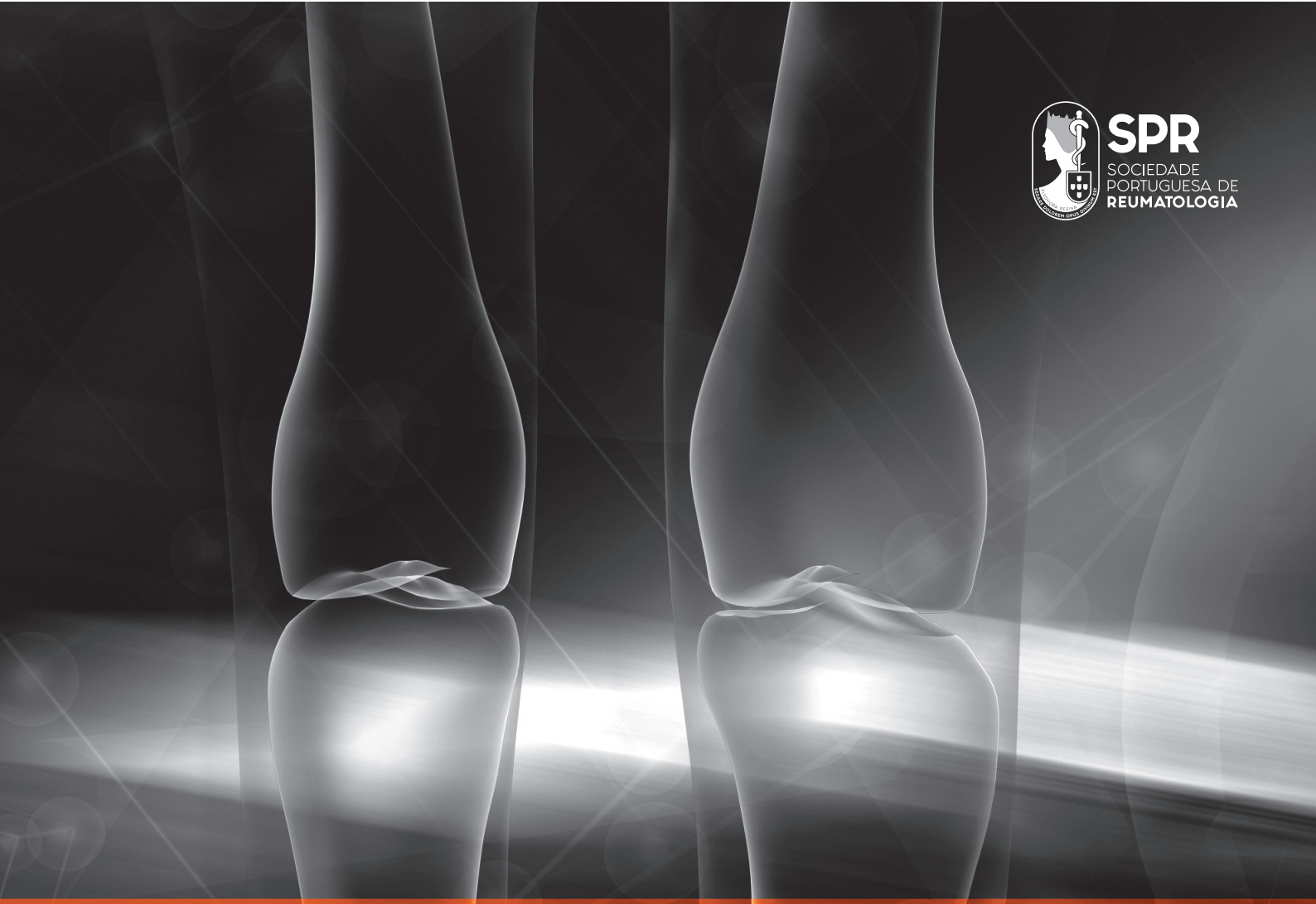

SPR
SOCIEDADE PORTUGUESA DE REUMATOLOGIA

ACTA REUMATOLÓGICA PORTUGUESA

VOL. 45 – N.º 4 | OUTUBRO/DEZEMBRO 2020

XXII

CONGRESSO PORTUGUÊS DE REUMATOLOGIA

ACTA REUMATOLÓGICA PORTUGUESA

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BOAS-VINDAS

Caros Colegas e Amigos

Nestes tempos estranhos, em que um pequeno vírus pode alterar a vida das pessoas à escala planetária, ficou provado que a qualidade científica e a formação médica é que fez e fará a diferença, entre quem tem a verdadeira capacidade de ajudar os outros. São tempos difíceis, desafiantes mas profundamente entusiasmantes para quem decidiu ser médico, especialmente para um Reumatologista. Estes momentos definem a inovação, a adaptabilidade, a qualidade mas, sobretudo, o compromisso perante os novos e antigos doentes e os médicos que deles cuidam.

As sociedades científicas têm contribuído de forma decisiva, com o apoio leal e ético da indústria farmacêutica, para a excelência da formação médica pós graduada em Portugal, colocando-nos ao nível do que o melhor se faz em todo o mundo. Foi por causa desse desígnio e compromisso, que a Sociedade Portuguesa de Reumatologia resolveu encarar esta pandemia, não como um obstáculo intransponível em termos de formação médica, mas antes um desafio. Adaptámos a nossa comunicação, introduzimos o Journal Club da Reumatologia e repensámos o nosso Congresso Português de Reumatologia.

E é nesse novo CPR, um e-CPR, que vos convidamos a participarem e se envolverem.

Este congresso é também um congresso de balanço final de uma direção, que tinha como estratégia lançar a SPR no século XXI, apostando numa nova imagem, num blogue para doentes e aposta nas redes sociais, num novo site, com maior ligação digital aos doentes e sócios, numa revista (Acta Reumatológica Pportuguesa) só em inglês e totalmente digital, na reedição do boletim informativo da SPR em formato digital, em novas formas de formação como o e-learning para MGF, em conceitos inovadores como o Fórum Art & Treat, num Reuma.pt com ecrãs tácteis para doentes nos hospitais de dia, etc, etc, etc. Em todas estas iniciativas tivemos um inexcedível apoio e entusiasmo de todos os reumatologistas e internos e essas foram apostas ganhas por todos. Aos coordenadores dos grupos de trabalho, Reuma.pt, ESPER, editor da ARP Rheumatology, Provedoria dos internos, o nosso agradecimento pelo tanto que fizeram no apoio à direção.

Igualmente temos de agradecer à Indústria Farmacêutica, que ao longo destes dois anos e meio soube apoiar de forma clara e desafiante a Reumatologia Portuguesa, contribuindo para muitos dos projetos e atividades da SPR, nomeadamente este CPR. Ter parceiros de qualidade e com visão facilita, mas responsabiliza-nos a querer mais e a fazer mais em prol da Reumatologia, sobretudo tendo em conta os doentes que são o foco da nossa atividade conjunta.

A todos os Reumatologistas e Internos o nosso profundo agradecimento pelo enorme envolvimento e trabalho, nomeadamente na criação de dezenas de brochuras da SPR, no livro Reumatologia/Clínica Geral, ou ainda nos júris e na avaliação dos trabalhos. Finalmente, um agradecimento público especial a todos os elementos da direção que trabalharam, partilharam e participaram em todos estes projetos. Para além de excelentes profissionais, são um grupo de excelentes pessoas e de amigos, do qual tivemos o prazer de fazer parte.

Este e-CPR será sem dúvida mais um grande desafio para a Reumatologia Portuguesa. Sejam bem-vindos a um futuro diferente da formação médica em Reumatologia

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Presidente da Comissão Científica do XXI CPR

Maria João Salvador
Presidente da Comissão Organizadora do XXI CPR

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Programa

13 DE OUTUBRO – TERÇA-FEIRA

13:30 Sessão 1 – Comunicações Orais
Moderadores: Sandra Falcão; Filipa Ramos

15:30 Sessão de abertura – Luís Cunha Miranda

15.45 Sessão – COVID 19
Moderador: Filipe Araújo
Outcome of COVID-19 in patients with RMDs – Pedro Machado
Covid 19, ontem hoje e amanhã – Filipe Froes

16.45 Sessão – Uso off label de DMARDs.
Moderadores: Tiago Meirinhos, Maria João Salvador
DMARDs sintéticos e biológicos: uso off label em idade pediátrica – Sandra Sousa
DMARDs sintéticos e biológicos: uso off label em idade adulta – Renata Aguiar

17.30 SIMPÓSIO Amgen – Fraturas Osteoporóticas. Velhos problemas. Novas soluções.
Moderador: José Canas da Silva
Palestrantes: Ana Rodrigues e José António Pereira da Silva

18.15 Sessão – Atualizações em Reumatologia
Moderadores: Lígia Silva; Lúcia Costa
Behçet state of the art – Nikita Khmelinskii
Distúrbios do Sono e doenças reumáticas: como gerir? – Susana Sousa

14 DE OUTUBRO – QUARTA-FEIRA

13:30 Sessão 2 – Comunicações Orais
Moderadores: Pedro Gonçalves; Walter Castelão

16.00 Sessão – Qua há de novo?
Moderadores: Ana Roxo Ribeiro, Joaquim Polido Pereira
ANAs: À descoberta da nova nomenclatura – Esmeralda Neves
Controvérsias em Osteoporose – Ana Rodrigues

17.30 SIMPÓSIO Lilly – Baricitinib (Olumiant®) – Resultados a Longo Prazo e Prática Clínica

17:30-17:35 Abertura – Chairman: João Eurico da Fonseca

17:35-17:50 Baricitinib – Resultados a Longo Prazo Eficácia e Segurança – Augusto Faustino

17:50-18:05 RWE de Baricitinib e prática clínica – Blanca Hernández Cruz

18:05-18:15 Q&A e fecho – João Eurico da Fonseca

18:15 Sessão – Doenças Reumáticas e Músculo-esqueléticas Induzidas por Fármacos
Moderadores: Marília Rodrigues; Anabela Barcelos
Doenças Reumáticas e Músculo-esqueléticas Induzidas por Fármacos – Vasco Romão

19:00 SIMPÓSIO Novartis – Joints + | A abordagem completa à Artrite Psoriática
Moderador: José Costa
O tratamento completo da AP... – Helena Santos
... Durante mais tempo – Patricia Nero

15 DE OUTUBRO – QUINTA-FEIRA

13:30 Sessão 3 – Comunicações Orais
Moderadores: Elsa Sousa; Alexandre Sepriano

17:15 SIMPÓSIO Pfizer – Practicalities of Tofacitinib Treatment
Chair: Cátia Duarte

17:15 Welcome and introduction
Chair:

17:20 How to use Tofacitinib in everyday practice
Joaquim Polido

17:35 Real World Data
Paul Hasler

17:55 Panel Discussion
Chair

18:00 Symposium close
Chair

18:00 Sessão – Gestão do doente em remissão
Moderadores: Graça Sequeira; Fernando Saraiva
AR – Cátia Duarte
SpA – Pedro Carvalho
AIJ – Daniela Peixoto
LES – Diogo Jesus

19:15 SIMPÓSIO MSD – Leading innovation. Changing lives.

19:15-19:20 Boas vindas – Elsa Vieira de Sousa, Sofia Ramiro

19:20-19:35 Espondilartrite axial: Ferramentas de avaliação e efetividade de tratamento anti-TNF – Sofia Ramiro

19:35-19:55 Dactilite: mais do que artrite – Elsa Vieira de Sousa

19:55-20:00 Conclusões – Sofia Ramiro, Elsa Vieira de Sousa

16 DE OUTUBRO – SEXTA-FEIRA

13:30 Sessão 4 – Comunicações Orais
Moderadores: José Bravo Pimentão; Rita Barros

16:00 APPSReuma – Consultas à distância – o que precisamos saber
Moderadores: Andréa Marques; Ricardo Ferreira
O que precisamos de saber para fazer uma teleconsulta? – Georgina Pimentel
Expressividade e emoções em vídeo (e telefone): dicas de uma expert – Mariana Figueira da Silva
Que ferramentas tecnológicas podemos recomendar aos doentes para promover a sua autogestão? – Elsa Mateus

16:45 Sessão – Dor & Inflamação:
Moderadores: Vera Las, António Vilar
AINEs – Armando Malcata
Opióides – Beatriz Craveiro Lopes

17:30 SIMPÓSIO Abbvie – Achieving sustained remission in RA: Upadacitinib, Not just another JAK in the pot
Dr. Luis Miranda
Dr. Tiago Meirinhos

18:15 Conferência plenária – Mabs e Nibs em 2025
João Eurico Fonseca

19:00 Encerramento do congresso
Entrega de prémios

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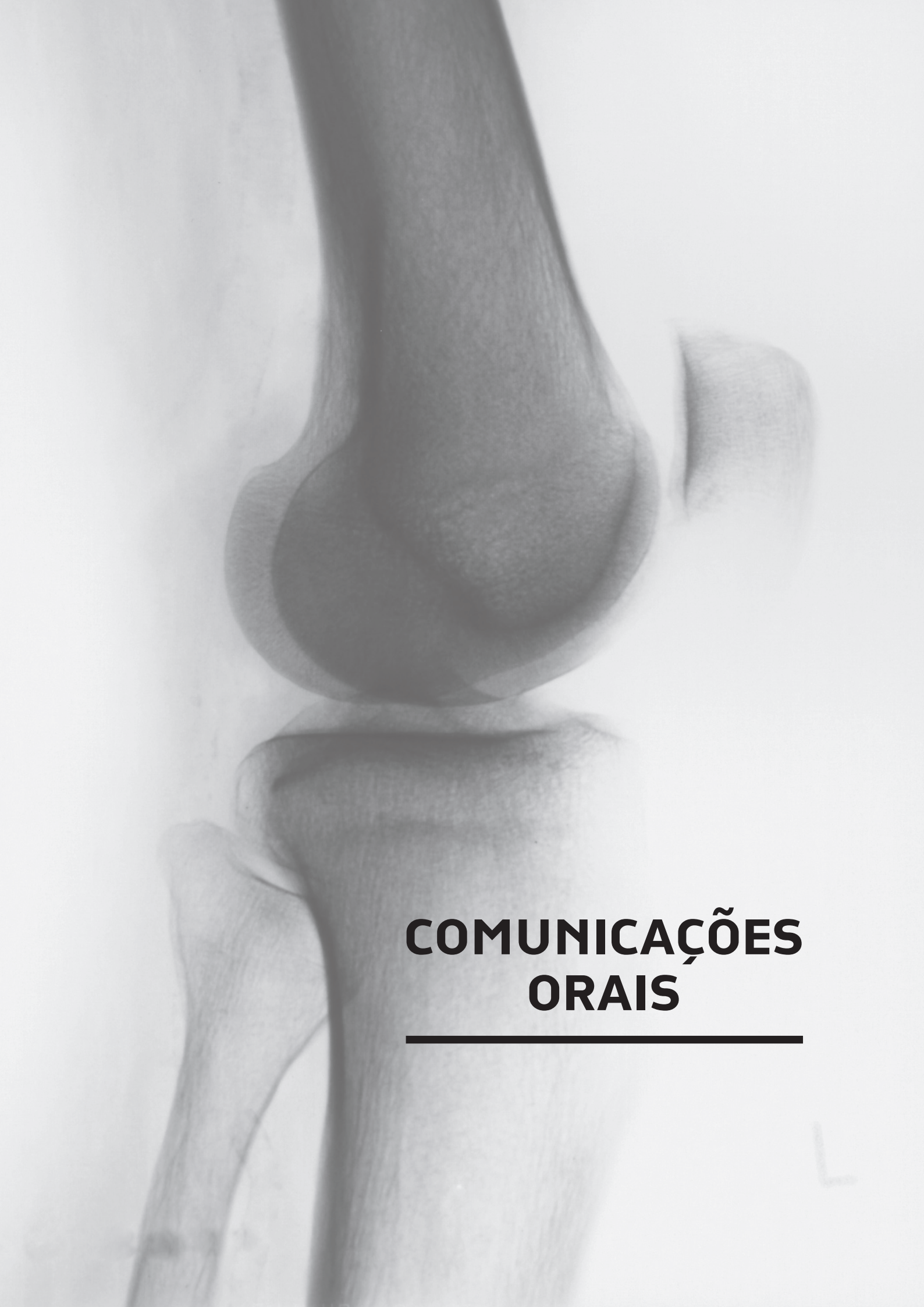

COMUNICAÇÕES ORAIS

Comunicações Orais

C0002 – PREDICTORS OF EARLY MORTALITY FOR GIANT CELL ARTERITIS AT THE TIME OF DIAGNOSIS

Eduardo Dourado[1, 2], Sofia C Barreira[1, 2], Ana Rita Cruz-Machado[1, 2], Martins-Martinho J[1, 2], Diana Raimundo[3], Luisa Brites[4], Helena Assunção[4], Vítor Teixeira[5], Nikita Khmelinskii[1, 2], Carla Macieira[1, 2], JAP da Silva[4], JE Fonseca[1, 2], Cristina Ponte[1, 2]

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Background: Giant cell arteritis (GCA) is the most common form of primary systemic vasculitis, typically affecting patients aged > 50 years. If left untreated, GCA can lead to permanent visual loss and other ischaemic complications. During the first two years of diagnosis, mortality is significantly greater in GCA than in the general population, with a significant contribution of infections to mortality in the first year of treatment. Identifying patients with a higher risk of mortality at the time of diagnosis could be crucial for prevention and tailored treatment; however, independent predictors of early mortality have never been reported in the literature.

Objective: To determine independent predictors of early mortality for GCA at the time of diagnosis.

Methods: Bicentric observational study using data from the Portuguese Register of Rheumatic Diseases (Reuma.pt) and hospital clinical records. Patients with biopsy-proven GCA or with the presence of "halo sign" on ultrasound were included. Early mortality was defined as death occurring in the first two years after diagnosis. Univariate analysis was performed using Chi-Square, Fischer's Exact Test and Mann-Whitney Test, as appropriate. Multivariate analysis was performed using binary logistic regression modelling. The linearity of the continuous variables with respect to the logit of the dependent variable was assessed via the Box-Tidwell procedure. Correlated variables, cases with missing information and outliers were excluded from the multivariate analysis in order to fulfil all assumptions

TABLE 1 – RESULTS OF THE UNIVARIATE ANALYSIS TESTING THE ASSOCIATION BETWEEN THE OUTCOME VARIABLE (EARLY MORTALITY) AND PREDICTOR VARIABLES

Variables	Early mortality (n=14)	Early survival (n=119)	p-value
Demographic and anthropometric data at diagnosis			
Age, median (IQR) in years	**83 (10)**	**74 (13)**	**0.001**
Female sex, n/mN (%)	7/14 (50)	81/117 (69)	0.226
Weigh, median (IQR) in kg	58 (13)	61 (13)	0.253
Height, median (IQR) in cm	157 (12)	153 (13)	0.486
Body mass index, median (IQR) in kg/m^2	24 (9)	26 (6)	0.426
Diagnostic delay, median (IQR) in days	38 (99)	34 (110)	0.456
Patient habits at diagnosis, n/mN (%)			
Smoking habits (current or previous smoker)	1/11 (9)	19/95 (20)	0.623
Drinking habits (current or previous drinking habits)	1/12 (8)	12/88 (14)	0.490
Disease manifestations at diagnosis, n/mN (%)			
Large vessel involvement documented by imaging	0/14 (0)	13/104 (13)	0.359
New-onset headache	11/14 (79)	100/117 (85)	0.449
Pain or altered pulse at the temporal arteries	8/11 (73)	59/93 (63)	0.742
Scalp hypersensitivity	2/14 (14)	25/117 (21)	0.733
Jaw claudication	9/14 (64)	50/117 (43)	0.126
Tongue claudication	**3/14 (21)**	**5/117 (4)**	**0.040**
PMR-like symptoms	6/14 (43)	54/117 (46)	0.851
Fever	1/14 (7)	13/117 (11)	1.000
Weigh loss	7/14 (50)	46/117 (39)	0.441
Fatigue	7/14 (59)	54/117 (46)	0.785
Cranial ischaemic event	**11/14 (79)**	**54/117 (46)**	**0.022**
Anterior ischaemic optic neuropathy	**9/14 (64)**	**33/117 (28)**	**0.012**
Central retinal artery thrombosis	1/14 (7)	7/117 (6)	1.000
Permanent loss of vision	**11/14 (79)**	**30/117 (26)**	**<0.001**
Transient loss of vision	0/14 (0)	14/117 (12)	0.361
Diplopia	1/14 (7)	6/117 (5)	0.568
Ischaemic transient attack	1/14 (7)	3/117 (3)	0.367
Cerebral vascular accident	2/14 (14)	7/117 (6)	0.247
Laboratory findings at diagnosis, median (IQR)			
Erythrocyte sedimentation rate, mm/h	85 (51)	82 (43)	0.552
C-reactive protein, mg/dL	4.2 (4.8)	5.0 (6.6)	0.738
Haemoglobin, mg/dL	10.4 (1.8)	11.7 (2.3)	0.080
Leucocyte count, x10^9	9.4 (4.3)	9.8 (3.8)	0.739
Creatinine level, mg/dL	**1.1 (0.6)**	**0.8 (0.3)**	**0.002**
Imaging and biopsy findings at diagnosis, n/mN (%)			
Positive temporal artery biopsy	4/6 (67)	37/58 (64)	1.000
Positive ultrasound	13/13 (100)	90/106 (85)	0.211
Comorbidities at diagnosis, n/mN (%)			
At least one comorbidity	14/14 (100)	115/117 (98)	1.000
Obesity	0/10 (0)	20/107 (17)	0.208
Arterial hypertension	11/14 (79)	79/116 (68)	0.548
Hypercholesterolemia	9/14 (64)	43/116 (37)	0.080
Hypertriglyceridemia	1/14 (7)	9/116 (8)	1.000
Hyperuricemia	3/14 (21)	12/115 (10)	0.209
Diabetes mellitus	5/14 (36)	39/116 (34)	1.000
Ischaemic heart disease	2/14 (14)	15/115 (13)	1.000
Cerebrovascular disease	3/14 (21)	14/115 (12)	0.396
Atrial fibrillation	**6/14 (43)**	**9/115 (8)**	**0.002**
Carotid atherosclerosis	1/14 (7)	9/116 (8)	1.000
Arterial peripheral disease	2/14 (14)	8/115 (7)	0.297
Chronic kidney disease	**6/14 (43)**	**13/115 (11)**	**0.007**
Thyroid disease	1/14 (7)	1/114 (1)	1.000
Treatments at diagnosis			
Glucocorticoid pulses, n/mN (%)	8/14 (57)	39/109 (36)	0.121
Prednisolone initial dose, median (IQR) in mg/day	60 (10)	60 (20)	0.380
NSAID's, n/mN (%)	0/14 (0)	9/115 (8)	0.596
Colchicine, n/mN (%)	0/14 (0)	4/115 (3)	1.000
Hypouricemic therapy, n/mN (%)	1/14 (7)	11/115 (10)	1.000
Bisphosphonates, n/mN (%)	**2/10 (20)**	**63/116 (54)**	**0.049**
Anticoagulants, n/mN (%)	**5/14 (36)**	**8/106 (8)**	**0.006**
Antiplatelet therapy, n/mN (%)	10/14 (71)	88/116 (76)	0.791
Statins, n/mN (%)	6/14 (43)	66/115 (57)	0.026
ACE inhibitors/ ARAs, n/mN (%)	8/14 (57)	65/115 (57)	0.965
Beta-blockers, n/mN (%)	6/14 (43)	32/115 (28)	0.244
Oral antidiabetics, n/mN (%)	5/14 (36)	32/115 (28)	0.542
Insulin, n/mN (%)	4/14 (29)	13/115 (11)	0.090

ACE – angiotensin-converting enzyme; ARAs – angiotensin II receptor antagonist; IQR – interquartile range; mN – modified N (= total N – missing data); NSAID's – non-steroidal anti-inflammatory drugs; PMR – polymyalgia rheumatica

necessary to assure the validity of the regression.

Results: The study included 133 patients with 85 (66.4%) females and a median age at diagnosis of 75.0 (interquartile range [IQR] 12.0) years. Fourteen (10.5%) deaths occurred during the first two years after diagnosis. Early mortality was significantly associated with: (i) cranial ischaemic event, anterior ischaemic optic neuropathy, permanent loss of vision and tongue claudication at disease presentation; (ii) older age, atrial fibrillation, chronic kidney disease, creatinine level and treatment with bisphosphonates at diagnosis; and (iii) treatment with anticoagulants before disease onset (Table 1). The multivariate analysis included 124 patients (5 patients had missing information, 4 patients were outliers) with 81 (65.3%) females, a median age at diagnosis of 75.0 (IQR 12.0) years, and 10 (8.1%) deaths in the first two years of diagnosis. The logistic regression model was statistically significant, $\chi 2$ (7) = 42.0, p<0.001. The model explained 66.9% (Nagelkerke R2) of the variance in early mortality and correctly classified 96.0% of all cases. Older age at diagnosis (OR 1.3/year, 95%CI: 1.0-1.6, p=0.032), tongue claudication at disease presentation (OR 2106.8, 95%CI: 4.2-1057334.5, p=0.016), previous treatment with anticoagulants (OR 42.1, 95%CI: 2.6-682.0, p=0.009) and treatment with bisphosphonates at diagnosis (OR 0.0, 95%CI: 0.0-0.4, p=0.019) were identified as independent predictors of early mortality and survival, respectively (Figure 1).

Conclusions: In our cohort, older age at diagnosis, tongue claudication at disease presentation and previous treatment with anticoagulants were independent predictors of early mortality. On the other hand, treatment with bisphosphonates at diagnosis was an independent predictor of early survival. These findings are novel and require replication. However, they highlight the need for a disease management not only focused on clinical manifestations but also on drug adverse effects and comorbidities.

FIGURE 1 – LOGISTIC REGRESSION PREDICTING THE LIKELIHOOD OF EARLY MORTALITY FOR GCA, BASED ON AGE, SEX, CRANIAL ISCHAEMIC EVENT, TONGUE CLAUDICATION, CHRONIC KIDNEY DISEASE AND TREATMENT WITH ANTICOAGULANTS AND BISPHOSPHONATES (AT THE TIME OF DIAGNOSIS)

Variables	*B*	SE	Wald	*df*	*p*	OR	95% CI for OR	
							Inferior	Superior
Age at diagnosis	0.257	0.120	4.583	1	**0.032**	1.293	1.022	1.636
Sex	3.155	1.867	2.856	1	0.091	23.456	0.604	910.522
Anticoagulant	3.739	1.422	6.918	1	**0.009**	42.051	2.593	681.981
Bisphosphonate	-5.915	2.518	5.520	1	**0.019**	0.003	0.000	0.375
Cranial ischaemic event	0.286	1.283	0.050	1	0.824	1.331	0.108	16.463
Tongue claudication	7.653	3.173	5.818	1	**0.016**	2106.788	4.198	1057334.541
Chronic kidney disease	0.015	1.121	0.000	1	0.989	1.015	0.113	9.131
Constant	-26.049	11.337	5.279	1	0.022	0.000		

B – coefficients in log-odds units; CI – confidence interval; *df* – degrees of freedom; OR – odds ratio, p – Wald 2-tailed p-value; SE – standard errors associated with the coefficients; Wald – Wald chi-square value.

C0015 – PORTUGUESE MULTIDISCIPLINARY RECOMMENDATIONS FOR NON-PHARMACOLOGICAL AND NON-SURGICAL INTERVENTIONS IN PEOPLE WITH RHEUMATOID ARTHRITIS

Eduardo Santos[1, 2, 3], Cátia Duarte[1, 4], Ricardo J. O. Ferreira[1, 3], JE Fonseca[5, 6], José António Canas da Silva[7], Helena Canhão[8, 9], Carlos Vaz[10], Catarina Aguiar Branco[11], Inês Campos[12], Elsa Mateus[13], Pinto, A.M.[14, 15], Iolanda Barbosa[16], Rui Costa[17], Ana Moreira[18], Maria Helena Mourão[19], JAP da Silva[1, 4]

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Background: People with rheumatoid arthritis (RA) report significant levels of disease impact. These are improved, but not fully abrogated, by immunosuppressive therapy, even when remission is achieved, thus imposing the need for adjuvant interventions targeting the uncontrolled domains of disease impact. Non-pharmacological are widely used to this purpose, but they have not been the object of professional recommendations or guidelines.

TABLE 1 – RECOMMENDATIONS FOR NON-PHARMACOLOGICAL AND NON-SURGICAL INTERVENTIONS IN PEOPLE WITH RHEUMATOID ARTHRITIS

	LoE*	GoR†	LoA (0-10)
Overarching principles			
A. The primary goal of non-pharmacological and non-surgical interventions in RA is to assist patients in maximizing their overall quality and enjoyment of life, through optimized control of the impact of disease, besides and beyond medications and surgery.	-	-	9.9 (0.3)
B. Patients must be given a decisive role in establishing the objectives and the nature of interventions in their particular case, in an informed and shared decision-making process.	-	-	9.7 (0.6)
C. The healthcare team must make sure that the patient has all the information deemed relevant to support his/her participation in self-care and in shared-decision making, including the short and long-term outlook of the disease and of the available scope of treatments and interventions.	-	-	9.8 (0.5)
D. Non-pharmacological and non-surgical interventions demand the involvement of a multiprofessional/ multidisciplinary team, and the adoption of an holistic bio-psycho-social model.	-	-	9.8 (0.5)
E. Non-pharmacological and non-surgical interventions should be selected on the basis of the available evidence and adjusted to the patient's specific clinical features, abilities, preferences and needs.	-	-	9.9 (0.5)
Recommendations			
1. Non-pharmacological and non-surgical interventions should be an integral part of standard care in people with RA and should be considered throughout the course of the disease, whenever they may provide relevant, objective or subjective, benefit to the patient, as adjuvant or as an alternative to symptomatic medication or surgery.	1a	A	9.6 (0.7)
2. Areas of intervention to be considered in this context include, but are not limited to: self-management, pain relief, energy management, joint alignment and support, thermotherapy, exercise, hydrokinesiotherapy, psychological interventions, daily, leisure and work activities, education, family involvement, social participation and social care, sleep hygiene and general management of comorbidities.	5	D	9.2 (1.3)
3. Dedicated educational programs and supporting materials should be made available to patients, providing information on the general aspects of the disease and its management, their role in the shared-care process and the specificities of common assessments and interventions.	1a	B	9.8 (0.6)
4. Patients should be regularly inquired for unmet needs, through the use of validated large-scope instruments, and referenced to the most appropriate health professional(s) in the team.	2b	B	9.1 (1.4)
5. The multiprofessional team must try to make interventions as attractive and relevant as possible to patients, as a mean to ensure the best possible adherence and long-term effectiveness.	2b	B	9.8 (0.5)
6. Interventions should be targeted to specific objectives that are relevant to the individual patient objectives, regularly monitored with validated instruments and adapted accordingly.	3	C	9.6 (0.7)
7. All patients should be stimulated to follow a personalized and regular physical exercise program to reduce pain, functional disability, fatigue and global impact of disease.	1a	B	9.7 (0.7)
8. Hydrokinesiotherapy should be considered, to reduce pain and global impact of disease.	2b	B	9.2 (1.0)
9. Orthoses should be considered as a mean to reduce joint pain, functional disability and global impact of disease.	1a	B	9.1 (1.1)
10. Psychosocial interventions should be considered as a mean to reduce pain, functional disability, fatigue and global impact of disease.	1a	B	9.6 (0.9)

These recommendations should be interpreted in the light of the clarifications provided in the body of the text and by the supporting SLR.

* 1a: systematic review of RCTs; 1b: individual RCT; 2a: systematic review of cohort studies; 2b: individual cohort study (including low-quality RCT; eg,<80% follow-up); 3a: systematic review of case-control studies; 3b: individual case-control study; 4: case-series (and poor quality cohort and case-control studies); 5: expert opinion without explicit critical appraisal, or based on physiology, bench research or 'first principles'.
† A: based on consistent level 1 evidence; B: based on consistent level 2 or 3 evidence or extrapolations from level 1 evidence; C: based on level 4 evidence or extrapolations from level 2 or 3 evidence; D: based on level 5 evidence or on troublingly inconsistent or inconclusive studies of any level.
LoE, level of evidence; GoR, grade of recommendation; LoA, level of agreement.

Objective: To propose multidisciplinary recommendations to inform clinical practice regarding the employment of non-pharmacological and non-surgical interventions in the management of patients with RA.
Methods: The EULAR standardized operating procedures for the development of recommendations were followed. First, a systematic literature review was performed. Then, a multidisciplinary Technical Expert Panel (TEP) including rheumatologists, physiatrists, allied health professionals and patient representatives, met to develop and discuss the recommendations and research agenda. For each developed recommendation i) the level of evidence and grade of recommendation were determined, and ii) the level of agreement among TEP members was set by an anonymous online survey using a numeric rating scale (NRS). The mean and range of the level of agreement for each recommendation was calculated. A recommendation was adopted if approved by ≥75% of the TEP members, and the level of agreement was considered high when NRS ≥8. All relevant national societies were included in this construction process to attain their endorsement.
Results: Based on evidence and expert opinion, the TEP developed and agreed on five overarching principles and 12 recommendations for non-pharmacological and non-surgical interventions in people with RA (table 1). The mean level of agreement between the TEP members ranged between 8.5 and 9.9.
Conclusions: These recommendations are based on the consensus judgment of clinical experts from a wide range of disciplines and patients' representatives from Portugal. Given the evidence for effectiveness, feasibility and safety, non-pharmacological and non-surgical interventions should be an integral part of standard care for people with RA. It is hoped that these recommendations should be widely implemented in clinical practice. The target audience for these recommendations includes all health professional involved in the care of patients with RA. The target patient population includes adult Portuguese people with RA.

C0024 – PREDICTIVE FACTORS OF RELAPSE AFTER METHOTREXTE DISCONTINUATION IN JIA PATIENTS WITH INACTIVE DISEASE

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Background: Methotrexate (MTX) is the most widely used first-line conventional synthetic disease-modifying antirheumatic drug (csDMARD) in the treatment of juvenile idiopathic arthritis (JIA).1,2 When remission is achieved, questions remain about discontinuing MTX. There is some evidence that a longer period of inactive disease before MTX withdrawal is associated with lower likelihood of relapse, while both rheumatoid factor (RF) positive polyarthritis and extended oligoarthritis categories are associated with higher probability of disease relapse.2,3

Objective: To identify predictive factors of relapse after discontinuation of MTX in JIA patients with inactive disease.

Methods: Prospective multicentre cohort study in patients diagnosed with JIA using real world data from the Portuguese national register database, Reuma.pt (Figure 1).4 We evaluated patients with JIA, according to the ILAR classification, who have reached JADAS27 inactive disease (JADAS 27 ≤1 and no active extra-articular manifestations) and discontinued MTX before the age of 18 years-old.5 Relapse was defined as recurrence (JADAS 27 >1 and/or extra-articular manifestations) or restarting a DMARD.5 To identify differences of relapse risk, univariate analyses were performed. Persistence in remission was estimated using the Kaplan-Meier method and groups compared with log-rank tests. Subsequently, Cox regression analyses were performed to identify predictors of relapse.

Results: 119 JIA patients discontinued MTX due to inactive disease (Figure 1). The majority (69.7%) were females and 60.6% had oligoarticular JIA. Sociodemographic and clinical characteristics are shown in Table 1. Relapse has occurred in 32.8%. Table 2 shows the disease characteristics at MTX initiation and discontinuation and at relapse or last visit.

In univariate analysis, relapse was associated with the use of NSAIDs at the time of MTX discontinuation (p=.027) and with a period of less than two years in inactive disease before MTX suspension (p=.040). We found no association with gender, race, immunology (RF, antinuclear and cyclic citrullinated peptide antibodies), MTX dose, discontinuation modality (tapering and spacing the doses or just tapering the dose), extra-articular manifestations, previous corticotherapy, family history, body mass index, JADAS, CHAQ index, inflammatory parameters, tender and swollen joint counts at MTX initiation or discontinuation nor with age at remission or at MTX suspension. Median persistence in inactive disease was significantly higher in patients with more than two years in remission before MTX discontinuation (p=.034) and in those who did not use NSAIDs at time of MTX discontinuation (p=.026) (Figure 2).

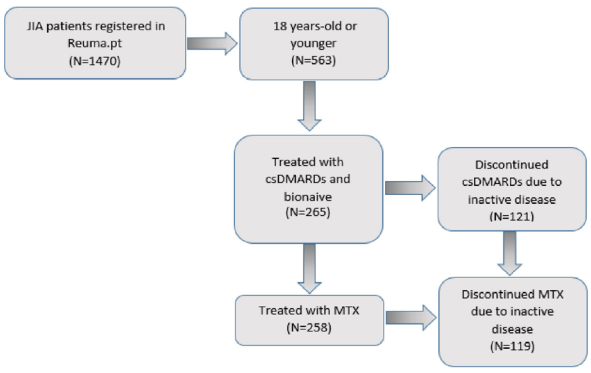
After adjustment for age at diagnosis, MTX tapering and JIA category, use of NSAIDs at the time of discontinuation (HR, 1.98 95%CI 1.03-3.82) and less than

TABLE 1: SOCIODEMOGRAPHIC AND CLINICAL CHARACTERISTICS OF THE STUDY POPULATION

Age at diagnosis, years (median (IQR))	6.23 (7.56)
Age at disease onset, years (median (IQR))	5.79 (7.82)
Gender %(n/N)	Female: 69.7% (83/119)
JIA categories %(n/N):	
Persistent oligoarthritis	47.1% (56/119)
Extended oligoarthritis	13.5% (16/119)
Systemic JIA	5.0% (6/119)
RF-negative polyarthritis	16.8% (20/119)
RF-positive polyarthritis	5.0% (6/119)
Psoriatic arthritis	5.9% (7/119)
Enthesitis-related arthritis	4.2% (5/119)
Undifferentiated arthritis	2.5% (3/119)
Race %(n/N):	
White of European origin	89.1% (106/119)
Other	10.9% (13/119)
Years from disease onset until DMARD initiation (median (IQR))	0.79 (1.55)
ANA POSITIVE %(n/N)	58.7% (61/104)
HLA-B27 positive %(n/N)	14.3% (9/63)
RF positive %(n/N)	7.6% (9/105)
ACPA positive %(n/N)	11.3% (6/53)
Family history of rheumatic diseases %(n/N)	10.1% (12/119)
Presence of extra-articular manifestations %(n/N)	28.6% (34/119)
Uveitis %(n/N)	41.2% (14/34)

ANA: antinuclear antibody; ACPA: anti cyclic citrullinated peptide antibody; HLA: human leucocyte antigen; IQR: Interquartile range; JIA: juvenile idiopathic arthritis; RF: rheumatoid factor

FIGURE 1: PATIENTE FLOWCHART



csDMARDs: conventional synthetic disease-modifying antiheumatic drugs; MTX: Methotrexate.

two years in remission (HR, 3.12 95%CI 1.35-7.13) remained associated with relapse.

Conclusions: In this large cohort we found that the use of NSAIDs at the time of MTX discontinuation was associated with two times the likelihood of relapse. Like in other studies we also showed that the time in remission before MTX discontinuation is the main predictor of relapse. We found no association between the JIA category and the risk of relapse.

C0032 – RISK OF CKD IN MEMBRANOUS AND PROLIFERATIVE LUPUS NEPHRITIS – ANALYSIS OF A NATIONWIDE MULTICENTRE COHORT

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Background: Lupus nephritis (LN) is one of the most severe manifestations of Systemic Lupus Erythematosus.

Objectives: 1) To compare proliferative (PLN), membranous (MLN) and mixed LN regarding clinical and laboratory presentation. 2) To investigate predictors of progression to chronic kidney disease (CKD).

Methods: Multicentre observational study, with retrospective analysis of a prospective cohort, using data from the Portuguese registry of rheumatic diseases – Reuma.pt. Patients with biopsy-proven PLN, MLN and mixed LN were included. Groups were compared using Pearson's Chi-Square for categorical variables and One-Way ANOVA or Kruskal-Wallis for numerical variables. COX regression analysis was used to investigate predictors of CKD (defined as estimated glomerular filtration rate [eGFR] lower than 60

TABLE 1: COMPARATIVE DESCRIPTION OF THE REUMA.PT COHORT OF PATIENTS WITH PROLIFERATIVE, MEMBRANOUS AND MIXED LN

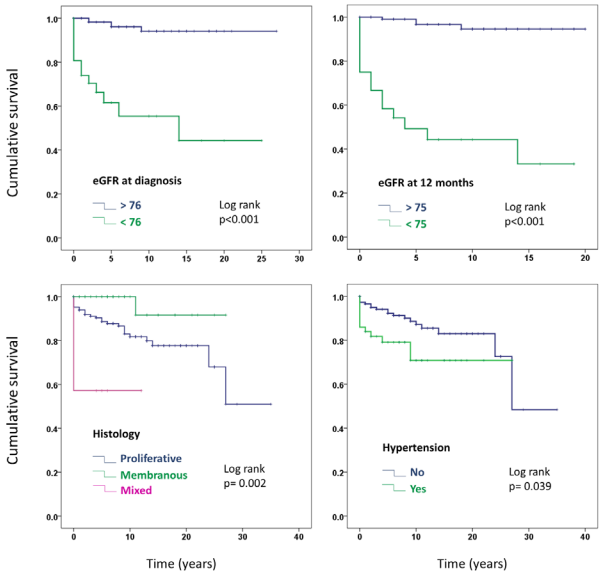
	PLN	MLN	Mixed	P
Total, N	186	42	8	
Females, N (%)	157 (85)	39 (95)	4 (50)	**0.004**
Ethnicity **White European**, N (%)	163 (90)	31 (78)	7 (88)	0.115
Other, N (%)	19 (10)	9 (23)	1 (13)	
Age LN diagnosis(y), median (IQR)	30 (20)	34 (16)	42 (25)	0.409
SLEDAI at LN diagnosis, median (IQR)	**16 (9)**	**10 (10)**	**21 (17)**	**0.006***
uPCR at LN diagnosis, median (IQR)	1675 (2598)	1698 (2153)	2160 (3320)	0.629
Creatinine at LN diagnosis, median (IQR)	**0.80 (0.32)**	**0.70 (0.20)**	**1.00 (0.95)**	**0.006***
eGFR at LN diagnosis, mean ± SD	**98 ± 33**	**112 ± 17**	**82 ± 45**	**0.019***
Albumin at LN diagnosis, mean ± SD	34 ± 7	34 ± 7	30 ± 6	0.390
C3 at LN diagnosis, mean ± SD	**0.65 ± 0.26**	**0.90 ± 0.35**	**0.53 ± 0.30**	**<0.001***
Positive anti-dsDNA LN diagnosis, N (%)	**115 (91)**	**11 (48)**	**6 (86)**	**<0.001***
Use of antimalarials, N (%)	166 (94)	36 (92)	8 (100)	0.688
Use of immunosuppressants, N (%)	163 (94)	33 (87)	8 (100)	0.245
Use of corticosteroids, N (%)	145 (84)	33 (85)	7 (100)	0.511
CKD after LN diagnosis, N (%)	**27 (15)**	**1 (3)**	**3 (38)**	**0.018***
ESRD, N (%)	**7 (4)**	**1 (3)**	**2 (25)**	**0.016**
Deaths, N (%)	14 (8)	2 (5)	0	0.610

uPCR: urinary protein-creatinine ratio, mg/g; y: years; Creatinine presented in mg/dL, eGFR in mL/min/1.73m2, albumin in g/L and C3 in g/L

Note: Baseline data (LN diagnosis) in grey; other data refer to the course of disease

*Significant difference between the proliferative and membranous groups

FIGURE 1: KAPLAN-MEIR CURVES SHOWING CUMULATIVE SURVIVAL FREE OF CKD IN PATIENTS WITH PLN, MLN AND MIXED LN



mL/min/1.73m2 for at least 3 months) and Kaplan-Meier curves were drawn.
Results: 236 patients were included. Median follow-up was 8 years (IQR 11; maximum 35 years). As seen in table 1, the level of proteinuria did not differ between groups; however, MLN patients presented with significantly lower serum creatinine. Levels of complement C3 and C4 were reduced in PLN but normal in MLN patients, and there were fewer patients with positive anti-dsDNA antibodies in the MLN group (p<0.001). On univariable COX regression, mixed histology was associated with progression to CKD (HR 26 [95% CI 3 – 255], p 0.005) (figure 1), however, it lost significance after adjusting for eGFR. In fact, eGFR≤75 at one year after the renal biopsy (HR 21 [95% CI 7 – 65], p<0.001) was the strongest predictor of CKD, even after adjusting for hypertension or histology.
Conclusions: Our results support previous findings from single-centre studies suggesting that MLN has a different serological profile than PLN, possibly reflecting different pathogenesis. Renal function at one year predicts long-term outcome in LN.

C0050 – CLINICAL DISEASE ACTIVITY, MRI SPINAL INFLAMMATION AND ENTHESITIS ARE KEY DETERMINANTS OF IMPAIRMENT OF SPINAL MOBILITY IN EARLY AXIAL SPONDYLOARTHRITIS – DATA FROM THE DESIR COHORT

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Background: It has been shown that spinal mobility impairment in axial spondyloarthritis (axSpA) is independently determined both by irreversible spinal damage and by reversible spinal inflammation. However, these relationships have only been investigated in patients with longstanding disease (ankylosing spondylitis). Moreover, only the composite score Bath Ankylosing Spondylitis Metrology Index (BASMI) has been evaluated rather than individual mobility assessments.
Objectives: Our aim was to investigate the determinants of spinal mobility in patients with early axSpA.
Methods: We analysed longitudinal data from the DEvenir des Spondyloarthrites Indifférenciées Récentes (DESIR) cohort, collected during the first five years of follow-up. We selected patients with a definite diagnosis of axSpA according to the treating rheumatologist, at the end of follow-up (month 60). Associations were tested using generalised estimating equations (GEE), a multilevel approach that adjusts for within-patient correlation. The Bath Ankylosing Spondylitis Metrology Index (BASMI) or the individual components of BASMI (lateral spinal flexion, tragus-to-wall distance, cervical rotation, anterior lumbar flexion, maximal intermalleolar distance) were used as dependent variables, and clinical and demographic variables were used as independent variables in univariable models. Spinal MRI inflammation was assessed using the Berlin scoring system and radiographic structural damage was assessed using the modified Stoke ankylosing spondylitis spinal score (mSASSS)]. As physical function and quality of life are considered to be hierarchically superior to spinal mobility, they were not included in the analysis. Multivariable models were built, adjusting for potential confounding. Variables with a p-value <0.10 were re-tested in the multivariable models. Six models were

TABLE: MULTIVARIABLE GEE RESULTS (AB; 95% CI) PRESENTING INDEPENDENT ASSOCIATIONS BETWEEN BASMI (OR ITS COMPONENTS) AND CLINICAL AND DEMOGRAPHIC VARIABLES

	Mobility measures where higher values represent worse mobility		Mobility measures where higher values represent better mobility			
	BASMI	Tragus-to-wall distance	Lateral spinal flexion	Cervical rotation	Anterior lumbar flexion (modified Schober)	Maximal intermalleolar distance
Age	1.02 (1.01-1.03)		0.91 (0.87-0.96)	0.79 (0.69-0.90)		0.71 (0.61-0.82)
Male gender	#	2.35 (1.73-3.18)	5.01 (2.15 11.67)	#		171.35 (16.85-1742.22)
Education*	#	2.10 (1.06-4.16)	#	#		388.23 (32.09-4696.38)
BMI	0.97 (0.95-0.99)	1.04 (1.00-1.09)			1.08 (1.05-1.11)	
HLA-B27 positive	0.82 (0.70-0.97)		#	13.99 (1.09-180.20)		19.07 (1.56-233.62)
Symptoms' duration			0.76 (0.65-0.88)		1.07 (1.03-1.12)	1.72 (1.30-2.27)
Currently employed					#	
Current arthritis		#		0.01 (0.00-0.86)	#	
ASDAS-CRP	1.23 (1.15-1.32)	1.28 (1.07-1.52)	0.56 (0.39-0.79)	0.12 (0.04-0.38)	0.87 (0.79-0.97)	0.14 (0.08-0.25)
Enthesitis score	1.02 (1.01-1.04)		0.92 (0.86-1.00)	0.73 (0.59-0.89)		0.78 (0.67-0.89)
mSASSS	#	1.17 (0.90-1.51)**	0.77 (0.610-0.98)	0.42 (0.16-1.11)**	#	
MRI inflammation score	1.13 (1.05-1.23)	1.33 (1.08-1.64)	0.59 (0.44-0.80)	0.41 (0.17-0.95)	0.92 (0.86-0.98)	

#Statistically significant in the univariable models but excluded from the best-fit multivariable model; *Education at baseline (university or equivalent); **mSASSS not significant in the multivariable model but included as it was found to be a contributory variable improving model-fit.

built, one regarding the BASMI total score and five regarding the individual components of BASMI.

Results: Data from 644 patients and 5152 visits were analysed. In the multivariable analyses (table), we found an independent association between higher BASMI values and age [adjusted B (aB)=1.02, confidence interval (CI)=1.01-1.03], Ankylosing Spondylitis Disease Activity Score-C Reactive Protein (ASDAS-CRP) (aB=1.23, CI=1.15-1.32), enthesitis score (aB=1.02, CI=1.01-1.04) and MRI inflammation score (aB=1.13, CI=1.05-1.23). All individual BASMI components were independently associated with ASDAS-CRP. Apart from maximal intermalleolar distance, all other mobility measures were associated with MRI spinal inflammation. Lateral spinal flexion, cervical rotation and maximal intermalleolar distance were associated with the enthesitis score. mSASSS was associated with lateral spinal flexion and a contributory factor to tragus-to-wall distance and cervical rotation.

Conclusions: In early axSpA, spinal mobility impairment is independently determined by clinical disease activity, MRI spinal inflammation and the severity of enthesitis. Maximal intermalleolar distance (which is not a true measure of spinal mobility) was the only measure not associated with MRI spinal inflammation. The influence of spinal inflammation prevails in the early phase of axSpA while spinal damage becomes more relevant in later disease stages.

C0085 – POOR RESPONSE TO HEPATITIS B VACCINATION IN RHEUMATIC PATIENTS TREATED WITH BIOLOGIC THERAPY – IMPLICATIONS FOR CLINICAL PRACTICE

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Background: Hepatitis B virus (HBV) vaccination is recommended for rheumatic patients starting biologic therapy. There is some evidence that HBV vaccination is effective in patients under conventional disease modifying anti-rheumatic drugs (DMARDs), but it is currently unclear whether this also applies to biologics.

Objectives: To assess the efficacy and safety of HBV vaccination in patients with rheumatic diseases treated with biologics.

Methods: We included patients with any inflammatory rheumatic diseases treated with any biologic, who were negative for anti-HBs and anti-HBc and had never been vaccinated for HBV. Engerix B® was administered at 0, 1 and 6 months, and anti-HBs was re-assessed ≥1 month after last dose. Response was defined as anti-HBs>10IU/L and compared against healthy controls (HC) undergoing Occupational Health immunization. Disease flare was evaluated before and until at least one month post-vaccination. We recorded serious adverse events (SAE) and immune-related disorders not previously present.

Results: We included 67 patients, most treated with TNF inhibitors (TNFi), and 70 HC (Table 1). Most patients were taking concomitant DMARDs (69%) and were in remission/low disease activity (59%). Only 20/67 patients (30%) had a positive response to vaccination, in comparison to 68/70 HC (97%, p<0.001). Mean post-vaccination anti-HBs titre was significantly lower in responding patients than HC (569 ± 772 vs 1316 ± 811U/L, p<0.001). Responders diagnoses were rheumatoid arthritis (RA; n=8 [25%]), psoriatic arthritis (PsA; n=7 [39%]), ankylosing spondylitis (n=4 [33%]) and inflammatory bowel disease -associated spondyloarthritis (n=1 [100%]). Response was seen in 19/53 patients treated with TNFi (36%), but only in 1/14 (7%) of the patients treated with non-TNFi (p=0.037). Importantly, some responders had to temporarily interrupt biologic therapy due to other intercurrences for at least one administration. Mean age was slightly lower in HC, who were more frequently female, but no clinical or demographic variables were associated with vaccine response, including age, sex and disease duration/activity. Fourteen patients (21%) experienced disease flares: 7 were mild and did not require therapy adjustment; 3 patients required minor treatment/dose adjustments; and 4 patients had secondary failures that led to treatment switch. There were 3 SAE occurring 1-4 months after the 1st/2nd dose, deemed not to be related to vaccination: acute diverticulitis in a RA patient on golimumab; serious abdominal infection in a PsA patient on infliximab; atrial fibrillation and urinary infection in a PsA patient on infliximab. One RA patient with

TABLE 1 – BASELINE CHARACTERISTICS OF STUDY PARTICIPANTS

	Patients (n=67)	Controls (n=70)	p
Age, years (mean ± SD)	56 ± 9	46 ± 9	<0.001
Female, n (%)	40 (60)	62 (89)	<0.001
Diagnosis, n (%)			
Rheumatoid arthritis	32 (48)		
Psoriatic arthritis	18 (27)		
Ankylosing spondylitis	13 (19)		
Systemic lupus erythematosus	2 (3)		
IBD-related spondyloarthritis	1 (1)		
Adult-onset Still's disease	1 (1)		
Disease duration, years (mean ± SD)	17 ± 10		
Biologic, n (%)			
TNF-inhibitor	53 (79)		
Tocilizumab	6 (9)		
Rituximab	2 (3)		
Belimumab	4 (6)		
Abatacept	1 (1)		
Anakinra	1 (1)		
Conventional DMARDs, n (%)			
Methotrexate	39 (58)		
Sulfasalazine	6 (9)		
Leflunomide	1 (1)		
Hydroxychloroquine	2 (3)		
Azathioprine	1 (1)		
None	21 (31)		
Glucocorticoids, n (%)	29 (43)		
Prednisolone dose, mg (mean ± SD)	5.6 ± 2.1		
Disease activity (mean ± SD)			
DAS28	3.1 ± 1.4		
ASDAS	2.2 ± 1.4		
SLEDAI	6.0 ± 2.8		
Disease activity class, n (%)			
Remission	25 (40)		
Low	12 (19)		
Moderate	18 (29)		
High	4 (6)		
Very high	4 (6)		

infliximab had bilateral uveitis 2 months after the 1st vaccine dose, which resolved with topical therapy. There were no adverse events in HC.

Conclusions: In this study, HBV vaccination response in rheumatic patients treated with biologic therapy was poor and lower than in healthy adults. Vaccination was overall safe but there were 4 severe flares and 3 SAE that lead to treatment switch/interruption, although causal association is unlikely. Our data reinforce the recommendation for HBV vaccination prior to starting biologic therapy, possibly even as soon as the diagnosis is established. Alternative HBV vaccination strategies should be investigated in patients already treated with biologics.

C0105 – SYSTEMIC INFLAMMATION AND COGNITIVE DYSFUNCTION IN JSLE PATIENTS

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Background: Neurocognitive dysfunction (NCD) is one of the most commonly reported neuropsychiatric symptoms in patients with juvenile systemic lupus erythematosus (SLE), even without overt CNS disease. Signs of NCD are often subtle and difficult to ascertain in daily clinical practice, requiring formal neuropsychological testing (NPT). Ischemic and inflammatory mechanisms are key components of its immunopathogenesis, including abnormalities of the blood–brain barrier and autoantibody-mediated production of proinflammatory cytokines. Several studies have identified a possible role of autoantibody activity, cerebral ischemia, disease duration, disease activity, therapeutics, pro-inflammatory cytokines and behavioural factors. Some studies have suggested that through the use of serum inflammatory markers such as C-reactive protein (CRP), it is possible to predict small vessel vasculopathy.

Objective: To assess the association between serum inflammatory markers and cognitive dysfunction in juvenile-onset SLE (jSLE) patients.

Methods: A cross-sectional sample of jSLE patients, currently aged ≥ 16 years, completed a psychosocial assessment including the SF-36, HADS, SHS, BriefCope and MMSE questionnaires, between October 2018- May 2019. Local Ethics Committee approved the study. All patients fulfilled both 2012 and 2019 EULAR/ACR classification criteria for SLE. Juvenile-onset was defined as age at diagnosis <18 years. Demographics and clinical characteristics were collected. Statistical analysis was performed with SPSS®. Variables were compared with spearman correlations tests.

Results: 30 jSLE patients were included in the study, 90%female, with median (min-max) age of 21 (16-35) years, with mean (SD) age at diagnosis of 15.8 ± 2.1 years. Median CRP and ESR serum levels were 1.9 (0.1-9.6) mg/L and 19 (2-75) mm/H, respectively. Mean (SD) platelets counts, leucocytes counts and haemoglobin levels were 248x109/L (12.5 x109/L), 6.2 x109/L (0.4 x x109/L) and 13.2 (0.3) g/dL, respectively. Mean values (SD) of psychosocial assessment were: MMSE of 27.7 (1.8); HADS – Depression 3.9 (3.3), HADS – Anxiety 9 (4.3), SHS 5.2 (1.02); Physical health SF-36 of 66.8 (9.9) and Mental health SF-36 of 68.9 (17.5). 23.3% showed mild cognitive impairment, 63.3% anxiety and 13.3% depression. We observed significant inverse correlations between both serum CRP levels and platelets counts and MMSE scores (p=0.044, ρ=-0.377; p=0.044; ρ=-0.377, respectively).

Conclusion: Our findings suggest that higher CRP serum levels and platelets counts were associated with lower MMSE scores, meaning more NCD in these patients. Identification of high-risk subgroups for NPS involvement could lead to earlier diagnosis with NPT and targeted interventions, thus improving their prognosis.

CO136 – IDENTIFICATION OF ARTHRITIS BY THE REFERRING PHYSICIAN IS THE CRUCIAL FACTOR ASSOCIATED WITH SHORTER REFERRAL TO THE EARLY ARTHRITIS CLINIC

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Background: Early identification and referral of patients with inflammatory rheumatic diseases are essential to improve long-term clinical and radiographic outcomes. The Early Arthritis Clinic (EAC) was created in our department in 2012 to ensure a prompt access of these patients to rheumatology care.
Objectives: To evaluated predictors of early referral to EAC.
Methods: Consecutive patients referred to the EAC, from 2012 to 2019 were included. Medical records were reviewed to retrieve the following data: gender, age and referencing criteria as used by the general practitioner – arthritis (Y/N), positive squeeze test (Y/N), morning stiffness >30 minutes(Y/N), arthralgias (Y/N), rheumatoid factor (RF) (<60Ul/ml/≥60Ul/ml), antinuclear antibodies (ANA, <160/≥160), erythrocyte sedimentation rate (ESR) (≤20 mm/h/>20mm/h), C-reactive protein (CRP) (≤0.5 mg/dl/>0.5mg/dl). Time to referral (in days) was defined as the gap between the beginning of symptoms and the date of referral to EAC. Correlation between age and time to referral was analysed through Pearson correlation. Comparison between groups was assessed through Man-Whitney test. Variables with p <0.1 in univariate analysis were included in multivariable linear regression analysis.
Results: In total, 277 patients (66.4% female, mean age (±SD) of 53.2 (±17.7) years) were included. Mean time to referral to EAC was 103.5 (±89.1) days. In univariate analyses, presence of arthralgias (109.1 vs 88.2 days, p=0.037) and morning stiffness (119.5 vs 92.6 days, p=0.037) were both associated with longer time to referral. The identification of "arthritis" was associated with shorter time to referral (94.6 vs 117.2, p=0.012). In multivariate analyses, identification of arthritis (β -23.1, 95% CI, [-45, 0; -1.2], p=0.039) and absence of morning stiffness (β 24.1, 95% CI, [2.0; 46.2], p=0.033) were the only independent predictors of shorter time to referral.
Conclusion: Arthritis was the only criterion associated with earlier referral to EAC. Longer morning stiffness has, surprisingly, the opposite effect. This shows that current referral criteria used in our EAC are not being effective in reducing the time to referral patients with suspicion of arthritis. Active training programmes for primary health care physicians to improve the quality of clinical assessment and expedite the referral of these patients are required.

CO140 – "PRO REUMA INITIATIVE": COLLECTING PATIENT-REPORTED OUTCOMES USING TOUCHSCREEN TECHNOLOGY (DATA FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTER)

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Introduction: Patient-Reported Outcomes (PRO) have become a pivotal part of the evaluation of patients with rheumatic diseases and are now commonly used in clinical practice. However, the impact on clinical workflow and the time consumption can represent barriers to a more widespread adoption of PRO in routine rheumatology care. The "PRO Reuma Initiative" is a national project promoted by the Portuguese Society of Rheumatology, designed to overcome these challenges and to increase data collection in the Rheumatic Diseases Portuguese Register (Reuma.pt). In this initiative, touchscreen devices (tablets or kiosks) were strategically installed in the waiting room or in the outpatient department of different centres, so that patients can easily complete the questionnaires while waiting for the visit.
Objective: To assess patients' response rates to PRO

TABLE 1. PREDICTORS OF PROS' COMPLETION AMONG PATIENTS IN THE MULTIPLE GEE MODEL ANALYSES

Predictor	OR	p-value
6 months	1.310	**<0.001**
12 months	1.380	**<0.001**
Age	0.990	**0.013**
Gender (males)	1.174	0.123
Years of education	1.093	**<0.001**
Disease duration	1.008	0.167
DMARD use	1.235	**0.040**

questionnaires before and after the implementation of "PRO Reuma Initiative"; to identify predictors of PROs' completion.

Methods: A multicenter, prospective, observational study was conducted. Patients from centres participating in the "PRO Reuma Initiative" up to January 2020, registered in Reuma.pt, were included. Patients who had lost to follow-up or who had died were excluded. Data included socio-demographic and clinical characteristics, participating centre and the completion of PROs at baseline (T0), 6 (T6) and 12 (T12) months after implementation. All PROs available in Reuma.pt, for each diagnosis, were taken into consideration. Categorical variables are displayed as percentages and continuous variables as means and standard deviations. A logistic regression analysis through a generalized estimating equations (GEE) model was performed to identify predictors of PROs' completion.

Results: A total of 7008 patients were included, 69.3% female, mean age 44.6 ± 15.7 years. The most frequent diagnoses were Rheumatoid Arthritis (39.6%), Ankylosing Spondylitis (21.8%) and Psoriatic Arthritis (13.4%). Almost 61.5% of patients were currently under at least one conventional or biological disease-modifying anti-rheumatic drug (DMARD).

Overall, there was an increase in the percentage of patients who fulfilled PROs at 6 and 12 months (33.2% and 39.7%, respectively), compared to the baseline (29.9%). Concerning the diagnosis of the rheumatic disease, an increase in the completion of PROs was only detected for patients with Ankylosing Spondylitis (33.8% at T0, 39.9% at T6 and 39.7% at T12), Erythematosus Systemic Lupus (0.8% at T0, 0.6% at T6 and 1.9% at T12) and Vasculitis (0.4% at T0 and T6, and 0.6% at T12).

In the multiple GEE model analysis, the odds of fulfilling PROs at 6M and 12M are estimated to be 1.310 and 1.380 times higher (respectively) than the odds of doing it at baseline (p<0.001), adjusting for age, sex, disease duration, completed school years and current use of DMARD. Moreover, younger patients (OR 0.990, p=0.013), those with higher level of education (OR 1.093, p<0.001) and those taking at least one DMARD (OR 1.235, p=0.040) were more likely to fulfil PRO questionnaires.

Conclusion: There was an increase in response rates to PRO questionnaires after the implementation of the "PRO Reuma Initiative". These data suggest that the integration of new electronic devices in rheumatology services can improve data collection in Reuma.pt and standard clinical care. Further efforts to promote the completion of questionnaires through the use of touchscreen devices are encouraged.

CO150 – FIRST CLINICAL ANALYSIS OF MYOSITIS PATIENTS REGISTERED AT REUMA.PT/MYOSITIS PROTOCOL: DATA FROM A SINGLE-CENTER

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Introduction: Idiopathic inflammatory myopathies are a group of rare heterogeneous diseases that require a multidisciplinary and standardized approach.

Objective: To clinically characterize the patients with inflammatory myopathies followed at the hospital's Rheumatology Department, using the Rheumatic Diseases Portuguese Register (Reuma.pt/Myositis protocol).

Methods: An observational transversal study of patients with inflammatory myopathies registered at Reuma.pt/Myositis protocol was performed. Data on demographic variables, clinical features, myositis-specific and -associated antibodies and treatment were collected and complemented with data from the hospital clinical records.

Results: One hundred and thirty-three patients were identified out of which 12 were excluded because an inflammatory myopathy was ruled out. Of the 121 included patients, 77.7% were female, with a median age of 56 [44-68] years and a median disease duration of 2 [0-4] years. Thirty-six patients had a muscular biopsy done, of whom 28 (77.8%) had histological myositis features. Electromyography was performed in 56 patients, of whom 34 (60.7%) had a myopathic pattern. The most frequent diagnosis was dermatomyositis (DM; n=28, 23.1%), followed by

TABLE 1. CLINICAL CHARACTERISTICS AND IMMUNOLOGICAL PROFILE OF THE PATIENTS WITH INFLAMMATORY MYOPATHIES

Diagnosis (n)	Clinical features					Myositis antibodies (n)
	Skin disease, median [IQR]/ (n)	MMT-8, median [IQR]	Lung disease (n)	Others (n)	Malignancy (n)	
Definite DM (28)	DAS Skin 2 [0-2] Calcinosis (3) Mechanic hands (2)	80 [75-80]	NSIP (2); COP (1)	Arthritis (6)	Breast cancer (1)	anti-Ro52 (6); anti-Mi2b (5); anti-PmScl100 (4); anti-MI2a (3); anti-NXP2 (3); anti-SAE (2); anti-MDA5 (2); anti-Ku (2); anti-Tif1g (1)
Antisynthetase syndrome (21)	DAS Skin 0 [0-0] Calcinosis (1) Mechanic hands (4)	80 [80-80]	NSIP (11); UIP (3); LIP (1)	Arthritis (15); RF (10)	–	anti-Jo1 (14); anti-Ro52 (12); anti-Mi2b (1); anti-PL7 (3); anti-PL12 (1)
Probable DM (19)	DAS Skin 0 [0-0]	80 [78-80]	COP (1)	Arthritis (5); RP (6)	–	anti-Mi2a (4); anti-Mi2b (2); anti-Tif1g (2); anti-Ku (2); anti-PmScl75 (2)
CADM (16)	DAS Skin 1 [0-2] Calcinosis (1) Mechanic hands (1)	80 [77-80]	NSIP (1); COP (1)	Arthritis (5); RP (4)	–	anti-Mi2b (5); anti-Ro52 (2); anti-Mi2a (1); anti-MDA5 (1); anti-EJ (1); anti-SAE (1); anti-SRP (1); anti-Ku (1); anti-Tif1g (1)
MCTD (12)	DAS Skin 0 [0-1]	80 [79-80]	NSIP (1)	Arthritis (8); RP (10)	–	anti-U1 RNP (12); anti-Ro52 (7)
PM (7)	DAS Skin 0 [0-0] Calcinosis (1)	80 [72-80]	NSIP (1)	Arthritis (1)	Ovarian cancer (1); non-Hodgkin lymphoma (1)	anti-Ro52 (2); anti-SRP (1)
UCTD (5)	DAS Skin 0 [0-2] Calcinosis (1) Mechanic hands (1)	80 [79-80]	NSIP (2)	Arthritis (1); RP (5)	–	anti-ThTo (2); anti-SRP (1); anti-PL12 (1)
Overlap syndromes (12)	DAS Skin 0 [0-2] Mechanic hands (2)	80 [77-80]	NSIP (3); UIP (1)	Arthritis (4); RP (8)	–	anti-PmScl75 (4); anti-PmScl100 (1); anti-Ro52 (1); anti-Ro60 (1); anti-RNAPIII (1); anti-NOR90 (1); anti-MDA5 (1); anti-Ku (1)
Necrotizing myopathy (1)	DAS Skin 0	80	–	–	–	–

DM – dermatomyositis; CADM – clinically amyopathic dermatomyositis; MCTD – mixed connective tissue disease; PM – polymyositis; UCTD – undifferentiated connective tissue disease; ILD – interstitial lung disease; NSIP – nonspecific interstitial pneumonia; LIP – lymphocytic interstitial pneumonitis; UIP – usual interstitial pneumonia; COP – cryptogenic organizing pneumonia; RP – Raynaud phenomenon.

antisynthetase syndrome (ASS; n=21, 17.4%). At the time of the analysis, the median Manual Muscle Test (MMT-8) was 80 [78-80] and CK was 95.5 [58-161.5] U/L. The median modified skin Disease Activity Score (DAS) was 0 [0-2] and global disease activity was 0.5 [0-0.75]. Calcinosis was found in 6% of patients (n=7), mostly with DM. Interstitial lung disease (ILD) was present in 29 patients (24%): of those, 51.7% (n=15) had ASS, 10.3% (n=3) had DM and 10.3% (n=3) clinically amyopathic DM. Three patients (2 with polymyositis, 1 DM) presented as a paraneoplastic syndrome, diagnosed with breast cancer, ovarian cancer and non-Hodgkin lymphoma. Ninety-nine patients (81.8%) had an autoantibody result: antisynthetase autoantibodies were the most commonly identified (n=20, 16.4%). At the time of the analysis, 62% of the patients were treated with glucocorticoids, 35.5% with two or more disease-modifying anti-rheumatic drugs, 9% with rituximab and 7.4% with intravenous immunoglobulin.

Table 1 depicts the main clinical characteristics and the immunologic profile for each diagnosis.

Discussion/Conclusion: In our cohort the most frequent myositis subtype was DM. Although most patients had mild disease activity, almost a quarter of them had associated ILD, which is an important cause of morbidity and mortality. ILD was more frequent in ASS patients and was most commonly related to anti-Jo1 antibodies, which is consistent with the literature. This is the first analysis of a cohort of myositis patients applying the Reuma.pt/Myositis protocol. The results here shown elucidate the potential applications of this protocol for further longitudinal studies.

C0162 – PREVALENCE AND CLINICAL MANIFESTATIONS ASSOCIATED WITH ANTINUCLEAR ANTIBODIES SEROCONVERTION IN PATIENTS UNDER BIOLOGICAL TREATMENT

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Background: Biological drugs used in the treatment of rheumatic diseases have been associated with induction of other immune-mediated disorders. Despite a known risk of development of antinuclear antibodies (ANA) – seroconvertion – in patients under biological treatment (especially anti-TNF drugs) its clinical significance is still little-known. The usefulness of routine ANA dosing remains to be defined.

Objectives: Evaluate the prevalence of ANA seroconversion in rheumatic patients under biological treatment, as well as the prevalence on the development of clinical manifestations associated with seroconversion.

Methods: All patients at Centro Hospitalar de Vila Nova de Gaia / Espinho registered in the database Reuma.pt with the diagnosis of rheumatoid arthritis (RA), spondylarthritis (spA) and psoriatic arthritis (PsoA) who started biological treatment for at least 12 months were identified. The lack of ANA measurement before and after the biological treatment was defined as an exclusion criterion. Data concerning clinical information, previous biological treatment and autoantibodies profile were collected. The version V26 IBM SPSS ® was used in the statistical analysis.

Results: Out of 98 patients initially identified, 29 were excluded, since there were no signs of ANA before and after biological treatment. Hence, 69 patients have been included: 39 (56,5%) with RA, 22 (31,9%) with spA and 8 (11,6%) with PsoA. The sample showed an average age of 51,0 years (standard deviation±14,00 years), with 41 (59,4%) female patients. Eighteen (26,1%) were under adalimumab; 14 (20,3%) under tocilizumab; 11 (15,9%) under etanercept; 11 (14,5%) under infliximab; 8 (11,6%)

under rituximab; 6 (8,7%) under golimumab; 1 (1,4%) under ustekinumab and 1 (1,4%) under secukinumab. Seroconversion occurred in 19 (27,5%) patients: in 5 (50%) under infliximab, 7 (38,9%) under adalimumab; 3 (27,3%) under etanercept; 3 (21,4%) under tocilizumab and 1 (12,5%) under rituximab. The prevalence difference of seroconversion during biological treatment showed no statistically-relevant significance. Time until seroconversion presented a median of 37,7 months (3-105). Anti-dsDNA new positivity occurred in 4 patients (21,1%) and anti-histones in 4 patients (21,1%). Seroconversion was associated with development of clinical manifestations in 4 patients (21,1%): 2 cases (10,55%) of induced lupus after beginning the treatment with infliximab and 2 cases (10,55%) of alopecia areata (AA) after beginning the treatment with etanercept and adalimumab. All 4 patients switched to a different biological treatment, with improvement of symptoms in 3; the remaining one switched recently and still didn't improved.

Conclusions: ANA seroconversion is a common event associated with biological drugs, especially anti-TNF. However, development of clinical manifestations occurs in a minority of cases. One should be aware of the importance of evaluating not only the cases classified as drug induced lupus, but also other manifestations that are associated with the event of seroconversion.

CO167 – MATERNAL AND PERINATAL OUTCOMES IN WOMEN WITH RHEUMATIC DISEASES – A 10-YEAR EXPERIENCE FROM A PORTUGUESE TERTIARY CENTRE

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Background: Pregnant women with rheumatic diseases (RD) represent a population at a higher risk for adverse pregnancy outcomes (APO). At our unit, these patients (pts) are surveilled at a high-risk pregnancy clinic by both rheumatologists and obstetricians.

Objectives: To assess pregnancy outcomes in pts with RD surveilled at our unit over the last decade.

Methods: Single-centre observational retrospective study of pregnant women with RD surveilled at a tertiary centre between 2009 to 2019.

Results: Overall, 353 pregnancies (preg) in 295 pts with RD were managed at our unit. Table 1 summarizes clinical data and the main APO recorded. Systemic lupus erythematous (SLE) was the leading diagnosis followed by spondyloarthritis (SpA) and rheumatoid arthritis (RA). Antiphospholipid syndrome (APS) was diagnosed in 49 (14%) preg. Mean maternal age at conception was 33±5.5 and mean duration of disease 8.2 ± 7.3 years. We documented 284 (78%) live births (9 twin preg), 32 (10%) miscarriages, 7 (2%) elective abortions and 2 stillbirths (1%); 35 (10%) preg were lost to follow up. Miscarriages occurred predominantly in pts with APS (34%). Mean gestational age at delivery was 38.36 ± 2.18 weeks, with a total of 36 (16%) preterm births recorded – mainly in APS, SLE and juvenile idiopathic arthritis pts. Fetal growth restriction was detected in 6% of preg, more than 1/3 of those in pts with APS. Preeclampsia complicated a total of 10 (4%) preg, 3 of those with HELLP syndrome, with SLE and APS underlying 60% of these cases. A total of 4 (1%) malformations were recorded – 1 case of central nervous system malformation and 1 of trisomy 18, both after warfarin exposure during the 1st trimester; 1 case of esophageal atresia and 1 of clubfoot, both in SpA pts not exposed to teratogenic drugs. Neonatal lupus ensued in 3 (4%) out of 80 preg of pts positive for anti-Ro/La antibodies. No neonatal deaths were recorded. SpA and RA represented the diseases which flared the most considering both pregnancy and the post-partum period. A total of 198 pts (56%) were treated with glucocorticoids during pregnancy, most of them (63%) at low doses (≤5mg/day of prednisolone or equivalent). Conventional synthetic (cs) disease-modifying antirheumatic drugs (DMARDs) were used in a total of 186 (53%) preg. In 27 pts (8%), a biologic (b) DMARD was used in the year prior to conception, but only in 6 pts were bDMARDs continued beyond the 2nd trimester of gestation – 1 pt with enteropathic SpA stayed on adalimumab until the 28 week (w) of gestation; 1 pt with Takayasu´s disease received tocilizumab until w 30; 1 axial SpA pt kept etanercept until w 36; 3 pts (2 SpA and 1 RA) took certolizumab throughout pregnancy. No serious infections were detected in the mothers or newborns. In 14 (4%) pts, methotrexate was not properly washed out before pregnancy or it was just stopped at a positive pregnancy test – 3 decided to perform medical termination of the pregnancy, the others had live infants with no malformations.

TABLE 1 – PREGNANCY OUTCOMES BY DISEASE

Main diagnosis	N (%)	Age at conception (years)	Gestational age at delivery (weeks)	BW, grams (g)	Low BW (<2500g), n/mN (%)	Miscarriages, n/mN (%)	FGR, n/mN (%)	PE, n/mN (%)	Preterm births, n/mN (%)	Flares during pregnancy, n/mN (%)	Flares during post-partum, n/mN (%)
SLE	116 (32.9)	32.6±5.3	38.0±2.3	2848.1±567.9	16/67 (23.9)	13/110 (11.8)	6/82 (7.3)	5/78 (6.4)	15/76 (21.1)	16/92 (17.4)	5/79 (6.3)
SpA	60 (17.0)	33.4±4.7	39.0±2.6	3136.5±575.6	2/30 (6.7)	2/55 (3.6)	5/44 (11.4)	1/45 (2.2)	5/40 (12.5)	18/40 (45.0)	5/23 (21.7)
RA	51 (14.4)	34.5±4.8	38.9±1.2	3106.4±580.1	2/26 (7.7)	6/49 (12.2)	1/35 (2.9)	1/33 (3.0)	1/29 (3.4)	10/37 (27.0)	7/30 (23.3)
Vasculitis	25 (7.1)	33.7±6	38.2±1.8	2870.3±498.0	4/19 (21.1)	1/24 (4.2)	1/21 (4.8)	0/19 (0)	4/21 (19.0)	7/23 (30.4)	2/17 (11.8)
Primary APS	22 (6.2)	33.3±4.1	38.2±1.6	2954.8±535.4	2/13 (15.4)	3/22 (13.6)	1/17 (5.9)	1/16 (6.3)	2/16 (12.5)	-	-
JIA	17 (4.8)	26.8±8.3	37.6±2.5	2979.5±641.0	2/10 (20.0)	1/17 (5.9)	0/12 (0)	0/11 (0)	4/12 (33.3)	2/13 (15.4)	1/8 (12.5)
UCTD	15 (4.2)	32.9±5	39.0±1.5	2902.5±561.3	2/10 (20.0)	2/15 (13.3)	0/11 (0)	1/10 (10.0)	1/11 (9.1)	0/15 (0)	0/15 (0)
Primary Sjögren Syndrome	12 (3.4)	36.5±4.6	38.5±1.4	3319.3±302.0	0/7 (0)	0/11 (0)	0/9 (0)	0/10 (0)	1/9 (11.1)	1/8 (12.5)	0/6 (0)
Others	35 (9.9)	32.9±5.5	38.3±2.3	2787.0±452.5	4/15 (26.7)	4/31 (12.9)	2/20 (10.0)	1/18 (5.6)	3/18 (16.7)	4/27 (14.8)	1/22 (4.5)
Total	353 (100)	33.0±5.5	38.4± 2.2	2951.3 ± 579.0	34/197 (17.3)	32/334 (9.6)	16/251 (6.4)	10/240 (4.2)	36/232 (15.5)	58/255 (22.7)	21/200 (10.5)
Secondary diagnosis											
APS	27	32.7±4.7	36.6±2.7	2537.0±426.3	7/15 (46.7)	8/26 (30.8)	5/16 (31.3)	2/12 (16.7)	5/15 (33.3)	-	-
Sjögren Syndrome	9	33.1±4.3	36.7±2.5	2719.8±765.4	2/6 (33.3)	1/9 (11.1)	1/7 (14.3)	0/7 (0)	3/6 (50)	-	-

APS – antiphospholipid syndrome; BW – birthweight; FGR – fetal growth restriction; JIA – juvenile idiopathic arthritis; PE – preeclampsia; RA – rheumatoid arthritis; SLE – systemic lupus erythematous; SpA – spondyloarthritis; UCTD – undifferentiated connective tissue disease. Continuous variables are presented as mean±SD; categorical variables as n/modified(m)N (total N – not applicable+missing data), %. "Others" accounts for diagnosis with N≤6 such as APS *non criteria*, cutaneous lupus, idiopathic uveitis and erythema nodosum, mixed connective tissue disease, myositis, overlap syndromes, Still's disease, systemic sclerosis and undifferentiated arthritis.

In the total cohort, a diagnosis of APS showed to significantly increase the risk of APO – OR 2.0, 95%CI 1.0-4.0. In SLE pts, renal involvement was associated with APO – OR 4.1, 95% CI 1.1-16.3.

Conclusions: In pregnant women with RD, it is of vital importance to be aware of the increased risk for APO. In our cohort, APS and SLE were the conditions most associated with APO, while SpA and RA were responsible for most maternal flares. Nevertheless, the majority of these pts, surveilled by a multidisciplinary team, had successful gestations.

CO180 – FRAX 10-YR FRACTURE RISK IN RHEUMATOID ARTHRITIS ASSESSED WITH AND WITHOUT BONE MINERAL DENSITY – ARE WE TREATING OUR PATIENTS UNDER BDMARDS?

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Background: Patients with rheumatoid arthritis (RA) have a higher risk of osteoporosis not only due to chronic inflammation status, but also due to the treatment with glucocorticoids. FRAX is a computer-based algorithm developed by the World Health Organization for estimation of the 10-year risk of a hip or major osteoporotic fracture. Inclusion of femoral neck bone mineral density (BMD) in the estimation is optional.

Objectives: The study aimed to identify the RA patients under treatment with biological disease-modifying antirheumatic drug (bDMARD), who have FRAX scores, calculated with and without BMD, classified as high fracture risk and evaluate if they are receiving treatment for osteoporosis. The authors also investigated the intra-individual agreement between FRAX fracture risk calculated with and without BMD.

Methods: Demographic and clinical data and BMD results from RA patients followed in a tertiary university hospital and registered in the Rheumatic Diseases Portuguese Register were used for analysis. Patients under 40 years of age at the last visit were excluded. McNemar test was applied for the identification of discordance of risk categories. The Wilcoxon test was used to characterize the intraindividual differences between paired FRAX risks with and without BMD. Correlations between pairs of variables were evaluated by the Spearman test. For independent variables Mann-Whitney test was used.

Results: A total of 303 patients were included, 244 were females (80.5%) and 49 current smokers (16.2%). Mean age was 59.5 ± 9.54 years. Two hundred and twenty patients (72.4%) and 243 (80.2%) were RF and ACPA positive, respectively, and 51.5% had erosive disease. Mean disease activity score (DAS28-4V-CRP) was 3.08 ± 1.18 and mean femoral neck BMD 0.84 ± 0.12 g/cm2. Among

all the patients, 35 (11.6%) had previous fractures and 19 (6.3%) have family history of fracture. The median 10-year risk of a major fracture and a hip fracture, calculated without BMD, was 6,0 (1,2-50) and 1,5 (0,1-39), respectively; with BMD it was 6.9 (1.3-61) and 1.7 (0-49). When FRAX score is calculated without BMD (n=303), 76 (25.1%) patients were categorized as high fracture risk. Among them, only 41 (54%) were receiving osteoporosis treatment. FRAX assessment with BMD (n=231) identified 99 (32.7%) patients with high fracture risk, 51 (51,5%) in treatment for osteoporosis. Thirty patients (21%) previously classified as low fracture risk using FRAX without BMD were recategorized as high risk ($p<0.001$). Despite that, there was a strong correlation between fracture risks assessed with and without BMD for both major and hip fracture ($r = 0.867$, $p < 0.0001$ and $r = 0.728$, $p < 0.0001$, respectively). ACPA and RF positive patients did not have higher FRAX scores (including or not BMA). Patients with erosive disease had a higher 10-year probability of major fracture evaluated by FRAX when it includes BMD ($p=0.041$).

Conclusion: It is very important to accurately assess the risk of osteoporotic fractures in RA patients to treat them properly. The authors highlight the high number of patients who are not receiving treatment according to FRAX categorization. In spite of the correlation between estimated fracture risk by FRAX with and without BMD, there is a discordance in fracture risk categorization, as one fifth of patients of low risk were reclassified as high risk. For the RA population treated with bDMARDS, our findings raise the need to request a DXA not only for patients classified as having an intermediate risk of fracture, but also for low-risk patients.

CO197 – FOLLOW UP OF INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES – THE EXPERIENCE OF ONE CENTRE

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Background: Interstitial Lung Diseases (ILD) may present features suggesting an underlying autoimmune process, which seem to differentiate them from idiopathic interstitial pneumonias, although without fully meeting the classification criteria (CC) for a specific connective tissue disease. Different terms had been used to describe these conditions and, to reach a consensus, the European Respiratory Society/American Thoracic Society proposed the CC for an entity named Interstitial Pneumonia with Autoimmune Features (IPAF). Clinical evolution and prognosis of this entity are still poorly understood.

Objectives: To evaluate clinical evolution and prognosis of a population of patients with IPAF.

Methods: Retrospective analysis of clinical files of patients followed by the Pulmonology Department since 02/2012 until 06/2019, who met the CC for IPAF, regarding clinical, functional and radiological evolution. Patients were considered to have a progressive phenotype in 24±3 months from their 1st evaluation if they fulfil 1 of the 4 criteria: relative decline in FVC ≥10% predicted; relative decline in FVC ≥5–<10% predicted and worsened respiratory symptoms; relative decline in FVC ≥5–<10% predicted and increased extent of fibrosis on High-resolution Computed Tomography (HRCT); worsened respiratory symptoms and increased extent of fibrosis on HRCT.

Results: 22 (7.4%) of 296 ILD patients met IPAF CC. 59.0% were female with an age at the 1st evaluation of 66.7±12.4 years. They were all non-smokers (63.6%) or ex-smokers (36.4%). Serologic and morphologic criteria were both present in 21 (95.4%) and clinical criteria in 5 patients (22.7%). Antinuclear antibodies (ANA) were identified in 19, rheumatoid factor in 4, SSA in 3 and anti-Jo-1 in 1 patient. HRCT patterns were identified in 21 patients: 15 nonspecific interstitial pneumonia (NSIP), 5 organizing pneumonia (OP) and 2 lymphocytic interstitial pneumonia (LIP). One NSIP and 1 LIP identified on HRCT were confirmed by histopathology. Three patients had inflammatory arthritis and 2 had Raynaud's phenomenon. Immunosuppressive therapy was introduced in most cases (18 patients, including systemic corticotherapy in 17, azathioprine in 4, mycophenolate mofetil in 1), azithromycin was prescribed in 2 patients and 3 remained without therapy. Regarding the follow up at 24±3 months from the 1st evaluation (3 patients were excluded due to too recent follow -up), 4 patients (18.2%) had progressive phenotype, 7 (31.8%) had a favourable evolution and 3 (13.6%) patients had died. During a follow-up of 31.1±19.8 months, this number rose to 6 patients (27.3%), all of them died by respiratory cause and had NSIP pattern. No differences were found in age, last FVC, therapy and time of disease evolution between those who died and the others.

Conclusion: Our study showed that a small proportion of IPAF patients had a progressive phenotype and the NSIP pattern seemed to be a poor prognosis factor for survival.

CO198 – PRESCRIPTION PATTERNS AND

DISEASE ACTIVITY IN PORTUGUESE WOMEN OF CHILDBEARING AGE WITH RHEUMATOID ARTHRITIS, PSORIATIC ARTHRITIS, ANKYLOSING SPONDYLITIS AND JUVENILE IDIOPATHIC ARTHRITIS

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Background: Disease activity (DA) at conception is one of the main predictors of pregnancy outcomes in women of childbearing age (WoCBA) with rheumatic diseases. Known teratogenicity of some disease modifying anti-rheumatic drugs (DMARDs) and uncertainty about iatrogenic obstetric events of the more recent ones might limit choice of treatment in WoCBA. Few studies have addressed this issue so far, most of them revealing a trend towards discontinuation of conventional synthetic and biological (b) DMARDs in up to half of WoCBA between 12 to 3 months before conception. Methotrexate (MTX), although teratogenic, remains one of the most prescribed DMARD during reproductive years (y). No study has evaluated prescription patterns in WoCBA compared to post-menopausal women (PMW) and male patients.

Objectives: To assess differences in prescription patterns between WoCBA with rheumatoid arthritis (RA), psoriatic arthritis (PsA), ankylosing spondylitis (AS) and juvenile idiopathic arthritis (JIA) and comparator groups, namely PMW and age-matched men. Evaluate DA in WoCBA comparing to the aforementioned groups.

Methods: Observational transversal study, using data from the Reuma.pt of 3 national centres. Adult patients with the diagnosis of RA, PsA, AS or JIA were allocated to the following groups: WoCBA (aged 18–44y), young men (YM) (18–44y), PMW (≥ 45y) and matched men (≥45y). Demographic and clinical variables are described as means or frequencies. Differences between groups regarding therapy and DA were assessed with Chi-square and ANOVA tests. Linear and logistic regression models were used to find predictors of DA and prescription patterns.

Results: 2133 patients were included, 69.9% female with a mean age of 55.96±15.85 y. 1437 patients were diagnosed with RA, 305 with PsA, 254 with AS and 137 with JIA. Demographic and clinical data are

TABLE 1 – DEMOGRAPHIC AND CLINICAL DATA

	Women of childbearing age (n= 317)	Young Men (n=186)	Postmenopausal Women (n= 1173)	Men (n=455)
Age (years), mean ± SD	34.07±7.29	32.51±7.97	63.39±10.22	62.82±10.17
Caucasian, n (%)	179 (56.5)	105 (56.5)	604 (51.5)	249 (54.8)
Smokers, n (%)	23 (7.3)	39 (20.9)	73 (6.2)	71 (15.6)
Body mass index (Kg/m^2), mean ± SD	25.4±7.0	24,1±4,4	27,8±5,1	27.8±4.3
Diagnosis				
- Rheumatoid arthritis, n (%)	173 (54.6)	22 (11.8)	1015 (86.5)	227 (49.9)
- Psoriatic arthritis, n (%)	36 (11.4)	50 (26.9)	96 (8.2)	123 (27.0)
- Ankylosing spondylitis, n (%)	39 (12.3)	63 (33.9)	53 (4.5)	99 (21.8)
- Juvenile idiopathic arthritis, n (%)	69 (21.8)	51 (27.4)	9 (0.8)	96 (1.3)
Disease duration (years), mean ± SD	11.5±8.2	11.1±6.9	15.8±11.9	17.5±13.3

FIGURE 1 – PRESCRIPTION PATTERNS AND DISEASE ACTIVITY

	A -WoCBA (N=256)	B - Young Men (N=161)	C - Post menopausal Women (N=927)	D - Men N=340	Chi-square test	
Medications, n (%)						
NSAIDs	143 (55.9)	111(68.9)	472 (50.9)	169 (49.7)	p<0.001	
Glucocorticoids	106 (41.4)	31 (19.3)	625 (67.4)	154 (45.3)	p<0.001	
csDMARDs						
- Methotrexate	149 (58.2)	60 (37.3)	663 (71.5)	197 (57.9)	p<0.001	
- Leflunomide	12 (4.7)	4 (2.5)	45 (4.9)	2 (0.6)	p=0.003	
- Sulfassalazine	9 (3.5)	5 (3.1)	39 (4.2)	9 (2.7)	NS	
- HCQ	36 (14.1)	4 (2.5)	117 (12.6)	18 (5.3)	p<0.001	
bDMARDs						
- Etanercept	48 (18.8)	29 (18.0)	140 (15.1)	66 (19.4)	NS	
- Infliximab	9 (3.5)	11 (6.8)	36 (3.9)	30 (8.8)	p=0.002	
- Adalimumab	17 (6.6)	15 (9.3)	47 (5.1)	27 (7.9)	NS	
- Golimumab	15 (5.9)	18 (11.2)	37 (4.0)	30 (8.8)	p<0.001	
- Certolizumab	10 (3.9)	0 (0)	2 (0.3)	2 (0.6)	p<0.001	
- Tocilizumab	12 (4.7)	2 (1.2)	71 (7.7)	10 (2.9)	p<0.001	
- Rituximab	5 (1.9)	0 (0)	50 (5.4)	4 (1.2)	p<0.001	
- Abatacept	0 (0)	0 (0)	9 (1)	0 (0)	NS	
- Secukinumab	1 (0.4)	4 (3.5)	8 (0.9)	2 (0.6)	NS	
- Ustekinumab	1 (0.4)	3 (1.9)	5 (0.5)	0 (0)	NS	
tsDMARDs						
- Baricitinib	2 (0.8)	0 (0)	5 (0.5)	0 (0)	NS	
- Tofacitinib	2 (0.8)	1 (0.6)	4 (0.4)	0 (0)	NS	
Disease activity and function					A vs B	A vc C
RA and peripheral PsA, mean± SD						
- DAS28	3.03±1.39	2.32±1.18	3.47±1.38	2.79±1.31	p=0.005	p=0.001
- HAQ	0.54±0.53	0.31±0.46	1.13±0.75	0.69±0.71	NS	p<0.001
Axial PsA and AS						
- BASDAI	3.55±2.01	2.43±1.66	3.96±2.23	3.28±2.14	p=0.014	NS
- BASFI	3.02±2.36	1.89±1.87	4.64±2.74	4.23±2.59	NS	p<0.001
JIA						
- JADAS27	7.33±6.76	6.32±6.27	9.58±4.47	7.54±7.76	NS	NS
- HAQ	0.44±0.56	0.20±0.30	1.25±0.47	0.85±0.60	NS	p<0.001

BASDAI – Bath ankylosing spondylitis disease activity índex, BASFI – Bath ankylosing spondylitis functional index, bDMARDs – biologic disease modifying antirheumatic drugs, csDMARDs – conventional synthetic disease modifying antirheumatic drugs, DAS – disease activity score, HAQ – Health assessment questionnaire disability índex, HCQ – hydroxychloroquine, JADAS – Juvenile Arthritis Disease Activity Score, NSAIDs – non-steroidal anti-inflammatory drugs, PsA – psoriatic arthritis, RA – rheumatoid arthritis, tsDMARD – targeted synthetic disease modifying antirheumatic drugs, WoCBA – women of childbearing age

presented in table 1. WoCBA and YM receive more NSAIDs than older patients. WoCBA were less likely to be treated with glucocorticoids than PMW (OR 0.66 95%CI 0.44-0.99), after adjusting for diagnosis, disease duration and DA. MTX was the main DMARD used in all groups. WoCBA were 1.76 times more likely to be treated with MTX than YM (95%CI 1.04-2.97). Regarding bDMARDs, etanercept was the most used across all groups. Certolizumab was specially prescribed in WoCBA (OR 13.8, 95%CI 1.4-132.8), although prescription rates were low. WoCBA had higher DA scores than YM: DAS28 and BASDAI were higher in RA/peripheral PsA and AS/axial PsA respectively, when adjusted for diagnosis, disease duration and medication (p=0.004). Functional indexes scores were similar between WoCBA and YM and lower in WoCBA than PMW. Patterns of prescription and DA are detailed in figure 1.

Conclusion: Certolizumab was prescribed preferentially in WoCBA reflecting concern and observation of Portuguese recommendations for bDMARDs safety in pregnancy. Despite receiving more MTX than YM, DA in WoCBA was not well controlled, which may influence future pregnancy outcomes. Ensuring tight DA control in WoCBA through proper medication remains an unmet clinical need.

CO224 – EFFICACY OF BIOSIMILAR INFLIXIMAB CT-P13 COMPARED TO ORIGINATOR INFLIXIMAB IN RHEUMATOID ARTHRITIS AND AXIAL SPONDYLOARTHRITIS PATIENTS: DATA FROM THE PORTUGUESE REGISTER REUMA.PT

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Background: Biosimilars were developed with the purpose of offering the same efficacy and safety as originator drugs at lower prices, thus generating significant cost-savings. Evidence continues to grow from clinical trials and data from 'real-life' settings supporting that there are no differences in efficacy or safety between biosimilar and biooriginator.

Objective: To compare the efficacy of the biosimilar infliximab CT-P13 with originator infliximab over 24 months of follow-up in patients with rheumatoid arthritis (RA) and axial spondyloarthritis (axSpA), using real-world clinical data from the Rheumatic Diseases Portuguese Register (Reuma.pt).

Methods: Biological-naive patients from Reuma.pt, with a clinical diagnosis of RA or axSpA, who were starting either the infliximab biosimilar CT-P13 or the originator infliximab, after 2014 (date of market entry of CT-P13 in Portugal), were included. The two treatment options (biosimilar and originator) were compared regarding different response outcomes at 3 and 6 months, in both conditions, adjusting for age, gender and baseline C-Reactive Protein, selected a priori on clinical grounds. The main outcome was the change in DAS28-ESR for RA and the ASDAS-CRP for axSpA. Additionally, the effect of infliximab biosimilar vs originator on different response outcomes over 24 months of follow-up was tested using multivariable generalized estimating equations (GEE) adjusted for the same confounders.

Results: In total, 140 patients were included, 66 (47%) of which with RA. The distribution of patients starting the infliximab biosimilar and the originator was the same between the two conditions (58% for the biosimilar [N=38 for RA and N=41 for axSpA] and 42% for

TABLE 1. ASSOCIATION BETWEEN TREATMENT AND RESPONSE OUTCOMES IN RA AND AXSPA PATIENTS OVER 24 MONTHS OF FOLLOW-UP

Variables	Biosimilar vs Originator
	Rheumatoid Arthritis
Outcomes	
DAS28 (3V) ESR, β (95% CI) [N=65, n=200 visits]	**0.6 (0.2;1.1)**
DAS28 (3V) ESR<2.6, OR (95% CI) [N=65, n=200 visits]	0.4 (0.1;1.4)
DAS28 (3V) ESR≤3.2, OR (95% CI) [N=65, n=200 visits]	0.5 (0.2;1.2)
CDAI, β (95% CI) OR (95% CI) [N=56, n=172 visits]	2.3 (-1.5;6.2)
CDAI≤2.8, OR (95% CI) [N=50, n=119 visits]	1.0 (0.9;1.2)
CDAI≤10, OR (95% CI) [N=50, n=119 visits]	1.0 (0.8;1.3)
SDAI, β (95% CI) [N=54, n=167 visits]	2.8 (-1.3;7.0)
SDAI≤3.3, OR (95% CI) [N=49, n=115 visits]	1.1 (0.3;4.0)
SDAI≤11, OR (95% CI) [N=49, n=115 visits]	1.2 (0.5;3.0)
HAQ, β (95% CI) [N=54, n=126 visits]	**0.4 (0.1;0.7)**
ACR-EULAR RC, OR (95% CI) [N=57, n=140 visits]	1.4 (0.4;5.5)
	Axial Spondyloarthritis
Outcomes	
BASDAI, β (95% CI) [N=73, n=284 visits]	0.1 (-0.7;0.9)
ASDAS, β (95% CI) [N=72, n=281 visits]	0.0 (-0.4;0.3)
BASFI, β (95% CI) [N=68, n=265 visits]	-0.3 (-1.3;0.7)
BASDAI50, OR (95% CI) [N=67, n=210 visits]	1.1 (0.5;2.5)
ASDAS CII, OR (95% CI) [N=65, n=207 visits]	1.5 (0.6;3.7)
ASDAS MI, OR (95% CI) [N=65, n=207 visits]	2.8 (1.0;8.2)
ASDAS LDA, OR (95% CI) [N=68, n=212 visits]	0.7 (0.2;2.1)
ASDAS ID, OR (95% CI) [N=140, n=57 visits]	1.0 (0.5;2.2)

GEE models with the treatment group as predictor (reference category: originator); all models adjusted for age, gender, and baseline CRP. OR or β in bold are statistically significant (p<0.05). β, Beta coefficient. OR, Odds Ratio. 95% CI, 95% Confidence Interval. DAS28, Disease Activity Score-28. DAS28 (3V) ESR<2.6, DAS28 Remission. DAS28≤3.2, DAS28 Low Disease Activity. CDAI, Clinical Disease Activity Index. CDAI≤2.8, CDAI Remission. CDAI≤10, CDAI Low Disease Activity. SDAI, Simple Disease Activity Index. SDAI≤3.3, SDAI Remission. SDAI≤11, SDAI Low Disease Activity. HAQ, Health Assessment Questionnaire. ACR-EULAR RC, American College of Rheumatology-European League Against Rheumatism Boolean Remission Criteria. BASDAI, Bath Ankylosing Spondylitis Disease Activity Index. ASDAS, Ankylosing Spondylitis Disease Activity Score. BASFI, Bath Ankylosing Spondylitis Functional Index. BASFI, Bath Ankylosing Spondylitis Functional Index. BASDAI50, BASDAI 50 Response. ASDAS CII, ASDAS Clinical Important improvement. ASAS MI, ASDAS Major Improvement. ASDAS LDA, ASDAS Low Disease Activity.

the originator [N=28 for RA and N=31 for axSpA]). From the 66 patients with RA, 82% were females with a mean age of 55.6 (SD 11.5) and mean DAS28-ESR of 5.0 (1.4) at baseline. As for the axSpA patients, 53% were males with a mean age of 46.4 (13.0) and a mean ASDAS-CRP of 3.7 (0.9) at baseline. There were no differences in efficacy between patients treated with the infliximab biosimilar and the originator for both RA (ΔDAS28 at 3 months: -0.6 (95% CI -1.3;0.1) vs -1.2 (-2.0;-0.4) and at 6 months: -0.7 (-1.5;0.0) vs -1.5 (-2.4;-0.7)) and axSpA (ΔASDAS at 3 months: -1.6 (-2.0;-1.1) vs -1.4 (-1.8;-0.9) and at 6 months: -1.5 (-2.0;-1.1) vs -1.1 (-1.5;-0.7)). Over 24 months of follow-up there was no difference in response to treatment between biosimilar and originator either.

Conclusion: Our results add to existing body of evidence by showing no differences in the efficacy profiles between the infliximab biosimilar CT-P13 and the infliximab originator in the treatment of patients with active RA and axSpA over 24 months.

CO240 – INFEÇÕES EM DOENTES REUMÁTICOS SOB TRATAMENTO BIOLÓGICO: O PAPEL DETERMINANTE DA CORTICOTERAPIA

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Introdução: Apesar dos anos de experiência com bDMARDs, pouco se conhece acerca do risco infecioso comparado entre diferentes bDMARDs, com diferentes mecanismos de ação. O objetivo deste estudo foi avaliar a incidência e tipo de infeções nos doentes sob bDMARDs, averiguar um eventual diferencial de risco para infeção grave entre os bDMARDs e determinar possíveis preditores.

Métodos: Estudo unicêntrico retrospetivo que incluiu todos os doentes registados no Reuma.pt com artrite reumatóide (AR), artrite psoriática (AP) e espondilartrite (SpA), com pelo menos um relato de infeção sob bDMARD como evento adverso até agosto de 2019. A classificação ICD-10 foi utilizada para definir o tipo de infeção e esta foi considerada grave sempre que motivou hospitalização. À data da infeção, foram colhidos dados demográficos e clínicos para estabelecer comparações entre os diferentes grupos de bDMARDs. Análise de regressão logística foi utilizada para avaliar preditores de infeção grave.

Resultados: Foram incluídas 121 infeções, das quais 98 (81,0%) no sexo feminino, com idade média de 55,1 ±13,5 anos [88 (72,7%) em doentes com AR, 20 (16,5%) com SpA e 13 (10,7%) com AP]. À data da infeção, 84 (67,7%) doentes estavam sob glucocorticoides, com dose mediana em equivalentes de prednisolona de 5 (5-7,5) mg/dia. Quanto a bDMARDs, 28 (23,1%) realizavam etanercept, 26 (21,5%) rituximab (RTX), 23 (19,0%) adalimumab (ADA), 17 (14,0%) tocilizumab (TCZ), 13 (10,7%) golimumab, 10 (8,3%) infliximab, 3 (2,3%) certolizumab e 1 (0,8%) abatacept. Em 28 (23,1%) casos ocorreu infeção da pele e tecidos moles, 27 (22,3%) respiratórias, 24 (19,8%) outras doenças infeciosas e parasitárias, 20 (16,5%) genitourinárias, 8 (6,6%) gastrointestinais, 11 (9,1%) musculoesqueléticas, 2 (1,7%) olhos

e anexos e 1 (0,8%) infeção do sistema nervoso. Em 52 casos (43,0%) a infeção foi considerada grave.
Verificou-se uma associação significativa entre o uso corrente de glucocorticoides e o surgimento de infeções graves (53,6% vs 18,9%, p<0,001), com claro efeito dose-dependente (p=0,006). O uso de csDMARDs ou bDMARDs, quer quando avaliados isoladamente, quer como grupo, não se associou com a ocorrência de infeção grave. A exposição aos glucocorticoides foi superior nos doentes sob TCZ (94,1% vs 65,4%, p=0,017) e RTX (92,3% vs 63,2%, p=0,004), sendo que para os sob TCZ a dose mediana foi superior à dos restantes bDMARDs [7,5 (5,0) vs 5 (7,5), p=0,003]. Curiosamente, após infeção, a frequência de switch foi superior nos sob TCZ face aos outros bDMARDs (47,1% vs 12,5%, p=0,002). Ser fumador associou-se a um OR de 27,66 de ter infeção grave (IC 95% 2,76-277,09), a presença de patologia pulmonar a um OR de 19,36 (IC95% 1,70-220,98) e o uso corrente de glucocorticoides a um OR de 443,59 (13,09-15031,23). Ajustando para estes fatores, verificou-se que nenhum bDMARD em particular se associava à ocorrência de infeções graves, sendo que ADA (OR 0,01; IC95% 0,01-0,34) e TCZ (OR 0,04; IC95% 0,01-0,84) conferiam menor risco de infeção grave. A diabetes mellitus, doença de base, idades à infeção e ao diagnóstico, as durações de doença e do tratamento com bDMARD não se associaram a infeção grave.
Conclusão: O risco de infeção grave parece ser determinado sobretudo pela corticoterapia concomitante, de uma forma dose-dependente, não havendo diferenças em função do bDMARD prescrito. Assim, por forma a diminuir o risco infecioso nos doentes sob bDMARD, os autores realçam a importância de se utilizar a menor dose possível de glucocorticoides e, sempre que possível, descontinuá-los.

CO263 – CONCORDANCE BETWEEN THE NEW SLE-DAS, DORIS AND DORIA REMISSION CRITERIA FOR SLE: ARE THEY DIFFERENT IN A REAL-LIFE CLINICAL SETTING?

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Background: The treat-to-target strategy in Systemic Lupus Erythematosus (SLE) aims to achieve remission. However, to define a target based on SLE Disease Activity Index (SLE-DAI-2K) is questionable, due to its limitations (especially its dichotomous nature). The currently used Doria and DORIS clinical remission criteria are both based on SLEDAI.
The Systemic Lupus Erythematosus Disease Activity Score (SLE-DAS) is a recently validated continuous disease activity score which demonstrated higher accuracy and improved sensitivity to change as compared to SLEDAI-2K. In addition, it comprises important manifestations ignored by SLEDAI, including hemolytic anemia, gastrointestinal and cardiopulmonary involvement.
Objective: Our primary goal was to compare the attainment of clinical remission defined by SLE-DAS, DORIS and Doria criteria in a real-life clinical setting. In addition, we aimed at evaluating disease activity distribution according to SLE-DAS in our cohort of lupus patients.
Methods Cross-sectional study of consecutive SLE patients fulfilling ACR'97 and/or SLICC'12 classification criteria followed at an academic lupus clinic from January to December 2019.
The SLE-DAS clinical remission criteria were defined as a score of 0 in all clinical items of SLE-DAS and current prednisolone dose ≤ 5 mg/day. The SLE-DAS cut-off values to define disease activity states were previously defined in the Padova Lupus Cohort: low disease activity (LDA) if SLE-DAS ≤ 3.77 in patients not fulfilling remission criteria; mild disease activity if 3.77 < SLE-DAS ≤ 7.64; and moderate-to-severe disease activity if SLE-DAS>7.64.
Fulfillment of DORIS, Doria and SLE-DAS clinical remission status was verified for each patient. The attainment of clinical remission according to these definitions was compared. We further classified all patients regarding SLE-DAS disease activity status.
Results The study population included 300 patients (female = 86%; mean age = 48.4±14.5 years; mean disease duration =14.1±9.3 years). The proportion of patients in clinical remission was 76% as defined by the DORIS and Doria criteria. Patients in clinical remission according to the SLE-DAS definition exactly matched those defined by either Doria or DORIS criteria, there being no discordant cases.
From patients in clinical remission, 18.4%, 92.5 %, and 30.4% were taking prednisone, antimalarials, and immunosuppressants, respectively.
In addition, the proportion of patients in LDA, mild disease activity and moderate-to-severe disease activity according to SLE-DAS were 9.7%, 6.7% and 7.7%, respectively (figure 1).
Conclusions In a real-life cohort of SLE patients, clinical remission is consistently defined by applying either

SLE-DAS, DORIS or the Doria criteria. Importantly, SLE-DAS definition is easier to apply, as it does not require the PGA or additional manifestations not included in SLEDAI. SLE presents a relapsing-remitting disease course despite maintenance treatment according to current good clinical practice. In accordance, in our cohort a few patients present moderate-to-severe activity and most patients achieved the target of remission, with <20% receiving prednisolone.

CO307 – PERFORMANCE OF MAJOR SALIVARY GLAND ULTRASOUND FOR THE DIAGNOSIS OF SJÖGREN'S SYNDROME IN CLINICAL PRACTICE

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Background: There is no specific test to diagnose Sjögren's Syndrome (SjS). Available classification criteria intended for research purposes help guide the diagnosis. Major salivary gland ultrasound (MSGUS) is an easily accessible, non-invasive, non-ionizing and inexpensive exam that may contribute to SjS diagnosis. Several available MSGUS scores have shown good diagnostic value.

Objectives: To study the performance of MSGUS for the clinical diagnosis of SjS and its relationship with other relevant markers of disease.

Methods: We included patients with a clinical suspicion of SjS that underwent MSGUS evaluation at our Department. Parotid and submandibular glands were classified on a 0-4 scale, based on tissue inhomogeneity (hypoechoic areas of variable size). Four rheumatologist ultrasonographers classified each gland at bedside. The score of the highest graded gland was considered and a score ≥2 was defined as a positive MSGUS. Anti-SSA, minor salivary gland biopsy (MSGB) and the diagnosis of all patients were reviewed. MSGB was considered positive if a focus score ≥1 was reported. We compared MSGUS findings across diagnoses and against MSGB result and anti-SSA status. Categorical variables were compared using the chi-square test. Continuous variables were compared using Student's t-test, Mann-Whitney U test, or ANOVA. P-value was considered significant at <0.05.

TABLE 1 – GENERAL CHARACTERISTICS OF THE SAMPLE

	Overall (n=219)	pSjS (n=131)	sSjS (n=17)	nSSS (n=62)	UCTD (n=9)	p-value
Age, years (mean ± sd)	60 ± 14	58 ± 17	56 ± 19	60 ± 13	48 ± 16	0.07
Female, n (%)	210 (95.9)	127 (97.0)	15 (88.2)	59 (95.2)	9 (100)	<0.001
Anti-SSA positive, n (%)	105 (47.9)	91 (69.5)	10 (5.8)	3 (4.8)	1 (11.1)	<0.001
Positive MSGB, n (%)	85 (46.4)	69 (63.3)	7 (46.7)	8 (15.7)	1 (12.5)	<0.001
MSGUS score, mean ± sd	1.70 ± 1.09	1.98 ± 1.09	1.76 ± 1.11	1.13 ± 1.09	1.44 ± 1.12	<0.001
Grade 0, n (%)	35 (16.0)	17 (13.0)	3 (17.7)	15 (24.2)	0 (0.0)	
Grade 1, n (%)	62 (28.3)	25 (19.1)	4 (23.5)	27 (43.6)	6 (66.7)	
Grade 2, n (%)	62 (28.3)	39 (29.8)	4 (23.5)	17 (27.5)	2 (22.2)	<0.001
Grade 3, n (%)	54 (24.7)	44 (33.6)	6 (35.3)	3 (4.9)	1 (11.1)	
Grade 4, n (%)	6 (2.7)	6 (4.6)	0 (0.0)	0 (0.0)	0 (0.0)	
MSGUS score ≥ 2, n (%)	122 (55.7)	89 (67.9)	10 (58.8)	20 (32.3)	3 (33.3)	<0.001

MSGB – minor salivary gland biopsy; MSGUS – major salivary gland ultrasound

Results: A total of 219 patients were included, 96% of which were female, with a mean age of 60 ± 13.6 years-old (Table 1). MSGUS was positive in a higher proportion of patients with primary and secondary SjS, in comparison to non-Sjögren's sicca syndrome (nSSS) and undifferentiated connective tissue disease (UCTD; p<0.001). In addition, mean MSGUS score was higher in SjS patients in comparison to nSSS and UCTD (p<0.001). A positive MSGUS was associated with anti-SSA positivity (61.4% vs 38.6% in anti-SSA negative patients; p<0,001). Patients positive for anti -SSA had a higher mean MSGUS score (2.12 ± 1.03) in comparison with anti-SSA negative patients (1.31 ± 0.97, p<0.001). Finally, while MSGUS and MSGB positivity were not significantly associated (p=0.237), the mean MSGUS score was higher in patients with a positive MSGB (1.95 ± 1.17) in comparison with those with a negative MSGB (1.61 ± 0.99; p=0.027).

Conclusions: MSGUS is a valuable resource in the investigation of SjS. It is associated with clinical diagnosis, anti-SSA and MSGB findings. Together with these other complementary exams, it can be used to assist in the diagnosis of patients presenting with sicca features suggestive of SjS.

CO323 – IDENTIFICATION OF MUSCLE ASSOCIATED KEY GENES TO SUPPORT AXIAL SPONDYLOARTHRITIS DIAGNOSIS BY TRANSCRIPTOMIC APPROACH, THE MYOSPA STUDY

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Background: Early diagnosis of axial Spondyloarthritis (axSpA) represents a major clinical challenge nowadays. Increasing evidence has determined that early diagnosis, prompt treatment initiation and early achievement of remission are the best predictors of long-term clinical, functional and radiographic outcomes. New tools to support the diagnosis are needed.
Objectives: This study aims to identify differentially expressed genes that may improve the current clinical diagnosis approach for early axSpA.
Methods: A cross-sectional study was conducted on 50 participants, 25 patients with axSpA (according to ASAS criteria) and 25 Healthy Controls, matched by gender, age and levels of physical activity. Peripheral blood samples were collected and RNA-Seq technology was performed. Normalization of raw data, and identification of differentially expressed genes was obtained using edgeR and limma-voom R packages. Gene Set Enrichment Analysis (GSEA) and Functional Enrichment analysis using Gene ontology (GO) and Kyoto Encyclopedia of Genes and Genomes (KEGG) annotations were also performed. A number of Differently Expressed Genes were highlighted.
Results: 311 genes were identified as being significantly differentially expressed between patients and controls. In details, 129 downregulated (7 genes have fold change more than 1) and 182 upregulated genes (3 genes have fold change more than 1) are highlighted. These genes are mostly involved in Myogenesis, Innate Immune Signalling and JAK/STAT pathways. Several genes with functions of skeletal muscle development and muscle contraction were identified.
Conclusions: The evidence disclosed that regulation of muscle development and contraction may be also engaged in physiopathology mechanisms of axSpA. These new cues open new perspectives for diagnosis and therapeutic approaches in axSpA.

C0324 – PROTEIN BIOMARKERS MAY DIFFERENTIATE RESPONDERS AND NON-RESPONDERS TO ADALIMUMAB, A TUMOUR NECROSIS FACTOR INHIBITOR, IN ANKYLOSING SPONDYLITIS (AS) PATIENTS – A BIOEFFICACY STUDY

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Background: Axial Spondyloarthritis (axSpA) is amongst the most common forms of inflammatory arthritis. Widely used in the treatment of axSpA, adalimumab is an engineered antibody holding only human peptide sequences that binds with high affinity and specificity to soluble and transmembrane TNFα, blocking its interaction with receptors p55 and p75. However, the response to TNFi is heterogeneous and might be linked with serious side effects. A priori identification of patients more propense to respond to TNFi is critical in clinical practice.
Aim: The aim of this work was to identify proteins for which the level of variation is tightly associated with the clinical response to TNFi (adalimumab) in radiographic-axSpA (r-axSpA/AS) patients.
Methods: The proteomic analysis involved 33 patients with r-axSpA/AS (19 responders and 14 non-responders) during a 14 weeks treatment with adalimumab. Serum samples were collected at baseline (before starting treatment), 3-5 days, 2 weeks and 14 weeks (W14) after treatment. Response to adalimumab was defined as the achievement of ASAS20. The experimental workflow combined immunoaffinity depletion of the high-abundant serum proteins, tryptic digestion of the depleted serum, MS/MS based identification and MS quantification of the detected proteins. Protein quantification was carried out to further normalize the amount of proteins to be analysed in the various samples. Uni- and multivariate statistical analysis of the differentially abundant proteins was

used to select the more specific protein biomarkers. Function protein association network analysis was performed with STRINGdb.

Results: LC-MS/MS method allowed the identification of 333 proteins with at least 2 non-ambiguous peptides. A set of new putative biomarkers was identified with 8 proteins displaying differences between responders and non-responders ($p < 0.05$) at baseline. Of these, 3 proteins were highly linked with a good clinical response at W14, while the other 5 proteins were strongly related with a non-response to adalimumab therapy. These proteins were measured simultaneously and confirmed to be predictive of response to treatment with an area under the Receiver Operating Characteristics (ROC) curve of 1. Additionally, it was possible to identify, for each phenotype, a unique protein profile when likening baseline with W14. Responders showed a significant increase in W14 of proteins related to inflammatory response and defence response, whereas W14 non-responders displayed an increase of platelet degranulation, acute inflammatory response, cell activation, very-low-density lipoprotein particle assembly and high-density lipoprotein particle remodelling process. The same comparison revealed that the differential proteins that decreased in responders in W14 were related to transport and regulation of response to external stimuli while in non-responders the differential proteins that diminished at W14 were associated with processing and regular turnover of intracellular proteins, collagen metabolism, calcium membrane-binding proteins, organization of the desmosomal cadherin-plakoglobin complexes and mediation of cell-cell adhesion.

Conclusions: Proteomic approaches constitute a very promising strategy to the identification of biomarkers for a particular therapy response. These results provide evidence that a panel of biomarker proteins might be able to predict the response to adalimumab therapy in r-axSpA/AS patients, even before the beginning of treatment.

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CO356 – CORRELATION BETWEEN ULTRASOUND FINDINGS AND CLINICAL AND LABORATORY DISEASE ACTIVITY MEASURES IN RA PATIENTS

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Introduction: Musculoskeletal ultrasound assessment is more sensitive than clinical examination to detect joint and soft-tissue inflammation. This image technique could add valuable information to guarantee the best possible outcome for patients with inflammatory articular diseases. Our aim was to examine the relationship between global synovitis/tenosynovitis/bursitis score and different variables used to measure disease activity.

Methods: Cross-sectional study including patients with Rheumatoid Arthritis (RA) fulfilling the ACR/EULAR 2010 classification criteria. Sociodemographic data, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), patient and physician global assessment (PGA and PhGA, respectively) were collected at the time of clinical visit. A blinded 44-joint clinical and US examination were performed in the same day by different rheumatologists. The presence of synovitis (synovial hypertrophy (SH) and power Doppler (PD), regardless of the presence of joint effusion) for each of the 22-paired joints comprised in the disease activity 44 was graded on a 0-3 point scale according to the OMERACT definitions, as well as 17-paired tendon sheaths and 3-paired bursas.(1) Composite synovitis, tenosynovitis and bursitis scores were calculated as the sum of all joints, tendon sheaths and bursas,

TABLE 1. CORRELATION BETWEEN ULTRASOUND FINDINGS AND CLINICAL AND LABORATORY FINDINGS (N=130 PATIENTS; N= 5720 JOINTS)

	Spearman correlation coefficient (r_s), p			
Variable	Composite synovitis score	Composite tenosynovitis score	Composite bursitis score	Global ultrasound score
44-TJC	r_s = 0.22, p= 0.012	NA	NA	r_s = 0.24, p= 0.006
44-SJC	r_s = 0.39, p<0.001	NA	NA	r_s = 0.43, p<0.001
ESR	r_s = 0.004, p= 0.962	r_s = 0.11, p= 0.205	r_s = 0.10, p= 0.276	r_s = 0.01, p= 0.892
CRP	r_s = 0.08, p= 0.380	r_s = 0.29, p= 0.001	r_s = 0.01, p= 0.910	r_s = 0.113, p= 0.202
PGA	r_s = 0.07, p= 0.438	r_s = 0.14, p= 0.118	r_s = 0.12, p= 0.173	r_s = 0.11, p= 0.209
PhGA	r_s = 0.53, p<0.001	r_s = 0.47, p<0.001	r_s = -0.04, p= 0.67	r_s = 0.57, p<0.001

CRP – C-reactive protein, ESR – Erythrocyte sedimentation rate, NA – Not applicable, PGA – patient global assessment, SJC – Swollen joint count, TJC – Tender joint count

respectively. Global US score (GUSS) was defined as the sum of all composite scores. Correlation between US findings and clinical and laboratory variables was determined using Spearman correlation coefficient (r_s). r_s values < 0.30 were considered poor, 0.30-0.59 fair, 0.60-0.79 moderate, ≥ 0.80 very strong. Variables with p<0.05 or considered clinically relevant were included in multiple linear regression analysis to identify predictors for GUSS. A p£0.05 was considered statistically significant.

Results: We included 130 RA patients (84% female, mean age of 63.1±12.2 years). We evaluated 5720 different joints (1300 metacarpophalangeal/proximal interphalangeal/metatarsophalangeal joints each; 260 shoulder/acromioclavicular/sternoclavicular/elbow/wrist/knee/tibiotalar joints each). The correlation level between US findings and clinical and laboratory disease activity measures is shown in Table 1. In multivariate analysis, only 44-tender joint count (β -0.43, 95% CI -1.71- (-0.52), p=0.00) and 44-swollen joint count (β 0.81, 95% CI 2.09-3.73, p=0.00) were associated to a higher GUSS.

Conclusion: GUSS had a poor correlation with tender joint count and a fair correlation with swollen joint count and PhGA. No correlation was found with PGA and ultrasound scores. In multivariate analysis, TJC and SJC remained as independent predictors of GUSS.

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CO358 – IMPACT OF THE MANDATORY CONFINEMENT DUE TO THE SARS-COV-2/ COVID-19 PANDEMIC IN PORTUGUESE PATIENTS WITH RHEUMATOID ARTHRITIS: RESULTS FROM THE COVIDRA SURVEY

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Introduction: As a result of the new SARS-CoV-2 pandemic, the state-of-emergency was declared in Portugal from the 18th March until the 3rd May, and included mandatory confinement as a containment strategy to control disease spread. As in most countries, healthcare resources were allocated to fight this pandemic at the expense of chronic disease management. The aim of this study was to evaluate the self-reported impact of the mandatory confinement in Portuguese patients with rheumatoid arthritis (RA), as a means to improve care in the pandemic era and to prepare for a likely second wave.

Patients and methods: The web-based survey COVIDRA was developed to assess 5 domains including RA symptoms, attitudes towards medication, employment status, physical exercise and mental health. The questionnaire was sent to RA patients through e-mail and social media of the Portuguese Society of Rheumatology, the National Association of Patients with RA and the Portuguese League Against Rheumatic Diseases. The questionnaire was also filled locally at two rheumatology centers in Lisbon. Recruitment took place during June and July 2020.

Results: We obtained 441 valid questionnaires. Most respondents were female (88.4%, N=388), caucasian (93.6%, N=410) with a mean age of 58 (+/-13) years. The majority (57.6%, N= 252) had longstanding disease (>10 years) and were treated with csDMARDs (63.2%, N=277) and bDMARDs/tsDMARDS (23,7%, N=104). Over 40% (N=178) experienced symptom worsening during confinement, almost half (17,8%, N=78) considered moderate or severe. Mobility restriction (34,0%, N=60) and increased stress, anxiety or depression (27,5%, N=49) were pointed out as the causes for this worsening. Only 2,5% (N=11) reduced or withheld their immunosuppressive medication due to fear of becoming infected with SARS-CoV-2. Another 2,5% (N=11) did so because they had no prescription, couldn't go to the community/hospital pharmacy or couldn't afford the medication. After confinement, 16,2% (N=32) of those previously employed were in a lay-off regime and 3% (N=6) lost their jobs. Most employed RA patients practiced telework during confinement (55,4%, N=87). The majority of patients decreased or did not practice any physical exercise (80,5%, N=350). Symptoms of anxiety and depression developed or worsened in 67,3% (N=297) and 51,9% (N= 228) respectively, approximately one third were moderate or severe.

Discussion: Literature on the patient-reported impact of the COVID-19 pandemic in rheumatic conditions is scarce. Cleaton *et al* found significantly worse HRQoL in rheumatic patients undergoing stringent self-isolation compared to those who do not. Seyahi *et al* assessed the psychological state of Turkish rheumatic patients but found lower rates of anxiety and depression compared to ours. Schmeiser *et al*, Pineda-sic *et*

al and Fragoulis *et al* reported very similar rates to ours of immunosuppressive discontinuation due to fear of being infected.

Conclusion: Portuguese RA patients experienced significant symptom worsening, anxiety and depression during state-of-emergency confinement. Only a minority changed their immunosuppressive treatment for fear of SARS-CoV-2 infection.

CO361 – AVALIAÇÃO DOS ÍNDICES DE ATIVIDADE DE DOENÇA (DAS) E HAQ EM DOENTES COM ARTRITE REUMATÓIDE SOB ETANERCEPT VS ETANERCEPT BIOSSIMILAR: ESTUDO DE VIDA REAL DO REUMA.PT

VAZ CC, INÊS L, CANHÃO H

Introdução: Apesar dos ensaios clínicos demonstrarem a equivalência dos produtos biológicos originais e biossimilares, os estudos de vida real são fundamentais para a decisão terapêutica na prática clínica. Os objetivos deste estudo foram comparar os índices de atividade da doença (DAS) e HAQ ("patient reported outcome": Health Assessment Questionnaire) em doentes com Artrite Reumatóide, de uma base de registo nacional, sob etanercept original vs etanercept biossimilar e explorar as características à baseline destes doentes.

Métodos: Trata-se de um estudo observacional comparativo longitudinal que inclui todos os doentes adultos com Artrite Reumatóide registados no reuma.pt que se encontravam sob terapêutica com etanercept original ou etanercept biossimilar à data da exportação dos dados em fevereiro de 2020. As avaliações entre o DAS e o HAQ em cada grupo foram feitas à baseline, aos 6 meses, 1 e 2 anos. Frequências, médias e desvios padrão (d.p.) foram calculados para descrever as características à baseline da amostra. A distribuição normal das variáveis foi examinada com o teste de Kolmogorov-Smirnov. Os valores descritivos foram comparados com o teste t de Student ou o teste U de Mann- Whitney, conforme apropriado, e a análise não paramétrica de amostras emparelhadas foi realizada usando o teste de Wilcoxon. As frequências foram comparadas usando o teste exato de Fisher, McNemar ou Qui-quadrado. Resultados significativos foram considerados quando p <0,05. Foi realizada a correlação de Pearson entre o DAS e o HAQ. Todas as análises foram realizadas por meio do Statistical Package for the Social Sciences (SPSS) para Mac, versão 26.0 (SPSS Inc., Chicago, IL, EUA).

Resultados: Foram incluídos um total de 6703 doentes na base de dados. Destes, 3070 doentes (45,8% do total) tinham o diagnóstico de Artrite Reumatóide. 1357 doentes encontravam-se sob etanercept (86,5%) e 212 sob etanercept biossimilar (13,5%). As características à baseline foram semelhantes entre os doentes tratados com o etanercept original em comparação com o biossimilar correspondente (Tabela 1). Em termos de atividade da doença (Tabela 2), o DAS à baseline e aos 6 meses foi diferente entre os dois grupos com significado estatísticamente significativo (respectivamente, p <0,001 e p=0,029), mas ao 1 ano e 2 anos os valores médios do DAS mostraram atividade da doença semelhante para os dois grupos. Em relação ao HAQ (Tabela 3), à baseline, foi diferente entre os dois grupos com significado estatísticamente significativo (p <0,001), mas não se verificou diferença com

TABELA 1 – CARACTERÍSTICAS *BASELINE* DOS DOENTES COM ARTRITE REUMATÓIDE SOB TERAPÊUTICA COM ETANERCEPT ORIGINAL OU BIOSSIMILAR

	Etanercept (n=1357)	Biossimilar (n=212)
Sexo n (%)		
Feminino	1153 (85,0)	165 (77,8)
Masculino	204 (15,0)	47 (22,2)
Duração da doença até introdução de biológico (anos) mediana (min-máx)	5,97 (0,01-50,46)	6,02 (0,05-48,70)
Morbilidades n (%)		
HT	293 (21,6)	27 (10,6)
A	91 (6,7)	11 (4,3)
DM	73 (5,4)	7 (2,7)
Dislipidemia	80 (5,9)	3 (1,2)
DCV		

HTA: Hipertensão Arterial; DM: Diabetes Mellitus; DCV: Doença Cardiovascular

TABELA 2 – DAS DE DOENTES COM ARTRITE REUMATÓIDE SOB ETANERCEPT VS. BIOSSIMILAR

DAS (média±d.p.)	Etanercept	Biossimilar	*P*
T0	5,38±1,32	4,50±1,54	<0,001
6m	3,63±1,40	3,37±1,35	0,029
1a	3,51±1,44	3,34±1,22	0,170
2a	3,37±1,27	3,04±0,99	0,155

DAS: Disease Activity Score; T0: Baseline; 6m: seis meses; 1a: um ano; 2a: dois anos

TABELA 3 – HAQ DE DOENTES COM ARTRITE REUMATÓIDE SOB ETANERCEPT VS. BIOSSIMILAR

HAQ (média±d.p.)	Etanercept	Biossimilar	*P*
T0	1,38±0,66	1,13±0,67	<0,001
6m	0,99±0,70	1,01±0,67	0,809
1a	0,97±0,66	0,99±0,71	0,810
2a	0,90±0,68	0,93±0,63	0,839

HAQ: Health Assessment Questionnaire; T0: Baseline; 6m: seis meses; 1a: um ano; 2a: dois anos

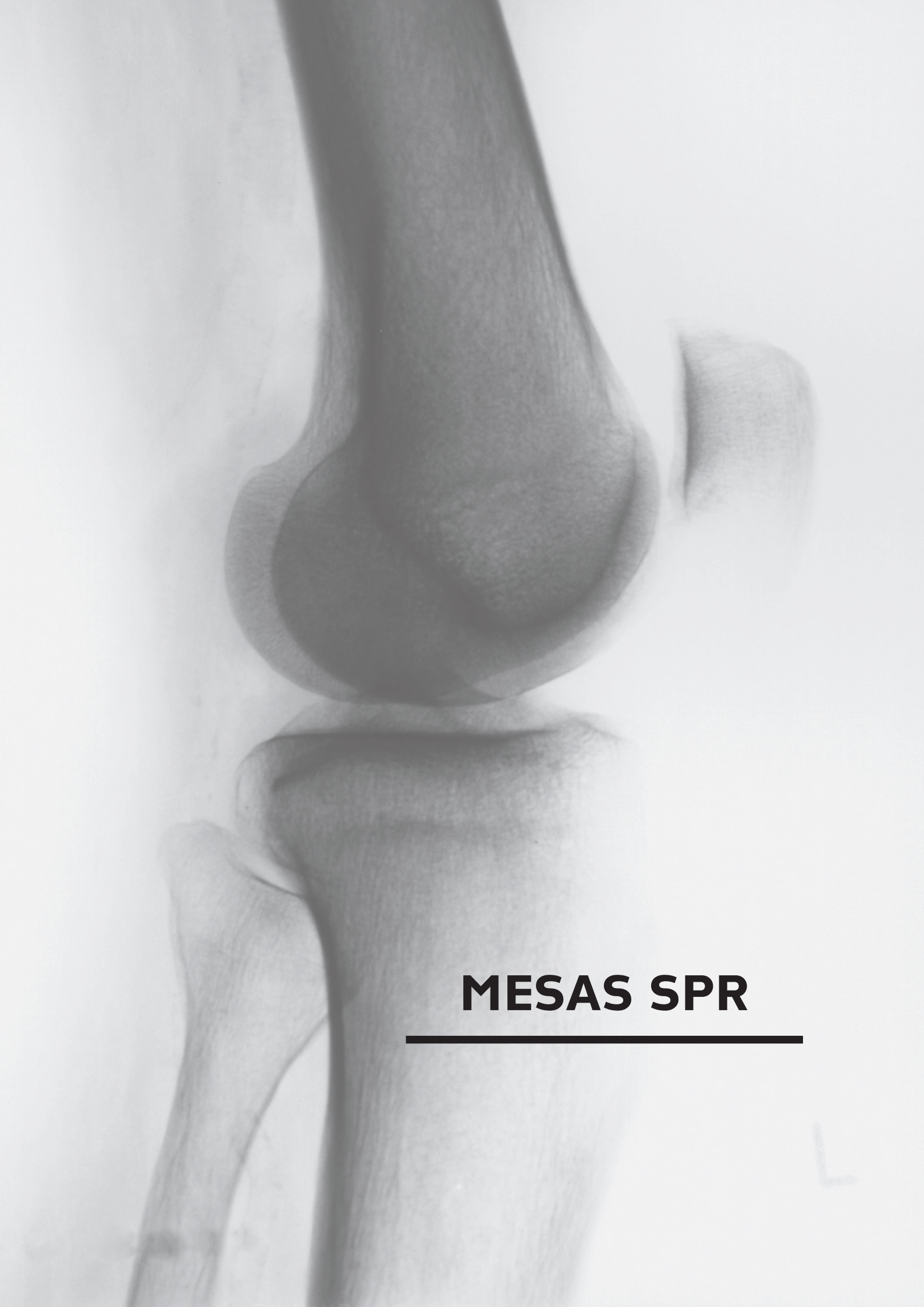
TABELA 4 – DAS VS. HAQ

	R	*p*
T0	0,53	<0,001
6m	0,55	<0,001
1ª	0,55	<0,001
2ª	0,53	<0,001

DAS: Disease Activity Score; HAQ: Health Assessment Questionnaire; T0: Baseline; 6m: seis meses; 1a: um ano; 2a: dois anos.

significado estatístico aos 6 meses, 1 ano e 2 anos. O DAS mostrou uma correlação linear moderada com o HAQ com significado estatístico à baseline, aos 6 meses, 1 e 2 anos (p< 0,001) – Tabela 4.

Conclusão: As semelhanças nas medidas de atividade média da doença (DAS) ao 1 ano e 2 anos em doentes tratados com o original versus biossimilar sugerem efetividade comparável na prática clínica. Para além da avaliação clínica expressa no DAS, o "patient reported outcome" HAQ corrobora a efetividade sob o ponto de vista do doente. Estes dados de vida real têm uma relevância significativa pois permitem aumentar a confiança na decisão terapêutica do biossimilar etanercept na artrite reumatóide.



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DMARDS SINTÉTICOS E BIOLÓGICOS: USO "OFF LABEL" EM IDADE PEDIÁTRICA

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Crianças e adolescentes com doenças reumáticas sistémicas necessitam frequentemente de terapêuticas imunossupressoras que modifiquem a atividade da doença reumática (DMARDs), sejam eles fármacos sintéticos ou biológicos.
A maioria dos DMARDs usados neste grupo de doentes, carece de documentação quanto a dados de eficácia, de segurança e dosagem nesta faixa etária, devido sobretudo às dificuldades inerentes à realização de ensaios clínicos pediátricos. Como resultado, a utilização "off-label" deste tipo de fármacos é comum nas crianças com doença reumática sistémica. A sua forma de administração e indicações terapêuticas são habitualmente extrapoladas de estudos realizados em adultos com artrite reumatoide ou com outras formas de artrite inflamatória.
Vários DMARDs biológicos foram aprovados nas últimas duas décadas para o tratamento de algumas doenças reumáticas pediátricas. Contudo, nem todas as crianças têm apresentado uma resposta clínica adequada aos medicamentos atualmente aprovados. A experiência publicada com fármacos considerados "off label" em crianças pode ser útil na tomada de decisão, embora esses relatos publicados não reduzam a necessidade de ensaios clínicos prospetivos na população pediátrica. A decisão relativamente à terapêutica mais adequada deve sempre basear-se na melhor evidência disponível e no binómio risco/benefício para cada criança.

DMARDS SINTÉTICOS E BIOLÓGICOS: USO "OFF LABEL" EM IDADE ADULTA

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Muitos DMARDs utilizados em Reumatologia foram importados de outras indicações e, de forma semelhante, várias terapêuticas com indicação primária em patologia reumática são correntemente utilizadas *off label* em indicações distintas das originais, com distintos graus de evidência subjacente.
Focando as indicações *off label* na idade adulta na Reumatologia, no caso dos DMARDs sintéticos, aponta-se o recurso ao metotrexato na espondilartrite periférica (com exceção da indicação na artrite psoriática); em várias conectivites como o lupus eritematoso sistémico (LES), síndrome de Sjogren (SS), esclerose sistémica (SSc) e miopatias inflamatórias; na doença de Still (AOSD) e nas vasculites de grandes e pequenos vasos. A sulfassalazina é frequentemente utilizada *off label* no tratamento da artrite psoriática e outras espondilartrites com envolvimento periférico. A leflunomida tem sido usada em doentes com AIJ, miosites refratárias e vasculites de pequenos vasos. Nas vasculites de grandes e pequenos vasos, a azatioprina também é utilizada *off label* como poupador de corticóide. O uso da ciclosporina, por seu turno, foi estendido ao LES, miopatias inflamatórias, doença de Behçet e AOSD.
Entre os fármacos biológicos, o rituximab destaca-se pela multiplicidade de utilizações *off label*: LES, SS, SSc, miopatias inflamatórias e crioglobulinémia. Os antiTNF têm sido utilizados com sucesso no tratamento da sarcoidose e doença de Behçet. O tocilizumab demonstrou benefício no tratamento da policondrite recidivante, AOSD, polimialgia reumática refratária e esclerose sistémica.
Sublinha-se que vários DMARDs têm vindo também a ser utilizados com eficácia em doenças dermatológicas, neurológicas, ofatlmológicas e hematológicas.

DISTÚRBIOS DO SONO E DOENÇAS REUMÁTICAS: COMO GERIR?

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Os distúrbios do sono são frequentes em doentes com patologia reumatológica quer pela elevada prevalência de algumas perturbações do sono nesta população, como a Síndrome de Apneia Obstrutiva do Sono (SAOS) ou a Síndrome de pernas inquietas, quer por se associarem a queixas álgicas que por si só condicionam alteração da estrutura do sono e diminuição da sua eficiência. A abordagem do sono torna-se por isso fundamental e deve ser incluída na avaliação global do doente. Existem várias ferramentas para a avaliação do sono, incluindo questionários sobre sonolência diurna, qualidade do sono, questionários de

auto-preenchimento sobre sintomas subjetivos de sono e questionários de rastreio para SAOS validados para populações específicas. Estas ferramentas devem ser aplicadas para a decisão da marcha diagnóstica e para a referenciação a consulta especializada de medicina do sono. A identificação de um distúrbio do sono e início do tratamento adequado e atempado tem impacto na qualidade de vida e nas comorbilidades, incluindo no controlo das doenças reumatológicas. Como identificar os distúrbios do sono e quando referenciar a consulta de patologia do sono será o tema da "Distúrbios do Sono e doenças reumáticas: Como gerir?"

DRUG-INDUCED RHEUMATIC AND MUSCULOSKELETAL DISEASES

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Drug-induced rheumatic and musculoskeletal disorders (RMDs) comprise a wide group of diseases and syndromes that affect the musculoskeletal and connective tissue, following exposure to a given drug. These important clinical entities are often difficult to recognise, as patients are commonly treated with a variety of medications, direct causal attribution is not always possible, and they often closely mimic the features of its 'primary' counterparts. As such, a high degree of suspicion, a detailed clinical history, and the acquaintance with classic and emerging drug-induced RMDs are paramount to allow for accurate identification and prompt treatment of these conditions. The spectrum of drug-induced RMDs is wide, with a plethora of clinical manifestations, and a multitude of culprit drugs. As new treatments emerge, it is important to acknowledge novel clinical entities, described on an ever more frequent basis. Drug-induced RMDs can be broadly divided into several groups, ranging from severe life-threatening disease to mild soft-tissue rheumatisms: (i) inflammatory and non-inflammatory arthropathies; (ii) connective tissue diseases (systemic lupus erythematosus, myositis and other myopathies, scleroderma and related fibrosing syndromes); (iii) vasculitis; (iv) metabolic bone disorders; and (v) periarticular soft-tissue rheumatisms. Specific entities include the autoimmune/autoinflammatory syndrome induced by adjuvants (ASIA) and the more recently defined immune-related adverse events of checkpoint inhibitor cancer immunotherapy. Recognition of the clinical picture of drug-induced RMDs is fundamental for appropriate treatment and, if indicated, early drug withdrawal.

ARTRITE REUMATÓIDE: GESTÃO DO DOENTE EM REMISSÃO

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A Artrite Reumatoide (AR) é uma doença inflamatória crónica com um elevado impacto na capacidade funcional, qualidade de vida e mortalidade dos doentes. Ao longo dos últimos anos, o diagnóstico precoce, o aparecimento de novos e mais eficazes fármacos, associados a uma estratégia *Treat-to-Target* têm contribuído para um melhor prognóstico destes doentes.
A monitorização da atividade com utilização de índices validados é, desta forma, imperativa. Os diversos índices de atividade incluem parâmetros objetivas como articulações dolorosas e tumefactas e parâmetros laboratoriais (VS ou PCR) e a avaliação da atividade da doença na perspetiva do doente (PGA), tendo todos eles critérios de remissão estabelecidos. De acordo com os critérios ACR/EULAR, remissão é ainda definida como articulações dolorosas e das articulações tumefactas (em 28 articulações), PCR (mg/dl) e PGA ≤1, constituindo a definição mais estrita de remissão.
Contudo diversos estudos têm demonstrado que uma grande proporção de doentes não atinge o estado de remissão apenas por elevado PGA, apesar da aparente ausência objetiva de inflamação, levando recentemente a que a definição de remissão tenha sido questionada.
Uma vez atingida a remissão, a descontinuação de terapêutica imunossupressora tem sido equacionada de modo a evitar um sobretratamento. Diversos ensaios clínicos e estudos observacionais têm sido desenvolvidos de modo a definir a melhor estratégia de descontinuação de terapêutica imunossupressora em doentes em remissão sustentada.
Ao longo desta apresentação irão ser apresentadas as limitações as implicações clínicas das atuais definições de remissão, assim como a abordagem do doente em remissão sustentada.

GESTÃO DO DOENTE EM REMISSÃO – ESPONDILARTRITES

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O aperfeiçoamento dos índices de atividade e dos critérios de remissão é uma realidade que tem cruzado de forma transversal grande parte das áreas da Reumatologia. A definição de remissão é, em particular, de extrema importância na identificação dos doentes a quem o Reumatologista pode poupar da escalada terapêutica. Uma definição desadequada deste conceito pressupõe consequências nefastas ao implicar sobretratamento ou controlo desadequado da doença.
Na área das espondilartrites, a definição de doença inativa *pelo Ankylosing Spondylitis Disease Activity Score (ASDAS)* tem sido cada vez mais utilizada, tanto em ensaios clínicos como em estudos observacionais. Valores de ASDAS inferiores a 1,3 demonstraram ter bom desempenho e validade externa. Felizmente, o número de doentes em remissão parece ter vindo a aumentar nas últimas décadas, muito provavelmente resultado do advento de fármacos biotecnológicos. Contudo, não é no dia em que o doente atinge a remissão que se despede do seu Reumatologista. Este marco é sim o início de uma nova etapa terapêutica.
Após identificação dos doentes em remissão sustentada, importa então saber se poderemos de forma segura diminuir a carga terapêutica. A decisão deve ser partilhada entro o clínico e o doente, após a explicação dos riscos e benefícios da mesma.
Contudo, mesmo nos doentes com remissão sustentada, a quem foi possível diminuir significativamente a carga terapêutica, o clínico deve manter-se atento a possíveis recaídas. A promoção de práticas de estilo de vida saudável e atividade física regular devem continuar a ser prioridades. A existência de outras comorbilidades é outra realidade que não deve ser esquecida no seguimento destes doentes.

GESTÃO DO DOENTE EM REMISSÃO – ARTRITE IDIOPÁTICA JUVENIL

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A artrite idiopática juvenil (AIJ) engloba um grupo heterogéneo de patologias que se iniciam antes dos 16 anos de idade e que cursam com artrite com duração superior a 6 semanas, cuja etiologia permanece ainda por esclarecer.
O seu curso natural é caracterizado por períodos de atividade e de quiescência com duração imprevisível. O tratamento, que deve ser individualizado, inclui o uso de anti-inflamatórios não esteroides (AINEs), tratamentos locais com corticoide, DMARDs clássicos ou biotecnológicos, de acordo com, entre outros fatores, a gravidade, o número de articulações afetadas e a presença ou ausência de manifestações extra-articulares.
A disponibilidade de novos fármacos, conduziu a uma melhoria significativa do prognóstico e permitiu o controlo da actividade da doença na grande maioria das crianças. Quando a remissão clínica é atingida com o tratamento com AINEs e/ou com tratamentos locais, parece consensual manter apenas uma vigilância clínica e analítica regular. No entanto, quando esta remissão é obtida com o recurso a DMARDs, clássicos ou biológicos, permanece a dúvida de qual o momento ideal para descontinuar estes fármacos e de como fazê-lo, traduzindo-se em grande heterogeneidade de prática clínica. Apesar de existirem poucos estudos relativamente a este tema, nesta apresentação serão abordados e discutidos os dados existentes na literatura.

GESTÃO DO DOENTE COM LÚPUS ERITEMATOSO SISTÉMICO EM REMISSÃO

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Nos últimos anos, com o advento de novas terapêuticas, a remissão tornou-se um aspeto central na gestão dos doentes com patologias reumáticas inflamatórias.
No Lúpus Eritematoso Sistémico (LES), várias definições de remissão têm sido propostas. Atualmente a mais aceite foi publicada em 2016 por um grupo de trabalho internacional liderado por van Vollenhoven (*Definition of Remission in SLE, DORIS*).
A gestão terapêutica dos doentes com LES em remissão, particularmente aqueles que se encontram em remissão prolongada, tem sido alvo de ampla investigação. Os benefícios da redução e, se possível, suspensão dos glucocorticóides têm sido demonstrados em diversos estudos. Por sua vez, a redução e/ou suspensão de imunossupressores deve ser avaliada e ponderada de forma individualizada. É importante salientar que a prescrição universal de antipalúdicos, bem como a adoção de medidas não farmacológicas preventivas do agravamento da atividade do LES (suspensão do tabagismo e proteção à exposição de radiação UV) assumem particular importância nestes doentes. Durante a redução/suspensão terapêutica deve ser mantida vigilância frequente, com o intuito de detetar precocemente sinais ou sintomas de ressurgimento da atividade da doença. Apesar do LES se encontrar em remissão, mantém-se a necessidade de

uma monitorização clínica e laboratorial.
Nesta palestra será apresentada a evidência das estratégias de gestão terapêutica nos doentes com LES em remissão e proposto um algoritmo de gestão terapêutica.

DOR E INFLAMAÇÃO: ANTI-INFLAMATÓRIOS NÃO ESTEROIDES (AINE)

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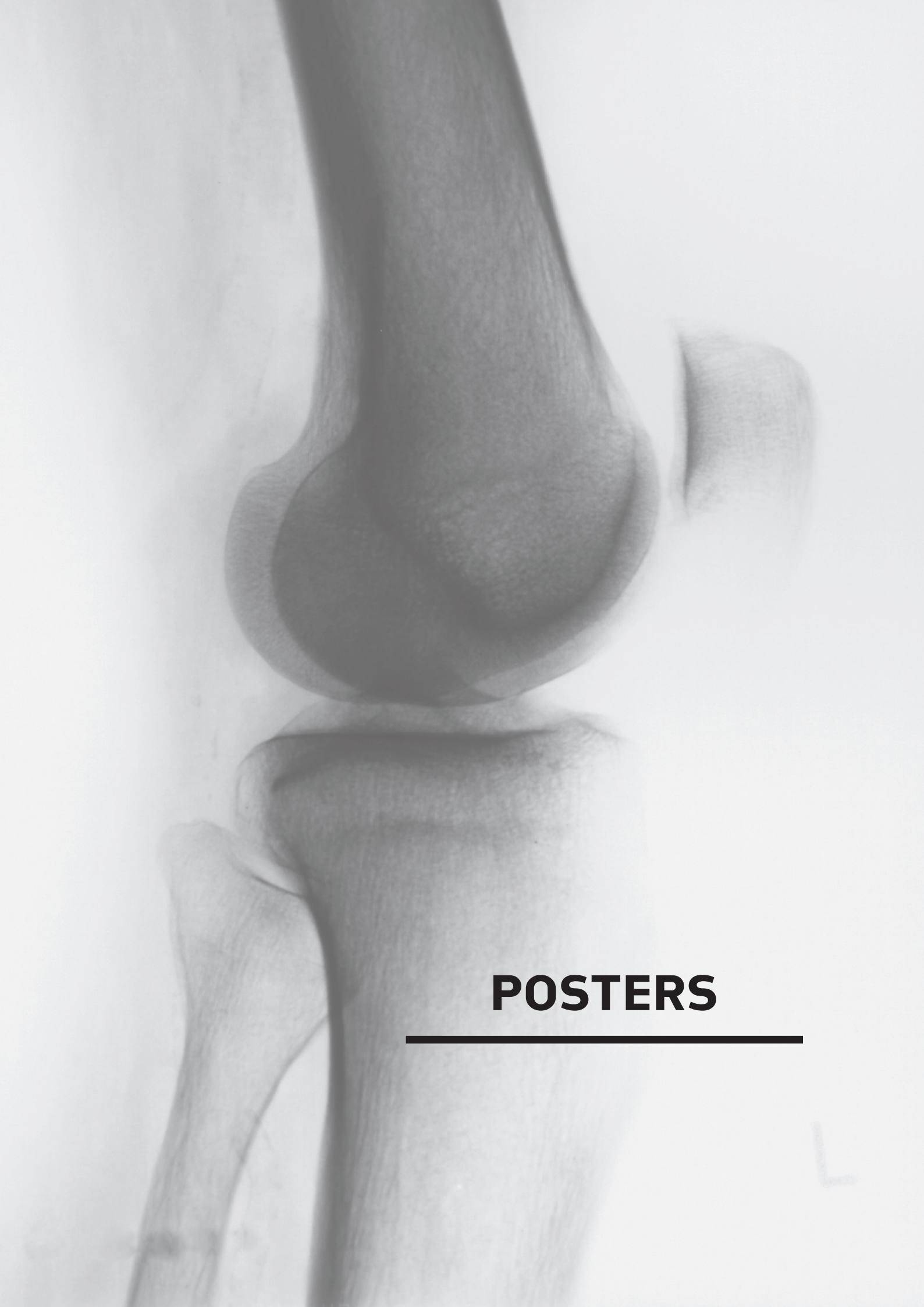
Os AINEs continuam sendo dos fármacos mais prescritos na prática médica. Apesar dos efeitos adversos que podem determinar, múltiplos e de gravidade variável, os seus efeitos analgésico, antipirético e anti-inflamatório estão na base da sua larga utilização.
Nas doenças reumáticas continuam a ser uma arma fundamental, usados quer em situações agudas quer crónicas, de modo pontual ou mais continuado, por períodos e em esquemas posológicos variáveis, tendo em conta a patologia em causa e o indivíduo em particular.
Estratégias de minimização de risco são usadas por forma a obter uma boa relação benefício/risco e utilidade. Novas evidências no conhecimento das doenças reumáticas, contribuem para a adopção de distintas estratégias terapêuticas, permitindo minimizar a exposição aos AINE, pela obtenção de remissão sustentada de muitas doenças reumáticas, nomeadamente inflamatórias sistémicas.
Contudo, o seu efeito multifactorial sobre a (in) capacidade funcional, constitui em grande medida a base da sua preferência pelos próprios doentes. Vem já do século passado, e permanece actual o debate sobre a escolha, o posicionamento, a segurança e, não menos importante, a adesão e os índices de satisfação dos doentes, em relação a analgésicos versus anti-inflamatórios.
Sendo que me não parece haver necessidade de fazer uma avaliação dicotómica desta problemática mas, de forma mais propulsiva, tentar usar essas armas terapêuticas, (e outras farmacológicas ou não) de modo racional e coerente, segundo a melhor evidência do conhecimento actual das distintas patologias, visando obter o resultado desejado: combatend a disfunção, preservar a capacidade e a participação.



POSTERS

Posters

P0009 – EFFECTIVENESS OF SWITCHING BETWEEN TNF INHIBITORS IN PATIENTS WITH AXIAL SPONDYLOARTHRITIS: IS THE REASON TO SWITCH RELEVANT?

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Background/Purpose: Over the last years, and mostly due to lack of alternatives, it has been common practice to start a second TNF inhibitor (TNFi) in patients with axial spondyloarthritis (axSpA) who discontinue their first TNFi. Evidence informing on the effectiveness of this strategy in clinical practice is limited. Importantly it remains unclear whether the reason for discontinuation of the first TNFi influences the response to the second. We aimed to assess whether the reason of discontinuation of the first TNFi influences the response to the second TNFi.

Methods: Patients with axSpA from the ReumaPt national registry, who discontinued their first TNFi and started a second TNFi were included in this analysis. In addition, patients were required to have complete data on Ankylosing Spondylitis Disease Activity Score (ASDAS) and Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) at baseline, 3 and 6 months after starting the first TNFi. Afterwards, patients were followed every 6 months up to 12 years. The main outcome was the ASDAS clinically important improvement (ASDAS CII). Secondary outcomes were ASDAS major important improvement (ASDAS MI); ASDAS low disease activity (ASDAS LDA); ASDAS inactive disease (ASDAS ID) and BASDAI 50. The reason for discontinuation of the first TNFi was defined as: i) Primary failure, if ASDAS CII was not achieved at 3 or 6 months; ii) Secondary failure if ASDAS CII was achieved at 3 or 6 months but lost in ≥1 visit during follow-up; iii) Adverse events; iv) Other (e.g. pregnancy, surgery). The response to the first TNFi at 3 and 6 months was compared to the response to the second TNFi at the same visits, adjusting for age, gender and C-reactive protein (CRP). The association between the reason of discontinuation of the first TNFi (predictor) and response the second TNFi over time was tested in generalized estimating equations (GEE) models, adjusted for age, gender and CRP.

Results: In total, 193 patients (53% male, mean age 45 (SD:11) years) were included, with a median follow-up time on the second TNFi of 1.5 years. Patients had a lower response to the second TNFi compared to the first TNFi according to the main outcome (ASDAS CII) at 3 months (41% vs 51%) and 6 months (35% vs 56%). There was no association between the reason to discontinue the first TNFi and response to the second TNFi as defined by ASDAS CII (Table). This association was present for the most stringent outcomes, namely ASDAS MI and ASDAS ID. Compared to patients who discontinued their first TNFi due to primary failure, patients were more likely to achieve ASDAS ID with the second TNFi when they discontinued their first TNFi due to secondary failure (OR: 7.3 [(95%CI: 1.9; 27.7]), adverse events (OR: 9.1 [2.5; 33.3]), or other reasons (OR: 7.7 [1.6; 37.9]).

TABLE – ASSOCIATION BETWEEN THE REASON FOR DISCONTINUATION OF THE FIRST TNFI AND RESPONSE TO THE SECOND TNFI

Reason to discontinue first TNFi*	Outcome for the second TNFi OR (95% CI)				
(ref Primary failure)	ASDAS-CII (N=135)	ASDAS-MII (N=135)	ASDAS-LDA (N=166)	ASDAS-ID (N=166)	BASDAI50 (N=147)
-Secondary failure	1.9 (0.7;4.8)	**4.8 (1.3;18.2)**	1.2 (0.6;2.4)	**7.3 (1.9;27.7)**	1.4 (0.6;3.0)
-Adverse events	1.5 (0.6;3.5)	2.4 (0.6;9.6)	0.9 (0.5;1.7)	**9.1 (2.5;33.3)**	1.1 (0.5;2.3)
-Other	1.0 (0.3;3.8)	1.7 (0.1;19.4)	1.0 (0.4;2.4)	**7.7 (1.6;37.9)**	0.5 (0.1;1.7)

*GEE models with the reason of discontinuation of the first TNFi as predictor (reference category: primary failure); all models adjusted for age, gender and C-reactive protein. OR in bold are statistically significant (p<0,05).

Conclusion: In patients with axSpA, response to the second TNFi is worse compared to the first TNFi. The reason to discontinue the first TNFi seems to influence the response to the second TNFi. Patients with a secondary failure to the first TNFi have a better response to the second TNFi compared to those discontinuing the first TNFi due to a primary failure, particularly when response is defined by the most stringent outcomes.

P0010 – SYSTEMIC SCLEROSIS – ARE PATIENTS WITH CALCINOSIS DIFFERENT FROM THOSE WHO DO NOT HAVE IT?

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Background: Systemic Sclerosis (SS) is a heterogenous disease with a broad range of organ involvement. Calcinosis is a common problem and although it may affect almost any body tissue, it is typically seen in the limbs. Its presence relates with higher risk of digital ulcers and infection. It is still unknown whether patients with calcinosis also have other clinical features that differentiate them from the remaining.

Objectives: To determine the prevalence of calcinosis in a SS cohort and to evaluate if its presence relates with specific clinical features.

Methods: A cross-sectional study was conducted evaluating a cohort of SS patients. Plain radiographs were taken to assess calcinosis at elbows, hands, knees and feet. Clinical data was obtained and analyzed using IBM SPSS Statistics 26®.

Results: We included 25 patients, 21 females [n= 21 (84%)], median (min, max) age was 58 (27, 75) years-old. Regarding disease classification, 16 (64%) had limited SS, 4 (16%) had diffuse SS, 3 (12%) had overlap syndrome and 2 (8%) had early SS. Ten (40%) patients had radiological calcinosis in at least one site, seven of which (70%) were subclinical. The most affected areas were knees and hands [n=6 (24%)]. Table 1 summarizes the clinical characteristics of patients with and without calcinosis. Limited SS was significantly more prevalent in the calcinosis group [n=9 (90%) vs. n=7 (46.7%), p=0.04]. All patients had Raynaud phenomenon [n=10 (100%) vs. 15 (100%)]. Current or past digital ulcers [n=5 (50%) vs. n=6 (40%), p=0.697], telangiectasias [n=9 (90%) vs. n=11 (73.3%), p=0.615], pulmonary hypertension [n=2 (20%) vs. n=1 (6.7%), p=0.550] and esophageal involvement [n=6 (60%) vs. n=6 (40%), p=0.428] were more frequent in the calcinosis group but with no statistical significance. Although late capillaroscopic pattern was more frequent in the calcinosis group, there was no statistical significance difference [n=4 (40%) vs. n=1 (6.7%), p=0.121]. Seropositivity for centromere-B antibodies was more frequent in the calcinosis group but with no statistical significance [n=7 (70%) vs. n=8 (53.3%), p=0.678].

TABLE 1: DEMOGRAPHIC AND CLINICAL DATA OF PATIENTS WITH AND WITHOUT CALCINOSIS

Demographic and clinical data	Calcinosis (n=10)	No calcinosis (n=15)	p-value
Female gender, n (%)	9 (90)	12 (80)	0.626
Age (years), median [min,max]	68.5 [27, 75]	52 [36, 73]	0.129
Cutaneous classification			
» Limited, n (%)	9 (90)	7 (46.7)	0.04
» Diffuse, n (%)	1 (10)	3 (20)	0.626
» Early, n (%)	0 (0)	2 (13.3)	0.500
» Overlap, n (%)	0 (0)	3 (20)	0.250
Clinical manifestations			
» Raynaud phenomenon, n (%)	10 (100)	15 (100)	-
» Current or previous digital ulcers, n (%)	5 (50)	6 (40)	0.697
» Interstitial lung disease, n (%)	2 (20)	4 (26.7)	1.000
» Pulmonary hypertension, n (%)	2 (20)	1 (6.7)	0.550
» Arthritis, n (%)	2 (20)	3 (20)	1.000
» Calcinosis, n (%)	3 (30)	0 (0)	0.052
» Esophageal involvement, n (%)	6 (60)	6 (40)	0.428
NFC patterns			
» Non specific abnormalities, n (%)	1 (10)	3 (20)	0.626
» Early scleroderma, n (%)	1 (10)	1 (6.7)	1.000
» Active scleroderma, n (%)	3 (30)	10 (58.8)	0.111
» Late scleroderma, n (%)	4 (40)	1 (6.7)	0.121
Autoantibodies			
» Centromere B, n (%)	7 (70)	8 (53.3)	0.678
» Scl-70, n (%)	1 (10)	4 (26.7)	0.615

Conclusions: The prevalence of calcinosis was similar to that reported in literature (18-49%). This study confirmed the association, already found in previous studies, between calcinosis and the limited form of SS and raises attention for the importance of calcinosis radiographic screening since there was a high prevalence of subclinical calcinosis. Although there were some clinical differences between patients with and without calcinosis, given the small cohort, statistical significance was not obtained. Larger studies are needed to increase statistical power.

P0011 – THE IMPACT OF SYSTEMIC SCLEROSIS ON BODY IMAGE PATIENTS

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Introduction: Satisfaction with body image has a major impact in quality of life. Systemic sclerosis (SSc) is a can result in disfiguring physical changes. Our aim was to determine the impact of systemic sclerosis

on body image using the Satisfaction with Appearance Scale (SWAP). (1)

Methods: Cross-sectional study including patients satisfying the 2013 American College of Rheumatology criteria for SSc diagnosis, aged ≥ 18 years, treated in a tertiary Rheumatology Department. Demographic and clinical data were collected from Reuma.pt and clinical records. All patients provided informed consent and fulfilled SWAP questionnaire, which consists of 14 questions in 4 subscales: satisfaction with facial appearance, satisfaction with non-facial appearance, social discomfort due to appearance and perceived social impact of appearance. Patients rate each item on a numerical rating scale from 1 (strongly disagree) to 7 (strongly agree). Scores for the facial and non-facial appearance range from 0-24 and scores for the social discomfort and perceived social impact subscales range from 0-18. Total SWAP score can range from 0-84 and higher values indicate greater dissatisfaction with appearance and poorer body image. A descriptive analysis was used to summarize demographic and clinical data; categorical variables were described using frequencies; and continuous data using mean and standard deviation. Correlation between variables [Rodnan, age, disease duration, Hospital Anxiety and Depression Scale (HADS) and Short Form Health Survey (SF36)] and SWAP score was tested with Pearson or Spearman coefficient, as appropriated. Scores of SWAP and its subscales in preclinical, limited and diffuse forms of SSc were compared using ANOVA test. Analyses were performed with SPSS Statistics, V.21 and $p<0.05$ was considered statistically significant.

Results: We enrolled 38 patients, 84.2% (n=32) female, with mean age 60.3±14.5 years and mean disease duration 13.3±6.5 years. All but one were caucasian. Fifty percent (n=19) had a limited form, 26.3% (n=10) had preclinical scleroderma and 23.7% (n=9) had a diffuse form of SSc. Regarding the autoantibody profile: 63.2% (n=24) had anti-centromere antibodies, 28.9% (n=11) had anti-Scl-70 antibodies, 5.3% (n=2) had anti-PM antibodies and 2.6% (n=1) had no positive antibodies. The median of Rodnan scores was 4 (IQR 0-9). The total mean SWAP score was 44.8±12.5 with worse results at "Satisfaction with facial appearance" subscale (mean score 14.4±6.1). There is no statistically significant difference in the SWAP score (or its subscales) between the three diagnosis subtypes. No statistically significant correlation was found between the total and subscale SWAP scores and any of the continuous variables considered and no statistically significant difference was found between the different forms of SSc.

Conclusion: We found no significant differences between preclinical, limited or diffused SS. SWAP scores were not significantly correlated with the total Rodnan score, age or disease duration. Contrary to our expectations SWAP did not show any relationship with depression, anxiety (HADS) or quality of Life (SF-36) However, our sample is too small to support definite conclusions. Further studies assessing body image in SSc and its impact in quality of life are warranted to support the holistic care of these patients.

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P0013 – IMPACTO SOCIO-ECONÓMICO DAS FALTAS NÃO JUSTIFICADAS NA CONSULTA DE REUMATOLOGIA

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Introdução: "Absentismo à consulta" é o termo utilizado para definir o fenómeno em que utentes com consulta agendada não comparecem à mesma na data, hora ou local correctos, sem pré-aviso da sua não comparência. A não comparência às consultas médicas reduz a eficiência dos serviços, condiciona importante impacto financeiro e prejudica a saúde do doente, além de poder atrasar o acesso a cuidados médicos aos utentes em lista de espera. Em Portugal, os dados relativos ao absentismo à consulta médica, e em particular entre doentes com patologia reumática, são bastantes escassos.

Objectivos: Avaliação e caracterização da prevalência de absentismo nas consultas de Reumatologia relativamente a variáveis sociodemográficas e clínicas e estimativa do impacto económico.

Metodologia: Revisão de um período mensal de consulta externa de Reumatologia Geral. Foram analisadas as consultas efectivadas e as consultas agendadas, mas não efectivadas (por falta, sem aviso prévio). Os custos directos das consultas não efectivadas foram estimados conforme o estabelecido no "Acordo Modificativo ao Contrato-Programa" para o hospital.

Resultados: Foram incluídos na análise 982 agendamentos decorridos durante o mês de Janeiro de 2018. Agendamentos para prescrição de terapêutica, relatórios e internamento programado foram excluídos. Verificaram-se 57 faltas, correspondendo a 5,8% do total de agendamentos. As consultas subsequentes

representaram 85,2% dos agendamentos efectivados e 80,7% das consultas não efectivadas por falta, correspondendo a 7,4% faltas à primeira consulta e 5,5% de faltas nas consultas consequentes. O sexo feminino foi o mais representado em ambos os grupos – 620 (67,0%) entre as consultas efectivadas e 37 (65,0%) na subamostra das faltas. A média de idades foi de 57±15 anos no primeiro grupo e 54±16 anos no segundo. A escolaridade média foi de 8±5 anos no grupo das consultas efectivadas e de 9±6 anos no grupo das faltas. Não se verificaram diferenças significativas entre grupos relativamente ao sexo, idade, escolaridade, diagnóstico, duração e actividade da doença. De igual modo, não houve diferenças quanto à modalidade de consulta (primeira ou subsequente). Estimou-se um valor de 2 438 euros em agendamentos não efectivados, o que poderá representar um custo de mais de 29 000 euros anuais, apenas em custos directos, sem entrar em linha de conta com a necessidade de remarcação de meios complementares de diagnóstico.

Conclusão: A falta à consulta poderá ser dependente de outros factores além do sexo, idade e escolaridade dos doentes. O impacto social e económico dos agendamentos não efectivados é indiscutível. Acrescem aos custos estimados neste estudo, os custos indirectos, como o potencial agravamento do estado de saúde do doente, a dificuldade de acesso aos cuidados de saúde e a penalização do sistema hospitalar. Torna-se pois necessária e urgente a implementação de medidas de sensibilização da população visando a optimização e efectividade do actual sistema de saúde.

P0014 – IMPORTÂNCIA DO DIAGNÓSTICO DIFERENCIAL DA ARTRITE SÉPTICA – REVISÃO A 5 ANOS DA ABORDAGEM DO DOENTE EM CONTEXTO DE URGÊNCIA

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Introdução: A artrite séptica (AS) constitui uma importante emergência médica, associando-se a elevada morbilidade. A patologia articular prévia (p.e., artrite reumatóide, osteoartrose, artropatia microcristalina) condiciona um risco séptico superior. A apresentação aguda da artropatia gotosa pode ser indistinguível da apresentação da AS e a sua existência não exclui uma AS concomitante, impondo dificuldades acrescidas no diagnóstico diferencial.

Objectivos: Caracterização sociodemográfica e clínica dos doentes avaliados por suspeita de artrite séptica no Serviço de Urgência (SU) entre 2014 e 2019.

Metodologia: Estudo transversal, retrospectivo, com recolha de dados sociodemográficos e clínicos relativos aos doentes observados no SU de Ortopedia por suspeita de AS entre Abril/2014 e Setembro/2019.

Resultados: Foram incluídos 110 doentes, maioritariamente do sexo masculino (n=61; 55,5%), com uma idade mediana de 70 (DIQ 20) anos. A HTA (50%), a dislipidemia (35,5%) e a diabetes mellitus (28,2%) foram as comorbilidades mais frequentes. Trinta e seis doentes (32,7%) apresentavam antecedentes de hiperuricemia/gota, ou tiveram este diagnóstico estabelecido de novo aquando do episódio. A monoartrite foi a apresentação mais comum (96,4%), sendo o joelho a articulação mais frequentemente envolvida (54,5%). O S. aureus foi o microorganismo identificado em maior percentagem no exame cultural do líquido sinovial (LS), embora as culturas de LS tenham sido positivas em apenas 50% dos casos. Nos casos com identificação de agente, a Proteína C Reativa (PCR) sérica e a contagem total de células (CTC) do LS foram significativamente mais elevadas (U=824,5; p=0,001 e U=73,5; p=0,026, respectivamente). A PCR sérica revelou-se um factor preditivo do isolamento de agente etiopatogénico, com valores ≥ 17,6 mg/dl a apresentar uma sensibilidade e especificidade de cerca de 60% e 77%, respectivamente (IC 95%=0,522 – 0,798). Doentes com hiperuricemia/gota apresentaram maior probabilidade de obterem um exame cultural de LS negativo (OR = 4,71 [IC 95% =1,92 -11,54]). As características bioquímicas do LS, a leucocitose e o valor de PCR não foram significativamente diferentes entre doentes com e sem história de hiperuricemia/gota.

Conclusão: Os indivíduos idosos e com múltiplas comorbilidades, nomeadamente factores de risco cardiovascular, são mais susceptíveis à ocorrência de AS. A hiperuricemia/gota é comum nos doentes abordados no SU por suspeita de AS. O valor de PCR parece ser o factor preditivo mais importante para a detecção de agente etiopatogénico. Os parâmetros bioquímicos do LS e os parâmetros inflamatórios séricos não parecem ser úteis na distinção entre a artropatia gotosa e AS. A maior probabilidade da obtenção de culturas de LS negativas nos doentes com hiperuricemia/gota deve alertar-nos para a possibilidade do diagnóstico erróneo de artrite séptica em doentes com episódios de artropatia gotosa aguda.

P0016 – PATIENT-PHYSICIAN DISCORDANCE IN ASSESSMENT OF DISEASE ACTIVITY IN RHEUMATOID ARTHRITIS

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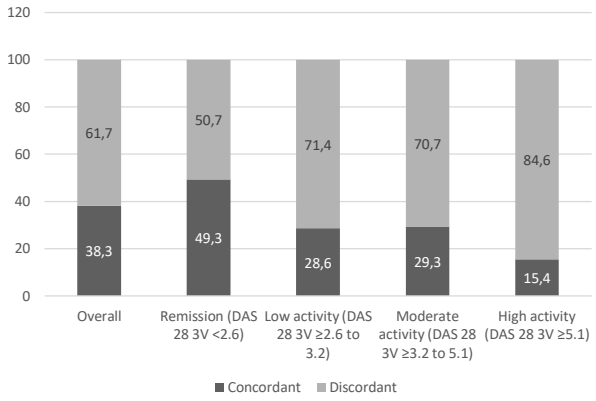
Background: In rheumatoid arthritis (RA), global disease activity is commonly assessed, from the patient's and the physician's perspective, through a 100mm VAS. Previous studies have commonly shown a considerable discrepancy between the patient's and physician's assessment. This study aimed evaluating patient-physician discordance in the assessment of disease activity and to explore its determinants.

Methods: Cross sectional study including RA patients (ACR/EULAR 2010 classification criteria), aged ≥ 18 years, followed in a single tertiary centre. Data were collected from the most recent evaluation including sociodemographic features, disease duration (years), disease activity (DAS 28 3V-PCR), tender and swollen joint count 0-28 (TJC and SJC), VAS-pain-patient, patient and physician global assessment (PGA and PhGA respectively), erythrocyte sedimentation rate (ESR), C-reactive protein (CPR), Health assessment questionnaire (HAQ) and EuroQol five-dimension scale (EQ5D). The discrepancy between patients and physicians (ΔPPhGA) was defined as PGA minus PhGA, and a difference > |20mm| was considered as "discordant". A descriptive analysis was performed and variables described as proportions or means (+/- SD), as adequate. Correlation between ΔPPhGA and other variables was assessed through Pearson's correlation and comparison between groups through t-test. Variables with p<0.05 or otherwise considered clinically relevant were included in multiple linear regression analysis to identify predictors for ΔPPhGA. A p£0.05 was considered statistically significant.

Results: In total, 467 patients with RA were included (81.2% female; mean age 63.9% ± 12.2 years). PGA and PhGA were discordant in 61.7% of the cases, the patient scoring higher than the physician in 95% of these cases. The proportion of concordance increased (p < 0.01) when considering only patients in remission (DAS 28 3V <2.6), (Graph 1). ΔPPhGA was moderately correlated with VAS-pain-patient (r = 0.59) and weakly correlated with SJC (r = -0.12), HAQ (r= 0.27), EQ5D (r = -0.28) and age (r = 0.21);

GRAPH 1. CONCORDANCE LEVEL (%) BETWEEN PGA AND PHGA ACCORDING TO THE DISEASE ACTIVITY



all p<0.01. In multivariate analysis, VAS-pain-patient (β 0.74, 95% CI 0.62-0.88, p=0.00) and TJC (β 0.16, 95% CI 0.45-0.48, p=0.02) remained associated with a higher ΔPPhGA.

Conclusion: Our study confirmed that a significant discrepancy between patients and physicians in the assessment of global disease activity is frequent in clinical practice, probably due to valorization of different parameters. This was much less pronounced among patients in remission. Higher VAS-pain-patient and TJC were independent predictors of greater discrepancy between patients and physician's assessment.

P0017 – AGREEMENT BETWEEN REFERRING PHYSICIANS AND RHEUMATOLOGISTS AND PREDICTORS OF INFLAMMATORY ARTHRITIS: ANALYSIS BASED ON 8 YEARS OF EXPERIENCE IN AN EARLY ARTHRITIS CLINIC

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Introduction: Early recognition of patients with arthritis is a crucial opportunity for optimal outcome. The Early Arthritis Clinic (EAC) of our department was created in 2012 to ensure a prompt access of these patients to efficient medical care. Patients may be referred based on a set of clinical criteria with less than 12 months duration and laboratory parameters: arthritis, inflammatory arthralgias, squeeze test, morning stiffness > 30 minutes, rheumatoid factor

GRAPH 1. CONCORDANCE LEVEL (%) BETWEEN PGA AND PHGA ACCORDING TO THE DISEASE ACTIVITY

Referral criteria	Kappa	p
Arthritis	0.23	0.05
Squeeze test	0.09	0.04
Inflammatory arthralgias	0.11	0.04
Morning stiffness	0.18	0.04
RF	0.27	0.04
ESR	0.26	0.04
CRP	0.25	0.04
ANA	0.02	0.47

ANA – antinuclear antibodies; CRP – C-reactive-protein; ESR – erythrocyte sedimentation rate; RF – Rheumatoid factor

(RF), erythrocyte sedimentation rate (ESR)>30mm/h and C-reactive-protein>0.5mg/dL (CRP).

Objectives: To assess the level of agreement between the referring physician and the rheumatologist, regarding the presence of each of the six referral criteria and to identify predictors of inflammatory arthritis.

Methods: Cross sectional study including patients aged ≥ 18-year-old observed in the EAC between January 2012 and October 2019. Subjects who were referred to the EAC by a rheumatologist and those without available referral letter/medical records from the first visit to the EAC were excluded. Demographic data, provenience, referral criteria (presence/absence) and the final diagnosis [presence or not of an inflammatory rheumatic disease (IRD)] were collected from medical records. For the six referral criteria, the agreement between the referring physician and the rheumatologist was assessed using the Cohen's Kappa. The presence of each referral criteria was compared between patients with and without an IRD using $\chi 2$ tests. Variables with $p<0.1$ or clinically relevant were included in forward stepwise multivariable logistic regression analysis to identify possible predictors for IRD. The statistical analysis was performed using SPSS® v21 and $p<0.05$ was considered statistically significant.

Results: 376 patients (70% female; mean age (±SD) 56.3±16.2 years) were included. Most patients were referred from primary care (84%); the remaining 16% include those referred from emergency department and other hospital specialties. We diagnosed an inflammatory arthritis in 62% (n = 232) of the patients. Table 1 shows the level of agreement between the referring physician and the rheumatologist, regarding the presence of the referral criteria.

In univariable analysis (IRD Vs non-IRD), inflammatory arthralgias (74% Vs 93%, p=0.01), squeeze test (24% Vs 55%, p=0.01), morning stiffness (49% Vs 63%, p=0.05), ESR (63% Vs 46%, p=0.01), CRP (62% Vs 48%, p=0.04) were associated to IRD. In multivariable analysis, only ESR (OR 5.0 [95% CI 1.9-13.0], $p < 0.05$) and inflammatory arthralgias (OR 0.15 [95% CI 0.04-0.52], $p < 0.05$) remained as predictors of IRD.

Conclusion: Agreement between the referring physicians and the rheumatologist regarding then presence/absence of the referral criteria was poor in all clinical criteria and fair in laboratory criteria. Elevated ESR was an independent predictor of IRD and the description of inflammatory arthralgias was negatively correlated with IRD. These findings suggest the need to clarify the referral criteria used and to improve education among the physicians referring patients to the EAC.

P0018 – COMPARING PATIENT-PHYSICIAN DISCORDANCE IN RA AND PSA PATIENTS

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Introduction: Patient global Assessment (PGA) of disease activity is considered a key patient reported outcome in Rheumatoid Arthritis (RA) and Psoriatic Arthritis (PsA), both being included in combined indices of disease activity. However, patients and physicians frequently disagree in their assessment. This study aimed at comparing the degree of this discrepancy and its determinants in RA and PsA.

Methods: Cross sectional study including 100 patients with RA (ACR/EULAR 2010 criteria) and 100 patients with PsA with predominant peripheral joint involvement (CASPAR criteria), aged ≥18 years, randomly selected from the electronic registry Reuma.pt. Data were collected from the most recent rheumatology visit during the last year: sociodemographic data, disease duration (years), tender and swollen joint counts 0-28 (TJC and SJC), disease activity (DAS28 3V-PCR), erythrocyte sedimentation

rate (ESR), C-reactive protein (CRP), patient's pain assessment, PGA and physician global assessment (PhGA). The discrepancy between patients and physicians (ΔPPhGA) was defined as PGA minus PhGA, and a difference > |20mm| was taken as "discordance". Categorical variables are presented as proportions and continuous variables as mean (±SD). Patient and clinical characteristics were compared between patients with RA and PsA using t- test and $\chi 2$ test, as adequate. Variables with $p<0.05$ or clinically relevant were included in multivariable logistic regression analysis to identify correlates for ΔPPhGA in the whole sample. A p£0.05 was considered statistically significant.

Results: Compared to PsA, patients with RA were more often female (90% Vs 49%, $p < 0.05$), older (66.7 ± 10.7 Vs 58.3 ± 12.2 years, $p < 0.05$) and had a shorter disease duration (18.2 ± 9.8 Vs 19.9 ± 9.7 years, $p = 0.202$). Regarding disease activity, the RA and PsA groups were comparable: DAS28 3V-PCR (2.3 ± 0.9 Vs 2.4 ± 1.0, $p = 0.34$). Patients with RA had a higher mean ΔPPhGA (30.4 ± 30.6 Vs 25.4 ± 27.5, $p < 0.05$), and were more frequently discordant to the physician (69% Vs 51%, $p < 0.05$). In univariable analysis, having RA, higher patient's pain assessment and higher ESR were associated to patient-physician discordance. In multivariable analysis, only patient's pain assessment (OR 1.04 [95% CI 1.03-1.06], $p = 0.00$) and TJC (OR 0.82 [95% CI 0.68-0.97], $p = 0.02$) remained as predictors of discordance.

Conclusion: Despite comparable disease activity scores in RA and PsA patients, RA patients tend to have a worst self-perception of their disease activity compared to their physician´s. Patient's pain assessment and TJC were the only predictors of patient-physician discordance, irrespective of the disease.

P0019 – THE IMPORTANCE OF A SYSTEMIC SCLEROSIS CLINIC IN A TERTIARY REFERRAL CENTER – A PORTUGUESE EXPERIENCE

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Background: Systemic sclerosis (SSc) is a rare systemic rheumatic disease (SRD) characterized by small vessel inflammation and fibrosis of skin and internal organs. Pulmonary and cardiac involvement contribute to both morbidity and mortality associated with the disease. A multidisciplinary approach with strict monitoring is therefore key to attain clinical success.

Objectives: To describe the organization and patient pathways of our SSc outpatient clinic.

Methods: Observational study using data extracted from Reuma.pt/SSc (a subset of the Rheumatic Diseases Portuguese Register). Data extracted included demographic variables and clinical and immunological manifestations. The disease was classified according to the 2013 ACR/EULAR criteria.

Experience: Our SSc clinic is managed by two dedicated Rheumatologists and up to two Rheumatology residents on a weekly basis, but it is a dynamic multidisciplinary clinic where various medical specialties collaborate closely. There are two associated subspecialty clinics (pulmonary hypertension and pulmonary fibrosis) where the Rheumatologists engage with pneumologists and cardiologists, allowing greater collaboration in the management of these patients. Patients' data is systematically registered in Reuma.pt/SSc as a part of the routine activity of this clinic, contributing to real-world data on SSc.

Results: A total of 220 patients were registered between July 2011 and June 2019. 196 (89.1%) were female, with a mean age of 58.9±14 years and a mean disease duration of 14.6±9 years. Ninety-seven patients (44.1%) had limited cutaneous SSc, 52 (23.6%) had diffuse cutaneous SSc, 35 (15.9%) had overlap SSc, 24 (10.9%) had preclinical SSc and 12 (5.4%) had SSc sine scleroderma. Raynaud phenomenon was present in 92% of the SSc patients and 40% had a history of digital ulcers. Gastrointestinal manifestations included esophageal dismotility in 39.5% of patients, gastric disease in 24.4% and intestinal involvement in 15.5%. Pulmonary involvement was found in 47.6% of SSc patients, heart disease in 43.6% and kidney involvement in only two patients. Antinuclear antibodies were positive in 92.2% of the patients, anti-centromere in 44.1%, anti-topoisomerase I antibodies in 39.1%, anti-U1RNP in 4.5% and only three patients had anti-PM-Scl and one had anti-RNA polymerase III. 31 patients were lost to follow-up and 32 died. 18 patients are currently being followed up in the pulmonary hypertension clinic and seven in the pulmonary fibrosis clinic.

Conclusion: The implementation of a standardized approach with regular multidisciplinary work has proven very helpful in evaluating patients with SSc. The continuous registry of patients in Reuma.pt/SSc has been essential for patient care, research and healthcare planning.

P0025 – DETERMINANTS OF PATIENT--PHYSICIAN DISCORDANCE IN GLOBAL ASSESSMENT IN SPONDYLOARTHRITIS

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Background: Patient's Global Assessment of Disease Activity (PtGA) and Physician's Global Assessment of Disease Activity (PhGA) are important measures in the evaluation of patients with Spondyloarthritis (SpA), but often provide discordant results.1 Both PtGA and PhGA are assessed as part of ankylosing spondylitis disease activity score (ASDAS), that is a measure of axial SpA disease activity endorsed by the Assessment of SpA International Society (ASAS) and Outcome Measures in Rheumatology.2,3 In peripheral SpA, although there are no formally validated indexes, the American College of Rheumatology (ACR) and Disease Activity Score 28 (DAS 28) response criteria have shown reliable discriminant characteristics and both include PtGA and PhGA.3 The lack of concordance between PtGA and PhGA may mislead treatment decisions, namely switches.

Objective: To assess the determinants of patient-physician discordance in SpA patients under biologic treatment.

Methods: Cross-sectional study, including 72 with SpA according ASAS criteria. Physicians' evaluation included comorbidities, parameters of inflammatory activity (erythrocyte sedimentation rate [ESR] and C-reactive protein [CRP], PhGA, ASDAS PCR and, DAS 28, and Participants completed patient-reported outcomes (PROs) and sociodemographic characteristics. For statistical analysis, SPSS was used and significance level was 2-sided $p<0.05$.

Results: Clinical and laboratory characteristics of patients are shown in table 1. PtGA and PhGA were significantly different (34.8±21.2 vs 7.8±12.5 mm, respectively, $p< .001$) and patient-physician discordance (ΔPtGA – PhGA) was 27.5±14.3 mm.

In peripheral SpA, patient-physician discordance had a correlation with patient age, Health Assessment Questionnaire (HAQ), Functional Assessment of Chronic Illness Therapy (FACIT), EuroQol-5 dimension (EQ5D), Short Form (36) Health Survey (SF-36), Hospital Anxiety and Depression scales (HADS), CRP, ESR, number of comorbidities and daily medication, and an association with employment status (employees had lesser discordance), anxiety/depression, fibromyalgia and osteoarthritis (OA). In multivariable analysis including employment status, SF-36, OA, number of comorbidities, and ESR (R2 adjusted= .505), the main predictors of patient-physician discordance were lower SF36, higher number of comorbidities and employment status.

In axial SpA, patient-physician discordance had a correlation with nocturnal back pain and total back pain VAS, FACIT, EQ5D, SF-36, HADS, Bath Ankylosing Spondylitis Functional Index (BASFI) and Bath Ankylosing Spondylitis Activity Index (BASDAI) scales, age, number of comorbidities and daily medication and an association with employment status (employees had lesser discordance), anxiety/depression and fibromyalgia. In multivariable analysis including employment status, SF-36, fibromyalgia, and number of comorbidities (R2 adjusted= .738), the main predictors of patient-physician discordance were lower SF36, higher number of comorbidities and concomitant diagnosis of fibromyalgia.

Neither for peripheral SpA nor for axial SpA an association with SpA subtype, HLA-B27 positivity, patient or physician gender, or patient education level was found.

TABLE 1: CLINICAL AND LABORATORY CHARACTERISTICS OF PATIENTS WITH SPONDYLOARTHRITIS.

	Spondyloarthritis
Age (years), *mean ± SD*	49.3 ± 10.6
Gender *%(n/N)*	Male: 61.1% (44/72)
Years from diagnosis, *mean ± SD*	10.4 ± 5.6
Biologic DMARD position, *%(n/N)*	1st: 84.7% (61/72) 2nd: 9.7% (7/72) Others: 5.6% (4/72)
Patient Global VAS, *mean ± SD*	34.8 ± 21.2
Patient pain VAS, *mean ± SD*	26.6 ± 28.1*
Patient back pain VAS, *mean ± SD*	27.5 ± 27.7**
Patient nocturnal back pain VAS, *mean ± SD*	26.2 ± 26.1**
Physician Global VAS, *mean ± SD*	7.8 ± 12.5
Patient-physician discordance *mean ± SD****	27.5 ± 14.3
Tender joints count *(n), median (IQR)*	0.0 (1.0)*
Swollen joints count *(n), median (IQR)*	0.0 (0.0) *
CRP (mg/dL), *median (IQR)*	0.30 (0.71)
ESR (mm/hr), *mean ± SD*	14.3 ± 16.5
HAQ, *median (IQR)*	0.7 (1.4)*
DAS28 4V, *mean ± SD*	2.3 ± 1.9*
BASDAI, *median (IQR)*	2.3 ± (4.0)**
BASFI, *median (IQR)*	2.4 ± (4.0)**
ASDAS PCR, *mean ± SD*	2.4 ± 1.1
Short Form (36) Health Survey (SF-36), *mean ± SD*	Physical functioning: 51.9 ± 27.0 Role limitations due to physical health problems: 47.5 ± 24.9 Pain: 42.6 ± 27.9 General health perceptions: 42.2 ± 15.7 Energy/fatigue: 38.1 ± 20.3 Social role functioning: 51.3 ± 35.1 Role limitations due to emotional problems: 52.5 ± 28.6 Mental health: 54.7 ± 21.3
FACIT, *mean ± SD*	35.6 ± 11.4
HADS, *median (IQR)*	Anxiety: 10.5 (6.8) Depression: 10 (5.3)
EQ5D, *mean ± SD*	0.3748 ± 0.3108
Comorbidities, *median (IQR)*	1 (3)
Mellitus Diabetes, *%(n/N)*	11.1% (8/72)
Depression/ Anxiety, *%(n/N)*	19.4% (14/72)
Osteoarthritis, *%(n/N)*	11.1% (8/72)
Fibromyalgia, *%(n/N)*	8.3% (6/72)
Osteoporosis, *%(n/N)*	2.8% (2/72)

Conclusions: This study shows the variability implied in patient-physician discordance. We have demonstrated that comorbidities, employment status, and other factors not directly related to the disease are determinants for the patient-physician discordance.

P0026 – DIFFERENCES AND DETERMINANTS OF PHYSICIAN´S AND PATIENT´S PERCEPTION IN GLOBAL ASSESSMENT OF RHEUMATOID ARTHRITIS

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Background: Patient's Global Assessment of Disease Activity (PtGA) and Physician's Global Assessment of Disease Activity (PhGA) are assessed as part of commonly used measures of disease activity in RA.1 Both are important measures in treat-to-target strategies in Rheumatoid Arthritis (RA), but often provide discordant results.2,3 This can provide an erroneous assessment of disease activity in patients under Biologic treatment and mislead treatment decisions, namely switches.

Objective: To assess differences and determinants of PtGA and PhGA in RA patients under biologic treatment.

Methods: Cross-sectional study, including 46 patients with RA diagnosed according to the ACR/EULAR criteria, under biologic treatment, consecutively evaluated in day-care unit. Participants completed patient-reported outcomes (PROs), including PtGA, and sociodemographic characteristics. Physicians collected comorbidities and parameters of inflammatory activity (erythrocyte sedimentation rate [ESR] and C-reactive protein [CRP]) and completed PhGA and disease activity score 28 with ESR (DAS28). SPSS was used for statistical analysis and significance level was defined as 2-sided $p<0.05$.

Results: Clinical and laboratory characteristics of patients are shown in table 1. PtGA and PhGA were significantly different (36.1±27.6 mm vs 8.7±14.2 mm, $p< 0.001$) and a positive discordance (PtGA>PhGA, more than 25mm in visual analogue scale [VAS]) was found in 54.3% of cases.

PtGA had a correlation with PROs (Pain VAS, 36-Item Short Form Health Survey [SF-36], Health Assessment Questionnaire [HAQ], Functional Assessment of Chronic Illness Therapy [FACIT], EuroQol [EQ5D] and Hospital Anxiety and Depression Scale [HADS]), CRP, tender and swollen joint counts and an association with comorbidities like fibromyalgia or osteoarthritis (OA). No association was found between PtGA and age, sex, education level, profession, employment status, extra-articular manifestations, positivity of rheumatoid factor, ESR, years of disease evolution or number of biologic treatments. In multivariable analyse including SF-36, CRP, tender joints count and OA (R2 adjusted= 0.672), the main predictors of PtGA were lower SF36, concomitant OA and higher CRP level.

PhGA had a correlation with PtGA, pain VAS, CRP, tender and swollen joints. No association was found between PhGA and patient or physician age, patient or physician sex, extra-articular manifestations, positivity of rheumatoid factor, ESR level, years of disease evolution or number of biologic treatments. In multivariable analysis including ESR, tender and swollen

TABLE 1: CLINICAL AND LABORATORY CHARACTERISTICS OF PATIENTS WITH RHEUMATOID ARTHRITIS

	Rheumatoid Arthritis
Age (years), *mean ± SD*	58.7 ± 12.3
Gender *%(n/N)*	Female: 69.6% (32/46)
Years from diagnosis, *mean ± SD*	14.7 ± 7.39
Biologic DMARD position, *%(n/N)*	1st: 58.7% (27/46) 2nd: 28.3% (13/46) Others: 13.0% (6/46)
Patient Global VAS, *median (IQR)*	40.0 (50.5)
Patient pain VAS, *median (IQR)*	31.0 (45.0)
Physician Global VAS, *median (IQR)*	0.0 (*15.0*)
Positive discordance *%(n/N)**	54.3% (35/66)
Tender joints *(n), median (IQR)*	0.0 (3.0)
Swollen joints *(n), median (IQR)*	0.0 (2.0)
CRP (mg/dL), *median (IQR)*	0.3 (0.9)
ESR (mm/hr), *median (IQR)*	14.0 (24.0)
HAQ, *median (IQR)*	1.0 (1.6)
DAS28 4V, *mean ± SD*	2.9 ± 1.9
SDAI, *mean ± SD*	6.6 ± 6.3
CDAI, *mean ± SD*	7.5 ± 7.3
Short Form (36) Health Survey (SF36), *mean ± SD*	Physical functioning: 49.5 ± 32.3 Role limitations due to physical health problems: 58.2 ± 30.3 Pain: 52.8 ± 26.3 General health perceptions: 41.2 ± 23.3 Energy/fatigue: 50.8 ± 23.3 Social role functioning: 66.0 ± 26.0 Role limitations due to emotional problems: 65.7 ± 30.9 Mental health: 62.5 ± 24.8
FACIT, *mean ± SD*	34.9 ± 10.3
HADS, *median (IQR)*	Anxiety: 7 (7) Depression: 6 (7.5)
EQ5D, *median (IQR)*	0.3248 (0.4462)
Comorbidities, *median (IQR)*	2 (3)
Mellitus Diabetes, *%(n/N)*	17.4% (8/46)
Depression/ Anxiety, *%(n/N)*	8.7% (4/46)
Osteoarthritis, *%(n/N)*	28.3% (13/46)
Fibromyalgia, *%(n/N)*	4.3% (2/46)
Osteoporosis, *%(n/N)*	15.2% (7/46)

VAS: Visual Analogic Scale; CRP: C-reactive protein; ESR: Erythrocyte Sedimentation Rate; HAQ: Health Assessment Questionnaire DAS 28: Disease Activity Score; SDAI: Simple Disease Activity Index ; CDAI: Clinical Disease Activity Index; EQ5D: EuroQol-5 dimension; FACIT: Functional Assessment of Chronic Illness Therapy; HADS: Hospital Anxiety and Depression; SD: Standard Deviation;

*Positive discordance: PtGA>PhGA, more than 25mm in VAS

joints count and CRP (R2 adjusted= .800), the main predictors of PhGA were swollen joint count and higher CRP level.
Conclusions: This study showed the variability implied on global assessment of RA activity. Overall PtGA is based on function and also in subjective and emotional experience of pain, whereas the PhGA is based on more objective measures, more related to disease activity.

P0028 – TIME-COURSE CHANGE OF NEUTROPHIL-LYMPHOCYTE AND MONOCYTE--LYMPHOCYTE RATIOS AND THE RESPONSE TO THE FIRST BDMARD IN A PORTUGUESE RHEUMATOID ARTHRITIS COHORT

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Background: Some data suggests that neutrophil -lymphocyte ratio (NLR) and monocyte-lymphocyte ratio (MLR) are related to systemic inflammation and may be associated with disease activity in rheumatoid arthritis (RA).
Objectives: To investigate the time-course change of NLR and MLR in a RA population after 6 and 12 months of bDMARD and its clinical significance in monitoring disease activity and response to bDMARDs.
Methods: Observational retrospective study including all RA patients under the first bDMARD followed at our Rheumatology Department. Data were collected from the national database. NLR and MLR were calculated by dividing neutrophil and monocyte count by lymphocyte count, respectively. The following data were collected at baseline and after 6 and 12 months of bDMARD: c-reactive protein [CRP], erythrocyte sedimentation rate [ESR], leukocyte formula ratios (NLR, MLR) and disease activity score 28 (DAS28 4V, 3V, 4V-CRP, 3V-CRP). The variation (Δ) of each parameter was calculated as the difference between the 6/12 months values and the baseline values, respectively. The correlations of ΔNLR and ΔMLR with ΔDAS28, ΔCRP and ΔESR were evaluated using the Spearman´s test.
Results: A total of 435 patients were included, 354 were females (81.4%). The mean age at baseline was 53 years (±11.4) and the median disease duration at the beginning of bDMARD was 11.0 years [0.7-45.0]. In total, 52.4% of the patients had erosive disease (n=228), 72.2% were positive for rheumatoid factor (n=314) and 77.0% for anti-citrullinated protein antibodies (n=335). Regarding treatment, etanercept (n=111; 25.5%) and adalimumab (n=64; 14.7%) were the most frequently used bDMARDs. Most patients (n=405; 93.1%) were also treated with csDMARDs, being methotrexate the most common (n=383; 88.0%). The median value of CRP was 0.99 mg/dL [0.02-30.42], 0.39 mg/dL [0.02-63.5] and 0.36 mg/dL [0.02-19.91] at baseline, 6 and 12 months, respectively. The median value of ESR was 30mm/h [2-110], 19mm/h [1-106] and 19mm/h [1-242] at baseline, 6 and 12 months, respectively. We found a similar decreasing trend in NLR and MLR values (NLR median: 2.2 [0.5-139.4], 1.7 [0.4-11.4], 1.6 [0.3-9.0]; MLR median 0.3 [0.2-252.2], 0.2 [0.1-1.2], 0.2 [0.1-1.1] at baseline, 6 and 12 months, respectively). At 6 months of bDMARD, we found positive correlations between ΔNLR and: ΔDAS28 4V ($r=0.304;p<0.001$), ΔDAS28 3V ($r=0.281;p<0.001$), ΔDAS28 4V-CRP ($r=0.325;p<0.001$), ADAS28 3V-CRP ($r=0.299;p<0.001$), ΔCRP ($r=0.316;p<0001$) and ΔESR ($r=0.264;p<0.001$). At 6 months, we also found correlations between ΔMLR and: ΔDAS28 4V ($r=0.157;p=0,016$), ΔDAS28 3V ($r=0.137;p=0.032$), ΔDAS28 4V-CRP ($r=0.202;p=0.002$), ΔDAS28 3V-CRP ($r=0.179;p=0.006$), and ΔCRP ($r=0.172;p=0.006$). At 12 months of bDMARD, ΔNLR correlated with: ΔDAS28 4V ($r=0.270;p<0.001$), ΔDAS28 3V ($r=0.266;p<0.001$), ΔDAS28 4V-CRP ($r=0.284;p<0.001$), ΔDAS28 3V-CRP ($r=0.268;p<0.001$), ΔCRP ($r=0.314;p<0.001$) and ΔESR ($r=0.231;p<0.001$). Also at 12 months, ΔMLR correlated with: ΔDAS28 4V ($r=0.149;p=0.031$), ΔDAS28 3V ($r=0.142;p=0.035$), ΔDAS28 4V-CRP ($r=0.155;p=0.025$), ΔDAS28 3V-CRP ($r=0.143;p=0.038$) and ΔCRP ($r=0.164;p=0.012$).
Conclusion: Our data demonstrate a correlation between the variation of leukocyte formula ratios (NLR, MLR) and not only the variations of inflammatory markers levels (CRP, ESR), but also the variations of disease activity scores (DAS 28). These results suggest that NLR and MLR may be seen as systemic inflammatory markers and may have a possible role in monitoring bDMARD´s effectiveness. Further studies are needed to validate these data.

P0029 – SPONDYLOARTHRITIS AND FRACTURE RISK: DOES DXA REALLY HAVE AN IMPACT IN THE RISK OF FRACTURE ESTIMATED BY FRAX?

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Background: Low bone mineral density (BMD) is common in ankylosing spondylitis (AS). The fracture risk (FR) is increased and its reduction with pharmacologic therapy is not clearly defined in this population. However, early screening and bisphosphonates as first-line treatment are recommended.
Objectives: To investigate the influence of dual-energy X-ray absorptiometry (DXA) in the ten-year risk of fracture assessed by FR Assessment Tool (FRAX) and to determine possible demographic or clinical factors associated with an increased FR in a spondyloarthritis (SpA) population.
Methods: Retrospective study including all the over 40 years-old SpA patients (ASAS classification criteria) followed at our Rheumatology Department and registered in the national database. Demographic, clinical and laboratorial data were collected at the time of the last follow-up visit. Data from the last DXA (until 3 years prior to the last visit) were collected. Indication for pharmacological treatment by FRAX was assessed according to the national recommendations.
Results: A total of 231 SpA patients were included: 126 males (54.5%), 53 (22.9%) smokers; 171 (74%) had AS, 23 (10%) had Inflammatory Bowel Disease Associated SpA and 37 (16%) had Undifferentiated SpA. At the last follow-up visit, the mean age was 52.9 years (±9.6) and the median disease duration was 21.9 years [1.0-55.5]. The mean ASDAS-CRP was 2.5 (±0.9) and the majority of patients had moderate (25.5%) or high (48.5%) disease activity (according to ASDAS). One hundred and thirty patients (56.3%) were taking NSAIDs, 45 (19.5%) were taking glucocorticoids, 85 (36.8%) were under csDMARDs and 170 (73.6%) under bDMARDs [157 (68%) under TNFi, 11 (4.8%) under secukinumab and 2 (0.9%) under ustekinumab].
Eleven patients (4.8%) had previous fragility fractures, 118 (51.1%) had DXA in the last 3 years and 167 (72.3%) were taking calcium and/or vitamin D supplements.
Sixteen patients (6.9%) had indication for treatment by FRAX without DXA and 9 of these (56.3%) were already under treatment. Similarly, 16 (6.9%) had indication for treatment by FRAX with DXA and 13 of these (81.3%) were already under treatment. Ten patients (4.3%) were reclassified in FRAX with DXA: 7 (3%) had no indication for treatment by FRAX without DXA but obtained it by FRAX with DXA and 3 (1.3%) had indication for treatment by FRAX without DXA but they lost it by FRAX with DXA. We found a moderate level of agreement in the indication for treatment between FRAX with and without DXA (kappa=0.595; p<0.001). The use of DXA in FRAX estimated a significant higher median FR, both for major osteoporotic fracture (2.4% [0.8-31.0] vs 1.8% [0.6-20.0]; p<0.001) and for hip fracture (0.5% [0.0-23.0] vs 0.2% [0.0-14.0]; p<0.001).
We found significant correlations between FR and some disease-related variables (table 1).
Conclusion: Our results showed that a similar number of patients had indication for pharmacological treatment by FRAX both with and without DXA. Although the inclusion of DXA resulted in a higher estimated FR by FRAX, the observed moderate level of agreement between FRAX with and without DXA suggests that the FR estimation by FRAX, even without DXA, may be a reasonable approach in SpA patients. In line with literature, we found significant associations between the estimated risk fracture by FRAX and some disease activity and function measures.

TABLE 1. CORRELATIONS BETWEEN THE RISK OF FRACTURE ESTIMATED BY FRAX AND DISEASE-RELATED VARIABLES

			Disease duration	BASDAI	ASDAS-CRP	BASMI	BASFI
Estimated fracture risk by FRAX:	without DXA	major osteoporotic fracture	r=0.352 p<0.001	r=0.204 p=0.002	r=0.214 p=0.001	r=0.301 p<0.001	r=0.317 p<0.001
		hip fracture	r=0.389 p<0.001	r=0.142 p=0.034	r=0.170 p=0.011	r=0.305 p<0.001	r=0.275 p<0.001
	with DXA	major osteoporotic fracture	r=0.227 p=0.014	r=0.314 p=0.001	r=0.356 p<0.001	r=0.293 p=0.002	r=0.379 p<0.001
		hip fracture	n.s.	r=0.197 p=0.036	r=0.269 p=0.004	r=0.271 p=0.004	r=0.258 p=0.006

ASDAS: Ankylosing Spondylitis Disease Activity Score; BASDAI: Bath Ankylosing Spondylitis Activity Index; BASFI: Bath Ankylosing Spondylitis Functional Index; BASMI: Bath Ankylosing Spondylitis Metrology Index; CRP: C-reactive protein; DXA: dual-energy X-ray absorptiometry; FRAX: Fracture Risk Assessment Tool; n.s. not significant

P0030 – FRACTURE RISK ASSESSMENT BY FRAX IN A SYSTEMIC LUPUS ERYTHEMATOSUS PORTUGUESE COHORT

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Background: Osteoporosis is commonly seen in patients with Systemic Lupus Erythematosus (SLE), even in pre-menopausal patients. The etiology is multifactorial and chronic glucocorticoid therapy seems to play a central role.
Objectives: To investigate the ten-year risk of fracture assessed by Fracture Risk Assessment Tool (FRAX),

with and without dual-energy X-ray absorptiometry (DXA) and to determine possible demographic or clinical factors associated with an increased risk of fracture in a SLE population.

Methods: Retrospective study including all the over 40 years-old patients with the diagnosis of SLE (2012 SLICC classification criteria) followed at our Rheumatology Department registered in our national database. Demographic, clinical and laboratorial data were collected at the last follow-up visit. Data from the last DXA (until 3 years prior to the last visit) were collected. Indication for pharmacological treatment by FRAX was assessed according to the national recommendations: estimated fracture risk, without DXA, ≥11% for major osteoporotic fracture or ≥3% for hip fracture and/or estimated fracture risk, with DXA, ≥9% for major osteoporotic fracture or ≥2.5% for hip fracture.

Results: We included 104 patients, 101 (97.1%) females, aged 54.5±9.9 years, with a median disease duration of 19.3 years [4.3-51.6]. Twelve patients (11.5%) were current smokers, 31 (29.8%) had elevated anti-dsDNA antibodies (≥100 IU/mL) and 27 (26.0%) had complement consumption (C3c<83mg/dL or C4<12mg/dL). Seventy-three patients (70.2%) were taking glucocorticoids with a mean daily prednisolone equivalent dosage of 4.4±5.2 mg/day. Regarding SLE treatment, 69 patients (66.3%) were under hydroxychloroquine, 22 (21.2%) under azathioprine, 16 (15.4%) under mycophenolate mofetil, 5 (4.8%) under belimumab, 4 (3.8%) under methotrexate, 1 (1.0%) under leflunomide and 1 (1.0%) under rituximab. Ten patients (9.6%) had previous fragility fractures, 54 patients (51.9%) had DXA in the last 3 years and 81 (77.9%) were taking calcium and/or vitamin D supplements. Sixteen (15.4%) had indication for treatment by FRAX without DXA and 8 of these (50%) were under treatment. Moreover, thirteen (12.5%) had indication for treatment by FRAX with DXA and 8 of these (61.5%) were under treatment. Five patients (4.8%) were reclassified in FRAX with DXA: 3 patients (2.9%) had no indication for treatment by FRAX without DXA but conquered it by FRAX with DXA and 2 patients (1.9%) had indication for treatment by FRAX without DXA but lost it by FRAX with DXA. We found a good level of agreement in the indication for treatment between FRAX with and without DXA (kappa=0.741; p<0.001). There was no significant difference in the risk of fracture estimated by FRAX with or without DXA, both for major osteoporotic fracture and for hip fracture. Correlations between fracture risk and some clinical variables can be seen in table 1.

TABLE 1. CORRELATIONS BETWEEN THE RISK OF FRACTURE ESTIMATED BY FRAX AND DISEASE RELATED FEATURES

			Age at SLE diagnosis	Disease Duration	ESR	SLEDAI
Estimated fracture risk by FRAX:	without DXA	major osteoporotic fracture	r=0.483 *p*<0.001	n.s.	r=0.249 *p*=0.012	r=-0.586 *p*=0.028
		hip fracture	r=0.481 *p*<0.001	n.s.	r=-0.552 *p*=0.041	n.s.
	with DXA	major osteoporotic fracture	r=0.386 *p*=0.005	r=0.299 *p*=0.033	n.s.	n.s.
		hip fracture	r=0.338 *p*=0.015	n.s.	n.s.	n.s.

DXA: dual-energy X-ray absorptiometry; ESR: Erythrocyte Sedimentation Rate; FRAX: Fracture Risk Assessment Tool; SLE: Systemic Lupus Erythematosus; SLEDAI: Systemic Lupus Erythematosus Disease Activity Index; n.s. not significant

Conclusion: A higher number of patients had indication for pharmacological treatment by FRAX with DXA in comparison with FRAX without DXA. However, we found no statistically significant difference in the estimated fracture risk with and without DXA. This, together with the good level of agreement between FRAX with and without DXA, suggests that the fracture risk estimation, even without DXA, may be an appropriate approach. The low number of patients with indication for pharmacological treatment by FRAX, with and without DXA, may be explained by their low mean age and the high number of them under vitamin D/calcium supplementation.

P0033 – BELIMUMAB IN THE TREATMENT OF 38 PORTUGUESE SLE PATIENTS: A REAL-LIFE MULTICENTER STUDY

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Background: Systemic Lupus Erythematosus (SLE) is a multisystemic disease in which therapeutic approach depends on the severity of the disease and organ involvement. Belimumab, a human monoclonal antibody targeting BLyS, is the first biotechnological drug approved for SLE. Clinical trials showed reduction of disease activity, number and severity of flares, steroid-sparing effects and improvement in health-related quality of life and fatigue. Better efficacy was demonstrated in patients with higher disease activity, anti-dsDNA positivity and low complement levels. Registries allow for a better understanding of real-world data on new drugs, particularly regarding efficacy and safety.
Objectives: To study belimumab´s effectiveness and safety in SLE patients followed in Portuguese rheumatology centers.
Methods: Multicenter cohort study including SLE patients (2012 SLICC classification criteria) treated with belimumab in rheumatology departments and registered at the Rheumatic Diseases Portuguese Register (Reuma.pt). Demographic and clinical features were collected at baseline and after 6, 12 and 24 months of belimumab treatment (until June 2019). The primary efficacy endpoint was SLE Responder Index (SRI) response rate at 6, 12 and 24 months of treatment with belimumab and safety was evaluated by the number of adverse events. The statistical analysis was performed using SPSS 24.0. Differences were considered statistically significant at p<0.05.
Results: Thirty-eight patients were included: 37 (97.4%) female, 34 (89.5%) Caucasian, with a mean age of 46.2±13.9 years and a median disease duration of 9.7 years (min:0.9; max:33.6) at the start of belimumab. Median SLEDAI at baseline was 8.2±3.9. 26/33 (78.8%) had elevated anti-dsDNA antibodies and 24/33 (72.7%) had complement consumption. Twenty-eight patients (73.7%) were concomitantly treated with hydroxychloroquine, 16 (42.1%) with azathioprine, 7 (18.4%) with methotrexate and 3 (7.9%) with mycophenolate mofetil. The reasons for prescribing belimumab were: multiorgan involvement in 20 (52.6%), haematologic disorders in 9 (23.7%), cutaneous manifestations in 5 (13.2%), arthritis in 3 (7.9%), necrotizing vasculitis in 1 (2.6%). Belimumab was administered intravenously for a median of 12 (min:1; max:76) months. SRI response was achieved in 14/27 (51.9%), 12/20 (60%) and 11/12 (91.7%) at 6, 12 and 24 months of belimumab treatment, respectively. Mean SLEDAI significantly decreased from 8.2±3.9 at baseline to 3.8±2.2, 4.1±3.2 and 3.1±1.6 at 6, 12 and 24 months, respectively. Anti-dsDNA antibodies significantly decreased at 6, 12 and 24 months and C3 increased at 12 months of belimumab treatment (table 1). We found a significant reduction in mean daily prednisolone dosage (p<0.001) from baseline (10.8±5.1mg) to the last evaluation under belimumab (5.5±3.0mg). Eleven (28.9%) patients discontinued belimumab: loss of efficacy in 4, lost to follow-up in 4, adverse events in 3 (urinary tract infections, acute myocardial infarction, breast cancer). Three presented infections related to belimumab (respiratory, skin and urinary tract infections).
Conclusion: We confirmed belimumab´s effectiveness, immunological response and steroid-sparing effect in real-life active SLE patients. Demographic features and SRI response rates were similar to the data from clinical trials. Of interest, about a quarter of the treated patients did not have elevated anti-dsDNA antibodies neither complement consumption. We also showed a good safety profile. As an observational study, the main limitation was the missing data.

TABLE 1. SRI RESPONSE AND EVOLUTION OF SLEDAI, ANTI-DSDNA ANTIBODIES AND C3 LEVELS AT 6, 12 AND 24 MONTHS OF BELIMUMAB AND ITS SIGNIFICANCE COMPARED TO BASELINE VALUES

	Baseline (n=38)	6 months (n=28)	12 months (n=21)	24 months (n=12)
SRI response - n (%)	-	14 (51.9%) (n=27)	12 (60%) (n=20)	11 (91.7%) (n=12)
SRI response LUNDEX adjusted - %	-	45.4%	45.0%	45.8%
SLEDAI - mean±SD	8.2±3.9 (n=37)	3.8±2.2 (p<0.001) (n=28)	4.1±3.2 (p<0.001) (n=20)	3.1±1.6 (p=0.002) (n=12)
Anti-dsDNA antibodies (IU/mL) - mean±SD	255.8±277.4 (n=33)	175.5±189.0 (p=0.015) (n=26)	177.4±333.9 (p=0.031) (n=20)	111.4±129.1 (p=0.001) (n=11)
C3 (mg/dL) - mean±SD	82.6±28.8 (n=33)	89.6±27.1 (p=0.223) (n=27)	94.6±25.8 (p=0.018) (n=21)	100.9±36.9 (p=0.139) (n=12)

SLEDAI: Systemic Lupus Erythematosus Disease Activity Index; SRI: Systemic Lupus Erythematosus Responder Index

P0035 – EARLIER TREATMENT OF NON-RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS WITH CERTOLIZUMAB PEGOL RESULTS IN IMPROVED CLINICAL AND PATIENT-REPORTED OUTCOMES

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Background/Purpose: Patients (pts) with axial spondyloarthritis (axSpA) often experience delayed diagnosis, which can lead to treatment delay. However, there are indications that earlier treatment with anti-tumour necrosis factors (-TNFs) can lead to a greater clinical response. Certolizumab pegol (CZP) has been shown to improve the signs and symptoms of non-radiographic (nr)-axSpA. However, it is not known if earlier CZP treatment has a greater impact on efficacy in nr-axSpA. Here we report clinical and pt-reported outcomes in pts with active nr-axSpA treated with CZP or placebo (PBO) over 52 weeks (wks) stratified by their symptom duration.

Methods: C-axSpAnd (NCT02552212) is a 3-year, phase 3, multicentre study including a 52-wk double-blind, PBO-controlled period (completed). Pts had previous inadequate response to ≥ 2 NSAIDs and were randomised 1:1 to PBO or CZP (400 mg at Wks 0/2/4, then 200 mg every 2 wks). This post-hoc analysis reports outcomes at Wk 12 and Wk 52 in pts stratified by their baseline symptom duration (< 5 and ≥ 5 years; key clinical outcomes also reported for < 3 and ≥ 3 years). Outcomes included: Ankylosing Spondylitis Disease Activity Score – Major Improvement (ASDAS-MI), Assessment in SpondyloArthritis international Society 40% response (ASAS40), Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), nocturnal spinal pain, fatigue (BASDAI Q1), morning stiffness (average of BASDAI Q5 and Q6), and the 36-Item Short Form Survey (SF-36) physical and mental component summary (PCS/MCS). Pts with missing values or values observed after discontinuing double-blind study treatment were considered as non-responders for binary measures or, for quantitative measures, had the last observation from double-blind treatment carried forward.

Results: Of 317 recruited pts, 159 were randomised to CZP, and 158 to PBO. The median (range) baseline (BL) symptom duration was 4.9 (1.0–41.9) years for CZP-treated pts and 5.2 (1.1–38.2) years for PBO pts. 50.3% (80/159) CZP pts and 48.7% (77/158) PBO pts had a symptom duration < 5 years. At Wks 12 and 52, ASDAS-MI and ASAS40 responder rates, and improvements in BASDAI, nocturnal spinal pain, fatigue, morning stiffness and SF-36 PCS were substantially better among CZP-treated pts with shorter symptom duration (< 5 years at BL) vs longer symptom duration (Table). Amongst PBO pts, responses were low and there was no consistent trend in outcomes by symptom duration (Table). Similarly, using a cut-off of 3 years, responder rates for ASDAS-MI and ASAS40 were greater in CZP-treated pts with shorter symptom duration: at Wk 52, 56.4% (31/55) and 42.3% (44/104) of pts with < 3 and ≥ 3 years symptom duration achieved ASDAS-MI, respectively, while 65.5% (36/55) and 51.9% (54/104) achieved ASAS40.

TABLE: CLINICAL AND PATIENT-REPORTED OUTCOMES IN PATIENTS TREATED WITH PBO OR CZP STRATIFIED BY SYMPTOM DURATION

		CZP (n=159)		PBO (n=158)	
	Wk	Symptom duration: < 5 yrs (n=80)	Symptom duration: ≥ 5 yrs (n=79)	Symptom duration: < 5 yrs (n=77)	Symptom duration: ≥ 5 yrs (n=81)
ASDAS-MI	**12**	46.3 (37)	24.1 (19)	9.1 (7)	3.7 (3)
% (n)	**52**	55.0 (44)	39.2 (31)	7.8 (6)	6.2 (5)
ASAS40	**12**	58.8 (47)	36.7 (29)	11.7 (9)	11.1 (9)
% (n)	**52**	65.0 (52)	48.1 (38)	18.2 (14)	13.6 (11)
BASDAI	**0**	6.7 (1.3; 80)	7.0 (1.5; 79)	6.9 (1.3; 77)	6.7 (1.3; 81)
Mean (SD; n)	**12**	3.4 (2.1; 79)	4.5 (2.2; 78)	5.8 (2.1; 77)	5.6 (2.1; 81)
	52	2.8 (2.3; 79)	3.7 (2.5; 78)	5.5 (2.4; 77)	5.4 (2.2; 81)
Nocturnal spinal pain	**0**	6.2 (2.7; 80)	7.1 (1.8; 78)	6.4 (2.2; 77)	6.8 (2.0; 81)
Mean (SD; n)	**12**	2.8 (2.7; 79)	4.1 (2.5; 78)	5.5 (2.6; 77)	5.6 (2.5; 81)
	52	2.2 (2.7; 79)	3.2 (2.7; 78)	5.2 (2.7; 77)	5.7 (2.9; 81)
Fatigue [a]	**0**	7.1 (1.5; 80)	7.2 (1.7; 79)	7.3 (1.3; 77)	7.2 (1.4; 81)
Mean (SD; n)	**12**	3.8 (2.2; 79)	4.9 (2.4; 78)	6.1 (2.1; 77)	6.1 (2.3; 81)
	52	3.3 (2.7; 79)	4.1 (2.7; 78)	5.9 (2.3; 77)	6.0 (2.4; 81)
Morning stiffness [b]	**0**	6.7 (1.9; 80)	7.2 (1.7; 79)	6.8 (1.8; 77)	6.6 (1.8; 81)
Mean (SD; n)	**12**	2.9 (2.3; 79)	4.3 (2.2; 78)	5.7 (2.6; 77)	5.4 (2.2; 81)
	52	2.2 (2.3; 79)	3.6 (2.4; 78)	5.3 (2.8; 77)	5.1 (2.4; 81)
SF-36 PCS	**0**	35.0 (7.1; 80)	34.2 (7.0; 77)	34.0 (6.8; 77)	33.5 (7.2; 80)
Mean (SD; n)	**12**	44.3 (8.4; 79)	40.9 (8.4; 78)	35.8 (7.3; 76)	36.0 (8.4; 81)
	52	47.6 (8.9; 79)	42.1 (9.4; 78)	37.3 (8.2; 76)	36.7 (8.3; 81)
SF-36 MCS	**0**	41.7 (11.0; 80)	42.3 (11.0; 77)	41.7 (9.3; 77)	40.6 (10.8; 80)
Mean (SD; n)	**12**	47.7 (9.5; 79)	45.2 (11.4; 78)	42.6 (10.6; 76)	43.7 (11.1; 81)
	52	47.1 (10.5; 79)	47.3 (11.2; 78)	41.9 (11.9; 76)	43.4 (11.0; 81)

Missing values were imputed using double-blind observation carried forward or, for ASDAS-MI and ASAS40, considered to be non-response. [a] BASDAI Q1; [b] Average of BASDAI Q5 and Q6. ASAS40: Assessment in SpondyloArthritis international Society 40% response; ASDAS-MI: Ankylosing Spondylitis Disease Activity Score – Major Improvement; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; CZP: certolizumab pegol; MCS: mental componente summary; PBO: placebo; PCS: physical componente summary; SD: standard deviation; SF-36: 36-Item Short Form Survey; wk: week; yrs: years.

Conclusion: In this post-hoc analysis, CZP-treated nr-axSpA pts with shorter symptom duration (< 5 vs ≥ 5 years) showed greater improvements across signs and symptoms of disease and in quality of life. To our knowledge, this is the first report indicating that early CZP treatment for nr-axSpA may be beneficial to pts.

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P0037 – ANA TESTING IN THE (VERY) ELDERLY: EXPECTATION VERSUS REALITY

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Introduction: Antinuclear antibodies (ANA) are frequently used as a screening tool for systemic autoimmune rheumatic diseases (SARD), although they are also present in 10-15% of the adult healthy population. SARD have their peak incidence in the young/ middle-aged adult. As age progresses, the incidence of SARD decreases while the prevalence of ANA tends to increase, with some series reporting up to 30% prevalence in older ages (1).
Aim: To determine the clinical significance and utility of ANA testing in a population over 85 years of age.
Methods: We conducted a retrospective study of patients over the age of 85 who underwent ANA testing due to a SARD suspicion at our hospital autoimmunity laboratory, from 2011 to 2018. Justification for ANA request was collected from patient's clinical records. Patients with pre-established diagnosis of SARD and patients with no justification given for ANA request were excluded from the analysis. ANA titer (positive ≥ 1:160) and cellular staining patterns were assessed by indirect immunofluorescence (Hep-2 cells).
Results: Ages ranged from 85 to 98 years, with 58.8% being females. The prevalence of ANA in this population was 61.5%, mostly in lower titers (1:160 in 45.0%, 1:320 in 31.9%, 1:640 in 20.3% and 1:1280 in 2.7%). Dense fine speckled pattern was by far the most common cellular staining pattern (79.1%). A suspicion of SARD was the reported reason for ANA testing in 34,5% (n=296) of the 854 patients submitted to this test. The main clinical clues justifying SARD suspicion were: arthralgia/arthritis (11.9%), thrombocytopenia (10.0%), pancytopenia (10.0%), spotless fever (8.2%), interstitial lung disease (4.8%), pleural (6.1%) and pericardial (4.1%) effusion. Over a median follow-up of 1.0 year, 10 patients (3.4%) were diagnosed with a SARD, only one being an ANA-related disease: 5 cases of polymyalgia rheumatica, 2 cases of rheumatoid arthritis, 1 case of giant cell arteritis, 1 case of Sjogren syndrome and 1 case of sarcoidosis. In 60% of patients with a confirmed SARD, the main reason for suspicion was the presence of arthralgia/ arthritis. Positive ANA testing showed a 90.0% sensitivity and a 39.6% specificity for SARD. This translates into a positive predictive value of 5.0%.
Conclusion: ANA are highly prevalent in elderly patients under SARD suspicion, while the incidence of SARD is very low, which explains the low positive predictive value of ANA testing. Interestingly, only one among the ten cases of SARD confirmed was indeed an ANA-related disease (Sjogren syndrome).

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P0039 – DETERMINANTS OF HAPPINESS AND QUALITY OF LIFE IN PEOPLE WITH SYSTEMIC SCLEROSIS: A STRUCTURAL EQUATION MODELLING APPROACH

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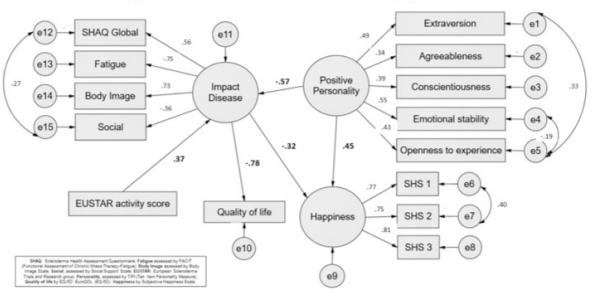
Background: In recent years more attention has been given to patients reported outcomes (PROs). Systemic sclerosis (SSc) is no exception. As there is no effective treatment or cure to SSc, it is important to recognize the relevance to patients of the different features of the disease to improve quality and enjoyment of life: the ultimate targets of therapy. Remarkably lacking in PROs is the evaluation of the overall perspective of subjective well being, equivalent to 'happiness' or "positive psychological dimensions".
Objective: To examine the determinants of happiness and quality of life (QoL) in patients with SSc with emphasis on disease activity, disease impact and personality traits.
Methods: This is an observational, cross-sectional and multicenter study from six rheumatology clinics in Portugal. A total of 113 patients with SSc with

FIGURE 1 – ESTIMATED STANDARDISED DIRECT EFFECTS FOR THE PROPOSED MODEL



*All with p<05; Circles represente latent factos. Squares represente measured variables. Arrows show a hypothesized direct relationship. Curved lines with an arrow represente a covariance.

a complete set of data on disease activity, disease impact, personality, quality of life and happiness were included.

Structural equation modelling (latent variable structural model) was used to estimate the association between the variables using a maximum likelihood estimation with Satorra-Bentler's correction and performed with STATA® 15.0. Two hypotheses were pursued: H1 – Disease activity and impact of disease are negatively associated to overall QoL and happiness; H2 – 'Positive' personality traits are related to happiness both directly and indirectly through perceived disease impact.

Results: Results obtained in the structural equation measurement model indicated a good fit [$\chi 2/df=1.44$; CFI=0.93; TLI=0.90; RMSEA=0.06] and supported all driving hypotheses (Figure 1). Happiness was positively related to 'positive' personality ($\beta=0.45$, p=0.01) and, to a lesser extent, negatively related with impact of disease ($\beta=-0.32$; p=0.01). This impact, in turn, was positively related to EUSTAR activity score ($\beta=0.37$; p<0.001) and mitigated by 'positive' personality traits ($\beta=-0.57$; p<0.001). Impact of disease had a much stronger relation with QoL than with happiness ($\beta=-0.78$, p<0.001). Quality of life and happiness had no statistically significant relationship.

Conclusion: Optimization of Qol and happiness in people with SSc requires effective control of the disease process. Personality and its effects upon the patient´s perception of the disease impact, seems to play a pivotal mediating role in these relations and should deserve paramount attention if happiness and enjoyment of life is taken as the ultimate goal of health care.

P0042 – REDUCTION OF ANTERIOR UVEITIS FLARES IN PATIENTS WITH AXIAL SPONDYLOARTHRITIS FOLLOWING 1 YEAR OF TREATMENT WITH CERTOLIZUMAB PEGOL: 48-WEEK INTERIM RESULTS FROM A 96-WEEK OPEN-LABEL STUDY

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Background/Purpose: Acute anterior uveitis (AAU), inflammation of the anterior uveal tract, is the most common extraarticular manifestation in patients (pts) with axial spondyloarthritis (axSpA), reported by up to 40% of pts. Uveitis is associated with a significant clinical burden; common symptoms include blurred vision, photophobia and pain. Previous studies have shown that anti-TNFs can reduce the incidence of AAU flares in pts with radiographic axSpA (ankylosing spondylitis), but few have focused on pts across the full axSpA spectrum. The aim of the C-VIEW study was to analyse the impact of certolizumab pegol (CZP) treatment on AAU flares in pts with active axSpA (radiographic and non-radiographic) and a recent history of AAU.

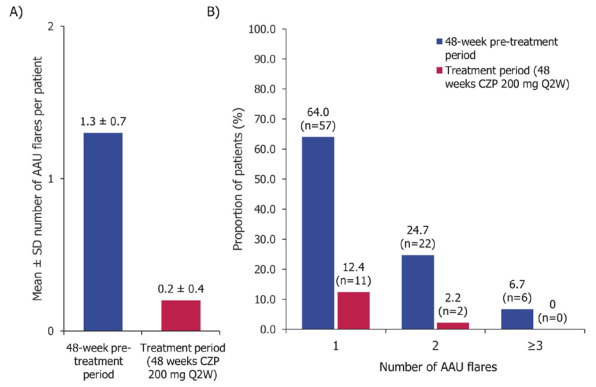
Methods: C-VIEW (NCT03020992) is an ongoing multicentre, open-label, phase 4 study. Pts had active axSpA according to the Assessment of SpondyloArthritis international Society (ASAS) classification criteria, a history of recurrent AAU (≥2 AAU flares in total and ≥1 AAU flare in the year prior to study entry), were HLA-B27 positive, and were eligible for anti-TNF treatment (active axSpA, previous failure of ≥2 NSAIDs, biologic naïve or had failed at most one anti-TNF). Pts received CZP 400 mg at Weeks (Wks) 0/2/4, then 200 mg every 2 wks through 96 wks. The primary variable was the incidence of AAU flares compared to historic rates. A pre-specified interim analysis compared AAU incidence in the 48 wks prior to CZP treatment initiation with the 48 wks of treatment, using Poisson regression adjusted

TABLE: BASELINE CHARACTERISTICS OF PATIENTS IN C-VIEW

	CZP 200 mg Q2W (N=89)
Age (years), mean ± SD	46.5 ± 11.2
Male, n (%)	56 (62.9)
Racial group, n (%)	
Caucasian	87 (97.8)
Other	2 (2.2)
Diagnosis, n (%)	
Radiographic axSpA	76 (85.4)
Non-radiographic axSpA	13 (14.6)
Duration of axSpA (years), mean ± SD	8.6 ± 8.4
Time since onset of first uveitis flare (years), mean ± SD	9.9 ± 9.0
ASDAS, mean ± SD	3.5 ± 0.9
BASDAI, mean ± SD	6.5 ± 1.5

ASDAS: Ankylosing Spondylitis Disease Activity Score; axSpA: axial spondyloarthritis; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; CZP: certolizumab pegol; Q2W: every 2 weeks; SD: standard deviation.

FIGURE: (A) MEAN NUMBER OF AAU FLARES EXPERIENCED BY PATIENTS IN C-VIEW AND (B) PROPORTION OF PATIENTS EXPERIENCING 1, 2 OR >3 AAU FLARES



AAU: acute anterior uveitis; CZP: certolizumab pegol; Q2W: every 2 weeks; SD: standard deviation.

for possible within-patient correlations, with period (pre- and post-baseline) and axSpA disease duration as covariates. Incidence rates (IR) were calculated based on the number of cases per pts at risk over 48 wks. Observed data are reported.

Results: Of 115 enrolled pts, 89 initiated CZP treatment and 85 completed Wk 48. Baseline characteristics are shown in the Table. The 48-wk interim analysis revealed significantly fewer AAU flares per pt during CZP treatment compared to before treatment (Figure; Poisson-adjusted IR: 0.2 vs 1.5, p<0.001). The number of pts experiencing 1 and ≥2 AAU flares (64.0% and 31.5%, respectively) was substantially reduced during CZP treatment (12.4% and 2.2%). In the 13 patients who had AAU flares both pre- and post-baseline, the mean duration of AAU flares was also reduced during CZP treatment from 97.4 to 58.4 days. After 48 wks CZP, pts' disease activity had improved substantially (mean ± SD Ankylosing Spondylitis Disease Activity Score [ASDAS]: 2.0 ± 0.9; BASDAI: 3.3 ± 2.1), with 31.4% of pts achieving ASAS partial remission and 29.1% ASDAS major improvement. No new safety signals were identified.

Conclusion: In this open-label study, we found a significant reduction in the AAU flare rate in axSpA pts with a history of recurrent AAU during the first 48 wks of CZP treatment. Pts also experienced significant improvement in axSpA disease activity during CZP treatment.

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P0044 – RISK FACTORS FOR RENAL FLARE IN PATIENTS WITH PROLIFERATIVE LUPUS NEPHRITIS

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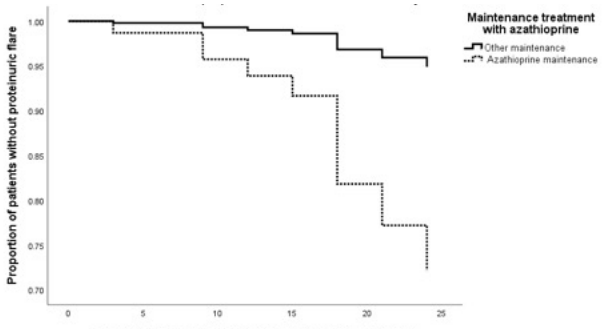
Background: Goals of lupus nephritis (LN) maintenance treatment include prevention of LN flares and long-term preservation of renal function, while minimizing drug iatrogenicity. There is an unmet need for identifying risk factors of LN flare in order to guide optimization of maintenance immunosuppression.

Aim: To identify risk factors of LN flare after attainment of complete renal response (CRR) in patients with proliferative LN.

Methods: Retrospective cohort study over 36 months including patients with SLE fulfilling the ACR'97 and/or the SLICC'12 classification criteria, enrolled in the CHUC Lupus Cohort between 1999 and 2018, with a biopsy-proven proliferative LN (class III/IV) and who attained CRR (proteinuria <0.5g/day and normal renal function, according to EULAR/ERA-EDTA definition) following induction treatment. Only proteinuric flares were considered and defined as doubling of proteinuria to >1g/day. Clinical-analytic characteristics at baseline (time of first CRR attainment after induction) were compared using survival analysis for time-to-flare. Variables with p<0.10 on univariate analysis with Log-Rank tests were further evaluated as predictors with multivariate Cox proportional hazards regression models (Backward Stepwise method, Wald-based), with estimation of hazard ratios (HR) with 95% confidence intervals (95%CI).

COX PROPORTIONAL HAZARDS MULTIVARIATE ANALYSIS



Time since complete remission of lupus nephririts (months)

Results: A total of 50 patients in CRR were included in the analysis (78.4% female, age at baseline 30.0 ± 12.5 years-old). Most patients had a diffuse proliferative nephritis (76.5%) with the remaining showing a focal proliferative type. At the time of CRR, 25.5% patients were under cyclophosphamide, 45.1% under mycophenolate mofetil and 25.5% under azathioprine. Over the follow-up period, 10 patients (20.0%) experienced a proteinuric flare, within a mean time of 29.1 months (95%CI 26.89-31.37). In univariate analysis, age <30 years-old (p=0.020), arterial hypertension (p=0.020) and presence of anti-RNP antibody (p=0.002) at baseline were associated with LN proteinuric flares. In multivariate analysis, age <30 years-old (HR 26.56; 95%CI 1.93-365.08; p=0.014), arterial hypertension (HR 8.30; 95%CI 1.21-56.92; p=0.031), use of antihypertensive antiproteinuric drugs (angiotensin-converting-enzyme inhibitors or angiotensin receptor blockers) (HR 11.18; 95%CI 1.24-100.66; p=0.031) and maintenance therapy with azathioprine (HR 6.23; 95%CI 1.51-25.66; p=0.011) (Figure 1) were identified as risk factor for LN proteinuric flares.

Conclusions: In patients with proliferative LN, proteinuric flares are a frequent event after induction treatment leads to CRR. Younger age, arterial hypertension, use of antihypertensive drugs and use of azathioprine as maintenance therapy were risk factors for LN proteinuric flare in this cohort. Given the retrospective non-randomized nature of this study, caution is needed when drawing conclusions, particularly regarding treatment efficacy.

P0045 – CLINICAL AND DEMOGRAPHIC CHARACTERISTICS OF PATIENTS WITH ANTISYNTHETASE AUTOANTIBODIES: DATA FROM A PORTUGUESE TERTIARY OUTPATIENT CLINIC

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Introduction: Antisynthetase syndrome (AS) may have different clinical phenotypes and outcomes associated with different anti-aminoacyl RNA-synthetase (anti-ARS). Patients may also present with incomplete/early phenotypes that do not fulfil the classification criteria.

Objective: To evaluate the clinical and demographic characteristics of patients positive for anti-ARS in our Myositis clinic.

Methods: Observational study using data from the Portuguese Rheumatic Diseases Register (Reuma.pt/Myositis protocol). Data extracted included demographic variables, clinical features and immunological expression of the disease.

Results: 17 patients were identified. All met the criteria for AS according to Connors criteria, while 3 did not met according to the Solomon criteria. Mean age at diagnosis was 60.1 years (26 – 80) and 76.5% were female. Mean follow-up time was 2.8 (0.5-9) years. Only 3 patients had history of smoking in the past. The autoantibodies detected were anti-Jo1 (n=12), anti-PL12 (n=2), anti-OJ (n=2) and anti-PL7 (n=1). 4 patients positive for anti-Jo-1 also had anti-Ro52 antibodies. The clinical information and treatment are

TABLE 1. DEMOGRAPHIC AND CLINIC CHARACTERISTICS OF OUR COHORT

Case	Serology	Major criteria (ILD [NSIP/UIP/non-specific] or PM/DM)	Minor criteria (Arthritis, MH, RP)	Other manifestations	Connors (2010)/ Solomon (2011) criteria	Prednisolone	DMARDcs (Methotrexate/MMF/AZA)	CYC/RTX
1	Jo1	PM	MH, arthritis	GS	Y/Y	Y	MMF	-
2	Jo1	NSIP	RP, MH	Asthenia	Y/Y	Y	AZA	-
3	Jo1	NSIP	MH, arthritis	Calcinosis, asthenia	Y/Y	Y	AZA	-
4	Jo1	NSIP; DM	-	GS	Y/Y	Y	-	-
5	Jo1/Ro52	PM	MH, arthritis	GS, asthenia	Y/Y	Y	MTX	-
6	Jo1	UIP	RP, arthritis	Asthenia	Y/Y	Y	-	RTX
7	Jo1/Ro52	NSIP; PM	arthritis	-	Y/Y	Y	MMF	CYC*
8	Jo1	NSIP; PM	-	-	Y/Y	Y	MMF	-
9	Jo1/Ro52	NSIP; PM	-	-	Y/Y	Y	AZA	-
10	PL7	-	RP, arthritis, MH	Dysphagia	Y/N	Y	-	-
11	Jo1	-	RP	Asthenia	Y/N	-	-	-
12	Jo1/Ro52	PM	MH, arthritis	GS, asthenia	Y/Y	Y	MMF	-
13	PL12	PM	RP, arthritis	GS, asthenia	Y/Y	Y	MMF	-
14	Jo1	UIP	RP, arthritis	Weight loss, asthenia	Y/Y	Y	-	RTX
15	OJ	Non-specific pattern	-	-	Y/N	Y	MMF	-
16	OJ	NSIP	-	-	Y/Y	Y	-	-
17	PL12	PM	RP, MH	-	Y/Y	-	-	-

AZA – azathioprine; CYC – Cyclophosphamide; DM – dermatomyositis; GS – Gottron's sign; ILD – Interstitial lung disease; PM – Polymyositis; NSIP – Nonspecific interstitial pneumonia; MH – mechanic hands; MMF – Mycophenolate mofetil; RP – Raynaud phenomenon; RTX – Rituximab; UIP – Usual interstitial pneumonia; Y – yes; N – no. Connors criteria – anti-ARS plus one or more of RP, MH, arthritis, ILD, fever. Solomon criteria – anti-ARS plus 2 major criteria or 1 major and 2 minor criteria. *induction therapy.

described in table 1. One patient presented as a paraneoplastic syndrome associated with anti-Jo-1.
Discussion/Conclusion: The most frequent autoantibody was anti-Jo-1, which is consistent with the literature. Interestingly, patients with anti-PL, usually described as having severe lung disease, in our series did not have it. Additionally, we found a trend for a younger age at diagnosis in Jo1 positive patients and remarkably more than half of these patients had been diagnosed with ILD, being the NSIP pattern the most frequently reported.

P0047 – RHEUMATOID ARTHRITIS: IS IT WORTH IT TO ADD LEFLUNOMIDE TO METHOTREXATE IN REFRACTORY DISEASE?

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Background: In refractory rheumatoid arthritis (RA), adding other classic synthetic disease-modifying antirheumatic drug (csDMARD) such as leflunomide (LFN) to methotrexate (MTX) is a suitable option. Yet, there are safety issues to consider that might limit this strategy, but also regarding its true effectiveness in avoiding exposure to biological DMARDs (bDMARD) or target synthetic DMARDs (tsDMARD).
Objectives: To assess the effectiveness and safety of adding LFN to MTX and to evaluate the predictors of drug retention, toxicity and inefficacy.
Methods: A retrospective, monocentric, clinical record review of adult RA patients in whom LFN was added to MTX was done. Sociodemographic information, comorbidities, disease related information, adverse reactions and disease activity according to Disease Activity Score 28 – C reactive protein (DAS28) were recorded at baseline and after 3, 6 and 12 months of combination therapy (3_DAS28; 6_DAS28; 12_DAS28, respectively). Information regarding toxicity (need to dose adjustment/suspension) and inefficacy (add/switch to bDMARD/tsDMARD) were recorded. Follow-up was considered until last medical record available. SPSS was used for statistical analysis. Kaplan Meier and Cox-regression were used for univariate and multivariate analysis, respectively, significant level was 2-sided p<.05.
Results: In total, 77 patients were included, 66.20% females, with a mean age of 56±11 years old. There was a significant reduction of DAS28 only after 3 months of therapy (4.01±1.01 to 2.57±1.52, p=.003; ΔDAS28=1.58±1.17). However, during a median follow up time of 64 (IQR 39-83) months, 58.44% of patients discontinued this treatment strategy, 66.67% due to toxicity (median time to toxicity 13 months, IQR 2-16) and 33.33% due to inefficacy (median time to inefficacy of 10 months, IQR 5.84-17.64). Gastrointestinal intolerance was the main reported toxicity (46.15%). In univariate analysis, anti-citrullinated protein antibodies (ACPA) positivity, alcohol consumption, lack of comorbidities, hepatic toxicity, higher 6_DAS28, swollen joint count and tender joint count on the 6th month were associated to lower retention rates.
In multivariate analysis, lack of comorbidities (HR=3.3, CI 95% 1.4-7.8, p=.006) and higher 6_DAS28 (HR=0.32, CI 95% 0.14-0.72, p=.006) were independent predictors of suspension of combination therapy. Moreover, both male gender (HR=2.87, 95%CI 1.2-6.56, p=.016) and positivity to ACPA (HR=0.1, 95%CI 0.01-0.73, p=.024) were independent predictors of toxicity. There was also higher tendency to toxicity, but without statistical significance, in alcohol consumers (p=.08). Regarding inefficacy, smoking habits (HR=0.15, 95%CI 0.04-0.52) and 3_DAS28 (HR=0.15, 95%CI 0.04-0.53) were independent predictors.
Conclusions: Addition of LFN to MTX showed an early positive response. Yet, it was frequently associated to toxicity, and less than half of the patients continued with this therapeutic strategy after 5 years of follow up. Male gender, smoking habits and positivity to ACPA were predictors of worse outcome, as already reported in literature. Lack of comorbidities was an independent predictor of suspension of this strategy. This can be explained by the fact that physicians tend to adopt a more aggressive strategy on patients without comorbidities, switching earlier to bDMARDs/tsDMARDs.
This study also showed that early response to combination therapy is an independent predictor on drug retention, suggesting that decisions on treatment strategy should be made early after the beginning of MTX/LFN.

P0052 – PARANEOPLASTIC RHEUMATIC SYNDROMES: LESSONS FROM A MULTICENTER CASE SERIES

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Background: Paraneoplastic rheumatic syndromes are uncommon conditions whose exact pathogenic mechanism remains elusive, currently thought to be caused by tumor-related soluble factors or immune mechanisms. To be considered truly paraneoplastic, a causal effect should be established, concerning temporal relationship, pathogenic plausibility or proof of regression of rheumatic manifestations after successful tumor treatment.
Objective: To describe the epidemiological and clinical profile of patients diagnosed with rheumatic manifestations and temporally related malignancies in five tertiary care Portuguese hospitals.
Methods: A retrospective multicenter study was performed, which included all patients diagnosed with paraneoplastic rheumatic syndromes during the period 2008-2019. Demographic, clinical and outcome data were obtained from their medical records. Descriptive statistics were performed.
Results: During the study period, 30 patients were identified (Table 1). The majority were female (56%) with a mean age at the diagnosis of rheumatic manifestations of 65.0 ± 13.5 years. Most neoplasms were solid (70%), namely 33% of primary lung cancers. Rheumatic syndromes were diagnosed at the same time or before the malignancies in 63%. The time gap between the two diagnoses was inferior to 24 months in 97% of patients. Polyarthritis was the most common rheumatological manifestation (28%), followed by polymyalgia rheumatica and dermatomyositis (20% each). In patients with sclerodermiform syndromes and myositis, antinuclear antibodies (>1/160) were present only in 42%. Musculoskeletal manifestations resolved after cancer treatment in all patients for which partial or complete malignancy remission was achieved (28%).
Conclusion: In our multicenter case series, polyarthritis, polymyalgia rheumatica, and dermatomyositis were the most found rheumatic paraneoplastic manifestations. This association has been based on plausible temporal correspondence, clinical course, and response to cancer treatment. Effective treatment of malignancy is likely to lead to the improvement of the paraneoplastic rheumatic disease. The coexistence of malignancies and several rheumatic syndromes, although rare, is of utmost clinical relevance, as clinician's awareness may allow early detection and prompt treatment of neoplasms.

TABLE 1. PATIENTS' CLINICAL RHEUMATOLOGICAL AND NEOPLASTIC DIAGNOSIS AND MAXIMUM CANCER DIAGNOSIS DELAY FOR EACH RHEUMATIC SYNDROME

RHEUMATIC MANIFESTATION	NUMBER OF CASES	ASSOCIATED MALIGNANCIES	MAXIMUM CANCER DIAGNOSTIC DELAY
Polyarthritis	8	3 Lung adenocarcinomas 1 Gastric adenocarcinoma 1 Colorectal adenocarcinoma 1 Myelodisplastic syndrome 1 DLBCL 1 Multiple myeloma	8 months
Polymyalgia rheumatica-like	6	2 DLBCL 1 Prostatic adenocarcinoma 1 Multiple myeloma 1 CLL 1 Endometrial adenocarcinoma	22 months
Dermatomyositis	6	1 Nasopharingeal carcinoma 1 Endometrial carcinoma 1 Undifferentiated non-small cell lung carcinoma 1 Lung adenocarcinoma 1 Melanoma 1 Papillary thyroid carcinoma	12 months
Sclerodermiform syndrome	5	2 Breast adenocarcinomas 1 Colorectal adenocarcinoma 1 Lung adenocarcinoma 1 Follicular lymphoma	6 months
Scleroderma/ myositis overlap syndrome	1	Multiple myeloma	32 months
Polymyositis	1	Epithelial ovarian carcinoma	0 months
RS3PE syndrome	1	Gastric adenocarcinoma	4 months
Hypertrophic osteoarthropathy	1	Lung adenocarcinoma	0 months
Vascular acral syndrome	1	Typical lung carcinoid tumor	23 months

DLBCL- diffuse large B-cell lymphoma; CLL- chronic lymphocytic leukemia; RS3PE – Remitting Seronegative Symmetrical Synovitis with Pitting Edema.

P0056 – THE IMPACT OF TREATMENT WITH A BIOLOGICAL DISEASE-MODIFYING ANTIRHEUMATIC DRUG ON SPINAL MOBILITY AND ITS CORRELATION WITH DISEASE ACTIVITY IN PATIENTS WITH SPONDYLOARTHRITIS

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Background: Bath Ankylosing Spondylitis Metrology Index (BASMI) is an instrument developed to assess spinal and hip mobility. The relationship between BASMI and disease activity is not always linear and data that correlate the variation in BASMI values (ΔBASMI) with the variation in disease activity scores

and response to treatment are not unanimous.
Objectives: Explore the effect of biological disease-modifying antirheumatic drugs (bDMARD) in BASMI and the associations between ΔBASMI and disease activity.
Methods: Observational retrospective study was performed including consecutive patients with the diagnosis of Spondyloarthritis (SpA). Demographic, clinical, including Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), BASMI, Ankylosing Spondylitis Disease Activity Score with erythrocyte sedimentation rate and C-reactive protein (ASDAS ESR and ASDAS CRP, respectively), and laboratorial data were collected from our national database at baseline, 6 and 12 months after initiation of a bDMARD. The variation of each parameter was calculated as the difference between the levels recorded at 6 and 12 months and the reference level and presented in the form of **Δ**. Correlations between variables were studied using Spearman correlation analysis and comparison between groups was performed using Wilcoxon and Kruskal-Wallis tests, using SPSS 23.0.
Results: Median age of patients (n=178) was 42 years old [34, 50], 92 (51.7%) were males with a median disease duration of 4.9 [1.0, 10.3] years. One hundred and twenty-six patients (70.8%) had Ankylosing Spondylitis, 15 (8.4%) Inflammatory Bowel Disease related SPA and 30 (16.9%) Undifferentiated SpA. Fifty four (30.3%) patients were taking glucocorticoids and regarding conventional synthetic disease-modifying anti- rheumatic drugs use before starting the bDMARD: Sulfasalazine (52, 29.2%), Methotrexate (31, 17.4%) and Leflunomide (3, 1.7%). Regarding the bDMARD, only one patient started Secukinumab and the others a Tumor necrosis factor inhibitor (TNFi) [Golimumab (n= 64, 36.0%), Adalimumab (n=36, 20.2%), Infliximab (n= 35, 19.7%), Etanercept (n= 32, 18.0%) and Certolizumab (n= 10, 5.6%)].
The majority of the patients had very high disease activity at baseline (86.0%, n=153); median ESR was 29 mm/h [15, 47], median CRP was 13.7 mg/L, [6.60, 27.3], median ASDAS CRP was 7.6 [6.0, 9.0] and median BASMI was 8.0 [7-0, 9.0]. After 6 and 12 months of treatment, mean ESR, CRP, ASDAS-CRP and BASMI were lower than mean baseline values ($p<0.01$), with median ASDAS-CRP at 12 months of 2.20 [1.50, 2.90] and median ΔBASMI of -4.10 [-5.50, -2.40]. BASMI at baseline showed correlations with ASDAS CRP, BASDAI and patient visual analogic scale (VAS) ($r=0.468$, $r=0.496$, $r= 0.563$; $p<0.01$, respectively). No correlations were found between BASMI and CRP, ESR, physician VAS or the consumption of nonsteroidal anti inflammatory drugs at baseline.
Correlations were found between ΔBASMI and ΔASDAS at 6 months and 12 months ($r=0.243$, $p=0.02$; $r=0.286$; $p<0.01$) and also between ΔBASMI and ΔBASDAI at 6 and 12 months ($r=0.183$, $p=0.04$; $r=0.291$, $p=0.02$). No correlations were found between ΔBASMI and ΔCRP or ΔESR. No differences were observed in ΔBASMI, regarding the bDMARD of choice.
Conclusion: Starting a bDMARD improved BASMI scores through a 12 month period and as such, a TNFi may retard the progression of spinal mobility dysfunction. We cannot draw conclusions regarding differences between TNFi and interleukin 17 inhibitors and further work is needed to clarify possible differences in their impact in improving spine mobility.

P0058 – ESTABLISHED RHEUMATOID ARTHRITIS PATIENTS HAVE INCREASED FREQUENCIES OF FOLLICULAR REGULATORY T CELLS IN PERIPHERAL BLOOD

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Background: Several studies have demonstrated that an immune dysregulation affecting both B and T cells occurs in rheumatoid arthritis (RA). Follicular helper T (Tfh) cells are crucial for B cell maturation, activation and class-switching as well as for germinal center (GC) formation, whereas follicular regulatory T (Tfr) cells can modulate the GC reaction by suppressing Tfh and B cells.
Aims: The main goal of this study was to analyze the phenotype and frequency of circulating follicular T cell subsets in established RA patients.
Methods: Blood samples were collected from established RA patients with active disease, treated with methotrexate (n=32) and from a group of age and sex-matched healthy donors (n=11). Peripheral blood mononuclear cells (PBMC) were isolated and Tfh (CD4+CXCR5+CD45RO+) and Tfr (CD4+ CXCR5+CD25+FoxP3+) cells, as well as from their three major subsets [CXCR3+CCR6- (Th1-like), CXCR3-CCR6- (Th2-like) and CXCR3-CCR6+ (Th17-like)] were evaluated by flow cytometry.
Results: The frequency of circulating Tfh cells was similar between established RA patients and controls.

Nonetheless, RA patients had a decreased frequency of Th1-like Tfh cells, and an increased frequency of Th2-like Tfh cells when compared to controls. No differences were observed in the frequencies of Th17-like Tfh cells between both groups. The frequency of circulating Tfr cells was significantly increased in RA patients in comparison to controls. Furthermore, Tfr cells from RA patients had a significantly increased CD69 median fluorescence intensity (MFI) values when compared to controls. No significant differences were found in the percentages and MFI values of PD-1, ICOS, CD28, CTLA-4, CD40-L and HLA-DR expressed by Tfh and Tfr cells in RA patients when compared to controls.

Conclusions: Established RA patients have increased circulating frequencies of Tfr cells, with higher CD69 expression levels, when compared to healthy controls. These results suggest a pre-activation state of Tfr cells in RA and a potential role in the disease physiopathology.

*RA Moura, JE Fonseca and LGraça are joint senior authors.

P0059 – BONE MINERAL DENSITY AND FRACTURE RISK IN A COHORT OF PORTUGUESE SYSTEMIC SCLEROSIS PATIENTS

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Background: Patients with Systemic Sclerosis (SSc) seem to have higher prevalence of low bone mineral density (BMD) and an increased spine fracture risk. We aim to determine, by conventional densitometry (DXA) and using the fracture risk assessment tool (FRAX), the prevalence of low BMD and the fracture risk, respectively, in a SSc cohort and its potential determinants.

Methods: Transversal study was performed including consecutive patients with the diagnosis of SSc. We collected data regarding demographics, BMD (lumbar spine and femoral neck) and occurrence of fracture. Ten-year risk of osteoporotic fracture was estimated using FRAX v4.1 with the Portuguese population reference. Statistical analysis was performed using SPSS 23.0; $p<0.01$ was considered statistically significant.

Results: Median age of patients (n=97) was 62 years old [56, 70], 88.7% females (n=86). Seventy-eight patients (80.4%) had limited cutaneous form, 5 (5.2%) presented a diffuse cutaneous form and 13 (13.4%) an overlap syndrome. Regarding clinical features: digital ulcers in 30 patients (30.9%), interstitial lung disease (ILD) in 16 (6.5%), gastrointestinal involvement in 16 (16.5%), miositis in 4 (4.1%) and pulmonary arterial hypertension in 3 (3.1%). Anti-topoisomerase I antibody (anti-Scl70) positivity was present in 15 patients (15.5%) and anti-centromere antibody (ACA) positivity in 63 (64.9%). Nine patients (9.3%) were smokers and 6 (6.2%) reported an alcohol consumption of 3 or more units/day. Median body mass index (BMI) was 25.4 Kg/m2[21.4, 29.1] and vitamin D insufficiency was reported in 19 patients (19.6%). Twenty-one patients (21.6%) have been exposed to oral glucocorticoids (GCT) for more than 3 months. Eleven patients (11.3%) had previous low impact fractures: 10 of which were vertebral and 1 wrist fracture. Low BMD was present in 45 patients (46.4%); median femoral neck BMD (FN-BMD) was 0.827 [0.709, 0.893].Ten year probability of fracture (%) was: median risk for major fracture was 5.1 [3.5, 9.7] and 3.8 [2.5, 8], with and without FN-BMD, respectively; for hip fracture the estimated risk was 1.2 [0.6, 3.1] and 1.0 [0.4, 2.5], with and without FN-BMD, respectively. According to FRAX thresholds for the Portuguese population, 25 patients (25.8%) met criteria to start AOP treatment, but only 10 patients (40%) started it, as the agreement between the indication to treat by FRAX and the onset of treatment was weak (k= 0.338). No agreement was found between FRAX risk without DXA and WHO threshold. FN-BMD presented a correlation with BMI (r = 0.393), an inverse correlation with major fracture risk with and without FN-BMD (r = -0.704, r=-0.412, respectively) and with hip fracture risk with and without FN-BMD (r = -0.799, r=-0.412, respectively). Major fracture risk with and without FN-BMD presented correlation with spine fractures (r = 0.350; r=0.397, respectively). No correlation was found between WHO threshold and spine fractures. No correlations were found between FN-BMD or fracture risk estimated by FRAX and disease manifestations, anti-Scl70 or ACA positivity, smoking or GCT use.

Conclusion: Low BMD was prevalent and had correlation with BMI. FRAX appears to be an useful instrument as it correlated with spine fractures, contrary to what was verified when we used the WHO threshold. Estimating fracture risk using FRAX appear to be an useful tool for the prevention of fractures in this population.

P0060 – THE USE OF A COMORBIDITY INDEX FOR PREDICTING CLINICAL RESPONSE IN RHEUMATOID ARTHRITIS PATIENTS RECEIVING THEIR FIRST BIOLOGICAL AGENT

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Background: Several studies in Rheumatoid arthritis (RA) have suggested that a greater number of comorbidities is associated with worse functional status and disease activity measures. To better understand the overall role of comorbidities in treatment outcomes it could be useful to use a composite comorbidity index, such as Rheumatic Disease Comorbidity Index (RDCI) that is validated for the use in patients with rheumatic diseases.

Objectives: To evaluate the impact of comorbidities in clinical response in a cohort of patients with RA treated with a first-line biologic disease-modifying antirheumatic drug (bDMARD), by using the RDCI.

Methods: Observational retrospective study was performed including consecutive patients with the diagnosis of RA. The prevalence of comorbidities was computed, and patients were stratified according to RDCI for evaluating its role in clinical response disease activity at baseline and follow up (6 and 12 months). Statistical analyses were performed using SPSS, 23.0.

Results: We included 251 patients: 83.7% (n=210) females, mean age of 58 (± 11.10) years old, with a median disease duration of 16.11 years [10.79 – 23.04]. The majority exhibited a very high or high disease activity at baseline (median DAS28 3V 5.48 [4.70 – 6.19]) and 90% (n=226) of them were concomitantly using corticosteroids and/or other disease-modifying anti-rheumatic drugs (129 with methotrexate (MTX), 96 with leflunomide and 35 with sulfasalazine). The most frequently reported comorbidities were cardiovascular disorders (37.5%), osteoporosis (7.6%) and depression (6.8%). The median RDCI score was 1.0 [0.0 – 2.0] and the majority of patients (63.6%) carried at least one comorbidity. When comparing baseline demographic and clinical characteristics of the 4 subgroups, stratified according to RDCI score (RDCI=0, 1, 2, or ≥3), we found statistically significant differences in age, age at diagnosis, sex and the prescribed anti-TNF agent ($p<0.05$). There was a progressive increase in the mean age as the RDCI score increased between the subgroups. Number of comorbidities (NC) was correlated with patient and physician global assessment of disease activity (pVAS and phVAS) ($r=0.183$, $p<0.01$ and $r=0.196$, $p=0.019$, respectively), DAS28 3V ($r=0.192$, $p=0.046$) and HAQ-DI ($r=0.301$, $p<0.01$) at 6 months. Moreover, RDCI correlated with CRP ($r=0.192$, $p=0.01$), pVAS ($r=0.183$, $p=0.02$) and HAQ-DI ($r=0.202$, $p<0.01$). Weaker correlations were also found at 12 months: NC with DAS28 3V ($r=0.216$, $p=0.01$) and HAQ-DI ($r=0.187$, $p=0.04$); RDCI with phVAS ($r= 0.196$, $p=0.04$). The 12-month DAS28 remission rate was 37.8% (n=95); 6.7% (n=17) achieved EULAR good response and 54.4% (n=137) a moderate EULAR response. RDCI was not an independent predictor of DAS remission (OR 0.794, 95% CI 0.561- 1.125, $p = 0.194$) nor it was of EULAR good/moderate response (OR 0,720, 95% CI 0.430- 1.206, $p= 0.212$).

Conclusion: Although our data point to a weak association between morbidities, assessed by the RDCI, and response to a first bDMARD, it is important to consider this simple and useful tool in future prospective and broader studies, since information bias regarding comorbidities may have been responsible for our results.

P0064 – PHYSICIAN'S GLOBAL ASSESSMENT OF DISEASE ACTIVITY IN RHEUMATOID ARTHRITIS IS A RELIABLE AND RESPONSIVE TOOL IN CLINICAL PRACTICE

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Background: Physician's global assessment of disease activity (PhGA) is highly influential upon treatment decisions taken by rheumatologists, surpassing the impact of DAS28. [1, 2]. However, data regarding its psychometric properties are scarce.

Objectives: To evaluate the reliability and responsiveness of PhGA.

Methods: We included two consecutive visits of RA patients followed in a Tertiary Rheumatology Department. Socio-demographic (age and gender) and clinical data were collected including tender (TJ28) and swollen (SJC28) joints in 28 count, C-Reactive Protein (CRP), Erythrocyte Sedimentation Rate (ESR), Disease activity Score (DAS28-3v-CRP, DAS28-3v-ESR, DAS28-4v-CRP, DAS28-4v-ESR), PhGA and Patient Global Assessment of disease Activity (PGA) through a Visual Analogic Scale (VAS) 0-100mm. Changes (Δ) between the two visits were calculated. Only patients without missing data were included. Correlations between ΔPhGA and change of other variables were assessed using Pearson's correlations. Reliability was evaluated through Intraclass Correlation Coefficient (ICC) between two consecutive appointments in a

subgroup of patients with stable disease activity (Δ DAS28-4vESR [-0.6 to 0.6]. An ICC above 0.8 was considered indicative of excellent reliability. Sensitivity to change was assessed in the subgroup of patients who improved their disease activity at least 0.6 on DAS28-4V-ESR, through Standardized Response Mean (SRM). The respective intervals of confidence were obtained through bootstrapping procedures. SRM above 0.8 were considered large. Independent factors associated with ΔPhGA were identified through multivariate linear regression analysis. p<0.05 was considered statistically significant

Results: 121 RA patients (84.3% female and 64.0±12.6 years) were included. Δ PhGA was weakly correlated with ΔCRP (r=0.23), Δ PGA (r=0.31) and Δ pain (r=0.37). Moderate to strong correlations were observed with Δ DAS28-3V-ESR (r=0.55), Δ SJC28 (r=0.56), Δ DAS28-3V-CRP (r=0.58), Δ DAS28-3V-CRP (r=0.60), Δ TJ28 (r=0.62) and ΔDAS28-4V-CRP (r=0.63). ICC between two consecutive visits was 0.7, [95%CI:0.47-0.83] and SRM was -1.01 [95%CI:-1.26-(-0.73)]. In the multivariate regression analysis, ΔSJC28 (β=4.01; 95% CI:3.07 to 4.96) and Δ Pain (β=0.18; 95%CI: 0.07 to 0.28) remained as independent factors associated with ΔPhGA (R2:0.49, p<0.01).

Conclusion: In this study, PhGA showed a high reliability and sensitivity to change regarding disease activity, in clinical practice. Changes in SJC had the strongest association with change in PhGA scoring, but Δ Pain was also significantly correlated.

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P0066 – PHYSICIAN'S GLOBAL ASSESSMENT OF DISEASE ACTIVITY IN RHEUMATOID ARTHRITIS: WHAT DO WE REALLY MEAN?

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Background: Physician's global assessment of disease activity (PhGA) is included in some scores of disease activity and, demonstrably, plays a major role upon treatment decisions in rheumatoid arthritis (RA) [1, 2, 3]. Therefore, understanding the reasons underlying the physician's assessment is crucial.

Objectives: To understand the reasons underlying the physician's assessment.

Methods: Cross-sectional study, including consecutive RA patients followed in a Tertiary Rheumatology Department.

Socio-demographic (age and gender) and clinical data were collected through a standardized protocol, including 28 tender (TJ28) and swollen (SJC28) joints count, C-Reactive Protein (CRP), Erythrocyte Sedimentation Rate (ESR), Disease activity Score (DAS28-4v-CRP and DAS28-4v-ESR), PhGA and Patient Global Assessment of disease Activity (PGA) through a Visual Analogic Scale (VAS) 0-100mm, Health Assessment Questionnaire (HAQ), European Quality of Life-5 Dimensions (EQ-5D) and Hospital Anxiety and Depression Scale (HADS). Correlation between PhGA and other continuous variables was evaluated through Pearson's Correlation Coefficient and variables with p<0.05 in univariate analysis were included in multivariable linear regression (stepwise model).

Results: 392 RA patients (80.6% female, 65.3±12.6 years) were included. PhGA was weakly correlated with CRP (r=0.23), TJC28 (r=0.35), PGA (r=0.26), HAQ (r=0.31) and EQ5D (r=-0.21). Moderate correlations were observed with SJC28 (r=0.45) and DAS-4V-CRP (r=0.48). In multivariable analysis, SJC28 (β=4.14, 95%CI:3.16-5.12), CRP (β=0.22; 95%CI: 0.02-0.03), HAQ (β=4.46, 95%CI:1.50-7.42) and PGA (β=0.08; 95%CI:0.00-0.16) remained as independent correlates of PhGA (R2=0.27, p<0,05).

Conclusion: In this study, PhGA was associated with SJC28, CRP, HAQ and PGA, suggesting that physicians adopt a comprehensive reading of the disease into account. However, a large proportion of the variance of PhGA remains unexplained. Given its driving role in treatment decisions, the need to standardize and better understand PhGA seems to deserve a closer attention.

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P0067 – BEYOND DISEASE ACTIVITY, PAIN, "TIME" AND "TIMING" ACCOUNT FOR DISABILITY IN PATIENTS WITH RHEUMATOID ARTHRITIS: RESULTS FROM A REAL-LIFE COHORT

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Background: Patients with rheumatoid arthritis (RA) suffer from joint pain, stiffness and fatigue and are therefore limited in their physical activities. Since functional disability is a major determinant of quality of life in patients with RA, an optimized approach should focus on the maintenance of functional ability.
Objectives: To evaluate self-reported disability and to identify its influencing clinical and demographic factors in a real-life cohort of patients with RA.
Methods: Cross-sectional study of consecutive patients with RA fulfilling the ACR/EULAR'10 and/or ACR'87 RA classification criteria, followed in a tertiary care centre. Variables collected included socio-demographic and clinical variables (disease duration; time from symptoms onset to diagnosis, classified as short (≤2 years) and long (>2 years); time of diagnosis, categorised as <2000,2000-2009,≥2010); DAS28-CRP-3V and its individual components; pain assessed through VAS (0-100 mm) and self-perception of anxiety/depression through EQ5D dimension 5. Disability was assessed through Health Assessment Questionnaire (HAQ) score and categorised as none-to-mild (<1) or moderate-tosevere (1-3). Comparison between groups was assessed through chi-square or T-student test, as adequate. Variables with p<0.1 and others clinically relevant in the researcher's perspective were included in a multivariable logistic regression model. Given the implementation of new strategies regarding diagnosis and treatment of RA in the last decade, a subgroup analysis was performed for patients with diagnosis performed after 2010).
Results: A total of 251 patients were included (78.9% female, aged 62.0±12.1 years, disease duration 16.7±11.2 years), with a mean DAS28-CRP-3V of 2.24 ±0.87, with 65.3% being in remission or low disease activity. The mean HAQ score was 1.2±0.8. Over half of the patients (56.2%) reported moderate-to-severe disability. In the univariate analysis, moderate-to-severe disability was more frequent in female patients (60.6% vs 39.6%, p<0,006), in patients with moderate-to-severe self-perception of anxiety/depressive symptoms (67.2% vs 44.2%,p<0.001) and in patients with diagnosis before the year 2000, 2000-2009 than ≥2010 (71.4% vs 63.1% vs 36.7%;p< 0.001). In addition, patients with moderate-to-severe disability tended to be older (65.05 vs 57.98,p<0,001), to have longer disease duration (20.07 vs 12.39,p<0.001), to report more pain (VAS 58.08 vs 28.62, p<0.001) and to have higher disease activity (2.48 vs 1.95,p=0.001). In the multivariable analysis, pain (OR=1.04; 95%CI 1.03-1.06, p<0.001), disease activity (OR=1.51; 95%CI 1.01-2.26, p=0.049), and time of diagnosis (OR=0.553, 95%CI 0.38 -0.81, p=0.002) remained as independent factors associated with moderate-to-severe disability (R2: 0.40,p<0,001). In the subgroup of patients diagnosed after 2010, a longer time to diagnosis (>2 years) (OR=7.97, 95%CI 1.88-34.06;p=0.005) and pain (OR=1.05, 95%CI 1.03-1.08;p<0,001) remained as independent factors (R2=0.44,p<0.001).
Conclusion: Functional disability remains a major problem in our patients with RA, despite clinical remission. Beyond non-modifiable factors, disease activity and pain are associated with higher disability. Moreover, in the subgroup of patients diagnosed after 2010 a long time to diagnosis was the major predictor of disability. However, a large variance of the reported functional disability remains unexplained. Hence, other factors should be properly evaluated in our patients in order to achieve a more holistic approach aiming at reducing functional disability.

P0077 – MESOTHERAPY FOR THE MANAGEMENT OF MUSCULOSKELETAL PAIN: A SINGLE-CENTRE EXPERIENCE

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Introduction: Over the last years, mesotherapy has been increasingly used as an alternative therapeutic strategy for the management of pain in locoregional disorders. It is generally well accepted by the rheumatology community and patients.
Objectives: To evaluate the effectiveness and safety of mesotherapy in musculoskeletal disorders in the rheumatology setting.
Methods: A prospective, observational study was conducted in our Rheumatology department. Adult patients with local musculoskeletal pain, with indication for undergoing mesotherapy by their rheumatologist were included. All patients were submitted

to 3 or 4 weekly sessions of mesotherapy, in which they received local microinjections of combined lidocaine (1 ml), piroxicam (1 ml) and thiocolchicoside (1 ml). Data were collected in each session and at 3 months of follow-up. Pain was assessed by the Visual Analogue Scale (VAS) 0-10 cm. Paired sample t-test was used to compare VAS scores. Fisher's exact test, chi-square test or independent t-test were used to compare differences between groups (reduction in VAS>=50% or <50%).

Results: A total of 53 patients were included, 92.5% were females. Mean age was 63.8 ± 12.7 years and mean pain duration was 12.1 ± 8.0 months. Treatments were performed in the lumbar (35.8%) and cervical regions (24.5%), shoulders (18.9%), trapezius muscle (13.2%), hip (3.8%), dorsal region (1.9%) and knee (1.9%). Most patients were diagnosed with periarticular disorders (30.2%) or muscle contractures (30.2%), followed by pain due to degenerative disease such as osteoarthritis or discopathy (26.4%) or fibromyalgia (13.2%). Nearly 72% of patients completed 4 sessions of mesotherapy, while 28% failed to complete them, being submitted only to 3 sessions.

At baseline, the overall mean VAS score was 7.2 ± 1.3. There was a statistically significant reduction in the VAS score at the second (6.6 ±1.5, p=0.010), third (4.9 ± 2.0, p<0.001) and fourth (3.8 ± 2.3, p<0.001) weeks, compared to the baseline values. At three months of follow-up, the mean VAS score remained significantly lower than baseline levels (4.1 ± 2.3 vs 7.2 ± 1.3, p<0.001), although there was a rise, in absolute terms, compared to the VAS score at the fourth session (4.1 ± 2.3 vs 3.8 ± 2.3, p=0.081).

Twenty-six patients (49.1%) presented a VAS score reduction of at least 50% at three months of follow -up. These patients had a shorter pain duration (8.5 ± 7.0 vs 15.6 ± 11.5 months, p=0.012) and more often completed the four sessions of mesotherapy (92.3% vs 51.9%, p=0.001), compared to those whose reduction in the VAS score was less than 50%. Moreover, there was a significant difference in the percentage of patients who achieved a reduction in VAS>=50% depending on the diagnosis (68.8% of patients with muscle contractures, 50% with periarticular disorders, 42.9% with degenerative disorders and 14.3% with fibromyalgia; p=0.048).

In addition, in patients with muscle contractures, a significant reduction in the VAS score was already detected from the second week (6.1 ± 1.5 vs 7.1 ± 1.2, p=0.005); in all other cases, this was only found from the third week.

Few reversible adverse effects were reported: ecchymosis or hematomas in 19 patients (35.8%) and a local allergic reaction in 1 case (1.9%).

Conclusion: Mesotherapy is an effective and safe treatment for the management of musculoskeletal pain. In our cohort, benefits in terms of pain reduction were observed, particularly in patients with a short pain duration, diagnosed with muscle contractures or those who completed four sessions of mesotherapy.

P0080 – FUNCTIONAL DISABILITY AND PAIN BUT NOT DISEASE ACTIVITY ARE ASSOCIATED WITH POOR HEALTH-RELATED QUALITY OF LIFE IN A COHORT OF RHEUMATOID ARTHRITIS PATIENTS

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Background: Rheumatoid Arthritis (RA) is a systemic autoimmune disease that presents with joint pain and inflammation leading to significant disability and poor health-related quality of life (HRQoL) (1,2). Optimizing long-term HRQoL is the primary goal of disease management in RA (3).

Objectives: To evaluate HRQoL and identify its influencing clinical and demographic factors in a Portuguese RA population.

Methods: This is a cross-sectional study including consecutive patients fulfilling the ACR/EULAR 2010 and/or ACR 1987 RA classification criteria, followed at a tertiary Rheumatology Department. Sociodemographic and clinical variables were collected. HRQoL was assessed using the EuroQoL 5-Dimensional Descriptive System (EQ-5D) total score (normal range from -0.496 to 1.000, lower values indicating poorer HRQoL). Independent t-test and Pearson's correlation coefficient were performed to evaluate EQ-5D differences between groups and examine its relationships with continuous variables, respectively. Variables with p<0.1 in univariate analysis were included in a stepwise multiple linear regression analysis to evaluate the independent association of variables with the EQ-5D score.

Results: 358 RA patients were included (80.20% female, mean age ± SD: 63.22 ± 0.66 years old). Mean EQ5D total score ±SD was 0.48 ± 0.01. Based on EQ-5D domains, 0.60% reported extreme problems with mobility, 3.40% extreme problems with self-care, 2.50% extreme problems with usual activities, 12.0% extreme pain or discomfort, and 7.30% extreme

anxiety or depression symptoms (Fig.1). There was a significant difference in EQ-5D scores between male (M=0.55, SD=0.24) and female gender (M=0.46, SD= 0.27); t(356)= -2.41, p=0.016. EQ-5D was weakly correlated with DAS-28-CRP (r=-0.32; p<0.001), moderately correlated with patient's global assessment of disease activity (r=-0.54; p<0.001) and pain-visual analogue scale (pain-VAS) scores (r=-0.58; p<0.001) and strongly with Health Assessment Questionnaire (HAQ) score (r=-0.72; p< 0.001). After multivariate analysis, HAQ-score (β=-0.57 [95% CI -0.24 to -0.17]; p<0.001) and pain-VAS ((β=-0.25 [95% CI -0.003 to -0.002]; p<0.001) remained as independent predictors of EQ-5D (R2=0.56, p<0.001).

Conclusions: Greater functional impairment and pain are associated with poor HRQoL in RA patients, and thus special attention must be given to treatment strategies providing the best patient-centred outcomes.

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P0081 – EVALUATION OF ENTHESITIS INDICES AND RESPONSE TO BDMARD THERAPY IN PORTUGUESE PATIENTS WITH SPONDYLOARTHRITIS

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Background: Enthesitis is a hallmark clinical feature of spondyloarthritis (SpA), but to date, few studies have investigated how the overall response to biological treatment relates to the evolution of enthesitis counts.

Objectives: Assess whether the variation in enthesitis indices reflects the overall response to bDMARD therapy in SpA.

Methods: This study included patients who met ASAS criteria for SpA followed at our Rheumatology Department under bDMARD therapy. Demographic, laboratorial and clinical data were collected, including Bath Ankylosing Spondylitis Metrology Index (BASMI), Bath Ankylosing Spondylitis Functional Index (BASFI), Bath Ankylosing Spondylitis Activity Index (BASDAI), Ankylosing Spondylitis Disease Activity Score-C-reactive protein (ASDAS-CRP), Maastrich Ankylosing Spondylitis Enthesitis Score (MASES), Leeds Enthesitis Index (LEI) and Spondyloarthritis Research Consortium of Canada (SPARCC) – LEI was included at a later stage in Reuma.pt, so it was calculated without medial femoral condyles entheses. All were evaluated at baseline and at 6, 12, 18 and 24 months after starting the first bDMARD. The variation in each parameter compared with the baseline values was calculated at 6, 12, 18 and 24 months and represented in the form of delta. Correlations between variables were assessed using Spearman test and comparison between groups using Wilcoxon, Mann-Whitney U and Kruskal-Wallis tests.

Results: We included 273 patients, 123 (45,1%) females, aged 42,0±12,3 years and with diagnosis of SpA for 15,4±11,2 years at the start of bDMARD therapy. At baseline, mean BASDAI was 6,43 ±1,62, ASDAS-CRP was 4,01 ± 0,86, median MASES was 1 (0-4), LEI 0 (0-1,75) and SPARCC 1 (0-4). Seventy-two patients (26,4%) started golimumab, 71 (26,0%) adalimumab, 66 (24,2%) infliximab, 54 (19,8%) etanercept, 9 (3,3%) certolizumab and 1 (0,4%) secukinumab. The indices were significantly higher at baseline in females [median MASES-females 2 (0-5) vs 0 (0-2), p<0,001; LEI-females 0 (0-2) vs 0 (0-1), p=0,03; and SPARCC-females 2 (0-5) vs 0 (0-2), p<0,001], and remained so at 24 months [median MASES-females 1 (0-3,5) vs 0 (0-0), p<0,001; LEI-females 0 (0-0,5) vs 0 (0-0), p<0,001; and SPARCC-females 1 (0-3) vs 0 (0-0), p<0,001]. There was a significant difference between each of the 3 indices when assessed at 6, 12, 18 and 24 months, compared to baseline (p <0.004). No differences were observed regarding the choice of bDMARD. At baseline, MASES had a significant correlation with patient visual analogic scale (pVAS) (r=0,18; p=0,01), BASDAI (r=0,36; p<0,001) and BASFI (r=0,21; p=0,003); LEI had a significant correlation with BASDAI (r=0,31; p<0,001) and BASFI (r=0,21; p=0,003); SPARCC had a significant correlation with pVAS (r=0,19; p=0,01), BASDAI (r=0,37; p<0,001) and BASFI (r=0,26; p<0,001). ΔLEI at 6 months had a significant correlation with ΔBASDAI (r=0,25; p=0,005), ΔASDAS (r=0,190; p=0,03), ΔpVAS (r=0,23; p=0,01) and Δphysician VAS (r=0,25; p=0,01), but not with ΔESR, ΔCRP and ΔBASMI; no correlation was found at 6 months for ΔMASES or ΔSPARCC. At 12 months, ΔMASES had a significant correlation with ΔBASDAI (r=0,18; p=0,03); ΔLEI with ΔBASDAI (r=0,23; p=0,01) and ΔpVAS (r=0,19; p=0,03); for ΔSPARCC no significant correlations were found. At 18 months and 24 months, no correlations were found.

Conclusion: The initiation of bDMARD led to improved enthesitis indices over a 24-month period. ΔLEI

correlates better with SpA activity scores and measurements than the other indices, especially at the first 12 months of initiation of bDMARD therapy.

P0082 – OSTEOPOROSIS TREATMENT IN PORTUGUESE PATIENTS WITH PSORIATIC ARTHRITIS – WHAT IS THE VALUE OF THE FRACTURE RISK ASSESSMENT TOOL (FRAX)?

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Background: Few studies have evaluated the prevalence and treatment of osteoporosis (OP) in patients with psoriatic arthritis (PsA), and many of these patients are not screened using dual-energy X-ray absorptiometry
(DXA). FRAX makes it possible to stratify the risk and define which patients may benefit from anti-osteoporotic treatment, but its usefulness in this population is not well established.
Objectives: The aim of this study was to determine whether the application of FRAX changes the indication for anti-osteoporotic treatment in PsA patients, according to the Portuguese guidelines.
Methods: In this cross-sectional study, we evaluated PsA patients from a tertiary hospital, registered in a national database (Reuma.pt), aged between 40 and 90 years and with a last consultation in 2019. FRAX was applied in all of them, regardless of being under anti-osteoporotic treatment and, when DXA was available, the femoral neck bone mineral density was used. Patients were stratified according to the risk of fracture, and those at high risk were considered candidates for anti-osteoporotic treatment, according to national guidelines [FRAX ≥11% for major osteoporotic fracture (MOF) or ≥ 3% for hip fracture (HF) without DXA; FRAX ≥9% for MOF or ≥ 2.5% for HF, with DXA].
Results: We included 100 patients, 52 females, with a mean age of 54,4 ±8,9 years and a median disease duration of 10 (6-17) years. Only 43 had already performed DXA and 6 had OP according to World Health Organization criteria. Seven patients were identified as having a high risk of fracture; applying femoral neck bone mineral density, 2 more patients with indication for treatment were recognized, totalizing 9 patients. There was a low agreement between the indication for treatment based only on DXA and FRAX (Cohen's k 0.066). There was a strong and significant correlation between percentage of risk of MOF by FRAX with and without DXA
Spearman's ρ 0.804, p <0.001); for the risk of HF by FRAX with and without DEXA the correlation was weaker but still significant (Spearman's ρ 0.439, p = 0.004). There was no association between the indication for treatment by FRAX and the performance of DXA (chi-square test, p = 0.597), nor the fact of performing DXA significantly affected the risk of MOF (Wilcoxon test, p = 0.185) or of HF (Wilcoxon test, p = 0.785) by FRAX.
Conclusion: In line with Portuguese guidelines, FRAX seems to be, in itself, a very useful tool in patients with PsA, and the performance of DXA does not significantly alter the indication for anti-osteoporotic treatment.

P0086 – REVISITING THE REMISSION CRITERIA FOR RHEUMATOID ARTHRITIS BY EXCLUDING PATIENT GLOBAL ASSESSMENT: AN INDIVIDUAL PATIENT META-ANALYSIS INCLUDING 5792 PATIENTS

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Background: Remission nowadays is the guiding target of management of rheumatoid arthritis (RA). However, no gold standard definition of remission exists and the inclusion of patient global assessment of disease activity (PGA) in remission definitions is debated.
Objectives: To determine the impact of excluding

TABLE 1: POOLED OUTCOMES AND MEASURES OF ASSOCIATION BETWEEN REMISSION CATEGORIES AND GOOD RADIOGRAPHIC OUTCOME (GRO, DEFINED AS ΔMTSS≤0.5), DURING THE SECOND YEAR OF FOLLOW-UP

Groups	*4V-remission (n=1,378)*	*4V-near-remission (n=1,085)*	*Non-remission (n=3,329)*
Percentage GRO (95%CI)	81 (74 to 87)	78 (70 to 86)	72 (62 to 81)

Comparisons	*4V-near-remission vs 4V-remission*	*4V-near-remission vs Non-remission*
Δ percentage GRO (95%CI)	−2.9 (−7.3 to 1.5)	6.2 (2.3 to 10.1)
Odds Ratio GRO (95%CI)	0.86 (0.68 to 1.07)	1.33 (1.11 to 1.60)

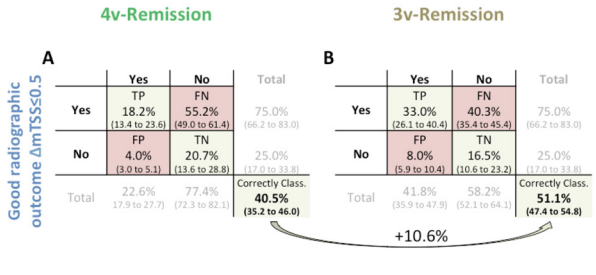
PGA from the ACR/EULAR Boolean remission criteria, upon prediction of radiographic outcome of RA.

Methods: Meta-analyses using individual patient data from randomized controlled trials (identified through PubMed and Clinicaltrials.gov until December 2016) testing the efficacy of biological agents on radiographic outcomes at ≥2 years. Remission was defined using the ACR/EULAR remission criteria with 4 variables: (i) tender and swollen 28-joint counts (TJC28/SJC28), C-reactive protein (CRP, mg/dl), and PGA≤1 (0-10=worst) all ≤1 (4V-remission), (ii) the same, except PGA>1 (4V-near-remission) (iii) 3V-remission (similar to 4V, but without PGA, i.e. equal to the combination of i) and ii), and (iv) non-remission (TJC28 and/or SJC28 and/or CRP >1). Meta-analyses were performed using the DerSimonian-Laird random-effects method. Good radiographic outcome (GRO) was defined as an increase of ≤0.5 modified Total Sharp score (mTSS) units. The relationship between the most stringent remission class achieved at 6 or 12 months and GRO during the second year was analysed. The pooled probabilities of GRO for the different definitions of remission were estimated and compared.

Results: Individual patient data (n=5,792) from eleven trials were analysed. 4V-remission was achieved by 23% of patients (95%CI: 18-28%) and 4V-near-remission by 19% (95%CI: 15-22%) and thus, 3v remission by 42% (95%CI: 36-48%). The probability of GRO in the 4V-near-remission group was similar to that of 4V-remission (78 vs 81%, ns) and significantly higher than that for non-remission (72%; difference 6%; 95%CI: 2-10%) (Table 1). These results were confirmed by meta-analyses of odds ratios of obtaining GRO of these groups (Table 1). 3V-remission showed a higher predictive value for GRO (51%, 95%CI: 47-55%) than 4V-remission (41%, 95%CI: 35-46%) (Figure 1).

Conclusions: 4V-near-remission and the original 4V-remission are similarly predictive of GRO, therefore combining these in the 3V-remission definition potentially reduces the risk of overtreatment compared to the 4V definition. This supports the use of 3V-remission as the target for immunosuppressive therapy. The patient's perspective, which must remain central, requires a separate treatment aim: a dual-target approach.

FIGURE 1 – POOLED META-ANALYTIC PREDICTION ACCURACY OF 4V- AND 3V-REMISSION STATUS FOR GOOD RADIOGRAPHIC OUTCOME (N=5,792 ANALYSED PATIENTS)



Legend: 4V-remission = SJC28. TJC28, CRP (in mg/dl), and PGA (0-10), all <1; 3V-remission=SJC28, TJC28, CRP (in mg/dl) <1; |mTSS = change in the modified Total Sharp Score from 12 months to 24 months. TP = True Positive; TN = True Negative; FP = False Positive; FN = False Negative; Correctly classified = TP + TN. Between brackets is the pooled 95% confidence interval.

Note: The sum of the meta-analytics percentages of TP, FN, FP, and TN is slightly Less than 100%, due to error estimation when multi-category (k>2) prevalence is estimated. We used in all meta -analyses double arcsine transformation as the preferred method to correct for this effect as much as possible.

P0088 – PATIENT GLOBAL ASSESSMENT LEVELS PRECLUDE THE MAJORITY OF RHEUMATOID ARTHRITIS PATIENTS OTHERWISE IN REMISSION, FROM REACHING THIS STATUS: SYSTEMATIC LITERATURE REVIEW AND META-ANALYSES INCLUDING 23,297 PATIENTS

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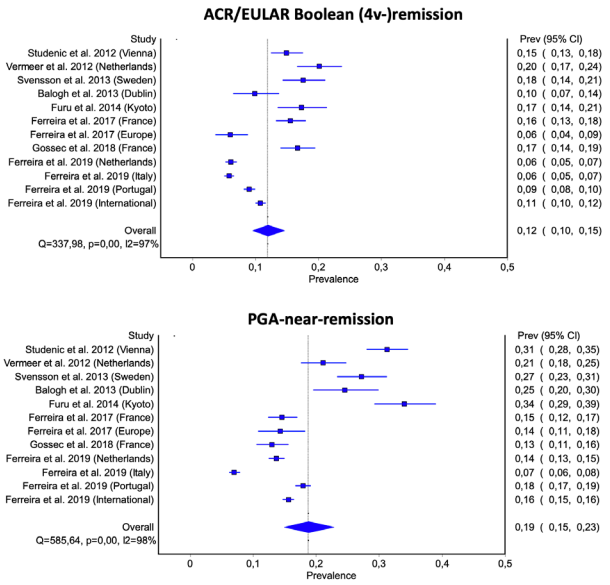
Background: Based on the ACR/EULAR Boolean-based remission definition for rheumatoid arthritis (RA), the main reason for near-misses (failing Boolean remission solely due to one criterion >1) is a patient

FIGURE 1 – META-ANALYSES OF PREVALENCE OF ACR/EULAR BOOLEAN-BASED AND PGA-NEAR-REMISSION IN CLINICAL COHORTS



4V-remission = SJC28, TJC28, CRP (mg/dl), and PGA (0-10), all ≤1; PGA-near-remission= the same, except PGA>1.

global assessment (PGA) >1/10 [1-3]. The frequency of this PGA-near-remission status has not been well established.

Objectives: To synthesize the overall prevalence of ACR/EULAR Boolean-based remission and PGA-near-remission cross-sectionally in published studies, and to explore association with disease duration.

Methods: We systematically searched PubMed database (from 1/jan/2011 till 15/jan/2020) for "Rheumatoid arthritis" AND Boolean [OR synonyms] AND PGA [OR synonyms], and carried out a hand search of the references of the included studies. Two reviewers independently assessed the study inclusion and extracted the data (n (%) patients in each remission status, mean (SD) disease duration, and country/city). Studies were excluded if any of the four Boolean criterion was not considered (e.g. CRP not assessed), or if only patients with low disease activity or remission were selected. If different time assessments were provided, we selected the 1y follow-up, as the most common. Random effects meta-analyses of proportions with double arcsine transformation were performed (also by disease duration subgroups) using MEDCALC®. Heterogeneity was assessed by I2.

Results: From 41 studies identified, 8 studies concerning 12 subsamples were analysed (n=23,297 patients; of which 22% had ≤2 years mean disease duration). The overall prevalence of Boolean remission was 12% (95%CI: 10-15%, p<0.005) compared to 19% of PGA-near-remission (15-23%, p<0.005) (Figure 1). In patients with shorter disease duration, PGA-near-remission was more prevalent (14% vs 18%), but even more so in longer disease duration (11% vs 19%).

Conclusions: The overall prevalence of PGA-near-remission in patients with RA followed in clinical practice was 1/3 more prevalent than Boolean remission, a difference more pronounced in patients with established disease. Overall, over 61% of all RA patients otherwise in remission failed to satisfy the Boolean definition of remission solely due to a PGA>1. The use of PGA in the definition of treatment target exposes a substantial proportion of patients to the risk of overtreatment with immunosuppressive agents, while being deprived of the adjunctive therapy they probably need.

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P0095 – OSTEOPOROSE INDUZIDA PELA QUIMIOTERAPIA DA NEOPLASIA DA MAMA: UM REFRESCAR DA MEMÓRIA

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Introdução: A menopausa prematura induzida pela quimioterapia da neoplasia da mama, que aumenta o risco de desenvolver osteoporose, parece ser um factor subestimado no momento da avaliação desta patologia. Como tal, os clínicos deverão estar sensibilizados para a importância da prevenção, bem como o momento do início de terapêutica específica para a osteoporose.

Objectivo: Para isso, avaliamos e categorizamos a existência de osteoporose / osteopenia em mulheres após realização de terapêutica hormonal para a neoplasia da mama.

Material e Métodos: Trata-se de um estudo observacional retrospectivo de 11 doentes referenciadas à consulta de Reumatologia da Unidade Local de Saúde da Guarda por queixas musculoesqueléticas, após quimioterapia para a neoplasia de mama. Foram explorados outros factores de risco para osteoporose,

tendo sido também realizada uma densitometria óssea (DMO). As características clínicas foram colhidas através do seu processo clínico.

Resultados: A idade média foi de 64,0 ± 12,8 anos e a idade média de diagnóstico de neoplasia da mama foi de 53,7 ± 10,8. O fármaco mais comum foi o anastrazol (54,5%), seguido do tamoxifeno (45,6%), goserelina (18,2%) e letrozol (9,1%). 4 doentes (36,4%) sofreram uma fractura de baixo impacto (vertebral, femoral, tibiotársica e costela), das quais 2 sofreram mais do que uma fractura. A mediana do risco a 10 anos de fractura (FRAX português) foi de 5,4% para fractura major e 1,7% para fractura da anca. 4 doentes (36,4%) apresentaram DMO compatível com osteoporose, 5 (45,5%) apresentaram osteopenia e 2 (18,2%) apresentaram densidade óssea normal. Das 4 doentes com osteoporose na DMO, a ferramenta de avaliação do risco de fratura foi <11% para fractura major e/ou <3% para fractura da anca em 3 (75%) mulheres.

Conclusão: Embora o FRAX se tenha vindo a mostrar como uma ferramenta inestimável para a população em geral, parece ser insuficiente na avaliação do risco de fractura em mulheres com osteoporose induzida pela quimioterapia para neoplasia da mama. Quando este importante factor de risco se encontra presente, outros elementos devem ser tidos em consideração para o início da terapêutica para a osteoporose, não devendo o FRAX ser usado isoladamente na mesma.

P0096 – AUMENTO DA TAXA DE VACINAÇÃO EM DOENTES SOB DMARDS BIOLÓGICOS APÓS CONSULTA DE AVALIAÇÃO DO RISCO INFECCIOSO (CARI)

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Introdução: As intercorrências infecciosas são uma grande preocupação nos doentes imunodeprimidos. Em 2019, na tentativa de melhorar este aspecto, a EULAR lançou uma versão actualizada das suas recomendações para a vacinação de doentes com doenças reumáticas. Uma vez que estes doentes são deliberadamente imunodeprimidos para que atinjam a remissão, a vacinação torna-se pertinente como forma de prevenção da infecção. Em 2018, foi criada no nosso centro uma Consulta de Avaliação do Risco Infeccioso (CARI), de forma a regularizar o calendário de vacinas e preparar os doentes para terapêutica biológica. Este trabalho resume a nossa experiência.

TABELA 1 – PRESCRIÇÃO DE VACINAS EM AMBOS OS GRUPOS (P<0.05)

	Ida à CARI	Sem ida à CARI	p
Pneumocócica conjugada 13-valente			0.24
Prescrita	23 (95.83%)	34 (82.93%)	
Não prescrita	1	7	
Pneumocócica polivalente			0.01
Prescrita	19 (79.17%)	19 (46.34%)	
Não prescrita	5	22	
Vírus da Gripe			<0.01
Prescribed	22 (91.67%)	12 (29.27%)	
Not Prescribed	2	29	
Hepatite A			0.05
Prescrita	3 (12.50%)	0 (0.00%)	
Não prescrita	21	41	
Hepatite B			<0.01
Prescrita	12 (50.00%)	0 (0.00%)	
Não prescrita	12	41	
Herpes Zoster			0.37
Prescrita	1 (4.17%)	0 (0.00%)	
Não prescrita	23	41	
Tétano/Difteria			0.14
Prescrita	3 (12.50%)	1 (2.44%)	
Não prescrita	21	40	

Objetivos: Comparar a taxa de vacinação entre doentes com doenças reumáticas sob terapêutica biológica antes e após a criação da consulta.

Materiais e Métodos: Foi realizado um estudo observacional retrospectivo no Serviço de Reumatologia e CARI, na Unidade Local de Saúde da Guarda. Todos os doentes sob terapêutica biológica seguidos no serviço entre 2010 e 2020 foram incluídos no estudo. As seguintes características sociodemográficas e clínicas foram colhidas: sexo, idade, diagnóstico de doença reumática, terapêutica biológica actual e histórico de vacinação. As vacinas estudadas foram a Pneumocócica conjugada 13-valente, Pneumocócica polivalente, Vírus da Gripe, Vírus da Hepatite A e B, Herpes Zoster e Tétano / Difteria. O doente foi considerado vacinado quando o timing correcto foi respeitado pelo médico assistente. Se a prescrição estivesse fora do prazo estipulado, este seria considerado não vacinado. Na análise estatística foram utilizados o teste de qui-quadrado e exato de Fisher para avaliar associações e foram consideradas como tendo significância estatística se p <0,05.

Resultados: Foram incluídos 65 doentes, dos quais 41 (63,10%) eram do sexo feminino, com uma idade média de 52,38 ± 11,11 anos. A distribuição diagnóstica foi a seguinte: artrite reumatóide (52,30%), espondilartrite axial (26,20%), artrite psoriática (12,30%), espondilartrite periférica (6,20%), 1 (1,50%) doente com Síndrome de Sjögren e 1 (1,50%) com esclerose sistémica. Os fármacos mais comuns foram anti-TNFα (64,60%), anti-JAK (15,40%), anti-IL17A (10,80%), anti-CD20 (7,70%) e anti-IL12 / IL23 (1,50%). Do total, 24 (36,90%) doentes foram observados na CARI antes de iniciar a terapêutica biológica. A tabela 1 mostra as vacinas prescritas nos dois grupos e suas associações.

Conclusão: Algumas conclusões podem ser tiradas

deste trabalho. Em primeiro lugar, a prevalência da vacinação aumentou após a criação da consulta, especialmente a da vacina Pneumocócica polivalente, Vírus da Gripe e Vírus da Hepatite A e B. Esperávamos os mesmos resultados nas restantes vacinas numa população mais ampla. Os resultados também foram afectados pela ruptura de stock de algumas vacinas. Em segundo lugar, este estudo mostra a importância de um protocolo estabelecido, que ajuda a sistematizar a avaliação do risco infeccioso antes da terapêutica biológica, analisando minuciosamente o histórico de vacinação e mantendo-o actualizado. Por fim, a responsabilidade compartilhada entre reumatologistas e infectiologistas permite-lhes tirar partido das suas competências, com ganhos últimos para o doente. Esperamos que este trabalho motive outros colegas a iniciar práticas semelhantes nos seus centros.

P0100 – ENTHESITIS AND CLINICAL RESPONSE IN PSORIATIC ARTHRITIS: REAL-LIFE DATA

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Background: Psoriatic arthritis (PsA) is an inflammatory arthritis that is characterized by a broad spectrum of clinical conditions, including axial skeletal involvement, enthesitis, dactylitis, uveitis and arthritis. Among those, enthesitis, the inflammation of the junction where the tendon, ligament or joint capsule inserts into the bone, is assigned to be the hallmark, affecting 35–50% of patients. Several clinical methods have been developed to measure it, including The Maastricht AS Enthesitis Score (MASES) index, which tests 13 entheses and the Spondyloarthritis Research Consortium of Canada (SPARCC) index that assesses 16.
Objective: To assess the relationship between enthesitis and clinical response in psoriatic arthritis.
Methods: Retrospective study including all the patients with PsA meeting the CASPAR criteria, beginning first-line biologic therapy at our centre. Demographic and clinical data including age, gender, body mass index (BMI), smoking status, physical examination findings such as presence of enthesitis, dactylitis, chronic back pain, tender and swollen joint counts (TJC/ SJC), ESR, CRP, DAS 28 4vESR , BASDAI, BASFI, BASMI, ASDAS, HAQ, patient VAS score , MASES and SPARCC were collected from the Portuguese database Reumapt. Statistical analysis was performed with SPSS. Continuous variables were analysed through Spearman correlations.
Results: We included 119 patients with PsA (60 female), of which 14.9% were active smokers. The mean age of patients was 46.3 ± 1.03 years. The median disease duration was 6.8 (0.3-33.8) years and the mean BMI was 26.8 ± 0.5 Kg/m2. Enthesitis, dactylitis, inflammatory back pain, peripheral arthritis, ungueal distrophy, and psoriasis were present in 53 (45.7%), 45 (38.8%), 76 (65.5%), 109 (94%), 45 (38.8%), 104 (89.7%) patients, respectively. At baseline, mean (SD) disease activity parameters were: DAS 28 4vESR 4.9 (0.2), ESR 33.2 (2.3) mm/h; CRP 2.35 (0.3) mg/dL, HAQ 1.3 (0.1), BASDAI 6.6 (0.2), ASDAS 3.9 (0.1), BASMI 3.7 (0.2), BASFI 5.8 (0.3), MASES 1.9 (0.3), SPARCC 2.3 (0.3). Median (min-max) values of TJC, SJC and patient VAS score at baseline were 4 (0-28), 3 (0-19), 76 (0-100), respectively. There were statistically significant positive correlations (0-12 months) between ΔMASES and ΔDAS 28 4vESR (p=0.02, rho=0.432), Δpatient VAS score (p=0.027, rho=0.307), ΔHAQ (p=0.02, rho=0.411), ΔBASDAI (p=0.025, rho=0.326), ΔBASFI (p=0.037, rho=0.315), ΔASDAS (p=0.023, rho= 0.331). Correlations between ΔSPARCC and ΔDAS 28 4vESR (p=0.023, rho=0.332), Δpatient VAS score (p=0.003, rho=0.402), ΔHAQ (p=0.012, rho=0.440), ΔBASDAI (p=0.011, rho=0.368), ΔBASFI (p=0.001, rho=0.445), ΔASDAS (p=0.002, rho= 0.437), ΔCDAI (p=0.039, rho=0.320) and ΔSDAI (p=0.039, rho=0.319), were also significant. However, there weren't strong correlations between ΔMASES neither ΔSPARCC and PsARC response at 12 months.
Conclusion: Our results suggest that enthesitis is correlated with clinical response in PsA, supporting the idea that it is a major determinant of disease activity. It should be given more importance, namely by incorporating it in daily clinical practice, due to its major role, both in establishing an early diagnosis and in assessing treatment response.

P0101 – THE IMPACT OF BODY MASS INDEX ON DISEASE ACTIVITY AND ENTHESITIS IN PSORIATIC ARTHRITIS

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Background: Overweight/obesity has increased exponentially in the last decades, becoming a huge Public Health problem. Moreover, an increase in adipose tissue is associated with an increased production of several proinflammatory cytokines and acute phase

reactants. Higher BMI has been related with new bone formation including syndesmophytes and enthesophytes. In fact, besides rheumatologic conditions including Psoriatic Arthritis (PsA), enthesopathy can be a consequence of several clinical conditions including metabolic syndrome, mechanical injuries and degeneration.

Objectives: To evaluate the effect of body mass index (BMI) on disease activity scores and enthesitis scores in Psoriatic Arthritis

Methods: Retrospective study including all the patients with PsA meeting the CASPAR criteria, beginning first-line biologic therapy at our centre. Demographic and clinical data were collected from the Portuguese database Reumapt. Statistical analysis was performed with SPSS. Continuous variables were compared through Spearman/Pearson correlations.

Results: The mean BMI was 26.8 (SD 0.5). In our sample of 119 PsA patients, 21.5% were overweight and 8.3% were obese. The mean age of patients was 46.3 ± 1.03 years; 60 female and 59 male. The median disease duration was 6.8 (0.3-33.8) years. At baseline mean (SD) disease activity variables were: DAS 28 4vESR 4.9 (0.2), ESR 33.2 (2.3) mm/h; CRP 2.35 (0.3) mg/dL, BASDAI 6.6 (0.2), ASDAS 3.9 (0.1), BASMI 3.7 (0.2), BASFI 5.8 (0.3), MASES 1.9 (0.3), SPARCC 2.3 (0.3). There were statistically significant positive correlations between BMI and MASES at baseline (p=0.024, r=0.411) but there weren't with SPARCC, DAS 28 4vESR, ESR, CRP, BASDAI, ASDAS, BASMI and BASFI.

Conclusion: The data showed that patients with higher BMI values had higher enthesitis scores suggesting that overweight/obesity may have a negative impact on enthesopathy. Further studies are still needed to further understand that possible relationship.

P0104 – SERUM ALBUMIN LEVELS AND DEPRESSION IN JSLE

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Background: Albumin is a negative acute phase response protein synthesized in the liver, being an important marker of inflammation. Under inflammatory conditions, the transcapillary escape rate of albumin may increase, leading to hypoalbuminaemia. Systemic lupus erythematosus (SLE) is a chronic condition involving multiple organ systems, inducing functional disability and psychological burden responsible for noteworthy depressive symptoms. Depression may be related with psychosocial, environmental and biological factors, disease activity and its severity, age and sex. Several studies show that immune activation and increased concentrations of positive and decreased concentrations of negative acute phase proteins are involved in the pathogenesis of depression. As albumin has the capacity to bind homocysteine, lowered serum albumin levels leads to hyperhomocysteinemia, a well-known risk factor for depression. Moreover, hypoalbuminaemia decrease the availability of tryptophan, an essential amino acid from which the neurotransmitter serotonin is derived, and induce oxidative stress, which further decreases antioxidant levels in people with depression.

Objective: To assess the association between serum albumin levels and depressive symptoms in juvenile -onset SLE (jSLE) patients.

Methods: A cross-sectional sample of jSLE patients, currently aged ≥ 16 years, completed a psychosocial assessment including quality of life (SF-36) anxiety and depressive symptoms (HADS) and cognitive assessment (MMSE), between October 2018- May 2019. Local Ethics Committee approved the study. All patients fulfilled both 2012 and 2019 EULAR/ACR classification criteria for SLE. Juvenile-onset was defined as age at diagnosis <18 years. Demographics and clinical characteristics were collected. Statistical analysis was performed with SPSS®. Variables were compared with spearman correlations tests.

Results: 30 jSLE patients were included (90%female) in the study, with median (min-max) age of 21 (16-35) years, with mean (SD) age of diagnosis of 15.8 ± 2.1. Median albumin serum level was 41.7 (16.7-46.3) g/dL. Psychosocial assessment revealed a mean (SD) score in HADS – Depression of 3.9 (3.3), HADS – Anxiety of 9 (4.3), MMSE of 27.7 (1.8), Physical health SF-36 of 66.8 (9.9) and Mental health SF-36 of 68.9 (17.5). 23.3 % jSLE showed mild cognitive impairment, 63.3% anxiety and 13.3% depression. We observed significant inverse linear relationships between serum albumin levels and depressive symptoms score (p=0.042, ρ=-0.380) and with anxiety symptoms score ((p=0.029, ρ=-0.406). No significant correlations were detected between albumin serum concentrations and cognitive assessment.

Conclusions: Our findings are consistent with studies previously reporting the potentially protective effect of high serum albumin levels on mental health

in different populations. A possible inflammation related aetiology for depression in jSLE patients is highlighted, further explained through the protective roles played by albumin in inflammation, infection, and oxidative damage.

P0106 – A EQUIPA DE ENFERMAGEM NO TRATAMENTO DOS DOENTES COM DOENÇA REUMÁTICA INFLAMATÓRIA SISTÉMICA – PROJETO DE UMA CONSULTA DE ENFERMAGEM

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As doenças reumáticas inflamatórias sistémicas têm um grande impacto na qualidade de vida dos doentes e suas famílias (Branco et al, 2016), assumindo-se como um problema com grande relevância na sociedade moderna, pois tendem a ser progressivas e a afetar, cada vez mais, diferentes dimensões da pessoa, nomeadamente física, social, espiritual e emocional.
Um cuidado multidisciplinar efetivo, alicerçado numa gestão global e integrando os diferentes saberes e as diferentes áreas de intervenção, constitui a chave do tratamento eficaz destes doentes, nomeadamente prolongando os períodos de remissão, evitando agudizações, prevenindo complicações e promovendo a qualidade de vida (Smith et al, 2018).
O enfermeiro, inserido na equipa multidisciplinar, assume um papel fundamental no apoio e cuidado ao doente, promovendo o empowerment na autogestão da doença, a educação em saúde do doente e sua família, constituindo um elo de ligação e um elemento de referência dentro da equipa, para a gestão global dos cuidados prestados (J. Hill, 2007).
A EULAR recomenda que os enfermeiros que prestam cuidados ao doente com patologia reumática inflamatória sistémica desenvolvam o seu trabalho no sentido de participarem na gestão da doença, ajudando a controlar a sua atividade, a reduzir os sintomas e a melhorar os resultados nas áreas preferenciais definidas pelo doente; identificarem, avaliarem e abordarem as questões psicossociais para minimizar o aparecimento de ansiedade e depressão nos doentes; e promoverem a capacidade de autogestão da doença, com a finalidade do doente obter uma maior sensação de conforto, auto-eficácia e capacitação (van Eijk-Hustings et al, 2019).
Neste contexto, entendeu-se que seria importante implementar uma consulta de enfermagem no Hospital Garcia de Orta, para o acompanhamento do doente com doença reumática inflamatória sistémica. Pretende-se que esta consulta tenha uma estrutura capaz de dar resposta às necessidades identificadas e que vise aumentar a efetividade do atendimento e otimizar o tratamento destes doentes.
Nesta consulta de enfermagem, cujo projeto pretendemos apresentar, ambiciona-se desenvolver um cuidado empático, promovendo uma relação de confiança e fornecendo ao doente a informação necessária à aceitação e adaptação à doença, nomeadamente relativa a sintomas, complicações, tratamentos e efeitos secundários, procurando aproximar os nossos cuidados das melhores práticas internacionais.

P0123 – WRIST INTERSECTION SYNDROME – THE ROLE OF ULTRASOUND ON A LESS DESCRIBED CAUSE OF WRIST PAIN

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Background: In our Rheumatology department, patients with regional pain syndromes are evaluated in a differentiated consultation with the possibility to use "on-time" musculoskeletal ultrasound. Wrist pain is a frequent reason for consultation. Although De Quervain's tenosynovitis seems the most common cause, other entities may be the cause of such pain. Intersection wrist syndrome is a less frequent cause, poorly described and reported in literature. It can occur proximally, where the first extensor compartment (abductor pollicis longus and extensor pollicis brevis) cross the second compartment (extensor carpi radialis longus and brevis), or distally, where the third compartment (extensor pollicis longus) cross the second compartment. When the cause of pain is not clinically evident, the use musculoskeletal ultrasound can contribute to a correct differential diagnosis with other conditions.
Objectives: To review the cases of intersection syndrome after ultrasound scanning among patients evaluated for wrist pain, describe epidemiological and clinical data and report outcomes from different therapeutic approaches.
Methods: Revision of all clinical appointments for wrist pain during 2019. Description of all cases of

wrist intersection syndrome, including patients' sociodemographic characteristics, occupation, symptoms, ultrasound findings, therapeutic measures and outcomes.

Results: Wrist pain was the cause for evaluation in 38 patients (38/684 – 5.6%) in this specific consultation, four of them reporting bilateral pain. De Quervain's tenosynovitis was the most common diagnosis (n=17; 44.7%), followed by wrist intersection syndrome (n=8; 21.1%) and inflammatory diseases (n=7; 18.4%). Proximal and distal intersection syndromes occurred in 4 patients each. All patients were females aged between 31 and 69 years old. Six of them had occupations that required repeated movement of the forearm and wrist. Two of the patients had already been submitted to surgeries for persistent wrist pain prior to Rheumatology evaluation, both De Quervain's tenosynovitis diagnosis and with no subsequent improvement. Musculoskeletal ultrasound allowed an accurate diagnosis. Most patients had effusions within involved tendon sheaths at the intersection site or in close proximity. In most patients, non-steroidal anti-inflammatory drugs (NSAIDs) and rest were unsuccessful measures and local steroid injection was performed in 5 patients with only transient improvement. The patients were then referred for surgery.

Conclusions: Wrist intersection syndrome is a less common cause of wrist pain, potentially being overlooked. Proximal and distal syndromes occurred in the same number of patients, although proximal syndrome seems more common, according to available reports. Musculoskeletal ultrasound is useful to correctly locate the origin of pain, allowing an accurate differential diagnosis and may also be useful for guidance of steroid injection. In our experience, conservative measures like NSAIDs and rest were unsuccessful and local steroid injection only allowed transient improvement.

P0128 – RISK FACTORS FOR ADVERSE PREGNANCY OUTCOMES IN SPONDYLOARTHRITIS: MATERNAL DISEASE PHENOTYPE AND DISEASE ACTIVITY MAY PLAY A ROLE

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TABLE 1 – DIFFERENCES BETWEEN PATIENTS WITH AND WITHOUT APO

	Pregnancies with APO	Pregnancies without APO	*p*
N, %	23, 41.1	33, 58.9	
Maternal age at conception, Median (IQR) (years)	33.0 (31.5-36.5)	33.0 (31.0-37.0)	0.9
Hx previous APO – N, %	4,17.4	3, 9.1	0.3
Disease duration, median (IQR) (months)	96 (36-132)	48 (24-96)	0.1
Axial dominant disease – N, %	6, 26.1	6, 18.2	0.5
Peripheral dominant disease – N, %	11, 47.8	20, 60.6	0.4
Hx enthesitis – N, %	9, 39.1	13, 39.4	1.0
Hx dactylitis– N, %	5, 21.7	11, 33.3	0.4
Hx psoriasis– N, %	10, 43.5	18, 54.5	0.6
Hx uveitis– N, %	1, 4.3	4, 12.1	0.6
Hx inflammatory bowel symptoms – N, %	4, 17.4	0, 0	**0.02**
HLA-B27– N, %	8, 34.8	7, 21.2	0.5
>1 cs or bDMARDs before conception – N, %	13, 56.5	10, 30.3	**0.05**
LDA during pregnancy – N, %	7, 30.4	17, 51.5	0.2
Active disease before pregnancy - N, %	5, 21.7	2, 6.1	0.1
Active disease 1st trimester - N, %	1, 4.3	4, 12.1	1.0
Active disease 2nd trimester - N, %	6, 26.1	5, 15.2	0.2
Active disease 3rd trimester - N, %	3, 13	0, 0	**0.03**

APO – adverse pregnancy outcomes; bDMARD – biological DMARD; cs –conventional synthetic; DMARD – disease-modifying antirheumatic drug; Hx – history of; LDA – low dose aspirin.

Background: Pregnant patients (pts) with spondyloarthritis (SpA) seem at increased risk for adverse pregnancy outcomes (APO), however limited and conflicting data have been published so far and risk factors for APO in these pts remain poorly understood.

Objectives: To assess APO and identify possible risk factors for those in a cohort of SpA pregnant pts.

Methods: Data on SpA pts prospectively-followed in a pregnancy clinic from 2010 to 2019 were retrospectively analysed before conception and during each trimester. Pregnancies complicated by APO were compared with those that were uneventful for demographic and clinical variables using Chi-square, Fischer's Exact Test and Mann-Whitney Test, as appropriate. Active disease was defined as a DAS-28-CRP>3.2 or an ASDAS-CRP ≥ 2.1 according to peripheral or axial dominant disease respectively.

Results: 56 pregnancies (median age 33 years, IQR 31-37; median disease duration 60 months, IQR 24-123) in 47 pts were analysed: 37 psoriatic arthritis, 7 axial SpA, 6 undifferentiated SpA, 3 enteropathic SpA, 2 reactive arthritis and 1 enthesitis-related juvenile idiopathic arthritis. APO were recorded in 23/56 (41%) pregnancies: 5 (9%) early miscarriages, 1 (2%) medical termination (central nervous system malformation), 3 (5%) preterm births (≥34 gestational week, all for preterm premature rupture of membranes – PROM); 2 (4%) PROM; 7 (13%) small for gestational

age newborns (SGA); 3 gestational diabetes and 2 cholestasis of pregnancy. Table 1 displays the comparison between pregnancies with and without APO. A higher number of pts with active disease were detected during the 2nd trimester in both groups, however differences between those were only significant at the 3rd trimester (p=0.03). History of inflammatory bowel symptoms (IBS) was also associated with an increased risk for APO (p=0.02). Although not reaching statistical significance, APO occurred more frequently in pts with a previous use of > 1 conventional synthetic or biological disease-modifying antirheumatic drugs (p=0.05), suggesting a more difficult to treat phenotype. Likewise, pts with APO were less often treated with low dose aspirin (LDA) during pregnancy.

Conclusions: SGA was the main APO recorded. History of IBS, a more difficult to treat phenotype and the presence of active disease during pregnancy influenced APO in this cohort, highlighting the need for tight disease control before and during pregnancy. Larger and prospective data are warranted to confirm these results and to assess the potential protective role of LDA.

P0130 – KNOWLEDGE AND AWARNESS OF PORTUGUESE HIGH SCHOOL STUDENTS: EFFECT OF A SINGLE SESSION PROVIDED BY NURSES AND PATIENT REPRESENTATIVES

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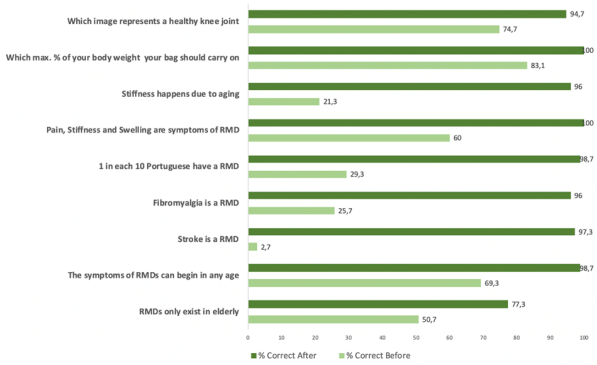
Background: International studies have shown that the awareness and knowledge of the general population about rheumatic and musculoskeletal diseases (RMDs) is poor. This is even lower in the young population, which are also affected by these disease but do not have as much awareness campaigns as adult community. Their professors and primary health care professionals may also play here also a key role, promoting early detection of signs and interpretations of symptoms, thus avoiding late health care referrals and diagnosis. (Vlieland, 2016).

Objective: To assess the knowledge of high school Portuguese students about the RMDs and raise awareness for RMDs in young people, their professors, vigilants, and primary care nurses, within the school environment.

Methods: A 1-hour educational session about RMDs was planned (with the inputs from members of the Portuguese EULAR Associations) and performed during school activities. The educational session started with a knowledge questionnaire about RMDs in a paper sheet (9 questions; Graph 1), repeated in the end. An interactive session, using slides, interactive questions (Sli.do®), and practical demonstrations to simulate RMD symptoms (e.g. stiffness and functional limitations) was then lead by a rheumatology nurse, with the testimony from a young patient representative. A primary care nurse assisted in order to be engaged and promote future sessions ("autonomously"). Change in knowledge was assessed with Wilcoxon-test and awareness was documented with "word clouds" (using Sli.do®).

Results: A total of 75 students participated in four sessions (mode=16 years). Half of students (52%) had

GRAPH 1 – CHANGE IN THE PERCEBTAGE OF CORRECT ANSWERS BEFORE AND AFTER AN EDUCATIONAL SESSION (N=75) (ALL P<0.001)



THE MOST COMMON WORDS REPRESENTING WHAT ARE THE MAIN SYMPTOMS A RMD

never heard about RMDs. Knowledge increased significantly in all questions (p<0.001; Graph 1). Figure 1 document the most common words representing what are the main symptoms a RMD.

Conclusion: Our results confirm that awareness and knowledge about RMDs are very low high school students. The single and educational session was very well received by all students, and the the knowledge increased. Post-educational feedback was that students especially liked the testimony of a peer. Other sessions are taking place in primary schools.

References:

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P0131 – RISK FACTORS FOR CRANIAL ISCHAEMIC EVENTS IN GIANT CELL ARTERITIS – A BICENTRIC POPULATION-BASED STUDY

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Background: Giant cell arteritis (GCA) is the most common primary vasculitis in patients (pts) aged over 50 years. It is considered a medical emergency due to the potential occurrence of cranial ischaemic events (CIEs), namely vision loss and cerebrovascular accidents (CVAs), largely responsible for the high burden of this disease. Identifying pts at a higher risk of developing CIEs is clinically relevant to determine prognosis and tailor treatment.

Objectives: To determine the risk factors for GCA-related CIEs in a cohort of pts with GCA managed at two tertiary Portuguese centres.

Methods: Bicentric observational retrospective study using data from the Rheumatic Diseases Portuguese Registry (Reuma.pt), complemented with information retrieved from hospital clinical records. Pts with biopsy-proven GCA or with the presence of "halo sign" on ultrasound of the temporal ± axillary arteries were included. Visual disturbances and CVAs (stroke or transient ischaemic attacks) were considered CIEs of interest. Clinical features of pts with or without CIEs were compared using Chi-square and Mann-Whitney U tests, as appropriate; multivariate logistic regression was used to identify predictors of CIEs.

Results: A total of 134 pts diagnosed with GCA between 1998 and 2019 were included. Median (IQR) age at diagnosis was 76.4 (70.2-82.4) years and 66% of pts were females. At least one CIE was reported in 65 (48.5%) pts; 8 pts had more than one CIE. A total of 73 CIEs were recorded: 44 cases of ischaemic optic neuropathy (29 anterior, 2 posterior, 13 not specified), 61% with unilateral involvement and 74% leading to permanent visual loss (PVL); 6 cases of central retinal artery occlusion with 100% PVL; 16 ocular events of non-specified cause (6 transient visual loss, 5 PVL, 5 diplopia); 6 ischeamic strokes; and 1 transient ischaemic attack. Table 1 presents the differences between

TABLE 1 – DIFFERENCES BETWEEN GCA PATIENTS WITH AND WITHOUT CRANIAL ISCHAEMIC EVENTS

	CIEs (N=65)	Non-CIEs (N=69)	p value
Age at diagnosis, years	79.2 (73.7-84.4)	74.2 (67.6-78.4)	**<0.001**
Time to diagnosis since first symptoms, days	37 (6-97)	36 (10-120)	0.65
Female sex, n/mN (%)	39/65 (60)	50/69 (72)	0.13
Comorbidities, n/mN (%)			
Hypertension	51/65 (78)	38/69 (55)	**0.005**
Dyslipidaemia	27/65 (42)	18/69 (26)	0.09
Diabetes mellitus	19/65 (29)	13/69 (19)	0.2
Hyperuricemia	6/62 (10)	7/61 (11)	0.75
Ischaemic heart disease	7/62 (11)	7/61 (11)	0.97
Cerebrovascular disease	8/62 (13)	6/61 (10)	0.59
Atrial fibrillation	10/62 (16)	5/61 (8)	0.18
Chronic kidney disease	10/62 (16)	8/61 (13)	0.64
Previous or current smoker	10/50 (20)	10/57 (18)	0.75
Baseline blood tests			
Erythrocyte sedimentation rate, mm/h	84 (65-109)	80 (61-103)	0.48
C-reactive protein, mg/dL	4.8 (3.0-8.6)	4.9 (2.6-9.4)	0.93
Leukocytes, x10^{9}cels	8.780 (6.810-10.900)	10.070 (7.600-11.600)	0.28
Haemoglobin, g/dL	11.5 (10.4-12.5)	11.9 (10.5-12.9)	0.38
Creatinine, mg/dL	0.9 (0.7-1.1)	0.8 (0.6-1.0)	**0.01**
Clinical features, n/mN (%)			
Constitutional symptoms *	30/65 (46)	38/69 (55)	0.26
Musculoskeletal manifestations	26/64 (41)	45/69 (65)	**0.005**
Arthralgia/myalgia/arthritis	24/64 (38)	39/69 (57)	**0.03**
PMR	21/64 (33)	29/69 (42)	0.27
New-onset headache	54/65 (83)	58/69 (84)	0.7
Jaw claudication	31/64 (48)	16/69 (23)	**0.002**
TA abnormalities on physical examination**	42/52 (81)	26/54 (48)	**0.001**
Medication before diagnosis, n/mN (%)			
Antiplatelet therapy	12/56 (21)	8/53 (15)	0.39
Statins or fibrates	12/54 (22)	3/39 (8)	0.06
Antihypertensive drugs	17/46 (37)	11/47 (23)	0.15

CIEs: cranial ischaemic events; GCA: Giant cell arteritis; PMR: polymyalgia rheumatica; TA: temporal artery.
Univariate analysis using Mann-Whitney U test for continuous variables and Chi-square for categorical variables. Continuous variables are expressed as median (IQR). Categorical variables are expressed as n/modified(m)N (total N – missing data), %.

* Fatigue, anorexia, weight-loss, fever, night sweats or lymphadenopathies.
** Thickness, tenderness, and reduced or absent pulse.

FIGURE 1 – MULTIVARIATE LOGISTIC REGRESSION MODEL FOR PREDICTION OF CRANIAL ISCHAEMIC EVENTS

	B	S.E.	Wald	df	p	OR	95% C.I. for OR	
							Inferior	Superior
Age at diagnosis	0,061	0,032	3,636	1	0,057	1,063	0,998	1,132
High blood pressure	1,353	0,562	5,788	1	0,016	3,869	1,285	11,652
Jaw claudication	1,251	0,609	4,226	1	0,040	3,496	1,060	11,527
Abnormalities on temporal artery palpation	1,465	0,606	5,842	1	0,016	4,328	1,319	14,199
Musculoskeletal features	-1,212	0,549	4,863	1	0,027	0,298	0,101	0,874
Baseline [creatinine]	1,082	0,696	1,459	1	0,227	2,950	0,510	17,072
Constant	-7,118	2,545	7,826	1	0,005	0,001		

B – coefficients in log-odds units; CI – confidence interval; df – degrees of freedom; OR – odds ratio; p – Wald 2-tailed p-value; SE – standard errors associated with the coefficients; Wald – Wald chi-squase value.

pts with and without CIEs. On univariate analyses, pts with CIEs were older (p<0.001), and more often had previous hypertension (OR: 3.0, 95%CI: 1.4-6.3), jaw claudication (OR 3.0, 95%CI: 1.5-6.5) and temporal artery (TA) abnormalities on examination (OR 4.5, 95%CI: 1.8-10). By contrast, musculoskeletal (MSK) features including polymyalgia rheumatica (PMR) were negatively associated with the occurrence of CIEs (OR 0.37, 95%CI: 0.2-0.7). In terms of laboratory results, only creatinine levels at diagnosis were significantly higher (p=0.014) in pts with CIEs; no differences were seen between groups for baseline inflammatory markers or haemoglobin levels. Likewise, antiplatelet therapy started before GCA diagnosis did not seem to prevent CIEs. By multivariate logistic regression (Figure 1), hypertension, jaw claudication and abnormalities on TA palpation were independently associated with CIEs, as opposed to age at diagnosis and baseline creatinine levels. The presence of MSK symptoms significantly decreased the risk of CIEs.

Conclusion: Almost half the pts in this cohort presented CIEs, highlighting the need for an earlier recognition of symptoms and a fast-track approach to pts with suspected GCA to improve outcome. High blood pressure, jaw claudication and abnormalities on TA palpation were associated with an increased risk for the occurrence of CIEs, while the presence of MSK symptoms including PMR had a protective role. The identification of pts with these high-risk features should prompt immediate treatment and tight follow-up. Future studies are warranted to determine the value of more aggressive immunosuppression in these cases.

P0132 – SACROILIAC JOINT INJECTIONS PERFORMED WITH ULTRASOUND GUIDANCE IN PATIENTS WITH SACROILIITIS

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Background: Sacroiliitis is the hallmark of axial Spondyloarthritis (axSpA). ASAS-EULAR management recommendations for axSpA, consider glucocorticoid injections directed to the local site of musculoskeletal inflammation as a treatment option for pain relief, besides treatment with oral nonsteroidal anti-inflammatory (NSAIDs) before starter biotechnological treatment. However, there are few studies to evaluate the efficacy of this technique with a small number of patients and a short follow-up. Ultrasonography has been used as a valuable option to guide this technique.

Objectives: To evaluate the efficacy and safety of ultrasound-guided injections of sacroiliac joints (SIJs) in patients with sacroiliitis using clinical and laboratory outcomes at baseline and 4-6th weeks.

Methods: This study involved patients with axSpA with acute sacroiliitis, ≥18 and ≤ 65 years old, with body mass index (BMI) < 30kg/m2 attending the Rheumatology Outpatient Clinic, which had been poorly controlled (ASDAS>2.1) by conventional therapy (physiotherapy, NSAIDs at the maximum tolerated dosing during ≥ 4 weeks). Sociodemographic, clinical (disease duration, BMI, BASDAI, BASFI, ASDAS) and laboratory (CRP) data were collected from the medical records at baseline and at 4-6th weeks. Statistical analyses were conducted using SPSS version 25. Continuous variables were described with mean/median ± standard deviation (SD).

SIJs injection was performed, under ultrasound guidance, using standard procedures with 2mL of lidocaine 1% and 40mg of methylprednisolone, with a 22-gauge needle. The procedure was performed by the same operator. Written informed consent was obtained from all patients.

Results: We performed eleven sacroiliac injection in eleven consecutive patients (one procedure per patient). Nine patients (81.8%) were female, mean age (±SD) of 40.6(±9.4) years, median disease duration(±SD) of 0.9(±6.2) years and median BMI(±SD) of 24.2(±3.3). Eight patients (72.7%) had Nr-axSpA. All patients were non-responders to NSAIDs. At 4-6th weeks there was a decrease in median (±SD) BASDAI (5.4±1.9 vs 4.1±1.9), BASFI (4.2±1.4 vs 3.5±2.3) and ASDAS (3.2±0.8 vs 2.2±0.6) indexes.

Conclusion: As previous studies demonstrated, this technique seems to be safe and quite effective. Our goal is to increase the number of patients undergoing this technique and have longer follow up to evaluate its efficacy. The study has several limitations: the mid- and long-term effects should be evaluated in the future based on the results of the short-term effects and the study was not conducted as a double-blinded, controlled study.

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P0138 – MUSCLE PHYSICAL PROPERTIES AND SEGMENTAL MUSCLE STRENGTH IN RADIOGRAPHIC AND NON-RADIOGRAPHIC AXIAL SPONDYLARTHRITIS PATIENTS: ANALYSIS OF THE MYOSPA STUDY

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Background: Axial Spondylarthritis (axSpA) is usually divided into radiographic (r-axSpA) and non-radiographic (nr-axSpA). Recently, increased resting lumbar myofascial stiffness was described in r-axSpA (Ankylosing Spondylitis) patients. However, there are no data comparing muscle physical properties (MPP) and strength between r-axSpA and nr-axSpA.
Objective: To investigate whether MPP and segmental muscle strength are different between patients with r-axSpA and nr-axSpA, having healthy subjects as controls.
Methods: Patients with axSpA (according to the ASAS classification criteria), aged 18 to 50 years, with symptoms duration ≤10 years were included in this cross-sectional study. Healthy individuals, matched by gender, age and levels of physical activity (1:1) were used as control group (HC).
MPP [stiffness, tonus, decrement (inverse of elasticity)] and strength of three different body segments (trunk, upper and lower limbs, on both sides) were measured by Myoton Pro® and resisted hand-held dynamometer, respectively, by a single reader. Physical activity was assessed by the International Physical Activity Questionnaire (IPAQ). Mann-Whitney U test, Fisher's exact test or chi-square test were used to compare differences between groups (axSpA vs HC; r-axSpA vs nr-axSpA).
Results: A total of 27 axSpA patients and 27 HC were included. Overall, mean age was 36.5 ± 7.5 years, 67% were males. There was no significant difference between patients and controls in terms of age, gender, body mass index and physical activity. axSpA patients had a mean symptoms duration of 6.5 ± 3.2 years, BASDAI 2.7±2.3 and BASFI 0.9±3.1; 68% were classified as r-axSpA.
Regarding MPP, axSpA patients had no significant difference in muscle stiffness, tone and decrement in the trunk, upper and lower limbs, compared to HC. Overall, there were also no significant differences in MPP between patients with r-axSpA and nr-axSpA, except for a lower decrement [(1.09 (0.92-1.11) vs 1.37 (1.14-1.89), p=0.026] and higher tonus [17.50 (15.80-19.70) vs 15.10 (14.10-15.30) Hz, p=0.039] in the dominant side of the trunk and lower limbs, respectively, in patients with r-axSpA.
In regard to segmental muscle strength, and compared to HC, axSpA patients had significant lower muscle strength in the upper [(axSpA 47.60 (40.15 – 73.20) vs HC 71.75 (51.93 – 80.50) Nm/s, p=0.023] and lower limbs [51.0 (38.5 – 57.1) vs 59.83 (54.6 – 64.45) Nm/s, p=0.001], but not in trunk [56.3 (37.6-67.2) vs 57.30 (51.2-63.0) Nm/s, p=0.669]. When comparing r-axSpA and nr-axSpA patients, there were no statistically significant differences in muscle strength of each segment (trunk, upper or lower limbs).
Conclusion: Young patients with r-axSpA showed lower decrement in the trunk and higher tonus in the lower limbs, compared with nr-axSpA. No differences were registered for strength. These results may speculate about the contribution of muscle to the disease physiopathology and to the identification of patients that may benefit from an early physiotherapy intervention.

P0144 – ANTI-KU ANTIBODIES: CLINICAL AND ANALYTICAL ASSOCIATIONS IN PATIENTS WITH CONNECTIVE TISSUE DISEASES

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Background: Ku is a protein that binds to double-strand DNA and is involved in repairing DNA breaks. Anti-Ku antibodies are associated with several connective tissue diseases (CTD). However, they are rare, and their clinical relevance needs to be clarified.
Objective: To determine the clinical and analytical manifestations associated with anti-Ku antibodies in patients with CTD.
Methods: We included all patients evaluated at our Rheumatology Department between January 2007 and December 2019 for a clinical hypothesis of CTD, whose blood tests for anti-nuclear antibodies performed at the Immunology lab of our hospital identified anti-ku antibodies. Anti-ku antibodies were detected by immunoblot assay. We reviewed patient files to retrieve the diagnoses of CTD, and the clinical-analytical and immunological features of the disease.
Results: Fifteen patients with anti-Ku antibody positive (female: 66%, mean age =59.6 years) were identified. At inclusion, the mean disease duration from first manifestations was 7.6 years. From these patients, 86.7% were diagnosed with a CTD (systemic lupus erythematosus: n=4; undifferentiated connective tissue disease: n=4; rheumatoid arthritis: n=2; myositis: n=1; systemic sclerosis: n= 1; overlap syndrome: n=1. Only two patients were asymptomatic carriers (13.3%).
At CTD inception, the most frequent presenting manifestations were: arthralgias =69%; myalgias and/or muscular weakness =38%; Raynaud's phenomenon =30%; malar rash =23%; sicca syndrome=15%; lymphopenia =15% and alopecia =7.7%.
Anti-ku positivity first presented during follow-up in 20% of patients. New CTD manifestations accrued during follow-up included cytopenias in 30.8%; skin thickening in 15.4%, digital ulcers in 7.7%, interstitial lung disease in 7.7% and nephritis in 7.7%.
Complement levels of C3 and C4 were low in 4 patients. In 5 patients, anti-ku antibody was the only positive autoantibody specificity. In 10 patients, other autoantibody specificities were present: anti-SSA (n=6); anti-double-stranded DNA (n=4); anti-nucleosome (n=3); anti-Sm (n=2); anticentromere (n=2); anti -Pm/Scl (n=2); anti-RNP (n=2); anti-ribosome (n=2); anti-U3RNP (n=1); anti-Th/To (n=1); anti-nucleolar (n=1); anti-SRP (n=1) and anti-Mi (n=1).
Conclusion: Positive anti-Ku is rare among patients with CTD, while most individuals with these antibodies do have a CTD. These patients, regardless of the specific type of CTD, present a common disease phenotype characterized by predominantly musculoskeletal features. Next step is to do a multicenter study with a larger, more representative study population.

P0149 – IMPACTO DA OBESIDADE NA GRAVIDADE DA ARTRITE PSORIÁTICA

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Introdução: Doentes com artrite psoriática (APso) têm uma elevada prevalência de obesidade quando comparados com a população em geral. Alguns estudos recentes têm sugerido uma associação entre a obesidade e a gravidade da doença e um efeito benéfico com a redução ponderal.
Objetivo: Avaliar a influência do índice de massa corporal (IMC) na gravidade da doença nos doentes com APso.
Materiais e métodos: Estudo transversal, retrospetivo e unicêntrico em doentes com APso propostos para início de terapêutica biotecnológica. A amostra obtida foi caracterizada segundo género, idade, duração da doença, hábitos tabágicos e alcoólicos e tratamento farmacológico. Para avaliação da atividade da doença foram utilizados os seguintes parâmetros: contagem articular (68 articulações dolorosas/66 tumefactas); proteína C reativa (PCR) e velocidade de sedimentação (VS); escala visual analógica (EVA) da dor do doente, da dor noturna na coluna e atividade global da doença segundo o doente e o médico; BASDAI (Bath Ankylosing Spondylitis Disease Activity Index); ASDAS (Ankylosing Spondylitis Disease Activity Score); MASES (Maastricht Ankylosing Spondylitis Enthesitis Score); BASFI (Bath Ankylosing Spondylitis Functional Index) e BASMI (Bath Ankylosing Spondylitis Metrology Index).
Resultados: Foram incluídos 94 doentes (50% do sexo masculino), 70 (74.5%) com idade acima dos 40 anos. Relativamente à forma da doença, 28 (29.8%)

apresentavam doença exclusivamente periférica e 48 (51.1%) doença axial. Quarenta e três (45.7%) tinham envolvimento das enteses e 38 (40.4%) história de dactilite. De acordo com os critérios da World Health Organization, 33 (35.1%) doentes têm peso normal ou baixo peso (IMC<24.9), 37 (39.4%) excesso de peso (IMC 25-29.9) e 24 (25.5%) obesidade (IMC³30). Oitenta e três (88.3%) estão medicados com anti-inflamatório não esteroide e 70 (74.5%) com classical disease-modifying antirheumatic drugs.

Na ausência de um critério único para avaliação da gravidade da APso, foi realizada uma análise de clustering (usando o algoritmo das K-médias) às variáveis associadas à atividade da doença. Foram identificados dois clusters: o grupo 1 (n= 45) com maior gravidade e o grupo 2 (n=34) com menor gravidade. Relativamente ao IMC, observaram-se as seguintes frequências absolutas entre os grupos 1 e 2, respetivamente: peso normal (18 vs 13), excesso de peso (14 vs 15) e obesidade (14 vs 6). Apesar do desequilíbrio marcado de frequências entre os obesos, não foi identificada uma associação estatisticamente significativa entre o IMC e a atividade da doença (p=0.318, teste do $\chi 2$). Também não foram encontradas associações significativas entre os grupos de gravidade e a idade, os hábitos tabágicos ou os hábitos alcoólicos (p=0.296, 0.910 e 0.344, respetivamente).

Discussão e **conclusão:** Neste estudo não se constataram associações estatisticamente significativas entre o IMC e a gravidade da doença na APso. No entanto, as frequências dos obesos nos dois grupos sugere uma distribuição desigual do IMC (70% dos obesos apresentam a maior gravidade da doença) pelo que a falta de significância estatística pode estar relacionada com o modesto tamanho da amostra. Outro fator confundidor da associação entre IMC e gravidade poderá ser o elevado índice de atividade apresentado pelos doentes da amostra, uma vez que todos eles estão propostos para terapêutica biotecnológica por falha das terapêuticas convencionais. Portanto, os valores amostrais sugerem a condução de um estudo mais abrangente, quer ao nível do tamanho amostral, quer ao nível da atividade da doença.

P0151 – IMPACTO DAS DOENÇAS REUMÁTICAS: UM QUESTIONÁRIO DE 8 ITENS

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Introdução: Em Portugal, mais de 50% da população apresenta pelo menos uma manifestação de doença reumática. As doenças músculo-esqueléticas limitam de forma significativa a mobilidade, levando muitas vezes à reforma antecipada e à redução da vida social do doente. Assim sendo, os doentes apresentam uma pior qualidade de vida e um aumento dos gastos em saúde. A qualidade de vida destes doentes é afetada de forma complexa, por factores individuais, clínicos e relacionados com a doença.

Objetivo: Avaliar o impacto das doenças reumáticas na qualidade de vida em diferentes patologias reumáticas.

Métodos: O estudo incluiu 220 doentes, seguidos em consulta de Reumatologia de dois centros (ULS-Guarda e CHULN). Os critérios de inclusão incluíam: idade> 18 anos, diagnóstico de doença reumática e capacidade cognitiva para compreender o questionário. O questionário era composto por uma lista de 8 queixas frequentes nas doenças reumáticas (dor, rigidez matinal, incapacidade física, cansaço, sono, disfunção social, bem-estar emocional e problemas sexuais). Foram registados dados demográficos e clínicos, sendo que para efeitos do estudo classificamos os doentes em grupos de diagnóstico. A correlação de Pearson foi usada para comparar as diferenças entre as queixas e as patologias. O estudo foi aprovado pela comissão de ética e todos os pacientes assinaram o consentimento informado.

Resultados: 71.7% dos pacientes eram do sexo feminino, média de idade de 58.0±15.0 anos. Seis grupos de diagnóstico foram estabelecidos: doenças reumáticas inflamatórias (58.2%), doenças do tecido conjuntivo (DTC) (15.5%), osteoartrose (9.1%), osteoporose (7.7%), doenças dos tecidos moles (6.4%) e doenças de deposição de cristais (2.7%).

A principal queixa foi a dor, reportada por 80.9% dos doentes, seguida pelo cansaço (60.9%) e incapacidade física (51.8%). A correlação de Pearson mostrou relação inversa da dor com as DTC e correlação positiva da dor com osteoartrose. As DTC mostraram correlação positiva com distúrbios do sono. Na comparação individual das doenças, foi encontrada uma diferença estatisticamente significativa entre a rigidez matinal e a Artrite Psoriática (AP). O Lúpus Eritematoso Sistémico (LES) correlaciona-se inversamente com a dor e directamente com os problemas sociais e sexuais. Quanto à Síndrome de Sjögren (SSj), correlacionou-se inversamente com a dor e directamente com os problemas sociais e bem-estar emocional.

Conclusões: Os pacientes com LES e SSj foram os que menos reportaram dor, explicando a correlação inversa que foi encontrada com as DTC. A dor foi a principal queixa reportada pelos doentes com osteoartrose, estando de acordo com a literatura. Quanto à relação da AP e da rigidez matinal, não existem publicações que destaquem esta associação, comparativamente a outras doenças reumáticas.
De acordo com a literatura, o impacto do LES na disfunção sexual foi menos estudado. Stein et al. reporta que 4% dos doentes com LES apresentam problemas sexuais, no nosso estudo 21% dos pacientes reportaram disfunção sexual.
Os distúrbios do sono estão descritos em mais de 50% dos doentes com LES, reportados neste estudo em 36% dos doentes. Entre as hipóteses dos mecanismos que possam explicar esta associação, foram apontados factores psicossociais e psicológicos. A prevalência de depressão e ansiedade é elevada em doentes com Sjögren. 42% dos pacientes reportaram que a doença afetava o bem-estar emocional.
Com este estudo, pretendemos destacar aspetos que afetam os doentes com patologia reumática e para os quais o médico deverá estar atento na sua avaliação.

P0152 – PREDICTING CARDIOVASCULAR EVENTS IN PATIENTS WITH SPONDYLOARTHRITIS AND PSORIATIC ARTHRITIS: THREE RISK ALGORITHMS

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Background: Inflammatory joint disorders have an increased cardiovascular (CV) risk when compared with general population. In 2009, the EULAR task force advocated the use of a 1.5 multiplication factor for these risk prediction models in Rheumatoid Arthritis patients with specific characteristics. The SCORE was not modified in patients with Psoriatic Arthritis (PsA) and Spondyloarthritis (SpA) in accordance with the present EULAR recommendations.
Objectives: To assess the accuracy of several CV risk algorithms to predict an event and determine its sensibility and specificity
Methods: A retrospective analysis of PsA and SpA patients, followed in our department in Local Health Unit of Guarda during one year (2019) was done. We determined CV risk profile of our patients using the following data: gender, age, smoking status, blood pressure, lipid values and diabetes mellitus status. These variables were used to calculate risk prediction algorithms such as Framingham, the American College of Cardiology/American Heart Association (ACC/AHA) risk score and the Systematic Coronary Risk Evaluation (SCORE). Discriminatory ability for CV risk prediction was evaluated by the area under the ROC curves. Sensibility and specificity were calculated for low-to-intermediate and intermediate-to-high risk cut-offs. Cut-off values that mark the high risk were defined in 5% for SCORE, 20% for Framingham and ACC/AHA.

TABLE 1: SENSIBILITY, SPECIFICITY, POSITIVE PREDICTIVE VALUE (PPV) AND NEGATIVE PREDICTIVE VALUE (NPV) OF CV RISK ALGORITHMS

	Sensibility (%)	Specificity (%)	PPV (%)	NPV (%)
SCORE > 1%				
SpA	75	63	18	96
PsA	100	41	5	100
SCORE > 5%				
SpA	NA	NA	NA	NA
PsA	0	93	0	97
Framingham >10%				
SpA	83	78	28	98
PsA	50	51	5	95
Framingham >20%				
SpA	33	86	20	93
PsA	50	69	8	96
ACC/AHA >5%				
SpA	67	77	29	94
PsA	100	44	9	100
ACC/AHA >20%				
SpA	17	86	14	88
PsA	50	83	14	97

Results: 112 caucasic patients were included, 61% female with a mean age of 52.15±14.18 years and mean BMI of 27.11±4.81kg/m2. 7 patients weren´t eligible to apply these cardiovascular scores. 8 patients were identified with non-fatal cardiovascular events (2 cases of stroke, 5 cases of myocardial infarction and 1 case of thrombophlebitis). 69 patients diagnosed with SpA and 43 with PsA, five CV events identified in SpA patients. Area under the ROC in SpA were 0.729 (95% CI 0.461 to 0.996) for SCORE, 0.839 (95% CI 0.735 to 0.943) for Framingham and 0.804 (95% CI 0.683 to 0.925) for ACC/AHA. Area under the ROC in PsA patients were 0.603 (95% CI 0.327 to 0.879) for SCORE, 0.660 (95% CI 0.326 to 0.995) for Framingham and 0.804 (95% CI 0683 to 0.925) for ACC/AHA. Sensibility and specificity were discriminated in table 1.
Conclusions: A good discrimination between patients with or without CV events has been demonstrated by area under the ROC curve, particularly to SpA patients. In PsA, the sample was smaller, which represents a limitation in this study. SCORE >5% did not identify CV events, therefore sensibility couldn't be calculated. Overall, the algorithms studied presented a low sensibility, underestimating CV risk. Main reason for this is

the fact that disease-related factors are not included in these scores. We are of the opinion that better algorithms are needed to correctly assess cardiovascular risk algorithms for SpA and PsA, since the majority of the events occur in patients with low-intermediate risk.

P0160 – SMOKING AS PREDICTIVE FACTOR FOR SPONDYLOARTHRITIS RELATED UVEITIS: RESULTS FROM A SINGLE CENTRE CROSS-SECTIONAL STUDY

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Background: Although spondyloarthritis (SpA) is primarily a musculoskeletal condition, ocular involvement is an important clinical feature and contributes to the burden of disease. Acute anterior uveitis (AAU) is classically described as the most frequent extra-articular manifestation of SpA and in some cases the first clinical presentation. The prevalence of AAU varies according to the subtype of SpA. In a systematic literature review, the mean prevalence of AAU was 32.7% and a positive association between HLA-B27 positivity, axial SpA, male sex and uveitis has been reported. More recently, some cross-sectional studies have described lower odds of spondyloarthritis-related uveitis (SpA-U) in smokers than in patients who are ex smokers or never smokers. Predictors of SpA-U are poorly defined in literature and the influence of smoking status remains controversial.
Objectives: To analyse the factors associated with uveitis in SpA patients in a Tertiary Rheumatology Center.
Methods: An observational cross-sectional study was performed including patients fulfilling the ASAS criteria for axial SpA with a follow-up visit between January and June 2019. Clinical patients' charts were reviewed and the following variables were considered: age, gender, history of uveitis (confirmed by ophthalmologist observation), number of AAU episodes, smoking status (never smoker or ever smoker), HLA-B27, disease duration, disease involvement (exclusively axial or axial and peripheral), history of enthesitis and syndesmophytes. History of AAU and associated variables were determined in this subset of patients.
Statistical analysis was performed with logistic regression model. P value <.05 was defined as statistically significant.
Results: The study included 164 patients (62.3% men) with median age of 44.0 years (IQR 37 to 54) and a median disease duration of 14.6 years (IQR 9.28 to 20.32). SpA diagnosis was ankylosing spondylitis in 70.7% cases and the remaining were non-radiographic axial SpA. HLA-B27 was positive in 84.8%, 31.1% of patients were ever smokers and 21% had both axial and peripheral joint involvement. Twenty four percent of patients had at least one AAU episode. Recurrence of uveitis occurred in 70% of patients. Ever smoking (OR=2.256; 95%CI [1.077-4.276]; p<.05) and syndesmophytes (OR=2.125; 95%CI [1.009-4.475]; p<.05) showed a statistically significant association with uveitis in univariated logistic regression. Althougth not statistically significant, a trend to association was found between smoking and recurrence of AAU (OR=2.235; 95%ICI [.973-5.135], p=.058). In multivariated logistic regression only ever smoking was independently associated with uveitis (OR=2.542; 95%CI [1.007-6.420]; p<.05). We did not find association between presence of uveitis and gender, age, disease duration, disease involvement, HLA-B27 positivity and enthesitis.
Conclusion: Contrary to few cross-sectional studies showing a possible protective effect of smoking in SpA-U, and in line with new data from Zhao et al (1), we report a statistically significant independent association between history of smoking and uveitis. Nevertheless, we emphasize the need of more studies to confirm these findings.

References:

1. Zhao S, et al. Smoking does not protect patients with axial spondyloarthritis from attacks of uveitis. Annals of Rheumatic Diseases 2019;78(9).

P0163 – GAIT PATTERN DIFFERENCES BETWEEN PATIENTS WITH RADIOGRAPHIC AND NON-RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS, THE MYOSPA STUDY

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Background: Axial spondyloarthritis (axSpA) is a chronic inflammatory disease classified as radiographic (r-axSpA) or non-radiographic (nr-axSpA). Defining the gait patterns associated with these two groups can improve its detection and promote early intervention. In normal walking, body segments move around the joints as struts of an inverted pendulum. The resultant cyclic rotations contribute to the forward translation of the body, while minimizing muscle work and maintaining stability. Recent literature describes a decline in this pendulum-like mechanism associated with aging and some neurological diseases (Parkinson and multiple sclerosis).
Objective: The aim was to compare the 3D gait kinematics of patients with r-axSpA and nr-axSpA.
Methods: A cross-sectional study was conducted on 54 participants (18-50 years old), 27 patients with axSpA (according to ASAS criteria, with less than 10 years since symptoms onset) and 27 healthy controls, matched by gender, age and level of physical activity. A sub-analysis was performed involving the whole group of patients classified as r-axSpA (n=14) and nr-axSpA (n=6). Subjects movement was reconstructed using a 3D full-body kinematic model (Kinetikos, Coimbra, Portugal) fed by 15 inertial sensors placed in the head, arms, trunk, pelvis, thighs, shanks and feet. 3D gait kinematics was characterised based on variables that analyse the body movement as a whole (e.g. center of mass displacement, speed), conventional spatiotemporal parameters (e.g. stance/swing time, step length) and joints kinematics time-normalized to 101 points, comprising the gait cycle from 0 to 100%. Nonparametric statistical tests were used.
Results: In the r-axSpA group, 71,4% were male, with a mean age of 34.43±7.84 years and a BASDAI of 2.84±2.39, whereas in the nr-axSpA, 50% were male, with a mean age of 41.83±6.27 years and a BASDAI of 2.99±0.58. A statistically significant difference was observed in the displacement of the center of mass (with respect to the pelvis local coordinate system) along the anteroposterior axis between the two studied groups ($H = 4.96$, $p = 0.03$), with a mean rank displacement of 8.6 for r-axSpA and 15.00 for nr-axSpa, corresponding to a reduction in displacement of 38% (mean 0.00986 vs 0.01579m), in the r-axSpa group.
Conclusion : Our preliminary results in r-axSpA subjects show a reduction of the pendulum mechanism. Although no significant segmental (kinematics) changes were observed, the sum of all studied variables result in a clear different gait pattern between the two groups. The observed decline can be an early sign of the inefficiency of the r-axSpA group to minimise the cost of transport of the center of mass during walking (i.e. increased instability). This study shows the potential of gait analysis to identify subjects who may benefit from early physiotherapy intervention.

P0168 – TIME-COURSE CHANGE IN AXIAL MOBILITY IN PSORIATIC ARTHRITIS PATIENTS UNDER BDMARD

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Background: Spinal mobility is assessed frequently in patients with psoriatic arthritis (PsA) using Bath Ankylosing Spondylitis Metrology Index (BASMI) to provide baseline measurement, monitor changes over time and to assess the impact of clinical interventions. BASMI comprises 4 measures of spinal mobility (cervical rotation, tragus-to-wall distance, modified Schober's test and lumbar lateral flexion) and one hip mobility measurement (intermalleolar distance).
Objectives: The aim of this study is to investigate the time-course change of BASMI in PsA patients after 6 months of Biologic Disease-modifying Antirheumatic Drug (bDMARD) therapy. The authors also pretend to evaluated, at baseline and after 6 months of treatment, the association between BASMI, disease activity scores and physical function.
Methods: An observational retrospective study was performed in patients with PsA under bDMARD followed in the Rheumatology department of a tertiary university hospital. Were included patients treated with only one bDMARD. Demographic and clinical data were collected from the Rheumatic Diseases Portuguese Register. For spinal mobility calculation BASMI was used. Disease activity was evaluated with ASDAS and BASDAI. Physical function was assessed with BASFI. The variation of BASMI, ASDAS, BASDAI and BASFI was calculated as the difference between values registered at 6 months and at baseline and presented as Δ. Correlations between ΔBASMI, ΔASDAS and ΔBASFI was calculated using Pearson test.
Results: A total of 55 patients were included. Thirty patients were males (54.5%). The mean age at diagnosis

was 44.6 ± 12.6 years and the median disease duration at start of bDMARD was 5.4 years (min: 0.30; max: 25.5). In total, 19 (34.5%) patients had predominant axial involvement, 36 (65.5%) peripheric and 36 (65.5%) enthesopathic. Almost all patients fulfilled the CASPAR criteria for PsA (n=50, 90.9%). According to ASDAS criteria, at the baseline 20 patients (36.4%) had high disease activity and 34 (61,8%) very high. The most used bDMARD was etanercept (n=21, 38,3%) followed by golimumab (n=19, 34.5%) and adalimumab (n=8, 14.5%). Three patients were treated with infliximab, two with certolizumab and other two with secukinumab. Axial PsA patients had more limitations in spinal mobility (BASMI mean 4.5 ± 1.5) and more functional limitation (BASFI mean 6.8±1.9) than patients with predominant peripheric involvement (BASMI mean 3.3± 1.2, p=0.004; BASFI mean 5.4±3, p=0,0048). Statistically significant differences in ASDAS and BASDAI in these two groups were not observed (p=0.332 and p=0.605, respectively). For all patients, BASMI did not vary significantly (p=0.691) at baseline (mean 3.7± 1.4) and after 6 months (mean 3.8±1.3) of treatment. Although the ΔBASMI for etanercept was negative (mean -0.12±0.9) and for golimumab positive (0.14±0.8), it was not statistically significant. At baseline there is a significant positive association between BASMI and ASDAS (r=0.435, p=0.001), BASMI and BASDAI (r=0.567, p<0.001) and BASMI and BASFI (r=0.510, p<0.001). However, there was not a statistically significant association between ΔBASMI and: ΔASDAS, ΔBASDAI and ΔBASFI (r=0.158; p=0.269, r=0.019; p=0.096 and r=0.121; p=0.397, respectively).

Conclusion: In PsA patients treated with bDMARDs, at least in short-term follow-up, BASMI does not improve with time. Changes in BASMI did not correlate with changes in activity disease and in functional outcome. Studies with longer follow-up and with more patients are needed to better evaluate these associations.

P0188 – RISK FACTORS FOR FLARES IN PREGNANT WOMEN WITH SYSTEMIC LUPUS ERYTHEMATOSUS: A 10-YEAR STUDY IN A TERTIARY CENTRE

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Background: Pregnant women with systemic lupus erythematosus (SLE) are at high risk for both adverse pregnancy outcomes (APOs) and disease flares. At least 50% of SLE patients may present active disease during pregnancy. Severe flares, however, are not frequent, and have been associated with a history of lupus nephritis (LN), disease activity at conception and hydroxychloroquine (HCQ) withdrawal.

Aim: To characterize the cohort of pregnant SLE patients followed at our centre and to identify risk factors for SLE flares.

Methods: Single-centre observational retrospective study of pregnant women with SLE followed at Hospital de Santa Maria between 2009 and 2019 in a Rheumatology-Obstetrician multidisciplinary clinic. Demographic and clinical data, medication, APOs and disease activity measured by the SLEDAI-2K on the periconceptional period and during each trimester of pregnancy were collected. Flare was classified according to the SELENA-SLEDAI flare index (SFI) as light/moderate or severe. Clinical variables were described as means ± standard deviation, medians with interquartile range (IQR) or frequencies, as appropriate. Univariate analysis was performed with Chi-Square test for categorical variables or Kruskal-Wallis test for continuous variables. Clinically and statistically relevant variables were introduced in a multinomial regression model to predict SLE flares.

Results: We followed 113 pregnancies in 88 patients with SLE, with a mean age at diagnosis of 24.9±6.8 years. Regarding comorbidities, 8 (9.1%) women had arterial hypertension, 2 (2.3%) were obese, and 1 (1.1%) had diabetes. Maternal age at conception was 32.6±5.3 years, with pregnancies occurring 7.5±5.6 years after diagnosis. APOs occurred in 43 (38.1%) pregnancies. Major APOs recorded were 16 (14.2%) preterm births, 16 (14.2%) small for gestational age, 15 (13.3%) miscarriages, 9 (7.9%) foetal growth restriction, 6 (5.3%) preeclampsias, 3 (2.7%) newborn infections in the first month, 2 (3.5%) malformations (1 trisomy 18 and 1 warfarin embryopathy), 2 (1.8%) foetal deaths, and 2 (1.8%) terminations of pregnancy (1 after exposure to methotrexate and 1 due to warfarin embryopathy). Neonatal lupus was recorded in 2 newborns from positive anti-SSA mothers (1 congenital heart block and 1 haematological neonatal lupus). Information on disease activity during pregnancy

TABLE 1 – CHARACTERISTICS OF PATIENTS ACCORDING TO FLARE CATEGORY AND REGRESSION MODEL FOR PREDICTION OF FLARES

Characteristic	No flare (N=68)	Mild/moderate flare (N=9)	Severe flare (N=7)	p-value	Multinomial regression – Mild/moderate flares			Multinomial regression – Severe flares		
					OR	95%CI	p-value	OR	95%CI	p-value
Disease duration (years)	7.6±5.9	9.7±6.6	6.6±5.6	0.624	0.97	0.82-1.15	0.667	1.14	0.91-1.44	0.251
Maternal age (years)	33.3±5.6	31±6.1	31.4±5.5	0.388						
Cutaneous involvement, N (%)	52 (76.5)	9 (100)	7 (100)	0.098						
Hematologic involvement, N (%)	28 (42.2)	7 (77.8)	4 (57.1)	0.099						
Articular involvement, N (%)	52 (76.5)	7 (77.8)	6 (85.7)	0.856						
Kidney involvement, N (%)	**6 (8.8)**	**4 (44.4)**	**1 (14.3)**	**0.014**	0.26	0.02-3.61	0.312	10.55	0.50-223.82	0.131
CNS involvement, N (%)	3 (4.4)	0 (0)	0 (0)	0.693						
Secondary APS, N (%)	13 (19.1)	2 (22.2)	0 (0)	0.425						
Secondary SS, N (%)	4 (5.9)	2 (22.2)	0 (0)	0.151						
HCQ at conception, N (%)	51 (75.0)	6 (66.7)	7 (100)	0.260	3.44	0.33-36.0	0.303	*	*	*
AZA at conception, N (%)	11 (16.2)	4 (44.4)	3 (42.9)	0.054	3.60	0.36-36.43	0.278	1.39	0.15-12.58	0.769
HCQ during pregnancy, N (%)	55 (80.9)	8 (88.9)	7 (100)	0.388						
AZA during pregnancy, N (%)	**13 (19.2)**	**6 (66.7)**	**4 (57.1)**	**0.002**						
GC during pregnancy, N (%)	**43 (63.2)**	**9 (100)**	**7 (100)**	**0.015**						
SLEDAI at conception	**1.62±2.8**	**4.1±4.2**	**7.2±5.9**	**0.005**	1.058	0.82-1.15	0.667	**1.50**	**1.12-2.02**	**0.007**
Active LN at conception, N (%)	4 (5.9)	1 (11.1)	1 (14.3)	0.633						
APOs, N (%)	23 (33.8)	3 (33.3)	3 (42.9)	0.914						

*Every patient with a severe flare was on HCQ
APO – adverse pregnancy outcomes; APS – anti-phospholipid syndrome; AZA – Azathioprine, CNS – central nervous system; GC – glucocorticoids; HCQ – hydroxychloroquine; LN – lupus nephritis; SS – Sjögren syndrome

was available in 84 pregnancies – flares occurred in 16 (19.0%) pregnancies: 9 (10.7%) were light/moderate and 7 (8.3%) severe. Characteristics of patients according to flare category are presented in table 1. In univariate analysis, higher SLEDAI at conception was associated with flares (p=0.005). Patients with flares were also more likely to need treatment with glucocorticoids (GC) and azathioprine (AZA) during pregnancy. In a multivariate multinomial regression model, adjusted for disease duration, SLEDAI at conception, history of LN, therapy with HCQ and AZA at conception, only higher SLEDAI at conception was associated with severe flares (OR 1.50, 95% CI 1.12-2.02, p=0.007).

Conclusions: SLE flares according to the SFI occurred in 19.0% of pregnancies, which is in line with data previously reported by other European cohorts. Flares led to more frequent use of GC and AZA during pregnancy. Higher SLEDAI at conception was independently associated with severe flares, highlighting the need for adequate disease control when planning a pregnancy in a patient with SLE.

P0196 – EFFICACY AND SAFETY OF METHOTREXATE IN GIANT CELL ARTERITIS: RESULTS FROM A BICENTRIC PORTUGUESE COHORT STUDY

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Background: Giant cell arteritis (GCA) is a vasculitis of large- and medium-sized arteries affecting patients over 50 years old. High-dose glucocorticoid (GC) therapy should be initiated promptly for the induction of remission in active GCA. Adjunctive therapy with methotrexate (MTX) was tested in four randomized-controlled trials but only one met its primary endpoint. In all studies, the dose of MTX used was generally low (7.5-15 mg per week), and more recent evidence suggests that MTX may still be a valid option for patients with GCA.

Objectives: Evaluate the effectiveness and safety of MTX in a cohort of Portuguese patients with GCA followed at two tertiary centres.

Methods: Bicentric observational study using data from the Rheumatic Diseases Portuguese Register (Reuma.pt), complemented with data from hospital clinical records. We included patients with biopsy -proven GCA or with the "halo sign" on ultrasound,

TABLE 1 – CHARACTERISTICS OF THE PATIENTS WITH GCA TREATED WITH GCS ALONE COMPARED WITH PATIENTS TREATED WITH GCS+MTX

Characteristics	GCs alone N=64	GCs+MTX N=66	p-value
Age at diagnosis, years	75.5 (13.0)	74.8 (10.6)	0.117
Female sex, n (%)	42 (65.6)	45 (68.2)	0.853
Body mass index, Kg/m^2	25.1 (7.9)	27.1 (6.3)	0.106
Diabetes, n (%)	16 (25.0)	27 (40.9)	0.064
Hypertension, n (%)	41 (64.1)	49 (74.2)	0.255
Delay in diagnosis, months	1.0 (2)	1.5 (4.0)	**0.046**
Ischemic event, n (%)	28 (43.8)	38 (57.6)	0.16
PMR features, n (%)	26 (40.6)	33 (50.0)	0.378
GC starting dose, mg	60.0 (20.0)	60.0 (20.0)	0.706
GC dose at index date, mg	15.0 (12.5)	20.0 (32.5)	**0.022**
GC cumulative dose at index date, mg	5335.0 (2677.2)	5515.0 (6662.5)	0.780
GC cumulative dose 6 months after index date, mg	6265.0 (2545.7)	91091.3 (6868.1)	0.069
GC cumulative dose 12 months after index date, mg	8050.0 (3751.6)	10395.0 (6160.0)	0.164
Total GC cumulative dose, mg	7327.5 (6470.0)	13710.0 (7767.5)	**<0.001**
ESR at diagnosis, mm/hour	85.0 (46.0)	80.0 (44.0)	0.466
CRP at diagnosis, mg/dL	4.6 (6.2)	4.87 (5.9)	0.777
Active disease at 6-months, n (%)	6 (9.4)	7 (10.6)	0.334
Active disease at 12-months, n (%)	3 (4.7)	6 (9.1)	0.916

Data for continuous variables are presented as median (IQR), and for categorical variables as absolute number (%). Index date relates to the time of MTX initiation in the GCs+MTX group and correspondent date in the GCs alone group.
CRP – C-reactive protein; GCs – glucocorticoids (prednisolone equivalent); ESR – erythrocyte sedimentation rate; MTX – methotrexate; PMR – polymyalgia rheumatica.

treated with GCs and/or MTX. Clinical features of patients treated with GCs alone or GCs+MTX were compared using exact Fisher and Mann-Whitney U tests, as adequate. Association between continuous variables was assessed with Spearman's correlation coefficient. For patients on the GCs alone group, an index date, correspondent to the median time to MTX start in the GCs+MTX group, was established for comparison of disease activity and total GC dose at 6- and 12-months. Active disease was defined according to the clinician's judgment upon presence of GCA-related symptoms and/or increased inflammatory markers. The relapse rate was defined as the number of events per patient-years (p-y) of follow-up, and relapse rates with or without exposure to MTX were compared using mid p-value test.

Results: We included 130 patients, with a median age at diagnosis of 74.9 years (IQR 11.8), 66.9% female. Sixty-six (50.8%) patients received MTX with a median time to start of 174 days (IQR 314), a median starting dose of 10 mg/week (range 5-15) and a median maximum dose of 15 mg (range 5-25). Characteristics of patients in the two treatment groups are summarized in Table 1. Inflammatory markers and proportion of patients with active disease at 6- and 12-months were not reduced in patients receiving MTX. Patients on the GCs+MTX group had a significantly higher median GCs dose at index date and total GC cumulative dose, but a longer time to start MTX positively correlated with total GC cumulative dose (r=0.32, p=0.01). Relapses occurred in 28 patients, in a median of 13.5 (IQR 43.0) months from diagnosis: the relapse rate in patients exposed to MTX was 2.75/100 p-y vs. 6.72/100 p-y in patients not exposed to MTX, with a rate ratio reduction of 0.41 (95%CI: 0.17-0.91). In subgroup analysis, comparing patients with or without relapses, cumulative GC dose at any time point was not different between groups. Twenty-one patients (31.8%) stopped MTX: 2 due to disease remission and 19 due to adverse events (AEs) – 10 mild, 7 moderate, and 2 severe AEs (pneumonitis and Kaposi sarcoma).

Conclusions: Treatment with MTX in GCA was associated with a reduced relapse rate during follow-up. Cumulative GC dose was higher in patients in the GCs+MTX group, probably reflecting a more severe disease; however, in subgroup analysis, GC dose was not different in patients with or without relapses. Discontinuation of MTX was higher than usually reported in literature, although the proportion of severe AEs was similar to previous studies. Therefore, adjunctive therapy with MTX should still be considered a safe and effective alternative for relapse reduction in patients with GCA.

P0205 – LONG TERM EXPERIENCE WITH RITUXIMAB IN RHEUMATOID ARTHRITIS PATIENTS

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Background: Rituximab (RTX) is a chimeric monoclonal antibody that selectively targets CD20-positive B cells, approved for the treatment of severe rheumatoid arthritis (RA) patients. It has been observed that some patients remain on RTX treatment for a long time, but in addition to rheumatoid factor (RF) positivity, other predictors of long-term persistence remain unclear.

Objectives: To describe efficacy and safety of rituximab (RTX) infusions in the treatment of RA and characterize long-term responders.

Methods: Data from RA patients registered at Reuma.pt, who started RTX before January 2019

TABLE 1 – BASELINE CHARACTERISTICS AND VARIATION OF DISEASE ACTIVITY, IMMUNOGLOBULIN AND CD19+ LEVELS AT 3MONTHS OF TREATMENT OF PATIENTS WHO WITHDREW RTX TREATMENT AND THOSE WHO PERSISTED ON TREATMENT FOR MORE THAN 42 MONTHS.

	Withdrew from RTX treatment	Long-term RTX persistence (> 42 months)	*p value*
Number of patients (N)	25	16	-
RF and/or ACPA positivity - N(%)	21 (84)	14 (87.5)	0.57
Erosive disease - N(%)	14 (63.6)	10 (76.9)	0.33
Number of previous bDMARD	2 ± 1.3	1.63 ± 0.6	0.22
Age at first bDMARD	53.1±13.4	52.7±12.4	0.93
Years of disease until first bDMARD	9.1 (2.6-15.1)	11.1 (4.6-13.5)	0.41
Concomitant LFN or MTX - N(%)	23 (92)	14 (87.5)	0.51
Concomitant corticosteroids - N(%)	19 (76)	14 (87.5)	0.31
Δ DAS-28 at 3 months	0.72 ± 0.6	1.1 ± 1.1	0.49
Δ IgA at 3 months	-22.1 (-38;-3)	16 (-64.3;60)	0.39
Δ IgG at 3 months	-48 (-106;42)	-32 (-59;141)	0.44
Δ IgM at 3 months	-36 (-56;-6)	-17 (-69;-0.5)	0.81
Δ CD 19+ %cells	-8.3 (-11.9;-3.9)	-11.8 (-15.2;-5.1)	0.39

RF – rheumatoid factor; ACPA – anticitrullinated protein antibody; bDMARD – biologic Disease-modifying anti-rheumatic drugs; LFN – Leflunomide; MTX – methotrexate; Δ- variation between baseline and 3 months of treatment; DAS – Disease Activity Score; Ig – immunoglobulin.

were analysed. Demographic and clinical variables, previous exposure to bDMARDs, number of RTX cycles, as well as EULAR response rate and adverse events are described. Baseline variables were compared between patients with long-term persistence on treatment (upper quartile of treatment duration) and those who withdrew from RTX treatment.

Results: A total of 60 patients were included, of which 88.3% were females, 90% were RF and/or anticitrullinated protein antibody (ACPA) positive and 61.5% had erosive disease. Before treatment with RTX, patients had already been treated in average with 1.85 ± 1.1 previous biologic agents. The median follow up time on RTX was 23.2 (IQR 14.3-43) months. The median number of RTX cycles per patient was 3 (IQR 2-5) and the median time for retreatment was 9.3 (IQR 6.7-13.7) months. In our cohort 58.3% discontinued treatment with RTX after a mean of 30 ± 21 months. The main reasons for RTX discontinuation were inefficacy (31.4%), adverse events (17.1%), death (14.3%) and remission (11.4%). In total, 87% maintain concomitant treatment with methotrexate and/or leflunomide. At baseline, patients had high disease activity (mean DAS 28 5.5±1.5). Immunoglobulins (Ig) levels remained stable along the first year of RTX treatment and as expected the number of CD19+ lymphocytes decreased significantly after RTX administration (10.2% at baseline versus 0.1% at 3 and 6 months).

The EULAR good/moderate response was achieved in 53.3/44.5/44.4 % at 3/6/12 months, respectively. Nineteen adverse events (AE) were recorded, the majority were infections.

There were no significant differences in the baseline characteristic between patients who discontinued and those who maintained long-term RTX treatment (Table 1). Improvement in disease activity, as well as variation of immunoglobulin or CD19 levels at 3 months was also similar between the two groups. Although statistically not significant, the group who withdrew from RTX treatment had lower decrease in CD19+ lymphocytes count and a higher number of previous biologics.

Conclusion: Our results are in line with the known for RTX in the treatment of RA patients, especially those who have failed previous bDMARD and have high baseline disease activity. Long-term persistence on treatment is good, although we could not identify any particular characteristics of these patients, possibly due to the small sample size.

P0226 – CONCORDANCE BETWEEN THE NEW SLE-DAS, DORIS AND DORIA REMISSION CRITERIA FOR SLE: ARE THEY DIFFERENT IN A REAL-LIFE CLINICAL SETTING?

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Background: The treat-to-target strategy in Systemic Lupus Erythematosus (SLE) aims to achieve remission. However, to define a target based on SLE Disease Activity Index (SLE-DAI-2K) is questionable, due to its limitations (especially its dichotomous nature). The currently used Doria and DORIS clinical remission criteria are both based on SLEDAI.

The Systemic Lupus Erythematosus Disease Activity Score (SLE-DAS) is a recently validated continuous disease activity score which demonstrated higher accuracy and improved sensitivity to change as compared to the SLEDAI-2K. In addition, it comprises important manifestations absent in SLEDAI, including hemolytic anemia, gastrointestinal and cardiopulmonary involvement.

Objective: Our primary goal was to compare the attainment of clinical remission defined by SLE-DAS, DORIS and Doria criteria in a real-life clinical setting. In addition, we aimed to evaluate disease activity distribution according SLE-DAS in our cohort of lupus patients.

Methods: Cross-sectional study of consecutive SLE patients fulfilling ACR'97 and/or SLICC'12 classification criteria followed at an academic lupus clinic from January to December 2019.

The SLE-DAS clinical remission criteria were defined as a score of 0 in all clinical items of SLE-DAS and current prednisolone dose ≤ 5 mg/day. The SLE-DAS cut-off values to define disease activity states were previously defined in the Padova Lupus Cohort: low disease activity (LDA) if SLE-DAS ≤ 3.77 in patients not fulfilling remission criteria; mild disease activity if 3.7 < SLE-DAS≤ 7.64; and moderate-to-severe disease activity if SLE-DAS>7.64.

Fulfillment of DORIS, Doria and SLE-DAS clinical remission status was verified for each patient. The attainment of clinical remission for each patient was compared according to these definitions. We further classified all patients regarding each disease activity state.

Results: The study population included 300 patients (female = 86%; mean age = 48.4±14.5 years; mean disease duration =14.1±9.3 years). The proportion of patients in clinical remission was 76% as defined by the DORIS and Doria criteria. Patients in clinical remission according to the SLE-DAS definition exactly matched those defined by either Doria or DORIS criteria and there were no discordant cases.

From patients in clinical remission, 18.4%, 92.5 %, and 30.4% were taking prednisone, antimalarials, and immunosuppressants, respectively.

In addition, the proportion of patients in LDA, mild disease activity and moderate-to-severe disease activity were 9.7%, 6.7% and 7.7%, respectively.

Conclusions In a real-life cohort of SLE patients, clinical remission is consistently defined by applying either SLE-DAS, DORIS or Doria criteria. Importantly, SLE-DAS definition is easier to apply, as it does not require the PGA or additional manifestations not included in SLEDAI. SLE presents a relapsing-remitting disease course despite maintenance treatment according to current good clinical practice. In accordance, in our cohort a few patients present moderate-to-severe activity and most patients achieved the target of remission, with <20% receiving prednisolone.

P0239 – EARLY RETIREMENT AND ITS DETERMINANTS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Work disability is a common consequence of systemic sclerosis (SSc) with economic implications for both the patient and society. However, there is scarse information available on work and disease-related factors associated with early retirement of patients with this condition in Portugal.

TABLE 1 – DEMOGRAPHICS AND CLINICAL CHARACTERISTICS OF THE STUDY POPULATION

	Early retired due to SSc (N= 17)	Professionally active (N=32)	p
Female, n (%)	13 (77)	24 (85.7)	0.5
Current age, years	60.7 (8.2)	50.0 (9.1)	P<0.01
Age of diagnosis	45.6 (10,9)	40,9 (10,7)	0.9
Age at retirement, years	53.3±9.3	NA	NA
Disease duration, years	15.0 (10.4)	10.4 (6.7)	0.01
Tertiary education, n (%)	2(12)	19 (60)	0.03
SSc subtype, n (%)			
Limited	12 (71)	27 (89.3)	0.26
Diffuse	5 (29)	5 (10.7)	0.26
Anti-Scl 70, n (%)	3 (18)	3 (10.7)	0.28
Anti-centromere, n (%)	9 (53)	18 (46.4)	0.82
Clinical manifestations, n (%)			
Raynaud's Phenomenon	17 (100)	31(96.9)	0.46
Lung involvement*	10 (59)	4 (7.1)	0.02
Digital ulcers	11 (65)	7 (28.6)	0.01
GI involvement**	5 (29)	4 (10.7)	0.2
Heart involvement***	8 (47)	4 (10.7)	0.02
Modified Rodnan skin score	8.9 (0-22)	2.5 (0-21)	0.01

NA – non-applicable

Results are expressed in mean (SD), unless otherwise stated.

*Defined as pulmonary involvement was defined as either pulmonary fibrosis (bilateral reticular nodular on chest X-ray, interstitial pneumonitis/ground glass opacities/fibrosis on HRCT), or DLCO <70% of predicted.

**Defined as persistent reflux/indigestion, diarrhea, constipation or dysphagia without another plausible cause.

***Defined as arrhythmias, pulmonary arterial hypertension, pericardial effusion or myocardial dysfunction.

Objectives: To evaluate the rate of early retirement due to SSc; and to identify its main determinants, both work and disease-related.

Methods: Cross-sectional cohort study including patients with SSc according with ACR/EULAR 2013, followed in our department. Patients retired prior to SSc diagnosis, never-employed or with missing information on current work status were excluded. Patients retired due to SSc versus professionally active were compared using T-test and Chi-2 test as appropriate. Variables with p<0.05 in univariate analysis and other potential predictors selected on clinical and epidemiological grounds were included in multivariable binary logistic regression.

Results: 74 SSc patients were included (83% female, aged 59.6±12.1, mean disease duration 10.6±8.0; 87% had the limited form of SSc). Until the present time, 56.8% (n=42) of the patients are retired, this being due to SSc in 23% of the cases. Early retirement due SSc translates into 10 years of active work lost, compared to retirement due to the other causes (53.3±9.3 vs. 63.0±4.0 years). Compared to patients that are still professionally active, patients retired due to SSc had longer disease duration (15.0 ± 8.8 years vs 10.4 ± 6.7 years p=0,018); shorter formal education (88% vs. 40% p=0.03); higher mRSS (8.9 vs. 2.5, p<0.01); and higher cumulative prevalence of digital ulcers (65% vs. 28.6% p=0.01); lung involvement

(59% vs. 7.1% p=0.02); and heart involvement (47% vs. 10.7% p=0.02).
After multivariate analysis the only probable independent predictor for early retirement due to SSc was lung involvement (OR: 13.98; 95% CI: 0.95-205.5).
Conclusion: In our cohort, SSc itself is frequently responsible for early retirement, being responsible for the loss of an average of 10 years of work. Pulmonary involvement is a probable predictor for early retirement associated with SSc. Further studies are warranted with a larger and more representative study population.

P0292 – NEW BIOLOGIC AND TARGETED SYNTHETIC DISEASE MODIFYING ANTIRHEUMATIC DRUGS FOR THE TREATMENT OF PSORIATIC ARTHRITIS. HOW ARE THEY POSITIONED IN CLINICAL PRACTICE?

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Introduction: Psoriatic arthritis (PsA) is a chronic heterogeneous disease that affects a variety of distinct anatomical sites including peripheral and axial joints and enthesis, skin and nails. Appropriate management of PsA requires early diagnosis, tight control of disease activity, and optimal use of available therapies, aiming at remission. The treatment options for PsA have recently broaden, including new biologic disease modifying antirheumatic drugs (bDMARDs) blocking the IL-23/IL-17 pathway and targeted synthetic (ts) DMARDs (Janus tyrosine kinase inhibitors- JAKi) with distinct mechanisms of action. The positioning of these newer therapies in PsA therapeutic algorithm is still debatable. In Portugal, ustekinumab-UST (anti -IL-12/23 p40), secukinumab-SEC (anti-IL-17A), and tofacitinib-TOFA (JAKi) are now approved and reimbursed for the treatment of PsA patients.
Objective: To describe the positioning (therapeutic line) of new bDMARDs and tsDMARDs for PsA patients in daily clinical practice, and to evaluate their effectiveness at 12 months of follow-up.
Methodology: PsA patients, registered at the Rheumatic Diseases Portuguese Register (Reuma.pt) treated with UST, SEC and TOFA were identified in three Portuguese Rheumatology Centres. Demographic (baseline) and disease activity parameters (baseline and first 12 months of treatment) were assessed.
Results: 60 patients (19 male and 41 female) (17 bDMARDs naïve and 43 bDMARDs experienced) with a mean (±SD) age of 47±12.1 years and a mean disease duration of 12.9±11.9 years were included. The most common subtype of PsA in this population was symmetric polyarthritis (41/60).
Forty patients received SEC, 18 UST and two TOFA. In 17/60 patients b/tsDMARDs were prescribed as the first therapeutic line after conventional DMARD failure (7 for UST and 10 for SEC); 11/60 as second line, 11/60 as third line and 21/60 as fourth or later therapeutic lines. One patient was under TOFA after 2 switches and the other one after 4 switches. Evaluating patients that were bDMARDs experienced, all received a tumor necrosis factor inhibitor (TNFi) as first line of therapy, with the exception of one patient under SEC that had been previously treated with UST. At baseline the mean (SD) DAPSA was 24.7 (±13.1) and DAS 28-4V 4.85±2.3. After 12 months of follow-up DAPSA and DAS28 4v decreased to 11.2 (±4.31) and 3.17±1.73, respectively.
Conclusion: In this PsA population, new b/tsDMARDs

TABLE 1 – BASELINE CHARACTERISTICS OF PSA PATIENTS TREATED WITH NEW BIOLOGIC AND TARGETED SYNTHETIC DISEASE MODIFYING ANTIRHEUMATIC DRUGS

Age (years)* mean (SD)	47±12.1
Male gender, n (%)	19 (31.7)
Disease duration (years±SD)	12.9±11.9
PsA subtypes	
Distal interphalangeal joint-predominant arthritis, n (%)	4 (6.7)
Symmetric polyarthritis-predominant arthritis, n (%)	41 (68.3)
Asymmetric oligoarthritis or monoarthritis, n(%)	11 (18.3)
Axial disease predominant spondylitis and/or sacroiliitis, n(%)	4 (6.7)
Arthritis mutilans, n (%)	0
PsA Disease activity	
DAPSA, mean (SD)	24.7 (±13.2)
DAS28, mean (SD)	4.85 (±2.3)
Treated with secukinumab, n (%)	40 (66.7)
Treated with ustekinumab, n (%)	18 (30)
Treated with tofacitinib, n(%)	2 (3.3)
1st line SEC or UST (bio-naives), n, (%)	17 (28.3)
Previous bDMARDs (bio-experienced), n (%)	43 (71.3)
2nd line (1 switch), n (%)	11 (18.3)
3rd line (2 switchs), n (%)	11 (18.3)
≥ 4th line (≥ 3 switchs), n (%)	21 (35)

n: number; PsA – Psoriatic Arthritis; DAPSA – Disease Activity in Psoriatic Arthritis; DAS28 4v – Disease Activity Score 28 joints; SEC- secukinumab, SD – Standard Deviation, UST- ustekinumab:

(UST/ SEC/TOFA) were prescribed after anti-TNF bDMARDs failure in 2/3 of the cases. Evidence of effectiveness after one year of treatment is provided inclusively after failure of several bDMARDs.

P0294 – CONSULTA MULTIDISCIPLINAR DE REUMATOLOGIA/OFTALMOLOGIA DO CENTRO HOSPITALAR LISBOA OCIDENTAL: CARACTERIZAÇÃO E IMPACTO NA POPULAÇÃO-ALVO

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Introdução e **objetivos:** A Consulta Multidisciplinar (CMD) de Reumatologia/Oftalmologia do Centro Hospitalar Lisboa Ocidental (CHLO) destina-se a doentes com patologia do foro reumatológico e/ou ocular que beneficiam da abordagem integrada de ambas as especialidades. Pretende-se caracterizar a população referenciada à CMD e analisar o seu impacto no diagnóstico e na terapêutica dos doentes.
Métodos: Análise retrospetiva dos doentes que estiveram presentes na CMD Reumatologia/Oftalmologia do CHLO, no período de Janeiro de 2017 a Dezembro de 2018 (24 meses). Foram registados os dados epidemiológicos, a origem e motivo da referenciação, os sinais e sintomas oculares, reumatológicos e outros no momento da consulta. Foram ainda recolhidos e quantificados os diagnósticos estabelecidos/presuntivos e as terapêuticas prescritas.
Resultados: Durante o período de tempo analisado, foram avaliados 72 doentes que estiveram presentes na CMD, pelo menos 1 vez. O número médio de consultas por doente foi 2,21 (±1,83). A média de idades observada foi 51,8±16,8 anos, 71% eram do sexo feminino. 52,8% dos doentes foram referenciados pela Reumatologia, 44,4% pela Oftalmologia, 1,4% pela Dermatologia e 1,4% pelo Centro de Saúde. A manifestação ocular mais frequente foi uveíte anterior aguda (52,8%), seguida de olho seco (12,5%), edema macular cistoide (6,9%), diminuição da acuidade visual (5,6%), episclerite (4,2%) e panuveíte (4,2%). Cerca de 8,3% dos doentes não apresentavam queixas oculares. Relativamente a manifestações osteoarticulares, 25,0% dos doentes referia lombalgia, 12,5% gonalgia e 12,5% artralgias das pequenas articulações mãos e/ou pés. 37,5% dos doentes não apresentavam manifestações osteoarticulares. Dos restantes sintomas, os mais frequentes foram xerostomia (9,7%), aftas orais/genitais recorrentes (8,4%), dermatológicos (8,3%) e gastrointestinais (4,2%). Os diagnósticos mais comuns foram Espondilartrite axial radiográfica (26,4%) e uveíte anterior associada ao HLA-B27 (18,1%). 19,4% do total de diagnósticos foi realizado em contexto de CMD, e 21,4% destes correspondem a diagnósticos reformulados a partir de um diagnóstico prévio. Outros diagnósticos relevantes incluem Artrite Reumatoide (6,9%), Síndrome de Sjögren (4,6%) e Doença de Behçet (4,2%). No período em estudo, apenas 15,3% dos casos foram considerados idiopáticos. Quanto às terapêuticas sistémicas mais frequentes, verificou-se que em 37,5% dos casos estava prescrita corticoterapia oral, em 69,4% DMARDs convencionais (27,8% sulfassalazina, 26,4% metotrexato e 5,6% azatioprina) e em 13,9% DMARDs biológicos (9,7% adalimumab, 4,2% etanercept e 1,4% tocilizumab). Por fim, 8,3% dos doentes iniciaram terapêutica biológica, 4,2% já tinham iniciado previamente e em 1,4% alterou-se o biológico que estava prescrito.
Conclusão: A CMD Reumatologia/Oftalmologia do CHLO torna possível a abordagem simultânea e sinérgica dos doentes, tendo um impacto fundamental no estabelecimento precoce do diagnóstico e da terapêutica. Será relevante avaliar a proporção de doentes em baixa atividade/remissão, o impacto em termos de custos e os níveis de satisfação dos doentes, comparativamente com os cuidados tradicionais.

P0295 – HOW TO IMPROVE EARLY DIAGNOSIS OF GIANT CELL ARTERITIS? A RETROSPECTIVE COHORT STUDY

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Background: Giant cell arteritis (GCA) is the most common form of primary systemic vasculitis, typically affecting patients aged > 50 years. If left untreated, GCA can lead to permanent visual loss and other ischaemic complications. Therefore, early diagnosis and effective treatment initiation are of utter importance.
Objective: To determine factors associated with early versus late diagnosis in GCA.

Methods: Retrospective cohort study in patients with GCA fulfilling ACR'90 criteria followed in a tertiary centre. Sociodemographic and clinical data were collected from medical records and the Portuguese Register of Rheumatic Diseases (Reuma.pt). Early and late diagnosis were defined as time from clinical onset until diagnosis ≤14 days and >14 days, respectively. Age was categorised as younger and older if the patient was <70 or ≥70 years old. Univariate analysis was performed using Chi-Square and Mann-Whitney Test, as appropriate. Logistic multivariate analysis including variables with p<0.01 in univariate analysis was planned.
Results: A total of 21 patients (female: 66%, mean age at diagnosis: 73±10.42 years) were included in the analysis, from which 14/17 had a compatible Doppler-ultrasonography and 14/18 biopsy-proven arteritis. At the time of diagnosis, all patients were >50 years old, had new onset headache and presented with an erythrocyte sedimentation rate (ESR) > 50 mm/1st hour. Most patients (76%) had temporal artery abnormalities on physical examination, including local inflammatory signs (52.4%), tender palpation (76%) and reduced pulsation (57%).
The median time from clinical onset to diagnosis was 14 days (IQR=21). Twelve patients (57.1%) and 9 patients (42.9%) were included in the early and in the late diagnosis group, respectively. There were no statistically significant differences between the groups regarding gender, age category, manifestations of polymyalgia rheumatica or visual disturbances. There were no differences regarding median age or ESR values between the groups.
Conclusions. We could not identify any factors associated with earlier diagnosis of GCA in this cohort. Interestingly, potential red flags for the hypothesis of GCA, such as polymyalgia rheumatic or visual abnormalities were not found to be associated with earlier diagnosis. Future directions are to perform a multicentre study with a larger and more representative population.

P0304 – IDENTIFICATION OF KEY GENES TO SUPPORT SYSTEMIC LUPUS ERYTHEMATOSUS, RHEUMATOID ARTHRITIS AND ANKYLOSING SPONDYLITIS DIAGNOSIS BY TRANSCRIPTOMIC APPROACH

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Background: Early diagnosis of inflammatory rheumatic diseases (IRD), as Systemic Lupus Erythematosus (SLE), Rheumatoid Arthritis (RA) and axial Spondyloarthritis (axSpA) represents in our days a major clinical challenge. Increasing evidence has determined that early diagnosis, prompt treatment initiation and early achievement of remission are the best predictors of long-term clinical, functional and radiographic outcomes. Therefore, identification of sensitive biomarkers to support an early diagnosis to enable early therapy is of utmost importance [1,2].
Objectives: This study aims to identify novel genes that may improve the current clinical diagnosis approach for early SLE, RA and axSpA.
Methods: A cross-sectional study was conducted on 44 participants, 12 with axSpA (according to ASAS criteria), 11 with RA (according to ACR/EULAR criteria for RA), 10 with SLE (according to ACR classification criteria for SLE) and 11 Healthy Controls (HC), gender and age matched. Patients with co-occurrence of other IRD or having received biological therapies were excluded. Peripheral blood samples were collected into PAXgene tubes and stored in -80°C. mRNA profiling by RNA-seq was performed. Unpaired t-tests with multivariate permutation correction were applied to identify differentially expressed genes (DEGs) between patients and HC for each disease and within diseases. Enrichment analysis, Gene ontology (GO) and Kyoto Enrichment of Genes and Genomes (KEGG) analysis were also performed. DEGs that allow to distinguish each disease from HC and between diseases. The top DEGs (axSpA n=2, RA n=2, SLE n=3) identified were confirmed by quantitative RT-PCR.
Results: For axSpA, genes involved in negative regulation of cytokines by JAK/STAT pathway and in osteoblast differentiation through STAT3 pathway, were confirmed. In SLE, genes involved in trap for immune complexes in peripheral blood and involved in nucleosome regulation, were also confirmed. Regarding RA, no genes were confirmed.
Conclusion: Our work provides new insights into

IRD pathogenesis, and discloses new biomarkers, which may be useful as either predictive biomarkers for diagnosis or therapeutic targets to improve IRD approach.Further validation are needed in different cohorts.

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P0305 – AS APLICAÇÕES DA TOXINA BOTULÍNICA NAS DOENÇAS REUMÁTICAS

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Introdução: A Toxina Botulínica (Btx) é produzida pela bactéria anaeróbia Clostridium botulinum, a sua alta toxicidade aliada a mecanismos de ação extremamente especificos confere-lhe características únicas e de alta utilidade em diversas patologias (oftalmológicas, urológicas, ortopédicas, dermatológicas, neurológicas, etc).
A Btx ao inibir a libertação de uma grande variedade de neurotransmissores das vesículas pré-sinápticas (acetilcolina, norepinefrina, substância P, PRGC) apresenta um mecanismo de ação vasto (ação antinocicetiva, atua no SNA e no SNC).
Com este trabalho pretendemos demonstrar a eficácia da Btx nas doenças reumáticas.
Métodos: Foi feita uma revisão sistemática da literatura para selecionar os estudos de casos, ensaios clínicos e revisões. Foram utilizadas as bases de dados da Pubmed e Cochrane Library entre 2005 e 2019 com as seguintes keywords: botulinum toxin, chronic pain, osteoarthritis, systemic sclerosis, reumatology.
Foram considerados para esta pesquisa um total de 13 artigos, dos quais 9 revisões, 1 ensaio clínico e 3 estudos de série de casos. Foram incluídos os artigos que estudassem a eficácia da aplicação da Btx (intervenção) em adultos com doença reumatológica (população), comparando com outras terapêuticas ou com placebo e relatando benefícios ou malefícios (resultados).
Resultados: Os estudos de maior dimensão e artigos de revisão encontrados demonstram a eficácia da Btx nas doenças reumatológicas associadas a algumas síndromes de dor crónica refratárias ao tratamento farmacológico e/ou de reabilitação. Na epicondilite lateral a eficácia da Btx tipo A (Btx-A) demonstra um nível A de evidência. A eficácia da Btx-A na fasceíte plantar, síndrome pirifome e lombalgia crónica demonstra um nível B de evidência. Na gonartrose associada a gonalgia a infiltração com Btx-A também se mostrou eficaz, com melhoria do quadro álgico, contudo quando comparado ao grupo submetido à infiltração com triancinolona, não se verificaram diferença estatisticamente significativas.
Nas síndromes miofasciais a aplicação de Btx-A demonstrou resultados contraditórios.
Mais recentemente têm sido publicados estudos sobre a eficácia da Btx no Fenómeno de Raynaud grave (FR) associado ou não a úlceras digitais (com ou sem esclerose sistémica). São estudos de menor dimensão, na maioria é utilizada a Btx-A, contudo num estudo é referida também a eficácia da Btx-B. As doses de Btx-A infiltradas variaram de 40 a 100 Unidades por mão e o local de infiltração foi na maioria dos estudos na face palmar e mais recentemente na face dorsal. Os resultados obtidos foram uma melhoria do FR, com diminuição da frequência, duração e sintomatologia associada, bem como a completa cicatrização das úlceras digitais.
Discussão e Conclusão: As doenças reumatológicas estão muitas vezes associadas a síndromes de dor crónica e nestes casos a Btx pode desempenhar um papel fundamental. O nível A de evidência só está demonstrado na epicondilite lateral, sendo que o efeito adverso mais frequente é a diminuição da força muscular na extensão dos dedos.
A utilização da Btx também se mostra eficaz no FR grave (com ou sem úlceras digitais), o seu mecanismo de ação ainda não está completamente esclarecido, mas pensa-se que se deva ao bloqueio simpático, com vasodilatação e aumento do fluxo sanguíneo
Em suma, as aplicações da Btx continuam a aumentar (recomendações baseadas na evidência), sendo que mais estudos são necessários para reforçar esta terapêutica promissora nas doenças reumatológicas.

P0306 – ANXIETY AND DEPRESSION IN PATIENTS WITH GIANT CELL ARTERITIS

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Background: Giant cell arteritis (GCA) is the most common primary vasculitis of the elderly causing blindness if left untreated. However, high doses of glucocorticoids (GCs) can lead to significant toxicity, including psychiatric manifestations. Few studies have investigated the association between these symptoms and GCA, mainly using Short Form 36 (SF-36), a generic patient-reported outcome (PRO) with a mental component summary. Hospital Anxiety and Depression Scale (HADS) is a validated PRO to assess depression and anxiety and to the best of our knowledge has never been evaluated in patients with GCA.

Objectives: To compare the prevalence of anxiety and depression in GCA with the general population using HADS. To explore GCA-specific contributive factors to mental status.

Methods: HADS and SF-36 questionnaires were prospectively collected from patients with biopsy- or imaging-proven GCA evaluated from July 2018 to January 2020 in the Vasculitis clinic of a tertiary centre. A cross-sectional analysis using data registered at the Rheumatic Diseases Portuguese Register (Reuma.pt) was performed. HADS results from an age- and gender-matched control group were retrieved from healthy individuals included in the EpiReumaPT study (the largest Portuguese epidemiologic study on rheumatic diseases). HADS-A and HADS-D ≥8 defined possible and HADS-A and HADS-D ≥11 defined probable anxiety and depression, respectively. Clinical features of GCA patients with HADS ≥8 or <8 were compared using T-student, Mann-Whitney, Chi-square, and Fisher's exact tests, as appropriate; association between continuous variables was assessed using Spearman's correlation coefficient; and binary logistic regression was used to identify independent predictors of HADS ≥8.

Results: We included 72 patients diagnosed with GCA, 52 (72.2%) females, with a mean ± SD age of 78.3 ± 7.7 years. The matched-control group consisted of 288 individuals. Patients with GCA had higher median [IQR] HADS-A than controls (7 [7] vs. 5 [5], p<0.001), as well as a higher prevalence of HADS-A ≥8

TABLE 1 – BASELINE CHARACTERISTICS OF PSA PATIENTS TREATED WITH NEW BIOLOGIC AND TARGETED SYNTHETIC DISEASE MODIFYING ANTIRHEUMATIC DRUGS

	HADS-A <8 (N=37)	HADS-A ≥8 (N=35)	*p-value*	HADS-D <8 (N=37)	HADS-D ≥8 (N=35)	*p-value*
Demographic data						
Mean age, mean ± SD	76.8 ± 7.3	79.9 ± 8.0	*p=0.093*	74.9 ± 7.4	81.9 ± 6.4	***p<0.001***
Female sex, n (%)	27 (73.0)	25 (71.4)	*p=0.884*	26 (70.3)	26 (74.3)	*p=0.704*
Clinical characteristics of the disease						
Ocular involvement, n (%)	17 (46.0)	12 (34.3)	*p=0.313*	15 (40.5)	14 (40.0)	*p=0.963*
LV involvement by imaging, n (%)	11 (29.7)	10 (28.6)	*p=0.914*	12 (32.4)	9 (25.7)	*p=0.531*
Disease duration, median (IQR) years	1.0 (4.2)	2.9 (4.91)	*p=0.383*	1.0 (4.0)	3.1 (6.5)	*p=0.481*
Disease duration > 3 months, n (%)	28 (75.7)	29 (82.9)	*p=0.453*	29 (78.4)	28 (80.0)	*P=0.866*
Disease duration > 1 year, n (%)	22 (59.5)	25 (71.4)	*p=0.286*	22 (59.5)	25 (71.4)	*p=0.286*
Comorbidities, n (%)						
Atherosclerosis	0 (0)	3 (8.6)	*p=0.110*	1 (2.7)	2 (5.7)	*p=0.609*
Atrial fibrillation	1 (2.7)	1 (2.9)	*p=1.000*	0 (0.0)	2 (5.7)	*p=0.233*
Cerebrovascular disease	2 (5.4)	4 (11.4)	*p=0.423*	1 (2.7)	5 (14.3)	*p=0.102*
Chronic renal disease	3 (8.1)	1 (2.9)	*p=0.615*	3 (8.1)	1 (2.9)	*p=0.615*
Diabetes mellitus	15 (40.5)	6 (17.1)	***p=0.029***	12 (32.4)	9 (25.7)	*p=0.531*
Hypercholesterolemia	5 (13.5)	4 (11.4)	*p=1.000*	5 (13.5)	4 (11.4)	*p=1.000*
Hypertension	26 (70.3)	23 (65.7)	*p=0.679*	25 (67.6)	24 (68.6)	*p=0.927*
Hyperuricemia/ gout	2 (5.4)	0 (0)	*p= 0.493*	1 (2.7)	1 (2.9)	*p= 1.000*
Ischaemic cardiac disease	1 (2.7)	3 (8.6)	*p=0.350*	1 (2.7)	3 (8.6)	*p=0.350*
Mourning	1 (2.7)	5 (14.7)	*p=0.098*	2 (5.6)	4 (11.4)	*p=0.429*
Neoplastic disease	5 (13.5)	8 (22.9)	*p=0.303*	5 (13.5)	8 (22.9)	*p=0.303*
Obesity	0 (0)	1 (2.9)	*p=0.486*	0 (0.0)	1 (2.9)	*p=0.486*
Peripheral arterial disease	1 (2.7)	2 (5.7)	*p=0.609*	0 (0.0)	3 (8.6)	*p=0.110*
Previous history of mental disease	2 (5.4)	2 (5.7)	*p=1.000*	1 (2.7)	3 (8.6)	*p=0.350*
Thyroid disease	3 (8.1)	2 (5.7)	*p=1.000*	3 (8.1)	2 (5.7)	*p=1.000*
Laboratory results, median (IQR)						
C-reactive protein, mg/dL	0.35 (0.5)	0.46 (1.2)	*p=0.556*	0.40 (0.5)	0.36 (0.9)	*p=0.581*
Erythrocyte sedimentation rate, mm/h	25 (20.0)	36 (33.0)	***p=0.027***	25.5 (26.5)	35 (43.5)	*p=0.318*
GC treatment at the time of questionnaires						
Patients under GCs, n (%)	29 (78.4)	34 (97.1)	***p=0.028***	29 (78.4)	34 (97.1)	***p=0.028***
Current GC dose, median (IQR) mg *	10 (42.5)	7.5 (42.5)	*p=0.994*	18.8 (42.5)	7.5 (42.5)	*p=0.827*
Patients under GCs >30mg, n (%) *	11 (37.9)	10 (31.3)	*p=0.583*	11 (39.3)	10 (30.3)	*p=0.462*
Treatment with GCs ≥1year, n (%)	16 (53.3)	24 (70.6)	*p=0.155*	16 (55.2)	24 (68.6)	*p=0.270*
Short-Form 36 components, median (IQR)						
SF-36 Physical function	55.0 (52.5)	25.0 (55.0)	***p=0.003***	70.0 (50.0)	20.0 (50.0)	***p<0.001***
SF-36 Role limitation (physical)	50.0 (75.0)	25.0 (50.0)	*p=0.103*	50.0 (59.4)	18.8 (31.2)	*p=0.001*
SF-36 Role limitation (emotional)	58.3 (75.0)	25.0 (66.7)	***p=0.006***	66.7 (66.7)	25.0 (41.7)	***p<0.001***
SF-36 Social function	87.5 (50.0)	50.0 (37.5)	***p<0.001***	87.5 (37.5)	50.0 (25.0)	***p<0.001***
SF-36 Pain	62.0 (43.0)	31.0 (29.0)	***p<0.001***	62.0 (43.0)	31.0 (40.0)	***p<0.001***
SF-36 Vitality	56.3 (42.5)	25.0 (33.8)	***p<0.001***	56.2 (32.5)	25.0 (27.5)	***p<0.001***
SF-36 Mental Health	80 (26.5)	40 (26.4)	***p<0.001***	76 (32.4)	40 (32.0)	***p<0.001***
SF-36 General Health	50.0 (27.5)	30.0 (20.0)	***p<0.001***	52.0 (30.0)	30.0 (20.0)	***p<0.001***

In bold statistically significant differences (p<0.05) * Prednisolone equivalent. GCs – glucocorticoids; HADS – Hospital Anxiety (A) and Depression (D) Scale; IQR – interquartile range; LV – large vessel; SF-36 – Short Form-36.

FIGURE 1 – LOGISTIC REGRESSIONS PREDICTING THE LIKELIHOOD OF HADS-A >8 AND HADS-D >8, BASED ON AGE, SEX, DISEASE DURATION AND CURRENT GC TREATMENT

HADS-A ≥8

	B	S.E.	Wald	df	p	OR	95% CI for OR Inferior	Superior
Sex	-0,150	0,581	0,067	1	0,796	0,860	0,276	2,685
Age at time of questionnaire	0,040	0,035	1,346	1	0,246	1,041	0,973	1,114
Disease duration >1year	0,772	0,533	2,099	1	0,147	2,164	0,762	6,147
GCs	2,342	1,124	4,341	1	**0,037**	**10,406**	1,149	94,230
Constant	-5,701	2,888	3,896	1	0,048	0,003		

HADS-D ≥8

	B	S.E.	Wald	df	p	OR	95% CI for OR Inferior	Superior
Sex	0,598	0,651	0,843	1	0,358	1,818	0,508	6,508
Age at time of questionnaire	0,183	0,054	11,236	1	**0,001**	**1,200**	1,079	1,336
Disease duration >1year	1,096	0,615	3,176	1	0,075	2,994	0,896	9,998
GCs	2,133	1,138	3,515	1	0,061	**8,442**	0,908	78,496
Constant	-17,372	4,626	14,105	1	0,000	0,000		

B – coefficients in log-odds units; CI – confidence interval; df – degrees of freedom; OR – odds ratio, p – Wald 2-tailed p-value; SE – standard errors associated with the coefficients

and HADS-A ≥11 (48.6% vs. 26.8%, p<0.001; 30.6% vs. 12.2%, p<0.001; respectively). Patients with GCA did not differ from controls in terms of median [IQR] HADS-D (7 [9] vs. 5 [6], p=0.130) or prevalence of HADS-D ≥8 (48.6% vs. 37.2%, p=0.075), but had more cases of HADS-D ≥11 (33.3% vs. 18.1%, p=0.004). Table 1 shows the differences between GCA patients with HADS ≥8 and <8. Patients with HADS ≥8 were more frequently under GC treatment (all cases of HADS ≥11 were on GCs). Patients had inferior levels of SF-36 in all categories except for physical role limitation and a correlation between values of HADS and SF-36 mental health was observed (HADS-A: r=-0.780, HADS-D: r=-0.742; both p<0.001). Patients with HADS-A ≥8 had higher levels of ESR and lower prevalence of diabetes mellitus, whereas HADS-D ≥8 was associated with older age. Figure 1 shows the logistic regression models for HADS. Treatment with GCs was identified as an independent predictor of anxiety and older age as an independent predictor of depression.

Conclusions: HADS appeared to be an efficient screening tool for depression and anxiety in GCA, correlating well with SF-36. Patients with GCA had more anxiety than general population but depression was only increased when using HADS-D ≥11 as the cut-off. GC treatment and older age were identified as independent predictors of anxiety and depression, respectively. Although these results require replication, they raise awareness for the fact that mental health should not be overlooked when managing GCA.

P0317 – RESULTADOS NA GRAVIDEZ E CERTOLIZUMAB PEGOL NA ARTRITE REUMATÓIDE: UMA SÉRIE DE CASOS

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Introdução: As doenças reumáticas geralmente afectam mulheres em idade fértil, muitas vezes quando contemplam a gravidez ou esta não foi planeada. A preparação e o controlo adequado da doença são factores cruciais para uma gravidez bem sucedida, o que tem levado a uma crescente consciencialização dos reumatologistas para estas questões particulares.

Objectivos: Descrever uma série de casos de duas doentes com artrite reumatóide (AR) sob certolizumab pegol (CZP) durante a gravidez.

Materiais e Métodos: Duas doentes com AR, que cumprem os critérios de classificação ACR/EULAR 2010, são seguidas no Serviço de Reumatologia da Unidade Local de Saúde da Guarda. Os processos clínicos foram revistos retrospectivamente e as características clínicas reumáticas e obstétricas foram colhidas.

Resultados: O Caso 1 é uma médica de 33 anos, com o diagnóstico de AR não erosiva seronegativa em 2017. Os sintomas iniciaram-se três anos antes, envolvendo as grandes articulações, mas com atingimento das pequenas articulações após a primeira gravidez e consequente diagnóstico. Iniciou metotrexato numa estratégia step-up de acordo com a actividade da doença e, rapidamente, escalou-se para terapêutica tripla com metotrexato, sulfassalazina e hidroxicloroquina. A doente alcançou baixa actividade da doença, mas mantinha dificuldade nas actividades relacionadas com o trabalho. Em 2018, a doente referiu o desejo de voltar a engravidar, com necessidade de suspender o metotrexato. De forma a manter a baixa actividade da doença, foi iniciado CZP em dose de indução de 400 mg nas semanas 0, 2 e 4 e dose de manutenção de 200 mg a cada 2 semanas. A sulfassalazina e a hidroxicloroquina foram suspensas e a corticoterapia desmamada, assim que se alcançou a remissão.

O Caso 2 é também uma mulher de 33 anos, com um cargo administrativo, diagnosticada com AR seropositiva e erosiva em 2012. A doente referia sintomas há seis anos, mas foi tardiamente referenciada para a consulta de Reumatologia. Nessa altura, já estaria a fazer corticoterapia há largos períodos e apresentava estigmas de síndrome de Cushing, bem como uma poliartrite grave com alta actividade da doença. Assim, iniciou metotrexato numa estratégia step-up de acordo com a actividade da doença, necessitando rapidamente de terapêutica tripla também. Infelizmente, manteve uma actividade moderada da doença e foi-lhe proposto iniciar um bDMARD, que a doente recusou. Suspendeu também, autonomamente, o metotrexato, uma vez que planeava engravidar. Sem o tratamento eficaz houve agravamento dos sintomas e aumento dos parâmetros inflamatórios, não tendo conseguido engravidar durante mais de um ano, já que a doença se encontrava em alta actividade. Foi assim proposta para CZP, que a doente aceitou. Foi utilizado o mesmo esquema do Caso 1.

Não foi observado nenhum aborto prévio. Não foi descrita nenhuma intercorrência durante ambas as gestações ou necessidade de suspender o CZP durante esse período. No Caso 1 o parto foi cesariana por distocia cervical e no Caso 2 o parto foi eutócico. Nenhum dos partos foi prematuro (38S4D no Caso 1 e 39S2D no Caso 2), e nenhuma malformação congénita foi detectada pelos neonatologistas na avaliação inicial. Ambas

as doentes iniciaram amamentação sob CZP.
Conclusão: Este trabalho mostra a importância do controlo da doença em todas as fases, seja antes da concepção ou durante a gravidez, para que não ocorram intercorrências. A nossa experiência, embora breve, corrobora os dados existentes de que o CZP parece ser uma escolha segura no tratamento da artrite reumatóide em mulheres em idade fértil.

P0320 – RITUXIMAB OFF-LABEL: EXPERIÊNCIA DE UM SERVIÇO DE REUMATOLOGIA

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Introdução: O Rituximab (RTX) é um anticorpo monoclonal IgG1 direcionado ao antigénio CD20, que promove a depleção de células B. Está aprovado atualmente para linfoma não-Hodgkin de células B, leucemia linfocítica crónica, artrite reumatoide (AR), vasculites associadas aos ANCA e pênfigo vulgar. Na Reumatologia, tem sido utilizado off-label em múltiplas patologias, frequentemente em casos desafiantes do ponto de vista terapêutico.
Métodos: Estudo retrospetivo longitudinal dos doentes com doença reumática sistémica (DRS) seguidos no Hospital de Dia do Hospital de Egas Moniz, entre 2015 e 2019, sob terapêutica com RTX off-label. Calcularam-se os dados de resposta terapêutica na baseline (S0) e às 24 semanas (S24) de acordo com a patologia e envolvimento clínico: Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), EULAR Sjogren Syndrome Disease Activity Index (ESSDAI), Disease Activity Score 28 articulações e 3 variáveis (velocidade de sedimentação) (DAS28(3V)). O restante envolvimento foi monitorizado clinicamente. Foram aplicados os protocolos de RTX 1000mg (dia 1 e dia 15) nos doentes com envolvimento articular e o protocolo RTX 375mg/m2 de superfície corporal/semana (4 semanas) nos doentes com o restante envolvimento.
Resultados: Dos 53 doentes sob RTX, 14 fizeram o fármaco off-label (93% género feminino; idade média 48 anos; mediana de doença 6 anos), 93% por doença ativa/refratária e 7% por iatrogenia a terapêutica prévia. Na amostra de 14 doentes: 4 Lúpus Eritematoso Sistémico (LES) por envolvimento articular, hematológico, renal, pulmonar, mucocutâneo, neurológico, vascular e ganglionar; 1 Síndrome dos Anticorpos Antifosfolipídicos (SAAF) catastrófico; 1 Síndrome de Sjogren primário (SS) por envolvimento articular e mucocutâneo; 1 Doença Mista do Tecido Conjuntivo (DMTC) por envolvimento pulmonar, 1 Doença Indiferenciada do Tecido Conjuntivo (DITC) por envolvimento articular, 1 Microangiopatia Trombótica/Síndrome Hemolítico Urémico atípico (MT/SHUa) por envolvimento renal e vascular; 5 síndromes de sobreposição (AR/LES – RHUPUS; LES/Dermatomiosite; Esclerose Sistémica/SS) por envolvimento articular, renal, hematológico, mucocutâneo, neurológico e vascular. Terapêutica modificadora da doença reumática (DMARD) pré-RTX na baseline por fármaco e dose média (m): 3 sob azatioprina 150mg/dia; 4 sob metotrexato 20mg/semana; 4 sob hidroxicloroquina 400mg/dia; 2 naïves a terapêutica. Nos doentes sob prednisolona (PDN) registou-se PDNm de 13mg/dia para envolvimento articular e 23mg/dia para envolvimento sistémico. Variação da atividade da DRS em S0 e S24 por índice de atividade: LES – SLEDAI S0: 9 e S24: 3, DAS28(3V) S0: 4.27 e S24: 2.46; SS – ESSDAI S0: 2 e S24: 1.3; DAS28(3V) S0: 6.38 e S24: 5.58; DITC – DAS28(3V) S0: 3.13 e S24: 2.8. Variação da atividade da DRS por evolução analítica: MT/SHUa – creatinina S0: 0.9mg/dL e S24: 0.86mg/dL; proteinúria S0: 149.8mg/dL e S24: 293.6mg/dL. Após RTX registou-se PDNm de 14mg/dia nos doentes com envolvimento articular e PDNm 16mg/dia nos doentes com envolvimento sistémico. Efeitos adversos reportados: hipogamaglobulinemia secundária (N=2) com necessidade de imunoglobulina endovenosa (1 dos quais com abandono terapêutico); prurido pós-perfusão (N=1).
Conclusão: Nesta população o RTX permitiu a redução de corticoterapia nos doentes com envolvimento sistémico e remissão articular nos restantes, mostrando-se uma alternativa terapêutica off-label no controlo das DRS.

P0331 – TENOSSINOVITE DE QUERVAIN, A PERSPETIVA MINIMAMENTE INVASIVA DO MÉDICO FISIATRA

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Introdução: A Tenossinovite de Quervain é caracterizada pelo espessamento, edema e muitas vezes fluido ao nível da baínha do 1º compartimento extensor do

punho. Sobrecarga repetitiva e contínua dos tendões do abdutor longo e extensor curto do polegar com ou sem traumatismo associado, condiciona um quadro inflamatório ao nível desse mesmo compartimento. Esta condição é mais frequente no género feminino e em idades laborais (30 a 50 anos). Existe uma panóplia de possibilidades de tratamento, desde o tratamento fisiátrico clássico à cirurgia com libertação do compartimento. Entre estes dois extremos existe a possibilidade de terapêuticas minimamente invasivas com o objetivo de controlar e mesmo normalizar o processo inflamatório. Associada à utilização de um guia ecográfico, a infiltração córtico-anestésica torna-se uma técnica cada vez mais segura e eficaz.

Objetivos: O objetivo deste trabalho visa demonstrar a eficácia da infiltração cortico-anestésica do 1º compartimento extensor do punho, guiada por ecografia, numa série de casos de Tenossinovite de Quervain avaliados e intervencionados no Centro de Reabilitação do Norte (CRN).

Materiais e Métodos: Foram identificados todos os doentes, com Tenossinovite do 1º Compartimento Extensor, referenciados para o Centro de Reabilitação para consulta de Fisiatria de Intervenção, no período assistencial do ano de 2019. Foi analisada a eficácia da intervenção (Infiltração Ecoguiada do 1º compartimento extensor do punho) nesta patologia, tendo em conta a diminuição da dor pela escala visual numérica (EVN) em diferentes tempos de consulta e a necessidade de recorrer a mediação SOS no período pós intervenção.

Resultados: Foram encaminhados para a consulta de Fisiatria de Intervenção do CRN 20 doentes com Tenossinovite de Quervain. A maioria era do género feminino (19) e a média de idade foi de 50,9 anos. Na avaliação clínica pré intervenção a EVN foi em média de 7,9 e cerca de 75% (15) realizavam medicação para a dor em SOS. Aos 2 meses pós-intervenção, a EVN média era 1,85 e apenas de 20% (4) realizavam medicação para a dor em SOS, sendo de destacar que 50% dos doentes ficaram totalmente assintomáticos após um único procedimento.

Conclusão: A Tenossinovite de Quervain é muito frequente na população ativa, sendo muitos casos encaminhados para a medicina física e reabilitação (MFR). Com a evolução do conhecimento e expertise em técnicas minimamente invasivas, o médico Fisiatra tem a possibilidade de associar ao tratamento convencional Técnicas diferenciadas com taxas de melhoria e até de cura muito interessantes. Com esta série de casos, verificou-se uma redução significativa na dor, sendo de destacar que metade dos pacientes ficaram totalmente assintomáticos e apenas 20% (4) mantiveram a necessidade de realizar medicação para a dor em SOS. As técnicas minimamente invasivas, situam-se na zona cinzenta entre o tratamento fisiátrico clássico e a cirurgia, fazendo parte da atividade assistencial em MFR, quer como abordagem inicial, potenciação dos programas de reabilitação preconizados inicialmente ou, em caso de falência destes, técnica de resgate terapêutico.

P0332 – LITERACIA PARA A SAÚDE NO DOENTE REUMÁTICO

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Introdução: O conceito de Literacia para a Saúde tem sido mencionado em diversos estudos, desempenhando um papel determinante na manutenção e melhoria das condições de saúde. Os serviços de saúde consideram-no extremamente importante, sobretudo, na área de cuidados de saúde primários e saúde pública. Um inadequado nível de Literacia para a Saúde pode ter implicações significativas na saúde individual/coletiva, podendo estar perante um contexto de desigualdades em saúde, com implicações na gestão de recursos e ganhos em saúde. Segundo a Sociedade Portuguesa de Reumatologia, atualmente as doenças reumáticas e músculo-esqueléticas têm um custo anual para o Estado superior a mil milhões de euros. O Governo Português, em 2016, estabeleceu como prioridade promover a saúde, criando o Programa Nacional para a Saúde, Literacia e Autocuidados.

Objetivos: Conhecer o nível de Literacia para a Saúde no doente reumático que recorre à consulta de reumatologia e identificar as áreas prioritárias de intervenção. Metodologia: Desenvolveu-se um estudo descritivo, transversal e quantitativo. A população alvo foram os utentes que recorreram à consulta de reumatologia. A amostra foi constituída por 109 utentes. Considerando como valor central de idade os 58 anos (x =58,3 e Md=61), aproximadamente, os elementos da amostra com mais de 61 anos teve uma frequência semelhante à dos que têm menos de 61 anos. A maioria (75.2%) era do sexo feminino. Para avaliar o nível de Literacia para a Saúde utilizou-se o Questionário Europeu de Literacia para a Saúde Health Literacy Survey in Portuguese (HLS-EU-PT), validado por Saboga-Nunes e Sørensen em 2013. Para o tratamento estatístico utilizou-se o programa de tratamento estatístico Statistical Package for the Social Science, versão 23 de 2016. As técnicas estatísticas aplicadas

foram frequências (absolutas e relativas), medidas de tendência central (média aritmética, média ordinal e mediana), medidas de dispersão ou variabilidade (valor mínimo, valor máximo e desvio padrão) e o coeficiente alpha de Cronbach.
Resultados: Apenas 17,5% da amostra apresentava um nível de Literacia para a Saúde geral suficiente ou excelente. Na amostra 82.6% tinha um nível de Literacia para a Saúde geral problemático ou inadequado. Nas dimensões cuidadas de saúde, prevenção da doença e promoção da saúde, observou-se a mesma tendência (83.5%, 80.7% e 83.5%, respetivamente). Na dimensão promoção da saúde observou-se uma percentagem mais elevada de pessoas com um nível de Literacia para a Saúde inadequado, 64.2%. Foi nos cuidados de saúde e promoção de saúde que os resultados foram mais preocupantes: 83.5% da amostra apresentava literacia limitada (inadequada/problemática).
Discussão/Conclusões: A amostra em estudo apresenta níveis de Literacia para a Saúde muito limitados, o que aponta para a necessidade dos profissionais de saúde, entidades de saúde, governo e sociedade em geral, investirem na educação da pessoa e comunidade, capacitando-os para a gestão da sua saúde, onde o enfermeiro especialista em enfermagem comunitária desempenha um papel primordial. Sugerem-se intervenções nas diferentes dimensões da Literacia para a Saúde, sobretudo na área de cuidados de saúde e promoção de saúde delineando estratégias/atividades que possibilitem mitigar a problemática identificada, procurando melhorar a qualidade da saúde da pessoa e comunidade, perspetivando-se implicações favoráveis na gestão de recursos e ganhos em saúde.
Palavras-Chave: Literacia para a Saúde; Enfermagem Comunitária; Reumatologia

P0345 – IMPACT OF ACTN3 AND VDR GENE POLYMORPHISMS ON STRENGTH, MUSCLE MASS AND MUSCULAR PERFORMANCE IN MUSCULOSKELETAL DISEASES – A SYSTEMATIC REVIEW

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Objectives: This systematic review aimed to identify genetic variants of ACTN3 and VDR genes associated with musculoskeletal diseases susceptibility, severity and progression; in addition, their potential impact in muscle properties such as muscle strength, muscle mass and muscle performance in this context.
Methods: A systematic literature review (SLR) was performed. The bibliographic databases, PubMed, Embase and Web of Science were systematically searched (from January 2000 to December 2019) using pre-defined search terms such as "polymorphism*", "actn3", "vdr", "genetic polymorphism", "muscle mass", "strength", "performance", "muscular", "exercise", "fitness", "physical performance", "muscle weakness", "obesity", "endurance", "muscle disease", "physical activity". The "Population(P)", "Intervention(I)", "Comparator(C)", "Outcome(O)" (PICO) criteria were used. "P", defined as musculoskeletal diseases "I" as snp of VDR and ACT3 genes, "C" as absence of studied snps and "O" musculoskeletal disease susceptibility, severity and progression and muscle properties.
Case control and prospective studies published in English, that examined the genetic variants association with musculoskeletal diseases and its muscle phenotypes, in subjects aged ≥ 18 years were included.
Results: Sixty studies were included in the final analysis. The VDR, FokI, ApaI, TaqI and BsmI, and the ACTN3 R577X are the most commonly studied polymorphisms.
Our results showed that ApaI, FokI, and TaqI snp are associated with susceptibility for systemic sclerosis, herniation and spinal tissue degeneration, and rheumatoid arthritis (RA) and osteoarthritis, respectively. FokI appearing to be a good biomarker for osteoporosis in women but seems to be the combination of TaqI recessive allele (t) with BsmI dominant (B) allele that would be associated with an increased bone loss on RA patients. On the other hand, ApaI and TaqI polymorphisms do not increase the susceptibility for Temporomandibular joint internal derangement/ osteoarthritis; the FokI is not correlated with fibromialgia.
The ApaI is associated with an increase of the erythrocyte sedimentation rate in systemic sclerosis. The heterozygous genotype of BsmI on adolescent idiopathic scoliosis is associated with a poor response to brace treatment and prognosis.
For the ACTN3 R577X polymorphism, we found no association of this snp.
R577X (RX or XX genotypes, specially XX) is associated with the development of idiopathic inflammatory miopathies (IIM) and dermatomyositis (DM). Despite that the X allele was not related with the severity of the disease.

R577X polymorphism is not associated with the risk of dystrophinopathy nor with adolescent diopathic coliosis in females.
There are no information for the association of the different studied snps and muscle properties in the context of musculoskeletal diseases.
Conclusion: Through the identification of key gene variants, this review furthers the elucidation of genetic associations with musculoskeletal diseases. However, no results are available regarding the association of the studied snps and muscle phenotype.
KEYWORDS: ACTN3, VDR, SNP, muscle
*IP and HM contributed equally for this review

PO350 – TEN-YEAR DRUG SURVIVAL OF BIOLOGICAL DISEASE MODIFYING ANTIRHEUMATIC DRUGS IN THE TREATMENT OF ANKYLOSING SPONDYLITIS: A RETROSPECTIVE ANALYSIS FROM HOSPITAL DE FARO

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Background: Ankylosing spondylitis (AS) is a chronic, progressive, axial inflammatory disease mainly involving the spine and the sacroiliac joints, as well as other sites of the axial skeleton. Treatment should be initiated as early as possible to prevent skeletal deformity and physical disability. The development of biologic agents (bDMARDs) has dramatically improved the management of AS. Drug survival has been reported as a composite measure of efficacy, safety and tolerability in clinical practice. Thus, long term survival of a bDMARD is an important indicator of its performance in terms of managing AS.
Objectives: to evaluate and compare the retention rate of bDMARDs in real-life practice and identify major reasons related to drug discontinuation in patients with AS.
Methods: retrospective study with the patients treated in our rheumatology department, starting bDMARD between January 2009 and September 2019 and fulfilling the ASAS criteria for AS. The data was collected from the data base Reuma.pt. Baseline demographic and clinical data were presented as means and standard deviations or absolute and relative frequencies. The drug survival rate was estimated using the Kaplan-Meier method and the predictor factors influencing of this rate were identified by multiple Cox proportional hazards models-regression analyses. The confidence level α=0.05 was considered throughout.

TABLE 1 – BDMARD SURVIVAL RATE

bDMARD	Survival % - 95% CI (lower 95%, upper 95%)					
	12 months	24m.	36m.	60m.	84m.	120m.
Adalimumab	76.9 (44.2, 91.9)	53.8 (24.8,76.0)	44.9 (17.7,69.0)	44.9 (17.7,69.0)	33.9 (11.7,61.3)	13.5 (0.98,41.9)
Etanercept	100 (1,1)	86.7 (56.4,96.5)	86.7 (56.4,96.5)	64.2 (33.3, 83.6)	64.2 (33.3, 83.6)	53.5 (22.8,76.7)
Golimumab	100 (1,1)	71.4 (25.8,92.0)	71.4 (25.8,92.0)	71.4 (25.8,92.0)	71.4 (25.8,92.0)	-
Infliximab	66.7 (28.2	55.6 (20.4,80.5)	33.3 (7.8,62.3)	22.2 (3.37,51.3)	11.1 (0.61,38.8)	-

Results: A total of 44 patients were included, 52% female (n=23) and 47% male (n=21).
Concerning the survival rate of the 1st bDMARD, at 84 months, GOL revealed the highest estimated rate (71.4%), although these results should bear in mind the low n with this bDMARD in this population. ETA had the second highest rate (71.4%). ADA only had a survival of 33.9% and INF presented the lowest survival (11.1%) (Table 1).
The highest global discontinuation rate was presented by INF (100%, n=9). ADA presented the highest discontinuation rate related to inefficacy (50%, n=5) and to adverse events (30%, n=3). The leading cause of drug discontinuation was inefficacy. GOL presented the lowest global discontinuation rate (28.6%, n=2) and the lowest discontinuation rates related to inefficacy (0%, n=0) and to adverse events (0%, n=0).
On the univariate analysis, taller patients seem to have a decreased risk (7%) of discontinuation. Most of the variables considered on the multivariate analysis seemed to have no impact on bDMARD survival, since they didn't show statistically significant results. However, ETA had a statistically significant decreased risk of discontinuation when compared to ADA.
The low n concerning the 2nd and 3rd bDMARDs wasn't adequate to a proper statistical analysis.
Conclusions: Over ten years, almost two thirds (61.4%) of patients discontinued their 1 st bDMARD. The leading cause of drug discontinuation was inefficacy. GOL presented a higher survival rate at 84 months, although bearing in mind the low n with this bDMARD in our population.

PO354 – MANIFESTAÇÕES PULMONARES DAS DOENÇAS REUMÁTICAS: CASUÍSTICA DA CONSULTA MULTIDISCIPLINAR DO INTERSTÍCIO DO CHUA – FARO

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Introdução: A Consulta Multidisciplinar do Interstício (CMDI) do Centro Hospitalar Universitário do Algarve (CHUA) – Faro realiza-se desde 10/2018 bimestralmente e nela participam as especialidades de Pneumologia, Reumatologia e Radiologia. Os principais objetivos são a discussão de situações em que existe dúvida diagnóstica, como a presença de patologia reumática inflamatória (PRI), o tipo e gravidade das manifestações pulmonares (MP) e decisão terapêutica conjunta.
Métodos: Análise e descrição das MP e eventual associação com PRI dos doentes seguidos entre 07/11/2018 e 31/12/2019 na CMDI do CHUA – Faro.
Resultados: Foram discutidos 24 doentes, 11 (46%) mulheres e 13 (54%) homens; a média de idades aquando das primeiras MP foi 61 anos.
Do total, 9 foram discutidos por patologia pulmonar e dúvida de associação a PRI, com ou sem alterações analíticas imunológicas, mas nos quais se concluiu não existir evidência de envolvimento clínico extra-pulmonar sugestivo de PRI (diagnósticos: fibrose pulmonar idiopática – FPI, pneumonia intersticial inespecífica – NSIP, NSIP fibrosante, micronódulos inespecíficos, pneumonite de hipersensibilidade, enfisema).
Os restantes 15 possuíam vários tipos de PRI concomitantes e a média de anos entre o diagnóstico da mesma e o surgimento de qualquer tipo de MP foi de 9 anos. Deste grupo, dos 4 doentes com diagnóstico de esclerose sistémica (ES) (3 ac anti-centrómero B+ e 1 ac anti-Scl70+), 1 tinha MP mistas de NSIP, pneumonia intersticial comum (UIP) e hemorragia alveolar difusa (HAD) associada a positividade ANCA PR3 e derrame pleural; os restantes tinham alterações inespecíficas (AI) (espessamento intersticial, bronquiectasias, nódulos). 1 doente tinha possível diagnóstico de doença mista do tecido conjuntivo e AI no TC. Os 3 doentes com artrite reumatóide (AR) seropositiva (anti-CCP e/ou FR em elevado título): 1 tinha alterações histológicas de pneumonite de hipersensibilidade (iatrogenia metotrexato) e os restantes tinham AI (bronquiectasias, enfisema). 2 doentes tinham possível síndrome de sobreposição de lúpus eritematoso sistémico e AR, 1 com alterações de UIP inicial e 1 com fibrose intersticial difusa. Dos 2 doentes com miopatias inflamatórias, 1 com polimiosite apresentava UIP e 1 com provável síndrome anti sintetase (anti-Jo1+) apresentava pneumonia organizativa. 1 doente tinha vasculite ANCA anti MPO+, com derrame pleural e HAD. 2 doentes tinham conectivite indiferenciada, com AI no TC (espessamento intersticial, bronquiectasias).
Nos casos de envolvimento pulmonar grave, os principais esquemas imunossupressores foram: ciclofosfamida (CYC) indução (i) + rituximab (RTX) manutenção (m); CYC (i) + micofenolato de mofetil (MMF) (m); MMF; RTX; e 1 caso fez azatioprina antes do diagnóstico de FPI, após o que iniciou anti fibrótico.
Discussão: A maioria dos doentes discutidos em consulta apresentavam PRI e MP concomitantes, tanto em associações bem conhecidas na literatura, como através de AI pulmonares que podem justificar vigilância. O doente com ES com manifestações graves de NSIP, UIP e HAD teve uma associação pouco descrita com vasculite ANCA positiva. Houve 1 caso de iatrogenia ao metotrexato com envolvimento pulmonar extenso. A CMDI aborda um grupo heterogéneo de patologias com potencial envolvimento pulmonar intersticial. Este pode ser determinante na decisão terapêutica e uma avaliação multidisciplinar é fundamental, representando a consulta uma importante mais valia no acompanhamento dos doentes.

P0357 – HIGH PGA SCORES IN PATIENTS WITH RHEUMATOID ARTHRITIS OTHERWISE IN REMISSION (NEAR-REMISSION) DOES NOT REFLECT SUBCLINICAL INFLAMMATION

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Introduction: Most recent treat-to-target recommendations propose that rheumatoid arthritis (RA) should strive for clinical remission as goal, as defined by American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) Boolean-based remission.(1) However, a large proportion (19%) of patients fail to achieve this aim only due patient global assessment >=1 (2), despite apparent abrogation of the inflammation leading to a risk of immunosuppressive overtreatment.
Objective: To evaluate and compare the presence of subclinical inflammation in RA patients, assessed by ultrasound (US), in different ACR/EULAR Boolean remission states.
Methods: Observational cross-sectional study of consecutive RA patients fulfilling the ACR/EULAR 2010 classification criteria. Patients were stratified

into (1) Remission (SJC28≤1, TJC28≤1, CRP≤1 mg/dl and PGA≤1), (2) Near-Remission (SJC28≤1, TJC28≤1, CRP≤1mg/dl and PGA>1) and (3) Non remission (any of TJC, SJC and CRP >1) adapting the Boolean criteria of remission ACR / EULAR.(1) Sociodemographic data, PGA, rheumatoid factor (RF), anti–citrullinated protein antibody (ACPA) and radiographic erosions were collected at the time of clinical visit. US assessment of the 44 joints, tendon sheaths and bursas (scored according to OMERACT definitions(3)) was performed at the same day, by a different rheumatologist, expertise in MSK ultrasound and blinded for the clinical patient status. Global US score (GUSS) was defined as the sum of composite US scores for all joints, tendon sheaths and bursas assessed. Patients classified as remission, near-remission and non-remission were compared using the Mann Whitney or the Kruskal Wallis tests as appropriate. A p£0.05 was considered statistically significant.

Results: We included 130 patients, 50 in non-remission state and 40 in both remission and near-remission state; 83.8% (n=109) female, mean age of 63.1±12.2 years. Most patients were positive RF (65.4%) and/or anti-CCP (58.5%) and 39.2% had documented erosions. The median of PGA was 3.3 (IQR 5); it was lower in remission patients (p<0.01) without significant difference between near and non-remission patients (4.8 (IQR 3) Vs 5.0 (IQR 3), p=1.00). The median GUSS was 7.0 (IQR 11) without statistically significant difference between remission and near-remission groups (6.0 (7) Vs 4.0 (8), p=0.80).

Conclusions: In our cohort, we found no differences regarding the presence of subclinical inflammation in RA patients between remission and near-remission groups. These findings reinforce the inadequate application of PGA in remission classification criteria leading to the risk of immunosuppressive overtreatment in a large proportion of patients.

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P0359 – NO RELATIONSHIP BETWEEN LUMBAR BONE MINERAL DENSITY AND SYNDESMOPHYTE FORMATION AT THE SAME LEVEL – A MULTILEVEL ANALYSIS IN PATIENTS WITH RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS

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Background/Purpose: In radiographic axial spondyloarthritis (r-axSpA) it has been hypothesized that inflammation-driven bone loss triggers bone repair but at anatomically distinct sites of the same vertebra (i.e. bone loss occurring in the trabecular bone and bone repair in the periosteum)[1]. However, the possible association between bone loss and new bone formation at the same individual vertebra has never been studied. The purpose of this study was to investigate if in r-axSpA low vertebral bone mineral density (BMD) is associated with development of new syndesmophytes at the same vertebral level.

Methods: In a post-hoc analysis from the ASSERT trial (infliximab vs placebo) dual-energy X-ray absorptiometry was used to measure baseline BMD (g/cm^2) of the lumbar spine L1 to L4. Syndesmophyte formation was assessed in the same vertebrae on conventional radiographs defined as an increase in modified Stoke Ankylosing Spondylitis Spine Score from 0 or 1 to 2 or 3 after 2 years. Radiographs were scored by two readers. Generalized estimating equations (GEE) adjusted for within-patient correlation across multiple vertebrae, taking potential confounders into account (Table 1).

Results: We analyzed 599 vertebrae in 165 r-axSpA patients (78% male, mean (SD) age 38 (10) years, 67% with at least one syndesmophyte anywhere in the spine). In total, 24 to 74 new syndesmophytes developed in 9 (5%) to 30 (18%) patients and 13 (2%) to 39 (7%) vertebrae, if either a syndesmophyte was seen by both or only one of the readers (i.e. specific and sensitive definitions) respectively. Analyses with both definitions, and both uni- and multivariable, showed

TABLE 1 – RELATIONSHIP BETWEEN BASELINE BMD AND 2-YEAR SYNDESMOPHYTE FORMATION AS REPORTED BY BOTH READERS AND AT LEAST ONE OF THE READERS – MULTIVARIABLE ANALYSIS (ADJOR (95% CI))

Independent variables	New radiographic syndesmophyte formation according to both reader 1 and reader 2	New radiographic syndesmophyte formation according to reader 1 or reader 2
BMD (g/cm^2)	0.56 (0.01, 44.45)	0.26 (0.03, 2.63)
Age (years)	1.03 (0.95, 1.11)	1.04 (1.00, 1.09)*
Gender (male)	0.82 (0.10, 6.85)	1.42 (0.50, 4.01)
Disease duration (years)	1.05 (0.97, 1.14)	1.00 (0.96, 1.05)
ASDAS-CRP	1.79 (0.66, 4.86)	1.09 (0.63, 1.87)
HLA-B27	0.13 (0.02, 0.89)	0.54 (0.15, 1.89)
Treatment with NSAIDs	0.41 (0.07, 2.47)	0.82 (0.21, 3.13)
Treatment with infliximab	1.82 (0.22, 15.31)	1.11 (0.45, 2.69)
Presence of MRI VCI at baseline	4.00 (0.99, 16.13)	4.32 (1.95, 9.60)*
Presence of MRI VCFD at baseline	0.69 (0.16, 3.01)	1.23 (0.60, 2.54)
Presence of syndesmophytes at baseline	20.20 (0.96, 424.63)	3.14 (1.14, 8.66)*

BMD, bone mineral density; VCI, vertebral corner inflammation; VCFD, vertebral corner fat deposition. *p<0.05

no significant association between baseline local vertebral BMD and new syndesmophyte formation after two years in the same vertebra (multivariable analysis adjOR (95%CI): 0.56 (0.01, 44.45) (specific definition) and 0.26 (0.03, 2.63) (sensitive definition)) (Table 1).

Conclusion: In patients with active and established r-axSpA, with an observed low incidence of lumbar spine syndesmophyte formation over two years, no relationship was found between baseline BMD and new radiographic syndesmophyte formation in the same vertebra.

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This study, carried out under YODA Project #2018-2761, used data obtained from the Yale University Open Data Access Project, which has an agreement with JANSSEN RESEARCH & DEVELOPMENT, L.L.C.. The interpretation and reporting of research using this data are solely the responsibility of the authors and does not necessarily represent the official views of the Yale University Open Data Access Project or JANSSEN RESEARCH & DEVELOPMENT, L.L.C..

P0360 – EFFECTIVENESS AND SAFETY OF ORIGINAL AND BIOSIMILAR ETANERCEPT (ENBREL® VS BENEPALI®) IN BDMARD-NAÏVE PATIENTS

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Background: The patent expiration of the original etanercept in Europe has facilitated the development of biosimilar products, creating the prospect of reduced treatment costs. In 2016, Benepali® became the first etanercept biosimilar to obtain marketing authorization in Europe. A biosimilar is required by the European Medicines Agency and the Food and Drug Administration to demonstrate similarity to the original product in terms of quality characteristics and biological activity. Furthermore, it must demonstrate comparable safety and effectiveness [1,2].

However, subtle differences in efficacy and safety outcomes have been noted and the potential clinical implications for this in daily practice are not established. The ability to extrapolate license indications without data supporting the use of that product in certain indications may create doubt. On-going vigilance by physicians in reporting adverse events and treatment outcomes is consequently essential[1,2,3].

Our **primary aim** was to compare the effectiveness and safety of original and biosimilar etanercept, in bDMARD-naïve patients, measured by persistence rates over 3 years of follow-up.

Methods: We performed a retrospective non-interventional study in our center, using data collected prospectively from Reuma.pt database (The Rheumatic Diseases Portuguese Register). Electronic clinical records were reviewed for all patients that fulfil the study inclusion criteria. Real-world anonymous patient-level data from the Reuma.pt database was used. The inclusion criteria were the following: age ≥ 18 years old; patients that fulfil the ACR/EULAR 2010 classification criteria for Rheumatoid Arthritis (RA); CASPAR criteria for Psoriatic Arthritis (PsA) or ASAS classification criteria for Spondyloarthritis (SpA) (axial and peripheral); patients with active rheumatic disease who initiated treatment with etanercept as first line of biological treatment.

Kaplan-Meyer was used to calculate persistence rate in biologic treatment. Reasons for discontinuing therapy were summarized using descriptive statistics and stratified by the treatment. *P*-value was considered significant at <0.05.

Results: One hundred and thirteen patients were included, 54 with RA, 36 with PsA and 23 with SpA. Demographic and clinical characteristics at baseline are listed in table 1. Disease characteristics are enumerated in table 2.

TABLE 1 – DEMOGRAPHIC AND CLINICAL BASELINE CHARACTERISTICS

	Rheumatoid Arthritis			Psoriatic Arthritis			Spondyloarhritis		
	Enbrel® (n=44)	**Benepali® (n=10)**	***p*-value**	**Enbrel® (n=30)**	**Benepali® (n=6)**	***p*-value**	**Enbrel® (n=16)**	**Benepali® (n=7)**	***p*-value**
Female, n (%)	37 (84.1)	3 (30.0)	**0.002**	17 (56.6)	4 (66.6)	0.65	5 (31.2)	5 (71.4)	0.42
Age, mean (±SD)	62 (12.2)	62.2 (7.6)	0.96	55.5 (12.2)	49.3 (6.7)	0.24	49.7 (13.5)	54.1 (6.7)	0.06
Smoke, n (%)	9 (20.4)	4 (40.0)	0.30	8 (26.7)	0	0.17	3 (18.8)	2 (28.5)	0.21
Comorbidities, n (%)									
- **Hypertension**	11 (25.0)	2 (20.0)	0.69	7 (23.3)	1 (16.7)	0.66	5 (31.3)	2 (28.6)	0.97
- **Dyslipidaemia**	8 (18.2)	3 (30.0)	0.45	5 (16.7)	1 (16.7)	0.95	4 (25)	1 (14.3)	0.61
- **Diabetes**	9 (20.4)	1 (10.0)	0.41	2 (6.7)	0	0.50	0	0	-
- **CV disease**	2 (4.5)	1 (10.0)	0.64	0	1 (16.7)	0.03	0	0	-
HLA B27 positivity, n (%)	-	-	-	5 (16.7)	0	0.19	12 (75.0)	4 (57.1)	0.20
Rheumatoid factor positivity, n (%)	34 (77.3)	7 (70.0)	0.63	1 (3.3)	0	0.68	-	-	-
ACPA positivity, n (%)	27 (61.4)	6 (60.0)	0.42	2 (6.7)	0	0.57	-	-	-
Disease duration, mean (±SD)	8.9 (7.1)	12.6 (8.1)	0.16	11.9 (9.4)	8.7 (4.8)	0.41	8.8 (7.6)	9.1 (6.4)	0.91
Treatment									
- **cDMARDs, n (%)**	34 (77.3)	9 (90.0)	0.46	24 (80.0)	4 (66.7)	0.42	10 (62.5)	4 (57.1)	0.78

CV: Cardiovascular; ACPA: Anti-Citrullinated Protein Antibodies; cDMARD: conventional Disease-Modifying Anti Rheumatic Drugs

TABLE 2 – DISEASE ACTIVITY AT BASELINE

	Rheumatoid Arthritis			Psoriatic Arthritis			Spondyloarhritis		
	Enbrel® (n=44)	**Benepali® (n=10)**	***p*-value**	**Enbrel® (n=30)**	**Benepali® (n=6)**	***p*-value**	**Enbrel® (n=16)**	**Benepali® (n=7)**	***p*-value**
CRP, mean (±SD)	2.2 (0.5)	1.6 (0.8)	0.51	2.9 (0.7)	2.9 (1.2)	0.99	5.9 (2.0)	3.5 (1.9)	0.49
ESR, mean (±SD)	29.6 (3.5)	24.6 (7.1)	0.50	41.6 (5.8)	32.2 (7.6)	0.46	42.9 (4.1)	30.2 (6.4)	0.11
SJC, mean (±SD)	8.5 (1.1)	5.7 (1.5)	0.19	4.6 (1.1)	2.0 (0.7)	0.25	1,0 (0.6)	1.2 (1.0)	0.88
TJC, mean (±SD)	11.5 (1.3)	7.2 (2.4)	0.12	5.9 (1.4)	2.0 (0.7)	0.18	2.1 (1.0)	2.0 (1.4)	0.97
Patient Global Assessment, mean (±SD)	55.7 (4.3)	44.5 8.2)	0.21	54.7 (4.6)	49.8 (5.7)	0.63	57.5 (7.7)	73.5 (8.0)	0.32
Pain VAS, mean (±SD)	53.4 (4.8)	41.8 (10.7)	0.27	56.6 (6.5)	51.4 (6.2)	0.69	52.2 (7.7)	48.5 (36.5)	0.88
DAS28, mean (±SD)	5.5 (0.2)	4.3 (0.5)	**0.01**	4.8 (0.3)	4.2 (0.2)	0.20	3.9 (0.2)	3.9 (0.7)	0.20
CDAI, mean (±SD)	31.1 (2.5)	20.7 (4.6)	0.06	17.1 (2.2)	13.6 (2.0)	0.35	-	-	-
SDAI, mean (±SD)	33.7 (2.8)	22.3 (4.9)	**0.05**	19.4 (2.6)	15.7 (1.5)	0.40	-	-	-
HAQ, mean (±SD)	1.4 (0.1)	1.0 (0.2)	0.15	1.4 (0.2)	0.5 (0.2)	**0.04**	-	-	-
DAPSA, mean (±SD)	-	-	-	22.7 (2.3)	17.0 (1.9)	0.20	-	-	-
ASDAS, mean (±SD)	-	-	-	4.2 (0.3)	3.6 (0.6)	0.38	4.0 (0.2)	3.8 (0.5)	0.77
BASDAI, mean (±SD)	-	-	-	6.6 (0.6)	5.3 (1.1)	0.32	5.9 (0.7)	5.9 (0.2)	0.97
BASFI, mean (±SD)	-	-	-	6.7 (0.6)	4.5 (1.6)	0.13	5.5 (0.7)	7.6 (0.6)	0.17

CRP: C-Reative Protein; ESR: Erythrocyte Sedimentation Rate; SJC: Swollen joint count; TJC: Tenderness joint count; VAS: Visual Analogue Scale; DAS: Disease Activity Score; CDAI: Clinical Disease Activity Index; SDAI: Simplified Disease Activity Index; HAQ: Health Assessment Questionnaire; DAPSA: Disease Activity in Psoriatic Arthritis; ASDAS: Ankylosing Spondylitis Disease Activity Score; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; BASFI: Bath Ankylosing Spondylitis Functional Index

The 3-year retention rate in RA patients was similar for both drugs: 60.0% with a median time-on-drug of 21.6 months for Benepali® and 55.8% with a median time-on-drug of 24.1 months for Enbrel® (p=0.851). In PsA patients, the retention rate was higher for Enbrel® (80.0% with a median time-on-drug of 32.3 months) from Benepali® (66.7% with a median time-on-drug of 27.8 months) but not statistically significant (p=0.226). In SpA patients, the persistence rate was significantly lower for Benepali® (42.9% with a median time-on-drug of 19.7 months) comparatively to Enbrel® (82.4% with a median time-on-drug of 32.4 months; p=0.025).

Overall, 38 (33.6%) patients stopped etanercept because of inefficacy (n=12 [31.6%]), adverse events (n=15 [39.4%]) or other reasons (n=11 [29.0%]). No differences were observed among both treatment subgroups in the cumulative incidence of discontinuation due to inefficacy, adverse events or other reasons (p>0.05). (Table 3).

Conclusion:

Our centre results presented above indicate similar effectiveness and safety of Benepali® compared to Enbrel® in RA and PsA. However, in SpA we observed

TABLE 3 – REASONS FOR DISCONTINUATION AND CUMULATIVE INCIDENCE OF EVENTS

	Rheumatoid Arthritis			Psoriatic Arthritis			Spondiyoarhritis		
Reasons for discontinuation	**Enbrel® (n=44)**	**Benepali® (n=10)**	***p*-value**	**Enbrel® (n=30)**	**Benepali® (n=6)**	***p*-value**	**Enbrel® (n=16)**	**Benepali® (n=7)**	***p*-value**
Inefficacy	5	4	0.06	1	1	0.84	0	1	0.20
- Primary no response	3	1		0	0		0	0	
- Secondary no response	2	3		1	1		0	1	
Adverse events	10	0	0.12	2	0	0.71	1	2	0.10
- Major infections	5	0		0	0		0	0	
- Malignancies	1	0		0	0		1	0	
- Injection reactions	1	0		0	0		0	0	
- Cutaneous diseases	0	0		0	0		0	0	
- Cardiovascular diseases	1	0		0	0		0	1	
- Liver toxicity	0	0		1	0		0	0	
- Deaths	2	0		0	0		0	1	
- Demyelinating disease	0	0		1	0		0	0	
Other reasons	4	0	0.37	3	1	0.11	2	1	0.58
Total (n=38)	**19**	**4**		**6**	**2**		**3**	**4**	

a statistically significant difference between Benepali® and Enbrel® (p=0.025), with a higher time-on-drug for Enbrel®. This could result of a lower sample of patients with SpA, one limitation of this study. In the future, we intend to expand this study using data from REUMA.PT to evaluate consistently the effectiveness and safety of both drugs in a large cohort.

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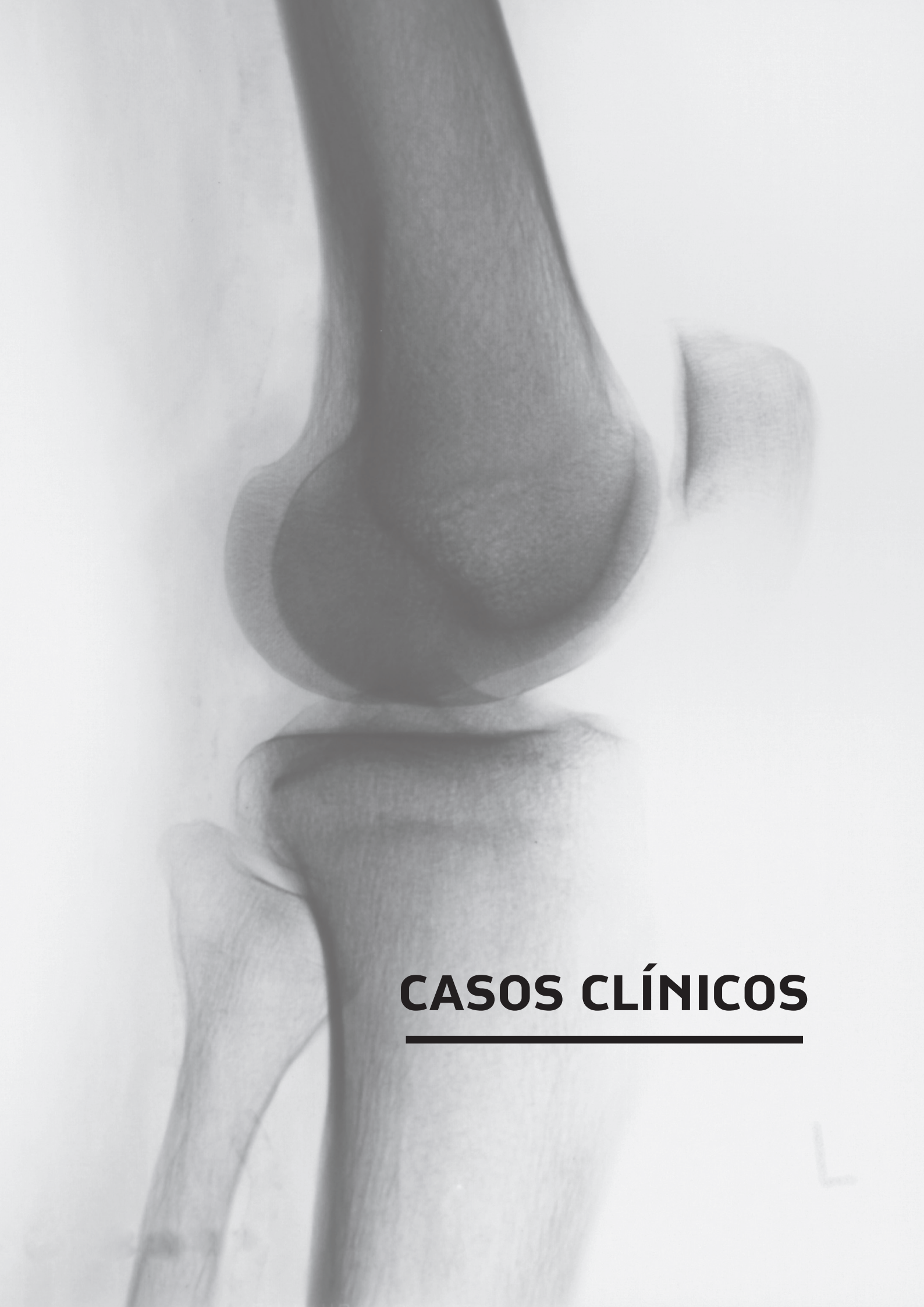

CASOS CLÍNICOS

Casos Clínicos

CC021 – HIPERPARATIROIDISMO PRIMÁRIO E HIPERCALCEMIA HIPOCALCIÚRICA FAMILIAR: UM CASO RARO

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Introdução: O hiperparatiroidismo primário e as neoplasias são as principais causas de hipercalcemia. O hiperparatiratiroidismo primário corresponde à hiperprodução de hormona paratiroideia (PTH) por um ou mais adenomas paratiroideus. A hipercalcemia hipocalciúrica familiar (HHF) é uma causa rara de hipercalcemia que se associa a baixa excreção renal de cálcio e a valores normais/elevados de PTH, ocorrendo como consequência de um distúrbio genético no gene do recetor sensível ao cálcio (CaSR), resultando em hipossensibilização generalizada ao cálcio com hipercalcemia compensatória.

Caso clínico: Doente do sexo feminino, 39 anos de idade, com espondilite anquilosante com 10 anos de evolução, medicada com golimumab 50mg s.c./mês (desde fevereiro de 2015), sulfassalazina 2g/dia e acemetacina 210mg/dia. Em julho de 2015, o estudo analítico revelou hipercalcemia (cálcio total 5.7 mEq/L, normal 4.1-5.1; cálcio ionizado 3.14 mEq/L, normal 2.26-2.64) e hipofosfatemia (2.6 mg/dL; normal 2.7-4.5). Foi alargado o estudo complementar, revelando PTH 66.3 pg/mL (normal 10-65), urina de 24h com normocalciúria (limite inferior da normalidade – 5.6 mEq/24h, normal 5-15) e normofosfatúria, proteinograma e enzima conversora da angiotensina normais. A ecografia renopélvica não revelou alterações compatíveis com litíase ou dilatação dos sistemas excretores e a ecografia cervical revelou nódulo hipoecogénico adjacente à vertente posterior do terço médio do lobo esquerdo. A cintigrafia das paratiróides revelou imagem de hipercaptação posterior ao terço inferior do lobo direito da tiróide que poderia corresponder a paratiróide anómala e a densitomeria óssea mostrou osteopenia lombar e femoral. A doente foi enviada à consulta de Cirurgia Endócrina onde realizou ecografia das paratiróides que revelou 3 formações nodulares hipoecoicas, homogéneas e bem delimitadas, compatíveis com hiperplasia das paratiróides. Foi proposta para cirurgia, que realizou em abril de 2018, com a exploração cirúrgica a revelar hiperplasia aparente de todas as paratiróides com doseamentos intra-operatórios de PTH aumentados, optando-se pela exérese de ambas as paratiróides superiores, da inferior direita e de 2/3 da paratiróide inferior esquerda, com descida da PTH para valores normais. A histologia revelou hiperplasia de células principais na paratiróide inferior esquerda e na superior direita. Em janeiro de 2019, por apresentar hipercalcemia persistentemente elevada (cálcio total 5.4 mEq/L e cálcio ionizado 3.0 mEq/L) com PTH normal (45.1 pg/mL) e hipocalciúria associada (3.0 mEq/24h) foi repetida a cintigrafia das paratiróides, que não revelou fixação anormal. Assim, foi requisitado estudo genético de HHF que detetou no exão 7 do gene CaSR, em heterozigotia, a variante provavelmente patogénica c.2243C>G (p.Pro748Arg), já associada a HHF tipo 1 de hereditariedade autossómica dominante. Foi orientada para as consultas de Genética Médica e de Nefrologia, mantendo-se assintomática e sob vigilância clinico-analítica.

Conclusão: Apresentamos o caso de uma doente com hiperparatiroidismo primário no contexto de hiperplasia das paratiróides, que manteve hipercalcemia após o tratamento cirúrgico, tendo o estudo complementar culminado no diagnóstico de HHF tipo 1. A associação das duas situações é rara, com escassos casos descritos na literatura. Este caso alerta para a importância do reconhecimento da HHF como causa rara e benigna de hipercalcemia, sem tratamento específico e na maioria das vezes assintomática, realçando-se o aumento da remodelação óssea que lhe pode estar associado.

CC022 – MONOARTRITE RECIDIVANTE E ESTUDO DO HLA: DESCRIÇÃO DE DOIS CASOS CLÍNICOS

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Introdução: Algumas variantes alélicas do HLA ou loci têm sido associadas a diversas patologias reumáticas.

O alelo HLA-B35 associa-se a um aumento do risco de doença de Still do adulto, artrite idiopática juvenil (AIJ), espondilartrite (SpA) associada à doença inflamatória intestinal, artrite reativa, psoríase e de hipertensão pulmonar nos doentes com esclerose sistémica; o alelo HLA-B39 associa-se a um risco aumentado de SpA axial, artrite reativa, SpA indiferenciada e AIJ; o alelo HLA- B44 está associado ao risco aumentado de AIJ, artrite reumatóide (AR), SpA associada à doença de Crohn e osteoartrose nodal erosiva; o HLA-B14 associa-se à espondilite anquilosante. Algumas mutações no alelo HLA-DRB1 estão associadas à AR, particularmente em asiáticos, assim como a outras doenças reumáticas.

Caso clínico 1: Doente do sexo masculino, 17 anos, que praticava andebol. Enviado pela consulta de Ortopedia em outubro de 2016 por episódios recorrentes de monoartrite do cotovelo esquerdo, autolimitados (resolução em 2 semanas) e com períodos intercríticos de 2 meses, desde os 14 anos de idade. Tinha já sido submetido a 3 sinoviorteses químicas e a uma ligamentoplastia por instabilidade do cotovelo. Não apresentava atingimento de outras articulações e referia úlceras orais recorrentes e fenómeno de Raynaud desde os 15 anos de idade. Tinha a seguinte história familiar: mãe com 2 episódios de uveíte anterior idiopática, pai com psoríase e irmão com úlceras orais recorrentes. O estudo imunológico foi negativo, a capilaroscopia e a ressonância magnética das articulações sacroilíacas foram normais. O estudo do HLA revelou negatividade para o HLA-B51 e positividade para HLA-B14 e B39. Foi medicado com deflazacort 6mg/dia e colchicina 1mg/dia e aconselhado a evitar a prática de andebol, com melhoria marcada das úlceras orais e dos episódios de monoartrite do cotovelo esquerdo, tendo necessitado apenas de mais uma sinoviortese com hexacetonido de triancinolona em setembro de 2017.

Caso clínico 2: Doente do sexo masculino, de 28 anos, com história de asma, enviado pela consulta de Ortopedia em fevereiro de 2017 por hidrartrose recidivante do joelho esquerdo desde os 12 anos de idade, com períodos intercríticos de 2 anos e já submetido a várias artroscopias, cuja biópsia da membrana sinovial demonstrou uma sinovite crónica com francos sinais de atividade e de hemorragia antiga. Apresentava resposta parcial a sinoviorteses químicas e não respondeu a AINEs e à sulfassalazina. O estudo imunológico foi negativo, assim como o estudo bacteriológico do líquido articular (bacteriológico, micobacteriológico e DNA de Chlamydia). A ressonância magnética mostrou derrame articular de grande volume, com sinovite difusa, exlcuindo patologia tumoral. O estudo genético revelou positividade para HLA-B35, B44 e DRB1*07,15. Iniciou colchicina 1mg/dia (com posterior desmame para 0.5mg/dia) e foi submetido a sinoviortese do joelho esquerdo com hexacetonido de triancinolona em julho de 2017, mantendo-se assintomático desde então.

Conclusão: Cada vez mais variantes alélicas dos genes do HLA têm sido identificadas como fatores de risco para algumas doenças reumáticas. Na prática, a sua presença no doente individual nem sempre se traduz num determinado diagnóstico. Os casos de monoartrite recorrente são casos desafiantes em que, após a exclusão de outras causas (nomeadamente patologia infeciosa ou tumoral), poderá ser útil o estudo genético dos alelos HLA, quer para um possível diagnóstico definitivo, quer para tranquilizar o doente.

CC023 – HIPOFOSFATÁSIA, UMA DOENÇA METABÓLICA ÓSSEA RARA: DESCRIÇÃO DE 2 CASOS CLÍNICOS

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Introdução: A hipofosfatásia é uma doença óssea metabólica rara que resulta de mutações no gele ALPL, responsável pela codificação da isoenzima da fosfatase alcalina (FA) não específica de tecido, cuja função passa pela desfosforilação de alguns substractos, como o pirofosfato inorgânico, responsável por inibir a mineralização óssea. O espectro clínico da doença é muito heterogéneo, variando desde a inviabilidade intrauterina até manifestações frustes, estando a reposição enzimática apenas indicada em idade pediátrica.

Caso clínico 1: Doente do sexo feminino, 48 anos, com o diagnóstico de espondilartrite axial pré-radiográfica desde fevereiro de 2018, sob acemetacina 90mg id. A doente apresentava história de dores ósseas generalizadas com vários anos de evolução, associada à perda de várias peças dentárias e a baixa estatura (149cm). O estudo complementar revelou metabolismo fosfocálcico normal (incluindo PTH), com exceção de hipovitaminose D (5.9 ng/mL) e FA sérica no limite inferior da normalidade (33 U/L; normal 30-120), com baixos níveis da FA óssea (9.8 U/L; normal 12-42). Tendo em conta a história clínica e os achados laboratoriais, foi pedido o estudo genético do gene ALPL que detetou a presença da variante genética c.658 G>A (p.Gly220Arg), associada à

hipofosfatásia autossómica recessiva do adulto. Foi pedida consulta de Genética Médica, que ainda não foi realizada, encontrando-se atualmente sob suplementação com vitamina D, mantendo-se a vigilância clínico-analítica.

Caso clínico 2: Doente do sexo feminino, 66 anos, enviada à consulta de Reumatologia em março de 2018 por osteoporose densitométrica (score T: lombar -2.3; fémur direito -2.3; cólo femoral -2.5), já medicada com ácido alendrónico semanal há poucas semanas e sob suplementação com vitamina D. Referia, também, perda de algumas peças dentárias e dores ósseas ocasionais. O estudo complementar revelou metabolismo fosfocálcico normal (incluindo PTH), com exceção da FA sérica, que se apresentava perto do limite inferior da normalidade (41 U/L), tendo sido pedido o doseamento da FA óssea, que veio diminuída (8.3 U/L). Nesse contexto, e com a suspeita de hipofosfatásia, foi pedida a sequenciação do gene ALPL, que revelou a presença da variante genética c.413 G>C (p.Arg138Pro) em heterozigotia, classificada como variante de significado clínico indeterminado. Foi referenciada à consulta de Genética Médica, cessou o alendronato semanal e mantem-se apenas sob suplementação com vitamina D. O seguimento em Genética Médica permitiu descobrir sintomas sugestivos de hipofosfatásia na filha da doente, estando a aguardar referenciação para a consulta.

Conclusão: Estes dois casos clínicos salientam a importância do reconhecimento da hipofosfatásia, doença na maioria das vezes com um grande atraso diagnóstico e na qual o uso da terapêutica anti-reabsortiva pode ser deletério, aumentando o risco de fraturas ósseas atípicas.

CC031 – SYSTEMIC SCLEROSIS AND NEPHROTIC SYNDROME: AN UNUSUAL ASSOCIATION

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Introduction: Renal complications are common in Systemic Sclerosis (SSc). The most common is scleroderma renal crisis (SRC), but several patterns of renal pathology are recognized. The appearance of nephrotic syndrome has been rarely described in SSc. We report a uncommon case of nephrotic syndrome in a patient with SSc.

Case Report: 39-year-old man diagnosed with limited cutaneous SSc at the age of 21, presenting with Raynaud's phenomenon, digital ulcers, joint and limited distal skin involvement, under treatment with naftidrofuryl and aminaftone. He was admitted to the emergency department with a two-weeks history of morning, bilateral and malleolar edema with progression up to the knees. One week before admission, he noted the appearance of foamy urine. He denied any history of exposure to toxins. Upon hospitalization, he had blood pressure of 140/50 mmHg, edema up to his knees bilaterally and digital ulcers in both hands. He had normal complete blood count; ANA >1/640 (homogeneous pattern), high titter Scl-70 (4.390 UQ), negative anti-DS-DNA and ANCA, elevated ESR (68mm/h), total protein of 5.2g/dL (6.6-8.7), pCreat. of 1.03mg/dl; total cholesterol of 363mg/dL and LDL cholesterol of 274mg/dL. The 24-hour urine analysis showed a volume of 2850 mL, with proteins 23g/L. The kidney ultrasound showed a discrete hyperechoic parenchyma. The kidney biopsy showed a renal cortex with 12 glomeruli, with a slight increase in cellularity and mesangial matrix, without changes of the glomerular basement membranes. Immunofluorescence showed 14 glomeruli with mesangial deposits of IgA (+++) and IgM (+). Electron Microscopy showed podocytopathy. These histologic findings were compatible with IgA nephropaty, with an uncommon clinical presentation characterized by nephrotic syndrome, suggesting a superimposed podocytopathy, with characteristics of minimal change disease. The patient was treated with prednisolone 0.5mg/kg/day for 10 weeks (tapering 5mg/week during the following 6 months) and mycophenolate mofetil 500mg, twice a day and proteinuria decreased. He is currently on complete remission (proteinuria 78.8 mg/24h), without edema and controlled blood pressure.

Conclusion: Few cases of nephrotic syndrome associated with SSc have been described and most of them were associated with D-penicillamine. Our case represents an exceptional form of nephropathy in a SSc patient, unrelated with the use of drugs usually associated with nephrotic syndrome. The prognosis of IgA nephropathy usually follows a benign course and end-stage renal disease develops only in 20% of patients at 20 years of follow up.

CC034 – FRATURA VERTEBRAL INSTÁVEL NUM DOENTE COM ESPONDILITE ANQUILOSANTE – A IMPORTÂNCIA DO DIAGNÓSTICO PRECOCE

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Introdução: A Espondilite Anquilosante (EA) é uma doença inflamatória crónica que altera as propriedades biomecânicas da coluna vertebral, o que conduz a um risco aumentado de fraturas vertebrais, mesmo com traumatismos ligeiros. Doentes com EA apresentam um risco de fratura ao longo da vida quatro vezes superior ao da população em geral.
Caso clínico: Doente do sexo masculino, de 52 anos, com EA com envolvimento axial e periférico, com 20 anos de evolução, medicado com anti-inflamatório não esteróide e metotrexato 20mg subcutâneo semanal. Foi avaliado por dorsalgia mecânica, sem sintomas neurológicos associados, com 2 meses de evolução, após queda da própria altura em contexto de lipotímia. Apresentava ao exame objetivo cifoescoliose dorsal, sem agravamento da dor à mobilização da coluna dorsolombar e sem áreas eletivas de dor à palpação. Realizou radiografia anteroposterior e lateral da coluna dorsal com evidência de "coluna em bambu" e com presença de traço de fratura ao nível da plataforma superior da 8ª vértebra dorsal (D8). A ressonância magnética evidenciou uma fratura recente transversal no espaço intersomático e plataformas D7-D8 com maior expressão na plataforma superior de D8 e irradiação aos elementos posteriores, nomeadamente pedículos, facetas articulares de D8, base das apófises transversas de D8, articulações costovertebrais e fraturas proximais das costelas, sem evidência de compressão mielorradicular nem componentes hemáticos intracanalares. Foi recomendado o uso de ortótese dorsolombar para estabilização e dois dias após o diagnóstico foi submetido a fixação da fratura por instrumentação pedicular. Teve alta hospitalar ao 3º dia pós-operatório, a deambular, sem défices neurológicos. Aos 6 meses de pós-operatório, o doente encontra-se assintomático e na radiografia da coluna é visível uma ponte óssea anterior ao nível da fratura.
Conclusão: As fraturas vertebrais nos doentes com EA são muitas vezes negligenciadas pelo médico e doente, pela dificuldade em distinguir a dor aguda da fratura da dor habitual. O diagnóstico tardio pode piorar o prognóstico, uma vez que estas fraturas são geralmente instáveis e requerem tratamento adequado para evitar lesões neurológicas. Assim, é de extrema importância alertar os doentes para os riscos dos traumatismos da coluna vertebral, de forma a minimizarem o risco de queda e a procurar assistência precocemente.

CC036 – CONGENITAL DISLOCATION OF PATELLA – A RARE CAUSE OF "MUSCLE WEAKNESS"

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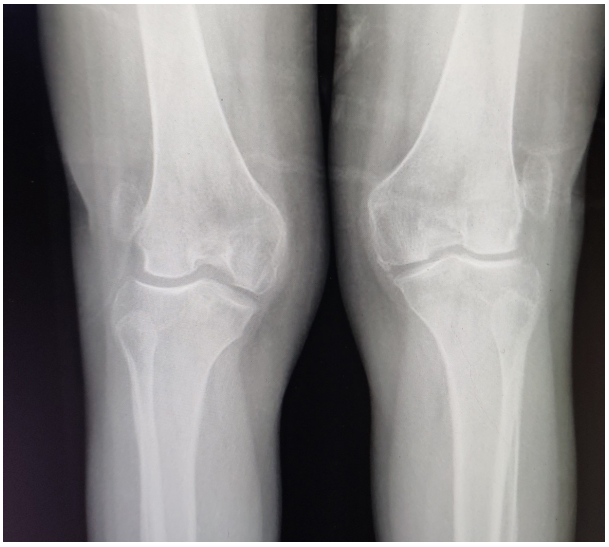
Introduction: Congenital dislocation of patella is a rare pathological condition characterized by permanent lateral dislocation of the patella with dysfunction of extensor mechanism of the knee. Infants present genu valgum and contracture of the flexed knee in association with external rotation of the tibia. When these deformities are not present, the diagnosis may be delayed until a skeletally mature patient.
Clinical Case: A 64 year-old woman was referred to our rheumatology department for workup of prolonged gait disturbance, lower limb muscle weakness and recurrent falls. She had a history of bilateral congenital absence of patella and suspected degenerative neuromuscular disease, under investigation at the neurology department. She started walking at 4 years-old. Child development was otherwise normal. There was no relevant family history. Physical examination revealed normal height, crutch walking, genu varum deformity, bilateral non-palpable patella and reduced muscle strength (grade 2/5) in proximal lower limbs with normal muscle strength (grade 5/5) in distal lower and upper limbs. Deep tendon reflexes and sensation were preserved. Laboratory studies revealed normal acute-phase reactants and muscle enzymes. She had already been tested for antinuclear antibodies, including myositis-specific and myositis-associated antibodies, which were all negative. Electromyography showed no signs of muscle fiber damage and muscle biopsy was also normal. Genetic screening for the main hereditary neuromuscular diseases came up negative. Radiographies of the knees were performed: in lateral view, patella was bilaterally absent with no other relevant abnormalities; in antero-posterior view, on the other hand, a small laterally displaced patella was seen on both sides (Figure 1), compatible with congenital dislocation of patella.
Conclusion: This case highlights the importance of differential diagnosis in the presence of a rather non-linear clinical picture. This patient was under investigation for a neuromuscular disease due to a proximal

FIGURE 1 – ANTERO-POSTERIOR KNEE RADIOGRAPHY SHOWING A BILATERAL SMALL LATERALLY DISPLACED PATELLA



muscle weakness in her lower limbs. However, this muscle weakness was not due to a pathology of the muscle fiber but instead of a compromise of the lower limb extensor system, explained by a displaced and dysfunctional patella. Surgical treatment is usually indicated in early childhood and aims to prevent disability and deformity. In the adult, these complications are usually already installed and thus surgical treatment is controversial.

CC048 – ANCA-ASSOCIATED VASCULITIS IN A PATIENT WITH ENTEROPATHIC SPONDYLARTHRITIS TREATED WITH TNFI: A CASE REVIEW

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Background: Co-existence of antineutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) and inflammatory bowel disease (IBD) is quite rare, but dozens of cases have already been reported. We describe an AAV in a patient with enteropathic spondylarthritis previously treated with Tumor Necrosis Factor α inhibitors (TNFi).

Clinical case: A 53-year-old female presented in our department with a 4-week history of malaise, anorexia, nauseas and worsening of her sinusitis symptoms. She had a 5-year history of enteropathic spondylarthritis, with axial and peripheral involvement. In the previous year, she was medicated with infliximab that was suspended after the third infusion due to an exuberant paradoxical palmoplantar psoriasis. A switch to adalimumab was made. Yet, after 16 weeks of therapy, there was a worsening of the psoriatic lesions, and adalimumab was then suspended, with the last administration being given 5 months prior to her admission.

On the current presentation, the patient was chronically medicated with sulfasalazine 2g/day, and in the previous weeks, she was medicated with deflazacort 12 mg/day and celecoxib 100 mg/day. On physical examination, patient was pale and afebrile. Blood tests revealed a microcytic anemia (hemoglobin 8.8 g/dL, medium globular volume 80 fL), elevated erythrocyte sedimentation rate of 110 mm, C-reactive protein of 8.36 mg/dL, and a serum creatinine (sCr) level of 2.11 mg/dL (eGFR of 26.2 ml/min/1.73 m2), which peaked two weeks later to 3.0 mg/dL (eGFR of 17.3 ml/min/1.73 m2).

Urinalysis showed a new onset haematoproteinuria, with urine protein to creatine ratio of 2.6 g/g. pANCA was positive with proteinase 3 (65.6 U/L). The remaining immunological study was negative. Kidney biopsy revealed a pauci-immune crescentic glomerulonephritis and acute interstitial nephritis (probably secondary to non-steroidal anti-inflammatory use). The diagnosis of granulomatosis with polyangiitis (GPA) pANCA positive was made. Treatment included intravenous methylprednisolone pulses, oral prednisolone and rituximab. Four months later, anemia has improved (Hg 11.2 g/dL), inflammatory parameters are normal, and her kidney function is stable, with a sCr level of 1.88 mg/dL (eGFR 29 ml/min/1.73 m2), and urine protein to creatinine ratio decreased to 1.7 g/g.

Discussion: Management of vasculitis in a patient with IBD may be troublesome because it can be difficult to discern if vasculitis is a new entity or an extraintestinal manifestation of the IBD. Moreover, in this case, a previous treatment with TNFi may be a confounding factor, regarding the fact that hundreds of TNFi-induced vasculitis cases have already been reported, and that the patient had previously experienced an exuberant paradoxical psoriasis. However, TNFi was suspended 5 months before of the current condition, and so this hypothesis is less likely, even though it was difficult to specify when symptoms truly began. Furthermore, extraintestinal manifestations of IBD are usually related with intestinal inflammatory activity and the commonest glomerular involvement is IgA nephritis. Yet, in the present case, the patient had a 20-year history of crohn's disease, with no signs

of disease activity in the last few years. Finally, a recent literature review identified 19 AAV patients in a total of 306 patients with IBD and vasculitis where, in most cases, the diagnosis of IBD preceded that of vasculitis by years, and IBD was usually not active at the time of the onset of vasculitis, which has also occurred in the reported case.

CC051 – ARTRITE REUMATÓIDE E DOENÇA RENAL SOB HEMODIÁLISE – ABORDAGEM TERAPÊUTICA COM ANTI-TNF É UMA OPÇÃO?

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Introdução: Os anti-TNF (Tumour Necrosis Factor) são agentes biológicos eficazes no tratamento da artrite reumatóide (AR). Contudo, em doentes com AR e doença renal crónica (DRC) terminal sob diálise existem muito poucos casos reportados sobre a sua eficácia e segurança. Os DMARDs (disease modifying antirheumatic drugs) sintéticos como o metotrexato ou a leflunomida têm um risco aumentado de toxicidade tornando-os pouco seguros nestes doentes. São poucos os casos clínicos descritos relativos à eficácia e segurança dos anti-TNF nos doentes com DRC sob diálise (1-4).
Objetivos e Métodos: Reportamos um caso de uso de etanercept (ETN) numa doente com AR e DRC sob hemodiálise.
Resultados: Mulher de 71 anos com AR positiva para Fator Reumatóide (FR), com cerca de 15 anos de evolução, tratada previamente com Metotrexato que foi interrompido por DRC de estadio terminal, tendo iniciado hemodiálise. A atividade da doença foi controlada durante 2 anos apenas com prednisolona em baixa dose (5mg/dia). Contudo, por agravamento clínico (DAS-28 (Disease Activity Score-28) de 5.25) e desenvolvimento de osteoporose fraturária, decidiu-se modificar a terapêutica para melhor controlo da AR e redução da dose da corticoterapia.
Os anti-TNF são hidrolisados por lisossomas e a sua excreção não parece ser influenciada pela função renal (1,5). Assim, estes agentes constituem uma possível alternativa terapêutica nos doentes com DRC. Existem algumas séries de casos com um número reduzido de doentes que mostram eficácia e segurança do infliximab (2), adalimumab (3) e ETN (4,5) em doentes com DRC sob diálise.
Com base nestes dados decidiu-se iniciar ETN em monoterapia e optou-se pela posologia de 25mg 2x por semana (injeção após sessão de hemodiálise), apesar de Don e colaboradores terem demonstrado que a hemodiálise não afetava a excreção de ETN (5). Aos 6 meses a doente apresenta uma boa resposta (DAS-28 de 3.01) e não se registaram eventos adversos, nomeadamente complicações infeciosas.
Conclusão: Apresentamos o primeiro caso do nosso centro com o uso de um anti-TNF numa doente com DRC sob hemodiálise, com boa resposta aos 6 meses e sem nenhum evento adverso registado. Os anti-TNF parecem ser seguros e eficazes na DRC sob diálise, contudo a experiência ainda é escassa nesta população.

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CC053 – POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME AND DIGITAL GANGRENE IN A PATIENT WITH GRANULOMATOSIS WITH POLYANGIITIS – A RARE CASE REPORT

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Introduction: Granulomatosis with Polyangiitis (GPA) is an antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV), classically associated with the presence of proteinase 3 (PR3)-ANCA. GPA has a wide range of clinical manifestations, involving almost every organ, particularly upper respiratory tract, lungs and kidneys. One of its rarest manifestations is digital ischemia or gangrene that accounts for <1% of cases. Posterior reversible posterior leukoencephalopathy syndrome (PRES) is a syndrome characterized by headache, visual changes, confusion, and seizures, accompanied by distinctive neuroimaging features, which are usually reversible. Rare cases of its association with vasculitis, including GPA, have been described.
Case report: A 64-year-old female was referred for Rheumatology observation at a University Hospital due to a one-week history of a rapidly progressive decrease in strength and sensitivity of the left hand, and 2nd, 4th and 5th left toes, associated with pain, swelling, and digital discoloration. Over the last 18 months, she had been complaining of weight-loss (30% of total body-weight), recurrent fever, arthralgia, purpura in the upper and lower limbs, and progressive decrease of lower limbs' strength for which she underwent nerve biopsy showing vasculitis of the small vessels and was started on 10 mg of prednisolone daily. She denied otorhinolaryngologic and cardiorespiratory symptoms. On physical examination, she was emaciated, with muscular atrophy, and presented purpuric lesions on extremities, necrosis of the 2nd, 4th, and 5th left toes, and necrosis of the 2nd to the 5th left fingers. Cardiopulmonary auscultation and abdominal examination were unremarkable. Laboratory tests showed normocytic normochromic anemia (Hb 7.8 gm/dL), ESR 84 mm/hr, CRP 5.8 mg/dL, creatinine 3.71 mg/dL, active urine sediment with a 24-hour proteinuria of 1106 mg, and positive ANCA-PR3 (390.8 UQ); other relevant autoimmunity screens and laboratory workup for infections were negative. One-week after hospital admission, she developed dyspnea and episodes of diffuse headache with associated confusion. Chest X-Ray and computed tomography scan showed pulmonary infiltrates, with subsequent bronchoscopy consistent with alveolar hemorrhage, and brain MRI revealed findings compatible with PRES. The patient was classified as having GPA4 and treated initially with IV methylprednisolone (three pulses of 1g), followed by oral prednisone (1mg/kg/day) and rituximab (two pulses of 1g). Given the presence of alveolar hemorrhage, digital ischemia and PRES plasmapheresis was additionally started. The patient showed significant improvement of renal and pulmonary function and complete resolution of neurological symptoms; follow-up brain MRI demonstrated resolution of PRES changes. Unfortunately, digital gangrene was irreversible and the patient underwent amputation of the necrotic areas.
Conclusion: PRES and digital ischemia or gangrene are unusual manifestations of GPA, a disease itself quite rare. To the best of our knowledge, only 6 cases of GPA-associated PRES and 17 cases of GPA-associated digital ischemia or gangrene have been reported in the literature, none of which with patients presenting both features. It is crucial that cases of AAV are treated at reference centers with multidisciplinary expertise in the management of these diseases. In the present case, early diagnosis and treatment could have improved disease progression and outcome.

CC063 – POLIARTERITE NODOSA CUTÂNEA – A GRANDE DESCONHECIDA

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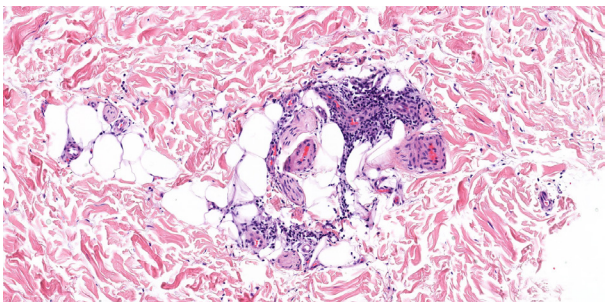
Introdução: A poliarterite nodosa (PAN) é uma vasculite sistémica que afecta a vasos de pequeno e mediano calibre e se caracteriza por envolvimento multi-orgânico. A poliarterite nodosa cutânea (PANc), é um subtipo mais raro que se caracteriza pelas lesões cutâneas, envolvimento articular, muscular e neuropatia periférica e que se distingue pelo mínimo envolvimento sistémico.
Caso clínico: Paciente do sexo masculino de 41 anos que apresenta mialgias, debilidade na cintura escapular/pelviana, lesões papulonodulosas violáceas palpáveis em todo o corpo, incluindo palmas e plantas, artralgias de grandes articulações e clínica de parestesias. O paciente não tem antecedentes de interesse, nega hábitos tóxicos, relações sexuais de risco e contacto recente com animais. Realizam-se várias provas complementarias: hemograma, serologias, bioquímica e estudo de autoimunidade que são normais, destacando unicamente elevação de proteínas de fase aguda. A biopsia cutânea das lesões e estudo de imunofluorescência directa destaca um infiltrado inflamatório na parede de vasos da derme constituído por linfócitos e neutrófilos e depósito de necrose fibrinoide, compatíveis com o diagnóstico de poliarterite nodosa. De forma a excluir envolvimento sistémico, procede-se à realização de análise da urina de 24h, radiografia de tórax e ecografia abdominal que são normais. A espirometria destaca uma leve descida da capacidade de difusão (DLCO) pelo que se decide fazer um TC de toráx que descarta envolvimento pulmonar. O estudo electromiografico destaca

FIGURA 1 – CORTE DE BIOPSIA DE PELE (DERME RETICULAR)



uma polineuropatia axonal sensitiva leve em extremidades inferiores. Iniciamos tratamento com corticoides a doses baixas, melhorando a clínica cutânea e articular mas, persistindo a alteração sensitiva.

Discussão: O nosso paciente apresentou uma forma cutânea de poliarterite nodosa, evidenciada pelas lesões compatíveis com livedo reticularis, mialgias, artralgias e envolvimento neurológico periférico. É importante descartar envolvimento sistémico de forma a fazer um diagnóstico diferencial com a poliarterite nodosa. A progressão da PANc a uma forma sistémica é pouco frequente, no entanto, há estudos que indicam que depois de anos de evolução, é possível e por este motivo, é fundamental fazer um seguimento dos pacientes.

Conclusão: A panarterite nodosa cutânea é uma vasculite benigna que afecta a vasos de pequeno e médio calibre. O diagnóstico é feito a partir da clínica, apoiando-se na biópsia cutânea e excluindo sintomatologia sistémica

CC069 – UM CASO RARO DE SOBREPOSIÇÃO LUPUS ERITEMATOSO SISTÉMICO E VASCULITE ASSOCIADA A ANTICORPO ANTICITOPLASMA DE NEUTRÓFILOS

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Introdução: A síndrome de sobreposição de vasculite associada a anticorpo anticitoplasma de neutrófilos (VAA) e lúpus eritematoso sistémico (LES) (LES/VAA) foi originalmente descrita em 2008. Este quadro raro é muitas vezes caracterizado por uma glomerulonefrite crescêntica agressiva em doentes que partilham critérios de classificação com LES e com VAA nomeadamente artrite, envolvimento cutâneo e renal e positividade para o anticorpo anticitoplasma de neutrófilos (ANCA).

Caso clínico: Mulher de 55 anos, com o diagnóstico de LES há 19 anos no contexto de rash malar típico, artralgias de ritmo inflamatório, anemia normocítica normocrómica e trombocitopenia discretas, ANA 1/1000 homogéneo e anticorpos anti-dsDNA positivos (acima do dobro do limite superior do normal). Previamente em remissão clínica, medicada com prednisolona 5 mg por dia, tendo interrompido hidroxicloroquina por intolerância gastrointestinal.

Assintomática até há 2 semanas, apresentou quadro de urina espumosa, negando outras alterações genitourinárias ou de outros orgãos ou sistemas. Colheu estudo analítico revelando anemia normocítica normocrómica (hemoglobina de 11.4 g/dL), lesão renal aguda (creatinina de 1.95 mg/dL para um valor prévio de 0.73 mg/dL e ureia de 73 mg/dL), elevação de velocidade de sedimentação (VS) de 68 mm/1ªh e de proteína C reativa (PCR) de 28.3 mg/L; colheu urina de 24h que mostrou proteinúria de 578 mg e leucoeritrocitúria. Não apresentava consumo de complemento e os anticorpos anti-dsDNA eram discretamente positivos (40 UI/mL para um normal inferior a 30). É internada em Reumatologia para estudo do quadro.

No internamento apresentou agravamento da função renal (creatinina máxima de 3.11 mg/dL), com manutenção da anemia. Realizou ecografia renovesical que excluiu foco obstrutivo e que revelou aumento da diferenciação corticomedular por aumento da ecogeneicidade cortical, em relação com provável nefropatia. Do restante estudo imunológico de ressalvar positividade para os ANCA relacionados com a mieloperoxidase (ANCA-MPO) de 69 UI/mL (para um valor normal inferior a 20), prova de Coombs e inibidor lúpico negativos. Realizou biópsia renal que mostrou 13 glomérulos esclerosas e 10 com crescentes (2 crescentes celulares e 8 crescentes fibrocelulares), sem necrose fibrinóide, focalmente com esclerose segmentar. No interstício apresentava infiltrado linfoplasmocitário e fibrose com marcação pela imunofluorescência para IgA, C3c, C1q e IgM mesangial. Foram entendidas como alterações a favor de vasculite com sinais de cronicidade o que, em conjunto com o achado da positividade para ANCA MPO, permitiu o diagnóstico de vasculite relacionada com ANCA MPO, num provável contexto de LES/AAV. A doente foi pulsada com metilprednisolona 1g/dia por 3 dias, tendo iniciado ciclofosfamida e prednisolona per os, segundo o protocolo CYCLOPS.

Apresentou uma boa evolução, em redução gradual da dosagem de prednisolona (atualmente de 17.5 mg/dia). Após a terceira administração de ciclofosfamida, a doente apresentava melhoria da função renal (creatinina de 1.98 mg/dL), VS normal, PCR de 5.1 mg/L; urina tipo 2 sem leucoeritrocitúria e ratio proteínas–

creatinina em urina tipo 2 de 327 mg/g.
Conclusão: O caso de glomerulonefrite rapidamente progressiva apresentado parece corresponder a um síndrome de sobreposição de LES/AAV. Mais estudos são necessários para melhor caracterizar esta síndrome e investigar a possível etiopatogenia imune compartilhada.

CC070 – DOENÇA DE STILL DO ADULTO COMPLICADA COM PNEUMONIA ORGANIZATIVA

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Introdução: A doença de Still de adulto (DSA) é uma doença rara, com envolvimento pulmonar descrito em cerca de 30% dos doentes. Apesar disso e, ainda que a pneumonia organizativa (PO) esteja associada a doenças do tecido conjuntivo, a associação com a DSA foi raramente descrita.
Caso clínico: Mulher de 29 anos, assintomática até há 2 semanas, altura em que inicia quadro de picos febris diários vespertinos, rash cutâneo evanescente não pruriginoso nas extremidades com relação com os picos febris e poliartralgias de ritmo inflamatório. De história familiar salienta-se irmã com artrite idiopática juvenil. No serviço de urgência é objetivada artrite da 3ª interfalângica proximal direita e tem alta com anti -inflamatório não esteróide (AINE) fixo por período de 8 dias. Foi reavaliada em consulta de Reumatologia após 2 semanas e, por agravamento do quadro e tosse seca de novo associada a dispneia progressiva para médios esforços, é internada para estudo. Analiticamente mostrou, de relevo, anemia normocítica normocrómica discreta de 11.4 g/dL, elevação da proteína C reativa de 165.1 mg/L, Velocidade de Sedimentação de 86 mm/1ªh e ferritina de 1416.6 ng/mL, não apresentando leucocitose ou trombocitose. Do estudo imunológico colhido: anticorpos anti nucleares, anticorpos anti-antigénios extraídos do núcleo, fator reumatóide e anti-dsDNA negativos. Do estudo de causa neoplásica: citologia cervicovaginal, mamografia, ecografia tiroideia e proteinograma sem alterações e a ecografia abdominal excluiu a presença de hepatoesplenomegalia ou outras massas identificáveis. Do estudo de causa infeciosa: ecocardiograma e exame sumário de urina sem alterações, hemoculturas negativas, pesquisa de RNA dos virus influenza A e B negativos, serologias negativas da sífilis, HIV, HBV e HCV e negativas para infeção atual para CMV, EBV, HSV1 e HSV2. Atendendo a IgM duvidoso para Parvovirus B19 e de Mycoplasma pneumoniae, foram colhidas pesquisas de DNA dos microorganismos que se revelaram negativas. Foram realizadas colheitas para micobacteriológico no aspirado gástrico e também no lavado broncoalveolar (LBA), negativas. Foi realizada uma tomografia computorizada torácica que mostrou no lobo inferior direito uma consolidação com broncograma aéreo, com focos de densificação em vidro despolido. No sentido de esclarecer a hipótese de pneumonia organizativa versus infeciosa foi realizado LBA que revelou alveolite linfocítica intensa e neutrofílica, com relação CD4/CD8 elevada (2.21) e biópsia pulmonar que mostrou rolhões de fibroblastos e tecido conjuntivo projetando-se para os alvéolos e focos de linfócitos, plasmócitos e histiócitos intra-alveolares, por vezes em localização intersticial, compatíveis com a suspeita clínica de PO.
Por recrudescência do quadro de artrite das pequenas articulações das mãos, rash cutâneo e manutenção de um pico febril diário, assumiu-se o diagnóstico de DSA, tendo iniciado tratamento com prednisolona (dose máxima de 20 mg) em esquema de redução gradual e metotrexato (dose actual de 15 mg/semanal). Atualmente melhorada do ponto de vista articular e pulmonar, com resolução da artrite, do quadro febril e em melhoria radiográfica.
Conclusão: A DSA é sobretudo um diagnóstico de exclusão, neste caso dificultado por um quadro respiratório que implicou o estudo exaustivo de causa infeciosa. Trata-se de um raro caso de DSA complicado com PO, que alerta para a necessidade de investigação diagnóstica exaustiva nestes doentes e que poderá justificar uma escalada terapêutica mais precoce.

CC072 – FRATURA FEMORAL DE BAIXO IMPACTO – MAIS UM CASO DE FRATURA OSTEOPORÓTICA?

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Introdução: As fraturas femorais atraumáticas ou causadas por traumas de baixo impacto incluem as fraturas de insuficiência e patológicas mas também fraturas atípicas, resultantes da alteração da normal via de remodelação óssea. São, sobretudo, descritas em doentes submetidos a tratamentos prolongados com fármacos que suprimem a atividade osteclástica como é o caso dos bifosfonatos e do denosumab.
Caso clínico: Homem de 65 anos, com anteceden-

tes de Osteoporose, sem fraturas prevalentes prévias conhecidas, e doença renal crónica estadio 3a. Apresentava baixo aporte de cálcio diário mas negava consumo tabágico ou consumo alcoólico médio superior a 3 unidade de álcool/dia. Negava corticoterapia crónica, negava fratura do colo do fémur dos pais ou imobilidade prolongada nos últimos 6 meses. Estava medicado com alendronato há 20 anos, tendo sido reavaliado com densitometria Hologic há cerca de 18 meses que revelava um T score no colo femoral de -1.9 e na coluna lombar de -2.9. Recorre ao ao serviço de urgência do nosso centro hospitalar por dor intensa referida à região trocantérica esquerda na sequência de queda de baixo impacto. Realizou radiografias do fémur esquerdo e da bacia que revelaram traço de fratura na diáfise femoral, distal ao pequeno trocanter com orientação sobretudo transversa lateralmente e com orientação mais oblíqua medialmente, formando um "spike" na região do córtex medial. Foi submetido a osteossíntese da fratura e teve alta orientado para consultas de fisiatria e reumatologia (segundo protocolo de avaliação de fraturas ósseas de fragilidade do nosso centro). Na consulta de Reumatologia é levantada a hipótese de fratura atípica pelas características radiográficas típicas e pela história clínica sugestiva. Foi feita biópsia óssea, com marcação dupla com tetraciclina, que demonstrou superfícies osteoblástica e osteoclástica francamente reduzidas, bem como superfície osteóide praticamente inexistente associada uma baixa taxa de formação óssea, achados que, no seu conjunto, são compatíveis com o diagnóstico de doença óssea de baixa remodelação/osso adinâmico.
O estudo analítico não apresentava alterações, nomeadamente do cálcio ionizado e fósforo inorgânico, 25-OH-vitamina D que era de de 35 ng/mL e da fosfatase alcalina (62 U/L para um intervalo de normalidade entre 30 e 120 U/L), paratormona (26.7 pg/mL para um intervalo de normalidade entre 10.0 e 65.0 pg/mL) e marcadores de remodelação óssea (beta-crosslaps 0.17 ng/mL e osteocalcina 18 ng/mL).
Realizou radiografias da coluna vertebral dorsal e lombar que não revelaram fraturas vertebrais.
Foi decidido iniciar tratamento com teriparatide uma administração subcutânea diária e suplementação de cálcio e vitamina D., mantendo a terapêutica (atualmente no 4º mês) e seguimento em consulta de Reumatologia.
Conclusão: Embora a evidência apoie claramente o efeito benéfico dos bisfosfonatos na prevenção de fraturas osteoporóticas, existe uma preocupação de que a terapia prolongada possa levar à supressão da remodelação óssea, com os resultados da maioria dos estudos observacionais mostrando um aumento no risco de fratura atípica nestes doentes. Estudos mais robustos são necessários para esclarecer qual a duração de tratamento com bifosfonato ideal e qual a orientação terapêutica destes doentes, que é também controversa. Este caso reforça a importância da reavaliação regular do risco fraturário e do tratamento anti-osteoporótico em curso, mas também para a relevância de ponderar os diagnósticos diferenciais de uma fratura de baixo impacto, pela implicância terapêutica associada.

CC078 – URTICÁRIA CRÓNICA NO LÚPUS ERITEMATOSO SISTÉMICO TRATADA COM COLQUICINA

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Introdução: A urticária crónica (UC) é caracterizada pela recorrência de lesões de urticária e/ou angioedema, num período de mais de 6 semanas, sem um desencadeante específico. O Lúpus Eritematoso Sistémico (LES) é uma doença imunomediada que pode afetar qualquer órgão incluindo a pele, podendo a UC constituir uma manifestação cutânea inespecífica de LES. A primeira linha de tratamento da UC são os anti-histamínicos, sendo, contudo, muitas vezes, uma condição de difícil controlo.
Objetivo: Apresentação do caso de uma doente com LES e UC em que as manifestações cutâneas do LES e a UC foram completamente controladas com a colquicina.
Caso clínico: Mulher de 47 anos, acompanhada em consulta de Reumatologia desde 2008 por LES com envolvimento cutâneo, articular e hematológico. Associadamente, em 2014, foi estabelecido o diagnóstico de UC (avaliada por Dermatologia e Imunoalergologia). A medicação com hidroxicloroquina na dose de 6mg/kg/dia, metotrexato 20mg/semana, prednisolona 5 mg/dia e levocetirizina 5mg/dia mantinha-a assintomática do ponto de vista articular e hematológico, continuando, no entanto, com episódios recorrentes de urticária, sem resposta a montelucaste, obrigando a doses mais elevadas de corticóides durante os episódios. Em maio de 2018, a terapêutica com hidroxicloroquina foi suspensa por toxicidade ocular, tendo-se verificado agravamento das lesões cutâneas com esta suspensão. Em agosto de 2018, iniciou terapêutica com colquicina, 1mg/dia, mantendo a restante tera-

pêutica. Desde então, não se verificou qualquer recidiva das lesões de urticária nem surgimento de outras manifestações de atividade do LES.
Discussão/Conclusão: As lesões de UC são de difícil tratamento e, após falência a anti-histamínicos, é recomendado o uso de terapêuticas imunossupressoras como a ciclosporina A e o omalizumab. Estes fármacos, de efeitos adversos não negligenciáveis, seriam de utilização mais difícil neste caso, pelo fato de a doente estar sob tratamento com outros imunossupressores. Há já alguma evidência de que a colquicina possa ter um papel no tratamento da UC, salientando-se que, normalmente, é uma terapêutica bem tolerada.

CC079 – ARTRITE REATIVA ASSOCIADA A ARTRITE SÉTICA POR STREPTOCOCCUS AGALACTIAE

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Introdução: A artrite sética é uma emergência médica, com uma mortalidade estimada em 11%.1 O Streptococcus agalactiae (Streptococcus do grupo B) é uma causa rara de artrite sética em adultos.1,2 A artrite reativa é atualmente integrada no espectro das espondilartrites, caracterizando-se pela presença de uma sinovite estéril, secundária a uma infecção distante, habitualmente do trato gastrointestinal ou geniturinário, sendo a Chlamydia trachomatis e as enterobacteriaceae os microorganismos mais frequentemente envolvidos.3 Os Streptococcus do grupo B, normalmente associados a infeções cutâneas, são uma causa rara de artrite reativa.4
Objetivo: Apresentação do caso de uma artrite reativa a artrite sética por Streptococcus agalactiae.
Caso clínico: Homem de 53 anos, com antecedentes de lúpus cutâneo discóide crónico, hipertensão arterial, dislipidemia e obesidade, avaliado no Serviço de Urgência por febre e artralgias, referenciado para observação no serviço de Reumatologia por suspeita de envolvimento sistémico. Apresentava artrite do joelho esquerdo e sinais inflamatórios francos no dorso da mão esquerda. Na avaliação ecográfica, foi constatado derrame septado de grande volume do joelho esquerdo e edema do tecido celular subcutâneo do dorso da mão esquerda, compatível com celulite. A artrocentese do joelho revelou líquido sinovial (LS) de aspeto purulento, hipercelular e sem cristais, tendo o doente sido internado no serviço de Ortopedia por suspeita de artrite sética, para realização de artrotomia. Analiticamente, apresentava anemia ligeira normocítica e normocrómica (hemoglobina: 12,8 mg/dL), leucocitose com neutrofilia, e aumento marcado dos parâmetros inflamatórios (proteína C reativa de 10,77 mg/dL e velocidade de sedimentação de 63 mm/h). As hemoculturas e as culturas do LS foram positivas para Streptococcus agalactiae sensível à antibioterapia em curso (ceftriaxone e vancomicina).
O doente apresentou apenas melhoria clínica transitória, com recidiva da artrite, pelo que foi submetido a mais duas artrotomias do joelho esquerdo, com lavagem repetida e colocação de dreno (3 e 10 dias após a primeira intervenção).
Ainda sob antibioterapia com ceftriaxone e vancomicina (14º dia), desenvolveu artrite do punho esquerdo e da tibiotársica esquerda. Foi reavaliado por Reumatologia, tendo-se confirmado, ecograficamente, a presença de artrite destas duas articulações. A artrocentese ecoguiada da articulação tibiotársica revelou também um líquido amarelo turvo, hipercelular, sem cristais, amicrobiano, tendo sido colocada a hipótese de artrite reativa. O doente veio a melhorar com a instituição de anti-inflamatório não esteroide e prednisolona na dose de 30 mg/dia em esquema de desmame durante 8 semanas, com resolução do quadro e sem recidiva da sintomatologia, corroborando a hipótese de quadro de artrite reativa à artrite sética e à bacteriemia por Streptococcus agalactiae.
Discussão/Conclusão: A raridade de uma artrite sética por Streptococcus agalactiae e de artrite reativa após uma artrite sética, justificam a apresentação do caso. Salienta-se a importância da intervenção multidisciplinar na observação e orientação dos doentes com patologia musculosquelética. Esta intervenção multidisciplinar no caso descrito permitiu o diagnóstico e correto tratamento da artrite sética, evitando o dano estrutural e da artrite reativa, evitando o prolongamento de antibioterapia e potenciais intervenções cirúrgicas.

CC089 – ARTROPATIA NA HEMOCROMATOSE HEREDITÁRIA

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Introdução: A Hemocromatose hereditária (HH) é uma doença metabólica autossómica recessiva, caracterizada pela sobrecarga de ferro sistémico e que, em

90% dos casos, associa-se a uma mutação em homozigotia C282Y no gene HFE. A artropatia associada à HH é uma alteração clínica comum e tanto pode ser a primeira manifestação da doença como surgir após início do tratamento. As queixas articulares na HH podem estar relacionadas com a deposição de hemossiderina e/ou com a formação de cristais de pirofosfato de cálcio, condição frequentemente associada.
Objetivos: Relatar a experiência clínica acumulada no seguimento dos doentes com artropatia associada à HH num Serviço de Reumatologia.
Métodos: Avaliação retrospetiva, com análise de registos clínicos, de todos os doentes com artropatia associada à HH observados nos últimos 8 anos. No total foram identificados 4 doentes com artropatia associada à HH, dois como manifestação inaugural e dois no decurso da doença.
Resultados: Dos 4 doentes com HH incluídos, todos eram do sexo masculino. O primeiro doente, 56 anos, foi referenciado à consulta de Reumatologia por quadro de poliartralgias de ritmo misto (punhos, pequenas articulações das mãos e tibiotársicas) com meses de evolução e pouco alívio com o recurso a anti-inflamatórios não esteroides. Associadamente referia diminuição da líbido e tinha história familiar (irmão) de hemocromatose, sob tratamento regular. Um segundo doente, 59 anos, referenciado à consulta de Reumatologia por quadro de poliartralgias de ritmo misto (punhos, pequenas articulações das mãos e ombros). Em ambos os doentes, foi objetivada tumefação e limitação da flexão da segunda e terceira metacarpofalângicas bilateralmente. Na radiografia das mãos, verificou-se a presença de "osteófitos em anzol" na segunda e terceira metacarpofalângicas, esclerose subcondral e quistos subcondrais assim como condrocalcinose no ligamento triangular do carpo. O estudo analítico documentou elevação marcada da ferritina (superior a 2x o normal), com saturação de transferrina de 97% e de 78%, respetivamente, sem outras alterações, nomeadamente alterações da função hepática, glicémia e o estudo imunológico foi negativo. O estudo genético confirmou homozigotia para C282Y e foi iniciado o tratamento. O terceiro e quarto doentes, de 50 anos e de 62 anos, respetivamente, com história clínica de HH sob tratamento com flebotomias , foram enviados à consulta de Reumatologia por início de artralgias de ritmo misto, com envolvimento das pequenas articulações das mãos (metacarpofalângicas e interfalângicas proximais). Objetivamente, verificou-se tumefação das segunda e terceira metacarpofalângica bilateralmente. Radiologicamente, constatou-se a presença de "osteófitos em anzol" e esclerose subcondral na mesma localização. Do estudo analítico não houve qualquer alteração de relevo. Foi iniciado tratamento sintomático com anti-inflamatório não esteróide com boa resposta clínica.
Conclusão: Apesar dos achados clínicos e radiológicos serem muitas vezes sugestivos da artropatia da HH, esta não é patognomónica da HH, o que pode dificultar o diagnóstico diferencial. Tratando-se de uma doença potencialmente devastadora, com uma elevada taxa de morbimortalidade, o diagnóstico precoce e o tratamento atempado da HH são essenciais. O tratamento sintomático com anti-inflamatórios não esteroides e outros analgésicos pode nos casos leves a moderados provocar um alívio franco dos sintomas.

CC092 – ENDOCARDITE INFECCIOSA SUBAGUDA – APRESENTAÇÃO ATÍPICA

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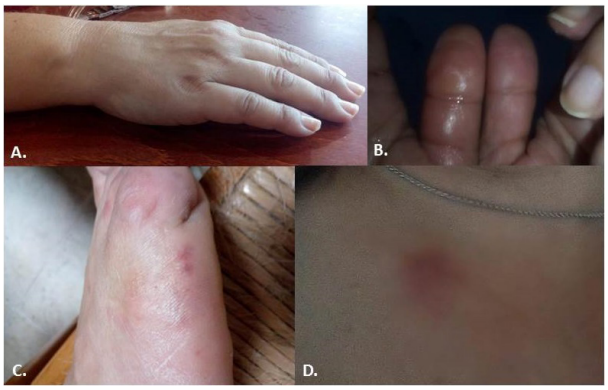
Introdução: A endocardite é uma infecção que afecta o endocárdio, habitualmente ao nível das válvulas ou de dispositivos intracardíacos. A manifestação clínica mais frequente é a febre.
Caso Clínico: Mulher de 44 anos, com antecedentes de febre reumática na infância e consequente doença reumática das válvulas aórtica e mitral, submetida a valvuloplastia com próteses mecânicas e hipocoagulada com varfarina. Foi enviada à consulta de Reumatologia por artralgias inflamatórias. À observação referia aparecimento de queixas migratórias desde há um mês: inicialmente dor no bordo lateral dos pés acompanhado de edema e de lesões cutâneas punctiformes, seguida de dor e tumefacção ao nível da 5ª falange distal esquerda e posteriormente com dor e tumefacção do punho com reaparecimento de lesões punctiformes dolorosas ao nível das polpas digitais. Associadamente descrevia hipersudorese diurna e nocturna, sem noção de emagrecimento e sem objectivação de febre ou outros sintomas sugestivos de conectivite. Ao exame objectivo constatado apenas edema e rubor da falange distal do 5º dedo esquerdo e tumefacção do maléolo externo esquerdo. Analiticamente apresentava velocidade de sedimentação de 92 mm/h, PCR de 7.27 mg/dL, factor reumatóide e anti-peptídeos cíclicos citrulinados negativos, anticorpos anti-nucleares de 1/160 com padrão mosqueado (imunoblot negativo), enzima de conversão da angiotensina negativa, serologias de Borrelia e Coxiella, pesquisa de sífilis

FIGURA 1 – MANIFESTAÇÕES CLÍNICAS APRESENTADAS PELA DOENTE



e marcadores víricos (HIV, HBV e HCV) negativos. O estudo radiográfico mostrou alterações degenerativas. Na segunda observação apresentava pequenas lesões punciformes arroxeadas nos pés pelo que foi complementado o estudo para excluir contexto infeccioso, vasculítico e paraneoplásico. Realizou TC toraco-abdomino-pélvico que mostrou foco de realce nodular com cerca de 10 mm, centrado à mucosa da face lingual da valécula direita, nódulo sólido tiroideu com cerca de 30 mm e esplenomegalia com áreas de hipoperfusão, à periferia do baço, em cunha, a sugerir focos de enfarte. Na terceira avaliação a doente não apresentava alterações no exame objectivo mas trazia fotografias com artrite do punho direito (1A), dactilite do 1º dedo da mão direita (1B), lesões sugestivas de nódulos de Osler (1C) e pequenas pápulas eritematosas migratórias e dolorosas (1D). Colheu nesse dia 2 sets de hemoculturas com isolamento (1 em 4) de microorganismo do grupo HACEK (Aggregatibacter actinomycetemcomitans) e fez ecocardiograma transesofágico: "sem imagens inequívocas de vegetações protésicas ou nas estruturas valvulares nativas". Foi assumido o diagnóstico de endocardite infecciosa subaguda, 3 meses após o início das queixas, tendo sido internada e medicada com ceftriaxone 2g e.v que cumpriu durante 6 semanas. Durante o internamento realizou biópsia do nódulo tiroideu que revelou lesão folicular de significado indeterminado e foi observada por Otorrinolaringologia que excluiu a existência de neoformação. Foi avaliada por Estomatologia que detectou cárie no dente 46, considerado como provável ponto de partida da endocardite, tendo-se procedido à sua exodontia. Após antibioterapia apresentou resolução do quadro músculo-esquelético e cutâneo com normalização dos parâmetros inflamatórios.

Conclusão: Este caso destaca-se pelo facto da endocardite infecciosa ter uma apresentação subaguda, com manifestações clínicas pouco comuns como artrite, tumefacção dos tecidos moles e lesões cutâneas migratórias. A colaboração da doente foi essencial no diagnóstico pois o exame objectivo era sempre muito fruste e inespecífico.

CC102 – ACQUIRED HAEMOPHILIA A, HAEMOLYTIC ANAEMIA, TYPE 1 DIABETES MELLITUS AND AUTOIMMUNE HYPOTHYROIDISM IN A SYSTEMIC LUPUS ERYTHEMATOSUS PATIENT

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Introduction: Acquired haemophilia A (AHA) is a rare disease caused by the spontaneous development of autoantibodies against endogenous factor VIII (FVIII). [1-6] Although most cases are idiopathic, AHA can be associated with pregnancy, malignancy and immune-mediated diseases, including Systemic Lupus Erythematosus (SLE). The authors describe a SLE patient who developed AHA due to the presence of a FVIII inhibitor (FVIII-i). Combination therapy with steroids and rituximab was effective in suppressing and controlling AHA.

Clinical case: A 56-year-old woman was admitted due to several spontaneous subcutaneous hematomas. Her medical history included a diagnosis of SLE 10 years ago with mucocutaneous, articular, immunologic (ANA >1/1000 homogeneous pattern, positive anti-Sm antibody) and hematologic (haemolytic anaemia) involvement, treated with hydroxychloroquine (HCQ) and azathioprine. One year after disease onset, she developed class IV lupus nephritis and received steroids and intravenous (IV) cyclophosphamide, followed by maintenance with mycophenolate mofetil (MMF). Due to remission, MMF was progressively reduced and stopped 1 month before current presentation, maintaining HCQ. The patient received no new medications and family history was negative for bleeding diathesis. Blood counts showed haemoglobin of 7.4 g/dL and normal platelets. Further laboratory work-up revealed a positive antiglobulin test, negative dsDNA, low complement levels, ESR 91 mm/h, CRP 20 mg/L. Haptoglobin <8 mg/dL (50-320), decreased reticulocyte production index (1.54) and normal lactate dehydrogenase and bilirrubins. Serum glucose 478 mg/dL (75-110), HbA1c 10.5%, anti-GAD antibodies 99.6 U/mL (N<1.5) and C-Peptide 0.05 ng/mL (N 1.10-4.40). Free T4 0.57 ng/dL (N 0.7-1.48), TSH

37.21 U/mL (0.35-4.94), anti-peroxidase antibody 483 U/mL (N<5.6). Renal and liver function were unremarkable, with no haematoproteinuria. Coagulation tests revealed a significantly prolonged activated partial thromboplastin time (aPTT) of 80.2 seconds (N 24.2-36.4) with no correction upon 1:1 (vol:vol) mixing of the patient with normal human plasma. Lupus anticoagulant was negative. FVIII was 0 U/mL (N 0.7-1.5). A Nijmegen-modified Bethesda revealed a FVIII-i of 10 Bethesda units. These findings clearly establish the diagnosis of AHA in a patient with SLE. Moreover, a diagnosis of autoimmune hypothyroidism and T1DM were concomitantly made. The patient received prednisolone 1 mg/Kg/day and IV rituximab 1000 mg fortnightly, along with levothyroxine and insulin. 2 months after initial presentation, coagulation tests were normal and FVIII-i disappeared.

Conclusion: We describe an unusual case of AHA ten years after SLE onset, during a period of inactivity, with sudden-onset of an autoimmune storm. Due to high cumulative dose of cyclophosphamide, we opted for rituximab.This is a rare but potentially severe and under-recognized manifestation in SLE patients. Rheumatologists must maintain a high index of suspicion in those presenting with bleeding or an isolated prolonged aPTT and negative lupus anticoagulant.

CC117 – EFICÁCIA DO TOCILIZUMAB NO TRATAMENTO DA DOENÇA DE STILL DO ADULTO – EXPERIÊNCIA DE UM CENTRO

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Introdução: A doença de Still do adulto é uma doença rara de etiologia auto-inflamatória caracterizada por febre, rash cutâneo evanescente, leucocitose com neutrofilia e sintomatologia articular inflamatória. Acompanha-se de síndrome bio-inflamatório exuberante e hiperferritinemia marcada. Na sua base fisiopatológica, as interleucinas (IL) 1 e 6 têm um papel de destaque. O tratamento usual passa por anti-inflamatórios esteroides e não-esteróides, havendo também lugar ao uso de disease modifying anti-rheumatic drugs clássicos e biotecnológicos, indicados no caso de refractariedade ou dificuldade no desmame aos corticoesteróides. Se os inibidores da IL1 encontram já aprovação, o papel dos inibidores da IL6 como o tocilizumab encontra-se menos estabelecido, com cerca de 160 casos descritos na literatura. Apresenta-se a experiência do nosso centro no tratamento da doença de Still do adulto com tocilizumab, correspondente a dois casos clínicos com apresentações e cursos clínicos diferentes.

Casos clínicos:

Caso 1: doente do sexo feminino, com 59 anos de idade; observada pela primeira vez com sintomas de febre, rash evanescente, odinofagia e oligoartrite crónica e erosiva dos punhos, acompanhada de síndrome bio-inflamatório e hiperferritinemia; tratada inicialmente com sucesso com corticoesteróides na dose de 1mg/Kg e metotrexato até 15mg/semana; verificou-se, com desmame de corticoterapia, recidiva do quadro de artrite, envolvendo punhos, joelhos e tornozelos; iniciou tocilizumab na dose de 8mg/Kg/mês com melhoria franca de ambos os componentes de doença – sistémica e articular – e possibilidade de desmame de prednisolona até 2.5mg/dia; mantém resposta satisfatória, sem sintomatologia de relevo ou evidência analítica de actividade inflamatória após 12 meses de tratamento, sem registo de efeitos laterais até à data.

Caso 2: doente do sexo feminino, com 25 anos de idade; diagnóstico de doença de Still na sequência de surgimento de febre, rash típico, poliartrite simétrica de pequenas articulações (artrite reumatóide-like) e hiperferritinemia, com curso policíclico. O quadro foi inicialmente responsivo à terapêutica com corticoesteróides na dose de 1mg/Kg e metotrexato até 20 mg/semana mas verificou-se flare sistémico (febre, rash e leucocitose neutrofílica) com a redução da corticoterapia; iniciou tocilizumab na mesma dose do caso 1, sendo possível, aos 6 meses de terapêutica, desmame de prednisolona até 2.5mg em dias alternados, com manutenção do controlo das componentes sistémica e articular de doença, verificando-se também normalização de ferritina e parâmetros inflamatórios,também neste caso, com boa tolerância.

Conclusões: Os casos descritos demonstram as duas "faces" da doença de Still: articular e sistémica. Em concordância com a (escassa) evidência encontrada na literatura sobre o papel da inibição da IL6 no tratamento da doença, a experiência do nosso centro é a de eficácia do tocilizumab no controlo de ambas as componentes desta condição. De facto, em ambos os casos – o primeiro com flare predominantemente articular e o segundo com flare sistémico – para além de um controlo de sintomas articulares, foi obtido um controlo sustentado de actividade inflamatória sistémica clínica e analítica.

CC119 – TERAPÊUTICA BIOTECNOLÓGICA EM DOENTE COM ARTRITE PSORIÁTICA APÓS ESPONDILODISCITE INFECCIOSA SOB ANTI-TNFA – EXPERIÊNCIA COM ABATACEPT

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Introdução: A artrite psoriática (AP) é uma artropatia inflamatória heterogénea, podendo envolver articulações periféricas e esqueleto axial, de múltiplas formas. No seu tratamento, quando as terapêuticas imunomoduladoras clássicas são insuficientes, estão aprovados vários fármacos biotecnológicos com diferentes mecanismos de acção. As complicações infecciosas destes fármacos são os efeitos laterais mais abordados na consulta do doente sob estas terapêuticas. De entre os fármacos biotecnológicos disponíveis, a evidência proveniente do tratamento da artrite reumatóide sugere que o abatacept (ABT) tenha um dos perfis mais seguros no que toca ao risco infecioso. O caso apresentado ilustra problemas do diagnóstico diferencial de lombalgia num doente com AP conhecida e explora as alternativas terapêuticas nesta patologia de acordo com os respectivos perfis de eficácia e segurança.
Caso clínico: Homem de 56 anos de idade, com diagnóstico de AP desde 2004, com envolvimento periférico predominante (articulações tibiotársicas e articulações dos pés, com dactilites). Até 2014, foi medicado com vários esquemas de imunomoduladores clássicos, incluindo metotrexato (MTX), leflunomida (LEF) e sulfassalazina, com controlo global da actividade da doença. Nessa altura, por recidiva de actividade inflamatória, apesar da associação de MTX e LEF, iniciou terapêutica biotecnológica com adalimumab (ADA), atingindo remissão num curto espaço de tempo.
Em 2017, iniciou queixas de lombalgia inflamatória insidiosa, com alterações imagiológicas de discite que foi, inicialmente, interpretada no contexto de actividade axial da AP, dada a ausência de alterações clínicas, analíticas ou imagiológicas que apontassem para outra etiologia, corroborada pela negatividade dos exames culturais de produtos biológicos (incluindo material de biópsia guiada por TAC do disco intervertebral envolvido). Contudo, a evolução arrastada e desfavorável, com posterior surgimento de febre e sintomas constitucionais, motivou repetição de vários exames e conduziu ao diagnóstico de espondilodiscite infecciosa por Staphylococcus aureus, complicada com abcesso local. Foi supensa toda a terapêutica imunomoduladora e esteve internado sob antibioterapia endovenosa prolongada, com evolução favorável.
Dois meses após suspensão da terapêutica, teve recidiva do quadro de artrite dos tornozelos e dactilite de vários dedos dos pés, reiniciando nessa altura MTX, com resposta insuficiente e necessidade de pequenas doses de corticoesteróides (CCT) e de anti-inflamatórios não-esteróides (AINEs). Até ao início de 2019 não foi reiniciada terapêutica biotecnológica pelo risco infeccioso e relutância do próprio doente. Nessa altura, por persistência de atividade inflamatória e após discussão com o doente, optou-se por iniciar ustecinumab, que cumpriu durante 9 meses sem eficácia. Fez switch para ABT em Setembro de 2019 e, com 6 meses de terapêutica, verifica-se remissão completa da artrite dos tornozelos e das dactilites, tendo sido possível a suspensão de CCT e AINE.
Conclusões: O caso descrito ilustra bem a necessidade de estar alerta para complicações infecciosas em doentes sob terapêutica biotecnológica, que podem ter um curso mais indolente e, por isso, mais difíceis de diagnosticar. O surgimento de uma complicação infecciosa grave motivou a exploração de terapêuticas biotecnológicas alternativas aos mais utilizados inibidores do TNFa. O ABT, escolhido pelo seu perfil de segurança mais favorável e apesar de ser pouco utilizado nesta indicação, revelou-se eficaz no controlo da actividade periférica da AP.

CC120 – POLIARTRITE CRÓNICA DE PEQUENAS ARTICULAÇÕES, FACTOR REUMATÓIDE POSITIVO – SERÁ SEMPRE ARTRITE REUMATÓIDE?

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Introdução: A artrite reumatóide (AR) caracteriza-se classicamente por um quadro de poliartrite crónica simétrica e aditiva de pequenas articulações. A positividade do factor reumatóide e/ou anticorpos anti -peptídeos citrulinados tem, nesse contexto, valor diagnóstico e prognóstico importantes. Perante um quadro clínico "clássico", o diagnóstico pode parecer linear. No entanto, os detalhes da história clínica e a evolução clínica podem trazer surpresas. De facto, outras doenças reumáticas podem ter um padrão de envolvimento articular semelhante ao da AR e várias condições, para além desta artropatia, podem conduzir à positividade de factor reumatóide.
Caso clínico: Apresenta-se o caso clínico de um homem de 64 anos de idade referenciado à consulta de Reumatologia por artralgias inflamatórias de pequenas articulações da mão e pés e surgimento

progressivo de tumefações subcutâneas localizadas nos cotovelos e superfícies extensoras do punho e articulações metacarpofalângicas. Destcavam-se antecedentes de neoplasia prostática, hipertensão arterial, dislipidemia e hiperuricemia. Na primeira avaliação, o doente apresentou-se com um quadro de poliartrite crónica envolvendo articulações metacarpofalângicas, punho e cotovelos. Na anamnese, referia episódios prévios sugestivos de monoartrite recorrente e autolimitada dos membros inferiores, de longa data, incluindo episódios sugestivos de podagra. O estudo analítico, revelou anemia, elevação de parâmetros inflamatórios, positividade de factor reumatóide em alto título (173 UI), bem como uma uricemia de 8.7mg/dL. Pelo quadro de poliartrite crónica, realizou rastreios para possível início de imunomodulador, e foi medicado com anti-inflamatório não-esteróide, verificando-se subsequente regressão dos sinais inflamatórios. A avaliação ecográfica músculo-esquelética posterior demonstrou que todas as tumefações identificadas eram compatíveis com tofos gotosos e revelou ainda sinal de duplo-contorno em várias articulações envolvidas. Concluiu-se pelo diagnóstico de artropatia gotosa tofácea com curso crónico e poliarticular.

Conclusões: O caso clínico descrito demonstra a importância da valorização da anamnese e, sobretudo, da evolução dos quadros sintomáticos para um diagnóstico correcto numa especialidade como a Reumatologia. Fica ilustrada a possibilidade de a artropatia gotosa, em especial quando em evolução crónica, poder simular quadros clínicos de outras artropatias inflamatórias com apresentação poliarticular persistente, contrastando com o quadro mais típico de monoartrite recorrente autolimitada. Por último, recorda-se a necessidade de estarmos atentos a patologias que podem conduzir à positividade do factor reumatóide (apesar do seu valor diagnóstico na AR), destacando-se ainda a importância da ecografia músculo-esquelética no diagnóstico diferencial das artropatias inflamatórias.

CC121 – DOENÇA POR DEPOSIÇÃO DE CRISTAIS DE PIROFOSFATO DE CÁLCIO – UMA CAUSA DE ARTRITE DESTRUTIVA

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Introdução: O diagnóstico diferencial de um quadro clínico de monoartrite destrutiva é extenso. No caso de uma apresentação aguda, as causas infecciosas são as mais temidas, requerendo tratamento urgente. Para além da artrite séptica, as causas mais comuns incluem a artrite pós-traumática, necrose avascular, artrite neuropática e as artrites microcristalinas. A doença por deposição de cristais de pirofosfato de cálcio (DCPC) raramente assume tal curso, sendo mais frequentemente identificada nas suas formas clínicas pseudo-reumatóide, de pseudo-gota, pseudo -osteoartrose ou condrocalcinose assintomática.

Caso clínico: Apresenta-se o caso clínico de uma mulher de 52 anos de idade, sem antecedentes médicos relevantes, que recorreu ao serviço de urgência por queixas de omalgia inflamatória com marcada limitação funcional do ombro esquerdo, de início súbito e sem antecedentes traumáticos ou queixas sistémicas, nomeadamente, febre. A radiografia evidenciou destruição exuberante da extremidade proximal do úmero e uma erosão grosseira na glenóide. O estudo analítico revelou síndrome bio-inflamatório importante. Foi realizada artrocentese com a obtenção de líquido sinovial (LS) de características inflamatórias, tendo a doente sido submetida a lavagem cirúrgica da articulação gleno -umeral por suspeita de artrite séptica. O exame cultural do LS foi negativo. Foi posteriormente avaliada no nosso serviço, por reaparecimento das queixas e do derrame articular exuberante, confirmado por ecografia. O estudo subsequente incluiu a realização de nova artrocentese, com pesquisa de cristais, e também de biópsia sinovial. Não sendo identificados cristais no líquido sinovial, foram identificados, no tecido sinovial, cristais com birrefringência negativa na microscopia de luz polarizada, compatíveis com pirofosfato de cálcio. Não foram identificados cristais de monourato de sódio ou hidroxiapatite, nem foram isolados quaisquer micro-organismos no líquido ou tecido sinovial. A doente iniciou terapêutica com colquicina e anti -inflamatório não-esteróide, verificando-se melhoria das queixas e achados do exame físico, apoiando o diagnóstico de DCPC.

Conclusões: A DCPC pode assumir múltiplas apresentações clínicas, devendo, por isso, ser incluída nos diagnósticos diferenciais de quadros de artrite, em particular em doentes acima dos 60 anos de idade. O caso clínico ilustra uma apresentação rara desta condição, não só por ser rapidamente destrutiva (forma pseudo-neuropática) mas também por ocorrer numa idade mais precoce.

CC124 – SPONDYLOARTHROPATHY ASSOCIATED WITH GLYCOGEN STORAGE DISEASE TYPE I: A CASE REPORT

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Introduction: Glycogen storage disease type I (GSD-I) is a group of rare inherited diseases resulting from a defect in the glucose-6-phosphatase system, which is crucial in the glucose homeostasis. The most common manifestations of this condition are hepatomegaly, hypoglycemia associated with hyperlactacidemia, hypertriglyceridemia and hyperuricemia. Previous research describes the association between GSD-I and arthritis, namely acute gout and chronic arthritis with uveitis, however, to our knowledge, no association between GSD-I patient and spondyloarthropathy has been described.

Case report: We report a rare case of a 35-year-old male, weighting 83kg and 1.62m tall (body mass index 31.6Kg/m2), with a diagnosis of GSD-I since childhood. At 20 years old, he was referred to the rheumatology consultation because of a low energy fracture (bone densitometry with lumbar spine Z-score equal to -3.7 and femoral neck Z-score equal to -0.9), having been medicated with zoledronic acid and supplemented with calcium and vitamin D. An improvement of lumbar spine and femur neck Z scores (+2.1 and +0.1, respectively) was observed after about two years. At 25 years old, patient was again observed in this consultation due to neck pain, inflammatory low back pain and bilateral hip pain with prolonged early morning stiffness for the last 4 months. There was no fever, skin rash, genital or oral ulcers, uveitis and gastrointestinal or genitourinary manifestations. General physical examination was normal. On musculoskeletal examination, there was no peripheral arthritis or enthesitis; however the FABER (Patrick) test on the right was positive. Besides that, he had of cervical rotation limitation bilaterally and 2.5 cm on the modified Schober's test. On investigation, he had a mild microcytic anemia (hemoglobin 11.8 g/dL) and an elevated erythrocyte sedimentation rate (ESR) (41mm/h) and C-reactive protein (CRP) (35mg/L). Leukogram, platelet, hepatic and renal function, and urinalyses were normal. He's HLA-B27 positive. Rheumatoid factor, anti–citrullinated protein antibody (anti-CCP), antinuclear antibody (ANA) and extractable nuclear antigens (ENA) were negative. Complement levels and quantitative immunoglobulin were normal. The X-rays revealed presence of syndesmophytes in the cervical spine and grade III bilateral sacroiliitis. Based mainly on the clinical course and radiographic findings, a diagnosis of HLA-B27 positive ankylosing spondylitis was made. A nonsteroidal anti-inflammatory drug was added to osteoporosis therapy, initially with improvement of clinical complaints and a decreased of inflammatory parameters. After two years of follow-up, there was worsening of low back pain and the onset of gastrointestinal symptoms with a persistent perianal fistula. The X-rays demonstrated progression of structural damage, with an anterior cervical spine block C2-C4 and C5-C6, syndesmophytes in the lumbar spine and grade IV bilateral sacroiliitis. The patient performed pelvic abdominal MRI with gadolinium, which confirmed a Crohn's disease. Infliximab was initiated, which resulted in progressive improvement of low back pain and gastrointestinal complaints, with a normal ESR (4.5 mm/h) and CRP (3 mg/L), despite radiographically present a bamboo spine. **Conclusion:** To our knowledge, this is the first description of a GSD-I patient with a spondyloarthropathy. A high index of clinical suspicion of rheumatic diseases, such as spondyloarthropathy, in these patients is needed to establish an early diagnosis and to target treatment to prevent structural damage.

CC126 – POLYMYALGIA RHEUMATICA-LIKE SYNDROMES: 5 ILLUSTRATIVE CASES

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Background: Polymyalgia rheumatica (PMR) is characterized by a bilateral shoulder and/or pelvic girdle pain and stiffness with an elevated erythrocyte sedimentation rate (ESR) and/or C-reactive protein (CRP) levels in patients older than 50 years. Differential diagnoses of PMR include other inflammatory systemic diseases and malignancy.

Objective: To describe paradigmatic cases of patients that presented with PMR-like symptoms whose final diagnosis was not PMR.

Case 1: An 83-year-old (yo) woman presented with a 2-year-long history of bilateral shoulder and hip pain, constitutional symptoms and elevated ESR (87

mm/h). Treatment with low-dose glucocorticoids (LDGC) ensured symptomatic relief but was stopped due to GC-induced psychosis. Knee radiographs revealed chondrocalcinosis and a diagnosis of calcium pyrophosphate deposition disease (CPPD) was assumed. Colchicine treatment led to great clinical and analytical improvement.

Case 2: An 82yo man presented with a 3-week-long history of bilateral shoulder pain, swelling and pain of the wrists and hands, constitutional symptoms and elevated ESR (59 mm/h) and CRP (7 mg/dL). Physical examination revealed polyarthritis. LDGCs were initiated with partial symptomatic relief and persistent arthritis was objectified at the second appointment. Blood tests revealed a positive rheumatoid factor and a diagnosis of late-onset rheumatoid arthritis (LORA) was established. Treatment with methotrexate induced sustained remission and complete GC tapering.

Case 3: A 79yo woman presented with a 1-month -long history of bilateral shoulder and hip pain and stiffness, constitutional symptoms and elevated ESR (64 mm/h), CRP (1 mg/dL) and creatinine (2 mg/dL). Physical examination revealed polyarthritis and generalized oedema. The analytical and clinical response of both PMR-like symptoms and oedema was suboptimal to treatment with LDGCs and furosemide and the protein electrophoresis showed hypogammaglobulinemia. Immunofixation revealed a light-chain clone and the myelogram had 12% of plasma cells, of which 97% were monoclonal. The patient refused multiple myeloma treatment and died 10 months later.

Case 4: A 73yo woman presented with a 6-month -long history of low-grade fever, bilateral shoulder and hip pain and stiffness, anorexia and elevated ESR (55 mm/h) and CRP (9 mg/dL). LDGCs were initiated with partial improvement. Three months later, the patient complained of temporal headache and an ultrasound of the temporal arteries revealed a "halo sign". Giant cell arteritis (GCA) was suspected and GC therapy was adjusted accordingly. All symptoms resolved with inflammatory markers normalization and the patient is currently tapering GCs.

Case 5: A 73yo man presented with a 1-year-long history of bilateral shoulder and hip pain, constitutional symptoms and elevated ESR (115 mm/h) and CRP (8 mg/dL). Physical examination revealed generalized oedema and polyarthritis that were treated with LDGCs and furosemide. Improvement was specifically suboptimal at the hands. Puffy fingers and sclerodactyly were noted and immunological characterization revealed a positive anti-Pm/Scl autoantibody, leading to the diagnosis of limited cutaneous systemic sclerosis. The patient is currently being screened for organ involvement.

Conclusions: All patients diagnosed with PMR should be warned about how to proceed if GCA symptoms occur. Patients should also be screened for CPPD and LORA at diagnosis. A poor response to LDGCs should raise the possibility of malignancy, subclinical GCA or an alternative diagnosis.

CC127 – GASTRIC ADENOCARCINOMA PRESENTING AS A RHEUMATOID FACTOR AND ANTI-CYCLIC CITRULLINATED PROTEIN ANTIBODIES ARTHRITIS, A CASE-BASED REVIEW OF SEROPOSITIVE PARANEOPLASTIC ARTHRITIS

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Introduction: A variety of malignancies may present as paraneoplastic syndromes with rheumatologic manifestations. Paraneoplastic arthritis (PA) presentation, both clinically and serologically, may be the same as rheumatoid arthritis (RA).

Case-report: A 64-year-old male presented with a six-month history of symmetric polyarthritis involving proximal interphalangeal joints and metacarpophalangeal joints of the hands, wrists and ankles. Associated symptoms included vomiting, fatigue and weight loss. Laboratory results showed microcytic anaemia, leucocytosis, thrombocytosis, elevated C-reactive protein and erythrocyte sedimentation rate and positive rheumatoid factor (RF) and anti-cyclic citrullinated protein (anti-CCP) antibodies. No erosions could be identified in joints radiographs. Upper endoscopy and gastric endoscopic ultrasonography showed a gastric adenocarcinoma with regional lymphatic involvement. Intraoperatively, peritoneal carcinomatosis was documented and the patient started palliative chemotherapy. A paraneoplastic seropositive arthritis was assumed and treatment with low-dose prednisolone and hydroxychloroquine was started. Arthritis remission was achieved and sustained up to 18 months of follow -up, despite documented gastric cancer progression.

TABLE 1 – CHARACTERISTIC FEATURES OF PUBLISHED CASES OF RF AND ANTI-CCP ANTIBODIES POSITIVE PARANEOPLASTIC ARTHRITIS

Reference	Age/ sex	Type of malignancy	Duration of symptoms preceding cancer diagnosis (months)	Family history of cancer	Arthritis location and pattern	Onset	Anaemia	ESR (mm/h)/ CPR (mg/dL)	RF (IU/ml)/ anti-CCP (IU/mL)	Joints radiographs	Response to NSAIDs	Response to corticosteroids	Cancer treatment used (response or evolution)
Kumar et al. [14]	58/M	Pancreatic cancer	2	N.M.	No arthritis; arthralgia involving hands, wrists, elbows, shoulders, lower back, and neck; asymmetric and intermittent	Gradual	Yes	++ / N.M.	+++ / ++	N.E.	N.M.	Yes	None (died)
Raja et al. [15]	40/M	Lymphomatoid granulomatosis	3-4	No	Wrists, knees, and ankles; symmetric	Gradual	No	- / +	++ / ++	N.E.	N.M.	Yes (partial)	None (died)
Larson et al. [16]	45/F	Lung adenocarcinoma	3	No	PIP, MCP, elbows, and knees; symmetric	N.M.	No	- / +	++ / +	N.E.	No	Yes (partial)	None (died)
Handy et al. [4]	61/F	T cell lymphoblastic leukaemia	1-2	No	MCP, wrists, knees, and ankles; symmetric	Acute	Yes	+++ / ++	+++ / ++	Erosions	No	No	Hyper-CVAD CMT (refractory)
Watson et al. [8]	80/F	Breast papillary cancer	1	N.M.	Shoulder arthritis; arthralgia involving wrists, shoulders, and knees; asymmetric and migratory	Acute	Yes	- /++	+ / +	N.E.	No	Yes	CMT (N.M) and RT (remission)
Present	64/M	Gastric adenocarcinoma	6	Yes (GI and lung)	PIP, MCP, wrists, and ankles; symmetric	Gradual	Yes	++ / +++	++ / ++	N.E.	No	Yes	5-FU and CIS CMT (not curative)

CPR – C-reactive protein; ESR – erythrocyte sedimentation rate; anti-CCP – anti-cyclic citrullinated peptide; RF – rheumatoid factor; NSAIDs – non-steroidal anti-inflammatory drugs; F – female; M – male; GI – gastrointestinal; MCP – metacarpophalangeal joints; PIP – proximal interphalangeal joints; 5-FU – fluorouracil; CIS – cisplatin; hyper CVAD – cyclophosphamide, vincristine, doxorubicin, and dexamethasone; CMT – chemotherapy; RT – radiotherapy; N.E. – no erosions; N.M. – not mentioned.
ESR: - if < 30, + if ≥ 30 and < 60, ++ if ≥ 60 and < 100, +++ if ≥ 100.
CPR: + if ≥ 0.5 and < 5.0, ++ if ≥ 5.0 and < 15.0, +++ if ≥ 15.0.
RF titre: + if ≥ 14 and < 100, ++ if ≥ 100 and < 300, +++ if ≥ 300.
Anti-CCP titre: + if ≥ 20 and < 100, ++ if ≥ 100 and < 300, +++ if ≥ 300.

Discussion: We describe a unique phenotype of PA presenting as seropositive (RF and anti-CCP antibodies positivity) arthritis with a good response to both corticosteroid and hydroxychloroquine therapy. This is the sixth case described in the literature of PA with RF and anti-CCP antibodies positivity (Table 1), being the first associated with gastric cancer. PA has clinical and serological diversity, being seronegative RA-like arthritis the most frequent presentation. Establishing PA diagnosis implies a high level of clinical suspicion and a set of more inclusive features that represent the overall heterogeneity of this entity. This case highlights the importance of considering underlying cancer in patients over 50 years old, especially male, presenting with polyarthritis and systemic symptoms, even in those with seropositive RA-like arthritis. Moreover, corticosteroid therapy and disease-modifying anti-rheumatic drugs might have a role in controlling the PA even in patients with metastatic cancer.

CC134 – THE MANY FACES OF RELAPSING POLYCHONDRITIS

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Background: Relapsing polychondritis (RP) is a rare immune-mediated disease characterized by relapsing-remitting episodes of inflammation and progressive destruction of the cartilaginous and other proteoglycan rich structures such as the eyes, heart, blood vessels and skin. Cardiovascular complications occur in 31% of cases and are the second cause of mortality in RP.
Clinical case: We report the case of a 73-year-old male patient attending our Rheumatology Department after a diagnosis of relapsing polychondritis (migratory oligoarthritis, pinna chondritis and episcleritis / keratitis). He was treated with non-steroidal anti-inflammatory drugs (NSAIDs) and corticosteroids without development of new episodes. Two years after the diagnosis, the patient presented at the outpatient clinic with a 5 days history of bitemporal headache, fever, and weight loss. He denied nausea, vomiting, diplopia, blurred vision, jaw claudication or scalp allodynia. Dysphonia and pharyngeal foreign body sensation were mentioned.
Laboratory tests showed a macrocytic anaemia of 10.1 g/dL (normal: 11.5-18 g/dL), VGM 107 fL (normal: 76.0 – 96.0 fL), C-reactive protein of 26.81 mg/dL (normal < 0.5mg/dL) and erythrocyte sedimentation rate of 102 mm 1thhour (normal < 20 mm 1thhour). Testing for HIV and viral hepatitis was negative. Anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies, rheumatoid factor and anti-cyclic

citrullinated peptide were undetectable. Serum protein electrophoresis revealed increased of α1, α2 and β2 protein fractions. Ferritin was elevated (800 ng/mL – normal range: 20-291 ng/mL). Procalcitonin and hemocultures were negative. An FDG-PET/CT showed changes compatible with active vasculitis of the aorta cross, common brachiocephalic, subclavian and carotid arteries as well as thyroid cartilage chondritis. Temporal biopsy histology presented no signals of giant cell arteritis.
After the fourth cyclophosphamide cycle, there were no signs of vasculitis in FDG-PET/CT, but cyclophosphamide had to be suspended, because of severe leukopenia and neutropenia. It was replaced by tocilizumab (162 mg sc weekly) with sustained clinical and laboratory remission.
Conclusion: RP can be associated with involvement of large vessels. This is a life-threatening manifestation of the disease and physicians should be aware of this condition.

CC139 – POLIRRADICULOPATIA NO CONTEXTO DE INFEÇÃO POR CITOMEGALOVÍRUS

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Introdução: A infeção pelo citomegalovírus (CMV) associa-se normalmente a uma síndrome mononucleose-like auto-limitada, condicionando um período de latência prolongado, podendo originar reativações periódicas. Em imunodeprimidos, pode haver envolvimento multiorgânico grave.
Caso Clínico: Homem de 33 anos de idade, com antecedentes de hipertensão arterial, que em setembro de 2018 iniciou quadro febril (38-39ºC), acompanhado de mialgias, fraqueza muscular dos membros inferiores e tosse pouco produtiva, com cerca de duas semanas de duração. O estudo complementar inicial revelou leucocitose (14.66x109/uL), linfocitose (5.04 x109/uL), trombocitopenia (93x109/uL), elevação das transaminases (AST 60U/L [normal 4-33] e ALT 138U/L [normal 4-50]) e da creatinaquinase (CK 459U/L [normal <172]) e positividade para anticorpos anti-CMV, tanto IgM como IgG, tendo sido assumida infeção aguda por CMV. Um mês depois, apresentou, de novo, dispneia para médios esforços e palpitações, mantendo mialgias e noção de diminuição da força muscular das coxas. Realizou radiografia do tórax, provas de função respiratória e ecocardiograma transtorácico, que não mostraram alterações. Por manter as queixas, recorreu a consulta de Reumatologia em março de 2019, com um estudo analítico a revelar linfocitose (5.07x109/uL) e elevação da CK (592U/L), da aldolase (9.9U/L [normal <7.6]) e da mioglobina (111.5ng/mL [normal <146.9]). Os estudos de condução nervosa e eletromiografia (ECN/EMG) excluíram alterações miopáticas e polineuropáticas e demonstraram sinais compatíveis com polirradiculopatia (L5 e S1 bilateralmente) em fase subaguda-crónica, possivelmente enquadráveis em infeção por CMV. Assim, foi internado em Reumatologia em maio de 2019 e, à admissão, apresentava défice motor proximal nos membros inferiores (grau 4/5 à esquerda e 4+/5 à direita) e hiporreflexia aquiliana bilateral, sem outras alterações ao exame objetivo. Manteve perfil de elevação das enzimas musculares de semelhante magnitude. O estudo microbiológico (VIH, VHC, VHB, parvovírus B19, VEB, Coxiella burnetii e TPPA) e imunológico (ANCAs, fator reumatóide, ANAs e anticorpos anti-ENA, anti-dsDNA e anti-cardiolipina) foram negativos/normais, excetuando-se a presença de anticorpos anti-CMV IgG positivos, embora com IgM e DNA correspondentes negativos. Repetiu ECN/EMG, que confirmaram a evolução para cronicidade dos achados previamente descritos. A ressonância magnética (RM) das coxas não mostrou achados sugestivos de miosite e a RM da coluna lombar excluiu etiologia compressiva. O estudo do líquor não demonstrou alterações de relevo, nomeadamente hiperproteinorráquia; a pesquisa de DNA de CMV foi negativa. Dada a coincidência temporal entre a seroconversão dos anticorpos de CMV e o início do quadro clínico, foi assumido como provável o diagnóstico de polirradiculopatia por CMV. Aos 6 meses após o internamento, o doente apresentou melhoria marcada das mialgias, com normalização da força muscular, e retomou a atividade laboral.
Conclusão: O atingimento do sistema nervoso periférico pelo CMV em imunocompetentes é raro, sendo a polirradiculopatia uma das manifestações possíveis. Nestes casos, a estratégia terapêutica não está bem definida, no entanto, a maioria dos autores defende uma atitude expectante sem terapêutica anti-viral, tendo em conta o bom prognóstico e a resolução completa, ainda que lenta, na maioria dos casos.

CC146 – ARTERITE DE CÉLULAS GIGANTES RECIDIVANTE: A IMPORTÂNCIA DOS POUPADORES DE GLUCOCORTICÓIDES

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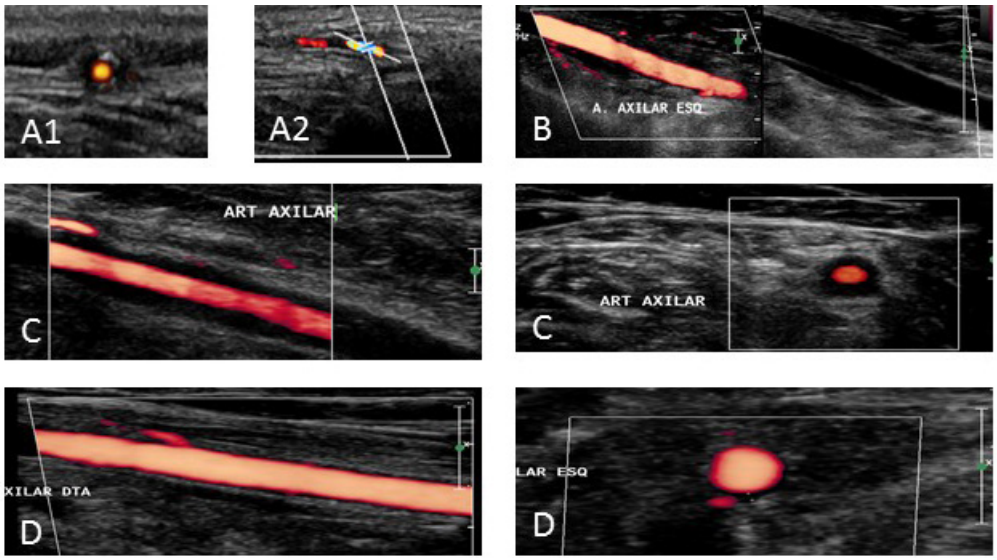
Introdução: A Arterite de Células Gigantes (ACG) é uma vasculite de grandes vasos, mais comum acima dos 50 anos. Pode ditar sequelas importantes, pelo que o tratamento com corticoterapia em alta dose deve ser precoce. O metotrexato e o tocilizumab provaram um efeito poupador de corticóide na ACG.
Caso Clínico: Mulher, 71 anos, com múltiplos fatores de risco cardiovascular (diabetes mellitus, hipertensão arterial e dislipidemia). Em janeiro de 2018 foi internada em Neurologia após 3 episódios de amaurose fugaz no olho direito (5 minutos de duração). Referia, desde há alguns meses, rigidez das cinturas escapular e pélvica, claudicação mandibular, astenia e anorexia com perda ponderal de 8 kg em 3 meses. Ao exame objetivo apresentava dor à palpação e pulso não palpável da artéria temporal esquerda. A avaliação por Oftalmologia excluiu patologia oftalmológica. O estudo complementar revelou anemia normocítica normocrómica (Hb 10.4g/dL), elevação dos marcadores inflamatórios (PCR 36.1 mg/L, VS 110mm/h) e trombocitose (421x109/L). A tomografia computorizada cerebral foi normal e o ecodoppler cervical, transcraniano e das artérias temporais superficiais mostrou espessamento segmentar concêntrico nos segmentos proximais das artérias temporais superficiais, com aceleração da velocidade de fluxo à esquerda, compatível com infiltrado vasculítico. As artérias axilares tinham aspeto ecográfico normal. Dada a elevada suspeita clínica e os achados neurossonológicos, foi dispensada a biópsia da artéria temporal para o diagnóstico de ACG e iniciou pulsos de metilprednisolona e.v. 1g/dia durante 5 dias, com melhoria clínico-analítica marcada, tendo alta sob prednisolona oral 60mg/dia (1mg/kg/dia), alendronato semanal e suplementação de cálcio/vitamina D, mantendo o controlo dos fatores de risco cardiovascular. Realizou desmame lento da prednisolona, mantendo-se assintomática até maio de 2019 quando, já sob prednisolona 2.5mg/dia, referia cefaleias temporais diárias à esquerda, rigidez de novo nas cinturas escapulares e astenia. Apresentava fácies cushingóide, com difícil controlo da diabetes mellitus apesar do início de insulinoterapia. O ecodoppler das artérias temporais e axilares revelou ligeiro espessamento de predomínio hiperecogénico (crónico) no segmento proximal da artéria temporal superficial esquerda e, nas artérias axilares, sinais de espessamento hipoecogénico concêntrico bilateralmente, com aceleração da velocidade de fluxo (figura 1). O estudo analítico revelou recrudescimento da anemia (Hb 9.9 g/dL) e da elevação da VS (97mm/h). Aumentou-se a prednisolona para 40mg/dia e foi requisitada consulta de Reumatologia. Tendo em conta a recidiva e os efeitos

FIGURA 1 – ASPETO ECOGRÁFICO DAS ARTÉRIAS TEMPORAIS SUPERFICIAIS E AXILARES AO LONGO DA EVOLUÇÃO DO CASO CLÍNICO



A) Jan/2018: espessamento concêntrico com componentes hipo e hiperecogénico (evolução crónica de infiltrado vasculítico?) nos segmentos proximais das artérias temporais (A1), com aceleração da velocidade de fluxo (A2); B) Dez/2018: normalidade do contorno vascular da artéria axilar esquerda; C) Maio/2019: sinais de espessamento hipoecogénico e concêntrico nos segmentos acessíveis das axilares, com aceleração local da velocidade de fluxo; D) Dez/2019: Normalização do espessamento hipoecogénico patológico nas artérias axilares.

da corticoterapia, foi pedido o tocilizumab, que não foi aprovado. Em agosto de 2019, foi internada em Ortopedia por fraturas compressivas de D6 e D12, para tratamento conservador e, 1 mês depois, iniciou o metotrexato 15mg/semana, com melhoria clínico-analítica, o que permitiu o desmame da corticoterapia, encontrando-se atualmente sob prednisolona 15mg/dia, mantendo o bifosfonato oral.

Conclusão: Trata-se de um caso de recidiva de ACG com atingimento das artérias temporais superficiais e mais tarde das axilares, com fraturas vertebrais no contexto da corticoterapia. Este caso salienta a importância da prevenção/orientação dos efeitos adversos da corticoterapia na ACG, tendo em conta a faixa etária dos doentes, desde os fatores de risco cardiovascular até à osteoporose. Realçamos a importância da instituição precoce dos poupadores de corticóides nas recidivas e nos doentes corticodependentes.

CC154 – PLEUROPARENCHYMAL FIBROELASTOSIS IN MIXED-CONNECTIVE TISSUE DISEASE

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Background: Pleuroparenchymal fibroelastosis (PPFE) is a newly described form of interstitial lung disease (ILD), characterized by progressive fibrotic thickening of the visceral pleura and subpleural parenchyma, involving predominantly the upper lobes. Although most cases are idiopathic, it can be associated with underlying conditions, namely connective tissue diseases.

Case report: A 27-year-old black man, non-smoker, was referred to our Rheumatology department by Pneumology with a 3-year history of exertional dyspnea, dry cough and non-quantified weight loss. There was no significant history of occupational or environmental exposures. When questioned about other complaints, he referred two-phase Raynaud's phenomenon in his fingers, puffy hands and xerostomia for nearly two years. He denied digital ulcers, xeropthalmia, arthralgias or other constitutional symptoms. On physical examination, he had puffy fingers, cold hands and globally diminished breath sounds on pulmonary auscultation.

Laboratory investigation revealed normal blood counts and normal inflammatory parameters. ANA titer was 1/640, with strong positivity for anti-U1 ribonucleoprotein (RNP), anti-SSA and anti-SSB; anti-dsDNA was negative, as well as ANCA and viral serologies. Videocapillaroscopy showed an early scleroderma pattern and the salivary gland biopsy was unremarkable. At this point, a presumptive diagnosis of mixed-connective tissue disease (MCTD) was established.

High-resolution computed tomography (HRCT) of the chest revealed multiple bilateral traction bronchiectasis with fibrotic thickening predominantly in the upper lobes and elevation of hilar opacities. In the mid- and lower zones, irregular bullae suggestive of emphysema were detected. Alpha 1 antitrypsin was, however, within normal values. Lung function tests revealed severe restrictive ventilatory impairment (FVC 42%, FEV1 48%, FEV1/FVC 116, TLC 56%) and decreased diffusion capacity of carbon monoxide (29%). Flexible bronchoscopy with bronchoalveolar lavage (BAL) was performed and no malignant cells or pathogenic microorganisms were observed.

The case was discussed in an ILD specialized multidisciplinary meeting and, based on clinical manifestations and imaging findings, he was diagnosed with PPFE. Due to the advanced stage of the disease, with great extent of pulmonary lesions and severe functional respiratory impairment, he was started on rituximab. Since then, he remains clinically stable and is under close monitoring.

Conclusion: We described a case of a patient with MCTD that fulfilled the imaging diagnostic criteria for PPFE proposed by Enomoto et al1. In this entity, the mid- and lower zones are initially spared, but they can be progressively envolved with time. Although there is no demonstrated effective treatment, immunosuppressive agents can be used in cases of progressive or severe disease. We should be aware of this condition, which is accompanied by a poor prognosis especially when associated with a connective tissue disease. Moreover, we reinforce the crucial role of multidisciplinary meetings in the diagnosis and management of these cases.

References:

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CC156 – GIANT CELL ARTERITIS AS A POSSIBLE PARANEOPLASTIC MANIFESTATION

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Introduction: Giant cell arteritis (GCA) is a vasculitis of large- and medium-sized arteries affecting patients aged >50 years. About half the cases are associated with polymyalgia rheumatica (PMR). Paraneoplastic syndromes have been described in rheumatic diseases, and musculoskeletal pain mimicking polymyalgia rheumatica (PMR) may occur in some malignancies. Conflicting results have been reported on the risk of malignancy in GCA and it is commonly believed that GCA should not be regarded as a paraneoplastic syndrome. We present four case reports of patients diagnosed with GCA who presented a neoplastic condition.

Case reports

Case 1: A 67-year-old man presented with weight loss, PMR symptoms, temporal headache, and left maxillary pain. C-reactive protein (CRP) was 2.4 mg/dL and erythrocyte sedimentation rate (ESR) 35 mm/hr. Temporal artery (TA) biopsy was negative and ultrasound of the TA and axillary (AX) arteries showed no signs of vasculitis. Despite these results, high clinical suspicion for GCA prompted glucocorticoid (GC) therapy with clinical improvement. During the investigation, computed tomography (CT) revealed a suspicious dense nodular lesion with 16mm in the inferior pole of the right kidney. The patient underwent partial nephrectomy and histology confirmed a renal clear cell carcinoma. Four years after initial diagnosis, symptoms recurred and large vessel imaging was consistent with vasculitis of the aorta, AX and subclavian (SC) arteries.

Case 2: An 84-year-old man with a clinical history of marked weight loss, PMR, headache, and jaw claudication was referred for Rheumatology observation. CRP was 3.26 mg/dL, ESR 89 mm/hr and ultrasound presented halo sign of the AX and SC arteries. He was started on GCs, with laboratory improvement. Due to his frail condition, a CT was requested showing a solid heterogeneous nodule in the left adrenal gland, suggestive of primary neoplasm. The patient died 3-months after the diagnosis due to a respiratory infection before the investigation was concluded.

Case 3: A 90-year-old man presented with severe weight loss, arthralgia, anterior ischaemic optic neuropathy, temporal headache, and jaw claudication. CRP was 2.8 mg/dL, ESR 93mm/hr and ultrasound showed halo sign of TA, AX, and SC arteries. GC treatment was started with moderate clinical improvement. After 2-months, the patient developed a respiratory infection and thoracic CT reported a mass with multiple surrounding nodules, most likely corresponding to primary lung cancer and metastases. Given the age and dependency status of the patient, no further investigation was made and he died within 1-week.

Case 4: A 79-year-old woman was referred for observation due to complaints of weight loss, PMR symptoms, severe headaches and jaw claudication. CRP was 8.1 mg/dL and ESR 119 mm/hr. The patient had TA halo sign on ultrasound. Treatment with GCs was initiated with marked improvement. At 9-months of follow-up, she reported nausea and epigastric pain; upper endoscopy showed a peptic ulcer and histology revealed moderately undifferentiated gastric adenocarcinoma. She now awaits surgery.

Conclusion: In all four cases, patients were diagnosed with both GCA and a malignant condition in less than one year apart, suggesting that GCA may have presented as a paraneoplastic syndrome. In three cases PMR features were also documented. Future work on this topic should explore the hypothesis of GCA acting as a true paraneoplastic syndrome and consequently, the potential resolution of vasculitis as a result of the malignancy treatment.

CC165 – EFEITOS ADVERSOS DA IMUNOTERAPIA NO CANCRO: UM DESAFIO DIAGNÓSTICO

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Introdução: Os inibidores dos checkpoints imunológicos revolucionaram o tratamento oncológico por aumentarem a sobrevida de algumas neoplasias, nomeadamente do cancro do pulmão de não-pequenas células. Apesar dos seus benefícios clínicos, estão associados a efeitos adversos, devido ao mecanismo inespecífico de ativação das células T, que podem afetar qualquer órgão ou tecido e que não devem ser negligenciados.

Caso clínico: Doente do sexo masculino de 64 anos, ex-fumador (30 UMA) com adenocarcinoma do pulmão estadio IV tratado com pembrolizumab (anti-programmed cell death receptor-1 – anti-PD-1) 200mg endovenoso, a cada 3 semanas. Após a 5ª toma do fármaco, recorreu à consulta de reumatologia por dor e tumefação do joelho direito, tibiotársica esquerda e 4ª articulação interfalângica proximal esquerda, com objetivação de artrite ao exame físico. Sem história pessoal ou familiar de psoríase, doença inflamatória intestinal ou uveíte. Negava raquialgias, lesões cutâneas, fenómeno de Raynaud, xerostomia ou xeroftalmia. Analiticamente apresentava elevação de parâmetros inflamatórios (VS 81mm, PCR 9.8mg/L) e fator reumatóide, anticorpos antipeptídeo citrulinado, anticorpos antinucleares e HLA-B27 negativos. O doseamento do ácido úrico sérico foi normal. Foi realizada artrocentese do joelho direito com drenagem de líquido sinovial de características inflamatórias. O exame microbiológico, incluindo bacteriológico e micobacteriológico, e a pesquisa de cristais no líquido sinovial foram negativos. Iniciou anti-inflamatório não esteróide e prednisolona 5mg/dia com melhoria franca do quadro articular. Após a 8ª toma de pembrolizumab, reaparecimento de gonartrite bilateral com incapacidade para a marcha, associada a febre e diarreia (4-5 dejeções/dia de fezes líquidas, sem sangue ou muco). Do estudo realizado destacava-se elevação de parâmetros inflamatórios (VS 89mm/h, PCR 146.9mg/L). O rastreio séptico alargado não evidenciou foco infecioso. A colonoscopia não mostrou qualquer lesão de relevo. A tomografia computorizada toraco-abdomino-pélvica não demonstrou lesões de novo e a tomografia por emissão de positrões com FDG-F18 não apresentou progressão da doença. Assumida provável toxicidade medicamentosa ao pembrolizumab, foi decidido a suspensão do fármaco e o início de prednisolona (1mg/kg/dia), com resolução da artrite, da diarreia e da febre.

Conclusão: Este caso ilustra a importância do reconhecimento das potenciais reações adversas da imunoterapia em oncologia, algumas delas requerendo uma abordagem e tratamento multidisciplinar. No entanto, devem ter-se em consideração diagnósticos diferenciais particularmente importantes neste grupo de doentes, nomeadamente a exclusão de progressão da doença bem como etiologia infeciosa. Este doente reúne várias reações adversas imunitárias secundárias à imunoterapia: artrite, colite e pirexia. Com base na gravidade da reação adversa, o pembrolizumab deve ser suspenso e devem ser administrados corticosteróides.

CC166 – DOENÇA DE SHULMAN E ANEMIA PERNICIOSA: UMA ASSOCIAÇÃO EXCECIONALMENTE DESCRITA

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Introdução: A doença de Shulman, ou fasceíte eosinofílica, é uma doença inflamatória sistémica fibrosante de etiologia desconhecida. O seu diagnóstico é um desafio, não só por ser uma entidade rara, mas também por poder mimetizar outras patologias como a esclerose sistémica. Associa-se a distúrbios hematológicos em 10% dos casos.

Caso clínico: Doente do sexo masculino de 75 anos, com antecedentes de asma bem controlada. Observado por quadro clínico com 4 meses de evolução, caracterizado por parestesias e edema bilateral dos membros inferiores, seguido de eritema e noção de espessamento cutâneo dos quatro membros, com franca limitação para a marcha. Referia perda ponderal de 4kg no último mês, sem outros sintomas constitucionais. Negava fenómeno de Raynaud, disfagia ou dispneia. Ao exame objetivo apresentava: espessamento cutâneo dos membros superiores e inferiores, poupando mãos, pés, face e tronco; antebraços com depressão linear ao longo do trajeto das veias superiores consistente com o sinal do sulco; limitação franca da mobilidade das articulações tibiotársicas e subtalares. Analiticamente, salientava-se anemia macrocítica (Hb 12.1g/dL, VGM 118 ft), eosinofilia periférica (1.8 x 109/L), elevação da proteína C reativa (31.1mg/L) e da aldolase (19.6U/L). A creatina cinase, velocidade de sedimentação e proteinograma eram normais, fator reumatóide e anticorpos antinucleares negativos. Realizou ressonância magnética (RM) dos membros inferiores com evidência de edema dos tecidos moles de predomínio fascial. A biópsia de pele, fáscia e músculo na região da coxa esquerda mostrou fibrose e inflamação com alguns eosinófilos compatível com a hipótese clínica de fasceíte eosinofílica. A colonoscopia e a tomografia computorizada toraco-abdomino-pélvica não evidenciaram alterações de relevo. O estudo da anemia revelou défice de vitamina B12 [171.0 pg/mL (N: 250-1100pg/mL)], ácido fólico normal, anticorpos anti-células parietais positivos e anti-fator intrínseco negativos. Foi realizada endoscopia digestiva alta

que evidenciou uma gastrite crónica ligeira e atrofia moderada. A RM da coluna cervical mostrou aspetos compatíveis com degenerescência subaguda combinada da medula e a eletromiografia dos membros inferiores revelou uma polineuropatia sensitiva do tipo axonal. Fez suplementação intramuscular de vitamina B12 com resolução da anemia e melhoria parcial das parestesias. O doente foi medicado com prednisolona 0.5mg/kg/dia com melhoria do espessamento cutâneo e resolução da eosinofilia e, posteriormente, com metotrexato 15mg/semana com desmame progressivo do corticoide.

Conclusão: A doença de Shulman é uma doença rara mas provavelmente subdiagnosticada. A RM e a biópsia de toda a espessura cutânea são fundamentais para o diagnóstico. O tratamento de primeira linha são os corticosteróides, isolados ou em associação com outros fármacos imunossupressores. Embora associada a distúrbios hematológicos, existem poucos casos descritos na literatura da associação entre a doença de Shulman e a anemia perniciosa.

CC176 – SUBLUXATION ARTHRITIS AND ANTISYNTHETASE SYNDROME

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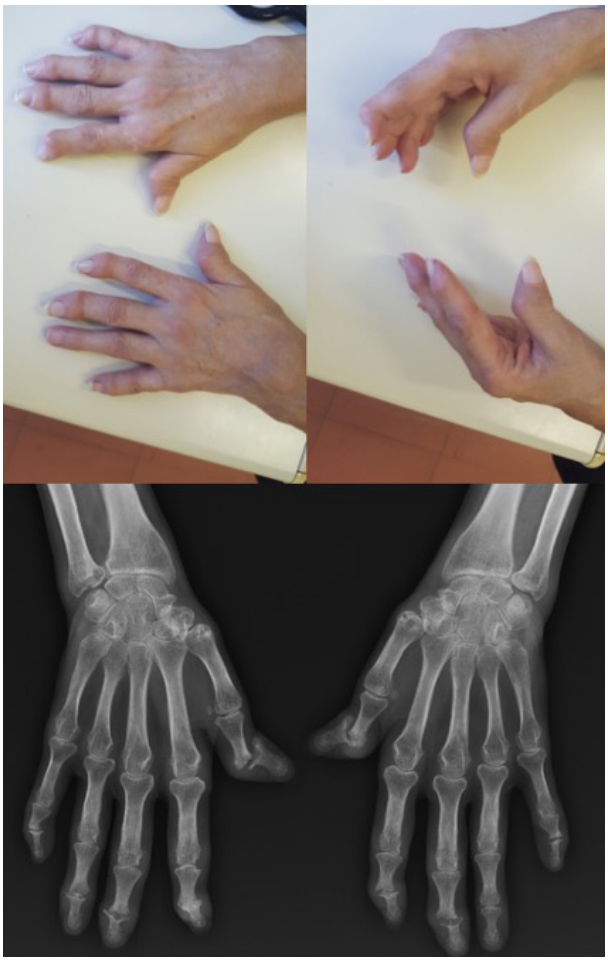
Background: Joint involvement occur in up to 90% of patients with antisynthetase syndrome (ASS), and it can be the initial manifestation. Joint involvement in ASS include arthralgia, polyarthritis, erosive arthritis and subluxation arthritis. Subluxation arthritis in ASS is uncommon, affects predominately distal interphalangeal joints (IP) and is associated with anti-Jo-1 antibodies.

Objectives and Methods: We report a case of ASS presenting with isolated subluxation arthritis.

Results: A 50-year old woman presented in our rheumatology department with inflammatory arthralgia of both hands beginning 3 years ago and already medicated with deflazacort 15mg/day since 4 months. She had arthritis and subluxation of DIP (figure 1) and had positive antinuclear antibodies (title 1/1280), anti-Jo1 antibodies and anti-mitochondrial M2. She started methotrexate with regression of arthralgias and was studied for other organ involvement. On pulmonary computorized tomography (CT) scan she had light changes of ground glass in the superior pulmonary lobes and some unspecify peripheral micronodules. Her pulmonary function tests were within the normal range. Although she doesn't yet have interstitial lung disease (ILD) she remains under close pulmonary monitorization.

FIGURA 1 – SUBLUXATION ARTHRITIS OF A PATIENT WITH ASS. UPPER PICTURES SHOWING PATIENTS HANDS AND LOWER PICTURE THE X-RAY



At disease onset, the classic clinical triad of ASS (arthritis, myositis and ILD) is only rarely observed. More frequently ASS articular presentation is symmetric polyarthritis of metacarpophalangeal and proximal IP with erosions or isolated inflammatory arthralgias. Subluxation arthritis is a rare type of articular involvement found on ASS and discovered to be exclusive to patients with anti-Jo1 antibodies.

Conclusion: Isolated arthritis is a common ASS clinical presentation and subluxation of DIP is a rare subtype of articular involvement that rheumatologists should be aware. This case highlights the existence of this rare articular subtype of joint involvement in ASS that is exclusive in anti-Jo1 positive patients.

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CC182 – METALOSE E INTOXICAÇÃO POR COBALTO: 2 CASOS DE FALÊNCIA DE PRÓTESE TOTAL DE ANCA METAL-METAL

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Introdução: As próteses de anca metal-em-metal foram amplamente utilizadas entre 2003 e 2010, mas verificou-se um elevado número de efeitos adversos como falência precoce da prótese por desgasto rápido da superfície articular, reações adversas locais e de hipersensibilidade a metais pesados, o que motivou a sua remoção do mercado. O presente trabalho demonstra 2 casos clínicos de intoxicação sistémica por cobalto após prótese total de anca meta-metal (PTAMM).
Caso 1: Mulher de 48 anos de idade, com espondilartrite seronegativa axial, não-radiográfica, submetida a PTAMM (crómio (Cr) e cobalto (Co)) bilateral em 2008 e história de eosinofilia com 5 anos de evolução. É internada por lesões urticariformes dispersas pelo corpo, coxalgia esquerda e massa nodular na face anterior da coxa esquerda, com cerca de 4 cm, não dolorosa, de consistência dura e de limites mal definidos e aderente aos planos profundos, com quatro meses de evolução. Da avaliação laboratorial, destacou-se eosinofilia severa (5570/uL). Fez ecografia da lesão, que sugeriu hematoma organizado, e posteriormente tomografia computorizada avançada que mostrou "na cavidade pélvica, em topografia retroperitoneal, à direita e na dependência do músculo íliopsoas, volumosa lesão ocupante de espaço com cerca de 8x10x11cm hipodensa". Fez-se biópsia da lesão que revelou "tecido conjuntivo fibro-adiposo com áreas de agregados de histiócitos com citoplasma vasto e pigmento castanho-escuro, sem achados sugestivos de neoplasia. Após discussão com imunoalergologia e ortopedia, é colocada a hipótese de reação a material de prótese com dermatite sistémica subsequente. A doente é submetida a revisão de prótese, verificando-se, no intraoperatório, metalose/pseudotumor extenso até ao terço distal ântero-lateral da coxa, cuja anatomia patológica confirmou os achados previamente obtidos em biópsia. Ao fim de 4 semanas, verifica-se resolução completa do quadro de coxalgia e urticária generalizada.
Caso 2: Mulher de 63 anos, com história de asma, osteoporose e poliosteoartrose, submetida a PTAMM esuqerda há 15 anos. Inicia quadro progressivo de coxalgia esquerda em associação a fadiga extrema, palpitações, dor generalizada, hipostesia dos membros inferiores e disestesias dos pés. Após extensa investigação etiológica, é solicitado o doseamento sérico de Co e Cr que revelou valores consideravelmente elevados de 52 µg/L (<1.2) e 12.2 µg/L (<5), respetivamente. Foi submetida a revisão de prótese em 2019, com consequente resolução do quadro de coxalgia. Mantém síndrome de fadiga crónica em associação a dor generalizada flutuante.
Discussão: Embora a coxalgia seja o sinal mais comum e precoce de falência protésica, reações cutâneas, pseudotumores e metalose são outras manifestações raras. O desgaste articular precoce de PTAMM cursa com a libertação de produtos metálicos (haptenos) para o espaço articular, que podem induzir a reações de corpo estranho e de hipersensibilidade, cujas manifestações podem cursar com massas ou outros sinais ou sintomas locais, ou terem apresentação inespecífica como urticária generalizada, sintomas cardiovasculares ou neurológicos. Não existe um teste de diagnóstico confirmatório, pelo que é necessário um alto índice de suspeição para considerar esta patologia num doente com história de PTAMM.

CC184 – LÚPUS ERITEMATOSO SISTÉMICO E HEMIPARÉSIA: UMA CAUSA NÃO-VASCULAR

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Introdução: Lúpus neuropsiquiátrico (LNP) é o termo usado para um largo espectro de manifestações neurológicas e psiquiátricas que podem surgir num doente com lúpus eritematoso sistémico (LES) e que estão associadas a significativa morbi-mortalidade. O envolvimento do sistema nervoso central (SNC) pode

decorrer por mecanismos vasculares ou parenquimatosos, com uma heterogeneidade de apresentação que continua a representar um desafio diagnóstico e terapêutico, com decisões tomadas caso-a-caso e com uma abordagem multidisciplinar.
Caso clínico: Mulher de 41 anos, com diagnóstico de LES há 20 anos, com envolvimento cutâneo (alopécia, rash malar), articular (artralgias das pequenas articulações das mãos), hematológico (anemia normocítica, normocrómica (NN)), perfil imunológico perfil imunológico anti-ds DNA positivo, ANAs em elevado título (anti-histonas, anti-U1-RNP), sem seguimento em Reumatologia, medicada com deflazacorte com ajustes sistemáticos, com mau controlo da doença. Inicia quadro de agravamento progressivo de fadiga, poliartralgias de ritmo inflamatório das pequenas articulações das mãos, parestesias e diminuição da força muscular no hemicorpo direito que motivou ida ao serviço de urgência. À entrada: lentificação psicomotora, desorientação têmporo-espacial, desvio da comissura labial e diminuição da força muscular no hemicorpo direito (grau 4/5). Mantinha anemia NN (Hb 9.8g/L) e aumento dos parâmetros de inflamação (VS>140mm e PCR 7.29mg/dL). Fez tomografia computorizada (TC) crânio-encefália (CE) que sugestionou lesão ocupante de espaço (LOE) insular à esquerda. Internada no serviço de Neurocirurgia com posterior colaboração do serviço de Reumatologia, com nova investigação etiológica: aumento persistente dos parâmetros de inflamação, anemia NN, hiperferritinémia (304 ng/mL), anti-dsDNA 111.7 IU/mL, ANAs positivos (1/640 com anti-U1-RNP) anticorpos antifosfolípidos negativos; hemoculturas, serologias virais, RPR, Toxoplasma gondii negativos; avaliação de líquido cefalorraquidiano com ligeiro aumento de proteínas sem pleocitose, eletroforese de proteínas sem bandas oligoclonais e exame cultural negativo; ressonância magnética nuclear (RMN) CE que mostrou "lesão difusa heterogénea, de limites mal definidos hipointensa em T1, hiperintensa em T2, na região temporal profunda esquerda, com extensão ao pedúnculo cerebral, núcleos da base e tálamo esquerdos, envolvimento das faixas óticas e parênquima cerebral à periferia do terceiro ventrículo e discreto efeito de massa sobre o terceiro ventrículo e o ventrículo lateral esquerdo", sem evidência de lesões vasculares; angio-TC-CE com ausência de oclusão de vasos intracranianos. Foi colocada como principal hipótese uma lesão parenquimatosa inflamatória (envolvimento do SNC em doente com LES em atividade). Iniciou terapêutica com dexametasona 8mg/dia com significativa melhoria clinica e analítica, com posterior switch para prednisolona 0.75mg/Kg/dia e início de azatioprina em dose crescente até 2,5mg/kg/dia. Após 1 mês de terapêutica estava sem sintomas, com nova RMN-CE que mostrou marcada diminuição da lesão intra-parenquimatosa, sem efeito de massa.
Conclusão: Não existem testes confirmatórios para o diagnóstico LNP e só uma detalhada avaliação clínica, laboratorial e imagiológica poderá estabelecer a sua presença. Neste caso, a ausência de dados sugestivos de infeção ou isquemia cerebral e a regressão da lesão parenquimatosa periventricular após terapêutica imunossupressora, reforçaram a hipótese de diagnóstico de envolvimento do sistema nervoso central numa doente com LES em atividade.

CC186 – TOCILIZUMAB FOR CORTICODEPENDENT POLYMYALGIA RHEUMATICA, REPORT OF A CASE AND REVIEW OF THE LITERATURE

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Introduction: Polymyalgia rheumatica (PMR) is an inflammatory disease that usually affects elderly people. It is characterized by bilateral pain and stiffness involving the shoulders, neck and pelvic girdle associated with elevated acute phase reactants. Oral prednisolone (PDN) is the cornerstone of treatment but concerns about glucocorticoid (GC) side effects remains an issue. Conventional immunosuppressive drugs, especially methotrexate (MTX) and leflunomide (LFN), are used as GC-sparing drugs but they have only moderate efficacy. Recently, observational studies indicate that the anti-IL6 receptor tocilizumab (TCZ) is useful in PMR.
Case-report: A 66-year-old male presented a nine-year history of corticodependent PMR treated with PDN (> 10 mg per day). His comorbidities were obesity, systemic hypertension, hypertensive nephropathy and hyperuricemia. Conventional disease-modifying antirheumatic drugs were used in an attempt to reduce corticoids. MTX (10 to 20 mg per week) had to be discontinued due to gastrointestinal intolerance. LFN (20 mg daily) was started with good tolerance, but still relapses occurred when the dose of PDN was less than 10 mg per day. Other diagnoses, namely cancer, giant cell arteritis, were excluded by colonoscopy, upper endoscopy, chest-abdomen-pelvis computed-tomography and ultrasonography of temporal and axillary arteries. Weekly subcutaneous

TCZ was added with adequate tolerance. Two adverse events occurred but they were unlikely related with TCZ (self-limited penile ulcers and toxidermic rash, possibly related to febuxostat that was recently started). At 18 months of follow up after TCZ initiation, LFN was suspended and PDN was decreased to 5 mg per day without relapses.

Discussion: We present a case of corticodependent PMR requiring long-term GC therapy because of recurrent flares despite the use of MTX and LFN. PDN dose reduction was only possible with the association of TCZ. Recent data confirm the utility of TCZ either as monotherapy or in association with GC for PMR. Moreover, TCZ used in corticodependent or refractory disease has a strong GC-sparing effect. Recent observational studies showed that TCZ added to GC therapy is more effective in achieving relapse-free remission off GCs than GC therapy alone and clinical trials are ongoing to clarify these observations. Its safety profile and good tolerance make it an attractive treatment option for PMR. This case highlights the need to consider the use of TCZ in PMR, especially in patients with risk factors for long-term GC therapy.

CC190 – OSTEOGENESIS IMPERFECTA AND BEHÇET DISEASE – TWO CASE REPORTS

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Background: Osteogenesis imperfecta (OI) is a group of rare inherited disorders of connective tissue with the common feature of bone fragility and recurrent fractures. It is mostly inherited as an autosomal dominant disorder with mutations in COL1A1 or COL1A2 genes. According to Sillence there are 4 types of clinical classification (type I OI is mild, type II is perinatal lethal, type III is progressive deforming and type IV is moderately severe). In the most mild cases, fractures usually decrease after puberty (1).

Behçet disease (BD) is a rare systemic vasculitis disorder of unknown etiology characterized mainly by recurrent oral aphthous ulcers, genital sores, cutaneous and ocular lesions. Half of patients may have a nonerosive arthritis (2). Familial aggregation of BD has been reported to occur in 1–18% of the population, with a higher incidence of familial association in juvenile patients (3). Effectiveness of biological disease modifying antirheumatic drugs (bDMARD) has been described in literature. Adalimumab was recently described as very effective and safe for severe and resistant BD uveitis, providing an appropriate and long-term control of ocular inflammation (4), although few cases are reported in literature.

Case-reports: A 30-year old man with OI type I is followed in our rheumatology department. He was previously medicated with pamidronate in his childhood and his last fracture was at 15 years old. He recently developed pseudofolliculitis cutaneous lesions in his dorsal and anterior chest, recurrent oral ulcerations, abdominal pain with diarrhea and arthritis specially of the writs. A diagnosis of BD was made and he was treated initially with methotrexate and colchicine. Because of persistent arthritis and need for maintained corticosteroids to control disease activity we decided to start Infliximab.

His younger sister, a 24-year old woman with OI type I is also followed in our rheumatology department and was previously medicated with pamidronate. Her last fracture was at 18 years old. One year after his brother BD diagnosis she also developed oral ulcerations, pseudofolliculitis cutaneous lesions and several episodes of anterior uveitis. She started treatment with prednisolone 1 mg/kg and methotrexate. She maintain severe ocular inflammation and had one new metatarsal phalange fracture. Because of her increased risk of fracture, prednisolone needed to be tapered, so we started Adalimumab to control ocular inflammation.

Conclusion: We report two siblings with two rare diseases, OI which is a inherit disorder and BD whose etiology is unknown but there is a reported familial connection. The association between the two conditions apparently does not exist, being probably an unfortunate coincidence. The association of these diseases is a therapeutic challenge since BD, especially with uveitis, is efficiently controlled with high dose corticosteroids and synthetic DMARD. However, corticosteroids contribute significantly to bone loss and can truly increase the risk of fracture in OI, so the use of other rapidly effective therapies like bDMARDs is an option which must be considered.

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CC193 – ADMINISTRAÇÃO DE CORTICOESTERÓIDES INTRAMUSCULARES EM DOENTE IMUNODEPRIMIDO – A DESCRIÇÃO DE

UM DESFECHO FATAL

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Introdução: A administração de corticoesteróides intramusculares (IM) é um procedimento invasivo minor e é frequentemente realizado em ambulatório para alívio sintomático no contexto de diversas patologias, em virtude do seu início de ação rápido e duração prolongada do efeito terapêutico. No entanto, ainda que raramente, pode associar-se a complicações potencialmente graves e ameaçadoras da vida como abcessos, bacterémia, sépsis e disfunção multiorgânica. A coexistência de imunossupressão não é considerada uma contraindicação formal ao procedimento.
Caso clínico: Homem, 56 anos, com antecedentes de bypass aorto-bifemoral por doença arterial periférica, seguido em Consulta Externa de Reumatologia por Síndrome de Sjogren com 4 anos de evolução, medicado com hidroxicloroquina 400mg/dia e prednisolona 5mg/dia. Por cervicalgia mecânica aguda no contexto de esforço físico intenso, realizou em agosto de 2019 betametasona IM com alívio da sintomatologia. Admitido ao internamento no nosso Serviço 10 dias depois, proveniente do Serviço de Urgência, por poliartralgias e sinais inflamatórios exuberantes no tornozelo direito, sem melhoria com anti-inflamatório não esteróide. Apresentava leucocitose 29.650/γl com neutrofilia 90,8% e PCR 347,9 mg/L. Realizou ecografia do tornozelo e pé que mostrou derrame articular de pequeno volume no recesso anterior da tibio-társica (TT), bem como tenossinovite do tibial posterior e edema do tecido celular subcutâneo da vertente medial do tornozelo. Fez artrocentese da TT direita com saída de líquido amarelo citrino, 5.133/γl células com 70% PMN, pesquisa de cristais e bacteriológico (disponível posteriormente) negativo. Colheu rastreio séptico e foi medicado com indometacina 125mg/dia e prednisolona 10mg/dia. A urocultura e hemoculturas foram positivas para Staphylococcus aureus meticilino-sensível, pelo que se iniciou antibioterapia com flucloxacilina. Por dor abdominal e vómito biliar e por surgimento de nódulo subcutâneo glúteo em local de administração IM prévia, fez TC abdomino-pélvico que mostrou múltiplos abcessos – perianal, intra-glúteos máximos, intra-psoas-ilíaco direito, espaço-periprostático/gordura isquiorretal esquerda, e infeção de prótese de bypass aorto-bifemoral com pseudoaneurismas na aorta torácica descendente e aorta abdominal. Decidido em conjunto com Cirurgia Vascular e Infecciologia manter antibioterapia e transferência para Unidade de Cuidados Intensivos. Apresentou nas 48h seguintes evolução desfavorável, tendo sido submetido a drenagem cirúrgica de abcesso perianal e alterada antibioterapia para piperacilina/tazobactam, e sofreu episódio de paragem cardiorrespiratória não reversível com saída de sangue abundante pela via aérea durante as manobras de reanimação. Foi assumida como causa de morte rotura de pseudoaneurisma da aorta torácica em contexto séptico com provável origem em abcesso intra-glúteo secundário a terapêutica IM.
Conclusão: Embora seja um procedimento de uso corrente na prática clínica, os autores reforçam a importância de se promover uma avaliação cuidada de cada doente, tendo em conta a patologia, comorbilidades e terapêuticas realizadas, e de se garantirem as condições de assepsia adequadas, sobretudo na presença de um doente imunodeprimido. Nestes casos, o recurso a outras formas de administração de terapêutica deve ser cuidadosamente ponderado.

CC199 – DOENÇAS IMUNOMEDIADAS INDUZIDAS POR PROPILTIOURACILO: DUAS APRESENTAÇÕES CLÍNICAS DIFERENTES

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Introdução: O propiltiouracil (PTU) é usado como 1ª linha no tratamento do hipertiroidismo. Este fármaco pode induzir respostas auto-imunes adversas como vasculite ANCA ou lúpus induzido por drogas. Este trabalho tem como objetivo apresentar uma serie de casos.
Caso Clínico 1: Mulher de 53 anos, enviada à nossa consulta por artralgias. Como antecedentes pessoais, destacava-se hepatite B crónica sob entecavir, hipotiroidismo subclínico (mas história de hipertiroidismo prévio sob PTU durante vários anos) e osteoporose sob zoledronato secundária a menopausa precoce por neoplasia do colo do útero. A doente referia poliartralgias inflamatórias, dispneia para médios esforços e sintomas gastrointestinais (náuseas, vómitos e enfartamento). Ao exame objectivo, apresentava dedos fusiformes mas sem artrite, alterações cutâneas ou da força muscular. Analiticamente, ligeira leucopenia (4x103/uL), teste de coombs indirecto positivo, VS 6, ANA 1/640 padrão AC-4 Nuclear finogranular (Ku 50 RU/

ml, RO-52 33 RU/ml), ANCA-MPO (59CU) e anticoagulante lúpico positivo em duas titulações, com consumo de complemento (C3 81mg/dL, C4 7mg/dL). Anticorpo anti-histonas e ds-DNA negativos. TC- tórax e ecocardiograma sem alterações relevantes. Ecografia renovesical a mostrar ligeira acentuação da diferenciação corticomedular. Quadro com sobreposição clínica e laboratorial de Lúpus Eritematoso Sistémico (LES) induzido por fármacos e vasculite por PTU. Iniciou hidroxicloroquina 400mg/dia e metilprednisolona 4mg/dia com resolução das queixas articulares.

Caso Clínico 2: Mulher de 57 anos, encaminhada por poliartralgias de ritmo inflamatório associadas a lesões cutâneas ao nível das mãos e joelhos com 3 meses de evolução. Referia também fenómeno de Raynaud com 1 ano de evolução, aftose oral frequente (>1 episódio/mês) e sintomas sicca. Como antecedentes pessoais destaca-se história trombótica (tromboflebites e AIT) com mutação MTHFR, encontrando-se hipocoagulada com varfarina e hipertiroidismo sob PTU. Ao exame objectivo, lesões arroxeadas nas mãos e livedo reticular, sem artrite. Analiticamente, leucopenia (2.76x103/uL), anemia (Hb 11.4mg/dL), VS 54, PCR 4.25mg/dl, ANCA PR3 positivo (242 CU), anticoagulante lúpico positivo e consumo de C3 (75mg/dL). Apresentava ainda creatinina de 0.71mg/dl, com razão proteínas/creatinina 196, e com proteinuria de 2g/24h tendo realizado biópsia renal, que não mostrou evidência de lesão renal. Por suspeita de vasculite ANCA induzida por PTU, suspende-se este fármaco e introduz-se tiamazol, associado a hidroxicloroquina e prednisolona 15mg/dia em esquema de desmame rápido até 5mg/dia. Dois meses após início de tratamento, melhoria das artralgias e remissão das lesões de vasculite cutânea, assim como redução do PR3 para 78.4 e aumento de C3 (87).

Conclusões: O risco de desenvolver LES induzido por drogas com o PTU é <1%, sendo que estes doentes apresentam maior risco de envolvimento músculo-esquelético, gastrointestinal e das serosas. Já na vasculite ANCA induzida por PTU o sistema respiratório e renal poderá ser envolvido. Os sintomas poderão iniciar-se de semanas a anos após início do tratamento, sendo que os doentes geralmente melhoram com a descontinuação do fármaco, podendo em 50% dos casos necessitar de corticóides ou imunossupressores. O objetivo deste trabalho é enfatizar a importância da alta suspeição clínica para a ocorrência de doenças imunomediadas pelo PTU e para as várias apresentações. O diagnóstico precoce e a suspensão do fármaco fazem a diferença na morbilidade da doença.

CC217 – SÍNDROME DE RAMSAY-HUNT EM DOENTE COM ARTRITE REUMATÓIDE SOB ETANERCEPT E LEFLUNOMIDA

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Introdução: Vários fatores contribuem para um aumento do risco infecioso na artrite reumatóide, tanto inerentes à própria doença, como relacionados com a medicação instituída, particularmente com o uso de terapêuticas biotecnológicas. A síndrome de Ramsay-Hunt (SRH) corresponde a uma reativação do vírus varicela-zoster (VVZ) no gânglio geniculado, com consequente clínica de parésia periférica no nervo facial, otalgia e exantema vesicular na região auricular. O Zoster Sine Herpete corresponde a um subtipo raro de SRH com parésia facial e otalgia sem exantema vesicular associado.

Caso Clínico: Doente do sexo feminino, 59 anos, com artrite reumatóide com 15 anos de evolução, tratada com hidroxicloroquina 400mg/dia e leflunomida 20mg/dia, após falência de resposta à terapêutica com metotrexato. Em maio de 2019, por manter atividade de doença, iniciou etanercept (50mg subcutâneo/semana). A 20 de junho de 2019 iniciou otalgia à esquerda e no dia seguinte, efetuou a toma semanal de etanercept. Um dia depois referia agravamento da otalgia com irradiação para a região cervical, náuseas e astenia, com temperatura axilar de 37.5ºC. A 22 de junho recorreu ao serviço de urgência com desvio da comissura labial para a esquerda e vertigem. Negava outras queixas de orgãos e sistemas. O exame neurológico demonstrou apenas parésia facial periférica à esquerda e nistagmo horizonto-rotatório com fase rápida para a direita. Não apresentava lesões cutâneas no pavilhão auricular ou noutros locais. A avaliação por Otorrinolaringologia não revelou alterações na otoscopia e na acumetria. A tomografia computorizada crânio-encefálica não mostrou lesões isquémicas agudas e o estudo analítico não evidenciou quaisquer alterações, nomeadamente elevação dos parâmetros inflamatórios ou alterações do sedimento urinário, e a radiografia do tórax foi normal. A punção lombar mostrou 146 células/uL (145 leucócitos/uL, com 4.1% de polimorfonucleares) com normoglicorrária e normoproteinorráquia. Foi assumida parésia facial de provável etiologia vírica com atingimento do sistema nervoso central (meningite de líquor claro) e iniciou aciclovir endovenoso no internamento do serviço de Infecciologia. As hemoculturas foram negativas e o

estudo microbiológico do líquor revelou pesquisa de DNA- VVZ positiva. Foram excluidas outras etiologias como Borreliose de Lyme. Durante o internamento cumpriu 10 dias de aciclovir (600mg e.v. 8/8 horas) e 5 dias de prednisolona (1mg/kg/dia), tendo alta medicada com pregabalina (200mg/dia) pela dor neuropática e beta-histina como terapêutica anti-vertiginosa. Foi orientada para reabilitação motora da parésia facial. Três meses após o internamento, a doente encontrava-se já sem dor neuropática, com melhoria parcial do desiquilíbrio e da parésia facial, pela qual mantem programa fisiátrico. Atualmente, encontra-se em ponderação a reintrodução da terapêutica biotecnológica, tendo em conta o recrudescimento das queixas articulares.

Conclusão: Apresentamos um caso raro de Zoster Sine Herpete, um subtipo raro de SRH, numa doente com artrite reumatóide sob etanercept e leflunomida. Este caso realça a importância da vigilância e do reconhecimento atempado de intercorrências infeciosas menos comuns nos doentes imunodeprimidos com patologia reumática inflamatória.

CC219 – PARANEOPLASTIC RHEUMATIC SYNDROME: A CHALLENGING DIAGNOSIS

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Introduction: Paraneoplastic rheumatic syndromes (PRS) are more frequently associated with hematologic, lung, breast and ovary malignancies and pose a diagnostic challenge to the physician. Differential diagnosis with primary rheumatic disorders might be complex since the onset of rheumatic manifestations can occur at any point in the neoplastic disease's timeline: up to two years before, at or after the diagnosis of malignancy. The most frequent presentations are arthritis (poli, oligo or, rarely, monoarthritis), hypertrophic osteoarthropathy, dermatomyositis/polymyositis and vasculitis. Hypertrophic osteoarthropathy is a syndrome characterized by abnormal proliferation of the skin and osseous tissue at the distal parts of the extremities, which includes digital clubbing, periostosis of tubular bones and synovial effusions. It can occur primarily or secondarily to other conditions, mainly intrathoracic neoplastic processes. The authors present two cases of hypertrophic osteoarthropathy as primary manifestation of lung adenocarcinoma.

Case reports:

Case 1: A previously healthy 49-year-old man was referred by his family doctor (FD) to our department due to a seven-month history of persistent pain in several joints (elbows, knees and ankles) which responded partially to naproxen 500mg twice a day. He denied personal or family history (FH) of rheumatic disease, psoriasis, inflammatory bowel disease or malignancy. The patient was an active smoker (25 pack-year). The only positive finding during clinical examination was the presence of digital clubbing. No arthritis, cutaneous abnormalities, cyanosis or lymphadenopathies were present. Despite having a normal chest X-ray from five months prior, a new x-ray was prescribed and a thoracic mass was present. Blood tests showed only a slight inflammatory markers' increase. Further investigation revealed a lung adenocarcinoma.

Case 2: A 70-year-old man, with previous history of curative prostate cancer surgery and 55 pack-years of active smoking, was referred by his FD to our department due to a 7-month onset of unilateral knee pain with a predominantly mechanic pattern, edema and functional limitation with a normal x-ray. There was a positive FH of malignancy (prostate, liver and ovarian cancer). During clinical examination, only digital clubbing and labial cyanosis were found. The patient showed a good response to NSAIDs. A chest X-ray was prescribed and a thoracic mass was present. Blood tests showed normocytic/normochromic anemia (Hb 11,3 g/dl), thrombocytosis and a slight increase in inflammatory markers. Further investigation revealed a lung adenocarcinoma.

Both cases are undergoing oncologic treatment, having shown resolution of the articular symptoms.

Conclusion: PRS are a group of manifestations with a variable spectrum of symptoms, making the diagnosis process a real challenge. Since there are no specific laboratory/imaging findings, the diagnosis of paraneoplastic depends on high clinical suspicion. The clinician should take into account the patient's characteristics – gender, age, arthritis presentation – and, simultaneously, exclude other etiologies for the clinical setting, such as primary rheumatic disorders or infection. Furthermore, the presence of hypertrophic osteoarthropathy should raise the hypothesis of intra-thoracic neoplasm, particularly lung cancer. Symptomatic treatment with analgesics and NSAID, as well as neoplastic treatment, relieves most cases.

CC222 – GOTA TOFÁCEA OU ALGO MAIS? – A IMPORTÂNCIA DA HISTÓRIA CLÍNICA EM REUMATOLOGIA

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Introdução: Existem mais de cem doenças reumáticas. Muitas delas apresentam manifestações articulares e extra-articulares, mais ou menos exuberantes e formas de apresentação muito variáveis, o que muitas vezes levanta incertezas diagnósticas. A realização de uma história clínica cuidada, de um exame objetivo pormenorizado e o pedido criterioso de exames auxiliares de diagnóstico, são a chave do diagnóstico. O caso em estudo ilustra as dificuldades que por vezes surgem com vista ao diagnóstico definitivo de certos quadros reumatológicos.
Caso clínico: Homem de 51 anos, condutor de pesados, com antecedentes de dislipidémia, hipertensão arterial, hiperuricémia, excesso ponderal, psoríase cutânea e ungueal, com hábitos etílicos e tabágicos mantidos e antecedentes de consumo de drogas ilícitas. Apresentava episódios recorrentes de mono/oligoartite de instalação súbita, afetando, ao longo do tempo, as articulações metacarpo-falângicas, punhos, tarsos e primeiras metatarso-falângicas, com boa resposta ao tratamento com anti-inflamatórios não esteroides e Colquicine e com períodos inter-críticos assintomáticos. Desenvolveu, recentemente, gonartrite bilateral, de instalação progressiva, astenia e mialgias difusas.
Ao exame objetivo apresentava lesões de psoríase cutânea e ungueal, tofos gotosos nas polpas digitais nas mãos e gonartrite bilateral.
Analiticamente com hemograma, função renal e parâmetros inflamatórios (VS e PCR) normais, Ac anti-CCP e Fator Reumatóide negativos, uricémia: 11,8 mg/dl, TGO: 45; TGP: 53, Ac e Ag HCV positivos, carga viral e genótipo para HCV ainda pendentes, serologias para HIV e HBV negativas. No estudo radiológico destacam-se erosões nas 1ª MTFs. A artrocentese de ambos os joelhos revelou um líquido com características inflamatórias, com pesquisa de cristais e exame cultural pendentes. A ecografia hepática mostrou esteatose difusa.
Tendo em conta as alterações analíticas e descrição das queixas, foram colocadas as hipóteses diagnósticas de gota tofácea em associação com quadro de eventual Artrite Psoriática ou manifestação articular da infeção por HCV.
De momento o doente está medicado com alopurinol e colquicine.
Discussão: Neste caso o raciocínio diagnóstico torna-se complexo, dado que se por um lado as queixas articulares anteriores têm características inflamatórias intermitentes e são compatíveis com artrite microcristalina por cristais de monourato de sódio (tanto mais que apresenta tofos gotosos), por outro lado o quadro sistémico atual e as queixas atuais dos joelhos podem estar associadas à psoríase ou à infeção por HCV, encontrando-se estas hipóteses ainda em estudo.
Conclusão: Este caso clínico realça as dificuldades diagnósticas com que nos deparamos na prática clínica diária e motivaram a realização de uma revisão aprofundada das manifestações musculo-esqueléticas da gota, artrite psoriática e associadas à infeção pelo VHC, com vista ao esclarecimento diagnóstico adequado deste quadro clínico.

CC232 – GRANULOMATOSE EOSINOFÍLICA COM POLIANGEÍTE (GEP): APRESENTAÇÃO ATÍPICA COM MIOCARDITE

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Introdução: A Granulomatose Eosinofílica com Poliangeíte (GEP) é uma vasculite de pequenos vasos que frequentemente se associa à presença anticorpos anti-citoplasma de neutrófilos (ANCA) [1, 2]. Apesar de incomum, o envolvimento cardíaco constitui uma das manifestações mais graves, associando-se a elevada mortalidade.
Caso Clínico: Mulher, 48 anos, com antecedentes de asma (diagnosticada aos 35 anos, sob montelucaste), pólipos nasais e rinite alérgica, recorreu às urgência por quadro clínico com 5 meses de evolução, de astenia, dispneia em repouso, ortopneia, edema dos membros inferiores e artralgias migratórias, de predomínio noturno, na região dos ombros, cotovelos e punhos bilateralmente, com noção de edema difuso das mãos. Associadamente referiu diminuição da sensibilidade sensitiva do membro inferior direito. Ao exame objetivo apresentava fervores bibasais e edema periférico dos membros inferiores, sem outras alterações. Do estudo complementar salientou-se leucocitose (21.1G/L), eosinofilia periférica (11.28G/L), elevação de PCR (PCR 6.63mg/dl), aumento de LDH e CK (775U/L e 420U/L, respetivamente), elevação da troponina (9698.7ng/L) e do péptido natriurético tipo B (BNP) 2047.2 pg/ml. O Ecocardiograma revelou depressão da função do ventrículo esquerdo, derrame pericárdico ligeiro e derrame pleural volumoso. Foi

assumido o diagnóstico de miocardite com disfunção cardíaca sistólica (fração de ejeção de 31%) tendo sido admitida na Unidade de Tratamento de Insuficiência Cardíaca Avançada. Realizou tomografia computorizada de alta resolução do tórax que mostrou áreas com padrão em vidro despolido.

Pelo elevado índice de suspeição de GEP, apesar de apresentação atípica, foi discutido o caso em equipa multidisciplinar de reumatologia e cardiologia tendo-se optado pela realização de biópsia do miocárdio e tratamento imediato com pulsos de metilprednisolona 1g, durante 3 dias. O resultado posterior da biópsia revelou infiltração eosinofílica do miocárdio, cumprindo critérios ACR 1990 para GEP. A doente iniciou regime Cyclops (15mg/Kg, até um máximo de 1g, associado a prednisolona 1mg/Kg/dia). Após estabilização clínica teve alta com diurético, anti-hipertensor, beta-bloqueante, corticoterapia e ciclofosfamida (CYC).

Durante o follow-up de 7 meses, a doente evoluiu favoravelmente, no entanto sem melhoria significativa da função cardíaca. Em reunião multidisciplicar e com expert internacional optou-se por completar 10 ciclos de tratamento com CYC, a cada 3 semanas, com redução após cada pulso da dose de prednisolona e switch para azatioprina 2mg/Kg/dia após completado o 10º ciclo. Atualmente, mantém seguimento multidisciplinar em cardiologia e reumatologia.

Conclusão: Este caso descreve uma situação particularmente incomum e potencialmente fatal, não só pelo envolvimento cardíaco (taxa de mortalidade até 50%), mas também por a dispneia ser o primeiro sintoma de alarme e que conduziu ao diagnóstico. Ilustra ainda a importância da elevada suspeição para o diagnóstico de GEP, a necessidade de ter presente esta entidade entre os diagnósticos diferenciais de insuficiência cardíaca, bem como a importância de serem criados consensos de tratamento para este tipo de envolvimento.

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CC247 – O USO DA PET-FDG NO DIAGNÓSTICO DE ARTERITE DE CÉLULAS GIGANTES – EXPERIÊNCIA DE UM CENTRO E BREVE REVISÃO DA LITERATURA

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O grupo das vasculites de grandes vasos inclui a arterite de Takayasu e a arterite de células gigantes (ACG), sendo esta última a vasculite primária mais frequente em adultos. Classicamente, o gold standard para o diagnóstico da ACG é a biópsia da artéria temporal, mas esta tem vindo a ser ultrapassada por métodos de imagem menos invasivos, mais rápidos e acessíveis e com capacidade de avaliar outros territórios arteriais, como é o caso da tomografia por emissão de positrões com [18]fluorodesoxiglucose (PET-FDG), particularmente útil nos doentes que se apresentam com sintomas inespecíficos e em que o diagnóstico é muitas vezes um desafio. Nas mais recentes recomendações

TABELA 1 – CARACTERÍSTICAS, FORMA DE APRESENTAÇÃO E EXAMES COMPLEMENTARES DE DIAGNÓSTICO DE CADA DOENTE

Doente	Sexo	Idade ao diagnóstico	Apresentação inicial	Territórios com hipermetabolismo na PET-FDG inicial	Ecografia das artérias temporais	Realização de biópsia da artéria temporal
1	F	75	Quadro consumptivo e queixas de polimialgia reumática	Tronco braquicefálico; carótidas comuns; artérias subclávias e axilares; aorta torácica e abdominal; artérias femorais.	Sem sinais de vasculite	Não
2	M	58	Febre, toracalgia e episclerites e esclerites de repetição	Aorta torácica; pequena extensão da carótida comum direita e da aorta abdominal	Sem sinais de vasculite	Não
3	F	60	Dor abdominal	Aorta torácica, aorta abdominal e região proximal das ilíacas comuns	Sem sinais de vasculite	Não
4	F	59	Febre	Artérias subclávias; carótidas; aorta torácica e abdominal; artérias ilíacas; femurais; porção proximal das tibiais posteriores	Sem sinais de vasculite	Não

da European League Against Rheumatism para o uso de métodos de imagem no diagnóstico de vasculites de grandes vasos, a PET está recomendada como método para detecção de inflamação em artérias extracranianas por forma a suportar o diagnóstico de ACG. Descrevemos a experiência de um serviço de Reumatologia com o uso da PET/CT-FDG no diagnóstico de ACG e fizemos uma breve revisão da literatura sobre este tema.

Foram identificados os casos de ACG diagnosticados entre 2017 e 2019 por meio de PET/CT-FDG. Trata-se de 4 doentes, 3 do sexo feminino, com idades compreendidas entre os 60 e os 75 anos, com apresentação clínica inicial com sintomas sistémicos e inespecíficos. Todos realizaram PET/TC-FDG como parte da abordagem diagnóstica, que revelou hipermetabolismo da parede arterial de múltiplos territórios arteriais. Todos foram submetidos a ecografia das artérias temporais que não revelou aspectos compatíveis com envolvimento vasculítico destes vasos. A tabela em anexo apresenta as características, apresentação e achados na PET-FDG de cada doente.

Vários estudos têm demonstrado a performance da PET-FDG no diagnóstico das vasculites de grandes vasos e em particular da ACG. Numa meta-análise de 2016, a pooled analysis revela uma sensibilidade de 83.3% e uma especificidade de 89.6% no diagnóstico de arterite de células gigantes. Mais recentemente, um estudo prospectivo duplo-cego demonstrou uma sensibilidade e especificidade da PET/TC-FDG de 92% e 85%, respectivamente, quando comparado com a biópsia da artéria temporal, e de 71% e 91% quando comparado com o diagnóstico clínico. Quando utilizada com intuito diagnóstico já após a instituição de terapêutica, a PET-FDG deve ser realizada o quanto antes de forma a aumentar o seu rendimento: um estudo de 2018 mostrou uma sensibilidade diagnóstica mantida ao fim de 3 dias de terapêutica com corticoesteróides, mas diminuída aos 10 dias.

Em conclusão, a nossa experiência, em linha com os dados da literatura, suporta a utilidade da PET-FDG no diagnóstico de ACG, particularmente em doentes que se apresentam com quadros clínicos atípicos e sem envolvimento craniano.

CC251 – ADALIMUMAB E ANGIOEDEMA: A PROPÓSITO DE UM CASO CLÍNICO

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Introdução: Os fármacos biotecnológicos têm sido cada vez mais utilizados nas doenças reumáticas inflamatórias crónicas, uma vez que conferem um melhor controlo clínico, analítico e imagiológico, modificando a progressão natural da doença. Destes, os mais utilizados são os anti-TNFα. Apesar dos seus benefícios, é importante conhecer o seu perfil de efeitos adversos. Os efeitos laterais mais frequentemente documentados na literatura são: reação no local da picada, rash cutâneo, intolerância gastrointestinal, aumento do risco infecioso e desenvolvimento de anticorpos antinucleares e anti-DNA nativo. A propósito de um caso clínico, os autores descrevem um efeito adverso pouco frequente: o angioedema.

Caso clínico: Mulher de 56 anos, com antecedentes de hipertensão arterial e dislipidemia, com o diagnóstico de artrite reumatoide erosiva e seropositiva (para fator reumatóide e anticorpos anti-CCP) desde há mais de 20 anos, com envolvimento predominante das pequenas articulações (mãos, punhos e pés). Esteve medicada, inicialmente, com metotrexato oral 20 mg/semana durante 4 anos, que suspendeu por ineficácia e, posteriormente, com leflunamida 20 mg/dia. Por manter agravamento clínico e imagiológico, iniciou tratamento com adalimumab (Humira) 40 mg s.c., quinzenal. Às 10 semanas de tratamento, por ainda manter elevada atividade da doença pelo DAS 28 4V (8.093), procedeu-se a um incremento na dose do adalimumab, que passou a ser de administração semanal. Ao primeiro mês após o ajuste posológico, apresentou o primeiro episódio de angioedema com envolvimento da região labial da hemiface direita, que resolveu com anti-histamínico e glucocorticoides. Posteriormente, apresentou mais três episódios no mês seguinte, sendo que o último foi mais grave, com envolvimento das vias aéreas superiores e a exigir tratamento endovenoso com adrenalina. Do estudo realizado, destacam-se: C3, C4, C1q e inibidor de C1 sem alterações; imunoglobulina E sérica total normal e testes de Prick sem alterações. A doente negou consumo de novos medicamentos ou alimentos diferentes dos habituais. Optou-se por interromper definitivamente o adalimumab, mantendo-se medicada com leflunamida 20 mg/dia. Após a suspensão do adalimumab, a doente apresentou apenas mais dois episódios de edema facial de menor intensidade nos primeiros três meses, sem recorrência desde então.

Após 6 meses da suspensão do adalimumab e face à necessidade de um melhor controlo da atividade da artrite reumatoide, iniciou golimumab em regime hospitalar, com vigilância nas primeiras 48 horas, o que decorreu sem intercorrências.

Conclusão: Os agentes anti-TNFα não são isentos de complicações e, raramente, associam-se a complicações graves, que devem ser reconhecidas. Segundo

a literatura, a segurança do adalimumab 40 mg s.c. em dose semanal é semelhante à em dose quinzenal, sendo muito equiparáveis em termos de perfil de eventos adversos. Esta evidência é transversal a várias patologias inflamatórias crónicas. Este caso clínico relata o aparecimento de angioedema após o incremento da posologia do adalimumab para toma semanal, com resolução após a suspensão do fármaco. Não foram identificados outros potenciais fatores causadores. A prevalência de angioedema com o adalimumab é inferior a 1% e estão descritos casos de angioedema secundário ao adalimumab vários meses após a primeira toma, parecendo ser um efeito lateral independente do tempo de exposição. É também de realçar que o aparecimento de efeitos laterais com um anti-TNFα, não contraindica o uso de um outro agente anti-TNFα, como este caso bem documenta.

CC252 – MANIFESTAÇÕES RARAS DE DOENÇAS RARAS – DOENÇA DE STILL DO ADULTO

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Introdução: A doença de Still do adulto (DSA) é uma entidade inflamatória sistémica rara de etiologia desconhecida. A apresentação clínica é heterogénea, sendo as manifestações mais típicas são a febre diária, um rash evanescente e artralgia/artrite. Não raras vezes, outros sinais e sintomas surgem em associação à doença tornando o diagnóstico, em parte, de exclusão e causando atraso na sua identificação. Manifestações como o síndrome de stress respiratório agudo (SSRA), o síndrome de ativação macrofágica (SAM) e a miopericardite são raras mas potencialmente graves e potencialmente fatais.

A doença pode seguir um curso monofásico, intermitente ou crónico com igual distribuição dos doentes nos três padrões.

Caso clínico: Apresenta-se o caso de uma mulher de 56 anos, com antecedentes de múltiplas faringites no último ano, previamente internada noutra instituição por quadro com 6 semanas de odinofagia, astenia, febre (>39.5ºC), rash maculopapular evanescente, não pruriginoso, distribuído pelo tronco, membros, com acometimento de palmas e plantas e com fenómeno de Koebner. Foi instituída terapêutica antibiótica, progressivamente escalada, mas sem evidencia de melhoria. Realizou extenso estudo etiológico do qual se destacam alguns achados relevantes: hepatomegalia, adenopatias simétricas em múltiplas cadeias ganglionares, anemia (9.2 g/dL), leucocitose (19.500/µL) com 94% de neutrófilos, elevação modesta das aminotransferases (AST 81 U/L; ALT 64 U/L), negatividade para anticorpos antinucleares e fator reumatoide, elevação de proteína C reativa (PCR) (182 mg/L) e aumento da velocidade de sedimentação (VS) (78 mm/h).

Foi transferida para a nossa instituição por suspeita de síndrome coronária aguda após início de queixas de dor retrosternal em peso e intensa dispneia. À admissão, documentado SSRA, miopericardite, derrame pleural, pericárdico e líquido ascítico em pequena quantidade. Analiticamente foi evidente pancitopenia (hemoglobina 6.1 mg/dL; leucócitos 3800/µL; plaquetas 113000/µL), ferritina de 60847 ng/mL, aumento marcado dos triglicerídeos (754 mg/dL) e o decréscimo paradoxal da VS (41 mm/h) acompanhado de aumento de PCR (248 mg/L).

Avaliada em regime de internamento após permanência em unidade de cuidados intermédios durante 5 dias, com o diagnóstico provisório de síndrome de resposta inflamatória sistémica em contexto de pneumonia, medicada com antibioterapia e corticoterapia endovenosa intermitente pelo broncospasmo. Apesar dos múltiplos fatores confundidores decorrentes do percurso complexo da doente nos cuidados de saúde, foi colocada a hipótese de se tratar de um caso de doença de Still do adulto, cumprindo critérios Yamaguchi, complicada com provável SAM, SSRA e miopericardite. Foi suspensa a antibioterapia e iniciada terapêutica com prednisolona na dose de 0.5mg/Kg/dia. A doente apresentou progressiva melhoria sintomática e analítica precoce reunindo condições para alta clínica. Reavaliada em consulta externa 1 mês após o episódio de internamento, sem evidencia de recidiva, normalização das contagens hematológicas e dos parâmetros inflamatórios.

Discussão: No caso apresentado retrata-se a complexidade no diagnóstico quando apresentações raras de doenças raras se conjugam.

Com a apresentação deste caso os autores pretendem alertar para a necessidade de reconhecimento desta entidade bem como de algumas formas de apresentação menos comuns na doença de Still do adulto como são o SSRA, o SAM e a miopericardite.

CC254 – ENVOLVIMENTO CARDÍACO NA DOENÇA DE BEHÇET – UMA APRESENTAÇÃO ATÍPICA

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Introdução: A Doença de Behçet é uma patologia inflamatória multissistémica, que se carateriza classicamente pela presença de aftose oral e genital recorrentes, uveítes e lesões cutâneas. No entanto, pode associar-se a diversas outras manifestações de órgãos e sistemas, nomeadamente neurológico, vascular, articular e gastrointestinal. O atingimento cardíaco é raro (afeta até 5% dos doentes) e inclui o envolvimento miocárdico, pericárdico, valvular, coronário ou a formação de trombos intracardíacos. A sua distribuição geográfica é também distinta, sendo mais frequente no médio e extremo orientes.

Caso Clínico: Mulher, 25 anos, de ascendência egípcia, com história passada de derrame pericárdico de ligeiro volume de etiologia desconhecida, documentado cerca de 2 anos antes no contexto de quadro de dor torácica e palpitações, cujo estudo analítico imunológico, serológico e virológico foi negativo e que resolveu com curso de prednisolona 50mg id durante 3 meses, foi referenciada à Consulta Externa de Reumatologia por quadro de poliartralgias de ritmo inflamatório e aditivas acometendo joelhos, tornozelos, antepés, ombros, cotovelos, metacarpofalângicas e interfalângicas proximais das mãos, com rigidez matinal de cerca de 30 minutos, associada a aftose oral recorrente com cerca de 1 ano de evolução. Sem outra sintomatologia associada e sem artrite objetivável ao exame físico. Repetiu estudo laboratorial extenso que não revelou alterações face à avaliação inicial, bem como o ecocardiograma transtorácico, tendo este evidenciado a presença de fina lâmina de derrame pericárdico, sem comprometimento da função sistólica. Após cerca de 2 anos de seguimento, a doente apresentou episódio de lesões de aftose genital, que após observação por Ginecologia e com a exclusão de outras causas mais frequentes, foram explicadas no quadro provável de Doença de Behçet. Colheu ainda análises com HLAB51, que foi negativo. Em concordância com a Cardiologia, a etiologia do derrame pericárdico enquadrou-se igualmente na doença inflamatória sistémica. Foi instituída colchicina 1mg id, com melhoria da sintomatologia.

Discussão/conclusão: O caso presente retrata uma apresentação atípica de Doença de Behçet, em que o atingimento cardíaco, para além de uma complicação rara, foi a manifestação inaugural da doença. Salienta também a importância da avaliação de outras manifestações sistémicas que numa fase inicial podem ser únicas numa patologia com tão amplo espectro clínico, e que apesar de excecionais, conferem habitualmente um prognóstico ominoso ao doente.

CC260 – RHUPUS – QUANDO A ARTRITE REUMATÓIDE SE SOBREPÕE AO LÚPUS ERITEMATOSO SISTÉMICO

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Introdução: A artrite é uma manifestação comum de diversas doenças imunomediadas, nomeadamente o lúpus eritematoso sistémico (LES) e a artrite reumatóide (AR), duas entidades patológicas com diferentes características clínicas e sorológicas. Em até 90% dos doentes com LES existe atingimento articular, frequentemente na forma de sinovite ligeira simétrica de pequenas e médias articulações, com deformidade articular tardia de caráter não erosivo descrita em até 35% dos doentes. Porém, em apenas 3 a 5% dos casos, artropatia severa erosiva indistinguível da presente na AR pode ser observada. Esta entidade clínica é tradicionalmente descrita como "rhupus", existindo controvérsia relativa à sua significância: síndroma de sobreposição entre AR com LES, ou quadro articular severo como parte integrante de LES. O seu diagnóstico é considerado na presença de artrite erosiva seguindo a distribuição característica de AR, associada a presença de marcadores serológicos de ambas as entidades: anticorpos antinucleares (ANAs), nomeadamente anti-SSA e anti-SSB, fator reumatóide (FR) e anticorpos anti-péptido citrulinado cíclico (anti -CCP). A sua prevalência estimada na população geral ronda os 0,09%.

Caso Clínico: Mulher de 48 anos de idade com diagnóstico prévio de LES e sob hidroxicloroquina 400 mg diárias que é referenciada à consulta de Reumatologia por quadro de poliartrite crónica afetando predominantemente os ombros, mãos e pés. Ao exame objetivo denotaram-se lesões cutâneas compatíveis com lúpus cutâneo crónico (confirmado anteriormente em biópsia cutânea). Do estudo complementar realizado, salienta-se a positividade para os ANAs (apresentando um padrão mosqueado), anti-histonas, anti-dsDNA (32 U/ml), FR e anti-CCP em títulos altos (637,0 UI/mL e > 300 UA /mL, respetivamente). No entanto, no estudo radiográfico das mãos e pés não foi evidenciada artropatia erosiva.

Foi assumido o diagnóstico de síndrome de sobreposição AR/LES ("Rhupus"), tendo sido introduzido tratamento com prednisolona 5 mg diárias e metotrexato 15 mg semanais suplementadas com 10 mg semanais de ácido fólico. O tratamento com HCQ foi mantido

durante o seguimento efetuado e a poliartrite melhorou progressivamente.
Discussão: Este caso retrata de forma evidente a forma como a interligação entre as várias doenças do tecido conjuntivo pode condicionar a apresentação de fenótipos de diferentes doenças em simultâneo num mesmo indivíduo. Devemos salientar que os autores estão cientes que a ausência de erosões poderá pôr em dúvida a sobreposição destas duas entidades. No entanto, tendo a doente sido identificada numa fase relativamente precoce do início das suas queixas articulares, é possível que esta seja uma das explicações para a ausência de dano estrutural. Além disso, a sinovite exuberante e a presença de FR e anti-CCP em títulos altos são, pelo menos, extremamente atípicos numa situação de LES isolado. Estes casos representam um desafio terapêutico acrescido, sendo a corticoterapia e os fármacos modificadores de doença (DMARDs) convencionais o esteio do tratamento na maioria dos doentes. No entanto, os condicionalismos inerentes a cada uma destas condições devem sempre estar presentes, o que deve contraindicar, por exemplo, o uso de agentes anti-TNF nos casos refratários, pois poderiam precipitar um agravamento do quadro de LES subjacente. Terapia com agentes biotecnológicos como o rituximab ou abatacept demonstrou resultados promissores nos doentes com reduzida resposta aos DMARDs convencionais.

CC261 – ENVOLVIMENTO PERICÁRDICO NAS DOENÇAS INFLAMATÓRIAS SISTÉMICAS – UMA REVISÃO DE CASOS

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Introdução: O envolvimento cardíaco não é incomum nas doenças inflamatórias sistémicas, podendo envolver o pericárdio sob a forma de pericardite aguda ou recorrente ou derrame pericárdico (assintomático), e constitui-se habitualmente como um fator de mau prognóstico. Apesar de para o diagnóstico definitivo ser mandatória a histologia ou alterações caraterísticas no derrame, na prática o diagnóstico é apenas de presunção. O tratamento é guiado pelos sintomas, sendo fundamental controlar a atividade da doença sistémica subjacente, o que pode requerer terapia imunossupressora mais agressiva.
Descrição dos casos: No ano de 2019 foram internados no nosso centro 5 casos de envolvimento pericárdico no contexto de doença inflamatória sistémica, dos quais 4 do sexo feminino, com idades compreendidas entre os 30 e os 60 anos, 2 com artrite reumatoide (AR), 2 com lúpus eritematoso sistémico (LES) e 1 com esclerose sistémica, com duração de doença entre os 3 e os 12 anos. Três dos doentes eram fumadores, 2 tinham dislipidemia, e nenhum tinha hipertensão arterial ou diabetes. No que toca a terapêutica para a doença de base, 2 realizavam corticoterapia, os 2 casos de LES estavam sob hidroxicloroquina e micofenolato de mofetil, e os outros 3 doentes realizavam metotrexato. Três casos apresentaram-se como pericardite aguda (um dos quais com quadro de síndrome gripal prévio), e 2 como derrame pericárdico assintomático, com valores de PCR entre os 2,3 e 176,8 mg/L, e VS entre 11-107 mm/1ª hora. Na avaliação ecocardiográfica, há a registar 2 casos sem derrame pericárdico objetivável, 1 derrame de pequeno, 1 de médio e outro de grande volume. Em nenhum dos casos foram identificadas complicações como hipertensão pulmonar ou redução da função sistólica biventricular. Colchicina foi usada em 2 casos e anti-inflamatórios não esteroides em 3, sendo que em 4 casos o controle da atividade da doença de base levou ao incremento ou introdução de corticoterapia. Nenhum doente foi submetido a pericardiocentese ou pericardiotomia para confirmação do diagnóstico, dada a evolução favorável em todos os casos. A doente que realizou apenas terapêutica sintomática, com AR, foi o único caso que foi reinternado pelo mesmo motivo, um mês após a alta.
Conclusão: O reconhecimento precoce deste tipo de envolvimento por parte das doenças inflamatórias sistémicas é fundamental. Apesar de a doença de base poder não exibir outros sinais de atividade e os parâmetros inflamatórios poderem não estar alterados, a imunossupressão adequada é fundamental para o prognóstico do doente com envolvimento pericárdico no contexto de doença inflamatória sistémica.

CC264 – ORELHAS A ARDER!

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Introdução: A condrite auricular é rara e inclui múltiplos diagnósticos diferenciais, incluindo traumatismos, infeções e doenças autoimunes. Entre estes diagnósticos inclui-se a policondrite recidivante (PR), uma patologia inflamatória sistémica rara e de etiologia desconhecida, que se caracteriza por episódios

recorrentes de inflamação de estruturas cartilaginosas que podem resultar em lesão ou destruição dos tecidos. Além disso, pode também atingir tecidos ricos em proteoglicanos como o olho, vasos sanguíneos, coração e pele.

Caso Clínico: Mulher de 52 anos, com antecedentes de tiroidite de Hashimoto e de diabetes mellitus tipo 2 não insulinodependente, referenciada à consulta de Reumatologia por quadro com 5 anos de evolução de episódios recorrentes de artralgias de ritmo misto, atingindo pequenas articulações das mãos, joelhos e tornozelos. As crises acompanhavam- se de dor, tumefação e eritema dos pavilhões auriculares, com noção de aumento da temperatura local. Referia ainda astenia e enxaquecas frequentes. Negava outros sintomas e entre crises ficava assintomática. Nas crises já tinha sido medicada com AINE's e/ou corticoides em esquema de desmame com resolução dos episódios. A doente referia que a frequência das crises aumentou de 1 para 3-4 /ano.

À data da primeira consulta, a doente não se apresentava em crise e o exame objetivo não apresentava quaisquer alterações.

Laboratorialmente, destacavam-se anticorpos antinucleares 1/320, com restante estudo autoimune negativo, nomeadamente ANCA, e PCR e VS dentro dos valores normais. Serologias infeciosas eram negativas.

O ecocardiograma revelou fibrose da válvula mitral com insuficiência ligeira e da válvula aórtica sem compromisso funcional significativo, e ainda derrame pleural mínimo, sem compromisso hemodinâmico. Nas ecografias abdominal e renal não se observaram alterações.

A doente recebeu indicação para voltar à consulta no caso de ter novo episódio de sintomas para reavaliação e repetição do estudo analítico.

Em novembro de 2019 regressa à consulta com novo episódio de condrite auricular e artralgias. Ao exame objetivo apresentava tumefação exuberante dos pavilhões auriculares sem artrite periférica. Repetiu estudo analítico, que não revelou alterações. Assumiu-se o diagnóstico de PR e foi medicada com prednisolona 15 mg em esquema de desmame, com resolução do quadro.

Em dezembro, após desmame de corticoide, volta à consulta com recidiva da condrite auricular. Atendendo à recorrência da condrite, foi iniciada terapêutica com metotrexato (4 cp/semana) e esquema de prednisolona 5 mg id durante 2 semanas.

Atualmente, a doente mantém-se assintomática sob terapêutica com metotrexato, com bom controlo dos sintomas articulares e sem novos episódios de condrite auricular.

Discussão: A PR é uma doença rara com uma incidência estimada de 3,5 casos por milhão. O seu pico de incidência é na quinta década de vida e pode associar-se, em 30% dos casos, a outras doenças autoimunes.

A condrite auricular é a forma mais frequente de apresentação, mas pode ter manifestações diversas. O seu diagnóstico é clínico mas desafiante pela variabilidade e carácter intermitente das manifestações. A abordagem terapêutica depende das manifestações clínicas. AINE's e corticosteroides são os fármacos de 1ª linha no tratamento das formas leves a moderadas da doença. Os agentes imunossupressores como a azatioprina, o metotrexato e a ciclosporina são opções terapêuticas nos casos refratários.

CC276 – MIELITE TRANSVERSA EM DOENTE JOVEM: CASO RARO DE NEURO-BEHÇET

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Introdução: A doença de Behçet trata-se de um distúrbio crónico multissistémico caraterizado por aftose oral e genital recorrente, lesões cutâneas e manifestações oftalmológicas, neurológicas e articulares. Não tem atingimento preferencial por género, observando-se um pico de incidência entre as 3ª e 4ª décadas de vida. A presença do alelo HLA-B51 foi descrita como estando associada à doença. Verifica-se envolvimento neurológico em 5 a 10% dos doentes, podendo-se manifestar na forma de síndromes do tronco cerebral e do trato corticoespinal ("Neuro-Behçet"). Atribui-se pior prognóstico a doença com envolvimento parenquimatoso ou do tronco cerebral, sintomas cerebelosos ou anomalias no líquor.

Caso clínico: Homem de 40 anos que recorre ao serviço de urgência por quadro de dor dorso-lombar aguda com 5 dias de evolução irradiando para o flanco direito, associada a astenia, parestesias no membro inferior direito e alteração do controlo de esfíncteres. Ao exame neurológico verificou-se dor à palpação da região dorso lombar mediana na região de D10, com nível sensitivo a esse nível, monoparésia dolorosa do membro inferior direito e reflexo cutâneo-plantar em extensão à direita. Ressonância magnética nuclear (RMN) dorso-lombar revelou imagens intramedulares hiperintensas em ponderação T2, a nível de D8-D9, sem realce após gadolínio e de características inespecíficas. Realizou RM cranioencefalica que não revelou alterações de relevo.

A nível analítico de salientar leucocitose com

neutrofilia (13,4 x 10^9), sem elevação de parâmetros inflamatórios, líquido cefalorraquidiano com características fisiológicas, sem alterações ao exame citoquímico, bacteriológico e pesquisa de vírus neurotrópicos negativa. Pesquisa de bandas oligoclonais, anticorpos contra a glicoproteína da mielina do oligodendrócito e contra a aquoporina-4 foram negativos, excluindo doença desmielinizante do SNC ou neuromielite óptica. Estudo neurofisiológico de potenciais evocados somatossensitivos e visuais normal.
Efetuou ciclo de 5 dias com metilprednisolona 1 g/dia com melhoria parcial dos défices neurológicos e das queixas álgicas, sem recuperação adicional com esquema de reabilitação.
Foi avaliado por Reumatologia, verificando-se quadro prévio de aftose oral recorrente, sem atingimento genital, sendo avaliado alelo HLA- B51 que se revelou positivo. Foi então assumido o diagnóstico de doença de Behçet com envolvimento neurológico (mielite transversa) iniciando tratamento segundo o protocolo CYCLOPS que incluiu prednisolona 60 mg diárias durante 2 semanas, com posterior desmame e ciclos de ciclofosfamida segundo protocolo. Realizou o 3º pulso a 13/01/2020 sem intercorrências, encontrando-se atualmente a cumprir pulsos de ciclofosfamida oral em intervalos de 3 semanas.
Registou-se melhoria marcada do quadro álgico e recuperação de força muscular com menor necessidade de auxiliares de marcha. A reavaliação imagiológica será agendada de acordo com a evolução clínica.
Discussão: As manifestações neurológicas da doença de Behçet, apesar de incomuns e potencialmente tratáveis, constituem um dos quadros clínicos mais sérios associados a esta entidade patológica. O neuro-Behçet deve ser equacionado no diagnóstico diferencial de distúrbios infeciosos, inflamatórios ou desmielinizantes do sistema nervoso central. Episódios agudos de neuro-Behçet devem ser prontamente tratados com terapêutica imunomoduladora e glucocorticóides em alta dose seguidos de desmame lento.

CC288 – BUERGER´S DISEASE OR CANNABIS-INDUCED ARTERITIS?

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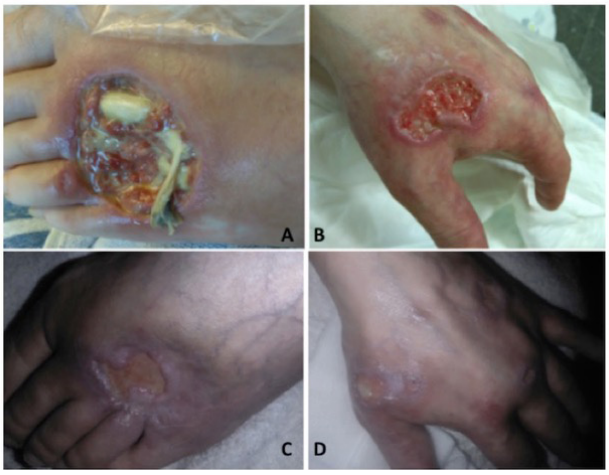
Introduction: Thromboangiitis obliterans (TAO), also called Buerger`s disease, is a nonatherosclerotic segmental inflammatory disease that affects the small and medium-sized arteries and veins in the upper and lower extremities. In general, affected patients are young male, and chronic tobacco smokers who present with distal extremity ischemia ulcers or gangrene of the fingers. The exclusion of a hypercoagulable state or an autoimmune disease is mandatory, essentially because anti-phospholipid syndrome shares clinical characteristics.

FIGURE 1 – SKIN LESIONS TO PRESENTATION (A,B); SKIN LESIONS AFTER REDUCE TOBACCO AND CANNABINOIDS CONSUMPTION



Clinical Case: A 34-year-old woman was referred to our Rheumatology department due to a one-year history of multiple ulcerative lesions and livedo reticularis in her limbs. She denied Raynaud phenomenon or other relevant complaints. She had consumptions of heroin, cocaine and cannabis in the past, and still keeps smoking tobacco and cannabis. She denied present consumption of injectable drugs. The patient had no other relevant antecedents. Previously, the patient was prescribed with methylprednisolone (16 mg/day) for a 6 months period for a suspected vasculitis, with very small improvement of lesions.
On the physical exam patient had ulcerative lesions over the hands and feet (these with tendon exposure) (figure 1: A,B), scars over right leg and livedo reticularis on limbs. She had a reduced dorsalis pedis arterial pulse and the remaining pulses were normal. The blood pressure was 126/66 mmHg, without significant changes in the four members. No other changes were found on the examination.
Laboratory findings revealed a complete normal blood count. C-reactive protein was increased, 4.98 mg/dL (normal <0,5 mg/dL) and normal sedimentation velocity 7 mm/h (normal range <20 mm/h); a chemistry panel including fasting blood sugar, liver enzymes, renal function, urinalysis, ionogram, lipid

profile, C3 and C4 tests were normal. Antinuclear antibodies, rheumatoid factor, myeloperoxidase anti-neutrophil cytoplasmic antibody, serine proteinase 3-anti-neutrophil cytoplasmic antibody and anti-phospholipid antibodies were all negative. Serologies for HVB, HVC and HIV were negatives. In addition, a complete hypercoagulability screen with coagulation tests, protein C, protein S, antithrombin III, factor V Leiden, and prothrombin gene were within normal values. On the toxicology panel, cocaine and amphetamines were negative but cannabinoids were positive in the urine.

The skin biopsy from the right foot lesion revealed acute thrombophlebitis. The computed tomography angiography showed absence of opacification in the distal parts of posterior tibial artery, dorsalis pedis artery in lower limbs and reduced vascularization of 2nd finger of left hand and 3th finger of right hand.

After smoking cessation counselling patient reduced tobacco and cannabis consumption, with very significant improvement of the lesions in two months (figure1: C,D).

Conclusion: This case highlights the importance of an extensive differential diagnosis in the presence of a non-linear clinical picture. However, the negativity of all laboratorial tests and a history of cannabis and tobacco consumption made us suspect of other causes. TAO is strongly associated with the use of tobacco products, and smoking cessation is essential to decrease the risk for amputation. Cannabis arteritis is clinically and pathologically indistinguishable from tobacco´s TAO but occurs less commonly.

CC296 – ORBITAL APEX SYNDROME MIMICKING GIANT CELL ARTERITIS: THE IMPORTANCE OF A CORRECT DIAGNOSIS

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Introduction: Giant cell arteritis (GCA) is a large- and medium-sized vessel vasculitis that most commonly affects older people. It typically manifests as constitutional features, headache, vision disturbances and elevated inflammatory markers. Around half the patients can present polymyalgia rheumatica (PMR). Due to its increased risk of blindness, GCA is considered a clinical emergency and thus treatment with high doses of glucocorticoids (GCs) should be initiated immediately. However, GCs can lead to significant toxicity, and other diseases may also present with similar manifestations to GCA. Therefore, a prompt and correct diagnosis in all patients with suspected GCA is essential for successful management.

Case report: We report a case of a 69-years-old woman who presented to the Emergency Department with a new onset of left hemicranial and retro-orbital headache, associated with diplopia and left eye amaurosis. Over the previous two months she had been complaining of weight loss (around 8% of total body weight) and PMR features. More recently she reported pain below the right shoulder blade. She denied jaw claudication. Her background history consisted of diabetes mellitus type 2 and there was no history of smoking or alcoholic habits. On physical examination cardiopulmonary auscultation was unremarkable and no abnormalities of the temporal arteries were found. Ophthalmic examination of the left eye revealed lack of light perception, relative afferent pupillary defect, ptosis, horizontal recti muscles palsy and hypoesthesia in ophthalmic division of the trigeminal nerve. On fundoscopy, diabetic macular oedema was observed and optic disc showed no signs of ischaemia. Erythrocyte sediment rate was 50 mm/hr and C-reactive protein 5.9 mg/dL. On initial imaging study, the thorax radiograph revealed a consolidation in the right inferior lobe and cranial computed tomography (CT) showed ischemic microangiopathic leukoencephalopathy. Cerebrospinal fluid analysis was normal. Due to the high suspicion of GCA at disease presentation, methylprednisolone pulses of 1g per day were initiated. On the 3rd day of treatment, the patient was observed by Rheumatology and underwent an ultrasound of the temporal, facial, axillary and subclavian arteries showing no signs of vasculitis and an ultrasound of the shoulders and hips without features suggestive of PMR. On further investigation, a thoracic CT was performed revealing a lung mass with multiple lung and adrenal nodules. Histology confirmed lung adenocarcinoma. In addition, the patient underwent cranial MRI showing fat obliteration of the left orbital apex with isointense tissue enhancement after gadolinium suggestive of orbital apex syndrome, a rare condition characterized by optic neuropathy and ophthalmoplegia due to the involvement of structures within or near the orbital apex. A dural and a parenchymal lesion were also detected consistent with metastases. Therefore,

metastatic lung cancer was assumed as the cause of orbital apex syndrome. The patient stopped GCs with a fast tapering scheme. However, two months after diagnosis she died due to an acute cholecystitis complicated by renal failure.
Conclusion: Other diseases may mimic GCA. In this case, the patient presented an orbital apex syndrome secondary to lung adenocarcinoma manifesting with headache, visual disturbances, PMR and high inflammatory markers. A fast track approach with immediate ultrasound at disease presentation could have prevented unnecessary treatment and help guiding clinical investigation without delay.

CC298 – ACIDENTE VASCULAR CEREBRAL – UMA APRESENTAÇÃO ATÍPICA DE ARTRITE DE CÉLULAS GIGANTES

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Introdução: A Artrite de Células Gigantes (ACG) é uma vasculite granulomatosa que atinge as artérias de médio e grande calibre. Surge habitualmente após os 50 anos, sendo mais comum nas mulheres e caracteriza-se classicamente por cefaleia persistente, claudicação mandibular e alterações visuais. Cerca de 50% dos casos de ACG podem ainda apresentar-se com clínica típica de Polimialgia Reumática (PMR), encontrando-se estas duas patologias intimamente relacionadas. A elevação marcada da velocidade de sedimentação (VS) associada a uma clínica típica, deve fazer suspeitar o seu diagnóstico. A ocorrência de AVC é uma complicação rara, com uma prevalência de 2-7%. Os fatores de risco para AVC mais descritos são o sexo masculino, idade avançada, Diabetes Mellitus e neuropatia óptica isquémica anterior. O tratamento do AVC nestes casos assenta na corticoterapia, podendo ser útil a combinação com antiagregantes plaquetares.
Caso clínico: Homem, 76 anos, com cefaleia holocraniana, ataxia e tonturas desde há 2 meses, com posterior surgimento de dor temporal bilateral, miodesópsias e fotópsias no olho direito, assim como noção de perda ponderal e fadiga generalizada. Antecedentes pessoais de hipertensão arterial e tabagismo, medicado com irbesartan. Exame objetivo com discreto desvio em pronação do membro superior direito, dismetria na prova calcanhar-joelho à direita e marcha atáxica com desequilíbrio sustentado para a direita. Analiticamente: anemia normocítica e normocrómica ligeira (Hb 13.7g/dL), VS 51mm/h e PCR 4.62mg/dL. Angio-RM CE com lesões isquémicas agudas a nível do lobo occipital esquerdo e hemisférios cerebelosos; Ecodoppler das artérias temporais com halo hiperecóico dos ramos frontais, parietais e tronco comum bilateralmente; e biópsia das artérias temporais compatível com ACG. Assim, foi confirmada a suspeita diagnóstica de ACG com AVC posterior. Iniciou terapêutica com pulsos de metilprednisolona e posteriormente prednisolona 1mg/kg/dia, apresentando diminuição dos parâmetros inflamatórios e recuperação gradual dos défices neurológicos.
Discussão: A paucidade de sintomas constitucionais e escassa clinica típica, torna o diagnóstico de ACG desafiante. Apesar de constituir uma complicação rara da ACG, a presença de AVC está associada a maior mortalidade e maior recidiva desta entidade. A distinção clínica e imagiológica entre os AVC de etiologia vasculítica e aterosclerótica é difícil, no entanto o envolvimento das artérias vertebro-basilares é mais comum na ACG (40-60%), pelo que deve ser considerada no diagnóstico diferencial em AVCs com esta localização.
Conclusão: A ACG tem uma apresentação clássica, contudo existem casos atípicos, como os AVC, que podem gerar atraso diagnóstico e início da terapêutica. A ACG deve ser ponderada em doentes com AVC, sobretudo acima dos 50 anos, na presença de VS aumentada, clínica típica e atingimento vertebrobasilar.

CC300 – BIFID MEDIAN NERVE AND CARPAL TUNNEL SYNDROME – A RARE ASSOCIATION DEMONSTRATING THE IMPORTANCE OF ULTRASOUND EXAM

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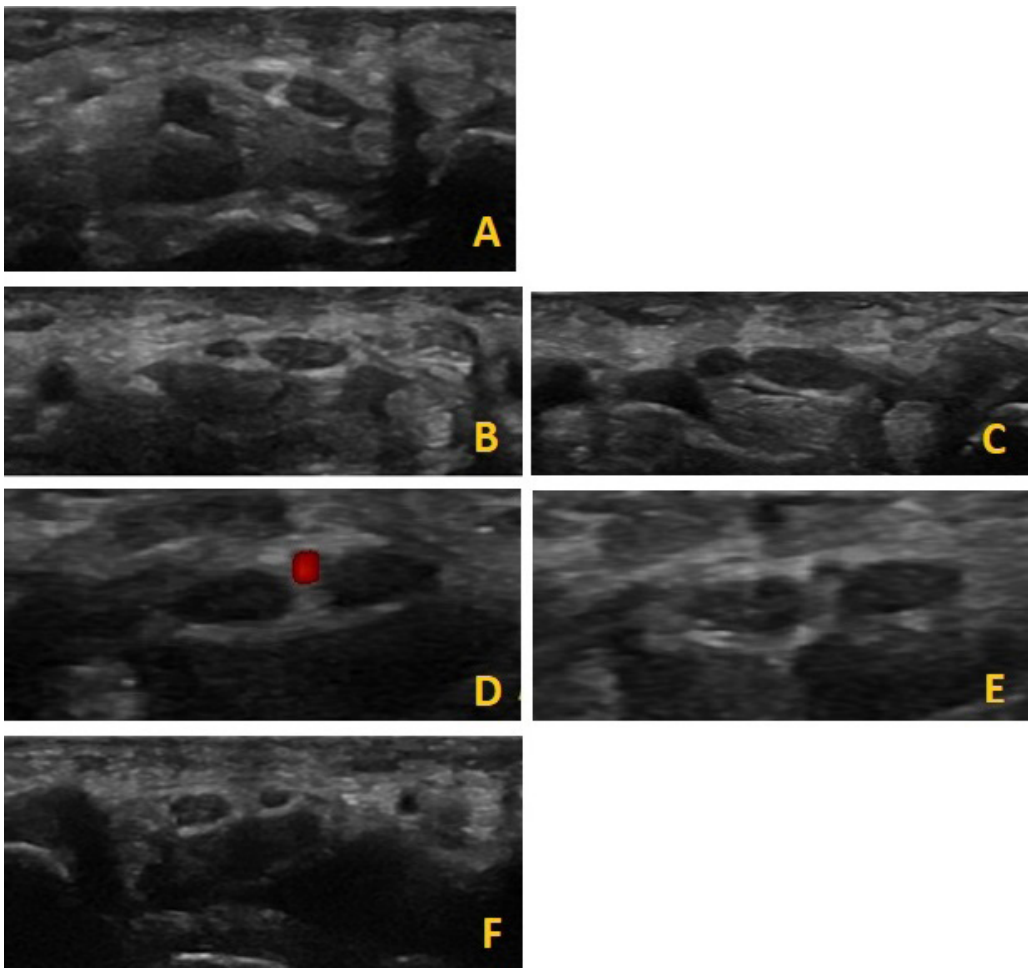
Introduction: Carpal tunnel syndrome (CTS) is a neuropathy caused by entrapment of the median nerve in the wrist. In some patients, it can be secondary to nerve median variations (like a bifid median nerve), to vascular anomalies (usually a persistent median artery) or both. Bifid median nerve is a rare anatomic variation characterized by its division proximally to the carpal tunnel and it occurs in 0,8% to 2,8% of patients with CTS. It may be associated with persistent median vessels (such as a persistent median artery) or with aberrant muscles. This variation may facilitate compression of the median nerve in the

FIGURE 1 – ULTRASONOGRAPHY IMAGES OF BIFID MEDIAN NERVES, FROM THE PATIENTS IN THE CLINICAL CASES



carpal tunnel because of its increased cross-sectional area, conducting to a CTS.

Clinical cases:

Clinical case 1: Female, 54 years old, housekeeper with a history of right rhizarthrosis and complaints of pain and paraesthesia in the median nerve distribution in both hands, that worsened at night. At examination, she had bilateral positive Phalen's test and in ultrasound, a bifurcation of the left median nerve (Fig 1A). The bifurcation happened right before the entrance in the carpal tunnel. The nerves overall cross-s-sectional area in the carpal tunnel of 10 mm2 the right one and of 13 mm2 the left one.

Clinical case 2: Female, 54 years old, cooker with a history of knee osteoarthrosis. She presented paraesthesia complaints in the median nerve distribution in both hands, which also worsened at night. Objectively she had positive Tinel's and Phalen's tests. The ultrasound revealed both right and left bifid median nerves. On the right hand, total cross-sectional area in the carpal tunnel of 10 mm2 (Fig 1B); left median nerve overall width in the carpal tunnel of 17 mm2 (Fig 1C).

Clinical case 3: Female, 63 years old, unemployed. Observed in Rheumatology consultation because of paraesthesia, pain, and numbness in the left hand that worsened at night and a recent complaint of hand muscle weakness. The Phalen's test was positive bilaterally and the ultrasonography showed a bifid left median nerve, with a cross-sectional area of 10 mm2 and with a median artery between both portions, that can be seen with power Doppler (Fig 1D e 1CE). At the right hand, the median nerve's anatomy was regular, with a cross-sectional area in the carpal tunnel of 9 mm2.

Clinical case 4: Female, 35 years old, with complaints of pain and paraesthesia in the median nerve distribution in the right hand, which worsened at night. Frequently she dropped objects off her hands due to muscle weakness. Physical examination did not present significant alterations. The ultrasonography revealed right bifid median nerve with an increased

overall cross-sectional area (Fig 1F) and left median nerve cross-sectional area of 9 mm2.
Conclusions: CTS is a frequent condition, mostly in middle-aged women, and bifid median nerve, as demonstrated in these 4 cases, is a potential cause of CTS, and must be considered, especially when the patient has unilateral symptoms. The ultrasonography examination, a wide method used by rheumatologists, with a sensitivity and specificity comparable to those of electrophysiological examinations, helps to identify median nerve anatomy variations and associated vascular or muscular anomalies, therefore minimizing lesion of these structures in surgery or by infiltrations.

CC303 – SÍNDROME HEMOFAGOCÍTICO EM DOENTE COM ARTRITE REUMATÓIDE: PERSEVERAR NAS ETIOLOGIAS MAIS PROVÁVEIS

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A artrite reumatóide (AR) predispõe ao desenvolvimento de doenças linfoproliferativas (DLP), sendo o risco maior em doentes sob terapêutica imunossupressora, como o metotrexato.
A Doença de Kikuchi-Fujimoto (DKF) é uma linfadenite necrotizante, tipicamente cervical, associada a febre persistente. A histologia ganglionar tende a revelar infiltrado histiocítico ou, em fases avançadas, necrose central. Tem um curso benigno e autolimitado. Contudo, estão descritos casos graves, raros, associados a síndrome hemofagocítico (SH), que é um estado de inflamação sistémica excessiva, resultante de uma desregulação imunológica. O SH cursa com febre persistente, citopenias, hiperferritinemia, hipertrigliceridemia e esplenomegalia. Pode ser idiopático ou secundário a infeção viral, DLP ou doenças autoimunes. Não tratado leva a disfunção multiorgânica, não raras vezes culminando em morte.
Caso Clínico: homem de 43 anos, com o diagnóstico de AR há 6 anos, fator reumatóide e anti-CCP positivos, com apresentação inicial de poliartrite assimétrica, e bem controlado com metotrexato 25mg/semanal. Iniciou queixas de febre, mialgias, sudorese noturna e astenia, associados a adenopatia cervical direita volumosa. Fez tratamento com anti-inflamatório, tendo o quadro regredido passados 10 dias. Episódios semelhantes ocorreram mensalmente desde esta altura, ao longo de 8 meses. Do estudo realizado neste período, salienta-se a TC de corpo, que revelou volumosas adenopatias cervicais à direita, a maior com 44x42mm, e adenopatias infracentimétricas em vários territórios do tórax e abdómen. A histologia do gânglio cervical, após punção aspirativa, revelou linfadenite necrotizante, sem presença de células neoplásicas ou histiócitos. As serologias virais foram negativas, os marcadores tumorais e a eletroforese de proteínas estavam normais e foi excluída tuberculose. Assumiu-se como causa etiológica a DKF, tendo o doente iniciado tratamento com prednisolona oral 0,5mg/kg/dia durante dois meses, com melhoria parcial das queixas, até à ocorrência de um novo episódio, com marcada deterioração do estado geral. Foi internado e fez-se biópsia excisional, que numa pré-análise revelou características similares de necrose. Enviou-se a amostra para caracterização em centro de referência. O estado clínico e laboratorial agravou nos primeiros dias, com febre persistente (>39ºC), perda ponderal, PCR 26,6 mg/dL, VS 80 mm/1h, Hb 6,9 g/dL, ferritina 26765ng/mL e triglicerídeos 400 mg/dL. Não houve resposta à antibioterapia de largo espectro. O medulograma caracterizou uma medula reativa, não sugerindo um diagnóstico específico. Por suspeita de SH, cumpriu três pulsos de metilprednisolona 1g IV e imunoglobulina humana 2g/kg IV, repartidos por 5 dias, seguidos de prednisolona oral 1mg/kg/dia. O estado clínico e laboratorial melhorou prontamente. A reanálise da amostra, disponível após esta ocorrência, revelou linfoma de Hodgkin clássico, de celularidade mista. O doente foi encaminhado para consulta especializada e iniciou tratamento dirigido.
Conclusão: descreve-se o caso de um SH que ocorreu em contexto de DLP num doente com AR. A gestão do caso foi dificultada pela inespecificidade do resultado de dois exames histológicos de uma adenopatia, que conduziu ao diagnóstico de exclusão de DKF. Compete ao clínico perseverar e excluir as causas que melhor podem justificar um quadro e onde a gestão atempada pode ditar o sucesso terapêutico, ou um desfecho trágico.

CC308 – DOENÇA DE BEHÇET COM ENVOLVIMENTO RENAL: UM DESAFIO TERAPÊUTICO

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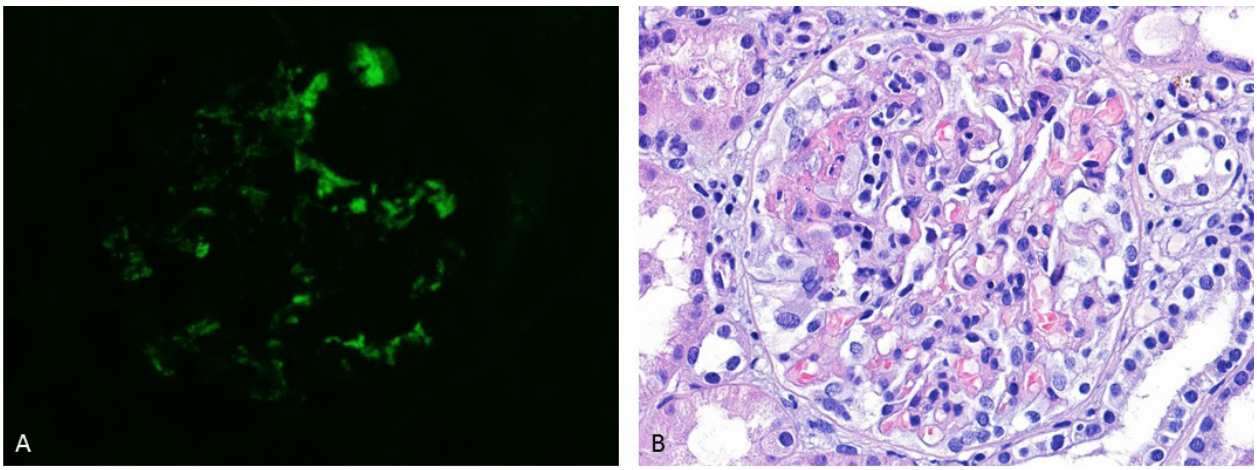
FIGURA 1 – HISTOLOGIA DA BIÓPSIA RENAL



A) Depósitos imunes mesangiais granulares [IgA, 200x]; B) Glomérulo com hipercelularidade mesangial e endocapilar e lesões de necrose fibrinóide [HE, 400x].

Introdução: A Doença de Behçet (DB) é uma vasculite multissistémica rara que constitui um desafio diagnóstico e terapêutico, particularmente em casos de envolvimento orgânico menos comum.

Caso Clínico: Mulher de 30 anos, com DB diagnosticada em 2010, com envolvimento mucocutâneo (úlceras orais recorrentes, teste da patergia positivo), neurológico (nevrite óptica, lesões da substância branca) e gastrointestinal (procto-colite), corticodependente e refratária, do ponto de vista intestinal, a múltiplas terapêuticas (azatioprina, metotrexato, infliximab, vedozilumab, adalimumab), com necessidade de colectomia segmentar (sigmóide e descendente distal) e ostomia em janeiro de 2019. Encontrava-se sob ustecinumab 90mg subcutâneo de 8/8 semanas (desde dezembro de 2018) e prednisolona 7.5mg/dia, mantendo seguimento em consultas de Reumatologia, Neurologia e Gastrenterologia. Em abril de 2019 referiu, de novo, hematúria macroscópica ligeira, assintomática. A angio-tomografia abdominopélvica não mostrou alterações no aparelho excretor e a citologia urinária foi negativa para células neoplásicas. O estudo analítico de maio de 2019 revelou, de novo, leucocitose (16.42x10^9/L) com neutrofilia (12.74x10^9/L), elevação de proteína C-reativa (84.3mg/L, normal<3.0), da velocidade de sedimentação (67mm/h) e da creatinina (1.58mg/dL, normal<0.95) com ureia normal (44mg/dL, normal<50), com o sedimento urinário a revelar proteinúria (0.3g/L) com eritrocitúria (52.1/uL). Repetiu o estudo uma semana depois, que mostrou: creatinina 1.5 mg/dL, urina ocasional com proteinúria de 0.3 g/L, eritrocitúria de 20-50 eritrócitos/campo e presença de numerosas sombras eritrocitárias e eritrócitos dismórficos. A doente referia astenia mais marcada, sem outras queixas. Foi internada em Reumatologia e proposta para biópsia renal. O estudo imunológico foi negativo (ANCAs, ANAs, anticorpos anti-ENA, anti-nucleossomas e anti-cardiolipinas negativos; ausência de imunocomplexos circulantes, crioglobulinas e consumo de fatores de complemento; inibidor lúpico e prova de Coombs direta negativos, imunoglobulinas normais) e a histologia renal mostrou quadro compatível com vasculite renal em atividade no contexto de uma nefropatia por IgA e nefrite tubulointersticial crónica (figura 1). Optou-se pela realização de pulsos de metilprednisolona 1g endovenosa (e.v.)/dia durante 3 dias e posteriormente ciclofosfamida e.v. mensal. A doente recusou as hipóteses de proteção/preservação gonadal e iniciou ciclofosfamida 1g e.v. mensal associada a esquema de prednisolona oral em desmame, vitamina D e cotrimoxazol. Ao quinto mês de ciclofosfamida, a colonoscopia revelou colite crónica com sinais de atividade, com positividade para citomegalovírus (PCR), tendo cumprido ciclo de valaciclovir. Após 6 meses de tratamento com ciclofosfamida, inciu azatioprina em dose crescente, estando atualmente sob azatioprina 100mg/dia e prednisolona 15mg/dia, revelando excelente resposta do ponto de vista renal, com creatinina 0.83 mg/dL e sedimento urinário sem hematoproteinúria.

Conclusão: O envolvimento renal na DB é pouco frequente e normalmente pouco grave. A apresentação inicial consiste mais frequentemente em hematúria e/ou proteinúria assintomáticas. O espectro do atingimento renal é variável, incluindo amiloidose, glomerulonefrite, nefrite intersticial, nefropatia por IgA ou vasculite renal. Apresentamos um caso grave de DB que apresentou hematúria macroscópica no contexto de vasculite renal, nefropatia por IgA e nefrite tubulointersticial crónica, com excelente resposta à ciclofosfamida.

CC312 – A RARE NON-INFECTIOUS CAUSE OF FEBRILE POLYARTHRITIS WITH GENERALIZED PALMOPLANTAR RASH

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Introduction: Schnitzler's syndrome is a rare autoinflammatory disease acquired during adulthood, clinically characterized by chronic urticaria, arthralgias or bone pain, adenomegaly and intermittent fever.
Clinical presentation occurs on average around 55 years of age, and it's always associated with IgM, or less frequently IgG, monoclonal gammopathy. The main complication is the development of lymphoproliferative disease in up to 45% of cases. Skin biopsy is an important, but not essential complementary exam for diagnosis.
Clinical case: A 61-year-old man complained of a 4 months history of generalized urticariform evanescent migratory rash, with episodes lasting around 7 days, involving the neck, trunk and limbs, affecting skin folds, palms and soles. He also presented an intermittent fever mainly at night-time with 40º C maximum temperature.
On the 3rd month the patient developed migratory polyarthralgia affecting small and large joints, without inflammatory signs, that were coincident with worsening of the cutaneous lesions.
Blood tests revealed leucocytosis (13,9 x 10 9/L), neutrophilia (11,2 x 10 9/L), raised CRP 27 mg/L and ESR 43 mm/hr, ferritin 337 ng/mL and were negative for ANAs, ANCAs, anti-dsDNA and rheumatoid factor. The search for infectious (serologies to cytomegalovirus, mycoplasma, coxsackie, coxiella burnetti, parvovirus, Epstein-Barr, hepatitis B and C virus, HIV, syphilis) and neoplastic (thoraco-abdomino-pelvic CT) causes was negative.
Oral deflazacort 60 mg daily was initiated with good clinical response but a relapse occurred with dosage reduction below 30 mg.
Electrophoretic proteinogram posteriorly revealed hypergammaglobulinemia with a peak on the gamma curve, an increase of free light lambda chains, and serum immunofixation revealed monoclonal IgG gammopathy.
A diagnosis of Schnitzler's syndrome was made, according to the Strasbourg diagnostic criteria.
Discussion: Schnitzler's syndrome is currently considered the paragon of the late onset acquired auto-inflammatory syndrome, deserving exclusion of other similar entities (adult Still's disease, urticariform vasculitis, cryopyrinopathies, cryoglobulinemia, acquired C1 inhibitor deficiency, hyper IgD syndrome). The palmoplantar involvement of the skin, often present in Schnitzler's syndrome, warrants exclusion of diseases such as syphilis, meningococcal infection, DRESS syndrome, SAPHO syndrome, acute cutaneous lupus erythematosus, among others.
Monoclonal IgG gammopathy may be more frequent than previously thought, being an important diagnosis as it has prognostic value, conferring risk of lymphoproliferative disease similar to that of the IgM subtype.

CC314 – TUBERCULOSE COM ENVOLVIMENTO PULMONAR E INTESTINAL EM DOENTE COM ARTRITE PSORIÁTICA SOB GOLIMUMAB

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Introdução: A artrite psoriática pertence à família das espondilartrites, afetando até 30% dos doentes com psoríase, sem atingimento preferencial por género.
Em caso de ineficácia de fármaco modificador do curso da doença (DMARD) convencional, pondera-se introdução de um DMARD biológico, estando a profilaxia de reativação de tuberculose latente sempre preconizada, entre outros rastreios serológicos e imunizações.
Caso clínico: Homem de 70 anos com diagnóstico de artrite psoriática forma poliarticular simétrica de predomínio periférico no 6º ano de terapia com golimumab 50 mg em intervalos de 4 semanas e 4ª ano de metotrexato 10 mg foi submetido a hernioplastia inguinal direita, com resultado histológico da peça operatória a revelar granulomas sugestivos de sarcoidose com implantação peritoneal.
Iniciou tratamento com prednisolona 60 mg diárias com desmame gradual até 10 mg ao longo de 7 meses, altura em iniciou quadro de declínio do estado geral, astenia, dor abdominal generalizada e febre. Recorreu ao serviço de urgência objetivando-se leucocitose (19 x 10^3 / ul) com neutrofilia (5,3 x 10^3) e elevação da PCR (91,94 mg/l), IGRA positivo, exame cultural positivo para Mycobacterium tuberculosis e TC-Tórax com padrão de nodularidade múltipla sugestivo de tuberculose miliar e achados em "vidro despolido". Suspendeu golimumab e metotrexato e iniciou terapia combinada de etambutol 1200 mg diárias, pirazinamida 1500 mg diárias, isoniazida 300 mg diárias e

piridoxina 40 mg 2/ 2 dias, com boa resposta clínica ao 10º dia de terapêutica.
No decorrer do internamento verificou-se quadro de sub-oclusão intestinal, observando-se áreas de espessamento e realce no íleo terminal em TC abdominal, havendo resolução do quadro com terapêutica conservadora.
Seis meses após alta recorre ao serviço de urgência por dor abdominal no hipogastro e fossa ilíaca esquerda de instalação súbita com vómitos alimentares associados. Observou-se em TC- abdominopélvica uma densificação edematosa difusa da gordura abdominal com plastron de ansas de intestino delgado, achados sugestivos de tuberculose com envolvimento intestinal. Por falência do tratamento conservador procedeu-se a laparotomia exploradora, onde se constataram aderências generalizadas, realizando-se ileocolectomia com anastomose.
Estudo histopatológico do líquido ascítico revelou processo inflamatório crónico transmural associado a granuloma com área de necrose, assumindo-se o diagnóstico de tuberculose com envolvimento pulmonar e intestinal em doente sob imunossupressão de longa data. Foi mantido o tratamento com tuberculostáticos e a vigilância de eventuais sintomas.
À reavaliação em consulta apresentava agravamento clínico da artrite psoriática, ponderando-se reintrodução de metotrexato via subcutânea ao 6º mês de terapêutica anti-bacilar.
Discussão: Foram registados casos de reativação de tuberculose em doentes sob Golimumab, independentemente da realização de terapêutica profilática prévia. O esquema adequado de reintrodução de terapia biotecnológica em doentes com tuberculose ativa com flare da doença reumática de base ainda não é consensual, estando também dependente do seu grau de atividade.
As recomendações da SPR sugerem que o tratamento com bDMARD poderá ser iniciado de forma segura 2 meses após o início do tratamento antibacilar efetivo. A vigilância apertada destes doentes em colaboração com o CDP é fundamental para que o tratamento em causa seja feito de forma segura.

CC319 – ENTESOPATIA DO DELTÓIDE NAS ESPONDILARTRITES: UMA CAUSA POUCO CONHECIDA DE OMALGIA

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Introdução: A Entesite é uma característica patológica típica das espondilartrites, podendo assumir aspetos variáveis e ocorrer em diferentes localizações. Nestas patologias, as enteses dos membros inferiores são as mais frequentemente envolvidas, e por isso, amplamente mais estudadas e documentadas1. A entesopatia dos membros superiores ainda permanece ignorada e subdiagnosticada.
O envolvimento do ombro nas espondilartrites é muito frequente, sendo na generalidade dos casos interpretado como patologia da coifa ou articular. A ecografia é o exame de eleição para o diagnóstico diferencial dos diferentes tipos de envolvimento.
Objetivo: Apresentação de três casos de espondilartrites que cursaram com omalgia por entesopatia do deltóide.
Casos clínicos:
Caso 1: Mulher de 51 anos, enviada para a consulta de reumatologia por suspeita de polimialgia reumática com quadro de dor, rigidez e limitação funcional dos ombros e ancas. No entanto, a avaliação ecográfica inicial foi normal. Posteriormente, por persistência das queixas na região posterior dos ombros, foi realizada nova ecografia que revelou edema, alteração da estrutura fibrilar e doppler na inserção do deltóide. Na mesma altura, iniciou queixas de fasceíte plantar bilateral e apresentou elevação dos parâmetros inflamatórios nas análises realizadas. O quadro clínico foi interpretado como espondilartrite com envolvimento entesopático (entesite do deltóide, do médio glúteo e fasceíte plantar).
Caso 2: Homem de 77 anos, seguido em consulta de Reumatologia por espondilite anquilosante, com tipagem HLA-B27 positiva, envolvimento articular axial, periférico e entesopático (fasceíte plantar). Após 20 anos de seguimento, iniciou omalgia bilateral persistente sem melhoria com anti-inflamatórios não esteróides, infiltração da bolsa subacromiodeltoideia e tratamento de reabilitação. Pela refratariedade à terapêutica instituída, foi realizada avaliação ecográfica que mostrou entesopatia do deltóide. Realizado tratamento local com corticosteróide com melhoria.
Caso 3: Homem de 61 anos, observado em consulta de Reumatologia por omalgia bilateral com 7 meses de evolução. Este quadro foi interpretado como patologia da coifa, tendo sido submetido a infiltração da bolsa subacromiodeltoideia, sem melhoria. Por manutenção da omalgia, foi realizada avaliação ecográfica do ombro que demonstrou entesite na inserção do deltóide no acrómio. Foi realizado novo tratamento local com corticosteróide, mas agora na entese. Desde então, sem queixas.

Discussão/conclusão: Nas Espondilartrites, a entese do deltóide, nomeadamente na sua inserção fibrocartilagínea lateral e superior do acrómio2 pode ser alvo de inflamação, passando frequentemente despercebida e subdiagnosticada3.
Dada a frequência de omalgia nestes doentes, é importante sublinhar a entesopatia do deltóide como manifestação integrante do quadro clínico, podendo auxiliar no seu diagnóstico e consequente tratamento. A ecografia é um exame excelente na avaliação desta entese.

CC329 – REMMITING SERONEGATIVE SYMMETRICAL SYNOVITIS WITH PITTING EDEMA SECONDARY TO HYPEREOSINOPHILIC SYNDROME

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Introduction: Remitting seronegative symmetrical synovitis with pitting edema syndrome (RS3PE) is characterized by symmetrical synovitis and hand and feet edema with a positive Godet sign. Most cases are idiopathic, however, on rare occasions, RS3PE might be associated with autoimmune, neoplastic or neurodegenerative disease. There is a known association of RS3PE with hematologic disease, more frequently with leukemia, although other, rarer, hematologic diagnosis can be at the base of RS3PE. Idiopathic hypereosinophilic syndrome (HES) is a rare entity that, even more infrequently, presents with distal edema and symmetrical polyarthritis and more often involves the heart, lungs and skin. A considerable number of patients are diagnosed with a hematologic malignancy in the months following the diagnosis.
Case report: 65 year old, male patient, with a personal history of HES since 2014, medicated with 20 mg of prednisolone, hypertension and complicated peripheral artery disease. The patient was referred to a Rheumatologist due the symptoms of inflammatory polyarthralgia with 6 months of evolution. The physical exam revealed symmetrical polyarthritis, involving the proximal interphalangeal articulations, wrists, knee and a pitting edema of both hands and feet. Blood analysis revealed marked eosinophilia (2600x10^9/L), erythrocyte sedimentation rate of 120 mm/s, C-reactive protein of 2.9 mg/dL, uric acid of 6.1 mg/dL, negative rheumatoid factor, anticyclic citrullinated peptide and antinuclear antibody. The ultrasound exam of the hand and wrist showed extensive synovitis and tenosynovitis of the wrists and edema. The patient was started on a daily dose of 40 mg of prednisolone, with resolution of all the articular symptoms. On the following consultation, the prednisolone dosage was adjusted to 30 mg/day, and the patient was started on 10 mg/week of methotrexate. One week later the patient was admitted in the emergency department with complaints of hand polyarthritis, edema of the hands and feet, fever and exanthematous rash of both arms, forearms, face and dorsal area.
The patient was medicated with intramuscular betamethasone and was admitted for diagnostic investigation. The rash was unchanged, and blood analysis revealed an exacerbation of the eosinophilia, but there was a regression of the polyarthritis and edema. All newly started drugs were suspended, including methotrexate, and the corticosteroid dosage was adjusted to 20 mg/day. On the days following the corticosteroid dosage reduction both the polyarthritis and low-grade fever resurged.
The patient underwent a thoracic CT scan that showed the presence of multiple axillary lymph nodes, the largest with a long axis of 33 mm, which was excised. Due to the persistence of the symptoms a PET scan was ordered and the result showed a cutaneous and periarticular infiltrative pattern and associated reactive nodular hypermetabolism. The skin biopsy put into evidence the presence of hyaline diffuse infiltrate suggestive of eosinophilic involvement. The patient was thus diagnosed with RS3PE secondary to HES, and was referred back to the hematology clinic, and the corticosteroid dosage was increased. The patient was diagnosed with a T-cell Lymphoma 6 months later.
Discussion: HES is a rare entity that does not spare any organ or system and might mimic primary rheumatologic conditions. RS3PE notably responds very well to corticotherapy, and its association with HES has been documented in the past.

CC330 – COXIELLA BURNETTI INFECTION – A RARE CAUSE OF POLYARTHRITIS

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Introduction: Most cases of oligoarthritis or polyarthritis caused by infectious agents are the result of reactive arthritis caused by Chlamydiae Spp. enterobacteriaceae, or, in the acute phase due to microorganisms associated with septic arthritis, such as Staphyloccocus aureus. However other bacteria, such as Coxiella Burnetti, can cause serious articular complications, which are often overlooked.

Acute infection caused by Coxiella Spp. can be associated with arthritis and, in some cases, with high titers of ANA.

Case Report: The authors report the case of a 51 year old, male patient, caucasian, farm worker, with a personal history of reflux esophagitis, active smoker. Referred to a Rheumatology clinic for the first time in 2014 due to migratory oligoarthralgia involving hands and wrists. At the initial physical observation the patient showed oligoarthritis with involvement of the first left metacarpophalangeal and left tibiotarsal. The blood analysis revealed a negative rheumatoid factor, negative CCP, ANA 1/160, negative anti-ds DNA, with no other significant alterations. The diagnosis of microcrystalline arthropathy was assumed and the patient was medicated with allopurinol and colchicine, with resolution of all the symptoms within 1 week. The patient remained asymptomatic until September 2019, at which time the patient recourse to an emergency consultation due to polyarthritis involving all metacarpophalangeal articulations, wrists, knees and feet. The patient suffered from a maculopapular rash, with punctiform areas, that spared only the legs, feet, palms and soles.

At this time the patient started complaining of dry cough and worsening of gastroesophageal reflux symptoms. Blood analysis was repeated and revealed ANA titer of 1/1280, equivocal anti-dsDNA level and ESR of 78mm/s. The patient underwent a chest radiography that showed a bilateral, diffuse interstitial infiltrate. Due to persistence of symptoms, in November of 2019, an extensive blood analysis with viral and bacterial serology was ordered. In this evaluation it was detected a positive Coxiella burnetii IgM (phase II). Clinically, the maculopapular rash now extended to the both legs. Empirical antibiotic therapy with doxycycline was initiated and the Cocxiella burnetti IgM positivity was confirmed. One week after starting doxycycline, the patient was evaluated and there was no evidence of arthritis. The patient completed 4 weeks of antibiotic.

Discussion: Although rare, Coxiella burnettii can be associated with polyarthritis or oligoarthritis. The symptoms tend to resolve with antibiotic therapy for at least 4 weeks. Sometime a second course of intravenous antibiotic may be necessary for symptoms improvement. Multiple cases of acute infection by Coxiella burnetti associated with high titers of ANA, ENA and anti-CCP were reported in the literature.

CC334 – ARTERITE DE GRANDES VASOS: UMA FORMA DE APRESENTAÇÃO RARA

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Introdução: A arterite de Takayasu é uma vasculite de grandes vasos que afeta primariamente a aorta e os seus ramos. Numa fase inicial, o doente pode apresentar sintomas de fadiga, perda ponderal e febrícula. A determinação e monitorização da atividade da doença são desafiantes, sendo que a utilização da velocidade de sedimentação (VS) como marcador, apresenta uma sensibilidade de 72% e especificidade de 56%.

Caso clínico: Mulher de 56 anos, com antecedentes de tabagismo ativo e hipertensão arterial, inicia em fevereiro de 2018 quadro de dispneia de início insidioso e agravamento progressivo, com desconforto torácico associado. Um ano depois recorre ao Serviço de Urgência, por precordialgia de novo, acompanhada de dor abdominal, e é internada por síndrome coronário agudo, por apresentar elevação do segmento ST em eletrocardiograma, contudo, analiticamente apresentava troponina T 21 ng/L, proteína C reativa 0,80 mg/dL e VS 42 mm/h. Em TC toraco-abdomino-pélvica verificou-se disseção segmentar 2,5 cm da aorta abdominal infra-renal e derrame pleural bilateralmente, sendo que em angio-TC descreveu-se presença de eventual trombo intramural concêntrico das porções ascendente, crossa e infra-renal da aorta. Realizou TC das artérias coronárias, que revelou um score de cálcio coronário igual a 0, excluindo-se assim doença arterial aterosclerótica; ecocardiograma transtorácico: dilatação do ventrículo esquerdo, hipocinesia difusa e disfunção sistólica bi-ventricular com fração de ejeção de 22%; alterações confirmadas posteriormente em ecocardiograma transesofágico. A ressonância magnética cardíaca evidenciou dilatação do ventrículo esquerdo, porém sem edema do miocárdio, com alterações segmentares tradutoras de enfarte não transmural com viabilidade. Por suspeita de processo vasculítico da aorta e dos seus ramos realizou tomografia por emissão de positrões, onde se confirmou suspeita de aortite, predominante dos ramos ascendente e de pequena porção infra-renal, com doença menos exuberante entre estes dois

troços. Durante toda a investigação diagnóstica, a doente não apresentou novo aumento dos parâmetros inflamatórios, ou alterações da contagem leucocitária. Por se admitir processo de arterite de grandes vasos, e após discussão multidisciplinar, foi medicada com metilprednisolona (1000mg/dia) seguidos de prednisolona na dose de 1mg/kg/dia, e dada a extensão e a gravidade do envolvimento vascular, realizou ciclofosfamida, de acordo com protocolo CYCLOPS (início em maio de 2019 e término em agosto de 2019). Em setembro integrou programa de reabilitação cardíaca e em ecocardiograma de controlo do mesmo mês apresentava apenas uma ligeira diminuição da fração de ejeção (48%), com hipocinesia limitada às paredes inferior e infero-lateral.

Conclusão: Este caso clínico põe em evidência que os parâmetros inflamatórios nem sempre são suficientes para o diagnóstico e monitorização destas patologias e destaca-se o papel dos exames de imagem neste contexto. A ciclofosfamida, a que se associa uma cardiotoxicidade entre 7-28%, foi essencial neste caso, para o controlo do processo inflamatório tendo revelado grande eficácia na reversão da disfunção sistólica inicialmente verificada.

CC335 – REFRACTORY THROMBOCYTOPENIA IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND SECONDARY ANTIPHOSPHOLIPID SYNDROME

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Background: Thrombocytopenia is a frequent hematological manifestation in patients with systemic lupus erythematosus (SLE). Its prevalence has beenestimated to range from 10% to 40%, but severe thrombocytopenia is relatively uncommon. Strokes and transient ischaemic attacks are considered the second most common clinical manifestations of antiphospholipid syndrome after venous thrombosis. There are no clear data on how to manage these events in patients with immune thrombocytopenia.

Case report: A 44-year-old woman diagnosed with SLE and secondary antiphospholipid antibody syndrome was admitted in our hospital, in July 2018, presenting severe renal and haematological involvement. She underwent treatment with methylprednisolone, followed by prednisolone, and mycophenolate mofetil, with no response. In August 2018, the patient started haemodialysis, human immunoglobulin (transient response) and plasmapheresis due to evidence of secondary thrombotic microangiopathy. The refractory severe thrombocytopaenia and lupus nephritis, justified two cycles of rituximab – platelet count rising lasted for five months. Seven months later, the patient presented pancytopenia and due to high hemorrhagic risk, there was a switch from warfarin to LMWH (prophylactic dosing). One month later, anti-coagulation was suspended due to spontaneous intracranial bleeding. In August 2019, the patient had an ischemic stroke of the right occipital lobe, secondary to APS, under prednisolone and hydroxychloroquine, and with platelet count of 24.000/uL. Because of the bleeding risk, no anti-aggregation was instituted, and she started tacrolimus – the initial good response disappeared two weeks later. After consulting a rheumatology board, tacrolimus was kept and rituximab repeated, regardless of the hypogammaglobulinemia and no lymphocytes CD19+ count.

Conclusion: We experienced a severe case of secondary thrombocytopenia, that was refractory to multiple therapeutic agents. A reasonable response was obtained under rituximab. Tacrolimus was kept due to the reported cases of late effect. Ischaemic stroke is a challenging condition in patients thrombocytopenic and further clinical guidance is warranted.

CC336 – MIOPATIA NECROTIZANTE IMUNOMEDIADA COM ENVOLVIMENTO CARDÍACO EXTENSO

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Introdução: A miopatia necrotizante imunomediada (MNIM) é um subtipo raro de miopatia inflamatória que se caracteriza por fraqueza proximal severa e rapidamente progressiva, com envolvimento extramuscular pouco frequente. Associa-se à presença de anticorpo anti-signal recognition particle (anticorpo anti-SRP).
Caso clínico: Mulher de 49 anos, previamente saudável, internada no serviço de Reumatologia por quadro súbito com duas semanas de evolução, traduzido por fraqueza muscular proximal simétrica, disfagia para sólidos e cansaço, com limitação funcional grave (Manual muscle testing grading system – MMT 27). Uma semana antes, avaliada no Serviço de Urgência, verificou-se aumento da creatina quinase (CK) de 20.000 U/L. No internamento, analiticamente, apresentava CK 12.300U/L, mioglobina 6456ug/dL, lactato desidrogenase 2245 U/L, troponina T 2639 ng/L, proteína C reativa 2 mg/dL e anticorpo anti-SRP positivo forte. Foi medicada inicialmente com imunoglobulina intravenosa (IGIV) e com metilprednisolona 1000 mg, durante três dias. Iniciou posteriormente, prednisolona oral, na dose de 1mg/kg/dia. A biópsia muscular e o eletromiograma dos membros superiores e inferiores, foram compatíveis com miopatia inflamatória. O ecocardiograma não apresentou alterações de relevo, contudo a ressonância magnética cardíaca mostrou aumento global dos tempos T1 e T2 miocárdicos e hipersinal nas sequências T2-STIR, sugestivo de edema miocárdico generalizado. A tomografia computorizada de corpo, mamografia e ecografia pélvica permitiram excluir neoplasia oculta, apenas parcialmente, visto a doente ter recusado estudo endoscópico e manometria esofágica. Por ausência de melhoria clínica, iniciou micofenolato de mofetil na dose de 1000mg de 12/12h e uma semana depois, a gravidade da situação clínica motivou início de rituximab (RTX). Apresentou resposta clínica parcial (MMT 32) e teve alta referenciada para unidade de reabilitação motora. Duas semanas depois, foi reinternada por insuficiência respiratória tipo I e disfagia grave (para sólidos e líquidos). Foi transferida para Unidade de Cuidados Intensivos, onde se mantém atualmente, por necessidade de ventilação mecânica invasiva; repetiu IGIV e perfusão de ciclofosfamida (CYC), que não se repetiu por subsequentes infeções.
Conclusão: O envolvimento cardíaco nesta patologia está descrito em menos de 20% dos doentes. Esta miopatia é reconhecidamente mais resistente ao tratamento convencional, como o caso descrito documenta. Poderá haver benefício em associar precocemente diferentes imunomodeladores, nomeadamente a associação de RTX e CYC.

CC337 – SARCOIDOSE: UM CASO RARO ASSOCIADO AO USO DE TNFI

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Introdução: Os inibidores do fator de necrose tumoral (TNFi) têm-se mostrado muito eficazes no controlo de várias doenças reumáticas inflamatórias. Porém, são bem conhecidos alguns efeitos adversos (EA) particularmente as infeções. Contudo, existem efeitos adversos que ocorrem muito raramente. A sarcoidose, uma doença granulomatosa multissistémica, tem sido descrita na literatura como um EA raro associado aos TNFi.
Caso clínico: Homem de 38 anos com história de artrite psoriática, sob adalimumab há dois anos. Desenvolve, em outubro de 2018, um quadro de cansaço, fraqueza generalizada e perda ponderal (10% nesse mês). Um mês depois, inicia febre de acentuação vespertina intermitente (temperatura máxima de 38,5ºC) e ligeiro desconforto torácico. Analiticamente, apresentava aumento dos parâmetros inflamatórios e em tomografia computorizada (TC) do tórax múltiplas formações nodulares preenchendo a região mediastínica e o parênquima pulmonar. Suspendeu-se adalimumab com resolução espontânea das queixas.
Foi internado em janeiro de 2019, altura em que apresentou de novo miodesópsias bilateralmente. Ao exame oftalmológico verificava-se flebite bilateral e edema dos discos ópticos, sugestivo de hipertensão craniana. Efetuou-se punção lombar para análise de líquido cefalorraquidiano, que revelou um predomínio de linfócitos e cujo exame cultural foi negativo. Ressonância magnética crânio-encefálica e angiografia ocular, sem alterações. Repetiu TC do tórax mantendo padrão multinodular. A broncofibroscopia não documentou lesões endobrônquicas; no lavado bronco-alveolar a razão linfócitos CD4+/CD8+ era elevada e o exame cultural negativo. O Interferon Gama Release Assay foi negativo. Analiticamente, mantinha aumento dos parâmetros inflamatórios (velocidade de sedimentação 59 mm na 1ª hora e proteína C reativa 0,95 mg/dL), com um aumento concomitante da enzima conversora de angiotensina (103 U/L). Após reunião

multidisciplinar, dado o alto grau de suspeita clínica e o envolvimento multissistémico a justificar urgência no início do tratamento, dispensou-se realização de biópsia pulmonar e de mediastinoscopia. Assumiu-se o diagnóstico de sarcoidose, tendo iniciado prednisolona na dose de 1mg/kg peso, tendo se verificado melhoria clínica e analítica. De referir que não se verificou exacerbação das lesões de psoríase ou sinais de artrite, entesite ou dactilite.

Conclusão: O desenvolvimento de síndromes sarcoidose-like em doentes sob terapêutica TNFi é uma complicação pouco comum. Neste caso, com clínica sugestiva, foi fundamental a alta suspeição para o início rápido da terapêutica. Não têm sido descritas recidivas das lesões após switch para outro TNFi.

CC341 – ARTERITE DE CÉLULAS GIGANTES: 2 CASOS COM CONSEQUÊNCIAS

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Introdução: As infeções oportunistas são uma consequência potencialmente fatal da terapêutica imunossupressora utilizada no tratamento das doenças reumáticas sistémicas. O presente trabalho ilustra dois casos clínicos com intercorrência infeciosa grave.

Caso 1: Mulher de 79 anos, com história de insuficiência cardíaca, cardiopatia valvular (submetida a valvuloplastia mitral), hipertiroidismo e osteoporose não-fraturaria, seguida em consulta de reumatologia por polimialgia reumática com um ano de evolução, controlada com 7,5mg/dia de prednisolona (PDN). Inicia quadro de perda súbita da visão, bilateral, sem resposta a corticoterapia em alta dose (1mg/Kg/dia). Admite-se neuropatia ótica isquémica arterítica bilateral. É proposta terapêutica com tocilizumab 162mg/semana. Manteve esquema terapêutico durante dois meses, altura em que inicia quadro de prostração, febre (38.6ºC), anorexia, náuseas, dor generalizada, incontinência fecal e dor torácica esquerda em associação a múltiplas equimoses ipsilaterais. Após investigação etiológica, tem o diagnóstico meningite e choque séptico a Neisseria meningitidis, é medicada com vancomicina e é transferida para a unidade de cuidados intensivos onde acaba por falecer 24 horas após a admissão por falência multiorgánica.

Caso 2: Homem de 88 anos, com história de doença renal crónica (3/5) e adenocarcinoma da próstata em remissão (status pós-prostectomia radical), seguido em consulta de reumatologia por artrite reumatoide seronegativa, não erosiva, medicado com metotrexato (MTX) 10 mg/semana e dose baixa de corticóide com desmame progressivo de setembro de 2014 a abril de 2016. Manteve remissão clínica até abril 2017 altura em que inicia quadro de perda súbita de visão do olho esquerdo, com recuperação espontânea após 30 minutos, sem qualquer outra queixa associada. Observado pela oftalmologia no serviço de urgência, assumido o diagnóstico de arterite de células gigantes, com confirmação histológica, medicado com pulso de 1g de metilprednisolona, em 3 dias consecutivos, seguido de 60mg/dia de PDN e AAS 100mg/dia, sem recuperação da acuidade visual. Após três meses de terapêutica, é internado por quadro de febre, dispneia e lesões ulcerativas na mucosa oral, com agravamento progressivo e transferência para a unidade de cuidados intensivos por insuficiência respiratória global grave com necessidade de ventilação invasiva. Após estudo, tem o diagnóstico de pneumonia a Pneumocystis jirovecii e infeção aguda a citomegalovírus, cumprindo terapêutica dirigida (25 dias de trimetoprim sulfametoxazol e ganciclovir) associado a 80 mg/dia de PDN, com resolução do quadro respiratório e recuperação da visão do olho esquerdo.

Conclusão: A terapêutica imunossupressora deve ser usada de forma ponderada e criteriosa, na mínima dose possível. Contudo, a existência de patologias reumáticas sistémicas graves, com manifestação de lesão de órgão-alvo, impõem a necessidade de imunossupressão em alta dose. O risco de infeção está diretamente relacionado com a dose cumulativa de corticoterapia, o uso concomitante de outras terapêuticas imunossupressoras e ainda com o status fisiológico do doente. O uso de terapêutica profilática, o ensino de sinais de alarme e uma avaliação regular constituem a melhor forma de prevenir ou detetar atempadamente um quadro infecioso, que pode ter um desfecho fatal.

CC351 – LOMBALGIA INFLAMATÓRIA BAIXA: QUANDO O "SINTOMA-CHAVE" NÃO SE TRADUZ NUMA ESPONDILARTRITE

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Introdução: A lombalgia inflamatória baixa (LIB) é um sintoma-chave do diagnóstico das espondilartrites axiais (axSpa), sendo incluída nos critérios de classificação destas patologias. Contudo, é uma

característica que se tem mostrado ser bastante inespecífica, tornando necessário ter em conta diversos outros diagnósticos diferenciais que se podem apresentar através deste sintoma, incluindo patologia não reumatológica. O presente caso ilustra esta mesma necessidade.

Descrição: Mulher, 45 anos, sem antecedentes pessoais ou familiares de relevo, com quadro de lombalgia baixa e glutalgia de ritmo inflamatório desde os 39 anos, notando agravamento em períodos prolongados na posição sentada e durante o período menstrual. Sem história de artrite periférica, dactilite, alterações gastrointestinais, cutâneas, oculares ou outras queixas concomitantes de órgão ou sistema.

Foi encaminhada para consulta de Reumatologia aos 41 anos. No exame objectivo a salientar manobras sacro ilíacas positivas. No estudo complementar, a realçar VS e PCR consistentemente baixas e positividade Ag HLA-B27. Nos exames imagiológicos dirigidos às articulações sacro-ilíacas (TC e RMN), a TC identificou alterações ligeiras e inespecíficas e a RMN não evidenciou alterações estruturais ou de atividade enquadráveis em sacroileíte. A TC da coluna lombar revelou vértebra transição da charneira lombo-sagrada, com mega-apófise transversa bilateral, condicionando à direita pseudartrose com a asa do sacro e sinais de possível instabilidade da charneira lombo-sagrada.

Instituiu-se AINE por ciclos, com controlo razoável da sintomatologia. Por queixas recorrentes axiais e posteriormente também coxalgia bilateral de ritmo inflamatório, optou-se por iniciar sulfassalazina, com resposta pouco significativa, pelo que se veio a suspender.

Após um período de 3 anos de seguimento, verificou-se agravamento da lombalgia e glutalgia, com diminuição da resposta ao AINE. Por fraca correlação com os achados clínicos e resultados do estudo complementar no âmbito das patologias do foro articular inflamatório, foram reequacionadas as hipóteses diagnósticas e enviada para avaliação em consulta de Ginecologia, sendo admitida pela primeira vez a possibilidade de endometriose em localização ano-rectal. A RMN pélvica confirmou a hipótese, revelando a presença de endometriose profunda expressiva, traduzida por nódulo (2.2 x 1.6 x 2.5 cm) localizado a nível subperitoneal, com infiltração até ao ligamento útero-sagrado esquerdo e infiltração ainda da parede pélvica lateral posterior esquerda, do paramétrio e paracolpos posterior, vagina, gordura pararrectal e parede do recto de forma transmural numa extensão de 1 cm; o nódulo apresentava o seu limite mais baixo a cerca de 9 cm da margem anal e estendia-se até às raízes sagradas S2-S3, comprometendo também o plexo hipogástrico inferior a este nível. A doente está atualmente em seguimento na consulta de Ginecologia.

Discussão: A endometriose consiste na presença de tecido mucoso endometrial normal implantado noutras localizações além da cavidade uterina. A deposição no ligamento útero-sagrado está entre as mais comumente descritas na literatura, podendo apresentar-se por LIB e/ou agudização cíclica coincidente com os períodos menstruais. Perante uma mulher em idade fértil e contexto clínico sugestivo, o reumatologista deve ponderar a inclusão desta hipótese no diagnóstico diferencial de uma axSpa. A RMN pélvica, quando justificada por uma elevada suspeição clínica, possui um papel chave no diagnóstico do acometimento desta localização.

CC352 – SÍNDROME ANTI SINTETASE – UM CASO CLÍNICO COM EXPRESSÃO INCOMPLETA

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Introdução: A síndrome anti sintetase (SAS) é uma patologia imunomediada, caracterizada por manifestações musculares, pulmonares e articulares e presença de anticorpos anti sintetase. Apresentamos um caso clínico altamente sugestivo da patologia.

Descrição: Mulher, 53 anos, sem antecedentes pessoais relevantes, antecedente de mãe com artrite reumatóide, iniciou quadro de 2 semanas de evolução de astenia, dispneia progressiva a médios esforços, lesões aftosas da mucosa oral, tosse seca, xerostomia, xeroftalmia e dor torácica inspiratória, sem febre, sendo estabelecido o diagnóstico de pneumonia adquirida na comunidade e realizada antibioterapia. Por manutenção das queixas, recorreu ao SU e foi internada. Destacam-se nos exames complementares: hemograma com leucocitose (12,2x109/L) e neutrofilia (10,2x109/L), PCR 16 mg/L (<5), sem alterações da função renal; Rx de tórax com hipotransparência basal bilateral; gasimetria com hipoxémia (pO2 61,4); angioTC sugestivo de pneumonia organizativa; broncofibroscopia com biópsia transbrônquica (histologia normal) e lavado broncoalveolar (leuc. 500 cél/mm3 – PMN 10 %, eos. 2 %, cél. mononucl. 22 %, linf. 66 %; límpido, incolor); estudo imunológico com ANA, FR, ac. anti-CCP, ac anti RNP, ac anti Scl70, ac anti centrómero B e ANCA MPO/PR3 negativos. Admitiu-se pneumonia organizativa e iniciou PDN 1mg/kg/dia (50 mg/dia) com melhoria clínica e analítica progressivas. Após alta,

coincidente com desmame da corticoterapia, iniciou edema difuso das mãos, rigidez matinal prolongada, poliartralgias inflamatórias das mãos e punhos, fissuração dos dedos ("mãos de mecânico"), síndrome de raynaud (SR) e perda ponderal >10% em 6 meses, sem mialgias ou défices de força muscular. As provas de função respiratória tinham diminuição grave de FVC (66,4%) e DLCO (4,6%) e o TC de controlo mostrou agravamento imagiológico (padrão subpleural de espessamento dos septos interlobulares, vidro despolido, áreas de consolidação na periferia dos lobos pulmonares, mais nos LIs; bronquiectasias de tração). Analiticamente, surgiu ANA positivo 1/640, anti-SSA 52kDa e anti Jo-1 positivos. Em análises sucessivas, houve aumento pontual de CK (265 UI/L), com posterior normalização. O ecocardiograma não mostrou hipertensão pulmonar ou outras alterações. Em reunião multidisciplinar de patologia do interstício pulmonar, admitiu-se provável SAS com envolvimento pulmonar. Fez indução com 6 ciclos mensais de ciclofosfamida EV (1º ciclo 500 mg/m2, seguintes 750 mg/m2), seguidos de terapêutica de manutenção com micofenolato de mofetil (2g/dia). Pelo SR foi medicada com amlodipina, nitroglicerina, pentoxifilina e ácido acetilsalicílico, mas por agravamento da fissuração dos dedos fez ciclo de alprostadilo EV (5 dias consecutivos), com boa tolerância e melhoria significativa dos sintomas.

Atualmente encontra-se clínica e analiticamente melhorada (PCR <3 mg/L). A TC de controlo mais recente mostra ligeira evolução favorável e houve também melhoria a nível da FVC (73%) e DLCO (46,6%).

Discussão: A SAS tem uma maior prevalência e gravidade de DPI dentro das miopatias. As diferentes manifestações podem ser assíncronas: aqui vemos uma expressão incompleta do SAS, sem miosite clinicamente valorizável até à data. As restantes características estão presentes, com predomínio do envolvimento pulmonar, que motivou a instituição de terapêutica imunossupressora. Conseguindo-se em bom controlo a este nível, pode ser expectável que o componente muscular não se venha a demonstrar.

CC353 – DESAFIO TERAPÊUTICO EM VASCULITE ANCA ASSOCIADA

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Introdução: As vasculites ANCA associadas (VAA) são patologias imunomediadas raras, mais frequentes em homens e idade avançada, com mortalidade elevada e necessidade de terapia imunossupressora a longo prazo. Descreve-se aqui um caso clínico com envolvimento multiorgânico grave.

Descrição: Homem, 62 anos, com antecedentes de tabagismo (50 UMA), recorreu ao SU por quadro de dispneia e hemoptises com 5 dias de evolução, de agravamento progressivo.

O estudo etiológico foi a favor de quadro compatível com vasculite ANCA anti MPO+ (>300 UA/mL) de apresentação inicial grave, com envolvimento pulmonar e hemorragia alveolar difusa (HAD) confirmada por TC e broncofibroscopia, e repercussão hemodinâmica com necessidade de suporte transfusional (valor Hb admissão 51 g/L). Cumpriu 2 pulsos de 1500 mg (3 g de dose cumulativa) de ciclofosfamida (CYC) e 5 pulsos de 1000 mg de metilprednisolona (MPDN), após o que iniciou doses de 1 mg/Kg/dia (60 mg) prednisolona (PDN) oral. Por melhoria clínica, fez desmame até 40 mg. Contudo, o TC de controlo aos 3 meses revelou novas áreas de HAD. Verificou-se ainda envolvimento oftalmológico compatível com vasculite retiniana bilateral e envolvimento renal, nomeadamente lesão renal aguda KDIGO 3 oligúrica intrínseca com necessidade transitória de diálise e posterior recuperação. Ao longo do internamento, desenvolveu pancitopenia em provável contexto de terapêutica efetuada (Hb 83 g/L, Leuc 2.100x109/L, N 1.400 x109/L, Plaq 102x109/L) e diversas intercorrências infeciosas com necessidade de antibioterapia: pneumonias associadas ao ventilador (p. aeruginosa multirresistente), peritonite biliar por colecistite isquémica perfurada e infeção do trato urinário. Atendendo ao elevado risco infecioso e citopenias, com elevada actividade da doença, optou-se por realizar 2 ciclos de imunoglobulina humana (400 mg/Kg/dia em 5 dias, dose total 2 g/Kg) e manteve PDN 40 mg/dia. Após melhoria clínica e analítica, teve alta para Unidade de Curta duração, onde ocorreu deterioração da função renal (Cl<15 ml/min/1.73m2) e proteinúria nefrótica (Prot/urina 24h 4,389 g). No reinternamento, realizou inicialmente pulsos de MPDN (500 mg, 3 dias) e posteriormente retomou dose de 60 mg/dia de PDN. A biópsia renal mostrou alterações de esclerose global sequela de glomerulonefrite, com fibrose marcada e sinais residuais de inflamação crónica. Em discussão com Nefrologia, considerou-se baixa probabilidade de resposta a repetição de terapêutica imunossupressora de indução e foi necessário iniciar terapêutica renal de substituição a curto prazo com hemodiálise. Foi decidida terapêutica de manutenção com Rituximab (500 mg aos 0 e 14 dias e depois 6/6 meses). Atualmente o doente já cumpriu as 2 perfusões iniciais e encontra-se em

esquema de desmame de PDN (dose actual 15 mg/dia), com estabilização do quadro.

Discussão: As VAA têm frequentemente envolvimento multissistémico e um pleomorfismo de manifestações clínicas que pode dificultar e atrasar o diagnóstico. O envolvimento renal e respiratório é frequente e a falência destes sistemas e as infeções constituem a principal causa de mortalidade nos doentes. No presente caso de apresentação grave, o diagnóstico e a terapêutica foram estabelecidos precocemente, mas as múltiplas intercorrências infeciosas representaram uma dificuldade adicional na abordagem e decisão terapêutica, não se conseguindo evitar lesão renal grave e irreversível.

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