confounding by treatment. Consecutive bacteremic patients were identified from an associated paediatric intensive care unit over the same period. Descriptive statistics and univariate logistic analyses were performed as appropriate.

**Results:** Patient characteristics are summarised in Table 1; bacteremic patients were younger. PCT was elevated in bacteremic patients, and was undetectable in all other subjects (Table 2). There were trends towards higher ESR and CRP in bacteremic patients, but these were not statistically significant.

**Conclusions:** Serum PCT levels appear to be a reliable biomarker to distinguish infection vs. active JIA at presentation, and can aid in directing therapy. However, PCT does not appear useful to assess disease activity in JIA. Further studies are needed to assess utility of serum PCT measurement in differentiating JIA flares from less severe infections.

#### **REFERENCE:**

[1] Milcent K, et al., "Use of Procalcitonin Assays to Predict Serious Bacterial Infection in Young Febrile Infants," JAMA Pediatrics 170, no. 1 (2016): 62– 69; Mohsen A, et al., "Predictive Values for Procalcitonin in the Diagnosis of Neonatal Sepsis," Electronic Physician 7, no. 4 (2015): 1190–95.

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AB1119

CLINICAL, THERAPEUTIC CHARACTERISATION AND TIME TO ACHIVE REMISSION ANALYSIS OF A COLOMBIAN COHORT WITH JUVENILE IDIOPATHIC MYOPATHY

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**Background:** The clinical characteristics of paediatric patients with idiopathic inflammatory myopathies difere from adutts in several aspects. Its clinical presentation can include amyopathic onset and the skin involvement has diferent charecteristics

**Objectives:** To describe a Colombian cohort with Juvenile Myositis (JM) recruited in a rheumatology facility.

**Methods:** A cross-section retrospective research with data collected between 2014 and 2017 from a population diagnosed before 16 years of age with Idiopathic Myopathy according to Peter and Bohan criteria and followed up for at least six months. Kaplan-Meier curves were performed to analize time to achieve remission

Results: Out of 37 patients, one was excluded for having a dystrophy myopathy gene, 73% fulfilled definitive and 16% probable Bohan and Peter criteria; most patients were female 75,8%,<sup>28</sup> with mean age of onset 7,2 years, and clinical remission was achieved on average at 4 years of disease. There was high prevalence of Gottron's sign and papules (89%), Heliotrope rash (62%) and Calcinosis (37%). Other involvements are described in Table 1. Antinuclear antibodies were positive in 52%. Electromyography (EMG) was positive for myopathy in 39% of the patients. Biopsy was compatible with myopathy in 10% and was negative in 32% of the patients. The most common treatment was metrothexate (91%) followed by antimalarials (72%) and corticoids (56,7%). Medication used in severe forms included Cyclophosphamide (5%), Rituximab (16%) and IV Immunoglobulin (5%). Kaplan-Meier curves showed an earlier time to remmision in patients without them (HR:8,25 HR CI95%IL:1,076–63,3;p=0042 and in childrens younger than 15 years compared to older patients (HR:2,529 HR95%IL:1,084–5,901, p:0,039).

Abstract AB1119 - Table 1. Clinical characteristics of Colombian patients with JM.

Cahracteritstics in JM n=37	N	%
Symmetrical muscle weakness	27	72,97
Gottron's papules	33	89,19
Heliotrope rash	23	62,16
Calcinosis cutis	14	37,84
Gastrointestinal involvement	6	16,22
Pulmonary involvement	4	10,81
Articular involvement	9	24,32
Amyopathic	9	24,32
ANA(+)	14/	37,84
	27	
EMG Myopathic changes	9/23	24,32
Biopsy-proven myopathy		
Positive	4	10,81
Negative	12	32,43

**Conclusions:** Our results agreed with those obtained in other multi-centred studies including latin america that evaluated clinical and therapeutic characteristics in children with myopathy, Gottron's sign and papules being the most common findings and with high rates of calcinosis and joint involvement. There was a significant difference between remission lapse in patients younger than 15 years compared to older ones.

### **REFERENCE:**

 , D. Guseinova, et al. Comparison of clinical features and drug therapies among European and Latin American patients with juvenile dermatomyositis. Clinical and Experimental Rheumatology 2011; 29: 117–124

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AB1120

THYROID HORMONE CONCENTRATIONS IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS FROM A SINGLE TERTIARY REFERRAL CENTRE

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**Background:** Despite mounting evidence linking both endocrine disorders and rheumatic diseases, there is a lack of studies investigating any association between the prevalence and clinical characteristics of thyroid disorders and juvenile idiopathic arthritis (JIA).

**Objectives:** The aim of this study is to assess the prevalence of abnormalities in thyroid function in patients with JIA, and to investigate the possible association between this endocrine disorders and specific disease activity markers.

**Methods:** Thirty patients diagnosed with JIA according to the International League of Association for Rheumatology were screened for thyroid diseases. We performed stratified analyses by sex, age, subtype of JIA, disease duration, the Juvenile Arthritis Disease Activity Score (JADAS-71), clinical peculiarities, laboratory values and ultrasound examination of thyroidal gland.

Results: Our results revealed that 67% of patients were girls. The mean age of the studied group was 127,56±8,8 months, the median age at diagnosis was 74,33±8,49 months and the median disease duration was 50,83±9,33 months. The most frequent types of JIA were oligoarticular (40%), polyarticular negative RF (34%) and systemic (20%). The median JADAS-71 score was 16,91±1,64 [range values from 5 to 34]. The status of the thyroid function in those patients was euthyroidism. Contrary to other findings in the literature, a high free triiodothyronine was recorded in 33% of cases. However, specific antibodies as antithyroglobulin and antithyroid peroxidase were not detected in any patients. The ultrasound examination of thyroidal gland revealed abnormalities in 30% cases, most of them cystic changes (26,6%) and hypo-echogenicity (23,33%). In 2 cases were detected 2 thyroid nodules. Furthermore, 2 patients presented mean thyroid volume above 2SDS according their age reference values. An increased vascular flow pattern on Doppler examination of thyroidal gland was found in 10% cases. Correlation and regression analysis showed low age at diagnosis and JADAS-71 score (more than 20) to be predictors for those thyroid disorders.

**Conclusions:** The goal of early identification of endocrine comorbidities in rheumatic diseases is to prevent and limit the clinical disease impact. The identification of autoimmune diseases in preclinical stage secondary to juvenile idiopathic arthritis allow a better disease control and quality of life.

### **REFERENCES:**

- [1] Tronconi E., Miniaci A., Pession A., The autoimmune burden in juvenile idiopathic arthritis, Italian Journal of Pediatrics, 2017, 43:56.
- [2] Elnady B.M., et all., Prevalence and clinical significance of nonorgan-specific antibodies in patients with autoimmune thyroiditis as predictor markers for rheumatic diseases, Medicine, 2016, 95:38 (e4336).
- [3] Robazzi T.C., Adan L.F., Autoimmune thyroid disease in patients with rheumatic diseases, Rev Bras Reumatol, 2012; 52 (3): 417–430.

**Disclosure of Interest:** None declared **DOI:** 10.1136/annrheumdis-2018-eular.7293

AB1121

EVALUATION OF CASES DIAGNOSED WITH CRMO; SINGLE CENTRE EXPERIENCE

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**Background:** Chronic recurrent multifocal osteomyelitis (CRMO); is a rare auto-inflammatory bone disease characterised by recurrent, sterile inflammatory

lesions occurring primarily in children and adolescents. Symptoms of presentation may range from mild unspecific bone pain, local swelling and warmth to severe pain, malaise, fevers and even fractures.

**Objectives:** In this study, we aimed to evaluate our patients who had a diagnosis of CRMO, retrospectively.

**Methods:** Six patients who were diagnosed with CRMO between 2010–2017 years were included in the study. The CRMO diagnosis was based on characteristic clinical features and magnetic resonance imaging findings. The clinical data were obtained from the records of electronic files.

**Results:** The female to male ratio of the cases was 4/2 and the median age was 11.15 years.  $^{6-12}$  The age of diagnosis was 10.35 years (4-12.5), the median period for diagnosis delay was 3 years (0.75-8). The most common complaint was localised pain  $(n=6,\ 100\%)$ . Accompanying diseases were detected in 3 patients; 1 case had inflammatory myositis, 1 case had PFAPA syndrome and 1 case had selective IgA deficiency. Multifocal bone involvement was present in 4 (66%) cases and unifocal bone involvement in 2 (33%) cases. The most common site of disease was femur. Acute phase reactants were high most of the cases; elevated erythrocyte sedimentation rate (ESR) in 5 cases  $(83.3,\ n=6)$ , elevated creactive protein level in 4 cases  $(66.6\%,\ n=6)$ , elevated serum amyloid a level in 3 cases  $(60\%,\ n=5)$ , and elevated fibrinogen in 2 cases  $(50\%,\ n=4)$  were present. ANA was found positive a low titer in only 1 case, whereas rheumatoid factor was negative in all cases. Non-steroidal anti-inflammatory drugs were prescribed in all cases and anti TNF drugs in 3 (Etanercept in 2 cases and adalimumab in 1 case). Clinical characteristics of the patients are given in Table 1.

Abstract AB1121 - Table 1. Clinical findings of the cases

Initial complaints of cases				
	n	%		
Pain	6	100		
Walking abnormality	4	66.7		
Swelling on bone	1	16.7		
Weight loss	1	16.7		
Distribution of involved bones				
Femur	4	66.7		
Iliac bone	2	33.3		
Tibia	2	33.3		
Calcaneus	1	16.7		
Vertebral column	1	16.7		
Acetabulum	1	16.7		
Treatment				
Ibuprofen	3	50		
Naproxen Na	2	33.3		
Aspirin	1	16.7		
Indomethacin	1	16.7		
Adalimumab	1	16.7		
Etanercept	2	33.3		

**Conclusions:** The diagnosis of CRMO is difficult and no consensus exist on diagnosis and treatment. Multifocal bone lesions with characteristic radiological findings are very suggestive of CNO. The first line treatment is usually NSAIDs, however, anti TNF treatment are needed in some patients to achieve for remission. Our case is the second one who had inflammatory myositis and CRMO according the literature.

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AB1122

## DIALYSIS TREATMENT OF SYSTEMIC VASCULITIS IN PAEDIATRIC PATIENTS

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**Background:** Childhood-onset systemic vasculitis is a rare but serious condition with high mortality rate even with proper treatment. Renal involvement at presentation is a high risk for end stage renal disease (ESRD).

**Objectives:** The aim of our study was to review the results of the dialysis treatment in paediatric patients with systemic vasculitis in a single dialysis centre for children in Bulgaria.

**Methods:** For a period of 20 years we observed 9 clinical cases of systemic vasculitis -5 cases of Goodpasture syndrome (GPS) -4 girls and 1 boy from 6 to 17 years old; one 6 year old girl and one 12 year old boy with Granulomatosis with polyangitis (GPA); an 8 month old boy with microscopic polyangiitis (mPAN), and one 8 year old girl with Takayasu arteritis. Seven of the children were treated by hemodialysis because of progressive kidney failure leading to ESRD, one of them was treated by continuous ambulatory peritoneal dialysis (CAPD) and one conservatively.

Results: In the GPS group one of the girls still continues on hemodialysis with no pulmonary symptoms for more than 14 years follow-up. The second girl was treated with immunoadsorption after which reminded antibody negative. On the follow-up she is managed conservatively for chronic kidney disease. Two of the girls died because of severe pulmonary bleeding caused by exposure to fragrant smoke during incense. The boy was treated conservatively because of mild pulmonary and kidney involvement and died from pulmonary bleeding caused by smoke. In the GPA group the girl underwent kidney transplantation and died one month later, and the boy died because of severe pulmonary bleeding. The 8 month old boy with mPAN was treated by dialysis with some clinical improvement but after six months died because of thrombus in the right atrium. The girl with Takayasu arteritis was treated with CAPD but died because of cardiopulmonary complications.

**Conclusions:** End stage renal disease is poor prognostic factor for survival in paediatric patients with systemic vasculitis. 75% of the children died in a period of 5 months after initiation of dialysis treatment.

**Disclosure of Interest:** None declared **DOI:** 10.1136/annrheumdis-2018-eular.5836

AB1123

DO CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS PLAY AN ACTIVE ROLE IN THEIR TREATMENT ADHERENCE? FIRST RESULTS OF THE RUMAJI STUDY

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**Background:** Adherence to DMARDs such as methotrexate and biologics is critical for patients with Juvenile Idiopathic Arthritis (JIA). Notwithstanding, few studies exists on that topic and we lack information to understand the grounds for adherence.

**Objectives:** The RUMAJI study aims, among others, to understand and decipher the parents and children adherence mechanisms and practices.

**Methods:** Qualitative methods were chosen in order to investigate parents' and children's everyday life with JIA and its treatment. An ethnographic study was designed by a multidisciplinary team including rheumatologists, paediatricians, patient associations members and anthropologists. The study involved 15 families (enough to reach saturation), recruited from 5 centres by diversity of clinical and sociological profiles. The panel included 17 children with JIA, 11 girls and 6 boys, median age 10,<sup>3; 17</sup> median disease duration 2.5.<sup>1; 15</sup> 4 children were treated with conventional DMARDs in monotherapy, 4 with biologic DMARDs in monotherapy, 5 with cDMARD-bDMARD association and 4 with NSAIDs only.

Interviews were conducted by anthropologists at family's home using in-depth semi directive and biographic methods. 3 fields were explored: organisation of everyday life with JIA, treatment practices, impact on school and social activities. Interviews were recorded and transcribed for analysis.

Results: Adherence results from an appropriation process of the JIA and treatment that require both an active role from parents and children, even before the transition. This active role played by children could be either stimulated or inhibited at home according to the family's structure, social background and parents' attitudes toward their child (participation to the decision, explanation of the disease)

Children's active role includes in particular: 1) negotiations with parents and physician, 2) experiments with the treatment (forgetting or involuntary switch from the parents, changing the dosage on their own initiative) and 3) participation to the treatment administration and ritualization.

The manner children consider and manage their DMARDs is the result of an arbitration depending on the positive (a) and side effects (b) they felt in their body and the effects noted by the doctors (c) during the examinations and test results. Dealing with these 3 dimensions requires to link together both a theoretical and practical knowledge of JIA. Thus, children build their own and singular knowledge of their disease and treatment, which is a source of control of their body and their life. **Conclusions:** Qualitative methods, through an ethnographic study starting from children's point of view, underline the active role they play in their care. Adherence to DMARDs could be improved by supporting children's implication as soon as the beginning of JIA.

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AB1124

### A UK STUDY: VOCATIONAL EXPERIENCES OF YOUNG ADULTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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**Background:** Little is known about the experiences of young adults living with Juvenile Idiopathic Arthritis (JIA) preparing for employment and career development.

**Objectives:** The purpose of this study was to understand the impact JIA has on career planning and early employment experiences of young adults (16–30 years).

**Methods:** Using existing literature (including grey literature), an online survey (consisted of 152 questions, 29 items related to young adults two of which were free text questions) was developed and sent to UK National Rheumatoid Arthritis Society (NRAS) members and distributed to non-members via social media tools including Facebook, Twitter and HealthUnlocked. Data collected included views and experiences in career planning and employment. The data pertaining to young adults are presented here.

Results: Of 1241 respondents 19 were young adults with JIA (range 16-30 years), 89% were female and 84% had university or equivalent qualifications. Due to incomplete responses there is missing data on all 19 young adults. 4/13 young adults were studying at university, 9/13 were in paid employment. 9/17 respondents reported their school did not offer additional work-related activities to students with disabilities and/or additional needs. 10/14 young adults felt their school did not provide advice about coping with possible limitations on placements/traineeships due to their arthritis. 11/14 respondents did consider their condition when thinking about future career plans e.g. "I wanted to work as a ranger or similar for the National Trust but it's a fairly physically demanding job and I knew my joints would suffer so I changed track slightly". However, 8/14 felt their career advisors at school/university did not take their arthritis into account e.g. "I had to cease my physiotherapy master's degree as my arthritis got too bad to continue and change career choice. I wish there would have been more discussion about it not being a reasonable choice for me at the time as we just didn't have the information then". 8/14 young adults changed their career plans because of their arthritis with managing JIA symptoms and a physically demanding role, as well as wanting to stay healthy, being the main reasons for changing career. Important aspects of employment included: "good relationships with your line manager, work you like doing and a job you can use your initiative".

Conclusions: Despite small numbers these results highlight potential current unmet vocational needs of young adults with JIA in the UK and the need for further research with this age group. There appears to be a lack of structured support within schools and universities offered to students with disabilities and/or additional needs, about work-related activities and careers. Young adults with JIA actively consider their condition whilst thinking about career opportunities and value a productive and challenging job with a good working environment, including relationships with colleagues and supervisors.

**Disclosure of Interest:** None declared **DOI:** 10.1136/annrheumdis-2018-eular.2099

AB1125

# URINARY SOLUBLE CD25 AS A BIOMARKER OF ACTIVE LUPUS NEPHRITIS IN EGYPTIAN CHILDREN WITH JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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**Background:** Lupus nephritis (LN) is more prevalent and severe in children than adult and considered a major predictor of poor outcome. Thus, early diagnosis and treatment is associated with better outcome. Soluble CD25 (sCD25), also known as Interleukin-2 receptor alpha chain, is a type I transmembrane protein present on activated T lymphocytes that play important role in the pathogenesis LN[.1]

**Objectives:** This study aimed to measure urinary levels of sCD25 in children with juvenile systemic lupus erythematosus (JSLE) and to investigate its role as a potential biomarker of activity in LN.

**Methods:** We measured sCD25 using enzyme-linked immunosorbent assay in urine samples from 53 JSLE patients and in urine samples from 30 healthy controls and these levels were normalised to creatinine excretion in urine. All JSLE patients underwent thorough clinical examination and disease activity assessment using Systemic Lupus Erythematous Disease Activity Index (SLEDAI).

Systemic Lupus International Collaborating Clinics (SLICC) renal activity score<sup>2</sup> was used to assess activity of LN.

Results: Urinary sCD25 normalised levels were highly significantly increased in JSLE patients (278.38±133.68 pg/mg) compared to urinary level in the healthy controls (187.33±83.59 pg/mg),p<0.001. Also, patients with active LN had significantly higher normalised urinary sCD25 levels (402.69±139.58 pg/mg) compared to urinary level in active JSLE patients without LN (262.18±98.35 pg/mg), p=0.002 and inactive JSLE patients (192.7±66.4 pg/mg), p<0.001. In JSLE patients, urinary sCD25 normalised levels significantly correlated with SLEDAI (r=0.48, p<0.05), renal SLEDAI (r=0.61, p<0.001), SLICC renal activity score (r=0.68, p<0.001) and C3 (r=-0.48, p<0.001).

**Conclusions:** JSLE patients have significantly increased urinary levels of sCD25 especially in those with active LN. Urinary sCD25 levels are remarkably correlated with the renal disease activity scores suggesting that it could be a useful marker to reflect active renal involvement in JSLE patients.

### **REFERENCES:**

- Gupta R, Yadav A, Misra R, Aggarwal A. Urinary sCD25 as a biomarker of lupus nephritis disease activity. Lupus 2015; Mar;24(3):273–9.
- [2] Petri M, Kasitanon N, Lee SS, Link K, Magder L, Bae SC, et al. Systemic Lupus International Collaborating Clinics (2008) Systemic lupus international collaborating clinics renal activity/response exercise. Development of a renal activity score and renal response index. Arthritis Rheum 2008; 58 (6):1784–88.

**Disclosure of Interest:** None declared **DOI:** 10.1136/annrheumdis-2018-eular.4494

AB1126

## NO RADIOGRAPHIC WRIST DAMAGE AFTER TARGETED TREATMENT IN JUVENILE IDIOPATHIC ARTHRITIS

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**Background:** Juvenile idiopathic arthritis (JIA) is characterised by chronic inflammation of the joints which can lead to structural bone damage.

Objectives: The objective of this study was to evaluate the response of new onset JIA patients to an early targeted treatment by conventional radiography.

Methods: JIA patients participating in the BeSt for Kids study (NTR 1574) were eligible in case of wrist involvement at inclusion and if conventional radiographs were available at baseline or within 6 months before or after study inclusion. Follow-up radiographs of hands and wrists after 12–36 months were available for comparison. Radiographic bone damage as reflected by carpal length was assessed using the Poznanski score<sup>1</sup>, providing 'Z' as indication of the deviation from a healthy population as measured by radiometacarpal length relative to the second metacarpal length (RM/M2). BoneXpert method<sup>2</sup> was used to automatically determine bone age and bone mineral density (BMD) of the wrist.

Abstract AB1126 - Table 1

	Baseline Z- score (95% CI)	Compared to healthy population	Follow-up Z- score (95% CI)	Compared to healthy population	Change in Z- score
Poznanski	0.047 (-0.32	p=0.795	0.055 (-0.28	p=0.744	p=0.937
score	to 0.41)		to 0.39)		
BMD	-0.71 (-1.12	p=0.001	-0.44 (-0.75	p=0.008	p=0.032
	to $-0.30$ )		to -0.12)		
Bone age	-0.08 (-0.44	p=0.651	-0.25 (-0.59	p=0.574	p=0.092
	to 0.28)		to 0.09)		

Results: Forty JIA (27 female) patients were evaluated for Poznanski score and BMD (mean age 7.2±3.4 years), 26 patients (15 female) were evaluated for bone age (mean age 9.3±2.2 years). Assessed by the mean Z-score of RM/M2, we did not detect wrist damage at baseline nor at follow-up. Assessed by the mean Z-score of the bone age, we did not detect deviating bone age at baseline nor at follow-up. At baseline BMD was significantly diminished compared to healthy

controls (Z-score -0.71, 95% CI=-1.12 to -0.30). BMD at follow-up improved significantly (Z-score -0.44, 95% CI=-0.75 to -0.12, p=0.032). Results are summarised in table 1

**Conclusions:** In this cohort of JIA patients treated early and targeted at inactive disease, we have detected no radiographic wrist damage at baseline or follow-up as detected by Poznanski score. BMD was significantly diminished at baseline but improved significantly after follow-up.

#### REFERENCES:

- [1] Poznanski AK, Hernandez RJ, Guire KE, Bereza UL, Garn SM. Carpal length in children – a useful measurement in the diagnosis of rheumatoid arthritis and some congenital malformation syndromes. Radiology 1978:129:661–8.
- [2] Anink J, Nusman CM, van Suijlekom-Smit LW, van Rijn RR, Maas M, van Rossum MA. Automated determination of bone age and bone mineral density in patients with juvenile idiopathic arthritis: a feasibility study. Arthritis Res Ther 2014:16:424.

**Disclosure of Interest:** None declared **DOI:** 10.1136/annrheumdis-2018-eular.5306

### Other orphan diseases\_

AB1127

PULMONARY ARTERIAL HYPERTENSION AND POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES) IN A PATIENT WITH ADULT ONSET STILL'S DISEASE

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**Background:** Pulmonary arterial hypertension is a rare complication of AOSD and there are only a limited number of case reports in the literature.

PRES is a rare acute neurological condition characterised by rapid onset of headache, seizures, altered consciousness, visual disturbances and usually very high blood pressure. Brain imaging characteristically shows high signal change in the subcortical white matter, predominantly in the posterior lobes which normalises within days to weeks. There are rare case reports of seizures and other neurological manifestations associated with AOSD but no published case reports of classic PRES

**Objectives:** To share this interesting case with our rheumatology colleagues.

**Methods:** We present a case of 24 year old Afro-Caribbean lady, diagnosed with AOSD in December 2015, presenting with recurrent fevers, weight loss, polyarticular synovitis, small volume lymphadenopathy, evanescent urticarial rash, hyperferritinemia (3700 ug/L) and raised CRP (146 mg/L). Rheumatoid factor, ANA, CCP, ENA and ANCA were negative. Infection screen was negative including blood-borne viruses and whole-body imaging was normal.

She was initially treated with pulsed Methylprednisolone 1 g IV for 3 days followed by 40 mg oral prednisolone. She had a good initial response (ferritin 1700 ug/L, CRP 36 mg/L), but subsequently we were unable to reduce her prednisolone below 35 mg due to recurrence of symptoms.

She had quite a stormy course over the rest of the year with a number of hospital admissions and her ferritin running as high as 35,000 ug/L and CRP more than 200 mg/L.

In February 2017 she had a further severe flare and was started on Anakinra. She initially responded well (Ferritin 400 ug/L, CRP 3 mg/L), but four months later started to flare again, requiring a further admission, treated with IV Methylprednisolone.

She was switched from Anakinra to Tocilizumab but was stopped after 4 doses due to poor response.

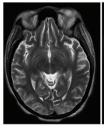
Following development of exertional dyspnoea, echocardiography and right heart catheter studies demonstrated a raised mean pulmonary artery pressure of 42 mmhg with severe TR, right sided volume overload and a BNP of 8741 ng/L warranting referral to the regional PAH centre.

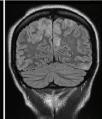
She also developed peripheral sensorimotor neuropathy in her lower limbs confirmed by NCS.

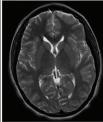
Results: In December 2017 she was admitted with severe shortness of breath, hypoxia and a ferritin of over 15,000 ug/L. She developed seizures with status epilepticus, very high blood pressure and ended up requiring mechanical ventilation. MRI and CT brain were suggestive of PRES with subcortical high signal change and symmetrical vasogenic oedema in occipital and parietal lobes. She was treated in Neuro ITU with anti-epileptics, anti-hypertensives, IV hydrocortisone and Anakinra was restarted. She made a rapid and full neurological recovery with resolution of changes on her brain scans.

She continues Anakinra, and Cyclosporine 2 mg/kg body weight has been added since. She has also been started on Tadalafil 20 mg BD for her pulmonary arterial hypertension

Prednisolone has been tapered to 15 mg and she is clinically well with a CRP of 26 mg/L and ferritin of 2600 ug/L.







Abstract AB1127 - Figure 1

**Conclusions:** We present a case of life threatening AOSD complicated by pulmonary arterial hypertension, PRES and peripheral neuropathy.

She has unusually severe disease, which is quite refractory to treatment and has been associated with rare manifestations.

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AB1128

EVALUATION OF SERUM VERSICAN LEVELS IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER (FMF)

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**Background:** Familial Mediterranean fever (FMF) is an autoinflammatory disease which has self-limiting inflammatory attacks placing in polyserositis. Versican is an extracellular proteoglycan which interacts with receptors that regulate immune system.<sup>2</sup>

**Objectives:** The aim of this study is to measure serum versican levels between FMF and control group.

**Methods:** Between June 2017 –, Semptember 2017 thirty-seven FMF patients with attack-free period that following-up at Cumhuriyet University Faculty of Medicine Department of Internal Medicine Rheumatology and thirty-five healthy volunteers without any rheumatic, systemic and metabolic diseases were enrolled in this study. Clinical findings of all patients were recorded. Blood tests were examined by Elisa method in Cumhuriyet University Department of Biochemistry.

**Results:** The median age of the FMF patients was 33(19-64) years. Of the FMF patients, twenty-one (56.8%) were female and sixteen (43.2%) were male. The median age of control group was 26<sup>18-38</sup> years. Of the control group fourteen (40%) were female and twenty-one (60%) were male. The median versican level was measured as 18.3 ng/ml in FMF group and 23 ng/ml in healthy group (p<0.05). There was no correlation between eritrosit sedimantation rate (ESR), CRP, fibrinogen, serum amyloid-A (SAA) protein other clinical manifestations, medications and versican levels (table 1).

Abstract AB1128 - Table 1. Subgroup analysis in patients with FMF.

	Serum Versican Levels ng/ml (median)	P value	
ESR>20 mm/h n=25	20.03		
ESR<20 mm/h n=2	16.5	.597	
CRP>10 mg/L n=25	19.2		
CRP<10 mg/L n=12	16.5	.713	
Fibrinogen>200 mg/dl n=9	18.2		
Fibrinogen<200 mg/dl n=28	18.7	.986	
>40 years n=15	18.2		
<40 years n=22	19.5	.591	