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.

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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.¹ Versican is an extracellular proteoglycan which interacts with receptors that regulate immune system.²

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

Methods: Between June 2017 – September 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^(18–38) 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 erythrocyte sedimentation 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

Abstract AB1128 – Figure 1. Serum Versican levels between FMF and Control groups

Conclusions: Serum versican level was statistically significant lower in FMF group than in control. On the other hand, there was no statistically significant correlation between versican levels and other parameters in FMF patients with attack-free period. Clinical trials including FMF patients with attack-period, attack-free period and amyloidosis would be designed in order to investigate the possible role of versican.

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AB1129 EPIDEMIOLOGICAL AND CLINICAL PROFILE OF PATIENTS WITH PARANEOPLASTIC RHEUMATOLOGIC SYNDROMES IN A SPANISH TERTIARY HOSPITAL

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Background: Paraneoplastic rheumatic syndromes (PRS) are defined as rheumatic symptoms caused by a malignancy, but which are not caused directly by the tumour or metastasis. PRS are characterised by a close temporal relationship with an underlying malignancy and fast regression of the symptoms after the treatment of the primary tumour.

Objectives: To describe the epidemiological and clinical profile of patients diagnosed with PRS in a Spanish tertiary hospital.

Methods: A retrospective observational study was performed, which included all patients who were diagnosed of PRS in our centre during the period 1985–2017. The demographic, clinical and outcome data were obtained from their medical records.

Results: During the study period 48 patients with suspected paraneoplastic syndrome were identified, but diagnosis was confirmed in 14 of them, 7 women (50%), with a mean age at diagnosis of 69.8±14.5 years. 42.9% of patients were smokers, 14.3% reported alcohol consumption, 35.7% and 7.1% had personal and family history of neoplasia, respectively; no patient reported personal or family history of autoimmune disease. The comparison between patients with and without PRS is shown in the table, but no significant differences were identified. The most common neoplasm was breast cancer (28.6%). 21.4% of the cases of PRS appeared as a consequence of a tumour recurrence. In 42.9% of patients the diagnostic of the tumour was concomitant to PRS, while the remaining 57.1% SPR was previous. The most frequent paraneoplastic syndrome was polymyalgia rheumatica (42.9%), followed by dermatomyositis (28.6%) and paraneoplastic polyarthritis (28.6%). The most common musculoskeletal manifestation was shoulder girdle pain and stiffness (35.7%) followed by polyarthritis and proximal

muscular weakness (28.5%). 85.7% of the patients had high erythrocyte sedimentation rate and C-reactive protein levels with a mean value of 60.2±39.4 mm/h and 77.6±9.5 mg/L, respectively. Antinuclear antibodies were positive in 7 patients (50%), while rheumatoid factor, anti-CCP and anti-RNP were positive in 1 patient each (7.1%). The response of PRS to the treatment of the tumour was available in 9 patients, of which 6 (66.7%) presented remission of the PRS. Regarding mortality, 5 patients (35.7%) died due to complications associated with the primary tumour.

Abstract AB1129 – Table 1

	PRS (n=14)	NO PRS (n=34)	P-value
Age (years)	69.78±14.57	73.23±10.81	0534
Sex (Male)	7 (50%)	14 (41,17%)	0575
Personal history of malignant neoplasm	5 (35,71%)	16 (47,05%)	0471
Personal history of autoimmune disease	0 (0%)	4 (11,76%)	0180
Family history of malignant neoplasm	1 (7,14%)	0 (0%)	0115
Family history of autoimmune disease	0 (0%)	1 (2,94%)	0115
Smoking	6 (42,86%)	13 (38,24%)	0766
Alcoholism	2 (14,29%)	4 (11,76%)	0810

Comparison of the demographic characteristics of patients with and without a diagnosis of paraneoplastic rheumatic syndrome.

Conclusions: Polymyalgia rheumatica was the most frequent PRS in our study, in contrast to several published series where other PRS are more common such as hypertrophic osteoarthropathy, polyarthritis and dermatomyositis/polymyositis. The laboratory data and response to treatment of the tumour were similar to those reported in the literature. The mortality in our series was high, but in none of the cases was related directly to PRS.

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AB1130 LATE ONSET TUMOUR NECROSIS FACTOR RECEPTOR (TNFR)–ASSOCIATED PERIODIC SYNDROME (TRAPS) CAUSED BY SOMATIC TNFRSF1 MOSAICISM

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Background: Tumour necrosis factor receptor (TNFR)–associated periodic syndrome (TRAPS) is an autosomal-dominant disease caused by gain-of-function mutations in the TNFRSF1A gene, which encodes the 55-kd TNFR type I (TNFRI) protein. Mosaicism has been identified in a single patient. A 60 year old male presented with a 6 year history of intermittent fever as high as 103.5, lasting 3–4 weeks with associated peritoneal symptoms, arthralgias, myalgias, lymphadenopathy, bilateral episcleritis, erythematous rash in his torso. Prednisone up to 60 mg daily and colchicine was ineffective where he responded fully to canakinumab at a dose of 150 mg every 4 weeks.

Objectives: To identify the extent of mosaicism in a patient with adult onset TRAPS phenotype.

Methods: DNA was extracted from the patient's whole blood, saliva and hair root. The TNFRSF1A gene was analysed by Sanger sequencing in all tissues, whole blood and fractionated cell subsets. *In silico* molecular modelling was performed to predict the structural and functional consequences of the tumour necrosis factor receptor (TNFR) type I protein mutation.

Results: Sanger sequencing revealed differential tissue and hematopoietic cell presence of a missense mutation at c.265 T>C p.Phe89Leu (F89L) Chr12 (GRCh37):g.6442960A>G ex3 rs104895245. The mutant allele was present in whole blood and buccal mucosa and absent in hair root, supporting the presence of somatic TNFRSF1A mosaicism. Additionally, quantitative analysis revealed allele frequency of 50% in B cells, 30% in NK cells and 30% in neutrophils. The mutation was absent in T cells and monocytes. *In silico* prediction modelling with SIFT and PolyPhen2 suggested that this mutation led to numerous structural rearrangements which resulted in changes in the protein surface profile.