prednisolone (30 mg/kg) for 3 days was given in 27 patients. Two patients received hydrocortisone in SJIA MAS secondary to infections (chickenpox, Hepatitis A). In patients unresponsive to steroids IVIG and/or cyclosporine was used. 19 (65%) patients survived whereas 10 (35%) died. Of the 10 who succumbed, the HLH 2004 protocol (including etoposide) was used in 2 who were refractory to pulse methylprednisolone +cyclosporine +IV Ig, but without any success. 2 patients on Tocilizumab had silent MAS.

Ferritin >50000 ng/ml was seen in 11 patients of which 8 died. Chi square test of significance was done and p value (0.001) was significant for mortality in patients with ferritin >50000 ng/ml, with a positive correlation (0.613).

Patients with cardiovascular involvement especially pericardial effusion in SJIA was observed to have higher mortality but no statistical correlation could be made. **Conclusions:** MAS is a fatal complication with a high mortality rate of 35% in our series. Ferritin levels >50,000 ng/ml was associated with high mortality. Early and aggressive intervention with optimal intensive care support maybe life saving.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3951

AB1117 ISOLATED CERVICAL ARTHRITIS AS THE SOLE MANIFESTATION OF FAMILIAL MEDITERRANEAN FEVER: A CASE REPORT

R. Dagher¹, M. Samarani¹, A. Chelala², M.C. Fadous Khalife¹. ¹pediatrics; ²radiology, Notre Dame de Secours University Hospital, Byblos, Lebanon

Background: Familial Mediterranean Fever (FMF) is an autosomal recessive disease affecting mainly eastern Mediterranean populations. Fever and Abdominal pain are the 2 most prevalent features. The most common arthritic manifestation of FMF is acute self-limiting monoarthritis. 5% of FMF patients develop chronic erosive arthritis. FMF mutation M694V has been associated to an increased risk of spondylarthropathy.

Objectives: We report the case of a child with cervical spine inflammation as the sole presentation of FMF.

Methods: A boy born to non-consanguineous parents presented at the age of 4 with complete blockage of his neck. He was described to have experienced progressive neck stiffness since the age of 3 years. His history is notable for intermittent limp at the age of 1 with complete spontaneous remission, and possible recurrent fever in the first 2 years of life. Family history is negative.

His physical exam revealed a painful and completely blocked neck in all movements, and stubby fingers. He had failure to thrive and a big belly without hepatosplenomegaly. Cognitive development was normal.

Laboratory tests revealed increased inflammatory markers. Other biological tests were insignificant. ANA, RF, anti-CCP and HLA B27 were negative. Lysosomal enzyme activities were normal ruling out mucopolysaccharidosis, mucopolidoses and multiple sulfatase deficiencies. Work-up for failure to thrive was noncontributory. Ophthalmic screening showed no abnormalities.

Cervical spine plain radiographs were normal. Cervical MRI showed global contrast enhancement of cervical vertebrae and joints with blurring of the osseous contours and synovial inflammation; bone oedema was noted and involved some posterior arcs, spinous processes and pedicles. Sacro-iliac MRI was normal.

The child was treated with oral steroids along with methotrexate and etanercept secondarily due to steroid dependence. Inflammatory markers normalised and clinical improvement was noted as the range of motion of neck increased and pain subsided. Repeated MRI after 6 months showed an almost normal image.

Results: Given this atypical isolated inflammation of the cervical spine, genetic testing for FMF was conducted. We identified 2 typical mutations (M694V and M694I) confirming the diagnosis of FMF. Colchicine treatment was started.

Abstract AB1117 – Figure 1

Conclusions: To the best of our knowledge, this is the first report of FMF masquerading as neck arthritis. Early spondylarthropathy is a possibility, but this

unusual neck inflammation might be an isolated arthritis associated to FMF. Based on this clinical presentation, in the setting of atypical arthritis, diagnosis of FMF is to be raised in at-risk ethnicities, even in the absence of familial history and common clinical signs.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2825

AB1118 PROCALCITONIN DIFFERENTIATES INFECTION FROM ACTIVE DISEASE IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA)

R. Trachtman¹, E.T. Murray², N. Pan¹, S.S. Toussi¹, M.E. Nellis³, J. Szymonińska², S.F. Taber¹, A.B. Adams¹, K.B. Onel¹, L.A. Mandl¹. ¹Hospital for Special Surgery/ Weill Cornell Medicine; ²Hospital for Special Surgery; ³Weill Cornell Medicine, New York, USA

Background: Patients with JIA often present with signs and symptoms suggestive of infection. However, differentiation of infectious from non-infectious presentation in routine clinical care is challenging. Procalcitonin (PCT) is a serum biomarker elevated in the setting of bacterial infection, but whether it can reliably differentiate infection from disease flare in patients with JIA is unknown.^[1]

Objectives: To test the hypothesis that PCT levels will differ between active JIA, quiescent JIA, bacteremic patients and healthy controls.

Abstract AB1118 – Table 1. Patient Characteristics

	Active Untreated JIA (n=12)	Quiescent JIA (n=15)	Healthy Controls (n=16)	Bacteremic Patients (n=5)
Age, years (median, IQR range)	9.0 [2.4–12.8]	14.5 [9.9–17.4]	14.4 [13.9–15.5]	1.1 [0.8–1.8]
Male Gender	5 (41.7%)	3 (20.0%)	6 (37.5%)	3 (60.0%)
Race				
Caucasian/White	7 (58.3%)	14 (93.3%)	12 (75.0%)	2 (50.0%)
AA/Black	1 (8.3%)	0 (0.0%)	3 (18.8%)	1 (25.0%)
Asian	3 (25.0%)	1 (6.7%)	0 (0.0%)	1 (25.0%)
Other	1 (8.3%)	0 (0.0%)	1 (6.3%)	0 (0.0%)
Hispanic Ethnicity	2 (16.7%)	2 (13.3%)	1 (6.3%)	1 (20.0%)
Private Insurance	7 (58.3%)	14 (93.3%)	14 (87.5%)	4 (80.0%)

Abstract AB1118 – Table 2. Laboratory Data

	Active Untreated JIA (n=12)	Quiescent JIA (n=15)	Healthy controls (n=16)	Bacteremic patients (n=5)	p-value
WBC (mean, SD)	8.9±4.4	7.7±1.6	6.7±1.7	13.1±12.1	0.06
ESR	6.0	8.0	8.0	43.0	0.18
Normal<10 (median, IQR)	[4.0–46.0]	[5.0–10.0]	[5.0–10.0]	[20.0–66.0]	
CRP	0.27	0.31	0.44	16.63	0.067
Normal<1 (median, IQR)	[0.12–6.48]	[0.16–2.65]	[0.12–1.85]	[7.76–25.68]	
PCT	0.00	0.00	0.00	5.78	<0.001
(median, IQR)	[0.00–0.00]	[0.00–0.00]	[0.00–0.00]	[4.26–52.00]	

Methods: From 10/16–4/17, consecutive children 6 months – 18 years with a) active untreated JIA b) quiescent JIA and c) healthy pre-surgical candidates were recruited from a musculoskeletal specialty hospital. JIA was defined according to ILAR criteria. Patients with active JIA despite treatment were excluded, to avoid

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.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7238

AB1119 CLINICAL, THERAPEUTIC CHARACTERISATION AND TIME TO ACHIEVE REMISSION ANALYSIS OF A COLOMBIAN COHORT WITH JUVENILE IDIOPATHIC MYOPATHY

R. Fuentes¹, M. Cañola¹, J.C. Diaz-Coronado², A. Rojas-Villarraga², D. Hernandez-Parra², P. Perez-Estrada², J.C. Salazar-Urbe³, R. Pineda-Tamayo².
¹Rheumatology Department; ²Clinical Research, Artmedica; ³National University of Colombia, Medellin, Colombia

Background: The clinical characteristics of paediatric patients with idiopathic inflammatory myopathies differ from adults in several aspects. Its clinical presentation can include amyopathic onset and the skin involvement has different characteristics.

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 analyze 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%,²⁸ 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%), Heliotope 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 methotrexate (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 remission in patients with Gottron sign compared to 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.

Characteristics in JM n=37	N	%
Symmetrical muscle weakness	27	72.97
Gottron's papules	33	89.19
Heliotope 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(+)	14/27	37.84
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.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3686

AB1120 THYROID HORMONE CONCENTRATIONS IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS FROM A SINGLE TERTIARY REFERRAL CENTRE

B. Eremciuc^{1,2}, N. Revenco^{1,2}. ¹Paediatric Department, State University of Medicine and Pharmacy "Nicolae Testemitanu"; ²Paediatric Rheumatologic Unit, Scientific Research Institute for Mother and Child Health Care, Chisinau, Moldova, Republic of

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.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7293

AB1121 EVALUATION OF CASES DIAGNOSED WITH CRMO; SINGLE CENTRE EXPERIENCE

S. Cekic, Y. Karali, S.S. Kilic. *Uludag University, Bursa, Turkey*

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