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

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

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

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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