Conclusions: One of the most common complaints seen during the childhood is musculoskeletal system pain. As shown by various studies performed, one of the significant reasons of extremity pain is GJH.¹ In our study, no significant correlation was found between GJH and joint pain. GJH is a disease that may cause musculoskeletal system pain during childhood. In our study which investigates the frequency of GJH in our region, we detected the GJH prevalence as 8.7. GJH is a clinical syndrome that is characterised with the fact that the joints have a range of motion above normal levels without a correlation with any systemic rheumatismal disease. The specific definition of GJH was shown by Kirk et al. in 1967.²

REFERENCES:

- [1] Larsson LG, Baum J, Mudholkar GS. Hypermobility: Features and differential incidence between the sexes. *Arthritis Rheum* 1987; 30: 1426–30.
- [2] Kirk JA, Ansell BM, Bywaters EG. The hypermobility syndrome. Musculoskeletal complaints associated with generalized joint hypermobility. *Ann Rheum Dis* 1967; 26: 419.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.1511

AB1114 SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS: A SINGLE CENTRE EXPERIENCE

K. Barut, G. Tarcin, G. Tahaoglu, S. Sahin, A. Adrovic, O. Kasapcopur. *Pediatric Rheumatology, Istanbul University, Cerrahpasa Medical School, Istanbul, Turkey*

Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood, divided into several subgroups. The sJIA could be presented by monocyclic, polycyclic or persistent polyarticular clinical course. Macrophage activation syndrome (MAS) represents the most devastating complication that could appear during the disease course. Studies on follow up, treatment response and disease complications of the sJIA patients are sparse and rare.

Objectives: To evaluate demographic and clinical characteristics and to explore the long-term treatment response and disease complications in a large cohort of sJIA patients from the single centre.

Methods: Demographic and clinical features of the sJIA patients were reached from the patients's archives. The frequency of disease flares, treatment response and side effects were recorded for each patient.

Results: A total of 168 sJIA patients were included in the study: 87 (51.8) female, 81 (48.2) male. The clinical features are shown in table 1. Fifty-three (31.5) patients had monocyclic while 23 (13.7) patients had polycyclic clinical course (mean recurrence of attacks 2.5 ± 2 (IQR:1–4)); in 38 (42 Polyarticular course was present in 92 (54.8) patients. Initially diagnosis of patients were: infection in 86 (51.1), sJIA in 34 (20.4), acute rheumatic fever in 19 (11.3), urticaria in 10 (5.9), Kawasaki disease in 4 (2.4) and juvenile systemic lupus erythematosus in 2 patients.

The most common disease complications were: MAS in 20 (11.9), growth retardation in 19 (11.3) and vertebral fracture due to osteoporosis in 3 (1.9) patients. Gastrointestinal symptoms secondary to methotrexate intolerance that led to cessation of treatment were present in 9 (7.1) patients. Among 5 (2.9) patients that developed tuberculosis, 4 (2.3) were under etanercept treatment.

All of the patients were treated with corticosteroids: a doses of 2 mg/kg/day in 118 (70.2) patients and pulse steroids in 50 (29.8) patients with severe clinical presentation. The methotrexate was used in 126 (75), leflunomide in 5 (3), cyclosporine A in 29 (17.3), intravenous immunoglobulin in 19 (11.3), anakinra in 27 (16.1), canakinumab in 27 (16.1), tocilizumab in 18 (10.7), etanercept in 50 (29.8) and adalimumab in 7 (4.2) patients. The median time to remission after the initial treatment with corticosteroids was 4 (IQR:2–4) months. The remission off medications was achieved in 82 (48.8) while remission on medications was achieved in 83 (49.4) of patients.

Table: Demographic, clinical features of sJIA

Female/male	87 (51.8)/81 (48.2)
Mean age at disease onset	76.7±54.5 months (IQR: 28–118)
Mean age at diagnosis	79.7±54.5 months (IQR: 33–121)
Clinical features, n(%)	
– Typical fever	160 (95.2)
– Typical rash	99(59)
– Lymphadenopathy	45 (26.8)
– Hepatosplenomegaly	70 (41.7)
– Arthritis/arthralgia	143 (85.1), 25 (14.9)

Conclusions: Systemic JIA is a subtype of JIA characterised by significant morbidity and mortality rate with macrophage activation syndrome being the most severe disease complication. Corticosteroids represent the main treatment modality. Biological agents should be considered in the steroid-resistant patients.

The clinical remission could be achieved and chronic arthritis sequelae could be prevented in a majority of patients with biological agents.

REFERENCE:

- [1] Barut K, Adrovic A, Sahin S, Kasapcopur O. Juvenile Idiopathic Arthritis. *Balkan medical journal*. 2017;34:90–101

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3594

AB1115 SYSTEMIC LUPUS ERYTHEMATOSUS IN PAEDIATRIC POPULATION: A SINGLE CENTRE STUDY FROM INDIA

P.P. Giri¹, P. Pal². ¹pediatrics; ²ich, kolkata, India

Background: systemic lupus erythematosus is an autoimmune disease that can be manifest in paediatric population in various ways. It is characterised by widespread inflammation of the blood vessels and connective tissues with positive autoantibodies. Though it is a chronic disease it can be fatal at times.

Objectives: 1. To study the diversity in clinical and laboratory profile in paediatric systemic lupus erythematosus patients at a tertiary care centre in Kolkata.

2. To identify the poor prognostic factors at the time of admission to the hospital.

3. To quantify the drug related adverse effects in follow up.

Methods: Both old known cases and newly diagnosed cases of paediatric SLE who presented to our rheumatology follow up clinic over last 18 months were retrospectively reviewed for their clinical and immunological presentation. SLICC diagnostic criteria has been applied to define a positive case.

Results: A total number of 64 patients were evaluated, among which 54 were girls and 10 were boys with a sex ratio of 5.4:1 favouring girls. Mean age on presentation was 9.9 years with a range of 2.5 to 16 years. Among the clinical presentation fever (72%) was the most common symptom, followed by skin manifestation (68.8%), musculoskeletal involvement (53.1%), haematological involvement (37.5%). Renal involvement was seen in 35.9%, among which 59.1% had stage IV lupus nephritis, and central nervous system involvement was observed in 10.9%. Among immunological profile, ANA was positive in 95.3%, anti-double-stranded DNA was positive in 92.1% and low complement levels were seen in 92.1%. Antiphospholipid antibody was seen in 7

patients (n=21) and anti-Smith antibody in 3 (n=4). All the patients required therapy with steroids and hydroxychloroquine. Steroid sparing agents like azathioprine (54.7%), cyclophosphamide (28.1%), mycophenolate mofetil (23.4%), methotrexate (18.7%), and rituximab (10.9%) were also used.

Conclusions: Paediatric SLE has got a varied presentation, and a high index of suspicion is needed for early diagnosis and timely management with multiple drugs of this dreadful disease

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3921

AB1116 MACROPHAGE ACTIVATION SYNDROME: AN EXPERIENCE FROM A TERTIARY PAEDIATRIC CARE SETTING IN EASTERN INDIA

P.P. Giri, P. PAL, J. bhatia. *pediatrics, institute of child health, Kolkata, India*

Background: Macrophage activation syndrome (MAS) is a rare but potentially fatal complication of systemic inflammatory disorders occurring most commonly in Systemic arthritis (SJIA) but also being increasingly reported in SLE, Kawasaki disease and Periodic Fever Syndromes. We present a series of 29 cases of MAS encountered in the last 9 years in a tertiary paediatric care setting in eastern India.

Objectives: The objective of this study is to evaluate the clinical features, laboratory findings and outcome in MAS; to assess the treatment response to different therapies and to identify the poor prognostic factors.

Methods: It is a prospective analysis of data of patients diagnosed as having MAS, between July 2008 and April 2017, admitted in the Department of Paediatrics at Institute Of Child Health, Kolkata.

All patients with Haemophagocytic Lymphohistiocytosis (HLH) secondary to autoimmune or inflammatory connective tissue diseases were included whereas HLH secondary to infections were excluded. Diagnosis of HLH was based on the HLH 2004 criteria. MAS diagnostic criteria for SJIA was laid down in 2014. Post 2014 we used those criteria in SJIA patients.

The data noted were the clinical and laboratory features, treatment details, the response to therapy and outcome.

Results: Twenty nine (n=29) patients were found to have MAS with the primary illness being SJIA in 24 (83%), SLE in 4 (14%) and Kawasaki Disease in 1 (3%). The mean age at presentation was 5 years 3 months. The male female ratio was 1.2:1. Neurological, cardiac, renal and pulmonary involvement was seen in 21 (72%), 14 (48%), 6 (20%) and 5 (17%) patients respectively. Pulse methyl

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.

REFERENCES:

- [1] Sönmez HE, Batu ED, Demir S, Bilginer Y, Özen S. Comparison of patients with familial Mediterranean fever accompanied with sacroiliitis and patients with juvenile spondyloarthritis. Clinical and Experimental Rheumatology. 2017 Nov-Dec;35 Suppl 108(6):124–127
- [2] Younes M, Kahn MF, Meyer O. Hip involvement in patients with familial Mediterranean fever. A review of ten cases. Joint Bone Spine. 2002 Dec;69(6):560–5.
- [3] Güncan S, Bilge NŞY, Cansu DÜ, Kaşifoğlu T, Korkmaz C. The role of MEFV mutations in the concurrent disorders observed in patients with familial Mediterranean fever. European Journal of Rheumatology. 2016;3(3):118–121.

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