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 $\pm$ 54.5 months (IQR: 28–118)
Mean age at diagnosis	79.7 $\pm$ 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