condition with high mortality rate. The rate of occurrence of the condition in AOSD patients is 5-10%.
Objectives: A 24-year-old female patient with fever, rash, joint pain having started 1.5 months ago was admitted to our hospital. In physical examination; She had hepatosplenomegaly and multiple LAPs in bilateral cervical region as well as tenderness on the left wrist and right ankle. The rash was concomittant with fever. CRP, erythrocyte sedimentation rate, white blood cell and ferritin elevation were determined. Hepatitis markers, HIV, brucella, syphilis, CMV, toxoplasma, EBV were found to be negative. The anti-nuclear antibodies, rheumatoid factor and anti-CCP were negative,
Methods: In the thorax CT, multiple lymph nodes were observed in the mediastinal (13 mm), bilateral hilar, axillary (15 mm largest) and bilateral lower neck. The patient's PET CT revealed increased metabolic rate in the neck, mediastinum, abdominopelvic region, mildly diffused increased metabolism in the bone marrow, and increased metabolism in the basal sections of both lungs. It was reported that lymphoproliferative disease should be included in the differential diagnosis. Right servical lymph node excisional biopsy was reported as reactive lymphoid hyperplasia. Bone marrow biopsy came as normocellular. Her bronchoscopy was normal. Thereforemalignancy and infection were excluded. Prednizolon 60 mg/d and methotrexate 15 mg/w were started.
Results: The patient developed pancytopenia after two weeks and exhibited an increase in her triglyceride, D-dimer, LDH and hypofibrinogenemia developed. The patient had no atypical cells in the peripheral smear. IVIG as 2g/kg was added to the treatment with the diagnosis of haemophagocytic syndrome. After IVIG, her fever reduced, leukopenia thrombocytopenia improved, her sedimentation CRP reached normal limits. IVIG was administered 3 times with an interval of four weeks. However, one month after, she was admitted to our clinic with common myalgia, subfebrile fever, sore throat, swelling of the elbow, rash on the back. Leukocytosis with neutrophil dominance, increased levels of ferritin, LDH, eryhtrocyte sedimentation rate and CRP were noted. Her methylprednisolone dose was increased and Tocilizumab treatment (8 mg/kg/4w) was begun. After three months, clinical and laboratory findings were normal.
Conclusion: Macrophage activation syndrome is a reactive form of haemophagocytic syndrome associated with rheumatic diseases. 71% of macrophage activation syndrome cases occur within the first month of AOSD diagnosis. Fever, cytopenia, hypertriglyceridemia, increased ferritin, LDH and liver enzymes, HSM, LAP,coagulopathy and elevation of fibrin degradation products are observed. The syndrome is characterized with haemophagocytosis in bone marrow and RES. Glucocorticoids, cyclosporine, anakinra, tocilizumab and IVIG are administered.

Consent for publication has been obtained from patient
Yes

Disclosure of Interest
None Declared

P1105

Drug retention rate and predictive factors of drug survival for interleukin-1 inhibitors in systemic juvenile idiopathic arthritis

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Introduction: The advent of biologic agents has revolutionized therapeutic approaches in systemic juvenile idiopatic arthritis (sJIA) as their introduction has been shown to modify disease course and improve overall outcomes, particularly when initiated early. Few studies have reported the drug retention rate (DRR) of biologic drugs in JIA, and none of them has specifically investigated the DRR of interleukin (IL)-1 inhibitors on sJIA.
Objectives: The primary aim of the study was to examine the overall DRR of IL-1 blockers in sJIA patients. Secondary aims of our study were to: (i) explore the influence of biologic line of treatment, adverse events (AEs), type of anti-IL-1 agent and the concomitant use of conventional disease modifying anti-rheumatic drugs (cDMARDs) on DRR; (ii) find eventual predictive factors associated with events leading to drug discontinuation. The corticosteroid sparing effect and the impact of disease duration and treatment delay on survival constituted ancillary aims.
Methods: sJIA patients – diagnosed according to the revised International League of Association for Rheumatology (ILAR) criteria – treated with anakinra (ANA) and canakinumab (CAN) were enrolled in 15 Italian tertiary referral centers. Demographic, clinical and

therapeutic data collected from medical records were retrospectively collected and statistically analyzed.
Results: Seventy seven patients were enrolled for a total of 86 treatment courses. The cumulative retention rate of the IL-1 inhibitors at 12-, 24-, 48-, and 60-months of follow-up was 79.9, 59.5, 53.5, and 53.5%, respectively, without any statistically significant differences between ANA and CAN (p = 0.056), and between patients treated in monotherapy compared to the subgroup co-administered with conventional immunosuppressors (p = 0.058). On the contrary, significant differences were found between biologic-naive patients and those previously treated with biologic drugs (p = 0.038) and when distinguishing according to AEs occurrence (p = 0.04). In regression analysis, patients pre-treated with other biologics (HR = 3.357 [CI: 1.341–8.406], p = 0.01) and those experiencing AEs (HR = 2.970 [CI: 1.186–7.435], p = 0.020) were associated with a higher hazard ratio of IL-1 inhibitors withdrawal. The mean treatment delay was significantly higher among patients discontinuing IL-1 inhibitors (p = 0.0002).
Conclusion: Our findings suggest an excellent overall DRR for both ANA and CAN that might be further augmented by paying attention to AEs and employing these agents as first-line biologics in an early disease phase.

Disclosure of Interest
None Declared

P1106
Clinic-laboratory profile of macrophage activation syndrome in children with systemic juvenile idiopathic arthritis
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Introduction: Systemic juvenile idiopathic arthritis (SJIA) is an autoinflammatory disorder with a propensity to develop macrophage activation syndrome (MAS). MAS is an emergent, fulminant, life-threatening condition which can be fatal if not recognized early.
Objectives: To study the clinic-laboratory profile of MAS in children with SJIA
Methods: Case records of children of SJIA who had MAS, diagnosed at the Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, during the period 2017-2018, were reviewed. Findings pertaining to history, clinical examination and laboratory investigations were recorded. Diagnosis of MAS was based on criteria given by Ravelli et al (2016).
Results: A total of 13 children (1-18 years) with 15 MAS events were included in the study. Two children had 2 episodes each of MAS during this period. Mean age was 6.7+ 4.8 years with male to female ratio of 1.17. Five children (38.5%) had MAS, at or during hospital admission on their first presentation. Common clinical features noted were fever (100%), pallor (100%), hepatomegaly and/or splenomegaly (73.3%), rash (46.7%), lymphadenopathy (46.7%), arthritis (33.3%), irritability (26.7%), mucocutaneous bleeding (20%) and jaundice (13.3%). In laboratory investigations (Table 1), all children had anaemia and elevated C-reactive protein (CRP). Leucopenia and leucocytosis was seen in 2 (13.3%) and 8 (53.5%) children respectively. Four (26.6%) children had thrombocytopenia (< 150 x 10^9/L), 6 (40%) had platelet count < 181 x 10^9/L and three children (20%) had thrombocytosis. Hypertriglyceridemia (93.3%), hypofibrinoginemia (73.3%), elevated aspartate aminotransferase (73.3%) and alanine aminotransferase (40%) were other prominent laboratory parameters. All children had high serum ferritin levels with mean of 8960.6 ± 13662.12 ng/ml. Nine children had serum ferritin/ESR (ratio) of > 80. Three (20%) children died. Two children, who died, had very high serum ferritin levels (52000 ng/ml and 42845 ng/ml).
Conclusion: MAS is a life threatening multisystemic complication of SJIA with high mortality rate (20%). Fever, pallor, organomegaly and persistent rash are prominent clinical features of MAS. Leucopenia and thrombocytopenia may not be early features of MAS but low or normal platelets can differentiate MAS from a disease flare in which we almost always get thrombocytosis. Very high serum ferritin, Hypertriglyceridemia, hypofibroginemia and transaminitis are better clue for MAS and serum ferritin levels may be a prognostic marker.

Disclosure of Interest
None Declared

Table 1 (abstract P1106). Laboratory parameters in children of SJIA with MAS

Variables	No. of patients	Mean ± SD (n=15)
Anaemia	15 (100%)	84 ± 19 g/L
Leucopenia	2 (13.3 %)	14.2 ± 11.7 x 10^9/L
Elevated CRP	15 (100%)	99.5 ± 63.3 mg/L
Platelet < 181 x 10^9/L	6 (40%)	283.5 ± 182.4 x 10^9/L
Elevated AST	11 (73.3%)	136.3 ± 137.3 IU/L
Elevated ALT	6 (40%)	109.6 ± 101.3 IU/L
Hypofibrinogenemia (<3.6 g/L)	11 (73.3%)	323 ± 189 mg/dl
Hypertriglyceridemia(>156 mg/dl)	14 (93.3%)	235.9 ± 132 mg/dl
Hyperferritinemia (ng/ml)	15 (100%)	8960.6 ± 13662.12

ALT, alanine aminotransferase; AST, aspartate aminotransferase;CRP, C-reactive protein; ESR, erythrocyte sedimentation rate

P1107
Serum soluble CD25: an useful biomarker of macrophage activation syndrome in systemic juvenile idiopathic arthritis
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Introduction: Systemic juvenile idiopathic arthritis (SJIA) is an autoinflammatory disorder secondary to innate immune dysfunction with a propensity to develop macrophage activation syndrome (MAS), a life-threatening condition. sCD25 has been used as a sensitive biomarker for the diagnosis of Hemophagocytic lymphohistiocytosis which has similarities in clinical features and pathogenesis to MAS.
Objectives: To assay serum soluble CD25 in children with systemic juvenile idiopathic arthritis (SJIA) and to compare levels of sCD25 in children with inactive disease, active disease and those with macrophage activation syndrome (MAS).
Methods: Thisprospective study was conducted in a tertiary care referral centre in North India from January 2017 to June 2018. All patients fulfilling the International League of Associations for Rheumatology (ILAR) 2001 criteria for SJIA were eligible for enrolment. At enrolment, all patients were examined clinically for signs of disease activity. Appropriate investigations were carried out and sCD25 was analyzed by using commercially available sCD25 / IL-2R ELISA kit.
Results: A total of 35 children (1-18 years) with 43 events were included in the study. Mean age at enrolment in the study was 7.3 ±3.59 years with male to female ratio of 2.5. Based on clinical features and investigations, events were categorized into 3 groups; SJIA with inactive disease (15; 34.9%), SJIA with active disease (15; 34.9%) and SJIA with MAS (13; 30.2%). Mean sCD25 levels in the study population were 10,966.02 ± 10,854.93 pg/ml. Children with inactive disease, active disease and disease with MAS had mean ± SD serum