present in 35 (17.5%) of children. These patients were determined to have significantly higher rate of persistent purpura (45.7% vs 20.7%), severe renal involvement (57.7% vs 30.8%), hypertension (28.6% vs 9.1%) and increased erythrocyte sedimentation rate (79.2% vs 55.9%). Obese patients also showed delayed improvement of skin (25 vs 14 days), joint (12.5 vs 10.0 days) and renal (280 vs 57 days) symptoms. While the rate of children requiring steroid treatment was not different between the two groups, obese children used steroids for significantly longer time (236 vs 40 days). Furthermore, need for immunosuppressive medications (azathioprine and cyclophosphamide) were higher in obese patients (40.0% vs 8.6%).

Conclusion: Obese children with HSP seem to have higher ESR, hypertension and severe renal involvement; show delayed improvement of skin, joint and renal findings; need more immunosuppressive medications and longer period of immunosuppressive treatment. These findings may be associated with the effect of adipose tissue on inflammation.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.3265

THU0524 ARE CHILDREN AND ADULTS HAVING DIFFERENT PHENOTYPE AND GENOTYPE OF FMF?

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Background: Familial Mediterranean fever (FMF) is an autosomal recessively inherited autoinflammatory disease and begins in childhood. In nearly 60% of patients, the first attack occurs before the age of 10, and in 90% of them before the age of 20 years. There isn't a prospective study designed for comparing childhood onset and adult onset FMF.

Objectives: To compare the demographic data, clinical features, genetic analysis, laboratory values and severity scores of both childhood and adult onset FMF. Compliance and resistance to colchicine, presence of accompanying diseases and complications due to FMF were also analyzed and reviewed.

Methods: The patients were divided into two groups; group I: children with FMF (symptoms begin before 18 years of age) and group II: adults with FMF (symptoms begin after 18 years of age). A questionnaire for collecting age at disease onset, sex, age at diagnosis, delay at diagnosis and duration of the disease, family history of FMF, consanguinity and accompanying diseases were filled. The questions were asked by an adult and pediatric rheumatologist to both groups by face to face interviews. Genetic analysis results and treatment protocols were taken from patient's charts. Laboratory data concerning complete blood count, ratio of urine protein to creatinine, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), serum amyloid A (SAA) levels were obtained during their routine follow-up at attack-free period.

Results: There were 178 (60.3%) children with the diagnosis of FMF; 73 female and 104 male and 117 (39.7%) adults diagnosed as FMF after 18 years of age; 69 female and 48 male. The mean±SD age at symptom onset, at the diagnosis and current age was 4.92±3.03, 6.45±3.61, 11.59±4.38 for group I and was 18.35±10.34, 32.23±11.62, 38±11.64 for group II, respectively. Consanguinity was significantly more frequent among children with FMF (36.1%). A positive family history of FMF was

similarly present in 102 (57.6%) of group I and 66 (56.4%) of group II. Twelve (10.2%) adult patient have FMF at their children. Family history of amyloidosis was equally distributed between groups; 6.2% in group I and 6.8% in group II. The median number of FMF attacks per year was 18 in children and 15 in adults. While children were having significantly more frequent attacks, the duration of attacks were longer in adults compared to children (p<0.001). The most frequent symptom was fever (91.5%) in children and abdominal pain (96.5%) in adults. Arthritis, chest pain and erysipelas like rash were seen similarly in both cohorts. Sacroileitis (10.2%), amyloidosis (5.1%) and chronic renal insufficiency (5.1%) were significantly more common in patients with FMF diagnosed after 18 years of age. Colchicine resistance and compliance were evenly distributed among groups. In genotypic evaluation, groups were compared for homozygotes, compound heterozygotes and heterozygotes and had put forth similar results. Attack free acute phase reactants like CRP; median 2.8 mg/dl and SAA; median 3.86 mg/dl were significantly higher in group I.

Conclusion: Both clinical features and acute phase response in FMF were less pronounced in patients diagnosed at adulthood. Children were having more frequent attacks, but accompanying diseases were more common in adults. While following patients taking age of the patient in to account will help to better understand the disease course.

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Acknowledgement: None

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7202

THU0524B EXTRA-OCULAR MANIFESTATIONS OF CHILDREN WITH SARCROID-LIKE UVEITIS

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Background: Paediatric sarcoidosis may represent a spectrum of disease. Early onset sarcoidosis & its genetically defined counterpart Blau syndrome associated with NOD2 mutations are characterized by fever, rash, arthritis and organomegaly. Later onset sarcoidosis has wider organ involvement including lungs, kidneys, lachrymal and extra-ocular glands. Both presentations may lead to long term complications due to end-stage organ damage. Although similar disease manifestations can be seen in adults and children, some entities are essentially pediatric at onset, namely Blau syndrome and most forms of immunodeficiency associated granulomatous diseases. Ocular sarcoidosis has a well described uveitis phenotype (1). We created a retrospective cohort of patients currently followed at GOSH with likely ocular manifestation of sarcoidosis (1)

Objectives: To describe a retrospective cohort of children with sarcoid-like uveitis, To describe their extra-ocular manifestations at the time of the study, To describe their management

Methods: retrospective case review of children currently followed at GOSH with a phenotype of ocular sarcoidosis with uveitis (ophthalmologist defn (5, 6)), or a diagnosis of idiopathic uveitis (anterior, posterior, intermediate or panuveitis) with raised ACE level at least once. We collected demographics and all extra-ocular involvement described in sarcoidosis

Results: 52 patients. 27/52 males. Median age onset of uveitis 4.20 yrs (1.41-15.16). 30/52 had onset <8 yrs. Median ACE 73 and mean ACE 77 at presentation (0-90U/L normal). Median max ACE 79 & mean max ACE 88.2 (14-420). NOD2 done in 12 patients: 6+, 6-. Only 4/6 patients Blau phenotype.

Ethnicity: black 13, asian 10, caucasian 13, unknown 16/52. 49/52 patients had bilateral uveitis. Uveitis anterior 17/52, intermediate 5/52, posterior 2/52, panuveitis 25/52, unknown 1/52. ANA+15/47(32%).

Extra-ocular (n=52): Lymphadenopathy: 15 (29%) clinically Liver: 16 (31%) - transaminitis 9, ultrasound(USS) abnormality (hepatomegaly, calcification, increased echogenicity) 5 & both abnormal USS & blood 2 Arthritis: 15 (29%) large joints of lower limbs & joints hands. Tenosynovitis 4 (7.7%) Renal: 15 (29%) (USS or Blood/urine tests – raised serum creatinine,