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

Nuray Aktay Ayaz¹, Cemal Bes², Hafize Emine Sonmez¹, Ayşe Tanatar³, Şerife Gül Karadağ¹, Nilüfer Alpay Kanitez², Selda Çelik², Ozan Cemal İçağan², Mustafa Çakan¹, Figen Çakmak¹. ¹University of Health Science, Kanuni Sultan Süleyman Research and Training Hospital, Pediatric Rheumatology, İstanbul, Turkey; ²University of Health Sciences, Bakırköy Sadi Konuk Research and Training Hospital, Rheumatology, İstanbul, Turkey; ³Istanbul University Faculty of Medicine, Pediatric Rheumatology, İstanbul, Turkey

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

Abhay Shivpuri¹, Emily Kalms¹, Ameenat Lola Solebo², Harry Petrushkin², Dhanes Thomas³, Elizabeth Graham², Clive Edelsten², Sandrine Compeyrot-Lacassagne¹. ¹Great Ormond Street Hospital, Paediatric Rheumatology, London, United Kingdom; ²Great Ormond Street Hospital, Ophthalmology, London, United Kingdom; ³Moorfields Eye Hospital, Ophthalmology, London, United Kingdom

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,

raised urinary ca creat ratio, raised urinary NAG/RBP — 4/15 on USS and 9/15 in blood/urine tests, 2/15 on both USS and blood/urine tests. **Lungs:** 7 (13.5%): 6 abnormal lung function test, 1 abnormal CXR, lung function & CT chest - severe interstitial lung disease & presented- progressive shortness of breath. **Skin:** 10 (19.2%), eczema-like/non-specific skin **Parotid/glandular:** 1 (1.9%) on USS **Splenomegaly:** 4 (7.7%) on USS **Sensorineural hearing loss:** 2 (3.8%)

Medications to treat uveitis and/or extra-ocular manifestations were: Methotrexate 13 (25%), Methotrexate + Adalimumab 11 (21.2%), MMF 6 (11.5%), topical steroids 8 (15.4%), systemic steroids 2 (3.8%). 10 patients no medication

Conclusion: Most of the sarcoid-like uveitis patients had at least one extra-ocular involvement. ACE does not appear to be a sensitive biomarker. Some patients (19.2%) have a mild phenotype and require no treatment. Our data demonstrate the importance of close monitoring for extra-ocular manifestations and highlight good clinical response to steroids, MTX, MMF and anti-TNF.

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THU0525

DEVELOPMENT OF A PREDICTIVE TOOL FOR RESPONSE TO ANTI-TNF-ALPHA THERAPY IN JIA USING GENE EXPRESSION PROFILES IN PERIPHERAL DERIVED MONONUCLEAR CELLS

Jedidja Baali¹, Rianne Scholman¹, Rae Yeung², Trang Duong², Nico Wulffraat¹, Joost F. Swart¹, Sebastian Vaster¹, Sytze De Rook¹. ¹UMC Utrecht, Department of Pediatric Rheumatology, Utrecht, Netherlands; ²Hospital for Sick Children, Department of Paediatrics, Toronto, Canada

Background: Heterogeneity in response to biological medication is a major challenge in the management of Juvenile Idiopathic Arthritis (JIA) (1). Biomarkers such as cytokines or whole blood RNA expression profiles that predict therapy efficacy prior to treatment pose a solution and could enable precision medicine. It was previously demonstrated that whole blood gene expression profiles can predict response to anti-TNF- α therapy in patients with JIA (2). These findings currently await validation in a multicenter prospective cohort study. Harmonization of data from different cohorts, however, is often hampered by site-dependent differences in biological sample collection and processing (3). For instance, the biological material that is retrospectively available from biobanks depends on what is stored exactly, e.g. whole blood versus peripheral blood mononuclear cells (PBMC). Furthermore, different blood collection tubes containing different additives can affect sample quality at an early stage and make interchangeability or comparability of data impossible.

Objectives: We evaluated the comparability of gene expression profiles when whole blood from patients with JIA is collected in different RNA collection tubes and processed accordingly. Next, we will investigate the predictive capacity for therapy response of RNA expression profiles from frozen PBMC of 63 JIA patients.

Methods: Peripheral blood from 11 children with non-systemic JIA with active disease was collected in PAXgene (PreAnalytiX) and Tempus (Applied Biosystems) tubes. All tubes were subsequently stored in the freezer at -80 °C. After thawing, RNA from the PAXgene tubes was extracted with use of the PAXgene Blood RNA Kit and RNA from the Tempus tubes was isolated by using the Tempus Preserved Blood RNA Purification Kit I. Gene expression of *CSNK1D*, *C1D*, *ASAP2*, *SRRD*, *PPP1R3B*, *HLA-DQA1*, *PDZK1IP1* and *MZB1* was determined with qPCR. For our next experiment, frozen PBMC derived from patients with non-systemic JIA who are included in our longitudinal Pharmachild registry biobank will be used. Of the 63 patients, 28 (44.4%) children were diagnosed with oligo articular JIA, 19 (30.2%) with poly articular JIA, 9 (14.3%) with enthesitis-related JIA, 4 (6.3%) with psoriatic JIA and 3 (4.8%) with undifferentiated JIA. The average age at sampling was 11.5

years $\pm$ 4.0 and 65% of the patients was female. These samples will be run on a 96-analyte NanoString panel and associated with clinical response at time points 3 and 6 months after start of TNF- α blockade.

Results: Both RNA blood collection systems yielded high-quality RNA, with overall higher RNA concentrations for blood collected in Tempus tubes in comparison to PAXgene tubes. qPCR data showed that gene expression of all measured genes is affected by the method of blood collection and processing. However, the inter-individual variation was similar between both collection tubes, indicating that similar RNA profiles were observed.

Conclusion: Gene expression profiles derived from whole blood collected in PAXgene or Tempus tubes are comparable, but are not interchangeable.

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THU0526

LONG-TERM OUTCOME IN JUVENILE IDIOPATHIC ARTHRITIS – A POPULATION-BASED STUDY FROM SWEDEN

Elisabet Berthold¹, Bengt Månsson¹, Robin Kahn². ¹Clinical science Lund, Lund University, Department of Rheumatology, Lund, Sweden; ²Clinical science Lund, Lund University, Department of Pediatrics, Lund, Sweden

Background: Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in children. The incidence rate among Caucasians is reported to be 8-15/100 000/year (1, 2). The long-term prognosis is insufficiently studied in JIA, although it is known that the disease often proceeds into adulthood and may cause joint damage leading to significant morbidity and physical disability. There are many challenges in elucidating the long-term prognosis. Amongst others, frequent occurrence of disease misclassification in diagnostic registers and the heterogeneity of JIA have made it difficult to interpret results (3, 4).

Objectives: To study the epidemiology, incidence and long-term outcome of JIA in southern Sweden using a population-based cohort of children with a validated diagnosis of JIA collected over nine years.

Methods: Potential cases of JIA, diagnosed between 2002 and 2010 were collected from a local search at the Department of Rheumatology in Lund and at the National Board for Health and Welfare, using the ICD-codes M08-M09. The study area is Skåne, the southernmost county of Sweden (population 1.24 million; 19.1% aged < 16 years) and the median follow-up time was eight years. The JIA-diagnosis was validated and subcategorised through medical record review based on criteria defined by The International League of Associations for Rheumatism. Parameters on disease activity and pharmacologic treatment were recorded annually until the end of the study period.

Results: 251 cases of JIA were confirmed. The mean annual incidence rate for JIA was estimated to be 12.8/100 000/year. The highest age specific annual incidence is at the age of two years (36/100 000/year). This peak is consistent in the female group, but the incidence peak among males is at twelve years of age. Almost all patients are at some point during their disease course prescribed non-steroidal anti-inflammatory drugs (98%). Intra-articular steroid injections are also frequently used (78.9% in the total cohort). Methotrexate is the most common disease modifying anti-rheumatic drug prescribed (60.6%). Tumor necrosis factor alpha-inhibitors are used as treatment in 23.9% of the children.

Oligoarthritis was the largest subgroup (44.7%), followed by undifferentiated JIA (16.3%), polyarticular rheumatoid factor negative JIA (13.9%), enthesitis-related arthritis (8.8%), polyarticular rheumatoid factor positive JIA (6.8%), juvenile psoriatic arthritis (6.8%) and systemic JIA as the smallest subgroup (2.8%).