challenging in the newborn. To our knowledge, interstitial lung involvement has never been described as onset feature in MKD.

Objectives: We report the case of a newborn affected by MKD characterized by interstitial lung disease.

Methods: The patient underwent laboratory and radiology evaluation as clinically indicated. Direct Sanger sequencing was used to screen the 10 exons of the MVK gene.

Results: A female neonate born at term from consanguineous parents was referred to our hospital at 16 days of life (DOL) for mild hypotonia and persistent raised inflammatory markers despite antibiotic therapy. Infectious work-up was negative. Chest x-ray revealed bilateral perihilar peribronchial thickening. Electroencephalography reported moderate diffuse anomalies of background activity without major abnormalities. On DOL 20 the first episode of fever was recorded. Due to worsening tachypnea and persistent abnormal chest x-ray, a pulmonary CT scan was performed and showed diffuse groundglass bilateral infiltrates consistent with alveolar-interstitial lung disease. On DOL 22 a maculo-papular skin rash appeared on feet and hands, vanishing spontaneously 24 hours later. Bone marrow examination and levels of perforins, neuron-specific enolase and urinary catabolites of catecholamines were normal. A total body MRI was normal except for a mild cerebellar hypoplasia and the known interstitial lung disease. The patient kept presenting hypotonia, relapsing episodes of fever and skin rashes, developed severe anemia and failure to thrive. Type-I IFN signature was negative. A genetic test was requested, as well as quantification of urinary levels of mevalonic acid, which were markedly elevated. Direct Sanger sequencing allowed to detect a homozygous c.709A>T missense mutation in the exon 8 of the MVK gene, coding for a protein substitution p.T237S, classified as pathogenic in the INFEVERS database and therefore consistent with the diagnosis of MKD. Both parents and her sister were heterozygous carriers of the same mutation. On DOL 38 treatment with anakinra was started, with prompt regression of fever and skin rash, decrease in inflammatory markers, increase in reticulocytes count and weight gain. Hypotonia improved but persisted. The patient was discharged from hospital on DOL 56 in good clinical conditions, with acute phase reactants within the normal range and mild hypotonia. She is now 4 months old, still on anakinra treatment without adverse events.

Conclusion: Autoinflammatory diseases in the neonatal period are a diagnostic challenge. Clinical suspicion is crucial in order to perform specific laboratory and genetic testing and start appropriate treatment. Interstitial lung involvement may be present in MKD and, together with increased inflammatory markers, could be the first manifestation of the disease.

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AB1064

LONG-TERM SAFETY AND EFFICACY OF ADALIMUMAB IN GREEK ADULTS WITH JIA OR NON-INFECTIOUS UVEITIS AT THE TRANSITION PERIOD

Artemis Koutsonikoli¹, Despoina Dimopoulou², Maria Trachana¹, Vassiliki Sgouroulou¹, Polyxeni Pratsidou-Gertsis¹, Alexandros Garyfallos².

¹Pediatric Immunology and Rheumatology Referral Center, 1st Dept. of Pediatrics, Hippokraton Hospital, Aristotle University of Thessaloniki, Thessaloniki, Greece;

²Rheumatology Unit, 4th Dept. of Internal Medicine, Hippokraton Hospital, Aristotle University of Thessaloniki, Thessaloniki, Greece

Background: Our previous publication on Juvenile Idiopathic Arthritis (JIA) revealed that half of the pts had flares in adulthood, despite previous administration of several anti-TNFs.

Objectives: a. To describe the long-term outcome of adults with JIA or non-infectious uveitis (NIU), exposed to Adalimumab (ADA) as the single

or last biologic during the transition period. b. To evaluate our model of transition in a subgroup of refractory to conventional DMARDs JIA or NIU pts, in the 1st Greek Transition Clinic for Pediatric Rheumatic Diseases.

Methods: Retrospective cohort analysis of Greek adults with JIA or NIU between 2004 and 2018. The JIA activity state was assessed by 2 quantitative tools: the Juvenile Arthritis Disease Activity Score-10 (JADAS-10, Clinical Remission [CR], Low, Moderate and High Disease Activity [LDA, MDA, HDA], respectively) and Wallace criteria (CR on/off ADA). Baseline was defined as the 1st ADA dose

Results: 28 pts were enrolled: 24 JIA pts (female 18), aged 20.6±3.1 yrs, for a f/u of 6.5±3 years (Group-A) and 4 female pts with active uveitis (2 with JIA-associated and 2 with NIU, mean aged: 18.9 yrs, f/u: 2.3 yrs) (Group-B). The age at baseline was 14.1±3.2 yrs in Group-A and 16.7 yrs in Group-B. The lag time from JIA onset to baseline was 6.3±3.8 yrs and the JIA subtypes were: 37.5% polyarthritis RF neg, 33.3% extended oligoarthritis, 20.8% enthesitis-related arthritis, 4.2% persistent oligoarthritis, 4.2% psoriatic arthritis. 11/24 (45.8%) pts (Group-A) and 3/4 pts (Group-B) were ANA positive, 5/24 JIA-pts had a history of uveitis (20.8%). Naïve to anti-TNF drugs were 18 pts (75%) of Group-A and 4/4 from Group-B. The baseline JADAS-10 was 13.9±5.9. The ADA yr-administration was 5.3±2.3 in Group-A and 2 in Group-B. Compliance to ADA had 17/24 (70.8%) pts in Group-A and all from Group-B. Regarding safety, 4/28 patients (14.3%) experienced Events of Special Interest (1/33.6 patient-years), herpes zoster (n=1), anorexia nervosa (n=1), breast fibroadenoma (n=1) and uveitis (n=1). Among the Group-A pts: 4/24 (16.7%) discontinued ADA due to inefficacy and 1/24 due to pregnancy planning. 6/24 pts (25%) achieved CR and discontinued ADA after 5.3±1.6 yrs. CR off ADA lasted 15.5±9.8 mo. The rest 13/24 continued ADA at the last f/u. In Group-B, 3/4 were still on ADA and 1 pt discontinued ADA due to NIU remission after 4.2 yrs and sustained CR 16 mo off ADA. The median% of CR remission on ADA total administration period was 75% in Group-A and 100% in Group-B. Among the ongoing receivers: a) the JIA activity state at the last assessment was: CR 84.6%, LDA 7.7% MDA 7.7% (median JADAS: 0, Group-A), b) 100% were in remission in Group-B.

Conclusion: 86% of adult pts with JIA or NIU tolerate ADA well. Pts exposed to ADA achieved CR on/off medication in 75% and 25%, respectively.

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- *: Despoina Dimopoulou and Maria Trachana contributed equally

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AB1065

THE UTILISATION OF S100 PROTEINS TESTING IN PEDIATRIC RHEUMATOLOGY PATIENTS IN A TERTIARY CARE INSTITUTION AND IMPLICATIONS FOR CARE

Elena Tronconi, Najla Aljaberi, Angela Merritt, Alexei Grom, Grant Schuler, Jennifer Huggins, Michael Henrickson, Hermine Brunner. *Cincinnati Children's Hospital Medical Center, Division of Rheumatology, Cincinnati, United States of America*

Background: S100 proteins are calcium-binding proteins of increasing value as biomarkers in various inflammatory conditions. The two highly studied members in this family, S100A8/9 and S100A12 have been considered sensitive biomarkers of disease activity in rheumatologic disorders as rheumatoid arthritis (1) and juvenile idiopathic arthritis (JIA) (2). They are considered more accurate than conventional inflammatory indices such as the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) (3).

Objectives: To evaluate the utilization of S100 proteins in the clinical setting as supportive diagnostic tools as well as their performance in evaluating disease activity.

Methods: Patients seen at the hospital's specialty clinics who had S100 proteins tested during their care from April 2017 to August 2018. A retrospective chart review was performed to collect data on serum S100 protein levels, other inflammatory markers as CRP and disease activity measured as active joint count (AJC) in JIA patients and the presence of systemic symptoms in systemic JIA (sJIA) patients.

Graphpad Prism Version 7.05 was used for data analyses. Descriptive statistics were calculated for all variables. Comparisons were done by the Mann-Whitney U test and the Kruskal-Wallis test. Spearman correlation were calculated to assess association between S100 proteins and other numerical variables.