Physical examination revealed weight bearing pain of foot or ankle in all patients. No other sensory, vasomotor, trophic or neurological signs and symptoms were detected. Vitamin D deficiency was found in all cases (range 10 to 22 ng/ml). All patients were thus treated with adequate vitamin D daily intake. Pain relief was achieved with paracetamol, low dose ibuprofen, or a short course of oral prednisone. Rest from intense physical activity and physical therapy, avoiding detrimental feet and ankle immobilization were recommended. All children (100%) fully recovered in 3-month lag period.

Conclusion: BMO in children is a cause of disability and it is often misdiagnosed and incorrectly treated. Environmental factors, such as underestimated articular or bone microtraumas, as well as joint hypermobility, typical of paediatric age, in a bone turnover milieu of vitamin D deficiency could be the cause of the clinical conditions. Adequate vitamin D supplementation, associated with physical and analgesic therapy, are crucial in the management of BMO. We highlight the importance of supplementing vitamin D as recommended in the current guidelines, in order to avoid its deficiency and prevent invalidating clinical complaints.

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AB1061 SHORT TERM FOLLOW-UP RESULTS OF CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER AFTER CESSATION OF COLCHICINE: IS IT POSSIBLE TO QUIT?

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Background Familial Mediterranean Fever (FMF) is considered as the prototype of autoinflammatory diseases, and colchicine is the main treatment option for FMF. However, the data on cessation of colchicine treatment in FMF patients is limited

Objectives To define the characteristics of children with familial Mediterranean fever (FMF) whose colchicine treatment was discontinued and then to compare these features of the patients whose colchicine was restarted with the ones not restarted.

Methods Sixty-four out of 1786 children with FMF whom colchicine was stopped by the physician or patients/parents own decision were enrolled. These patients were grouped into two as: group 1; children whose colchicine was re-started and group 2; children whose colchicine was not re-started. The demographic, clinical and genetic data were collected and compared between group 1 and group 2.

Results Colchicine was stopped in 59.4% (38/64) by the physician and 40.6% (26/64) of them had stopped colchicine by patients/parents will. Colchicine was ceased at a median of 10.6 (2.120.5) years of age, and attack- and inflammation-free periods of 18.2 (6-148) months. The median follow-up of 64 patients after colchicine cessation was 37.4 (6.4-154.7) months. It was re-started in seventeen patients due to attacks (n=11) or elevated acute phase reactants (n=6), while remaining 47 patients did not require colchicine. The age at cessation of the colchicine was lower (p= 0.04) and duration of colchicine treatment until its cessation was shorter (p= 0.007) in group 1 than group 2.

Conclusion Even though the results of our study are not satisfactory enough to endorse the hypothesis that colchicine may be discontinued by close follow-up; older age and long duration of colchicine treatment before cessation may be two important features that should be considered in the future studies.

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AB1062 INTER-LABORATORY COMPARISON OF TYPE I INTERFERON SIGNATURE ANALYSES: PAVING THE WAY TO SHARE RECOMMENDATIONS.

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Background Interferon (IFN) signature analysis is experimentally used to classify pathological conditions characterized by a type I IFN dysregulation (i.e. monogenic interferonopathies, dermatomyositis, systemic lupus erythematosus), and to formulate targeted therapy approaches. Indeed, IFN signature is used to differentiate patients with IFN-related inflammation from conditions predominantly mediated by other cytokines (e.g. JIA and periodic fevers), through the calculation of an IFN score (IS). However, at the moment, there is not a shared threshold able to discriminate among different inflammatory conditions.

Objectives To characterize the IS in different inflammatory diseases and evaluate the concordance of IFN signature results among the laboratories of the three Italian Hospitals.

Methods Assessment of the expression in peripheral blood of six IFN-induced genes (IFI27, IFI44L, IFIT1, ISG15, RSAD2, SIGLEC1) in Real Time PCR, referring to a calibrator sample (healthy controls). Calculation of the "interferon score" (IS) for each patient using the median fold change of the six relative quantifications.

50 patients with different inflammatory disorders considered not related to IFN (e.g. Crohn, JIA, hereditary periodic fevers) were compared with established type I IFN-related diseases (SLE, monogenic interferonopathies, dermatomyositis). We analysed the concordance of the clinical classification of different inflammatory diseases and the IS results. To assess the inter-laboratory variability, twenty patients with different degree of type I IFN inflammation were analysed at three different laboratories.

Results Despite the IFN signature showed a similar and comparable trend, the IS was not consistent between different laboratories, in particular for samples with higher IS score. Thus, the concordance between clinical classification and IS could be assessed only separately for each laboratory. Although in each series IFN-related disorders showed a IS significantly higher than other kind of inflammatory disorders, preliminary results suggest there is no clear threshold able to differentiate among these groups.

Conclusion Analysis of type I IFN activation in peripheral blood showed a relevant inter-laboratory variability between our three centers, limiting the possibility of identifying a shared defined threshold to differentiate inflammatory syndromes, and suggesting the need to determine reproducible calibrator samples. Cooperation between different institutes could facilitate the collection of clinical and laboratory data in a common database, to exchange expertise in type I IFN-mediated diseases, and provide practical advices for IFN signature assessment and interpretation in the clinical practice.

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AB1063 INTERSTITIAL LUNG DISEASE IN A NEWBORN AFFECTED BY MEVALONIC ACIDURIA

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Background: Mevalonic aciduria (MA) is the most severe phenotype of mevalonate-kinase deficiency (MKD), with early onset and poor prognosis. Given its rarity and its unspecific symptoms, MA diagnosis may be

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

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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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AB1065

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

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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.