Objectives: To co-design components of an RCT of corticosteroid regimens with CYP and parents living with JIA.

Methods: A focus group was conducted with CYP with JIA and parents as part of a wider consensus and discussion group meeting within the Steroid Induction Regimen for Juvenile Idiopathic Arthritis (SIRJIA) study in December 2018. The discussion focused on two components of the RCT design: i) Discussing the most appropriate treatment protocols; and ii) Addressing practicalities associated with an RCT.

Results: Two RCT protocol options, chosen through an online survey by a clear majority out of a possible eight protocols, were discussed and critiqued: i) Protocol A (intravenous vs intraarticular corticosteroid delivery); and ii) Protocol B (intravenous vs intraarticular vs intramuscular vs oral corticosteroid delivery). Several issues pertaining to both protocols were raised, related to the influence of age and past experience, routes of administration and concerns over randomisation. Participants emphasised the importance of clinicians/researchers discussing all of the potential risks with them. Participants also wanted enough information to make an informed choice. Participants emphasised the usefulness of combining trial visits with regular follow-up appointments to minimise the burden of taking part in an RCT and had a preference for their usual hospital being the site they visited. Some participants remarked that videos could be a useful way of conveying information beyond traditional participant information sheets. Some also felt that awareness of research opportunities is not equally accessible to them either, depending on where they lived in the country. Participants would want to be kept regularly updated about the progress of the RCT and felt that incentives were a good way of keeping people engaged, although some were trepidatious to hear negative treatment results. With regards to dissemination, participants felt that study results should be readily available to them in an accessible format, should they wish to view them.

Conclusion: CYP and parents have a considerable amount of knowledge and experience which can shape the design of RCTs. With adequate support, complex concepts such as treatment protocols can be discussed and critiqued. Involving CYP and parents at the design stage of an RCT has been shown to eliminate some potential challenges in the future.

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AB1059

CLINICAL MANIFESTATIONS OF CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING WITH CYTOPENIA

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Background: Childhood-onset Systemic Lupus Erythematosus is a rare disease, with more than 50% of patients presenting with cytopenia of a decrease in at least one cell line.

Objectives: The purpose of this study was to evaluate whether clinical characteristics differed according to hematologic manifestation.

Methods: We retrospectively reviewed the presenting clinical and laboratory manifestations for 40 pediatric SLE patients (mean age 13.4 years, range 0-18 years), with leukopenia (n=12) and thrombocytopenia (n=15). Variables including age, inflammatory markers (ESR and CRP), prothrombin time, complement system (C3, C4 and CH50), immunoglobulin (IgG, IgA and IgM), Lupus anticoagulant ratio, antinuclear antibody, anti-phospholipid antibody, anti-platelet antibodies, anti β 2-glycoprotein antibody, anti-cardiolipin antibody, anti-smith antibody and anti-dsDNA antibody levels were compared according to grouping by presence of leukopenia ($<4000/\text{mm}^3$) or thrombocytopenia ($<100,000/\text{mm}^3$).

Results: Patients with cytopenia showed younger age at diagnosis than without (mean 13.1 vs. 15.2 years, $p=0.0031$ for leukopenia and 12.8 vs. 15.2 years, $p=0.014$ for thrombocytopenia), although the onset of puberty might not contribute to cytopenia. Patients with leukopenia showed lower C3 level than without (mean 44.7 vs. 73.7, $p=0.013$). Patients with thrombocytopenia showed lower anti-dsDNA titer than without (mean 94.1 vs. 259.8, $p=0.038$), and was more often negative for anti-platelet antibody (Ib/IX). There were no statistical differences among other variables. Also, there was no significant difference in one cell line decrement compared with both.

Conclusion: Pediatric SLE patients with leukopenia and thrombocytopenia in our study may independently decrease the levels of C3 and anti-dsDNA/anti-platelet (Ib/IX) antibody, suggesting respective pathologic pathways that do not affect each other.

DISCLOSURE OF INTERESTS

None declared

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AB1060

BONE MARROW FOOT OEDEMA IN CHILDREN: THE ROLE OF VITAMIN D

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Background: Bone marrow oedema (BMO) in children is a rare clinical condition characterized by joint and bone extremity pain, out of proportion to the clinical findings, exacerbated by weight bearing, in the absence of a known etiologic cause. It is associated with typical increased signal intensity on T2-weighted MRI. Management is still under debate. Treatment has mostly been reported in adult case series encompassing analgesic drugs, a variety of pharmacological treatments (corticosteroids, bisphosphonates, vasodilators), physiotherapy, reduction of weight-bearing, or core decompression. No treatment guidelines for children are to date available.

Objectives: Recently it has become evident that BMO is associated by an increase in bone turnover, in which vitamin D plays a pivotal role. In literature association between hypovitaminosis D and BMO of the foot and ankle in adult patients is reported. No data are reported in cohorts of children. The purpose of this study is to investigate the incidence of hypovitaminosis D in a paediatric population with primary bone BMO of the feet and the role of a vitamin D supplementation therapy.

Methods: A retrospective study involving 12 paediatric patients (range age 8-14 years) referred to our Rheumatologic Paediatric Clinic of Verona University in the period 2015-2018 with persistent foot pain and MRI compatible with BMO of the foot has been performed. They had all been misdiagnosed in other institutions as affected by algodystrophy or complex regional pain syndrome. Data collection included sex, age, medical and surgical history, recent or remote trauma history, symptoms at presentation, clinical examination, laboratory bone turnover markers, vitamin D levels, MRI, treatment and outcome.

Results: 2/12 patients are male and 10/12 female (male to female ratio: 1:5). 2/12 had a previous diagnosis of juvenile idiopathic arthritis ANA + with the disease in remission at the moment of evaluation. 10/12 were previously healthy.

In all cases history of minor ankle strain or recurrent microtraumas of feet prior to symptom onset had been reported. Joint hypermobility was observed in 75% of cases. One child had been previously treated with bisphosphonates and 5 with limb immobilization, without any improvement.

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