AB1056 THE EXPERIENCES OF BIOLOGICAL THERAPIES IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS ASSOCIATED UVEITIS, SINGLE CENTER STUDY

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Background: Juvenile idiopathic arthritis (JIA) is the most common systemic disease causing uveitis in childhood, with a prevalence of 10 per 100 000 persons. JIA-associated uveitis is estimated to have a poor prognosis and has a high rate of complications. JIA-associated uveitis can manifest in various forms, depending on the location and severity of the ocular inflammation, as well as on the type of arthritis. The most of JIA patients with uveitis have oligoarthritis. Therefore, other types of JIA are rarely accompanied by uveitis. Topical corticosteroids are the first line therapy, and disease conventional and biologic modifying anti-rheumatic drugs (DMARDs) are used.

Objectives: The purpose of the present study was to report on the clinical characteristics, ocular complications, treatment, and visual outcome in children with JIA-associated uveitis who were examined in recent years at a single tertiary pediatric rheumatology and ophthalmology center in Turkey.

Methods: We retrospectively analyzed the data of 41 JIA patients (14 males, 27 females).

The duration between the initial evaluation and the final visit was recorded as follow-up time. Juvenile idiopathic arthritis was defined according to the International League of Associations for Rheumatology (ILAR) classification criteria. Uveitis was classified according to the SUN classification.

Two approaches were utilized to evaluate the change of visual acuity (VA) during the disease course: (i) VA was measured on Snellen chart. The equivalent logarithm of the minimum angle of resolution acuity (logMAR) was calculated and used for analysis.

Results: The study included 31 patients (57 eyes) of whom 22 (71%) were females. Mean age (±SD) at uveitis diagnosis was 8.42 (±4.13) years (median 8, range: 34 month–17 years) and there was no significant difference between genders. The mean age at jia diagnosis was 8.42 (±4.7), respectively. Nine patients were ≤ 7 years of age at the time of JIA diagnosis. Anterior uveitis (AU) was the most common type, diagnosed in 57 (76%) eyes. All patients had methotrexate, therefore biologic therapy was used in 29/31 patients (93.5%) at the follow up time (infliximab in 12, adalimumab in seventeen, and tocilizumab in three patients) and 9 children (31%) required ≥2 biologics over the follow up period. Thirteen patients switched between infliximab and adalimumab (10 patients switched from infliximab). The reason for treatment switch included treatment failure and treatment-related side-effects (n=3). Systemic and topical steroids treatment were gradually tapered and discontinued in all patients after initiation of biologics. Of the all affected eyes, posterior synechiae (n=24) was the predominant complication on presentation. During the follow-up period new complications were seen in 11 eyes (13%). Posterior synechiea (6 eyes, 7,5%) was the most frequent complication observed followed by cataract (3 eyes, 3,%) and glaucoma (2 eyes, 2,5%) Improvement or preservation of visual acuity (VA) was noted in 77 eyes (%94,3) at the last visit.

Conclusion: We report a large cohort of children with JIA uveitis managed in a Turkey tertiary unit. Low complication rates and favorable visual outcomes are found in the present study. The high rate of biologic use), and close monitoring of affected children with pediatric rheumatology and uveitis clinic may have contributed to our improved outcomes.

Disclosure of Interests: None declared

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AB1057 DIFFICULTIES IN THE DIAGNOSIS OF THE PFAPA SYNDROME IN THE REAL CLINICAL PEDIATRICIAN PRACTICE

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Background: Symptom complex of PFAPA syndrome is characterized by a rather diverse clinical picture, including recurrent fever, aphthous stomatitis, pharyngitis, adenitis. The annual incidence of PFAPA syndrome does not exceed 0,2‰ [1], but it is believed that this disease is much more common than diagnosed [2]. The absence of specific genetic and biochemical markers of this syndrome determine the complexity of timely diagnosis and treatment of this auto-inflammatory disease.

Objectives: The object is to identify objective reasons that impede timely diagnosis of PFAPA syndrome in the clinical practice of a pediatrician.

Methods: We conducted a follow-up analysis of the medical documentation of patients with verified PFAPA syndrome who were examined and treated at the hospital pediatrics department of the university clinic for the period from 2015 to 2019 year. A survey of pediatricians and pediatric otolaryngologists was carried out on the knowledge of diagnostic criteria, protocol for the management and treatment of patients with PFAPA syndrome.

Results: During this period, in the department of PFAPA syndrome was diagnosed in 7 patients. Among patients with a diagnosis of PFAPA syndrome, there were 4 boys and 3 girls, the average age of children at the time of diagnosis was 3,4 ± 0,7 years. Before the diagnosis was verified, all patients were observed by a pediatrician or otorhinolaryngologist regarding repeated manifestations of tonsillitis and received antibacterial therapy at each episode of exacerbation of the disease. The average period from the first manifestations of the disease to the verification of the diagnosis was 1,4 ± 0,5 years. Diagnostic algorithm for verification of the diagnosis was completed by setting prednisolone test. According to the results of this test, in 100% of cases it was possible to completely arrest the phenomena of the inflammatory process (fever, tonsillitis, pharyngitis, stomatitis) and to normalize biochemical markers of inflammation (CRP) without the use of antibacterial therapy. With further observation, two of 7 patients recorded a decrease in the effectiveness of glucocorticoid therapy, which required a tonsilectomy, and one of them noted the temporary effectiveness of colchicine.

According to the results of a questionnaire survey of pediatricians and children's otorhinolaryngologists of the outpatient service, a low level of knowledge was revealed on the issues of the clinic, diagnosis and therapy of auto-inflammatory diseases. Only 40% of respondents were able to specify the diagnostic criteria for setting PFAPA syndrome and determine the further routing of these patients. It should be noted that more than half of the respondents (57%) had a clear idea about the methods of rational therapy of patients with PFAPA syndrome.

Conclusion: Thus, the complexity of the primary diagnosis of PFAPA syndrome is associated both with the clinical features of the disease and the insufficient level of knowledge of primary outpatient specialists on this issue, which is largely due to the low incidence of PFAPA syndrome. This conclusion was the reason for the inclusion of additional topics in the educational course of pediatricians.

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AB1058 CO-DESIGNING A COMPARATIVE RANDOMISED CONTROLLED CLINICAL TRIAL OF CORTICOSTEROID REGIMENS WITH CHILDREN, YOUNG PEOPLE AND PARENTS LIVING WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Previous research has identified the need for a randomised controlled trial (RCT) evaluating the most appropriate corticosteroid induction regimen to be used for children and young people (CYP) with juvenile idiopathic arthritis (JIA) (1). A recent qualitative study found that parents and CYP understood trial concepts and were able to identify potential flaws in a proposed RCT. This confirms the need to involve parents and CYP in co-designing RCTs to best meet the needs of future trial participants (2).

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 β2-glycoprotein antibody, anti-cardiolipin antibody, anti-smith antibody and anti-dsDNA antibody levels were compared according to grouping by presence of leukopenia (<4000/mm^3) or thrombocytopenia (<100,000/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

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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.