AB1056

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

Beül Sözeri¹, Esra Kardes², Ferhat Demir¹, Fatma Ciftci¹, Zeynep Demircioglu¹.

¹University of Health Sciences, Istanbul, Umraniye Training and Research Hospital, Pediatric Rheumatology, Istanbul, Turkey; ²University of Health Sciences, Istanbul, Umraniye Training and Research Hospital, Ophthalmology, Istanbul, Turkey

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 (log-MAR) was calculated and used for analysis.

Results: The study included 31 patients (57 eyes) of whom 22 (71%) were females. Mean age ($\pm$ SD) at uveitis diagnosis was 8.42 ($\pm$ 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 ($\pm$ 4.7), respectively. Nine patients were $\leq$ 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 $\geq$ 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 synechia (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

Anna Spivakovskaya, Yuri Spivakovskiy, Yuri Cnemenkov, Aisiya Dushanova. Saratov State Medical University named after V. I. Razumovsky, Saratov, Russian Federation

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 $3,4 \pm 0,7$ 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 $1,4 \pm 0,5$ 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

Simon Stones¹, Heather Bagley², Frances Sherratt³, Louise Roper³, Eileen Baildam⁴, SIRJIA Trial Management Group. ¹University of Leeds, School of Healthcare, Leeds, United Kingdom; ²University of Liverpool, Institute of Translational Medicine, Liverpool, United Kingdom; ³University of Liverpool, Institute of Population Health Sciences, Liverpool, United Kingdom; ⁴Alder Hey Children's NHS Foundation Trust, Paediatric Rheumatology, Liverpool, United Kingdom

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).