AB1047 DYNAMICS OF GROWTH AND BONE AGE IN CHILDREN WITH JIA DURING BIOLOGICAL THERAPY

Valentina Serayia. I.M.Sechenov First Moscow State Medical University, children's rheumatology, Moscow, Russian Federation

Background: In children with severe juvenile idiopathic arthritis (JIA), growth impairment is often accompanied by retardation of bone age. Dynamics of growth and bone during biological therapy is an important criterion for the effectiveness of treatment.

Objectives: To assess the effect of biological therapy on growth and bone age in children with JIA.

Methods: The study included 45 patients with JIA and (30 boys and 15 girls, mean age 12.2 ± 2.7 years; disease duration before the start of biological therapy 6.5 ± 3.3 years, 18 children with systemic JIA and 27 with polyarticular JIA). All children due to the ineffectiveness of standard therapy were transferred to biological therapy with TNF- α and IL-6 inhibitors. The study assessed growth SDS and bone age (according to x-ray imaging) before starting biological therapy (initially) and after 24 months of therapy.

Results: Of the 45 children, the initial growth retardation marked in 13 children, all with systemic JIA (growth SDS -3.19 ± 0.36 (-6.67; -2.04)). All these children also showed a lag of bone age from the passport by 2.9 ± 0.5 years, which corresponded to growth retardation. During biological therapy, the growth retardation decreased within 24 months: growth SDS -2.73 ± 0.34 (-5.87; -1.17). The lag of bone age was maintained at 2.3 ± 1.5 years, which is comparable to the baseline ($p = 0.053$). In 32 children, the initial growth was within the normal range (growth SDS 0.12 ± 1.07 (-1.97; 1.76)); 8 children with sJIA, 24 with polyarticular JIA. In all these children, the lagging of bone age from the passport at the time of the start of biological therapy was not detected (deviation from the passport age at most 1 year). During biological therapy for 24 months, growth rates remained normal: growth SDS was 0.22 ± 0.99 (-1.58; 1.67). Indicators of bone age in all children were corresponding passport age.

Conclusion: In children with severe JIA and normal growth, the bone age is more likely to correspond to the passport age; during therapy with biological agents, there is no significant change in bone age in these children. In children with severe JIA, growth impairment and bone age may be observed, most likely in children with systemic JIA. During biological therapy, there is an improvement in growth, while the lagging of bone age from the passport remains, which indicates the preservation of growth potential and the possibility of achieving higher final growth.

REFERENCES

- Uettwiller F, Perlberg J, Pinto G, Bader-Meunier B, Mouy R, Compeyrot-Lacassagne S, Melki I, Wouters C, Prieur AM, Landais P, Polak M, Quartier P. Effect of biologic treatments on growth in children with juvenile idiopathic arthritis. *J Rheumatol*. 2014 Jan;41(1):128-35

Acknowledgement: The staff of children's rheumatology department of Pediatric Clinic of I.M.Sechenov First Moscow State Medical University

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6131

AB1048 SYSTEMIC LUPUS ERYTHEMATOSUS WITH UNUSUAL PRESENTATION : SINGLE CENTRE EXPERIENCE FROM NORTH INDIA

yamini sharma, Ranbir Jaswal, Piyush Gautam, Jyoti Sharma, Sanjeev Chaudhary, Avinash Sharma. *dr RPGMC, PEDIATRICS, kanga, India*

Background: Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease. Presentation can range from constellation of non specific constitutional symptoms to definitive signs and symptoms. Systemic Lupus International Collaborating Clinics (SLICC) criteria help in making a diagnosis of SLE. SLE mimics many clinical entities and diagnosis may become difficult at times, especially if presentation is atypical. We describe children who presented with unusual manifestations and were finally diagnosed to have pediatric SLE.

Objectives: To highlight unusual presentation of pediatric SLE.

Methods: Retrospective case review of all children diagnosed as SLE from July 2017–November 2018 at a single tertiary care hospital in north India was done. We here describe 5 children who presented with unusual manifestations.

CASE 1: 8 years, girl, presented with fever for 1 month and bilateral pleural effusion. She had had 2 hospitalizations in the past. Once with fever, epistaxis and haemorrhagic shock and subsequently with fever, hepatosplenomegaly and bicytopenia and was managed as tropical infections at both times. She was evaluated this time and was diagnosed to have SLE and managed for the same.

CASE 2: 11 years, boy, presented with arthritis in multiple small and large joints of body. No other features of SLE were seen. On evaluation ANA and anti dsDNA was found to be positive and he fulfilled criteria for SLE. He was started on subcutaneous, weekly methotrexate and responded to treatment. However, he developed lupus nephritis during disease course after 6 months of diagnosis. Renal biopsy revealed Class III+V lupus nephritis. Intravenous Cyclophosphamide pulses were started for induction along with tapering doses of corticosteroids.

CASE 3: 8 years, girl, presented with early morning periorbital puffiness and abdominal distension. She also had a history of photosensitivity and Raynaud phenomenon. Urine routine examination revealed nephrotic range proteinuria with no hematuria. Clinical possibility of nephrotic syndrome secondary to SLE was kept. Renal biopsy revealed Class V Lupus nephritis. She was started on oral prednisolone and enalapril.

CASE 4: 15 years old, girl, presented with non-specific generalized body pains with joint pains associated with low grade fever. There were no other complaints. Examination was unremarkable. Investigations, however, were suggestive of SLE.

CASE 5: 17 years old, girl, presented with high grade fever and pain in bilateral knee joints and malar rash. Investigation were suggestive of SLE with MAS (macrophage activation syndrome). Child was started on iv antibiotics and iv immunoglobulins but died during the disease process.

ANA positivity, hypocomplementemia and Anti ds DNA positivity were seen in all the 5 cases.

Results: out of 11 children diagnosed to have SLE, 5 presented with unusual manifestations as described above. All patients were initiated on hydroxychloroquine and photoprotection other drugs used were prednisolone, naproxen, subcutaneous weekly methotrexate, cyclophosphamide and Azathioprine.

Conclusion: In this report we present unusual presentations as the predominant manifestation of SLE and emphasize the fact that early recognition and awareness of unusual presentation of SLE help institute timely initiation of appropriate treatment.

REFERENCES

- Iqbal S, Sher MR, Good RA, et al. Diversity in presenting manifestations of systemic lupus erythematosus in children. *J Pediatr*. 1999;135:500-505
- Bennarroch LMK, Sterba G, Bosque M. Macrophage activation syndrome as an early presentation of lupus erythematosus. *J Allergy Clin Immunol* 2006;117:s210.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.560

AB1049 CO-MORBIDITY INDEX IN CHILDREN WITH SYSTEMIC LUPUS ERYTHEMATOSUS

Ljudmila Bohmat^{1,2}, Irina Bessonova^{1,2}, Natalia Shevchenko^{1,3}, Elmira Akhnazariants^{1,2}, Tetiana Holovko³. ¹Institute of Children and Adolescents Health Care, Department of cardiorheumatology, Kharkiv, Ukraine; ²V.N. Karazin National University, Department of pediatrics, Kharkiv, Ukraine; ³V.N. Karazin National University, Department of pediatrics №2, Kharkiv, Ukraine

Background: Comorbid conditions in patients with rheumatic diseases (RD) aggravate the course of diseases, worsen the prognosis, impede the response to therapy, and reduce the quality of life of patients. Initial manifestations of comorbid lesions of organs and systems are already manifested in childhood and adolescence, and in the future are the causes of persistent morphologic and functional disorders, disability and mortality of patients.

Objectives: The purpose was to identify comorbid conditions in children with systemic lupus erythematosus (SLE), followed by an assessment of the comorbidity index (CI).

Methods: 41 patients with SLE aged 7-18 years were examined. The average duration of the disease at the time of the survey was 44.5 ± 2.9 months, 20 persons (48.78%) had a period of illness up to 3 years, 21 (51.22%) - over 3 years. Among the patients female subjects predominated - 37 (90.24%). The comorbidity index (CI) was determined by the total score (points) of the disturbances in all systems of the organism without specific diagnoses.

Results: Pathological changes of the one or two systems were noted in 19.51% of patients, between three and four systems - in 24.4%, and in some cases - five (7.32%) and six systems (2.44%).

The mean values of CI among patients with SLE were 2.6 ± 0.3 points and did not differ significantly depending on the duration of the disease (2.5 ± 0.7 points before 3 years and 2.4 ± 0.3 points after 3 years; $p > 0.05$), as well as did not differ in female and male patients (2.6 ± 0.3 and 2.8 ± 0.5 respectively, $p > 0.05$). The IC was higher in the cases of development of SLE before the age of 7 years than in older patients (3.6 ± 0.5 vs 2.4 ± 0.3 respectively, $p < 0.05$).

The activity of the pathological process significantly influenced the indicators of the CI: 2.4 ± 0.4 with its minimal activity; 2.7 ± 0.4 with an average degree of activity and 3.0 ± 0.6 with a high degree ($p < 0.05$). The frequency of the smallest CI (0 or 1) turned out to be associated with the lowest activity of the disease.

The CI with ANA-positivity association was defined: 2.8 ± 0.3 points among patients with ANA that has twice exceeded the CI of ANA-negative patients (1.4 ± 0.5), $p < 0.05$. Its highest level (like 4, 5 and 6 points) was registered only among the ANA-positive patients.

In patients with SLE with high CI, changes in the system of coagulation due to elevated fibrinogen levels predominated (3.7 ± 0.4 g/l vs 2.8 ± 0.3 g/l ($p < 0.05$). Similar changes were recorded with the atherogenic coefficient increased (2.7 ± 0.7 vs 1.3 ± 0.5 ; $p < 0.05$).

Conclusion: According to the research, it was found that in children and adolescents with SLE the formation of lesions of systems and organs occurs while maintaining the activity of the process, especially immunological (ANA-positivity). Disruption of lipid metabolism and blood coagulation were associated with the formation of comorbid states.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.3808

AB1050 TOCILIZUMAB MODIFIES CLINICAL MANIFESTATIONS AND LABORATORY FEATURES OF SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS ASSOCIATED MACROPHAGE ACTIVATION SYNDROME

Masaki Shimizu¹, Mao Mizuta¹, Takahiro Yasumi², Naomi Iwata³, Yuka Okura⁴, Noriko Kinjo⁵, Hiroaki Umebayashi⁶, Tomohiro Kubota⁷, Yasuo Nakagishi⁸, Kenichi Nishimura⁹, Masato Yashiro¹⁰, Junko Yasumura¹¹, Hiroyuki Wakiguchi¹², Nami Okamoto¹³, Masaaki Mori¹⁴. ¹Graduate School of Medical Sciences, Kanazawa University, Kanazawa, Japan; ²Kyoto University Graduate School of Medicine, Kyoto, Japan; ³Aichi Children's Health and Medical Center, Obu, Japan; ⁴KKR Sapporo Medical Center, Sapporo, Japan; ⁵University of the Ryukyus, Nakagami-gun, Japan; ⁶Miyagi Children's Hospital, Sendai, Japan; ⁷Graduate School of Medical and Dental Sciences, Kagoshima University, Kagoshima, Japan; ⁸Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan; ⁹Yokohama City University Graduate School of Medicine, Yokohama, Japan; ¹⁰Okayama University Hospital, Okayama, Japan; ¹¹Hiroshima University Graduate School of Biomedical and Health Sciences, Hiroshima, Japan; ¹²Yamaguchi University Graduate School of Medicine, Ube, Japan; ¹³Osaka Medical College, Takatsuki, Japan; ¹⁴Graduate School of Medical and Dental Sciences, Tokyo Medical and Dental University, Department of Lifetime Clinical Immunology, Tokyo, Japan

Background: Previous studies including a systematic literature review revealed clinical manifestations and laboratory features of systemic juvenile idiopathic arthritis (s-JIA) associated macrophage activation syndrome (MAS) could be modified in patients treated with tocilizumab (TCZ) ^{1,2}.

Objectives: To clarify whether TCZ modifies clinical manifestations and laboratory features of s-JIA associated MAS, and to assess performance of the 2016 MAS classification criteria for patients with s-JIA associated MAS while treated with TCZ in the real world.

Methods: A combination of expert consensus and analysis of real patient data was conducted by a panel of 15 pediatric rheumatologists. Clinical manifestations and laboratory features of s-JIA associated MAS at the MAS diagnosis in 12 patients while treated with TCZ and 18 patients not treated with TCZ were evaluated. Possible MAS was defined as having characteristic laboratory features but lack of clinical features of MAS, or atypical MAS, or early treatment that prevented fulminant MAS ^{3,4}.

Results: Among 12 patients while treated with TCZ, only 2 patients were diagnosed with definite MAS, and other 10 patients were diagnosed with possible MAS. On the other hand, among 18 patients not treated with TCZ, 10 patients were diagnosed with definite MAS, and other 8 patients were diagnosed with possible MAS. MAS classification criteria could classify the patients diagnosed with definite MAS while treated with TCZ as having MAS as well as the patients not treated with TCZ (100% and 100%, respectively). However, this criteria were less likely to classify the patients diagnosed with possible MAS while treated with TCZ as well as the patients not treated with TCZ (60% and 75%, respectively).

Furthermore, the patients with possible MAS while treated with TCZ were less likely febrile and significantly less often had rash, and had notably lower ferritin levels (587 versus 8518 ng/ml; $P = 0.0021$), compared to the patients with possible MAS not treated with TCZ. Other laboratory features of MAS including lower platelet counts, lower fibrinogen were more pronounced in patients treated with TCZ.

Conclusion: These findings show TCZ could modify clinical manifestations and laboratory features of s-JIA associated MAS. When evaluating s-JIA patients while treated with TCZ, care must be taken to not underdiagnose MAS based on MAS classification criteria.

REFERENCES

- [1] Shimizu M, et al. Cytokine 2012;58:287-294.
- [2] Sculert GS, et al. Arthritis Care Res 2018;70:409-419
- [3] Grom AA, et al. Arthritis Rheumatol 2016;68:218-228.
- [4] Yokota S, et al. J Rheumatol 2015;42:712-722.

Disclosure of Interests: Masaki Shimizu: None declared, Mao Mizuta: None declared, Takahiro Yasumi Shareholder of: Takeda, Speakers bureau: AbbVie, Novartis, CSL Behring, Naomi Iwata: None declared, Yuka Okura: None declared, Noriko Kinjo: None declared, Hiroaki Umebayashi Speakers bureau: AbbVie, Eisai, Novartis, Ono, Chugai, Tomohiro Kubota: None declared, Yasuo Nakagishi: None declared, Kenichi Nishimura: None declared, Masato Yashiro: None declared, Junko Yasumura: None declared, Hiroyuki Wakiguchi: None declared, Nami Okamoto: None declared, Masaaki Mori Grant/research support from: Tokyo Medical and Dental University (TMDU) received unrestricted research grants for Department of Lifetime Clinical Immunology from AbbVie GK, Ayumi Pharmaceutical Corporation, Chugai Pharmaceutical Co., Ltd., CSL Behring K.K., Japan Blood Products Organization, Mitsubishi Tanabe Pharma Corporation, Nippon Kayaku Co., Ltd., Ono Pharmaceutical Co., Ltd., Towa Pharmaceutical Co., Ltd., UCB Japan Co. Ltd.

DOI: 10.1136/annrheumdis-2019-eular.5479

AB1051 TRANSITION CARE IN A NEW PEDIATRIC RHEUMATOLOGY CENTER IN BRAZIL: CARING FOR ADOLESCENCE BEYOND RHEUMATISM

Larissa Pinho, Marco Felipe Silva, Ana Paula Pessoa, Carlos Nobre Rabelo Junior. Hospital Geral de Fortaleza, Pediatric Rheumatology Unit, Fortaleza, Brazil

Background: Transition care is the planned transfer of adolescents with chronic health conditions from children-centered to adult-oriented health systems. Its challenges include gaps in continuity of care, poor adherence, delays in adult services attendance, differences between pediatric and adult centers, difficulties in autonomy over care and unstable medical conditions. EULAR/PRES recently published recommendations, standards and quality indicators on transitional care for young people with rheumatic diseases, which serve as a guide for transitional outpatient clinics. In Brazil, few centers focus on transition from pediatric rheumatology. The Pediatric Rheumatology Unit of Hospital Geral de Fortaleza is a reference in North and Northeast of the country, intending to be pioneer in transitional medicine in region.

Objectives: To evaluate the adolescents and their families' perception regarding follow-up and autonomy in health care, as well as to identify the main risks inherent to this age group.

Methods: A longitudinal and descriptive study was carried out through interviews, questionnaires and medical records analyzes. Adolescents between 15 and 18 years old, regularly followed, who had at least three planned visits/year were included. Patients with cognitive impairment or without definite diagnosis were excluded. Epidemiological and clinical data were collected; self-management and health care utilization skills were evaluated by Transitional Readiness Assessment Questionnaire (TRAQ), a self-administered questionnaire.

Results: Eighty two patients were recruited into the transitional service (61 female [74.4%]). Patients' ages ranged from 15.0 to 18.7 years (median 16.9). Regarding religion, 41 patients (54.7%) were Catholic and 27 were Protestant (36%). Concerning to ethnicity, most of them declared themselves brown (61%) or white (22%). Fifty-nine adolescents (72%) considered themselves able to take care of their own follow-up, whereas 39 relatives (50%) considered the patient capable of doing so. The most frequent diseases were juvenile idiopathic arthritis (31.7%) and systemic lupus erythematosus (29.3%), of which 37 (45.1%) were in remission. Median disease duration and follow-up were 48.5 (5 - 180) and 34.5 (1 - 175) months, respectively. The median TRAQ value was 3.27 (1.61 - 4.55). There were 191 transitional visits (median 2.0 visits/patient). Fifty three patients had 2 or more visits, out of which 42 (79.2%) had at