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AB0981 10-YEARS EXPERIENCE OF ETANERCEPT USE IN TREATMENT OF JUVENILE IDIOPATHIC ARTHRITIS IN CHELYABINSK REGIONAL PEDIATRIC HOSPITAL

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Background: Juvenile idiopathic arthritis (JIA) in recent decades has changed its course, thanks to the use of genetically engineered biological drugs. If therapy with methotrexate is inefficient etanercept is the drug of choice for JIA treatment. Etanercept has been used in Chelyabinsk regional pediatric hospital during 10 years.

Objectives: Performance and safety assessment of etanercept in patients with JIA in Chelyabinsk regional pediatric hospital.

Methods: 51 children aged from 3 to 17 (mean age 10,0 years) diagnosed with JIA were under monitoring (12 boys, 39 girls). Disease duration was from 2 to 15 years (mean duration 5,4 years). JIA was diagnosed based on ILAR diagnostic criteria. Oligo arthritis was diagnosed in 9 children, sero-negative polyarthritis was diagnosed in 34 children. 3 patients had systemic JIA (without active systemic presentations), 5 children had enthesitic JIA. X-ray stage 1-2 was observed in 45 children and stage 3-4 in five. Enhancement antigens HLA B 27 were found in 11 children. In all children methotrexate was ineffective in dose of 15 mg/m² during 6-12 months. Etanercept was introduced in dose of 0.8 mg QW. Therapy duration was from 3 months to 8 years (mean duration 29 months). Assessment of disease activity and therapy efficiency was conducted in accordance with ACR pedi criteria. Nonparametric statistical methods were used to compare results.

Results: Prior to etanercept use high disease activity was observed in all children. Mean number of joints with active arthritis was 8 [4, 10] (Me[25;75%]). Mean number of joints with functional impairments – 4 [2;10]. Mean ESR (according to Panchenkov) – 23 [10;35] mm/h, CRP 12,0 [5,7;32] g/L. Assessment of functional activity according to CHAQ questionnaire – 1,25 [1;2]. Activity assessment according to VAS by doctor – 70 [60;70]. Assessment of parents/patients according to VAS 70 [60;80]. No active systemic presentations and eye lesions were found in children under monitoring.

On the background of etanercept therapy a decrease in disease activity was observed in 50 patients. Mean number of joints with active arthritis was 0 [0;2] (Me[25;75%]) ($P \leq 0.0001$). Mean number of joints with functional impairments – 0 [0;2] ($P \leq 0.0001$). Mean ESR was 5 [3,6] mm/h ($P \leq 0.0001$), CRP 3 [2;4] g/L ($P \leq 0.0001$). Assessment of functional activity according to CHAQ questionnaire was 0,125 [0;0,5] ($P \leq 0.0001$). Activity assessment according to VAS by doctor – 10 [5;20] ($P \leq 0.0001$). Assessment of parents according to VAS 10 [5;20] ($P \leq 0.0001$).

Clinical disease remission (according to ACR pedi criteria $\geq 90\%$) was observed in 32 patients after 6-12 months of treatment. Remission duration up to now is from 1 to 7 years. Efficiency according to ACR pedi criteria is 70% in 11 children, 50% in 5, 30% in 2.

Etanercept was well-tolerated. Drug was cancelled in 9 patients. 6 patient (11,8%) developed bilateral uveitis, one patient had an allergic reaction (rash), one - systemic manifestations, one - urinary tract infection. There were no cases of tuberculosis.

Conclusion: Etanercept therapy was highly effective and safe in patients with JIA. Clinical remission was achieved in 62,7% children. Decrease in disease activity was observed in 98% of children. 11,8% patient developed uveitis, on average, after a year of using etanercept.

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AB0982 CENTRAL NERVOUS SYSTEM ANGIITIS: THE EXPERIENCE OF A TERTIARY PEDIATRIC RHEUMATOLOGY BRAZILIAN CENTER

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Background: Central nervous system (CNS) angiitis is a severe and rare inflammatory brain disease whose course varies from patient to patient. May be associated with infections, malignancies, metabolic diseases or systemic collagen vascular disorders. It is classified as primary or idiopathic in the absence of associated systemic diseases. The exact incidence is still undetermined.

Objectives: To describe the clinical and laboratory data, the response to therapy and outcome of patients with CNS angiitis seen at a Tertiary Pediatric Rheumatology Center.

Methods: This is a retrospective, single-center chart review study of pediatric patients with clinical diagnoses of CNS angiitis, followed up in a Tertiary Pediatric Rheumatology Center, from January 2009 to December 2018. Diagnosis was confirmed by magnetic resonance angiography (MRA) or after exhaustive exclusion of other causes.

Results: Nine patients (4 girls) were enrolled in this study. Mean age at inclusion and at diagnosis was 11.5 years (7.8 to 19.9) and 8 years (3,5 to 12,5), respectively. Among the 9 patients, 4(44%) were diagnosed with primary CNS angiitis. Polyarteritis nodosa (n=4) and Behçet disease (n=1) were the aetiologies of secondary CNS angiitis. Main clinical features were sudden onset of seizures (67%) and headache (55%). Other important symptoms were: decreased level of consciousness, hemiparesis and neurocognitive dysfunction. In the cerebral spinal fluid, protein was elevated in 22% of patients. The pattern of lesions was bilateral in 67% and multifocal in 55%, being suggestive of ischemic lesions by CNS magnetic resonance in 78% of the cases. The MRA was conclusive in all cases of secondary and normal in only one case of primary CNS angiitis. Increased erythrocyte sedimentation rate, C-reactive protein and leukocyte count were more frequently observed in patients with secondary CNS angiitis as compared to patients with primary angiitis. In one case of primary CNS angiitis with negative vascular study, the Von Willebrand factor antigen was positive, being useful in the differential diagnosis. Steroids were administered in 100% of patients, associated with immunosuppressants in 7/9 cases. Induction therapy with intravenous cyclophosphamide was used in 78% of the cases and in the maintenance phase, azathioprine in 78% of them. No relapses occurred. The most commonly clinical sequel observed was residual epilepsy (55%).

Conclusion: In this study, seizures were the most frequently symptom found. Steroids plus immunosuppressants were effective in the disease control. MRA was positive in all cases of secondary CNS angiitis, being effective in the diagnosis of this condition. Recognition of findings and adequate diagnosis guides the treatment, which should be specific to the underlying cause, aiming to provide a good neurologic outcome.

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AB0983 PENTRAXIN 3 AS A MARKER OF DISEASE ACTIVITY IN JUVENILE IDIOPATHIC ARTHRITIS PATIENTS

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Background: Pentraxin-3 (PTX3) is a secretory acute phase protein which is produced and expressed in many immune cells especially macrophages, fibroblasts and endothelial cells at different inflammatory sites [1].