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AB0979

CORTICOSTEROID TREATMENT IN PEDIATRIC RHEUMATIC DISEASES AND SUPPRESSION OF THE HYPOTHALAMIC-PITUITARY AXIS

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Background: Corticosteroids are the mainstay in the treatment of several chronic inflammatory diseases. Long-term treatment may lead to suppression of hypothalamic-pituitary-adrenal axis, which in turn can be dangerous in stress situations such as after surgical interventions. Oral replacement therapy may be needed in some cases. Data on pediatric populations are scanty.

Objectives: The aim of our study has been to evaluate hypothalamic-pituitary-adrenal function during corticosteroid tapering by measuring serum cortisol.

Methods: During long term corticosteroid treatment, serum cortisol levels were evaluated when oral prednisone dose was decreased to 7.5 mg/day and also after 4-6 weeks, and if below range ACTH levels were also determined.

All patients aged < 18 years seen in our center during the last 6 months who were on corticosteroid treatment for > 1 month were included. Serum cortisol levels were considered normal if in the range of 5-25 µg/dL in a morning fasting blood drawing, while ACTH levels were considered normal in between 6 and 55 ng/L (with chemiluminescent method). Clinical and demographic data were recorded from clinical charts in a customized database.

Results: We have included in this preliminary study 12 patients (7F, 5M) affected by uveitis (n=4), JIA (n=4), scleroderma (n=3), lupus (n=1). The mean age (at the time of sampling) was 11.5 years, with a median of 10 and a range of 8-17. Prednisone starting dose was 1 mg/kg/day.

Four out of 12 patients had decreased cortisol levels. Characteristics of these four patients (none of whom had additional steroid pulses) are detailed in the table.

Conclusion: One third of our patients had decreased cortisol levels, after three months of prednisone treatment. In one case, who showed persistent low levels, oral supplementation with hydrocortisone 20 mg/day for two months was needed. Our study is ongoing and results could help in identifying patients at risk for adrenal crisis.

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AB0980

EFFICACY OF ETANERCEPT TREATMENT FOR CHILDREN WITH KAWASAKI DISEASE INTRACTABLE TO INTRAVENOUS IMMUNOGLOBULIN THERAPY IN RUSSIAN CHILDREN

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Background: Kawasaki disease (KD) is recognized in developed countries as a fundamental reason of acquired heart diseases in children nowadays. The main treatment is infusion of intravenous immunoglobulin (IVIG) at a dose of 2 g/kg. However for about 10-30% of children is reported the development of resistance to standard therapy: continuance of fever ($\geq 38,0^{\circ}\text{C}$) or recurrency of fever after 36 hours afebrile period ($T < 37,5^{\circ}\text{C}$), but not later than on the 7th day after completion of the IVIG infusion at a dose of 2 g/kg. Until now there are no unified recommendations for treatment of such patients. As alternative therapy researchers empirically apply second infusion of IVIG (77%), high doses of glucocorticoids (18%), blockers of tumor necrosis factor α (3%) and interleukin-1, cyclosporine and other cytostatic drugs.

Objectives: Efficacy evaluation of therapy by inhibitor of TNF-alpha (Etanercept) in treatment of KD immunoglobulin-resistant forms according to the data from Morozovskaya Children's City Clinical Hospital, Moscow.

Methods: There were examined 152 patients (boys:girls – 2:1, median age Me = 21 months [10;36]) with KD hospitalized in Morozovskaya Children's City Clinical Hospital in 2014-2018. The frequency of complete form of KD was 80,6%. All children had a standard therapy – IVIG. Efficacy evaluation was based on normalization of body temperature, decrease of neutrocytosis, decrease of C-reactive protein (CRP) level. In case of ineffectiveness of therapy the second infusion of IVIG, pulse-therapy and inhibitor of TNF-alpha (Etanercept) were used.

Results: The resistance to standard IVIG therapy was revealed in 16 children (10,5%), mostly in boys (4,3:1) at the age of Me = 21,5 months [9,5; 34]. Complete form of KD was diagnosed in 11 patients (68,8%), incomplete form – in 5 children (31,2%). Cardiovascular lesions were noted in 14 children (87,5%): pericarditis – in 2 (12,2%), coronary arteries (CA) lesions – in 12 (75%), with formation of aneurysms in 8 (50%), including the giant ones in 3 children (18,8%); peripheral arteries damages were noted in 2 children (12,5%), thrombosis and thromboembolism – in 4 children (25%).

To all of children was made the second IVIG infusion, 13 of those children (81,2%) had a positive effect: 11 children with complete form of KD, 2 children with incomplete form of KD. For 3 children (18,7%) with incomplete form of KD and significant cardiovascular changes (pericarditis, giant aneurysm of coronary and peripheral arteries, thrombosis and thromboembolism) it was needed an additional therapy. As a second-line therapy the methylprednisolone pulse-therapy was used in 2 children (12,5%) at a dose of 10 mg/kg, without sufficient effect. For these three patients (3) as a third-line therapy was applied Etanercept at a dose of 0,8 mg/kg/a week subcutaneously. In all children normalization of body temperature, decrease of leukocytosis by factor of 1,5-3 and decrease of CRP by factor of 10-15 occurred after three injections of Etanercept. However, for 1 child it was needed the additional fourth injection of Etanercept because of repeated fever, increased laboratory parameters, development of thrombosis of the left coronary artery after 2 weeks upon discharge.

Conclusion: The frequency of immunoglobulin-resistant forms was 10, 5%. The first-line therapy of resistance (second IVIG-infusion) was efficient in 13 children (81,2%). Methylprednisolone pulse therapy was ineffective. The use of 3 subcutaneous injections of Etanercept at a dose of 0,8 mg/kg/ a week resulted in decrease of intensity of inflammatory changes and in fever relief, but it did not prevent the development of relapse and thrombosis of coronary arteries.

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Patient	Gender	Age	Disease	PDN initial dose [mg/Kg/die]	PDN duration of therapy up to 7.5mg/day	Cortisolemia (1) [mcg/dl]	Cortisolemia (2) [mcg/dl]	ACTH [ng/L]
1	M	17	Uveitis	0.9	3 months	4,37	N/A	N/A
2	F	13	Scleroderma	1	3 months	2,87	5,22	15,90
3	F	8	JIA	1.4	4 months	0,20	2,68	7,98
4	M	10	Uveitis	1.3	3 months	4,3	N/A	25,01

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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].