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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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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