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CLINICAL AND PREVENTIVE EFFICACY OF PHYTOPREPARATIONS IN PRE-SCHOOL CHILDREN WITH ADENOTONSILLAR PATHOLOGY

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Aim Assessing efficacy of Tonsilgon N in pre-school children as an etiotropic drug in complex therapy of adenotonsillar pathology at the rehabilitation stage.

Materials Within the period from 2011 to 2015, 1,076 children (aged from 2 to 5 years) with a history of pathology of nasopharyngeal and palatine tonsils in 100% of cases were reviewed. Pre-school children were given Tonsilgon N phytopreparation in the form of drops in age-specific doses within 30 days. The control group included 200 children. The phytopreparation efficacy was analyzed before and after the preventive therapy.

Results A year prior to the preventive therapy, degree II nasopharyngeal tonsil hypertrophy complicated by adenoiditis was noted in 71.3% of children in the treatment group and in 74.5% of children in the control group. Degree III nasopharyngeal tonsil hypertrophy made 28.7% and 25.5%, respectively. A year after the rehabilitation therapy, only 32.9% of patients in the treatment group did not show improvement in the clinical presentation. Symptoms of adenoiditis were practically relieved in half of the patients, nasal breathing was restored in 95.8% of patients, the size of a nasopharyngeal tonsil was reduced from degree II to degree I in 64.9% of patients. Normalization of the rhinoscopy presentation was noted in 82% of patients, 77.9% of patients showed reduction of the ARI rate from 5–8 to 2–3 times a year. Endoscopic control confirmed reduction of size of both nasopharyngeal and palatine tonsils in 64.9% of patients. The hypertrophy degree of nasopharyngeal and palatine tonsils in pre-school children not receiving Tonsilgon N phytopreparation did not show any change. In addition, hypertrophy demonstrated increase from 74.5% to 83% over time.

Conclusions Prospective study of children receiving Tonsilgon N as a rehabilitation method demonstrated its efficacy on immune development of the respiratory tract. The obtained data allow us to recommend Tonsilgon N to be included in programs of immune rehabilitation and prevention of exacerbation of adenotonsillar pathology.

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BAC-SET® FORTE MULTISTRAIN PROBIOTICS COMPLEX IN THE PREVENTION OF ADENOTONSILLAR PATHOLOGY IN PRESCHOOL CHILDREN

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Goal to study the efficacy of the probiotic complex Bac-Set Forte Multistrain in the medical treatment of children with adenotonsillar pathology.

Methods during the period of 2016–2019, 346 children (3–6 years old) with chronic pathology of pharyngeal and Palatine tonsils, suffering from recurrent respiratory infections, were examined. The treatment group (n=230) was getting a multistress probiotic complex Bac-Set Forte (UK) daily, 1

capsule per day for 30 days, as well as irrigation and elimination therapy, as prevention of exacerbations of chronic nasopharyngeal pathology. The group under control (n=116) was getting only irrigation and elimination therapy.

Results before the start of the therapy with the probiotic complex Bac-Set Forte, pharyngeal tonsil hypertrophy of the 2nd degree with complicated adenoiditis was observed in 76.3% of children in the treatment group and in 75.8% of children in the control group (P=0.2376). By the end of the study nasal breathing was restored in 62.7% of patients of the treatment group; symptoms of adenoiditis were stopped in 51.8% of patients (P=0.000); 82.7% of patients had a decrease in the volume of the pharyngeal tonsil to the 1st degree (P=0.000); 78.9% of patients had a normal rhinoscopic picture (P=0.000); endoscopic control confirmed a decrease in the size of the palatine tonsils in 56.8% of patients (P=0.000), a decrease in the frequency of recurrent respiratory infections in 72.6% of children from 5–8 times to

2–3 times per year (P=0.000). In the control group, the degree of hypertrophy of the pharyngeal and Palatine tonsils did not change and even increased in dynamics in 81.4% of patients.

Conclusion the results of the study confirmed the effectiveness of the probiotic complex Bac-Set Forte Multistrain. Prospective monitoring of children who received Bac-Set as prevention of exacerbations of adenotonsillar pathology confirmed its effectiveness in forming the immunity of the respiratory tract.

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SEVERE PRESENTATION OF NETHERTON SYNDROME: A CASE REPORT

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Netherton syndrome (NS; MIM256500) is a rare genodermatosis with autosomal recessive inheritance characterized by the triad of ichthyosiform erythroderma, hair shaft abnormality and an atopic diathesis. The primary defect in NS is loss-of-function mutations in the gene coding for serine protease inhibitor Kazal-type 5 (SPINK5). The defective expression and function of this inhibitor induce a severe skin barrier defect. It is also considered a primary combined immunodeficiency with associated or syndromic features as many patients have an increased tendency for infections and abnormal levels of various immunoglobulins.

We report a case of a female patient born as a first child of nonconsanguineous parents that presented in early infancy as a severe systemic disease with generalized erythroderma, dryness and skin desquamation, hypernatraemic dehydration, failure to thrive and recurrent severe skin and systemic infections (Staphylococcus aureus, Klebsiella pneumoniae, Pseudomonas aeruginosa). During follow-up, she presented with elevated IgE levels, atopic dermatitis and food allergies (including anaphylaxis to egg proteins). Flow cytometry of peripheral blood lymphocytes and lymphocyte proliferation test were within the reference range. Due to severe life-threatening infections, the patient receives monthly immunoglobulin infusions. Genetic analysis revealed that our patient has two pathogenic SPINK5 variants, c.[153delT], p.(Gln52LysfsTer6) and c.[1431-12G>A], p.(?). It has been reported that cases of

NS associated with c.153delT are particularly severe as demonstrated in our patient.

Testing of both parents was recommended as each parent of a child with autosomal recessive disease is generally considered to carry at least one copy of an aberrant allele. We found that her father has a pathogenic variant 153delT], p. (Gln52LysfsTer6), while her mother has no pathogenic variants. Genetic counseling emphasized that the genetic finding in our patient was not due to uniparental disomy and that the likely cause is de novo mutation in the maternal germinal cell. Although no specific treatment is available for Netherton syndrome, the first year of life has to be carefully managed, with rigorous care procedures that include multidisciplinary care. This approach should include careful monitoring and treatment of infections and hypernatremia, individual recommendations for calorie and protein needs for adequate growth, careful monitoring of allergic sensitization and dietary advice to optimise their nutritional intake, local skin treatment and supportive immunoglobulin therapy.

Following introduction of immunoglobulins, the patient had no infections during 2-years follow-up. However, despite intensive and individualized dietetic approach and nutritional treatment, our patient is still severely malnourished (WHO z-score: weight-for-age -2.49; length-for-age: -1.29; weight-for-length: -2.5).

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FOLLOW-UP OF TWO CHILDREN WITH VERY LOW IGE FROM BIRTH

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Background Immunoglobulin E (IgE) is best known for its association with allergic diseases. Very low IgE may be a marker for other immunodeficiencies. Deficiency or low IgE is defined with IgE concentration lower than 2.5 IU/ml. A majority of studies suggest a prevalence of 1-2.6 %.

The clinical significance of an isolated low IgE („selective IgE deficiency”) is unknown and is not currently included in the consensus classification of Primary Immunodeficiency Diseases (PIDD). Low IgE is suggestive of a primary humoral immunodeficiency, like Common variable immunodeficiency (CVID). In recent studies a connection between low serum IgE and low levels of one or more classes of immunoglobulins are found. The children's group was characterized with a higher prevalence of asthma.

Both groups (children and adults) had significantly higher prevalence of chronic sinusitis, otitis media, autoimmune, and oncological diseases than controls. In our case report two cases of very low IgE are presented. Both of them have been followed up from birth.

Case presentation: a cohort of 72 children has been followed up from birth up to 10 years of age. In two of them a very low serum IgE was found. Serum IgE was assessed at birth from cord blood, and reassessed at 1, 2 and 10 years of age with specific IgE (sIgE) for standard palette of food and inhalant allergens. At 10 years of age skin prick test, measurement of fractional exhaled nitric oxide (FeNO) and spirometry have been conducted.

The girl had an unmeasurable level of IgE in cord blood, and also at 1 and 2 years, and her IgE at 10 years was 2.0

IU/ml. The boy's IgE in cord blood, and at 1, 2 and 10 years was 0.11, 1.0, 8.2 and 0.1 IU/ml, respectively.

They both had symptoms characteristic of asthma and allergic rhinoconjunctivitis throughout their childhood, with negative sIgE at 1, 2 and 10 years of age and no evidence of type I hypersensitivity. Their other immunoglobulin (IgG, IgM, and IgA) levels at 10 years was within reference range for age, and they showed no symptoms suggestive of immunodeficiency or autoimmunity.

Conclusion here we report of two cases with selective IgE deficiency in children who had been followed up from birth. Very low serum IgE may serve as a marker of immune dysregulation and autoimmunity, and should trigger appropriate investigation (immunoglobulin quantification). The children will be closely monitored.

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52-WEEK LABORATORY SAFETY FINDINGS FROM AN OPEN-LABEL EXTENSION STUDY OF DUPILUMAB IN ADOLESCENT PATIENTS WITH ATOPIC DERMATITIS (LIBERTY AD PED-OLE)

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Objective No clinically significant changes in hematology/chemistry parameters were reported after clinical trials of dupilumab in adults and adolescents with moderate to severe atopic dermatitis (AD). Here, we report long-term laboratory outcomes in adolescent patients (aged ≥ 12 to < 18 years) from an ongoing long-term open-label extension (OLE) trial (LIBERTY AD PED-OLE; NCT02612454).

Methods Patients with moderate-to-severe AD who had previously participated in 1 of 3 prior dupilumab studies were enrolled in this OLE study. In March 2017, weight-based weekly dosing (2 or 4 mg/kg) was amended to a fixed-dose regimen (300 mg every 4 weeks), which could be up-titrated in case of inadequate clinical response at Week 16 as: patients < 60 kg, 200 mg every 2 weeks (q2w); patients ≥ 60 kg, 300 mg q2w. Concomitant topical corticosteroids and topical calcineurin inhibitors were allowed. Laboratory safety data for the adolescent cohort were evaluated (data cutoff: December 15, 2018) from OLE baseline (n = 299) to Week 52 (n = 105).

Results Hematology assessments at Week 52 demonstrated that mean (standard deviation [SD]) counts of eosinophils ($0.58 \times 10^9/L$ [0.52]), basophils ($0.01 \times 10^9/L$ [0.03]), monocytes ($0.46 \times 10^9/L$ [0.17]), lymphocytes ($2.17 \times 10^9/L$ [0.65]), neutrophils ($4.11 \times 10^9/L$ [1.58]), leukocytes ($7.35 \times 10^9/L$ [2.03]), hemoglobin (139.7 g/L [13.80]), and platelets ($283.5 \times 10^9/L$ [65.28]) remained within the normal reference range for this adolescent population. Similarly, chemistry parameters at Week 52 showed no clinically relevant changes from baseline as seen from mean [SD] levels of creatinine kinase (141.6 U/L [117.3]), albumin (45.9 g/L [3.0]), protein (72.5 g/L [4.7]), bilirubin (7.0 $\mu\text{mol/L}$ [5.5]), potassium (4.3 mmol/L [0.3]), alkaline phosphatase (160.1 U/L [92.0]), creatinine (57.2 $\mu\text{mol/L}$ [13.4]), blood urea nitrogen (4.2 mmol/L [1.2]), lactate dehydrogenase (186.1 U/L [39.7]), and glucose (4.9 mmol/L [0.7]).

Conclusions No clinically meaningful changes in hematology/chemistry parameters were seen in adolescents with 52 weeks