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).

160

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.

161

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

of dupilumab treatment, consistent with results in adults and adolescents. These data suggest that no routine laboratory monitoring for hematology/chemistry parameters is required in adolescents with AD prior to or during dupilumab treatment.

162

CHARACTERIZATION OF PEDIATRIC PATIENTS UNDER ALLERGEN-SPECIFIC IMMUNOTHERAPY AT A LEVEL II HOSPITAL

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Allergen-Specific Immunotherapy (ASI) is being used on the treatment of respiratory allergies for over a century, aiming to reduce individual hypersensitivity to specific allergens, and is proven to be very effective on the long term.

Goal Evaluation of the patients followed in the Paediatric Allergology clinic at a level II Hospital treated with ASI.

Retrospective and descriptive study of the efficiency of ASI in patients attending our Allergology inpatient clinic between 2016 and 2020.

The sample was characterized according to patients' gender, age, initial diagnosis, method of allergy testing (blood IgE or Skin Prick Testing) and the type of ASI used.

During the estimated period, a total of 44 patients were treated with ASI.

Of these, 27 (61%) are male, with a median age of 11 years old on the first day of treatment (minimum age: 6 years old, maximum age: 17 years old).

As to the diagnosis: 34 (77%) have Allergic Rhinitis, 20 (46%) have Allergic Asthma, 9(21%) Allergic Conjunctivitis, 3 (7%) Urticaria, and 2 (5%) Atopic Dermatitis. The majority of patients have more than one condition identified.

All the patients underwent Skin Prick Testing and 60% also did blood tests.

Grasses and domestical mites were the agents more frequently identified as inducers of Hypersensitivity, and 36% of the patients received monotherapy.

Subcutaneous administrations was the preferred method in 2/3 of the patients.

There were no adverse effects observed until today and the ASI was proven to be effective to reduce allergic symptomatology.

ASI is the only treatment modality known to effectively change the natural course of Allergic disease.

However, it is essential to thoroughly select the patients that can qualify for the treatment, as to the specific allergen extracts being used, so that we can optimize therapy benefits.

163

AMOXICILLIN ALLERGY IN CHILDREN... COMMON OR UNCOMMON? (#151 WITH CHANGES)

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β -Lactam antibiotics are safe and cost-effective antibiotics, being amoxicillin the most common antibiotic used among the paediatric population.

Many patients report allergic reactions to this antibiotic, but amoxicillin allergy range between 1-10%. However, clinicians hesitate to prescribe it when a suspected, but unproven, allergy exists. Our aim is to confirm amoxicillin allergy in children with clinical suspicion.

This study was done between January 2018 and December 2020, in children younger than 18 years, admitted to the emergency room with suspicion of clinical allergic reaction to amoxicillin. According to the protocol of our hospital, they were referred for pediatric allergology appointment to perform prick tests and afterwards oral provocation test.

A total of 57 cases were referred for evaluation. The average age was 8.4

(1-17) years old, and 53% were female. The suspicion was based on late rash reaction presented in 68.4%, urticarial exanthema in 19.3%, edema in 8.8%, vomit in 8.8%, cutaneous rash in 7% and dyspnea in 2.1%. In 18 patients, specific IgE screening for amoxicillin was performed, but all results were negative. All children did a prick test for amoxicillin and oral provocation test. There were no positive results for prick tests, but two positive results in the oral provocation test (3.5%).

Confirmation of amoxicillin allergy, before deciding to use it is or not, is an important tool for antimicrobial stewardship and, consequently, to decrease the rate of antibiotic resistance. So far, in our hospital, there were only two positive results.

164

SERUM PERIOSTIN AS A POTENTIAL BIOMARKER FOR ASTHMA SYMPTOMS IN CHILDREN WITH HISTORY OF RESPIRATORY SYNCYTIAL VIRUS INFECTION

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Periostin is a matricellular protein upregulated in response to IL-4 and IL-13, that have a role in development of allergic diseases. Previous studies reported that periostin can be a non-invasive biomarker of T2-driven inflammatory response in asthma in adults, with inconsistent results in children. None of the studies examined the association of serum periostin levels with asthma symptoms in children who have been infected with respiratory syncytial virus (RSV) in the first two years of life. The aim of this study was to determine the usefulness of serum periostin levels as a potential biomarker for asthma, especially recent asthma symptoms in children.

This prospective study observed 72 children from birth. RSV infection was confirmed with positive serum specific RSV Immunoglobulin G (IgG) at one and/or two years of age. Asthma was diagnosed according to International Study of Asthma and Allergies in Childhood (ISAAC) questionnaire at 10 years of age. Fractional exhaled nitric oxide (FeNO), lung function, skin prick test and blood samples for analysis of specific immunoglobulin E (sIgE) on standard pallet of inhalant allergens, total IgE (tIgE) and periostin were provided.

At 10 years of age, asthma was diagnosed in 23 (31.9%) of the observed children. In 15 (20.8%) of them who reported asthma symptoms during the last

12 months, median serum periostin levels were 40.04 ng/ml. In 57 (79.2%) children who were free of asthma symptoms in the last 12 months, mean serum periostin levels were 30.57 ng/ml. Serum periostin levels correlated significantly