and swollen lips. Following parenteral administration of epinephrine and glucocorticoid together with intravenous bolus of fluids the boy recovered. Two hours earlier he had eaten tuna steak. His mother tasted a piece of it and reported mild tongue pruritus.

Therefore the boy was diagnosed of having scombroid poisoning. Two days later he performed some experiments at home. He realized that his hand in a container with cold water, as opposed to a hand in warm water, has swollen and blushed. When he drank a small amount of an ice tea he felt throat tightness. Barefoot walking on the cold floor caused to him itching of the soles of the feet. The ice cube test was performed at hospital. The test resulted positive and diagnosis of cold urticaria and anaphylaxis was made.

For the whole next year the boy suffered from hypersensitivity reactions to cold. Continuous oral antihistamine therapy made symptoms less severe.

However, it is interesting that the boy got over an acute enteral infection caused by *Campylobacter jejuni* just a week before the reported episode of anaphylaxis to cold occurred. Moreover, two days before the symptoms of the enteral infection started the boy swam in a pool with very cold water (colder than sea water) and had no reactions. Conclusion: The exact mechanisms of hypersensitivity reaction and histamine release to cold are not completely elucidated. There are many infective agents that have already been associated with cold urticaria but *Campylobacter jejuni* was not one of them. Future investigations are needed to reveal the pathophysiologic pathways that connect these two conditions.

153 ATYPICAL FOOD-PROTEIN INDUCED ENTEROCOLITIS SYNDROME

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Food-protein induced enterocolitis syndrome (FPIES) is a non-IgE mediated gastrointestinal hypersensitivity, categorised into two major phenotypes: acute and chronic. Acute FPIES manifests as profuse, repetitive vomiting, 1-4 hours after exposure to a triggering food, sometimes with diarrhoea 5-10 hours later, leading to dehydration and lethargy. In chronic FPIES symptoms develop over days or weeks with intermittent emesis, progressive diarrhoea, abdominal distension and failure to thrive. The pathophysiology of FPIES remains obscure, but there is evidence of profound activation of innate cells, eosinophils, and pan-T-cell activation. Most FPIES cases are induced by cow's milk proteins and soy in infants, although other single or multiple foods can be triggers, including solid foods, where there are significant geographical differences. The incidence ranges from 0.015 to 0.5%. There is a significant lack of awareness among professionals about FPIES and significant delay of diagnosis. There are some adult patients, too. Diagnosis is purely clinical, and there are diagnostic criteria for patients presenting with possible PFIES and diagnostic criteria for the interpretation of oral food challenges (OFC), as shown here. The most important aspect of management in patients with FPIES is the avoidance of triggering food. The natural course is favourable, and there are no reports of long-term complications in children with FPIES and it is largely a self-limiting, generally benign disorder of childhood.

Patients with FPIES usually have undetectable specific IgE to triggering food, except in rare cases of atypical FPIES. In addition to classic acute FPIES symptoms, children with atypical FPIES can also have symptoms consistent with an acute IgE-mediated hypersensitivity such as urticaria, angioedema, flushing, and wheezing, and are likely to have a more protracted course. Rarely, patients with IgE-mediated food allergies develop non-IgE-mediated food allergies and vice versa. Our patient is a child who had been avoiding cow's milk due to immediate cutaneous reaction on ingestion, with positive specific IgE and skin prick-to-prick test. After ten months of avoidance, and negative new specific IgE, she was challenged for resolution. She tolerated cow's milk during the 2 hour observation period, but then developed delayed profuse vomiting. FPIES was confirmed with another, FPIES designed oral food challenge, and she has been advised to continue using an extensive hydrolysate formula. Due to the known possibility of developing FPIES to other proteins, their cautious introduction is advised. Further follow-up will be taken, considering OFC every 18-24 months.

154 FOOD ALLERGY WITH IDIOPATHIC ANAPHYLAXIS AND MAST CELL ACTIVATION SYNDROME DIAGNOSTIC CHALLENGE

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Background Food-related anaphylaxis is common in a paediatric age group. Allergic reactions with multisystemic involvement can potentially point towards mast cell activation syndrome.

Case Presentation 6-year-old child initially presented with allergic reaction after eating strawberries. He developed lip swelling, breathing difficulties, itchy rash. Symptoms resolved with oral antihistamine and GP prescribed adrenaline autoinjector (AAI) with suspected anaphylaxis reaction.

At 8 years of age, he had an allergic reaction to orange fruit shoot with lip and tongue swelling, itchy rash. He responded well to oral antihistamine. Similar episode occurred few days later along with breathing difficulty and he received IM adrenaline in the hospital. He also had provocation test with orange fruit shoot and developed anaphylaxis requiring IM adrenaline, IV chlorphenamine.

Later, severe anaphylaxis reactions occurred in school, but there was no identifiable cause found. At one stage, required AAI once a week over 5 weeks period at school with no trigger. On few occasions there was no itchiness but only lip and facial swelling present. He also had chronic urticarial rash for which was prescribe oral cetirizine and was in control.

Once he developed urticarial rash after contact with latex tablecloth in school, after which plan made in place to avoid contact with latex cloth. At this point of time, he was referred to tertiary care centre for further opinion.

Over the next 2 years anaphylactic episodes resolved. Later, developed neurological symptoms of tremor and feeling of being disoriented. School noticed that, his concentration has been lacking. Eye symptoms with redness, itchiness and discomfort were developed.

Investigations Skin prick test showed positive 6mm, Peanut-3mm, Walnut-4mm, Hazelnut-4mm, Almond-3mm, Strawberry-3mm, House dust mite-2mm, Birch pollen- 3mm. Total serum

IgE is 268IU/ml. Specific IgE to Tomato- 0.02, Strawberry- 0.01, Kiwi fruit- 0.03, Peanut- 1.25, Hazelnut-0.97, Orange- 0.87, Latex-1.18, Birch pollen-1.15. Thyroid function test, MRI brain were normal. Investigations performed for Wilson's disease, which were unremarkable. Coeliac screen was normal. Hereditary angioedema screen is normal. There was no record of basal serum tryptase level.

Treatment Initial diagnosis of idiopathic anaphylaxis made. Later with new neurological and eye symptoms, diagnosis of mast cell activation syndrome suspected, and he was started on trial of oral sodium cromoglycate capsules.

In next clinic follow up, his symptoms were completely resolved and feedback from school was excellent in terms of concentration.

Discussion Symptoms of allergic disorders are very wide. Mast cell activation syndrome especially with multisystem involvement should be a differential to Idiopathic Anaphylaxis.

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CLINICAL, IMMUNOLOGICAL AND GENETIC FINDINGS IN 22 PATIENTS WITH COMBINED IMMUNODEFICIENCY TREATED IN A SPECIALIZED CENTER IN CROATIA

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To gain insight into the peculiarities of clinical presentation and time of diagnosis in the population of patients with combined immunodeficiency treated in our center.

We retrospectively analyzed 22 patients with combined immunodeficiency treated in our center in the past 25 years. Clinical findings were collected, including age of presentation and age of diagnosis, genetic defects, consanguinity, family history, number and site of infections, presence of diarrhea, failure to thrive, skin rash, vaccine complications and type of treatment. Immunological findings included WBC with absolute counts, lymphocyte subsets and immunoglobulin levels.

Patients (n=22) with severe combined immunodeficiency (n=17) and other combined immunodeficiencies (n=5) were included in the study. Sixteen patients (77%) were male. The most common clinical manifestations were opportunistic infections. Most of the patients were diagnosed after 3 months of age. Prenatal diagnosis was performed in one patient (4.5%), two patients

(9%) did not undergo genetic analysis. The spectrum of genetic defect in our cohort show wide heterogeneity. Seventeen patients (77%) underwent HSCT, one patient (4.5%) is on enzyme replacement therapy. None of our patients have undergone gene therapy. Twelve patients (54.5%) died, of which four (18%) died before HSCT.

Combined immunodeficiencies are fatal without definitive therapy (HSCT or gene therapy). The outcomes are better if infants are diagnosed and treated in pre-symptomatic stage, preferably before 3.5 months of age. More and more countries are implementing newborn screening for primary immunodeficiency diseases in their health care systems to diagnose patients as early as possible and to prevent infections. For the

same reason, we advocate for expanding our national neonatal screening program

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OMALIZUMAB FOR CHRONIC URTICARIA IN A PATIENT YOUNGER THAN 12 YEARS – CASE REPORT

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Urticaria is common condition in children and most cases are acute and self-limiting. Chronic urticaria in children is rare and defined with daily/almost daily presence of symptoms lasting for at least six weeks. It has two forms, chronic spontaneous urticaria (CSU) and chronic inducible urticaria.

CSU is usually considered as a disease of unknown origin but in some children an underlining autoimmune mechanism or atopic disease may be present.

Current treatment guidelines recommend elimination of revealed trigger factors, second generation H1-antihistamines and in unresponsive patients, ciclosporin or omalizumab (anti-IgE antibody).

Data about effectiveness and safety of omalizumab treatment in paediatric population are limited.

We present a 9-year-old girl with CSU who was successfully treated with omalizumab.

A 9-year-old girl presented with urticaria and angioedema of the eyelids lasting for 4 months. She was treated with H1-antihistamines without success. Her mother reported sun exposure, exercise, egg and milk consumption as trigger factors. Laboratory investigations including inflammatory markers, complete blood count, general biochemistry, immunoglobulin levels and protein electrophoresis were normal. Total IgE level was 15.9 kIU/L and specific IgE for milk and egg was negative. Thyroid function tests were pathologic, antithyroid antibodies were increased but not requiring substitute levothyroxine therapy. Thyroid ultrasound showed diffusely enlarged thyroid gland with a heterogeneous echotexture. She also had homogenous pattern of antinuclear antibodies on indirect immunofluorescence without other symptoms/signs of a rheumatologic disease.

Initial weekly Urticaria Activity Score (UAS7) was 28-30. Therapy with H1-antihistamines was continued with the four-fold increase in dosage without success. She also received systemic corticosteroids in acute flares. Further investigations revealed negative autologous serum skin test and positive basophil activation test. Due to unresponsiveness to H1-antihistamines, almost daily need for systemic corticosteroids and high UAS7, treatment with omalizumab was initiated. The therapy was started with 150 mg subcutaneously

(s.c.) every 4 weeks for 3 months. There was no expected improvement after four doses and her UAS7 was 30-36, the dose was increased to 300 mg s.c.

After two doses disease activity reduced and UAS7 was 14. Simultaneously, further impairment in thyroid function tests was observed and levothyroxine therapy was started. Omalizumab therapy lasted for 12 months without side-effects and with significant reduction in disease activity and UAS7, and with improvement in overall quality of life of the patient and her family.