

lesions was carried on: 5 polyps were removed from duodenum (max 2.5 cm) and 3 from jejunum (one “cauliflower” polyp 4 cm). An intraoperative ileo-enteroscopy was conducted and all ileum was investigated: no polyps were found.

Conclusion. Simone was an infant when first became symptomatic. Many studies suggest that a correct screening programme should be performed after 10 years of age, but in some patients PJS may present at a very young age and in a very aggressive pattern. For this reason our opinion is that even in first childhood, a careful gastrointestinal and oncologic follow up is mandatory in Peutz Jeghers patients to prevent complications (bleeding, bowel obstructions) and malignancies (pancreas, thyroid. . .).

A conservative surgical approach has been carried on because of the young age of our patient and because he will probably need other surgical treatments in the future.

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PA51

SHWACHMAN-DIAMOND SYNDROME IN A 9 MONTHS OLD GIRL: A NEW CASE OF UNUSUAL PATHOLOGY

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Background. Shwachman-Diamond Syndrome (SDS) is an autosomal recessive genetic disorder probably tied to multifactorial origin and often to mutation of SBDS gene.

The SDS is characterised mainly from exocrine pancreatic insufficiency, short stature, bone marrow deficiencies with neutropenia, and sometimes chemotaxis defects, anaemia, thrombocytopaenia, skeletal alterations, moreover hepatic and cutaneous interest. It involves a risk of evolution in mielodisplastic syndrome and acute myeloid leukaemia between the 8 and 33%.

Materials and methods. We present our regarding experience on a 9 months old girl affected from SDS with hepatic presentation. From the first months of life she had insufficient growth, neutropenia, dermatitis, dispepsia. At the income to the ward she presented 3rd centile lower increase, diffuse dermatitis, marked neutropenia, light anaemia and thrombocytopenia and hypertransaminase (AST 272 UI/ml, ALT 435 UI/ml, Gamma-GT 130 UI/ml), steatocrito 14%, faecal elastasis inferior to 20 µg/g, hepatosplenomegaly, hyperechogenus pancreas. In relation to persisting of elevated values of AST, ALT and Gamma-GT has been executed RNM abdomen that evidenced the presence of a solid expansive formation, compatible with inflammatory formation with miofibromatosis component. The patient therefore underwent to digestive RX study with negative outcome, and to

seriate abdomen TC that have shown the progressive volumetric reduction of the formation whose context was present multiple aerial images. Such report has been attributed to the presence of partially compacted internal handles compatible with the malnutrition state in which was the small patient.

As a result of the search of the genetic mutations for the SDS the patient has turned out carrier of mutations: 258 +2T/183–184ta > ct respective of paternal and maternal origin. There was a isochromosome of the long arm of the chromosome 7i(q10)c.

The patient underwent therapy with pancreatic extract.

Results. The small patient has shown height and weight increase attested after 2 months between 5th and 10th centile, didn't present infectious diseases, therefore it has not been necessary to execute GCSF treatment, also because the absolute values of the neutrophils have been maintained within 500/µl.

Conclusions. It is important to study this pathology even if rare, to obtain more accurate epidemiological knowledges, about the frequency and the clinical manifestations.

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PA52

VALIDATION OF THE DIAGNOSTIC ACCURACY OF A NEW PROTOCOL FOR FOOD CHALLENGE IN CHILDREN WITH SUSPECTED FOOD ALLERGY

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Introduction. Although controlled oral food challenge (FC) is considered the gold standard for the diagnosis of food allergy (FA), the interpretation of this test is sometime difficult especially in the presence of subjective signs and there is no consensus regarding the optimal procedure to objectively interpret the results. We have recently developed a protocol to perform FC in children with suspected gastrointestinal symptoms related to FA. This procedure is based on an objective evaluation of gastrointestinal signs before and after FC, and also includes non-invasive tests to assess intestinal inflammation and permeability.

Aims. To investigate the diagnostic accuracy of this new protocol for FC in children with suspected FA and gastrointestinal symptoms.

Methods. Children (age <36 m) with suspected FA consecutively observed in our Department were enrolled in a prospective study. The children were fully evaluated and all data were recorded in a special form. Intestinal permeability, faecal calprotectin and eosinophilic cationic protein levels were determined before and after FC.

Results. FC were performed in 45 patients (mean age 16.7 m, range 3–36 m, 28 male), 37 as open and 8 as double-

blind procedure. The most common food allergen was cow's milk proteins (82.2%). Data of each patient were collected from 4 days before FC and during the 7 days following the procedure. The results of the forms were interpreted by 3 physicians unaware to the patient's history. The protocol has a specificity and sensitivity of 100% and 83.3%, respectively. The positive and negative predictive values were 100% and 97.3%, respectively.

Conclusions. Our data showed that the new protocol to perform FC including an accurate registration of gastrointestinal symptoms and a objective evaluation of intestinal inflammation and permeability has a high specificity and sensitivity. This protocol could be helpful for the diagnosis and the follow up of children with FA.

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PA53

THE EFFICACY OF NUTRITIONAL INTERVENTION IN FOOD ALLERGY AFFECTED CHILDREN

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Background. We have previously demonstrated that an unbalanced diet with deficient caloric intake is a frequent feature of food allergy (FA) affected children, determining a significant impact on body growth.

Aims. To evaluate the efficacy of a nutritional intervention in paediatric patients on elimination diet because FA.

Participants and methods. Prospective multicenter controlled study including FA affected children (3–36 m of age) on elimination diet for at least 30 d, consecutively observed in 2 Tertiary Centers of Paediatric Gastroenterology and Allergy. For comparison healthy children were also examined. At the enrolment and after 2, 4 and 6 m the anthropometric indices (z-score for weight and z-score for length) were collected and dietary intake was assessed by a 3-day diary. The daily nutrient intake was analysed using a computer program (Nutriclick) based on the Italian food composition tables. A professional dietary intervention was then provided by a nutritionist and specialized paediatric dieticians.

Results. The study included 151 children. Of these, 86 (54 boys, median age 19.8, 95% CI 17.0–20.6 m) had FA, and 65 were healthy controls (41 boys, median age 20.5, 95% CI 18.0–23.0 m). Six FA children were lost at the follow up and were excluded from final analysis. Clinical symptoms of FA were gastrointestinal (80.2%, vomiting, chronic diarrhoea, and hematochezia), atopic dermatitis (54.7%), asthma (7.0%), acute urticaria and angioedema (4.7%) and anaphy-

laxis (1.2%). Seventy six patients presented allergy to cow's milk (CMA) (88.4%), alone or in combination with hen's egg (45.2%), wheat (5.8%), tomato (5.8%), fish (8.1%), soybean (6.1%), rice (3.5%); the 10 patients without CMA were allergic to hen's egg (5), soybean (2), fish (2) and rice (1). The mean length of the elimination diet was 10.8 m (8.5–13.1, 95% CI). At the enrolment, FA-affected patients presented a median z-score for weight (−0.48 vs 0.45, $p < 0.001$) and for length (−0.82 vs 0.65, $p < 0.005$) lower than healthy controls. Thirty out of 86 (34.8%) FA affected children presented at least one major dietary reference intake (DRI) below <85% compared to only 2 subject (3.0%) in the control group ($p < 0.001$). The only general exception was protein intake. Already after two months of controlled dietary intervention the DRI became normal in all subjects (average 101% of RCI, 97.0–105.4, 95% CI), the intake of proteins became more balanced and DRI remain stable on normal values up to 6 months of follow up.

Conclusions. Nutritional counselling is able to provide optimal nutrient intake and normal body growth and development in paediatric patients on elimination diet because FA. Our data should encourage the application of a paediatric medical nutrition protocol for FA affected children.

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PA54

HEPATIC PRESENTATION OF KAWASAKI DISEASE

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Objective. Kawasaki disease (KD), an acute systemic vasculitis of childhood, is diagnosed when 5 of the 6 principal features (fever ≥ 5 days, bilateral conjunctivitis, changes of lips and oral cavity, extremity changes, polymorphous rash and cervical lymphadenopathy) are present. Liver involvement does not belong to the classic diagnostic criteria of KD and includes mainly symptomatic or asymptomatic gallbladder hydrosis and in very few cases intrahepatic cholestasis of acute onset. We report here two children admitted in the last 12 months with febrile cholestatic jaundice as presenting symptom of KD. The aim of this report is to provide a high index of suspicion of KD allowing to early diagnose and treat these patients with hepatic presentation to reduce the risk of developing coronary arterial disease.

Report of cases. First patient was a 1-year-old child with fever and scarlet-like exanthema who abruptly developed jaundice. Clinical examination revealed jaundice, mild bilateral conjunctival injection, dry cracked lips, pharyngitis and homogeneous hepatomegaly. Blood test showed leukocytosis ($14.1 \times 10^9/\text{mm}^3$ with 76% neutrophils), Hb 11, 8 g/dl, platelet count $306 \times 10^9/\text{mm}^3$, ESR 62 mm/h and evidence of cholestatic jaundice [total/conjugated bilirubin