Special Issue

Euromediterranean Biomedical Journal
for young doctors
(formerly: Capsula Eburnea)

ABSTRACT BOOK
ONSP DAYS 2014—11th ITALIAN MEETING OF NATIONAL RESIDENTS PEDIATRIC OBSERVATORY

Pavia (Italy) 17-20 September 2014

ISSN 2279-7165 - Euromediterranean Biomedical Journal [online]

Vitiligo after bone marrow transplantation for SCID

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Allogeneic bone marrow transplantation (BMT) is a well-established therapy used for the treatment of an expanding number of pediatric diseases. One of the main complications of BMT is chronic *graft-versus-host disease* (c-GVHD). The major target tissues of c-GVHD include skin, gastrointestinal tract and bile ducts. Autoimmune disease may occur after BMT in absence of c-GVHD as a result of an imbalance between autoregulatory and autoreactive lymphocytes. Vitiligo (V), an autoimmune condition characterized by selective destruction of melanocytes, has been reported to be associated with c-GVHD. We report an 18 year old boy who developed perioral V with progression to axillary folds and legs 12 years after BMT performed for SCID T-B-NK+ at the age of 3 months. He received a transplant from HLA identical sibling donor without conditioning with complete immune reconstitution. No complications in the immediate post-BMT period and in the long term follow-up occurred before the appearance of V. The donor is still otherwise healthy without history of autoimmune or skin conditions. At time of examination, the patient had normal blood cell count, metabolic panel, liver function and thyroid tests. No other deregulation of immune system has been found.
Our case is interesting due to the fact that V occurred without c-GVHD, so far not reported in the literature. It could be considered an isolated manifestation of c-GvHD or an *ex novo* late onset of autoimmune disease after BMT.

####

A child with a swollen hand…

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A 7 year old boy who had been stung by a an honeybee about 24 hours before, presents with swelling, pain and itching extending from the sting site, on the left hand. The symptoms started a short time after the sting and progressively increased. He had been previously stung by honeybees without any reaction. He had no history of allergy or past medical problems. He was afebrile. A remarkable angioedema of the left hand involving also the left wrist was evident, with *rubror*, *calor* (except for the area of the sting where topical corticosteroid had been applied) and normal consistency of the skin. Mobility of the hand and fingers was limited by pain and oedema. Secretion was absent. A diagnosis of *large local reaction* (LLR) to hymenoptera venom was established. He was treated with oral corticosteroids and antihistamines for a few days. 4 days later the symptoms had almost completely disappeared.
LLRs is a frequent clinical manifestation of hymenoptera venom allergy (HVA) in children. LLRs cause pain, itching, swelling and erythema extending from the sting site, resembling a cellulitis, usually peaking at 24–48 h and resolving within 5–10 days. LLRs tend to decrease with time and the risk of a systemic reaction if restung is very low, hence neither diagnostic test for HVA or immunotherapy are necessary. Cold compresses, oral antihistamines and oral or topical corticosteroids for a few days are generally used to control symptoms.

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A child with rough nails…

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CASE REPORT: a healthy six year old boy presents with nail dystrophy for several months. On examination the nails appeared rough with ridging and loss of luster. There were neither muco-cutaneous lesions nor other clinical signs. Past clinical

history was silent and familiar anamnesis was positive for vitiligo. The clinical picture was in keeping with a diagnosis of idiopathic Twenty Nail Dystrophy (TND) of childhood. With a three-week course of topical periungual corticosteroid there was a significant improvement which persisted to the last examination, one month later.
DISCUSSION: TND or trachyonychia is a disease of the nail matrix characterized by excessive ridging and roughness of the nail. TND can be idiopathic or associated with autoimmune disorders such as alopecia areata, lichen planus, psoriasis, and vitiligo. Idiopathic TND is the most common form among children with a peak incidence at 3-12 years of age. The histopathology shows spongiosis and exocytosis of the inflammatory cells into the nail epithelia. Nail biopsy is not necessary when idiopathic TND is suspected because itis a benign condition with tendency to spontaneous resolution without scarring. There is no evidence-based therapy for idiopathic TND. Considering the favorable outcome, simply reassuring parents can be enough. As an alternative, a trial with periungual topical corticosteroids aimed to reduce the inflammation of the nail matrix may help, as in the case reported here.

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A dangerous banana

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Peter first came to our attention when he was 6 months old,he had never shown health problems before. He was brought by his family to the Emergency ward because of projectile vomiting and somnolence a few minutes after eating a carrot, courgette and millet mash. He had eaten one banana one hour and a half before. When he arrived at Hospital he had diarrhea and he presented with lethargy, hypotonia and pallor. He had worsening hypotension and hypothermia. No other clinical alteration was revealed. As a result of his deteriorating condition, he was given epinephrine and corticosteroid ev therapy. Liquid intravenous infusion was started with a good response. Blood tests showed thrombocytosis, leucocytosys, negative RCP and regular liver and kidney function. Echocardiogram and stool tests were normal and prick tests werepositive for egg proteins (that were later introduced without problem). Patch tests were positive for banana after 72 h. The infant was breast fed and banana was excluded from his diet. After an in-depth interview with the parents, 2 episodes of vomit after the only 2 attempts of giving some banana to the infant were discovered. Clinical history together with laboratory and allergy tests strongly suggested the diagnosis of acute FPIES. FPIES is an under-recognized non-IgE -mediated food hypersensitivity disorder with challenging differential diagnosis. This is the first severe case elicited by the ingestion of banana.

####

A case of retropharyingeal abscess (RPA)

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Livia is a one year old baby with high fever, sore throat, rhinitis, neck swelling and laterocervical adenomegaly, who arrived at the Pediatric Emergency Room in Pavia. The laboratory exams showed elevated inflammatory parameters and the US of the neck revealed colliquative necrosis of local lymph nodes.
She was immediately admitted and treated with intravenous antibiotics (ceftriaxone). The worsening of clinical conditions precipitated the performance of a CT, showing evidence of RPA, which was treated with transoral surgical approach. Microbiological culture detected Staphylococcus aureus infection which was further treated with teicoplanina.
After a few days, there was a good improvement in general conditions, con-

firmed by the CT..
Retropharyngeal abscess (RPA) is an uncommon complication of upper-respiratory infections in children resulting from spread of infection to and eventual suppuration of retropharyngeal lymph nodes. The diagnosis is based on the use of CT. The patients can be managed with intravenous antibiotics or with surgical intervention, if necessary. Complications of RPA are uncommon, but there is a high correlation between MRSA infection and mediastinitis. Many authors have reported a marked rise in the incidence of RPA over recent years, so it could be considered an emerging disease. According to the last classification of head and neck masses, RPA must be distinguished from neoplastic, congenital and other inflammatory masses, and first of all from epiglottitis.

####

When medical therapy is not enough: a pediatric case of osteomyelitis

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Most cases of long-bone osteomyelitis are post-traumatic or postoperative. Culture-specific antibiotics play a major role in decreasing the incidence of acute osteomyelitis. Surgical debridement, wound irrigation, and muscle-flap or vascularised tissue grafts play major roles in prevention and treatment by removing dead tissue, decreasing bacterial load, and filling dead space with vascularised tissue. Internal fixation of contaminated dead bone inevitably leads to osteomyelitis and must be avoided.
An 11 year old underwent osteotomy of the left femur, with placement of an external fixator and bilateral tibial epiphysiodesis plaques due to a congenital discrepancy in limb-length (right > left), attributed to the reduced growth of ¾ left ipsilateral femoral and tibial ¼.
During rehabilitation physiotherapy, G. reported a pathological fracture to the left femur. For this reason, he underwent surgery again for internal fixation with plate and screws and release, according to Judet.
Following the onset of fever, asthenia and suppuration of the wound, G. was brought to the attention of the Department of Orthopaedics at our hospital.
G. was subjected to femoral NMR and CT scan, which detected a loss of bone substance as a result of extended osteolysis and persistent inflammatory-necrotic tissue associated with cutaneous fistula on the external side of the middle third. Diagnosis of osteomyelitis of the femur, stage 4 according to Cierny-Mader classification (diffuse osteomyelitis involving the entire thickness of the bone, with loss of stability, as in an infected non-union).
The child underwent three revision surgeries, surgical wound debridement and placement of VAC-therapy at the cutaneous fistula ducted up to the bone surface below. Intra-venus antibiotic therapy with oxacillin (170 mg / kg / day) and cotrimoxazole (30 mg / kg / day) was continued for 8 weeks.
Bone samples were intraoperatively collected and they were found to be positive for Staphylococcus Aureus producing beta-lactamase and Enterococcus faecalis. Cutaneous swabs, collected weekly, were negative.
It was the third surgical intervention which lead to the radical removal of the outbreak and resolution of the wound and subsequent formation of callus.

####

Wait and watch approach in a pediatric case of Langherans cell histiocytosis.

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A 14 year old boy was admitted to our emergency room because of two painless tumefactions localized in the under-chin region. He was afebrile and did not have any other systemic symptom. Clavulanate amoxicilline treatment did not have any effect. A soft tissue ultrasonography revealed reactive lymph nodes with a maxi-

mal diameter of 3,5 cm, without internal colliquation or vascular alterations. Laboratory evaluation revealed mild leukocytosis, with eosinophilia, no signs of inflammation, normal thyroid function, previous EBV immunization and other negative viral serologies. Pediatric infectious and otolaryngologist evaluation did not reveal any disorder. Neck MRI showed two oval tumefactions with regular borders, without necrosis, surrounded by other smaller reactive lymph nodes. He underwent excision biopsy that revealed overthrown architecture determined by polymorphic cellular infiltrate. Immunostaining revealed that these cells, accompanied by abundant eosinophils, were positive for CD1a and Langherina, S100, supporting a diagnosis of Langherans cell histiocytosis (LCH). For this reason, following the International Collaborative Treatment Protocol for Children and Adolescents with LCH, the child underwent a staging based on total body X-ray, abdomen ultrasonography and positron emission tomography that excluded other LCH localizations rather than the lymph nodal ones. He was signed up to the LCH VI Protocol as single-system LCH (SS-LCH) and he started a clinical and instrumental follow up, without taking any treatment. After a period of 9 months, he is in good health, without signs or symptoms of systemic diseases, and has not developed any other lesion. Patients with SS-LCH of the lymph nodes, skin or skeleton have an excellent prognosis and sometimes need no treatment at all.

####

Herpes Simplex Virus reactivation manifested through Stevens Johnson-syndrome.

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S. is an 8 year old healthy boy. In January 2012, he presented herpetic stomatitis; given antiviral therapy with acyclovir, an erythema multiforme appeared. In the next few months, S. presented three episodes of herpes labialis. In October 2012, vescicular stomatitis reappeared, associated with aphthosis, gingival hyperemia and hemorrhagic erosions of mouth mucous membranes, and was treated with oral acyclovir. The medical case evolved with the presentation of worsening oral lesions, and consequent difficulties in feeding and hydration, followed by the appearance of erythematous-papular lesions on the trunk and limbs. Therefore, acyclovir was continued intravenously; a recurrence of erythema multiforme was clinically identified and confirmed by skin biopsy. After additional clinical deterioration, steroid therapy was introduced. Then, Stevens Johnson-syndrome (SJS) due to HSV reactivation was clinically diagnosed due to further worsening of the skin lesions, gradually evolving into vesicles, and of stomatitis with labial and lingual edema. Improvement until complete resolution of lesions was achieved after immunoglobulins infusion. All microbiological tests performed were negative.
SJS is an emergency, characterized by erythematous skin, extensive (limited to less than 10% of BSA) detachment of the epidermidis and hemorrhagic erosions of mucous membranes; it is principally caused by drugs or infections.

####

Weight, height, saturation and…arterial pressure: don't forget this parameter!

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P.G. is a 20 month old boy, who came to our attention with cough without fever, vomiting and dehydration. In our emergency room, he expereinced mild tachypnea and metabolic acidosis. Intravenous hydration was started with the onset of dyspnea,desaturation and rapid clinical deterioration. Chest x-ray showed edema and cardiomegaly. Blood exams indicated an increase in cardiac enzymes. The patient's blood pressure value was persistently elevated (230/100mmHg), so he underwent echocardiography which revealed a significant left ventricular hypokinesia and dilatation, associated with mild hypertrophy, without evidence of coarctation. He was admitted to the intensive care unit for intubation and to start intravenous antihypertensive therapy. Four drugs were necessary to stabilize blood pressure between 170/90 and 150/75mmHg. After 8 days, blood pressure value reached 130/70mmHg, and he started oral medication. The whole-body CT, PET, fundus oculi and urinary catecolamines levels were normal. Viral and bacterial serologies and tests for autoimmune diseases came back negative. Endocrine tests and serum electrolytes showed normal values. The renin level was elevated, the Captopril test resulted positive. Our hypothesis was renovascular hypertension, probably caused by renal small-vessels stenosis, not visible with traditional imaging. The last echocardiography showed a mild hypertrophy of ventricular septum and currently, he has good blood pressure control with a 3 drug oral therapy.

####

Carbapenem-resistant Klebsiella pneumoniae infection in a patient treated with haplo-identical hemopoietic stem cell transplantation.

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Carbapenem-resistant Klebsiella pneumoniae (CR-KP) is a multidrug resistant pathogen. In hemopoietic stem cell transplantation (HSCT) recipients, colonization/ infection by CR-KP is associated with increased therapeutic failure and mortality.
We describe a 14 year old boy with a resistant form of Acute Myeloid Leukaemia (M4) eligible for HLA-matched unrelated donor HSCT. Since a CR-KP was detected in urine and from perianal cellulitis and considering the high risk of sepsis-related death, the HSCT was postponed in order to clear the infection with a 4 antibiotic therapy.
Unfortunately, leukaemia progressed rapidly and with the aim of avoiding any further delay in transplant, we proceeded with a haplo-identical HSCT from the mother. The procedure was followed by primary graft rejection and thus, after 1 month, a second haplo-identical HSCT from the father was performed. Full donor engraftment was regularly achieved and maintained together with cytogenetical remission 2 months after transplant.
Overall, the neutropenia lasted about 60 days and during hospitalization strict environmental isolation was maintained. The poli-antibiotic therapy was initially administered in 3 febrile neutropenic episodes pending evaluation for possible CR-KP reactivation, and suspended in the absence of microbiological isolation (de-escalation approach).
Comments: strict environmental surveillance and cautious management reduced the risk of severe transplant complications.

####

A challenging case of "Red Eye". Acute anterior uveitis as the first manifestation of Juvenile Systemic Lupus Erythematosus.

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"Red eye" is an ordinary event in pediatrics. Most cases are infectious in origin and can be easily treated. However, diagnosis may sometimes be challenging, as it can be the first manifestation of severe systemic dis-

eases.
We report the case of an 11 year old girl, presenting at our Clinic with a diagnosis of Vernal Keratoconjunctivitis. She had had red and painful eyes associated with tearing, photophobia and mild visual loss for one month. Ophtalmological evaluation with slit-lamp led to a diagnosis of acute anterior uveitis. Clinical examination also revealed oral aphthosis, nasal mucosal ulceration and low-grade fever. Differential diagnosis work-up excluded infectious uveitis, celiac desease, IBD as well as JIA and systemic autoimmune diseases. We defined the case as "Idiopathic Uveitis" and started local corticosteroid treatment. Two months later, she was readmitted for thoracic pain: biochemistry showed neutropenia and chest X-ray evidenced exudative pleuritis. Considering the presence of Anti-Nuclear and anti-DNA antibodies, Juvenile Systemic Lupus Erythematosus (JSLE) was finally diagnosed and appropriate systemic therapy started.
Red eye is frequent in pediatrics but misdiagnosis is also frequent and may lead to inappropriate treatment. Anterior uveitis is an uncommon cause of red eye in children, and often has an infectious etiology. Exceptionally, anterior uveitis can be the first manifestation of JSLE, as in the reported case.

####

Epidemiology characteristic of respiratory viruses in pediatric ospedalisation for respiratory tract infection

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Respiratory tract infection (RTI) is the most common problem in paediatric clinical practice. Rate of admissions for RTI is often much higher than predicted.
To compare the demographic characteristics, viral prevalence, age distribution, and seasonal distribution of paediatric patients with RTI admitted to our hospital.
Retrospective review of 1359 children, under 16 years old, admitted to our hospital from 1 January 2013 to 31 December 2013. Nasopharyngeal swabs were collected according to a standard procedure in 526 of them, to test forseveral respiratory viruses and bacterial infections (influenza virus, parainfluenza virus, respiratory syncytial virus, adenovirus, human metapneumovirus, human coronavirus, human bocavirus, enterovirus, rhinovirus, and b. pertussis e parapertussis, l. pneumofila, m. pneumoniae, c. pneumoniae, s. pneumoniae, h. influenzae).
Viral prevalence varied by season, being more prevalent in winter (47%), and spring (28%), and less in autumn (16%) and summer (9%).
In all four seasons, the detection rate of rhinovirus was the highest of all respiratory viruses tested, followed by parainfluenza and influenza viruses.
To analyse the age distribution of the viral infection, we organised the children into three groups: aged <1 years old, 2-5 years old, and >6 years old.
This study contributes to the knowledge of seasonal variation of respiratory viral infection in southern Italian hospitals.

####

Incidental radiological diagnosis of portal cavernoma in a teenager with abdominal pain: a case report

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C. was born at 40W of gestation and received neonatal umbilical vein catheterization, due to severe respiratory distress.
She is now a 14 y.o. healthy looking teenager. In the last 2 months, she has been suffering from asthenia and bilateral abdominal recurrent pain, unrelated to menstrual cycle. Regular bowel function.
Physical examination: soft abdomen with left hypochondrium tenderness. Spleen palpable at left costal arch.
Normal liver blood tests. Iron deficiency

anemia (Hb 11.2 g/dl, MCV 70 fl; Ferritin 5.5 mg/l) in treatment with oral iron supplementation.
Abdominal US (*Image 1*): multiple periportal anechoic vascular formations.
Abdominal CT (*Image 2*): numerous periportal venous ectasia (43x27mm) consistent with a portal cavernoma. Threadlike caliber of the left portal branch. Regular right portal branch. Reduction of left hepatic lobe perfusion, without evidence of collateral circulation. Mild splenomegaly (129 mm).
EGDS (*Image 3*): slightly hyperemic antral mucosa, no peri-esophageal or gastric varices.
Conclusions: C. has an incidental diagnosis of cavernous transformation of the portal vein with a mild extra-hepatic portal hypertension, which only needs follow up. Should it worsen in future years, a retrograde portography will be needed to verify the feasibility of a meso-Rex by-pass [interposition of a vascular graft between the Mesenteric Vein and the Rex recessus (Left Portal Vein system)] to restore a physiological hepatopetal flow.

####

A "creeping" disease

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B.R. was hospitalized at 18 days of life due to lethargy, muscular hypotony, and poor feeding; after a seizure, he was given a lumbar puncture, and he started empiric antibiotic therapy with ampicillin, gentamicin, and cefotaxime. Considering the results of CSF exam (180 cells/mm3, lymphocytes 62%, negative Gram coloration), and suspecting a viral infection, acyclovir therapy was also started (dosage: 60 mg/kg/d iv). The following day, the result of PCR showed the presence of 240 copies of HSV-2 DNA, confirming the diagnostic suspicion of herpetic encephalitis.
Herpetic encephalitis (30% of total HSV infections) usually occurs at 16-19 days of life, presenting with lethargy, irritability, tremors, poor feeding, seizures, bulging anterior fontanelle, temperature instability and cutaneous lesions. Despite the fact that the introducion of specific antiviral therapy, has decreased mortality from 50% to 4%, there have been no differences in the incidence of neurological sequelae.
The most common collateral effects related to the use of acyclovir are neutropenia and nephrotoxicity, both of which can be easily monitored.
Considering the risk of death and neurological damage, and based on evidence from the literature,, it was decided that the balance between risk and benefit could justify a higher dosage of the drug to guarantee adequate exposition. For this reason the acyclovir dosage was raised to 90/mg/kg/d. The intravenous administration was suspended after 21 days, and suppressive therapy with oral acyclovir was continued for six months.

####

The "fire" of FIRES (febrile infection-related epilepsy syndrome)

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Fires is a catastrophic epileptic encephalopathy with a yet undefined etiology. The onset age is from 4 to 15 years. It has an annual incidence of 1: 10^6.
The early clinical course is frequently biphasic: a febrile infection (mainly consisting of URTI or gastroenteritis, without any

identified infectious agent) and, after 2-14 days, subsequent acute onset of refractory seizure. The disease develops into Status Epilepticus, requiring intensive care management. Acute seizures are resistant to treatment with a variety of antiepileptic drugs and immunomodulatory agents.
The prognosis is poor, with a death rate of up to 30%.
We present the case of a 6 year old male, with no relevant personal or family history; he was born after normal pregnancy and delivery, and he also had normal acquisition of psychomotor developmental milestones.
In the week prior to admission, the patient experienced a flu-like illness with fever, myalgia and headache. Within a week, he developed a partial seizure secondarily generalized, evolving into status epilepticus, which required treatment with a high-dose of barbiturates and propofol accompanied by mechanical ventilation for 20 days. The immunologic and metabolic evaluations were always silent.
The EEGs showed a progressive and definitive electrical-disorganization.

####

A case of Pelvic Inflammatory Disease caused by Chlamydia in an adolescent

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We report a case of Pelvic Inflammatory Disease (PID) by *C. trachomatis* in a 13 year old girl who presented with worsening abdominal pain, in the lower left quadrant, and fever. She was admitted into a surgical ward where treatment with amoxiclavulanate was started with no clinical improvement. Although medical evaluation and USs were inconclusive, an abdominal CT scan showed peritoneal thickening with adhesions of uncertain interpretation. The patient was transferred to our tertiary paediatric surgery where a laparoscopy showed inflammation of müllerian structures and ovaries and confirmed the intestinal adhesions. The suspicion of a Sexually Transmitted Infection (STI) was given. A deeper personal history was then collected and the patient who was found to have been sexually active for a few months, had also had -unprotected intercourse. Microbiological examinations of vaginal discharge performed before the admission detected a *C. trachomatis* infection, both in our girl and her friend with whom she had shared a partner. Doxycyline and Cefoxitin therapy was started and a progressive improvement was seen both clinically and in US appearances of müllerian structures and ovaries. The partner and friend were treated with a single dose of azithromycin for uncomplicated infection. The age of becoming sexually active has been decreasing over the last decades; therefore, the possibility of STIs and their complications, must be borne in mind even in paediatric practice, especially when it comes to the differential diagnosis of abdominal pain or other genito-urinary symptoms.

####

An atypical lung opacity

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Klaudio, a 5 year old boy, was evaluated for fever and persistent cough, in spite of antibiotic therapy. Laboratory examination showed mild anemia (already known in his medical history and treated with oral iron therapy), mild lymphocytosis and increase in IgE (823 kU/l). Chest X-ray revealed a pulmonary consolidation in the right lower lobe, as a round pneumonia. He had moderate splenomegaly, confirmed by ultrasonography which showed a 6 cm round cystic lesion. Abdominal CT showed a splenic fluid mass without solid components, with a thick, hypoechoic outline and no contrast enhancement. Therefore, chest CT was performed which revealed the liquid content of the lung opacity. Serological examinations were negative for *Bartonella hensale, Chlamydia pneumoniae, Mycoplasma pneumoniae* and *Aspergillus spp*, while *Echinococcus granulosus* infection was detected by hemagglutination, ELISA and immunoblotting assays. The diagnosis was confirmed by the presence of *Echinococcus* protoscolex in the cyst fluid. Finally, there was no evidence of infection in

the other organs. CT of the brain showed no cerebral cysts. The treatment was surgical (echo-guided aspiration of the splenic cyst and thoracoscopic resection of the pulmonary cyst) and pharmacological (albendazole), with complete resolution of the pulmonary involvement. Klaudio is now being followed up by a team ofRadiologists, an Infectologist, Surgeons and Pediatricians.

####

Emergency differential diagnosis in a girl with fever, convulsions and altered conscious state.

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Convulsions associated with fever and altered conscious state are the most common neurological disorder of the paediatric age in Africa. When facing this clinical presentation, itis mandatory to quickly take in to consideration the main differential diagnoses: febrile convulsions with intercurrent illness, central nervous system infections (meningitis and encephalitis), hypoglycaemia or electrolyte disturbance of any cause, cerebral malaria (CM) or poisoning by indigenous drugs. Awa, a 4 year old girl, was admitted at Ziguinchor Hospital for fever and convulsions; she presented with high fever (40°C), lethargy and slight neck stiffness. Blood count and culture, RCP, electrolytes and glycaemia, lumbar puncture, malaria rapid diagnostic test and thick blood film were immediately performed (Hb 8g/dl, platelets 115.000/ mm3, RCP 12mg/dl, glycaemia 65mg/dl, clear CSF with normal pressure and thick film test positive for malaria). Since malaria rapid test was positive, an antimalarial agent (intravenous quinine) was immediately started. She rapidly improved, with complete symptom resolution after 3 days of treatment; negativity of blood and CSF cultures allowed us to confirm the suspected diagnosis of CM. This pathology is an acute encephalopathy which is a complication of infection by *Plasmodium falciparum,* commonly affecting children in hyperendemic areas. Considering its high mortality rate (up to 50%) and the residual neurological abnormalities, an early diagnosis and prompt therapy are essential.

####

Iron deficiency without anemia in children at time of diagnosis of celiac disease

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Celiac disease (CD) causes various hematologic disorders in children. Iron deficiency anemia (IDA) is the most common one, varying from 15 to 46% in patients (Pt) with CD. To the best of our knowledge, there are no studies concerning conditions of iron deficiency without anemia (IDWA). In this regard, we retrospectivly investigated a cohort of pediatric Pt at diagnosis of CD for anemia and iron imbalance. We enrolled 400 children at diagnosis of CD, 268 F and 132 M (mean age 6y/2m, range 6m-18y), admitted to our Pediatric Unit (Jan 1990-Jan 2012). IDA was defined as Hb<-2SD of the normal values and serum ferritin (SF)<12 ng/ml; IDWA was defined as SF<12 ng/ml and normal Hb. 119 Pt (29,75%) were anemic, 77 F (19,25%) and 42 M (10,5%); among them 94 presented IDA, 6 IDA associated to heterozygous β-thalassemia and 19 heterozygous β-thalassemia with normal SF. IDA was observed in overall 100 Pt (25%), 79 of them less than 6 year old. IDWA was present in 81 Pt (20,25%), 64 of them less than 6 years old. A total of 181 (44,25%) children showed iron store depletion. Our study demonstrated that iron imbalance is very frequent at the time of CD diagnosis. About ¼ of them had IDA and, of note, IDWA was present in another 20,25%, demonstrating that many children, newly diagnosed as celiac, already have an iron store depletion. These data underline the need for further evaluation of Pt with unexplained low SF, irrespective of the presence of IDA, for a precocious diagnosis of CD.

####

The use of Sirolimus in Lymphangioma-

tosis and Cystic Hygroma: our experience.

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L.Z. was born at 37 weeks + 5 days of G.A. by spontaneous delivery (Apgar 9-10) in Bisceglie's Hospital. She was transferred to our ward a result of detection of a laterocervical mass. She was in good clinical condition except for an expiratory groan. During hospitalization, she underwent a neck' ultrasound that showed the presence of left laterocervical lymphangioma and surgical consultation gave no indication for surgical treatment.
The patient was discharged at 5 days of life and started outpatient follow-up. Three days afte , she was admitted to our unit again for persistent cough and breathing difficulties. On admission, the patient appeared hypotonic and weak, with faint crying, fever, pain resulting from cystic hygroma and difficulty in breathing (retractions, breath sounds, SatO2 <80%). Consequently, she began oxygen in incubator (carried out for two days) also high flows for 24 hours and nCPAP for additional 48 hours, and started antibiotic therapy with Meropenem and Vancomycin for 7 days.
Blood tests revealed leukocytosis (30110 / mm3 with 88% neutrophils), and a high level of CRP (35.2 mg / L); normal level of VMA (0.30 mg / 24h), high level of alpha-fetoprotein (AFP) (2638.9 ng / mL), high level of enolase (22.3 microg / l) and normal standard karyotype (XX).
These symptoms indicated diffuse lymphangiomatosis. For this reason, at 33 days of life, she underwent NMR (Nuclear Magnetic Resonance) (neck-thorax, abdomen, skeleton) which confirmed the suspected diagnosis.
Multiple cysts were localized at the level of the neck, (max length about 6 cm),other cysts were localized at the level of Mediastinum, in the "Brety's loggia", in the third segment of liver and also at the skeletal level. (millimeter areas referable to cysts.)
At 34 days of life, she began therapy with propranolol, beginning at a dose of 2mg / kg twice a day, and after 9 days at a dose of 4 mg / kg once a day. As she did not respond to the drug, at 45 days life, she began therapy with sirolimus, first at a dose of 0.8 / mg / m2 twice daily and after 9 days at the dose of 0.4 / mg / m2. This therapy was ongoing with progressive improvement in the appearance and clinical ultrasound.
At 75 days of life, after 30 days of therapy, she repeated NMR (neck-thorax-abdomen-skeleton) which highlighted the volumetric regression of cervical, hepatic and mesenteric lesions.
At 76 days of life she was discharged in good condition, and started outpatient checks.She repeated blood tests (blood level of sirolimus, liver function, albumin, total protein, electrolytes, glucose, cholesterol, HDL, LDL, LDH, blood count, CRP, and urinanalysis) every month. .
Outpatients detected anemia (possible side effect of sirolimus therapy), which was treated with iron therapy.
Ecography, repeated every month, showed the full regression of the lesions. She had a cardiac evaluation that was normal, and at nine months of life, she performed a new NMR (neck-thorax-abdomen-skeleton) .
She continues with bi-monthly controls (blood tests and ecography). She also continuous therapy with sirolimus, dose established through blood level of sirolimus (between 8-10 microg / l in the first year of life and between 6-8 microg / l in the second year).

####

Cerebral hemorrhage during chemotherapy in a patient with acute lymphoblastic leukemia

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The clinical case presented is a 13 year old female patient who came to our observation for fever and cough. Chest radiographs showed lung condensation with consensual pleural effusion. By execution of routine blood examination and bone marrow aspiration, acute T-cell lymphoblastic leukemia was diagnosed and chemotherapy was started. During the induction phase of chemotherapy, there

was the sudden onset of confusion, aphasia, right hemiparesis and pale skin. A neuro TC-scan performed in emergency showed an intraparenchymal hematoma with slight shift of the ventricular system respecting the midline. So the patient underwent neurosurgical treatment. Since then a close neurological follow up, logopedy and physiotherapy have been started with the purpose of improving neurological outcome of cerebral hemorrhage. This case teaches us that we must not underestimate the potential bleeding complications of chemotherapy, which are rarer than thrombotic complications, which are already rare in themselves.

####

Elevation of serum creatine kinase as the main manifestation of Duchenne and Becker muscular dystrophy

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Background: Duchenne and Becker muscular dystrophy (DMD/BMD) are X-linked diseases resulting from a defect in the dystrophin gene located on Xp21. Although deletions in the dystrophin gene represent 65% of mutations in DMD/BMD patients, a clear genotype-phenotype correlation is not well-defined. We report a 12 year old boy who presented with muscle cramps and increase in serum CK (creatine kinase) caused by DMD/BMD.
Case report: The 12 year old boy was admitted to our pediatric unit because of recurrent abdominal pain with sporadic leg cramps. Laboratory evaluation showed fluctuant elevated CK, LDH and AST (maximum values of 1584 UI/L, 566 UI/L and 92 U/L, respectively). Neurological examination, muscle power and gait were normal. The Gowers' sign was negative. Electrocardiography (ECG) showed a short PR-interval (85 ms). Elettromiography (EMG) was normal. Muscle biopsy (MB) revealed nonspecific myopathic alterations. Immunohistochemistry for dystrophin showed normal pattern and intensity. DNA analysis of the dystrophin gene detected a deletion within the central rod domain involving the exons 48-51. The same mutation was detected in the mother.
Conclusions: Spectrum phenotype of DMD/BMD may be variable with no genotype-phenotype correlation, thus leading to a misdiagnosis. Therefore, molecular analysis of the dystrophin gene should be performed in the differential diagnosis of elevated CK, even in the presence of normal EMG and MB. Cardiological follow-up is needed.

####

A strange back pain

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Case report
A 14-year old boy complained of persistent lower back pain after a low-energy sport trauma. Back pain was localized at L5-S1 with a positive Lasegue sign on the right side. Areas of paresthesias and sensory loss followed the dermatomal distribution of S1 on the right side; achilles reflex was diminished on the left side.
MRI scan of lumbosacral district was performed, showing a T2-weighted hypointense and a T1 and FAT-SAT-weighted hyperintense signal area, compressing the dural sac. According to the clinical history, this finding was consistent with an epidural hematoma. Oral steroid therapy was started, without significant improvement.
CT scan confirmed the previous findings but raised questions about the nature of the lesion.
Laboratory tests revealed myeloblasts in the peripheral blood. Bone marrow aspiration showed a pattern compatible with acute myeloid leukemia AML-ETO positive with leukemic meningosis and extranodal localizations.
Discussion
Back pain in childhood is an uncommon situation, which deserves further diagnostic evaluation if symptoms persist; neo-

plasm should be considered in the differential diagnosis.
Myelocytic sarcoma is rare even related to AML or myelodysplatic syndromes. Imaging is helpful for diagnosis but misinterpretation of its findings is frequent. Bone marrow may confirm the diagnosis, but myelocytic sarcoma cannot be excluded if BM is normal. This may appear months or years before bone marrow involvement.

####

Dangerous breastfeeding

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We describe the case of a previously healthy 16 monthold girl presenting with a 1-month history of pallor, fatigue, anasarca and vomiting.
She was the first daughter of vegan parents, born at 39 weeks, after an uneventful pregnancy. The child had been exclusively breastfed up to the age of 16 months. Weaning had not yet been started. Normal development was observed, but she couldn't walk without support because of peripheral oedema.
Laboratory findings showed severe megaloblastic and iron deficiency anemia (Hb 5g/dl), hypoalbuminemia (16g/l) and vitamin B12 deficiency. Urine analysis documented no proteinuria.
We performed blood transfusion, iron and albumin infusion, and then we successfully started weaning the child. The girl presented progressive reduction of peripheral oedema and improvement in laboratory values.
Organic causes of malabsorption like celiac disease, cystic fibrosis and renal problems were excluded.
In this case anemia and hypoalbuminemia were caused by malnutrition due to prolonged exclusive breastfeeding and to delayed weaning.
Malnutrition is a very common problem in the developing world, but can be seen also in developed countries. Paediatricians should keep this problem in mind especially where parents are on a restricted diet, such as vegans.

####

Acute cerebellitis caused by Mycoplasma pneumoniae

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We describe a patient affected by acute cerebellitis caused by Mycoplasma pneumoniae.
A previously healthy 7 year- old boy presented a three day-history of ataxia, vomiting and altered behaviour, after mild respiratory symptoms that occurred almost 7-10 days before.
Ocular fundus and brain magnetic resonance were performed and showed no significant alterations. Serum titers for Mycoplasma antibodies indicated a recent infection. A thoracic ultrasound showed a pneumonia affecting the lower lobe of the left lung. The boy underwent antibiotic treatment with azytromicin for seven days, and showed complete resolution of both neurological and respiratory pattern.
Mycoplasma pneumoniae is generally responsible for upper and lower respiratory tract infections in school-age children; however rare multisystemic manifestations of Mycoplasma pneumoniae infections can also occur, including central nervous system inflammation. The most common ones are meningoencephalitis, aseptic meningitis, polyradiculopathy, cranial nerve palsy, cerebellitis, ADEM (acute disseminated encephalomyelitis), transverse myelitis, and cerebrovascular thromboembolic events or a combination of these. Two mechanisms have been postulated to explain these neurological complications: Mycoplasma direct invasion of the central nervous system or an autoimmune mechanism caused by brain-reactive antibodies.
We describe a case report of acute cerebellitis caused by Mycoplasma pneumoniae which occurred a few days after a respiratory infection. Our patient presented a benign and self-limited course, but cases complicated by cerebellar atrophy have been described.

####

A case of dumping Syndrome in a patient undergoing oesophageal atresia surgery

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Introduction: Dumping syndrome (DS) is a cluster of non-specific symptoms that typically occur when the stomach is emptied too quickly and the small intestine is filled with undigested food.
Case description: A newborn girl was diagnosed with congenital oesophageal type 1 atresia, requiring oesophageal replacement by gastric transposition. She was jejunally fed for the first 3 months after surgery and then oral feeding was started. Two months later she presented with severe weight loss, vomiting and dehydration, requiring a naso-jejunal tube. Subsequently, she presented daily episodes of vomiting, tachycardia, prostration, sweating, and anxiety after meal alleviated by vomiting. Abnormally high serum glucose level after meals (310 mg/dl) and late hypoglycaemia (48 mg/dl 120 minutes after meal) precipitated a diagnosis of DS. She was treated with dietary measures including: fractionating meals, delaying liquid intake, increasing the thickness of foods, avoiding simple sugars and adding uncooked starch. At 14 month follow-up she was exclusively orally fed and showed a significant clinical improvement with complete resolution of "dumping episodes".
Discussion: In children surgically treated for oesophageal atresia, even without gastroesofageal reflux surgery, DS should be considered and ruled out in the presence of suggestive gastrointestinal symptoms. The management of DS can be challenging and requires careful nutritional intervention.

####

Use of camel milk in children with cow milk allergy

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Objective: Cow's milk (CwM) is an alternative to human milk when breastfeeding is not possible, but some children develop an allergy to CwM proteins (CMA). In these cases, formula milks (*hydrolysed protein* or *amino acid*) or milks from other mammals such as goat, are necessary. Arab and Asian populations use camel milk (CM) which is gaining interest among researchers for its unique composition, which is without beta-lattoglobulin and has a different percentage of casein subclasses. We decided to investigate CM reactivity in CMA children.
Methods: We tested six CMA patients aged from 14 months to 13 years using the Prick Test with food antigens and Prick by Prick test with powder CM and full cream CwM. Neither antihistamine nor cortisone had been administered in the previous week.
Results: Children were positive at the Prick Test for CwM. The size of the wheal was between 6-12 mm. At Skin Prick Test the size of the wheal was 3 to from 6 mm for casein, from 5 to 11 mm for α-lactalbumin, and from 3 to 10 mm for β-lattoglobulin. All children were positive for full cream CwM and negative for CM at Prick by Prick test.
Conclusions: Our results are consistent with the study of Ehlayel *et al*. CM could represent a possible alternative to infant formulas. Larger studies are needed to confirm our findings.

####

Recurrent Rhino-ocular-cerebral Mucormycosis in a Leukemic child: Case Report and review of pediatric literature

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We describe a case of a 12 year old girl affected by T-cell ALL who experienced aggressive rhino-oculo-cerebral mucormycosis during the first "reinduction" schedule. L-AMB intravenous therapy was administered and a double rhino-endoscopic debridement of maxillary sinus and ethmoidal cells and dacryocystorhinostomy were performed. One week later, an angio-MRI showed a pseudo-aneurysm of the left internal maxillary artery that resolved during the angiographic procedure. Gradual symptom and imaging improvement followed. However, six months later, during posaconazole prophylaxis, the girl exhibited a rapid reactivation of the palatal infection, necrosis of upper left maxillary bone and spontaneous avulsion of 3 teeth. High-dose L-AMB was restarted and left hemimaxillectomy was essential, together with palatal and orbital base prosthesis implant. Chemotherapy was stopped. At present, 6 months from the surgical procedure, the girl shows no functional deficits, mild cosmetic impairment and complete hematological remission. Mucormycosis is a rare life-threatening fungal infection that is becoming increasingly common in immunocompromised patients. Therefore, it should be included in the differential diagnosis for facial pain, edema, paralysis and paresthesia. Our case demonstrates that aggressive multisite surgical de-bulking of disseminated fungal foci together with antifungal therapy can result in survival despite the grave prognosis associated with disseminated mucormycosis.

####

Pathologic fracture: a rare situation in children.

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Introduction: Pathological fracture is a rare condition in the paediatric patient. It should be suspected if the fracture involves an unusual district, or if it is associated with minimal trauma.
Case report: An 11 year old girl presented a spiroid fracture of the tibia, after an ankle sprain occurred during low-intensity sport activity. Leg X-ray showed a 5 cm multi-loculated radiolucent area surrounded by a thin corticalization layer associated with the fracture. A conservative treatment was initially chosen with closed reduction of the fracture and immobilization. An MRI showed a lesion with a hypointense signal on T1-weigthed sequences and a dishomogenous hyperintense signal onT2-weigthed images, with typical marrow oedema surrounded by haemorrage secondary to the fracture. The imaging was consistent with non-ossifying fibroma. As the child suffered from a consolidation delay, after 3 months, she was treated with curettage and bone grafting.
Discussion: Non-ossifying fibroma (NOF) is the most common benign bone lesion in children. It is estimated to occur in between 30% and 40% of people <20 years of age. Usually, it is asymptomatic and needs only to be monitored without intervention. NOF should be treated only if it interferes with the bone healing process.

####

Anderson-Fabry disease in children: clinical features

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We present a case of Anderson-Fabry disease (AFD), an X-linked lysosomal storage disorder of glycosphingolipid catabolism, due to deficiency or absence of a galactosidase A (a-gal A) enzyme, with multiorgan involvement and reduced life expectancy. A.T., a 8-year old boy, presented with throbbing headache (RMN and EEG negative), burning pain located at the lower extremities, triggered by exercise (playing football or climbing the stairs) and tingling and pain in the hands, sometimes so intense as to interfere with every-day life.

Healso experienced abdominal pain, easy fatigue, heat intolerance and intermittent tinnitus. Objectively presented angiokeratoma at the right popliteal fossa
AFD was suspected and biochemical and genetic tests were performed with the subsequent detection of pathological a-gal A activity in plasma, but normal a-gal A activity in leukocytes. Direct sequencing analysis of the GLA gene coding regions identified nucleotide substitution c.352>T (p.Arg118Cys) in exon 2 of the GLA gene; coding for a protein with reduced enzymatic activity, 29% of residual activity compared to the native protein.
Since December 2009 A.T. has been in enzyme replacement therapy (ERT) with agalsidase-alfa (Replagal®) at a dose of 0,2 mg/kg every 14 days, with improvement of symptoms, and reduction in the use of FANS.

####

Esophageal atresia… with surprise

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G.B.newborn girl, cesarean delivery at 40+2 weeks gestation; birth weight 2630g; length 47cm, head circumference 33,5 cm; Apgar 1'=8, 5'=10. Maternal hypertension during pregnancy; prenatal ultrasound without detection of any fetal malformation; no family history for other pathologies.
Examination at birth was normal, without evidence of associated malformations; the baby showed profuse sialorrhea; nasogastric tube (NG-tube) stopped during positioning, and Chest-Abdomen X-Ray showed tube tip at D1 and gastric air (Fig 1).
Esophageal atresia with tracheaesophageal fistula was suspected. At 1 day of life the baby underwent general anesthesia; tracheoscopy confirmed the presence of a large tracheal fistula; at surgery the fistula was sutured and esophagus-esophageal anastomosis was wrapped.
Postoperative care was uneventful. Eight days after surgery, esophageal X-ray with Gastrografin® was performed before starting oral feeding. The exam showed repeated opacification of the distal third of the trachea and of the bronchial tree on the right side (fig.2). A NG-tube was repositioned and oral suction was combined to avoid inhalation. A new esophageal X-ray with Gastrografin® through a tube was performed (fig.3 - 4) and a thin fistula, approximately 2 cm above the bronchial fork and along the left posterolateral wall of the esophagus, was detected.
The baby underwent tracheoscopy to confirm the suspicion of an upper fistula. The endoscopic evaluation showed the main trachea-esophageal fistula closed. Slightly above, on the left posterolateral wall, a proximal native fistula of 3mm diameter was found.
An attempt at endoscopic closure with direct injection of cyanoacrylate was made. Three weeks after, a new X-Ray showed the persistence of the fistula (fig.5). At the age of 2 months, the baby underwent surgery and the proximal fistula was sutured and divided.
Postoperative course was uneventful and 10 days after surgery oral feeding started. The baby was discharged 4230g weight, with a normal X-ray (fig.6). Follow up at 3-months was normal.

####

Not all nodes come to comb

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A 4 year-old child presented with fever twice without other symptoms, which resolved spontaneously. After two months she had fever again with erythematous like nodular lesions of about 2cm, that appeared bilaterally at the level of the lower limb extensor .
The overlying skin appeared hot, red and characterized by pain in acupressure. After about a week the lesions disappeared but reappeared after a few days so she was admitted to hospital. We performed: complete blood count, erythrocyte sedimentation rate, C-reactive protein, tuberculin skin test, throat culture, antistreptolysin O

titer, chest X-ray, stool examination for parasites and calprotectin, ANA, anti-dsDNA, ASCA and ANCA. Enzyme-linked immunoassay tests for M. and C.pneumoniae, HBV, HCV, EBV, CMV and B. burgdorferi. B_2-microglobulina, ACE and calcemia to exclude sarcoidosis's diagnosis. All these exams came back negative except for respiratory Multiplex, Corona and Rhinovirus, but she showed new nodular lesions. E.N. is an acute inflammatory dermatosis with painful non ulcerative nodules, mainly located to the extensor surface of the lower legs. The nodules are often accompanied by fever and resolve without sequelae. EN has been associated with infectious and non infectious diseases. Age and sex distributions vary according to etiology; women are affected more than men. In more than half of cases a specific etiology can not be traced despite correct diagnostic procedure, so we can considerate EN idiopathic.

####

Intrathoracic Mass in Girl with Neurofibromatosis type 1

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An 8 year old girl with neurofibromatosis type 1 (NF1) was put on treatment with amoxicillin with clinical suspicion of right lung pneumonia. Within 2 days fever disappeared but a worsening cough appeared over the following fortnight. Consequently, she visited our hospital: BP was 99/70 mmHg, PR 140/min, SpO2 100% and RR 26/min. The physical examination revealed a pale girl with poor respiratory sounds on the right lung and a systolic heart murmur.
The lab evaluation showed mild anemia with normal white cell blood count and mild increase of inflammatory markers.
A chest ultrasound scan revealed a pulmonary consolidation involving most of the right lung, confirmed by chest X-ray. The CT better defined a large mass occupying the right thoracic cavity (16x12x10 cm) causing tracheal deviation, atria and vena cava compression.
Due to her quickly worsening condition a trucut biopsy was performed showing an undifferentiated high-grade sarcoma. Pleuropulmonary blastoma and malignant peripheral nerve sheath tumor in NF1 were included in the differential diagnosis. She received cytoreductive chemotherapy and radiotherapy in order to reduce the mass and attempt the surgical excision.
Patients with NF1 have an increased incidence of tumors. In addition to neuroblastoma and leukaemia, sarcomas are more common in children with NF1 than in the general population. The most common are malignant peripheral nerve sheath tumor and rhabdomyiosarcoma.

####

A "complicated" case of juvenile idiopathic arthritis

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A 13 year old girl was admitted to hospital with persistent fever, rash (as in Figure) and generalized myalgia and arthralgia. During hospitalization she presented intermittent fever, evanescent urticarial rash (associated with fever spikes), cervical lymphoadenopathy, hepatosplenomegaly (confirmed to ultrasound), and arthritis especially bilaterally in the upper limbs. Initial laboratory findings showed neutrophil leukocytosis, elevated markers of inflammation and negative results for investigations for the most common causes of arthritis. A diagnosis of systemic juvenile idiopathic arthritis (sJIA) was made on the basis of the ILAR classification criteria for the disease. During hospitalization, she presented a progressive deterioration in her general condition with continued fever and severe abdominal pain. Laboratory evaluation showed anemia, thrombocytopenia, hypertriglyceridemia, marked in-

crease of liver enzymes, ferritin and D-dimers. CT thorax and abdomen showed minimal pericardial and pleural effusion, hepatosplenomegaly and thin fluid collections at peri-hepatosplenic and pelvic levels. Macrophage activation syndrome (MAS) was suspected and a bone marrow aspiration was performed. It revealed signs of emophagocytosis, so corticosteroid therapy was begun.
MAS is a clinical entity characterized by serious liver diseases, hematologic anormalities, coagulopathy resembling disseminated intravascular coagulation and neurologic involvement. Its association with sJIA is rare.

####

Severe malnutrition in celiac disease: the essential role of gluten free diet compliance.

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Celiac disease is an immune-mediated gluten-dependent enteropathy with a wide range of clinical manifestations in genetically susceptible individuals. Infection and poor adherence to diet can cause celiac crises characterized by diarrhea, vomiting, dehydration, acidosis, hypoproteinemia and shock. Here ee describe a case report. R. is a 2 yr old male admitted to our Paediatric Unit for vomiting, diarrhea and weight loss (2Kg) over 2 months. The investigation performed at home showed high titer of Ab anti-tTg. On admission R. presented poor general condition and severe malnutrition (Weight <3°) with "tobacco-bag-like" buttocks, muscle wasting and abdominal distension (1-2). R. showed indifference, irritability, and game refusal. Blood tests showed metabolic acidosis, anaemia and confirmed celiac disease. Hand X-ray showed osteopenia. I.v. rehydration and steroid treatment was started. The child was gradually fed a gluten free diet. Parents were also provided with adequate dietary instructions. Following nutritional rehabilitation, R's clinical condition gradually improved with significant weight gain and he recovered interest in food, play and his environment.
Celiac crisis in children is a rare complication and few cases have been reported in the literature; its major cause is transgression of diet in patients with poor nutritional status often after intestinal infection. Nutritional education and regular follow up are essential for good diet compliance and to prevent complications.

####

Common variable immunodeficiency and autoimmune haemolytic anemia in a patient with CHARGE syndrome

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The following case is about a 9 year old patient with CHARGE syndrome who developed autoimmune haemolytic anemia and common variable immunodeficiency.
At the age of 7 months, leucopenia and thrombocytopenia occurred after a febrile episode and the patient was administered folic acid supplementation, IVIG and steroids until normalization of CDC and healing of bleeding manifestations.
At the age of 7, the child underwent surgical exeresis of two subcutaneous nodules. The hystological diagnosis was cutaneous T-cells lymphoma-like and inflammatory granulomatous dermatitis. With the suspicion of lymphoproliferative disease, a total body TC-scan was performed which showed enlarged retroclavear, supraclavicular, axillary, mediastinal and retroperitoneal lymphnodes and small lung nodules. The bone and bone marrow biopsies as well as culture tests were negative. Blood tests showed monoclonal hypergammaglobulinemia, serum positivity of EBV and HHV-6.
After an episode of dark urines associated to weakness, pallor and subicterum, blood tests showed severe anemia with elevation of haemolysis indices which did not respond to steroid and IVIG and required blood transfusion.
Last month, since the child had frequent infections of the airways and candidiasis of the mouth, the immunologic profile was analyzed showing slight deficit of humoral bilinear immunity and lack of vaccinal immunity compatible with a condition of

common variable immunodeficiency and treatement with IVIG has been already started.

####

A rare condition of crying

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We studied a 2 year old Caucasian girl with psychiatric symptoms. The patient had an uncomplicated perinatal history. Her medical past and family history were unremarkable.
The child was admitted to our Emergency Department suffering from persistent, inconsolable crying lasting > 3 hours, psychomotor agitation and aggressive behavior. This condition occurred within 5 days of gastroenteritis. On admission her neurological examination was normal and her blood chemistry results were normal. There was no history of fever. The patient was therefore referred to the pediatric clinic for a neurological workup. In the first 48 hours a functional cause was hypothesized after negative results for brain magnetic resonance imaging (MRI) and electroencephalographic exam (EEG) were obtained. Due to the persistence of symptoms, a lumbar puncture was performed which gave normal biochemical results. Polymerase chain reaction (PCR) performed on cerebrospinal fluid (CSF) showed positivity for DNA of Herpes-virus type 7 (HHV-7), and the same test applied on peripheral blood mononuclear cells gave a positive result.
These results are highly supportive of an infection of the central nervous systems, caused by HHV7. We emphasize the difficulties of early diagnosis at the first episode of acute psychiatric disorder associated with acute gastroenteritis. Therefore, it is necessary to consider viral encephalitis, and perform neuroradiological exams, cerebrospinal fluid analysis and blood test.

####

Unusual paroxysmal autonomic manifestations in a 22 month old girl

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G, 22 months old, was referred to the Pediatric Department because of cyclic vomiting and masticatory automatisms reported for approximately 30 days. She was born at term after an uneventful pregnancy, no history of hereditary disease or neurological disorders were reported in either parental nuclei. Her clinical history was characterized by the appearance of “gastroesophageal reflux” at the age of 21 days of life which was subsequently investigated for hypertrophic pyloric stenosis, cow's milk protein allergy and fructose intolerance. All examinations performed were within the normal range and the pattern of symptoms gradually settled to about 3 episodes per month. During hospitalization, we decided to perform a video-EEG that revealed paroxysmal activity in the right temporal-parietal derivations. A brain MRI was subsequently carried out showing a mild asymmetry of the hippocampal right regions. These elements taken together allowed us to make the diagnosis of Temporal Lobe Epilepsy (TLE). In addition, the patient was placed on therapy with cambamazepine without recurrence of symptoms. Semiology of TLE in children older than six years resembles adult semiology, but younger children exhibit features reminiscent of frontal lobe seizures. The main distinctive features of TLE seizures in infants are a predominance of behavioral arrest with possible impairment of consciousness, no identifiable aura, automatisms that are discrete and mostly orofacial, more prominent convulsive activity and a longer duration (more than one minute).

####

Acute pain management in a child with sickle cell disease: a case report

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An 11 year old African girl affected by sickle cell disease (SCD) presented at the Emergency Department with a history of back pain of 5 days duration and fever. The pain was reported as severe in nature, persistent, restricting her physical activity and with no-response to anti-inflammatory drugs (acetaminophen, codeine). She rated her pain as 7 on numerical scale (1-10). The patient was in therapy with hydroxyurea, amoxicillin and folic acid. Laboratory results showed: Hgb 9.4 g/dL (HbA2 3.2%, HbF 10%, HbS 81.6%); WBC 9080/uL; CRP 17.99 mg/dl; LDH 706 mU/ml. Antero-posterior spine and chest X-ray were normal.
The girl was febrile and hemodinamically stable with normal vital signs. Pheripheral blood culture was negative. Abdominal US showed normal results. Ketoprofen, idratation and Ceftriaxone were promptly started. We decided on NSAIDs because of their effectiveness in relieving the inflammatory component of infarctive bone pain. The patient's pain was adequately controlled, as verified in following clinical evaluations. When the acute pain began to resolve, the ketoprofen dose was tailed off gradually.
The patient's painful vaso-occlusive episode (PVOE) was appropriately treated and the girl was discharged on oral drugs. The most common SCD manifestation is painful PVOE and inappropriate treatment leads to unnecessary suffering and potentially fatal complications. Physicians must evaluate, treat and follow the pain evolution to improve the patients' quality of life.

####

Our experience in Palliative cure and pain management in a terminal stage galglioneuroblastoma patient

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A 6yr old girl affected by final stage abdominal ganglioneuroblastoma presented at the Emergency Department with severe neuropathic back and abdominal pain and left leg paresis caused by the primary tumor. Pain was incompletely controlled by acetaminophen, tramadol and gabapentin. Metabolic radiotherapy with 177gLu was performed 4 months before to delay cancer growth and to treat the pain, with temporary gain. The patient was admitted to the palliative care department. The pain was rated as 9 on numerical scale (1-10), associated with agitation and anxiety. Morphine iv 2.5 mg/h was started with incomplete pain management, so was increased to 3.5 mg/h, with oral etoricoxib 120 mg/day and gabapentin. Thanks to a temporary pain relief, the therapy was gradually switched to fentanyl transdermal (100 mcg/h), then sc morphine was started (15 mg/dose) and gabapentin increased. Clinicians continued to assess pain whenever possible. The elevated pain scores reflected the need for sedation. Fentanyl was reduced and the iv morphine was properly escalated till 140 mg/day with rescue doses, associated with midazolam (for effective palliation of agitation and anxiety) and ketoprofen. The girl was comfortable, feeding and sleeping well until death. There is a lack of epidemiological data on pediatric pain oncology. Management of children's pain is somewhat different from that in adults. Physicians need to achieve good pain control to improve quality of life in children with advanced malignancies.

####

Two unusual presentations of mononucleosis.

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INTRODUCTION: acute Epstein-Barr-Virus (EBV) infection can result in a broad spectrum of clinical manifestations. We report 2 unusual presentations of infectious mononucleosis (IM) in children: one with ocular involvement, and another with neurological features.

CASES REPORT: **Case 1:** a 3 year old girl was admitted to our hospital for swelling and purulent discharge from the left eye for 5 days - treated with antibiotic eye drops with no effect - and fever for 1 day. On admission, she presented with hyperemia and swelling in the left medial canthal region, with purulent discharge. Tonsils were hypertrophic with exudate. Blood sample showed lympho-monocytosis. EBV serology was compatible with IM. The dacryocystitis was treated with systemic and topical antibiotics and oral steroid with consequent resolution. **Case 2:** a 7 year old boy was admitted to our hospital for difficulty during the masticatory phase and dysarthria. The previous week he had presented fever and clinical signs of IM. Physical examination showed signs of hypoglossal nerve palsy (HNP). Neuroimaging excluded intracranial lesions. IgM EBV-VCA were positive. Oral steroid was initiated with complete recovery.
CONCLUSIONS: acute dacryocystitis and HNP are uncommon in children. Regarding dacryocystitis, IM should be considered in children without previous similar episodes. Concerning the isolated cranial nerve palsy - once organic lesions have been excluded - it is important to consider IM.

####

The use of ultrasonography in the diagnostic-therapeutic approach in a case of pleural empyema

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Case: Pleural effusion and empyema are the most frequent complications of pneumonia in children. A 4 year old child was hospitalized for fever, abdominal pain and diarrhea. He was in pain, chest auscultation revealed rhonchi and rales, but oxygen saturation was good. His chest x-ray showed parenchymal consolidation on the left pulmonary base. He started empiric therapy with Ceftriaxone, without clinical improvement. The thoracic ultrasound showed a pleural effusion. We performed a diagnostic and therapeutic thoracentesis. He started a new therapy with Ampicillin sulbactam and Amikacin i.v. A further thoracic ultrasound was performed which showed an increase in pleural effusion with fibrin septa. The pleural fluid was positive forStaphylococcus H. and the IgM for Mycoplasma P., so clarithromycin and vancomycin were administered with rapid clinical improvement. Fibrin persisted in the pleural effusion, despite pleural washings with urokinase, so he was treated with a thoracotomy with pleural decortication.
Conclusion: Staphylococcus Hominis, has long been considered a contaminant of culture. In this case, we sawa Mycoplasma pneumonia with Staphylococcus superinfection, which was responsible for pleural effusion and antibiotic resistance. Thoracic ultrasound played a key role in monitoring. Early pleural decortication, when pleural washes with urokinase are ineffective, is often the only treatment option toprevent a lobectomy.

####

Tibial Brodie's abscess in a child

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A 4 year old child presented with a 10 day history of fever, intermittent pain in his left knee and refusal to walk. 2 weeks before she had had an accidental trivial trauma of the left knee, without any visible skin lesion. Magnetic resonance imaging (MRI), performed because of a slight increase of blood inflammatory markers, revealed a hyperintensity signal area in T2-W sequences of the left knee, surrounded by intense signal alteration of the bone tissue. This finding was interpreted as a post-traumatic oedema. The child was completely asymptomatic after a few days, and the blood inflammatory markers were normalized. An MRI and radiograph control performed 20 days later revealed a well-delineated pseudo-oval lesion in the proximal metaphysis of the left tibia, with intense enhancement and a thin layer of hypointense signal around the central part

of it. The likely explanation of the MRI findings was that of a thin layer of subacute abscess of an osteomyelitic process. The child underwent surgery: cultures were found to be negative and no bacteria were seen microscopically. Histopathology confirmed the diagnosis of subacute osteomyelitis (Brodie's abscess). The patient was administered intravenous ceftriaxone and oxacillin for 8 weeks with complete recovery. Brodie's abscess is a type of subacute osteomyelitis, which may present without any symptoms and with normal laboratory parameters. Cases involving the proximal tibia are diagnosed incidentally, as in our patient.

####

Protein-losing enteropathy in a child: Langerhans cell histiocytosis involving gastrointestinal tract

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Background: Gastrointestinal tract (GIT) involvement in Langerhans cell histiocytosis (LCH) is very rare and associated with a poor outcome. We report a 20-month-old female presenting with protein-losing enteropathy (PLE) caused by LCH involving GIT.
Case report: A 20 month old girl was admitted to our unit because of diarrhea, vomiting, oedema and dystrophy. Laboratory evaluation revealed a PLE (hypoprotinemia, hypoalbuminemia and elevated fecal alpha-1-antitrypsin concentration). Abdominal MRI showed hepatosplenomegaly, ascites and mesenteric lymphadenopathy (Fig. 1-3). GI endoscopy showed abnormal duodenal mucosa with edema, erythema, friability and an overlying exudates and lymphoid hyperplasia of the colon (Fig. 4-5). Duodenal and ileum biopsies showed a mononuclear cell infiltrate and medium-sized cells with folded nuclei in the lamina propria. These cells, accompanied by abundant eosinophils, were positive for CD1a by immunostaining, supporting a diagnosis of LCH (Fig. 6). A bone marrow aspirate confirmed LCH. Chemotherapy was initiated using the first-line treatment of LCH-IV protocol for multisystem LCH. The patient showed clinical improvement with improved weight gain and resolution of GI symptoms and edema. However chemotherapy is still ongoing.
Conclusions: LCH involving GIT, a rare but highly fatal disease, should be considered in infants with refractory and severe gastrointestinal symptoms; endoscopic biopsy is strongly recommended for immediate diagnosis.

####

Blackwater fever: our experience in an hospital in Cameroon

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Malaria is a major public health problem in Africa. P. falciparum (PF) is the main species associated with complicated malaria cases such as blackwater fever (BWF), a clinical syndrome characterized by an acute intravascular haemolysis presenting with dark urine, jaundice, anemia and fever. Abdominal pain, vomiting, hepatosplenomegaly, bone pain, dyspnea, tachycardia, generalized malaise and dizziness can also be found. Bryan, a 10 year old boy was admitted to Mary Health of Africa (Fontem, Cameroon) as a result of a 6 day history of fever, drowsiness, dark urine and jaundice. He had previously been treated with quinine for suspected malaria. On admission, GCS was 10/15, heart rate was 150/bpm and temperature was 38.5 ° C. He was jaundiced and dehydrated, with mild hepatomegaly and diffuse abdominal tenderness. Bloods showed haemolysis and renal failure (Hb:6,5 g/dl, bilirubin:3 mg/dl, GOT 65 mg/dl, creatinine 2,7 mg/dl), thick film tested positive for malaria. He was transfused, IV fluids and treatment

with artemisinin were started. Urine output was initially reduced. He quickly improved with resolution of fever and neurological status returned to normal on the 4th day of stay. BWF is a severe complication of malaria with a high mortality rate. Susceptible individuals are patients previously treated with quinine and G6PD deficiency. Such a condition requires prompt treatment with an alternative anti-malarial drug, and can be particularly challenging in a low income country.

####

Lung ultrasound: a useful tool in diagnosis and management of bronchiolitis

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Objective: To evaluate the accuracy of lung ultrasonography in the diagnosis and management of bronchiolitis in infants.
Study design: This was an observational cohort study of infants admitted to our Pediatric Unit with suspected bronchiolitis. A physical examination and lung ultrasound assessment were performed on each patient. An exploratory analysis was used to assess correspondence between the lung ultrasound findings and the clinical evaluation and to assess the inter-observer concordance between the two different sonographs.
Results: 106 infants were studied (average age 71 days). According to our clinical score, 74 infants had mild bronchiolitis, 30 had moderate bronchiolitis and 2 had severe bronchiolitis. The control group was composed of 25 infants. Agreement between the clinical and sonographic diagnosis was good (90.6%), with a high inter-observer ultrasound diagnosis concordance (89.6%).
Lung ultrasound allows for the identification of infants who are in need of supplementary oxygen with a specificity of 98.7 %, a sensitivity of 96.6 %, a positive predictive value of 96.6% and a negative predictive value of 98.7%. An aberrant ultrasound lung pattern in posterior chest area was collected in 86% of infants with bronchiolitis. In all patients, clinical improvement at discharge was associated with disappearance of the previous LUS findings. Subpleural lung consolidation of 1 cm or more in the posterior area scan and a quantitative classification of interstitial syndrome based on intercostal spaces involved bilaterally, correlate well with bronchiolitis severity and oxygen use.
Conclusion: The lung ultrasound findings strictly correlate with the clinical evaluations in infants with bronchiolitis and permit the identification of infants who are in need of supplementary oxygen with high specificity. Scans of the posterior area are more indicative in ascertaining the severity of bronchiolitis.

####

Uvulitis, what's that?

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CASE REPORT: a 12 year old boy presents with drooling, difficulty with swallowing and inability to speak. Two days before he had undergone surgical extraction of lower molars in deep sedation with a laryngeal mask. The procedure was uneventful. Upon examination he was eupneic, with a swollen, elongated uvula covered by whitish exudate. Submandibular and laterocervical lymphadenomegaly was evident. Further evaluation by an otolaryngologist confirmed the suspecion of uvulitis. He was treated with oral amoxicillin-clavulanate and ibuprofen with dramatic recovery in 4 days.
DISCUSSION: uvulitis is an acute cellulitis with oedema and erythema of the uvula. Causes may be both infectious (mainly bacterial, such as S.pyogenes and H. influenzae) and non-infectious, including trauma during airway invasive management, allergic reactions, vasculitis or irrita-

tion by chemicals including cannabis. Patients may present with fever, sore throat, pain with swallowing, drooling and respiratory distress. Uvula appears red and swollen, often with a whitish discoloration. The diagnosis is clinical, and a bacterial culture is recommended to identify infectious forms. The differential diagnosis includes epiglottitis, pharyngitis, retropharyngeal or peritonsillar abscess and angioedema. Antimicrobial therapy is necessary for infectious uvulitis while traumatic forms can be treated with analgesics or topical anesthetics. Generally, traumatic uvulitis resolves spontaneously without sequelae, as in our patient.

####

Ovarian dysgerminoma: clinical and immunohistochemical features

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We present a case of ovarian dysgerminoma (OD) in a 13 year old girl. She presented with recurrent abdominal pain, weight gain of about 3-4 kg, nausea, asthenia and anorexia. On physical examination an abdominal mass was detectable. Pelvic ultrasound showed the presence of a voluminous mass within the right ovary (RO), with an irregular echostructure and a mainly solid aspect. Within the left ovary (LO) two cystic formations were detectable. Abdominal magnetic resonance imaging confirmed the presence of a capsulated solid mass of 12 cm diameter, with some cystic parts inside (5mm to 5 cm), causing bladder and rectum dislocation. The mass showed a gradual and irregular contrast enhancement. Serum markers demonstrated AFP, beta HCG, CEA, CA 19-9, CA125 within normal range and increased LDH levels. Further investigations included karyotype analysis (46, XX) and SRY-gene expression analysis (negative). Surgical treatment and staging included laparoscopic right ovariectomy, prophylactic salpingectomy and appendectomy. A sample of cystic fluid from LO was taken for cytology. Histology features allowed the diagnosis of dysgerminoma, with immunohistochemical phenotype CD117+, PLAP+, AFP-, beta-HCG-, Calretinin-, Cytokeratines-, alfa-inhibin-.
OD, common during adolescence, can be related to aberrant sexual differentiation, which may be clinically unapparent, thus only the detection of Y-chromosomal DNA expression within tumor cells can confirm the presence of gonadal dysgenesis.

####

Stevens-Johnson Syndrome and Mycoplasma Pneumoniae: history of a case with atypical presentation

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Introduction: The SJS, immune complex-mediated disorder, is caused by drugs (macrolides, penicillins) or infection (M.P., HSV). Diagnosis is based on the involvement of two mucosal surfaces (eyes, mouth, esophagus, anus-genital region) with vesicle necrotizing of the skin.
Case: AD, male 9 years, presented cough associated with thrush two weeks before admission. This was treated with clarithromycin, which was suspended, after 24 hours, because of the appearance of a vesicle in the lower labial mucosa. On admission, the child was feverish andin pain, presented bilateral conjunctivitis, confluent bullous lesions in the oral and labial mucosa andskin vesicles causing itchiness on the palms and soles. Remaining O.E. is negative. In doubt about the possible infectious or pharmacological causes we looked at serology and suspended clarithromycin. The serology was positive for M.P. and the chest X-ray showed pulmonary accentuation of right lung. He started broad-spectrum antibiotic therapy and,because of poor feeding, started parenteral nutrition. His clinical

condition deteriorated and sowe started infusion of IGIV (2g/kg in 12 hours), with progressive improvement.
Conclusions: This case presented difficulty with regard to the treatment of complications. It is classified as atypical SJS because of the lack of periorificial involvement . When pharmacological causes are suspected, it is necessary to suspend any antibiotic treatment; however, often the causes are infections (M.P. in 30% of cases). Treatment with IGIV was efficacious.

####

Which came first, the virus or the newborn?

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We present a case report of a Caucasian term newborn who was admitted to the intensive care unit in the 4th day of life to undergo phototherapy for jaundice. Her clinical conditions began to deteriorate rapidly, with desaturation episodes, irritability and anorexia. Blood exams showed leukocytosis and thrombocytopenia with negative inflammation indices. Sepsis was suspected and broad-spectrum intravenous antibiotic therapy was initiated. As a result of the persistent thrombocytopenia, neonatal alloimmune form was suspected, but it was not possible to screen the neonatal serum for platelet antibodies. From the moment that NEC and bacterial sepsis were ruled out, viral infections were considered. After three platelet transfusions and IVIG we saw a normalization of the platelet count. The molecular viral DNA/RNA blood analysis was positive for Enterovirus (Coxsackie virus B). A careful history of an unknown maternal viral illness in the prepartum period confirmed this etiologic hypothesis. Currently, newborn growth and general conditions are good, but we have been observing an unusual prolonged viremia for a period of 8 weeks. The cerebral ultrasonography was negative. Immunological sistem was evaluated and no defect was found. Enterovirus infections are very common in newborns, with consequences ranging from asymptomatic infection and benign illness to severe disease. In this case it is very difficult to understand the vertical or horizontal origin of the viral transmission.

####

A strange allergy

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L, a 5-month old boy, was admitted for profuse vomiting, floppy, pale skin, and lethargy which occurred 1 hour after the ingestion of a meal consisting of rice, water, olive oil, turkey and apple. Three similar episodes were reported in the previous 2 weeks after ingestion of meals containing rice, barley, lamb, cheese and vegetables.
Laboratory findings showed normal white blood cell count, hemoglobin levels, renal and epatic function. Serum specific IgE against food allergens were negative. Electrocardiogram, heart and abdominal echocardiography were normal. A prick-by-prick against rice, turkey, lamb, chicken and vegetables was negative. Weight, height, and cranial circumference were normal. Based on personal history and clinical features, a diagnosis of food-protein induced enterocolitis syndrome (FPIES) was proposed.
For food avoidance guidance, oral food challenges (OFC) were performed with lamb, turkey, chicken, and vegetables, with no symptoms. Parents were instructed to avoid rice, milk and soy until the 2nd year of age, when a specific OFC will be performed. At a 2-month follow-up the child had not presented any other relevant episodes.
FPIES is a severe non-IgE mediated hypersensitivity, common triggers are cows' milk and soy. However, almost any food can cause an FPIES reaction, including rice, cereals, vegetables and meats.

####

What strange behaviour!

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Abstract: Acute disseminated encephalomyelitis (ADEM) is a rare immune-mediated acute inflammatory demyelinating disorder of the central nervous system commonly affecting children. G.S., 2 years 10 months, was admitted to our Emergency Department due to the presence of fever, apathy, vomiting and refusal to walk. Her recent history was evocative of a banal febrile upper respiratory tract infection treated with a short-course of antibiotic therapy, which had been suspended because of the development of strange behaviour and psychomotor agitation. On admission, she was under-reactive but responsive under painful stimulus, her neurological examination revealed global weakness, right esotropia, higher muscular tone at lower limbs and Babinski sign bilaterally. The diagnosis of ADEM was made by T2-weighted and FLAIR MRI images showing hyperintense areas in nucleo -capsular regions, periacqueductal area and spinal cord. Cerebrospinal fluid was negative except for the presence of pleiocitosis (35 mononuclear cells/uL), in particular its total protein content was normal; polymerase chain reaction (PCR) tests for viruses detected EBV and mild HHV6 positivity on blood and Adenovirus positivity on oropharyngeal swab. She was treated with high dose methylprednisolone (30mg/kg daily) for 5 days followed by gradual steroid tapering with a rapid improvement of neurological signs.

####

Esophageal atresia. Twice

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Newborn, male, only son of healthy non-sanguineous parents. Family history negative for hereditary diseases. Pregnancy complicated by poliidramnios. Born by means of a vaginal delivery, induced at 41 weeks g.e.. One-minute Apgar score = 9. In the first few hours of life, he presented marked sialorrhea. Nasogastric tube could not be introduced to more than 12 cm. Chest X Ray with contrast showed esophageal atresia associated with tracheoesophageal fistula of the distal portion, and atelectasis of the superior pulmonary lobe. Infusion of glucosate solution and antibiotic therapy were administered, and a nasogastric tube was placed in aspiration. At 2 days of life, surgical end-to-end esophageal anastomosis and fistula closure were performed. At 13 days of life, aChest X Ray showed stenosis of the surgical anastomosis. Dilatation by means of EGDS could not be managed because of stenosis of the medial portion of the esophagus. A second surgical correction was then mandatory, with resection of the previous anastomosis and reconstruction of a new end-to-end one. After a week, Chest X Ray showed a plain esophageal tract and stomach in situ. Oral alimentation was then gradually undertaken, with satisfying weight gain.

####

Blood pressure alterations in young people with cardiovascular risk: a comparison study between type 1 diabetic patients and overweight subjects

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Adolescents with type 1 diabetes and obesity present higher cardiovascular risk and ambulatory blood pressure measurements (ABPM) which have been shown to predict vascular events more accurately than office blood pressure or random blood pressure measurements. The aim of our observational cross-sectional case-control study conducted in adolescents with type 1 diabetes, overweight subjects and healthy controls was to assess mean blood pressure parameters to identify subclinical cardiovascular risk.
The study included adolescent patients with type 1 diabetes. For each subject we performed systolic and diastolic ABPM

during wakefulness and sleep in the non dominant arm recording blood pressure every 30 minutes for 24 h. We compared the data of patients with those of overweight subjects and healthy controls. ABPM revealed no significant difference between type 1 diabetic and overweight patients, but we did see significantly different values in night-time diastolic blood pressure values.With regard to type 1 diabetic patients and healthy controls we found significant difference in all 24 h Systolic, 24 h Diastolic ,Day-time Systolic Night-time Systolic, Diastolic Blood Pressure values. We detected blood hypertension in 12/60 type 1 diabetic patients and in 10/60 overweight subjects, whereas not one of the healthy controls presented hypertension.We did not find a correlation between alteration of blood pressure circadian rhythm and HbA1c, duration of diabetes, total cholesterol,BMI or microalbuminuria.