ERCİYES MEDICAL JOURNAL

Erciyes Pediatrics Academy Winter Congress
3-5 March 2016, Kayseri, Turkey

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WISCOTT ALDRICH SYNDROME IN A GIRL! WHAT IS THE MYSTERY?

Aslıhan Akdemir[1, 2], Maximilian Witzel[3], Türkan Patiroglu[4], Mehmet Akif Ozdemir[4], Musa Karakükcü[4], Christoph Klein[5], Ekrem Ünal[4]

[1]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Division of Hematology-Oncology, Dr von Hauner Children's Hospital, Ludwig Maximilians University, Munich, Germany

[3]Department of Pediatrics, Division of Pediatric Hematology Oncology, Dr. von Hauner Children's Hospital, Munich, Germany

[4]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey,

[5]Department of Pediatrics, Division of Pediatric Hematology Oncology, Dr von Hauner Children's Hospital, Munich, Germany

OP01

Background: Wiscott-Aldrich Syndrome (WAS) is an X-linked recessive inherited immunodeficiency; which is presented with micro-trombocytopenia, eczema, recurrent infections, and increased incidence of autoimmune diseases, and malignencies. WAS gene is located on the short arm of X chromosome, and encodes WAS protein (WASP). WASP is expressed in hematopoietic stem cell-lineages and responsible for cytoskeleton reorganization affecting the functions of T, B, NK- cell, granulocytes, dendritic cells and platelets. Mutation in the WAS gene ends up with X-linked thrombocytopenia (XLT) or classical WAS. The prevalence of WAS is, 1 in 100 thousand live births.

Aims: In this presentation we would like to share our experience about a girl diagnosed with WAS.

Methods: A 7 year old female patient was followed with trombocytopenia since she was born. She had been hospitalized for recurrent infections, gastrointestinal bleedings, and CMV pneumonia. Bilateral ventilation tube regarding to persistent otitis media was also performed. Serum immunoglobulin levels were checked, Ig A and Ig E levels were increased, Ig M level was decreased and Ig G level was normal. Western blot studies confirmed the reduced WAS protein expression in peripheral mononuclear blood cells. The complete WAS gene was sequenced, one heterozygous mutation in Exon 7, leading to a premature stop codon p.G219*, c.655G>T was found.

Results: WAS is an X-linked recessive disorder, which is seen in male patients due to the transition. But, in case of X gene inactivation, it can also be presented in female patients.

Conclusion: The clinicians must be vigilant about the possibility of X linked diseases such as WAS in females.

Note: This poster is also submitted to the 21st Congress of European Hematology Association, June 9-12, 2016, Copenhagen, Denmark.

A CASE REPORT: FACIAL AND DENTAL MANIFESTATIONS IN A FEMALE WITH APERT SYNDROME

Ayşe Feyda Nursal[1], Serbülent Yiğit[2], Mehmet Kemal Tümer[3]

[1]Department of Medical Genetics, Giresun University School of Medicine, Giresun, Turkey

[2]Department of Medical Biology, Gaziosmanpasa University School of Medicine, Tokat, Turkey

[3]Department of Oral and Maxillofacial Surgery, Gaziosmanpasa University School of Dentistry, Tokat, Turkey

OP02

Background: Apert syndrome (AS) is a rare congenital disorder with autosomal dominant inheritance and is characterized by irregular craniosynostosis, syndactylia of hands and feet, mid-facial hypoplasia, hypertelorism and anomalies of central nervous system, heart and kidneys. AS has been associated with mutations in Fibroblast growth factor receptor 2 *(FGFR2)*gene located on chromosome 10q (10q26). Dental anomalies are common in AS. We report a 6-year-old AS patient with complex dental anomalies.

Case Report: A 6 year-old female patient with AS presented to the dental clinic with complaints of teeth decay and embedded teeth. She had dysmorphic facial symptoms including mid-facial hypoplasia, low-set ears, hypertelorism, prognathic mandible, steep wide forehead, downslanting lateral canthi and palpebral fissures. She had syndactyly of third and fourth digits of both hands. Arachnoid cyst was diagnosed previously. She had intellectual disability. Upper second incisors absent and canine teeth were displaced. Her maxilla and mandible were narrow. The maxillary dental arch was v-shaped. Radiography showed that there were multiple embedded teeth. Orthodontic treatment was planned for the future because the patient was too young.

Conclusion: The aim of the present report is to show the dental manifestations in a case with AS. The treatment and management of AS require a multidisciplinary approach.

A DRESS SYNDROME CASE RELATED WITH CARBAMAZEPINE

Nazan Ülgen Tekerek, Adem Dursun, Başak Nur Akyıldız

Department of Pediatric Intensive Care, Erciyes University School of Medicine, Kayseri, Turkey

OP03

Introduction: Fever and rash can be seen in many diseases. In this study we are presenting 3 years old girl, who was admitted to our hospital with complaints of fever and rash and diagnosed DRESS Syndrome. In her medical history it was learnt that carbamazepine was added to her treatment to control seizures for one month before of admission. We are presenting this study to increase awareness of DRESS Syndrome among physicians.

Case Report: Three-year-old girl was admitted to our hospital with complaints of fever and generalized skin rash of one week duration. She had a medical history of seizure and she was under valproic acid therapy for six month and carbamazepine for one month. The body temperature was 39°C, respiratory rate being 35 breaths/min, pulse rate 152 beats/min, blood pressure 85/55mm Hg. The patient manifested with maculopapular rashes that were scattered and fused on the face, trunk and extremities. Nikolsky sign was negative. Edema was detected on her face and scalp. Physical examination revealed bilateral submandibular lymphadenopathy and hepatomegaly. Laboratory findings were as follows: hemoglobin, 11.1 gr/dL ; white blood cell count, 7130/mm^3; platelet, 115.000/ mm^3; eosinophils, 3540/mm^3; AST, 278 IU/L; ALT, 148 IU/L. Kidney function tests , bilirubin, alkaline phosphatase, total protein, albumin and gamma glutamyl were normal. Serological tests for hepatitis A, B, C and tests for Epstein-Barr virus, cytomegalovirus, herpes-simplex virus, human immunodeficiency virus and ANA were negative. Urine, blood and throat cultures were sterile. Our patient's score was 7 and diagnosed as definite DRESS Syndrome based on RegiSCAR scoring system.

Carbamazepine and valproic acid treatment were immediately withdrawn. We initiated the treatment with intravenous methyl prednisolone 20 mg/kg followed by oral prednisolone 2mg/kg/day. After three days of the prednisolone treatment plasmapheresis was started due to her clinical deterioration and worsening of skin rash. Despite 2 cycles of plasma exchange the patient did not show any improvement. So we treated the patient with intravenous immunoglobulin 600 mg/kg/day for 5 days. On the 8th day of the PICU admission the patient showed improvement and discharged 20th day.

Discussion: DRESS syndrome is is an uncommon, life-threatening drug reaction in childhood and has a mortality rate of %10-20. Due to fever and rash can be seen in wide range of disease such as infectious, rheumatologic and allergic diseases, thediagnose of DRESS syndrome may delay. Difficulties in diagnosis, delaying in appropriate treatment plan increase the potential morbidity and mortality rates. In the differential diagnosis, DRESS syndrome should be considered particularly in patient using anticonvulsant drugs and presenting with fever and rash.. The awareness of DRESS syndrome should be increased among physicians. Mortality rates should be reduced with early diagnosis and treatment of disease.

ANALYSIS OF UCP2 EXON 8 INS/DEL POLYMORPHISM IN CHILHOOD OBESITY

Ömer Ates[1], Samet Özer[2]

[1]Department of Medical Biology, Gaziosmanpasa University School of Medicine, Tokat, Turkey

[2]Department of Pediatrics, Gaziosmanpasa University School of Medicine, Tokat, Turkey

OP04

Aim: Changes in lifestyles resulting in energy intake and expenditure imbalance have led to an increase in obesity prevalence all over the world. The role of genetic and environmental factors in the emergence of obesity is well known. In parallel to the increasing of childhood obesity, the studies which for elucidating the genetic structure that play a role in the etiology of obesity gained momentum. Human uncoupling protein 2 (UCP2) is widely expressed in many tissues including white adipose tissue and contributes to the regulation of energy metabolism. Also, it is thought to play a role in thermogenesis, obesity and diabetes. Thus, it's hypothesized that change in UCP2 gene involved in the development of obesity. In this study, ıt was aimed to evaluate the association between the exon 8 ins/del gene polymorphism is thought to affect the level of expression of UCP2 and childhood obesity.

Methods: In this study, ıt was investigated the frequency of the UCP2 gene exon 8 ins/del genotypes and allelic variants in the range of 6-17 years of school age that 300 children and adolescents patients with obesity and 200 healthy controls using polymerase chain reaction- restriction fragment length polymorphism (PCR–RFLP) method.

Results: DD, II and ID genotype frequencies of the UCP2 gene exon 8 ins/del polymorphism was detected 56%, 37%, 7% in patient group and 51%, 36%, 13% in control group, respectively. Allelic frequencies was detected 74% for D allele, 26 % for I allele in patients and 69% for D allele, 31% for I allele in controls ($P = 0.051$). There was no significant difference in the genotype frequency of UCP2 gene exon 8 ins/del polymorphism between patients with obesity and controls, if the allele frequency was observed a difference close to the significance limit.

Conclusion: Our findings suggest that the presence of the UCP2 D allele may be one of the many genetic factors to genetic susceptibility in childhood obesity.

CLINICAL MANAGEMENT IN SECONDARY PSEUDOHYPOALDOSTERONISM: A CASE SERIES

Sabriye Korkut[1], Leyla Akın[2], Nihal Hatipoğlu[2], Zübeyde Gündüz[3], İsmail Dursun[3], Ahmet Özdemir[1], Selim Kurtoğlu[1,2]

[1]Department of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatric Endocrinology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Pediatric Nephrology, Erciyes University School of Medicine, Kayseri, Turkey

OP05

Background: Secondary PHA is a transient aldosterone resistance condition mostly occurring in relation with urinary system infection and/or malformations. Secondary PHA cases and very few case series have been reported in the literature. In this article, we reported a case series of 8 patients including different clinic presentations which have not as yet been reported in the literatüre and their long-term follow-ups.

Methods: Patients who have secondary PHA reasons in addition to hyponatremia (<130 mEq/L), hyperpotassemia (>6 mEq/L) and high serum aldosteron levels for the age in Erciyes University Faculty of Medicine Pediatrics Department were included in the study.

Results: All the patients in our case series were younger than 3 months old. Among eight patients in our case series, seven patients were diagnosed with PHA secondary to obstructive uropathy (OUP), one patient was diagnosed with PHA secondary to ileostomy. Six patients were diagnosed with OUP together with urinary tract infection (UTI) and in all except one patient, secondary PHA recovered with only UTI treatment before applying surgical correction. All the patients in our case series were younger than 3 months old. Three patients with PUV diagnosis, salt wasting recurred in an UTI attack recurring under 3 months of age. Although they had an UTI attack in later follow-ups, salt wasting did not develop. Salt supplementation was made with IV/oral NaCl of 3 mEq/kg/day at least and 32 mEq/kg/day at most. The salt supplementation lasted between 3 days and 6 months.

Conclusion: PHA should be considered in the differential diagnosis in patients refered with salt wasting crisis in newborn and early infancy period. In the initial evaluation of patients referring with salt wasting crisis, the aim should be to exclude the reasons which might be responsible for secondary PHA. Therefore, urine analysis and renal/surrenal ultrasonography should be considered first. On the other hand, infants known to have UOP should be closely observed for salt wasting in the presence of urinary tract infection, especially in the early infancy period.

NEONATAL ENDOCRINOLOGICAL PROBLEMS IN COLLODION BABIES

Ahmet Ozdemir[1], Sabriye Korkut[1], Selim Kurtoglu[1,2], Nihal Hatipoglu[2], Tamer Gunes[1], Mehmet Adnan Ozturk[1]

[1]Department of Pediatrics, Division of Neonatalogy, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Division of Pediatric Endocrinology, Erciyes University School of Medicine, Kayseri, Turkey

OP06

Objective: To identify endocrinological problems, and particularly those concerning growth, in 42 collodion babies (CBs).

Method: Clinically identified newborn CBs were included in the study group (Group 1). Since CBs are generally premature and/or born small for gestational age (SGA), a control group matched to the patients in the study group in terms of gestational age (± 7 days) and birth weight (100 gr ±) (Group 2) was established. Blood specimens were collected between the 3rd and 7th days of life from both groups for thyroid function tests [thyroid-stimulatinghormone (TSH), triiodothyronine (T3), thyroxine (T4) andthyroglobulin (TG)] and to measure serum GH, IGF-I and IGFBP-3 levels.

Results: Group 1 consisted of 42 CBs (25 males and 17 females) with gestational ages between 32 and 42 weeks and birth weights between 1,400 and 4,000 gr. Twelve patients were assessed as premature and 17 as SGA. Serum IGF-I and IGFBP-3 levels were lower and serum GH levels higher compared to the controls. Primary hypothyroidism was diagnosed in 10 patients in the study group, subclinical hypothyroidism in two and central hypothyroidism in one. A statistically significant difference was determined between the groups in terms of primary hypothyroidism (p=0.01). A weak positive correlation was determined among birth weight and serum IGF-I and IGFBP-3 levels (r=0.23, p=0.06) (r=0.21, p=0.07). Serum GH levels were weakly negatively correlated with birth weight (r=-0.32, p=0.04) and serum IGF-I (r=-0.38, p=0.001) and IGFBP-3 (r=-0.36, p=0.002) levels.

Conclusion: Premature birth and SGA are more common in CBs. High GH and low IGF-I and IGFBP-3 levels in cases indicate malnutrition-like GH resistance. In addition, the greater prevalence of hypothyroidism in babies is noteworthy.

PERCUTANEOUS PDA CLOSURE IN EXTREMELY LOW BIRTH WEIGHT BABIES

Nazmi Narin[1], Ozge Pamukcu[1], Ali Baykan[1], Suleyman Sunkak[1], Ayse Ulgey[2], Kazim Uzum[1]

[1]Department of Pediatric Cardiology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Anesthesiology, Erciyes University School of Medicine, Kayseri, Turkey

OP07

Aim: Patent Ductus Arteriosus(PDA) is an important cause of morbidity and mortality in preterms. As birthweight decrease, risks increase. Main aim of our study is to emphasize the effectiveness and safety of percutaneous PDA closure even in extremely low birth infants (less than 1000 gr).

Material and Method: In our center between the dates June2014-December2015, PDA of eight patients less than1000gr were closed percutaneously. To our knowledge this study includes the largest cohort of infants less than 1000g in the literature, whose PDA were closed percutaneously.

Results: Symptomatic patients, less than1000gr having PDA were included in the study. All have3times medical therapy for PDA closure but it didnotwork. PDA was decided to be contributor of this medical state of them. The mean patient age 16±5.9days. The mean weight of patients was 923±75.9gr. Mean gestational age was 27.2±1.28weeks. Mean PDA diameter was 2.48±0.5mm. Mean Qp/Qs was 1.7±0.2. Morphology of PDA: 5of them were conical, 3of them were tubular. In all patients ADOII-AS device were used for PDA closure (Table1). Steps of percutaneous PDA closure procedure was shown by Figure1. In all patients, we have done closure by venous route. We did not ever used arterial route in 4 patients. There were no major complications reported. Left pulmonary arterial stenosis was detected in 2 patients which were all resolved in 6 months duration.

Conclusion: Interventional catheterization procedures are more commonly used, in the recent years. The advantages of percutaneous PDA closure include a high success rate, shorter length of hospital stay, reduced blood loss, low morbidity rate, and no traumatic scars. Since the length of hospital stay decreases with catheterization, it is much more cost-effective than surgery. We want to emphasize that in experienced centers percutaneous closure of PDA can be an alternative to surgery even in the extremely low birth weight babies.

RETROSPECTIVE ANALYSIS OF POISONING CASES WHICH FOLLOWED IN PEDIATRIC INTENSIVE CARE UNIT

Nazan Ülgen Tekerek, Adem Dursun, Başak Nur Akyıldız

Department of Pediatric Intensive Care, Erciyes University School of Medicine, Kayseri, Turkey

OP08

Background: Although poisonings represent a significant number of preventable cause of morbidity and mortality admissions to the hospital and pediatric intensive care unit (PICU), however data about poisonings requiring PICU care level are limeted. The aim of this study was to evaulate the posining patients treated in the PICU of Erciyes University Faculty of Medicine.

Methods: The records of 186 patients admitted to the PİCU with acute poisoning between 2010 and 2015 were evaluated retrospectively.

Results: Poisoning cases aged between 14 months- 17 years and the ratio of female/male was 1.6:1. Poisoning mostly occurred in the home (87.6%), via the oral route (91.4%). It was noted that 59,1% of poisoning cases were accidental, whereas 28,5% were suicidal and 12.4% were a result of a therapeutic error. Nearly two-thirds (60.2%) of cases were drug-related, while 39.8% were non-drug-related. Central nervous system drugs (27.6%) were the most comon agent in drug related poisoning however corrosive substances were the morst common in nondrug related poisoning. The overall mortality rate in this study was 5.4%. Mortality from non-drug poisoning (4.3%) was higher than from drug-related causes (1.1%).

Conclusion: The results of this study emphasise the need for regulations in industrial and health policies related to the aim of increasing awareness regarding potential toxins, appropriate storage of potential toxins, and general precautions to promote safety in the home.

SATISFACTION LEVEL OF THE PARENTS OF THE HOSPITALISED PEDIATRIC PATIENTS; KUTF HOSPITAL

Selda Bülbül, Osman Dursun, Esra Dursun

Department of Pediatrics, Kırıkkale University School of Medicine, , Kırıkkale, Turkey

OP09

Objectives: This study was planned to determine the satisfaction level of parents of the patients hospitalized for any reason at the pediatric clinics of a university hospital, factors affecting the satisfaction levels and to make suggestions on things to be done to improve the quality of patient services.

Methadology: 179 parents of the children hospitalized between the dates April-December 2014, established the study group. Self-filled questionnares were filled by whoever (mother/father) was with the child. The attendants were asked to assess their satisfaction level by marking between 0-100 for each question.

Results: Among all 67% were the mothers. Mean age of the mothers, fathers and the inpatiened children were 36.34±7.15 years, 42.64±5.23 years and 91,25±52 months (min 0 – max. 192 months) respectively. Mean hospital stay was 2,77±3,37 days (min 1 – max. 37 days). Reasons for hospitalization were; 15% surgery, 68,4% relatively mild reasons such as; epistaxis, fever, rash and joint pain, 16,5% more serious reasons such as; poisining, burns and seizures. In general, satisfaction levels were high. The mean satisfaction levels from the hospital, doctors, nurses, given information and therapy were also high [respectively; *76.74*, *95.31*, *80.28*, *85.65*, *85.75*]. Satisfaction levels of the parents of the patients with serious reasons were significantly higher (p<0,05). Among all, 74.3% mentioned that they feel comfortable by the doctor, and 86,4% reported the clinics as clean. Reasons of dissatisfaction were mainly the general functioning of the hospital (cleaning toilets, lack of hot water, the water flow, safety concerns etc).

Conclusion: Self-assessment demonstrated that, satisfaction with the hospital and pediatric clinic were high. Several deficiencies were detected in the pediatrics clinics, and reformative efforts were made to improve the inadequacies reducing the patient satisfaction and to meet patient demands and needs.

THE EFFECT OF OBESITY ON QTC INTERVAL IN CHILDREN

Esra Akyüz Özkan[1], Hashem E. Khosroshahi[2], Halil İbrahim Serin[3], Zeynep Tuğba Özdemir[4], Mahmut Kılıç[5], U. Aliye Geçit[1], Meral Ekim[6]

[1]Department of Pediatrics, Bozok University School of Medicine, Yozgat, Turkey

[2]Department of Pediatric Cardiology, Bozok University School of Medicine, Yozgat, Turkey

[3]Department of Radiology, Bozok University School of Medicine, Yozgat, Turkey

[4]Department of Internal Medicine, Bozok University School of Medicine, Yozgat, Turkey

[5]Department of Public Health, Bozok University School of Medicine, Yozgat, Turkey

[6]Department of Biochemistry, Bozok University School of Medicine, Yozgat, Turkey

OP10

Background: Obesity has an increased risk for arrhythmia and they have an increased incidence of sudden death without cardiac dysfunction. Delay in cardiac repolarization reflected as QTc prolongation on electrocardiogram (ECG). We aimed to investigate the effect of obesity on the QTc interval and correlation between the QTc interval and body mass index (BMI).

Methods: A total of 45 obese children (mean age 11.14±2.98 years) and 87 healthy controls (mean age 10.8±3.13 years) were compared regarding ECG. BMI was calculated as weight (kg) / height (m)^2. Obesity was defined as BMI exceeding 95th percentile. Because it varies depending on the heart rate, heart rate corrected QTc values were calculated according to Bazett's Formula. For each patient, three consecutive QTc averages were calculated. QTc > 440 msec was considered as prolonged QTc. The correlation between the QTc intervals and BMI was recorded.

Results: The average BMI in the obese children was 26.64±3.93 kg/m^2 and 18.12±3.47 kg/m^2 for control group (p=0.000). QTc intervals were found to be longer in obese children. Mean OTc interval was 413.89±23.26 msec in obese children and 398.95±24.28 msec for healthy controls (p=0.001). Five children in obese group and two children in control group had QTc value over 440 msec. Prolonged QTc value was longer in obese children than controls. There was no correlation between QTc value and BMI in Pearson correlation analyses.

Conclusion: The prolongation of QTc interval indicates impaired ventricular repolarization. In present study there was no correlation between QTc and BMI in obese children. These results suggest that prolongation in QTc value was independent of the degree of obesity and thought to be; this result was due to the effects of obesity on systemic and cardiovascular system. Some studies demonstrated that the QTc interval back to normal value with the treatment of obesity.

THE INITIALSIGNS OF CYSTIC FIBROSIS: THE EXPERIENCE OF THREE CENTER IN THE MIDDLE ANATOLIA

Tuğba Şişmanlar[1], Ayşe Tana Aslan[1],Mehmet Köse[2], Sevgi Pekcan[3]

[1]Department of Pediatric Pulmonology, Gazi University School of Medicine, Ankara, Turkey

[2]Department of Pediatric Pulmonology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Pediatric Pulmonology, Necmettin Erbakan University Meram School of Medicine, Konya, Turkey

OP11

Background: Cystic fibrosis (CF) is the most prevalent inheritable chronic disease in Caucasian children, resulting in recurrent sinopulmonary infection, chronic airway obstruction, and exocrine pancreatic insufficiency. Additionally, CF can be associated with early childhood mortality. Patients classically present with failure to thrive, steatorrhea, recurrent chest infections, meconium ileus, rectal prolapse edema/hypoproteinemia/'kwashiorkor' skin changes, severe pneumonia, salt depletion syndrome, prolonged neonatal jaundice, and vitamin K deficiency with bleeding diathesis. We aimed to review the initial signs of cystic fibrosis patients in three center in middle Anatolia.

Methods: The initial signs of all CF patients seen at 3 pediatric CF centers between 2006 and 2015 in Ankara (Gazi University, Medicine Faculty Hospital), Kayseri (Erciyes University, Medicine Faculty Hospital), and Konya (Necmettin Erbakan University, Meram Medicine Faculty Hospital) all cities located in Central Anatolia were retrospectively reviewed. Patient age, gender, initial signs, pancreatic sufficiency/insufficiency, CF transmembrane conductance regulator (CFTR) gene mutations at the time of diagnosis were recorded.

Results: The study included 231 CF patients that were followed at 3 CF centers between 2006 and 2015. The mean age was 40.83±40.75 months and there were 103 (45%) female and 128 (55%) male patients. The most common initial signs were respiratory tract infections (24%), pseudobartter syndrome (17.7%), diarrhea (10.8%), vomiting (9.9%), failuretothrive (8.6%). Pancreatic insufficiency was detected in 207 (89%) patients. 107 (46%) patients had severe CFTR mutations (class I, II, III) and 21 (0.9%) patients had mild mutations (class IV and V).

Conclusion: Cystic fibrosis is a common chroniclung disease in our country and allover the World. It is expected to increase in the incidence of the disease with newly used newborn screening. Lower respiratory tract infections and diarrhea are some of the most common leadingcauses of childhood death in Turkey and cystic fibrosis, which presents with these signs, can be considered to be important in childhood mortality in our country.

CEREBRAL STROKE IN A CHILD WITH CONGENITAL DYSERYTHROPOIETIC ANEMIA TYPE II

Alper Özcan[1], Türkan Patıroğlu[1], Hamit Acer[2], Hakan Gümüş[2], Serkan Şenol[3], Musa Karakükcü[1], Mehmet Akif Özdemir[1], Ekrem Ünal[1]

[1]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Division of Pediatric Neurology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Radiology, Divison of Interventional Radiology, Erciyes University School of Medicine, Kayseri, Turkey

P01

Background: Congenital Dyserythropoietic Anemia type II (CDA II) belongs to a subtype of bone marrow failure syndromes characterized by monolineage involvement and typical morphological abnormalities in erythroid precursor cells resulting with different degree of hyporegenerative anemia. Moreover reticulocytosis, which is not corresponding to the degree of anemia (ineffective erythropoiesis) with jaundice and splenomegaly are major diagnostic criteria. Causative gene is located at SEC23B. Although stroke among children is rare, it can cause significant morbidity and mortality.

Aims: Herein we present three years old boy who had diagnosed with CDA II and experienced stoke.

Methods: A newborn male baby referred to us with complaints of icterus and anemia. From his medical history it was learned that his parents were consanguineous. Initial physical examination showed pallor, icterus, hepatosplenomegaly and cryptorchidism. Laboratory finding showed anemia, reticulocytosis, hyperbilurinemia. Bone marrow aspiration showed morphological abnormalities of the erythroblasts. The genetic studies showed double heterozygous mutations in SEC23B.Regular transfusions were started. At age of four he admitted to emergency department with complaints of aphasia and physical examination showed facial paralysis. The MRI revealed acute infarcts at left frontal lobe and digital subtraction angiograph showed occlusion of left internal carotid artery suggestive of fibromusculer dysplasia. Enoxiparine was started. And he is under outpatient control without any neurological sequel.

Results: Pediatric stroke is an important cause of long-term disability. Risk factors for stroke in childhood are different from those traditionally observed in adults. Over 100 risk factors for stroke in children have been reported, but in up to one third of patients, no cause is identified, and these cases are classified as idiopathic. In literature search we did not encounter any individual with CDA II who had stroke.

Conclusion: To best of our knowledge this case presentation reports an interesting combination of CDA II and stroke. This combination can be coincidental but clinicians who manage patients with CDA II must be vigilant about the neurological complications including stroke.

Note: This poster is also submitted to the 21st Congress of European Hematology Association, June 9-12, 2016, Copenhagen, Denmark.

ALLOGENEIC BONE MARROW AND MESENCHYMAL STEM CELLS THERAPY FOR DYSTROPHIC EPIDERMOLYSIS BULLOSA

Musa Karakükcü, Ebru Yılmaz, Gülşah Uçan, Mehmet Akif Özdemir, Türkan Patıroğlu, Ekrem Ünal

Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey

P02

Background: Epidermolysis bullosa (EB) represents a group of inherited blistering skin diseases; some forms of the disease are associated with considerable morbidity and increased mortality. The severe forms of the disease are characterized by mutilating scarring, blisters covering large proportions of the body surface. Later in the disease course, mitten deformities, joint contractures, esophageal strictures, corneal erosions, chronic cutaneous infections, and aggressive squamous cell carcinoma can be seen.

Aims: In this presentation, we presented patient who was treated by allogeneic bone marrow and mesenchymal stem cell transplantation for epidermolysis bullosa.

Methods: A 42 day-old male infant was presented to our newborn clinic with vesiculobullous lesions on more than his lower legs and forearms including the entire body. The patient diagnosed with dystrophic EB and was performed hematopoietic stem cell transplantation (HSCT) from human leukocyte antigen fully matched sibling donor. The bone marrow stem cell graft was infused by the intravenous route after the patient was treated with a non-myeloablative conditioning regimen of cyclophosphamide, rabbit antithymocyte globulin, and fludarabine. Hematopoietic stem cells, mesenchymal stem cells derived from the donor were transfused but no engraftment was achieved. After 29 month from the first HSCT, second myeloablative HSCT with conditioning regimen of busulfan, fludarabine, and rabbit antithymocyte globulin was performed. The infused nucleated cell and CD34 doses were 6.09×108 cells/kg, 9.51x106 cells/kg, respectively. In addition to the hematopoietic stem cell, mesenchymal stem cell derived from the donor (2.8x106 nucleated cells/kg) was also transfused by the intravenous route on the -9, 0, +12, +26+35 days of HSCT. The mesenchymal stem cells were reinfused to the patient on the 13th day of HSCT. Micofenolate mofetil and cyclosporine were used for graft versus host disease (GVHD) prophylaxis. The neutrophil, and platelet engraftment was achieved at day +13, +36 respectively. The chimerism was evaluated 99.6%, 100%, and 100% in +33rd, +64th, +77th days of transplantation, respectively. GVHD prophylaxis was discontinued because chimerism was evaluated 1%. We observed markedly healing of the skin lesions after the HSCT.

Results: Concominant use of hematopoietic and mesenchymal stem cells transplantation may be a promising treatment option for patients with epidermolysis bullosa. The non-myeloablative regimen may be a possible reason for graft failure. The myeloablative regimen of busulfan, fludarabine, and rabbit antithymocyte globulin with immunosupression resulted with satisfactory preliminary results in the presented case.

Conclusion: Further studies with long term follow up are necessary to evaluate the optimal HSCT modalities for patients with EB.

MULTIFOCAL OSTEONECROSIS AND SEPTIC ARTHRITIS: AS UNUSUAL MANIFESTATIONS OF A CHILD WITH ACUTE LYMPHOBLASTIC LEUKEMIA

Alper Özcan[1], Deniz Göl Koçak[2], Mehmet Akif Özdemir[1], Selim Doğanay[3], İbrahim Kafadar[4], Türkan Patıroğlu[1], Musa Karakükcü[1], Ekrem Ünal[1]
[1]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Radiology, Division of Pediatric Radiology, Erciyes University School of Medicine, Kayseri, Turkey
[4]Department of Orthopedic, Erciyes University School of Medicine, Kayseri, Turkey

P03

Background: Although, arthritis and joint pain are common symptoms of children with acute leukemia; osteonecrosis and septic arthritis are rarely seem as initial manifestations.
Aims: In this presentation, 9 years old girl with acute lymphoblastic leukemia (ALL) with initial manifestation of septic arthritis and osteonecrosis was presented.
Methods: A 9 year-old female patient referred to our clinic with pain and swelling of left ankle and movement restriction. Physical examination revealed swelling, warmth and redness over the area of the infection. Joint effusion of left ankle, osteonecrosis of left femur, tibia and navicular bone was identified by radiological imaginings. The laboratory results revealed a white blood count of 1540/mm^3, absolute neutrophil count of 270/mm^3, hemoglobin of 6.4 gr/dL, platelet of 10.000/mm^3. Microscopic examination bone marrow and blood smear revealed 90% lymphoblasts (ALL-L1 morphology). Flow cytometry was performed and revealed the presence of CALLA (+) B precursor ALL. Synovial fluid analysis is compatible with septic arthritis and osteonecrosis is radiologically identified. The patient was treated according to ALLIC BFM 2009 protocol. After splinting the joint and antibiotic treatment, patient's condition improved. Our patient is in remission and continuing consolidation treatment.
Results: The initial presentation of ALL may be nonspesific. Fever, cytopenias, organomegaly, pallor and bleeding anomalies often are present. Although, musculoskelatal symptoms of leukemia are common, joint paint in patients with leukemia may be misdiagnosed as juvenile idiopathic arthritis, reactive arthritis and osteomyelitis.
Conclusion: It must be kept in mind that the rheumatologic and orthopedic disorders may mask or co-exist with acute leukemia.

UNUSUAL MANIFESTATION OF A CHILD WITH NEUROFIBROMATOSIS TYPE 1: T-CELL LYMPHOBLASTIC LYMPHOMA

Cem Geyik[1], Salih Akbaş[1], Alper Özcan[2], Mehmet Akif Özdemir[2], Türkan Patıroğlu[2], Musa Karakükcü[2], Ekrem Ünal[2]
[1]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey

P04

Background: Neurofibromatosis (NF) is the most common multisystem neurocutaneous genetic disorder. It has autosomal dominant pattern of inheritance. NF-1 occurs in approximately 1 in 3.000 births. Although NF-1 was associtted with nerve tumor, a few other malignancies such as lymphoma was reported.
Aims: In this presentation, we represent T-cell lymphoblastic lymphoma in a patient of NF-1.
Methods: An 8 year-old male patient was referred to clinic with cough, nodules on his neck and swelling on both eyelids in 2012. Because of venous distention in the neck and distended veins in the upper chest and arms, a thorax CT was performed; and it revealed a mediastinal mass. A needle biopsy performed; and resulted as T cell lymphoblastic lymphoma. Microscopic examination bone marrow was completely normal. MRI scan of brain shows no evidence of infiltration. The patient was treated according to ALLIC BFM 2009 protocol. Maintenance protocol was administered between 2012 and 2014. After the maintenance protocol, thorax CT shows no evidence of mediastinal mass. Remission period of patient lasted only one year. After one year, mediastinal mass identified radiologically. After relaps, the patient's treatment started according to ALL- REZ BFM; and the patient achieved remission.
Results: In neurofibromatosis, norofibromin is responsible for malignencies and it affects Ras–MAPK pathway. The clinic presentations of sporadic cases are more likely to become more aggressive.
Conclusion: Although optic glioma and other nerve sheath tumors are more likely to occur, lymphoma should be also kept in mind. Malignancies are rare but have an important role in mortality and morbidity of the disease and systemic follow-up is important because of this reason.

CUTANEOUS LUPUS IN A CHILD PRESENTED WITH HYPEREOSINOPHILIC SYNDROME

Alper Özcan[1], Yunus Güler[1], Demet Kartal[2], Betül Sözeri[3], Mehmet Akif Özdemir[1], Türkan Patıroğlu[1], Musa Karakükcü[1], Ekrem Ünal[1]
[1]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Dermatology and Venereology, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Pediatrics, Division of Pediatric Rheumatology, Erciyes University School of Medicine, Kayseri, Turkey

P05

Background: Hypereosinophilic syndrome is characterized by increased number of eosinophils in peripheral blood and bone marrow with infiltration of various organs with eosinophils. Cutaneous lupus is uncommon in children; and characterized by papulosquamous discoid lesions, and annular plaques usually around the neck, the back.
Aims: This paper reports a child with cutaneous lupus presented with hypereosinophilic syndrome.
Methods: A 7 years old boy referred to our department with increased eosinophils. His medical history and family background were normal. On physical examination, the rash at face, neck and trunk was detected. No hepatospelenomegaly, nor lymphadenopathy were found. Hemoglobin was 10.1 g/dL, WBC 39.850/mm^3, the absolute number of eosinophils was 17240/mm^3 (43%), platelet count was 537000/mm^3. Stool examination, chest radiography were performed for the etiology of hypereosinophilia. On the other hand serological studies for toxoplasmosis, toxocara, cytomegalovirus, Ebstain Barr Virus were within normal limits. The patient was assessed by a pediatric allergy department but no allergic disease was considered. Abdominal ultrasonography, and echocardiography were normal. Bone marrow aspiration was normal. FIP1L1-PDGFRA mutation was negative. Corticosteroid therapy (1 mg/kg/day dose) decreased the number of eosinophils. The skin biopsy was performed because the rash did not resolve; cutaneous lupus was proven.
Results: The patient is under follow up by the departments of pediatric rheumatology and dermatology without any complication.
Conclusion: Physicians should consider cutaneous lupus as differential diagnosis in children with hypereosinophilia.

EVERY FACIAL PARALYSIS IS NOT BELL'S PALSY!

Özlem Gül Kırkaş, Pelin Kaçar, Cem Geyik, Alper Özcan, Mehmet Akif Özdemir, Türkan Patıroğlu, Musa Karakükcü, Ekrem Ünal

Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey

P06

Background: Peripheral facial paralysis (FP) is seen in childhood with a incidence of approximately 20/100000. The idiopathic facial paralysis (Bell Palsy) is the main cause however infection, trauma, inflammatory and metabolic disorders, vascular malformations, cancer can be underlying disorder of is located. Patints wit acute lymphoblastic leukemia (ALL) can rarely present with FP.

Aims: In this paper, two cases of ALL were reported whom steroid treatment had initiated after a mis-diagnosis of Bell's palsy.

Methods: Case 1: Thirteen-year-old boy were evaluated in ENT clinic with complaints of swelling behind his left ear, facial asymmetry, incomplete closure of left eyelid. It was learned that steroid therapy was started because of suspected Bell's palsy. It was learned that the decline of symptoms were experienced with the treatment. In laboratory studies on the recurrence of the same symptoms WBC was 48,000/mm^3, hemoglobin 12 g/dL, platelet 112000/mm3, BUN 19 mg/mL, creatinine 2.74 mg/mL, uric acid: 10 mg/mL, LDH: 5451 IU/mL. Lymphoblasts were observed in the peripheral blood smear. The cerebrospinal fluid (CSF) revealed leukemia involvement, The ALL IC-BFM ALL 2009 protocol was initiated, and the symptoms of the facial palsy were diminished with the treatment.

Case 2: Five years old girl with symptoms of toothache, and facial asymmetry was started a steroid therapy with a diagnosis of peripheral FP by the child neurology in another hospital. The patient was referred to use because the leukocyte was 145.870/mm^3, hemoglobin 12.9 g/dL, platelets 108,000/mm^3. The peripheral blood smear and flow cytometric analysis showed T-ALL so the 2009 ALL-IC BFM chemotherapy protocol was initiated. CSF cytology showed class V with lymphoblast at CSF. The facial paralysis was improved after the chemotherapy treatment.

Results: Albeit it is rare, facial paralysis can occur as an initial sign of leukemia in children.

Conclusion: The detailed physical examination of the patient with acute facial paralysis, the evaluation of the complete blood count and peripheral blood smear is very important for suspected children. This strategy will prevent the delay of the diagnose underlying diagnosis of the underlying disease will reduce the morbidity and mortality rates.

A CASE REPORT OF FAMILIAL HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS WITH CNS DEMYELINATION COMPLICATED WITH THROMBOSIS

Saliha Çıracı[1], Alper Özcan[2], M Mustafa Özdemir[2], Samuel C. C. Chiang[3], Bianca Tesi[3], Mehmet Akif Özdemir[2], Musa Karakükcü[2], Türkan Patıroğlu[2], Selim Doğanay[1], Hakan Gümüş[4], Yenan T Brysecon[3], Ekrem Ünal[2]

[1]Department of Radiology, Division of Pediatric Radiology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Medicine, Centre for Infectious Medicine, Karolinska Institutet, Karolinska University Hospital, Stockholm, Sweden

[4]Department of Pediatrics, Division of Pediatric Neurology, Erciyes University School of Medicine, Kayseri, Turkey

P07

Background: Hemophagocytic lymphohistiocytosis (HLH) is a fatal disease affecting infants and very young children, including high fever, hepatosplenomegaly and pancytopenia. Hemophagocytic syndrome may be spontaneous or secondary to infection, malignancy or autoimmune disease, and mechanisms involved are poorly understood. The main histopathologic feature is increased proliferation and activation of macrophages with hemophagocytic lymphohistiocytosis throughout the reticuloendothelial system.

Aims: We present a case with cranial involvement of HLH showing diffuse infiltration of white matter complicated with intracranial thrombosis.

Methods: A 5 year-old girl with fever, and pancytopenia was referred to our hematology unit. Also she had a history of recurrent infections. Her parents were consanguine. Lymphadenopathy, and hepatosplenomegaly were detected in physical examination. Ultrasound examination displayed hepatosplenomegaly and intraabdominal free fluid. HLH was revealed on bone marrow aspiration biopsy. Anomaly in NK, and T lymphocyte cytotoxicity and degranulation tests was determined. İn genetic analysis, syntaxin gene mutation was depicted. Immunosuppressive therapy was performed to the patient, diagnosed with familial HLH. Brain MR imaging was performed because of the suspicion of cranial involvement. On MRI diffuse hyperintense signal changes of cerebral white matter on T2-W and T2 FLAIR images, showing demiyelination were detected. There wasn't any mass effect, contrast enhancement and restricted diffusion on MRI. A repeated brain MR performed a month after the first cranial imaging, showed an acute infarct involving left temporooccipital region. Follow up images showed that the infarct was disappeared but white matter lesions was stable on the brain MR imagines. The cerebral white matter lesions were stable but hyperintense signal changes were appeared in cerebellar white matter, accepted as progression. She was died in despite of immunosuppressive therapy.

Results: HLH is a syndrome of pathologic immune activation, in association with a variety of triggers and is prominently associated with cytopenias and combination of clinical signs and symptoms of extreme inflammation.

Conclusion: CNS involvement may occur at the beginning or during the treatment. Patients with CNS involvement should be treated with intrathecal agents. Depiction of the cranial involvement is important for patient's survival and treatment. All patients with HLH should had brain MRI, even if asymptomatic.

Note: This poster is also submitted to the 21st Congress of European Hematology Association, June 9-12, 2016, Copenhagen, Denmark.

FIVE CASES OF A RARE PANCREATIC TUMOR: SOLID PSEUDOPAPILLARY TUMOR

Mehmet Akif Özdemir[1], Selim Doğanay[2], Mustafa Küçükaydın[3], Ceyda Arslanoğlu[4], Alper Özcan[1], Türkan Patıroğlu[1], Musa Karakükcü[1], Kemal Deniz[5], Tahir Patıroğlu[5], Süreyya Burcu Görkem[2], Ekrem Ünal[2]
[1]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Radiology, Division of Pediatric Radiology, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Pediatric Surgery, Erciyes University School of Medicine, Kayseri, Turkey
[4]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[5]Department of Pathology, Erciyes University School of Medicine, Kayseri, Turkey

P08

Background: Solid pseudopapillary tumor (SPT) is one of the rarely seen primary pancreatic tumors and composed 2-3% of primer pancreatic tumors among all age groups. It is mostly observed in young women (91%), especially in adolescent girls. SPT has low malignant potential that mostly emergesas a benign lesion.

Methods: Five patient admitted to our institution and generally complaint us with abdominal pain. The patients age was 13-16 year, median age was; 13 year). Gender was female/male; 4/1. All of patient reveliaeded with ultrasonography (USG) and Magnetic Resonance Imaging (MRI) and detected abdominal mass. All of patient underwent to surgery. One of them underwent a surgery operation including mass resection and splenectomy, one of them underwent a surgery operation including distal pancreatectomy andmass excision. The other patients had a surgery operation just solid mass excised. Histopathological examination was consistent with SPT.

Results: Solid pseudopapillary tumor is a rarely seen pancreatic mass with a low malignancy rates. Its diagnosis may be delayed due to be asymptomatic usually and not to cause any descriptive symptoms. The complete survival rate is extremely high with surgical resection of the tumor.

Note: This poster is also submitted to the XIX. Congress of Turkısh Pediatric Cancer, May 5-8, 2016, Çeşme, İzmir.

OVARIAN SCLEROSING STROMAL TUMOUR

Alper Özcan[1], Emrah Türk[2], Mustafa Küçükaydin[3], Figen Öztürk[4], Mehmet Akif Özdemir[1], Musa Karakükcü[1], Ekrem Ünal[1], Türkan Patıroğlu[1]
[1]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Pediatric Surgery, Erciyes University School of Medicine, Kayseri, Turkey
[4]Department of Pathology, Erciyes University School of Medicine, Kayseri, Turkey

P09

Background: Sclerosing stromal tumours of the ovary (SST) is a rare benign tumors the originate from ovarian stroma. The most common presenting symptoms are menstrual disorders, pelvic or abdominal pain and an adnexial mass. SST treated with excision or salpingo-oophorectomy. Here we present an extremely rare case of a sclerosing stromal ovarian tumor in a child patient.

Methods: A 14-year-old girl patient was presented to the emergency unit with abdominal pain. On ultrasonography examination, right ovarian was 34x18x14 mm, left ovarian 29x18x14 mm measured and millimeter-sized cysts on both ovaries were monitored. Midline in the pelvis containing probably left over origin 88x65 mm size lobuler, well-defined, cystic necrosis and milimeter echogenicity it revealed a solid mass. On doppler examination showed a slight increase in the mass of the peripheral vascularization. There was no ascid in the pelvis. Patient who detected ovarian mass underwent salphingo-oophorectomy by pediatric surgery. Washing cytology came class 2. The final diagnosis was that of sclerosing stromal tumour of the ovary.

Results: Approximately 2% of all cancers of the reproductive system in girls, 60-70% of these lesions are due over. The most common signs and symptoms are a palpable pelvic mass, menstural irregularity and pelvic pain related to the ovarian mass and our patient is complained of pelvic pain. These tumors are usually hormonally inactive. It is expected that long-term survival with complete resection by surgery.

Note: This poster is also submitted to the XIX. Congress of Turkısh Pediatric Cancer, May 5-8, 2016, Çeşme, İzmir.

PORTAL VEIN THROMBOSIS IN A CHILD WITH PROTEUS SYNDROME

Alper Özcan[1], Emir Gökalp[2], Tuğba Yılmaz[2], Neslihan Karacabey[3], Ekrem Ünal[1], Duran Arslan
[1]Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Pediatrics, Divison of Pediatric Gastroenterology, Erciyes University School of Medicine, Kayseri, Turkey

P10

Background: Proteus syndrome (PS) is a rare genetic disorder. Somatic mosaic mutation in AKT1 gene, which has main role in cell signaling is shown in 90% of PS patients and this explains characteristic clinical findings. Major clinical findings are progressive overgrowth, cerebriform connective tissue nevi, linear epidermal nevus, adipose dysregulation, overgrowth of other tissues and tumors.

Methods: A 15 year-old male patient with Proteus Syndrome was referred to clinic with after a week og abdominal pain. In patients history, portal hypertension and esophageal varices noticed. Our patient presented with dysmorphosis of left 3rd and 4th finger, tenderness of abdomen. Portal vein thrombosis, cavernous transformation and thrombophlebitis of peripancreatic and parailiac collateral veins identified radiologically. Due to portal vein thrombosis, 2 mg/kg/day low molecular weight heparin started for treatment. Genetic and biochemical parameters for thrombosis were normal. Fifth day of treatment, patient's symptoms improved. 1 month after treatment, thrombus of dilated veins in left lower quadrant resorbed but thrombus of dilated veins in right lower quadrant is persisting. Thus portal vein thrombosis and cavernous transformation identified. Thrombophlebitis of peripancreatic and parailiac dilated veins resorbed. Low molecular weight heparin treatment is stopped and warfarin treatment started. Our patient is still continuing the treatment.

Results: PS usually has no signs after birth, it develops and progresses rapidly in the toddler period, continues through childhood. Progressive overgrowth, adipose dysregulation, overgrowth of other tissues (commonly spleen, liver) are the major clinical findings for diagnosis of PS. Thrombosis and pulmonary embolism are very rare and life threatening complications of PS.

Note: This poster is also submitted to the XIX. Congress of Turkısh Pediatric Cancer, May 5-8, 2016, Çeşme, İzmir.

A CASE REPORT OF SWALLOWING DYSFUNCTION (NASOPHARYNGEAL PENETRATION), FOLLOWED BY APNEIC CONVULSION MISDIAGNOSIS

Sabriye Korkut[1], Selim Doğanay[2], Mahir Ceylan[1], Ahmet Özdemir[1], Safiye Gözübüyük[3], Pınar Aslan[3], Elif Şüheda Kocaer[3], Selim Kurtoğlu[1]

[1]Department of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatric Radiology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Pediatry, Erciyes University School of Medicine, Kayseri, Turkey

P11

Background: Penetration and aspiration are different degrees of swallowing dysfunction related with the protection of the airway. Penetration is the occurrence of food entering the laryngeal zone yet staying at the same level as the real vocal chords whereas aspiration describes the occurrence of the aspired material in the airway going below the vocal cord level.

Case: In the case of the 50 day old male case with complaints of scattered cyanosis, was born in the 27th week of gestation and was followed up for 45 days in the newborn unit due to prematurity. Under observation, the patient going through episodic desaturation accompanied with apneic episodes has been observed. Patient underwent examination for apnea etiology. Electroencephalography (EEG) determined sporadic sharp wave activity in the left centro-occipital and temporo-occipital regions. Anti-convulsion treatment was started following apneic convulsion diagnosis. During periods where oral feeding were increased patient experienced apnea accompanied by desaturation and bradycardia with each feeding episode and was observed to be in good condition in between feedings. Choanal atresia was eliminated through examinations. Video EEG during feeding was found to be normal. Videofluoroscopic swallowing activity showed nasopharyngeal penetration during swallowing. Neither laryngeal penetration nor gastro-esophageal reflux were found. During feeding of patient with swallowing dysfunction diagnosis suitable positions were applied, the formula's thickness and feeding duration was increased. After these adjustments the patient tolerated oral feeding and no apnea was observed. Anti-convulsion treatment was discontinued.

Conclusion: It is important to know that premature babies are in the risk group for swallowing dysfunction, and the swallowing dysfunction should be considered under apnea etiology. It should be known that with these babies laryngeal or nasopharyngeal penetration alone with no gastro-esophageal reflux or aspiration may be a cause for apnea.

A CASE WITH PRIMARY LYMPHEDEMA

Halil Tangüner, İsmail Dursun, Ayşenur Kısaarslan, Fatmagül Hisarcıklıo, Betül Sözeri, Ruhan Düşünsel, Zübeyde Gündüz, Hakan Poyrazoğlu

Department of Pediatrics, Erciyes University School of Medicine

P12

Lymphedema refers to swelling localized to a bodily region due to accumulation of lymph fluid at subcutaneous tissue as a result of abnormal lymphatic drainage. Although it is most commonly observed at lower extremities, it may also occur at upper extremities, face, neck and external genitalia. The interruption in lymphatic drainage may be either primary or secondary. In primary lymphedema, lymphatic abnormality can be aplastic, hypoplastic or hyperplastic. Secondary lymphedema refers to those caused by decreased lymphatic drainage due to acquired causes. Causes of secondary lymphedema include trauma, recurrent infection (cellulitis, lymphangitis or parasitic diseases), surgical interventions, metastatic malignant disease, several syndromes (e.g. Klippel-Trenanunaysyndrome) and lymphangiosarcoma among others. It is important to discriminate primary from secondary lymphedema by clinical and radiological evaluations. Here, we will discuss a patient presented with swelling at dorsum of left foot and diagnosed as primary lymphedema. A 5-years old boy presented with swelling at dorsum of left foot. The patient had no concurrent symptoms of infection or chronic diarrhea. On scintigraphy, an abnormality involving lymphatic drainage of left lower extremity was detected and no additional pathology regarding lympatic system was detected. The patient was considered as primary lymphedema. A specific therapy involving vascular surgery and physical therapy was recommended to the patient who is still attending to follow-up visits.

A NEWBORN WITH FAMILIAL HEMOPHAGOCYTICLMPHOHISTIOCYTOSISCOMPLICATED WITH TRANSFUSION ASSOCIATED GRAFT VERSUS HOST DISEASE

Ahmet Özdemir[1], Tamer Güneş[1], Samuel C. C. Chiang[2], Yenan T. Bryceson[2], Ekrem Ünal[3]

[1]Department of Pediatrics, Division of Neonatalogy Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Medicine, Centre for Infectious Medicine, Karolinska Institutet, Karolinska University Hospital Huddinge, Stockholm, Sweden

[3]Department of Pediatrics, Division of Pediatric Hematology-Oncology, Erciyes University School of Medicine, Kayseri, Turkey

P13

Introduction: HemophagocyticLmphohistiocytosis (HLH) is a life-threatening hyperinflammatory syndrome. HLH may be primary (familial) or secondary to malignancy, metabolic diseases, collagen vascular diseases, and infections. Transfusison associated graft versus host disease (TA-GVHD) is a rare condition developing after transfusion of blood products containing vividlymphocytes. The disease especially develops among patients with insufficient immune system and has high mortality rate even with prompt medical treatment.

Case Report: A thirteen-day-old male newborn was referred to our neonatal intensive care unit (NICU) with complaints of hepatosplenomegaly, hypoglycemia and bicytopenia. At 41th week of gestation, he was born to a 26-year old gravida 3, para 2 female via normal spontanous vaginal route. Birth weight was 3600 gr; and was under follow-up due to hypoglycemia in another hospital. It was learned that he had received un-irradiated erythrocyte suspension transfusions. In physical examination, the height, weight and head circumference were within normal range His body temperature was 38.5. At the time of admission, his liver and spleen had enlarged 3 cm and 4 cm below his costal margins, respectively. The patient had wide spread macular eruptions extending over the whole body including the head, neck, hands and feet. The following laboratory data were recorded as follows: WBC of 6.23010^9/L; Hb of 4.8g/dL; platelet count of 6×10^9/L; glucose 30 mg/dL; bilirubin total/direct 135 µmol/L/36 µmol/L; AST of 363 IU/L; ALT of 115 IU/L; triglyceride301 mg/dl; ferritin 55357 ng/mL; fibrinogen of 1.24 g/L; PT/APTT of >100/>120 sec; D-dimer of 8960 µg/L; triglyceride of 2.24 mmol/L and LDH of 2369 IU/L. Abdominal B-mode ultrasound examination showed massive hepatosplenomegaly with ascites. A bone marrow aspiration examination revealed numerous hemophagocytichistiocytes consistent with a diagnosis of HLH. Bone marrow aspiration examination revealed histiocytic proliferation and hemophagocytosis. Natural killer (NK) cell killing activity and NK cell degranulation as and determined via coincubation with K562 cells showed reduced but above FHL diagnostic levels (Fig II). Interestingly, antibody dependent cellular cytotoxicity in NK cells as determined by coincubation with P815 cells coated with anti-CD16 antibody and cytotoxic T cell degranulation showed abnormally low degranulation. Skin biopsy of the patient confirmed grade I TA-GVHD. Fresh frozen plasma, erythrocyte suspension, thrombocyte transfusion and intravenous immunoglobuline (IVIG) were applied. In follow-up at NICU, the patient's general status worsened and developed cardiopulmonary arrest. The patient did not respond to ressusitation and died.

Result: HLH is an immune dysregulation disorder involving mainly infants and children. Clinical sign and symptoms include excessive inflammation, cytopenia, hepatitis, life-threatening severe central nervous system dysfunction.HLHshould be considered among patients with persistent fever, hepatosplenomegaly, cytopenia, hyperferritinemia and hypertriglyseridemia. Additionally,clinicians must be vigilant about the importance ofirradiated blood products should be for neonatal transfusion. Additionally, to our best knowledge, this is the first HPS newborn case with GVHDT.

A RARE ENDOCRINE CAUSE OF TACHYCARDIA: REFETOFF SYNDROME

Süleyman Sunkak[1], Özge Pamukçu[1], Selim Kurtoğlu[2], Ali Baykan[1], Kazım Üzüm[1], Nazmi Narin[1]

[1]Department of Pediatric Cardiology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatric Endocrinology, Erciyes University School of Medicine, Kayseri, Turkey

P14

Introduction: Thyroid hormone resistance (Refetoff syndrome) is a syndrome that caused by the decrease in susceptibility to thyroid hormone in end-organs. The incidence was reported as one in 40,000 live births. Mutations in thyroid hormone receptors play role in the patogenesis. Typical laboratory findings are elevated T3, T4 levels and normal or mildly elevated TSH levels. Same mutations may lead to different symptoms in different patients because of different end organ expressions for a mutation.

There are different therapy strategies like thyroid hormone replacement for patients with hypothyroidism or antithyroid treatment for patients with hyperthyroidism. However only propranolol therapy may be sufficient for patients who has only tachycardia compliant. We aimed to report a case with Refetoff Syndrome, tachycardia caused by thyroid hormone resistance ameliorated with propranolol treatment.

Case Report: Nine years old boy admitted to hospital with palpitation. In physical examination, he had tachycardia with 126/minute heart rate. Other physical examination findings were normal. His electrocardiogram revealed sinusal tachycardia. No other rhythm problem was seen in 24 hour electrocardiogram monitorisation. In laboratory examination, free T_4 level was 4,27 ng/dL (0,96-1,77) and TSH was 2,87 µIU/mL (0,7-5,97). Thyroid auto antibodies were negative and thyroid ultrasonography was normal. Laboratory findings indicated thyroid hormone resistance. Genetic analysis was performed. c.926>G mutation which is responsible for Y321C amino acid was detected as heterozygous positive. He was given propranolol therapy: 2 mg/kg/day and after one month, in the second control he didn't have any tachycardia compliant.

Discussion: The sensitivity of peripheral tissues to thyroid hormone is different. While some patients have hyperthyroidism or hypothyroidism signs, some of them are asymptomatic. So that, there is no certain consensus about treatment. Propranolol takes control adrenergic symptoms and decrease T4 to T3 conversion by inhibiting 5-deiyodination path. Propranolol treatment only by itself may be sufficient for the treatment of a this rare disease called Refetoff syndrome.

ACUTE POSTINFECTIOUS GLOMERULONEPHRITIS AND ACUTE PYELONEPHRITIS OCCURED AT THE SAME TIME IN A CHILD

Fatma Hisarcıklı, İbrahim Halil Tangüner, İsmail Dursun, Ayşenur Kısaarslan, Ruhan Düşünsel, Betül Sözeri, Zübeyde Gündüz, Hakan Poyrazoğlu

Department of Pediatric Nephrology, Erciyes University School of Medicine, Kayseri, Turkey

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Acute post streptococcal glomerulonephritis (APSGN) is the most common cause of acute glomerulonephritis in children and characterized by microscopic/gross hematuria, transient hypocomplementemia, hypertension, oliguria and edema. Clinical symptoms of APSGN can mimic acute pyelonephritis (APN) such as abdominal pain, fever and hematuria. We report a child who had both APSGN and APN at the same time.

A nine and half year old girl presented with cola-colored urine, abdominal pain and painful urination. Her history was unremarkable for oliguria, diarrhea, rash and arthritis except upper respiratory tract infection 10-14 days ago. On admission, her physical examination was normal. Laboratory tests revealed pyuria (60/HPF), hematuria (>500/HPF), elevated creatinine (1.1 mg/dL), hypocomplementemia (C3 low, C4 normal) and markedly increased ASO (600 IU/L). Urine culture was positive for ESBL positive K. pneumonia. She was treated with seven days course of ciprofloxacin. Fourth day of admission, her urine color was normal and serum creatinine was 0.6 mg/dl. Complement C3 levels returned to normal within 6-8 weeks.

In conclusion, Both APSGN and APN can be seen in a child at the same time possibly as a coincidentally although there has never been published in English literature. Detailed clinical history and laboratory evaluation based on clinical findings should be done for the early diagnosis of these disorders.

CASE REPORT: PROPOFOL ASSOCIATED ACUTE PANCREATITIS AFTER TRANSESOPHAGEAL ECHOCARDIOGRAPHY

Özge Pamukçu, Süleyman Sunkak, Ali Baykan, Kazım Üzüm, Nazmi Narin

Department of Pediatric Cardiology, Erciyes University School of Medicine, Kayseri, Turkey

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Transesophageal echocardiography is a technique that enables to display the heart through the esophagus with high quality without intervening structures such as lung and bone. Propofol is an anesthetic agent which is used during transesophageal echocardiography procedure often combined with ketamine. Although there are many advantages of using this agent but also it has some disadvantages, such as hypotension, desaturation and hemodynamic instability.

In this case we presented acute pancreatitis as a rare side effect of propofol which is used for transesophageal evaluation.

Case Report: 12-year-old female patient admitted hospital because of heart murmur.

In transthoracic echocardiographic examination we suspected ASD. So that, we performed transesophageal echocardiography under anesthetized with ketamine and propofol.

In the follow up, patient had epigastric pain and vomiting. Erect abdominal X-ray and hemogram were normal. However, amylase and lipase levels were detected higher than normal. Abdominal ultrasonography was performed for acute pancreatitis it revealed that pancreas is edematous. Intravenous fluid replacement and proton pump inhibitors were given as treatment.

Approximately after 24 hours symptoms were regressed and amylase and lipase levels were back to normal. Than patient was discharged.

Discussion: In pediatric cardiology practice, TEE is important diagnostic tool. Because cardiac examinations is assessed clearer by this way. However, the necessity of using anesthesia in pediatric patients is an important limitation.

Propofol is an agent commonly used in anesthesia induction. Pancreatitis is a rare side effect of propofol, although hypotension, desaturation and hemodynamic instability are common.

As well as hypertriglyceridemia may explain the pathophysiology of acute pancreatitis as a side effect of propofol but acute pancreatitis has been reported after single dose.

In this case, we reported acute pancreatitis as a rare side effect of propofol which is used commonly in pediatric cardiology practice.

CASE REPORT: PULMONARY HYPERTENSION AFTER ARTERIAL SWITCH OPERATION

Kazım Üzüm, Süleyman Sunkak, Özge Pamukçu, Ali Baykan, Nazmi Narin
Department of Pediatric Cardiology, Erciyes University School of Medicine, Kayseri, Turkey

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The transposition of the great arteries is a common cyanotic congenital heart disease in neonatal period. The incidence is 5-7% among all congenital heart diseases. Arterial switch (AS) operation is the most popular surgical technique recently. Expected complications following this procedure are coronary artery stenosis, anastomotic supravalvular pulmonary stenosis, neo-aortic valve regurgitation and supravalvular neo-aortic stenosis. But severe pulmonary hypertension in late period after surgery has been reported very rarely in the literature.

In this case, we reported a girl who developed severe PHT 15 months after AS procedure in newborn.

Case Report: A newborn who had cyanosis complaint was admitted to the neonatal service. The echocardiography findings were compatible with transposition of the great arteries. Atrial septostomy was performed in Cath Lab. In the first day of life due to severe cyanosis. She went under AS operation after 7 days.

Except minimal neo-aortic and neo-pulmonic valve regurgitation, postopeTrative echocardiography findings were normal until 12 months.

Patient admitted to our department with fatigue (NYHA, stage 3) and dyspnea in 15 months old. echocardiographic assessment revealed tricuspid valve regurgitation with a high jet velocity (4 m/s, equalization of 74 mmHg RV end-sistolic pressure). Angiographically, pulmonary artery pressure was measured 69 mmHg. Bosentan therapy was started with the diagnosis of PHT. In the second week of therapy, fatigue and dyspnea complaints were decreased partially (NYHA, stage 2) and tricuspid valve regurgitation was regressed in echocardiography (4 m/s jet velocity, equalization of 74 mmHg RV end-sistolic pressure).

Discussion: Common late onset complications after AS procedure are right ventricul outflow tract obstruction and neo-pulmonic valve regurgitation. Late onset PHT after this procedure is a very rare complication.

Pathophysiology of late onset PHT after AS procedure is not clear exactly. A possible etiopathogenesis are altered hemodynamics in the fetal pulmonary vascular bed. The another one is more delicate pulmonary vascular bed during early postoperative period.

The treatment of PHT after AS procedure is controversial. In the cases reported in literature, alone bosentane and bosentan additional sildenafil or prostacyclin combined therapies were used. In our patient we gave only bosentan and planned other combined therapies according to patients clinical status.

PHT which has poor prognosis should be remind after AS procedure, although it is a rare complication.

COLLODION BABY: CASE PRESENTATION

Ahmet Özdemir[1], Cihangir Şahin[2], Müşerref Bağırova[2], Sabriye Korkut[1], Mahir Ceylan[1], Tamer Güneş[1], Selim Kurtoğlu[1], Mehmet Adnan Öztürk[1]
[1]Division of Neonatology, Departmant of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[2]Departmant of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Collodion baby is a rare skin disease characterized with abnormal keratinisation in the skin. Today it is accepted to have a autosomal recession. Eclabium, ear deformities and congenital ectropion secondary to abnormal keratinisation on the skin are typical face appearance findings in these babies.

Case Presentation: Being a male infant born in term, with a birth weight of 3740 gr, with caesarean operation in an external center as the 4th alive child from the 5th pregnancy of a 36 year old healthy mother. It was learned that his parents were first cousins and their first child was born with similar findings and died when 3 months old. In the physical examination, it was observed that the skin covered the whole body as a membrane and it had a hyperemic appearance. Bilateral ectropion and hyperemia in the eyes were striking. Consultation was made with dermatology and eye departments. Vaseline, skin care, synthetic tears and antibiotic containing eye drops were started. The patient whose skin and eye findings had a tendency for recovery was discharged with available care suggestions and inviting for polyclinic controls.

Result: Complications such as dehydration, electrolyte imbalance, sepsis and pneumonia related to squamous cell aspiration are common in collodion babies. Early detection of skin and eye findings and treatment in appropriate conditions are important. Also close observation of metabolic changes and early and wide spectrum antibiotic use in case of sepsis development significantly decreases the death ratio in these infants.

CONGENITAL GINGIVAL GRANULAR CELLED TUMOR IN A NEWBORN

Ahmet Özdemir[1], Emir Gökalp[2], İbrahim Halil Tangüner[2], Füsun Görözen[3], Sabriye Korkut[1], Mahir Ceylan[1], Tamer Güneş[1], Mehmet Adnan Öztürk[1], Selim Kurtoğlu[1]
[1]Department of Pediatrics, Division of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Pediatrics, The World Hospital, Kayseri, Turkey

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Introduction: Congenital gingival granular celled tumor (congenital epulis) is a rare benign pathology in newborns. It was defined by Neumann in 1871 for the first time and is known as Neumann tumor. It is 8-10 times more common in girls. The hospital referral reason for the patients is a mass preventing respiration and/or nutrition. Later enlargement is not expected in the epulis dimensions present at birth. Resection of the mass is satisfactory for treatment. When examined histopathologically, gathered and widespread polygonal cells which include a small round or oval nucleus and a dense granular cytoplasm are observed. In this essay, a rare congenital epulis case which didn't have any problems in prenatal follow-ups but was referred to the hospital with a mass inside the mouth at birth is presented.
Case report: A female newbornwho had a caesarean operation in an external centre three hours ago was transferred after her mother observed an exophytic mass in the oral cavity and was hospitalized in Erciyes University Faculty of Medicine Newborn Clinic. A nearly 1 cm mass hanging outside the mouth and leading with its stem towards the right side of the uvula inside the mouth was observed in the laryngoscopic examination. Enteral feeding of the patient who didn't have a significant respiration problem was cut against the risk of aspiration and oesophagus obstruction. It was considered that congenital epulis which is a rare disease was present in the patient who referred to the hospital due to a mass inside the mouth which didn't constitute a problem in prenatal follow-ups but was present in the mouth at birth and the mass was detected outside the oral cavity. No other congenital anomalies were detected and the patient was operated. The mass was excised easily from the gingiva it was based in with a monopolar cautery. The patient who didn't have any complications and wound recovery problems in postoperative period and enteral feeding was started on the 1st day after the operation and the patient was discharged on the third day. No recurrence was observed on the follow-ups. Pathological diagnosis was congenital granular celled tumor.
Result: As a result, diagnosis and treatment can be provided by the surgical resection made in congenital epulis in early period. No recurrence was reported in literature. Although the size of the mass and the urgency of the operation according to the clinic changes, primary aim of excision in general is to solve feeding and/or respiration problem of the infant. No recurrence finding was detected in the eight-month follow-up after the simple excision applied in our case.

DIAGNOSTIC ROLE OF INFLAMMATION MARKERS IN CHILDHOOD BRUCELLOSIS

Şefika Elmas Bozdemir[1], Yasemin Ay Altıntop[2], Salih Uytun[1], Hümeyra Aslaner[1], Yasemin Altuner Torun[1]
[1]Department of Pediatrics, Kayseri Training and Research Hospital, Kayseri, Turkey
[2]Department of Microbiology, Kayseri Training and Research Hospital, Kayseri, Turkey

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Background: Brucellosis is an endemic zoonotic disease which causes significant health problem in Turkey. Brucellosis patients present various non-specific clinical symptoms, such as fever, fatigue, sweating, jointpain, arthritis, myalgia and headache. Based on the nonspecifity of its clinical signs and symtoms, we decided to evaluate whether mean platelet volume (MPV), neutrophil to lymphocyte ratio (NLR), and platelet to lymphocyteratio (PLR) will contribute to the diagnosis on admission.
Methods: In this retrospective study, were viewed hospital-records of 60 children with a confirmed diagnosis of brucellosis in Kayseri between January 2013 and January 2016, and compared the hematological parameters; White blood cell count (WBC), hemoglobin (Hb), neutrophilcount, lymphocytecount, plateletcount, MPV, NLR and PLR with 55 healthyageandgendermatchedchildren.
Results: We found significant difference among the hemoglobin, plateletcount, MPV and NLR values between the patient and control groups ($p<0.05$). There was no difference between WBC, neutrophilcount, lymphocytecount and PLR between the patient and control groups ($p>0.05$). The presence of brucellar arthritis was not related to serum aglutinationtiter ($p=0.507$) and blood culture positivity ($p=0.646$). When the patients were divided into groups as arthritis positive and arthritis negative and compared to the control group; we found that the NLR is moresignificant in between the arthritis positive and control group ($p=0.013$). Most of the patients' (36.7%; 22/60) drug regimen consisted of a combination of rifampicin plus doxycycline.
Conclusion: The results of this study indicates that MPV and NLR values can be used as markers of inflammation in childhood brucellosis. Also, NLR is more valuable in children with brucellar arthritis.

DRAMATIC RESULT OF FOLIC ACID DEFICIENCY: ANENCEPHALY-ENCEPHALOCELE

Ahmet Özdemir[1], Musharraf Baghırova[2], Cihangir Shahin[2], Sabriye Korkut[1], Mahir Ceylan[1], Tamer Gunesh[1], Selim Kurtoğlu[1], Mehmet Adnan Özturk[1]
[1]Department of Pediatry, Service of New Born, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatry, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Anencephaly is the congenital deficiency of a significant part of brain, cranium and sculp. Neural tube defect (NTD) is the most commonly detected problem in prenatal period. Cranial defects are diagnosed easily in early fetal examination and prenatal consultancy should be provided to the family. A newborn case with NTD developing due to folic acid (FA) deficiency is presented in this article.

Case Presentation: Male patient, with a birth weight of 3020 gr, Mother didn't use FA during pregnancy. NTD was detected in intauterine 20th week. There was anencephalic appearance in the physical examination and a pouch in line with encephalocele. There was no significance in other system inspection. Encephalocele including a 4 cm heterogeneous brain parenchyma was detected in the occipital area. Brain magnetic resonance imaging findings were in line with ultrasonography. An operation is planned for the case taking consent from the family and the follow-ups are continued.

Result: The incidence of anencephaly and encephalocele gradually decreases with the prenatal ultrasonography and maternal serum scannings becoming widespread. It can be seen mainly due to deficiency of maternal FA. At least 400 µgr FA should be given in order to provide protection against NTD in reproductive age. In a female who had given birth to an anencephalic child before, 4000 µgr FA per day should be started at least a month before conception. This case is presented to re-emphasize the importance of FA use which is very important to decrease NTD incidence.

EVERY FACIAL PARALYSIS, IS NOT BELL'S PALSY!

Özlem Gül Kırkaş, Pelin Kacar, Cem Geyik, Alper Özcan, Mehmet Akif Özdemir, Türkan Patıroğlu, Musa Karakükcü, Ekrem Ünal
Department of Pediatrics, Division of Pediatric Hematology Oncology, Erciyes University School of Medicine, Kayseri, Turkey

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Background: Peripheral facial paralysis (FP) is seen in childhood with a incidence of approximately 20/100000. The idiopathic facial paralysis (Bell Palsy) is the main cause however infection, trauma, inflammatory and metabolic disorders, vascular malformations, cancer can be underlying disorder of is located. Patients with acute lymphoblastic leukemia (ALL) can rarely present with FP.
Aims: In this paper, two cases of ALL were reported whom steroid treatment had initiated after a mis-diagnosis of Bell's palsy.
Methods: Case 1: Thirteen-year-old boy were evaluated in ENT clinic with complaints of swelling behind his left ear, facial asymmetry, incomplete closure of left eyelid. It was learned that steroid therapy was started because of suspected Bell's palsy. It was learned that the decline of symptoms were experienced with the treatment. In laboratory studies on the recurrence of the same symptoms WBC was 48,000/mm3, hemoglobin 12 g/dL, platelet 112000/mm^3, BUN 19 mg/mL, creatinine 2.74 mg/ml, uric acid: 10 mg/mL, LDH: 5451 IU/mL. Lymphoblasts were observed in the peripheral blood smear. The cerebrospinal fluid (CSF) revealed leukemia involvement, The ALL IC-BFM ALL 2009 protocol was initiated, and the symptoms of the facial palsy were diminished with the treatment.
Case 2: Five years old girl with symptoms of toothache, and facial asymmetry was started a steroid therapy with a diagnosis of peripheral FP by the child neurology in another hospital. The patient was referred to use because the leukocyte was 145.870/mm3, hemoglobin 12.9 g/dL, platelets 108,000/mm^3. The peripheral blood smear and flow cytometric analysis showed T-ALL so the 2009 ALL-IC BFM chemotherapy protocol was initiated. CSF cytology showed class V with lymphoblast at CSF. The facial paralysis was improved after the chemotherapy treatment.
Results: Albeit it is rare, facial paralysis can occur as an initial sign of leukemia in children.
Conclusion: The detailed physical examination of the patient with acute facial paralysis, the evaluation of the complete blood count and peripheral blood smear is very important for suspected children. This strategy will prevent the delay of the diagnose underlying diagnosis of the underlying disease will reduce the morbidity and mortality rates.

HOME MECHANICAL VENTILATION IN CHILDREN: ERCİYES UNIVERSITY EXPERIENCE

Nazan Ülgen Tekerek, Adem Dursun, Başak Nur Akyıldız
Department of Pediatrics, Pediatric Intensive Care Unit, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Advances in pediatric critical care have reduced childhood mortality; however the nunber of children with chronic respiratory failure requiring long-term ventilation support is increasing. Over the last 30 years, there has been unprecendented emphasis on home care for chronically ill children. As a consequence of technological advances for children who are chronically disabled but medically stable and dependent on ventilatory assistance to live, home care is an approriate alternative. The aim of this study to evaluate underlying diseases, prognosis and ventilation modes of home ventilator-dependent children.
Methods: We retrospectively analyzed the medical records of 19 children with chronic respiratory failure in whom Home Mechanical Ventilation initiated between January 2008 and December 2015 at Erciyes Erciyes University Medical Faculty Pediatric Intensive Care Unit.
Results: Nineteen patients were identified on HMV in the present study. Eight patients were female and 11 patients were male. The median age at the time of HMV initiation was 77,6 month (7-204 month). Fifteen patients were ventilated using tracheostomy with SIMV-PS mode and 4 patients were ventilated using oronasal mask with S-T mode. The diagnosis of patiens were muscular dystrophy(n= 5), neurometabolic disease (n=59, spinal muscular atrophy (n=4), cerebral palsy (n=4), osteogenesis imperfecta (n=1). Five patients died on follow-up.
Conclusion: HMV is currenlty used in a variety of conditions. It reprsents a valid alternative to long hospitalization for children with stable chronic respiratory failure. HMV can be safely applied in selected children.

GIGANTIC SACROCOCCYGEALTERATOMA WITH BLEEDING: CASE PRESENTATION

Ahmet Özdemir[1], Tamer Güneş[1], Aysun Albayrak[2], Alper Özcan[3], Sabriye Korkut[1], Mahir Ceylan[1], Ekrem Ünal[3], Mehmet Adnan Öztürk[1], Selim Kurtoğlu[1]

[1]Department of Pediatrics, Division of Newborn, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Pediatrics, Division of Pediatrics, Division of Pediatric Hematology and Oncology, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Sacrococcygealteratoma (SCT) is the most common germ celled tumor in newborns and it is seen once in every 27.000-40.000 births in average. Neurological, cardiac, urogenital and skeletal-muscle system-related anomalies may rarely accompany these tumors. Teratomas reaching very big dimensions by growing in in-utero period were reported. A mortal gigantic SCT case with bleeding diagnosed in intrauterine 20th week in a new born was presented in this essay.

Case Presentation: 31 weeks old, 2950 gram male patient born with caesarean operation (SCT diagnosis reason for intauterine 20th week) as the first alive birth from the first pregnancy of a twenty five year old mother was intubated after resuscitation in delivery room and was hospitalized in the newborn intensive care unit. A 17x15x10 cm teratoma with intense vascularization in sacrococcygeal area and active bleeding was present. Erythrocyte suspension, frozen fresh plasma and thrombocyte suspension was given to the patient who had anaemia, thrombocytopenia and active bleeding from the lesion area. In the craniospinal magnetic resonance imaging, it was detected that the spinal cord ended in L1-2 vertebra level and a gigantic SCT with the dimensions of 16x11x14 cm in sacrococcygeal area leading to the interior of the pelvis up to sacrum anterior, containing cystic components with partial levelling, solid areas demonstrating contrast involvement and microcystic areas with intense content were detected. Alphafetoprotein level was detected as >50000 U/mL and human chorionic gonadotrophin level as <2.00 ng/mL. Patent ductus arteriosus, tricuspid valve regurgitation and pulmonary hypertension was detected in the ecocardiographic examination made for cardiac anomalies which may be accompanying. An operation was planned for the patient but it was impossible due to tumorlysis syndrome development and unstable general condition. The patient who had a cardiopulmonary arrest and didn't respond to resuscitation was accepted as exitus. The result of the biopsy taken postmortem was detected as sacrococcygealteratoma.

Result: Although the etiology of the teratomas is not completely known, the diagnosis can be made with prenatal ultrasonography in general. A close hemodynamic observance is important since bleeding is frequent in SCT cases reaching big dimensions.

KRABBE LEUKODYSTROPHY: A CASE REPORT

Adem Topcu[1], Gül Demet Kaya Özçora[2], Sefer Kumandaş[2], Mehmet Canpolat[2], Fatih Kardaş[3], Songül Gökay[3], Süreyya Burcu Görkem[4]

[1]Department of Paediatrics, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Paediatric Neurology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Child Nutrition and Metabolism, Erciyes University School of Medicine, Kayseri, Turkey

[4]Department of Paediatric Radiology, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Krabbe leukodystrophy (or globoid cell leukodystrophy) is an autosomal recessively transmitted progressive neurodegenerative disorder characterized by accummulation of galactosyl ceramide, lactocyl ceramide and galactosyl sphingosine due to deficiency lysosomal galactocerebroside, beta galactocidase enzyme. In our case with feeding difficulty and no head control since birth has been reported here to emphasize that Krabbe leukodystrophy may be a factor to consider in definitive diagnosis of patients presenting with swallowing dysfunction.

Case Report: A 6 months old male infant admitted to the hospital with the complaints of feeding difficulty and irritability. On physical examination; no head control, no eye tracking of objects or smile, increased deep tendon reflexes, bilateral babinski signs, hypertonicity, and tonic spasms dependent on touch and voices were detected. Cranial MRI revealed reduced bilateral thalamus signal intensity and thickened optic chiasm whic is suggesting Krabbe leukodystrophy. To confirm the diagnosis, the patient was referred for EMG and was consistent with neuropathy. The lysosomal enzyme levels studied and galactoserebrosidase level found 0.08 (N: 0.8-4). Genetic counselling was given to the family. In the follow up gastrostomy tube was inserted into the stomach for feeding diffuculty. Baclofen treatment was given for spasticity.

Discussion: Krabbe leucodistrophy is a lysosomal storage disease affecting deep white matter in its early stages. Due to galactocerebrosidase enzyme deficiency, galacto-sphingosine accumulates, which causes central and peripheral nervous system neurotoxicity by inhibiting myelin formation. In the infantile form, symptoms are usually present between 2 to 5 months following birth. Decerebrate posture develops quickly and many cases die around the age of two. Our case presented with irritability and feeding difficulty at 2 months, followed by contractions. The main difference between the juvenile and early onset type is that no peripheral neuropathy is present in the former. In our case, EMG findings were indicative of neuropathy. The MRI scan in our case was evaluated as compatible with the infantile type. The ultimate diagnosis is established by measurement of galactocerebrocidase enzyme activity in peripheral blood leukocytes or fibroblastic cell cultures. In symptomatic patients, supportive treatment for irritability and spasticity is provided. Although it is reported that stem cell transplants may be beneficial for infantile and juvenile Krabbe cases in the pre-symptomatic period, the long-term effects of this application are unknown. Due to the fact that the symptoms were obvious in our case, no such option for treatment was taken into consideration. In cases where there is irritability and extensor spasms occur due to touch or voices, and where cranial MRI shows white matter involvement and thickness of optic nerve, the possibility of Krabbe disease should be taken into consideration.

LIVER DISFUNCTION AS A RESULT OF CHILD PHYSICAL ABUSE: A CASE REPORT

Neslihan Günay[1], Adil Bozpolat[1], Meda Kondolot[2], Duran Arslan[3]

[1]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

[2]Division of Social Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

[3]Division of Pediatric Gastroenterology, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Child abuse is a global problem with short and long term physical and mental effects and social consequences**.** Abdominal trauma is the second cause of mortality after head trauma. Abdominal trauma may result in spleen and liver injuries, stomach, small intestine, bladder perforation or laceration, or pancreatic pseudocysts. In this paper, a case of liver disfunction as a result of physical abuse was discussed.
Case Report: Sixteen-month-old female patient was referred to our hospitalby her family doctor because of the detection widespread bruises on her body and her doctor also gavenotice to the police. The child was brought with complaint of vomiting to the Family Health Center. She was healthy beforeand she was in the care of her stepmother during the last three months. In the physical examination, her body weight was 7500 grams(<3p) and multiplebruises in different stages on her face, body, arms, and legsand hepatomegalywere detected. Laboratory testingrevealed AST:2585 u/L, ALT:3376 u/L, PT:22 sec, aPTT:41 sec, and albumin:2.8 g/dL. Increase in hepatic echogenicity and free fluid were reported in the ultrasonography. We did not detect any other reason that could be causeliver dysfunction. The clinical and laboratory findings were improved with symptomatic treatment. (AST:43 u/L, ALT:29 u/L, PT:11 sec, aPTT:26 sec, albumin:3,5 gr/dl). Left distal radius fracture was identified on her full skeletal survey. There was no retinal hemorrhages. Cranial CT was normal. Child protection team filed a report with the police for physical abuse. The child'sgeneral condition improved and adequate weight gain were observed. Social services was informed and she was delivered to the child protection agency official with the decision of the prosecutor.
Conclusion: Abdominal injuries due to physical abuse usually occurafter blunt trauma and early recognition is importantbecause of the mortality risk. Liver was reported as the most frequentlyinjuredorganand liver injuriescan be asymptomatic ormay cause life-threatening bleeding. In this case, liver disfunction as a result of physical abuse was diagnosed according to history, physical examination with different stages of bruises and hepatomegaly, laboratory and radiologic findings. However, it was not detected any other reason that could be causeliver dysfunction. It is important to make a careful assessment of the abdominal injury in cases of physical abuse, and physical abuse should also be considered in the differential diagnosis of liver dysfunction.

ONYCOMADESIS AFTER HAND-FOOT AND MOUTH DISEASE IN A CHILD

Zeliha Güzelküçük
Department of Pediatrics, Ahi Evran University School of Medicine, Kırşehir, Turkey

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Entry: Eruptive disease is the wide group disease that is mostly seen at childhood period and also its diagnosis is quite wide. Hand-foot and mouth disease which is taken part in eruptive diseases, is consisted of fever and hand-foot and mouth with vesicular lesions. Entereroviral infections are seen among especially the children under 5 years old. Here the case is presented with the progress of onychomadesis that has skin eruptive and fever, extension of time in recovery.
The case: The 10 years old male patient is evaluated for the fever and the rubor on hands at the pediatrics polyclinic. On his physical examination; there was orafarinks hiperemic, mouth mucosa is dry, and partly vesicular desquamations on palm and sole. The other system examinations are normal except his weakness; his eyeballs are a little decadent and dehydrated. The intravenous treatment is given to patient because of the decrease in oral taken and the appearance of dehydrated.
The result: Hand-foot and mouth disease is a kind of enteroviral infection and it's really infectious. Especially infections following with enterovirus 71; rarely fatal complications such as encephalitis, aseptic meningitis, acute paralysis, myocarditis, pulmonary edema could be accompanied. There is only improved onychomadesis in our case and in the following it is regress automatically. The enteroviruses are essential in the progress of onychomadesis that is connected to direct or indirect effects in nail beds. There isnt antiviral agent in the treatment of hand-foot and mouth disease. It isn't suggestible inhibiting direct contact, disinfecting the contamined belongings, washing hands and not squeezing vesiculitis in the protection period. We aim to draw attention to the hand-foot-mouth disease which is increasing nowadays by presenting the case at the out of ordinary age range whose skin eruption are get better with desquamation, onychomadesis on hand and foot nails.

POSSIBLE RISK FACTORS AND INCIDENCE OF FEMORAL VASCULAR COMPLICATIONS IN THE CHILDREN AFTER CARDIAC CATHETERİZATION

Özge Pamukçu[1], Süleyman Sunkak[1], Burcu Süreyya Görkem[2], Nazmi Narin[1], Ali Baykan[1], Kazım Üzüm[1], Selim Doğanay[2]
[1]Division of Pediatric Cardiology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Division of Pediatric Radiology, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Nowadays as the frequency of cardiac catheterization increase the complication rates also increase. The most common complications related to catheterisation are the femoral vascular ones. Therefore in this study we aimed to determine the frequency of vascular complications related to cardiac catheterisation, to find out possible risk factors and take precautions.
Material Method: In our center between the dates 2014-2015 years, 106 patients who underwent cardiac catheterisation were included in the study. Patients' age, weight, procedure type, sheath diameter, duration after procedure, number of catheterisations that were done, complications during or after the procedure, additional health problems were gathered from the patients' records. Bilateral external iliac, superficial femoral artery, vein of each patient were evaluated with ultrasonography and flow patterns were measured by Doppler ultrasound.
Results: Totally 106; 45 (42,5%) male, 61 (57,5%)female patients were included. Mean age of male: 21 (6-69), female 42 (5, 75-72) months. Weight of patients atthe time of catheterisation were; male: 10kg (6, 35-15, 5), female: 13,5kg (6,8-18,8). Mean of total time period after catheterisation was 28,5 (7,75-60,25) months. Two patients had 4 times, 9 patients had two, three times and 86 patients had only one time of catheterisation (Table1). Mostly right femoral vein was used (76 times, 41,9%), the least used vessel was left femoral artery (22 times,12,1%). Right femoral artery catheterised 56 times (30,9%), left femoral vein was 27 times (14,9%). Most common used sheath diameter was 5 French (122 times, 67%), the largest sheath that was used: 10Fr (3times right femoral vein, once left femoral vein). The smallest sheat was 4 French (right femoral artery 4, right femoral vein 2, left femoral artery 2 times). Mean procedure time was 44 (30-50)minutes.
No pathological situation was found in Doppler examination of total 105 patients. Only one patient, 18months old followed for VSD had hematoma in right inguinal region that was spontaneously regressed in 3months interval.
Conclusion: As a conclusion; risks of having complications get decreased as improving technology, using smaller sized sheath, deliveries and good anticoagulation.

HOLOPROSENCEPHALY CASE ACCOMPANIED BY A SINGLE NOSTRIL IN NEWBORNS

Mahir Ceylan[1], Aslıhan Akdemir[2], Nazlı Sultan Özsoy[2], Ahmet Özdemir[1], Sabriye Korkut[1], Tamer Güneş[1]
[1]Department of Pediatrics, Division of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatrics, Division of Hematology-Oncology, Erciyes University School of Medicine, Kayseri, Turkey
[3]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Accompanied by facial median line defects, holoprosencephaly is a complex developmental brain malformation caused by incomplete division of prosencephalon and resulting in insufficient development of forebrain structures. They are classified as alobar, semilobar and lobar according to the degree of septation deficiency. Prognosis of holoprosencephaly, which may also accompany syndromes as well as being isolated, is related to the severity of the brain and facial deformities and the presence of related anomalies. Microcephaly, hypotelorism, single nostril, choanal atresia and microgenitalia accompanied holoprosencephaly in our case, too. We thought that it would be appropriate to present the case due to its rareness and accompanying findings.

Case Report: Male patient born term as the first alive child from the third pregnancy of a twenty three year old mother was hospitalized in Newborn Intensive Care Unit after being intubated since he turned blue and was not crying right after birth. It was learned that his parents were third degree relatives and the first pregnancy was terminated in the 21st week due to fetal anomaly and the second pregnancy was ended with abortus at the seventh week. No chromosomal anomaly was detected in the chromosome analysis made on the amniotic fluid of the patient diagnosed with fetal ultrasonography on the 19th week of pregnancy. Microcephaly, hypotelorism, single nostril, choanal atresia and microgenitalia were detected in the physical examination. Cranial magnetic resonance (MR) imaging was reported as AlobalHoloprosencephaly. It was observed that cerebral hemispheres were agiric and thalamus and basal ganglions were fusioned. While ventricules had the structure of enlarged single ventricule, interhemispheric fissure, falx and corpus callosum were not observed. Nasal cavity, choana and nasopharynx were observed in atresic appearance in paranasal sinus computed tomography.

Result: We wanted to present our case since it is important as it is presented with AlobarHoloprosencephaly which is the severe form of Holoprosencephaly, its rareness and anomalies which may be accompanying.

NEUROBLASTOMA CASE REFERRED WITH RESPIRATION PROBLEM IN NEWBORN PERIOD

Mahir Ceylan[1], Alper Özcan[2], İbrahim Halil Tangüner[3], Aykut Poyraz[3], Sabriye Korkut[1], Ahmet Özdemir[1], Tamer GüneŞ[1], Selim Kurtoğlu[1], Ekrem Ünal[2], Mehmet Adnan Öztürk[1]

[1]Department of Pediatrics, Division of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Division of Hematology-Oncology, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Tumors are rare in newborns and nearly 2% of all childhood malign tumors are seen in newborns. The most common malign tumor seen in this period is neuroblastoma. Since neuroblastoma is an embryonal tumor caused by neural crest cells, there are cases diagnosed in prenatal period. Although congenital neuroblastoma has a benign course in general, it may also have an aggressive course. The first clinical finding in infants emerges mostly as metastasis related complications rather than the tumor itself. We thought that it would be appropriate to present the case since it is rare in newborns.

Case Report: Male patient, with a birth weight of 2750 gr, born term with caesarean operation in an external center as the third alive child from the fifth pregnancy of a thirty seven year old healthy mother was hospitalized in Newborn Intensive Care Unit with pneumonia diagnosis on postnatal 12th day. In the neck ultrasonography and magnetic resonance (MR) imagings made due to the swelling in the right side of the neck after postnatal third week, a solid component containing lesion with multicystic appearance was detected . Tracheotomy was opened due to the pressure on respiratory tract. Incisional biopsy on the mass was in line with neuroblastoma. Bone marrow aspiration (BMA) was made and no bone marrow spreading was detected. Whole body MR was planned and a metastatic spreading was not detected. A very good response was detected in the patient in whom TPOG Neuroblastoma 2009 chemotheraphy protocol was applied.

Result: Neuroblastoma can be rooted from any area where sympathic neural tissue is located and on the other hand it is caused by surrenal medulla and abdominal pelvic ganglions in more than half of the cases. In the presented surrenal area and abdomen images of the newborn, no tumoral existence was observed. BMA was normal. It should be kept in mind that respiratory problem in newborns is a rare referral characteristic of neuroblastoma.

THANATOPHORIC DYSPLASIA: CASE REPORT

Ahmet Özdemir, Mahir Ceylan, Sabriye Korkut, Tamer Güneş, Mehmet Adnan Öztürk, Selim Kurtoğlu

Department of Pediatrics, Division of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Thanatophoric dysplasia is the most common lethal neonatal skeletal dysplasia among skeletal dysplasias recognizable at birth. Being a lethal chondroplasia, it is seen once in every 16.000-20.000 births. A case born from a mother whose pregnancy couldn't be ended due to late intauterin diagnosis died in an hour because of respiration inability was presented.

Case report: Male patient, with a birth weight of 2380 gr (<3p), birth height of 46 cm (10p) and head circumference of 39 cm (>97p), born term with caesarean operation in our hospital as the third alive child from the third pregnancy of a thirty-seven year old mother was hospitalized in our Newborn Intensive Care Unit due to respiration problem and syndromic appearance. It was learned that the parents didn't have a consanguineous marriage and had two healthy sons aged 12 and 8. The mother didn't have any diseases or didn't use any medicine during pregnancy. It was stated that the mother didn't have regular pregnancy follow-ups and there were multiple anomalies in the baby in the 32nd week of pregnancy. The patient who had a severe respiration problem and whose oxygen saturation couldn't be increased with 100% oxygen support was intubated and connected to mechanic ventilator. Bradycardia developed in the follow-ups. The patient had an arrest and he died as he was not responding to resuscitation.

Result: Thanatophoric Dysplasia (TD) is characterized with short-limb dwarfism. It has two types; Type 1 and Type 2. While the mutations are observed in 7, 10, 15 and 19th exons in TD Type I patients, c.1948A>G (p.Lys650Glu) mutation is detected in exon 15 in all patients diagnosed with Type II. These patients whose clinical findings were similar to homozygote achondroplasia die a short time after birth due to narrow thorax and hypoplasic lungs. TD was presented since it is a rare disease and as it was considered that reminding would be advantageous to be able to provide genetic support to the family if diagnosed.

HEREDITARY TYROSINEMIA TYPE 1: NEWBORN CASE PRESENTATION

Ahmet Özdemir[1], Narmin Guluzade[2], Fatih Kardash[3], Songül Gökay[3], Sheyda Duzcuosmanogh[2], Safiye Gözübüyük[2], Sabriya Korkut[1], Mahir Ceylan[1], Mustafa Kendirci[3], Mehmet Adnan Öztürk[1]

[1]Department of Pediatrics, Division of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey

[2]Department of Pediatrics, Erciyes University School of Medicine, Kayseri, Turkey

[3]Department of Pediatrics, Division of Pediatric Metabolism, Erciyes University School of Medicine, Kayseri, Turkey

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Introduction: Hereditary tyrosinemia is a disease group caused by enzyme defects in tyrosine amino acid metabolism and is diagnosed in newborns or children. In type 1 hypertyrosinemia, hepatorenaltyrosinemia in other words, liver, kidney and nervous system are affected due to fumaryl acetoacetate hydroxylase enzyme defect. A case diagnosed with hereditary tyrosinemia type 1 diagnosis in newborns was presented in this article.

Case presentation: Male patient, with a birth weight of 3500 gr, was referred to the hospital due to coagulopathy and was hospitalized in newborn service for advanced examination and treatment. He had a seven year old brother who was diagnosed with hereditary tyrosinemia type 1 and his parents were relatives. In the controls, it was observed that his coagulopathy had improved. Tyrosinemia type 1 was diagnosed due to the metabolic examinations of the patient for whom consultation was made with paediatric metabolism department and the patient was fed with a special formula containing low tyrosine. Nitisinon treatment preventing the accumulation of more toxic metabolites was also added to his treatment. The patient's stable follow-ups are still being continued in newborn service and his discharged.

Result: Most of the infants die with an acute hepatic crisis before they are two years old in type 1 tyrosinemia. Consanguineous marriage, siblings with metabolic disease diagnosis are the most important tips in diagnosis. Tyrosine limitation, vitamin C replacement, use of NTBC use in diet are palliative solutions and liver transplantation is a treatment method providing cure. Early diagnosis is very important and our patient was diagnosed in newborn period.

TWO CASES WHICH ARE RECURRENT HIGH FEVER: PFAPA SYDROME

Zeliha Güzelküçük
Department of Pediatrics, Ahi Evran University School of Medicine, Kırşehir, Turkey

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Entry: PFAPA sydrom is a clinical pictures that is characterised with periodical fever, aphthous stomatitis, pharyngitis, servical lymphadenopathy. It should be thought as a differential diagnosis on the child who has repeater fever attacks.The high fever that brings to mind the infection diseases and not respond to antipyreticis the most important symptom of PFAPA sydrome. Here we present you the two cases in our hospital which are given treatment with antibiotic because of resistive high fever at epicentre.
Case 1: 26 months male patientis taken into the hospital. In his background; his fever is very high in last 6 months and not respond to antipreticis and hospitalized several times. there is not any characteritic in his family story. His anthropometric test are coherent with the age. In his physical examination; his body temperature is 39 °C and the other vital signs are at the normal limit. The tonsillar tissue is covered with the exudate lesions. In the frontier cervical there is not abnormality except too much micro lympadenopathy. It is demanded the complete blood cell count, C reactiveprotein (CRP), sedimentation, immunoglobulin levels and throat culture. It is not determine abnormality except at the border high white blood cell count and CRP. There is not reproductionin thorat culture. After two haurs of 1mg /kg applying intramuskular methyl prednisolone; there is not any high fever and not repeated.
Case 2: The 4.5 years old male patient consulted the our clinic. It is learned that in the epicentre; his high fever is depended the throat infection and started the treatment of intramuscular ceftriaxone. There is not any characteristic in his background and family story. His height and weight is coherent with his age. His body temperature is 39.2°C there is generalised exudative lesions on tonsiller tissue. There are bilateral two 1*1 cm, moving lymphadenopathy in frontier cervical. The other system examinations are normal. There aren't any abnormality except elevated white blood cell count in his laboratory assessment. After one haurs of 1mg /kg applying oral methyl prednisolone; there is not any high fever and not repeated.
Result: The PFAPA sydrome is clinically diagnosed after excluding the other diseases. Its exceedig the 39 °C and duration 3-6 days and repeating once of 3-8 weeks is the main characteistic of this disease. The case are completely healthy among the attacks. There is not any laboratory determiner special to the disease. The repeater high fever attacks can be hold years, but by the growing its freqvency is decreasing. During the disease; there is not long run requel. Nowadays by the rational drug use; we present you the two case that are used antibiotic by unnecessarily and we aim to remind you the PFAPA sydrome that is take part in high fever's rare causes.

XLAG SYNDROME CASE ACCOMPANYING A NEW ARX MUTATION AND HAS A INTERHEMISPHERIC CYST

Sabriye Korkut[1], Selim Doğanay[2], Ahmet Özdemir[1], Serkan Kurtgöz[3], Selim Kurtoğlu[1], Tamer Güneş[1]
[1]Department of Neonatology, Erciyes University School of Medicine, Kayseri, Turkey
[2]Department of Pediatric Radiology, Süleyman Demir University School of Medicine, Isparta, Turkey
[3]Department of Medical Genetic, Süleyman Demir University School of Medicine, Isparta, Turkey

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Background: XLAG (X-linked lissencephaly with ambiguous genitalia) syndrome which is a clinical spectrum of ARX mutations is presented with severe growth deficiency, abnormal genitalia and resistant seizures in neonatal period. We present a XLAG case which was formed due to a new ARX mutation and has an appearance of a huge interhemispheric cyst different from classic neuroradiologicalimagings.
Case: The case which was diagnosed with prenatal hydrocephalus and had prematurity, low birth weight and respiration problem was taken to the newborn intensive care unit. In the physical examination, hypotonic, spontaneous movements were very few in macrocephalic appearance and the patient had highly arched palate and the face was dysmorphic (hypertelorism, micrognathia, long filtrum, thin upper lip, low ear). Fallus height was measured 1.1 cm in the genital inspection, ventral hypospadias was present and the gonads were inpalpable. Starting from the postnatal second day, cyclic movements in facial muscles and multifocal clonic seizures began. Serological tests were negative when TORCH infections were considered. Uterus and gonad were not observed in pelvic ultrasonography. Corpus callosum agenesis, gigantic interhemispheric cyst, lissencephalia and olfactory gyrus absence were detected in brain magnetic resonance imaging. Karyotype analysis 46XY was found. A new homozygote p.Thr357Asnfs*175 (c.1068_1069dupA) mutation was detected in DNa sequence analysis.
Conclusion: While karyotype is 46XY in ARX mutations, external genital abnormalities can change between hypoplastic penis or undescended testicle and complete female appearance. So genital examinations of lissencephalic cases should be made carefully, clinical spectrums of ARX mutations should be considered in the presence of indefinite genitalia.