ROLE OF KALLISTATIN AND PRO, ANTI-INFLAMMATORY CYTOKINES IN PULMONARY HYPERTENSION

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Introduction: Kallistatin, serine proteinase inhibitor exhibits its properties in inhibiting inflammation, oxidative stress and apoptosis, angiogenesis. Severe pulmonary hypertension caused by left to right shunt congenital heart disease and Eisenmenger syndrome is progressive obliterative vasculopathy; the pathogenesis endothelial dysfunction and function of ion channels, calcium homeostasis, changes in platelet and endothelial function, intravascular thrombosis proliferation reactivity increased vascular inflammation and remodeling. In this study, we aimed to show the possible relationship between kallistatin levels, pro-inflammatory TNF-alfa and anti-inflammatory cytokine IL-10 levels in pulmonary arterial hypertension.

Material and Method: The study was performed in Erciyes University Medical Faculty Hospital. Patient group was formed from pulmonary hypertensive patients (Eisenmenger syndrome and primary Pulmonary Hypertension). We enrolled the patients with pulmonary arterial hypertension diagnosis that confirmed by the angiocardiography in our center. Pulmonary arterial hypertension is defined as mean pulmonary arterial pressure greater than 25mmHg. Serum kallistatin, TNF-alfa, IL-10, NtProBNP levels were studied in each patient.

Results: The study included total 50 patients: 25 patients in pulmonary hypertension group (16with Eisenmenger syndrome, 9 with primary pulmonary arterial hypertension). Control group consisted 25 patients with innocent murmur.Median value of kallistatin in pulmonary hypertension group was 1.42(1.0-1.58), control group 2.27(1.57-3.38).Serum levels of kallistatin were significantly lower (p<0.05) in pulmonary hypertensive patients. Negative correlation was detected between mean pulmonary arterial pressure and serum kallistatin levels. However no correlation was found for proinflammatory cytokine TNF-alfa and anti-inflammatory cytokine IL10.

Conclusion: Anti-inflammatory and anti-apoptotic features of Kallistatin were shown in various diseases like arthritis, pneumonia etc. Our study is the first one that shows the anti-inflammatory effect of kalistatin in pulmonary hypertension. Kallistatin levels were low in pulmonary hypertension because of increased lung inflammation. Since no correlation between IL10 and TNF-alfa was found, probably they used different pathways in the pathogenesis of inflammation.

CASE REPORT: IGA VASCULITIS WITH RESISTANT GASTROINTESTINAL FINDINGS

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Introduction: IgA vasculitis is a systemic small vessel vasculitis accompanied by gastrointestinal tract (GIT) and renal involvement. In these patients, abdominal pain accompanied by purpura is a major presentation at the time of admission.

Case report: An 11-year-old male patient was admitted to the hospital because of severe abdominal pain and purpura involving the buttocks and lower extremities. He was diagnosed as IgA vasculitis with severe GIT involvement. In laboratory evaluation, he had normal levels of hemoglobin (13.4 gr/dL) and hematocrit (%38) red blood cell 5.11 106 /mm3, leukocyte count 6900 /mm3, thrombocyte count 213000 /mm3, and ESR level 9mm/h. CRP was elevated as 22.1 mg/L (N: 0-5 mg/L). He had normal levels of PT, aPTT and INR, and normal urinalysis. The patient with severe abdominal pain was admitted to the hospital. Pediatric surgery consultation with abdominal ultrasonography showed no invagination and surgical pathology was not considered. Oral prednisolone 1 mg/kg/day was administered; however, the response to the treatment was not received. Resistant GIT involvement was considered, IV pulse methylprednisolone of 30 mg/kg/day was given consecutively for 3 days. Further investigations were planned and endoscopic biopsy revealed Helicobacter pylori infection, which was thought to worsen the prognosis. Factor 13 level was normal. Regarding the association of resistant IgA vasculitis with FMF, MEFV analysis resulted in M694V/R202Q compound heterozygosity. H. pylori eradication therapy together with daily colchicine for FMF resulted in complete remission.

Conclusion: IgA vasculitis is often accompanied by severe gastrointestinal symptoms. One should emphasize the association of FMF as a monogenic autoinflammatory disease, and coexistent H. pylori infection as contributing factors of resistant severe gastrointestinal involvement.
PERCUTANEOUS VSD CLOSURE UNDER 1 YEAR OF AGE

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Introduction: Main aim of this study is to share our experience about percutaneous Ventricular Septal Defects (VSD) closure under 1 year of age. Untreated large VSD are important reason of congestive heart failure in early infancy. This population usually fails to grow and surgical closure is challenging because of congestion in their lungs prone to respiratory infection and their bad nutritional status.

Material and Method: We have performed VSD closure of 11 patients under 1 year of age between the dates December 2014- January 2017 in our university Pediatric Cardiology Department.

Results: Age of patients ranged between 2 months-12 months. Weight of the patients during the procedure was between 7±2.7 kg. Median VSD diameter was 3±0.84mm. One of defects was muscular, rest of them were perimembranous. All defects were closed with Amplatzer Ductal occluder II (ADO-II). Mean fluoroscopy duration and total radiation dosage were 22.6 ± 18.7 minutes and 1674 ±851 cGy/min respectively. We did not face with any major complication except in one patient: complete AV block was seen one month after the procedure. Pacemaker was implanted. No aortic regurgitation associated with device was seen in patients.

Conclusion: The procedure of VSD closure, whether it is surgical or percutaneous, is very risky. The risks were higher when the children were smaller than 1 year of age and low body weight. Percutaneous VSD closure may be an alternative to surgery in early infancy that carry the similar risks but less invasive.

THE ROLE OF ENDOCAN AND KALLISTATIN IN THE DIAGNOSIS AND PROGNOSIS OF COMMUNITY ACQUIRED PNEUMONIA DISEASES

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Introduction: Measurement of plasma endocan and kallistatin levels at diagnosis and at day 4 of pediatric patients with community-acquired pneumonia (CAP). Determination of the role of these markers in diagnosis and prognosis.

Material and Method: 53 patients with clinical and radiologic diagnosis of pneumonia who were admitted to the Pediatric Chest Diseases Polyclinic and the control group were enrolled of 55 healthy children who applied to the general child polyclinic. Plasma kallistatin and endocan levels were determined using the ELISA method and the control group and patients were compared. In addition, the patient group was divided into 3 subgroups according to who were hospitalized and non-hospitalized, and who developed complication and no complication, who followed-up mechanical ventilation, and no follow-up. Plasma kallistatin, endocan and other inflammatory markers (CRP, ESR, PCT) were evaluated separately for each group.

Results: The plasma kallistatin level of the patient group was 1.604 (0.606- 9.700) ng/ml and the control group was 0.900 (0.550-2.250) ng/ml at the time of diagnosis. Kallistatin levels in the patient group were higher than control group, and the difference was statistically significant (p<0.001), but there was no statistically significant difference between patient and control group endocan levels (p>0.05). The patients were divided into 3 groups: complications, no complications, and deaths. In patients who died, kallistatin levels was statistically significantly higher than the other patients (p=0.022, p=0.026) at the time of diagnosis and on the fourth day of treatment, but there was no significant relationship between endocan levels (p>0.05). The kallistatin level of patients with mechanical ventilation (MV) was significantly higher than the non-MV patients at the time of diagnosis and fourth day of treatment (p<0.05). In addition, the patients who were followed up in MV, the level of the 4th day of the endocan was statistically higher than non-MV patients (p=0.018).

Conclusion: Serum endocan level has not been observed efficiency in the diagnosis and prognosis of CAP. Higher plasma kallistatin level was found to be significant in CAP. Patients with high kallistatin levels should be carefully monitored for unwanted side effects such as MV need and death.
UNCOMMON TYPE OF ASTROCYTOMA: PLEOMORPHIC XANTHOASTROCYTOMA

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**Introduction:** Pleomorphic xanthoastrocytoma (PXA) is a rare astrocytic tumor constitutes less than 1% of all astrocytic glial neoplasms. It was first described by Kepes et al. in 1979. As WHO classification of tumors, PXA is grade II tumor. PXA typically presents in later childhood or early adult life, presents itself first as seizures and focal neurologic deficits. Its most effective treatment procedure was not established because of its rarity.

**Case Report:** 9 years old male patient is evaluated for the new onset seizures and meaningless talking. He had resistant fever in that period, therefore he was followed in intensive care unit. Lumbar puncture was performed and antbiotherapy and steroid was started. His ECO, EEG, immune function tests were normal. His cranial MRI revealed 2 cysts in right cerebellum and 1.5cm hyperintense area in left occipital lobe. After that contrast MRI was consistent with the meningoencephalitis therefore screened for viral, bacterial and mycobacterial etiology. MRI of 1,5 months after event was revealed expansion of lesion. He was discussed at neuroradiology council and consequence was suggest of cranial biopsy. Histopathology of biopsy material demonstrated a pleomorphic xanthoastrocytoma. After surgery, he received postoperative therapy, which consisted radiotherapy and chemotherapy (cisplatin, vincristine, CCNU).

**Conclusion:** Pleomorphic xanthoastrocytoma (PXA) is an uncommon, low grade neuroepithelial tumor that occur in later childhood or early adult life, with predilection of the supratentorial brain without dural involvement, especially temporal lobe. Seizure is mostly present in PXA clinic. Tumor resection is mostly curative, but more likely to undergo spontaneous malignant transformation compared to other low-grade tumors. 9 to 20 % PXA may undergo malignant change can be recur and demonstrate aggressive clinical behavior with a mortality rate between 15% and 20%. Understanding of PXA’s natural behavior and prognosis have been hindered by its infrequency and the lack of studies with long follow up.

COMBINATION OF WERDNIHG HOFFMANN DISEASE AND CHRONIC RENAL FAILURE

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**Introduction:** Spinal muscular atrophy (SMA) is generally a neuromuscular disease with autosomal recessive inheritance and occur due to the mutations in survival motor neuron (SMN) gene. Pathogenesis of the disease is not completely known. Deletion and small intragenic mutations are detected in SMN1 gene. SMA type 1 or Werdnig Hoffman disease is the severest. The disease starts with birth, the infant is too loose, cannot hold his/her head, turn or sit, a severe respiratory difficulty is present and cannot survive without mechanic respiratory support. Suckling problems, respiratory difficulty, gastroesophageal reflux and tendency to aspiration pneumonia and infections can be seen in SMA patients in newborns.

**Case report:** The infant was born through caesarean from a twenty-five year old mother as the fourth alive birth from the third pregnancy in the 34th week with a birth weight of 2450 gr. It was learned that fetal movements didn’t decrease in prenatal period. Oligohidramniosis developed in the intrauterine thirty third week and early membrane rupture occurred and the patient was hospitalized in newborn intensive care unit after intubating due to superficial respiration. Low-set ear and weak hypotonic and newborn reflexes were detected in the physical examination. It was learned that the infant’s sister/brother had renal failure in newborn period and died when a week old. In the renal ultrasonography made due to lack of urination in the follow-ups in the first 48 hours, the left kidney dimension was 35mm and was evaluated to be in the lower limit of normal values. Creatinine increased to 1.7 mg/dl. Peritoneum dialysis was started. Gene analysis was made as the patient could have SMA since the reflexes were weak and hypotonicity was present. Deletion was detected in exon 8 area in SMN1 gene and SMA type 1 (Werdnig Hoffman disease) was diagnosed also considering postnatal age. The patient is being observed on mechanic ventilator in newborn intensive care unit and in elective conditions, kidney biopsy is being planned.

**Conclusion:** We wanted to present our case as it is the first newborn case with the combination of Werdnig Hoffman disease and non-functional renal failure according to the literature.
RARE CAUSE OF UNSTOPPABLE BLEEDING: HIRUDOTHERAPY
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Introduction: Hirudotherapy (leech therapy) is a popular treatment method that has gained popularity in recent years and has been widely used in previous times. It is used in many countries in the world, but has side effects such as bleeding, infection, allergy. In this case we wanted to discuss an unstoppable bleeding case after hirudotherapy in a twelve year-old male patient with back pain due to scoliosis.

Case report: A 12-year-old male patient who was followed up for scoliosis, was treated with hirudotherapy for the first time because of back pain. Right after the treatment, bleeding had started in the form of leakage in the back and waist region immediately. The child was referred to the emergency service because of the unceasing bleeding that continued more than ten hours. The patient has weakness and paleness when he arrived. The wound location on the back with the leech bite was cleaned with antiseptic solution and closed tightly with gauze. The patient was observed for the complications that could develop. The patient’s hematological tests (Hemoglobin: 13.2 g/dl, thrombocyte 214000 mm³, prothrombin time 13.6 secs, partial thromboplastin time secs: 29.3, bleeding time 4 min) and vital signs were normal. There was no history of drug use and bleeding disorders. The patient’s wound was re-cleaned after the three hours. Following the patient’s observation his hemorrhage was reduced and he had been discharged because his general condition was good.

Conclusion: Hirudotherapy is a long-standing complementary medical treatment method. But the indications and contraindications should be determined with scientific studies and should be applied by educated specialists. Even if the patient is already healthy, the patient should be investigated for any bleeding disorders before treatment. Since complications such as unstoppable bleeding, hypotension, anaphylaxis, sepsis and death may occur, the specialists should be careful during the treatment and the treatment should be administered in the centers where the complications could be interfered.

NEONATAL THYROTOXICOSIS CASE OCCURRING DUE TO STARTING NON-REQUIRED L-THYROIDINE IN TEMPORARY NEONATAL HYPERTHYROTROPINEMIA
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Introduction: While thyrotoxicosis is a presentation developing due to extreme thyroid hormone released from extrathyroidal tissues or giving exogen extreme thyroid hormone, the presentation caused by thyroid gland is called hyperthyroidism. While fetal and neonatal thyrotoxicosis presentation can be observed with the TSH receptor stimulating antibodies transferred to the fetus and fetal TSH response maturing after the 20th week, hyperthyroidism may also occur rarely in addition to the temporary hypothyroidism in neonatals depending on the antithyroid antibody type and level transferred from the mother in maternal Hashimoto disease. Discomfort, flushing, tachycardia, hypertension, inadequate weight gaining, enlargement of thyroid gland and exophthalmos may be observed as clinical findings.

Case Report: While st4 value of the female infant born interm with caesarean as the first live birth of the second pregnancy of a 27 year old healthy mother in the 38th week of her pregnancy was normal on postnatal 14th day, L-thyroxine treatment was started with a dose of 12.5 mcg/kg/day in an external center due to high TSH level. The patient was transferred from the neonatal polyclinic she was referred to with complaints such as discomfort, sweating, innutrition and fast breathing and was hospitalized in the service (Figure 1). The patient was monitored. P waves were present in the electrocardiogram and were in sinus tachycardia. The patient had thyrotoxicosis presentation with st4>7.7 ng/dl (0.89-2.2) TSH:0.016 mıu/ml (0.72-11.0) according to the TFT (thyroid function tests) sent and L-Thyroxin treatment was cut and the patient was followed-up. Furosemide treatment was started to increase diuresis. TSH was found 0.381 mıu/l according to the control TFT sent in the first week of hospitalization. St3:2.85 pg/ml (1.95-6.04) and st4:1.12 ng/dl were in normal borders. The patient whose general condition and symptoms recovered was discharged and came for polyclinic controls.

Conclusion: Decision should be made by following whether the neonatal hyperthyrotropinemia presented with normal St4 and high TSH is temporary or permanent. Follow-up without starting L-thyroxine is generally adequate. It shouldn’t be forgotten that unnecessary medicine use may cause neonatal thyrotoxicosis.
### A RARE CONDITION IN A NEWBORN DELIVERED BY CESAREAN SECTION: PING PONG SKULL FRACTURE

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**Introduction:** Depressed skull fracture in newborn is uncommon. It can occur as a result of two mechanisms. The first is iatrogenic (using forceps or obstetric manoeuvres) and the second is spontaneously because of the cranial vault flexibility. Other factors include maternal trauma, large uterine fibroids and foetal malpresentation. Skull depressions rarely have been observed in atraumatic deliveries. Here in, a newborn delivered by cesarean section with ping pong fracture was reported.

**Case report:** A 3180 gram male baby was born at 38 weeks of gestation by caesarean section. Pregnancy was uncomplicated with no history of maternal trauma. Physical examination revealed a 3x2.5 cm depression in the right temporo-parietal region. His neurological examination was normal. No instruments were used during the birth and no history of postnatal trauma was reported. There were no complaints reported by the family. He was admitted to department of Neurosurgery when he was 13-day-old. Computed tomography (CT) of the head revealed a ping pong fracture in the right temporo-parietal bone. Depressed fracture was elevated surgically and the newborn was discharged home uneventfully.

**Conclusion:** Neonatal skull depression can occur spontaneously without any history of trauma and also occur in the newborns born by cesarean section. They usually has a good prognosis. However, there is no consensus to treat these newborns. Treatment options depend on the severity of the fracture or on the degree of brain damage as determined by clinical examination or imaging. Clinicians must be aware of this kind of fractures and treatment options should be evaluated individually.

### UNCONTROLLED POWER IS NOT POWER AND UNCONTROLLED HYPOTHERMIA IS NOT A TREATMENT

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**Introduction:** HIE (Hypoxic ischemic encephalopathy) is an important cause for neonatal death, cerebral pulse and neurological dysfunction which may occur in late phase despite all developments in neonatal care. Efficiency of hypothermia which is the most effective treatment used for decreasing morbidity and mortality in HIE cases is directly related to the start of treatment in early phase (first 6 hours).

**Case report:** The case was born as the first live birth from the 1st pregnancy of an 18 year old mother in the 39th week of her pregnancy with a birth weight of 3200 grams through normal spontaneous vaginal route in an external center. The patient who had a story of a difficult delivery and perinatal asphyxia diagnoses was transferred after entubation due to hypothermia treatment requirement. Inside of the transport catheter was full of ice cubes when the patient arrived. Also the whole body of the patient was covered with ice cubes. It was detected that the peripheral circulation was extremely damaged and there was spread bleeding from umbilicus, mucosae and intervention places. Blood gas ph was 6.49, bicarbonate was 5.4, base deficit was -36.4 and measured axilliary body temperature was 27 °C at arrival. The case was heated in stages and then was taken in hypothermia treatment. Plasma and cryoprecipitate support was provided for DIC occurring due to asphyxia and deep hypothermia. Acute renal failure occurred in the patient followed-up and the creatinine value increased up to 6.14 mg/dl. Peritoneum dialysis was started by locating peritoneum dialysis catheter. In the cranial MR imaging, suspected minimal ischemic-hemorrhagic areas and appearance in line with resorbed subdural hemorrhage were detected. Electroencephalography was normal. As the renal failure, DIC presentation and general condition recovered, the patient was started to be fed and discharged and came back for controls.

**Conclusion:** While transporting the patient for hypothermia treatment which can be made in relatively new and limited number of centers, it should be based on correct application of passive cooling and principle of not harming the patient should form the basis.
HUMAN PARVOVIRUS B19 INFECTION PRESENTED WITH FEVER AND MACULOPAPULAR RASH IN NEONATES

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**Introduction:** Human parvovirus (HPV) B19 may cause syndromes with a different clinic such as erythema infectiosum, non-immune hydrops fetalis, temporary aplastic anemia and arthropathy. Main clinical presentations are fever, anemia, rash, arthropathy, hepatitis and optic neuritis.

**Case report:** A mature newborn baby was admitted to NICU with two days history of fever and rashes. His birth weight was 3800 grams. His grandmother and sister had a recent viral infection story. His physical examination was normal other than fever and rashes and splenomegaly on admission. After urine, blood cultures and viral serology were taken, he was given antibiotics. Peripheral blood smear and bone marrow aspiration were normal in terms of hemophagocytosis. His immunologic tests including immunoglobulin levels and lymphocyte sub-groups were normal except increased levels of immunoglobulin. Cerebrospinal fluid culture was also normal. Results for HPV IgG and IgM were positive. So HPV PCR was taken from the patient and HPV serology was taken from the mother, grandmother and sister. HPV of the patient was positive and parvovirus IgM and IgG of the grandmother and sister were positive and IgG of the mother was positive. In the follow-up, it was observed that the rashes increased and changed places during the day with a maculopapular character. Skin prick and patch test were normal. Brain MR was unremarkable for intracranial infection focus and abscess. Eye and fundus examination for optic neuritis was normal. His rashes was regressed and he was discharged on the 20th day of hospitalization.

**Conclusion:** HPV infection should also be considered in neonatal cases that do not have rashes, fever or any bacterial growth in the cultures taken from body fluids.

TRISOMY 13 CASE

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**Introduction:** Trisomy 13 is a rare disease seen nearly in 1 out of 10,000 live births. It generally ends with spontaneous abortus. 50% of the cases born alive die in the first month and 90% die in the first year. Most common accompanying anomalies are cardiac anomalies, cleft palate-lip, eye anomalies and nervous system anomalies. The final diagnosis is determined by chromosome analysis.

**Case Report:** The infant was born with caesarean in the 38th pregnancy week of a 34 year old mother with a birth weight of 4070 gr in an external center. Anal atresia, bilateral pes equinovarus, polydactyly, buffalo hump, squat nasal root, low and small ear and micropenis were detected in the delivery room examination of the patient who didn’t have any problem in intrauterine follow-ups. The patient was transferred to our unit for examination and treatment and was operated by Pediatric surgery for anal atresia. Subaortic ventricular septal defect and patient ductus arteriosus were detected in the ecocardiography made and the patient was followed-up. Microphthalmia (+) was detected in the left eye and coloboma was detected in the optic disk and macula in the right eye in eye examination. In brain magnetic resonance imaging, while simplified gural pattern, millimetric hyperintense focus in right parietal area (asphyxia, microhemorrhage?), trigonocephalic appearance, right bulb of the eye were normal, primary hyperplastic vitreous appearance in the left and also edema were detected in bilateral pre-auricular and mastoid area (Figure 1). Antiepileptic treatment was started as convulsion was also observed in the follow-ups. Grade 1 echo increase was detected in bilateral kidneys in the abdominal ultrasonographic imaging. The patient was followed-up as diuresis and kidney functions were normal. Chromosome analysis sent was interpreted as 47 XY, +13 Trisomy 13. The patient requiring full enteral feeding with nasogastric catheter and oxygen in incubator was discharged.

**Conclusion:** Trisomy 13’s survey is short due to multiple anomalies despite multidisciplinary approach and follow-up.
NEONATAL CASE WITH PANHYPOPITUITARISM RELATED TO CONGENITAL ARACHNOID CYST

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P13A

Introduction: Arachnoid cysts are cavities associated with the subarachnoid space, which contain cerebrospinal fluid. They constitute for 1% of all occupants in the brain. They are mostly asymptomatic. Most frequent clinical findings are headache, macrocephaly, convulsion, focal neurological symptoms and increased intracranial pressure symptoms. They can also rarely cause endocrine anomalies, optic neuritis, cerebellar dysfunction and hydrocephaly. In this report, we present a newborn case with antenatal detected arachnoid cyst presented with postnatal hypernatremia due to panhypopituitarism.

Case report: A female neonate, born from a healthy 24 year old mother on the 38th gestational week at 2700 grams, admitted to our hospital on her postnatal 9th day because of an arachnoid cyst seen on the 32nd gestational week in her antenatal follow-ups. On admission, her mother complained about her hypoactivity, absence of suckling and vomiting. Her serum Na level was 154 and reached to 166 mEq/L. Her urine output was 7cc/kg/hour. Desmopressin was started with a suspicious of central diabetes insipidus. Hormones secreted from the pituitary gland were low (ACTH, LH and FSH). Hydrocortisone was added because of low cortisol. Her cerebral MRI showed a mass measuring 71x48x50 mm sized located at the left lateral ventricle, compatible with arachnoid cyst. A minimal midline shift to the right was observed. On the 2nd day of medical treatment, her Na levels and urine output returned normal. However, the cyst was drained by neurosurgery on the 6th day of treatment to decrease intracranial pressure and prevent midline shift.

Conclusion: Conservative approaches are recommended for arachnoid cysts, which are usually asymptomatic and detected coincidentally. Cysts that show a mass effect or that are symptomatic should be treated surgically. This case was presented because no arachnoid cyst cases had been reported in the literatures that show endocrine symptoms at this early stage of life.

NEWBORN WITH EXTENSIVE MONGOLIAN SPOTS: A CASE REPORT

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Introduction: Mongolian spots, also known as congenital dermal melanocytosis are benign, congenital, single or multiple, blue-grey colored skin lesions. Usually, they are seen in healthy newborns and are inclined to disappear in the first years of life. While they are mostly sacral and lumbar located, they can also be gluteal, leg, back and shoulder located. Spots can be multiple, and their sizes can reach from a few centimeters to twenty centimeters. They usually don’t exceed 5% of the body surface area. They can develop at birth, as well as appear afterwards. Recent publications have reported that extensive, darkly pigmented mongolian spots which don’t disappear in childhood are related to mucopolysaccharidosis type 2 (Hunter Syndrome). In this report, we present a newborn with mongolian spots covering 70% of her body surface area.

Case report: A full term female neonate was hospitalized because of extensive skin lesions mimic Mongolian spots. The mother had goiter not required hormone replacement during pregnancy. Her uncle was also born with similar skin lesions completely disappeared in his childhood. Physical examination showed patchy macular skin lesions on the back, anterior torso and four extremities without any life threatening findings such as sepsis. Hemogram and coagulation work-up were normal. We considered her diagnosis as extensive Mongolian spots. Skin biopsy could not be performed due to family decision. One month after discharge, her growing was normal and skin lesions were partially regressed.

Conclusion: Mongolian spots usually do not need medical treatment and regress spontaneously in childhood. They can be confused with nevus of Ito and blue nevus if they appear on scalp, arms and face. Child abuse should always be considered as a differential diagnosis if there are multiple purple Mongolian spots. Multiple and extensive spots that don’t regress after many years should be regarded as an alert for metabolic diseases.
BERDON SYNDROME: MEGACYSTIS-MICROCOLON-INTESTINAL HYPERPERISTALYSIS; CASE REPORT

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Introduction: Berdon Syndrome is a rare autosomal recessive fatal congenital disorder characterized by megacystis-microcolon-intestinal hypoperistalsis. A mutation has been detected in the ACTG2 gene. The main reasons of poor outcome are malnutrition, sepsis, renal insufficiency or liver failure due to total parenteral nutrition. We will present a neonate with Berdon Syndrome in this article.

Case report: The baby girl who was delivered by a 23-year-old mother by G3P3A1 cesarean birth at 37th gestational week with a weight of 3100 grams was admitted to NICU due to distended abdomen. Radiologic investigations performed at antenatal period showed dilated cystic structure filling the abdomen and pelvis completely and bilateral pelvialectasis. Postnatal evaluation confirmed antenatal findings. Urine catheter was inserted and 690 ml urine was drained that decreased abdominal distention. X-ray imaging of the lower GI tract via contrast and abdominal MRI showed characteristics findings of microcolon and hypoperistalsism. Dilation was detected also in the bowel loops of the upper part of the microcolon. Ileostomy was performed because of absent of stool discharge at 13 th day of life. Total parenteral nutrition was started and intestinal transplantation was planned. Unfortunately, she passed away on the 32 nd day of the life. The genetic test was obtained from the baby and parents.

Conclusion: Berdon Syndrome is a rare disease characterized by impaired motility in the intestinal and bladder muscles. Berdon’s Syndrome should be kept in mind in patients with antenatal detected bladder dilatation and hydronephrosis, dilated the bowel loops. Suspecting from this disease in the prenatal period will affect our birth method and neonatal approach in this syndrome which is mostly fatal.

SPONTANEOUS PNEUMOMEDIASTENIUM AFTER SEVERE COUGHING; CASE REPORT

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Introduction: Pneumomediastinum, the mediastinal localization of the air after perforation in the respiratory or gastrointestinal system organs, is rarely seen in children. Most common symptom is chest pain. Typical sign of this condition is Hamman sign which is synchronized crackle sound at auscultation with the hearth beat. Although many underlying disorders caused increased intrathoracic pressure can lead to pneumomediastinum, it can be occurred without any reason. Treatment is directed at the cause, if necessary surgical treatment is performed. In this article, we present a child with severe cough and spontaneous pneumomediastinum due to tracheitis.

Case report: A 4-year-old girl was admitted with severe cough and shortness of breath. Her cough started four days ago and progressed, shortness of breath has just begun. On admission, her vital signs were within normal range. Oxygen saturation measured by pulse oximetry (SpO₂) was at 93 % in room air. On her physical examination, bilateral coarse crackles on chest and crepitations on both anterolateral sides of neck were detected. No pathology was detected in laboratory tests. AP and lateral chest X-ray showed intense subcutaneous air bronchograms on the neck which was compatible with pneumomediastinum. Chest CT detected free air particles around the clavicle and sternum, in cervical region, between muscles, at subcutaneous tissue, in peritracheal and esophageal region, at superior mediastinum and anterior thymus, at retrosternal localization, around of arcus aorta and its branches, in mediastinal and deeper neck fascia’s without any positive findings of intestinal perforation. Patient was diagnosed as spontaneous pneumomediastinum secondary to severe cough. Antibiotic treatment for the upper respiratory tract infection, hydration support and symptomatic treatment for cough was given. After 2 days, patient’s crepitation on the neck was disappeared. At the 5th day of treatment, chest X-Ray was normal and she was discharged from hospital with a good condition.

Conclusion: Although it is rare disease, spontaneous pneumomediastinum should be thought in children with severe cough as a differential diagnosis.
FOREIGN OBJECT RECOGNIZED RANDOMLY IN THE LEG: A CASE REPORT

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Introduction: The World Health Organization defines an accident as “an unexpected incident that occurs beyond human will, leading to physical and mental damage”. When classifying according to place and type, accidents are investigated as traffic, working, industrial, sport, school and home accidents. Home accidents are the ones that occur “at home, in the garden or around”. They fall into significant issues of community health care by being preventable, frequent, leading to death or disability and their high disease burden. Children are under a high risk of home accidents due to not being aware of dangers, being open to environmental risks and their interest about finding-learning. According to data from Turkey, accidents are ranked 4th at top death reasons of children aged 1-4 years. Raising families awareness about home accidents and improving their prediction is a significant factor in protecting children from home accidents. In this paper, we aimed to present a case where a foreign object was recognized randomly in the case’s leg with intention to raise awareness of home accidents that children experience.

Case report: A 10 month old male patient with fever and complaints of cough admitted to the Pediatric Emergency Polyclinic of our hospital. The patient who was hospitalized with a diagnosis of lower respiratory tract infection had a history of being born at gestational week 30 and being screened for 2 months in the Neonatal Unit. At day 2 of hospitalization, upon his mother referring about palpable stiffness under the knee, a long-fine mass at about 3-4 cm of size under the left knee on the front face of the tibia was detected with palpation. After an x-ray scan was obtained, a foreign object that was thought to be compatible as a needle was seen on the front face of the tibia. The family’s thought was that it could have been from the child’s hospitalization at the neonatal unit. However, it was thought that the incident was likely to be an accident, because the localization of the object was not compatible with regions for establishing vascular access. The foreign object was extirpated from our patient’s under local anesthesia by the department of orthopedics. The extirpated object was found to be a pin without top that had been used for the mother’s scarf. The fallen pin was thought to be located subcutaneously by accident while crawling because the patient was in his infancy. The patient was discharged after the lung infection also recovered after the extirpation of the foreign object.

Conclusion: Home accidents that place a material and nonmaterial burden to our community and affect especially children may greatly be avoided by primarily raising awareness of the families and the whole community.

COMBINATION OF BILATERAL PERSISTANT HYPERPLASTIC PRIMARY VITREOUS AND TOXOPLASMA INFECTION

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Introduction: Persistent hyperplastic primary vitreous (PHPV) is a rare benign disease in the globe and defines lenticular pathologies occurring due to dissolved embryological hyaloid artery and secondary changes in retina and globe. This condition is the second most common cause of leukocoria. PHPV is unilateral in 90-98% of the cases. Herein, we present a case with both bilateral PHPV and toxoplasma infection.

Case report: The male infant born to a twenty-nine year old mother in the 39th week of gestation through caesarean G1P1; was hospitalized due to cord entanglement and meconium aspiration syndrome. The body weight was 3300 gr (25-50p), height was 48cm (10-25p) and head circumference was 32cm (10p) on physical examination of the patient in whom intrauterine corpus callosum agenesis and pericardial effusion were detected. Pericardial effusion measuring 17 mm was detected in the postnatal echocardiography. Pericardiotest was performed and ibuprofen treatment was started. Corpus callosum agenesis and polymicrogyria were detected in the ultrasonography and magnetic resonance imaging (MRI). Toxoplasma immunoglobulin M was found positive in viral serology. Toxoplasma Polymerase Chain Reaction (PCR) was found positive in peripheral blood and cerebrospinal fluid PCR was found negative. Ophthalmologic examination showed bilateral PHPV. Orbital MRI was remarkable for increased vitreous density on right bulbus oculi and significantly hyper dense appearance on bilateral lens. Toxoplasmosis treatment containing pyrimethamine-sulfadiazine-folinic acid combination was started due to central nervous system involvement. The patient’s follow-up and treatment were continued and he was discharged due to stable vital findings and called for outpatient follow-up.

Conclusion: The incidence of bilateral PHPV is quite rare in the literature. Ophthalmoscopy and bio microscopic examination is the first method for the diagnosis. To best our knowledge; this is the first case with both toxoplasma infection and bilateral PHPV in the literature.
COMBINATION OF GLUTARIC ACIDURIA TYPE 2 AND BILATERAL POLYCYSTIC KIDNEY

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**Introduction:** Glutaric aciduria type 2 is an autosomal recessive neurodegenerative disease also affecting various organs in the body. A newborn case diagnosed with glutaric aciduria type 2 accompanied by bilateral polycystic disease was reported in this article.

**Case report:** A full term female infant was born with respiratory problem was admitted to NICU. Mom had two miscarriages. Her mom and dad were first degree cousin. On her physical examination revealed body weight 3030 gr (50-90p), height 52cm (90p), head circumference 35cm (50-90p) and the skin was pale and hypotonic. Abdominal wall was loose, abdomen was flat and the kidneys were palpable. Patient's urine smelled like sweaty feet. A Glutaric aciduria type 2 diagnoses was made with urine organic acid analysis and tandem mass panel. Micro cysts scattered in the kidneys were detected by abdominal ultrasonography. Ecocardiographic examination was normal. Continuous Renal Replacement Treatment (CRRT) was started due to metabolic acidosis and ammonia value at 295.6µmol/L. Ammonia increased to 1335µmol/L in the follow-up. CRRT was ended and peritoneum dialysis was started due to technical problems and the ammonia level of the patient decreased to 181.2 µmol/L. Carritine and rhodavin support was provided. Lipid restriction was made in total parenteral nutrition. On the follow-up, she experienced with sudden cardiac arrest did not respond to cardiopulmonary resuscitation and she died.

**Conclusion:** Characteristic sweaty feet smell is detected in glutaric aciduria type 2 patients depending on isovaleric acid accumulation as in our patient. As a result, we think that glutaric aciduria type 2 should be kept in mind in children with abnormal urine odour and polycystic kidney disease.

**NEONATAL WHITE ADDISON PRESENTATION: PRESENTATION OF TWO 21-HYDROXYLASE DEFICIENCY CASES**

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**Introduction:** 21-hydroxylase deficiency constitutes most of salt-depleting congenital adrenal hyperplasia (CAH) cases in newborns. Pigmentation may not develop in the cases. These are called "non-pigmented CAH" (white Addison). Two cases were presented in the article to emphasize the fact that pigmentation may not always be present in 21-hydroxylase deficiency cases and the significance of surrenal ultrasonography in diagnostic study.

**Case reports:** Case 1: It was learned that the 23 days old infant was taken to the hospital with complaints of discomfort-inability to gain weight was born in-term with a birth weight of 3260 gr and couldn't gain weight after the 10th day and she was restless. In the physical examination, her weight was measured as 3220 gr, height as 51 cm, head circumference as 35.5 cm and blood pressure as 93/56 mm/Hg. External genital structure had a masculine appearance, fallus was 2.3cm, bilateral scrotum was empty and gonad was inpalpable. Case 2: It was learned that the male infant brought to the hospital with diarrhoea and vomiting complaints on postnatal 20th day was born in-term with a birth weight of 3800 gr and diarrhoea-vomiting complaints started when the infant was 13 days old and that he couldn't gain weight. His height was 53cm, weight was 3650 gr, head circumference was 38cm and average blood pressure was 55 mm/Hg in his physical examination. In the genital examination of the infant with a low fontanel and dry mouth, it was observed that the testicles were in scrotum and the penis length was 4.5cm (normal: 2.5-4.5 cm). There was no pigmentation in the nipples-genital areas of the patients. Dehydration and surrenal deficiency were considered since thrombocytosis was present in addition to hyponatremia and hyperpotassemia. Surrenal glands were larger than normal in the ultrasonography. The hormonal findings of the infant taken in dehydration-surrenal crisis treatment were in line with salt depleting 21-hydroxylase deficiency. Genetic 21 hydroxylase deficiency and deletion in CYP21A2 gene in line with salt depletion form were detected in patients for whom hydrocortisone, mineralocorticoid and salt replacement treatment were started.

**Conclusion:** Presenence of pigmentation in CAH diagnosis which is the main reason for neonatal surrenal crisis threatening life and which should be treated with early diagnosis is important. But pigmentation increase may not always take place as in our case. Even though there is no pigmentation increase in newborn cases with presentation of dehydration, hyponatremia, hyperpotassemia and thrombocytosis, we would like to emphasize the necessity of making urgent surrenal-renal ultrasonography in addition to hormonal evaluations for CAH diagnosis.
MILLER-DIEKER LISSENCHEPHALIA SYNDROME
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**Introduction:** Miller-Dieker syndrome is characterized by dysmorphic facial appearance accompanied by microcephaly, lisencephalia and prominent forehead, bitemporal hollowing, upward slanting of the nostrils and micrognathia. Autosomal dominant transfer and 17p13.3 mutation or deletion is determined. Other findings accompanying the disease are fallot tetralogy (TOF), ventricular septal defect (VSD), pulmonary valve stenosis (PVS), intrauterine growth retardation (IUGR), cystic dysplasia of kidneys, cleft palate and cataract. A newborn case diagnosed with rarely seen Miller-Dieker syndrome was presented in this article.

**Case report:** The female infant born from a twenty-nine year old mother in the 38th week of her pregnancy through caesarean G6C6 with a birth weight of 1760gr was hospitalized in newborn clinic due to respiratory problem. No specialities were observed in her medical story. The body weight was 1760gr (<3p), height was 44cm (3p) and head circumference was 31cm (3p) in the physical examination. In the head-neck examination, prominent frontal area, micrognathia and low-set ears were present. TOF and pulmonary hypoplasia in the ecocardiographic examination made due to dysmorphic appearance, dilatation-colpocephalic appearance in lateral ventricules in transfontanel ultrasonography, corpus callosum agenesis and colpocephalic dilatation were detected in type-1 lissencephalia and bilateral lateral ventricles in brain MRG. TORCH serology was detected negative. 17p13.3 deletion was detected positive in the FISH analysis sent by considering Miller-Dieker syndrome due to SGA, cardiopathy and lissencephalia present in the patient. The patient was discharged in a stable condition and she will be followed by outpatient clinic in advance.

**Conclusion:** Lissencephalia is one of the most severe problems seen in the migration period of intrauterine brain development and is a severe developmental anomaly formed by normal gyrus pattern loss in cerebral hemispheres, disorganization of cerebral cortical architecture and generally accompanying severe neurological deficit and seizures. Miller-Dieker lissencephalia syndrome seen rarely in newborn may present with IUGR, TOF, lissencephalia and dysmorphic facial appearance.

PRIMARY GLAUCOMA: NEWBORN CASE PRESENTATION
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**Introduction:** Glaucoma is a general term defining the damage in visual field caused by or related to high intraocular pressure and optic disk damage. Primary glaucoma results from isolated anomaly on eye trabecular meshwork. More than 50% of congenital glaucoma is primary. Incidence of primary congenital glaucoma is 0.03% and a newborn case with primary glaucoma is presented in here.

**Case report:** In the first day examination of a female infant born from a twenty-two year old mother in the 39th week of her pregnancy through caesarean G2C2, she was hospitalized for examination and treatment due to corneal opacity detected. No specialities were observed in her medical story. Body weight was 2700 gr (10-50p), height was 47 cm (10-50p), head circumference was 37cm (>97p), chest circumference was 31cm and abdominal circumference was 28cm in her physical examination. In the ophthalmologic examination, the patient had bilateral mildly blurred-cloudy grey coloured corneas and very little enlargement in eyeballs in favour of glaucoma (Figure 1a-1b). Also bilateral aniridia and bilateral secretion increase-conjunctival hyperemia were detected. Eye drops containing acetazolamide, aminoglycoside, brinzolamide and timolol were started. According to the evaluation made under general anaesthesia during follow-up, right eye average pressure was 18.5mmHg and left eye average pressure was 22.5mmHg and blurity and aniridia were present in the right cornea. Bilateral cornea diameters were measured as 11.5 mm horizontally and vertically. These findings were evaluated as congenital glaucoma and corneal opacity. The patient was examined for neurometabolic disease and WAGR. The results of the examinations were normal. TORCH serology was detected negative. Transfontanel and complete abdomen ultrasonography was normal. Polymicrogyria was detected in cranial MRG. The patient was discharged in a stable general condition and the polyclinic follow-ups were continued.

**Conclusion:** 69.6% of childhood visual loss cases in our country are due to preventable causes. Eye examination-visual scanning is very important in newborns in order to detect risk factors to prevent normal development of sight and cases with unsatisfactory sight in early period.
CONTINUOUS RENAL REPLACEMENT THERAPY: NEWBORN CASE PRESENTATION

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Introduction: Even though peritoneal dialysis is the dialysis method generally preferred in renal failure in newborns, in literature it is reported that haemodialysis at intervals, continuous veno-venous hemofiltration or hemodiafiltration can be applied with success. Two cases with acute renal deficiency and hyperammonemia for whom continuous renal replacement treatment (CRRT) was applied were presented in this article.

Case reports: Case 1: The female infant born as one of the twins of a thirty three year old mother in the 28th week of her pregnancy through G1C2 caesarean with a birth weight of 1150gr was transferred to our hospital for patent ductus arteriosus (PDA) after living in an External center for 45 days. Her body weight was 2495gr (10-50p), height was 39cm (10-50p) and head circumference was 33.5cm (10-50p) in the physical examination and she was ligated with an urgent operation after vein rupture during PDA closure with angiography. Then CRRT was made after stabilizing the hemodynamics of the anuric patient. Peritoneal dialysis of the patient was made after a successful CRT lasting seven days and creatinine increase and diuresis were provided. Follow-ups are continued for the patient in our service. Case 2: Female infant born from a forty year old mother with a birth weight of 3030 gr in the 37th gestational week was hospitalized in newborn intensive care unit due to respiratory problem. Body weight was 3030gr (50-90p), height was 52cm (90p) and head circumference was 35cm (50-90p) in the physical examination and CRRT was applied to the patient diagnosed with glutaric aciduria type-2 and had multicystic dysplastic kidneys and hyperammonemia. CRRT was ended due to technical problems after an operation lasting fourteen hours and peritoneal dialysis was started. Cardiopulmonary resuscitation was applied since cardiorespiratory arrest occurred in the patient followed-up for hypoglycemia-metabolic acidosis. But the irrespnsive case died.

Conclusion: CRRT should be preferred in newborns due to advantages such as providing a treatment similar to kidney working due to extension to a longer period of time, better toleration by patients with poor hemodynamics, providing the drawal of more fluid and removal of materials with medium-large molecular weight.

HERLYN-WERNER-WUNDERLICH SYNDROME IN A NEWBORN CASE

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Introduction: Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital anomaly in female urogenital system and is related to mesonephric channel anomalies-müller channel anomalies. This syndrome is characterized by uterus didelphys, obstructive hemivagina and ipsilateral renal agenesis tirade. The objective of this article is to draw attention to the early diagnosis-treatment of HWWS in newborns which is observed rarely and diagnosed due to suspicion.

Case report: Female infant born in the 31st pregnancy week from a twenty-seven year old mother G2P2C2 through caesarean with a birth weight 1600gr was hospitalized in newborn clinic due to early membrane rupture (EMR) in the first 72 hours. The mother had renal agenesis-vaginal prolapus story and the body weight of the infant was 1600gr (50-75p), height was 41cm (50p) and head circumference was 28cm (50p) in the physical examination. Antibiotherapy was started for EMR for the patient with positive infection markers. Right renal agenesis was confirmed in the abdominal ultrasonography made on the 3rd day of hospitalization of the patient with intrauterine right renal agenesis. Pelvic USG and MRG were made for the inner genitourinary evaluation of the patient in whom prolapsus was detected in vaginal entrance especially when abdominal distension is present. Bilateral over agenesis, fluid collection in uterus, septum appearance in vagen distal 1/3 wall level and nearly 23x9mm cystic formation with a lobulated contour and full of fluid which is leaning towards the right adnexal area were detected. The case was evaluated as mini puberte since 123.7pg/mL estradiol, 2.13µg/dL cortisol, 192.2ng/mL prolactin, 22.08mIU/mL FSH, 3.74mIU/mL LH and 19.9-42.20U/mL CA were detected. Hymen was intact, vagina was normal, cervix uteri had irregular limited-prolapsed appearance in the cystopic examination and no cystic-tumoral structures were observed. In the light of the present information, HWWS was diagnosed and the patient was discharged due to a stable general condition in the follow-up and polyclinic follow-ups were continued.

Conclusion: HWWS is a rare disease with an etiology not clearly known should be considered in female infants with urogenital system anomaly and renal agenesis and family story should be questioned. We think that the studies in the future would enlighten the inheritance way of HWWS.
A RARE CAUSE FOR INTRATAUTERINE ASPHYXIA: CARBONMONOXIDE INTOXICATION

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Introdution: Carbonmonoxide (CO) is a colourless, odourless, tasteless and non-irritant gas and causes cellular anoxia showing 210-280 times more affinity to hemoglobin than oxygen. Fetus whose mother was exposed to CO poisoning is affected more than the mother as CO interest of fetal hemoglobin is higher and cleaning of carboxihemoglobin from fetal blood takes four-five times more time than the mother. This explains extremely severe fetal complications which may occur in pregnant women with average severity of complaints who were exposed to CO poisoning. Toxic effects such as fetal death, intrauterine malformations and functional changes occur in pregnant women acutely exposed to CO gas.

Case report: Female infant born as the first live birth from the first pregnancy of a 17 year old mother with caesarean in the 30th week of her pregnancy was hospitalized in neonatal service after birth as her mother was hospitalized due to maternal CO poisoning a week before and cranial hemorrhage was detected in the infant in the fetal MR imaging. Carbohemoglobin value was normal for the mother and cord blood gas. Cortex involvement and widespread white matter in both cerebral hemispheres, widespread cystic encephalomalacic changes in these locations, acute influence related diffusion restriction in both cerebral hemispheres, especially in frontoparietooccipitotemporal and left parietooccipital areas were observed and enlarged appearance, more significant in both lateral ventricule occipital horns was detected secondary to volume loss (Figure 1). Treatment was arranged for the renal failure in follow-ups.

Conclusion: The patient should be followed-up more closely and carefully in order to prevent the potential destructive effects of intrauterine CO exposure in the fetus, the mother should be referred to a center where she can receive hyperbaric oxygen treatment as soon as possible and application of birth decision without delay if required are very important during follow-up and treatment.

A RARE CASE FOR DIRECT HYPERBILIRUBINEMIA IN NEONATAL PERIOD: PATENT DUCTUS VENOSUS

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Introduction: Ductus venosus is a connection present in fetal circulation between left umbilical vein and right hepatocardiac channel. Ductus venosus staying open although it should normally be closed in a few hours after birth in term infants and in a few days in preterms may cause high liver enzymes and direct hyperbilirubinemia.

Case report: The patient born with a birth weight of 2260 grams in the 39th pregnancy week of a 29 years old mother was sent to our center due to direct hyperbilirubinemia based on the examinations made in the medical center they referred due to anxiety and jaundice when the infant was 20 days old. The patient didn’t have any known diseases and both the patient and the mother didn’t have any story of medicine use and direct hyperbilirubinemia and high liver enzymes were detected in the examinations of the case. Suspected venous malformation combining with dilated left portal vein lying inside the left lobe passing inside the wide hepatic vein branch separated from vena cava inferior was detected in the abdominal ultrasonography taken. Although PDV (patent ductus venosus) was detected in the abdominal CT and MR portography made when compared to the radiological evaluations made before (Figure 1). Viral serology and metabolic examinations were normal. Prothrombin time and activated partial thromboplastin time, albumin levels and alpha-1 antitrypsin value checked for liver failure were normal. Left peripheral pulmonary stenosis and thin PDA were detected in the ecocardiography. No additional operation was considered by interventional radiology for the patient whose PDV calibration decreased and liver function test results and bilirubin levels decreased. The patient was discharged and followed-up. In the control ultrasonography taken by the polyclinic, it was detected that the ductus venosus was closed.

Conclusion: We wanted to present our PDV case with a very rare reason for liver function disorder and direct hyperbilirubinemia.
HYPOCALCEMIA SECONDARY DILATED CARDIOMYOPATHY

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Introduction: Dilated cardiomyopathy was characterized by dilatation of the left or both ventricles and decreased contraction. Hypocalcaemia is a very rare cause of dilated cardiomyopathy, as it may be caused by idiopathic, genetic, viral, immunological or toxic. In this article an 8 month-old male infant with hypocalcaemia secondary dilated cardiomyopathy was presented.

Case report: An 8 month-old male patient was referred to our pediatric intensive care unit because of sudden widening of the tachycardia, tachypnea and general condition disorder. The glascow come score was 5 at his admission. He was intubated because of cardiac and respiratory system failure and he connected to the mechanical ventilation. The patient is diagnosed with decompensated heart failure and his drugs were regulated. In the laboratory results, calcium: 4,6 mg/dl, other biochemical and hematological examinations were normal. Calcium gluconate infusion was initiated as 75 mg/kg/day. Echocardiographic examination revealed dilated cardiomyopathy, decreased left ventricular function, ejection fraction %33. The he submitted 25-OH vitamine D3 for hypocalcemia etiology was 6,773 ng/ml (20-120), parathormone: 113,8 pg/ml (15-65). The patient was diagnosed with rickets due to vitamine D deficiency. The general condition of the follow-up was worse and the patient was lost on the 8th day of his admission.

Conclusion: Although hypocalcemia reduces heart contractility, hypocalcemia associated heart failure is rare. However, long-term hypocalcemia can seriously impair cardiac contractility and heart failure cannot be controlled without hypocalcemia. We would like to highlight that a clinician must be careful about serum calcium levels in cases with cardiomyopathy.

A PATIENT WHO HAD LIVED WITH THE LOWEST PH IN PEDIATRIC INTENSIVE CARE UNIT

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Introduction: Shock is an acute clinical condition in which tissue perfusion is not sufficient to meet metabolic requirements. If insufficient perfusion persists for a long time, various systemic and metabolic responses occur. As long as the shock continues, the compensation mechanisms are insufficient to meet the increased metabolic need at the tissue level. The first response is tachycardia when the cardiac output falls for any reason. When tachycardia cannot provide the sufficient cardiac output and oxygen distribution, the acidosis develop because of tissue hypoxia. We present a patient with the lowest pH in our case which developed secondary to hemorrhagic shock and followed up in pediatric intensive care unit.

Case report: A 9-month-old girl, who was operated by neurosurgery for craniosynostosis, was taken to the pediatric intensive care unit from the neurosurgical intensive care unit due to general condition disorder on the second postoperative day. The blood pressure was 50/30 mmHg, central venous pressure was 3 cmH2O at her admission. Patient’s capillary withdrawal time was prolonged (>5 sec) and were given inotropic agents, vasopressor support and saline challenge twice a day (20 cc/kg) Her blood gases were; pH: 6,56, pCO2: 104 mmHg, HCO3: 9,2 mmol/L, Be:-31,6, lactat: 16,32 mmol/L at her admission. Patient with trombocytopenia who had active bleeding from the incision site was treated with erytrocyte and platelete suspension, and vitamine K was administered. The cefotaxime and vancomycin therapy was empirically initiated because of fever. The patient with disseminated intravascular coagulopathy (aPTT: 100 sec, PT: 26 sec) was supplemented with fresh frozen plasma. Phenytoin was loaded and maintenance dose was given. The blood gas in the following, pH:7,47, pCO2: 35,6 mmHg, HCO3: 25,4 mmol/L BE:-2,1 lactate:1,32 mmol/L. The patient was extubated and followed with high flow oxygene. On the 9th day of follow-up, the general condition improved and vitally stable patient was transferred to the neurosurgery service.

Conclusion: Treatment of hemorrhagic shock and secondary acide base disorder is to remove the cause and provide adequate tissue perfusion. For this reason clinicians must use the appropriate fluid therapy and inotropic vasopressor agents to obtain satisfactory results.
THREE DIFFERENT ICD INDICATIONS IN CHILDHOOD IN OUR INSTITUTE

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**Introduction:** An implantable cardioverter-defibrillator (ICD) is a device that can be implantable under the skin, able to perform cardioversion, defibrillation, and also pacing of the heart. The ICD is the first-line treatment and prophylactic therapy for patients at risk for sudden cardiac death due to ventricular fibrillation (VF) and ventricular tachycardia (VT). Recommendations for secondary prevention in children are similar to ICD implantation guidelines developed for adults, despite a paucity of pediatric randomized controlled trials. Several retrospective studies demonstrate efficacy of ICD therapy in young patients. In these cases we reported 3 different ICD indications in childhood, which are implanted in our institute.

**Case reports:** Case 1: A 9-year-old male patient who had Hypertrophic cardiomyopathy (HCMP) diagnosis was admitted to the pediatric intensive care unit (PICU) after resuscitation because of cardiac arrest. The emergency team stated that the patient was cardiac arrested when they found and the monitorization showed asystoly. In PICU, left QRS axis, significant R wave in V1, deep S wave in V6, T negativity in V1 and V2 were observed. The patient was evaluated as HCMP-associated VT-VF after 5 days of follow-up, discharged with beta blocker, and implanted ICD in elective conditions. Case 2: A 14-years old male patient admitted hospital because of seizures. In PICU, VT was noticed at the time of seizure. 24 hour holter rhythm monitorization revealed recurrent catecholaminergic polymorphic VT. Anti-arrhythmic therapy was started with propranolol. A laboratory sample was sent for etiology-directed genetic factors and a new mutation was detected by the genetic laboratory. Case 3: A 10-years old female patient admitted hospital because of recurrent syncopes. In PICU, QTc interval was calculated 0.60 msn. With 24 hours ECG holter monitorization, sustained VT attacks were detected. So that we gave propranolol therapy and implanted ICD.

**Conclusion:** ICD implantation is an effective management for malignant arrhythmias in selected pediatric patients. Patients who have survived sudden cardiac arrest due to malignant arrhythmias would benefit from ICD placement. The indications for ICD implantation in pediatric patients and those with congenital heart disease have developed in recent years with adult randomized clinical trials. Similar to adults, ICD indications have evolved from the secondary prevention of sudden cardiac death. In these 3 cases, all of them have sudden cardiac death risk and have class Ila indications for ICD implantation.

A NEWBORN CASE DIAGNOSED WITH TUBEROUS SCLEROSIS

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**Introduction:** Congenital cardiac tumors are quite rare and the most common is rhabdomyoma (RM). Its prevalence is nearly 0.027-0.08% in autopsies. Cardiac RMs are manifested with heart murmur, cardiac failure and arrhythmia in newborns. RM is reported to be the most common finding of cardiac tuberous sclerosis (TS) with a ratio of 50-80%. TS diagnosed newborn case with central nervous system, skin, cardiac and renal involvement was presented because it is a rare case.

**Case report:** Male patient born in the 40th pregnancy week from a 26 years old mother through G2/P2 caesarean with a birth weight of 3500 gr in our faculty was hospitalized in newborn service due to respiratory problem. Hypopigmented lesions were observed in his examination; five on the back and one on each leg. When the familial medical history was questioned, it was learned that his 31 years old father had multiple sebaceous adenomas in his face. It was considered that rhabdomyoma could be present according to the echocardiography since a hyperechogenic appearance of 10.9x7.8mm on mitral valve, 7x7mm leading from the mitral valve to right ventricular exit way, 5.7x6.8mm inside right ventricle was observed (Figure 1b). Mobitz type 2 block developed in the patient followed-up with ECG monitorization. In the examinations made for multi-organ involvement, minimal floor rhythm irregularity in EEG and calcified subependymal hamartoma, calcified tuber and astrocytomal appearance were observed. Renal cortical-subcortical located regular contoured simple cystic formation were observed in abdominal ultrasonography. Eye examination was normal. Hypopigmented lesions, cortical dysplasia, rhabdomyomas, subependymal nodules were detected as major findings and renal cysts as minor findings and TS was diagnosed. The patient was discharged due to a stable general condition and polyclinic follow-ups were continued.

**Conclusion:** TS is a rare disease in newborns and occurs with multiple organ involvement. Patients with TS in familial story should be evaluated for intracardiac mass and rhabdomyoma should be considered first when a mass is detected. It shouldn’t be forgotten that it may cause narrowness in cardiac mechanics and arrhythmias and thus severe and life-threatening results.
ATRIOVENTRICULAR (AV) COMPLETE BLOCK: A RARE CLINICAL PRESENTATION OF ACUTE RHEUMATIC FEVER

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**Introduction:** In electrocardiography (ECG), prolongation at the PR distance is a minor criteria of diagnostic criterias of rheumatic fever. Complete atrioventricular (AV) block, which is seriously affected by the transmission pathways, is only presented as case reports in the literature. We report three ARA cases that developed AV block in this article.

**Case reports:**

Case 1: A 15-year-old male patient was admitted to the hospital emergency department due to syncope. The ECG was compatible with complete atrioventricular block with ventricular rate 33 / min. 10 mitral valve insufficiency and minimal aortic valve insufficiency were detected by ECO. Transient pacemaker therapy was performed because of syncope caused by a complete AV block. On the third day of antiinflammatory treatment, transient pacemaker treatment was terminated upon complete AV block recovery. Case 2: A 13-year-old girl was referred from another center on the right knee pain and right shoulder pain starting 5 days ago, and on the physical examination, murmur and irregular hearing of the heart sounds. It was seen that the complete AV block in the ECG at the time of admission. First degree mitral regurgitation was detected in ECO. ARA was evaluated as carditis, steroid therapy was started. On the 3rd day of the follow-up of the patient, the rhythm improved. Case 3: A 17 years old male patient was followed up with ARA carditis for 8 years and admitted to the hospital with mimic loss in the right half. He was hospitalized with a diagnosis of facial paralysis. Second degree AV block was observed on ECG. There was an increase in mitral and aortic regurgitation in ECO. Steroid therapy started. On the 3rd day it was seen to return to normalcy.

**Conclusion:** Complete AV block and ARA association is very rare. Of the 21 cases with complete AV block reported in the literature, 19 are in the pediatric age group and 7 of them have syncope. Five of these seven patients underwent transient pacing therapy. All of the reported cases of AV blocks recovered within 8 days at the latest with antiinflammatory treatment. As in our patient (Case 1), complete recovery occurred on the third day in accordance with the literature. Permanent cardiac pacemaker therapy has not been performed because complete AV block recovery is predicted with anti-inflammatory therapy. In cases with ARA, the most common AV block is first degree and seen as a disturbance of transmission but a complete AV block is seen very rarely and in these cases just antinflammatory treatment is sufficient. However, in cases with symptomatic complete AV block, transient transvenous pacemaker should be kept in mind until the block is corrected.

PERIANAL ABSCESSSES ON PEDIATRIC PATIENTS REPORTING ACUTE GAIT DISTURBANCE

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**Introduction:** Perianal abscess is an infectious condition which is characterized by a form of mass located in the anal area that is accompanied by aches and fever. This condition is rare in infants and children with it having less of a chance of being seen spontaneously. It can be caused by the inflammatory bowel disease, diabetes mellitus, immune deficiency, tuberculosis, radiotherapy and trauma in patients.

**Case report:** A 2,5 year old female infant was reported to the external center 2 days ago with complaints regarding gait disturbance and imbalance. After spotting the increasing amount of acute phase reactants (APR) that were inspected in her report, the infant has been redirected to our emergency clinic. There were signs of discomfort, limping and ataxia during the examination that was done on the infant at her arrival. However, the neurologic, jointal and other systemic examinations were classified as normal. In the blood tests that were done, an extraordinarily high amount of APR was spotted. The direct graphs and limbic ultrasonographies were reported normal, as well. However, the patient developed the opistotonus position and an increase in gait disturbance. The magnetic resonance imaging that was taken reported “a 75x37x44 mm sized placeholder apparition in the belly button area that is primarily compatible with the abcess, extending to the right gluteal area”. The patient was taken to abscess drainage. Empirically, cephexon and clindamycin treatment has been started. There has also been E.coli reproduction in the pill cultures that were taken. The immunologic evaluation of the patient showed no anomalies. The decrease of APR has also been spotted in the controlled blood tests that were checked. The treatment has worked on the patient.

**Conclusion:** Perianal abscess separative diagnostics should be taken into account, although with a low possibility, when evaluating patients, especially younger ones, that report with instantaneous gait disturbances.
**A RARE CAUSE OF JOINT PAIN: SYNOVIAL HAEMANGIOMA**

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**Introduction:** Synovial haemangioma is a rare type of tumor which affects the knee joint. Patients who are usually female and their second decade of life, often apply nonspecific symptoms like pain and limitation of motion. When this uncommon tumor is untreated, it can cause cartilage erosion and degenerative joint disease. Because of its rarity, physician awareness is low that results in delayed diagnosis. We presented a synovial haemangioma of the knee to increase awareness of pediatricians.

**Case report:** A 17-year-old girl was admitted with complaints of pain, swelling and tenderness in her left knee since she was 3 years old. With the previously magnetic resonance imaging of the knee joint, she was diagnosed as chronic arthritis on the knee joint. Non-steroidal anti-inflammatory drugs were recommended. Patient applied to our hospital because of ongoing complaints. There was no any arthritis finding including tenderness, pain on motion, deformity, and limitation of motion. On the upper part of left knee, she had localized swelling measuring 3,5x4 cm in diameter. Magnetic resonance imaging (MRI) showed synovial haemangioma characterized by a well-circumscribed contour with a lobule filling the left suprapatellar bursa and space-filling formation with heterogeneous intense contrast enhancement after contrast agent administration. It was required open total synovectomy and mass resection, and histopathological findings were compatible with cavernous haemangioma.

**Conclusion:** Tumor and tumor like lesions such as synovial haemangiomas should be kept in mind in patients with arthritis- compatible findings in single joint. In case of doubt, especially intra-articular lesions, imaging methods should be used for early diagnosis and prevent long term sequel.

**MULTIPLE HIT HYPOTHESIS: A CASE WITH HERBAL MEDICINE INDUCED ATYPICAL HUS**

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**Introduction:** The thrombotic microangiopathies (TMA) consist of a heterogeneous group of diseases which have common clinical manifestations including nonimmune hemolytic anemia, thrombocytopenia, and acute kidney injury. The noninfectious HUS is rare in children. Atypical hemolytic uremic syndrome (aHUS) is the term commonly used for this set of diseases. There are many factors that were reported associations with the aHUS like non-Shiga toxin infectious agents, drugs, cancers, vaccination, transplant, autoimmune disease etc. These data for whether these agents are the primary cause of disease or triggers of an alternative complement system is limited. According to the “multiple-hit” hypothesis, aHUS is a consequence of both genetic predisposition to alternative complement dysregulation as well as the occurrence of events or conditions that may precipitate TMA by activating complement and/or damaging endothelium. We discussed a patient who was diagnosed with atypical hemolytic uremic syndrome possibly triggered by over the counter medication, herbs.

**Case report:** 11 years old boy, whose brother was dead because of renal insufficiency possibly secondary to TMA, admitted to hospital with flank pain. A renal biopsy was done because of the presence of proteinuria and high blood pressure. The complement levels and hematologic parameter were normal but haptoglobin was low. Biopsy was remarkable for hypertensive nephrosclerosis. He had been following up at the outpatient clinic with ACE inhibitor with normal renal function test and abnormal proteinuria for 5 years until he admitted to hospital with acute renal failure. It was learned that he had been using many herbal drugs for 3 months. On admission, his blood pressure was high and kidney was not working well. In the follow up, his platelet and hemoglobin levels were decreased and LDH levels were increased. With his family history, he was diagnosed as an aHUS which was trigger by herbs. The patient who is currently on eculizumab needs hemodialysis three times a week.

**Conclusion:** It is known that atypical hemolytic uremic syndrome results from a defect on the alternative complement system. We think that herbal drugs acting like antigen may trigger alternative pathway of complement especially in patients who have defects in their complement pathway.
GIANT HYDRONEPHROSIS DUE TO URETEROPELVIC JUNCTION OBSTRUCTION: A CASE REPORT

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Introduction: Ureteropelvic junction obstruction (UPJO) is the most common cause of prenatal hydronephrosis and has an incidence of 1 in 1,000-1,500 newborns. Hydronephrosis caused by UPJ does not always progress, but may increase rapidly and without warning in adult life. Giant hydronephrosis is defined as a hydronephrotic kidney containing more than 1 lt of fluid and the cause is usually due to a delay in diagnosis and treatment. We will present a 16 years old girl who was admitted with complain of flank pain and diagnosed as giant hydronephrosis required surgery.

Case report: A 16-year-old girl, who was attended to another center with flank pain, was admitted our hospital with seriously increased right kidney size measuring 220 mm in diameter with thinning of the renal parenchyma and nephrolithiasis. Computerized abdominal tomography was remarkable for UPJO with a lesion in a multicystic structure extending to the pelvis grim in the size of 25x13x15 cm. Her biochemistry was normal in terms of kidney functions. Markedly decreased renal uptake in right kidney (% 20) was detected by Tc-99m. Pyeloplasty was done. Then, her flank pain was gradually decreased.

Conclusion: Giant hydronephrosis may be detected at an advanced age with nonspecific complaints such as wasting distention and flank pain. In general, nephrectomy is performed if there is non-functioning kidney. Our patient underwent pyeloplasty because of having functioning kidney. Giant hydronephrosis should be kept in mind in children having chronic flank pain even in teenagers.

PYOURETER RELATED TO VCUG IN THE DIFFERENTIAL DIAGNOSIS OF UROSEPSIS

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Introduction: Voiding cystourethrography (VCUG), is a diagnostic tool that is frequently used in pediatric urology. However, urinary tract infection can occur and after the procedure. Pyoureter is a very rare urinary tract infection, in which ureter is filled with pus and in some cases, it can result in fatal urosepsis and obstruction. In this study, we are presenting a pediatric patient that is admitted with urosepsis after VCUG and pyoureter that we operated surgically.

Case Report: 2-year-old male patient, who has been followed-up for VUR in the right kidney, had a VCUG with prophylaxis for control. 5th day after the procedure, the patient admitted with unrest while urinating, vomiting, and high fever. Pain in the suprapubic area and high fever (39 °C) was detected on the physical exam. After laboratory results showed an increase in pyuria and inflammation markers (WBC 14960 / mm3, CRP 175 mg/l), antibiotics are initiated. Patient’s fever didn’t go down, urinary USG showed that right ureter was extremely dilated (5cm) and echogenicities on all calyces of right kidney, pelvis, and ureter. After admitting double j-stent for treatment purposes, patients fever and acute phase reactants were normalized within 48 hours. The catheter was removed, the patient has normal kidney function and follow-up is been continued as an outpatient.

Conclusion: Pyoureter is a very rare complication that develops after manipulations to the urinary system. Even though appropriate prophylaxis used before the procedure, this complication can occur. This shows that complying sterilization procedures are very important.
**OUR PEDIATRIC TULAREMIA CASES**

**P37**

**Introduction:** Tularemia is an infectious zoonic disease caused by Francisella tularensis, a gram-negative, nonmotile, aerob cocccobacillus. The disease can be transmitted to humans through direct contact of skin and mucosal surfaces with the tissues or body fluid of infected animals, arthropod (acarid, phthirius and fly) bite, consumption of contaminated water or food, or breathing infected aerosols. Consumption of water and food contaminated by infected with animal tissue is a major mode of transmission in epidemic of tularemia, particularly in our country. No human-to-human transmission has been reported. Tularemia is classified in 6 six clinic forms according to the entry of the factor into the host, i.e. ulceroglandular, glandular, oculoglandular, oropharyngeal, pneumonic and typhoidal. All clinic forms are characterized with sudden fever, shivering, headache, fatigue, myalgia and arthralgia. Skin rash is the most common complication in tularemia, with being seen in 3-25% of the patients. Erythma nodosum and erythma multiform are skin manifestations that may be seen in the process of the disease. In this paper, 4 pediatric cases with tularemia who were followed in our clinic in the recent year are presented with the intention to raise awareness to childhood tularemia.

**Case reports:**

**Case 1:** A 6 years old female patient admitted with complaints of erythema and swelling on front site of leg. When physically examined, lesions congruent with erythema nodosum were present on the lower extremity. Laboratory findings were as follows; WBC: 19.7 (10³ u/lt), CRP: 32.9 (mg/l), ESR 56 (mm/h). Gentamicin treatment was initiated. Case 2: A 13 years old female patient admitted with complaints of swelling, pain and tenderness in left cervical region, fatigue and tiredness. When physically examined, a mobile, 5x4.5 cm sensitive to contact, LAP in firm consistency in left frontal cervical region, anterior to the sternocleidomastoid muscle was present. Laboratory findings were as following: WBC: 10.4 (10³/u/lt), CRP: 5.1 (mg/l), ESR 29 (mm/h). Treatment with gentamicin and surgical drainage was initiated.

**Case 3:** A 4 years old female patient admitted with complaints of swelling posterior to left ear and on left neck site. When physically examined, a nonmotile, painful 4x4 cm LAP in left cervical region and a 1x0.5 cm LAP in left axillary region were present. Laboratory findings were as following: WBC: 14.9 (10³/u/lt), CRP: 9.3 (mg/l), ESR 32 (mm/h). Treatment with Streptomycin and surgical drainage was initiated.

**Conclusion:** Tularemia continues to be a common public health issue in our region. All presented studies were treated with several antibiotics but their complaints did not regress. Therefore, if unresponsiveness to antibiotics used against common lesions of lymphadenopathy or erythema nodosum in childhood is present at first step, tularemia should be considered in the etiology definitively.

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**A RARE CAUSE IN A PATIENT PRESENTING WITH VISUAL LOSS; HODGKIN LYMPHOMA**

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**Introduction:** In children with visual loss, one of the reasons is the inflammation of the uvea, which is a rich middle layer of veins and pigment of the eyebrow. Uveitis is seen less frequently in childhood than adults, but worse in visual prognosis. There is a very wide spectrum of differential diagnosis. Patients should be evaluated sarcoidosis, infections (tuberculosis, viral infections, hematologic diseases and malignancies. Here we present Hodgkin lymphoma associated with uveitis in a patient who presented with loss of vision.

**Case report:** A 9-years old male patient was presented with pain, redness and vision loss in both eyes to the external center and diagnosed with panuveitis and papillary edema, sent to our center for further examination and treatment. The patient has no known systemic disease. There is no similar health problem before that. The patient was consulted with Pediatric Pulmonology for sarcoidosis and tuberculosis, pediatric rheumatology department for the evaluation of rheumatological diseases, and pediatric neurology department for papillary edema. The patient was examined for sarcoidosis and tuberculosis, which may cause panuveitis. WBC was 4130/mm³, hemoglobin: 11.3 g/dL, platelet 247.000/mm³, urine Ca/Creatin 0.019, serum Ca:9.56 md/dl P:4.45 md/dl sedimentation:57mm/h, CRP:46.6mg/L. PPD was anergic. Bilateral hilar and mediastinal mass were found in chest X-Ray. Since these findings couldnt be distinguished from malignancy, firstly bone marrow aspiration sampling was performed. Atypical and malignant cells were not seen in bone marrow aspiration, and eosinophil was found to be increased. Lymph node biopsy was performed for the diagnosis of sarcoidosis in the patient and for tissue sampling to exclude malignancy. Pathology report was evaluated as mixed cell type Classical Hodgkin lymphoma.

**Conclusion:** Uveitis is a rare disease in children than adults. According to the etiological classification, two groups are separated as infectious and non-infectious. The correct distinction between non-infectious types is of vital importance in planning adequate treatment. In this case report, we emphasized that Hodgkin lymphoma, which may be rare in the differential diagnosis of uveitis, should be remembered.
### MYOCARDIAL BRIDGE APPEARING IN CHILDREN WITH CHEST PAIN

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<th>P39</th>
<th>Introduction: Chest pain in children is a frequent complaint. Causes of chest pain in children are: idiopathic (most common), musculoskeletal, respiratory, gastrointestinal, cardiovascular, nervous system diseases. The most feared reason of chest pain is cardiac pathologies although it is seen %5 of the time. Myocardial Bridge is a rare cardiac condition which causes chest pain. Coronary arteries and large branches usually travel on the surface of the heart. Myocardial bridge which is part of the coronary arteries are embedded in the myocardium. The myocardial bridge is commonly seen in the middle segment of the left anterior descending artery (LAD), rarely in the right coroner (RCA) and circumflex (Cx) arteries. The incidence increases in men. It is usually a benign disease with a good long-term survival. However, myocardial ischemia, infarction, tachycardia can be the cause of death.</th>
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<td>Case report: A 13-years-old male patient who had complaints of intermittent chest pain for 1 week had chest pain in a detectable manner. There was no pain spreading to the neck, arm and shoulder. When the patient got tired he had chest pain and the pain would increase when he laid on his left side. The patients' mother has coronary artery disease. Consanguinity was present between parents. Chest pain is present on the patient's physical examination. In the laboratory tests performed, CK-MB was 42.3 ng / ml, Troponin I was 26738 pg / ml, Pro-BNP was 132 pg / ml and AST was 77 U / l. ECG: Sinus rhythm speed was 66 / min. No pathological findings were found. Echocardiography detected: valve structures and functions are normal, there is no wall motion disorder. Myocardial bridge was detected in long segment of LAD in coronary angiography and RCA, Cx were normal. The patient was started on beta blocker therapy and the operation was recommended.</td>
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<td>Conclusion: Myocardial Bridge is a rare cause of heart disease in chest pain. The myocardial bridge is a part of the coronary arteries is embedded in the myocardium. The myocardial bridge is commonly seen in the medial segment of LAD. Coronary angiography was performed because of the fact that the patient was a child and a family history of coronary artery disease. Myocardial bridge was diagnosed due to the milking effect of systolic narrowing on the coronary artery. Chest pain should be remembered as a rare cause of myocardial bridging.</td>
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### THREE RARE MYOKYMIA CASES IN CHILDHOOD

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<th>P40</th>
<th>Introduction: Myokymia is a spontaneous recurrent muscle movement that causes a fluctuating or wavy condition. It is most commonly seen in focal form and in the face area.</th>
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<td>Case report: Case 1: A 14-year-old girl presented with twitching under the right eye for 3 months. While their complaints disappeared during sleep, there were fasciculations that occurred 2-3 times a second in the wake. Cranial imaging and EEG were normal, with no drug use history and no known disease. The levels of Vitamin D and Vitamin B12 in the case are low. Complaints of the patient who received Vitamin B 12 and Vitamin D treatment disappeared after 20 days. Case 2: A nine-year-old girl presented with a complaint of throwing in the neck region lost in sleep, repetitive every second during the last 20 days. There was no known disease or drug use in his medical history. Complete blood count, biochemistry, acute phase reactors, thyroid hormones and Cranial MRI were evaluated as normal. Neck USG was performed, it was normal except for a few millimetric lymphadenopathy that were reactive. A decrease in the complaints of the patient who came to the control one month after the upper respiratory tract infection treatment was seen. Case 3: A 10-year-old girl with asthma was admitted to our emergency pediatric clinic with asthma. Fasciculations were seen in the left jugular region 30 minutes after ventolin nebulus was delivered to the patient. It was learned that the same complaints were seen 2 months ago, after ventolin nebulus using. The laboratory and radiological examinations performed were normal.</td>
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<td>Conclusion: Myokymia may be associated with fatigue, anxiety, stress, exercise and excessive caffeine use, as well as secondary etiologic agents such as genetic, immunological, autoimmune, radiation, vascular, demyelination. All three cases were presented because of their relatively uncommon appearance.</td>
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HEREDITARY HYPERFERRITINEMIA AND CATARACT SYNDROME: A CASE REPORT
Ulviye Esra Bayat Güler, Alper Özcan, Yunus Güler, Ekrem Ünal, Hüseyin Per
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**Introduction:** The causes of hyperferritinemia in children include infections, chronic inflammatory diseases, autoimmune diseases, hematological diseases and liver diseases. High ferritin levels can cause genetic diseases. One of the rare genetic causes of serum ferritin elevation is hereditary hyperferritinemia and cataract syndrome (HHCS). HHCS is an autosomal dominant disorder. Associated symptoms of HHCS are high serum ferritin levels in the absence of iron overload accompanied by early onset of bilateral cataracts. It may give no findings other than visual impairment. Point mutations in the iron-responsive element (IRE) in the 5' UTR region of the L-ferritin gene (FTL) result in increased expression of the L-ferritin protein resulting in hyperferritinemia. In this report, we present a 10-year-old girl with HHCS who was admitted to the general polyclinic of children's hospital with ferritin height.

**Case report:** A 10-year-old girl was referred to my hospital when she had serum ferritin > 2000 ng/ml, even though there was mild anemia in the hemogram seen at the health center with pain, anorexia and weakness. There was no pathological examination on physical examination. Hemoglobin: 11.9 mg / dl, MCV: 79.5 fl and RDW: 13.7% transferrin saturation: 9%, iron, iron binding, vitamin B12 normal. The level of ferritin (1630 ng / ml) was high in the case of peripheral dissemination. Renal and liver function, GGT, bilirubin, LDH, triglyceride levels were normal. In the case of family medicine, mother and monument were cataracts, the case department was cared for. Bilateral cortical and nuclear concentrations (<3mm) were consulted for hematology. Molecular genetic analysis was sent in cases where hyperferritinemia was detected in individuals with cataracts in the family.

**Conclusion:** HHCS was first reported in 1995 by Girelli et al. Family members affected in three generations have bilateral cataracts, hyperferritinemia and an autosomal dominant inheritance pattern. Although the prevalence is not clearly known, it has been found that there is about 1 / 200,000 of prevalence in a study in Australia. The effects of HHCS in other tissues are unknown at this time. As a result, HHCS patients are subjected to numerous stressful and recurrent medical examinations before being diagnosed. Patients with unexplained hyperferritis should be consulted to ophthalmologist for cataract screening.

EVALUATION OF HEART RATE VARIABILITY WITH HOLTER MONITORIZATION IN VSD CHILDREN CASES WHO WERE REPAIRED WITH TRANSCATHATER METHOD
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**Introduction:** Heart rate variability (HRV) is a well-known sign of cardiac autonomic nervous system and it is based on variability of RR distances on electrocardiogram. Evaluation of HRV in pediatric VSD cases before and after transcatheter repair will enlighten us in term of cardiac autonomic control.

**Material and method:** 25 VSD cases who were treated with transcatheter closure method and 13 control healthy children were enrolled in the study. 24hr Holter rhythm monitoring was applied to all children. HRV was calculated before transcatheter closure, after 1 day, 1, 3 and 6 th months with Holter rhythm monitorization.

**Result:** 15 patients whose VSD closed percutaneously and 13 healthy children were enrolled in study. There were no significant difference between 2 groups in terms of HRV parameters. In 3 months period, any difference was detected for HRV parameters in the patient group. HRV parameters were similar in daylight holter monitorization.

**Conclusion:** In our study when transcatheter repaired VSD cases and control group were compared, there were no significant difference between them. This finding may be result of small number of cases or short follow-up period. Another reasons of similarity may be normal pulmonary arterial pressure and absence of congestive heart failure. Statistically significant results can be obtained when the study is completed with more patients with 6-month follow-up. In the literature, there is no another study that evaluates HRV in VSD pediatric cases closed by transcatheter method. We think that our study will contribute to the literature when the number of cases increases and long-term follow-up (6 months) is completed.
THE FACTORS AFFECT ORGAN AND RENAL INVOLVEMENT IN PEDIATRIC HENOCH SCHONLEIN PATIENTS
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Introduction: IgA vasculitis is the most common vasculitis seen in childhood. Though kidney involvement is a major factor affecting prognosis, every organ in body might be affected. The aim of this study is to determine the organ involvement and clinics, to identify the risk factors that increase the nephritis and to determine the long-term prognosis of patients with IgA vasculitis.

Materials and Methods: 415 IgA vasculitis patients who were followed in 1990 and 2016 in Erciyes University Pediatric Nephrology and Rheumatology Department included the study. Patients' files were scanned retrospectively. The patients' presenting symptoms, organ involvement, treatment and long-term prognosis were determined.

Results: In our study, 415 patients diagnosed with IgA vasculitis (HSP) were evaluated. Of the patients, 173 (41.7%) were female and 242 (58.3%) were male. The mean reference age was 8.3 ± 3.1 years. Skin involvement was found 100%, joint involvement was 77.1%, GIS involvement was 58.6%, renal involvement was 38.3%, scalp edema was 10.1%, scrotal involvement was 5.3% and central nervous system involvement was 1.6%. When risk factors for nephritis were assessed, it was found that gastrointestinal involvement was more frequent in those with nephritis (p=0.01). Age, diastolic blood pressure and GIS involvement were correlated with renal involvement, and GIS involvement and diastolic blood pressure are found to be risk factors for renal involvement.

Conclusion: Age at diagnosis, elevated blood pressure and GIS involvement are important findings in predicting the development of nephritis in patients. Patients with GIS involvement should be carefully monitored for kidney involvement.

RESPIRATORY DISTRESS IS SECONDARY OF BRANCHOCONSTRICTION IS RELATED WITH HYPOCHALCEMICA IN NEWBORN
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Introduction: Regulation of calcium is controlled by many factor in newborn. Newborn’s vitamin D levels are closely associated with his mother’s vitamin D levels.

Case Report: Patient was burned at 34 weeks and 2650 gram and his mother’s age was 21. Patient admitted to newborn service because of respiratory distress and complaint of unease and groaning. Postero-anterior lungs x-ray was normal. The patient’s vascular access was established and parantheral fluid was regulated. Empiric treatment was started. CPAP (Continue Positive Airway Pressure) was performed with nasal mask because patient’s complaints has increased. Although serum calcium levels was normal initially, it was 7.21 mg/dl at 24h hours and 5.8 mg/dl at 48 hours after birth. Vitamin D and PTH was studied for patient and his mother. PTH level was in normal range and Vitamin D level has decreased. Ca Laktat was started as 50 mg/kg/per day and Vitamin D was also started as 1*1200 ü/day. Because of magnesium deficiency magnesium replacement was started. Calcium levels has increased gradually and finally it has been in normal range after five days. And suddenly patient has gotten breath back and his necessary for CPAP with nasal mask vanished without atrace. He has got seizures, consequently TF USG, cranial MR and EEG was made but there is no patological material. Anticonvulsan treatment wasn’t started. If it would be necessary for clinical signs, EEG at sleep-awake for the cerebral maturation was suggested. PA Luns x-ray was normal, blood culture was sterile at conrolled. So that it is considered, patient’s respiratory distress is secondary of bronchoconstriiction is related with hypochalcemica. And when it has come to normal interval, respiratory distress is healed without suquel.

Conclusion: Hypochalcemica is common electrolyte imbalance in newborn can respiratory distress is secondary of bronchospsasm. And this situation should not be forgotten.
SEPSIS AND COMPLETE BLOOD COUNT: THERE'S STILL MUCH TO SAY

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**P45**

**Introduction:** Despite advances in vaccination and modern antibiotics, sepsis remains one of the leading causes of admission to pediatric emergency and intensive care units in the world and in our country. Procalcitonin (PRC) and C-Reactive Protein (CRP) are successful in showing bacterial infections; The most important limitations are high costs and laboratory results take a long time. Complete blood count (CBC) is a commonly used test because of its low cost, ease of use, and its ability to provide valuable information in many diseases. In this study, we aimed to evaluate the association between CRP and PRC with the parameters of CBC, NLR, and MPV, in the diagnosis of sepsis.

**Materials and Methods:** We retrospectively reviewed the medical records of patients who had been followed up in our department between January 2015 and January 2017. Hematologyoncology patients were excluded. PRC level of 0.5 ng/mL was defined as sepsis.

**Results:** A total of 125 patients enrolled into the study. PRC with CRP (p < 0.01; rho = 0.279), NLR (p = 0.02, rho = 0.186) and MPV (p < 0.01; rho = 0.243) were found to have a weakly statistically significant positive correlation. The NLR (1.8) produced a sensitivity of 78%, and specificity of 43%. At a cut-off value of 8.5 fl, the sensitivity of the MPV was 42%, and specificity of 80%. At a cut-off value of 10 mg/L, CRP level yielded a sensitivity of 79%, and a specificity of 44%.

**Conclusion:** To our knowledge, our study is the first to investigate NLR and MPV as an early diagnostic marker for sepsis in pediatric patients.

ALICE IN WONDERLAND SYNDROME: CASE REPORT

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**P46**

**Introduction:** Some episodic syndromes have been identified as migraine precursors. In these cases, the sensory impairment (Syndrome of Alice in Wonderland) is an episodic syndrome characterized by a smaller (micropsia) or greater (macropsia) perception disorder of the patient's own body or surroundings. This phenomenon is not only associated with migraine, but also with infection. We presented this case because of the infection-related Alice in Wonderland syndrome is very rare.

**Case Report:** A 7-year-old male patient referred with nausea, vomiting, and fever that started 1 week before. In the last week, especially when he has fever and just awake, he saw objects and his body as bigger. 3 years ago, there was similar complaints in the period when he had fever and after the illness symptoms was lost. On physical examination, there was a case of membranose tonsillitis. His neurological examination was normal. In blood count, number of white blood cell was normal but lymphocyte was low. EBV IgM, CMV IgM, and HSV IgM were negative. The EEG was normal. The Alice in Wonderland Syndrome was considered. The patient was hospitalized for two days because of the fever and followed up with IV fluid. The patient was discharged when the fever fell and the complaints did not recur.

**Conclusion:** Alice in Wonderland syndrome was considered because of clinical and laboratory findings were compatible with viral infection. This phenomenon may also be associated with major depressions, seizures, medications (such as topiramate), and right medial temporal lobe vascular lesions. In our case EEG and Magnetic Resonance imaging showed no pathological findings. Alice in Wonderland Syndrome is a rare clinical picture. Although it is known as a migraine-related syndrome, it should be considered in relation to EBV infection, temporal lobe epilepsy, and treatment should be given for etiological factors.

SPASMUS NUTANS: CASE REPORT

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**P47**

**Introduction:** Spasmus nutans is a rare disease characterized by head nodding, torticollis, and rapid, asymmetric, low amplitude nystagmus triad, usually seen in 4 to 14 months of age. Nystagmus can persist until the age of 12. It's a self-limiting benign disease.

**Case Report:** A 6-month-old male patient was referred for swinging in his eyes lasting 2-3 days. On physical examination, horizontal nystagmus, head nodding and torticollis were observed. His neurological examination was normal. In this case spasmus nutans were considered and eye examination was normal. Cranial magnetic resonance imaging was normal. In the videos of the case, nystagmus, head nodding and torticollis were detected and spasmus nutans were diagnosed.

**Discussion:** Spasmus nutans is a rare benign disease characterized by head nodding, torticollis, and rapid, asymmetric, low-amplitude nystagmus. The diagnosis is based on these three symptoms and distinguish the other nystagmus causes. The age of onset varies from 4 to 14 months. Before Spasmus nutans are diagnosed, other possible diagnoses must be excluded. Optic nerve glioma and congenital nystagmus are important in differential diagnosis. Normal ophthalmologic and neurological examination findings and normal brain MR are necessary for diagnosis. Our diagnose based on having three characteristic symptoms and eliminating other possible causes of nystagmus.

Spasmus nutans is a benign disease that disappears spontaneously without treatment within 1-4 years, but nystagmus can persist to 12 years. Although the pathophysiology is unknown, it is thought that the head nodding is the result of the normal compensatory oculo-vestibular reflex.

**Conclusion:** Spasmus nutans are a rare benign disease. Diagnosis made by clinical observations, but differential diagnosis is important.
WHAT A PROBLEM OF DERMATITIS
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Introduction: Langerhans cell histiocytosis is a rare disease that affects infants and children and characterized by localized or generalized proliferation of Langerhans type cells. Our case is in a patient who has skin rash which started on the 4th month and after noticing cephalic swelling, the patient has been diagnosed with an x-ray which has lesions that are concordant with lytic lesions.

Case report: Four months old patient admitted to another center with Upper respiratory tract infection and Rash, and after symptoms got better, the patient was discharged. At the routine outpatient exam of the patient at the 8th month, the cephal swelling was detected, a head x-ray was taken and lytic lesions were seen. The patient was dispatched to our clinic for further examination and treatment. The initial dermatologic exam showed erythematous, sporadic eroded patches around suprapubic, inguinal, sacral and perianal area; there were yellow squam papules on the erythematous skin. There were no peripheral LAP or hepatosplenomegaly in physical examination. There were lesions in both parietomastoid zones, the right zone had 1.5x1 cm and left zone had 2x2 cm, left frontoparietal zone had 6x6 mm lesions. These lesions were round shaped lumen images and concordant with lytic lesions in head x-ray. Patient admitted to pediatrics department with pre-diagnosis of Langerhans cell histiocytosis. Skin biopsy showed were CD163 and CD68 and histiocytes were positive for CD1A and S100 immunohistochemical staining. Abdominal USG and PA-lung x-ray were normal. Bone marrow aspiration made by department consultant, and cells that were twice the size of lymphocyte, blue cytoplasm, loose chromatin structure, and blep like dentations was seen. Langerhans cell histiocytosis was concurrent for bone marrow involvement.

Conclusion: Definitive diagnosis is made by showing the presence of Langerhans cell or by showing CD1a surface antigen with immunohistochemical methods. In this poster, we present the case of a 8-month-old female baby who hasn’t get diagnosed for a long time.

A RARE ETIOLOGY FOR MEDIASTINAL NECROTIZING GRANULOMATOUS INFLAMMATION: TULAREMIA
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Introduction: Tularemia is a zoonotic infection caused by the Gram-negative bacterium Francisella tularensis. People living in rural areas, farmers, hunters, forest workers, laboratory staff are at the highest risk group. In this report a case of tularemia, a very rare cause of mediastinal necrotizing granulomatous lymphadenopathy was presented.

Case Report: A 17-year-old male was admitted to another hospital a month ago with fever, weight loss (8 kg in the last month) and fatigue. He had been treated with antibiotic therapy nonspecifically. In the follow-up, since his increased sedimentation rate and fever still persisted despite antibiotic therapy he presented to our clinic for further examination and treatment. In admission, Hb was 9.4 gr/dl, WBC was 16530/mm³, ESR was 120 mm/h and CRP was 34.7 mg/l. Liver function tests, renal function tests and electrolytes were within normal limits. The thoracoabdominal tomography of the patient had numerous lymphadenopathies around the mediastinal, celiac truncus, superior mesenteric artery. Tuberculin skin test was anergic. Malignant cells were not observed in peripheral smear and bone marrow aspiration. Excisional biopsy was performed from the mediastinal lymph nodes by mediastinoscopy for differential diagnosis. Routine cultures of the lymph node material were negative for fungi and acid-fast organisms. Histological examination was compatible with necrotizing granulomatous lymphadenitis. After the current clinical findings of the patient could not be explained by tuberculosis, sarcoidosis and lymphoma, upon further questioning, the patient’s life in the rural areas, hunting and feeding rabbit meat stories, the blood samples were sent to the Refik Saydam Public Health Laboratory for tularemia, listeria, cat scratch and Lyme diseases. Microagglutination test for tularemia was found positive with a titer of 1/1280, the patient was diagnosed with tularemia and ciprofloxacin treatment was initiated. After 28 days of treatment complaints of patient were delayed completely.

Conclusion: In the differential diagnosis of mediastinal lymphadenopathies, a rare etiology of necrotizing granulomatous inflammation, tularemia should be considered together with tuberculosis, sarcoidosis and lymphoma.
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Introduction: Gastric volvulus is a twisting of the stomach by more than 180 degrees and causing obstruction (1). It’s hard to diagnose gastric volvulus (2). Although it doesn’t usually have clinical findings, it causes some patients to have respiratory or gastrointestinal complaints. Here, we present a gastric volvulus case of 3-year-old that was found incidentally.

Case Report: A 3-year-old girl who was scheduled for operation due to strabismus applied to the pediatric chest diseases polyclinic because of cough complaints and preoperative evaluation. The general condition was good, blood pressure 80/50 mmHg, respiratory rate 25/ min, temperature 37.6°C, pulse rate 115/min, oropharynx hyperemic, rare bilateral rales on respiratory examination, other systemic examination was detected naturally. Blood tests revealed white cure 9300/mm3, hemoglobin 14.6 g/dl, platelet 286000/mm3, CRP 6.5 mg/dl, sedimentation 13 mm/h. Bilateral paracardiac infiltrative areas were present on the anterior-posterior chest X-ray. The notable point was that the deterioration of endothelial integrity plays a major role in the development of vascular diseases. Endothelial microparticles (EMP) are considered as markers of endothelial dysfunction. In this study, we aimed to examine whether there is an endothelial dysfunction in patients with Familial Mediterranean Fever (FMF) and the relationship between EMPs and activation of the disease in children with FMF.

Material and method: This cross-sectional study included 65 FMF patients (41 attack free, 24 attack period), and 35 healthy controls. Circulating EMPs, serum amyloid A (SAA) and other inflammation markers were measured. Circulating EMPs were measured by flow cytometry. Study groups were compared for circulating EMP and inflammatory markers. The relationship between EMPs and activation of the disease was evaluated.

Results: The levels of SAA, CD144+ and CD146+ EMP in the FMF attack period group were significantly higher than those in control group (p<0.05). Additionally, the levels of CRP, erythrocyte sedimentation rate and white blood cell count in the attack period group were significantly higher than those in the control and attack free groups (p<0.05). In FMF attack group, the CD144+ and CD146+ EMP were significantly correlated with CRP.

Conclusion: Our results suggest that endothelial damage is present especially in active period of disease in FMF. The endothelial dysfunction becomes overt parallel with inflammation. EMPs may be a new reliable marker of the inflammation in children with FMF.
**A RARE DISEASE: ENCEPHALITIS AFTER EXANTHEM SUBITUM**

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**Introduction:** Exanthem subitum (ES) a benign, inflammatory disease caused by human herpes virus type 6 (HHV-6), which is seen especially in children during the first two years of life. It is characterized by sudden onset of fever which resolves abruptly and is followed by development of a rash. The most common complications of ES are central nervous system (CNS) complications such as febrile seizures and encephalitis. Here, we present a case of encephalitis after ES, who admitted with complaints of eruption, decreased appetite and dizziness.

**Case report:** Antibiotic therapy was started with a diagnosis of upper respiratory tract infection to a 7 months old boy who applied another hospital with the complaints of nose discharge, anorexia, restlessness and fever of 40 °C. Fever had continued for three days. After the fever returned to normal levels, a rash had occurred on chest firstly and spread out to neck, arm, leg, and facial region. The patient had admitted to our emergency department with complaints of rash, decreased appetite, and dizziness. On physical examination the general condition was moderate and tending to sleep. Body temperature was 36.7 °C, heart rate was 120 / min, respiratory rate was 28/min and blood pressure was 105/60 mmHg. The anterior fontanel was pulsatile and bulged. There was maculopapular rash on the whole body. Other system examinations were normal. Cerebrospinal fluid (CSF) sample analysis revealed, protein 23.5 mg/dl, glucose 63 mg/dl, and concurrent blood glucose 96mg/dl. Microscopic examination was normal. Intravenous acyclovir treatment was started. CSF culture analysis revealed negative and HHV-6 viral DNA was detected in CSF by using the DNA isolation + Real Time PCR + multiplex PCR method. The patient was discharged with complete recovery after 21 days of acyclovir treatment.

**Conclusion:** Although CS is a benign disease characterized by fever and rash, careful attention should be paid for the development of central nervous system complications. HHV-6 encephalitis should be considered in differential diagnosis in patients with fever, rash, and general impairment.

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**A RARE CAUSE OF MENINGITIS: LACTOBACILLUS CASE**

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**Introduction:** In recent years, microorganisms with multiple drug resistance predominate in recurrent meningitis. Lactobacillus is a genus of bacteria found in normal flora of the mouth and the intestinal tract which rarely cause infections. Here we report a boy with Lactobacillus casei meningitis, who has cerebral palsy and history of recurrent meningitis.

**Case report:** An 8-years-old boy who was followed-up with the diagnoses of cerebral palsy and hydrocephaly, with ventriculoperitoneal shunt was admitted to our pediatric emergency clinic with the complaints of fever and vomiting. He had had a neuromotor sequela caused by a car accident happened 8 months ventriculoperitoneal shunt had been implanted because of hydrocephaly, subsequently he had experienced recurrent meningitis for 3 times, so that ventriculoperitoneal shunt had been removed a month before, acinetobacter was detected in his last cerebrospinal fluid (CSF) culture, susceptible to ceftazidime and he had been discharged after 14 days of treatment. On his physical examination he was unconscious and he could not localize pain stimulus. Nuchal rigidity was remarkable. Pupillary reflex was bilateral negative and the pupils were isochoric. Respiratory system examinations revealed bilateral crepitant rales. He had gastrostomy. The patient was hospitalized to pediatric intensive care unit with the diagnoses of menengitis and pneumonia. An external shunt drainage catheter was placed. CSF analysis revealed, protein level 3201 mg/dl, glucose level 161 mg/dl and simultaneous blood glucose level was measured as 197 mg/dl, and 14300/mm3 white blood cell was detected on microscopic examination. Vancomycin and cefotaxime treatments were started. Magnetic resonance imaging was concordant with hydrocephaly, ventriculitis and meningitis. Lactobacillus casei was detected in CSF culture. The patient was discharged subsequent to the completion of 14 days antibiotic treatment.

**Conclusion:** It seems likely that rarely pathogenic bacteria will be increasingly dominant because of the reasons including prolonged hospitalization in intensive care units, increased rates of invasive procedures, prolonged use of antibiotics. Further investigations are needed in this topic.
INVASIVE ABDOMINAL ASPERGILLOSIS CASE IN A NEONATAL
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Introduction: Neonatal aspergillosis is the second most common opportunistic fungal infections in immuno-suppressed hosts after candida. Risk factors for neonatal invasive aspergillosis are use of wide-spectrum antibiotic and steroid, premature and necrotizing enterocolitis. The final diagnosis is the positivity of the culture taken from the tissue and histopathological examination.

Case report: In the infant born from a 27 year old mother in an external center in the 26th week of her pregnancy with a birth weight of 1000 grams and followed up with prematurity and respiratory distress syndrome diagnoses, antibiotherapy, positive inotropic treatment and pentoxifylline treatment were started due to sepsis and disseminated intravascular coagulation presentation occurring on the second day of hospitalization and widespread intraparenchymal bleeding, asphyxia and also left shift in median line structures were detected in the brain tomography taken due to generalized convulsions occurring on the third day of the follow-up of the patient who was given septic exchange due to the continuing severe septic presentation despite the treatment. The brain surgery department recommended follow-up. The patient was transferred when seven days old due to heart murmur occurring during follow-up. Umbilical catheter was leaking when the patient arrived. Due to umbilical hernia in appearance and sensitivity in examination and the presence of widespread crusted infected appearance in whole abdominal skin, the umbilical catheter was removed (Figure 1). Systemic antifungal treatment was started considering that widespread abdominal fungal infection was present in the case with systemic antibiotic use and prematurity. Diffused mild edematous appearance in intestinal loops, significant edematous intestinal loop in upper left quadrant and mild free fluid in the abdomen and dirty appearance in mesentery observed in the abdominal ultrasonography were in line with fungus intraabdominal dissemination. Aspergillus reproduction was present in the umbilical catheter culture sent. The patient whose general condition got worse and had a anuric course despite the treatment was exitus on postnatal 14th day.

Conclusion: Despite systemic antifungal treatment in immuno-suppressed and preterm neonatals with lengthened antibiotic usage, invasive abdominal aspergillosis infection is an important clinical entity with high mortality.

CYSTIC ADENOMATOID MALFORMATION CASE PRESENTED WITH RESPIRATORY PROBLEM STARTING AFTER BIRTH
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Introduction: Congenital cystic adenomatoid malformation occurs due to non-functional solid lung tissue formed by bronchioalveolar matura-
tion stopping and mesenchymal cells over multiplying in the fourth-eighth week of pregnancy. Its incidence is one in 10.000 live births and it affects male infants more (1.8:1). It occurs most commonly with acute respiratory problem in neonatal period. Cyst excision is required as infection and malignation risk is present even if it is asymptomatic.

Case report: The infant born through urgent caesarean from the first pregnancy of a 21 year old mother in the 36th week of pregnancy with a birth weight of 2080 grams was hospitalized due to fetal distress. The patient had a chest x-ray (Figure 1). In the first thorax CT taken, an appearance in line with cystic adenomatoid malformation and consolidation was present in left lung upper lobe (Figure 2). Left lung upper lobe lobectomy operation was made for the patient with a significant respiratory problem and high-parameter ventilation requirement. The pathological examination was reported to be in line with Type II cyst adenoid malformation and extralobular sequestration. In the contrasted thorax CT taken on postnatal 17th day, both lungs had an almost completely collapsed appearance and ventilated lung areas had diffused iced glass appearance. The patient whose general condition got worse and gram negative reproduction was detected in blood culture despite the ventilation treatment and supporting treatments was exitus on postnatal 44th day.

Conclusion: It is important to follow the cases with rare cystic adenomatoid malformation with a multidisciplinary approach and to operate them early if respiratory problem findings are significant.
CONGENITAL GIANT HEMANGIOMA CASE CAUSING KASABACH MERRITT SYNDROME IN NEONATALS

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Introduction: Kasabach-Merritt Syndrome (KMS) is a deep thrombocytopenia and coagulopathy presentation observed due to infantile vascular tumors. This consumption coagulopathy develops into thrombocyte activation based on endothelium defect in hemangioma, thrombosis formation by thrombocyte and fibrin and decrease in coagulation factors based on use. Transfusions should be at the lowest amount required. Especially giving thrombocytes more than required exacerbates KMS. Medical treatment containing high dose corticosteroids, antifibrinolytics and chemotherapeutics and surgical excision are the options.

Case report: The infant was born in the 37th week of pregnancy of 33 year old mother as the fifth live birth from her fifth pregnancy with a birth weight of 3600 gr in an external center. The infant was transferred to our unit after entubation due to respiratory problem as a giant mass was present in left neck anterolateral (Figure 1). The infant was followed-up on mechanic ventilator in neonatal intensive care unit. In the cranial-neck magnetic resonance imaging, huge mass lesion (11x10x10 cm) appearance in face-neck left anterolateral made us consider infantile hemangioma primarily and hyperdense areas which may belong to hemorrhage were present inside the mass in the computer tomography angiography imaging made and external karotis artery mass extended from lesion inferior to the inner part of the lesion in branches. KMS was considered in the patient who had wide spread vascularized giant hemangiomateous mass with thrombocytopenia and consumption coagulopathy. Embolisation operation was made on inner-mass external karotis branches by Interventional Radiology unit and external karotis and its branches feeding the mass were embolized with a ratio of 80% with the operation. Even though a temporary contraction was observed on the mass after operation, it was observed that it enlarged again in the follow-ups and in the control ultrasonography made, it was detected that the mass was recanalized. Re-embolisation was not considered. Oral propranolol and steroid treatment was started.

Conclusion: We wanted to share a rare KMS case occurring together with thrombocytopenia and consumption coagulopathy.

REPORTING A CASE WITH INFANTILE HEMANGIOMA DIAGNOSIS WITH THE MASS

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Introduction: Infantile hemangiomas are the most common tumors of childhood. Hemangiomas occur in 1.1-2.6% of term babies. Although the cause is unknown, hemangiomas are 2-5 times more common in females. At least 50-60% of hemangiomas are in the head and neck region. 25% of hemangiomas are located in the body, 15% are located in the extremities. Here we presented a case with a mass on the shoulder.

Case report: A 6-month-old male patient was admitted to our clinic with the complaint of a swelling in his left shoulder region which his mother saw it the day before and brought him to the hospital. In the physical examination; his general condition was good, In the systemic examination there was a mass which was approximately 2-3 cm in size and covered under the skin in the superior region of the left shoulder, and had no hyperemic appearance and no temperature increase on the region. The patient’s laboratory study results were Hgb: 10.9g/dl, Hct:31.1% , WBC: 7.730/mm3, MCV:73fl, Platelet: 308000/mm3, Free T4:1.27mIU/l, TSH:3.11mIU/l, fibrinogen:225,66mg/dl, aPTT:34.7sec, PT:13.6 sec, INR:1.12. Transfontanellar and abdominal ultrasound was normal. In Contrast magnetic resonance imaging of left shoulder reported that there was a well-defined, T1A hypointense, T2A hyperintense lesion on the left subclavian artery and ven superior, extending subcutaneously between the anterior muscle planes in the left shoulder region, with a massive lesion with diffuse contrast enhancement with hypotense signal loss foci and tubular structures selected (infantile hemangioma). Before the treatment, ECO was performed and the patient was evaluated as normal. The patient was terated with propranolol hydrochloride The 5th day was a shrinkage of the size of the mass. The patient was monitored for complications. There was no complication in the patient during the procedure. The patient was scheduled for a 6-month treatment for 2mg/kg/day of propranolol hydrochloride.

Conclusion: Patients who are consulted with mass should be evaluated with hemngioma and make study for diagnosis.
A CASE OF HYDATID CYST DETECTED BY FLEXIBLE FIBEROPTIC BRONCHOSCOPY

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Introduction: Hydatid cyst is a parasitic infestation that caused by echinococcus granulosus. In this article, we reported a pulmonary hydatid cyst case that the material is acquired by flexible bronchoscope to diagnostic purpose and confirmed pathologically.

Case Report: A sixteen years old male patient was admitted to our clinic with the complaints of an occasional shortness of breath and a chest pain in the form of stinging during inhalation for one week. He stated that he did not have cough, sputum and any other complaints. The general condition of him was good, blood pressure was 110/70 mmHg, respiration rate was 20 / min, body temperature was 36.3 °C, pulse was 88 / min. In his physical examination, otopathy was naturally detected, fine crackles and cyberlan ronkus were detected in left hemithorax. Other system examinations were detected naturally. Blood tests revealed white cell count 9300/mm3, hemoglobin 14.5 g/dl, platelet 340000/mm3, C-reactive protein 5.4 mg/L, sedimentation 12 mm/h. An opacity was seen in left paracardiac area in anteroposterior pulmonary graphy. Prominently, the lesion appear like a mass. Computed tomography revealed a non-contrasting cystic structure with a size of 4x2.5 cm in the anterior segment of the upper lobe of the left lung. Bronchocele was considered first. Eosinophilic acellular material in cyst of which texture consisting of basophilic amorphous material were detected on the pathological examination of the specimen taken by flexible bronchoscopy. Findings was compatible with cyst hydatid disease. Therefore, therapy was started and the patient was directed to the chest surgeon.

Discussion: Endobronchial extent of membrane structures on fiberoptic bronchoscopy, and radiologic pathologies such as in our case is rare in the literature. Patients are usually asymptomatic; but cough, chest pain, dyspnea, sputum, hemoptysis, fever and allergic reactions can be observed. Our present had also reported cough and shortness of breath for 1 week. Our case had chest pain and dyspnea. Radiological methods are important in diagnosis. Calcification is frequently observed hydatid cysts of the liver, but not seen in pulmonary disease. In the case of air-fluid level, air cysts and hydropneumothorax; hydatid cyst should be keep in mind. Imaging methods in the diagnosis of cystic formation weren’t sufficient for definitive diagnosis. Fiberoptic bronchoscopy was performed for this reason. Hydatid cyst detected. Our case is presented to emphasize that flexible bronchoscopy may be beneficial because of the diagnosis with histopathology when we think hydatid cyst in cases with properly limited heterogeneous mass appearance on radiology. (Note: This poster is also submitted to the XX. Congress of Turkish Thorax Society, April 5-9, 2017, Antalya)

CASE REPORT: TRACHEOSTOMY MYIASIS WOUND CAUSED BY SARCOPHAGA HAEMORRHOIDALIS

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Introduction: Tracheostomy is a term used to describe the surgical method of drilling through the trachea to provide airway patency to patients who need long-term mechanical ventilation with have respiratory failure. For patients with tracheostomy who are looked after at home, to provide enough training for tracheostomy patient care decrease both infection risk and rate of hospitalization. Wound myiasis is the infestation of open wounds of mammalian hosts caused by larvae of various species of flies. In this case report, because of not giving enough care training to the relatives of a patient with SMA diagnosis, reproduction of sarcophagahemorrhoidalis larvae in tracheostomy wound will be presented.

Case Report: In July 2016, the tracheostomy was opened in the external center to an 11-month-old male patient who was hypotonic from birth. About a month later after he was discharged from hospital, he applied to the same center for a complaint of seeing many whitish larvae around the tracheostomy. All accessible larvae were cleared by otorhinolaryngology clinic. Because of the suspicion of larval aspiration, the patient was directed to our center for bronchoscopy. In his history, we learned that the patient was discharged without training to his family in basic subjects such as tracheostomy mouth and skin care, recognition of infections and airway management and emergency tracheostomy cannula replacement. The patient was hospitalized for wound myiasis infection treatment. Bronchoscopy was planned for the patient. Collected larvae were sent to our university department of the veterinary for microscopic and parasitic analysis. It was found that the isolated parasite was originated from the species sarcophaga hemorrhoidalis. Because of the suspicion of larval aspiration, a flexible fiberoptic bronchoscope was performed to the patient and it was seen that there wasn’t any presence of larvae in the airway, as well as any damage to the wall of the trachea.

Conclusion: The success of the pediatric tracheostomy and prevent the infection is directly related to the quality of the relationship between family, patient, and physician, as well as to the family’s having adequate training about patient care.