PS-08-027
Fetal kidney and bladder agenesis: a rare case
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Objective: Reporting a case of a fetus with 25 w + 6 days gestation which underwent pregnancy interruption because of urologic anomalies.
Method: Mother was 29 years and case report corresponds to the 3rd pregnancy. Twenty-four week ultrasound scan confirmed previous ultrasound results of left kidney and bladder agenesis. X-ray of fetus showed fusion of C2-C3 spinous apophyses. Termination of pregnancy was decided. Autopsy was performed.
Results: Fetus had 826,76 g and female external genitalia. Other malformations besides the ones previously reported were observed: agenesis of left ureter, uterus, fallopian tubes and proximal portion of vagina; multicystic dysplasia of right kidney; fistula from the right ureter to the lower portion of the vagina. Lungs were hypoplasic. Ovaries and other organs were normal.
Conclusion: Ultrasound, x-ray and autopsy findings allow us to make the diagnosis of Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) with MURCS association (Mullerian agenesis, Renal agenesis, Cervicothoracic Somite abnormalities). MRKH often involves missing internal genital organs in genetic females. MURCS is rare and a complex version of MRKH affecting approximately 1/4 500 female newborns. It can be sporadic or autosomal dominant. No responsible gene was yet found. Autopsy remains the gold standard exam for fetal diagnosis.

PS-08-028
Congenital cytomegalovirus infection: an autopsy case
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Objective: Human cytomegalovirus (CMV) is the most prevalent congenital viral infection. Primary, reactivation, or recurrent CMV infection can occur in pregnancy and can lead to congenital CMV infection. Congenital CMV infection can result in sensorineural hearing loss, cerebral palsy, microcephaly, cognitive impairments, and mental retardation. Although 90–95 % of infants with congenital CMV infection are normal at birth, some of these infections are very serious and can be fatal.
Method: We report an autopsy case of congenital CMV infection.
Results: A male fetus at 37 week gestation was delivered unexplained intrauterine fetal death. The fetus weighed 3,200 g and had a crown heel lenght of 50 cm. The fetus was severely macerated and internal organs revealed autolysis. Microscopically, a diagnosis of CMV infection was made based on the presence of cytomegalic intranuclear and intracytoplasmic inclusion bodies were noted in the thyroid, lungs and kidneys. Immunohistochemically, cytomegalic inclusion bodies were positively stained by anti-CMV antibody.
Conclusion: We consider fetal autopsies can provide understanding of main causes in stillbirth. If autopsy can not be performed, needle biopsies taken from thyroid, lung and kidney may help in the diagnosis of congenital CMV infection.

PS-08-029
Pulmonary Interstitial Glycogenosis (PIG): a developmental disorder?
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Objective: PIG, previously described as infantile cellular interstitial pneumonia, is included in the pediatric interstitial lung diseases (ILDs) group and seen exclusively in infants, especially under 6 months. PIG is characterized by an increased number of mesenchymal cells with abundant intracytoplasmic glycogen expanding the interstitial septa.
Method: We present the case of a 12-h-old, full-term newborn admitted to the neonatal unit with history of tachypnea and diffuse bilateral interstitial opacities on chest x-rays. An open lung biopsy was performed. Paraffin sections were stained with H-E, PAS and thricrome. Immunohistochemistry and electron microcopy were also performed.
Results: H-E sections showed a diffuse involvement of the lung parenchyma with thickening of the interalveolar septa by mesenchymal cells with abundant cytoplasm, containing granular PAS-positive material. Ultrastructurally, these cells showed scarce organelles and abundant intracytoplasmic dot-like material, consistent with glycogen. Adjacent alveolar spaces presented immature features. Inflammatory cells were absent.
Conclusion: PIG is an unusual and underreported entity which we must consider in infants with tachypnea and a diffuse bilateral interstitial radiological pattern, since it has a better prognosis than other ILDs if treated with steroids. The presence of areas with immature alveolar structures and the abnormal differentiation of mesenchymal cells support a developmental origin of this disorder.

PS-08-030
Epithelial Wilms Tumours (nephroblastoma) in the SFCE/SIOP2001 protocol: clinical, pathological and molecular analysis
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Objective: To focus on clinical, pathological and molecular data of epithelial Wilms tumor (WT) included in the SFCE/SIOP2001 protocol.
Method: Out of 250 WT, 19 epithelial WT: clinical, pathological, Molecular Ligand Probe Amplification (MLPA) using P380-*X*2 Wilms'tumour probmix in 10 cases and SNP Illumina in one case were analyzed. All children were treated according to SIOP2001.
Results: Mean age at diagnosis: 23 months (7 months–9 years). Mean response to chemotherapy: 19 % (0–60 %). Mean epithelial component: 85 % (70–95 %). Stage 1 (n=14), stage 2 (n=1), stage 3 (n=1), stage 4 (n=3). Nephrogenic rests in 4 cases. MLPA anomalies in 3 out of 10: 1q and N-Myc gain, 16q loss and 16p gain and 1p loss and 1q gain. In this last case, SNP showed mosaic 1p loss and 1q gain. One relapse and one death at follow-up (51–360 months).
Conclusion: Epithelial WT represented 7.6 % of WT. They affected children at a mean age of 23 months and were mainly stage 1 (73 %). MLPA profile was normal in 70 %. Molecular anomalies were similar to the other WT types. Relapses/deaths were observed in 10 %.

PS-08-031
New pathomorphological approaches to study of appendicitis in children
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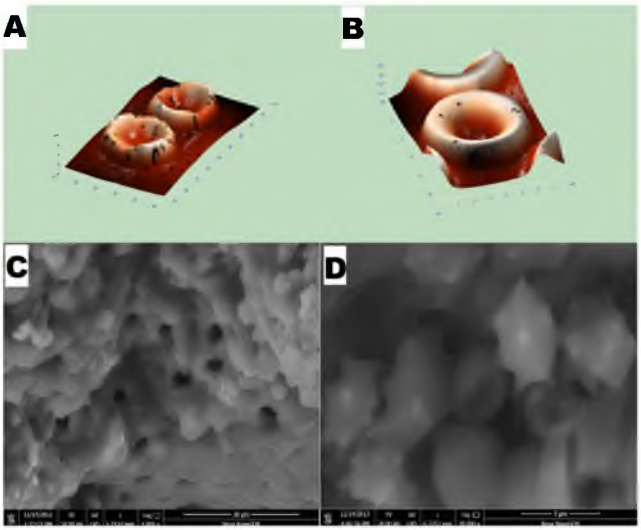
Objective: The modern setting of indications to surgical treatment of appendicitis is still complex problem in spite of reached success and wide usage of modern diagnostic methods.
Method: The researches of erythrocytes of venous blood of children with superficial (catarrhal) (10) and phlegmonic(destructive) (35) appendicitis were performed. The cells were explored with help of atomic power microscopy. The removed appendixes were explored with help of light

microscopy, scanning probe microscopy as well as atomic power microscopy.

Results: The increasing of surface area and diameter of central fossa of erythrocyte are characteristically for destructive appendicitis(7.85± 0.36 um and 5.69±0.43 um) in contrast to catarrhal appendicitis(7.37± 0.18 um and 5.1±0.28 um). Also there are a tendency to increasing of cell's surface (98.83±10.54 um2 and 91.36±6.44 um2), decreasing of height of erythrocyte(534±125 um and 553±105 um), and depth of its central fossa (373±114 um and 395±155 um) and presence of big quantity of pores to 5 nm in diameter.

Conclusion: The increasing of erythrocyte's diameter and its central recess are characteristically for all forms of acute appendicitis and they can be additional criteria for diagnosis and estimation of severity of the patient's condition.

Fig. 1. A. The alteration of form and size of erythrocytes at acute destructive appendicitis. B. Normal erythrocyte. Atomic power microscopy. Three-dimensional histogram. C. D. The wall of appendix at phlegmonic appendicitis. SEM. C. x5000 D. x20000:



PS-08-032

Rare congenital pulmonary malformation with diagnostic challenging: congenital Pulmonary Lymphangiectasia, report of four autopsy cases

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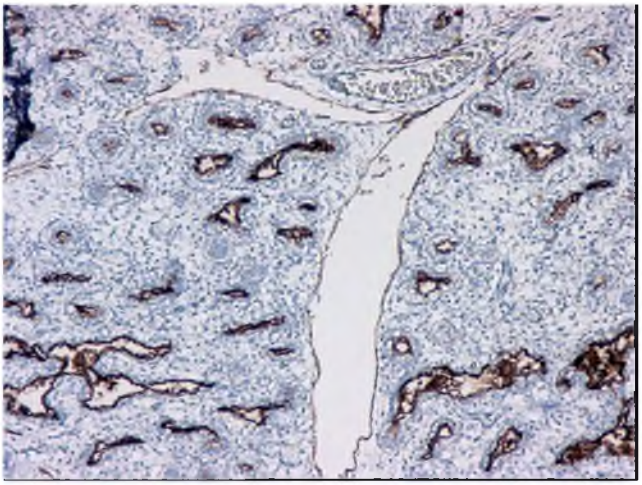
Objective: Congenital pulmonary lymphangiectasia (CPL) is a rare congenital disorder that typically presents with intractable respiratory failure in the first few days of life. There is an association non-immun hydrops and CPL.

Method: In this study we reviewed four CPL cases between January 2006–January 2014 among 684 fetal-pediatric autopsies.

Results: All cases were in the second trimester. In light microscopy there were marked dilatated channels in the subpleural -peribronchial-subseptal region of the lungs. The channels were lined with flattened cells which were expressing CD 31 and D2-40, negative for CD34. Although pulmonary interstitial emphysema (PIE) was considered an important differential diagnosis, a giant cell reaction surrounding the interstitial cystic lesions, a histological hallmark of PIE.

Conclusion: In fetal autopsy examination CPL should be recognized if there is a fetus with pleural effusion, non-immune hydrops. There is no clinical evidence for CPL.

D2-40 positivity in CPL:



PS-08-033

Diagnostic pitfall for placeta: placental mesenchymal dysplasia

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Objective: Placental mesenchymal dysplasia (PMD) is an increasingly recognizable abnormality. Early cases of PMD have been confused with partial hydatidiform mole. PMD first described in 1991 as a rare lesion of placenta which is also known as mesenchymal stem villous hyperplasia. PMD is probably under-diagnosed because of being an unfamiliar clinical entity and also mistaken for gestational trophoblastic disease because of similar sonographic findings of two entities. Because of relatively recent recognition of PMD, the data is limited about the effected pregnancies.

Method: In this report we describe clinical, macroscopical, histopathological findings of PMD among two cases.

Results: Thirythree-week-preterm baby of a 26-year-old women with cardovascular disease had 342 g placenta. And 19-week fetus with trisomy 21 of a 40 year-old women was terminated. In both cases macroscopically thick walled vessels and microscopically hydropic villous with peripherically localised thick walled vessels, without trophoblastic cell proliferation were observed.

Conclusion: These two cases represents a rare placental anomaly which is benign but it is challenging to distinguish PMD from a complete mole. PMD should be included in differatial diagnosis for sonographic findings that show a normal appearing fetus with cystic lesions of placenta. PMD has a relation-ship with pregnancy related hypertension.

PS-08-034

Embryonal rhabdomyosarcoma of the urachus in a child with Costello Syndrome

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Objective: Costello syndrome (CS) is a rare inherited autosomal dominant disease. It is characterized by mental retardation, cardiovascular, musculoskeletal abnormalities, tumour predisposition, and HRAS gene mutation. Solid tumors, most commonly rabdomyosarcomas, occur more frequently in CS than in the general population.

Method: We report a case of a 2-years-old boy with CS genetically confirmed, that was admitted in our hospital with acute abdominal complaints and fever. An intestinal intussusception with 50 mm of extension was seen in a routine abdominal