

P18.09

Prediction of prognosis and outcomes in pregnancies with a prenatal diagnosis of congenital diaphragmatic hernia compared to postnatal detectionP. Conner¹, J. Hammarqvist Vejde¹, B. Frenckner², C. Mesas Burgos²¹*Department of Obstetrics and Gynecology, Karolinska University Hospital, Stockholm, Sweden;* ²*Department of Pediatric Surgery, Astrid Lindgren's Children's Hospital, Stockholm, Sweden*

Objectives: To assess the ability to estimate the prognosis using ultrasound measurements of observed to expected lung to head ratios (O/E LHR) and compare postnatal outcomes in pregnancies with a prenatal detection of congenital diaphragmatic hernia (CDH) with outcomes of treated children lacking a prenatal diagnosis at the same institution.

Methods: A retrospective review of all children with a diagnosis of CDH and evaluated at the department of Pediatric surgery during 2006–2014. Comparisons were made between 45 infants referred to the hospital for surgery without a prenatal diagnosis and 36 pregnancies with a known diagnosis of CDH during pregnancy managed through the Fetal medicine unit.

Results: The distribution of left and right sided hernias was similar in both groups (79%/16%). In the prenatal group, observed to expected lung to head ratios were significantly different between survivors and cases with neonatal demise at all three observations during 21, 29 and 34 gestational weeks. A cut off was observed at 35% O/E LHR with lower survival below this level. Neurologic and pulmonary morbidity among survivors was observed in 15% and 25% respectively. Two infants with bilateral hernias and one child with Trisomy 18 were not operated due to poor prognosis at prenatal assessment. Survival to discharge and 1 year survival were significantly higher in the postnatal group 95% and 91% vs. 77% and 72% in the prenatal group ($p < 0.05$, OR:3.9 (1.2–13.9). Need for ECMO was 24% vs. 42% and significantly fewer 27% vs. 75% ($p < 0.001$) required repair with a patch in the postnatal group compared to the prenatally diagnosed respectively.

Conclusions: Children born with CDH having a prenatal diagnosis represent a population with a more severe condition compared to treated infants with a late postnatal diagnosis. They have considerably poorer outcomes with higher needs of ECMO, use of patch and lower survival rates. Risk for lower survival was possible to predict when O/E LHR was below 35% at prenatal evaluation.

P18.10

Prenatal sonographic signs of biliary atresiaO. Shen¹, H. Nagar⁴, R. Rabinowitz¹, E. Yaakobowitz³, E. Granot³, D. Chen²¹*Department of Obstetrics and Gynecology, Shaare Zedek Medical Center, Jerusalem, Israel;* ²*Tel Aviv University, Tel Aviv, Israel;* ³*Kaplan Medical Center, Rehovot, Israel;* ⁴*Pediatric Surgery, Shaare Zedek Medical Center, Jerusalem, Israel*

Objectives: This is a retrospective study designed to identify possible signs of biliary atresia on prenatal ultrasonography.

Methods: We identified patients with non syndromic biliary atresia, who underwent a Kasai procedure from two pediatric gastroenterology clinics from Tel Aviv and Jerusalem. We retrieved the prenatal records, fetal anomaly scan reports and digital images. The reports and images were scrutinized for the presence of the fetal gallbladder, cysts in the porta hepatis region, liver echogenicity, thickened wall of the portal veins (triangular cord sign) and other sonographic signs associated with biliary atresia in children.

Results: 55 cases were identified. 30 cases were excluded from the study; 13 did not have anomaly scans, in 16 cases prenatal images were not found or were of insufficient quality to be reliably assessed and one case had syndromic biliary atresia. 25 cases with non syndromic biliary atresia had good quality images for review and these cases constituted the study group. 7/25(28%) cases had an absent or abnormal gallbladder. Six of these had non visualisation of the gallbladder and one had an abnormally small gallbladder. In no cases was an abnormal cyst shown in the area of the hepatic hilum and in no cases was an abnormally thickened wall of the right or left portal veins depicted. One of the cases with a non-visualised gallbladder had interrupted inferior vena cava without heterotaxy.

Conclusions: Prenatal non-visualisation of the fetal gallbladder or a small gallbladder may be associated with biliary atresia. None of the other signs found on pediatric sonography were present.

P19: CASE REPORTS: ABNORMALITIES OF THE NERVOUS SYSTEM, FACE, HEART AND VESSELS

P19.01

Prenatal MRI diagnosis of frontonasal dysplasiaS.L. Laifer-Narin¹, R. Ayyala¹, C. Gyamfi-Bannerman²¹*Department of Radiology, Columbia University Medical Center, New York, NY, USA;* ²*Columbia University College of Physicians and Surgeons, New York, NY, USA*

Frontonasal dysplasia (also known as median facial cleft syndrome) is a rare congenital anomaly associated with midline phenotypic abnormalities such as midline facial clefting involving the nose, upper lip and/or palate. We present a case of a patient with 23-week intrauterine gestation who was referred for fetal MRI to evaluate for a suspected nasal mass and possible agenesis of the corpus callosum detected on prenatal screening ultrasound. Multi-planar fetal MRI was performed demonstrating agenesis of the corpus callosum, mild hypertelorism, bifid nose with lack of formation of a nasal tip and soft tissue protuberance at the tip of the nose. Postnatal imaging confirmed absence of the corpus callosum and physical examination confirmed abnormal bifid nose with soft tissue protuberance. We propose the diagnosis of frontonasal dysplasia should be considered when agenesis of the corpus callosum is seen in conjunction with 2 or more of these findings; abnormal central clefting of the nose, hypertelorism, cleft lip/palate, or incomplete formation of the anterior cranium (with or without encephalocele).

P19.02

Prenatal diagnosis of syntelencephaly: two case reports

J.H. Ochoa, J. Pautasso, G. Lovagnini Frutos

DIAGNUS Center for Prenatal Diagnosis, Córdoba, Argentina

Syntelencephaly, also known as “middle interhemispheric variant” is a rare anomaly considered a type of lobar holoprosencephaly. About 2% to 15% of holoprosencephaly cases present this variant. It is characterised by fusion of the hemispheres in the posterior frontal and parietal regions.

We report here two cases diagnosed in our Centre between 2010 and 2015. **Case1:** A 44 year-old woman, gravida 6, parity 5, was referred at 32 weeks of gestation. She had history of obesity, chronic hypertension, Chagas disease and preeclampsia in a previous gestation. Neurosonography showed absence of septum pellucidum, a corpus callosum dysgenesis with thickening of genu and splenium. The posterior aspect of the frontal lobe, the parietal lobe and lateral ventricles were fused. At this level the interhemispheric fissure