

P09: HEAD, NECK AND BONES

P09.01

Otocephaly-agnathia, proboscis and holoprosencephaly diagnosed in a fetus with Trisomy 18 using three-dimensional ultrasound and postmortem magnetic resonance imagingA. Alhousseini^{1,2}, L. Yeo^{1,2}, E.A. Hernandez-Andrade^{1,2}, S. Luewan^{1,3}, M. Garcia¹, J. Neelavalli⁴, R. Romero¹¹Perinatology Research Branch, NICHD/NIH, Detroit, MI, USA; ²Department of Obstetrics and Gynecology, Wayne State University School of Medicine, Detroit, MI, USA; ³Department of Obstetrics and Gynecology, Chiang Mai University, Muang, Chiang Mai, Thailand; ⁴Department of Radiology, Wayne State University School of Medicine, Detroit, MI, USA

Introduction: The otocephaly complex is usually a lethal syndrome characterised by agnathia and ventromedial malposition of the ears. The etiology of otocephaly is poorly understood and has never been reported in a fetus with Trisomy 18. We describe for the first time a case of otocephaly-agnathia complex with proboscis and holoprosencephaly diagnosed using three-dimensional ultrasound (3D US) and postmortem magnetic resonance imaging (MRI) in a fetus with Trisomy 18.

Case: A 43 year old Gravida 7, Para 4024 female presented at 20 5/7 weeks for ultrasound examination due to an increased risk for Trisomy 18 on maternal cell-free DNA screening. A targeted two- and three-dimensional ultrasound examination was performed. The following congenital anomalies were diagnosed (figure 1): agnathia, low-set fused ears (“bow-tie” appearance), large proboscis, holoprosencephaly, and atrioventricular septal defect. In addition, there was fetal growth restriction and polyhydramnios (amniotic fluid index of 35 cm). Amniocentesis revealed Trisomy 18, and the patient underwent a vaginal delivery at 22 weeks of gestation. Both autopsy and 7.0T postmortem MRI examinations were performed. Autopsy confirmed otocephaly with anteriorly set ears fused in the midline (synotia), lobar holoprosencephaly, and an atrioventricular septal defect. MRI demonstrated a hypoplastic maxilla, absence of the mandible and tongue, and the presence of two orbits, thereby adding further diagnostic information.

Comment: We report for the first time a case of otocephaly-agnathia complex with proboscis and holoprosencephaly diagnosed in a Trisomy 18 fetus. The use of 3D US and postmortem MRI were invaluable in making the diagnosis.

Supporting information can be found in the online version of this abstract

P09.02

Typical and atypical dural sinus malformation imaging in the fetus: based on 4 cases

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Objectives: To describe the atypical imaging characteristic of dural sinus malformation in fetuses by ultrasound and MRI.

Methods: We report a series of four fetuses with dural sinus malformation in Hubei Maternal and children's hospital from July, 2013 to February 2016. All the mothers undertook the prenatal MR imaging because of the intracranial space occupying lesions discovered by prenatal ultrasound.

Results: In all four cases, two of them represented the typical MR imaging of dural sinus ectasia with thrombosis in the vicinity of

torcular Herophili (also known as sinus confluence), while in the other two cases lesions located mainly in superior sagittal sinus and transverse sinus separately. Two pregnancies were terminated, while the outcomes of other two cases were not available.

Conclusions: Atypical location of the dural sinus malformation, such as superior sagittal sinus and transverse sinus, should be paid special attention to. Sonographer should realise the atypical sign of dural sinus malformation, and prenatal MRI is necessary to identify the lesion.

P09.03

MR of fetal neck and face masses: prediction of high morbidity neonatal outcome, including ex utero intrapartum therapy (EXIT)D.M. Twickler^{1,2}, A.A. Bailey^{1,2}, A. Beavers¹, Y. Xi¹¹Radiology, University of Texas Southwestern Medical Centre, Dallas, TX, USA; ²Obstetrics and Gynecology, University of Texas Southwestern Medical Centre, Dallas, TX, USA

Objectives: To determine MR fetal imaging parameters which predict high morbidity neonatal outcomes, including EXIT, in the setting of fetal face and neck abnormalities.

Methods: This retrospective study (2004-2016) included parameters of polyhydramnios, based on largest vertical pocket (LVP), mass effect on the trachea, mass midline extension, size, and morphologic grading characteristics. Morbid outcomes included those requiring an EXIT procedure, difficult intubation at delivery or lethal outcome. Statistical analysis included univariate analysis (Fisher's exact test, Wilcoxon and multivariable logistic regression on complete data).

Results: Of 41 cases, 5 delivered by EXIT procedures, with 1 demise within 12 hours of delivery, 1 with multiple intubation attempts, and 1 with fetal hydrops and intrauterine fetal demise. The remaining 33 delivered with no interventions or neonatal morbidity. Receiver operating characteristic curve of the largest vertical pocket of amniotic fluid and mass effect on the trachea yielded an area under the curve of 0.949 (95% CI, 0.8795 to 1) $f = -6.8141 + 0.5051 * \text{Largest Vertical Pocket} + 2.9926 * \text{Mass effect on trachea}$.

Conclusions: The LVP measurement and mass effect on the trachea were the most contributory MR parameters that predicted significant morbidity in fetuses with abnormalities of the face and neck, along with other significant parameters. These parameters predict high morbidity neonatal outcomes, including the need for EXIT procedures.

P09.03: Table 1.

MRI parameters	No Morbidity (N=33)	Morbidity (N=8)	P value
Midline extension	15(52%)	7(100%)	0.0288
Mass effect on trachea	6(21%)	7(100%)	0.0002
Cystic Vs. Solid			0.0207
1. Simple Cystic	9(31%)	0(0%)	
2. Few simple septations	6(21%)	1(14%)	
3. Thick/multi septations	5(17%)	1(14%)	
4.	4(14%)	1(14%)	
5. >50% solid	5(17%)	4(57%)	
Largest Vertical Project	5.92±2.29	10.55±3.47	0.0006
Maximum dimension (mm)	57.0±35.7	80.71±26.3	0.0525