Congenital High Airway Obstruction Syndrome

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Case Summary

A screening obstetrical ultrasound performed at 29 weeks' gestation demonstrated polyhydramnios with an amniotic fluid index (AFI) of 27.1cm (normal 10.5-18.9). Several abnormalities were present in the fetus, including shortened long bones, mesocardia with enlarged lungs, and massive ascites. A diagnostic fetal ultrasound and a fetal MRI were performed.

Imaging Findings

Fetal ultrasound (Figure 1) showed enlarged, echogenic lungs, mesocardia, a dilated fluid-filled trachea, and marked ascites. Fetal MRI (Figure 2) failed to demonstrate a cervical tracheal fluid column and confirmed the dilated, fluid-filled thoracic trachea, enlarged lungs causing mass effect upon the cardiomediastinal structures and eversion of the diaphragm, and fetal hydrops.

Diagnosis

Congenital high airway obstruction sequence (CHAOS)

Discussion

CHAOS is a rare proximal congenital airway obstruction that leads to distal airway dilatation, expanded lungs with mass effect on the heart, ascites, and hydrops.[1] The condition is characterized by partial or complete obstruction of the upper airway. The obstruction can vary in location and severity. Types of obstruction include laryngeal atresia (the most common), laryngeal stenosis and dysgenesis, subglottic stenosis, tracheal aplasia, and tracheal stenosis.[2]

If the condition goes unrecognized in utero, the fetus does not usually survive. However, if an ex-utero intrapartum treatment (EXIT) procedure is performed, survival can occur but depends on the associated congenital anomalies.

CHAOS is predominantly sporadic.[3] However, it can be seen in syndromes such as Fraser, cri-du-chat, short-rib polydactyly, and velocardiofacial (DiGeorge syndrome).[4,5] Thus, a fetus with CHAOS should be evaluated for other anomalies that may be part of a genetic syndrome.

CHAOS is diagnosed via prenatal imaging. The initial imaging study of choice is fetal ultrasound. Typical findings include enlarged hyperechoic lungs, dilated fluid-filled airways, a compressed and centrally positioned heart, flattened/inverted diaphragms, ascites, hydrops, and polyhydramnios.[6,7] Guimaraes et al recommend that the presence of these signs prompt consideration of genetic counseling and fetal MRI. The modality can demonstrate increased lung volumes with flattened or inverted hemidiaphragms, a dilated airway below the level of obstruction, massive ascites, mesocardiac, and placentomegaly, in which the placenta is disproportionately large or thicker than 4 cm.[8]

Improvements in prenatal imaging and treatment modalities have led to increased survival in fetuses with CHAOS. Prenatally, treatment can include amnioreduction.[9] While there are a small number of reports of spontaneous resolution of CHAOS,[9] the initial intervention is the EXIT procedure, which allows surgeons to secure an airway, commonly in the form of a tracheostomy, below the obstruction.[9] After birth, the airway obstruction is definitively treated via surgery. More recently, an in-utero treatment has been developed that relies on fetoscopic, ultrasound-guided decompression of the laryngeal atresia. In this technique a wire is passed across the atretic region of the airway. This is followed by balloon dilation of the airway with stent placement and laser laryngotomy.[9]

Conclusion

CHAOS is characterized by the partial or complete obstruction of the fetal upper airway that can occur sporadically or as part of a syndrome. Fetal imaging is a mainstay of diagnosis and management of the condition. Advances in imaging and treatment may reduce neonatal mortality.

References

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Figure 1. (A) Axial ultrasound through the fetal chest at 29 weeks' gestation demonstrates markedly enlarged, echogenic lungs and mesocardia. (B) Longitudinal ultrasound of the fetal chest and abdomen demonstrates the dilated lower trachea (arrow), surrounded by echogenic lungs, and the marked ascites.

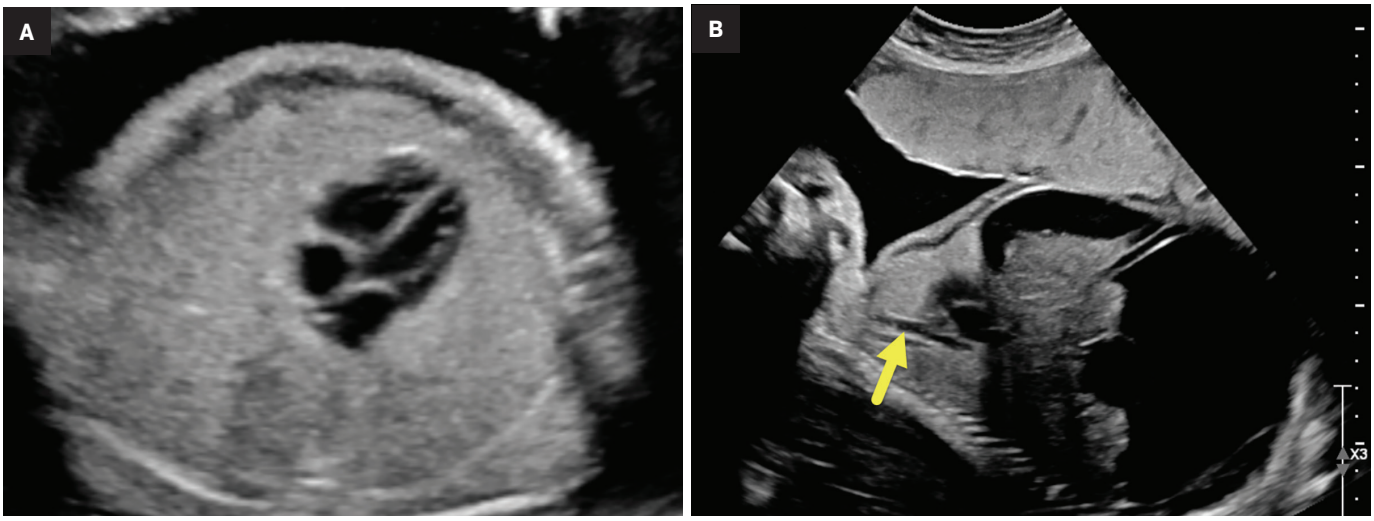
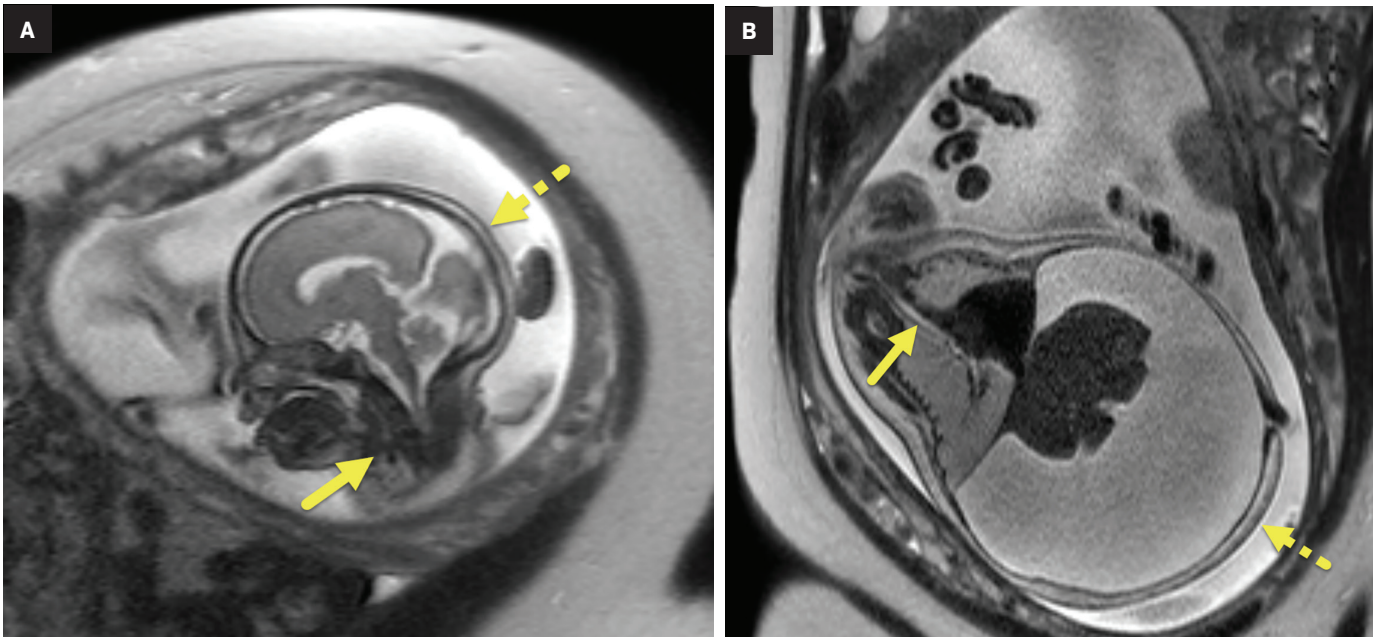


Figure 2. (A) MRI of the fetal brain and neck demarcates the atresia (arrow) of the cervical trachea indicated by the absence of fluid signal. Scalp edema (dashed arrow) results from the hydrops. (B) MRI obliquely through the fetal chest and abdomen demonstrates the dilated thoracic trachea (arrow), enlarged lung, eversion of the diaphragm, marked ascites, and skin edema (dashed arrow) from hydrops.



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