Congenital high airway obstruction sequence (CHAOS): A Case Report

Mia Ma, M.D., Beatrice Knisely, M.D., Brian Craig, M.D., Karyn Ledbetter, M.D.

Introduction:

Congenital high airway obstruction sequence (CHAOS) was coined by Hedrick and colleagues to describe fetuses with upper airway obstruction and ultrasound findings that were incompatible with life. CHAOS is often an isolated finding, but it can also be linked to certain genetic syndromes, such as Fraser syndrome.

The pathogenesis of CHAOS remains largely unknown. It is believed to be due to atresia or stenosis of the larynx or trachea, which leads to obstruction of the fetal upper airway. The obstruction results in the tracheobronchial tree's inability to reabsorb the fluid secreted by the fetal lung. Subsequently, the accumulated fluid causes increased intratracheal pressure, expands the fetal lungs and may result in oligohydramnios. The overdistended lungs compress the heart, resulting in a dysfunctional and centrally deviated cardiac system. The lack of venous return causes an accumulation of ascitic, pleuritic and pericardial fluid, which directly contributes to nonimmune fetal hydrops and eventual polyhydramnios.

Patient History:

A 35-year-old gravida 10 parity 4 female presents at 18 4/7 weeks gestation for evaluation of a possible twin pregnancy with one viable fetus with an omphalocele. She has a history of 2 spontaneous abortions and 3 elective abortions. She has a past medical history of abnormal Pap smear, chlamydia, and postpartum depression. Her past surgical history includes cervical cerclage and cesarean section. Patient has a family history of autism, Down syndrome, stillbirth, and miscarriage. The only illness during pregnancy was a pimply rash. She denies any vaginal bleeding. She denies recent travel and new sexual partners. She denies drug use, tobacco use, and alcohol use. Ultrasound imaging was obtained for further evaluation.

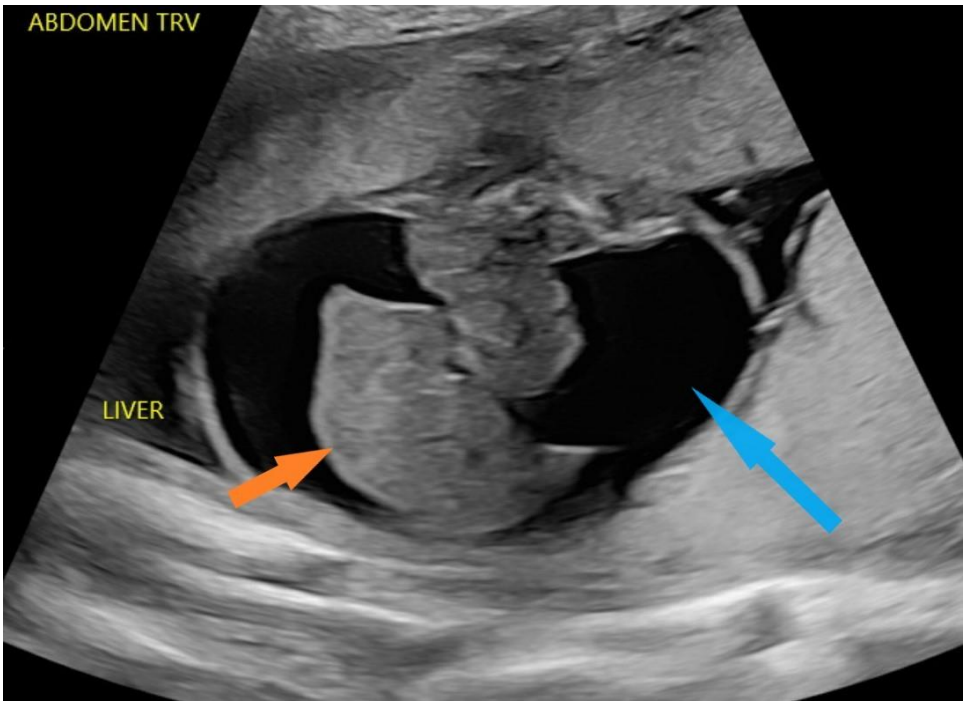

Figure 1: This axial greyscale ultrasound image demonstrates the normal liver (orange arrow) surrounded by large volume ascites (blue arrow).

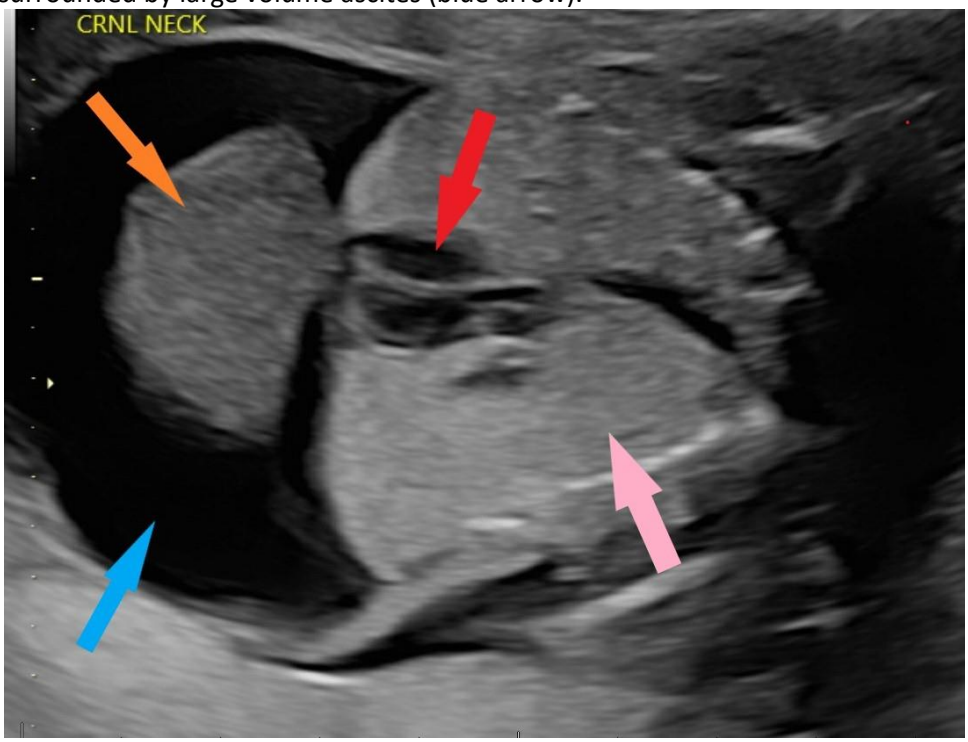

Figure 2: This coronal greyscale ultrasound image demonstrates the normal liver (orange arrow) surrounded by significant ascites (blue arrow). Additionally, the enlarged hyperechoic lungs (pink arrow) are visualized with resultant eversion of the diaphragm. The heart (red arrow) is midline and appears compressed.

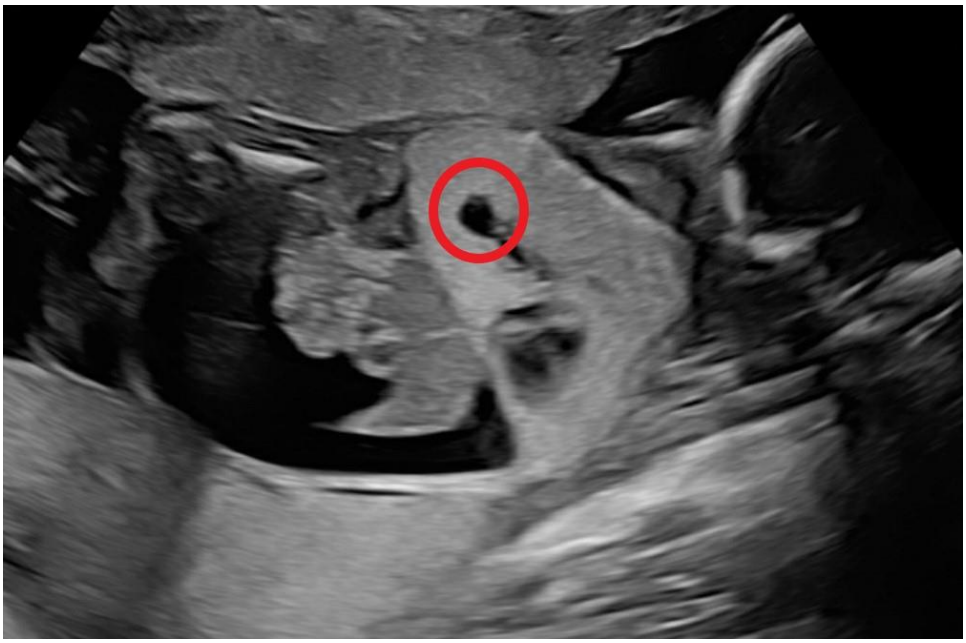

Figure 3: This coronal oblique greyscale ultrasound image demonstrates a fluid filled structure (red circle) opposite of the heart, in the right lung. Presumably, this structure is causing the high airway obstruction, leading to CHAOS syndrome.

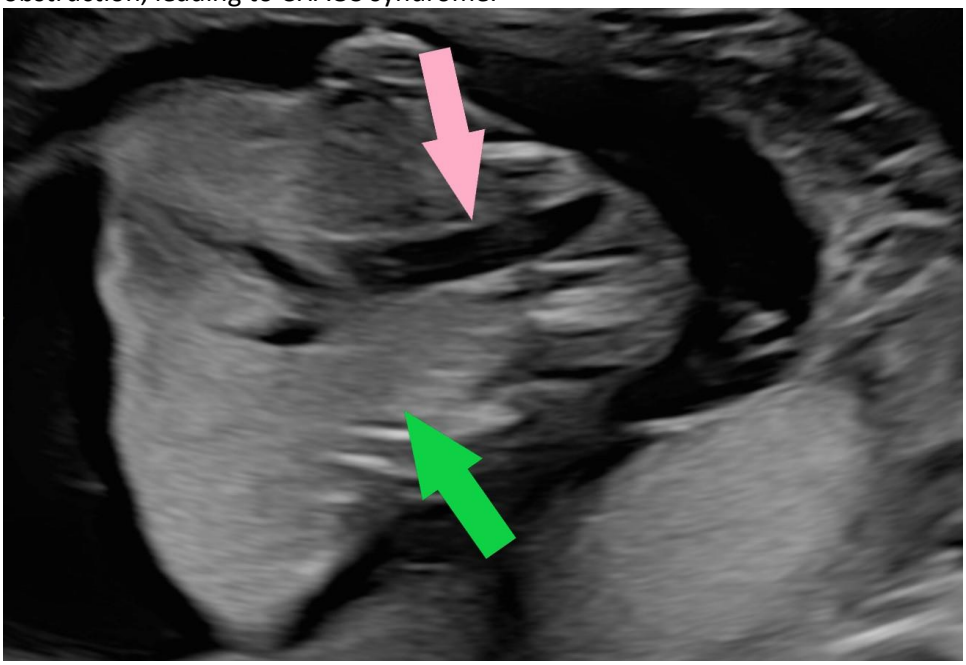

Figure 4: This greyscale ultrasound image in the coronal plane demonstrates an abnormal fluid-filled trachea (pink arrow). The enlarged hyperechoic lungs (green arrow) are again noted.

Discussion:

The classic sonographic findings of congenital high upper airway obstruction (CHAOS) include bilateral symmetric enlarged hyperechoic fetal lungs, flattened or everted hemidiaphragms, and a dilated, fluid-filled trachea. These findings are diagnosed on prenatal ultrasound, and CHAOS may be

detected in the fetus as early as 16 weeks of gestation. Displacement of the heart and severe ascites may also be seen on ultrasound. MRI can also be used to diagnose CHAOS. MRI allows for improved visualization of dilated airways with accurate detection of the level and structure of the obstruction. Although CHAOS often results in fetal demise, there is a spectrum of disease severity that may aid in indicating prognosis. The presence of fetal hydrops, ascites and severe oligohydramnios upon early imaging is an ominous sign and often results in unfavorable outcomes.

The true incidence of CHAOS remains unknown. Nevertheless, among fetuses that have been identified to have CHAOS, the morbidity and mortality rate is extraordinarily high. The ex-utero intrapartum treatment (EXIT) is one of the most recognized procedures for treating CHAOS. This technique enables clinicians to maintain placental blood circulation to the fetus while maintaining the airway by tracheostomy or intubation. The EXIT procedure should only be considered in fetuses with obstruction at a level that is compatible with an artificially established airway and in fetuses that do not have additional findings that are associated with poor prognosis, such as hydrops or genetic syndromes.

CHAOS is a rare syndrome that is important to detect early to offer interventions, such as the EXIT procedure or pregnancy termination, in a timely manner. CHAOS presents with a spectrum of clinical severity and may be associated with factors that indicate a poor prognosis, such as early hydrops or ascites. Pregnancies complicated by CHAOS may be associated with genetic disorders, resulting in intellectual disability, recurrent miscarriages and karyotypic aberrations. Therefore, it is important for radiologists to recognize the key radiological findings of CHAOS to accurately guide patient management.

References:

Ekmekci E GS, Kiziltug N. Prenatal ultrasonography findings of fetus with congenital high airway obstruction (chaos): A case report and review of literature. Clin Obstet Gynecol Reprod. 2017;3.

Lago Leal V, Cortes LM, Seco Del Cacho C. Prenatal diagnosis of congenital high airway obstruction syndrome. Indian J Radiol Imaging. 2018;28(3):366-368.

Masahata K, Soh H, Tachibana K, et al. Clinical outcomes of ex utero intrapartum treatment for fetal airway obstruction. Pediatr Surg Int. 2019;35(8):835-843.

Nolan HR, Gurria J, Peiro JL, et al. Congenital high airway obstruction syndrome (CHAOS): Natural history, prenatal management strategies, and outcomes at a single comprehensive fetal center. J Pediatr Surg. 2019;54(6):1153-1158.

Roybal JL, Liechty KW, Hedrick HL, et al. Predicting the severity of congenital high airway obstruction syndrome. J Pediatr Surg. 2010;45(8):1633-1639.

Witters I, Fryns JP, De Catte L, Moerman P. Prenatal diagnosis and pulmonary pathology in congenital high airway obstruction sequence. Prenat Diagn. 2009;29(11):1081-1084.