

Antenatal Diagnosis of Congenital High Airway Obstruction (CHAOS)

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Abstract

Congenital High Airway Obstruction is a rare and life-threatening condition which occurs due to obstruction of upper airways in the fetus. Ultrasonography is the modality of choice to diagnose this condition, characteristic features being enlarged echogenic lungs, compressed cardia occupying central axis, fetal ascites, inverted diaphragm and dilated airways. We discuss the ultrasonography features of this condition, differential diagnosis, the role of Magnetic resonance Imaging and available treatment options in such condition in this case report.

Case Report

A 26-year-old women Gravida 3, Para 2, L2 A0 was referred for an anomaly scan at 19weeks. Scan performed outside had commented on fetal ascites and patient came for anomaly scan. There was no history of consanguinity. Family history from both sides was unremarkable. Ultrasonography demonstrated bilateral echogenic lungs, centrally located compressed heart, fetal ascites, dilated trachea and inverted diaphragm. There was no abdominal wall edema, amniotic fluid was normal and other systems were unremarkable. Based on these findings a diagnosis of CHAOS was made and the patient was explained

regarding the prognosis of the pregnancy. She decided to continue her pregnancy and around 32 weeks there was fetal demise. Patient refused for autopsy.

Introduction

CHAOS (Congenital high airway obstruction) is a life-threatening condition, which occurs due to congenital obstruction of fetal airway due to tracheal or laryngeal atresia¹. This condition was first observed by Hendrick in 1994². The incidence of this condition is unknown, but if this condition is undiagnosed antenatally it progresses to stillbirth or death after delivery³. Most of the cases are diagnosed antenatally due to improvements in imaging². Classical antenatal findings include bilateral enlarged hyperechogenic lungs, inverted diaphragm, dilated airways, fetal ascites and non-immune hydrops³.

Discussion

CHAOS is a condition associated with many findings due to obstruction of upper airway tract⁴. It results due to congenital obstruction of fetal airway due to tracheal or laryngeal atresia, other causes being laryngeal cysts, obstructing tumours of the oropharynx and in the cervical region¹. The incidence of this condition is unknown⁴. Airway obstruction causes reduced clearance of fluid produced by fetal lungs which leads to increase in

intratracheal pressure, hyperexpansion of lungs and abnormal development². Further progression of this condition leads to thinning of alveolar walls and reduction of type II pneumocytes and surfactant. This progresses to hyper-expansion of lungs and compresses the cardia and inferior vena cava and reduce the venous return and results in non-immune hydrops⁴. Diaphragm becomes flat or inverted depending on the severity of the condition³. Identification of fluid-filled trachea and bronchus is very important in diagnosing this entity¹.

Ultrasonography is the main diagnostic modality in diagnosing CHAOS antenatally³. On antenatal USG (ultrasonography), CHAOS presents as enlarged echogenic lungs, flattened or inverted diaphragm, dilated airways, and fetal ascites¹. The heart is displaced anteriorly and abnormally positioned with a central axis and dilated proximal airway up to the level of obstruction. CHAOS must be differentiated from other external causes of laryngotracheal obstruction, such as cervical teratoma, lymphatic malformation and vascular rings like a double aortic arch⁴. CHAOS is associated with polyhydramnios as fetal swallowing is impaired, however, impaired swallowing can also cause oligohydramnios, the authors are of the opinion that amniotic fluid quantity is not a reliable parameter in this condition³.

There are instances of spontaneous inutero improvement of findings, which is attributed to spontaneous perforation or fistulization of airway obstruction⁴. Various studies found improvement in lung volumes, hyperechogenicity, reduction in ascites and diaphragmatic eversions between 22 and 32 weeks⁵. CHAOS has to be differentiated from CPAM and other causes of external airway obstruction and vascular causes such as double aortic arch, identification of dilated airway is a very important finding to differentiate CHAOS from other causes of lung masses². MRI is an additional imaging tool to evaluate the

dilated airway and assess the level of obstruction in case of planned surgical intervention³. On MRI, enlarged lungs show increased signals, other features of CHAOS are also well visualized⁴.

CHAOS is associated with certain syndromes. It is associated with Fraser's syndrome which is characterized by tracheal or laryngeal atresia, cryptorchidism, microphthalmia, renal agenesis, facial clefting, mental retardation, syndactyly or polydactyly and musculoskeletal abnormalities⁴. Other syndromes associated with CHAOS are short rib-polydactyly syndrome (SRPS), Shprintzen-Goldberg Omphalocele syndrome (SRPS) and VATER / VACTERL association, in addition to these CHAOS is also associated with some chromosomal abnormalities like deletions of 22q11.2, deletion of chromosome 5p, 47XXX, partial trisomy 16q and partial trisomy 9¹. Detailed evaluation of all cases of CHAOS for coexistence of any genetic syndromes is very important to evaluate the implication of inheritance in future pregnancies³.

Prenatally diagnosed cases of CHAOS can be managed by EXIT procedure where fetal head and chest is delivered first and airway is secured while maintaining the uteroplacental circulation with an option of tracheostomy to be kept open if required⁴. Chances of neonatal survival are better in case of a well-planned EXIT procedure which is performed at the time of controlled near term caesarean section⁵. Intrauterine foetoscopic laser laryngotomy is benefitted in a small section of patients⁴.

Figures and Legends

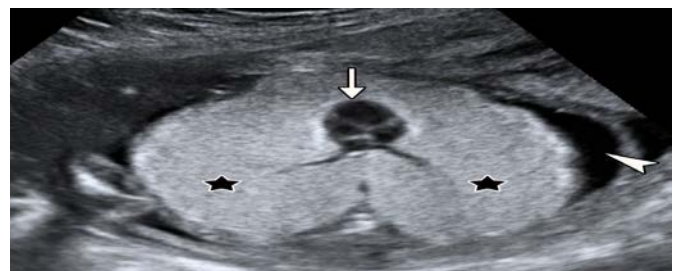


Fig1. Axial ultrasound image of a 19weeks foetus at the level of thorax shows bilateral echogenic lungs (star) causing cardiac compression with central displacement of the heart (arrow) and ascites (arrowhead).



Fig 2. Coronal ultrasound image of a 19weeks foetus shows bilateral echogenic lungs (star) causing cardiac compression with central displacement (arrow) ascites (arrowhead) and inverted diaphragm (curved arrow).



Fig 3. Coronal ultrasound image of a 19weeks foetus at the level of thorax shows bilateral echogenic lungs and dilated trachea (arrow).

Conclusion

CHAOS is an uncommon cause of echogenic lungs. Antenatal ultrasonography is the investigation of choice to diagnose this entity. CHAOS has various characteristic features which can be diagnosed by ultrasonography. CHAOS is associated with certain genetic syndromes and

chromosomal abnormality so it is very important to methodically evaluate other systems for the syndromic association. MRI is an adjunct imaging investigation for evaluating the level of obstruction in case of planned intervention. A well-planned EXIT procedure gives a very good chance of survival in fetuses with CHAOS.

Abbreviations: CHAOS- congenital high airway obstruction, USG- ultrasonography, CPAM-congenital pulmonary airway malformation, MRI-magnetic resonance imaging, EXIT-ex utero intrapartum treatment.

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