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CASE REPORT

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Creating Order Out of Chaos – Role of Antenatal Ultrasound in Diagnosis

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Summary

Background: Congenital high airway obstruction syndrome (CHAOS) is a rare fetal anomaly characterized by obstruction of the higher fetal airway. This could be either complete or incomplete and is more commonly seen at the subglottic level, resulting in a spectrum of characteristic secondary features.

Case Report: In this case study, we report two cases of CHAOS with one showing laryngeal atresia and the other, tracheal atresia. Both these cases showed characteristic findings on a detailed, meticulous USG examination which led to this diagnosis.

Conclusions: Early and accurate diagnosis offers a window of opportunity for parental counseling and management using procedures such as EXIT (ex-utero intrapartum procedure). Earlier, CHAOS was thought to be incompatible with life; however, with the advent of ex-utero intrapartum procedure, a few cases of post-natal survival have been noted in the literature. In this article, we emphasize the sonographic findings found in CHAOS. Early diagnosis offers an opportunity for a intrauterine fetal intervention in potentially lethal cases.

MeSH Keywords: Prenatal Diagnosis • Stillbirth • Ultrasonography

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Background

Congenital high airway obstruction syndrome (CHAOS) is defined as a complete or incomplete obstruction of the fetal upper airways. The obstruction may be either due to luminal narrowing or extrinsic compression.

We report two cases of CHAOS diagnosed by antenatal ultrasound. A transabdominal obstetric scan was performed in both cases using the PHILIPS HD11XE device with a low frequency, (2–5 MHz) curvilinear probe.

Case Report

Case 1

A 30-year-old, gravida 3, patient was referred to our ultrasound unit at 19 weeks of gestation. A transabdominal obstetric scan revealed mild fetal hydrops with moderate ascites (Figure1). Both lungs appeared bulky and echogenic with a resulting increase in chest circumference. The diaphragm was everted due to the grossly enlarged lungs. The fetal heart was central, small in size and seen to be squeezed between the echogenic lungs. However, the heart was structurally normal (Figure 2). There was abrupt transition in the fetal laryngeal lumen with the dilatation of fluid-filled tracheobronchial tree, suggestive of laryngeal atresia (Figure 3). On the basis of the above findings, a diagnosis of CHAOS due to laryngeal atresia was made. No other congenital abnormality was detected.

Case 2

A 23-year-old, primigravida female was referred to our ultrasound unit at 20 weeks of gestation. A transabdominal obstetric scan revealed an abrupt transition in the lumen of trachea with a dilatation of the tracheobronchial tree. Both fetal lungs appeared bulky and echogenic (Figure 4). This resulted in eversion of the diaphragm. The fetal heart was displaced to the center of thorax and appeared small in size (Figure 5). Minimal fetal ascites was also noted. No other significant abnormality was noted. A diagnosis of CHAOS secondary to tracheal atresia was made.

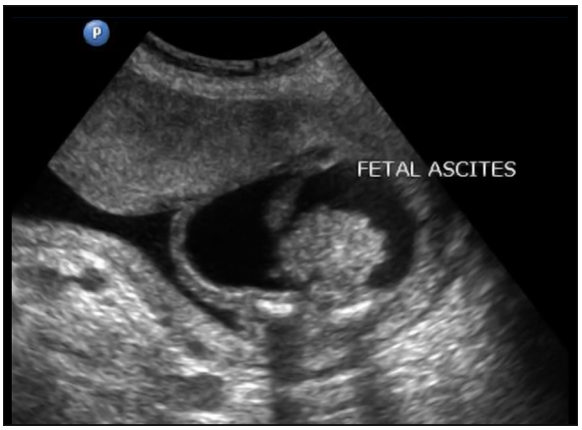

Figure 1. Antenatal ultrasound at the level of abdomen showing gross fetal ascites.

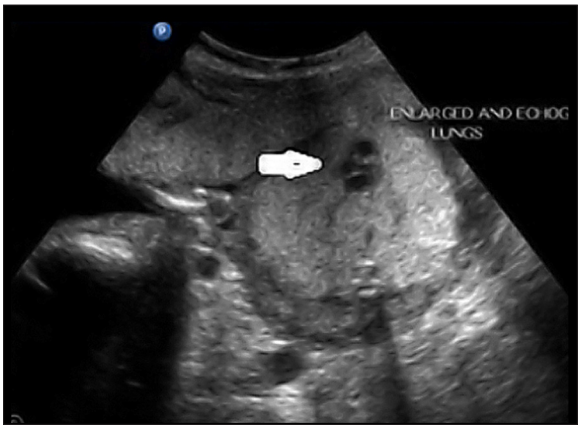

Figure 2. Antenatal ultrasound image at the level of mid-thorax showing bilateral, bulky echogenic lungs with centrally compressed heart.

Both pregnancies underwent medical termination following consultation with respective families. On religious grounds, autopsy of fetuses could not be performed.

Discussion

The term CHAOS was coined by Hedrick MH et al. and it broadly denotes a prenatal diagnosis of upper airway obstruction without a specific cause.

Congenital high airway obstruction syndrome is a constellation of fetal abnormalities due to obstruction of the high fetal airways. This obstruction can be due to either intrinsic or external compression. Extrinsic compression can be caused by lymphatic malformations, vascular rings or masses such as teratomas. Intrinsic causes are mostly secondary to laryngeal atresia or tracheal atresia [1]. Embryologically, laryngeal atresia results from failure of recanalization of the larynx, whereas tracheal atresia results from unequal partitioning of the foregut into the esophagus and trachea [2]

CHAOS can occur in isolation or in association with various syndromes, the most common of which is "Fraser syndrome"[3]. CHAOS due to tracheal atresia is relatively rare, with two cases described by Artunc Ulkumen B et al. [3].

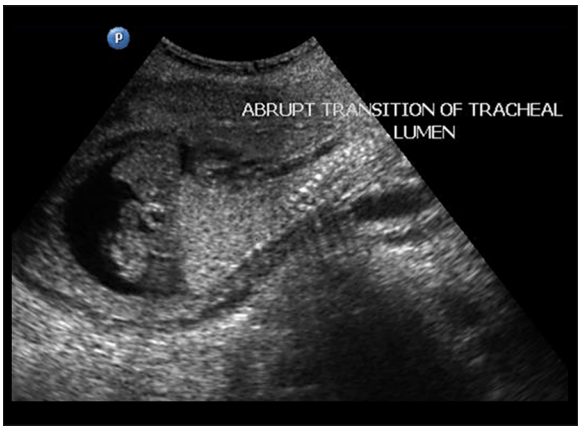

Figure 3. US showing a dilated tracheobronchial tree and an abrupt cutoff of the laryngeal lumen.

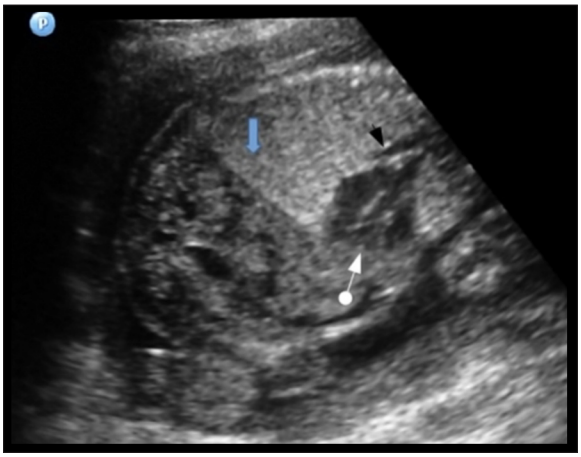

Figure 4. Coronal image showing everted diaphragm (blue arrow), dilated bronchi (black arrowhead), central heart (white arrow).

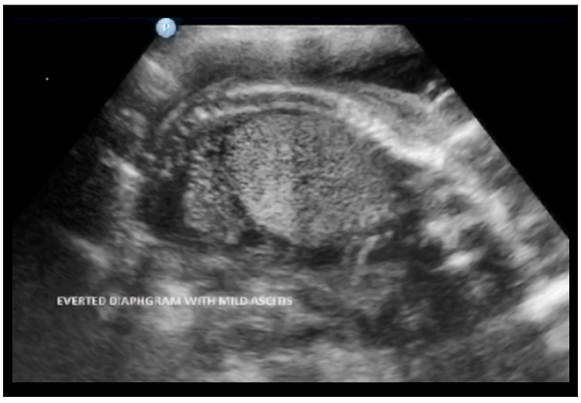

Figure 5. Bulky, echogenic lungs with everted diaphragm and mild ascites.

The prevalence is lower than 1: 50000, with a male-to-female ratio of 2: 1 [4]. Unilateral findings of CHAOS highly suggest a diagnosis of bronchial atresia [5].

There are three possible presentations of CHAOS:

1. Complete laryngeal atresia without tracheoesophageal fistula.

2. Complete laryngeal atresia with tracheoesophageal fistula.
3. Near-complete high airway obstruction.

Understanding the pathophysiology helps considerably in making a prenatal diagnosis and instituting perinatal management.

In the normal fetus, tracheobronchial secretions are gradually cleared out. In case of obstruction of higher airways, there is retention of secretions in the tracheobronchial tree. The increased intra-tracheal pressure causes fluid accumulation in both lungs.

Usually, normal amniotic fluid index is seen. However, polyhydramnios is commonly noted in cases of laryngeal atresia due to the decreased fetal swallowing of amniotic fluid. This could be due to compression of the esophagus by the lung and/or compression of the stomach by ascites. Oligohydramnios may be present in certain cases, such as Fraser syndrome, or when there is impaired swallowing or renal agenesis [6].

There are numerous case reports demonstrating both high sensitivity and specificity of ultrasound in antenatal diagnosis of CHAOS [7,8]. Dilated airways, enlarged lungs with flattened diaphragm, fetal ascites and severe nonimmune hydrops can be observed [4]. Other indirect signs are curved descending aorta and inferior vena cava due to mass effect. In the majority of cases, fetal ascites is present, possibly due to a pressure effect on IVC. The fetal heart appears small and in a more central and anterior position. This eventually leads to heart failure which in turn leads to placentomegaly and hydrops fetalis [7].

There are certain sonographic maneuvers which may help in localizing the site of narrowing and they include visualization of the fetal neck in the coronal plane and the use of color Doppler over fluid-filled, dilated tracheobronchial tree for detecting loss of flow signal at the onset of respiration [9]. Transvaginal ultrasonography is a better alternative than transabdominal sonogram for early detection of laryngeal atresia.

The major differential diagnoses include CCAM (congenital cystic adenoid malformation) type III, pulmonary sequestration, congenital diaphragmatic hernia and mediastinal teratoma. Most of the above-mentioned disorders are unilateral, whereas CHAOS is invariably bilateral. The most specific sonographic finding that helps in differentiating CHAOS from other conditions is the dilatation of the tracheobronchial tree.

Although previously thought to be totally fatal, neonatal survival is now possible by ex-utero intrapartum treatment (EXIT) procedure where the fetal airway is repaired before the cessation of fetomaternal circulation [10]. Fetal laryngoscopy has been used in to achieve decompression of obstruction of the larynx or trachea in CHAOS [11,12].

Conclusions

CHAOS is a rare, lethal, fetal malformation due to a primary obstruction of fetal higher airways. Accurate and early diagnosis of this condition allows for parental counseling and may aid in a proper management. In our opinion, the role of MRI should be reserved for postpartum confirmation of diagnosis in situations where consent for pathological autopsy is not obtained either on religious grounds or due to personal preferences. Ultrasound, if carefully performed, should allow for an accurate and prompt diagnosis of this condition. We have found ultrasound to be a reliable method for diagnosing CHAOS.

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Conflict of interest

No conflict of interest from all authors.

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