Original Research Article

A case series study in CHAOS syndrome

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Abstract

Background: Congenital high airway obstruction syndrome (CHAOS) is a series of events due to a obstruction of the fetal airway that blocks the larynx or trachea. Prenatal diagnosis of patients with CHAOS is necessary so that perinatal treatment can be undertaken or elective termination of pregnancy can be done. Combined approach of several academic disciplines comprising of paediatricians, radiologists and obstetricians is key to effective management. We also briefly review the available literature. We report two cases of CHAOS syndrome diagnosed on antenatal ultrasonography.

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INTRODUCTION

Congenital high airway obstruction syndrome (CHAOS) is a series of events due to a obstruction of the fetal airway that blocks the larynx or trachea, which may be partial or complete. It is usually a fatal syndrome. Antenatal diagnoses is key to effective management. Due to advances in imaging techniques, the syndrome can be recognized prenatally. The true incidence of CHAOS is unknown, an incidence of 1 per 50,000 newborns is described. Our aim of this paper was to familiarize the radiologists and OBGYN specialists with the classical USG features of this syndrome. We describe here two cases of CHAOS diagnosed on antenatal ultrasonography.[1,2]

MATERIALS AND METHODS

We describe the classical radiological features of CHAOS syndrome in two patients referred to our tertiary care hospital. Ultrasound was performed on Philips IU22 machine. PCPNDT formalities were taken care of.

OBSERVATIONS AND RESULTS

Case 1:

A 24 year-old G2P1 female came for routine second trimester antenatal ultrasound known as the anomaly scan. Clinical history did not reveal any predisposition to increased risk for genetic or familial disorder. Initial first trimester ultrasound scan was done at 8 weeks which did not show any significant abnormality. USG showed a single foetus with: bilaterally symmetrical enlarged echogenic lungs, dilated airways below the level of obstruction, centrally placed and compressed heart, fetal ascites with crumpled organs – CHAOS syndrome. Other differentials like cystic adenomatous malformation of the lung and Bronchopulmonary sequestration were ruled out. Patient underwent medical termination of pregnancy. Autopsy confirmed the findings and also showed that left renal agenesis (a case of Fraser syndrome). However, because of ascites and crumpled nature of organs, we were unable to properly visualized either kidneys of the fetus.

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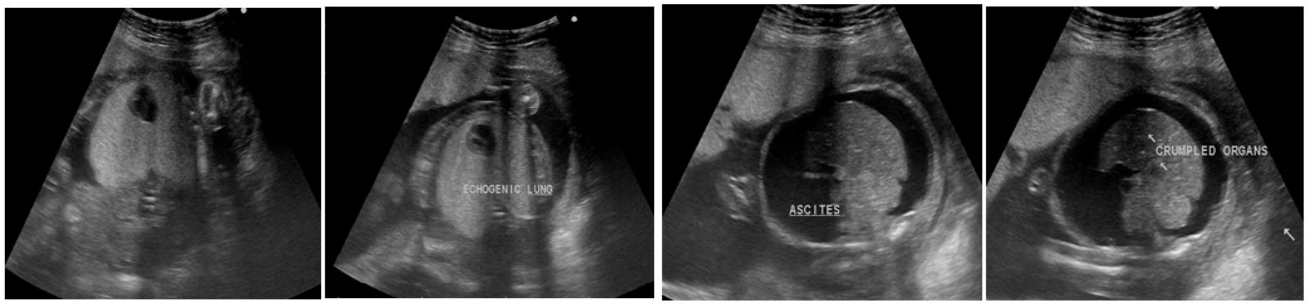

Figure 1a **Figure 1b** **Figure 1c** **Figure 1d**

Figures 1a and 1b: show bilateral enlarged echogenic lungs with compressed heart; **Figures 1c and 1d**: show gross fetal ascites with resultant central compression of abdominal organs.

Case 2:

A 22-year-old G2L1 was referred for obstetric sonography at around 21 weeks of gestation. There was no significant family or personal history. USG showed a single gestation corresponding to 21 weeks 4 days with bilaterally symmetrical enlarged echogenic lungs, flattened diaphragm, centrally placed and compressed heart, fetal ascites, bilateral cystic dysplastic kidneys and anhydramnios – CHAOS syndrome. Other differentials like cystic adenomatous malformation of the lung and Bronchopulmonary sequestration were ruled out. We could not find any literature describing association between CHAOS syndrome and bilateral multicystic dysplastic kidneys (MCDK). More literature / studies are needed to study the association between CHAOS and bilateral MCDK. As gestational age was over 20 weeks – medical termination of pregnancy was not be done. Patient and concerned OBGYN opted for follow up. On follow up sonography, approximately 7 weeks later showed resolution of ascites. Rest of the findings showed no interval change. There are very few cases of CHAOS described with spontaneous antenatal improvements. The literature explains that this may be owing to spontaneous perforation or tracheoesophageal fistula with the refining of the obstructed fluid leading to decrease in the pressure of airways and reversal of the process.

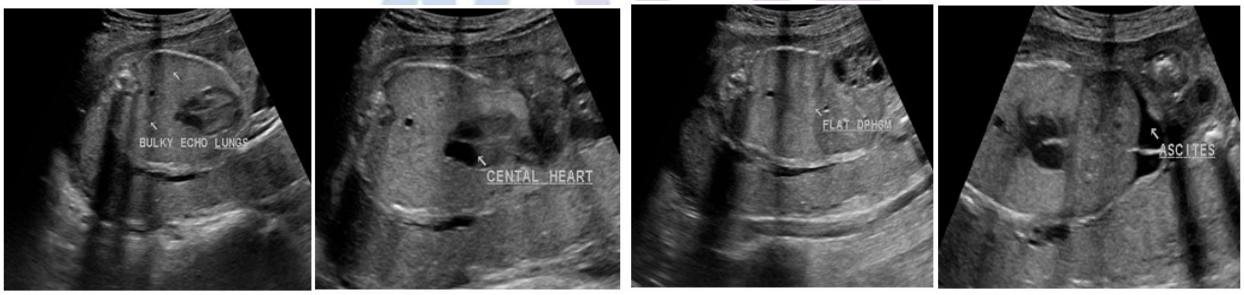

Figure 2a **Figure 2b** **Figure 2c** **Figure 2d**

Figures 2a and 2b: show bilateral enlarged echogenic lungs with centrally displaced heart; **Figure 2c:** shows flattening and eversion of diaphragm; **Figure 2d:** shows moderate foetal ascites.

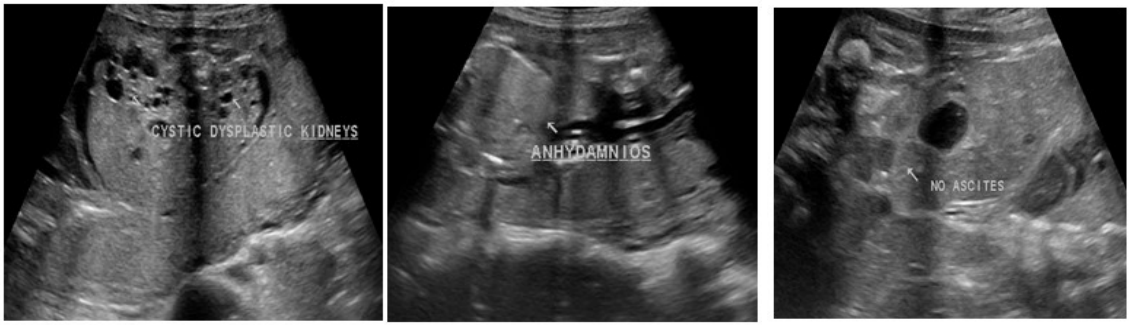

Figure 2e **Figure 2f** **Figure 2g**

Figure 2e: shows bilateral multicystic dysplastic kidneys; **Figure 2f:** shows anhydramnios; **Figure 2g:** shows resolution of ascites on follow up scan.

DISCUSSION

CHAOS Syndrome is a very rare congenital disease with etiopathogenesis being deficient recanalization of the upper airways around the 10th week of gestation. The root cause of CHAOS therefore is an intrinsic obstruction of the upper airway**.** These include laryngeal atresia, laryngeal web, laryngeal cyst, and tracheal atresia or stenosis. In a normal foetus, the fluid secreted by foetal lung is absorbed by the tracheobronchial tree. In case of obstruction in the tracheobronchial tree, there is decreased clearance of fluid produced by the foetal lungs with resultant increase in intratracheal pressure and as a result hyperexpansion of lungs. The hyperexpanded lungs cause compression of the heart and inferior vena cava which causes decreased venous return and dysfunctional cardiovascular system. This ultimately ends in ascites and non immune hydrops. The diaphragm flattens or inverts according to the severity of the mass effect of lungs. These series of events are responsible for the prenatal imaging characteristics of CHAOS.[3] Cases with resolution of pulmonary enlargement on follow up in later pregnancy have also been described indicating less severe course of the disease due to development of pharyngotracheal or laryngotracheal communications. Early in pregnancy oligohydramnios is seen due to decrease in amniotic fluid secondary to obstruction. Later on in pregnancy polyhydramnios may develop because of oesophageal compression and thus decreased foetal swallowing**.** Oligohydramnios is also seen in Fraser syndrome (renal or ureteral agenesis, cryptophthalmous, syndactyly, ambiguous genitalia and laryngeal atresia) which is the most common association with CHAOS.[4]

The main diagnostic tool for prenatal diagnosis of CHAOS is sonography. USG findings are *(5):*

a. Bilateral large hyperechoic lungs
b. Dilated tracheobronchial tree
c. Small, compressed, and centrally placed heart
d. Flattened or inverted diaphragm
e. Ascites

Syndromic associations: As told earlier, the most common associated genetic disorder with CHAOS is Fraser's syndrome. CHAOS may be also a part of Cri-du-Chat syndrome, short-rib polydactyly syndrome, and velocardiofacial syndrome . This warrants the necessity of a detailed evaluation of all CHAOS suspected cases to avoid significant implications of inheritance in future pregnancies. The other reported associations include short long bones, facial cleft, flexion deformities, radial and tibial aplasia, renal agenesis, oesophageal atresia, microphthalmia, and single umbilical artery.[6]

Role of fetal MRI : All the structural findings described on USG can also be recognized on MRI. MRI is especially needed if any fetal surgical intervention is planned. MR imaging can more accurately locate the dilated airway up to the level of obstruction.[7]

*Treatment***:** CHAOS is thought to be equivalent to a foetal death. However, especially if CHAOS due to incomplete obstruction , is diagnosed in the late 2nd or in the 3rd trimester and if severe hydrops has not occurred yet, the EXIT procedure (ex utero intrapartum treatment) can be offered. The common objective of the procedure being to settle an intact airway for the baby before the foetomaternal circulation is stopped.[8]

CONCLUSION

CHAOS has various peculiar imaging appearances and familiarity with the imaging presentations of CHAOS is of key importance for the radiologists and OBGYN specialists, resulting in timely diagnosis and thereby effective management of this disease. In summary prenatal early diagnosis of patients with CHAOS is necessary so that perinatal management can be undertaken successfully or elective termination of pregnancy can be undertaken.

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