International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Congenital High Airway Obstruction (CHAOS) Syndrome: A Rare Case Presentation

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Received: 16/08//2014 *Revised: 05/09/2014* *Accepted: 07/09/2014*

ABSTRACT

Congenital high airway obstruction syndrome (CHAOS) is a near fatal condition of multifactorial inheritance, in which the fetus has a dilated trachea, enlarged echogenic lungs, an inverted or flattened diaphragm, and ascites. A case of CHAOS, diagnosed antenatally on USG at 19 weeks of gestation, is being reported here.

Key words: Congenital high airway obstruction syndrome; Echogenic lungs; Dilated trachea; Ascites

INTRODUCTION

Congenital high airway obstruction syndrome (CHAOS) is a condition in which the fetus has hyperinflated, enlarged, and highly echogenic lungs; an inverted or flattened diaphragm; a dilated tracheobronchial tree; and ascites. It occurs as a result of congenital obstruction of the fetal airway secondary to laryngeal atresia, tracheal atresia, or a laryngeal cyst. [1-2] The disease is generally incompatible with life and, therefore, antenatal USG diagnosis is desirable.

CASE REPORT

A 22-year-old multiparous (gravida) woman at 19 weeks gestation was referred for a fetal well-being examination. There was no history of consanguinity and the family history was unremarkable. Her first pregnancy was still birth in a local hospital, the cause could not be established due to lack of follow up.

USG showed bilateral enlarged hyperechoic lungs (Fig1b), a dilated trachea (Fig 2) and principal bronchi (fig 3), inferiorly displaced and flattened diaphragms (fig 5), moderate fetal ascites (fig 5), a small heart because of compression by the obstructed lungs(fig 1b). However, amniotic fluid volume was normal. These findings were diagnostic of CHAOS. We discussed the possible unfavorable outcome of the pregnancy with the parents who chose not to terminate the pregnancy because of personal reasons.

Fig 1 (a) (b)

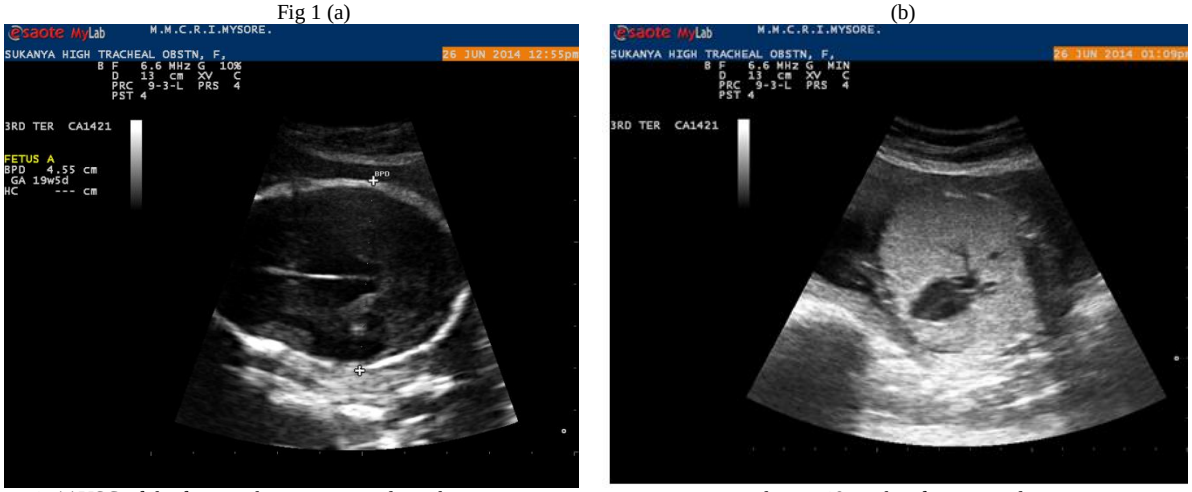

Figure 1 :(a)USG of the fetus in the transverse plane shows intrauterine gestation corresponding to 19 weeks of gestational age.
(b)USG of the fetus in the transverse plane shows bilateral enlarged hyperechoic lungs.

Fig 2 (a) (b)

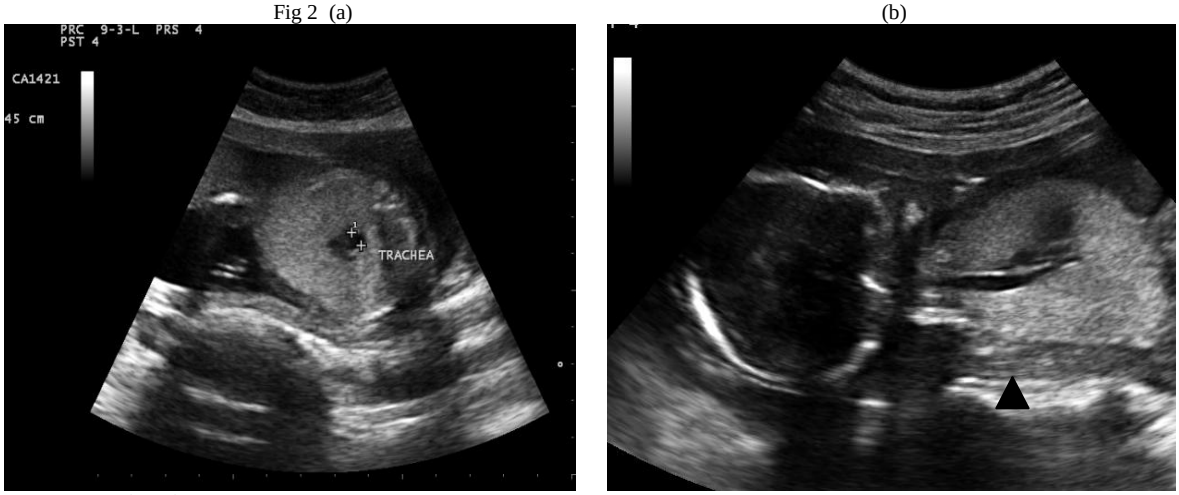

Figure2 :(a)USG of the fetus in the transverse plane shows dilated trachea
(b) USG of the fetus in the saggital plane shows dilated trachea (arrowhead)

Fig 3 (a) (b)

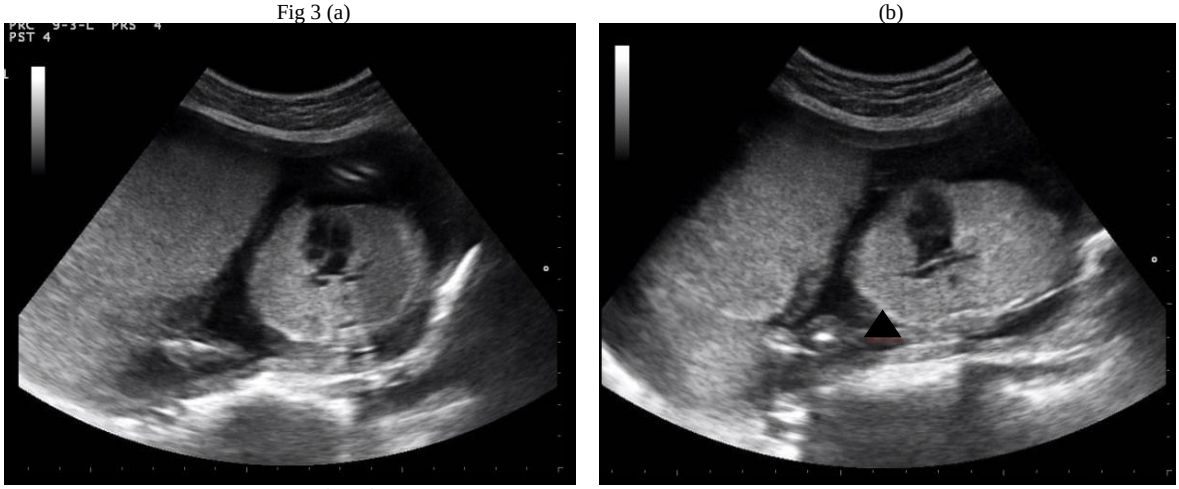

Figure3 :USG of the fetus in the transverse plane shows dilated principal bronchi (arrowhead)

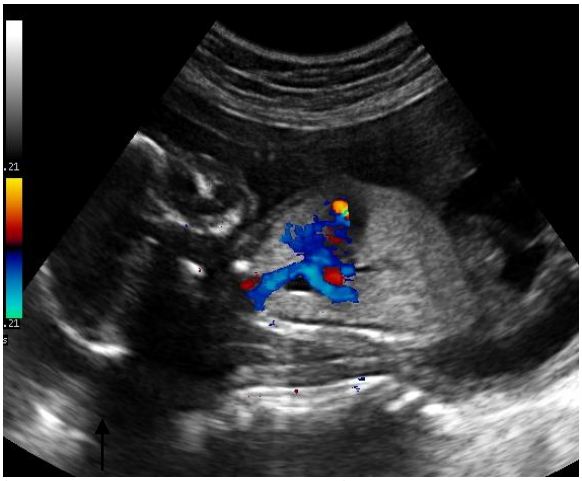

Figure 4: USG of the fetus in the sagittal plane, at the level of the thorax shows a dilated trachea (black arrow).

Fig 5 (a) (b)

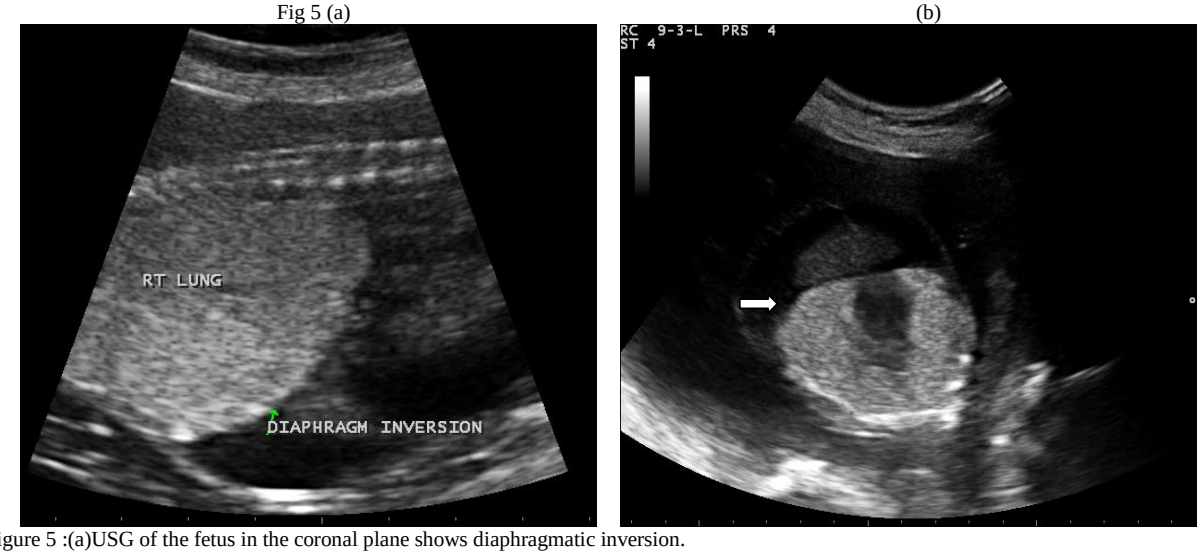

Figure 5 :(a)USG of the fetus in the coronal plane shows diaphragmatic inversion.
(b)USG of the fetus in the transverse plane shows ascites(white arrow)

DISCUSSION

Congenital High Airway Obstruction Syndrome **(**CHAOS) was defined by Hedrick et al in 1994 as upper airway obstruction that is diagnosed in utero by ultrasound, with concomitant findings of large echogenic lungs, flattened or inverted diaphragms, dilated airways distal to the obstruction, and fetal ascites or hydrops. [3]

The obstructed airway results in decreased clearance of the fluid produced by fetal lungs and increased intratracheal pressure which causes the lungs to expand and develop abnormally. This causes thinning of the alveolar walls, reduction of Type II pneumocytes, and reduced surfactant. This further leads to hyperexpanded lungs which cause compression of the heart and inferior vena cava. Ultimately, these events culminate in decreased venous return and lead to non-immune hydrops. [4]

The identification of airway obstruction is important to establish the diagnosis of CHAOS and to distinguish it from other causes of echogenic lungs such as bilateral congenital cystic adenomatoid malformation (CCAM) or sequestrations. It is also necessary to distinguish CHAOS from extrinsic causes of airway obstruction.

The level of airway obstruction needs to be established with accuracy if any intervention is planned as it can help to choose between foetal and neonatal interventions. [4]

Oligohydramnios can develop before 20 weeks because of the absence of the lung liquid contribution to the total volume of the amniotic fluid. Polyhydramnios may be an associated feature, occurs subacutely, usually after 28weeks, and develops after the compressive obstruction of the esophagus and after stopping of the fetal swallowing. Non immune hydrops and placentomegaly may be the result of the decrease venous return after the increase of the pressure on the heart and on the great veins. [5]

Although most cases of CHAOS are sporadic, some cases have been linked to genetic syndromes, the commonest being Fraser's syndrome. This comprises of laryngeal or tracheal atresia, cryptophthalmos, microphthalmia, renal agenesis, orofacial clefting, mental retardation, musculoskeletal anomalies and syndactyly or polydactyly. Other syndromes which have reported in association with CHAOS are Cri-du-chat syndrome, short-ribpolydactyly syndrome, and velo-cardio-facial syndrome. [6-8]

Antenatally diagnosed cases of CHAOS may be offered an EXIT (ex utero intrapartum treatment) procedure which consists of delivery of the foetal head and chest to secure an airway while simultaneously maintaining the uteroplacental circulation,with tracheostomy being necessary in most cases. Cases of spontaneous antenatal improvement in CHAOS due to spontaneous perforation also suggest that intrauterine foetoscopic laser laryngotomy may be beneficial in a small subset of these patients. [9-11]

The lung lesion that has to be considered in the differential diagnosis of CHAOS is the microcystic solid form of CCAM (type 3). In contrast to laryngeal or tracheal congenital obstruction, CCAMs are generally unilateral lesions that are very rarely associated with diaphragm flattening and never with dilated airways.

CONCLUSION

Congenital high airway obstruction syndrome is a rare cause of congenital airway obstruction which is incompatible with life. Antenatal imaging with ultrasound usually shows typical findings which can lead to a diagnosis. The accuracy of prenatal diagnosis is useful in planning perinatal surgical assistance.

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How to cite this article: Nanjaraj CP, Basavaraj, Manupratap N et. al. Congenital high airway obstruction (CHAOS) syndrome: a rare case presentation. Int J Health Sci Res. 2014;4(10):342-346.

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