Congenital high airway obstruction syndrome—antenatal diagnosis of a rare case of airway obstruction using multimodality imaging

Surg Cdr Piyush Joshi*, Col Lovleen Satija†, Col RA George#, Col S Chatterjee**, Surg Cmde J D'Souza‡, Sqn Ldr Abdul Raheem##

MJAFI 2012;68:78–80

INTRODUCTION

Congenital high airway obstruction syndrome (CHAOS) is a rare condition which occurs as a result of congenital foetal airway obstruction which may be complete or partial. The disease is usually incompatible with life and leads to stillbirth or death shortly after delivery. However, due to advances in antenatal imaging by ultrasound and magnetic resonance imaging (MRI), more cases are being detected in the antenatal period. Commonly, CHAOS occur secondary to atresia or stenosis of the larynx or trachea, which may be partial or complete or due to a laryngeal cyst. We report a case of CHAOS due to laryngeal atresia diagnosed by antenatal ultrasonography and foetal MRI. We also briefly review the pertinent literature.

CASE REPORT

A 27-year-old primigravida presented for a routine second trimester antenatal ultrasound at 18 weeks gestational age. Ultrasound examination using a convex transducer (4.0–5.5 MHz, Logiq P5, GE Medical Systems, Milwaukee, USA) revealed that the foetus had large echogenic lungs causing inversion of the diaphragm. The heart was centrally placed and appeared compressed, and there was minimal ascites. The trachea and principal bronchi appeared dilated (Figure 1). However, liquor quantity was normal.

An MRI study was carried out on a 1.5 Tesla scanner (Symphony, Siemens AG, Germany) using T2 weighted (half-Fourier acquisition single shot turbo spin echo [HASTE]; TR 4.3 ms, TE 2.15 ms) and TrueFISP (TR 1000 ms, TE 83 ms) sequences in sagittal, coronal, and axial planes which showed large markedly hyperintense lungs (as compared to foetal skeletal muscle) causing inversion of the diaphragms. The dilated trachea was exquisitely demonstrated as increased signal with a gap at the level of the larynx. The cardiac findings and ascites were also confirmed and no additional anomaly could be demonstrated (Figure 2).

Based on ultrasound and MRI findings, diagnosis of CHAOS due to laryngeal atresia was made. The parents were counselled regarding the relatively poor prognosis and the pregnancy was terminated.

DISCUSSION

Congenital high airway obstruction syndrome is a constellation of findings which arise due to obstruction of the upper airway tract. The causes of obstruction include laryngeal atresia, stenosis or laryngeal cysts, and tracheal atresia or stenosis.^{1–3} Cases of CHAOS are sporadic and the exact incidence is not known.

Figure 1 Antenatal ultrasound coronal image showing dilated trachea (long arrow), large echogenic lungs, inverted diaphragm (short arrows) and foetal ascites.

*Classified Specialist (Radiodiagnosis), Command Hospital (SC), Pune – 40, †Senior Advisor (Radiodiagnosis), Command Hospital (CC), Lucknow, #Senior Advisor (Radiodiagnosis), Military Hospital, Mhow, **Associate Professor, Department of Radiology, AFMC, Pune – 40, ‡Consultant (Radiology), INHS Asvini, Mumbai, ##Graded Specialist (Radiology), 5 Air Force Hospital, Jorhat.

Correspondence: Surg Cdr Piyush Joshi, Classified Specialist (Radiodiagnosis), Command Hospital (SC), Pune – 40.
E-mail: piyushjoshi@hotmail.com

Received: 16.12.2010; Accepted: 19.09.2011
doi: 10.1016/S0377-1237(11)60111-1

Figure 2 Antenatal magnetic resonance imaging sagittal half-fourier acquisition single shot turbo spin echo image demonstrating large lungs with increased signal causing diaphragmatic inversion, dilated trachea (long arrow) and foetal ascites (short arrow).

Though till 2007 only about 52 cases had been reported,⁴ it is suspected that the actual incidence is higher as increasingly more cases are being recognised.³

Congenital high airway obstruction syndrome should be distinguished from extrinsic causes of tracheolaryngeal obstruction such as lymphatic malformation, cervical teratoma, and vascular rings like double aortic arch. Accurate distinction of these conditions is essential for planning subsequent management.⁵

The obstructed airway results in decreased clearance of the fluid produced by foetal 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.³

This sequence is responsible for the typical imaging appearance of CHAOS. Sonography shows large echogenic lungs and their enlargement relative to the chest wall may cause flattening or inversion of the diaphragm. The proximal airway appears dilated up till the level of the obstruction. The heart may appear displaced anteriorly with an abnormal centrally positioned axis and may appear small due to compression by the lungs. Ascites and other features of non-immune hydrops may be present. Polyhydramnios is commonly seen though some workers have also reported oligohydramnios. In addition some workers have reported abnormal jerky foetal breathing movements.^{1,2}

The MRI findings include increased lung volumes with abnormally increased signal. There may be flattening or inversion of the diaphragm, small anteriorly displaced heart with centrally positioned axis, ascites, and other features of non-immune hydrops. The MRI also shows a dilated airway up to the level of the obstruction and is better at identifying the level of obstruction due to higher intrinsic soft tissue contrast.³

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.³

There have been few reports of spontaneous improvement in utero which have been attributed to spontaneous perforation or fistulisation of the airway obstruction.^{3,6} Some workers have also proposed that a subset of patients have small laryngo-tracheal communications and are associated with a milder course and perhaps even resolution. However, they caution that a high index of suspicion needs to be maintained as the primary pathology may not have resolved even after resolution of the ultrasound findings.⁷

Antenatal ultrasound is the primary modality used for the diagnosis of CHAOS. Increasingly MRI is being used as an adjunct for confirming of the level of obstruction, to exclude extrinsic causes of obstruction, and to assist in the differential diagnosis. The MRI is considered superior to ultrasound in identifying the level of obstruction.⁸

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.^{1,3,4} Other syndromes which have reported in association with CHAOS are Cri-du-chat syndrome, short-rib polydactyly syndrome, and velo-cardio-facial syndrome.

It has been proposed that CHAOS may be a part of an association, which has been given the acronym TACRD (Tracheal Agenesis, complex Cardiac anomalies, Radial ray defects, and Duodenal atresia). This is distinct from the more common VACTERL (Vertebral, Anorectal, Cardiac, Tracheo-Esophageal, Renal, Limb anomalies) association which has trachea-oesophageal fistula and not tracheal agenesis as a component.⁴ Hence, a suspicion of CHAOS on imaging should prompt a search for associated anomalies too.

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.¹⁻³ 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.³

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. However, MRI has an adjunctive role in demonstrating the level of obstruction which may not always be identified on ultrasound and in excluding extrinsic causes of obstruction. This is especially important in case any intervention is being contemplated.

CONFLICTS OF INTEREST

None identified.

REFERENCES

1. Gareth P, Seaward R. The fetal chest. In: *Diagnostic Ultrasound* 3rd ed. Rumack CM, Wilson SR, Charboneau JW, Johnson JM, eds. St Louis: Mosby 2005:1303–1321.
2. Garg MK. Case report: antenatal diagnosis of congenital high airway obstruction syndrome – laryngeal atresia. *Indian J Radiol Imaging* 2008;18:350–351.
3. Guimaraes CV, Linam LE, Kline-Fath BM, et al. Prenatal MRI findings of fetuses with congenital high airway obstruction sequence. *Korean J Radiol* 2009;10:129–134.
4. Cadichon SB. Congenital high airway obstruction syndrome. In: *Congenital Malformations: Evidence-based Evaluation and Management*. Kumar P, Burton BK, eds. New York: McGraw-Hill Professional 2007:135–138.
5. Courtier J, Poder L, Wang ZJ, Westphalen AC, Yeh BM, Coakley FV. Fetal tracheolaryngeal airway obstruction: prenatal evaluation by sonography and MRI. *Pediatr Radiol* 2010;40:1800–1805.
6. Lim FY, Crombelholme TM, Hedrick HL, et al. Congenital high airway obstruction syndrome: natural history and management. *J Pediatr Surg* 2003;38:940–945.
7. Vidaeff AC, Szmuk P, Mastrobattista JM, Rowe TF, Ghelber O. More or less CHAOS: case report and literature review suggesting the existence of a distinct subtype of congenital high airway obstruction syndrome. *Ultrasound Obstet Gynecol* 2007;30:114–117.
8. Mong A, Johnson AM, Kramer SS, et al. Congenital high airway obstruction syndrome: MR/US findings, effect on management, and outcome. *Pediatr Radiol* 2008;38:1171–1179.