International Journal of Radiology Research
www.radiologyjournal.in
E-ISSN: 2663-4562, P-ISSN: 2663-4554
Received: 15-07-2020; Accepted: 30-07-2020; published: 16-08-2020
Volume 2; Issue 2; 2020; Page No. 01-03

Congenital high airway obstruction syndrome: A rare congenital disorder

Dinesh Chataut[1], Sundar Suwal[2], Kamal Subedi[3], Bishnu Gautam[4], Prashant Khatiwada[5]
[1-4] Department of Radiology, Tribhuvan University, Institute of Medicine, Tribhuvan University Teaching Hospital, Kathmandu, Nepal
[5] Department of Radiology, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal

Abstract
Congenital High Airway Obstruction Syndrome (CHAOS) is a very rare congenital disorder with partial or complete laryngeal or tracheal atresia. Prenatal diagnosis is important as immediate post-natal intervention only can improve survival from this otherwise fatal disorder. Ultrasonography is primary imaging modality for its diagnosis, which can be supplemented by MRI by identifying level of obstruction and ruling out extrinsic compression to trachea or larynx. We present a case of 28 years old woman presented for routine anomaly scan at 18-19 weeks with fetal ultrasound features of CHAOS.

Keywords: CHAOS, prenatal diagnosis, ultrasonography

1. Introduction
Congenital high airway obstruction syndrome (CHAOS) is a rare congenital disorder in which there is complete or partial blockage of the fetal airway, either due to intrinsic atresia of the trachea or glottis, or due to extrinsic compression. Although the true incidence of CHAOS is unknown, incidence of 1 per 50,000 newborns has been described in literatures [1].
In normal fetus, the fluid secreted within the lungs are eventually absorbed within the tracheobronchial tree. However, when tracheobronchial obstruction occurs, the secretions within the lungs are not drained. Thus enlarged lungs in the fetus squeezes heart and mediastinal great vessels. The heart may become small and dysfunctional. Also there will be decreased venous return, which along with dysfunctional heart results in fetal ascites and hydrops fetalis [2]. It may be associated with other congenital anomalies as well [3]. Prompt post-natal airway intervention may allow survival of the baby in this otherwise fatal condition. Thus, antenatal diagnosis desirable, which can be done with antenatal ultrasonography (USG) or Magnetic Resonance Imaging (MRI) [4].

2. Case Report
A 28- year old woman presented for routine anomaly scan at 18-19 weeks of gestation in the department of radiology. She had previous bad obstetric history as fetus was terminated in second trimester due to suspected omphalocele in prenatal USG. This time, 1st trimester scan did not show significant anomalies, thus amniocentesis was not done in 1st trimester. The anomaly scan at 18-19 weeks of gestation showed bilateral enlarged and homogeneously hyperechoic lungs occupying most of the thoracic cavity with compressed heart in between. Posterosuperiorly, trachea was dilated. Bilateral diaphragms were inverted. Moderate free fluid collection and floating bowel loops were noted in the fetal abdomen. No significant pleural fluid collection seen. No other congenital anomalies were detected in the fetus. Based on these ultrasound pictures, diagnosis of congenital high airway obstruction syndrome (CHAOS) was made. (Figure 1 and 2)
The husband was explained about the prognosis of the CHAOS and advised for fetal MRI. But they decided to terminate the fetus. Patient was also asked for postmortem of the terminated fetus but they refused to do so.

3. Discussion
Congenital high airway obstruction is caused by complete or partial obstruction of the fetal upper airways most likely by laryngeal atresia. The other underlying causes of CHAOS are tracheal atresia, laryngeal agenesis, subglottic stenosis or atresia, and laryngeal webs or cysts [5]. In our case the exact cause could not be detected as the post-mortem was refused, however it was suspected due to laryngeal atresia as the trachea was dilated.
The malformation is caused by non-development of the 6th branchial arch during normal embryological development. CHAOS spectrum is related to the deficient recanalization of the upper airways around the 10th week of gestation, which manifests later as airway obstruction. Due to the obstruction, normal fluid secretions in lungs cannot be cleared. The retained secretions will lead to lung parenchymal hyperplasia and massive distension of lung and results in cardiac and caval compression, which will develop heart failure, ascites, pleural effusion and hydrops in utero [6, 7]. Our case presented with distended lungs, dilated trachea and ascites. However, no e/o pleural effusion and hydrops was seen in our case. CHAOS can be associated with trisomy 9 and 16 [6]. No obvious gross features of trisomy seen in our case, however trisomy could not be totally ruled out as amniocentesis was not done.
The true incidence of CHAOS is unknown but is a very rare and life-threatening condition with high mortality and morbidity.

Prenatal diagnosis of CHAOS can be made with antenatal USG. Characteristic USG features in CHAOS are bilateral enlarged hyperechoic lungs, dilated trachea, compressed centrally displaced heart and flattened or inverted bilateral diaphragms. Ascites, pleural effusion and fluid collection in other body parts can also be seen [5]. MRI can confirm the diagnosis of CHAOS by identifying the level of obstruction and excluding extrinsic obstruction to airways as well [2].

The differential diagnosis of CHAOS includes other causes of hyperechoic lungs including bilateral congenital cystic adenomatoid malformation (CCAM) type III and sequestration. Other differential diagnosis can be different causes of external compression of airway including lymphatic malformation, vascular rings like double aortic arch and cervical teratoma [2, 5]. CHAOS may be linked to some genetic disorders like Fraser's syndrome, Cri-du-chat syndrome, short-rib polydactyly syndrome and velo-cardio-facial syndrome. CHAOS may be a part of TACRD (Tracheal agenesis, Complex cardiac anomalies, Radial ray defects and Duodenal atresia), which is a distinct entity from VACTERL (Vertebral, Anorectal, Cardiac, Tracheo-Esophageal, Renal, Limb anomalies) as VACTERL is associated with trachea-esophageal fistula rather than tracheal atresia [2].

A multidisciplinary team of obstetrician, neonatologists, laryngologists, anesthesiologist and pediatric surgeons is necessary for in-utero intervention in CHAOS [8]. Due to unavailability of sophisticated treatment facility in our setting and the worst prognosis of the condition, the patient party decided to terminate the fetus. Fetoscopic tracheoscopy with venting tracheostomy or creation of a pharyngotracheal fistula have been described, however there are reports which prove that chronic drainage of fetal lung fluid results in pulmonary hypoplasia in sheep [9, 10]. There are only few cases treated in utero and there are no uniform algorithms for CHAOS treatment. In the past, fetus with CHAOS has been at high risk of anoxia or death after conventional cesarean section or vaginal delivery due to time required to secure the airway. The EXIT (Ex Utero Intrapartum Treatment) procedure gives much time (even 8–66 minutes) to establish newborn airways while the uteroplacental circulation is preserved. Successful application of the EXIT procedure have been done in management of giant fetal neck masses, CCAM or unilateral pulmonary agenesis [9, 11]. Complete airway occlusion is a lethal malformation if not diagnosed prenatally and without appropriate intrapartum airway establishing procedure. Even if baby survives the EXIT, there are still a lot of problems to face. The consequences include capillary leak syndrome, respiratory distress syndrome, tracheobronchomalacia, diaphragmatic dysfunction. The newborn might be mechanical ventilation dependent for months and many complications including death can occur during that time. A laryngotracheal reconstruction is performed when the child is older. Most of the patients with complete larynx atresia are not able to speak even after reconstruction operations. Literature describes only a few cases of long-term survival with CHAOS [9, 12].

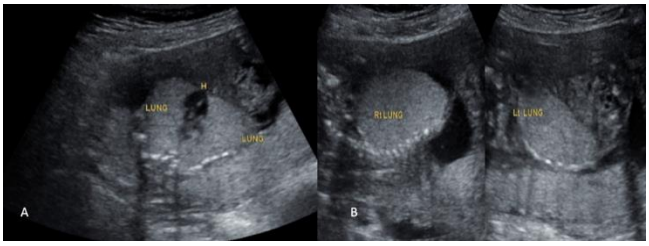

Fig 1: Ultrasound image of the fetus with congenital high airway obstruction syndrome at 18-19 weeks of gestation. (A) Enlarged and echogenic both lungs. (B). Echogenic lungs with compressed heart in between, the diaphragms are inverted.

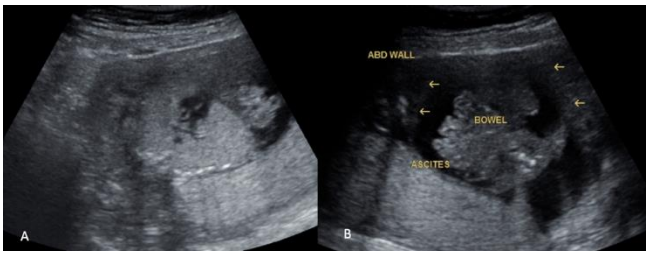

Fig 2: Ultrasound image of the fetus with congenital high airway obstruction syndrome at 18-19 weeks of gestation. (A). Dilated trachea behind the great vessels. (B). Ascites with free floating bowel loops.

5. Conclusions

Early diagnosis, detailed fetal assessment and an adequate postnatal intervention for establishing fetal airways are most important in management of CHAOS. Even though it is the rare condition and true incidence is unknown, prenatal diagnosis is possible with imaging. The teamwork of experienced CHAOS multidisciplinary team (obstetrician, neonatologist, laryngologist, pediatric surgeon and anesthesiologist) is crucial in these otherwise fatal cases.

6. References

1. Victor LL, Martinez CL, Seco DCC. Prenatal diagnosis of congenital high airway obstruction syndrome. Indian J Radiol Imaging [Internet]. 2018; 28(3):366-8. Available from: http://www.ijri.org/text.asp?2018/28/3/366/242176
2. Joshi P, Satija L, George R, Chatterjee S, D'Souza J, Raheem A. Congenital high airway obstruction syndrome-antenatal diagnosis of a rare case of airway obstruction using

multimodality imaging. Med journal, Armed Forces India [Internet]. 2012/01/18. 2012; 68(1):78-80. Available from: https://pubmed.ncbi.nlm.nih.gov/24669041

3. Aslan H, Ekiz A, Acar DK, Aydiner B, Kaya B, Sezer S, *et al*. Prenatal diagnosis of congenital high airway obstruction syndrome (CHAOS). Five case report. Med Ultrason [Internet]. 2015; 17(1):115-118. Available from: https://doi.org/10.11152/mu.2013.2066.171.haek
4. Mong A, Johnson A, Kramer S, Coleman B, Hedrick H, Kreiger P, *et al*. Congenital high airway obstruction syndrome: MR/US findings, effect on management, and outcome. Pediatr Radiol. 2008; 38:1171-9.
5. D'Eufemia MD, Cianci S, Di Meglio F, Di Meglio L, Di Meglio L, Vitale SG, *et al*. Congenital high airway obstruction syndrome (CHAOS): discussing the role and limits of prenatal diagnosis starting from a single-center case series. J Prenat Med [Internet]. 2016; 10(1–2):4-7. Available from: https://pubmed.ncbi.nlm.nih.gov/28725339
6. Garg MK. Case report: Antenatal diagnosis of congenital high airway obstruction syndrome - laryngeal atresia. Indian J Radiol Imaging [Internet]. 2008; 18(4):350-1. Available from: https://pubmed.ncbi.nlm.nih.gov/19774196
7. Artunc Ulkumen B, Pala HG, Nese N, Tarhan S, Baytur Y. Prenatal diagnosis of congenital high airway obstruction syndrome: report of two cases and brief review of the literature. Case Rep Obstet Gynecol [Internet]. 2013/10/22. 2013; 2013:728974. Available from: https://pubmed.ncbi.nlm.nih.gov/24251054
8. Adzick NS, Harrison MR. Fetal surgical therapy. Lancet. 1994; 343(8902):897-901.
9. Hamid-Sowinska A, Ropacka-Lesiak M, Breborowicz G. Congenital high airway obstruction syndrome. Neuro Endocrinol Lett. 2011; 32:623-6.
10. Lanman JT, Schaffer A, Herod L, Ogawa Y, Castellanos R. Distensibility of the Fetal Lung with Fluid in Sheep. Pediatr Res [Internet]. 1971; 5(11):586-90. Available from: https://doi.org/10.1203/00006450-197111000-00002
11. Bouchard S, Johnson MP, Flake AW, Howell LJ, Myers LB, Adzick NS, *et al*. The EXIT procedure: Experience and outcome in 31 cases. J Pediatr Surg [Internet]. 2002; 37(3):418-26. Available from: https://doi.org/10.1053/jpsu.2002.30839
12. Lim F-Y, Crombleholme TM, Hedrick HL, Flake AW, Johnson MP, Howell LJ, *et al*. Congenital high airway obstruction syndrome: natural history and management. J Pediatr Surg [Internet]. 2003; 38(6):940-5. Available from: https://doi.org/10.1016/S0022-3468(03)00128-3