International Journal of Contemporary Pediatrics
Khan R. Int J Contemp Pediatr. 2024 Apr;11(4):485-489
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20240740

Case Report

The silent storm: a case report of missed antenatal diagnosis leading to fatal respiratory distress and death in a rare case of congenital high airway obstruction syndrome

Roohi Khan*

Department of Pediatrics, Fortis Flt. Lt. Rajan Dhall Hospital, Vasant Kunj, New Delhi, India

Received: 12 February 2024
Revised: 07 March 2024
Accepted: 14 March 2024

***Correspondence:**
Dr. Roohi Khan,
E-mail: doctorroohi@yahoo.com

ABSTRACT

Congenital high airway obstruction syndrome in neonates is a rare, life-threatening condition characterized by upper airway obstruction. Typically diagnosed prenatally through advanced imaging, CHAOS results from anomalies such as laryngeal atresia or web or tracheal stenosis. Neonates with CHAOS face respiratory distress at birth, necessitating prompt intervention. Management often involves ex-utero intrapartum treatment procedure or tracheostomy to establish a secure airway. Timely diagnosis and multidisciplinary collaboration are crucial for optimizing outcomes in affected infants. Despite its rarity, CHAOS demands heightened clinical awareness to ensure swift, tailored interventions and improve the chances of neonatal survival. We report here a case of CHAOS which was not diagnosed on antenatal ultrasonography.

Keywords: CHAOS, Antenatal diagnosis, Upper airway obstruction

INTRODUCTION

Congenital high airway obstruction syndrome (CHAOS) is characterized by complete or partial blockage of the fetal upper airways. The recognition of this clinical condition dates back to the late 1900s when Hedrick first highlighted it.[1] The true incidence of CHAOS remains unknown. Failure to identify the syndrome during the prenatal period often leads to stillbirth or early post-delivery mortality.[2] Fortunately, contemporary advancements in prenatal imaging contribute to increase in utero recognition. Common sonographic indicators include bilaterally enlarged hyperechoic lungs, dilated airways, and altered diaphragm shape. Fetal ascites and non-immune hydrops may also accompany this condition.[3] The emerging management strategies underscore the growing significance of prenatal identification, offering prospects for enhanced neonatal outcomes.[4] Here, we report a case of congenital high airway obstruction syndrome due to laryngeal web which was not diagnosed antenatally. We also review the relevant literature in brief.

CASE REPORT

A 36-year-old female (G2P1L0A1), IVF conception, PIH on labetalol, Hypothyroidism on thyronorm, antenatal Scan done at 18weeks showed SLIUF AFI 16 Doppler normal. Another scan done at 30 weeks showed cardiac fetal echogenic speck and no other abnormality. At 37±4 weeks, she presented in our hospital, and was taken for elective LSCS in view of breech presentation. Male infant delivered by Elective LSCS breech presentation, but had difficult extraction, forceps assisted, born limp

and cyanosed, after initial steps of resuscitation as per NRP, PPV given for 60 sec HR increased color improved but had labored breathing. APGAR at 1 and 5 mins was 3 and 8 respectively. Infant was shifted to NICU on neo puff in view of respiratory distress and labored breathing. Postnatal Infant had weak cry with muffled sound, labored breathing and stridor.

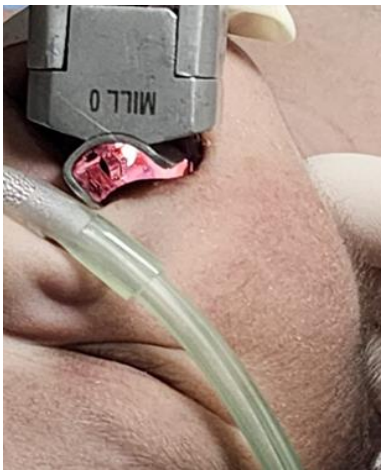

Figure 1: Direct laryngoscopy view of vocal cord during intubation showing some web/ band like membrane difficult to pass ET tube.

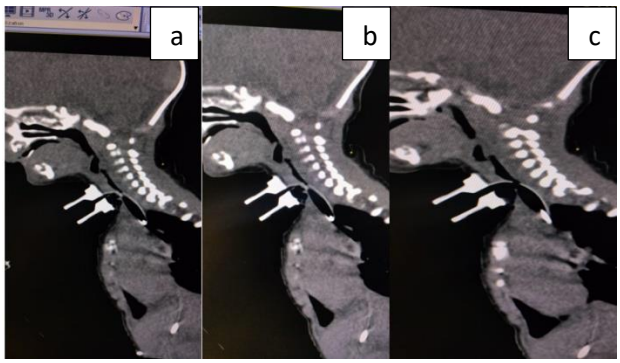

Figure 2: (a) Sagittal reconstruction of the neck showing tracheostomy tube; (b) sagittal reconstruction image shows thin web in trachea travesing anteroposteriorly at the level of C3 vertebra; (c) sagittal reconstructed image show patent tracheal lumen above and below the web with tracheostomy tube noted inferiorly.

ENT review was taken immediately in view of stridor, abnormal voice, they diagnosed laryngomalacia and advised no active intervention required. In view of respiratory distress, infant was Put on NIV with FiO2 60%. Within 1 hour of birth infant had seizures for which was loaded with injection phenobarbitone. The respiratory distress kept increasing and did not settle on NIPPV, hence in view of increasing and worsening distress intubation tried but was unsuccessful and difficult, even smallest ETT failed, hence anticipated some congenital high airway obstruction and as per protocol call for help was done and anaesthetist and ENT/pediatric surgeon were called.

Anaesthetist help was asked but still intubation was failed due to obstruction visualized between vocal cord. ENT consultation was sought in view of worsening distress and need for tracheostomy, till then infant was put LMA, bag and LMA and if required bag and mask as per need with 100% FiO2 was required intermittently. ENT review was taken immediately in view of stridor, abnormal voice, they diagnosed laryngomalacia and advised no active intervention required. In view of respiratory distress, infant was Put on NIV with FiO2 60%.

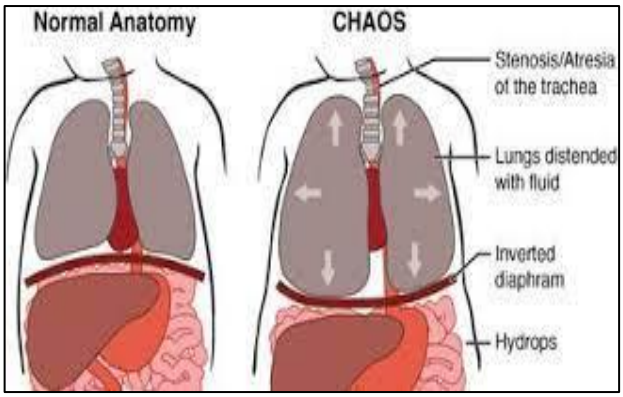

Figure 3: Schematic representation of normal and CHAOS anatomy.[15]

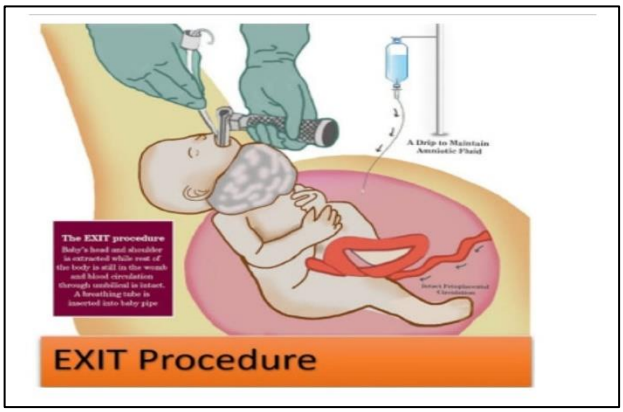

Figure 4: EXIT procedure.[14]

Within 1 hour of birth infant had seizures for which was loaded with injection phenobarbitone. The respiratory distress kept increasing and did not settle on NIPPV, hence in view of increasing and worsening distress intubation tried but was unsuccessful and difficult, even smallest ETT failed, hence anticipated some congenital high airway obstruction and as per protocol call for help was done and anaesthetist and ENT/pediatric surgeon were called. Anaesthetist help was asked but still intubation was failed due to obstruction visualized between vocal cord. ENT consultation was sought in view of worsening distress and need for tracheostomy, till then infant was put LMA, bag and LMA and if required

bag and mask as per need with 100% FiO2 was required intermittently.

Around 2 hours of life, infant had sudden bradycardia and cardiac arrest twice revived after CPR and injection adrenaline. Post resuscitation started on inotropes and fluids. Continues bag and mask given. Immediately pediatric surgeon (as ENT surgeon was not available at that time, nor video laryngoscope or neonatal bronchoscope was also not available) was called for emergency tracheostomy and tracheostomy was done by surgeon, infant was put on invasive ventilator on SIPPV mode. After initial stabilization CECT was done and MRI was planned. CECT with 3D reconstruction showed thick membrane like stricture of thickness 1.2 mm seen 10 mm below level of epiglottis causing complete stenosis of lumen s/o web, Area of consolidation with multiple air bronchogram and ground glassing is seen in basal segment of both lobes with bilateral mild pleural effusion. Severe reflux of contrast seen in right hepatic vein and Subsegmental venosus channels opacifying the right lobe of liver.

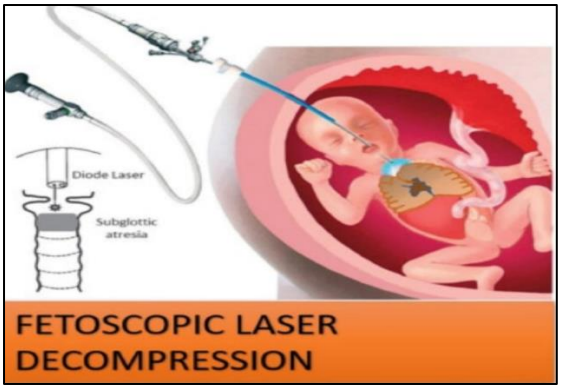

Figure 5: Fetoscopic Laser decompression.[14]

ECHO: Small ASD 6 mm left to right, PDA 2.2 mm left to right, Normal ventricular function. USG whole abdomen showed Gall bladder wall thickened, Minimal free fluid in pelvis, Portal triad appears prominent in both lobes. MRI was refused by parents. Infant was very sick on ventilation, on multiple inotropes dopamine, dobutamine and nor epinephrine, IV antibiotics like Meropenem, at 12 hours of life infant had multiple episodes of seizures which was not aborted with levetiracetam and phenobarbital, hence Midazolam infusion started. Also, infant didn't pass urine for which urinary catheter was inserted which showed meconium suggestive of vesicorectal fistula, also infant had hypospadias.

In view of multiple congenital anomalies and poor neurological outcomes, future surgeries required was discussed with parents right in the beginning itself. In hospital stay with very sick condition, parents opted no escalation hence only palliative treatment and supportive symptomatic treatment continued in the form of ventilation, fluids, inotropes, seizure medications. Infant again went in cardiac arrest at around 42 hours of life and could not be revived and was declared dead. Diagnosis: Term 37+4/ AGA/male/multiple congenital anomalies/ congenital high airway obstruction syndrome with laryngeal web/ neonatal seizure/encephalopathy/vesico-rectal fistula/ASD/PDA.

DISCUSSION

CHAOS is an exceedingly rare condition with a grim prognosis, where the proximal airway faces complete or partial obstruction. While the true incidence remains unknown, emerging evidence suggests it might be more prevalent than previously believed.[1,2,5,6] The medical literature has documented over 100 case reports and several case series detailing CHAOS cases.[7] This condition is typically fatal if not addressed, as fetal lung fluid outflow obstruction triggers pulmonary hyperplasia. Prenatal ultrasound examinations reveal distinct characteristics, including symmetrically enlarged and highly echogenic lungs, a dilated and fluid-filled trachea, and bronchi, along with inverted hemidiaphragms. The compressed lungs often lead to a small heart, centrally positioned within the thorax. Fetal ascites and occasional hydrops are common findings.[3,5,6,8-11]

CHAOS is a rare and often fatal syndrome, with reported associations with certain genetic syndromes and chromosome aberrations, although the exact cause remains unidentified. Antenatal sonographic examinations reveal distinct patterns, facilitating early diagnosis, a crucial factor if fetal intervention is being contemplated. The only potential lifeline for an affected fetus appears to be the ex-utero intrapartum treatment (EXIT). The primary aim of this procedure is to establish a functional airway for the infant before terminating the fetomaternal circulation, enhancing the chances of survival.

In instances of unidentified syndromes during the prenatal phase, it commonly leads to stillbirth or death shortly post-delivery. The overall prognosis remains grim, and survivors face significant medical and surgical challenges even after undergoing fetal intervention. Enhancing neonatal survival is achieved through the implementation of a carefully orchestrated EXIT procedure during a controlled near-term caesarean section.[2,12,13] As a result, the significance of prenatal diagnosis becomes crucial in such scenarios.

If a syndrome goes unnoticed during the prenatal period, it commonly leads to stillbirth or post-delivery fatality. The prognosis is generally grim, and survivors often face substantial and enduring medical and surgical difficulties even after successfully navigating fetal intervention. Without interference, CHAOS carries a 100% perinatal mortality rate. Although chances of survival may increase with treatments like fetoscopic procedures and EXIT, CHAOS is associated with elevated morbidity and

mortality. In a case series by Maria et al ultrasound diagnoses were conducted between 13 and 22 weeks of gestation in six reported cases.[16]

Five pregnancies were terminated at the parent's request, and one infant born at 8 months via caesarean section for placenta previa died shortly after birth. Amniocentesis in two cases revealed aneuploidy karyotype (18 trisomy). Another study by Lee et al reported on 13 prenatally diagnosed CHAOS cases, with 7 lost to follow-up and 6 postnatally confirmed.[17] Except for one, all fetuses were delivered via caesarean section with an EXIT procedure.

Two patients had concurrent congenital heart diseases requiring multiple cardiac surgeries, leading to developmental delay. The remaining 4 showed normal development, except for expressive language. Two infants succumbed to respiratory complications, while the other 4 survived. All 4 live patients underwent tracheostomy with planned reconstruction surgery, and three children are now capable of phonation.

Key takeaways

Key takeaways include the need for meticulous observation of deviations from normal fetal anatomy to comprehend the pathophysiology of the disease. High-quality imaging, capturing sagittal, parasagittal, and sectional fetal anatomy, significantly enhances diagnostic accuracy. The multidisciplinary approach emphasizes the importance of a well-supported team comprising fetal surgeons, pediatric and ENT surgeons, and neonatologists. Furthermore, it underscores the necessity for appropriately sized neonatal instruments, such as neonatal video laryngoscopes, tracheostomy tubes, and bronchoscopes, acknowledging potential challenges in resource-limited settings.

CONCLUSION

In conclusion, the timely prenatal identification is pivotal not only for the survival of the fetus but also for strategic planning of EXIT procedures. It is essential to adopt a multidisciplinary approach, involving ENT and pediatric surgeons at an early stage. This proactive collaboration aims to anticipate challenges in airway management, necessitating the presence of backup ENT and pediatric surgeons for emergency tracheostomy. Additionally, the preparation should include the availability of neonatal video laryngoscopes, tracheostomy tubes, and bronchoscopes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Note required

REFERENCES

1. Roybal JL, Liechty KW, Hedrick HL, Bebbington MW, Johnson MP, Coleman BG, et al. Predicting the severity of congenital high airway obstruction syndrome. J Pediatr Surg. 2010;45:1633-9.
2. Joshi P, Satija L, George R, Chatterjee S, D'Souza J, Raheem A, et al. Congenital high airway obstruction syndrome- antenatal diagnosis of a rare case of airway obstruction using multimodality imaging. Med J Armed Forces India. 2012;68:78-80.
3. Garg MK. Case report: antenatal diagnosis of congenital high airway obstruction syndrome—laryngeal atresia. Indian J Radiol Imaging. 2008;18:350-1.
4. 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;40:1800-5.
5. Mudaliyar US, Sreedhar A. Chaos syndrome. BJR| Case Rep. 2017;3(3):23-9.
6. Kuwashima S, Kitajima K, Kaji Y, Watanabe H, Watabe Y, Suzumura H. MR imaging appearance of laryngeal atresia (congenital high airway obstruction syndrome): unique course in a fetus. Pediatr Radiol. 2008;38(3):344-7.
7. Kanamori Y, Abdelrahman AM, Senna A, Alsaad F. Congenital High Airway Obstruction Syndrome (CHAOS): No Intervention, No Survival-A Case Report and Literature Review. Case Rep Radiol. 2020;2020:1036073.
8. Ekmekci E, Gencdal S, Kiziltug N. Prenatal ultrasonography findings of fetus with congenital high airway obstruction (chaos): a case report and review of literature. Clin Obstet Gynecol Reprod Med. 2017;3(5):21-9.
9. Arthurs OJ, Chitty LS, Judge-Kronis L, Sebire NJ. Postmortem magnetic resonance appearances of congenital high airway obstruction syndrome. Pediatr Radiol. 2015;45(4):556-61.
10. Artunc UB, 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. 2013;2013:728974.
11. Mong A, Johnson AM, Kramer SS, Coleman BG, Hedrick HL, Kreiger P, et al. Congenital high airway obstruction syndrome: MR/US findings, effect on management, and outcome. Pediatr Radiol. 2008;38(11):1171-9.
12. Lim FY, Crombleholme TM, Hedrick HL, Flake AW, Johnson MP, Howell LJ, et al. Congenital high airway obstruction syndrome: natural history and management. J Pediatr Surg. 2003;38(6):940-5.
13. Guimaraes CV, Linam LE, Kline-Fath BM, Donnelly LF, Calvo-Garcia MA, Rubio EI, et al. Prenatal MRI findings of fetuses with congenital high airway obstruction sequence. Korean J Radiol. 2009;10(2):129-34.
14. Pathare S, Varghese K, Limbad K. A rare case: Antenatally diagnosed congenital high airway obstruction syndrome. Radiol Case Rep. 2021;16(9):2668-71.

15. CHAOS. Available at: https://fetus.ucsf.edu/chaos/. Accessed on 20 November 2023.
16. 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. 2016;10(1-2):4-7.
17. Jeong SH, Lee MY, Kang OJ, Kim R, Chung JH, Won HS, et al. Perinatal outcome of fetuses with congenital high airway obstruction syndrome: a single-center experience. Obstet Gynecol Sci. 2021;64(1):52-61.

Cite this article as: Khan R. The silent storm: a case report of missed antenatal diagnosis leading to fatal respiratory distress and death in a rare case of congenital high airway obstruction syndrome. Int J Contemp Pediatr 2024;11:485-9.