 DOI: 10.18231/2455-6793.2017.0029

Congenital High Airway Obstruction Syndrome (CHAOS)

Mohit Goyal[1,*], Sunita Sattavan[2], Nandini Sharma[3], Sonal Gupta[4]

[1,2,3,4]Resident, Dept. of Pediatrics, MG Medical College & Hospital, Jaipur, Rajasthan

***Corresponding Author:**
Email: mohitgoyal008@gmail.com

Abstract

This is a case report of preterm male child weighing 1.9 Kg, APGAR 2/10 -3/10 with tracheal stenosis, web or agenesis. Mother was taking phenytoin and on ANC showing oligohydramnios. It was done in 2nd trimester showing age 21 weeks 2 days with congenital high airway obstruction syndrome with enlarged hypoechoic lungs, compressed tubular heart and massive ascitis. This syndrome is associated with a poor prognosis. Baby was delivered via NVD with signs suggestive of fetal hydrops. Inspite of best efforts baby could not be saved.

Introduction

CHAOS or congenital high airway obstruction syndrome is an extremely rare clinical entity defined as enlarged echogenic lungs, flattened and inverted diaphragm, dilated airway, fetal ascitis, hydrops on antenatal scan.(1)

CHAOS is a blockage of the fetus's trachea or larynx due to a number of factors including narrowing of the airway, a web-like membrane or even tracheal atresia. In the uterus, the fetal lungs constantly produce fluid and as a result of this airway blockage in the trachea, the lung fluid cannot escape out of the fetal mouth. Because of this the fetus's lungs become distended with fluid and over distended lungs can put pressure on the heart and affect the heart's ability to function. If the heart cannot beat effectively hydrops or congestive heart failure can occur.It is associated with poor prognosis and long term outcome. Now advances in prenatal imaging, early in utero diagnosis, in utero surgery and delivery with EXIT to tracheostomy has improved outcome.(2)

There have been 2 descriptions of airway reconstruction procedure for complete laryngeal atresia causing CHAOS in literature.(3)

Case Report

A 27 year old female with 2nd trimester antenatal scan showing a 21 +2 week fetus with congenital high airway obstruction syndrome with oligohydramnios. The family was adviced to undergo MRI for other associated congenital anomalies. Due to unaffordability and the poor prognosis associated with this syndrome the family was unwilling to undergo any expensive investigations. The mother went into labour at 30 weeks delivering a PT male baby weighing 1.9 kg, with massive ascitis via NVD. Apgar 2/10 followed by 3/10 at 1 and 5 minutes respectively. Tracheotomy was planned after failure to intubate but the baby landed up in cardiac arrest and could not be saved.

Discussion

CHAOS is likely more common than reported, as many fetuses with CHAOS die in utero or are stillborn.

Currently, all viable patients with CHAOS are offered an EXIT delivery followed by surgical reconstruction surgery later on after 2 years of age.(2)

Outcomes may depend on other anomalies, as fetuses with CHAOS frequently have additional congenital abnormalities diagnosed postnatally.

In our case the mother ideally should have undergone MRI in 2nd trimester, followed by an EXIT procedure during delivery.

Conclusion

Early diagnosis, detailed fetal assessment and an adequate postnatal intervention for establishing the fetal airway are prerequisites for survival in CHAOS patients.

However due to limited resources and inadequate antenatal scans in our setup this is usually not possible.

References

1. Hedrick MH, Ferro MM, Filly RA, Flake AW, Harrison MR, Adzick NS. Congenital high airway obstruction syndrome (CHAOS): a potential for perinatal intervention. *J Pediatr Surg*. 1994;29(2):271-274.

2. Rahman T, Khalequezzaman S, Ahsan S, Alam J, Sarkar MK. A case report of CHAOS caused by complete laryngeal obstruction. *Pulse Vol*.2014;7:38-41.
3. Hartnick CJ, Rutter M, Lang F, Willing JP, Cotton RT. Congenital high airway obstruction syndrome and airway reconstruction: an evolving paradigm. *Arch Otolaryngol Head Neck Surg*. 2002;128(5):567-570.