Congenital High Airway Obstruction Syndrome (CHAOS): Peripartum Planning, Airway Management, and Resuscitative Measures

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Introduction

Congenital high airway obstruction syndrome (CHAOS) is a condition usually diagnosed in utero which is caused by complete or near-complete obstruction of the airway and ranges widely in clinical severity. This syndrome, usually characterized by tracheal agenesis in the absence of tracheoesophageal fistula (TEF), presents unique challenges not only for the anesthesiologist, but also for a multidisciplinary team usually including neonatologists and otolaryngologists. Obstruction upon delivery sets into motion events that can ultimately lead to fetal demise, but in certain circumstances airway decompression can lead to complete reversal of syndromic findings.

Case Description

A 44 year old G2P1 female presented at 24 weeks with imaging consistent with CHAOS. The EXIT procedure was strongly recommended but was declined by the family due to inconsistencies on later MRI imaging. The OB team planned for an induction of labor at 38 weeks with both pediatric anesthesia and airway teams on stand-by. Upon delivery, the neonate was taken immediately to the OR. Initial assessment by the NICU team revealed no spontaneous respiratory effort and a HR in the 70s despite stimulation. The neonate was handed to the Pediatric ENT/Anesthesia team. Mask ventilation was unsuccessful, and direct laryngoscopy revealed a pinpoint depression in the expected location of the vocal cords. CPR was initiated. ENT attempted an emergent tracheostomy, but the initial attempt was unsuccessful and caused significant blood loss. A rigid scope placed from above was unsuccessful. After obtaining hemostasis, a tracheostomy was successfully completed, and heart rate returned. The neonate received 30 minutes of cardiopulmonary resuscitation.

After volume resuscitation the profoundly acidic neonate was transferred to the NICU on an epinephrine infusion. Over the next 24 hours, the neonate had increasing ventilation and vasopressor requirements. The family elected to withdraw care due to the high likelihood of mortality and severe neurological impairment secondary to prolonged hypoxia.

Summary

CHAOS results from a rare, usually lethal defect with an incidence of approximately 1 per 50,000 newborns. The characteristic findings of enlarged hyperechogenic lungs, dilated trachea, compression of the heart, and flattened diaphragm allow for in utero diagnosis, but the presence of TEF masks these ultrasonographic findings by allowing lung fluid to pass through the fistula to the stomach or amniotic sac. While an EXIT procedure is the preferred management, this case illustrates an alternative approach for parturients who decline. Associated malformations can include VACTERL association, and in this case postmortem analysis revealed a constellation of abnormalities perhaps consistent with Fraser syndrome, including ambiguous genitalia, hydrocolpos, unicorne uterus, partial syndactyly, and bi-lobed right lung. This case highlights the significant challenges that providers face upon the delivery of a neonate with CHAOS and illustrates that poor outcomes can inevitably result despite the preparedness of a multidisciplinary team.

References:

Figures 2 & 3: Postmortem pathology reveals severe cricoid stenosis and tracheostomy site.