Congenital High Airway Obstruction Syndrome (CHAOS)

Cynthia Hayes PGY-3
Genesys Regional Medical Center
Objectives

• Define what is CHAOS.
• Most common cause of CHAOS.
• Diagnosis of CHAOS.
• Treatment options of CHAOS.
• Review two cases of patients with CHAOS.
CHAOS

• First described by Dr. Rossi in 1826, but not named until 1994.
• Incidence equal in males to females.
• Some reports of an autosomal dominance inheritance pattern, but reported in only four cases.
CHOAS

• Intrauterine obstruction above the lungs that creates a closed system.
• Results in accumulation of fetal lung fluid leading to hyperinflation of the developing lungs with flattening of the diaphragm.
CHAOS

- Laryngeal atresia (Most common cause)
- Laryngeal stenosis
- Subglottic stenosis
- Tracheal aplasia and/or stenosis
CHOAS

• Diagnosis
  • Diagnosis can be possible as early as 15 weeks of gestation.
  • Prenatal ultrasound (US)
    • Large echogenic lungs
    • Flattened or inverted diaphragms
    • Dilated distal airways
    • Fetal ascites or hydrops
  • If fetus has a tracheoesophageal fistula the characteristic findings on US may be absent.
CHAOS

- Echogenic lungs
- Ascites

Image courtesy of Chaemsaithong 2012.
• inverted diaphragm.

Image courtesy of Chaemsaithong 2012
More detailed imaging recommended through magnetic resonance imaging (MRI) as up to 55.2% fetuses have additional anomalies.

- Tetralogy of Fallot
- Pulmonary atresia
• Treatment
  • Intrapartum intervention (Payam et al. 2012)
    • Decompress upper airway to allow more functional airway development.
    • Passage of wire through obstruction.
    • All fetuses required further intervention.
CHAOS

• Treatment continued.
  • Ex utero intrapartum therapy (EXIT) procedure at the time of delivery to secure an airway through tracheostomy placement.
  • Allows time to further delineate the anatomy and stabilize patient for reconstruction.
CHAOS
Treatment-EXIT

Image courtesy of Mychaliska 1997.
CHAOS

- Reconstruction
  - Twelve patients reconstruction at 17 months of age or older.
  - Majority of the patients were tracheostomy dependent.
CHAOS

• Case #1
  • Prenatal US at 24 weeks of gestation
    • Enlarged echogenic lungs.
    • Flattening of diaphragm.
    • Mild ascites
  • Amniocentesis revealed a normal karyotype.
CHAOS

• Case #1 continued.
  • Delivered by Cesarean section at 28 weeks with an EXIT procedure performed to secure an airway.
• At one week of age a direct laryngoscopy and bronchoscopy was performed.
  • Blind pouch in subglottic region.
• Case #1 continued.
  • Reconstruction
    • Currently patient is 15 months old and still requiring ventilator support due to mild tracheobronchomalacia.
    • Once weaned from the ventilator, laryngeal reconstruction will be performed.
Case #2

- Prenatal US at 20 weeks of gestation
  - Hyperinflated echogenic lungs.
  - Flattened diaphragm.
  - Dilated distal trachea.
  - Bilateral clubbed feet.
- Amniocentesis revealed a normal karyotype.
Case #2 continued.

Delivered at 31 weeks via Cesarean section with an EXIT procedure at the University of Michigan.

At two weeks of age a direct laryngoscopy and bronchoscopy was performed.

Laryngeal atresia.
Image courtesy of Dr. Green at the University of Michigan.
CHAOS

• Case #2 continued.
  • Reconstruction
    • At 3 months of age.
    • Arytenoids were divided using a CO2 laser.
    • Resection of anterior cricoid and rudimentary thyroid cartilage.
    • Trachea was advanced superiorly to laryngeal notch.
    • A 7mm Montgomery stent was placed and removed one week post operatively.
• Case #2 continued.
  • Post reconstruction follow-up.
    • Direct laryngoscopy the CO2 laser was used for excision of granulation tissue.
    • Spontaneous movement of right true vocal cord.
  • At six months of age still requiring ventilator due to tracheobronchomalacia.
CHAOS
Case #2

Image courtesy of Dr. Green at the University of Michigan.
CHAOS

- Other things to consider
  - Aspiration
    - Behavior modifications
    - Diet modifications
  - Voice outcomes
References

Thank you.

Any questions.