Western States Case Conference

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Outline

• Case Presentation
• Diagnosis
• Management
• Summary
• Teaching Points
Case Presentation

• 4 month old Caucasian female: 36 week GA, h/o cyanotic congenital heart disease with multiple associated medical anomalies who has developed nasal congestion, cough and supplemental oxygen requirement.

• Initiated Xopenex every 4 hours as needed with benefit.

• Mother of the patient had recently been ill with a URI.

• 3 weeks prior to the consultation:
  – Brief intubation during cardiac catheterization procedure
  – ET aspirate positive for Stenotrophomas maltophilia and Serratia marcescens.
  – Treated with 7-day course of Bactrim; negative nasopharyngeal culture obtained 4 days prior.
Case Presentation

BIRTH HISTORY:
• Former 36 week GA, prenatally diagnosed with congenital heart disease. Her mother had routine prenatal care.
• The pregnancy was complicated by chronic hypertension, treated with labetalol and magnesium as well as type 2 diabetes requiring insulin therapy. The infant was noted to have a 2-vessel cord on prenatal ultrasound.

REVIEW OF SYSTEMS:
• A 14-point review of systems was obtained and otherwise noncontributory aside from what is noted above.
Case Presentation

PAST MEDICAL AND SURGICAL HISTORY:

• TOF with PA, MAPCAs, left hemitruncus of the left pulmonary artery arising from the aorta and PAPVR.
• Status post bronchoscopy on DOL#1 by ENT to assess for presence of tracheobronchial abnormalities.
• Status post aorticopulmonary shunt to the right pulmonary artery and banding of the left pulmonary artery on DOL#10.
• Ligation and division of a right ventricular outflow tract to the main pulmonary artery on DOL#10.
• History of right diaphragm paralysis, s/p diaphragm plication on DOL#42.
• Evaluated by Genetics, normal FISH and chromosomes.
Case Presentation

DEVELOPMENTAL HISTORY:
• Developmental delay, normal brain MRI. Followed by PT and OT who suggest that she may need a G-tube, but the family is against this at this time.

FEEDS: Elecare 28kcal/oz; 22ml/hr via NGT.

IMMUNIZATIONS: Up to date.

FAMILY HISTORY: Noncontributory.

SOCIAL HISTORY: Parents are united in marriage and actively involved during this care.

MEDICATIONS:
• Diuril 80 mg per NG tube q.12 h.
• Xopenex 0.63 mg/3 mL inhaled neb q.4 h.
• Prilosec 3.2 mg/1.6 mL per NG tube daily.
Physical Examination

MEASUREMENTS: Wt 4.28 kg (> than 2 SD below the 3%) and Ht is 55.5 cm (2%).

VITAL SIGNS: T 36.4, HR 132, RR 33, BP 68/31; O2 sat 79% on 2 L NC with FIO2 of 28%.

GENERAL: Small for age, 4-month-old female in no acute distress.

HEENT: NCAT, nares patent, NG tube and NC is in place. Trachea midline.

RESPIRATORY: Well-healed sternotomy scars. Good aeration throughout. Decreased BS in RUL compared to LUL, otherwise CTA. There is no stridor, nasal flaring, grunting, tachypnea or increased work of breathing.

CARDIOVASCULAR: Normal precordial activity, tachycardia, no murmurs, rubs or gallops. Capillary refill less than 2 seconds. Brachial pulses palpable and equal bilaterally.

EXTREMITIES: No clubbing or cyanosis.
Echocardiogram

- Large membranous to outlet VSD.
- Mildly dilated right ventricle.
- TOF with PA and MAPCAs.
- Normal LV systolic function.
- Mild right ventricular hypertrophy.
- Left to right shunting at the atrial level.
- The right atrium is moderately dilated.
CXR obtained on the day of our consultation
Upon further review of her imaging studies...
3D Reconstructed images of the airway
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And the diagnosis is....

Congenital Tracheal Stenosis with a Bifid Trachea!
Congenital Tracheal Stenosis

- Characterized by structural tracheal constriction.
- Associated with pulmonary, cardiovascular and gastrointestinal malformations.
- Incidence ~ 1 in 64,500.
- Represents ~ 1% of laryngotratheal stenosis.
- Prior to the advent of current surgical techniques, mortality was reported to be as high as 79%.
Development of the Tracheoesophageal Septum

A: Foregut
   Ventral diverticulum
   Esophagus

B: Tracheoesophageal septum
   Lung buds
   Distal esophagus
   Carina

C: Esophageal atresia
   Distal tracheoesophageal fistula

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Embryologic Aortic Arch Complex

- Ventral aorta
- Dorsal aorta
- Internal carotid
- External carotid
- Pulmonary arteries
- Ductus arteriosus

Normal heart
- Esophagus
- Trachea

Double aortic arch
- Aorta

AP View
- Rt. Aortic Arch
- Lt. Aortic Arch
- Esophagus

Lateral View
- Heart

Double Aortic Arch Barium Swallow - shows impingement of the vascular malformation upon the esophagus both posteriorly and bilaterally. The impingement upon the trachea as well can result in tracheomalacia.
Tracheal Anatomy and Embryology

- The tracheal consists of a fibromuscular sheath supported by approximately 15 to 20 C-shaped cartilaginous rings and the trachealis muscle.
- Average diameter of the full-term newborn trachea is 6mm.
- The inner lumen is lined with pseudostratified ciliated columnar epithelium.
- Vital for the mucociliary transport that prevents mucus obstruction.

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Clinical Symptomatology

- Classically presents with biphasic stridor.
- Brassy, nonproductive cough, nasal flaring, wheezing, intercostal retractions, intermittent cyanosis, acute life threatening respiratory failure.
- Difficult intubation, failure to extubate or trouble with ventilation.
- Symptoms may not become apparent until ~50% stenosis, dyspnea at rest is likely to present at 75% stenosis.

Ho et al, 2008
Clinical Symptomatology

• Presentation may be delayed until the infant develops an acute respiratory infection that exacerbates the narrowed lumen.
Diagnosis

- Clinical suspicion.
- Initial priority must be to secure the airway.
- Exclude other causes of acute respiratory distress.
- Imaging modality.
- Bronchoscopy remains the most reliable method of diagnosing tracheal stenosis.
Imaging

Ho et al., 2008
Imaging

Berrocal T et al. Radiographics 2004;24:e17-e17
CXR obtained on DOL#1
Classification schemes

- **Tracheal Stenosis**
  - Congenital
  - Acquired

- **Compression**
  - Intrinsic
  - Extrinsic

- **Segment Size**
  - Short
  - Long
Classification Schemes
Medical Management

• Conservative management.
  – Observation is a safe, viable approach for clinically mild tracheal stenosis to determine if an operation will be needed eventually.
  – One longitudinal study found that stenotic tracheas naturally display catch up growth.
  – Treatment includes antireflux treatment, antibiotics, chest physiotherapy, and humidified air.

• Endoscopic treatments include laser excision, balloon dilation and stent placement.

Interventional Strategies

Ho et al, 2008
Interventional Strategies

Ho et al, 2008
Surgical Management: Resection and Anastomosis
Surgical Management:
Slide Tracheoplasty

Ho et al, 2008
Surgical Management:
Pericardial Patch Tracheoplasty
Getting back to our patient…

• Tracheoplasty of the bifid trachea with a patent bronchus.
• Ligation and division of the MAPCAs.
• Central patch augmentation of the left and right branch pulmonary arteries.
CXR on POD#1
Case Presentation

• Unremarkable post-operative course.
• Discharged home on room air without evidence of respiratory distress on POD #10.
• Close follow up with Pulmonology and ENT.
CXR 4 ½ months after her tracheoplasty
Summary

• Congenital tracheal stenosis is associated with pulmonary, cardiovascular, and gastrointestinal malformations.
  – Tracheal bronchi are seen in up to 20% of cases of congenital tracheal stenosis.
  – Vascular malformations are seen in as many as 50% of all cases of tracheal stenosis.

• The management of congenital tracheal stenosis has dramatically improved the outcomes of affected patients in the past several decades.
Teaching points

• Associations with other malformations.
  – Know your embryology!
• Imaging pearls.
• Index of suspicion.
Questions? Further thoughts?
Thank you very much for participating!
References

8. micro.magnet.fsu.edu