AIRWAY ABNORMALITIES IN CONGENITAL HEART DISEASE

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CT vs MR for Assessing Airway Abnormalities

CT - better spatial resolution
- faster
- ionizing radiation

MR - no ionizing radiation
- longer study
- more difficult to evaluate airway and lung

CHD and Abnormalities of the Lower Airways

Congenital/Intrinsic
Obstruction
- Tracheal / bronchial atresia
- Tracheal rings
- Tracheobronchial web/stenosis
- Fistulae
  - Tracheoesophageal fistula (TEF)
  - Bronchobiliary fistula
- Branching anomalies
  - Tracheal bronchus
  - Bridging bronchus
  - Heterotaxy syndrome
  - Scimitar syndrome

Tracheal Agenesis

- Very rare
- Associated with other anomalies similar to the VACTERL spectrum
- Three types:
  Type 1 - Atresia proximally with normal distal segment and TEF
  Type 2 - Complete tracheal agenesis with normal carina and bronchi + TEF
  Type 3 - Complete agenesis, mainstem bronchi join esophagus separately

Reconstruction Techniques

Raw Images
MPR
MIP
Volume Rendering
VR Cine
Virtual Bronchoscopy

Airway Obstruction
Tracheal Agenesis, Type 3
VSD
Airway Obstruction
Tracheal Rings

- Complete cartilage rings
- Absent trachealis muscle
- Segmental narrowing
- Associated with
  - Tracheal bronchus
  - Pulmonary sling
  - Structural heart disease
  - H-type TEF

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Ring/Sling Complex
Tracheal rings/stenosis & LPA sling

Type 2 LPAS strongly associated with tracheal stenosis

49 yo F Prior TOF Repair
Rt diaphragmatic paralysis, decreased exercise tolerance

Pulmonary Sling
Tracheal rings/
stenosis, branching
anomalies
CT virtual bronchoscopy

Normal proximal trachea
Stenotic lower trachea

Pre & Post Surgical Repair

Ring/Sling with long tracheal stenosis,
T-carina, proximal bronchial stenoses & right bridging bronchus
Vertebral Anus
Cardiac anomalies
Tracheoesophageal fistula
Renal Limb

Branching anomalies
Heterotaxy, Asplenia
Horizontal liver, bilateral right sided bronchial branching

Branching Anomalies
Scimitar Syndrome
Rt PAPVR, small RPA, hypoplastic right lung, absent RUL bronchus

CHD and Abnormalities of the Lower Airways
Acquired/Secondary
- Extrinsic compression
  - Cardiac
  - Vascular
  - Post-cardiac surgery
- Intrinsic obstruction
  - Tracheo/bronchomalacia
  - Tracheal/bronchial stenosis
  - Foreign body
- Other
  - Tracheomegaly
  - Tracheobronchial calcification

Extrinsic Airway Compression
- Cardiac
- Vascular
- Aorta
- Innominate artery
- Pulmonary artery/branches
- Pulmonary Veins
- Surgical repair for
  - Truncus arteriosus
  - Tetralogy of Fallot
  - Pulmonary atresia
  - Interrupted aortic arch

Vascular Extrinsic Compression
2YO NOISY BREATHING
Tracheomalacia- innominate artery syndrome
Vascular Extrinsic Compression
Truncus Arteriosus
Enlarged aorta compressing trachea

Vascular Extrinsic Compression
14yo with repaired Truncus Arteriosus with Double Aortic Arch & RV/PA conduit. Bilateral SVC's. Mild airway compression

Vascular Extrinsic Compression
6mo with large PDA, cardiomegaly, enlarged PA's & RML CLE (Rudhe Syndrome)
- reversible post shunt closure

Vascular Extrinsic Compression
Large PDA, RML emphysema, bronchus compressed by enlarged RPA

Vascular Extrinsic Compression
3 NB infants
- Same underlying diagnosis
- Unilateral or bilateral perihilar mass and air trapping

Tetralogy of Fallot
Enlarged aorta compressing Lower Trachea + Tracheomalacia
TOF absent pulmonary valve leaflets
• Severe PR
• Aneurysmal central PA’s
• Airway compression

Vascular Extrinsic Compression
NB – TAPVR
• Venous confluence drains to SVC medially
• Compresses LMSB
• RMSB web

Cardiac Extrinsic Compression
5yo DORV, S/P Rastelli
• Prosthetic pulmonary Valve
• RV failure, cardiomegaly
• LMSB compressed between heart and descending aorta

Post-cardiac surgery
Airway narrowing
7yo TOF PAT
• S/P Unifocalization
• Trachea flattened by Ao arch
• Markedly compressed prox LMSB

7yo TOF PAT S/P Unifocalization
Minip
• Proximal LMSB almost obliterated

7mo TOF PAT
• Postop opacified RUL
• MAPCA clip obstructing RULB
SUMMARY

• There is a strong association between Congenital Heart Disease and a broad spectrum of primary and secondary airway abnormalities
• Recognition and detailed evaluation of intrinsic airway anomalies as well as extrinsic airway compression may greatly influence patient management
• CT is usually the best study for airway evaluation 2D and 3D image reconstructions allow optimal display of anatomic relationships