CONGENITAL ABNORMALITIES OF THE GREAT VESSELS

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INTRODUCTION

- Congenital abnormalities of the great vessels are very common
- Frequently identified in adults
- Many vascular anomalies exist – some incidental, others symptomatic
- MDCT is increasingly being used
- Protocols can be tailored specific scenario
CT PROTOCOL - AORTA

Technique
- Initial precontrast scan
- 100 ml injection
- 4-6 cc/sec
- Bolus triggering in desc Ao
- ECG-gating, retro vs prospective
- 1-2 mm slice collimation
- 50% overlap for 2D/3D reconstructions
- Iterative recon, often 100 kV
ADULT THORCIC CONGENITAL VASCULAR DISEASE

CLASSIFICATION

- Thoracic Venous Anomalies
- Thoracic Aortic Anomalies
- Pulmonary Artery Anomalies
THORACIC VENOUS DISEASE

- Congenital systemic venous anomalies
  - Left superior vena cava
  - Interrupted IVC (azygos continuation of IVC)

- Congenital anomalies of pulmonary venous return
  - Partial anomalous pulmonary venous return
  - (TAPVR) - infants
SYSTEMIC VENOUS ANOMALIES

- Cardinal Vein System
  - Anterior (pre) cardinals
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  - Common cardinals
SYSTEMIC VENOUS ANOMALIES

- Cardinal Vein System
  - Anterior (pre) cardinals
  - Common cardinals
  - Subcardinals
SYSTEMIC VENOUS ANOMALIES

- Duplicated (Left SVC)
  - L precardinal persists
  - Normal R precardinal involutes or is small
Left SVC (prevalence – 0.3%)
- MDCT - best visualized lateral to aortic arch
- absence of left brachiocephalic vein (65%)
- absence of right SVC (15%)
- dilated coronary sinus
PERSISTENT LEFT SVC
SYSTEMIC VENOUS ANOMALIES

Interrupted IVC (azygos continuation)
- intrahepatic IVC (subcardinal) interrupted
- flow diverted posteriorly to azygos vein, large arch
- MDCT – absence of suprarenal, intrahepatic IVC, large azygos
- hemiazygos continuation occasionally
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- associated with polysplenia
PULMONARY VENOUS RETURN

Partial APVR
- Partial failure of primitive veins to fuse with left atrium
- Right: left APVR = 2:1 overall
- In adults, left APVR is more common
- Isolated or associated with CHD (especially ASD)
Scimitar Syndrome

- Also, venolobar, hypogenetic lung syndrome
- Involves RLL pulmonary vein and variable RUL/RML vein
- Typically drains to IVC or right atrium
RIGHT PAPVR

RUL PAPVR

- One or several veins drain into the lower SVC
- Strong association with sinus venosus ASD
PULMONARY VENOUS RETURN

Left partial APVR
- usually left upper lobe (vertical vein)
- MDCT - best visualized lateral to aortic arch
- DD$_x$ - left SVC
PULMONARY VENOUS RETURN
LUL PAPVR/LEFT SVC

<table>
<thead>
<tr>
<th>Left SVC</th>
<th>LUL PAPVR (vertical vein)</th>
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<tbody>
<tr>
<td>- Absent BV/ ?right SVC</td>
<td>- Large BV and right SVC</td>
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<tr>
<td>- No vessels from lung</td>
<td>- Vessels coming for LUL</td>
</tr>
<tr>
<td>- Dilated coronary sinus</td>
<td>- Normal coronary sinus</td>
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![Image of chest x-ray showing LUL PAPVR/LEFT SVC with parenchymal and mediastinal findings]
TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)

TAPVR – all pulmonary veins drain to right heart (has obligatory R to L shunt)

- Type 1 – supracardiac (most common)
- Type 2 – cardiac
- Type 3 – infracardiac
- Type 4 – mixed

Associated with complex CHD, esp. asplenia
ADULT CONGENITAL HEART DISEASE
EXTRACARDIAC

CLASSIFICATION

- Thoracic Venous Anomalies
- Thoracic Aortic Anomalies
- Pulmonary Artery Anomalies
CONGENITAL AORTIC ANOMALIES

- Arch anomalies
  - Aberrant Right Subclavian Artery
  - Right Aortic Arch
  - Double Aortic Arch
- Aortic (Pseudo) Coarctation
- Pulmonary Sling
CONGENITAL ARCH ANOMALIES

- Edwards’ model - paired arches
- left fourth arch normally persists, right regresses
- variation causes arch anomalies
NORMAL LEFT ARCH
Aberrant right subclavian artery (ARSA)

- Prevalence - 0.5% (most common type)
CONGENITAL ARCH ANOMALIES

ARSA

» Right subclavian is last vessel and courses posterior to esophagus
» Usually asymptomatic
» Uncommonly “dysphagia lusoria”
» Dilated origin = Diverticulum of Kommerell
» Origin may become aneurysmal
ARSA - DIVERTICULUM
CONGENITAL ARCH ANOMALIES

- Right aortic arch
  - Etiology - involution of left arch with persistence of right arch
  - Two major types
    - Mirror image branching
    - Right arch with aberrant left subclavian artery (ALSA)
RIGHT AORTIC ARCH

• Mirror image branching
  » >95% association with congenital heart disease (esp. TOF, truncus)
  » In adults, often isolated
RIGHT ARCH - MIRROR IMAGE

- Cross-sectional imaging
  - Three arch vessels
  - No retroesophageal vessel
  - In adults, CHD often absent or repaired CHD
**CONGENITAL ARCH ANOMALIES**

- Right aortic arch with ALSA
  - Slight increase in CHD
  - Potential vascular ring with left ductus
  - Aorta usually descends on right
CONGENITAL ARCH ANOMALIES

- Double aortic arch
  - Less common than other lesions
  - Often presents in childhood - vascular ring
  - Right arch higher, larger than left
    - Left arch may be atretic
CONGENITAL ARCH ANOMALIES

- Double aortic arch
  - On CT or MRI, arches pass on either side of trachea and esophagus
  - Most descend on left
  - Arch vessels come from respective arches
  - If left arch atretic, lesion may be mistaken for a right arch and ALSA
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DOUBLE AORTIC ARCH
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<td>Aortic pseudocoarctation</td>
</tr>
<tr>
<td>● more common in adult</td>
</tr>
<tr>
<td>● kinked or “cervical” arch</td>
</tr>
<tr>
<td>● no collaterals</td>
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<td>● discrete stenosis of arch at ligamentum</td>
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COARCTATION

Courtesy of S. Abbara MD
ADULT CONGENITAL HEART DISEASE

CLASSIFICATION

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- Pulmonary Artery Anomalies
PULMONARY SLING

- Anomalous origin of Left PA from Right PA
- Vessels courses between trachea and esophagus
- Associated with airway abnormalities
- Rare in adults
CONGENITAL UNILATERAL ABSENCE OF THE PULMONARY ARTERY

- Agenesis thought to be due to failure of migration of embryonic 6th aortic arch
- Typically associated with bronchial collateral circulation » hemoptysis
- Isolated or associated with congenital heart disease (e.g. TOF)
CONGENITAL ABSENCE OF THE PULMONARY ARTERY
CONCLUSIONS

- Congenital thoracic vascular anomalies in adults are common
- Most are benign but the anatomic variation may have consequences during invasive procedures of surgery
- A few may be symptomatic or life-threatening

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