Congenital Heart Disease
Systematic Interpretation of CT

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Disclosures

Medical Advisory Board Member
Partners Imaging

Consultant / Editing / Authoring (honoraria):
Perceptive Informatics
Amirsys, Inc., Elsevier

Research Funding
NIH
Bayer
Guerbet

Contrast not FDA approved for coronary CTA or MRA
CT Approach to Congenital Heart Disease

Acquisition:

- **ALARA Principle**
  - Can CT sufficiently answer the clinical question?
  - Will CT results affect management?
- **Protocol depends on specific question**
  - May require right heart injection protocols

Interpretation:

- **Segmental Approach**
- **Footsteps of a Surgeon**
- **Know surgical procedures and their complications**

Inglessis I, Abbara S, de Moor M. Which diagnostic modality to choose for Adult Congenital Heart Disease? JCCT 2008; 2(1):23-25
Segmental Approach

(1, 2, 3)

1. Visceroatrial situs
   \( S = \) Solitus, \( I = \) Inversus, \( A = \) Ambiguus

2. Ventricular loop
   \( D = \) dextro, \( L = \) levo

3. Position of Great vessels
   \( S = \) solitus, \( I = \) inversus, \( D-TGV, L-TGV \)
   \( D-MGV = D\)-malposition
   \( L-MGV = L\)-malposition

Visceroatrial Situs

- Situs solitus  **S**
  - Normal (<1%)
- Situs inversus  **I**
  - Abdominalis, Thoracalis, Totalis
  - Usually no cardiac abnormality (3-5%)
- Situs ambiguous  **A**
  - AKA *Heterotaxy Syndromes*
    - Right isomerism (99-100%, severe)
    - Left isomerism (90%, mild)

Heterotaxy Syndromes

Hetero = “different”; -taxy = “arrangement”

- Disturbance of normal left-right asymmetry
- Paired organs often mirror images of one another
  - Lungs, kidneys, atria
  - Malrotation → volvulus
  - Midline liver and biliary atresia
  - MSK, CNS, and urinary tract
- Spleen may be dysfunctional → infections
Heterotaxy Syndromes

Right (atrial) isomerism

- Asplenia = Ivemark's syndrome
- Bilateral rightsidedness
- Two morphological right atria
- Midline liver, malrotation
- Bilateral eparterial bronchi and tri-lobed lungs
- Associated with severe congenital defects:
  - TAPVR, TGA, pulmonary atresia, single ventricle
  - Often cyanotic at birth or soon thereafter
Heterotaxy Syndromes

Left (atrial) isomerism

- Polysplenia syndrome
- Bilateral leftsidedness
- Bilateral hyparterial bronchi
- Bilateral bi-lobed lungs
- Intestinal malrotation
- two morphologic left atria
- ASD, VSD, ECD, PAPVR, double outlet RV, PS
Transverse Midline Liver - Polysplenia
Normal Bronchial Anatomy – Situs Solitus
Situs Inversus

Normal Bronchial Anatomy
Situs Inversus Totalis

→ Kartagener’s Syndrome

Bronchiectasis

Rt Ao Arch

LV

RV

IVC

L Liver

Rt Spleen
Bilateral Eparterial Bronchi

→ Bilateral Rightsidedness → Asplenia

NO Pulmonary artery arching over bronchi
→ Bilateral eparterial bronchi
→ Right isomerism
→ Asplenia
Bilateral Hyperarterial Bronchi

Azygos arch (Azygos continuation of IVC)

Pulmonary arteries arching over both bronchi
→ bilateral hyperarterial bronchi
→ Left isomerism
→ Polysplenia
Cardiac Loop (1, 2, 3)

RV rotates

- either to right (dextro) = D-Loop ‘D’
- or to left (levo) = L-Loop ‘L’

Identify Morphologic Right & Left Ventrices

- Systemic ventricle = connected to aorta

- Morphologic LV
  - Fibrous continuity of AV- and arterial valves
  - No septal trabeculations

- Morphologic RV
  - Conus = complete muscular ring separating AV valve and arterial valve
  - Septal leaflet closer to apex
  - Moderator band, heavier trabeculated
Morphologic **Right Ventricle**

Conus (complete muscular ring) separates AV-valve from arterial valve
Moderator Band
Septal leaflet closer to apex

- Bilateral conus = double outlet right ventricle
- Bilateral absence of conus = double outlet left ventriocle
- Subaortic conus - transposition
Morphologic **Right Ventricle**
Morphologic **Left Ventricle**

Direct communication of AV-Valve with Arterial Valve via Intervalvular Fibrosa (No conus)
No septal Trabeculations
Papillary muscles attached to free wall

Loop Rule

Assumption used if determination of morphologic ventricles is difficult:

• In presence of a **right-sided** aortic valve, the **RV** is to the **right** of the **LV** (d-loop)

• In presence of a **left-sided** aortic valve, the **RV** is to the **left** of the **LV** (l-loop)
Position of Great Arteries

S = Solitus normal
PA anterior and to the left

I = inversus mirrored
PA anterior and to the right

D-TGV dextro transposition
Ao anterior and to the right

L-TGV levo transposition
Ao anterior and to the left

(1, 2, 3)
Normally Related Great Arteries

S = Solitus, NORMAL

I = Inverted
Transposition of Great Arteries

D-TGA or D-MGA

L-TGA or L-MGA
Apply Segmental Approach
Step 1: Visceroatrial Situs \((S, 2, 3)\)

- Right liver, left single spleen, left stomach
- Left sided left atrium
- Cardiac apex leftsided
- Left hyparterial bronchus
- Right eparterial bronchus

→ Situs Solitus
Step 2 - Cardiac Loop  \( (S, L, 3) \)

- Moderator Band, heavy trabeculations & Complete muscular ring of systemic ventricle
- AV-valve hinge-point more apical
- IVC and SVC to RA, PAs to LV

→ Atrio-Ventricular Discordance
→ L-Loop
Step 3 - Great Arteries \( (S, L, L-TGV) \)

- Ao arises from systemic morphologic RV
- PA arises from morphological LV
- Aorta anterior & left of Pulmonary artery
- \( \rightarrow \) Ventriculo-Arterial Discordance
- \( \rightarrow \) L-TGV
Footsteps of a Surgeon
44yom w. SOB. Remote heart surgery for unknown condition.
ECHO: PR, dilated RVOT, RPA stenosis, gradient 27-49mmhg, LPA not seen
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ECHO: PR, dilated RVOT, RPA stenosis, gradient 27-49mmhg, LPA not seen
Footsteps of a Surgeon

L Subclavian BT Shunt, surgically ligated

Membranous VSD Patch Repair

& RV Hypertrophy & Overriding Aorta

Footsteps of a Surgeon

RVOT/PA Patch Repair of Pulmonic Stenosis

Know Complications

Branch Pulmonary Artery Stenosis

Know Complications

Aorto-Pulmonary Systemic Collaterals (MAPCAs)

Thank you!

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