Congenital Heart Disease & Echocardiography

- A Congenital Echocardiologist ……
  - Presumes that every vein, artery, chamber or valve is abnormal, until it is shown to be normal
  - Will use an organized method to examine the cardiovascular system (a step by step approach to avoid missing extra abnormalities)
Segmental Approach to CHD
Definition

- A detailed, sequential description of CV anatomy including
  - Cardiac Position and Axis
  - Apex Orientation
  - Determination of Sidedness (“situs”)
  - Abdominal Organ and Atrial Spatial Arrangements
  - Anatomy and Function of each
  - CV Segment and
  - Connections between Segments

Segmental Approach to CHD
Position vs Axis

Cardiac Position
Where is the Heart?
Left, Right or Midline

Cardiac Axis
How is the Heart Aligned?
- Apex Left – Levocardia
- Apex Right – Dextrocardia
- Inferior / Midline - Mesocardia
Segmental Approach to CHD
Position vs Axis

Levocardia  Dextrocardia

Why Distinguish Between Position and Axis?

- Cardiac Position can change
- Axis is constant
Abdominal Sidedness

- Normal
- Ambiguous

Segmental Approach to CHD
Visceral Anatomy

Tal Geva: Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult
Segmental Approach to CHD
Visceral Anatomy

Situs Inversus Totalis

Situs Ambiguous
Segmental Approach to CHD
Systemic Veins

- Inferior vena cava
- Azygos venous system
- Hepatic veins
- Superior vena cava
- Coronary sinus
Segmental Approach to CHD

Systemic Veins

- **IVC**
  - Always drains into mRA

- **Interrupted IVC**
  - Azygos continuation to SVC

Echo Correlates of Atrial Morphology

**RA Findings**
- Pectinate muscles
- Broad appendage
- Thick septal limbus
- Coronary sinus
- Supra-hepatic IVC

**LA Findings**
- Smooth walls
- Finger-like appendage
- Thin valve of the atrial septum
Segmental Approach to CHD
Atrial Anatomy


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Segmental Approach to CHD
Ventricular Morphology

How do I tell the RV from the LV ??
AV Connection, the Internal Crux & Ventricular Morphology

- AV Valve morphology is directly correlated with ventricular type
  - TV $\Rightarrow$ RV
  - MV $\Rightarrow$ LV
- Internal Cardiac Crux
  - Septal TV leaflet always inserts slightly apical to anterior MV leaflet

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Additional RV Markers
- TV attachments to septum
- Moderator Band
- Multiple, small PMs

Additional LV Markers
- No MV septal attachments*
- Smooth endocardium
- Distinct, large PMs (usually just 2)
Types of AV Connection

Biventricular

- Connection is determined by the annulus
- The 50% rule
- Chordal Straddling does not impact connection

Impact of Annular Override on AV Connection
Types of AV Connection
Univentricular

- Cannot use the internal crux
- Degree of myocardial trabeculation
- Papillary muscle anatomy
  - Septal attachments = TV / RV

Ventricular Morphology in Univentricular AV Connection

Tricuspid Atresia
- Cannot use the internal crux
- Degree of myocardial trabeculation
- Papillary muscle anatomy

HLHS

Ventricular Morphology in Univentricular AV Connection

- The position of the hypoplastic ventricular remnant is most the reliable indicator
  LV's are Posterior
  RV's are Anterior

Ventricular to Arterial Connections

[Diagrams showing different types of connections: Concordance, Discordance, Double outlet, Single outlet, Common outlet]
Aortic Arch Sidedness & Branching

Left Aortic Arch with Normal Brachiocephalic Branching

Right Aortic Arch
Components of the Segmental Approach

CV Segments
- Position, Anatomy, Size and Function of each component of the segment
- State of Septation between the “right” and “left” components of the segment

CV Connections
- Position, Anatomy, Size and Function of each connection between segments
- Relationship of each connector to the preceding and subsequent segments
  - Malalignment or Straddling?

Segmental Approach

CV Segments
- Great Veins
- Atria
- Ventricles
- Great Arteries

Connections
- Veno-Atrial Connection
- Atrial-Ventricular Connection
- Ventriculo-Arterial Connection
Using the Segmental Approach to Evaluate Patients with CHD

CV Segments
- Great Veins
- Atria
- Ventricles
- Great Arteries

Connections
- Veno-Atrial Connection
- Atrial-Ventricular Connection
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= Understanding of CHD