Segmental approach And Evaluation Of Cardiac Morphology In Congenital Heart Disease

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Segmental Approach to the Heart

• Any imaginable combination of atrial, ventricular and great vessel morphology can and does occur in congenital heart disease

• A simple, logical, step-by-step approach is essential to understanding and describing CHD
Segmental Approach to the Heart

Layout of a 3-storied house

• 3 floors (Major Segments)
  – Viscero-atrial situs
  – Ventricular loop
  – Conotruncus
Or, more simply:
  – Atria
  – Ventricles
  – Great Arteries

• 2 staircases (Segmental Connections)
  – AV junction
  – Conus or Infundibulum

• 2 Entrances
  – Systemic veins
  – Pulmonary Veins
Steps in the segmental approach to heart disease

• Anatomic type of each of the 3 major segments. For example \{S,D,D\}, \{I,L,L\}, \{S,D,L\} etc.
• How each segment is connected to the adjacent segment (CAVC, DILV, TGA etc. )
• Associated anomalies within each segment, or between them (TAPVR, ASD, VSD, cleft mitral valve etc.)
• How the segmental combinations and connections, with or without the associated malformations, function

Accurate anatomical and physiological diagnosis allows selection of therapeutic options: medical, surgical, or both.
Steps in the segmental approach to heart disease

Anatomic type of each of the 3 major segments

- Atria, Ventricles and Great arteries
- Segmental set \(\{S,D,S\}\) provides a shorthand description of the floor-plan of the heart
- Right and left do not refer to the side of the body, but to specific morphologic criteria that identify each component of the heart
Viscero-atrial Situs

• Situs refers to the position of the atria and viscera relative to the midline
• Three types: solitus, inversus, ambiguous
Atrial Identification

Right Atrium

- Receives IVC
- Receives coronary sinus
- Broad based triangular appendage with pectinate muscles extending into body of RA
- (Receives SVC)

Left Atrium

- Narrow based finger like atrial appendage
- (Receives pulmonary veins)
IVC as a marker of the RA
Morphology of atrial appendages on MRI

Triangular trabeculated broad based RAA

Tubular smooth narrow based LAA
## Ventricles

<table>
<thead>
<tr>
<th>Right Ventricle</th>
<th>Left Ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Moderator band</td>
<td>• No moderator band</td>
</tr>
<tr>
<td>• Septal attachment of AV valve is more</td>
<td>• Smooth septal surface</td>
</tr>
<tr>
<td>apical</td>
<td>• Septal attachment of AV valve is more</td>
</tr>
<tr>
<td>• Infundibulum (conus)</td>
<td>cranial</td>
</tr>
<tr>
<td></td>
<td>• No conus</td>
</tr>
</tbody>
</table>
Ventricles
**Great Arteries**

<table>
<thead>
<tr>
<th>Pulmonary artery</th>
<th>Aorta</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Branches to the lungs</td>
<td>• Branches to the body</td>
</tr>
<tr>
<td>• No branch to the body</td>
<td>• Coronary artery</td>
</tr>
</tbody>
</table>
Great Arterial Relations

Solitus (S)  Inversus (I)
Great Arterial Relations

D-malposition
Great Arterial Relations

L-malposition
Segmental possibilities at each level

- Atria: S, I, A
- Ventricles: D, L
- Great Arteries: S, I, D, L
Segmental Combinations

<table>
<thead>
<tr>
<th>Types of Human Hearts: Segmental Combinations and Connections</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Normal</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
</tr>
<tr>
<td><strong>Isolated Ventricular Discordance</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
</tr>
<tr>
<td><strong>Isolated Atrial Noninversion</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
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<tr>
<td><strong>Transposition of the Great Arteries</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
</tr>
<tr>
<td><strong>Anatomically Corrected Malposition of the Great Arteries</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
</tr>
<tr>
<td><strong>Double Outlet Right Ventricle</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
</tr>
<tr>
<td><strong>Double Outlet Left Ventricle</strong></td>
</tr>
<tr>
<td><img src="image.png" alt="Diagram" /></td>
</tr>
</tbody>
</table>
Segmental Connections: AV Connection

• Biventricular AV connections

• Univentricular AV connections
Biventricular AV connections

Concordant

Discordant
AV connections
Common AV Canal
Univentricular AV connections

- Absent left AV connection
- Absent right AV connection
- Double inlet LV
- Double inlet RV
Univentricular AV connections

Double inlet left ventricle

Tricuspid atresia
# VA connection: Conus

<table>
<thead>
<tr>
<th>Frontal View</th>
<th>Inferior View</th>
<th>Subpulmonary Conus</th>
<th>Subaortic Conus</th>
<th>Bilateral Conus</th>
<th>Absent or Very Deficient Conus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sup</td>
<td>Ant</td>
<td>TV</td>
<td>Conus</td>
<td>Conus</td>
<td>TV</td>
</tr>
<tr>
<td>Rt</td>
<td>Rt</td>
<td>PV</td>
<td>AD</td>
<td>AoV</td>
<td>MV</td>
</tr>
<tr>
<td>Lt</td>
<td>Lt</td>
<td>Conus</td>
<td>Subaortic</td>
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</tr>
<tr>
<td>Inf</td>
<td>Post</td>
<td>Conus</td>
<td>Conus</td>
<td>Conus</td>
<td>Conus</td>
</tr>
</tbody>
</table>
Conus in D - TGA
Conus in L - TGA
Infradiaphragmatic TAPVR

Associated Anomalies:
Systemic Veins and Pulmonary Veins

Bilateral SVC
Physiology

- Too much Pressure (Afterload)
  - Right sided obstruction
  - Left sided obstruction

- Too much Volume (Preload)
  - Left to right shunts
  - Valvular incompetence

- Intermixing: Right to left shunts / cyanosis

- Poor contractility: Bad myocardium
Steps in the segmental approach to heart disease

• Anatomic type of each of the 3 major segments. For example \{S,D,D\}, \{I,L,L\}, \{S,D,L\} etc.
• How each segment is connected to the adjacent segment (CAVC, DILV, TGA etc.)
• Associated anomalies within each segment, or between them (TAPVR, ASD, VSD, cleft mitral valve etc.)
• How the segmental combinations and connections, with or without the associated malformations, function

Accurate anatomical and physiological diagnosis allows selection of therapeutic options: medical, surgical, or both.
MRI Evaluation of extra-cardiac vasculature

- Aortic coarctation
- Anomalous pulmonary veins
- Scimitar syndrome
- Systemic venous anomalies
- Branch pulmonary artery stenosis
- Anomalous coronaries

- Relatively high incidence of failure of echo to characterize extra-cardiac vascular pathology due to lack of acoustic windows
- Failure rate increases in the post-operative setting, and in older children, when acoustic windows diminish
- Role of MRI in these settings is well established
MRI Evaluation of Cardiac Morphology in CHD

- Atrial, ventricular and great arterial situs
- Segmental connections
- Status of the atrial and ventricular septum
- Cardiac valves
- Ventricular function
- Myocardium

- Echo is successful in delineating intra-cardiac pathology at all ages
- But, even in expert hands, some intra-cardiac defects remain difficult to diagnose by echo
- Role of MRI as a trouble-shooting modality in such situations has not been well evaluated
Common MR sequences used for evaluating cardiac morphology

• Black blood sequences
  ❖ Fast spin echo double inversion recovery
  ❖ Fast spin echo with EPI readout

• Cine sequences
  ❖ Steady state free precession (SSFP)
  ❖ Segmented k-space gradient echo

• 3-D sequences
  ❖ Gadolinium enhanced 3-D MR angiography
  ❖ Navigator respiratory gated isotropic 3-D SSFP

• Cine phase contrast imaging
• Myocardial perfusion and delayed enhancement
Indications for evaluating cardiac morphology by MRI
Clarifying complex segmental cardiac anatomy

- MR is an excellent technique for defining the morphologic features of each atrium and ventricle.
- Provides anatomical data which is easily related to the surrounding structures of the body.
- Provides reliable diagnoses in heterotaxy with a sensitivity approaching 100%.
- MR may be primary imaging technique in the setting of complex intra-cardiac anatomy so as to maximize non-invasive information prior to catheterization.
Criss-cross ventricles, \{S,D,L\} TGA
Atria - Secundum atrial septal defects

• Sensitivity of echo for diagnosing secundum ASD is close to 100% in infancy, but drops to 85 to 90% in older children and adults

• ASD sizing by MRI using SSFP and phase-contrast protocols correlates well with TEE estimation

• MRI provides additional information on ASD shapes and proximity to adjacent structures

• MR guidance used recently to navigate endovascular catheters and deliver ASD closure devices
Secundum ASD
Atria - Sinus venosus defect

- Superior sinus venosus defect difficult to detect by echo due to the extreme rightward and superior position of the defect
- Inferior type is also difficult to depict by echo because of its infero-posterior location to the fossa ovalis
- MRI has become the gold standard for depiction of sinus venosus defects
Atria - Sinus venosus defect
Atria – Atrial septal defects

• Successfully distinguish a common AV canal from an AV canal type of VSD or a primum ASD
• Identify atrial septal defect (ASD) or partial anomalous pulmonary venous connection in adults with right-sided chamber enlargement, hypertrophy or dysfunction of unknown etiology
• In all forms of septal defects, quantification of shunt size (Qp:Qs ratio) by flow velocity mapping enables decision making regarding conservative therapy versus surgery
Atria - Cor triatriatum

- Common pulmonary vein is usually largely incorporated into the left atrium and forms the part of the left atrial posterior wall
- Membrane develops between the common pulmonary vein and the left atrium resulting in stenosis
- Membrane lies between the entrance of the pulmonary veins posteriorly and the foramen ovale and the left atrial appendage anteriorly, in contrast to supra-mitral ring which attaches between the foramen ovale and the left atrial appendage posteriorly and the mitral annulus anteriorly
Cor triatriatum  Supramitral ring
Atria - Atrial appendages

MRI has been used to demonstrate a broad range of pathology involving the appendages:

• Abnormal appendage symmetry in heterotaxy
• Juxtaposition of the atrial appendages in association with other complex malformations
• Thrombi within the appendages
• Aneurysm of the atrial appendage
Atrioventricular connections

• Demonstrate discordant atrioventricular connections and crisscross atrioventricular connections

• Echocardiography is usually employed initially for demonstrating double inlet ventricle, straddling atrioventricular valve, tricuspid atresia, and mitral atresia, and CMR is used to supplement this information

• CMR is superior to echo for quantifying ventricular volumes in these abnormalities, which may be critical for surgical decisions regarding biventricular repair versus the Fontan procedure
Straddling and Over-riding TV
Ventricles - VSD

- MR is highly sensitive and specific for the quantification and detection of ventricular septal defects.
- Certain types of VSD that are difficult to evaluate by echo are well suited to imaging by MR including supracristal defects, LV-RA shunts, and apical muscular VSDs.
- Precisely depict the location of the ventricular septal defect in relation to the great arteries in double outlet ventricles.
Anterior Malalignment VSD in TOF  Apical muscular VSD in ectopia cordis
Ventricular Morphology

- Ventricular anatomy in complex anomalies like double chambered RV, tricuspid atresia, Shone’s syndrome, subaortic stenosis, and univentricular heart
- Differentiation of a single RV from a single LV in functional or morphologic single ventricle
- Most accurate technique for quantifying left and right ventricular mass and volumes
Double Chambered RV
Infundibular outlet chamber in DILV
Ventricles – Single vs biventricular repair

Help surgical decision making regarding univentricular repair (Fontan), one and a half ventricle repair or biventricular repair in patients who have two functioning ventricles, but also have factors preventing biventricular repair:

• straddling AV valves
• Unfavorable location of the VSD
• Suboptimal ventricular morphology or function
Valvular pathology

• High temporal resolution, spatial resolution and interactive real-time nature of echo makes it the primary imaging modality for defining valve morphology
• MR velocity mapping is more accurate and reproducible means of quantifying the severity of regurgitation
• MR helpful in morphologic depiction of tricuspid atresia, Shone’s syndrome and Ebstein's anomaly
• Precise ventricular volumetric and functional assessment in setting of valvular pathology
Ebstein’s Anomaly
Tricuspid regurgitation in ventricular inversion
Surveillance of Corrected CHD

- Ventricular function after Fontan
- Sequential measurements of RV volumes, mass and function in TOF, TGA
- Surveillance of grafts, conduits and baffles
- Early detection of complications
- Determine timing of surgical intervention
Restrictive ASD after Norwood 3
Restrictive atrial communication after Norwood 3
Conclusion

• Important complementary role to echocardiography in evaluation of cardiac morphology and function in children with congenital heart disease in the pre-operative and post-operative period

• Promote awareness amongst clinicians about potential of MRI for cardiac morphology