Echocardiography Conference

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Goals for conference

• Better understanding of transposition of great vessels
  – D/L, ‘congenitally corrected’

• Review of ASD types and board pearls
Case 1

- 40 yo WM with hx Congenitally corrected Transposition of the Great Vessels presenting with increased DOE
- 1994 L Subclavian to L PA shunt
- Pulmonary Stenosis
  - Porcine valve replacement
- VSD
  - Patch repair
- Hx Left AV Regurgitation
  - Repaired 2000
Transposition of the Great Vessels

A – Normal Cardiac development
B – Complete transposition of the Great Vessels
   Note VSD and PDA
C – Congenitally corrected transposition

Hornung et al.
Vocabulary

• Atrioventricular concordance vs. discordance
• Ventriculoarterial concordance vs. discordance
  – Transposition features Ventriculoarterial discordance
• D-loop – Complete transposition; Aorta arises right of pulmonary artery
• L-loop – Congenitally corrected; Aorta arises left of pulmonary artery
The “Broderick” classification

- Atria are in their usual position
- The tricuspid valve attaches to the morphologic RV
- The mitral valve attaches to the morphologic LV
- The aortic valve is connected to the aorta
- The pulmonary valve connects to the pulmonary artery
Complete Transposition

• Most common
  – cyanotic congenital heart disease
  – presenting in infancy
• 5-7% of all congenital heart disease
• 20-30/100,000 live births
• 2:1 male:female

Hornung et al.
Complete Transposition D-Loop

- Aorta arises from RV
  - Coarses right and anterior to Pulmonary Artery
- Pulmonary artery arises from LV
- Coronaries typically from aorta but various origins

Hornung et al.
Associated lesions

- 50% have another heart defect
- 40-45% with VSD
- 25% with pulmonary outflow tract obstruction
- 5% with aortic coarctation
Natural History
No intervention

• 90% die in infancy
• Those WITHOUT other defects do worse
  – Only 30% survive beyond 1 month
• Those with LVOT (Pulm Artery) obstruction and VSD tend to do better
Surgical Therapy

• Mustard and Senning procedures started in 1960’s
• Result in normal Pulmonary->systemic blood flow
• RV and TV still function systemically
• Operative mortality 5% in 1970’s
  – Worse for patients WITH other defects

Hornung et al.
Mustard operation

- Atrial baffle diverts blood from SVC and IVC toward the MV and LV
- Pulmonary blood returned to TV and RV

Hornung et al.
Following surgery

- 85-90% 10 year survival
- 80% 20 year survival
- Late causes of death
  - Presumed arrhythmia
  - RV failure
- Good functional capacity (60-80% NYHA class I)
- Many patients have chronotropic incompetence

Hornung et al.
Right Ventricle

- Morphologic RV destined to provide systemic circulation
- RVEF of 50% considered ‘normal’
- Concern for degeneration of function in 3rd-4th decade
- Geometry yields echo insufficient to evaluate function
- MRI considered gold standard*
- RV typically hypertrophies

Hornung et al.
Tricuspid Valve

• Mild TR (1-2+/4) common
• Significant TR may be due to RV dilitation
• In patients with VSD
  – Tricuspid may be damaged during repair of VSD; may necessitate further repair
Coronary anatomy

• Various origins
• Left cor supplies ANATOMIC LV
• Right cor supplies ANATOMIC RV
Arrhythmia

- Loss of sinus rhythm
- Atrial tachycardia
- May require permanent pacemaker
- Atrial flutter is marker of increased risk of sudden death
Late surgical therapies

• Baffle leaks
  – May close with percutaneous occluder

• Pulmonary or systemic venous stenosis
  – Amenable to balloon dilatation

• Late arterial switch procedure
  – Attempted for RV dysfunction, refractory TR
  – Not very successful, requires ‘retraining’ LV
Medical therapies

• ACE inhibitor
  – No data, ongoing clinical trial

• Beta blockers
  – Presumed effect from ‘normal’ CHF patients
  – No data
Arterial Switch Procedure

• Initially high mortality, but theoretically better
• Should be performed age 3-4 weeks
• Operative mortality 5%
  – Increased risk with other defects, prior surgery
• 5 year survival good
• Patients in 20’s now
Arterial Switch Procedure

- Great arteries transected above the valves and switched
- Cors are removed from aorta with cuff of wall and re-attached to the aorta after the switch
Late complications

• RVOT obstruction
  – LeCompte maneuver to flatten main PA
  – 18% reintervention in 10 years

• Aortic regurgitation
  – 9% incidence at 15 years

• Coronary occlusion/obstruction
  – 18% incidence
Transposition with VSD and Pulmonary Stenosis

- Rastelli procedure
- Operative mortality 5%
- 80% survival at 10 years
- 50% survival at 20 years
- High incidence of late arrhythmia
Rastelli procedure

- VSD closed to become part of LVOT
- RV-PA conduit bypasses stenosis
- Conduit stenosis is inevitable!
Congenitally corrected transposition
L-Loop

- Rare, atrioventricular discordance and ventriculoarterial discordance
- 0.5% of congenital heart disease
- Blood flow pattern is normal
- RV supports systemic circulation
- 80-90% incidence of other defect
  - 60-80% with VSD
  - 30-50% with Pulmonary outflow obstruction
  - 90% with Ebstein’s anomaly (clinical picture variable)
  - Complete heart block 2%/year
Natural history of congenital corrected transposition

- Variable
  - If no other defect, may only be discovered at autopsy
- Mean age of death about 38% in 1 series
- Various associated lesions make stats difficult
- Subnormal exercise capacity
Right ventricle

- Progressive dysfunction
- 67% with CHF by age 45
- No data to support medical therapy, though ACE inhibitor used
Tricuspid valve

• Regurgitation common
• Repair is feasible but replacement may be more beneficial
Double switch operation

- Small number of patients have
  - Great vessel arterial switch
  - Atrial baffle to redirect blood flow
- Anatomic LV->Aorta
- Anatomic RV-> Pulm Artery
## Echo features

<table>
<thead>
<tr>
<th>Feature</th>
<th>RV</th>
<th>LV</th>
</tr>
</thead>
<tbody>
<tr>
<td>AV valve</td>
<td>Trileafet</td>
<td>Bicuspid (unless cleft)</td>
</tr>
<tr>
<td>Annulus</td>
<td>More apical</td>
<td>More basal</td>
</tr>
<tr>
<td>Trabeculation</td>
<td>Prominent</td>
<td>Less prominent</td>
</tr>
<tr>
<td>Moderator band</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Infundibulum</td>
<td>Present</td>
<td>Absent</td>
</tr>
</tbody>
</table>

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Cardiac Catheterization

- CX – OM1, OM2
- LAD small
- RCA – large RV branches, PLV, PDA branches
Cardiac Catheterization
Hemodynamics

- RA 10
- LV 118/6 EDP 18
- PA 139/79, mean 103
- RV 92/2 EDP 2
- AO 91/52 mean 70
- Oxygen saturation on RA: PA 23%, Ao 80%
- Following Oxygen administration: PA 35%, Ao 88%
- No change in Right sided pressures with Oxygen
- CO 2.34, CI 1.15
- Severe RV dysfunction EF 25%, 4+ regurgitation
Followup of Pt

• Referred initially to Mayo, declined reintervention
• Accepted at Cleveland clinic (site of prior surgeries) for possible TV repair/replacement
• Ultimately received Heart-lung transplant
Patient #2

- 24 yo WF with ‘heart murmur’
- Mild DOE
- Previously negative TEE for ASD
Types of ASD

• **Secundum** – most common (7% of all congenital heart disease)
  – Failure of secundum atrial septum to cover foramen secundum

• **Primum**
  – Abnormal septum primum with failure of closure
  – Associated with *cleft mitral valve*

• **Sinus venosus**
  – Least common (2-3% of ASD)
  – Abnormal fusion of sinus venosus and atrium
  – Near junction of IVC/SVC and atrium

Otto, 408
FIGURE 16–15. Sinus venosus atrial septal defect in a 74-year-old man. Although the transesophageal four-chamber view shows marked volume overload of the right side of the heart, the atrial septum appears intact in this view.
FIGURE 16-16. In the same patient as in Figure 16-15, the region of the atrial septum just inferior to the superior vena cava is absent (above), and color flow demonstrates left-to-right flow across this defect (below). (See Color Plate 19, p. 386.)
Pearls for ASD

• **Secundum**
  – Most common type
  – Commonly presents in adulthood

• **Primum**
  – Associated with cleft MV

• **Sinus venosus**
  – Associated anomalous pulmonary vein return
  – Difficult to visualize
  – Catheter may ‘slip’ easily into pulmonary vein
References