Interventions in Adult Congenital Heart Disease: Role of CV Imaging

Sangeeta Shah MD, FACC, FASE
Associate Professor

ACHD mortality

Adult Congenital Heart Disease

Heterogeneity Symptoms

Treatment

Percutaneous/Surgical
Electrophysiologic
Medications

Adult Congenital Heart Disease and CV imaging

- Atrial septal defect
- Tetralogy of Fallot
- D- TGA with atrial switch
Guidelines for the Echocardiographic Assessment of Atrial Septal Defect and Patent Foramen Ovale: From the American Society of Echocardiography and Society for Cardiac Angiography and Interventions

Frank E. Silvestry, MD, FASE, Chair, Meryl S. Cohen, MD, FASE, Co-Chair, Laurie B. Arrasibay, MD, FSNAI, Nidin J. Burkle, MD, DM, FASE, Craig E. Ficialman, MD, FASE, Ziyad M. Hijazi, MD, MPH, FSCAI, Roberto M. Lang, MD, FASE, Jonathan J. Rome, MD, and Yan Wang, RD, Philadelphia, Pennsylvania; Portland, Oregon; Thane, India; Orlando, Florida; Doha, Qatar; and Chicago, Illinois

Recommendation for Closure

CLASS I

Closure is indicated for RA or RV enlargement with or without symptoms

CLASS IIa

Paradoxical embolism

Documented orthodeoxia-platypnea
<table>
<thead>
<tr>
<th>PERCUTANEOUS</th>
<th>SURGERY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secundum</td>
<td>All other types</td>
</tr>
<tr>
<td></td>
<td>Fenestrated ASD</td>
</tr>
</tbody>
</table>

**Secundum: Percutaneous**

- Size
- Location
- Adequate Rims: >5mm
Size

- Amplatzer Septal Occluder
  - up to 38mm diameter
- Helex Occluder
  - up to 18mm diameter

Rims

- At least 75% Rim
- > 5mm lip in all views
Rims and Views

- Bicaval 90°
- Posterior Superior + Posterior Inferior Rims

Fenestrations

- TEE with color flow Doppler: Swiss Cheese
- 3D: Swiss Cheese
Case

- 36 y.o. woman with increased dyspnea on exertion noted to have an enlarged heart on Echo
- TEE performed for assessment of ASD prior to percutaneous closure

Mid-Esophageal 4 chamber

Anterior Inferior + Posterior
Mid-esophageal SAX 45°

Superior Anterior + Posterior

Bicaval 90°

Posterior Superior + Posterior Inferior
Summary

Fenestrated ASD with an atrial septal aneurysm

Surgical Closure

Tetralogy of Fallot
## Complications

<table>
<thead>
<tr>
<th>RIGHT SIDE</th>
<th>LEFT-SIDED</th>
</tr>
</thead>
<tbody>
<tr>
<td>Significant PI</td>
<td>Left side arrhythmia</td>
</tr>
<tr>
<td>Right heart dilation with dysfunction</td>
<td>LV dysfunction</td>
</tr>
<tr>
<td>Right-side arrhythmias</td>
<td>Aortic aneurysm</td>
</tr>
<tr>
<td>RV hypertension secondary to pulmonary artery stenosis</td>
<td></td>
</tr>
</tbody>
</table>

**Sudden Cardiac Death of 6%**

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**GUIDELINES AND STANDARDS**

*Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology*

Anne Marie Valente, MD, FASE, Co-Chair; Stephen Cook, MD, Pierluigi Festa, MD, H. Helen Ko, J5, RDMS, RDMS, FASE, Rajesh Krishnamurthy, MD, Andrew M. Taylor, MD, Carole A. Warnes, MD, Jacqueline Kreutzer, MD, and Tal Geva, MD, FASE, Co-Chair; Boston, Massachusetts; Pittsburgh, Pennsylvania; Mason, Italy; New York, New York; Houston, Texas, London, United Kingdom; Rochester, Minnesota

*(J Am Soc Echocardiogr 2014;27:111-41.)*
Case 2

- 32 y.o. male with sudden cardiac death has been resuscitated
- PE: BP 110/70 with HR of 77 bpm; height 70in; weight 200 lbs; BSA 2.1 m2
- Right thoracotomy scar and mid sternal scar
- 3/6 systolic EM at RUSB with a 1/6 early peaking diastolic murmur

EKG
Tetralogy of Fallot

RVOT View - Conduit
CW Doppler

3.7 m/sec; mean 31mmHg; Moderate-severe PI

Cardiac MRI

- Gold standard
- Left and Right ventricular volume and ejection fraction
- MRA
  - conduit
  - pulmonary artery
  - aorta
Cardiac MRI

- Ventricular function
- LVEDV 54cc/m2 LVESV 24cc/m2; EF 56%
- RVEDV 121cc/m2 RVESV 68cc/m2 EF 44%

RVOT

Phase Contrast

Peak velocity 3.2m/sec
<table>
<thead>
<tr>
<th>VALVE</th>
<th>TYPE</th>
<th>APPROVED USE</th>
<th>EXPANDABLE DIAMETER</th>
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</thead>
<tbody>
<tr>
<td>MELODY</td>
<td>Bovine jugular venous valve in covered stent</td>
<td>RVOT conduits &gt;16mm</td>
<td>20,22</td>
</tr>
<tr>
<td>SAPIEN</td>
<td>Bovine pericardial valve on stainless steel stent</td>
<td>Conduit &gt;21mm</td>
<td>23,26</td>
</tr>
<tr>
<td>SAPIEN XT</td>
<td>Bovine pericardial valve on stainless steel stent</td>
<td>Aortic, mitral</td>
<td>20.23.26.29</td>
</tr>
<tr>
<td>NATIVE OUTFLOW DEVICE</td>
<td>Porcine pericardial valve on nitinol stent</td>
<td>Investigational</td>
<td></td>
</tr>
<tr>
<td>VENUS-P</td>
<td>Porcine pericardial valve in covered self-expanding stent</td>
<td>Investigational</td>
<td></td>
</tr>
</tbody>
</table>

Melody Valve
27 y.o. female with history of repaired Tetralogy of Fallot with right BT shunt (15 months) followed by complete repair (3 yo) and subsequent RV to PA conduit (18 yo)
Significantly reduced Peak Vo2

Kemeny. European Heart Journal 2012; 33:1386-96

Cardiac MRI

LVEDV 50cc/m2 LVESV 22cc/m2; EF 56%
RVEDV 93cc/m2 RVESV 69cc/m2 EF 24%

SV 36 cc
Regurgitation volume 4cc
Peak velocity of 3.2m/sec
Percutaneous Valve not an option

- Surgical repair
  - 25mm St Jude bioprosthesis
  - Right PA augmentation

D-TGA - Atrial Switch
Complications with percutaneous options

- Baffle Obstruction/Leak
Case D-TGA- Atrial switch

- 32 y.o. male
- Viral syndrome for one week; treated with antibiotics & prednisone; still with dyspnea on exertion and fatigue
- Vitals: BP 110/70; HR of 150 bpm; Pox 92%
- Cardiovascular exam: tachycardia with systolic murmur; JVP of 10cm; HJR; +1 LE edema
Baffle Leak

- Saturations poor sensitivity for detection of baffle leak
- Incidence of baffle leak of 50-75%
- Especially important if deauration with exercise or cardiac lead placement
- Agitated Saline injection superior to MRI for evaluation of baffle leak

Wilhelm. Echocardiography 2016;33:437
Amplatzer Closure

Ms Teddy

- 32 y.o. D-TGA with ABP complains of progressive dyspnea on exertion.
- G2P2 with a 6 year old and 3 year old who was noted
- BP 100/70 P 80 Pox 80%
- Cardiovascular: RRR 3/6 systolic on RSB and 2/6 at the mid clavicular line. JVP is 10cm
TEE Agitated Saline

- She
Baffle Obstruction

- 35 y.o. male with D-TGA with atrial switch. He has been experiencing palpitations with exercise intolerance.

Has a TEE and undergoes DCCV. Develops 4-6 second sinus pauses.
Percutaneous Stenting

- 1/3 D-TGA atrial switch develop baffle obstruction
- Stenting suggested
- mean gradient of 6mmHg
- <10mm diameter
- prior to device placement

Poterucha. CCI 2016
Adult Congenital Heart Disease

- Heterogeneity Symptoms
- CV imaging
- Percutaneous/Surgical Intervention