Congenital Heart Disease I: The Unrepaired Adult

Doreen DeFaria Yeh, MD FACC
Assistant Professor, Harvard Medical School
MGH Adult Congenital Heart Disease Program
Echocardiography Section. No disclosures
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Overview:
Unrepaired Adult Congenital Heart Disease

- Case review of common and uncommon congenital lesions
Natural History of Unrepaired CHD

Growing Adult CHD population

- 1.2M Adults in the US with Congenital Heart Disease

Williams et al. Report of the NHLBI working group on research in ACHD. J Am Coll Cardiol 2006;47:701-7
46M history of a restrictive VSD new diastolic murmur
46M asymptomatic. You recommend:

- A. Serial echo monitoring as the defect is restrictive
- B. Percutaneous closure to the VSD and aortic root fistula
- C. Surgical valve sparing aortic root replacement and VSD closure
- D. Monitoring for LV volume load, then surgical correction
**Ventricular Septal Defects**

- **Inlet:**
  - AV septal defect, may be associated with ASD

- **Outlet / Supracristal:**
  - can lead to Ao RCC prolapse

- **Membranous:**
  - Commonly closes spontaneously

- **Muscular:**
  - May be multiple

**Associated Lesions:**
- Pulmonic stenosis, BAV, coarctation, subaortic membranes

**Complications of VSDs:**
- Left Heart Enlargement
- Atrial Arrhythmias
- Endocarditis
- Aortic Cusp Prolapse; Aortic Insufficiency
- Sinus of Valsalva Aneurysm → Fistula (continuous murmur)
- Pulmonary Hypertension/ Eisenmenger Physiology
ACHD Guidelines: VSD

**CLASS I**
- Catheterization to assess operability of adults with VSD and PAH
- Closure for $Qp/Qs > 2.0$ and clinical evidence of LV volume overload ($B$)
- History of endocarditis

**CLASS IIa**
- Closure is reasonable:
  - Net L>R shunt with $Qp/Qs > 1.5$ and PASP $< 2/3$ systemic, PVR $< 2/3$ SVR ($B$)
  - Net L>R shunt with $Qp/Qs > 1.5$ in the presence of LV systolic or diastolic failure ($B$)

**CLASS IIb**
- Pulmonary vasodilators for VSDs with progressive/severe pulmonary vascular disease ($B$)

**CLASS III**
- VSD closure is not recommended in patients with severe irreversible PAH ($B$)

Warnes, Circ 2008

64F year old male with new atrial flutter

- Exertional fatigue and two 'normal' prior echos
RV dilation must be explained...
Superior Sinus Venosus Defect
Atrial septal defects

- **Sinus Venosus**: (rare)
  - ECG: junctional or low atrial rhythm
  - Anomalous pulmonary venous drainage into RA or vena cavae

- **Ostium Primum**:
  - MR, cleft MV leaflet, VSD
  - ECG: RBB morphology, LAD 1st degree AVB (75%)

- **Ostium Secundum**:
  - MVP (10-20%)
  - ECG: RBB morphology, RAD
Atrial septal defects

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Complications related to ASDs in adults

- Paradoxical embolization
- Atrial arrhythmias
- Right heart enlargement; Exertional fatigue
- Pulmonary hypertension

ACHD Guidelines

CLASS I
- RA or RV enlargement
  - With or without symptoms
- Percutaneous closure for secundum defects
- Surgical closure: sinus venosus, coronary sinus, or primum ASD

CLASS IIa
- Surgical closure of secundum ASD is reasonable:
  - when the anatomy of the defect precludes the use of a percutaneous device. (C)
- paradoxical embolism (C)
- orthodeoxia-platypnea (B)
30M recently emigrated to the US. VSD murmur and leg claudication with radial-femoral delay
The following statement is true.

- A. Surgical correction of coarctation is indicated if the peak gradient by echo exceeds 20mmHg
- B. Surgical correction of coarctation is indicated if the mean gradient by echo exceeds 20mmHg
- C. Surgical correction of coarctation is indicated if the peak to peak cath gradient exceeds 20mmHg
- D. Surgical correction of coarctation is not indicated in the presence of collateral vessels

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Aortic Coarctation

- 6-8% of all congenital heart disease
- 4/10,000 live births
- Male: Female 2:1
- Diffuse arteriopathy
- **Hypertension**
- Berry aneurysm screening

**Associated abnormalities:**
- Bicuspid aortic valve (50-60%)
- Mitral valve abnormalities
- Subaortic membrane
- VSD, PDA
- Aortic arch anomalies/ aberrant subclavians

Guidelines for intervention

**CLASS I:**
- Peak to peak gradient > 20 mmHg by catheterization
- Gradient <20mmHg with evidence of collaterals
- Persistent hypertension

Role for Functional testing? Exercise ABIs?
Percutaneous Interventional approaches

- Coarctation Stenting:

Images courtesy Ignacio Inglessis, MD
Coarctation of the Aorta
Extra-anatomic Bypass


Coarctation of aorta: take home points

- Think about coarctation among patients with BAV
- Search for associated abnormalities
- **Always evaluate descending aorta with Doppler
- **Always evaluate abdominal aortic Doppler profile
- Advanced imaging may be necessary to determine percutaneous vs. surgical candidacy
- Hypertension is common; diastolic dysfunction
Case: 65M with progressive fatigue and heart failure

- Known congenital heart disease and kidney disease
- He has a leaky valve
- No prior intervention

- Several recent heart failure admissions, presented in cardiogenic shock, cardiorenal syndrome
Putting it all together: [S, L, L] CC-TGA

- Levocardia with normal visceroatrial situs (S)
- Atrioventricular discordance, bialtrial dilation
  - Severe systemic TR
- Ventriculararterial discordance (L- ventricular looping)
  - Biventricular dilation and dysfunction
- L transposed great vessels
- Elevated LVSP without LVOT obstruction or PS → significant pulmonary arterial hypertension

Long term issues: CC- TGA (or L-TGA)

- Systemic tricuspid regurgitation

- Systemic RV dysfunction—50% risk of HF by age 35

- Progressive conduction disease, need for pacer
  - Rate of complete heart block approximately 2%/year
Summary

- ACHD: growing and aging population

- Unrepaired adults are not uncommon, if you are not picking up congenital heart disease you are missing it

- Look for associated lesions

Thank You
ddefariayeh@mgh.harvard.edu
ACHD Growth Implications

Hospital admissions for ACHD in the U.S. more than doubled between 1998 and 2005. Total national hospital charges increased from $691 million in 1998 to $3.16 billion in 2005.

Opotowski JACC. 2009