

# Congenital Heart Disease I: The Unrepaired Adult

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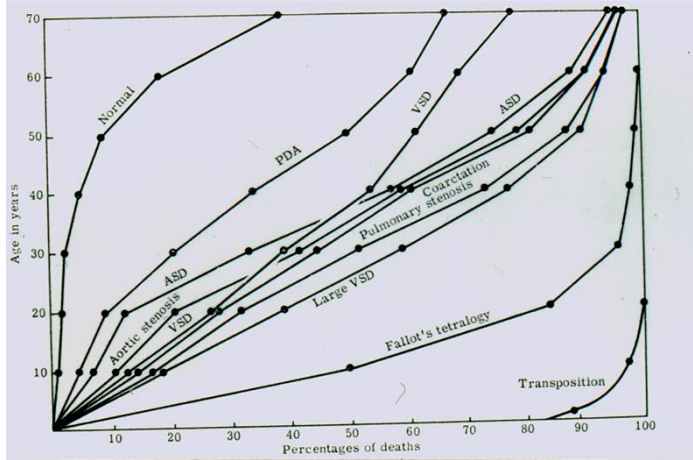
## Overview: Unrepaired Adult Congenital Heart Disease

- Case review of common and uncommon congenital lesions



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# Natural History of Unrepaired CHD

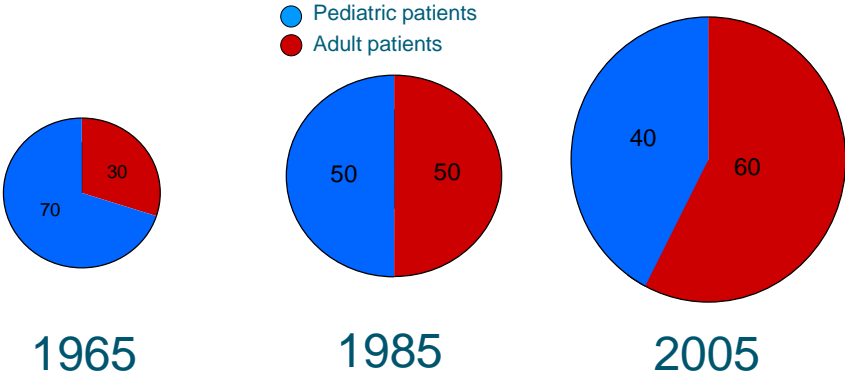


J Hoffman, 1965

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# Growing Adult CHD population

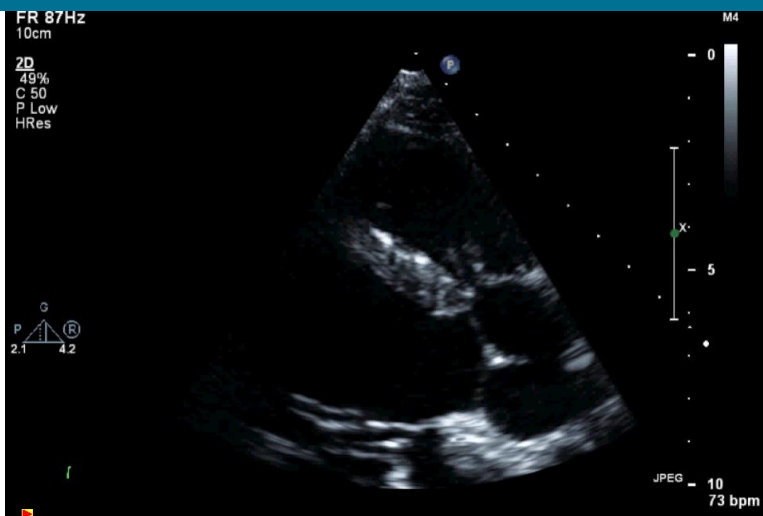
- 1.2M Adults in the US with Congenital Heart Disease

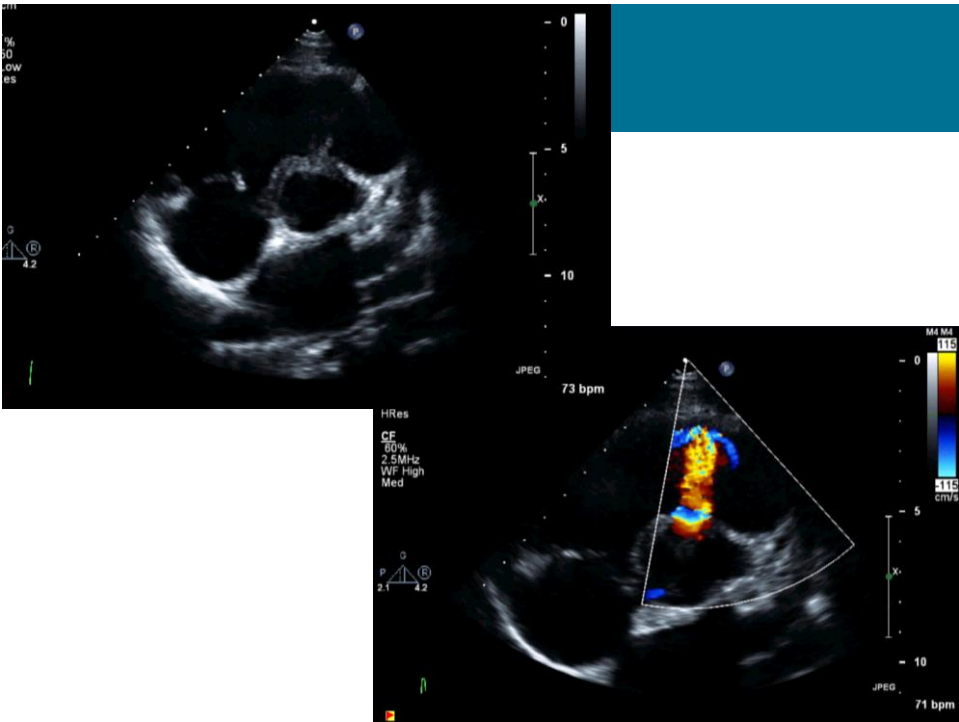
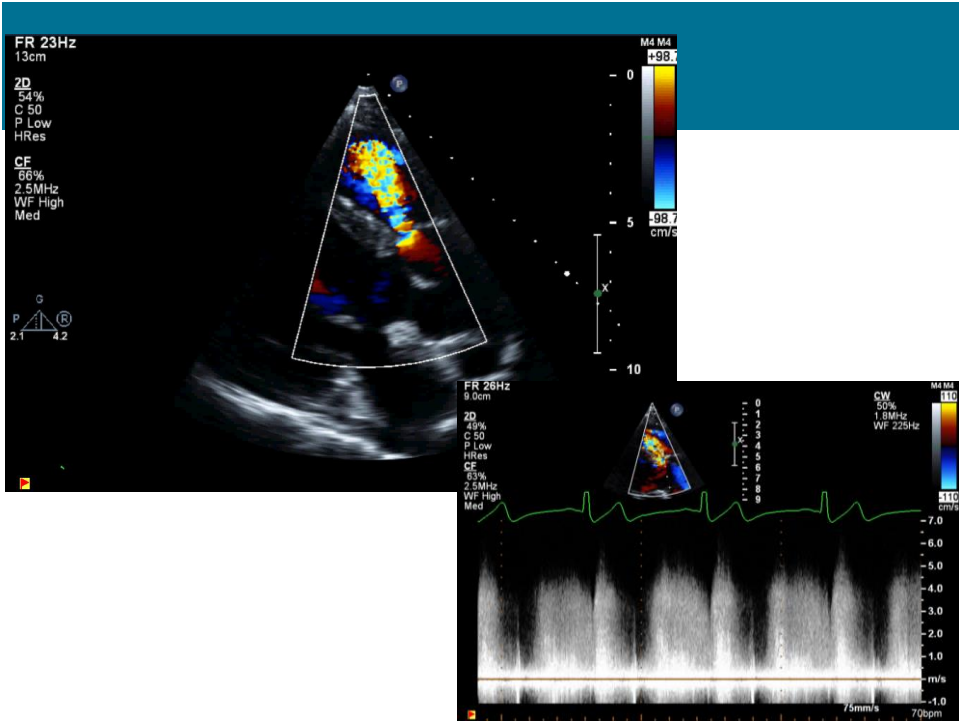


Williams et al. Report of the NHLBI working group on research in ACHD.  
*J Am Coll Cardiol* 2006;47:701-7

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# 46M history of a restrictive VSD new diastolic murmur





## 46M asymptomatic. You recommend:

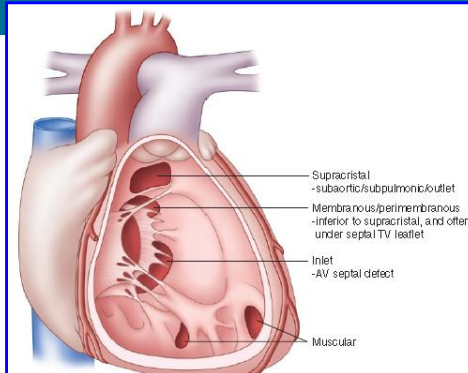
- A. Serial echo monitoring as the defect is restrictive
- B. Percutaneous closure to the VSD and aortic root fistula
- C. Surgical valve sparing aortic root replacement and VSD closure
- D. Monitoring for LV volume load, then surgical correction

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# 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



DeFaria, Liberthson, Bhatt. 2013

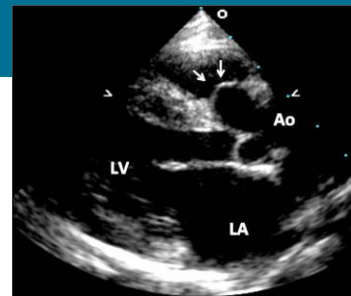
## Associated Lesions:

- Pulmonic stenosis, BAV, coarctation, subaortic membranes

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## 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



DeFaria, King Curr Card. 2015

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# ACHD Guidelines: VSD

## CLASS I

- Catheterization to assess operability of adults with VSD and PAH
- Closure for **Qp/Qs of > 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)**

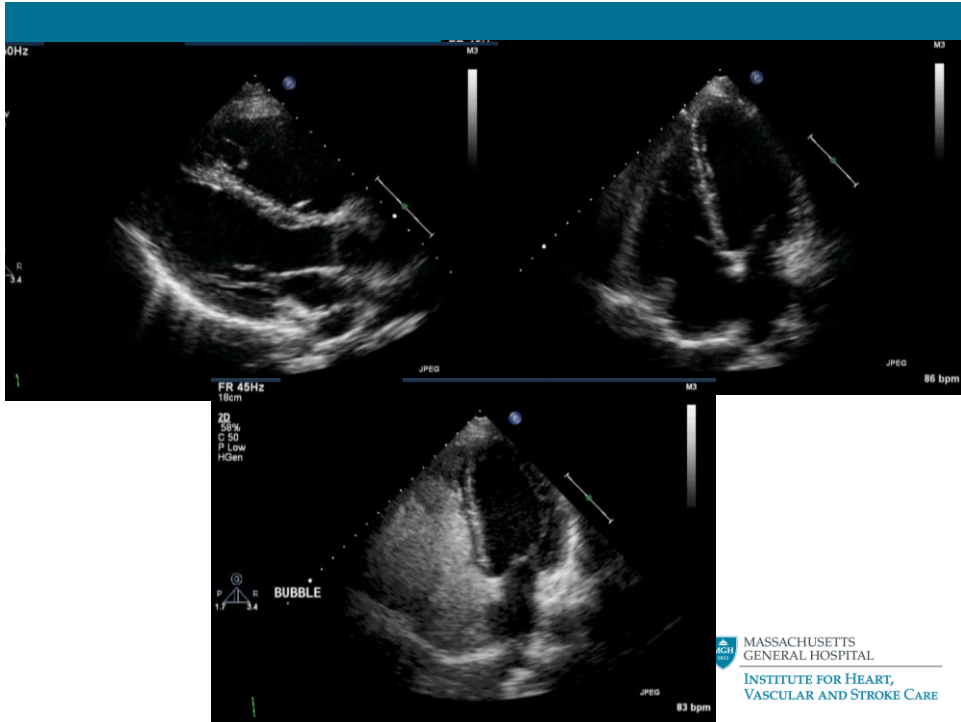
Warnes, Circ 2008



# 64F year old male with new atrial flutter

- Exertional fatigue and two 'normal' prior echos





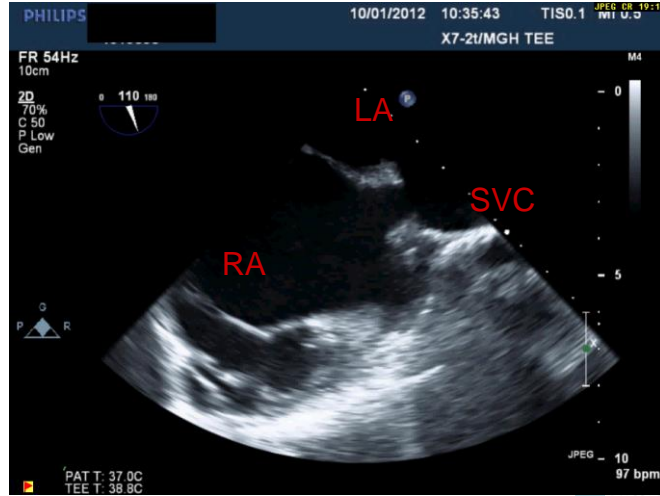
RV dilation must be explained...



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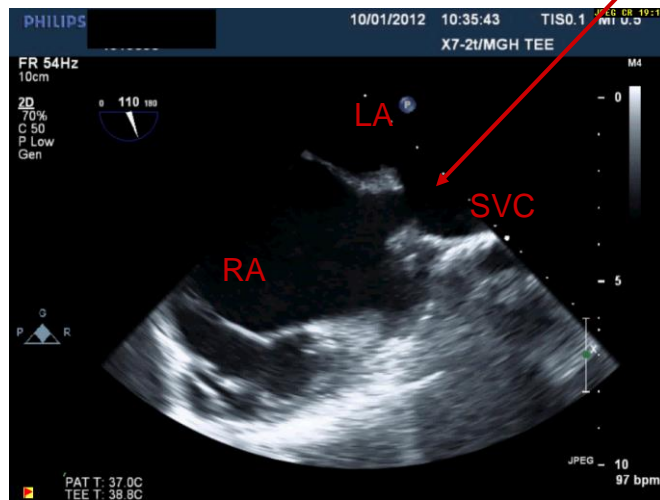
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# Superior Sinus Venosus Defect

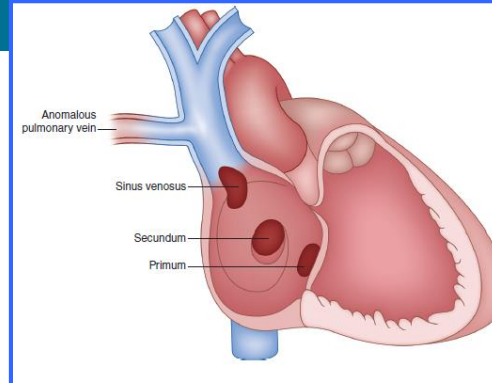
defect



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## Atrial septal defects

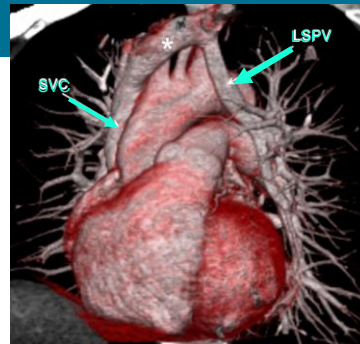
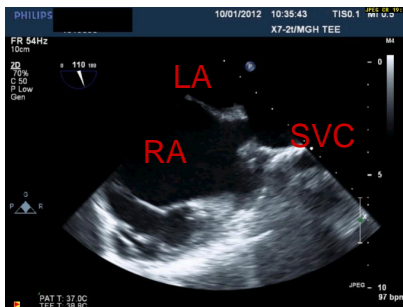
- **Sinus Venosus: (rare)**
  - ECG: junctional or low atrial rhythm
  - Anomalous pulmonary venous drainage into RA or vena cavae
- **Ostium Primum:**
  - MR, cleft MV leaflet, VSD
  - ECG: RBB morphology, LAD 1st degree AVB (75%)
- **Ostium Secundum:**
  - MVP (10-20%)
  - ECG: RBB morphology, RAD



DeFaria, Liberthson, Bhatt. 2013

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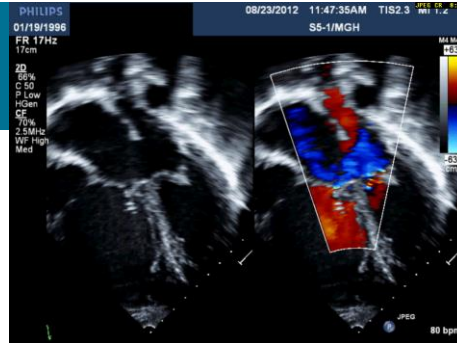
## Sinus Venosus Defects



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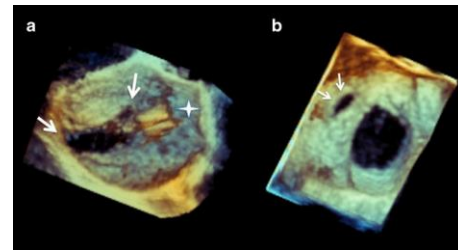
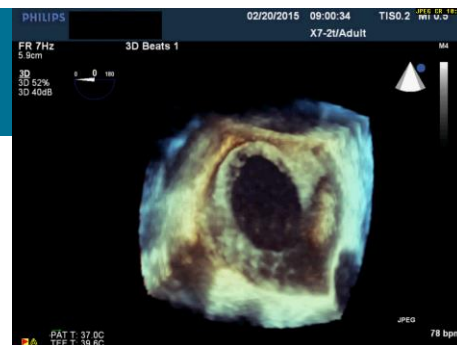
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DeFaria King, Curr Card 2015

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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



Amplatzer PFO Occluder  
Illustration courtesy of  
AGA Medical Corp.



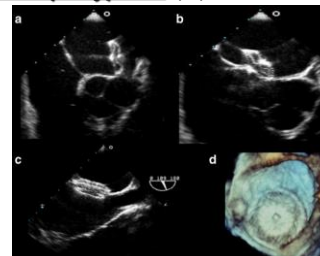
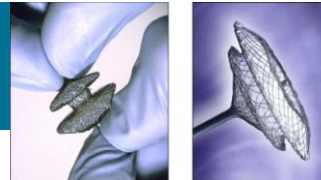
CardioSEAL Septal Occluder  
Illustration courtesy of  
MMF Medical



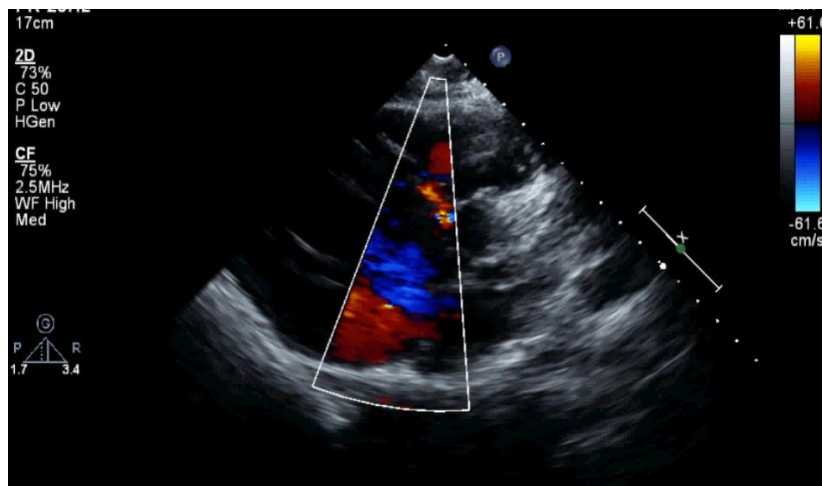
Gore Helix Septal Occluder  
Illustration courtesy of  
W. L. Gore & Associates, Inc.

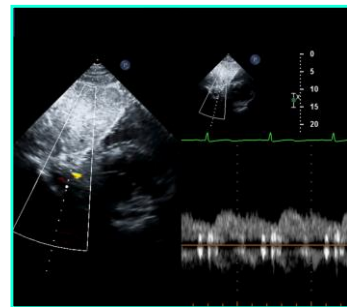
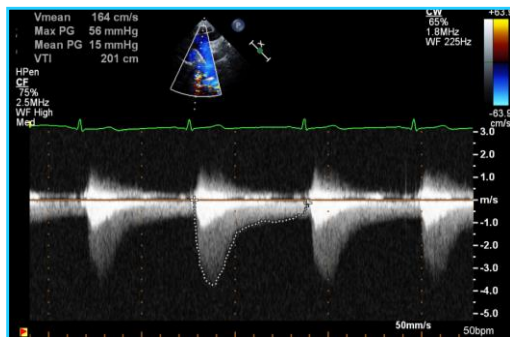
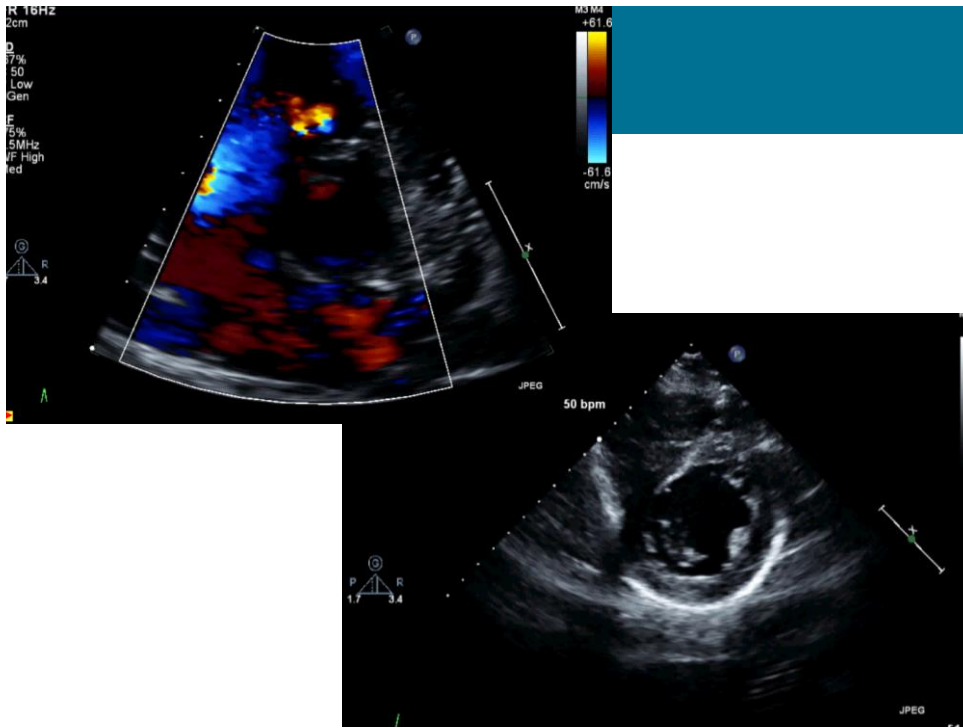
### CLASS IIa

- 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

29

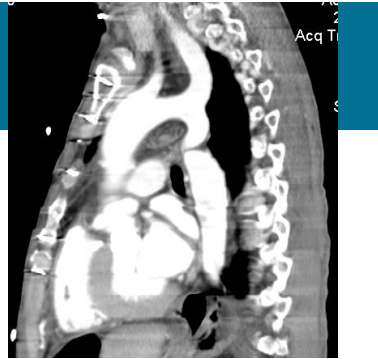
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30

## 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

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## Guidelines for intervention

### **CLASS I:**

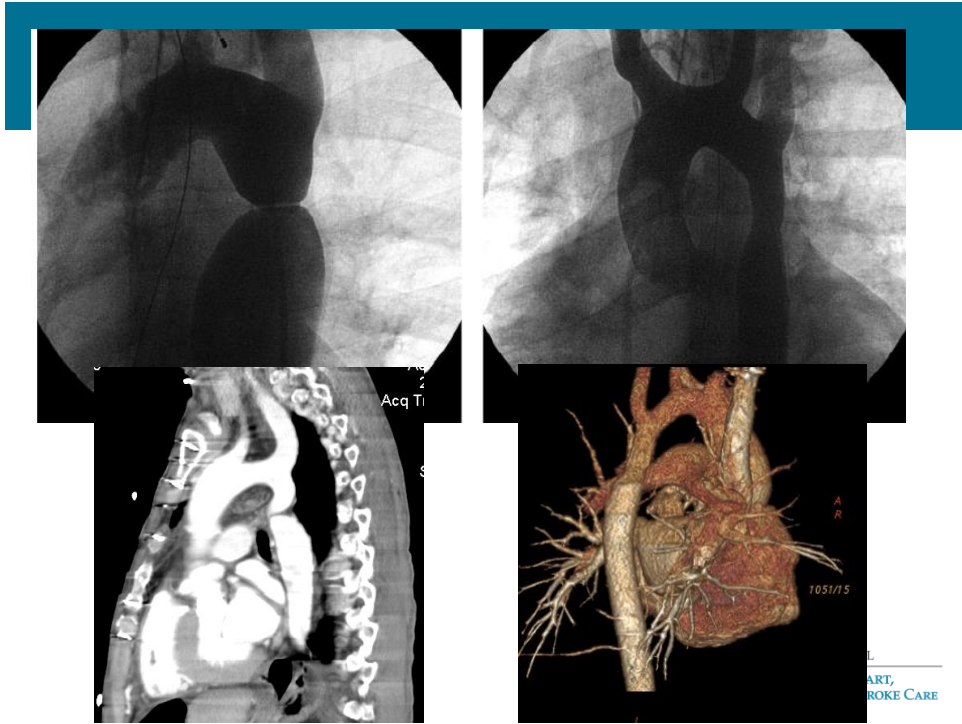
- Peak to peak gradient  $\geq 20$  mmHg by catheterization
- Gradient  $<20$ mmHg with evidence of collaterals
- Persistent hypertension

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Role for Functional testing? Exercise ABIs?

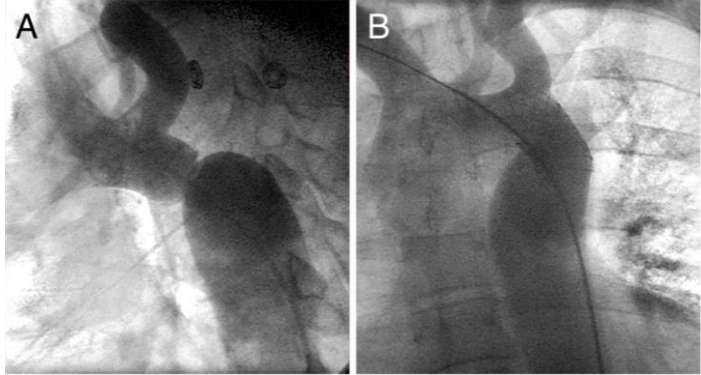
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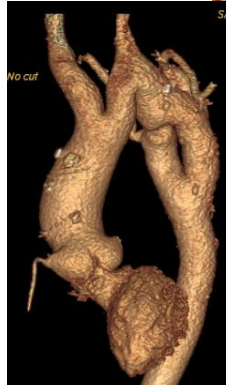
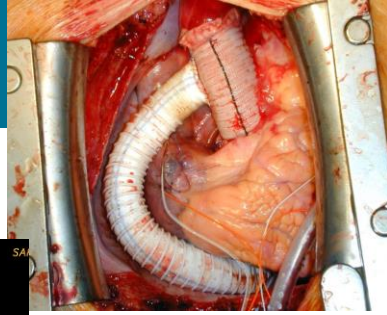
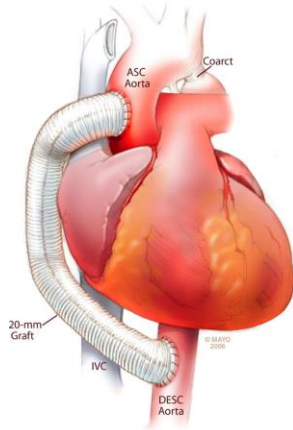
## Percutaneous Interventional approaches

- Coarctation Stenting:



Images courtesy Ignacio Inglessis, MD

## Coarctation of the Aorta Extra-anatomic Bypass



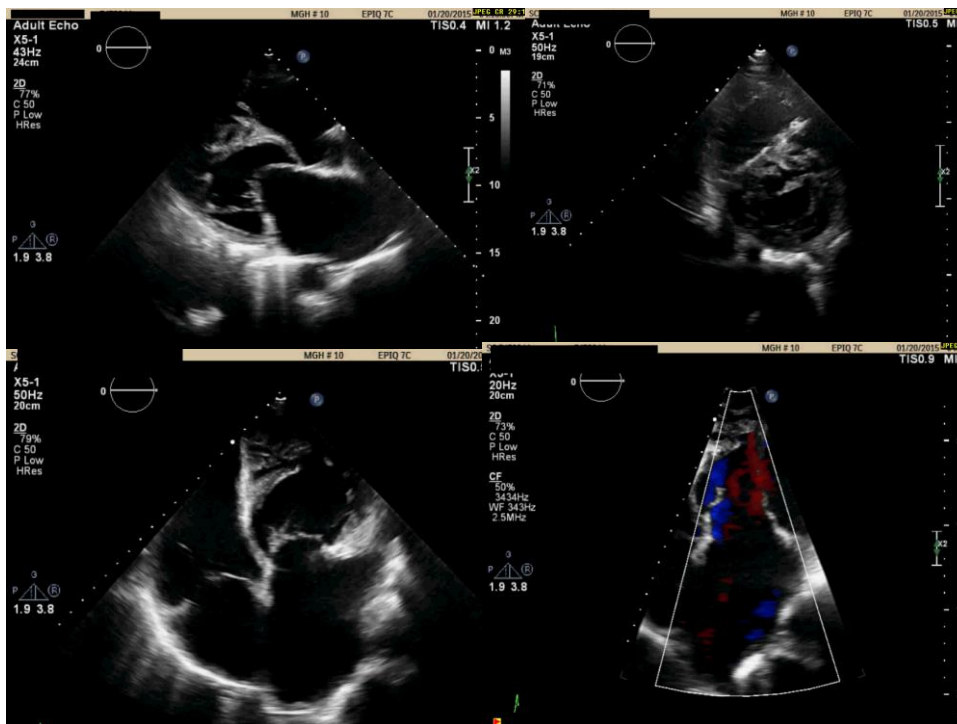
McKellar SH et al. J Thorac Cardiovasc Surg 2007 5  
Schnhoff FS et al. Ann Thorac Surg 2008.

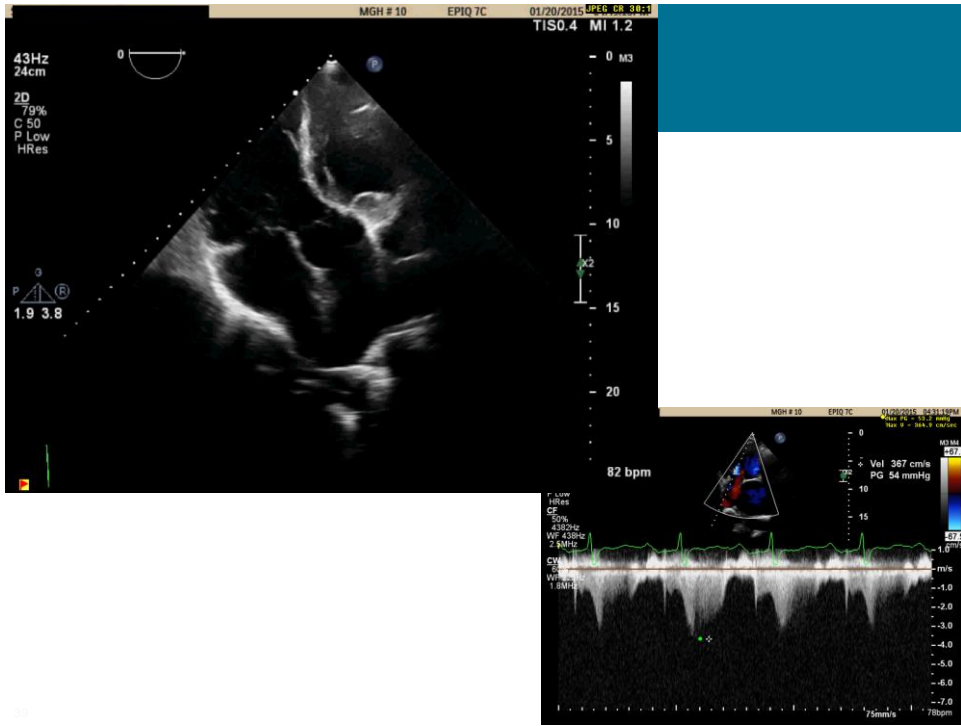
## Coarctation of aorta: take home points

- 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

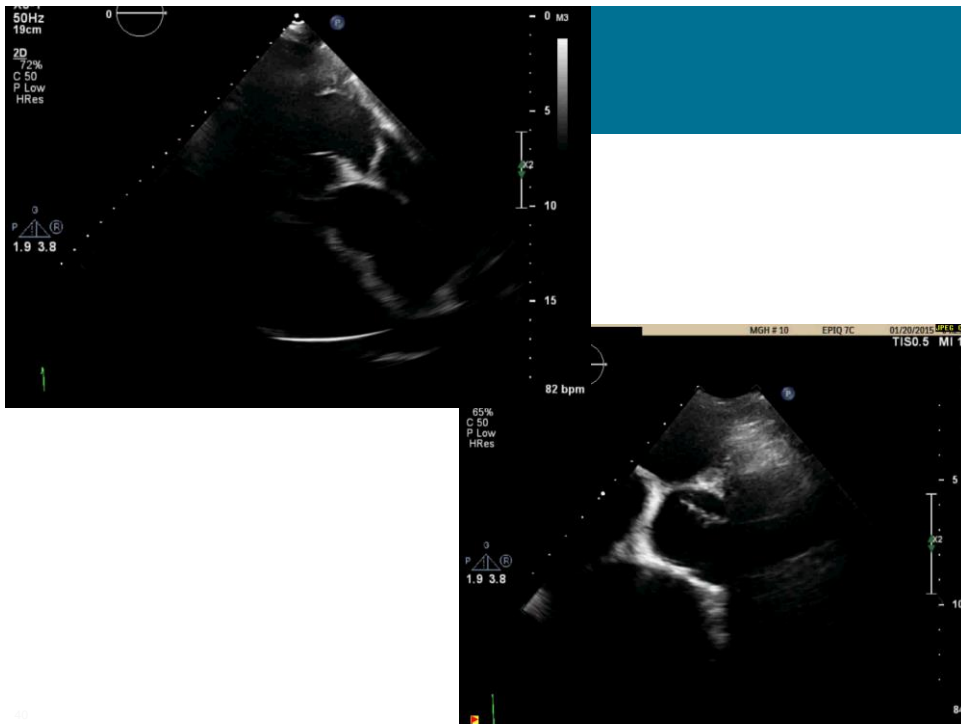
## Case: 65M with progressive fatigue and heart failure

- 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





39

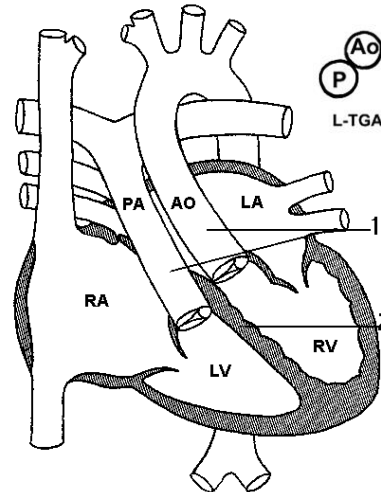


40

## Putting it all together: [S, L, L] CC-TGA



- Levocardia with normal viscerotrial situs (S)
- Atrioventricular discordance, biatrial 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



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## 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

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## 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  
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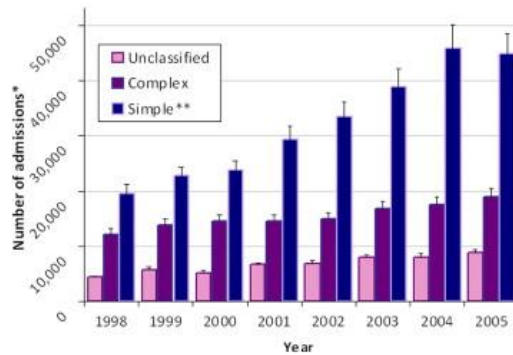
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45

# ACHD Growth Implications



Opotowski JACC. 2009