

ADULT CONGENITAL HEART DISEASE

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 - Research Focus: Imaging and Outcomes

Disclosures

I have no relevant financial relationships with ineligible companies.

Objective

- To highlight the top 10 take-home messages from the recently published AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease



2025 ACC/AHA/HRS/ISACHD/SCAI Guideline for the Management of Adults With Congenital Heart Disease

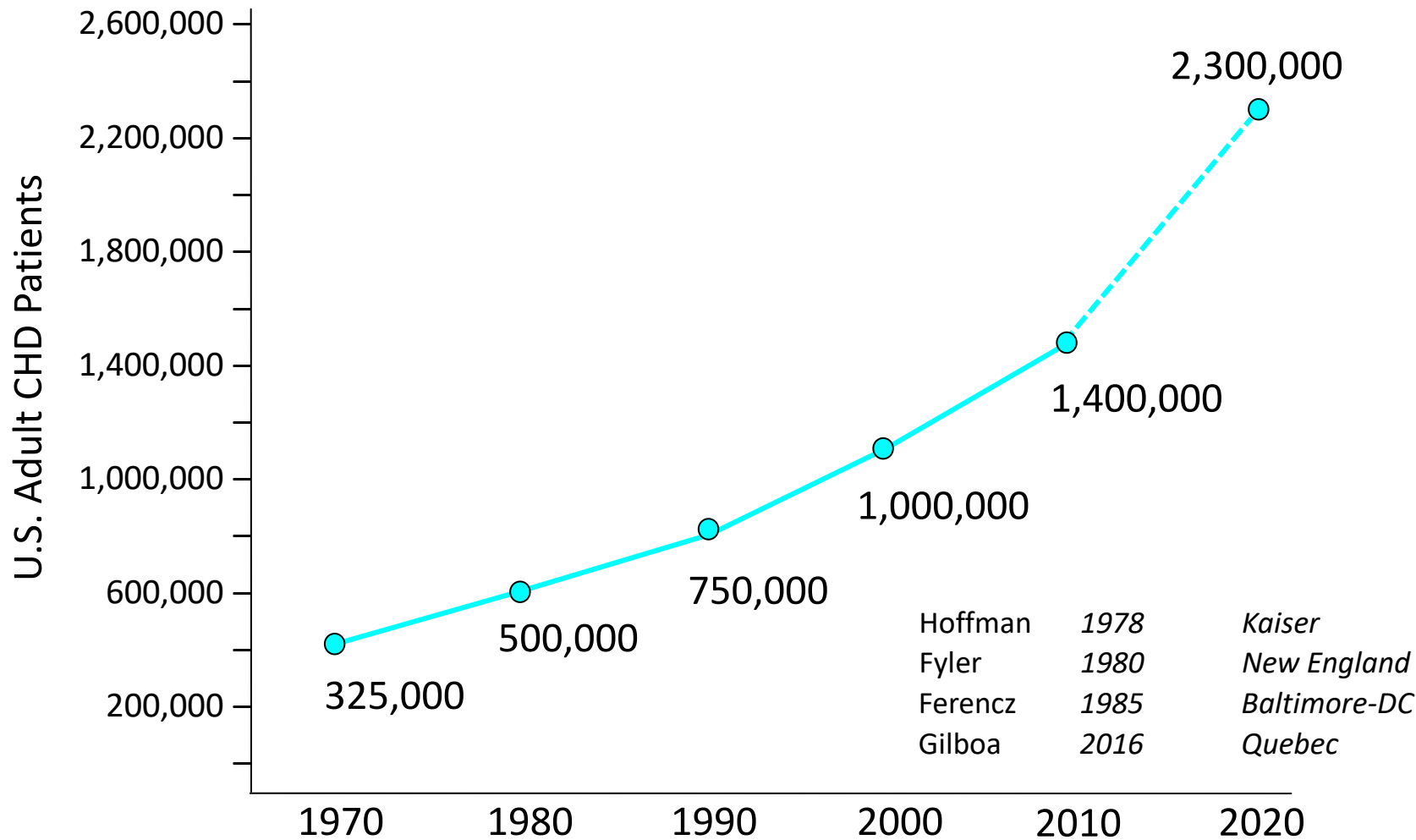
A Report of the American College of Cardiology/American Heart Association Joint Committee
on Clinical Practice Guidelines

*Developed in Collaboration With and Endorsed by the Heart Rhythm Society, International Society for Adult Congenital Heart Disease, and
Society for Cardiovascular Angiography and Interventions*

#1. ACHD Access to Care

- Adults with congenital heart disease benefit from routine care at adult congenital heart disease (ACHD) centers and in collaboration with ACHD cardiologists. Multidisciplinary teams are useful for complex-care decision-making.

A Growing ACHD Population



85% increase in adults living with **complex** CHD between 1985 and 2000

By 2030, it is estimated that 11% of the European ACHD population will be >60 years old

#2. ACHD Anatomic and Physiologic Class

- Patients with anatomic or physiologically moderate or complex ACHD who undergo cardiac or noncardiac procedures are recommended to have an ACHD cardiologist involved in their care to offer expert guidance on procedures, anesthesia, and postprocedural management.

Anatomic / Physiologic Classification

Congenital Anatomy

- **I: Simple Complexity**
VSD, PDA, secundum ASD
- **II: Moderate Complexity**
Anomalous coronary, coarctation, Ebstein anomaly, tetralogy of Fallot, sinus venosus defect
- **III: Great Complexity**
Fontan, systemic right ventricle, unrepaired cyanotic CHD

Physiological Stage

- **Stage A**
No symptoms, no anatomic sequelae
- **Stage B**
Mild residual sequelae (e.g., mild ventricular dilation)
- **Stage C**
Moderate sequelae (e.g., moderate valvular dysfunction)
- **Stage D**
Severe sequelae (e.g., severe hypoxemia)

#3. Emphasis on SBE

- The possibility of endocarditis is important to evaluate in acute or subacute malfunction of bioprosthetic pulmonary valves.
- Endocarditis has been added to the physiological classification system (subacute bacterial endocarditis in the past year is stage D).

SBE Prophylaxis Guidelines

High-risk Patients

Prosthetic valves, including mechanical, bioprosthetic, homografts

Prosthetic material for valve repair, including annuloplasty rings

Prior history of IE

Unrepaired cyanotic CHD

Repaired CHD with residual shunts or valvular regurgitation at or adjacent to the site of the prosthetic patch or prosthetic device

Repaired CHD with catheter-based intervention involving an occlusion device or stent for 6 months

Valve regurgitation due to a structurally abnormal valve in a transplanted heart

High-risk Procedures

Dental work including abscess drainage, extractions, or routine cleaning

NOT NEEDED FOR:

Procedures in GI or GU tract, vaginal or cesarean delivery, respiratory tract procedures

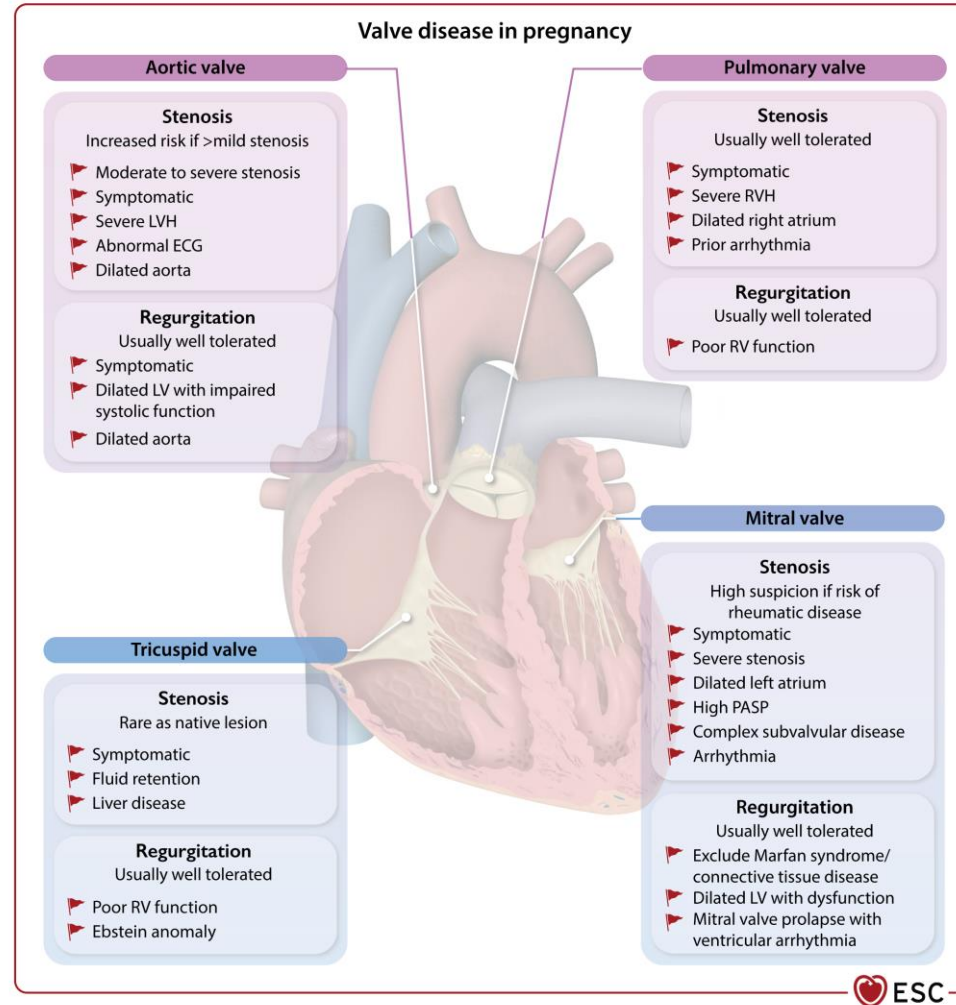
#4. Pregnancy Risk Stratification

- Most pregnant patients with ACHD can undergo vaginal delivery safely, with appropriate risk stratification and monitoring.

2025 ESC Guideline Recommendations

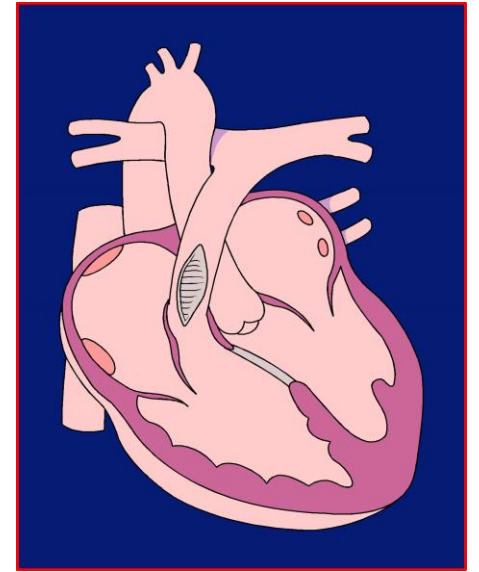
Recommendations	Class	Level
Vaginal delivery is recommended in most women with ACHD.	I	B
It is recommended that all women with Fontan circulation who wish to become pregnant receive counselling from the Pregnancy Heart Team regarding the high risk of pregnancy-related adverse events.	I	C
It is recommended that women with a systemic RV (Mustard/Senning or congenitally corrected TGA), in NYHA class III/IV, systemic ventricular dysfunction (EF <40%), or severe TR wishing to become pregnant are counselled by the Pregnancy Heart Team regarding the high risk of pregnancy-related adverse events.	I	C
In women with significant haemodynamic lesions, discussion about guideline-directed interventions is recommended prior to pregnancy.	I	C

Valvular Heart Disease in Pregnancy



#5. Repaired Tetralogy of Fallot (TOF)

- Referral for pulmonary valve replacement in patients with repaired tetralogy of Fallot, according to right ventricular end-systolic volume criteria ($>80 \text{ mL/m}^2$) and other metrics rather than end-diastolic volume
- New approaches to arrhythmia management, including ablation of ventricular tachycardia



#5. Updated Recommendations for TOF

1

B- NR

4. In adults with repaired TOF and native RVOT anatomy being considered for transcatheter pulmonary valve replacement, cardiac CT is recommended to determine anatomic suitability.^{4,5}

2a

B-NR

5. In adults with repaired TOF and moderate risk for future sustained ventricular tachycardia and/or SCD, invasive electrophysiology evaluation is reasonable to inform clinical management.⁶⁻⁸

1

B-NR

9. In symptomatic adults with repaired TOF and moderate or greater pulmonary valve dysfunction,* pulmonary valve replacement (surgical or transcatheter) is recommended for relief of symptoms.¹⁵

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B-NR

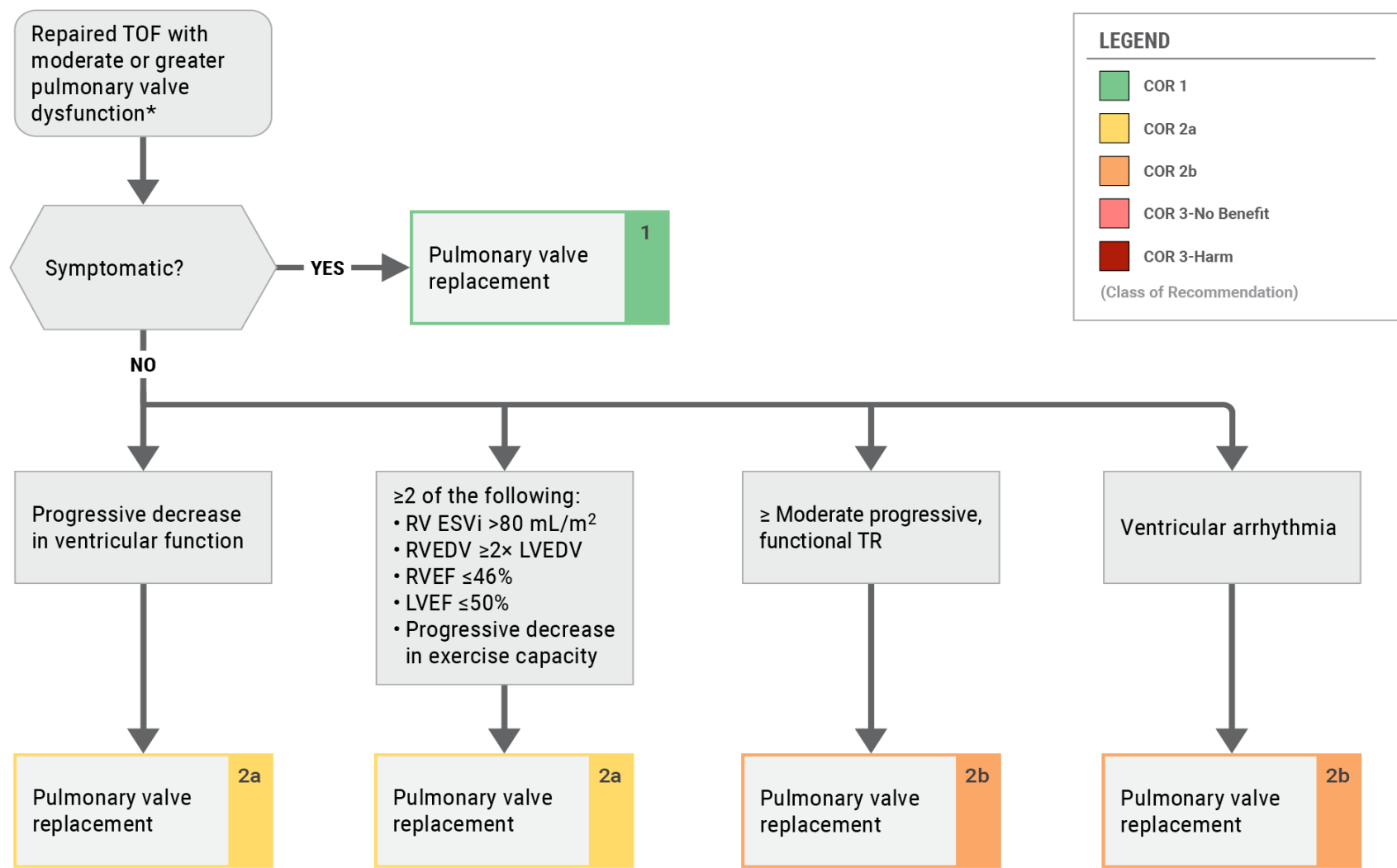
11. In adults with repaired TOF and high risk (by clinical markers and/or multivariable score†) for SCD, placement of an ICD is reasonable to prevent SCD.¹⁷⁻¹⁹

2a

B-NR

12. In adults with repaired TOF and appropriate ICD therapies for monomorphic ventricular tachycardia, adjunctive catheter ablation is reasonable to reduce ventricular tachyarrhythmia burden.^{20,21}

Indications for Pulmonary Valve Replacement in rTOF



*Pulmonary valve dysfunction defined as moderate PR (CMR-derived RF ≥25%) or RVSP >2/3 systemic pressure due to RVOTO.

CMR indicates cardiovascular magnetic resonance; ESVi, end-systolic volume index; LVEDV, left ventricular end-diastolic volume; LVEF, left ventricular ejection fraction; PR, pulmonary regurgitation; RF, regurgitant fraction; RV, right ventricular; RVEDV, right ventricular end-diastolic volume; RVEF, right ventricular ejection fraction; RVOTO, right ventricular outflow tract obstruction; RVSP, right ventricular systolic pressure; TOF, tetralogy of Fallot; and TR, tricuspid regurgitation.

#6. Atrial Septal Defect Closure

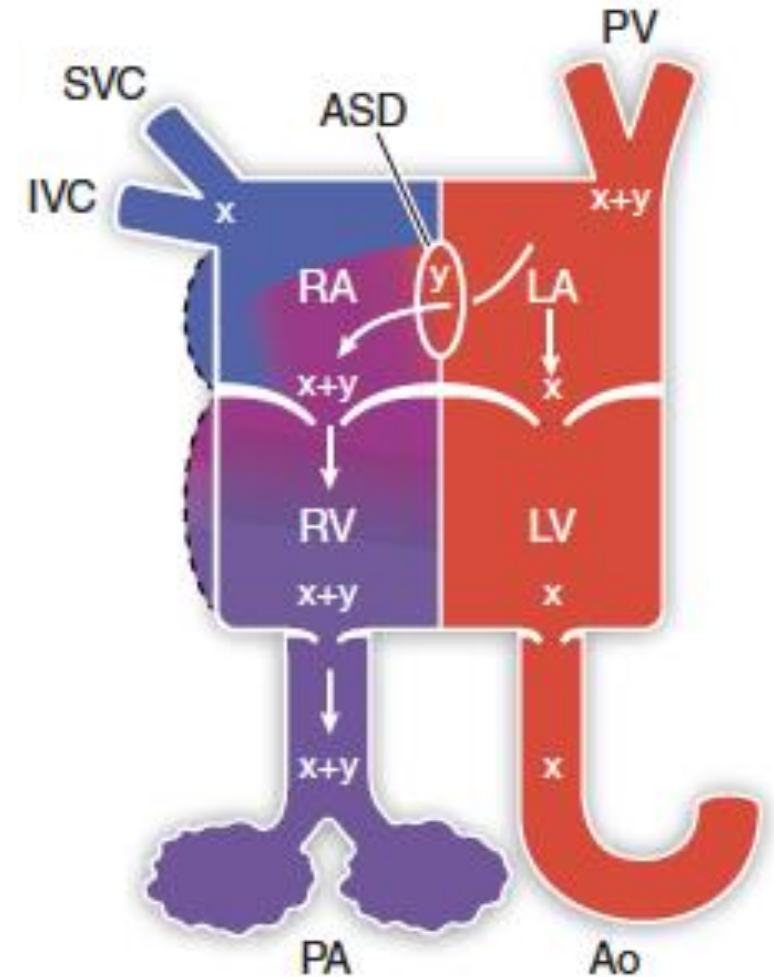
- Strategies for patients with secundum atrial septal defect and pulmonary arterial hypertension now include recommendations for closure for many patients with a significant left-to-right shunt and pulmonary vascular resistance ≤ 2 Wood units or >2 to <5 Wood units.

Atrial Septal Defect (ASD)

- Left-to-right shunt causing pulmonary overcirculation and R heart dilation
- Palpitations, dyspnea, exercise intolerance
- Exam findings: Fixed, split S2
- EKG: RBBB, RAD*

*for secundum ASDs (most common)

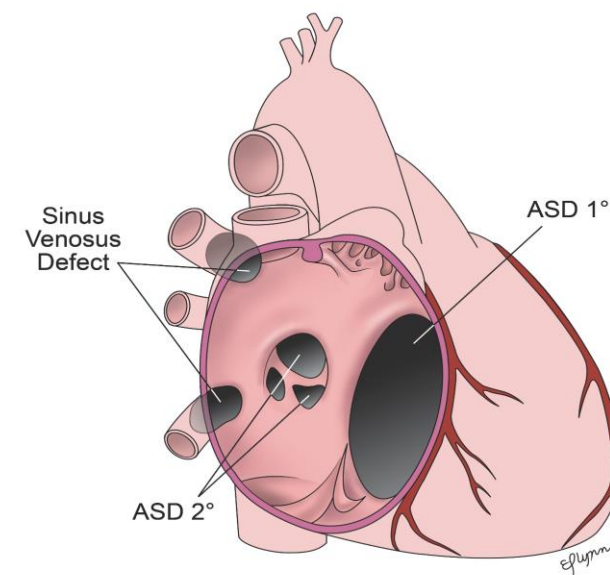
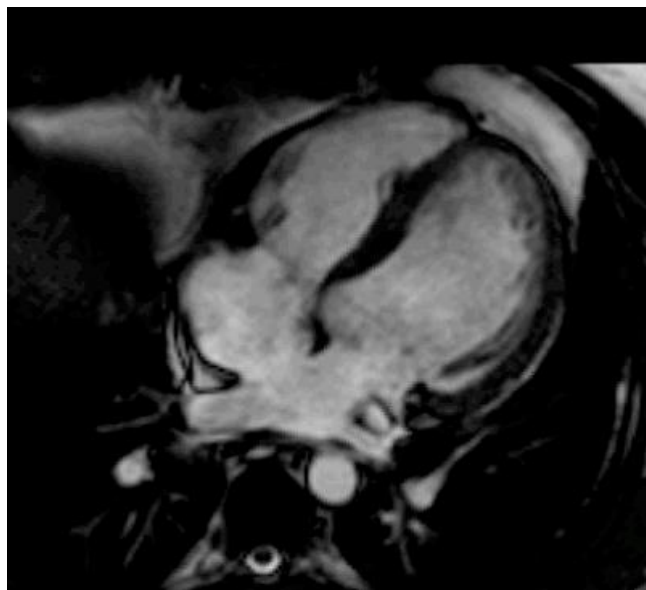
If LAD -> think primum ASD



Types of ASD

- **Secundum ASD:** most common
- **Primum ASD:** on the spectrum of AVC defects; associated with a cleft mitral valve
- **Sinus venosus defect*:** associated with partial anomalous pulmonary venous return

*Not in the atrial septum



Expected Chamber Enlargement with Cardiac Shunts

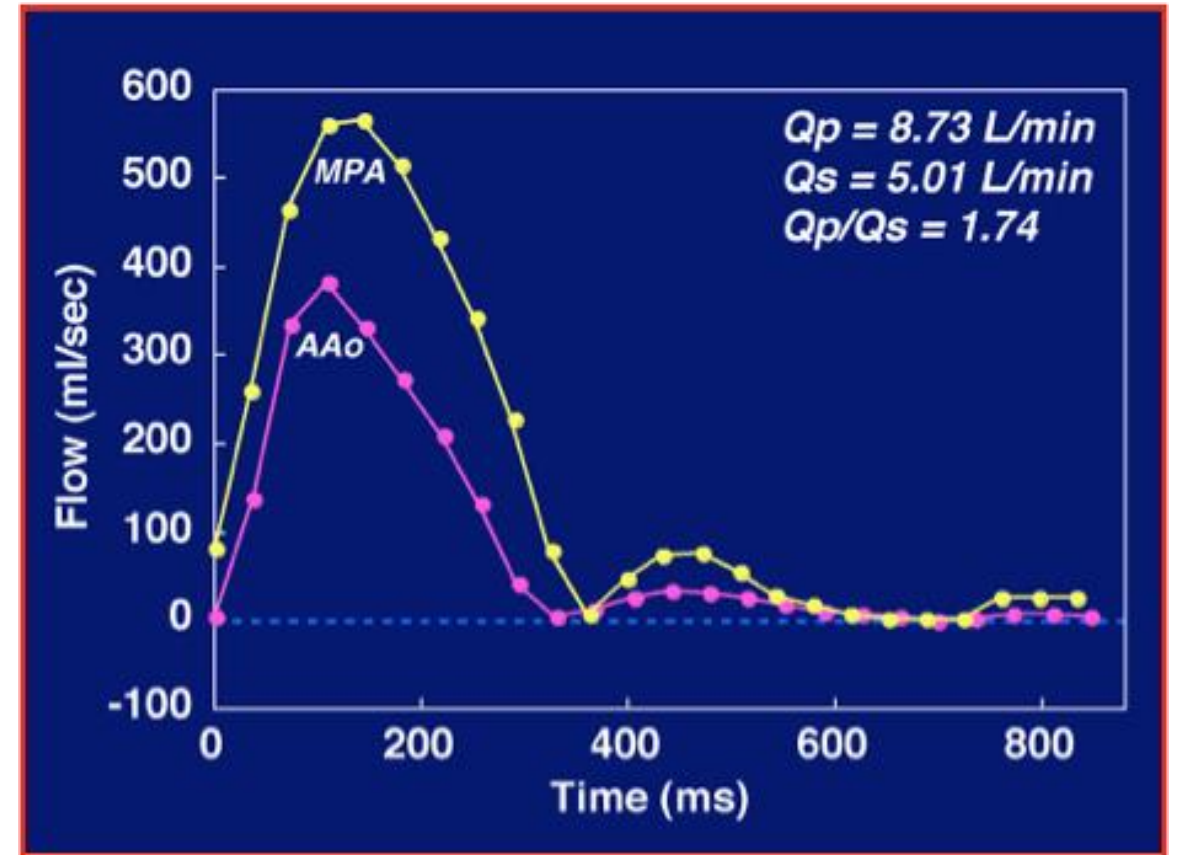
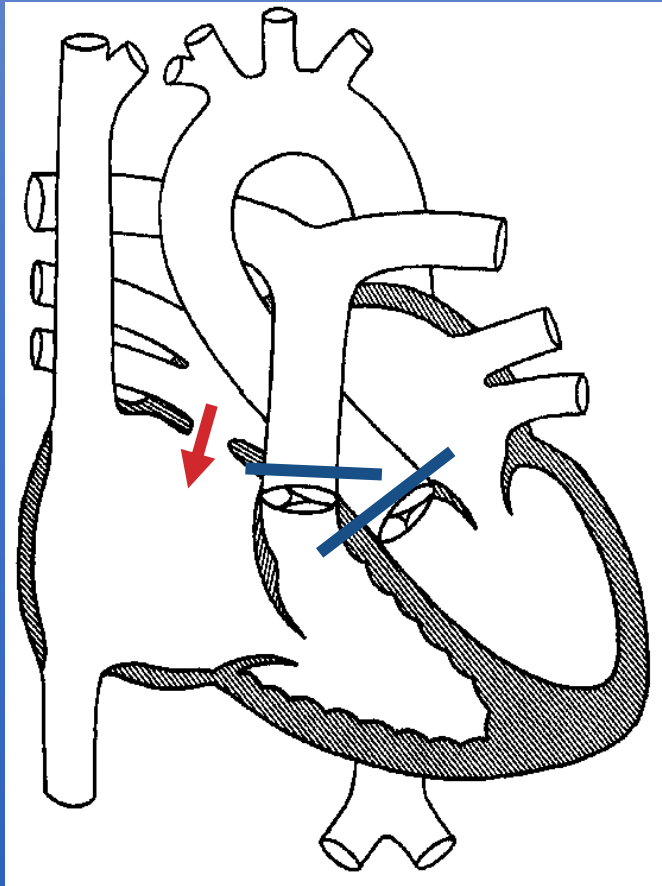
To dilate a chamber, it needs to fill during diastole

VSD ejects *through* the right ventricle (and into the PA) during systole

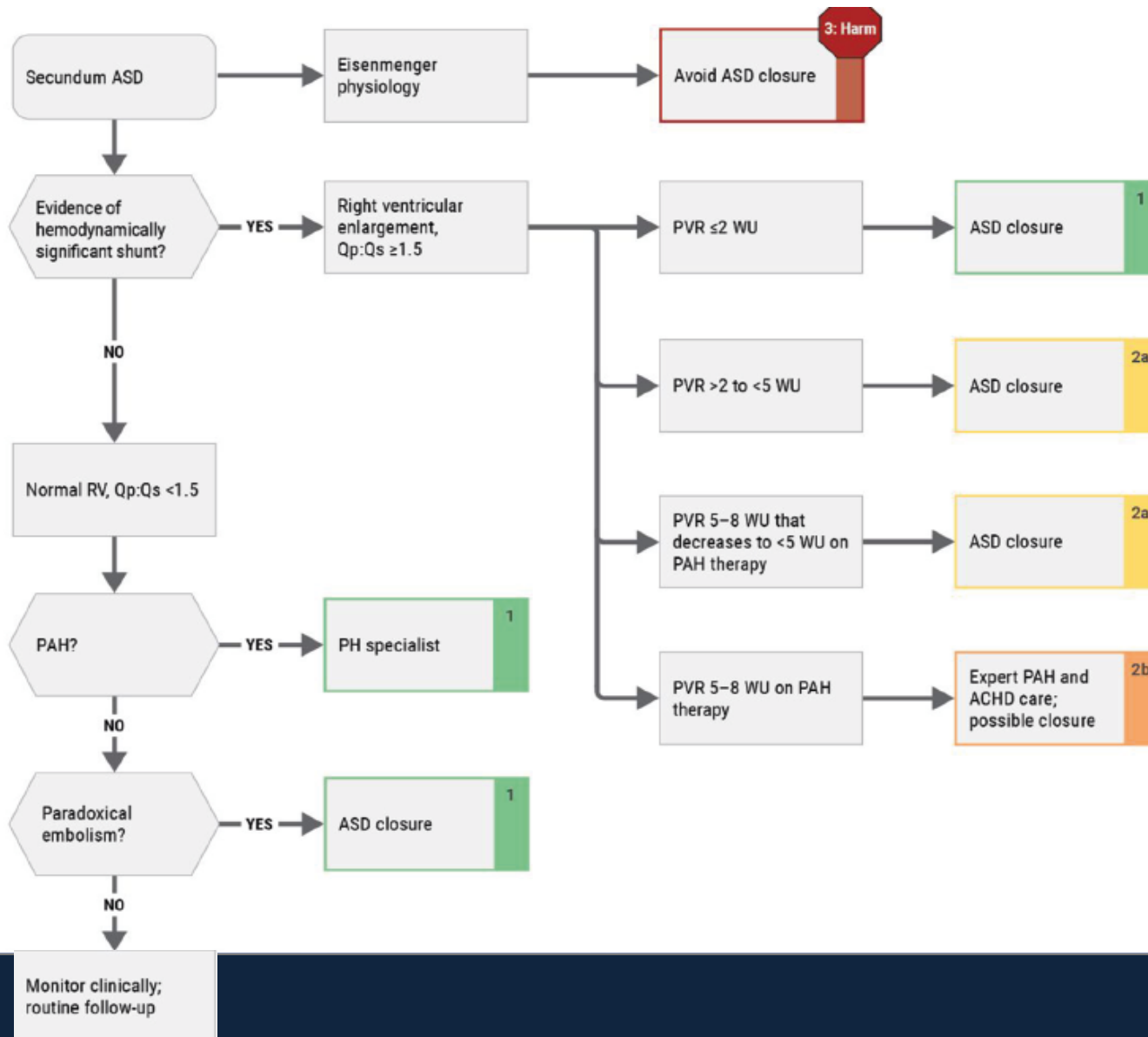
Shunted blood returns to the left heart and fills (and dilates) during diastole

Shunt	RA	RV	PA	LA	LV	Aorta
ASD	+	+	+			
VSD		+/-	+	+	+	
PDA			+	+	+	+

ASD: Quantification of Shunt



Shunt Closure

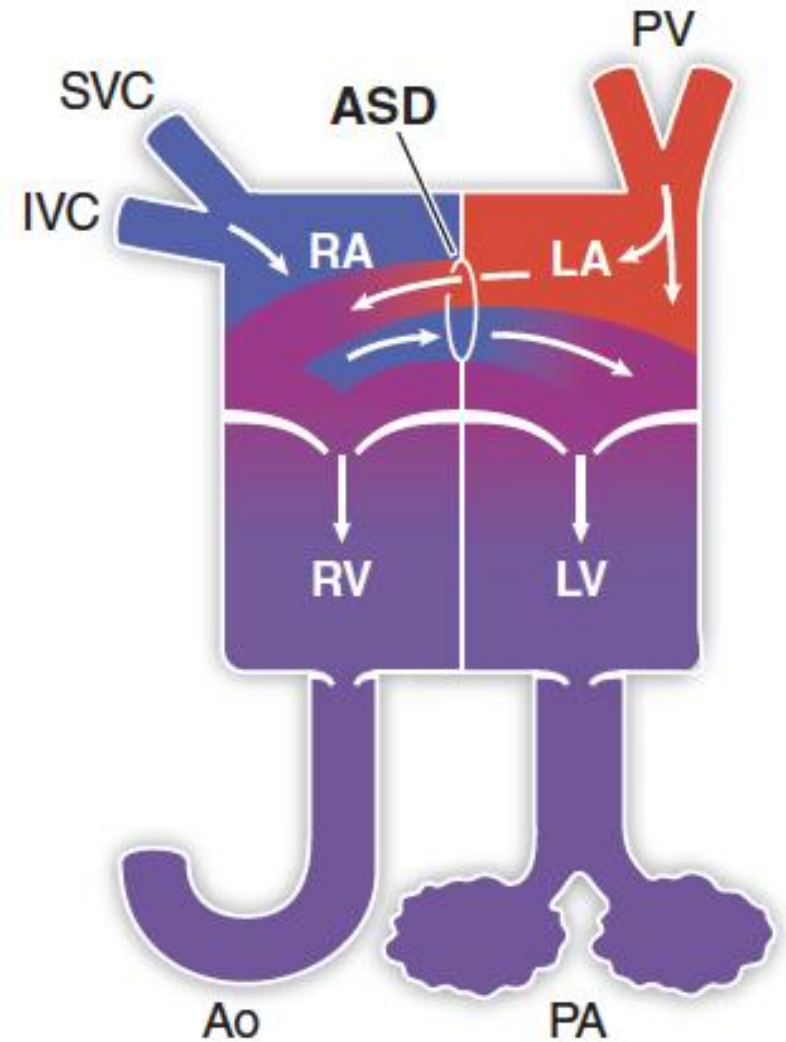
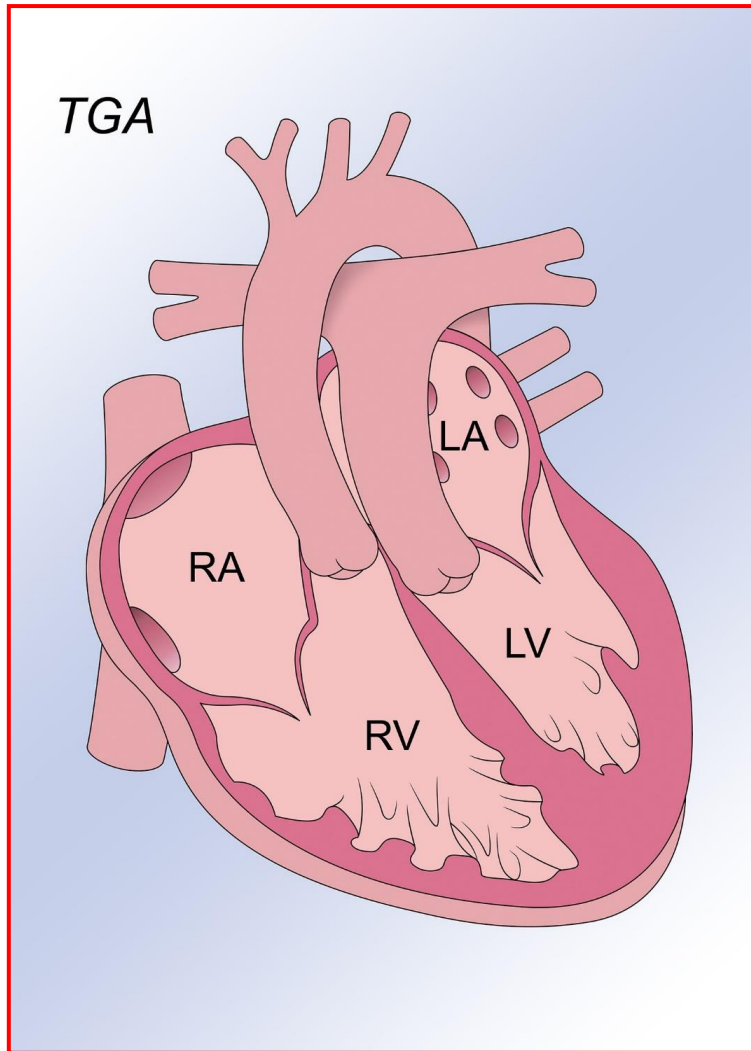


- Criteria changed from PA systolic pressure thresholds to PVR
- Management changed to include treat-to-close strategies

#7. Rhythm Control Is Preferred in Complex CHD

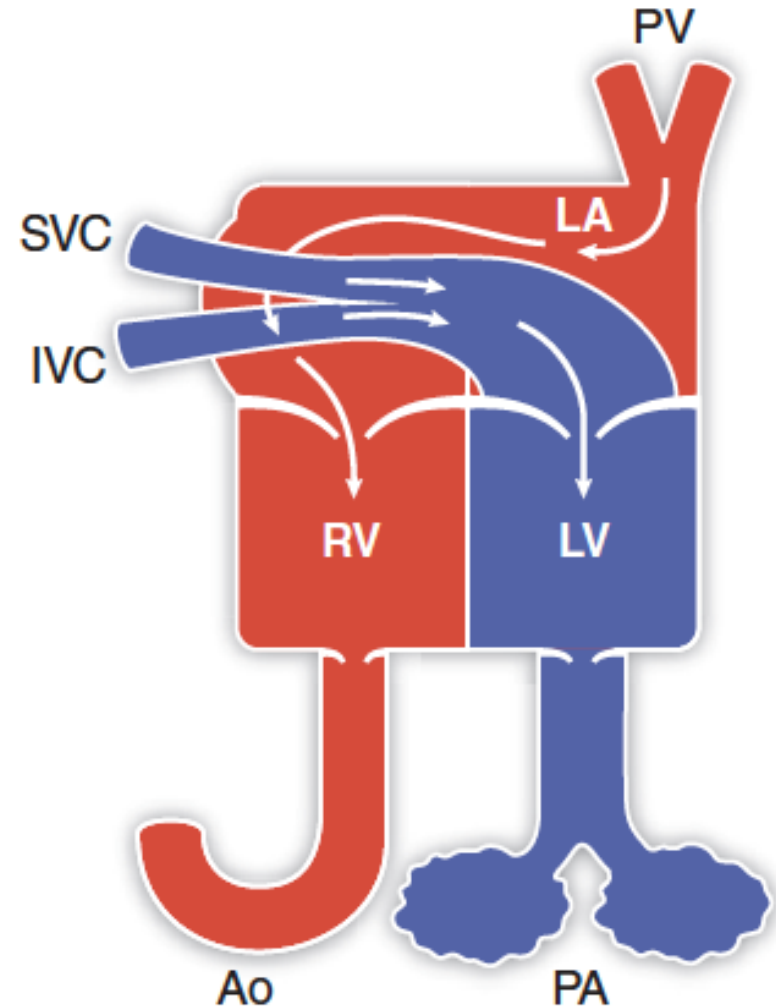
- Rhythm control is typically preferred over rate control for atrial arrhythmias in complex patients, such as those with a systemic right ventricle or Fontan circulation.

Transposition of the Great Arteries (TGA)



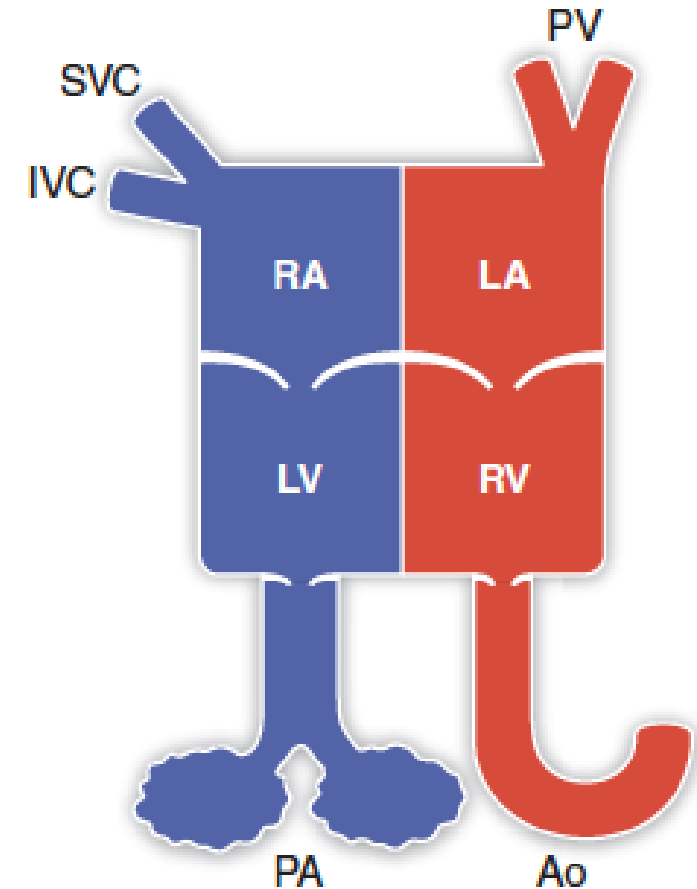
TGA s/p Atrial Switch (Older Patients)

- Senning procedure (1957)
- Mustard procedure (1963)
- Redirects blood flow to allow systemic venous return to get to the lungs
- Long-term complications:
 - **Systemic RV dysfunction**
 - Tricuspid regurgitation
 - Atrial baffle complications
 - Atrial arrhythmias



Congenitally Corrected Transposition of the Great Arteries (cc-TGA)

- AV and VA discordance
- Symptoms depend on associated lesions:
 - ASD
 - Tricuspid valve dysplasia $\pm$ TR
 - Pulmonary stenosis
- 2% incidence of **complete heart block** per year in adulthood
- May be diagnosed in adulthood

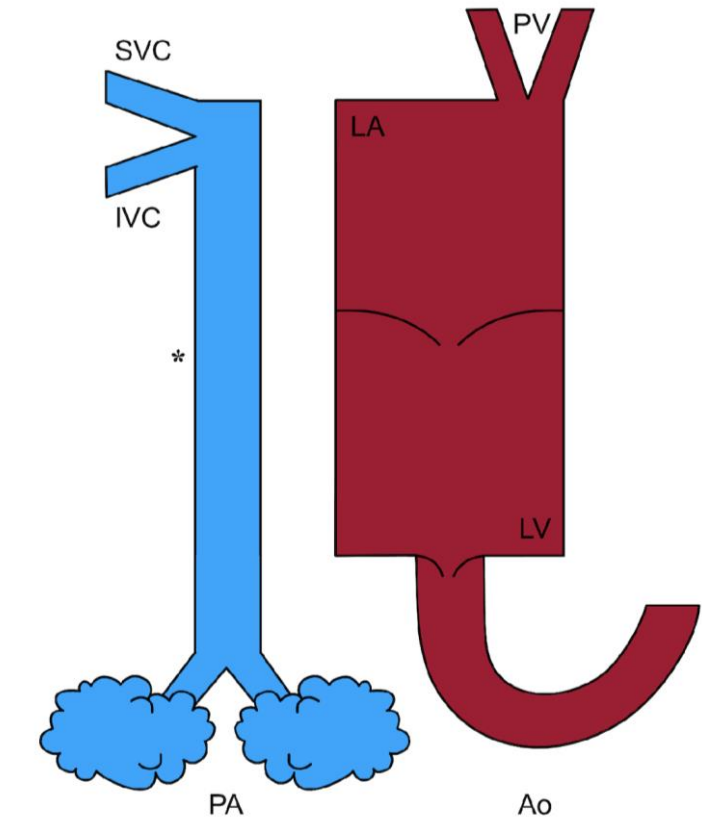


#8. Intensified Heart Failure Therapies

- New recommendations on guideline-directed medical therapy for heart failure in patients with ACHD include people with a systemic right or left ventricle and discussions of pacing strategies for a systemic right ventricle and Fontan circulation.

Fontan Physiology

- Complications:
 - Arrhythmias
 - Ventricular dysfunction
 - Valvular regurgitation
 - Thromboembolic events
 - Protein-losing enteropathy
 - Renal dysfunction
 - Fontan-associated liver disease
 - Low cardiac output, hypoxemia



#9. Eisenmenger Syndrome

- Patients with Eisenmenger syndrome can be treated with pulmonary vasodilators, using either phosphodiesterase-5 inhibitors or endothelin receptor antagonists as initial therapy.

Eisenmenger Syndrome

Deoxygenated blood from right side of the heart shunts over to the left side



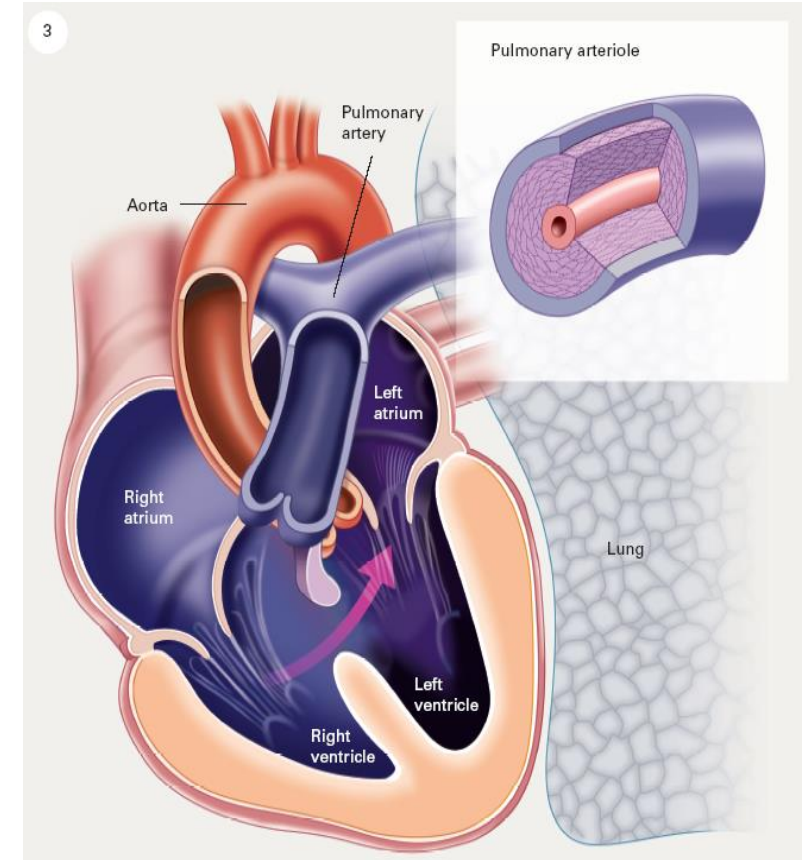
Takes blood from the right side of the heart and pulmonary blood flow decreases



Since pulmonary blood flow decreases, no increase in ventricular volumes



Systemic arterial desaturation (cyanosis)



PVR ↑'s: shunt reverses:
R-to-L → Eisenmenger
syndrome: ↑ cyanosis

#10. Fontan-Associated Liver Disease

- Fontan screening for liver disease includes recommendations for at least annual imaging and laboratory evaluation, including alpha-fetoprotein, and at least 1 consultation with a hepatologist.

ACHD Co-Morbidities

Hepatorenal dysfunction ~30%

- 10% of adults with right-sided heart lesions have severe HRD (Egbe, *Can J Cardiol* 2022)

Atherosclerotic Risk Factors ~50%

- 47% of ACHD patients have >1 ASCVD risk factor (Egbe, *JACC Advances* 2022)

Neuropsychological Disorders ~3-fold increased risk

- Adults with repaired TOF have a 3-fold higher risk of mental illness (Hsu, *J Clin Psych* 2021)

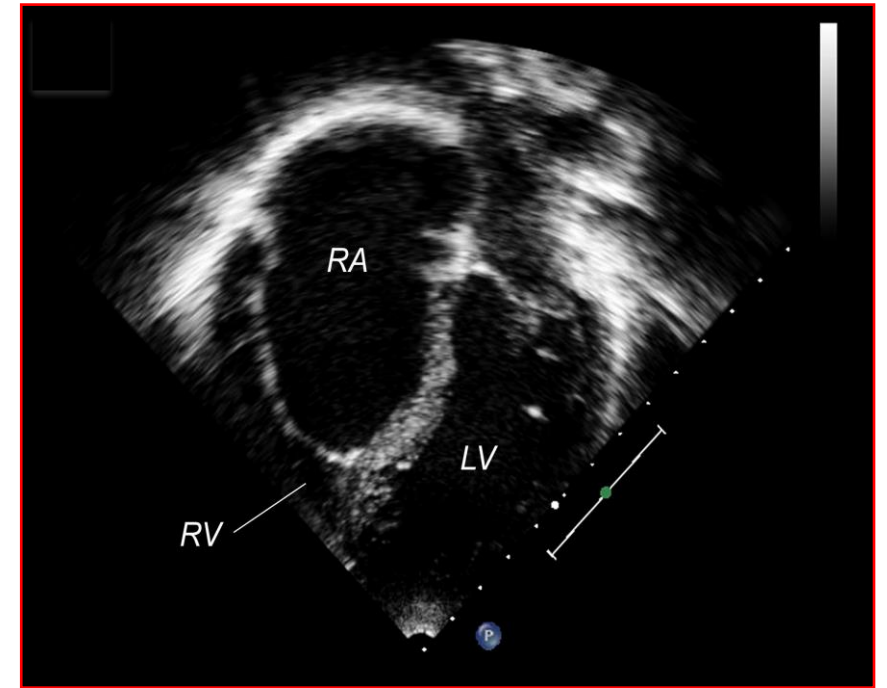
Malignancy ~2-fold increased risk

- Adults with CHD have ~2x greater prevalence of malignancy (Gurvitz, *Am J Cardiol* 2016)

ACHD Question #1

A 21-year-old person presents with progressive shortness of breath and palpitations. An echocardiogram is performed (still frame shown). Which ECG finding is most likely in this person?

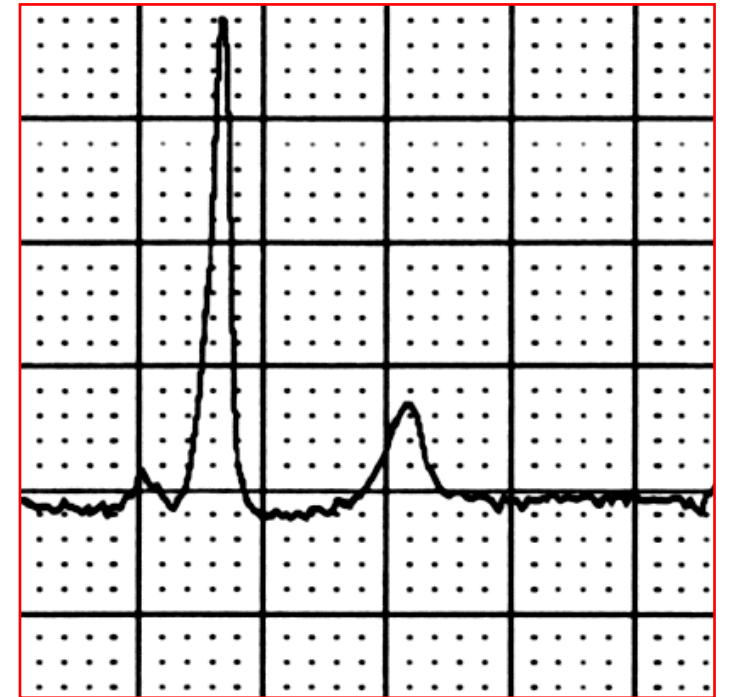
- a) Prolonged PR interval
- b) Short PR interval with delta wave
- c) Prolonged QTc
- d) Atrial fibrillation
- e) Complete heart block



ACHD Answer #1

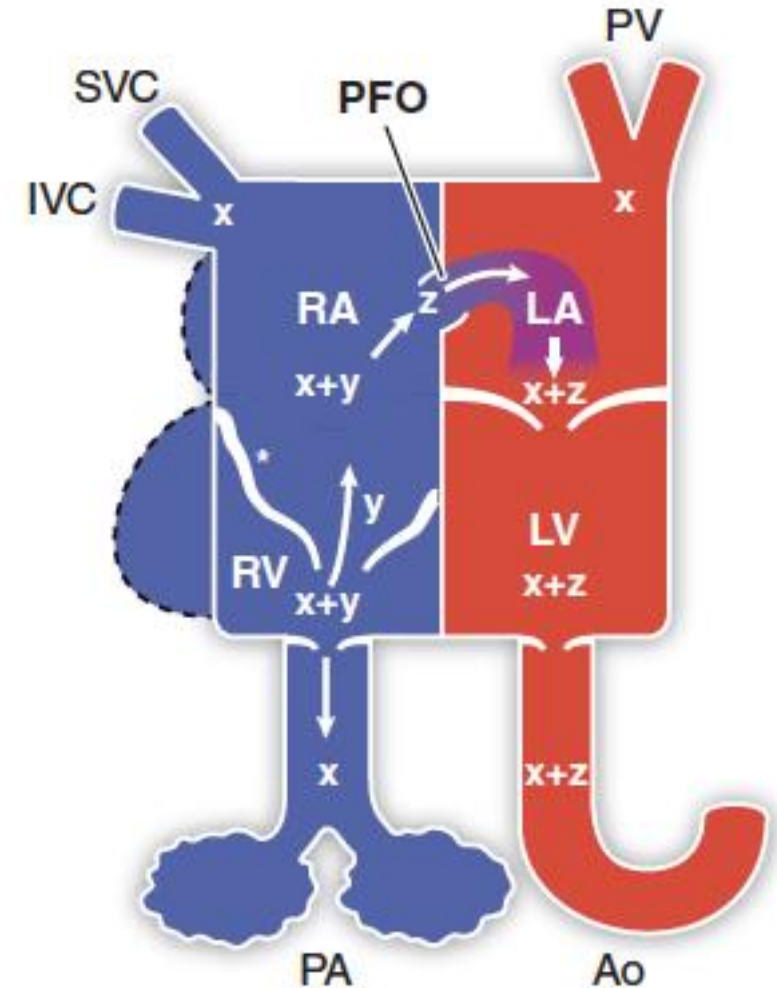
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Ebstein Anomaly

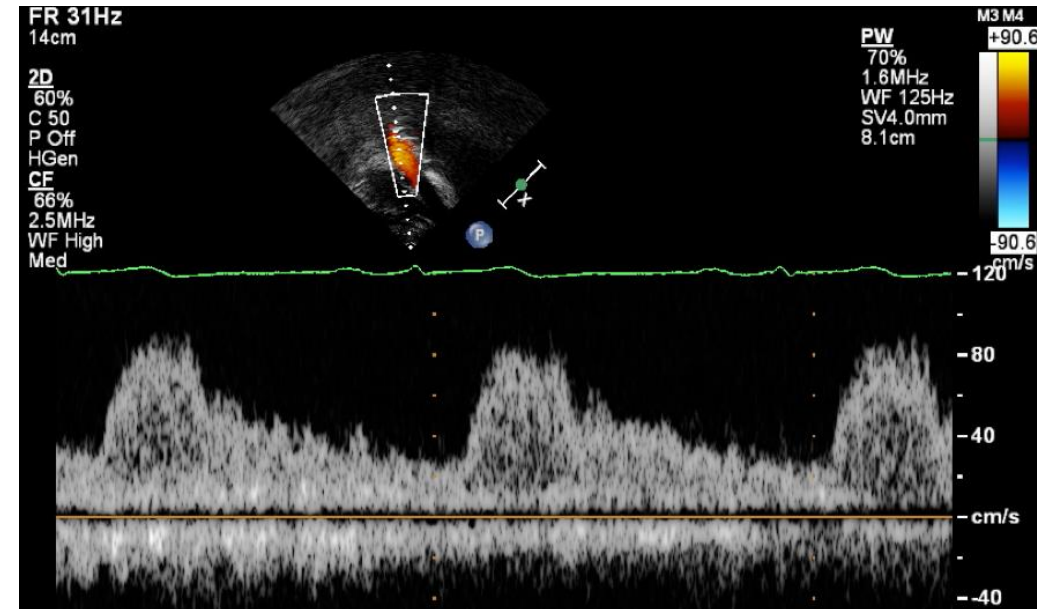
- Failure of delamination of the septal leaflet of the TV
- Results in “atrialized” portion of the RV
- Symptoms depend on severity of TR and associated lesions
 - Signs of R heart failure
 - Cyanosis (if ASD/PFO)
- Exam: Widely split S1 “sail sign”
- 20% of patients have WPW



ACHD Question #2

A 28-year-old is seen for evaluation of headaches. On exam, BP is 172/94 mmHg. A 3/6 systolic ejection murmur is heard at the left sternal border. An echocardiogram is performed which demonstrates normal left ventricular function, left ventricular hypertrophy, and no significant valve disease. Abdominal aorta Doppler is shown here. What is the most common additional physical examination finding with this condition?

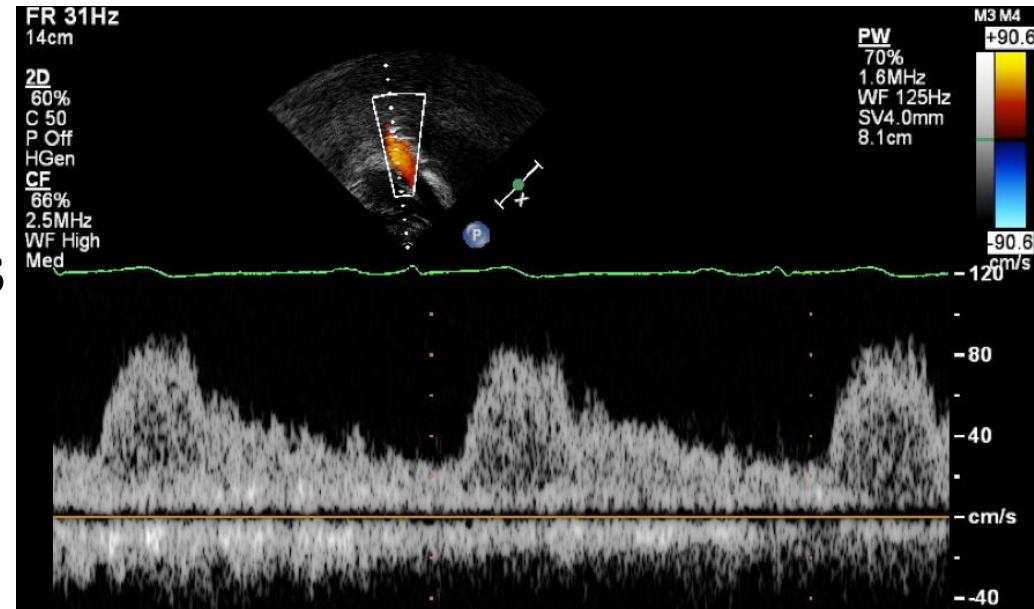
- a) Clubbing of the fingernails
- b) Bifid uvula
- c) Diminished pulses in the lower extremities
- d) An abdominal bruit



ACHD Answer #2

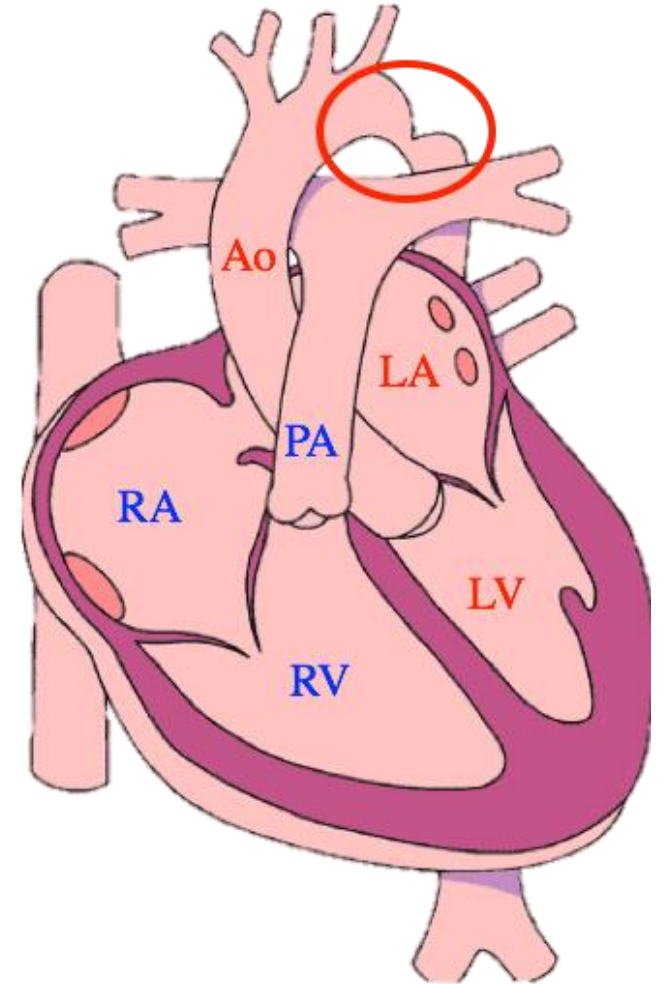
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Coarctation of the Aorta

- Presents with hypertension
- Exam: Brachial-femoral pulse delay and diminished LE pulses
- Associations:
 - Bicuspid aortic valve
 - Ascending aortic dilation
 - Aneurysms
 - LVH
 - Coronary artery disease
 - Cerebral aneurysm



MOC Reflective Statement

- **Updated Guidelines for ACHD Care incorporate evidence to emphasize.**
 - Coordinated care with ACHD expertise
 - Anatomic and Physiologic Stages
 - Risks associated with endocarditis
 - Pregnancy risk stratification: Vaginal delivery safe in majority of cases
 - Pulmonary valve replacement in repaired TOF symptomatic patients
 - ASD closure strategies including treatment of elevated PVR
 - Rhythm (over rate) control for complex CHD
 - Intensified heart failure therapies including pacing for complex CHD
 - Pulmonary vasodilator therapy for Eisenmenger syndrome
 - Recognition of co-morbidities in adults with CHD

References

- Gurvitz M et al. 2025 ACC/AHA/HRS/ISACHD/SCAI Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol* 2026 Feb 24;87(7):822-976. doi: 10.1016/j.jacc.2025.09.006.
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Thank You from the BACH Team



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