

Acyanotic Congenital Heart Diseases

May30th 2020

Iowa ACC – Lecture Series

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Disclosures

- *I have no relevant financial disclosures or conflicts of interest.*

Outline of Today's Talk

- Introduction of Acyanotic Congenital Heart Disease
- Specific Types Cardiac Lesions discussed today:
 - ASD
 - VSD
 - AVSD
 - PDA
 - Aortic Stenosis
 - Pulmonary Stenosis
 - Ebstein's Anomaly
 - Anomalous Pulmonary Veins

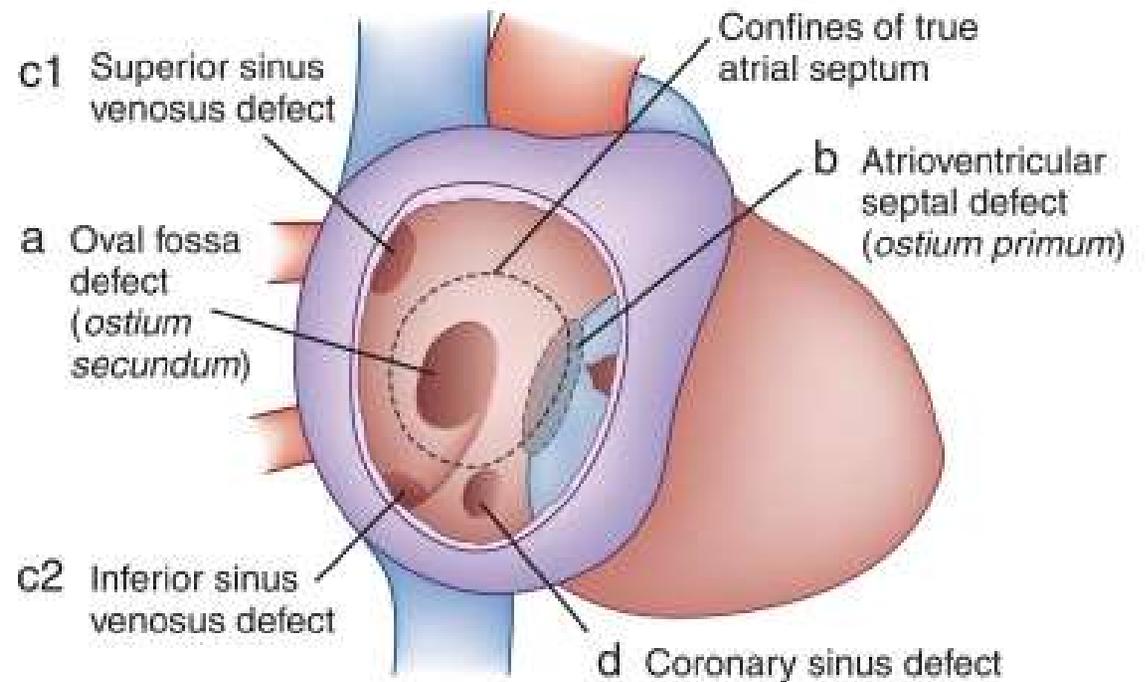
Acyanotic Congenital Heart Disease

- Increased pulmonary blood flow: Left to right shunt lesions
 - Ventricular septal defects
 - Atrial septal defects
 - Atrioventricular canal defect
 - Patent ductus arteriosus
 - Partial Anomalous Pulmonary Venous Return (PAPVR)
- Normal pulmonary blood flow: Left, right obstructive lesions
 - Aortic stenosis
 - Pulmonary stenosis
 - Coarctation of the aorta*

Atrial Septal Defects

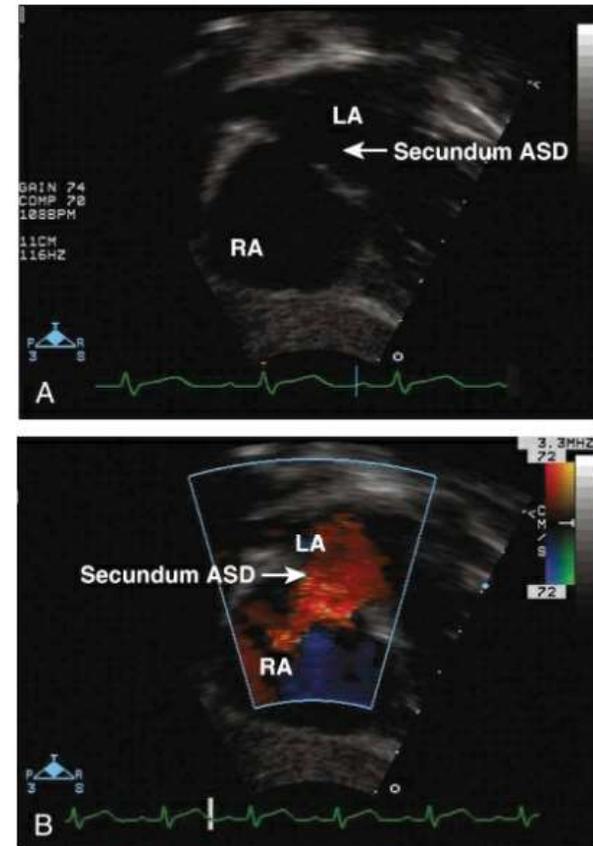
Atrial Septal Defects - Types

- Patent foramen ovale (PFO).
- Secundum atrial septal defect (ASD).
- Primum ASD.
- Sinus venosus ASD.
- Coronary sinus (CS) ASD.



Secundum Atrial Septal Defect

1. Most common type of ASD - 70% of ASDs are secundum defects.



Clinical Features – Presentation in Adults

- Dyspnea on exertion and palpitations.
- Abnormal cardiac exam, observation of cardiomegaly on routine chest imaging, or incidentally during cardiac imaging.
 - RV heave, Ejection systolic murmur,
 - Fixed split S2
 - Diastolic rumble (Increased flow across tricuspid valve)
 - Right atrial dilatation – atrial fibrillation
- Alternatively, patients may present with stroke or systemic ischemic event due to a paradoxical embolism.

ASD - ECG

- Incomplete RBBB
- Right axis deviation due to RV hypertrophy



Atrial Septal Defect - CXR



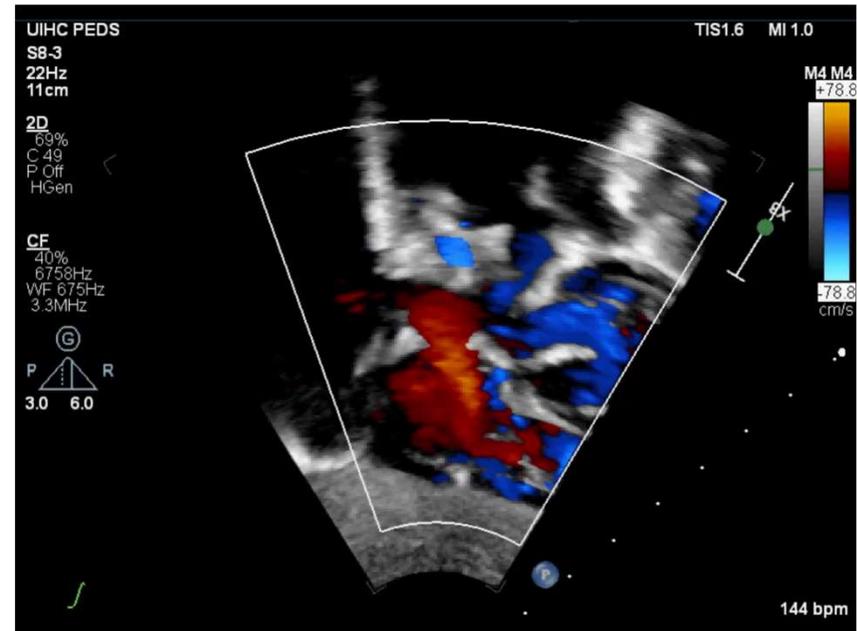
2 year old



22 year old

- Right atrial and right ventricular enlargement
- Prominent pulmonary arteries
- Increased pulmonary plethora

ASD - Echo



Evaluate

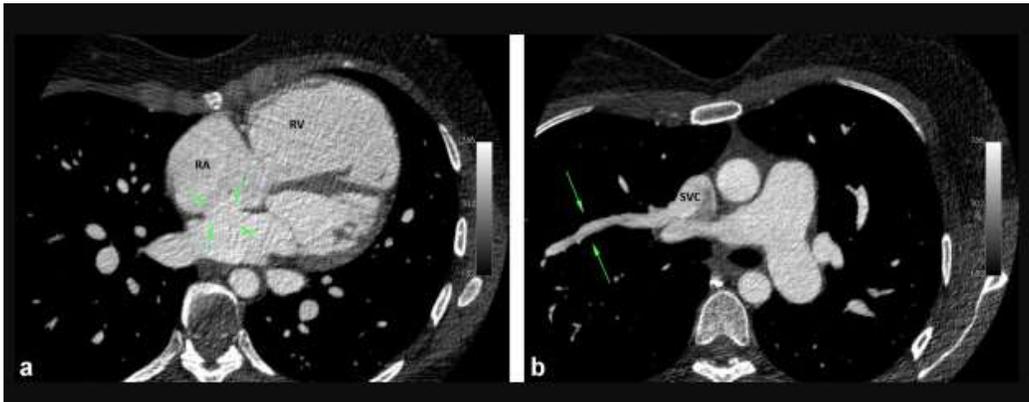
- size and direction of shunting
- Chamber size
- Right sided pressures
- evaluate ASD Rims

Cardiac MRI/Cardiac CT



Cardiac MRI performed in a 60-year-old woman with exertional dyspnea

- Size of ASD
- Qp:Qs
- Pulmonary vein anatomy
- RV volume



Cardiac CT performed in a 38-year-old woman. Note the ASD (arrows) on the axial view. Incidental finding of abnormal partial venous return from the right upper lobe (b, arrows) into the superior vena cava (SVC).

Natural History

- Right Atrial dilatation – atrial fibrillation
- Earlier the ASD is closed less likely to have atrial fibrillation or paradoxical embolism.

Management – Class I recommendation

In adults with isolated secundum ASD causing:

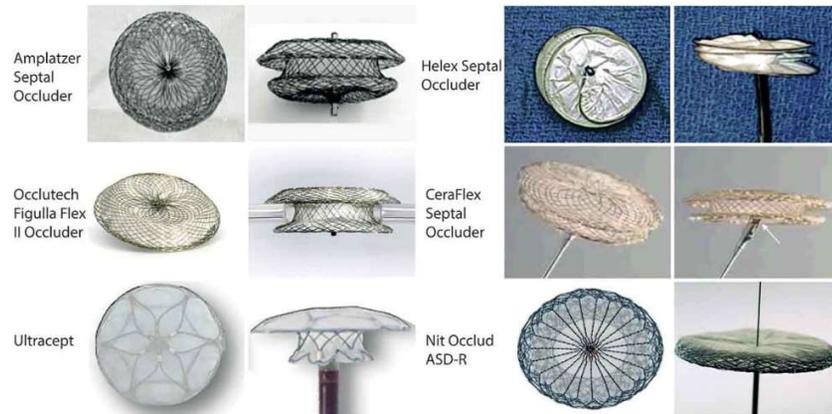
- impaired functional capacity, right atrial and/or RV enlargement
- net left to-right shunt [Qp:Qs] $\geq 1.5:1$) without cyanosis at rest or during exercise,

Transcatheter or surgical closure to reduce RV volume and improve exercise tolerance is recommended, provided that systolic PA pressure is less than 50% of systolic systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance.

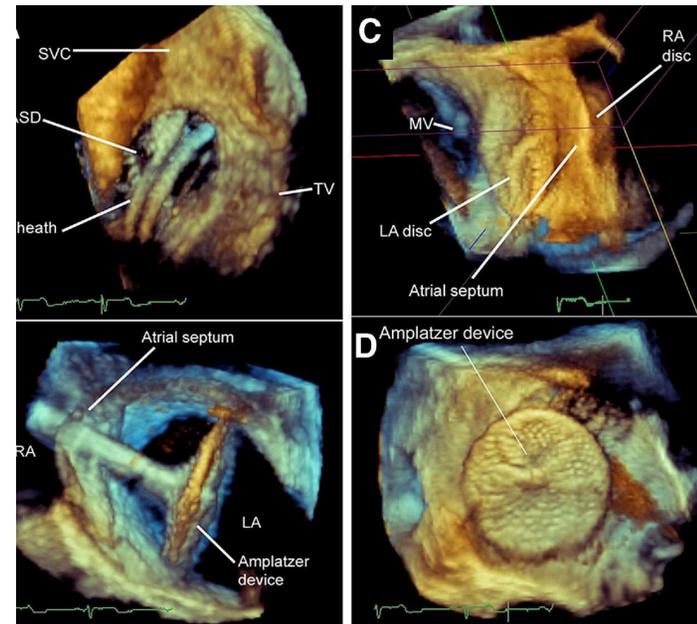
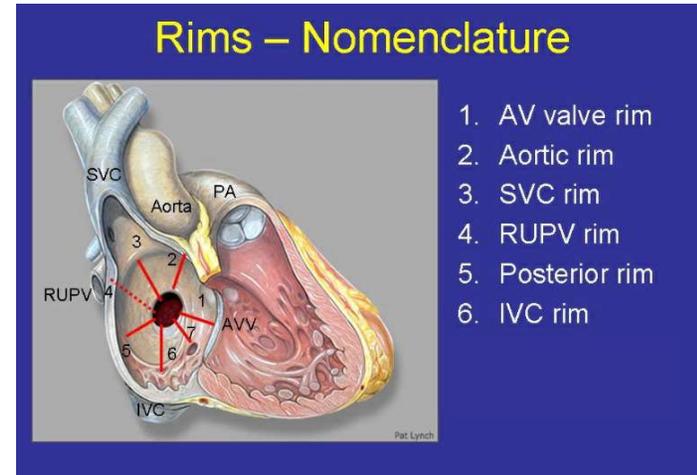
Transcatheter Closure

Adequate Rims – Transcatheter device closure.
Currently approved devices include:

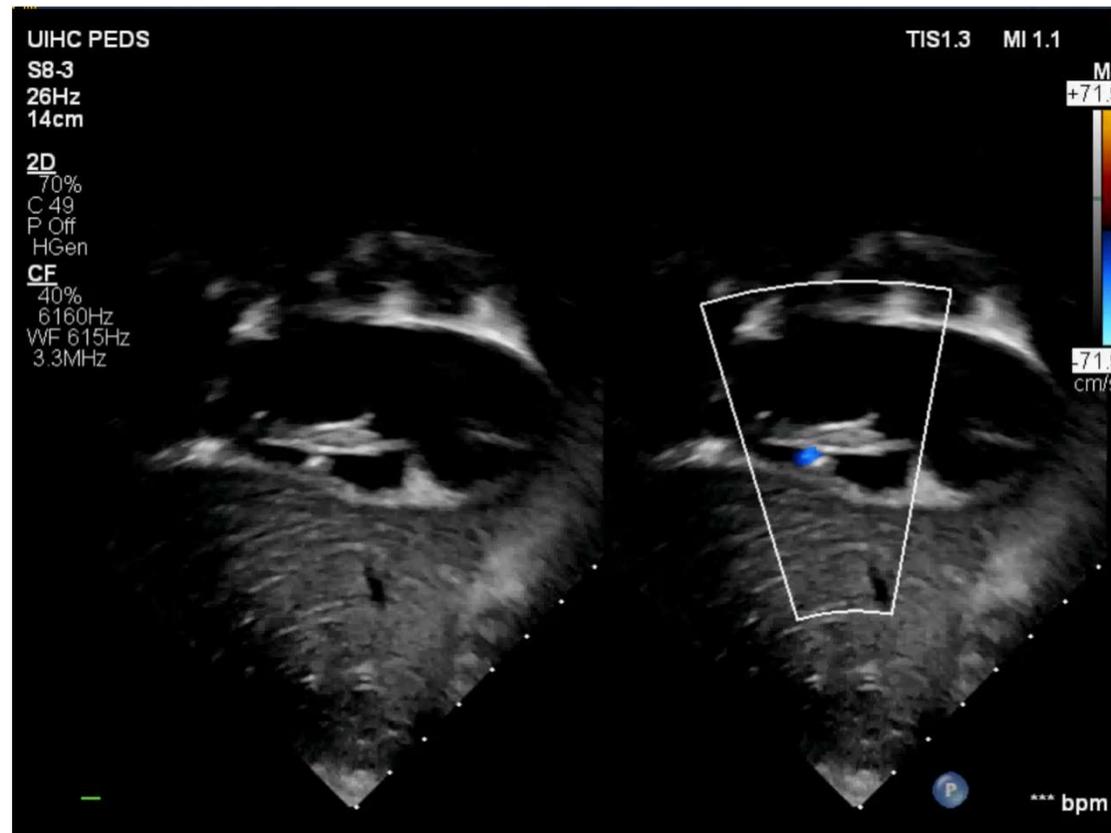
- Amplatzer septal occluder
- Gore Helex
- CardioSEAL devices



Not amenable to cath closure – Surgical closure



Secundum ASD – Post GORE Cardioform Device Closure



Atrioventricular Septal Defect

Atrioventricular Septal Defect

- AVSDs represent about 4% to 5% of congenital heart defects.
- Deficient atrioventricular (AV) septation.
 - Associated with Trisomy 21.
 - Usually seen a cleft in the anterior leaflet of the mitral valve, although primum defects can occur in isolation.
 - Both AV valves (AVVs) appear at the same level.
 - The tricuspid valve (TV) will not be apically displaced.

Types of AV Septal Defects

1. Complete AV Septal Defect
2. Partial AV Septal Defect
3. Transitional AV Septal Defect

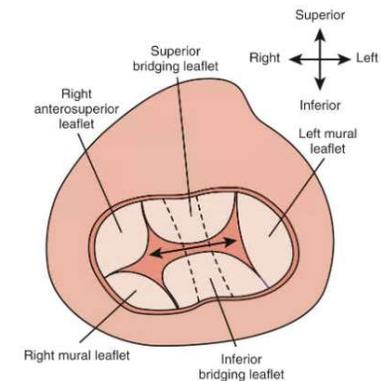
Complete AV Canal

A complete AVSD has a common AV junction,

- a primum ASD,
- an IVSD, and
- a common AVV.

The primum ASD is anterior and inferior to the fossa ovalis, adjacent to the AVVs.

The AVV consists of five leaflets: superior and inferior bridging leaflets, a left mural leaflet, a right mural leaflet, and a right anterosuperior leaflet.

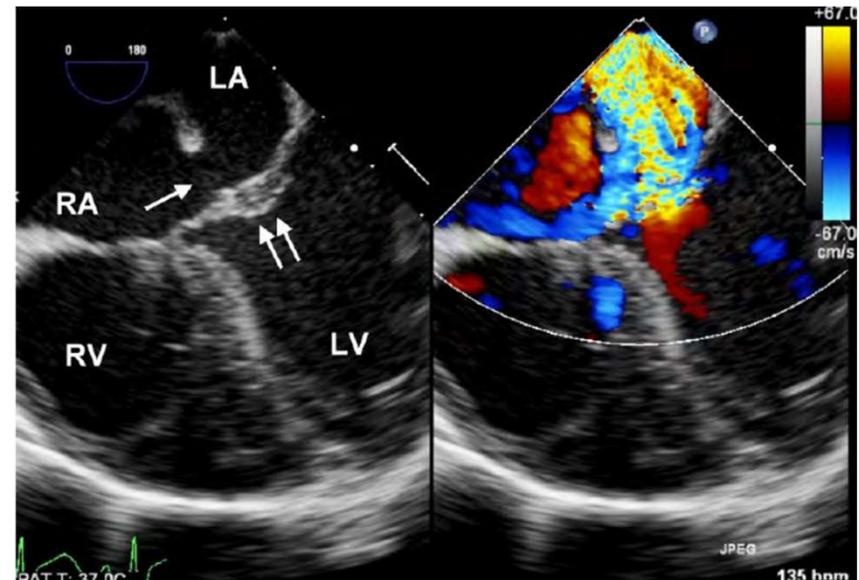


Partial AV Septal Defect

A partial AVSD usually has:

- a primum ASD and two separate AVVs
- cleft in the anterior leaflet of the left-AVV.

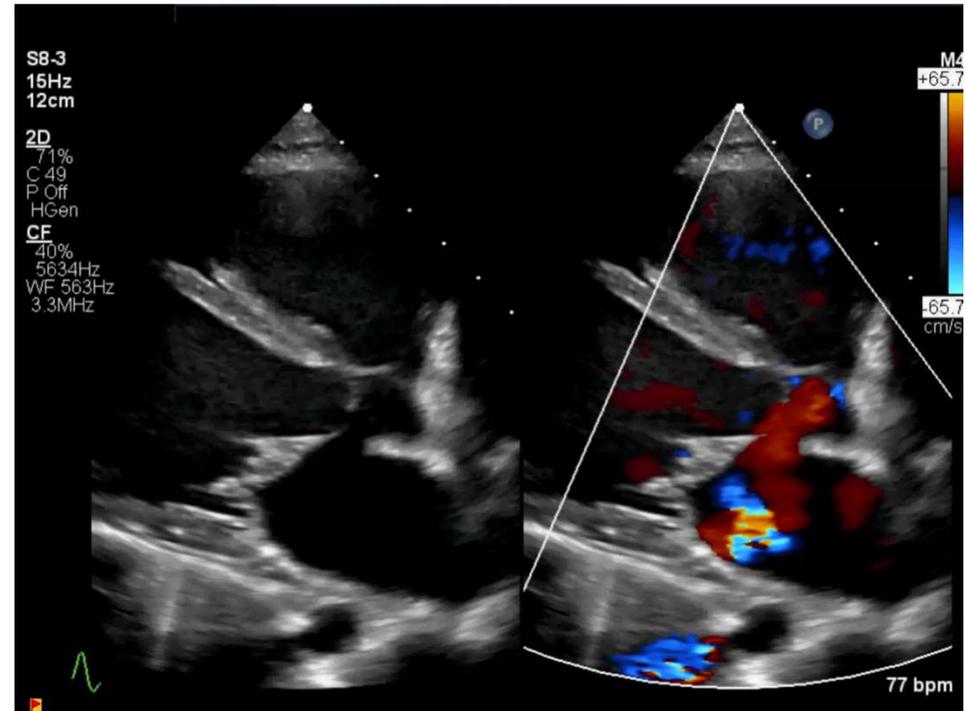
The cleft in the left-sided AVV usually results in some degree of regurgitation of the valve.



Partial atrioventricular septal defect.
TEE: The mid-esophageal four-chamber view demonstrates a primum atrial septal defect (atrial) and mitral cleft (double arrow) with severe mitral regurgitation.

Primum Atrial Septal Defect

- Located in the most anterior and inferior aspects of the atrial septum.



Presentation in Adults with repaired AVSD

- Left atrioventricular valve regurgitation and stenosis
- Left ventricular outflow tract (LVOT) obstruction attributable to the abnormal shape of the LVOT
- Arrhythmias – late onset complete heart block.

Left atrioventricular valve regurgitation is the most common reason for later surgical reintervention.

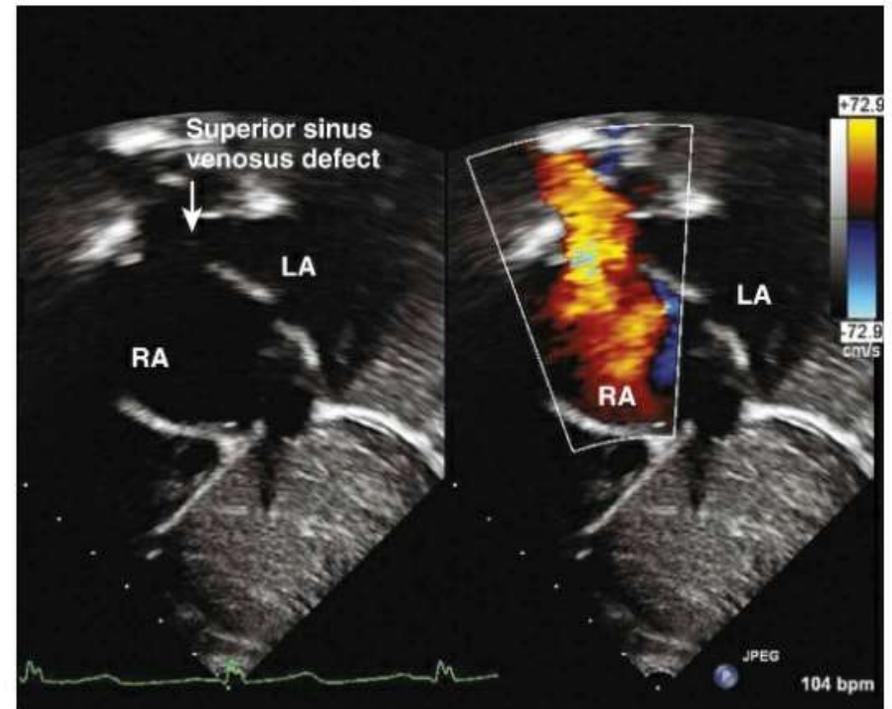
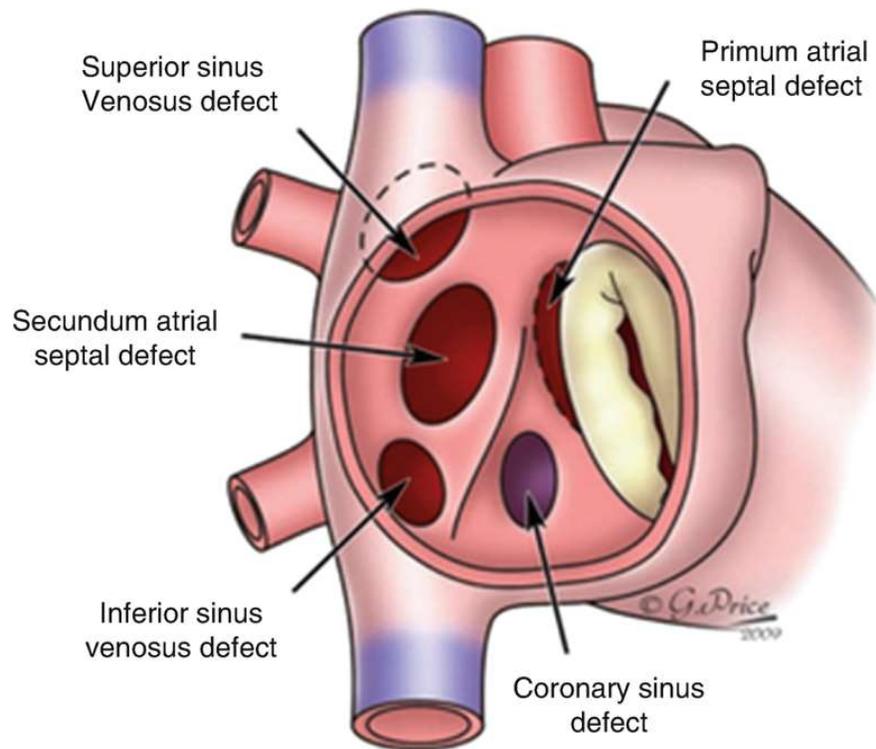
Recommendations for Atrioventricular Septal Defect

| COR | Recommendations |
|-----|--|
| I | Surgery for severe left atrioventricular valve regurgitation is recommended. |
| I | Surgery for primary repair of atrioventricular septal defect or closure of residual shunts in adults with repaired atrioventricular septal defect is recommended when there is a net left-to-right shunt (Qp:Qs \geq 1.5:1), PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic. |
| IIa | Operation for discrete LVOT obstruction in adults with atrioventricular septal defect is reasonable with a maximum gradient of 50 mm Hg or greater , a lesser gradient if HF symptoms are present, or if concomitant moderate-to-severe mitral or AR are present . |

COR - Class of Recommendation

Sinus venosus ASD

Sinus venosus ASD



Often associated with partial anomalous pulmonary venous drainage of the right pulmonary veins

Clinical Features

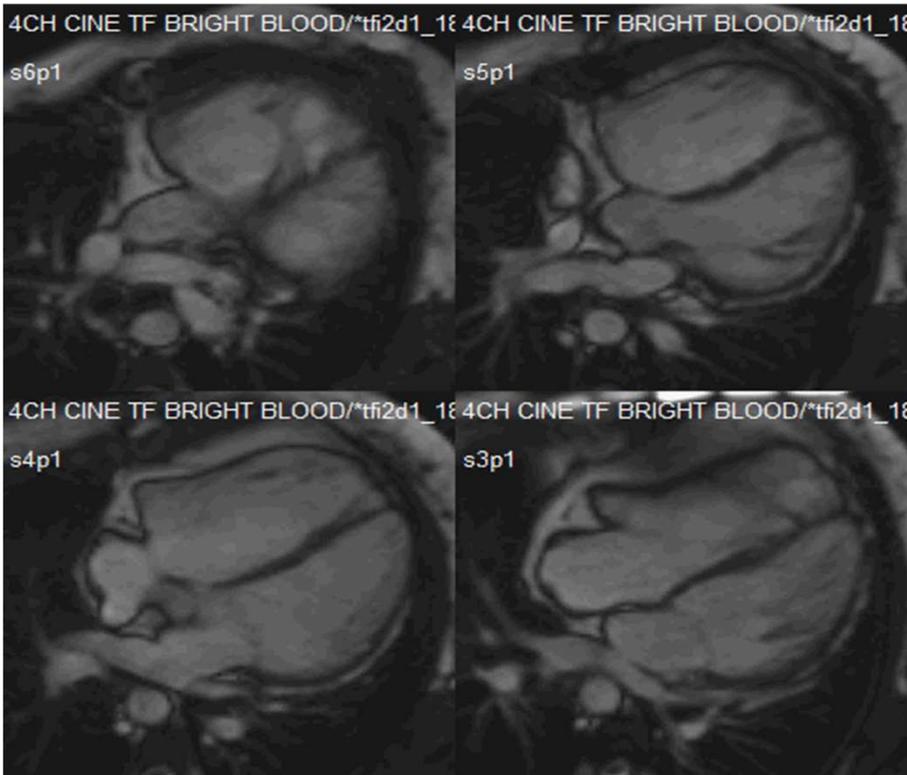
- The age at which symptoms appear is highly variable and is not exclusively related to the size of the shunt.
- Exercise intolerance is the most common initial presenting symptom.
- Atrial fibrillation or flutter is an age-related reflection of atrial dilation
- Less commonly, decompensated right heart failure in the older patient
- Tricuspid regurgitation (secondary to severe right heart and tricuspid annular dilation)
- Pulmonary arterial hypertension of variable severity
- Cyanosis being more common in inferior sinus venosus defects.

Diagnosis

CMR, CCT, Cardiac catheterization and TEE maybe superior to TTE in diagnosis of sinus venosus ASD.

Advanced cardiac imaging helps better define anomalous pulmonary veins

Cardiac MRI – in a 35 yo F with Inferior sinus venosus defect



LEFT VENTRICULAR VOLUME RESULTS

ED volume: 155.09 ml (96 - 174 ml) 1.05

ED volume index: 96.64 ml/m² (56 - 100 ml/m²)

Ejection fraction: 55.70 % (54 - 74 %)

LV mass ED index: 56.73 g/m² (37 - 67 g/m²)

RIGHT VENTRICULAR VOLUME RESULTS

ED volume: 245.44 ml (83 - 178 ml)

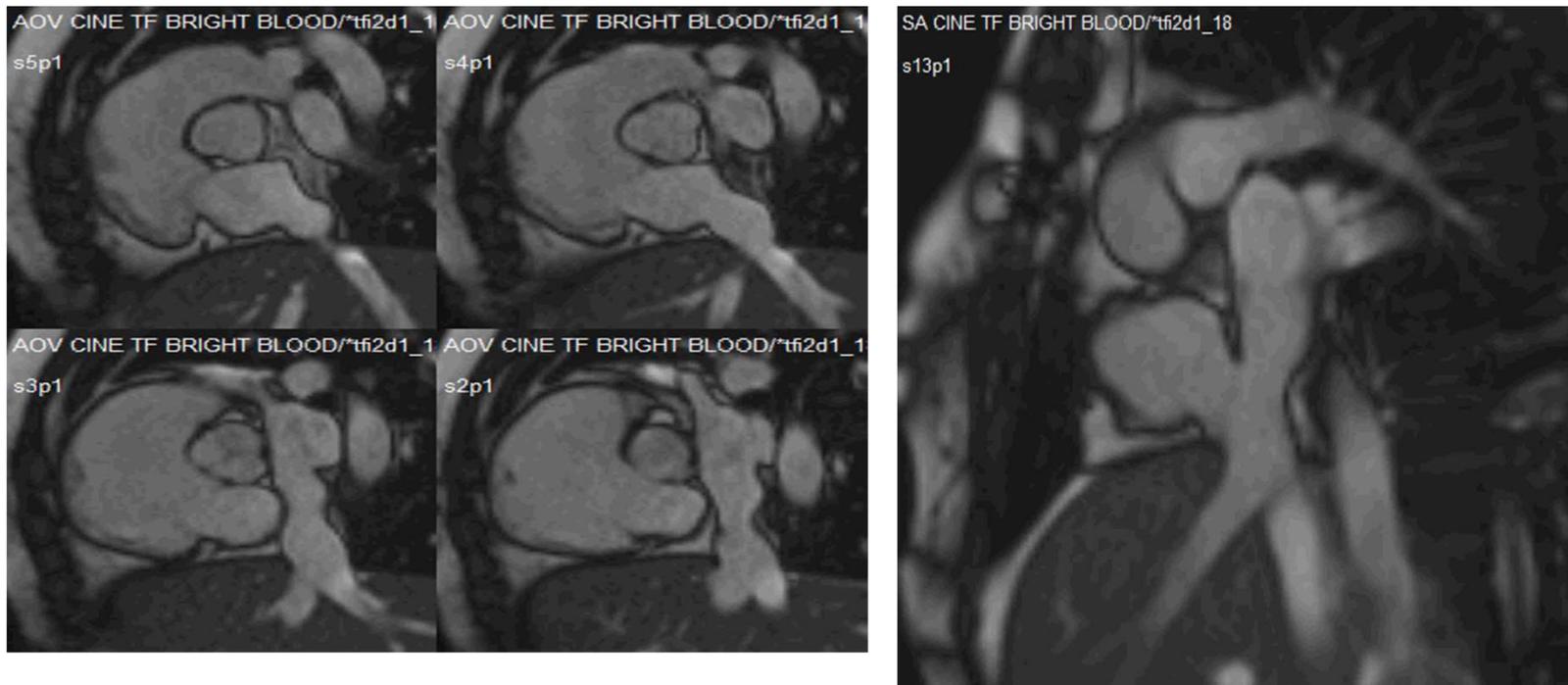
ED volume index: 152.94 ml/m² (47 - 103 ml/m²)

ES volume: 96.39 ml (32 - 73 ml)

Ejection fraction: 60.73 % (49 - 70 %)

Qp:Qs = 1.7:1

Over-riding IVC to inter-atrial septum



Recommendations for Primum and Sinus Venosus Atrial Septal Defects

| COR | Recommendations |
|-----|--|
| I | Adults with primum ASD, sinus venosus defect or coronary sinus defect causing impaired functional capacity, right atrial and/ or RV enlargement and net left-to-right shunt sufficiently large to cause physiological sequelae (eg, Qp:Qs \geq 1.5:1) without cyanosis at rest or during exercise, should be surgically repaired unless precluded by comorbidities , provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance. |

COR - Class of Recommendation

Ventricular Septal Defects

Ventricular Septal Defects

Most common isolated congenital cardiac defect.

VSDs are frequently associated with more complex cardiac malformations such as:

- Conotruncal malformations including

 - Double-outlet RV.

 - Truncus arteriosus.

 - Tetralogy of Fallot (TOF).

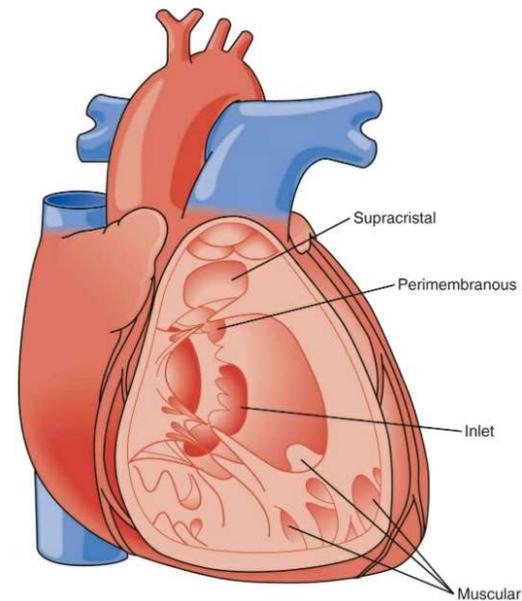
 - Interrupted aortic arch.

 - Complete AVC defects.

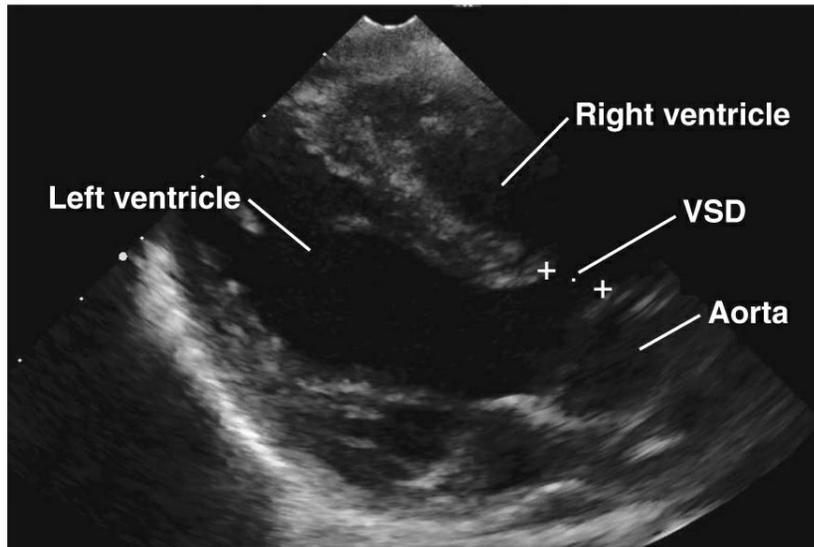
 - Coarctation of the aorta (Ao).

Types of Ventricular Septal Defects

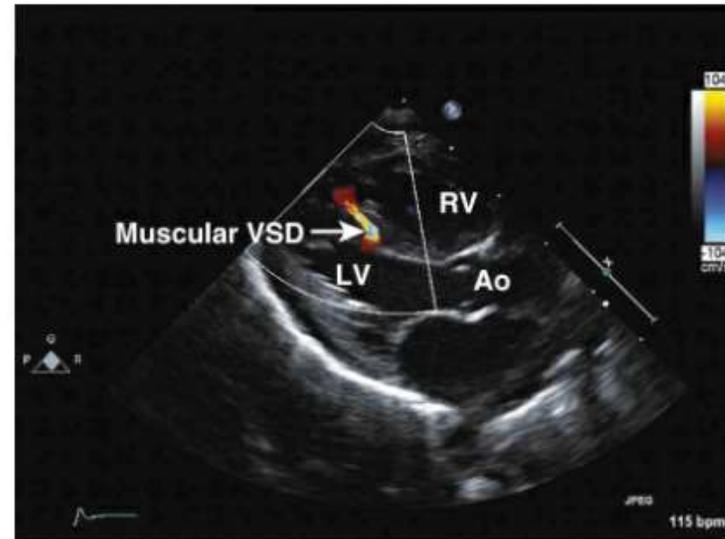
- VSDs may overlap two or more classifications depending on the size and shape of the defect.
 - **Perimembranous – 70%**
 - **Muscular – 20%**
 - Inlet – AV Septal Defects 5%
 - Supracristal (subpulmonary) 5%



Common Types of VSD

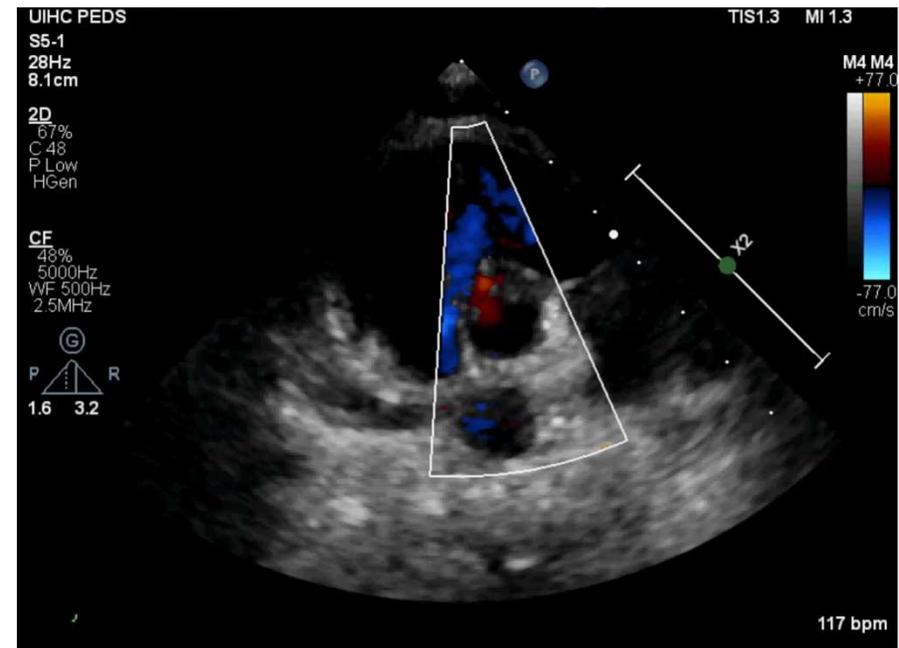
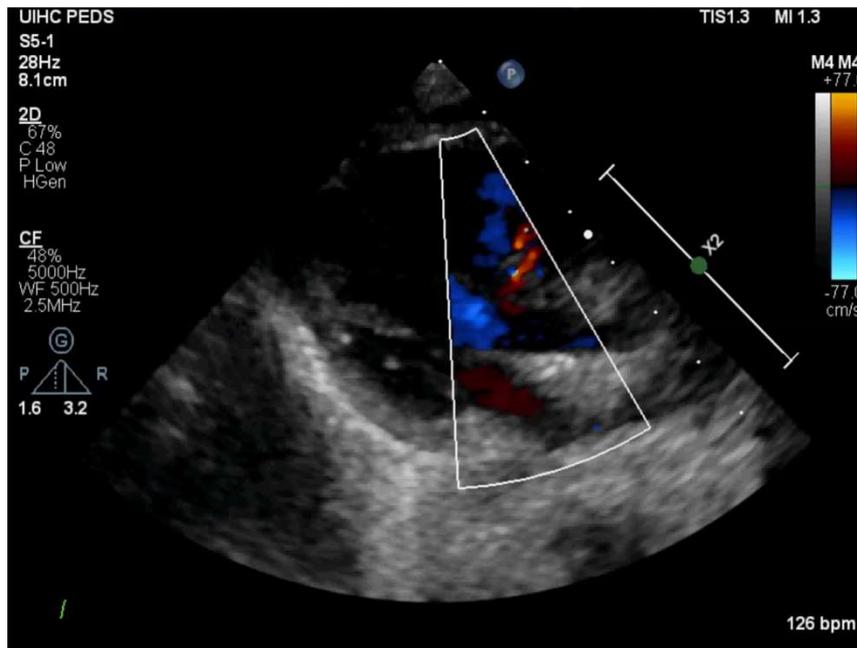


Perimembranous

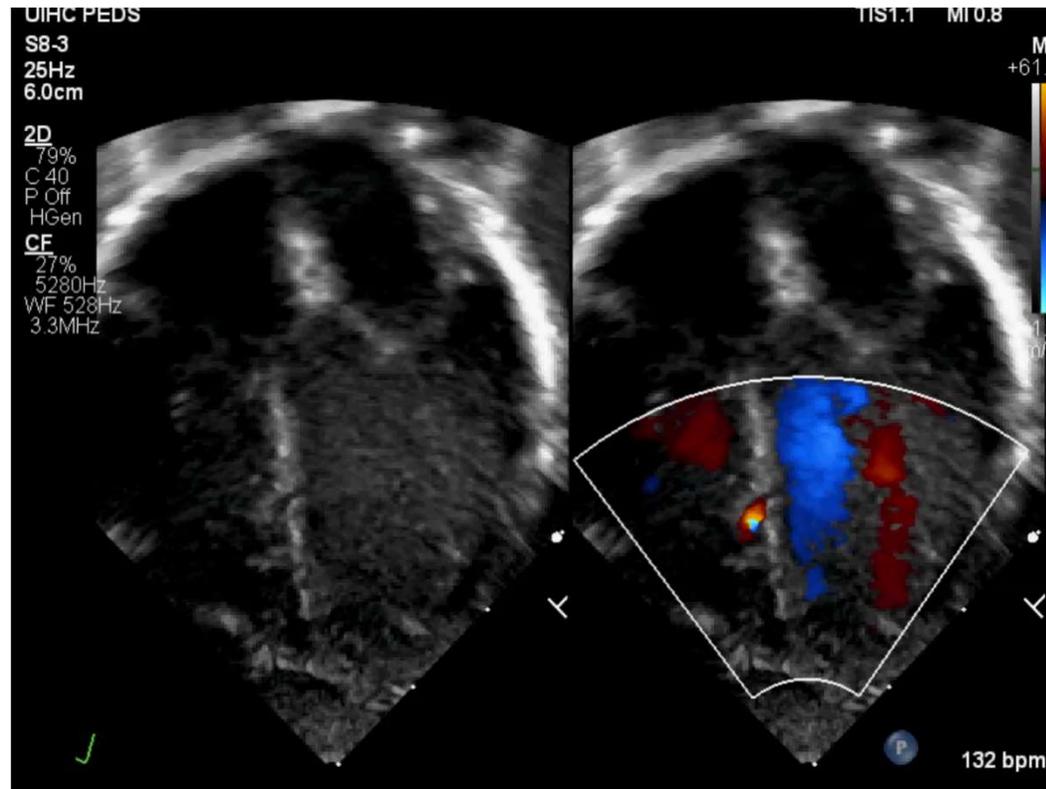


Muscular

Perimembranous VSD



Muscular VSD



Clinical Features in Adults

- Patients repaired earlier in life are typically asymptomatic sometimes with small residual VSDs are usually small.
- Exercise intolerance as a result of LV dilation due to a moderate VSD.
- With age the LV becomes less compliant, LA pressures rise, and symptoms develop.
- Young adults with large defects that were missed can present with severe pulmonary hypertension and systemic cyanosis consistent with Eisenmenger syndrome

Clinical Features

- VSD– Restrictive defects will produce a loud, holosystolic, high-pitched murmur.
- Small muscular septal defects may only cause early systolic murmur.
- Large defects may produce only a soft or absent murmur due to laminar, nonturbulent flow
- Aortic cusp prolapse in patients with supracristal defect can cause diastolic murmur of aortic insufficiency.
- In patients with unrepaired VSD, there is an increased risk of IE, typically involving the tricuspid and pulmonic valves

Diagnosis

ECG: Left-sided pressure/volume overload – LVH; LA enlargement.

CXR: Normal or LA and LV enlargement.

In pulmonary hypertension – Prominent pulmonary artery

Echo: size and location

LA, LV size

Right sided pressures from TR

Aortic valve prolapse

RV outflow tract obstruction—double-chamber RV

Cardiac Cath and Advanced Imaging in VSD

Cardiac cath:

Qp:Qs and response to inhaled iNO in pulmonary hypertension.

Cardiac MRI:

Can better define the size and location.

Estimate ventricular size

Calculate Qp:Qs

Associated valvular lesions and regurgitant fraction.

Recommendations for Ventricular Septal Defect

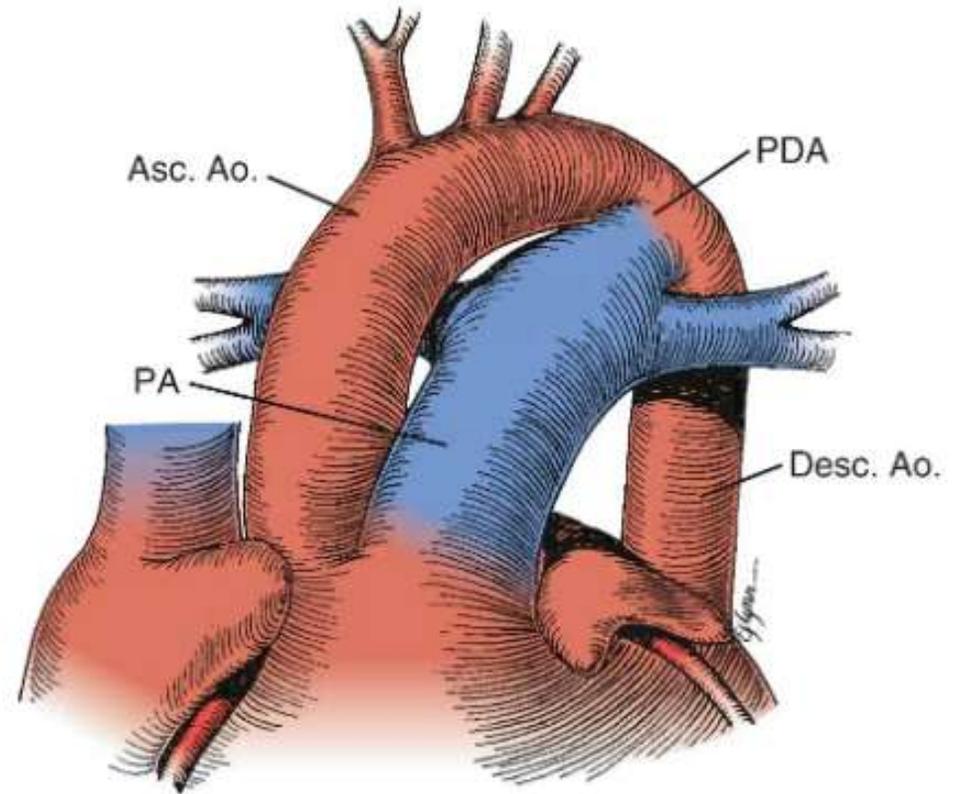
| COR | Recommendations |
|----------|--|
| I | Adults with a VSD and evidence of left ventricular volume overload and hemodynamically significant shunts ($Q_p:Q_s \geq 1.5:1$) should undergo VSD closure , if PA systolic pressure is less than 50% systemic and pulmonary vascular resistance is less than one third systemic. |
| III Harm | VSD closure should not be performed in adults with severe PAH with PA systolic pressure greater than two thirds systemic, pulmonary vascular resistance greater than two thirds systemic and/or a net right-to-left shunt |

COR - Class of Recommendation

Patent Ductus Arteriosus

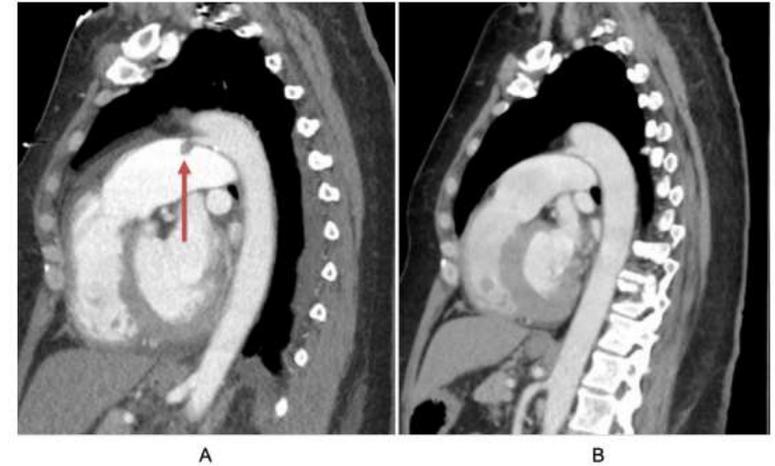
Patent Ductus Arteriosus

- A patent ductus arteriosus is a persistent patency of a fetal connection (6th left branchial arch) between the descending aorta (6th left branchial arch) just distal to the origin of the left subclavian artery and PA.
- PDA occurs in 5- 10% congenital heart defects.



Clinical Features

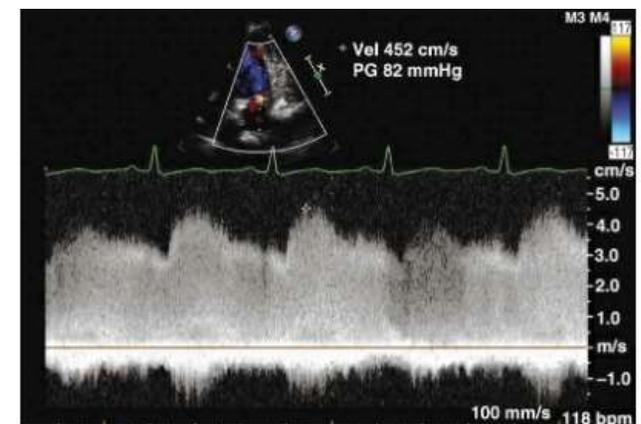
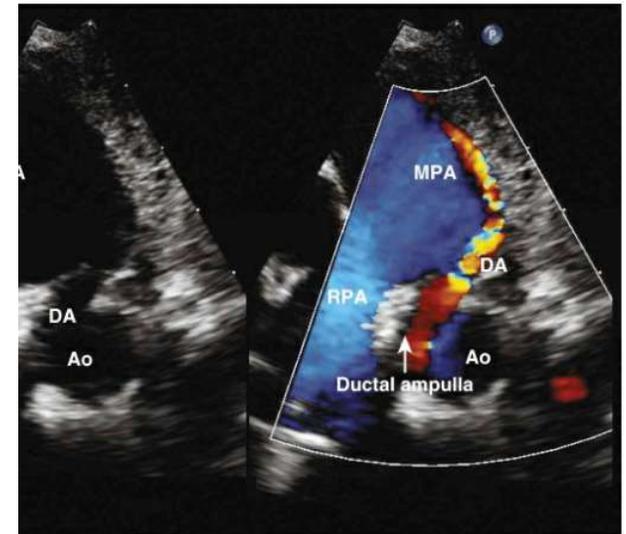
- Small PDA $Q_p:Q_s < 1.5$. usually asymptomatic; rarely infective endarteritis.
- Moderate PDA - $Q_p:Q_s < 1.5-2.2$; left-sided volume overload and congestive heart failure, often unmasked by the onset of atrial fibrillation.
- Large PDA – $Q_p:Q_s > 2.2$. who present late may have already progressed to severe pulmonary hypertension.



Contrast enhanced CT showing filling defect representing PDA Endarteritis (A) and follow up CT after completion of IV antibiotics (B).

Diagnosis:

- CXR:
 - LA/LV enlargement
 - Pulmonary HTN: Prominent pulmonary artery with increased pulmonary vascular markings. – Distal pulmonary vascular pruning in severe PHTN.
 - PDA calcification may be seen
- Echo:
 - Shunt direction.
 - PDA velocity - aortopulmonary pressure gradient.
 - Aortic arch and isthmus anatomy
- Cardiac MR/CCT:
 - can be useful to fully define the anatomy of a patent ductus arteriosus in preparation for surgical or percutaneous closure



Recommendations for Patent Ductus Arteriosus

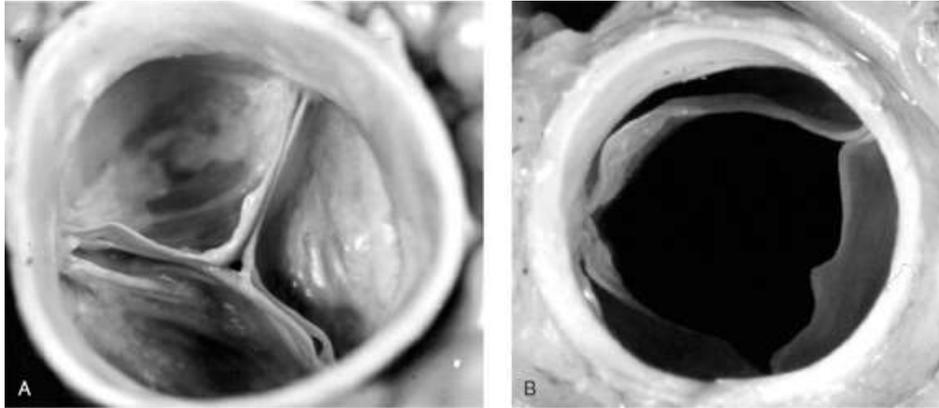
| COR | Recommendations |
|-----|--|
| I | PDA closure in adults is recommended if left atrial or LV enlargement is present and attributable to PDA with net left-to-right shunt, PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic. |

3D Aortic Arch with PDA Device – 2 yo

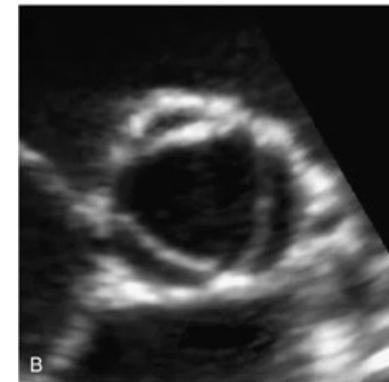
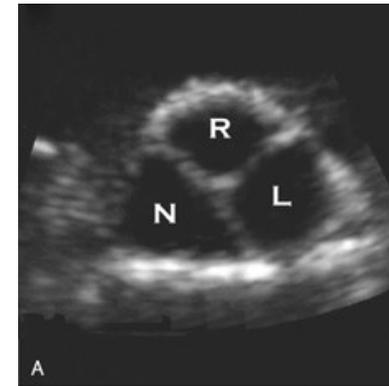


Aortic Valve Anomalies

Normal Aortic valve



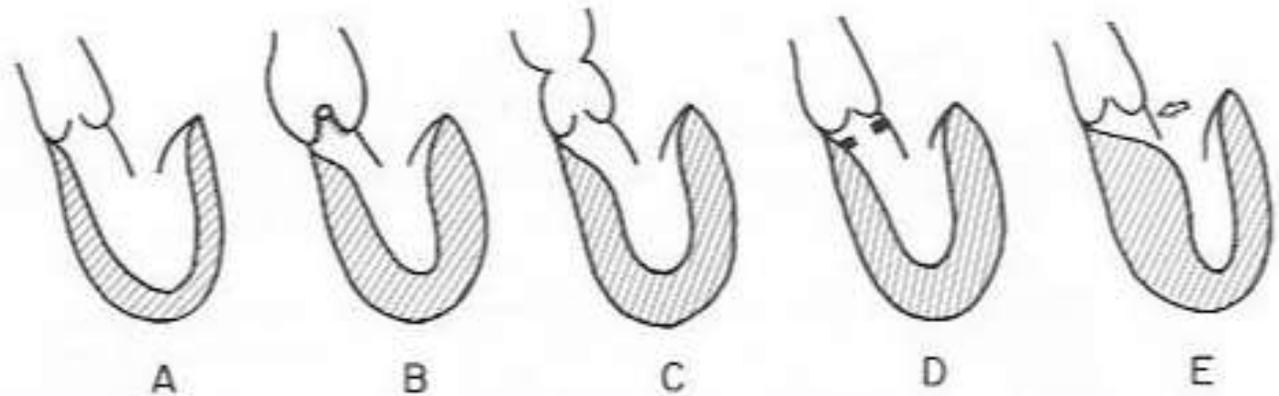
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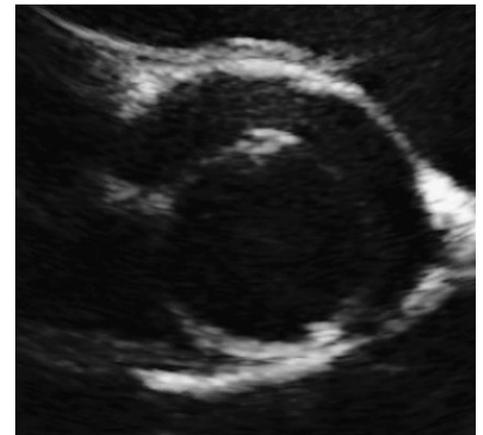
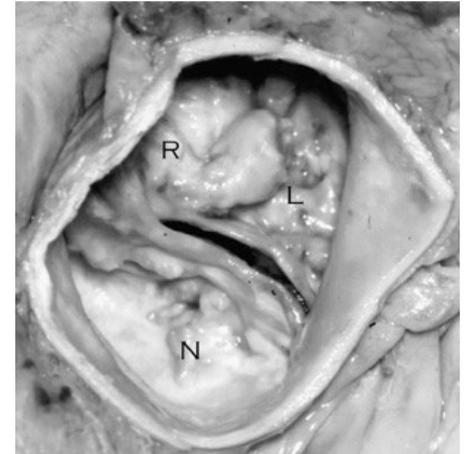
Aortic Stenosis

- Aortic Stenosis Types:
 - Valvular
 - Subvalvular
 - Supravalvular



Valvular Aortic Stenosis- Pathology

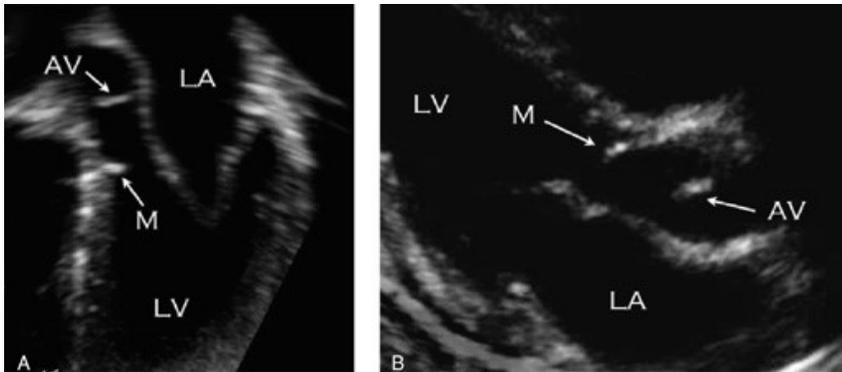
- Thickening, increased rigidity, diminished commissural separation
- Bicuspid/Unicuspid/Quadricuspid
- Under development of aortic valve ring
- Thickening and calcification in later life



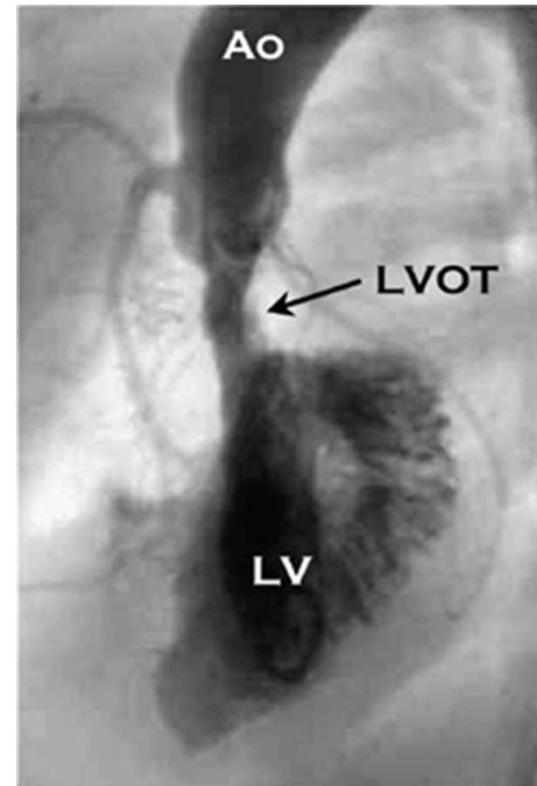
Subaortic Stenosis

- The prevalence of subaortic stenosis is 6.5% among patients with adult congenital heart disease.
- Subaortic stenosis is often caused by obstruction in the left ventricular outflow tract (LVOT) due to:
 - a discrete membrane (type I) or
 - less commonly by diffuse and tunnel-like obstruction (type II).
- Discrete subaortic membranes are occasionally associated with:
 - Primum atrial septal defects
 - membranous ventricular septal defects
 - coarctation of the aorta,
 - double outlet right ventricle
 - tetralogy of Fallot

Subvalvular Aortic Stenosis



M- Sub-aortic Membrane 10% of AS

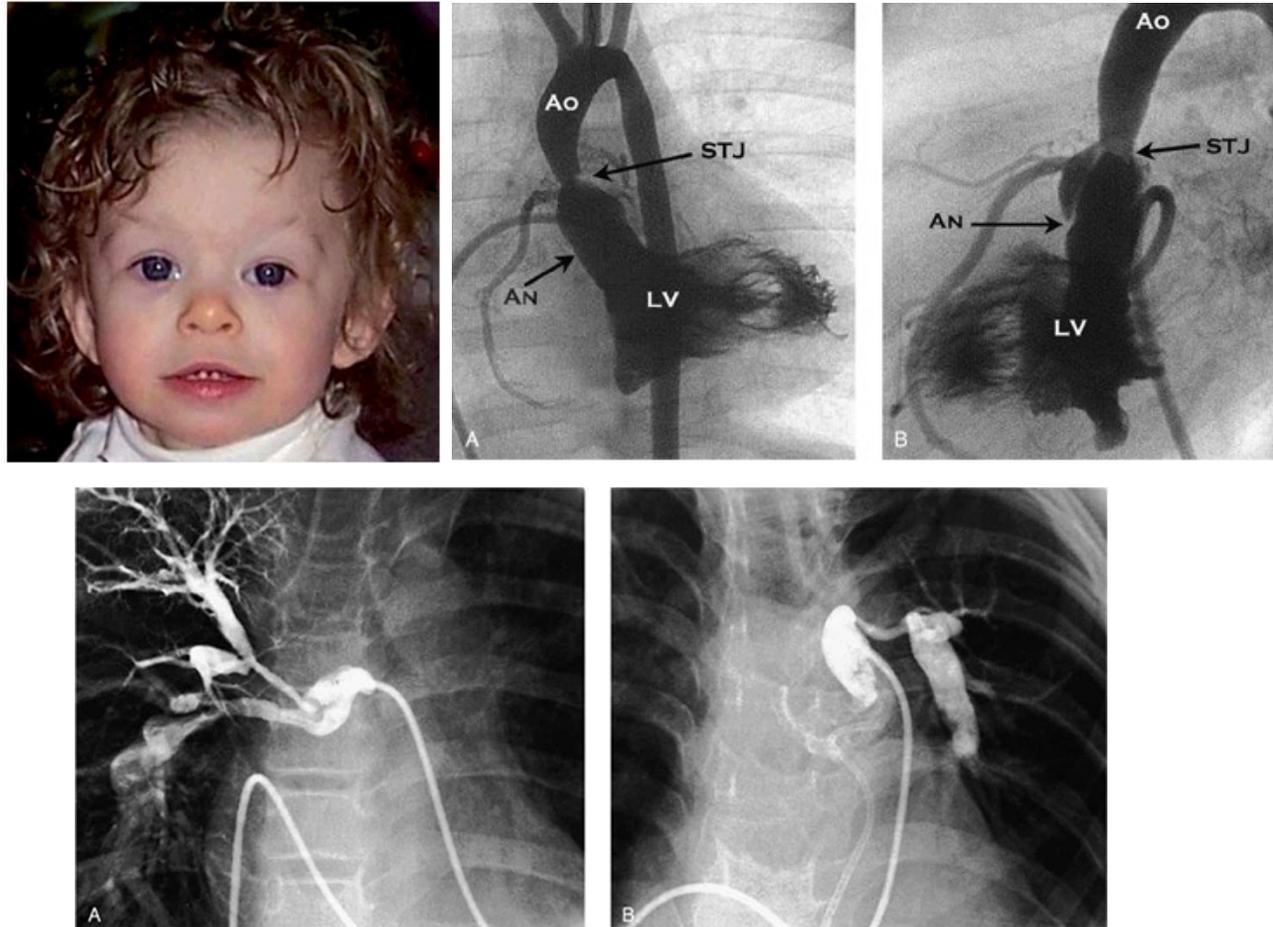


Fibromuscular narrowing of the LVOT

Associated Defects

- BAV is associated with aortopathies - may lead to ascending aortic dilation and an increased risk of aortic dissection over time.
- BAV is also associated with atrial septal defects, ventricular septal defects, mitral valve prolapse, and aortic coarctation.
- Certain genetic syndromes and diseases have an association with BAV including:
 - Turner syndrome
 - LoeysDietz syndrome,
 - Shone's complex
 - aortic coarctation.

Supravalvular Aortic Stenosis - William Syndrome



Clinical Presentation

Adults with aortic stenosis may present with:

- dyspnea with exertion
- chest pain with exertion
- fatigue
- angina
- arrhythmia
- heart failure
- endocarditis

Diagnosis:

ECG: Increased QRS voltage may be seen on electrocardiogram in those patients who have developed left ventricular hypertrophy as a result of the aortic valve/LVOT obstruction.

Chest X-Ray: Usually Normal; Late stages of aortic stenosis may show cardiomegaly and pulmonary vascular congestion.

Echo:

- Define the anatomy of valve
- severity of aortic stenosis
- level and type of subaortic obstruction.
- Aortic insufficiency
- LV size and mass
- Aortic root dilatation

Cath and Advanced Imaging

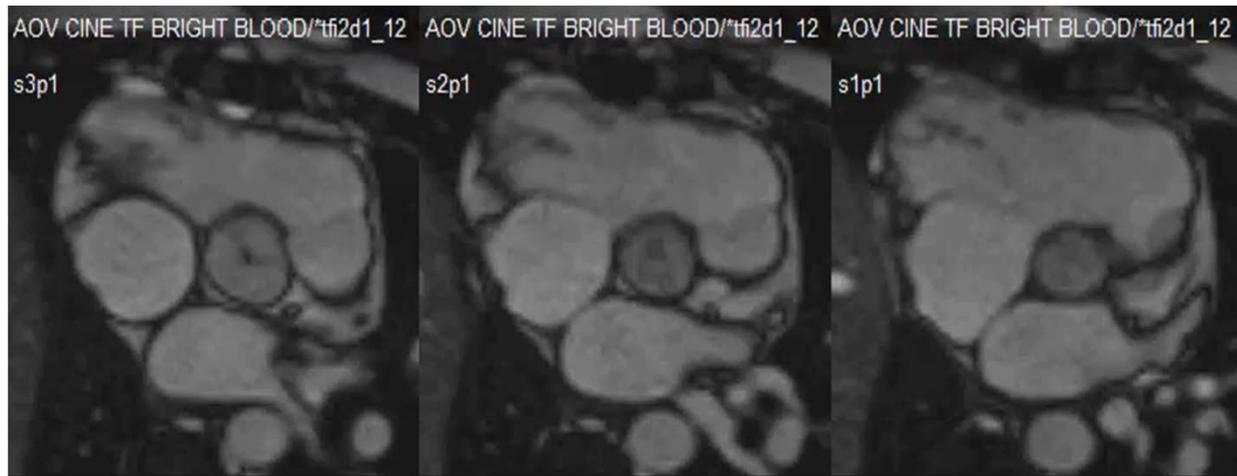
Cardiac catheterization may be indicated in those patients whose echocardiographic Doppler measurements do not correlate with the patient's symptoms.

Cardiac MRI:

- serial imaging of the aorta to follow aortic dimensions
- to calculate left ventricular ejection fraction
- to calculate aortic regurgitant volume.
- better define the anatomy of sub-aortic membrane.

Bicuspid Aortic Valve

Thickened and stenotic bicuspid aortic valve secondary to fusion of right and left coronary cusps.



Flow Measurements:

Aorta: Above Aortic Valve:

Peak flow velocity: 311.21 cm/s

Peak pressure gradient: 38.74 mmHg

Regurgitant fraction: 12.50 %

LVOT

MRI flow measurements suggest at least moderate aortic stenosis



Flow Measurements:

Aorta: Above Aortic Valve:

Peak flow velocity: 311.21 cm/s

Peak pressure gradient: 38.74 mmHg

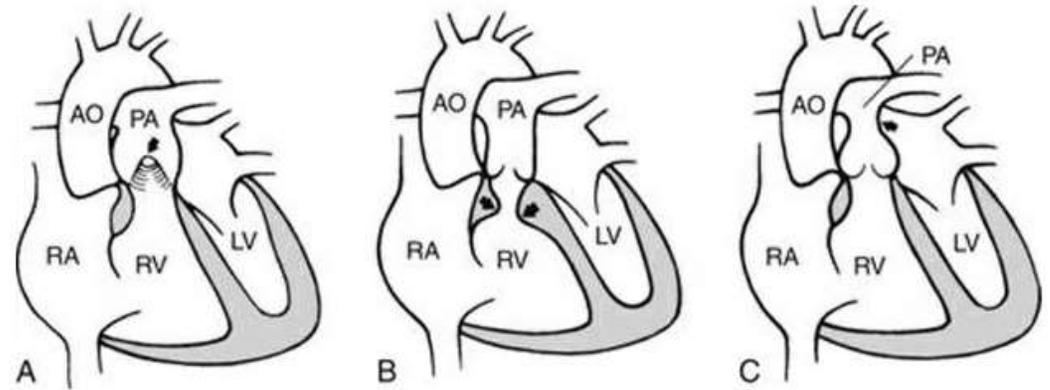
Regurgitant fraction: 12.50 %

Aortic Stenosis- Management

- **Balloon valvuloplasty** in a young patient with a stenotic BAV and no significant calcification or aortic regurgitation.
- **Surgical or transcatheter aortic valve replacement (TAVR)** is recommended for symptomatic severe aortic stenosis.
- **Subaortic stenosis:**
- **COR I Recommendation:**
 - Surgical intervention is recommended for adults with subAS, a maximum gradient >50 mm Hg
 - Surgical intervention is recommended for adults with subAS and less than 50 mm Hg maximum gradient and HF or ischemic symptoms, and/or LV systolic dysfunction attributable to subAS

Pulmonary Stenosis

PS



A. Valvular PS

- PV thickened, one-three leaflets, fused or absent commissures - Noonan's

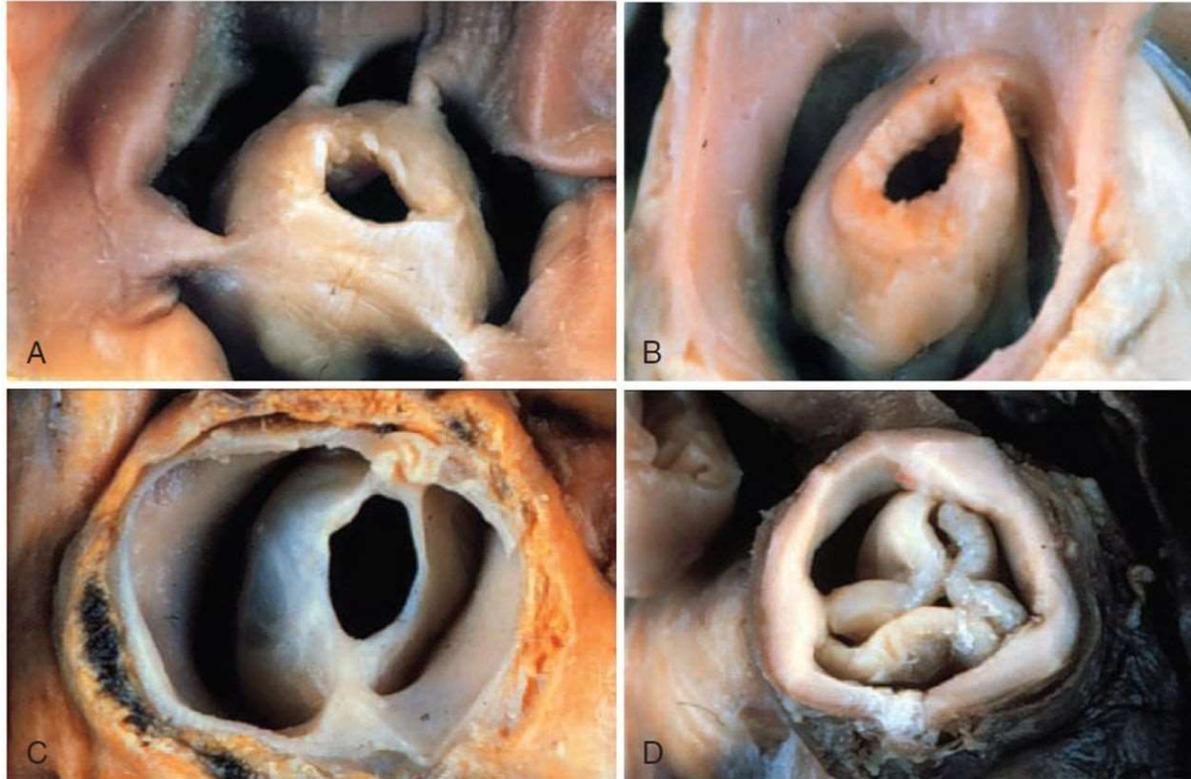
B. Subvalvular PS

- Infundibular - TOF

C. Supra-valvular PS

- Stenosis of MPA - William's Syndrome

Types of pulmonary valve stenosis.



Classification of pulmonary valve stenosis. The pulmonary valve can be (A) a commissural or (B) unicommissural with prominent systolic doming of the cusps and an eccentric orifice. (C) Bicuspid pulmonary valve is shown with fused commissures. (D) Dysplastic pulmonary valve is severely thickened with deformed valve cusps.

Emily Ruckdeschel, and Yuli Y Kim *Heart* 2019;105:414-422

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Heart

PS- Pathology

- Hypertrophy of RV, infundibulum -> dynamic subvalvar obstruction
- Tricuspid Regurgitation - RA dilation
- Post stenotic dilation of PA-LPA in classic form
- CHF in severe stenosis.

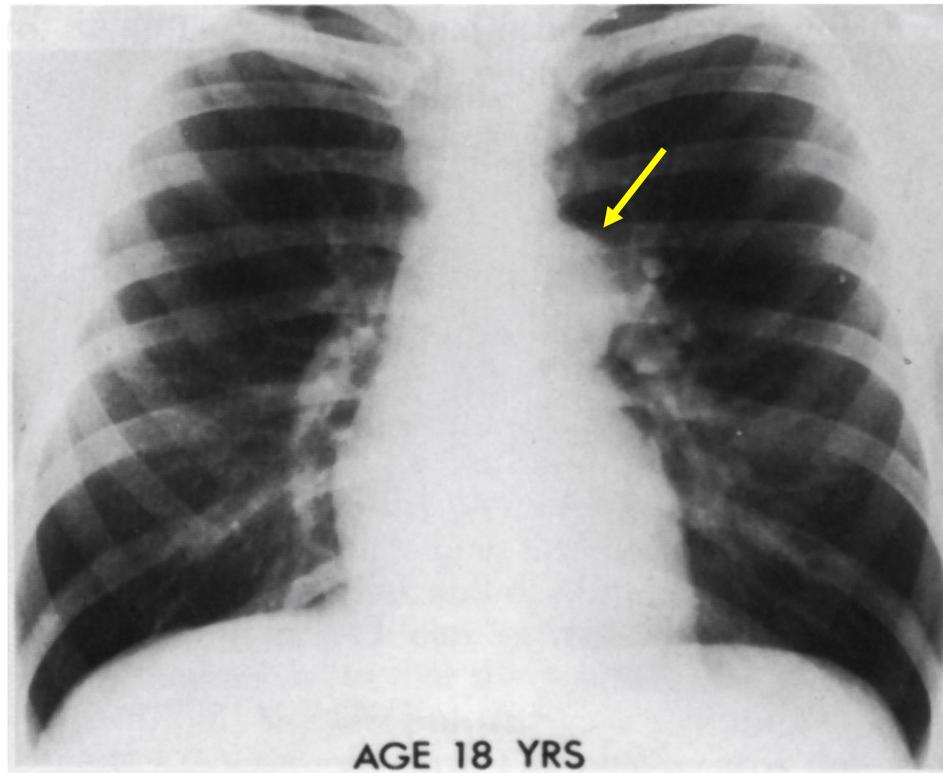
- Association: Noonan Syndrome :
 - 60% of patients with Noonan Syndrome have some level of PS

PS- Clinical Presentation

- Most asymptomatic
- Exertional dyspnea, fatigue; chest pain syncope, sudden death
- Occasionally - infective endocarditis can develop.

- Physical Exam findings:
 - “a” wave on JVP
 - RV heave
 - Ejection click with a systolic ejection murmur (maybe absent in Noonan).
 - Soft and late P2

PS- CXR



Post-stenotic Dilatation with normal vascularity

Diagnosis:

Echo:

- Pulmonary valve leaflet morphology and thickness
- Annular, main pulmonary artery, and branch dimensions
- Continuous wave Doppler peak and mean gradient
- Degree of regurgitation
- Subvalvular hypertrophy or obstruction
- Right ventricular size, wall thickness, and function
- Doppler tricuspid regurgitant peak velocity

Cath: RV pressures and transvalvular gradients

Cardiac MRI:

- precise quantification of pulmonic regurgitation
- serial assessment of RV size and function

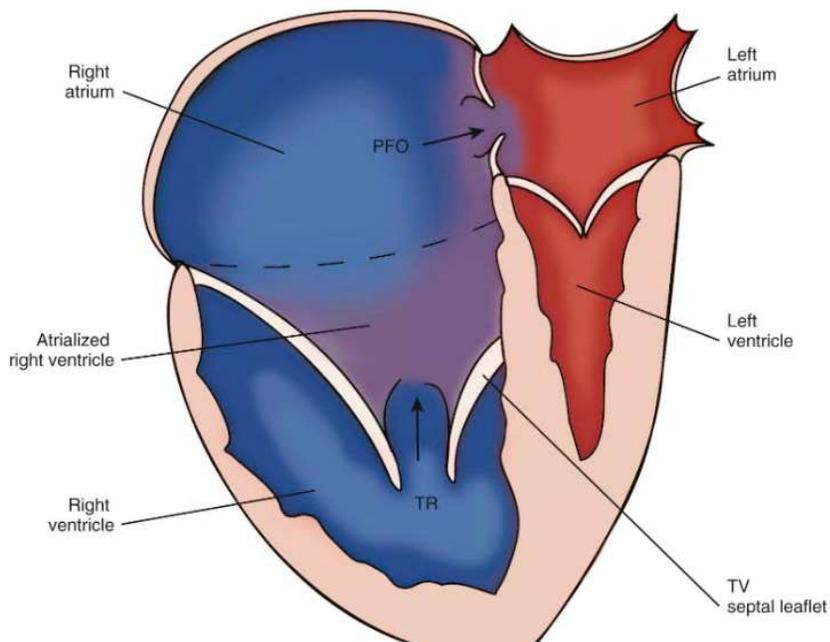
Recommendations for Valvular Pulmonary Stenosis

| COR | Recommendations |
|-----|---|
| I | In adults with moderate or severe valvular pulmonary stenosis and otherwise unexplained symptoms of HF, cyanosis from interatrial right-to-left communication, and/ or exercise intolerance, balloon valvuloplasty is recommended. |
| I | In adults with moderate or severe valvular pulmonary stenosis and otherwise unexplained symptoms of HF, cyanosis, and/ or exercise intolerance who are ineligible for or who failed balloon valvuloplasty, surgical repair is recommended. |
| IIa | In asymptomatic adults with severe valvular pulmonary stenosis, intervention is reasonable. |

COR - Class of Recommendation

Ebstein's Anomaly

Ebstein's Anomaly



Ebstein's anomaly of the tricuspid valve (TV) accounts for <1% of all cases of congenital heart disease.

The fundamental lesion in Ebstein's anomaly is failure of delamination of the tricuspid leaflets from the right ventricular (RV) myocardium.

tethered "sail-like" anterior TV leaflet and apically displaced septal and posterior leaflets.

Pathologic Specimen Cut in the 4-Chamber Plane From a Patient With Ebstein Anomaly



Carole A. Warnes JACC 2009;54:1903-1910

Physiology

- Progressive tricuspid regurgitation and RV failure.
-
- The functional RV can be quite small, myopathic, and RV contractility and compliance can be impaired, contributing to RV failure.
- Free tricuspid regurgitation can contribute to significant right-sided volume overload with both RA and RV dilatation.
- Associated defects:
 - ASD or PFO
 - Atrioventricular bypass tracts: WPW syndrome is present in ~25% of patients, and multiple bypass tracts are common
 - Pulmonic stenosis or pulmonary atresia

Clinical Features in Adults:

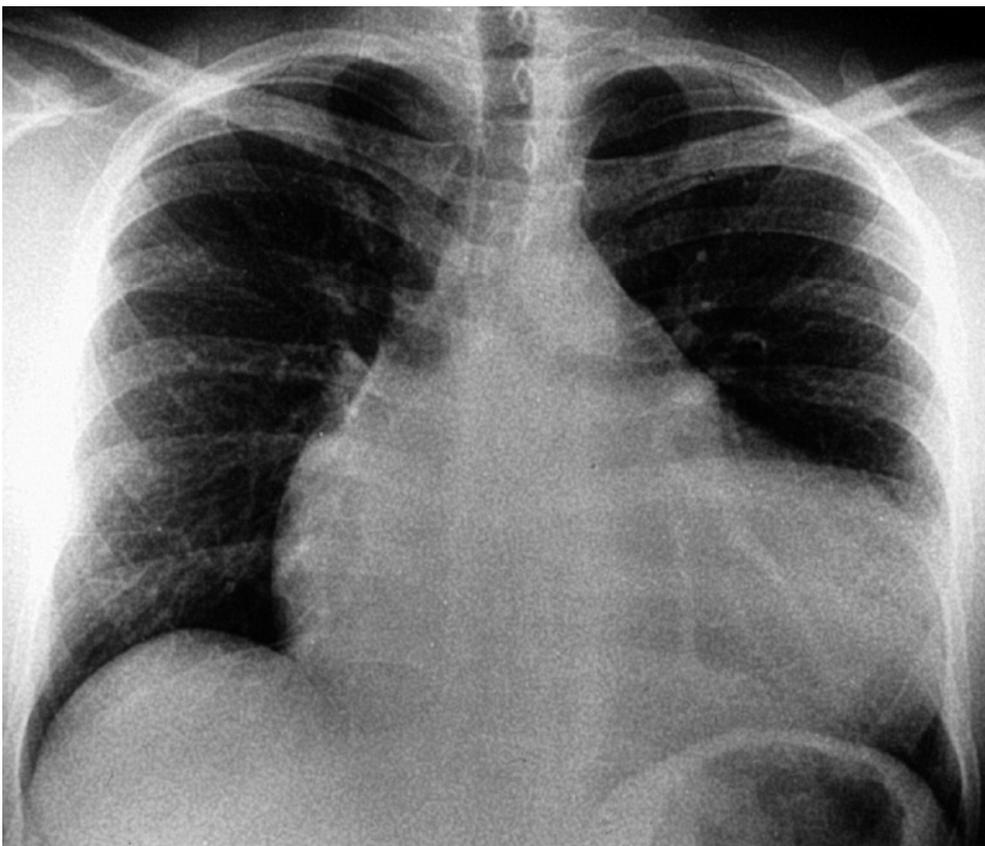
- Symptoms or deteriorating exercise capacity
- Cyanosis (oxygen saturation less than 90%)
- Paradoxical embolism
- Progressive cardiomegaly on chest radiography
- Progressive RV dilation or systolic dysfunction

ECG



ECG of an adult patient with Ebstein's anomaly. The electrocardiogram demonstrates right axis deviation, first-degree AV block, and complete right bundle branch block.

PR interval shortening can occur.



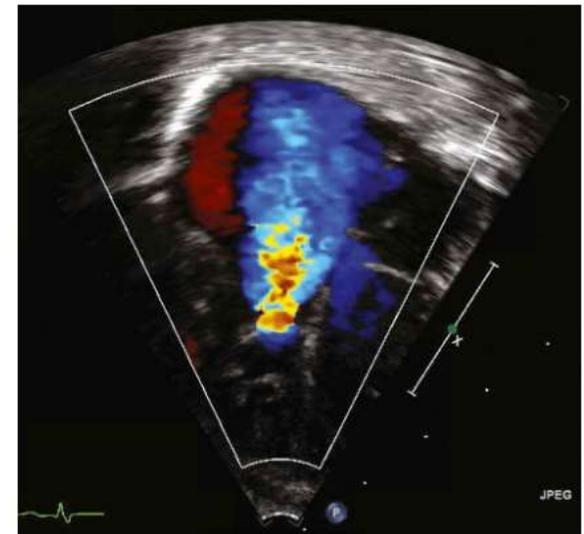
Chest Radiograph of a 32-Year-Old Man With Severe Ebstein Anomaly

The patient was referred late with severe right ventricular (RV) enlargement, severe RV dysfunction, and severe tricuspid regurgitation. He was cyanotic secondary to a small atrial septal defect and could walk only a few yards.

Carole A. Warnes JACC 2009;54:1903-1910

Echo

- TV anatomy, severity of tricuspid regurgitation, RV size, and associated abnormalities:
 - apical displacement of the septal tricuspid leaflet of more than 8 mm in the apical four chamber view.
 - Identifies the redundant, elongated anterior tricuspid leaflet and evaluates the functional RV's size and function



Advanced Imaging

- In adults with Ebstein's anomaly, CMR can be useful to determine anatomy, RV dimensions, and systolic function.
- EP study - can be useful in the diagnostic evaluation of adults with Ebstein's anomaly and ventricular preexcitation but without supraventricular tachycardia.

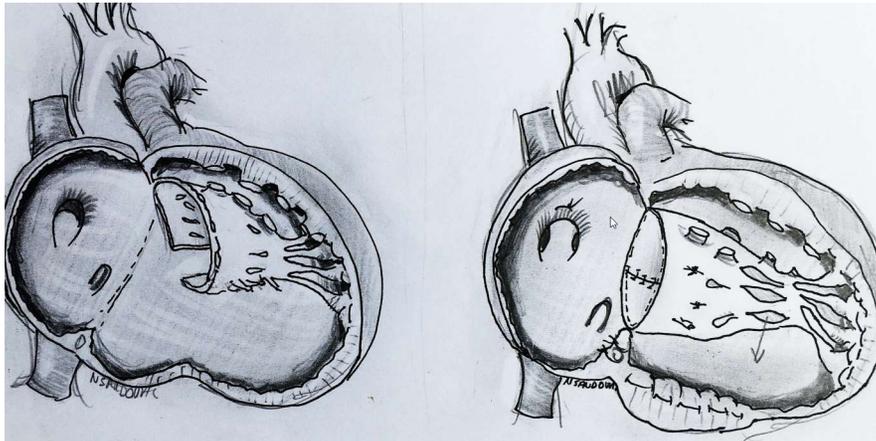
Recommendations for Ebstein's Anomaly

| COR | Recommendations |
|-----|--|
| I | Surgical repair or reoperation for adults with Ebstein anomaly and significant TR is recommended when one or more of the following are present: HF symptoms, objective evidence of worsening exercise capacity, progressive RV systolic dysfunction by echocardiography or CMR. |
| I | Catheter ablation is recommended for adults with Ebstein anomaly and high-risk pathway conduction or multiple accessory pathways |
| IIa | Surgical repair or reoperation for adults with Ebstein anomaly and significant TR can be beneficial in the presence of progressive RV enlargement, systemic desaturation from right-to-left atrial shunt, paradoxical embolism, and/or atrial tachyarrhythmias |

COR - Class of Recommendation

Management - Options for tricuspid valve intervention

1. “Cone” procedure
2. Tricuspid valve replacement
3. Bidirectional Glenn procedure (or bidirectional cavopulmonary connection) - This procedure is often referred to as the “1.5 ventricle repair,”



Cone Procedure:

- anterior and posterior tricuspid valve leaflets are mobilized in their entirety from their anomalous attachments in the RV.
- The free edge of this complex is rotated clockwise to be sutured to the septal border of the anterior leaflet, creating a “cone,” the vertex of which remains fixed at the right ventricular apex

Anomalous Pulmonary Veins

Anomalous Pulmonary Veins - Types

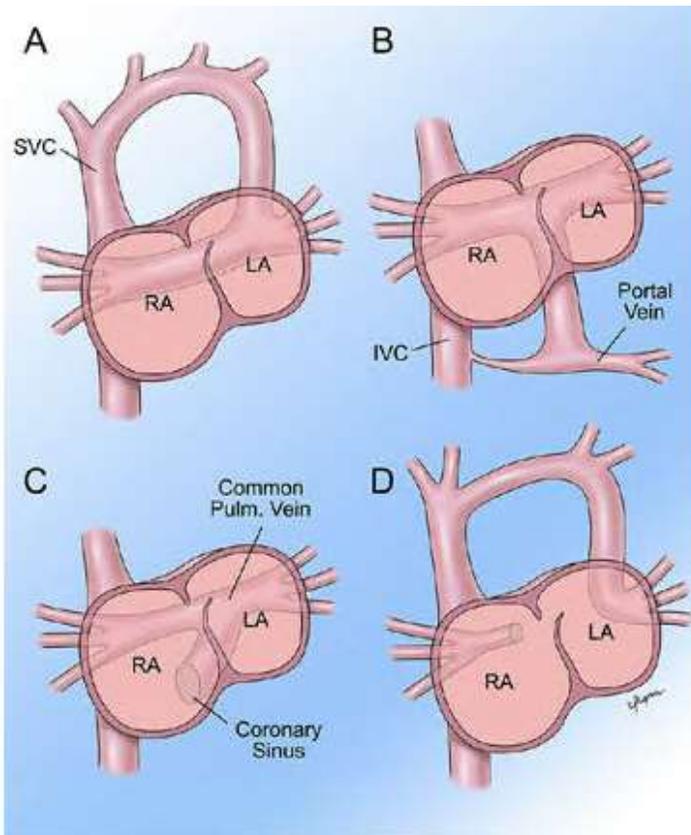


Diagram depicting the various types of total anomalous pulmonary venous return. For each type, there is an atrial communication enabling blood to reach the left side of the heart.

A: Supracardiac- the right and left pulmonary veins join in a confluence posterior to the left atrium and are connected via an ascending vertical vein to the innominate vein, which drains to the superior vena cava.

B: Infracardiac (Infradiaphragmatic): the confluence of pulmonary veins drains via a descending vertical vein to the portal venous system.

C: Cardiac (Coronary sinus): the right and left pulmonary veins drain into the coronary sinus.

D: Mixed pattern: the diagram depicts the left pulmonary veins draining via a vertical vein to the innominate vein and the right pulmonary veins connecting directly to the right atrium.

Partial anomalous venous return (PAPVR)

- **Right lung drainage to:**

- SVC
- RA
- Coronary sinus
- Azygos vein
- Inferior vena cava (IVC)—This lesion is known as the “Scimitar syndrome” due to the radiographic appearance, which resembles a Turkish sword or “scimitar.” Usually drains the entire right lung .

- **Left lung drainage to:**

- Brachiocephalic vein
- Coronary sinus
- Hemiazygos vein

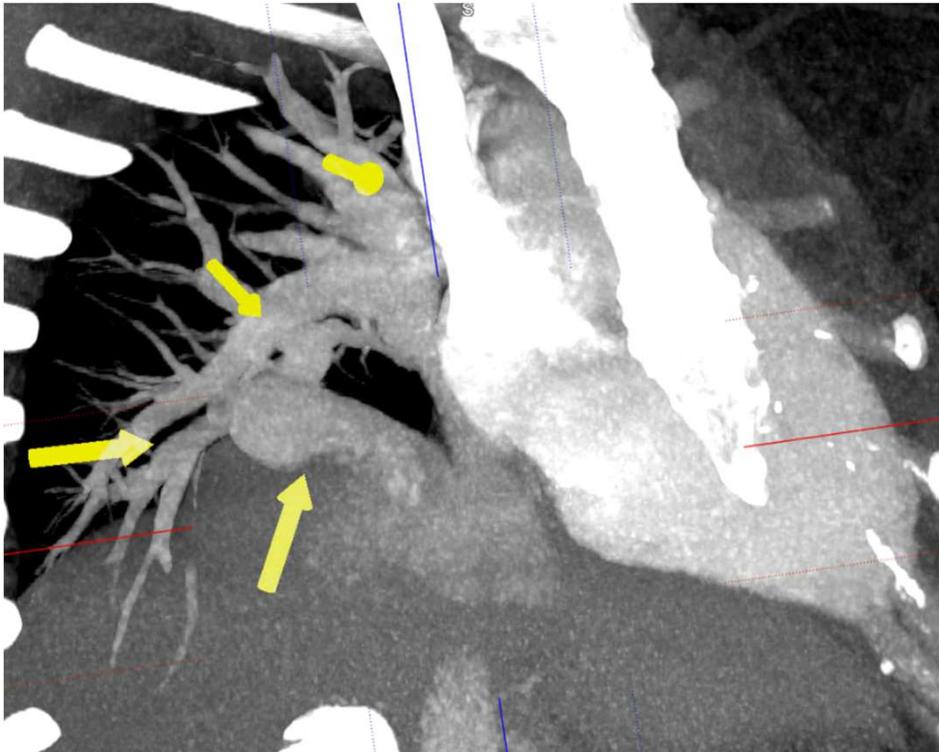
Clinical Presentation in Adults

- Presentation is dependent on the volume of the shunt and the presence and significance of associated cardiac defects.
 - With large-volume shunt lesions, patients may present early in childhood.
 - With moderate-volume lesions, patients may present in adulthood with dyspnea on exertion, palpitations, and symptoms of pulmonary hypertension and right heart failure.
 - With low-volume shunt lesions, patients may never develop symptoms, and PAPVR may be discovered as an incidental finding on chest or cardiac imaging.
 - PAPVR should be considered in patients with unexplained RV enlargement and no intracardiac shunt identified by echocardiography.

Diagnosis:

- Physical Examination – findings similar to an ASD.
- ECG: RA enlargement, RV hypertrophy and Right axis deviation.
- CXR:
 - RA and right ventricular enlargement.
 - Prominent pulmonary artery with an increase in pulmonary vasculature.
- Echo:
 - RA and RV enlargement
 - Cannot adequately assess anomalous veins
- Cardiac MRT/CCT
 - modality of choice and is able to definitively define the anatomy of the anomalous venous return
 - Also helps calculate Qp:Qs

PAPVR – a recent case



35 yo F who used to be able to run half marathons who recently developed shortness of breath.

Echo: Borderline RV Dilatation
Chest CT: Anomalous drainage of RMPV and RLPV to IVC.

Cath: Qp:Qs of 1.9:1

Surgical Repair: Using autologous pericardial patch the anomalous pulmonary vein was baffled to LA.

Recommendations for Anomalous Pulmonary Venous Connections

| COR | Recommendations |
|-----|--|
| I | Surgical repair is recommended for patients with partial anomalous pulmonary venous connection when functional capacity is impaired and RV enlargement is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae (eg, Qp:Qs \geq1.5:1), PA systolic pressure is less than 50% systemic pressure, and pulmonary vascular resistance is less than one third of systemic resistance |
| I | Repair of partial anomalous pulmonary venous connection is recommended at the time of closure of a sinus venosus defect or ASD. |
| I | Repair of a scimitar vein is recommended in adults when functional capacity is impaired, evidence of RV volume overload is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae (eg, Qp:Qs \geq1.5:1), PA systolic pressure is less than 50% systemic pressure and pulmonary vascular resistance is less than one third systemic. |

COR - Class of Recommendation

References:

1. *Echocardiography in Congenital Heart Disease-* by Mark B. Lewin, Karen K Stout
2. *Adult Congenital Heart Disease in Clinical Practice* Editors: Yeh, Doreen DeFaria, Bhatt, Ami (Eds.)
3. *2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines*
4. *Pulmonary valve stenosis in the adult patient: pathophysiology, diagnosis and management* Emily Ruckdeschel, Yuli Y Kim.
5. *Adult Congenital Heart Disease; Importance of the Right Ventricle;* Carole A. Warnes.
6. *To close or not to close!! A case of pda endarteritis;* Srishti Nayak, Purvi Parwani and Ahmed Kheiwa

