ACYANOTIC CONGENITAL HEART DISEASE

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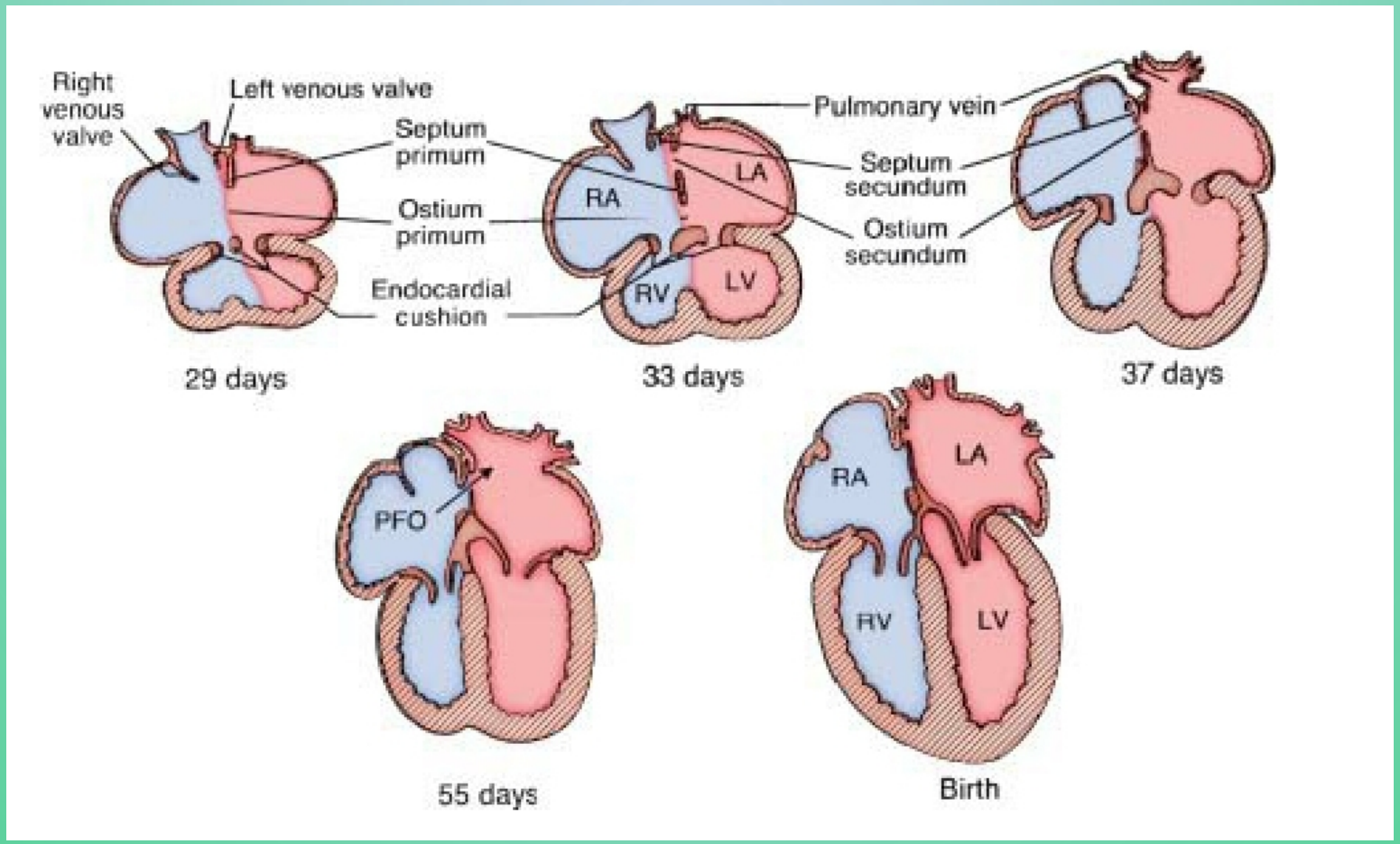
- **1- Left to Right shunts** => Atrial Septal Defect
- Ventricular Septal Defect
- Atrioventricular Septal Defect
- Patent Ductus Arteriosus

- 2- **Obstructive Lesions** => Pulmonary stenosis
- Aortic stenosis
- Coarctation of Arota

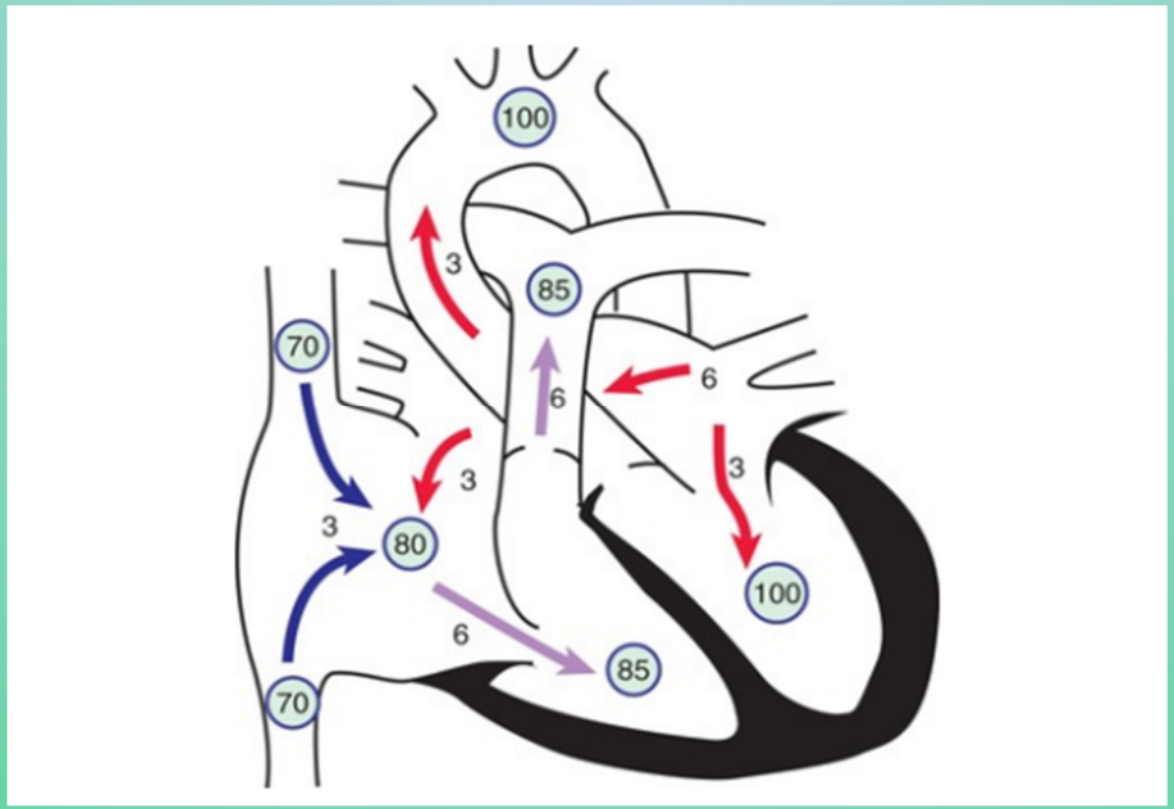
- **Left-to-Right Shunt Lesions :**
- ***Atrial Septal Defect :***
- ***TYPES:***
- Secundum , Primum , Sinus venosus , Coronary sinus
- Sporadic(mostly), hereditary (Holt-Oram syndrome-AD)
- Patent foramen ovale (PFO) => common in infancy, no hemodynamic significance


Right venous valve
Left venous valve
Septum primum
Ostium primum
Endocardial cushion
RA
LA
RV
LV
Pulmonary vein
Septum secundum
Ostium secundum
29 days
33 days
37 days
PFO
55 days
RA
LA
RV
LV
Birth

- A valve-competent but probe-patent PFO in 15–30% of adults.
- Isolated PFO => no surgical treatment, risk for paradoxical systemic embolization.
- Device closure in adults with a history of thromboembolic stroke

- ***Ostium Secundum Defect :***
- The most common
- ***Associated lesions:***
- Mitral valve prolapse ,Partial anomalous pulmonary venous return (PAPVR), usually of RUPVs
- Females/males =>3 : 1

- *The degree of shunt depends on :*
- 1-the size of the defect
- 2-the relative compliance of the right and left ventricles
- 3-the relative vascular resistance in the pulmonary and systemic circulations.
- Enlargement of the right atrium and ventricle and dilation of the pulmonary artery
- Normal PVR throughout childhood, may increase in adulthood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- ***Clinical Manifestations :***
- Asymptomatic
- Subtle failure to thrive
- Varying degrees of exercise intolerance
- ***The physical findings :***
- 1- mild left precordial bulge
- 2- RV systolic lift

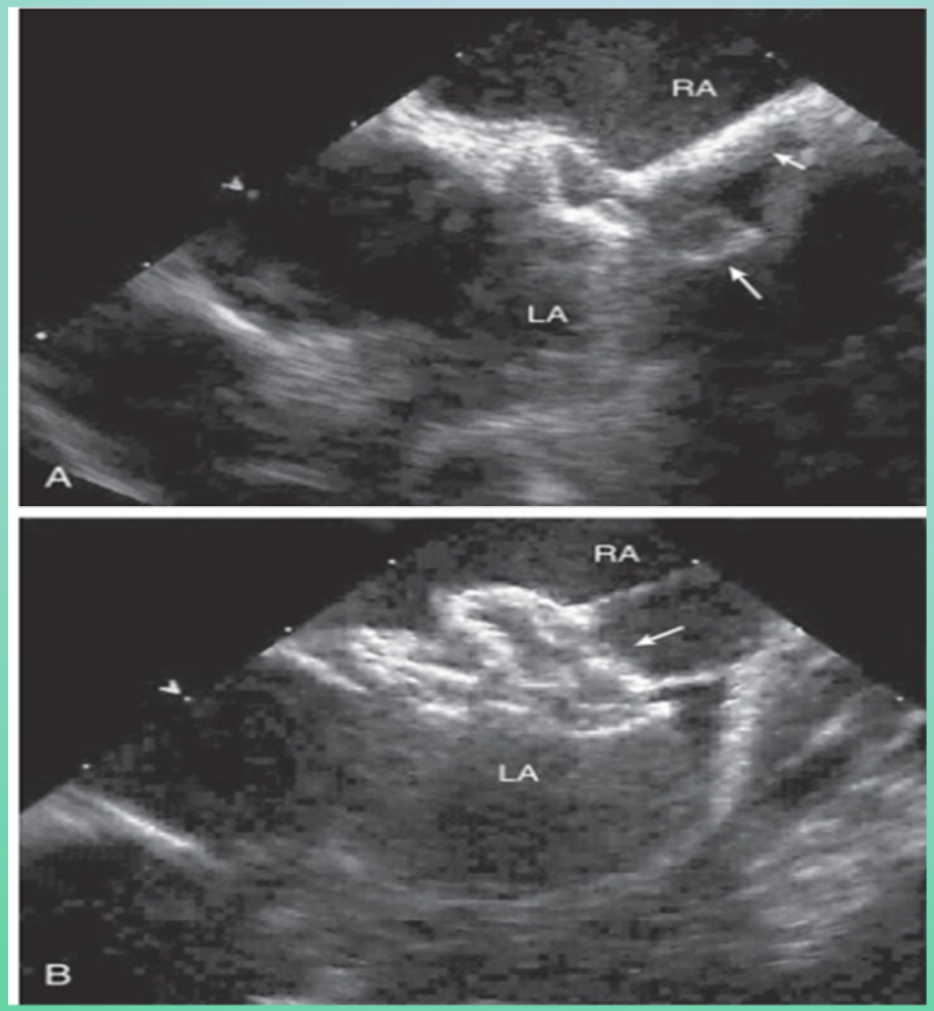
- 3- pulmonic ejection click
- 4- widely split and fixed S2
- 5- Systolic ejection murmur
- 6- Rumbling mid-diastolic murmur at LLSB

- ***Diagnosis :***
- *1- CXR:*
- Varying degrees of enlargement of the RV,RA and MPA
- Increased pulmonary vascularity
- Cardiac enlargement

- *2-ECG:*
- Normal or Rt axis deviation
- rsR′ pattern in the right precordial leads
- Right ventricular hypertrophy

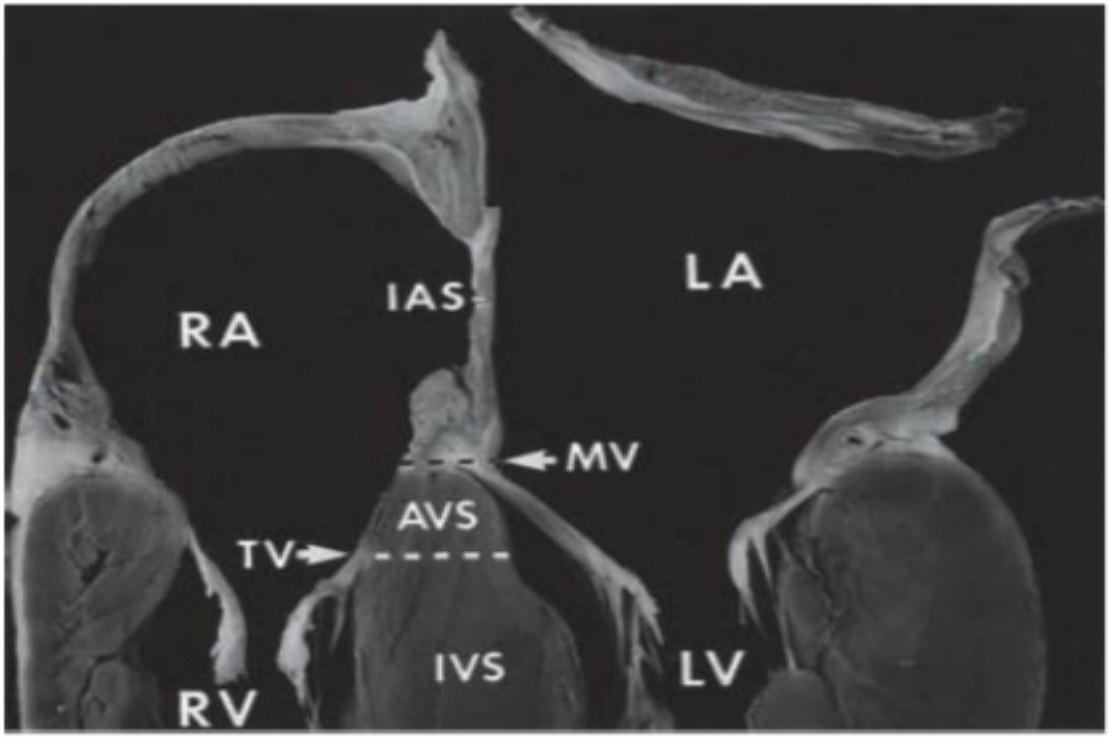
- *3-Echocariography :*
- Increased RV end-diastolic dimension
- Flattening and abnormal motion of the ventricular septum
- *4-Catheterization:*
- Pulmonary vascular disease suspected

- ***Treatment :***
- *Transcatheter device or surgical closure:*
- 1-Symptomatic patients
- 2-Asymptomatic patients => Qp:Qs ratio of at least 2 : 1
- RV enlargement.
- Elective closure is usually after the 1st yr of life and before entry into school


RA
LA
A
RA
LA
B

- ***Prognosis :***
- Small to moderate-sized ASDs => may grow smaller or close spontaneously
- ***Late manifestations:***
- Pulmonary hypertension
- Atrial dysrhythmias
- Tricuspid or mitral insufficiency
- Heart failure
- No antibiotic prophylaxis for infective endocarditis

- ***Atrioventricular Septal Defects (Atrioventricular Canal or Endocardial Cushion Defects) :***
- ***TYPES:***
- *Complete form* => single AV valve , primum ASD, inlet VSD, more common in Down syndrome
- Intermediate form => two AV valves, primum ASD, inlet VSD
- *Transitional form* => two AV valves, primum ASD, small inlet VSD, MV cleft
- *Partial form* => two AV valves, primum ASD, MV cleft


RA
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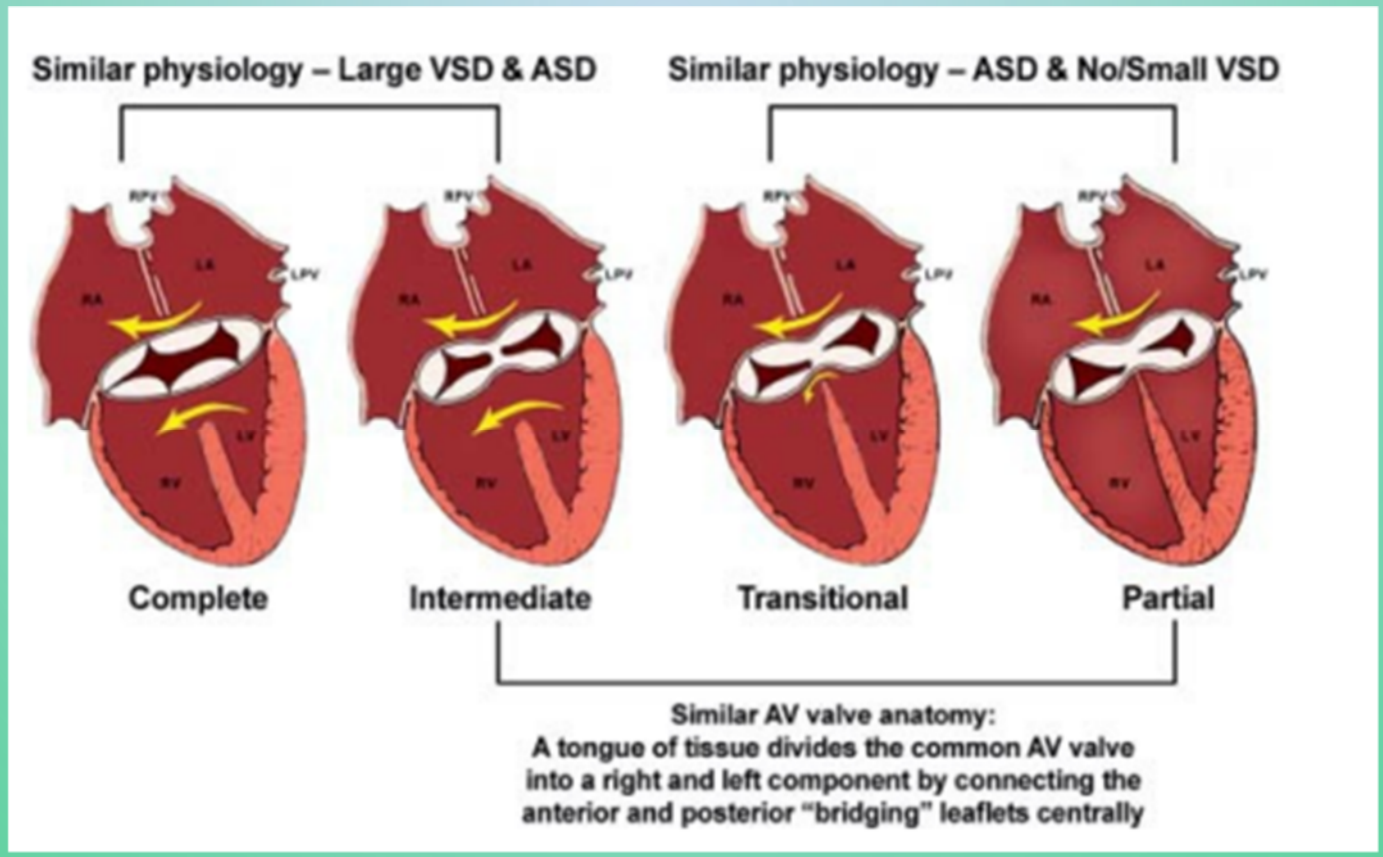
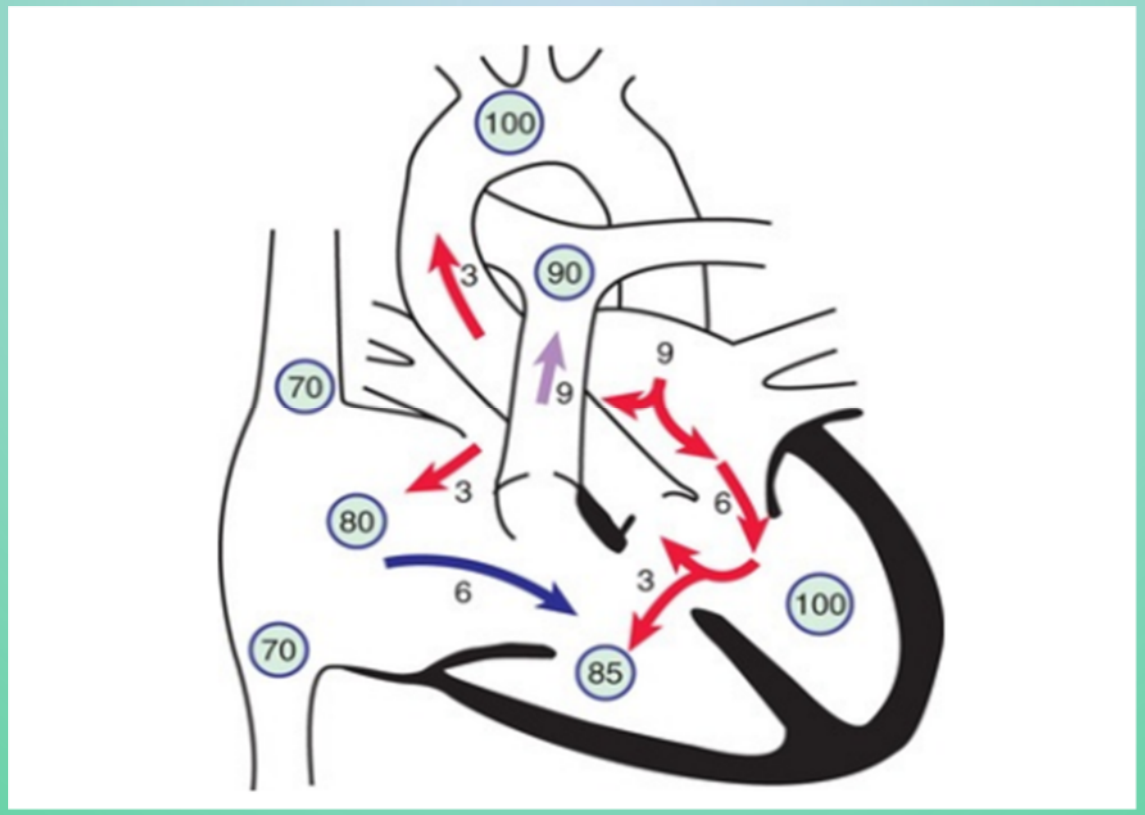
RV


Similar physiology – Large VSD & ASD
Similar physiology – ASD & No/Small VSD
RPV
LA
LPV
RA
LV
RV
Complete
Intermediate
Transitional
Partial
Similar AV valve anatomy:
A tongue of tissue divides the common AV valve into a right and left component by connecting the anterior and posterior "bridging" leaflets centrally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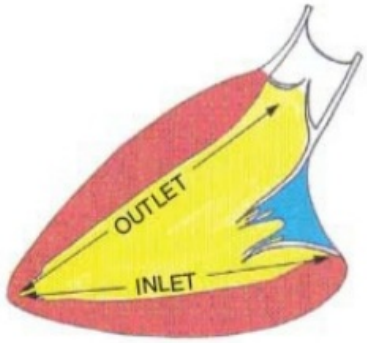
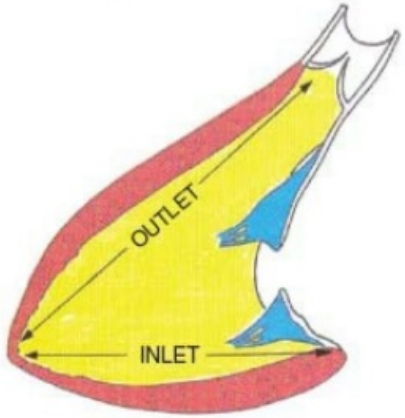
- ***Complete AVSD:***
- **Clinical Manifestations :**
- Exercise intolerance
- Easy fatigability
- Recurrent pneumonia
- Pulmonary overflow sysmptoms

- ***Physical examination:***
- Wide, fixed splitting of S2
- Systolic ejection murmur ,mid-diastolic rumbling murmur ,holosystolic murmur that radiates to the left axilla
- Hyperdynamic precordium

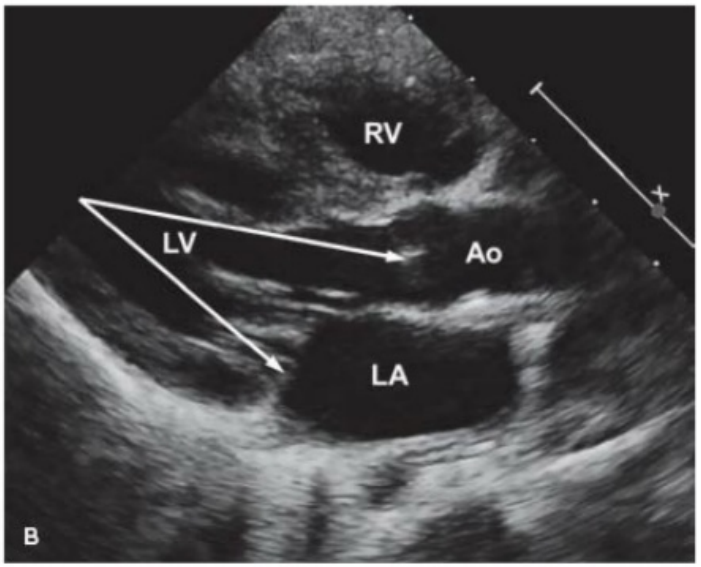
- **Diagnosis :**
- *CXR :*
- Cardiac enlargement, enlarged pulmonary artery, and increased pulmonary vascularity.
- *Echocardiography :*
- RV enlargement
- “Gooseneck” deformity of the LVOT



Normal



A Atrioventricular septal defect



- *Catheterization:*
- Rarely required to confirm the diagnosis unless pulmonary vascular disease is suspected

- ***ECG :***
- (1) Right superior axis(QRS negative in both lead I and lead aVF)
- (2) Q wave in leads I and aVL
- (3) Biventricular hypertrophy or isolated RV hypertrophy
- (4) RV conduction delay (rSR′ pattern in leads V3 R and V1)
- (5) Normal or tall P waves
- (6) Occasional prolongation of the P-R interval

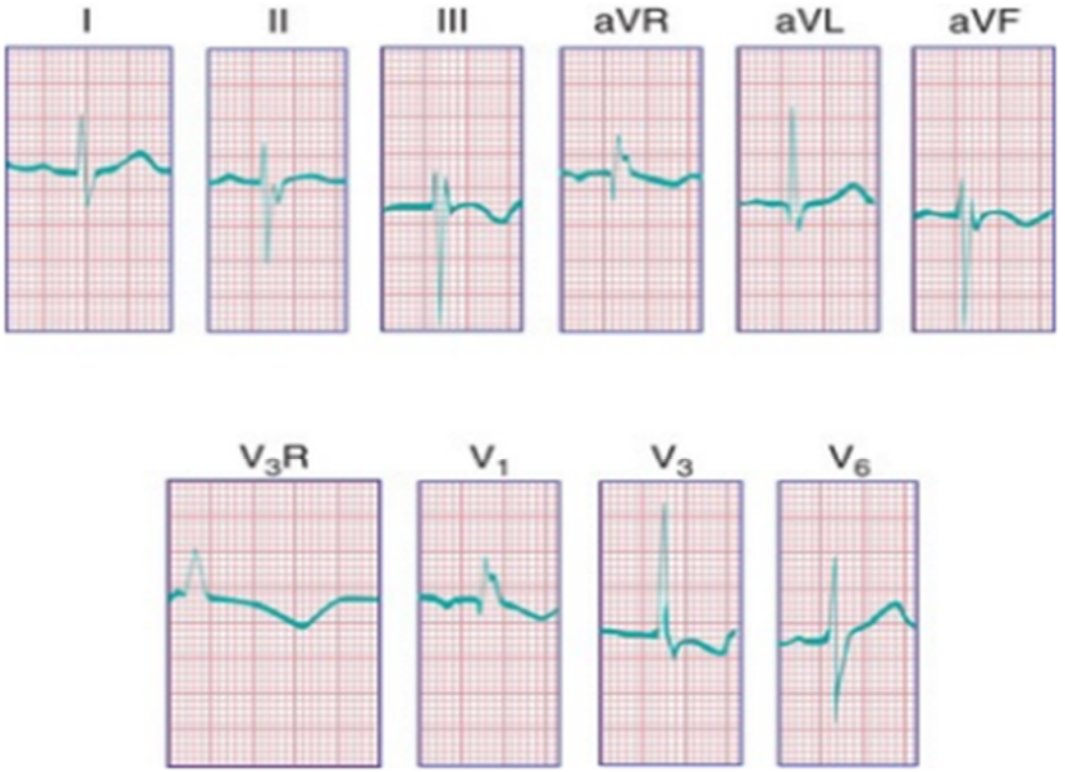
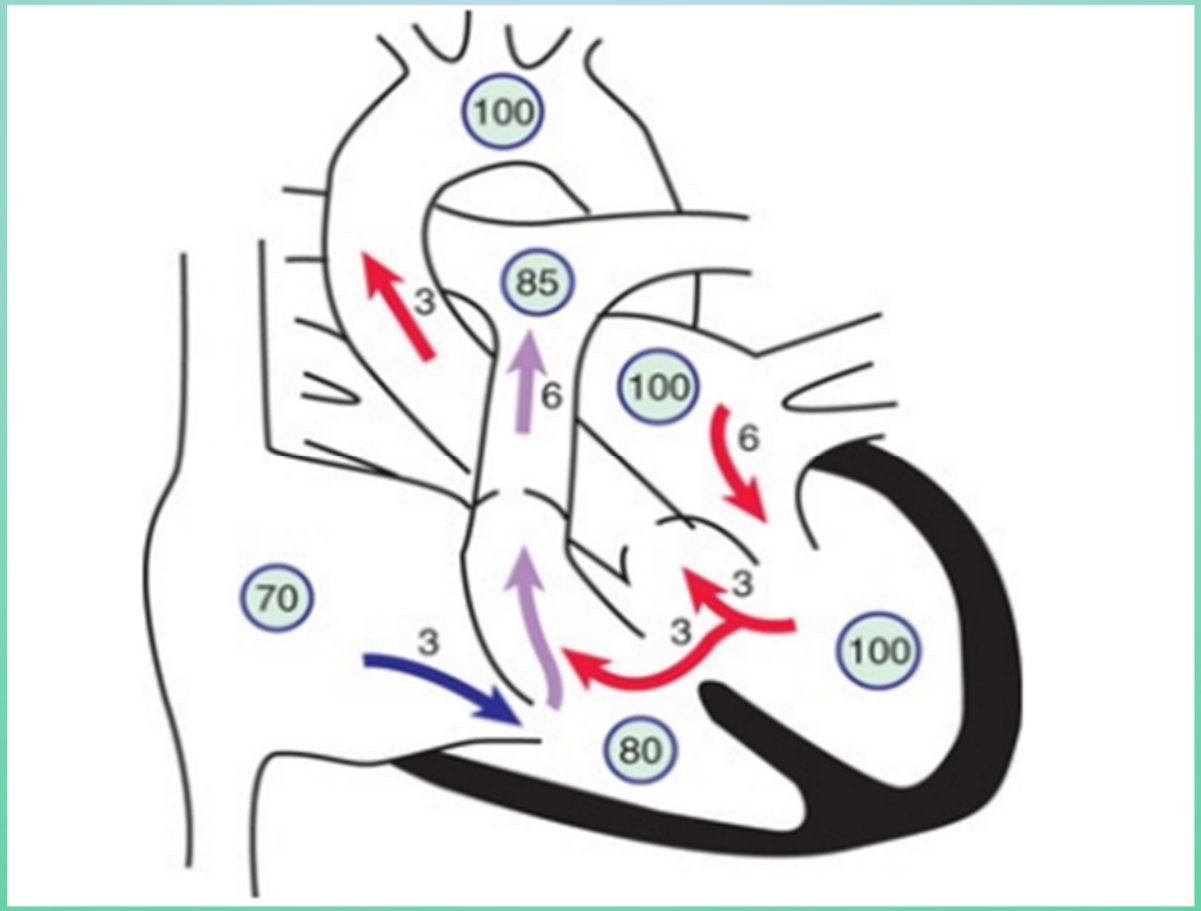


FIG. 453.5 Electrocardiogram from child with atrioventricular septal defect. Note the QRS axis of −60 degrees and the right ventricular conduction delay with an RSR′ pattern in V_1 and V_3 R (V_3 R paper speed = 50 mm/sec).

- ***Treatment :***
- Surgical intervention during infancy
- **Corrective:**
- Patching of atrial and ventricular defects and reconstruction of the AV valves.
- **Palliative:**
- Pulmonary arterial banding => not much effective in large amount of AV valve regurgitation.

- ***Postoperative complications:***
- Atrial arrhythmias
- Heart block
- Progressive narrowing of the LVOT
- AV valve regurgitation

- ***Ventricular Septal Defect :***
- The most common (25% CHD)
- ***TYPES:***
- 1-Memberanous: the most common, posteroinferior position, anterior to the septal leaflet of the tricuspid valve
- 2- Supracristal : less common, superior to the crista supraventricularis, aortic insufficiency
- 3-Muscular: midportion or apical region of the septum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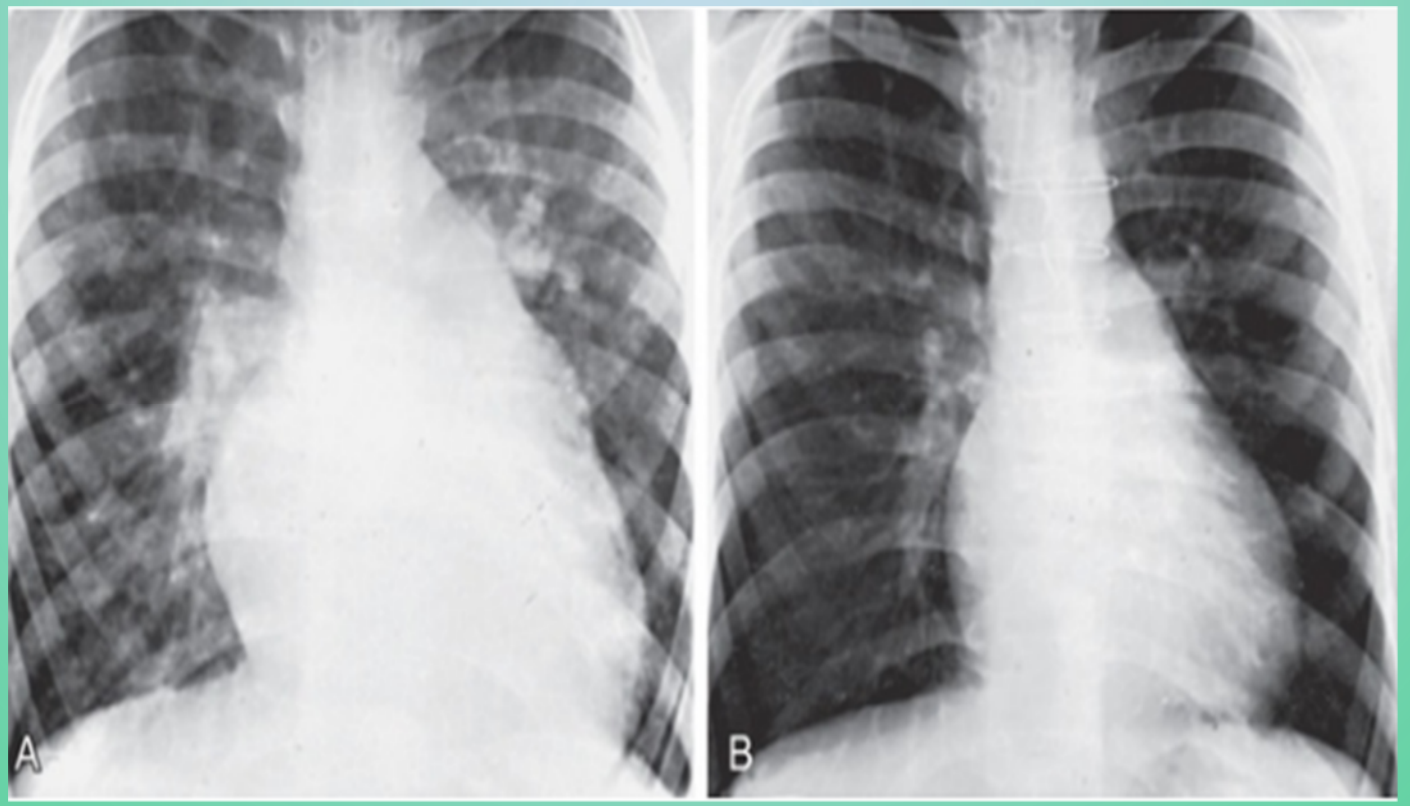
- **Clinical Manifestations :**
- Small ones :
- Asymptomatic
- Loud, harsh, or blowing holosystolic murmur and thrill in the lower left sternal border

- Large ones :
- Congestive heart failure signs: dyspnea, feeding difficulties, poor growth, profuse perspiration, and recurrent pulmonary infections
- Prominent left precordium, palpable parasternal lift, less harsh holosystolic murmur, loud P2, mid-diastolic rumble

- **Diagnosis :**
- *Small* => minimal cardiomegaly and a borderline increase in pulmonary vasculature, normal ECG or LV hypertrophy.
- *Large* => gross cardiomegaly, increased pulmonary vascular markings , pulmonary edema, pleural effusions.
- **The ECG** => biventricular hypertrophy; notched P waves
- **Echocardiography and catheterization** are other diagnostic modalities


A
B

- **Treatment :**
- Small defects close spontaneously during the 1st year of life(30–50%), muscular > membranous
- Unoperated small VSDs => increased incidence of arrhythmia, subaortic stenosis, and exercise intolerance
- No prophylaxis for infective endocarditis except for dental hygiene

Surgery indications:

- 1-Symptomatic large defects uncontrolled medically
- 2-Infants 6-12 mo of age with moderate to large defects and pulmonary hypertension, even if the symptoms are controlled by medication
- 3-Qp:Qs ratio greater than 2 : 1.
- 4-Supracristal VSD of any size due to AR

- ***Patent Ductus Arteriosus :***
- Functional closure soon after birth, usually within the 1st wk of life
- Female/males ratio 2 : 1
- Associated with maternal rubella infection during early pregnancy
- Common problem in premature infants

- **Clinical Manifestations :**
- Heart failure
- Retardation of physical growth
- Bounding peripheral arterial pulses and a wide pulse pressure
- Prominent apical impulse

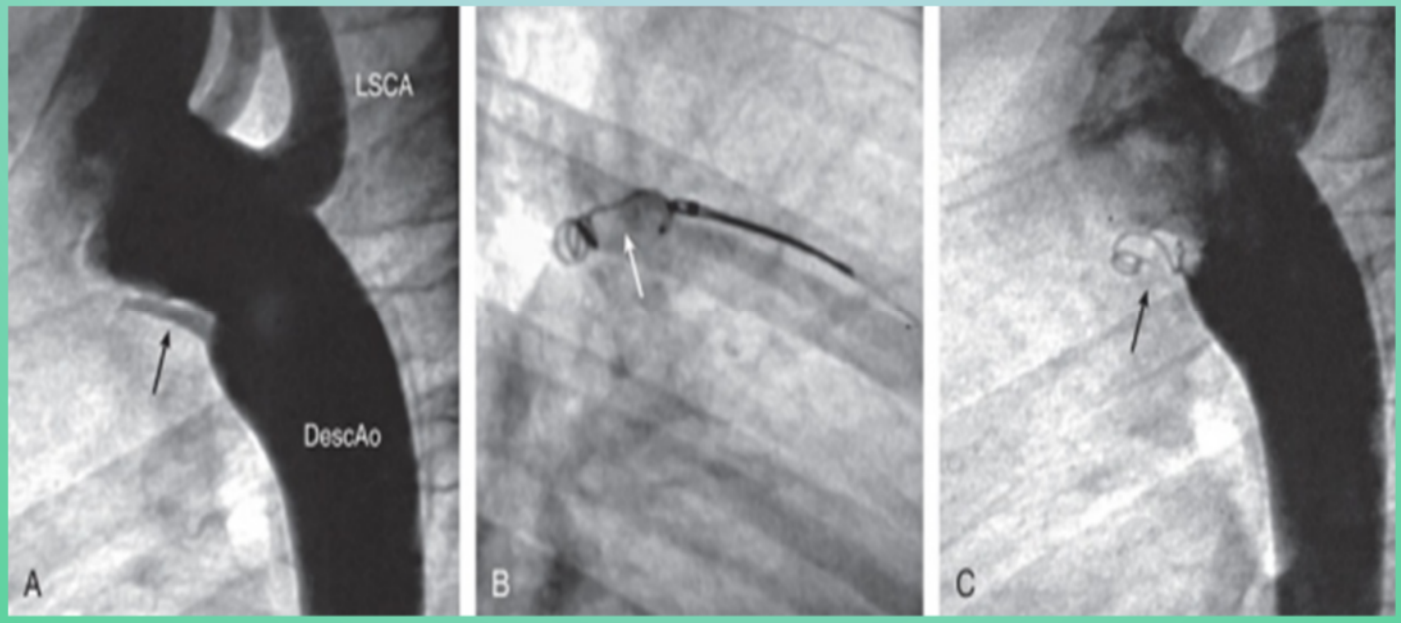
- A thrill in the 2nd left interspace may radiate toward the left clavicle, down the left sternal border, or toward the apex
- Continuous machinery-like murmur(systolic in PH cases)
- a low-pitched mid-diastolic murmur at the apex

- **Diagnosis :**
- *CXR* : increased cardiothorasic ratio and PVM , prominent Aortic knob
- *ECG* : LVH or BVH
- *Echocardiography:* LAE and LVE, systolic or diastolic (or both) retrograde turbulent flow in the pulmonary artery
- *Catheterization* : to evaluate PVR in special cases

- **Prognosis and Complications :**
- Spontaneous closure of the ductus after infancy is extremely rare
- Infective endarteritis
- Pulmonary or systemic emboli
- Pulmonary hypertension (Eisenmenger syndrome)

- ***Rare complication :***
- Aneurysmal dilation of the pulmonary artery or the ductus
- Calcification of the ductus
- Noninfective thrombosis of the ductus with embolization
- Paradoxical emboli

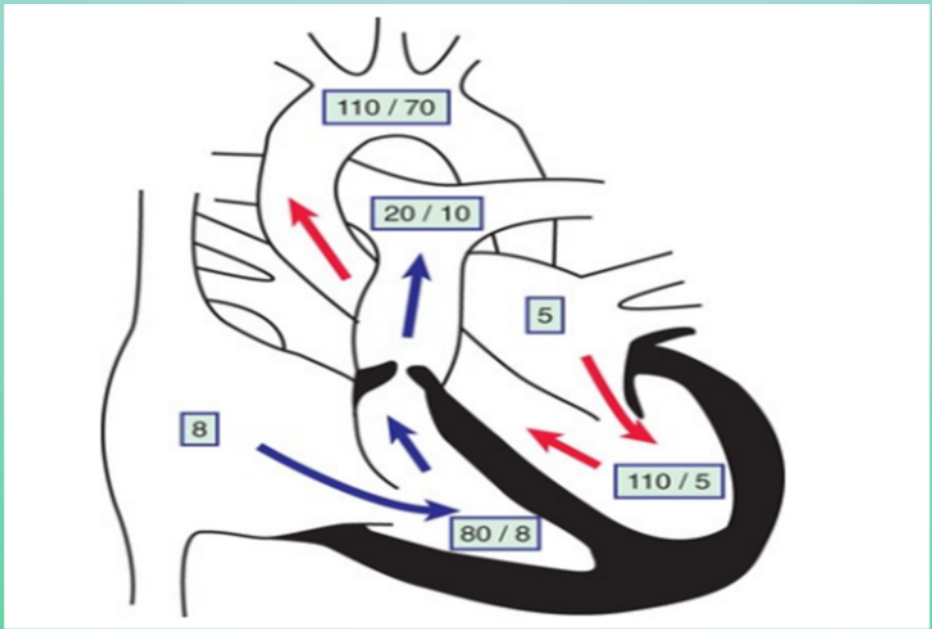
- **Treatment :**
- Catheter or surgical closure.



- **Obstructive Lesions :**
- ***Pulmonary Valve Stenosis With Intact Ventricular Septum :***
- 7–10% of all congenital heart defect
- *Associations :*
- Noonan syndrome

- LEOPARD syndrome : lentigines, electrocardiographic abnormalities, ocular hypertelorism, pulmonary stenosis, abnormalities of genitalia, retardation of growth, deafness syndrome
- Alagille syndrome

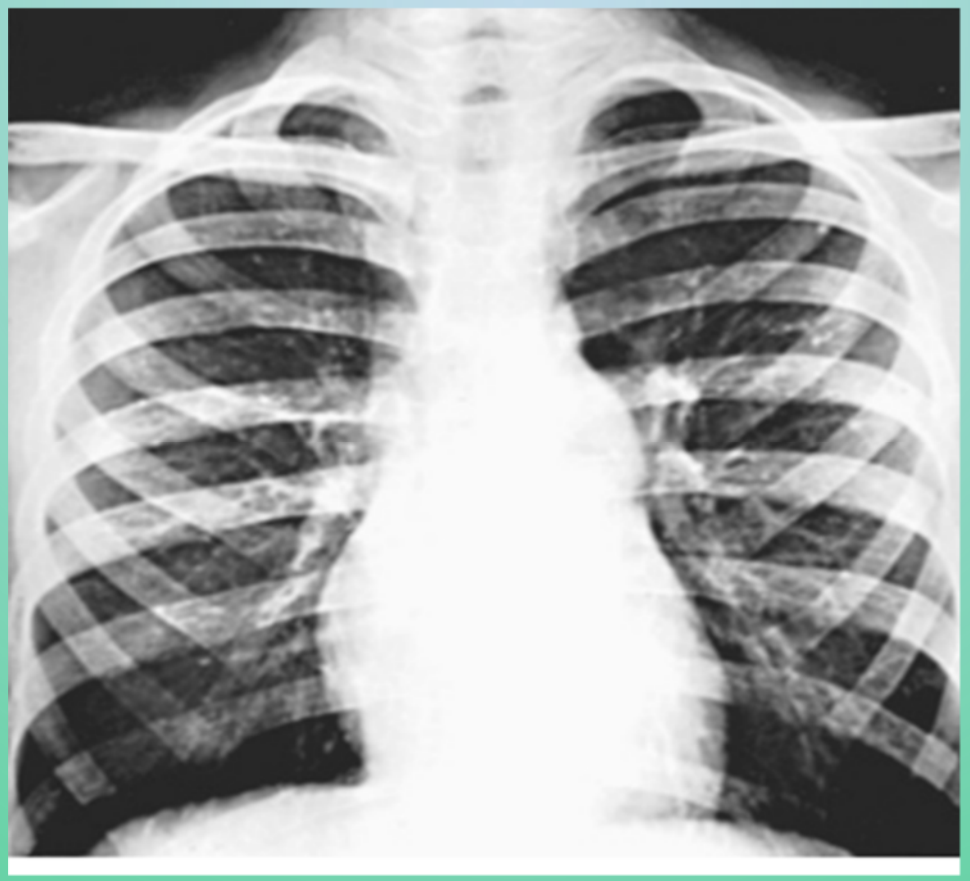
- Elevated RV pressure
- Normal or decreased PAP
- RV hypertrophy
- Cyanosis in case of critical PS and intracardiac shunt



- **Clinical Manifestations :**
- ***Mild pulmonary stenosis :***
- Normal venous pressure and pulse
- Sharp pulmonic ejection click at LUSB
- Wide S2 splitting
- Normal ECG with mild RVH
- RV-PA pressure gradient of ≤30 mm Hg

- ***Moderate pulmonic stenosis :***
- Slightly elevated venous pressure(prominent a wave)
- RV lift
- Wide S2 splitting
- RVH with a prominent spiked P wave in ECG
- Mild cardiomegaly , pulmonary vascularity may be normal or slightly decreased
- RV-PA pressure gradient of 30-60 mm Hg

- ***Severe pulmonary stenosis :***
- RV failure(hepatomegaly, peripheral edema, and exercise intolerance)
- Mild to moderate cyanosis(in case of ASD,PFO)
- Elevated venous pressure(prominent a wave)
- Cardiomegaly, RV lift
- Inaudible P2
- Loud, long, and harsh systolic ejection murmur
- RVH, tall, spiked P wave.



- **Treatment :**
- *Moderate or severe cases:*
- Balloon valvuloplasty => choice
- Severely thickened pulmonic valves(Noonan syndrome) may require surgical intervention.
- Recurrence is unusual after successful treatment except in those patients with extremely dysplastic valves

- ***Peripheral Pulmonary Stenosis :***
- *Associations:*
- Congenital heart disease
- Congenital rubella syndrome.
- Williams syndrome
- Alagille syndrome
- Mild and transient form of peripheral pulmonic stenosis in the immediate newborn period

- Systolic ejection murmurs over the chest, either anteriorly or posteriorly
- Severe PPS => RVH , RAE, increased CT ration
- Echo has limitation in visualizing distal pulmonary branches
- CT, MRI and catheterization are diagnostic.

- ***Aortic Stenosis:***
- 5% of cardiac malformations
- Bicuspid aortic valve, one of the most common congenital heart lesions
- More frequent in males (3 : 1)

- ***TYPES:***
- *Valvular*
- *Sub valvular* : fibromuscular shelf, associated with MS, COA(shone complex), progressive
- *Supra valvular*: the least-common , sporadic, familial, or associated with Williams syndrome

- **Williams syndrome :**
- Developmental delay (IQ range: 41-80)
- Elfin facies (full face, broad forehead, flattened bridge of the nose, long upper lip, and rounded cheeks)
- Idiopathic hypercalcemia of infancy
- Loquacious personality
- Hypersensitivity to sound, hypoplastic nails, dental anomalies, joint hypermobility, nephrocalcinosis, hypothyroidism

- **Clinical Manifestations :**
- Heart failure
- Fatigue, angina, dizziness, or syncope
- Sudden death
- **Critical AS :**
- LV failure and signs of low cardiac output, cardiomegaly, pulmonary edema, weak pulses , pale or grayish skin ,diminished urine output

- ***Physical exam :***
- Early systolic ejection click which does not vary with respiration
- Diminished S1, paradoxically split S2
- Systolic ejection murmur at the right upper sternal border
- Suprasternal notch thrill
- The louder, harsher (higher pitch), and longer the murmur, the greater is the degree of obstruction

- **Diagnosis :**
- **ECG:** left ventricular hypertrophy (LVH) and LV strain (inverted T waves in left precordial leads)
- **CXR:** prominent ascending aorta, normal aortic knob and heart size, valvular calcification

- **Echocardiography :** LVH and the thickened and domed aortic valve, associated anomalies, endocardial fibroelastosis
- **Catheterization :** usually performed in conjunction with aortic balloon valvuloplasty, demonstrates the magnitude of the pressure gradient

- **Treatment :**
- ***Balloon valvuloplasty :***
- Moderate to severe valvular aortic stenosis.
- Peak-to-peak systolic gradient exceeds 60-70 mm Hg at rest or less when symptomatic or ECG changes

- ***Surgery :***
- Extremely dysplastic aortic valves not amenable to balloon therapy or in patients who also have subvalvar or supravalvar stenosis.
- Regardless of whether surgical or catheter treatment has been carried out, aortic insufficiency or calcification with restenosis is likely to occur again

- **AVR :**
- Homograft => no anticoagulant, less longevity
- Mechanical => anticoagulant, more longevity
- Ross procedure => translocation of aorta and pulmonary valve

- **Prognosis :**
- Moderate to severe degrees of AS => no participation in active competitive sports.
- Mild AS=> sports participation is less severely restricted
- Annual follow up
- No Prophylaxis against infective endocarditis

- ***Coarctation of the Aorta :***
- Juxtaductal : 98% , occur just below the origin of the left subclavian artery at the origin of the ductus arteriosus
- Male/ female :2/1
- ***Pathophysiology :***
- 1-Decreased anterograde blood flow through the aortic valve
- 2-Abnormal extension of contractile ductal tissue into the aortic wall.

- ***Associations:***
- Turner syndrome
- BAV (>70%)
- Mitral valve abnormalities (a supravalvular mitral ring or parachute mitral valve)
- Subaortic stenosis

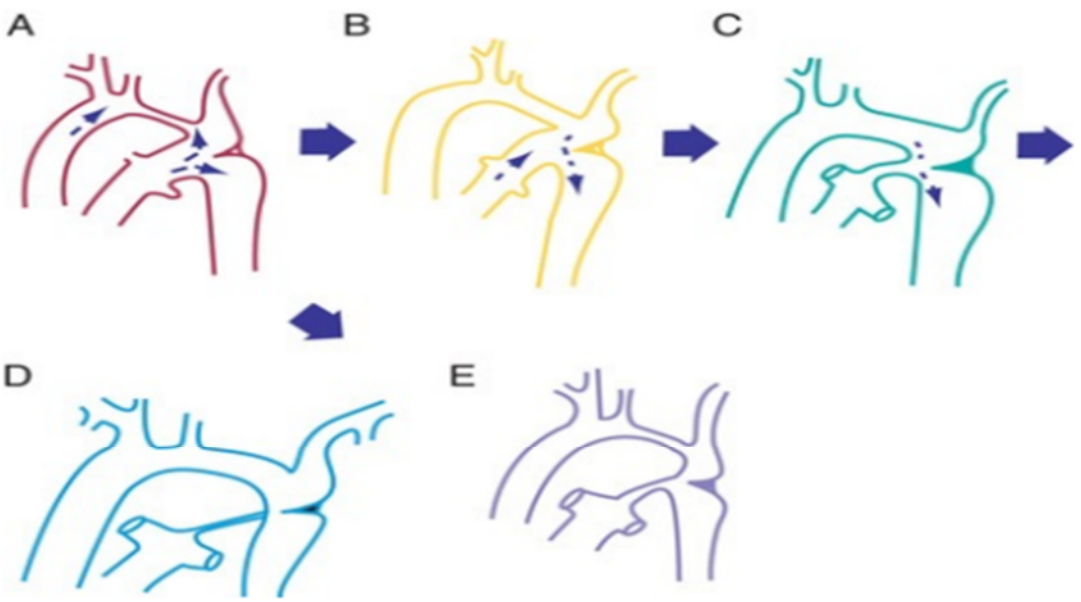
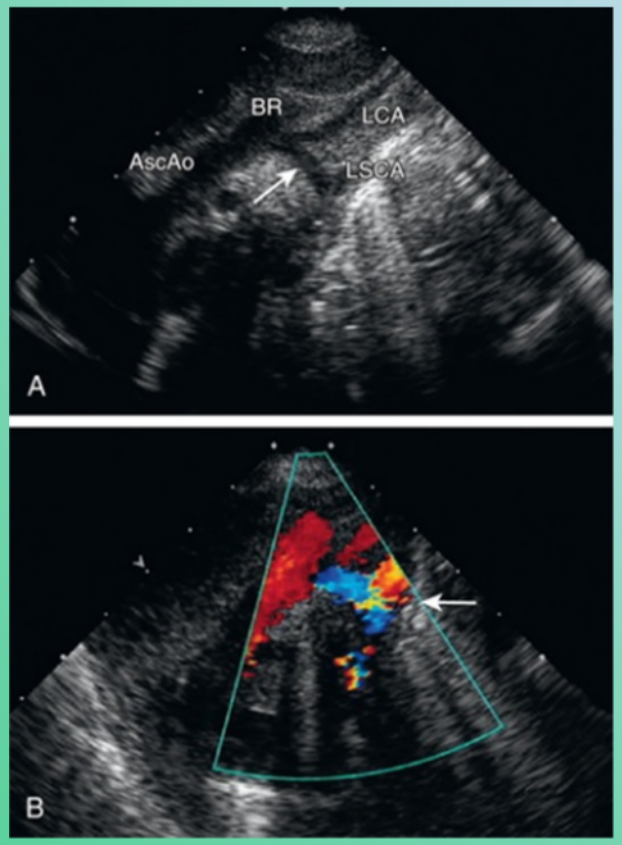


FIG. 454.7 Metamorphosis of coarctation. A, Fetal prototype with no flow obstruction. B, Late gestation. The aortic ventricle increases its output and dilates the hypoplastic segment. Antegrade aortic flow bypasses the shelf via the ductal orifice. C, Neonate. Ductal constriction initiates the obstruction by removing the bypass and increasing antegrade arch flow. D, Mature juxtaductal stenosis. The bypass is completely obliterated, and intimal hypoplasia on the edge of the shelf is aggravating the stenosis. Collaterals develop. E, Persistence of the infantile-type fetal prototype. An intracardiac left-sided heart obstruction precludes an increase in antegrade aortic flow before or after birth. Both isthmus hypoplasia and a contraductal shelf are present. Lower-body flow often depends on patency of the ductus. (From Gersony WM: Coarctation of the aorta. In Adams FH, Emmanouilides GC, Riemenshneider T, editors: *Moss heart disease in infants, children, and adolescents,* ed 4, Baltimore, 1989, Williams & Wilkins.)

- **Clinical Manifestations :**
- Weakness or pain/claudication (or both) in the legs after exercise
- Asymptomatic mostly
- Differential cyanosis
- Hypertension
- Disparity in pulsation and BP in the arm and leg
- Radial-femoral delay

- Signs of lower-body hypoperfusion, acidosis, and severe heart failure(neonates)
- ***Physical exam :***
- A short systolic murmur along the left sternal border at the 3rd and 4th intercostal spaces, well transmitted to the left infrascapular area and occasionally to the neck
- Systolic or continuous murmurs over the chest laterally and posteriorly

- **Diagnosis :**
- ***CXR*:**
- Cardiac enlargement and pulmonary congestion, Rib notching
- ***ECG* :**
- LVH, BVH
- ***Echocardiography* :**
- ***CT, MRI***
- ***Catheterization***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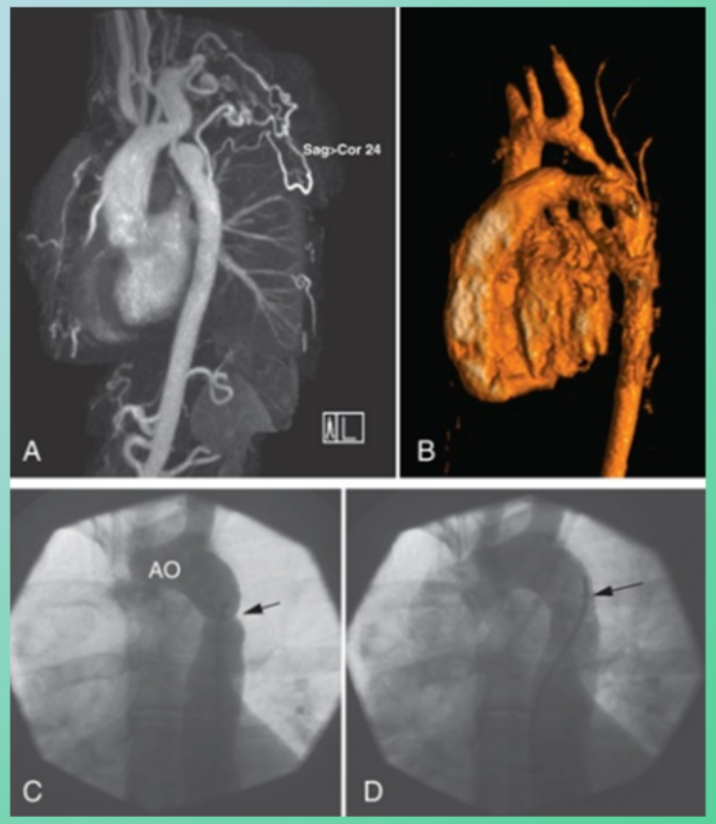
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- **Treatment :**
- Neonates => PGE1, surgical repair
- Infants => anticongestive drugs, surgical repair
- Surgery(choice in native COA)
- Aortic angioplasty(choice in ReCOA)
- Stent replacement
- Rebound hypertension: In the immediate postoperative course, requires medical management

- ***Operative complications:***
- Spinal cord injury from aortic cross-clamping (if the collaterals are poorly developed)
- Chylothorax
- Diaphragm injury
- Laryngeal nerve injury
- If a left subclavian flap approach is used, the radial pulse and BP in the left arm are diminished or absent