Cardiology review lecture 1

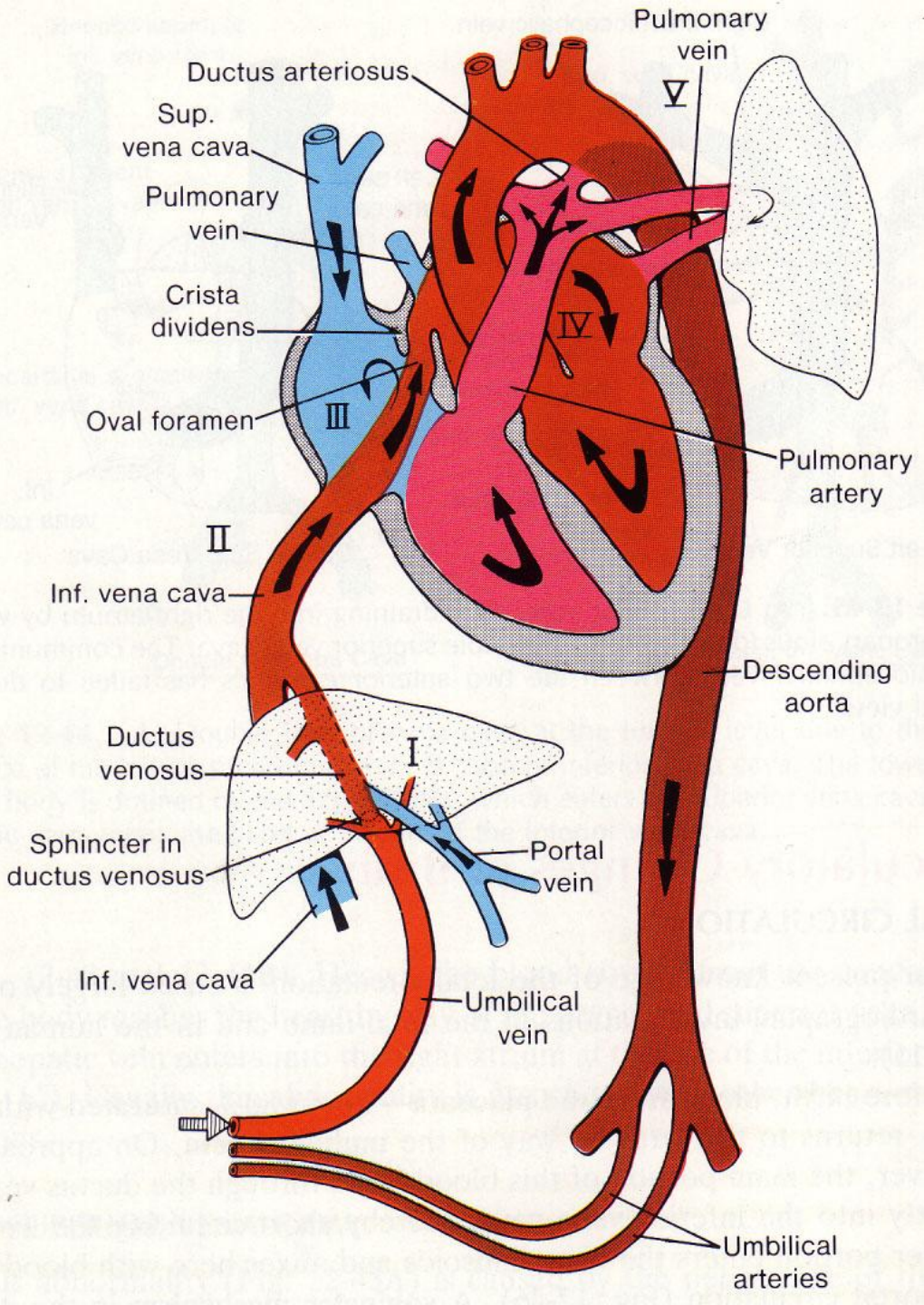
Congenital heart disease

Congenital Heart Disease

- Commonest group of life threatening anomalies
- 8/1000 live births
  - VSD 30-50%, PDA 10%, ASD 7%.
  - PS 7%
  - Coarctation 6%

Principle differences in fetal circulation compared to post-natal circulation:

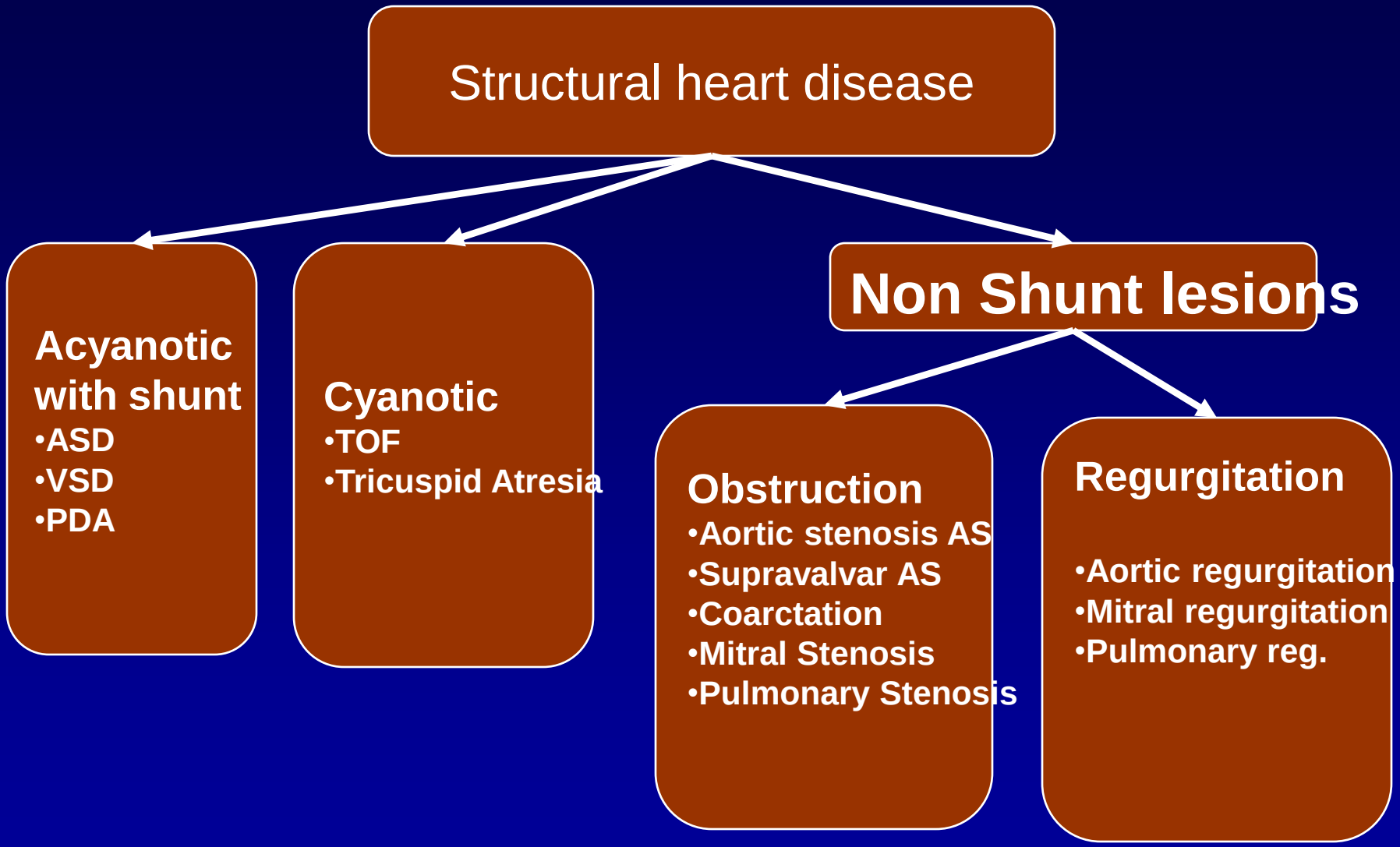
- Combined ventricular output
- Three critical anatomic communications
  - PFO (from RA to LA)
  - PDA (from PA to AO)
  - Ductus venosus (from UV to IVC)
- Organ responsible for oxygenation is Placenta



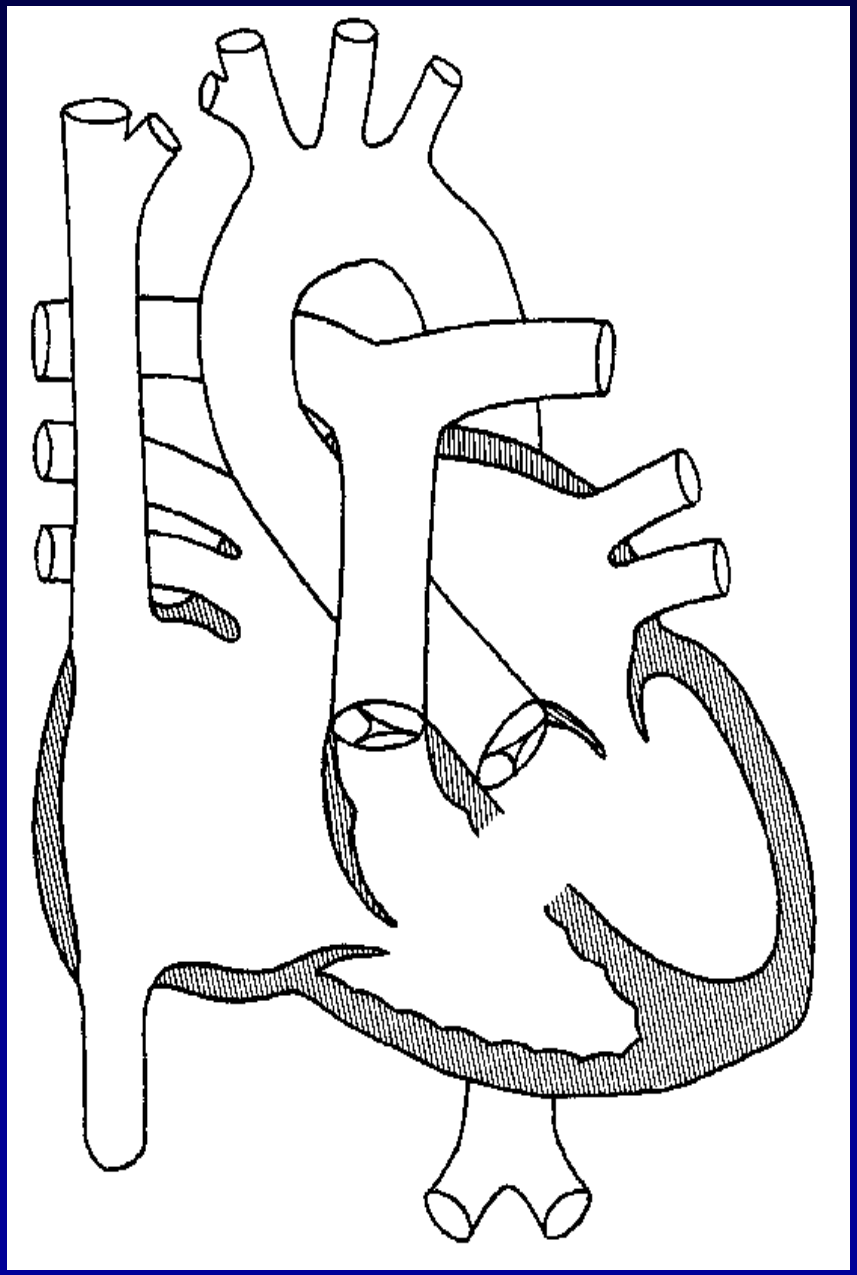
Transition From the Fetal Circulation

- Pulmonary vascular resistance falls
- Ductus venosus and ductus arteriosus close
- Right-to-left shunting through foramen ovale ceases

Timing of these events determines the timing of presentation of congenital heart defects


Structural heart disease
Acyanotic with shunt
•ASD
•VSD
•PDA
Cyanotic
•TOF
•Tricuspid Atresia
Non Shunt lesions
Obstruction
•Aortic stenosis AS
•Supravalvar AS
•Coarctation
•Mitral Stenosis
•Pulmonary Stenosis
Regurgitation
•Aortic regurgitation
•Mitral regurgitation
•Pulmonary reg.

How will this baby look at 12 hours of age?



Presentation First 24 Hours

- **Critically ill" - like asphyxia**
- **Cyanosis (may be mild)**
- Pure heart failure is uncommon
- Murmur

Symptoms of cardiac failure in the infant

- Poor feeding
- Failure to thrive
- Fatigue

Left to Right shunt

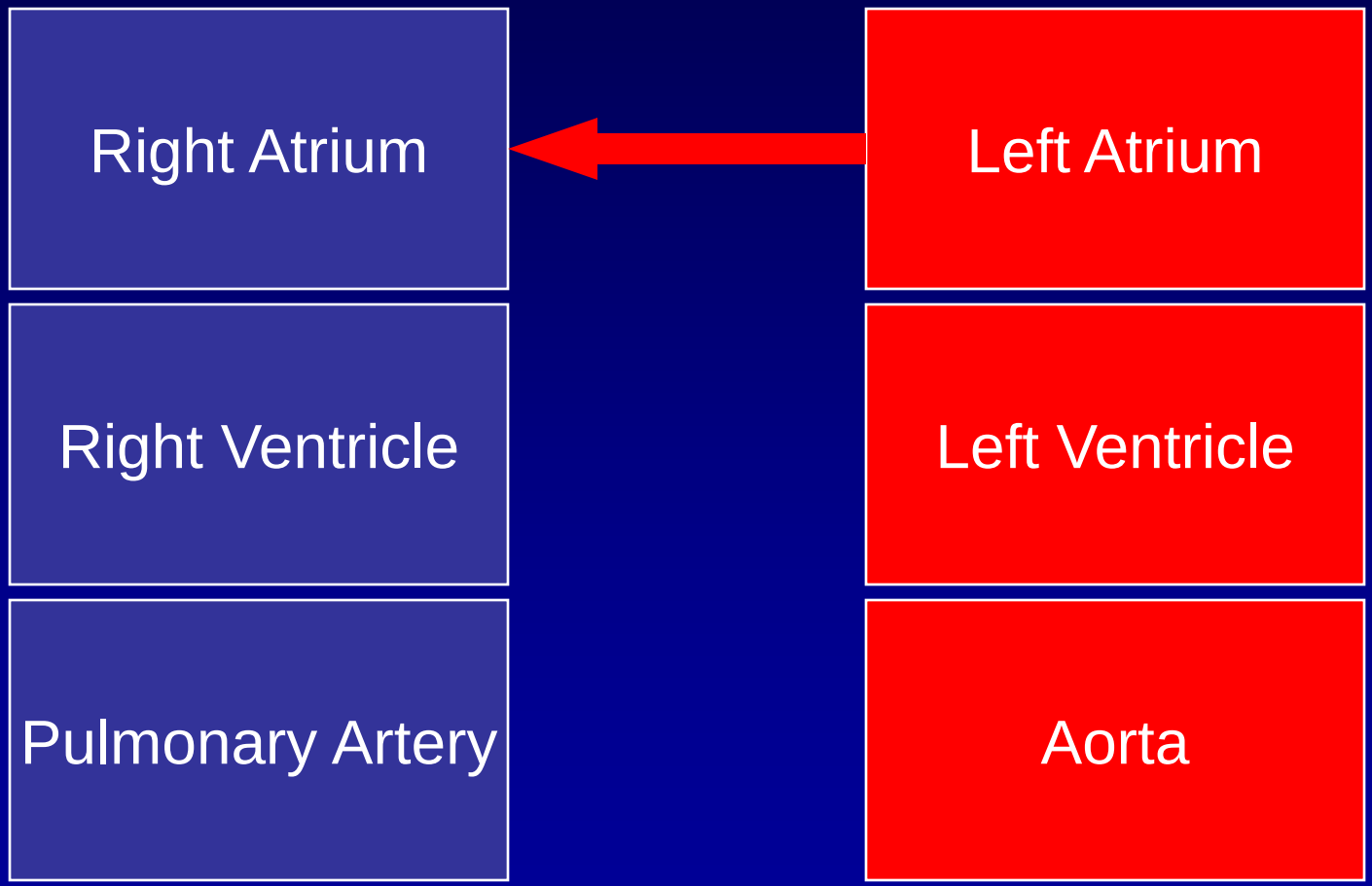
- Portion of fully oxygenated pulmonary venous blood bypassing the systemic flow and going back to the lungs
- ➔ In-effective pulmonary blood flow
- ➔ S&S of Increased pulmonary blood flow

Left to right shunts

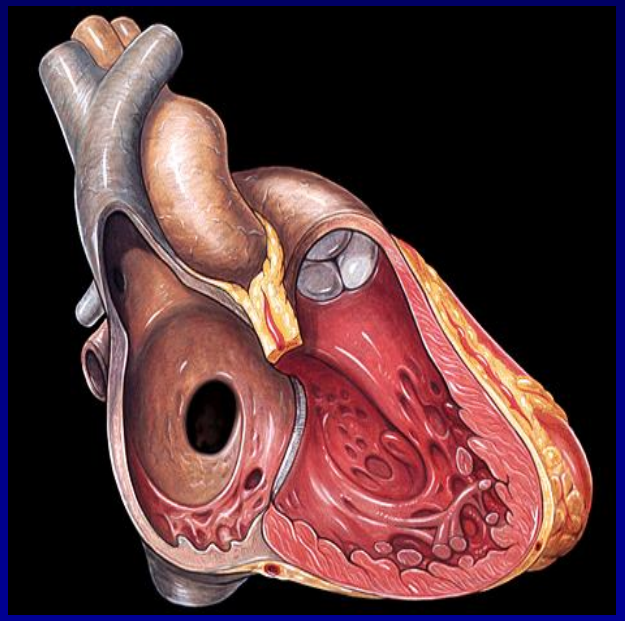
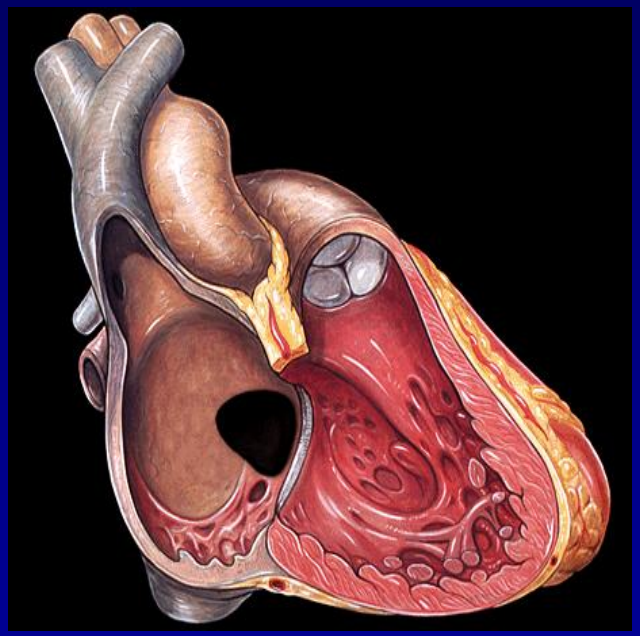
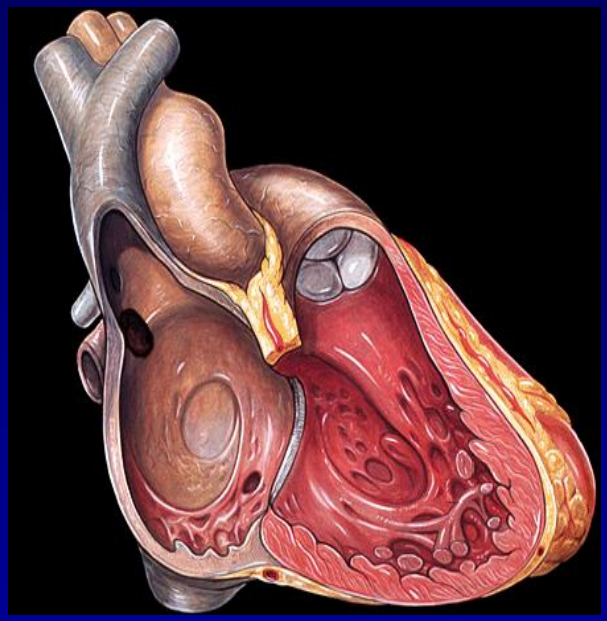
Physiologic effect of the shunt is dependent on three factors:

1) Location of the shunt
2) Size of the defect
3) Relative pulmonary and systemic vascular resistance (or ventricular compliance in case of atrial level shunts)

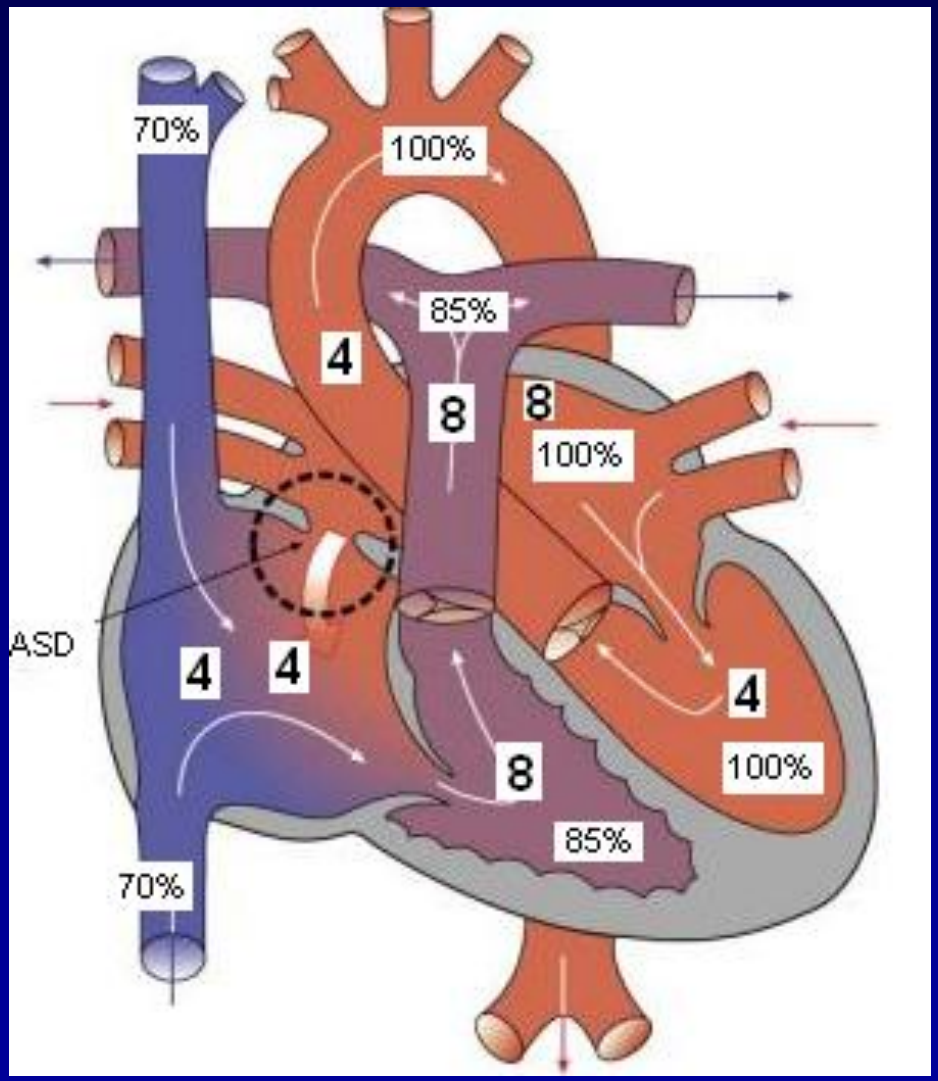
Atrial level shunts



Atrial Level shunt: Anatomy

Atrial Level Shunt: Physiology



- ASD causes **volume** load on the right atrium, and right ventricle leading to dilation of these structure
- Majority of the shunt occurs during diastole (ventricular filling)
- Cardiac output is well maintained even in large ASD's

Symptoms: ASD

- Usually asymptomatic
- Symptoms of CHF, tachypnia, or decreased exercise tolerance are rare
- Most ASD's are discovered incidentally or due to a heart murmur

Examination

- Normal in young infants
- Wide, fixed S2
- Ejection systolic murmur

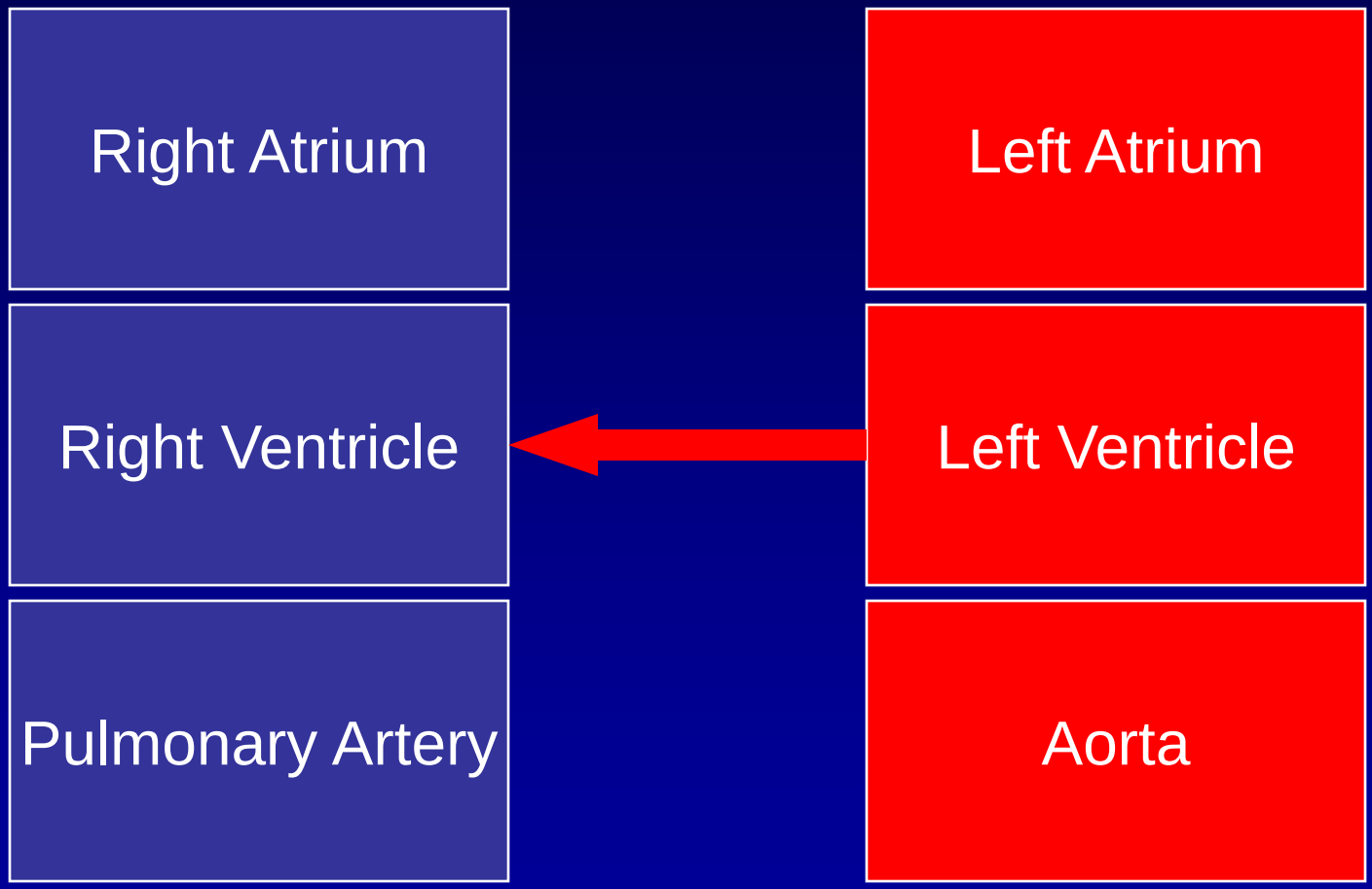
Diagnostic studies

- ECG: Usually Normal, but may reflect right atrial and right ventricular dilatation
- CXR: May show increased pulmonary vascular markings and dilated pulmonary trunk
- Echocardiography is diagnostic, and important to determine the type and physiologic effect of the defect

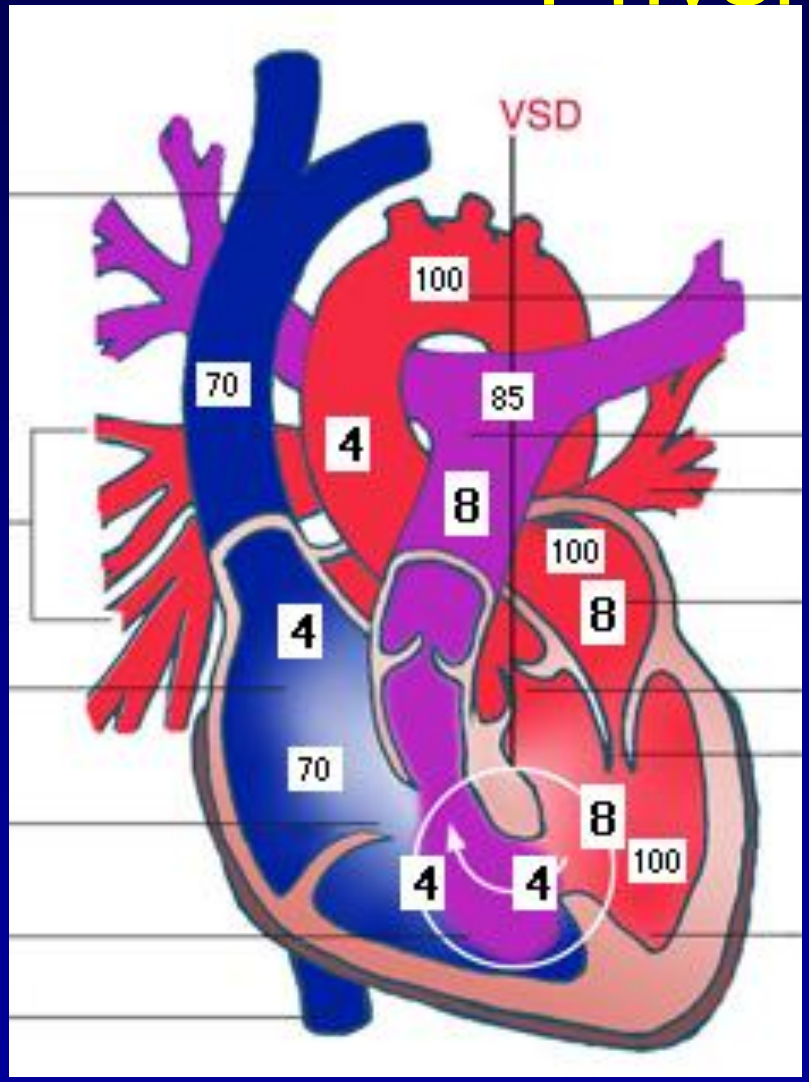
Management

- No restriction from activity
- No medications
- Observation for spontaneous closure if secundum type and no significant volume overload on the right ventricle

Ventricular level shunt



Ventricular Level Shunt: Physiology



- VSD causes **Pressure** load on the right ventricle causing RVH, and Volume load on the left atrium and ventricle leading to dilation
- Shunt occurs during Systole (ventricular emptying)
- Cardiac output is well maintained even in large VSD's

Symptoms: VSD

- Newborns with VSD are usually well
- Mod-Large VSD in infants may cause CHF symptoms once Pulmonary vascular resistances
- Decreased feeding, diaphoresis and respiratory distress
- Compensated patients deteriorate rapidly with infection

Examination

- Newborns may not have murmur (High PVR)
- Displaced apex beat
- Pan-systolic murmur
- Small muscular defects may have short murmurs
- Loud S2
- S3

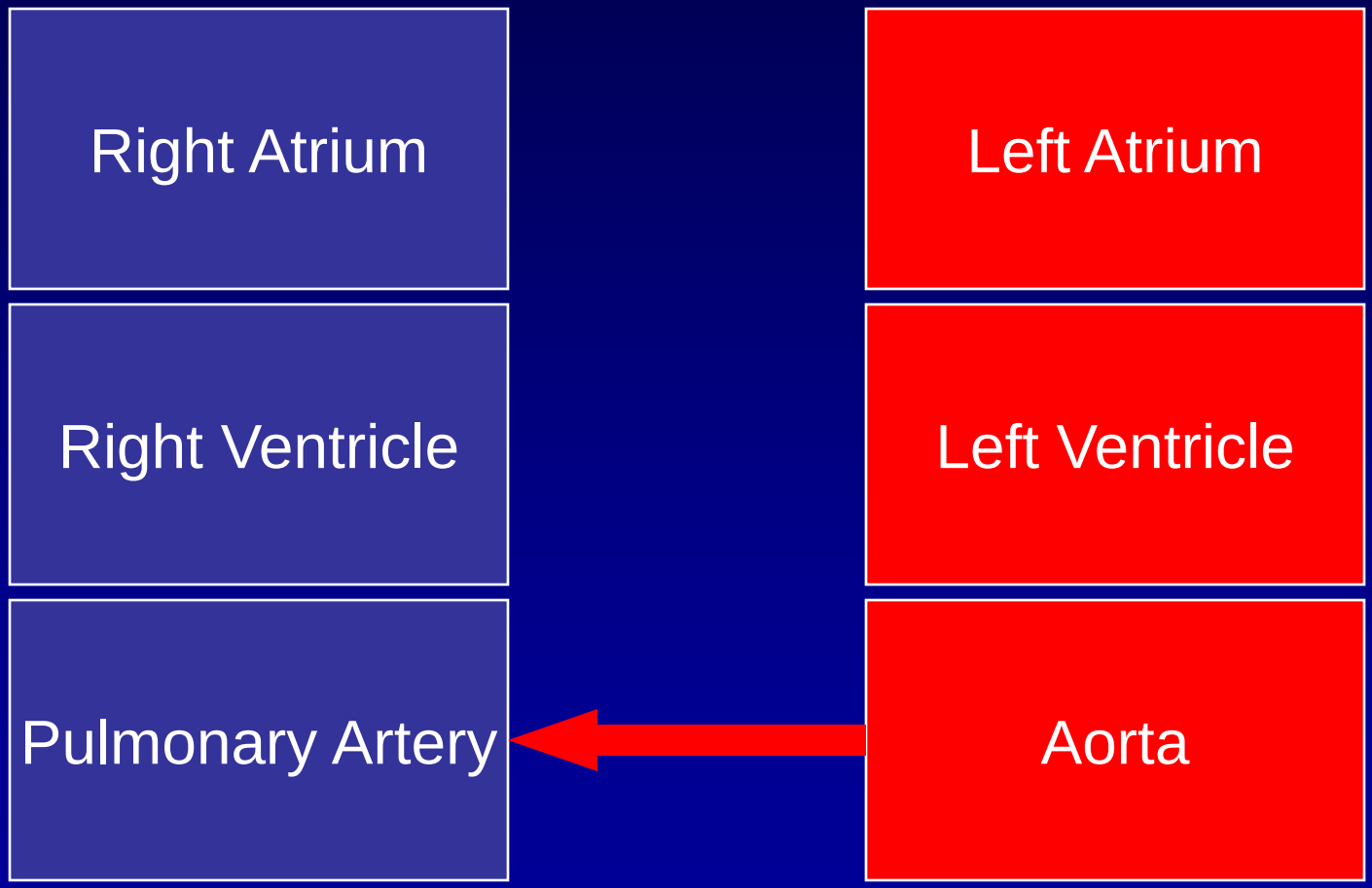
Diagnostic studies

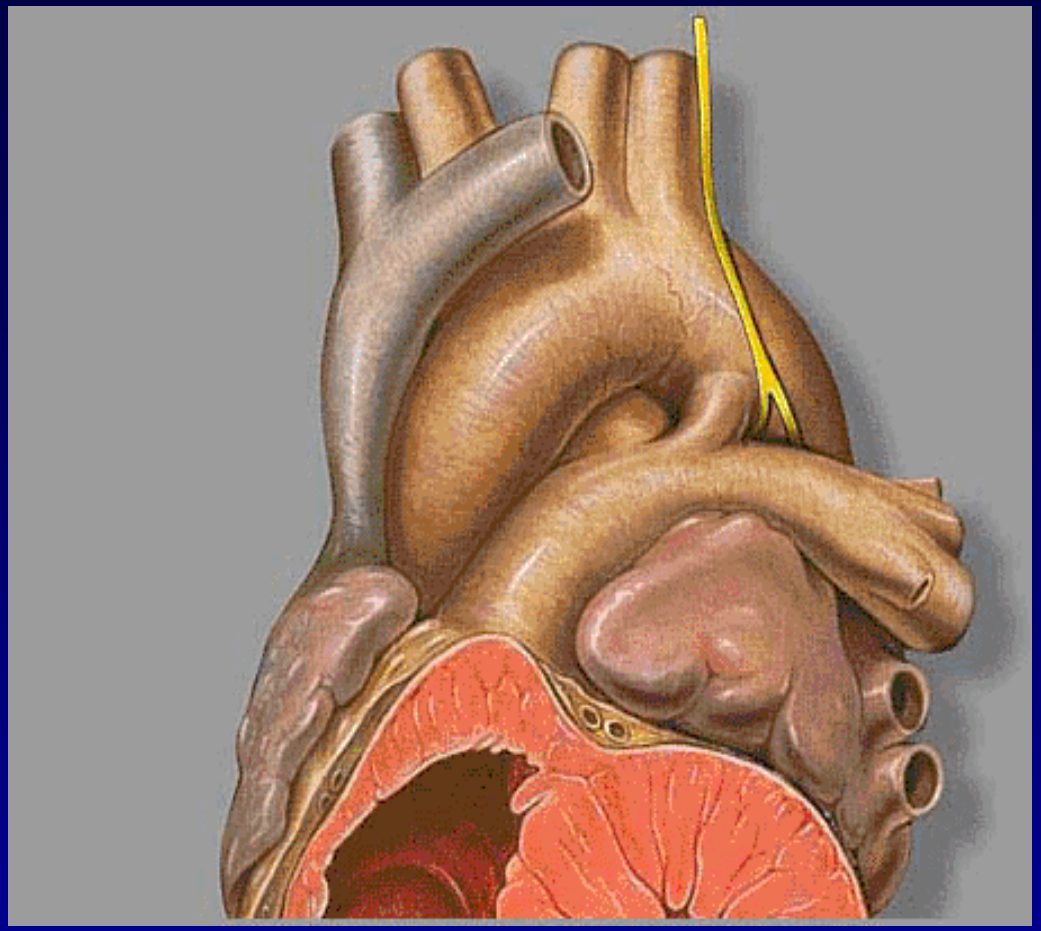
- ECG: (beyond infancy)
  - Left axis deviation
  - LVH
  - Left atrial dilation
- CXR shows cardiomegally, and increased pulmonary vascular markings in significant VSD’s

Management

- No restriction from activity
- No SBE prophylaxis (the newer guidelines)
- Spontaneous closure is common in small and moderate perimembranous and muscular defects
- Surgical treatment is the standard treatment for symptomatic VSD’s

PDA


Right Atrium
Left Atrium
Right Ventricle
Left Ventricle
Pulmonary Artery
Aorta



- Located just distal to the origin of the left subclavian artery
- 1 / 2500 to 1/ 5000 live births
  - Increased incidence with prematurity
- 12% of all CHD
- Female to male ratio = 2:1

- **Associations:** Asphyxia,Chromosomal anomalies, Birth at high altitudes, Congenital Rubella, Drugs, Genetic?

Normal postnatal closure

Functional closure

- Usually occurs within the first 24 hours
- Stimulated by:
  - High pO2 (more prominent effect with increased GA)
  - Interruption of Prostaglandins
    - Most important factor in ductal patency
    - Produced by the placenta and the ductal tissue
    - Metabolized by the lungs
    - Ductal tissue is much more sensitive to prostaglandins at earlier gestations

Complete “anatomic” closure (fibrosis)

- Usually occurs in the first 2-3 weeks

PDA physiology

- Flow across the PDA depends on
  - size
  - Pressure gradient
  - relative resistence (SVR vs. PVR)
- Cardiac consequences
  - Increased pulmonary blood flow and venous return
  - Left ventricular dilation (volume load on LV)
  - Right ventricular hypertrophy (pressure load on RV)
  - ➔ Similar physiologic effect as VSD
  - Unlike VSD it causes diastolic hypotension ➔ decreased myocardial coronary perfusion ➔ may worsen congestive heart failure

Presentation

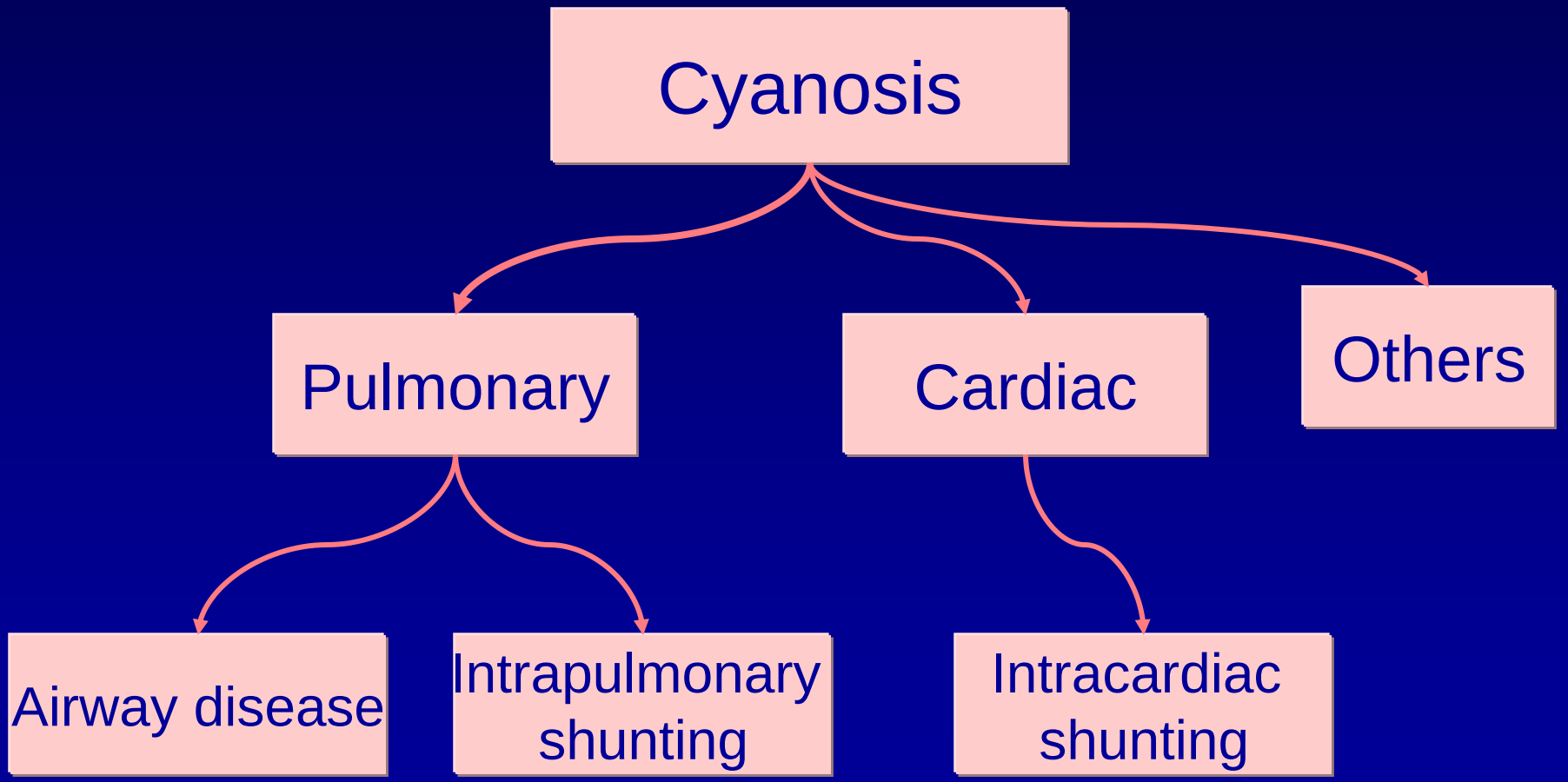
- In premature infants:
- Hyperdynamic circulation
  - Wide pulse pressure
  - Heart murmur
  - Increased oxygen requirement occurs due to pulmoanry congention, and increased respiratory distress
- In Term infants: Usually asymptomatic in the first few weeks of life (similar to VSD's)

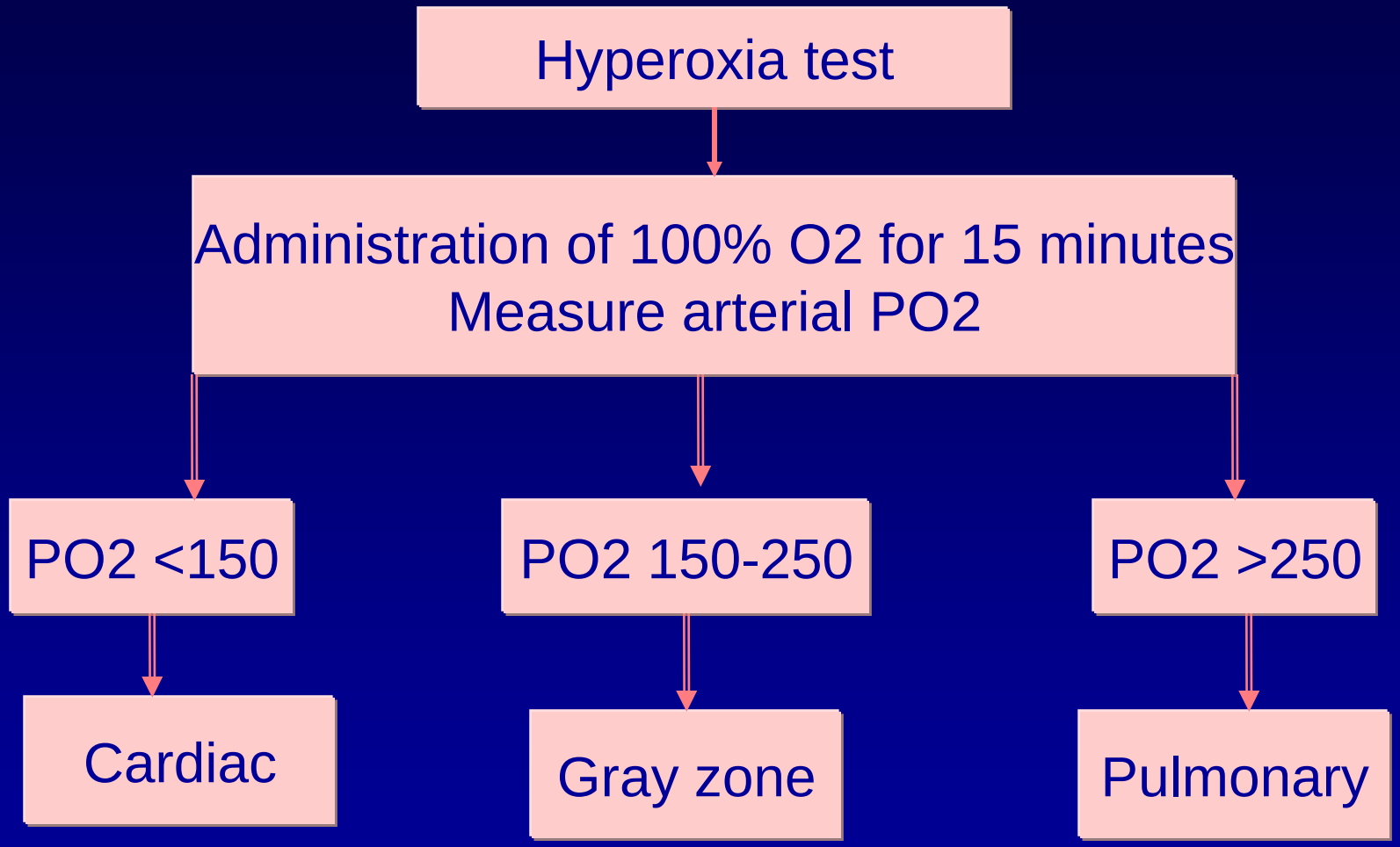
Management

- Asymptomatic PDA's require no treatment before age of 1 year, elective closure can usually be done by catheterization
- Symptomatic PDA's
  - Symptomatic treatment of CHF (diuresis, inotropic support, and vasodilators)
  - Avoid lowering PVR (avoid oxygen, alkalosis, NO)
  - Medical closure (Indomethacin IV, preferably before day 10 of life, Ibuprofen IV has similar effect)
  - Surgical closure in refractory cases

Cyanotic heart disease (right to left shunt)

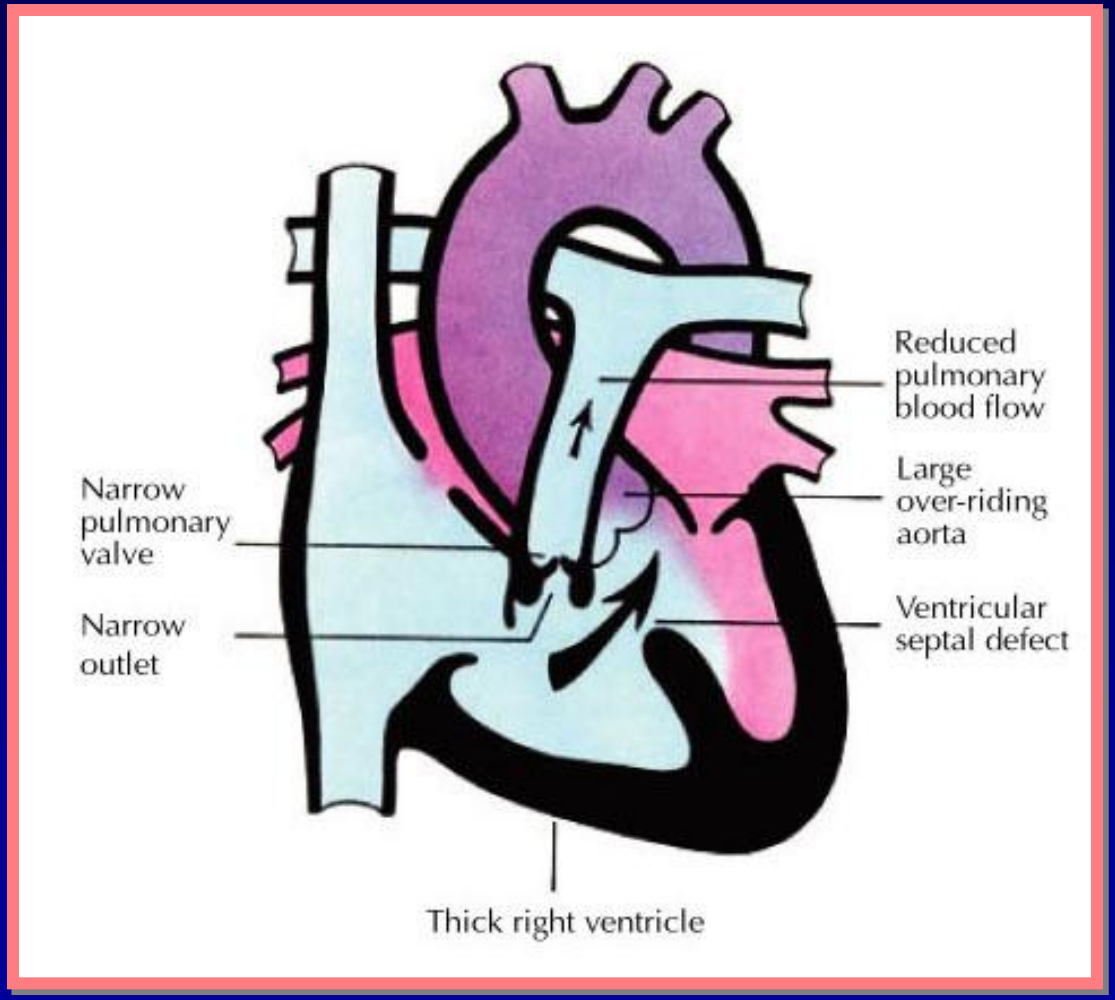
General causes of Cyanosis




Hyperoxia test
Administration of 100% O2 for 15 minutes
Measure arterial PO2
PO2 <150
PO2 150-250
PO2 >250
Cardiac
Gray zone
Pulmonary

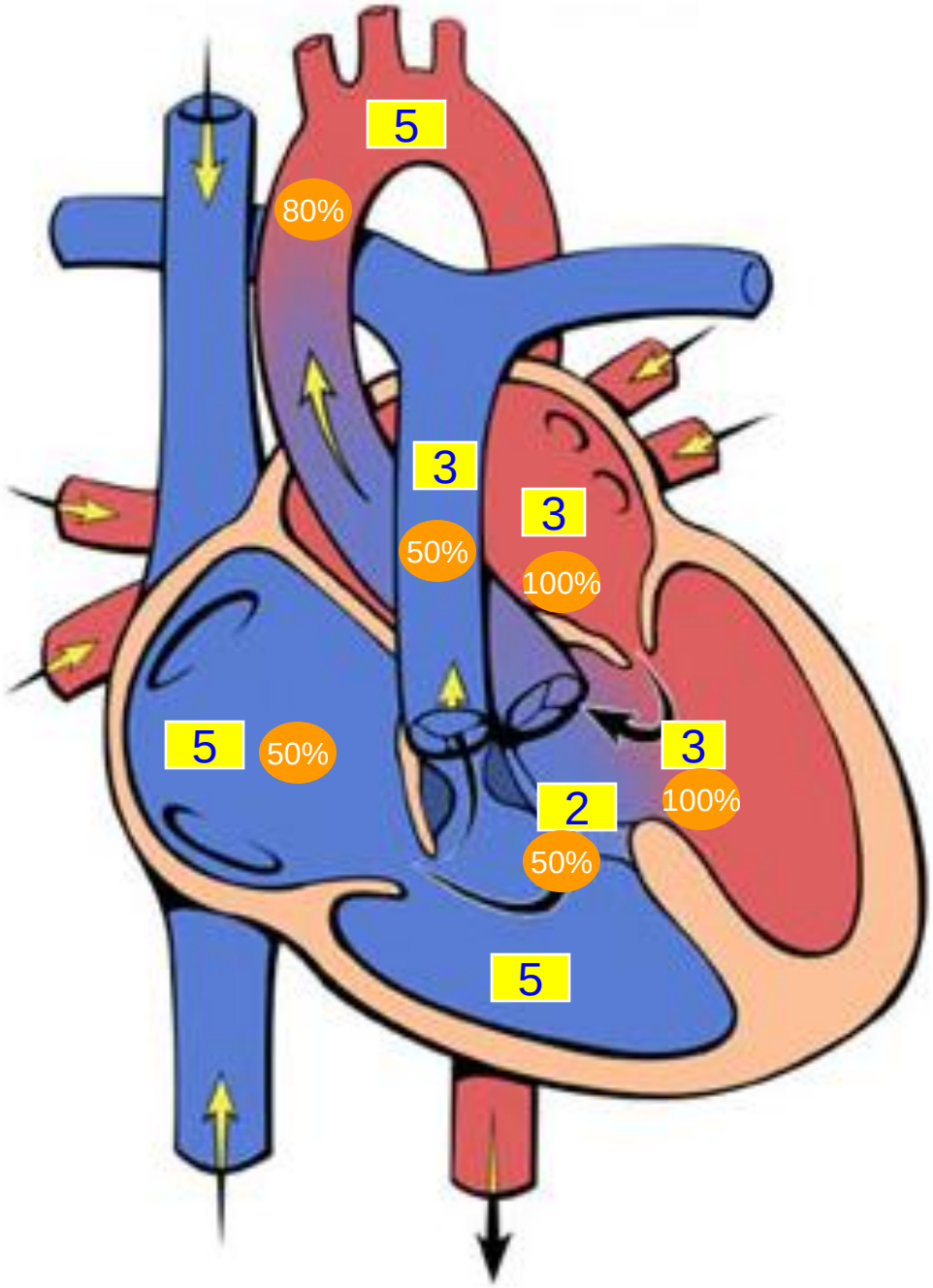
Tetralogy of Fallot (TOF)

1. ASD
2. VSD
3. Overriding aorta
4. RV hypertrophy



TOF

- Most common cyanotic heart lesion
- 3rd most common overall
- TOF is approximately 10% of CHD
- In the extreme form there is complete pulmonary atresia (PA/VSD), 2% of CHD
- Exact cause of TOF is uncertain
- Generally both TOF and PA/VSD are isolated findings
- Not typically found with syndromes
  - De Lange, Goldenhar, Klippel-Feil
- TOF is seen with malformation assoc
  - VACTERL, CHARGE, Velo-cardio-facial

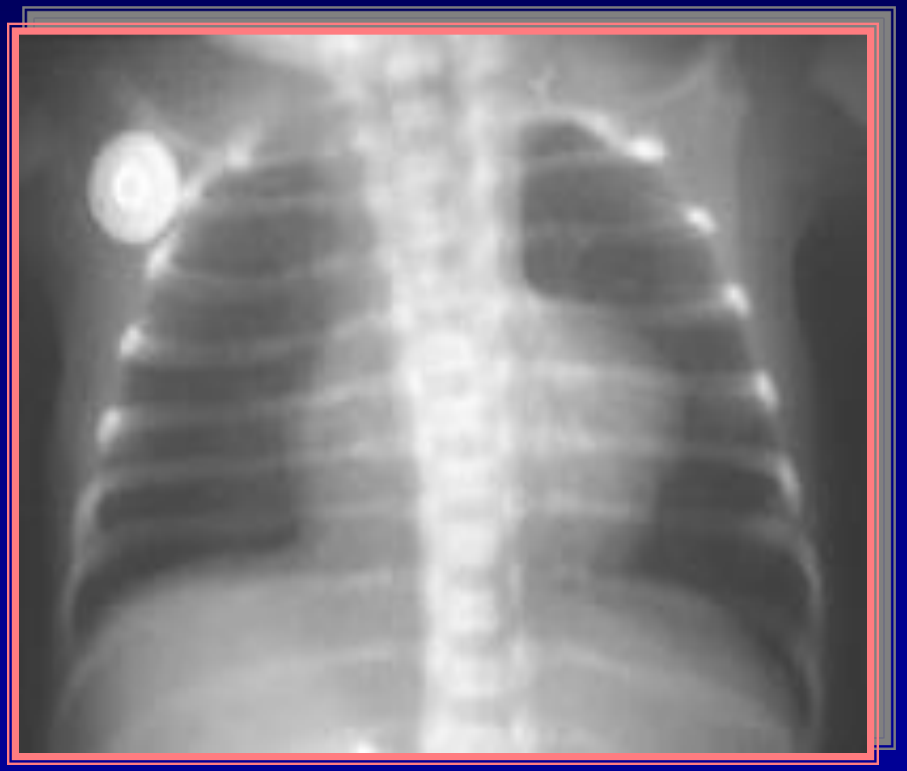

5
80%
3
3
50%
100%
5
50%
3
100%
2
50%
5

Clinical Features

- Asymptomatic infant with murmur is very common in the usual TOF patients
- Murmur of RVOT obstruction can be confused with VSD in infancy
- Cyanosis
  - Typically appears between 6wks and 6 months in the unrepaired infant
  - Nail beds and mucous membranes
  - May be present at rest or only with agitation/exercise
- Persistent cyanosis and clubbing if not repaired

Diagnostic studies

- CXR “coeur en sabot”
  - Normal heart size
  - Upturned apex
  - Concave upper left heart border
  - Normal or decreased pulmonary vascular markings
- Echocardiography is diagnostic

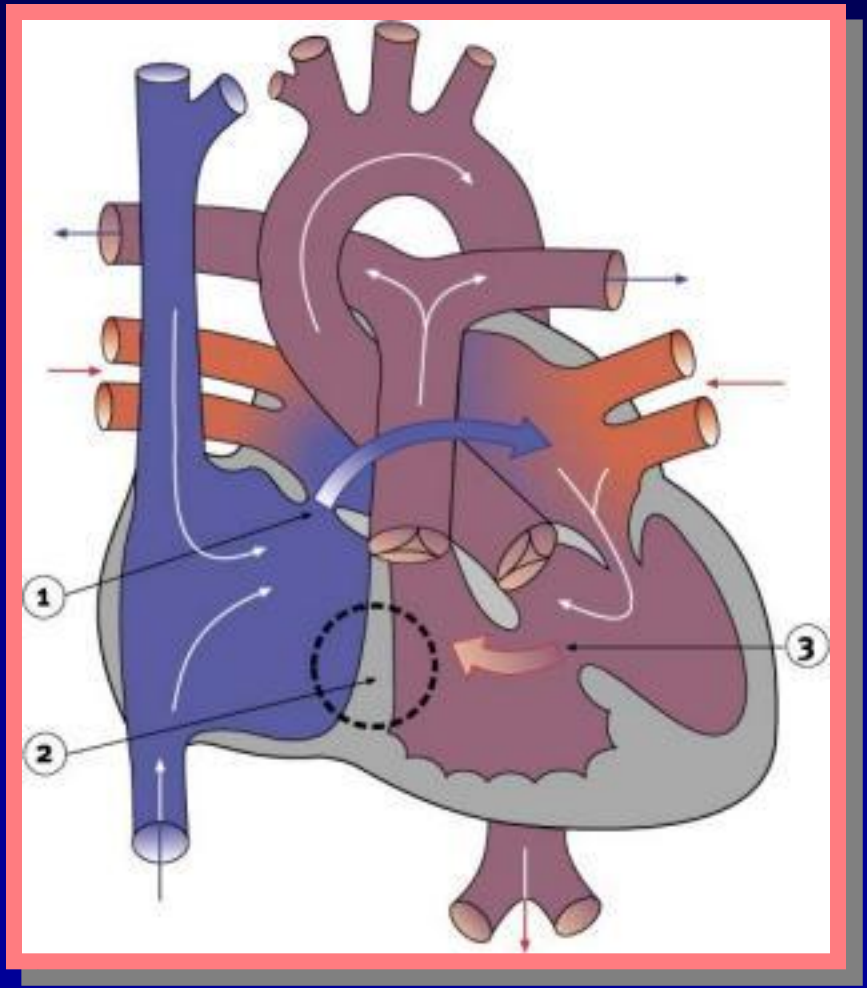


Surgical Management

- VSD closure
  - transatrial access if possible
  - Infundibular resection for visualization
  - Patch closure

Trcuspid Atresia

- Complete absence of communication between the right atrium and right ventricle
- About 3 % of congenital heart disease



Tricuspid Atresia

- There is an obligate interatrial communication
- Usually associated with VSD
- The pulmonary blood flow is dependent on the size of the VSD
- Pulmonary blood flow can be increased or decreased causing variable presenting symptoms
- If there is no VSD (also called Hypoplastic right ventricle) the pulmonary blood flow is dependent on the PDA

Tricuspid Atresia- presentation

- The presentation will depend on the amount of pulmonary blood flow
  - cyanosis
  - congestive heart failure
- CXR will also reflect the amount of pulmonary blood flow

Acyanotic-Non shunt lesions

non-shunt
Acyanotic heart lesions

Obstruction
- Aortic stenosis AS
- Supravalvar AS
- Coarctation
- Mitral Stenosis
- Pulmonary Stenosis

Regurgitation
- Aortic regurgitation
- Mitral regurgitation
- Pulmonary regurgitation

Generally cause pressure overload

Generally cause Volume overload

Coarctation of aorta

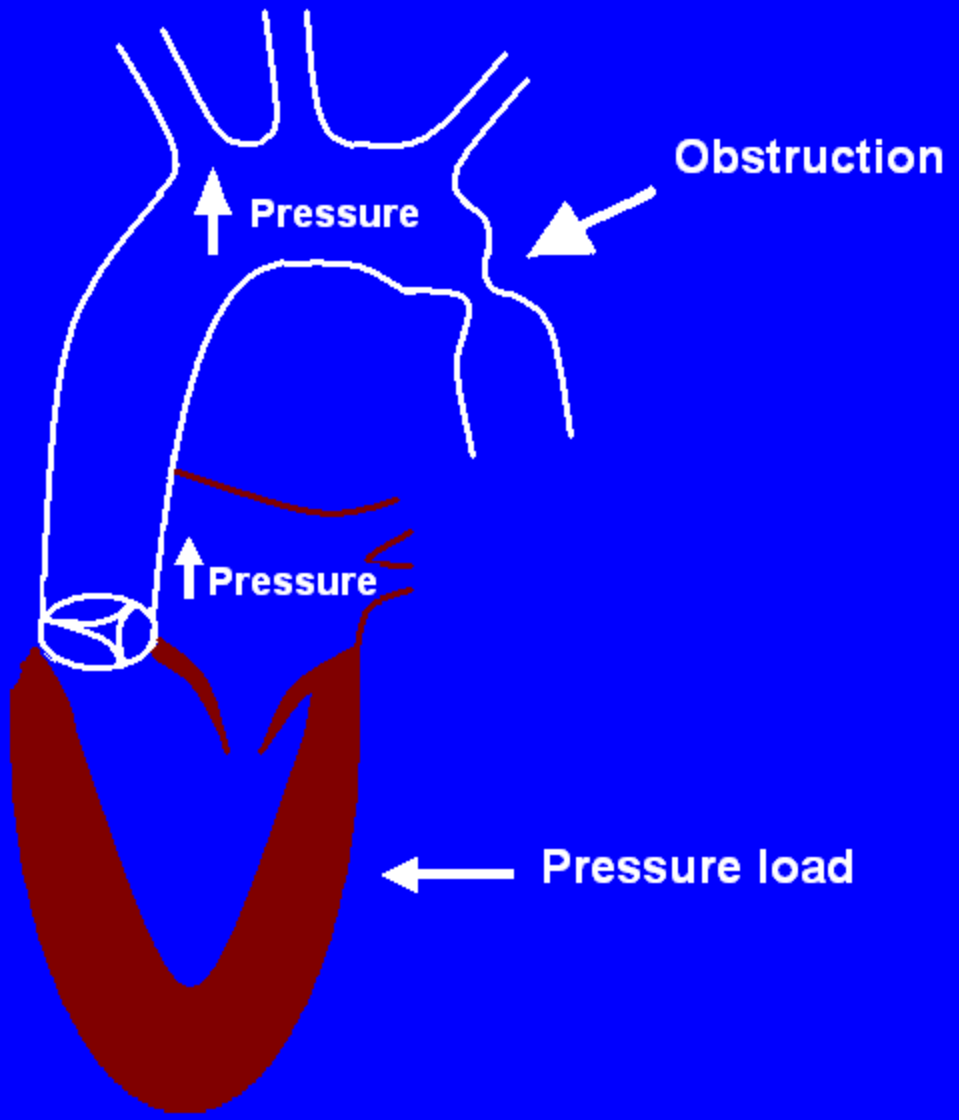
- Narrowing of the aortic lumen that causes an obstruction to blood flow.
- Variable degrees of obstruction, anatomic location, and hence clinical manifestations and outcome after treatment.

Hypertension
Pressure gradient

Left ventricular
Hypertrophy

Heart Failure
Pulmonary Edema

Pulmonary
Hypertension



CLINICAL FEATURES

- 3 typical patterns.

- Infant with congestive heart failure.
  - Catastrophic illness on day 8 - 12
  - Shock, CHF

- Child with a murmur
  - Subtle signs, pressure gradient, radiofemoral delay on examination

- Adolescent
  - Systolic hypertension, Chest pain

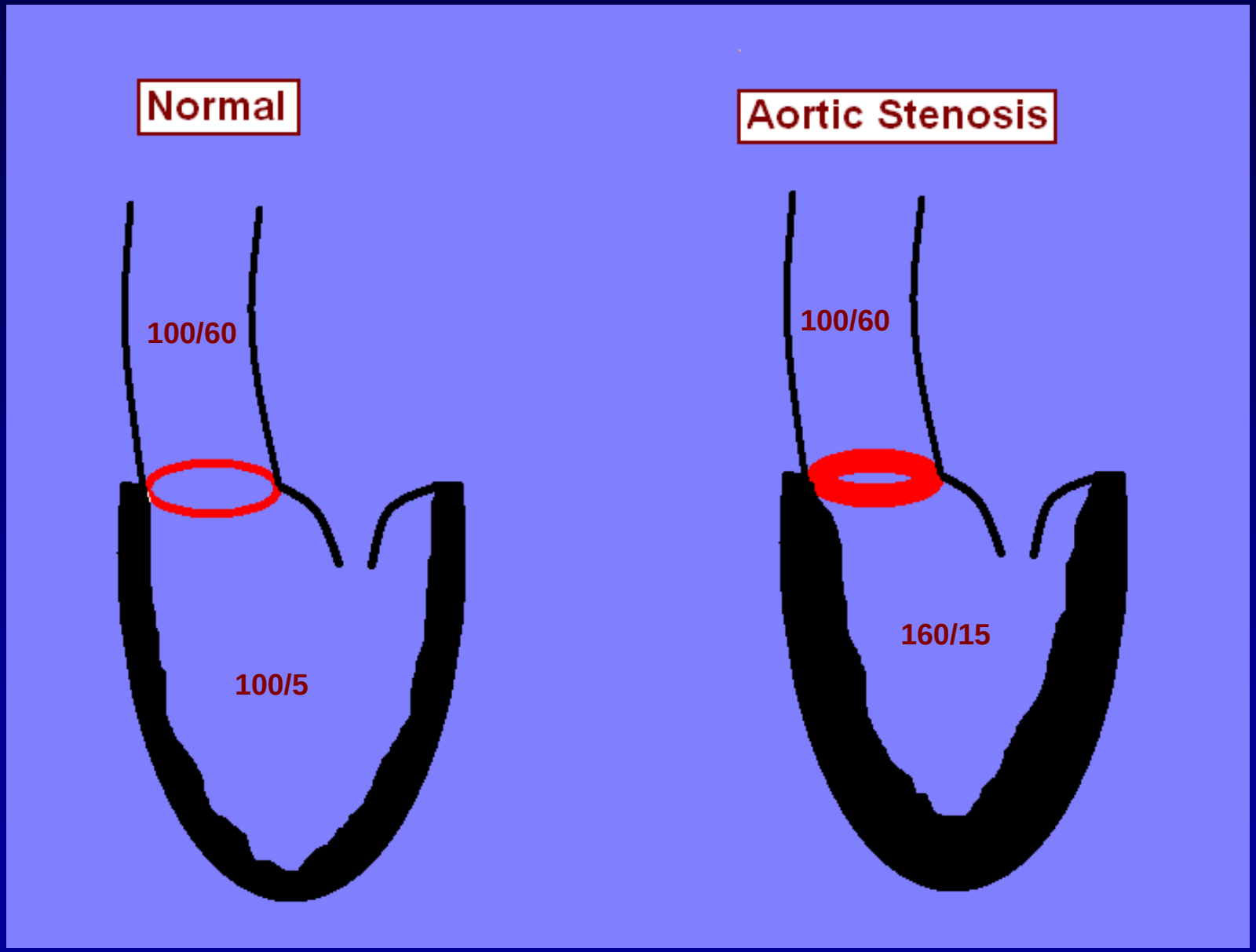
TREATMENT

- Untreated it has a a poor prognosis.
- Treatment depends on the clinical presentation:

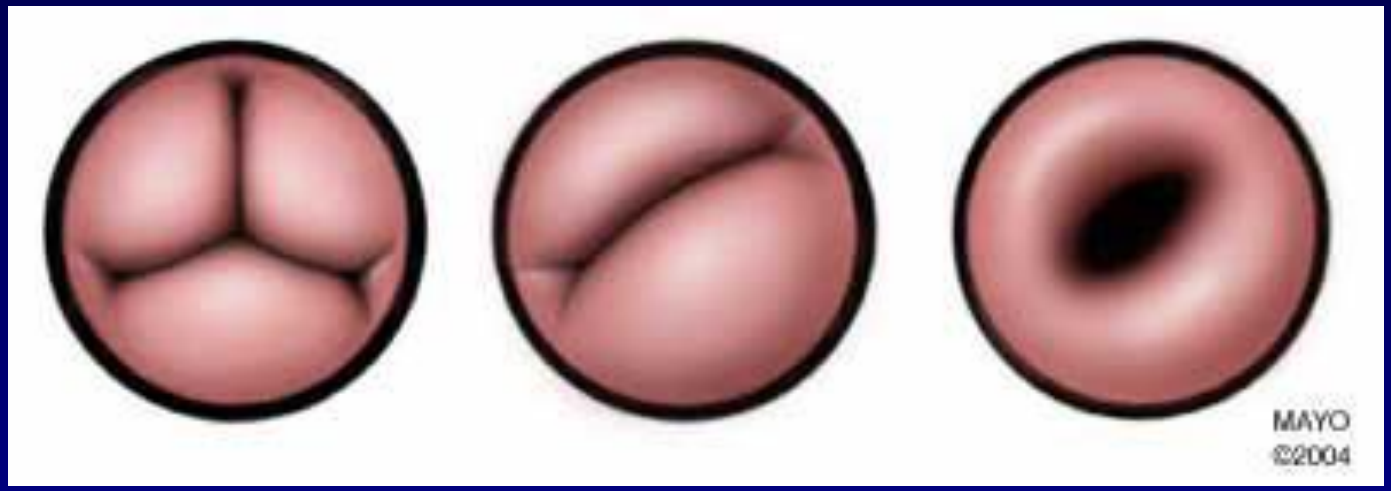
Infants presenting with shock require medical management and immediate surgical treatment.

For patients presenting during childhood timing is more elective.

Aortic stenosis



Aortic stenosis



- Clinical presentation depends on the degree of stenosis
  - Ranges from asymptomatic to Chest pain with exercise to heart failure
  - May present with sudden cardiac death

Presentation

Volume overload results in ventricular dilation and congestive heart failure

Atrial dilation may lead to atrial arrythmias

Examination typically shows holosystolic murmur radiating to the axilla, with displaced apex beat and S3

Management

Diuresis, and afterload reduction are the mainstay of treatment

Surgical repair of replacement of the valve in refractory CHF

Heart Failure in Children

Definition

- Pathophysiological state in which an abnormality of cardiac function is responsible for the failure of the heart to pump blood to meet the metabolic requirements of the body
- Chronic heart failure is a clinical syndrome in which heart disease reduces cardiac output, increases venous pressures, and is accompanied by molecular abnormalities that cause progressive deterioration of the failing heart and premature death of myocardial cells

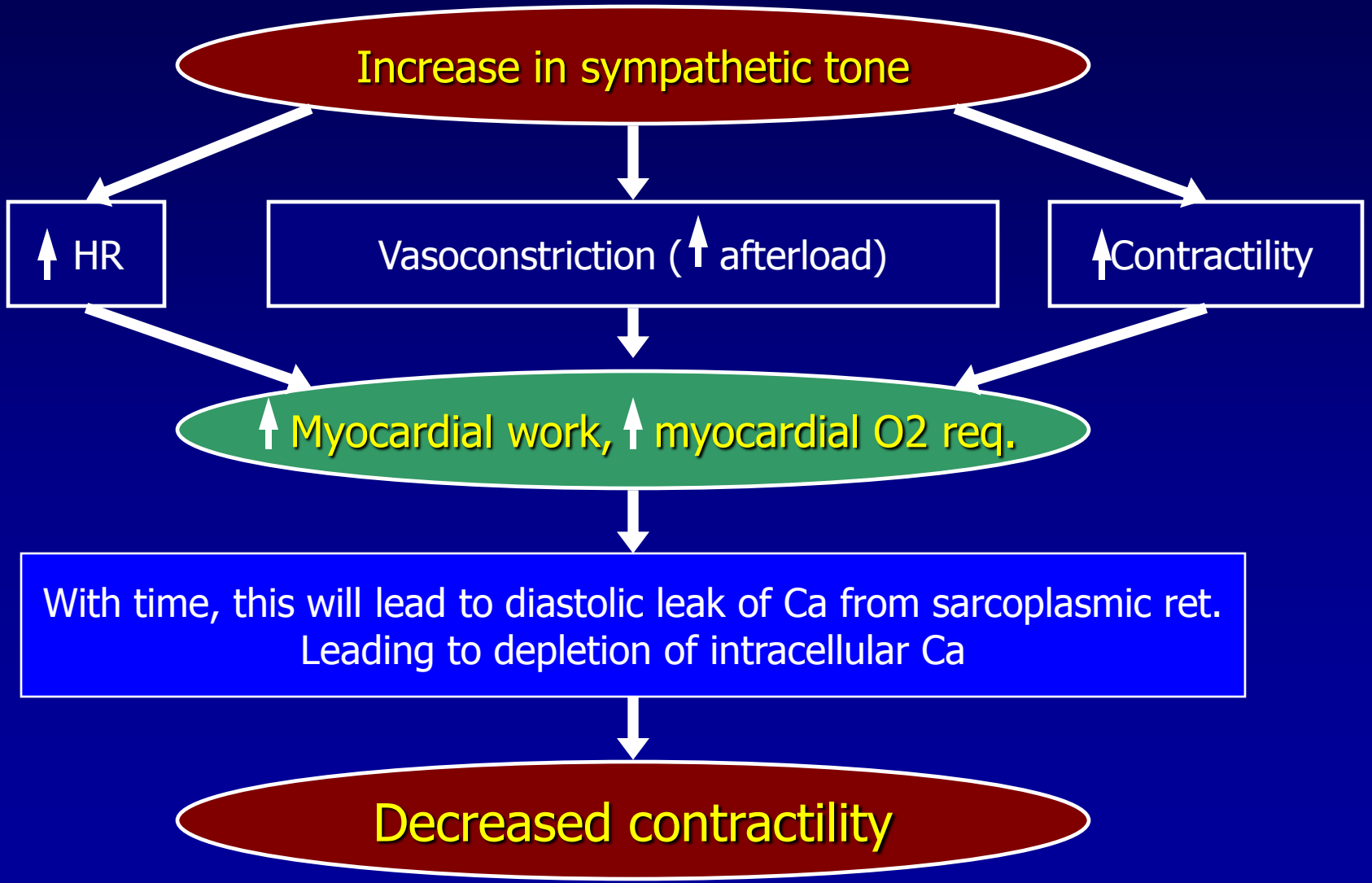
How may the heart fail?

Too much preload	Too much afterload	Impaired contractility	Impaired filling
Streching of muscle fibers • Lt-Rt shunts • Complete heart block	**Increased wall stress** • Hypertension • Coarctation • Aortic stenosis	• Dilated cardiomyo-pathy • Myocarditis	• Hypertrophic cardiomyo-pathy • Constrictive pericarditis

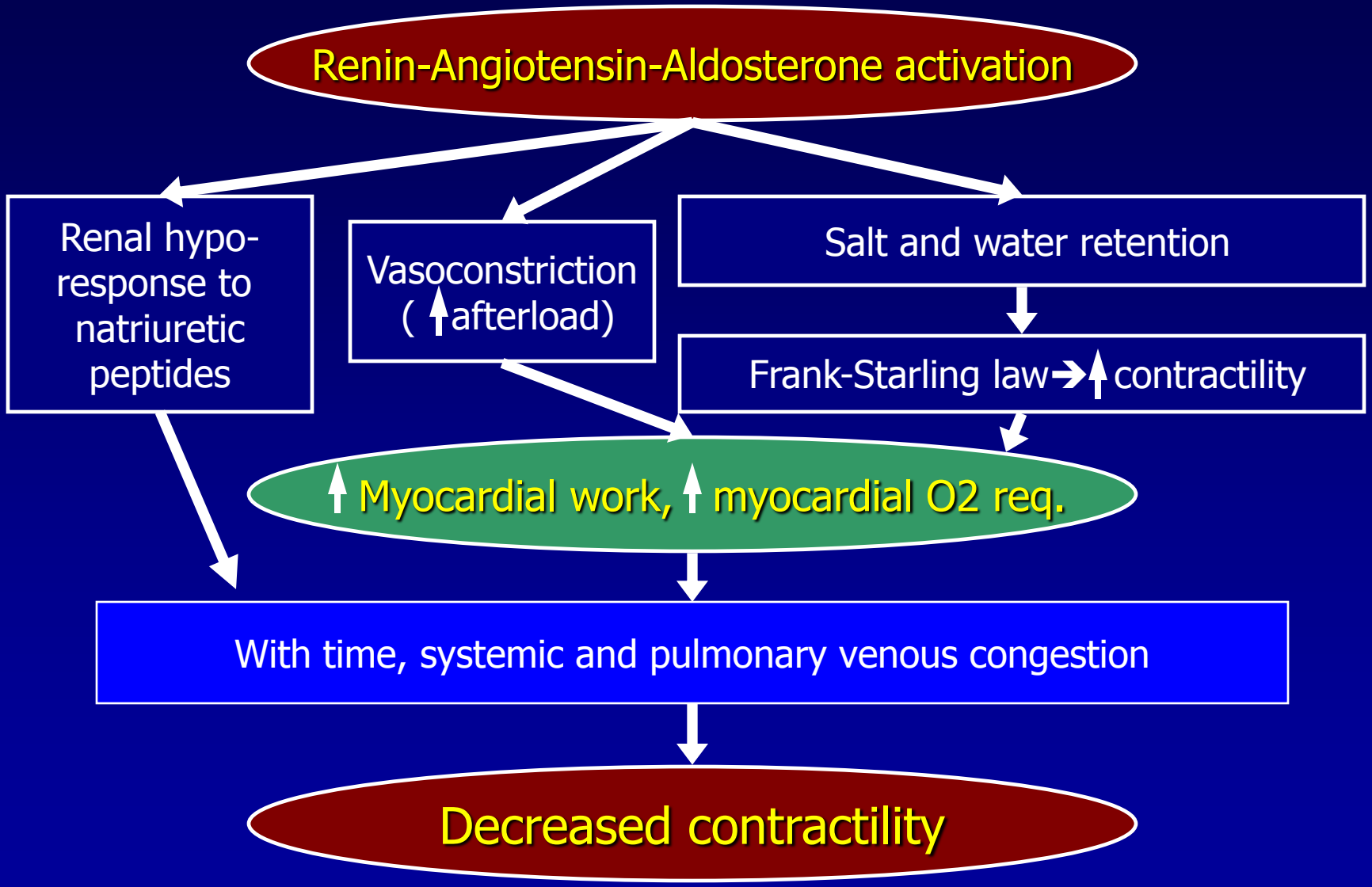
What causes heart failure in children?

- Congenital heart disease:
  - Volume overload
  - Pressure overload
  - Post-op. patients with residual lesions
  - Pulmonary vascular disease
  - Chronic hypoxia
- Coronary artery disease
  - Congenital anomalies
  - Acquired disease
- Endocarditis
- Cardiomyopathies
  - Dilated
  - Hypertrophic
  - Restrictive
- Chronic anemia
- Primary pulmonary hypertension
- Rheumatic heart disease

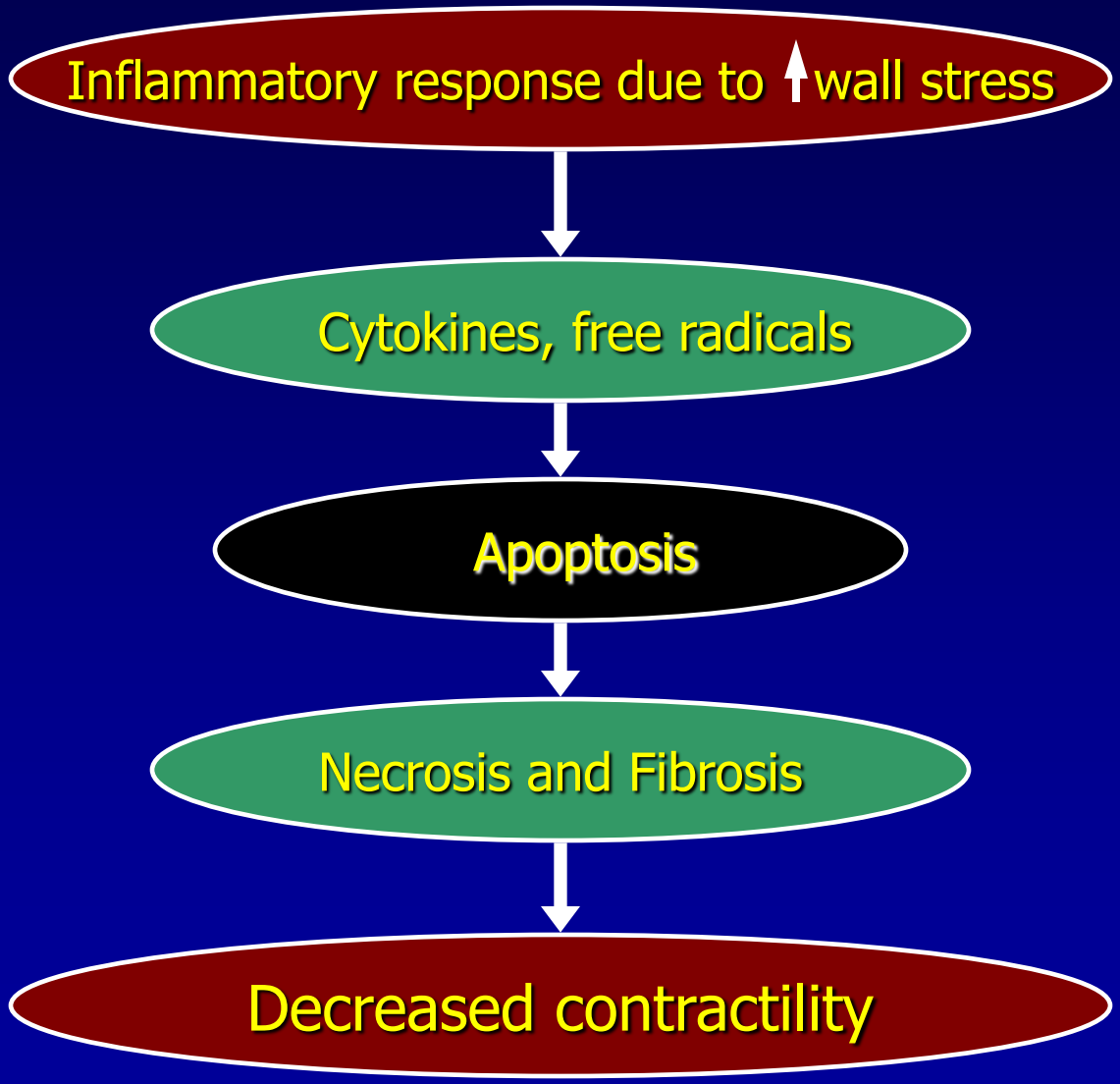
What happens when the heart fails? (Compensatory mechanisms)



(More Compensatory mechanisms)



(More Compensatory mechanisms)



Clinical Features

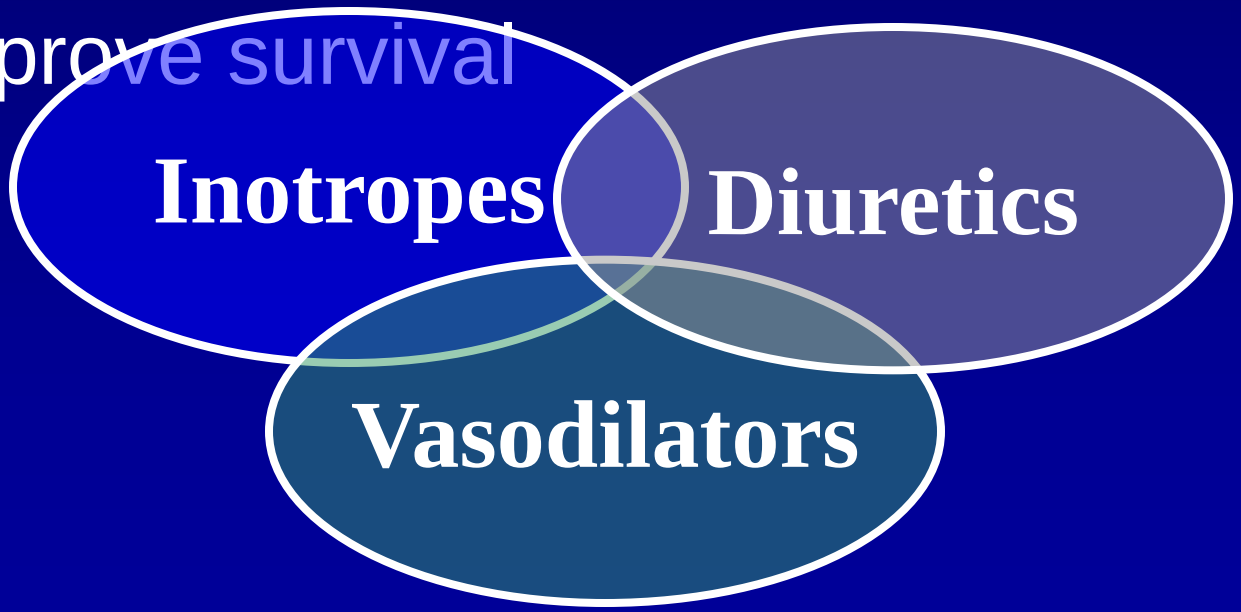
- Reflection of:
  - Compensatory mechanisms
  - Manifestation of decreased tissue perfusion
  - Manifestation of pulmonary and systemic venous congestion

Clinical features

- Tachycardia, cardiomegally,
- Cold extremities, weak pulses, hypotension, skin mottling
- Metabolic acidosis
- Dyspnia, tachypnia, retractions, grunting
- Poor feeding, poor growth
- Central cyanosis (pulmonary edema)
- Peripheral cyanosis (decreased perfusion)
- Hepatomegally, peripheral edema
- Exercise intelerance in older children

Management goals

- Improve symptoms
- Slow progression of ventricular remodeling
- Improve survival



Diuresis

- Decreases volume overload
- *BUT* stimulates Renin-Angiotensin-Aldosterone axis, and sympathetic nervous system

Inotropic support

- Digoxin Inhibits sarcolemmal Na-K ATPase pump ➔ Increase intracellular Na
  - ➔ Inhibition of Na/Ca exchange
  - ➔ Increase intracellular Ca
  - ➔ Increase contractility
- Decrease sympathetic tone and norepinephrine

Therefore; Improves symptoms and decreases hospital stay

In the acute setting, inotropic support is usually accoplished by intravenous medications: **Dopamine, Dobutamine, Epinephrine, Milrinone**

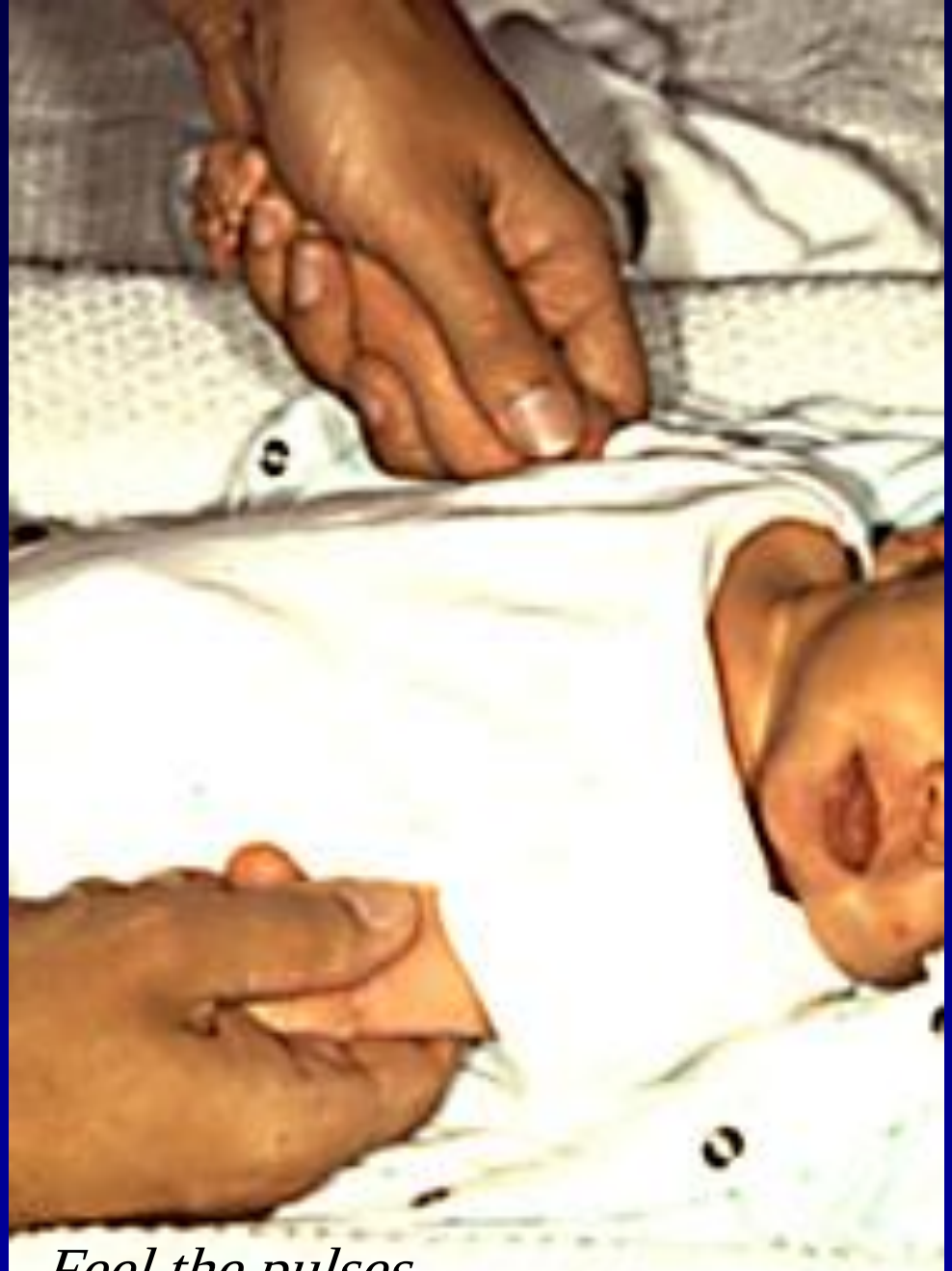
Improving cardiac output will promote more effective diuresis

Vasodilation

- Most common approach is ACE inhibition
- Favorable ventricular remodeling
- Improvement of renal function
- Blunts hypertrophic and apoptotic effect of angiotensin II

Therefore; improves cardiac output, and decreases wedge pressure (left atrial pressure), and improves symptoms

Angiotensin receptor antagonists (e.g Losartan) has similar effect

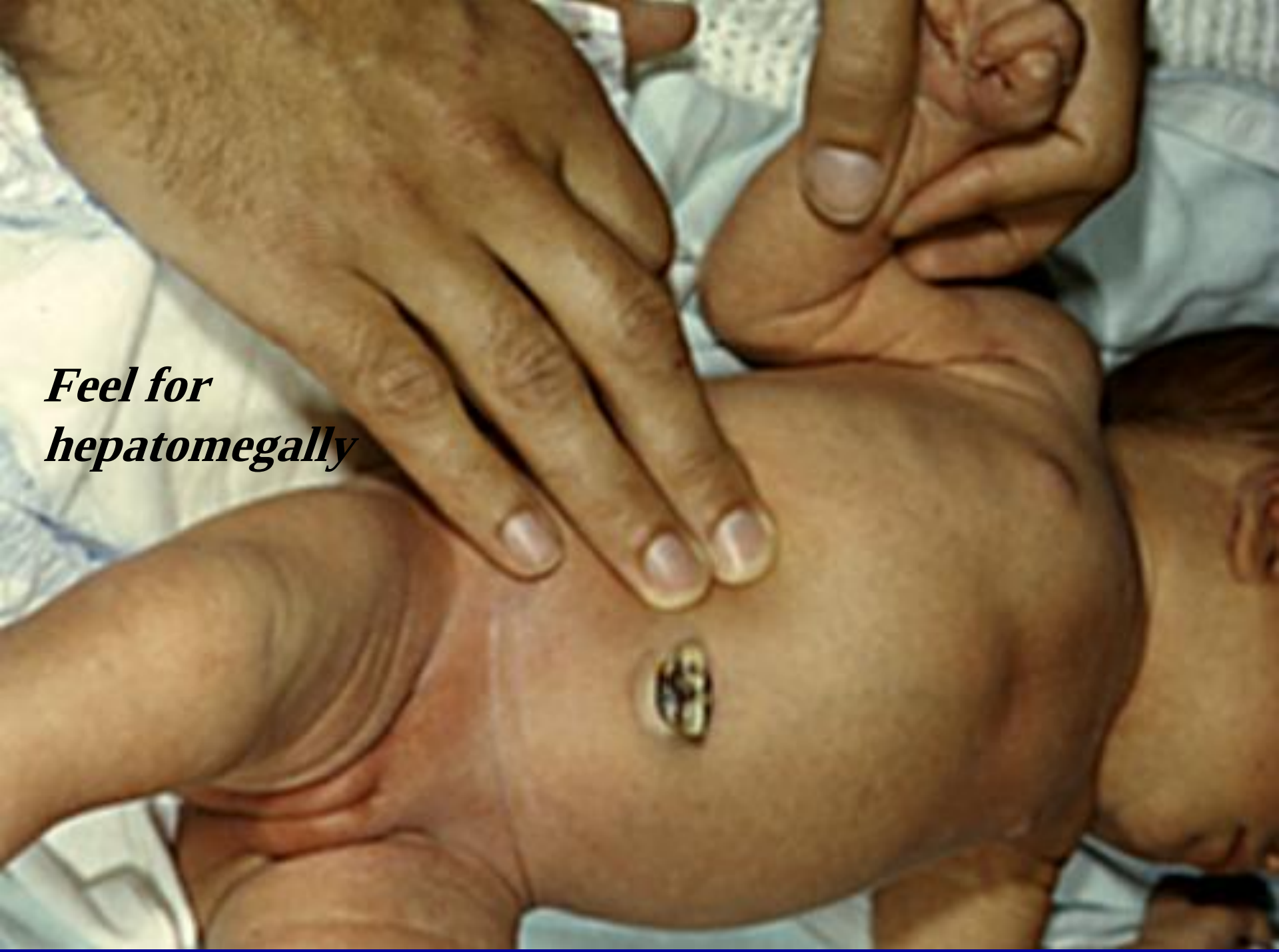


Feel the pulses
especially brachial and femoral

Look at the respiratory pattern and for evidence of rec

Feel the precordium for hyperactivity and for thrills


Locate the apex beat

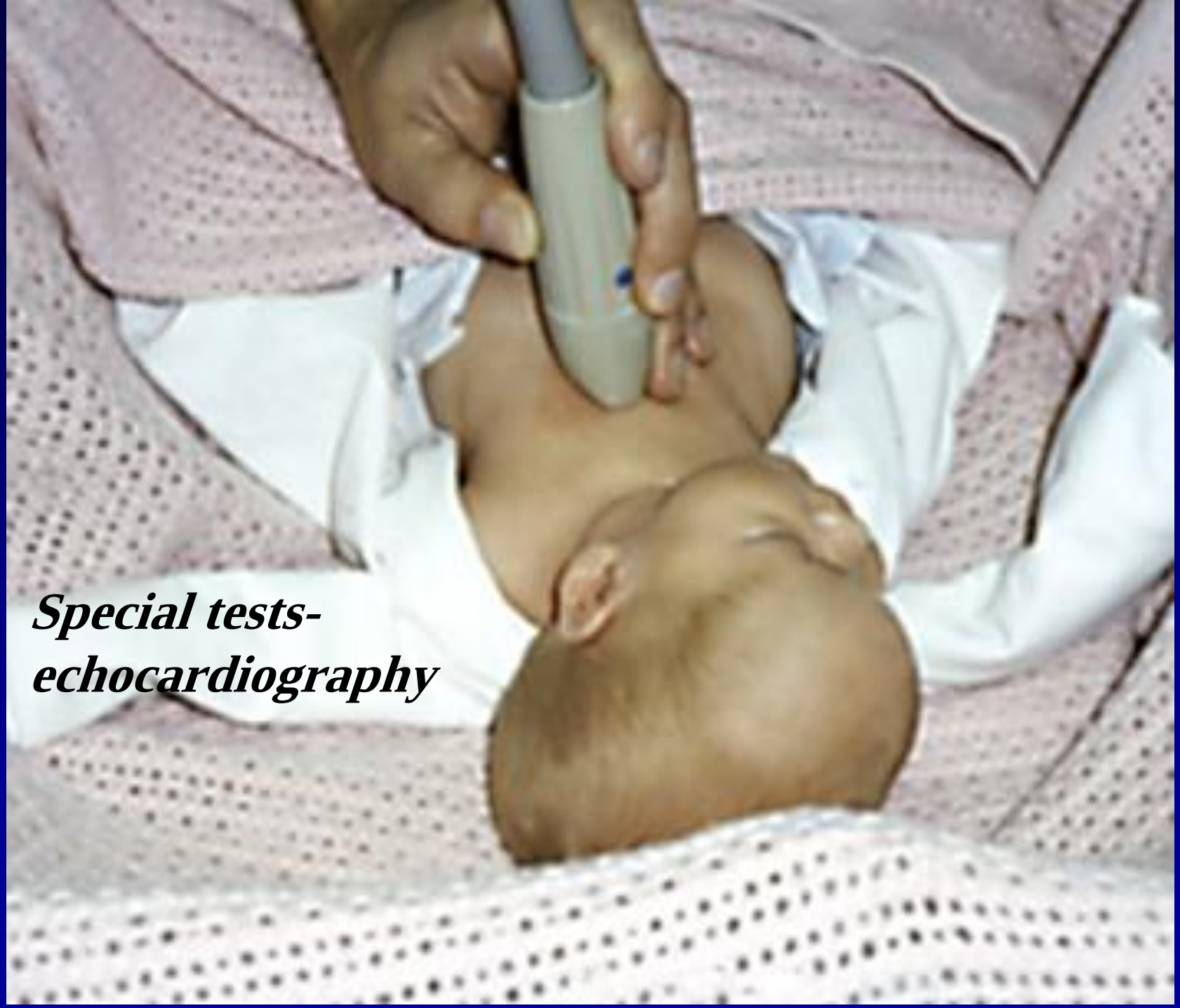

Feel for hepatomegally


Do what is necessary to calm the baby down!

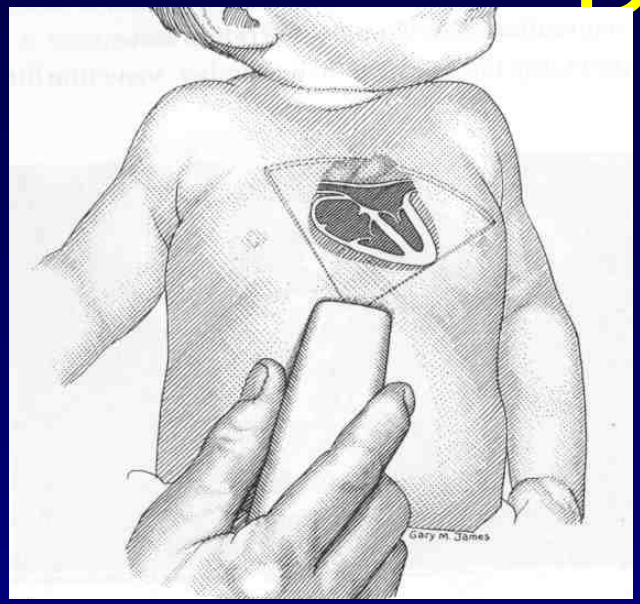

Do what is necessary to calm the baby down!

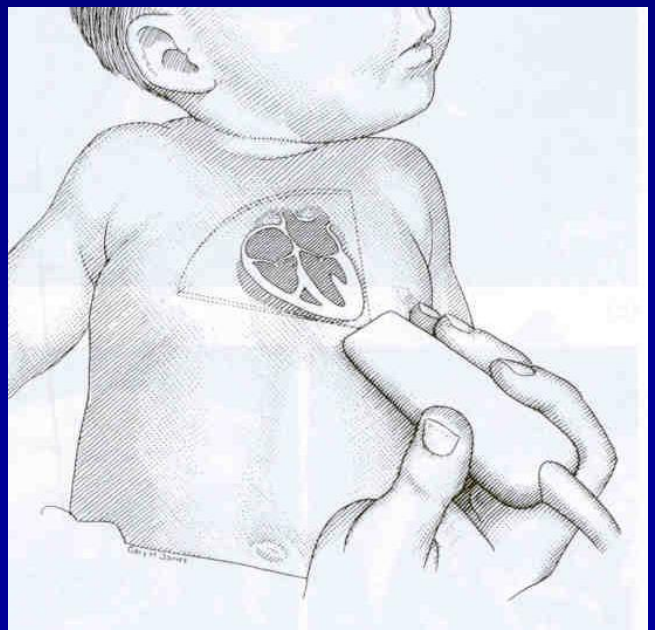
Listen at the back for radiation of murmurs


3 months old- severe failure to thrive

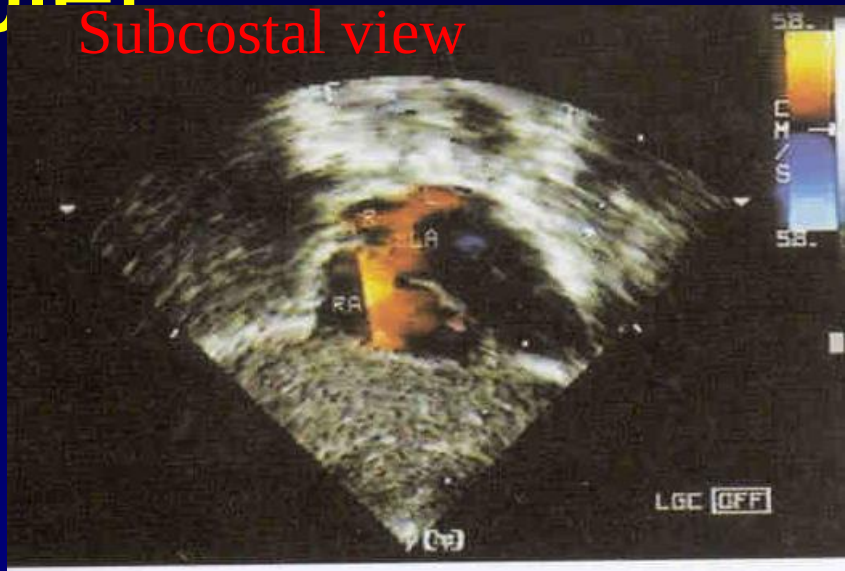

Special tests-
echocardiography

Pulmonary veins - colour Doppler

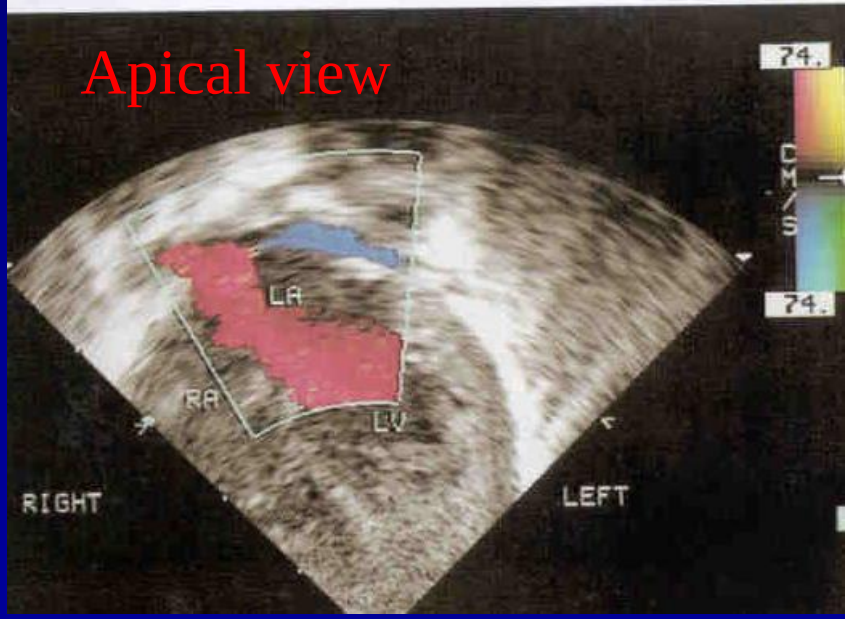




Subcostal view



Apical view



Thank you