Introduction to CHD
VSD and TOF as examples

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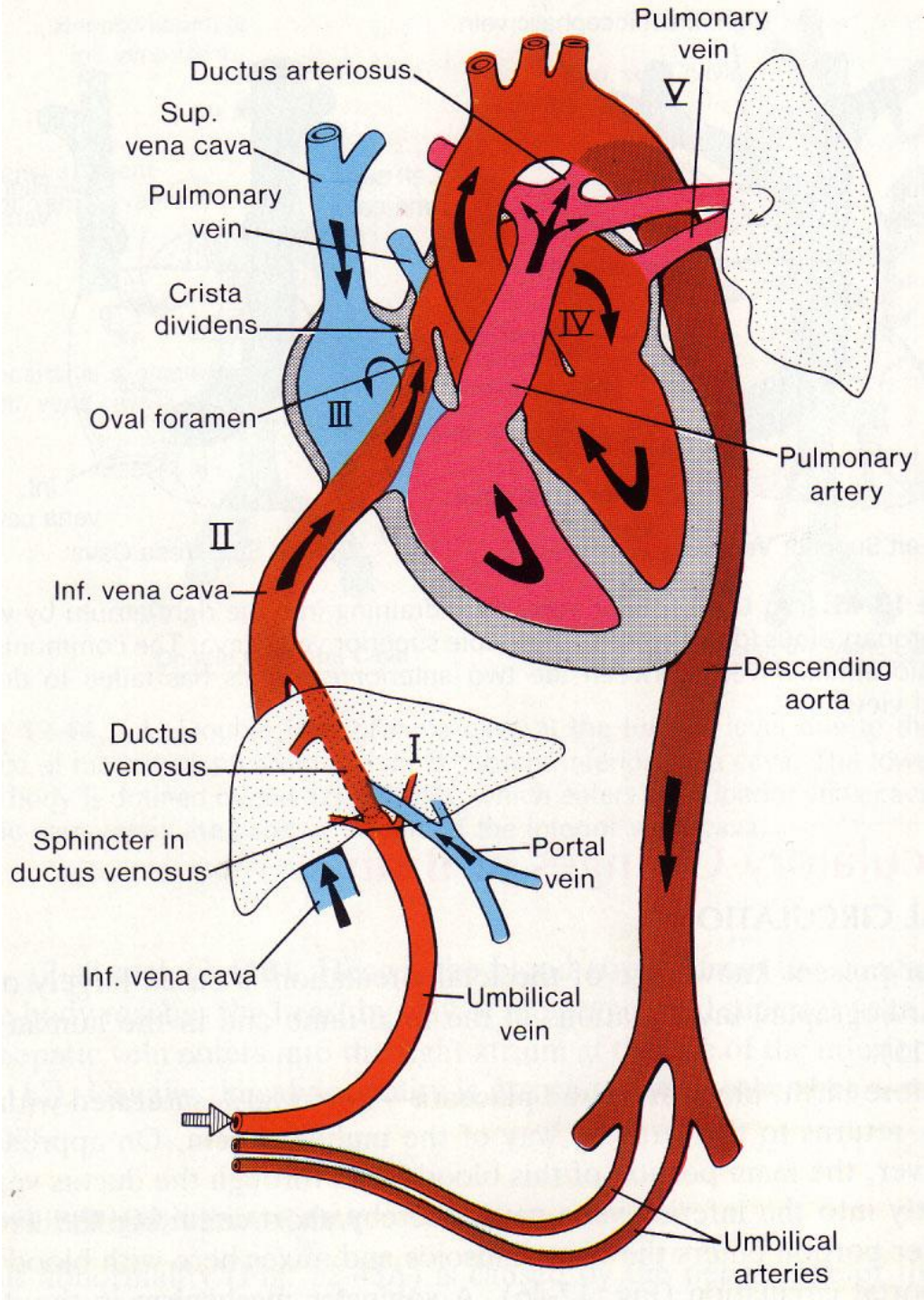
2022

Outline

- The Fetal circulation and postnatal changes
- Possibilities and classification of congenital heart disease
- Presentations of CHD
- Mechanisms of Heart Failure in CHD
- Mechanisms of Cyanosis in CHD
- VSD as an example of acyanotic heart diseae
- TOF as an example of cyanotic heart disease

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

- 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
- Pulmonary vascular resistance is high due to constricted pulmonary vessels



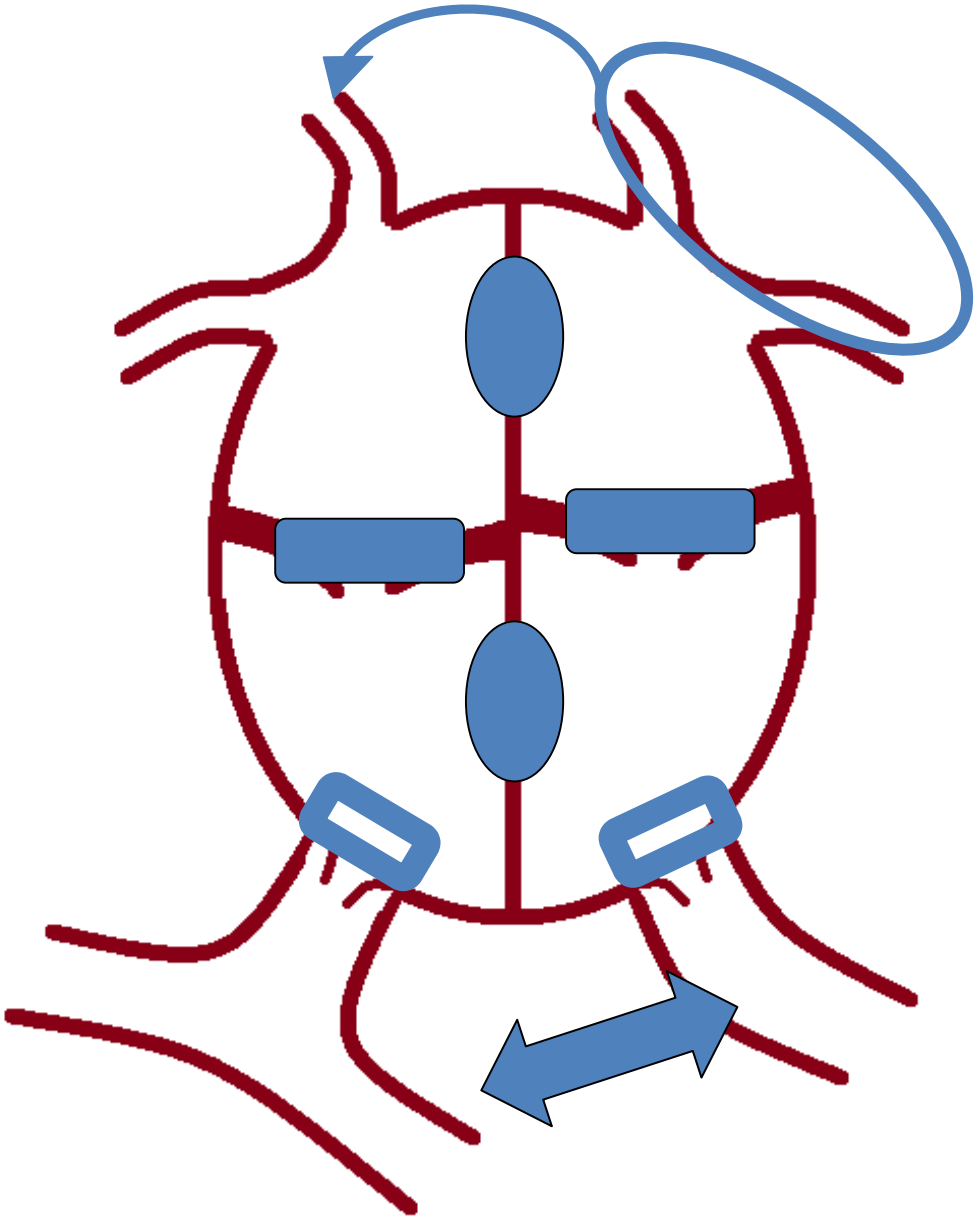
PFO and PDA during fetal life

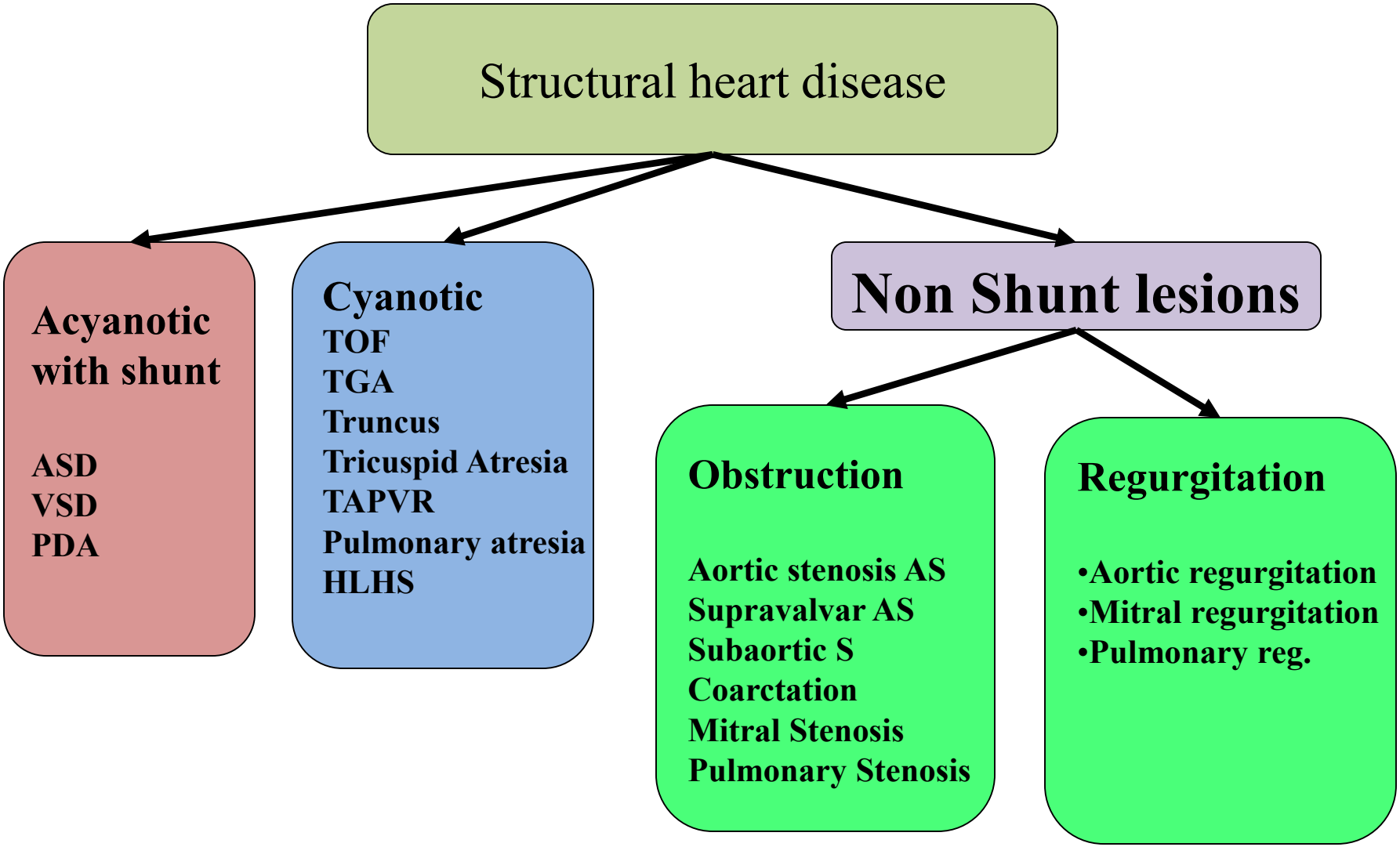
- PFO: directs blood coming from inferior vena cava (from umbilical vein) to the left atrium
- PDA: directs blood from the pulmonary artery to the descending aorta because the lungs are not being used for oxygenation in the fetus
- Both PFO and PDA are important for fetal survival

PFO and PDA after birth

- PFO: when blood flow to the left atrium increased because of more blood flow to the lung after the baby is born, the flap of foramen ovale closes
- PDA: Increased oxygenation and absence of placental prostaglandins once the cord is clamped causes constriction and closure of the PDA within few days after birth

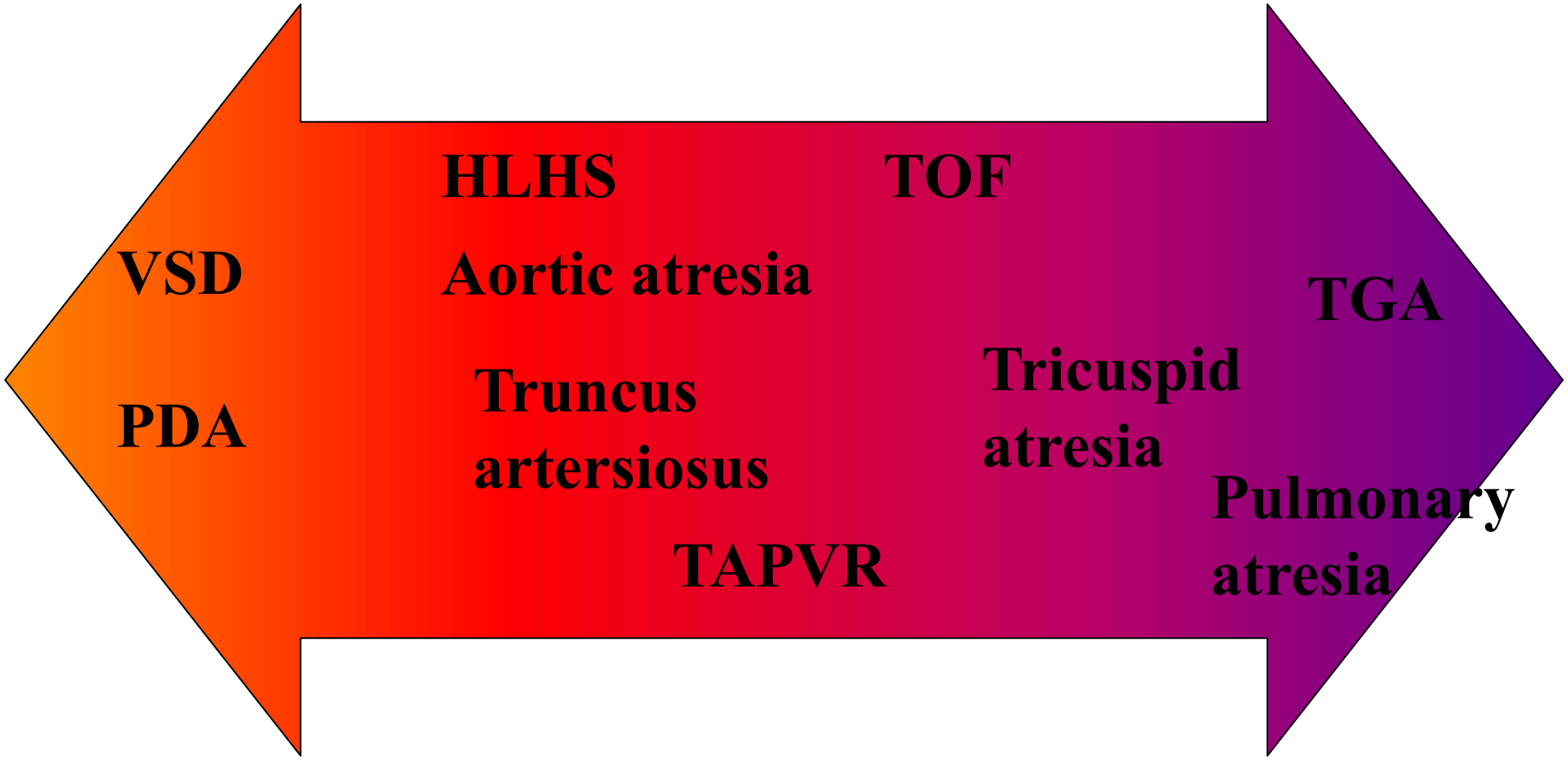
Congenital heart disease – Many possibilities




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

Symptoms of congenital heart disease

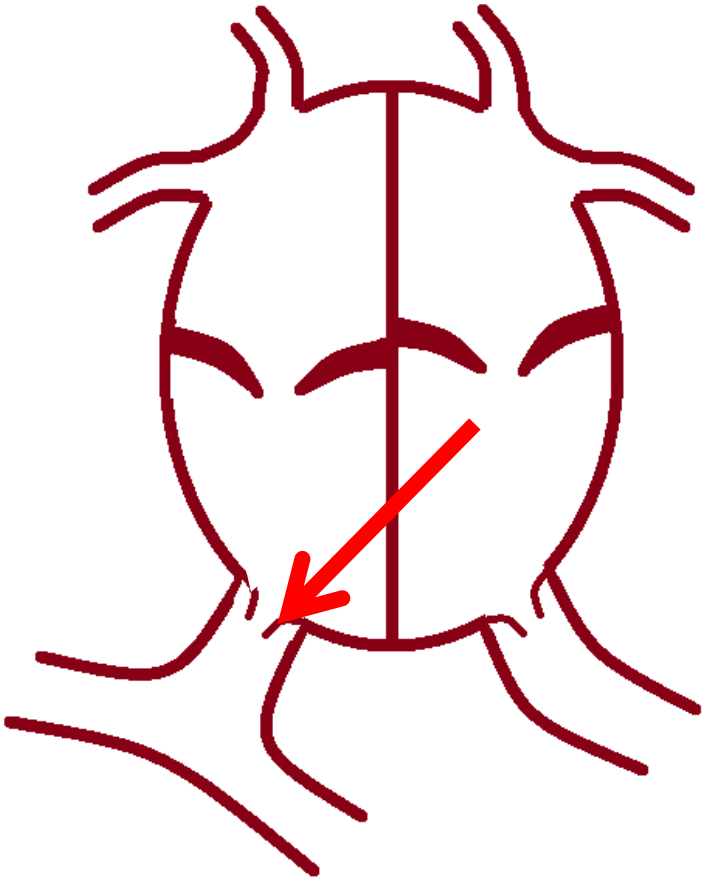
Lesions with shunts



Many asymptomatic patients

Mechanisms of heart failure in CHD

- Pulmonary overcirculation with high pressure
  - VSD
  - PDA
  - AVCanal
  - Truncus arteriosus
  - Single ventricle without pulmonary stenosis



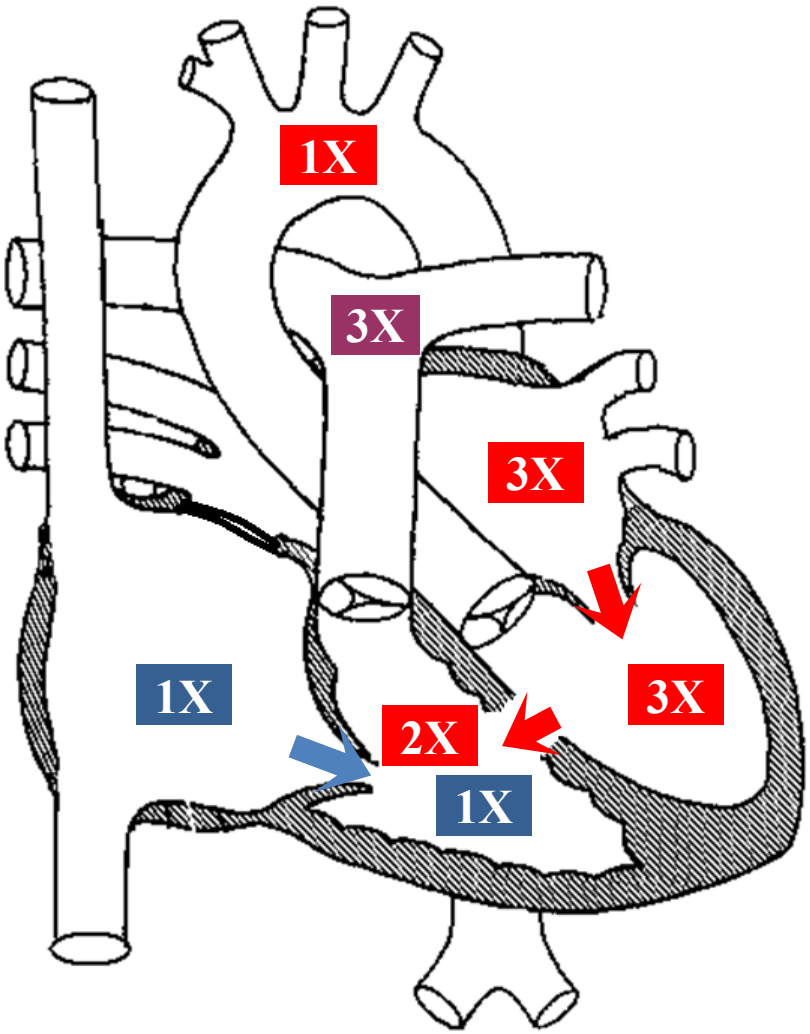
Mechanisms of heart failure in CHD

- Critical obstruction to systemic blood flow causing metabolic acidosis and shock
- Presents when PDA starts to constrict
  - Critical aortic stenosis
  - Critical coarctation of aorta
  - Hypoplastic left heart syndrome

Mechanisms of heart failure in CHD

- Left ventricular dysfunction, takes long time to happen
- Due to:
  - Long standing pressure overload (eg, AS, COA)
  - Long standing volume overload (eg, VSD, mitral or aortic regurgitation
  - Rarely due to myocardial ischemia (severe cyanosis, severe diastolic hypotension, relative ischemia due to severe hypertrophy, coronary anomalies)

VSD physiology



QS=1X

QP=3X

L-R shunt= 2X

Qp:Qs= 3:1

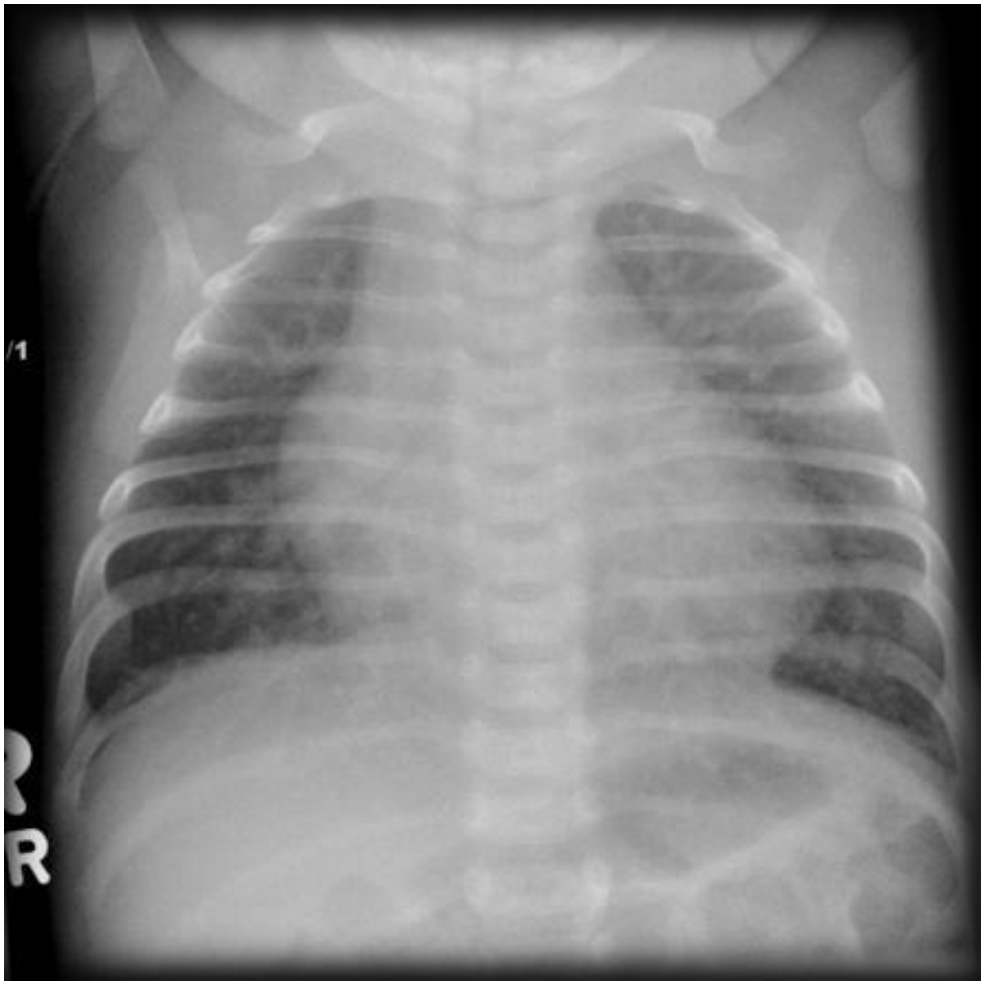
- **VSD causes Pressure load on the right ventricle causing RVH, and Volume load on the left atrium and ventricle leading to dilation**
- **Shunt occurs during Systole (ventricular emptying)**
- **Cardiac output is well maintained even in large VSD's**

Presentation of VSD

- Depends on the size of the defect
- Large defects cause significant increase in pulmonary blood flow resulting in respiratory symptoms
  - Progressive Tachypnia and dyspnea in infancy
  - Interrupted feeding with resultant failure to gain weight
  - Excessive diaphoresis, cold extremities
  - Recurrent respiratory infections
  - If untreated, patients may develop Eisinminger syndrome with right to left shunting due to pulmonary vascular hypertension, usually takes many years to develop
- Small defects are usually asymptomatic

Signs in large VSDs

- Physical examination:
  - Signs of respiratory distress, grunting
  - Tachycardia
  - Displacement of apical impulse
  - Hepatomegally
  - Crepitation in cases of pulmonary edema
  - Cold extremities, signs of decreased perfusion in severe cases
- ECG: Left ventricular (hypertrophy), left atrial dilatation
- CXR: Cardiomegally, increased pulmonary vascularity



Treatment

Nutritional support

Increase caloric intake (fortified formulas, more frequent feeds, NG feeding if needed)

Medications

Diuretic (furosemide): decrease the congestion, improves respiratory distress

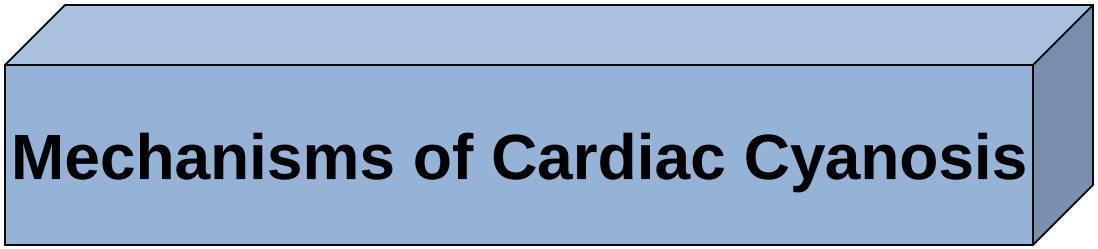
Afterload reduction (captopril): decrease the amount of left to right shunting

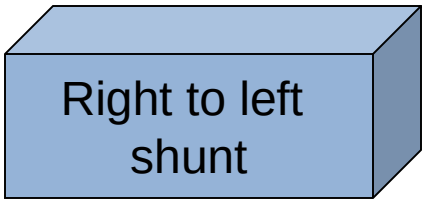
Inotrope (digoxin): rarely needed, helps if systolic function is depressed

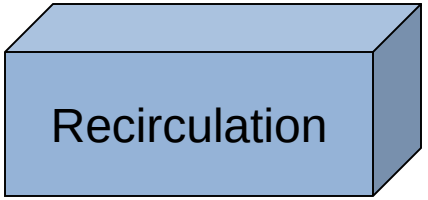
Surgery

Surgical paliation (Pulmonary artery constriction by a band for temporary relief of symptoms if correction cannot be done)

Surgical repair (VSD closure) is the definitive therapy







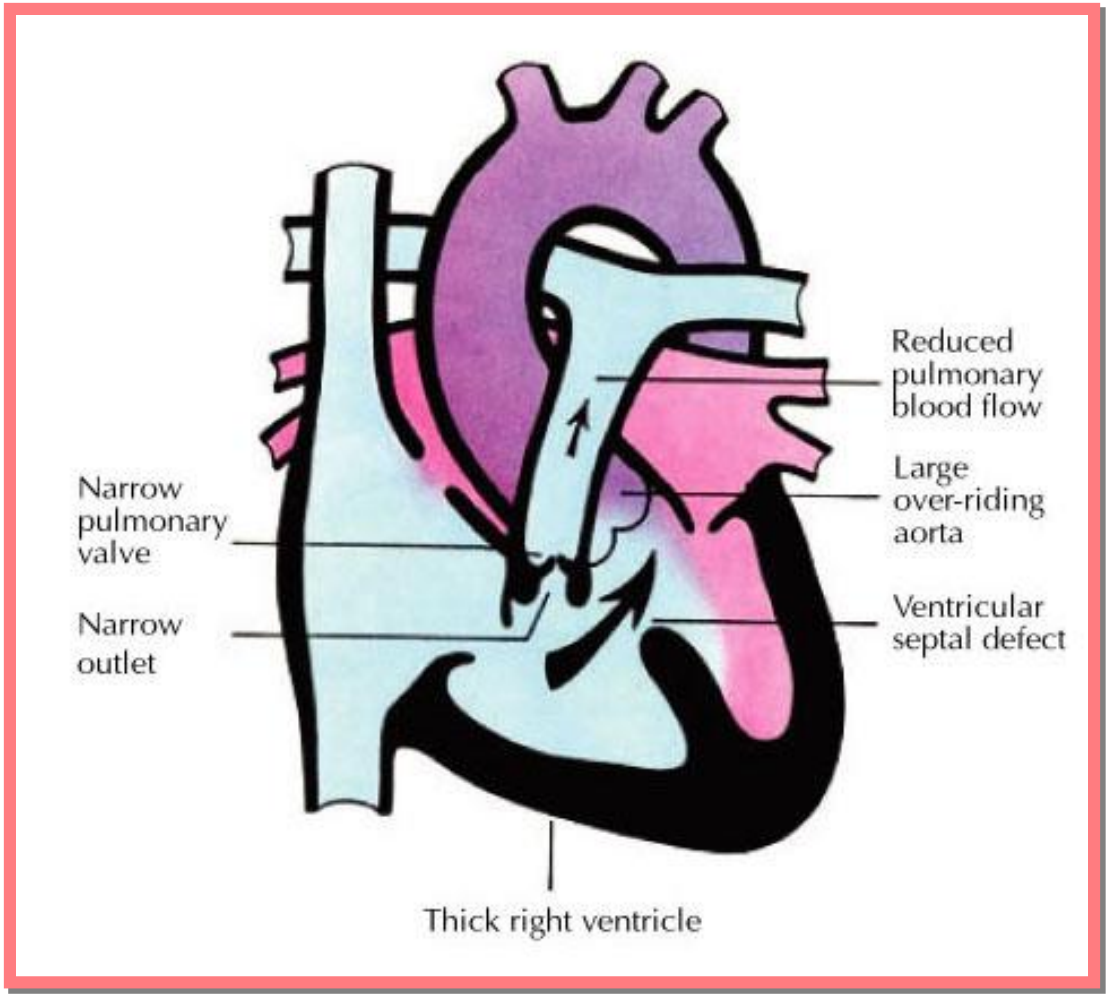
TOF

Tricuspid atresia
Truncus arteriosus
Single ventricle

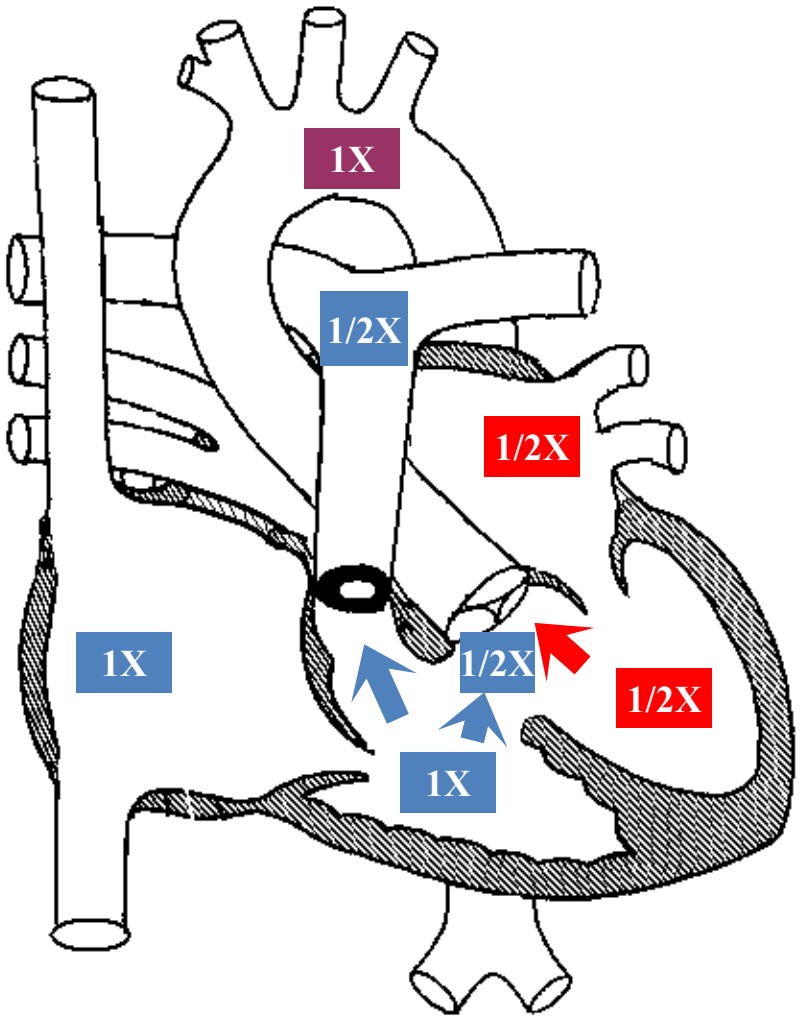
TGA

Tetralogy of Fallot (TOF)

1. RVOT obstruction
2. VSD
3. Overriding aorta
4. RV hypertrophy



Tetralogy of Fallot physiology



QS=1X (cardiac index)

QP=1/2X

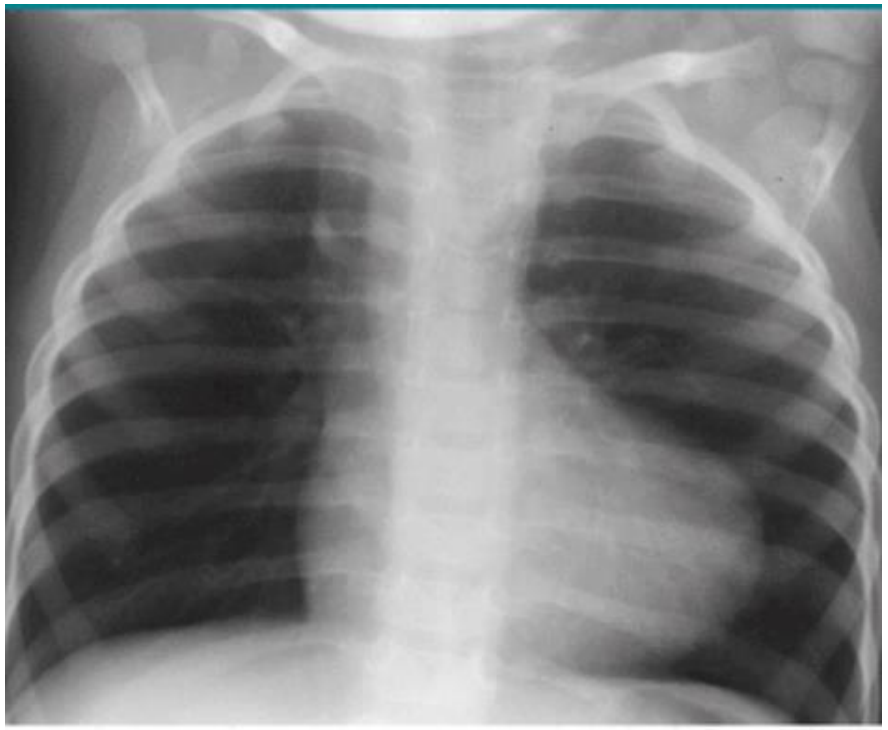
R-L shunt= 1/2X

Qp:Qs= 0.5:1

- The degree of right to left shunt depends on the degree of pulmonary stenosis
- No chamber is receiving any volume overload➔ no cardiomegaly
- Lungs receive less than normal amount of blood ➔ no congestion, no respiratory distress.

Presentation in TOF

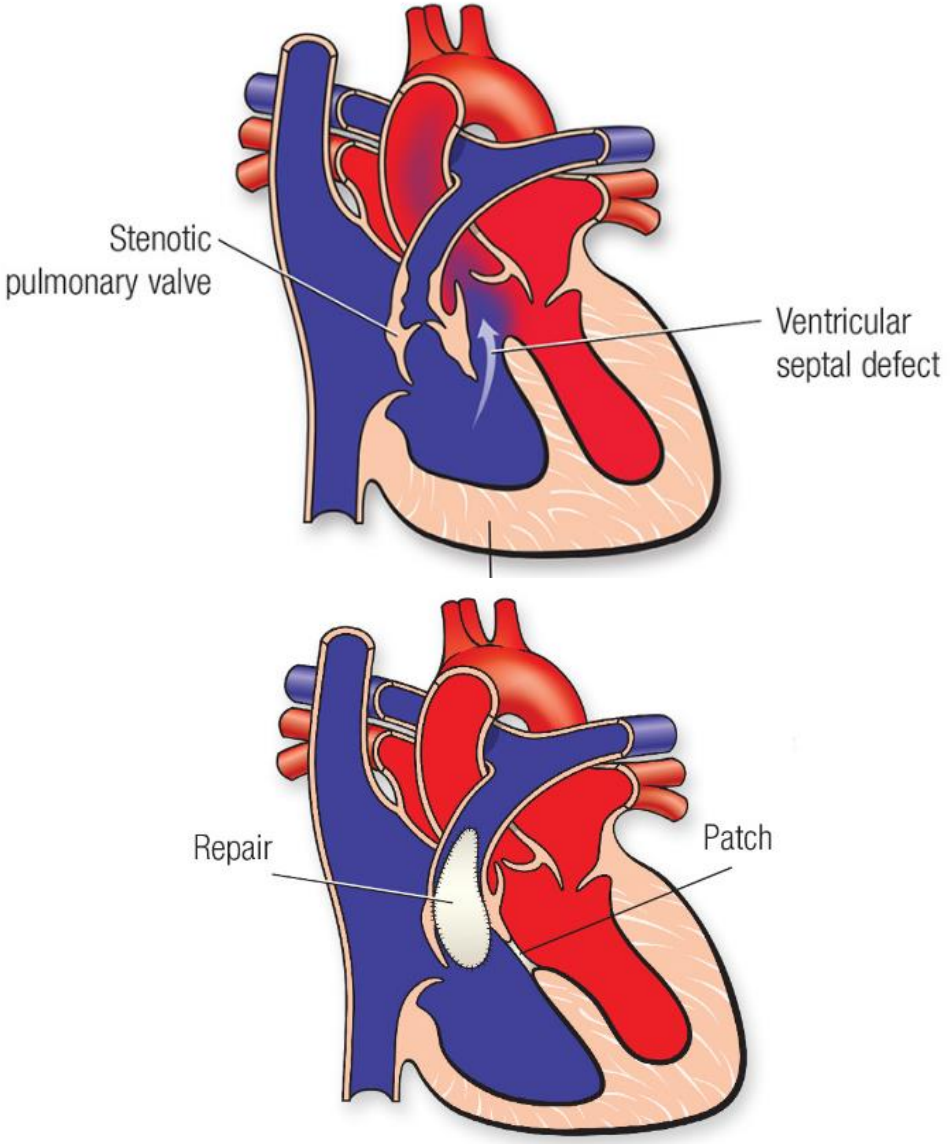
- Cyanosis (variable degree, depending on the degree of PS)
- Hypercyanotic spells (episodes of life-threatening deep cyanosis)
- Usually well thriving infants
- No distress
- Clubbing (in childhood)
- Prominent RV heave
- Loud systolic murmur at the left sternal border
- CXR: boot shape heart, oligemic lungs (no cardiomegaly)
- ECG: right axis deviation and right ventricular hypertrophy



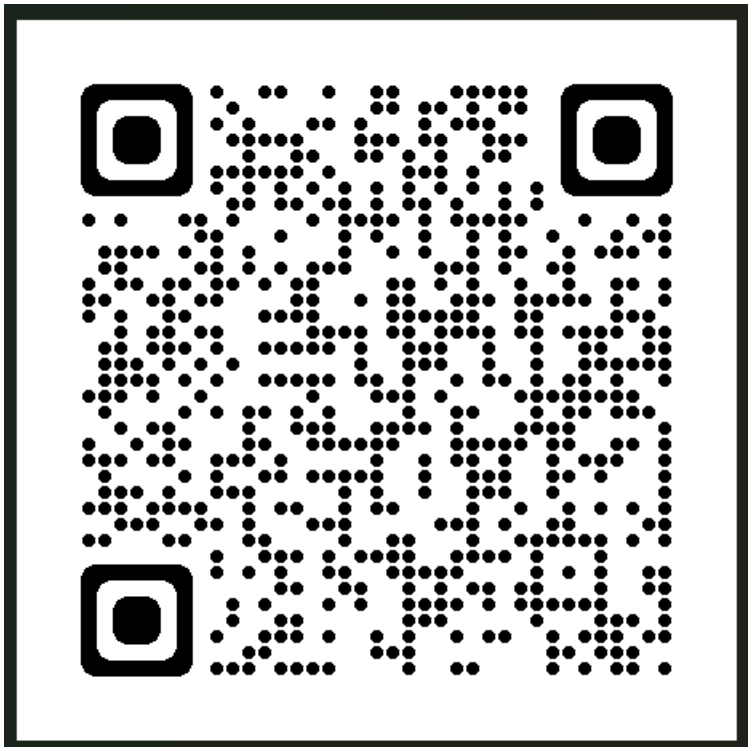
Management

Hypercyanotic spells: Oxygen, knee chest position, calm baby down, IV fluids, systemic vasoconstrictors (phenylephrine), Morphine.

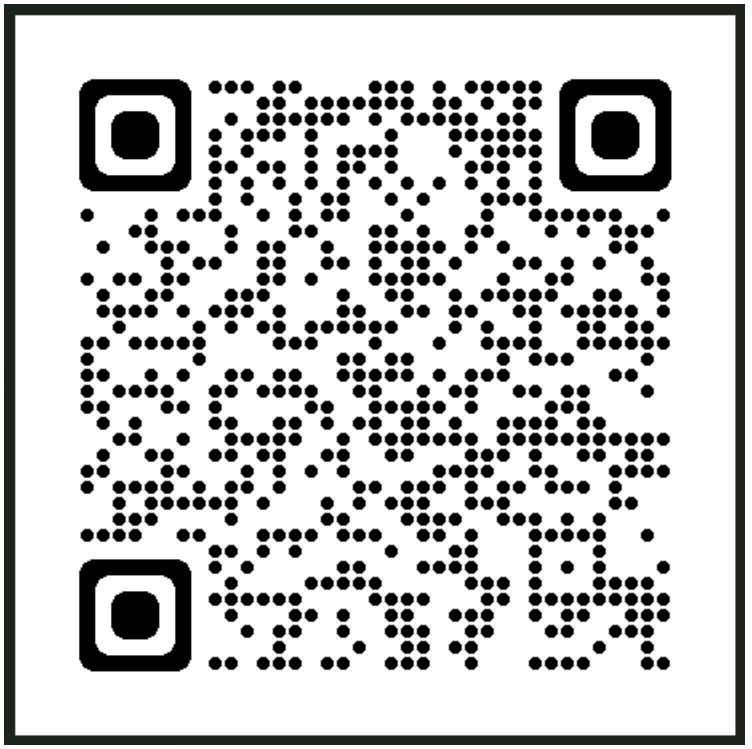
Surgical repair



Extra-readings



VSD review article



TOF review article

The end