Congenital Heart Disease

- Abnormalities of the heart/ great vessels since birth
- Incidence higher in premature infants
- Faulty embryogenesis during 3-8 weeks of IU life
- Cause – unknown

  - genetic or environmental

  - rubella infection, drugs, heavy drinking during pregnancy

Classification

1. Malpositions of Heart
   Dextrocardia
   may be accompanied by situs inversus
2. Shunts (Cyanotic CHD)
3. Obstructions (Obstructive CHD)

Shunts

A. *Left to Right shunts (Acyanotic or Late Cyanotic group) cyanosis months or years after birth*

1. Ventricular septal defect (VSD) -25-30%
2. Atrial septal defect (ASD)- 10-15%
3. Patent ductus arteriosus (PDA)- 10-20%

B. *Right to Left shunts (Cyanotic group)*

1. Tetralogy of Fallot (TOF)- 6-15%
2. Transposition of great arteries -4-10%
3. Persistent truncus arteriosus – 2%
4. Tricuspid atresia and stenosis 1%

Obstructions

1. Coarctation of Aorta 5-7%
2. Aortic stenosis and atresia 4-6%
3. Pulmonary stenosis and atresia 5- 7%

Shunts

Abnormal communication b/w 2 chambers or blood vs; blood flows according to pressure gradient

R→L shunt:

- ↓pulm blood flow →poor oxygenation of blood → enters Lt heart →systemic circulation → dusky blueness of mucus membranes and skin (**Cyanosis**)
- Functional anemia → increased synthesis of Hb + RBC mass (**polycythemia**)
- Emboli from peripheral veins donot undergo filteration action of lungs →enter Lt heart →embolize to systemic circulation(**paradoxical emboli**) → cause brain infarction & abscess
- **Clubbing (hypertrophic osteoarthropathy**) of tips of fingers and toes

Shunt

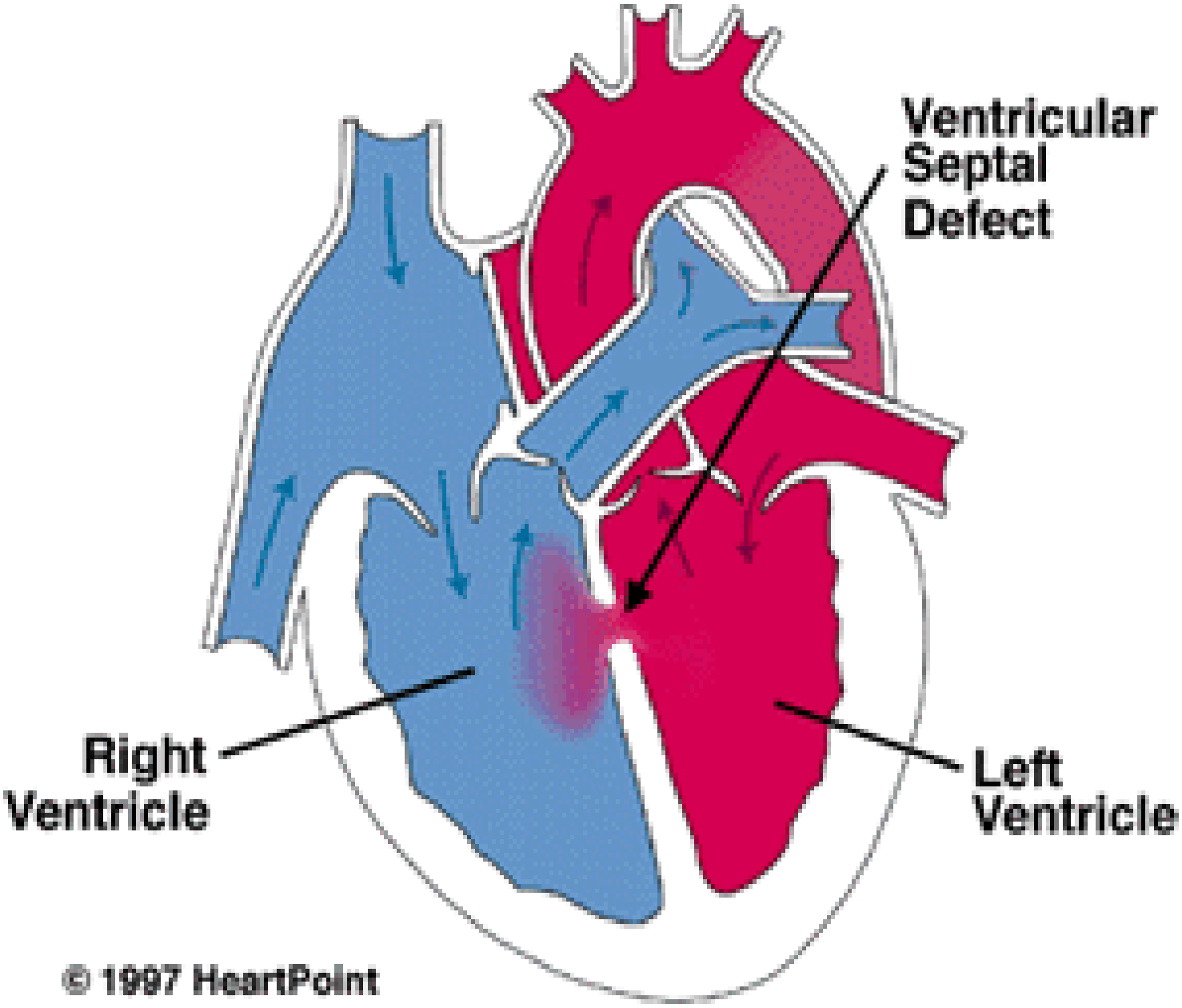
L→R shunt:

- ↑pulm blood flow → ↑pulm pressure →RVH →potential cardiac failure
- ↑pulm blood flow → medial hypertrophy + intimal proliferation to prevent pulmonary edema.
- prolonged ↑pulm pressure → (> even systemic pressure) → reversing the flow from R →L: unoxygenated blood in systemic © → late cyanosis or Eisenmenger syndrome
- Once significant pulmonary HT develops, surgical Rx of cardiac defects not possible

Ventricular Septal Defect (VSD)

- Hole between the two ventricles, incomplete closure of ventricular septum
- Left to right shunt – majority
- Dilated right heart – too much blood to lungs – increase in pulmonary pressure
- Morphology:

- 90% in membranous septum
- 10% lie below pulm valve or within muscular septum
- Mostly single. Multiple VSDs in muscular septum: Swiss cheese septum


Ventricular
Septal
Defect
Right
Ventricle
Left
Ventricle
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VSD

Cl features: depend on size of lesion

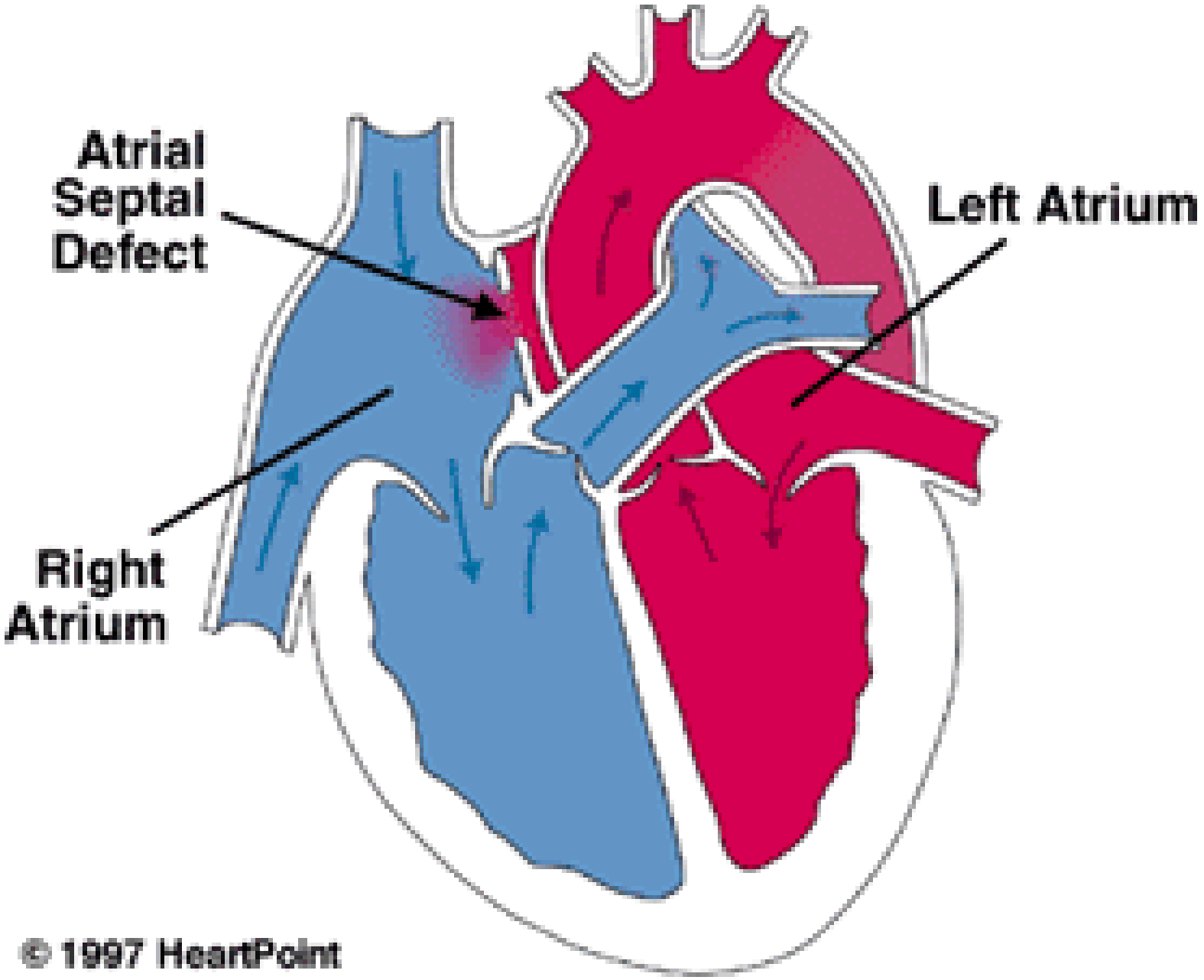
- Small lesions recognized later or may spontaneously close
- Large VSDs recognized early in life, cause Lt- Rt shunts → volume hypertrophy of RV → pulmonary HT since birth. Ultimately, shunt reversal, cyanosis, death
- Not corrected till 1 yr to wait for spontaneous closure

Atrial Septal Defect

- Abnormal fixed opening in atrial septum caused by incomplete tissue formation
- Not to confuse with patent foramen ovale present in 30% of normal individuals
- Unnoticed in infancy and childhood
- Usually presents late in life (30), late cyanotic heart dis.
- L→R shunt at atrial level (pulm vascular resistance is less than systemic and compliance of Rt ventricle is greater than Lt)
- Pulm blood flow increased to 2-4 times, hypertrophy of RA and RV
- Pulmonary HT, RHF unusual

Morphology: 3 types according to location

- Secundum ASD (90%)- deficient or fenestrated oval fossa near centre of septum
- Primum ASD- occur adjacent to AV valves
- Sinus venosus- near entrance of SVC
- AVSD – Atrio ventricular septal defect


Atrial
Septal
Defect
Left Atrium
Right
Atrium
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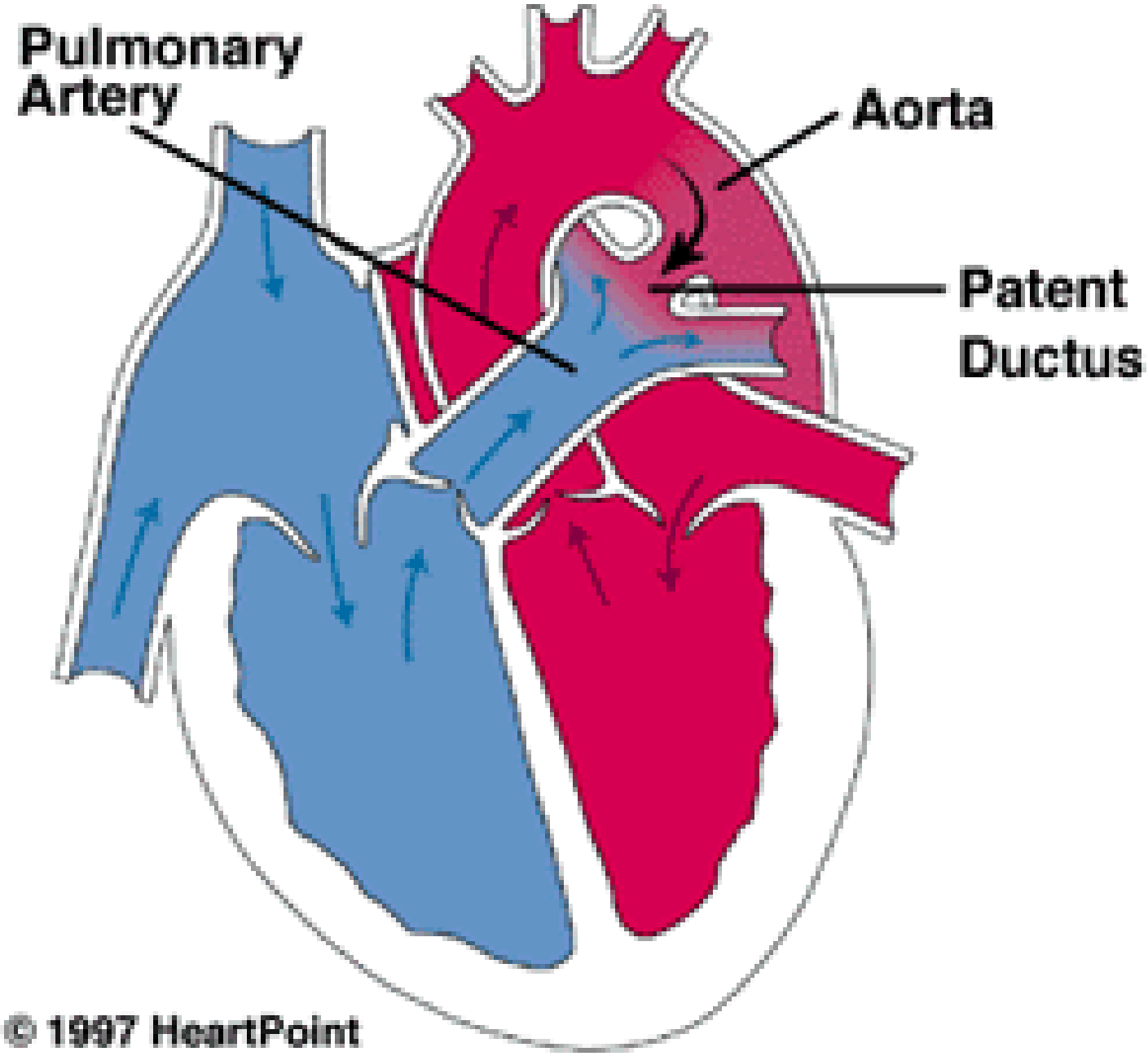
Patent Ductus Arteriosus

- Ductus arteriosus is normal connection b/w aorta and bifurcation of pulmonary A
- Normally closes at 1st or 2nd day of life, > 3months persistence is abnormal
- Cause: possibly due to ↑ levels of PGE2 after birth

- seen in children with respiratory distress syndrome

- pharmacologic closure with indomethacin (PGE2 inhibitor)

- Most often does not produce functional difficulties at birth
- A narrow ductus: no effect on growth and development during childhood
- Harsh machinery like murmur


Pulmonary Artery
Aorta
Patent Ductus
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Right to Left shunts (Cyanotic CHD)

1 Tetrology of Fallot (TOF)- 6-15%

2 Transposition of great arteries -4-10%

3 Persistent truncus arteriosus – 2%

4. Tricuspid atresia and stenosis- 1%

Cyanosis in early postnatal life

Tetralogy of Fallot

- Combination of shunts with obstruction with functional shunting of blood
- **Most common cyanotic heart disease**
- 4 features:

1. VSD
2. Displacement of aorta to right side so that it overrides the septal defect
3. Sub-pulmonary stenosis (obstruction)
4. Right ventricular hypertrophy

- Clinical manifestations dependant on extent of pulmonary stenosis & VSD

- 2 types : Cyanotic and Acyanotic (**pink tetralogy**)
- **Cyanotic**:

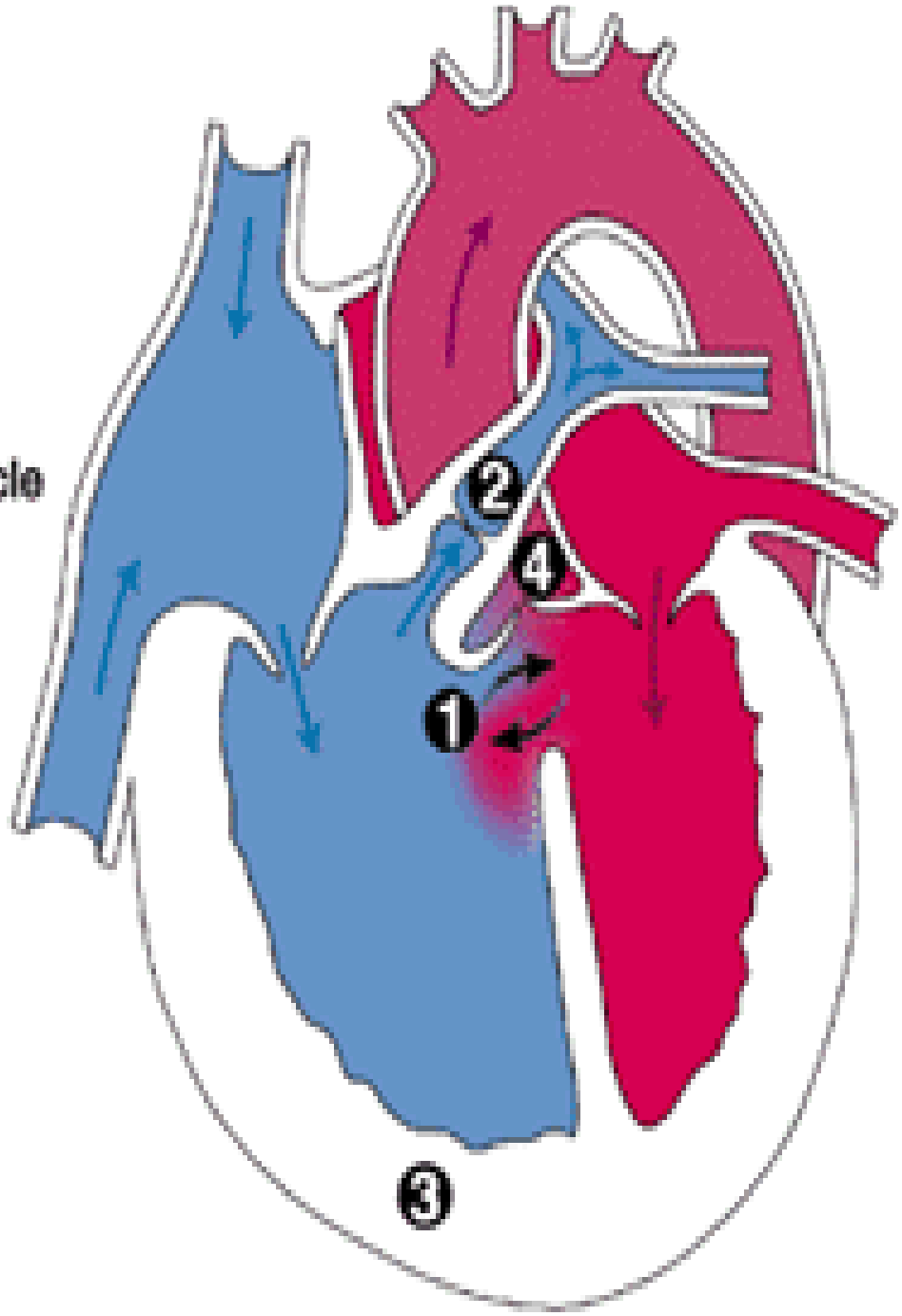
  - Pulm stenosis is greater →↑ resistance to flow of blood in RV → it flows to LV→ Cyanosis

  - Effects: - pressure hypertrophy of RA and RV
    - small tricuspid valve
    - small lt atrium & ventricle
    - enlarged aortic orifice
- **Acyanotic tetrology:**

- VSD larger, pulmonary stenosis mild: L → R shunt, behaves like isolated VSD

Boot shaped heart

1. Ventricular Septal Defect
2. Pulmonary Stenosis
3. Hypertrophy of Rt. Ventricle
4. Overriding Aorta



Transposition of Great arteries

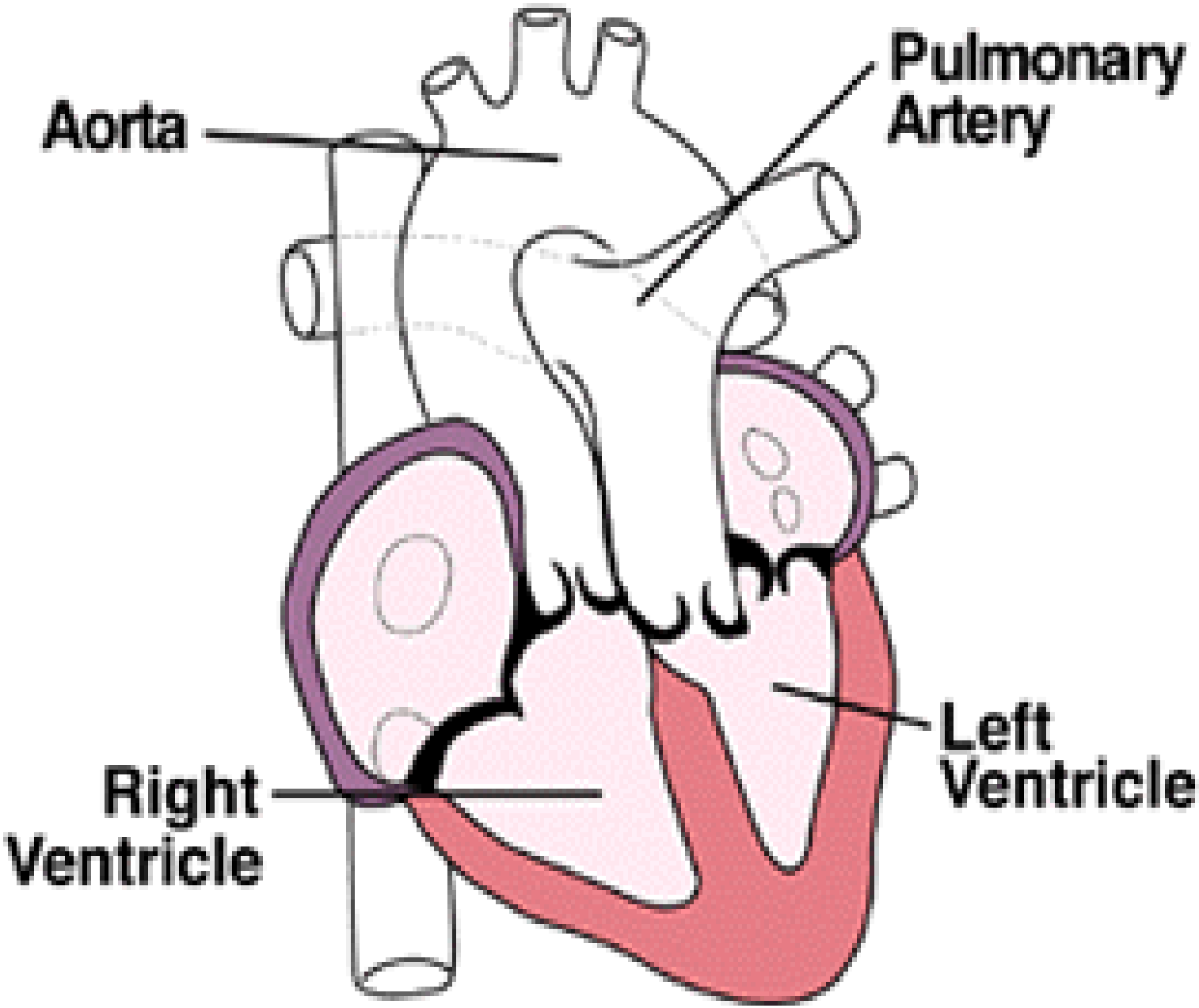
- **Regular transposition**:

  - Aorta arises from RV and Pulmonary A from LV - cyanosis from birth

- **Corrected Transposition**:

  - Aorta arises from RV, Pulmonary A from LV + Pulm veins drain into RA, Sup and Inf vena cava into LA

  - Physiologically corrected circulation


Aorta
Pulmonary Artery
Left Ventricle
Right Ventricle

Persistent Truncus Arteriosus

- Arch that separates aorta from pulmonary A fails to develop. A single large vessel receives blood from both the ventricles
- Often associated VSD
- Early systemic cyanosis
- Poor prognosis

Tricuspid Atresia and stenosis

- Often associated with pulmonary stenosis or atresia
- Atresia- absence of tricuspid orifice, there is dimple in floor of rt atrium
- Stenosis- tricuspid ring is small

Obstructions (Obstructive CHD)

Coarctation of Aorta:

- Localised narrowing in any part of the aorta
- More common in males, females with Turner syndrome
- **Postductal or adult type**

  - Obstruction is just distal to ductus arteriosus which is closed

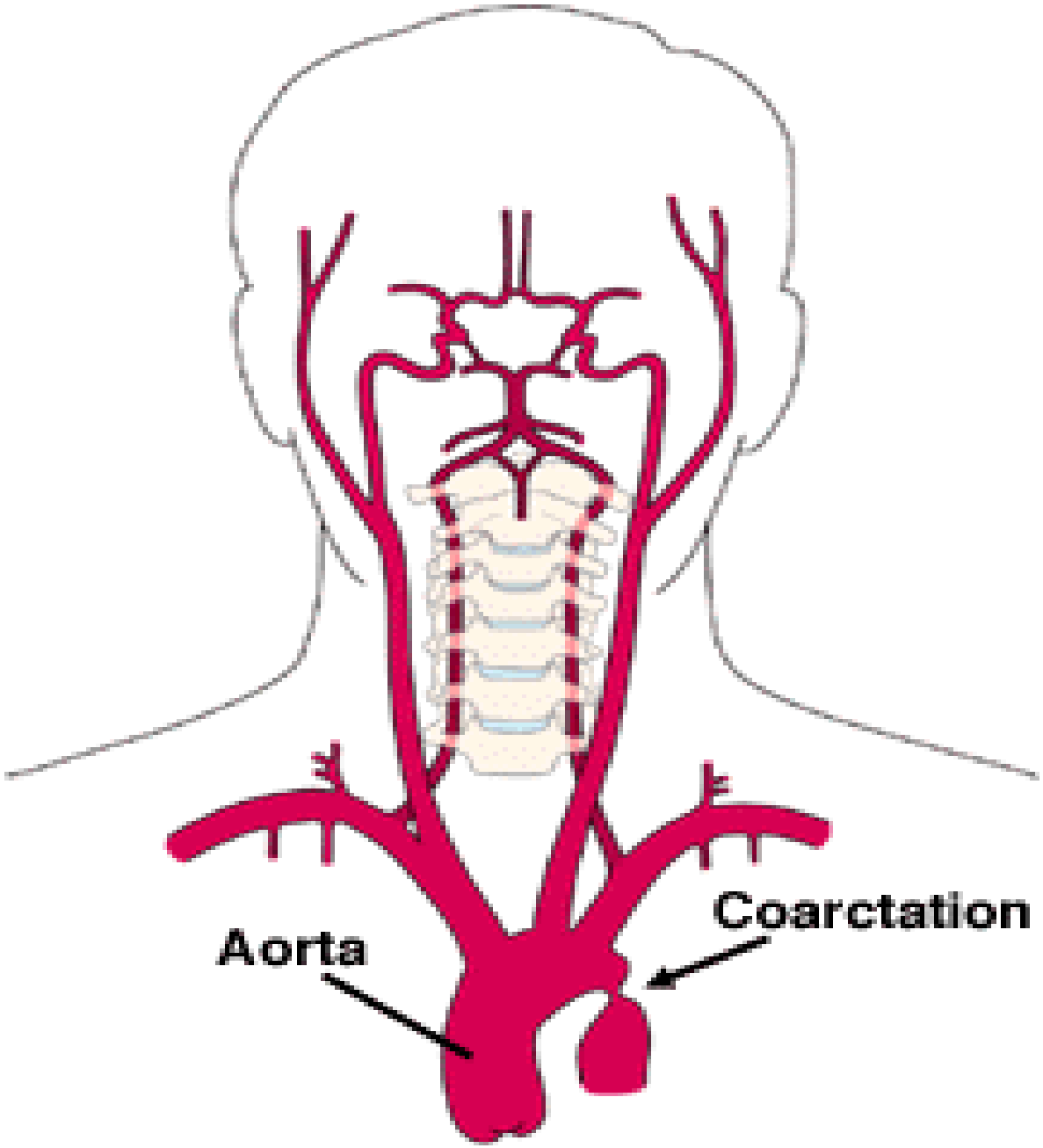
  - Characterized by HT in upper extremities, weak pulses and low BP in the lower extremities, effects of arterial insufficiency such as coldness and claudication

  - With time, development of collaterals b/w pre-stenotic and post- stenotic segment with enlargement of intercostal arteries → rib erosion

- **Preductal or Infantile type**:

  - narrowing proximal to ductus arteriosus which remains patent

  - lower half of body cyanosed while upper part of body receives blood from aorta


Aorta
Coarctation
© 1997 HeartPoint

Aortic stenosis and atresia

- Most common anomaly of aorta is congenital bicuspid valve. Not much functional significance except predisposes it to calcification
- Congenital aortic atresia rare & incompatible with life
- Aortic stenosis- congenital or acquired(RHD)
- 3 types of congenital AS:

1. **Valvular**: cusps thickened and malformed
2. **Subvalvular**: thick fibrous ring under the aortic valve
3. **Supravalvular**: uncommon

- May be assoc with hypoplastic heart synd: fatal in neonates

Pulmonary Stenosis and Atresia

- Stenosis

  - commonest form of obstructive CHD

  - occurs as component of TOF or isolated defect

  - fusion of cusps of pulmonary valve forming diaphragm like obstruction

- Atresia

  - no communication b/w rt ventricle & lungs

  - blood goes to left heart through interatrial septal defect and enters lungs via PDA