



Congenital Heart Disease

Mary Rummell, RN, MN, CPNP, CNS
Clinical Nurse Specialist
Pediatric Cardiology/Cardiac Surgery



Objectives

♥ The learner will:

- ♥ Identify 3 areas in the developing heart that result in congenital heart defects.
- ♥ Discuss 3 important components of a cardiovascular assessment.
- ♥ List the 3 cardinal signs of congenital heart disease.
- ♥ Describe the clinical symptoms in each category.
- ♥ Identify 3 defects that are included in each category.



Congenital Heart Disease

A “congenital” anomaly originating in the developing fetus is often considerably modified, at least physiologically, by the dramatic circulatory adjustments at birth. Weeks, months, or even years may then elapse before the anomaly evolves into the “typical” clinical picture. Both physiologic and structural changes subsequently continue, or conversely, the malformation may “vanish.” Joseph Perloff (1978)

♥ Definition:

- ♥ Congenital (Latin) Con = together, Genital = Born
- ♥ Actually occurs seven months before birth
- ♥ Most defects are compatible with intrauterine life until a term birth



Incidence of Congenital Heart Disease

- ♥ 8-10 per 1000 live births (approx. 1 %)
 - ♥ (Does not include preterm infants with PDAs or people with slight abnormalities of aortic valve)
- ♥ Increases to approx 3% with subsequent siblings
- ♥ Risk increases with left sided lesions – some quote 15%.
- ♥ Also increases in offspring of parents with CHD
- ♥ Remains one of the greatest causes on neonatal mortality & morbidity
- ♥ Leading expense for neonatal health care



Etiology of Congenital Heart Disease: Multifactorial

♥ Maternal:

- ♥ Disease: Diabetes, Collagen disease (Lupus),
- ♥ Seizure disorders
- ♥ Medications: anticonvulsants, diazepam
progesterone/estrogen, alcohol, street drugs (cocaine) ,
Retin-A, lithium, thalidamide, lithium, warfarin, aspirin, ACE
inhibitors

♥ Genetic:

- ♥ Chromosome abnormalities: 13, 18, 21;
- ♥ Turner's Syndrome,
- ♥ DiGeorge Syndrome (22q11 deletion),
- ♥ Williams Syndrome

♥ Environmental:

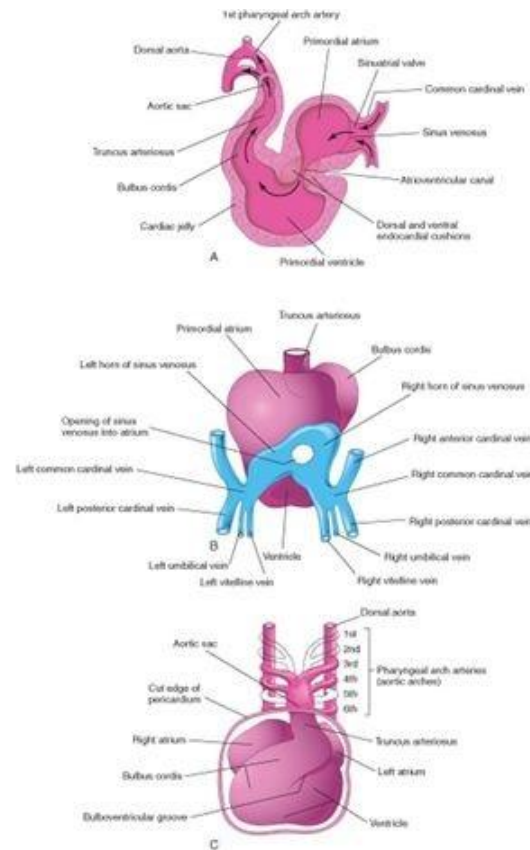
- ♥ Viral infections (Rubella, CMV), ????
- ♥ Toxins



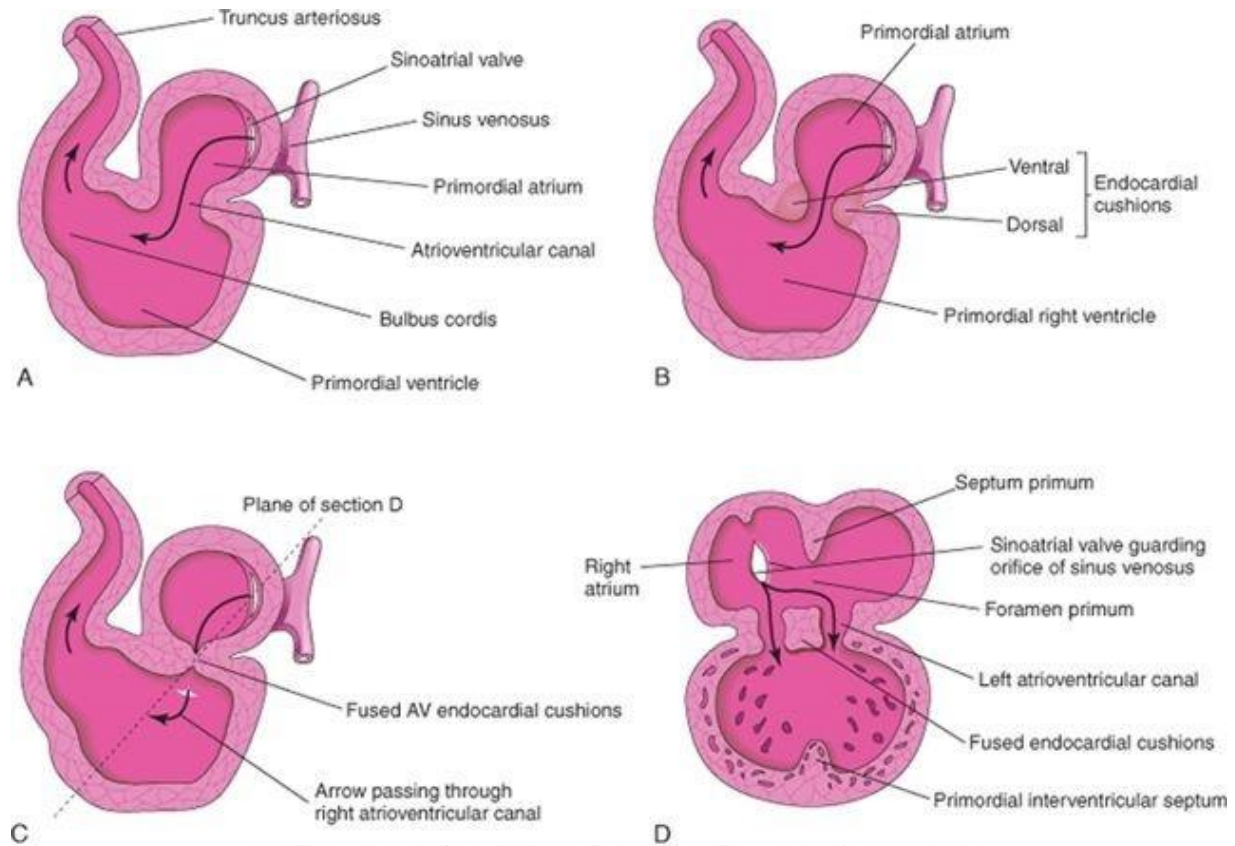
Embryologic Timeline

- ♥ **Week 3:** Cardiogenic plate, endocardial tubes
- ♥ **Week 4:** Fusion of endocardial tubes to single median tube, first contraction at day 22, cardiogenic looping
- ♥ **Week 5:** Beginning of true circulation, septation of primum atrium, AV valves to 3 chamber heart
- ♥ **Week 6:** Septum secundum, septation of bulbus and ventricle, divided truncus arteriosus
- ♥ **Week 7:** Four chambered heart, absorption of pulmonary veins

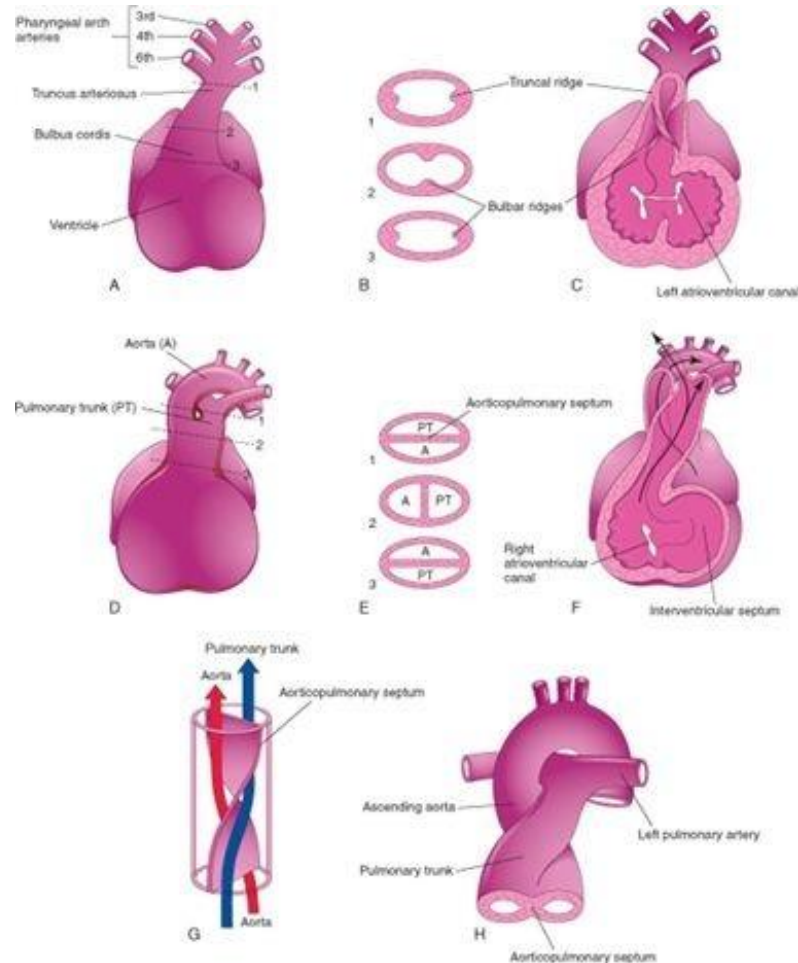
Cardiac Looping



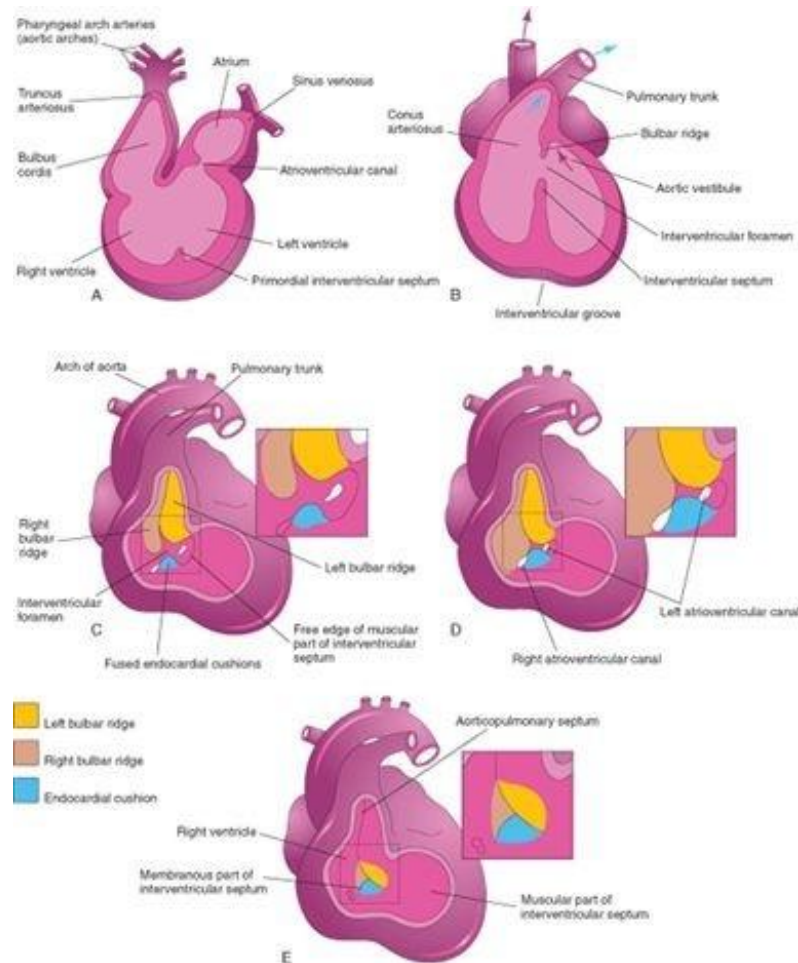
Blood Flow Through Partitioning Heart



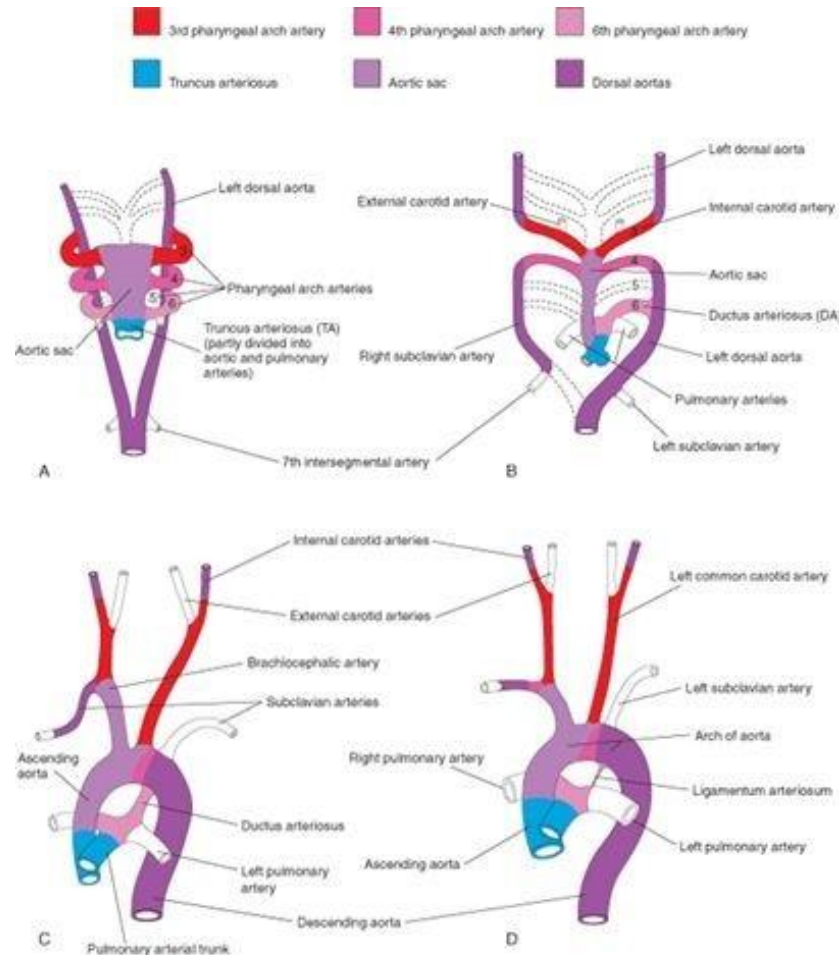
Partitioning of Truncus Arteriosus



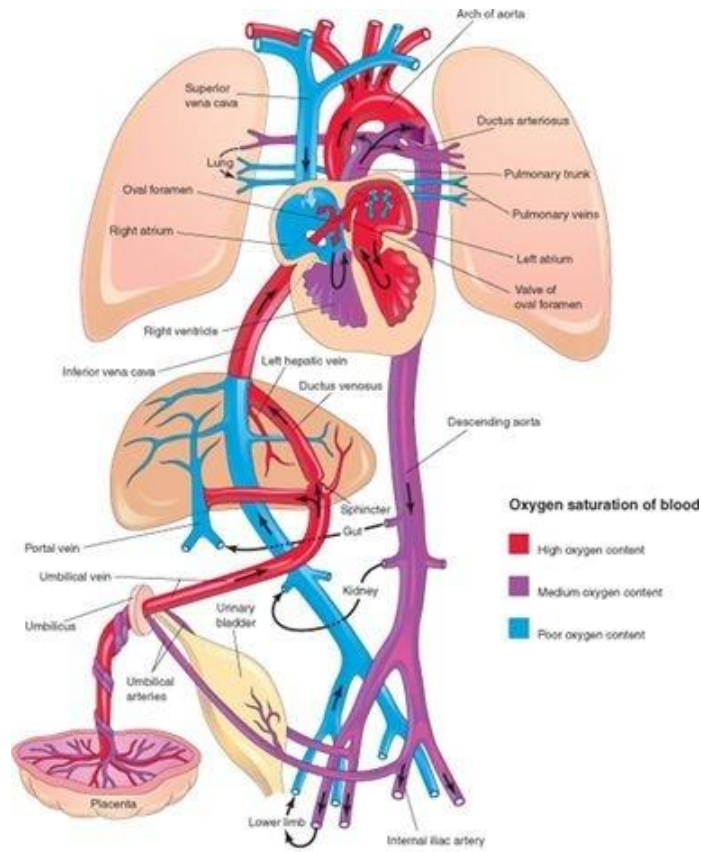
Development of the Ventricular Septum



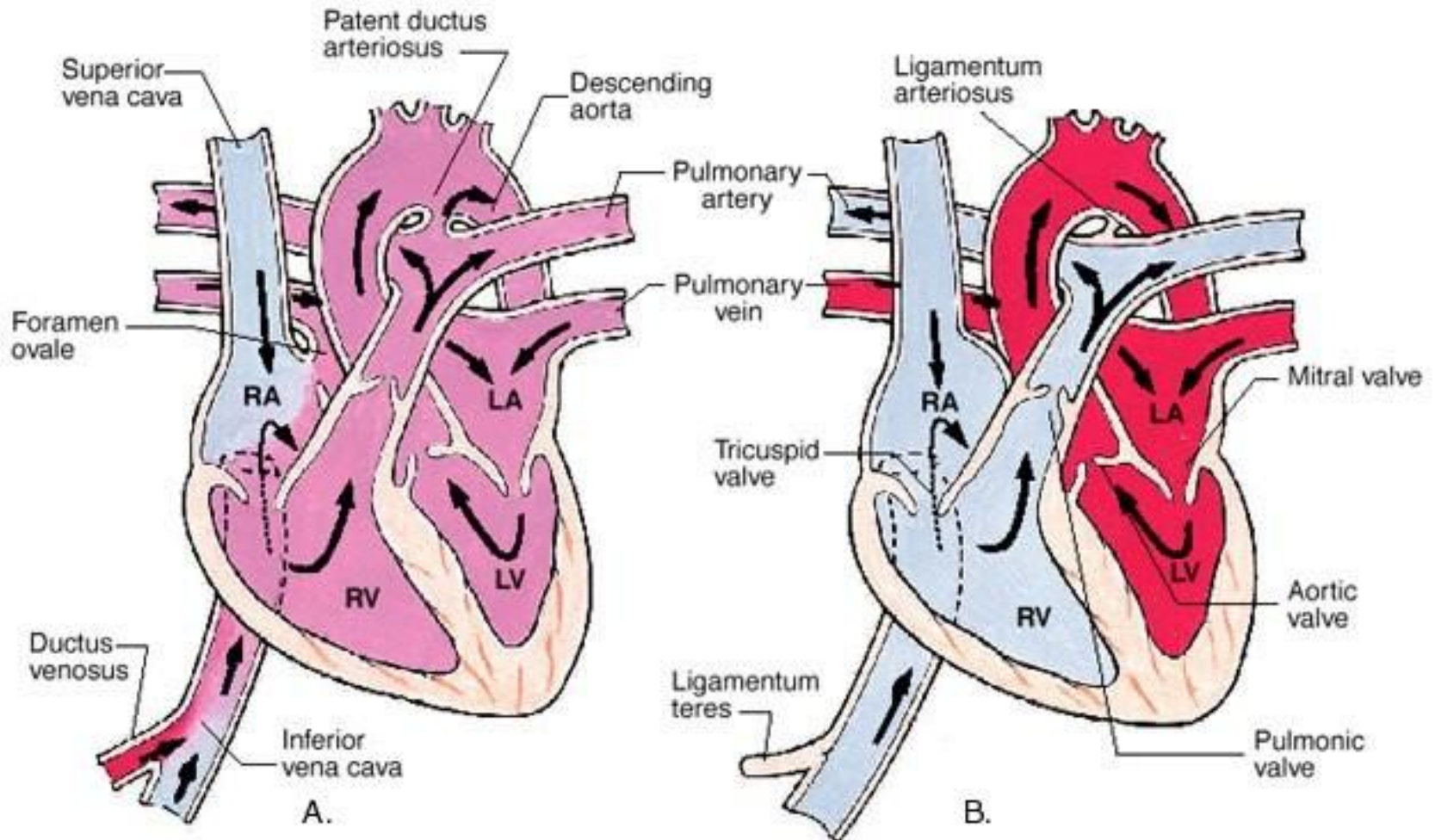
Development of Aortic Arch



Fetal Circulation



Newborn Physiology





Closure of fetal shunts

♥ Ductus arteriosus

Constriction by 10 to 14 days. Anatomic closure and formation of ligamentum arteriosus by 12th week

May remain open much longer with prematurity or hypoxia.

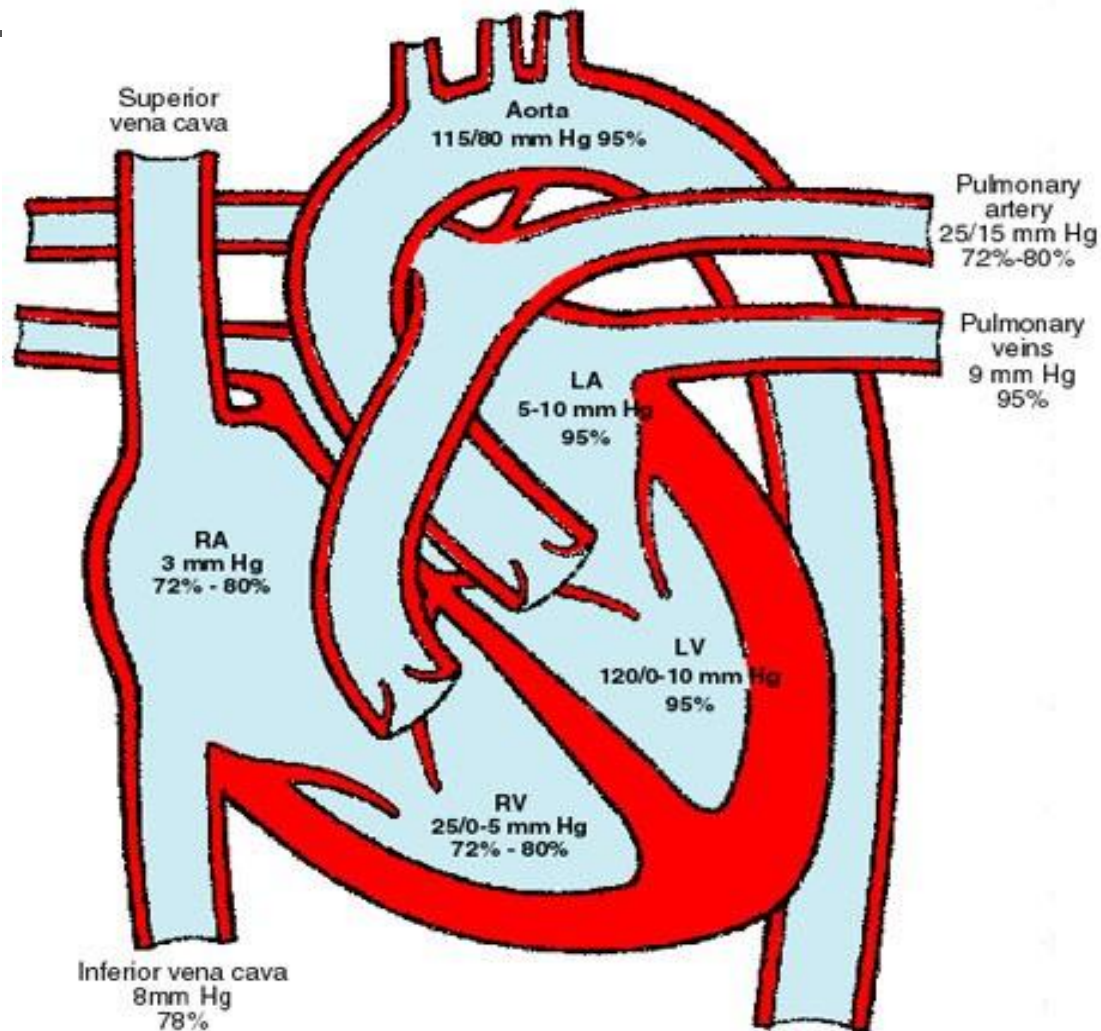
♥ Foramen ovalae

Functional closure at birth, anatomic closure has been seen by 3 months, 50% by 5 years, 25% of adults still have PFO.

♥ Ductus venosus

Constriction of vessel within hours of birth as portal pressure falls with removal of placenta

Normal Heart Pressures and Saturations





Cardiac Output:

Heart Rate X Stroke Volume

♥ Normal Stroke Volume

- Newborn = 400 mL/kg/min
- Infant = 200mL/kg/min
- Child = 100mL/kg/min

♥ Increased oxygen demand
(consumption) increase cardiac output:
Heart rate and/or stroke volume



Heart Rate:

The number of contractions/minute

Principles:

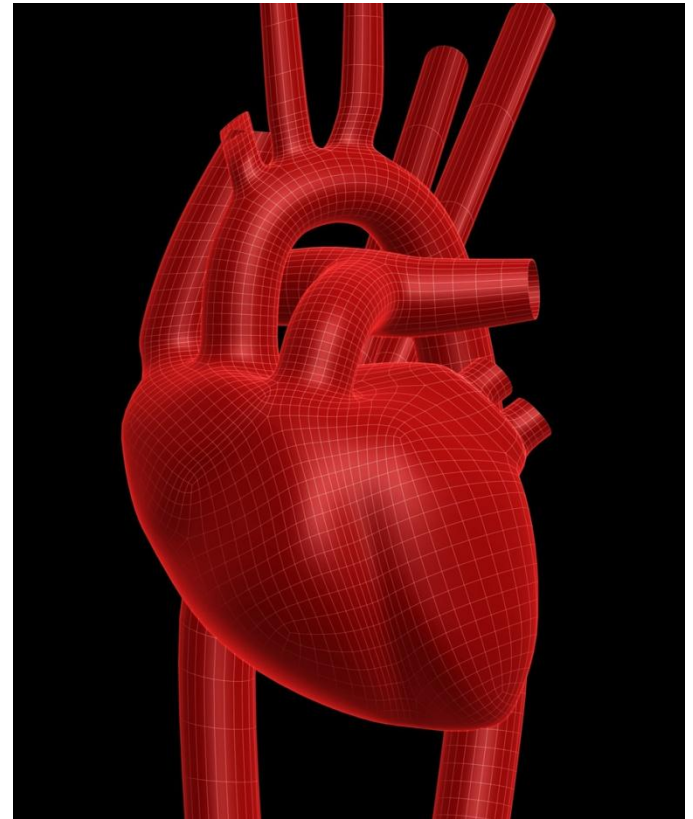
- ♥ Increased heart rate improves Cardiac Output until diastolic filling time and coronary perfusion are compromised
- ♥ Decreased heart rate necessitates an increase in stroke volume to maintain Cardiac Output
- ♥ Neonates are heart rate dependent for Cardiac Output

Stroke Volume:

The volume of blood ejected by the ventricle with each contraction

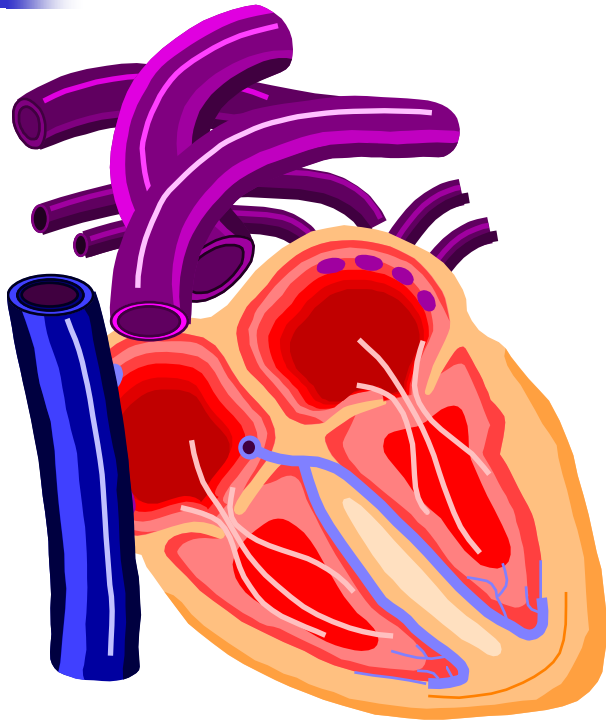
♥ Factors affecting Stroke Volume:

- ♥ Preload
- ♥ Afterload
- ♥ Contractility



Cardiac Output:

Amount of blood pumped from the ventricle/minute



- ♥ MUST meet the metabolic demands of the body.
- ♥ Normalized to BSA (Body Surface Area)
- ♥ The cardiovascular assessment is your measurement of how well your patient is meeting the metabolic demands.



Assessment

- ♥ Systematic
- ♥ Consistent
- ♥ Head to toe
- ♥ Assess other systems with CV system
- ♥ Always start with inspection and note general appearance
SICK or WELL



Cardiovascular Assessment

- ♥ End-organ perfusion
- ♥ Skin color and temperature
- ♥ Pulses (peripheral & central)
- ♥ Capillary refill
- ♥ Heart rate/rhythm
- ♥ Heart sounds
- ♥ Blood pressure



Inspection

♥ Head/neck

Color

Cyanosis- central vs peripheral

Pallor - hemoglobin

Work of breathing

Nasal flaring, retractions

Neck veins

Palpation



Peripheral pulses

Rate/rhythm

Quality

Equality

Location

Carotid

Central – Brachial,
femoral

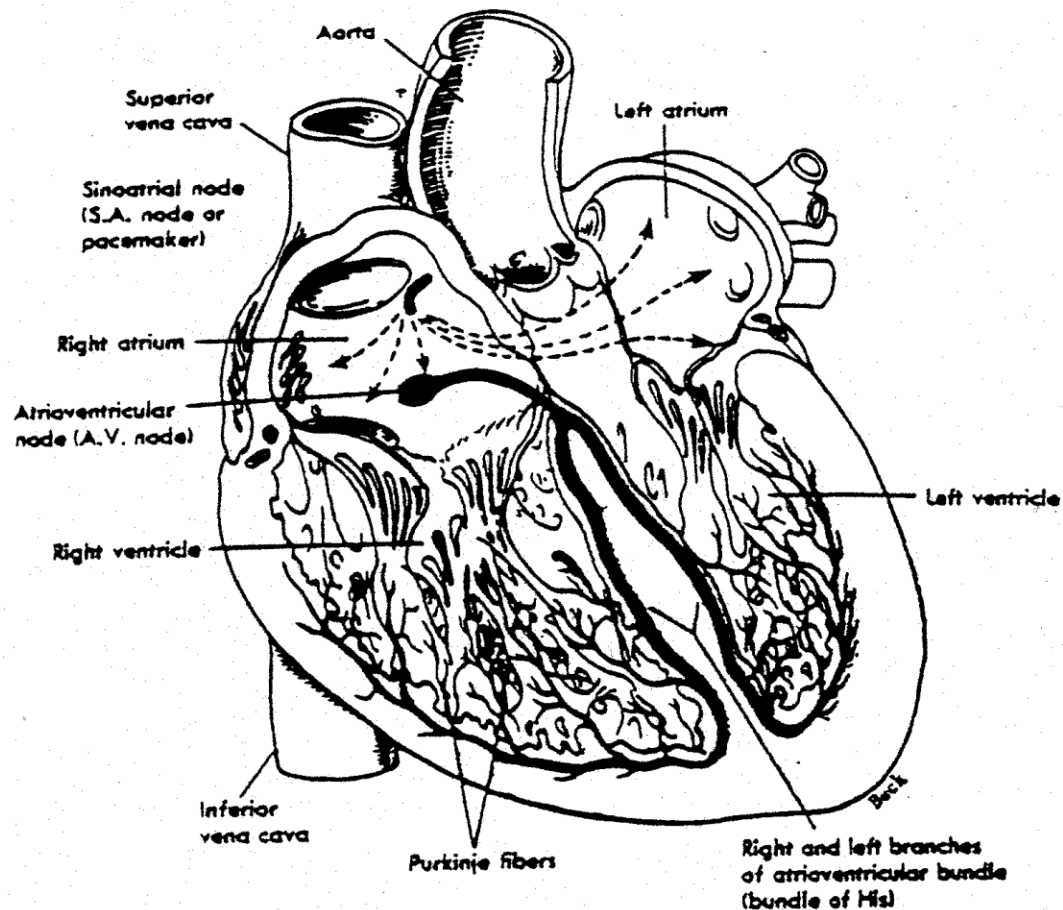
Peripheral – Radial,
dorsalis pedis,
posterior tibial

Palpation



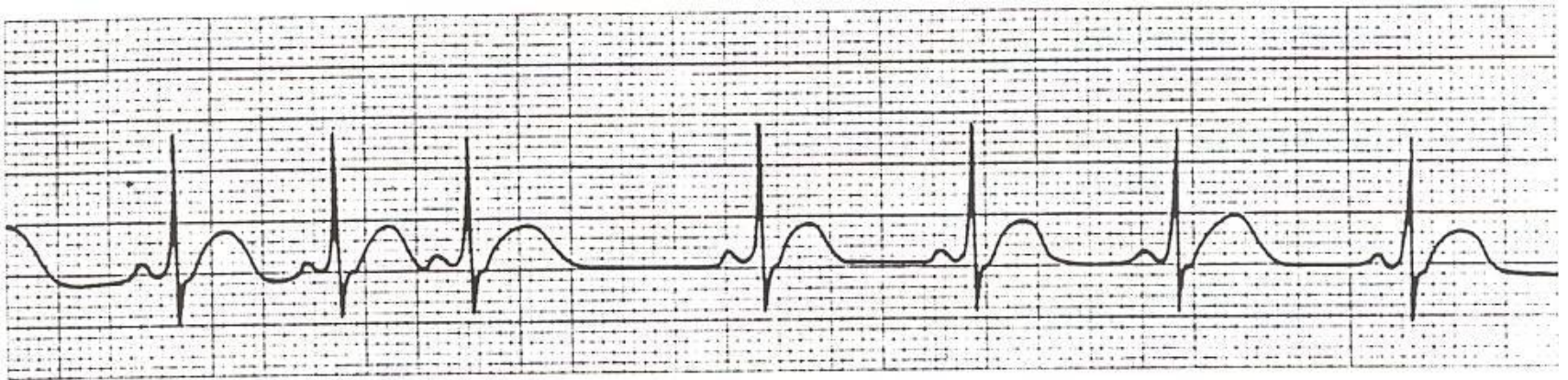
- ♥ Anterior chest
 - Thrills
 - Precordium – Point of Maximum Impulse
- ♥ Skin
 - Peripheral perfusion
 - Temperature – one of best ways to assess CO
 - Moisture
- ♥ Abdomen
 - Liver

Conduction System

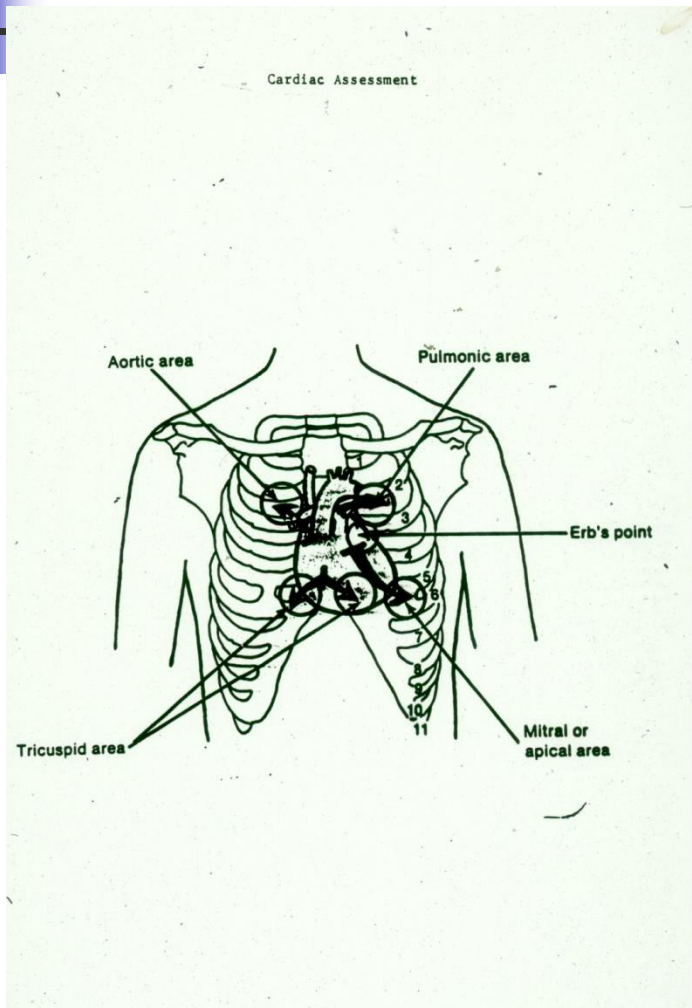


Rhythm Interpretation

♥ Sinus Arrhythmia

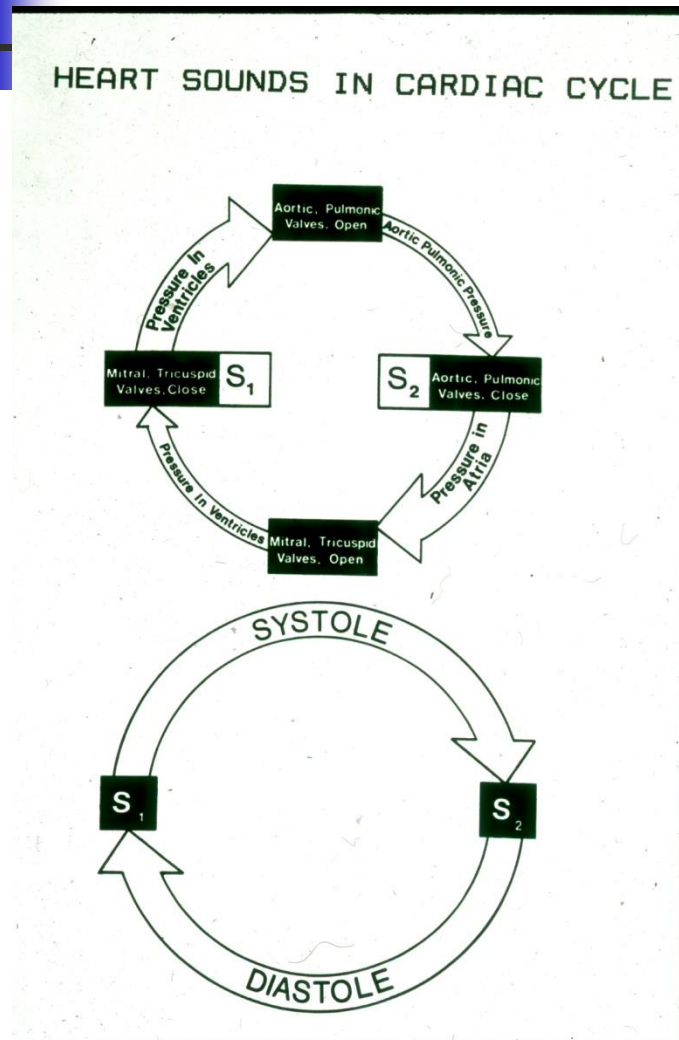


Auscultation



- ♥ Key is to listen to one sound at a time!
- ♥ Listen in 2 positions.
- ♥ Listen in systematic pattern.

Heart Sounds



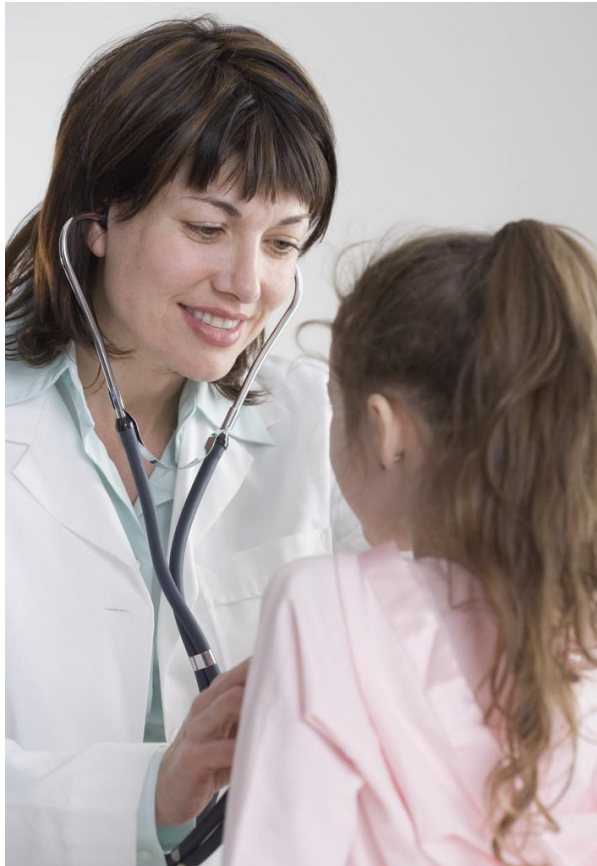
- ♥ S₁— beginning of systole, AV valves close
- ♥ S₂ — end of systole, closure of semilunar valves
- ♥ S₃— normal in children, related to rapid filling of ventricle
- ♥ S₄—abnormal, heard late in diastole or early systole, heard in CHF, decreased ventricular compliance



Murmurs

- ♥ Definition: “Whooshing” sounds that occur in various stages of diastole and/or systole due to turbulent blood flow.
- ♥ Causes:
 - ♥ Normal heart murmur
 - ♥ Increased rate of blood flow
 - ♥ Blood flow forced forward through an incompetent/constricted/irregular valve
 - ♥ Blood flows back (regurgitant) through an incompetent valve, a septal defect, or a patent ductus arteriosus

Characteristics of Murmurs



- ♥ Timing
- ♥ Frequency or pitch
- ♥ Location
- ♥ Intensity (Graded I – VI)
- ♥ Radiation
- ♥ Quality
- ♥ Effect of respiration
- ♥ Effect of position



Significance of Murmurs

- ♥ A systolic murmur may be normal. A diastolic murmur is never normal.
- ♥ The intensity of the murmur does NOT indicate the severity of heart disease.
- ♥ You may not hear a murmur in serious CHD.
- ♥ You may hear a Grade 4-5 murmur in insignificant CHD.

Normal Heart Murmurs

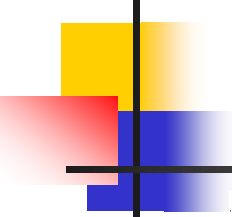


- ♥ Newborn – Peripheral pulmonary stenosis
- ♥ Infant/child – Pulmonary ejection murmur
- ♥ Child – Still's murmur, vibratory murmur
- ♥ Teenager – Venous hum

Blood Pressure

- ♥ Cuff size – EXTREMELY important
- ♥ Extremity – use right arm
- ♥ Normal Range
- ♥ LAST vital sign that will indicate that the child is in trouble





Normal Dinamap Blood Pressure Ranges

Normal Dinamap BP Values
[Systolic/Diastolic (Mean Arterial Pressure)]

AGE-GROUP	MEAN	RANGE	90 TH PERCENTILE	95 TH PERCENTILE
Newly born (Term Birth)	65/41 (50)	50-70/ 25-45	75/49 (59)	78/52 (62)
Neonate (To 28 days)	84/	60-90/ 20-60	100/	104/
Infant (1 – 12 mos)	94/52	87-105/ 53-66	105/68 (83)	110/70 (86)
Toddler (2 – 5 yrs)	94/56	90-105/ 53-66	108/68 (82)	114/70 (85)
School Age (6 – 10 yrs)	98/62	97-112/ 57-71	114/74	118/78
Adolescent (11 - 13 yrs) (14 – 18 yrs)	106/66 (B)116/68 (G)108/66	112-128/ 66-80	120/78 (B)130/82 (G)122/80	124/82 (B)136/86 (G)126/82



Cardiac Reserve

- Adaptive mechanisms activated to increase cardiac output.

Mechanical factors

Hypertrophy

Dilation

Biochemical factors

Increase energy production

Activation of the adrenergic nervous system



Cardinal Signs of Heart Disease

- ♥ Cyanosis
- ♥ Decreased Systemic Perfusion
- ♥ Tachypnea



Cyanosis

Congenital Defects

- Transposition of the Great Arteries
- Tetralogy of Fallot
- Tricuspid Atresia
- Pulmonary Atresia

Clinical Presentation

- ♥ Central rather than peripheral.
- ♥ May not be immediately present.
- ♥ Not evident until a significant amount of reduced hemoglobin is present.
- ♥ Source of systemic arterial blood flow.
- ♥ Tachypnea without distress



Consequences of Cyanosis

- ♥ Polycythemia
- ♥ Clubbing
- ♥ Hypoxic spells and squatting
- ♥ Central nervous system complications
- ♥ Bleeding disorders



Key Questions

- ♥ How does blood get to the lungs ?
- ♥ How does blood get to the body?
- ♥ Is the pulmonary/systemic blood flow dependent on the ductus arteriosus?
- ♥ If the answer to any of these questions is yes, immediate intervention is mandatory.
 - ♥ Prostaglandin E1 (PGE)
 - ♥ Atrial septostomy
 - ♥ Arterial to pulmonary shunt
 - ♥ Palliative/reconstructive surgery



Prostaglandin E1 (PGE)

♥ Indications for use:

- ♥ Right heart outflow obstruction
- ♥ Transposition of the great arteries
- ♥ Left heart outflow obstruction

♥ Contraindications for use:

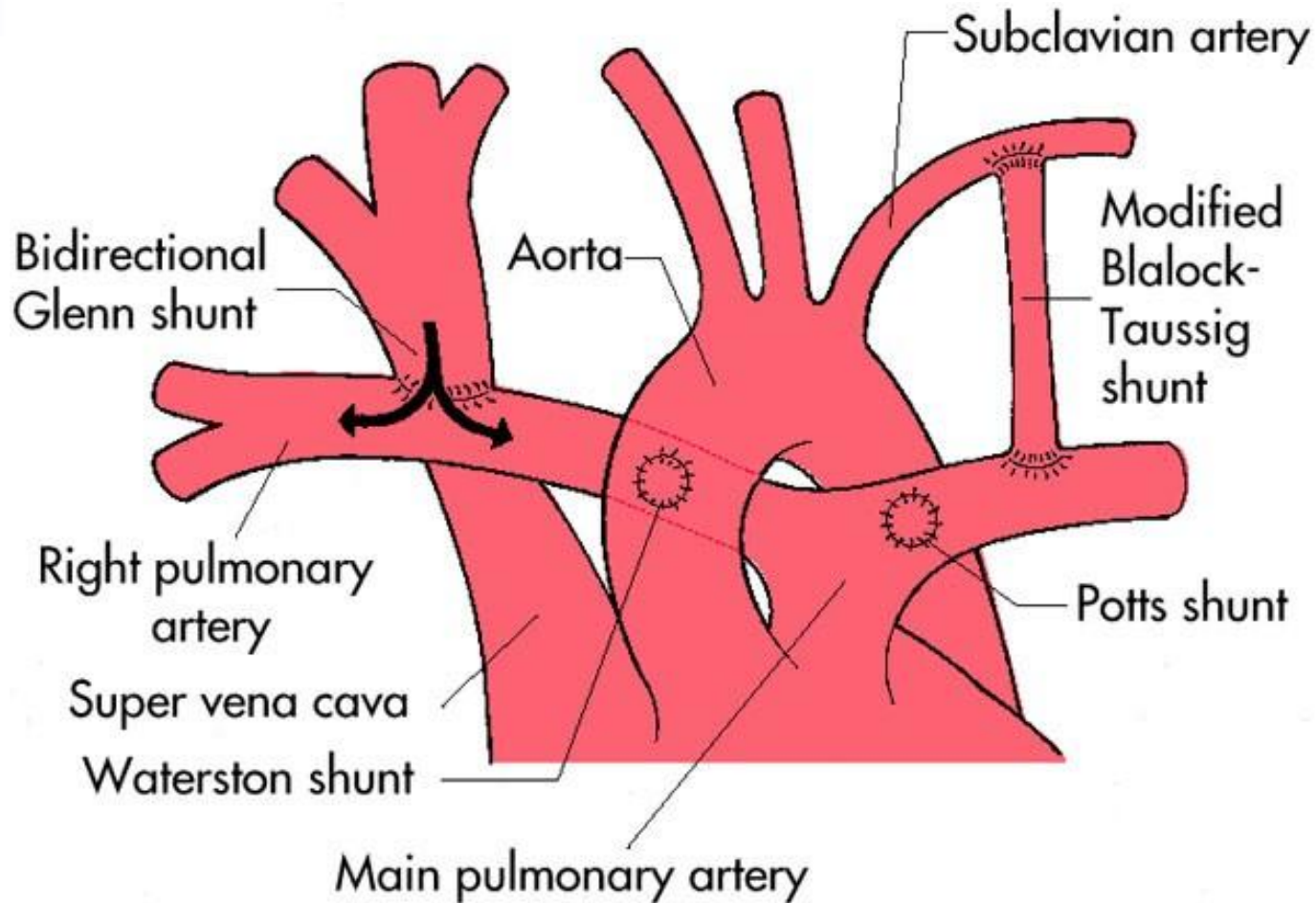
- ♥ Total anomalous pulmonary vascular return (TAPVR)
- ♥ Respiratory distress
- ♥ Left-to-right shunts with increase pulmonary blood flow



Prostaglandin E1

- ♥ Maintains patency of the ductus arteriosus in ductal dependent congenital heart defects to provide either pulmonary or systemic circulation.
- ♥ Side effects:
 - ♥ Cardiovascular: Vasodilation, arrhythmias, hypotension, edema
 - ♥ Neurological: Seizures, hyperthermia, irritability, lethargy
 - ♥ Respiratory: Apnea, hypoventilation, bronchospasm, tachypnea
 - ♥ Renal: anuria
 - ♥ Hematologic: Thrombocytopenia, hemorrhage, DIC

Modified Blalock-Taussig Shunt

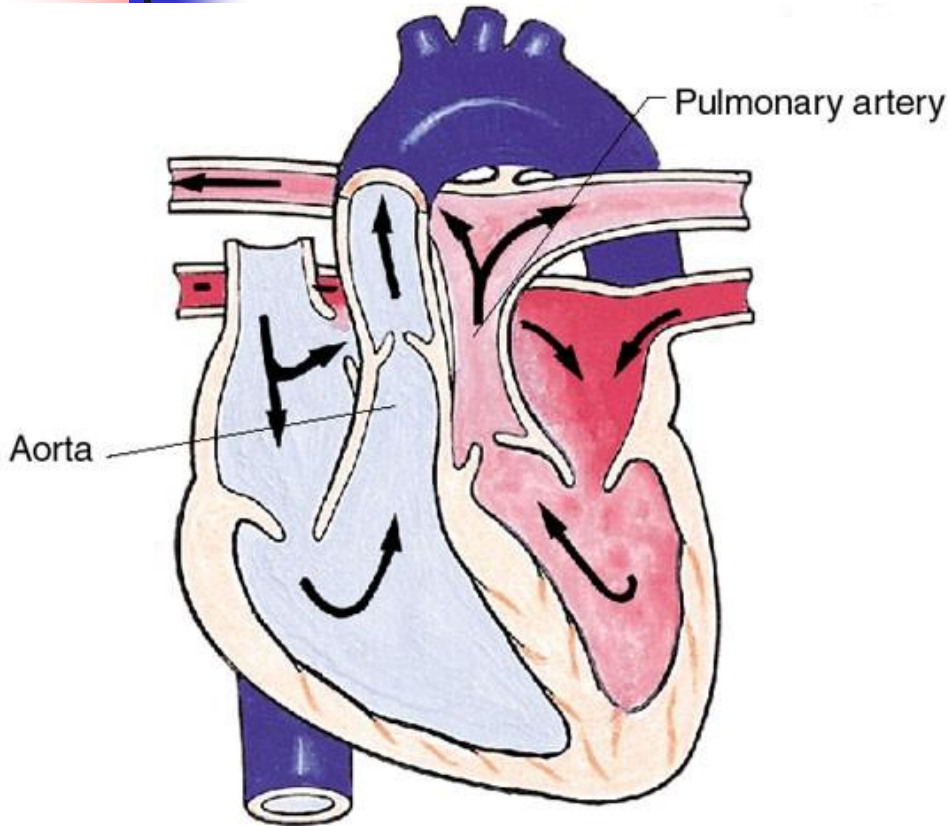




Nursing Considerations:

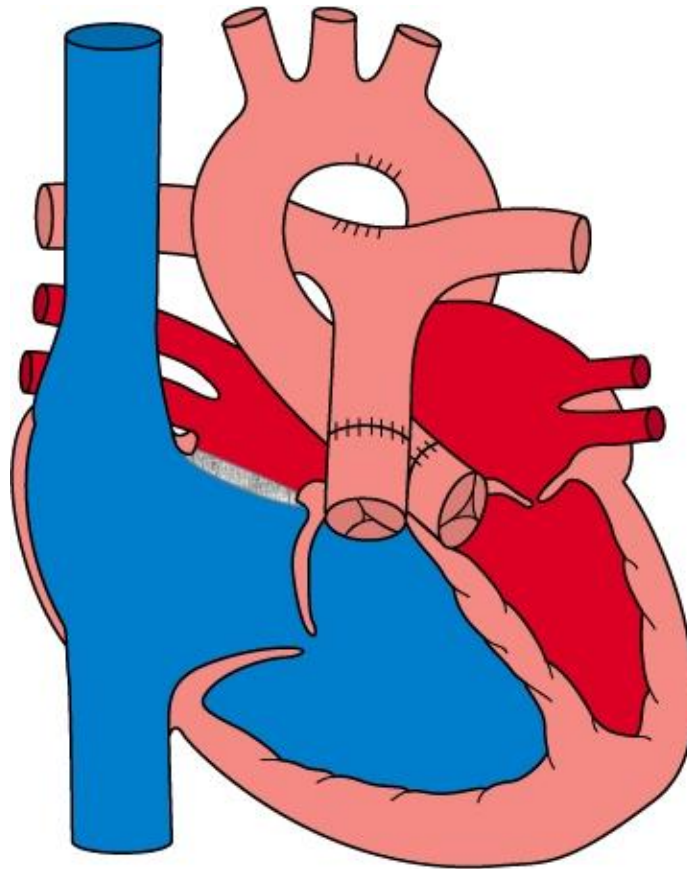
- ♥ Intracardiac Mixing of Systemic and Pulmonary Circulation
- ♥ Assessment of Cardiac Output:
- ♥ Shunt
 - ♥ Murmur present?
 - ♥ Hydration status?
 - ♥ Oxygen saturation?

Transposition of the Great Arteries

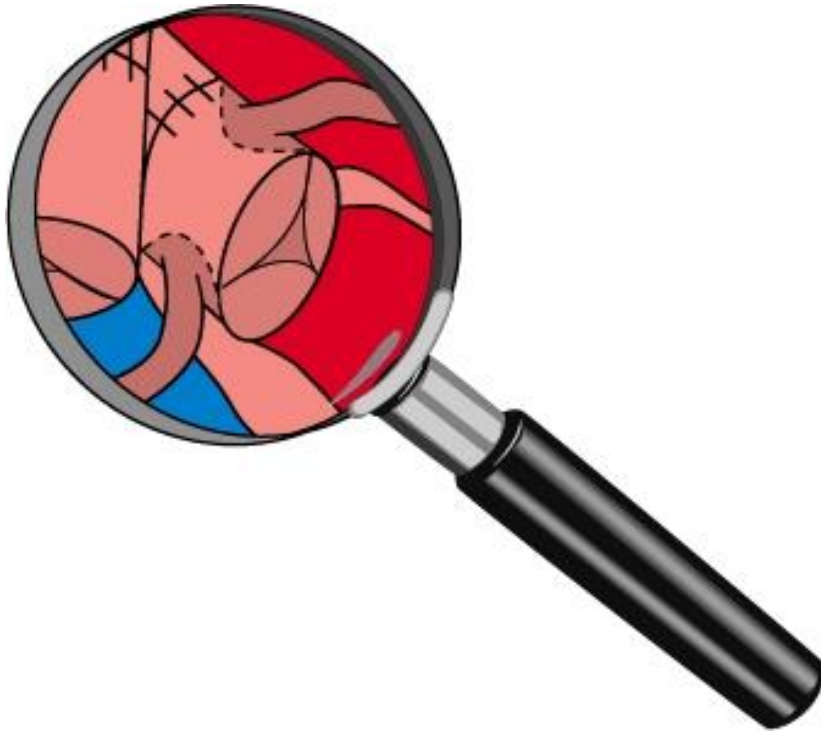


- ♥ 5% of all CHD
- ♥ Males > female by 3:1
- ♥ Must have communication between left and right sides of the heart
- ♥ Surgical repair before left ventricular muscle strength decreases
- ♥ Oxygen saturation may be greater in lower extremities

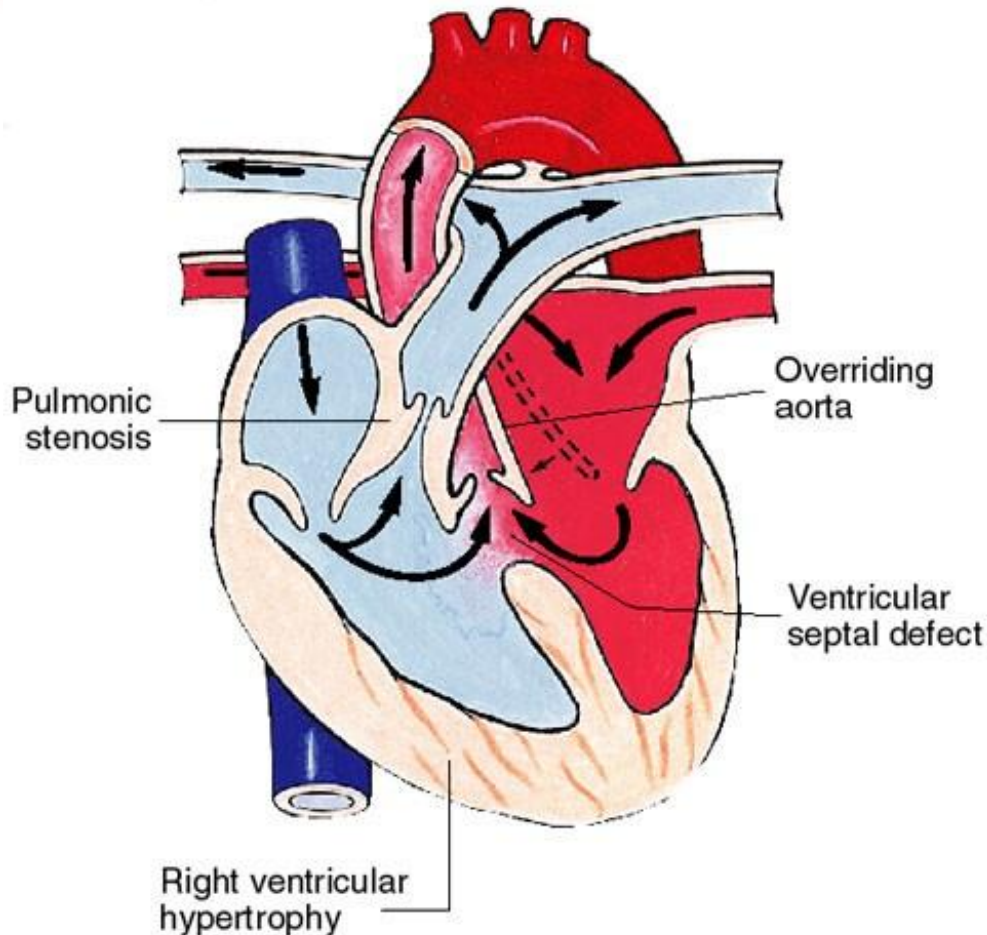
Arterial Switch Procedure



Coronary Artery Repair for TGA



Tetralogy of Fallot



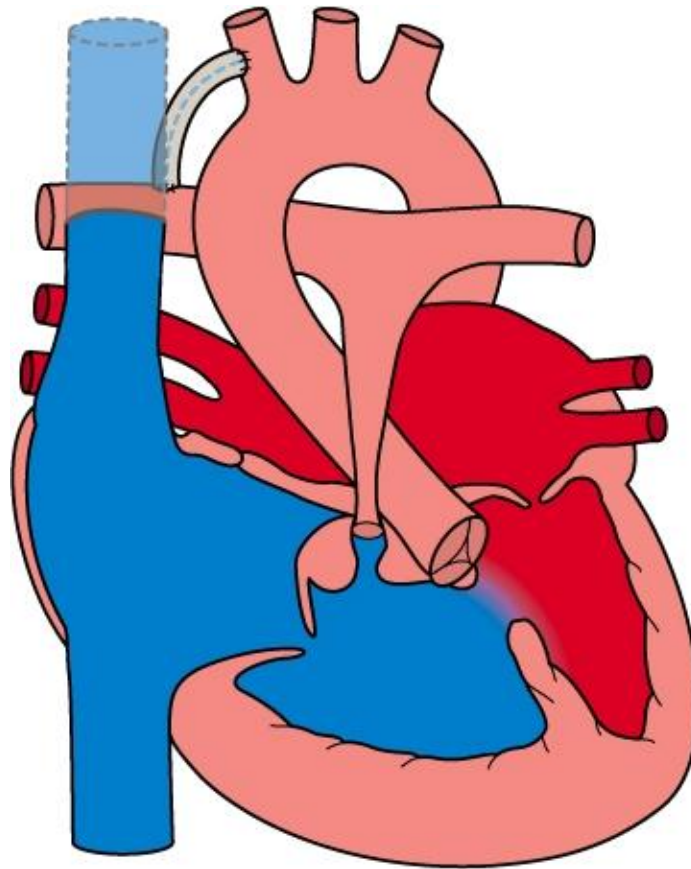
- ♥ 10 % of all CHD
- ♥ Was most common cyanotic defect beyond infancy
- ♥ Cyanosis varies by degree of pulmonary stenosis
- ♥ 4 components:
 - ♥ Large VSD
 - ♥ Right ventricular outflow tract obstruction
 - ♥ Overriding aorta
 - ♥ Right ventricular hypertrophy



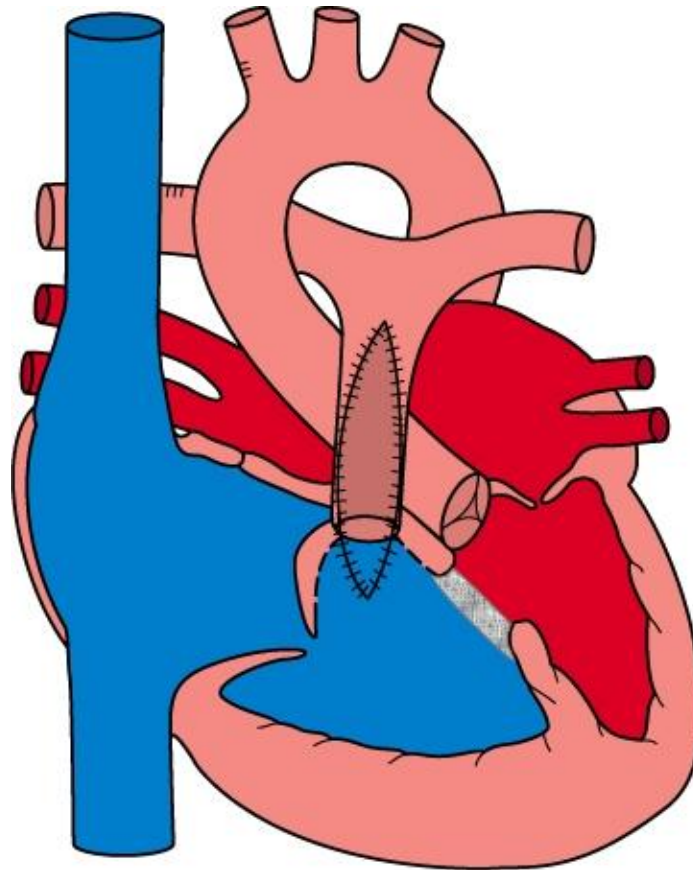
"Tet" Spells

- ♥ Most frequently seen in unrepaired patients with Tetralogy of Fallot.
- ♥ Symptoms:
 - ♥ Rapid and deep respirations (hyperpnea)
 - ♥ Increasing cyanosis
 - ♥ Irritability and prolonged cry
 - ♥ Decreased intensity or absence of heart murmur
- ♥ Treatment:
 - ♥ Pick up and hold infant
 - ♥ Knee chest position
 - ♥ Morphine sulfate: 0.1-0.2 mg/kg IM
 - ♥ 100% oxygen
 - ♥ Phenylephrine 0.02 mg/kg IV bolus followed by infusion
 - ♥ Sodium bicarbonate: 1 mEq/kg IV

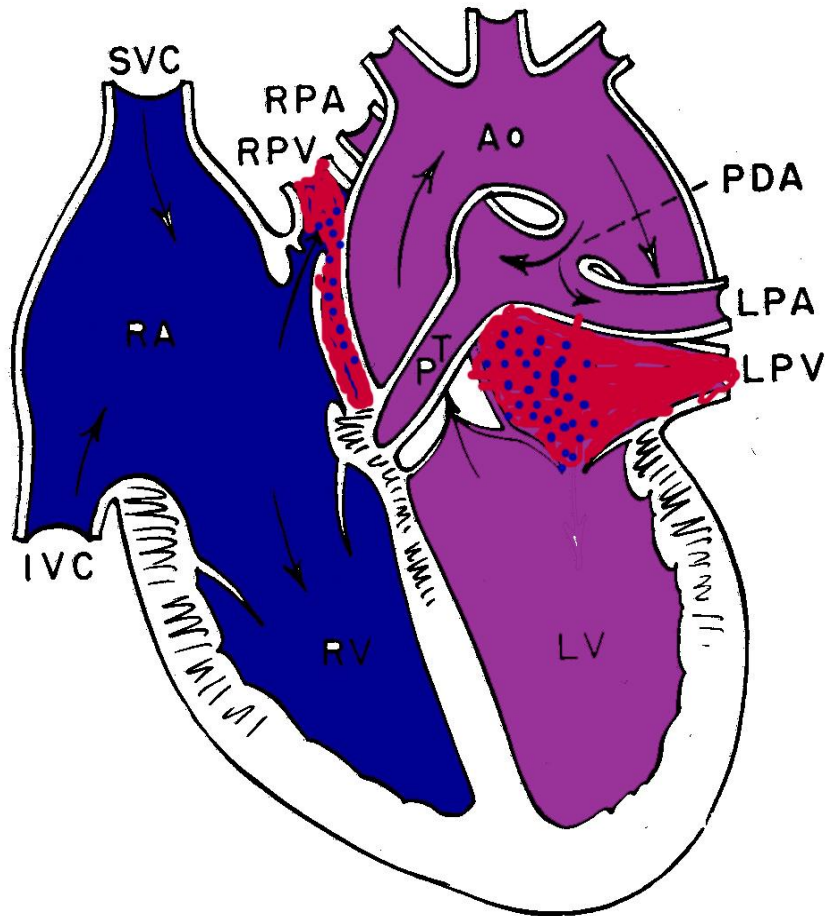
Tetralogy of Fallot with BT Shunt



Repair of Tetralogy of Fallot

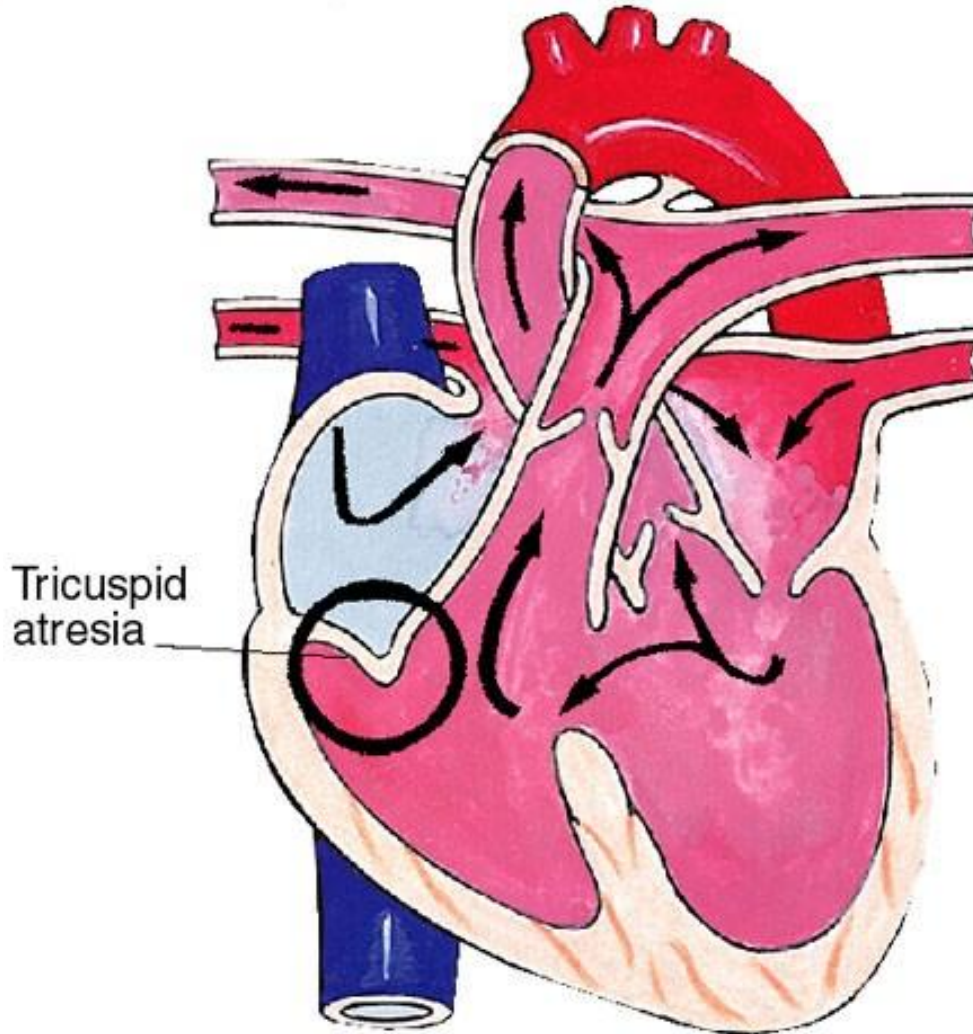


Pulmonary Atresia



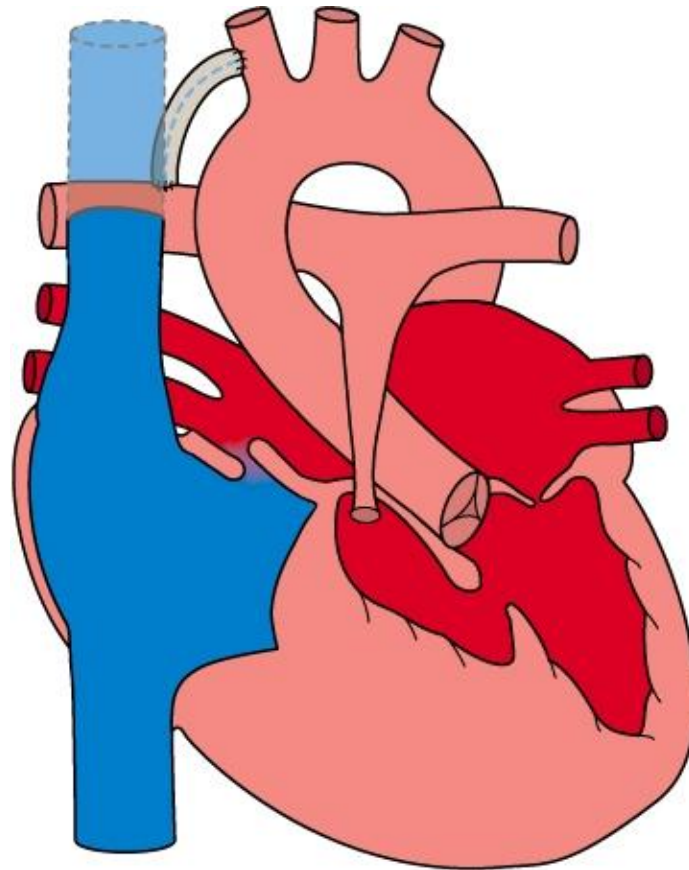
- ♥ With Intact Ventricular Septum 1% of all CHD
- ♥ With VSD 2% of all CHD
- ♥ Is ductal dependent
- ♥ Intervention necessary within first few hours after birth.

Tricuspid Atresia

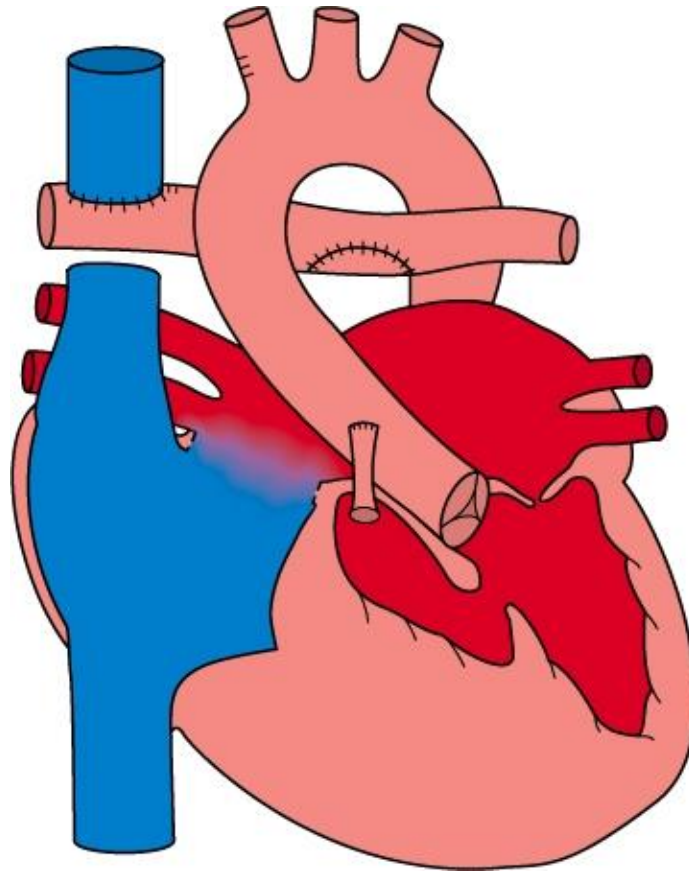


- ♥ 1-2 % of all CHD
- ♥ Male slightly more than female
- ♥ An associated defect is necessary for survival.
- ♥ Systemic saturation is directly related to the amount of pulmonary blood flow.

Tricuspid Atresia with BT Shunt

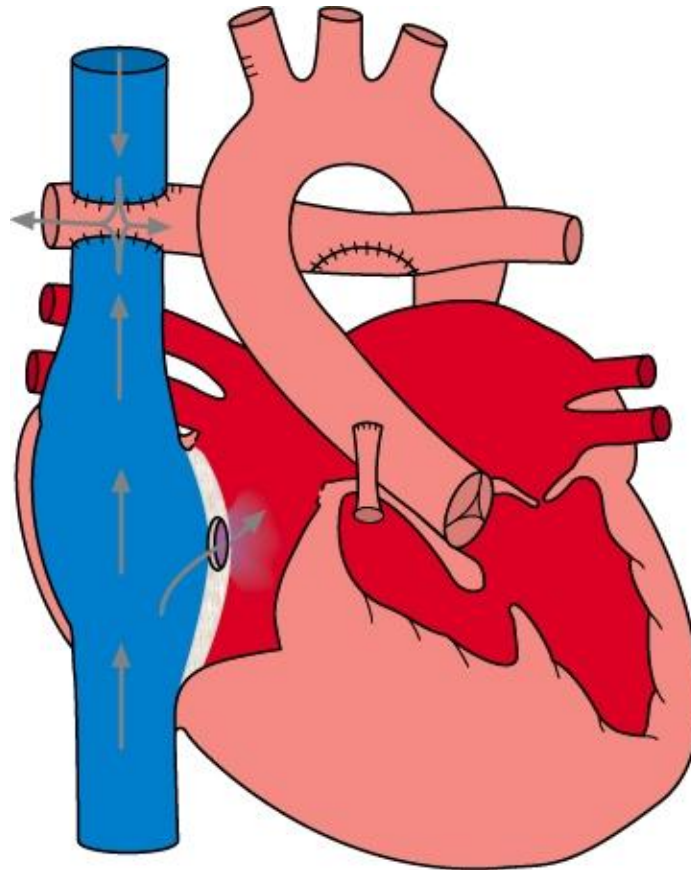


Tricuspid Atresia with Glenn Shunt



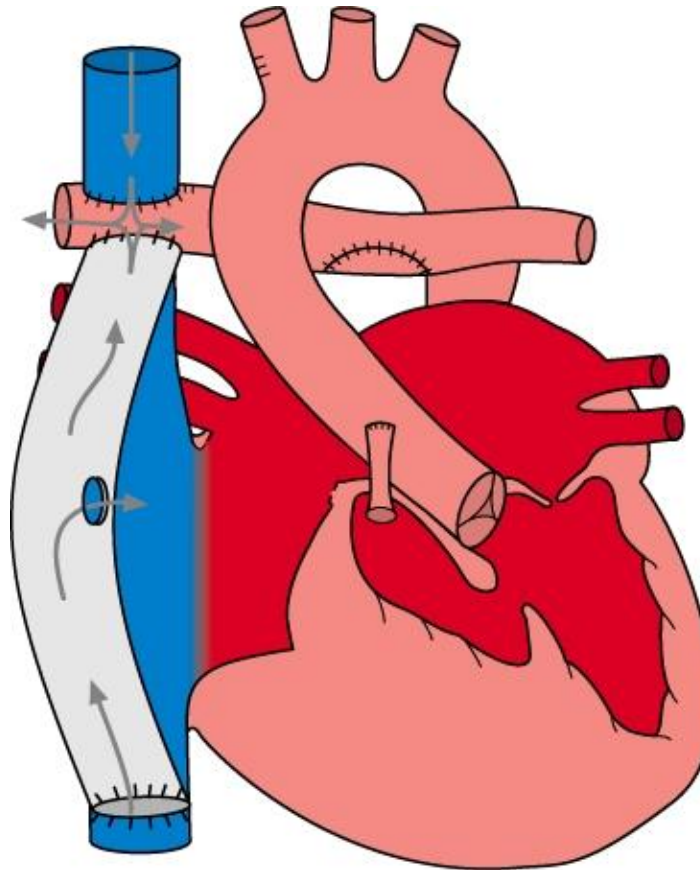
Tricuspid Atresia

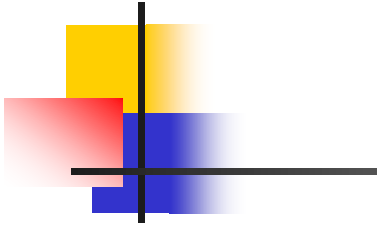
Lateral Tunnel Fontan



Tricuspid Atresia

External Conduit Fontan







Decreased Systemic Perfusion

Clinical Presentation

- Poor feeding
- Pallor/grey
- Diaphoresis
- Tachypnea with respiratory distress
- Irritability

Congenital Defects

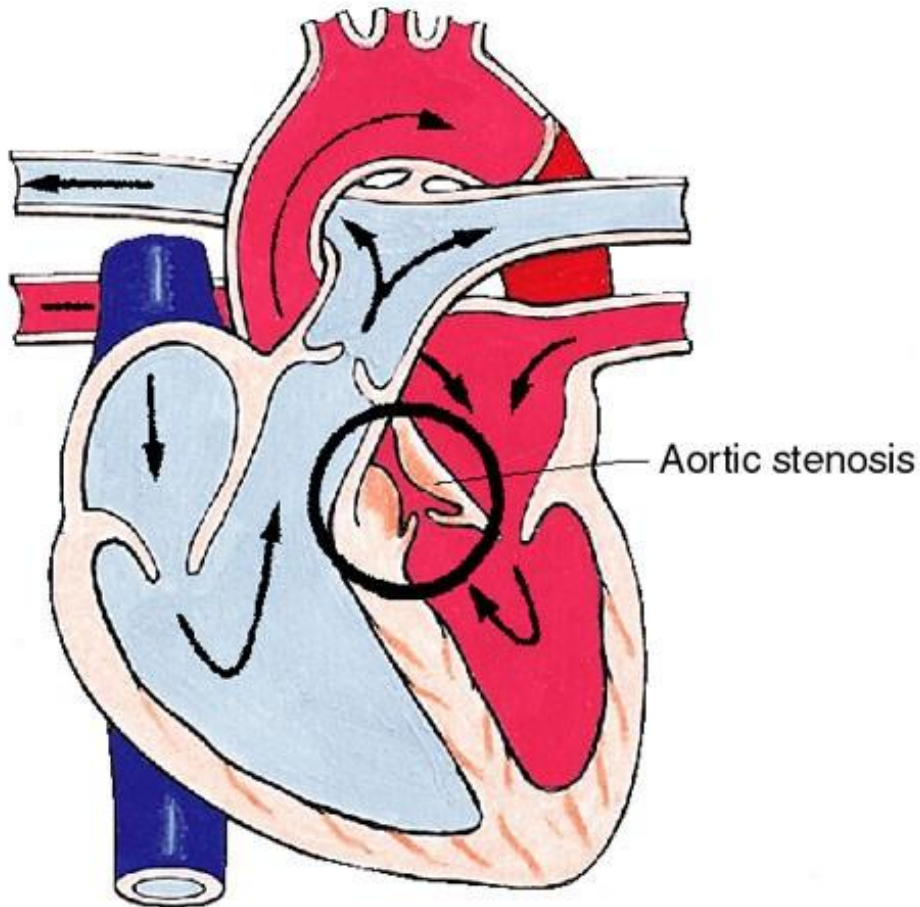
- ♥ Aortic Valve Stenosis
- ♥ Coarctation of the Aorta
- ♥ Hypoplastic Left Heart Syndrome



Nursing Considerations:

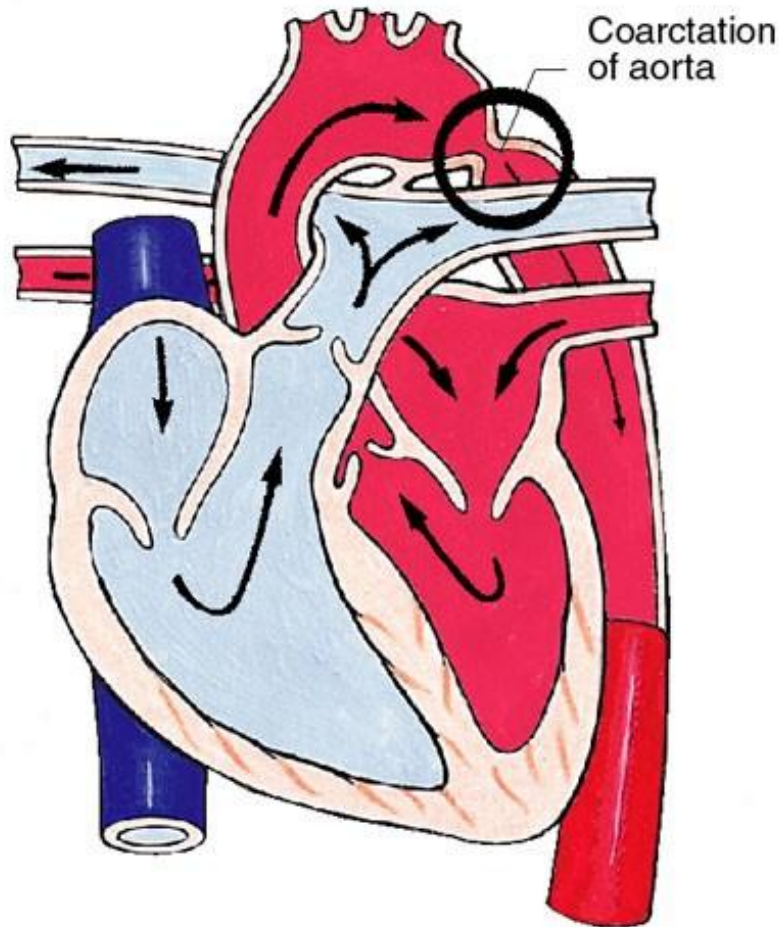
- ♥ Systemic Blood Flow?
- ♥ Intracardiac Mixing of Systemic and Pulmonary Circulations?
- ♥ Assessment of cardiac output
 - ♥ Color
 - ♥ Temperature
 - ♥ Perfusion
 - ♥ Blood Pressure
 - ♥ Heart Rate/rhythm

Aortic Valve Stenosis



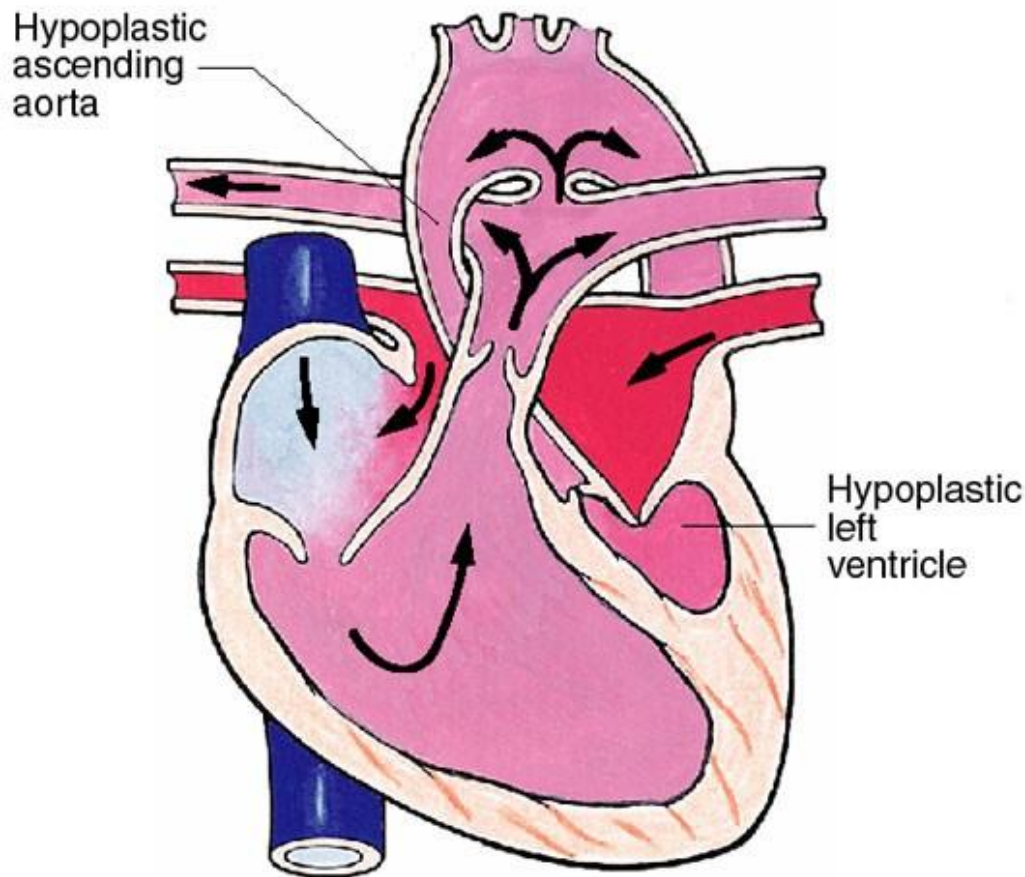
- ♥ 5 % of all CHD
- ♥ Males > female by 4:1
- ♥ Wide spectrum of disease.
- ♥ Babies with severe stenosis may be ductal dependent.
- ♥ Half of patients with Coarctation also have abnormality of aortic valve.
- ♥ Risk of reoccurrence increases by 15 % or greater.

Coarctation of the Aorta



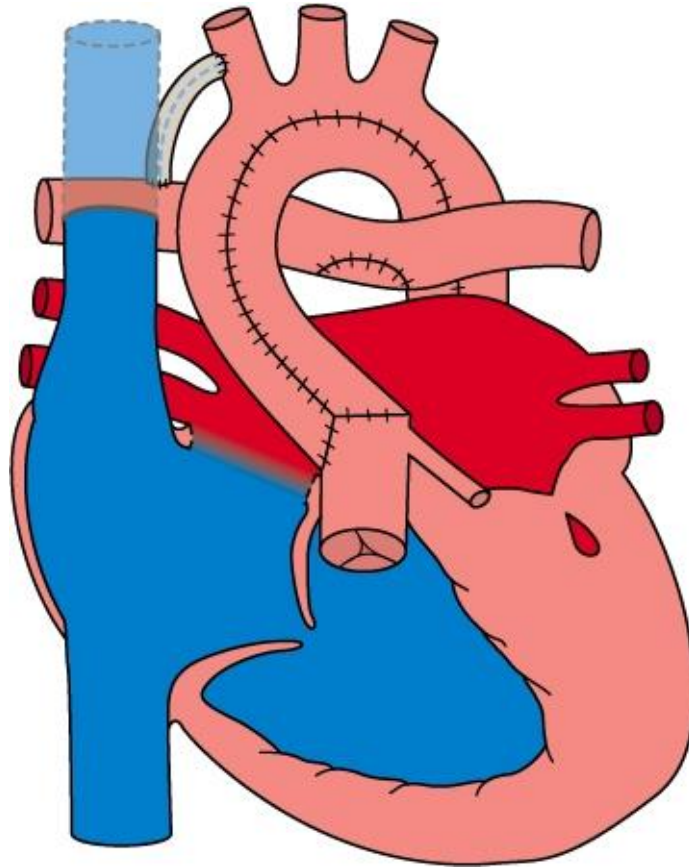
- ♥ 8 % of all CHD
- ♥ Male > female by 2:1
- ♥ 30 % of children with Turners Syndrome
- ♥ May not be evident until ductus arteriosus closes
- ♥ Can present with severe cardiogenic shock

Hypoplastic Left Heart Syndrome

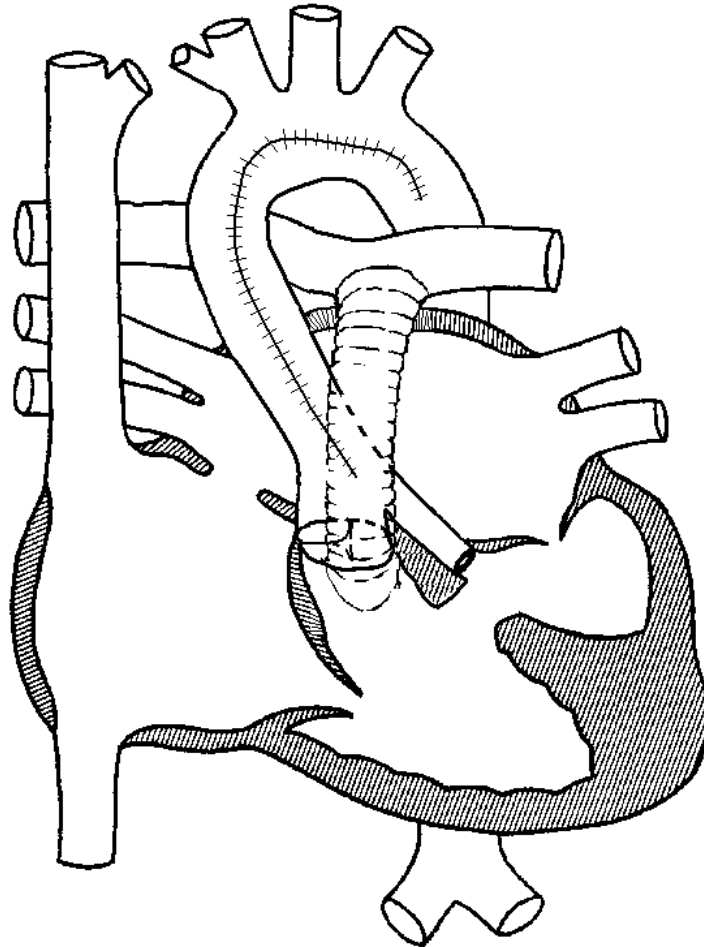


- ♥ 2 % of all CHD
- ♥ 10% of all CHD presenting within first month of life
- ♥ May not be identified until ductus arteriosus closes.
- ♥ Presents in shock.
- ♥ Treatment with oxygen will increase severity by decreasing pulmonary vascular resistance resulting in increase pulmonary blood flow and decreased systemic circulation.

Norwood Procedure for HLHS



Norwood Procedure/Sano



Glenn Shunt

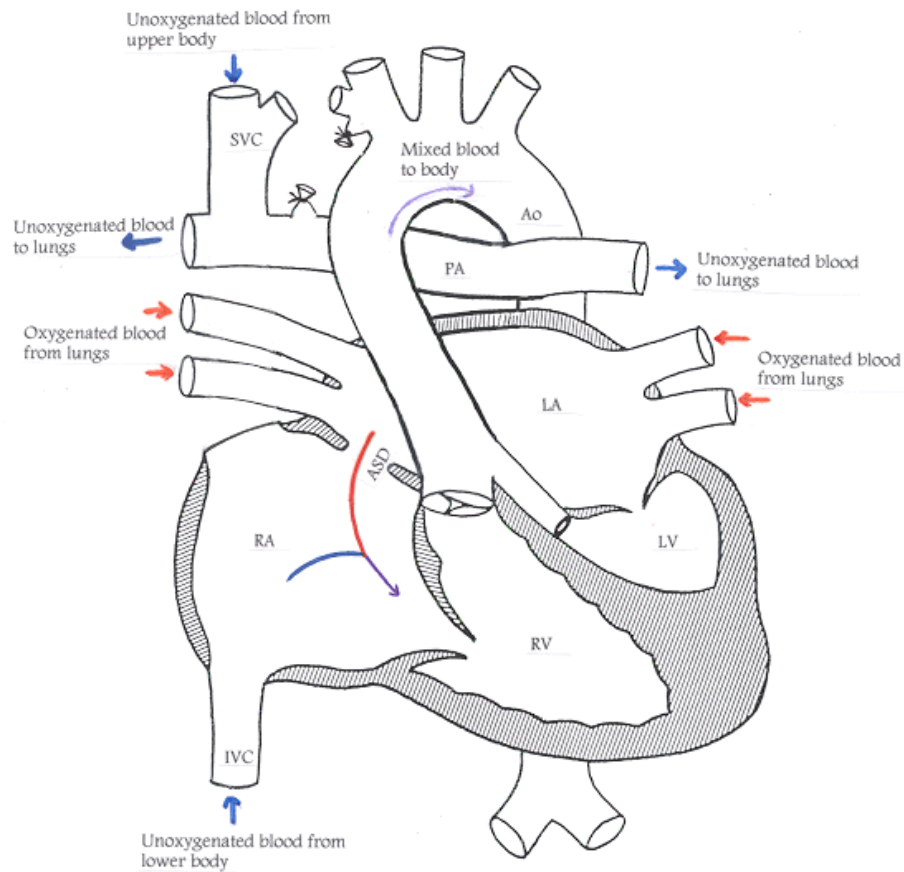
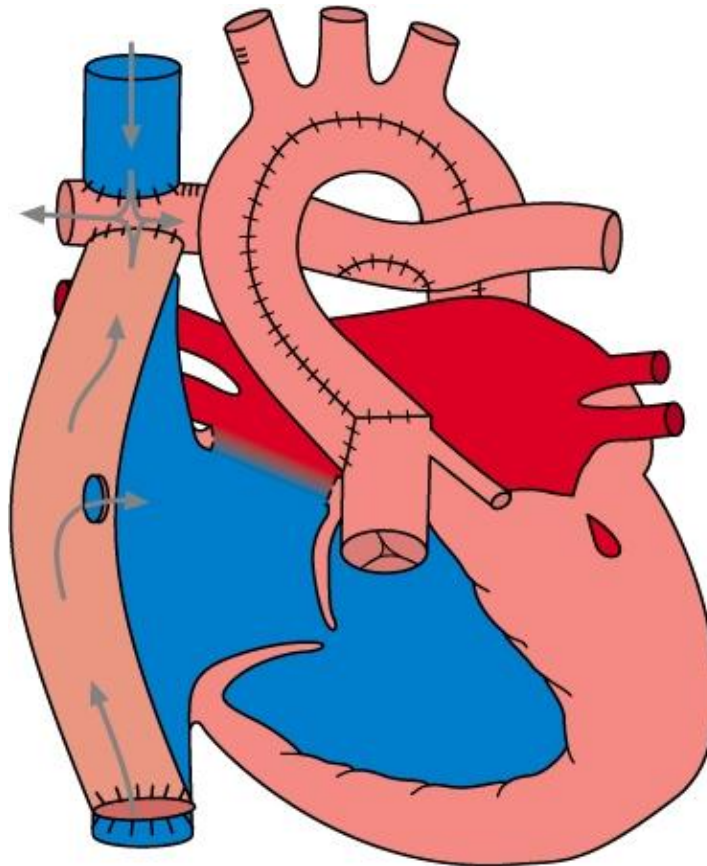


ILLUSTRATION 3 – Hemi-Fontan Procedure

HLHS with External Conduit Fontan





Tachypnea

Clinical Manifestations

- ♥ Tachypnea with mild respiratory distress
- ♥ Hepatomegaly
- ♥ Tachycardia
- ♥ Diaphoresis (secondary to sympathetic response to increased stress)
- ♥ Failure to thrive
 - ♥ Increased oxygen consumption
 - ♥ Increased work of breathing

Congenital ♥ Defects

- ♥ Patent Ductus Arteriosus
- ♥ Atrial Septal Defect
- ♥ Ventricular Septal Defect
- ♥ Atrioventricular Septal Defect
- ♥ Total Anomalous Pulmonary Venous Return (TAPVR)
- ♥ Truncus Arteriosus



Tachypnea:

Excessive Pulmonary Blood Flow

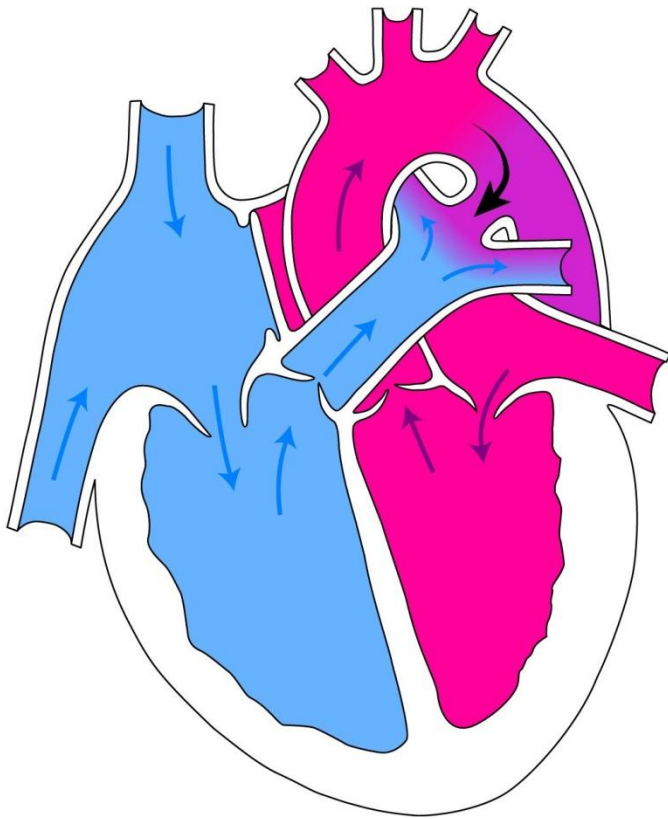
- ♥ Increased pulmonary blood flow results in pulmonary edema and congestive heart failure.
- ♥ Increasing inspired oxygen will decrease pulmonary resistance thereby increasing pulmonary blood flow. CHF symptoms will increase and may even have acute decompensation.
- ♥ Exception is TAPVR below diaphragm. This results in pulmonary venous congestion.



Nursing Considerations:

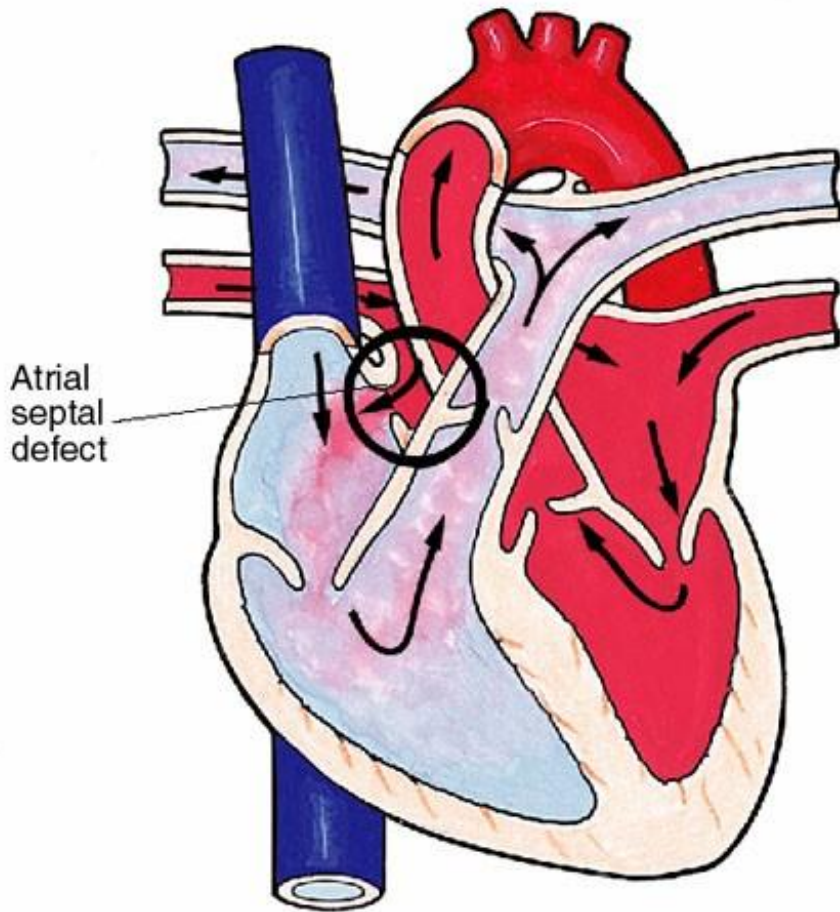
- ♥ Intracardiac Mixing of Systemic and Pulmonic Blood
- ♥ Supplemental Oxygen
- ♥ Assessment of Cardiac Output
- ♥ Failure to Thrive
 - ♥ Increased work of breathing
 - ♥ Increased heart rate
 - ♥ Decreased caloric intake

Patent Ductus Arteriosus



- ♥ 5-10 % of CHD except for preterm infant
- ♥ Female > male by 3:1
- ♥ Elective closure except in preterm infant
- ♥ Non-surgical closure with coils

Atrial Septal Defect



- ♥ 5-10 % of all CHD
- ♥ 4th most common
- ♥ Females > males by 2:1
- ♥ Usually asymptomatic
- ♥ Primary types:
 - ♥ Secundum (site of fossa ovalis)
 - ♥ Sinus venosus (often associated with PAPVR)
 - ♥ Ostium primum (AVSD type defect)

Types of Atrial Septal Defects

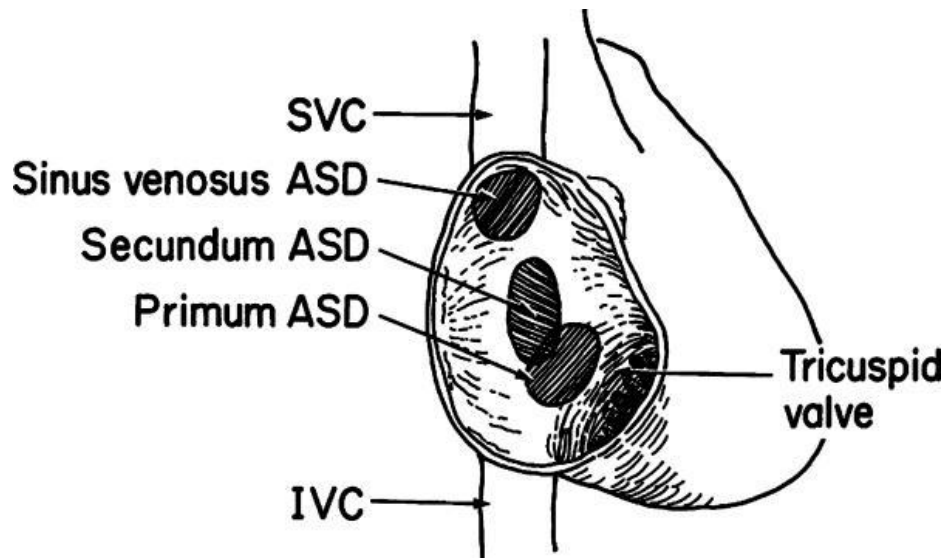


Figure 12-1

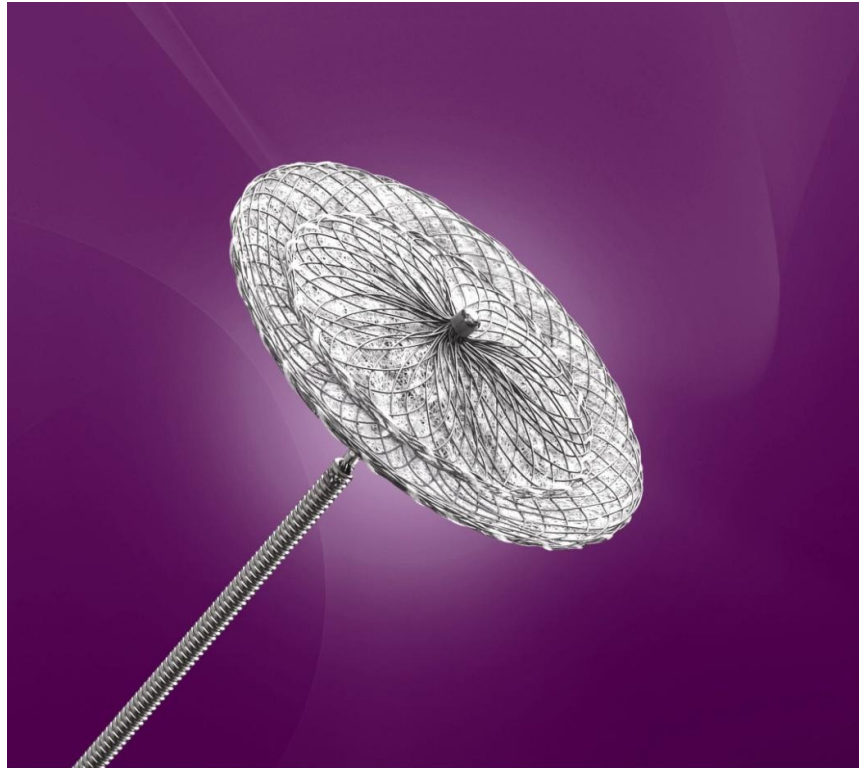
Anatomic types of atrial septal defects (ASDs) viewed with the right atrial wall removed. IVC, inferior vena cava; SVC, superior vena cava.

Park: Pediatric Cardiology for Practitioners, 5th ed.

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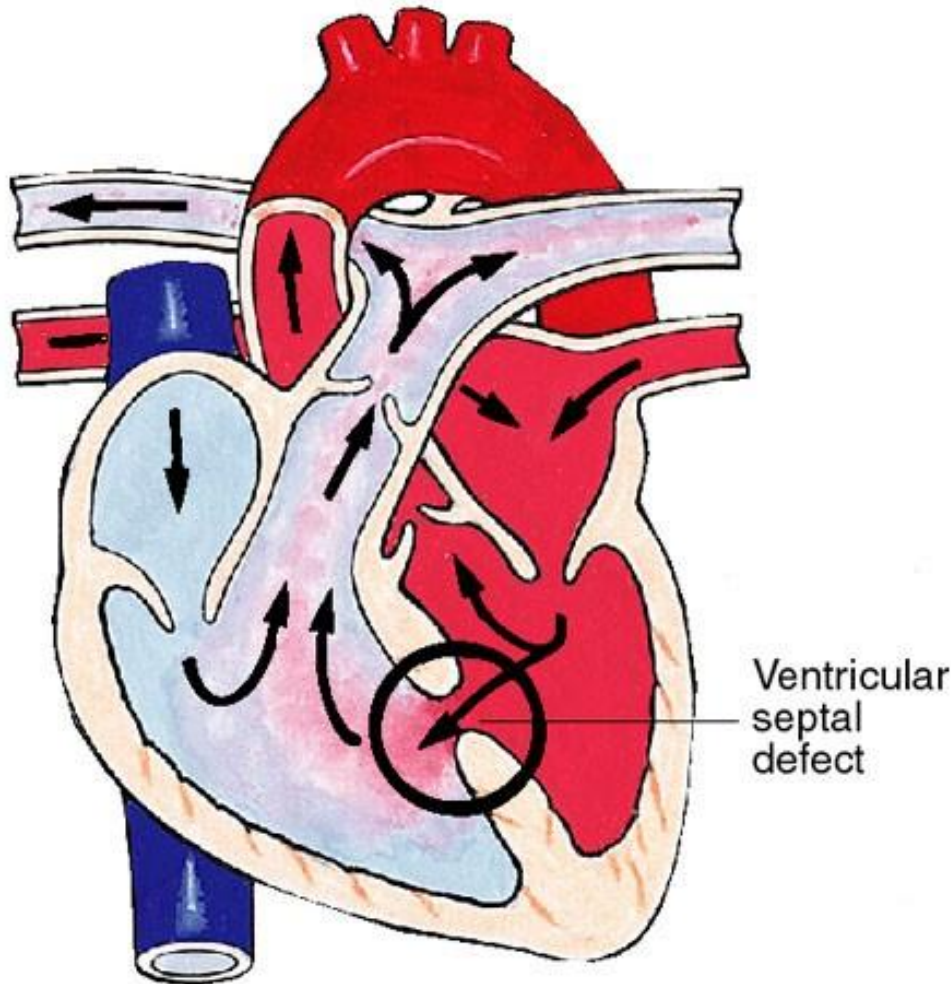
Non surgical closure of ASD



AMPLATZER® PFO Occluder*
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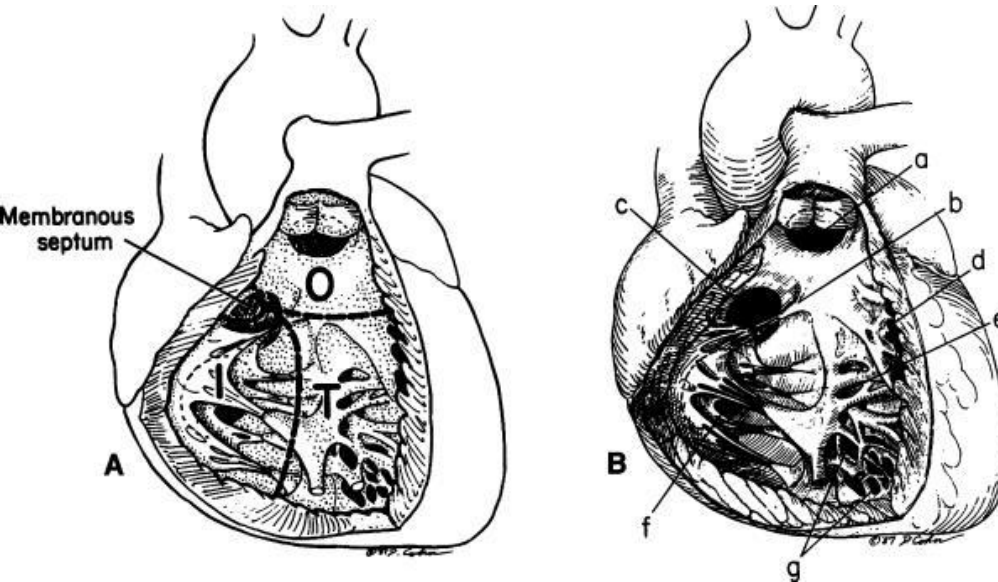
*Not available in all markets. Not for sale in the U.S.

Ventricular Septal Defect



- ♥ 20-25 % of all CHD
- ♥ Most common defect
- ♥ Many will close spontaneously
- ♥ Intervention depends on location and symptoms
- ♥ Increasing use of cath closure devices

Anatomy of Ventricular Septal Defects



- ♥ Figure 12-6 Anatomy of ventricular septum and ventricular septal defect (VSD). A, Ventricular septum viewed from the right ventricular (RV) side. The membranous septum is small. The large muscular septum has three components: the inlet septum (I), the trabecular septum (T), and the outlet (or infundibular) septum (O). B, Anatomic locations of various VSDs and landmarks, viewed with the RV free wall removed. a, outlet (infundibular) defect; b, papillary muscle of the conus; c, perimembranous defect; d, marginal muscular defect; e, central muscular defect; f, inlet defect; g, apical muscular defect. (From Graham TP Jr, Bender HW, Spach MS: Ventricular septal defect. In Adams FH, Emmanouilides GC, Riemenschneider TA (eds): Moss' Heart Disease in Infants, Children and Adolescents, 4th ed. Baltimore, Williams and Wilkins, 1989.)



Types of Ventricular Septal Defects

♥ Membranous

- ♥ Also called perimembranous
- ♥ Most common
- ♥ Located below aortic valve
- ♥ May require surgical repair

♥ Muscular

- ♥ 70 % close spontaneously

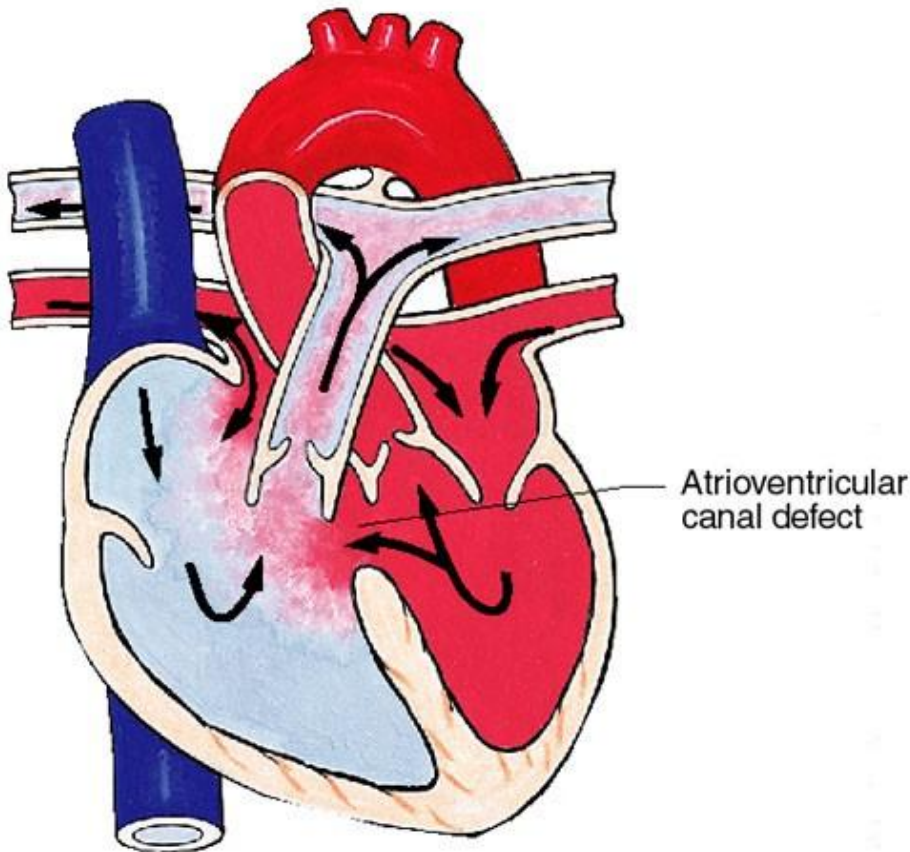
♥ Outlet

- ♥ Located just below pulmonary valve
- ♥ Risk of prolapse of aortic valve leaflet into defect
- ♥ Requires surgical repair, important to protect integrity of aortic valve

♥ Inlet

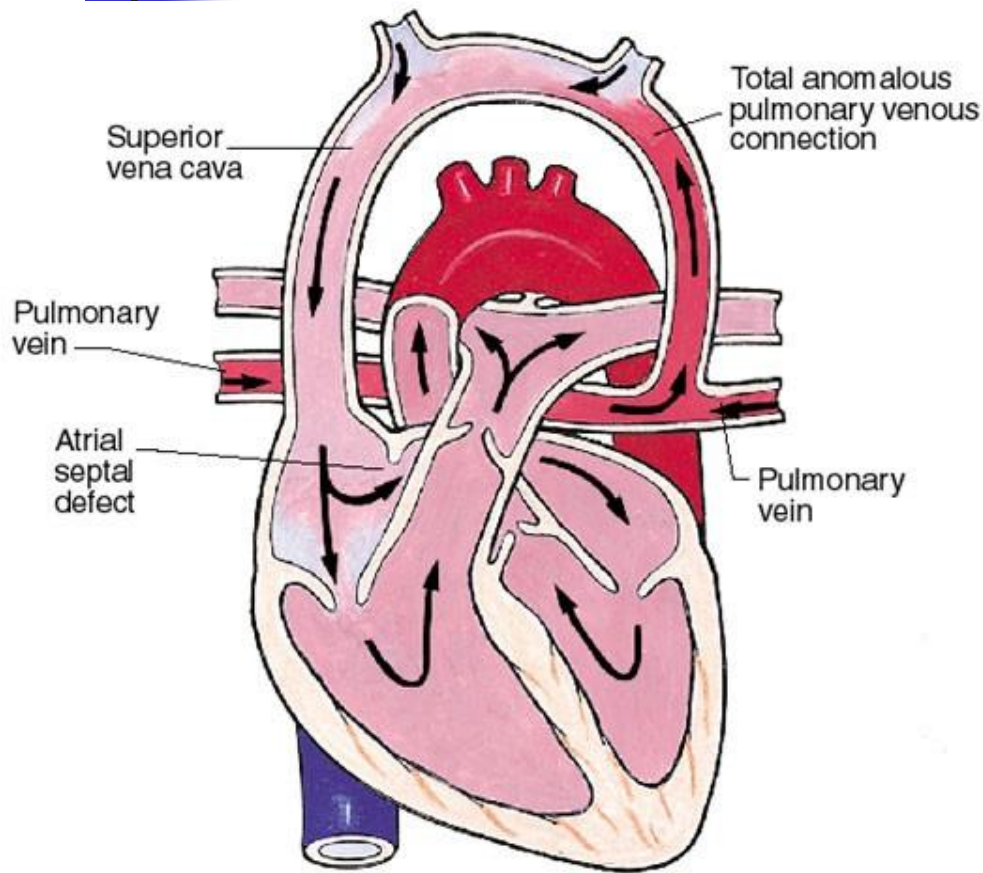
- ♥ Located below the leaflet of the tricuspid valve
- ♥ Will need surgical repair

Atrioventricular Septal Defect (AVSD) (AV Canal Defect, Endocardial Cushion Defect)



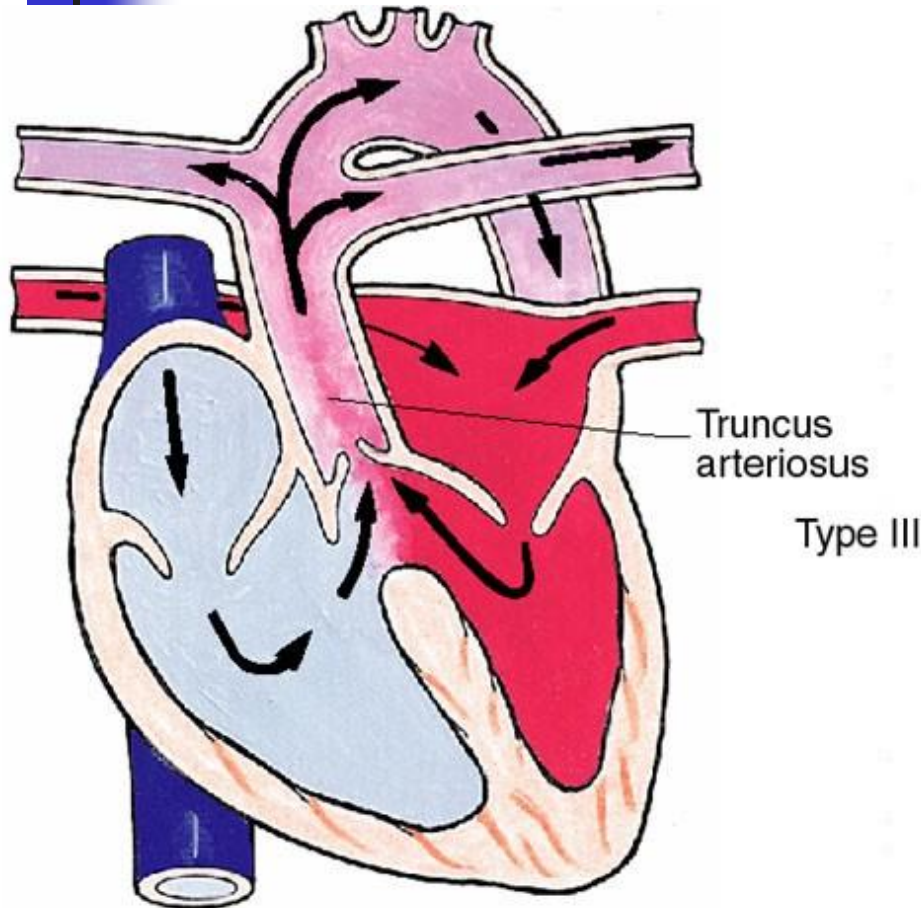
- ♥ 2 - 5 % of all CHD
- ♥ 30-50 % of children with Down's Syndrome
- ♥ Usually surgically repaired by 4- 6 months of age. Earlier if increasing CHF.

Total Anomalous Pulmonary Venous Return



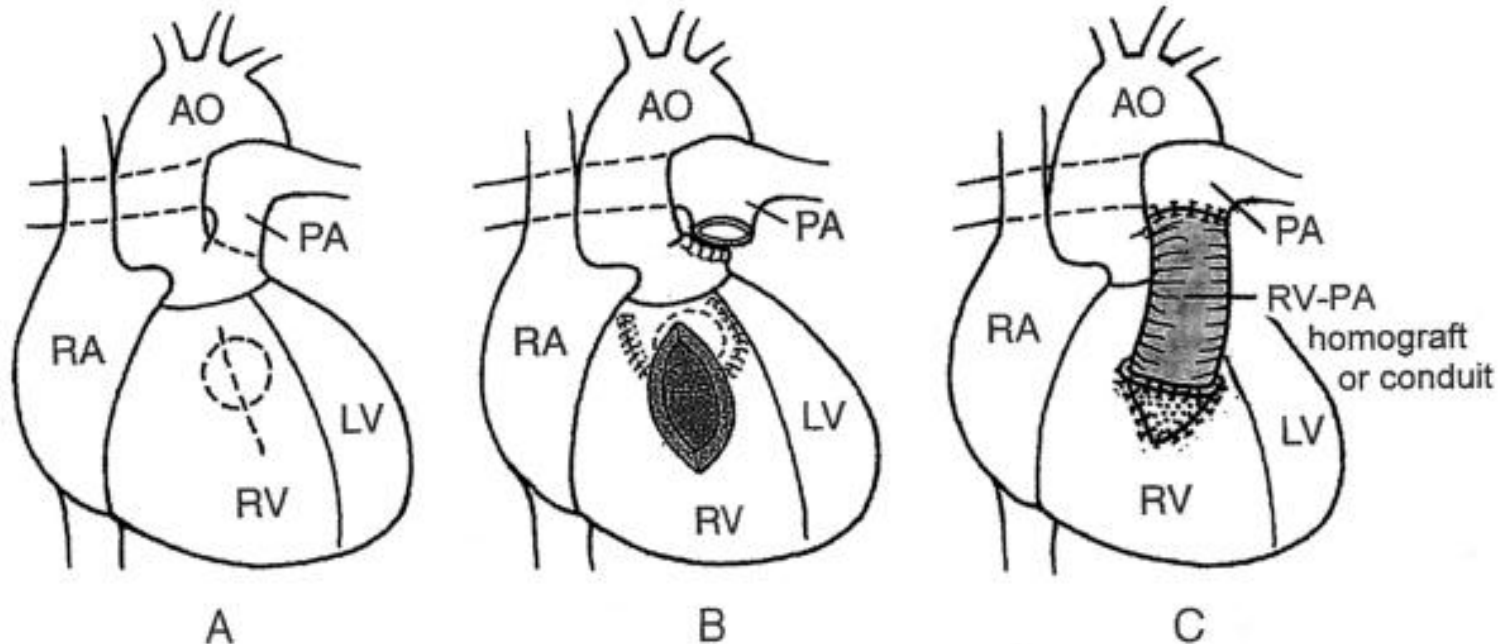
- ♥ Less than 1 % of all CHD
- ♥ No direct connection between PV and LA
- ♥ Must have an atrial communication for survival
- ♥ Types:
 - ♥ Supracardiac (50%)
 - ♥ Male = female
 - ♥ Infracardiac (20%)
 - ♥ Male > female by 4:1
 - ♥ Cardiac (20%)
 - ♥ Usually to coronary sinus
 - ♥ Mixed - Right veins involved twice as often as left

Truncus Arteriosus



- ♥ <1 % of all CHD
- ♥ Single trunk from both ventricles
- ♥ Must have a VSD
- ♥ Truncal valve may have 3 or 4 cusps
- ♥ Usually presents shortly after birth.
- ♥ Surgical repair when diagnosed.

Surgical Repair: Truncus Arteriosus





Ready for Discharge:

- ♥ Newborn Discharge Considerations

- ♥ Hearing Test
- ♥ Car Seat Angle Test
- ♥ Newborn Metabolic Screen

- ♥ Medications

- ♥ Wound Healing

- ♥ Interstage monitoring

- ♥ Oxygen Saturation
- ♥ Weight Gain
- ♥ Activity/fussiness



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