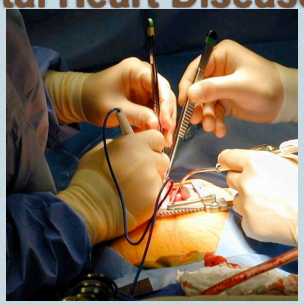
Congenital Heart Disease (CHD)



David Lopez, Ed.D, RCP, RRT

RSTH: 421

NeoNatal Pediatrics

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Objectives

- Describe in writing the major congenital heart defects described in the lecture.
- Describe in writing the possible correction for each congenital heart defect-connect Surgeries to defects.
- Identify the definition and consequence of a "right to left shunt" and a "left to right" shunt.
- Describe the four (4) main hemodynamic consequences of congenital heart defects.
- Describe the Respiratory Care interventions with those patients with congenital heart defects
- Given a case scenario provide the diagnosis and treatment for the major congenital heart defects described in the lecture.

2

Introduction

- In aggregate, congenital heart disease (CHD) comprises a relatively large percentage of all malformations.
- The effect on the patient depends on the severity and type of the malformation, but can range from lethal to clinically insignificant.

3

Introduction

- Prevalence:
  - All births: 1.56 to 7.7/1000
  - Stillbirths: 1 to 34.5/1000
  - Live births: 2.0 to 10.2/1000
  - Affected by mortality, diagnostic criteria, length of follow-up, etc.

4

Introduction

- **Congenital heart disease (CHD) is the most common major birth defect in live births, occurring in approximately 8 per 1,000 live births. CHDs occur in association with other defects, frequently the CNS, gastrointestinal, or genitourinary system, approximately 30 percent of the time. The etiology of these defects may be chromosomal, teratogenic, multifactorial, or Mendelian.**

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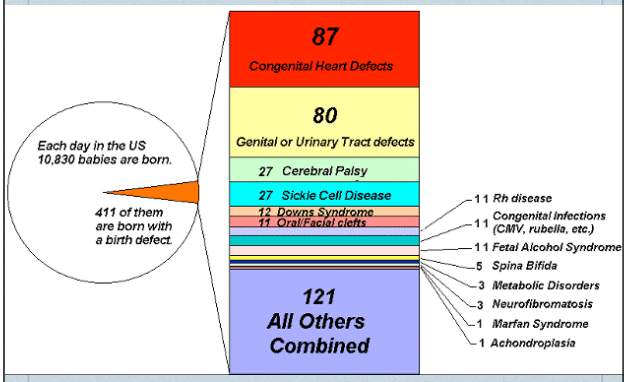

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Introduction-Causes

1. Genetic:

   Trisomies (21, 18 and 13) and Turner syndrome (monosomy X) are strongly associated with CHD.

   45% with CHD have other developmental abnormalities.

   When one child in a family is affected with CHD, the relative risk for siblings is empirically increased (incidence in future children 2-6% rather than the 0.2-1% underlying incidence)

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Introduction-Causes

2. Environmental:

   Rubella, maternal phenylketonuria, diabetes, thalidomide, isotretinoin (retinoic acid) and others

3. Idiopathic:

   Over 90% not clearly genetic or environmental. Multifactorial genetic and environmental factors?

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Congenital Cardiovascular Defects Statistics

- 4,310 Americans died from cardiovascular defects in 2009.

http://www.americanheart.org/presenter.jhtml?identifier=4576 8

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Congenital Cardiovascular

- About 40,000 babies are born each year with cardiovascular defects. Of these,
  - 4-10 percent have atrioventricular septal defect.
  - 8-11 percent have coarctation of the aorta.
  - 9-12 percent have tetralogy of Fallot.
  - 10-11 percent have transposition of the great arteries.
  - 14-17 percent have ventricular septal defects.

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Congenital Cardiovascular Defects Statistics

- About 1,000,000 Americans with cardiovascular defects are alive today.
- 2000 death rates per 100,000 people for congenital cardiovascular defects were 1.7 for white males, 2.1 for black males, 1.4 for white females and 1.8 for black females.
- Crude infant death rates (under 1 year) are 45.7 per 100,000 white babies and 62.8 per 100,000 black babies.
- From 1990 to 2000 death rates for congenital cardiovascular defects declined **27.1** percent, while the actual number of deaths declined 26.1 percent.

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Toddler development (1 -3 years)

Physical:
- able to stand alone well by 12 months
- can stoop over, pick up something, and recover to standing
- usually walking well by 12 to 14 months
- once walking well, learns to walk backwards and up steps
- can kick ball; throw ball overhand (15 - 18 months)
- may jump in place; pedal a tricycle (20 - 24 months)
- demonstrates good grasp with thumb opposing fingers
- able to pick up small object with pincer grasp (13 months)
- scribbles spontaneously at 12 to 15 months
- can dump items from a bottle at 12 to 16 months
- able to stack 2 cubes (12 - 17 months); 4 cubes (15 - 20 months); 8 cubes (20 - 26 months)
- copy a vertical line (15 - 20 months); circle (18 - 24 months)

Language:
- uses 3 words (other than Mama or Dada) at 12 to 15 months
- combines 2 words at 14 to 20 months
- able to point to named body parts at 14 to 20 months
- able to name pictures of items, animals at 16 to 22 months
- begins to use plurals at about 20 months
- may be able to state first and last name (22 - 24 months)
- may develop 5 to 8 word sentences
- begins to put series of events into chronological order
- may explore story telling

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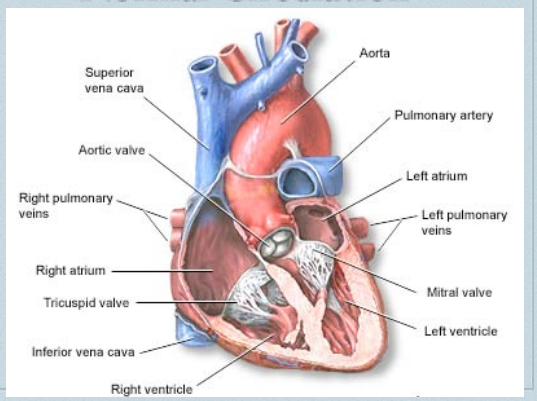
Normal Circulation



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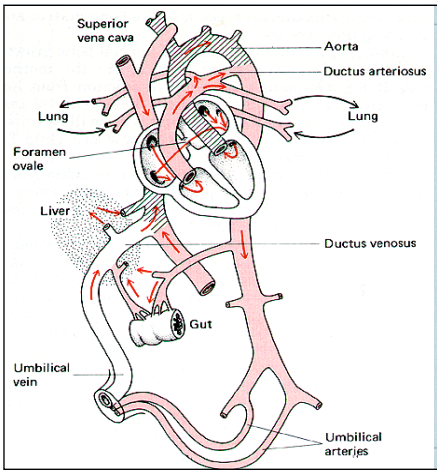

Normal Fetal Circulation

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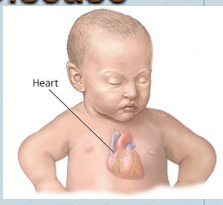
Congenital Heart Disease

- Patent ductus arterious PDA
- Atrial septal defect ASD
- Ventricular septal defect VSD
- Coartation of the aorta
- Tetrology of Fallot
- Transposition of the great vessels
- Truncus arteriousus
- Hyoplastic Left Heart Syndrome
- Total anomalous pulmonary venous return TAPVR(D)



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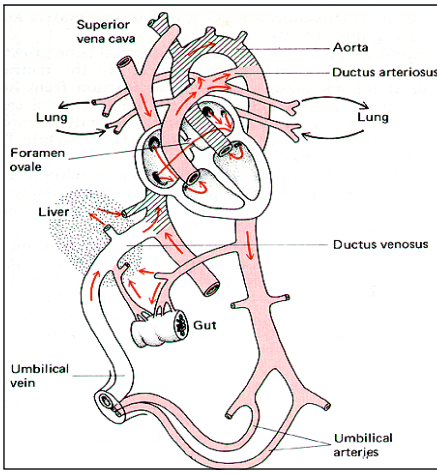

Normal fetal circulation

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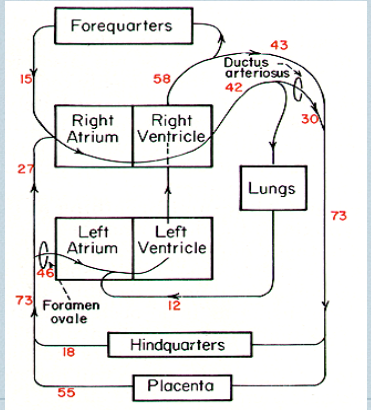
% Fetal Cardiac output



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4 Hemodynamic Consequences

- Volume overload: increased ventricular volume load resulting in an increased workload on the ventricle, hypertrophy to failure.
- Pressure overload: Obstruction to ventricular outflow resulting in an increased workload on the ventricle, hypertrophy to failure.
- Desaturation, R to L shunt: hypoxia, acidosis, decrease myocardial work/effectiveness.
- Decreased cardiac output: combined effect or primary.

Single or combined effect

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Left-to-right shunts

- The direction of the shunt is defined by its presentation, not by the terminal features.
- Do NOT have cyanosis as an early feature. Cyanosis may develop if pulmonary hypertension causes reversal of flow late in the course of the disease.
- L-to-R: The most common group of disorders with the single most common malformation (VSD).
- Pulmonary congestion
- Increased left ventricular work, due to increased volume.
- Failure to thrive or gain weight.

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Left-to-right shunts-ASD

- There are three classical types of ASDs, but other septal defects may occur. The naming of ASDs is related to an understanding of the development of the septum
- Secundum defects are the most common
  - Occur in the region of the foramen ovale
  - Usually asymptomatic initially

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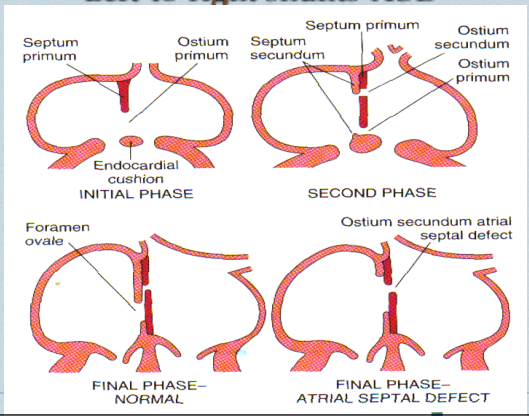
Left-to-right shunts-ASD



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Left-to-right shunts-ASD

- Primum defects occur inferiorly and often have associated VSDs (endocardial cushion defects)
  - Often have heart failure due to mitral valve abnormality
- Sinus venous defects are the least common
  - Associated with anomalous pulmonary venous drainage.

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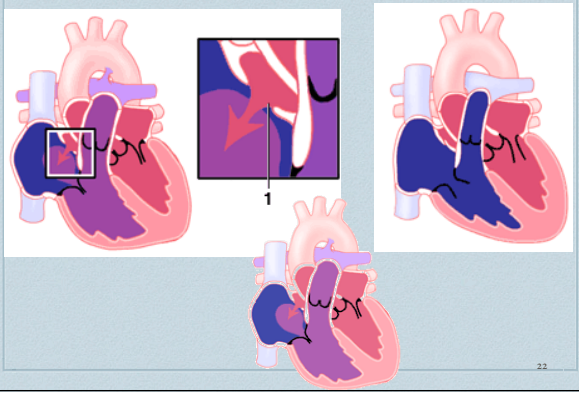
Atrial septal defect ASD



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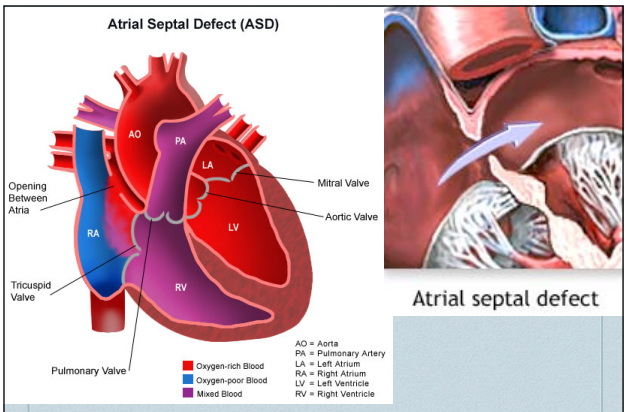

Atrial septal defect

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Left-to-right shunts-VSD

- The intraventricular muscular ridge grows from the apex toward the endocardial cushions. The membranous portion is the last to form and is the location for most VSDs.
- VSDs are the MOST COMMON congenital defect and occur in isolation less than one-third of the time.

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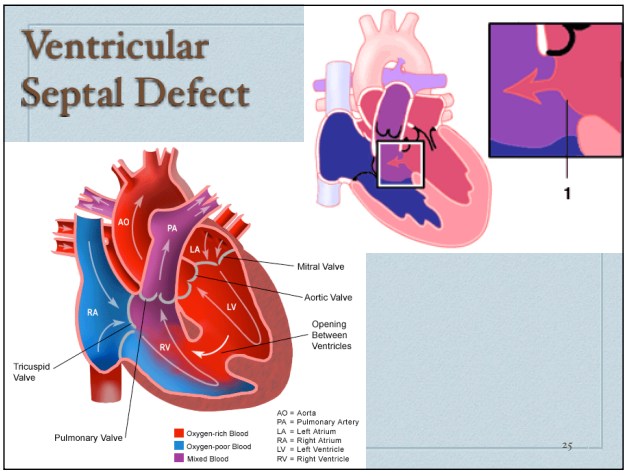
Ventricular Septal Defect



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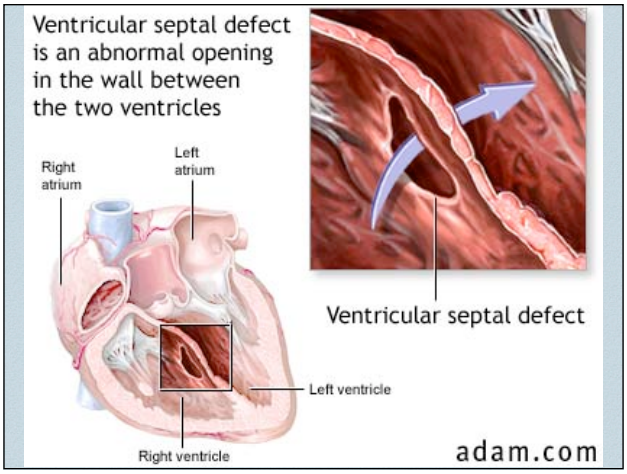

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Left-to-right shunts-VSD

- Have associated right ventricular hypertrophy if a significant L-to-R shunt.
- Small VSDs may close spontaneously, especially if in the muscular wall.
- If large or if they do not close then pulmonary hypertension develops, reversal of flow (R-to-L) and cyanosis.
- Infective endocarditis-risk in all congenital

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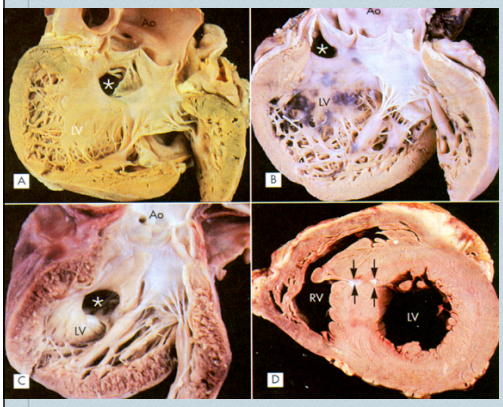

VSDs

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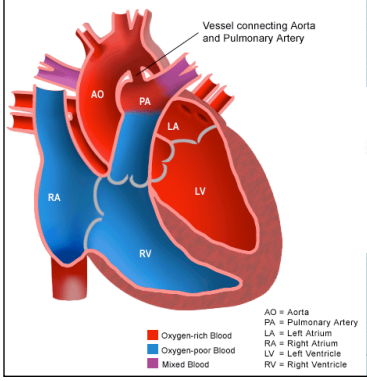
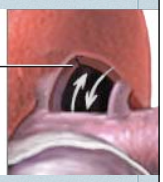
Patent Ductus Arterious PDA

Patent Ductus Arteriosus (PDA)



Patent ductus arteriosus



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Left-to-right shunts-PDA

- Ductus arteriosus
  - In-utero connects the pulmonary artery to the aorta to allow for normal fetal circulation.
  - Closes in response to elevated oxygen tension, decreasing vascular resistance, decreasing prostaglandin E2.
  - Normally closes (functionally) 1-2 days after birth in term infants.

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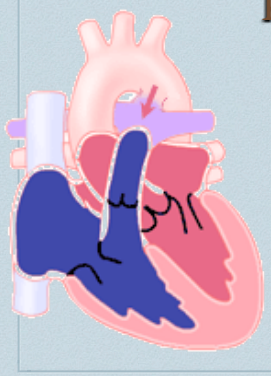
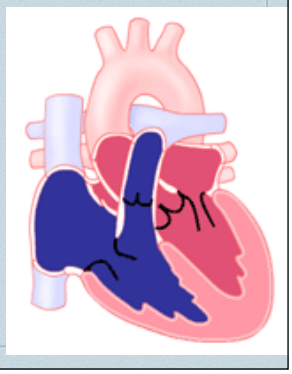
PDA





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Left-to-right shunts-PDA

- PDA
  - Ductal closure may be delayed with hypoxia, prematurity, or cardiac disease.
  - May be isolated or with other CHD (VSD).
- Causes L-to-R volume and pressure overload,
  - May see left atrial and ventricular dilation and hypertrophy.
- Clinically: Holosystolic "machinery murmur"

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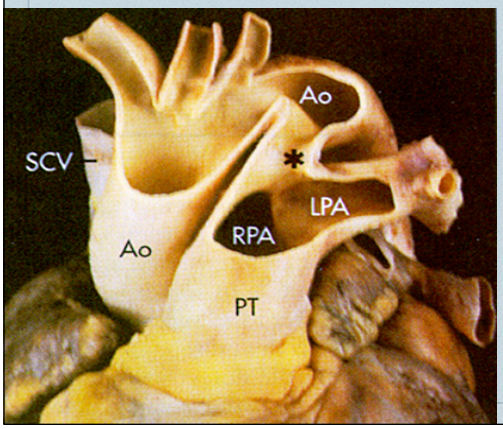

Patent ductus arteriosus

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Right-to-left shunts

- Cyanotic disorders
- Severe hypoxemia

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Right-to-left shunts
Tetralogy of Fallot

- R-to-L shunts: CYANOSIS at or near the time of birth (or diagnosis if tetralogy of Fallot).
- Tetralogy is the most common cause of cyanotic CHD, causing ~10% of all CHD.
- Four key components: VSD, overriding aorta, right ventricular outflow tract obstruction and right ventricular hypertrophy
  - Minority may have an ASD or a PDA

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Tetralogy of Fallot

Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.

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Tetrology of Fallot

1. **pulmonary stenosis** (narrowing of the pulmonary valve and outflow tract creating an obstruction of blood flow from the right ventricle to the pulmonary artery);
2. **ventricular septal defect** or VSD;
3. **overriding aorta** (the aortic valve is enlarged and appears to arise from both the left and right ventricles instead of the left ventricle as occurs in normal hearts);

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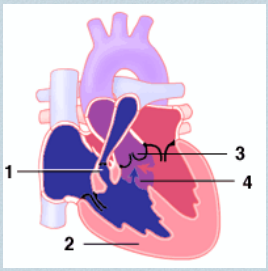
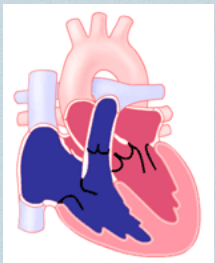
Tet. Of Fallot





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Right-to-left shunts
Tetralogy of Fallot

- VSD-Membranous location-malaligned
- Overriding aorta. The aorta is shifted to the right (dextroposition)
- Right ventricular outflow tract obstruction
  - Most severe form is pulmonary valve atresia. Pulmonary artery is relatively hypoplastic.

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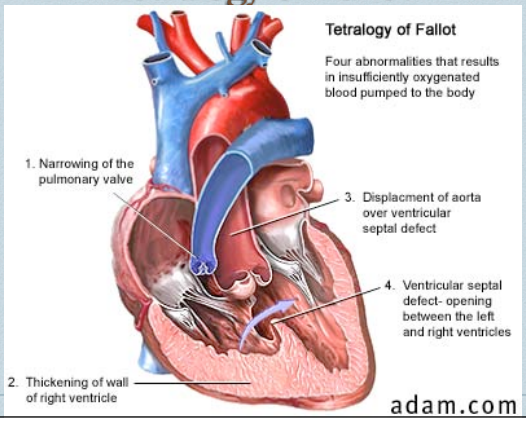
Tetralogy of Fallot



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Right-to-left shunts
Tetralogy of Fallot

- Degree of the right to left shunt is related to the degree of right ventricular outflow obstruction.
  - Outflow tract stenosis becomes worse with growth.
- Pulmonary hypertension is not a problem, but infective endocarditis and "paradoxical emboli" (including brain abscesses) are potential complications.

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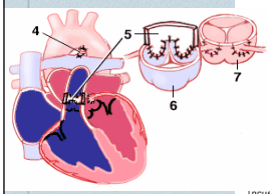
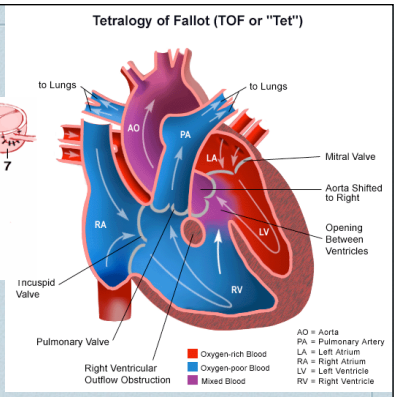
Tet. Repair





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Right-to-left shunts
Transposition of the great vessels

- Abnormal development of the truncus arteriosus.
- Aorta arises from the right ventricle and the pulmonary artery arises from the left.
- Incompatible with life without a connection. In-utero that connection is the foramen ovale and the ductus arteriosus. Ex-utero an ASD, VSD or PDA is required.

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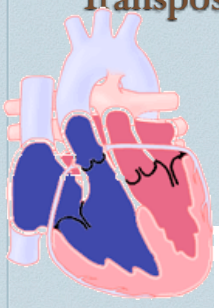
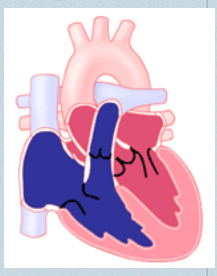
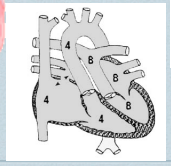
Transposition of the great vessels







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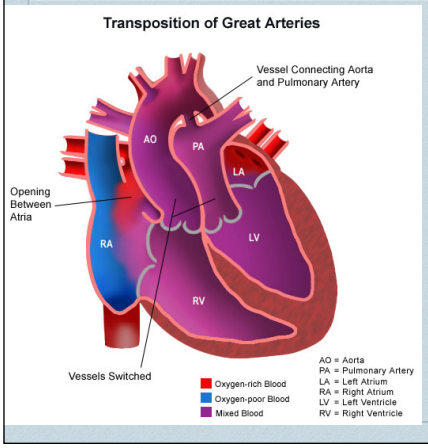

Complete transposition of the great vessels w/out VSD

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Congenital obstructive lesions Coarctation of the aorta

- Other congenital obstructive lesions (valvular stenosis for example) occur.
- Coarctation is isolated (no other CHD) in half of cases
- Is more common in males than females
- Is increased in frequency in patients with Turner syndrome

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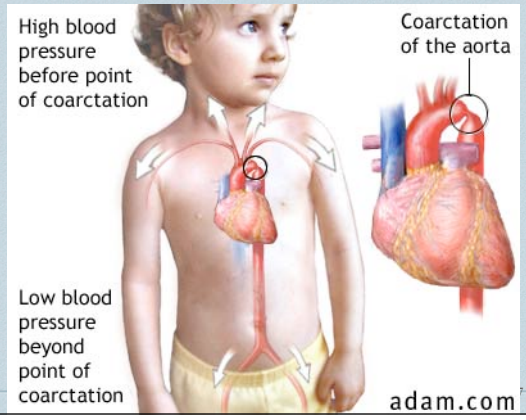
Coarctation of the aorta



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Congenital obstructive lesions
Coarctation of the aorta

- Preductal-narrowing of the aortic isthmus between the left subclavian artery and the entry of the ductus arteriosus.
  - Can be focal or diffuse with hypoplasia of the aortic arch.
  - Usually seen in infants with congestive heart failure, decreased femoral pulses.
  - Require correction early in life.

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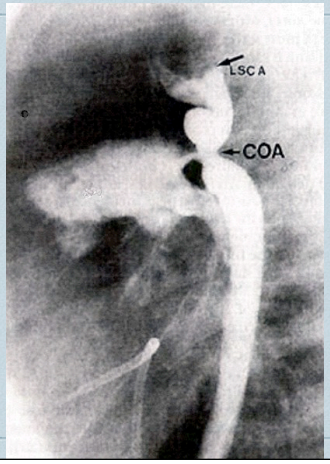

Preductal Coarctation of the aorta

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Congenital obstructive lesions
Coarctation of the aorta

- Postductal-More common than preductal.
  - Sharp constriction distal to the ductus arteriosus.
  - Proximal dilation of aorta.
  - Collateral flow through other arteries.
- More common in older children and adults. Have upper extremity hypertension.

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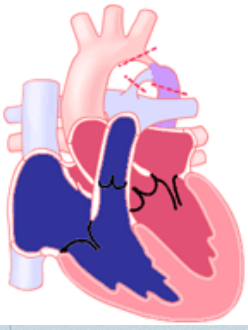
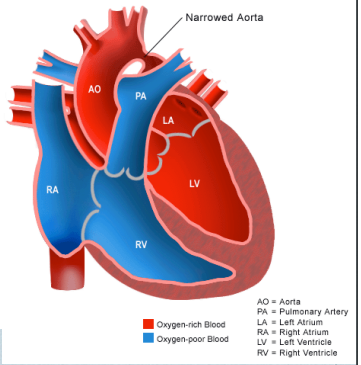
Coart. Repair





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Clubbing

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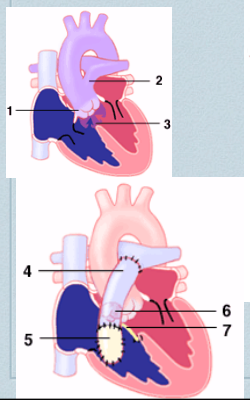
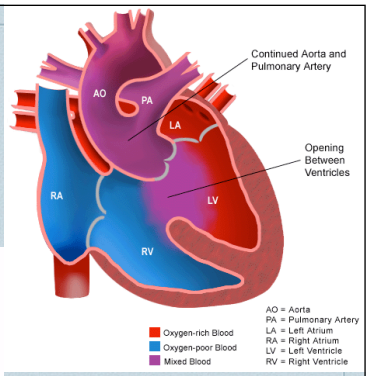
Truncus





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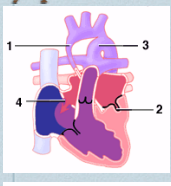
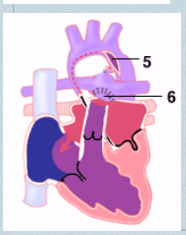
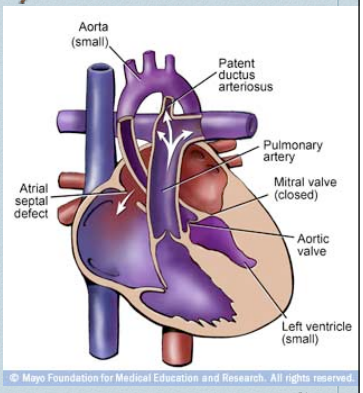
Hyoplastic Left Heart Syndrome







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Hyoplastic Left Heart Syndrome

- Hypoplastic left heart syndrome (HLHS), also known as aortic atresia, is characterized by hypoplastic left ventricle with aortic and mitral atresia. Diagnosis is made by diminished left ventricle size in comparison to right ventricle size. Also, aortic atresia and hypoplasia of the ascending aorta may be visualized. The prognosis for this condition is poor without transplant. The etiology of HLHS is not known, but the recurrence risk is approximately 4 percent after the birth of one affected infant.

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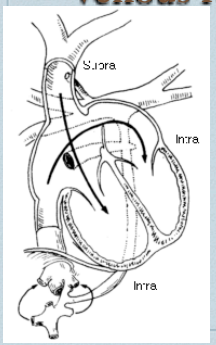
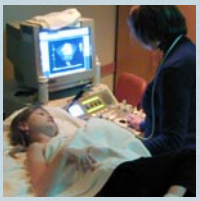
Total anomalous pulmonary venous return TAPVR(D)





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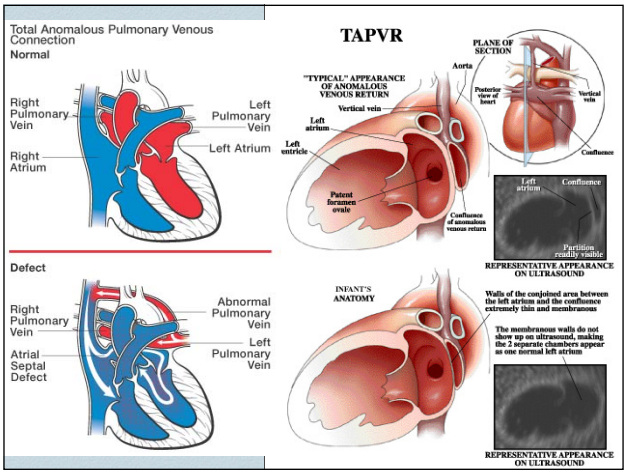
57



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TAPDR

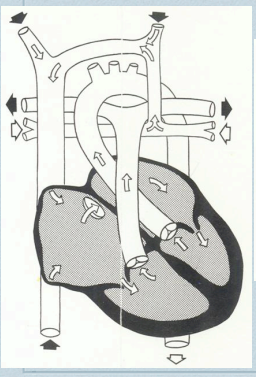

Fig 9. Total anomalous pulmonary venous connection.
All the veins drain into the right atrium, either directly or by way of the systemic veins. An interatrial communication is necessary to allow blood to reach the left side of the heart and from there the systemic circulation. The admixture of blood in the right atrium may flow in one of two pathways: through the atrial communication into the left atrium and the left ventricle, or into the right ventricle. The direction and magnitude of blood flow from the right atrium depend on the relative compliances (distensibilities) of the ventricles.

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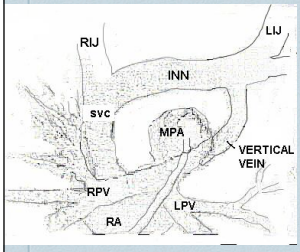

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- The preceding is an angiogram in the AP projection with the catheter positioned in the main pulmonary artery. The anomalous pulmonary veins join in a confluence, do not enter the left atrium in a normal fashion but instead travel upward in a vertical vein to join the innominate vein. The pulmonary veinous return then flows back into the right atrium**(RA)** in a continuous circle. The only blood entering the left ventricle is that which shunts across the patent foramen ovale. A small amount of contrast is seen refluxing into the proximal right jugular vein **(RIJ)**. Similarly a small washout of contrast is seen where the vertical vein joins the innominate **(INN)**.

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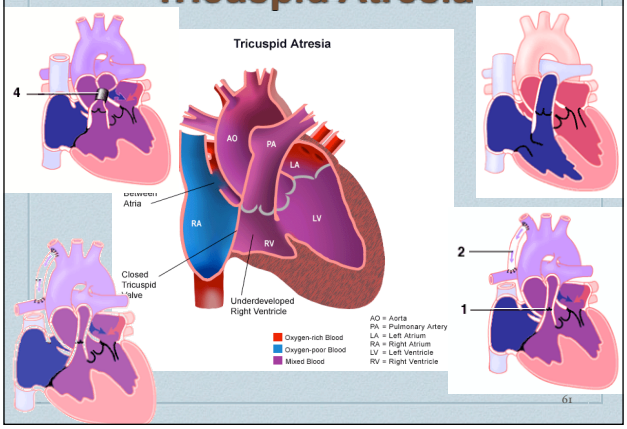
Tricuspid Atresia



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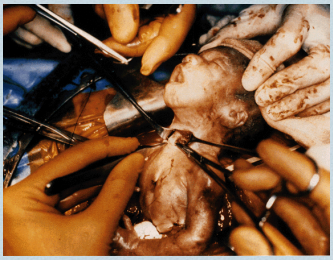
Surgical Repairs



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Surgeries-assignment

Glenn (classic)

Fontan

Blalock-Taussig

Jatene

Norwood

Mustard

Ross

Atrio-ventricular Septal Defect

Tetralogy of Fallot

-Senning Damus-Kaye-Stansel

Rastelli

Blalock-Hanlon

Rashkind

Pulmonary Artery Band

Potts

Waterston

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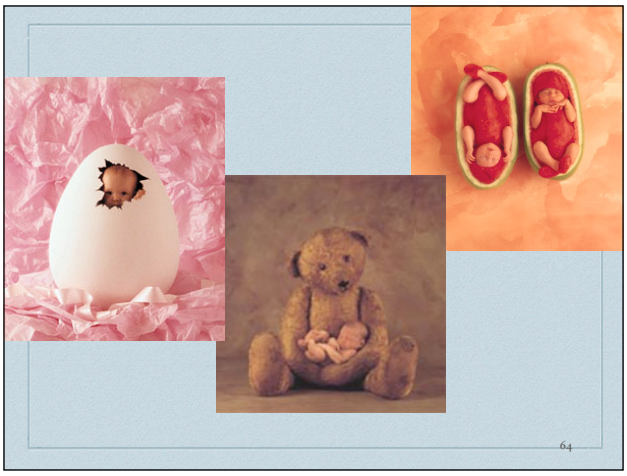

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