CONGENITAL HEART DISEASES

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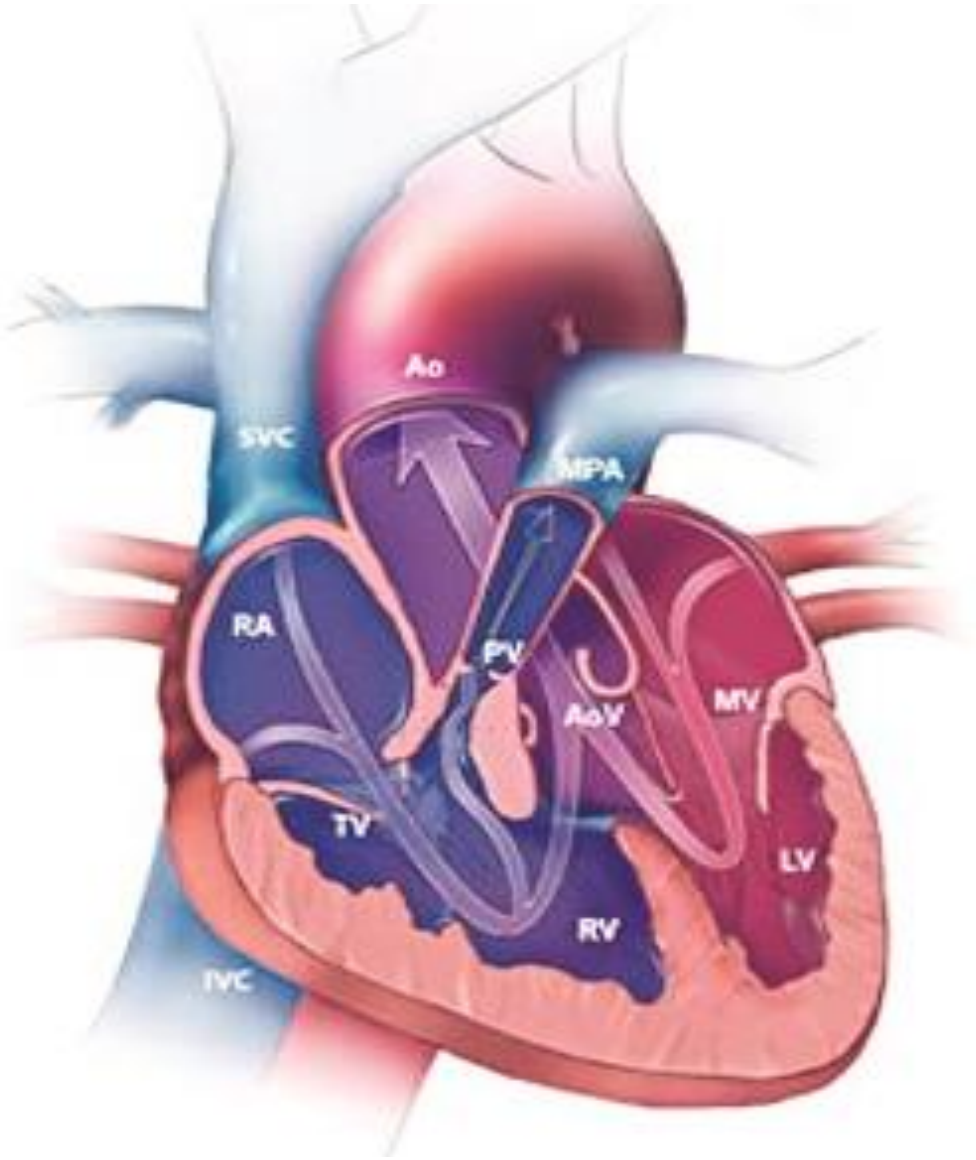
CYANOTIC CHD

TETRALOGY OF FALLOT

- 10% of all CHD.
- TOF include four abnormalities:
  1. **Overriding of the aorta.**
  2. **Large VSD**
  3. **RVOT obstruction.**
  4. **RVH**



Clinical Manifestations

- Most infants with TOF are symptomatic, with **cyanosis**, clubbing, dyspnea on exertion, **squatting**, or **hypoxic spells**.

- Right ventricular heave and a systolic thrill at the LSB are usually found.

- A loud and single S2, and a loud systolic ejection murmur at the middle and upper LSB are present (from the RVOT obstruction, not from the VSD)

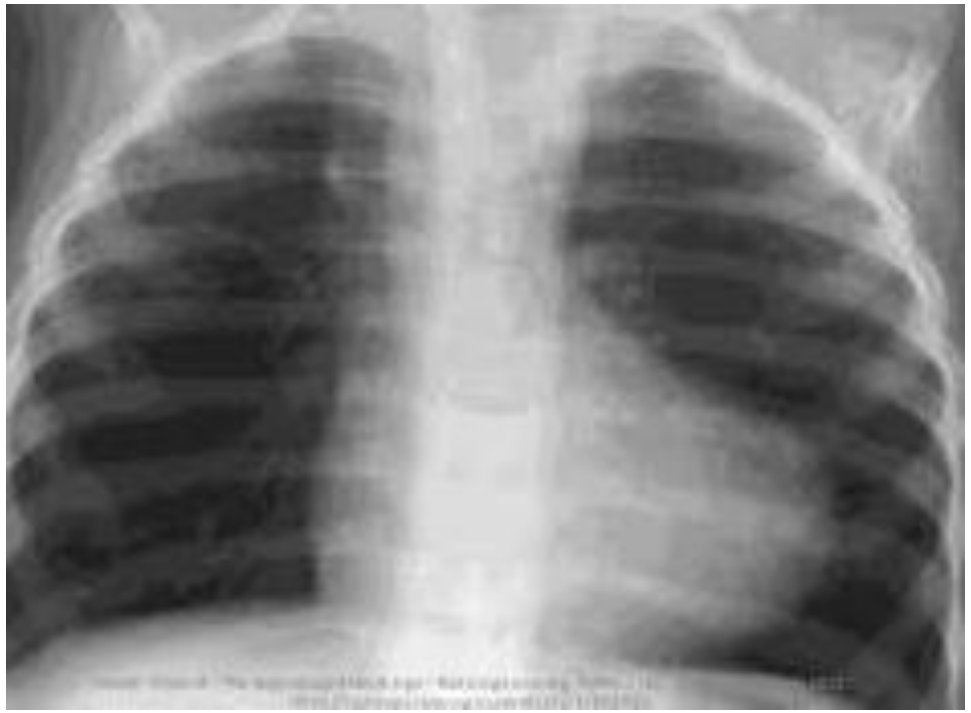
- Hypoxic spells may develop in infants. Brain abscess, cerebrovascular accident, and SBE are rare complications.

- Polycythemia is common, but relative iron deficiency state (hypochromic) with normal hematocrit may be present.

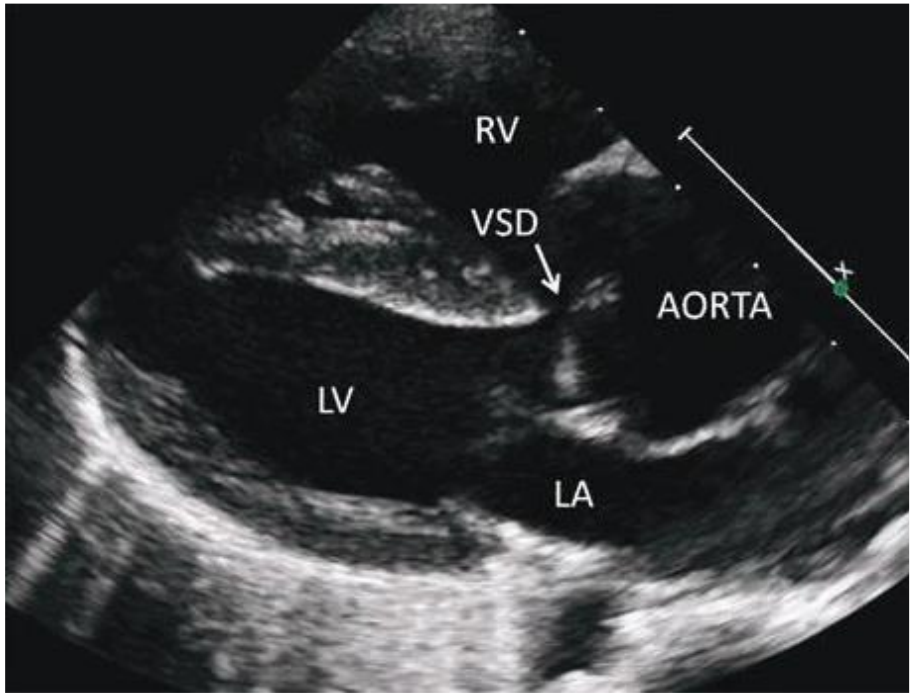
- Coagulopathies are late complications of a long-standing severe cyanosis.

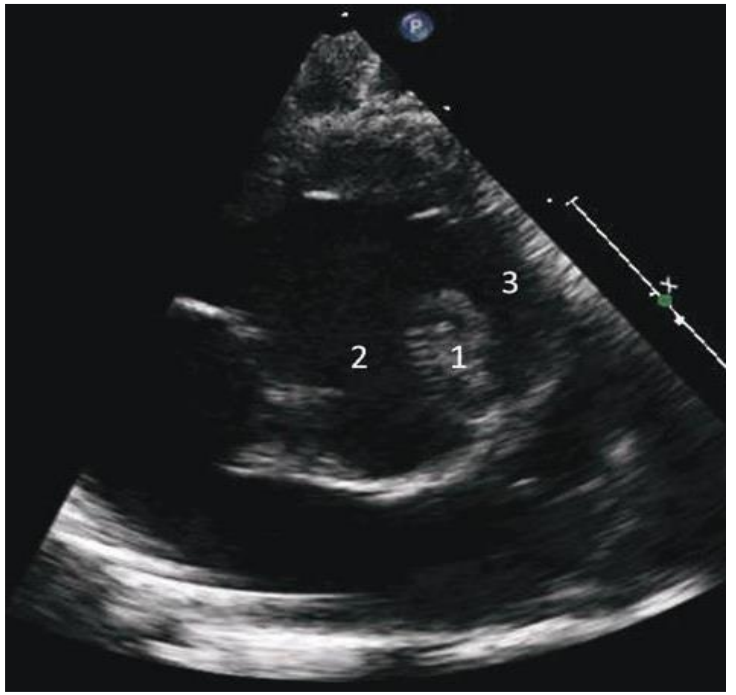
- **ECG** → RAD and RVH.

- **CXR** → a boot-shaped heart with a concave MPA segment.
  → Right aortic arch is present in 25% of the cases.



- **Echo** → a large subaortic VSD and an overriding of the aorta in the parasternal long-axis view.



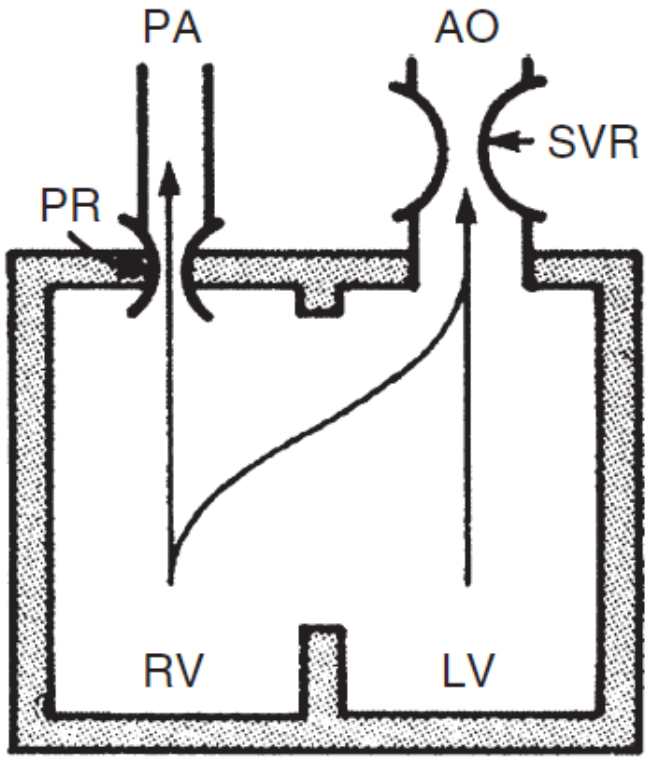


Hypoxic Spell

- Hypoxic spell (also called cyanotic spell or “tet” spell) is characterized by
  (1) a paroxysm of hyperpnea (rapid and deep respiration).
  (2) irritability and prolonged crying.
  (3) increasing cyanosis.
  (4) decreased intensity of the heart murmur.
- A severe spell may lead to limpness, convulsion, cerebrovascular accident, or even death.
- It occurs in young infants, with peak incidence 2-4 months of age.

Pathophysiology of hypoxic spell:

- In TOF, the RV and LV can be viewed as a single pumping chamber, as there are large VSD equalizing pressures in both ventricles



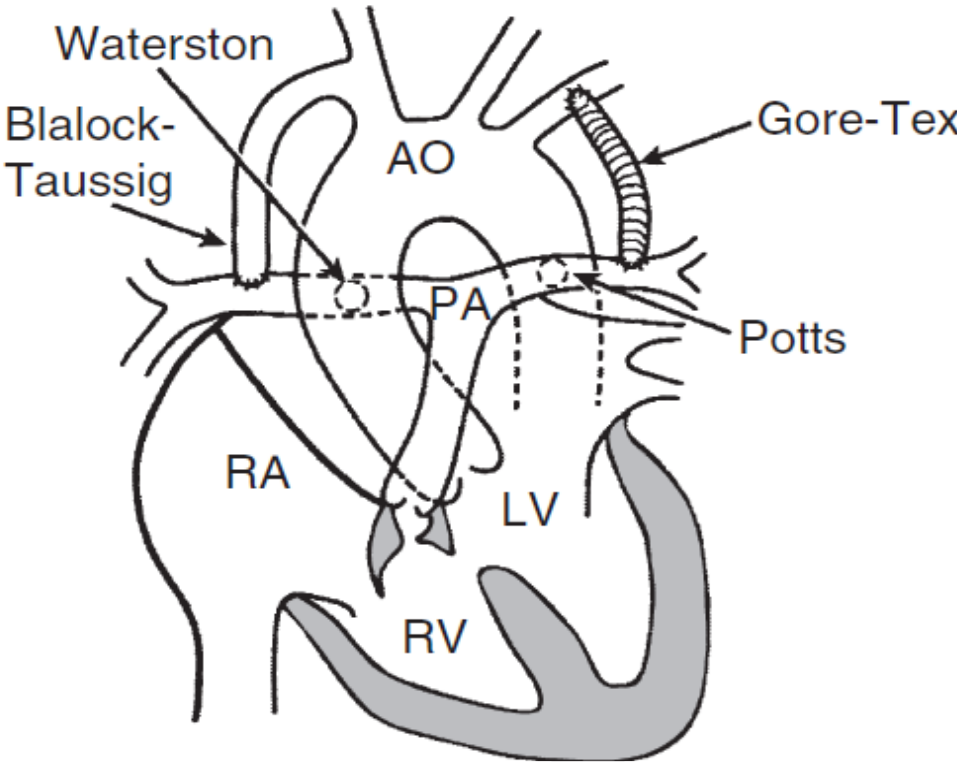
Medical therapy

- The infant should be picked up and comforted as soon as an episode begins, ideally while being held in a position of flexed knees and hips that kinks or compresses the femoral arteries and increases peripheral systemic vascular resistance.
- If no improvement is seen within a few minutes, oxygen should be administered and intravenous access obtained.

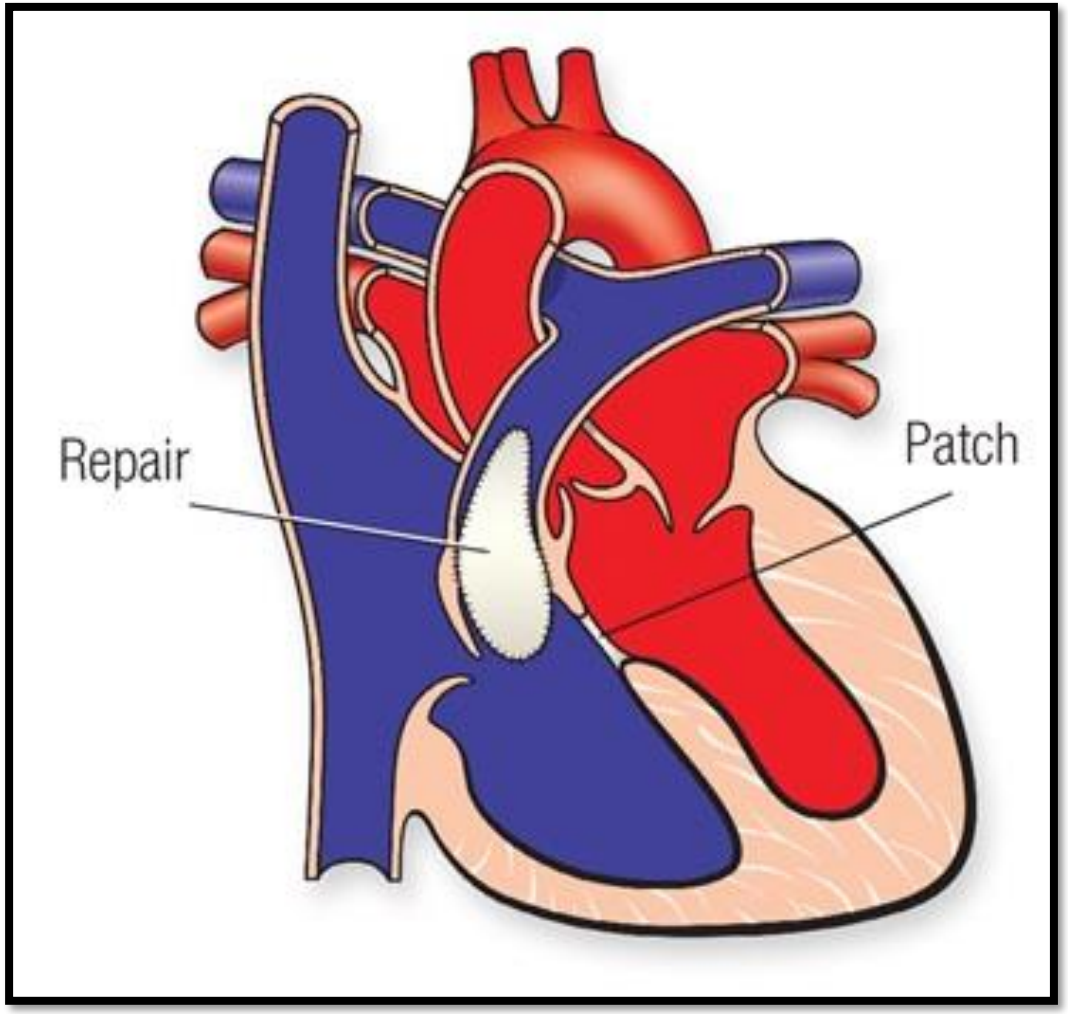
1. An intravenous bolus of **colloid or crystalloid fluid**.
2. Intravenous (or intramuscular) **morphine** (0.1 to 0.2 mg/kg).
3. Intravenous **propranolol** (0.015 to 0.02 mg/kg).
4. Intravenous **sodium bicarbonate** (1 mEq/kg) may be required if there is evidence of worsening acidosis.
5. In unremitting cases, intravenous systemic **vasoconstrictors**, for example, phenylephrine (boluses of 0.005 to 0.001 mg/kg), or norepinephrine (0.05 to 1.0 *m*g/kg/min) may be required.
6. **Anesthesia**, intubation, and ventilation may ultimately be required.
7. Very occasionally, severe life-threatening spells may require emergent surgical intervention or mechanical circulatory support

Surgical

1. **Palliative procedures** are indicated to increase PBF in infants with Severe cyanosis or uncontrollable hypoxic spells on whom the corrective surgery cannot safely be performed, and in children with hypoplastic PA on whom the corrective surgery is technically difficult.
   - **The Blalock-Taussig (B-T) or modified B-T shunt**
   - Waterston shunt.
   - Potts operation.

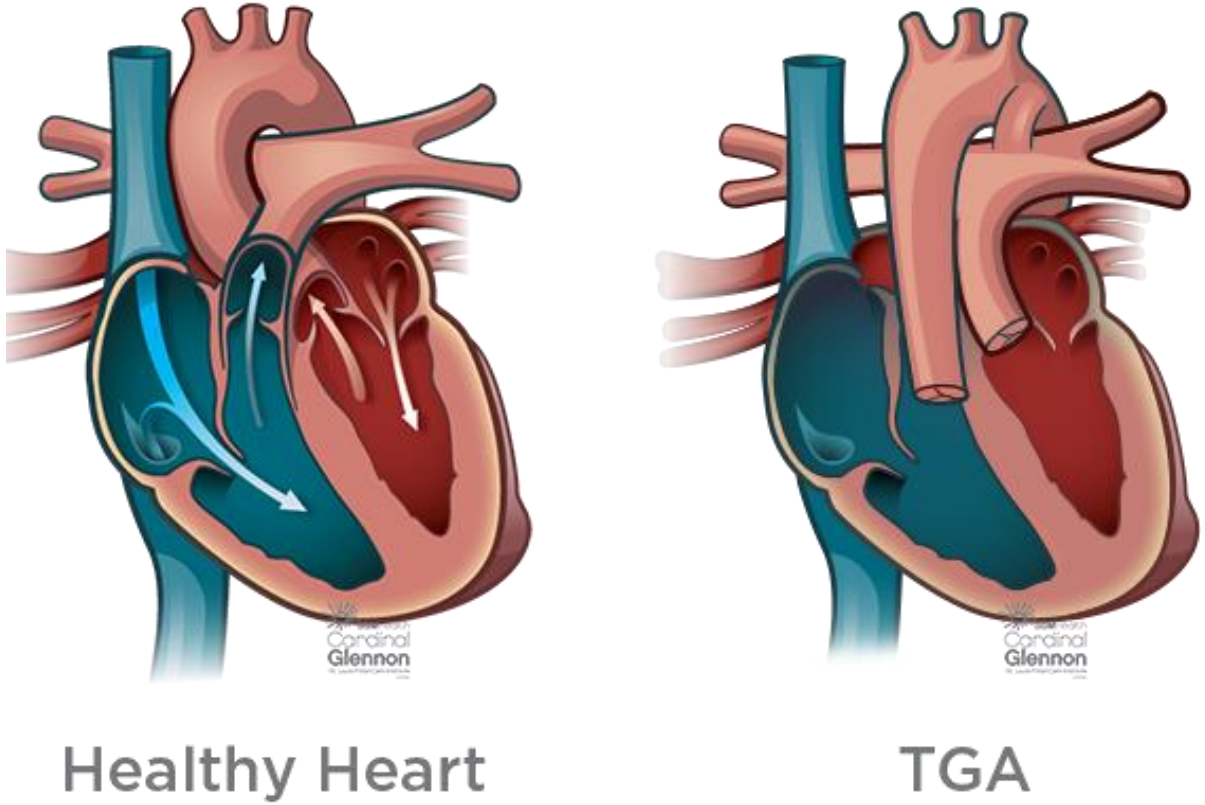


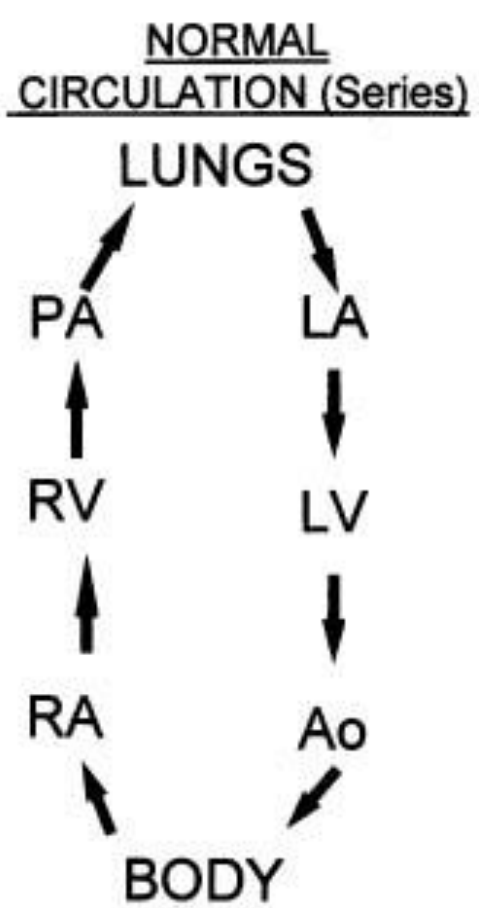
2. Complete repair surgery

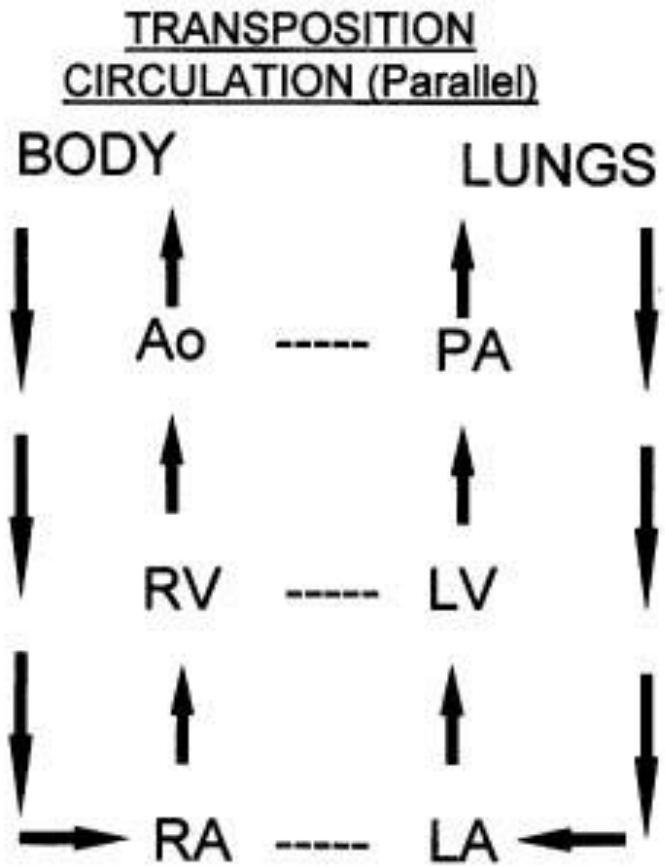


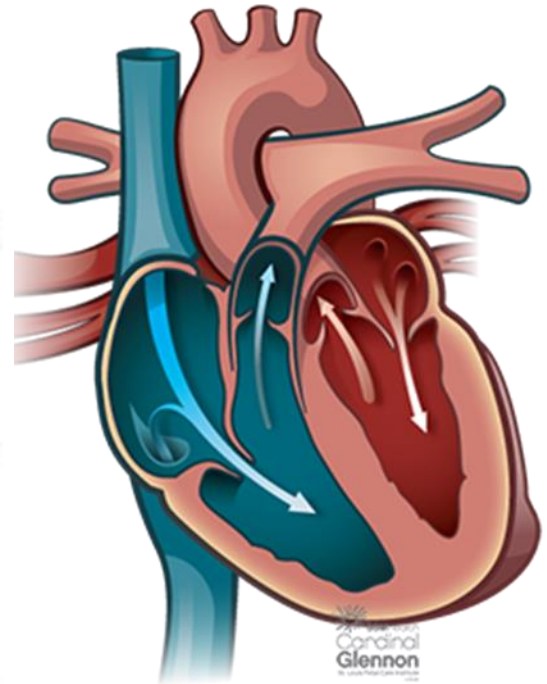
Transposition of the great arteries

- TGA constitutes 5% of all CHD. It is more common in boys (3:1).
- The aorta (AO) and the pulmonary artery are transposed, with the AO arising anteriorly from the RV, and the PA arising posteriorly from the LV.

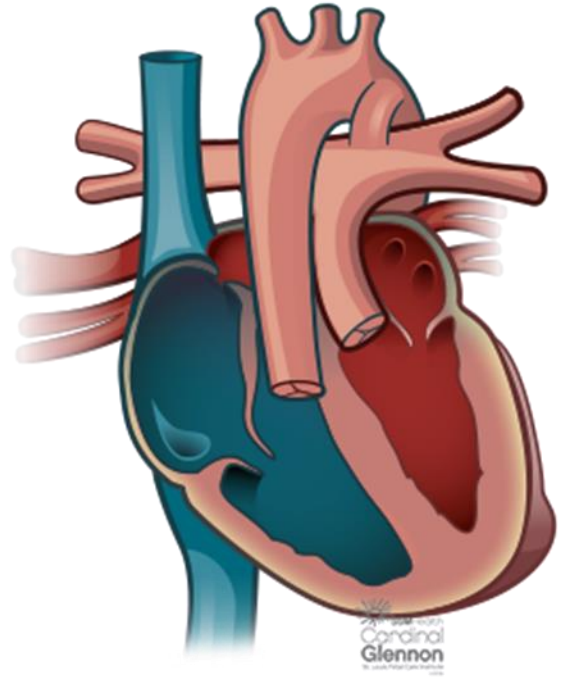








Healthy Heart

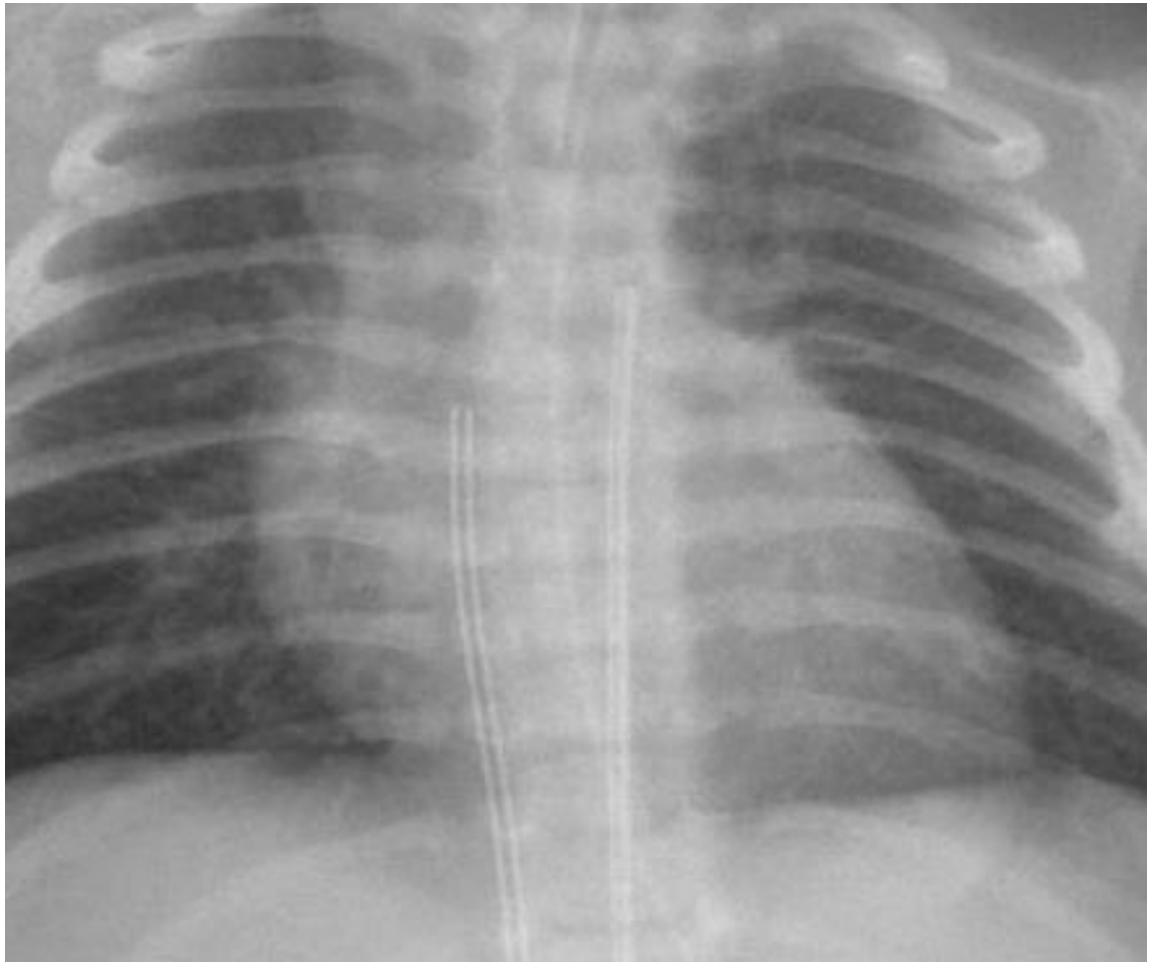


TGA

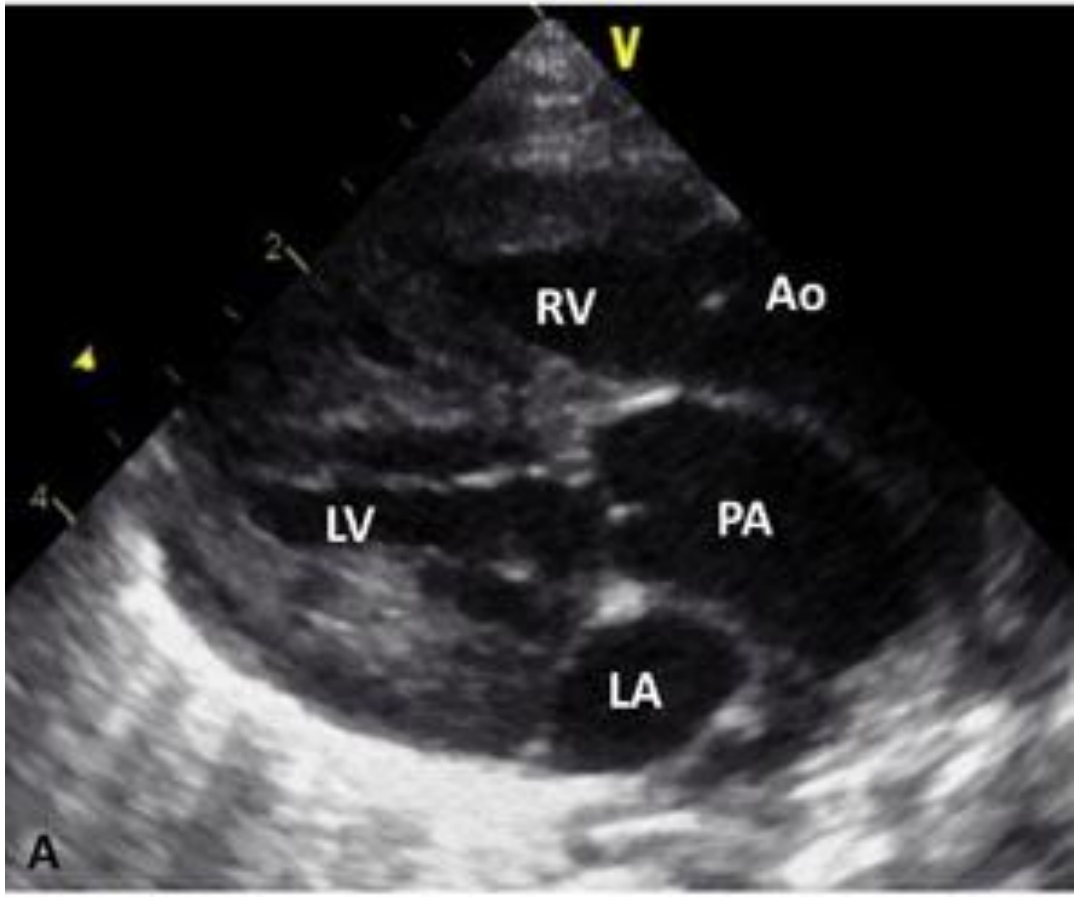
Clinical Manifestations

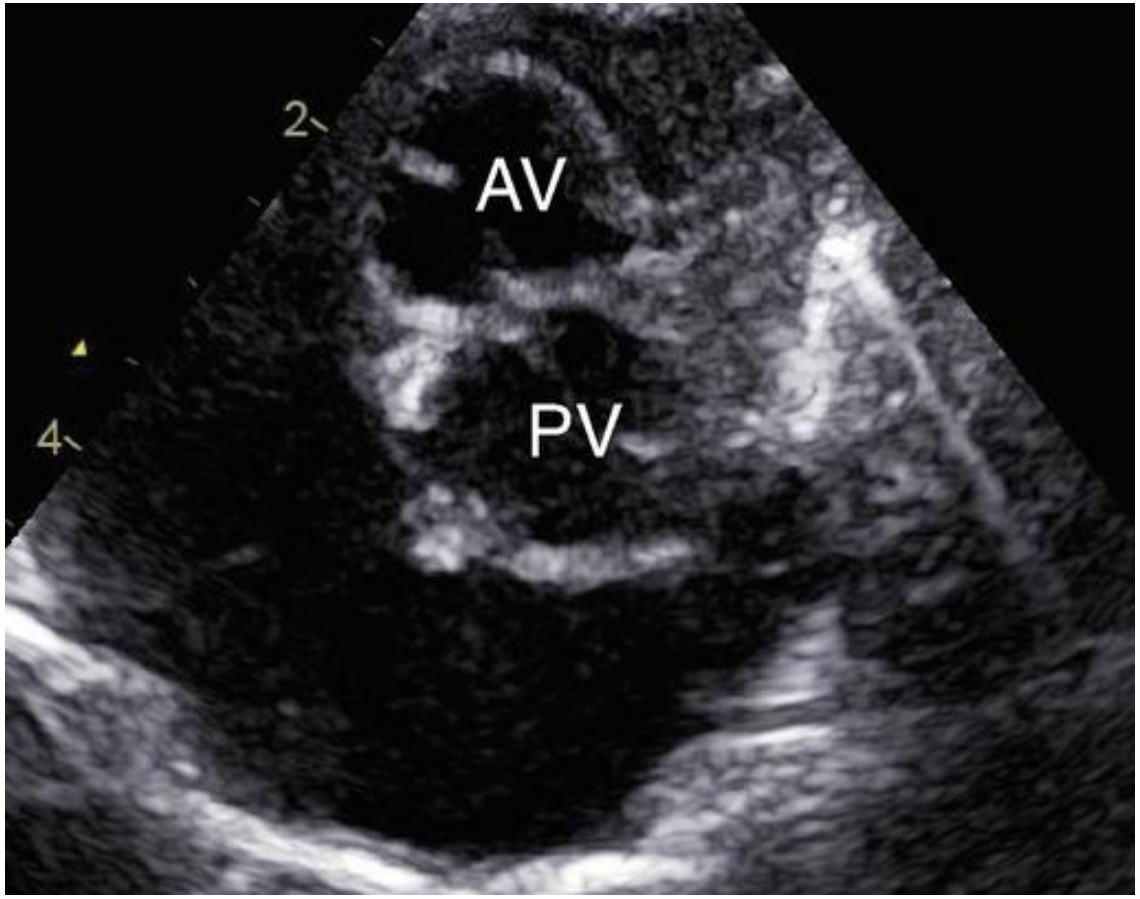
- Cyanosis and signs of CHF develop in the newborn period. Severe arterial hypoxemia unresponsive to oxygen inhalation and acidosis are present in neonates with poor mixing.
- Auscultatory findings are nonspecific. The S2 is single and loud. No heart murmur is audible in infants with intact ventricular septum. When TGA is associated with VSD or PS, a systolic murmur of these defects may be audible.

- **ECG** → RAD and RVH.
- **CXR** → cardiomegaly with increased PVMs. An **egg-shaped** cardiac silhouette with a narrow superior mediastinum is characteristic.



- **Echo** → study is diagnostic.





Natural history and prognosis depend on anatomy.

- Infants with intact ventricular septum are the sickest group, but they demonstrate the most dramatic improvement following PGE1 infusion or the Rashkind balloon atrial septostomy.

- Infants with VSD or large PDA are the least cyanotic group but are most likely to develop CHF and PVOD (beginning as early as 3 or 4 months of age).

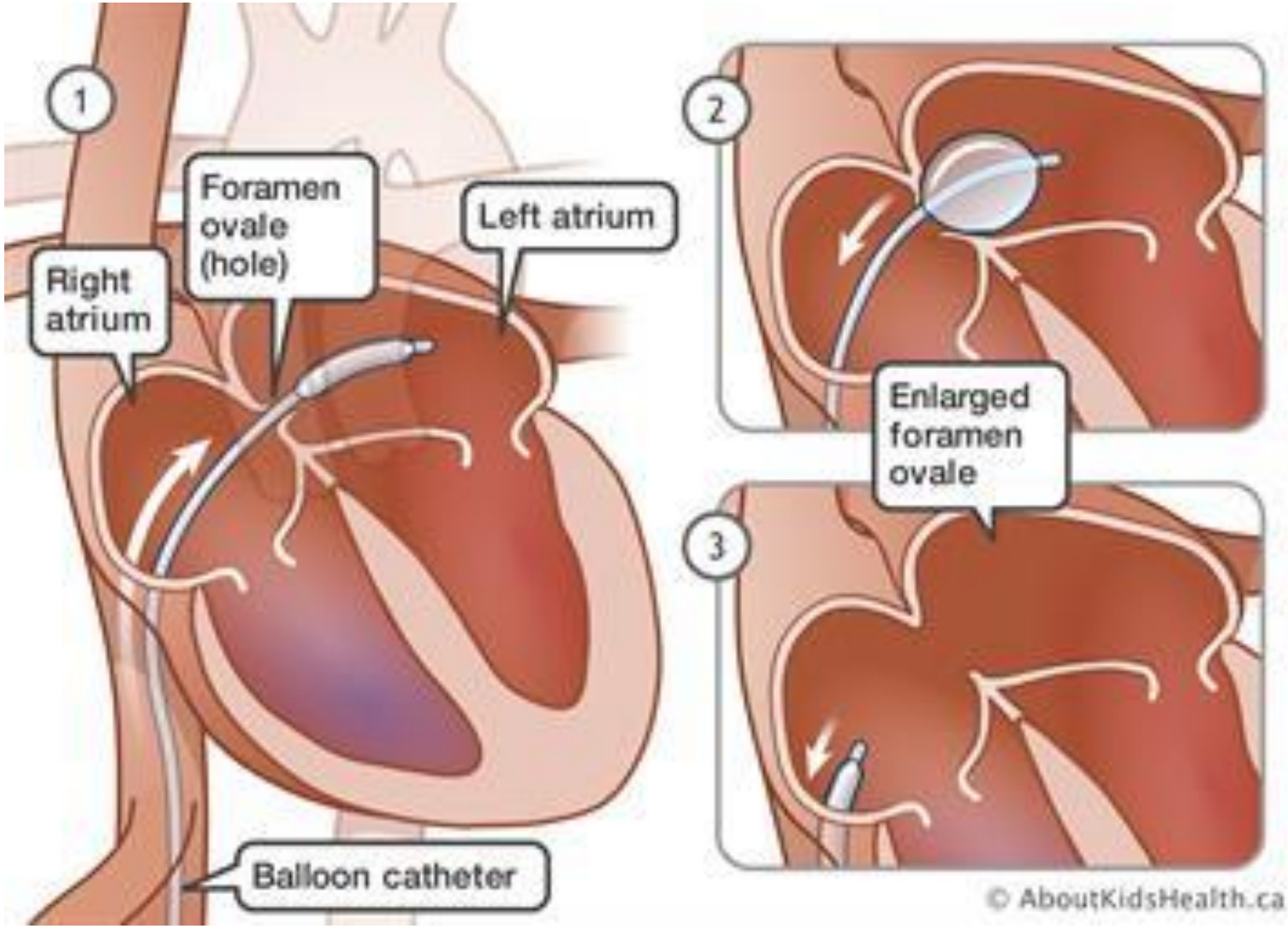
- Cerebrovascular accident and progressive PVOD, particularly in infants with large VSD or PDA, are rare late complications.

Management

Medical

- Metabolic acidosis, hypoglycemia, and hypocalcemia should be treated if present.
- PGE1 infusion is started to raise arterial oxygen saturation by reopening the ductus.
- Administration of oxygen may help raise systemic arterial oxygen saturation by lowering PVR and increasing PBF, with resulting increase in mixing.

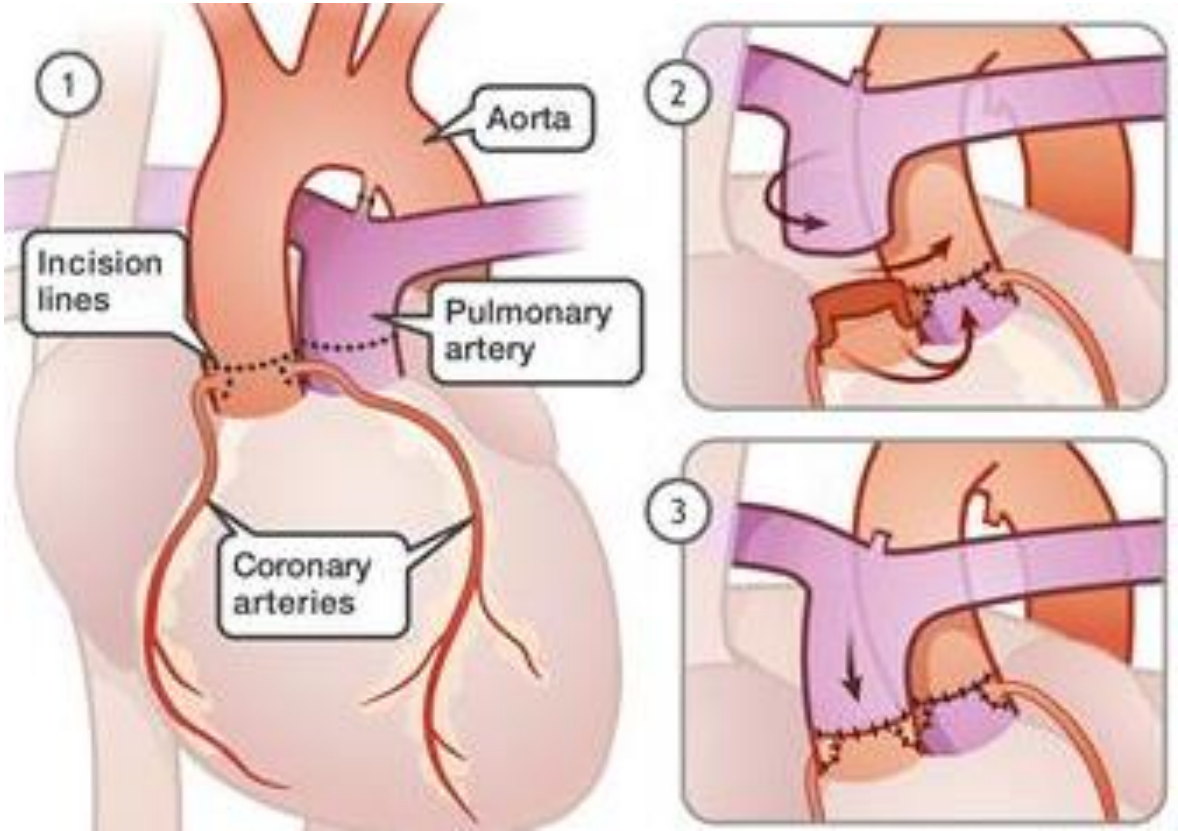
- A therapeutic balloon atrial septostomy (**Rashkind procedure**) may be performed.



Surgical

As a definitive surgery, the right- and left-sided structures are switched

- at the atrial level (Senning operation)
- at the ventricular level (Rastelli operation).
- **at the great artery level** (**arterial switch operation**).



Congestive Heart Failure

- Congestive heart failure (CHF) is a clinical syndrome in which the heart is unable to pump enough blood to the body to meet its needs, to dispose of systemic or pulmonary venous return adequately, or a combination of the two.

Causes

- The heart failure syndrome may arise from diverse causes.
- By far the most common causes of CHF in infancy are CHDs.
- Beyond infancy, myocardial dysfunction of various etiologies is an important cause of CHF.
- Tachyarrhythmias and heart block can also cause heart failure at any age.

1. Congenital heart disease

- Volume overload lesions such as VSD, PDA, and AVSD are the most common causes of CHF in the first 6 months of life.

- Large L-R shunt lesions, such as VSD and PDA, do not cause CHF before 6 to 8 weeks of age because the pulmonary vascular resistance (PVR) does not fall low enough to cause a large shunt until this age. CHF may occur earlier in premature infants (within the first month) because of an earlier fall in the PVR.

CAUSES OF CONGESTIVE HEART FAILURE RESULTING FROM CONGENITAL HEART DISEASE

AGE OF ONSET	CAUSE
At birth	HLHS
	Volume overload lesions
	Severe tricuspid or pulmonary insufficiency
	Large systemic arteriovenous fistula
First wk	TGA
	PDA in small premature infants
	HLHS (with more favorable anatomy)
	TAPVR with pulmonary venous obstruction
	Critical AS or PS
	Systemic arteriovenous fistula
1-4 wk	COA with associated anomalies
	Critical AS
	Large left-to-right shunt lesions (VSD, PDA) in premature infants
	All other lesions previously listed
4-6 wk	Some left-to-right shunt lesions such as ECD
6 wk-4 mo	Large VSD
	Large PDA
	Others such as anomalous left coronary artery from the PA

AS, aortic stenosis; COA, coarctation of the aorta; ECD, endocardial cushion defect; HLHS, hypoplastic left heart syndrome; PA, pulmonary artery; PDA, patent ductus arteriosus; PS, pulmonary stenosis; TAPVR, total anomalous pulmonary venous return; TGA, transposition of the great arteries; VSD, ventricular septal defect.

2. Acquired heart disease.

- **Viral myocarditis** (in toddlers).
- **Myocarditis associated with Kawasaki disease** (1 to 4 years of age).
- **Acute rheumatic carditis** (in school-age children).
- **Rheumatic valvular heart diseases**, such as MR or AR (older children and adults).
- **Dilated cardiomyopathy** (at any age during childhood and adolescence).
- **Doxorubicin cardiomyopathy** (months to years after chemotherapy).
- **Cardiomyopathies** associated with muscular dystrophy and Friedreich's ataxia (in older children and adolescents).

3. Miscellaneous causes

- **Metabolic abnormalities** (severe hypoxia, acidosis, hypoglycemia, hypocalcemia) (in newborns)
- **Hyperthyroidism** (at any age)
- **Supraventricular tachycardia** (in early infancy)
- **Complete heart block** associated with CHDs (in the newborn period or early infancy)
- **Severe anemia** (at any age)
- **Primary carnitine deficiency** (2-4 years)
- **Acute cor pulmonale** caused by acute airway obstruction (during early childhood)
- **Acute systemic hypertension with glomerulonephritis** (school-age children)

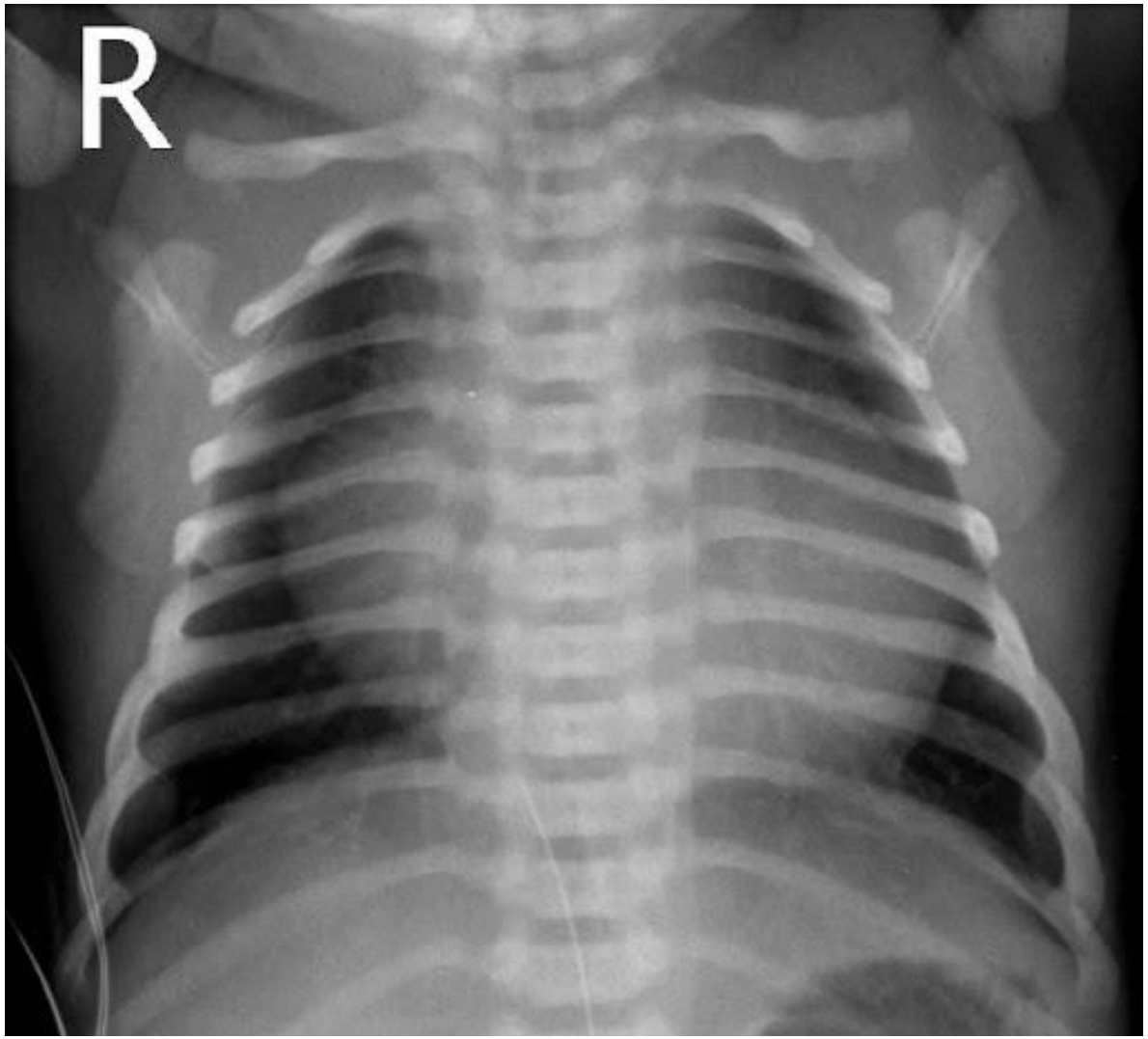
Diagnosis of CHF

- Poor feeding of recent onset, tachypnea, poor weight gain, and cold sweat on the forehead suggest CHF in infants.
- In older children, shortness of breath, especially with activities, easy fatigability, puffy eyelids, or swollen feet may be presenting complaints.

Diagnosis of CHF

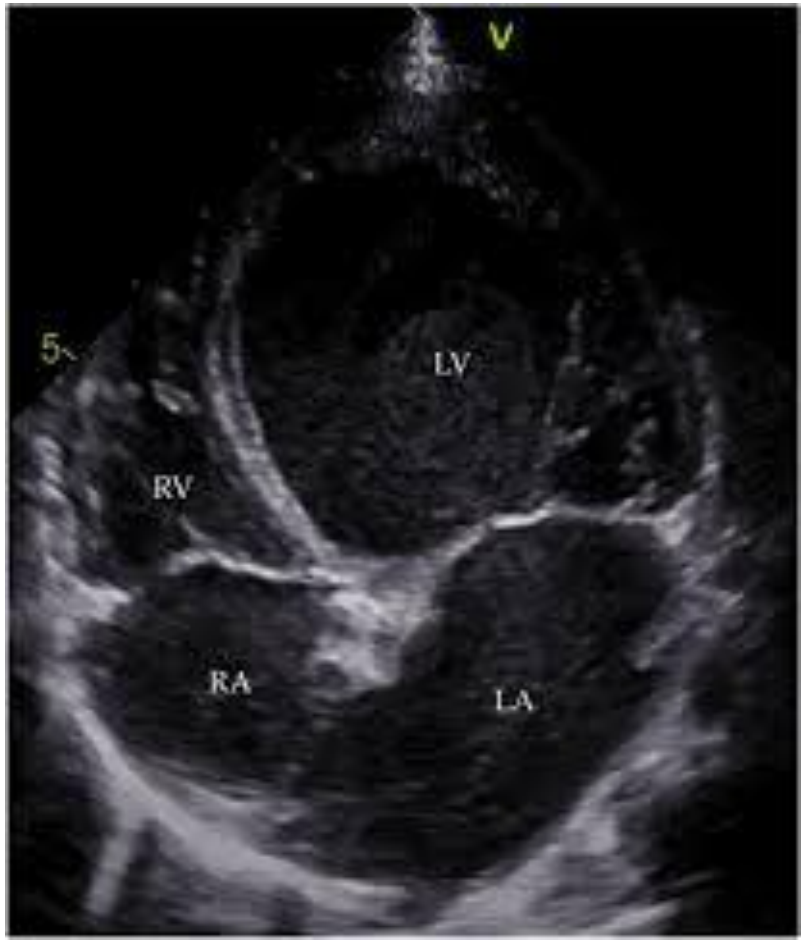
- Physical findings can be divided by pathophysiologic subgroups.
- a. Compensatory responses to impaired cardiac function.
    - (1) Tachycardia, gallop rhythm, weak and thready pulse, and cardiomegaly on chest radiographs.
    - (2) Signs of increased sympathetic discharges (growth failure, perspiration, and cold wet skin).
- b. **Signs of pulmonary venous congestion** (left-sided failure) include tachypnea, dyspnea on exertion (or poor feeding in small infants), orthopnea in older children, and rarely wheezing and pulmonary crackles.
- c. **Signs of systemic venous congestion** (right-sided failure) include hepatomegaly and puffy eyelids. Distended neck veins and ankle edema are not seen in infants.

- Cardiomegaly on chest radiograph is almost always present.



- The ECG is not helpful in deciding whether the patient is in CHF, although it may be helpful in determining the cause.

- Echo studies confirm the presence of chamber enlargement or impaired LV function and help determine the cause of CHF.



- Increased levels of plasma natriuretic peptides (atrial natriuretic peptide [ANP] and B-type natriuretic peptide [BNP]) are helpful in diagnosis and follow up.

Management

- Elimination of the underlying causes or correction of precipitating or contributing causes (e.g., infection, anemia, arrhythmias, fever, hypertension)

- General supportive measures including: Bed rest, Nutritional supports

- Control of heart failure state by use of drugs, such as inotropic agents, diuretics, or afterload-reducing agents.

Drug therapy

Diuretics.

- Diuretics remain the principal therapeutic agent to control pulmonary and systemic venous congestion. Diuretics only reduce preload and improve congestive symptoms.

DIURETIC AGENTS AND DOSAGES

PREPARATION	ROUTE	DOSAGE
THIAZIDE DIURETICS		
Chlorothiazide (Diuril)	Oral	20-40 mg/kg/day in 2 to 3 divided doses
Hydrochlorothiazide (HydroDIURIL)	Oral	2-4 mg/kg/day in 2 to 3 divided doses
LOOP DIURETICS		
Furosemide (Lasix)	IV	1 mg/kg/dose
	Oral	2-3 mg/kg/day in 2 to 3 divided doses
Ethacrynic acid (Edecrin)	IV	1 mg/kg/dose
	Oral	2-3 mg/kg/day in 2 to 3 divided doses
ALDOSTERONE ANTAGONIST		
Spironolactone (Aldactone)	Oral	1-3 mg/kg/day in 2 to 3 divided doses

Rapidly acting inotropic agents.

- In critically ill infants with CHF, rapidly acting catecholamines with a short duration of action are preferable.

SUGGESTED STARTING DOSAGES OF CATECHOLAMINES

DRUG	DOSAGE AND ROUTE	SIDE EFFECTS
Epinephrine (Adrenalin)	0.1-1 μg/kg/min IV	Hypertension, arrhythmias
Isoproterenol (Isuprel)	0.1-0.5 μg/kg/min IV	Peripheral and pulmonary vasodilatation
Dobutamine (Dobutrex)	2-8 μg/kg/min IV	Little tachycardia and vasodilatation, arrhythmias
Dopamine (Intropin)	5-10 μg/kg/min IV	Tachycardia, arrhythmias, hypertension or hypotension Dose-related cardiovascular effects (μg/kg/min): Renal vasodilatation: 2-5 Inotropic: 5-8 Tachycardia: >8 Mild vasoconstriction: >10 Vasoconstriction: 15-20

Afterload-reducing agents.

- Reducing afterload tends to augment the stroke volume without a great change in the inotropic state of the heart and therefore without increasing myocardial oxygen consumption.

- Combined use of an inotropic agent, a vasodilator, and a diuretic produces most improvement in both inotropic state and congestive symptoms

DOSAGES OF VASODILATORS

DRUG	ROUTE AND DOSAGE	COMMENTS
ARTERIOLAR VASODILATOR		
Hydralazine (Apresoline)	IV: 0.15-0.2 mg/kg/dose, every 4 to 6 hr (maximum 20 mg/dose) Oral: 0.75-3 mg/kg/day, in 2 to 4 doses (maximum 200 mg/day)	May cause tachycardia; may be used with propranolol May cause gastrointestinal symptoms, neutropenia, and lupus-like syndrome
VENODILATORS		
Nitroglycerin	IV: 0.5-1 μg/kg/min (maximum 6 μg/kg/min)	Start with small dose and titrate based on effects
MIXED VASODILATORS		
Captopril (Capoten)	Oral: Newborn: 0.1-0.4 mg/kg, TID-QID Infant: Initially 0.15-0.3 mg/kg, QD-QID. Titrate upward if needed. Max dose 6 mg/kg/24 hr. Child: Initially 0.3-0.5 mg/kg, BID-TID. Titrate upward if needed. Max dose 6 mg/kg/24 hr. Adolescents and adults: Initially 12.5-25 mg, BID-TID. Increase weekly if needed by 25 mg/dose to max dose 450 mg/24 hr.	May cause hypotension, dizziness, neutropenia, and proteinuria Dose should be reduced in patients with impaired renal function
Enalapril (Vasotec)	Oral: 0.1 mg/kg, once or twice daily	Patient may develop hypotension, dizziness, or syncope

Digoxin

- Digoxin act by inhibition of Na+/K+ ATPase mainly in the myocardium → increases the cardiac output (or contractile state of the myocardium).

- Use of digoxin in infants with large L-R shunt lesions (e.g., large VSD) is **controversial** because ventricular contractility is normal in this situation. However, studies have shown that digoxin improves symptoms in these infants, perhaps because of other actions of digoxin, such as parasympathomimetic action and diuretic action.

ORAL DIGOXIN DOSAGE FOR CONGESTIVE HEART FAILURE

AGE	TOTAL DIGITALIZING DOSE (μg/kg)	MAINTENANCE DOSE* (μg/kg/day)
Premature	20	5
Newborns	30	8
<2 yr	40-50	10-12
>2 yr	30-40	8-10

*The maintenance dose is 25% of the total digitalizing dose in two divided doses. The IV dose is 75% of the oral dose.

β-Adrenergic blockers

- β-adrenergic blockers have been shown to be beneficial in patients with **Chronic CHF**, who were treated with standard anticongestive drugs to overcome the adrenergic overstimulation. (may have detrimental effects on the failing heart -Acute HF- by inducing myocyte injury and necrosis).

- Carvedilol, Metoprolol, Bisoprolol.

THANK YOU