Cyanotic Congenital Heart Diseases

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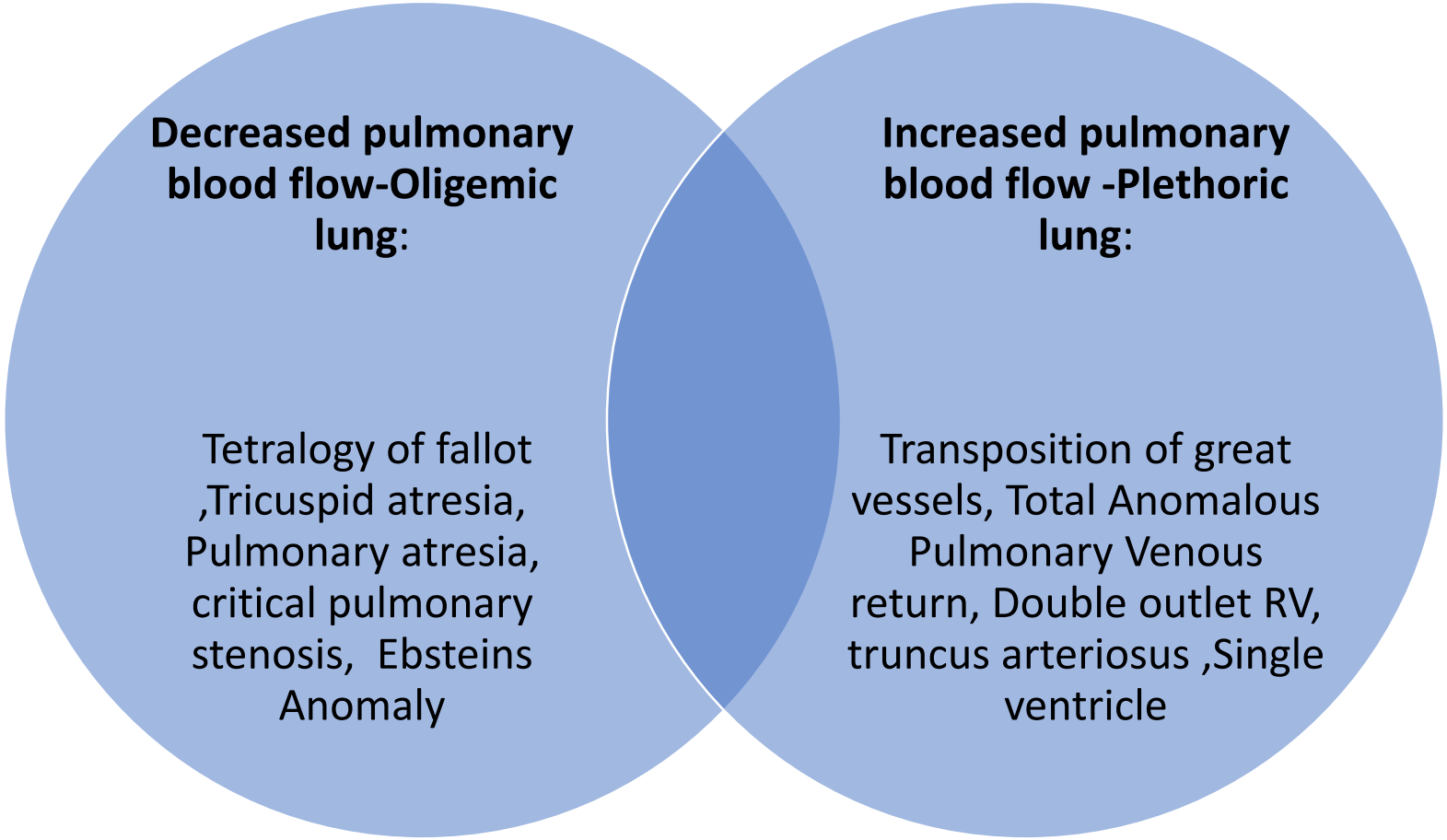
- Cyanosis – bluish discoloration of the skin and nail beds and mucus membranes appears when tissues are deprived of adequate amount of O2.
- It becomes visible when hemoglobin, **approximately 5 gms/dl circulates unbound to O2** and **the measured O2 saturation drops below 85%**

- Cyanosis can occur when:
  - **Blood flow to the lungs is decreased or insufficient (ebstien anomalies , TA+restrictive VSD , PA +intact septum,…)**
  - **Deoxygenated or desaturated blood is pumped to the body** due to:
    - **Right to left shunting lesions** when blood flow to the lungs is decreased and there is a communication between the intracardiac chambers or great vessels **(ex. TOF).**
    - **Mixing lesions** where blood flow to the lungs is increased or normal and there is a connection between the great arteries **(ex. Single ventricle without pulmonary stenosis or tricuspid atresia)**
    - **Pulmonary hypertension or pulmonary vascular disease** and a communication between the intracardiac chambers or great vessels **(A-V CANAL with Eisenmerger's complex)**

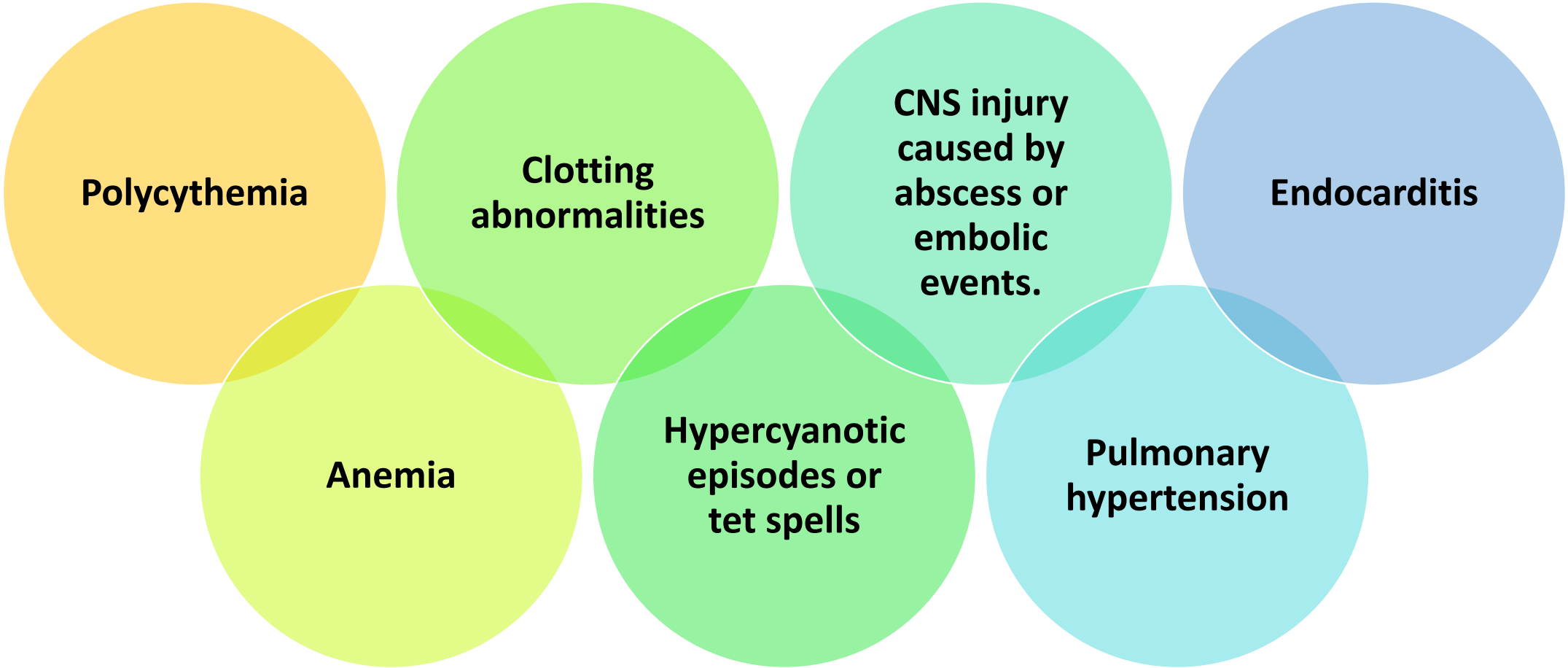
Cyanotic congenital heart disease

Classification according to Pulmonary Flow



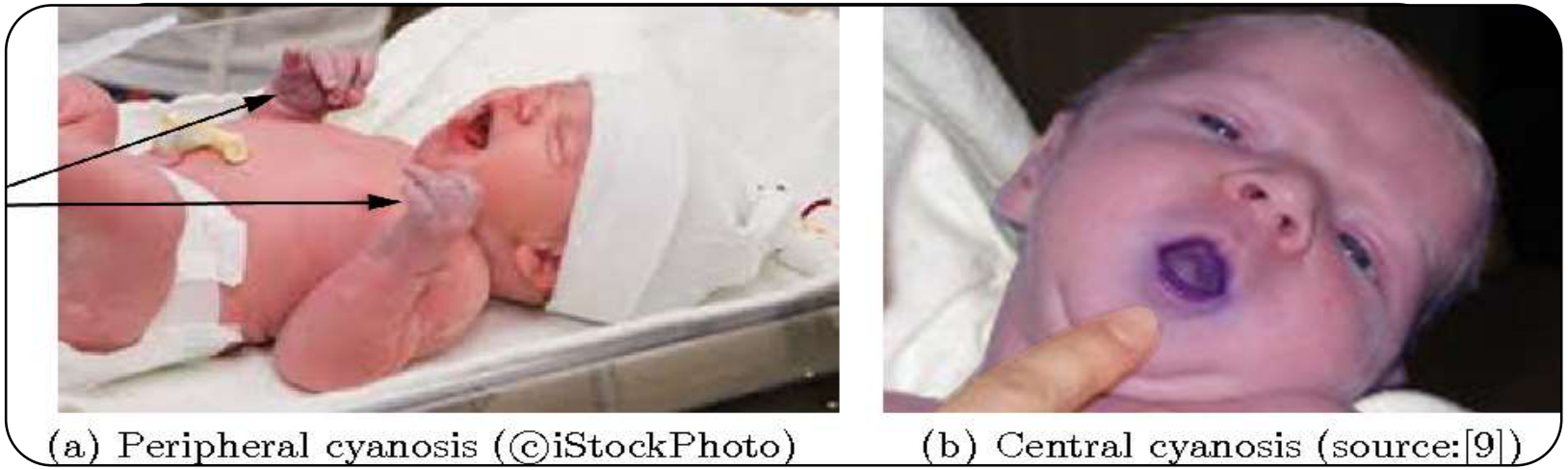
Cyanotic congenital heart disease

Clinical consequences of cyanosis



Cardiac findings in cyanotic heart diseases

Tachypnea **but usually without retraction**	**Lack of crackles or abnormal breath sounds** unless congestive heart failure supervenes	**Heart murmur that may be absent in serious forms of cyanotic heart defects**	A **continuous murmur (of patent ductus arteriosus) that may indicate restricted pulmonary blood flow through the ductus**.	Chest x-ray films that may **show cardiomegaly, abnormal cardiac silhouette, increased or decreased pulmonary vascular markings**	**Little or no** increase in **PO2 with oxygen administration**



(a) Peripheral cyanosis (©iStockPhoto)

(b) Central cyanosis (source:[9])

Arterial blood gases in room air

Arterial blood gases in room air confirm or reject central cyanosis.

An elevated Pa CO2 suggests pulmonary or central nervous system problems.

A low pH may be seen in sepsis, circulatory shock, or severe hypoxemia.

Hyperoxic test. Repeating arterial blood gases while the patient breathes 100% oxygen helps separate cardiac causes of cyanosis from pulmonary or central nervous system causes.

Hyperoxia test results in neonates with cyanosis

	PaO_2 (percent saturation) when $FiO_2 = 0.21$		PaO_2 (percent saturation) when $FiO_2 = 1$	$PaCO_2$
Normal	>70 (>95)		>300 (100)	35
Pulmonary disease	50 (85)		>150 (100)	50
Neurologic disease	50 (85)		>150 (100)	50
Methemoglobinemia	>70 (<85)		>200 (<85)	35
Cardiac disease				
Parallel circulation*	<40 (<75)		<50 (<85)	35
Mixing with reduced PBF¶	<40 (<75)		<50 (<85)	35
Mixing without restricted PBFΔ	40 to 60 (75 to 93)		<150 (<100)	35
	Preductal	**Postductal**		
Differential cyanosis◇	70 (95)	<40 (<75)	Variable	35 to 50
Reverse differential cyanosis§	<40 (<75)	>50 (>90)		

PaO_2: partial pressure of oxygen; FiO_2: fraction of inspired oxygen; $PaCO_2$: partial pressure of arterial carbon dioxide; PBF: pulmonary blood flow.

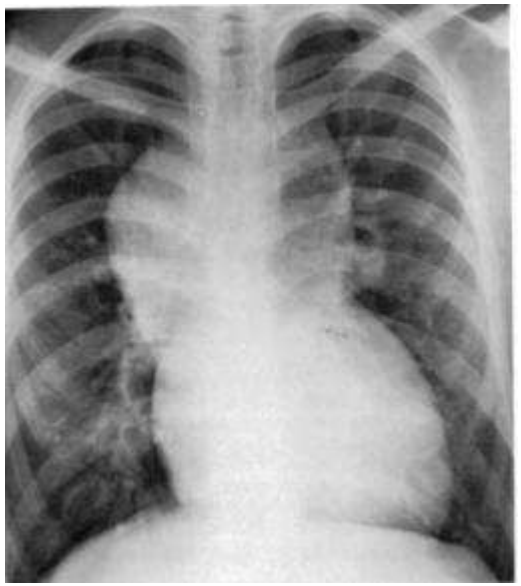
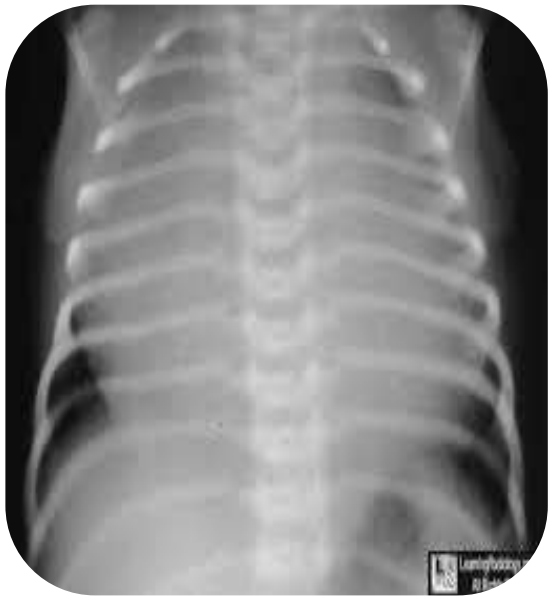
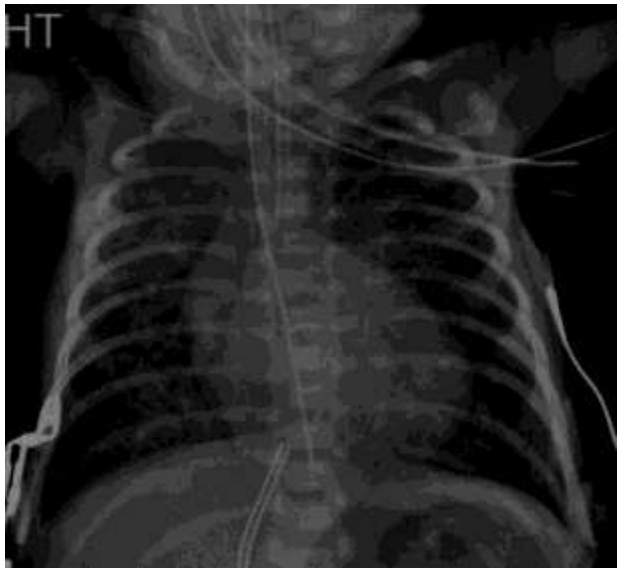
* D-transposition of the great arteries with or without ventricular septal defect.

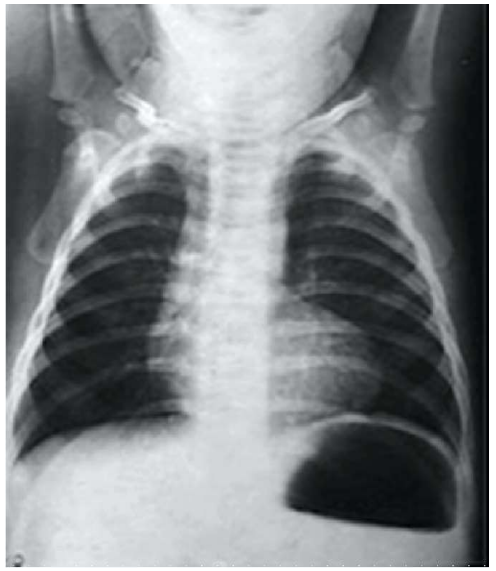
¶ Tricuspid atresia with pulmonary stenosis or pulmonary atresia, pulmonary atresia or critical pulmonary stenosis with intact ventricular septum, tetralogy of Fallot.

Δ Truncus arteriosus, total anomalous pulmonary venous connection without obstruction, hypoplastic left heart syndrome, single ventricle without pulmonary stenosis or pulmonary atresia.

◇ Persistent pulmonary hypertension of the newborn, interrupted aortic arch, severe coarctation.

§ D-transposition of the great arteries associated with either coarctation or suprasystemic pulmonary vascular resistance.

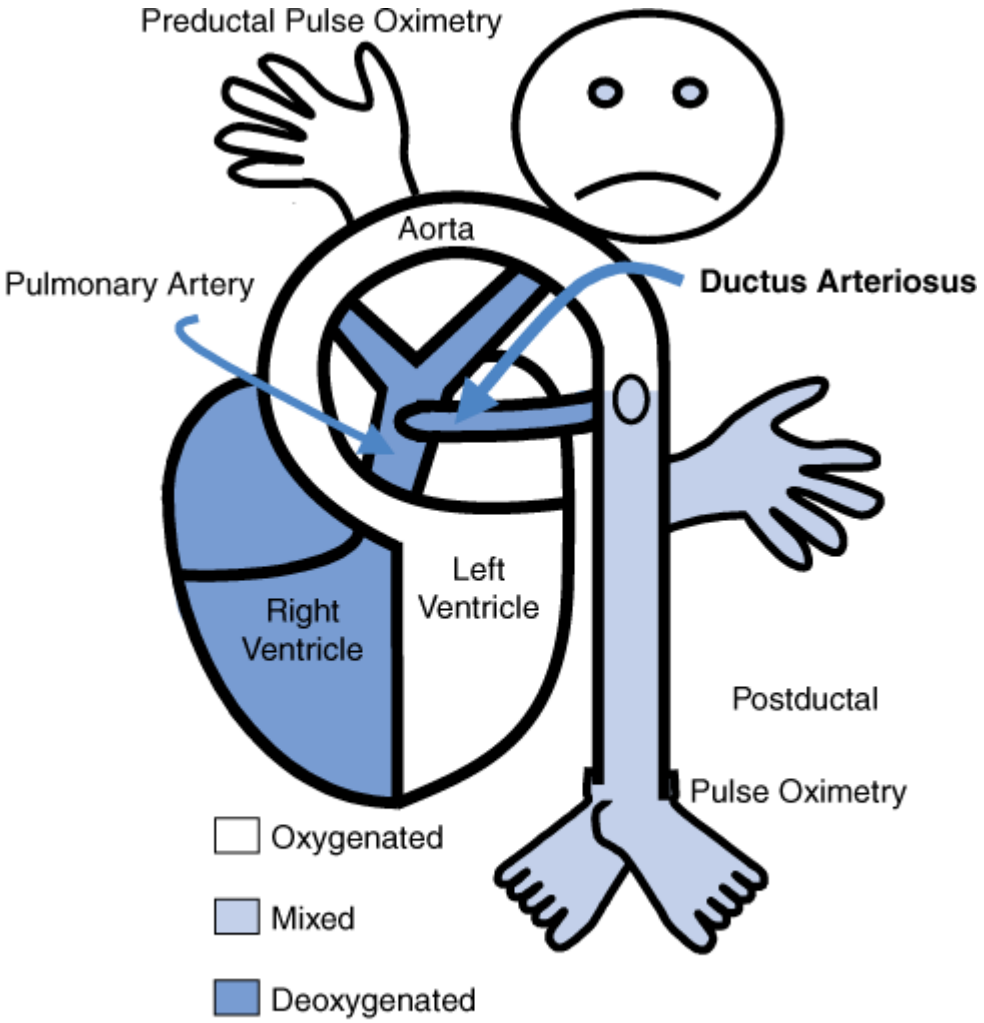


Chest x-ray films

Chest x-ray films may reveal pulmonary causes of cyanosis and the urgency of the problem. They can also hint at the presence or absence of cardiac defects and the type of defect.

Oximeter probes can be placed on preductal (right hand) and postductal (feet) sites to assess for right-to-left shunting at the level of the foramen ovale and ductus arteriosus

- **A difference of o2 sat greater than 3% between preductal and postductal oxygen saturations –is abn.**
- **A difference of o2 sat greater than 10% or PaO2 greater than 10 mmhg between preductal and postductal oxygen saturations correlates to right-to-left ductal shunting.**



Source: Lowry AW, Bhakta KY, Nag PK: *Texas Children's Hospital Handbook of Pediatrics and Neonatology*: www.accesspediatrics.com

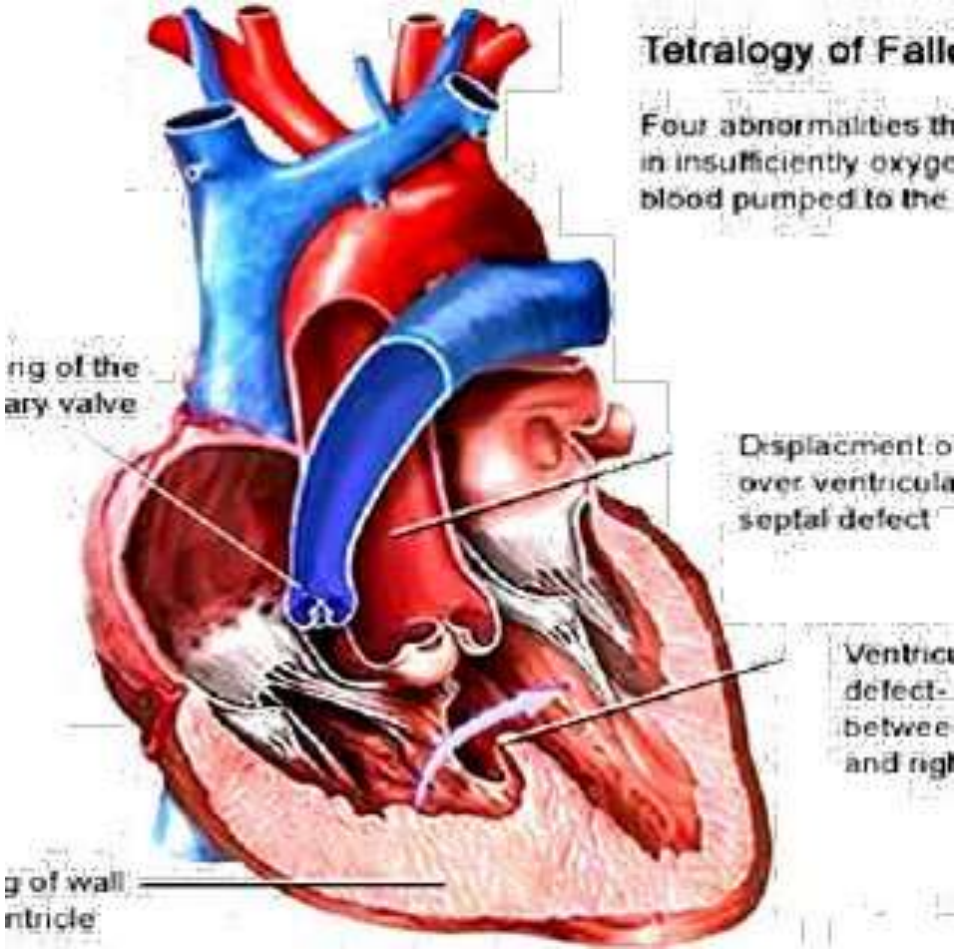
Prostaglandin E1 . If a cyanotic defect is suspected based on these laboratory tests, prostaglandin E 1 should be started or made available

CYANOTIC DISEASE
The 5 T's of cyanotic heart disease

- TOF(Tetralogy of fallot)
- TGV(Transposition of great vessels)
- Tricuspid atresia
- Truncus arteriosus
- Total anomalous of pulmonary venous drainage
- Hypoplastic left heart syndrome
- DORV, single ventricle , PA, Critical Pulmonary stenosis

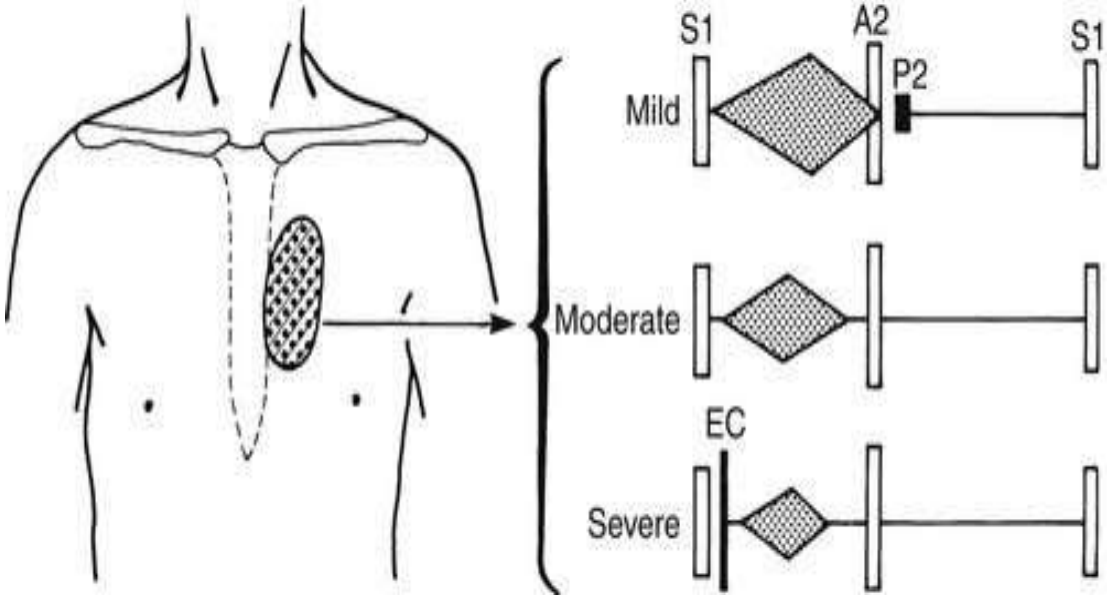
Tetralogy of Fallot

- TOF is **the most common cyanotic heart disease.**
- 4 features
  - **VSD**
  - **Overriding Aorta**
  - **Infundibular Pulmonic Stenosis(more severe with advancing age with R to L shunt increase)**
  - **RVH**
- Severity depend on PS.



Clinical features

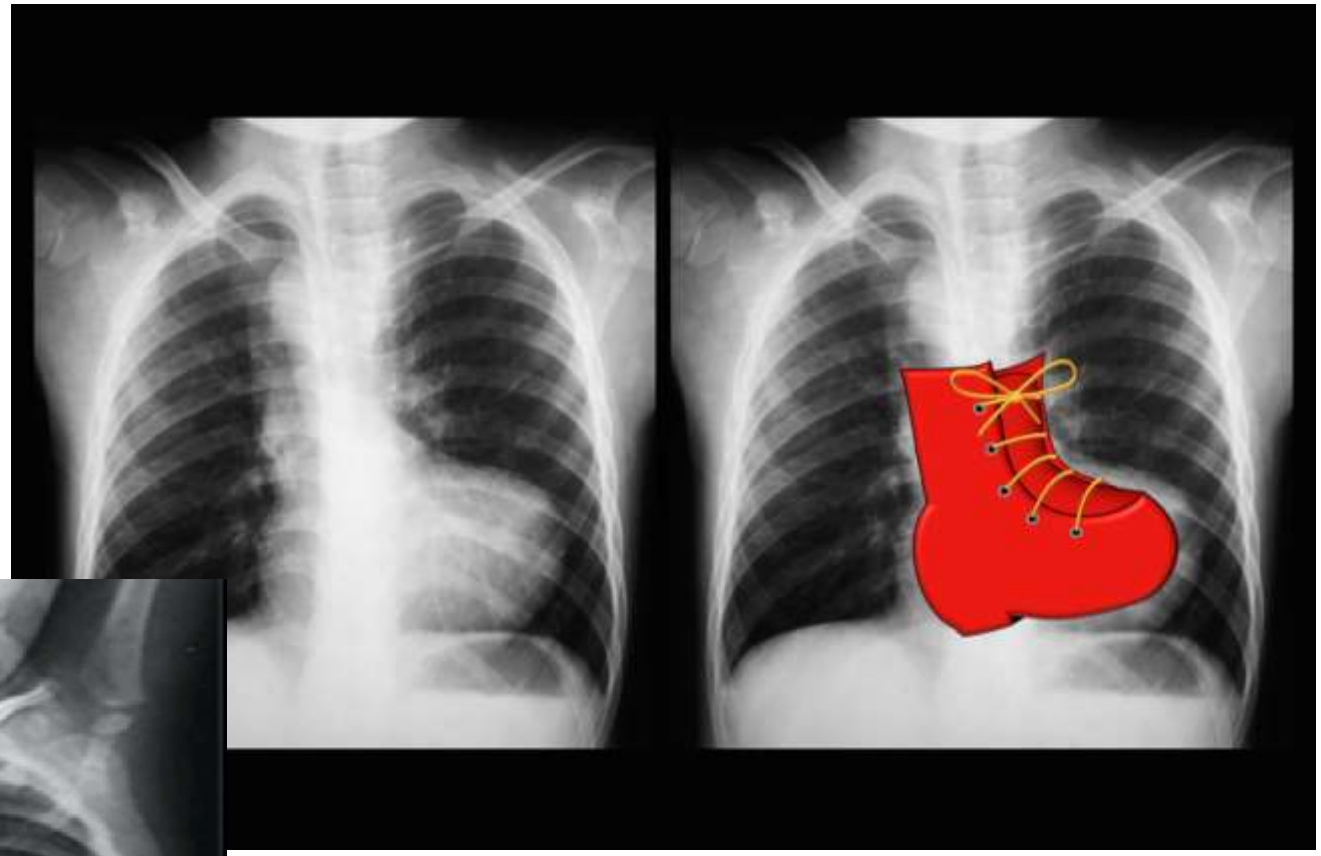
- Mild degree of RV out flow obstruction may present by **congestive heart failure due to ventricular septal defect**
- **By time** RV hypertrophy increase and out flow become more narrow and cyanosis will appear in mucous membrane +lips + mouth +finger nails + toe nails
- **FTT +dyspnea**
- **Hyper cyanotic spells** after exercise/cry and squatting position.
- **Clubbing**
- Chronic hypoxia –Polycythemia - Thrombosis(CVA)
- Infective endocarditis-Cerebral abscess - Hemiplegia

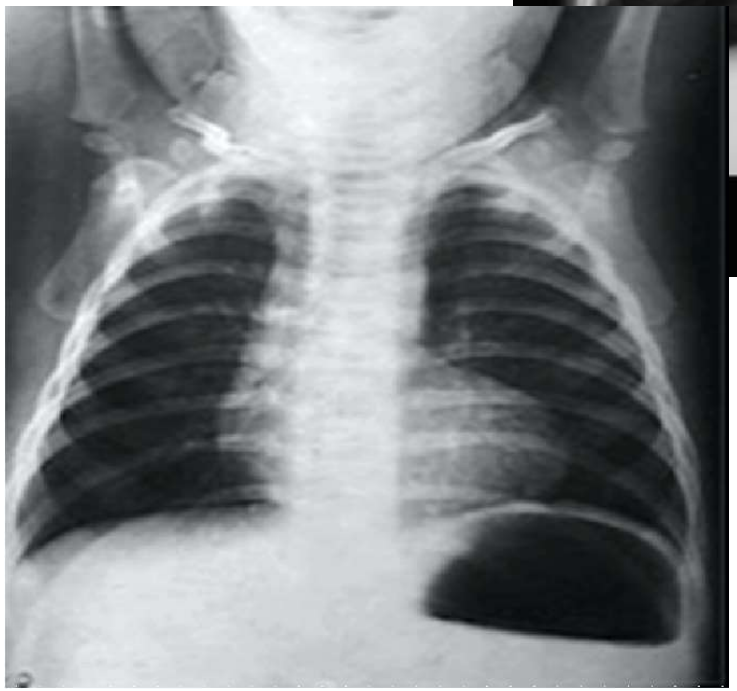


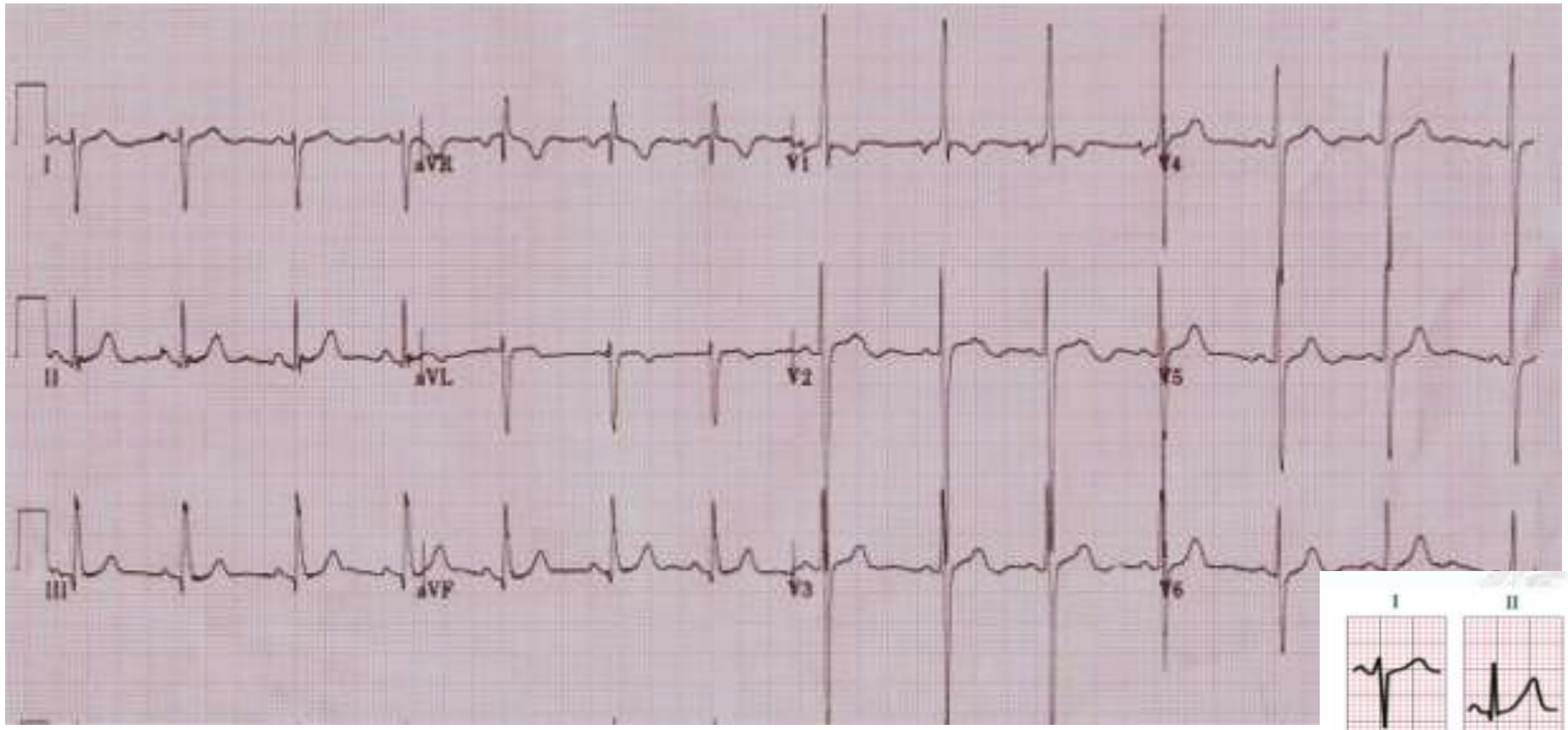
- **Long loud ejection systolic murmur(Due to PS)**
  - No PSM!
  - During cyanotic episodes murmur is inaudible

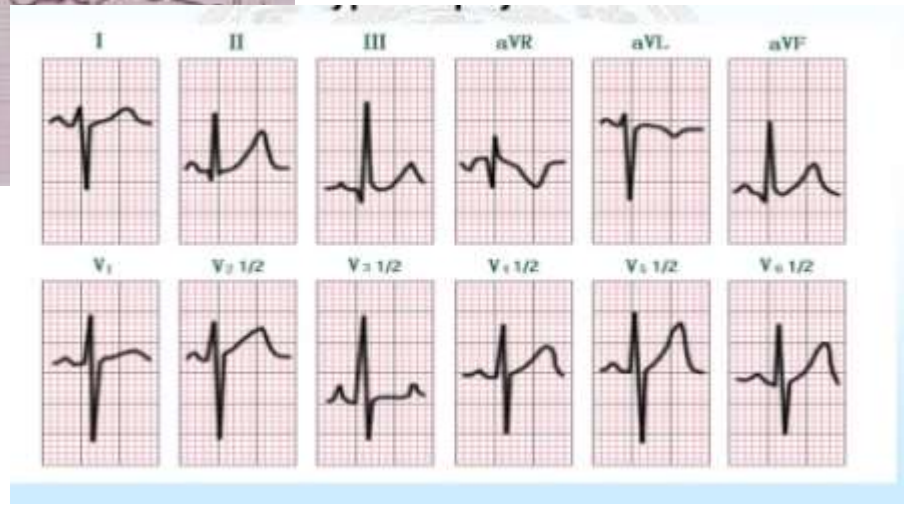
Chest X-Ray

- Cardiac shape like wooden shoe
- Large aorta
- Oligemic lungs









Right axis deviation.
RVH.

TOF: Complications

- **Paroxysmal Hypercyanotic Attacks**
- **Cerebral thrombosis** due to polycythemia or dehydration
- **Brain abscess** (headache+ nausea + vomiting +convulsion)
- **Bacterial endocarditis**

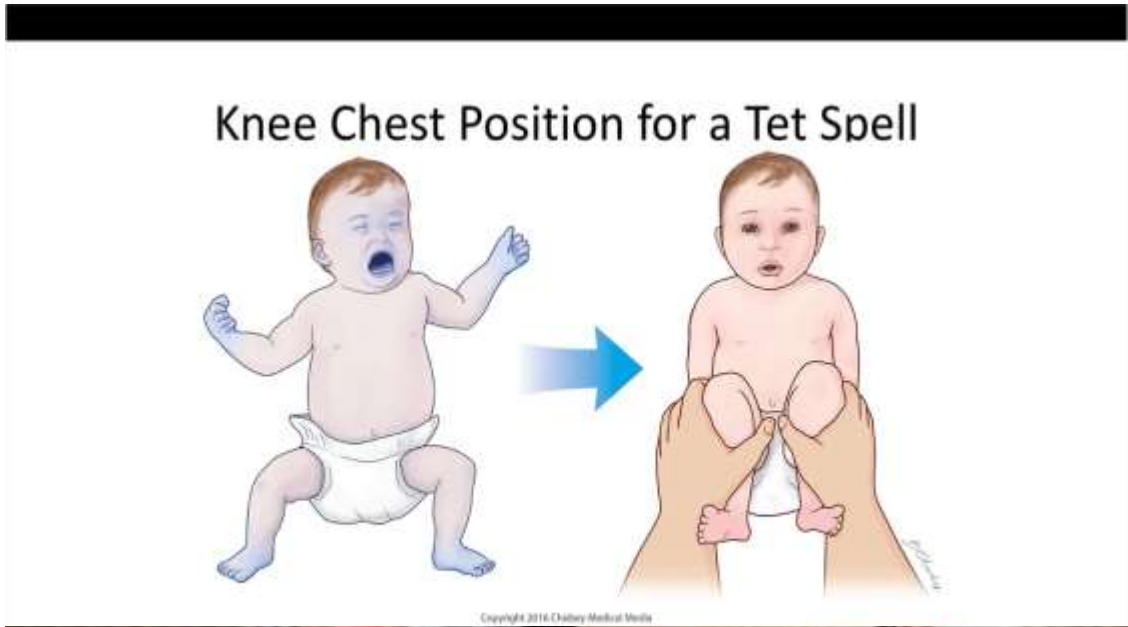
Paroxysmal Hypercyanotic Attacks

- **Known as hypoxic or TET spells**
- In **1st 2 ys of life**
- The child become hyperpnenic and restless -increased cyanosis -gasping – shock.
- Most frequently in the morning or with vigorous crying
- The murmur disappear due to diminished blood flow to the lungs
- The spells lasts from few min to hours rarely fatal
  - Short spells followed by generalized weakness and sleep
  - Severe spells progress to unconsciousness convulsion +hemi paresis
- Decrease in pulmonary blood flow will lead to hypoxia and acidosis

Management of Hypercyanotic Spell

Usually self-limiting

- If prolonged need Rx
  - **Calming** the infant, placing the infant in **the knee-chest position**
    - Increases systemic vascular resistance
  - **Oxygen** is usually administered
    - A pulmonary vasodilator and a systemic vasoconstrictor.
  - **Intravenous bolus of fluid**

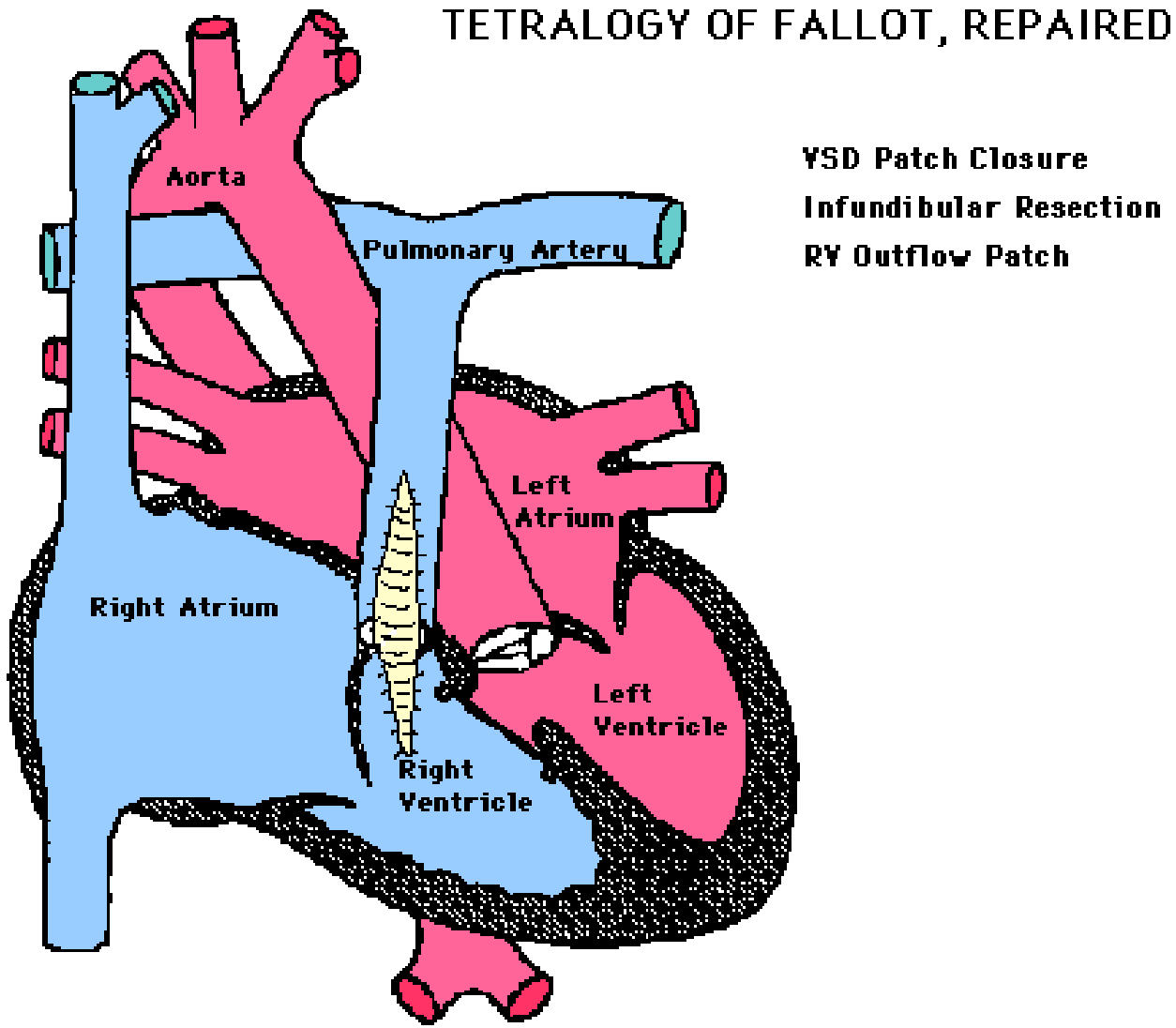


Management of Hypercyanotic Spell (Continue)

- **Morphine sulfate, 0.1 mg/kg** –S/C or IM, suppresses the respiratory center and abolishes hyperpnea
- **Acidosis** should be treated with **sodium bicarbonat**e (NaHCO3), 1 mEq/kg , IV. The same dose can be repeated in 10 to 15 minutes.NaHCO3 reduces the respiratory center-stimulating effect of acidosis.
- **Propranolol** or **esmolol** (0.1 mg/kg) administered by slow IV push
  - -Relaxation of the right ventricular outlet tract
- **Phenylephrine-**Vasoconstrictor , 0.005- 0.02 mg/kg bolus followed by infusion
  - - Increases systemic afterload
- **Ketamine**, 1 to 3 mg/kg (average of 2 mg/kg) ,IV over 60 seconds, works well.
  - -Increases the systemic vascular resistance and sedates the infant

Surgical correction of TOF

- **In NB: PGE1 to keep the ducts open, it will cause relaxation of the smooth muscles of the ductus arteriosus.**
- **Total surgical correction consists of two main steps: closure of the VSD with a patch and reconstruction of the right ventricular outflow tract.**
- **TOF is typically treated by open heart surgery for total repair electively in the first year of life (the best time is Complete Repair at age of 4-6 months but depending on pulmonary artery anatomy and symptoms and size of the baby)**



Tricuspid atresia

It represents approximately 2%-3% of all CHDs.

It is **the 3 rd most common cyanotic cardiac condition.**

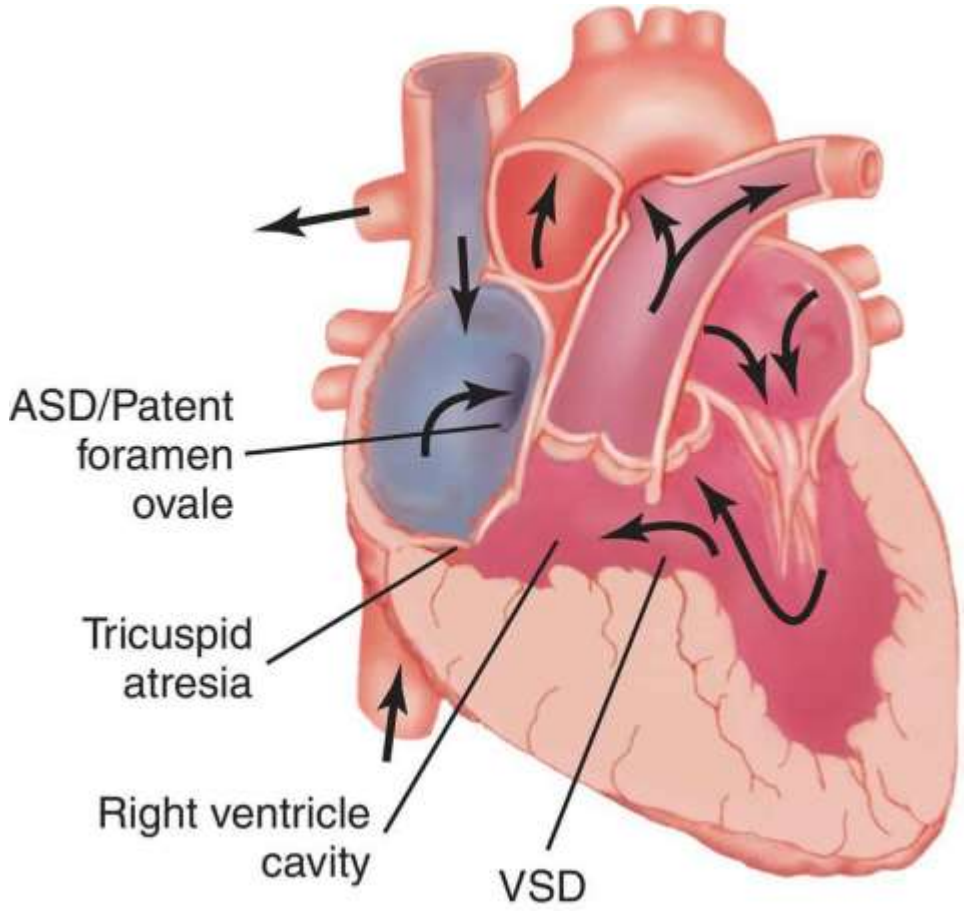
It is a complex lesion with many variations.

<u>In this lesion:</u>

The tricuspid valve does not develop.

An ASD or patent foramen ovale must be present for the fetus or infant to survive.

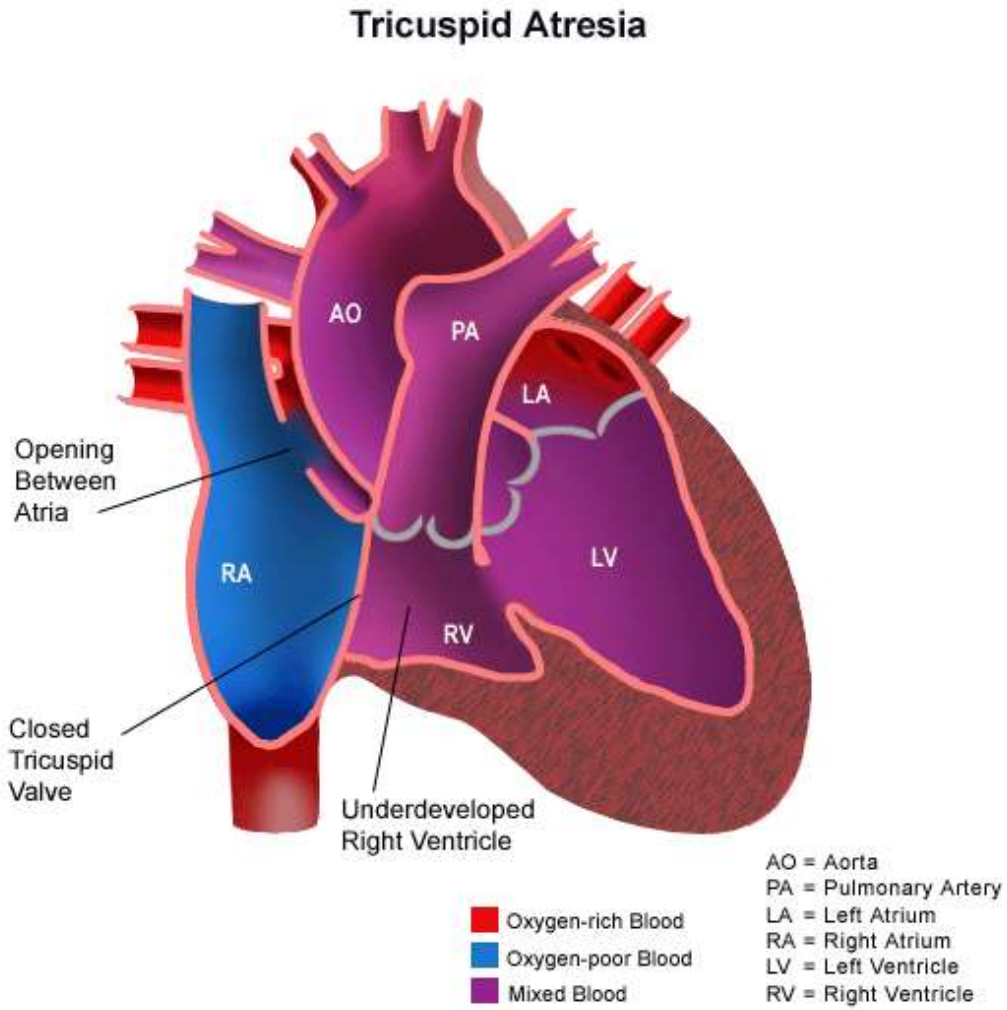
The RV is hypoplastic (underdeveloped).



- The Newborn may depend on **the ductus arteriosus** for pulmonary blood flow.

- Tricuspid atresia is usually classified according to the presence or absence **of PS and TGA.**

- The great arteries **are normally related in about 70% of cases and are transposed in 30%**.

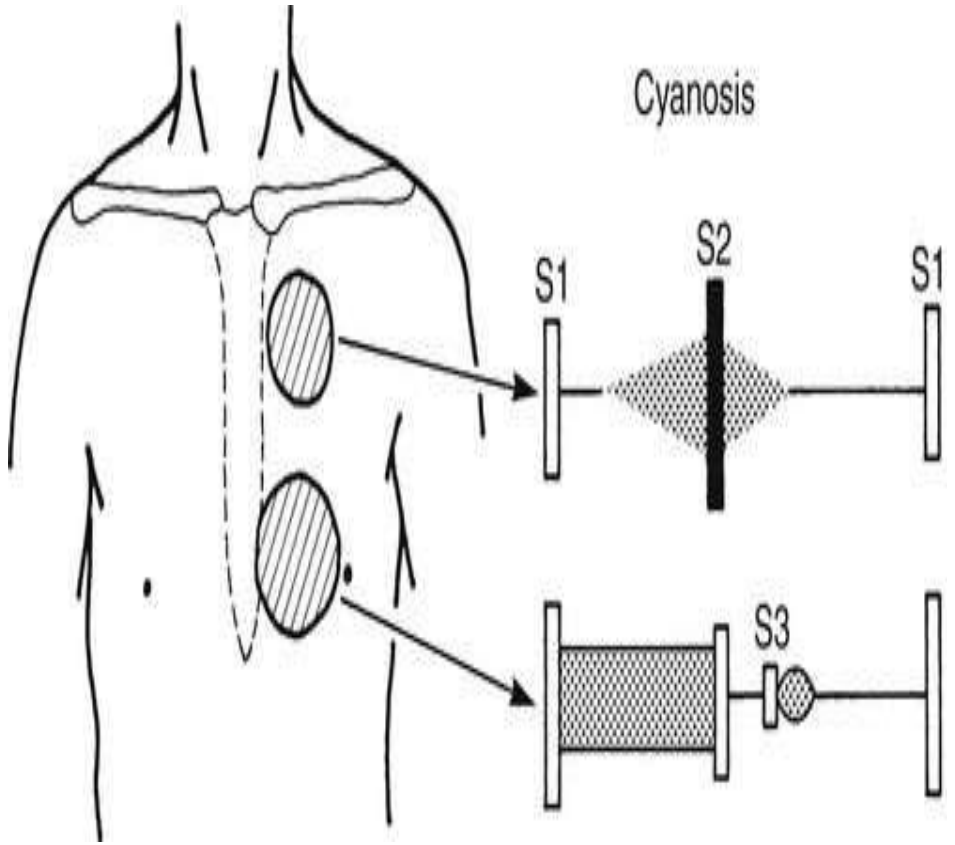


CLINICAL MANIFESTATIONS

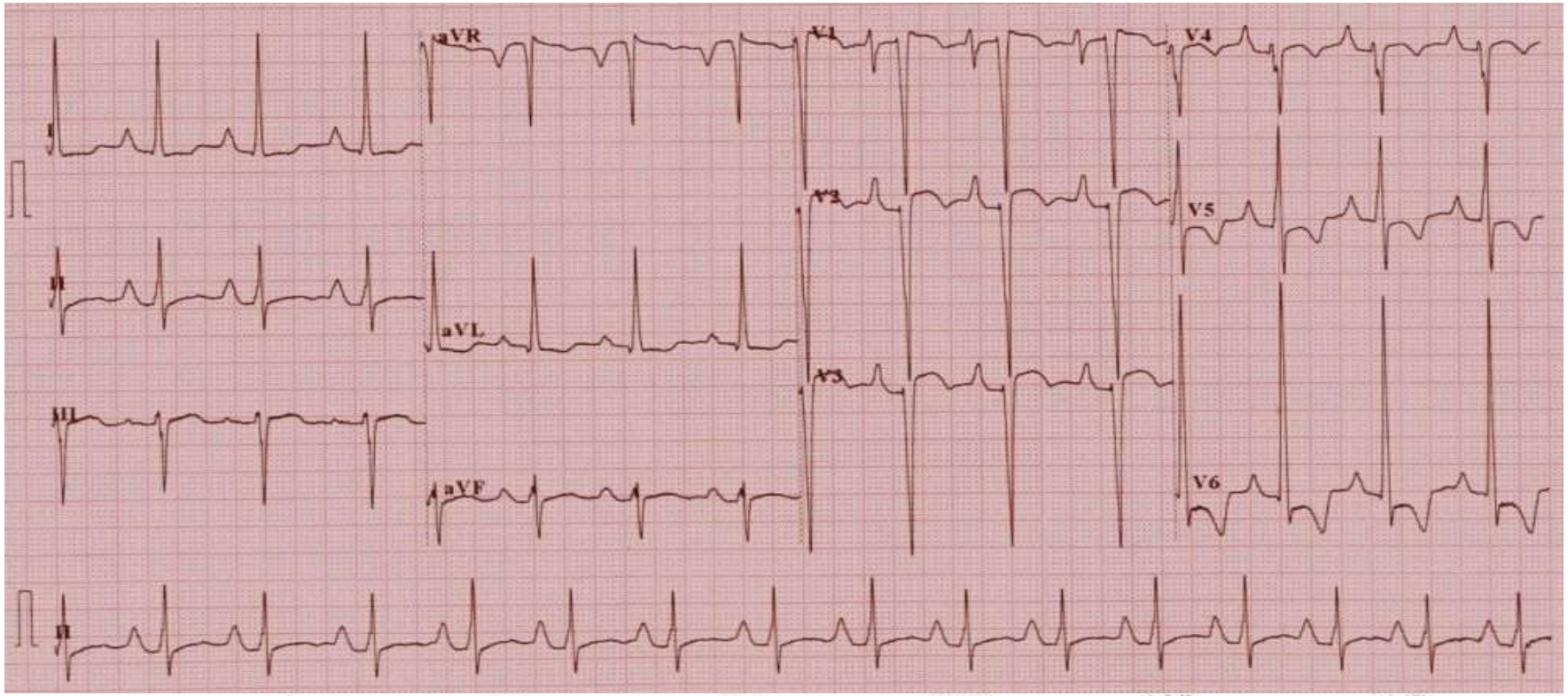
Cyanosis is usually severe from birth.

On Physical Examination :

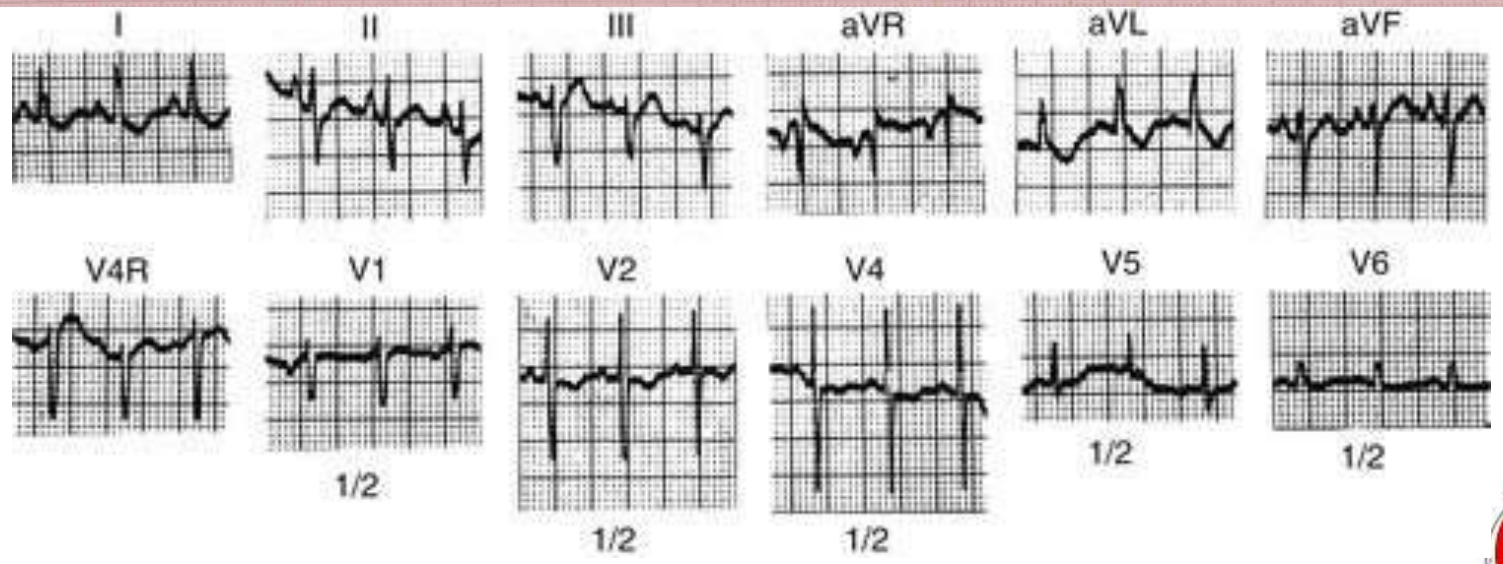
- **Cyanosis is always present.**
- A systolic thrill is **rarely palpable** when associated with PS.
- **Hepatomegaly** may indicate an inadequate interatrial communication or CHF.
- **Auscultation :**
  - **The S2 is single**.
  - **A grade 2-3/6 systolic murmur of VSD** is **usually present** at the lower left sternal border.
  - A continuous murmur of PDA is **occasionally present**.



ECG Findings:

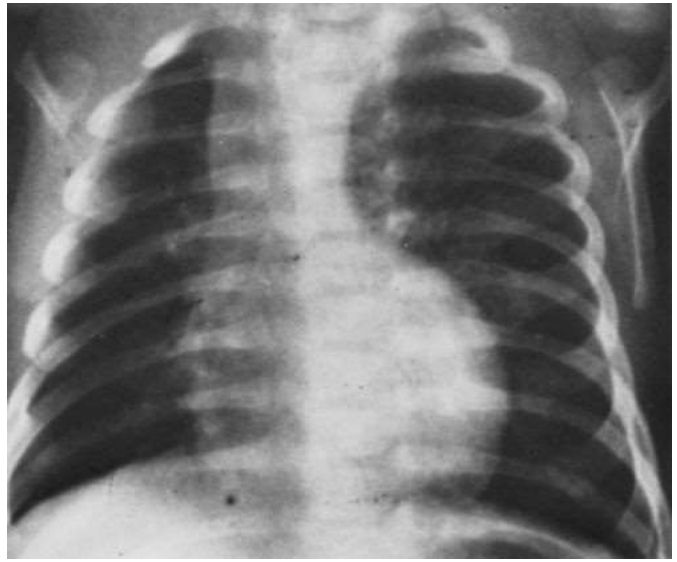


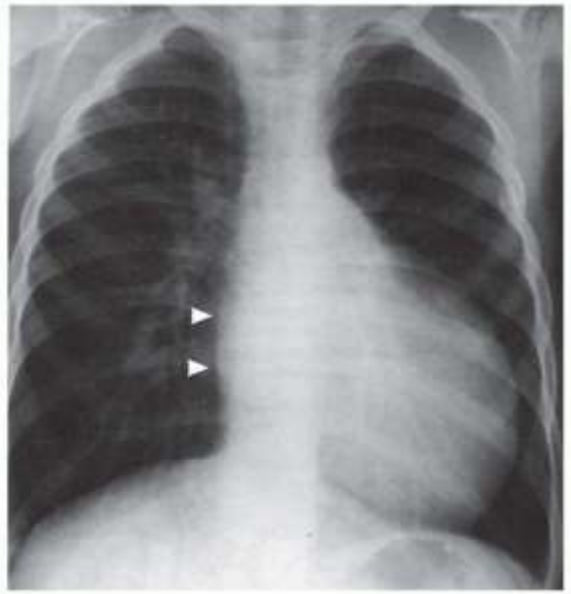
- Left “Superior” QRS axis (between 0 and -90 degrees) is characteristic.
- LVH is usually present
- RA dilatation or combined atrial Dilatation (CAH) is common.

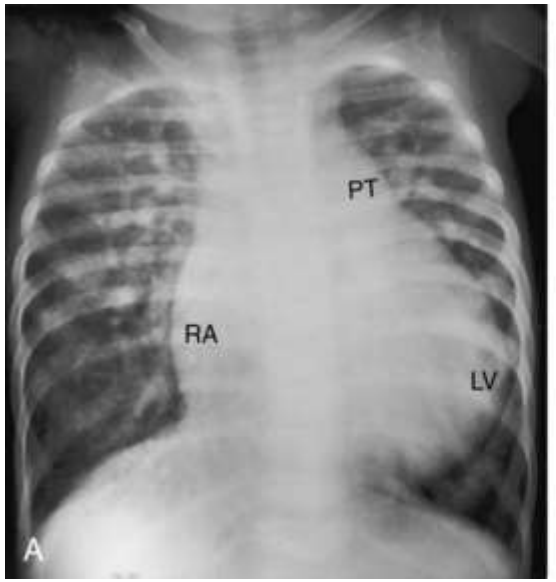


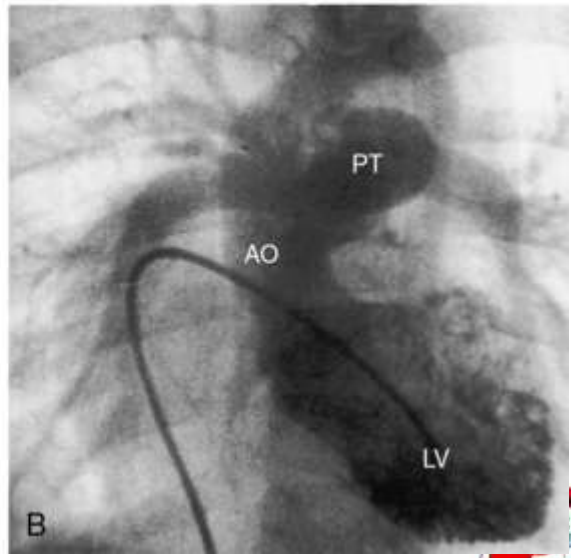
Chest X-ray findings:

- **The heart size is normal or slightly increased**, with enlargement of the RA and LV.
- **Pulmonary vascularity decreases in most patients,** although it may increase in infants with TGA.
- **Occasionally**, the concave PA segment may produce a boot-shaped heart, like the x-ray findings of TOF.





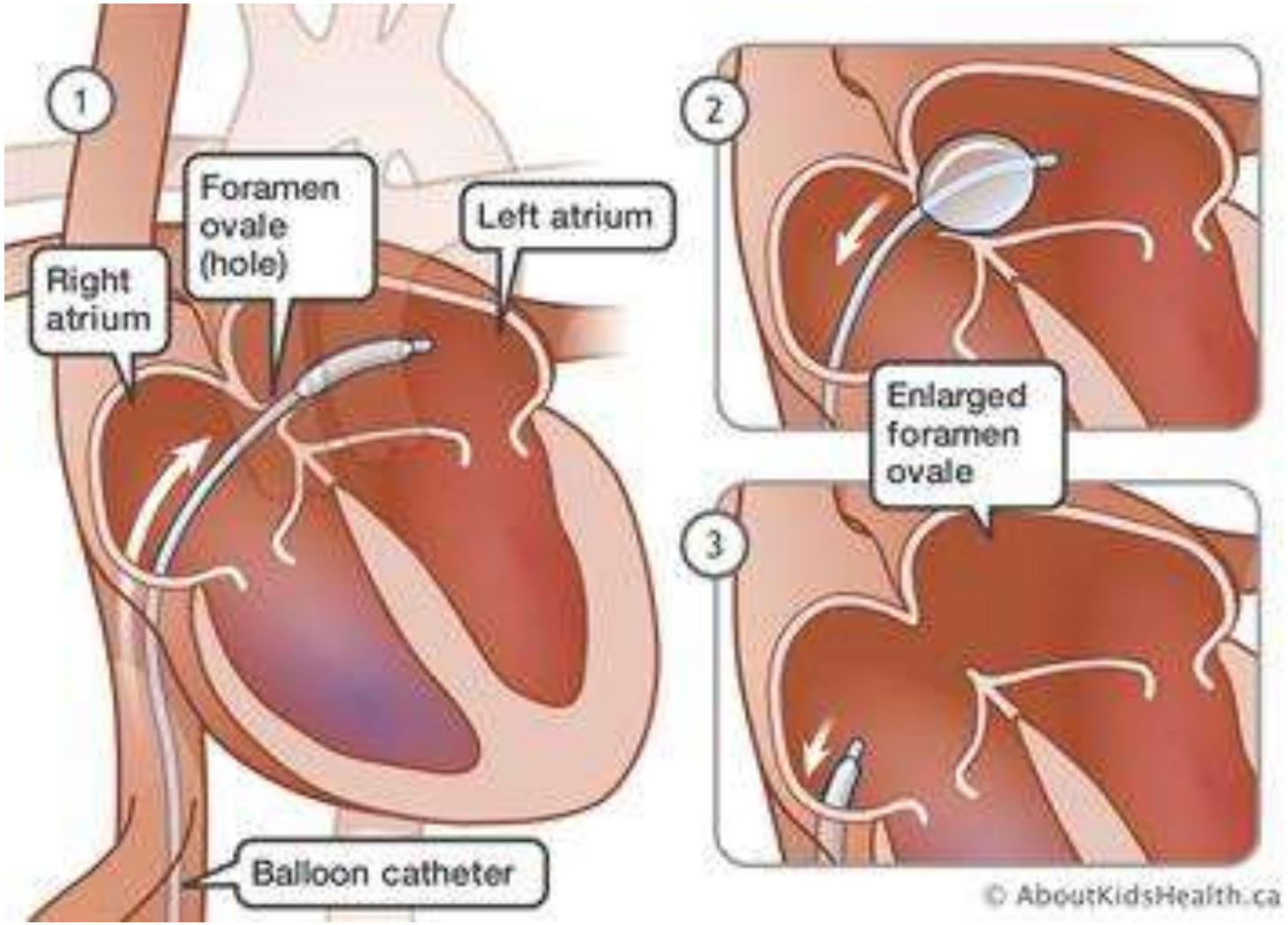




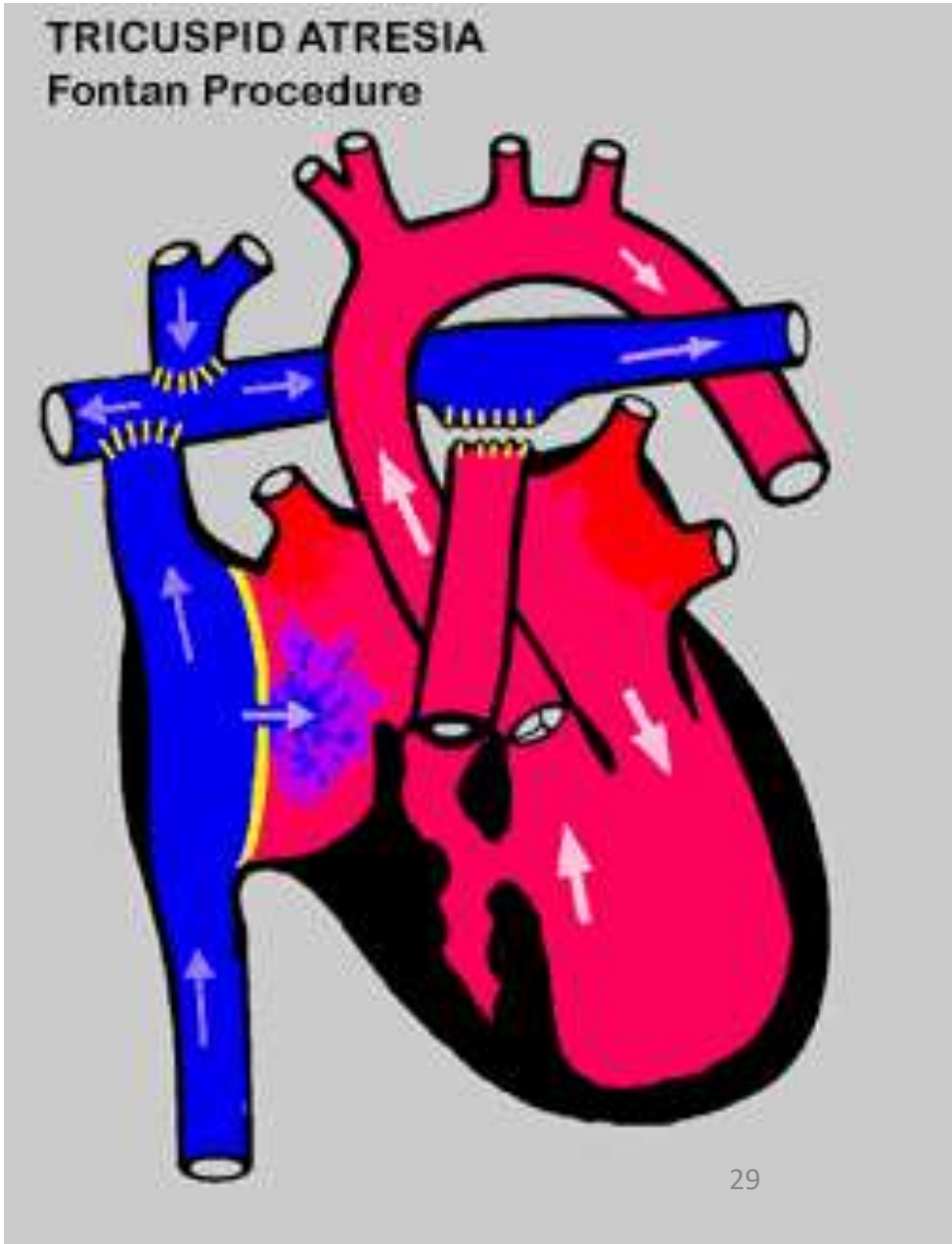
Management of TA

- **Medical:**
  - Prostaglandin E1 should be started in neonates with severe cyanosis to maintain the patency of the ductus before planned cardiac catheterization or cardiac surgery.
- **SURGICAL- SINGLE VENTRICLE STAGED CORRECTION:**
  - **Shunt or pulmonary bandage**
  - **Bidirectional Glenn shunt**
  - **The modified Fontan operation**

- **The Rashkind procedure (balloon atrial septostomy) to improve the RA-to-LA shunt, especially when the interatrial communication is considered inadequate by echo studies.**

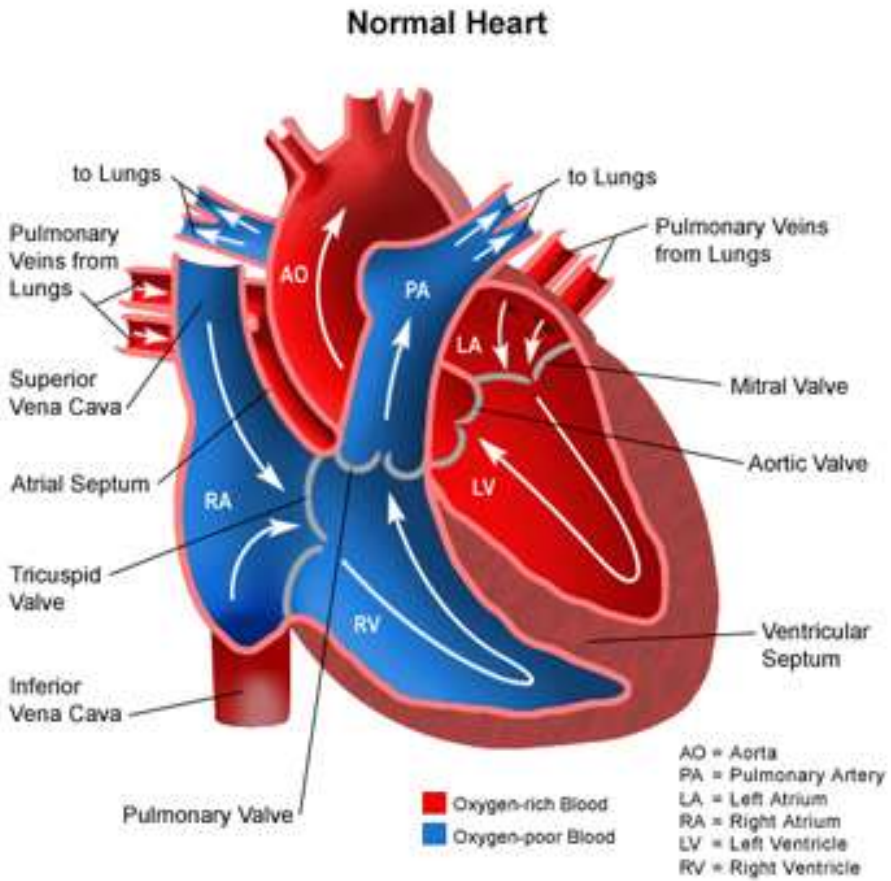


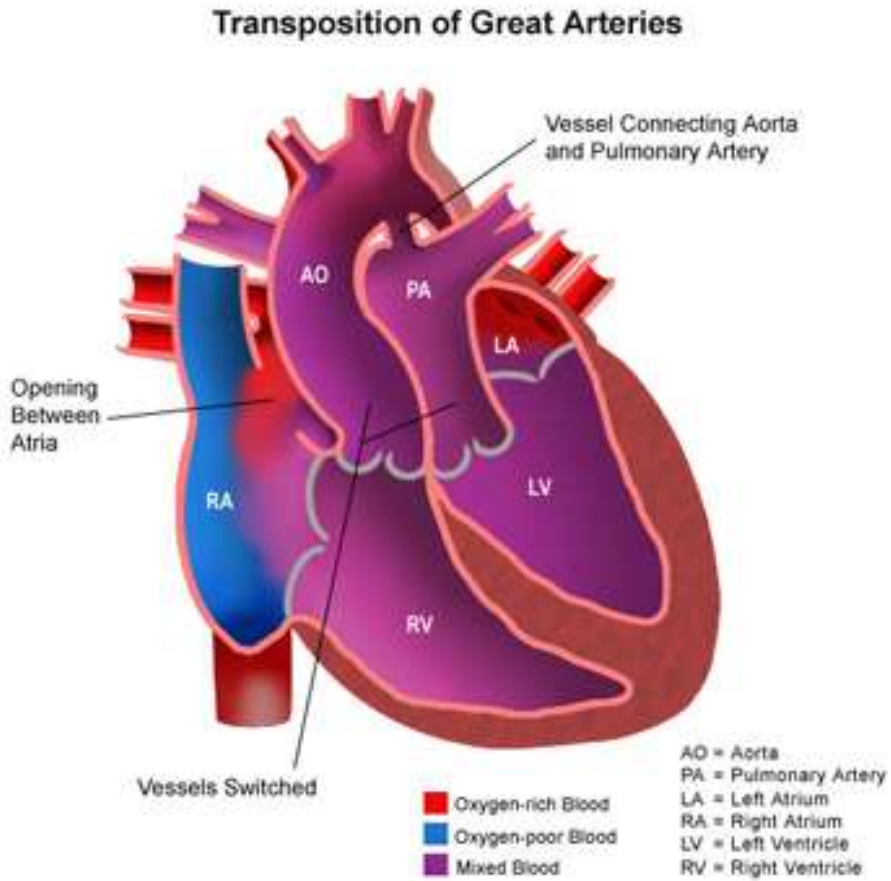
- **Shunt or pulmonary bandage**
- **Bidirectional Glenn shunt:**
  - the creation of an anastomosis between the superior vena cava and the pulmonary arteries.
  - Usually between 3 and 6 mo. of age.
  - To reduces the volume load on the left ventricle and may decrease the chance of left ventricular dysfunction developing later in life.
- **The modified Fontan operation**
  - Usually performed between 1.5 and 3 yr of age,
  - Was performed by anastomosing the right atrium or atrial appendage directly to the pulmonary artery, and now is performed by anastomosing IVC to the pulmonary artery.

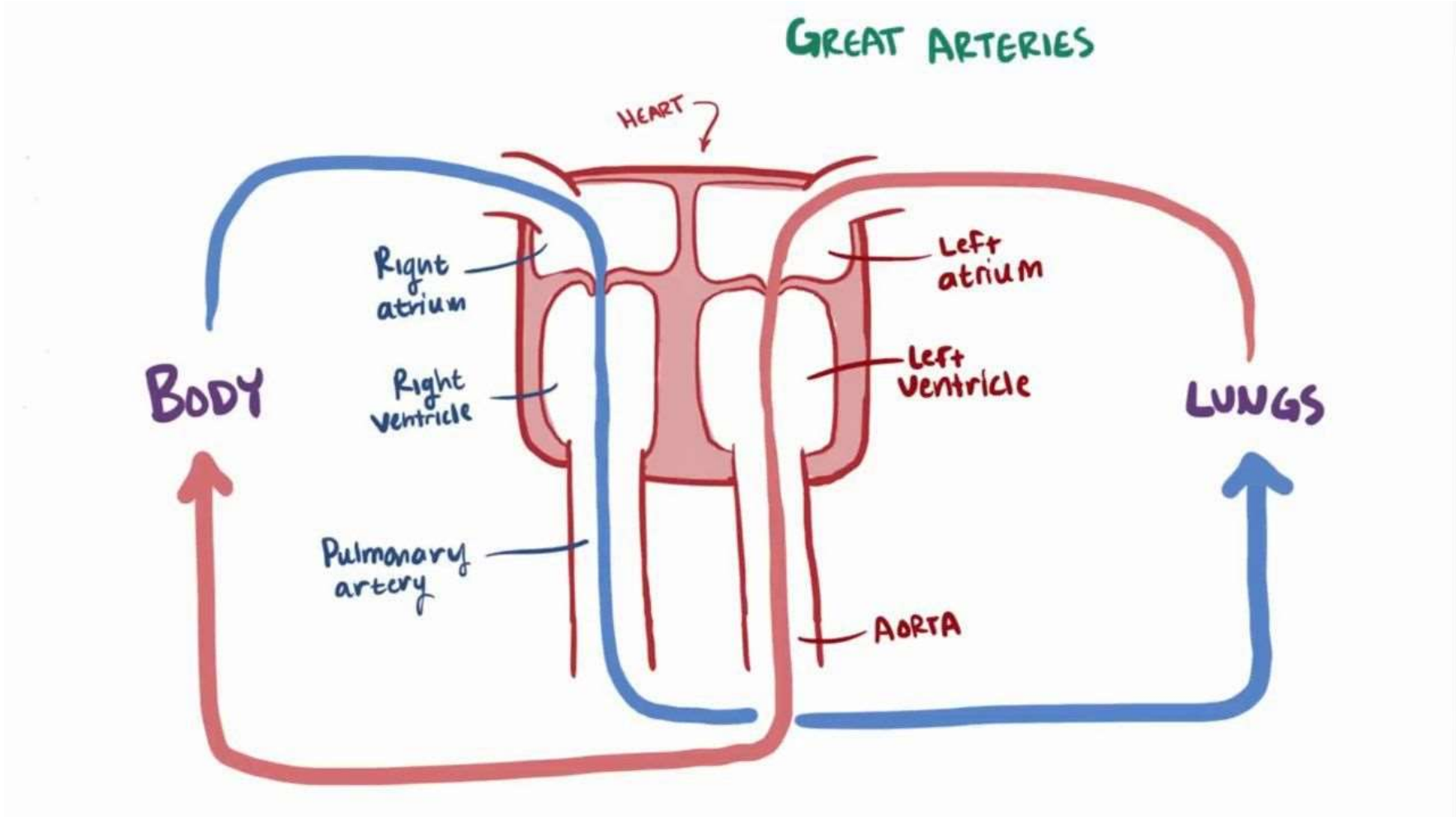


D-transposition of the Great Arteries

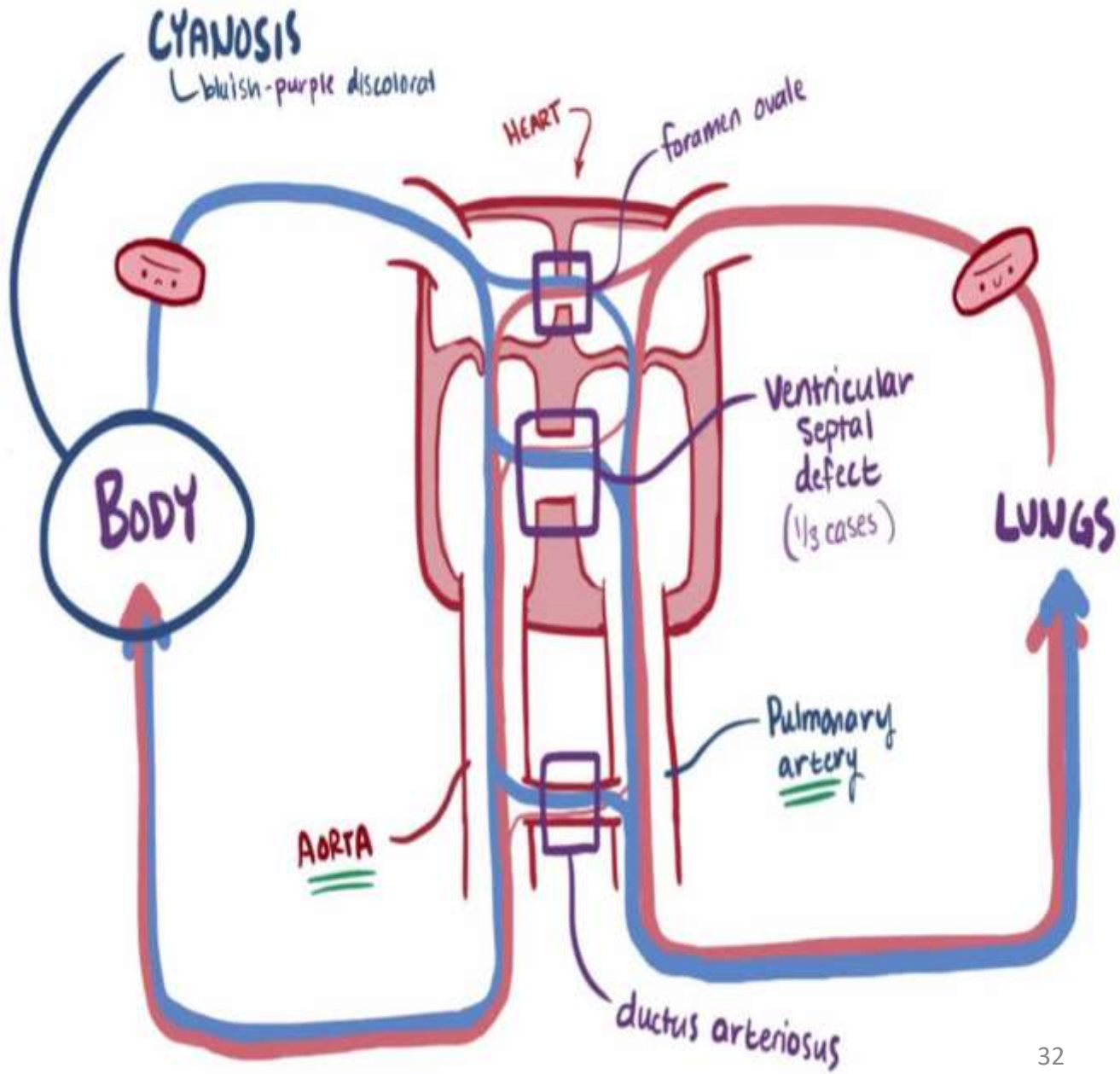
The most common cyanotic defect presents with cyanosis at neonatal period






GREAT ARTERIES
HEART
Right atrium
Right ventricle
Pulmonary artery
Left atrium
Left ventricle
AORTA
BODY
LUNGS

- **The degree of saturation will depend on the degree of mixing of the 2 “parallel” circuits.**
- The mixing sites are: **ASD, PDA, and VSD.**
- The more mixing, the higher the **“effective pulmonary blood flow”**



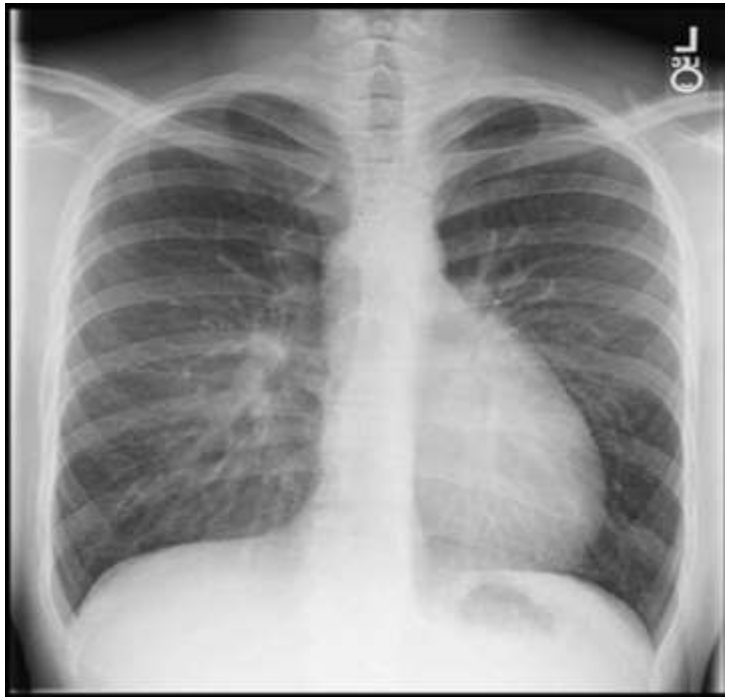
CLINICAL Findings

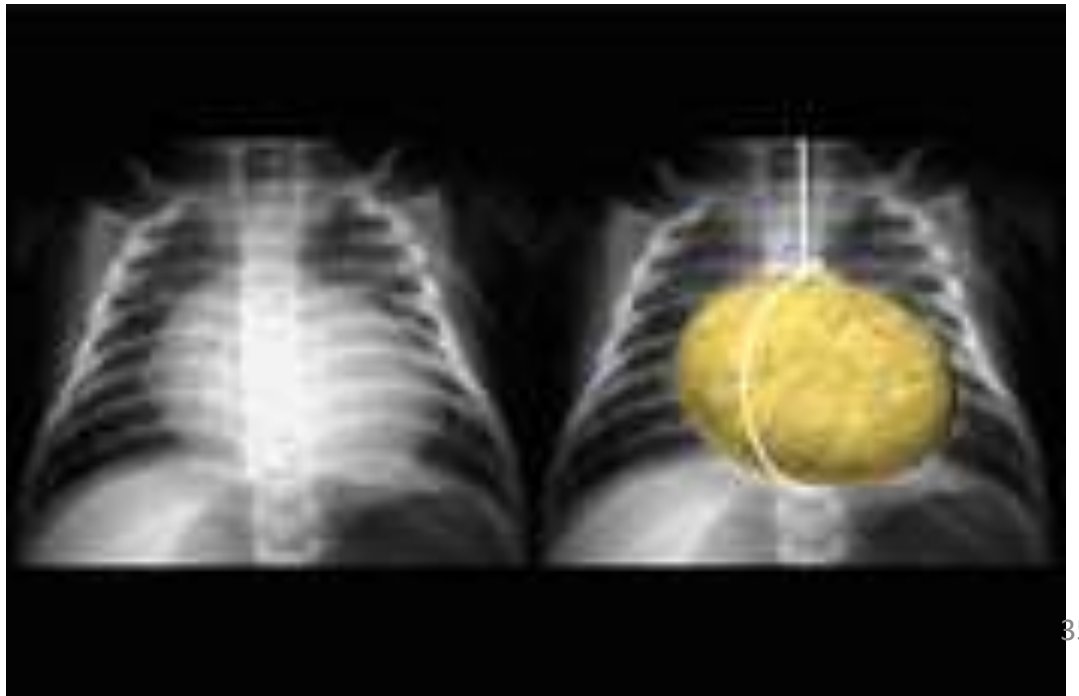
- **History**
  - History of cyanosis from birth **is always present.**
  - Signs of **congestive heart failure (CHF) with dyspnea and feeding difficulties** develop during the newborn period?
- **Physical Examination**
  - **Moderate to severe cyanosis** is present, especially in large, male newborns. Such an infant is tachypnic but without retraction unless CHF supervenes.
  - **The S2 is single and loud**. **No heart murmur is heard** in infants with an intact ventricular septum.
  - **If CHF supervenes, hepatomegaly and dyspnea develop**

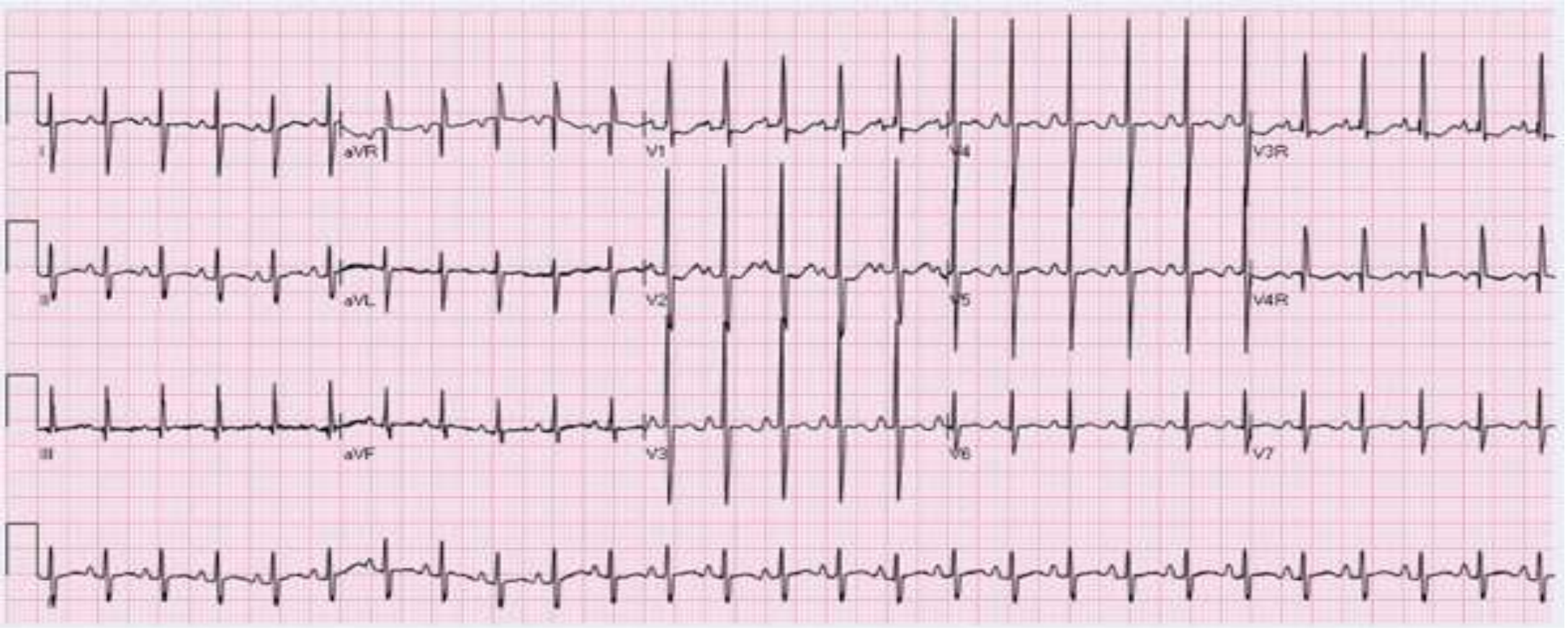
- **Laboratory Studies**
  - **Severe arterial hypoxemia** with or without acidosis is present. Hypoxemia does not respond to oxygen inhalation .
  - **Hypoglycemia and hypocalcaemia** are occasionally present.

Chest X-ray findings:

- Narrow mediastinum due to anterior-posterior orientation of great arteries and small thymus
- Cardiomegaly is present w/ increased pulmonary vascular markings
- d-TGA CXR: “egg on a string”



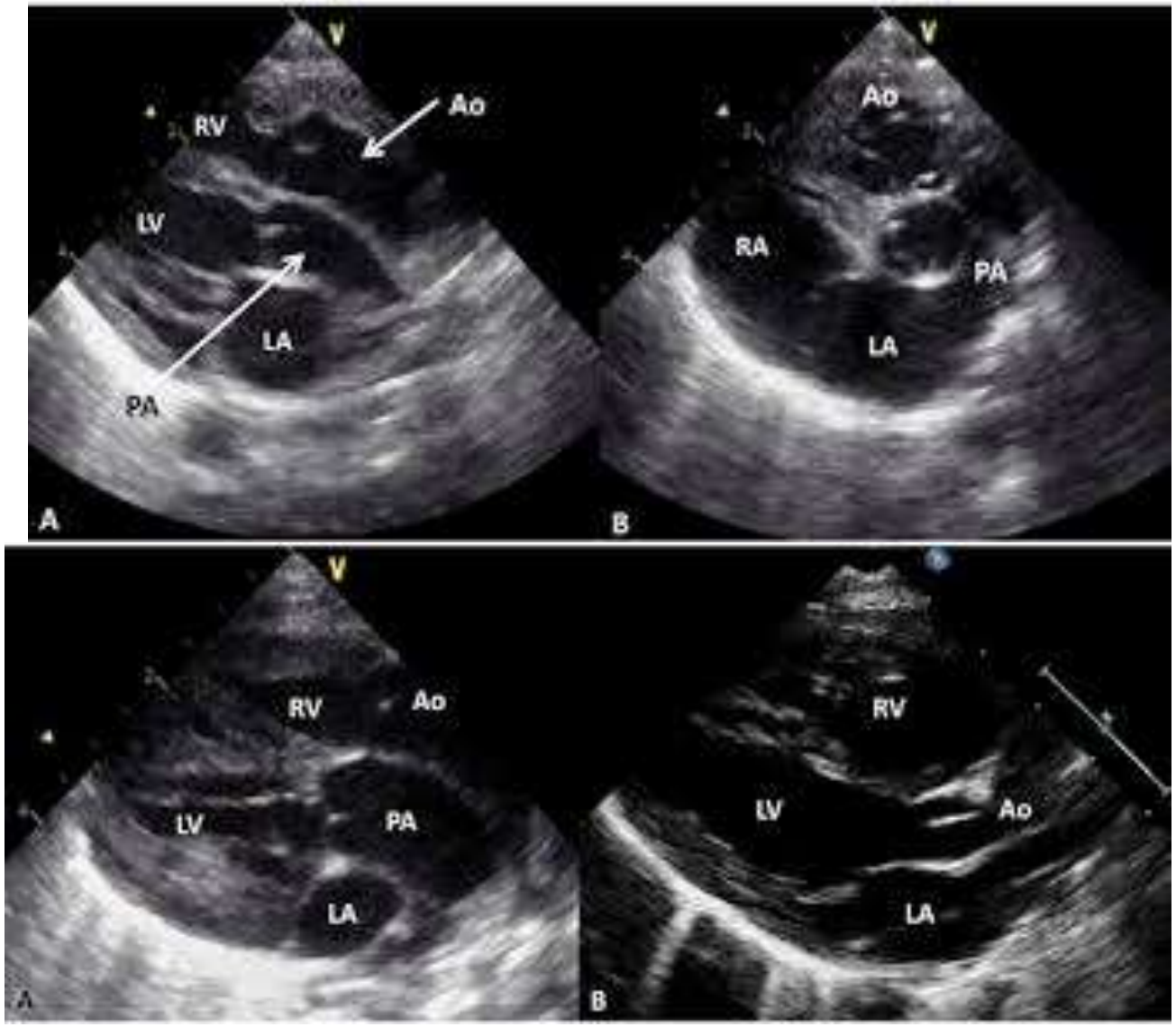




- There is a rightward QRS axis (i.e., +90 to +200 degrees).
- **Right ventricular hypertrophy (RVH)** is usually present after the first few days of life.
- **Combined ventricular hypertrophy (CVH) may be present in infants with large VSD, PDA, or pulmonary vascular obstructive disease**, because all these conditions produce an additional left ventricular hypertrophy (LVH).

Echocardiography

- Two-dimensional echo and color flow Doppler studies usually provide all the anatomic and functional information needed for the management of infants with D-TGA.



Medical RX

Initial management of d-TGA: goal is to improve mixing

The following measures should be carried out before an emergency cardiac catheterization (if performed) or a surgical procedure:

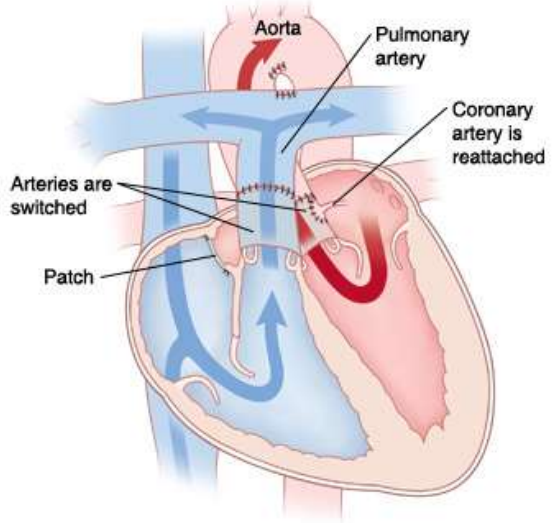
- **Prostaglandin E1 infusion** should be started to improve arterial oxygen saturation by reopening the ductus.
- **Oxygen** should be administered for **severe hypoxia**
- **Metabolic acidosis** should be corrected
- H**ypoglycemia** and **hypocalcemia** should be treated if present.
- **CHF may be treated with digoxin and diuretics**

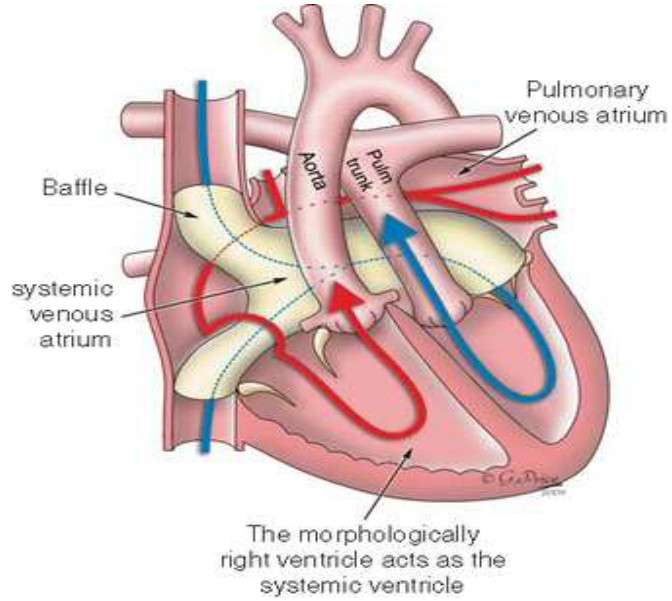
- **Before surgery, cardiac catheterization and a balloon atrial septostomy (i.e., the Rashkind procedure) are often carried out to have some flexibility in planning surgery- <u>if atrial shunt is not sufficient.</u>**

- **An increase in the oxygen saturation of 10% or more and a minimal interatrial pressure gradient are considered satisfactory results of the procedure**

Surgical management of d-TGA:

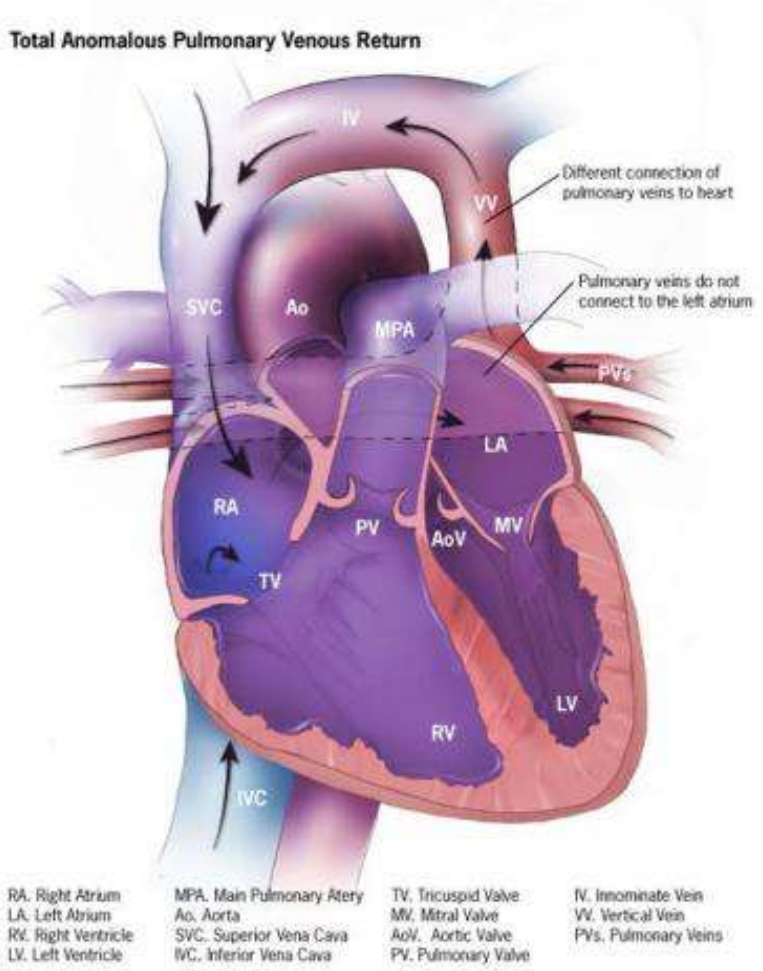
- **Arterial switch procedure –surgical of choice**
  a) The arterial trunks are transected and “switched” to restore “normal” anatomy
  b) The coronary arteries are resected and re-implanted.
- There are other types of surgical correction of d-TGA depends on many clinical and anatomical factors as:
  - Intra-atrial repair surgeries such as the **Senning , Mustard operation**
  - **Nikaidoh** operation, or others
  - **REV**





Total Anomalous Pulmonary Venous Return(TAPVR)

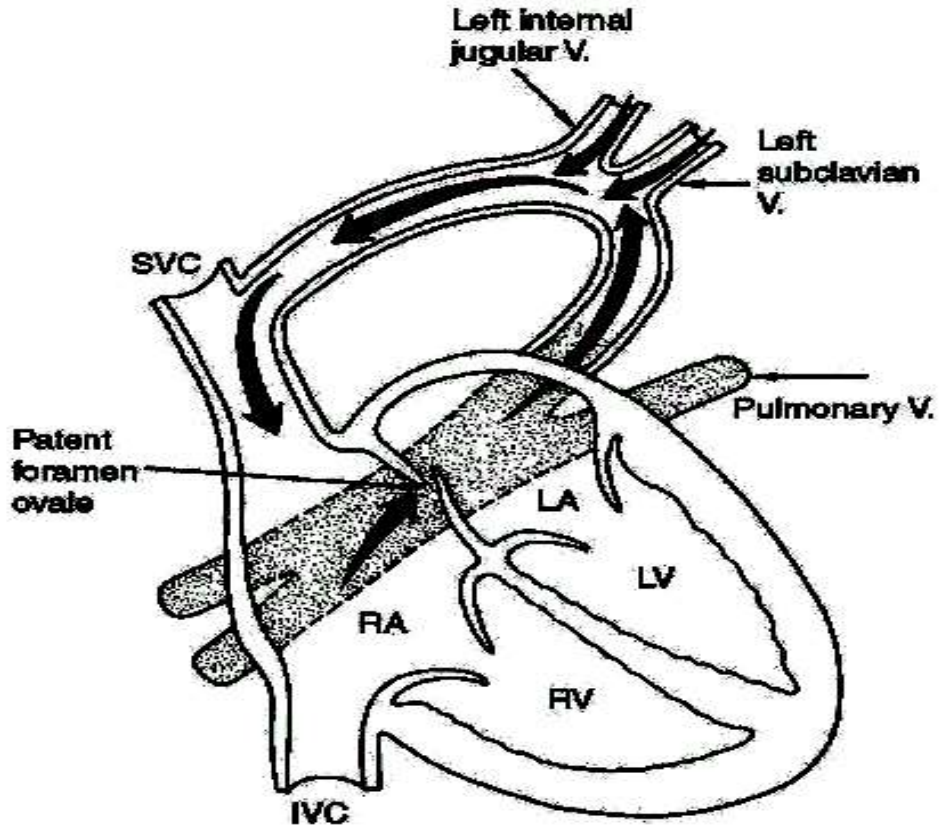
- **TAPVR accounts for 1% of all congenital heart defects.**
- **No direct communication exists between the pulmonary veins and the LA. Instead, they drain anomalously into the systemic venous tributaries or into the RA.**



PATHOLOGY

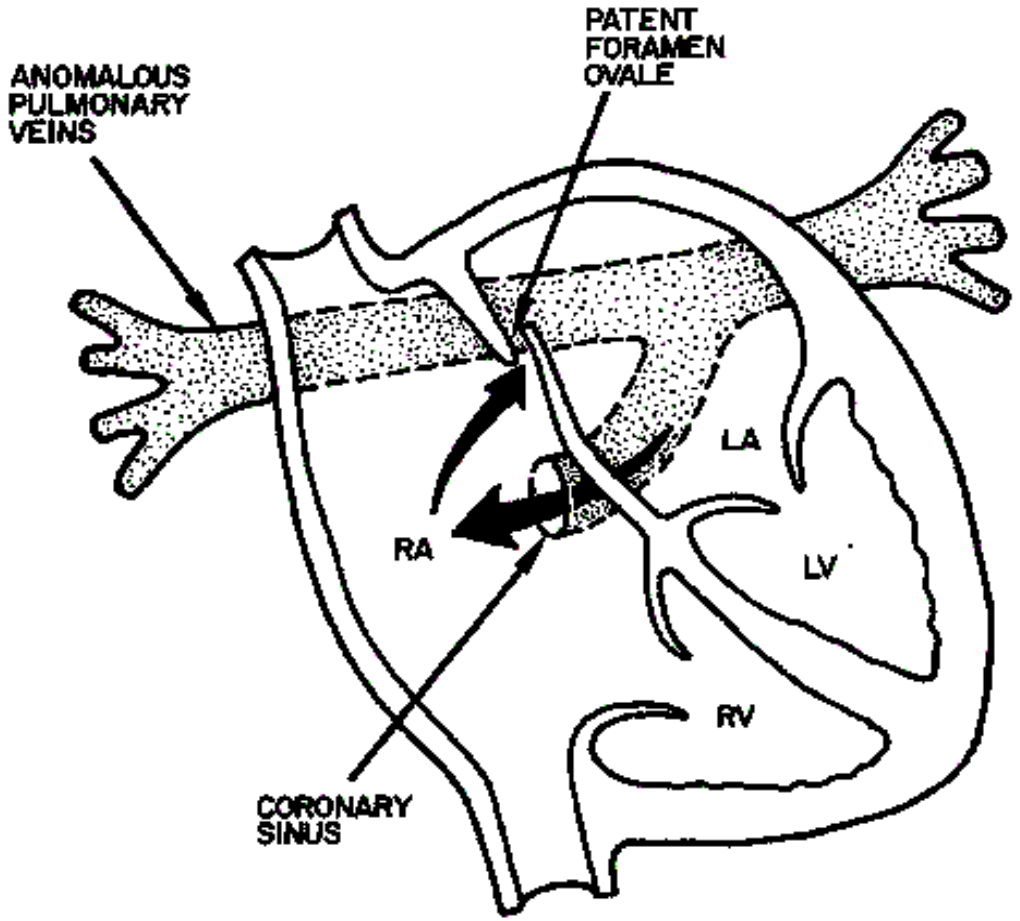
- Depending on the drainage site of the pulmonary veins, the defect may be divided into the following four types :

  - **Supracardiac:**
    (50% of TAPVR patients).
    The common pulmonary venous sinus drains into the right SVC through the left vertical vein and the left innominate vein



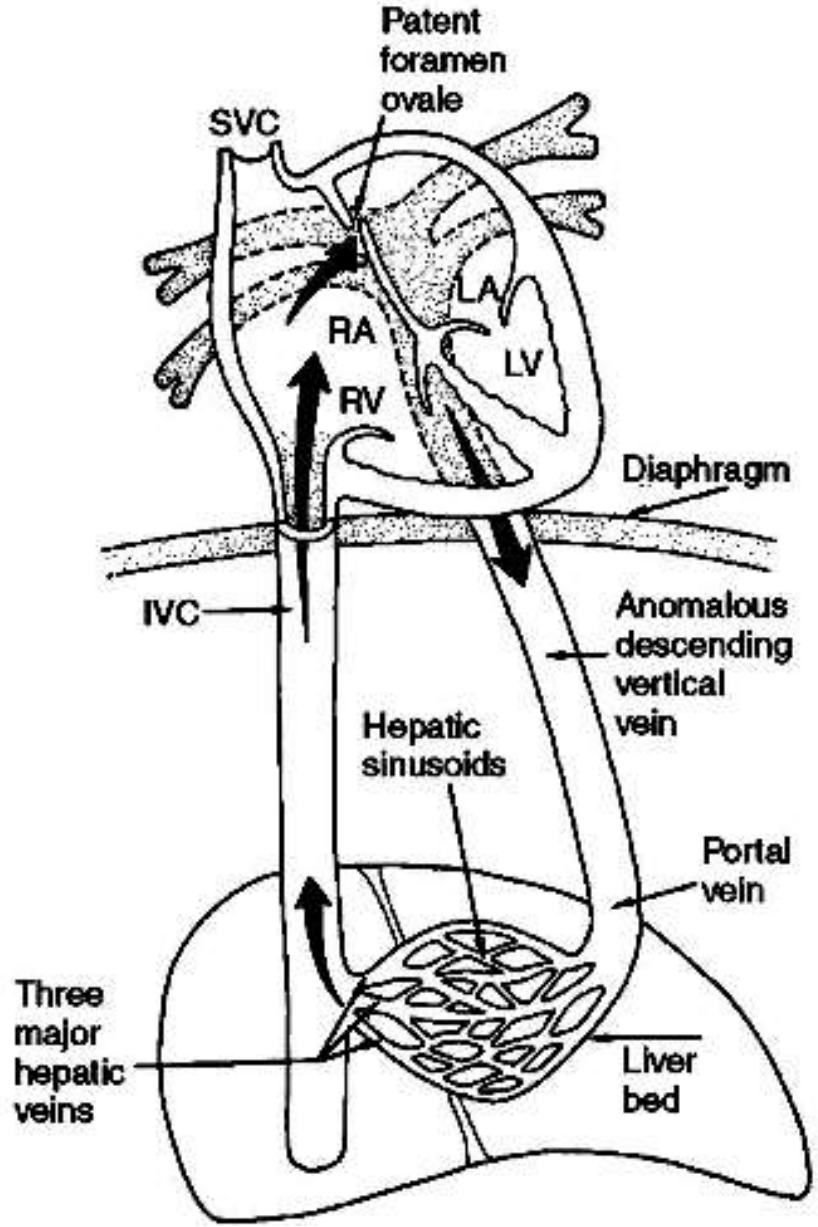
- **Cardiac:**

  (20% of TAPVR patients). The common pulmonary venous sinus drains into the coronary sinus or right atrium.

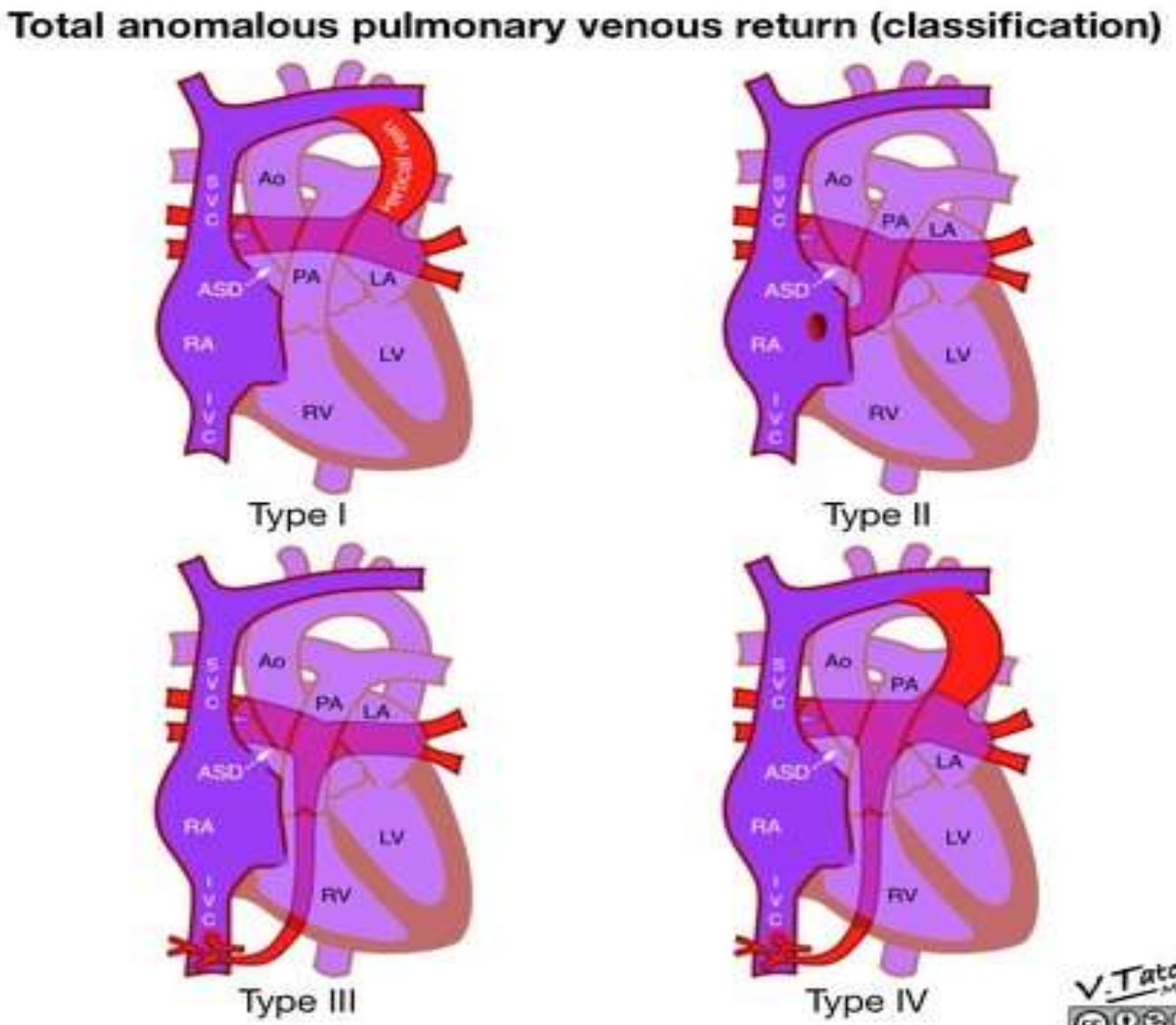


- **Infracardiac (subdiaphragmatic):**

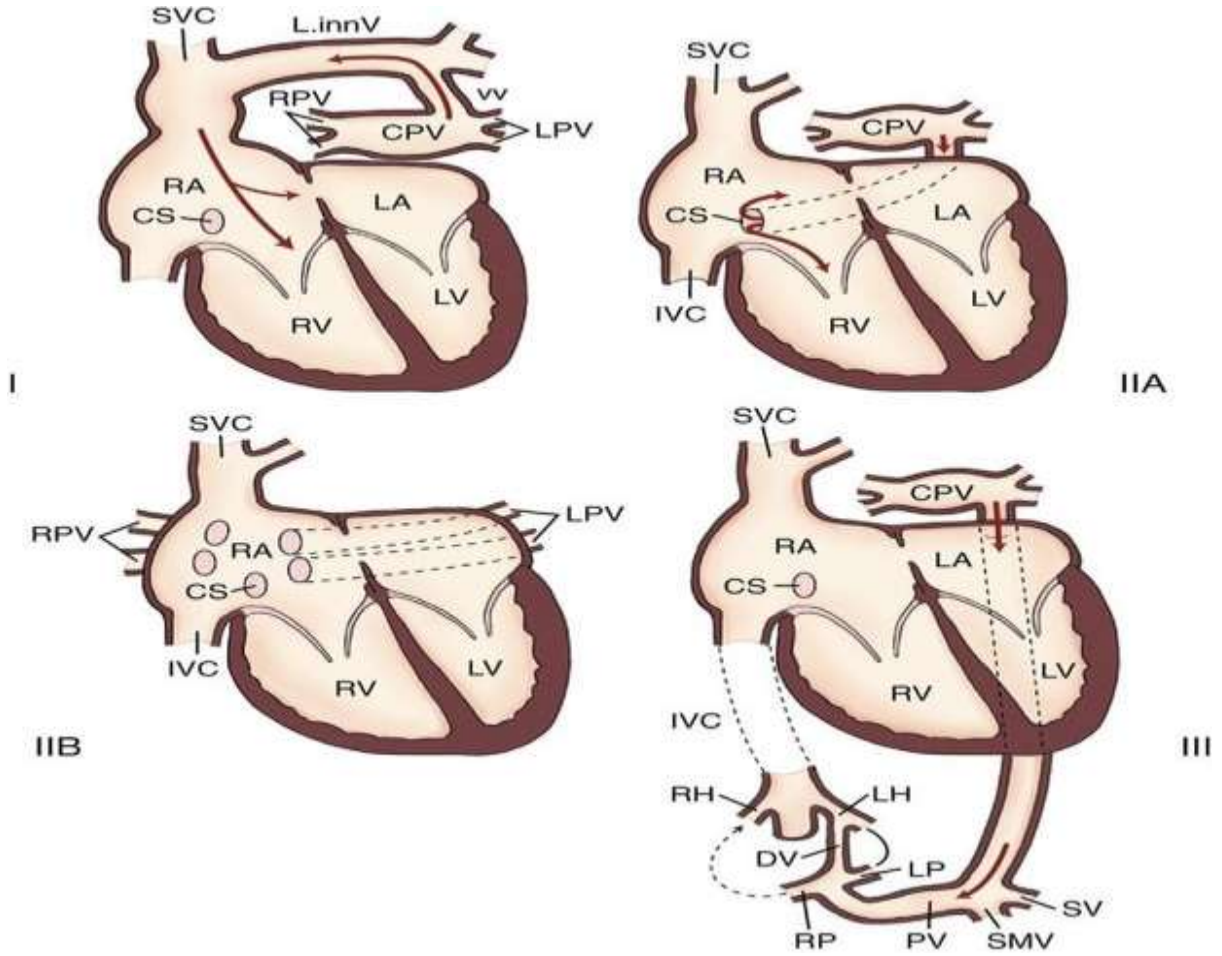
  (20% of TAPVR patients). The common pulmonary venous sinus drains to the portal vein, ductus venosus, hepatic vein, or inferior vena cava (IVC).



Mixed type: (10% of TAPVR patients). This type is a combination of the other types.



- An interatrial communication, either an ASD or PFO, is **necessary for survival.**
- The left side of the heart is **relatively small.**
- **<u>Many</u> patients** with supracardiac and cardiac types of TAPVR and **<u>most</u> patients** with the infracardiac type have pulmonary hypertension secondary to **obstruction of the pulmonary venous return.**



CLINICAL MANIFESTATIONS

Clinical manifestations differ, depending on whether there is obstruction to the pulmonary venous return

Without Pulmonary Venous Obstruction

- **History**

  1. **CHF with growth retardation and frequent pulmonary infection are common in infancy.**

  2. A history of mild cyanosis from birth is present.

- **Physical Examination**
  - The infant is undernourished and mildly cyanotic.
  - Signs of CHF (e.g., tachypnea, dyspnea, tachycardia, hepatomegaly) are present.
  - Precordial bulge with hyperactive RV impulse is present.
  - **Characteristic quadruple rhythm is present**.
  - The S2 is **widely split and fixed**, and the **P2 may be accentuated**.
  - A grade 2-3/6 ejection systolic murmur is usually audible at the upper left sternal border.
  - A mid-diastolic rumble is always present at the lower left sternal border

With Pulmonary Venous Obstruction

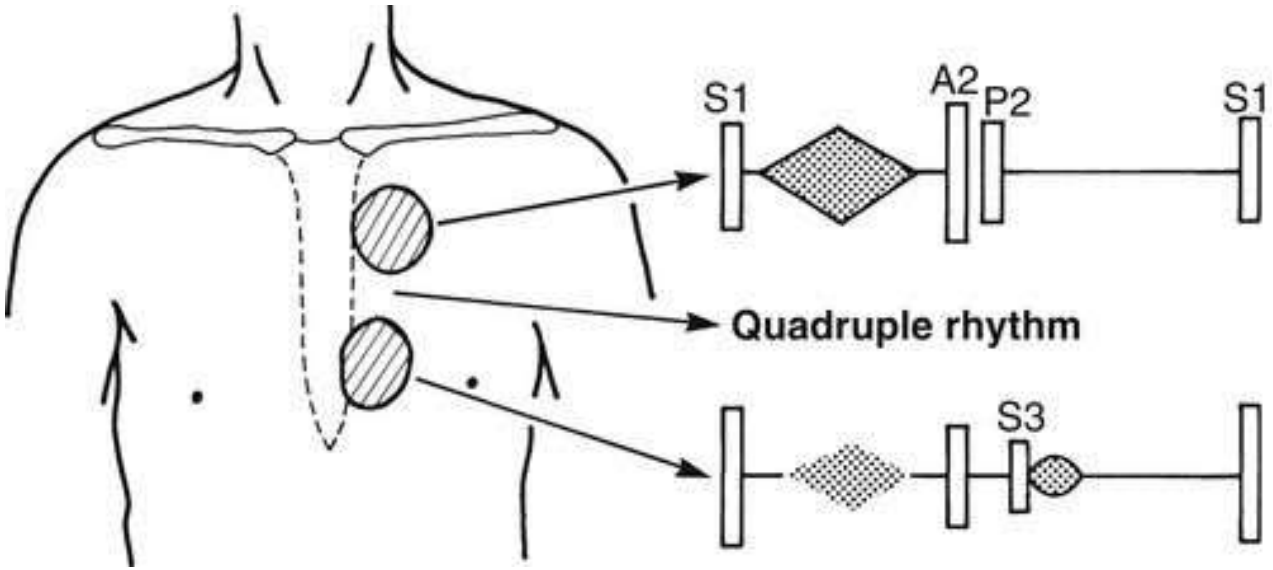
- **History**
  1. Marked cyanosis and respiratory distress develop in **the neonatal period with failure to thrive.**
  2. Cyanosis worsens with feeding, especially in infants with the **infracardiac** type, resulting from compression of the common pulmonary vein by the food-filled esophagus.
- **PHYSICAL EXAMINATION**

1. Moderate to marked cyanosis and tachypnea with retraction are present in newborns
2. ***A loud, single S2 and gallop rhythm are present***.
3. **Heart murmur is usually absent**. If present, however, it is usually a faint ,ejection-type systolic murmur at the upper left sternal border.
4. **Pulmonary crackles and hepatomegaly are usually present**.

ECG

- **WITHOUT OBSTRUCTION**
  - RVH of the so-called <u>volume **overload type** (i.e**., rsR' in V1)**</u> and **o**ccasional RA dilatation are present.
- **WITH OBSTRUCTION**
  - Invariably, **<u>RVH in the form of tall R waves in the right precordial leads is present</u>.** RA dilatation is occasionally present.

Auscultation



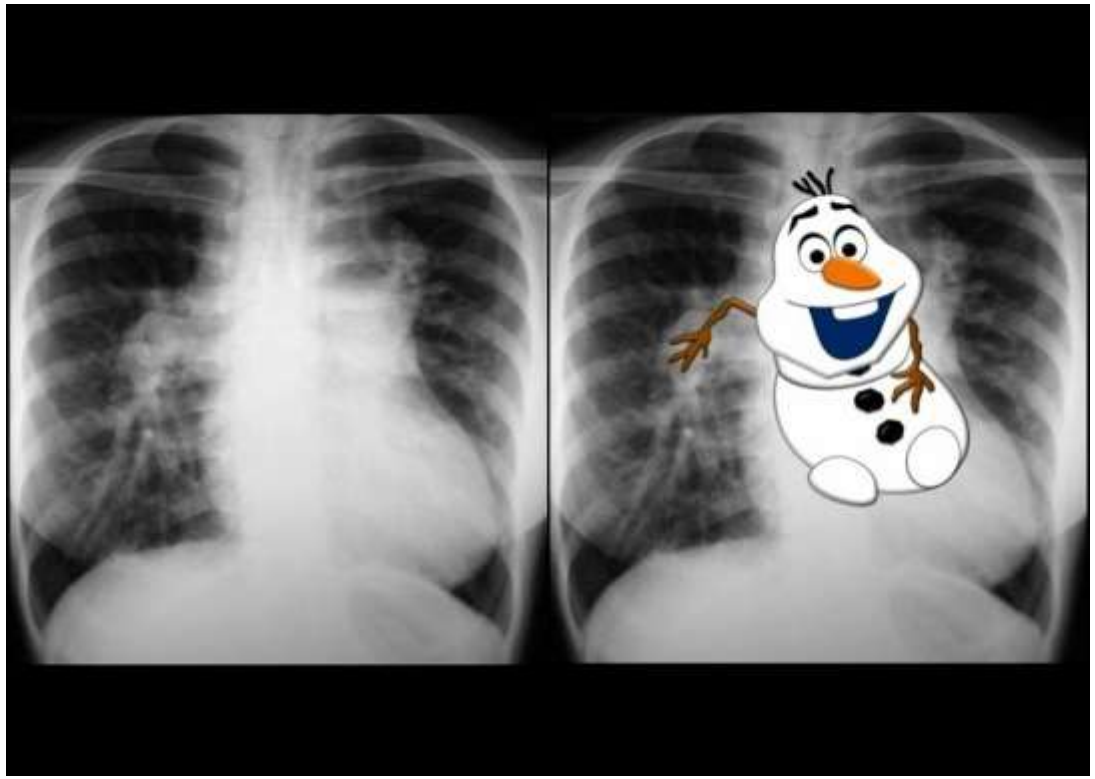
Chest X-ray Studies

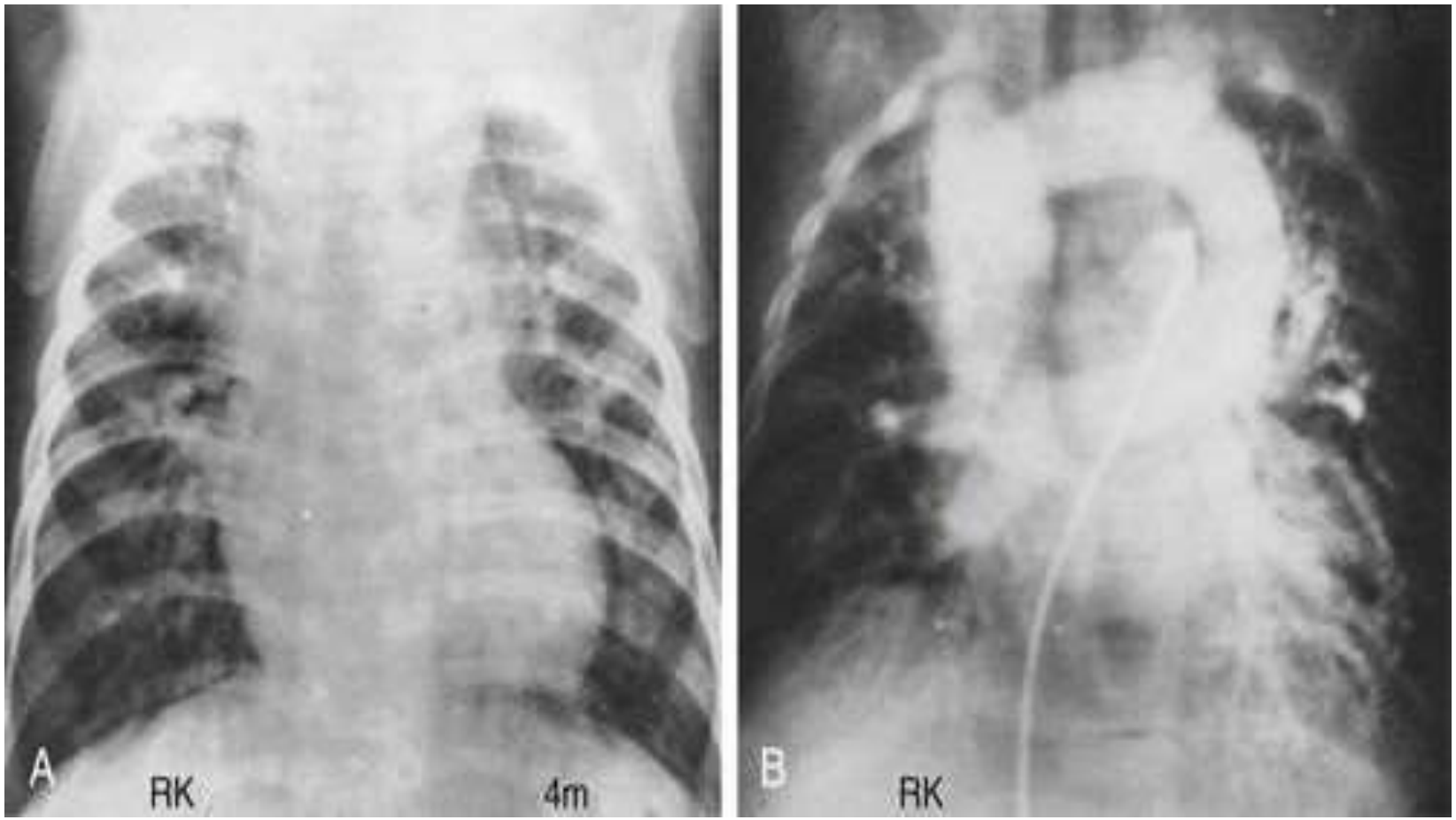
WITHOUT OBSTRUCTION

1. Moderate to marked cardiomegaly involving the RA and RV is present with increased pulmonary vascular markings.
2. “Snowman” sign or figure-of-8 configuration may be seen in the supracardiac type, but **rarely before 4 months of age**

WITH OBSTRUCTION

1. The heart size is normal or slightly enlarged. The lung fields reveal findings of pulmonary edema ,These findings may be confused with pneumonia or hyaline membrane disease.




A
RK
4m
B
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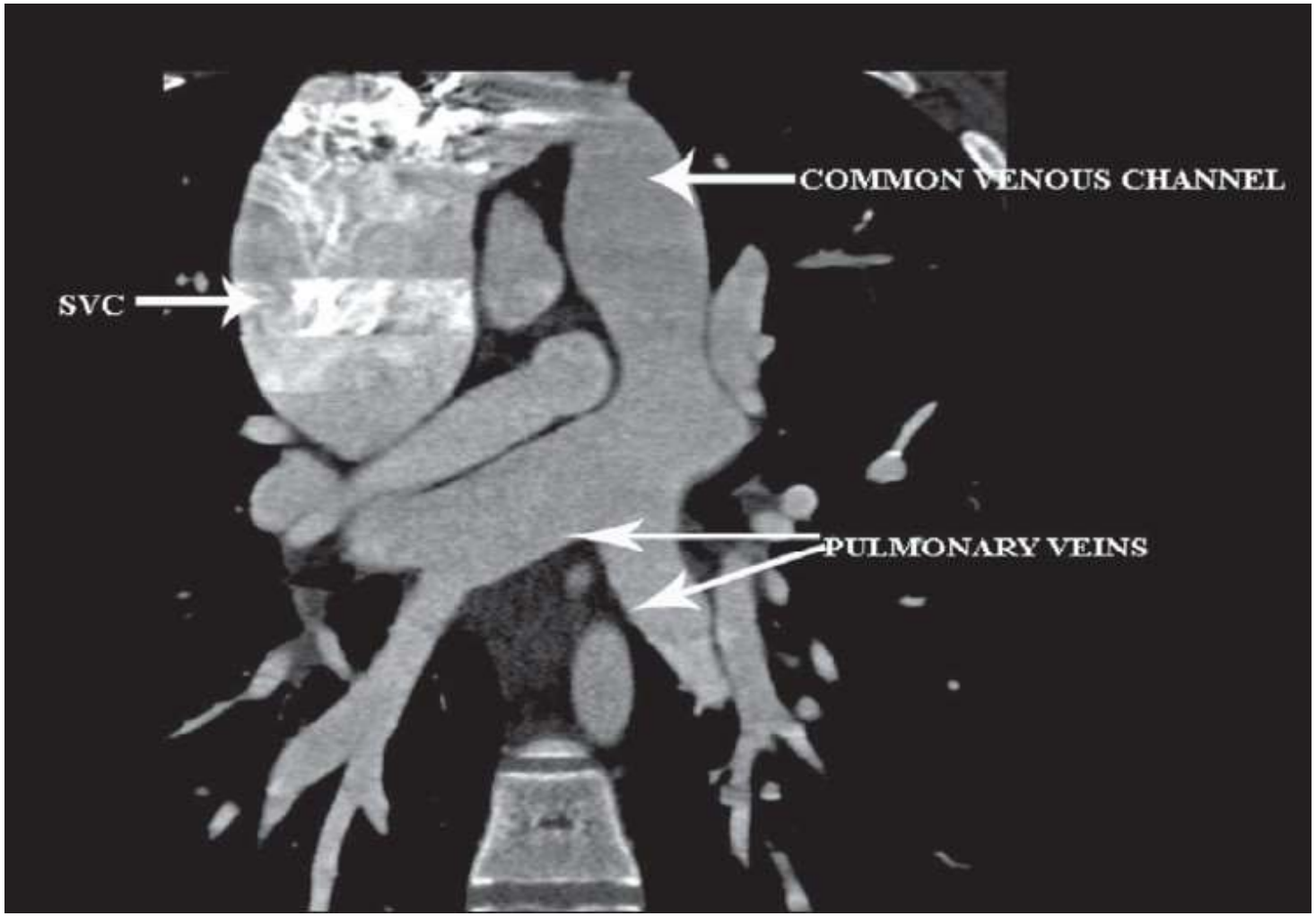
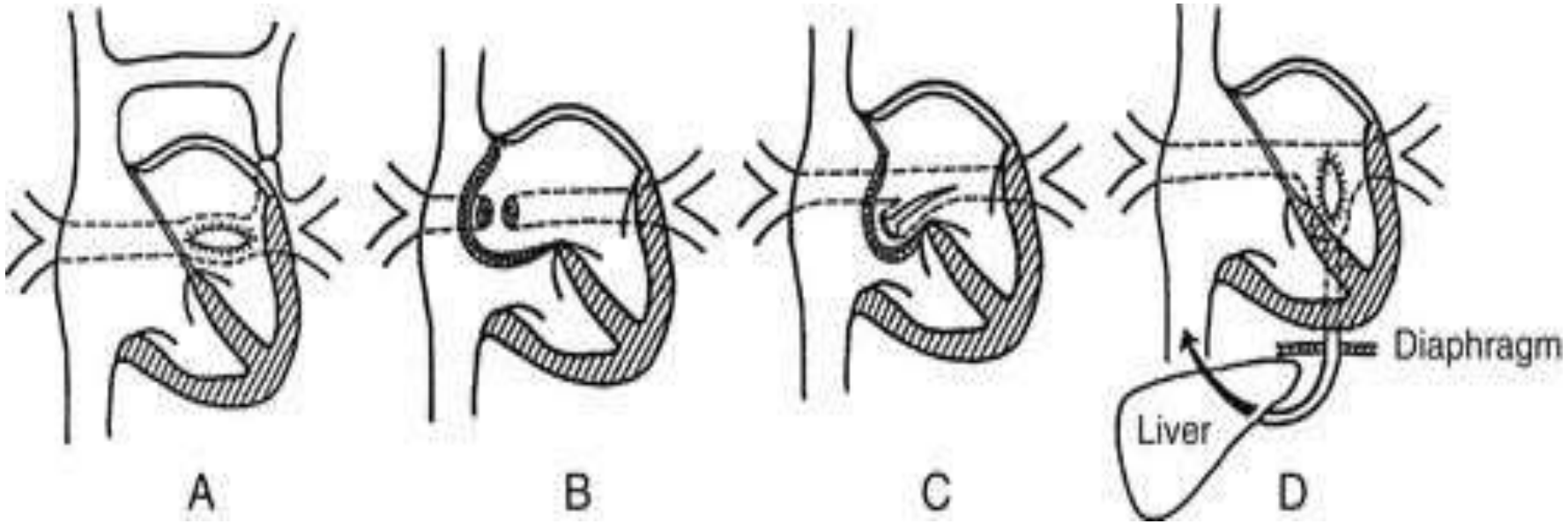

COMMON VENOUS CHANNEL
SVC
PULMONARY VEINS

Management of TAPVR

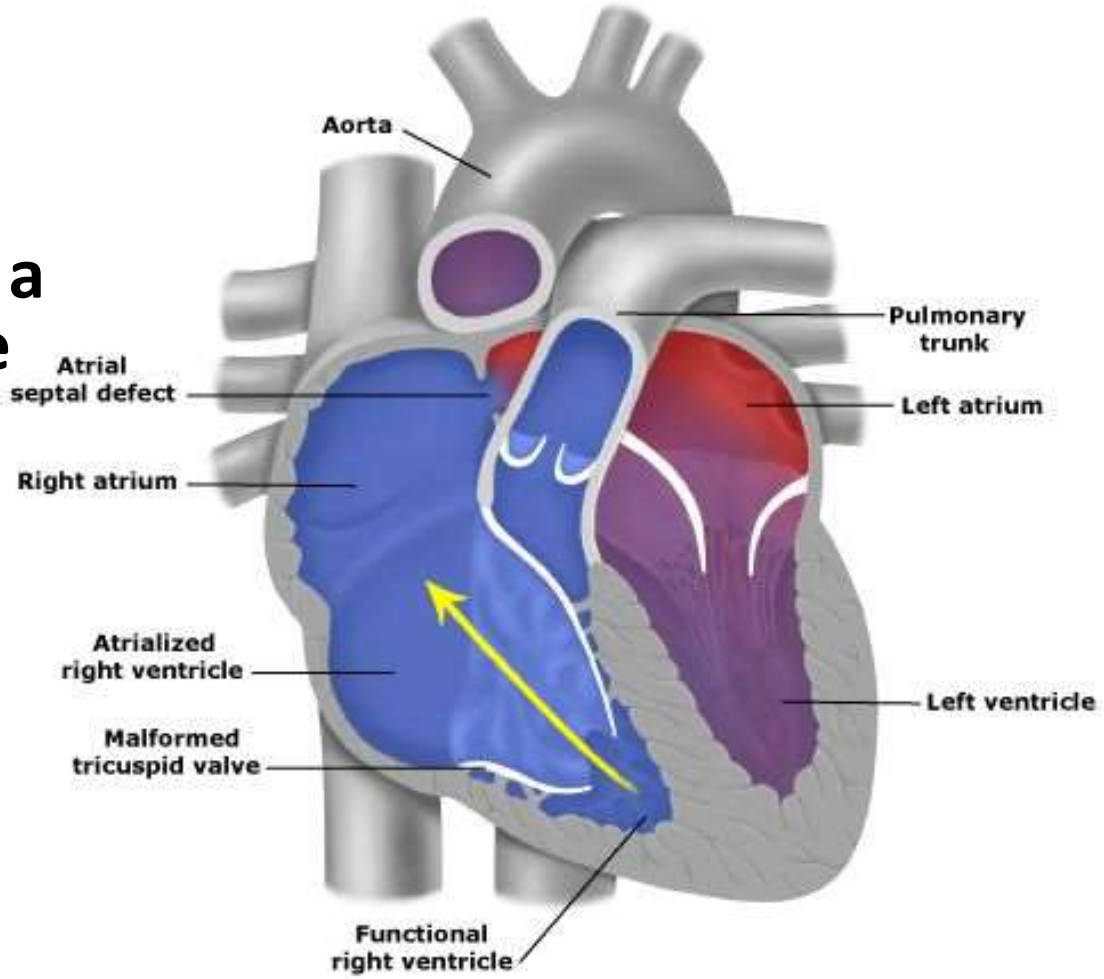
- **Medical and surgical:**
- **O2** supplementation for desaturation
- **Loop Diuretics** for pulmonary edema
- **NAHCO3** for Metabolic acidosis correction.
- Proper anti-failure therapies with **diuretics and/or digoxin** should be given to baby
- Surgical redirection can be done during neonatal period, all of the four pulmonary veins are reconnected to the left atrium, and any other associated heart defects must be closed surgically.
- Infants with severe pulmonary edema resulting from the infracardiac type and from other types with obstruction should be **intubated and receive ventilator**
    - **Therapy with oxygen and positive end-expiratory pressure, if necessary, before cardiac catheterization and surgery.**
    - If the size of the interatrial communication appears to be small and immediate surgery is not indicated, **balloon atrial septostomy or blade atrial septostomy may be performed to enlarge the communication**.

- Surgical Correction of TAPVR:



Ebstein anomaly

- There is downward displacement of the septal and posterior leaflets of the tricuspid valve into the RV cavity, so that a portion of the RV is incorporated into the RA (i.e., atrialized RV) and functional hypoplasia of the RV results.
- TR is usually present.
- RA is dilated and hypertrophied.
- An interatrial communication (e.g., PFO, true ASD) with a right-to-left shunt is present in all patients.



Clinical manifestation

- In **severe cases**, **cyanosis and CHF** develop during **the first few days of life**. Some subsequent improvement coincides with reduction of the pulmonary vascular resistance.
- In **milder cases**, children may complain of dyspnea, fatigue, cyanosis, or palpitation on exertion.

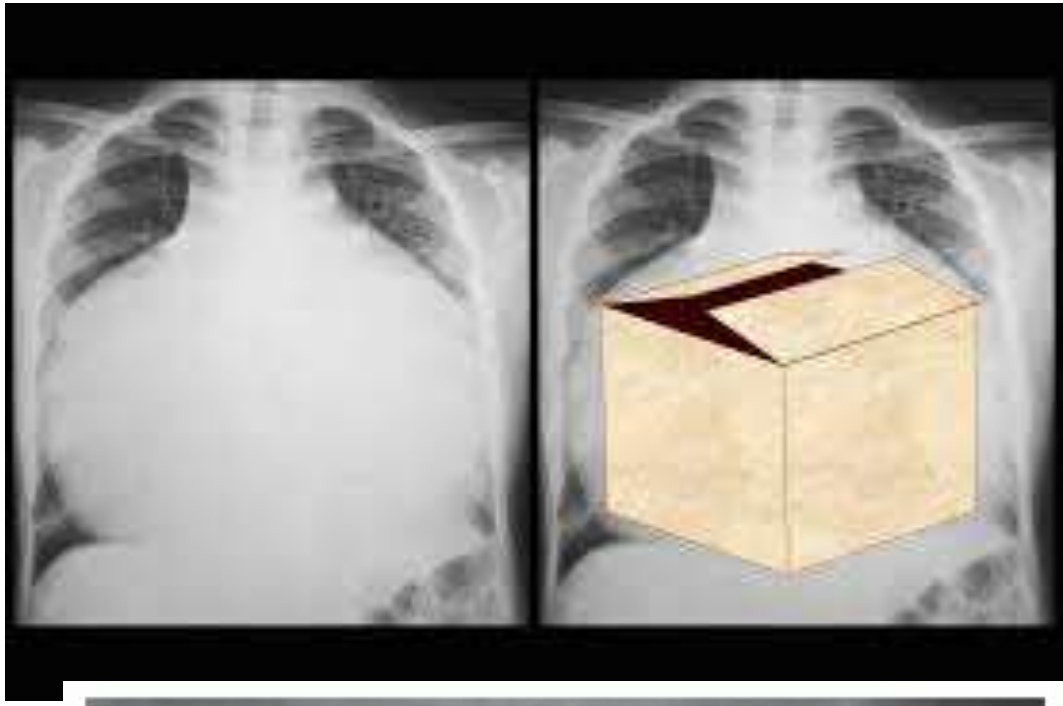
- **PH. EXAM:**
  - **Mild to severe cyanosis is present**
  - **Characteristic triple or quadruple rhythm is audible.** This rhythm has a widely split S2, in addition to split S1, S3, and S4.
  - **A soft, systolic regurgitant murmur of TR is usually audible at the lower left sternal border .**
  - A soft, scratchy, mid-diastolic murmur is present at the same location.
  - **Hepatomegaly is usually present.**

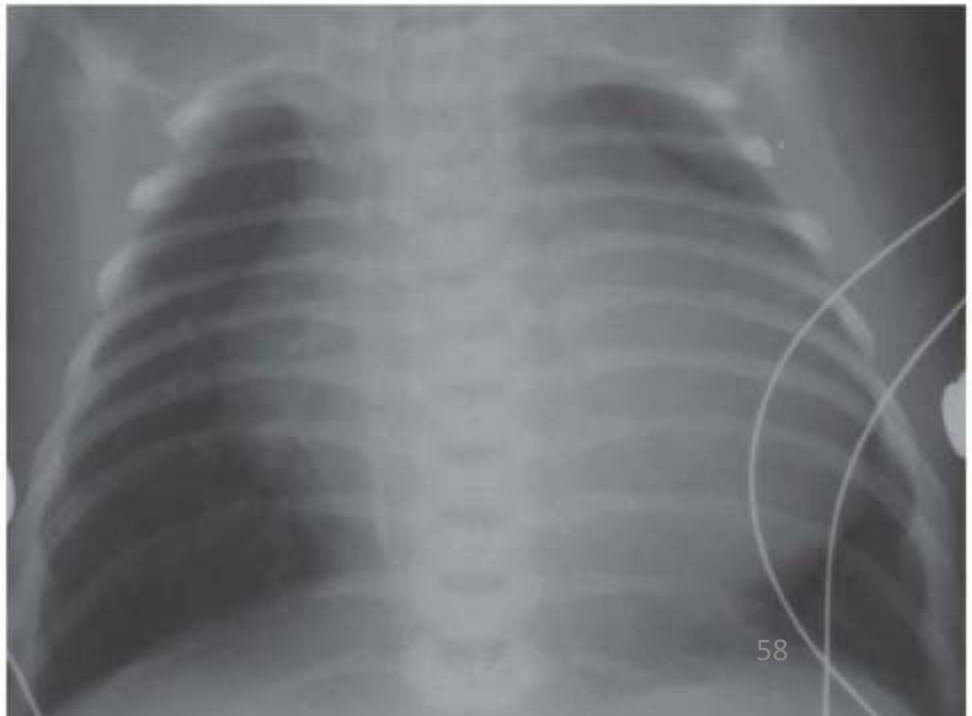
ECG:

- Characteristic ECG findings of **RBBB and RA dilatation** are present in most patients with this condition
- **First-degree AV block is frequent**, occurring in 40% of patients.
- **WPW syndrome is present in 20% of patients with occasional episodes of SVT**

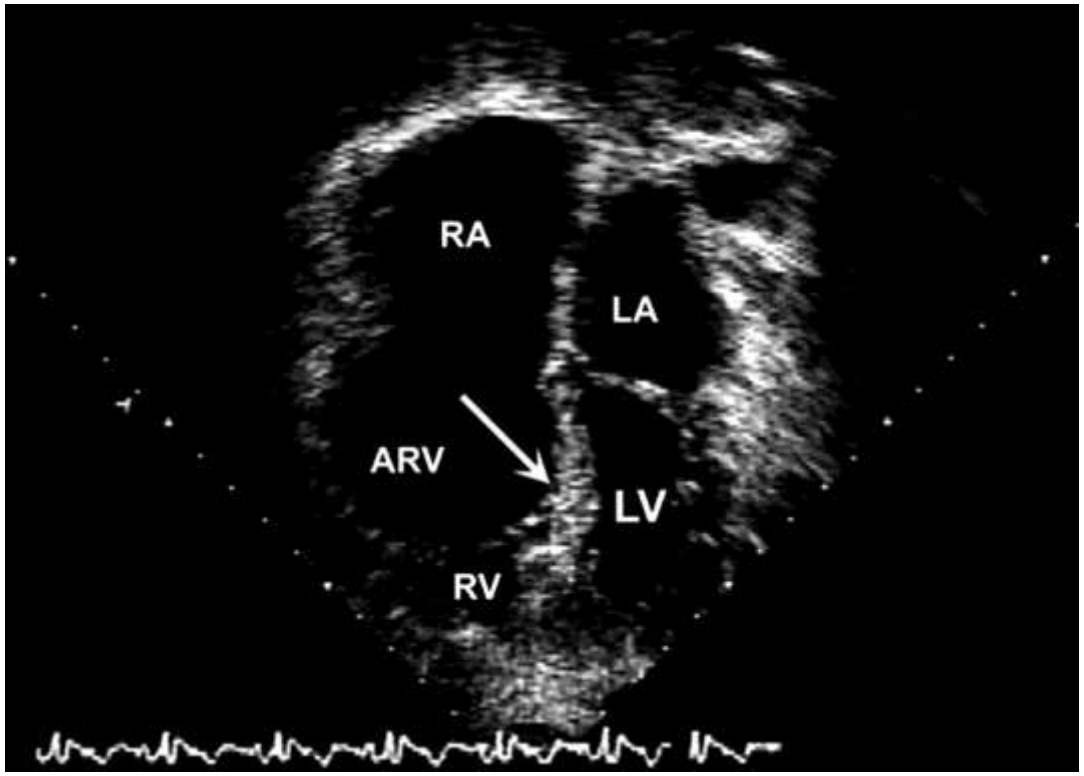
Chest x ray findings

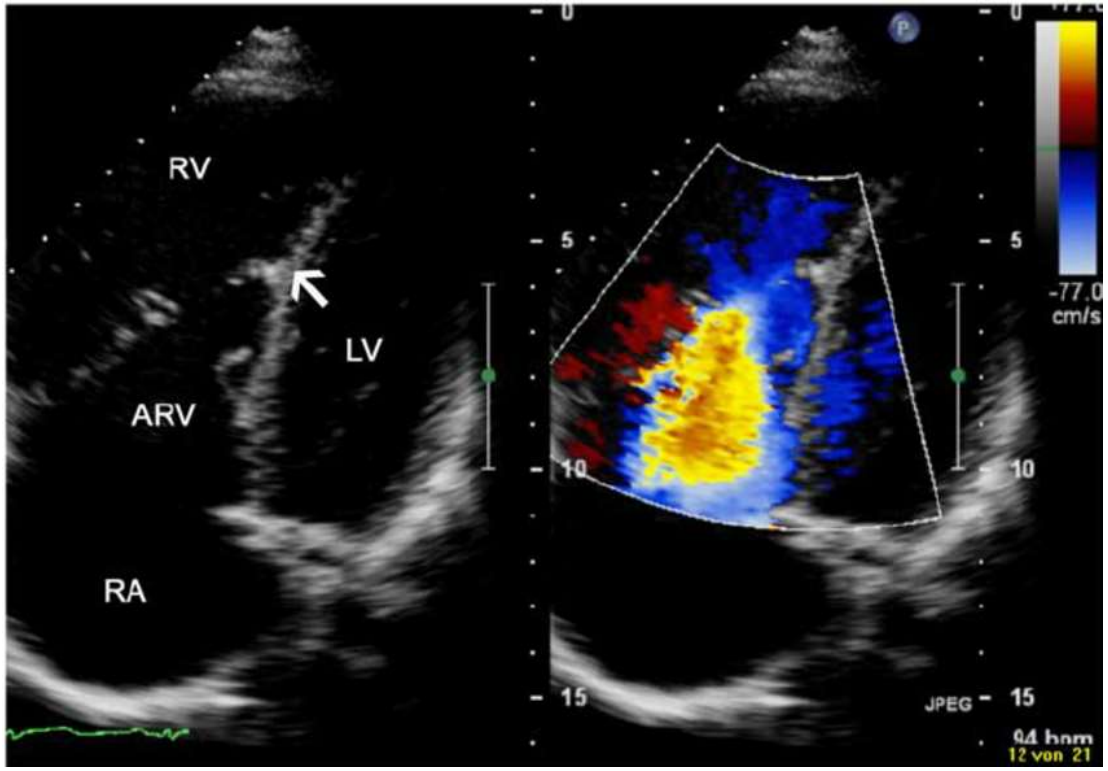
- In **mild cases**, the heart is **almost normal in size and has normal pulmonary vascular markings.**
- In **severe cases**, an extreme cardiomegaly (principally involving the RA) with **a balloon-shaped heart** and **decreased pulmonary vascular markings are present**. Some of the largest heart sizes are found in newborns with this condition.





2D-Echocardiogram





Management:

- **Medical management in a newborn with cyanosis:**
  - **In case of extreme cyanosis, prostaglandin E1 infusion is indicated**
  - **In case of Critically ill newborns may require inotropic support (Milrinone infusion) until surgical intervention is done or the clinical situation is improved**
  - **IN severe cases with small RV: PGE1, systemic to pulmonary shunt, then staged surgery as Glenn procedure and fontan operation (it is preformed to reduce the blood volume on the right ventricle)**

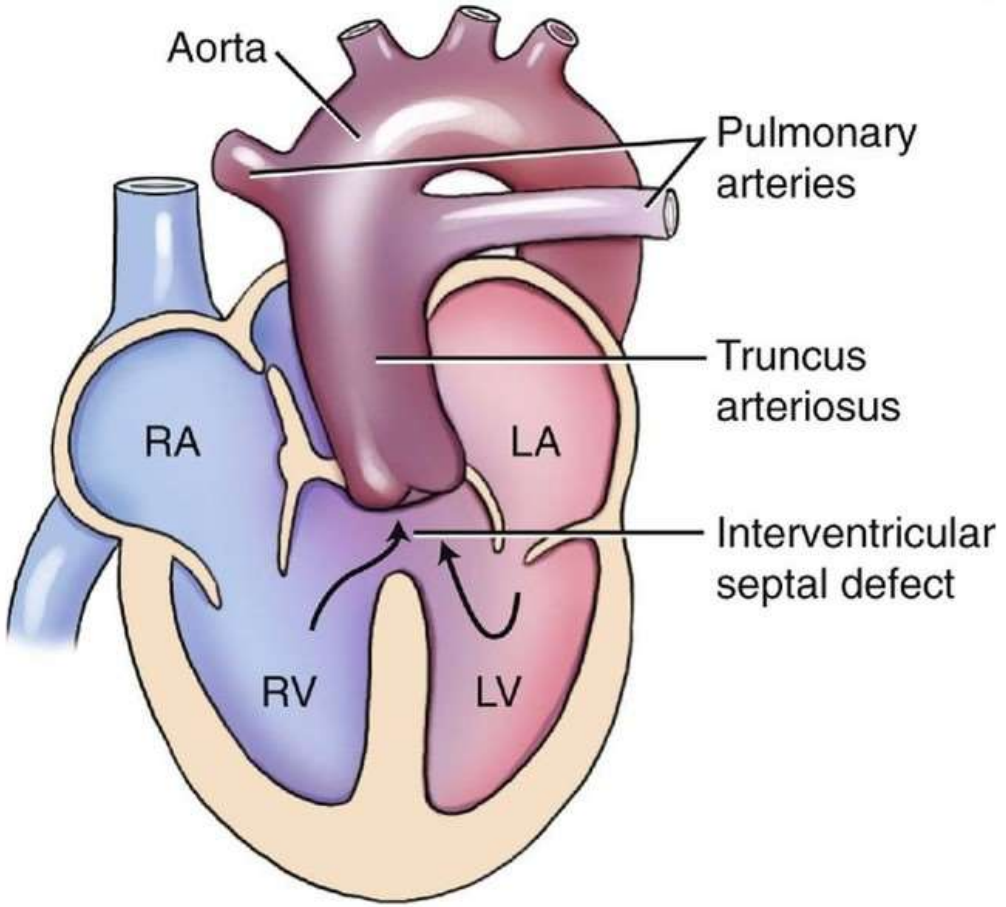
Management:

- **Medical management of symptomatic Infant and children Ebstein patients includes:**
    - **Heart failure management to get the best clinical situation with inotropic agent (Milrinone, digoxin), and the loop diuretic before evaluation for early planned surgical correction**
- **Medical management of symptomatic adult Ebstein patients includes:**
    - **Diuretic therapy for right side heart failure**
    - **Antiarrhythmic drug if there is any abnormalities**
    - **Anti-failure medication for left side systolic dysfunction**
    - **Surgical intervention**

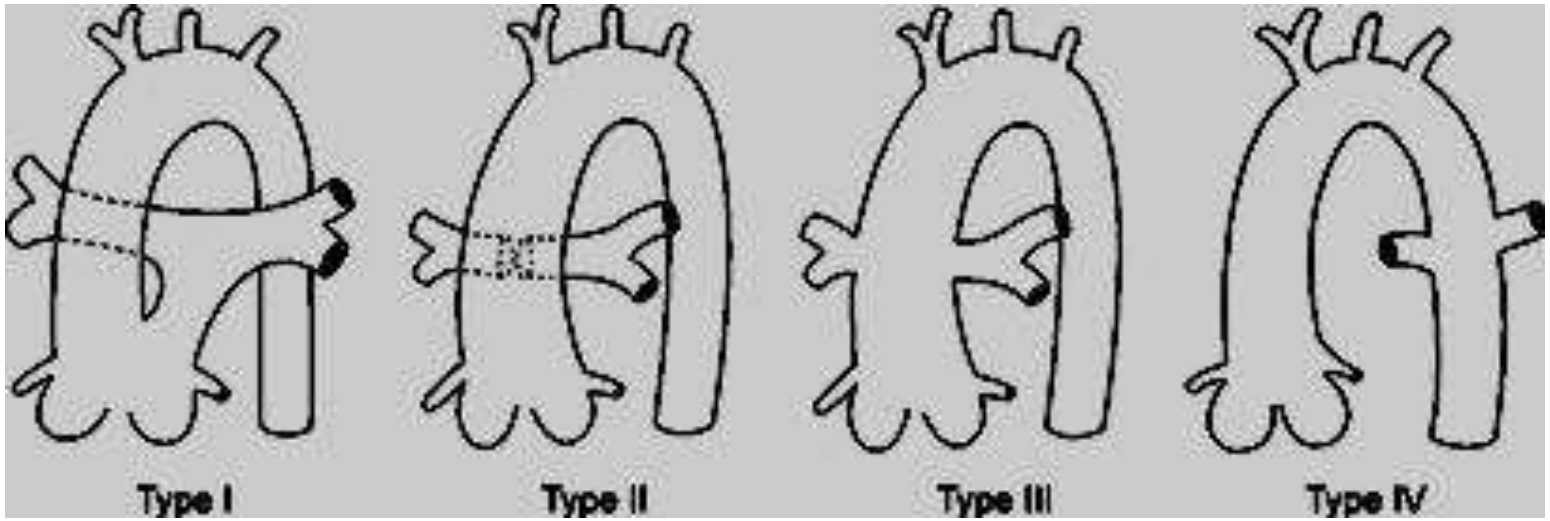
Truncus Arteriosus

- **Persistent truncus arteriosus** occurs in <1% of all congenital heart defects
  - **<u>Only a single arterial trunk with a truncal valve leaves the heart and gives rise to the pulmonary, systemic, and coronary circulations.</u>**
  - **A large perimembranous,infundibular VSD is present directly below the truncus**

- Evidence of **DiGeorge syndrome with hypocalcemia is present in 33% of patients.**



- This anomaly is divided into four types according to Collett and Edwards' classification .



Clinical manifestation

- Cyanosis may be **seen immediately after birth.**
- Signs of **CHF develop within several days or weeks after birth**
- History of dyspnea **with feeding, failure to thrive, and frequent respiratory infections** is usually present in infants

Physical Examination

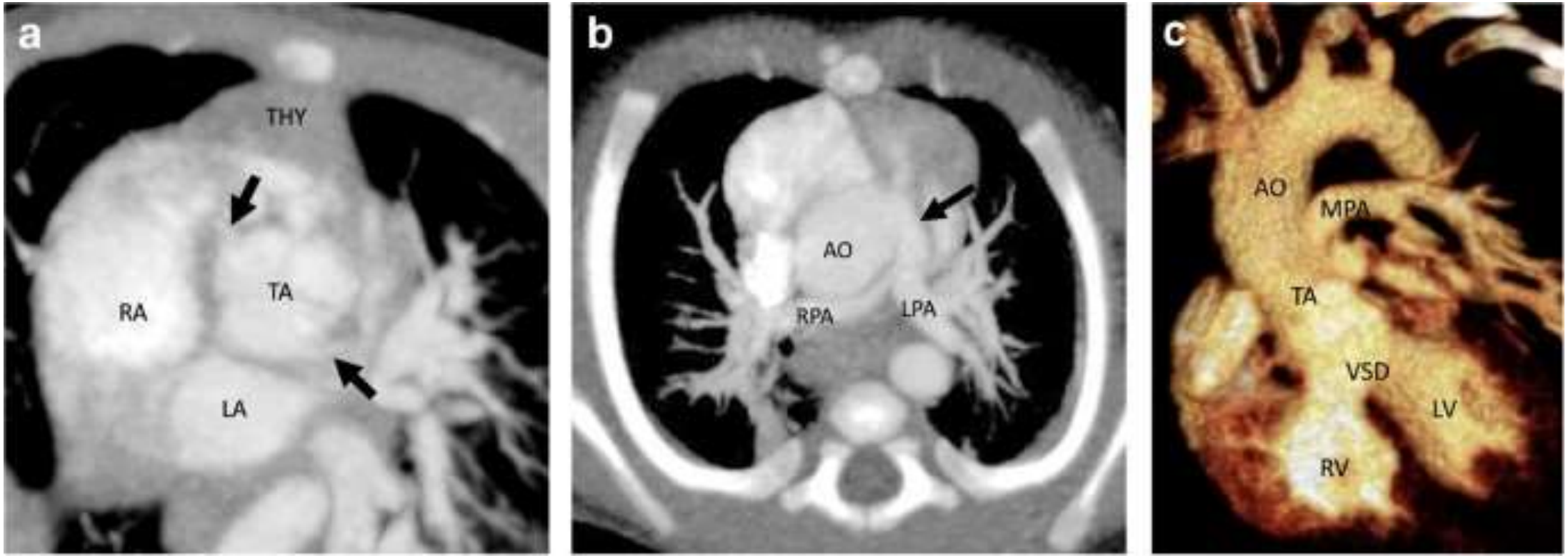
- **Varying degrees of cyanosis and signs of CHF with tachypnea and dyspnea are usually present.**
  - **The peripheral pulses are bounding, with a wide pulse pressure.**
  - **The S2 is single.**
  - **An apical rumble with or without gallop rhythm** may be present **when the PBF is large.**
  - **Truncal valve regurgitation(if present)** A high-pitched, early diastolic, decrescendo murmur **may be audible.**
  - Rarely is a continuous murmur heard over either side of the chest

Chest x ray findings

- Depend on TA type
- Cardiomegaly is usually present, with increased pulmonary vascularity, specially in type one

ECG

The QRS axis is normal (+50 to +120 degrees). BVH is present in 70% of cases.


a
THY
TA
RA
LA
b
AO
RPA
LPA
c
AO
MPA
TA
VSD
LV
RV

Management:

- In case of normal pulmonary arteries with increased pulmonary blood flow (In the 1st few weeks of life):
    - **Anti congestive medications**
- In case of small hypoplastic pulmonary arteries with decreased pulmonary blood flow and severe cyanosis :
    - **PGE1 is started and shunt is done to increased PBF and augment growth of pulmonary arteries after proper imaging for evaluation of pulmonary arteries with catheterization or CT angiogram**

THANK YOU FOR YOUR ATTENTION