

Pediatrics cardio-vascular disorders

Congenital Heart Disease (CHD)

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Congenital Heart Disease (CHD)

Epidemiology of CHD

Incidence - 8/1000 live births

- 3-4/100 still born

- 2/100 premature infants excluding PDA

- 10-25/100 abortuses

- Most congenital defects are well tolerated during fetal life.

Etiology - Unknown in most cases

- Genetic factors - single gene defect

- Chromosomal abnormality.

- Environmental factors

CHD ...

Clues for Evaluation of an Infant with suspected CHD

1. On History and Physical Examination color)

- Acyanotic
- Cyanotic

2. Chest roentgenogram

- Normal
- Increased/Plethora
- Decreased/Oligemia

pulmonary blood flow

CHD ...

3. **Electrocardiogram**

- Right
 - Left
 - Biventricular
- } hypertrophy

Final diagnosis - Precordial
examination

- Echocardiography

CHD...

Relative Frequency of Congenital Heart Lesions

Lesions	% of all Lesions
- Ventricular septal defect	25-30
- Atrial septal defect (Secundum)	6-8
- Patent ductus arteriosus	6-8
- Coarctation of aorta	5-7
- Tetralogy of Fallot	5-7
- Pulmonary Valve Stenosis	5-7
- Aortic Valve Stenosis	4-7
- d-Transposition of great arteries	3-5
- Hypoplastic left ventricle	1-3

CHD...

Relative Frequency ...

Lesions	% of all Lesions
- Hypoplastic right ventricle	1-3
- Truncus arteriosus	1-2
- Total anomalous PVR	1-2
- Tricuspid atresia	1-2
- Single ventricle	1-2
- Double-outlet right ventricle	1-2
- Others	5-10

I. Acyanotic Congenital Heart Diseases

1. Left to Right Shunt Lesions

1.1 Atrial Septal Defect

- ☛ Defect occur in any portion of the atrium
 - Ostium secundum
 - Ostium primum (ECD)
 - Sinus venosus

Pathophysiology

- ☛ Left to right shunt
 - Transatrial in OS & SV
 - Transatrial & transventricular in OP

Acyanotic CHD...

Clinical Manifestations

- ☛ Most are asymptomatic
- ☛ Right ventricular lift
- ☛ Wide & fixed split of 2nd heart sound
- ☛ Systolic ejection murmur
- ☛ Mid-diastolic murmur at tricuspid area
- ☛ Holosystolic murmur at mitral area in OP (ECD)

Acyanotic CHD...

Diagnosis

- ☛ Clinical
- ☛ CXR
 - Right. V & A enlargement
 - Large pulm. artery
 - ↑ed pulm. vascularity
- ☛ ECG - volume overload, Rt ventricle & atrium
- ☛ Echocardiography
- ☛ Catheterization

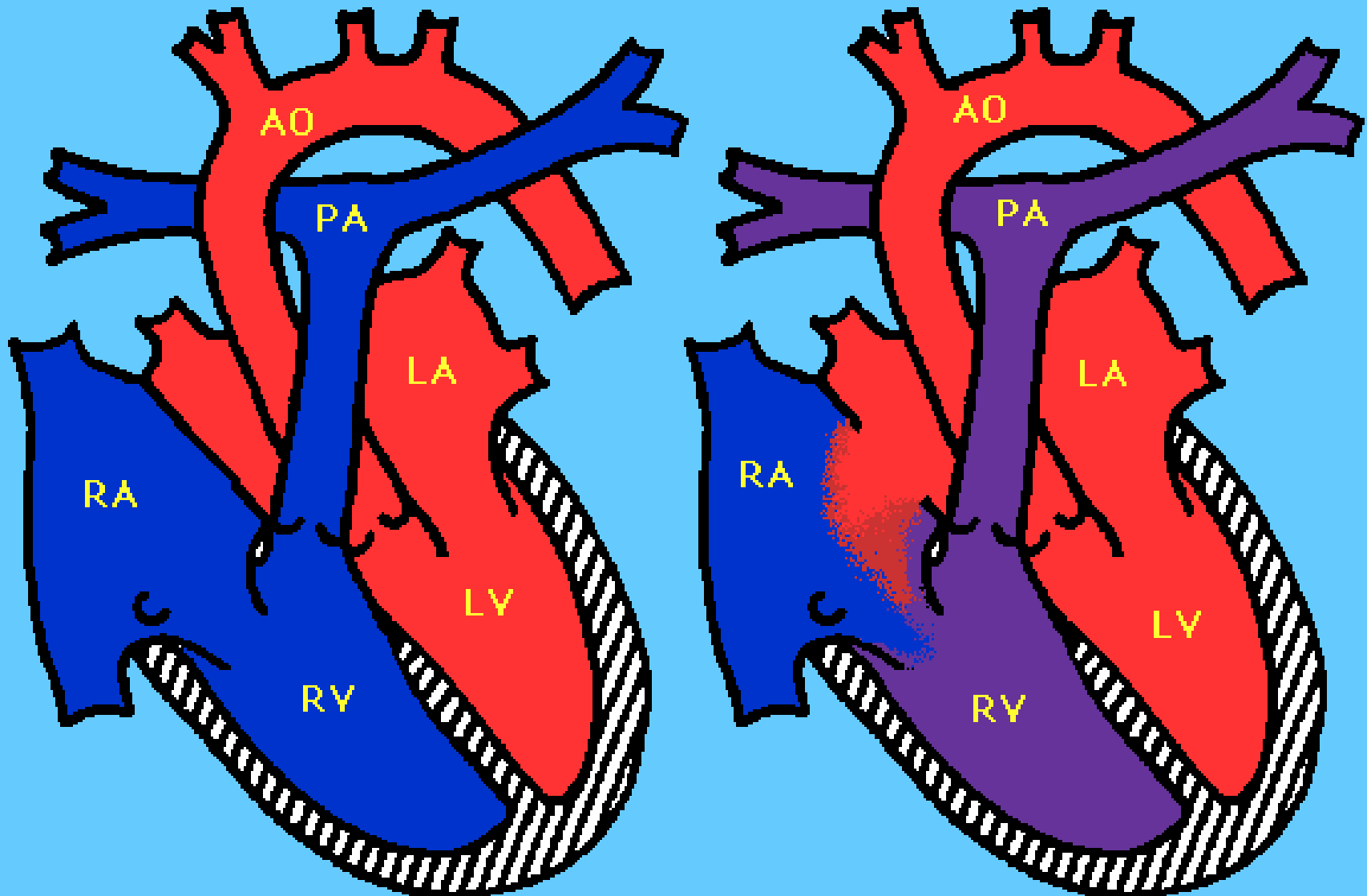
Prognosis - Well tolerated

Complications - pulm. Hypertension Eismenger syndrome

Treatment

- ☛ Surgery-for all symptomatic

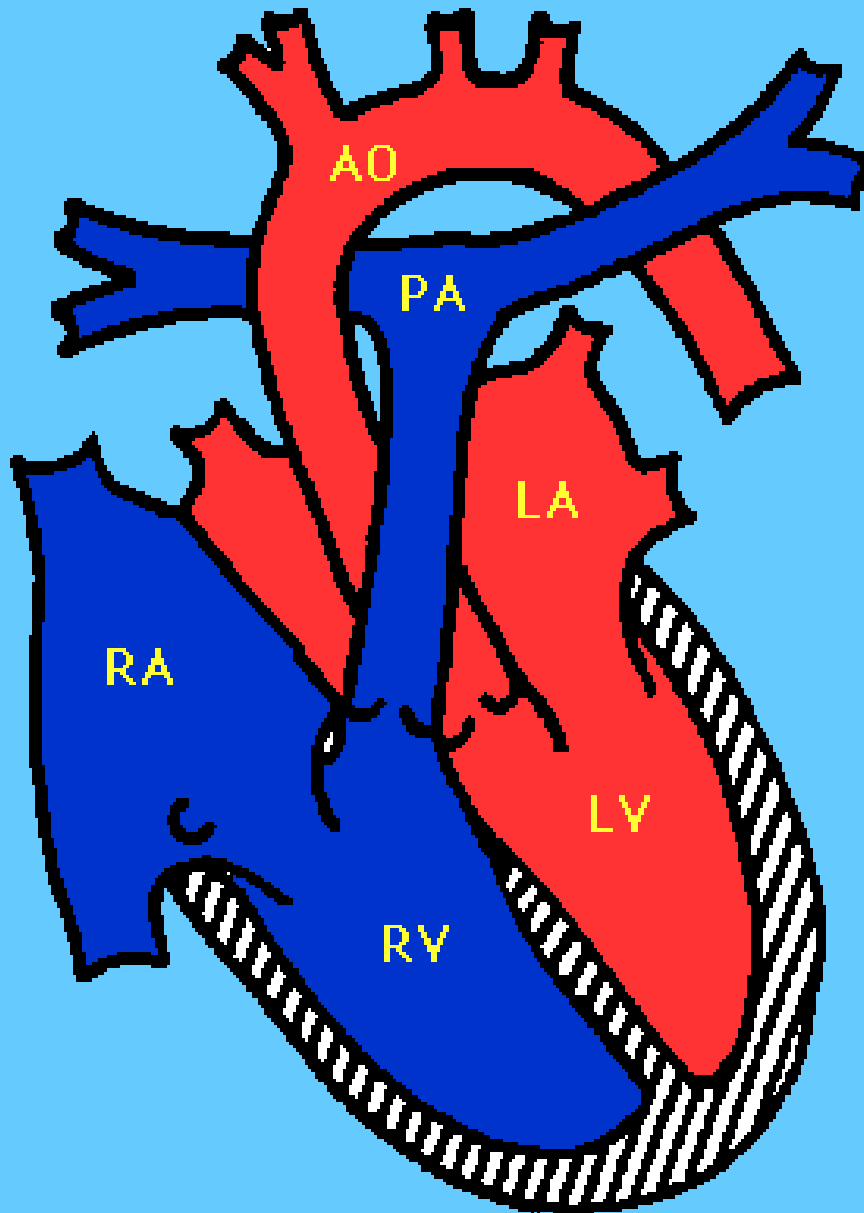
Atrial Septal Defect



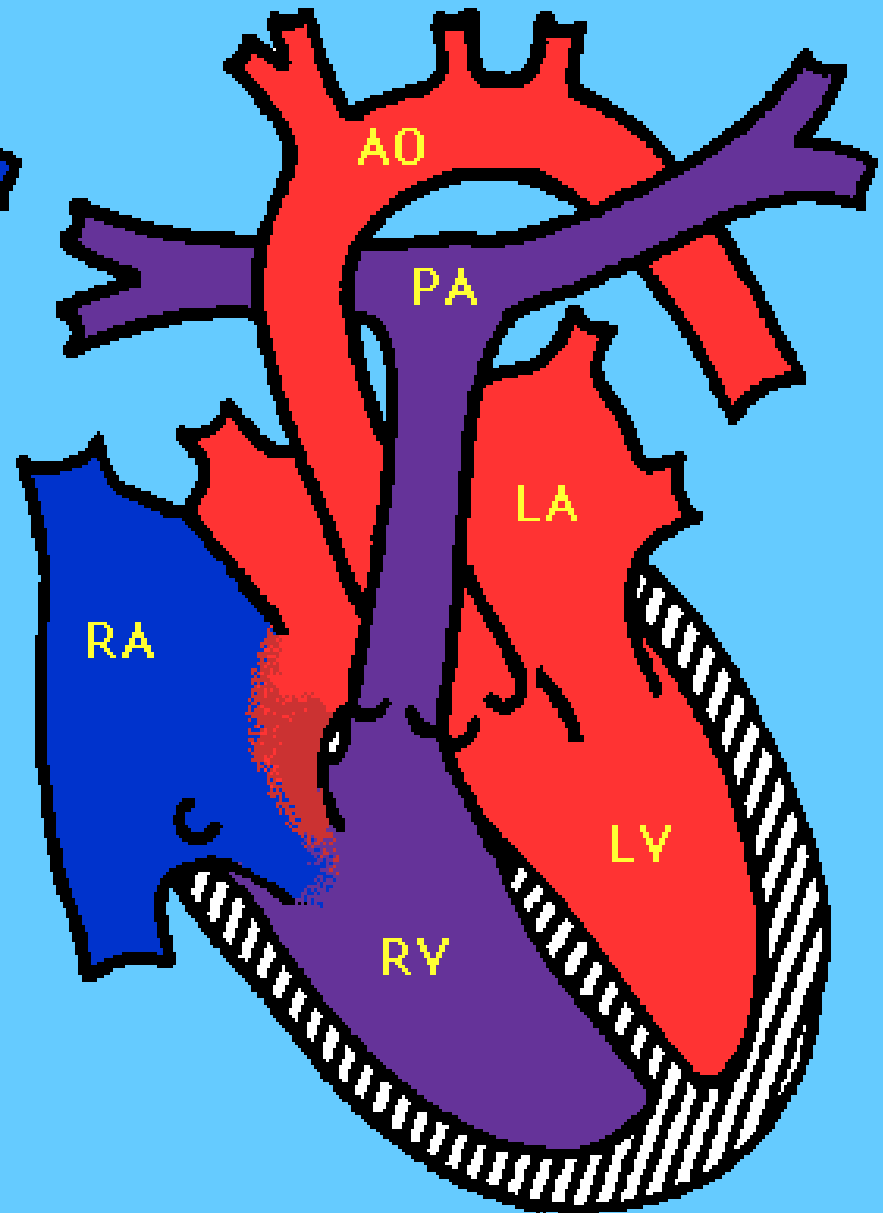
Normal

Atrial Septal Defect

Primum Atrial Septal Defect

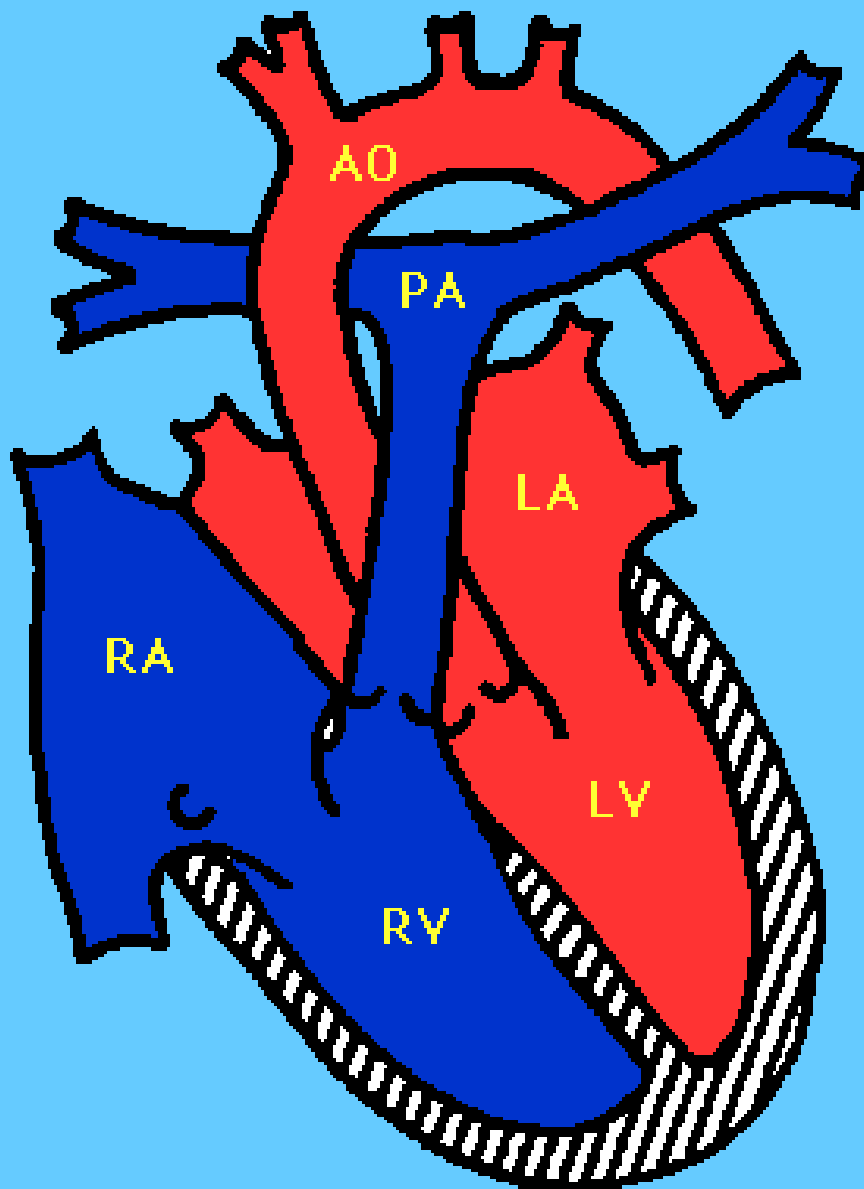


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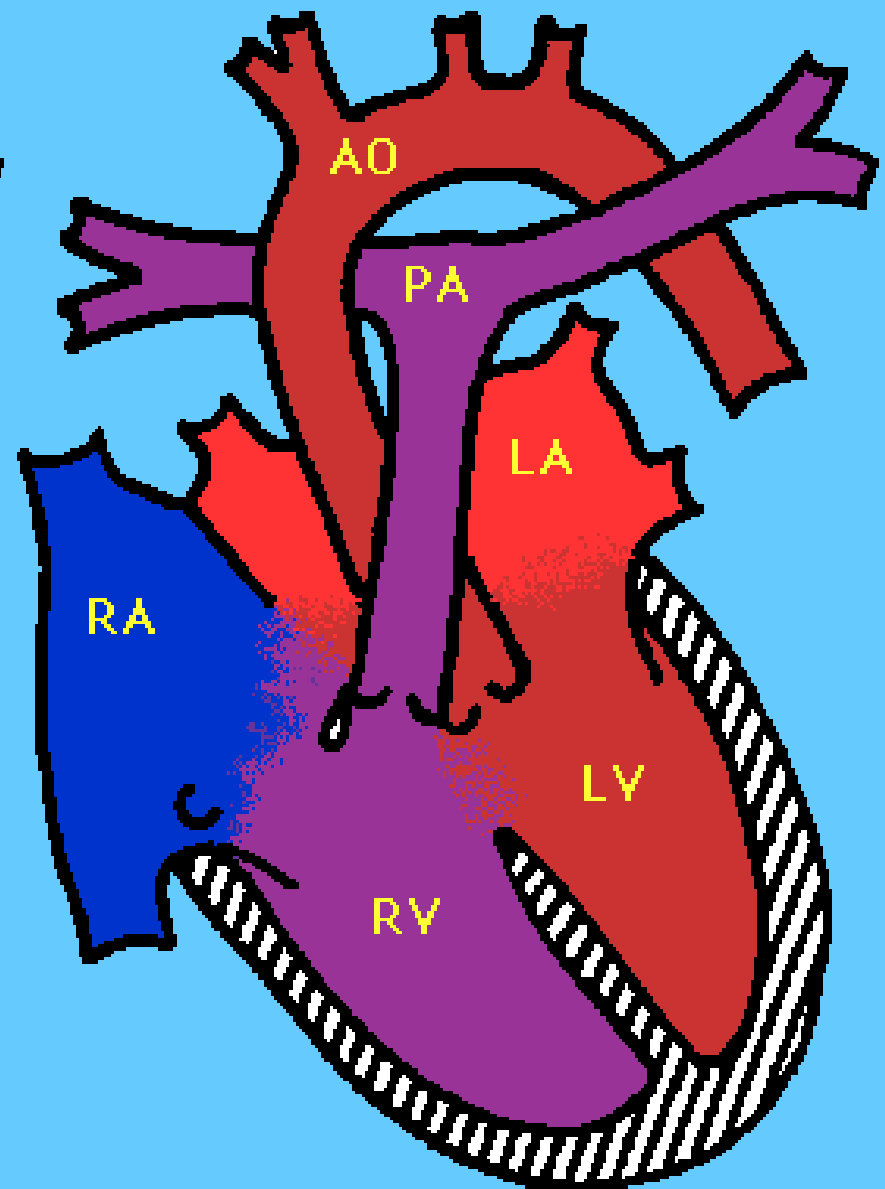


Primum Atrial Septal Defect

Atrioventricular Canal Defect



Normal



Atrioventricular Canal Defect

Acyanotic CHD...

1.2 Ventricular Septal Defect

- ☞ The most common cardiac malformation
- ☞ Defect occur in any portion of the septum
 - Majority -membranous
 - Swiss-cheese defect

Acyanotic CHD...

Clinical Manifestation

☞ Large defects

- Excessive pulmonary blood flow
- Pulmonary hypertension
- Dyspnea, feeding difficulties, poor growth, perspiration, recurrent plum. infection, heart failure
- Less harsh but more blowing holosystolic murmur
- Accentuated 2nd heart sound
- Mid-diastolic apical murmur

Acyanotic CHD...

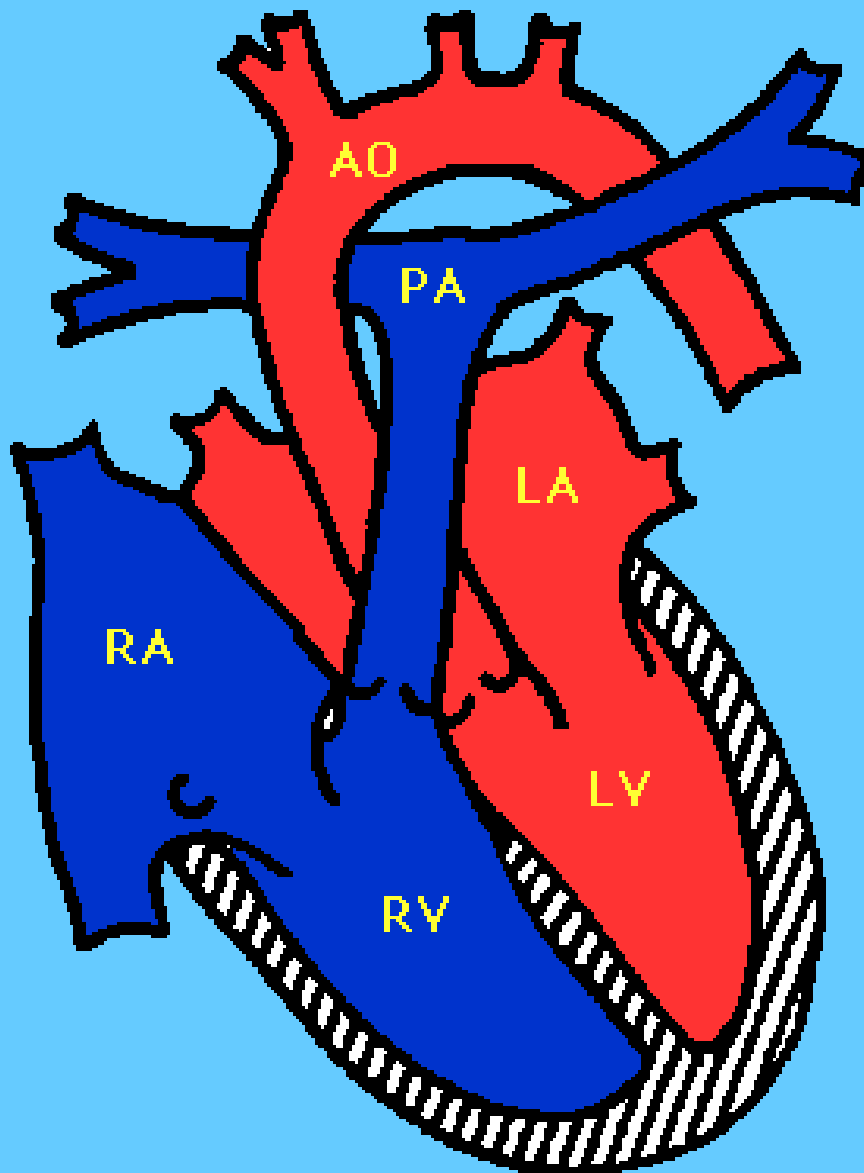
Diagnosis

- **Clinical**
- **CXR** - **Cardiomegaly**
 - **Plethoric lung**
- **ECG**
- **Echocardiography**

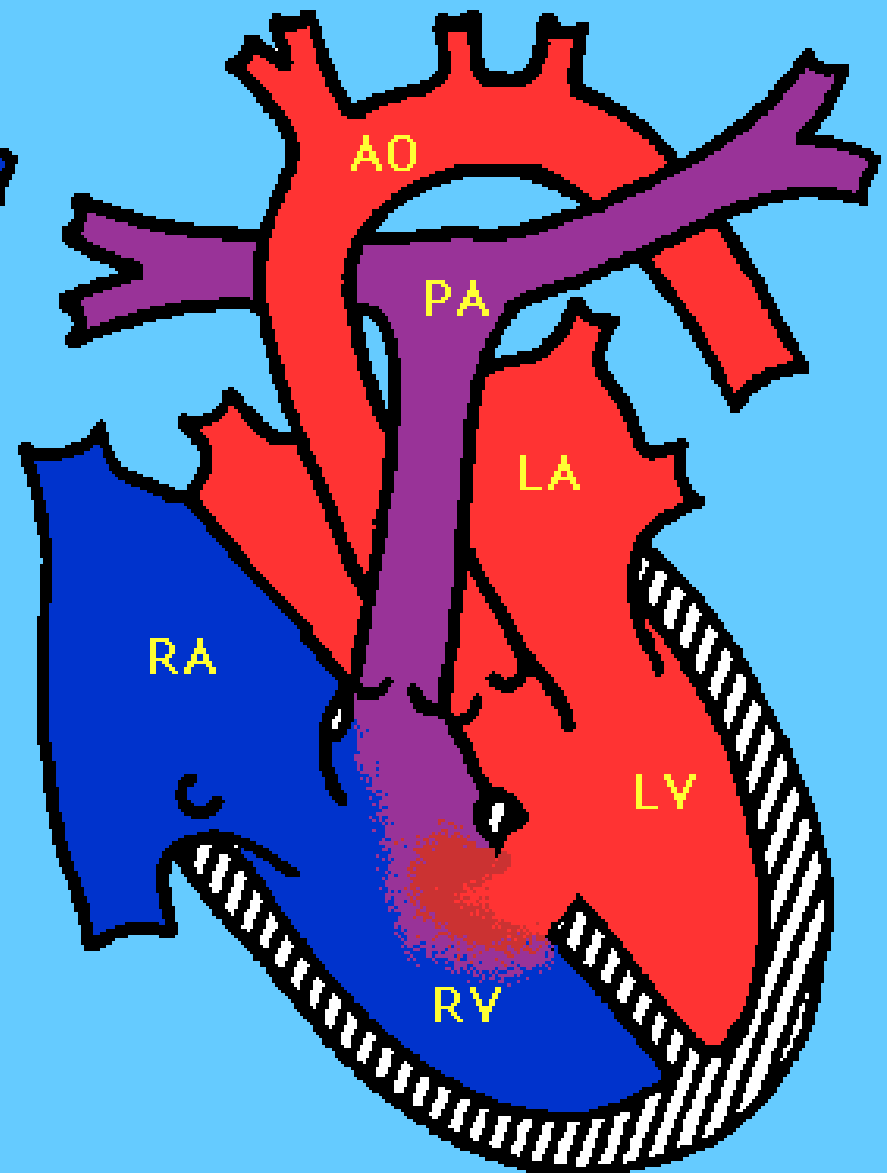
Prognosis

- **30-50% small defects close**
- **Rarely moderate to large defects close**

Ventricular Septal Defect



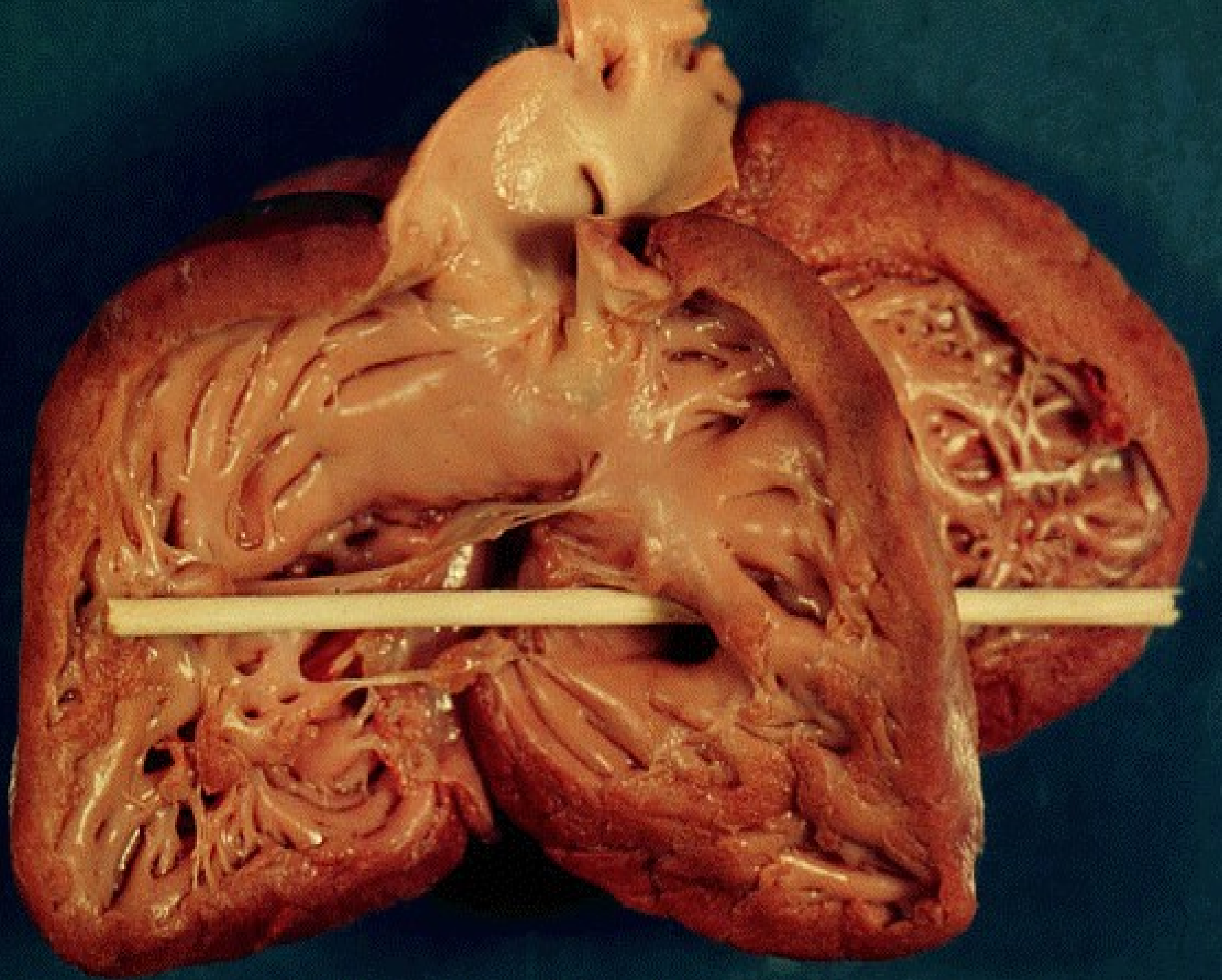
Normal



Ventricular Septal Defect



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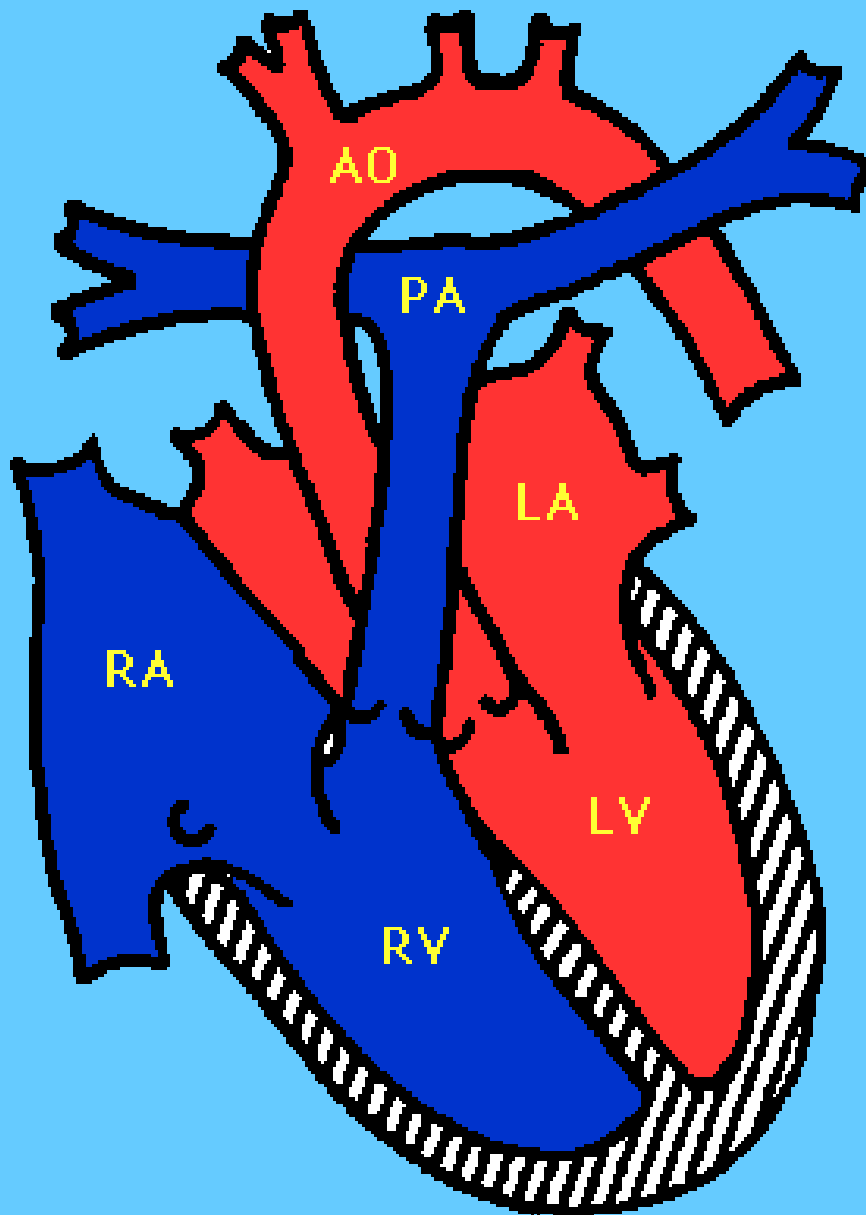


Acyanotic CHD...

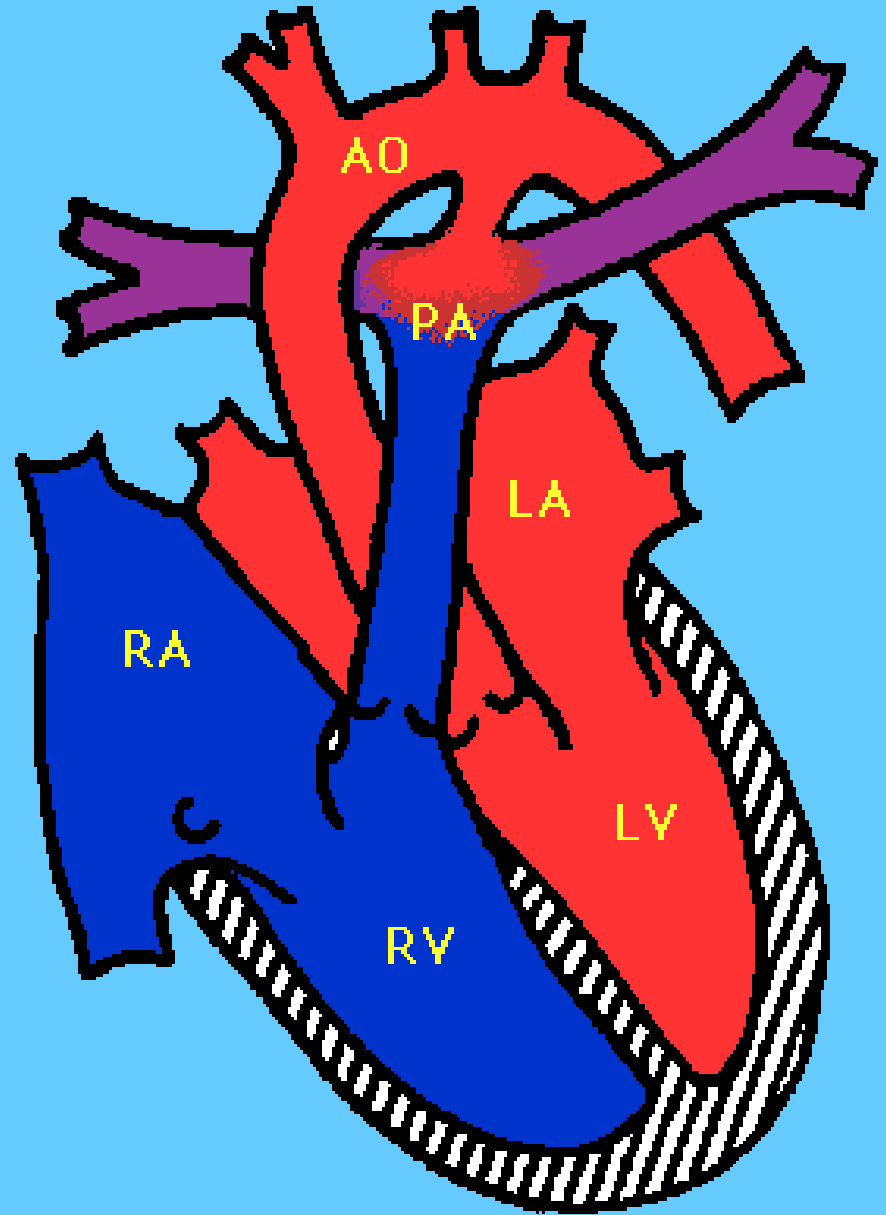
1.3 **Patent Ductus Arteriosus**

- ☞ Functional closure soon after birth
- ☞ Aortic end of the ductus distal to the origin of subclavian artery and the other end of bifurcation of pulmonary artery.
- ☞ Male to female ratio 1:2

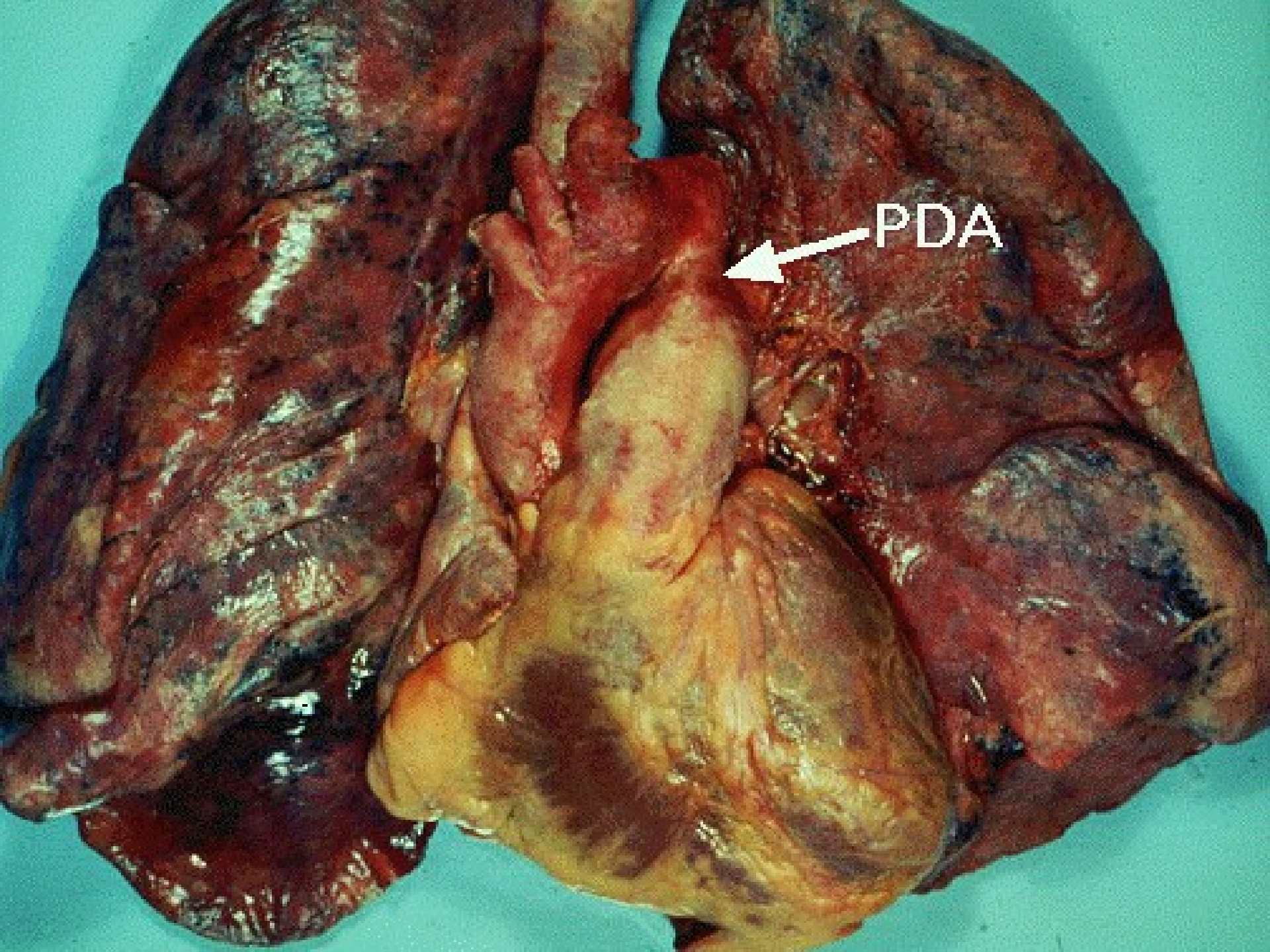
Patent Ductus Arteriosus



Normal



Patent Ductus Arteriosus



PDA

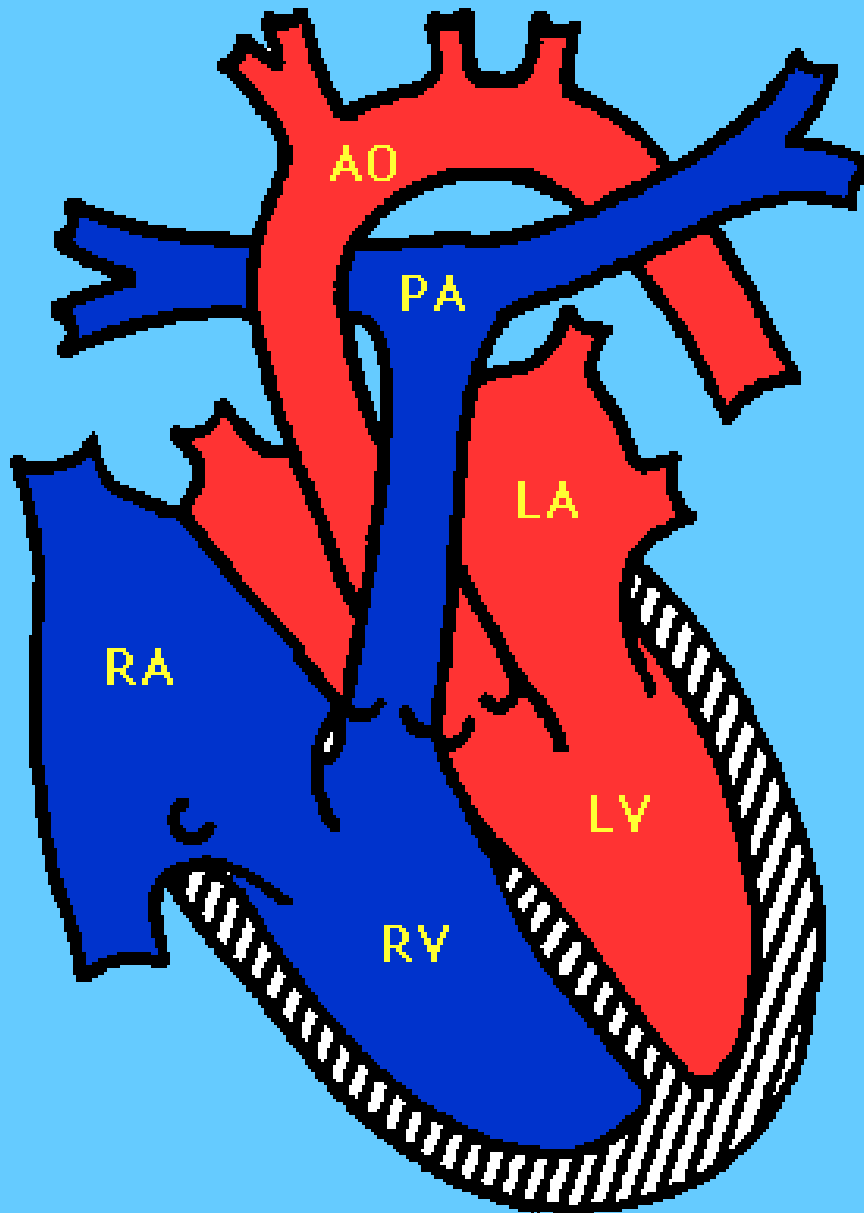
Acyanotic CHD...

2. Obstructive Lesions

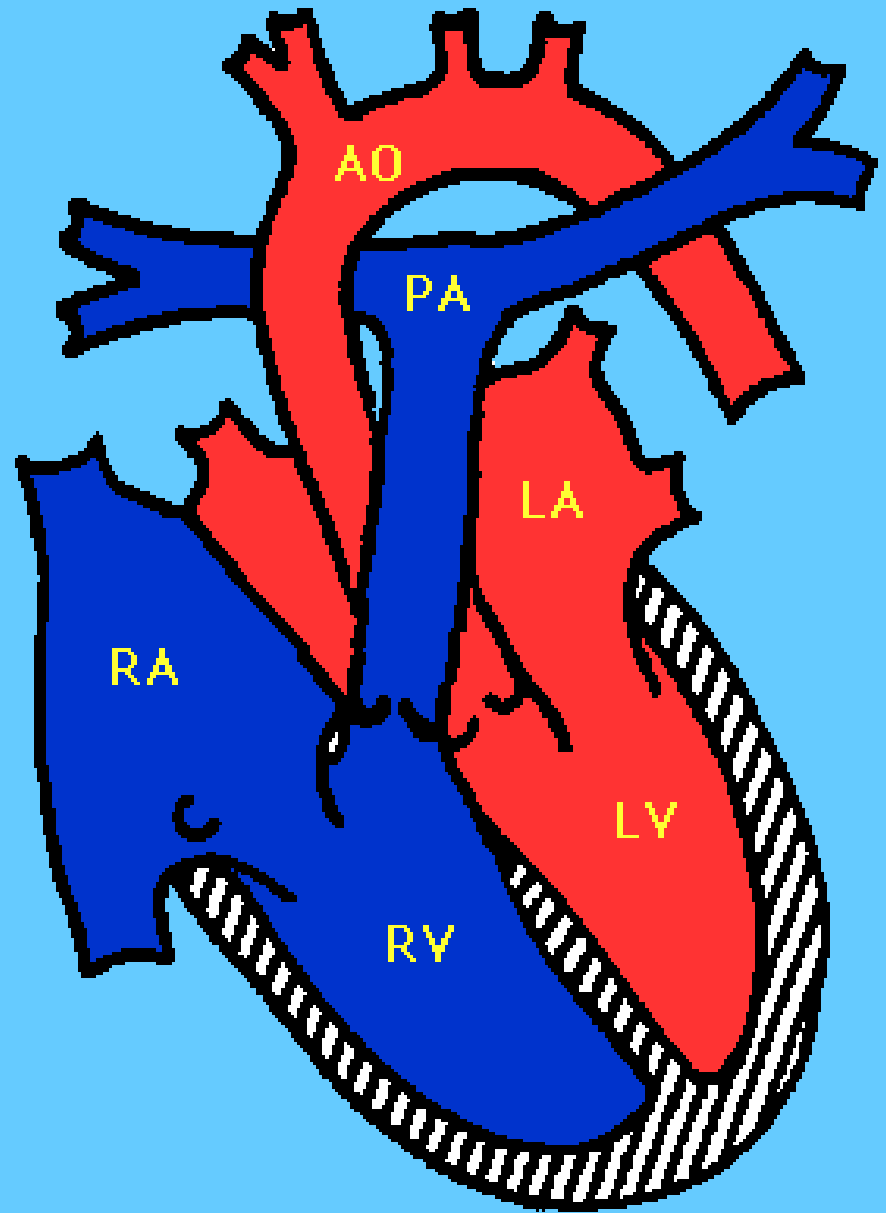
2.1 Pulmonic Stenosis - 4 types

- Valvular
- Infundibular
 - Supra valvular
- Peripheral

Pulmonary Valve Stenosis



Normal



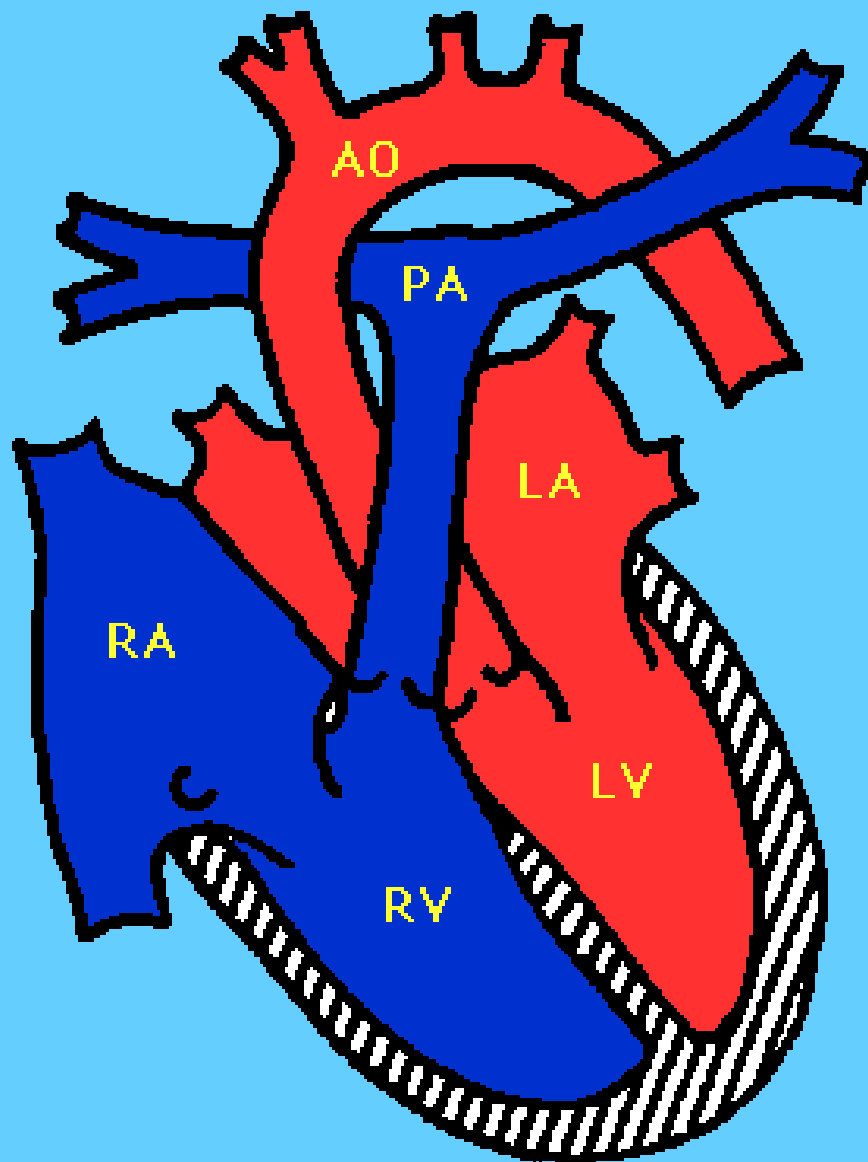
Pulmonary Valve Stenosis

Acyanotic CHD...

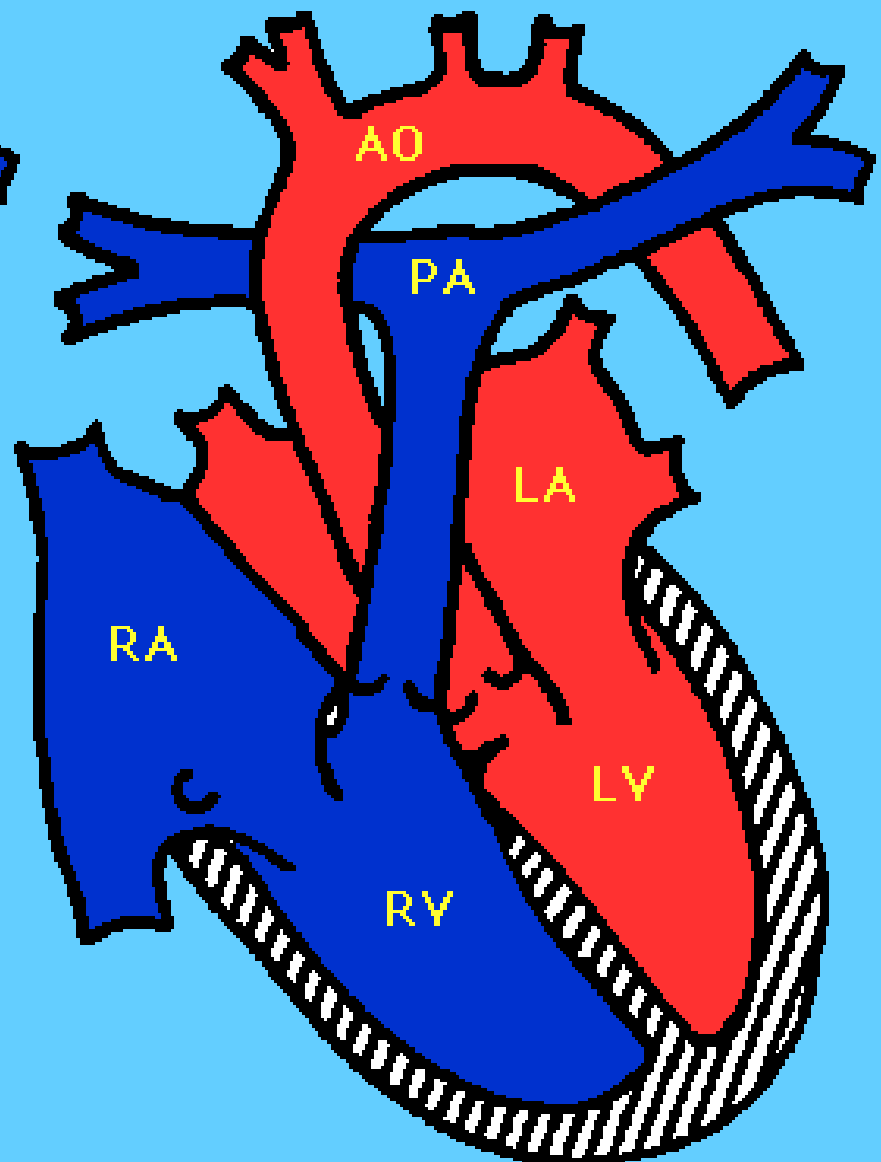
2.2 Aortic Stenosis

- Valvular - the commonest
- Supra valvular
- Subvalvular (subaortic)

Subvalvular Aortic Stenosis

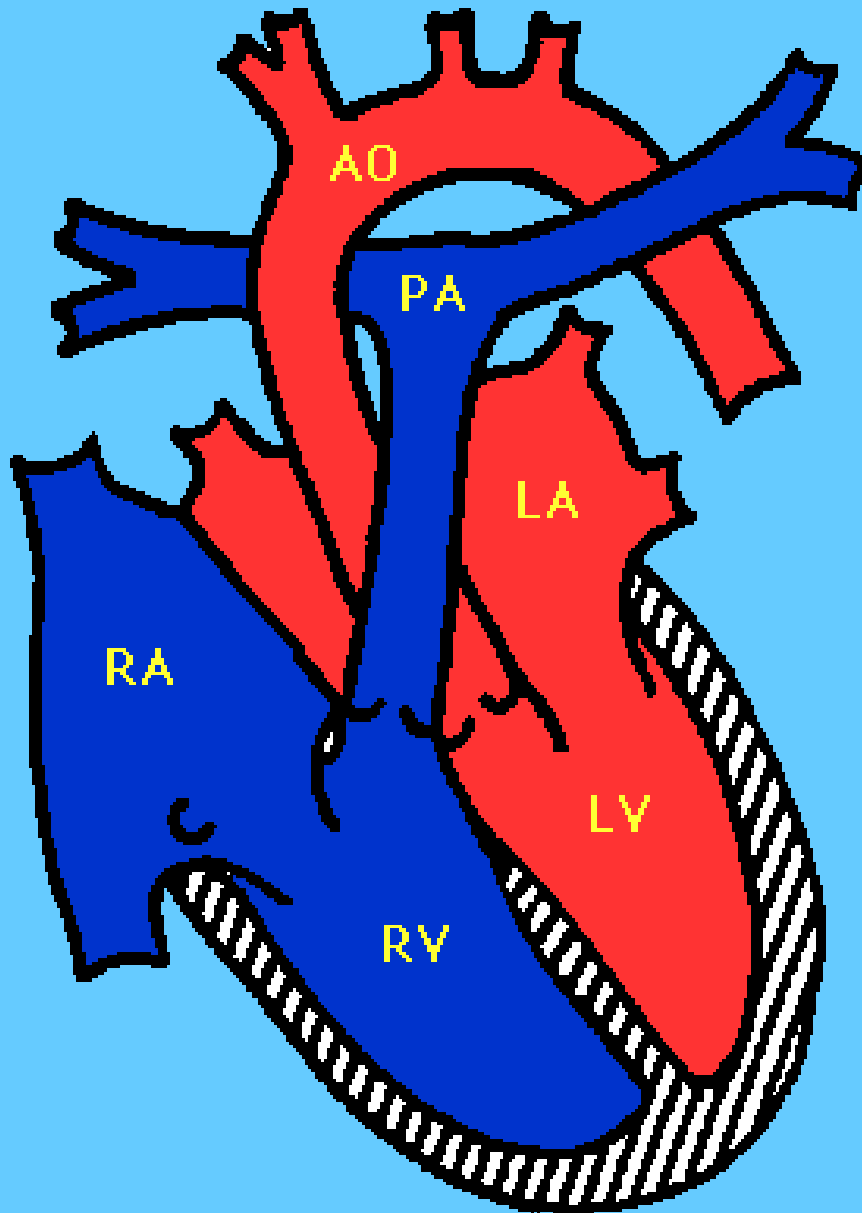


Normal

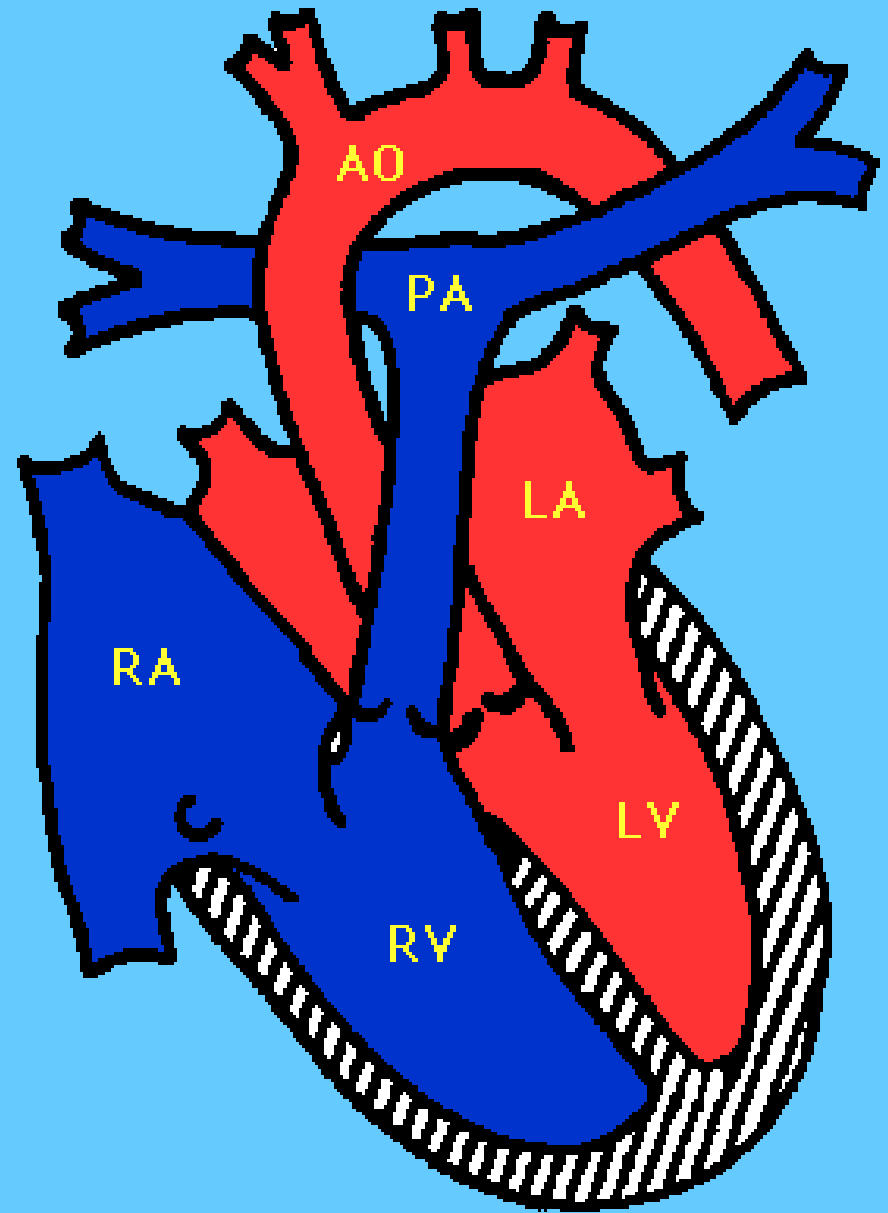


Subvalvular Aortic Stenosis

Aortic Valve Stenosis

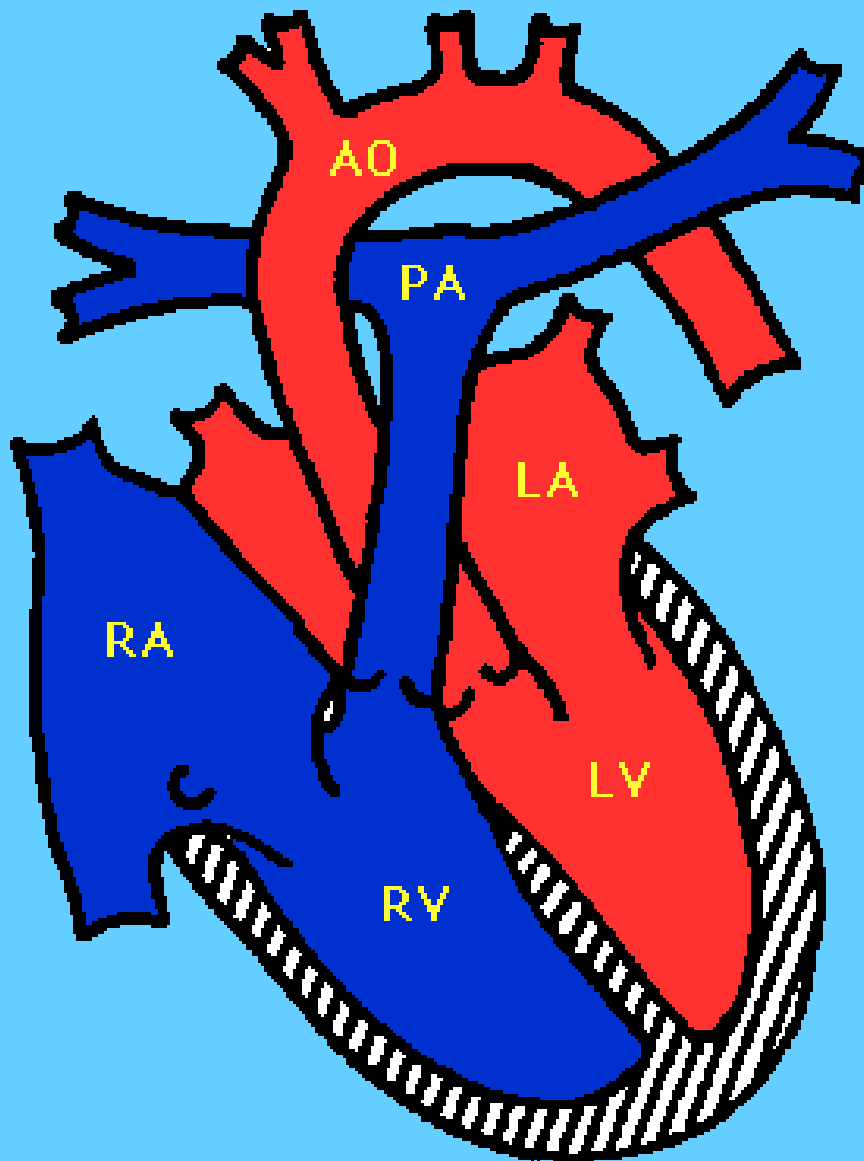


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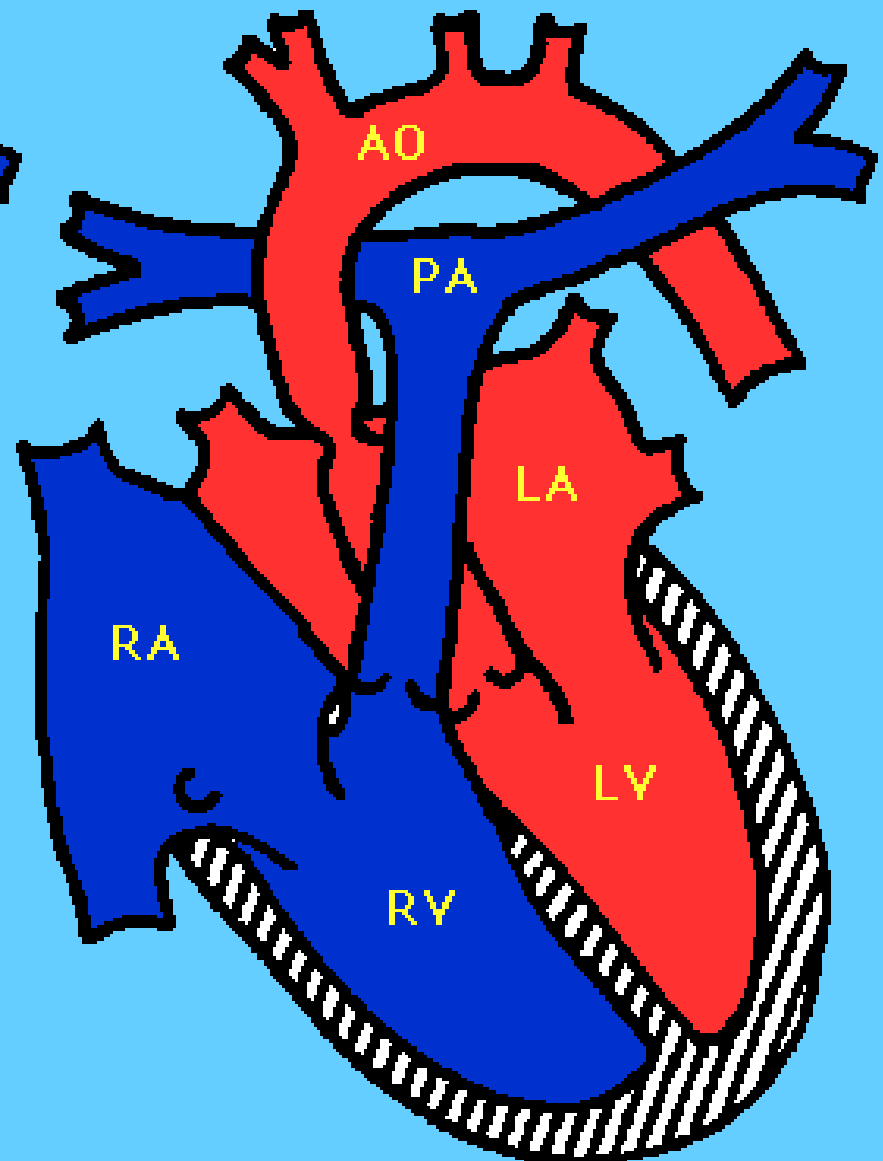


Aortic Valve Stenosis

Supravalvular Aortic Stenosis



Normal



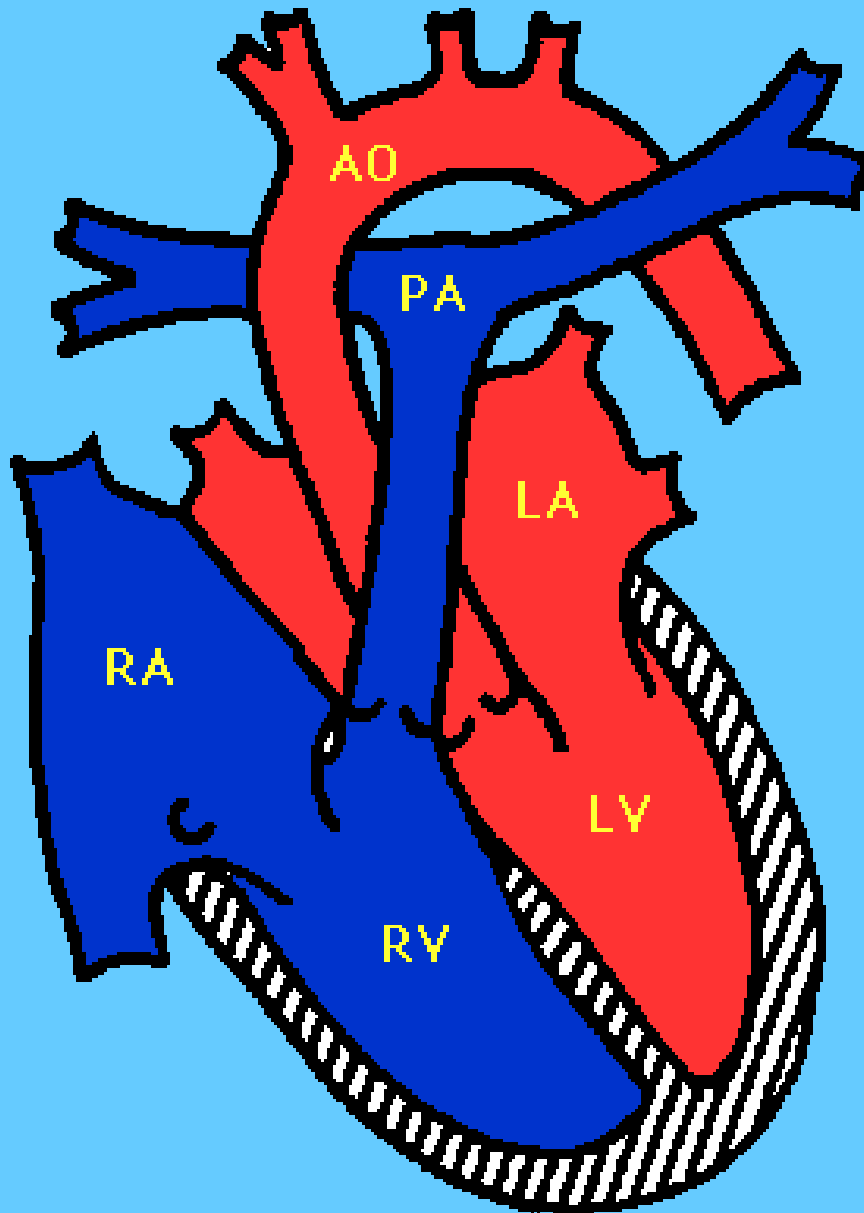
Supravalvular Aortic Stenosis

Acyanotic CHD...

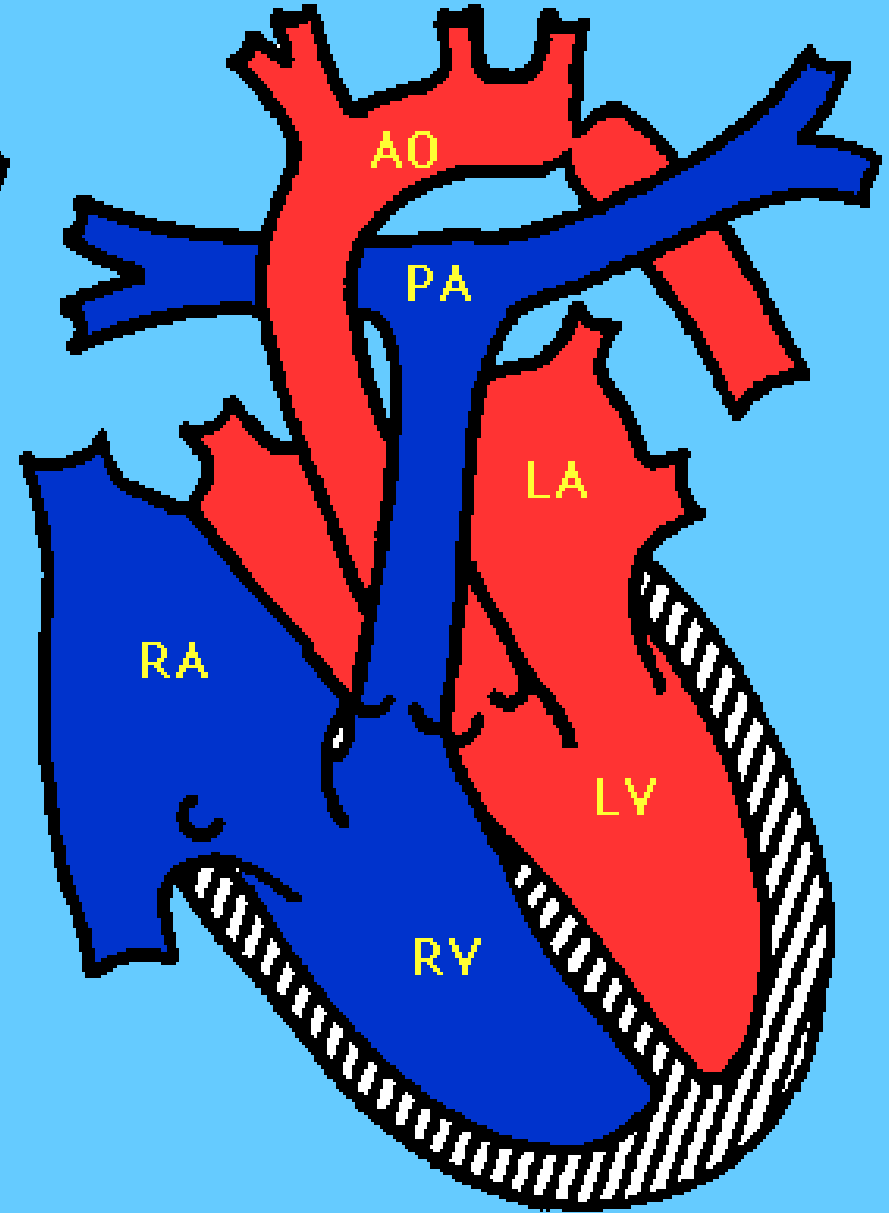
2.3 **Coarctation of the Aorta**

- Occur at any site from the arch of aorta to iliac bifurcation
- 98% of it is juxta ductal

Coarctation of the Aorta



Normal



Coarctation of the Aorta

Acyanotic CHD...

3. Pulmonary Vascular Disease

(Eismenger syndrome)

- Occur in shunt lesions

VSD - mainly

ASD

PDA

- Reversal of shunt due to pulm.

HTN

→ Cyanosis

Acyanotic CHD...

4. Regurgitant Lessons

- Pulmonary valvular insufficiency
- Congenital mitral valve insufficiency
- Mitral valve prolapse
- Tricuspid regurgitation

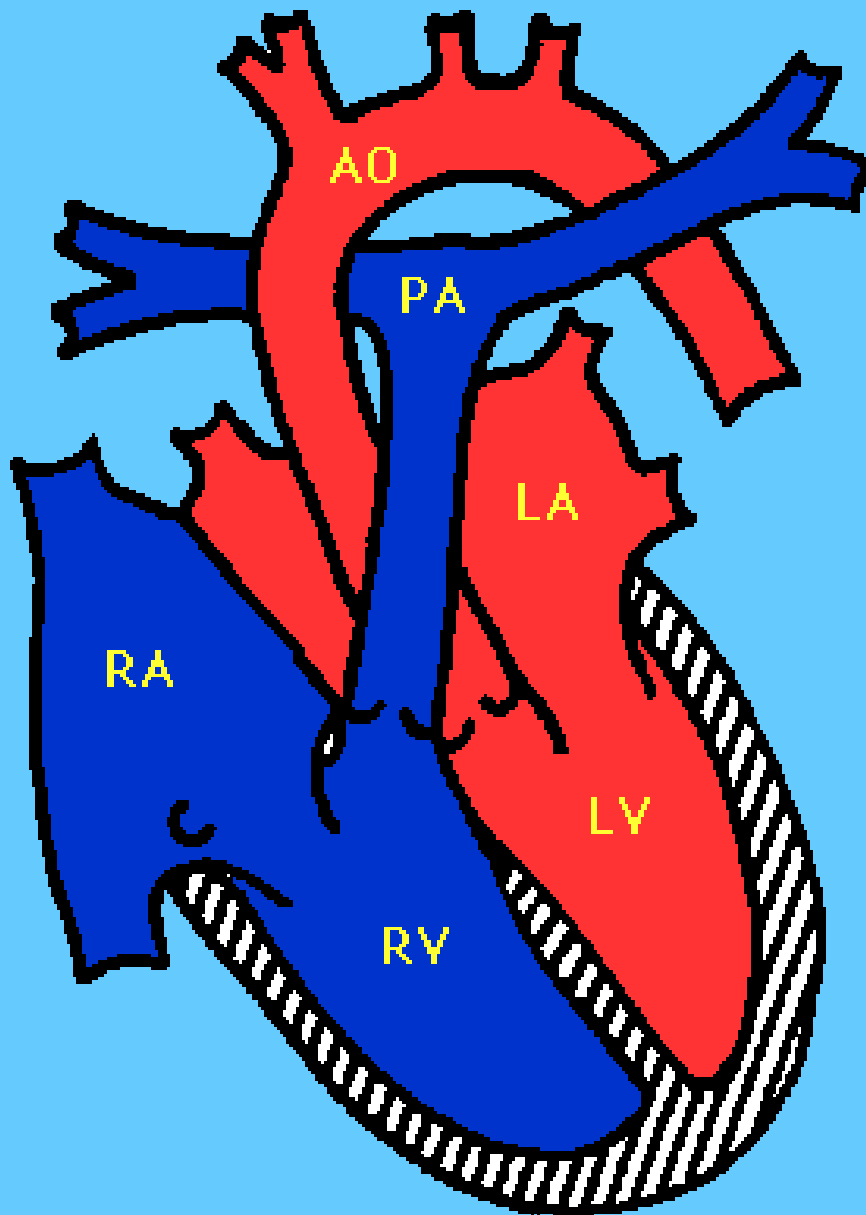
II. Cyanotic Congenital Heart Disease

1. Cyanotic lesions with decreased pulmonary blood flow

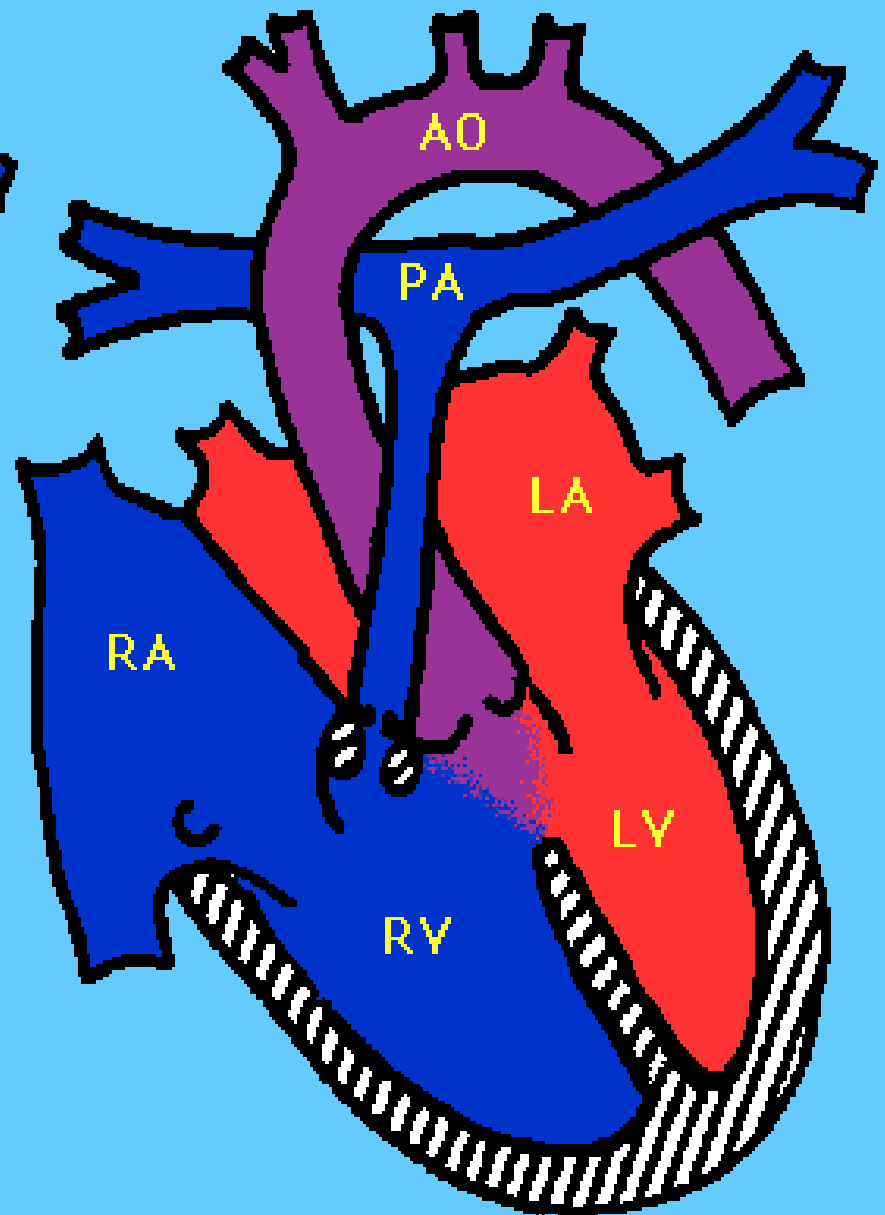
1.1 Tetralogy of Fallot

- ☞ **Consists:**
1. Rt ventricular outflow obst.
 2. Ventricular septal defect
 3. Dextroposition of the aorta
 4. Right ventricular hypertrophy

Tetralogy of Fallot

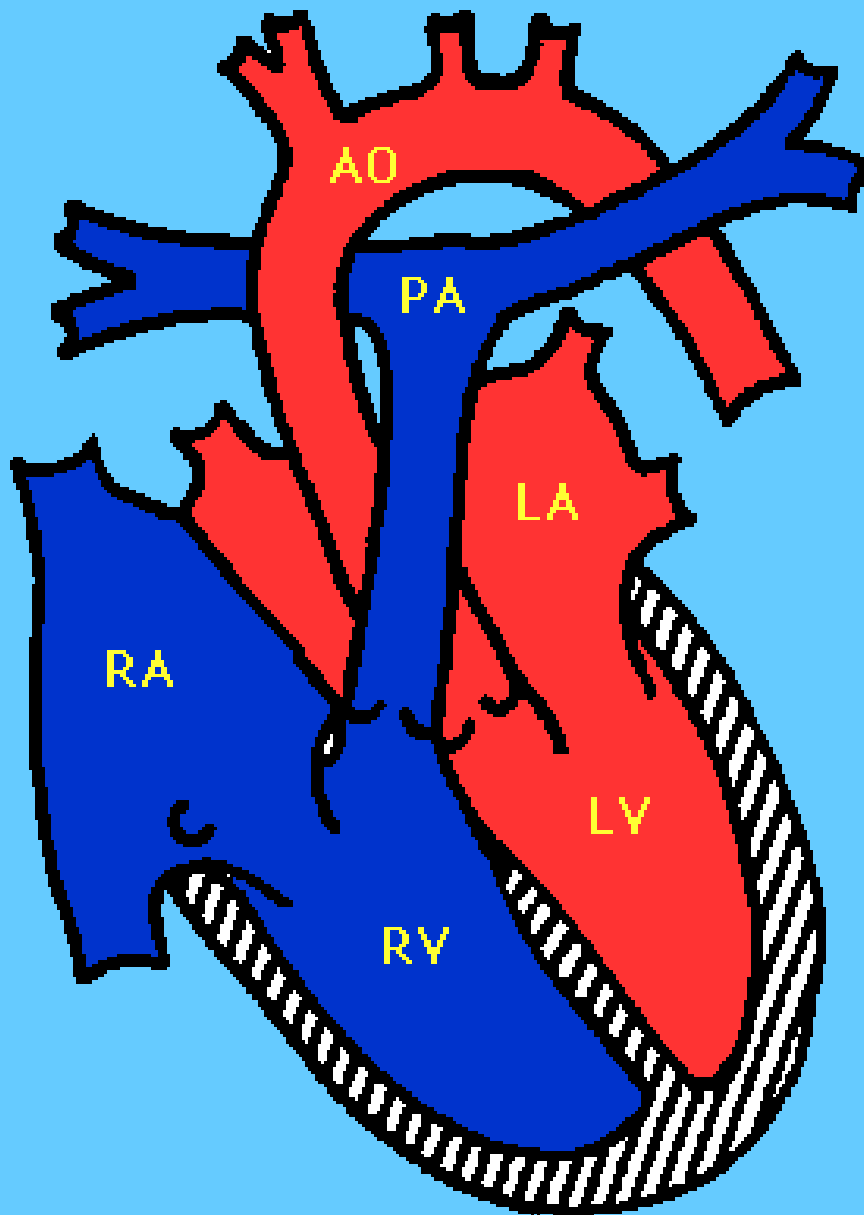


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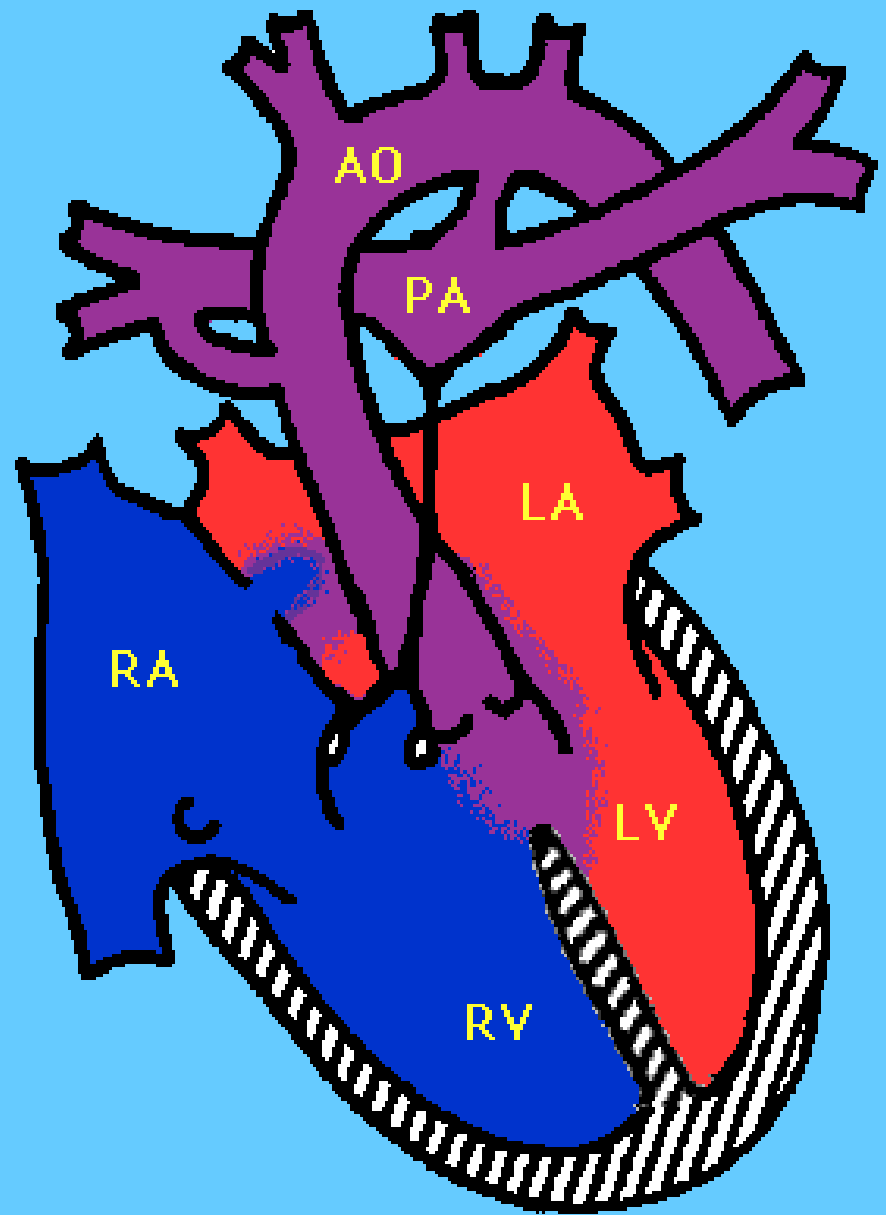


Tetralogy of Fallot

Pulmonary Atresia with VSD



Normal



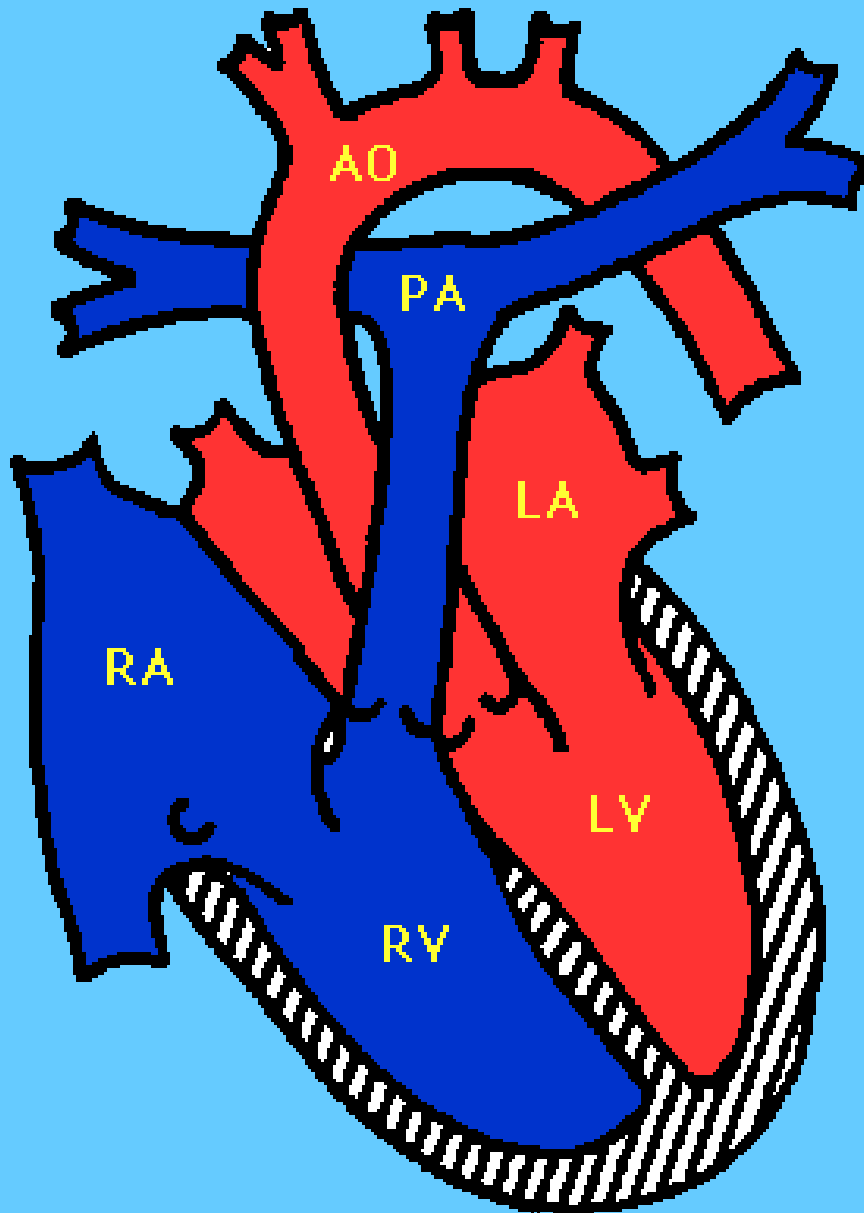
Pulmonary Atresia with VSD

Cyanotic CHD...

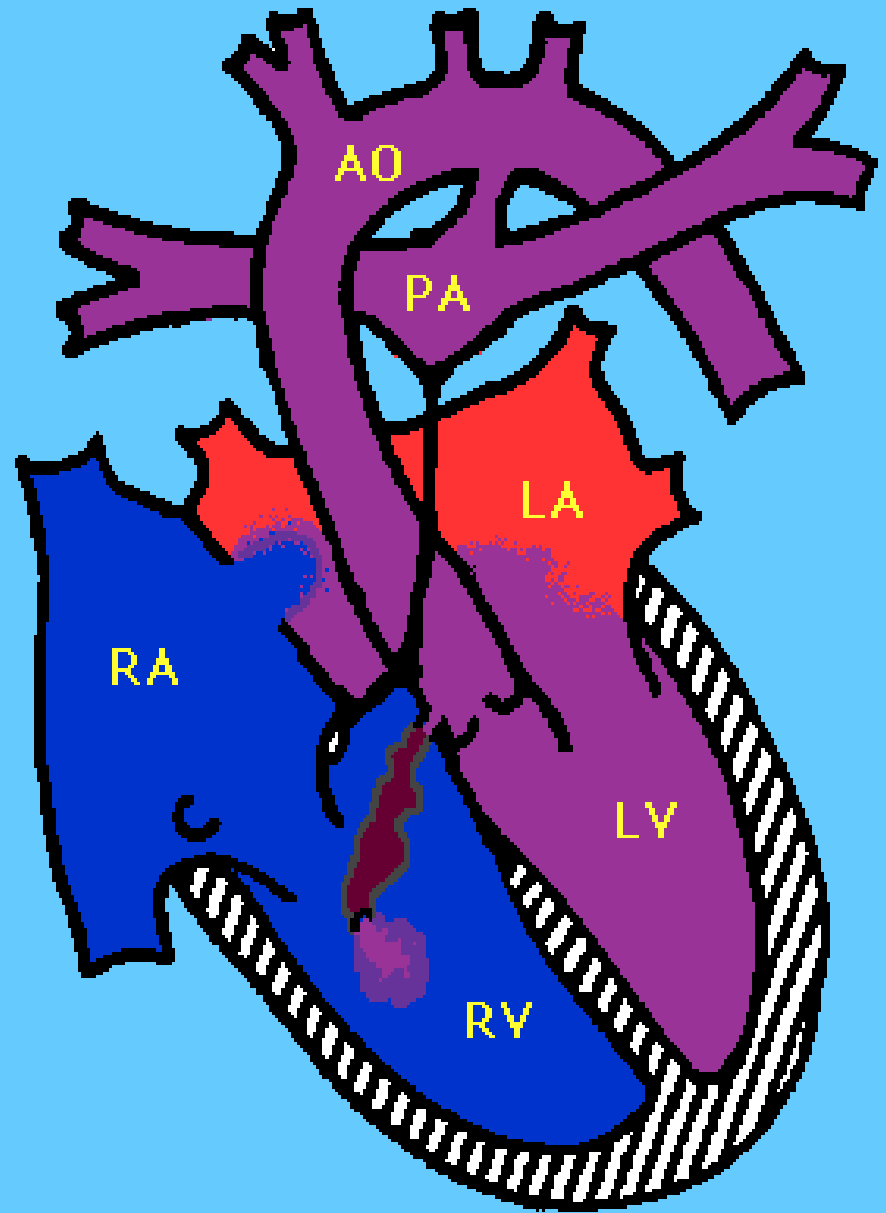
1.2 Pulmonary Atresia

- With VSD - Extreme form of TOF
- Without VSD - No egress of blood from Rt vent.
atrium
 - Shunt through foramen ovale to Lt

Pulmonary Atresia without VSD



Normal

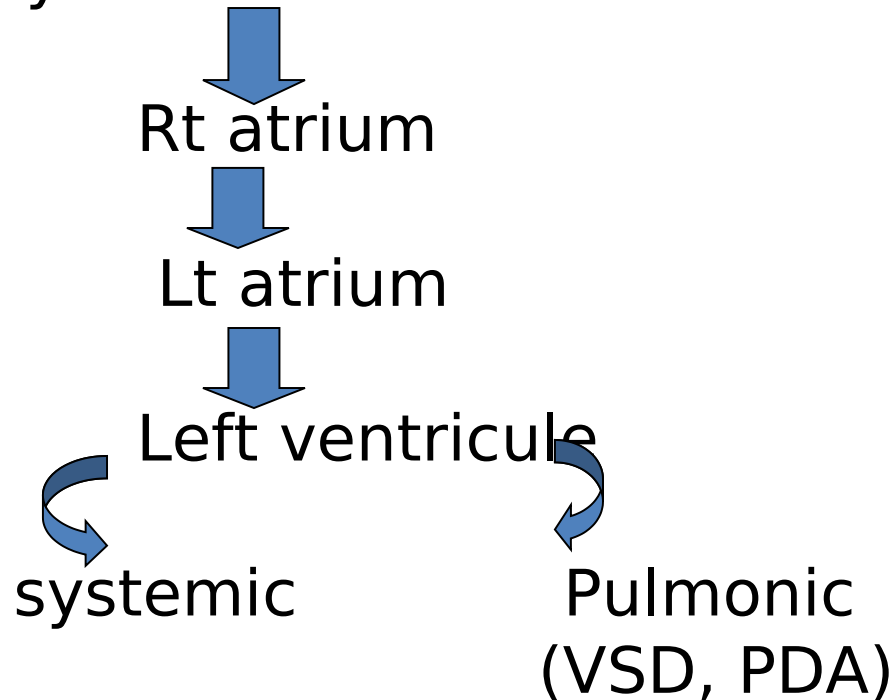


Pulmonary Atresia without VSD

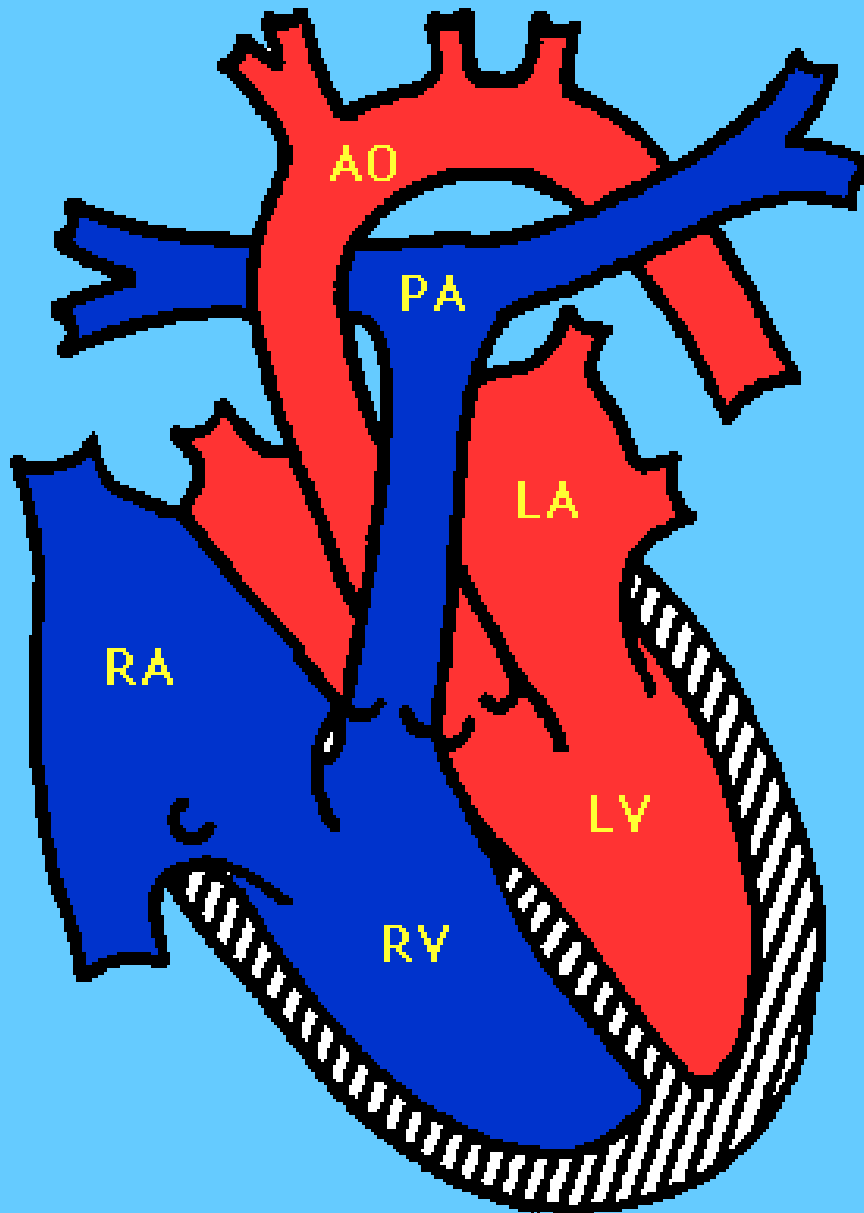
Cyanotic CHD...

1.3 Tricuspid atresia

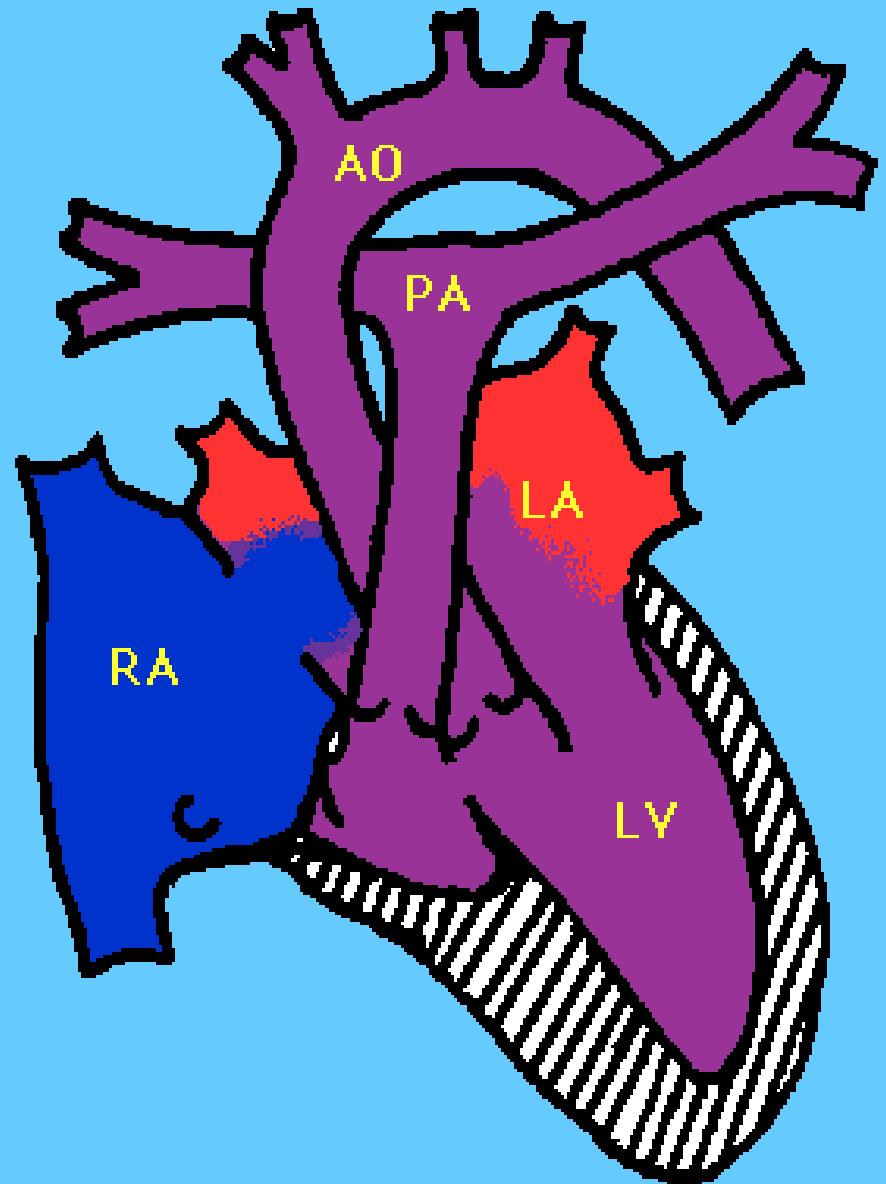
- No outlet from Right atrium to right vent.
- Systemic venous return



Tricuspid Atresia



Normal



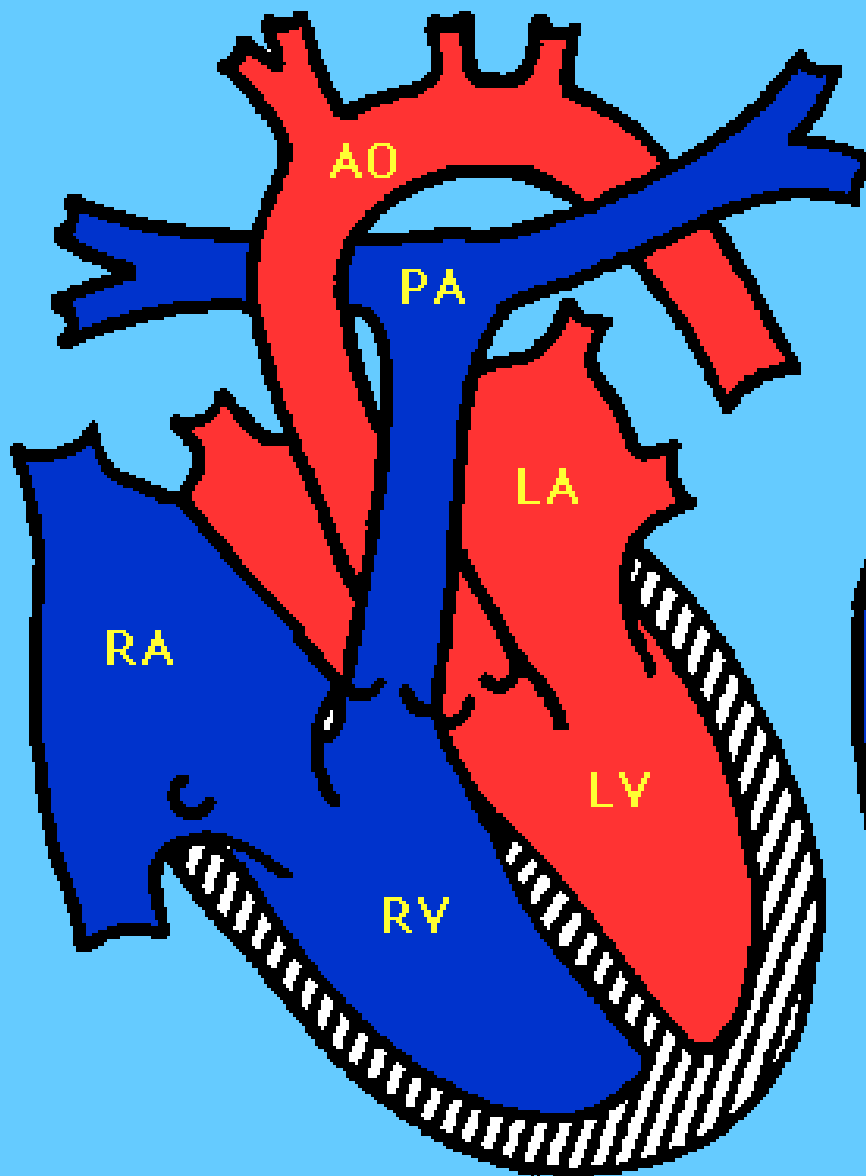
Tricuspid Atresia

Cyanotic CHD...

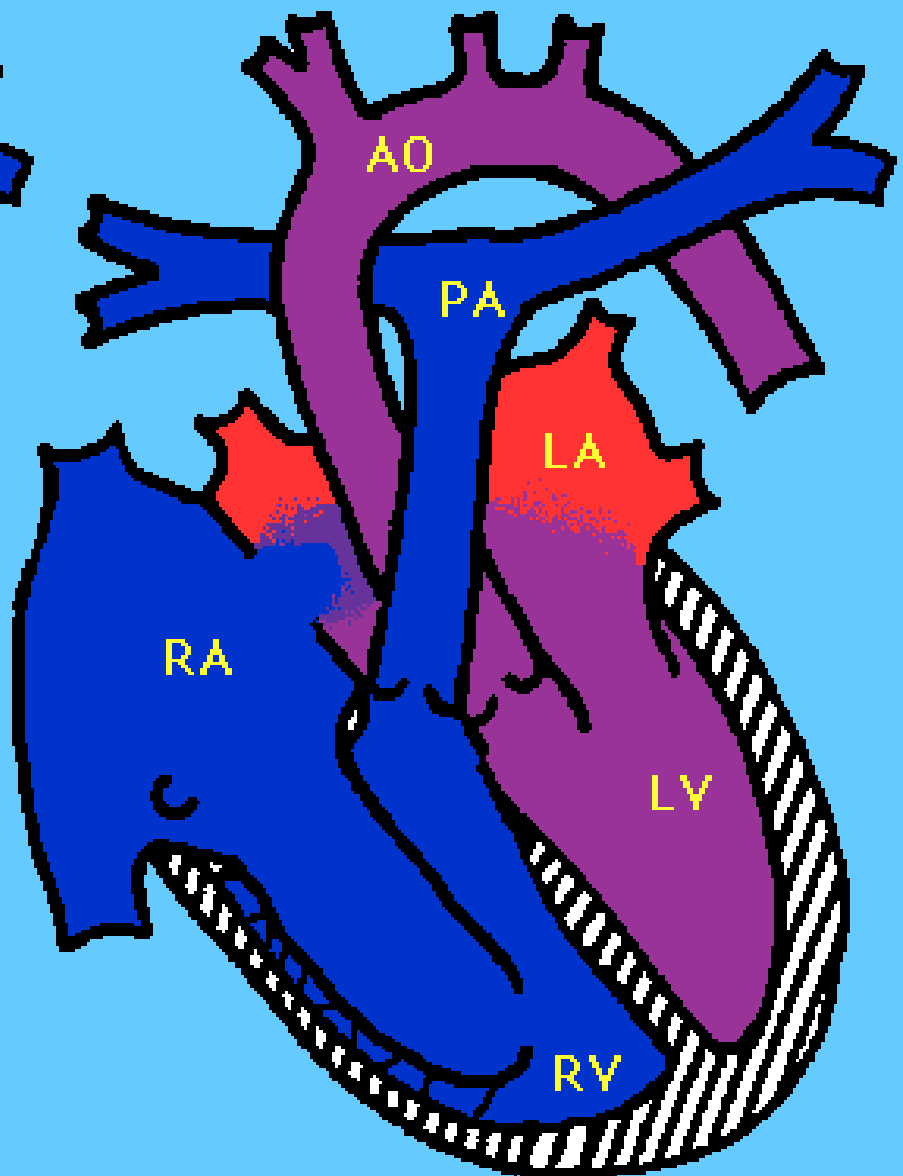
1.4 Ebstein Anomaly of the tricuspid valve

- Down ward displacement of the tricuspid valve
- Right ventricle with two parts
 - atrialized
 - normal ventricular myocardium
- Abnormal tricuspid valve
- Huge Rt atrium
- Tricuspid regurgitation
- Compromised Rt ventricular function

Ebstein Anomaly



Normal



Ebstein Anomaly



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Cyanotic CHD...

2. Cyanotic CHD With increased pulmonary blood flow

2.1 Transposition of GA

a. D -TGA (uncorrected)

- Systemic venous return to Rt atrium

Normal

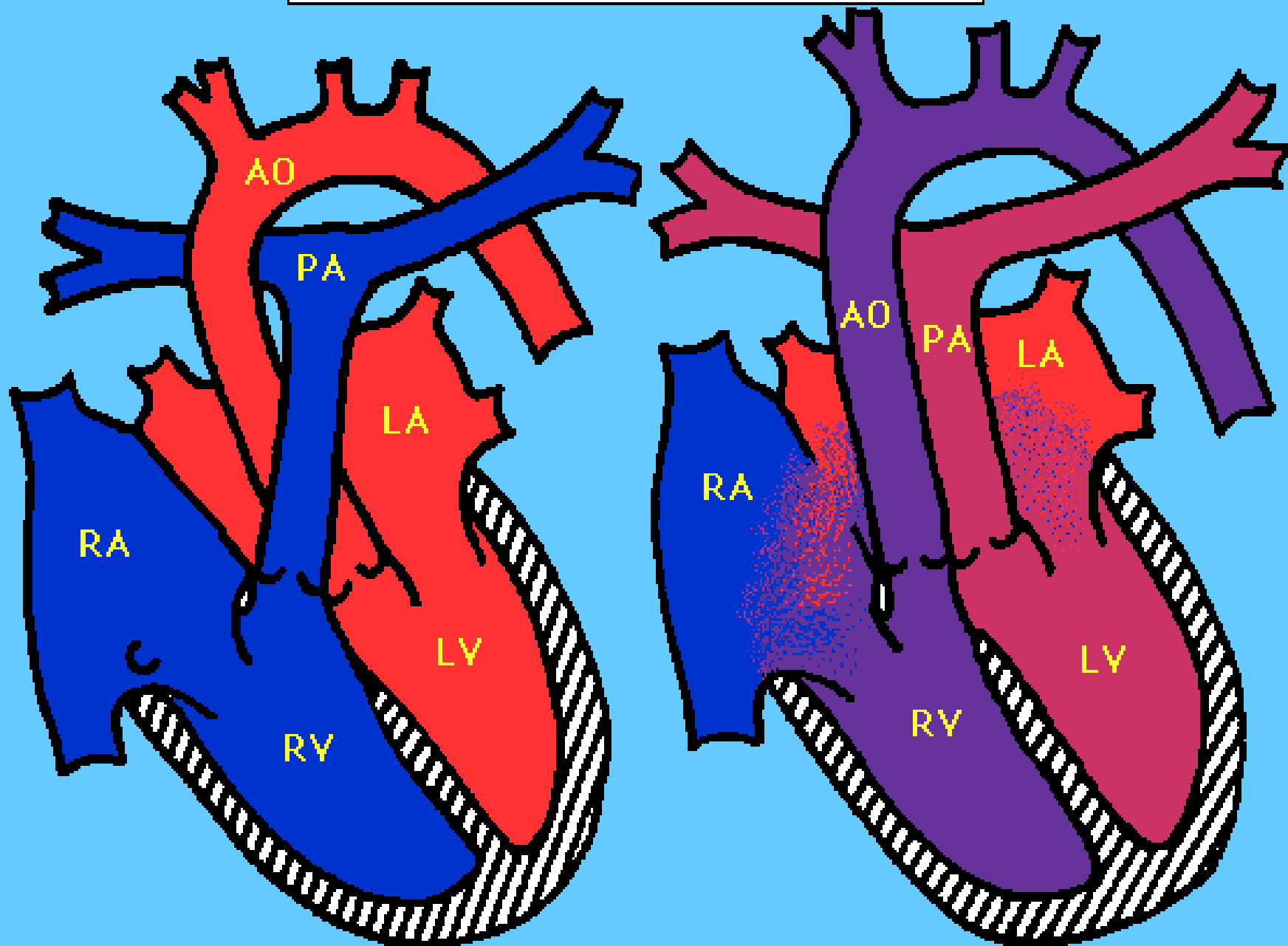
- Pulmonary venous return to Lt atrium

- Aorta arises from Right ventricle

- Pulm. artery arises from Lt vent.

Pathology

Transposition of the Great Vessels



Normal

Transposition of the Great Vessels

Cyanotic CHD...

2.2 Truncus arteriosus

- Single arterial trunk for both pulm. & systemic circ.
- 4 types depending the origin of pulmonary artery

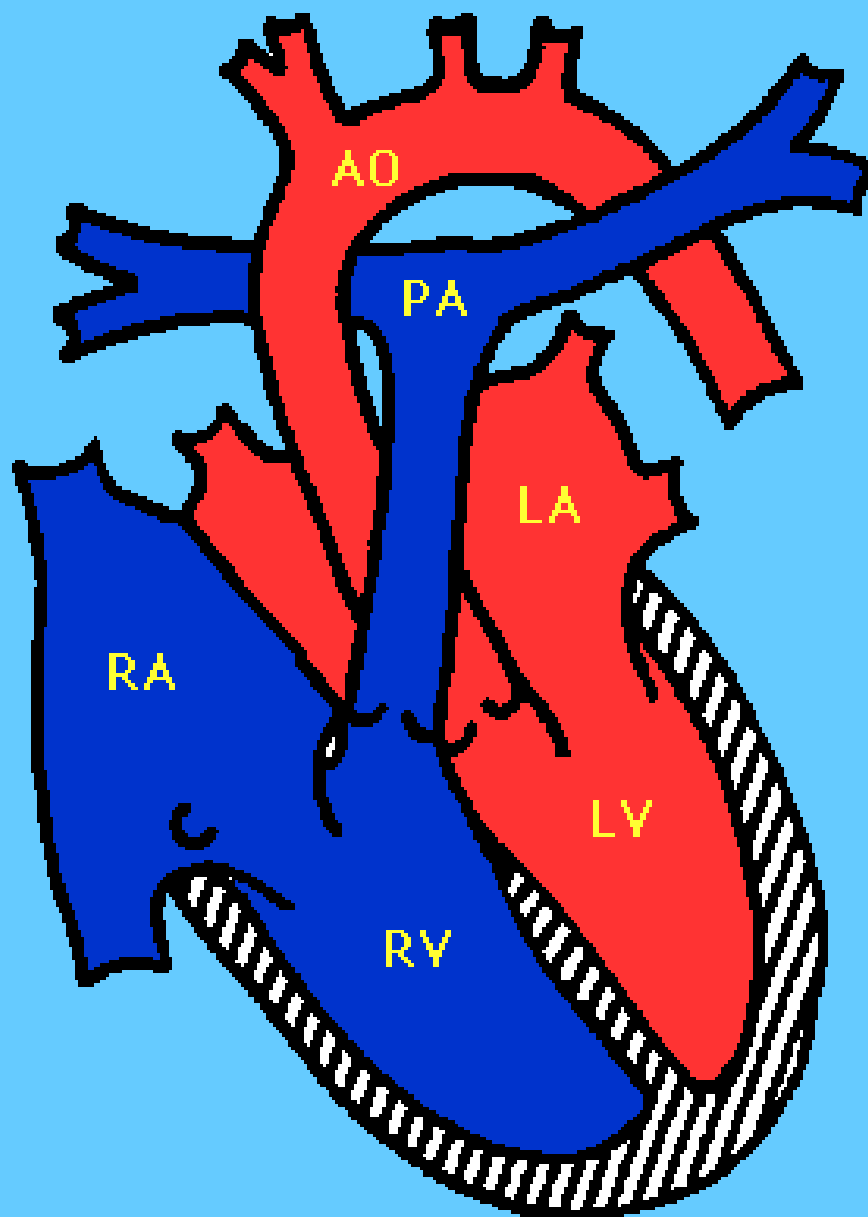
Clinical Manifestation

- Cyanosis
- CHF
- Systolic ejection m

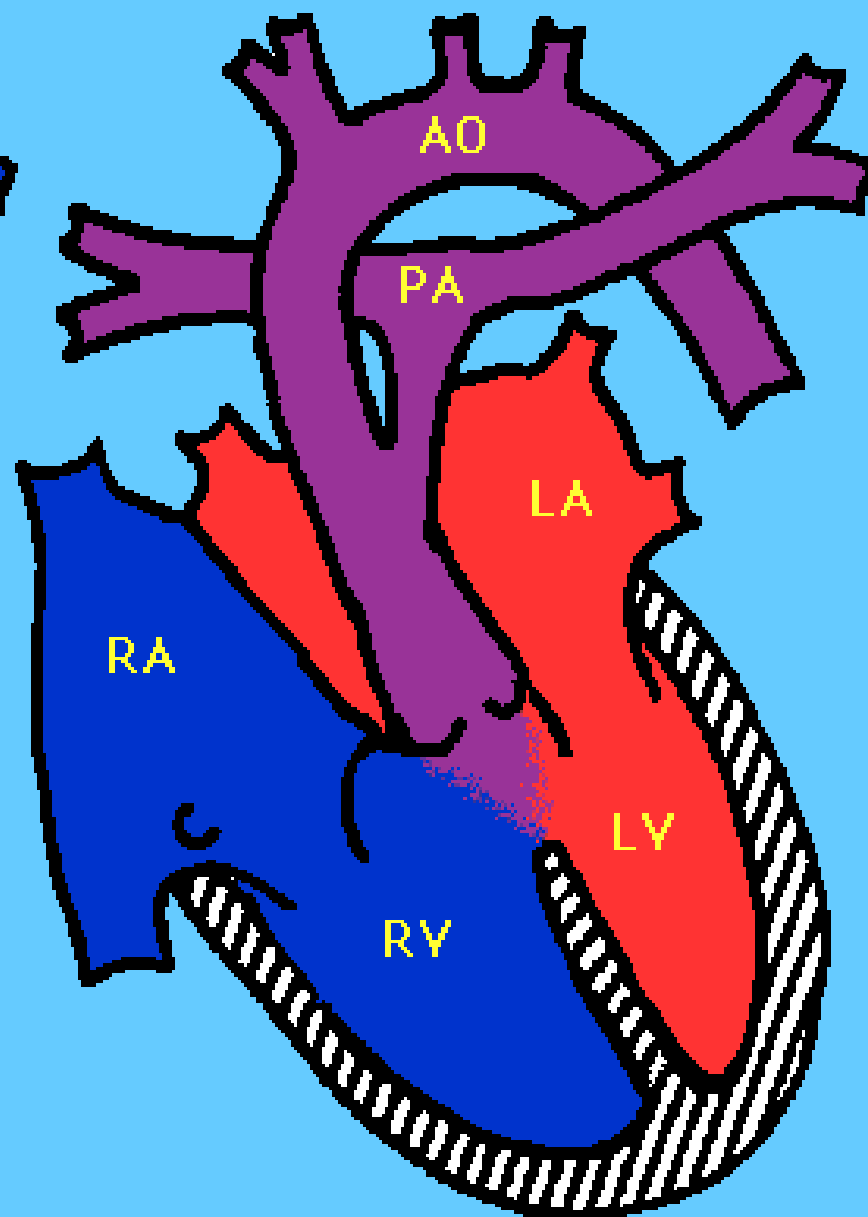
at 2-3rd m

Treatment - surgery

Truncus Arteriosus



Normal

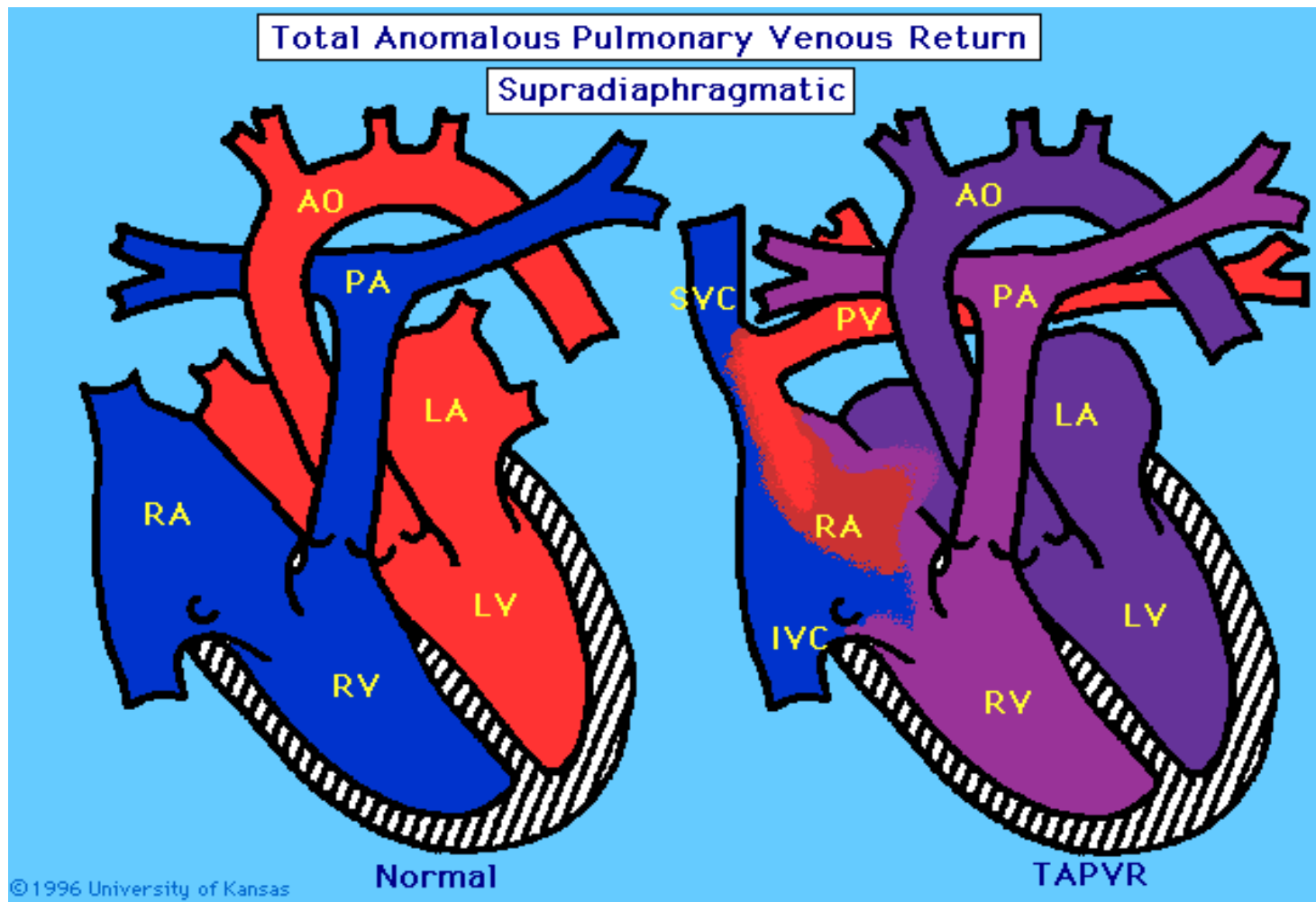


Truncus Arteriosus

Cyanotic CHD...

2.3 Total Anomalous Pulm. Venous return

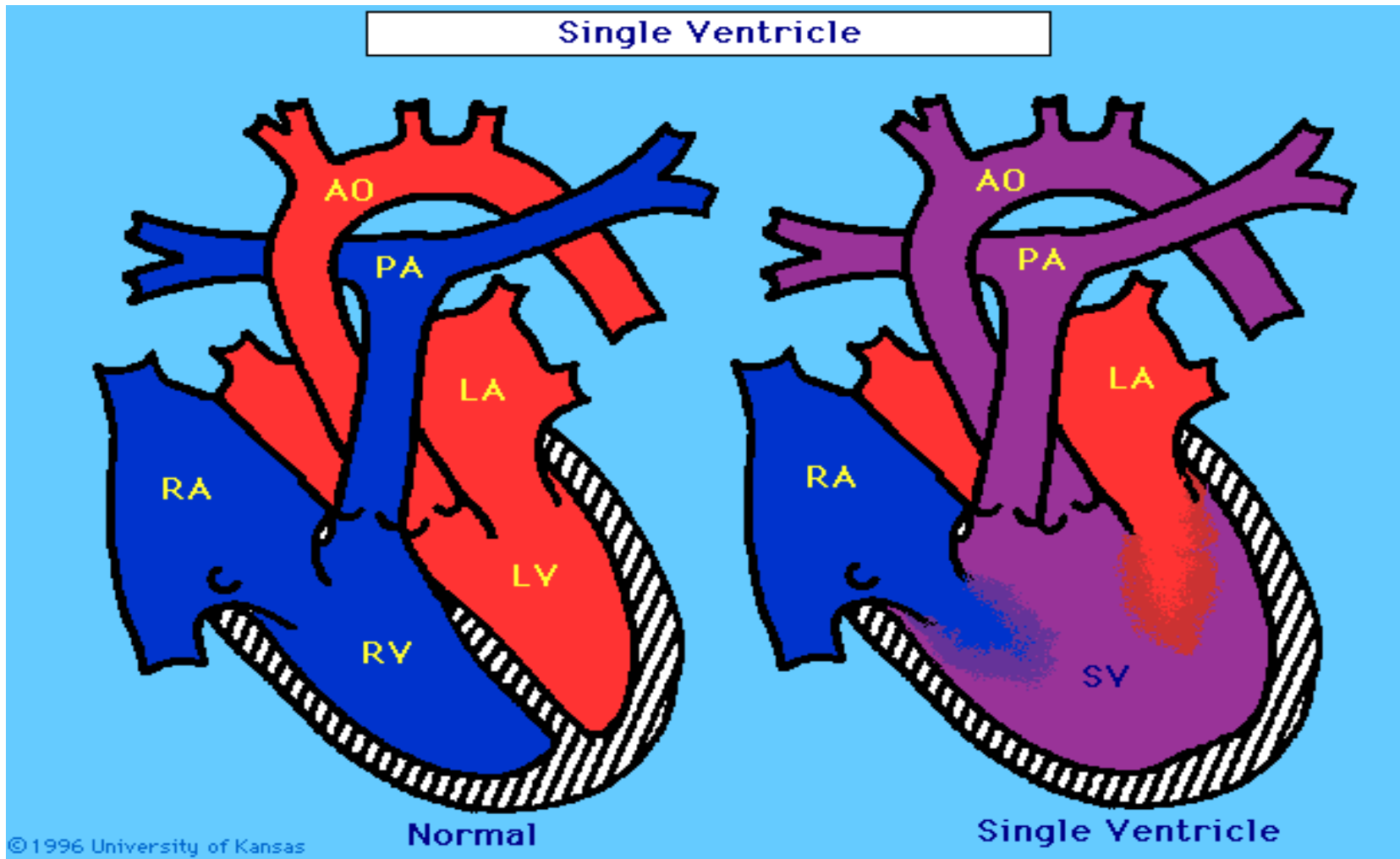
- Pulm. drainage into systemic vein



Cyanotic CHD...

2.4 Single Ventricle

- No interventricular septum

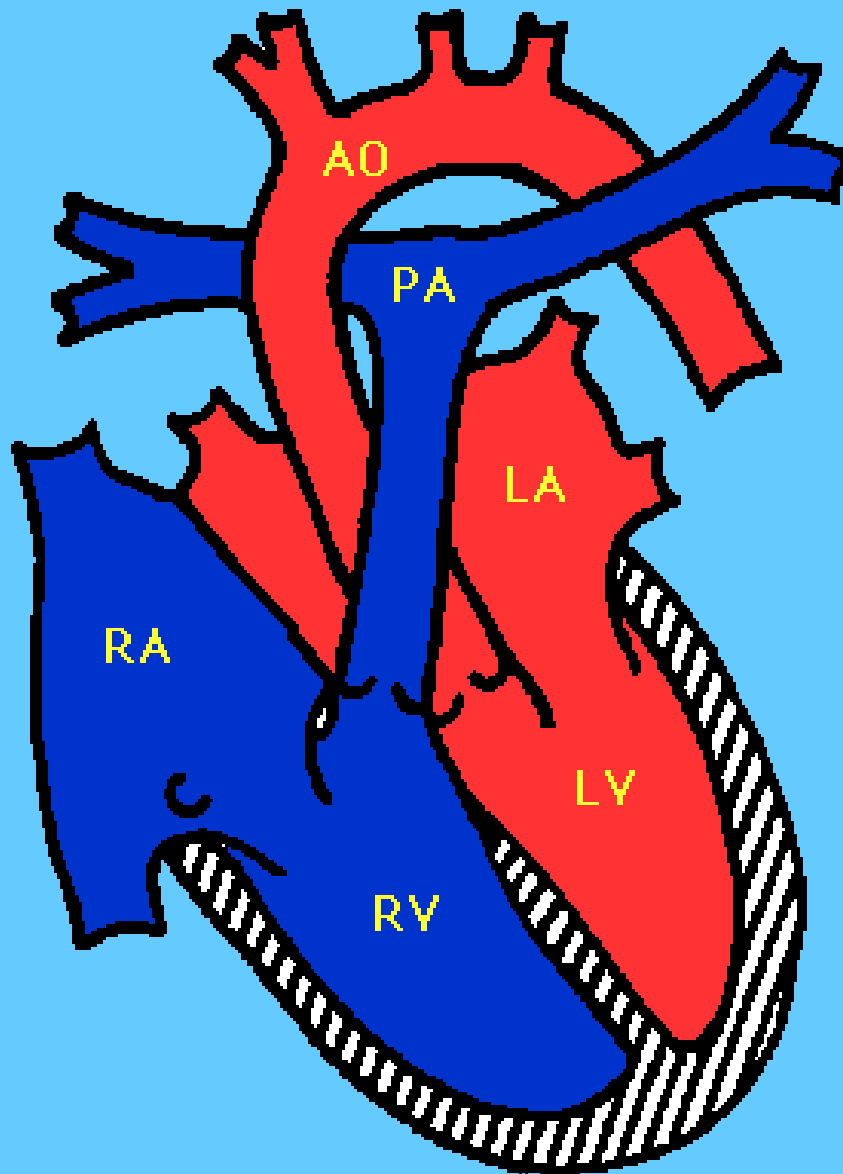


Cyanotic CHD...

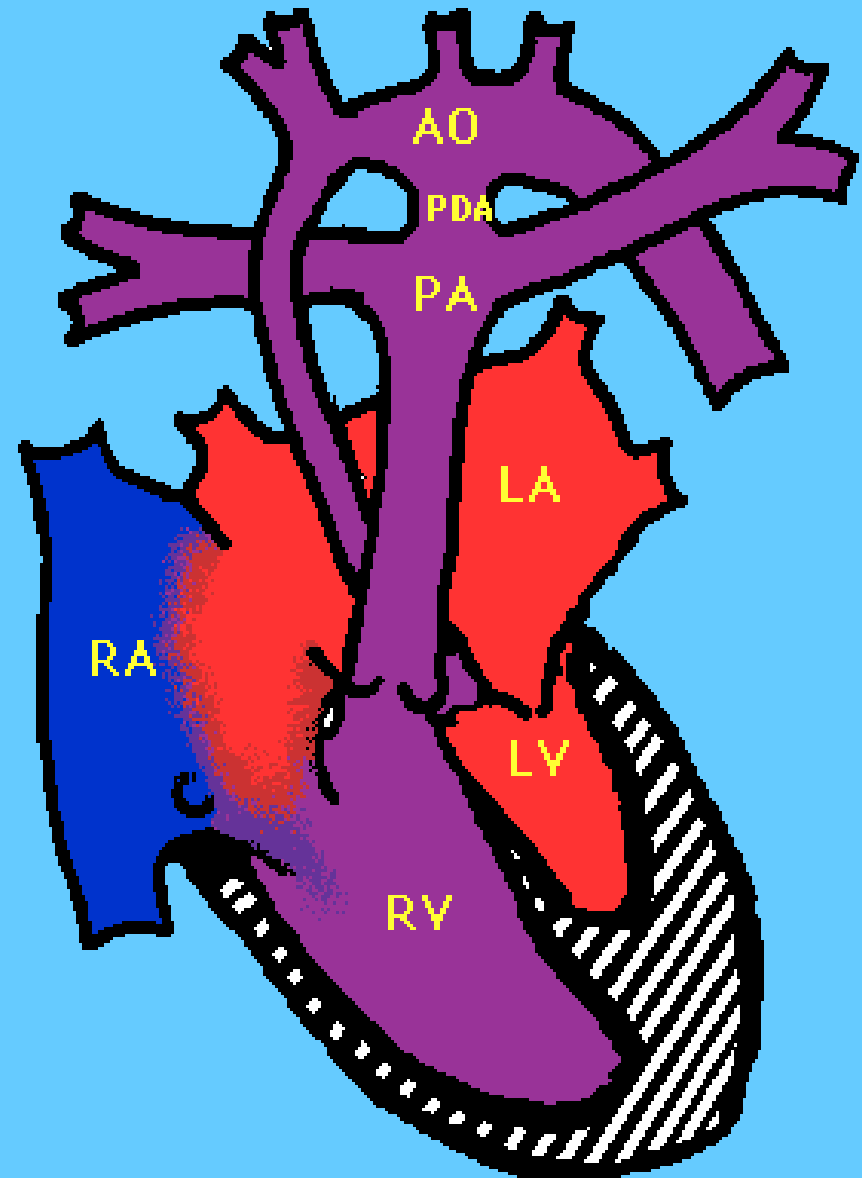
2.5 Hypoplastic Left Heart Syndrome

- Under development of Lt Side of the heart
 - Atretic aortic or mitral orifice
 - Non functional Lt ventricle
 - Hypoplasia of ascending aorta
 - ☛ Right ventricle supplies both pulm.
- &
- systemic circulation

Hypoplastic Left Heart Syndrome



Normal



Hypoplastic Left Heart

Approaching a child with congestive heart failure

CHF



CONTENT

- ☐ **Definition**
- ☐ **Etiology**
- ☐ **Pathophysiology**
- ☐ **Classification**
- ☐ **Approach to a patient with CHF**
- ☐ **Clinical manifestation**
- ☐ **Diagnosis**
- ☐ **Management**

Definition (CHF)

- ❖ Heart failure is a clinical condition that results from impairment of the ventricle to fill with or eject blood.
- ❖ Heart failure occurs when the heart cannot deliver adequate cardiac output to meet the metabolic needs of the body.

ETIOLOGY

❖ Fetal

- Severe anemia
- Supraventricular Tachycardia
- Ventricular tachycardia
- Complete heart block

❖ Premature neonate

- Fluid over load
- PDA
- VSD

❖ **FULL TERM NEONATE**

- Asphyxia cardiomyopathy
- Lt sided obstructive lesion
(coarctation of aorta, hypoplastic Lt heart syndrome)
- Large mixing cardiac defects (single ventricle, truncus arteriosus)
- Viral myocarditis
- Transposition of great arteries
- Anemia

➤ Supraventricular Tachycardia

➤ Complete heart block

❖ **Infant-Toddler**

➤ Left-to-right cardiac shunts

➤ Metabolic cardiomyopathy

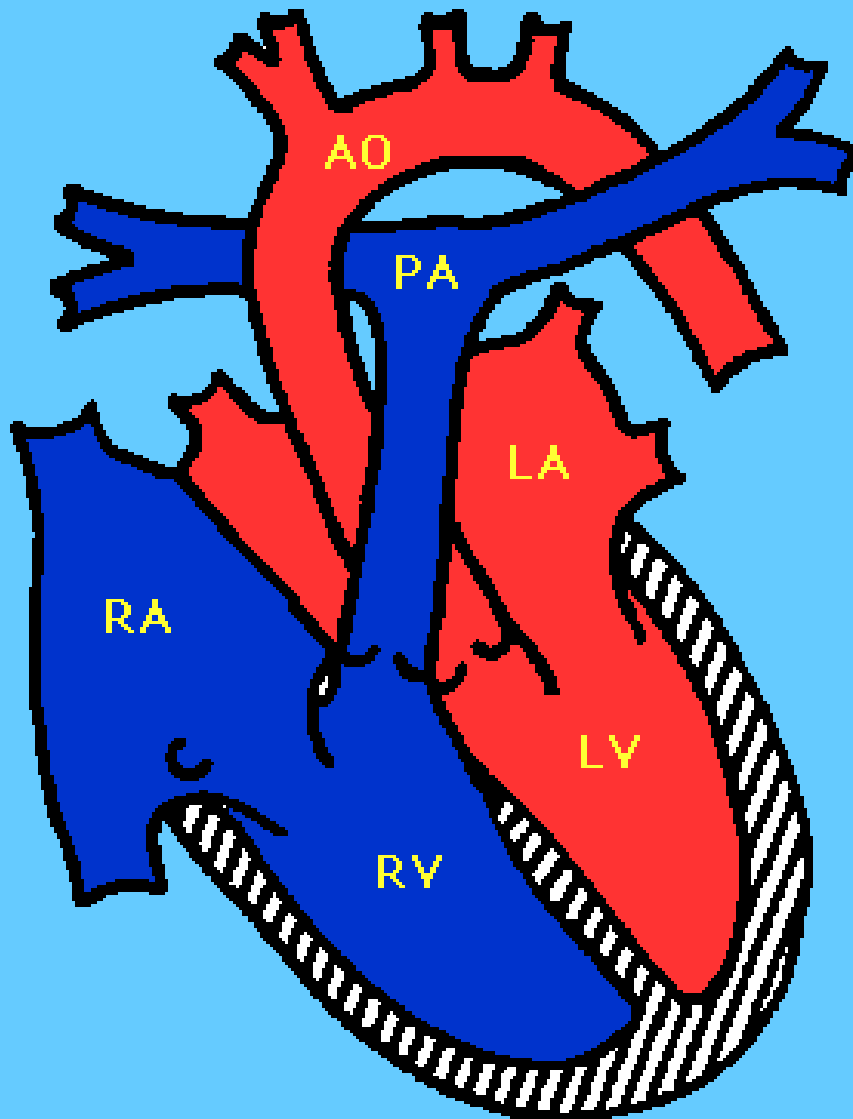
➤ Acute hypertension

➤ Supraventricular tachycardia

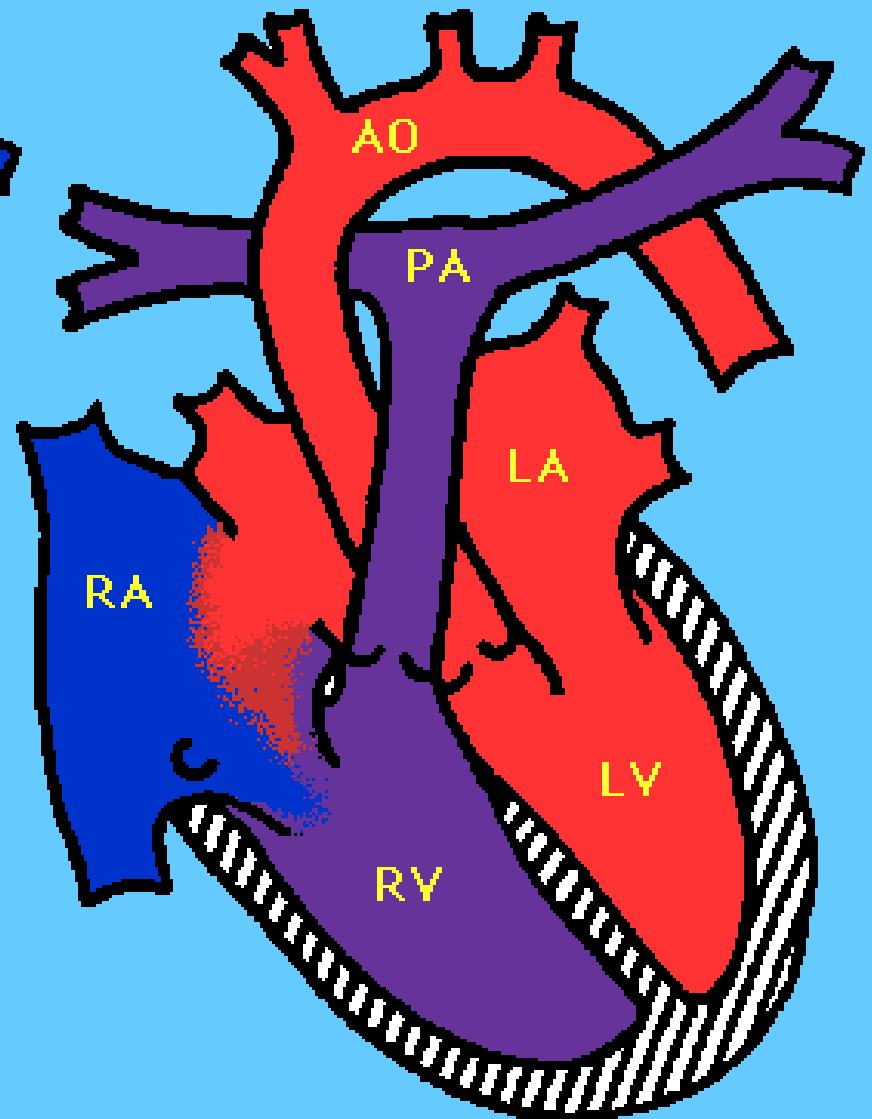
❖ **Children-Adolescents**

- L→R shunt (VSD, ASD, PDA)
- Viral myocarditis
- Rheumatic fever
- Acute hypertension
(glomerulonephritis)
- Viral myocarditis
- Thyrotoxicosis
- Endocarditic
- Cardiomyopathy(hypertrophic,
dilated)

Atrial Septal Defect

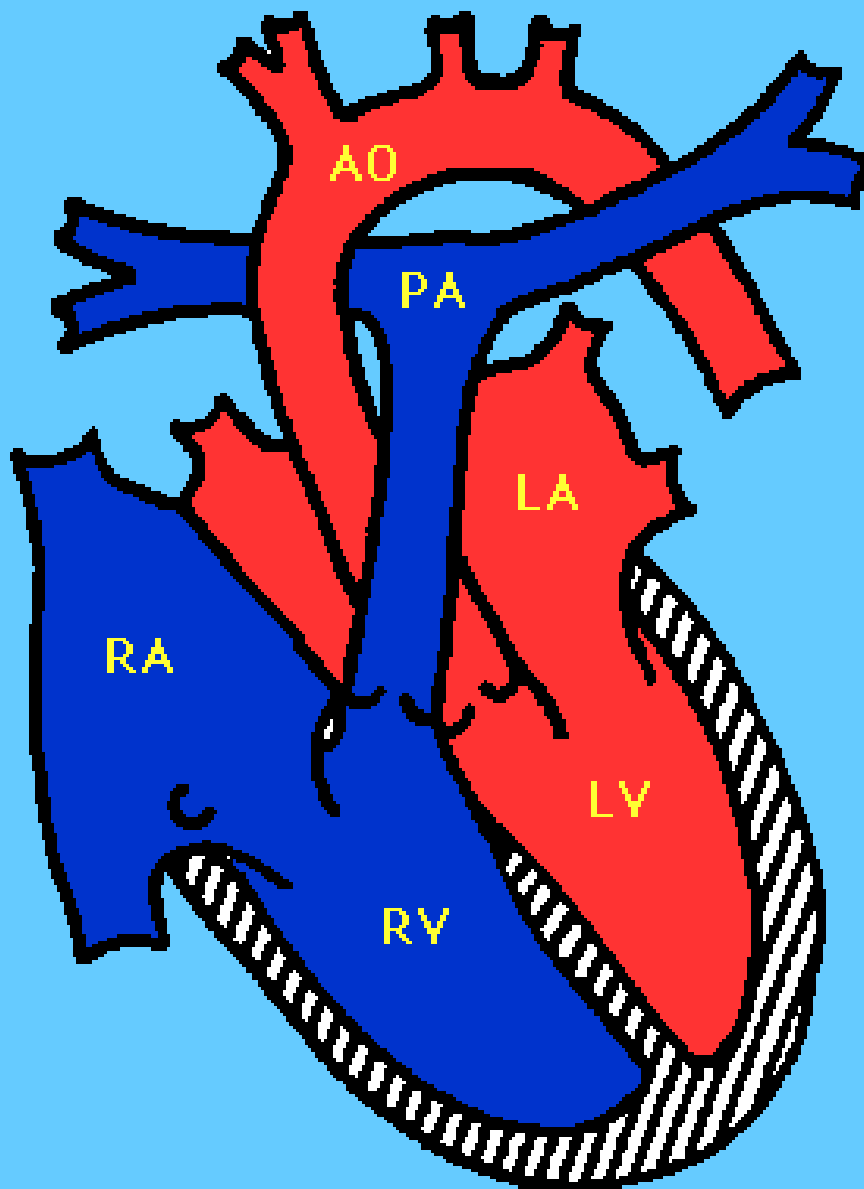


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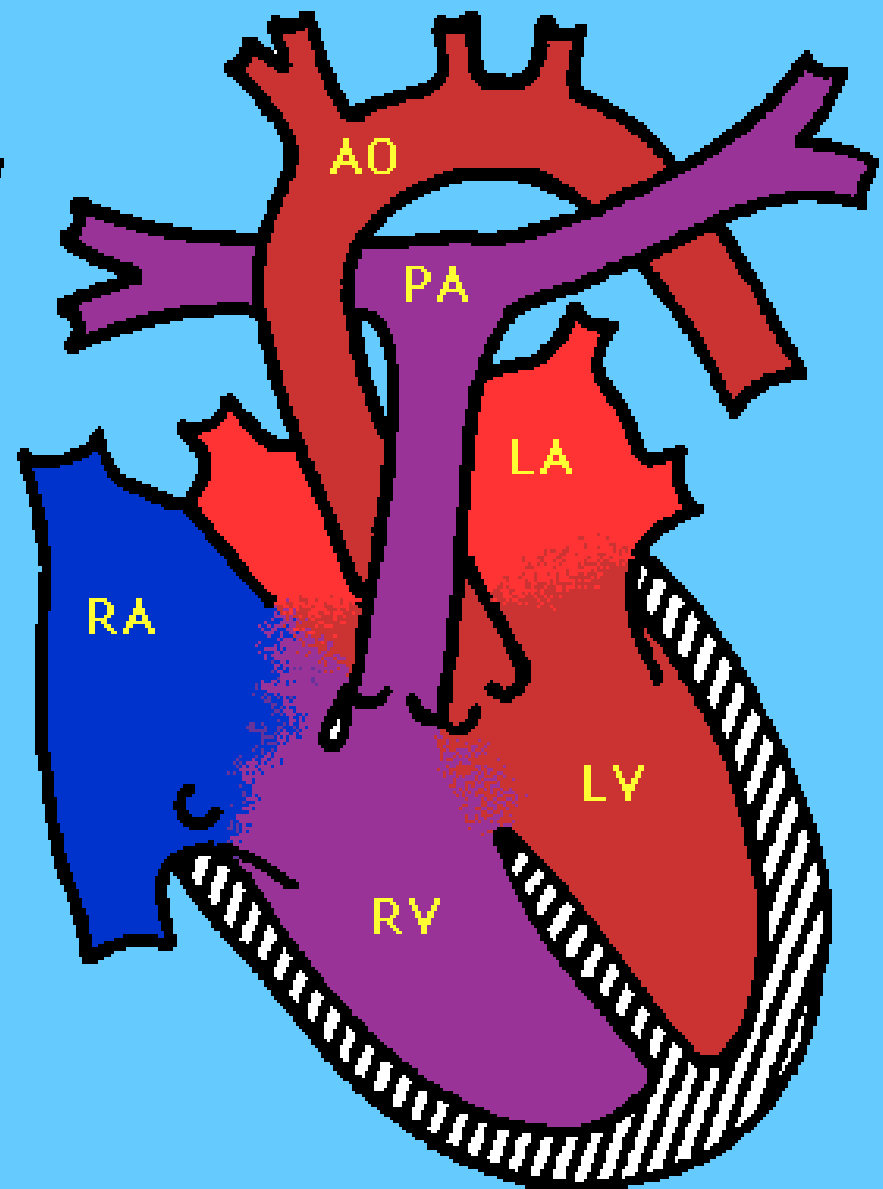


Atrial Septal Defect

Atrioventricular Canal Defect

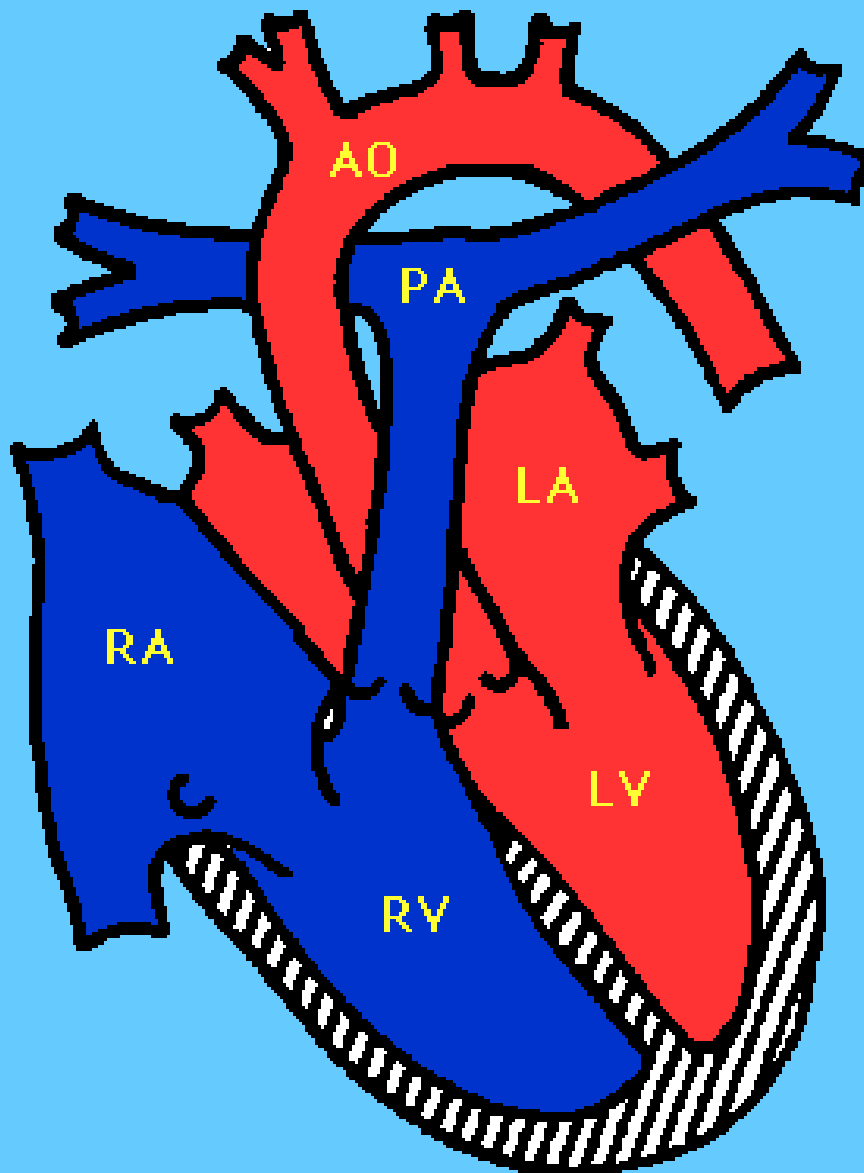


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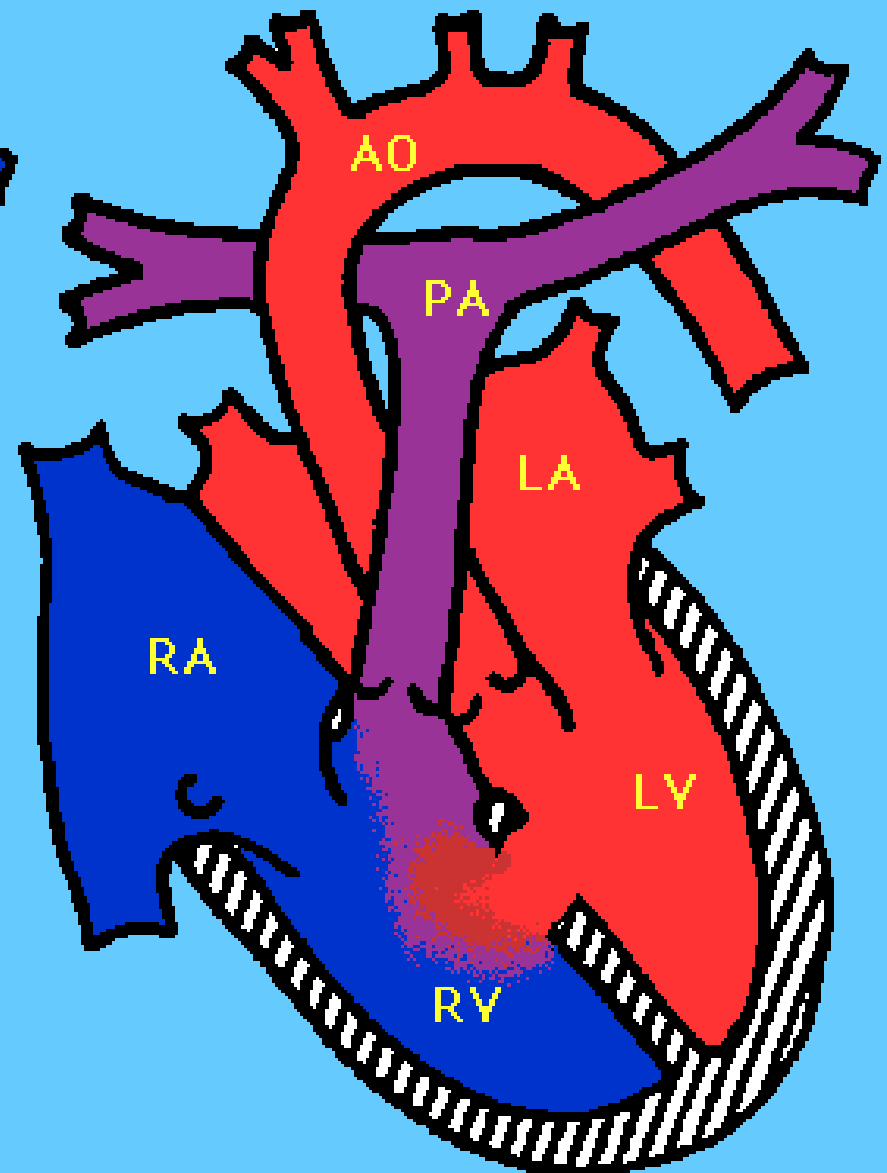


Atrioventricular Canal Defect

Ventricular Septal Defect

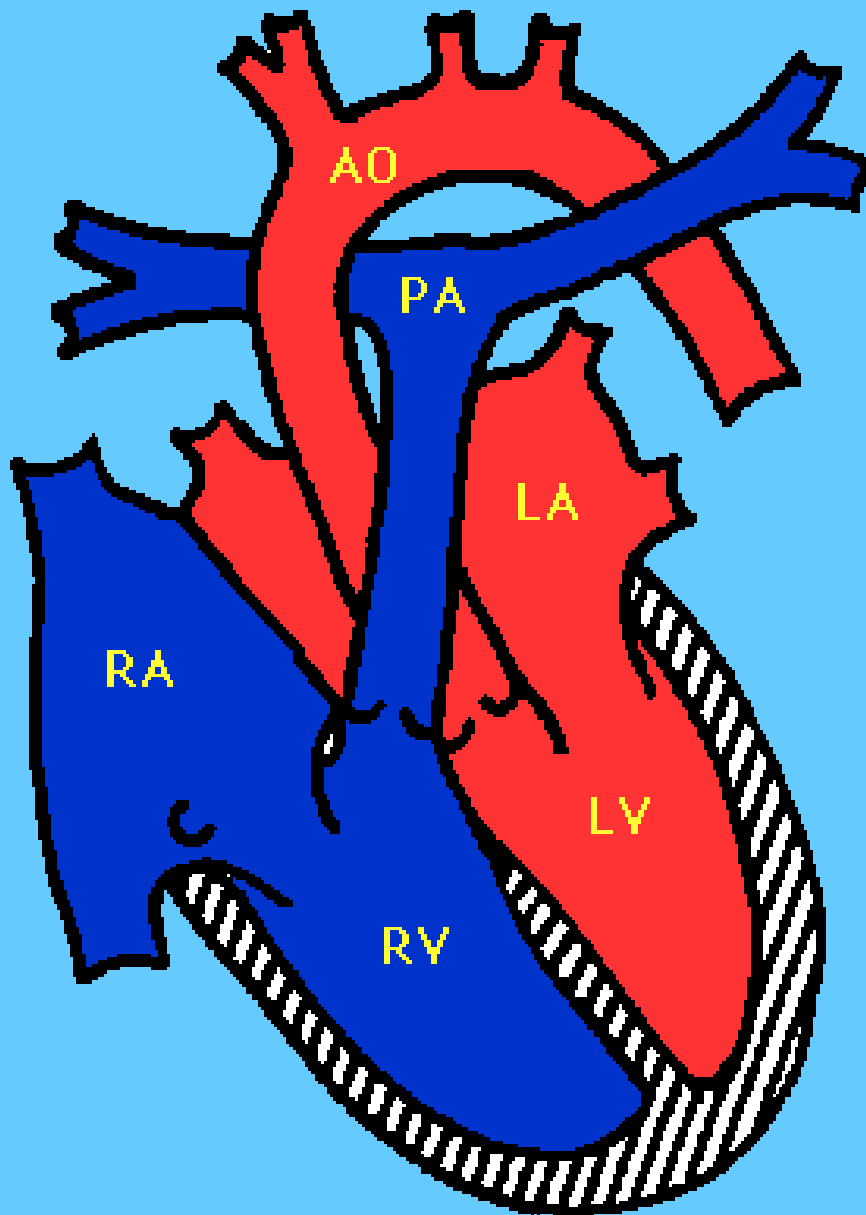


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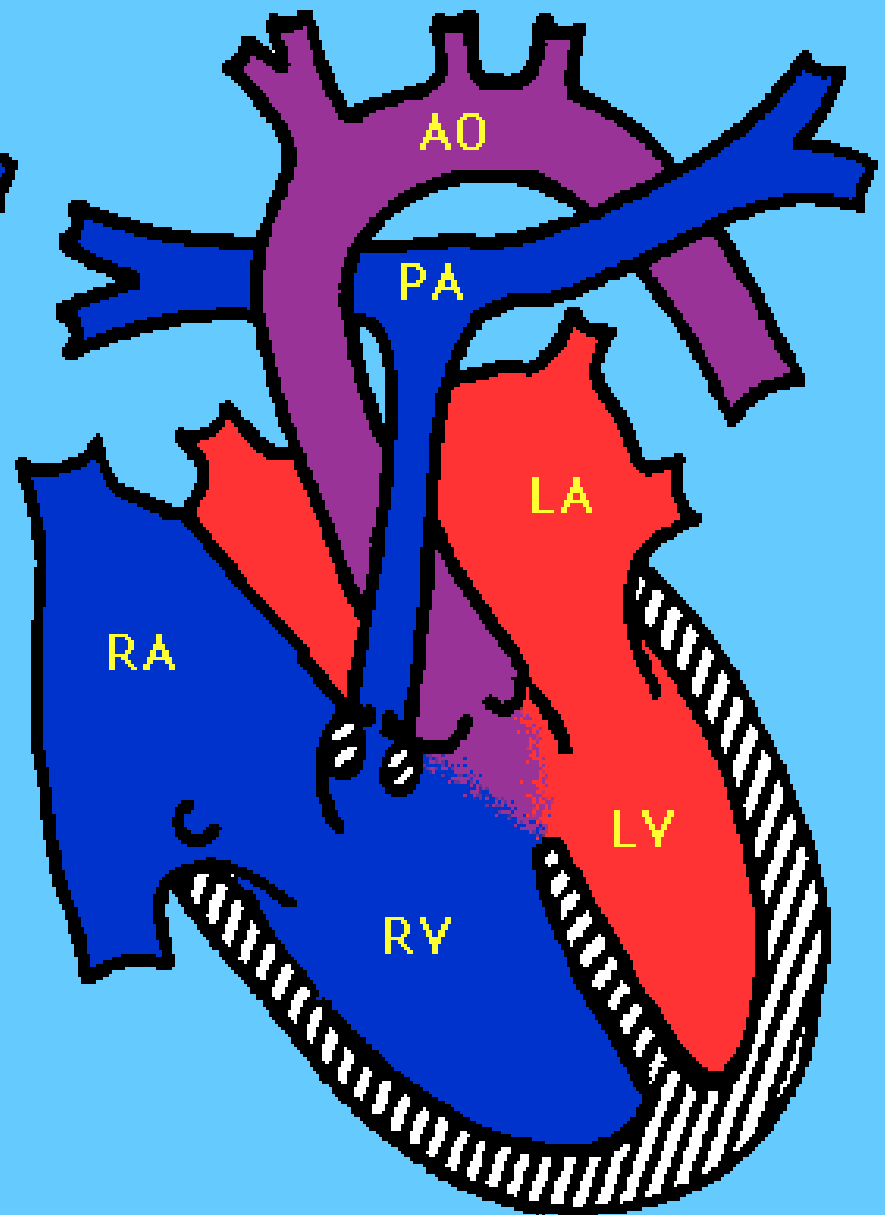


Ventricular Septal Defect

Tetralogy of Fallot

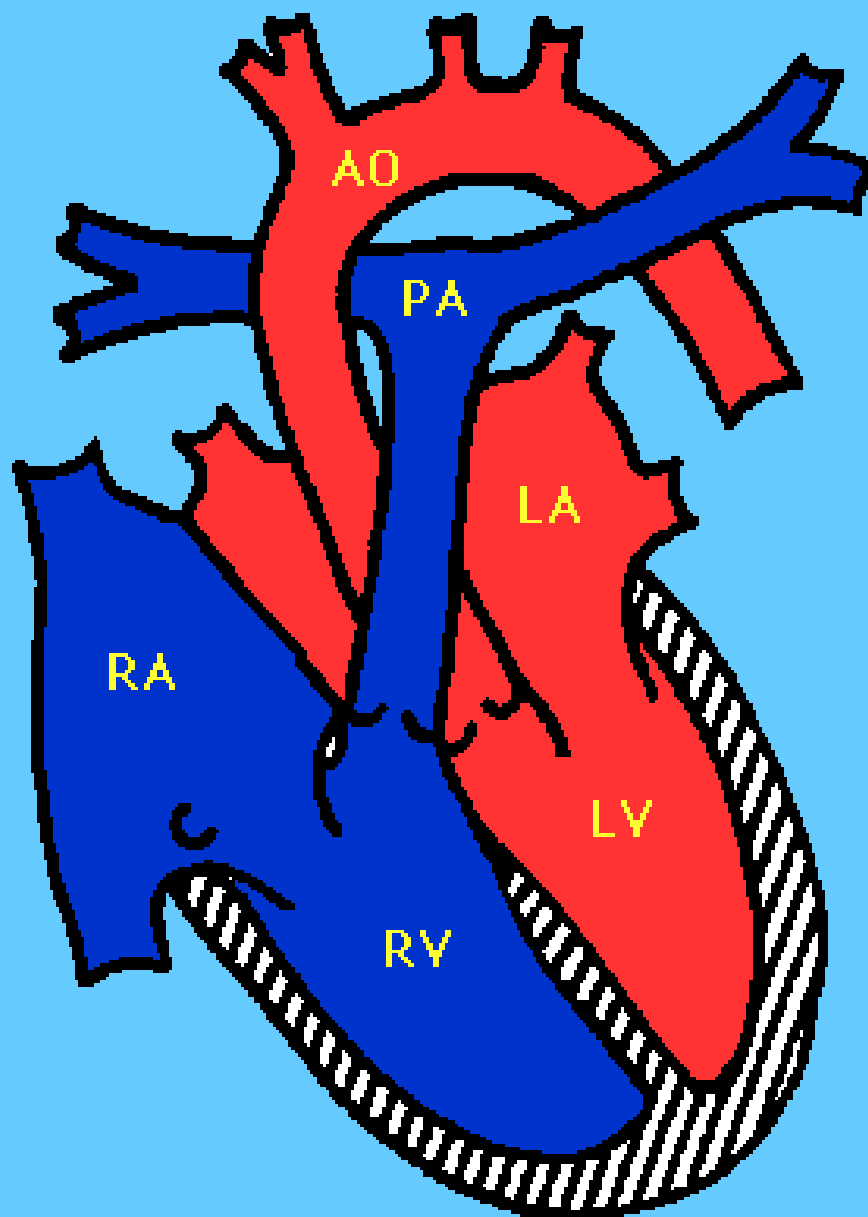


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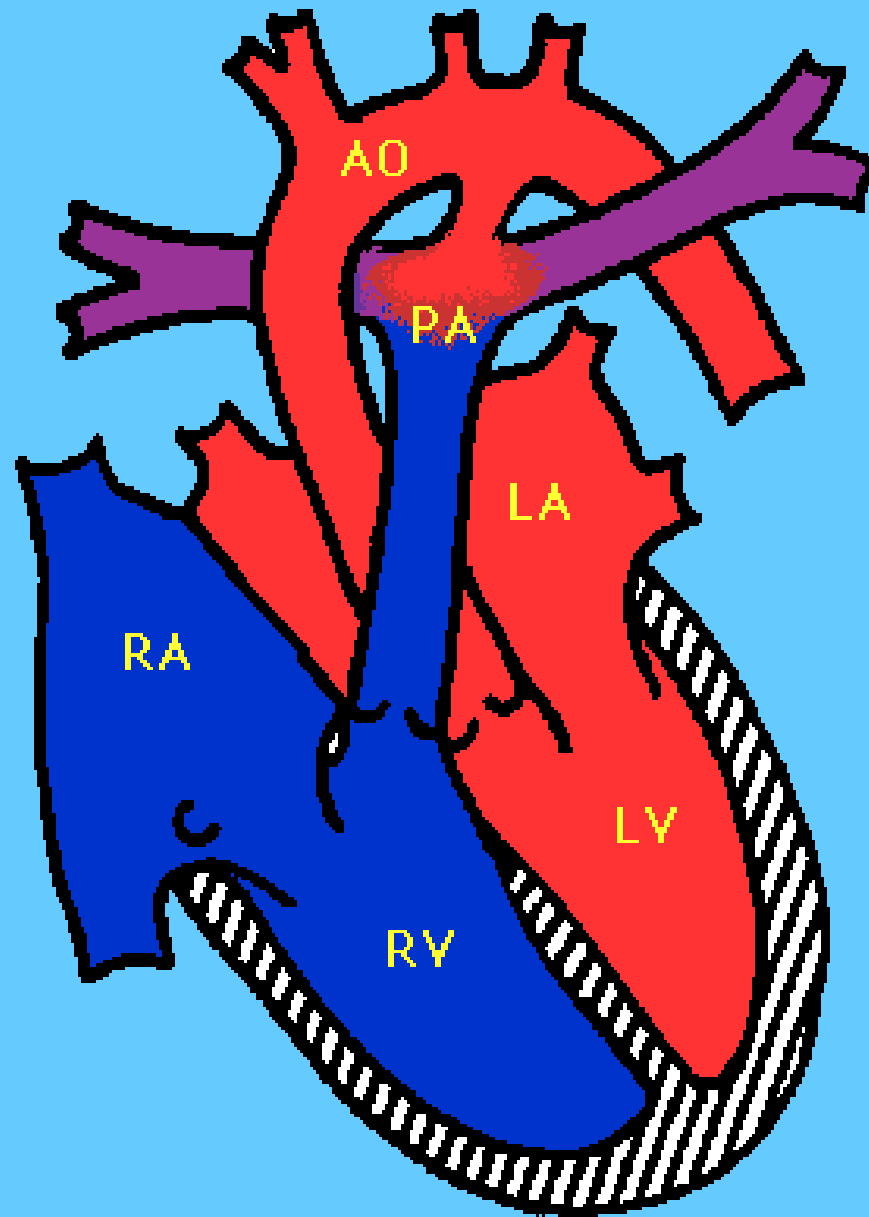


Tetralogy of Fallot

Patent Ductus Arteriosus

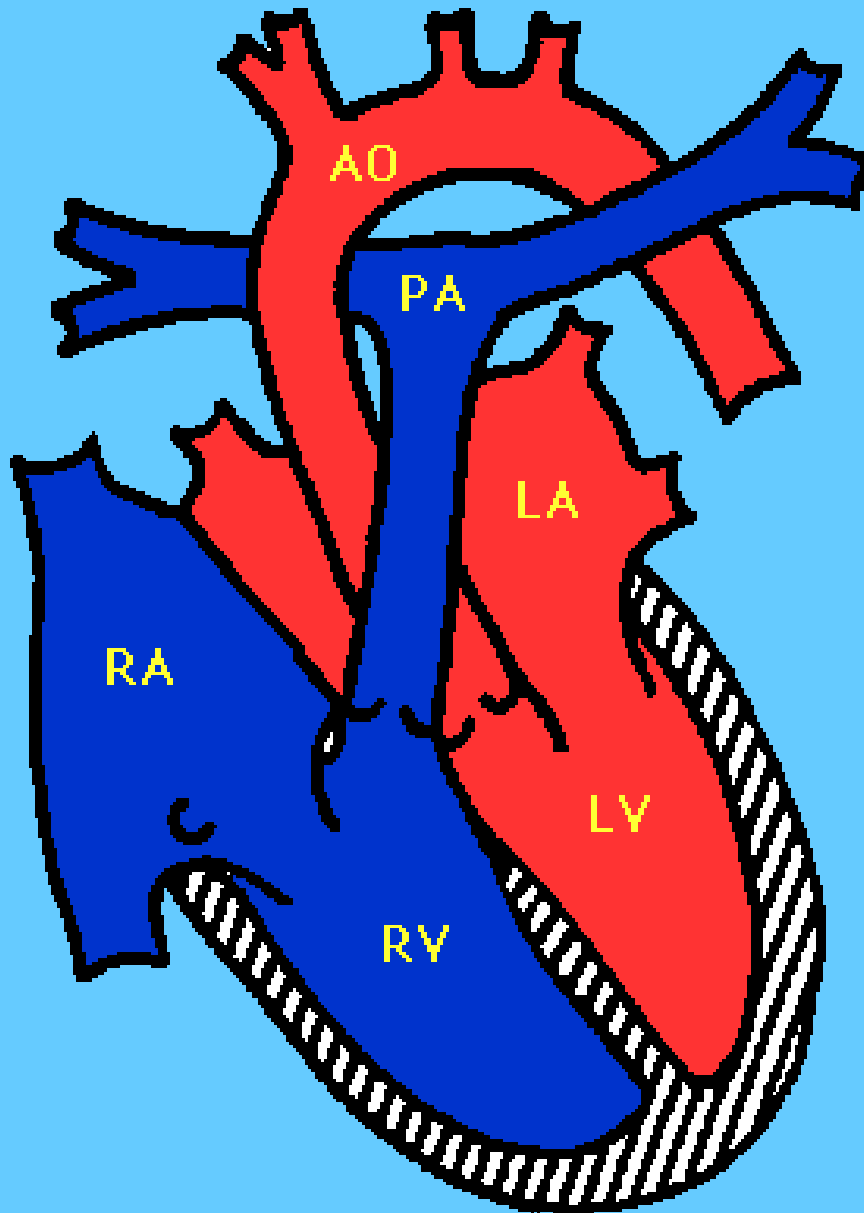


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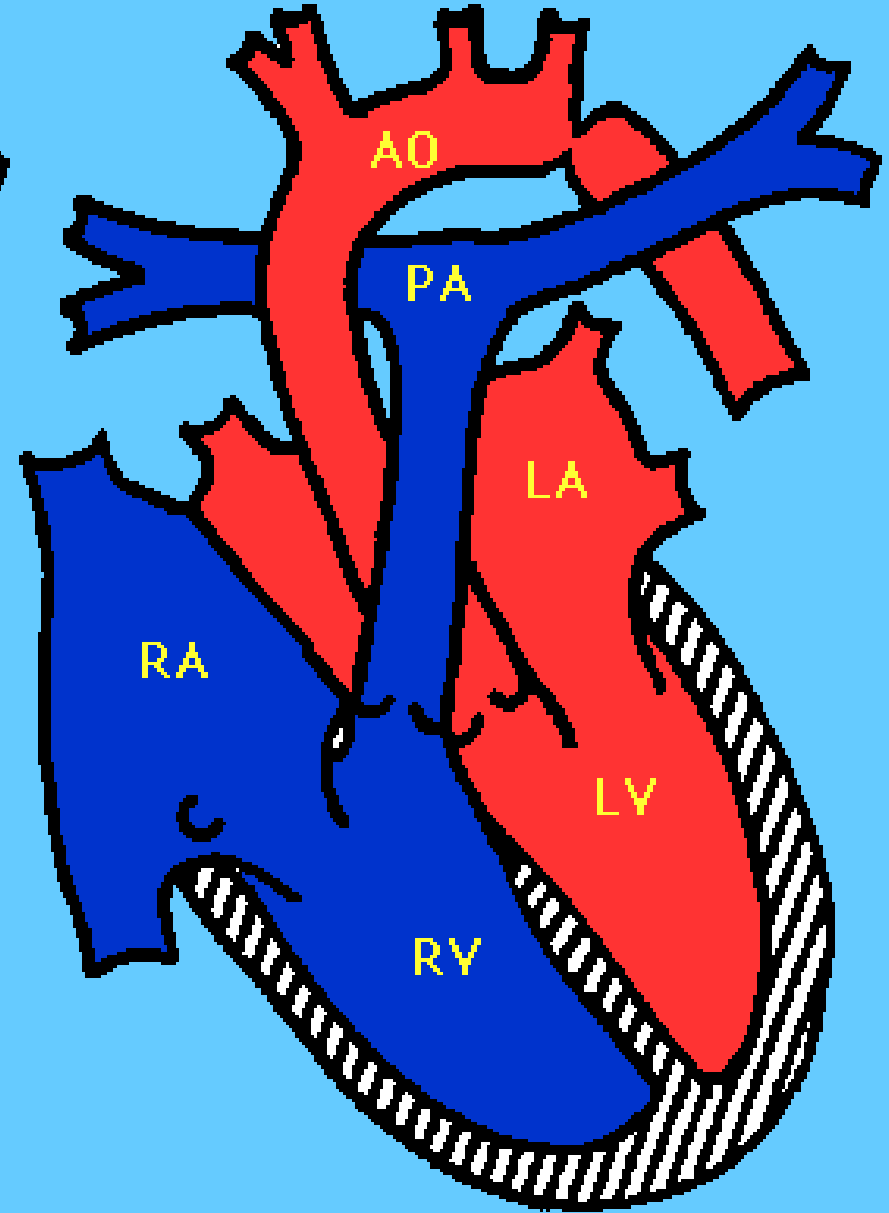


Patent Ductus Arteriosus

Coarctation of the Aorta

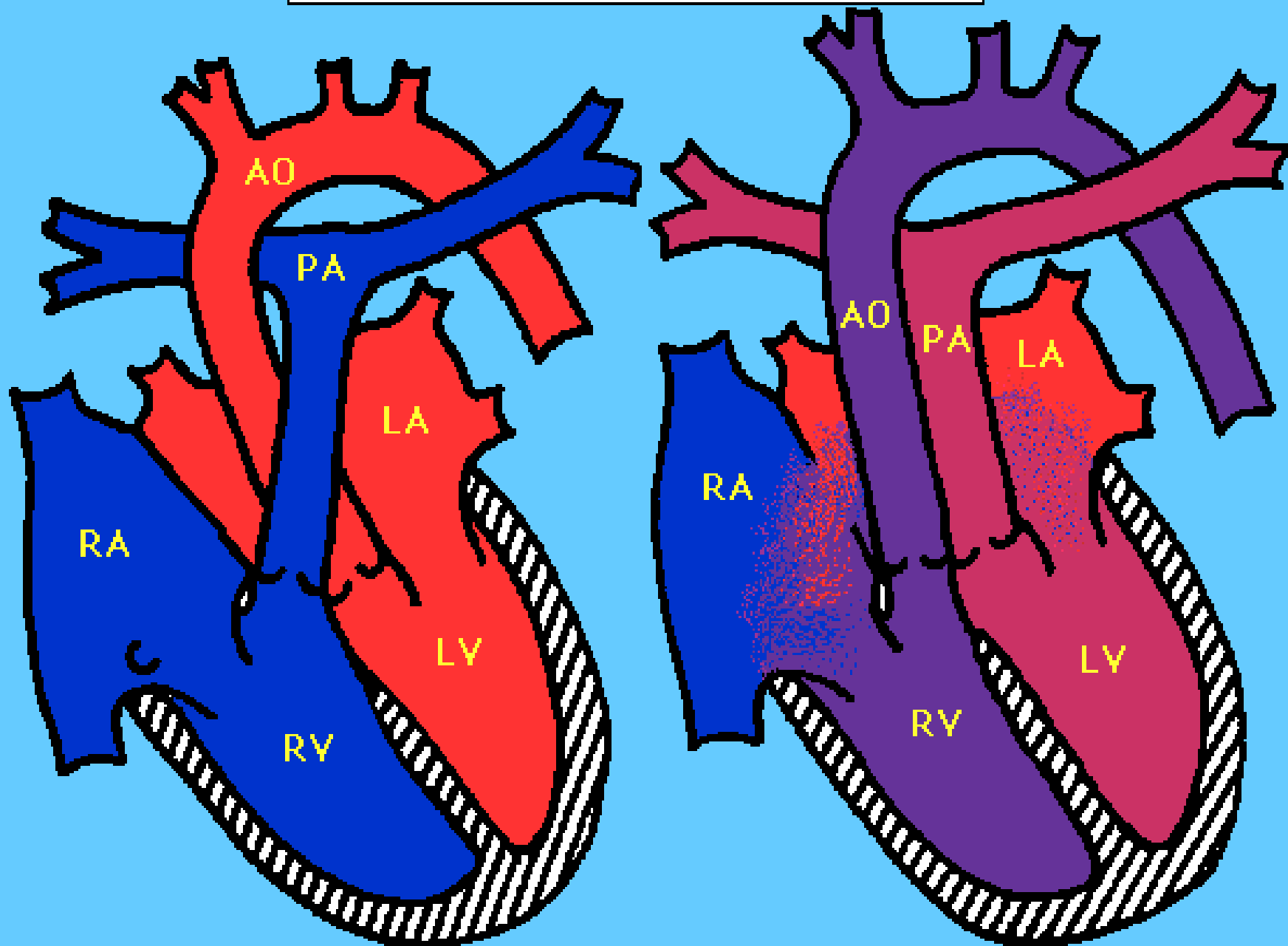


Normal



Coarctation of the Aorta

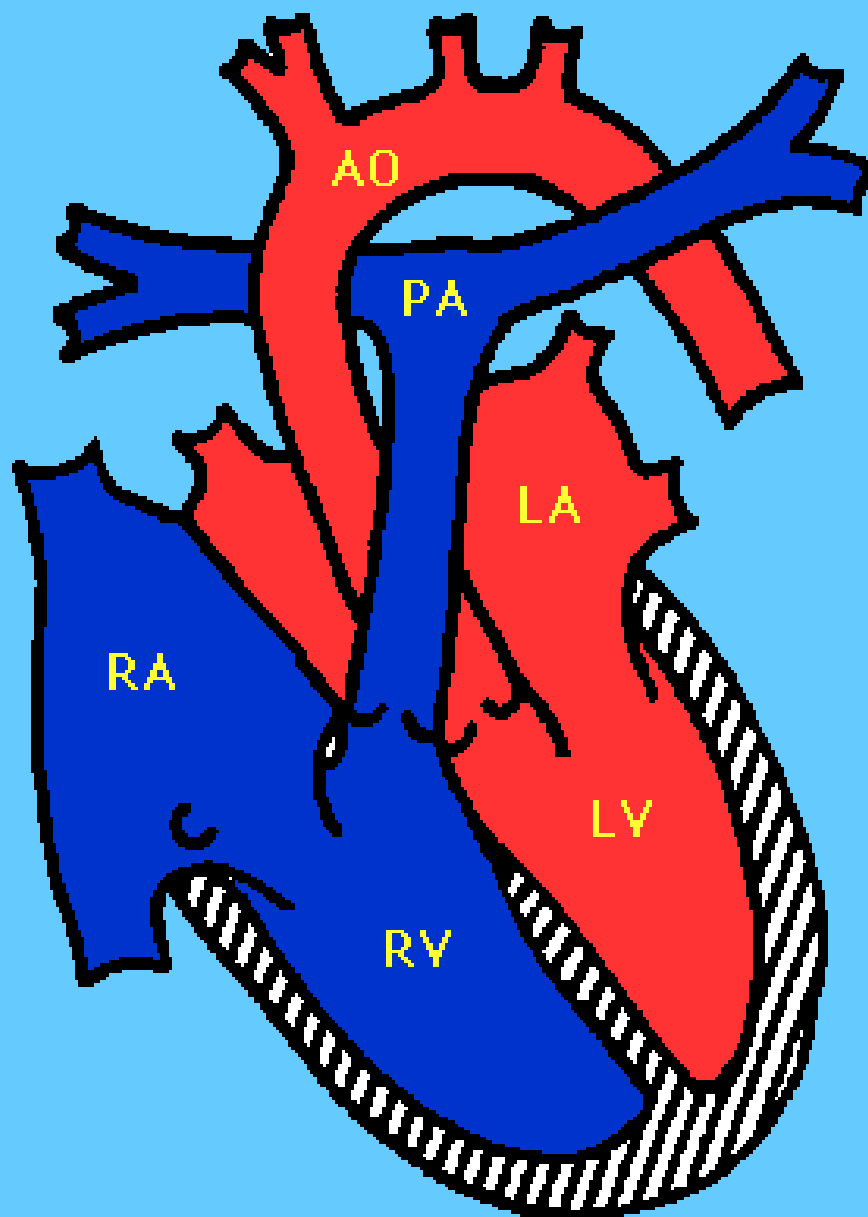
Transposition of the Great Vessels



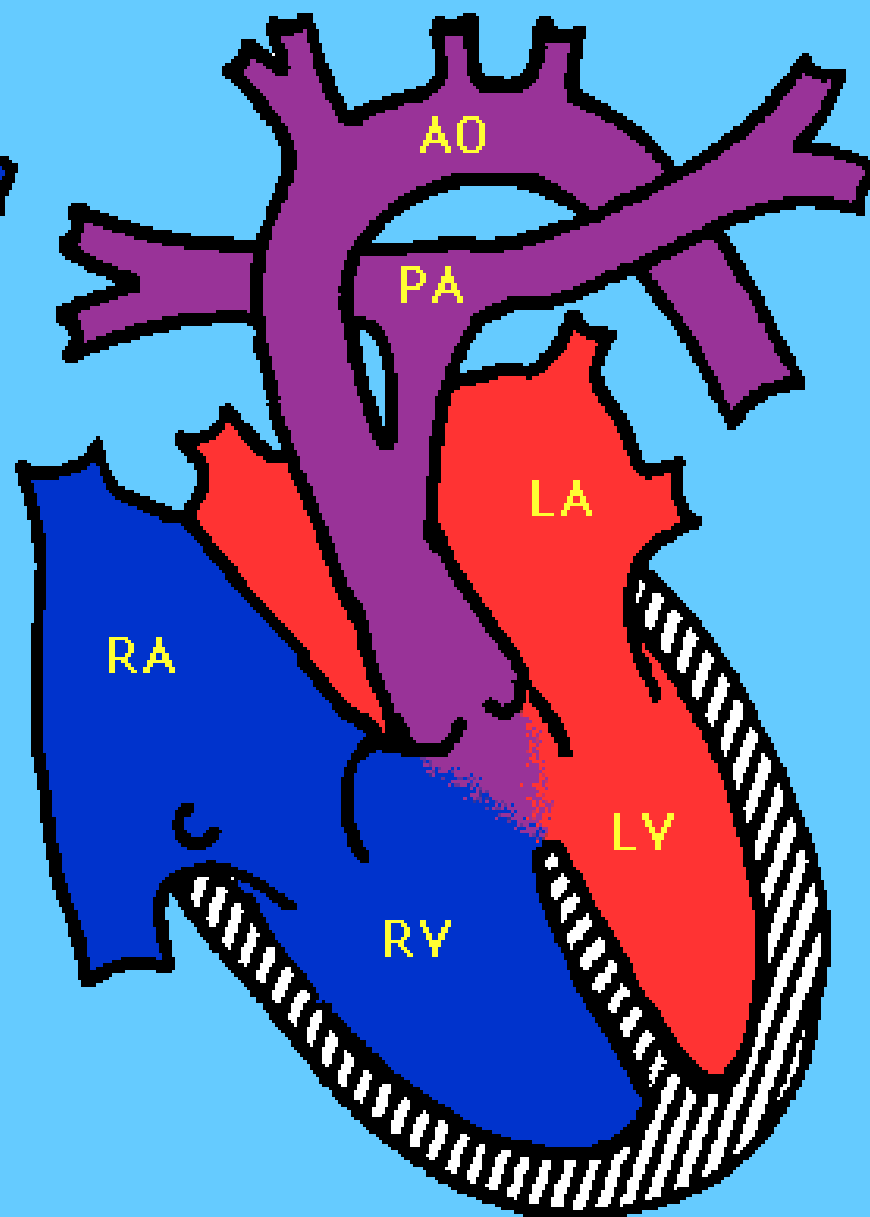
Normal

Transposition of the Great Vessels

Truncus Arteriosus



Normal



Truncus Arteriosus

Pathophysiology

- Heart failure leads to a neurohormonal response→which contributes to the symptoms associated with heart failure and increased morbidity and mortality.

Factors Affecting Cardiac Performance:

- A. Preload
- B. Afterload
- C. Contractility
- D. Heart Rate

After-load vs pre-load pathology

After-load

Obstruction



High Lt ventricle systolic pressure



Increase contractility due to pressure overload.



Prolongation cause diminish cardiac contractility



HF

After-load vs pre-load pathology.....

❖ Preload

- Lt → Rt ventricle
- Rt ventricle → P. artery
- Lungs → Lt atrium
- Lt atrium → Lt ventricle
- Lt ventricle gets high load



- Result in compensation by remodeling (eg. dilatation, hypertrophy).
- Since Lt atrium gets high pressure, result in increase pulmonary venous pressure
- Pulmonary edema

Abnormal Loading Conditions

- **Afterload(Pressure overload)**

1. Aortic Stenosis
2. Pulmonary Stenosis
3. Coarctation of the Aorta

- **Preload(Volume overload)**

1. VSD
2. PDA
3. Valvular Insufficiency

Contractility (force and rate of contraction)

- Is the ability of the heart to change force of contraction dependent on the change of the preload.
- Myocardium abnormalities either congenital or acquired myocarditis, cardiomyopathy, muscular dystrophy result in compromised intrinsic contractility
- **Heart Rate :**
- Tachyarrhythmias shortens the diastolic time interval for ventricular filling.
- It also affect the time for coronary perfusion.

Compensatory Mechanism during low CO:-

1. Frank-Starling Mechanism (Cardiac dilatation)
2. Neurohormonal System Activation
 - **RAAS**
 - **Sympathoadrenal Axis**
3. Myocardium Structural Changes
 - **Eg. Hypertrophy**

Compensatory mechanisms in heart failure:-

1. Cardiac compensation

- Increased HR and cardiac contractility
- Cardiac dilatation (The Frank-Starling mechanism)
- Myocardial hypertrophy

2. Systemic compensation

- Increase the blood volume
- Redistribution of blood flow

- Increment of erythrocytes
- Increased ability of tissues to utilize oxygen

3. neurohormonal compensation

- Sympathetic nervous system
- Renin-angiotensin system
- Atrial natriuretic peptide; endothelin

Classification of heart failure

❖ According to the course of disease

- 1) Acute HF
- 2) Chronic HF

❖ According to the severity

- 1) Mild HF or complete compensation
- 2) Middle HF or incomplete compensation
- 3) Severe HF or decompensation

❖ According to the cardiac output (CO)

- 1) Low-output HF: due to volume overload, pressure overload & contractility problems.
- 2) High-output HF: Heart Rate is primarily affected; 3A (Anemia, arrhythmia, AV Fistula)

❖ **According to the function impaired**

- 1) Systolic failure :Myocarditis, hypertension
- 2) Diastolic failure: restrictive cardiomyopathy, cardiac tamponate.

❖ **According to the location of heart failure**

- 1) Left -side heart failure (LHF)
- 2) Right-side heart failure (RHF)
- 3) Biventricular failure (whole heart failure)

❖ **Left-sided heart failure.**

- Result in reduced left ventricular ~~output~~ → High pressure in left atrium and pulmonary vein → Pulmonary congestion.

❖ **Right-sided heart failure.**

- Result in reduce right ventricular output, for any given right atrium pressure. Eg. Chronic lung disease → Systemic venous congestion

❖ **Biventricular heart failure.**

- Resulted from secondary to the **progression of the disease.**
Eg. Dilated cardiomyopathy, ischemic heart disease, etc.

New York Heart Association Classification

❖ Class I

- ✓ Patients with cardiac disease but without resulting limitation of physical activity. Ordinary physical activity **does not cause** undue fatigue, palpitations, dyspnea.

❖ Class II

- ✓ Patients with cardiac disease resulting in **slight limitation** of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea.

❖ Class III

- ✓ Patients with cardiac disease resulting in **marked limitation** of physical activity. They are comfortable at rest. Less than ordinary activity causes fatigue, palpitation, dyspnea.

❖ Class IV

- ✓ Patients with cardiac disease resulting in **inability to carry on any physical activity without discomfort**. Symptoms of heart failure syndrome may be present even **at rest**. If any physical activity is undertaken, discomfort is increased.

APPROACH A Patient WITH CHF

- ❖ **History: In an infant;** Comprehensive cardiac hx with detail of the **perinatal** period including the presence of **cyanosis, respiratory distress or prematurity.**
- Maternal complication such as gestational DM, medications, SLE or substance that can be associated with cardiac problems.
- Hx of feeding difficulty, volume of food taken, time spent on each breast, dyspnea, diaphoreses while feeding, apnic breathing.
- **In older children;** exercise intolerance, difficulty of keeping up with peers during sport or need for a nap after coming home from school poor growth, orthopnea, PND, hx of chest pain.

GENERAL PHYSICAL EXAMINATION

- Presence of cyanosis, abnormality in growth, any evidence of respiratory distress.
- Presence **of hepatomegaly** and occasionally splenomegally.
- **Cyanosis** – look over nail beds, lips, tongue, mucosa membrane.
- **The site of peripheral edema** - age dependant, in infant – edema is usually seen around the eye, and over the flank, in older children – edema seen on both periorbital, and pedal.
- **Heart rate** : New borns- have **120 to 140 b/min** and was increase to **170/mint** during crying and activity or drop **70 - 90/mint** during sleep.

Clinical manifestation

- ❖ **It depends on the degree of cardiac reserve**
 - Generalized edema and usually involve the eyelid and the sacrum less often the leg and feet.
 - Noisy labored respiration with inter costal and sub costal retraction as well as flaring of nose.
- ❖ **In infant --**
 - Tachypnea
 - Feeding difficulty
 - Poor wt gain
 - Excessive perspiration
 - Irritability
 - Cardeomegaly
 - Gallop rhythm
 - Hepatomegaly

❖ In children --

- fatigue
- Effort intolerance
- Anorexia
- Abdominal pain
- Dyspnea and cough
- Enlarged heart
- Raise JVP
- Hepatomegaly
- Orthopnea
- Anasarca may be present
- Gallop rhythm
- Holocystolic murmur of mitral or tricuspid valve regurgitation may be heard

Investigations

❖ Chest X-ray

- Cardiomegaly
- Pulmonary plethora
(enlargement of blood vessels
in the lungs)
- Pulmonary oedema

❖ ECG

- Shows chamber hypertrophy
- Used to assess the cause of HF
(Not diagnosis)
- Used also to evaluate rhythm
disorder

❖ **Echocardiography- is used**

- To assess ventricular function –parameters :
 - Children – fractional shortening(normal, between 28% and 40%)
 - Adult – ejection fraction(normal between 55%-65%)
- To delineate underlying structural defects.
- To assess cardiac chamber dilatation and hypertrophy.
- To demonstrate decreased myocardial contractility and cardiac function.
- To determine response to therapy.

❖ Doppler Ultrasound

- To estimate CO
- To assess cardiac function, wall motion abnormalities

❖ MRA (Magnetic Resonance Angiography)

- Lt Rt ventricle: function, volume, mass.

❖ Blood investigation:

- CBC , CRP

— Sepsis screen

— Renal function

— Liver function

— Electrolyte

— ABG

— Biomarkers of heart failure (Sérum B-type Natriuretic Peptide (BNP)).

Management of a child with CHF

- **The objective of management include:**
 1. To remove or alleviate the underlying cause of cardiac failure if possible
 2. To improve myocardial contractility
 3. To decrease work load on the heart
 4. To remove retention of sodium and water
 5. To improve tissue oxygenation

❖ **General measures:-**

- Strict bed rest is rarely necessary.
- Patients feel better sleeping in semi-upright position
- for infant with heart failure, an infant chair may be advisable.
- Competitive and strenuous sport activities are usually contraindicated
- For patient with severe pulmonary edema, positive pressure ventilation may be required along with other drug therapy.
- Dietary management

Treatment principles

❖ Remember 5 Ds

1. **Direct** ; Correct the underlying causes of HF
2. **Diet**; (low salt and high calories)
3. **Digitals**; Improve the cardiac contractility
4. **Diuretics**; Reducing preload: frusemide
5. **Dilators**; Reducing afterload; ACE and vasodilators.

1. Diuretics

❖ **furosemide**

- Is the most commonly used diuretic in patients with heart failure.
- It inhibits reabsorption of fluid in the distal tubules and the Loop of Henle.
- An initial IV or IM dose of **1-2 mg/kg** is will be given.
- For chronic **furosemide** therapy give a dose of **1-4mg/kg** and monitor hypokalemia.
- Kcl supplementation is necessary if spiro lactone is not given concomitantly

❖ **Spironolactone** –

- Is an inhibitor of aldosterone and enhance potassium retention.
- If we administer this drug, no need of oral potassium with furosemide.
- The usual dose of spironolactone is 2-3 mg/kg/24hr in two divided dose.

❖ 2. Digitalis

- **Digoxin** is the digitalis glycoside used in pediatric patients.
- Its half-life is 36 hrs which is long enough to allow daily or twice daily administration and short enough to limit toxic effect from over dosage.
- It is well absorbed by GI tract (60-85%) even in infants.
- The usual dose of digoxin is 0.04mg/kg
- Any form of arrhythmia occurring after institution of digitalis therapy must be considered to be drug related until proved.
- A cardiac arrhythmia that develops in a child who is taking digitalis may also be related to the primary cardiac disease rather than the drug.
- succeeding dose should be withheld until the issue is resolved.

3. After load reducing agents and ACE inhibitors:-

- These are groups of drugs that reduce ventricular after load by decreasing peripheral vascular resistance and increase myocardial performance.
- Not recommended in Lt ventricular outlet stenosis.

❖ captopril

- Is an orally active ACE inhibitor that produces arterial dilatation by blocking the production of angiotensin II, thereby resulting in significant after load reduction.

- ❖ Enalapril – is longer acting ACE inhibitor that can be taken once or twice daily

❖ **Nitroprusside** -

- a potent peripheral vasodilator that affects both arterioles and venules
- Monitor BP to prevent sudden hypotension.
- It is contraindicated for those who had hypotension.

Nursing Management

❖ **Maintain the cardiac output**

- Administer digoxin as ordered.
- Take apical pulse and listen to heart sounds regularly, especially before and after each dose of digoxin.
- Use cardiac monitor if ordered.
- Prevent injury by monitoring for digoxin side effects and serum potassium level.
- Provide for rest periods each hour.

Nursing Management



❖ **Maintain adequate oxygenation**

- Place child in semi-Fowler's position.
- Evaluate respiratory rate and sounds.
- Take pulse oximetry readings to determine oxygen saturation.
- Provide oxygen and humidification if ordered.
- Observe for diaphoresis, a sign of increased respiratory effort.

Nursing Management



❖ **Maintain fluid balance**

- Measure intake and output carefully, weigh diapers to obtain output of young child.
- Maintain fluid-restricted diet if ordered.
- Administer diuretics as ordered.
- Monitor electrolytes.
- Weigh daily.
- Measure abdominal girth daily if present.
- Observe for peripheral oedema.

Nursing Management



❖ **Maintain Skin integrity and Nutritional Status**

- Change position frequently
- Hold infant at 45° angle for feeding.
- Record intake carefully.
- Weigh child daily.
- Give frequent small meals with rest periods in-between.
- Give high-calorie snacks.

Nursing Management



❖ **Reduce the anxiety of the family**

with questions.

- Encourage parents to room in or stay with child.
 - Explain procedures and treatment.
 - Involve parents in care as much as possible.
 - Have parents plan child's play periods.
 - At discharge, provide clear instructions and information about what to do in an emergency, and whom and where to call
- Allow parents to verbalize questions, concerns, and feelings.
 - Refer parents to support groups or other resources as needed.

Infective Endocarditis

Mekonen. A

Out lines

- Definition
- Classification
- Etiology
- Epidemiology
- Pathogenesis
- Clinical manifestations
- Diagnosis
- Treatment
- Prognosis
- Complications
- Prevention.

Definition and classification

- Infective endocarditis (IE): Is an infection of the heart's endocardial surface.
- It is a form of endocarditis, or inflammation of the inner tissue of the heart (such as its valves) caused by infectious agents.
- Classified into four groups:
 - Native Valve IE
 - Prosthetic Valve IE
 - Intravenous drug abuse (IVDA) IE
 - Nosocomial IE

Further classification

- **Acute**

- Affects normal heart valves
- Rapidly destructive
- Commonly caused by *S. aureus*
- If not treated, usually fatal within 6 weeks

- **Subacute**

- Often affects damaged heart valves
- If not treated, usually fatal by one year.

Etiologic agents in Pediatric Infective Endocarditis

❖ **For native valve or other**

bovis, S. faecalis)

cardiac lesions:

➤ **un common causes**

➤ **Common causes include:**

include:

– Viridans group streptococci

– Streptococcus pneumoniae

(S. mutans, S. sanguis, S. mitis)

– Haemophilus influenzae

– *Staphylococcus aureus*

– Neisseria gonorrhoeae

– Group D streptococcus (*S.*

– Brucella

Un common causes

- *Chlamydia trachomatis*
- *Chlamydia pneumoniae*
- *Legionella*
- *Bartonella*
- HACEK group(Haemophilus, Actinobacillus,
- Cardiobacterium, Eikenella, and Kingella).
- *Streptobacillus moniliformis*
- *Pasteurella multocida*
- *Campylobacter fetus*

Etiology



Prosthetic valve IE



- *Pseudomonas aeruginosa*
- *Serratia marcescens*
- Diphtheroids
- *Legionella* species
- HACEK group.
- Fungi
- Staphylococcus epidermidis
- Staphylococcus aureus
- Viridans group streptococcus

Epidemiology

- IE is often a complication of congenital or rheumatic heart disease.
- It can also occur in children without any abnormal valves or cardiac malformations.
- It is rare in infancy; if it occurs, it usually follows open heart surgery or is associated with a central venous line.

Epidemiology.....

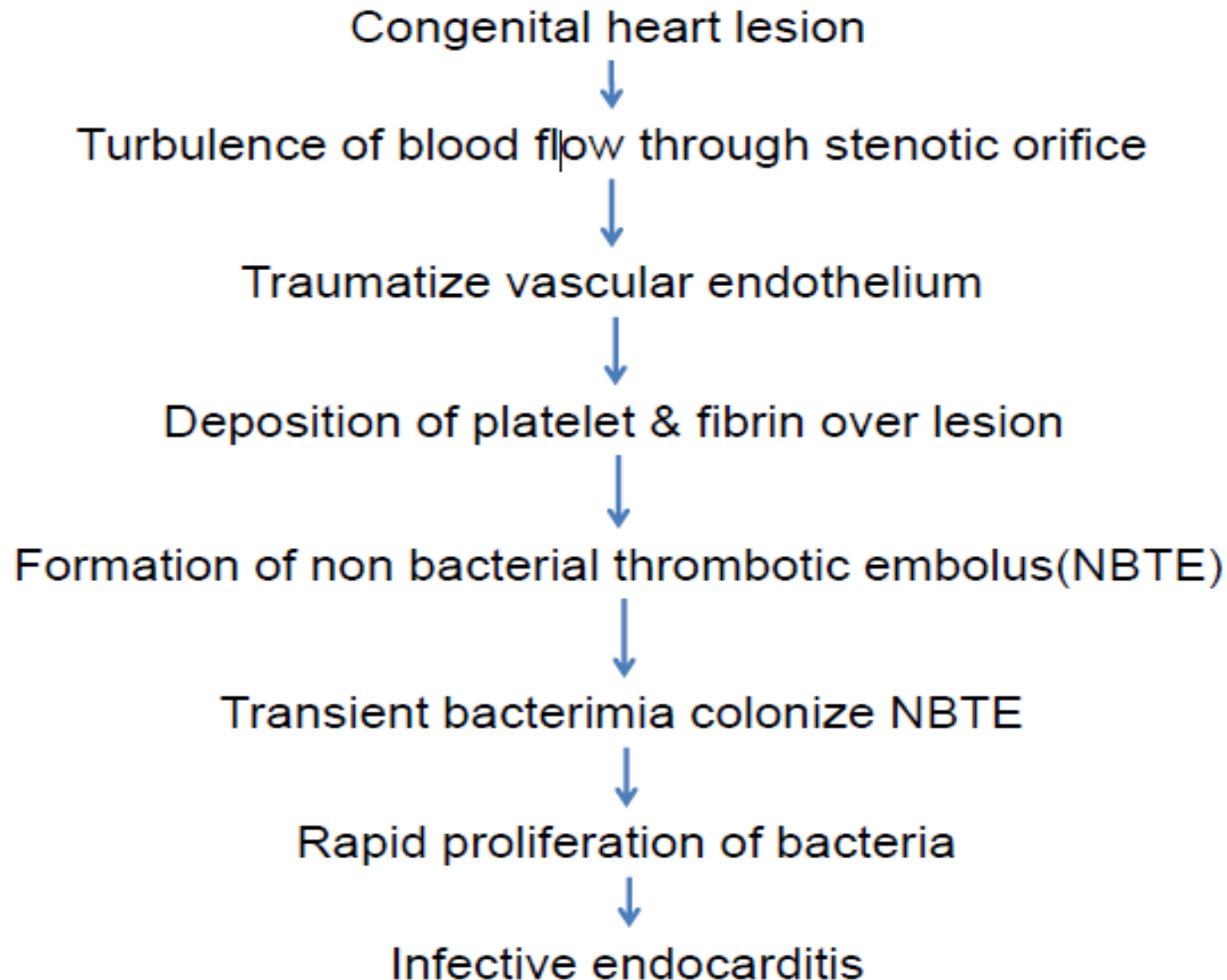
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- In $\approx 30\%$ of patients with infective endocarditis, a predisposing factor is recognized.
- A surgical or dental procedure can be implicated in $\approx 65\%$ of cases in which the potential source of bacteremia is identified.
- Poor dental hygiene in children with cyanotic heart disease results in a greater risk of endocarditis.

Pathophysiology

- **In normal heart,**
 1. Turbulent blood flow disrupts the endocardium making it “sticky”
 2. Bacteremia delivers the organisms to the endocardial surface
 3. Adherence of the organisms to the endocardial surface
 4. Eventual invasion of the valvular leaflets

Pathogenesis in congenital heart lesion



Clinical manifestations

❖ History

- Prior congenital or rheumatic heart disease.
- Preceding dental, urinary tract, or intestinal procedure
- Intravenous drug use
- Central venous catheter
- Prosthetic heart valve

Clinical manifestations.....

❖ Symptoms

- Fever
- Chills
- Chest and abdominal pain
- Arthralgia
- myalgia
- Dyspnea
- Malaise
- Night sweats
- Weight loss
- CNS manifestations (stroke, seizures, headache)

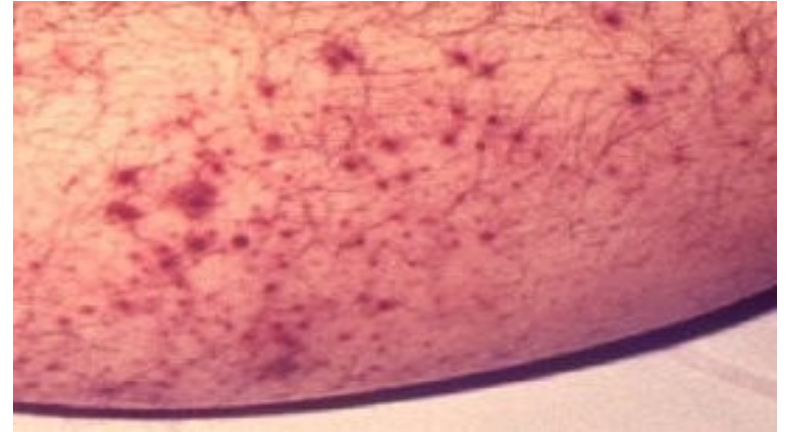
Clinical manifestations.....

❖ Signs

- Elevated temperature
- Tachycardia
- Vascular- Embolic phenomena
(Roth spots, petechiae, splinter nail bed hemorrhages, CNS or ocular lesions)
- Immune complex phenomena
(glomerulonephritis, arthritis, Osler nodes, Roth spot,)
- Janeway lesions
- New or changing murmur
- Splenomegaly
- Arthritis
- Heart failure
- Arrhythmias
- Metastatic infection (arthritis, meningitis, mycotic arterial aneurysm, pericarditis, abscesses, septic pulmonary emboli)
- Clubbing

Petechiae

1. Nonspecific
2. Often located on extremities or mucous membranes



Splinter Hemorrhages



1. Linear reddish-brown lesions found under the nail bed
2. Nonspecific
3. Nonblanching
4. Usually do NOT extend the entire length of the nail

Osler's Nodes



1. Painful and erythematous nodules located on pulp of fingers and toes
2. More specific
3. More common in subacute IE

Janeway Lesions



1. Erythematous, blanching macules located on palms and soles
2. More specific
3. Nonpainful

❖ **Laboratory**

- Positive blood culture
- Elevated ESR; may be low with **heart or renal failure**
- Elevated C-reactive protein
- Anemia
- Leukocytosis
- Hypocomplementemia
- Hematuria
- Renal failure: azotemia (high BUN and creatinine

(glomerulonephritis)

- Chest radiograph: bilateral infiltrates, nodules, pleural effusions
- Echocardiographic evidence of valve vegetations, prosthetic valve dysfunction or leak, myocardial abscess, new-onset valve insufficiency

Diagnostic (Duke) Criteria

1) Definitive infective endocarditis:

- **Pathologic criteria**

- ✓ **Microorganisms or pathologic lesions:** demonstrated by culture or histology in a vegetation, or in a vegetation that has embolized, or in an intracardiac abscess

- **Clinical criteria**

- ✓ Two major criteria, or one major and three minor criteria, or five minor criteria

Diagnostic (Duke) Criteria.....

2) Possible infective endocarditis

- Findings consistent of IE that fall short of “definite”, but not “rejected”

3) Rejected

- Firm alternate Dx for manifestation of IE
- Resolution of manifestations of IE, with antibiotic therapy for ≤ 4 days
- No pathologic evidence of IE at surgery or autopsy, after antibiotic therapy for ≤ 4 days

Diagnostic (Duke) Criteria.....

- **Major criteria**

- Positive blood culture
- Echocardiographic evidence of endocardial involvement

- **Minor criteria**

- Predisposing conditions (heart condition or IV drug use)
- Fever of 100.4°F or higher
- Embolic – vascular phenomena
- Immunologic phenomena
- Microbiologic evidence not meeting major criteria
- Echocardiographic evidence not meeting major criteria

Duke's Major Criteria

1) **Blood Cultures**

- Minimum of three blood samples
- Three separate venipuncture sites
- Obtain 10-20mL in adults and 0.5-5mL in children

➤ **Positive Result**

- Typical organisms present in at least 2 separate samples
- Persistently positive blood culture (atypical organisms)
 - ✓ Two positive blood cultures obtained at least 12 hours apart
 - ✓ Three or a more positive blood cultures in which the first and last samples were collected at least one hour apart

Duke's Major Criteria.....

2) Evidence of endocardial involvement

(Echocardiography)

- Intracardiac mass on a valve or other site
- Regurgitant flow near a prosthesis
- Abscess
- Partial dehiscence of prosthetic valves
- New valve regurgitant flow

Complications

- **Four mechanisms:**
 - Embolic
 - Local spread of infection
 - Metastatic spread of infection
 - Formation of immune complexes –
glomerulonephritis and arthritis

Local spread of infection

1. Heart failure
 - Extensive valvular damage
 - Myocardial abscesses
 - Toxic myocarditis
2. Paravalvular abscess (30-40%)
 - Most common in aortic valve, and *S. aureus* infection
 - Higher rates of embolization and mortality
3. Arrhythmias
4. Heart block
5. Mycotic aneurysms
6. Rupture of sinus of Valsalva(aortic sinus)

Metastatic spread of infection

- Metastatic abscess
 - Kidneys, spleen, brain, soft tissues
- Meningitis and/or encephalitis
- Vertebral osteomyelitis
- Septic arthritis

Treatment

1. Medical
2. Surgical

Medical treatment

- Pre-antibiotic era - a death sentence
- Antibiotic era
 - microbiologic cure in majority of patients
- Parenteral antibiotics
 - High serum concentrations are needed to penetrate vegetations
 - Prolonged treatment is important to kill dormant bacteria clustered in vegetations

Medical treatment.....

- A total of 4–6 wk of treatment is recommended, with serumcidal levels.
- Depending on the clinical and laboratory responses, antibiotic therapy may require modification and, in some instances, more prolonged treatment is required.
- In nonstaphylococcal disease, bacteremia usually resolves in 24–48 hr, whereas fever resolves in 5–6 days with appropriate antibiotic therapy.
- Resolution with staphylococcal disease takes longer

Medical treatment: bacterial

- ❖ Commonly used antibiotic combination:

Aqueous crystalline penicillin G sodium

or

Ceftriaxone sodium

plus

Gentamicin sulfate

Plus

Vancomycin hydrochloride

Medical treatment:

❖ Fungal

- **Amphotericin B**
- **5-fluorocytosine**
- Treatment of complications
 - Heart failure
 - ✓ Digitalis
 - ✓ Salt restriction
 - ✓ Diuretic therapy

Surgical treatment

- Removal of vegetations
- Valve replacement

Indication of surgical treatment:

- Severe aortic or mitral valve involvement with intractable heart failure
- Mycotic aneurysm (infected aneurysm)
- Rupture of an aortic sinus
- dehiscence of an intracardiac patch
- failure to sterilize the blood despite adequate antibiotic level
- recurrent emboli
- new heart block
- myocardial abscess
- increasing size of vegetation

PROGNOSIS

- In the pre-antibiotic era, infective endocarditis was a fatal disease.
- Despite the use of antibiotic agents, mortality remains at 20–25%.
- Serious morbidity occurs in 50–60% of children with documented infective endocarditis

Poor prognostic factors

- Femaleness
- S. aureus
- Vegetation size
- Aortic valve
- Prosthetic valve
- Older age
- Diabetes mellitus
- Low serum albumen
- Heart failure
- Paravalvular abscess
- Embolic events

PREVENTION.

- In patients with high or moderate risk heart conditions, antimicrobial prophylaxis before various procedures:
 - dental and oral procedures
 - surgery of the upper respiratory tract
 - Surgery of the GI tract
 - Urinary tract procedures
- Continuing education regarding the important of prophylaxis
- Proper general dental care and oral hygiene

PREVENTION:

- **Antibiotic prophylaxis**

- Standard general prophylaxis;
 - amoxicillin
- If unable to take oral medication
 - ampicillin
- If allergic to penicillin;
 - clindamycin

- cephalexin

- azithromycin

- clarithromycin

- If allergic to penicillin and unable to take oral medications;
 - clindamycin
 - cefazolin

ACUTE RHEUMATIC FEVER

Mekonen A.

Introduction

- Rheumatic fever is an **immunologically mediated inflammatory disorder**, which occurs as a sequelae to group A streptococcal pharyngeal infection.
- It is a multisystem disease affecting the heart, joints, brain, cutaneous and subcutaneous tissues.
- Acute rheumatic fever remains an important preventable cause of cardiac disease.

- **Etiology:**

- Rheumatogenic strains of GAS serotypes (M- 1, 3, 5, 6, 18, 24)
 - $\frac{2}{3}$ of the patients with an acute episode of rheumatic fever have a history of an upper respiratory tract infection several weeks before.

Epidemiology

- **Most common** form of acquired **heart disease** in all age groups world wide
- Important cause of chronic heart disease and death in developing world
- An estimated 30 million people suffer from ongoing heart disease from ARF, 70% of them die at average age of 35 years old.
- Accounts for
 - 50% of all cardiovascular disease
 - 50% of all cardiac admissions in many developing countries
- Males and females are equally affected.

Factors associated with acute rheumatic fever

– Socioeconomic status

- ✓ Overcrowding,
- ✓ Poverty,
- ✓ Lack of access to medical care

– Virulence of strain of GAS

- ✓ Serotypes of GAS (M types 1, 3, 5, 6, 18, 24) are associated with ARF

– Host factors

- ✓ Age : Peak incidence in children 5 - 15 years old
- ✓ Previous history of acute rheumatic fever

Pathogenesis:

- The pathogenesis is still not clearly defined.
- Group A strep pharyngeal infection precedes clinical manifestations of ARF by 2 - 6 weeks

❖ **Two seriously considered theories regarding pathogenesis:**

1. The cytotoxicity theory

- Suggests that cytotoxic effect of **GAS streptolysin O toxin** may be responsible for the pathogenesis.

2. The immunologic theory (theory of molecular mimicry)- more acceptable now a days.

- Antibodies made against group A strep cross-react with human tissue (e.g. heart, brain, joint).

The immunologic theory.....

- There is no direct invasion to the tissue by the microorganism but its an **autoimmune** disease that involves Ag-Ab interaction.
- It is due to an immunologic reaction that is a delayed sequelae of GAS infections of the pharynx (not skin infection).

The immunologic theory.....

❖ **Group A streptococcal(GAS) pharyngeal infection**

- Body produce **antibodies** against streptococci ->
- These antibodies **cross react with human tissues** because of the **antigenic similarity** between streptococcal components and human connective tissues (molecular mimicry)[there is certain amino acid sequence that is similar between GAS and human tissue]->
- **Immunologically mediated inflammation & damage**
(autoimmune) to human tissues which have antigenic similarity with streptococcal components- like heart, joint, brain connective tissues.

Clinical features

- Following upper airway infection with GAS
 - Silent period of 2 - 6 weeks
 - Sudden onset of fever, pallor, malaise, fatigue
 - RF is characterized by:
 - Arthritis
 - Carditis
 - Sydenham's chorea
 - Erythema marginatum
 - Subcutaneous nodules
- Called "major Jones's criteria"
- 



Subcutaneous nodules



*Erythematous patches
with central clearing*

Erythema marginatum

❖ **Minor Jone's criteria**

- Clinical features

✓ Fever

✓ Arthralgia

- Laboratory features

✓ Elevated c-reactive protein or

✓ Erythrocyte sedimentation rate

✓ Prolonged PR interval on EKG

- **Migratory polyarthrititis (in about 75%):**

- Most common major manifestation but least specific.
- Almost always asymmetrical and migratory.
- Commonly involves larger joints (knees, ankles, elbows, wrists).
- No permanent joint deformity.
- If untreated – lasts 2 to 3 weeks.
- Dramatic response to salicylates – a hallmark of this symptom.

- **Rheumatic Carditis:**

- Characterized by mitral regurgitation – is the hallmark of rheumatic carditis
- Is the most serious manifestation
- occurs in about 50–60% of all cases of acute rheumatic fever
- May lead to death in acute phase or at later stage
- Any cardiac tissue may be affected
- Endocarditis (valvulitis), is a universal finding.
- Most common valvular lesions are : mitral and aortic lesions.
- Serious and long-term illness is related entirely to **valvular heart disease**

❖ Clinical signs of carditis include:

- Tachycardia
- Murmurs
- Cardiomegaly
- Rhythm disturbances (prolonged PR interval)
- Pericardial friction rubs
- Cardiac failure
- peripheral and pulmonary edema

- **Chorea (involvement of Basal ganglia & caudate nucleus)**
 - Seen in about 20% of patients with RF.
 - Delayed manifestation – usually 3mo or longer.
 - Characterized by purposeless and involuntary movements, muscle incoordination, weakness and emotional lability.
 - May disappear with sleep.
 - Seen in < 5% of cases.
 - Erythematous, macular nonpruritic rash with pale centers and rounded or serpinginous margins.
 - Mostly trunk and proximal extremities.
 - May be induced by application of heat.

- **Erythema marginatum:**

- **Subcutaneous nodules**

- Seen in less than 3% of patients with RF.
- Is a firm, painless, freely movable nodules (0.5 – 2cm in size).
- Most often seen in patients with carditis.
- Usually located over the extensor surfaces of the joints (elbows, knees and wrists), in the occipital portion of the scalp, or over the spinous processes.

Diagnosis

❖ Jones criteria

- Criteria developed to prevent overdiagnosis even though some criticism regarding validity
- Still important as guidelines

❖ The probability of ARF is high with:

- Evidence of previous infection with streptococcal upper respiratory infection plus

- 2 major criteria

or

- 1 major criteria and 2 minor criteria

- ❖ Supporting evidence for antecedent streptococcal pharyngitis:
 - **Positive throat culture or rapid streptococcal antigen test.**
 - **Elevated or rising streptococcal antibody titer.**
- There are **3 circumstances** in which the diagnosis of acute rheumatic fever can be made without strict adherence to the Jones criteria.
 - Chorea.
 - Indolent carditis (carditis of insidious onset and slow progression)
 - Recurrences of acute rheumatic fever.

Treatment

❖ Supportive

- Bed rest .
- For carditis with heart failure
 - ✓ Digoxin,
 - ✓ Fluid and salt restriction,
 - ✓ Diuretics,
 - ✓ Oxygen

❖ Antibiotic therapy

- 10 days of orally administered penicillin or erythromycin,
or
- Single intramuscular injection of benzathine penicillin

Anti-inflammatory therapy

- Agents such as acetaminophen can be used to control pain and fever while the patient is being observed for more definite signs of acute rheumatic fever or for evidence of another disease.
- ASA in patients with typical migratory polyarthrititis and those with carditis.
- The usual dose of aspirin is 100 mg/kg/day in 4 divided doses PO for 3–5 days, followed by 75mg/kg/day in 4 divided doses PO for 4 wk.

- ❖ In patients with carditis and cardiomegaly or congestive heart failure, corticosteroids will be given.
 - The usual dose of prednisone is 2 mg/kg/day in 4 divided doses for 2–3 wk followed by a tapering of the dose that reduces the dose by 5 mg/24 hr every 2–3 days.
 - At the beginning of the tapering of the prednisone dose, aspirin should be started at 75 mg/kg/day in 4 divided doses for 6 wk.

Prognosis

- The prognosis for patients with acute rheumatic fever depends on:
 - ✓ The clinical manifestations present at the time of the initial episode,
 - ✓ The severity of the initial episode
 - ✓ The presence of recurrences.
- Approximately 70% of the patients with carditis during the initial episode of acute rheumatic fever recover with no residual heart disease;
- The more severe the initial cardiac involvement, the greater the risk for residual heart disease.

- Patients who have had acute rheumatic fever are susceptible to recurrent attacks following reinfection of the upper respiratory tract with GAS.
- Therefore, these patients require long-term continuous chemoprophylaxis
- Approximately 20% of patients who present with “pure” chorea who are not given secondary prophylaxis develop rheumatic heart disease within 20 yrs.
- Therefore, patients with chorea, even in the absence of other manifestations of rheumatic fever, require long-term antibiotic prophylaxis.

Prevention

❖ Primary Prevention

- Appropriate antibiotic therapy

OR

instituted before the 9th day of
symptoms of acute GAS
pharyngitis

- Penicillin V 250 mg, twice a day
Oral

OR

❖ Secondary Prevention

- Penicillin-G benzathine , every 4 wk
IM

- < 27 kg: 600,000 units

- >27 kg: 1,200,000 units

- Sulfadiazine or sulfisoxazole

- For people who are allergic to
penicillin and sulfonamide drugs:

- Erythromycin 250 mg, PO, BID

Duration of prophylaxis

- Patients with carditis with their initial episode of acute rheumatic fever should receive antibiotic prophylaxis well into adulthood and perhaps for life.
- Patients who did not have carditis : until they reach their early 20s and after at least 5 yr have elapsed since their last episode of acute rheumatic fever.
- The decision to discontinue prophylactic antibiotics depends on epidemiologic factors such as the risk for exposure to GAS infections.

any questions?

"A sore throat
can lead to a
broken heart"

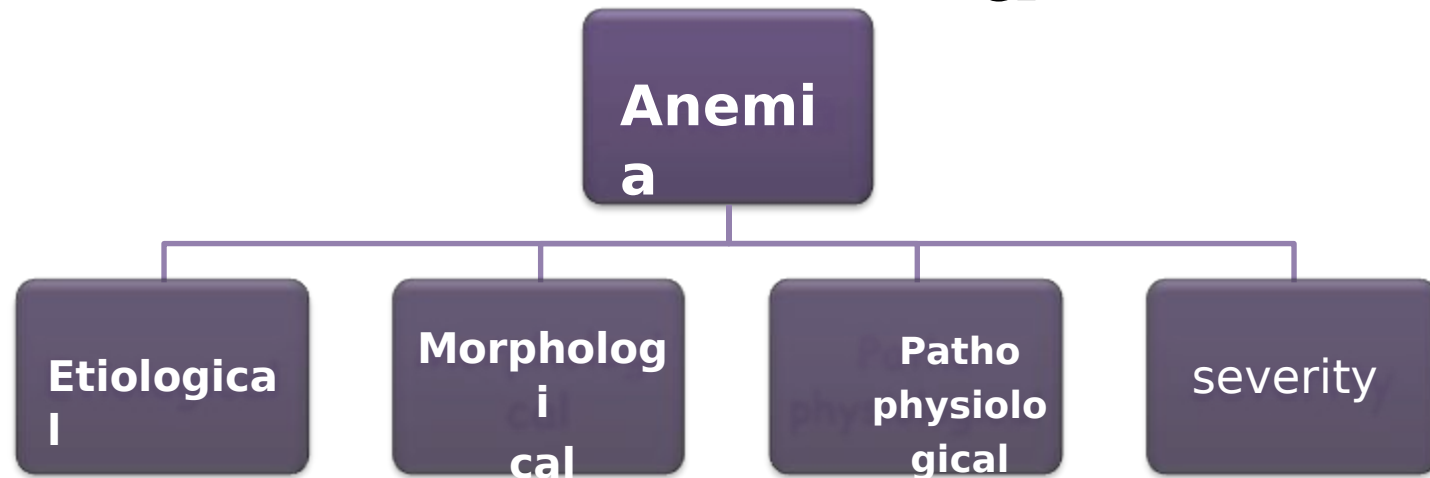


Anemia

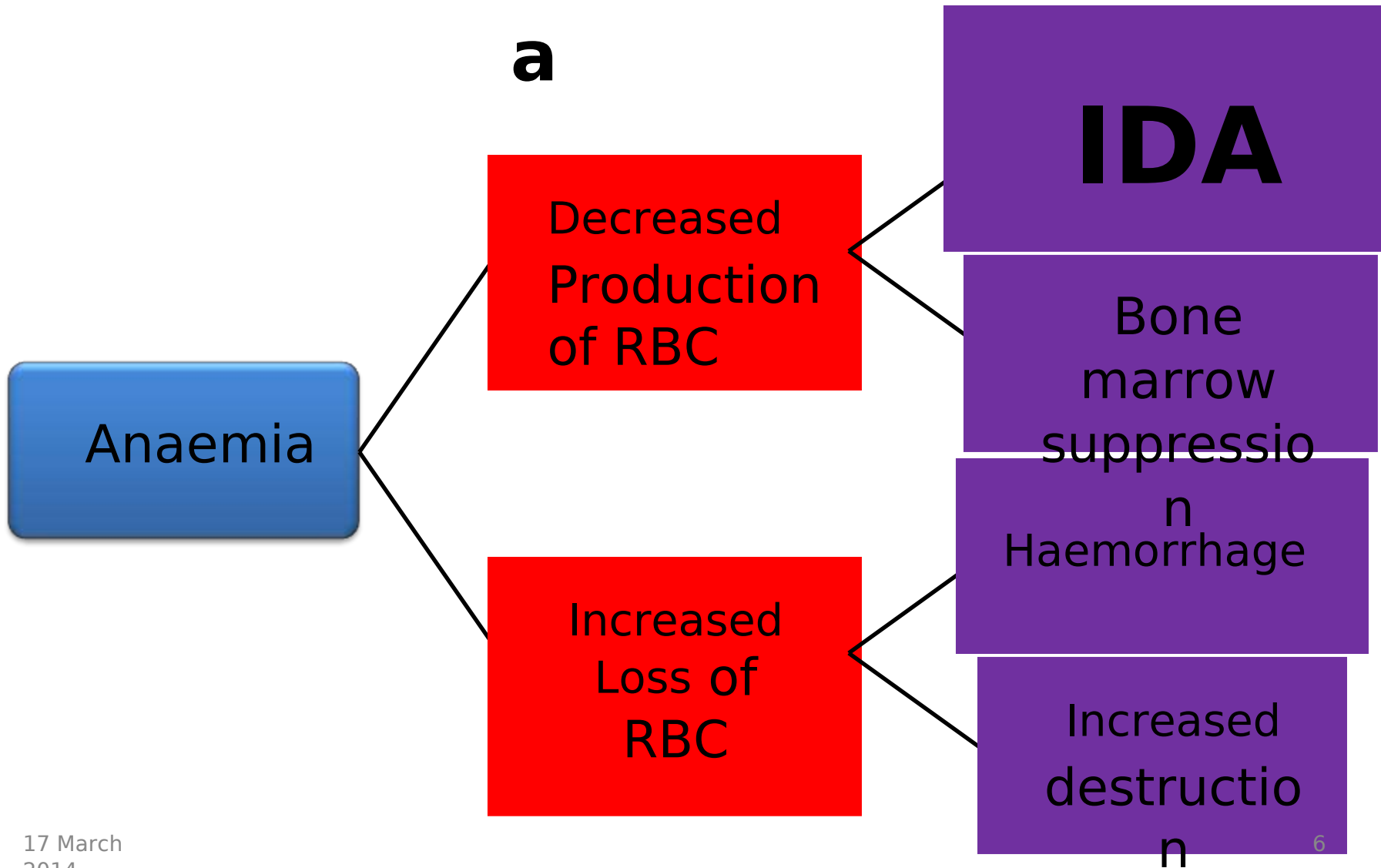
Anemia

- Anemia is a Greek word
- “Ana” = absent/decreased
- „emia“ = blood
- Anemia refers to a lower than normal number of red blood cells in the blood.
- The principal function of red blood cells is to carry oxygen from the lungs and distribute it throughout the entire body. In general, anemia is caused by one of the following:
 - An inability of the body to make red blood cells
 - Premature destruction of the red blood cells
 - Bleeding, or loss of red blood cells

classification of Anaemia



Etiological classification of Anaemia



A) Etiological classification(Classification according to cause)

1. Anemia due to blood loss:

- a) Acute post hemorrhagic: It occurs due to any accident, which cause large amount of blood loss. Anemia is normocytic normochromic anemia
- b) Chronic post hemorrhagic: When small amount of blood is lost continuously from our body E.g. in Hookworm infestation, chronic duodenal ulcer, bleed piles. The anemia is initially normochromic normocytic but later changes to Hypochromic microcytic anemia.

Etiological classification.....

2. Anemia due to impaired red cell formation:

function not due to deficiency of factor required for erythropoiesis

a) Due to disturbance of bone marrow function or due to deficiency of factors necessary for erythropoiesis

I. Iron deficiency anemia

II. Megaloblastic anemia

1. Anemia associated with chronic infection like renal failure, liver disease, disseminated malignancy

2. Bone marrow infiltration

3. Aplastic anemia

4. Anemia associated with myxedema,

b) Due to disturbance of bone marrow Hypopituitarism

Etiological classification.....

3. Anemia caused by excessive red cell destruction (Hemolytic anemias):
 - I. Intracorpuseular causes
 - Decreased production of Hb (Thalassemia)
 - Abnormal production of Hb (hemoglobinopathies) – sickle cell anemia.
 - II. Extracorpuseular causes
 - III. Mechanical damage and antibodies to RBCs etc.

Morphological classification

I

Size & color of RBCs

```
graph TD; A[Size & color of RBCs] --> B[Normocytic normochromic]; A --> C[Microcytic hypochromic]; A --> D[Macrocytic anemia];
```

**Normocytic
normochromic**

**Microcytic
hypochromic**

**Macrocytic
anemia**

B).Morphological classification:

- Classification based on characteristics of red cell as determined by blood examination (MCV, MCH, MCHC)
 1. Normocytic normochromic anemia: Here MCV, MCH, MCHC are normal.
 - E.g. aplastic anemia, acute post hemorrhagic anemia.
 2. Microcytic hypochromic anemia: here, MCV: Decreased; MCHC: Decreased and MCH: Decreased **E.g. iron deficiency anemia**
 3. Macrocytic anemia: MCV: Raised. E.g. megaloblastic anemia

Patho-Physiological

```
graph TD; A[Patho-Physiological] --> B[Increased demand]; A --> C[Decreased production]; A --> D[Increased loss];
```

Increased
demand

Decreased
productio

Increased
loss

C. Pathophysiological classification(how anaemia occurs)

- I. Increased demand. eg. Infancy, childhood, reproductive age and pregnancy in female.
- II. Decrease production.
- III. Increased loss

Severity
WHO

```
graph TD; A[Severity WHO] --> B[Moderate 7-10.9]; A --> C[Severe 4-6.9]; A --> D[Very severe <4 g/dl];
```

A hierarchical flowchart showing the WHO classification of anemia severity. The root node is a red box labeled 'Severity WHO'. It branches into three green boxes: 'Moderate 7-10.9', 'Severe 4-6.9', and 'Very severe <4 g/dl'.

Moderate
7-10.9

Severe
4-6.9

Very severe
<4 g/dl

Common form of anaemia or types in children

1. Iron deficiency anaemia
2. Aplastic anaemia
3. Sickle cell anaemia
4. B- Thalassemia

IRON DEFICIENCY ANEMIA

- A common form of anaemia in childhood is due to **iron deficiency.**

❖ Causes

- Inadequate iron intake in the diet.
- Chronic diseases
- Iron malabsorption ,as in chronic diarrhoea and malabsorption syndromes, such as celiac disease and pernicious anaemia
- Blood loss secondary to drug- induced GI bleeding

Complication of IDA

- Infection and pneumonia
- Pica, causing the child to eat lead based paint, which result in poisoning
- Bleeding
- Hemochromatosis(iron overload)

Clinical manifestation OF IDA

- When a child has a mild anaemia, he or she may appear almost normal;
- Signs of anaemia may include:
 - paleness, headache or dizziness, fatigue
 - In cases of sudden very severe anemia, a child may become listless, extremely pale and may feel faint,
 - Dyspnoea,
 - Inability to concentrate
 - Tachycardia
 - Susceptibility to infection

Investigation

- Hematocrit (HCT) and Hb and serum ferritin - levels are low
- Mean corpuscular Hb level is decreased
- RBC count is low, with microcytic and hypochromic cells
- Serum iron level- are low with high binding capacity

WHO's Hb thresholds used to define anemia

Age or gender group	Hb threshold (g/dl)	Hb threshold (mmol/l)
Children (0.5–5.0 yrs)	11.0	6.8
Children (5–12 yrs)	11.5	7.1
Teens (12–15 yrs)	12.0	7.4
Women, non-pregnant (>15yrs)	12.0	7.4
Women, pregnant	11.0	6.8
Men (>15yrs)	13.0	8.1

Management of IDA

- The treatment of anemia in general depends on the cause of the anemia.
- In general, iron deficiency anemia can be treated successfully with prescription-strength iron by mouth and with changes in diet by increase iron intake by adding rich in iron to diet .
- Drug thereby
 - Oral preparation of iron or a culmination of iron and ascorbic acid (which enhance iron absorption)
 - Vitamin supplement : cyanocobalamin (vitamin B 12) if intrinsic factor is lacking
 - Iron supplements : iron dextran
 - In some cases, blood transfusions must be given.

APLASTIC ANEMIA

- **Definition**
- Group of disorders characterized by pancytopenia (anemia, granulocytopenia, and thrombocytopenia) resulting from the decreased functional capacity of a hypoplastic, fatty bone marrow.
- **Causes**
- **Congenital:** Congenital hypoplastic anemia (Blackfan – Diamond anemia); develops between ages 2 and 3 months
- **Acquired :** Drug – induced; occurs as an adverse effect from chemotherapy, chloramphenicol ,phenytoin (Dilantin), some antibiotics, and some recreational drugs

Clinical manifestations of AA

- Anorexia
- Dyspnea, tachypnea
- Epistaxis
- Fatigue, weakness
- Gingivitis
- Headache
- Melena
- Multiple infections, fever
- Palpitations, tachycardia
- Purpura, petechiae, ecchymosis, pallor

Investigation

- Bone marrow biopsy shows fatty marrow with reduction of stem cells
- Faecal occult blood test is positive
- Haematology shows decreased granulocytes, thrombocytes, and RBCs
- Peripheral blood smear shows pancytopenia
- Urine chemistry reveals haematuria

Medical management

- Dietary changes, including establishing a high- protein, high-calorie, high-vitamin diet
- Transfusion of platelets and packed RBCs
- **Drug therapy:**
 - Analgesics: ibuprofen, acetaminophen; androgens: fluoxymensterone, oxymetholone; Antibiotics (if infection present); Hematopoietic growth factor : epoetin alfa; Human granulocyte colony – stimulating factor: filgrastim
 - Neulasta (a man-made form of a protein that stimulates the growth of white blood cells in our body)
 - Bone marrow transplantation
 - Peripheral stem – cell transplants

• **Nursing interventions**

- Assess respiratory status to detect hypoxemia caused by low Hb levels
- Assess vital signs for signs of hemorrhage, infection, and activity intolerance
- Assess cardiovascular status to detect arrhythmias or myocardial ischemia
- Monitor and record intake and output and urine specific gravity to determine fluid balance

Nursing care con.....

- Assess stool, urine, and emesis for occult blood loss caused by decreased platelet levels
- Monitor for infection, bleeding , and bruising caused by decreased levels of WBCs and platelets
- Encourage fluids and administer I.V. fluids to replace fluids lost by fever and bleeding
- Administer oxygen to improve tissue oxygen because low Hb levels reduce the blood's oxygen-carrying capacity
- Administer transfusion therapy, as prescribed, to replace low levels of blood components
- Diet

SICKLE CELL ANEMIA

- It is a congenital haemolytic disease resulting from characterized by the partial or complete replacement of normal haemoglobin with abnormal haemoglobin S (HbS)
- Sickle cell anemia occurs primarily in blacks people.

➤ **Causes**

- Autosomal recessive inheritance (homozygous inheritance of the Hb S-producing gene.

Complication of SCA

- Retinopathy , nephropathy, and cerebral vessel occlusion caused by organ infarction
- Hypovolemic shock and death
- Necrosis
- Infection and gangrene

Clinical Manifestation of SCA

❖ Infants:

- Affected children are usually asymptomatic until 4 to 6 months of age because sickling is inhibited by high levels of fetal haemoglobin.
- Colic from pain caused by an abdominal infarction
- Dactylitis or hand foot syndrome from infarction of the small bones of the hand and feet
- Splenomegaly

Clinical manifestation of SCA.....

❖ Toddlers and pre-schoolers

- Hypervolemia and shock from sequestration of large amount of blood in spleen
- Pain at site of vaso- occlusive crisis

❖ School age children and adolescents

- Delayed growth and development , delayed sexual maturity
- Enuresis
- Extreme pain at site of crisis

Clinical manifestation of SCA.....

- Suspect any of the following crisis in a patient with sickle cell anemia - who has:
 - Pale lips, tongue , palms or nail beds
 - Lethargy
 - Restlessness
 - Sleepiness
 - Irritability
 - severe pain and fever.

Management

- Bed rest with crises
- Avoidance of stress and extreme temperatures
- Hydration with I.V. fluid (may be increased to 3 L/day during crisis)
- Short-term oxygen therapy (long-term oxygen decreases bone marrow activity, further aggravating anemia)
- Transfusion therapy as needed
- Drug therapy
 - Analgesic: meperidine
 - Antineoplastic : hydroxyurea
 - Vaccine: pneumococcal vaccine

Leukemia

LEUKEMIA

- Leukaemia is the abnormal, uncontrolled proliferation of white blood cells (WBCs)
- Acute lymphocytic leukaemia (ALL) is the most common type of leukaemia and cancer in children
 - Peak age is 2 to 5 years
 - 90% to 95% of children with ALL achieve a first remission
 - Almost 80% live 5years
 - The child between ages 3 and 7 with ALL and an initial WBC count of less than 10,000/ μ l at the time of diagnosis has the best prognosis

- Acute myeloblastic (myeloblastic) leukaemia (AML) is more common than ALL in adolescents
 - 50% to 70 % of adolescents with AML achieve a first remission
 - 40% live 5 years
- Monozygotic twin with leukaemia
- Congenital disorders, such as Down syndrome, Bloom syndrome, and ataxia-telangiectasia
- Viruses
- Ionizing radiation
- Exposure to the chemical benzene and cytotoxins such as alkylating agents

❖ **Causes**

- The exact cause of most leukemias remains unknown
- Increasing evidence suggests a combination of contributing factors.

❖ **Predisposing factors**

- Familial tendency

Pathology

- Acute leukemia:- is characterized by clonal expansion of immature hematopoietic or lymphoid precursors.
- Chronic leukemia – refers to conditions characterized by the expansion of mature marrow elements

Clinical features

- **General Systemic Effects**

- Fever (60%).
- Lassitude (50%)
- Pallor (40%)

- **Hematologic Effects Arising from Bone Marrow Invasion**

- Anaemia- pallor, fatigability, tachycardia, dyspnoea & CHF
- Neutropenia – fever, ulceration of buccal mucosa and infection.
- Thrombocytopenia – petechial, purpura, easy bruisability, bleeding from mucous membrane and internal bleeding.¹⁹⁸

Clinical features.....

- **Clinical Manifestations Arising from Lymphoid System Infiltration**
 - Lymphadenopathy
 - Splenomegaly.
 - Hepatomegaly
- **Clinical Manifestations of Extramedullary Invasion**
 - CNS- Seizures
 - Genitourinary - painless testicular swelling
 - Bone, joints- bone pain
 - Skin - bleeds

Investigation

- Bone marrow aspirate showing a proliferation of immature WBCs (greater than 25% blasts)
- Bone marrow biopsy of the posterior superior iliac spine can help confirm diagnosis
- Blood counts show thrombocytopenia, neutropenia, and anemia
- Differential leukocyte count determines cell type.
- Lumbar puncture detects meningeal involvement (if central nervous system (CNS)is involved)

Medical management

- Systemic chemotherapy aims to eradicate leukemic cells and induce remission
- ALL – combination therapy that may include vincristine, prednisone, high-dose cytarabine
- Intrathecal chemotherapy to prevent or treat CNS infiltration by leukemic cells
- Antibiotic, antifungal, and antiviral drugs and granulocyte injections to control infection
- Transfusions of platelets to prevent bleeding and of red blood cells to prevent anemia
- Possible bone marrow transplant especially in children with the Philadelphia chromosome(a specific genetic abnormality in *chromosome* 22 of leukemia cancer cells particularly chronic myeloid leukemia)

Complications

- Organ malfunction, especially the spleen and liver
- Anemia
- Infection
- Bleeding (petechiae)
- Death

Nursing interventions

- Prevent infection by placing the patient in a private room, giving special attention to mouth care, and screening staff and visitors for contagious diseases
- Inspect the skin frequently; avoid taking a rectal temperature or administering rectal medications
- Use interventions for anemia and thrombocytopenia
- Give increased fluids to flush to flush chemotherapy through the kidneys
- Provide a high-protein, high-calorie , bland diet

Nursing

care.....

- Provide pain relief
- Monitor the CNS for involvement (confusion, lethargy, and headache)
- Gear nursing measures toward easing the adverse effects of radiation and chemotherapy
- Help the child and the family verbalize their fears and guilt; allow the family to participate in the child's care, as appropriate
- Help the child adjust to changes in body image
- Refer parents and adolescents with cancer to support groups

Hemophilia

What is Hemophilia?

- Hemophilia is an X-linked recessive disorder characterized by deficiency of coagulation factor.
- It is a common **hereditary coagulation blood disorder** due to deficiency or reduced activity of commonly **clotting factor VIII** or **clotting factor IX**.
- Hemophilia is a bleeding disorder that slows down the blood clotting process.
- It's **transmitted via females** to **males**. Female who carry a single mutated gene, are generally asymptomatic and not affected.
- Hemophilia is more common in males than females.

Types of Hemophilia

Disease	Factor deficiency	Inheritance
Hemophilia A	VIII (8)	X linked recessive
Hemophilia B	IX (9)	X linked recessive
Hemophilia C	XI (11)	Autosomal recessive
Parahemophilia	V (5)	Autosomal recessive

Hemophilia A

- Also known as classic hemophilia or Factor **VIII Deficiency**.
- People with this type of hemophilia have **low levels of a blood clotting factor called factor 8 (FVIII)**. It's the **most common** type of hemophilia.
- **Mild Hemophilia A**: Do **NOT** have spontaneous bleeding but unusual bleeding occurs with surgery and tooth extractions.
 - People are usually diagnosed with this in later life.
- **Moderate Hemophilia A**: spontaneous bleeding, delayed oozing after minor injury, and usually diagnosed before they are 5 to 6 years old.
- **Severe Hemophilia A**: Spontaneous joint or deep muscle bleeding. Usually diagnosed within first two years of life.

Hemophilia B

- Also known as **Christmas disease** or Factor IX Deficiency
- People with this type of hemophilia have low levels of a blood clotting factor called factor 9 (FIX).
- The two different types of hemophilia are caused by permanent gene changes (mutations).
- Mutations in the factor VIII gene cause Hemophilia A. Mutations in the factor IX gene cause Hemophilia B.
- **Hemophilia C.** Some doctors use this term to refer to a lack of clotting factor **XI**.

What causes hemophilia in children?

- Hemophilia types A and B are inherited diseases. They are passed on from parents to children through a gene on the X chromosome.
- Females have two X chromosomes, while males have one X and one Y chromosome.
- A female carrier has the hemophilia gene on one of her X chromosomes.
- When a hemophilia carrier female is pregnant, there is a 50/50 chance that the hemophilia gene will be passed on to the baby.
- If the gene is passed on to a son, he will have the disease.
- If the gene is passed on to a daughter, she will be a carrier.
- If the father has hemophilia but the mother does not carry the hemophilia gene, then none of the sons will have hemophilia disease, but all of the daughters will be carriers.

- In about one-third of the children with hemophilia, there is no family history of the disorder. In these cases, it's believed that the disorder could be related to a new gene flaw.
- Carriers of the hemophilia gene often have normal levels of clotting factors but may:
 - Bruise easily
 - Bleed more with surgeries and dental work
 - Have frequent nosebleeds
 - Have heavy menstrual bleeding
- Hemophilia C usually doesn't cause problems, but people may have increased bleeding after surgery.

- Woman can have hemophilia in the ff occasion:
 - Lyonization(inactivation) of the normal X chromosome
 - Turner syndrome (XO)
 - Father with hemophilia/ mom as a carrier

What are the symptoms of hemophilia?

- The most common symptom of this disorder is heavy, uncontrollable bleeding.
- The severity of haemophilia depends on the amount of clotting factors in the blood.
- Those affected with haemophilia that have levels greater than 5% most often have bleeding only with major surgeries or tooth extractions.
- These children may not even be diagnosed until bleeding complications from a surgery occur.
- Severe haemophilia occurs when the factor VIII or IX is less than 1%. Bleeding can occur in these children, even with the minimal activities of daily life.
- Bleeding may also occur from unknown injury.
- Bleeding most often occurs in the joints and in the head.

❖ **Patient's symptoms may also include:**

- **Bruising.** Bruises can occur from even small accidents.
- **Bleeds easily.** A tendency to bleed from the nose, mouth, and gums with minor injury.
- **Bleeding into a joint.** Hemarthrosis (bleeding into a joint) can cause pain, immobility, and deformity if not treated. This is the **most common site of complications** due to hemophilia bleeding.
- **Bleeding into the muscles.** Bleeding into the muscles can cause swelling, pain, and redness. This can cause permanent damage and deformity.
- **Bleeding in the brain from injury or spontaneously.** Bleeding in the brain, is the most common cause of death in children with hemophilia and the most serious bleeding complication.
- Small bleeds in the brain can result in blindness, intellectual disability, a variety of neurological deficits. It can lead to death if not spotted and treated right away.

Diagnosis?

- The diagnosis of haemophilia is based on patient`s family history, patient`s medical history, and a physical exam.
- ❖ Blood tests include:
 - Complete blood count (CBC).
 - Clotting factors. To check the levels of each clotting factor.
 - Bleeding times. To test the speed that blood clots.
 - Genetic or DNA testing. To check for abnormal genes.

treatment

- Treatment for hemophilia is aimed at preventing bleeding complications (mainly head and joint bleeds).
- Patient's healthcare provider will refer to a hematologist, an expert in blood disorders.
- Patient's health care provider will figure out the best treatment based on:
 - How old patient(child) is.
 - His or her overall health and medical history
 - How sick he or she is
 - How well patient(child) can handle specific medications, procedures, or therapies
 - How long the condition is expected to last
- Treatment depends on the type and severity of the hemophilia.

- Treatment may include:

- Bleeding in the joint may need surgery or immobilization.

Patient may need rehab of the affected joint. This may include physical therapy and exercise to strengthen the muscles around the area.

- Blood transfusions may be needed if major blood loss has occurred. This is when patient gets donated blood.
 - Self-infused factor VIII or IX can allow a child with hemophilia to lead a near normal lifestyle.

Treatment.....

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❖ Factor VIII concentrates:

- Hemofil M(Baxter)
- Immunate(Baxter)
- Koate-DVI(Bayer)

❖ Factor IX concentrates:

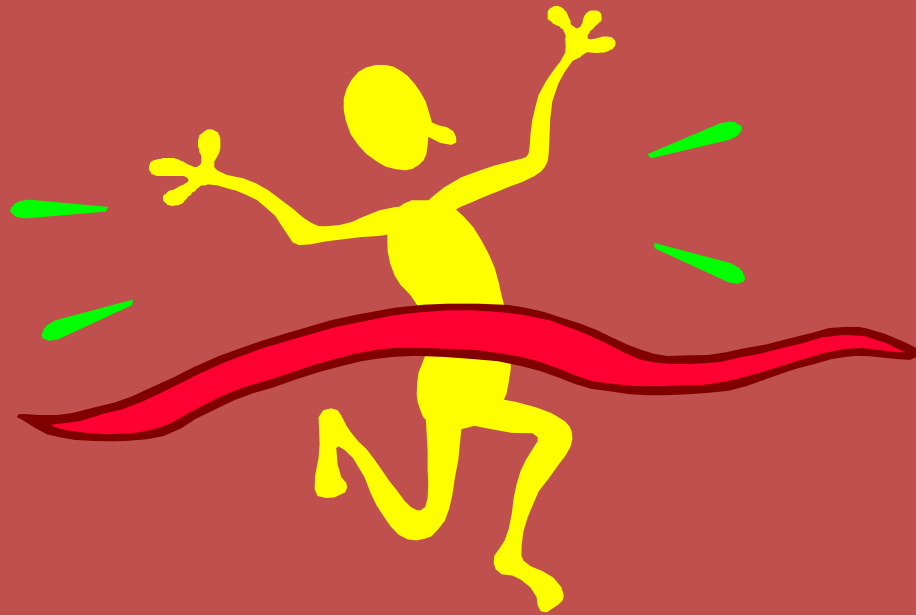
- Aimafix(Kedrion)
- Immunine(Baxter)

Complications

❖ **Complications of hemophilia include:**

- Bleeding in the joints or muscles
- Inflammation of the joint lining
- Long-term joint problems
- Very serious tumor-like enlargements, of the muscle and bone
- Development of antibodies against clotting factors
- Infections from transfusions

Thank you



you

Thank