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Pediatrics

CARDIOLOGY

FIRST EDITION
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FIRST EDITION
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TABLE OF CONTENTS

“Two men looked out from prison bars, one saw the mud, the other saw stars.”

Which one will you be?

INTRODUCTION TO CONGENITAL HEART DISEASE (CHD)

It is the most common single group of malformations in infants.

Classification

According to presence/absence of cyanosis:

Acyanotic

- **Shunts (potentially cyanotic)**: ASD, VSD, PDA, and AVSD
- **Semilunar valve lesions**: PS, AS
- **Obstructive heart lesions**: Coarctation of the aorta
- **Atrioventricular valve lesions**: Congenital MR, MS (rare)

Cyanotic

- **With ↑ pulmonary flow (Plethora)**
  - Transposition of Great Vessels
  - Truncus Arteriosus
- **With ↓ pulmonary flow (Oligemia)**
  - Tetralogy of Fallot
  - Tricuspid Atresia

Others

- Cardiac malposition
- Coronary abnormalities

Incidence

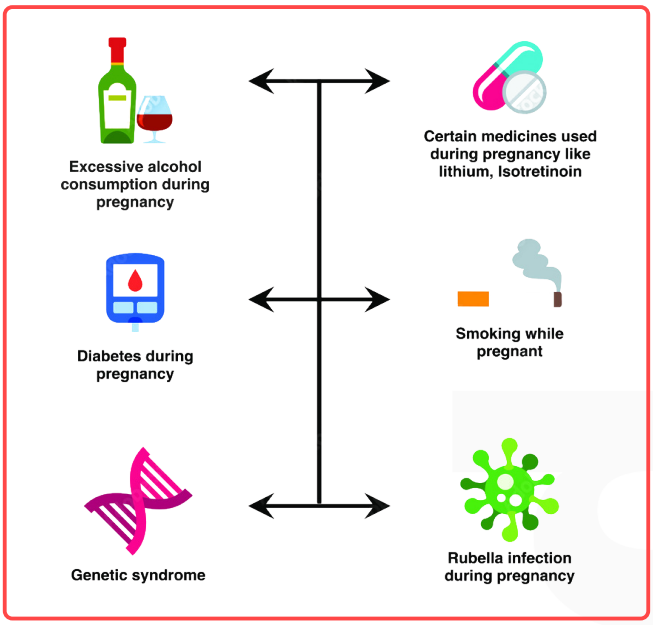
- The incidence is around ***8/1000 live births*** and is **higher in stillbirths and abortions.**
- ***10-15%*** have **complex anomalies** (i.e. more than one cardiac abnormality).
- ***10-15%*** have an associated **extra-cardiac anomaly.**

VSD	25-30%	**ASD**	6-8%	**PDA**	6-8%	**ToF**	5-7%
Coarctation	5-7%	**PS**	5-7%	**AS**	4-7%	**TGA**	3-5%

Relative frequency of the common types of CHD Disease

Etiology

- **Sporadic**
- **Genetic:** More in some families
- **Teratogens:** ie drugs
- **Part of a syndrome**
- **Multifactorial inheritance**
- **Maternal:**
  - Infection
  - Chronic disease
  - Irradiation
  - Teratogenic drugs



Recurrence risk in genetic CHD

- After 1 affected sibling is: 2-6%
- if 2 siblings are affected: 20-30%

Down syndrome (Trisomy 21)	AVSD (ECD), VSD, ASD
Edward syndrome (Trisomy 18)	VSD, ASD, PDA, Coarctation, Bicuspid aortic valve or pulmonary valve
Patau syndrome (Trisomy 13)	
Turner Syndrome (XO)	Bicuspid Aortic Valve, Coarctation of the Aorta
Noonan Syndrome	PS, ASD, Cardiomyopathy
Infant of diabetic mother	Hypertrophic cardiomyopathy, VSD, Conotruncal anomalies
Marfan syndrome	MVP, MR, dilated aortic root
Congenital Rubella	PDA, peripheral PS

Syndromes with Congenital Heart Disease

2 CONGENITAL ACYANOTIC HEART DISEASE

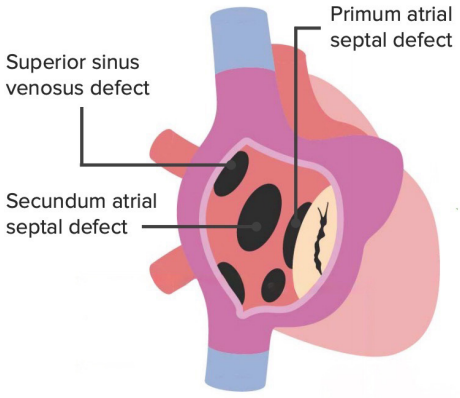
1. Atrial Septal Defect (ASD)

Types of ASD:

Ostium Secundum: defect in the **interatrial septum** at the fossa ovalis **(most common)**

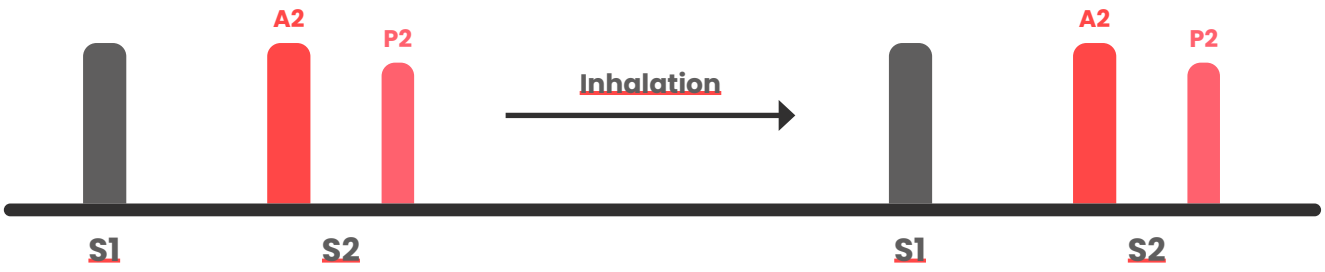
Ostium Primum: defect is at the **atrioventricular septum** (lower part of the septum). Cleft mitral leaflet, Abnormal AV junction and abnormal AV valves are found.

Sinus Venosus: defect in the upper part **near the SVC.**



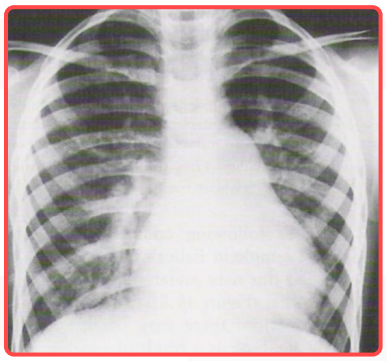
Clinical manifestations

Symptoms	Physical signs
Commonly presents in the second decade • None (commonly) • Growth failure • Recurrent chest infection/wheeze • Mild dyspnea and fatigue • Heart failure • Arrhythmias in old age • Cyanosis on effort	***Heart sounds:*** • **Wide fixed splitting** of S2 due to delayed closure of the pulmonary valve. ***Murmurs:*** • A **systolic flow murmur** over the pulmonary area. • A **diastolic rumbling murmur** over the tricuspid area.



Investigations

Chest X Ray



- The heart is enlarged
- Prominent pulmonary arteries
- Increased pulmonary vascular markings

ECG

Secundum ASD:

- Sinus rhythm
- Right axis deviation
- Partial or complete Rt. BBB (rsR' pattern)

Primum ASD:

- Left axis deviation with a negative deflection in lead aVF.

Echocardiography

Transthoracic echocardiography can demonstrate the anatomy in **neonates and children.**

Transesophageal echocardiography is needed in **older children and adults.**

Complications

- Pulmonary hypertension
- Pulmonary vascular disease
- Heart failure
- Pulmonary emboli
- Paradoxical emboli
- Brain abscess
- **Arrhythmias:** atrial fibrillation, atrial flutter.

Management

For complications

- Treat repeated **chest infection**, **heart failure** and **growth delay.**

For the defect: closure, either surgical or by catheter device.

Early closure: in **symptomatic** infants and infants with evidence of rt side volume overload.

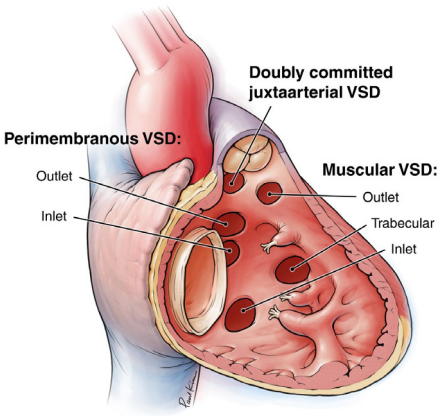
Elective closure: is usually done after 4-5 yrs **if no spontaneous closure.**

2. Ventricular Septal Defect (VSD)

VSDs constitutes 30% of all cases of congenital heart disease

Classification

According to size	Small	Moderate	Large
According to site	Muscular	Perimembranous	
According to site	Single	Multiple	



Clinical Picture

Presentation is usually early with a loud murmur noted on routine examination

Symptoms

- **Asymptomatic**
- **Heart failure** with breathlessness and FTT
- **Recurrent chest infections**
- **Cyanosis:** due to pulmonary vascular disease
- **Endocarditis**

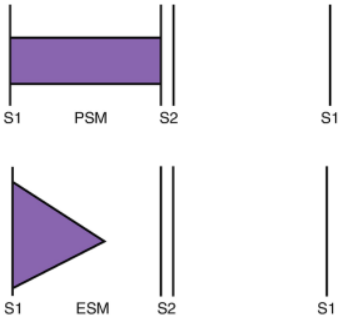
Physical signs

General:

- **Growth failure**
- **Heart failure:** Tachypnea, tachycardia and enlarged tender liver.

Local:

- Hyperactive precordium
- Parasternal thrill
- **Murmurs:**
  - **In small defects:** Loud pansystolic at 3rd & 4th left sternal edge.
  - **In large defects:** unimpressive ejection systolic murmur of increased pulmonary blood flow.
- **Variable intensity of pulmonary component of 2nd heart sound;** normal in small defects, loud in pulmonary HTN.



Natural history and complications of VSD

Usually asymptomatic

Small defects usually close spontaneously within the first few years of life with <10% requiring surgical closure.

Chest infection and congestive heart failure (CHF). Symptoms of HF may later resolve due to development of pulmonary hypertension.

Pulmonary hypertension:

- **Large left to right shunt**, increased pulmonary flow and pulmonary hypertension will lead to irreversible damage of the pulmonary capillary vascular bed (pulmonary vascular disease (PVD)).
- **Small defect:** usually no pulmonary hypertension

Acquired pulmonary stenosis

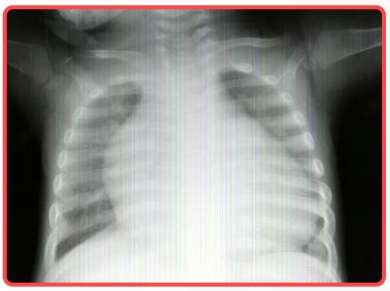
Infective endocarditis

Eisenminger syndrome: Advanced pulmonary vascular disease with ***cyanosis*** due to intracardiac right to left shunting, commonly **after the first 10 years of life.**

Investigations

Chest X Ray

- Enlarged heart
- Enlarged pulmonary arteries
- Increased pulmonary vascular markings



ECG

Varies from normal to grossly abnormal:

- Starts with LVH, then LVH & RVH
- P pulmonale and RVH in pulmonary hypertension (PH).

RVH is suggestive of PH and always warrants further investigation

Echocardiography

- Demonstrates the **ventricular septal defect** and determines its size, site and number as well as the size of the different cardiac chambers
- Demonstrates the **hemodynamic effects** (pulmonary artery pressure)

Management

Follow up in small defects

Medical therapy

- Treat repeated **chest infection**
- Manage **growth failure**
- Treat **heart failure if symptomatic:**
  - **Diuretics:** Fursemide or thiazide and spironolactone
  - **Angiotensin Converting Enzyme Inhibitor (ACEI).**
  - **Digoxin**

Closure: Surgical or by catheter device (device closure).

Early closure:

- If **severe symptoms** and failure to thrive
- **Pulmonary hypertension** with possible progression to PVD

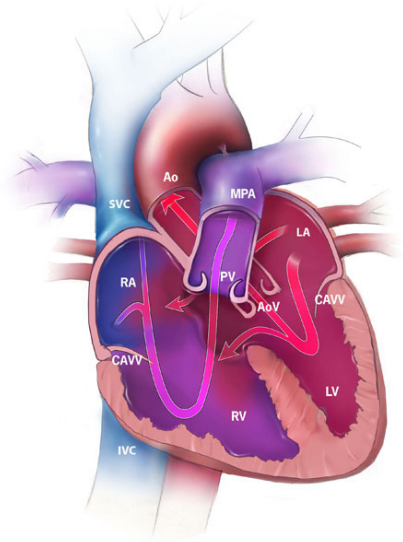
Elective closure.

3. Atrioventricular septal defect (AVSD)

Formerly known as endocardial cushion defect, and is commonly seen in *Trisomy 21*

Types

- **Complete:** ASD, VSD, and a common AV valve.
- **Incomplete:** Ostium Primum ASD and cleft mitral leaflet.



Clinical picture

Symptoms:

- **Partial AVSD:** may produce symptoms similar to ostium secundum ASD.
- **Complete AVSD:** usually have symptoms such as failure to thrive, tachypnea, diaphoresis with feeding, or recurrent bouts of pneumonia.

Auscultation:

- Murmur often inaudible in neonates.
- Loud pulmonary component of S2.
- Parasternal systolic murmur that is usually not as harsh as that of an isolated VSD

Management

As for VSD, but surgical correction is more hazardous.

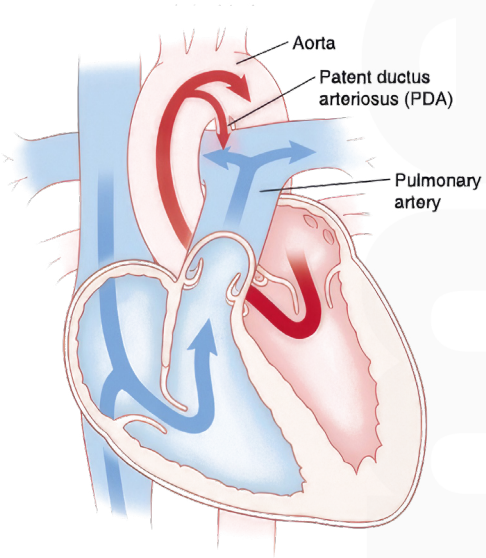
- In children with **Trisomy 21** AVSDs must be corrected **early in infancy.**
- Require **prophylaxis against infective endocarditis.**

4. Patent ductus arteriosus (PDA)

The ductus arteriosus normally connects the PA to the descending aorta in fetal life.

In utero:

- in the presence of collapsed alveoli and elevated right sided pressures, it diverts blood from the lung to descending aorta.
- It is ***maintained open by low PO2 and PGE.***



In preterm neonates, **failure to close** shortly after birth frequently occurs:

- It should spontaneously **close by 3 months**
- It may be suspected if a bounding pulse and a systolic murmur at the left sternal edge are present.
- When severe, heart failure can result.

In full term infants: it should undergo functional closure a **few hours after birth** due to the sudden increase in PO2.

Clinical Picture

Symptoms in older children

- Asymptomatic
- Repeated chest infection
- Dyspnea
- Growth retardation
- Heart failure
- Infective endocarditis
- Eisenminger syndrome

Physical examination

- Wide pulse pressure
- Water hammer pulse
- A continuous thrill and a ***machinery murmur*** beneath the left clavicle figure.

> **Note**
>
> The characteristic murmur of PDA—continuous "machinery" murmur radiating to the back—is **rarely heard before 6 months of age.**

Investigations

CXR

- Enlarged heart
- Enlarged pulmonary arteries
- Increased pulmonary vascular markings

ECG

Usually normal, or:

- Left ventricular hypertrophy with large left to right shunt
- Right ventricular hypertrophy with pulmonary hypertension

Echocardiography

- Ducts are readily identified by cross-sectional echo and Doppler.

Management

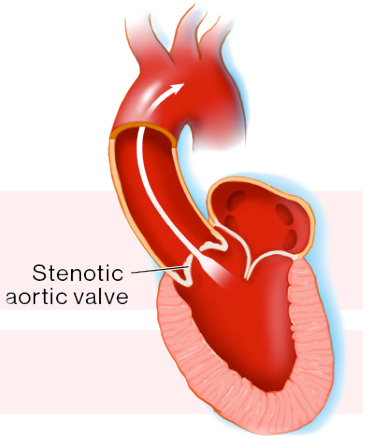
In preterm infants: Closes spontaneously in most.

If symptomatic in neonates: fluid restriction, diuretics, close it by indomethacin, ibubrufen or surgical ligation.

In young children, even with an asymptomatic PDA: closure is recommended to abolish the risk of infective endocarditis, using:

- Surgical ligation
- Coil device transvenous occlusion

5. Congenital aortic stenosis



Narrowing of the AV prevents the valve from opening properly and obstructs the blood flow from the left ventricle to the aorta.

This can ↓ the amount of blood that flows forward to the body.

It may be associated with **mitral stenosis or coarctation of the aorta.**

Clinical Picture

Symptoms

- Breathlessness, fainting or weakness with activity.
- Sensation of feeling the heart beat
- Chest pain, angina-type

> **Note**
> Aortic stenosis may show no symptoms until late in the course of the disease.

Physical signs

- **Pulse:** Small volume, slow rising, plateau-type pulse (pulsus parvus et tardus.)
- Carotid thrill.
- **Blood pressure:** may be low.
- **Palpable chest thrill** (1st aortic area) or heave
- **Auscultation:**
  - Ejection systolic murmur heard maximally in aortic area and radiating to both sides of the neck
  - Diminished aortic component of S2
  - Ejection click heard at apex.

Investigations

Chest X-ray: cardiomegaly and/or post stenotic dilatation.

ECG: may show left ventricular hypertrophy

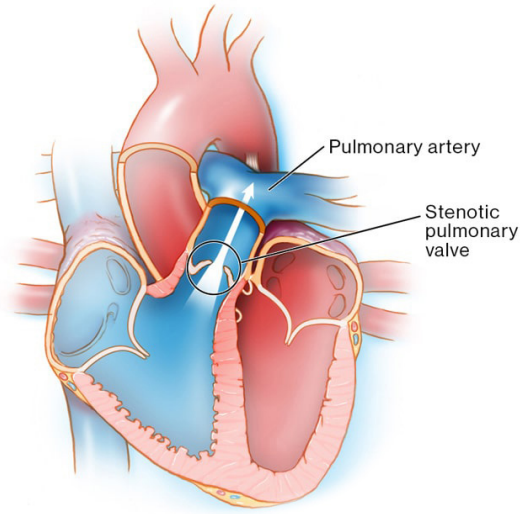
Echocardiogram: is usually ***diagnostic.***

Complications

- LVH (enlargement) due to the extra work of pushing blood through the narrowed valve
- Left-sided heart failure
- Arrhythmias and sudden death
  - → **avoid strenous exercise**
- Endocarditis and valve infection
- Valve calcification

Treatment

- Anti-failure measures
- Infective endocarditis prophylaxis
- Balloon valvuloplasty
- Surgery: Aortic valve replacement



6. Pulmonary Stenosis

Clinical picture

Symptoms

- **Frequently asymptomatic** and accidentally discovered
- ***Critical PS:*** severe pulmonary stenosis that presents in the neonatal period in the first few days of life with duct dependent pulmonary blood flow and cyanosis.

Physical signs

- Ejection systolic murmur at 2nd and 3rd left intercostals space, radiating to the back
- Ejection click

Investigations

Chest X-ray: Post stenotic dilatation of the pulmonary artery.

ECG: Evidence of right ventricular hypertrophy.

Echocardiography: ***Diagnostic*** (valvular affection and the RVH if present).

Management

Prophylaxis against infective endocarditis

In severe stenosis with marked right ventricular hypertrophy:

- Transvenous balloon dilatation of the pulmonary valve
- Surgery

7. Coarctation of the Aorta

The key to the clinical diagnosis of coarctation of the aorta is the recognition of *weak or absent femoral pulses.*

Coarctation of the Aorta is often associated with other cardiac anomalies, especially **bicuspid aortic valve and ventricular septal defect.**

> **Note**
>
> Palpation of the femoral pulses must be routinely performed during the cardiovascular examination of any child

Clinical picture

Symptoms:

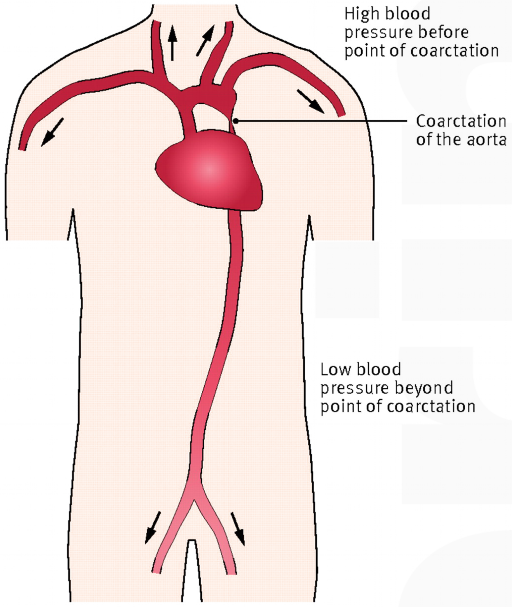
In neonates:

- Circulatory collapse on PDA closure
- Severe heart failure

Older children: Hypertension

Physical examination

- Weak femoral pulses or radio-femoral delay
- ABP in LL < UL
- Murmur heard over the back between the scapulae



Investigations

CXR

- May show **cardiomegaly and increased bronchovascular markings** in the presence of **HF.**
- **Rib-notching** in **older children** due to dilated collateral arteries.

ECG: LVH in children

Echocardiography: ***Diagnostic***

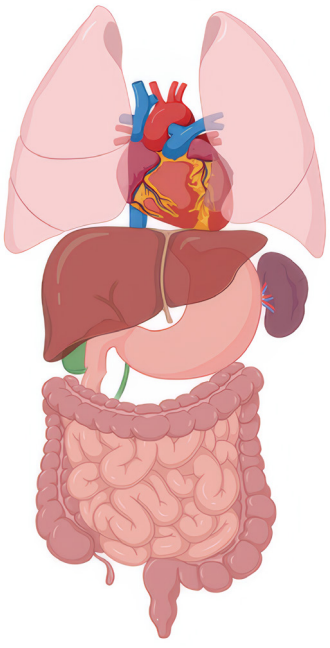
Management

- Surgery or catheter dilatation

8. Congenital cardiac malposition

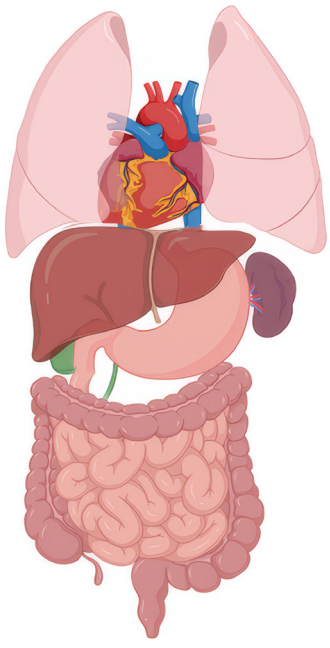
Situs solitus

Liver to the right + stomach to the left + aorta to the left + IVC to the right + cardiac apex to the left



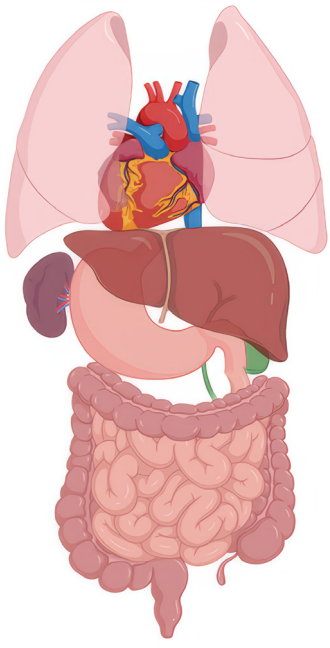
Isolated Dextrocardia

Liver to the right + stomach to the left + **aorta to the right** + **IVC to the left** + **cardiac apex to the right**



Situs inversus totalis

Liver to the left + **stomach to the right** + **aorta to the right**+ **IVC to the left** + **cardiac apex to right**



CONGENITAL CYANOTIC HEART DISEASE

Cyanotic Heart Disease may be associated with:

↑ Pulmonary flow (Plethora)

- Transposition of Great Vessels
- Truncus Arteriosus

↓ Pulmonary flow (Oligemia)

- Tetralogy of Fallot
- Tricuspid Atresia

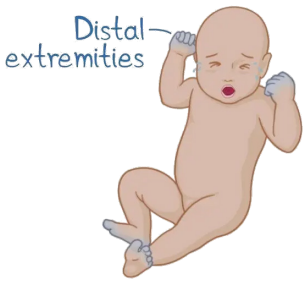
Other congenital heart defects that can cause cyanosis include:

- Ebstein's Anomaly
- Total Anomalous Pulmonary Venous Return
- Hypoplastic Left Heart Syndrome

Cyanosis

Bluish discoloration of the skin and mucous membranes due to increased amount of reduced hemoglobin *above 5 gm/dl* in the superficial capillaries.

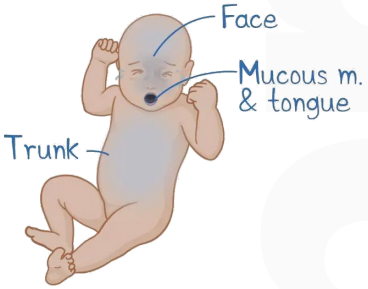
Peripheral cyanosis

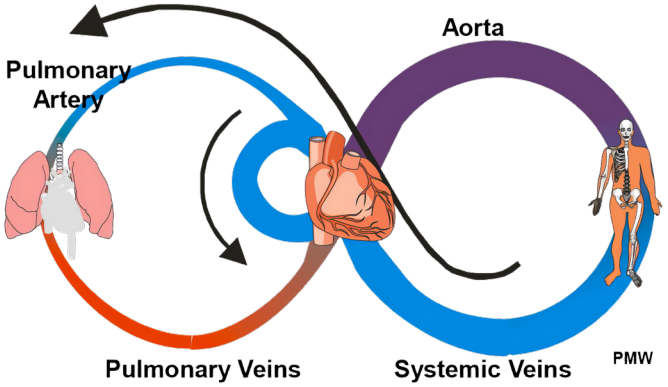


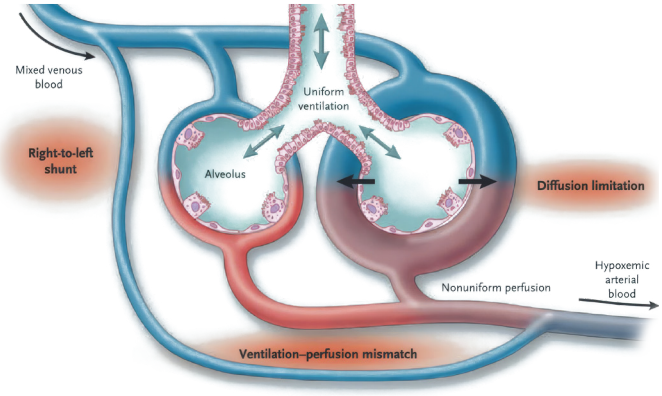
- **C** • Cold
- **O** • Obstruction
- **L** • LVF and shock
- **D** • Decreased cardiac output

Central cyanosis

- **P** • Polycythemia
- **A** • Altitude
- **L** • Lung disease
- **M** • Met - sulfhemoglobinemia
- **S** • Shunt



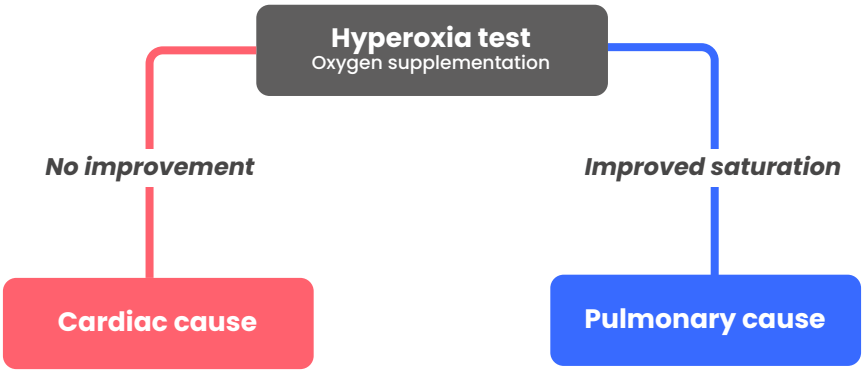




Pathophysiology of cardiac cyanosis

Testing for cardiac cyanosis

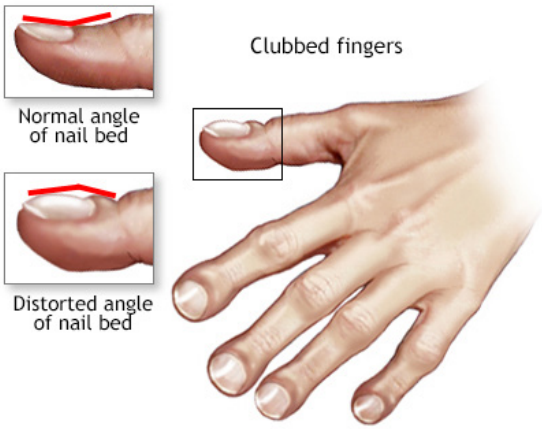
Pulmonary vs cardiac cause



Clubbing

Degrees of clubbing

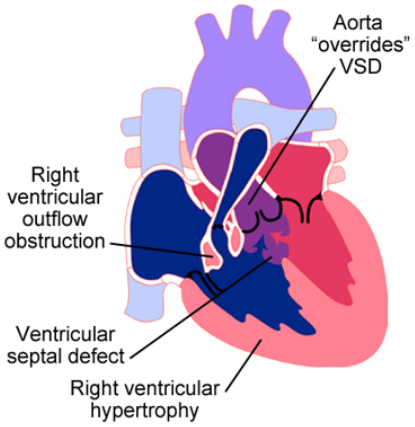
Grade 1	Fluctuation of the nail bed
Grade 2	Obliteration of the Lovibond angle
Grade 3	Parrot beak or drumstick appearance
Grade 4	Hypertrophic osteo arthropathy (HOA)



Complications of Persistent Cyanosis

- Polycythaemia
- Relative anaemia
- CNS abscess
- Thromboembolic stroke
- Clubbing
- Infection
- Poor growth

1. Tetralogy of Fallot (TOF)



It occurs slightly more often in males than in females.

Most common cyanotic heart disease

The characteristic four abnormalities in Tetralogy of Fallot (TOF) are:

1. Pulmonary stenosis

Right ventricular outflow obstruction by a narrowing (stenosis) that is either:

- Valvular
- Just below the pulmonary valve (infundibular)

2. Ventricular septal defect

The defect is at the 'outlet septum'

In the majority of cases is single and large.

3. Overriding Aorta

The aortic root overrides VSD:

- The aortic valve is not restricted to the left ventricle, thus having biventricular connections.
- The degree of override is quite variable, being between 5-95% of the valve being connected to the right ventricle.

4. Right ventricular hypertrophy

The right ventricle is more muscular than normal due to the obstruction to the right outflow tract.

Types

The severity of cyanosis in tetralogy of Fallot depends on the severity or extent of the anatomical defects and encompasses a spectrum:

- **"Extreme" Fallot or "pseudotruncus":** In which the pulmonary stenosis is so severe that the pulmonary artery is almost atretic or absent
- **"Pink" Fallot:** In which a mild pulmonic stenosis and a discrete overriding of the aorta over the interventricular septal defect result in minimal cyanosis.
- **Pentalogy of Fallot:** Tetralogy of Fallot and ASD
- **Trilogy of Fallot:** PS, RVH, and ASD.

'Pink' Tetralogy of Fallot

'Blue' Tetralogy of Fallot

'Profound cyanosis' Tetralogy of Fallot

Clinical picture

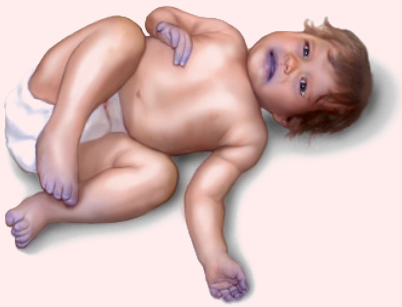
Presentation

1. Cyanosis: is usually observed weeks after delivery.

- Absence of cyanosis at birth is attributed to the presence of a **patent ductus arteriosus** in the early postnatal life that redirects a large portion of the partially oxygenated blood into the lungs, increasing flow through the pulmonary circulation with relatively better oxygenation.

Cyanotic spells

- Acute severe cyanosis or hypoxic "tet spells" may result from an increase in resistance to blood flow to the lungs with increased preferential flow of desaturated blood to the body.
- Tet spells are potentially lethal and are characterized by a sudden, marked increase in cyanosis, syncope, +/- hypoxic brain injury and death.



2. Clubbing develops and progresses with time

3. Birth weight is low; growth is retarded. Development and puberty may be delayed.

4. Poor feeding, breathlessness and agitation.

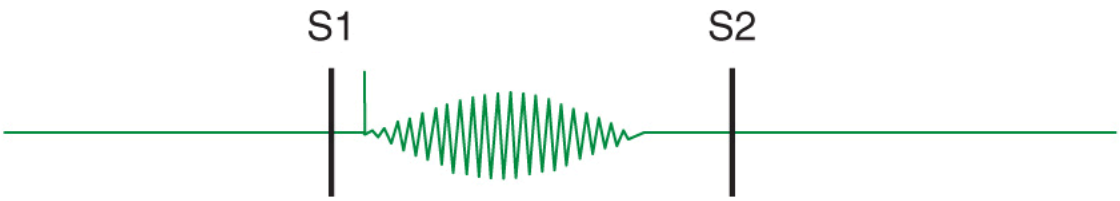
5. Dyspnoea on exertion is common, usually after prolonged crying.

6. Squatting to rest whilst exercising is a feature of a right-to-left shunt in children.

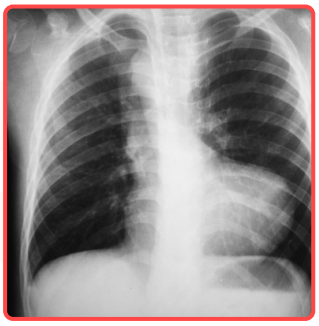
Cardiac Examination

In older children with long-standing cyanosis without surgical treatment:

1. **Right ventricular predominance** on palpation or possibly a **bulging left hemithorax.**
2. **Systolic thrill** at the lower left sternal border.
3. **Single S2** (Pulmonic valve closure not heard).
4. **Systolic ejection murmur** over **Pulmonary area.**



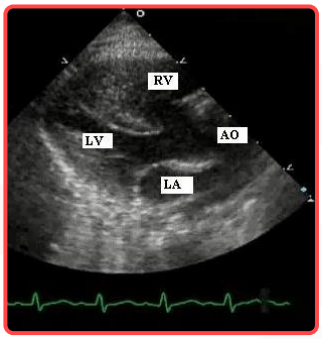
Investigations



1. Fetal echocardiography: is performed on high risk mothers

2. Chest X-ray

- Normal heart size
- Concavity in area of main pulmonary artery (silhouette compared to a boot or wooden shoe – ***'coeur en sabot'***)
- The lung fields are **oligemic**
- The **aorta** is usually large



3. Echocardiography: *For definitive diagnosis.*

- It demonstrates the abnormal anatomy.

Management

Medical

1. In severe form in neonates:

- **IV prostaglandin E1** is used to keep the ductus arteriosus open whilst waiting for surgery.
- Oxygen, keep warm, check blood glucose.

2. Iron supplementation due to relative iron deficiency in polycythemia.

3. Emergency management of 'Tet' spells

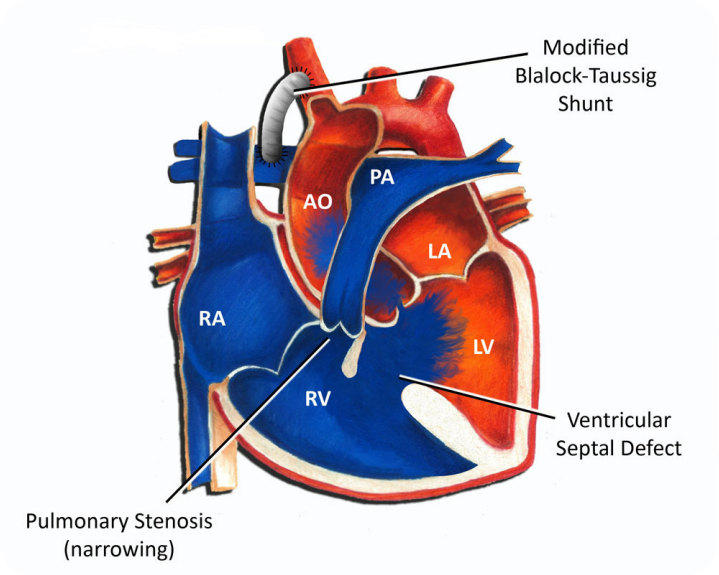
a. Placing infant on abdomen in **knee-chest position** with calming.
b. **Oxygen**
c. **Morphine**
d. **+/- Intravenous Propanolol** (0.1mg\kg/dose).

Oral propranolol (1mg/Kg/dose) may reduce the number and severity of attacks but best to refer for surgery as soon as possible.

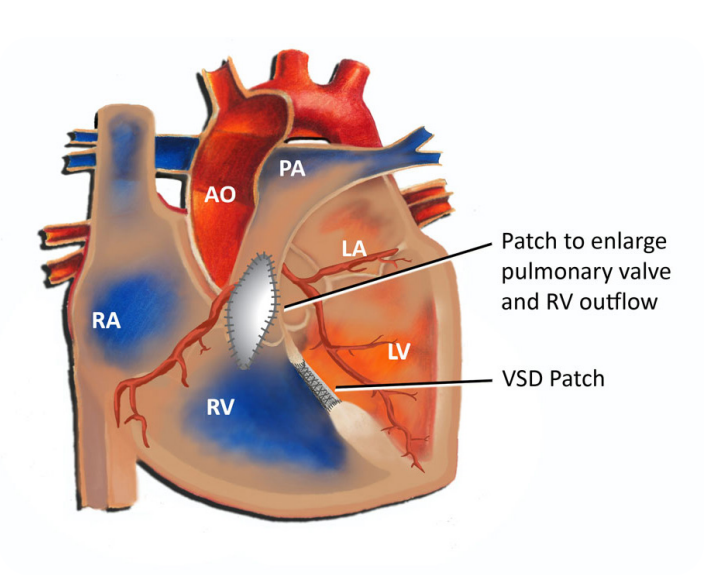
Surgical

Choice is between early palliative systemic-to-pulmonary artery shunt or corrective open heart surgery:

1. **Palliative surgery:** usually a **modified Blalock-Taussig shunt** from subclavian to pulmonary artery. This procedure is performed on infants with severe variants.
2. **Total surgical repair:** is now done in infants (less than 1y) with little risk.



Modified Blalock-Taussig shunt



Corrective surgery

Complications

- **Polycythemia and hyperviscosity** syndrome with secondary thrombosis.
- **Iron deficiency anemia** (risk factor for thrombosis)
- **Thrombocytopenia** is common in patients with cyanotic CHD leading to bleeding.
- **Infective Endocarditis**
- **Pulmonary Tuberculosis**

Prognosis

This depends on the severity of the right ventricular outflow tract obstruction and the efficacy of surgical treatment in the proper time.

In untreated patients:

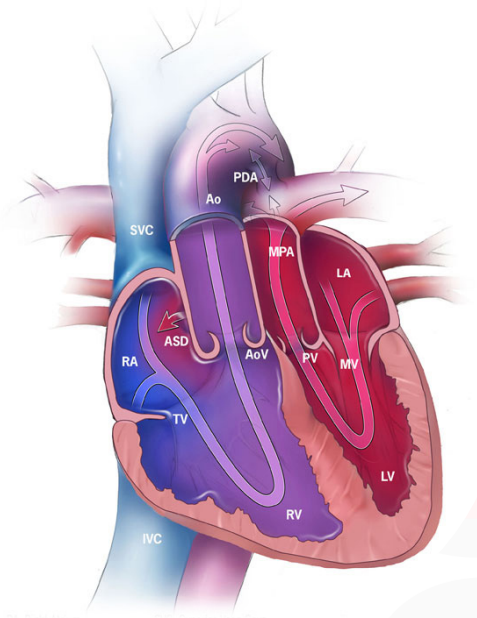
- ***25%*** with TOF and RVOT obstruction die within the **first year of life.**
- ***95%*** of patients die **by 40 years.**
- Delayed growth and development including puberty.

After surgical correction:

- Many patients are usually without symptoms and can lead normal lives.
- However, some may have mild outflow obstruction and/or mild to severe pulmonary insufficiency.

2. Transposition of The Great Arteries

The aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle.



Most common cyanotic heart disease in neonatal period

Epidemiology

- More in boys.
- Aetiology is multifactorial.

Pathophysiology:

- The pulmonary and systemic circulations function in parallel, rather than in series.
- A shunt has to be present for the patient to live.

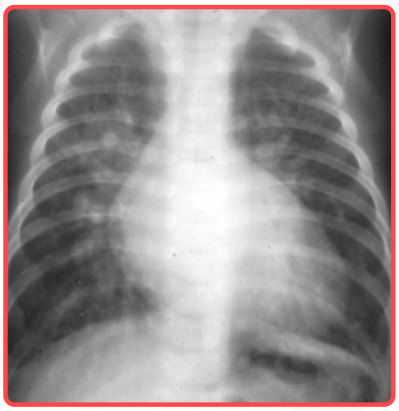
Presentation

- Patients with TGA usually present with ***cyanosis in the newborn period***
- Clinical manifestations and course are influenced predominantly by the degree of inter-circulatory mixing and the presence of associated anatomic lesions

Investigations

CXR

- May appear normal or may show the classic ***"egg on a string" or "egg on its side"*** appearance (1/3 of patients)



Echocardiography

Management

Must be surgically corrected

Management in the neonate

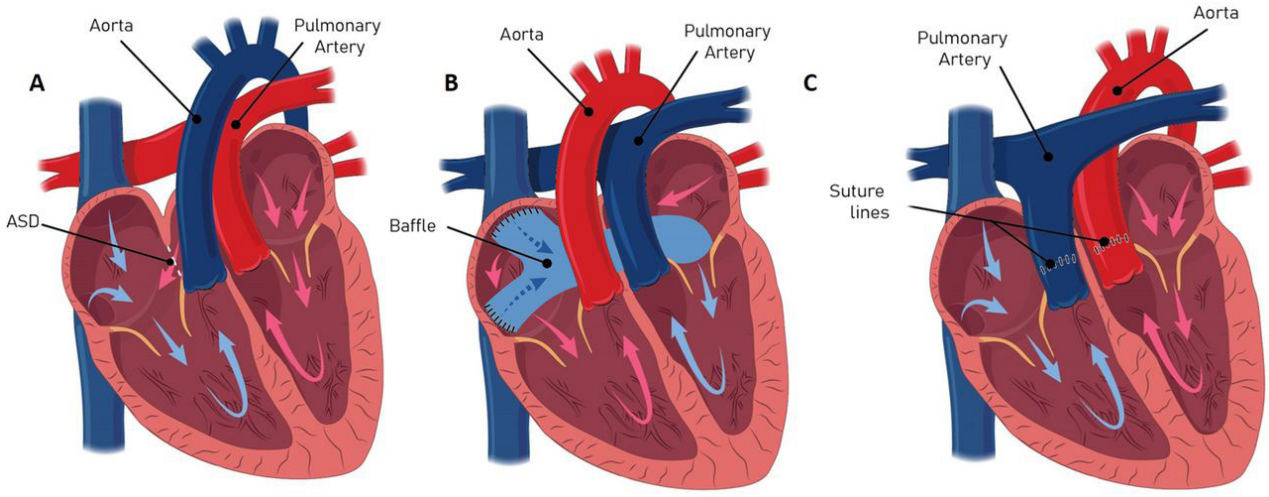
- **IV prostaglandin E1 infusion:** essential for maintaining ductal patency.
- Correction of metabolic acidosis with **fluid replacement.**
- **Mechanical ventilation** may be necessary if pulmonary edema develops.

- **Cardiac catheterization and balloon atrial septostomy** in **severely hypoxemic patients** with an inadequate atrial communication and insufficient mixing.

Surgical Care

- Surgical approach depends on the age of the patient at presentation, the presence of associated congenital cardiac lesions, and experience of the cardiothoracic surgeon.

> **Note**
>
> Although the arterial switch operation requires moving the coronary arteries to the neo-aorta, the incidence of coronary a. insufficiency after the switch operation is low.



Complications

- **Polycythemia with hyperviscosity and thrombocytopenia** as in TOF.
- **Iron deficiency anemia** (together with polycythemia predisposes to thrombosis)
- **Congestive heart failure**
- **Arrhythmia**

Prognosis

The mortality rate in untreated patients is approximately:

- **30%** in the first week
- **50%** in the first month
- **90%** by the end of the first year.

With improved diagnostic, medical, and surgical techniques the short and mid-term survival rate **exceeds 90%.**

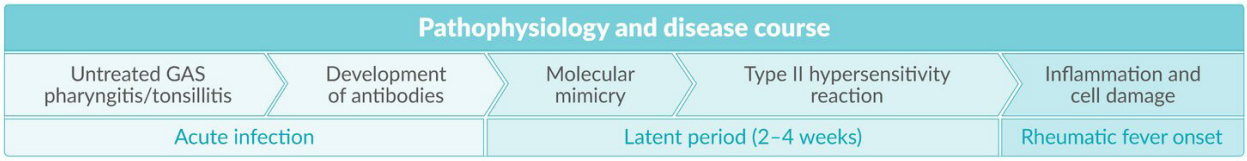
4 ACUTE RHEUMATIC FEVER

It is an acute autoimmune inflammatory process that occurs 2-3 weeks following a rheumatogenic Group A B hemolytic streptococcal pharyngeal (upper respiratory tract) infection.

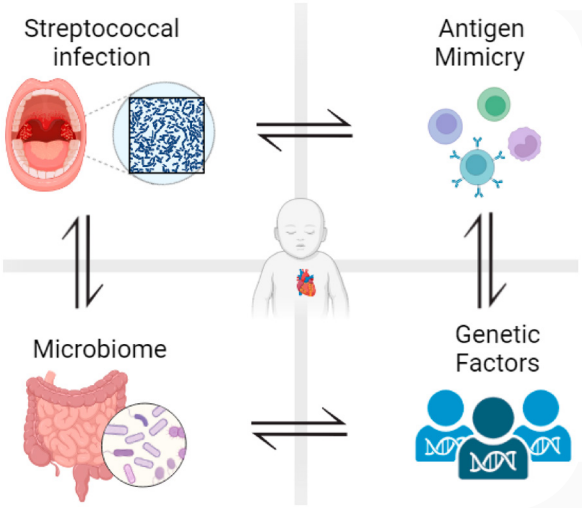
It affects mainly the joints heart and CNS and less frequently the skin, and SC tissues.

It is the commonest cause of acquired heart disease (rheumatic heart disease=RHD) in developing countries.

Etiology



- The most accepted theory for ARF and is an **immune-mediated pathogenesis.**
- Common epitopes are shared (antigenic mimicry) between certain components of **group A streptococcus** (e.g., M protein, protoplast membrane, cell wall group A carbohydrate, capsular hyaluronate) and specific mammalian tissues (e.g., heart, brain, joint).
- Certain serotypes of group A streptococcus (e.g., M types 1, 3, 5, 6, 18, 24) are more frequently isolated from patients with ARF than others.
- **Low socioeconomic standards**, crowding and poverty are predisposing factors for the spread of group A streptococcal infections and ARF.
- Following an infection with a rheumatogenic strain of GABHS there is a chance of developing ARF in 3% of patients but if the patient has a previous history of ARF the chance of developing a recurrence rises to 65%.
- **Genetic predisposition** to ARF is suggested.



Clinical Manifestations

Age:

- Most commonly 5 - 15 years age.
- In Egypt: 4-12yrs

History of streptococcal pharyngitis

- 2 - 3 weeks before the disease.
- In chorea it may be up to 6 weeks - 6 months.

Diagnosis of ARF is based on the "Modified Jones' Criteria"

Major Manifestations	Minor Manifestations	Evidence of Strep. infection
1. Carditis 2. Polyarthritis 3. Chorea 4. Erythema marginatum 5. Subcutaneous nodules	1. Fever 2. Arthralgia 3. +ve Acute phase reactants 4. Prolonged P-R interval.	1. Recent history of scarlet fever. 2. +ve throat cultures. 3. ↑ ASO (> 333 IU) & other Strep. antibodies.

Other findings (not included in Jones' Criteria):

1. Abdominal pain. 2. Rheumatic pneumonia. 3. Epistaxis.

Diagnosis

Evidence of recent streptococcal infection +

2 major manifestations

OR

1 major manifestation AND **2 minor manifestations**

Jones Criteria are not completely fulfilled in:

- **Rheumatic chorea:** it may occur as an isolated finding, not accompanied by any minor or major manifestations, or evidence of recent GABHS infection.
- **Insidious onset carditis.**
- **Patients with RHD:** only one major, fever, arthralgia or raised CRP may suggest recurrence.

Note

If arthritis is present: arthralgia is not considered a minor manifestation.

If carditis is present: prolonged P-R interval is not a minor manifestation.

1. Carditis

It affects about 50–60% of all cases of ARF.

It is the most serious manifestation of ARF due to the residual damage affecting the valves.

- ***Pancarditis affecting the:*** pericardium, myocardium and endocarduim varies in severity from fulminant to mild, transient cardiac involvement.
- Moderate-to-severe rheumatic carditis can result in cardiomegaly and congestive heart failure with hepatomegaly and peripheral and pulmonary edema.

a. Pericarditis:

- Stitching pain + pericardial rub (dry pericarditis) or mild effusion.

b. Myocarditis

- Tachycardia out of proportion of fever.
- Dilatation of the heart.
- Dilatation of A-V rings (leading to incompetent valves).
- Muffled heart sounds.
- Gallop rhythm.
- Prolonged P-R interval.
- Heart failure.

c. Endocarditis

Affecting the heart valves, leading to:

1. **Mitral Regurge (the most common rheumatic valve lesion in children)**
2. **Mitral Stenosis:** due to edema of mitral valve → transient functional mid-diastolic murmur "Carrey Combs" murmur.
3. **Aortic Regurge.**
4. In children with previous rheumatic heart disease (RHD), there will be:
   - **A change of the character of the old murmur.**
   - **Appearance of new murmurs.**

Note

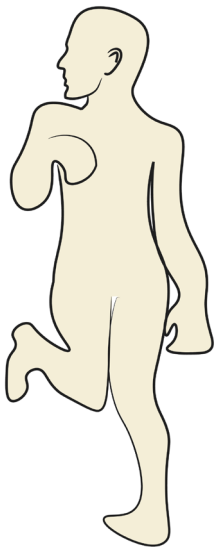
- **Echocardiographic findings include:** pericardial effusion, and aortic and/or mitral regurgitation.
- **Once carditis, always carditis:** Recurrence prefers first site affected.

2. Polyarthritis

- Affects mainly **large joints** as knee, ankle, hip, shoulder, elbow, or wrist joints.
- **The affected joints are:** red, hot, swollen, painful, and tender with loss of function.
- It is a **fleeting** arthritis, migrating from one joint to another, leaving no residual damage or deformity in the affected joints.
- There is an apparent inverse relationship between the severity of arthritis and the severity of cardiac involvement.

Shows dramatic response to salicylates (within 24 hr).

3. Chorea



- Sydenham's chorea occurs in about 10–15% of patients with ARF and usually presents as an isolated, frequently subtle, neurologic behavior disorder.
- The latent period from acute group A streptococcal infection to chorea is usually longer than for arthritis or carditis and can be **months.**
- It is due to involvement of **basal ganglia** (especially Caudate nucleus).
- **Females** are twice affected than males, usually at age 10-15 years.
- It may present alone (isolated rheumatic chorea); without any other major or minor manifestations. Acute phase reactants are usually normal. Sometimes, it may be associated with carditis.

Onset is usually insidious characterized by involuntary, semi- purposeful movements that are sudden, jerky, irregular and non-repetitive. They affect face, tongue and limbs.

Manifestations include:

Emotional lability and facial grimaces:

- Emotional instability and behavioural disorders are usually present, with laughing or crying with no reason.
- In **severe cases** with persistent excitement, insomnia, irritability, and confusion might occur → **Maniacal chorea.**

Tongue and palate:

- Dysarthria, and failure to sustain the tongue outside the mouth except by biting it.

Upper limb:

- Dropping of objects.
- Inability to do fine movements properly (hand-writing).

- **Milkmaid's grip sign:** irregular contractions of the muscles of the hands while squeezing the examiner's fingers.
- **Pronator sign:** Pronation of forearm if asked to hold the upper limbs above the head.
- **Spooning of the hand:** stretching the hands and upper limbs → (flexion of wrist and hyperextension of fingers).
- **Piano sign:** piano-like movements in the extended fingers

Lower limb: walking may become difficult, with frequent falling.

Weakness, hypotonia and hypo-reflexia:

- Are usually associated by a weak grip.
- The limb appears paralyzed (Chorea mollis or paralytica) from the severe weakness and hypotonia.

Involuntary movements are accentuated by emotions, excitement and stress and disappear during sleep.

It may be bilateral, asymmetrical, or may be unilateral (Hemi-chorea) or monochorea.

Prognosis

- The course is self limited, but may be prolonged up to **months.**
- **Frequent recurrences** occur.
- It might recur during pregnancy → **Chorea gravidarum.**
- It **does not** lead to **permanent neurologic sequelae.**

4. Subcutaneous Nodules

Rare and present in severe attacks usually with carditis.

- Nodule are firm, non-tender, and range in size from 0.1 - 1 cm in diameter. The skin over it is freely mobile and not inflamed.

Site: Most commonly over the extensor surfaces of small and large joints, and bony prominences as knuckles, elbows, scapula, vertebral spines, knees, chin of tibia, scalp and mastoid process.

5. Erythema Marginatum

Rare but characteristic rash of ARF.

- It starts as **pink, slightly raised macules**, which **fade in the center and extend peripherally.**
- The rings coalesce to form a **serpiginous** pattern, with an **irregular** border.
- It occurs primarily on the **trunk and extremities**, but **not on the face.**
- It can be accentuated by warming the skin.
- They are **not painful, not itchy, and not markedly elevated.**

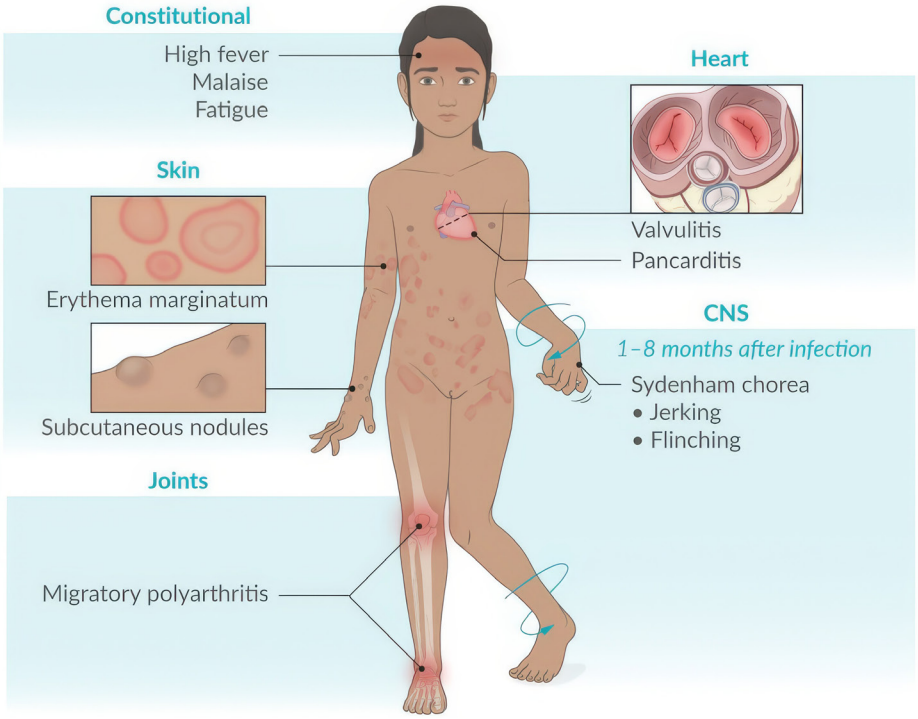
Acute rheumatic fever

Etiology
Previous GAS pharyngitis/tonsillitis without antibiotic treatment

Peak incidence
5–15 years of age

Diagnosis
Evidence of a previous GAS infection (e.g., throat culture, rapid antigen detection test, antistreptolysin O, antistreptococcal DNase B test)
Revised Jones criteria

Treatment
Antibiotic treatment of underlying GAS infection
Symptomatic treatment of arthritis and fever



Investigations

1. Acute Phase Reactants

Collectively, these are considered as one minor manifestation.

They are non-specific indicators of inflammation. There is:

- Leukocytosis.
- ↑↑ ESR (N = 0-13 mm/hr).
- ↑↑ C-Reactive protein.

2. ECG

- Prolonged P-R interval.
- Evidence of carditis or pericarditis.

3. Evidence of previous streptococcal infection

- ↑↑ ASO titre > 333 IU in children.
- ↑↑ Other streptococcal antibodies.

4. X-ray chest and heart: Cardiomegaly and Pericardial effusion may be seen.

5. Throat culture: GABH streptococci might be demonstrated.

6. Echo: for cardiac function, valvular affection, effusion, and follow up.

Treatment

A. Prevention

1. Prevention of 1st attack (1ry prevention)

Preventing Streptococcal infections: by improving housing conditions, good nutrition, avoiding over-crowding and contact with infected children.

TTT of Strept throat infections: by giving 10 days antibiotics to eradicate GABHS from the throat.

Drugs used:

- A single dose of **long-acting penicillin (LAP)** is effective.
- Oral Amoxicillin, Erythromycin or Cephalosporins for 10 days or Azithromycin for 5 days may also be uesd.

Early and proper treatment of streptococcal pharyngitis is highly effective in preventing first attacks of ARF.

2. Prevention of 2nd attack of Rheumatic Fever (2ry prevention)

Drugs used:

- **Long acting penicillin 1.2 million units/2 weeks** → is the best.
- Oral penicillin V (Ospen) : 250.000 IU once or twice/day.
- Oral sulfadiazine 0.5 - 1 gm once/day.
- Erythromycin 250 mg twice/day.

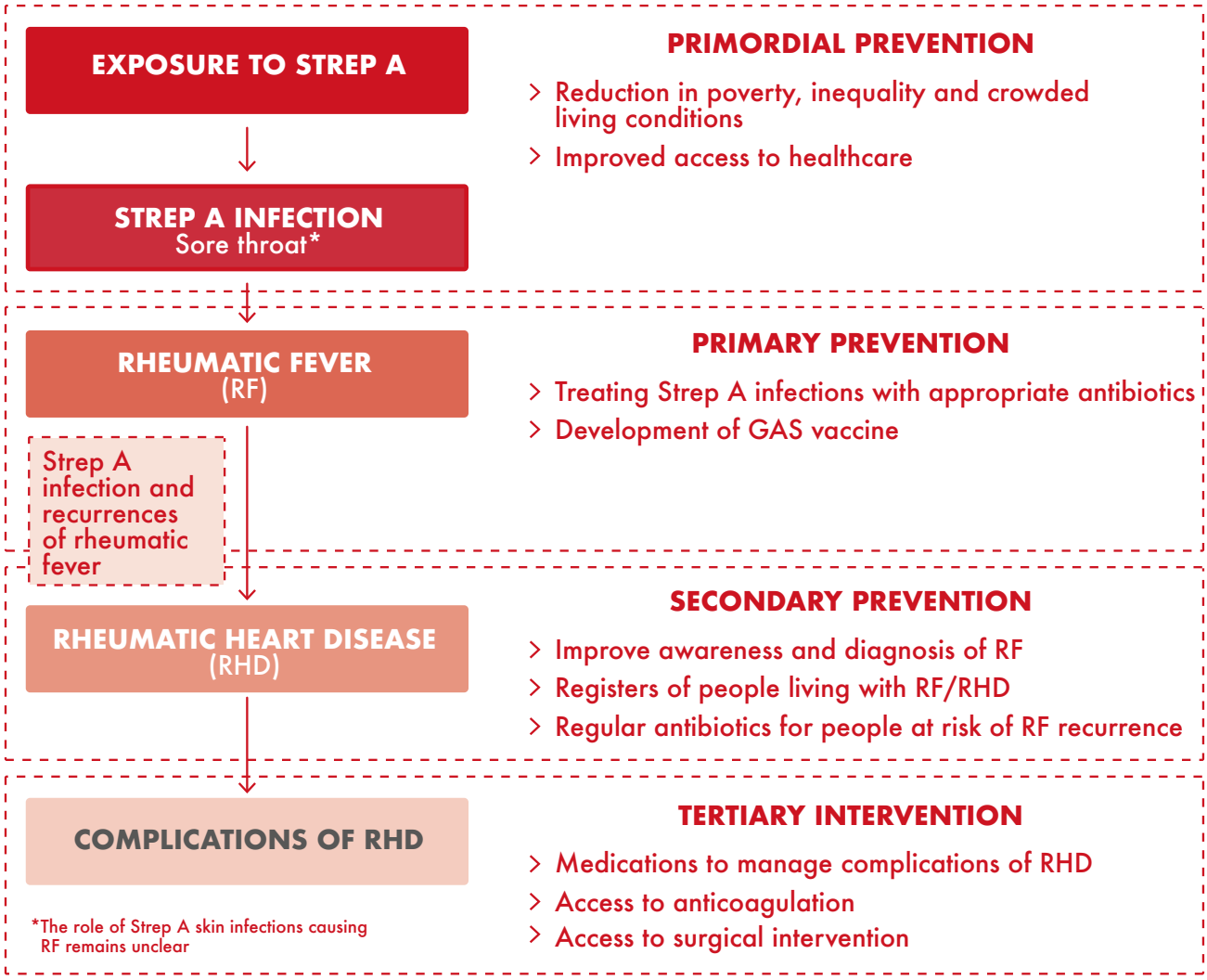
3. Prevention of Infective Bacterial Endocarditis

- Procedures with possible bacteremia should be done under an umbrella of antibiotics.

Continuation of prophylaxis:

- **In absence of cardiac involvement:** Prophylaxis against Strep. infection should be continued for at least **5 years after the last attack** or **21 years** (whichever is longer).
- **If cardiac affection is present:** it should be continued up to at least **25 years age.**

Recent WHO recommendations recommend continuation of prophylaxis for life in RHD.



B. Curative TTT

1. Antibiotics

- Once the diagnosis of acute rheumatic fever has been established and regardless of the throat culture results.
- ABs are given to eradicate group A streptococcus from the upper respiratory tract.

Drugs used:

- 10 day course of **Procaine penicillin IM. Inj 600.000 iu/d**, or
- **Orally administered penicillin or erythromycin** for 10 days, or
- **A single intramuscular injection of benzathine penicillin**.

After this initial course of antibiotic therapy, the patient should be started on long-term long acting penicillin prophylaxis/2 weeks.

2. Anti-inflammatory drugs

1. **Patients with typical migratory polyarthritis:** should be treated with **oral salicylates.**
2. **Patients with carditis and cardiomegaly or congestive heart failure:** should receive **corticosteroids (prednisone).**
3. **Patients with both arthritis and carditis** should receive **prednisone.**

a- Salicylates: Is the 1st choice for **isolated arthritis.**

- A dose of **100 mg/kg/day** in 3-4 divided doses usually induces rapid clinical improvement.
- After improvement, lower doses of **75 mg/kg/d** in divided doses PO are used for **4 wks.**

b- Steroids:

- Prednisone is used if there is **Rheumatic carditis,** for its more powerful anti-inflammatory action.
- The usual dose is **2 mg/kg/24 hr** in 4 divided doses until sedimentation rate is normal, then tapered gradually over 2 weeks; by reducing the dose by 5 mg/24 hr every 2–3 days.
- Prednisone tapering should be done **under an umbrella of aspirin** at 75 mg/kg/24 hr in 3-4 divided doses for 6 wk. to prevent rebound.

3. Bed rest: Until ESR returns to normal, especially if carditis is present.

4. Treat congestive heart failure accordingly (see later)

5. Treatment of Rheumatic Chorea

A. Usually it needs Supportive care, and understanding attendants.

B. Antistreptococcal prophylaxis should be given.

C. Drugs:

- Phenobarbitone: 3 - 5 mg/kg 3 times daily. Is the drug of choice, or
- Chloropromazine: 1 mg/kg/d, or
- Diazepam (Valium): 0.2 mg/kg/d, or
- Haloperidol: 10-30 ug /Kg/day in 2 divided doses (observe for toxicity).

Complications

- The arthritis and chorea of acute rheumatic fever resolve completely without sequelae.
- Therefore, the long-term sequelae of rheumatic fever is usually **limited to the heart (RF licks the joints and bites the heart).**
- RHD increases the risk for developing **infective endocarditis** during episodes of transient bacteremia.

Prognosis

It depends on the clinical manifestations, the severity and the recurrences:

- **Patients without carditis during the initial episode** are unlikely to have carditis with recurrences.
- **Patients with carditis in the initial episode** are likely to have carditis with recurrences. The risk of permanent heart damage increases with each recurrence.

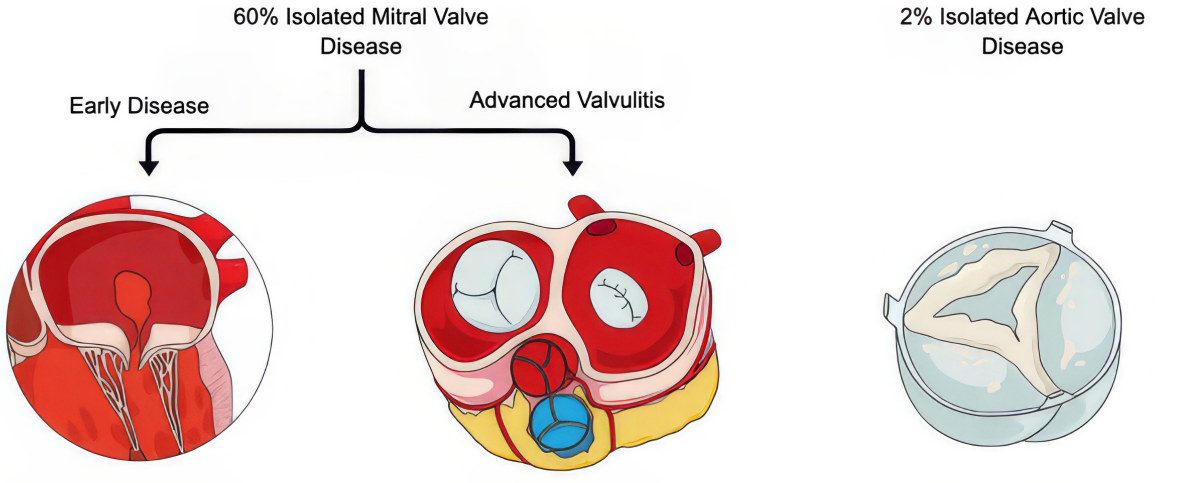
Approximately 20% of patients who present with "pure" chorea develop rheumatic heart disease within 20 yr.

Therefore, patients with chorea, even in the absence of other manifestations of rheumatic fever, require long-term antibiotic prophylaxis.

Rheumatic Valvular Heart Diseases

Almost all acquired heart diseases are rheumatic.

- **Mitral valve involvement** occurs in **75%** and **aortic valve involvement** in about **25%** of all cases of rheumatic heart diseases.
- Stenosis and regurgitation of the same valve may occur **together**, **isolated aortic stenosis is almost always never rheumatic.**
- Tricuspid valve involvement is very rare , that of the pulmonary valve almost never occurs.



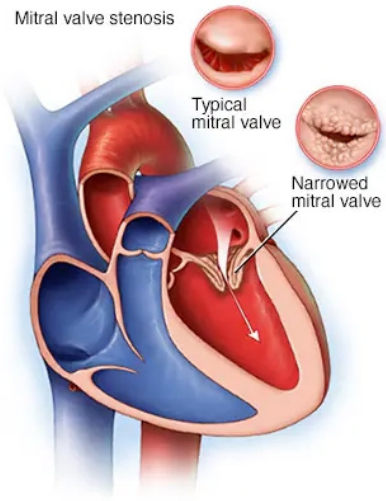
Mitral Regurgitation: Thickened leaflets began to retract towards the annulus, limiting coaptation

Mitral Stenosis: Valve thickening limits normal opening

Other Valves: The aortic valve is the second most commonly affected valve, most often with a stenotic lesion. The tricuspid valve can also be affected

1. Mitral Stenosis

It requires 5 to 10 years from the initial attack of ARF for mitral stenosis to develop.



Pathology: Mitral stenosis results from fibrosis of the mitral valve, commissural adhesions, and contracture of valvular leaflets, chordea and papillary muscle.

Symptoms

- **Mild cases** are asymptomatic.
- **Moderate to severe cases** show:
  - **Manifestaions of pulmonary congestion:** cough expectoration, dyspnea, orthopnea, paraxosymal nocturnal dyspnea.
  - **Manifestations of heart failure.**

Examination

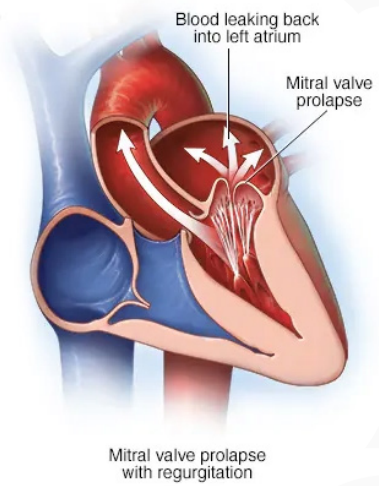
Palpation: diastolic thrill at apex.

Auscultation:

- **Narrow splitting of S2** with **accentuated 2nd heart sound if pulmonary hypertension develops.**
- Opening snap followed by **low frequency mid diastolic rumble** is audible at the apex.
- Occasionally a high frequency diastolic murmur of pulmonary regurgitation (Garham Steel murmur).

2. Mitral Regurgitation

Mitral valve regurgitation is the most common valvular involvement in children in rheumatic heart disease.



Symptoms

- **Moderate to severe cases** show:
- **Manifestations of pulmonary congestion**, cough expectoration, dyspnea, orthopnea, paraxosymal nocturnal dyspnea.
- **Recurrent chest infections** and **palpitations.**

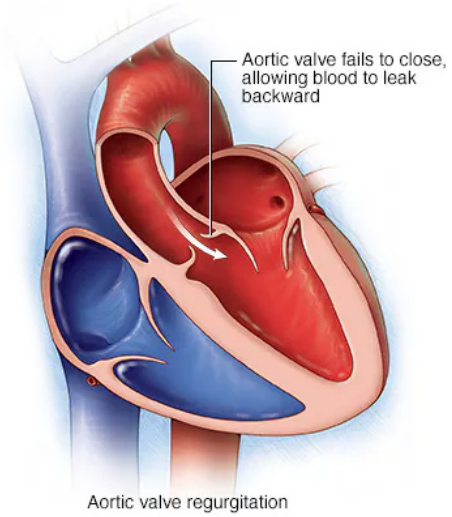
Examination

Palpation: Hyperdyanmic apex, an apical systolic thrill might be found.

Auscultation:

- S1 is diminished, S2 becomes loud after the development of pulmonary hypertension.
- Grade 2-4/6 **pan systolic murmur** is present at the apex radiating to the axilla.
- A short low frequency diastolic flow rumble is present at the apex.

3. Aortic Regugitation



Aortic valve regurgitation

Less common than MR, most patients have associated mitral valve disease

Symptoms

Mild AR: asymptomatic.

Severe AR:

- Exercise intolerance or CHF may occur with.
- Palpitations
- Manifestations of left sided heart failure
- Manifestations of pulmonary congestion: cough, expectoration, dyspnea, orthopnea and paraxosymal nocturnal dyspnea.
- **Chest pain (angina)** due to myocardial insufficiency as the coronary arteries fill during the diastole.

Examination

Auscultation: A **high pitched early diastolic decresendo murmur** best heard at the second aortic area (while the patient is leaning forward) is heard

Blood pressure shows: Wide pulse pressure → High systolic, very low diastolic.

Peripheral signs:

- **Collapsing pulse (Water-hammer pulse)** → best felt on brachial artery.
- **Corrigan's sign** → Marked arterial pulsations in the neck.
- **Pistol shot** → best heard on femoral artery.
- **Capillary pulsations** → detected by pressing the nail tip

Investigations

- X-RAY chest and heart
- ECG: Left ventricular hypertrophy
- Echocardiography
- Cardiac cathetariztaion and angiography

Treatment

Medical treatment:

- Primary prevention of ARF by giving 10 days of oral penicillins.
- Secondary prevention of RH by giving long acting penicillins
- Good oral hygiene and prophylaxis against bacterial endocarditis.
- Treatment of heart failure if associated.
- After load reducing agents are useful in maintaining forward strok volume

Surgical:

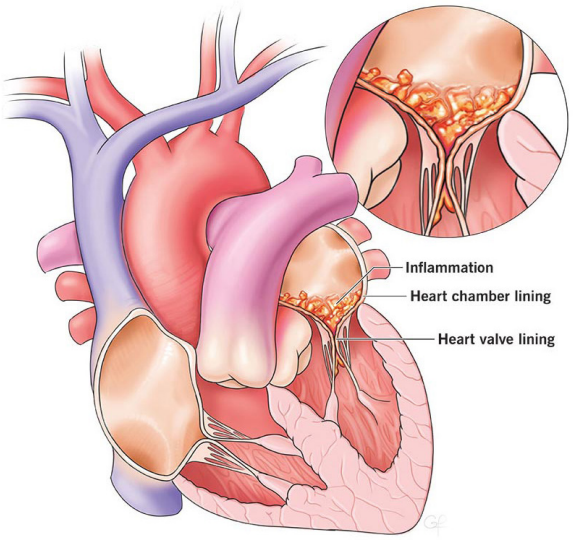
- Valvular repair and prosthetic valve replacement in severe cases
- Anticoagulant must be maintained

TTT of Rheumatic valvular lesions (very important)

- Prophylaxis against streptococcal infection (by long acting penicillin).
- Prophylaxis against infective endocarditis

5 INFECTIVE ENDOCARDITIS

IE is an infection of the endocardium and/or heart valves that involves thrombus formation (vegetation), which may lead to their damage.



Etiology

- IE is caused by bacteria in most cases that enter the bloodstream and settle in the heart lining, heart valves or blood vessels. Other organisms may be encountered.
- IE is uncommon, but people with some heart conditions have a greater risk of developing it.

Most common causative organisms:

- **Streptococcus viridans** (50% of cases) → after **dental procedures.**
- ***Streptococcus fecalis*** (enterococcus) → after **lower GIT or urinary procedures.**
- **Staphylococcus aureus** (***may occur in normal heart***, and is acute in course).

Other less common organisms include: Pneumococcus, Hemophilus sp. Staph. Albus, Neisseria, anaerobes, Clamydia, Fungi, etc.

Pathogenesis

Features of bacterial endocarditis are due to bacteremia, local cardiac invasion by organisms, peripheral embolization, and the formation of immune complexes.

Blood turbulence caused by high velocity flow through a stenotic or incompetent valve or an abnormal communication between systemic and pulmonary circulations damages or denudes the endothelium, to which platelets and fibrin can adhere, and a small, sterile nonbacterial thrombotic endocardial lesion forms.

Traumatized endocardium or valvular endothelium due to indwelling catheters to which circulating bacteria and inflammatory cells adhere and grow in thrombi, forming an infected vegetation. Once formed, the constant blood flow may result in embolization to any body organ. A brisk immunologic response is produced.

Narcotic drugs abusers, immunocompromized patients, and post cardiac surgery patients may be affected in absence of a predisposing cardiac lesion

Culture-negative endocarditis

Culture-negative endocarditis occurs when a patient has typical clinical or echocardiographic findings of endocarditis, with persistently negative blood cultures.

Common causes include recent antibiotic therapy, or infection caused by a fastidious organism that grows poorly in vitro.

Clinical Picture

Is attributed to Toxaemia, Embolization, Circulating immune complexes and Cardiac damage.

Toxaemia

Insidious onset of fever, anemia, and toxic earthy look, in a patient with Rheumatic or CHD (+/-History of a minor surgical procedure)

Fever:

- is persistent, usually low grade, but high spiky fever may be noticed especially with acute endocarditis.
- It may persist for months before diagnosis is established.

Toxaemic manifestations may occur: fatigue, anorexia, arthralgia, weight loss, and headache and at times, chills, nausea, and vomiting.

Embolic Manifestations

Renal infarction: Pain + tenderness in renal angle + hematuria.

Pulmonary emboli occur in Rt. sided vegetations, while **systemic emboli** occur in Lt. sided vegetations

Neurologic manifestations:

- Cerebral embolism, and brain abscess due infected emboli.
- Cerebral Hge or subarachnoid Hge due to rupture of mycotic aneurysms.

Cardiac damage

- Change of character of an existing murmurs or appearance of new murmurs.
- Purulent pericarditis due to rupture of myocardial abscess
- Heart failure.

Circulating immune complexes

Soft tender Splenomegaly.

Pale finger clubbing: occurs after 5-6 wks

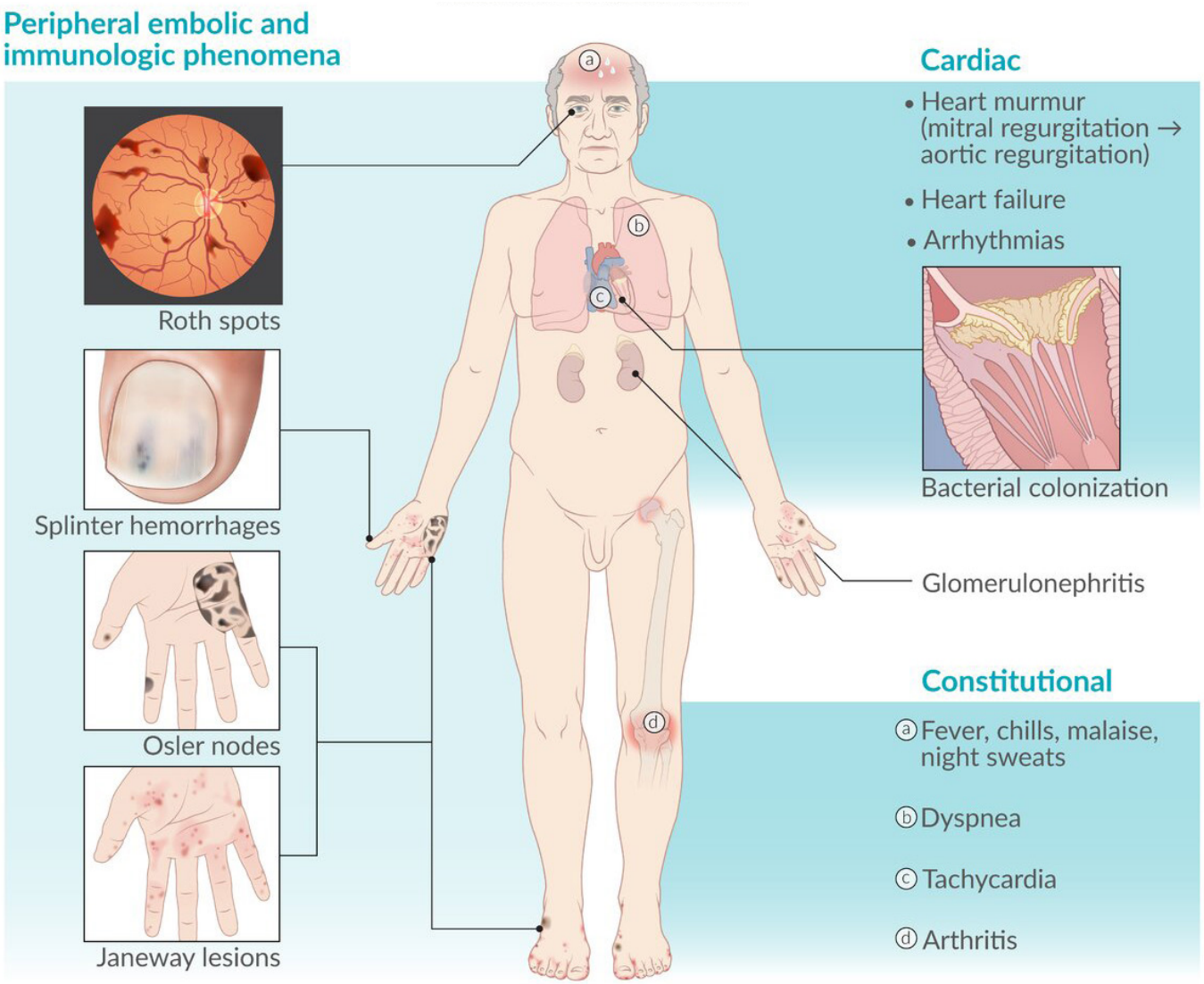
Hematuria + picture similar to acute glomerulo-nephritis.

Renal failure in advanced cases.

Skin Manifestations:

- **Osler nodules:** tender intra-cutaneous purple nodules, in pulp of fingers, toes.
- **Janaway lesions:** painless erythematous or hemorrhagic lesions on palms and soles.
- **Splinter Hges:** streaky hemorrhages under the nails.

Fundus: may show **Roth spots** (small pale spots surrounded by hemorrhagic area).



Complications

- **Systemic and pulmonary embolization**
- **Acute heart failure** due to valve destruction or distortion and/or rupture of the chordae tendineae.
- **Chronic heart failure** due to progressive valvular insufficiency with worsening ventricular function.
- **Vasculitis** may result from circulating immune complexes that may deposit on various endothelial surfaces.
- **Renal involvement** leading to renal failure.

Without treatment, the disease is lethal.

Investigations

Infective endocarditis must be suspected in any patient with a known cardiac lesion who develops an unexplained fever.

Blood culture

- For microbiologic documentation, obtaining 5-7 mL of blood from children (1-3 mL in infants) in **3 separate samplings within 1 - 24 hours** is recommended, according to the clinical presentation.
- Done under **complete aseptic conditions**, for aerobic & anaerobic bacteria, and fungi.
- **Antibiotic sensitivity** must be tested for +ve cases. Antimicrobial pretreatment of the patient reduces the yield of blood cultures to only 50–60%.
- Culture may be -ve in about 10% of cases. Therefore, antibiotic TTT is ***started based on clinical diagnosis.***

CBC: Anemia, associated with leukocytosis and ↑↑ ESR, and CRP.

Urine examination: hematuria microscopic or macroscopic with RBCs casts..

Transthorathic echo: can show vegetations as small as 2 mm in diameter.

Diagnosis is based on the "Modified Duke Criteria"

Using the modified Duke Criteria, the diagnosis of endocarditis is based on pathologic or clinical findings

Pathologic criteria

Pathologic criteria for definite infectious endocarditis include:

- Microorganisms on cultures of a vegetation
- Histology in a vegetation
- Histologic confirmation of active disease in a vegetation or intracardiac abscess.

Clinical criteria

Major Criteria	Minor Criteria
Positive blood cultures: **2 separate cultures for a typical endocarditis microorganism:** • Streptococcus viridans. • a HACEK organism*. **Persistently positive blood cultures** **Coxiella organism and/or Q fever.** **Positive echocardiographic findings:** • Oscillating mass and/or vegetation. • Paravalvular abscess. • Dehiscence of a prosthetic valve. **New valvular regurgitation.**	**Predisposition:** history of **IV drug use** or **congenital heart disease.** **Fever** with a temperature of **more than 38°C**. **Vascular phenomena:** Arterial emboli, septic pulmonary infarcts, intracranial hemorrhage, conjunctival hemorrhage, Janeway lesions. **Immunologic phenomena:** Glomerulo-nephritis, Osler nodes, Roth spots, a positive result for rheumatoid factor) **Positive blood culture findings:** without meeting the criteria above or serologic evidence of active infection consistent with endocarditis.

***HACEK ORGANISMS:** Haemophilus parainfluenzae, H aphrophilus, H paraphrophilus, Actinobacillus actinomycetemcomitans, Cardiobacterium hominis, Eikenella corrodens, or Kingella species.

Diagnosis

2 major criteria **OR** (**1 major criterion** **AND** **3 minor criteria**) **OR** **5 minor criteria**

Differential Diagnosis

- Rheumatic activity
- Collagen-Vascular Disease
- Heart Failure
- Myocarditis
- Vasculitis and Thrombophlebitis
- Other causes of fever in patients with previous heart disease.

Management

Prevention

Prophylactic antibiotic cover before procedures that may cause bacteremia for patients with rheumatic or CHD in the form of:

- **Oral amoxicillin** 50 mg/kg is recommended, 1 hr before procedure, and 25 mg/kg 6 hours after the initial dose.
- **If penicillin allergy**, oral erythromycin 20 mg/kg 2 hours before procedure, and 10 mg/kg 6 hours after the initial dose.

For GIT and urinary tract procedures:

- **Parenteral Ampicillin** 50 mg/kg + **Gentamicin** 2 mg/kg are given ½ hr before procedure.
- **If penicillin allergy**, Vancomycin is used instead of ampicillin in a dose of 20 mg/kg slow IV infusion 1 hour before procedure + Gentamicin

Early TTT for upper respiratory infections and septic foci, to prevent bacteremia.

Dental care and oral hygiene.

Treatment

Mortality is high (20-25%) even with antibiotic TTT

1. Antibiotic TTT

- For at least 4-6 wks should be instituted immediately once a definitive diagnosis is made. The choice is determined according to culture & sensitivity.

The initial TTT of choice:

- Usually **IV crystalline penicillin + Gentamicin.** This combination is active against Streptococcus viridans & Strept. Fecalis.
- IV Crystalline penicillin 200,000 IU/kg/d, in 6 divided doses, for 4-6 wks.
- IV Gentamicin 3-5 mg/kg/d, in 3-divided doses for 2 weeks.

If Staph. is suspected:

A penicillinase resistant antibiotic must be used instead of penicillin as:

- Nafcillin, methicillin, oxacillin, or **vancomycin** for 6 weeks.

If on prosthetic tissue:

- Is treated with nafcillin or oxacillin plus rifampin for at least 6 weeks, in combination with gentamicin for 2 weeks.

Methicillin-resistant S aureus (MRSA) infection:

On native valves:

- Is treated with vancomycin for at least 6 weeks, with or without 3-5 days of gentamicin.

On prosthetic tissue:

- Is treated with vancomycin plus rifampin for at least 6 weeks, in combination with gentamicin for 2 weeks.

2. TTT of heart failure

3. Surgical TTT

Done in:

- Severe aortic or mitral valve involvement with intractable HF.
- Myocardial abscess
- Recurrent emboli
- Increasing vegetations size while receiving therapy.

Done by:

- Removal of vegetations
- Sometimes, valve replacement may be lifesaving

Sustained antibiotic administration will most often prevent re-infection

6 HEART FAILURE

Failure of the heart to produce the cardiac output required to sustain the metabolic needs of the body without evoking compensatory mechanisms (cardiac reserve).

Congestive heart failure refers to:

Clinical syndrome characterized by:

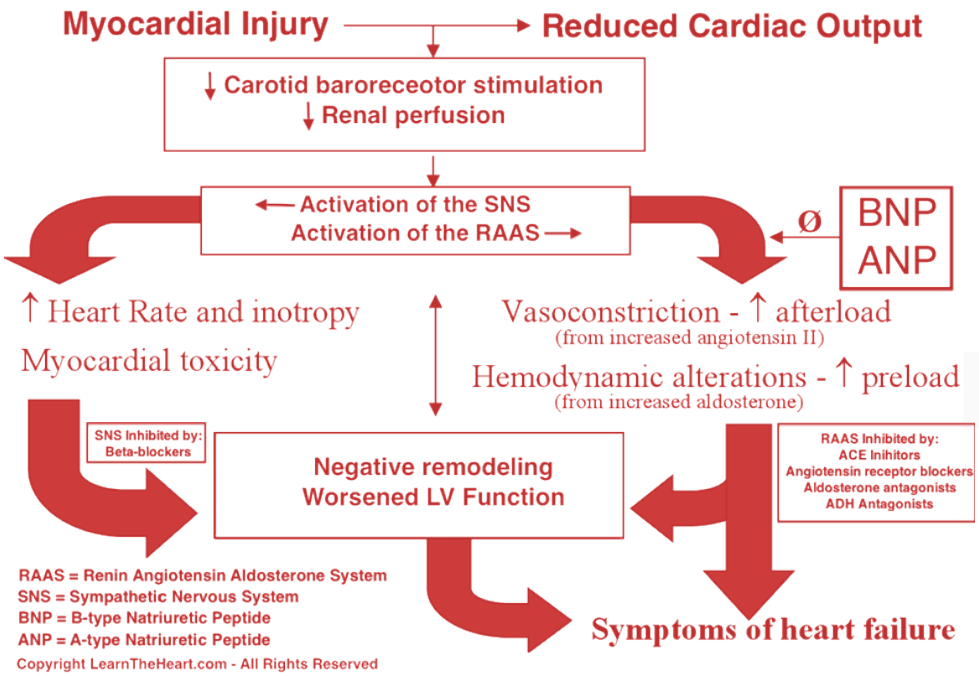
- **Symptoms:** of dyspnea, reduced exercise tolerance, orthopnea.
- **Signs:** such as tachycardia, rales, gallop rhythm and edema.

The disease is a multi-system response to an initial reduction in ventricular function.

Pathophysiology

The heart is viewed as a pump whose output is directly proportional to its filling volume.

COP = SV x HR



- **Failure due to increase preload** → Lt to RT shunts, valvular lesions.
- **Failure due to severe afterload** → PS,AS and Ao Coarctation.
- **High output failure** → AV fistulas, thyrotoxicosis and anaemia

Pathogenesis of major findings

Ankle, sacral edema

- **"Backward failure"** of **right ventricle** → increased venous pressure → fluid transudation.

Hepatomegaly

- **Increased venous pressure** → increased resistance to portal flow.

Pulmonary congestion

- **"Backward failure"** of **left ventricle** → increased pulmonary venous pressure → pulmonary venous distention and transudation of fluid into air spaces.

Dyspnea on exertion

- **Failure of left ventricular output to rise** during exercise → in creased pulmonary venous pressure.

Paroxysmal dyspnea & pulmonary edema

- Probably **sudden failure of left heart output** to keep up with right heart output → acute rise in pulmonary venous and capillary pressure → transudation of fluid into air spaces.

Orthopnea

- Normal pooling of blood in lungs in supine position added to already congested pulmonary vascular system (**increased venous return** not putout by left ventricle).
- Relieved by sitting up, raising head of bed, lying on extra pillows.

Weakness & exercise intolerance

- **"Forward failure"** of **left ventricle**
- Cardiac output inadequate to perfuse muscles; especially, failure of output to rise with exercise.

Cardiac dilation

- Greater ventricular end-diastolic volume

Causes of heart failure

Age	Causes	Diagnostic features
Fetal	• Severe anemia (hemolytic, fetal-maternal transfusion, parvovirus, B19-induced An.) • Supraventricular tachycardia. • Ventricular tachycardia. • Complete heart block.	**History of** • Maternal fever • Hemorrhage **Fetal echo**
Premature	• Fluid over load. • PDA. • VSD. • Cor pulmonal (bronchopulmonary dysplasia). • Hypertension	**History of** • Neonatal intensive care admission • Prolonged administration of oxygen **Presence of:** murmur
Full term	• Asphyxial cardiomyopathy. • **Arteriovenous malformation:** vein of galen, hepatic. • **Left-sided obstructive lesions:** coarction of aorta, hypoplastic left heart. • **Large mixing cardiac defects:** single ventricle, truncus arteriosus. • Viral myocarditis.	**History of** Prolonged resuscitation **Presence of:** • Cardiac murmur • Associated pneumonia
Infants and toddlers	• Left to right shunts (vsd). • Hemangioma (AV malformation). • Anomalous left coronary artery. • Metabolic cardiomyopathy. • Acute hypertension (hemolytic - uremic $). • Supraventricular tachycardia. • Kawasaki disease.	**Presence of:** • Cardiac murmur • Tachycardia • Associated hepato-megaly • Other manifestations of Kawasaki disease
Child - adolescent	• Rheumatic fever. • Acute hypertension (glomerulonephritis). • Viral myocarditis. • Thyrotoxicosis. • Hemochromatosis-hemosiderosis. • Cancer therapy (radiation, adramycin). • Sickle cell anemia. • Endocarditis • Corpulmonale (cystic fibrosis). • Cardiomyopathy (hypertrophic, dilated, postviral)	**History of** recurrent tonsilitis **Presence of:** • Hematuria • Hand tremors • Tachycardia • Miliary shadow in chest x-ray • Fever • Recurrent chest infections

Diagnosis of heart failure

History

Careful history taking including family history

Signs and symptoms of heart failure

Infancy	Children & Adolescednts
1. Decrease feeding-dyspnea on suckling. 2. Perspires profusely and irritability. 3. Intermittent sleep + poor weight gain. 4. Weak cry-noisy resp-chest retraction. 5. **O/E:** Hepatomegly and +/- Edema.	1. Rapid fatigue –anorexia. 2. Abdominal pain & cough. 3. Dyspnea-orthopnea-PND. 4. **O/E:** underweight, congested pulsating neck veins and edema.

Examination

General exam

Laying stress on:

1. **Blood pressure**
2. **Pulse:** rate, character, volume
3. **Edema:**
   - Eyelids in neonates
   - Lower limbs in ambulatory child
   - Sacrum in bed ridden child
4. **Abdominal exam:**
   - Hepato-spleno megaly
   - Ascites.

Auscultation

Chest:

1. Pulmonary rales,
2. Hydrothorax: bilateral or unilateral.
3. Ronchi and wheezes.

Heart:

1. Murmurs of the etiology
2. Tachycardia
3. Gallop rhythm.

Investigation

1. Chest X-ray
2. ECG
3. Echo
4. Radionuclid studies using (Tcm99).
5. Blood gases
6. Serum electrolytes.

Treatment

The main stay of therapy is to detect and treat the underlying cause if possible and give appropriate inotropic drugs

1. General measures: hospitalization and bed rest in severe cases.

2. Drug therapy: is aimed to improve cardiac contractility, treating the symptoms.

3. Treatment of the underlying causes

Drugs used

Digoxin: +ve inotropic action improves myocardial contractility.

Precautions:

- ECG monitoring prior to each digitalizing dose
- Base line serum electrolytes prior & after digitalis.
- If already taking digoxin use maintenance dose.

Digoxin doses:

- **Premature** = 0.02 - 0.025 mg/kg
- **Neonates** = 0.03 - 0.04 mg/kg.
- **Infant or child** = 0.04 - 0.05 mg/kg
- **Digitalizing dose** =75% of PO doses.

Rapid digitalization:

- **In CHF:** IV 0.5 (immediate) + 0.25 (12 hrs later) + 0.25 (12 hrs later).
- Maintenance dose 12 hrs later.

When to measure serum digoxin level?

- When standard dose is not having beneficial effects.
- When unknown amount of digoxin has been ingested.
- When renal function is impaired.
- When toxicity is suspected.

Dopamine & Dobutamine: B-agonists used mainly in patients with low C.O.P.

Dopamine:

- **Dose:** 5-15 ug/kg/min
- Increases myocardial contractility & has selective renal vasodilator action.
- Dopamine in high doses (above 15 ug/kg/min) results in vasoconstriction.

Dobutamine:

- A derivative of dopamine with the advantage over dopamine.
  - Direct inotropic action.
  - Decreases peripheral vascular resistance
- **Dose:** 0.5-1 mcg/kg/min IV continuous infusion initially, then 2-20 mcg/kg/min; not to exceed 40 mcg/kg/min

Milrinone: is a bi-pyridine positive inotrope and vasodilator with little chronotropic activity.

- It differs in mode of action from both digitalis glycosides and catecholamines.
- This agent is used for the **short-term management of acute decompensated heart failure.**
- **Dose:** 50 mcg/kg loading dose by IV push over 10-60 minutes, then 0.25-0.75 mcg/kg/min IV Monitor electrolytes, renal function, blood pressure

Diuretics

Frusemide (loop diuretic):

Infants and children:

- 1-2 mg/kg IV/IM/PO once initially; increased by 1-2 mg/kg q6-8hr (PO) or 1 mg/kg q2hr (IV/IM)
- Individual dose not to exceed 6 mg/kg

Neonates (<28 days):

- 0.5-1 mg/kg IV/IM q8-24hr
- Individual dose not to exceed 2 mg/kg

Spironolactone: (aldosterone antagonist)

- 1-3.3 mg/kg/day PO or divided q12hr; not to exceed 3.3 mg/kg/day or up to 100 mg/day

Captopril: ACE inhibitor, causing arterial and venous dilatation production

Doses:

Neonates: 0.05-0.1 mg/kg/dose q8-24hr, titrate dose up to 0.5 mg/kg/dose q6-24hr

Infants: 0.15-0.3 mg/kg/dose; titrate dose upward to maximum 6 mg/kg/day in 1-4 divided doses; 2.5-6 mg/kg/day usually required.

Children: 0.3-0.5 mg/kg/dose; titrate to maximum 6 mg/kg/day divided q6-12hr

Sodium Nitroprusside: Directly dilates arterial & venous vessels **(CI in hypotension).**

- Given IV **only in ICU.**
- Given to **critically ill patients & for a short period of time** as it may **exacerbate myocardial ischemia** by increasing heart rate.

Steps in evaluation of ventricular dysfunction

1. Make the diagnosis of heart failure or asymptomatic ventricular dysfunction.
2. Determine the type and severity of cardiac dysfunction.
3. Determine the patient's prognosis.
4. Provide treatment and follow-up.
5. Recommendations.

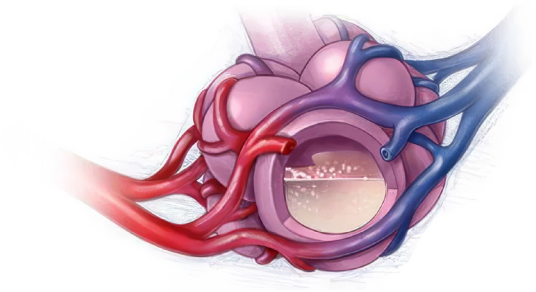
Pulmonary edema

It is not a diagnosis, it is a finding due to cardiac or non cardiac causes.

Cardiogenic causes

Left ventricular failure complicating:

- Valvular heart diseases
- Dilated cardiomyopathy
- Tight mitral stenosis
- Ischemic heart disease
- Rarely, left atrial myxoma.



Non cardiogenic causes

Adult respiratory distress $

- Altered alveolar capillary membrane permeability,
- Caused by pn. Toxins, allergens, smoke inhalation ,gastric aspiration, radiation pnemonitis.

Others: drugs, narcotics overdose, severe hypoalbumina ,uremia.

Treatment

Oxygen

Morphia

Frusemide: 16 mg/kg. increase dose if blood pressure is stable.

Dopamine: If hypotesive. 3 - 10 ug/kg/min

Aminophyllin:

- Loading dose 2 5 mg/kg, slowly over 20 to 30 min
- Continuous infusion by 0.5mg/kg.

Mechanical ventilation: If **pco2 > 50mmHg** & **po2 < 50mmHg.**

Positive end expiratory pressure (PEEP): Reduces cardiac output and hypotension.

Antibiotics

CARDIOMYOPATHIES

Diseases of the myocardium associated with cardiac dysfunction.

They are classified as:

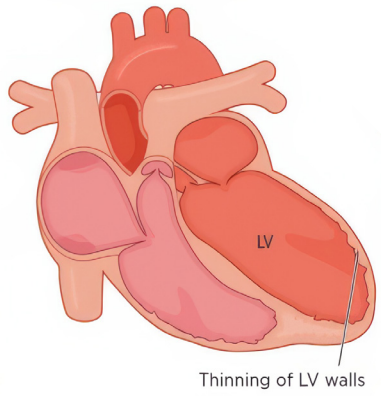
1. Dilated cardiomyopathy
2. Hypertrophiccardiomyopathy
3. Restrictive cardiomyopathy
4. Arrhythmogenic right ventricular cardiomyopathy.
5. Unclassified cardiomyopathies

Investigations

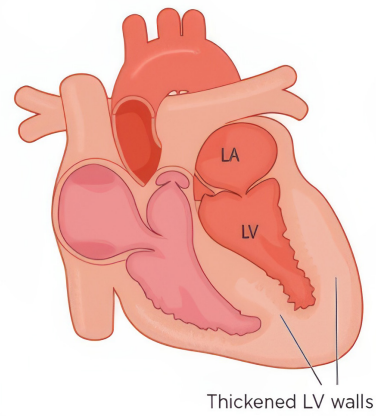
- Chest X-ray
- ECG
- Echo **(diagnostic)**
- Cardiac MRI

The common types of cardiomyopathy seen in children include:

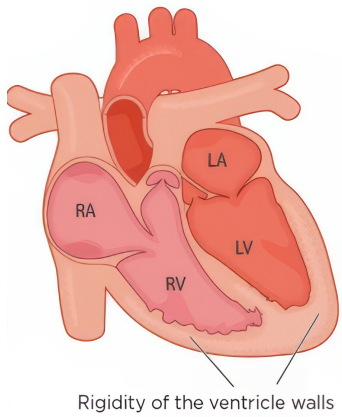
Dilated Cardiomyopathy



Hypertrophic Cardiomyopathy



Restrictive Cardiomyopathy



Dilated (congestive) cardiomyopathy

Dilated cardiomyopathy is the most common form of cardiomyopathy

Causes

- Infectious, toxic, or metabolic agents & immunologic defects.
- **Idiopathic: most common (60%)** followed by, familial cardiomyopathy, active myocarditis, and other causes.
- Hyper and hypothyroidism, excessive catecholamines, nutritional disorders (e.g. kwashiokor, beriberi & carnitine deficiency).
- Cardiotoxic drugs (doxirubicin).

Endocardial fibro elastosis and doxorubicin cardiomyopathy have clinical features similar to those of dilated cardiomyopathy

Pathology and pathophysiology

- In dilated cardiomyopathy, a weakening of systolic contraction is associated with dilatation of all four cardiac chambers.
- Interacavitary thrombus formation is common in the apical portion of the ventricular cavities and in atrial appendages.
- It may give rise to pulmonary and systemic embolization.

Clinical manifestations

Symptoms: Fatigue, weakness, and symptoms of left heart failure

Signs: Signs of CHF (e.g. tachycardia, pulmonary crackles, weak pulses, distended neck veins, hepatomegaly) may be present.

Auscultation:

- A prominent S3 with or without gallop rhythm is present
- A soft systolic murmur of MR or TR may be audible

Investigations

ECG: Sinus tachycardia , LVH , and ST T changes are common ECG findings

CXR films show generalized cardiomegaly , often with signs of pulmonary venous congestion

Echo: Diagnostic.

- The LV and RV are dilated with ↓ fractional shortening (FS) and ejection fraction (EF).
- Intracavitary thrombus and pericardial effusion may be present.

Doppler: The mitral inflow Doppler tracing demonstrates a reduced E velocity and a decreased E/A ratio.

Progressive deterioration is the rule rather than the exception; About two thirds of these patients die within years of the onset of symptoms due to arrhythmias, systemic or pulmonary embolization, or CHF.

Management

CHF is treated: Digoxin, diuretics, vasodilators (captopril, enalapril, hydralazine) bed rest, and restriction of activity.

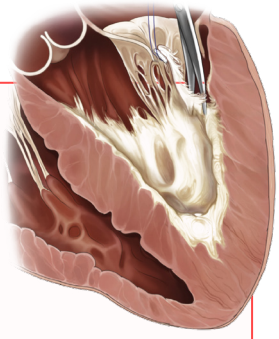
Anticoagulation: (Coumadin or heparin) is recommended because of the frequency of embolization.

Arrhythmias: Amiodarone, other antiarrhythmic agents, or a pacemaker.

Beneficial effects of B adrenergic blockers (somewhat heretical given poor LV contractility) are under investigation.

Growth hormone (under trial in adults): It may increase LV wall thickness, reduce of chamber size, and improve cardiac output.

Many of these children may need cardiac transplantation.



ENDOCARDIAL FIBROELASTOSIS

Endocardial fibro elastosis (EFE) is a form of dilated cardiomyopathy of unknown origin. seen in infants and children.

Viral agents, especially mumps, have been implicated in the past and again recently in some cases of EFE.

- **The condition is characterized by:**
  - Diffuse changes in the endocardium, with a white, opaque, glistening appearance.
  - The left side of the heart is dilated and hypertrophied, with poor contractility.
- **Clinical manifestations:**
  - Symptoms and signs of CHF develop in the first 10 months of life
  - By auscultation, No heart murmur is audible in most patients occasionally a heart murmur of MR is audible , gallop rhythm , hepatomegaly is usually present
- **Investigations:**
  - CXR: marked cardiomegaly with normal PVMs or pul. venous congestion patterns.
  - ECG: LVH with strain, occasionally , myocardial infarction
- **Treatment:**
  - Early and long term (years) treatment with Digoxin, Diuretics and after load-reducing agents is recommended.

DOXORUBICIN CARDIOMYOPATHY

Doxorubicin cardiomyopathy is becoming the most common cause of chronic CHF in children

It occurs in up to 50 % of patients who have received more than 1 g/m 2 of doxorubicin

- **Risk factors include:**
  1. Age younger than 4 years
  2. The cumulative dose exceeding 400 to 600 mg/m^2
  3. A regimen of larger, infrequent doses.
- **Clinical manifestations:**
  - Patients are usually asymptomatic until signs of CHF develops.
  - Symptoms may develop 2 to 4 months, and rarely years, after completion of doxorubicin therapy.
  - History of exertional dyspnea, palpitation, cough, or substernal discomfort may be present.
  - Signs of CHF develop with hepatomegaly and distended neck veins.
  - Gallop rhythm may be audible with occasional soft systolic murmur of MR or TR
- **Investigations:**
  - CXR: cardiomegaly with or without pulmonary congestion or pleural effusion.
  - ECG sinus tachycardia with rare ST-T changes
  - Echo: dilated LV with decreased contractility.
- **Treatment:**
  - Anticongestive measures with inotropic agents (digoxin), diuretics, and after load reducing agents (captopril) are useful.
  - Doxorubicin administered as a continuous infusion can reduce cardiac injury.
  - The advisability of the modification of anthracycline therapy is controversial .
  - Beta blockers (metoprolol , 0.1 mg/kg/dose twice daily, increasing to a maximal dose of 0.9 mg/kg/day) have been shown to be beneficial in some patients with chemotherapy induced cardiomyopathy, similar to what has been reported in adults.
  - Cardiac transplantation may be an option for selected patients.
- **Prevention:**
  - The cardiology committee of children's cancer study group has recommended close monitoring for cardiac toxicity by: Echo, radionuclide angiography, and endomyocardial biopsy
  - If these tests show abnormalities of LV systolic function, either withholding anthracycline therapy or limiting the total cumulative dose to 400 to 500 mg/m^2

Hypertrophic cardiomyopathy

Heterogeneous cardiomyopathy, usually familial disorder of heart muscle about 30% to 60% of cases.

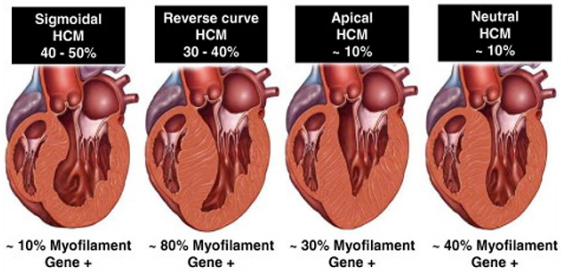
- Massive ventricular hypertrophy with an enhanced ventricular contractility.
- Ventricular filling is impaired due to relaxation abnormalities.

Inheritance:

- Cardiomyopathy appears to be genetically transmitted, as an autosomal dominant trait
- It may occur sporadically.
- It may be seen in children with **LEOPARD** syndrome.

Pathology and pathophysiology

- Massive ventricular hypertrophy is present.
- **Asymmetric septal hypertrophy (ASH)**, formerly known as idiopathic hypertrophic subaortic stenosis (IHSS), is the **most common type**.
- Aconcentric hypertrophy with symmetric thickening of the LV sometimes occurs
- **Interacavitary obstruction** may develop during systole partly because of systolic anterior motion (SAM) of the mitral valve against the hypertrophied septum, called ***hypertrophic obstructive cardiomyopathy (HOCM).***



Clinical manifestations

30 % to 60 % of cases are seen in adolescents and young adults with positive family history.

Symptoms: Easy fatigability , dyspnea & palpitation.

Signs: A sharp upstroke of the arterial pulse is characteristic.

Auscultation: A late systolic ejection murmur of medium pitch, best audible at the middle and lower LSB or at the apex.

Investigations

ECG: may show LVH, ST-T changes, abnormally deep Q waves with diminished or absent R.

CXR: Mild LV enlargement

Echo: Septal hypertrophy and/or LV free wall.

Doppler: Mitral inflow demonstrates a decreased E velocity ,increase A velocity& a decreased E/A ratio.

Management

Moderate restriction of physical activity.

Drugs:

- **Beta adrenergic blockers** as, propranolol, atenolol, or metoprolol, acts by **reducing the obstruction & have antiarrythemic action.**
- **Calcium channel blockers**, principally verapamil , may be equally effective

Dual chamber pacing was shown to reduce the pressure gradient.

Digitalis and other cardiotonic and vasodilators are contraindicated

Transaortic left ventricular septal myotomy may be usefull in selected patients.

Restrictive cardiomyopathy

Denotes a restriction of diastolic filling of the ventricles (usually infiltrative)

- ↓ Contractile function of the ventricle.
- There is a marked dilatation of both atria.
- It is the least common of the three types of cardiomyopathy.

It is characterized by

- An **abnormal diastolic ventricular filling** owing to excessively stiff ventricular walls.
- The **ventricles remain normal in size** and maintain normal contractility, but the **atria are enlarged** out of proportion to the ventricles.

Causes:

- Often caused by **infiltrative disease processes** (e.g., sarcoidosis , amyloidosis)
- **Endomyocardial biopsy** may be useful in identifying causes of restrictive cardiomyopathies (e.g., amyloidosis, hemochromatosis, glycogen deposit)

Clinical manifestations

Symptoms: History of exercise intolerance, weakness and dyspnea, or chest pain.

Signs: Jugular venous distention.

Auscultation:

- Gallop rhythm
- Systolic murmur of MR or TR.

Investigations

ECG: may show atrial fibrillation and paroxysms of SVT.

CXR: show cardiomegaly, pulmonary congestion, and pleural effusion.

Echo:

- Biatrial enlargement, with normal cavity size of the LV and RV.
- LV systolic function is normal until the late stages of the disease.
- Atrial thrombus may be present.

Doppler: The mitral inflow Doppler tracing shows an increased E velocity and increased E/A ratio.

Management

Diuretics are beneficial by relieving congestive symptoms.

Digoxin is not indicated, because systolic function is unimpaired.

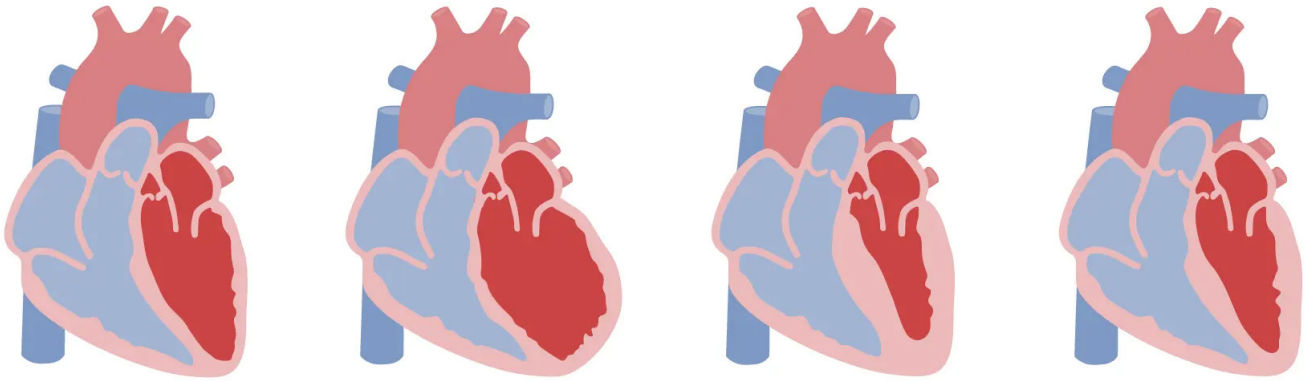
Calcium channel blockers may be used to increase diastolic compliance.

Anticoagulants (warfarin) and antiplatelet drugs (aspirin and dipyridamole) may help prevent thrombosis .

Corticosteroids and immunosuppressive agents have been suggested.

Permanent pacemaker is indicated for **complete heart block**

Cardiac transplantation may be an option.



Normal | Dilated | Hypertrophic | Restrictive

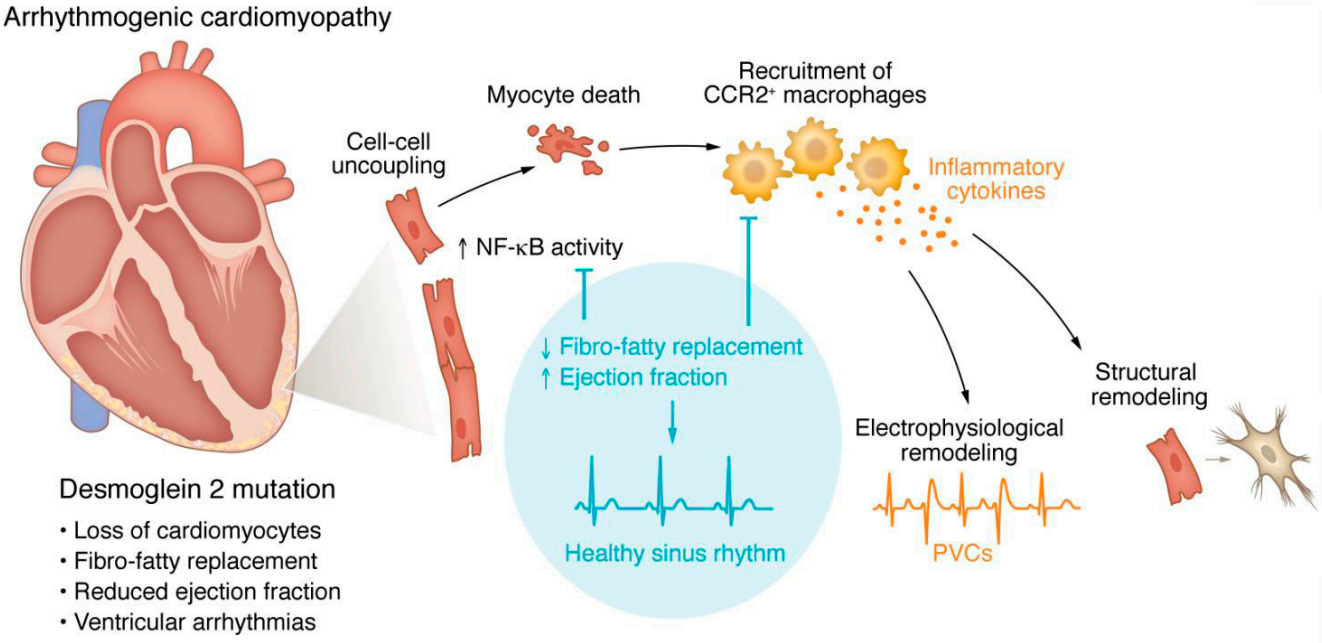
RIGHT VENTRICULAR DYSPLASIA (ARRHYTHMOGENIC CARDIOMYOPATHY)

RV dysplasia, or RV cardiomyopathy, is a rare abnormality of unknown etiology in which the myocardium of the RV is partially or totally replaced by fibrous or adipose tissue.

The LV is usually spared.

Most cases appear to be sporadic. It is prevalent in northern Italy.

- **Clinical manifestations:**
  - **Onset:** in infancy, childhood, or adulthood (but usually before age 20 years)
  - **History:**
    - Palpitation, syncopal episodes , or both.
    - Sudden death due to arrhythmias (VT , SVT arrhythmias)
    - Signs of CHF
- **Investigations:**
  - **CXR:** Usually shows cardiomegaly.
  - **ECG** most often shows tall p waves in lead II (RAH), decreased RV potentials, T wave inversion in the right precordial leads, and PVCs or ventricular tachycardia of LBBB morphology.
  - **Echo:** Selective RV enlargement and often areas of akinesia or dyskinesia.
- **Treatment:**
  - **Various antiarrhythmic agents** may be tried, but they are often unsuccessful in abolishing ventricular tachycardia.
  - **Surgical intervention** (ventricular incision or disconnection of the RV free wall) may be tried if antiarrhythmic therapy is unsuccessful
- **Prognosis:**
  - A substantial potion of patients **die before age 5 years** of CHF and intractable ventricular tachycardia .



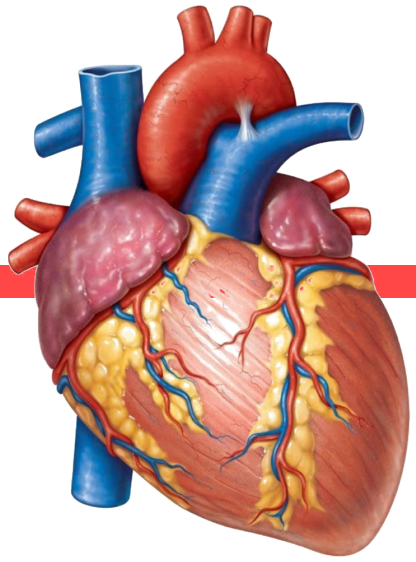
	Dilated Cardiomyopathy	Hypertrophic Cardiomyopathy	Restrictive Cardiomyopathy
Definition	Characterized by **dilatation and impaired contraction** of the left ventricle or both ventricles.	**AD inheritance with variable expression** It is characterized by **left ± right ventricular hypertrophy** and **asymmetrical interventricular septal hypertrophy** Accompanied by **obstruction of the left ventricular outflow tract.**	Characterized by **poor ventricular compliance** and **inadequate restrictive ventricular filling** with **normal or near-normal systolic function** and **wall thickness.**
Common Causes	• **Idiopathic** • **Familial/genetic** • **Viral** (coxackie B & adeno) and/or **immune** mediated. • **Alcoholic/toxic**		• **Idiopathic** • **Associated with other disease** (eg, amyloidosis; endomyocardial disease with or without hypereosinophilia)
Histology	Nonspecific	**Typical morphological changes include:** • Myocyte hypertrophy and disarray • Surrounding areas of increased loose connective tissue	**Increased interstitial fibrosis** may be present
Presentation	Usually **heart failure**, which is often progressive	• Dyspnea, PND • Chest pain, syncope • Palpitations due to arrhythmias • **Premature sudden death** (might be the presenting symptom)	
Treatment	**Antifailure measures:** • Lanoxin • ACEI • Diuretics.	**B-blockers and Ca channel blockers.**	
Complications	**Common and may occur at any stage:** • Intractable heart failure • Arrhythmias • Thromboembolism • Sudden death	**Ventricular tachycardia/fibrillation** is the usual cause of death in hypertrophic cardiomyopathy	Prognosis is **poor usually fatal**

Specific Cardiomyopathies

The term specific cardiomyopathy describes a heart muscle disease that is associated with specific cardiac or systemic disorders.

They include:

1. **Muscular dystrophies:** e.g. Duchenne muscular dystrophy; Becker muscular dystrophy
2. **Neuromuscular disorders:** e.g. Friedreich's ataxia; Noonan's syndrome.
3. **Inflammatory cardiomyopathy:**
   - **Idiopathic**
   - **infectious** especially viral (e.g. Adenovirus; CMV; coxsackie A and B; HIV; Enterovirus)
   - **Autoimmune**
4. **Metabolic cardiomyopathy**
   - **Endocrine:** e.g. Thyrotoxicosis, Hypothyroidism, Pheochromocytoma.
   - **Familial storage disease and infiltrations:** e.g. Hemochromatosis, Glycogen storage disease; Hurler's syndrome; Niemann-Pick disease.
   - **Nutritional disorders and deficiencies:** e.g. Beri-beri, Kwashiorkor, Anemia.
5. **Systemic disease**
   - Connective tissue disorders (e.g. SLE, rheumatoid arthritis)
   - Infiltrations and granulomas (e.g. sarcoidosis, leukemia)
6. **Sensitivity and toxic reactions:** e.g. anthracyclines, irradiation
7. **Valvular cardiomyopathy**
8. **Hypertensive cardiomyopathy**
9. Dilated cardiomyopathy **secondary to tacharrhythmia**
10. **Coronary abnormalities:** congenital or acquired (2ry to Kawasaki)



END.

Questions

1. One of the congenital cyanotic heart diseases:

A. ASD
B. PDA
C. VSD
D. PS
E. Fallot tetralogy

E

2. The organism responsible for the development of rheumatic fever is:

A. Group B streptococcus
B. Pneumococci
C. Staphylococcus aureus
D. Group A B-hemolytic streptococcus
E. None of the above

D

3. Mitral Insufficiency is characterized by all of the following EXCEPT:

A. Apical pansystolic murmur
B. Weak "muffled" second heart sound.
C. Cardiac apex may be shifted down and out
D. Mostly due to rheumatic carditis.

B

4. You are evaluating a 3-month-old boy in your office who you know to have tetralogy of Fallot. His mother informs you that the infant has had fever, diarrhea, and poor feeding in the last 24 hours. On physical examination, you note cyanosis of the extremities and perioral area, tachypnea, hyperpnea, and a heart rate of 180 beats per min. At the last visit you recall a harsh 3/6 systolic ejection murmur; today you do not hear a murmur.

Of the following, the MOST appropriate management strategy is to:

A. Administer antipyretics for fever.
B. Reassure his mother because the murmur is gone.
C. Order echocardiogram to evaluate the pulmonary valve.
D. Encourage oral intake of fluids.
E. Place him in the knee–chest position with oxygen

E

5. A 12 –hour-baby was found on the postnatal ward to be grunting and dusky. Hewas born at term by NVD after uneventful pregnancy. O/E he has increased work of breathing, with retractions, systolic murmur and palpable liver 2 cm below coastal margin.CRT 3 sec and HR 190 bpm. Femoral pulsations well felt. His oxygen saturation 72% which remains low despite oxygen supplementation.

What is the most probable diagnosis?

A. Fallot tetraology.
B. Transposition of great vessels.
C. Coarctation of the aorta.
D. Ventricular septal defect.
E. Esinmenger syndrome.

B

6. You are evaluating a recently adopted 2-year-old from overseas. His new parents were told that he has a murmur. They noticed he loves to run around the house, but he often takes breaks squats down and breathes heavily. He has dark skin, but they wonder if he looks dusky when he runs. On examination, he has room air saturations of 80%, a right ventricular heave, and a harsh systolic ejection murmur at the LUSB that radiates to the lung fields. There is no hepatomegaly. Pulses are equal in all extremities. His upper and lower extremity blood pressures are normal for age.

Which congenital cardiac lesion do you suspect?

A. ASD
B. PDA
C. VSD
D. PS
E. Fallot tetralogy

E

7. Two-month old with tetralogy of Fallot presents to clinic with saturations of 60%. All are immediate management steps you would like to take EXCEPT:

A. Knees to chest
B. Oxygen
C. Digoxin
D. Beta-blocker

C

8. Which of the following is a symptom of rheumatic heart disease?

A. Orthopnea
B. Subcutaneous nodules
C. Polyarthralgia
D. Carditis

A

9. One is not considered as a criterion for the diagnosis of infective endocarditis.

A. Persistently positive blood cultures
B. Hypogammaglobulinemia
C. Echo evidence: positive for IE (oscillating intracardiac mass in path of regurgitant jet or on implanted material, abscess)
D. Leukocytosis and high acute phase reactants

B

10. You are evaluating a patient who presents with orthopnea and peripheral edema. Given his history with multiple untreated group A streptococcal infections, you are concerned about rheumatic heart disease. On exam you note a diastolic decrescendo murmur heard at the upper left sternal border. Which of the following valve lesions do you expect to see on echocardiography?

A. Mitral valve regurgitation
B. Mitral valve stenosis
C. Aortic valve regurgitation
D. Aortic valve stenosis

C

11. Rheumatic heart disease results from of which of the following?

A. Recurrent episodes of infective endocarditis
B. Initial group A streptococcal infection
C. Damage to the heart valves from recurrent episodes of acute rheumatic fever
D. Untreated rheumatic arthritis

C

12. Rheumatic Fever usually follows an infection with:

A. Streptococcus group A
B. Staphylococcus aureus
C. Streptococcus viridans
D. H.Influenza

A

13. One of the tests that is done to confirm diagnosis of R.F:

A. Alpha 1 antitrypsin
B. Rheumatoid factor
C. Creatine kinase
D. Anti-streptolysin O

D

14. Which of the following is one of Jone's major criteria?

A. Elevated ESR
B. Fever
C. Subclinical carditis
D. Previous RF

C

15. Acute and sub-acute endocarditis are caused by which bacteria respectively?

A. S. Pyogenes, S. Aureus
B. S. Viridans, S Aureus
C. S. Aureus, S. viridans
D. S. Pyogenes, S. Viridans

C

16. Which of the following valves is mostly affected in patients with central venous catheters?

A. Aortic
B. Mitral
C. Bicuspid
D. Tricuspid

D

17. Persistent cyanosis in the newborn can be caused by one of the following:

A. TGA
B. VSD
C. Hyperbilirubinemia
D. Coarctation of the aorta
E. Eisenmenger syndrome

A

18. Collapsing pulse (water hammer) is a sign of the following EXCEPT:

A. Tricuspid regurge
B. Large PDA
C. Anemia
D. Hyperthyroidism
E. Aortic regurge

19. The organism responsible for the development of rheumatic fever is:

A. Group B streptococcus
B. Pneumococci
C. Staph aureus
D. Group A B hemolytic streptococcus

D

20. A 3months old girl, presented with breathlessness and excessive sweating on feeding. Her weight is 4.3 kg. O/E her saturation is 96% with precordial thrill and pansystolic murmur at the lower left sternal border. CXR: cardiomegaly with plethoric lungs. The most likely diagnosis:

A. ASD
B. Coarctation of the aorta
C. PDA
D. VSD
E. Fallot tetralogy

C

21. Fallot tetralogy is characterized by all EXCEPT:

A. Infundibular PS
B. RV hypertrophy
C. Aortic overriding
D. Central cyanosis in the first week of life

D

22. Major criteria for diagnosis of rheumatic fever include all of the following EXCEPT:

A. Carditis
B. Arthritis
C. Arthralgia
D. Subcutaneous nodules
E. Chorea

C

23. Cardiac cause of chest pain in children should be suspected in the following conditions EXCEPT:

A. Exertional syncope
B. Family history of sudden death < 35 year old
C. Stitching localized pain to the left hemithorax
D. Hypercoagulable states
E. Presence of connective tissue disorder

C

24. A medical student working in the emergency department sees a female baby, born 2 weeks ago, who is brought in by her anxious mother. The mother tells the student that her baby seems "purple," especially her fingers and toes, and looks extremely blue when crying. On physical examination the sleeping baby has mild cyanosis of the face and trunk, but moderate cyanosis of the extremities. Which of the following is the most common cause of cyanosis within the first few weeks of life?

A. Atrial septal defect
B. Patent ductus arteriosus
C. Tetralogy of Fallot
D. Transposition of the great vessels
E. Ventricular septal defect

CASE 1

A 3 months old baby presents to ER referred due to faltering growth. He was born at term after a normal antenatal course. His birth weight was on the 50th centile and he was entirely bottle fed from birth. He grew along 50th centile for the first 2 months of life but started to drop off this centile over the last month. His parents described him as increasingly sweaty with feeds and becoming fatigued halfway through the bottles. On examination, his pulse rate was 180 beats / min. He has a pansystolic murmur loudest over the lower left sternal border with palpable 3 cm hepatomegaly.

Q1: What is your provisional diagnosis?

A case of congenital acyanotic heart disease, most probably VSD, complicated with heart failure.

Q2: What is the initial management for this patient?

1. Oxygen therapy
2. Anti-failure measures (diuretics , captopril)
3. Added calories
4. Surgical closure at age of 5-6 months

Q3: What are the other complications suspected in this case ?

- Severe pulmonary hypertension up to Eisenmenger syndrome
- Infective endocarditis
- Arrhythmia

CASE 2

A five-months-old baby with a postnatal diagnosis of trisomy 21 (Down syndrome) is brought to the pediatric outpatient department for assessment after referral by the General Practitioner (GP). The parents have arrived in your region approximately 6 months ago from Uganda and speak very limited English and no Arabic . They could not quite understand why the GP referred their baby to your department, but are generally happy with the baby's clinical progress. They also try to explain that the baby had a heart scan at about four weeks of age and that a visiting pediatric cardiologist wrote to them to explain that the baby will require open heart surgery soon. They recall the nurses checking vital signs during the cardiology consultation and recall that the health professionals were happy with all numbers. The baby's health record shows weight and length markedly below average and states that the oxygen saturations were 99% during the first cardiac assessment.

Your clinical examination reveals a happy looking baby with typical features of trisomy 21. Peripheral perfusion and pulses are normal. There is cyanosis without signs of respiratory distress, but the baby is making some upper airway noises which you assume are due to the large tongue while lying supine. The lungs appear clear on auscultation with good bilateral air entry. The first heart sound is normal. The second heart sound appears single and louder than the first. There is a soft systolic murmur near the left sternal edge. The liver edge is palpable approximately 3 cm below the right costal margin. The abdomen is slightly distended but soft with normal bowel sounds. The baby's general muscle tone is slightly reduced and the anterior fontanel feels normal. The babies' reflexes are unremarkable for age. You request a set of clinical observations and a 12 lead electrocardiogram (ECG) as there are no other clinical notes available

Vital signs: Oxygen saturations 87 % - Heart rate: 116/min - Temperature: 37.0 C - Blood pressure (right arm): 85/43 mmHg - Respiratory rate: 28/min.

1. What is the most likely explanation for the baby's symptoms? (MCQ)

A. Pulmonary hypertension in complete atrioventricular septal defect (AVSD)
B. Methemoglobinemia due to glucose-6-phosphate dehydrogenase deficiency
C. Increasing upper airway obstruction with sleep apnea
D. Congenital hypothyroidism with faltering growth

A

2. Which investigation is most likely to reveal the diagnosis? (MCQ)

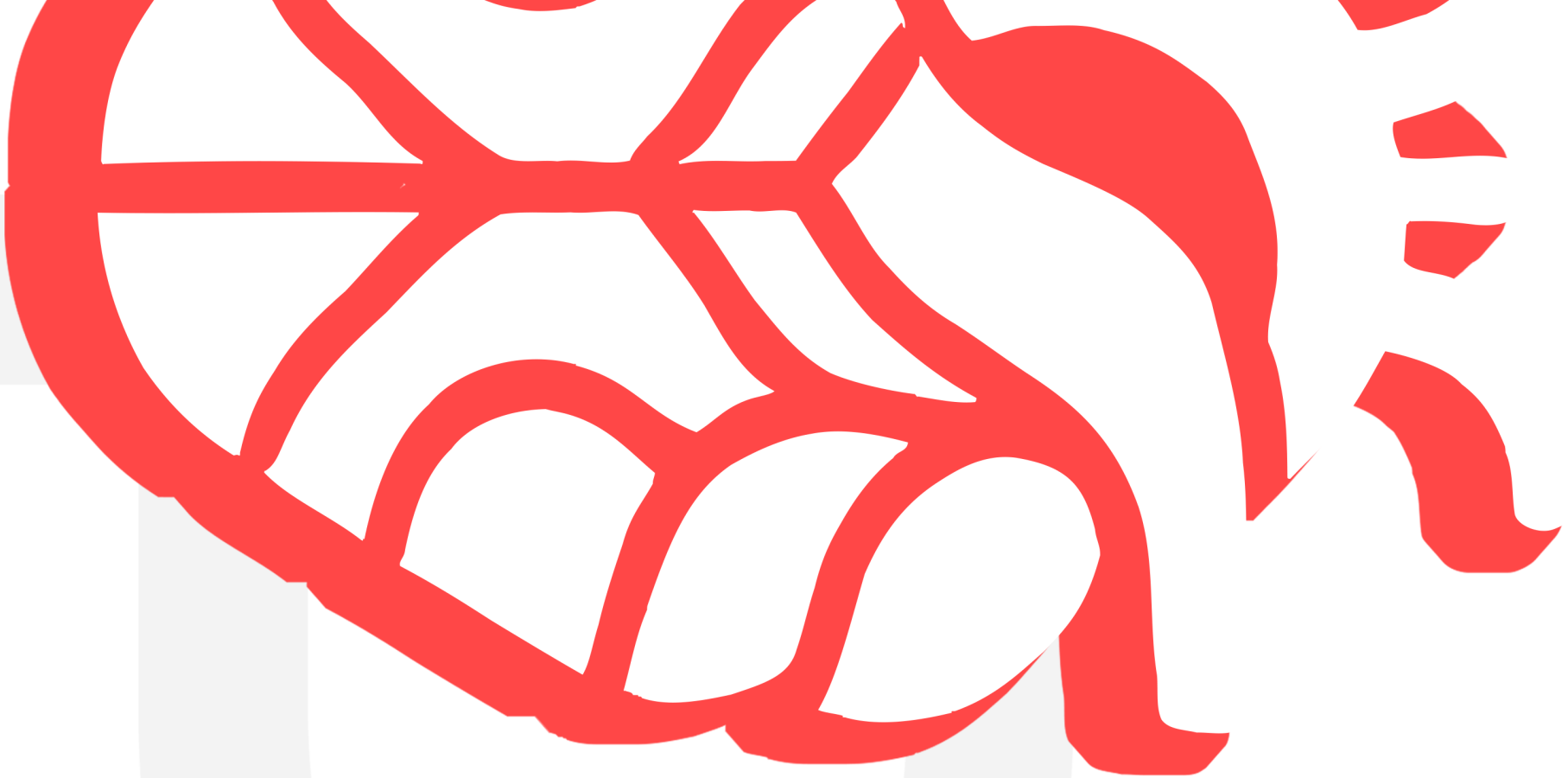
A. Transthoracic echocardiography
B. Methemoglobin measurement and co-oximetry
C. Sleep study and micro laryngoscopy under general anesthesia
D. Thyroid function tests and neck ultrasound

A

3. What would be the preferred acute management of this patient? (MCQ)

A. Urgent reversibility assessment of pulmonary hypertension in pediatric cardiology
B. 1% methylene blue administration at 1 mg/kg I.V. followed by family counselling
C. Prone positioning with continuous monitoring and possible nasogastric tube feeding
D. Rapid initiation of thyroid hormone replacement as ambulatory patient

A



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Pediatrics