By
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L1. Cardiovascular System Diseases

K:1,4,5,6,7,8,11,13. S: 1, 3,4,7,13,17,18,21,23,24 . AB: 1,3,4,5

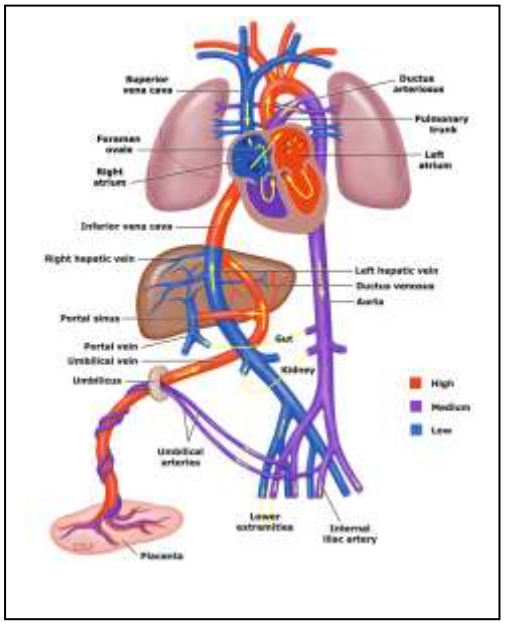
Objectives: The student should know:

- The fetal circulation and cardiovascular changes to transmit from intrauterine to extra-uterine life.
- How to classify the congenital heart diseases.
- The main clinical manifestations of cardiovascular diseases according to the age.
- How to diagnose and manage atrial septal defects.
- How to diagnose and manage ventricular septal defects.

The Fetal Circulation:

In the fetus, the placenta is responsible for the gas exchange and the lungs are functionless, so the pulmonary vessels are vasoconstricted causing shifting of the blood away from the pulmonary circulation.

In the fetal circulation, the Rt. and Lt. ventricles present in parallel circuits rather than series circuits as in newborn or adult. This parallel circulation achieved by THREE cardiovascular structures which are ***ductus venosus, foramen ovale and ductus arteriosus.***



The blood enter to the fetus through the umbilical vein where approximately 50% of it enter to the hepatic circulation and the rest 50% will bypass the liver and joint the inferior vena cava (IVC) through the ductus venosus where it will mixed with poorly oxygenated IVC blood from the lower part of the fetal body.

This combined blood enter the Rt. Atrium and is mainly directed by ***Eustachian valve*** (flap of tissue at the Rt atrial-IVC junction) across the foramen ovale into the Lt atrium then to Lt ventricle, then ejected into the ascending aorta to supplies mainly the fetal upper body and brain.

Fetal superior vena cava (SVC) blood enter the RT atrium and then mainly into the RT ventricle through the tricuspid valve then ejected into the pulmonary artery. Because the pulmonary circulation is vasoconstricted, so only 5% of the pulmonary artery blood enter the lungs and the remaining blood shifting through the ductus arteriosus to the descending aorta (Rt-to-Lt shunt) to supply the lower part of the fetal body and placenta via 2 umbilical arteries.

So, the upper part of the fetal body (including coronary arteries, cerebral arteries and upper extremities) is supplied exclusively by the Lt ventricle, while the lower part of the fetal body is supplied mainly by the Rt ventricle.

At Birth: the following changes will occurs:

1. Mechanical expansion of the lungs and increases in the arterial Po2 lead to rapid decrease in the pulmonary vascular resistance (PVR) to become much lower than systemic vascular resistance (SVR).
2. Removal of the placenta lead to increase SVR more than PVR and the closure of ductus venosus.

** These changes will lead to:

1. The blood will shift from Lt-to-Rt through the ductus arteriosus due to increased SVR and rapid decrease in the PVR and the ductus will closed over several days to form **ligamentum arteriosum** and now the entire Rt ventricle output will flows into the pulmonary circulation.
2. Increase the volume of pulmonary blood flow returning to the Lt atrium from the lungs will increase the Lt atrium volume and pressure causing functioning closure of foramen ovale although it may remain anatomically patent for several years.

Classification of the Congenital Herat Diseases (CHD):

I. Acyanotic CHD:

A. Acyanotic CHD causing increased volume load:

1. Atrial Septal Defect (ASD).
2. Ventricular Septal Defect (VSD).
3. Atrio-Ventricular Septal Defect (AV canal).
4. Patent Ductus Arteriosus (PDA).

Pathophysiology: Presence of communication between the pulmonary and systemic circulations causing shifting of oxygenated blood back to the lungs (Lt-to-Rt shunt) with increased pulmonary blood flow.

B. Acyanotic CHD causing increased pressure load:

1. Valvular pulmonary stenosis.
2. Valvular aortic stenosis.
3. Coarctation of the aorta.
4. Tricuspid stenosis.
5. Mitral stenosis.
6. Pulmonary veins obstruction.

Pathophysiology: Presence of obstruction to the normal blood flow, which is either obstruction to ventricular out flow(more common, as in the first 3 causes) or obstruction to ventricular inflow (less common, as in the last 3 causes).

II. Cyanotic CHD:

A. Cyanotic CHD with decreased pulmonary blood flow:

1. Tetralogy of Fallot (TOF).
2. Pulmonary atresia with intact septum.
3. Tricuspid atresia.
4. Total anomalous pulmonary venous return with obstruction.

Pathophysiology: In this group, there is must be presence of obstruction to the pulmonary blood flow and a communication between the pulmonary and systemic circulation through it the systemic deoxygenated venous blood shunt from right to left enter the systemic circulation (via PDA, foramen ovale, ASD and VSD).

B. Cyanotic CHD with increased pulmonary blood flow:

1. Transposition of Great Arteries (TGA).
2. Single ventricle.
3. Truncus arteriosus.
4. Total anomalous pulmonary venous return without obstruction.

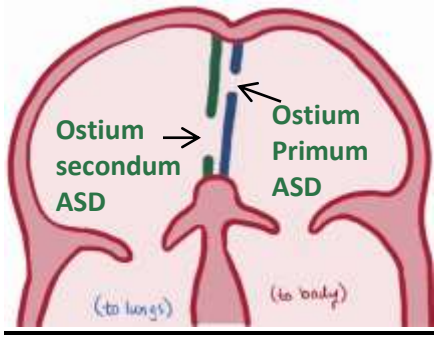
Pathophysiology: This group is not associated with obstruction of pulmonary blood flow. Cyanosis caused by either abnormal ventricular-arterial connection, or from total mixing of systemic venous and pulmonary venous blood within the heart.

Atrial Septal Defect (ASD)

ASDs represent approximately 10% of all congenital heart defects.

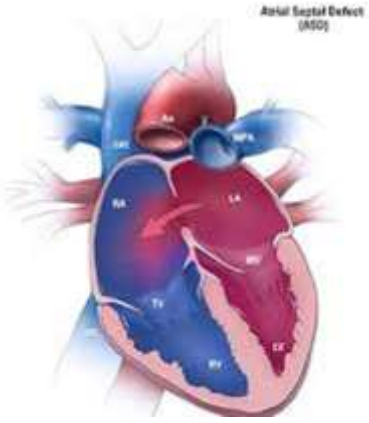
Types:

1. **Ostium secondum ASD:** is the most common type, in which the hole in the region of the foramen ovale.
2. **Ostium primum ASD:** in which the hole located near the endocardial cushions, and it may be part of a complete atrioventricular canal defect
3. **Sinus venosus ASD:** is the least common type, which may be associated with anomalous pulmonary venous return.



Pathophysiology:

The degree of left-to-right shunting is dependent on:
The size of the defect, the relative compliance of the right and left ventricles, and the relative vascular resistance in the pulmonary and systemic circulations.
In large defects, a considerable shunt of oxygenated blood flows from the left to the right atrium. This blood is added to the usual venous return to the right atrium and is pumped by the right ventricle to the lungs. With large defects, the ratio of pulmonary to systemic blood flow (Qp : Qs) is usually between 2 : 1 and 4 : 1.



Clinical features:

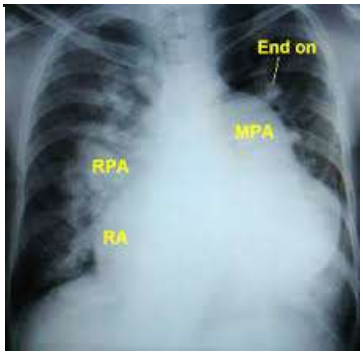
1. ASD is often asymptomatic and discovered during physical examination.
2. Even extremely large ASD, it rarely produce clinical heart failure during childhood, but subtle failure to thrive may be present.
3. Various degrees of exercise intolerance might present in older children.

On Examination:

1. There is might be mild left precordial bulge due to cardiomegaly.
2. Rt ventricular impulse can palpated on Lt. lower sternal border.
3. On auscultation:
    a. Fixed splitting of second heart sound (S2).
    b. Soft (grade I or II) systolic ejection murmur best heard in the middle and upper sternal border due to increase blood flow across the Rt ventricle into the pulmonary artery and not due to low pressure flow across the ASD.
    c. Mid-diastolic murmur at Lt lower sternal border due to increase flow across the tricuspid valve.

Diagnosis:

1. CXR: shows cardiomegaly, Rt atrial enlargement, prominent pulmonary artery and increased pulmonary vascularity.
2. ECG shows Rt axis deviation and Rt ventricular hypertrophy.
3. Echocardiography demonstrate the definitive size and location of the defect.



Treatment:

1. Asymptomatic child required no treatment.
2. Transcatheter or surgical closure indicated in:
    a. All symptomatic children.
    b. Asymptomatic patient with Qp:Qs ratio of 2:1 and more.
    c. Severe Rt ventricular enlargement.

Prognosis:

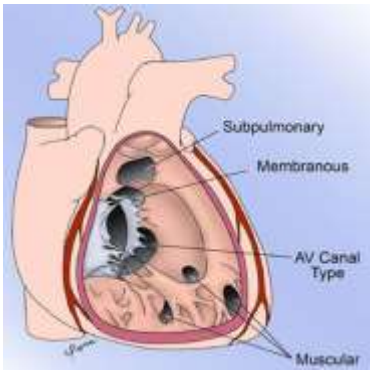
1. Small-moderate ASD in term infants may close spontaneously.
2. Secondum ASD is well tolerated during childhood and complications don't appear until the third decade or later including pulmonary HT, atrial dysrhythmias, tricuspid or mitral insufficiency and heart failure.

Ventricular Septal Defects (VSD):

Is the most common cardiac malformation, accounts for 25% of CHD.

Types:

1. **Membranous type:** Is the most common type located in the membranous portion of the septum in the posteroinferior position.
2. **Supracristal type:** Is less common, located superior to crista supraventricularis, just beneath the pulmonary valve.
3. **Muscular type:** is located in the mid portion or apical region of the ventricular septum and it may be single or multiple (Swiss cheese septum)



S):

vascular resistance (PVR) is high it will limit the Lt-to-Rt shunt. After birth, the PVR is high due to pulmonary vasoconstriction, so there is limitation of the Lt-to-Rt shunt through the VSD. When the PVR begin to fall during the first 6-8 wk after birth, the shunt will be increase and the clinical manifestations will appears.

When the PVR=SVR (Qp:Qs is 1:1), the shunt will become bidirectional and patient become cyanosed (Eisenmenger physiology).

When the PVR>SVR (Qp:Qs is 2:1 or more), the shunt will reverse to become Rt-to-Lt and the cyanosis will increase more.

Clinical Features:

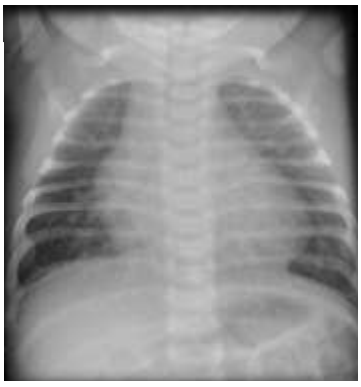
A. Small VSD with minimal Lt-to-Rt shunt and normal pulmonary pressure:

1. The patient is asymptomatic and discovered during routine examination.
2. Characteristic loud, harsh, halosystolic murmur, best heard over the lower Lt sternal border and frequently associated with thrill

B. Large VSD with excessive pulmonary blood flow and pulmonary HT:

1. Symptoms of heart failure in early infancy (Dyspnea, feeding difficulties, poor growth and profuse sweating) and recurrent chest infections.
2. Cyanosis is usually absent, but duskiness may note during infection or crying.
3. Lt precordium bulge and palpable parasternal lift is common.
4. The halosystolic murmur is less harsh than that of small VSD.
5. Mid-diastolic, low-pitched murmur at the apex due to increased blood flow across the mitral valve.

Diagnosis:



1. CXR: may be normal in small VSD, but in large VSD there is may be cardiomegaly and increased pulmonary vascularity.
2. ECG: It may be normal in small VSD, but in large VSD there is biventricular hypertrophy with notched or peaked P wave.
3. Echocardiography: for size and location of the defect, direction and magnitude of the shunt, and measurement of pulmonary pressure.

Natural coarse(natural history) of VSD: It depend on the size of VSD.

1. 30-50% of small VSDs closed spontaneously, mostly during the first 2yr of life.
2. Small muscular VSDs are more likely to close (up to 80%) than membranous VSDs (up to 35%)
3. Closed VSD often have ventricular septal aneurysm which limit the shunt magnitude.
4. Most children with small restrictive VSD remain asymptomatic.
5. Non operated VSD might increase incidence of arrhythmia, subaortic stenosis and exercise intolerance.

Treatment:

1. Prophylactic antibiotic to prevent subacute bacterial endocarditis (SBE).
2. Treatment of heart failure in large VSD by diuretics and digoxin.
3. Surgical closure indicated in continued poor growth or pulmonary HT despite medical treatment.

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L2. Patent Ductus Arteriosus (PDA)

K: 1,4,5,6,8,9. S: 1,3,4,7,13,17,18,21,23,24 .AB: 1,3,4,7 .

Objectives: The student should know:

- How to diagnose and manage patent dactus arteriosus.
- How to diagnose and manage Coarctation of aorta.
- How to diagnose and manage Tetralogy of Fallot.
- How to diagnose and manage Transposition of great arteries

PDA represent approximately 5-10% of CHD (excluding premature infants).

Etiology: Ductus arteriosus allow blood flow from the pulmonary artery to the aorta during fetal life. Failure of normal closure of this vessel result in PDA. With falling of PVR after birth, there is Lt-to-Rt shunt and increased pulmonary blood flow.

Clinical Features:

It depend on the magnitude of Lt-to-Rt shunt, which determined by the size of PDA and PVR (as in VSD).

1. Small PDA is asymptomatic.
2. Moderate-large PDA causing congestive heart failure (CHF) as the PVR decrease over the first 6-8 wks of life
3. Findings on physical examination:
   a. Wide pulse pressure.
   b. Continuous machinery murmur at Lt infra-clavicular area, it may be associated with thrill.
   c. Hyperdynamic precordium in large PDA.
   d. Splitted S2 occur with high pulmonary pressure.

Diagnosis:

1. CXR: shows full pulmonary artery shadow and increased pulmonary vascularity in large PDA. But CXR might be normal in small PDA.
2. ECG: it varies from normal to evidence of LV hypertrophy or RV hypertrophy in presence of pulmonary HT.
3. Echocardiography: for the size of PDA, magnitude of the Lt-to-Rt shunt and pulmonary pressure.

Treatment:

1. Spontaneous closure of PDA after few wks of age is uncommon in full term infants.
2. Treatment of CHF with diuretics and digoxin.
3. Closure the PDA through the catheterization is recommended even in small PDA to prevent subacute bacterial endocarditis.

COARCTATION OF THE AORTA:

occurs in approximately 10% of all CHD. It is almost always **juxtaductal** in position. During development of the aortic arch, the area near the insertion of the ductus arteriosus fails to develop correctly, resulting in a narrowing of the aortic lumen.

Clinical Manifestations

Infants with severe coarctation frequently dependent on a PDA to provide descending aortic flow. Symptoms develop when the ductus close. Clinical manifestations include:

1. Symptoms including poor feeding, respiratory distress, and shock, may develop before 2 weeks of age.
2. Classically the **femoral pulses** are weaker and delayed compared with the right radial pulse.
3. The blood pressure in the lower extremities is lower than that in the upper extremities.

Older children are usually asymptomatic:

1. There may be a leg pain with exercise, headache, or epistaxis.
2. Decreased or absent lower extremity pulses and **hypertension** (in upper extremity).
3. The machinery murmur is typically best heard in the left interscapular area of the back. If significant collaterals have developed, continuous murmurs may be heard throughout the chest.
4. An abnormal aortic valve is present approximately 50% of cases, causing systolic ejection click and systolic ejection murmur of aortic stenosis.

Diagnosis:

1. In infants, the ECG show evidence of right ventricular hypertrophy .
2. CXR show marked **cardiomegaly** and **pulmonary edema**.
3. Echocardiography shows the site of coarctation and associated lesions.
4. In older children, the ECG and chest x-ray usually show left ventricular hypertrophy and a mildly enlarged heart. **Rib notching** may also be seen in older children (>8 years of age) with large collaterals.
5. Echocardiography shows the site and degree of coarctation, presence of left ventricular hypertrophy, and aortic valve morphology and function.

Treatment:

1. IV infusion of **prostaglandin E1** (chemically opens the ductus arteriosus), digoxin, diuretics, and other supportive care.
2. **Balloon angioplasty**, especially in critically ill infants, but **surgical repair** of the coarctation is most commonly performed.

Tetralogy of Fallot (TOF):

It is the most common cyanotic CHD, represent about 10% of all CHD.

It consist of:

1. Large, non-restrictive VSD.
2. Rt ventricular out flow obstruction (pulmonary stenosis) which is most commonly in the Rt ventricular infundibulum (supvalvular) or less commonly in the pulmonary valve.
3. Rt sided aortic arch which overriding the ventricular septum (overriding of aorta).
4. Rt ventricular hypertrophy (RVH).

Pathophysiology:

Systemic venous return to right atrium and right ventricle is normal.
When the Rt ventricle contracts in the presence of marked pulmonary stenosis, blood is shunting across the VSD into the Lt ventricle then into aorta (Rt-to-Lt shunt) causing arterial desaturation and cyanosis. The severity of cyanosis depend on the degree of pulmonary stenosis. In the severe pulmonary stenosis the pulmonary blood flow is supplied by PDA.
Degree of Rt ventricular outflow obstruction determine the timing of onset of symptoms, the severity of cyanosis and the degree of RVH.
In mild-moderate Rt ventricular outflow obstruction, there's balanced shunt across the VSD and the patient is not visibly cyanosed (Acyanotic or "pink" TOF).
When the obstruction is severe, the cyanosis present at birth and worsen with closure of ductus arteriosus.

Clinical features:

1. Infants with mild pulmonary stenosis may be initially presents with heart failure due to severe Lt-to-Rt shunt across large VSD.
   Cyanosis is absent at birth, but it will appear latter in 1st year of life due to increased pulmonary stenosis secondary to hypertrophy of right ventricular infundibulum.
2. Infants with severe pulmonary stenosis have cyanosis immediately in neonatal period and pulmonary blood flow is depend on the ductus arteriosus. When the ductus begin to close over the 1st few hours or days of life it will cause severe cyanosis and circulatory collapse.
3. Older children with long-standing cyanosis have dusky blue skin, gray sclera with engorged blood vessels and marked clubbing of fingers and toes.
4. Systolic ejection murmur due to pulmonary stenosis present in all patients which best heard at the 2nd Left intercostal space and radiate to the back.
5. Paroxysmal hypercyanotic attacks ("cyanotic" or "blue" or "tet" spells):
   It's particular problem during the first 2 years of life occurs due to reduction of an already compromised pulmonary blood flow precipitated by excessive crying, vigorous exercise or systemic infection. If the spell is prolong, it will result in severe systemic hypoxia and metabolic acidosis.
   These spells are characterized by:
   a. The onset is spontaneous and unpredictable.
   b. Spells occurs most frequently in the morning on awaking or after vigorous crying.
   c. Infants developed hyperpnea, increasing cyanosis, gasping breathing and might end with syncope or death.
   d. Temporary disappearance or reduction of ejection systolic murmur due to decrease blood flow across the aortic valve.
   e. The spells may last from a few minutes to a few hours. Short spells followed by generalized weakness and sleep, while prolong severe spells may progress to unconsciousness or convulsion or hemiparesis.
   f. Toddler patient adapt squatting position during the spell in attempt to increase systemic venous return and then increase pulmonary blood flow.

Note:

Infants who are only mildly cyanosed at rest are more liable to develop cyanotic attacks because they lack the homeostatic mechanisms to tolerate rapid decrease of arterial oxygen saturation such as polycythemia.

Diagnosis:

1. ECG: shows Rt axis deviation and RV hypertrophy.
2. CXR: shows **boot-shaped heart** (created by small main pulmonary artery and upturned apex secondary to RVH) and **oligemic lung fields.**
3. Echocardiography: shows the anatomical features of TOF.

Treatment:

1. Treatment of hypercyanotic attacks:

   Depending on the frequency and severity of hypercyanotic attacks,1 or more of the following procedures should be done in sequence:

   a. Knee-chest position of the infant which may prevent progression of an early spell.
   b. Oxygen administration (although increasing oxygen will not reverse cyanosis because it's due to intracardiac shunting).
   c. S.C morphine in a dose not in excess of 0.2 mg/kg.
   d. IV infusion of sodium bicarbonate if the spell is severe and not responding to above measures.
   e. Intubation and sedation are often sufficient to break the spell which is severe and not responding to other treatments.
   f. Drugs that increase systemic vascular resistance, such as intravenous phenylephrine, can improve right ventricular outflow, decrease the right-to-left shunt, and improve the symptoms.
   g. IV propranolol (0.1 mg/kg given slowly to a maximum of 0.2 mg/kg).

2. Complete surgical repair with closure of VSD with removal of pulmonary stenosis can be performed during infancy.
3. Palliative shunt surgery **(Blalock-Tausing shunt)** in which a palliative shunt created between subclavian artery (systemic circulation) and pulmonary artery. It done for a complex TOF with complete repair later in life.
4. Prophylactic antibiotic to prevent subacute bacterial endocarditis.

Complications:

1. **Cerebral thrombosis:** Occurs as a complication of severe polycythemia and dehydration. It most often in patient younger than 2 years.
2. **Brain abscess:** It less common than cerebral thrombosis and it usually occurs in children older than 2 years.
3. **Subacute bacterial endocarditis.**
4. **Heart failure:** Is unusual in TOF except in young infants with "pink" or "Acyanotic" TOF and as the degree of pulmonary stenosis increase with age the symptoms of heart failure resolved and the patient develop cyanosis often at 6-12 months of age.

Transposition of Great Arteries (TGA):

It represent about 5% of CHD, but it is the most common cyanotic disease present in the neonatal period.

In TGA the aorta arises from Rt ventricle and pulmonary artery arises from the Lt ventricle. This will result in desaturated blood return to the Rt side of the heart and pumped back to the body via the aorta, while the oxygenated blood from the lungs enter the Lt side of the heart and pumped back to the lungs via the pulmonary artery.

Without mixing of the tow circulations, death occurs shortly after birth. This mixing may occurs in atrial level (through a patent foramen ovale or ASD) or at ventricles (through a VSD) or at great vessels (through PDA).

Clinical Features:

1. Cyanosis is always present and its severity depend on the amount of mixing between systemic and arterial circulations.
2. Tachypnea.
3. Single S2, and if there's no VSD there's no murmur.
4. Children with TGA and large VSD may present with signs of heart failure with palpable Lt and Rt ventricular impulses, single S2 and load VSD murmur (pansystolic murmur).

Diagnosis:

1. ECG: shows Rt axis deviation and RVH.
2. CXR: shows **increase pulmonary vascularity** and classical cardiac shadow which called **egg on a string** caused by narrow superior mediastinum.
3. Echocardiography: shows the transposition of great artery, site and amount of the shunt and any associated anomalies.

Treatment:

1. Prostaglandin E1 infusion immediately after birth to maintain the patency of ductus arteriosus and allow mixing of the blood.
2. Balloon atrial septostomy done if there's no response to prostaglandin infusion.
3. Complete surgical repair by **arterial switch operation**, in which the aorta is reconnected to the Lt ventricle and the pulmonary artery is reconnected to the Rt ventricle. It is usually done within the first 2 weeks of life.

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L3. Acquired Heart Diseases

K: 1,4,5,6,8,9,11,13. S: 1,3,4,7,13,17,21,23,24 . AB: 1,3,4,5.

Objectives: The student should know:

How to diagnose, manage and prevent the infective endocarditis.

Infective Endocarditis (IEC):

It include acute and subacute bacterial endocarditis in addition to non-bacterial endocarditis caused by viruses, fungi and other microorganisms.

It is a significant cause of morbidity and mortality in children and adolescents.

IEC is often complicate CHD or rheumatic heart diseases and prosthetic heart valves, but it can occur in children without any cardiac abnormalities (native heart).

Pathogenesis:

IEC is a consequence of jet streams of turbulent blood (from PDA, VSD, or systemic-pulmonary shunt) which cause damage of vascular endothelium causing nonbacterial thrombotic embolus which thought to be the initiating lesion for IEC.

Predisposing factors:

1. Prior congenital or rheumatic heart diseases.
2. Preceding dental, urinary tract or intestinal procedures.
3. Intravenous drug use.
4. Central venous catheter.
5. Prosthetic heart valve.

Causative MO (Etiology):

- ***Streptococcus Viridans*** (alpha-hemolytic streptococci) and ***Staphylococcus aureus*** are the most common causative agents for bacterial endocarditis.
- Other MO e.g. *St. pneumoniae, H. influenzae*, coagulase-negative staphylococci and fungi are less common.
- In about 6% of cases, the blood culture is negative for any microorganism.

Clinical Features:

A. Symptoms:

1. Early manifestations are usually mild especially when St. Viridans is the infective MO which include prolonged fever without other manifestations (except occasionally weight loss) which might persist for several months.
2. Or the onset might be acute and sever with high intermittent fever and prostration
3. Other "nonspecific" symptoms include fatigue, myalgia and arthralgia, headache, chills, nausea and vomiting, chest and abdominal pain, dyspnea, night sweating and CNS manifestations (stroke and seizures).

B. Clinical signs:

1. Elevated body temperature.
2. Tachycardia.
3. Petechiae.
4. New or changing heart murmur.

5. Splenomegaly.
6. Signs of heart failure and arrhythmias.
7. Clubbing.
8. Metastatic infections (Arthritis, meningitis, Mycotic arterial aneurysm, pericarditis, abscesses, septic pulmonary emboli).
9. Classical skin signs: it developed later in the course of the disease and they may represent vasculitis caused by circulating Ag-AB complexes. These signs are:
    a. **Osler nodes** (tender, pea-size intradermal nodules in the pads of the fingers and toes)
    b. **Janeway lesions** (painless small erythematous or hemorrhagic lesions on the palms and soles).
    c. **Splinter hemorrhages** (linear lesions beneath the nails).

Diagnosis:

1. Blood culture: Is the main way for confirmation of diagnosis. 3-5 blood samples should obtained for culture after perfect sterilization of skin to prevent contamination because the bacteria which might contaminated skin might themselves the cause of IEC.
   In 90% of the cases, the causative agent is recovered from the first 2 blood cultures. The timing of blood collection is not important because the bacteremia is relatively constant.
   The bacteremia is low grade in 80% of cases (<100 colony units\ ml of blood), so the laboratory should be notified that IEC is suspected, so that, the blood cultured on enriched media for longer than usual (>7 days) to detect fastidious bacteria or fungi.

2. Elevated ESR and C-reactive protein.
3. CBC shows anemia and leukocytosis.
4. Immune complexes detection.
5. Positive rheumatoid factor.
6. GUE shows hematuria.
7. Echocardiography shows evidence of valve vegetation, prosthetic valve dysfunction, myocardial abscess, and new-onset valve insufficiency.

Complications:

1. Heart failure (most common) due to vegetation involving aortic or mitral valves.
2. Systemic emboli: CNS and pulmonary embolisms.
3. Mycotic aneurysm.
4. Valvular obstruction by large vegetation.
5. Acquired VSD.
6. Heart block due to involvement of conducting system.
7. Other complications: meningitis, osteomyelitis, arthritis, renal abscess and immune complex-mediated GN.

Treatment:

1. Antibiotic therapy should be started immediately once the diagnosis is made.

- Empirical antibiotic therapy (before obtaining blood culture results) include **Vancomycin** (40mg\kg\24 hr in 2-3 divided doses) ***Plus*** **Ceftriaxone** (100mg\kg\24 hr once a day).
- Antibiotic therapy should continue for 4-6 weeks and modified according to the blood culture results.
- If the causative agent is *St. Viridans* the drug of choice is **crystalline penicillin G** (200,000 IU\kg\day) or **ceftriaxone**
  If the patient is sensitive to penicillin, is treated with **ceftriaxone** *plus* **gentamycin** or **Vancomycin**.

- If the causative agent is staphylococci, so it will treated by **naficillin** or **oxacillin** for 6 wk *plus* gentamycin for 3-5 days (optional).
- If the patient is allergic to penicillin, the treatment is **ceftazolin** for 6 wk *plus* **gentamycin** for 3-5 days (optional).
  If the patient is infected by oxacillin-resistance strains then treated with **Vancomycin** for 6 wks.

2. Treatment of heart failure by diuretics, after load reducing agents and digoxin.

Prevention:

Previously, antibiotics prophylactic therapy was recommended for any patient at risk for development of IEC before doing any dental, genitourinary or lower GIT procedures. But nowadays the prophylactic antibiotics indicated only before dental procedures in patient with cardiac conditions which are associated with poor prognosis of IEC which are include:

1. Prosthetic cardiac valve.
2. Previous IEC.
3. Congenital heart diseases:
   a. Unrepaired cyanotic CHD including palliative shunts.
   b. Completely repaired CHD with prosthetic materials during the first 6 mo after the procedure.
   c. Repaired CHD with residual defect at the site or adjacent to the prosthetic device.
   d. Cardiac transplant recipients.
4. Permanently damaged valve by rheumatic heart disease.

Prophylactic antibiotic regimens for dental procedures:

1. Oral amoxicillin 50 mg\kg.
2. If the patient is unable for oral intake, the prophylaxis by:
   - Ampicillin 50mg\kg IM or IV
   - **Or** Ceftriaxone 50mg\kg IM or IV
3. Patients allergic to penicillin and able for oral intake:

- Cephalexin 50 mg\kg
- **Or** clindamycin 20mg\kg.
- **Or** Azithromycin or clarithromycin 15mg\kg.

4. Patients allergic to penicillin and unable for oral intake:
   - Ceftriaxone 50mg\kg IM or IV.
   - **Or** clindamycin 20mg\kg IM or IV.

Prognosis:

Mortality rate of IEC is 20-25% and serious morbidity occur in 50-60% despite of antibiotic therapy.

By
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L4. Acute Rheumatic Fever (RF)

K: 1,2,3,6,7. S: 1,3,4,7,13,17,18,21,23,24 .AB: 1,3.4,5,8,9.

Objectives: The student should know:

How to diagnose, manage and prevent the acute rheumatic fever.

It is a delayed sequela of group A beta-hemolytic streptococcal (GAS) pharyngitis as immunological reaction to the infection.

It is rarely followed skin infection by GAS and it is commonly affect children between 6 and 15 years of age.

It is the most common acquired heart diseases in all age groups, accounting for about 50% of all cardiovascular diseases. Its commonly occurs in overcrowded and poor communities and there might be genetic predisposition for rheumatic fever.

Clinical features:

Acute RF typically develops 2-4 wk after the acute GAS pharyngitis, and the disease characterized by high rate of recurrence after the initial attack of acute RF.

The clinical features consist of **5 major** and **4 minor criteria,** which are called **revised Jones criteria** *PLUS the evidence of recent GAS infection***.**

Major criteria:

1. Migratory polyarthritis:

Is the earliest manifestation of acute RF, occurs in about 75% of all patients. It typically involve the large joints (knees, wrists, and elbows) while the involvement of spine, small joints of hands and feet or hip is uncommon.

The affected joint is hot, red, swollen and extremely tender that the patient is not tolerate even the friction of bed sheets. These manifestations may preceded by sever joint pain (arthralgia).

The arthritis is non-deforming (completely resolve without residual joint destruction) and have dramatic response to NSAID (salicylates).

The arthritis is characteristically migratory in nature, that mean the affected joint become normal within 2-3 days without treatment, while one or more other large joints become involved.

Monoarticular arthritis is unusual unless the salicylate is prematurely used during the early course of acute RF which stop the progression of migratory arthritis.

2. Carditis:

It occurs in about 50% of all cases of acute RF. It's either **subclinical carditis** (echocardiac evidence of valvulitis but without murmur) or **clinical carditis** (echocardiac evidence of valvulitis plus new cardiac murmur).

It is the most serious manifestation of acute RF and it characterized by pancarditis (inflammation of pericardium, myocardium and endocardium "valvulitis") and the endocarditis (valvulitis) is universal finding in rheumatic carditis, whereas the presence of pericarditis or myocarditis is variable.

Presence of myocarditis and\or pericarditis without clinical evidence of endocarditis (Valvular disease) it is almost never rheumatic heart disease.

The most common rheumatic heart disease is isolated mitral regurgitation or combined aortic and mitral regurgitations.
Clinical manifestations of carditis include tachycardia, new murmur (aortic or mitral regurgitation), pericarditis, cardiomegaly and signs of congestive heart failure.

3. Chorea "Sydenham Chorea":

Occurs in 10-15% of patients with acute RF and usually presents as isolated, subtle movement disorders.
It characterized by emotional liability, incoordination, poor school performance, uncontrollable movements and facial grimacing. These manifestations are exacerbated by stress and disappearing with sleep.
The latent period between the acute GAS infection and chorea usually longer than that for arthritis or carditis and it might be as long as months (so it usually presents as isolated manifestation) and it rarely lead to permanent neurological sequelae.

Examination methods of chorea:

1. Milkmaid's grip: irregular contractions and relaxations of fingers muscle while squeezing the examiner's fingers.
2. Spooning and pronation of the hand while the arms extended.
3. Warmian darting movement of the tongue on protrusion.
4. Examination of hand writing for fine motor movements.

Diagnosis of chorea based on clinical examination and raised GAS antibody titers, but because of the prolonged latent period between the GAS infection and onset of chorea, the antibody titers may return to normal levels.

4. Erythema Marginatum:

It is rare manifestation of acute RF occurs in about 1% of all cases, consist of nonpruritic erythematous macular rash with pale centers and serpiginous borders, occurs on the trunk and extremities (but not on the face) and it accentuated by local heat application.

5. Subcutaneous nodules:

It's rare manifestation (<1%) seen predominantly with chronic or recurrent RF. They are firm, painless, nonpruritic, mobile nodules found on the extensor surfaces of the large and small joints, scalp and spine.
There's a correlation between the presence of these nodules and significant rheumatic heart disease.

Minor criteria:

1. Arthralgia ((joint pain without physical signs of arthritis)), it used as minor criteria only if the arthritis is not used as major criteria.
2. Fever (38.2-38.9 C).
3. Elevated acute phase reactant (ESR, C-reactive protein or leukocytosis).
4. Prolonged PR-interval on ECG (unless the carditis is a major criteria).

Evidence of recent group A streptococcal (GAS) infection:

1. Scarlet fever.
2. Positive throat swab culture for GAS (rarely positive).
3. Raised ASO (antistreptolysin O) titer and other anti-streptococcal Abs e.g. anti DNase B antibodies.

Diagnosis of Rheumatic fever:

The diagnosis of acute RF made if the patient have **2 major criteria** or **1 major and 2 minor criteria** *PLUS* **the evidence of recent GAS infection.**

Recurrent RF diagnosed by presence of 3 minor criteria plus evidence of recent GAS infection.

There are THREE conditions in which the diagnosis of RF done without strict adherent to Jones criteria, these are:

1. When the chorea is only the major manifestation of RF.
2. When the indolent carditis is the only manifestation.
3. In limited number of patients with recurrent RF in high risk population.

Treatment of RF:

1. General measures: bed rest and closed monitoring for evidence of carditis.

2. Antibiotics therapy: It should be started regardless to the throat swab culture results by given 10 days course of oral penicillin or amoxicillin or single I.M. injection of benzathin penicillin G to eradicate GAS from URT.

If the patient is allergic to penicillin give 10 days course of erythromycin or 5 days course of azithromycin or clindamycin.

3. Anti-inflammatory therapy:

Anti-inflammatory drugs (aspirin or corticosteroid) should be delayed if arthralgia or atypical arthritis is the only manifestation of RF because premature treatment with one of these drugs may interfere with the development of characteristic migratory polyarthritis and obscure the diagnosis of acute RF.

Acetaminophen (paracetamol) can be used for treatment of pain and fever while patient observed for more definite signs of acute RF.

Oral salicylate (aspirin): indicated for patient with typical migratory arthritis or those with carditis without cardiomegaly or congestive heart failure.

Dose: 50-70 mg\kg\day in 4 divided doses orally for 3-5 days followed by 50 mg\kg\day in 4 divided doses orally for 3 wk then half of the dose for another 2-4 wk.

Corticosteroid (prednisone): indicated in patients with carditis associated with cardiomegaly and\or congestive heart failure.

Dose: 2mg\kg\day in 4 divided doses orally for 2-3 wk followed by half of the dose for another 2-3 wk and then gradual tapering of the dose by 5 mg\day every 2-3 days. When prednisone is being tapered, the aspirin at dose of 50 mg\kg\day in 4 divided doses orally should be given for 6 wk to prevent rebound of inflammation.

4. Treatment of congestive heart failure: by digoxin, fluid and salt restriction, diuretics and oxygen.

5. Treatment of Sydenham chorea: Phenobarbital is the drug of choice. If it is ineffective, then haloperidol or chlorpromazine should be used as alternative therapies. Anti-inflammatory agents are not indicated in treatment of chorea because it often occurs as isolated manifestation after resolution of acute phase of disease, but some patients may get benefit from a few weeks course of corticosteroid if the other drugs are ineffective in treatment of chorea.

Prevention of RF:

A. Primary prevention: Is prevention of first attack of acute RF by identification and eradication of GAS pharyngitis (as in the treatment of RF).

B. Secondary prevention: Is prevention of recurrent attacks of RF in patients with previous attack of acute RF by continuous prophylactic antibiotic therapy to prevent any GAS infection of URT.

This secondary prevention should be started as soon as the diagnosis of acute RF made and immediately after a full course of antibiotic therapy.

Duration of antibiotic prophylaxis therapy:

1. Patients who did not have carditis during the first attack have low risk of carditis with recurrent RF so the prophylactic antibiotics should continue until the age of 21yr or until 5yr after the last attack of RF, whichever is longer.
2. Patients who have initial RF with carditis but without valvular heart disease required prophylaxis for 10 years or until 21yr of age, whichever is longer.
3. Patients with carditis and Valvular heart disease in initial attack of acute RF required continuous antibiotic prophylaxis into the adulthood (40yr) or for long life because they are at high risk of having carditis with recurrent RF and for additional cardiac damage.

CATEGORY	DURATION of prophylactic therapy
Rheumatic fever without carditis	5 yr or until 21 yr of age, whichever is longer
Rheumatic fever with carditis but without valvular heart disease	10 yr or until 21 yr of age, whichever is longer
Rheumatic fever with carditis and persistent valvular heart disease	For adulthood (40yr), sometimes lifelong prophylaxis

Regimen of secondary prevention:

1. Single I.M. injection of benzathin penicillin G every 4wk or every 3wk in populations with high incidence of RF.
2. Oral penicillin V, 250 mg twice daily.
3. Sulfadiazine or sulfasoxazole once daily.
4. Macrolides (erythromycin or clarithromycin) or azithromycin are used for patients who are allergic to penicillin and sulfonamide.

Prognosis of Rheumatic fever:

It depend on the degree of permanent cardiac damage. Cardiac involvement may resolve completely especially in the first attack and followed by prophylactic therapy. The severity of cardiac involvement worsens with each recurrent R

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L5. Congestive Heart Failure (CHF)

K:1,4,5,7,11, S: 1,3,4,7,13,17,18,21,23,24 .AB: 1,3,4,5 .

Objectives: The student should know:

- The diagnostic criteria of CHF.
- Causes of CHF according to age groups.
- How to diagnose and manage CHF

The doses, regimen, side effects of common drugs used in treatment of CHF

Definition: Is inability of the heart to deliver adequate cardiac output to meet the body metabolic needs.

Causes:

Age group	Causes
Fetus	1. Severe anemia e.g. hemolysis (severe Rh incompatibility), fetal-maternal transfusion, hypoblastic anemia or parvovirus B19 infection. 2. Supraventricular or ventricular tachycardia. 3. Complete heart block as in maternal SLE. 4. Arteriovenous (A-V) malformations. 5. Myocarditis.
Premature neonate	1. Fluid overload. 2. PDA 3. VSD 4. Bronchopulmonary dysplasia causing cor pulmonale.
Full term neonate	1. Asphyxial cardiomyopathy. 2. A-V malformation. 3. Lt sided obstructive heart lesions e.g. coarctation of aorta, or severe aortic stenosis. 4. Transposition of Great Arteries (TGA). 5. Severe anemia. 6. Supraventricular tachycardia. 7. Complete heart block. 8. Myocarditis.
Infant-Toddler	1. VSD (Lt-to-Rt shunt). 2. A-V malformation. 3. Metabolic or genetic cardiomyopathy. 4. Acute hypertension (hemolytic uremic syndrome). 5. Supraventricular tachycardia (SVT). 6. Kwasaki disease. 7. Myocarditis.

Child-adolescent	1. Rheumatic fever. 2. Acute hypertension (glomerulonephritis) 3. Viral myocarditis. 4. Thyrotoxicosis. 5. Hemochromatosis. 6. Cancer therapy (radiation, doxorubicin) 7. Sickle cell anemia. 8. Endocarditis. 9. Arrhythmias. 10. Cor pulmonale e.g. cystic fibrosis and chronic air way obstruction 11. Cardiomyopathy.

Clinical features:

Infant: poor feeding, failure to thrive (FTT), tachypnea, tachycardia, diaphoresis (sweating) with feeding and hepatomegaly.

Older children:

Symptoms: dyspnea, easy fatigability, and edema.

Clinical signs: tachycardia, gallop rhythm, weak pulse.

If the Lt-sided heart failure is predominant, there is tachypnea, orthopnea, wheezing and pulmonary edema.

If the Rt-sided heart failure is predominant, there is hepatomegaly, edema and distended neck veins.

Investigations:

1. CXR: is not specific but absence of cardiomegaly usually rules out diagnosis of CHF.
2. ECG: is not specific but it useful for assessment of heart chambers (Lt, Rt, or biventricular hypertrophy) and for detection of some underlying causes e.g. arrhythmias or ischemia.
3. Echocardiography: It diagnostic, it assess the chamber sizes, myocardial function, and detection of congenital heart diseases if present.
4. Blood gas analysis: shows hypoxia and metabolic or respiratory acidosis in sever CHF.
5. Serum electrolytes levels especially serum sodium level because infants with CHF often have hyponatremia due to water retention which decrease more with chronic diuretic therapy

Treatment:

A. Removal of underlying cause, if possible.

B. General measures:

1. Rest: child allowed for rest during the day time and adequate sleep at night, but strict bed rest is not necessary.
2. Position: patient sleep in semi-upright position (45 degree).
3. Oxygen supplementation especially in presence of pulmonary edema.

4. Diet: infant with CHF usually have FTT due to increased metabolic demands and decrease intake, so the caloric intake should increase to 24 calories\oz but not more because it will cause diarrhea and increase solute load on already compromised kidneys.
   N\G tube may be beneficial in infants or children with poor feeding because of dyspnea or gastro-esophageal reflux (GER).
   Low-sodium formula is not recommended because it poorly tolerated and may exacerbate diuretic-induced hyponatremia. Breast milk is ideal low sodium diet.

C. Preload-reducing agents (diuretics):

They interfere with renal reabsorption of water and sodium, so they reduce the pulmonary and cardiac fluid overload.

1. **Furosemide (Lasix):**is the most common diuretic used in pediatrics. It potent diuretic because it inhibits sodium and chloride reabsorption in distal tubules and loop of Henle.
   Dose: Acute treatment 1-2 mg\kg\24hr I.V. until clinical improvement, then chronic treatment with 1-4mg\kg\day in 1-4 doses orally.
   Side effects: hypokalemia, so the patient required oral potassium chloride supplementation or potassium-sparing agent (spironolactone) with furosemide.

2. **Spironolactone (aldactone):** it aldosterone inhibitors, it enhance potassium retention eliminating the need for oral potassium chloride supplementation which is poorly tolerable.
   Dose: 2mg\kg\day orally in 2 divided doses.

3. **Chlorothiazide:** is less potent than furosemide because it affect electrolyte reabsorption in the renal tubules only.
   Dose: 10-40 mg\kg\day in 2 divided doses.
   Potassium chloride or spironolactone is must be administer with chlorothiazide because of its hypokalemic effect.

D. Afterload reducing agents (angiotensin converting enzyme inhibitors (ACEI) and angiotensin II receptor blockers:

They decrease ventricular afterload by decreasing peripheral vascular resistance (arterial dilatation) to improve myocardial performance.
They also decrease systemic venous tone (venodilatation) which significantly decrease the preload.
They are not used in patients with Lt ventricular out flow obstruction as in aortic stenosis because it reduce the coronary perfusion. These agents include.

1. **Nitroprusside:** it should be given by IV infusion in the intensive care units(ICU) only for critically ill patients and for short time only due to its serious side effects.
   Side effects:
   1. Sever hypotension due to its arterial and venous dilation effect.

2. Cyanide poisoning, because it metabolized in the body to cyanide which detoxified in the liver into thiocyanate and excreted in the urine. With large dose of Nitroprusside, cyanide toxicity will occurs causing fatigue, nausea, disorientation, acidosis and muscle spasms.

2. ACEI (captopril and enalpril): they have the following effects:

1. Arterial dilation by blocking angiotensin II production, so they are afterload reducers.
2. Causing venodilatation so they reduce the preload.
3. They have aldosterone inhibitor effect, so the control salt and water retention.

Doses:

Captopril: 0.3-6mg\kg\day in 3 divided doses orally.

Enalpril: 0.05-0.5 mg\kg\day in 1-2 doses orally.

Side effects: hypotension, hyperkalemia, pruritic maculopapular rash, neutropenia, renal toxicity and chronic cough.

E. Digoxin:

Is the mainstay in treatment of CHF, although the combination of diuretics and ACEI significantly reduce need to digoxin or even is no longer use in some centers.

Dose: 0.01 – 0.04 mg\kg\day according to the patients age.

Rapid digitalization is achieved within 36hr by given half of the total dose (IV) immediately then 1\4 of the total dose after 12hr and then the last 1\4 after the next 12hr.

Important considerations (cautions) with digoxin administration:

1. ECG should be done before each of 3 doses of digoxin and digoxin should stop if new rhythm disturbance developed.
   Prolonged P-R interval alone is not an indication for digoxin stopping, but indicate delay the next dose or decrease the dose according to the patient status.
   Minor ST segment or T-wave changes are commonly noted with digoxin administration and should not affect the digitalization regimen.
2. Close monitoring of serum electrolytes especially in concomitant use of diuretics because hypokalemia and hypercalcemia increase digoxin toxicity.
3. Digoxin is better to be avoided in patients with myocarditis if possible or given in half of the total dose only because it increase the risk of arrhythmias in these patients.

Factors increase digoxin toxicity:

1. Hypokalemia.
2. Hypomagnesemia.
3. Hypercalcemia.
4. Myocarditis.
5. Prematurity.
6. Renal failure.