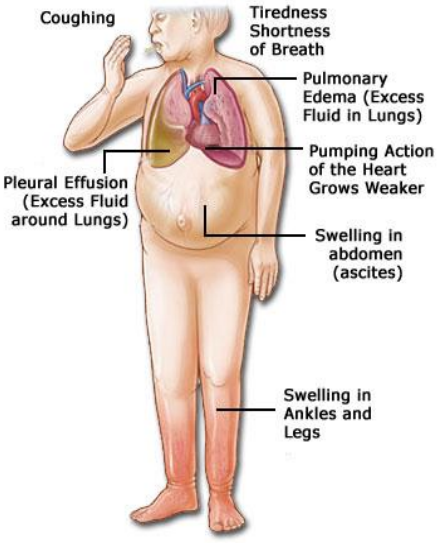
- Congestive Heart Failure (CHF):

- It is defined as inability of the heart to pump blood normally thus leading to hypoperfusion and reduced oxygen delivery to tissues. Hypoperfusion will result in the following:
  - ✓ Increase in hear rate and contractility (trying to increase cardiac output).
  - ✓ It will be sensed by kidneys as reduced blood volume thus activating renin-angiotensin system which leads to retention of salts and water.
  - ✓ Release of catecholamines from SNS will also increase heart rate and contractility.
- **Causes of CHF:**

Congenital	**Increased pulmonary blood flow**: large VSD, transposition of great arteries, truncus arteriosus and total anomalous pulmonary venous connection
	Obstructive lesion: aortic, pulmonary or mitral stenosis; corarctation of aorta
	Others lesions: mitral or tricuspid regurgitation
Acquired	Viral myocarditis (most common), infections (endocarditis, pericarditis), hypothyroidism, IHD, cardiomyopathies, dysarrhythmias and drugs (chemotherapeutic agents)
Others	Severe anemia

- **Clinical features:**

Pulmonary congestion	Tachypnea, cough, wheezing, rales and pulmonary edema on CXR
Impaired myocardial performance	Tachycardia, sweating, reduced urine output, pale skin and enlarged cardiac shadow on CXR
Systemic venous congestion	Hepatomegaly and peripheral edema
Late manifestations	Cyanosis and shock



- **Management:**

Cardiac glycosides	Digoxin
Loop diuretics	Furosemide
Surgical repair	CHD secondary to congenital heart diseases

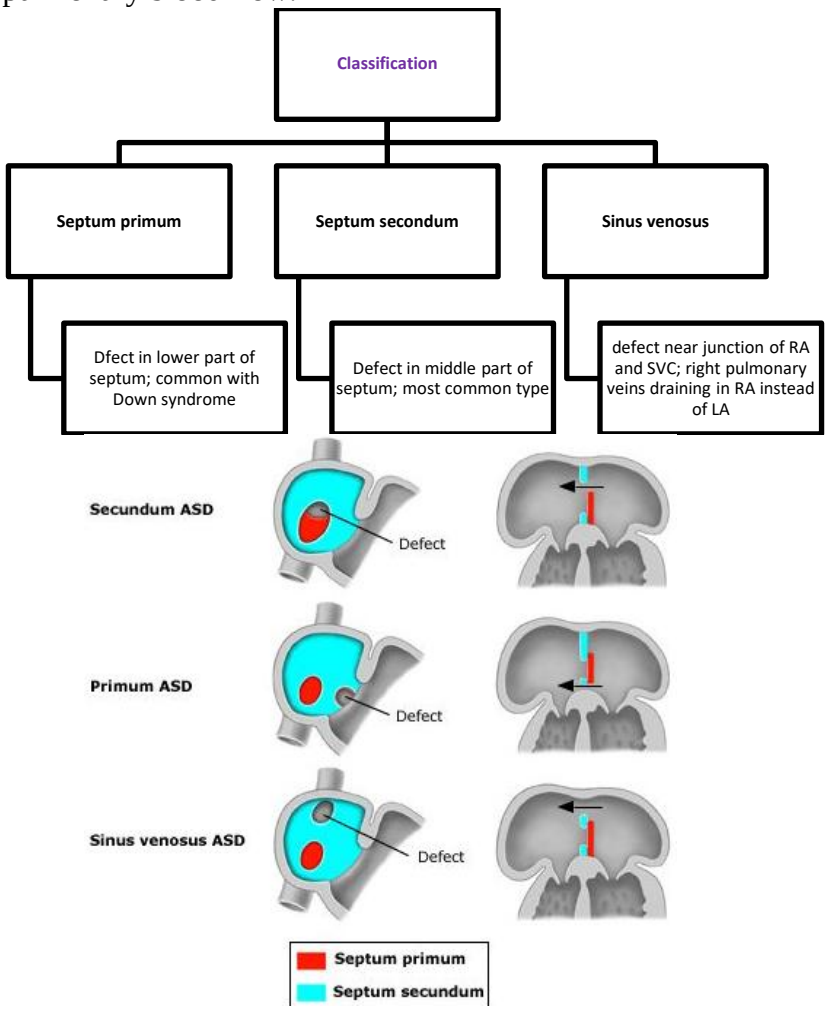
- **Innocent cardiac murmurs:**
  - Those which are caused by turbulent blood flow with no structural abnormalities of the heart. This occurs in 50% of children at some point during childhood.
  - **Grading of heart murmurs:**

Grade-I	Soft; heard under quiet conditions
Grade-II	Soft; heard under noisy conditions
Grade-III	Easily heard prominent murmur
Grade-IV	Loud murmur with thrill
Grade-V	Loud murmur with edge of stethoscope tilted against the chest plus a thrill
Grade-VI	Very loud murmur heard 5-10 mm from the chest plus a thrill

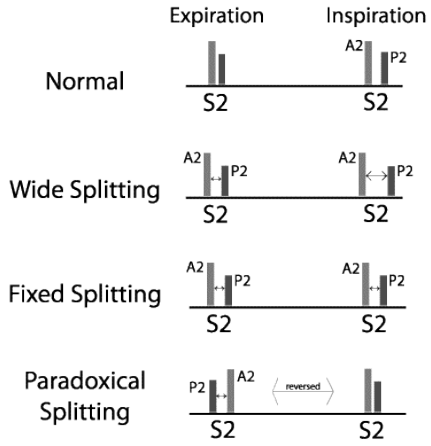
  - **Clinical features of innocent heart murmurs:**

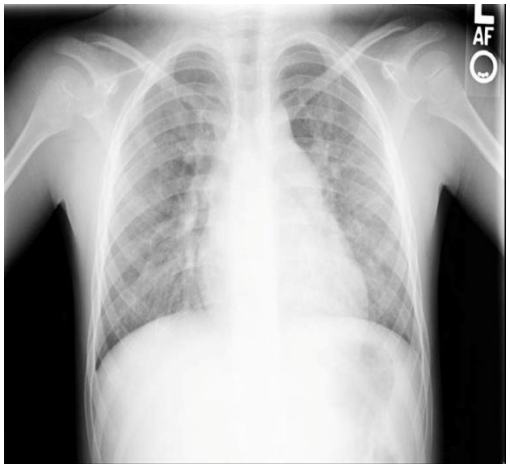
Murmur	Age	Location	Characteristics
Still's murmur	2-7 years	Mid-left sternal border	Grade I-III; systolic, vibratory, loudest when supine; louder with exercise
Pulmonic systolic murmur	Any age	Upper-left sternal border	Grade I-II; early systole; blowing; loudest when supine; louder with exercise
Venous hum	Any age	Neck and below clavicles	Continuous murmur, heard when sitting or standing

- **Acyanotic congenital heart diseases:**
  - **Atrial Septal Defect (ASD):**
    - ✓ It is a left-to-right shunt in which blood is flowing from left atrium (area of high resistance) to right atrium (area of low resistance). Increased blood flow to right atrium and right ventricle through this septal defect will result in increased pulmonary blood flow.

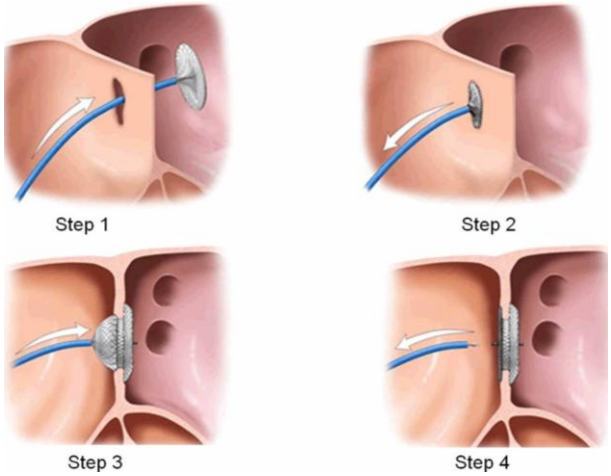


- ✓ Clinical features and physical examination:
  - ❖ Mainly asymptomatic and might not be discovered until adulthood!
  - ❖ There will be a systolic ejection murmur at the upper-left sternal border.
  - ❖ Fixed split second heart sound: because increased pulmonary blood flow will lead to disappearance of the normal variation in closure of aortic and pulmonary valves during respiration.
- ✓ CXR: right atrial and ventricular enlargement + ↑pulmonary vascular markings.



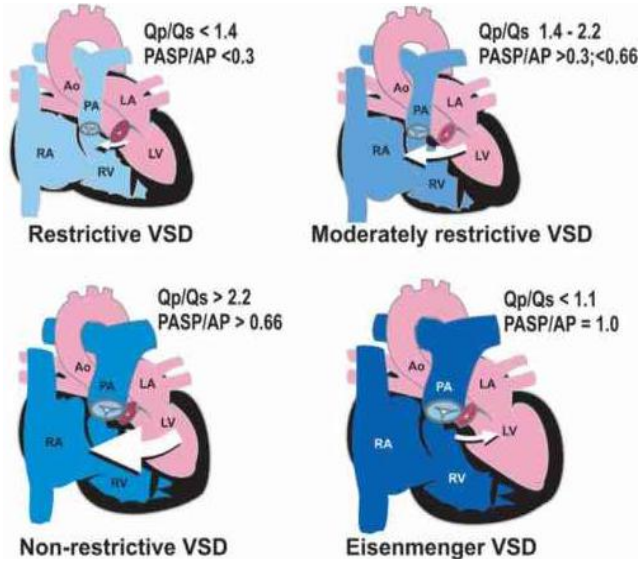


- ✓ Management: closure by open heart surgery to prevent pulmonary hypertension and right-sided heart failure.

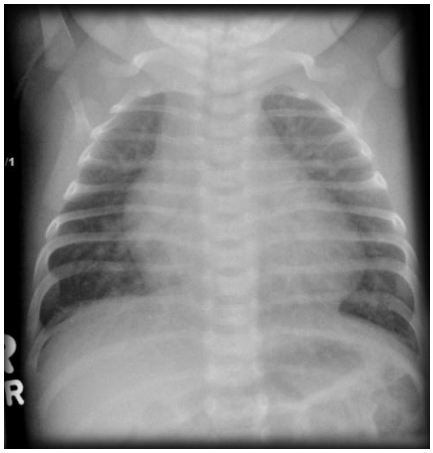


- **Ventricular Septal Defect (VSD):**
  - ✓ Classification: mainly being membranous or muscular (notice that ventricular septum is composed of both a muscular and a membranous parts).
  - ✓ It is a left-to-right shunt in which blood is flowing from left ventricle (higher resistance) to right ventricle (lower resistance). Thus pulmonary blood flow will increase resulting eventually in pulmonary hypertension. Notice that in moderate and large VSDs chronic pulmonary resistance might result in irreversible pulmonary hypertension that changes the shunt to become right-to-left (this is known as Eisenmenger syndrome).
  - ✓ Amount of blood flowing through the shunt depends on: size of VSD and degree of pulmonary vascular resistance.
  - ✓ Clinical features:

Small VSDs	Usually close spontaneously; grade-IV holosystolic murmur at the lower-left sternal border
Moderate VSDs	May result in signs and symptoms of CHF; a holosystolic murmur is usually present
Large VSDs	Often results in signs and symptoms of CHF; systolic murmur is shorter and lower in pitch.

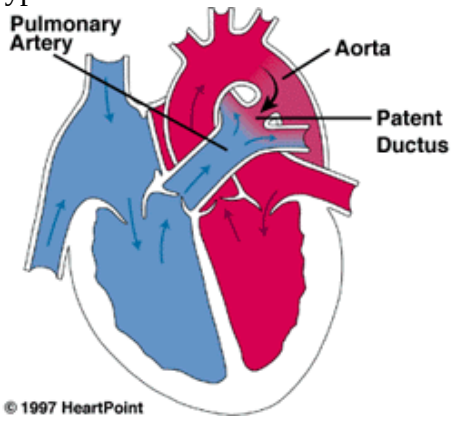


- ✓ CXR: cardiomegaly and ↑pulmonary vascular markings (with moderate/large VSDs).



- ✓ Management:
  - ❖ *If there are signs and symptoms of CHF* → cardiac glycosides and loop diuretics.
  - ❖ *Small-moderate VSDs*: closed surgically between 2-6 years of age.
  - ❖ *Large VSDs with pulmonary hypertension*: closed surgically between 2-6 months of age.

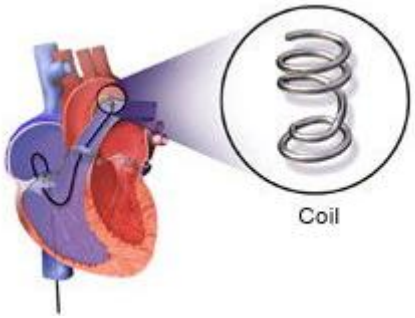
- **Patent Ductus Arteriosus (PDA):**
  - ✓ Ductus arteriosus is a connection between aorta and pulmonary arteries which is supposed to be closed after birth and converted to ligamentum arteriosum. If it remains open this is called PDA and it is more common in premature infants.
  - ✓ It is a left-to-right shunt in which blood flows from aorta –through PDA- to pulmonary arteries thus increasing pulmonary blood flow and risk of pulmonary hypertension.



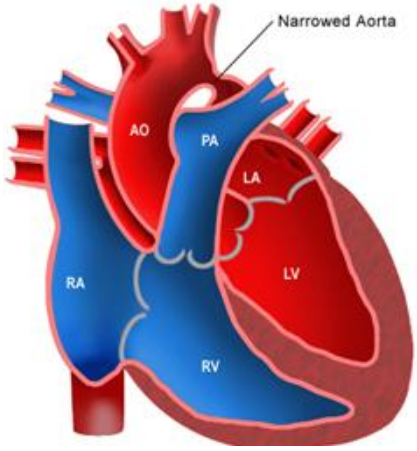
- ✓ Clinical features and physical examination:

Small PDA	Asymptomatic
Moderate-large PDAs	Signs and symptoms of CHF due to increased pulmonary blood flow
Physical examination	Machinery-like continuous murmur at upper-left sternal border

- ✓ CXR: cardiomegaly with increased pulmonary vascular markings.
- ✓ Management: indomethacin (closing PDA medically in prematures) or surgical closure (coil embolization).



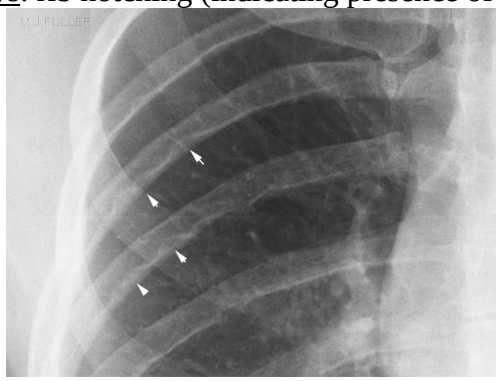
- **Coarctation of aorta:**
  - ✓ There is a narrowing of the aortic arch below the origin of left subclavian artery resulting in decreased blood flow from proximal to distal aorta.



- ✓ Clinical features and physical examination:

Neonates and infants	Those with severe coarctation of aorta will depend on a right-to-left shunt through PDA for perfusion or distal aorta. When PDA closes, signs and symptoms of CHF will develop.
Older children	↑BP in right arm and ↓BP in lower extremities; radio-femoral delay, aortic stenosis with systolic murmur is present in 50% of patients; bruit of turbulence through coarctation may be heard at left upper back near scapula.

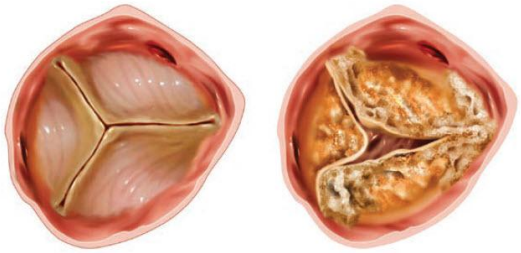
- ✓ CXR shows: rib notching (indicating presence of collateral blow flow).



- ✓ Management:

Medical	Prostaglandin E to keep ductus arteriosus open in neonates
Surgery	• Removal of narrowed segment with end-to-end anastomosis. Recurrence = 50% • Balloon angioplasty is the therapy of choice for recurrent coarctation.

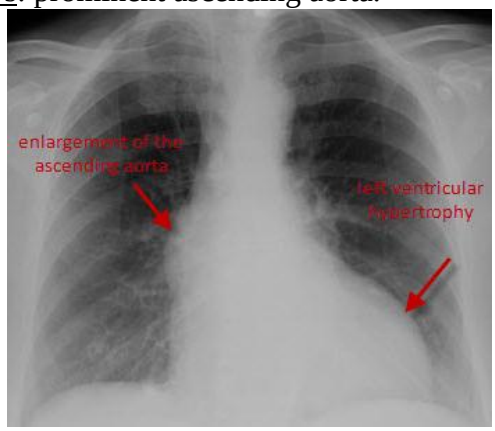
- **Aortic stenosis:**
  - ✓ It is defined as narrowing of the aortic valve in which the valve becomes bicuspid or unicuspid (calcified).
  - ✓ Aortic stenosis results in reduced left ventricular output which may predispose to myocardial ischemia due to reduced blood flow through coronary arteries which are supplying the heart will blood and oxygen.



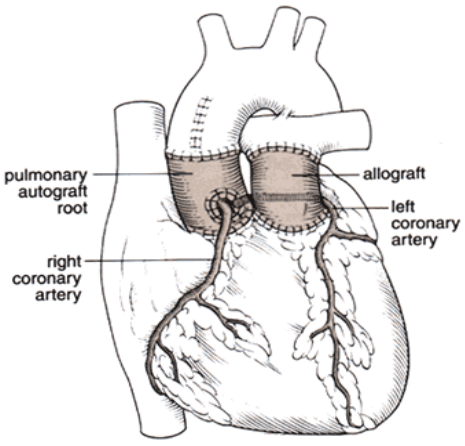
  - ✓ Clinical features and physical examination

Neonates with severe aortic stenosis	Developing signs and symptoms of CHF 12-24 hours after birth
Older children	Asymptomatic until stenosis is severe causing: exercise intolerance, chest pain, syncope and death
Physical examination	Systolic ejection murmur at upper-right sternal border

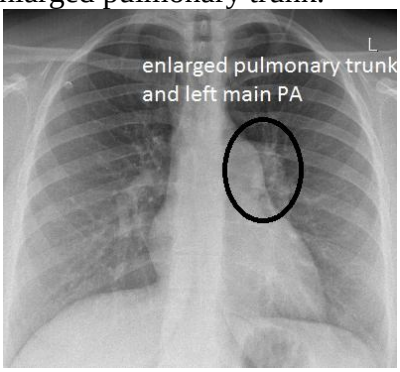
  - ✓ CXR shows: prominent ascending aorta.



  - ✓ Management:
    - ❖ Initially palliative balloon valvuloplasty for aortic stenosis without insufficiency.
    - ❖ After 5-10 years, surgery for aortic stenosis with insufficiency using patients own pulmonary valve (Ross procedure) or prosthetic valve.

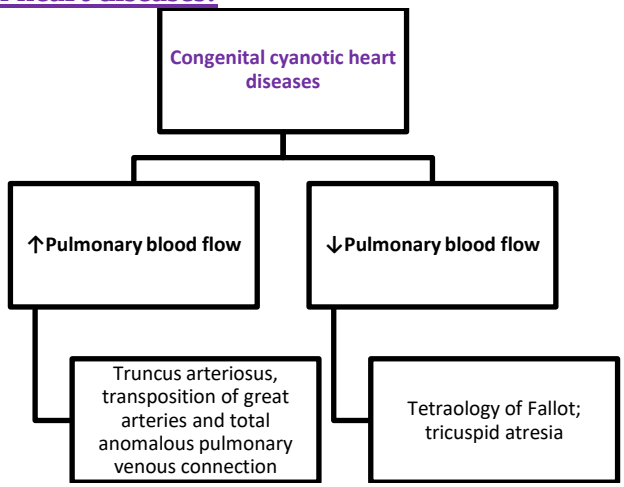


- **Pulmonary stenosis:**
  - ✓ It is defined as narrowing of the pulmonary valve which results in decreased right ventricular output.
  - ✓ Clinical features: asymptomatic unless severe in neonates where there will be cyanosis due to right-to-left shunt through from right atrium to left atrium through patent foramen ovale. On physical examination, you will hear a systolic ejection murmur at the upper-left sternal border.
  - ✓ CXR shows: enlarged pulmonary trunk.

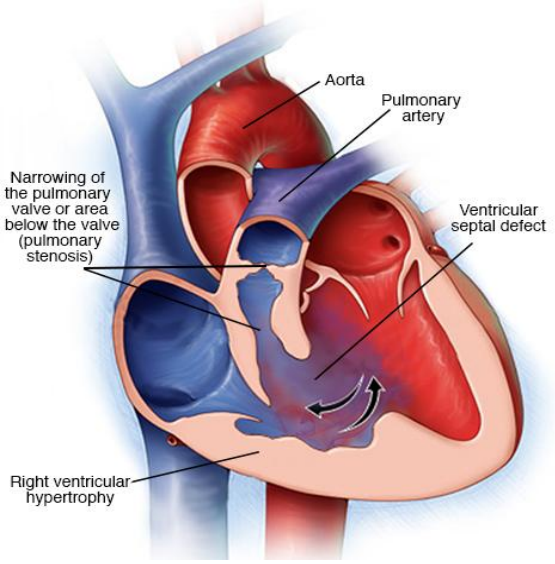


  - ✓ Management: balloon valvuloplasty when symptomatic.

- **Cyanotic congenital heart diseases:**



- **Tetraology of Fallot (TOF):**
  - ✓ It is the most common cause of central cyanosis after neonatal period and is characterized by:
    - ❖ *Pulmonary stenosis.*
    - ❖ *Right ventricular hypertrophy.*
    - ❖ *Overriding aorta.*
    - ❖ *VSD.*
  - ✓ Due to pulmonary stenosis, there is a right-to-left shunt in which blood is flowing from right ventricle –through the VSD- to left ventricle (mixing with oxygenated blood) and being pumped through the overriding aorta.

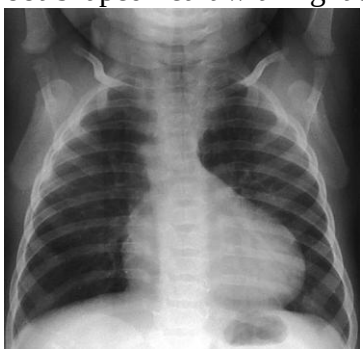


  - ✓ Clinical features and physical examination:
    - ❖ Systolic ejection murmur at the upper-left sternal border; increased right ventricular impulse and cyanosis.
    - ❖ Cyanosis results with conditions that decrease systemic vascular resistance (e.g. exercise, vasodilation and volume depletion) but

decrease with conditions that increase systemic vascular resistance (e.g. volume infusion, hypertension and valsalva maneuver).

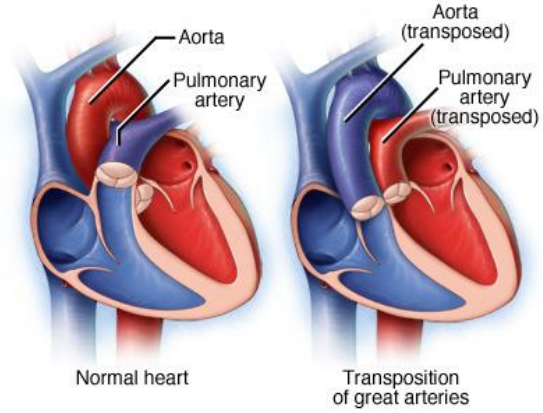
- ❖ TOF hypercyanotic or "tit" spells: this condition is characterized by sudden onset of cyanosis and decreased murmur intensity. It occurs when there is ↓oxygen saturation → baby cries and becomes irritable → ↑ right-to-left shunt. Baby will compensate by squatting (knee-chest position) which is increasing systemic vascular resistance thus decreasing the shunt.

✓ CXR shows: a boot-shaped heart with right aortic notch.



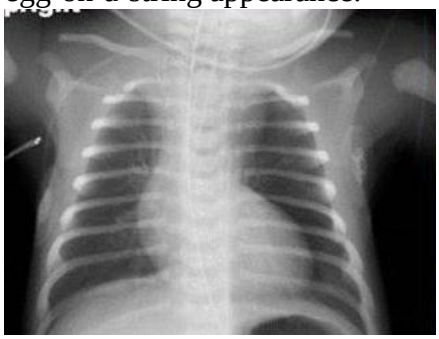
✓ Management: complete surgical repair at 4-8 months of age.

- **Transposition of great arteries:**
  - ✓ It is defined as aorta which is arising from right ventricle and pulmonary trunk which is arising from left ventricle.
  - ✓ There has to be a shunting of blood from one circulation to the other through: PDA, ASD, patent foramen ovale or VSD. Notice that id transposition of great arteries is accompanied by dextrocardia → the condition will be corrected!



✓ Physical examination: central cyanosis, single S2 and no murmur.

✓ CXR shows: egg-on-a-string appearance.

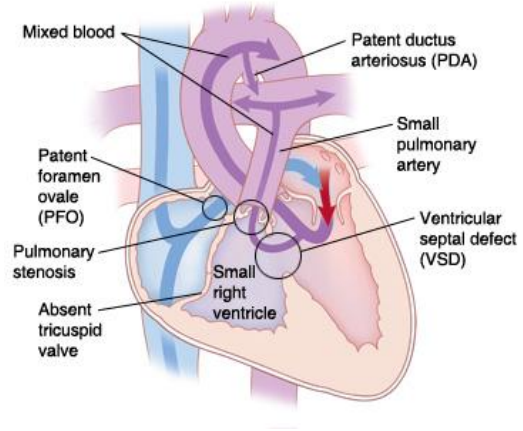


✓ Management:

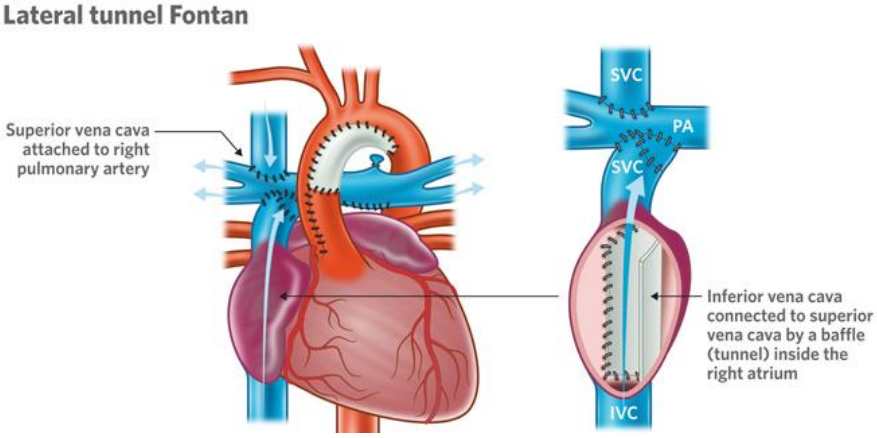
Neonates	Palliative management by giving PGE to keep ductus arteriosus open or emergency atrial balloon septostomy to increase the size of ASD/PFO
Definitive treatment	Arterial switch operation

- **Tricuspid atresia:**
  - ✓ It is characterized by the presence of a plate of tissue in the floor of the right atrium instead of tricuspid valve. An ASD or Patent Foramen Ovale (PFO) is always present.
  - ✓ <u>Pathophysiology:</u>

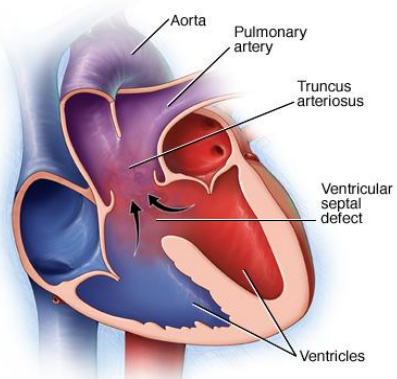
VSD NOT present	• Pulmonary atresia will occur because there is no blood reaching the right ventricle and flowing through the pulmonary trunk. • A PDA must be present to allow flow of blood through pulmonary arteries to the lung. As it closes after birth, visible cyanosis will occur • Patients have single S2 and no murmur
VSD PRESENT	• Blood will flow from left ventricle to the right ventricle through the VSD and subsequently to the lungs through pulmonary trunk. Therefore, providing adequate saturation. • Patients have holosystolic murmur at lower-left sterna borders.



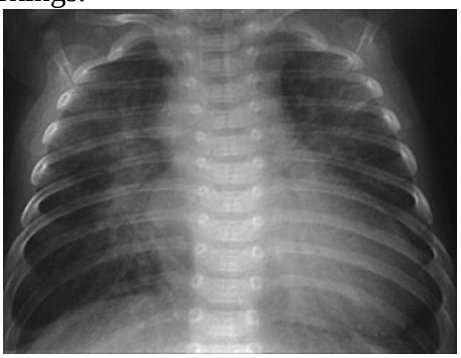
  - ✓ <u>Management</u>: Fontan procedure (3-6 years) in which blood flow from IVC will be directed directly into pulmonary trunk.



- **Truncus arteriosus:**
  - ✓ It is a congenital cyanotic heart disease in which there is no septum separating the aorta from pulmonary trunk and they both appear as one large vessel (the truncus). Therefore, oxygenated and deoxygenated blood will mix together and there will be excessive blood flow to the lungs eventually resulting in signs and symptoms of CHF.

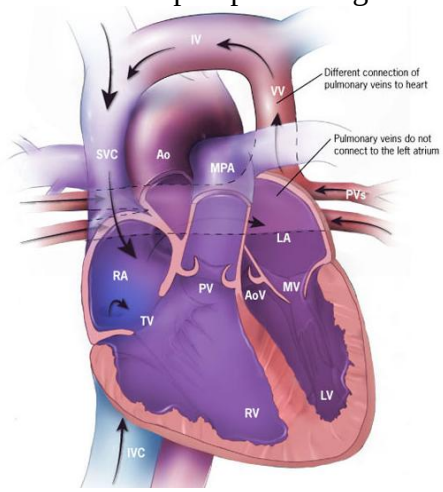


- ✓ Physical examination: systolic ejection murmur along the left sternal border; single S2; diastolic murmur at the apex (resulting from excessive pulmonary blood flow going back to the left atrium through the mitral valve).
- ✓ CXR shows: enlarged heart, right aortic notch and increased pulmonary vascular markings.

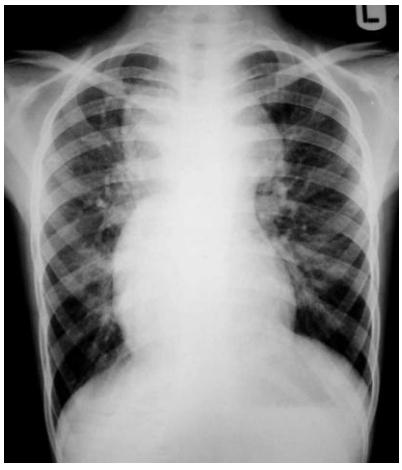


- ✓ Management: treatment of CHF with cardiac glycosides and loop diuretics; closure of VSD and homograft placement between right ventricle and pulmonary trunk.

- **Total Anomalous Pulmonary Venous Connection (TAPVC):**
  - ✓ It is a congenital cyanotic heart disease in which pulmonary veins drain into systemic venous system (such as right atrium or SVC) instead of draining into the left atrium. Therefore, mixing of oxygenated and deoxygenated blood will occur and then it will pass from right atrium –through ASD or PFO- to the left atrium and ventricle to be pumped through the aorta.



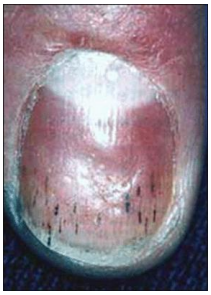
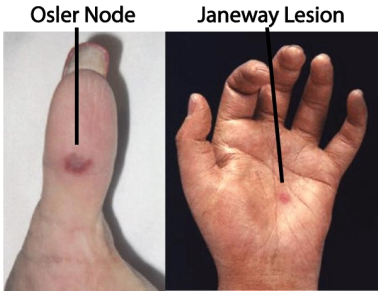
  - ✓ Physical examination: pulmonary ejection murmur along the left sternal border.
  - ✓ CXR shows: snow-man appearance with increased pulmonary vascular markings.

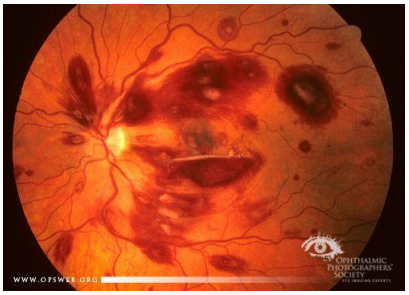


  - ✓ Management: surgical repair in which pulmonary veins will be connected to the posterior aspect of left atrium and ASD or PFO will be closed.

- **Acquired heart disease:**
  - **Infective endocarditis:**
    - ✓ It is a microbial infection of the internal surface of the heart.
      - ❖ *In 80% of cases* → there is a structural abnormality of the heart.
      - ❖ *In 50% of cases* → it occurs after cardiac surgery.
    - ✓ Causative organisms: α-hemolytic streptococcus (Streptococcus viridians) and Staphylococcus species. The organism will be introduced to the blood and then affect the injured cardiac endothelium. Subsequently, fibrin and platelets will adhere creating vegetations that might cause valve incompetency and embolic phenomena.
    - ✓ Clinical features:

Fever	The most common symptom
Splinter hemorrhages	Linear hemorrhages beneath the nails
Osler's nodes	Small, pink, swollen, tender lesions on palms or soles
Janewy lesions	Small, erythematous hemorrhagic lesions on palms or soles
Roth's spots	Round white spots seen in the retina

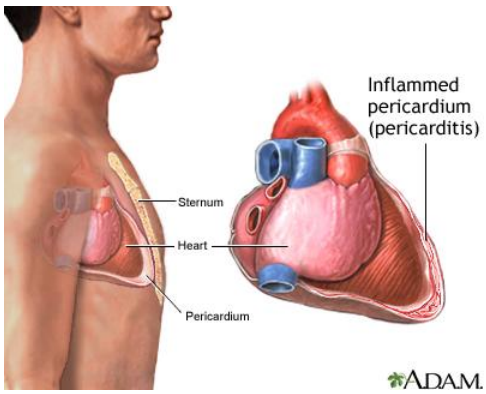





    - ✓ Investigations: blood culture (most important); ↑ESR; ↑RF (50% of patients); transesophageal echocardiography (more sensitive in detecting vegetations).
    - ✓ Management: IV antibiotics directed against the identified organism (for 4-6 weeks). Antibiotic prophylaxis is given for:
      - ❖ All patients with structural heart abnormalities (except those with secundum ASD).
      - ❖ All patients after having a cardiac surgery (for 6 months after the repair).
  - **Pericarditis:**
    - ✓ It is inflammation of pericardial space.
    - ✓ Causes:

Infection	Viral (most common): coxsackievirus or EBV
	Purulent pericarditis = bacterial infection: S.aureus or S.pneumoniae
Collagen vascular disease	SLE
Uremia	-
Postpericardiotomy syndrome	Inflammatory response after cardiac surgery

    - ✓ Pathophysiology: inflammation of pericardial layers (visceral and parietal) will result in transudation/exudation of fluid into the pericardial space that leads to reduced venous return and filling of the heart. Cardiac temponade may occur (fluid building up in pericardium resulting in cardiac compression).

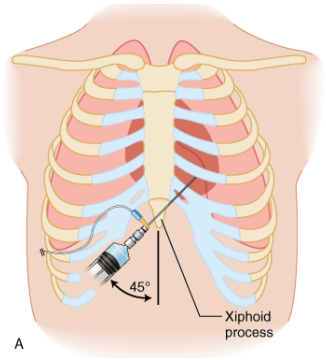


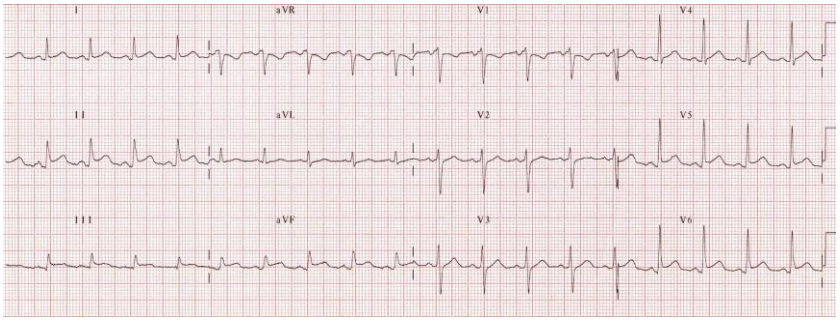
✓ Clinical features:

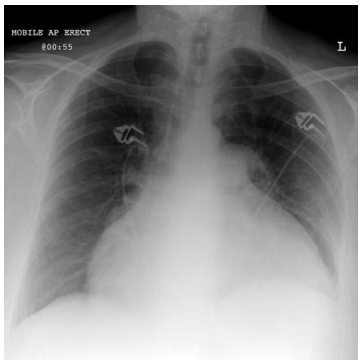
Symptoms	Fever, dyspnea and chest pain (which becomes more severe in supine position and relieved when sitting upright)
Physical examination	Pericardial friction rub, distal heart sounds (if effusion is large), pulses paradoxus (> 10 mmHg reduction in systolic blood pressure with deep inspiration) and hepatomegaly

✓ Diagnosis:

Pericardiocentesis	A needle is inserted into the pericardial space and fluid is aspirated (this is both diagnostic and therapeutic)
ECG	Low-voltage QRS complex in patients with large pericardial effusion
CXR	Enlarged cardiac shadow with large effusion
ECHO	Demonstrates extent and quality of pericardial effusion







✓ Management:

Bacterial pericarditis	Appropriate antibiotics
Viral pericarditis	Anti-inflammatory agents (e.g. aspirin)

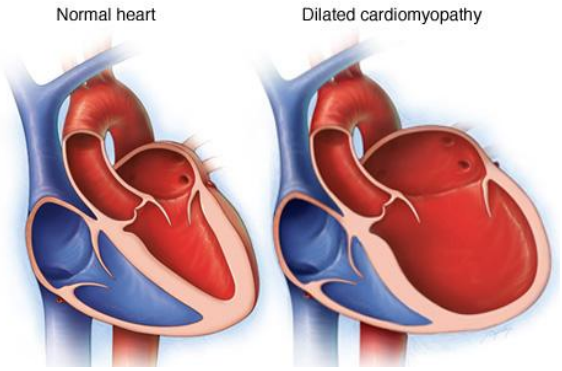
- **Myocarditis:**
  - ✓ It is inflammation of myocardium which is characterized by: cellular infiltrate and myocardial cell death. It is one of several common causes of sudden death in young athletes.
  - ✓ Causes:

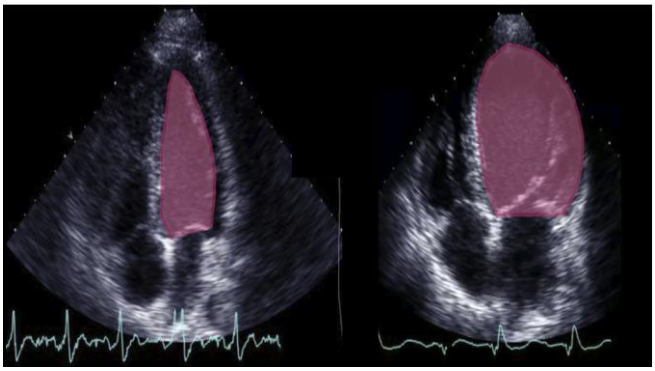
Viruses	Coxsackievirus
Bacteria	S.aureus or S.pyogens
Fungi	Candida or Cryptococcus
Protozoa	T.cruzi (Chagas' disease)
Autoimmune	SLE
Kawasaki disease	-

  - ✓ Clinical features:
    - ❖ It is usually preceded by viral or flu-like illness.
    - ❖ There is dyspnea and malaise.
    - ❖ Physical examination shows: resting tachycardia, muffled heart sounds, gallop heart rhythm and hepatomegaly.

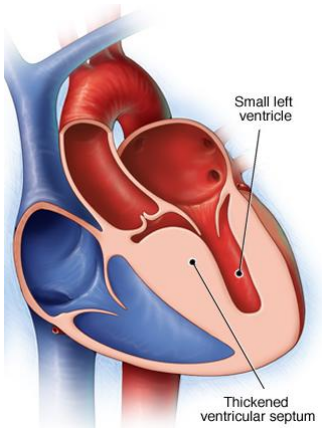
- ✓ Investigation:
  - ❖ ↑ESR, ↑CRP, ↑creatinine kinase MB fraction.
  - ❖ Endomyocardial biopsy can be obtained with PCR to detect the virus.
  - ❖ ECHO: global ventricular dysnsfunction.
- ✓ Management: supportive with treatment of CHF.

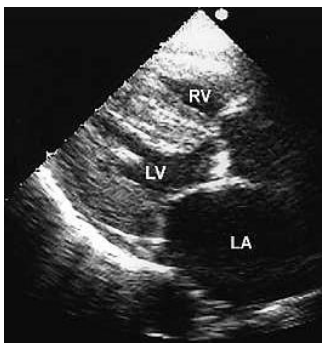
- **Cardiomyopathies:**
  - **Dilated cardiomyopathy:**
    - ✓ It is characterized by ventricular dilation and reduced cardiac output.
    - ✓ Cause: idiopathic but can be associated with viral myocarditis, mitochondrial abnormalities, carnitine deficiency, nutritional deficiency (thiamine or selenium), hypocalcemia and medications (e.g. doxorubicin).
    - ✓ Clinical features: signs and symptoms of CHF.
    - ✓ Investigations:
      - ❖ Viral serologies and carnitine level.
      - ❖ *ECG*: sinus tachycardia, low cardiac voltage and ST-segment and T-wave changes.
      - ❖ *ECHO*: dilated left ventricle with poor ventricular function.





    - ✓ Management:
      - ❖ Treatment of CHF.
      - ❖ Treatment of underlying metabolic or nutritional problem.
      - ❖ Surgical repair of ALCAPA (Anomalous origin of Left Coronary Artery from Pulmonary artery).
      - ❖ Cardiac transplantation if CHF is unresponsive to medical therapy.
  - **Hypertrophic cardiomyopathy:**
    - ✓ It is defined as left ventricular hypertrophy in which the typical anatomic lesion is asymmetric septal hypertrophy. It is the most common cause of sudden death in athletes.
    - ✓ Cause: AD in 60% of cases.
    - ✓ Pathophysiology:
      - ❖ Poor ventricular filling.
      - ❖ Left ventricular outflow tract obstruction because mitral valve presses against the septum during systole leading to obstruction of blood flow.
      - ❖ Mismatch between myocardial oxygen demand and supply (owing to hypertrophy) may result in myocardial ischemia.
    - ✓ Clinical features: there might be no symptoms until syncope or sudden death occurs or patient might suffer from exercise intolerance and chest pain. Physical examination reveals a harsh systolic ejection murmur at the apex.





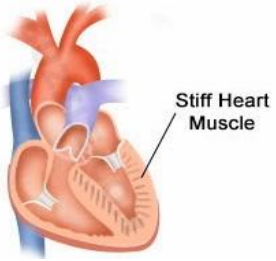
✓ Diagnosis:

ECG	LVH; ST-segment and T-wave changes
ECHO	Hypertrophy

✓ Management:

Medical	• B-adrenergic blockers or calcium-channel blockers: reducing left ventricular outflow tract obstruction • Anti-arrhythmic drugs because ventricular dysrhythmias are common
Surgery	Myomectomy done for patients with severe obstruction which is not responding to medical treatment

- **Restrictive myopathy:**
  - ✓ It is characterized by excessively rigid ventricular walls which impair normal diastolic filling of the heart.

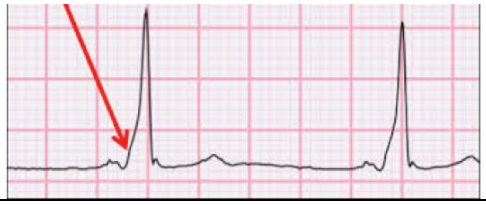


  - ✓ Causes: amyloidosis; inherited infiltrative disorders (Gaucher disease, hemochromatosis or hemosiderosis).
  - ✓ Clinical features:

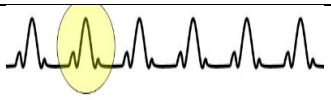
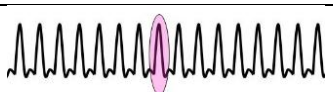
Symptoms	Exercise intolerance, dyspnea and weakness
Physical examination	Edema, ascites and hepatomegaly

  - ✓ Management: diuretics; β-clockers or calcium-channel blockers.

- **Dysrhythmias:**
  - **Supraventricular tachycardia:**
    - ✓ It is the most common type of dysrhythmia in childhood in which there is accelerated heart rhythm originating proximal to the bifurcation of bundle of his.
    - ✓ There are two type of supraventricular tachycardia:

Atrio-Ventricular Re-entrant Tachycardia (AVRT)	Retrograde conduction through an accessory pathway
Atrio-Ventricular Node Re-entrant Tachycardia (AVNRT)	Conduction abnormality occurs in different pathways within the AV node itself
Wolff-Parkinson-White syndrome	Characterized by the presence of delta wave and can result in sudden death 

    - ✓ Comparison between sinus tachycardia and supraventricular tachycardia:

Feature	Sinus tachycardia	Supraventricular tachycardia
Rate (beats/min)	< 230 (in newborns) < 210 (in children)	> 250
P-wave	Present and normal	Absent
Predisposing factors	Fever, infection and anemia	None
Response to adenosine	Gradual	Rapid
ECG		

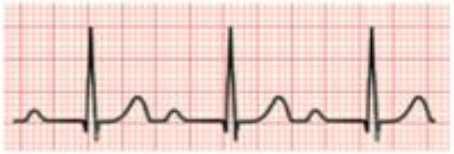
- ✓ Clinical features: palpitations, chest pain, dyspnea and altered level of consciousness.
- ✓ Management:

Vagal maneuvers	Valsalva, placement of ice-pack on face, unilateral carotid message or placing the child upside down
IV adenosine	For conversion to sinus rhythm (other drugs which can be used include propranolol or digoxin)

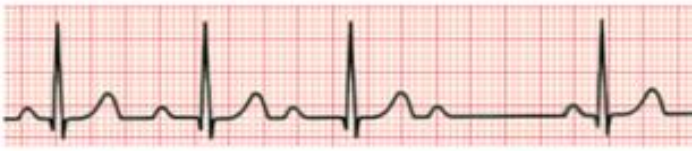
- **Heart block:**
  - ✓ It is defined as delayed or interrupted conduction of impulses from atria to ventricles.
  - ✓ Types:

Type	Characteristic(s)
First-degree	Prolongation of PR interval
Second-degree	Type-I: progressive prolongation of PR interval with sudden drop of QRS complex
	Type-II: sudden drop of QRS complex without prolongation of PR interval
Third-degree	Complete block with no conduction of atrial impulses to ventricles. It occurs in children born to mothers with SLE

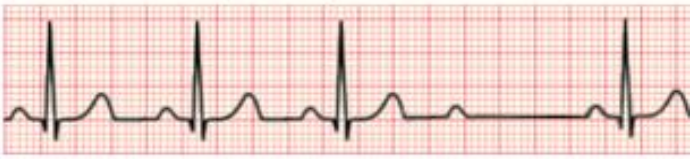
First degree AV block



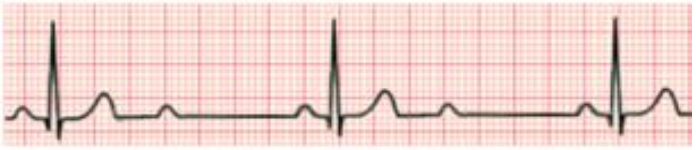
Second degree AV block (Mobitz I or Wenckebach)



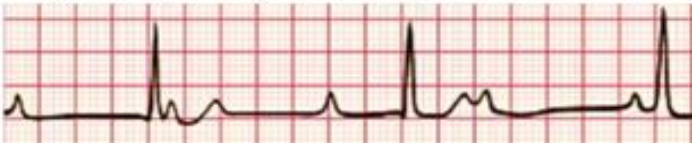
Second degree AV block (Mobitz II)



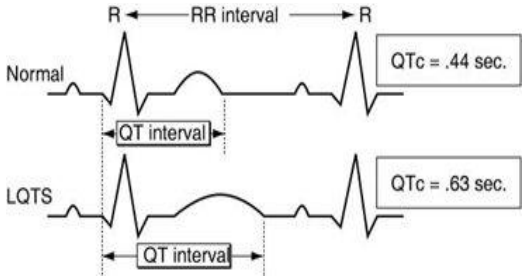
Second degree AV block (2:1 block)

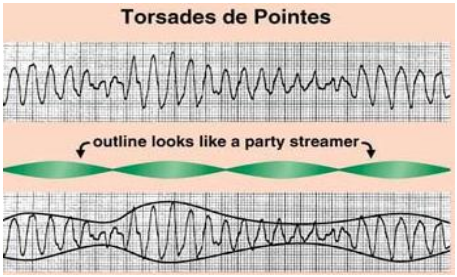


Third degree AV block with junctional escape



  - ✓ Management: cardiac pacemaker.
- **Long QT syndrome:**
  - ✓ It is defined as prolongation of QT interval which increases the risk of a lethal ventricular arrhythmia known as “torsades de pointes”.





- ✓ Causes:

Inheritance (50% of cases)	AD (Romano-Ward syndrome): not associated with deafness
	AR (Jervell-Lange-Nielsen syndrome): associated with deafness
Drugs	Tricyclic antidepressants or erythromycin
Idiopathic	

- ✓ Clinical features: syncope (most common) or sudden cardiac arrest.
- ✓ Management: β-blockers