CYANOTIC GROUP

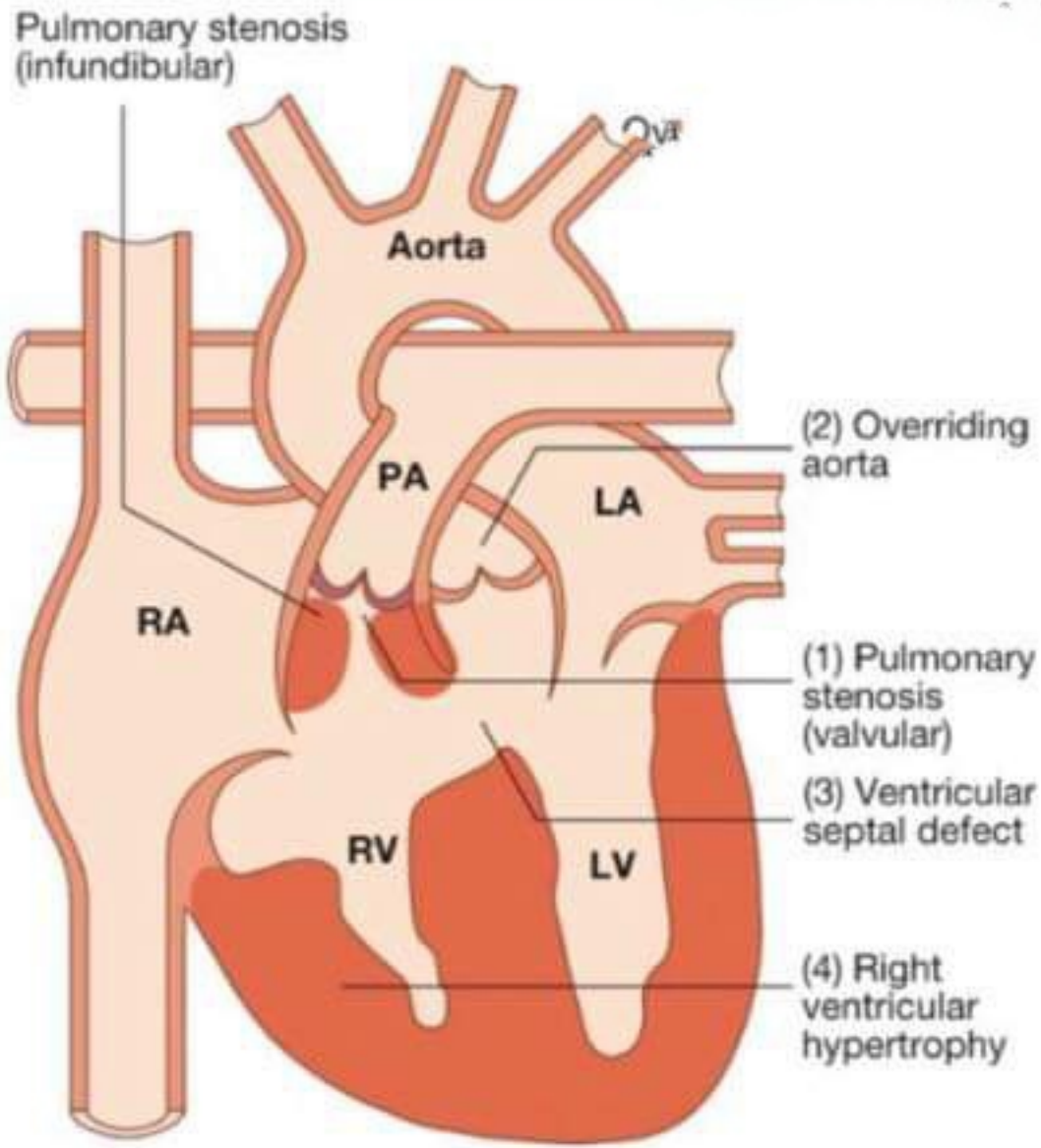
TETRALOGY OF FALLOT

Fb/Nurse-Info

TOF

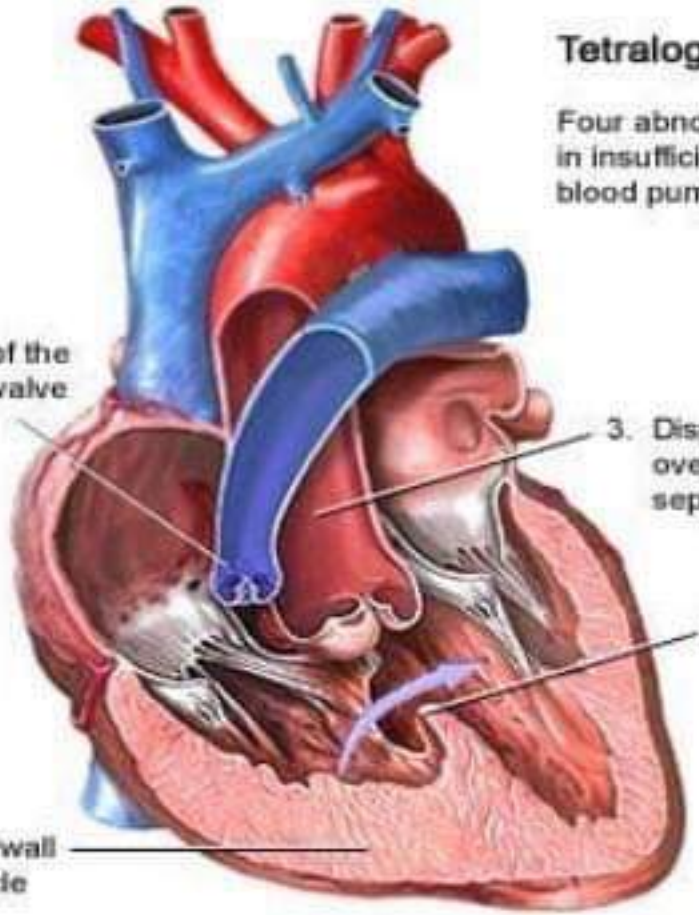
- Tetralogy of Fallot is the most common **cyanotic** congenital heart disease, found in about 1 in 2000 births.
- Classically there are 4 defects:
  1. Ventricular septal defect
  2. Pulmonary stenosis
  3. Right ventricular hypertrophy
  4. Overriding aorta

TOF


Pulmonary stenosis
(infundibular)
Aorta
PA
LA
RA
RV
LV
(2) Overriding
aorta
(1) Pulmonary
stenosis
(valvular)
(3) Ventricular
septal defect
(4) Right
ventricular
hypertrophy

Tetralogy of Fallot

Four abnormalities that results in insufficiently oxygenated blood pumped to the body



1. Narrowing of the pulmonary valve
2. Thickening of wall of right ventricle
3. Displacment of aorta over ventricular septal defect
4. Ventricular septal defect- opening between the left and right ventricles

A.D.A.M.

DEFINITION:

This condition is characterized by the combination of four defects:

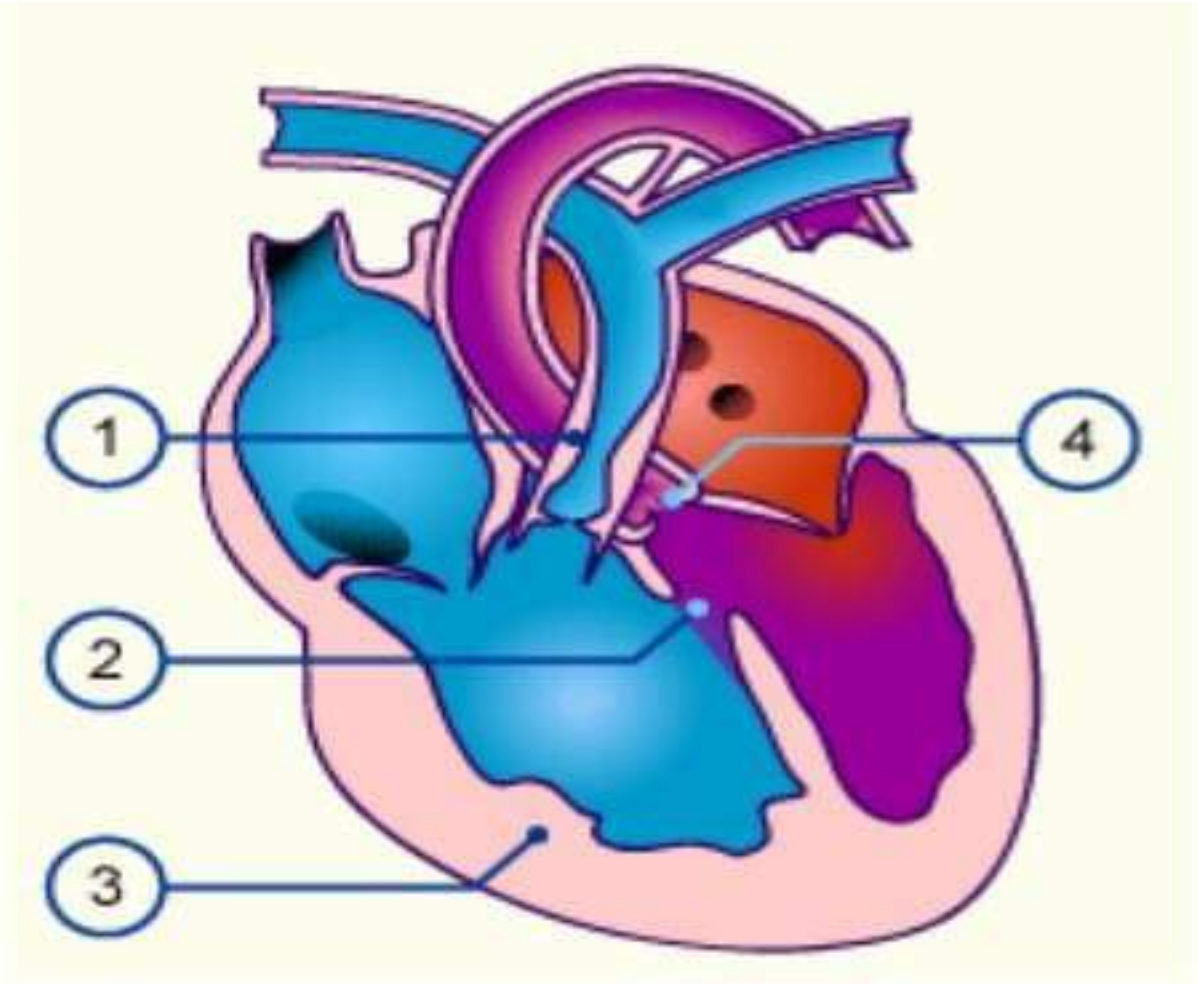
1. Ventricular septal defect (VSD)
2. Pulmonic stenosis
3. overriding or dextroposed aorta, and
4. Right ventricular hypertrophy

INCIDENCE:

- It is the most common cyanotic heart defect, and the most common cause of blue baby syndrome.
- It accounts for 6-10 percent all CHDs

Causes:

- Not clear at this time
- Children born to mothers with PKU are highly susceptible, as are mothers who drink during birth.
- Genetic


1
2
3
4

Pathophysiology:

Physiologically the pulmonic stenosis causes concentric right ventricular hypertrophy without cardiac enlargement and an increase in right ventricular pressure

when the right ventricular pressure is as high as the left ventricular or the aortic pressure, a right to left shunt appears to decompress the right ventricle

once the right and left ventricular become identical, increasing severity of pulmonic stenosis reduces the flow of blood into the pulmonary artery and increases the right to left shunt

as the systolic pressures between two ventricle are identical there is little or no left to right shunt and the **VSD is silent**

- the flow from the right ventricle into the pulmonary artery occurs across the pulmonic stenosis producing an **ejection systolic murmur**

- more severe the pulmonic stenosis, the less the flow into the pulmonary artery and the bigger the right to left shunt more the cyanosis

- thus the severity of cyanosis is directly proportional to the severity of pulmonic stenosis

- The VSD of TOF is always large enough to allow free exit to the right to left shunt

congestive failure never occurs in TOF.

CLINICAL MANIFESTATIONS:

- It depends upon the size of VSD and degree of right ventricular flow obstruction.
- Blue baby (cyanosis of lips and nail beds with dyspnea is found initially with crying and exertion)
- Tired easily with exertion.
- May have difficulty in feeding.

CONTD…

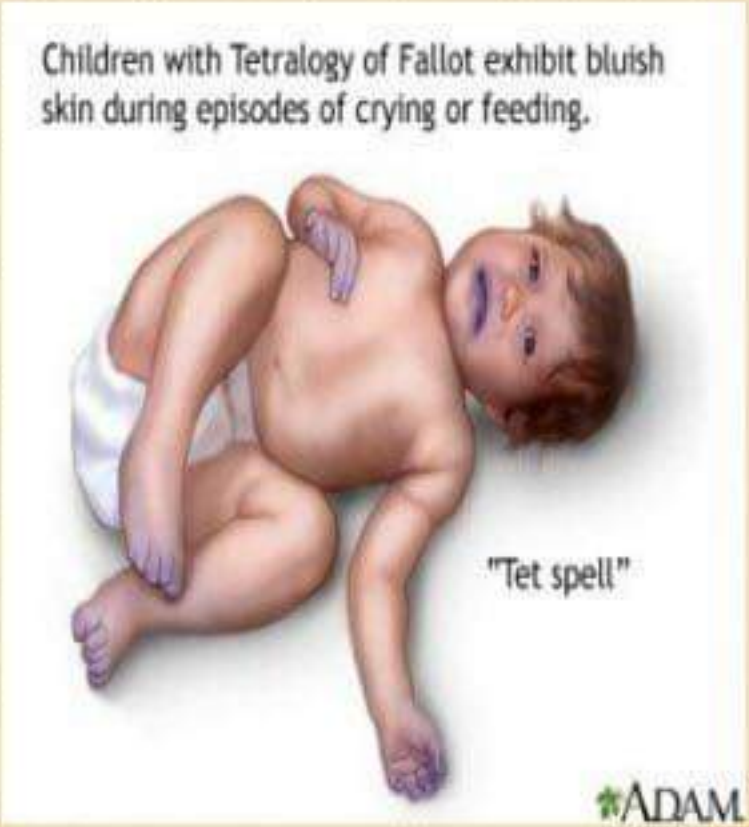
- Normal growth and development depend on a normal Babies who have tetralogy of Fallot may not gain weight or grow as quickly as children who have healthy hearts because they **tire** easily while feeding.

- **Squatting** (a compensatory mechanism) is uniquely characteristic of a right-to-left shunt that presents in the exercising child. Squatting **increases the peripheral vascular resistance**, which diminishes the right-to-left shunt and increases pulmonary blood flow.

- Sign of chronic hypoxemia.
- Harsh systolic murmur, often accomplished by a palpable thrill.
- In radiograph the heart is boot-shaped(i.e because the poor development of pulmonary artery).

TET SPELLS:

Tet spells (hyper cyanotic spells)due to cerebral anoxia – it consists of irritability, dyspnea, cyanosis, flaccidity with or without unconsciousness. It is found in the morning after awakening, during or after feeding and painful procedures.



Causes of spell:

- Due to "spasm" or contraction of a band of muscle in the right ventricle just under the pulmonary valve. When this muscle contracts, it further narrows the channel for blood flow into the lungs. As a result, oxygen delivery becomes further reduced. This causes a spell

CLUBBING:

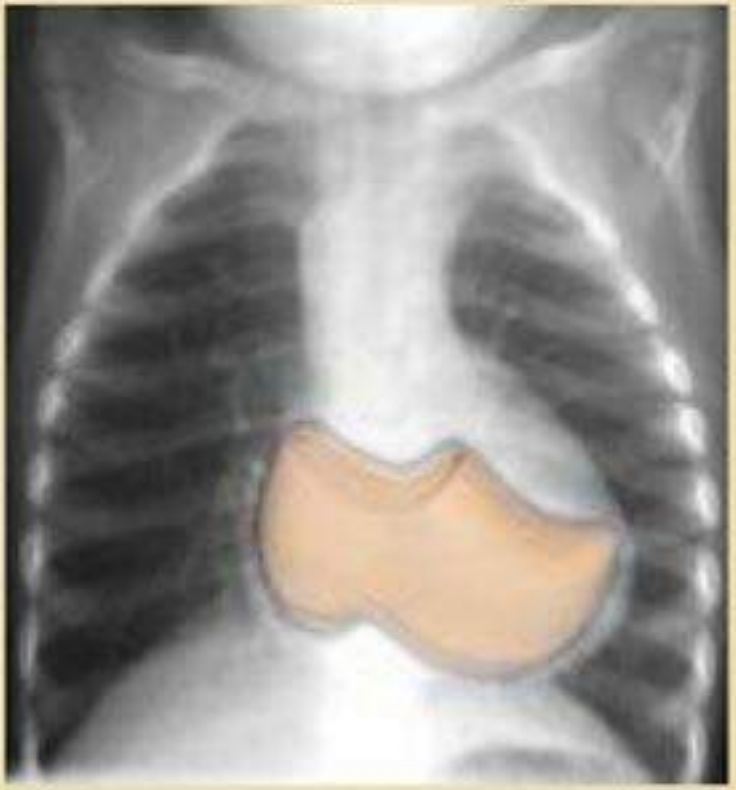
Clubbing of fingers and toes

DIAGNOSTIC EVALUATION:

- History collection
- Physical examination
  - Cyanosis
  - Clubbing,

- Heart sound-Normal first sound
- Single second sound and
- An ejection systolic murmur.
- ECG- right axis deviation and right ventricular hypertrophy
- Echocardiography : identify the large overriding aorta, right ventricular hypertrophy and outflow obstruction.
- Cardiac catheterization

- **Boot shaped heart** (it means apex is lifted up & there is a concavity in the region of pulmonary artery)
- **Oligaemic lung** fields
- Hilar vessels are few, lung vessels also few, large rt. Ventricle.



Chest X Ray:

Medical management:

- Monitoring for hypoxemia.
- Hemoglobin levels and hematocrit values may be evaluated to assess the anaemia.
- Monitoring for hypercynotic episodes. (10-15minutes).
- Balloon dilatation of R.V outflow tract.

- beta-blockers such as propranolol
- acute episodes may require rapid intervention with morphine to reduce ventilatory drive and a vasopressor such as epinephrine, phenylephrine, or norepinephrine to increase blood pressure.

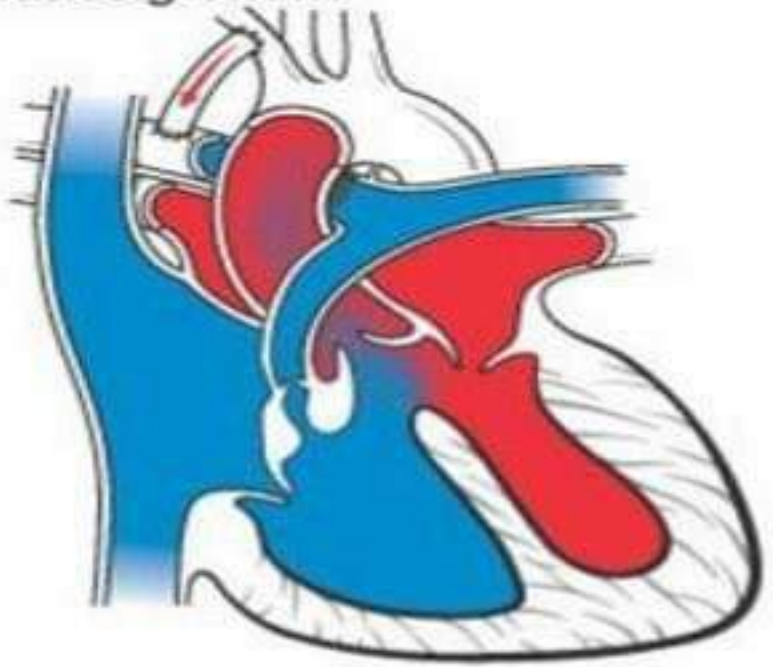
Surgical management:

- **Blalock-Taussig operation** : connection between the right subclavian artery, and the right pulmonary artery, which increases the amount of oxygenated blood reaching the lungs, relieving cyanosis.
- **Pott`s shunt** : descending aorta is anastomosed to the pulmonary artery
- **Waterstont`s shunt** : ascending aorta right pulmonary artery anastomosis

Treatment for spells:

- **Knee chest position** to increase aortic resistance. The increased aortic and left ventricular pressure reduces the rush of blood through the septal hole from the right ventricle and improves blood circulation to the lungs
- **Oxygen**

Tetralogy of Fallot with Modified Blalock-Taussig Shunt



- **Total correction**: The hole in the ventricular septum is closed with a patch and the obstruction to right ventricular outflow, pulmonic stenosis, is opened.
- These corrections allow blood flow to the lungs for oxygenation before being pumped out into the body.

COMPLICATIONS:

- Brain abscess
- Bacterial endocarditis
- Ventricular arrhythmias

THANK YOU