

TETRALOGY OF FALLOT

TOF is the **most common cyanotic** CHD, representing \approx **10%** of all CHD. It consists from:- 1. **Obstruction to right ventricular outflow** (pulmonary stenosis at **both**

right ventricular infundibulum & pulmonary valve). 2. **Ventricular septal defect** which usually **large** and nonrestrictive. 3. **Dextroposition of the aorta** which override of the ventricular septum.

4. **Right ventricular hypertrophy** due to \uparrow R.V. pressure. **Note:** *When aorta overrides >50% of the VSD & associated with subaortic conus, this is called "double-outlet right ventricle".*

C.M. The **degree of right ventricular outflow obstruction** determines;

the onset of symptoms, severity of cyanosis, & the degree of R.V. hypertrophy.

☐ **Mild to moderate obstruction** \rightarrow **balanced** shunt or even **Lt to Rt shunt** across the VSD, thus patient may present **initially** with symptoms of **HF** "without cyanosis", thus called **"Acyanotic" or "Pink" TOF**. However, as the infant **grow**, there is **increasing hypertrophy** of

the right ventricular infundibulum with increasing obstruction which eventually \rightarrow **Rt to Lt shunt** with **cyanosis later in the 1st yr** of life.

☐ **Severe obstruction** \rightarrow **early Rt to Lt shunt with cyanosis at birth**

which **worsen** when the ductus arteriosus begins to close. Older children with **long-standing cyanosis** may have dusky blue skin, gray sclerae and marked **clubbing** of the fingers and toes. Extracardiac

manifestations also may occur (*see later*).

Dyspnea occurs on exertion. Patient usually play actively for a short

time, then sit or lie down or assume a **"squatting position"**, then resume

playing again within few minutes. The pulse, venous and arterial pressure are usually **normal!**. The left anterior hemithorax may **bulge with impulse & thrill** due to R.V. hypertrophy. The 2nd heart sound is either **single** or the pulmonic component is soft. The **ejection systolic murmur** is caused by **turbulence** through the right ventricular outflow tract, it usually loud and harsh & may be transmitted widely, especially to the lungs, but is most intense at the left

sternal border. It tends to become louder, longer, and harsher as the severity of pulmonary stenosis increases. Infrequently, a **continuous murmur** may be audible due to multiple

Major Aorto-Pulmonary Collateral Arteries (MAPCAs) arising from the ascending and descending aorta to the pulm artery. **Inv.**

☒ **CXR**; the **heart is generally normal in size** but hypertrophied right ventricle causes the rounded apical shadow to be uptilted make the

cardiac silhouette similar to a **boot** or **wooden shoe** "*coeur en sabot*".

The lungs fields are **oligemic** & the aortic arch is **right sided** in **20%** of

cases. ☒ **ECG**; right axis deviation & evidence of right ventricular hypertrophy. A dominant R wave appears in the right precordial chest leads.

☒ **2D Echo** is diagnostic. ☒ **Catheterization** demonstrates a systolic pressure in the right ventricle equal to systemic pressure, whereas the pressure is markedly decreased in the pulmonary artery. ☒ **Selective right ventriculography** can demonstrates the anatomy of TOF.

☒ **Aortography or Coronary arteriography** is important before surgery because **5-10%** may have **aberrant major coronary artery** crosses

over the right ventricular outflow tract (which must not be cut during surgical repair). 2D Echo can also delineate the coronary artery anatomy (although less sensitive). **Cx.**

☒ **Paroxysmal Hypercyanotic, Hypoxic, Blue, or "Tet" Spells:-**

It usually occur in the **1st 2 yr** of life in patient who has **mild cyanosis** at rest because they have **not** yet acquired the homeostatic mechanisms to

tolerate rapid lowering of arterial oxygen saturation e.g. polycythemia. It is manifested as **dyspnea (or gasping respirations), restlessness, increased cyanosis** and may be followed by **syncope, convulsions or hemiparesis, but rarely fatal**. The onset is usually **spontaneous** & unpredictable which last few min to few hrs then usually end by generalized weakness and sleep.

Chest exam during the attack reveal ↓ **intensity of the systolic murmur** (or may **disappear**) due to ↓ blood flow through the right ventricular outflow by **severe infundibular spasm**.

Rx of these spells depends on their severity. Early spells can be **aborted**

by **calming** the infant & **holding** him in knee-chest position on the mother's shoulder & **avoiding** of early blood sampling as it may aggravate the spell. The following steps should be instituted in **following sequence**:- 1. **Placement** of the infant on the abdomen in the knee-chest position &

make certain that infant's cloths are not constrictive. 2. **Oxygen** administration, although it will not reverse the spell. 3. **Morphine** injection, 0.1-0.2 mg/kg SC. 4. **Sodium bicarbonate** to correct acidosis if spell is unusually severe. 5. **Phenylephrine** IV, to ↑ systemic vascular resistance. 6. **Propranolol**, 0.1 mg/kg slowly IV to ↓ infundibular spasm. 7. For spells that are resistant to this therapy, **intubation and sedation**

are often sufficient to break the spell. ☒ **Bacterial Endocarditis**; which need antibiotic Px. ☒ **Cerebral Thrombosis**; it common in patient <2 yr of age, especially when there is **severe dehydration & polycythemia**, although it may occur in patients with **iron deficiency anemia** with Hb levels in the normal range but too low for cyanotic heart disease!. **Rx** is supportive e.g. adequate **hydration** and **phlebotomy** for symptomatic polycythemia with volume replacement by albumin or NS.

☒ **Cerebral Abscess**; it less common & mainly occur in patient >2 yr; the

onset is usually **insidious** as low-grade fever or gradual change in

behavior, or both. Some have **acute** symptoms e.g. headache, nausea, vomiting, seizures, or localized neurologic signs with ↑ ICP. **Inv.** CT or MRI of head. **Rx. Antibiotics & surgery.**

☒ **Delayed growth & development** may occur in severe untreated TOF with chronic hypoxia, especially when O₂ saturation is chronically <70%.

🔗 **Anomalies that may associated with TOF:-**

☐ Right sided aortic arch (20%), PDA, multiple VSDs, AV canal defect (especially in patient with Down synd) & aberrant major coronary artery crosses over the right ventricular outflow tract (5-10%). ☐ Congenital absence of the pulmonary valve → cardiomegaly with loud to-and-fro murmur, whereas cyanosis may be absent. It also may cause marked aneurysmal dilatation of the main and branch pulmonary arteries results in compression of the bronchi and produces secondary tracheomalacia or bronchomalacia → stridor or wheezing respirations

with recurrent pneumonia. ☐ Absence of a branch pulmonary artery (mainly the left) which often associated with hypoplasia of the affected lung. ☐ TOF is one of the conotruncal family of heart lesions that may be

associated with **CATCH 22** (cardiac defects, abnormal facies, thymic

hypoplasia, cleft palate, hypocalcemia) or **DiGeorge syndrome**.

Rx of TOF. It depends on the **severity** of right ventricular outflow obstruction; but however, **all patients** with TOF require surgical

correction (sooner or later). ☐ **Severe obstruction** with neonatal cyanosis require **urgent infusion of**

PGE1, 0.01-0.20 µg/kg/min to keep PDA open, then plan for surgery. ☐ **Less severe obstruction** in an infants who are stable and awaiting surgical intervention require careful observation with the following:- 1. **Pv & Rx of dehydration & polycythemia**. 2. **Antibiotic Px** for bacterial endocarditis. 3. **Iron Rx** for IDA (which also may ↓ the frequency of Tet spells). 4. **Propranolol**, 0.5-1 mg/kg every 6 hr orally may also ↓ the frequency &

severity of Tet spells. 5. **Surgery** (palliative or corrective) is required **once Tet spells begin**.

☐ **Palliative surgery** involve the **modified Blalock-Taussig shunt** by

either Gore-Tex conduit that anastomose subclavian artery to the

homolateral branch of the pulmonary artery, or by a Central shunt that directly anastomose ascending aorta to the main pulmonary artery. **Note:** *Development of a continuous murmur over the lung fields indicates a*

functioning anastomosis. However, Blalock-Taussig shunts are currently reserved for patients with comorbidities e.g. other major congenital anomalies or prematurity

Postoperative Cxs include: chylothorax, diaphragmatic paralysis, Horner synd, HF (if the shunt is large) & long-term arm length discrepancy with ↓ radial pulse. Rapidly progressive cyanosis may indicate thrombosis of the shunt which requires emergency surgery. As

the child **grows**, the palliative shunt become **inadequate**, so patient may need corrective surgery. ☒ **Corrective surgery**; here the surgeon should make a full repair (with removal of all artificial shunts). **Postoperative Cxs** include: R.V. failure, transient right heart block, residual VSD with Lt to Rt shunting, myocardial infarction (due to

interruption of the aberrant coronary artery) & long-term pulmonary valvular insufficiency.