Tetralogy of Fallot

What is it?

- Four common features; VSD, overriding aorta, enlarged right ventricle & pulmonary stenosis.
- Most common form of cyanotic heart defect after age one.
- 15% of ToF patients have deletion of chromosome 22q11.
- Associated lesions are ASD, additional muscular VSD, right aortic arch, anomalous LAD and complete AVSD.
- Surgical treatment as baby/children includes VSD closure and pulmonary valvotomy.

How is it diagnosed?

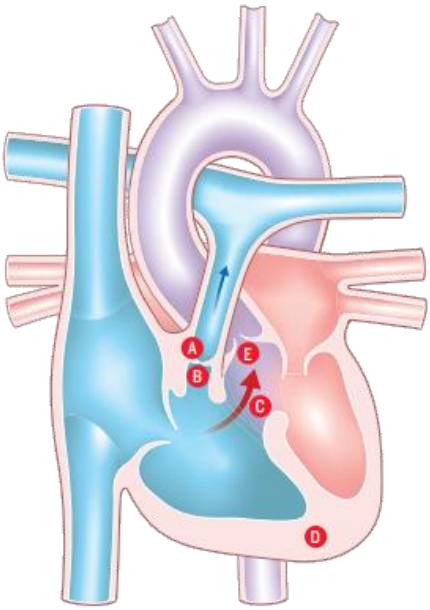
- Diagnosed as baby by heart murmur and cyanosis.
- Echocardiogram. CMR, CT, CPET and cardiac cath can be done for diagnostic work up of repaired patients.

How does it affect the heart?

- High risk arrhythmia & sudden cardiac death.
- LV dysfunction due to shunts, cyanosis and LV volume overload.
- Residual VSDs can occur.
- RV dilation & dysfunction due to residual PR
- Aortic root dilation with AR seen in 15% adults with ToF.
- Residual right ventricular outflow tract obstruction (RVOTO).

What is the treatment/long term management?

- Pulmonary valvuloplasty /surgical valvotomy as a child
- Pulmonary valve replacement surgically or percutaneously.
- Enlargement of pulmonary artery may be necessary/stenting to branch PAs
- EP testing +/- ablation for symptomatic arrhythmia.
- ICD for secondary prevention of SCD.
-
- What is the follow up?
- Regular 1-2 year follow-up in a ACHD centre.
- Echocardiogram at each follow-up.
- Monitor aortic root size.
- All patients should have a CMR if RV enlarging



A Narrowed pulmonary valve
B Narrowing beneath pulmonary valve
C Ventricular septal defect
D Thickened muscle
E Overriding aorta

J.S/ACHD 2018

Reference: The European Society of Cardiology (2012) *Guidelines for the Management of Grown-Up Congenital Heart Disease.* Oxford University Press: Oxford. Image *with permission* www.bhf.org