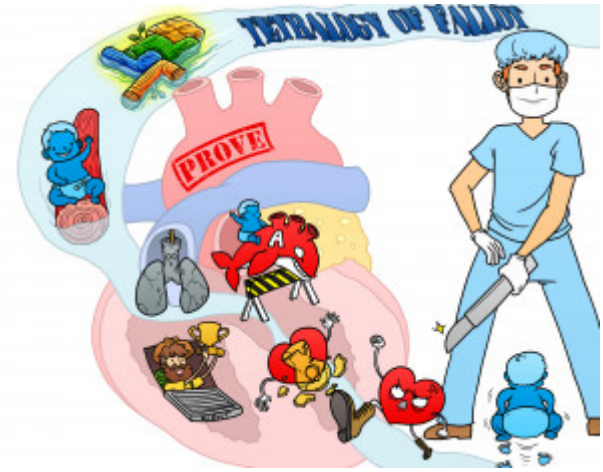
Tetralogy of Fallot (TOF)

Tetralogy of Fallot (TOF) is a congenital birth defect of the heart that leads to cyanosis. It presents with the combination of four different heart defects and is the most common cause of blue baby syndrome. The four pathologies seen in this condition are pulmonary infundibular stenosis, right ventricular hypertrophy (RVH), overriding aorta, and ventricular septal defect (VSD).



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Features

Blue Baby Syndrome
Blue Baby
TOF is the most common cause of blue baby syndrome in newborns. Babies are "blue" due to the cyanosis experienced in this heart disorder.

Mechanism

PROVe Acronym
Prove-proof Stamp
The acronym “PROVe” can be used to help remember the four specific abnormalities generally associated with TOF: P for pulmonary infundibular stenosis, R for RVH, O for overriding aorta, V for VSD, and the “e” is silent without an associated pathology.

Pulmonary Infundibular Stenosis
Lungs made of Stone
One of the tetralogies that can occur is pulmonary infundibular stenosis. Pulmonary infundibular stenosis is described as a narrowing of the right ventricular outflow tract. It occurs at the pulmonary valve, or just below at the infundibulum.

Right Ventricular Hypertrophy
Right Vent Hiker-trophy
Due to the increased pressure and resistance of the right ventricular outflow tract, the right heart becomes pathologically hypertrophied. This RVH can cause the heart to be visualized as "boot-shaped."

Overriding Aorta
Over-riding A-orca
In TOF, the aorta sits above the VSD, giving it biventricular access. This is problematic because deoxygenated blood from the right ventricle can then make its way into systemic blood flow.

Ventricular Septal Defect (VSD)
Vase-hole-heart
Patients with TOF have a VSD which allows deoxygenated blood from the right ventricle to mix with oxygenated blood from the left ventricle. Because pulmonary stenosis causes right-sided outflow obstruction, blood is preferentially pushed toward the left side, and a right-to-left shunt is seen.

Diagnosis and Treatment

Boot-Shaped Heart
Boots on Heart
Patients show a "boot-shaped" heart on X-ray due to RVH.

Squatting for Tet Spells

Squatting

Often patients, especially children, squat to alleviate cyanotic tet spells. This is a therapeutic action because squatting increases systemic resistance. The increase in systemic resistance allows more blood to be pumped into the pulmonary system and for improved oxygenation.

Surgery

Surgeon

The only definitive treatment for TOF is surgical correction of the respective heart malformations.