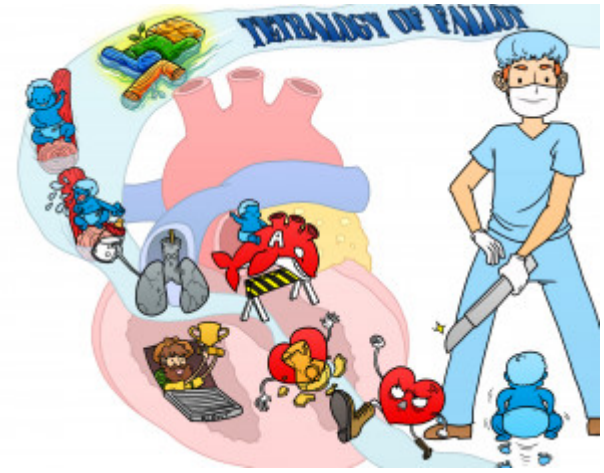
Tetralogy of Fallot

Tetralogy of Fallot (ToF) is a congenital birth defect that is comprised of a combination of four heart defects: pulmonary stenosis, right ventricular hypertrophy, overriding aorta, and a ventricular septal defect (VSD). Symptoms do not usually appear until after the ductus arteriosus begins to close within the first week of life. Tetralogy of Fallot is a common cause of cyanosis in neonates and requires surgical intervention within the first year of life.



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Blue Baby Syndrome (Cyanosis)
Blue Baby
Tetralogy of Fallot is the most common cause of blue baby syndrome in newborns. Babies are "blue" due to the cyanosis experienced in this heart disorder. This cyanosis directly correlates to the level of pulmonary stenosis, which prevents blood flow to enter the lungs from the right ventricle and forces deoxygenated blood through a VSD and into the systemic circulation.

Crying or Feeding
Baby Crying and Feeding
Neonates often become cyanotic during times of feeding or crying when oxygen demand is greater than supply. These babies are sometimes termed “blue babies.”

PROVe

Pulmonary Stenosis
Lungs made of Stone
One of the tetralogies which can occur is pulmonary infundibular stenosis. Stenosis is another term for narrowing. In ToF the outflow tract of the right ventricle is narrowed. The degree of cyanosis is directly correlated to the severity of narrowing.

Right Ventricular Hypertrophy
Right Vent Hiker-trophy
Increased pressure caused by the narrowed outflow tract (pulmonary stenosis) causes a compensatory increase inside of the right ventricle. This often causes a boot shaped heart in the neonate on diagnostic exams.

Overriding Aorta
Over-riding A-orca
In Tetralogy of Fallot, the aorta is shifted slightly to the right anteriorly to be on top of the VSD (ventricular septal defect). This causes the aorta to receive blood flow from both the right and left ventricle allowing deoxygenated blood to circulate throughout the body.

Ventricular Septal Defect (VSD)
Vase-hole-heart
A defect/hole in the wall of the ventricles allows blood to shift between the ventricles. As pressures change between the left and right ventricle, deoxygenated blood can bypass the lungs and shunt into the left ventricle allowing deoxygenated blood to enter the systemic circulation.

Diagnosis and Treatment

Boot-Shaped Heart
Boots on Heart
Due to right ventricular hypertrophy (RVH), patients show a "boot-shaped" heart on X-ray.

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Squatting for Tet Spells

Squatting

Cyanotic episodes are termed “tet spells” and classically children will squat to relieve the cyanosis. Squatting increases the systemic vascular resistance, which decreases the amount of blood that crosses improperly through the VSD. This allows more blood flow to properly flow through the lungs.

Surgery

Surgeon

The only definitive treatment for Tetralogy of Fallot is surgical treatment to correct the heart malformations.

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