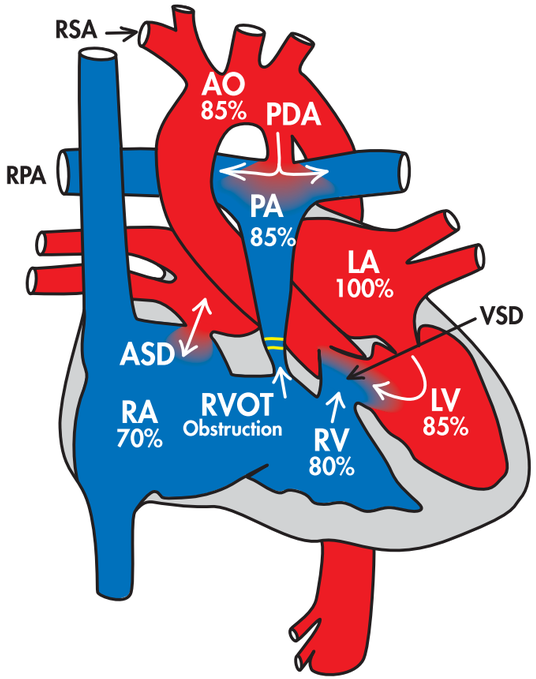
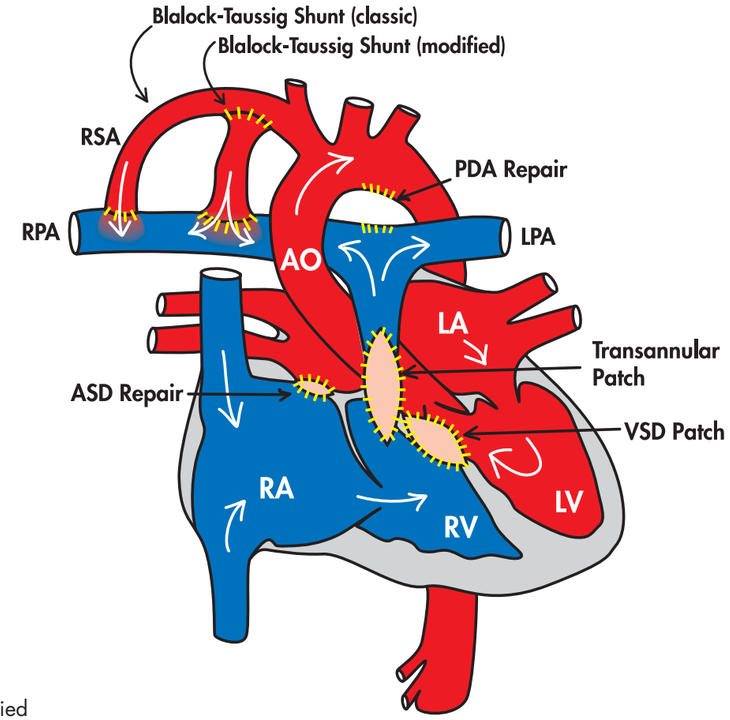
Tetralogy of Fallot (TET)



- **Large malalignment VSD**
- **Right ventricular outflow tract obstruction (Pulmonary Stenosis)**
- **Overriding aorta**
- **Right ventricular hypertrophy**

SURGICAL REPAIR



Tetralogy of Fallot (TET)

In General

Tetralogy of Fallot (TET, TOF) is a complex cyanotic abnormality resulting from failure of the interventricular septum to properly attach to the annulus during fetal (conus) development. As a result, the outlet portion of the right ventricle (conus arteriosus, or infundibulum) is malaligned, leading to the combination of 4 congenital abnormalities that define TET:

- Ventricular septal defect (VSD) associated with malalignment of the infundibulum
- Right ventricular outflow tract (RVOT) obstruction or ***Pulmonary Stenosis*** (refer to page 48)
- Overriding aorta (AO)
- Right ventricular hypertrophy

The degree of RVOT obstruction determines the severity of the abnormality. In cases where pulmonary stenosis is minimal, shunting across the VSD may be bi-directional or left to right resulting in acyanotic TET. At the other end of the spectrum, TET with pulmonary atresia and complete closure of the RVOT is the most severe form of the disease. Acyanotic TET can progress to cyanotic TET. Associated cardiac abnormalities may complicate treatment.

- Typically, the VSD is large and unrestrictive. Right-to-left shunting predominates.
- An ASD (secundum type) is sometimes present.
- Pulmonary valve (PV) stenosis is common. The pulmonary arteries may be hypoplastic.
- Rarely, the PV is absent (pulmonary atresia).
- The outflow obstruction may be located at the subvalvular level.
- The AO is dilated and displaced to the right, overriding the VSD. Some degree of aortic regurgitation may be present.

Echocardiography:

- Evaluate the direction of flow and gradient of the VSD.
- Demonstrate and grade the degree of aortic malalignment.
- Measure the gradient across the pulmonic valve (PV) and assess the degree of RVOT obstruction.
- Measure the diameter of the pulmonary artery (PA) and its branches.
- Evaluate coronary artery anatomy (particularly the left anterior descending coronary artery).

Associated Cardiac Abnormalities May Include:

- ***Right Aortic Arch*** (refer to page 16)
- ***Anomalous Coronary Arteries*** (refer to page 14)
- ***Partial Anomalous Pulmonary Venous Return*** (refer to page 40)
- ***Persistent Left Superior Vena Cava*** (refer to page 42)
- ***Pulmonary Atresia with VSD*** (refer to page 46)

Surgical Repair

TET is the most common cyanotic heart defect among infants, and surgical intervention is required. In the past, 2 to 4 years before corrective surgery was peformed, a palliative shunt procedure was done in order to establish a systemic venous-to-pulmonary artery connection. Three types of shunts were commonly used: Classic Blalock-Taussig shunt, Waterston shunt, and Potts shunt. These procedures are no longer widely used (see ***Surgical Procedures,*** page 76*)*.

Presently, complete single-stage surgical repair at approximately 6 months of age is the preferred approach. However, severely cyanotic neonates may still require palliative shunting. In these patients, a modified Blalock-Taussig shunt is used employing a tubed homograft or synthetic conduit.

Complete repair generally involves the following:

- If a modified **Blalock-Taussig shunt** was used, it is detached and removed.
- The **VSD** is closed with a synthetic patch or autograft (pericardium). If an **ASD** is present, it will be closed. The PDA will be closed if it has remained open.
- Pulmonary stenosis is relieved by resection of obstructive muscle tissue in the right ventricular outflow tract (**RVOT reconstruction**), pulmonary valvotomy, and reconstruction using a transannualar patch.