Repair of Tetralogy of Fallot (TOF)

Surgery to repair tetralogy of Fallot involves closing the ventricular septal defect (VSD) and removing the obstruction of blood flow to the lungs. In TOF, there can be narrowing beneath the pulmonary valve (sub-valvar stenosis) due to thickened right ventricle muscle, the pulmonary valve itself can be small (valvar stenosis), or the area above the pulmonary valve can be small (supra-valvar stenosis). Sub-valvar stenosis can be relieved by resection of enlarged ventricular muscle or with a patch. When combined with valvar stenosis, a patch can be placed from the main pulmonary artery to the right ventricle across the pulmonary valve, which is known as a “transannular patch.” If the narrowing is limited to the supra-valvar area, a patch can be placed across the narrowing on the main pulmonary artery. Repair of TOF is usually done between 3-9 months of age.

In TOF/absent pulmonary valve, as the name implies, the patient is missing a pulmonary valve. Due to the unrestricted blood flow to the lungs, the pulmonary arteries are often very large. If they are large enough to compress the patient’s airway, the patient will have surgery in the neonatal period. During surgery to correct this type of congenital heart disease, a right ventricle to pulmonary artery (RV-PA) conduit is used to replace the pulmonary valve. There are many types of materials used for RV-PA conduits. Depending on the surgical plan and patient’s anatomy, conduits made of Gore-tex®, homograft (cadaver valved tissue), Contegra® conduits (Medtronic) (valved bovine (cow) jugular vein), or Hancock® conduits (Medtronic)(Dacron tube graft containing a porcine (pig) valve) can be used.

During surgery, a median sternotomy (incision through the middle of the chest) is done. The patient is placed on cardiopulmonary bypass (heart–lung machine). The right atrium is opened and thickened right ventricle muscle is removed through the tricuspid valve. A Dacron patch is cut to the appropriate size and sutured over the VSD. The tricuspid valve is tested to insure competency, and stitches are placed to repair any leaks. If needed, the pulmonary artery is opened and a patch of bovine (cow) pericardium (sac surrounding the heart) is cut to the appropriate size. Oftentimes, thickened right ventricle muscle can be removed through the pulmonary valve if necessary. If indicated, the bovine pericardium is sewn either as a sub-valvar, transannular, or supra-valvar patch.

If the patient has TOF/absent pulmonary valve, incisions are made on the pulmonary artery and right ventricle. An appropriate sized RV-PA conduit is selected. One end of the conduit is sewn onto the incision on the pulmonary artery and the other end is sewn onto the incision on the right ventricle. If the right and left pulmonary arteries are large enough to compress the patient’s airway, they can be plicated to narrow their effective size to prevent further airway compression.

Typical Post-Operative Course:

- **Surgery Length**: 4 hours
- **Typical Lines**: Most patients will return to the Cardiovascular Care Center after surgery with a breathing tube, an arterial line to monitor blood pressure, a central venous line (for giving IV medicines and drawing labs), a peripheral IV, chest tubes to drain fluid, temporary pacing wires, and a foley catheter to drain urine.
- **Typical Post-Operative Recovery**: The breathing tube is generally removed within 24-48 hours after surgery. The arterial line is usually removed within a few days, once most IV medicines are stopped. The central venous line is removed once most IV medicines are stopped and labs no longer need to be drawn. Chest tubes are usually removed 24-48 hours following surgery, once the output of fluid is minimal. If used, depending on the type of conduit placed and surgical plan, the patient may be placed on Aspirin for a period of time after surgery.
- **Typical Length of Stay**: A patient usually stays in the hospital for 6 days following repair of tetralogy of Fallot.

**Typical Home Medications**: Children will require one or more medications at home following repair of TOF such as:
- Diuretics (Lasix) to control fluid
- Anticoagulant (Aspirin) to prevent clotting (if a conduit was placed)