13 Tetralogy of Fallot

Antonio G. Cabrera, Patricia Bastero, Stuart R. Hall, Carlos M. Mery

Tetralogy of Fallot (TOF) is the most common cyanotic congenital anomaly. It is characterized by anterosuperior deviation of the infundibular septum, leading to 4 components: a large anterior malalignment conoventricular VSD, RVOT obstruction (valvar and subvalvar), aortic override, and RVH (Figure 13-1).

Coronary anomalies are present in up to 10-15% of patients with TOF, most commonly an LAD that arises from the right coronary and crosses the RVOT inferiorly. Approximately 25% of the patients have a right-sided aortic arch. TOF is associated with genetic syndromes in 20% of the patients, most notably DiGeorge syndrome (22q11 deletion), trisomy 21, and VACTERL association (vertebral anomalies, imperforate anus, cardiac anomalies, tracheoesophageal fistula, renal anomalies, and limb abnormalities).

Pathophysiology and Clinical Presentation

Progressive subpulmonary obstruction causes shunting of deoxygenated blood across the VSD into the LV (right-to-left shunting) leading to a decrease in systemic oxygen saturation. The degree of shunting is related to the degree of RVOT obstruction and the relative SVR. As such, children may present with cyanosis at birth (rare) or may develop progressive cyanosis as the subpulmonary stenosis progresses.

On physical exam, in addition to varying degrees of cyanosis, patients present with a high-pitched systolic ejection murmur at the left sternal border. Clubbing can be seen in advanced cases, usually after 2-3 years of age.

Children with TOF may present with a hypercyanotic spell ("tet spell"), an acute and sustained episode of profound cyanosis, hyperpnea, agitation, and acidosis. It is the result of complete or near complete obstruction of pulmonary blood flow with worsening right-to-left shunting. This profound cyanosis can lead to acidosis due to poor oxygen delivery. Acidosis may then reduce SVR, thus worsening right-to-left shunting and causing further acidosis. Clinically, patients present with muffling or shortening of the systolic ejection murmur. Treatment of hypercyanotic spells involves:

- Have a family member hold the patient in a quiet room to calm the patient
- Oxygen administration
- Use alpha-agonists (phenylephrine) to increase SVR
- Treat acidosis with sodium bicarbonate replacement
- Decrease agitation/sedation with morphine and/or ketamine

Diagnosis

- **CXR (Figure 13-2).** RVH will produce a "boot-shape" appearance with oligemic lung fields from decreased pulmonary blood flow.
- **ECG.** RA enlargement, RVH, or dominant right-sided forces.
- **Echocardiogram (Figure 13-3).** Important features to assess include details on the VSD (perimembranous vs. doubly-committed juxta-arterial defect), degree of

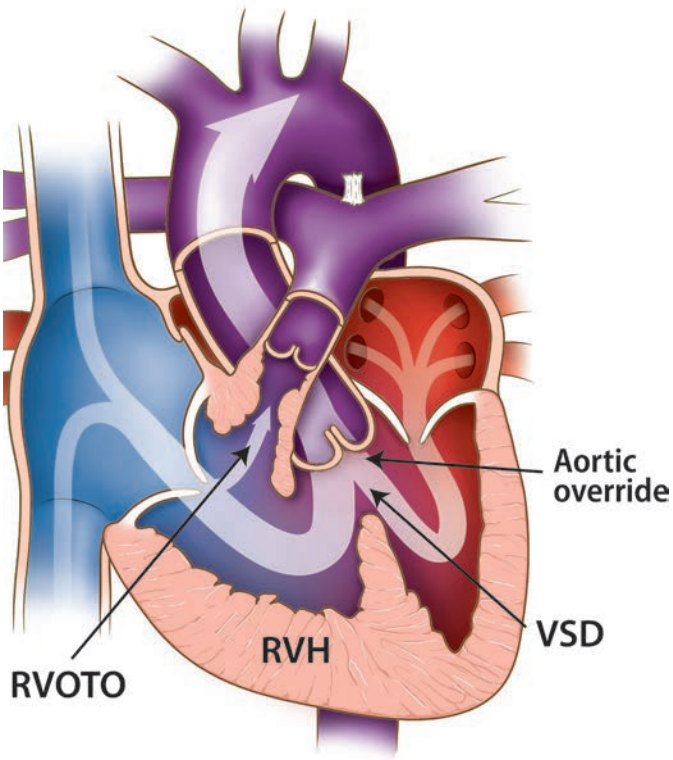

Figure 13-1. Elements of TOF. Anteroseptal deviation of the infundibular septum leading to: 1) a large anterior malalignment conoventricular VSD, 2) valvar and subvalvar RVOT obstruction (RVOTO), 3) aortic override, and 4) right ventricular hypertrophy (RVH).

valvar/subvalvar pulmonary stenosis, aortic arch anatomy, coronary anatomy, and associated anomalies.

- **Cardiac catheterization.** Not necessary for diagnosis. Can be helpful in small children in which stenting of the arterial duct can provide a stable source of pulmonary blood flow as an alternative to a modified Blalock-Taussig-Thomas shunt (mBTTS).

Indications / Timing for Intervention

TOF is repaired using an infundibular-sparing transatrial/transpulmonary repair. This repair is best performed after 4-6 months of age due to the technical difficulty in smaller children and the possibility of JET from surgical retraction in neonates.

Indications for early intervention (mBTTS vs. full repair) include progressive cyanosis (oxygen saturation <85%) or the occurrence of a cyanotic spell (Figure 13-4). Otherwise, patients are repaired electively after 4-6 months of age.

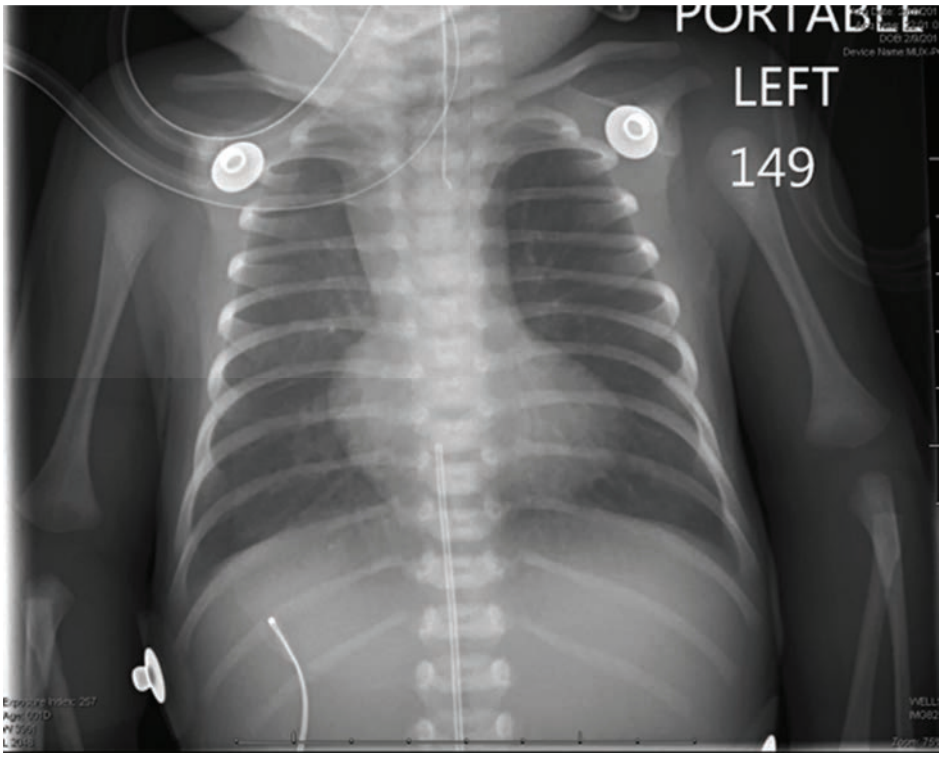

Figure 13-2. Classic CXR on TOF showing a "boot-shaped" heart with oligemic lung fields.

Anesthetic Considerations

The main goals of the pre-CPB period are to maintain adequate preload and SVR to prevent hypercyanotic spells. Should a hypercyanotic spell occur: increase the FiO_2, give phenylephrine 5-10 mcg/kg, increase preload, and deepen the anesthetic. Abdominal compression or surgical compression of the aorta can also augment SVR and force more blood into the pulmonary circulation. Esmolol, starting at 50-200 mcg/kg/min, can also help relieve infundibular spasm and is easily titrated. If all measures fail, rapid cannulation and initiation of CPB may be needed.

Post-CPB, inotropic agents are rarely necessary. Given the possible presence of some dynamic narrowing after repair (see below), esmolol is routinely used to slow down the heart rate and allow adequate time for RV filling. Milrinone can be considered for its lusitropic effects but its concomitant decrease in SVR may make the child more hypotensive than desired.

Surgical Repair

The main goal of the TOF infundibular-sparing repair is to relieve RVOT obstruction while preserving as much contractile infundibular muscle as possible. A ventriculotomy

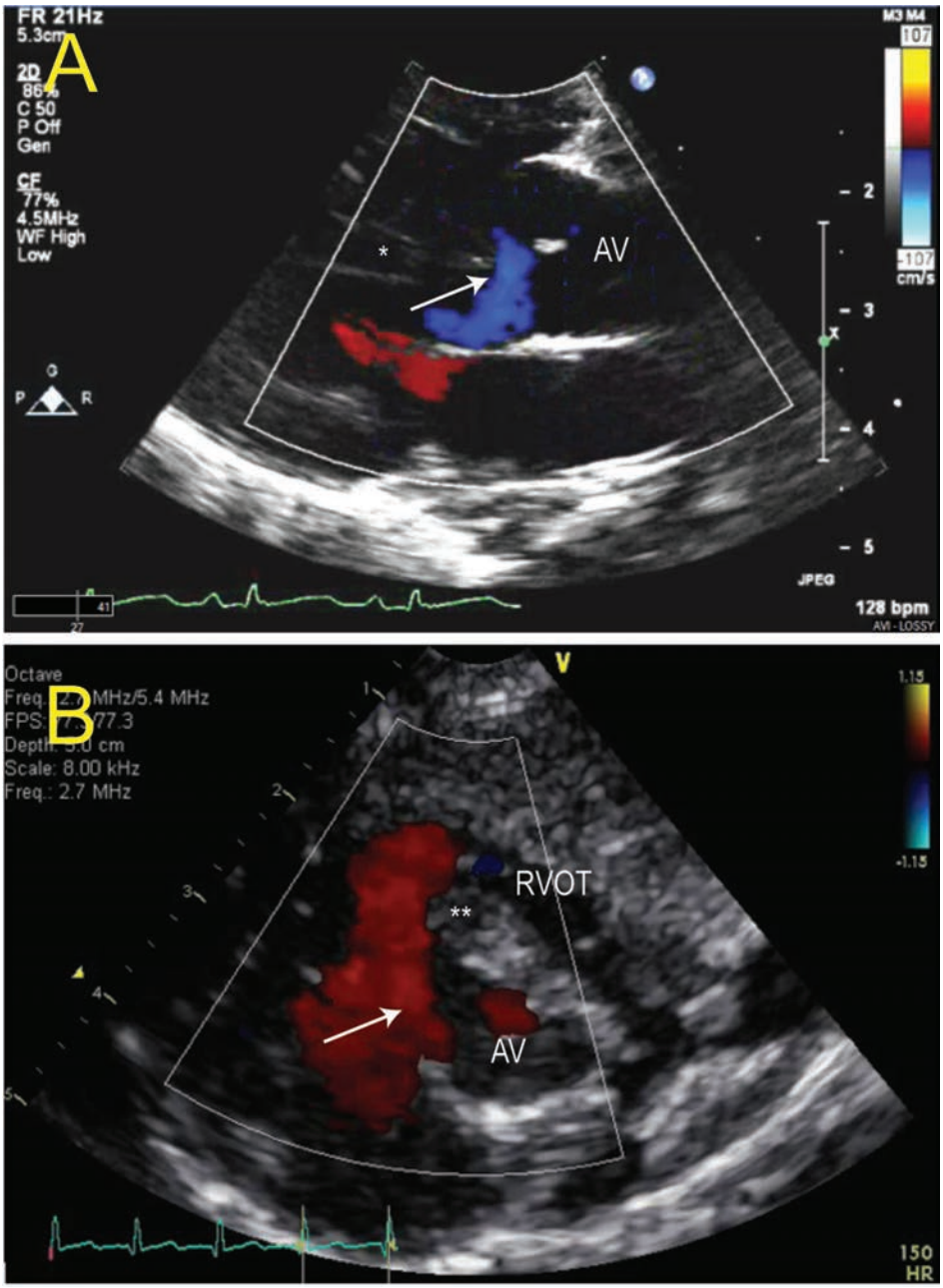

Figure 13-3. Echocardiography in TOF. The parasternal long-axis view (A) shows a conoventricular septal defect (arrow) with override of the aortic valve (AV) on the interventricular septum (*). The short-axis parasternal view (B) shows an anteriorly deviated infundibular septum (**) causing significant narrowing of the RVOT. Flow is seen from the VSD (arrow) toward the RVOT.

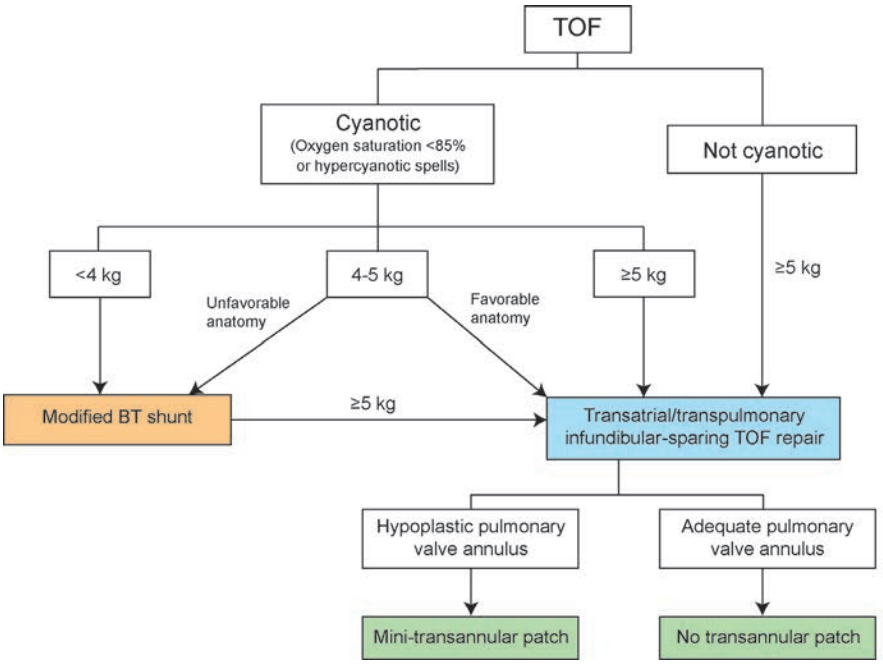

Figure 13-4. Decision process for surgical intervention on TOF.

is avoided to prevent substituting functional muscle for a noncontractile patch. The preservation of the infundibulum may lead in the long term to improved function and better tolerance against PI.

The procedure is performed using standard aorto-bicaval cannulation and moderate hypothermia (28 °C). Working through the tricuspid valve, the hypertrophied septoparietal trabeculations are excised from the free wall of the infundibulum. The infundibular septum is left alone to serve as the anchor for the VSD baffle. The anterior and septal leaflets of the tricuspid valve may be taken down for visualization. Significant traction is avoided to prevent postoperative JET.

The VSD is repaired by creating a baffle between the LV and the anteriorly displaced aortic valve using an autologous pericardial patch tanned on glutaraldehyde. The conduction system (bundle of His) travels on the posteroinferior edge of the VSD and can be injured during repair.

A longitudinal incision is performed on the main pulmonary artery and the pulmonary valve is inspected. If after performing a pulmonary valvotomy the valve accepts a Hegar dilator corresponding to the normal size of the pulmonary valve based on BSA, the valve is preserved (25%). Otherwise, the longitudinal incision is extended proximally across the pulmonary annulus for a few millimeters, creating a transannular incision (75%). The incision is extended minimally until the corresponding Hegar dilator is allowed to pass, therefore leaving the infundibulum intact. An autologous pericardial patch is used to reconstruct the main PA.

TCH experience on TOF (2007-2014)
TOF repairs performed at TCH: 22-35 per year
Median age at surgery: 9 months
Patients that require mBTTS prior to full repair: 17%
Median ICU length of stay: 3 days
Median hospital length of stay: 7 days
Perioperative mortality: 0.9%

An LA line is placed in all patients to help with postoperative management (Chapter 66). Since these patients have diastolic RV dysfunction due to RV hypertrophy, CVP is not an accurate reflection of volume status. Temporary atrial pacing wires are placed in all patients (for postoperative management of JET or other arrhythmias). Ventricular wires are placed selectively based on AV conduction.

Mild dynamic RVOT obstruction at the subvalvar area (<2.5 m/s) is tolerated and expected to improve within a few weeks as the RV remodels. If the obstruction is more significant on postoperative TEE, RV pressure may be directly measured. If the RV pressure is greater than 2/3 systemic, the patch is further extended proximally.

Postoperative Management

The postoperative management is mainly directed to address diastolic dysfunction, for which volume resuscitation and heart rate control are key.

General Management

- **Fluids.** 25% maintenance with D5%/0.45% NS is standard. However, these patients will require volume resuscitation due to diastolic dysfunction.
- **Analgesia and sedation.** Analgesics and sedatives are adjusted for patient's comfort. A fentanyl infusion is used for intubated patients and morphine PCA and/or intermittent morphine is used for short-term mechanical ventilation needs or extubated patients. Scheduled acetaminophen q6h (enteral, rectal, or IV) is used as an adjuvant. *Sedation* is achieved with a combination of dexmedetomidine (both intubated and extubated patients) and/or intermittent benzodiazepines. Midazolam as a drip (only for intubated patients) or intermittently is commonly used; lorazepam is used for longer-term benzodiazepine needs.
- **Vasoactive drugs.** Most patients will arrive from the OR on an esmolol infusion 25-400 mcg/kg/min and a low-dose milrinone infusion 0.25 mcg/kg/min.
- **Mechanical ventilation.** Usually ventilated on SIMV-VC with pressure support and Vt 8-10 ml/kg, aiming for a pH of 7.35-7.45 and SaO_2 >93%. Postoperative TOF patients are expected to be extubated within 12-24 hours after surgery (usually within 6 hours), unless a significant respiratory comorbidity or postoperative complication prevents extubation.

What to Expect in the First 24 Hours Postoperatively

- **Vasoactive drugs.** Patient on no vasoactive drugs or a low-dose of milrinone and/or low-dose esmolol, ready to be started on propranolol if esmolol infusion cannot be stopped. Patients who were on propranolol preoperatively are usually placed on

the same dose of propranolol in the CICU once the esmolol drip has been stopped and slowly weaned off in the next few weeks.
- **Ventilation.** Extubated on regular nasal cannula with low O_2 requirements.
- **Fluids.** Positive fluid balance ~300-350 ml.
- **Nutrition.** Clear fluids are started PO 4 hours after extubation if the patient is stable. Progress to regular diet ad lib.

Complications

The most common postoperative complication after TOF repair is LCOS, usually related to diastolic dysfunction.

- **LCOS.** It will present with tachycardia, normal or low BP, high CVP with normal LAP (low LAP if low intravascular volume), lactic and/or metabolic acidosis, oliguria, low cerebral NIRS, and increased core-toe temperature gradient. Management consists in optimizing RV preload (volume administration, usually no more than 40 mL/kg), improving ventricular filling time (rate control with esmolol), and optimizing lusitropy (milrinone) to optimize RV output, as well as optimizing RV afterload (milrinone and early extubation).
- **Arrhythmias.** Arrhythmias compromising cardiac output are not common among our patients; they include JET (2%) and complete heart block (0.3%). For the management of these arrhythmias, see Chapters 74 and 75. Right-bundle-branch block is a common finding after TOF repair and has no hemodynamic impact.

Suggested Readings

McKenzie ED, Maskatia SA, Mery CM. Surgical management of tetralogy of Fallot: in defense of the infundibulum. Semin Thorac Surg 2013;25:206-212.

Morales DL, Zafar F, Heinle JS, et al. Right ventricular infundibulum sparing (RVIS) tetralogy of Fallot repair: a review of over 300 patients. Ann Surg 2009;250:611-617.

Niu MC, Morris SA, Morales DL, et al. Low incidence of arrhythmias in the right ventricular infundibulum sparing approach to tetralogy of Fallot repair. Pediatr Cardiol 2014;35:261-269.