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Tetralogy of Fallot

9.1 Morphology

The important features of this lesion are:

1. Right ventricular (RV) outflow tract obstruction (RVOTO) which is infundibular and/or valvular stenosis.
2. Unrestricted and malaligned VSD (ventricular septal defect) associated with anterior misalignment of the conal septum.
3. Dextroposition of aorta which overrides VSD, i.e., an aorta that to varying degrees originates from the RV.
4. The anterior displacement and rotation of the infundibular septum also causes RV obstruction of variable degree and location.
5. The RVOTO leads to secondary right ventricular hypertrophy (see Figure 9.1).

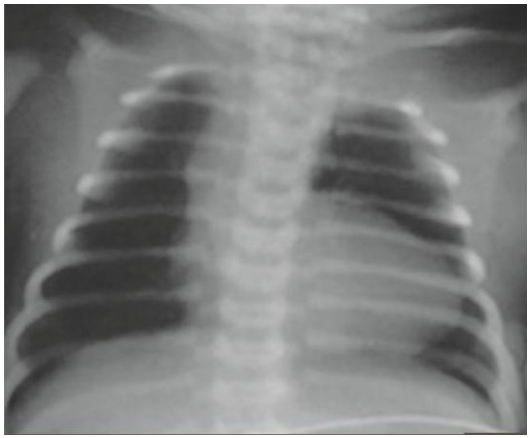

Figure 9.1 *Chest X-ray showing boot shaped heart (Coeur en sabot) characteristic of tetralogy of Fallot in a 3 month old infant. Note the decreased pulmonary vascularity and absent PA (pulmonary artery) shadow in the superior mediastinum with leftward shift of ventricular apex due to RV hypertrophy.*

The pulmonary arteries vary in size and distribution, and they may be atretic or hypoplastic. In some, a varying degree of peripheral pulmonary arteries stenosis occurs. The coronary anatomy may also be abnormal, i.e., the left anterior descending (LAD) coronary artery arises from the proximal right coronary artery which crosses the RV outflow at variable distances from the pulmonary valve annulus. This abnormality makes placement of a patch across the pulmonary annulus risky, possibly, requiring an external conduit. During the VSD repair, the anomalous LAD coronary artery is also prone to injury.

9.2 Pathophysiology

The VSD is usually large (nonrestrictive), therefore, RV and left ventricular (LV) pressures are equalized. The relative resistance of the RVOT and systemic vascular bed determines the direction of a shunt. If the RVOT obstruction is severe, the intracardiac shunt is from right to left, and pulmonary blood flow may be markedly diminished resulting in cyanosis and hypoxemia.

Some infants at birth with tetralogy may not show signs of cyanosis, but they may later develop episodes of bluish pale skin during crying or feeding, called “tet spells”. These episodes occur even in non-cyanotic patients, and the mechanism is thought due to spasm of the infundibular septum, which acutely worsens the right ventricular (RV) outflow tract obstruction (RVOTO). The older child by squatting increases systemic vascular resistance, thus decreases the magnitude of the right-to-left shunt with increase in pulmonary blood flow.

In tetralogy, the predominant shunt can also be from the left to right if the infundibular stenosis is minimal. These infants are not cyanotic (pink tetralogy), but they often have systemic oxygen desaturation. Signs and symptoms of tetralogy progress due to increased RVOTO and hypertrophy of the infundibular septum that leads to secondary RV hypertrophy, increased right-to-left shunting,

and systemic hypoxemia. Dehydration, acidosis, stress, infection, beta-adrenergic agonists, and closure of the ductus arteriosus can all worsen the cyanosis.

9.3 Management of Hypercyanotic Tet Spells

"Tet spells": Paroxysms of hyperpnoea, prolonged crying, intense cyanosis, and ↓ intensity of the murmur of pulmonic stenosis.

9.3.1 Mechanism

It is secondary to infundibular spasm which acutely worsens RVOTO, and/or ↓ systemic vascular resistance (SVR) with ↑right-to-left shunting at VSD. This results in diminished pulmonary blood flow. If left untreated, it may result in syncope, seizure, stroke, or a death.

9.3.2 Treatment

IV beta-blockers such as propranolol may be needed in a severe infundibular spasm. *Acute episodes usually require rapid intervention as mentioned below:*

1. 100% Oxygen: It is a potent pulmonary vasodilator and a systemic vasoconstrictor. This increases pulmonary blood flow to the lungs.
2. Morphine sulfate: Give 0.1-0.2 mg/kg IM or subcutaneously (SC). May reduce the ventilatory drive and ↓ systemic venous return.
3. Phenylephrine: Give 0.02 mg/kg IV, the dose may be repeated. It is used to increase SVR.
4. Dexmedetomidine: Give a low dose of 0.1-0.125 mcg/kg/hour infusion *without a bolus.* In some hypercyanotic neonates, this infusion has ameliorated the symptoms.
5. Metabolic acidosis: Corrected by $NaHCO_3$. It reduces the respiratory drive of acidosis.

6. If feasible, hold the infant in a knee chest position.

9.4 Surgical Treatment

Most infants with adequate oxygen saturation should undergo an elective repair, but progressive hypoxemia and the occurrence of hypercyanotic spells are the indications for an early surgery.

The infusion of prostaglandins to maintain the patency of ductus in critically ill cyanotic-hypoxic infants has decreased the need to perform urgent surgery such as the systemic-to-pulmonary artery shunt.

The current trend is to perform a total surgical correction at presentation though studies have shown that the surgery is preferably done at or about 12 months of age.

Primary repair avoids prolonged right ventricular (RV) outflow obstruction and the subsequent right ventricular hypertrophy (RVH), prolonged cyanosis, distortion of pulmonary artery (i.e., PA) secondary to the palliative shunt, and postnatal angiogenesis.

Multiple VSDs, severe annular hypoplasia, pulmonary artery atresia, small pulmonary arteries, and a low birth weight are some of the factors, which increase the risk for early repair.

The infants with tetralogy of Fallot and pulmonary atresia or an anomalous left anterior descending coronary artery that crosses the RV outflow tract may not be surgical candidates for establishing transannular patch but may require placement of a conduit.

Infants with extremely small pulmonary arteries may not tolerate total correction with a conduit placement in infancy. These infants may require palliation with the Blalock-Taussig shunt instead of primary corrective surgery.

9.5 Operative Procedure

Principle: Perform correction to provide unobstructed pulmonary blood flow, close VSD and ASD, preserve the pulmonary valve if feasible, and adequately resect all the obstructing and hypertrophied infundibular muscle bands.

The procedure is performed on CPB (cardiopulmonary bypass) with moderate hypothermia and cardioplegic arrest. In small infants and neonates deep hypothermia and circulatory arrest is often used.

RV approach: Through a small infundibular incision the hypertrophied and obstructing muscle bands are resected. The VSD is closed with a Dacron patch. The pulmonary valve and/or annulus are evaluated, and if it is small (by Z value), it is enlarged by transannular incision and closed with a patch covering both the annulus and infundibulum. The patent foramen ovale (i.e., PFO) should be left open in neonates to assist systemic output in the early postoperative period.

Right atrial approach: Through the right atrium, the VSD and ASD are closed. Pulmonary arteriotomy incision is made to evaluate the pulmonary valve and/or annulus. If the valve is stenotic or the annulus is small (may be assessed by Z values), either valvotomy and/or or transannular incision is performed. The hypertrophied and obstructing muscle bands of the infundibulum are resected through the atrium or through the infundibular incision. Transannular and infundibular patch is placed as necessary.

Some of the risk factors for an early death are severity of annular hypoplasia, small size of pulmonary arteries, need for transannular patch (debatable), and a high peak RV to LV pressure ratio.

9.6 Postoperative Management

The postoperative course can be variable and depends upon the morphology of the lesion (i.e., RV outflow tract, Z value of pulmonary valve, size of PAs

etc.) and the surgical course (i.e., technique and choice of surgical treatment). The hospital stay is variable but may average 1 to 2 weeks.

9.6.1 Hemodynamic Management

Adequate right ventricular (RV) function is a determinant of postoperative hemodynamic stability.

RV function determines transpulmonary blood flow, left ventricular filling, and LV output.

Significant resection of the right ventricular muscle, coupled with pulmonary insufficiency subsequent to the transannular patch repair can lead to an early postoperative hemodynamic instability.

RV diastolic pressure might be higher than normal secondary to pulmonary insufficiency (as in cases of liberal annular patch re-construction).

CVP may be elevated, the indicating right ventricular failure, secondary to pulmonary insufficiency and / or liberal right ventriculotomy incision.

Some degree of right ventricular failure, however, is common.

RV pressures usually return to normal as the right ventricular performance improves.

9.6.2 To Maintain Adequate Right Ventricular Function Do the Following

1. Maintain atrioventricular synchrony.
2. Infuse adequate volume to maintain RV preload.
3. Maintenance of low pulmonary vascular resistance.
4. Maintain systemic mean arterial pressure either normal or slightly elevated.
5. Avoid excessive reduction of afterload.

Arterial oxygen saturation (SaO_2) following the repair should be normal.

In neonates after corrective surgery (by transannular patch and VSD closure), SaO_2 may fall subsequent to elevations in pulmonary vascular resistance and / or right ventricular dysfunction as a result of right to left shunt through a patent foramen ovale.

9.6.3 Invasive Pressure Monitors

Arterial, central venous catheter, and LA (left atrial) line.

An oximetric catheter may be placed in the PA to assess cardiac output.

9.6.4 Vasoactive Drug Infusions

Dopamine or dobutamine, milrinone, epinephrine, and nitroprusside (see Section I Chapters 4 & 16).

9.6.5 Postoperative Bleeding

It is a potential in patients who have undergone extensive pulmonary artery patch angioplasty.

9.6.6 AV Conduction Abnormalities

They may occur occasionally due to edema around the margins of the VSD, resulting in transient AV (atrioventricular) block. Temporary pacing (atrioventricular pacing) should be available.

9.6.7 Pleural Effusion

It is a common, especially in patients with the presence of aortopulmonary collaterals. Prolonged chest tube drainage may be required.