Anesthesia for right-sided obstructive lesions

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Introduction

Right-sided obstructive congenital heart lesions encompass a wide variety of presentations of congenital heart disease. Some minimally affected teenagers or adults may present only with vague complaints of exercise intolerance or fatigue. At the other extreme, right-sided obstructive congenital heart disease (CHD) may be immediately apparent in the neonate who manifests severe cyanosis or congestive heart failure (CHF). All lesions of this category have the potential for right-to-left shunting of blood flow. The severity of the disease depends upon the degree of structural malformation of the heart and great vessels.

Congenital malformations that impede blood flow through the right heart may occur at a single or combination of critical anatomical areas. These include the right atroioventricular (AV) valve, the outflow tract of the right ventricle (RV), the pulmonary valve (PV), and the main pulmonary artery (MPA) and/or branch pulmonary arteries (BPAs). Commonly, congenital malformations affect several of these critical areas simultaneously, such as in the tetralogy of Fallot (TOF).

Malformations may occur directly as a result of aberrant movement of tissues during development, or indirectly as a result of impaired flow hemodynamics due to malaligned structural anatomy. Often the resultant congenital heart deformity is a combination of both processes.

Patients with obstructive right-sided congenital heart anomalies may present in the neonatal period with either cyanosis or CHF. Right-sided lesions, which have potential for right-to-left shunting, may produce cyanosis, such as with right-to-left shunt through an atrial septal defect (ASD) or patent foramen ovale (PFO) in severe Ebstein’s anomaly or through a ventricular septal defect (VSD) in TOF. The shunt direction may vary, becoming right-to-left as right-sided pressures exceed those in the comparable left-sided chamber, providing a “pop-off” mechanism for right-sided obstructive hypertension. Neonates with restrictive right-to-left communications or without anatomical potential for shunt develop congestive right heart failure. Infants with obstructive right-sided lesions such as critical pulmonary stenosis (PS) or pulmonary atresia (PA) may be ductal-dependent, achieving pulmonary flow either in part or entirely from a patent ductus arteriosus (PDA). Unless patency is maintained by exogenous prostaglandin, increasing cyanosis may occur when the ductus arteriosus begins to close shortly after birth.

The physiology of right-sided obstructive defects and the changes that occur with surgical intervention in the context of perioperative anesthetic care and planning for such patients are described in this chapter for Ebstein’s anomaly, TOF, PS/PA with intact ventricular septum (IVS), and PA/VSD with multiple aortopulmonary collateral arteries (MAPCAs). Other right-sided obstructions such as those that result in a single functional ventricle (e.g. tricuspid atresia) are covered elsewhere in this volume.

Ebstein’s anomaly

Anatomy

Ebstein’s anomaly is by far the most common congenital malformation of the tricuspid valve (TV). The earliest description of TV malformation was by Ebstein in 1866.1 Ebstein’s anomaly is present in only about 0.3–0.7% of patients with CHD and occurs in approximately 1 in 20 000 live births.2 Other tricuspid anomalies such as TV stenosis, TV insufficiency (TI), and various malformations of leaflets, chordae tendoneae, and papillary muscles are much less common.3

Ebstein’s anomaly consists of: (i) a downward displacement of septal and posterior leaflet attachments at the junction of the inlet and trabecular portions of the RV; (ii) an “atrialized” portion of the RV between the tricuspid annulus and the attachment of the posterior and septal leaflets; and (iii) a malformed RV chamber (Fig. 20.1). The dysplastic characteristics of the anomaly are quite variable in functional severity, leading to a wide range of functional presentations from infancy to adulthood.
The position, size, and shape of the posterior and septal leaflets are quite variable. The posterior and septal leaflets may insert at varying distances below the AV annulus or may be closely adherent to the ventricular wall rather than displaced in one-third of patients. Shortened chordae often attach to papillary muscles that may be deformed. Over one-third of the hearts have an ASD, while most of the remaining two-thirds contain a PFO. The anterior leaflet is attached at the AV annulus superior to the other leaflets, but it is always abnormal. It is often large and redundant, shaped like a sail, with abnormal attachments to the border of the inlet and trabecular portions of the RV. The anterior leaflet and/or the chordae can act as a barrier to blood flow from the atrium/atrialized RV to the trabecular RV. The aperture between the atrialized and trabecular portions of the RV may be restricted to slits or perforations in the anterior leaflet. As a result of the distally displaced valves the trabecular portion of the RV is often very small, lacking an inlet chamber. The walls of the RV may be normal, or thin with impaired contractile function.

The RV wall above the line of insertion of the distally displaced leaflets functions as part of the right atrium (RA), but is anatomically ventricular. This inlet portion of the RV is often thin and dilated. Although it is exposed to atrial pressures, this atrialized RV manifests electrical conduction of an abnormal ventricular pattern. In some cases, the walls of the inlet portion is so thin that it moves paradoxically during ventricular systole, dilating with RA contraction. The RA is dilated, sometimes massively.

Left ventricular geometry may be compromised by the abnormal position of the interventricular septum, resulting in a small left ventricle (LV) chamber. In addition, mitral valve prolapse may occur because the chordae tendoneae of the normally situated mitral valve leaflets are altered in shape and size by the LV distortion.

Pathophysiology and natural history

The clinical presentation of Ebstein’s anomaly varies greatly depending upon the extent of the downward displacement of the TV leaflets. Severe hemodynamic compromise may present in the neonate, and when it does death may occur from massive right heart failure, hypoxemia and arrhythmias. The neonate with Ebstein’s anomaly shows rapid improvement of hemodynamics in the postnatal period due to gradual reduction of pulmonary vascular resistance (PVR). Ebstein’s cases with lesser anatomical aberration may have no signs during the neonatal period and only mild to moderate signs and symptoms later in childhood. Unless the foramen ovale is not patent, there is little exercise intolerance. Paroxysmal supraventricular tachycardia may occur in up to 20–25% of children, but other electrophysiologic abnormalities are also common (Table 20.1).

Without surgical intervention, death from Ebstein’s anomaly is usually secondary to CHF in the second or third decades of life. The more severe the cyanosis in the child or young adult, the poorer the prognosis. The onset of CHF often is a harbinger of death within a few years.

Surgical approach

The natural history of the disease varies with its severity and accordingly, the management of Ebstein’s anomaly is based on its severity. The size of the trabecular portion of the RV usually determines whether the patient is eligible for a two ventricular, one and a half ventricular, or single ventricular repair/palliation.

The first TV replacement was performed in 1963 as valvuloplasty techniques were rapidly evolving. Large numbers of patients have survived with a valvuloplasty technique.
Dysrhythmias are often problematic after surgical repair of Ebstein's anomaly, and temporary pacing wires placed on the RA and RV during surgery may be useful in some patients for monitoring of rhythm postoperatively or pacing. For teenaged and adult patients with preoperative arrhythmias, intermediate follow-up post-repair indicates substantial reduction of arrhythmia in survivors who did not require placement of pacemakers.\(^9\) Outcomes analysis\(^10\) has shown a hospital mortality of 10% (largely due to acute postoperative RV failure), but a long-term actuarial survival of 75% at 10 years for children and adults (no infants). High-risk patients (severely impaired RV function, difficult tricuspid valve repair, and/or permanent atrial fibrillation) seemed to benefit from a cavopulmonary anastomosis.

Surgical intervention is infrequently necessary in the infant and child unless tricuspid incompetence results in progressive right heart failure. Of 189 patients aged 11 months to 64 years, 75% had successful atrial plication and annuloplasty, with the remainder having atrial plication and placement of a bioprosthetic valve. Operative mortality was 4.9%. Carpentier\(^6\) et al.\(^6\) (Fig. 20.2) and Quaegebeur\(^7\) et al.\(^7\) (Fig. 20.3) have described variations on the Danielson repair. Quaegebeur reported nine of 10 patients had good TV function on intraoperative echocardiography, and that seven of the nine continued to have reduced TV regurgitation by echocardiography on follow-up (2–23 months, mean 11.7 months).\(^8\)

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**Table 20.1** Major electrophysiologic abnormalities in Ebstein’s anomaly.

- Intra-atrial conduction disturbance—“RA P wave abnormalities,” PR interval prolongation
- AV nodal conduction disturbance—PR interval prolongation
- Infranodal conduction disturbances
  - (a) Intra- or infra-His disturbances
  - (b) Right bundle branch block
  - (c) Bizarre “second” QRS attached to preceding “normal” complex
- Type B Wolff–Parkinson–White
- Supraventricular tachycardia
- Atrial fibrillation or flutter
- Electromechanical dissociation in atrialized RV
- Irritability of atrialized RV
- Q waves in leads V\(_1\)–V\(_4\)

RA, right atrium; RV, right ventricle.


**Fig. 20.2** Carpentier repair of Ebstein’s malformation. (a) Anterior and posterior leaflets of tricuspid valve are detached at annulus. (b) Atrialized chamber is obliterated in a circumferential direction. The anterior and posterior leaflets are reattached to the new, smaller annulus. Modified with permission from Casteñeda AR, Jonas RA, Mayer ME, Jr, Hanley FL. Cardiac Surgery of the Neonate and Infant. Philadelphia, PA: Saunders, 1994: 279.

**Fig. 20.3** Operative technique as described by Quaegebeur et al.\(^8\) (a) Surgeon’s view after opening the right atrium. a, anterior leaflet of the tricuspid valve; ac, atrialized ventricular chamber; p, posterior leaflet. (b) Detachment of the anterior and posterior tricuspid valve leaflets and their chordal attachments to the ventricular wall. The dashed lines denote the suture insertion points. (c) Longitudinal plication of the atrialized portion of the right ventricle. (d) Clockwise spread of the anterior and posterior leaflets on the newly created tricuspid valve annulus, and direct closure of the atrial septal defects without right atrium reduction. Modified with permission from Quaegebeur JM, Sreeram N, Fraser AG et al. Surgery for Ebstein’s anomaly: The clinical and echocardiographic evaluation of a new technique. \(J\) Am Coll Cardiol 1991; 17: 722–8.
cardiac output (CO), resection of redundant atrialized RV tissue and realignment of TV leaflets or placement of a prosthetic TV have all provided reasonable surgical outcomes.

Although severe TV dysplasia in the neonate is often not reparable with surgical valvuloplasty, a recent report suggests that aggressive two ventricular repair can be successful in some cases. Favorable 5 year follow-up was found for three neonatal repairs that included reconstruction of a monocuspid TV, ventriculorrhaphy, reduction atrioplasty, subtotal closure of the ASD, and repair of other associated defects. In the early 1990s, the Starnes procedure was proposed for prostaglandin E1 (PGE1) dependent neonates with Ebstein’s anomaly and physiologic PA, converting the cardiac physiology effectively to that of the single ventricular system. A pericardial patch was placed over the TV in such a fashion as to include the coronary sinus on the ventricular side, the foramen ovale was enlarged and the RA free wall plicated. A central 4 mm Gore® aortopulmonary shunt was placed. Ultimately, infants were able to undergo definitive surgical palliation with a Fontan procedure.

Patients with a severely hypoplastic or poorly functioning RV may ultimately require a single ventricle repair with cavopulmonary anastomosis or Fontan circulation. However there are instances when a hypoplastic or small RV is still capable of ejecting partial CO to the pulmonary arteries. These patients may benefit from a one and a half ventricular repair, allowing the diminutive RV to pump part of the systemic venous return to the lungs. The venous drainage of the upper body returns by passive flow via a cavopulmonary anastomosis to the pulmonary circulation. In brief, the one and a half ventricular repair includes valvuloplasty, possible repair of the ASD, and creation of a cavopulmonary anastomosis. A small ASD may be left if there is an anticipated need for a “pop-off” for systemic venous return to the “half” pulmonary ventricle. The pulmonary arteries must be of adequate size and PVR must be low for successful implementation.

Advantages exist in utilizing a semifunctional pulmonary ventricle. Preservation of some pulsatile flow to the pulmonary artery may, possibly, reduce the risk of development of MAPCAs. Also, a hypoplastic pulmonary ventricle may be able to respond to increased demand by increasing CO beyond what might result with a Fontan circulation. Van Arsdell et al. have proposed that the one and a half ventricular repair may be of benefit to the patients with Ebstein’s anomaly who have a partial RV outflow tract (RVOT) obstruction due to billowing of the anterior leaflet.

Reported mortality with the one and a half ventricular repair for all lesions (including Ebstein’s anomaly) is variable between 0% and 12%. Long-term outcomes have not been compared to the Fontan procedure, but the one and a half ventricle repair seems not to have the short-term and intermediate-term complications of cyanosis, chronic atrial arrhythmias, and protein-losing enteropathies associated with the Fontan physiology. However, an increase in perioperative effusions and chylothorax has been found. Other complications have included chronically increased superior vena cava (SVC) pressure, early-morning periorbital edema, and one instance of a SVC aneurysm. Another instance is reported of development of pulmonary arteriovenous fistulas with a one and a half ventricle repair in combination with the classic Glenn procedure.

Decision-making for the type of surgical repair or palliation relies on two critical assessments: the morphology of the TV and the size of the pumping chamber of the pulmonary ventricle. Valvuloplasty is preferred in infants and young children due to the need to upsize valves as the child grows. The teenager who has reached near adult size may do better with a prosthetic valve as the native valve may have incurred much damage due to abnormal dynamics over time. Patients with less than adequate pumping chambers will generally present for definitive surgical management in infancy or early childhood.

Perioperative anesthetic management

Preoperative

Pre-anesthetic evaluation of the child or infant with TV abnormalities includes an assessment of severity of the disease. Specifically, the patient is assessed for symptoms of fatigue, dyspnea, and increasing frequency and severity of cyanotic episodes. An assessment of exercise tolerance may often be delineated in reference to the child’s healthy peers. In the infant, questions are focused on the usual baby activities; poor feeding ability, failure to thrive, and/or signs of dyspnea, irritability, cyanosis, or diaphoresis are indicative of a poorly functioning heart. A history of syncope, chest pain, and palpitations suggests arrhythmia in the older child.

With Ebstein’s anomaly, physical examination may be notable for triple or quadruple heart sounds, often with a soft, high-pitched systolic murmur. A soft, scratchy mid-diastolic murmur heard best at the left sternal border and apex may be present. The second heart sound is widely split with little respiratory variation due to delayed emptying of the RV. With failure, the child may be diaphoretic, tachypneic, and irritable with rales present on chest auscultation and hepatomegaly on abdominal palpation. The chest roentgenogram may reveal moderate to severe cardiomegaly with a large RA and diminished pulmonary vascular markings. The heart often has a globular shape. Electrocardiogram (ECG) usually suggests RA hypertrophy, an increased PR interval, and complete or incomplete right bundle branch block. Interestingly, the pre-excitation patterns of Wolff–Parkinson–White syndrome are seen in 10–15% of individuals. Two-dimensional echocardiography is usually diagnostic, revealing a large tricuspid orifice complete with apical displacement of the septal leaflet of the TV. Cardiac catheterization is seldom indicated and may be complicated by induction of tachyarrhythmias.
Anxiolysis may be accomplished with midazolam, either given orally (0.5–0.75 mg/kg up to 15–20 mg) or intravenously (0.1–0.15 mg/kg up to 2–4 mg). Infants who manifest stronger fear (approximately 9 months of age and older) may also benefit from such sedation.

Intraoperative

Inhalation induction of anesthesia may be accomplished with nitrous oxide and sevoflurane for those infants and children with mild to moderate disease. Lowered CO or a small right-to-left shunt at the atrial level may slow induction by the inhalation route. Alternatively, intravenous induction with ketamine (1–4 mg/kg) or thiopental (4 mg/kg) will be consistent with reasonable induction hemodynamics. For patients with moderate to severe TV pathology, intravenous induction with glycopyrrolate and ketamine (1–4 mg/kg) can be accomplished smoothly in most instances without excessive myocardial depression or reduced afterload. Etomidate may also be used as an alternative (see Chapter 4). Since these patients are dependent upon adequate preload, increases in vascular compliance due to anesthetic vasodilation need to be met with intravenous volume replacement such as with 5% albumin. Choices of muscle relaxant depend upon the expected duration of the procedure and the need for rapid sequence or modified rapid sequence induction techniques. Pancuronium, a long-acting muscle relaxant, is sufficient for most cases and provides vagolysis via ganglionic blockade for a sustained increase in baseline heart rate. The maintenance technique is largely narcotic based (fentanyl 30–50 µg/kg) with low dose isoflurane (e.g., 0.4%) for myocardial preconditioning prior to institution of cardiopulmonary bypass (CPB). For repeat sternotomy, antifibrinolytic drugs such as plasmin binding inhibitors (e.g., ε-aminocaproic acid) or the plasmin active site inhibitor, aprotinin, may reduce blood loss during the pre- and post-CPB period.

Five-lead ECG with an ability to display multiple lead tracings is useful in monitoring changes in rhythm both during the pre- and post-repair periods. Other than standard monitoring, near-infrared spectroscopy is useful for monitoring brain tissue oxygenation during periods of cannulation and CPB. Transcranial Doppler-flow velocity may provide alternative information about cerebral blood flow during cannulation and CPB as well (see Chapter 8).

Patients with severely dilated right hearts are at high risk for potentially lethal ventricular arrhythmias post-repair. Prior to separation from CPB, intravenous infusion of an antiarrhythmic agent such as lidocaine or amiodarone may prophylactically protect against ventricular arrhythmias. Inotropic support that encourages forward flow in the right heart (e.g., milrinone 0.3–0.5 µg/kg/minute or dobutamine 5 µg/kg/minute) may improve hemodynamics for hearts with pre-existing myocardial dysfunction in the post-CPB period. Generous RV filling pressures may be needed to maintain adequate preload with a poorly functioning ventricle.

Postoperative

At the end of surgery, patients are transported to the cardiovascular intensive care unit (ICU) with continuous monitoring for rhythm and arterial blood pressure. Pain can be well controlled with narcotic infusions such as morphine sulfate (20–80 µg/kg/hour, depending on the need for sedation beyond analgesia). Patients with minimal pre-existing myocardial dysfunction may be weaned from mechanical ventilation and extubated within hours of arrival in the cardiovascular ICU. For other patients, it is prudent to allow the patient to emerge more slowly from narcotic sedation and inotropic support in order to assess the remodeled tricuspid competency and allow more time for recovery of myocardial function. Midazolam (0.1–0.2 mg/kg/hour) may be added simultaneously with narcotic analgesic infusion to provide long-term sedation for patients who need longer myocardial recovery times (beyond 1–2 days) (see Chapter 27).

As mentioned previously, dysrhythmias are common in the immediate postoperative period after repair of Ebstein’s anomaly, and may persist as a late complication of repair. Supraventricular tachycardia, junctional rhythm, or intermittent AV block may complicate recovery. Risk for ventricular arrhythmias and sudden death persists through the first postoperative month. Those patients who demonstrate perioperative ventricular tachycardia or ventricular fibrillation are likely at greatest risk. Patients with intermittent AV block or junctional rhythm may benefit from temporary pacing to enhance CO in the immediate postoperative period. As myocardial edema subsides, return of functional conduction pathways may allow return of normal sinus rhythm. As mentioned above, intravenous amiodarone or lidocaine may be helpful in the early postoperative period, and switching to oral amiodarone for several months may be warranted for high risk individuals.

In the early postoperative period, echocardiography often shows poor coaptation of the TV leaflets. This finding is likely due to post-bypass dysfunction of the papillary muscle bundles (possibly of ischemic etiology) as the leaflet coaptation often improves with subsequent echocardiographic examinations.

Tetralogy of Fallot

Anatomy

Tetralogy of Fallot represents 10% of all congenital heart defects and is the most common form of cyanotic heart disease. A Danish scholar, Nicholas Steno, first described the defect in 1673, 200 years before Fallot. One hundred years
Coronary abnormalities occur in 5–12% of patients with TOF. Failure to detect these preoperatively can have serious consequences for a successful outcome because they may be damaged during surgery. The most common abnormality consists of a left anterior descending artery that originates from the right coronary artery, and crosses the RVOT inferiorly. This arrangement makes it very susceptible to damage if the transannular incision is carried too far inferiorly across the RVOT. Indeed, an alternative surgical approach may be needed in relieving the subpulmonary obstruction, or a RV to MPA conduit may be required. Other coronary anomalies include a right coronary artery originating from the left coronary artery, and left coronary artery originating from the MPA. Precise definition of the coronary anatomy may be possible with echocardiography alone. If there is uncertainty, then aortic root or selective coronary angiography may be used.

Other coexisting cardiac lesions include left SVC (LSVC), AV septal defect, PDA, ASD, and interrupted inferior vena cava. All of these require modifications to the surgical repair. It is particularly important to be aware of the presence of a LSVC because this usually drains into the coronary sinus in the RA. During surgery a venous drainage cannula is placed in the LSVC and this vessel is usually tied off at the end of the procedure. Therefore, a central venous line placed via the left internal jugular route will be rendered useless postoperatively.

Two important variants of TOF are PA with VSD and the absent PV syndrome. Pulmonary atresia with VSD is characterized by hypoplasia of the central and peripheral pulmonary arteries. The MPA may be absent or the branch PAs may be non-confluent or stenotic. Pulmonary blood supply is usually via MAPCAs. The surgical correction of this lesion is very different from that of classical TOF as described later in this chapter. The absent PV syndrome is characterized by combined PS and incompetence which in utero leads to increased pulsatile pulmonary blood flow (PBF) producing massive enlargement of the main and branch PAs. This produces the central characteristic feature of airway compression and tracheobronchomalacia. These babies typically present in the neonatal period with severe respiratory distress, cyanosis, and air trapping. Tracheal intubation with high levels of positive end-expiratory pressure (PEEP) may be useful in stenting the airways. Prone positioning may also be useful in relieving some of the obstruction. Infants with significant lung disease require urgent surgical intervention. However, symptoms commonly persist due to the underlying intrinsic airway abnormalities and such patients may need long-term ventilator support.

In the common form of TOF, the obstruction to the RVOT usually has dynamic and fixed components. The dynamic component consists of hypertrophied infundibulum and muscle bundle fibers in the RVOT. The hypertrophy occurs in response to the pressure load on the RV. Fixed obstruction

![Fig. 20.4 Schematic diagram of tetralogy of Fallot illustrating the infundibular stenosis (stippled area), ventricular septal defect, overriding aorta, and right ventricle hypertrophy.](image-url)
may be valvar, consisting of a thickened, hypoplastic, and often bicuspid PV. In 15–25% of patients the PV is atretic producing complete obstruction to the RVOT. These patients require an alternative of PBF such as a PDA, bronchial arteries or MAPCAs.

Beyond the PV, impedance to RV outflow occurs due to abnormalities in the pulmonary arterial tree. There is usually some degree of central pulmonary artery hypoplasia in all patients. There may also be localized narrowing of the MPA or BPAs. Atresia or discontinuity of the MPA or BPAs may occur, which will further complicate surgical correction, as restoration of continuity or augmentation of the pulmonary arteries will be required.

There is a weak association of familial inheritance of TOF. Indeed TOF is associated with major extracardiac malformations and may occur as part of a syndrome. Some examples are the VACTERL (vertebral, vascular, anal, cardiac, tracheoesophageal, renal, and limb anomalies) association, DiGeorge syndrome, velocardiofacial syndrome, and CHARGE (coloboma, heart anomaly, choanal atresia, retardation, and genital and ear anomalies) association. Recent genetic studies have shown that TOF is associated with chromosome 22q11 deletion (catch 22 syndrome). This chromosomal abnormality is also responsible for DiGeorge syndrome, velocardiofacial syndrome, and conotruncal anomaly face syndrome. In one study of TOF patients, the prevalence of 22q11 deletion was 13%. This deletion is considered to be the most common genetic cause of TOF-associated syndromes.

Pathophysiology and natural history

The clinical manifestation of TOF ranges from extreme cyanosis at one end of the spectrum, because of profound right-to-left shunting through the VSD, to normal saturation for patients who have minimal RVOT obstruction and who exhibit a net left-to-right shunt. The latter group is sometimes known as “pink tets” because of the absence of cyanosis. They may even show signs of CHF from pulmonary overcirculation. The presentation of symptoms with TOF is determined primarily by the degree of RVOT obstruction because the large non-restrictive VSD effectively equalizes the pressures in both ventricles. If the VSD is restrictive RV pressure may be suprasystemic. The RV undergoes hypertrophy in response to the high afterload and the wall thickness may become similar to the LV. Right ventricle hypertrophy is undesirable for several reasons. It can lead to diastolic dysfunction; surgical correction of the VSD and RVOT obstruction becomes more difficult through a thickened ventricle; and it becomes more difficult to protect the hypertrophied RV during aortic cross-clamping, which may contribute to postoperative RV dysfunction. In order to limit RV hypertrophy, surgical correction is undertaken in early infancy.

With a non-restrictive VSD and equalization of RV and LV pressure, the main determinants of the degree of shunting are the relative resistances of the systemic and pulmonary circuits. Both of these change depending on various factors and, therefore, the degree of cyanosis will vary. Right ventricle outflow tract obstruction usually has a dynamic and fixed component; producing different propensities for cyanosis.

An acute form of this RVOT obstruction occurs during a hypercyanotic or “tet spell” producing severe cyanosis and hypoxemia, which can lead to syncope or a stroke. The obstruction is thought to result from infundibular spasm leading to an acute decrease in PBF and shunting of desaturated blood into the systemic circulation. These spells can occur spontaneously, but are usually in response to crying, defecation, agitation, injury or fright which increases sympathetic tone leading to increased contractility producing infundibular spasm. As well as acute increases in RVOT obstruction, “tet spells” may be precipitated by an acute fall in systemic vascular resistance (SVR). This may happen during induction of anesthesia (especially if the patient is hypovolemic), and it is very important that the anesthesiologist is well prepared in advance to treat a spell. The goal of treatment is to use maneuvers (described later in this chapter) which reverse the direction of right-to-left shunting. Other factors which can worsen cyanosis are anemia, acidosis, infection, stress and posture. “Tet spells” in the awake patient are usually accompanied by hyperventilation secondary to the metabolic acidosis and the hypoxemia. Children classically adopt a squatting posture during a spell to alleviate discomfort. Squatting increases intra-abdominal pressure leading to increased venous return and RV preload, which helps to “enlarge” the RVOT. This position also increases SVR, which favors left-to-right shunting.

Clinical features

The presenting features of TOF are variable depending on the degree of RVOT obstruction. Prenatal diagnosis is possible with ultrasonography. In the neonate, cyanosis and the presence of a murmur will lead to further diagnostic evaluation. In newborns with critical PS and ductal-dependent PBF, the clinical presentation may be delayed until ductal closure. Very rarely, the baby will be in CHF from pulmonary overcirculation if there is only mild RVOT obstruction—the so-called “pink tet.” Sudden severe desaturation will occur during a hypercyanotic spell characterized by infundibular spasm and severe reduction in PBF.

Physical findings are not specific for TOF. The degree of cyanosis will vary, and pulse oximetry will demonstrate low hemoglobin saturation. Clubbing is a relatively late finding. Cardiac auscultation reveals a crescendo–decrescendo systolic murmur best heard at the upper left sternal border. The intensity of the murmur will be diminished during a hypercyanotic spell. Chest radiograph shows a characteristic “boot
shaped” heart, which is a reflection of RV hypertrophy and a concave upper left heart border from a small or absent MPA. The lung fields are oligemic from diminished blood flow. The ECG usually shows RV hypertrophy and right axis deviation.

**Morbidity and mortality**

Survival beyond the fourth decade is very rare in untreated patients. Without surgery, 25–35% of children with TOF die in the first year of life, 40–50% by year 3, 70–76% by year 10, 90% by year 21, and 95% by year 40. The outcome is even worse for the subset of patients with PA. Without surgery, most of these children do not survive beyond infancy. Even those children who are completely palliated show delayed growth and development compared with their normal counterparts. This is usually due to the associated non-cardiac conditions. The clinical course of patients with TOF is determined primarily by the degree of PS. Mortality in untreated patients is usually a result of hypoxemia or its hematologic consequences, or the result of problems such as endocarditis or brain abscess. With complete repair in early infancy or childhood, over 85% of patients are expected to survive to adulthood.

**Surgical approach**

In the present era, most of the diagnostic information needed for surgical decision-making can be obtained from echocardiography, with cardiac catheterization only necessary in selected cases. As well as delineating the anatomy of the heart chambers, PV, MPA, BPAs, and aortic arch, Doppler studies can demonstrate the severity of RVOT obstruction and the location, type, and number of VSDs. In addition, it aids in localizing alternative sources of PBF. Coronary anatomy can also be described in many cases. However, echocardiography cannot reliably image pulmonary artery anatomy beyond the proximal branches. If echocardiography is insufficient in providing all the information needed for determining the surgical plan, then cardiac catheterization and angiography are performed. If only a palliative procedure is planned, echocardiography may be adequate. However, for planned complete surgical correction, and for patients who have had a palliative procedure (where there may be distortion of the vascular anatomy), cardiac catheterization may be indicated.

Cardiac catheterization is not a benign procedure in the tetralogy patient, especially for those at high risk for hypercyanotic spells which may be precipitated by manipulation of the catheter across the RVOT. Such patients should be managed by experienced personnel with appropriate emergency drugs and equipment immediately available for the management of “tet spells” in the catheterization laboratory.

Interventional catheterization procedures are having an increasing role in the management of CHD. In some centers, balloon dilation of the RVOT is considered as a palliative alternative to a systemic to pulmonary artery shunt. Correct technique avoids the disadvantages of surgical intervention, including distortion of pulmonary vascular anatomy, which can occur with shunting procedures. Balloon dilation and stent placement also has an important role in those patients who have undergone complete surgical correction, but are left with residual pulmonary artery or conduit stenosis.

All patients diagnosed with TOF require some form of surgical intervention. However, there is ongoing debate regarding the timing of repair, and whether this should be done as a staged technique (palliative shunt followed by complete repair) or primary total repair. There is a trend now towards early total correction, whether the patient is asymptomatic or not, with some institutions performing surgery in the neonatal period with good results. Others prefer to adopt a slightly more conservative approach, opting for repair at 3–6 months of age or even later. Other factors to consider are the institution’s capability of providing peroperative critical care of neonates and small infants undergoing complicated cardiac surgery, or specific anatomical features that are contraindications to primary repair. Examples of unfavorable anatomy include the presence of coronary abnormalities such as the left anterior descending arising from the right coronary and crossing the RVOT, the presence of multiple VSDs, and inadequate pulmonary artery anatomy. In these cases it is reasonable to place a palliative shunt and allow the baby to grow, facilitating the eventual complete repair. The two-stage approach does have certain disadvantages. It subjects the baby to an additional surgical procedure with attendant risks and complications. These include injury to the recurrent laryngeal and phrenic nerves, inadequate or excessive PBF requiring subsequent shunt revision, potentially fatal shunt obstruction, and potential distortion of the pulmonary artery at the anastomotic site. If the shunt is placed centrally through a median sternotomy, subsequent surgery may be more hazardous due to increased risk of damage to the heart or major vascular structures during dissection. As far as timing of surgery, there are certain disadvantages to operating in the neonatal period. In addition to the risks of performing complicated surgery on tiny neonates and the effects of CPB (and possibly circulatory arrest) on immature organ systems, the surgical procedure is technically more challenging. Although most centers perform the repair using a transatrial–transpulmonary approach, smaller patients more commonly require a ventriculotomy to facilitate repair. Ventriculotomy may result in late RV dysfunction and dysrhythmias. Nevertheless, some centers strongly favor this approach. Proponents of early repair point out the desirability of operating before significant RV hypertrophy has occurred. Right ventricle hypertrophy may require extensive myectomy, making VSD closure more difficult. Severe RV hypertrophy may also impair myocardial protection during the period of aortic cross-clamping. Early repair may also lead to more normal growth and development of
There are three major goals of the TOF repair: (i) maximal relief of RVOT obstruction; (ii) separation of the systemic and pulmonary circulation by closure of the VSD; and (iii) preservation of RV function in the short and long term. It is particularly important to delineate the coronary anatomy, determine the levels of obstruction of the RVOT, and the size and continuity of the pulmonary arteries. The details of the operative procedures are well described elsewhere. After cardioplegic arrest the repair is done using a transatrial–transpulmonary approach. Right ventriculotomy is avoided if possible to preserve RV function. The PV is examined through a longitudinal incision in the MPA, and if necessary, a commissurotomy is performed. The RVOT is exposed through the RA and TV and resection of the infundibular septum is carried out. Hegar dilators are passed through the TV into the MPA to calibrate the RVOT. If the size of the RVOT is judged to be inadequate, the MPA incision is extended downwards across the annulus and onto the RV free wall. The VSD is then closed through the RA. Any ASD is also closed although some surgeons prefer to leave an atrial communication to act as a “pop off” valve in case of severe RV dysfunction postoperatively. This will produce some hypoxemia via right-to-left shunting. The adequacy of repair is assessed using several methods. The RV : LV pressure ratio can be measured, less than 0.75 being considered acceptable. It is important to emphasize, however, that pressure measurements in the early post-CPB period do not reflect measurements made at follow-up and may lead to unnecessary revisions of the repair. Transesophageal echocardiography (TEE) is useful in demonstrating gradients across the RVOT, showing residual VSDs, and assessing ventricular function. Blood gas measurements from the vena cava and the MPA are also useful in detecting residual shunts.

In patients who have a coronary artery which crosses the RVOT, a transatrial–transpulmonary repair is still feasible if the transannular incision is limited. Many of these patients, however, require a valved RV–PA conduit to avoid damage to the coronary artery.

Surgical palliation

The proposal by Blalock and Taussig to anastomose the subclavian artery to the pulmonary artery in an end-to-side fashion to alleviate cyanosis resulted in the first successful palliation of TOF in 1944. This “classic” Blalock–Taussig (BT) shunt is very rarely used today. Potts and Waterston later described direct aorta to pulmonary artery shunts. Although these shunts provided good palliation, their size was difficult to control and commonly resulted in too much PBF. They were also extremely difficult to take down during corrective surgery and thus were largely abandoned. A “modified” BT shunt (MBTS) is created by interposition of a graft of synthetic material, usually polytetrafluoroethylene, between the subclavian artery or brachiocephalic artery and the ipsilateral pulmonary artery. This is the most common palliative procedure carried out in the current era (Fig. 20.5). The advantages of the MBTS are many: (i) it preserves blood flow to the arm; (ii) it can be used on either side, although most are done on the right side because the pulmonary anastomosis can be placed more centrally allowing easier control of the shunt during subsequent repair; and (iii) it avoids excessive PBF when appropriately sized. A central shunt, between the ascending aorta and the MPA, using graft material is used as an alternative to the MBTS when the vascular anatomy precludes placement of the latter.

Complete surgical repair

Lillehei carried out the first successful repair of TOF in 1954. There are three major goals of the TOF repair: (i) maximal relief of RVOT obstruction; (ii) separation of the systemic and pulmonary circulation by closure of the VSD; and (iii) preservation of RV function in the short and long term. It is particularly important to delineate the coronary anatomy, determine the levels of obstruction of the RVOT, and the size and continuity of the pulmonary arteries. The details of the operative procedures are well described elsewhere. After cardioplegic arrest the repair is done using a transatrial–transpulmonary approach. Right ventriculotomy is avoided if possible to preserve RV function. The PV is examined through a longitudinal incision in the MPA, and if necessary, a commissurotomy is performed. The RVOT is exposed through the RA and TV and resection of the infundibular septum is carried out. Hegar dilators are passed through the TV into the MPA to calibrate the RVOT. If the size of the RVOT is judged to be inadequate, the MPA incision is extended downwards across the annulus and onto the RV free wall. The VSD is then closed through the RA. Any ASD is also closed although some surgeons prefer to leave an atrial communication to act as a “pop off” valve in case of severe RV dysfunction postoperatively. This will produce some hypoxemia via right-to-left shunting. The adequacy of repair is assessed using several methods. The RV : LV pressure ratio can be measured, less than 0.75 being considered acceptable. It is important to emphasize, however, that pressure measurements in the early post-CPB period do not reflect measurements made at follow-up and may lead to unnecessary revisions of the repair. Transesophageal echocardiography (TEE) is useful in demonstrating gradients across the RVOT, showing residual VSDs, and assessing ventricular function. Blood gas measurements from the vena cava and the MPA are also useful in detecting residual shunts.

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Perioperative anesthetic management

Management for surgical palliation

Many of these patients are critically ill due to severely reduced PBF. They may be mechanically ventilated and receiving an infusion of PGE1 to maintain ductal patency. If intravenous access is available then anesthesia can be induced with a combination of ketamine and fentanyl and maintained with low concentrations of a volatile agent. It is important to maintain adequate SVR in order to limit right-to-left shunting through the VSD; in this regard, sevoflurane is a good choice as this agent has the least effect on SVR. The myocardial depressant effect of volatile agents is also useful in limiting infundibular spasm. Low SVR is treated with phenylephrine or...
norepinephrine; and preload is augmented with fluid boluses. It is important to avoid most inotropes, as these will worsen infundibular spasm by increasing heart rate and contractility. If there is no intravenous access, induction can be carried out rapidly and smoothly with sevoflurane. An alternative is to use intramuscular ketamine in unstable patients. Central venous access is obtained for the infusion of fluids and vasoactive agents. A radial arterial line is placed on the side opposite to that of the MBTS in order to get a true assessment of the blood pressure because after the shunt is opened there may be significant “steal” from the ipsilateral subclavian artery. A femoral arterial line can also be placed as long as care is taken to observe for evidence of distal lower extremity ischemia.

Most MBTS are performed via a thoracotomy. The median sternotomy approach is used when the surgeon feels that the patient will not tolerate lung retraction or side-clamping of the PA, when there is a possibility that CPB may be required, and for central shunt placement. Low dose heparin (100 U/kg) is administered prior to shunt placement. Lung retraction can severely impair oxygenation and ventilation, and intermittent reinflation may be required. Similar decompensation can occur during partial clamping or obstruction of the PA during the construction of the anastomosis. Such decompensation is managed with fluids, vasopressors, and ventilation adjustments. However, attempting to normalize $P_{\text{CO}}$, during single lung perfusion may overdistend the dependent lung, increasing $P_{\text{VR}}$ and impairing venous return. For central shunts, partial clamping of the ascending aorta is required, but it may be poorly tolerated in the presence of LV dysfunction. Inotropic support with dopamine is usually helpful. Once the shunt is open, oxygen saturation usually improves immediately. However, blood pressure may drop significantly, requiring volume infusion and vasopressors. If the diastolic pressure becomes very low, coronary flow will be reduced and ischemic changes may be seen on the ECG. Ventilation and inspired oxygen are adjusted to mimic spontaneous, non-anesthetized values for an accurate assessment of the shunt flow. An oxygen saturation of near 80% is optimal as this represents balanced pulmonary and systemic blood flow. A high saturation suggests pulmonary overcirculation and the shunt size may have to be reduced. Conversely, a low saturation suggests inadequate $P_{\text{BF}}$, and a larger diameter shunt may be needed. In cases of persistent hypoxemia after apparently uneventful shunt placement, it is important to rule out the possibility of endobronchial intubation because failure to do so may lead to unnecessary shunt revision or even sternotomy. After chest closure, the patient is transferred to the cardiac ICU where mechanical ventilation is typically necessary for at least 12–24 hours. Increased $P_{\text{BF}}$ can cause unilateral pulmonary edema or pulmonary hemorrhage. Diastolic hypotension may cause myocardial ischemia, requiring close monitoring of inotropic medications and volume. Other complications include injury to the phrenic and recurrent laryngeal nerves, Horner’s syndrome, chylothorax, and shunt thrombosis. Patency of the shunt can be clinically confirmed by briefly disconnecting the patient from the ventilator and auscultating over the end of the endotracheal tube. The murmur is transmitted via the tracheal tube due to the proximity of the shunt to the bronchus. A low dose heparin infusion is started (8–10 U/kg/hour) to maintain shunt patency when the risk of post-surgical hemorrhage has diminished. After enteral intake has begun, the patient is prescribed aspirin until the time of corrective surgery. Platelet transfusions are generally avoided for patients undergoing shunt placement due to the risk of shunt thrombosis.

### Management for complete repair

Procedures for complete repair necessitate additional considerations of the effects of CPB. The anesthetic induction does not differ from that described above. Generally, we utilize a total dose of fentanyl to 20–50 µg/kg and administer inhalational agents to supplement anesthesia. A recent study showed that a ketamine infusion provided more hemodynamic stability by preserving $SVR$ in the pre-CPB period when compared with isoflurane.\(^3\)\(^9\) The lower dose of fentanyl usually allows for extubation within 4–8 hours after surgery. Transesophageal echocardiography is used for almost all patients. However, it is important to monitor the effects of probe insertion on ventilation. The TEE probe may compress the trachea or mainstem bronchi, compromising ventilation, requiring removal. If TEE is not possible or unavailable, epicardial echocardiography can be performed post-CPB to assess repair. In addition to routine monitors, brain oxygen saturation trends can be followed with near infrared spectroscopy. Other monitoring alternatives include electroencephalography and transcranial Doppler. Neurologic monitoring is discussed elsewhere in this volume.

Patients who do not have a palliative shunt may develop a “tet spell” during the pre-CPB period and without prompt and aggressive treatment severe hypoxemia may progress to cardiovascular collapse. Particularly vulnerable periods are during anesthetic induction before surgical stimulation, when reduced sympathetic tone causes a fall in the $SVR$ leading to increased right-to-left shunting. Manipulation of the great vessels by the surgeon may also result in sudden right-to-left shunting. The primary goal of management of a spell is to correct the hypoxemia by relieving the infundibular spasm and reversing the shunt. Some or all of the following maneuvers can be employed:

1. **Administer oxygen.** This does not relieve the spasm but helps reduce hypoxic pulmonary vasoconstriction.
2. **Phenylephrine,** 5–10 µg/kg and titrated to effect to increase $SVR$.
3. **Volume infusion** to support the blood pressure and increase right heart filling thereby reducing RVOT obstruction during systole.
4. Compress the abdomen to directly compress the aorta and place the child in a knee–chest position to increase the SVR.

5. Titrate esmolol, 50 µg/kg to effect. The negative inotropic effect and reduction in heart rate will help to reduce the infundibular spasm. Propranolol (0.1 mg/kg given slowly) also works but is slower in onset.

6. Increase the depth of anesthesia with a volatile agent to decrease contractility thereby reducing the infundibular spasm. Although halothane is traditionally used for this purpose, a recent echocardiographic study showed that sevoflurane has less effect on the SVR index than halothane or isoflurane at 1.5 minimum alveolar concentration and therefore may be superior. Isoflurane is a poor choice because it is a potent vasodilator and also causes tachycardia, which increases contractility. Although morphine is frequently recommended for the treatment of “tet spells” in the awake patient, it produces excess vasodilation under anesthesia and is, therefore, not recommended.

7. If all these measures fail and the patient continues to deteriorate, the chest may have to be opened quickly, and the aorta may need to be compressed to reverse shunting.

During the rewarming phase of CPB, preparations are made for weaning from CPB. In general, three problems may be anticipated:

1. Right ventricle dysfunction, especially if the transannular incision was extended down the RV free wall. The mainstays of treatment are fluid loading to higher filling pressures, inotropic support, and reduction of RV afterload.

2. Arrhythmias and heart block. These are common after VSD repairs because of the close proximity of the conduction system. Epicardial pacing may be needed to accomplish weaning from CPB. In most instances heart block is a transient phenomenon due to the edema around the VSD patch. If it does not resolve after 7–10 days, permanent pacing may be required. Junctional ectopic tachycardia is seen occasionally, although the onset is usually 12–24 hours later. This is characterized by AV dissociation and rapid junctional rates as high as 200–230 beats/minute. Treatment consists of cooling the patient to 34–35°C, and drug therapy with amiodarone or procainamide. Atrial overdrive pacing can also be used to re-establish AV synchrony.

3. Post-CPB bleeding. Coagulopathy results from hemodilution of coagulation factors and the effects of CPB on platelet number and function, and may require transfusion of multiple component blood products. The use of antifibrinolytics such as ε-aminocaproic acid, or protease inhibitors such as aprotinin, may reduce post-CPB bleeding and minimize the use of blood products.

Once the chest is closed the patient is transferred to the ICU. Analgesia is provided with a continuous infusion of morphine at 20–40 µg/kg/hour, supplemented with intermittent boluses of midazolam for sedation. Hemodynamically stable patients with minimal bleeding are good candidates for early extubation, usually within 4 hours. After the patient is extubated, analgesia can be reliably provided with a combination of acetaminophen and a non-steroidal anti-inflammatory agent such as ibuprofen, with morphine as needed.

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**Pulmonary stenosis with intact ventricular septum**

**Anatomy**

Pulmonary stenosis/intact ventricular septum is relatively common, accounting for 8–10% of congenital heart defects. Pulmonary stenosis may be valvular, subvalvular, or supravalvular. In valvular PS, the PV is dome-shaped with a centrally placed orifice. The RV is usually normal in dimension with the exception of infundibular hypertrophy that occurs as a result of outflow obstruction. Valvar PS is frequently associated with Noonan’s syndrome. Although isolated subvalvular PS is rare, obstruction may occur within the RV cavity due to abnormal hypertrophied muscle bands, which run between the ventricular septum and the anterior wall, effectively dividing the RV cavity into a proximal high-pressure chamber and a distal low-pressure chamber (“double chambered RV”). Supravalvular PS involving the MPA may be seen with congenital rubella and Williams’ syndrome. The etiology of the defect is unknown, but there is likely a genetic factor as the incidence of the defect in siblings of the affected patient is 2–4%.

**Pathophysiology and natural history**

In its most severe manifestation, PS/IVS presents in the neonatal period with cyanosis and right heart failure. However, most children develop signs and symptoms more slowly, with the onset based on the severity of the PS and the relative sizes of a PFO or ASD. Many patients are initially identified by the presence of a harsh systolic ejection murmur and perhaps a thrill over the PV auscultation area. There is often post-stenotic dilation of the MPA and BPAs which may be visible on chest roentgenogram. Radiographic cardiomegaly is a late sign, coincident with signs of failure. Electrocardiogram often shows right axis deviation, prominent P waves, and evidence of RV hypertrophy. Echocardiography with Doppler evaluation of the valve gradient can be
used to measure the severity of the lesion and serial measurement used for follow-up studies. Cardiac catheterization, in addition to its value in obtaining further measurements, can also be used to perform balloon valvuloplasty.

Surgical approach
Symptomatic patients and those with severe gradients and impending RV failure are treated primarily with balloon valvuloplasty, which has replaced surgery as the first line treatment. In fact, repeat balloon valvuloplasty is employed for cases of recurrent stenosis. The incidence of pulmonary insufficiency after balloon valvuloplasty is 80%, but is usually mild in clinical severity. Surgical repair may be attempted with or without CPB via a median sternotomy approach. With the CPB technique, a transverse incision is made in the MPA. Fused valve leaflets are incised. The annulus may be enlarged with Hegar dilators. Subvalvar obstruction in the infundibular region may be excised. Rarely, a transcaval patch may be needed. A right atriotomy is used to close a PFO or ASD, and may also be used to perform an infundibular resection through the TV. Surgical pulmonary valvotomy may also be performed off CPB via a transventricular approach through a purse string suture in the anterior RV. Hegar dilators are inserted in increasing diameters across the valve. The CPB pump is kept primed and on standby for this approach. With either surgical technique, residual valve gradients may be measured utilizing needle pressure transducers.\(^{40}\)

Perioperative anesthetic management
Optimizing inotropic therapy, instituting adequate diuresis, and correcting metabolic acidosis and electrolyte abnormalities medically optimizes the patient with CHF prior to surgery. Neonates with critical PS should be stabilized with PGE, and taken for cardiac catheterization without delay. Most patients will be eligible for elective repair.

After CPB, attention is given to optimizing RV filling pressures and keeping pulmonary arterial pressures low to induce forward right-sided CO. Pulmonary vasodilators begun in the early postoperative course or in the late CPB period may increase pulmonary flow and reduce RV afterload. Although most patients tolerate pulmonary insufficiency that results from either open or closed pulmonary valvotomy, inotropic support is often needed to assist the transient RV dysfunction that often presents after anterior right ventriculotomy with the closed approach. Inotropic support is utilized cautiously in patients that may have a dynamic subvalvar obstructive component due to infundibular hypertrophy, but may be needed to achieve adequate RV function for a few days after repair. Residual infundibular hypertrophy often resolves with time after the valvular obstruction is relieved.

Pulmonary atresia with intact ventricular septum
Anatomy
Unlike PS/IVS, PA/IVS does not have a familial association. The defect comprises approximately 1.0–1.5% of congenital heart defects. Although the etiology of the defect is unknown, the inciting event appears to be severe intrauterine RVOT obstruction, leading to maldevelopment of the TV, RV, and coronary arteries. The degree of abnormality varies with the gestational age at which the RVOT obstruction occurred. Patients with a diminutive RV, small TV, and extensive RV to coronary artery communications would be presumed to have incurred PA at an earlier stage of gestation. Multiple morphologic abnormalities occur with this lesion, all of them proximal to the PV (in contrast to PA with VSD in which the major associated defects occur distal to the valve). There is almost always a PFO or secundum ASD, restrictive in 5–10%. The TV is usually smaller than normal, but may range from extremely stenotic to the dilated annulus of Ebstein’s anomaly (5–10%). The RA is dilated proportionately to the degree of tricuspid regurgitation. The RV is hypertrophic with reduced size of the cavity. In about 50% of cases, there are endothelial-lined blind channels within the RV myocardium known as sinusoids. These sinusoids are in direct communication with the RV cavity and may form coronary artery to RV fistulae. The prevalence of these sinusoids is inversely proportional to the diameter of the TV, RV cavity size, and magnitude of TI, but directly proportional to RV systolic pressure. In the least affected individuals, RV blood may be sent as part of a dual supply of blood to small areas of myocardium in tandem with normal aortocoronary flow. But approximately 20% of patients with PA/IVS have a RV-dependent coronary circulation with absence of anterograde aortocoronary flow. In these patients the coronary bed is perfused with desaturated systemic venous blood directly from the RV and, therefore, the myocardium may be chronically ischemic. Sometimes the PV is seemingly intact, but with fused commissures. Most often, there is a fibrous tissue at the ventriculoarterial junction. The pulmonary arteries usually have normal branching and may be hypoplastic in about 6% of cases. There is almost always a PDA. The LA is enlarged and hypertrophied, sometimes exhibiting fibroelastosis. Sub-aortic stenosis has occurred due to bulging of the ventricular septum into the LV from RV hypertension.\(^{41}\)

Pathophysiology and natural history
Untreated, PA/IVS results in death in 50% of neonates, and in 85% of infants by 6 months of age. Fetuses with small, hypertrophied ventricles often survive to birth; those with
dilated RVs and severe TI may die of fetal hydrops. In the presence of moderate to severe TI, RV pressure will remain low and sinusoids and coronary fistulae will not evolve. Alternatively, if TI is mild or nil, the RV will hypertrophy and remain small, developing systolic hypertension. The increased flow across the foramen ovale in utero causes a volume overload of the left heart, resulting in neonatal LV hypertrophy and dilation, and potential aortic root dilation.

The affected newborn is dependent upon the PDA and is resuscitated with PGE1. Generally, the left heart functions normally, and CO is maintained with the presence of an adequate PDA. If there is LV hypertrophy from septal hypertrophy/LV outflow tract obstruction, there may be coronary fistulae and resulting myocardial ischemia. Single ventricle physiology is manifested. Tricuspid valve insufficiency is common, partly because of the RVOT obstruction and, in approximately one-third of cases, due to structural abnormalities of the TV. Over 90% of patients will present with cyanosis and a ductal flow murmur within the first 3 days of life. Electrocardiogram reveals lack of RV forces and often a large P wave, indicative of RA enlargement. Chest roentgenogram can show decreased to normal pulmonary vascular markings depending upon the amount of ductal flow. The cardiac silhouette is normal unless RA and RV enlargement occur due to severe TI. Echocardiography can define the RVOT, RV dimensions, the TV, and the PDA. Right ventricle pressure can be derived from Doppler measurement of the TI. Ventricular function can be assessed, but dependency of coronary blood flow cannot be determined solely with echocardiography. Cardiac catheterization is essential in all cases to define major stenoses and fistulae in the coronary anatomy.

Surgical approach

In the early 1960s, palliative shunts and closed pulmonary valvotomies were done. But survival was dismal given that an estimated 2.5% of patients lived 3 years of age. Right ventricle outflow procedures were combined with systemic to pulmonary shunts in the 1970s. Since that time, repair techniques have varied among surgeons, partly based on the spectrum of anatomic dysmorphology and partly on the individual surgical outcome experiences. Current corrective procedures include: (i) neonatal RVOT patch augmentation with continued infusion of PGE1 (average of 6 days); (ii) neonatal RVOT patch augmentation with concurrent systemic to pulmonary artery shunt; and (iii) pulmonary valvotomy (open or closed) and a systemic to pulmonary artery shunt. Success rates in achieving ultimate biventricular repair have varied from 40% to 60%. However, all congenital heart surgeons generally avoid RV decompression if there is a complete dependency of myocardial blood supply on the RV. In these cases, initial palliation consists of placing a systemic to pulmonary artery shunt. Patients with partial RV dependency for myocardial blood supply are managed variably by different surgeons, but regional LV wall motion abnormalities can worsen after RV decompression.41

The major determinants of the most appropriate surgical approach for a particular patient are: (i) the degree of RV and TV hypoplasia; (ii) presence of RV-dependent coronary circulation; and (iii) the degree of tricuspid regurgitation. The surgical options include: (i) complete biventricular repair with later closure of the interatrial communication; (ii) biventricular repair with allowable mixing of blood at the atrial level (ASD/PFO left open, or surgically adjustable ASD41), but using the RV to pump blood to the lungs; (iii) one and a half ventricular repair using cavopulmonary anastomosis to reduce RV load; (iv) ultimate modified Fontan procedure; and (v) cardiac transplantation (last resort).42,43

Reddy and Hanley40 outline the goals of initial surgical therapy as: (i) minimize mortality; (ii) promote growth of the RV such that chances are improved for a later two ventricular repair; and (iii) minimize the need for non-definitive later surgeries. They point out that: survival after systemic to pulmonary artery shunt is at least as successful as any other initial surgical procedure; the RV will not grow if it is not decompressed and RVOT relief will be needed if two ventricular repair is thought to be possible later; and the ultimate functional potential of the RV is often unclear in the neonate with PA/IVS. The initial procedure often determines the final repair/palliation outcome (Fig. 20.6).41

Perioperative anesthetic management

Given the variety of surgical options available for this particular lesion, general considerations are outlined. For any patient with a small hypertrophic, hypertensive RV, RV filling pressures must be maintained such that the RV cavity does not collapse, causing it to be an ineffective pump. This is especially important after RV outflow obstruction is relieved with the biventricular repair. Inotropic RV support is often essential as RV dysfunction is present after CPB in the presence of increased afterload of an unadjusted pulmonary vascular circulation. Minimizing ventilation pressures and vasodilating the pulmonary vasculature with drugs such as milrinone or dobutamine may reduce RV afterload. With severe pulmonary hypertension after RVOT relief, nitric oxide is useful post-repair to aid in pulmonary vasodilation for the immediate post-repair period until the pulmonary vascular bed adjusts to the increased flow. Also, pulmonary edema causes oxygenation difficulties and bronchospasm after pulmonary flow increases acutely. The one and a half ventricular repair requires a balance of adequate preload to a partially unloaded RV and maintenance of low PVR for upper body passive venous return to the pulmonary vasculature. Along with the ventilation and pharmacological
maneuvers listed above for the biventricular repair, these patients are positioned with 30° head-up to assist upper body venous return to the pulmonary vascular bed. When RV function becomes adequate to support work of breathing, spontaneous respiration in an extubated patient generating negative intrathoracic pressure will boost pulmonary flow. With palliative aortopulmonary shunt placement, consideration is given to continued balance of pulmonary and systemic parallel circulations (described elsewhere in this volume).

Low CO in the postoperative period may occur secondary to unrecognized RV-dependent coronary circulation or from a phenomenon known as a “circular shunt.” The latter occurs in patients who have had a transannular patch and a systemic to pulmonary artery shunt. Because the transannular patch produces free pulmonary regurgitation, blood ejected from the LV flows through the systemic to pulmonary artery shunt and enters the RV in a retrograde fashion. If there is significant TI, blood flows back into the LA and then into the RA through the interatrial communication. This effectively "steals" blood from the systemic circuit and may lead to hypoperfusion of the distal organs and metabolic acidosis. Conservative measures such as raising the PVR and reducing the SVR may be helpful, but quite often surgical intervention to revise the shunt and treat the tricuspid regurgitation is required.

Sedation with benzodiazepines and pain control with narcotic infusions such as morphine are balanced with the patient’s condition and anticipation of duration of mechanical ventilation and time needed for pulmonary vascular adjustment/RV recovery.

**Pulmonary atresia/ventriculoseptal defect/multiple aortopulmonary collateral arteries**

**Anatomy**

In its simplest form, with normal pulmonary vasculature, this lesion can be considered an extreme variation of TOF. However, in most cases, there is great morphologic variability regarding pulmonary artery architecture and sources of PBF, posing major challenges for corrective surgery. With PA, there is no continuity between the RV and the pulmonary trunk. The VSD is usually large and malaligned. The pulmonary arteries may be normal in size or have varying degrees of hypoplasia. The left and right pulmonary arteries may be confluent or non-confluent. An additional major source of PBF is derived from multiple collateral arteries arising from the aorta or its major branches. A given lung segment may be supplied solely from the true pulmonary arteries, solely from the aortopulmonary collaterals, or from both.

**Pathophysiology and natural history**

The great heterogeneity in PBF determines the natural history of this lesion. Excessive PBF through the collateral arteries will produce pulmonary congestion and a clinical picture of CHF with LV volume overload. Moderate stenoses of the collateral arteries may result in a balanced PBF and minimal symptoms. Severe stenoses of the collateral arteries will lead
to inadequate \( PBF \), cyanosis, and hypoxemia. Survival is poorest in those who have either excessive or inadequate \( PBF \). Patients with a balanced blood flow can survive to adulthood with minimal symptoms, but eventually LV failure and aortic insufficiency ensue from chronic left-to-right shunting and volume overload.

Although the diagnosis of PA/VSD/MAPCAs can be made with echocardiography, virtually all patients require cardiac catheterization to delineate the true pulmonary artery architecture and collateral artery anatomy in order to plan the surgical approach.

**Surgical approach**

The ultimate goal of surgery is to achieve a biventricular repair by completely separating the systemic and pulmonary circulations by closing the VSD and restoring continuity between the RV and the pulmonary circuit. The success of this repair is dependent on having an adequate pulmonary vascular bed, which will accommodate the entire RV output. An inadequate pulmonary vascular bed will eventually cause RV failure due to chronic increased afterload. The most important factor for a successful outcome is the post-repair RV pressure, which should be as low as possible. The pulmonary vascular bed is reconstructed by a procedure known as “unifocalization” in which as many of the aortopulmonary collateral arteries as possible are detached from the aorta and anastomosed to the central pulmonary arterial tree, in order to provide unobstructed blood flow from the RV to the pulmonary microcirculation. This centralization of multiple sources of \( PBF \) was traditionally done in two or three stages via bilateral thoracotomies, followed by a definitive repair through a median sternotomy.\(^{44}\) However, some centers are obtaining good results with a single-stage unifocalization and repair, and this approach does have some advantages.\(^{20,45}\)

It avoids subjecting the patient to multiple surgeries, which if performed via thoracotomies, can make subsequent procedures extremely hazardous (especially lung transplants) due to increased adhesions and the potential for massive bleeding. Additionally, serious neurological injury can occur during CPB-assisted unifocalization because increased runoff into the pulmonary circuit can result in cerebral hypoperfusion despite adequate pump flows. Obviously, a single-stage approach will not be applicable in all patients, and this group will need a systemic-to-pulmonary artery shunt or a conduit from the RV to the pulmonary artery to allow growth before definitive repair. Reddy et al.\(^{20}\) in their series of 85 patients, were able to complete one-stage unifocalization and intracardiac repair in 56 patients. In 23 patients single-stage unifocalization was done but the VSD was left open, and six patients required staged unifocalization through sequential thoracotomies. In the subgroup of patients with excessive \( PBF \), the collateral arteries may have to be narrowed at their origin to limit overcirculation (conceptually similar to pulmonary artery banding).

**Perioperative anesthetic management**

The anesthetic management will vary according to whether a staged approach to unifocalization via a thoracotomy or one-stage unifocalization with intracardiac repair is being contemplated. The general principles for induction, maintenance, and monitoring are similar to those described above for TOF repair. There are several major anesthetic challenges for unifocalization via a thoracotomy.\(^{46}\) These include difficulties with oxygenation and ventilation from one-lung anesthesia, hemodynamic instability, and metabolic acidosis. In the older child, lung separation with either a double-lumen tube or a bronchial blocker will greatly facilitate surgical exposure and minimize lung contusion from surgical retraction. Extensive intrapulmonary and major airway bleeding from multiple vascular Anastomoses will also compromise ventilation. In addition, major blood loss should be anticipated, and large-bore intravenous access is essential. Warming of fluids before transfusion will reduce the chance of hypothermia. Finally, thoracotomies are extremely painful, and a thoracic epidural catheter will provide excellent postoperative analgesia. The benefits of epidural anesthesia are balanced against the potential risk of neurological damage from catheter placement in an anesthetized child.

One-stage unifocalization (with or without definitive repair) is carried out via a median sternotomy or bilateral trans-sternal thoracotomy (“clamshell” incision). As many MAPCAs as possible are ligated, mobilized, and unifocalized without CPB. As each MAPCA is ligated, the arterial saturation will decrease because a proportion of the \( PBF \) is being cut off. At the point at which the patient nears compromise from arterial desaturation, CPB (with moderate hypothermia and a beating heart) is initiated. The rest of the unifocalization is then completed. As mentioned above, it is vital to control as many of the MAPCAs as possible prior to initiating CPB to prevent cerebral injury due to increased runoff into the pulmonary circulation. A valved conduit is then placed between the RV and the central pulmonary artery to restore continuity. Next, the surgeon determines whether the VSD should be closed at the time of one-stage unifocalization. This is a critical step because if the VSD is closed, and the “new” pulmonary vascular bed is inadequate to receive all of the \( CO \), RV failure will rapidly ensue. One approach\(^{19,47}\) is to do an intraoperative pulmonary flow study to estimate the resistance of the new vascular bed. The lungs are perfused with the equivalent of one \( CO \), and if the mean pulmonary artery pressure is less than 30 mmHg, the VSD is closed.

Three major problems can be anticipated post-CPB:

1. RV dysfunction, which is usually secondary to increased afterload due to an inadequate pulmonary vascular bed.
The mainstays of treatment are inotropic support with dopamine, dobutamine, or milrinone; increased preload; and ventilatory maneuvers to lower PVR. Nitric oxide may also be helpful as a pulmonary vasodilator.

2 Intrapulmonary bleeding due to multiple vascular suture lines, systemic anticoagulation, and the effects of CPB.

3 Lung reperfusion injury due to increased blood flow to many previously underperfused lung segments. This injury manifests as pulmonary edema, bronchospasm and difficulties with ventilation and oxygenation. Frequent endotracheal suctioning, fiberoptic bronchoscopy, and PEEP will be helpful in managing this problem. In general, these patients are not good candidates for early extubation. These patients benefit from sedation with benzodiazepines and aggressive pain control to keep pulmonary and systemic blood pressures from becoming excessive as multiple arterial anastomoses and suture lines may predispose to postsurgical bleeding.

**Summary**

Right-sided obstructive congenital heart lesions manifest a variable presentation. Depending upon the severity of the structural anomaly(-ies), such lesions may present a spectrum of illness in the congenital heart patient, ranging from the ductal-dependent, cyanotic neonate in CHF to the minimally affected young adult who manifests mild to moderate exercise intolerance. All are characterized by the nearly universal presence of a septal defect that has the potential for right-to-left flow. An understanding of the physiology of the defect and effects of the proposed surgical intervention are essential for the meticulous and well-planned perioperative anesthetic management of such patients.

**Key points**

**Ebstein’s anomaly**

1 Patients with extreme cardiomegaly or perioperative ventricular arrhythmias should receive prophylactic antiarrhythmic treatment, such as amiodarone.

2 Patients with poorly functioning RVs may be dependent upon high filling pressures to maintain adequate CO.

**Tetralogy of Fallot**

1 Hypercyanotic spells are treated with oxygen, phenylephrine, intravenous fluid and esmolol.

2 During BT shunt placement difficulties may be encountered with oxygenation and ventilation, and hypotension after the shunt is open.

3 Postoperative ventilation is recommended for 12–24 hours after BT shunt because of the risk of pulmonary edema.

4 Following complete repair, RV dysfunction, heart block, arrhythmias, and bleeding should be anticipated.

**Pulmonary stenosis or atresia with intact ventricular septum, and pulmonary atresia with ventricular septal defects and major aortopulmonary collaterals**

1 Inotropes are used cautiously with patients with PS/IVS in which there is a component of dynamic infundibular stenosis, so as to not worsen outflow obstruction.

2 Small hypertrophic RVs require meticulous attention to preload to maintain function.

3 An RV-dependent coronary circulation requires maintenance of RV intracavitary pressure to prevent myocardial ischemia.

4 After repair of PA/IVS and PA/VSD, severe RV dysfunction is often present due to the high afterload of an insufficient pulmonary vasculature: inotropes, ventilation adjustments, and vasodilators are helpful.

5 Enhanced post-repair lung perfusion may result in pulmonary edema, intrapulmonary bleeding, bronchospasm, and difficulties with oxygenation and ventilation.

6 Antifibrinolytic agents may be useful for minimizing bleeding after unifocalization of MAPCAs.

**References**


PART 5 Anesthesia for specific lesions


