Chapter 47

VACTERL Syndrome

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Case Scenario

A 1-day-old neonate weighing 2.5 kg, born at 36 weeks estimated gestational age, is scheduled for esophageal atresia/tracheoesophageal fistula repair after initial feeding attempts led to choking, respiratory distress, and cyanosis. A Replogle tube could not be advanced and a chest radiograph showed the tube curled in the upper esophagus, with the presence of gas in the stomach.

An echocardiogram was obtained and showed the following:

- *Tetralogy of Fallot with severe infundibular stenosis*
- *Hypoplastic pulmonary arteries*
- *Right aortic arch*
- *Patent ductus arteriosus*

The pulse oximeter is consistently reading 85% with the patient breathing room air.

Key Objectives

- Describe the common anatomic forms of esophageal atresia/tracheoesophageal fistula.
- Understand the anesthetic implications of VACTERL association.
- Describe airway and ventilatory management strategies during surgical repair of a tracheoesophageal fistula.
- Understand the intraoperative management of ventilatory problems during tracheoesophageal fistula repair.
- Understand the perioperative management of patients with unrepaired tetralogy of Fallot undergoing noncardiac surgery.

Pathophysiology

What is the "VACTERL association"?

An "association" is a group of pathogenetically unrelated malformations occurring together more often than expected by chance without evidence of a single unifying cause. The original acronym **V**ertebral abnormalities, **A**nal atresia, **T**racheoesophageal fistula with **E**sophageal atresia, **R**adial and Renal dysplasia was described in 1973 and later updated to VACTERL with the inclusion of **C**ardiac anomalies and **L**imb defects rather than radial anomalies. VACTERL association occurs in approximately 1 in 10,000 to 1 in 40,000 live births with the diagnosis confirmed by the presence of at least three of the aforementioned abnormalities [1]. Patients with VACTERL association frequently require surgery for anal atresia or repair of esophageal atresia/tracheoesophageal fistula (EA/TEF) in the first days of life.

While the cardiac anomalies most commonly associated with VACTERL association are ventricular septal defects (VSD) and tetralogy of Fallot (TOF), a variety of simple and complex cardiac defects are possible, and therefore multidisciplinary perioperative planning in conjunction with cardiac surgery and pediatric cardiology is necessary. Patients with a ductal-dependent cardiac lesion are the most critically ill. In this patient group severe obstruction to either systemic or pulmonary blood flow (PBF) exists, requiring maintenance of a patent ductus arteriosus (PDA) with prostaglandin E_1 (PGE_1) infusion and balancing of the pulmonary and systemic blood flows for survival.

Clinical Pearl

Many of these neonates will require surgery in the first few days of life and will present significant anesthetic challenges, mostly due to cardiac anomalies, which occur in 40%–80% of patients with VACTERL syndrome.

What is esophageal atresia and what is the most common subtype?

Esophageal atresia (EA) is a congenital malformation of esophageal continuity, associated in over 90% of cases with a fistulous communication of the esophagus with the

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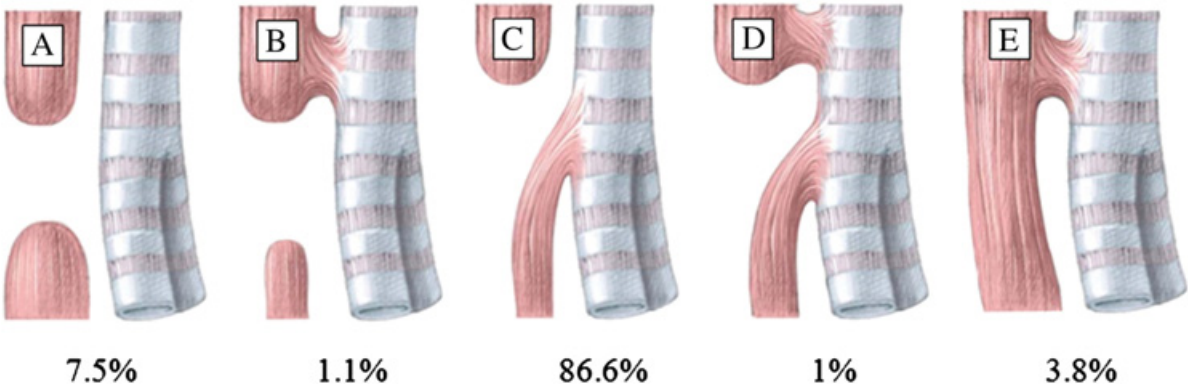

Figure 47.1 Gross classification and frequency of anomalies of the esophagus and trachea. (A) Isolated esophageal atresia without fistula. (B) Esophageal atresia with proximal tracheoesophageal fistula. (C) Esophageal atresia with distal tracheoesophageal fistula. (D) Esophageal atresia with proximal and distal tracheoesophageal fistula, "K-type." (E) Tracheoesophageal fistula without esophageal atresia, "H-type." From Parolini F., et al. Esophageal atresia with proximal tracheoesophageal fistula: A missed diagnosis. *J Pediatr Surg* 2013; **48**: E13–18. With permission.

trachea or a main bronchus. It occurs in approximately 1 in 2500 to 3500 livebirths [2]. Although two main classification systems (Gross and Voght) show the different subtypes, it is preferable to use descriptive language to avoid confusion. The most commonly occurring tracheoesophageal fistula (TEF) is Gross Type C (over 85%), consisting of a blind upper esophageal pouch and a distal tracheoesophageal fistula. (See Figure 47.1.) This results in pulmonary aspiration of upper pouch contents, with ventilation of the stomach via the fistula producing abdominal distension, increased ventilatory difficulty, and aspiration of gastric contents into the trachea via the fistula, producing aspiration pneumonitis. Embryologically, EA/TEF results from incomplete separation of the cranial part of the foregut into respiratory and esophageal parts during the fourth week of gestation. Most cases arise sporadically, but there is an association with certain major chromosomal abnormalities – specifically, trisomy 21, 18, and 13. Trisomy 18 has a very poor prognosis, and it may be necessary to rule out this chromosomal anomaly by urgent karyotyping before surgical planning. VACTERL association occurs in about 25% of EA/TEF infants.

Clinical Pearl

Approximately 50% of patients with EA/TEF have additional congenital abnormalities. Congenital heart disease is common and can have a significant impact on survival.

What is tetralogy of Fallot?

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease (CHD). The key morphologic abnormality producing the features of the tetrad is anterior and cephalad deviation and malalignment of the infundibular (or outflow) septum of the right ventricle (RV) resulting in the following:

- Right ventricular outflow tract (RVOT) obstruction
- Large, unrestrictive VSD
- Overriding of the aorta
- Right ventricular hypertrophy (RVH)

Right ventricular outflow tract obstruction may be subvalvular secondary to hypertrophied muscle bundles, valvular, or supravalvular due to variable degrees of hypoplasia of the pulmonary annulus and main/branch pulmonary arteries.

Echocardiography is the mainstay of diagnosis. The large VSD and the malaligned infundibular septum are easily demonstrated in the parasternal long axis view. The parasternal short axis and subcostal views are used to define the degree of RVOT obstruction. Additional findings may include an atrial septal defect (ASD), right aortic arch (RAA), left-sided superior vena cava (SVC), and coronary abnormalities. Cardiac catheterization is rarely required for diagnostic purposes and may be risky, as it may provoke a "tet spell" due to catheter manipulation in the RV. However, it is increasingly being used in selected patients for palliative procedures such as stenting of the PDA or the RVOT, as an alternative to surgical aortopulmonary shunt placement. (See Chapter 7 and Figure 7.1.)

What is an acceptable systemic saturation for a neonate with unrepaired TOF?

The systemic hemoglobin–oxygen saturation of a patient with unrepaired TOF can be quite variable, depending on

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the presence and size of the PDA and the degree of existing RVOT obstruction.

- "***Pink tets***" have normal systemic oxygen saturation due to their mild RVOT obstruction; they predominantly shunt from left-to-right (L-to-R) across the VSD, producing a picture similar to that of congestive heart failure.
- ***If the PDA has closed,*** systemic saturations will decrease as the RVOT obstruction worsens, and shunting across the VSD then becomes bidirectional. Systemic oxygen saturations in the mid- to high 80s are acceptable as long as they remain stable until the patient undergoes palliation or definitive repair. At the most severe end of the spectrum, shunting across the VSD is mostly right-to-left (R-to-L), producing severe cyanosis.
- ***Patients with a PDA*** should have adequate PBF and systemic oxygen saturations in the 90s even with RVOT obstruction, as the PDA also provides PBF.

Clinical Pearl

The systemic hemoglobin–oxygen saturation of a patient with unrepaired TOF can be quite variable, depending on the presence and size of the PDA and the degree of existing RVOT obstruction.

What is a "tet" spell? How should one be managed?

Infants can experience hypercyanotic episodes ("tet spells") resulting from infundibular spasm; these spells can occur even in "pink tets." These spells can be provoked by painful stimuli, feeding, or bowel movements, and unless treated promptly can lead to cardiovascular collapse due to severe hypoxemia and lack of PBF.

Treatment of a tet spell includes:

- ***Airway***: Administration of 100% oxygen and tracheal intubation if necessary
- ***Fluid***: Crystalloid or colloid boluses (15–20 mL/kg) to increase preload and promote antegrade PBF
- ***Increase SVR (pharmacologic)***: Phenylephrine bolus(es) titrated to increase SVR and reduce R-to-L shunting across the VSD. Patients with severe RVOT obstruction and recurrent tet spells can require institution of a phenylephrine infusion.
- ***Increase SVR (mechanical)***: Mechanical measures include bilateral femoral artery compression or placing the infant into a knee-to-chest position.
- ***Sedation/anesthesia***: Intravenous (IV) sedation with morphine (0.05–0.1 mg/kg). If in the operating room, or intubated, fentanyl (2–4 mcg/kg) can be used to reduce sympathetic overstimulation or increase anesthetic depth.
- ***Reduce infundibular spasm***: Esmolol bolus(es) of 50–100 mcg/kg can be administered to reduce the heart rate and infundibular spasm via its negative inotropic effect. Infants who are prone to tet spells are frequently maintained on oral propranolol until surgical repair.

The frequent or continuing occurrence of tet spells resulting in severe hypoxemia and cardiovascular collapse is the main concern for unrepaired TOF patients. In this patient, if adequate PBF is maintained via a PDA maintained by PGE_1, then EA/TEF repair can be performed. After repair, PGE_1 can be stopped to assess whether PBF continues to be adequate without the PDA. If not, due to this patient's size and prematurity, he is most likely not currently a candidate for a complete TOF repair and therefore will require establishment of a stable source of PBF to allow time to grow and await complete TOF repair at 4–6 months of age. This initial palliative procedure can occur either in the cardiac catheterization laboratory in the form of a PDA or RVOT stent, or in the cardiac operating room via creation of a modified Blalock–Taussig shunt (mBT shunt). (See Figure 47.2.)

Clinical Pearl

The mainstays of treatment for a perioperative tet spell are phenylephrine and fluid boluses to increase PBF and reduce R-to-L shunting.

After EA/TEF repair, what is the impact of CHD on patient outcomes?

The two strongest predictors of morbidity and mortality in this patient population are low birth weight (<1500 grams) and the presence of major congenital anomalies. Spitz's prognostic classification from 1994 identified three groups: ***Group 1*** – birth weight >1500 grams and no major cardiac anomalies (98% survival); ***Group 2*** – birth weight <1500 grams OR major cardiac anomalies (59% survival); ***Group 3*** – weight <1500 grams AND major cardiac anomalies (22% survival). Although contemporary outcomes are improved, the overall trend is similar. Perioperatively, patients with CHD have significantly more complications, specifically, difficulties with ventilation and oxygenation, need for inotropic therapy, longer duration of mechanical ventilation, and longer intensive care unit (ICU) and hospital stay [3]. Patients with ductal-dependent cardiac lesions are an even higher risk group compared to non-ductal-dependent lesions [4].

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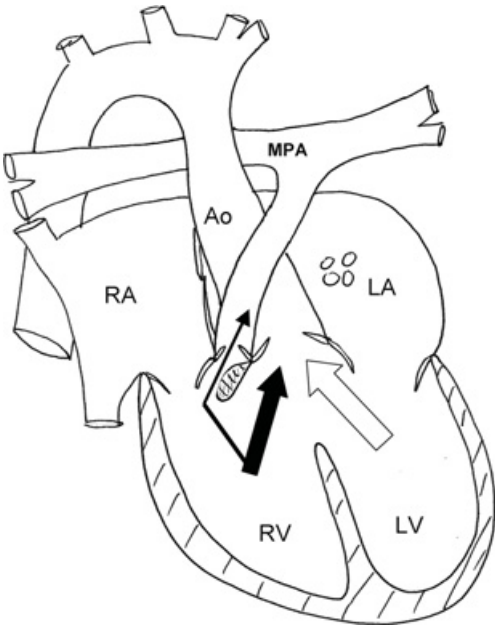

Figure 47.2 Tetralogy of Fallot. Malalignment of conal septum results in overriding aorta, subvalvular pulmonary stenosis, and VSD. With narrowing of RV outflow, desaturated blood flow will be divided (black arrows), a portion to the lungs and a portion across the VSD to the aorta, depending on the relative resistance of each pathway. Systemic saturation will be determined by the amount of desaturated blood mixing with fully saturated blood (white arrow). A right aortic arch is shown. Ao, aorta; LA, left atrium; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RV, right ventricle. From Sommer R. J. et al. Pathophysiology of congenital heart disease in the adult. Part III: Complex congenital heart disease. *Circulation* 2008; **117**: 1340–50. With permission.

What is the significance of a RAA for surgical management in this patient?

The incidence of RAA is much higher in patients with CHD. It is commonly associated with TOF (approximately 25% of patients), vascular rings, prematurity, and lower birth weight. Because the RAA courses above the right mainstem bronchus, it can complicate surgical exposure of the TEF, making the repair more difficult. In these cases, the surgeon may prefer a left thoracotomy.

Anesthetic Implications

What are the most important aspects of preoperative workup in this patient?

- ***Confirm the diagnosis of EA/TEF***: These patients are frequently premature and have low birth weights. The diagnosis may be suspected prenatally by the presence of polyhydramnios and a small or absent stomach bubble, resulting from the inability of the fetus to swallow amniotic fluid, but this is not a specific finding. The first feed will result in choking, coughing and possibly cyanosis due to pulmonary aspiration. Attempts to pass a 10 Fr orogastric tube beyond about 10 cm will fail. A whole-body radiograph ("babygram") will reveal the tube curled up in the upper esophageal pouch, and the concomitant appearance of gas in the stomach will confirm the presence of a distal TEF. A "gasless" abdomen usually indicates EA without a fistula. The Replogle tube should be connected to low continuous suction, and the neonate kept nil per os after diagnosis.
- ***Echocardiogram***: Every patient with VACTERL association or an EA/TEF needs an echocardiogram to rule out CHD and determine aortic arch sidedness, as both will influence intraoperative management. Repair of EA/TEF is usually done via a right thoracotomy, therefore the presence of a RAA significantly hinders surgical exposure, making the repair more complicated and riskier. In these cases, a left thoracotomy is usually preferred. A RAA, which passes over the right main bronchus, is present in approximately 4% of the normal population, but is much more common in patients with CHD. The presence of a ductal-dependent cardiac lesion (indicating severe systemic or pulmonary outflow obstruction) requires initiation of a PGE_1 infusion to maintain ductal patency until a more definitive cardiac palliation or repair is performed; this will significantly complicate intraoperative management and postoperative recovery for the patient.

Clinical Pearl

Every EA/TEF patient must have a preoperative echocardiogram both to determine aortic arch sidedness for the surgical approach and to define any existent CHD to aid in intraoperative management and prognosis.

What are the airway and ventilation management considerations in this patient?

Until the TEF is surgically controlled, management of intubation and ventilation present the biggest challenges for the anesthesiologist. ***The main concerns are loss of tidal volume through the fistula leading to gastric distension and further difficulties in ventilation and oxygenation***. This is more likely to occur if the fistula is large and there is poor lung compliance due to preexisting lung disease secondary to prematurity or aspiration pneumonia. Massive gastric distension and rupture has been reported. Immediate needle

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decompression of the abdomen may be needed in this situation, followed by either immediate laparotomy and temporary ligation of the distal esophagus or rapid thoracotomy and ligation of the fistula.

Clinical Pearl

There is no consensus on the optimal anesthetic management of the airway. The choice depends on the experience and preference of the anesthesiologist, patient comorbidities, and institutional surgical practices.

What options exist for anesthetic induction in this patient? What intubation technique is preferable?

Awake intubation is traumatic and stressful to the neonate and does not provide optimal intubating conditions. In premature babies, it may also increase the risk of intraventricular hemorrhage due to the hypertensive surge associated with laryngoscopy. It is rarely practiced and cannot be routinely recommended.

Spontaneous ventilation, using a combination of sevoflurane and intravenous drugs (propofol, ketamine, or remifentanil infusion) is widely practiced and has many advantages. However, as propofol may reduce SVR and promote R-to-L shunting, it should either be avoided in this patient or utilized in reduced doses. Due to the negative intrathoracic pressure generated during spontaneous ventilation, inspired gas tends to favor the lungs rather than passing through the fistula into the stomach. As anesthesia is deepened gentle manual assistance with ventilation is usually needed, especially with low birth weight premature neonates. It is widely advocated to maintain spontaneous ventilation until the fistula is ligated, but in practice this is difficult to do with the chest open. Intravenous induction and the use of neuromuscular blockade allow rapid optimal intubating conditions even though gentle positive pressure ventilation (PPV) is needed. It is the preferred technique in smaller, fragile infants with coexistent cardiac anomalies who may not tolerate deep inhalational anesthesia.

Is preoperative bronchoscopy necessary before surgery?

Routine pre-repair bronchoscopy is performed in only approximately 50%–60% of cases [5]. However, there are several strong arguments in favor of bronchoscopy [6, 7]. It can be done in less than 5 minutes, and in experienced hands, has minimal complications. It allows assessment of the position, size, and number of fistulas, which can be very helpful in correctly positioning the tracheal tube. Approximately 30% of TEFs are within 1 cm of the carina, so precise positioning of the tracheal tube is crucial. Intraoperative difficulties with ventilation or oxygenation can be managed more confidently if one knows the anatomy of the tracheobronchial tree. If the fistula is large or carinal (which can be diagnosed only with a bronchoscope), it can be helpful to occlude it with a Fogarty balloon catheter to improve intraoperative ventilation. A large fistula is also more likely to be inadvertently intubated during the procedure. This is especially important in premature babies with lung disease resulting in poor compliance that will preferentially result in inspired gas flowing into the fistula. Bronchoscopy can identify an upper pouch fistula and may indicate a long-segment atresia if the fistula is located higher in the trachea, which has potential surgical implications. (See Figure 47.3.)

Clinical Pearl

Preoperative bronchoscopy can be extremely valuable in facilitating correct endotracheal tube positioning, diagnosing additional airway abnormalities, and when possible, occluding the fistula with a Fogarty balloon catheter.

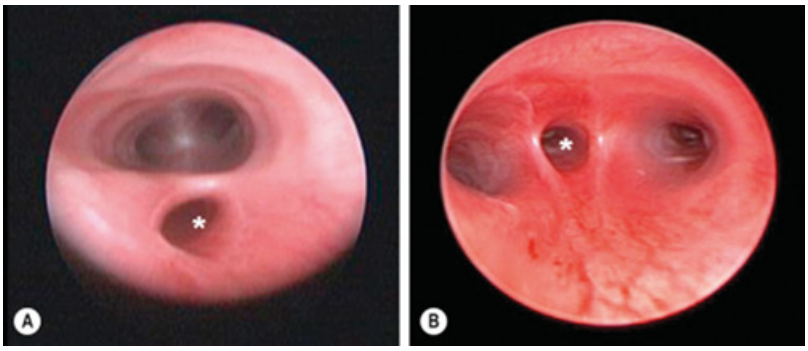

Figure 47.3 Bronchoscopic views showing distal TEF entering the trachea at different levels. (A) The fistula (asterisk) enters in the mid-trachea. (B) The fistula (asterisk) enters at the carina. These images emphasize the importance of preoperative bronchoscopy during airway management and repair. From Rothenberg S. Esophageal atresia and tracheoesophageal fistula malformations. In Holcomb G. W., Murphy J. P., and St. Peter S. D., eds. *Holcomb and Ashcraft's Pediatric Surgery*, 7th ed. Elsevier; 2020: 437–58. With permission.

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How should the endotracheal tube be positioned?

The trachea should be intubated with a cuffed endotracheal tube (ETT) positioned below the fistula to seal the airway and minimize ventilation of the stomach. Presurgical bronchoscopy is extremely helpful in aiding accurate ETT placement and predicting ventilation difficulties during the procedure. Traditional teaching is to deliberately intubate the right main stem bronchus, and then, while auscultating over the left chest, slowly withdraw the tube until breath sounds are heard on the left side, indicating the tube position is just above the carina. Rotating the tube to place the bevel anteriorly may also help to occlude the fistula, which lies posteriorly. This will be satisfactory for most cases. However, in approximately 10% of patients, the fistula is located at the carina or the main stem bronchus. Positioning the tube just above the carina may cause gastric distension and ventilatory compromise. In these cases, deliberate endobronchial intubation may be needed to allow adequate ventilation. In cases with a large or a carinal fistula, a balloon catheter may also be employed via a rigid bronchoscope to occlude the fistula. This will improve ventilation and may prevent inadvertent intubation of the fistula during surgical manipulation.

After administration of muscle relaxant, the stomach is increasingly distended with mask ventilation, chest rise is diminished, and oxygen desaturation is occurring. What should be done next?

This scenario suggests loss of pulmonary ventilation through the fistula into the stomach and can rapidly lead to cardiorespiratory compromise. The trachea should be intubated immediately. If the abdomen becomes massively distended, there is a risk of gastric rupture and pneumoperitoneum. Needle decompression or opening the abdomen and placing a temporary ligature around the gastroesophageal junction may be required. If the clinical situation is stable, an immediate thoracotomy and ligation of the fistula may be required to stabilize the patient before proceeding with complete repair.

Is invasive monitoring necessary? What sites could be used for arterial and central access?

Umbilical arterial and venous access can be extremely useful but are not always possible. An arterial line should be placed in all cases, if possible, as there is significant lung retraction and manipulation of the great vessels. In patients with coexisting cardiac anomalies, it is essential for hemodynamic monitoring. The right arm should be avoided for IV and arterial access because it is positioned above the head and may be attached to a bar above the patient's head. Patients with VACTERL association can also have radial abnormalities which may preclude arterial line placement in the arms. In critically ill patients, or those with complex cardiac malformations, central venous access (internal jugular or femoral) is recommended in the event that administration of inotropes becomes necessary. Many neonates may have peripherally inserted central catheters placed prior to surgery.

Clinical Pearl

An arterial line should be placed in all cases, if possible, as there is significant lung retraction and manipulation of the great vessels. In patients with coexisting cardiac anomalies, it is essential for hemodynamic monitoring.

What are the pros and cons of thoracoscopic repair of TEF?

Thoracoscopic TEF repair requires significant technical skill and experience. Contraindications include prematurity, low birth weight, major cardiac defects, and significant abdominal distension. Advantages are avoidance of a thoracotomy with its associated risks of muscle and nerve damage to the chest wall, less pain, better visualization due to magnification, and less retraction on the lung and other mediastinal structures. Disadvantages are mostly due to carbon dioxide (CO_2) insufflation, which can result in significant acidosis that may not be tolerated in patients with delicately balanced pulmonary and systemic blood flows.

What are the key surgical steps in an open surgical repair of EA/TEF?

The patient is positioned for a right posterolateral thoracotomy in the fourth to fifth intercostal space with the right arm positioned above the head. A Replogle tube should be placed in the upper esophageal pouch and will be manipulated by the anesthesiologist to allow recognition of the upper pouch. Extrapleural dissection is carried out to the posterior mediastinum to reveal the azygous vein, which usually overlies the fistula. The azygous vein is usually (but not always) ligated and divided. A very important caveat to this is that the surgeon must be certain that there is no interrupted inferior vena cava (IVC) and azygous continuation that carries the venous drainage from the lower body to the SVC. In this situation, test clamping of the azygous will significantly and rapidly reduce cardiac output. An interrupted IVC with azygous continuation is a common finding in the heterotaxy syndromes and should

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be ruled out with preoperative echocardiography. The TEF is then identified, ligated, and divided. This usually improves ventilation significantly, and the subsequent surgery is then relatively uneventful. The two esophageal ends are then anastomosed to restore continuity over a feeding tube. Before chest closure, the chest cavity is irrigated with warm saline as positive pressure is applied to the tracheal tube to rule out an air leak.

During dissection, pulse oximetry rapidly decreases from 90% to 50%. What are the possible causes and management?

Problems with ventilation and oxygenation are common during TEF repair and require constant vigilance and close communication with the surgeon. It is important to emphasize that even with initially perfect ETT placement, patient positioning and surgical manipulation can frequently cause ventilatory difficulties. It is helpful to have a flexible fiberoptic bronchoscope immediately available in the operating room. Moderate hypoxemia (SpO_2 80%–90%) usually results from lung retraction and may necessitate increasing inspired oxygen concentration and intermittent reexpansion of the lung to maintain oxygenation. Traction on the small pliant trachea can easily obstruct ventilation. Secretions, blood, or mucus can obstruct small tracheal tubes, and frequent suctioning may be necessary. A balloon catheter placed in the fistula may be displaced into the trachea and cause total obstruction of the airway. Deflation of the balloon and gentle withdrawal of the catheter may solve the problem. Sudden loss of the capnogram with severe desaturation may result from intubation of the fistula. If withdrawing the tube slightly does not restore oxygenation, the airway should be rapidly evaluated by passing the fiberscope through the ETT if time allows. Otherwise, the patient will need emergent reintubation.

In patients with ductal-dependent PBF undergoing a left thoracotomy due to a RAA, surgical retraction on the ductus or pulmonary arteries will cause hypoxemia due to diminished PBF. This may present as decreasing expired CO_2 on the capnogram. A tet spell is less likely to manifest in patients with a PDA, but acute infundibular spasm accompanied by hypotension and tachycardia may be enough to increase R-to-L shunting across the VSD causing profound desaturation. The surgeon should be asked to remove any retraction if possible. Manual ventilation should be used to reexpand the upper lung. If these measures do not restore appropriate oxygenation, phenylephrine and a fluid bolus should be administered to raise the SVR and reduce the shunting across the VSD. It is essential to check that the PGE_1 infusion has not been interrupted, as this may cause ductal closure.

Clinical Pearl

Mild intraoperative desaturation is common, but a complete loss of ventilation and oxygenation can be due to blockage of the ETT by blood or secretions, intubation of the fistula, or displacement of the Fogarty balloon into the trachea.

When should this patient be extubated?

Patients who undergo uncomplicated EA/TEF repairs can be extubated early, even in the operating room, to minimize trauma to the trachea from the ETT and suctioning. In unstable patients, and those with significant tension on the esophageal anastomosis, it may be beneficial to keep the patient intubated, sedated, and paralyzed for several days with the head in a slightly flexed position.

What modalities can be used for postoperative analgesia?

Opioids are the mainstay of analgesia. Caudal and thoracic epidural catheters can be placed if there are no vertebral or spinal cord anomalies. Intercostal nerve blocks and intrapleural catheters can also be used.

What are some common complications of EA/TEF repair?

Early complications include tracheomalacia, which may be severe enough to require reintubation or surgical aortopexy; anastomotic leaks, most of which can be managed conservatively; esophageal stricture requiring repeated dilations; and recurrence of the fistula. The major late complication is gastroesophageal reflux which may require surgical management if medical management fails.

Ten days later, the patient is extubated and progressing well. A trial of discontinuing PGE_1 resulted in severe hypoxemia. Based on size, the patient is not considered a candidate for complete TOF repair yet. What other management options exist?

Neonatal TOF repair, although feasible, is associated with higher morbidity and mortality [8]. In addition, several factors may mitigate against neonatal repair, including small patient size, gestational age, comorbidities, coronary anatomy, and inadequate branch pulmonary artery size and arborization. Repair is generally deferred until 3–6 months of age.

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For ductal-dependent patients, there are several options. Recently, stenting the PDA or the RVOT in the cardiac catheterization laboratory has become a viable alternative with outcomes either equivalent or superior to surgical shunt placement. It is a less invasive procedure with shorter ICU and hospital length of stays. Surgical creation of a systemic-to-pulmonary shunt such as a mBTS can also be considered [9].

What anesthetic issues should be considered for ductal stent placement?

Although relatively simple in principle, this procedure requires significant experience and planning for a successful outcome [10]. The key points are outlined here.

Location: The procedure should be performed in a cardiac catheterization laboratory with general endotracheal anesthesia.

Pharmacologic: Inotropic infusions (dopamine or epinephrine, and phenylephrine) should be available. A heparin infusion will also be needed after stent placement to prevent in-stent thrombosis.

Monitoring: Invasive monitoring is usually not necessary.

Surgical team back-up including extracorporeal membrane oxygenation (ECMO) is essential in case of irreversible ductal spasm or ductal tearing.

Procedural considerations:

- Knowledge of ductal morphology is essential, including its origin (head and neck vessels, underside of aorta, descending aorta), length, and tortuosity.
- PGE_1 may be stopped several hours before the procedure to allow the PDA to shrink and allow a better assessment of the ductal size to choose the appropriately sized stent. It is important to leave the PGE_1 infusion in line so that it may be restarted immediately in case of ductal spasm during the procedure.
- Vascular access is chosen based on the site yielding the straightest route to the ductus. This may be the carotid or axillary artery.
- The most critical period occurs when the wire is being manipulated in the ductus, as this can cause sudden ductal spasm and rapid hemodynamic collapse, requiring rapid management with stent placement, resuscitation with inotropes and vasopressors, and reinstitution of PGE_1. ECMO rescue may be required if these measures fail.

Post-procedure: Heparin infusion and aspirin are usually started to prevent in-stent thrombosis and reduce thrombotic complication in the access vessel.

Complications: Vascular injury, stent thrombosis, and the need for reintervention for stent dilation or additional stent placement.

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Suggested Reading

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Puri K., Morris S. A., Mery C. M., et al. Characteristics and outcomes of children with ductal-dependent congenital heart disease and esophageal atresia/tracheoesophageal fistula: a multi-institutional analysis. *Surgery* 2018; **163**: 847–53.

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