Anesthetic Management of Tracheoesophageal Fistula in a Premature Newborn with VACTERL Association and Hypoplastic Left Heart Syndrome

Moderators:

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Goals:

1. Understand the anatomy and anesthetic implications of esophageal atresia (EA)/tracheoesophageal fistula (TEF)
2. Discuss the anesthetic implications and increased morbidity and mortality of neonates with unrepaired hypoplastic left heart syndrome (HLHS) undergoing noncardiac surgery
3. Review VACTERL association and other syndromes associated with EA/TEF and with HLHS
4. Discuss the combined anesthetic implications of HLHS, TEF, and prematurity during a thoracotomy procedure

Case history:

You are called by the general surgery team for input on a 2-day old, 34-week premature infant with a probable diagnosis of VACTERL association, a prenatal diagnosis of hypoplastic left heart syndrome (HLHS), and newly diagnosed esophageal atresia/tracheoesophageal fistula (EA/TEF). The surgeons want to know what the anesthetic risks are so that they can advise the family regarding possible interventions.

Questions:

*What are the different types of esophageal atresia (EA) and TEF? What other associated anomalies should be evaluated in a newborn with EA/TEF? What is VACTERL association, and how is it different than VATER, CHARGE, or Velocardiofacial Syndrome? What are the options for surgical timing in isolated EA/TEF?*

Case history and physical exam (cont.):

The patient was born with an uncomplicated vaginal delivery, and with the prenatal diagnosis of HLHS, he was immediately transported to the NICU and started on prostaglandin E₁ (PGE₁) at 0.01 mcg/kg/min. Failure to pass an NG tube led to the diagnosis of EA. Several hours after birth, he had an increase in the work of breathing, and the NICU began high-flow nasal cannula at FiO₂ 0.21 with good effect. On day of life 2, the neonate demonstrated worsening respiratory distress, and he was placed in a sub-ambient hood of FiO₂ = 0.17 with good effect over several hours.

- Extremity survey: shortened left radius, absent right radius
• Renal U/S: mild echogenicity of bilateral kidneys consistent with prematurity
• Cranial U/S: consistent with prematurity
• Abdominal U/S: normal abdominal situs
• Genetic studies: pending
• Echo:
  o Hypoplastic left heart syndrome
  o Indistinguishable left ventricular cavity
  o Aortic and mitral valve atresia
  o Mild to moderate tricuspid valve insufficiency
  o Severely hypoplastic ascending aorta, 0.12 cm.
  o Patent ductus arteriosus, bidirectional shunt, large and unrestrictive
  o Continuous flow noted through the branch pulmonary arteries
  o Unrestrictive atrial septal defect

Questions:

What is the anatomy and physiology of HLHS? What will influence pulmonary versus systemic blood flow in this neonate? How would aspiration due to the TEF influence the cardiac physiology? What additional pulmonary challenges might be encountered simply due to prematurity?

What are the side effects of PGE? If the patient is on PGE and sub-ambient FiO₂, may TEF repair proceed? How does the degree of tricuspid regurgitation and size of the ASD impact the cardiac physiology?

What input do you have to the family and surgical team at this time?

Preoperative planning:

In your discussions with the surgeons, intensivists, and cardiologists, the decision is made to perform repair of the TEF first thing in the morning, prior to the patient undergoing a Norwood repair in a week.

Questions:

What vascular access would you want for this anesthetic? If umbilical venous and/or arterial catheters were present, would you place additional central venous and/or arterial catheters? Does the upcoming Norwood repair influence your decision?

If the ASD was restrictive, would you advocate balloon atrial septostomy prior to the EA repair? Should it be done preoperatively in the NICU or under the same anesthetic as the EA repair in the cath lab and operating room? What about a hybrid procedure prior to the TEF repair?

What are your thoughts on post-operative pain management? Is an epidural appropriate? Thoracic or caudal?

Intraoperative care:

The surgeons request that prior to intubation a bronchoscopy is performed to characterize the overall anatomy and the location of the fistula. The surgical plan is for complete repair (ligation of TEF and anastomosis of EA) via a left thoracotomy.
What is your plan for induction of anesthesia? Is a rapid sequence induction necessary? What FiO₂ would you use? Is a rigid bronchoscopy prior to intubation an appropriate request? How would you manage anesthesia and ventilation in this patient during bronchoscopy if not intubated?

Immediately after intubation, which is confirmed with end-tidal CO₂ and auscultation, increasing gastric distension occurs and the SpO₂ drops 20% from baseline.

What is the differential diagnosis? How will you place the endotracheal tube and verify position? What are the pros and cons of intentional mainstem intubation in this patient? What are your thoughts on using a fogarty catheter? Cuffed vs. non-cuffed endotracheal tube?

After optimal placement of the endotracheal tube, the patient’s saturations stabilize and surgical prep begins.

What maintenance anesthetics would you use? How would you plan to manage the FiO₂? What is Qp:Qs and what is your plan to manage Qp:Qs during the anesthetic?

Two minutes after beginning lung retraction, the SpO₂ drifts down to the 60s, and the blood pressures are stable if not slightly increased.

How would you manage desaturations during lung retraction? Would you expect to see any differences on the pre- and post-ductal saturations during this time? Is the end-tidal CO₂ representative of the pCO₂? Would a thoracoscopic approach be better for this patient?

While continually optimizing ventilation and communicating with the surgeons, the SpO₂ again drifts down to the 60s and the blood pressure drifts down to 36/17.

What is the differential diagnosis? How will you treat? Which vasopressors would you consider using during this anesthetic? What are your thoughts about fluid management and the effects on the Starling curve in this patient?

Surgery finishes with successful repair.

Would you extubate the patient? Would you continue paralysis? What FiO₂ would you use during transport?

Discussion

Congenital EA/TEF has an incidence of 1:2500-1:3000 live births. The majority of patients with congenital EA have an associated TEF, the most common arrangement being a blind-ending upper oesophageal pouch with a distal fistula. Other arrangements are much less common (Table 1). Nearly 50% of patients with EA/TEF possess one or more additional congenital malformations (Table 2). Cardiac defects consistently account for the greatest number of anomalies and may be a subset of VACTERL (Vertebral, Atresia [duodenal and anorectal], Cardiac, Tracheoesophageal, Renal, Limb) or CHARGE (Coloboma, Heart, Atresia choanal, Retarded growth, Genital hypoplasia, Ear deformities) syndromes. EA/TEF occurs as a subset of VATER/VACTERL associations in 10% of cases and is also associated with chromosomal anomalies such as trisomy 18 and 21 and DiGeorge syndrome.
Table 1: EA/TEF Classification

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<thead>
<tr>
<th>Name</th>
<th>% of total</th>
<th>Description</th>
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<tbody>
<tr>
<td>Gross C; Vogt IIb</td>
<td>86%</td>
<td>EA with distal TEF</td>
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<tr>
<td>Gross A; Vogt II</td>
<td>7%</td>
<td>Isolated EA</td>
</tr>
<tr>
<td>Gross E, “H-Type”</td>
<td>4%</td>
<td>TEF without EA</td>
</tr>
<tr>
<td>Gross B; Vogt III</td>
<td>2%</td>
<td>EA with proximal TEF</td>
</tr>
<tr>
<td>Gross D; Vogt IIIa</td>
<td>&lt;1%</td>
<td>EA with proximal and distal TEF</td>
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Table 2: Associated anomalies in patients with EA/TEF

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<tr>
<th>Organ system</th>
<th>Incidence</th>
<th>Examples</th>
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<tbody>
<tr>
<td>Cardiovascular</td>
<td>29%</td>
<td>VSD, PDA, TOF, ASD, and right sided aortic arch</td>
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<tr>
<td>Gastrointestinal</td>
<td>14%</td>
<td>Duodenal atresia, imperforate anus, malrotation, pyloric stenosis, omphalocele</td>
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<tr>
<td>Genitourinary</td>
<td>14%</td>
<td>Renal agenesis, hypospadias, horseshoe or polycystic kidneys, ureteric abnormalities</td>
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<tr>
<td>Musculoskeletal</td>
<td>10%</td>
<td>Radial limb abnormalities, polydactyly, lower limb defects, hemivertebrae, rib defects, and scoliosis</td>
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<tr>
<td>Respiratory</td>
<td>6%</td>
<td>Trachea-bronchomalacia, pulmonary hypoplasia, tracheal agenesis/stenosis</td>
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Infants with EA are unable to swallow saliva or milk and have excessive salivation requiring repeated suctioning. Diagnosis, if not established prenatally, is quickly suspected with failure to pass a suction tube >10 cm. Repair of EA is not a surgical emergency; however, pre-term infants with severe respiratory distress must be managed with intubation preoperatively. Positive pressure ventilation risks ongoing gastric distension and respiratory embarrassment, and urgent surgery may be considered. The surgical goal in most newborns is a right thoracotomy, ligation of the TEF, and anastomosis of the esophagus. For the past decade, a thoracoscopic technique has been used with similar results. Very low birthweight infants (< 1500 g) may benefit from a two-staged repair which involves ligation and division of the TEF with concurrent gastrostomy placement followed by delayed primary repair when the infant has achieved a weight > 2000 g.

HLHS is rare, accounting for 2-3% of all congenital heart disease and occurring in approximately 2 per 10,000 live births per year. During fetal development, the structures of the left side of the heart are underdeveloped and unable to function properly at birth. Specifically, the left ventricle, the aortic and mitral valves, the ascending aorta, and the aortic arch are hypoplastic or atretic, thus with varying degrees of severity. Ductal-dependent heart diseases such as HLHS impact anesthesia in that the balance of pulmonary (Qp) and systemic (Qs) circulations must be carefully managed during all phases of the anesthetic. Neonates with lung prematurity, history of prior aspirations, and who undergo surgical compression of the lung may have increased pulmonary vascular resistance, low Qp:Qs, and may exhibit marked hypoxemia.

Usual surgical palliation of HLHS involves three stages: Norwood operation in neonates, bi-directional Glenn shunt at 6-8 months of age, and Fontan procedure at 18 months to 5 years of age. Currently, survival is approximately 65% at 5 years of age and 55% at 10 years. The hybrid procedure (balloon atrial septostomy, surgically placed pulmonary artery bands, and stenting of the ductus arteriosus) is an alternative procedure to the Norwood operation. The hybrid procedure is most often used in those neonates with HLHS who are at significantly increased risk of undergoing cardiopulmonary bypass or in those who are not likely to survive through complete palliation and require a palliative bridge to heart transplant (e.g., poor ventricular function, tricuspid valve disease).
The presence of congenital heart disease confers higher mortality in the perioperative period. The major predictors of survival in infants undergoing repair of EA/TEF are birth weight, presence or absence of cyanotic heart disease, and pneumonia. A recent study looking at TEF/EA repair and survival rates demonstrated survival rates of 100% for infants >2 kg and without heart disease; however, survival was 72% and 27% for infants with major cardiac anomalies and with birth weights greater than or less than 2.0 kg, respectively (Okamoto 2009). In a small, recent series of infants undergoing EA/TEF repair (Diaz 2005), infants with ductal dependent lesions had a significantly higher mortality rate (57% vs. 10%) than those with non-ductal dependent lesions. Also, infants with CHD had a higher rate of intraoperative events including difficult ventilation, desaturation > 10% from baseline, increased use of inotropic agents, and transfusion of blood products.

In summary, this case illustrates the complex interplay of cardiac and respiratory physiology that a neonate with ductal dependent heart disease and EA/TEF presents to the anesthesiologist.

References: