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Difficult esophageal atresia: Trick and treat

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ABSTRACT

Although most patients with esophageal atresia (EA) and tracheo-esophageal fistula (TEF) may benefit from "standard" management, which is deferred emergency surgery, some may present unexpected elements that change this paradigm. Birth weight, associated anomalies, and long gap can influence the therapeutic schedule of the patients with EA/TEF and can make their treatment tricky. As a consequence, detailed information on these aspects gives the power to develop a decision-making process as correct as possible. In this article, we will review the most important factors influencing the treatment of patients with EA/TEF and will share our experience on the diagnostic and therapeutic tips that may provide pivotal help in the management of such patients.

Introduction

In the last 50 years, the concept of esophageal atresia (EA) in pediatric surgeon's mind has not changed much. Willis J. Potts, pioneer of pediatric surgery, once stated, "to anastomose the ends of an infant's esophagus, the surgeon must be as delicate and precise as a skilled watchmaker. No other operation offers a greater opportunity for pure technical artistry." Since then, the approach to EA patients has been poorly modified, with no consensus on preoperative assessment, perioperative treatment, and postoperative care.

EA, with or without tracheo-esophageal fistula (TEF), remains the most common congenital anomaly of the esophagus. The current mortality is minimal[1,2] in spite of the frequent association with other malformations and the reported decline in trainee experience.[3] The survival improvement is probably dependent on several factors such as advances in neonatal intensive care, anesthesia and intraoperative monitoring, refined surgical approach and techniques, management of complications, parenteral nutrition, and antibiotics. Despite the outcome being generally good, EA may still represent a challenge to pediatric surgeons.[4] Nowadays, the centralization of these patients and their rigorous and homogeneous preoperative assessment are not yet a common standard.

The focus of surgeons with EA subspecialty interest shifted recently to the "more difficult" cases. Two main aspects can be included in the definition of "more difficult": (1) *general patient's characteristics* such as prematurity and associated life-threatening cardiac and bowel anomalies and (2) *specific surgery-related issues* including laryngotracheal anatomy, esophageal anatomy, and redo surgery. For these babies, the optimal timing of procedure and preoperative assessment, the definition of the so-called "long gap," and the possible approaches to difficult or impossible anastomosis (including technical refinements) are of paramount importance but still source of controversies.

In this article, we will endeavor to browse all these topics to provide the most recent data and knowledge regarding perioperative diagnostic assessment and therapeutic approaches, to highlight helpful refinements in difficult cases, and to suggest a possible personal flowchart as a guide.

General patients' characteristics

In the vast majority of patients with EA/TEF, operative repair is urgent but not an emergency, and overrides the treatment of other associated problems. However, in specific clinical conditions, it may be necessary to step out from this paradigm. The most important patients' characteristics that can change the treatment schedule include birth weight (and prematurity) and some associated anomalies, which are also the most important factors in determining survival in historic risk classifications.[1,5]

Very low-birth-weight (VLBW) neonates (< 1500 g) with EA/TEF represent a distinct subgroup of patients that need particular therapeutic attention. Prematurity and low birth weight are relatively frequent in association with EA/TEF, especially in patients without distal TEF, due to reduced amniotic fluid absorption and subsequent polyhydramnios. VLBW infants are prone to develop respiratory distress syndrome and require mechanical

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ventilation.[6,7] In case of decreased lung compliance, increased airway resistance, or need for mechanical ventilation, the fistula may act as a low-resistance vent through which air may preferentially pass into the esophagus. As a consequence, the loss of ventilation and the progressive gastric distension will worsen the respiratory distress syndrome, also facilitating gastroesophageal reflux and inhalation, increasing the risk of gastric perforation with high mortality rate.[8] For VLBW neonates with EA/TEF and respiratory distress, several maneuvers have been suggested to prevent or to stop the air leak through the fistula, including gastric division,[9] silastic banding of the distal esophagus,[10] distal positioning of the endotracheal tube with the bevel pointing anteriorly,[11] antegrade[12] or retrograde[13,14] occlusion of the fistula with a Fogarty balloon, water-seal gastrostomy,[15] and high-frequency ventilation.[16] Most recent series suggest early thoracotomy with ligation of the fistula.[17,18] In the emergency setting, the approach should be transpleural to expedite the operation. If the patient is stable, the extrapleural approach is preferable due to the possibility of more severe consequences of an anastomotic leak after a transpleural approach. The fistula should be ligated in continuity, ideally with non-absorbable sutures. After the fistula is closed, if the patient is stable and the anatomy is favorable (see section Preoperative assessment of EA), the fistula may be divided and a primary anastomosis can be attempted. Otherwise, a delayed primary anastomosis should be programmed. In VLBW infants, the esophageal tissue is extremely fragile. Therefore, lengthening maneuvers should be performed very cautiously as, with the words of Dr. Rickham, "the tissue paper thick walls of the 2 segments will disintegrate."[19] Minimal handling is recommended on the ends of the esophagus completing the anastomosis with no more than 8 stitches. When the fistula is ligated in continuity and a delayed anastomosis is programmed, this should be attempted within 1–2 weeks, as failure to do so may result in recanalization of the TEF.[20,21] Other authors[18] wait until the patient is clinically stable and reaches 2 kg. In such cases, to avoid the risk of refistulization, the fistula is ligated, divided, and anchored to the spine with moderate traction if needed. In addition, a cervical esophagostomy (CE) should be considered to make the nursing of these patients easier and to reduce the risk of saliva aspiration (see section Difficult/"impossible" anastomosis).

VLBW infants with EA/TEF and respiratory distress are at high risk of gastric rupture. In a large series of 623 patients, 6 developed a gastric rupture, 5 of which were preterm.[8] If such a complication occurs, tension pneumoperitoneum may lead to diaphragmatic splinting, further reducing the lung compliance and deteriorating cardiorespiratory status. Emergency abdominal decompression should be performed, quickly followed by laparotomy, and emergency gastrostomy for air leak control with a Foley catheter in the lower esophagus. In the series from Maoate et al.,[8] this had an immediate beneficial effect on ventilation allowing the authors to proceed to the division of the fistula through an extrapleural approach and primary esophageal anastomosis.

Associated anomalies are very frequent in patients with EA/TEF. The cardiovascular system is the most commonly affected with a prevalence reaching 50%.[22] Major cardiac anomalies may cause a hemodynamic instability so severe that it does not allow prolonged thoracic surgery. In these cases, emergency treatment of the cardiac anomaly may represent a priority over esophageal surgery. Healey et al.[23] report that out of 118 patients with EA/TEF, 23 required delayed primary repair. The presence of a life-threatening associated cardiac anomaly was the second most frequent cause of primary anastomosis delay after long-gap EA.[23] In a small series of 6 patients with both EA/TEF and congenital cardiac anomalies, Hayashi et al.[24] report delayed primary anastomosis in 4 of them. They had a gastrostomy performed the first day of life, and the anastomosis was delayed between 3 days and 3 months of life. All above data indicate that early echocardiography is strongly recommended to tailor the treatment specifically to each patient.

Right aortic arch (RAA) and right descending aorta may cause operative difficulties in patients with EA/TEF. Ideally, preoperative cardiac ultrasound should be also performed to define the situation of the aortic arch. However, preoperative recognition of a RAA may be difficult with a low detection rate even in experienced hands.[25] As a consequence, the surgeon should be prepared to face a RAA even with a negative preoperative ultrasound scan. With preoperative diagnosis of RAA, a left thoracotomy is suggested.[25,26] Other authors[27,28] also achieved satisfactory results via a right thoracotomy in all patients with a RAA, and they support this approach in all EA patients.

Duodenal obstruction (DO) in the form of duodenal atresia or stenosis may be associated with EA/TEF in up to 6% of patients.[29] Several treatment options have been proposed for this association. Spitz et al.[30] recommend that when a TEF is present, the primary approach should be to divide the fistula and repair the EA, performing a wide gastrostomy to decompress the stomach. For patients with pure EA, they recommend duodenoduodenostomy and gastrostomy leaving a transanastomotic feeding tube. Others suggest to repair the DO first followed by the delayed repair of EA/TEF, ideally after 1 week.[31,32] The advantages of staging the repair with DO first would be to give the opportunity for improvement in growth and pulmonary function and to allow some resolution of the delayed gastric emptying of DO, thus stressing less the esophageal anastomosis. Finally, in patients with pure EA, it offers the possibility for the distal esophageal segment to elongate. As a general principle, when a distal TEF is present, the priority is to avoid the risk of inhalation. Thus, emergency decompressive gastrostomy, immediately followed by the closure of the fistula, is recommended. Then, if clinical conditions are stable, esophageal anastomosis and duodenoduodenostomy should be considered in the same operation. If the clinical conditions remain critical, the esophageal and/or duodenal anastomoses should be delayed after stabilization. In patients with no distal TEF, duodenoduodenostomy and gastrostomy are the first surgical steps. The esophageal gap can be measured (see section Preoperative assessment of EA) and a delayed esophageal anastomosis planned.

Congenital diaphragmatic hernia (CDH) is very rarely associated with EA, with an estimated incidence of 1:200,000.[33] The coexistence of these anomalies carries a very poor prognosis, with a survival rate below 30%.[34] The presence of a distal TEF poses serious management problems when associated to CDH as it greatly limits the possibility of positive pressure ventilation. On the one side, the escape of gasses through the fistula reduces the ventilation to the lungs, and on the other side, it leads to progressive inflation of the bowel loops that further compress the hypoplastic lungs. For that reason, urgent fistula closure is necessary. A delayed esophageal anastomosis can be done when stabilization is obtained, at the time of CDH repair.

Chromosomal anomalies are found in up to 10% of patients with EA.[35] The most common is Edwards syndrome (trisomy 18), followed by Down syndrome (trisomy 21), and more rarely Patau syndrome (trisomy 13). The occurrence of a chromosomal anomaly in a patient with EA may represent an ethical quandary. This is well illustrated by the case of Baby Doe who was born in April 1982 with Down syndrome and associated EA.[36] Currently, that case would not pose the same ethical problems, as the presence of Down syndrome is not regarded anymore as a motivation for treatment withhold. Nowadays, Edwards syndrome and Patau syndrome are facing the same debate. In such syndromes, with poor long-term survival, surgical treatment of associated EA may be seen as cause of unjustified, prolonged suffering to the infant and a waste of the scarce medical resources. A recent report[37]