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Surgical Approaches to Congenital Anomalies of Esophagus

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Abstract

With prevalence of about 1 in 3000 live births, pediatric surgeons commonly deal with esophageal abnormalities, which may provide substantial clinical complications. Surprisingly, the embryologic processes underlying esophageal atresia (EA) with or without tracheoesophageal fistula (TEF), one of the hallmark disease entities of pediatric surgery, have only lately been largely uncovered. When it comes to the treatment of congenital esophageal abnormalities, notably esophageal atresia and tracheoesophageal fistula, surgical methods are essential. In order to address the anatomical abnormalities and restore normal function, surgical correction is often necessary in the care of congenital esophageal anomalies, including esophageal atresia and tracheoesophageal fistula. In this review we are going to cover surgical approaches to repair those malformations, long-term outcomes, and latest developments in esophageal surgical approaches.

Keywords: *Tracheoesophageal fistula (TEF), esophageal atresia (EA), abnormalities, surgery*

1. Introduction

Congenital anomalies of the esophagus refer to a group of abnormalities that affect the structure and function of the esophagus, which is the tube that connects the throat to the stomach. These anomalies can occur during fetal development and are present at birth. Esophageal congenital problems affect around 1 in every 3000 births, making them very common conditions. Owing to the respiratory system and the esophagus having similar embryologic ancestry, associated anomalies in the lungs and trachea often correspond with esophageal abnormalities. These abnormalities consist of multiple types of tracheoesophageal fistula and esophageal atresia. It makes sense that the abnormalities might cause problems eating and, in some situations, breathing, necessitating immediate medical intervention during the perinatal stage. Obstetrical abnormalities involving the esophagus may cause a hard delivery situation caused by the reduced capacity to swallow. Awareness of the relationship between congenital esophageal and tracheal anomalies requires a fundamental awareness of the present understanding of esophageal embryology, notwithstanding the enormous complexity of the organogenesis process and

the ongoing research into cellular mediators [1]. Some common congenital anomalies of the esophagus include esophageal atresia, tracheoesophageal fistula, and esophageal dysmotility. Esophageal atresia is the most common congenital anomaly of the esophagus [2]. Esophageal atresia and/or tracheoesophageal fistula (EA/TEF) occurs when the esophagus fails to grow normally and ends up as a pouch in the neck or upper thorax. It is typical to have abnormal communication between the trachea and the distal esophagus. When left untreated, esophageal continuity and the separation of the airway from the digestive tract are compromised, making (EA/TEF) a potentially fatal condition for newborns. Even though survival has significantly increased over the past few decades, EA/TEF-related morbidity and anomalies continue to be highly prevalent [3-6].

The epidemiology, survival, and variables associated with mortality are well described by recent reviews of EA/TEF management using large administrative databases, such as the Pediatric Health Information System (PHIS) and the Kid's Inpatient Database (KID). The inability to distinguish crucial clinical components, such as operative anatomic findings, long-term follow-up, and the requirement for additional outpatient intervention, limits these investigations [3-5, 7, 8].

The absence of esophageal continuity is a characteristic of esophageal atresia, which can occur with or without tracheoesophageal fistula, an embryonic abnormality of the foregut. Three categories exist for EA/TEF: (1) a category based on the presence of tracheoesophageal (TE) anomalies in a known genetic syndrome (syndromal or non-syndromal), (2) the anatomical category relying on the existence and place of atresia and fistula, (3) a category based on the association with other congenital anomalies (isolated or non-isolated) [9, 10]. The atresia is linked to a TEF, or a distal esophageal–tracheal connection, in the great majority of patients (78.0–91.8%). The remaining patients consist of those with an atresia alone (5.0–13.0%), a fistula (2.4-6.5%), an atresia plus a proximal tracheal connection (0.4-5.7%), or an atresia plus a distal and proximal fistula (0.1-2.6%). TE anomalies are linked to other congenital defects in about half of the patients. Many genetic and environmental factors are thought to play a multifactorial role in the development of non-syndromal EA/TEF [9, 10].

Over the past seven decades, there has been progress in the prognosis and course of treatment for infants with congenital abnormalities such as EA, TEF, and others. Improving the care provided to expectant and newborn babies has been crucial in lowering the morbidity and death rates related to these illnesses. Adjacent congenital defects and pulmonary complications are currently the main causes of unfavorable outcomes [2].

A poor outcome is more likely in infants with severe cardiac abnormalities or very low birth weights. The prognosis and survival rates of these high-risk newborns would increase with advancements in prevention and care. Furthermore, improved prenatal screening for congenital anomalies such as TEF and/or EA enables improved prenatal counseling and delivery preparation at a tertiary medical facility [2, 11]. Currently, thoracoscopy is used to repair esophageal defects; in the future, robotic-assisted surgery may be employed. The treatment of these infants may eventually be further improved by tissue engineering for esophageal replacement, in-utero intervention, and minimally invasive procedures like thoracoscopy and robotic assistance [2, 12].

Surgical Approaches to Esophageal Anomalies

Surgical approaches play a crucial role in the management of congenital anomalies of the esophagus, particularly in cases of esophageal atresia and tracheoesophageal fistula. The management of congenital anomalies of the esophagus, particularly esophageal atresia and tracheoesophageal fistula, often involves surgical repair to correct the structural abnormalities and restore normal function. The goal of surgical approaches in the management of congenital anomalies of the esophagus is to establish a functional connection between the proximal and distal ends of the esophagus, allowing for the passage of food and liquids from the mouth to the stomach. Various surgical approaches may be used to address congenital anomalies of the esophagus. These approaches can include primary repair, esophageal elongation procedures, and esophageal substitution [13].

The focus is now on lowering morbidity and improving the quality of life for these patients since pediatric surgery facilities have a survival record for these children that are higher than 90%. There are several surgical approaches that can be used in the management of congenital anomalies of the esophagus. One common surgical approach is primary repair, which involves directly connecting the proximal and distal ends of the esophagus. This is typically done using sutures to create an anastomosis and restore the continuity of the esophagus. Another surgical approach is esophageal elongation procedures, which are used when there is a gap between the proximal and distal ends of the esophageus that cannot be directly repaired [14].

A bronchoscopy and laryngoscopy should be performed on every newborn prior to an open surgical correction of TEF/EA. Prior to primary treatment, tracheomalacia, tracheobronchitis, and the fistula's levels are all determined via bronchoscopy and laryngoscopy. Additionally, bronchoscopy helps clarify laryngeal anomalies such as aortic arch location, various fistulas, laryngomalacia, posterior laryngeal cleft, and vocal cord dysfunction. The results of a bronchoscopy may be used to design surgical repairs. Mid-tracheal fistulas are linked to minimum gap atresia, while carinal fistulas are related with large gap atresia [14].

A right posterolateral thoracotomy, fistula closure, and the formation of a main esophageal anastomosis are all necessary for open surgical repair of TEF/EA. Echocardiography is required for preoperative assessment because a right-sided aortic arch, which is present in 2.5% of patients, indicates a greater risk of morbidity and requires a left thoracotomy. Additional VACTERL (Vertebral, Anorectal, Cardiac, TracheoEsophageal, Renal and Limb) defects may be prevented via a renal ultrasound, spinal ultrasound, and limb radiography. Anastomotic leak, chronic second upper pouch fistula, esophageal stricture, recurrent fistula, persistent laryngeal nerve damage leading to voice cord paralysis, and mortality are among the complications after initial repair. It is quite uncommon for a recurrent TEF to spontaneously close. Usually, when there are more than two vertebral bodies dividing the upper and lower esophageal segments, a primary anastomosis cannot be accomplished. Surgical alternatives in this case include the Foker procedure, mobilization of the distal esophageal segment to the diaphragmatic hiatus, and lavaditis myotomy. The patient is at risk for reflux disease, an esophageal stricture, and a higher incidence of leakage from an esophageal anastomosis made under strain [14]. Tom Lobe and Steve Rothenberg performed the first minimally invasive thoracoscopic TEF repair sixty years after the first successful primary repair. Only highly skilled pediatric surgical facilities should use minimally invasive procedures, which have not been shown to reduce the risk of stricture and anastomotic leak [15]. If done correctly, thoracoscopic surgery reduces morbidity by avoiding an open thoracotomy and offers great visualization of anatomic features [16]. Aside from probable chest wall deformity, scoliosis, rib fusion, muscular contractures, and persistent discomfort, avoiding open surgical treatment also avoids these issues [17]. In the past, proximal pouch decompression, a gastrostomy, and measuring the distance between the proximal and distal esophageal segments have all been used in the management of esophageal reflux disease. Those handlings provide patients enough time to develop linearly, which might result in esophageal lengthening [14]. However, in order to prevent aspiration, immediate surgical care entails creating a gastrostomy for feeding and maintaining continuous suction of the blind esophageal pouch. Primary repair utilizing the natural esophagus or replacement operations using sections of the stomach or large intestine are the options for reconstruction. Since replacement operations increase the risk of repeated aspiration and persistent respiratory problems, preservation of the original esophagus is preferred. If primary repair is not possible, a phased surgery may be carried out as the baby becomes older and the esophagus elongates. Although results are still preliminary, techniques including bougienage, electromagnetic stimulation, and graded strain applied to the severed esophageal segment using traction sutures may mechanically extend the esophageal segment [13, 18]. Implementing a tiered strategy has been linked to better outcomes for babies with very low birth weight. The process of repairing H-type fistulae involves dissecting the cervical neck to reveal the area where the fistula has to be separated and fixed [19]. There is a chance of operational trauma and recurrent laryngeal nerve damage with this surgical treatment. Although little experience has been gained, the Nd:YAG laser has also been used to treat H-type fistulas [13].

Pros and Cons of Different Surgical Techniques

Different surgical techniques have their pros and cons when it comes to the management of congenital anomalies of the esophagus. Primary repair is a direct and straightforward approach that aims to restore the continuity of the esophagus. However, it may not be feasible in cases with a significant gap between the proximal and distal ends of the esophagus. Esophageal elongation procedures can be effective in bridging this gap, but they may require multiple surgeries and have a higher risk of complications. Esophageal substitution is another surgical approach that may be used in the management of congenital anomalies of the esophagus. Esophageal substitution involves replacing a portion of the esophagus with a segment of tissue from another part of the body, such as the stomach or colon. This approach can be effective in cases where primary repair or elongation procedures are not possible [20].

Long-term Outcome of Esophageal Surgery in Children

In the first year after repair, follow-up appointments must be made often. Until the kid reaches school age, visits may be reduced to once or twice a year if they are performing well. The kid may only be able to handle pureed food up to the age of 12 to 18 months and then diced food until the age of 5 years due

to damage at the anastomosis. By the time they are five years old, the youngster usually knows how to chew food properly before swallowing it and has grown enough teeth to help with it. The symptoms of GER, recurring fistula, tracheomalacia, and other consequences should be explained to the child's parents [2].

GER

GER is often seen after EA/TEF repair. It is characterized by the reflux of stomach contents that results in symptoms including coughing, poor weight gain, irritation, heartburn, and recurrent regurgitation with or without vomiting. Congenital dysmotility of the esophagus, alterations to the angle of His, damage to the vagus nerve after surgery, and wider esophageal hiatus are the causes of gastric reflux disease (GER). Incompetence of the lower esophageal sphincter mechanism may also contribute to GER. Peptic esophagitis, eosinophilic esophagitis, metaplasia, and recurring anastomotic strictures are only a few of the serious GER sequelae [21-24]. Specifically, prolonged gastric reflux disease (GER) may result in Barrett's esophagus, a premalignant ailment marked by metaplastic alterations in the esophageal epithelium. In order to evaluate and correlate non-acid GER with symptoms in a subset of patients, including those who are symptomatic on antireflux medication (proton pump inhibitors, or PPI), on continuous feeding, experiencing extra-digestive symptoms, and having a normal pH probe and endoscopy, the ESPGHAN-NASPGHAN guidelines for the evaluation of GER in children with EA recommend using pH-impedance [25]. Following TEF/EA repair, there are several different ways to treat GER, ranging from conservative anti-reflux medication to various anti-reflux surgical techniques [26].

Dysphagia

With rates as high as 75–100%, dysphagia is one of the most prevalent symptoms and complaints among children and adults who had EA/TEF repair [27]. According to recent research by Coppens et al., the prevalence of dysphagia declines with age: 51% in patients under 1 year old, 51% in those between 1 and 4 years old, 17% in patients between 5 and 11 years old, and 21% in patients between 12 and 18 years old [28]. Studies using manometry have shown either no or weak esophageal peristalsis together with a compromised or nonexistent contraction pattern [29]. Rat experiments have shown an aberrant intrinsic innervation of the distal esophagus that impacts intramural nerves that are both excitatory and inhibitory [23]**.**

With 36% and 75% of patients experiencing problems with the oral and pharyngeal stages of swallowing after EA/TEF repair, respectively, the video fluoroscopic swallow study is useful in providing an objective evaluation. Using the Functional Oral Intake Scale, which has seven levels ranging from nothing by mouth (level 1) to a complete oral meal without limits (level 7), the degree of dysphagia after TEF repair may be non-invasively assessed**.** In addition to structural airway abnormalities such laryngomalacia, vocal cord paralysis, tracheomalacia, oropharyngeal abnormalities, and laryngeal clefts, children with TEF/EA and long-term dysphagia often also have these conditions. As a result, symptoms including choking episodes, aspiration, chest pain, or food impaction may be present in both dysphagia and respiratory dysfunction. In children with corrected EA, dysphagia is also highly linked to reflux reflux disease (GER), independent of whether fundoplication surgery was performed as an anti-reflux measure [28, 30].

Latest Developments in Esophageal Surgery

Latest developments in esophageal surgery aim to improve the outcomes and minimize complications of surgical approaches for congenital anomalies of the esophagus. These developments include advancements in minimally invasive techniques, such as laparoscopic or robotic-assisted surgery, which can lead to reduced postoperative pain and faster recovery times for patients. In addition, tissue engineering and regenerative medicine approaches are being explored as potential alternatives for esophageal substitution. These approaches involve using biocompatible materials or stem cells to create functional esophageal tissue that can be implanted in patients with esophageal defects [31].

The goal of robot-assisted minimally invasive esophagectomy (RAMIE) is to retain favorable oncological results while perhaps overcoming the technical constraints of minimally invasive esophagectomy (MIE). Using a console, the surgeon manipulates the robotic arms aimed at the patient during RAMIE. This allows for more control over the operating field in three dimensions, increased range of motion with the articulated tools, and stabilization of the patient's natural tremor. Robotics can be used for both the laparoscopic and thoracoscopic portions of the procedure; however, in this study, we concentrated on the thoracoscopic portion (esophagectomy, lymphadenectomy, and

esophagogastrostomy), while non-robotic laparoscopy was used for the abdominal part (formation of the gastric conduit) [32].

4. Conclusion

To reduce the associated morbidity and death, significant congenital abnormalities of the newborn must be detected early and treated promptly. Most of these defects manifest in the early newborn period with respiratory distress or evident exterior deformities if no prenatal identification was made. In order to get the right diagnosis as soon as possible, clinicians need to have a high index of suspicion for these very uncommon illnesses. Surgical repairs have shown to be effective in these abnormalities, thus more surgical developments which are mainly less invasive are being studied.

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