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The History and Legacy of the Foker Process for the Treatment of Long Gap Esophageal Atresia

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ABSTRACT

Historically, children afflicted with long gap esophageal atresia (LGEA) had few options, either esophageal replacement or a life of gastrostomy feeds. In 1997, John Foker from Minnesota revolutionized the treatment of LGEA. His new procedure focused on "traction-induced growth" when the proximal and distal esophageal segments were too far apart for primary repair. Foker's approach involved placement of pledgeted sutures on both esophageal pouches connected to an externalized traction system which could be serially tightened, allowing for tension-induced esophageal growth and a delayed primary repair.

Despite its potential, the Foker process was received with criticism and disbelief, and to this day, controversy remains regarding its mechanism of action - esophageal growth versus stretch. Nonetheless, early adopters such as Rusty Jennings of Boston embraced Foker's central principle that "*one's own esophagus is best*" and was instrumental to the implementation and rise in popularity of the Foker process. The downstream effects of this emphasis on esophageal preservation would uncover the need for a focused yet multidisciplinary approach to the many challenges that EA children face beyond "just the esophagus", leading to the first Esophageal and Airway Treatment Center for children. Consequently, the development of new techniques for the multidimensional care of the LGEA child evolved such as the posterior tracheopexy for associated tracheomalacia, the supercharged jejunal interposition, as well as minimally invasive internalized esophageal traction systems. We recognize the work of Foker and Jennings as key catalysts of an era of esophageal preservation and multidisciplinary care of children with EA.

1. Early history of long gap esophageal atresia

In 1941, Cameron Haight of Michigan successfully performed the first single-staged repair of esophageal atresia and tracheoesophageal fistula (EA/TEF) in a 12-day-old baby harkening in an era of hope for these children who previously would succumb to this rare condition [1]. Despite this, primary repair remained challenging in a cohort of infants where the esophageal ends seemed too far apart for primary repair. The definition of what constitutes a "long-gap" esophageal atresia (LGEA) has varied over time, from definitions based on distance between the two esophageal ends (i.e. >2 vertebral bodies), to functional definitions (i.e. inability to perform a single-stage primary repair), to image based definitions (i.e. lack of intra-abdominal air on initial X-ray) [2,3]. Regardless of definition, before the 1990's, infants with LGEA had few options. Some would be relegated to an early life of nasoesophageal suctioning of the proximal pouch for months, and gastrostomy feeds while awaiting spontaneous growth of the esophageal segments, with the hopes of a delayed attempt at primary repair [4]. Others would be treated initially with an esophagostomy, gastrostomy tube, and delayed esophageal replacement (ER), often with colon or a gastric tube [5–8]. Each interposition exposed the child to its own host of complications and morbidity.

As early as 1954, many were attempting to find a surgical solution to preserve the native esophagus in children with LGEA. Rehbein of Germany tried to connect the esophageal pouches by means of a catheter with metal olives and a thread running through the mediastinum but, unfortunately, after 4 of the 5 children he attempted this on died, the method was abandoned [9]. In 1965, Howard and Myers of Australia introduced elongation bougienage, utilizing intraluminal distention of the proximal esophagus to

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promote esophageal growth, often using a special feeding tube, but this method had risks of perforation and leak [10,11]. In 1975, Hardy Hendren of Boston developed an electromagnet technique built upon Howard and Myers' technique to draw the esophageal ends together but risks of the uncontrolled nature of the magnetic forces alongside significant logistical concerns forced termination of the project [12]. In 1972, the Livaditis technique was introduced which focused on the creation of a circular esophagomyotomy for elongation of the upper pouch but fell out of favor due its risk of leak and diverticular formation due to exposure of the underling mucosa [13,14].

In 1974, Alan Shafer and Tirone David of Ohio described their attempts at an operation in which patients with LGEA had both esophageal pouches mobilized and merely approximated using rethreaded ends of silk to close the lower pouch but allowing the upper pouch to be left open for spontaneous fistula formation [15]. Unfortunately, this technique has lost popularity due to the associated risks for uncontrolled leak and failure of a fistula to occur in some cases [16,17]. In 1994, Ken Kimura described a technique in which a cutaneous cervical esophagostomy of the proximal esophagus was serially translocated down the anterior chest wall until sufficient length was obtained for primary anastomosis, however, it is not widely used due to the need for externalization, and later reinternalization, of the esophagus [18–20].

2. John Foker and the foker process

John Foker, a pediatric cardiothoracic surgeon at the University of Minnesota, recognized the complications and morbidity of the previously described approaches to LGEA repair. Foker postulated that the ends of the esophagus would grow if tension was applied allowing eventual primary esophageal anastomosis, expanding on Kimura's work. Foker would say that *"One can easily see that the esophagus can withstand a great deal of tension."* [21]. He began by placing intra-operative tension on the esophageal ends in cases of moderate sized gaps to promote growth. Later he would devise a method to externalize the traction system and allow the tension to be adjusted outside of the operating room for infants with LGEA [21]. His main objective was to move away from a "*one size fit all*" approach and create a truly versatile technique. This suture-assembled traction system would allow delayed primary repair of the full spectrum of esophageal defects and avoid the use of ERs. In an interview, Foker would say that *"The old way … wasn't good … not even close. So, we tried something new."* In 1993 at the University of Minnesota, Foker would perform his first Foker process with an external traction system, ushering in an era of preservation of the native esophagus [21].

The Foker process generally requires three fundamental stages. The Foker "1" was the initial operation that included esophageal mobilization and placement of pledgeted traction sutures on the esophageal pouches, externalizing them and tying them onto a silastic disc (Fig. 1). This system would allow for incremental tension adjustments on the sutures by placing small segments of a feeding tube under the sutures on a regular basis in the intensive care unit (ICU). The Foker "2" was the subsequent primary repair performed after the esophageal pouches overlapped. Historically, the Foker "3" was the final step in which a Nissen fundoplication would be performed to combat gastroesophageal reflux disease (GERD) that can be prevalent in infants following LGEA repair [21]. It is now selectively performed due to the improvements in medical management of GERD [22].

The Foker process was met with criticism from the academic community. Two of Foker's critics, Lewis Spitz and Arnold Coran, had concerns regarding the safety and effectiveness of this new treatment paradigm. Nevertheless, over time, Spitz and Coran understood the value and implication that the Foker process could have on the future surgical management of those with EA and would ask Foker to write a chapter focused on the updated management of LGEA in their 7th edition of Operative Pediatric Surgery [23].

3. Downstream effects and lessons learned

By 2004, Foker had performed 63 successful repairs using his technique, but the pediatric surgery community was slow to embrace this innovative approach as Foker's results were difficult to reproduce [20,24,25]. With his passion to treat children with EA, Rusty Jennings of Boston Children's Hospital (BCH) contacted Foker who was eager to share his work and demonstrate its reproducible nature. After visiting Foker in Minnesota and witnessing the procedure in person, Jennings would bring this technique back to Boston and spend his career refining the Foker process.

The first patient to undergo a Foker process from BCH, was brought by Jennings to Minnesota in 2004 to perform the joint case with Foker. In 2005, Jennings began to perform the Foker process at BCH for the management of LGEA cases that were not amenable to an initial primary repair. 2009 marked an important milestone for the development of the Foker process, as Foker became a visiting

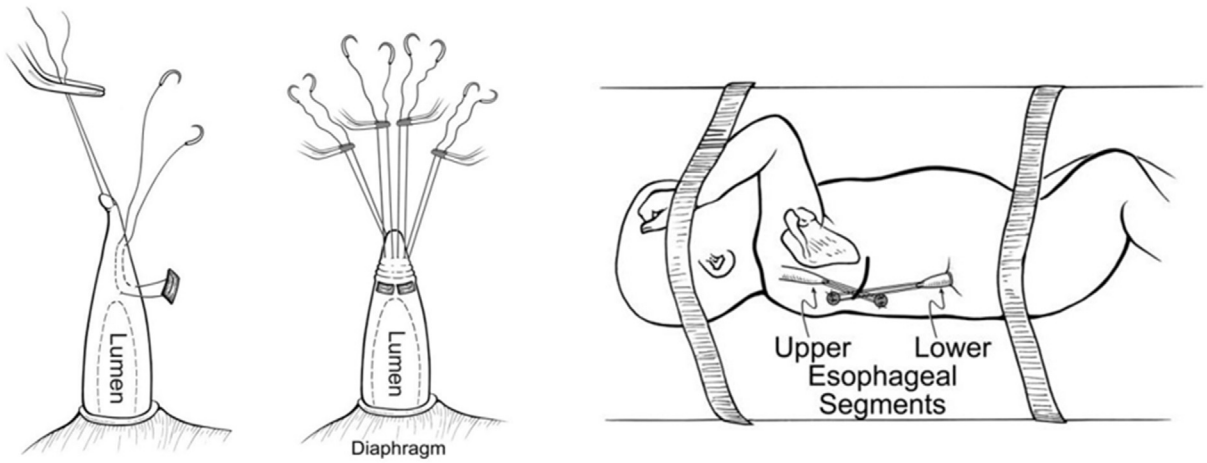

Fig. 1. Example of the Foker "1" with placement of traction sutures on the esophageal pouches and externalization onto silastic discs.

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professor at BCH spurring the creation of the Esophageal and Airway Treatment (EAT) Center, the first of its kind in the United States, and the establishment of a multidisciplinary team and infrastructure required to care for these complex infants [26]. The team included surgery, critical care services, nutrition, gastroenterology, pulmonology, neonatology, otorhinolaryngology, anesthesia, nutrition, and social work, among others.

An uptick in referrals (mostly parent driven) to BCH from all parts of the country for LGEA occurred after 2009 when many parents began hearing word of the innovative surgery to preserve the esophagus and started to formalize modes of communication between each other, forming support groups primarily through social media [27]. From 2005 through 2014, Jennings and the BCH team performed 52 Foker procedures for LGEA, followed by 102 Foker procedures from 2014 to 2020, highlighting the exponential rise in cases as experience accrued and success became evident [28,29]. Foker and Jennings worked hand in hand to continue to grow the program which eventually included transfers from across the world for complex salvage or re-operative cases. The learning curve from 2005 through 2014 presented many challenges as earlier cases at BCH dealt with the consequences of leaks, traction system malfunctions, and strictures in conjunction with new postoperative challenges. Jennings, alongside the multidisciplinary EAT team at BCH, would devise innovative solutions for these challenges, progressing the field forward (Table 1).

As Jennings and BCH gained experience with the Foker process and more complex referrals accrued, the need to have a reliable fallback ER option became evident in situations where a “rescue” Foker was not possible due to lack of healthy esophagus, or in patients with severe esophageal dysmotility and/or refractory reflux, or with dysfunctional or failed alternative ER [29]. Specifically, the referral patterns driven by the Foker process and establishment of the BCH EAT center were key drivers for the implementation and refinement of what is now the supercharged Roux-&-Y jejunal interposition as BCH's and The International Network of Esophageal Atresia's ER of choice [3,30].

The influx of complex esophageal pathology and the postoperative management of patients undergoing the Foker process at BCH also paved the way for significant advances in the field of endoscopic management of EA patients. Michael Manfredi, a pediatric gastroenterologist, working collaboratively with Jennings spearheaded several advances in the endoscopic management of these patients such as endoluminal vacuum assisted therapy (eVAC) for esophageal leaks, endoscopic incisional therapy, and stenting for refractory strictures [31–33].

At the same time, minimally invasive surgery (MIS) continued to rise to the forefront of the surgical world and the benefits of such an approach became evident within the EA community. Thom Lobe and Steven Rothenberg of Colorado undertook the first successful thoracoscopic repair of EA in 1999 demonstrating that an MIS approach, even for EA/TEF, was possible [34].

Later, David van der Zee from the Netherlands and Dariusz Patkowski from Poland, began publishing on their experiences and successes with their MIS iterations of the Foker process [35]. While Patkowski focused on the creation of an internal traction system where the individual esophageal pouches acted as their own

Table 1
Challenges and lessons learned with the implementation of the foker process and management of children with LGEA.

Technical/Operative Challenges	
Challenge	Solution/Lesson
Esophageal Leaks on Traction	Endoscopic guidance during suture placement to prevent full thickness sutures
Adhesions Preventing Esophageal Growth	- Use of thin (0.005 mm) silastic sleeves around each esophageal pouch - Bioresorbable hydrophilic adhesion barrier - Optimal alignment of traction suture bundles with expected vectors of growth so pouches can overlap and not bump into each other
Recurrent Laryngeal Nerve Injury	- Routine pre- and post-operative screening for vocal fold movement impairment regardless of symptoms [45] - Intraoperative recurrent laryngeal nerve monitoring
Large leftward upper pouch and/or Prior failed right sided repair	Consideration of left-sided repair to avoid distorting/shifting esophagus rightward/posterior to trachea and affecting airway/worsening TBM, or to start fresh in a clean operative field [37]
Short upper esophageal pouch (above clavicles)	Incorporate neck dissection to approach upper esophageal pouch
Difficult exposures/re-entry, access to esophageal pouches, rib fractures	Total muscle sparring approach via posterior thoracotomy (auscultatory triangle) with low threshold for multiple interspace entries to approach entire chest, as opposed to one single large incision
Perioperative Management Challenges	
Challenge	Solution/Lesson
Monitoring of Traction System	Placement of leading and trailing clips on each esophageal pouch [46]
Traction System Disruption	- Utilization of thin monofilament sutures (often 5–0 prolene) - Chemical muscle paralysis (if external traction) - Standardization of traction adjustments - Early reoperation if disruption suspected
Optimal Frequency of Traction System Adjustments	Bedside traction adjustments every other day to every third day, allowing the esophagus time to heal
Consequences of Muscle Paralysis (DVT, fractures, volume overload, pressure ulcers) [47,48]	- Consider minimally invasive approach whenever appropriate - Routine chemoprophylaxis while paralyzed - Daily paralytic holiday - Judicious use of diuretics and minimizing loop diuretics - Fluid restriction - Daily physical therapy with passive range of motion on extremities - Padding of bony prominences and frequent repositioning
Proactive Endoscopic Therapy and Surveillance	Expect stricture formation and treat it pro-actively upfront. First endoscopy at one-month post-anastomosis and treat based on initial diameter [49].
Great Vessel Anomalies	Routine preoperative CT scan of the chest with IV contrast and incorporation of arch or vessel anomalies into decision/treatment algorithm [50].
Management of Associated TBM	- Thorough pre- and intra-operative airway evaluation - Potential posterior tracheopexy at the time of esophageal repair [51]

*Numbers in parenthesis correspond to their appropriate reference numbers.

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traction system being pulled together by traction sutures tied to one another, van der Zee would mobilize and place the traction sutures thoracoscopically yet externalize the traction systems and utilize clamps to maintain tension compared to incremental adjustments with feeding tube segments characteristic of the Foker process [35].

Although cases of short to medium gap EA type A cases, amenable to primary repair, began being performed thoracoscopically in 2014 at BCH, it wasn't until 2016 when Jason Smithers and Benjamin Zendejas of BCH would innovate the MIS Foker to tackle cases of truly long gap EA - a thoracoscopic approach with internal traction around the ribs [28] (Fig. 2). They would utilize a different type of internal traction system from Patkowski, one in which the esophageal pouches used an opposing rib as the point of tension, allowing for differential tension to be adjusted on each pouch, as they often grow at different rates, and they can each have a different degree of tolerance to tension. More importantly, this allowed the esophageal pouches to achieve overlap while on traction - a key principle of the classic Foker process. Smithers would continue to broaden this, describing the experience with a left sided MIS approach (even in the setting of a left aortic arch) to create less tracheal intrusion or worsen existing tracheomalacia due to having to move to the right a large leftward upper esophageal pouch. This approach became ideal for those without a thoracic TEF, no significant tracheobronchomalacia (TBM), or as a salvage plan after prior failed right-sided repair [36,37]. Recent studies have reported that whether undergoing a traditional open vs. MIS approach, complications related to the traction system have been limited (i.e., esophageal pouch leaks, traction suture pull-outs, etc.), but unfortunately, esophageal strictures remain a challenge that affects the long-term morbidity of these infants [28,38]. Efforts to decrease anastomotic tension are ongoing including the evaluation of botulism toxin injections to facilitate traction-induced growth and EA repair [39,40].

Prior to the development of an MIS traction system, children who underwent the traditional open Foker process remained sedated and paralyzed in the ICU while on traction. Though the relative harms of sedation and paralysis are often seen as the main deterrents to the Foker process, many have attempted to perform the external traction Foker process without paralysis and have invariably encountered traction system disruptions and esophageal leaks [28]. Hence, the MIS Foker process, which combines the benefits of MIS surgery while maintaining traditional traction principles with an internalized traction system allows children to

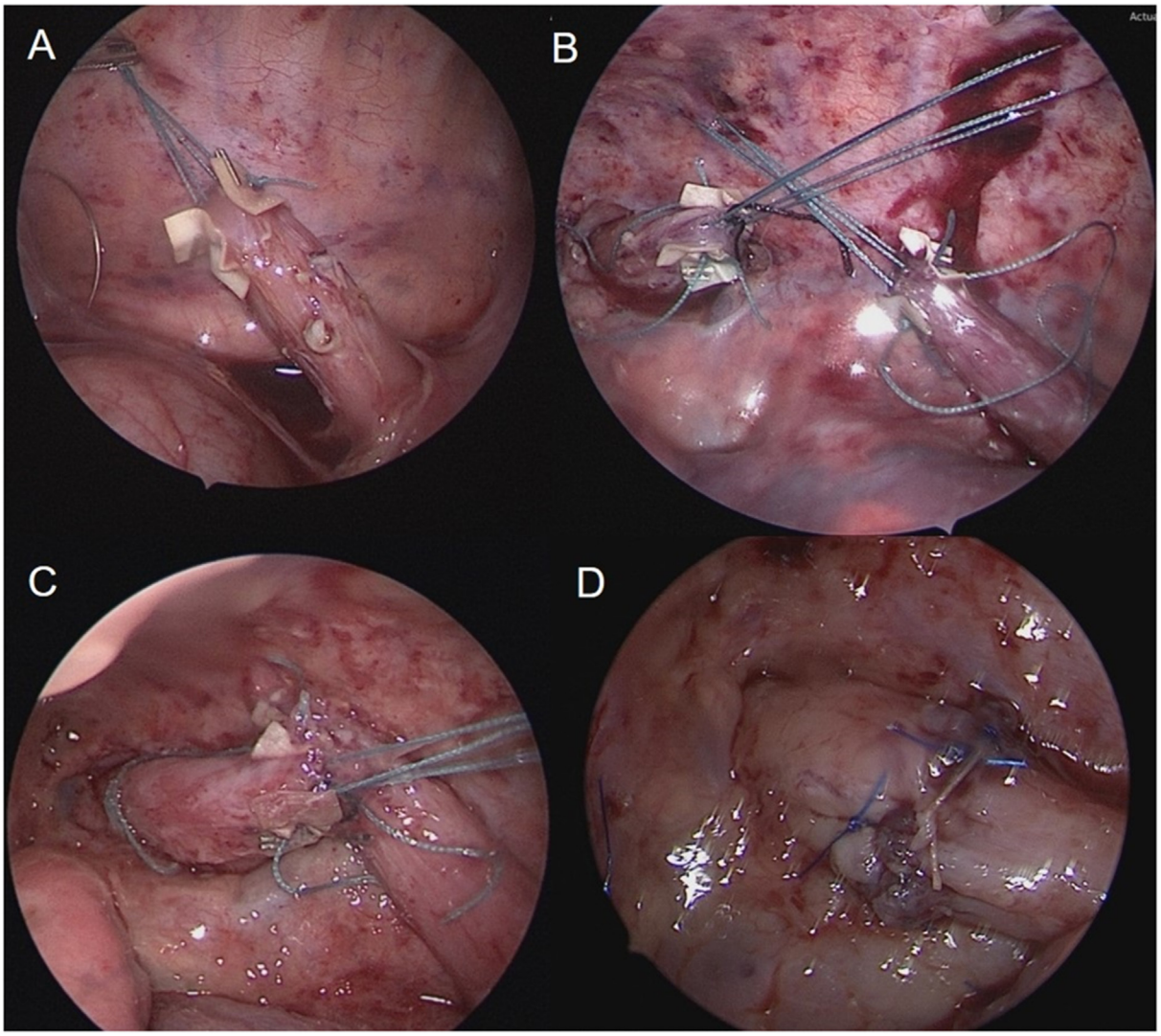

Fig. 2. The minimally invasive foker process. A) Esophageal traction system on lower pouch, B) Esophageal traction system – around the ribs, C) Overlap of esophageal pouches after traction process, D) Anastomosis.

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forgo paralysis while on traction, without increasing traction-related complications, resulting in a true milestone for EA care.

As 2020 approached, the surgical community had now seen not only the stepwise advancement of the Foker process, but the cumulative long term follow-up data to demonstrate its clinical efficacy and ability to achieve normalcy in this subset of patients [28,41]. Led by Jennings, the EAT Center at BCH would serve as a foundational element in aiding the implementation of multidisciplinary care for children with EA. Such philosophy of care has expanded to other centers including All Children's Hospital in Florida, and The Children's Hospital of Philadelphia led by BCH trained EAT surgeons who continue to carry on the work of Foker and Jennings'.

4. The future of the foker process: where do we go from here?

In 2019, The American Pediatric Surgical Association published evidence-based guidelines highlighting the recommendation of utilizing any variant of a staged traction technique to perform a primary anastomosis in the event of failure to complete one in a traditionally delayed fashion - a testament to Foker and Jennings' work [2]. What was once a fatal condition and considered the pinnacle of challenges for pediatric surgeons has undergone stepwise advancement, resulting in new surgical techniques and a new framework for thinking about how surgeons approach the multidisciplinary management of infants born with EA (Table 1).

The Foker process has evolved as experience has grown. Variables such as the esophageal gap length, characteristics of the proximal/distal esophageal pouch, findings on pre-operative bronchoscopy, and prior operative history are some of the features that alter the surgical approach for each patient. Concomitant procedures are now frequently added to treat co-existing pathologies including posterior tracheopexy for those with tracheobronchomalacia (TBM) and intraoperative nerve monitoring to reduce the incidence of recurrent laryngeal nerve injury, especially in re-operative cases [42]. Patkowski and Smithers, among others in select circumstances, have begun forgoing the placement of a gastrostomy tube and the ability to perform a formal gapogram at the index operation, but simply beginning the traction process in the first few days of life. Others, such as Mario Zaritsky and Bethany Slater of Chicago have reinstituted the non-surgical use of magnets in select patients but concerns remain in the form of severe or refractory strictures [43,44].

The evolution of the Foker process can be traced from a once fatal condition to one where not just survival, but normalcy became the expected outcome. History recognizes Foker and Jennings for their work with EA, not only with the creation and evolution of a novel repair emphasizing esophageal preservation, but for its widespread implementation and lifelong dedication to the training of others to carry on the practice.

Conflicts of interest

We declare that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

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