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The first steps in the management of esophageal atresia



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Called to see an infant who could not swallow, Thomas Gibson in 1696 saw the first described case of esophageal atresia (EA) with tracheoesophageal fistula (TEF), the most common of congenital defects of the foregut. Surgeons struggled with the condition from the first attempt by Charles Steele in 1888. Their persistence, epitomized by Thomas Lanman's 1940 review of 32 cases of EA that all ended in death, reached a climax when Cameron Haight in 1941 reported the first patient to survive primary anastomosis for EA. In the 75 years that have followed advances in intraoperative care, neonatology, and surgical technique have produced survival rates that approach 100 percent among babies with EA uncomplicated by prematurity and associated conditions such as cardiac malformations.

Discovery

In 1670 William Durston made the first English description of esophageal atresia (EA) in a pair of conjoined twins. One of the twins, who did not survive, had an esophagus "[that] from the mouth of the right head descended no lower than a little above half an inch off the mid-riff, and there it ended."¹ In 1697 Thomas Gibson made the first description of the most frequently encountered combination of tracheoesophageal anomalies, a proximal EA with a distal tracheoesophageal fistula (TEF).² His careful description clearly describes the symptoms and anatomy.

About November 1696 I was sent for to an infant that would not swallow. The child seem'd very desirous of food, and took what was offer'd it in a spoon with greediness; but when it went to swallow it, it was like to be choked, and what should have gone down returned by the mouth and nose, and it fell into a struggling convulsive sort of fit upon it. It was fleshy and large, and was two days old when I came to it but the next day died. The parents being willing to have it opened, I took two physicians and a surgeon with me... We blew a pipe down the gullet (esophagus), but found no passage for the wind into the stomach. Then we made a slit in the stomach, and put a pipe into its upper orifice, and blowing, we found the wind had a vent, but not by the top of the gullet. Then we carefully slit open the back side of the gullet from the stomach upwards, and when we were gone a little above half way towards the pharynx, we found it hollow no further. Then we began to slit it open from the pharynx downward, and it was hollow till within an inch of the other slit... the imperforated part ... did not seem ever to have been hollow ... the parts were here smooth as the bottom of an acorn-cup. Then searching what way the wind had passed when we blow from the stomach upwards, we found an oval hole on the fore-side of the gullet opening into the aspera arteria [trachea].²

In 1840 Thomas Hill described a case in that was associated with rectal agenesis,³ the first observation of one of the spectrum of associated anomalies known familiarly by the acronym VACTERL (Vertebral, Atresia - duodenal and anorectal - Cardiac, Tracheoesophageal, Renal, Limb).

Failure

In the latter half of the 19th century anesthesia and aseptic surgery allowed bolder operations, including the repair of EA. Timothy Holmes proposed in 1869 a primary anastomosis for EA. While never performing the operation, he described how he would set about finding the two ends of the esophagus.

[The] object would be to cut down upon the point of a catheter passed down the pharynx, and then attempt to trace the obliterated oesphagus down the front of the spine until the lower dilated portion is found.⁴

Charles Steele made the first attempt in 1888 in an infant with EA under chloroform anesthesia. Approaching the defect

through an upper midline abdominal incision he found the gap between the upper pouch and the distal esophagus was an inch-and-a-half, too wide for a primary repair. He abandoned the attempt, and his patient died within 24 hours.⁵

The hazard of the communication between the distal esophageal segment and the trachea was demonstrated by Joseph Brennemann in 1913. He attempted to feed one patient through a gastrostomy, and in a second a jejunostomy. The procedures did not address the communication between the gastrointestinal and respiratory tracts and both died from pulmonary aspiration.⁶

Harry Richter directly addressed the TEF through a thoracic approach in two patients that he reported in 1913. He entered the right sixth interspace, extended the posterior aspect of the incision superiorly, and divided “three or four” ribs. While an assistant’s finger held exposure by depressing the ipsilateral lung, Richter tried to assure adequate inflation of the lung (and presumably the contralateral lung) by maintaining positive pressure within the trachea using a homemade pump. He still was confronted by the unusual anatomy.

*Lack of familiarity with the surgical anatomy of the parts will obviously be a source of embarrassment to most general surgeons. It was to the author. The smallness of the parts in a new-born infant and the peculiarly difficult site of the operation made the hazard apparently.*⁷

He closed the fistula and performed an anastomosis between the ends of the esophagus. Without the benefit of intravenous infusion, positive pressure ventilation, and having only auscultation to verify complete inflation of the lung after operation, he was able to complete both procedures only to have both infants die after operation.⁷ His approach, without the superior extension and division of the ribs above the thoracotomy, would become standard 50 years later.

Thomas Lanman’s (Figure 1) 1940 review of the experience of the Children’s Hospital in Boston with EA is a landmark in its thorough and candid appraisal of the management of these desperately ill infants.⁹ All 32 patients died, including 30 who had undergone a variety of operative procedures. Among the hard-won lessons learned involved care of the infant before surgery. Early diagnosis and treatment was a priority. Aspiration was an ever-present threat, and delays only assured that the baby would be malnourished, dehydrated, and subject to an increasing risk of pneumonia. The diagnosis could be made without giving the child oral barium, a tube defining the limit of the proximal EA and air in the gastrointestinal tract below the diaphragm confirming the presence of a TEF. Children died of pulmonary infection even after technically successful operations. Babies could not tolerate injudicious fluid administration, as one infant who had undergone a successful primary anastomosis appeared to die in pulmonary edema.⁸

The immediate surgical priority was to prevent aspiration by exteriorization of the proximal EA and division of the TEF. The former was a comparatively simple task compared with the latter. Brennemann had showed already that gastrostomy alone was not going to work. The Boston surgeons tried Gage and Ochsner’s operation, dividing the cardia at the level of the esophagogastric junction. It proved to be a failure because still attached to the trachea was a long diverticulum that once filled would spill into the lungs.⁸

A thoracotomy would be necessary to divide the fistula. Surgeons in Boston had adopted a posterior approach to the mediastinum with the baby prone, using a longitudinal incision to the right of the erector spinae, resecting the fourth rib, and dividing the posterior aspects of the ribs above and below. In the pre-antibiotic era they recognized the hazard of an esophageal leak and sought to contain it in the posterior mediastinum. Thus they kept the dissection out of the free



pleural cavity, another principle followed by many surgeons today. They tried to exteriorize the distal esophagus through the incision, but learned a hard lesson about the segmental blood supply of the distal esophagus when it became ischemic and retracted into the chest.⁸

When the two ends of the esophagus appeared sufficiently close and the child's condition would permit, they attempted primary anastomosis. Between 1936 and 1939 they made the attempt in five patients. One patient died during the operation and two within hours after surgery. However, two survived nine days after repair, proving that survival after primary anastomosis was possible. Lanman said, "[Every] reasonable risk should be taken to secure a primary anastomosis."⁸ Despite 32 deaths he said,

That this method will eventually be successful I have no doubt.... Given a suitable case in which the patient is seen early, I feel that, with greater experience, improved technic and good luck, the successful outcome of a direct anastomosis can and will be reported in the near future.⁸

Success

In 1935 a report in the literature appeared of a child with EA without TEF who survived with a gastrostomy and cutaneous esophagostomy for 16 years.⁹ In 1939 William Ladd in Boston and Logan Leven in Minneapolis had babies with EA and TEF born under their care on consecutive days. Both divided the TEF and performed an esophagostomy and gastrostomy. Once survival was assured, each surgeon began to painstakingly construct in stages skin tubes from bipedicle skin grafts from the anterior thorax. Situated on the surface of the chest, the tube received swallowed material from the esophagus above and drained by gravity into the stomach below. The children were the first long-term survivors of EA with TEF.^{10,11}



Finally, in 1941, Cameron Haight of Ann Arbor (Figure 2) fulfilled Lanman's prediction and had a baby survive a primary anastomosis of an EA.¹² Hiram Langston gives an entertaining account of the operation.¹³ The infant, a girl, was in remarkably good shape, surviving 12 days without significant pulmonary contamination, and avoiding dehydration by means of intravenous fluids administered by her pediatrician. She had also

evaded the hazard from a barium swallow, despite aspirating some of the material.

Under local anesthesia Haight entered the posterior mediastinum through a vertical incision in the left chest by resecting the posterior portions of the 2nd through 5th ribs, keeping the parietal pleura intact. The aorta and left subclavian artery were clearly in the field, the latter requiring retraction during the rest of the case. Haight's illustration shows the entire field but clearly exposure was a problem, as he notes that the operative field had to be shifted during the case.¹² He ligated and divided the TEF. The blind end of the upper esophageal segment was nearby. Its mobilization required a deeper plane of anesthesia so open drop ether was administered. Using a single layer of silk sutures, Haight sewed the proximal end to the open distal segment that he had just freed from the trachea. He closed the thoracotomy over a rubber drain that he left near the anastomosis. The child received rectal sulfathiazole by rectum in an effort to address the possibility of perioperative infection.

Saliva appeared in the drain a few days after surgery, the leak at the anastomosis verified by giving the child radio-opaque lipiodol. One of Haight's associates placed a gastrostomy, through which feedings were cautiously begun. They tried to probe the anastomosis with a catheter placed through the baby's mouth. Imagine the alarm when the catheter emerged out of the thoracotomy incision! They reduced the volume of gastrostomy feedings in the hope of avoiding any of it refluxing into the esophagus and out the repair.

On the 21st day after operation the baby happened to burp some evaporated milk from her mouth. Correctly taking it as a sign that the anastomosis was patent, they repeated the lipiodol study. The leak had sealed, the first example of a lesson well known to pediatric surgeons, that esophageal leaks often seal after EA repairs. The child eagerly accepted oral feedings, the only problem being a stricture that required dilation when the child was 17 months old.¹³ The baby was discharged to her family 18 months after her birth. Haight proudly showed a photo of his patient, aged 16, at his presidential address to the American Association for Thoracic Surgery in 1957.¹⁴

She was the last patient Haight saw before his death in 1970. As of a review published in 2005 she was still alive.¹³ Her survival was a signal achievement, being sole survivor and number 10 of a series of 15 patients in his series. The remaining 14 all died within two weeks of their operation.¹²

Epilogue

Still, Haight proved that survival after EA repair was possible. Other reports of successful EA repairs appeared after Haight's signal achievement. Improvements in positive pressure ventilation in newborn infants, intraoperative care, antibiotic therapy, and neonatal intensive care produced a survival

rate of nearly 100 percent among infants that were free from extreme prematurity, other malformations, and heart defects. Advances in surgical technique produced solutions to long-gap EA and obviated the unwieldy skin tubes of Leven and Ladd. Right, not left, thoractotomy exposures are used. No longer do surgeons resect ribs to expose the posterior mediastinum. They routinely enter the pleural cavity to perform thoracoscopic repairs, avoiding a thoracotomy altogether. The 75-year story of surgery for EA demonstrates why one writer described it as the “epitome of modern surgery.”¹⁵

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Legend

- 1 Thomas Lanman, standing fourth from left. The famous 1939 portrait of the surgical staff, The Children’s Hospital, Boston. William Ladd, chief, is to the right of Lanman, Robert Gross is standing at the far right, Orvar Swenson and seventh from the left, seated.
- 2 Cameron Haight. Courtesy University of Michigan Millennium Project.