



Type A esophageal atresia: a critical review of management strategies at a single center

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Received 23 January 2010; accepted 2 February 2010

Key words:

Isolated esophageal atresia;
Primary repair;
Esophagomyotomy;
Collis gastroplasty;
Gastric tube replacement

Abstract

Purpose: The purpose of was to study the short- and long-term outcomes in the management of isolated esophageal atresia with different operative strategies.

Methods: All patients undergoing type A atresia repair over a 15-year period were included. Demographic data, birth weight, gestational age, incidence of associated anomalies, management, and long-term outcomes were studied.

Results: Fifteen patients with type A atresia (9 male) were treated in the study period. The mean gestational age was 35.5 weeks (range, 27-39 weeks), and the mean birth weight was 2179 g (range, 670-3520 g). Eight babies had associated anomalies. Thirteen patients underwent gastrostomy as the initial procedure, and 2 underwent the Foker procedure. In the delayed management group, 9 patients underwent primary anastomosis, with 2 patients needing proximal pouch myotomy. Two patients underwent a Collis gastroplasty. Two patients underwent a cervical esophagostomy and a gastric tube replacement at 4 months and 1 year, respectively. Eight patients (60%) in this group had anastomotic leaks. All patients are currently on prokinetics and proton pump inhibitors. Seven required antireflux surgery. The median length of hospital admission was 4 months (range, 3-19 months). The native esophagus was preserved in 13 (85%) of 15 babies. All patients are alive, and 14 of 15 are capable of feeding orally.

Conclusions: Type A esophageal atresia continues to be associated with significant morbidity despite advances in surgical technique and intensive care.

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The prognosis for babies born with pure esophageal atresia has improved dramatically in the last 2 decades [1]. However, the sequelae of this malformation can be severe with significant adverse effects on growth and development. Many infants remain on parenteral nutrition to overcome

limitations in oral energy intake owing to dysphagia and abnormal deglutition. Furthermore, the remnant esophagus is unable to protect itself against acid reflux, predisposing it to esophagitis, strictures, Barrett dysplasia, and ultimately adenocarcinoma. However, the consensus remains among practicing pediatric surgeons that every effort should be made to conserve the native esophagus, as no other conduit can replace its function in transporting food from the oral cavity to the stomach [2]. This has proven difficult in type A atresia where the distal end can be completely absent and a

Presented at the 41st Annual Meeting of the Canadian Association of Paediatric Surgeons, Halifax, Nova Scotia, Canada, October 1-3, 2009.

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Table 1 Clinical features and demographics

Patient	Birth weight	Age (wk)	Associated anomalies	Initial surgery	Definitive surgery
1	1890	35	Duodenal atresia cloaca hemivertebrae horseshoe kidney	G tube	Delayed end to end
2	2120	36	—————	G tube	Delayed end to end
3	2830	38	—————	G tube	Delayed end to end
4	2870	37	Duodenal atresia glycogenosis IV bicuspid aortic valve	G tube	Initial esophagostomy, Graviiliu gastric tube reconstruction
5	1100	34	ASD, VSD	G tube	Delayed end to end
6	3075	39	—————	G tube	Delayed end to end
7	670	28	Fanconi anemia radial and digital anomalies	G tube	Collis gastroplasty
8	1549	31	Multicystic kidney mosaic karyotype (46XY; 47 XYY)	—	Foker technique
9	2575	37	Neurologic	G tube	Delayed end to end anastomosis with proximal circular myotomy
10	1660	35	Encephalopathy, scoliosis	—	Foker technique
11	2320	39	Duplex kidney, malrotation	G tube	Delayed end to end anastomosis with proximal circular myotomy
12	2840	38	—————	G tube	Esophagostomy, Graviiliu gastric tube reconstruction
13	1570	34	—————	G tube	Collis gastroplasty
14	3520	39.4	—————	G tube	Delayed end to end anastomosis
15	2760	38	—————	G tube	Delayed end to end anastomosis

Table showing the demographics, clinical features, surgical strategy, and outcomes for patients with pure esophageal atresia. PO indicates per os. ASD, atrial septal defect; VSD, ventricular septal defect.

primary anastomosis is all but impossible. Surgeons have devised various ingenious techniques to overcome this “long gap.” In this article, we describe our experience over the last 15 years with managing this complex condition.

1. Methods

After institutional review board approval, the charts of all newborn infants diagnosed with pure esophageal atresia

Age at definitive surgery	Complications	Other surgical interventions	Result	Follow-up
63 d	Reflux, stricture	Toupet, multiple dilatations	Full PO	13 y
98 d	Reflux, Barrett	Boix-Ochoa, Collis Nissen	Full PO	17 y
63 d	Anastomotic leak, peptic esophagitis, hiatal hernia		Full PO	11 y
1 y 2 mo	Anastomotic leak, reflux, gastrostomy necrosis	Antireflux	Partial PO (supplemental jejunostomy feeds)	9 y
85 d	Anastomotic leak, reflux, stricture, gastrostomy complication	Dilatations, mitomycin, Nissen, pyloroplasty gastrojejunostomy	Partial PO (supplemental gastrostomy feeds)	6 y
87 d	Anastomotic leak, Barrett		Full PO	13 y
186 d	Anastomotic leak	Esophagostomy, awaiting colon interposition	Jejunostomy feeds; sham PO feeds	8 y
18 d	Anastomotic leak, esophageal diverticulum, recurrent esophageal diverticulum	Resection of esophageal diverticulum, aortopexy, dilatations, Collis gastroplasty, Nissen, pyloroplasty esophagogastric disconnection, Roux-en-Y esophagojejunostomy	Partial PO feeds	4 y
88 d	Reflux	Toupet	Full PO feeds	13 y
57 d	Pull-through of traction sutures, alkaline reflux		Full PO feeds	<1 y (care transferred elsewhere)
90 d	Anastomotic dehiscence, stricture esophagitis	Thoracotomy and reanastomosis, multiple dilatations	Full PO feeds	5 y
1 y 2 mo	Anastomotic stricture, dysphagia, small bowel obstruction	Laparotomy and intestinal resection, dilatations	PO feeds (some dysphagia to solids)	14 y
107 d	Anastomotic leak, reflux	Decortication multiple dilatations (complicated once by perforation)	Full PO feeds	9 y
84 d	Anastomotic dehiscence, reflux	Thoracotomy and reanastomosis multiple dilatations (mitomycin)	Full PO feeds	1 y
87 d	Anastomotic leak		Full PO feeds	1/2y

were reviewed. Demographic data such as sex, birth weight, and gestational age were obtained. The incidence and types of associated anomalies were noted. The diagnosis of this condition was made based on the inability

to successfully pass a 10F Replogle tube into the stomach while noting a gasless abdomen on the abdominal radiographs. All babies with this anomaly at our institution were admitted to the neonatal intensive care unit. Upper

pouch suction was established through the Replogle tube to prevent aspiration. Intravenous access was established, and broad-spectrum antibiotic coverage was started. Babies underwent an echocardiogram, ultrasound of the abdomen, and radiographs of the spine and limbs. Except for a short period during this study when we tried the Foker procedure, the primary operation performed on these babies (on exclusion of a proximal tracheoesophageal fistula) was a gastrostomy. Care was taken to position the gastrostomy close to the lesser curvature of the stomach so as to preserve the greater curvature and its vascular arcade for potential gastric tube reconstruction in case a primary repair would fail because of the inadequate length of the distal pouch. Associated anomalies needing surgical intervention were dealt with at this time. Babies were admitted to the regular surgical floor once they no longer needed neonatal intensive care, with continuous upper pouch suction. The Replogle tube was irrigated every 4 hours to ensure patency. Gastrostomy feeds were started, and the baby's weight was monitored. After a period of approximately 3 months or when the body weight of the infant doubled its birth weight, thoracotomy was scheduled to reestablish esophageal continuity. If the 2 ends did not come together easily despite adequate mobilization of the proximal pouch, we performed a circular myotomy of the proximal pouch [5]. If the anastomosis was under some tension, we preferred to leave the baby intubated and on a muscle relaxant for approximately 10 days after the procedure. A Collis gastroplasty was performed if there was near-complete atresia of the distal pouch, and we could confidently perform a satisfactory anastomosis with a proximal gastric pouch only. If we felt that a satisfactory anastomosis could not be performed despite all of the above maneuvers, we sutured the proximal and distal pouch to the prevertebral fascia as close to each other as possible. Opacification of both proximal and distal pouches with contrast was then planned before a redo thoracotomy in 2 to 3 months. At this point, if we still felt that a satisfactory anastomosis could not be performed, we performed a cervical esophagostomy and let the infant sham feed orally so as to allow for the development of the oropharyngeal musculature and coordinated deglutition. We noticed microgastria in many of these babies and hence allowed for sufficient time (about a year) to elapse with gastrostomy feeds to ensure that a satisfactory antiperistaltic gastric tube (based on the left gastroepiploic arcade) could be developed. All children were started on antireflux medication, and prokinetic agents were added on a case-by-case basis. An esophagram and upper gastrointestinal series were performed to confirm the integrity of the anastomosis at the seventh postoperative day. Children were discharged from the hospital when they tolerated adequate oral intake, supplemented by gastrostomy feeds, if necessary, at night. Anastomotic strictures were dealt with by elective balloon dilations performed under fluoroscopic guidance. Mitomycin was used topically on

the scar tissue if the stricture was particularly dense. Anastomotic leaks were managed conservatively with adequate chest tube drainage of the pleural cavity and appropriate antibiotic coverage. In our experience, most leaks healed with conservative management. In the case of complete anastomotic dehiscence, we reexplored the chest to better drain the mediastinum and pleural cavity, as well as realign the 2 ends with a transanastomotic stent. Children were followed subsequently in a multidisciplinary esophageal atresia clinic where they were reviewed by a surgeon, pulmonologist, gastroenterologist, dietician, and a physical therapist. Elective 24-hour pH probe studies along with endoscopy were carried out a year after the initial surgery to determine the optimal time to stop antireflux medications. If studies showed ongoing acid reflux, medications were continued; and in case of complicated reflux disease with esophagitis, strictures, and/or Barrett metaplasia, antireflux surgery was planned. Details of medication use, surgical complications, subsequent surgical interventions, and antireflux procedure and its timing were noted. The eating habits of the child were documented, including quantity of oral intake, dysphagia, and the need for enteral tube feeds. The ability of the child to obtain 100% of his energy needs orally without supplementation was considered a success. Statistical analysis was carried out using the Fisher's Exact test for categorical variables and the Student's *t* test for continuous variables.

2. Results

Fifteen infants with type A atresia were managed during the study period. There were 9 males and 6 females. The clinical features and demographics are shown in Table 1. The mean gestational age was 35.5 weeks (range, 27-39 weeks), and the mean birth weight was 2179 g (range, 670-3520 g). Associated anomalies were diagnosed in 50% of the infants (8/15). Six babies had multiple anomalies associated with the VACTERL syndrome, whereas 2 had neurologic abnormalities. Thirteen neonates underwent an initial gastrostomy tube insertion followed by a delayed definitive repair. In this group, 9 patients underwent an end-to-end esophageal anastomosis, with 2 aided by circular myotomies of the proximal pouch. The mean duration to definitive repair in this group was 83 days (range, 63-90 days). The average duration of hospital stay in this group was 4 months (range, 3-4 months). Six (66%) of the 9 babies in this group had anastomotic leaks. Four healed spontaneously with conservative management, whereas 2 needed reexploration for complete dehiscence. Eight of 9 had documented reflux on upper endoscopy, and 4 underwent partial or complete fundoplication. Five patients had multiple dilatations for strictures. Two were documented to have Barrett dysplasia and remain on regular endoscopic surveillance protocols. All patients remained on proton pump inhibitors, and some were

placed on prokinetic agents for dysmotility of the lower esophagus. Eight of 9 children in this group were on full oral feeds, whereas one needed some supplemental gavage feeds to enable adequate energy intake. Average follow-up duration was 9 years (range, 1-17 years).

Two infants had a Collis gastroplasty with a simultaneous Dor fundoplication. One was performed at 107 days of life and the other at 186 days of life. Both were complicated by anastomotic leaks, with one patient undergoing a second thoracotomy for mediastinal debridement. This infant developed an esophageal stricture needing dilation but is now in his sixth year of follow-up with excellent oral intake. The outcome for the other infant was not as good. After repeated dilatations for an intractable stricture, he underwent a resection of that stricture; but with persistent dysphagia despite a new antireflux procedure, he underwent a cervical esophagostomy (performed at 6 years of age) and is currently awaiting a colon transposition. These children have been followed for 8 and 9 years, respectively.

Two patients underwent an antiperistaltic gastric tube placement 1 year after initial gastrostomy for a completely absent distal pouch. Both patients had a cervical esophagostomy created before stomach tube reconstruction. Both had problems with reflux and remain on proton pump inhibitors, despite antireflux surgeries. One was unable to tolerate full oral intake and receives supplemental jejunostomy feeds at night, whereas the other has some dysphagia to solid food but an adequate oral caloric intake. These patients were followed for 8 and 14 years, respectively.

Two babies underwent a Foker procedure. One baby had approximation of the 2 ends at 18 days of life. This, however, was complicated by an anastomotic leak that healed spontaneously. His subsequent course was complicated by multiple respiratory tract infections and severe tracheobronchomalacia for which he underwent an aortopexy, esophageal diverticulum excision (that has recurred despite resection), and severe reflux not corrected by antireflux surgery for which he finally underwent a Bianchi esophago-gastric disconnection. This child is on partial oral intake with some supplemental jejunostomy feeds. This child has been followed for 4.5 years. The second baby who underwent the Foker procedure lost the traction sutures placed on the esophagus during postoperative elongation and underwent a delayed primary anastomosis 59 days after initial thoracotomy. After 4 years of follow-up, this child continues to have alkaline reflux on pH probe studies and was transferred to a peripheral center for logistical reasons. She had full oral intake until her last follow-up appointment.

Patients who had a delayed primary anastomosis, with conservation of the native esophagus, had a better probability of being able to obtain their entire energy requirement orally without nutritional supplementation as compared with infants who had esophageal replacement or attempts at early primary anastomosis ($P = .04$). However,

there was no statistical difference in the time to final reconstruction between the group that underwent delayed primary anastomosis as compared with the other surgical strategies ($P = .064$).

3. Discussion

Many surgical strategies have been reported for the management of infants with type A esophageal atresia [3-8]. In our experience, the conservation of the native esophagus has worked well in offering a long-term solution to a potentially devastating malformation. We strongly believe in a conservative approach, waiting 3 months to allow for the greater differential growth of the esophagus as compared with the thoracic vertebral column, to facilitate an anastomosis between the 2 ends [9]. Others have suggested that this growth is secondary to the swallowing reflex in the upper pouch and gastroesophageal reflux (GER) into the lower pouch [3]. In our series, babies who were able to successfully undergo a delayed primary anastomosis were most likely to eat normally while having equivalent hospital stays compared with infants managed by other strategies. Recent studies have shown that the period of waiting, with upper pouch suction before definitive surgery, can be carried out at home [10]. This was not practical in our setting because many of the babies were transferred from remote regions. Furthermore, we believe strongly in the safety of frequent irrigation and continuous aspiration of the upper pouch, albeit on the regular surgical floor, to prevent aspiration pneumonia in these tenuous infants. Our leak rate of 66% is higher than the 37.5% to 45% rate quoted in the literature [2,11]. However, the distance between the proximal and distal pouches in all of these cases, as documented by contrast studies, was greater than 6 cm at the time of diagnosis; almost all anastomoses were performed under some tension. Only 2 infants needed reintervention for anastomotic dehiscence, with the others having small leaks that healed spontaneously. Although 2 patients underwent circular myotomies in this series, we have moved away from this technique because of reported problems with diverticula formation [12].

Gastroesophageal reflux remains a problem in these patients. Indeed, 4 patients required antireflux operations, whereas 2 of 8 patients with symptomatic GER developed Barrett dysplasia. The cause of GER is likely multifactorial in these cases. We have noticed motility problems in the distal esophagus, and excessive traction on the distal pouch to facilitate the anastomosis likely disrupts the intraabdominal antireflux mechanism. Four patients have needed multiple dilatations. Gastroesophageal reflux remains a leading cause for morbidity in these patients, and long-term follow-up is very important to detect potential complications owing to reflux. Our multidisciplinary tracheoesophageal fistula clinic visits transition into fol-

low-up by adult gastroenterologists as the patient reaches 18 years of age.

Conserving the native esophagus in ultralong gap esophageal atresias is not usually feasible, and a "conduit" is required to reestablish alimentary tract continuity. There continues to be an active debate in pediatric surgical circles as to the optimal conduit to replace the esophagus [13-15]. We believe that the stomach is a superior conduit because of its innate acid resistance, its ability to retain a tubular shape without dilatation, and its ability to bridge long gaps because of an excellent and reliable blood supply [16]. Whereas the Collis gastropasty was performed around the fourth month of life, the gastric tubes with cervical anastomosis were performed at 1 year of life. There are reports of excellent results in early neoesophageal reconstruction using the stomach either as a gastric pull-up or a gastric tube [17,18]. Our strategy, however, revolves around conserving the native esophagus wherever possible; and hence, we prefer to reserve esophageal replacement using the stomach only if all other attempts have failed. Proponents of the early reconstruction technique claim the advantages of lower costs of hospitalization, a lower incidence of aspiration pneumonia, and the early restoration of oropharyngeal feeding to decrease the risk of oral aversion. However, this is often achieved at the expense of an intrathoracic anastomosis and a high incidence of GER. We prefer using a cervical esophagostomy at the second exploration (around the third month) and to allow for sham feeding while the infant grows.

Microgastria is a frequent finding in this population of patients. Gavage feedings into the stomach during the period of waiting allow for its enlargement and facilitate the construction of a long gastric tube that reaches easily into the neck. Both of the infants who underwent this procedure are capable of oral intake, with one needing some supplemental jejunostomy feeds. Reflux has been a problem in 1 of our 2 patients. Gastric tube replacement has been associated with Barrett dysplasia in the proximal retained esophagus [19]. The 2 patients who underwent Collis gastropasty with intrathoracic anastomosis (isoperistaltic tube) have not fared as well. Severe reflux has led to strictures in both patients. Although one was successfully treated with multiple dilations, the other ended up with a cervical esophagostomy and is awaiting a colon transposition. This finding is consistent with recent reports on this subject. The isoperistaltic gastric tube is thought to secrete a greater amount of acid leading to a higher incidence of stricture and Barrett dysplasia [20].

We attempted the Foker technique, based on the principle of stimulating growth of the pouches by longitudinal traction, twice during the study period and were unable to replicate the success reported by the authors [7]. One patient had lost her traction sutures during the elongation process and underwent a delayed anastomosis with a good outcome. This is one of the drawbacks with the Foker technique and has been described by other authors as well [21]. The other

patient developed an esophageal diverticulum and problems with repeated aspiration pneumonia. This infant ultimately underwent a Bianchi procedure with esophagogastric disconnection [22]. Although this patient is tolerating a major portion of his energy intake orally, he still needs some supplemental gastrostomy feeds. We have moved away from this technique because of our experience.

In this series, the native esophagus was conserved in 10 (67%) of 15 patients. This compares favorably to other recent reports [23]. In our experience, infants whose native esophagus is conserved have better outcomes than those with replacement conduits such as colon or stomach. The ultimate success of repair of pure esophageal atresia rests in the ability of the infant to sustain growth and development on his/her own through oral feeding. All except one of the children in our study are capable of doing this. Long-term follow-up is imperative to detect and treat potential complications.

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