

Esophageal Replacement for Long-Gap Esophageal Atresia in a Resource-Limited Setting

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Summary

The management of esophageal atresia in a resource-limited environment is plagued with challenges that often lead to poor outcome. The diagnosis and management of babies with long-gap esophageal atresia adds a new dimension to these challenges. We report the success of esophageal replacement surgery for a baby with long-gap esophageal

atresia in a resource-limited setting. One year after surgery, she was eating well and her weight gain was appropriate.

Keywords: Long-gap, Esophageal Atresia, Esophagoplasty, Resource-Limited

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Introduction

Management of esophageal atresia (EA) in a resource-limited environment is a major challenge to the pediatric surgeon. Esophageal atresia occurs in various forms in approximately 1 in every 3000 – 4500 live births (1). Although almost 100 variants of this anomaly have been described, EA with distal tracheo-esophageal fistula (TEF) is the commonest, occurring in approximately 86% of cases. Pure esophageal atresia (isolated EA with no fistula) occurs in approximately 8% of cases (1). Survival of babies with EA is firmly hinged on effective pre- and post-operative management. Many pediatric surgical units in sub-Saharan Africa have reported survival of 10 - 40% (2).

In long-gap esophageal atresia (LGEA), the interval between the upper pouch and the distal esophagus does not permit primary anastomosis at surgery. Fortunately, this is rarely encountered in EA with distal TEF. However, it is the norm in pure EA (3).

The diagnosis of pure EA is confirmed by the inability to pass a naso-gastric tube into the stomach and a gasless abdomen on plain x-ray (Figure 1). The surgical management of babies with this variant is equally challenging and controversial (3). The majority of pediatric surgeons consider delayed primary anastomosis of the native esophagus the optimum approach, a strategy that demands meticulous nursing care, physiotherapy and proper nutrition by supervised gastrostomy feeding (4).

Whereas this care can easily be provided in the developed world, in Sub-Saharan Africa, it is still a challenge. But even in the established pediatric surgical centers, the desire to preserve the baby's native esophagus must be carefully weighed against humility to know when to consider a replacement procedure (3).

To replace the esophagus of a baby is a major decision in many pediatric surgical units world over. Fortunately, the need for this procedure has decreased in the recent years due to the increase in the variety of techniques of lengthening the upper esophageal pouch of babies with LGEA, that have enabled successful delayed primary anastomosis (5). Despite the procedure being adopted for use in children in 1955, its practice in the sub-Saharan Africa has hardly been reported.

We report the success of esophageal replacement for a baby with LGEA in a resource-limited setting.

Case Report

Baby EB, a female neonate was born at a preterm age of 35 weeks gestation. She was the first twin to a 35 year old Para 6+0 mother. The baby was born at Nyanza Provincial General Hospital, Kisumu, via an emergency Caesarian Section due to breech presentation. Her birth weight was 1665g. She was referred to the pediatric surgery unit of Moi Teaching and Referral Hospital on her 6th day of life with a history of frothing in her mouth, excessive drooling

of saliva and regurgitation of feeds. Ante-natal history revealed that the mother had polyhydramnios.

On examination, she was in respiratory distress, premature, with moderate pallor. Her head circumference, spine and limbs were normal. Her respiratory rate was 40 per minute and the axillary temperature was 38°C. Her chest was normal in shape and expansion except for intercostal recession. She had bilateral coarse crepitations. Her cardiovascular and abdominal examinations were normal. A stiff nasogastric tube (Fr 8) could not be passed into the stomach.

The baby was admitted to the newborn unit and was kept warm in an incubator. The initial management included half-hourly nasogastric tube suction, broad-spectrum intravenous antimicrobials, intravenous ranitidine and the baby was positioned on the left lateral position with head propped up at 30°.

Her hemogram revealed a moderate anemia with a hemoglobin level of 11.0g/dl. The babygram showed the nasogastric tube unable to pass beyond the level corresponding to C3 vertebra, right upper lobe lung opacity and a featureless (gasless) abdomen. There were no vertebral or long bone anomalies (Figure 1). The echocardiogram showed a small secundum atrio-septal defect and a left sided aortic arch. A diagnosis of pure EA (Isolated EA with no fistula) was confirmed and the baby was subsequently prepared for surgery. A cervical esophagostomy and a feeding gastrostomy using a Foley's Catheter Fr 12 were done under general anesthesia. Post-operative recovery was gradual and uneventful. This was done in the newborn ward due to lack of neonatal intensive care facility.

Feeding through the gastrostomy was commenced on the 3rd post-operative day and gradually increased to the optimal volumes after 10 days. The mother was carefully trained on care of the esophagostomy and sham oral feeding. She was discharged on day 18 of life weighing 1850g.

She was closely followed-up in the pediatric surgical out-patient clinic and at 1 month of age, her weight was 2800g and had a leaking gastrostomy. The Foley's catheter was changed and she started retaining feeds. At 2 months of age, her weight was 3510g and at 5 months 4500g.

She was re-admitted for definitive surgery at the age of 7 months by esophageal replacement with reversed gastric tube. This involved creating a tube from a portion of the greater curvature of the stomach whose blood supply is dependent on the left gastro-epiploic artery (Figure 2). The tube was reversed and placed in the posterior mediastinum and gastro-esophageal anastomosis was done in the neck. Pyloroplasty was then performed.

In the post-operative period the baby was managed in the general pediatric surgical ward due to lack of intensive care facility. Enteral feeding was recommenced via the trans-anastomotic naso-gastric tube on the 3rd post-operative day. Her recovery was gradual except for the minor anastomotic leak that was managed non-operatively and healed within 10 days. The patient was discharged on the 14th post-operative day for close follow up in the pediatric surgical out-patient clinic. She was eating well and her weight was 9.2 kg at 12 months of age.

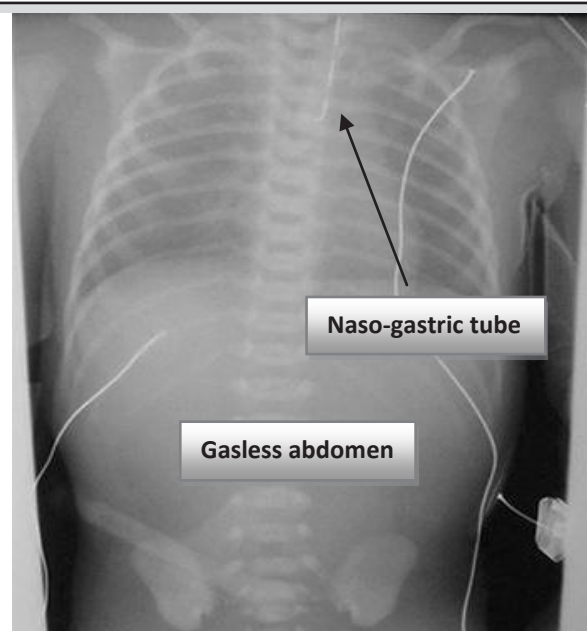


Figure 1: Plain radiograph of abdomen and chest

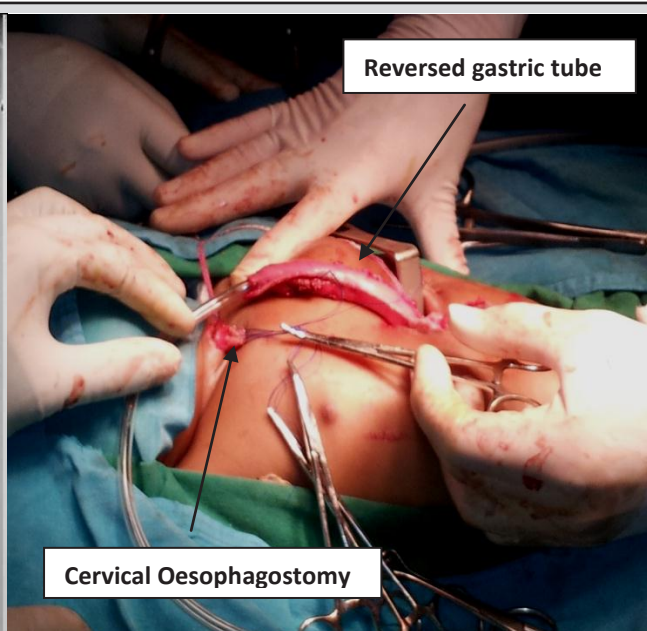


Figure 2: Photo of a fashioned reversed gastric tube

Ethical approval was obtained from the Moi Teaching and Referral Hospital Institutional Research and Ethics Committee (IREC) and the patient's father gave informed consent for the publication of the case report.

Discussion

The management of esophageal atresia (EA) in a resource-limited environment is plagued with challenges that include delay in diagnosis, lack of facilities and inadequate experience in many of these pediatric surgical units that often lead to poor outcome (6). The more commonly seen variant, the EA with a fistula between the distal esophagus and the trachea is often amenable to primary anastomosis with good long-term functional outcome. Significant determinant of survival is the effectiveness of pre- and post-operative management of these patients (7). Many budding pediatric surgical centers in Africa still face great challenges in managing this common variant of EA.

The long-gap esophageal atresia (LGEA) adds a new dimension to the existing problems as its management is still a major challenge worldwide. There is consensus among most pediatric surgeons that the preservation of the native esophagus is key in achieving better post-operative outcomes. This has led to the development of various techniques of lengthening the upper esophageal pouch that include bougienage, modified Scharli and Foker procedures, which enable delayed primary anastomosis (8,-11).

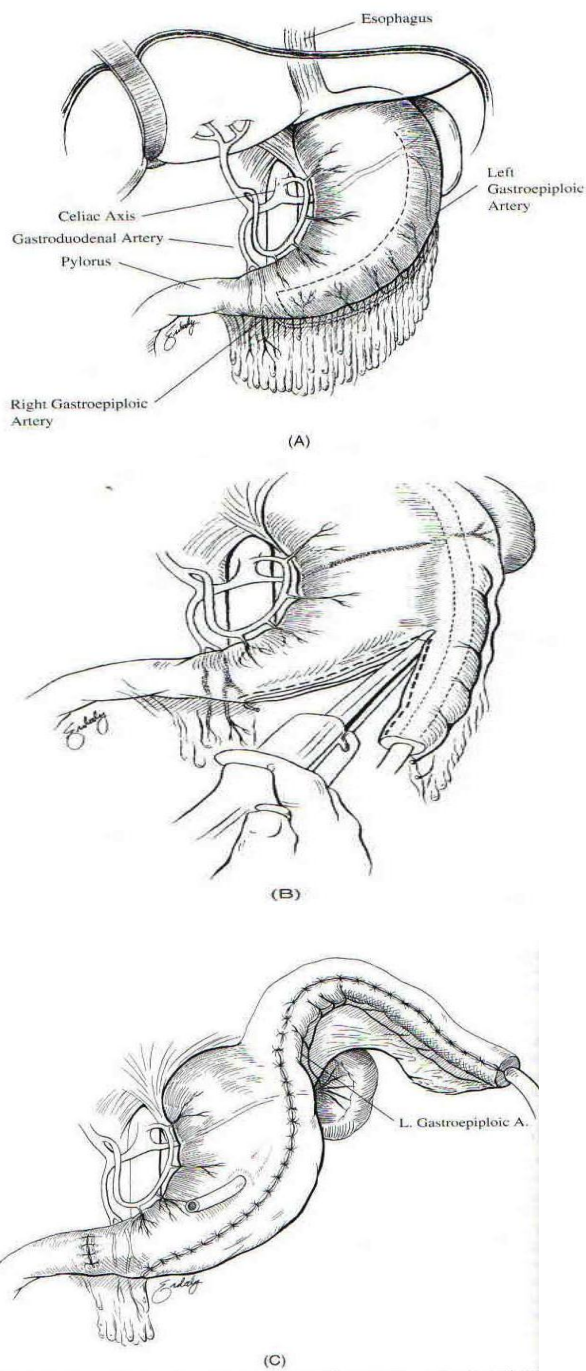
However, achieving adequate length to enable delayed primary anastomosis requires time to allow for esophageal growth, hence the proximal esophageal pouch must be managed adequately to minimize the morbidity and mortality that arises from aspiration of saliva. Continuous low-pressure suction through a Replogle tube inserted into the proximal esophageal pouch, which is the norm in many first world pediatric surgical centers, is hardly available in many centers in the third world (7). Parenteral nutrition, another basic requirement for the success of delayed primary esophageal anastomosis, is scarce. These factors make this option of managing LGEA untenable in resource-limited settings.

Esophageal replacement was traditionally indicated in failed lengthening techniques, unfavorable native

esophagus with stricture or significant dysmotility post-repair. An ideal esophageal substitute in an infant should provide efficient conduit from the mouth to the stomach with minimal gastric reflux. It should not impair respiratory or cardiac function and the operative technique should be non-challenging. The conduit must grow with the child to adulthood. In reality, none of the available esophageal conduits fulfill the set criteria (5). The stomach has been used either as gastric tube esophagoplasty or gastric interposition. Colon interposition is well described particularly in adult patients. The jejunum has also been used as a pedicled interposition or a free graft.

Gastric transposition, first described by Lewis Spitz in 192 cases, involves a laparotomy and a left-sided cervical incision incorporating the cervical esophagostomy. A left-sided thoracotomy is done if difficulty is experienced in the blunt dissection of the posterior mediastinum. The stomach is mobilized transhiatally and proximal gastro-esophageal anastomosis is done. Pyloroplasty is then performed to improve gastric emptying. Spitz reported a mortality rate of 9% and significant morbidity that made the procedure unpopular (12).

Gastric tube esophagoplasty, which was successful on our patient, involves a laparotomy, closure of the gastrostomy and mobilization of the stomach to the gastro-esophageal junction. The esophageal stump is resected and the duodenum is extensively mobilized via Kocher maneuver. A gastric tube is created along the greater curvature of the stomach that is pedicled on the left gastro-epiploic artery, as illustrated by the diagrams in Figure 3 (5). The cervical esophagostomy is then dissected. Transhiatal blunt dissection of the posterior mediastinum through the thoracic inlet into the neck is done. The gastric tube is reversed through this route. Gastro-esophageal anastomosis and pyloroplasty are then performed (5). The advantages of this procedure include the relative ease with which adequate length and appropriate size of the conduit is achieved. Moreover, it is pedicled on a dependable blood supply and allows rapid transit of food boluses. However, its drawbacks include the very long suture line that leads to the high incidence of anastomotic leak and strictures. It is also prone to gastro-esophageal reflux and Barret's metaplasia (13,14).



Anatomic considerations for a reversed gastric tube based on the fundus. A distal incision is made 2 cm proximal to the pylorus at the point of the division of the right gastro-epiploic artery

The stomach is opened along the greater curvature at the site of the eventual proximal end of the gastric tube. A 20 to 24 Fr chest tube is inserted and passed along greater curvature to the fundus to help define the circumference of the gastric tube. The tube is then created using multiple firings of a GIA stapler along the superior border of the chest tube.

The completed reversed gastric tube, gastrostomy, and pyloroplasty.

Figure 3: Gastric tube interposition Images used with permission from Textbook of Operative Pediatric Surgery (5).

Figure 3. Gastric tube interposition Images used with permission from Textbook of Operative Pediatric Surgery

Colon interposition involves a laparotomy, mobilization of the left colon with the preservation of the marginal and the left paracolic artery. The posterior mediastinal route is then developed by blunt dissection. The esophagostomy is then taken down. Esophago-colic anastomosis and gastro-colic anastomosis are performed. The procedure is completed with a 2700 ante-reflux wrap at the gastro-colic anastomosis (15). The advantages of colon interposition include the ease with which adequate

length of the conduit is achieved and gastric reflux is rare. However, its major setback is the redundant growth of the conduit over time that leads to slow transit of food boluses.

Jejunal interposition and free jejunal grafts are used when the other approaches have failed or when neither the stomach nor the colon is available for esophageal reconstruction (16).

In conclusion, although we certainly subscribe to the principle that the baby's own esophagus is best

and that the native esophagus can be preserved in a majority of cases, esophageal replacement offers pragmatic solutions to the challenges faced by surgeons managing LGEA in a resource-limited environment. This approach allows for cervical esophagostomy and feeding gastrostomy be performed early in the neonatal period which enable survival and growth of the baby. Reversed gastric tube esophagoplasty can then be performed once the baby is fully nutritionally optimized.

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