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Jejunum for bridging long-gap esophageal atresia

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KEYWORDS
Esophageal atresia;
Long-gap esophageal atresia;
Esophageal replacement;
Jejunum

OBJECTIVE: Exploring pros and cons of bridging long-gap esophageal atresia with an orthotopic jejunal pedicle graft. Retrospective series of 19 patients.
METHODS: From 1988 through 2005, 19 patients with long-gap esophageal atresia received a jejunal graft. Median age at reconstruction was 76 days. The technique involved an initial right-sided thoracotomy or thoracoscopy to confirm the diagnosis of long-gap esophageal atresia. Through a median laparotomy, a small pediculated jejunal graft was prepared and placed transmesocolically and transhiatally in an orthotopic position in the right chest.
RESULTS: All patients survived and none of the grafts were lost. Four intrathoracic and one intraabdominal leak occurred. One intrathoracic and one intraabdominal leak were surgically repaired. One early distal stenosis was reoperated as well. There were always signs of distal functional subobstruction, responding to dilation in all but one patient. Gastroesophageal reflux was not a problem except for one patient whose distal esophagus was eventually resected because of ongoing distal obstruction with dilation of the graft. Except for one patient, all patients are eating normally and most of them grow well. Respiratory problems were rare. Grafts did not become redundant and retained peristaltic activity.
CONCLUSION: Orthotopic jejunal pedicle graft reconstruction of the esophagus in children is a demanding operation with considerably early morbidity but good long-term results. It should be part of the pediatric surgical armamentarium for reconstruction of the esophagus.

Long-gap esophageal atresia for me is an esophageal atresia without distal fistula. I realize that there may be occasions when the distance between the upper pouch and the distal fistula is just too long for making an anastomosis, but in my experience, this is very rare. As the esophageal body in esophageal atresia without distal fistula is largely absent, and as the morbidity related to delayed primary anastomosis in this group of patients is considerable, I opted in 1988 for bridging the esophageal gap with an orthotopic jejunal pedicle graft.[1] The purpose of this publication is to discuss the advantages and disadvantages of this procedure on the basis of my own experience.

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Patients and methods

From 1988 through 2005, 19 children received an orthotopic jejunal pedicle graft reconstruction of the esophagus in the context of esophageal atresia.[2] All but six reconstructions were done at the Wilhelmina Children's Hospital in Utrecht. Eighteen children had no distal fistula, but 6 of them had a proximal fistula. Only 1 had a delayed primary anastomosis, performed in another center. This child underwent esophageal replacement at 3 years of age because of poor function of the esophagus. The only child with esophageal atresia and distal fistula developed a recurrent fistula

and long stenosis after the primary repair. Four of the 19 children had trisomy 21, and 1 had the CHARGE association. Two more children were mentally retarded, 1 due to cerebral hemorrhage complicated by hydrocephalus, 1 on an idiopathic basis. One child had a duodenal atresia and another child a rectovestibular fistula. There were no other life-threatening associated anomalies. Only 1 child had a cervical esophagostomy. Median age at replacement was 76 days (range 16-1080).

Technique

The technique has been described in detail before but is briefly repeated here.[2]

In the waiting period between birth and the jejunal interposition, the upper esophagus is emptied by means of Replogle catheter suction. A gastrostomy is created for feeding. As the incidence of proximal fistula in patients without distal fistula is high, a proximal fistula is excluded early.[3] It is also wise to exclude intestinal malrotation, which has a higher incidence in esophageal atresia.[4,5] In the event of malrotation, the vascular supply of the jejunum may preclude its use for pedicle graft interposition, as happened in one of our patients. In this patient, not included in this series, a gastric pull-up was performed.

Timing of the jejunal interposition depends on how problematic the conservative approach is. In the event of repetitive lung problems, the interposition procedure is not postponed; otherwise, the jejunal interposition is performed at 8 to 12 weeks. The day before surgery, antegrade whole-bowel lavage is performed to leave all options open. In a patient not included in this series, a pedicle ileal graft was used.[6]

If a cervical esophagostomy had been created, it is taken down first.

I prefer a right-sided posterolateral thoracotomy, which avoids interference with the aortic arch and allows for a more straight posterior mediastinal position of the graft. A classic but large posterolateral thoracotomy is performed through the bed of the 6th or 7th rib. After verification of the diagnosis of long-gap esophageal atresia, the skin is closed with a running nylon suture. Verification in the last patient was done thoracoscopically.

Next, the patient is placed in a supine position and a midline laparotomy is performed. The fundus is detached from the diaphragm, and the upper short gastric vessels are severed. The left crus is freed and the posterior hiatus is opened. Access is gained behind the distal esophageal pouch into the right pleural cavity. The tunnel from the abdomen into the right chest is dilated with Hegars.

The creation of the pedicle graft is the most critical part of the operation. The first two or three mesenterial vessels are divided between ligatures close to the main mesenteric route (Figure 1). The jejunum is transected close to Treitz ligament, leaving enough proximal jejunal length for restoration of continuity. The jejunum is severed again opposite the level of the third mesenteric branch. Only jejunum is taken, not the arcade. The jejunum thus isolated is far too long. No more than maybe the 5 proximal centimeters are needed. The rest therefore is removed, starting at the distal end of the graft. Bowel continuity is restored behind the pedicle of the graft. The graft is passed through the left mesocolon, behind the remaining short gastric vessels, and through the posterior hiatus behind the esophageal remnant into the right chest.

It is best to close the abdomen temporarily, to reposition the child and to quickly access the right chest to check the graft. The distal esophageal pouch is opened longitudinally in the posterolateral area. One should make sure that the distal part is widely patent. In one of our patients, the distal segment was stenotic over about 2 cm, which went unnoticed, however, until the graft had been trimmed, resulting in an anastomosis under tension and postoperative leak developed. Next, the upper pouch is opened.

An anastomosis is made between the upper and lower esophagus and the graft. Before finishing the anastomosis, a nasogastric tube is passed through the graft into the stomach. A chest drain is inserted and the thoracotomy closed in layers.

Finally, a gastrostomy is recreated and the abdomen closed.

The patient is weaned from the ventilator when appropriate. Gastrostomy feedings are started when there are no gastric retentions. The drain is removed after 5 to 6 days, provided a contrast study shows that there are no leaks. Oral feeding is then started. It is important to monitor the patient for stenosis. In case of distal stenosis, the graft will dilate, which must be avoided at all times.

Results

General

None of the patients died and none of the grafts were lost.

Specific results

Early results

Gaining enough jejunal length was not a problem, even not in the patient with an esophagostomy.

The median duration of endotracheal intubation was 5 days (range 1-43). One child was excluded from this analysis because of iatrogenic stenosis of the trachea and long-term intubation as a result. Three patients developed ARDS, two in connection with leakage.

Five leaks occurred, four in the chest, and one in the abdomen. The leak in the abdomen as well as one leak in the chest were treated surgically. In one patient, a proximal fistula was missed and this required ligation through the neck in a second operative session. One patient developed early distal graft anastomotic occlusion requiring surgical correction. Dilation in this patient could not be performed as the nasogastric tube had been removed prematurely and no lumen could be seen at esophagoscopy.

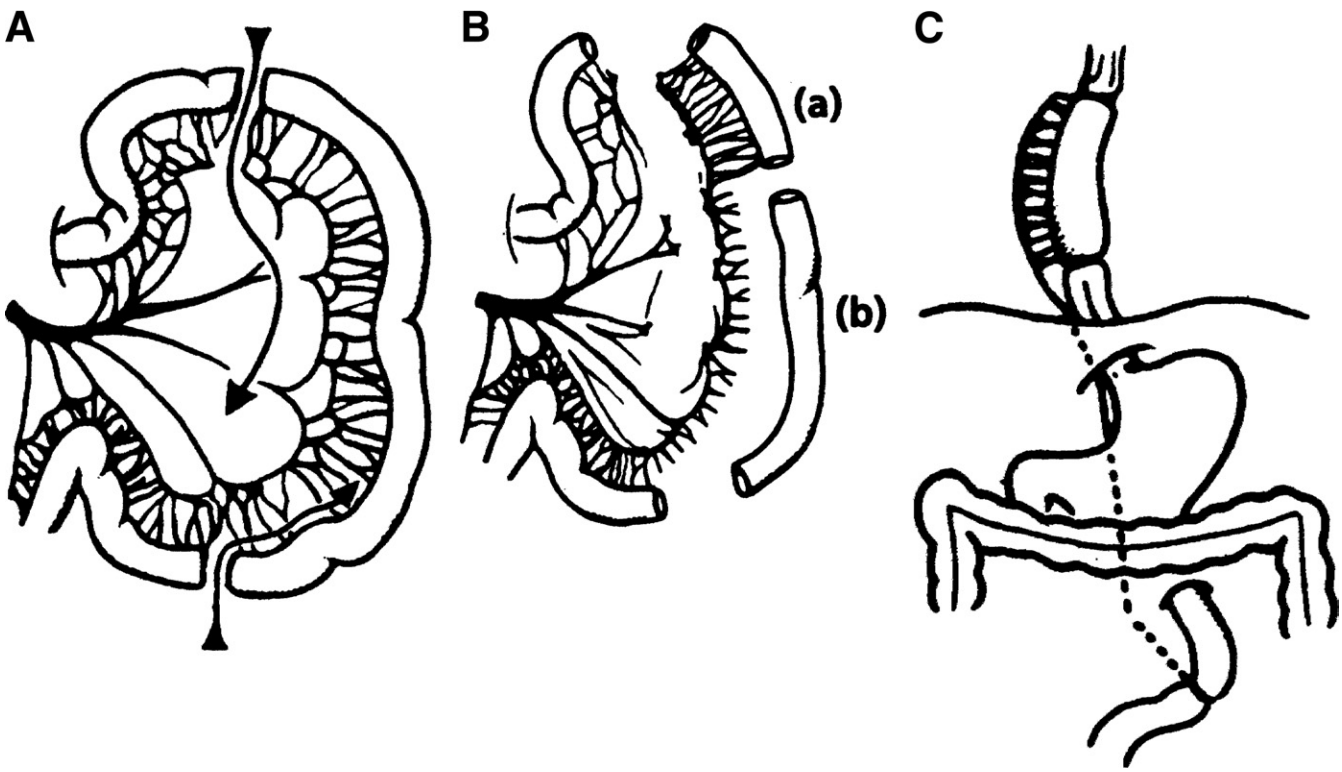

Figure 1 Schematic representation of the technique of orthotopic jejunal pedicle graft reconstruction of the esophagus (Reproduced with permission, J Pediatr Surg Int 1994). (A) The jejunum is transected close to Treitz ligament. The first two mesenteric artery branches are centrally divided between ligatures, leaving the peripheral arcades intact. The jejunum is transected again at the level of the third mesenteric artery branch. The distal part of the upper jejunum is skeletonized close to the bowel wall. (B) The distal part of the upper jejunum (b) has been removed, leaving the uppermost part (a) for transfer into the chest. (C) The uppermost part of the jejunum has been transferred through the left mesocolon, behind the stomach, and through the posterior part of the hiatus into the right chest, where a double anastomosis has been made.

Late results

The median follow-up period was 5.5 years (range 1-17.5).

Ten of the 19 patients required dilation, which had to be repeated 5 times or more in 6 of the 10 patients. Sometimes both anastomoses in the chest had to be dilated, but in the long run, the distal anastomosis was more problematic. On one occasion, the conduit was perforated, which necessitated a second thoracotomy and closure of the perforation. In one patient, functional obstruction of the distal anastomosis caused marked dilation of the conduit. A widening of the anastomosis (plasty) was unsuccessful. Finally, the distal esophagus was resected and the dilated conduit was anastomosed to the stomach. This led to significant gastric reflux and pulmonary problems. The other patients showed no signs of reflux.

One patient with trisomy 21 developed a functional but not anatomical short-bowel syndrome and stayed on gastrostomy feedings in an institution until he died from pulmonary problems at the age of 10 years. All other patients acquired normal eating habits and most patients grow well. Bolus obstruction was rare, and so were repeat respiratory problems.

With time, the graft did not tend to elongate, but in some patients it widened slightly as a result of functional obstruction at the distal anastomosis. In all patients, the transplant showed vigorous contractions at follow-up (Figures 2 and 3).

Discussion

Affirming that the patient's own esophagus is best would seem too dogmatic.[7,8] After all, a critical look reveals that the long-term results regarding the esophagus after esophageal atresia repair are not very good. This contrasts with quality of life assessments.[9-12] The explanation for this may well be that the patient does not know what to expect. In esophageal atresia without distal fistula, the esophagus is largely absent. Of course, the ends can be brought together, depending on traction and the amount of mobilization. Still, the crux of the matter is restoring function. The likelihood of stricture and severe gastroesophageal reflux is as high as 30%.[13] Reflux in these patients is very difficult to treat. Moreover, the extensive mobilization of the esophagus even further impairs esophageal function.

Whether true esophageal lengthening by traction can be achieved, as is claimed for the Foker procedure, remains to be proven.[14] In the patient who had a rather uncomplicated delayed primary repair but was reoperated at the age of 3

Figure 2 Barium meal 3 months after jejunal grafting. (A, B) Anteroposterior view, showing vigorous contractions. (C) Lateral view. (Reprinted with permission.[2])

years because of poor esophageal function, the esophagus at surgery looked like a guitar string. After transection of esophagus at the site of the previous anastomosis, the ends retracted leaving a considerable gap to bridge. I realize this is just a case observation, but even after 3 years the esophagus was under tension.

When bridging esophageal defects, there is a discrepancy between the difficulty of the operation and the long-term results to achieve. Gastric transposition is certainly one of the easier procedures, followed by colonic interposition and finally by jejunal graft interposition. Free jejunal graft interposition with microvascular anastomoses would be the most demanding one.

The vast majority of adult patients show good swallowing function after free jejunal graft following cervical pharyngoesophagectomy for cancer.[15,16] Swallowing function was also good in the 10-year-old child who had such an operation.[17] This is certainly related to the fact that jejunum retains peristaltic activity. Preservation of peristaltic activity has been described for free jejunal grafts[18] as well as for pedicle grafts.[19] We also have noted good peristaltic activity in all our jejunal pedicle grafts.

In most studies of children receiving a jejunal pedicle graft esophageal replacement, the distal part of the graft was anastomosed directly to the stomach.[19,20] Cusick and coworkers put the graft in a retrosternal position and added a dual blood supply by anastomosing the terminal arcade vessels to vessels in the neck.[20] The so-called supercharge technique for jejunal or colonic pedicle grafts in adults is gaining popularity,[21] but its feasibility in small children remains questionable.

One of the reasons for not using the distal esophageal remnant has been the observation that the distal esophageal remnant may contain abnormal embryonic tissues such as cartilage.[22,23] Up until now, we have preserved the distal esophagus with the idea to prevent reflux. To compensate for a caliber discrepancy distally, the distal esophagus was opened longitudinally, thus allowing for an end-to-side anastomosis with the distal part of the jejunal graft. In most of the patients, functional distal obstruction was noted after the jejunal interposition, which often required dilation. In one patient, we eventually resected the distal esophagus, but this created a common channel of dilated jejunum and stomach with reflux and respiratory symptoms. In view of the functional obstruction seen to some degree in all patients, removal of most of the distal esophagus may be better. In two patients not included in this study, the distal esophagus was replaced with a pedicle graft jejunum because of a long peptic stricture. In these patients, the graft was anastomosed directly to the stomach. No signs of distal functional obstruction were noted. It seems that absence of dilation and retainment of good peristaltic activity are the reasons why no pathological reflux was observed.

Gaining enough jejunal length was not a problem. In long-gap esophageal atresia, it is the middle part of the esophagus that is missing, usually not the proximal part. If the more proximal esophagus has to be replaced, getting enough jejunal length may be a problem. Under those circumstances, a pedicle ileal graft may be better.[6]

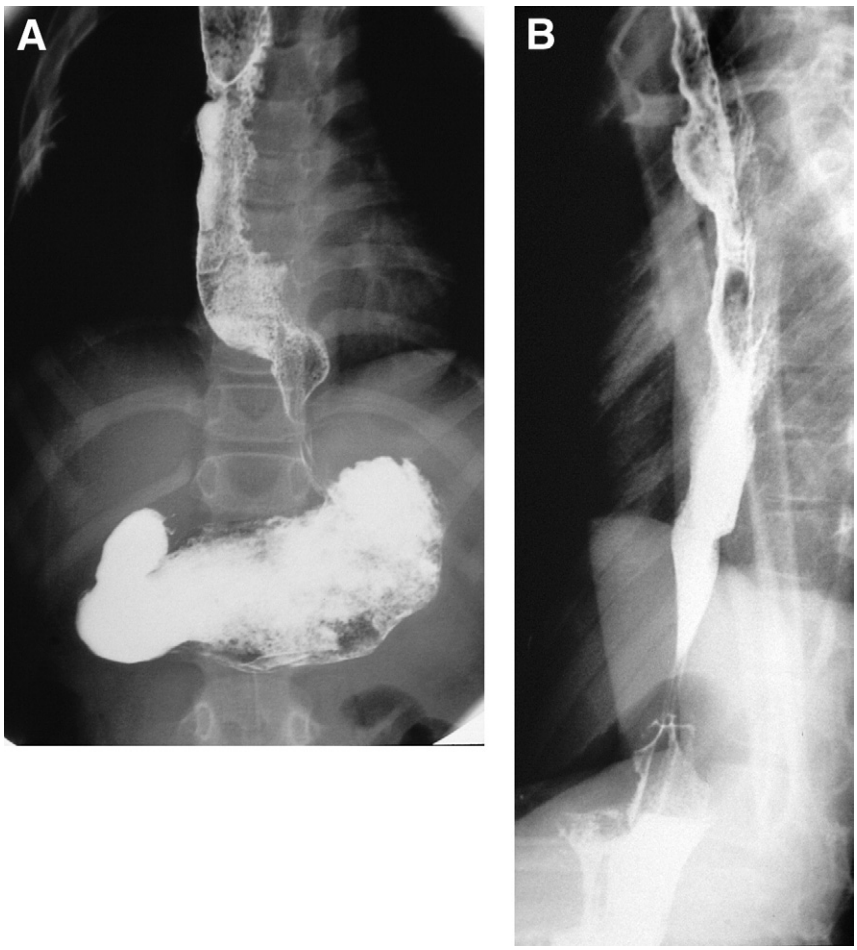

Figure 3 Barium meal in another patient, 7 years after jejunal grafting. (A) Anteroposterior view. (B) Lateral view. (Reprinted with permission.[2])

Esophageal replacement with jejunum in children is a demanding operation with considerable morbidity, but in this series, there was no mortality and none of the grafts were lost. The technique of jejunal pedicle grafting should be part of the pediatric surgical armamentarium for esophagus reconstruction.

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