Definitive management of isolated esophageal atresia: Experience at NICH Karachi

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ABSTRACT

Background: Definitive treatment of isolated esophageal atresia (IEA) is still controversial. A study was conducted to review cases of IEA in our department with a view to evaluate the long term results of definitive surgery in these patients.

Materials and Methods: Nine consecutive patients with IEA were included in the study. All babies initially underwent cervical esophagostomy and feeding gastrostomy. One baby also had anorectal atresia and needed a colostomy. In 6 babies, end esophagostomy was performed while in 3 babies, lateral esophagostomy was performed. Of the 6 babies with end esophagostomy, two underwent jejunal Interpositioning, two had serial extra-thoracic lengthening and two reverse gastric tube interpositioning. Three babies with lateral esophagostomy were planned for delayed primary repair.

Results: Both babies with jejunal interpositioning initially did well; serious gastro-esophageal reflux occurred in one baby who needed antireflux surgery. At 6 years follow-up both children had redundant Jejunum. Recurrent Respiratory tract infection remained a problem in one child while other is doing well. Of the two babies with extrathoracic lengthening it was possible to perform end to end anastomosis in one baby but in the other extrathoracic lengthening did not work and gastric transpositioning was performed. Of the two babies with reverse gastric tube interpositioning one developed esophageal stenosis at the site of anastomosis and is on serial dilatations. Other died due to anastomotic leak and mediastinitis. In all the three babies with lateral esophagostomy spontaneous lengthening of esophagus was noted. End to end anastomosis was possible in two of these babies who are doing well. One is waiting definitive surgery. Number of complications, hospital stay and cost of surgery was least in patients with lateral esophagostomy.

Conclusion: Various methods of definitive treatment have been proposed for isolated esophageal atresia but each having its own complications. Treatment therefore has to be individualized. Lateral esophagostomy with spontaneous esophageal growth may be a good option in developing countries with limited facilities.

KEY WORDS: Esophageal atresia, jejunal interpositioning, esophageal substitution, lateral esophagostomy.

INTRODUCTION

Definitive treatment of isolated esophageal atresia is still a challenge for the treating surgeons. Due to wide gap between the two esophageal segments primary end to end anastomosis is often not possible. Retained secretions in upper pouch is a major nursing issue in these babies thus cervical esophagostomy and feeding gastrostomy is a preferred procedure for initial management of these cases. Esophageal continuity may then be restored by esophageal substitution, extra-thoracic lengthening or by serial stretching of stoma by soft bougies.[1] In some centers, primary gastric transpositioning is performed with satisfactory results. Lateral esophagostomy has recently gained popularity due to simplicity of the procedure. In these cases, spontaneous lengthening of esophagus is expected and a delayed primary repair is often possible.

The danger of aspiration of secretions is avoided, patient may be nursed at home and surgery is planned in optimal conditions. This procedure is especially useful for countries with limited resources as cost of repeated admissions is avoided.

MATERIALS AND METHODS

Data of patients with IEA was analyzed in our department with a view to evaluate various management options in these patients. 9 patients having IEA were included in the study. There was no exclusion criterion. Cervical esophagostomy and feeding gastrostomy was performed in all babies however the type of cervical esophagostomy was individualized according to the definitive management plan [Table 1]. 6 babies had end oesophagostomy and 3 had lateral esophagostomy. Of the
6 babies who had end esophagostomy 2 had extra-thoracic lengthening procedure and 4 were planned for esophageal substitution. Of these two had Jejunal Interpositioning and two had reverse gastric tube interpositioning. Extrathoracic lengthening was performed at 2-4 weeks interval under general anesthesia; Stoma was mobilized and brought down serially at a lower level usually 2-3 cm down the previous stoma site. Jejunal interpositioning was performed using long jejunal isoperistaltic loops through retrosternal route. Vascularity of the both ends was ensured and an end to side anastomosis performed proximally with esophagus and distally with the stomach. Reverse gastric tubes were constructed using anti-peristaltic loops from the greater curvature of the stomach with vascular pedicle based on left gastro-epiploic vessels. Gastric transposition was performed by mobilizing the duodenal loop with Mikulics pyloroplasty. A transthoracic repair was then performed. In patients with lateral esophagostomy initial stoma was made in lower neck. During definitive surgery, stoma was closed and end to end thoracic repair of esophagus was performed.

RESULTS [FIGURES 1-3]

Table 2 depicts the complications encountered. The first two babies had jejunal interpositioning, both initially did well. After few years one child developed severe GER and needed Fundoplication. Same child came back at the age of 6 years with recurrent chest infections due to stasis and aspirations. There was no reflux on barium swallow. He was treated conservatively with some improvement and is planned for reduction Jejunoplasty if symptoms persist. The other child did well after surgery but also had few episodes of chest infections. Of the two patients with reverse gastric tube one developed esophageal stricture and is on repeated dilatations. The other patient had uneventful surgery but developed anastomotic leak and died due to severe mediastinitis. Small stomach and gastrostomy site were problems in both cases. Of the two patients with extra thoracic lengthening, in one child good lengthening was achieved and it was possible to perform an end to end anastomosis. The child needed dilatations twice for mild dysphagia but is doing well. He also has craniotubal synostosis. The other child developed repeated strictures at stoma site after extra thoracic lengthening and we failed to achieve adequate length even after 9 months of repeated lengthening procedures. This child later had gastric pull-up as stomach was too small for reverse gastric tube. Postoperative course remained uneventful and baby remained symptoms free. He also needed few dilatations for dysphagia for solids.

First Lateral esophagostomy was incidental by the resident. Contrast studies after few months in this child showed that there was significant growth of the esophagus. Later we performed it on two more cases. In two children good length of esophagus was achieved and end to end anastomosis was performed. One of these babies needed esophageal dilatations but is otherwise well. One is waiting for definitive surgery and contrast studies have shown good esophageal length after few months.

DISCUSSION

Definitive treatment of isolated esophageal atresia remains a challenge to the treating surgeons. There is usually a long gap between the two ends and primary end to end anastomosis is not possible. Various techniques to achieve esophageal continuity have been used with successes and failures but none of the technique has been...
adapted universally due to unpredictable results. This suggests the difficulty in standardizing the treatment of IEA. Successful primary repair of IEA has been reported as early as 1967 by Ibay RS Jr et al but in most babies the gap is often too long to achieve a primary repair.\textsuperscript{[2]} Delayed primary repair of esophagus has been reported to show good initial results but high incidence of gastroesophageal reflux and esophageal stricture.\textsuperscript{[3]} These complications are well explained by the fact that forced pulling of esophagus results in displacement of esophagus in the thorax increasing the incidence of reflux. Native esophagus is the preferred conduit and substitution is only performed when it is not possible to have a satisfactory end to end repair. Current management of IEA in most centers is to perform a cervical esophagostomy and feeding gastrostomy at birth and delayed repair of the esophagus is planned. To achieve adequate length for a tension free anastomosis various ingenious techniques have been described. Stretching of the esophagus remained the most popular technique for gaining esophageal length. It is achieved by stretching of esophageal segments by soft and firm dilators, Magnetic devices, nylon threads and metal olives. Extra thoracic lengthening was described by Kimura K et al.\textsuperscript{[4]} They showed high incidence of adhesions therefore the technique was modified by Dessanti et al.\textsuperscript{[5]} They reported a case where they successfully used Gortex mesh to prevent adhesions in multistage extra thoracic lengthening. The procedure was later adapted by others with good results.\textsuperscript{[6]} In cases where esophageal lengthening cannot be achieved by the above mentioned techniques esophageal substitution may be required. Colon or jejunum are the preferred conduits. The complication rates are high with these substitutes and these are now used in selected cases where other methods of delayed primary repair fail or are not possible.

Spitz described primary gastric transposition for IEA.\textsuperscript{[7]} He gave excellent results with this technique obviating the need for initial diversions. Although the number of complications was relatively high, the follow-up showed good quality of life with primary gastric transposition.

Lateral esophagostomy was described by Aloisi AS et al\textsuperscript{[8]} and it was possible to perform a delayed primary repair after elongation of the proximal pouch due to stretching by the ingested saliva. Lateral esphagostomy has the advantage that child may be treated at home avoiding the risk of aspirations and sufficient time may be given for elongation of the proximal pouch for a delayed primary repair.

IEA was universally fatal in our setup until the neonatal care, parerteral nutrition and concept of proper
preoperative stabilization of the baby was introduced. Our first baby with IEA also had anorectal atresia and thus also needed a colostomy in the newborn period. For his definitive treatment various options were evaluated. Colonic interpositioning was not possible due to colostomy; Reverse gastric tube was not possible due to small capacity stomach. With limited options we preferred to perform a Jejunal interpositioning. He developed minor leak which settled with conservative treatment Encouraged by the good short term results in this baby we also performed jejunal interpositioning in the second baby. Both however developed long term complications like gastroesophageal reflux, difficulty in swallowing due to redundant jejunum and recurrent chest infections. We noticed that Jejunal inter-positioning is a difficult conduit to construct and is associated with significant complications therefore we have not used it since our two cases and shall only be performed if other options are not available and for segmental substitution.

Reverse gastric tube interposition has been our favorite substitute in cases of corrosive injuries and peptic strictures but we found that reverse gastric tube has high incidence of complications in IEA. Initial gastrostomy and small capacity stomach damages the blood supply for the reverse gastric tube and thus the incidence of complications like anastomotic leak is high. We lost one patient who had an anastomotic leak leading to severe mediastinitis and baby died inspite of vigorous treatment. We therefore consider reverse gastric tube only in selected cases. Other studies have however shown good results of reverse gastric tube interpositioning in long gap EA.

Ludman and Spitz have shown excellent results with primary gastric pull-up. We performed it in one patient where extra thoracic lengthening had failed and are satisfied with the initial results. As we are not performing a primary gastric transpositioning thus gastric transpositioning is not our preferred technique for IEA.

Extra-thoracic lengthening gave satisfactory result in one of two patients. The main problem has been difficulty in serially mobilizing the esophagus due to dense adhesions. The use of Gortex mesh as advised by Dessanti et al may be helpful in making it a suitable procedure. This procedure however does involve repeated general anesthesia and we feel that it gives good results in selected cases.

Lateral esophagostomy is a simple procedure where instead of end stoma a lateral stoma is made. The baby may be sent home on gastrostomy feeding and the risk of aspiration are minimal. We advise the family to keep the baby in prone position. This helps in draining the secretions. The esophageal length grows over the period of time and no extra procedure is necessary. The length of the proximal segment may be monitored by single stout X-ray with contrast in esophagus. Once adequate length of the esophagus has been achieved a delayed primary repair of the esophagus may be performed. The procedure itself is especially suitable for the developing countries where the risk of major neonatal surgical intervention like gastric tranpositioning may be high. We therefore recommend that lateral esophageotomy and gastrostomy may be performed in all babies with IEA with delayed primary repair. In some babies it may not be possible to perform a lateral esophageotomy due to very short proximal pouch. In those cases, other options may be used to achieve esophageal continuity.

CONCLUSION

Isolated Esophageal atresia is a difficulty management problem. None of the esophageal substitutions have given the optimal results. Lateral esophageotomy may be a good technique for IEA as it causes spontaneous lengthening of the proximal segment without need of admission or manual stretching. It may be the treatment of choice in developing countries. Results may however differ in patients and will depend upon the initial length of proximal segment.

REFERENCES