

## The role of fiberoptic endoscopy in the evaluation and management of long gap isolated esophageal atresia

Erica Rachel Gross · Ari Reichstein ·  
Jeffrey W. Gander · Charles J. H. Stolar ·  
Arnold G. Coran · Robert A. Cowles

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**Abstract** Accurate measurement of gap length is useful for operative planning in cases of esophageal atresia (EA) without distal fistula. This paper demonstrates how fiberoptic endoscopy of the distal esophagus enables measurement of the gap in the case of isolated EA, and compares other commonly practiced techniques.

**Keywords** Isolated esophageal atresia · Esophageal gap · Fiberoptic endoscopy · Rigid dilator

### Introduction

In long gap isolated esophageal atresia (EA), in order to restore the continuity of the esophagus with the least morbid and most functional technique, the distance of the gap between the proximal and distal esophageal pouches should be estimated pre-operatively. Inaccurate measurement can lead to premature or ill-advised attempts at primary repair, anastomotic leak, fistulization, stricture and multiple salvage operations. A standardized approach to gap measurement aids pre-operative planning and may decrease the morbidity related to reconstruction of pure EA. Although a variety of techniques exist for the pre-operative measurement of esophageal gap, the benefits of one approach versus another have not been compared. This report describes the use of a flexible endoscope to measure gap length in long gap EA and compares it with two other common gap measurement techniques: (1) attempted placement of a rigid

dilator to delineate the distal pouch, and (2) injection of contrast via the gastrostomy tube.

### Materials and methods

A female child was born at 36 weeks of gestation with prenatal ultrasound findings of polyhydramnios and a poorly distended stomach, raising concern for isolated EA. At birth, the baby had no respiratory distress. A Repleglo tube was placed but could not be advanced. The initial chest radiograph revealed the tip of the Repleglo tube just below the heads of the clavicles and a gasless abdomen, suggesting the diagnosis of EA without distal tracheoesophageal fistula. On physical examination, the only abnormality identified was a single palmar crease and a sprengele deformity of the left scapula. All other pre-operative investigations were within the normal limits.

The patient was taken to the operating room and underwent bronchoscopy, followed by open gastrostomy tube placement. No proximal fistula was identified, and a diminutive stomach was noted at laparotomy. A 16 French (F) Malecot tube was placed for feeding access. A Repleglo tube remained in the upper esophageal pouch to drain oral secretions, and gastrostomy tube feedings were initiated the next day.

At 6 weeks of age, she returned to the operating room for the measurement of the esophageal gap. Under general anesthesia, the Repleglo and gastrostomy tubes were removed. A mercury-filled bougie was inserted into the oropharynx and advanced into the upper esophageal pouch. Then, the gastrostomy was dilated with an 8-mm Hegar dilator. Using fluoroscopy, the dilator was inserted into the gastrostomy with an attempt to intubate the distal

E. R. Gross (✉) · A. Reichstein · J. W. Gander ·  
C. J. H. Stolar · A. G. Coran · R. A. Cowles  
Morgan Stanley Children's Hospital, New York, USA  
e-mail: eg2561@columbia.edu

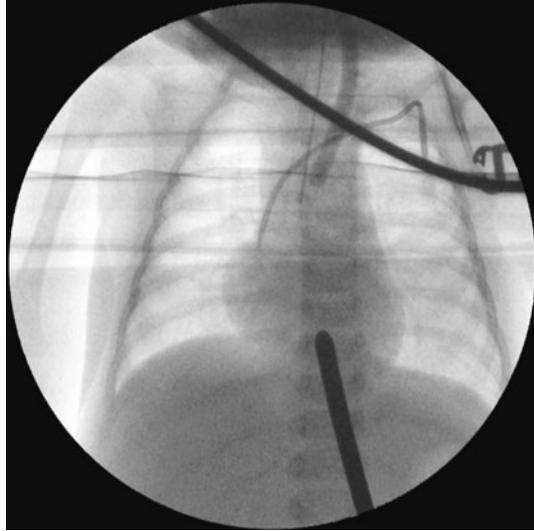
esophageal pouch (Fig. 1). Using this technique, the gap was measured as two vertebral bodies.

The Hegar dilator was removed, and a neonatal endoscope (Pentax EG-1690 K, 2.0; 5.4 mm in diameter) was inserted via the gastrostomy. The stomach was insufflated, and the pylorus and incisura angularis were identified. The gastroesophageal junction (GEJ) was visualized, and the distal esophagus was intubated with the endoscope. A fluoroscopic image was obtained (Fig. 2). The gap was estimated to be 3.5 vertebral bodies with this technique. Given the discrepancy in measurements, the decision was made to delay reconstruction and remeasure the gap length in 6 weeks, allowing for continued growth of the esophagus.

The child was brought back to the operating room at 12 weeks of age for final evaluation of the esophageal gap and definitive surgical reconstruction of the esophagus. Again, the Replogle and gastrostomy tubes were removed and a mercury-filled bougie was inserted into the proximal esophageal pouch. An initial measurement with the Hegar dilator was performed (Fig. 3), and on this occasion, the gap length was estimated at five vertebral bodies. Next, the gastrostomy tube was reinserted and contrast was injected into the stomach. No distal esophagus was visualized on this contrast study (Fig. 4). Finally, the neonatal endoscope was inserted into the stoma. The GEJ was visualized and appeared to be contracted. After intubation of the distal



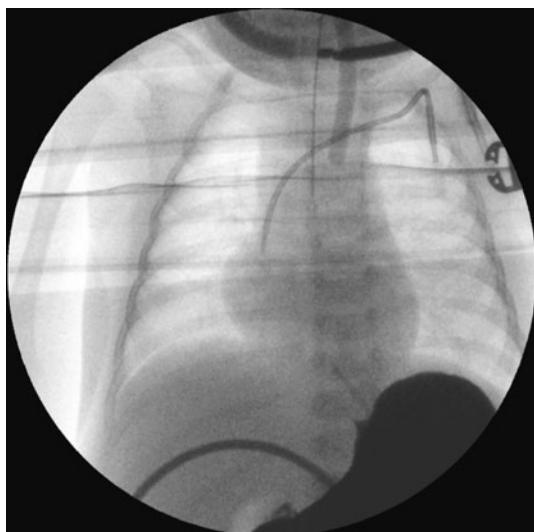
**Fig. 1** Hegar dilator inserted through the gastrostomy into the distal pouch with fluoroscopic guidance at 6 weeks of age



**Fig. 3** Hegar dilator inserted through the gastrostomy into the distal pouch with fluoroscopic guidance at 12 weeks of age



**Fig. 2** Fluoroscopic image of endoscope in distal esophageal pouch at 6 weeks of age



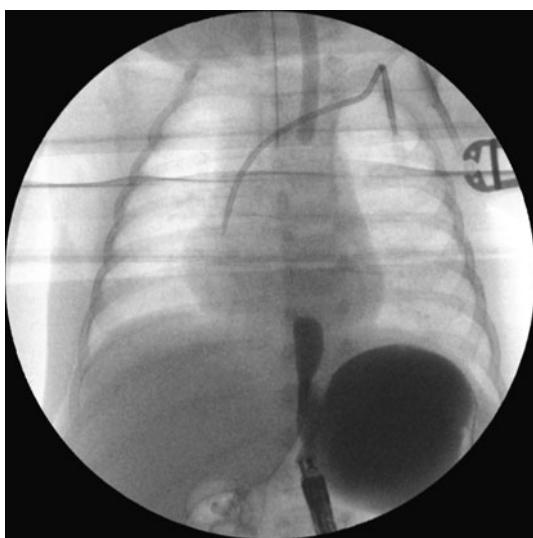
**Fig. 4** Fluoroscopic image of water-soluble contrast administered through the gastrostomy tube at 12 weeks of age

esophageal pouch (Fig. 5), contrast was injected through the endoscope and a final fluoroscopic image was taken (Fig. 6). Both images using the endoscope revealed an esophageal gap measurement of 3.5 vertebral bodies.

With these findings, the patient underwent primary repair through a standard right thoracic extra-pleural approach. The flexible endoscope was left in the distal esophageal pouch during the thoracic dissection to aid in its intra-operative identification. The pouches were mobilized, and an anastomosis was performed under moderate tension. A 6.6 F Broviac catheter was used as an esophageal stent. A contrast study was performed 7 days after surgery and no leak was noted. Oral feedings were then begun in addition to gastrostomy tube feedings, and the infant was discharged



**Fig. 5** Fluoroscopic image of endoscope in distal esophageal pouch at 12 weeks of age



**Fig. 6** Fluoroscopic image after injection of water-soluble contrast through endoscope at 12 weeks of age

home 23 days after surgery, receiving oral and bolus tube feedings.

The Institutional Review Board of Columbia University has waived the need for written informed consent.

## Results

At both 6 and 12 weeks of age, the measurements of esophageal gap that were obtained using the Hegar dilator were inconsistent when compared with images using a neonatal endoscope. At 6 weeks, the fluoroscopic image with the Hegar dilator in the distal segment suggested the gap measurement to be only two vertebral bodies. At 12 weeks, however, the same technique estimated the gap at five vertebral bodies. In contrast, placement of the endoscope into the distal pouch via the gastrostomy yielded a more consistent measurement of 3.5 vertebral bodies at both time points. The study using contrast injected through the gastrostomy tube also failed to adequately identify the lower esophageal pouch. In this image, no distal esophagus was delineated. This imprecision is clearly seen when compared to the study with contrast injected through the endoscope (Fig. 6).

Especially, both images using an endoscope to identify the distal pouch, with and without contrast, display a gap measurement of 3.5 vertebral bodies at both 6 and 12 weeks of age. This indicates that there was no significant growth of the esophageal pouches during the interval time period.

## Discussion

Some authors have suggested that post-operative morbidity and mortality associated with EA depend on the length of esophageal gap. Hand and Dudley [1] analyzed follow-up data for neonates after repair of EA to identify predictors of morbidity and mortality and found that neonates with a long gap length (>2 cm at operation) had significantly more post-operative respiratory complications and failure to thrive. Brown and Tam [2] also proposed a classification based on the length of esophageal gap to predict surgical outcome in the cases of EA. They found that gap length was strongly associated with mortality, rate of anastomotic leak, stricture and gastroesophageal reflux after repair, failure to thrive, and length of hospital stay during initial admission. In addition, these studies suggest that gap length appears to be an important indicator of disease severity in EA, and that accurate measurement of esophageal gap is imperative.

Many methods of measurement of esophageal gap have been described, but no comparison of these techniques has

been made. Some authors have described placement of a rigid dilator to measure the distal esophageal remnant [3–5]. Rossi et al. [4] state that this technique is simple, fast, safe, and accurate. While the simplicity of this method is self-evident, its ease and accuracy, however, are debatable. Without anesthesia, blind passage of a rigid dilator through the GEJ into the distal pouch may be difficult [5]. The lower esophageal sphincter is tonically contracted, as seen in our second endoscopic measurement, and there is an evidence that some children with EA retain an intact gasto-esophageal mechanism. We believe that even with the use of anesthesia, it is difficult to intubate the distal esophageal pouch consistently, and the present case illustrates this challenge. The dilator may elevate the cardia giving the impression on fluoroscopy of a distal pouch.

Caffarena et al. [6] first described placing an endoscope in both the upper and lower esophageal pouches as a means of measurement of esophageal gap. Chan and Saing [7] described a similar approach, using a flexible bronchoscope via the gastrostomy to visualize and intubate the distal esophageal pouch. Endoscopy often requires the use of general anesthesia and maturation of the gastrostomy tube site. Therefore, the first measurement should be performed at 4–6 weeks of age, when the gastrostomy site is mature enough to safely allow intubation. A gastrostomy tube should be placed at birth and be of sufficient size to allow passage of the scope. Despite the need for general anesthesia, there is an assurance of localization of the lower pouch. Additionally, the endoscope may be left in the distal esophageal pouch during reconstruction allowing intra-operative illumination and visualization. This is especially helpful when the distal esophageal pouch is small and, thus, difficult to find at thoracotomy.

Injection of water-soluble contrast via the gastrostomy tube may be the simplest and oldest technique performed to evaluate the lower esophageal pouch. No special equipment is needed, and the study can be performed without anesthesia. The GEJ, however, must be incompetent to allow the reflux of contrast into the distal pouch. In the case presented, a contracted GEJ was visualized, and there was no reflux of contrast.

Another published technique uses computed tomography (CT) scanning for evaluation of EA [8, 9]. CT can offer early detection of the fistula and direct measurement of the gap. CT is accessible, quick, and simply requires placement of the neonate perpendicular to the plane of the scanner (sagittal imaging) [9]. The accuracy of identifying the distal esophageal pouch using CT, however, has the same drawback as using water-soluble contrast. Visualization of the lower pouch relies on gaseous distention of the stomach and reflux of air through the GEJ. The dose of radiation is also significantly higher than fluoroscopy, especially considering that neonates with pure EA will

likely require multiple images over time as the esophageal pouches grow. Sedation of the neonate for CT imaging can also be a concern, further limiting the utility of this technique.

A problem common to the use of a rigid dilator, as well as fiberoptic endoscopy of the distal pouch is the amount of tension applied during the measurement. This can differ between operators as well as between different measurements performed at by the same operator. While applying tension gives more information regarding mobilization of the pouches, tension is a variable that cannot be quantified or standardized.

Finally, although this represents only one case, the concept of using endoscopy to directly delineate the distal pouch is valid and useful. In fact, the senior author (AGC) of this report has successfully employed this technique for the past 15 years in over 15 patients.

## Conclusion

Accurate measurement of esophageal gap is critical for operative planning. The presented case illustrates that gap measurement with a rigid dilator or contrast is not reliable and that a neonatal endoscope should be used. A technique that employs endoscopy allows the distal pouch to be intubated under direct vision. Injection of contrast through the endoscope can yield additional information by outlining the distal esophageal pouch.

**Conflict of interest** None.

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