Management Strategies and Outcomes of Distal Congenital Esophageal Strictures in the Setting of Long-Gap Esophageal Atresia

Ali Kamran MD¹, C. Jason Smithers MD², Somalia Mohammed MD¹, Shawn Izadi MD¹, Farokh Demehri MD¹, Hester Shieh MD², Peter Ngo MD¹, Jessica Yasuda MD¹, Denis Chang MD¹, Michael J. Wilsey MD², Benjamin Zendejas MD, MSc¹

¹Boston Children’s Hospital, Boston, MA, USA. ²John Hopkins All Children’s Hospital, St. Petersburg, FL, USA

Abstract

Background: The management of neonates with long-gap esophageal atresia (LGEA) combined with distal congenital esophageal strictures (CES) is challenging. We sought to review our approach for this rare set of anomalies.

Methods: We reviewed children with LGEA+CES surgically treated at two institutions (2018 - 2024). LGEA repair was performed using the Foker technique (traction-induced esophageal lengthening). A CES strategy was chosen based on preoperative evaluations and intraoperative findings. The configuration, and length of the CES were assessed using retrograde flexible esophagoscopy via gastrostomy with contrast fluoroscopy (Figure).

Results: Eight patients (75% male) with LGEA+CES were treated: Five had type A and three had type B EA, median gap length was 3.5 cm. Three underwent thoracoscopic esophageal lengthening. After a median follow-up of 18 months (IQR: 9-25), all retained their native esophagus. However, those who had CES resection concurrent with the lengthening process or at the time of EA anastomosis had more challenging perioperative courses: one required additional time on traction, and another required esophageal anastomotic stricture resection.

Conclusions: Our experience with LGEA and distal CES emphasizes tailoring surgical approaches to each patient’s unique condition, avoiding a one-size-fits-all strategy. However, if the esophageal tissue above the distal CES is in good condition, our preference has shifted towards retaining the CES during traction, performing gentle dilation at anastomosis time, and conducting definitive endoscopic management subsequently. We would caution against making the assumption that salvage of the native esophagus is not possible or that resection of the CES is always needed.