Endoscopic Features in Esophageal Atresia: From Birth to Adulthood

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Since the first successful surgery in 1941 (1), the survival of patients with esophageal atresia (EA) has improved greatly. Consequently, the follow-up of these patients has revealed gastrointestinal (GI) complications unrecognized 60 years ago, such as gastroesophageal reflux and esophagitis with their related consequences. Among these complications, 1 of the major concerns in long-term GI follow-up is the increased incidence of gastric metaplasia of the esophagus (2,3), intestinal metaplasia (4,5), and esophageal adenocarcinoma (6–8), all of which have been reported in young adults with EA (5–7). However, the exact incidence and natural history of these complications are unknown in patients with EA, and whether systematic upper GI endoscopic screening and surveillance in the follow-up of patients with EA is recommended is not yet established.

In this article, we review the upper GI endoscopy findings in patients with EA, the specificities of upper GI endoscopy in patients with EA (How to scope them? What about landmarks in EA? Do we need to scope the patients? When? How often?), and present a proposed algorithm for the surveillance of patients, children, and adults with EA. We do not address the problem of congenital stenosis or anastomotic and peptic strictures.

WHAT IS ACTUALLY OBSERVED IN PATIENTS WITH EA?

Esophagitis and Long-Term Consequences of Esophagitis

Peptic esophagitis (2,9) and Barrett esophagus (2,9) have been reported in children (2,3,9,10) and more recently in adults with repaired EA (Fig. 1) (4,5,11,12). We recently reported on a systematic cross-sectional endoscopic evaluation in 45 children with EA (median age 7.3 years, range 0.4–17.9). Twenty-six patients (58%) had normal endoscopy, 14 (31%) had esophagitis, and 16 (36%) had gastric metaplasia (10). No intestinal metaplasia or adenocarcinoma was detected, similar to other reports. Data in adults are sparse and have been published recently (4,5,11,12). Maynard et al reported preliminary results of a systematic endoscopic screening in 21 adults ages 27 ± 7 years born with EA (personal communication). Esophagitis was found in 29% of patients ($n = 6$) and Barrett endoscopically suspected in 24% of patients ($n = 5$) in whom intestinal metaplasia was documented in 3 cases. These data demonstrate that EA, even in childhood, is a significant risk factor for the development of esophagitis and Barrett esophagus.

Miscellaneous

Various anecdotal features can also be observed in patients with EA. Some of them may have clinical consequences (eg, diverticulum) and may be related to symptoms. Others such as a high incidence of heterotopic pancreas (13) could lead to a new pathophysiological hypothesis whether or not this association between EA and heterotopic pancreas is related to the same developmental mechanisms. Eosinophilic esophagitis should be ruled out in patients with EA for whom dysphagia worsen without stricture (14).

SPECIFICITIES OF UPPER GI ENDOSCOPY IN EA

Do the Patients Need Endoscopy? When? How Often?

Performing a systematic endoscopic assessment in patients with EA is largely debated in the pediatric (9,15) and, more recently, adult literature (11). The predictive value of an early (<3 years old) normal upper gastrointestinal endoscopy is unknown regarding development of peptic esophagitis and Barrett esophagus later in life. In a study designed to determine whether clinical symptoms may predict endoscopic lesions (10), 63 patients with EA were recruited. Eighteen had dysphagia related to an esophageal stricture needing dilatation and were not included in the analysis. Forty-five patients (26 girls) with a median age of 7.3 years (range 0.4–17.9) were evaluated. Mucosal abnormalities at endoscopy were observed in 19 of 45 patients (42%). Symptoms were reported by 62%, but none could be identified as statistically associated with an abnormal endoscopy. Furthermore, 6 asymptomatic patients had abnormal endoscopies, reflecting the difficulty in adequately deciding which patients need to be investigated (10). Similarly, in adults, the symptoms are frequent but underreported by the patients who do not seek medical attention.

Pitfalls: Of the Importance of Correct Landmarks in Patients With EA

Hiatal hernia or traction on the stomach during surgery can complicate landmark recognition and lead to erroneous diagnosis of gastric metaplasia, mostly in patients with a long-gap atresia. In this regard, the esophageal mucosa should be determined by carefully delimiting the gastroesophageal junction, identified as the proximal margin of the gastric mucosal folds, as defined by Prague C & M criteria (16).

CONCLUSIONS AND PERSPECTIVES

The present data demonstrate the high incidence of esophageal mucosal lesions in pediatric and adult patients with EA without

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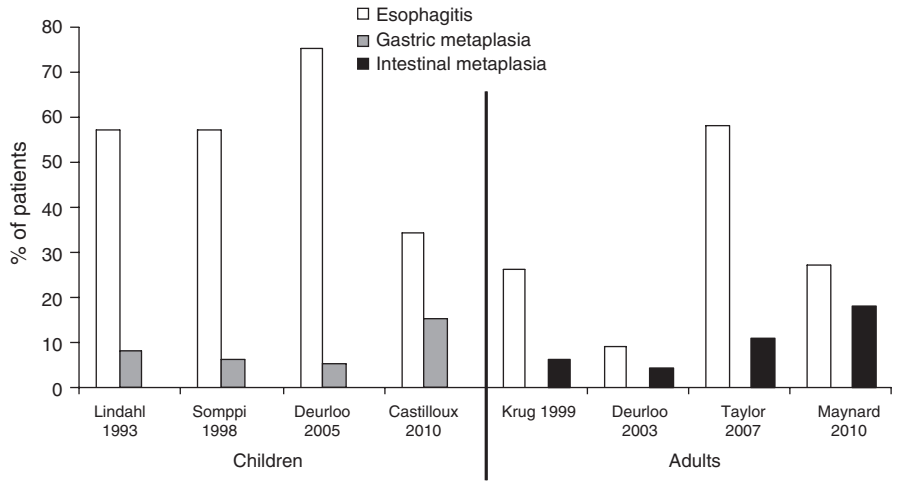

FIGURE 1. Incidence of macroscopic mucosal anomalies, esophagitis, and gastric and intestinal metaplasia in patients with surgically intervened esophageal atresia. (Data are summarized from references 2, 3, 9, and 10 for children and references 4, 5, 11, and 12 for adults.)

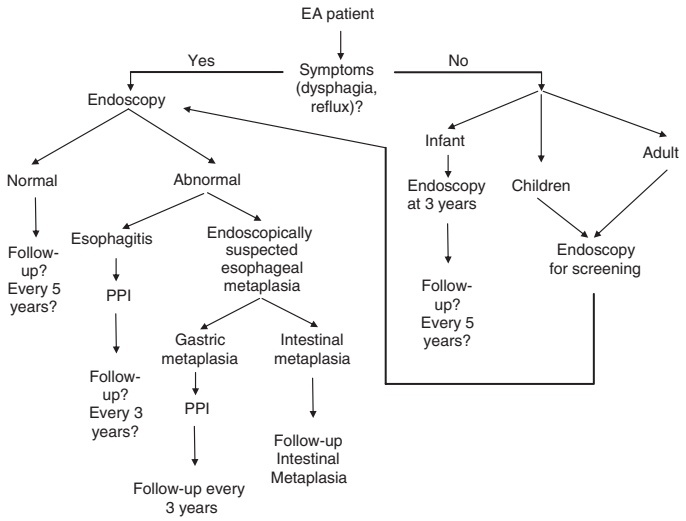

FIGURE 2. Proposed algorithm for the evaluation and surveillance of esophageal mucosa by upper gastrointestinal endoscopy in pediatric and adult patients with EA. The question marks depict the lack of data in the literature. EA = esophageal atresia; PPI = proton pump inhibitor.

any predictive clinical symptoms. This argues for a systematic screening of endoscopic lesions in all patients with EA even in the absence of upper GI symptoms; a proposed algorithm for the evaluation and surveillance of esophageal mucosa by upper endoscopy in patients with EA is provided (Fig. 2). However, the evaluation of systematic endoscopic follow-up with esophageal biopsies in patients with EA to evaluate accurately the complications, the response to treatment, and the outcome are urgently warranted. The predictive value of a normal endoscopy should be assessed as well as the value of a new technology such as narrow band imaging in the characterization of the gastroesophageal junction of these patients. Finally, a close collaboration should be established with pathologists, the lesions should be described according to standardized landmarks, and biopsy samples should be taken with special care.

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