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Mini-symposium: Oesophageal Atresia and Tracheo-oesophageal Fistula

Feeding Difficulties in Children with Esophageal Atresia

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EDUCATIONAL AIMS

- To describe the mechanism of esophageal dysphagia and oropharyngeal dysphagia/aspiration as mechanisms for feeding difficulties in patients with esophageal atresia
- To highlight the prevalence of respiratory symptoms in patients with esophageal atresia
- To review methods for diagnosing aspiration
- To discuss treatment strategies for management of aspiration

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SUMMARY

The current available literature evaluating feeding difficulties in children with esophageal atresia was reviewed. The published literature was searched through PubMed using a pre-defined search strategy. Feeding difficulties are commonly encountered in children and adults with repaired esophageal atresia [EA]. The mechanism for abnormal feeding includes both esophageal and oropharyngeal dysphagia. Esophageal dysphagia is commonly reported in patients with EA and causes include dysmotility, anatomic lesions, esophageal outlet obstruction and esophageal inflammation. Endoscopic evaluation, esophageal manometry and esophograms can be useful studies to evaluate for causes of esophageal dysphagia. Oropharyngeal dysfunction and aspiration are also important mechanisms for feeding difficulties in patients with EA. These patients often present with respiratory symptoms. Videofluoroscopic swallow study, salivagram, fiberoptic endoscopic evaluation of swallowing and high-resolution manometry can all be helpful tools to identify aspiration. Once diagnosed, management goals include reduction of aspiration during swallowing, reducing full column reflux into the oropharynx and continuation of oral feeding to maintain skills. We review specific strategies which can be used to reduce aspiration of gastric contents, including thickening feeds, changing feeding schedule, switching formula, trialing transpyloric feeds and fundoplication.

INTRODUCTION

Esophageal atresia, with or without tracheoesophageal fistula (TEF), is a congenital anomaly of the esophagus affecting one in every 2500-3000 live births [1]. Infant survival of this condition is high, with reported survival rates of over 90% [2]. However, gastrointestinal and respiratory complications are well documented in children, adolescents and adults with repaired EA [3–9]. Feeding disorders in children with esophageal atresia are common in clinical practice but the literature supporting these observations is limited. Between 6% and 52% of patients have some abnormalities of feeding [7,10,11]. The majority of studies focus on esophageal abnormalities as source of feeding difficulties. There are no prospective studies on oropharyngeal dysfunction or aerodigestive abnormalities in patients with esophageal atresia.

OVERVIEW OF FEEDING DIFFICULTIES

Feeding difficulties have been described in patients with EA. Puntis et al., [11] first characterized feeding difficulties in 124 children with EA. Compared to healthy controls, children born with EA were significantly more likely to eat slowly, refuse meals, cough or choke during eating and vomit with meals. Chetcuti et al., [12] described similar feeding difficulties in childhood, but noted that overall, these difficulties lessen with

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Abbreviations: EA, Esophageal atresia; TEF, Tracheoesophageal fistula; VFSS, Videofluroscopic Swallow Study.

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age, with < 10% of patients age 15 or older reporting pervasive feeding difficulties. Patients who have undergone primary repair of long-gap esophageal atresia achieve major feeding milestones in a similar pattern to normal control infants, although do have much greater variability in achieving these milestones [13]. Baird et al., [10] administered a validated feeding questionnaire to 30 caregivers of children with EA. They found that, in comparison to controls, 17.5% of children with EA have feeding scores one standard deviation above the mean feeding difficulty score and 6.7% of cases are greater than two standard deviations above the mean. However, overall, feeding difficulties were classified as mild and in the subclinical range in the majority of patients. Schier et al., [14] administered questionnaires to 128 parents involved in an EA support group. 68% of respondents experienced feeding difficulties which included pain, regurgitation, vomiting and burping. Patients generally avoided meats and other tough or bulky foods and 69% of patients experienced at least 1 food impaction. Given the high prevalence of feeding difficulties in this population, Ramsey et al., [15] advocated for early involvement of a multidisciplinary team comprised of occupational therapy, nutrition and psychological support to assist families with feeding-related difficulties. However, despite the fact that feeding difficulties are common in patients with EA, only 11% of parents report discussing their concerns with medical staff [11].

MECHANISM OF ABNORMAL FEEDING

Esophageal dysphagia as a cause for feeding difficulty

Dysphagia is a common complaint in patients with EA and causes include dysmotility, anatomic lesions, esophageal outlet obstruction and esophageal inflammation. The reported prevalence of dysphagia in patients with EA ranges from 38% to 85% [3,8,9,12,16,17]. A recent systematic review and meta-analysis by Connor et al. found an overall pooled estimated prevalence of 50.3% (95% CI 35.7 – 64.8) [5]. The evaluation of dysphagia involves (1) an esophagram to assess for strictures or, when present, pooling above the fundoplication; (2) videofluroscopic imaging during swallowing to assess for aspiration; (3) upper endoscopy to assess for inflammation and; (4) esophageal motility study to assess for adequate contraction pressures, and when paired with impedance, to assess for bolus stasis.

Endoscopic evaluation: Esophageal inflammation is common in patients with EA and is often is often implicated as a cause of dysphagia. In a cross-sectional study by Castilloux et al. of 45 patients with esophageal atresia undergoing endoscopic assessment, 31% had histologic evidence of esophagitis and 36% had gastric metaplasia [16]. Interestingly, there was no association between symptoms of dysphagia and endoscopic findings, either grossly or histologically in this cohort. Similarly, Sistonen et al. reported histologic esophagitis in 25% of patients, however there were no significant differences in rates of dysphagia between patients with esophagitis and those with normal biopsies [17]. Deurloo et al. found that while patients who reported dysphagia more often had disturbed motility on esophageal manometry, there was no association between reported dysphagia and biopsy confirmed esophagitis [18]. Further supporting this observation is the finding that 38% of patients with food impactions have normal esophageal biopsies, suggesting that food impactions can occur even in the absence of inflammation and may be more related to abnormal esophageal motility [16].

Esophageal Manometry: Esophageal dysmotility is common in patients with EA. Sistonen et al. report normal propagating peristalsis in only 20% of the 101 adult patient studied [17]. Deurloo et al. described similar findings, with 70% of patients having low or moderate amplitude of esophageal body contractions and 35% of

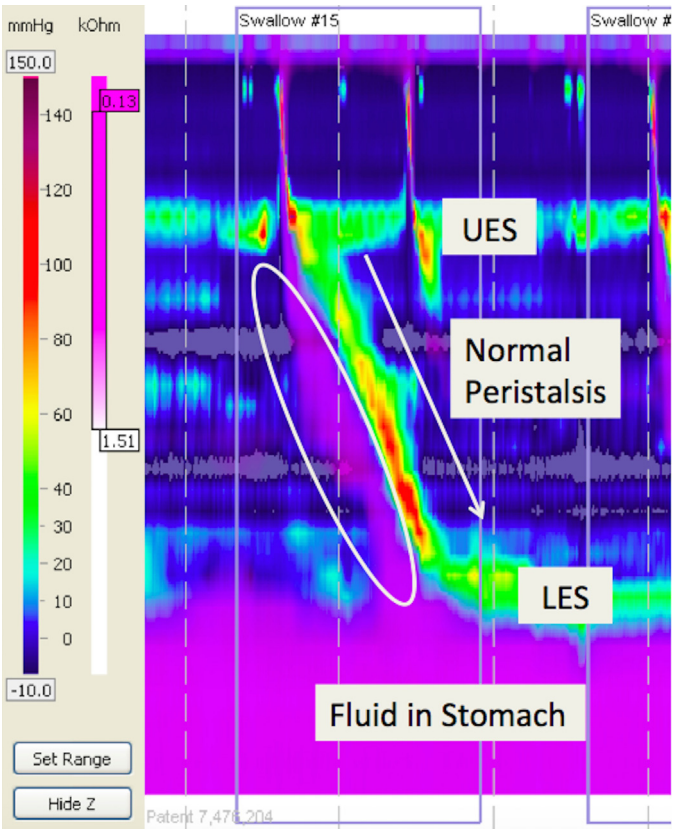

Figure 1. Normal peristalsis with normal bolus clearance using high resolution esophageal manometry with impedance. Purple: Liquid. Note that with each peristaltic wave in yellow/red there is complete bolus clearance with no residual purple in the esophagus (white circle).
LES: Lower esophageal sphincter; UES: Upper esophageal sphincter.

patients having retrograde contractions [18]. Furthermore, they found that patients reporting dysphagia more often had disturbed motility and significantly lower scores on a variety of health-related quality of life scales. Kawahara et al. also described a lack of distal esophageal contractions on esophageal manometry in patients with EA [19]. For centers where esophageal motility studies are not available, even radionucleotide esophagogastric studies reveal significantly longer esophageal transit time in patients with a history of long-gap EA, compared to those with non-long-gap EA suggesting that imaging may be helpful in identifying dysmotility. In these patients, the bolus accumulated mainly in the lower 2/3 of the esophagus below the anastomosis and persisted in this area for several minutes before being cleared into the stomach [20] (see Figures 1 and 2). This suggests that impaired clearing capacity may be playing a role in the dysphagia in these patients. There appears to be overall improvement in esophageal peristaltic function on manometric studies as patients age [21]. More recently, high-resolution manometry has been used in the EA population to better characterize both esophageal dysmotility and extraesophageal symptoms. Lemoine et al. used high-resolution manometry in 40 children with a history of EA repair [22]. All patients had abnormal manometry results: 38% of patients had aperistalsis, 15% had pressurization and 47% had abnormal distal contractions. They found both gastroesophageal reflux and pulmonary symptoms more commonly in the aperistalsis group. Kawahara et al. found the absence of significant contractions in the middle esophagus just below the anastomosis in 29 patients with repaired EA [19]. Lack of distal esophageal contractions with significantly correlated with development of gastroesophageal reflux in this population ($P < 0.001$). Patients