Clinician knowledge of societal guidelines on management of gastrointestinal complications in esophageal atresia

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ABSTRACT

Objectives: The aim of this study was to assess whether clinicians approached the management of children with esophageal atresia (EA) in accordance with the 2016 European Society of Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN)/North American Society of Paediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) guidelines on the management of gastrointestinal and nutritional complications in this cohort.

Methods: We invited expert physicians and surgeons closely involved in the care of children with EA (members of the International network on esophageal atresia [INoEA], ESPGHAN EA working group, French national EA registry, European pediatric surgical association (EUPSA), and European rare disease reference network [ERNICA]) to participate in an anonymous online survey containing 15 multiple choice questions concerning the management of gastrointestinal and nutritional complications in children with EA. Questions were based on the management of gastroesophageal reflux disease (GERD) dysphagia, cyanotic spells, feeding and nutrition, anastamotic strictures, and transition to adult care as detailed in the 2016 guidelines.

Results: Median concordance with ESPGHAN/NASPHAN EA Guidelines was 69% (16–100%, SD 16%) across all responders. Areas of greatest concordance were in the fields of surveillance endoscopy and medical management of GERD. Areas for potential educational opportunities include: the differential diagnosis and appropriate investigation of dysphagia and the diagnostic evaluation of extraesophageal symptoms.

Conclusions: This survey highlights the importance of improving the understanding and adherence to the EA guidelines amongst clinicians involved in the care of these patients.

Key Words: anastomotic stricture, dysphagia, esophageal atresia, fundoplication, gastroesophageal reflux, guidelines, transition

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What Is Known

- European Society of Paediatric Gastroenterology, Hepatology and Nutrition/North American Society of Paediatric Gastroenterology, Hepatology and Nutrition guidelines are internationally compiled societal guidelines on the management of gastrointestinal and nutritional complications in children with esophageal atresia.

What Is New

- This is the first study to assess expert concordance with the European Society of Paediatric Gastroenterology, Hepatology and Nutrition/North American Society of Paediatric Gastroenterology, Hepatology and Nutrition guidelines on esophageal atresia.
- Overall, this study showed that specialist health professionals involved in the care of children with esophageal atresia had good concordance with the societal guidelines.
- This study also identifies some areas in which there was lack of concordance with the esophageal atresia guidelines, which needs to be addressed.

Esophageal atresia (EA) is one of the most common congenital malformations of the digestive tract affecting 1 in 2500 to 1 in 4500 live births (1–3). Improved operative and perioperative care has resulted in survival rates between 90% and 100% (4), with management of EA patients now focused on improving

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management of associated symptoms and complications. Gastroesophageal reflux disease, gastric metaplasia and Barrett esophagus, anastomotic strictures (AS), eosinophilic esophagitis (EoE), feeding disorders, dysphagia, esophageal dysmotility, cyanotic spells/BRUEs (brief resolved unexplained events), and tracheomalacia are frequent complications seen in these children (5). Due to the growing awareness of medical and surgical morbidities of these patients combined with the improved natural history studies, the European Society of Paediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) and the North American Society of Paediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) jointly published the ESPGHAN/NASPGHAN guidelines for the management of gastrointestinal and nutritional complications in children with repaired EA. Overall, there were 36 recommendations/statements on the management of these complications in the guidelines. These guidelines were the first to provide evidence-based recommendations for the evaluation and treatment of gastrointestinal complications and follow-up in children with EA. To assess the impact of these guidelines on clinical practice, we emailed a 15-question questionnaire to specialist providers who care for these patients.

METHODS

Study Population

We emailed a 15-question questionnaire to health professionals with expertise in the care of EA patients who were on mailing lists from the International Network on Esophageal Atresia (INoEA), the European Society of Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) Esophageal Atresia working group, the French national Esophageal Atresia registry, the European Pediatric Surgical Association (EUPSA), and European Rare Disease Reference Network (ERNICA). Only those clinicians closely involved in the care of EA patients—gastroenterologists, surgeons and respirologists, and otolaryngologists were included as the aim was to initially test the awareness and understanding of the guidelines amongst the specialist clinicians who would be most familiar with them. An initial invitation email was sent with a link to a Survey Monkey questionnaire, with 3 follow-up reminder emails sent.

Questionnaire

We developed a questionnaire containing 15 multiple choice questions. The questions assessed familiarity with the 2016 ESPGHAN/NASPGHAN gastrointestinal (GI) guidelines, with specific reference to gastroesophageal reflux disease (GERD), fundoplication, dysphagia, AS, cyanotic spells, EoE, vascular anomalies, feeding and nutrition, and transition to adult care. Several questions required more than 1 correct answer. In addition to fact-based recall questions, we included 3 clinical scenarios wherever applied knowledge was required. The multiple-choice questions are shown in Supplemental Table 1, *http://links.lww.com/MPG/B972* with the correct answers highlighted in bold.

RESULTS AND DISCUSSION

On the basis of the databases discussed in the methods, 332 email addresses were initially included. After removing duplicate email addresses and incorrect email addresses, 182 correct email addresses remained, and surveys were sent out to these care providers. There were 123 (67%) responders who completed the questionnaire.

Median concordance with ESPGHAN/NASPHAN EA Guidelines was 70% (16%–100%, standard deviation [SD] 16%) across all responders (Fig. 1), with an average completion time of 10 minutes. Percentage of correct responses in each of the areas tested is shown in Table 1. Average scores with standard deviations for each of the questions in the survey is shown in Table 2.

GASTROESOPHAGEAL REFLUX IN ESOPHAGEAL ATRESIA PATIENTS

Acid Suppressive Therapy

Guideline statement: It is recommended that GER be treated with acid suppression in all EA patients in the neonatal period (5).

Eighty-seven percentage of responders agreed that all neonates with repaired EA should be systemically treated with anti-acid treatment for the first year of life. This good concordance with respect to the management of GER in EA patients is at odds with the

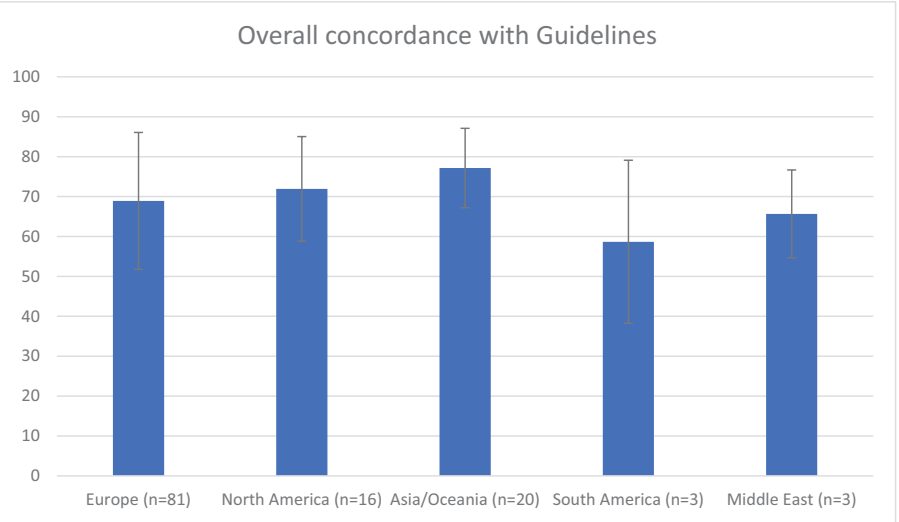

FIGURE 1. Average concordance in percentage of correct responses overall with European Society of Paediatric Gastroenterology, Hepatology and Nutrition/North American Society of Paediatric Gastroenterology, Hepatology and Nutrition EA Guidelines by continent with error bars representing standard deviation.

TABLE 1. Percentage of correct responses in each of the areas tested

Subject	Percentage in agreement
Gastroesophageal reflux	
Proton pump inhibitor therapy	87
pH/impedance	54
Endoscopy	91
Fundoplication	
Clinical indications for fundoplication	26
Investigations before fundoplication	16
Dysphagia	
Causes of dysphagia in EA patients	15
Presentations of dysphagia	36
Dysphagia post fundoplication	45
Cyanotic spells	
Causes of cyanotic spells	30
Clinical scenario: cyanotic spells	16
Feeding and nutrition	79
Anastomotic stricture	58
Vascular anomalies	29
Eosinophilic esophagitis	96
Transition to adulthood	53

EA = esophageal atresia.

study by Quitadamo et al (6), which evaluated the implementation by pediatricians of the 2009 NASPGHAN/ESPHAGAN GER guidelines, and found acid-suppressive treatment to be over-used amongst general pediatricians in the management of uncomplicated GER in normal infants and children, despite lack of proven efficacy especially in infants (7).

GER is the most frequent GI tract complication with a reported prevalence of 22% to 45% (5). It is associated with gastrointestinal complications, such as peptic esophagitis, recurrent stricture, feeding difficulties, and pulmonary complications, such as atelectasis, aspiration pneumonia, asthma/increased airway reactivity, chronic lung disease with bronchiectasis, and worsening tracheomalacia. Chronic GER can lead to Barrett esophagus and intestinal metaplasia, with a well-documented risk for esophageal adenocarcinoma. In patients with repaired EA because of their inherent esophageal dysmotility, any refluxate that enters the esophagus is poorly cleared because of profound disturbances of peristalsis. Proton-pump inhibitors (PPI) are recommended in all patients with EA for the first year of life, because of their efficacy in reducing reflux symptoms and preventing long-term GER-associated complications. There has been, however, some recent evidence, since the publication of the guidelines which have questioned the severity of acid reflux in these patients and the causal relationship between acid reflux and AS, and esophagitis (8–11). It is important to note, however, that the long-term toxicity of PPIs and their potential adverse effect on respiratory tract infections, bone health, food allergy, and kidney injury as well as changes in the gut microbiome are currently unknown (12). Therefore, there is a need for prospective studies to test this recommendation by evaluating for the presence of pathological GERD with gastroscopy and pH/impedance (where available) testing off PPI treatment at 6 and 12 months of age and by evaluating the benefit of such treatment in the prevention of AS and GER-associated complications.

Role of pH Impedance Monitoring

Guideline statement: All EA patients (including asymptomatic patients) should undergo monitoring of GER (impedance/pH-metry and/or endoscopy) at time of discontinuation of anti-acid treatment and during long-term follow-up (5).

Guideline statement: pH-impedance monitoring is useful to evaluate and correlate nonacid reflux with symptoms in selected patients (symptomatic on PPI, on continuous feeding, with extra-digestive symptoms, ALTE, GER symptoms with normal pH-probe and endoscopy) (5).

pH impedance (MII-pH) testing is a useful investigation to quantify GERD. The advantage of pH impedance monitoring lies in its ability to quantify both acid and nonacid esophageal reflux and to determine symptom association with all reflux episodes. Fifty-four percentage of responders agreed with both of these statements when asked what the role was of 24-hour pH/pH impedance monitoring in EA patients.

Thirty-eight percentage of responders, however, believed that only EA patients with severe esophagitis on endoscopy should have pH monitoring. It is important to note that the utility of impedance is to evaluate the association between clinical symptoms especially extra-esophageal ones, such as ALTE/BRUE with acid and nonacid reflux episodes as well as quantify its proximal migration. As patients with normal endoscopy and biopsy may still have a positive symptom association with extra esophageal symptoms, especially in the cohort with BRUE, it is important not to restrict doing the MII-pH testing to those with reflux esophagitis alone. At the same time, it is also important not to avoid doing MII-pH testing in those with reflux esophagitis on biopsy as having an abnormal biopsy does not necessarily mean that the GER symptoms (which have multifactorial etiologies in the EA patient) are because of reflux. Lastly, it is important to realize that the automated software analysis is often unreliable because of the presence of low baseline in patients with esophagitis or motility disorders as in the EA cohort, which 11% of responders did not recognize.

Guideline statement: Routine endoscopy in asymptomatic EA patients is recommended. The expert panel recommends 3 endoscopies throughout childhood (1 after stopping PPI therapy, 1 before the age of 10 years, and 1 at transition to adulthood) (5).

Ninety-one percentage of responders agreed that endoscopy with biopsies for surveillance is mandatory even in asymptomatic patients and a minimum of 3 endoscopies recommended throughout childhood.

Ten percentage believed that biopsies should only be taken if abnormalities were seen, 7% believed needed only in symptomatic patients.

Routine surveillance in EA patients has demonstrated that up to 80% of patients had moderate-to-severe esophagitis or gastric metaplasia (13). Asymptomatic patients are still at risk of esophagitis with up to a third of symptom-free patients showing mucosal abnormalities in some studies (14).

The goal of regular surveillance is to detect early esophagitis and avoid the development of strictures, Barrett esophagus, and cancer.

Fundoplication

Guideline statement: EA patients may benefit from fundoplication in: recurrent anastomotic strictures, especially in long-gap EA, poorly controlled GERD despite maximal PPI therapy, long-term dependency on trans-pyloric feeding, cyanotic spells (5).

Whilst medical management with a PPI is the first-line approach for those with GERD in EA, there is a subset of patients who might benefit from fundoplication surgery. Responders were

TABLE 2. The average scores with standard deviations for each of the questions and the maximum possible score for each question

Subject	Average score	Maximum score
Gastroesophageal reflux		
Which of these statement(s) about acid suppressive treatment in EA patients in the first year of life are true?	0.9 (0.34)	1
What is the role of 24 hour pH/pH-impedance monitoring in EA patients?	1.5 (0.62)	2
What is the role of upper gastrointestinal endoscopy in the management of EA patients?	0.9 (0.29)	1
Fundoplication		
Which EA patient(s) may benefit from fundoplication surgery?	2.6 (1.08)	4
What investigation(s) should be performed before fundoplication?	2.6 (0.92)	4
Dysphagia		
What are the causes of dysphagia in EA patients?	6.1 (2.56)	10
How do EA patients with dysphagia present?	6.1 (1.94)	8
A 5-year-old long gap EA patient with history of fundoplication presents with new onset of difficulty swallowing, what investigation(s) would you do?	1.35 (0.82)\	3
Cyanotic spells		
What are the causes of extra esophageal symptoms including ''cyanotic spells'' in EA patients?	6.7 (2.85)	10
A 6-month-old type C, EA patient on once a day PPI therapy presents with symptoms of coughing, choking, and bluish discolouration of lips, not always related to feeds. How would you investigate and manage this patient?	3.9 (1.52)	6
Feeding and nutrition		
Which of the following statement(s) is (are) TRUE with regards to feeding and nutrition in EA patients?	1.58 (0.41)	2
Anastomotic stricture		
Which of the following statement(s) is (are) TRUE with regards to anastomotic stricture (AS) in EA patients?	2.4 (0.85)	3
Vascular anomalies		
When should we investigate for vascular anomalies in EA patients?	1.16 (1.17)	4
Eosinophilic esophagitis		
A 12-year-old male EA patient with an atopic history of asthma and food allergies, who has never had an endoscopy previously presents to your rooms with a new 6-month history of symptoms of dysphagia, choking at meal times, and clinical evidence of food bolus impactions. You do an upper GI contrast study, which does not show a stricture. What is an important diagnosis to exclude in this patient, which can only be confirmed with an endoscopy with biopsies?	0.96 (0.20)	1
Transition		
You see an 18-year-old EA patient in your outpatient clinic/rooms. His parents ask whether he still needs to see a doctor in adulthood. What do you tell them?	2.3 (0.82)	3
Overall	43 (9.9)	62

EA = esophageal atresia.

asked, ''Which EA patients may benefit from fundoplication surgery?''

Of all the above indications, 94% agreed with fundoplication for poorly controlled GERD with persistent symptoms and or reflux esophagitis on maximal PPI therapy, 68% with an acute life-threatening event (ALTE)/BRUE shown to be secondary to GERD, 62% for those with long-gap EA with recurrent AS and 41% for those with dependency on trans-pyloric feeds. Eleven percentage believed it was indicated for chronic cough—despite data indicating that fundoplication has not been shown to protect against respiratory symptoms (15).

Only 26% of responders agreed with all of these indications for fundoplication (Table 1), which likely represents paucity of data on long-term postsurgical outcomes to inform management. There remains a lack of multicenter controlled studies evaluating the outcomes of fundoplication in these patients.

Guideline statement: Barium-contrast study, endoscopy with biopsies, and pH-metry should at least be performed before fundoplication (5).

Before performing a fundoplication, it is necessary to perform investigations to justify the need for antireflux surgery. Investigations recommended include, pH with or without impedance depending on availability at the individual center is required to quantify acid and nonacid reflux, proximal migration of reflux episodes and symptom association, barium contrast study to look for hiatal hernia, associated congenital stenosis, assessment of the gastric cardiac region, exclusion of microgastria, and other intestinal malformations including malrotation and pyloric stenosis and endoscopy to screen for peptic esophagitis and Barrett esophagus and to exclude EoE. Exclusion of EoE is especially important as its symptoms may mimic GERD and not all patients with EoE respond to PPI therapy resulting in persistent symptoms despite being on maximal PPI therapy. When asked, which investigations should be performed before fundoplication, only 15% of responders agreed that all 3 investigations were required.

Thirty-five percentage of responders also indicated that an esophageal manometry was required before fundoplication. Whilst there is a paucity of evidence currently regarding role manometry

before fundoplication in EA patients with GERD, there is some evidence that high-resolution manometry combined with impedance may be predictive of postfundoplication dysphagia. One study using high-resolution impedance manometry and ''Pressure Flow Metrics'', has indicated that elevated clearance pressure and/or pressure-flow indexes may predict postoperative dysphagia in children undergoing fundoplication (16).

The low concordance between the surveyed population with the ESPHAGAN/NASPGHAN guidelines likely represents the lack of understanding about multifactorial etiology for esophageal and extra-esophageal symptoms in the EA cohort in addition to an underestimation of the increased risk of complications because of a dysmotile esophagus resulting in poor bolus clearance post antireflux surgery in this population.

There is a need for future research comparing outcomes in children with and without esophageal atresia post fundoplication to assess true outcomes, symptom resolution, complication rates, and postoperative quality of life.

Dysphagia

Dysphagia is common in repaired EA patients with incidence quoted between 21% and 84% and is associated with a lower quality of life (17). It can be present in up to 48% of patients 10 years post surgical repair (18). Correct identification of the etiology of dysphagia can facilitate diagnosis and management in these patients. It has a multifactorial etiology, and can result from AS, peptic esophagitis, EoE, congenital stenosis, peptic stricture, postfundoplication, vascular anomalies, anastomotic diverticulum, mucosal bridge, and inlet patch. Only 15% of responders correctly identified all of these causes of dysphagia. Not surprisingly, 97% identified that AS is a cause of dysphagia. This highlights the need for increased recognition that a broad range of etiologies in addition to AS can cause dysphagia in the EA patient. This is particularly relevant to this cohort because of the negative impact that dysphagia can have on feeding, nutrition, and growth.

Guideline statement: Dysphagia should be suspected in patients with EA who present with food aversion, food impaction, and difficulty in swallowing, odynophagia, choking, cough, and pneumonia, alteration in eating habits, vomiting, and malnutrition (5).

Dysphagia is often difficult to diagnose in children (5). ESPHAGAN/NASPHGAN guidelines highlight the fact that dysphagia can present as food aversion, choking, cough, pneumonia, alteration in eating habits, vomiting and malnutrition in addition to the more common presentations of food impaction, difficulty swallowing, and odynophagia.

Only 36% of responders indicated that dysphagia should be suspected in EA patients with any of these symptoms. Difficulty swallowing, food aversion, and coughing/choking during swallowing were the most well recognized symptoms and pneumonia was the least recognized, likely as AS is the most well-known cause of dysphagia.

Guideline statement: We recommend that all EA patients with dysphagia undergo evaluation with upper GI.

Contrast Study and Esophagoscopy With Biopsies

Responders were also presented with a clinical scenario of a 5-year-old with long-gap EA with a history of fundoplication who presented with dysphagia and were asked, which investigations they would do (5). Eighty-nine percentage chose barium contrast study, 80% endoscopy with biopsies, and 51% esophageal manometry with or without pH impedance. In total, only 45% of responders identified that all investigations have a role in work-up of dysphagia in an EA patient post fundoplication. A barium contrast study has the potential to show strictures, hiatal or para-esophageal hernias, and a slipped or unwrapped fundoplication. An endoscopy can demonstrate EoE or peptic or fungal esophagitis. Finally, manometry can show hold-up at the level of the fundal wrap and a pH-impedance can demonstrate symptom association with reflux events. All these investigations are useful in investigating causality as this can inform specific treatment.

Extra-esophageal Symptoms

Guideline statement: The etiology of life-threatening events is multifactorial and merits a multidisciplinary diagnostic evaluation before surgical intervention (5).

Guideline statement: Anatomic issues (strictures, recurrent or missed fistulae, congenital esophageal stenosis, vascular rings, and laryngeal clefts) and aspiration need to be excluded in children with ALTE (5).

Extra-esophageal symptoms including cyanotic spells and BRUEs in this cohort can have a multifactorial etiology. Only 30% of responders, however, correctly identified that direct aspiration during swallowing because of laryngeal clefts/vocal cord paralysis, reflux aspiration, esophageal AS, congenital esophageal stenosis, esophageal dysmotility, recurrent or missed trachea esophageal fistula, EoE, esophageal pooling over fundoplication wrap, food retention in proximal pouch in esophagus and vascular rings can all cause cyanotic spells.

Responders were presented with a clinical scenario where a 6-month-old type C EA patient on daily PPI therapy presented with coughing, choking, and bluish discoloration of their lips, not always related to feeding. When asked how they would investigate and manage this patient, only 16% correctly identified that pH-impedance, upper GI contrast study, modified barium swallow, gastroscopy with biopsies, laryngo-bronchoscopy, and a multidisciplinary team review were all appropriate, despite the multifactorial etiology for these symptoms in an EA patient. This poor concordance is likely to be secondary to a lack of multidisciplinary program in all tertiary pediatric centers managing EA patients, which limits the discussion of these complex patients amongst the various specialists involved in their care.

Feeding and Nutrition

Guideline statement: No data are available on the most efficacious methods of avoiding feeding disorders in children with EA. However, the committee recommends a multidisciplinary approach to prevent and treat feeding difficulties (5).

Guideline statement: Intensive early enteral and oral nutrition intervention and advances in neonatal care and surgery have reduced the risk of long-term malnutrition in children with EA; however, other associated comorbidities may increase this risk (5).

Seventy-nine percentage of responders correctly identified that early enteral nutrition intervention reduced the risk of malnutrition and a multidisciplinary approach is needed to prevent and treat feeding difficulties. Early enteral nutrition postsurgical repair in EA patients results in reduced sepsis and reduced hospital stays (19,20). There are multiple causes of feeding difficulties in children with EA—including GERD, EoE, aspiration, dysphagia, and AS. Whilst no studies address how to prevent or treat abnormal feeding behaviors, given the broad etiology, it is recommended that this is addressed in an MDT setting.

Anastomotic Strictures

Guideline statement: In addition to relative esophageal narrowing at the level of the anastomosis (by contrast and/or endoscopy), significant functional impairment and associated symptoms need to be present for anastomotic strictures to be considered clinically significant (5).

AS is one of the most common postoperative complications post EA repair (21). ESPGHAN/NASPGHAN guidelines state that AS should be considered clinically relevant and dilated only in patients with symptoms. These symptoms are feeding and swallowing difficulties, regurgitation and vomiting, mucus and food retention in the proximal pouch, cough, drooling, recurrent respiratory infections, foreign body impaction, and poor weight gain. There is currently a dearth of quality evidence that routine screening and dilatation is superior to evaluation and stricture dilation only in symptomatic patients.

Responders were asked to identify true statements in regards to AS in EA. Fifty-eight percentage responded that they agreed that only strictures associated with clinical symptoms are clinically relevant, strictures detected during the evaluation of symptomatic patients should be dilated and those detected during evaluation of EA children who are unable to achieve feeding milestones should be dilated. Seven percentage of responders indicated that all AS detected on routine screening, even in asymptomatic patients need to be dilated and 5% that all AS seen on contrast studies are clinically relevant.

There remains a lack of good-quality evidence regarding AS management in EA children. Not only is AS difficult to diagnose clinically given its similar clinical picture with esophageal dysmotility, recurrent fistula, GERD, tracheomalacia, laryngeal cleft, and vocal cord dysfunction but also there is no evidence to support ideal interval between dilatation sessions (21). There has been 1 retrospective study by Koivusalo et al indicating that dilatations in only those that are symptomatic versus dilating all strictures seen in contrast studies results in a reduction in dilatations without a difference in clinical outcomes. With known complications of perforation, hemorrhage, and airway compression, it is thought that restricting stricture dilation to symptomatic EA patients with strictures would improve outcomes and result in a reduction in routine dilatations with associated risk of complications.

Vascular Anomalies

Guideline statement: Even though congenital vascular malformations are usually asymptomatic, they may be the underlying etiology for dysphagia, dyspnea, or cyanosis, by causing external compression on the esophagus and/or trachea. We recommend that congenital vascular malformations be excluded in these situations by chest CT or MR angiography (5).

Vascular anomalies have been shown to be present in up to 18% of children with EA (22). These abnormalities may be the cause of respiratory symptoms, such as dyspnea, cough, and cyanosis in EA patients and may exacerbate symptoms, such as dysphagia. ESPGHAN/NASPHAN guidelines recommend that congenital vascular malformation be excluded in those with dysphagia, dyspnea, or cyanosis. This is particularly relevant before stenting because of the risk of anomalous right subclavian artery-esophageal fistulas.

Less than 50% of responders agreed vascular anomalies should be investigated before stenting despite there being evidence that severe gastrointestinal hemorrhage secondary to fistula formation after stenting or prolonged nasogastric tube placement is a potential life-threatening complication of failing to investigate for vascular anomalies (20). Fifty-three percentage, however, agreed that vascular anomalies be investigated for in patients with dyspnea, dysphagia, and cyanotic spells.

Eosinophilic Esophagitis

Guideline statement: EoE needs to be excluded in EA patients of all ages with dysphagia, reflux symptoms, coughing, choking, or recurrent strictures that are refractory to PPI and before proceeding to antireflux surgery (5).

The association between EoE and EA was first published in a retrospective case study by Dhaliwal et al (23), where an incidence of 17% was seen. This has been corroborated in multiple subsequent studies, which have also shown an increased prevalence of EoE in the EA cohort compared with the general pediatric population (24), although there is limited case-control data on true prevalence.

Responders were presented with a clinical scenario of a 12-year-old boy with an atopic history of asthma and food allergies who has never had an endoscopy who presented with a 6-month history of dysphagia, choking at meal times, and clinical evidence of food bolus impactions. An upper GI contrast study has shown no stricture. They were asked, which diagnosis was important to exclude, which would require an endoscopy with biopsies to confirm the diagnosis. Ninety-six percentage of responders identified EoE as the most likely diagnosis, with 13% identifying GERD and 12.2% esophageal dysmotility as the likely diagnosis, suggesting that most clinicians are aware of the EoE-EA association.

Transition of Esophageal Atresia Patients

Guideline statement: We recommend transition of young adults from pediatric care to an adult physician with expertise in EA (general practitioner, surgeon, gastroenterologist, pulmonologist, or any informed specialist aware of the specificities of the care of adults operated for EA) (5).

With ever improving mortality and morbidity, it is becoming more important to ensure EA patients have a smooth transition into the care of adult physicians with experience in EA. Case reports have suggested that EA patients might be at a higher risk of Barrett and esophageal carcinomas at a younger age (25). A clinical scenario was given of an 18-year-old patient asking if he needs to be followed up in adulthood. Ninety-one percentage of responders agreed that regular endoscopy with biopsy at the time of transition and every 5–10 years if asymptomatic is indicated. Seventy-one percentage of responders agreed that follow-up with an adult physician trained in EA to monitor for dysphagia, GERD, respiratory symptoms, and anemia and 68% that esophageal cancer remains a concern in EA patients in adulthood. Only 53% of responders, however, identified all 3 answers.

Study Limitations

This study had some limitations. Firstly, demographic data was limited to location, and information regarding specific role (type of specialist doctor, allied health, nursing staff, other) was neither collected nor was years of experience looking after EA patients. Information regarding the size of the center the professional worked and the number of EA patients treated at the center per year was also not collected. This would have provided interesting information on guideline concordance between different professionals at centers of varying sizes. Secondly, there was no survey conducted before the publication of the ESPGHAN/NASPGHAN guidelines, which would have provided interesting information on whether publication of the guidelines improved their understanding of the gastrointestinal and nutritional complications in the EA

cohort. The survey also did not look to see whether these specialists were adhering to these guidelines in their day-to-day management of these complex EA patients. It is expected that a future survey will address these limitations, and which will specifically look at adherence to these guidelines in the care of EA patients.

CONCLUSIONS

The ESPGHAN/NASPGHAN Guidelines for the Evaluation and Treatment of Gastrointestinal and Nutritional Complications in Children with Esophageal Atresia-Tracheoesophageal Fistula were the first EA guidelines published in 2016. This study evaluated the awareness and understanding of these guidelines amongst expert clinicians involved in the care of these patients, by the means of multiple-choice questions and clinical scenarios based on the recommendations/statements in the published guidelines. Overall, this study showed that specialist health professionals involved in the care of children of EA had good concordance with the ESPGHAN/NASPGHAN Guidelines (70%), and there was no difference in guideline concordance between continents. There was also a good response rate with a 67% uptake of the survey amongst interest groups. We found that, responders scored highly on the role of endoscopy in EA, role of acid suppression, and link between EA and EoE. Of concern is the fact that responders overall scored lower in the questions on importance of thorough investigations before fundoplication, multifactorial causes, and presentation of dysphagia and extra-esophageal symptoms. With respect to treatment of AS, just over half (58%) agreed that only symptomatic patients with AS need to be dilated, which reflects a paucity of evidence in this field, comparing routine dilation with on-demand dilation. The low concordance with societal recommendations in certain areas could reflect either a poor understanding of the reasoning behind specific recommendations, or a lack of awareness of the guidelines. However, some of the responses may also have been because of a paucity of high-quality evidence behind some of the recommendations in the guidelines. There is a need for making more clinicians aware of these guidelines and also further research and prospective collaborative multicenter projects in these areas to strengthen the evidence behind the recommendations/statements in current ESPGHAN/NASPGHAN guidelines.

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