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Characterization of Esophageal Motility Disorders in Children Presenting With Dysphagia Using High-Resolution Manometry

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Abstract

Purpose of Review The Chicago classification was based on metrics derived from studies in asymptomatic adult subjects. Our objectives were to characterize esophageal motility disorders in children and to determine whether the spectrum of manometric findings is similar between the pediatric and adult populations.

Recent Findings Studies have suggested that the metrics utilized in manometric diagnosis depend on age, size, and manometric assembly. This would imply that a different set of metrics should be used for the pediatric population. There are no standardized and generally accepted metrics for use in the pediatric population, though there have been attempts to establish metrics specific to this population.

Summary Overall, we found that the distribution of esophageal motility disorders in children was like that described in adults using the Chicago classification. This analysis will serve as a prequel to follow-up studies exploring the individual metrics for variability among patients, with the objective of establishing novel metrics for the pediatric population.

Keywords Esophageal motility disorders · Dysphagia · Manometry · Pediatric gastroenterology · Chicago classification

Introduction

The Chicago classification of esophageal motility disorders has been largely utilized in the diagnosis of esophageal diseases in the adult and pediatric population and was based on metrics derived from studies in asymptomatic adult subjects. These metrics have been established to be dependent on age, size, and manometric assembly [1–4]. Esophageal motility disorders in the pediatric age group can often present a diagnostic challenge. This is largely because of children's inability to accurately describe their symptoms as well as the nonspecificity of symptoms frequently associated with impaired esophageal motility [5–7]. A lot of progress has been made with the use of high-resolution manometry with esophageal pressure topography (HRM-EPT) in the evaluation of esophageal motility disorders, but most of the studies have been in the adult population [8–12]. The greatest limitation to obtaining similar metrics in the pediatric population is the ethical considerations involved in subjecting healthy children to the rigors of testing with no immediate benefit to them. Hence, there is a dearth of studies specifically characterizing motility disorders in the pediatric population.

Furthermore, there are no studies that have attempted to characterize esophageal motility disorders in the pediatric population using the current Chicago classification. The prevailing belief based on scant evidence is that the spectrum of esophageal motility disorders seen in children is like what has been described in adults [6, 13, 14].

Although the clinical implications of some esophageal motility disorders, such as achalasia, are clear, there is

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considerable controversy over the significance of other esophageal motility abnormalities found on esophageal manometry testing and whether the patient's symptoms can be attributed to these findings. In adults with achalasia, type I (characterized by 100% failed peristalsis and lower esophageal sphincter (LES), integrated relaxation pressure (IRP) greater than 15 mmHg) is known to be better treated with Heller myotomy than balloon dilation, and type II (characterized by pan-esophageal pressurization and IRP greater than 15 mmHg) responds well to both Heller myotomy and balloon dilation, while type III (characterized by spastic contractions with distal latency >4.5 s and IRP >15 mmHg) has an overall poor prognosis irrespective of treatment option pursued [15–17].

The clinical utility of esophageal manometry studies, high resolution (HRM) or conventional, is in demonstrating contraction patterns that deviate from what has been described in asymptomatic subjects, and in suggesting whether these abnormalities correlate with symptoms and can ultimately guide patient management.

Esophageal manometry has been used for several decades to diagnose esophageal motility disorders. The advent of high-resolution manometry (HRM) with esophageal pressure topography (EPT) has improved the diagnostic accuracy and characterization of esophageal motility disorders.

High-Resolution Manometry vs Conventional Manometry

The fundamental difference between conventional manometry and HRM is the number of sensors used and the spacing between them. In contrast to conventional manometry, in which sensors are spaced 3–5 cm apart, HRM sensors are typically spaced 1 cm apart along the length of the manometric assembly [18]. The HRM with EPT assembly is used to characterize accurately the esophagogastric junction (EGJ) and esophageal body peristalsis.

Using conventional esophageal manometry, primary esophageal motility disorders were classified as achalasia and "other" abnormal motility patterns, which are further sub-classified as hypercontracting, hypocontracting, or discoordinated motility. On the other hand, the Chicago classification based on HRM and EPT utilizes a hierarchical approach, sequentially prioritizing (i) disorders of esophagogastric junction (EGJ) outflow (achalasia subtypes I–III and EGJ outflow obstruction), (ii) major disorders of peristalsis (absent contractility, distal esophageal spasm, hypercontractile esophagus), and (iii) minor disorders of peristalsis characterized by impaired bolus transit [19–21].

The recent updates in the new version of the Chicago classification (v3.0) incorporated some changes in the algorithm for the diagnosis of esophageal motility disorders. For instance, the category "minor disorders of peristalsis" was renamed, small breaks (2–5 cm) in the 20-mmHg isobaric contour was eliminated as a criterion for abnormality, and contractile front velocity (CFV) was eliminated because the clinical relevance of these metrics is uncertain. Other changes included elimination of the designation of hypertensive peristalsis (distal contractile integral (DCI) 5000–8000 mmHg.s.cm) and frequent failed peristalsis as a distinct diagnostic entity. The new version also adopted the term "ineffective esophageal motility" (IEM), defined by poor bolus transit in the lower esophagus [19–22]. No distinction was made between failed swallows and weak swallows, thereby getting rid of the term "frequent failed peristalses."

Clinical Presentation of Motility Disorders in Children

Symptoms of abnormal esophageal motility in children are like those in adults, although for the younger children, symptoms are usually as perceived and reported by the caregiver. The range of symptoms is broader for the pediatric population than the adult population. Presenting complaints can range from chest pain and dysphagia to pyrosis, regurgitation, heartburn, nausea, vomiting, diarrhea, constipation, or even change in feeding. Poor eating can potentially lead to malnutrition and weight loss. Similarly, chronic cough and recurrent pneumonia can sometimes be attributed to an underlying esophageal motility disorders.

Methods

High-resolution manometry with esophageal pressure topography recordings of liquid swallows of patients <21 years old seen at the Neurogastroenterology and Motility Disorders unit at Cincinnati Children's Hospital Medical Center (CCHMC) were extracted from a database of studies conducted between April 2011 and May 2015. Studies were reviewed to ensure optimal recording quality of at least ten liquid swallows. Access to patient charts was approved by the institutional review board.

Manometry Protocol

A 3.2-mm-diameter solid-state manometric and impedance catheter incorporating 36 pressure sensors spaced 1 cm apart with 12 adjoining impedance segments (Unisensor USA Inc., Portsmouth, NH, USA) was used. Pressure and impedance data were acquired at 20 Hz (Solar GI acquisition system, MMS, Enschede, USA). Patients were intubated after application of topical anesthesia (lidocaine spray or gel). Some patients were directly intubated in the endoscopy suite during upper endoscopy. Studies were performed with the patients sitting or being held in the semi upright positions. Testing was performed with ten boluses of saline (0.9% saline at room temperature). Boluses of 5 to 10 ml were administered with a syringe at intervals of 20 s between swallows.

Data Analysis

All tracings were manually scanned for artifacts and studies of poor quality were excluded. Tracings were analyzed with MMS automated analysis software version 8.23. Swallowing onset was determined by upper esophageal sphincter (UES) relaxation. Standard EPT metrics were obtained using the analysis software after manually adjusting to localize accurately landmarks such as upper esophageal sphincter, gastric lower esophageal sphincter, pressure inversion point, and crural diaphragm. EPT metrics utilized included distal contractile integral (DCI), distal latency (DL), 4-s integrated relaxation pressure (IRP4s), and peristaltic 20-mmHg isobaric contour break size (BS).

Manometric diagnosis was based on ten swallows, using the established hierarchical Chicago classification algorithm. Per this algorithm, the first assessment of esophageal motility is whether there is EGJ outflow obstruction as defined by IRP. Disorders of EGJ obstruction are then further classified into achalasia subtypes and EGJ outflow obstruction. The next step was to look for major disorders of peristalsis, which include distal esophageal spasm (defined as normal IRP and ≥20% of premature contractions distal latency <4.5); hypercontractile esophagus (defined as ≥20% of swallows with a DCI >8000); and absent contractility (defined as normal EGJ relaxation with 100% failed peristalsis). The final step was to look for minor disorders of peristalsis classified as ineffective esophageal motility, defined by a DCI <450 mmHg-s-cm with ≥50% ineffective swallows.

Patients who did not demonstrate any of these disorder subtypes were classified as normal [10, 19].

Statistical Analysis

Data were analyzed using the Statistical Package for the Social Sciences (SPSS) version 20 (SPSS Inc., Chicago, IL, USA). Statistical analysis was purely descriptive. Parametric data are expressed as mean ± SD and non-parametric data are expressed as median (range).

Results

A total of 137 manometric studies were extracted from our database. Of these studies, six were excluded due to age >21 years or inadequate study. Patient characteristics, such as age, gender, body mass index, and presenting symptoms, are displayed in Table 1; 54.2% ($n = 71$) were male and 45.8% ($n = 60$) were female; 80.9% ($n = 106$) had two or more symptoms. The most commonly reported comorbid condition was allergic diathesis in 34.4% ($n = 45$). Others were neurodevelopmental delay 24.4% ($n = 32$), eosinophilic esophagitis 11.5% ($n = 15$), and Nissen fundoplication 10.7% ($n = 14$). Esophageal manometry was defined as normal in 43.5% ($n = 57$), abnormal in 54.2% ($n = 71$), and inadequate in 2.3% ($n = 3$). Of those with abnormal manometry, 35.9% ($n = 28$) of cases had normal EGD, while 57.7% ($n = 41$) had abnormal findings at EGD as well as manometry. EGJ outflow obstruction was reported in 5.3% ($n = 7$) of cases. The most common esophageal body peristalsis disorder was IEM in 22.9% ($n = 30$), followed by absent peristalsis in 16.8% ($n = 22$), DES 7.6% ($n = 10$), and hypercontractions 4.6% ($n = 6$). There was one case of non-relaxing upper esophageal sphincter. Achalasia was present in 11.5% ($n = 15$), with type 2 being the most common (7.6%, $n = 10$) followed by type 1 (3.1%, $n = 4$) and type 3 (0.76%, $n = 1$) achalasia.

Table 1 Patient characteristics of our study population

Patient characteristics	Number
Median age in years (range)	12 (1–21)
Median BMI (range)	17.54 (10.76–46.92)
Number male (%)	71 (54.2)
Number female (%)	60 (45.8)
Presenting symptoms	
• Dysphagia (%)	87 (66.4)
• Nausea (%)	59 (45)
• Vomiting (%)	54 (41.2)
• Weight loss (%)	43 (32.8)
• Regurgitation (%)	42 (32.1)
• Abdominal pain (%)	31 (23.2)
• Diarrhea or constipation (%)	27 (20.6)
• Feeding difficulty (%)	17 (13.0)
• Chest pain (%)	16 (12.2)
• Cough (%)	3 (2.3)
Comorbid conditions	
• Eosinophilic esophagitis (%)	15 (11.5)
• Neurodevelopmental delay (%)	32 (24.4)
• Hiatal hernia (%)	5 (3.8)
• Allergic diathesis (%)	45 (34.4)
• Tracheoesophageal fistula (%)	2 (1.5)
• Status post Nissen fundoplication (%)	14 (10.7)

Table 2 characterizes the esophageal motor disorders using the Chicago classification algorithm.

Discussion

This study describes commonly encountered esophageal motility disorders in children presenting with dysphagia. We

Table 2 Diagnoses based on Chicago Classification

Findings	*N* studies (%)
Achalasia subtype	
• Type 1	4 (3.1)
• Type 2	10 (7.6)
• Type 3	1 (0.8)
EGJ outflow obstruction	7 (5.2)
Distal esophageal spasm	10 (7.6)
Jackhammer esophagus	6 (4.6)
Ineffective esophageal motility	30 (22.9)

characterized esophageal motility findings in 131 patients using HRM with EPT recordings and the Chicago classification algorithm. There were more males than females with dysphagia but this difference was not statistically significant. The spectrum of symptoms reported is similar to what has been described in other smaller studies with children [23, 24] with the caveat that symptoms are mostly as perceived by the caregiver because the very young children are unable to characterize their symptoms. The most commonly identified abnormality was ineffective esophageal motility. This diagnosis has been documented in adults as having very good prognosis with little or no worsening of symptoms after initial diagnosis [25]. Achalasia was also demonstrated in children, with type 2 being the most common and type 3 being the least common. One patient had a non-relaxing UES, and this has not been reported in any other studies to the best of our knowledge. Allergic diathesis was the most common comorbid condition in our study population, although there was no statistically significant relationship between this and any specific esophageal manometric abnormality. At least one study has reported increased incidence of abnormal esophageal manometry in children with allergic diathesis [26]; 43.5% of the patients had normal manometry findings even in the presence of symptoms and the absence of obstructive or neuromuscular swallowing disorders. This raises an important question about applying adult metrics from the Chicago classification in the evaluation of the pediatric population. The Chicago classification was based on normative data of healthy adult controls and has been broadly applied to the pediatric population. It is well recognized that children are not "little adults," and any evaluation or care involving them must take into cognizance their weight, body surface area, and other characteristics [27]. One study has proposed adjustment of the distal contractile integral (DCI) for esophageal length, while a second study suggested adjustment of distal latency (DL) and 4-s integrated relaxation pressure (IRP4s) for age and esophageal length but did not find any relationship between lower DCI values and either younger age or shorter nares to EGJ length [1, 3]. Our study did not seek to make any adjustments for age or esophageal length as there are currently no standard validated metrics based on age or esophageal length. More studies are needed with respect to this issue, to improve the diagnostic accuracy of HRM with EPT in the pediatric population. Even though there remains a pressing need to define age-specific metrics for diagnosing esophageal motility disorders in healthy children, this may not be ethically possible.

Next Steps

A few studies have tried to look at the variations and changes that occur in diagnoses when metrics were adjusted for age and esophageal length. However, a standardized and generally accepted way to accurately measure these metrics, taking into consideration the unique features in children, does not yet exist. Once we acquire more data in children, there will need to be an expert, evidence-based consensus to design a pediatric version of the software.

Conclusion

This study was descriptive and aimed at documenting the commonly observed esophageal manometry findings in children. As is the case in numerous studies in the adult population, disorder of esophageal body peristalsis, notably ineffective esophageal motility, was the most commonly observed abnormality in our cohort. As in adults, achalasia was also notable, with type 2 being the most common.

Study Limitations

The study used the Chicago classification, which was developed from data acquired from a healthy adult population, and may not be directly applicable to children. We did not attempt to make any adjustments in the metrics for age and size. Symptoms reported in the study were sometimes as perceived by the caregiver in instances where the patient could not verbalize symptoms.

Author Contribution Ajay Kaul: Study concept and design, acquisition of data, interpretation of data, revision of abstract and manuscript for intellectual content, study supervision.

Adeel Malik: Data collection.

Francis Edeani: Data collection, data analysis, statistical analysis, initial draft of abstract and manuscript.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest. Ajay Kaul, MD, is a speaker for Laborie.

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