



A 12 Year Experience with Conventional and High Resolution Esophageal Manometry in Children

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Abstract

Objectives: Once anatomical causes are ruled-out, esophageal manometry is the test of choice for diagnosis for esophageal dysmotilities in children. There are currently two types: water-perfused conventional manometry (conventional EM) and high-resolution esophageal manometry (HREM). HREM has been introduced recently and has become the standard of care. There is scarcity of information on pediatric esophageal dysmotilities and therefore, the aim of our study was to observe and describe the etiologies of these disorders in children at our center over a period of 12 years.

Methods: We retrospectively reviewed medical records of a cohort of patients who underwent esophageal manometry studies from January 2001 to June 2013. We recorded age, gender, year of study, indication and results.

Results: A total of 94 subjects were selected for our study. The median age of children in our study was 13 years. In our cohort, the most common indication for manometry was dysphagia (80%). We found that 38% of our subjects had normal motility. The most frequently diagnosed conditions were achalasia (22%) and ineffective esophageal motility disorder/minor peristaltic abnormalities (HREM) (22%). Other diagnoses included hypotensive LES, found in 8%, esophageal spasms found in 4%, and hypertensive LES/EGJ outflow obstruction (HREM) which was found in 4% of subjects. Two percent of our subjects had scleroderma, while 0% was found to have hypertensive peristalsis of the esophagus (Nutcracker esophagus/Jackhammer esophagus).

Conclusion: As in adult studies, our observation in children revealed that achalasia and ineffective esophageal motility disorder/minor peristaltic abnormalities (HREM) accounted for most of the motility disorders that were detected. Further studies will be useful to investigate the characteristics of patients diagnosed with these disorders.

Keywords: Dysmotilities; Dysphagia; Esophageal manometry; Achalasia

What is known about the Subject?

- Esophageal dysmotility disorders can cause nutritional deficiencies, weight loss and feeding problems in children.
- Esophageal manometry is the gold standard for diagnosis for esophageal motility disorders.

What is New?

- The frequency with which these disorders occur in children is not well addressed in the literature.
- We aimed to determine the frequency of these diagnoses in a group of children undergoing esophageal manometry at our institution.
- We now have an idea of the demographics of esophageal dysmotility disorders in the pediatric population at our practice.

Introduction

Esophageal dysmotility is an uncommon disorder usually presenting with dysphagia. Population-based studies have estimated the prevalence at 0.25/100,000 in young adults [1], but

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Received Date: 24 Oct 2017

Accepted Date: 10 Dec 2017

Published Date: 17 Dec 2017

Citation:

Waseem S, Jafri FW, Kisseih ED, Erum Rizvi BS, Bennett Jr WE, Croffie J. A 12 Year Experience with Conventional and High Resolution Esophageal Manometry in Children. *J Gastroenterol Hepatol Endosc.* 2017; 2(6): 1033.

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Table 1: Demonstrates the demographics associated with the results of WP versus HRM.

	WP	HRM	p-value
N	58	36	
Age			
Mean	12.0	12.7	0.37
Median	13	13.5	
< 5 years	1	0	
5-9 years	16	11	
10-14 years	21	9	
15-18 years	20	16	
Gender, # females (%)	28 (49%)	18 (50%)	0.71
Race			0.93
White	52 (90%)	32 (89%)	
Black	3 (5%)	2 (6%)	
Asian	1 (2%)	1 (3%)	
Native American	1 (2%)	0 (0%)	
Unknown/Other	1 (2%)	1 (3%)	
Dysphagia as primary presenting complaint	49 (84%)	36 (75%)	0.39
Final diagnosis			0.04 (all)
Normal	26 (45%)	10 (28%)	0.11 (just abnormal)
Abnormal	32 (55%)	26 (72%)	
Achalasia	10 (17%)	10 (28%)	
Esophageal spasms	4 (7%)	1 (3%)	
Hypertensive LES	1 (2%)	4 (11%)	
Hypotensive LES	1 (2%)	4 (11%)	
Nonspecific Esophag. Motility Disorder	14 (24%)	7 (20%)	
Scleroderma	2 (3%)	0 (0%)	

the prevalence in children is not well known. It occurs as a result of disorganized esophageal muscular activity and can have multiple etiologies. Esophageal manometry is a diagnostic test used to evaluate the function of the esophageal muscles in patients presenting with difficulties with deglutition once anatomic and inflammatory disorders are ruled out.

There are currently two types of equipment used to record intraluminal pressure activity in the esophagus: water-perfused EM and solid state. The water-perfused system utilizes a pneumohydraulic pump to perfuse the manometry catheter with water through capillary tubes within the wall of the catheter. Pressure changes in the columns of water exiting side holes along the catheter, and generated by lumen-occluding contractions of the esophageal body and sphincters are referred to external transducers which convert these into electrical signals that are then displayed as pressure waves on a computer monitor. The solid state system uses catheters which have internal micro-transducers made from metal diaphragm strain gauges or piezoresistive silicon chips [2]. Pressure changes detected by these micro-transducers are transmitted along the catheter to a computer and displayed as pressure graphs on a monitor. High resolution manometry (HREM) is a more recent and advanced solid state system which utilizes a catheter that has multiple miniaturized strain gauge sensors that are closely spaced together, allowing for detection of rapid pressure changes and therefore more detailed pressure mapping of esophageal motor function. Whereas the conventional water perfused and solid state catheters have only 4 to 8

longitudinal pressure monitoring ports, the high resolution catheter may have as many as 16 to 36 longitudinally and circumferentially positioned pressure monitoring sites. Because the high-resolution manometry catheter has multiple sensors, it allows for simultaneous recording from the pharynx to the lower esophageal sphincter and beyond, without the need for station pull-through as is necessary with the conventional solid state and water-perfused catheters. Specific abnormalities traditionally diagnosed with esophageal manometry can be classified under 4 main categories [3]: 1) Disorders of lower esophageal sphincter (LES) relaxation, which includes achalasia and atypical disorders of LES relaxation; 2) Uncoordinated contractions in the esophageal body 3) Hypertensive contractions of the esophageal body; and 4) Hypotensive contractions of the esophageal body. The frequency with which these disorders occur in children undergoing esophageal manometry is not well addressed in the literature. The primary aim of this study, therefore, was to determine the frequency of these diagnoses in a group of children undergoing esophageal manometry at our institution. The secondary aim was to compare results obtained with the conventional water perfused system that we previously used and results obtained with the high-resolution system which we currently use, to determine if there are any significant differences.

Methods

This was a retrospective study of children between the ages of 0 and 18 years old who underwent esophageal manometry from

January 2001 to December 2013 at our large, tertiary care center, located in Indianapolis, IN. Data were collected after obtaining Institutional Review Board (IRB) approval from Indiana University. Inclusion & Exclusion criteria: All children aged 0-18 who underwent diagnostic esophageal manometry from January 2001 to December 2013 were included in our study. Exclusion criteria: Absence of pertinent medical record and age over 18 years. Medical charts were reviewed and patients were assigned to one of two groups: 1) those who underwent conventional EM and 2) those who underwent HREM. Data collected for each patient included age, race, the year of study, the indications for the procedure and the results. We assessed all patients and did not eliminate any based on co-morbid conditions. All patients had sufficient data for analysis, so no patients were excluded due to incomplete data. Conventional EM was performed using an eight channel low compliance polyvinyl catheter assembly with 4 pressure ports located circumferentially 1 cm from the tip and 4 pressure ports located longitudinally along the catheter and separated by 5 cm. Patients were unsedated, but topical anesthesia was used to anesthetize the nasal passage through which the catheter was passed. All patients were fasting at the time of the procedure. Patients were positioned semi-upright. The catheter was perfused with sterile water by a low compliance pneumohydraulic pump (Andorfer Medical Specialities, Greendale, WI) at a rate of 0.5 ml/min. The station pull-through method was used to identify the sphincters [4]. Patients were given 10 to 15 5-ml boluses of water to swallow at 30 second intervals. Analysis was performed using the GIPC™ software (REDTECH, Calabasas, California). For assessment of LES, we used LES resting pressure and LES residual pressure following swallows. For assessment of UES, we used UES resting pressure and UES residual pressure following swallows. For the esophageal body, we used mean distal and proximal esophageal pressure amplitudes with swallows. For distal esophagus, the distal pressure port was placed at 3 cm above the LES while for the proximal esophagus the proximal pressure port was placed 1 cm below the UES. The average velocity of contractions at these sites was also determined. We used atmospheric pressure at zero for reference. Conventional EM tracings were interpreted by two authors (SW and JC), and were unblinded. HREM was performed using a solid-state high-resolution esophageal manometry catheter with 36 sensors spaced 1 cm apart. Analysis was performed using the ManoView™ analysis software (Given/Sierra Scientific, Los Angeles California). The procedure for HREM was similar to that used for conventional EM except a pull-through was not necessary. The catheter was advanced transnasally until the UES and LES high-pressure zones were identified. After a 5-minute resting period baseline pressures of UES and LES were obtained and patient was given 10 to 15 5-ml boluses of water to swallow at 30 second intervals. LES and UES pressures, integrated LES relaxation pressure and esophageal peristalsis were analyzed. HREM tracings were interpreted by two authors (SW and JC), and were unblinded. Assignment of diagnosis was performed according to the Chicago classification [5].

Statistical analysis

The median ages for patients undergoing conventional EM and HREM were compared using the Welch two sample t-test. Gender, race, the presence of dysphagia as a primary indication, and the final diagnosis from manometry between the two groups were all compared using the chi-square test for independence. The frequency of normal manometry as well as the frequency of a diagnosis of achalasia was subsequently subjected to post hoc analysis using logistic regression.

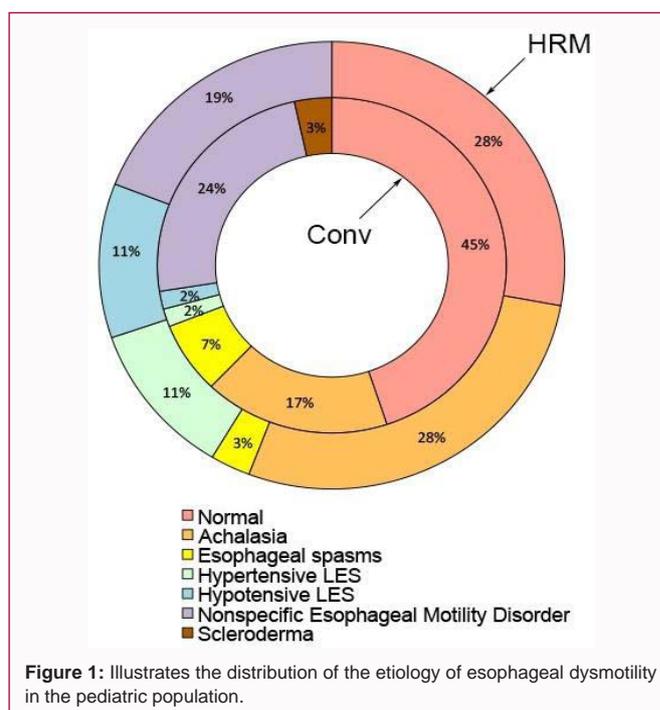


Figure 1: Illustrates the distribution of the etiology of esophageal dysmotility in the pediatric population.

The presence or absence of a normal exam and the presence or absence of achalasia as a final diagnosis were modeled as dependent variables, while age, gender, race, and the presence of dysphagia as a primary indication were modeled as independent variables. A p-value of 0.05 or less was used to determine statistical significance. All statistics were performed using the R software package (<http://www.r-project.org>). Since our response variable (the presence or absence of the performance of a procedure) was binary, we chose a binomial distribution function and log it link function.

Results

A total of 94 subjects (48 males and 46 females) were included in the study. Fifty-eight patients (30 males and 28 females) underwent conventional EM from 2001 to 2011 while 36 patients (18 males and 18 females) underwent HREM from 2011 to 2013. The median age of our subjects was 13 years. The most common indication for manometry was dysphagia (80%, n=75), followed by vomiting (9%), regurgitation (5%), and chest pain (4%). Thirty-six subjects (38%) had a normal esophageal manometry. The most frequently diagnosed conditions were achalasia, and ineffective esophageal motility disorder characterized by non-transmitted or low amplitude (<30 mmHg) contractions in $\geq 30\%$ of swallows (conventional)/minor peristaltic abnormalities characterized by $\geq 30\%$ but <70% failed swallows or peristaltic breaks, or abnormally low DCI (HREM), with either condition being found in 22% (n=20) of subjects. Among the children found to have achalasia, 40% were males and 60% were females. Other diagnoses made were hypotensive LES (LESP < 10 mmHg, with complete relaxation or normal IRP and normal distal peristalsis) in 8%, esophageal spasms/DES (HREM) in 4%, and hypertensive LES/EGJ outflow obstruction (HREM) in 4%. Two percent of our subjects had scleroderma, while none of the patients were found to have Nutcracker esophagus (conventional) or Jackhammer esophagus (HREM) (Figure 1). There were no significant differences between conventional EM and HREM in age (p=0.37), gender (p=0.71), race (p=0.93), or the presence of dysphagia as a presenting symptom (p=0.39). The distribution of final diagnosis

of manometry was, however, significantly different between the two groups ($p=0.04$). When subjected to logistic regression, there were no differences in the rate of normal/abnormal diagnoses or the rate of diagnosis of achalasia (Table 1). We were unable to assess referral patterns to determine the cause of these differences, but we are the only pediatric motility center in the state of Indiana, and most such referrals are seen at our institution. Therefore, we re-analyzed the final diagnosis using just patients with abnormal results, and there was no difference ($p=0.11$).

Discussion

The true prevalence of esophageal motility disorders in children is unknown, and in a search of the literature we found very few studies that have looked at the frequency of classic esophageal motility disorders such as achalasia, esophageal spasm, Nutcracker esophagus/Jackhammer esophagus and ineffective esophageal motility disorders in children undergoing esophageal motility testing. Glassman et al. [6] reviewed the spectrum of esophageal disorders in 83 children undergoing conventional esophageal manometry for chest pain in 1992. The most common diagnosis in their patients was diffuse esophageal spasm in 33%, followed by achalasia (19%), hypotensive LES (14%), nonspecific esophageal motility disorder (14%) and Nutcracker esophagus and hypertensive LES (10% each). Rosen et al. [7] reviewed the frequency of diffuse esophageal spasms in 278 children undergoing water-perfused conventional esophageal manometry in their institution from 1994 to 2004 and reported that 13% of their patients had diffuse esophageal spasms. Twenty percent of their patients were diagnosed with nonspecific esophageal motility disorder while 4% were diagnosed with achalasia. The patients in their study who were diagnosed with diffuse esophageal spasm presented with food refusal or chest pain. The majority of our patients undergoing esophageal manometry presented with dysphagia, and in this group the most common diagnoses were ineffective esophageal motility disorder and achalasia, being found in 24% and 17% of our patients undergoing conventional EM and 19% and 28% of our patients undergoing HREM respectively. Esophageal spasm was found in only 7% and 3% respectively. Hypotensive and hypertensive LES/EGJ outflow obstruction (HREM) were found in 2% and 11% respectively while hypercontractile disorders such as Nutcracker esophagus (conventional) and Jackhammer esophagus (HREM) were not found. This is similar to adult studies of patients undergoing esophageal manometry for dysphagia. Katz et al. [8] diagnosed achalasia in 19% and ineffective esophageal motility disorder in 27% of 251 patients undergoing conventional esophageal manometry for dysphagia. Diffuse esophageal spasm was seen in 7% and nutcracker esophagus in 5% [8]. A larger percentage of our patients were diagnosed with achalasia using HREM (28%) than with conventional EM (17%). This could be due to increased incidence of achalasia during the time period 2011 through 2013 when we switched to HREM, although we cannot be certain about this. Marlais et al. [9] and Sadowski et al. [10] reported that the rate of achalasia has been increasing in the last 2 decades.

Conclusion

In conclusion, the most common diagnoses made in children with a predominant symptom of dysphagia undergoing both high

resolution and conventional water-perfused esophageal manometry in our institution was ineffective esophageal motility disorder and achalasia. Esophageal spasm, Nutcracker esophagus, isolated hypertensive or hypotensive LES (conventional) and Jackhammer esophagus (HREM) were less common. A limitation of our study is its retrospective nature, which is not ideal. Furthermore, although both HREM and conventional EM are studying the same gross abnormalities of esophageal motility; that is abnormalities of peristalsis and sphincter relaxation, the advancement in HREM allows for more detailed and specific characterization of these abnormalities which is not achievable with conventional manometry, making it challenging to compare the two modalities. Moreover, our HREM diagnoses were made using adult criteria; it has subsequently been shown that some of these criteria are influenced by age and size and could affect the diagnosis if age adjustments are not made in children [11]. A prospective analysis collecting data in a registry would be a superior study design and with HREM now replacing conventional EM in most centers, a multicenter study will be ideal to give us a better perspective on the incidence of these motility disorders in children.

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