The assessment of the esophageal motility of children with esophageal disorders by the detailed observation of the pH-multichannel intraluminal impedance waveform and baseline impedance: screening test potential

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Running head: Motility assessment of pediatric esophageal disorders

Abstract

Background

The present study aimed to evaluate whether the detailed observation of pH/MII waveforms and the analysis of baseline impedance (BI) values could detect esophageal dysmotility in pediatric patients with esophageal disorders.

Patients and Methods

Eleven patients with congenital esophageal disorder in whom pH/MII was conducted from April 2011 to June 2015, were enrolled in this study. The diagnoses of the patients were as follows: postoperative esophageal atresia (EA), n=6; esophageal achalasia (EAch), n=4; and congenital esophageal stenosis (CES), n=1. The characteristics of the pH/MII waveform, pathological GERD, BI value, and the average BI value of the 2 distal channels (distal BI; DBI) were analyzed in each disorder.

Results

Two EA (33%) patients and 1 EAch (25%) patient were diagnosed with GERD. The mean DBI values of the EA, EAch and CES patients was 912 ± 550, 2153 ± 915 and 1392Ω, respectively. The EA patients showed consistently low DBI values. One CES patient and 2 infantile EAch patients showed postprandial prolonged low DBI values. Whereas, the pH/MII waveforms of the adolescent EAch patients were difficult to interpret due to their extremely low BI values.

Conclusions

The present study demonstrated that the detailed observation of the pH/MII waveforms in all channels and the analysis of BI was useful for evaluating esophageal motility in children with congenital esophageal disorders. In particular, infantile patients with EAch showed DBI findings that were distinct from those of adult EAch patients.

Considering the difficulty of performing esophageal manometry in young children, the detailed observation of the pH/MII waveform may help in the diagnosis of esophageal dysmotility in children.

Key words: baseline impedance, multichannel intraluminal impedance measurement, esophageal motility, waveform pattern, pediatric

Abbreviations

pH/MII	esophageal combined pH–multichannel intraluminal impedance measurement
EA	esophageal atresia
EAch	esophageal achalasia
CES	congenital esophageal stenosis
BI	baseline impedance
DBI	distal baseline impedance
BPT	bolus presence time
TBTT	total bolus transit time

Introduction

The main role of the motor function of the esophagus is to clear swallowed material into the stomach and to prevent the reflux of gastric content into the esophagus. Motility disturbances of the esophagus are always observed to a greater or lesser extent in children with congenital esophageal disorder.

Esophageal manometry, using either a liquid or a viscous material, is widely considered to be the gold standard for assessing esophageal motility; several previous studies have reported its usefulness [1]. This conventional technique provides detailed information about the esophageal pressure pattern as well as the sphincter function but

does not provide information about bolus transit. Thus far, esophageal motility studies using either radiology or scintigraphy have been performed as alternative examinations; however, these techniques are associated with limitations, including radiation exposure, which is especially problematic in children.

Recently, esophageal combined pH-multichannel intraluminal impedance measurement (pH/MII) has emerged as a novel technique for evaluating both GERD and esophageal motility [2, 3]. Impedance channels can detect bolus transit and the direction of bolus movement [4]. Validation studies have found an excellent correlation between the findings of pH/MII and videofluoroscopy. There is also a good correlation between the findings of pH/MII and manometry in healthy subjects and patients with GERD [5].

Several previous reports that have assessed esophageal motility by calculating various pH/MII parameters during bolus transit have revealed that pH/MII, which can detect direct bolus transport in the esophagus, is useful for evaluating the esophageal function [6]. Esophageal bolus clearance can be assessed by measuring the average bolus presence time (BPT) or the total bolus transit time (TBTT) and by classifying swallows as complete bolus transit or as incomplete bolus transit [7]. However, this method of evaluation has a weakness in that it cannot assess the esophageal disorders associated with passage obstruction or severe motility disorders, such as esophageal achalasia (EAch) and esophageal atresia (EA).

Previous studies have shown that adult patients with various degrees of esophageal motor abnormalities show low baseline impedance (BI) values in comparison to healthy controls [8-10]. It has been also reported that low BI levels, reflecting chronic fluid retention in the lower esophagus, are associated with EAch [11].

In contrast, there are no reports have focused on the application of BI in the assessment of esophageal motility disorders in children. Furthermore, no studies have evaluated esophageal motility in children with congenital esophageal disorders. We also hypothesized that the detailed observation of the pH/MII waveform can yield important information about what is happening in the esophagus. Thus, the aim of the present study was to evaluate whether the detailed observation of the pH/MII waveform and the analysis of BI values can detect the characteristics of esophageal dysmotility in pediatric patients with esophageal disorders.

Patients and methods

The study population of this retrospective study included 11 patients with congenital esophageal disorders (mean age, 5.0 ± 4.3 years; range, 10 months to 14 years) in whom pH/MII was conducted at Kurume University hospital from April 2011 to June 2015. The diagnoses of the patients included esophageal atresia (EA), n=6; esophageal achalasia (EAch), n=4; and congenital esophageal stenosis (CES), n=1. In preparation for this study, medications that were administered for GERD (*i.e.,* proton pomp inhibitors, prokinetics, and a herbal medicine [Rikkunshito]), were stopped at least 3 days before the patients entered the study. The study protocol was approved by the Kurume University Ethical Committee (No. 2575). Informed consent was obtained from the families of the patients before starting this study.

Two multiple intraluminal impedance catheters with two pH antimony electrodes and seven impedance electrodes (patient's height <150 cm, CZPN-BG-57; ≥150 cm: ZAN-BG-44, Sandhill Scientific, Inc., Highlands Ranch, Co., USA) were used. The catheter was inserted transnasally through the esophagus, and the pH sensor was fluoroscopically placed above the two vertebrae of the diaphragm. The impedance

data were automatically evaluated using the BioVIEW analysis software program and each tracing was manually reviewed by the same investigator (S.F.)

The pH index (pHI) was defined as the percentage of time when pH was <4 and included acid reflux events and pH-only events. An abnormal pHI was defined according to the criteria published by the German Pediatric Impedance Group, as a value of of ≥10% in patients of <1 year of age or ≥5% in patients of > 1 year of age [12]. The BI values of the 2 channels of the distal esophagus (Z5 and Z6) were assessed every 6 h over the first stable 60 s time period by using the automatic calculating function (electronic ruler) of the software program and the obtained waves were averaged, according to the method described by Borrelli et al. [13].

The characteristics of the waveform pattern of pH/MII, pathological GERD, the BI values (Z5 and Z6) and the average BI value of the 2 distal channels (distal baseline impedance, DBI) were evaluated in each patient [10].

Results

Patients with EAch were diagnosed by a combination of esophagography, upper-gastrointestinal endoscopy and esophageal manometry. In EAch patients, manometry revealed the increased resting pressure of the LES with the absence of relaxing on swallowing and simultaneous contractions of the esophagus.

A boy with CES was also diagnosed based on the findings of esophagography, upper-gastrointestinal endoscopy and esophageal manometry. He presented vomiting, dysphagia and refusal of solid food. Esophagography revealed a dilated esophagus with an air-fluid level and esophageal stenosis at the lower esophagus **(Figure 1A)**. Endoscopy showed alimentary bolus retention in the esophagus and esophageal stenosis

(Figure 1B). Manometry revealed no simultaneous contraction and relaxation of the LES on swallowing **(Figure 1C)**.

Two patients with EA (33%) and one with EAch (25%) were diagnosed with GERD based on the pH/MII findings **(Table 1)**.

The mean BI values of Z5 to Z6 in the EA, EAch and CES patients were 1066 ± 734, 755 ± 374 and 2391 ± 988,1915 ± 937 and 1659,1124Ω, respectively. The DBI values of patients with EA, EAch and CES were 912 ± 550Ω, 2153 ± 915Ω and 1392Ω, respectively **(Table 2)**.

The esophageal disorders demonstrated distinctive waveform patterns on pH/MII. The patients with EA showed consistently low BI values during all examination periods **(Figure 2)**. One patient with CES and 2 infant patients with EAch showed postprandial prolonged low DBI **(Figures 3A and 3B).** Whereas, the pH/MII waveforms of adolescent patients with EAch were difficult to interpret due to their extremely low BI values during all examination periods **(Figure 4A)**. However, after the balloon dilatation procedure, the waveform was significantly improved **(Figure 4B)**.

Discussion

Esophageal manometry is considered to be the gold standard for studying esophageal motility. However, it is often difficult for pediatric patients to perform wet swallows in standard manometry studies. Moreover, sedation is often required for such patients, which makes this standard examination invasive.

pH/MII has been reported to be useful as a novel alternative method of examining esophageal motility. It shows high sensitivity and specificity in the detection of bolus transit abnormalities in comparison to multichannel intraluminal impedance

and esophageal manometry [14]. The advantage of pH/MII is that it is able to collect the data for a long period and does not require the cooperation of the patient (*i.e.,* wet swallows or remaining in the supine position) once a thin catheter is inserted into the esophagus. These characteristics make it a suitable method for examining pediatric patients. A study by Tutuian et al., revealed that most patients with major esophageal dysmotility show abnormal MII findings [15], which indicated that pH/MII is not likely to miss important findings associated with esophageal motility disorders or GERD. On the other hand, pH/MII has a weakness in that parameters related to bolus transit (*i.e.,* bolus presence time and total bolus transit time) cannot detect stagnant fluid in the esophagus. Heard et al. reported that pH/MII was not accurate for analyzing patients suffering from EAch, GERD, scleroderma or ineffective esophageal motility due to their extremely low BI values <500Ω [10].

Conversely, regarding the relationship between esophageal motility and BI, the previous adult studies have indicated that the patients with various degrees of esophageal motor abnormalities show low BI values in comparison to healthy controls [8, 10].

Additionally, a study of adult patients with GERD suggested that the presence of a low BI is commonly associated with the mucosal inflammation related to esophageal acid exposure [13, 16].

As mentioned above, we hypothesized that such BI findings imply the presence of severe GERD or esophageal dysmotility. Thus, a detailed overall observation of the pH/MII waveform patterns to assess the changes in the BI values might be useful for indicating further examinations (*i.e.,* endoscopy or manometry) for pediatric patients.

It is a well-known fact that almost all EA patients suffer some degree of esophageal dysmotility due to congenital abnormal neural development in the

esophagus and complications after surgical repair. Several studies using conventional water-perfused stationary manometry or high resolution esophageal manometry demonstrated impaired esophageal peristalsis and an impaired LES function in EA patients [17-20] Furthermore, GERD is a common problem in children with this esophageal disorder. Tambucci et al. reported that patients with EA showed low BI values in both the proximal and distal channels, regardless of the presence of GERD, suggesting a close association between esophageal dysmotility and the BI value [21]. They also reported that the patients with EA showed the lowest distal BI values (895 [615-1189]Ω) among patients with GERD (2231 [1170-3032]Ω) and controls (3522 [2927-3994Ω]).

It remains controversial as to whether or not mucosal inflammation due to GERD or esophageal motility disorder has a greater effect on the BI value. Stanley et al. reported that the DBI values of EA patients tended to be low and that biopsies revealed esophagitis in 57% of EA patients [22]. In contrast, Pedersen et al. reported that the BI values of EA patients with erosive esophagitis (552Ω [411-967Ω]) were significantly lower than those of EA patients without erosive esophagitis (794Ω [578-1040Ω]) [23]. Thus, the DBI may be useful for identifying patients who should undergo endoscopy.

In the present study, more than half of the patients with EA were diagnosed with GERD based on pH/MII and showed low DBI values, which was consistent with the previous studies that reported that a low DBI reflects inflammation of the esophageal mucosa as well as esophageal motility disorder in EA patients. The esophageal motility is profoundly impaired in almost all children with EA; this may, in turn, parallel both the impaired bolus transit and low distal BI values detected by pH/MII [24]. We could not definitively determine whether poor esophageal motility or

esophageal inflammation was the true cause of the low DBI values in EA patients; however, the results indicate that the detection of a low DBI may be a useful indication for endoscopy.

Needless to say, manometry is the most sensitive and specific method for establishing the diagnosis of EAch. Several studies on the relationship between EAch and BI have reported that adult EAch patients showed low BI values. Nguyen et al. reported that the mean BI values of EAch patients (999Ω) were significantly lower in comparison to healthy volunteers (2749Ω) [25]. Moreover, a recent study by Conchillo et al. revealed that the BI values in the distal esophagus were low in EAch patients [11]. Heard et al. also reported that more than half of EAch patients showed low DBI values (<300Ω) [10].

In the present case, adolescent EAch patients showed extremely low BI values in all channels during all examination periods, which made pH/MII difficult; this was consistent with the findings of previous reports. On the other hand, EAch is a relatively rare condition in pediatric patients; its incidence is reported to be 0.1–0.3 cases per 100000 children per year in the United Kingdom. The age-dependent alteration of the nerve endings in the LES of EAch has been reported [26]. As EAch develops, the numbers of NOS-positive neurons may gradually decrease in patients with EAch; this may be why the duration of symptoms is prolonged before the treatment of EAch [27]. Morera et al. reported that the LES function varies with time and that it may appear normal in the early stages of EAch. They also showed that only 4 of 29 EAch patients had no LES relaxation in all swallows and that the LES function of children with EAch was heterogeneous, with some showing an intermittent degree of LES relaxation and occasional normal relaxations [28]. In our study, the infantile

patient with EAch did not show a low BI value; however the spontaneous decrease of the BI value in the distal channel was observed during postprandial period based on the detailed observation of the pH/MII waveform. Thus, the present results might reflect age-dependent alteration or an individual stage of Each; however, a study of a larger population would be necessary to confirm this result.

CES is a rare disorder that is characterized by intrinsic esophageal narrowing due to an esophageal wall abnormality. The incidence is 1 in 25000-50000 live births, with the highest incidence in Japan [29]. The diagnostic criteria and standard treatment have not been completely established. Although esophagography and esophagoscopy are frequently conducted as diagnostic examinations, the accurate diagnosis of CES is often difficult in actual clinical practice. To the best of our knowledge, no reports have analyzed the BI values of CES patients. In the present study, the BI value of the patient with CES spontaneously decreased in the postprandial period (similarly to the infantile patient with EAch), indicating that the detailed observation of the pH/MII waveform might also help in the detection of CES.

In summary, the present study investigated 3 types of the congenital esophageal disorders in pediatric patients and indicated that the detailed observation of the pH/MII waveform patterns would enable the detection of esophageal passage disturbance in infantile patients with EAch and CES. Furthermore, this method of evaluation could confirm the dramatic improvement of the waveform pattern after a pneumatic dilatation procedure in adolescent patients with EAch.

Thus, the combination of the detailed observation of the waveform and the evaluation of the BI by pH/MII enabled the detection of esophageal passage obstruction in pediatric patients and could be useful for indicating further examinations such as manometry or

endoscopy. We created a flowchart to illustrate the method of diagnosing children with congenital esophageal disorders such as EA, EAch , and CES (**Figure 5)**. Moreover, this approach can also be used to evaluate the effects of therapy in patients with EAch.

The present study is associated with some limitations, in particular, the small study population. Furthermore, the present approach of evaluating pH/MII can indicate the possibility of the esophageal motility disorder but cannot evaluate the degree or type of disorder. A study of a larger population is necessary to further elucidate the pathophysiological mechanisms of esophageal disorders.

In conclusion, the present study demonstrated that the detailed observation of the pH/MII waveform pattern in all channels and the analysis of the BI value is useful for evaluating the esophageal motility of children with congenital esophageal disorders. In particular, the DBI findings in an infant patient with EAch were distinct from those of adult EAch patients. Considering the difficulty of performing esophageal manometry in young children, the detailed observation of the pH/MII waveform pattern may help in the diagnosis of pediatric patients with esophageal dysmotility and be useful for indicating further examinations, such as manometry or endoscopy.

Author contributions

Daisuke Masui and Suguru Fukahori designed the research study, analyzed the data and wrote the paper; Minoru Yagi & Yoshiaki Tanaka designed the research study and wrote the paper; Shinji Ishii & Naoki Hashizume analyzed the data; Nobuyuki Saikusa, Motomu Yoshida, Naruki Higashidate ,Saki Sakamoto, Shiori Tsuruhisa & Hirotomo Nakahara performed the research.

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Ethical Statement

The protocol of this study has been approved by the Kurume University Ethical Committee (No. 2575). Informed consent was obtained from the families before starting this study.

Conflict of interest

Drs. Masui, Fukahori, Ishii, Hashizume, Saikusa, Yoshida, Higashidate, Sakamoto, Tsuruhisa, Nakahara, Tanaka and Yagi have no conflicts of interest or financial ties to disclose.

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Table 1. The pHI and BI values in each patient

Type of disease	Age (y)	gender	pHI (%)	BI:Z5 (Ω)	BI:Z6（Ω）
EA	8	F	0.3	2166	1250
EA	7	F	7.1	584	657
EA	5	M	9.2	272	326
EA	2	M	4.7	603	408
EA	0.83	F	5.5	1703	1132
EA	2	F	0.4	1069	755
EAch	1	F	0.1	1564	1719
EAch	3	M	1.4	2125	1092
EAch	14	F	0	3485	2935
EAch	10	M	8.3	#	#
CES	2	M	1.5	1659	1124

BI, baseline impedance; pHI, pH index; EA, esophageal atresia; CES, congenital esophageal stenosis; EAch, congenital esophageal achalasia
The analysis was difficult due to an extremely low BI value.

Table 2. The mean Z5 and Z6 and DBI values in each disease

Type of disease	No. of patients	Mean BI:Z5 (Ω)	Mean BI:Z6 (Ω)	Mean DBI (Ω)
EA	6	1066 ± 734	755 ± 374	912 ± 550
EAch	3	2392 ± 988	1915 ± 937	2153 ± 915
CES	1	1659	1124	1392

BI, baseline impedance; DBI, distal baseline impedance; EA, esophageal atresia; CES: congenital esophageal stenosis; EAch, congenital esophageal achalasia.

Figure legends

Figure 1.

Esophagography revealed a dilated esophagus with an air-fluid level and esophageal stenosis at the lower esophagus **(A)**. Endoscopy showed alimentary bolus retention in the esophagus and esophageal stenosis **(B)**. The manometry findings showed no simultaneous contraction and relaxation of the LES on swallowing **(C)**.

Figure 2.

The waveforms of EA patients showed consistently low BI values during all examination periods.

Figure 3.

One patient with CES and 2 infants with EAch showed postprandial prolonged low DBI **(A and B).**

Figure 4.

The MII-pH waveforms of adolescent patients with EAch were difficult to interpret due to their extremely low BI values during all examination periods **(A)**. However, the waveform showed significant improvement after the balloon dilatation procedure **(B)**.

Figure 5.

Flowchart of the method of diagnosing children with congenital esophageal disorders such as EA, EAch, and CES.