Introduction

Achalasia is an uncommon swallowing disorder characterized by absence of esophageal body peristalsis associated with an impaired relaxation of lower esophageal sphincter (LES) and elevated LES pressure, leading to passage disturbance.\textsuperscript{1,2} The incidence of achalasia is expected to be 1 in 100000 persons per year with a prevalence of 10 in 10000. This disorder can occur at any age, with a two peaks incidence at 20~40 and 70~80 years, without gender prevalence.\textsuperscript{3} Achalasia results from the degeneration of neurons in the esophageal wall. The cause of this inflammatory degeneration of neurons in achalasia is not known, however, it is thought to be associated with viral infection, autoimmune disease or other degenerative disorders.\textsuperscript{4}

With advances in diagnostic technique, such as high resolution manometry (HRM), an international consensus process has evolved over recent years to define esophageal motility disorders using HRM and newly devised classification (Chicago Classification) was firstly published in 2009\textsuperscript{5} and was subsequently updated in 2012.\textsuperscript{6}

Evaluation for diagnosis of achalasia

1. Symptoms

The diagnosis of achalasia should be suspected in any patient complaining of dysphagia for solids and liquids with regurgitation of undigested food and saliva. Most common symptoms are dysphagia and regurgitation in 70~90\% of patients. Other frequent symptoms are chest pain, heartburn, weight loss and respiratory complications such as nocturnal cough and aspiration.

Eckardt score is very useful tools most widely used to assess the severity of symptoms and success of treatment of achalasia. This score which range from 0 to 12 is based on the clinical symptoms including dysphagia, regurgitation, chest pain and degree of weight loss (Table 1). Usually drop in Eckart score to $\leq 3$ has been considered as treatment success. Eckardt score should be evaluated before and after treatment of patients with achalasia.

2. Barium esophagography

In the diagnosis of achalasia, both barium esophagography and endoscopy are less sensitive than manometry. In early stage of achalasia endoscopic and barium study may be normal. Barium esophagography is used to visualize the

Table 1. Eckardt Score

<table>
<thead>
<tr>
<th>Score</th>
<th>Weight loss</th>
<th>Dysphagia</th>
<th>Regurgitation</th>
<th>Chest pain</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>1</td>
<td>$&lt; 5 \text{ kg}$</td>
<td>Occasional</td>
<td>Occasional</td>
<td>Occasional</td>
</tr>
<tr>
<td>2</td>
<td>5--10 kg</td>
<td>Daily</td>
<td>Daily</td>
<td>Daily</td>
</tr>
<tr>
<td>3</td>
<td>$&gt; 10 \text{ kg}$</td>
<td>Each meal</td>
<td>Each meal</td>
<td>Each meal</td>
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</table>
anatomic structure and motility of esophagus. It typically reveals a “bird beak” appearance at the esophagogastric junction (EJG) with a dilated proximal esophageal body and an air-fluid level in the absence of an intragastric air bubble. Esophagography has some advantage over endoscopic examination in the diagnosis of achalasia. This advantage is that an assessment of esophageal motility and function is available. To assess motility and emptying of the esophagus, a timed barium swallow can be performed, in which the height of the barium column 5 minutes after ingestion of diluted barium is a measure for emptying. This study is important in assessing response to therapy.

3. Endoscopy

Endoscopy is also helpful in the diagnosis of achalasia although it may be normal at early stage. One of the earliest finding in achalasia is the resistance as the endoscopy pass through the lower esophageal sphincter (LES) into the stomach due to impaired LES relaxation. In patient with long standing achalasia, endoscopy reveals a dilated esophagus with retention of food material. Most important role of endoscopy in patients with symptoms suspicious achalasia is the exclusion of other disease which are mimic achalasia such as esophageal cancer or cardio cancer. Endoscopic examination should be performed for differential diagnosis from other disease.

4. Manometry

Manometry is the gold standard for the diagnosis of achalasia and will provide diagnostic certainty in approximately 90% of the cases. Conventional manometry examination typically reveals 3 abnormalities in patient with achalasia: high pressure in LES at rest, impaired LES relaxation after swallowing and the absence of peristaltic contraction in lower esophagus. The later two features are the most important and are required to make the diagnosis of achalasia.

Recently high-resolution manometry (HRM) is increasingly performed providing more detailed information on esophageal motility. Using catheters incorporating 36 or more pressure sensors spaced only 1cm apart, HRM allows in detail pressure recording of the whole esophagus, and is currently considered the gold standard to diagnose achalasia. In HRM system, new parameter is used to quantify LES relaxation, the integrated relaxation pressure (IRP). During swallowing, LES relaxation is evaluated using the IRP, which has been defined as the mean of the 4 seconds of maximal deglutitive relaxation in the 10-second window beginning at deglutitive UES relaxation. With application of HRM, Chicago Classification has subdivided achalasia into 3 distinct phenotype (Fig. 1), the clinical relevance of which has been verified in several studies.

Type I achalasia was characterized by 100% failed peristaltic contraction and no esophageal pressurization, type II achalasia was defined as 100% failed peristaltic contraction and preseverd peristaltic contraction, type III, there is at least 20% premature contraction, D; EGJ outflow obstruction is characterized by impaired LES relaxation and preseverd peristaltic contraction.

Fig. 1. Subtype of achalasia. A; Type I is characterized by impaired LES relaxation and 100% failed peristalsis, no esophageal pressurization, B; Type II is characterized impaired LES relaxation and panesophageal pressurization and absence of peristaltic contraction, C; in type III, there is at least 20% premature contraction, D; EGJ outflow obstruction is characterized by impaired LES relaxation and preseverd peristaltic contraction.
panesophageal pressurization for at least 20% of swallows, and type III achalasia was defined as the presence of pressed fragments of distal peristalsis or premature contractions for at least 20% of the swallows in the 2012 Chicago Classification. The Chicago Classification also included another potential achalasia phenotype, which is classified as esophagogastric junction outflow obstruction (EGJOO), defined by impaired LES relaxation with some degree of preserved peristalsis.

5. **Endoscopic Ultrasound**

Endoscopy, barium esophagrapy, HRM are major esophageal function testing methods. These diagnostic tools provide key information on function of the esophagus that allows one to treat patients based on the physiologic understanding of patient’s symptoms. Ultrasound imaging of esophagus is another potential esophageal function testing method which is not widely used yet. Ultrasound imaging can also provide other important information such as muscular hypertrophy and luminal disention during peristalsis. A recent study reported that hyperophy of both circular and longitudinal muscle layers were more common in patients with primary or nonspecific motor disorder compared to control patients. In a study of 98 consecutive patients with dysphagia, nearly all patients with primary motility disorders, based on manometry, were found to have an muscular hypertrophy, and 25% of patients with normal manometry also had thicker esophageal muscle than normal subject. This study suggest that normal manometry does not guarantee normal esophageal function, and ultrasound imaging could be a more sensitive modality than manometry to detect esophageal motor dysfunction.

**Conclusions**

New manometric technology and recently described analysis algorithms summarized in the Chicago Classification have greatly improved the clinical recognition of achalasia. However, there are many patients complaining dysphagia in spite of normal manometric or endoscopic findings. They might be a patients with achalasia, other nonspecific motility disorder or functional dysphagia. Further studies are needed to increase the sensitivity of esophageal motility disorders.

**References**