Several papers have reported [1] about “functional disorders of the esophagus” in European countries and the USA; however, few reports on these disorders have been found in Japan except concerning achalasia [2]. Having an opportunity to study with Dr. DeMeester, a well-known surgeon in this field, I had the personal experience of measuring the esophageal motility and pH recording in many patients with these disorders from June 1987 to August 1988 in Omaha, Nebraska, USA.

The question I kept asking myself during this period in the USA was why there were so few reports about benign esophageal disorders in Japan and why gastroenterologists in Japan had so little interest in these clinical entities. In other words, is there, in fact, low incidence of these disorders among Japanese or is there simply no concern with these diseases among Japanese gastroenterologists? The only way to clarify this question is to measure esophageal motility and pH monitoring in patients with dysphagia, non-cardiac chest pain or heartburn in Japan, too. It is also important to perform the examination in the same way as occidentals do and to compare our results with theirs. As there is confusion about the criteria for the definition and name of these disorders in Japan, it may be necessary to define these relatively new clinical concepts as follows.

Functional disorders of the esophagus are defined as the disorders of the esophagus that are not derived from organic diseases such as malignant or benign tumor, diverticulum or stricture. Since the function of the esophagus is to transport material from the mouth to the stomach, functional disorders are related to disturbance of motility. They are classified as primary or secondary. Primary functional disorders may be subdivided into the three anatomical regions: upper, middle, and lower esophagus. Neuromuscular diseases cause motility disorders in the pharyngeal region, diffuse esophageal spasm (DES), nutcracker esophagus (high amplitude peristalsis), which is a disorder of the esophageal body, and achalasia or hypercontracting sphincter, a dysfunction of the lower esophageal sphincter (LES). Secondary functional disorders include motility disorders derived from gastroesophageal reflux (GER) or collagen vascular diseases.

To make a precise diagnosis of functional esophageal disorders, specific diagnostic tests are required in addition to a detailed history and physical examination. In practice, these include video roentgenographic barium studies and endoscopy, esophageal manometry, 24-hour pH monitoring, and sometimes ambulatory esophageal motility.

Manometry may be performed using a perfused multi-lumen catheter connected to a pneumohydraulic capillary infusion pump. The amplitude, duration, propagation, and efficiency of contractions in the esophageal body as well as the incidence of abnormal contractions may be recorded manually or by a computer. The location, length, pressure, and percentage relaxation of the LES are recorded. The purpose of manometry is twofold: first, to record esophageal motor
activity and second, to place the pH probe accurately for 24-hour monitoring. It is customary in our laboratory to place the probe 5 cm above the LES. The distance above the sphincter is critical for comparing patterns of reflux with normal controls. Reflux episodes are defined by the pH falling below 4.0 and the following parameters: the number of reflux episodes > 5 min., the longest episode in minutes, the total number of episodes and percent time while supine and upright are used to calculate a pH score (the DeMeester Score). In cases with unexplained chest pain or dysphagia, 24-hour ambulatory esophageal manometry may provide further detailed diagnosis [3].

The most common causes of pharyngoesophageal dysfunction are neuromuscular diseases such as Parkinson's disease or multiple sclerosis. These patients show, in manometry, failure of the pharyngeal pump, cricopharyngeal incoordination and/or incomplete relaxation of the cricopharyngeal muscle (upper esophageal sphincter, UES).

Diffuse esophageal spasm (DES) was first described by Osgood (1989) [4]. It is characterized by symptoms of intermittent chest pain with or without dysphagia. Manometry shows that contractions in the body tend to be simultaneous in the distal esophagus and these contractions are of high amplitude and often prolonged and repetitive. The LES, on the other hand, usually relaxes normally. When the results of conventional manometry are normal but symptoms are still suspected, cholinergic provocative tests are recommended.

There are other motor disorders which are thought to be related to diffuse spasm. Vigorous achalasia shows simultaneous but high amplitude contractions in the body with failure of relaxation of the LES in contrast to findings in patients of classic achalasia, who have simultaneous low-amplitude contractions but incomplete relaxation of the LES. In patients with nutcracker esophagus, high-amplitude contractions are for the most part peristaltic rather than simultaneous.

Achalasia may be secondary to local ganglionic denervation and this is demonstrated manometrically as hyperreaction to cholinergics or pentagastrin provocative tests. The distinction between achalasia and diffuse esophageal spasm is not always clear [5].

In reflux disease a secondary motility disorder results which may lead to a reflux-induced aspiration pneumonitis. Manometric characteristics of reflux patients are not specific. They usually show lower amplitude than normals, existence of pathologic waves and lower percentage of synchronous contractions. In such cases, 24-hour pH monitoring is of great value.

In patients with collagen vascular diseases, motility disorders are common. In progressive systemic sclerosis, manometry shows a decreased LES pressure and amplitude in the distal esophagus because it is a disease of smooth muscle, which comprises the distal esophagus. On the other hand, the decrease in esophageal pressure in patients with polymyositis is seen in the proximal esophagus.

Esophageal manometry and 24-hour pH monitoring are essential to obtain a precise diagnosis of esophageal functional disorders. There is still much controversy about the categorization of each motility disorder, and each entity may not by discrete but represent points in a continuous spectrum. Further research must be performed in order to classify and treat these functional disorders of the esophagus.
NOTE  FUNCTIONAL DISORDERS OF THE ESOPHAGUS:

Organic Disorders
- malignant (cancer)
- benign (benign tumors, diverticula, strictures)

Functional Disorders
- primary
  - UES - neurological deficit
  - BODY - DES, Nutcracker
  - LES - achalasia, hypercontracting sphincter
- secondary
  - GER - incompetent cardia
  - scleroderma - competent cardia
  - myositis

Figure. Disorders of the esophagus

UES : upper esophageal sphincter
BODY : esophageal body
LES : lower esophageal sphincter
DES : diffuse esophageal spasm
GER : gastroesophageal reflux

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