ACHALASIA OF THE CARDIA IN CHILDREN

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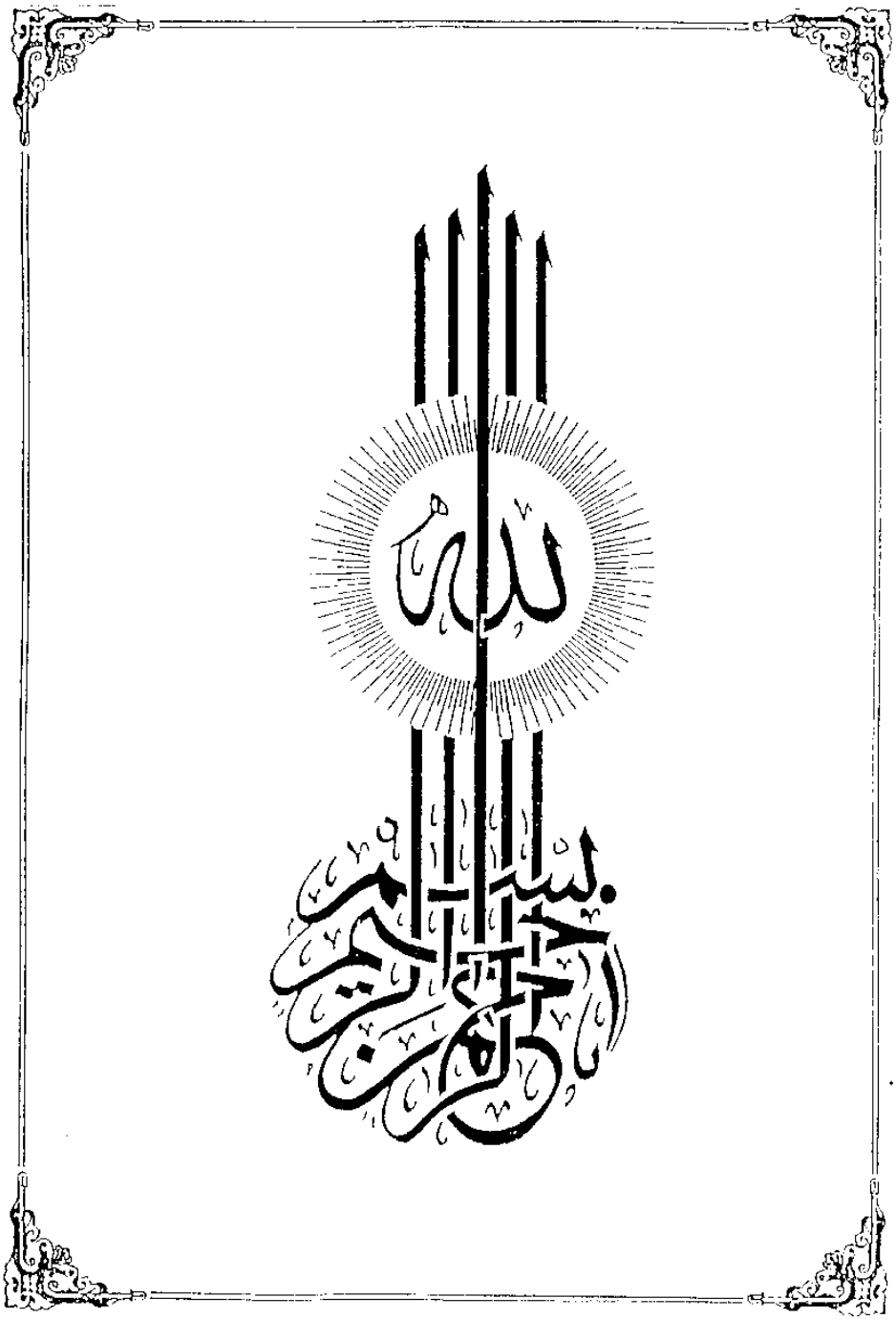

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Introduction

Achalasia is one of the oldest recorded diseases of the esophagus. It is relatively rare problem in children, with more than 250 cases reported in the worlds literature since the turn of the century.

The disease was first appreciated by Willes in 1674, who employed esophageal bongienage as successful therapy.

The etiology of achalasia of the esophagus remains poorly understood. However, occurrence of achalasia in the first year of life implies that achalasia in childhood may represent a congenital problem (Moazam F., 1976).

The ganglion cell abnormalities noted in Aurbach's plexus in the esophagus suggest that the pathogenesis of achalasia may be similar to aganglionosis of colon. This disorder is relatively rare, occuring in approximately one in 100,000 individuals in the general population.

Achalasia in children accounts for only 3% - 4% of total number of cases (Ellis FH, 1961). Modern surgical therapy for the disorder was described by Hellar in 1913, and with certain modifications, esophagomyotomy remains the surgical treatment of choice.

The use of pneumatic dilatation for achalasia in children was first used and reported by Moersch in 1929.

Like adults, many children require more than one dilatation to respond.

Some authors have suggested that children who respond to pneumatic dilatation are older than 9 years, (Aziz Khan RG, 1980). Although many reports of successful dilatation in children younger than 5 years have appeared (Berquist WE, 1983).

This essay will study the mysterial aspects of the disorder and methods of treatment, surgically and non surgically (Medical & Balloning dilatation). It will discuss also the experience of Aim Shams University in management and caring of children perioperatively.

Embryology Of The Esophagus

The elements of esophageal wall are derived from : -

(1) An internal tube of endoderm which is the primary tissue that eventually becomes the epithelial lining.

(2) An investing layer of splanchnic mesoderm which differentiates into lamina propria, submucosa and muscularis mucosa.

(3) The neurogenic elements are derived from the neural crest.

The esophagus develops from primitive foregut as a part of fusiform dilatation- that develops into stomach. A tubular esophagus becomes recognizable between the primitive pharynx and stomach around the third week gestation (Botha, 1963).

Initially the esophagus and, the trachea are a single tube. In its proximal portion, the lateral and anterior proliferation of the Layrgo-tracheal fissure forms the early trachea and the larynx. The

seperation of the trachea and esophagus is completed by the 36th day (Smith, and Taylor, 1972).

During the 4th week, the embryonic esophagus elongates rapidly by ascent of the Larynx, and also by the descent of the stomach in the thorax to enter the abdomen before the lateral diaphragmatic components fuse with the septum transversum.

Concentric deposition of mesodermal elements are found around the esophagus, these elements differentiate into muscles and connective tissue.

The inner circular layer of muscle differentiate at the 10 - 40 mm stage (6th week), and the outer longitudenal layer at the 20 - 72 mm stage (8th week).

The muscularis mucosa differentiates even later and becomes apparent at the 30 - 90 mm stage (10th week) (Botha, 1963).

Neuroblasts from the neural crest migrate in the mesoderm along, the vagi before 10 mm stage, and from complete circle outside the circular muscle layer (the myentric plexus) before the longitudenal muscle layer develops (Smith, and Taylor, 1972). During the 7th week, the epithelium lining the esophagus proliferates and vacuoles

appear. Thus, irregular channels appear in the esophagus but normally the lumen never become totally occluded.

By the 10th week, the vacuoles have disappeared, and a single lumen is restored. The epithelium becomes two-layered between the 23-24 mm stage. It subsquently changes into a multilayered sheath. It begins to acquire cilia after the 70 mm stage (John, 1952). Superficial glands develop during the 5th month, and deep glands develop mostly after birth (Arey, 1952).

The esophagus never acquires typical mesentry or serosal tissue (Gibbon, and Gamishon, 1969). Blood vessels from aorta and its branches penetrate the wall of lthe esophagus during the 7th week.

Anatomy of the Gastroesophageal Junction

The gastro-esophageal junction can be defined in 3 different ways :

1- The junction of esophageal squamous epithelium with columnars epithelium of gastric cardiac type.

2- The 2nd definition is " the point at which the muscular wall of the digestive tract changes from an esophageal to a gastric pattern.

The latest work of Liebermann- Meffert (1982) has demonstrated a special internal architecture of the lower esophagus and esophagogastric junction, consisting of two layers of smooth muscle - an internal with circular fibres (stratum circulae) and an external layer with longtudinal fibres (stratum Longitudinal).

The internal muscle bundles form a circle around the esophagus when contracting and narrow the esophageal lumen (Fig.). There fibres appear 1-2 cm proximal to the cardiac angle, extending down over the fundus of the stomach, particularly the greater curvature.

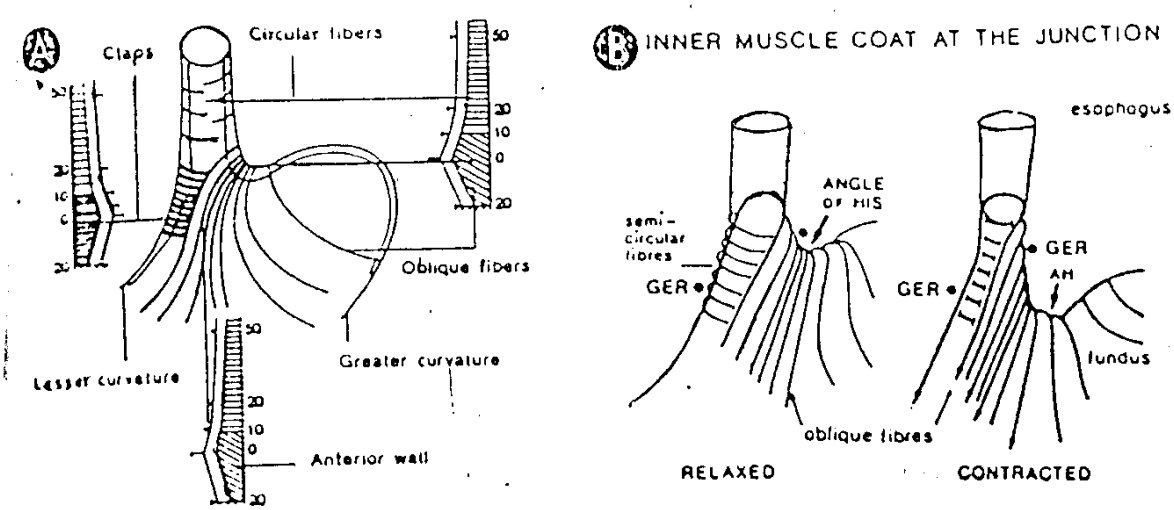

Fig. 1 : Masculature of gastroesophageal junction. (A) Muscle thickness marked to side of muscle structure. (B) contraction of the oblique fibres together with the mascular bands at the lesser curvature produces a ring - shaped mechanical closure. (From Liberman-Mefferet).

The gastric sling fibres are called the collar of Helvetius and, on contraction tend to sharpen the esophageal gastric angle. Contraction of the internal muscle layer, with consequent narrowing of the esophageal lumen and sharpening of the angle of his. strengthens the hypothesis that these muscle fibres play a role in the sphincteric mechanism. The thickest portion of this layer is directly proximal to its entry into the stomach corresponding to the high

The muscular thickness of this segment correlates with strength of the lower esophageal sphincter (Boix - Ochoa, 1986). There is an oblique or sling type fibres course from the lesser curvature toward the left and upward as a U-shaped sling around the greater curvature aspect of stomach and gastro-esophageal junction .

In dissection of human or subhuman primate as cat esophagus, no discrete anatomically identifiable circular sphincter muscle in the distal esophagus is never demonstrated.

Leibermann - Mefferet (1979) demonstrated the clustering of oblique gastric sling fibres around the greater curvature of the esophageal orifice lead to some thickening in the left posterior quadrant of the gastro-esophageal junction.

The thickening is not uniform around the distal esophagus nor the muscle fibres complete in a sphincter like pattern.

3- *The most useful definition of gastro-esophageal junction is "the gastric point part at which the tubular distal esophagus abruptly joins the dilated gastric digestive pouch".*

The problems with this definition : -

(a) The junction moves proximally during stomach in over distention.

This strectching of the stomach efface the opening of the tubular esophgus below the diaphragm gives an inverted funnel shaped structure. This sets the stage for normal post prandial physiological reflux (Bockus, 1985).

(b) The junction can be less certain during gastric outlet obstruction or when stomach is full.

The stomach orifice is normally Slit-like and tilted posteriorly from right to left.

By esophagoscopy, it is seen as a snug fold highly applied around the esophagoscope. When viewed internally through gastrostomy the normal orifice into stomach (the cardia) is never seen because it is covered by folds of gastric mucosa, when passing the finger 2-3 cm into the distal esophagus - a delicate ring can be palpated which represents the mucosal junction. When this ring is unduly tight, it is seen radiologically and called Schatzki ring. This ring does not indicate the presence of hiatal hernia but rather signifies the mucosal junction normally placed within the distal tubular esophagus.

The muscles of esophagus as it has inner circular and outer Longitudenal layer. The mucosa is easily stripped away from the muscle layer. This feature is evident when the paediatric surgeon performs circumferential myotomies to lengthen a shortened esophagus after tracheo - esophageal fistula repair, and by the surgeon who performs a modified Heller's cardiomyotomy for achalasia or diffuse esophageal spasm (Botha, 1963).

The upper 1/3 of the muscle layers consists of striated muscle and the lower 2/3 consist of smooth muscle. There is a short transition zone of both striated and smooth muscle between these segments.

Blood Supply of Esophagus

1- Arterial supply to the esophagus :

From (Damel, 1924) till Liebermann - Mefferet, 1987) show that there are three principal sources of the arterial supply to esophagus

(A) In neck :

The upper superior and inferior thryroid arteries send small arteries to the cervical esophagus. Immediate branching reduces the diameter of these vessels that are already minute when seen in peri-esophageal tissues before they enter the esophagal wall.

(B) At the level of the aortic arch :

A group of 3 to 5 tracheo - bronchial arteries arising from the concavity of the arch and a single tracheobronchial artery arising more caudaly from anterior surface of the aorta give rise to several esophageal tributaries, occasionally one or two esophageal arteries proper arise from the anterior thoracic aorta. Again, all esophageal vessels are reduced to small diameter by branching when they approach the esophageal wall.

(C) At the cardia :