DISORDERS OF ESOPHAGEAL MOTILITY

VOLUME

XVI

IN THE SERIES

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MAJOR PROBLEMS IN INTERNAL MEDICINE

Lloyd H. Smith, Jr., M.D., *Editor*

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EDITOR'S FOREWORD

The gullet is not an organ of glamour or versatility. Even the name "esophagus" reflects by etymology its function of carrying food —from the pharynx to the stomach. In brief it is simply a conduit which transfers but does not modify the diverse solids and liquids which traverse its span of 20 to 24 centimeters. The esophagus has been much maligned, however, in being considered only a membrane-lined tube for gravity flow. It must propel its contents, widely differing in composition and consistency, through the low pressure area of the thorax into the abdomen, where pressures may vary considerably. Furthermore, it must impede inappropriate reversal of this flow despite the variables of posture and pressure. Occasionally, of course, reversal is appropriate, when emesis is a protective activity to empty the stomach. In order to carry out these functions, the esophagus has a specialized musculature. The process of swallowing requires the complex coordination of striated and smooth muscle through voluntary and reflex actions to propel food in the proper direction and to protect the respiratory system. The pharynx is a common conduit for food and air, and hazards exist in that anatomical fact.

Where there is physiology, there is pathophysiology. This holds as true for a comparatively simple organ, such as the esophagus, as it does for the intricacies of the central nervous system. Disturbances of motility in the esophagus are common and may result in distressing dysfunction and symptomatology. These abnormalities may be functional disorders limited to the esophagus, or they may represent esophageal involvement by a more generalized disease, such as systemic sclerosis. From the work of many investigators, sophisticated methods have been developed to measure normal and abnormal motility of the esophagus and to sort out in a rational way the types of disorders which occur. This is of great importance, since therapy and prognosis can be shown to depend on this selective definition of abnormal function.

In this monograph Drs. Hurwitz, Duranceau, and Haddad have called upon their own experience, as well as summarized the work of others, to present the current state of knowledge concerning the disorders of esophageal motility. The result is an authoritative review, but one which is crisply focused on the problems faced in clinical medicine. As such it is a ready source of information for the general internist, as well as for the gastroenterologist in specialty practice.

LLOYD H. SMITH, JR.

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CONTENTS

CHAPTER 1

INTRODUCTION

Since the original publication of *An Atlas on Esophageal Motility* by Code and coworkers in 1958,[1] a large amount of material has been published about esophageal physiology and pathophysiology.

Of particular importance is the development of several new instruments and techniques that have improved the quality of the esophageal motility tracing.[2] In parallel with these developments, a more sophisticated understanding has evolved regarding the limitations of both the manometric technique and its interpretation.[3] These advances have led to new concepts in the pathogenesis of esophageal disease. For example, the lower esophageal sphincter dysfunction characteristic of achalasia was fully appreciated only after the development of water-perfused esophageal motility catheters.

In clinical practice an esophageal motility study should be considered in any patient whose esophageal complaints are not readily explained by a structural abnormality. The study may be diagnostic in achalasia, symptomatic idiopathic diffuse esophageal spasm, and esophageal scleroderma. It may be useful in the differential diagnosis of chest pain. The preoperative assessment of esophageal motility provides the surgeon with physiologic data that may influence the approach taken and quantify the results. Finally, manometric evaluation of the esophagus enhances one's understanding of esophageal function in certain diseases such as esophageal diverticula, gastroesophageal reflux, and oropharyngeal dysphagia.

This book has two purposes: first, to present normal and diseased esophageal function through the use of manometric tracings and second, to provide a rational approach to the patient with esophageal motor disease. The interpretation of motility tracings is emphasized, and how to perform an esophageal motility study is described in precise detail.

Motor diseases of the upper esophageal sphincter, the esophageal body, and the lower esophageal sphincter are covered sequentially in separate chapters. Both primary and systemic diseases known to influence esophageal function are discussed. An effort has been made to consider all factors that influence esophageal motility (e.g., the influence of gastric cancer and medications on the lower esophageal sphincter). A final chapter on the therapeutic and adverse effects of surgery on esophageal function discusses recent developments in this rapidly changing field.

The intent of the authors has been to provide a sufficient understanding of esophageal motility. Such understanding should lead to a more systematic approach to the patient with a swallowing disorder.

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CHAPTER 2

EMBRYOLOGY, ANATOMY, HISTOLOGY, AND CONTROL MECHANISMS

EMBRYOLOGY[1-9]

The esophagus appears in tubular form with the formation of the foregut at approximately the twentieth day after fertilization. The early esophagus then occupies the major part of the foregut between the stomodeum and the gastric dilatation.

Initially, the esophagus and the trachea are a single tube (Fig. 2–1A). In its proximal portion, the lateral and anterior proliferation of the laryngo-tracheal fissure forms the early trachea and larynx. More distally, lateral ridges build a septum between the anterior and posterior walls of the primitive gut (Fig. 2–1B). During the septation process, cells at the union of the septa undergo necrosis and form coalescent vacuoles. A collapse of the basement membrane then occurs, allowing a passive filling by mesenchymal cells (Fig. 2–1C). The separation of the trachea and esophagus is complete by the thirty-sixth day (Fig. 2–1D). While this separation proceeds, a rapid elongation of the esophagus takes place, mainly through the ascent of the larynx.[1, 2]

At the proximal end of the foregut, the endoderm and ectoderm fragment progressively to open the digestive tube. The surrounding mesoderm differentiates to form the various layers of the pharynx and esophagus. Striated muscle progressively spreads as an envelope around the primitive pharynx. At the 12.5 mm stage, the inferior constrictor muscle can be recognized.[8] At six weeks of gestation, the circular layer of esophageal muscle can be identified, and nerve cells appear just peripheral to this layer. By the ninth week, the longitudin-

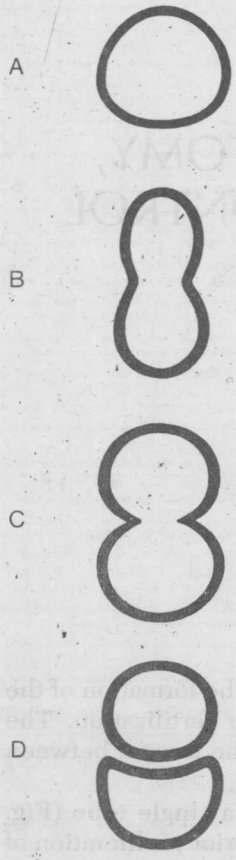

Figure 2-1. The embryologic development of the esophagus and trachea. Progressive division of the two structures is complete at the thirty-sixth day of gestation.

al muscle layer has covered the circular one. The muscular layer is a definite structure at 12 weeks.[3] Striation appears in the muscle of the proximal esophagus at a later stage than the somite period. This suggests that these striations originate from a primary differentiation of the esophageal muscle itself and not from pharyngeal extension.[4] The muscularis mucosae differentiates from a longitudinal myoblast layer at the same time that the longitudinal muscle layer appears.[5]

Blood vessels from the aorta and its branches penetrate the wall of the esophagus during the seventh week.

Innervation of the pharynx and pharyngoesophageal junction is based on the original branchial arches involved in their formation. The fourth arch is responsible for the formation of the pharyngeal constrictors and receives its innervation from the superior laryngeal nerve. The fifth and sixth arches give rise to pharyngeal and laryngeal musculature; they are innervated by the recurrent laryngeal nerve.[2, 9]

Neuroblasts from the neural crest migrate in the mesoderm adjacent to the endoderm of the foregut and form the myenteric plexus between the muscle layers of the esophagus. These cells are second-order or postganglionic parasympathetic neurons. Preganglionic neurons located in the mesencephalon and medulla oblongata originate from the neuroblasts of the neural tube. Through elongation the axons of these neurons will synapse with postganglionic parasympathetic neurons in the wall of the esophagus.[2, 9]

The gastroesophageal junction is the result of the coordinated development of the esophagus, stomach, and diaphragm and the innervation of these structures.

UPPER ESOPHAGEAL SPHINCTER (UES)

Anatomy

At rest, the proximal esophagus is closed by a functional sphincter that creates a high pressure zone measurable by perfusion manometry[10] (Fig. 2–2). The cricopharyngeus muscle is classically described as responsible for this pressure zone. This muscle is attached anteriorly on both ends of the cricoid cartilage and encircles the proximal end of the esophagus as an uninterrupted muscular sling. Located at the level of the sixth cervical vertebra, the muscle appears as a posterior indentation on a barium swallow (Fig. 2–3). The extensive study by Zaino et al.[8] describes this muscle as independent of the esophageal musculature in most of the dissected specimens. In nearly one-third of the dissections, however, the muscle was fused with the longitudinal layer of the esophagus. Five specimens revealed no muscle distinction at the pharyngoesophageal junction. While the cricopharyngeus muscle can be identified as the extrinsic component of

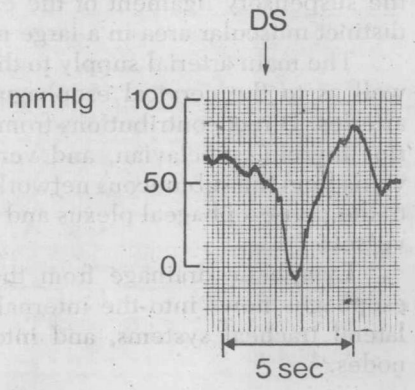

Figure 2-2. The proximal esophageal sphincter. A high pressure zone that is present at rest relaxes on swallowing. (*DS*, dry swallow.)

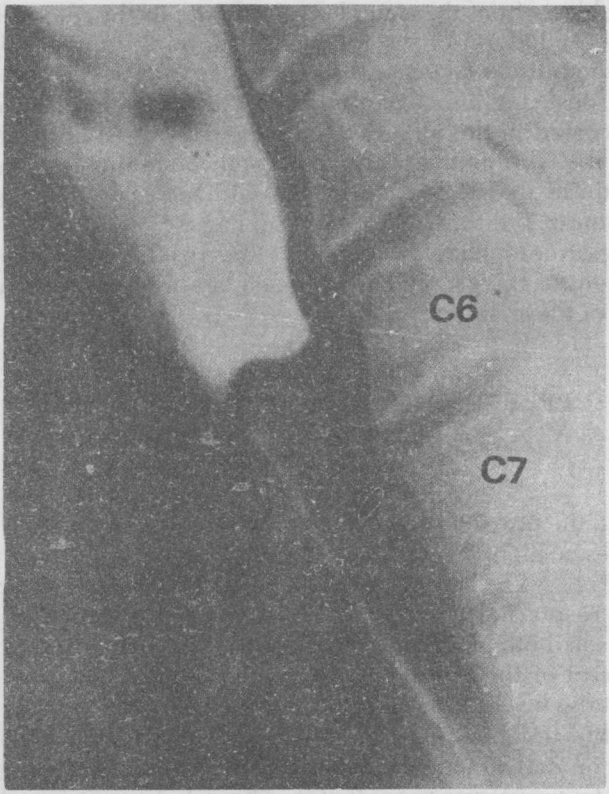

Figure 2-3. Posterior indentation of the cricopharyngeus muscle at the level of the sixth and seventh cervical vertebrae (*C6* and *C7*).

the sphincter (Fig. 2–4A), the innermost circular muscle layer of the very proximal part of the esophagus may be the true intrinsic component of the UES (Fig. 2–4B). This circular musculature is attached to the suspensory ligament of the esophagus and was interpreted as a distinct muscular area in a large majority of Zaino's observations.

The main arterial supply to the proximal esophageal sphincter as well as to the cervical esophagus comes from the inferior thyroid arteries. Direct contributions from the aorta or from the carotid, superior thyroid, subclavian, and vertebral arteries are possible.[11] The veins from the submucous network drain through the wall of the organ to the peri-esophageal plexus and from there into the brachiocephalic vessels.

Lymphatic drainage from the proximal sphincter and cervical esophagus flows into the internal jugular system, the right and left lateral tracheal systems, and into the intertracheobronchial lymph nodes.[12]