

HOSPITAL PHYSICIAN®

GASTROENTEROLOGY BOARD REVIEW MANUAL

PUBLISHING STAFF

PRESIDENT, GROUP PUBLISHER

Bruce M. White

EXECUTIVE EDITOR

Debra Dreger

SENIOR EDITOR

Becky Krumm, ELS

ASSOCIATE EDITOR

Lamont Williams

ASSISTANT EDITOR

Jennifer M. Vander Bush

EDITORIAL ASSISTANT

Renee Autumn Ray

EXECUTIVE VICE PRESIDENT

Barbara T. White, MBA

PRODUCTION DIRECTOR

Suzanne S. Banish

PRODUCTION ASSOCIATES

Tish Berchtold Klus

Christie Grams

Mary Beth Cunney

ADVERTISING/PROJECT MANAGER

Patricia Payne Castle

NOTE FROM THE PUBLISHER:

This publication has been developed without involvement of or review by the American Board of Internal Medicine.



**Endorsed by the
Association for Hospital
Medical Education**

The Association for Hospital Medical Education endorses HOSPITAL PHYSICIAN for the purpose of presenting the latest developments in medical education as they affect residency programs and clinical hospital practice.

Esophageal Motility Disorders

Series Editor:

Robert M. Craig, MD

*Professor of Medicine, Department of Medicine,
Division of Gastroenterology and Hepatology, Northwestern
University Medical School, Chicago, IL*

Contributing Author:

Ikuo Hirano, MD

*Assistant Professor of Medicine, Department of Medicine,
Division of Gastroenterology and Hepatology, Northwestern
University Medical School, Chicago, IL*

Table of Contents

Preface	ii
Introduction	1
Normal Esophageal Motility	1
Idiopathic Achalasia	2
Secondary Achalasia and Pseudoachalasia	7
Diffuse Esophageal Spasm	9
Scleroderma Esophagus	11
Nutcracker Esophagus	12
References	13

Cover Illustration by Christine Schaar

Copyright 2001, Turner White Communications, Inc., 125 Strafford Avenue, Suite 220, Wayne, PA 19087-3391, www.turner-white.com. All rights reserved. No part of this publication may be reproduced, stored in a retrieval system, or transmitted in any form or by any means, mechanical, electronic, photocopying, recording, or otherwise, without the prior written permission of Turner White Communications, Inc. The editors are solely responsible for selecting content. Although the editors take great care to ensure accuracy, Turner White Communications, Inc., will not be liable for any errors of omission or inaccuracies in this publication. Opinions expressed are those of the authors and do not necessarily reflect those of Turner White Communications, Inc.

HOSPITAL PHYSICIAN®

GASTROENTEROLOGY BOARD REVIEW MANUAL

Esophageal Motility Disorders

Series Editor:

Robert M. Craig, MD

Professor of Medicine

Department of Medicine

Division of Gastroenterology and Hepatology

Northwestern University Medical School

Chicago, IL

Contributing Author:

Ikuo Hirano, MD

Assistant Professor of Medicine

Department of Medicine

Division of Gastroenterology and Hepatology

Northwestern University Medical School

Chicago, IL

INTRODUCTION

The 3 classic esophageal motility disorders are achalasia, diffuse esophageal spasm, and scleroderma esophagus. Gastroesophageal reflux disease—one of the most prevalent conditions affecting the Western population—is often classified as an esophageal motility disorder because reflux events result from transient lower esophageal sphincter (LES) relaxations or in association with a hypotensive LES. Additionally, a number of disorders have been *proposed* to be motility disorders of the esophagus, but associations between their respective clinical presentations and the existence of motility abnormalities in the esophagus have not yet been definitively established (**Table 1**). Thus, it is not clear whether these candidate disorders should be considered distinct motility disorders of the esophagus or simply viewed as manometric “curiosities” or nonspecific motility abnormalities. In some instances, these disorders have been observed to transform over time into one of the classic esophageal motility disorders.

This review begins with a brief description of some of the principles of normal esophageal motility. It then provides case discussions that highlight the important pathophysiologic and clinical features of several major esophageal motility disorders: achalasia, diffuse esophageal spasm, scleroderma esophagus, and nutcracker esophagus.

NORMAL ESOPHAGEAL MOTILITY

Esophageal motility can be divided according to the functions of the esophageal body and the LES. The principal function of the esophageal body is that of peristalsis. In general, peristalsis involves the movement of material down a tubular structure by waves of muscular contractions, with each period of contraction (or excitation) being preceded by a period of relaxation (or inhibition). Peristalsis in the esophagus involves sequential contractions that propel liquids or solids from the upper esophageal sphincter through the LES and into the stomach. Peristalsis of the proximal striated muscle portion of the esophagus is regulated by the sequential activation of vagal efferent neurons. Peristalsis of the distal, smooth muscle portion of the esophagus is under the direct control of inhibitory and excitatory neurotransmitters arising from motor neurons situated in the myenteric plexus within the wall of the esophagus.

Relaxation of the distal, smooth muscle portion of the esophagus is primarily mediated by inhibitory neurotransmitters such as nitric oxide, which has recently been identified as a primary inhibitory neurotransmitter in the esophagus. Contractile distal esophageal activity is mediated by excitatory neurotransmitters such as acetylcholine.

In the esophagus, the period of inhibition increases in duration from the proximal to the distal aspects of