

# HOSPITAL PHYSICIAN®

## GENERAL SURGERY BOARD REVIEW MANUAL

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## Gastrointestinal Carcinoid Tumors: Case Studies

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#### I. INTRODUCTION

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Carcinoid tumors are a heterogeneous group of neuroendocrine tumors that can arise in any organ originating from the endoderm. They arise from enterochromaffin or enterochromaffin-like cells (in the amine precursor uptake and decarboxylation system) with the potential to produce several different biologically active amines and peptides. Serotonin is the best known of these biologically active substances. Carcinoid tumors may also secrete corticotropin, histamine, dopamine, substance P, neurotensin, prostaglandins, and kallikrein.<sup>1</sup> The release of vasoactive substances into the systemic circulation is responsible for the development of the carcinoid syndrome, which is a marker of advanced disease (*see* Section VIII.).

The incidence of carcinoid tumors is 1 to 2.5 per 100,000 annually; they are most commonly found in the bronchial tree and the gastrointestinal (GI) tract.<sup>1-4</sup> **Figure 1** depicts the anatomic location of carcinoid tumors as identified in review of the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) database, which accumulated data from 1973 to 1991. In the small intestine and appendix, carcinoid

tumors account for approximately 33% and 77% of all tumors, respectively.<sup>2,3</sup> In organs such as the colon, stomach, and lung that develop tumors more frequently, carcinoid tumors make up only 1% of tumors.<sup>2</sup>

Carcinoid tumors have traditionally been classified according to their presumed derivation from different embryonic divisions of the GI tract. Foregut carcinoid tumors originate in the lungs, bronchi, stomach, or duodenum; midgut tumors in the small intestine, appendix, and proximal large intestine; and hindgut tumors in the distal colon and rectum.<sup>6</sup> The biologic and clinical characteristics vary within these groups. This review will examine GI carcinoid tumors by location including the stomach, duodenum, small intestine, appendix, colon, and rectum. The carcinoid syndrome will also be reviewed. Two case patients are presented to emphasize clinical features and management issues. Sample board review questions with detailed answers are provided at the end of this manual for self-assessment.

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#### II. GASTRIC CARCINOID TUMORS

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Carcinoid tumors of the stomach are uncommon, accounting for 0.54% of all gastric malignancies and