



AAVBC

AMERICAN ACADEMY OF VALUE BASED CARE

Carcinoid Tumors

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Carcinoid tumors are a subtype of **neuroendocrine neoplasms (NENs)** arising from diffuse neuroendocrine cells in the gastrointestinal tract, lungs, and other sites.¹ The 2019 WHO Classification of Tumours of the Digestive System grades these tumors by **Ki-67 proliferative index** and **mitotic rate** into **well-differentiated** G1/G2 neuroendocrine tumors (NETs) and **poorly differentiated** G3 neuroendocrine carcinomas (NECs).^{1,2} Their defining clinical feature is the capacity to secrete vasoactive hormones, primarily serotonin, **driving carcinoid syndrome:** episodic flushing, watery diarrhea, bronchoconstriction, and, over time, carcinoid heart disease (CHD) affecting right-sided cardiac valves. Classical carcinoid syndrome typically requires liver metastases (so hormones bypass hepatic degradation), but primary pulmonary and ovarian carcinoids can secrete **directly into the systemic circulation regardless of liver involvement.**³

ICD-10 Codes:⁴

Malignant carcinoid tumors are coded by site under C7A — small intestine (**C7A.010** duodenum, **C7A.011** jejunum, **C7A.012** ileum, **C7A.019** unspecified small intestine), appendix and large intestine (**C7A.020** appendix, **C7A.021** cecum, **C7A.022** ascending colon, **C7A.023** transverse colon, **C7A.029** large intestine unspecified), bronchus/lung (**C7A.090**), and other sites (**C7A.094** foregut, **C7A.095** midgut, **C7A.096** hindgut, **C7A.098** other).³⁰ Secondary (metastatic) carcinoid tumors are coded **C7B.0x** — most commonly **C7B.02** (liver), **C7B.01** (distant lymph nodes), **C7B.03** (bone), **C7B.04** (peritoneum). Benign carcinoid tumors (**D3A.0x**) and carcinoid syndrome (**E34.0**; **E34.01** with cardiac involvement) do not map to any HCC. Personal history (**Z85.030**) applies only after definitive treatment with no active disease. Code **E34.0** is always an additional code beside the tumor classification, never the sole code.

Prevalence and Burden: Approximately **8,000** new GI NETs are diagnosed annually in the United States. NET incidence rose from **1.09 per 100,000** in 1973 to **6.98 per 100,000** in 2012, with continued increases through 2021.⁵ In patients **≥65**, **incidence reached 28.44 per 100,000** by 2021 — a greater than 5-fold increase from earlier decades.⁵ GI carcinoids often present in the early 60s. **Primary sites: GI tract >50% (principally ileum, appendix, rectum); lungs/bronchus ~27%, and ~6% in the pancreas.**⁶ Incidence is slightly higher in **women** and in **Black Americans.**⁷

HCC/RAF V28 Mapping

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
C7A.0x (localized,primary)	HCC 22 — Bladder, Colorectal and other Cancers	0.363	Active malignant carcinoid tumor. State 'malignant neuroendocrine tumor' or 'malignant carcinoid' explicitly. Specify primary site (ileum, appendix, lung, etc.) and grade (G1, G2, G3) per WHO 2019

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