AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Carcinoid Tumors

Quick Reference Guide

2026

Table of Contents

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