AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Small Intestine Cancer

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: **Small intestine cancer** (small bowel cancer) is a group of rare malignancies of the **duodenum**, **jejunum**, and **ileum**.[1] Although the small intestine makes up roughly **75%** of the length of the gastrointestinal tract, these tumors account for fewer than **3%** of GI neoplasms and about **0.6%** of all new US cancers.[1,2] **Four histologies dominate** and each behaves, stages, and codes differently: **adenocarcinoma** (most common, typically duodenal),[3,4] **neuroendocrine tumors** (NETs/carcinoids, most often ileal),[1] **gastrointestinal stromal tumors** (GIST), and **lymphoma**.[5] The defining clinical problem is **diagnostic delay**: nonspecific symptoms mimic benign disease, so adenocarcinoma is delayed an average of **6–8 months** from symptom onset and small bowel NETs up to **36 months**.[1,6]

ICD-10 Codes:

Small bowel adenocarcinoma is reported with anatomic-subsite codes: **C17.0** (duodenum), **C17.1** (jejunum), **C17.2** (ileum), **C17.3** (malignant Meckel diverticulum), **C17.8** (overlapping sites), and **C17.9** (small intestine, unspecified).[7] **Histology drives code family**:[3,7–10] neuroendocrine/carcinoid tumors use the **C7A.019** series (not **C17.x**), GIST uses **C49.A3**, and confirmed metastasis to the small bowel uses **C78.4**. The single most common coding error is defaulting to **C17.9** when pathology or imaging **has identified the subsite** → document the anatomic location **explicitly** (e.g., 'adenocarcinoma of the jejunum'). Code complications and surgical status separately **at every encounter**: malignant bowel obstruction (**K56.69**), cancer-related anemia (**D63.0**, not **D64.9** unspecified), and ileostomy status (**Z93.2**).[7] Reserve **Z85.068** (personal history) only for **true remission with no active treatment**, surveillance, or complications; premature use during adjuvant therapy or watchful waiting **eliminates the active-malignancy picture entirely**.

Prevalence and Burden: The National Cancer Institute estimates approximately **14,450** new cases and **2,170** deaths from small intestine cancer in the US in 2026.[11] The global age-standardized incidence is about **0.60 per 100,000**, and incidence has been rising for both adenocarcinoma and NETs over the past two decades.[2] Median age at diagnosis is in the **6th decade**, with peak incidence in adults **60–79 years**.[4,12] **Age itself confers risk**: the hazard ratio for adenocarcinoma is **3.12** (95% CI 1.33–7.31) in adults ≥65 versus 50–55 years, and increasing age is an independent predictor of mortality (**HR 1.1** per year).[5,13] SEER data show a median survival of **11 months** in patients >60 versus **24 months** in those ≤60 ($p<0.0001$).[13]

HCC/RAF V28 Mapping

ICD-10 CODE(S)[7]	HCC CATEGORY (V28)[14]	RAF (CNA)*[15]	DOCUMENTATION REQUIREMENT[3,7,8]
C17.0/C17.1/C17.2 Malignant neoplasm of duodenum/ jejunum/ileum	**HCC 22** Bladder/ Colorectal/and **Other Cancers**	0.363	Active SBA confirmed by pathology; **specify subsite** (duodenum, jejunum, or ileum); **document histologic subtype and AJCC 8th Edition stage**; for duodenum, distinguish from **ampullary origin**