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 21 — Lymphoma and Other Cancers	0.671	**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

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
C7A.1 or C7A.8 (poorly differentiated malignant, high grade)	HCC 21	0.671	Document poorly differentiated features, aggressive, small/large cell morphology, or high-grade neuroendocrine carcinoma. Specify the primary site and active treatment status
C7B.0x (secondary carcinoid tumors)	HCC 17 — Cancer Metastatic to Lung/ Liver/Brain	4.209	**Secondary carcinoid** (most commonly **C7B.02** liver). **MUST code alongside the primary C7A code**; document site of metastasis
D3A.0x (benign)	No HCC	—	**AVOID: Benign carcinoid tumors do NOT map to any HCC in V28.** When pathology confirms malignancy (invasion, metastasis, high-grade features), transition to C7A
E34.0 (Carcinoid syndrome, unspecified)	No HCC (additional code only)	—	**Carcinoid syndrome** — always code alongside the tumor code **(C7A/C7B)**, never as the sole code. **E34.01** specifically documents carcinoid heart syndrome
E34.01(Carcinoid heart syndrome)/	HCC 17	4.209	Hormone-secreting tumors cause **damage to the heart valves**, leading to valve dysfunction. Document echocardiographic findings (valve involvement, severity, structural changes), biomarkers (5-HIAA, CgA, NT-proBNP), primary NET location/ stage, and heart failure status with functional class

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
E34.09(Other carcinoid syndrome)	HCC 17	4.209	Document specific manifestations (flushing, diarrhea, bronchospasm, skin changes), elevated biomarkers (24-hr 5-HIAA or CgA), primary NET site/treatment status, and a normal echo **ruling out carcinoid heart disease** (E34.01)
Z85.030 (history)	No HCC	—	**AVOID: Personal history of malignant carcinoid.** Use ONLY after definitive treatment with no active disease and no ongoing surveillance for known disease. Active surveillance patients retain C7A
C7A.00 (Malignant carcinoid tumor of unspecified site, avoid)	HCC 21	0.671	**Unspecified site** — avoid when documentation supports a specific site. Use C7A.012 (ileum), C7A.020 (appendix), C7A.090 (bronchus/lung), etc.

ABBREVIATIONS: AML = acute myeloid leukemia; CgA = chromogranin A; CMS-HCC V28 = Centers for Medicare & Medicaid Services Hierarchical Condition Category Model V28; CNA = Community Non-Dual Eligible, Aged segment; HCC = Hierarchical Condition Category; ICD-10 = International Classification of Diseases, 10th Revision; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; RAF = Risk Adjustment Factor; WHO = World Health Organization

***RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting**

Risk-Adjusted Care Resources per Patient/Year[8,9]

Risk-adjusted care resource allocation — MA base rate (~$10,402.34) × RAF coefficient

Carcinoid Tumors (Malignant/Local)
$6,980
HCC 21 · RAF 0.671

Carcinoid Tumors (Secondary)
$43,780
HCC 17 · RAF 4.209

AAVBC PERSPECTIVE

AAVBC encourages primary care providers to approach carcinoid tumors as a complex, potentially fatal condition requiring specialized multidisciplinary staging and long-term management, rather than a condition that is inherently slow-growing or self-limiting. ***Not all***

***neuroendocrine tumors follow an indolent course**, and all require formal staging regardless of grade. In elderly patients, even localized low-grade tumors carry meaningful prognostic weight due to competing comorbidities, and the clinical framework must reflect this.*
AAVBC supports a management approach centered on preventing avoidable hospitalizations, minimizing polypharmacy-driven complications, and preserving functional quality of life, rather than the tumor-suppression framework that standard oncology guidelines prioritize.
*This shift is particularly relevant in the geriatric population, where **aggressive intervention may compound comorbidity burden without proportionate benefit**.*
*AAVBC encourages providers to maintain active longitudinal ownership of these patients in partnership with oncology and endocrinology, ensuring that management decisions are guided by functional status and quality of life alongside tumor biology. **This helps ensure the right patients receive the right treatment at the right time.***

2 RECOGNITION AND DIAGNOSIS

Medicare Screenings and Diagnostic Testing (Adult and Geriatric)

TEST	COVERAGE	FREQUENCY	CPT/HCPCS	CLINICAL INDICATION
No population-based carcinoid screening[10]	Not covered — no Medicare or guideline-endorsed screening	—	—	**Carcinoid is too rare for population screening**; incidental detection (e.g., colonoscopy under HEDIS COL) is the most common asymptomatic entry point
24-hour urine 5-HIAA[10]	Medicare Part B; medically necessary	Baseline; q3–12 months per NCCN surveillance; pre-collection: avoid avocados, bananas, pineapples, eggplant, tomatoes, walnuts 48 hours prior	83497	**Initial syndrome screening** (flushing + diarrhea + wheezing); ongoing surveillance; ≥300 µmol/24h triggers cardiac screening[11]

TEST	COVERAGE	FREQUENCY	CPT/HCPCS	CLINICAL INDICATION
Serum chromogranin A (CgA)[10]	Medicare Part B; medically necessary	Baseline; serial during surveillance	84439	**Initial syndrome workup**; trend monitoring; interpret with caution — PPIs, renal failure, and cardiac disease cause false elevations
Cross-sectional imaging (multiphase CT or MRI abdomen/ pelvis)[10]	Medicare Part B; medically necessary	Baseline; q3–12 months active treatment; q6–12 months stable on SSA ≥2 years	74177 (CT)/ 74183 (MRI)	Localization, staging, and treatment response monitoring
Somatostatin-receptor PET/CT (^{68}Ga-DOTATATE)[10]	Medicare Part B; medically necessary	At staging; before PRRT eligibility evaluation; at significant clinical change	78814/ G0273 (DOTATATE)	**Defines SSTR-positive status**; required for PRRT eligibility (uptake > normal liver, Krenning scale)
Transthoracic echocardiogram (TTE)[10]	Medicare Part B; medically necessary	Baseline at carcinoid syndrome diagnosis; q1–3 years without CHD; at least annually with established CHD	93306	NT-proBNP >260 pg/ mL, urinary 5-HIAA ≥300 µmol/24h, new cardiac murmur, or new dyspnea/edema → **urgent TTE**[11]
NT-proBNP[11]	Medicare Part B; medically necessary	Baseline at carcinoid syndrome diagnosis; serial	83880	>260 pg/mL triggers urgent TTE for CHD evaluation
Serum tryptophan and niacin/vitamin B3[10]	Medicare Part B; medically necessary	Baseline in all carcinoid syndrome patients; during SSA therapy	84510 (tryptophan) 84591 (niacin)	**Detect tryptophan diversion to serotonin synthesis**; preempt pellagra; routine B3 supplementation in elderly

TEST	COVERAGE	FREQUENCY	CPT/HCPCS	CLINICAL INDICATION
Comprehensive Geriatric Assessment (CGA)[12]	Bundled into E/M (no specific CPT)	Before treatment intensification; periodically	Embed in 99213/ 99214 E/M	**Guides treatment intensity, polypharmacy review, and SSA tolerability monitoring in frail patients**
Advance Care Planning (AWV)[10]	Medicare Part B; no copay during AWV	Annual + at major changes	99497/ 99498	**Goals-of-care alignment**; particularly relevant in G3/NEC and progressive metastatic disease

ABBREVIATIONS: ACP = advance care planning; AWV = Annual Wellness Visit; CGA = Comprehensive Geriatric Assessment; CHD = carcinoid heart disease; CMS = Centers for Medicare & Medicaid Services; CPT = Current Procedural Terminology; CT = computed tomography; E/M = evaluation and management; HCPCS = Healthcare Common Procedure Coding System; 5-HIAA = 5-hydroxyindoleacetic acid; HEDIS = Healthcare Effectiveness Data and Information Set; MRI = magnetic resonance imaging; PPI = proton pump inhibitor; PRRT = peptide receptor radionuclide therapy; SSA = somatostatin analog; SSTR = somatostatin receptor; TTE = transthoracic echocardiogram

Carcinoid Syndrome Signs and Atypical Presentations

SIGN/SYMPTOM	CLINICAL SIGNIFICANCE
Episodic facial/upper-chest flushing (dry, not sweat-related)[10]	**Hallmark of carcinoid syndrome** — often misattributed to rosacea, menopause, or anxiety in older adults
Unexplained watery diarrhea persisting beyond 4 weeks[10]	**Most common syndromic symptom**; frequently misdiagnosed as IBS or microscopic colitis
Adult-onset wheezing or bronchoconstriction without atopic history[10]	**Carcinoid-driven bronchospasm**; often misattributed to COPD or late-onset asthma in elderly
Dyspnea + peripheral edema in patient with carcinoid syndrome[10]	**Suggests carcinoid heart disease** — fibrotic right-sided valve damage; clinical overlap with CHF/ COPD masks the diagnosis
Pellagra triad — dermatitis, diarrhea, dementia[13]	Niacin deficiency from tryptophan diversion to serotonin synthesis; **particularly common in elderly patients**

SIGN/SYMPTOM	CLINICAL SIGNIFICANCE
Right upper quadrant pain or hepatomegaly[10]	**Often the first sign of liver metastasis (C7B.02)**; palpate the liver in every patient with suspected carcinoid syndrome
Unexplained anxiety, palpitations, or new-onset tachycardia[14]	Vasoactive hormone release may **mimic primary anxiety disorder** — consider syndrome screening before psychiatric attribution
Cognitive impairment or new confusion in older adult with chronic diarrhea[15]	**May reflect pellagra** (niacin deficiency) **or carcinoid-related dehydration**; screen tryptophan/niacin and rehydrate
Refractory peptic ulcer disease or recurrent GERD[10]	May reflect **gastrin-secreting NET** (Zollinger-Ellison) — check fasting gastrin off PPI and consider SSTR-PET
Rectal bleeding with incidental NET on colonoscopy[16]	**Rectal NETs** are commonly found incidentally during colorectal cancer screening (HEDIS COL); workup before assuming benign

ABBREVIATIONS: CHF = congestive heart failure; COPD = chronic obstructive pulmonary disease; GERD = gastroesophageal reflux disease; IBS = irritable bowel syndrome; NET = neuroendocrine tumor; PPI = proton pump inhibitor; SSTR-PET = somatostatin receptor positron emission tomography

Geriatric Risk Factors

FACTOR	RISK SIGNAL	NOTES
Age ≥65 years	Incidence **28.44 per 100,000** by 2021 (5-fold increase)[5]	Mean age at diagnosis **61.4 years**[5]
Age >70 years	HR 1.66 (95% CI 1.26–2.18);[17] Median OS 28 months across all NETs vs. substantially longer in younger cohorts after adjusting for stage and grade[18]	**Frailty and limited physiologic reserve** constrain treatment intensity
Female sex[5]	**Slightly higher** incidence than men overall	Lung NETs may present earlier in women (ages 45–55)
Black American race[19]	**Higher incidence rates** compared to White Americans	**Document race/ethnicity at baseline**; address SDOH
First-degree family history of NET[10]	MEN1, MEN2A, NF1, VHL syndromes predispose to NET	**Refer for genetics evaluation** if family history of pituitary, parathyroid, pancreatic, or adrenal tumors

FACTOR	RISK SIGNAL	NOTES
MEN1 (multiple endocrine neoplasia type 1)[10]	Strong NET predisposition (pancreatic, duodenal, and lung NETs)	**All MEN1 patients require lifelong NET surveillance per NCCN**
Type 2 diabetes mellitus	Modestly **elevated NET risk** (OR 2.76 for gastric NET in some series)[20]	Coexists frequently with SSA-induced dysglycemia; monitor closely
Long-standing atrophic gastritis or PPI use[21]	Type 1 gastric NETs arise in chronic hypergastrinemia from atrophic gastritis	**Distinguish low-risk type 1 from aggressive type 3 gastric NETs**; gastrin level + biopsy
Prior carcinoid diagnosis[10]	Multifocal disease is common in midgut NETs; full bowel evaluation at diagnosis	**Annual surveillance imaging** per NCCN even after curative resection
Tobacco/smoking history (lung NETs)[22]	Atypical carcinoid more associated with smoking than typical carcinoid	**Smoking cessation counseling at every encounter**

ABBREVIATIONS: CI = confidence interval; HR = hazard ratio; MEN = multiple endocrine neoplasia; NET = neuroendocrine tumor; NF1 = neurofibromatosis type 1; OR = odds ratio; OS = overall survival; PPI = proton pump inhibitor; SDOH = social determinants of health; VHL = von Hippel-Lindau

Diagnostic Thresholds and Classification

TEST/SYSTEM	DIAGNOSTIC CRITERION	NOTES
24-hour urine 5-HIAA	**Sensitivity ~73%, specificity ~100%** for carcinoid syndrome; dietary restriction 48 hours prior is essential[23]	**Elevated >300 µmol/24h triggers cardiac (CHD) screening** per Davar et al. JACC expert statement[11]
Serum chromogranin A (CgA)	**Sensitivity 68–100%** depending on tumor burden; rises with tumor progression[23]	**PPIs cause false elevation** — discontinue ≥1–2 weeks before measurement when feasible; renal failure also falsely elevates CgA10

TEST/SYSTEM	DIAGNOSTIC CRITERION	NOTES
WHO 2019 grading (Ki-67 + mitotic rate)	**G1:** Ki-67 <3%, mitoses <2/10 HPF; **G2:** Ki-67 3–20%, mitoses 2–20/10 HPF **G3:** Ki-67 >20% **(NET if well-differentiated; NEC if poorly differentiated)**, mitoses >20/10 HPF[10,24]	**Grade drives treatment selection; G3 NEC requires platinum-based therapy** similar to small-cell lung cancer rather than SSA-based pathways
Cross-sectional imaging (multiphase CT or MRI)[10]	Primary localization and staging; multiphase essential for hypervascular liver metastases	**Detection sensitivity varies by site**; small ileal primaries may be missed
Somatostatin-receptor PET/CT (^{68}**Ga-DOTATATE)**[10]	Krenning scale: uptake greater than normal liver **indicates SSTR-positive** disease eligible for SSA therapy and PRRT	**Higher sensitivity than older OctreoScan**; standard at initial staging and PRRT planning
Endoscopic ultrasound (pancreatic and duodenal NETs)[10]	Localizes small **(<2 cm)** pancreatic NETs not seen on cross-sectional imaging	Tissue acquisition for grading and molecular studies
Echocardiography (TTE)[11]	Tricuspid regurgitation, pulmonary stenosis, right-sided valve thickening =carcinoid heart disease	Baseline at syndrome diagnosis; q1–3 yr without CHD; ≥annual with established CHD
NT-proBNP[11]	**>260 pg/mL** triggers urgent TTE for CHD evaluation	**Easier to obtain than baseline echo**; serial monitoring during SSA therapy
Comprehensive Geriatric Assessment (CGA)[12]	Function (ADL/IADL, TUG), cognition (MoCA/MMSE), comorbidity (Charlson), polypharmacy, nutrition, social support	**Guides treatment intensity calibration for elderly NET patients**

ABBREVIATIONS: ADL = activities of daily living; CgA = chromogranin A; CGA = Comprehensive Geriatric Assessment; CHD = carcinoid heart disease; CT = computed tomography; HPF = high-power field; 5-HIAA = 5-hydroxyindoleacetic acid; IADL = instrumental activities of daily living; MMSE = Mini-Mental State Examination; MoCA = Montreal Cognitive Assessment; MRI = magnetic resonance imaging; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PET = positron emission tomography; PPI = proton pump inhibitor; PRRT = peptide receptor radionuclide therapy; SSA = somatostatin analog; SSTR = somatostatin receptor; TTE = transthoracic echocardiogram; TUG = Timed Up and Go; WHO = World Health Organization

Clues to Dig Deeper

CLINICAL CLUE	WHY IT MATTERS	NEXT DIAGNOSTIC STEP
Patient with two or more of: episodic flushing, watery diarrhea, wheezing[10]	**Most common missed diagnostic pattern in primary care**	**24-hour urine 5-HIAA + serum CgA**; GI/oncology referral within 1–2 weeks
Diarrhea attributed to IBS not responding to standard management[25]	Persistent unexplained diarrhea may reflect carcinoid syndrome rather than functional disorder	**Stool studies + 5-HIAA + CgA**; small-bowel imaging if biomarkers elevated
New 'asthma' diagnosed after age 50 without prior atopic history[14]	**Adult-onset wheezing in elderly should prompt syndrome screening**; bronchoconstriction is a classic carcinoid feature	**5-HIAA + CgA**; chest CT if persistent and screening positive
Right-sided heart failure of unclear etiology in patient with chronic flushing/diarrhea[11]	Carcinoid heart disease causes right-sided valve fibrosis distinct from left-sided HF; symptoms mimic CHF and COPD	TTE + NT-proBNP; 5-HIAA + CgA; **cardio-oncology referral if CHD confirmed**
Dementia + diarrhea + dermatitis in elderly carcinoid patient[26]	Classic pellagra triad from niacin deficiency — tryptophan diverted to serotonin synthesis	Serum tryptophan and niacin; B3 supplementation; engage caregiver for adherence
Incidental NET on colonoscopy or CT[16]	Rectal and appendiceal NETs are commonly found incidentally; small ileal primaries may be subtle on imaging	**Endoscopic pathology + Ki-67 grading**; staging CT and SSTR-PET; multidisciplinary NET center referral
Patient on long-term PPI with elevated CgA[27]	PPIs cause false-positive CgA elevation; misinterpretation leads to unwarranted escalation	**Hold PPI ≥1–2 weeks if clinically safe**; repeat CgA; interpret in context with imaging and 5-HIAA
Family history of pituitary, parathyroid, pancreatic, or adrenal tumors[20]	MEN1 syndrome — strong predisposition to multiple NETs	**Genetics referral**; full endocrine workup; lifelong NET surveillance

ABBREVIATIONS: CgA = chromogranin A; CHD = carcinoid heart disease; CHF = congestive heart failure; CT = computed tomography; COPD = chronic obstructive pulmonary disease; GI = gastrointestinal; HF = heart failure; 5-HIAA = 5-hydroxyindoleacetic acid; IBS = irritable bowel syndrome; MEN1 = multiple endocrine neoplasia type 1; NET = neuroendocrine tumor; PPI = proton pump inhibitor; SSTR-PET = somatostatin receptor positron emission tomography; TTE = transthoracic echocardiogram

Common Oversights

OVERSIGHT	WHY IT MATTERS — WHAT TO DO INSTEAD
Treating chronic diarrhea, adult-onset wheezing, and flushing as isolated conditions[10]	**These are syndrome-screening triggers** — order 24-hour urine 5-HIAA and serum CgA when two or more co-occur; refer GI/oncology within 1–2 weeks
Assuming 'carcinoid' means benign and slow-growing	The term is largely archaic; G3 NEC is highly aggressive (median OS ~10 months metastatic). **Behavior is defined by grade**, not the historical label[24,28]
Aggressive antidiarrheal use (loperamide) in frail elderly to chase normal bowel movements[10]	**Over-aggressive use causes ileus, AKI, and dehydration hospitalizations**. Optimize SSA dosing first; target symptom control, not biochemical normalization[1,23]
Using epinephrine or beta-agonists for hypotension in suspected carcinoid crisis[29]	Catecholamines and beta-agonists can precipitate or worsen crisis by triggering massive hormone release — **preferred vasopressors are vasopressin, phenylephrine**
Failing to flag the carcinoid diagnosis prominently in the chart[11]	Anesthesia, pulmonology, and emergency providers may inadvertently administer contraindicated agents; **ensure problem list highlights diagnosis and rescue SC octreotide**
Interpreting elevated CgA without checking for PPI use[27]	**PPIs falsely elevate CgA**; renal failure and cardiac disease also elevate. Hold PPI ≥1–2 weeks when clinically safe before measurement
Skipping TTE in carcinoid syndrome patients[11]	**Carcinoid heart disease is underdiagnosed**; baseline TTE at syndrome diagnosis and q1–3 years thereafter is standard. NT-proBNP >260 pg/mL or 5-HIAA ≥300 µmol/24h triggers urgent TTE
Not screening for niacin deficiency/pellagra[30]	**Tryptophan diversion to serotonin synthesis depletes niacin**; routine B3 supplementation and serum tryptophan/niacin monitoring should be standard in carcinoid syndrome patients, particularly elderly

OVERSIGHT	WHY IT MATTERS — WHAT TO DO INSTEAD

ABBREVIATIONS: AKI = acute kidney injury; CgA = chromogranin A; CHD = carcinoid heart disease; GI = gastrointestinal; HCC = Hierarchical Condition Category; 5-HIAA = 5-hydroxyindoleacetic acid; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; NT-proBNP = N-terminal pro–B-type natriuretic peptide; OS = overall survival; PPI = proton pump inhibitor; RAF = Risk Adjustment Factor; SC = subcutaneous; SSA = somatostatin analog; TTE = transthoracic echocardiogram

Key Differentials in Elderly

PRESENTATION	DIFFERENTIAL	KEY TESTS
Chronic episodic diarrhea in patient ≥55[31]	Carcinoid syndrome vs IBS-D **vs** microscopic colitis vs bile acid malabsorption vs pancreatic exocrine insufficiency vs small-bowel bacterial overgrowth vs medication-induced	**24-hour urine 5-HIAA + serum CgA**; stool studies; fecal elastase; SeHCAT or trial of cholestyramine
Adult-onset wheezing without atopic history[23]	Carcinoid-related bronchospasm **vs** late-onset asthma vs COPD vs cardiac asthma vs tracheomalacia vs vocal cord dysfunction	**Spirometry with bronchodilator**; chest CT if persistent; 5-HIAA + CgA if other syndromic features present
Recurrent unexplained flushing[32]	Carcinoid syndrome **vs** rosacea vs menopausal vasomotor symptoms vs mastocytosis vs pheochromocytoma vs medullary thyroid carcinoma vs alcohol vs medications (niacin, calcium channel blockers)	**5-HIAA + CgA**; metanephrines if pheochromocytoma suspected; tryptase if mastocytosis
Right-sided heart failure with chronic diarrhea/flushing[11]	Carcinoid heart disease **vs** primary pulmonary hypertension vs left-heart disease with secondary RV failure vs constrictive pericarditis vs restrictive cardiomyopathy	TTE + NT-proBNP; 5-HIAA + CgA; **right-heart cath if PH suspected**
Hepatomegaly with elevated liver enzymes[10]	Liver metastasis from C7A primary **vs** primary HCC vs cholangiocarcinoma vs hepatic adenoma vs cysts vs congestive hepatopathy	**Multiphase CT or MRI liver**; AFP; serum CgA; liver biopsy if indeterminate
Dementia + diarrhea + dermatitis in carcinoid patient[33]	Pellagra (niacin deficiency) **vs** B12 deficiency vs vitamin D deficiency vs medication-induced cognitive impairment	Serum tryptophan + niacin; B12; vitamin D; medication reconciliation[1]

PRESENTATION	DIFFERENTIAL	KEY TESTS

ABBREVIATIONS: AFP = alpha-fetoprotein; CgA = chromogranin A; COPD = chronic obstructive pulmonary disease; CT = computed tomography; HCC = hepatocellular carcinoma; 5-HIAA = 5-hydroxyindoleacetic acid; IBS-D = irritable bowel syndrome with diarrhea; MRI = magnetic resonance imaging; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PH = pulmonary hypertension; RV = right ventricular; SeHCAT = selenium-75 homotaurocholic acid test; TTE = transthoracic echocardiogram

Comorbidity Screening

CONDITION*	PREVALENCE/ASSOCIATION	SCREENING APPROACH
Carcinoid heart disease (E34.01)[11]	Develops in patients with chronic carcinoid syndrome and hormone exposure; right-sided valve fibrosis	Baseline TTE at syndrome diagnosis; q1–3 years without CHD; ≥annually with CHD; **NT-proBNP >260 pg/mL** or **5-HIAA ≥300 µmol/24h → urgent TTE**
Pellagra/niacin deficiency[13,30]	Tryptophan diversion to serotonin synthesis depletes B3; particularly common in elderly with high tumor burden	**Serum tryptophan and niacin at baseline**; routine B3 supplementation; engage caregiver for adherence
Cholelithiasis (long-term SSA use)[34]	Common with sustained octreotide/lanreotide therapy	**Hepatobiliary imaging q1–2 years on long-term SSA**; symptomatic stones may require GI/surgery referral
Glucose dysregulation (SSA-induced)[35]	SSAs can cause both hyperglycemia and hypoglycemia depending on baseline pancreatic function	**Glucose at SSA initiation**; quarterly during stable therapy; adjust antidiabetic regimen as needed
Bowel obstruction/mesenteric fibrosis[14]	Midgut NETs can cause desmoplastic mesenteric fibrosis leading to obstruction, mesenteric ischemia, or bleeding	**Imaging-based surveillance per NCCN**; surgical evaluation for primary tumor resection in low-burden disease

CONDITION*	PREVALENCE/ASSOCIATION	SCREENING APPROACH
Polypharmacy[36]	SSAs, telotristat, antidiarrheals, cardiac medications, antidiabetics, AEDs create high interaction risk	**Medication reconciliation every visit**; pharmacist review before treatment escalation; deprescribing review
Atrial fibrillation or other arrhythmias[11]	Carcinoid heart disease and electrolyte derangements from chronic diarrhea increase arrhythmia risk	**Baseline ECG**; monitor electrolytes (K, Mg) during chronic diarrhea; anticoagulation per CHA_2DS_2-VASc
Renal impairment[10,37]	Common in elderly; required GFR ≥30 for PRRT; affects gadolinium contrast administration	**Baseline eGFR**; before PRRT; at every SSA visit during active diarrhea; minimize nephrotoxic co-administration
Depression/anxiety[38]	**Common in cancer patients**; vasoactive hormones may mimic primary anxiety	**PHQ-9/GAD-7** baseline and during treatment; SSRIs generally safe
Frailty (CGA-defined)[36]	Limits treatment intensity; informs SSA titration and PRRT/systemic-therapy eligibility	**G-8 screening**; full CGA when indicated; reduced-intensity treatment plans for frail patients

ABBREVIATIONS: AED = antiepileptic drug; CGA = Comprehensive Geriatric Assessment; CHA_2DS_2-VASc = Congestive heart failure, Hypertension, Age, Diabetes, Stroke, Vascular disease, Sex; CHD = carcinoid heart disease; ECG = electrocardiogram; eGFR = estimated glomerular filtration rate; GAD-7 = Generalized Anxiety Disorder-7; G-8 = Geriatric 8 screening; 5-HIAA = 5-hydroxyindoleacetic acid; K = potassium; Mg = magnesium; NCCN = National Comprehensive Cancer Network; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PHQ-9 = Patient Health Questionnaire-9; PRRT = peptide receptor radionuclide therapy; SSA = somatostatin analog; SSRI = selective serotonin reuptake inhibitor; TTE = transthoracic echocardiogram

***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

⚠ RED FLAG — EMERGENCY/ACTIVATE CRISIS RESPONSE

- **Carcinoid crisis** — sudden severe flushing + hypotension + tachycardia + bronchospasm → Call 911 immediately; administer **SC octreotide rescue 100–500 µg** if prescribed
- **Epinephrine and beta-agonists are CONTRAINDICATED** as vasopressors in carcinoid crisis — precipitate or worsen hormone release → Preferred vasopressors: **vasopressin, phenylephrine, or angiotensin II**
- **Acute severe abdominal pain** with distension and no bowel movement — may reflect mesenteric fibrosis with obstruction or ischemia → Emergency evaluation
- **Acute decompensated right-sided heart failure** — severe dyspnea, ascites, or anasarca in known carcinoid heart disease → ED transfer
- **Sudden confusion or severe cognitive change** — may reflect pellagra, brain metastasis, or carcinoid crisis → Urgent evaluation
- Ensure **carcinoid diagnosis is flagged prominently in the chart** — anesthesia, pulmonology, and emergency providers must avoid contraindicated agents

⚠ RED FLAG — URGENT (24-72 HOURS)

- **NT-proBNP >260 pg/mL** or **urinary 5-HIAA ≥300 µmol/24h** in carcinoid syndrome patient → Urgent TTE to evaluate for carcinoid heart disease
- **New or progressive right-sided heart failure signs** (dyspnea on exertion, peripheral edema, ascites) in known carcinoid → Urgent TTE and cardio-oncology
- Referral **rising chromogranin A trend** or **new abdominal pain** in active disease → Expedited imaging and oncology contact
- **New hepatomegaly or right upper quadrant pain** — evaluate for liver metastasis → Multiphase CT or MRI; code **C7B.02** if confirmed
- **Severe refractory diarrhea** unresponsive to SSA optimization → Assess for non-carcinoid causes — pancreatic exocrine insufficiency, bile acid malabsorption, short-gut syndrome — before further escalation
- **Carcinoid syndrome patient requiring elective surgery, endoscopy, or anesthesia**
  → Pre-procedure SSA prophylaxis and crisis-prevention protocol communication required

3 MEAT DOCUMENTATION ESSENTIALS

Accurate carcinoid tumor documentation supports clinical continuity, quality measurement, and care coordination across specialties. Common documentation gaps include: coding **D3A** (benign) when pathology confirms malignant behavior; using **E34.0** (carcinoid syndrome) as the sole code without a corresponding tumor code; applying **Z85.030** (personal history) while the patient remains under active surveillance or an active treatment plan; omitting **C7B.02** when liver metastases are documented; and failing to specify primary site and histologic grade per WHO 2019 classification. Each of these gaps misrepresents the patient's clinical status and undermines coordinated decision-making with oncology, gastroenterology, cardiology, and palliative care.

Case vignette: *A 72-year-old female presents to her PCP with 6 months of episodic facial flushing (not menopausal — patient is **22 years** post-menopause), watery diarrhea attributed to IBS for 4 years that has worsened, and new wheezing diagnosed as adult-onset asthma without atopic history. Past medical history: hypertension on amlodipine, hyperlipidemia on atorvastatin, GERD on omeprazole. No prior cancer. Functional status: independent ADLs; G-8 score 13 (borderline). PCP recognizes the symptom triad and orders syndrome screening.*

MONITOR: "Presenting symptom triad: **episodic facial flushing** (onset 6 months ago; not menopausal — **22 years post-menopause**), **chronic watery diarrhea** (4 years, previously attributed to IBS, now worsening), and **new wheezing** (diagnosed adult-onset asthma without atopic history). BP 132/80 mmHg**. Weight stable. No hepatomegaly on abdominal exam.** Functional status: independent ADLs. **G-8 score 13** — borderline frailty signal; monitoring functional trajectory. No prior NET diagnosis. Symptom pattern prompted syndrome screening."

EVALUATE: "**24-hour urine 5-HIAA elevated at 195 µmol/24h** (reference <50 µmol/24h). **Serum CgA 480 µg/L** — omeprazole held **2 weeks prior** to draw to avoid false elevation; substituted H2-blocker during washout period. Multiphase CT abdomen/pelvis (date): **2.1 cm** distal ileal mass; three hypervascular liver lesions, largest **4.8 cm**. 68Ga-DOTATATE PET/CT (date): intense SSTR-positive uptake **(Krenning score 4)** at ileal primary and all hepatic lesions. Baseline TTE (date): mild tricuspid regurgitation noted; no significant valve thickening; **NT-proBNP 198 pg/mL**— below 260 pg/mL threshold for urgent cardiology referral; annual TTE surveillance initiated. Baseline serum tryptophan and niacin obtained. WHO 2019 histologic grade pending biopsy; imaging pattern consistent with G1 vs. G2."

ASSESS: "Malignant carcinoid tumor of the ileum **(C7A.012)**; secondary carcinoid tumor of liver **(C7B.02)**; carcinoid syndrome **(E34.0)**; essential hypertension **(I10)**; hyperlipidemia **(E78.5)**; GERD **(K21.9)**. Carcinoid diagnosis flagged prominently in problem list with EHR-level alerts —anesthesia, pulmonology, and emergency providers must avoid epinephrine and beta-agonists. Referred to multidisciplinary NET center; endocrinology and oncology engaged."

TREAT: "**Octreotide LAR 30 mg IM every 4 weeks** initiated per NCCN; short-acting octreotide **100–250 µg** SC TID prescribed as bridge during **10–14-day LAR onset window**. SC octreotide rescue prescription documented in chart and reviewed with caregiver. **Niacin (vitamin B3) supplementation** initiated to preempt pellagra given chronic tryptophan depletion from serotonin overproduction. Omeprazole substituted with H2-blocker on planned CgA monitoring dates to preserve assay accuracy. Surveillance plan: multiphase CT abdomen and **24-hour urine 5-HIAA every 3 months on active SSA therapy**; TTE annually given confirmed carcinoid syndrome. Advance care planning conversation initiated given progressive metastatic disease trajectory; surrogate documented"

Clinical Documentation Elements

- **Document the specific site** (**C7A.012** ileum, **C7A.020** appendix, **C7A.090** lung/bronchus, etc.) when pathology and imaging support it — **avoid C7A.00** (unspecified) when the site is known
- **Document grade per WHO 2019** — G1, G2, G3 (NET), or NEC — with Ki-67 percentage and mitotic rate. Grade drives treatment selection
- **Document carcinoid syndrome (E34.0)** — and specifically **E34.01** if cardiac involvement is present — as an additional code alongside the tumor code, never as the sole code
- **Code every metastatic site separately: C7B.02** (liver), **C7B.01** (distant lymph nodes), **C7B.03** (bone), **C7B.04** (peritoneum) — each documented site supports clinical complexity
- **Document active vs. history status at every encounter:** Active C7A while on treatment or active surveillance under a treatment plan; transition to **Z85.030** only after definitive treatment, no further treatment directed at the site, and no evidence of disease
- **Document SSA regimen** (octreotide LAR dose and interval; lanreotide dose and interval), telotristat ethyl when added for refractory diarrhea, dose-titration rationale, and tolerability findings
- **Document carcinoid heart disease screening** — TTE date, NT-proBNP value, urinary 5-HIAA trend; flag **E34.01** if cardiac involvement confirmed
- **Document niacin (B3) supplementation and serum tryptophan/niacin status;** engage caregiver for adherence in elderly
- **Document carcinoid-crisis precautions** — chart flag for diagnosis, SC octreotide rescue prescription, communication to anesthesia/pulmonology/emergency providers about contraindicated agents (epinephrine, beta-agonists)
- **Document CGA findings** (function, cognition, comorbidity, polypharmacy, nutrition, social support) when treatment intensification is being considered
- **Document advance care planning** — ACP within 8 weeks of diagnosis for G3/NEC or progressive metastatic disease; annual or at major changes for stable disease

Reframing Common Documentation Shortcuts

INSTEAD OF...	DOCUMENT...
'Carcinoid tumor'	'Malignant carcinoid tumor of the ileum **(C7A.012)**, WHO 2019 grade 1, with hepatic metastases **(C7B.02)** and carcinoid syndrome **(E34.0)**; on octreotide LAR 30 mg IM q4 weeks per NCCN v1.2026'

INSTEAD OF...	DOCUMENT...
'Possible NET'	'Suspected neuroendocrine tumor — biopsy pending; coding presenting symptoms (**R19.7** diarrhea, **R23.2** flushing, **R06.2** wheezing) until pathology confirms malignancy and assigns **C7A** code'
'Carcinoid syndrome'	'Carcinoid syndrome **(E34.0)** — additional code alongside **C7A.012** ileal primary with **C7B.02** liver metastases; not a sole code'
'History of carcinoid'	**Use Z85.030 ONLY after definitive treatment with no active disease and no ongoing surveillance for known disease**; AS patients retain **C7A**
'On octreotide'	'Octreotide LAR 30 mg IM every 4 weeks; short-acting octreotide 100 µg SC TID prn bridge; targeting symptom optimization not biochemical normalization per Crook 2022'
'On telotristat'	'Telotristat ethyl 250 mg PO TID added for refractory diarrhea persisting despite SSA optimization per TELESTAR; baseline LFTs and periodic monitoring documented'
'Frail elderly patient'	'G-8 score 12 → comprehensive geriatric assessment completed; CIRS comorbidity burden moderate; functional status ECOG 1; SSA dose calibrated to symptom optimization, not biochemical normalization'
'Carcinoid heart disease'	'Carcinoid heart syndrome **(E34.01)** — confirmed on TTE with severe tricuspid regurgitation and pulmonary stenosis; NT-proBNP 980 pg/mL; cardio-oncology engaged'
'Liver metastases'	'Secondary carcinoid of liver **(C7B.02)** — three hypervascular lesions on multiphase CT; SSTR-positive on Ga-DOTATATE PET; primary ileal **C7A.012** documented'
'Diarrhea worsening'	'Watery diarrhea 6–8 stools/day worsening near end of 4-week SSA cycle; shortened octreotide LAR interval to q3 weeks per NCCN v1.2026; renal function and electrolytes monitored'

ABBREVIATIONS: AS = active surveillance; CGA = Comprehensive Geriatric Assessment; CIRS = Cumulative Illness Rating Scale; CT = computed tomography; ECOG = Eastern Cooperative Oncology Group; G-8 = Geriatric 8 screening; IM = intramuscular; LAR = long-acting release; LFT = liver function test; NCCN = National Comprehensive Cancer Network; NET = neuroendocrine tumor; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PET = positron emission tomography; PO = per os (orally); SC = subcutaneous; SSA = somatostatin analog; SSTR = somatostatin receptor; TID = three times daily; TTE = transthoracic echocardiogram; WHO = World Health Organization
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

4 TREATMENT AND REFERRAL QUICK GUIDE

Carcinoid tumor treatment is directed by multidisciplinary NET centers.

Treatment intensity is calibrated to functional status using a CGA-based assessment and to **histologic grade/tumor burden**, not biochemical normalization alone. **NCCN does not prescribe a fixed treatment sequence.** Decisions are **individualized** based on grade, somatostatin receptor status, tumor burden, symptom burden, and rate of progression.

CLINICAL PEARL

The primary care role is to **recognize the syndrome, confirm with biomarkers and imaging, and refer promptly**. Ongoing PCP responsibilities include longitudinal symptom management coordinated with endocrinology and oncology, annual cardiac surveillance with TTE and NT-proBNP, carcinoid crisis prevention, niacin supplementation, nutritional support, and polypharmacy review. Accurate active-versus-history coding is maintained throughout.

Therapy Escalation Criteria

TRIGGER	ACTION
Two or more of: episodic flushing, watery diarrhea, wheezing[10]	**Order 24-hour urine 5-HIAA + serum CgA**; refer GI/oncology within 1–2 weeks
Elevated 5-HIAA or CgA, or NET on imaging[10]	**Refer to multidisciplinary NET center**; full staging (multiphase CT + ^{68}Ga-DOTATATE PET) and biopsy for grading
Confirmed C7A diagnosis	**Initiate SSA if symptomatic or SSTR-positive**: octreotide LAR 20–30 mg IM q4 weeks (PROMID HR 0.34)[39] or lanreotide 120 mg SC q4 weeks (CLARINET HR 0.47)[40]
SSA bridging needed for acute symptom control[10]	**Short-acting octreotide 100–250 µg** SC TID during the 10–14-day LAR-onset window
Symptoms worsen near end of 4-week SSA cycle[10]	**Shorten octreotide LAR injection interval to q3 weeks**; above-label dosing (octreotide LAR up to 60 mg/month, lanreotide up to 120 mg q14 days per CLARINET FORTE)[41]
Refractory diarrhea despite SSA optimization[42]	Add **telotristat ethyl 250 mg PO TID** (TELESTAR); symptomatic benefit may be delayed several weeks

TRIGGER	ACTION
Rule out non-carcinoid diarrhea before escalation[10]	**Assess pancreatic exocrine insufficiency** (fecal elastase), bile acid malabsorption (SeHCAT or cholestyramine trial), short-gut syndrome before further escalation
Carcinoid syndrome confirmed[11]	Baseline TTE and NT-proBNP; q1–3 years TTE without CHD; ≥annually with CHD; **NT-proBNP >260 pg/mL** or **5-HIAA ≥300 µmol/24h → urgent TTE**
Disease progression on optimized SSA[10]	**Multidisciplinary review for next-line therapy**: 177Lu-Dotatate PRRT, everolimus, cabozantinib, capecitabine/temozolomide; pancreatic NETs may use sunitinib
SSTR-positive midgut NET with progression on SSA	**Refer for 177Lu-Dotatate (Lutathera)**[37] 7.4 GBq IV q8 weeks × 4 cycles (NETTER-1 PFS HR 0.18); **GFR ≥30 required**[43]
Patient on long-term PPI needing CgA assessment[27]	**Hold PPI ≥1–2 weeks when clinically safe before CgA measurement**; substitute H2-blocker if continued acid suppression needed
Elective surgery, endoscopy, or anesthesia in carcinoid patient[11]	**Pre-procedure prophylactic SSA per neuro-endocrine protocol**; ensure carcinoid diagnosis flagged in chart so anesthesia avoids epinephrine and beta-agonists
G3 NEC or rapidly progressive disease[10]	**Multidisciplinary review**; platinum-based chemotherapy similar to small-cell lung cancer rather than SSA-based pathway; early palliative care integration
CGA-defined frailty or G-8 ≤14[36]	**Reduced-intensity treatment plans**; SSA dose calibrated to symptom optimization, not biochemical normalization

ABBREVIATIONS: CGA = Comprehensive Geriatric Assessment; CgA = chromogranin A; CHD = carcinoid heart disease; G-8 = Geriatric 8 screening; GFR = glomerular filtration rate; 5-HIAA = 5-hydroxyindoleacetic acid; IM = intramuscular; IV = intravenous; LAR = long-acting release; LN = lymph node; NCCN = National Comprehensive Cancer Network; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PFS = progression-free survival; PO = per os; PPI = proton pump inhibitor; PRRT = peptide receptor radionuclide therapy; SC = subcutaneous; SeHCAT = selenium-75 homotaurocholic acid test; SSA = somatostatin analog; SSTR = somatostatin receptor; TID = three times daily; TTE = transthoracic echocardiogram
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NCCN v1.2026-Aligned Treatment by Subtype, Grade, and Patient Profile

SUBTYPE/SETTING	PREFERRED REGIMEN(S)	KEY NOTES
Asymptomatic, low-burden G1 NET[10]	Observation with imaging every **3–12 months** per NCCN v1.2026	VBC-aligned first step for asymptomatic, low-tumor-burden G1 disease in elderly
Symptomatic or SSTR-positive G1/G2 NET — first-line[10]	Octreotide **LAR 20–30 mg IM q4 weeks** (PROMID HR 0.34)[39] OR **lanreotide 120 mg SC q4 weeks** (CLARINET HR 0.47)[40]	Therapeutic LAR levels in 10–14 days; bridge with s**hort-acting octreotide 100–250 µg** SC TID for **acute symptom control**
Worsening symptoms near end of 4-week cycle[10]	**Shorten injection interval to q3 weeks**; above-label dosing per CLARINET FORTE	Titrate to symptom optimization, not biochemical normalization
Refractory carcinoid-syndrome diarrhea despite SSA optimization[10]	**Telotristat ethyl 250 mg PO TID** (TELESTAR)[42]	**Symptomatic benefit may be delayed several weeks**; monitor LFTs at baseline and periodically given hepatotoxicity risk
Progressive disease on optimized SSA (G1/G2)[10]	**Everolimus 10 mg PO daily** (category 1 nonfunctional NETs; RADIANT-3 and RADIANT-4)[44,45]	**Monitor for stomatitis, hyperglycemia (challenging in diabetics), immunosuppression, and pneumonitis**
Progressive SSTR-positive midgut NET[10]	**177Lu-Dotatate 7.4 GBq IV every 8 weeks × 4 cycles** (NETTER-1 PFS HR 0.18)[43]	**GFR ≥30 required**; baseline and serial renal function and CBC monitoring; myelodysplastic syndrome in ~2% of NETTER-1 patients
First-line PRRT (selected SSTR-positive)[10]	Now an option for SSTR-positive tumors with **Ki-67 ≥10%** and clinically significant tumor burden per NCCN v1.2026	Specialist-directed; eligibility requires confirmed SSTR positivity and adequate renal function
Second-line after everolimus or PRRT[10]	Cabozantinib (category 1 in pretreated patients)	Hypertension, diarrhea, hand-foot syndrome — significant QoL impact in elderly; **monitor closely**

SUBTYPE/SETTING	PREFERRED REGIMEN(S)	KEY NOTES
Pancreatic NET — additional first-line option[10]	**Sunitinib 37.5 mg PO daily**	Hypertension, diarrhea, hand-foot syndrome; greater cytotoxic activity reported with capecitabine/ temozolomide in pancreatic NETs
Cytoreduction needed (tumor response priority)[10]	Capecitabine + temozolomide	**Greater activity reported in pancreatic NETs**; consider in patients needing rapid burden reduction
Liver-predominant metastatic disease[10]	Hepatic arterial embolization (TAE/ TACE/TARE)	**Highly effective for symptom palliation**; multidisciplinary review at NET center
Low-burden primary midgut NET with risk of obstruction[10]	Primary tumor + regional lymph node resection	**Reduces risk of bowel obstruction, mesenteric ischemia, bleeding, perforation**
G3/NEC, any stage[10]	Platinum-based chemotherapy (etoposide + cisplatin/carboplatin) similar to small-cell lung cancer; early palliative care	**Median OS ~10 months metastatic**; warrants early ACP and goals-of-care discussion
Stable disease on SSA ≥2 years[10]	Consider extending imaging intervals to **every 6–12 months**	**Nonfunctional tumors with clinically significant progression**: **discontinue SSA** (continue only for functional tumors)
Frail patient with limited life expectancy from non-NET comorbidities[36]	Prioritize symptom management and QoL over tumor-directed therapy; integrate palliative care early	Reduced-intensity SSA dosing; advance care planning per ASCO 2024

ABBREVIATIONS: ACP = advance care planning; ASCO = American Society of Clinical Oncology; CBC = complete blood count; GFR = glomerular filtration rate; HR = hazard ratio; IM = intramuscular; IV = intravenous; LAR = long-acting release; LFT = liver function test; LN = lymph node; NCCN = National Comprehensive Cancer Network; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; OS = overall survival; PFS = progression-free survival; PO = per os; PNET = pancreatic neuroendocrine tumor; PRRT = peptide receptor radionuclide therapy; QoL = quality of life; SC = subcutaneous; SSA = somatostatin analog; SSTR = somatostatin receptor; TACE = transarterial chemoembolization; TAE = transarterial embolization; TARE = transarterial radioembolization; TID = three times daily; VBC = value-based care

***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

CLINICAL PEARL

Functional status, not chronologic age, guides treatment intensity. A fit 80-year-old may tolerate SSA therapy and PRRT as well as a younger patient; a frail 65-year-old may not. Use validated tools such as the **G-8 Questionnaire, CGA, or Fried Frailty Criteria** to assess capacity before escalating therapy.

In frail older adults, **titrate SSAs to symptom control, not biochemical normalization.** Pursuing normal biomarker levels in this population risks ileus, gastroparesis, AKI, and preventable hospitalizations.

Observation is a valid first step for asymptomatic, low-tumor-burden G1 disease. Systemic therapy is not appropriate for every patient.

Non-Pharmacologic Care and Supportive Interventions

INTERVENTION	TARGET/RECOMMENDATION*	NOTES
Symptom diary[10]	Record frequency, severity, and triggers of flushing, diarrhea, wheezing, and cardiac symptoms	**Bring to every visit**; supports SSA titration decisions; trigger avoidance does not replace medical management
Dietary trigger avoidance[10]	Avoid alcohol, aged cheeses, fermented foods; avoid avocados, bananas, pineapples, eggplant, tomatoes, walnuts **48 hours before 5-HIAA collection**	**Helpful but does NOT replace SSA therapy**; reinforce with each prescription cycle
Niacin (vitamin B3) supplementation[10]	Routine in all carcinoid syndrome patients, particularly elderly, to preempt pellagra	**Monitor serum tryptophan and niacin** at baseline and during SSA therapy
Small frequent meals[46]	**Reduce GI hormonal stimulation**; practical for symptom management	Coordinate with nutrition referral when significant **weight loss** is present
Hydration plan[47]	**≥2 L/day when active diarrhea burden**; document hydration plan in chart	Reduce or pause fluid-restriction medications if present; **monitor electrolytes**
Carcinoid-crisis precaution flag[11]	Prominent EHR alert; SC octreotide 100–500 µg rescue prescription; caregiver and patient trained on use	Communicate diagnosis to anesthesia, pulmonology, emergency, and cardiology providers **before procedure**

INTERVENTION	TARGET/RECOMMENDATION*	NOTES
TTE surveillance for CHD[10]	Baseline at carcinoid syndrome diagnosis; q1–3 years without CHD; ≥annually with CHD	NT-proBNP >260 pg/mL or 5-HIAA ≥300 µmol/24h → urgent TTE11
Cholecystogram or RUQ ultrasound[10]	**q1–2 years** with long-term SSA use	**Cholelithiasis is common**; symptomatic stones may need GI/surgery referral
Caregiver engagement[36]	**At every encounter**; particularly important when pellagra-related cognitive impairment present	**Self-injection programs** (lanreotide) may improve adherence; home nursing for IM octreotide
Advance care planning[47]	**Within 8 weeks of diagnosis** for G3/NEC or progressive metastatic disease; annual + at major changes for stable disease	**Document surrogate, code status, and goals of care**; ACP 99497/99498 during AWV
Palliative care integration[47]	Concurrent with active treatment in symptomatic or progressive disease, not end-of-life only	**Reduces hospitalizations and ED visits**; improves QoL

ABBREVIATIONS: ACP = advance care planning; AWV = Annual Wellness Visit; CHD = carcinoid heart disease; ED = emergency department; EHR = electronic health record; GI = gastrointestinal; 5-HIAA = 5-hydroxyindoleacetic acid; IM = intramuscular; NT-proBNP = N-terminal pro–B-type natriuretic peptide; QoL = quality of life; RUQ = right upper quadrant; SC = subcutaneous; SSA = somatostatin analog; TTE = transthoracic echocardiogram
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Medication Safety and Key Interactions

DRUG/CLASS*	INTERACTION/TOXICITY	CLINICAL ACTION
Catecholamines/beta-agonists (epinephrine, albuterol) in carcinoid crisis[29]	**CONTRAINDICATED** — precipitate or worsen crisis by triggering massive hormone release	Flag diagnosis prominently; **preferred vasopressors are vasopressin and phenylephrine**; rescue SC octreotide 100–500 µg
PPIs + CgA measurement[23]	**PPIs cause false elevation of CgA**; risks unwarranted escalation	**Hold PPI ≥1–2 weeks before CgA** measurement when clinically safe; substitute H2-blocker if needed
Loperamide (over-aggressive antidiarrheal use)[10]	Constipation, paralytic ileus, AKI, dehydration in **frail elderly**	**Use cautiously and only when carcinoid-specific diarrhea confirmed**; optimize SSA dose first; monitor renal function and bowel function

DRUG/CLASS*	INTERACTION/TOXICITY	CLINICAL ACTION
Telotristat ethyl (long-term)[48]	**Hepatotoxicity** (LFT elevation), constipation, abdominal pain; possible CYP-mediated DDIs	Baseline LFTs; monitor periodically during therapy; monitor for constipation and ileus
SSAs + antidiabetic agents (insulin, sulfonylureas)[10]	**SSAs cause both hyperglycemia and hypoglycemia** depending on baseline pancreatic function; unpredictable in elderly	**Glucose at SSA initiation**; quarterly during stable therapy; adjust antidiabetic regimen as needed
SSAs and cholelithiasis[49]	Cholelithiasis common with long-term octreotide/lanreotide	**Hepatobiliary imaging q1–2 years**; symptomatic stones may need GI/surgery referral
Everolimus (mTOR inhibitor)[10]	Stomatitis, hyperglycemia (worsens diabetes), immunosuppression, pneumonitis, hyperlipidemia	**Oral care**; glucose monitoring; vigilance for new dyspnea; lipid monitoring; periodic dose review
PRRT (177Lu-Dotatate) + nephrotoxic agents (NSAIDs, contrast, aminoglycosides)[43]	Compounded renal injury; required **GFR ≥30 for PRRT**; myelodysplastic syndrome in ~2% of NETTER-1 patients	**Baseline and serial renal function and CBC**; minimize nephrotoxic co-administration; long-term hematologic monitoring
Cabozantinib[50]	Hypertension, diarrhea, hand-foot syndrome, fatigue	**Daily home BP monitoring**; antidiarrheal support; dermatologic management; dose modification per oncology protocol
Sunitinib (pancreatic NET)[51]	Hypertension, diarrhea, hand-foot syndrome, hypothyroidism	**BP monitoring**; TSH q3 months; antidiarrheal support; dermatologic care
Capecitabine + temozolomide[10]	Myelosuppression, mucositis, hand-foot syndrome, nausea	**CBC monitoring**; oral care; dose modification per protocol
Anticoagulation in carcinoid patient with AF or VTE[52]	CHD with right-sided valve disease and electrolyte derangements increase arrhythmia risk	ECG and electrolytes; CHA_2DS_2-VASc-based anticoagulation; cardio-oncology coordination if CHD

ABBREVIATIONS: AF = atrial fibrillation; AKI = acute kidney injury; BP = blood pressure; CBC = complete blood count; CgA = chromogranin A; CHA_2DS_2-VASc = Congestive heart failure, Hypertension, Age, Diabetes, Stroke, Vascular disease, Sex; CHD = carcinoid heart disease; CYP = cytochrome P450; DDI = drug-drug interaction; ECG = electrocardiogram; GFR = glomerular filtration rate; H2 = histamine-2 receptor; LFT = liver function test; mTOR = mammalian target of rapamycin; NET = neuroendocrine tumor; NSAID = nonsteroidal anti-inflammatory drug; PPI = proton pump inhibitor; PRRT = peptide receptor radionuclide therapy; SC = subcutaneous; SSA = somatostatin analog; TSH = thyroid-stimulating hormone; VTE = venous thromboembolism

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When to Refer

CRITERION	SPECIALIST	URGENCY
Carcinoid crisis (hypotension + flushing + tachycardia + bronchospasm)[10]	Emergency department/ anesthesia/oncology on call	**Emergent**
Acute bowel obstruction or severe abdominal pain in known NET[10]	Emergency department/GI surgery	**Emergent**
Acute decompensated right-sided heart failure in CHD[11]	Emergency department/cardio-oncology	**Emergent**
Two or more carcinoid syndrome symptoms with positive biomarkers[10]	Multidisciplinary NET center/GI/ oncology	**Urgent** (within 1–2 weeks)
NET on imaging or biopsy[10]	Multidisciplinary NET center	**Urgent** (within 1 week)
Progressive disease on optimized SSA[10]	Medical oncology for PRRT/ everolimus/cabozantinib[1,17,25]	**Urgent** (within 2–4 weeks)
G3 NEC or rapidly progressive disease[10]	Medical oncology + palliative care	**Urgent** (within 1 week)
NT-proBNP >260 pg/mL or 5-HIAA ≥300 µmol/24h[11]	Cardiology/cardio-oncology for TTE[19]	**Urgent** (within 1–2 weeks)
Liver-predominant disease for embolization[10]	Interventional radiology + medical oncology	Routine (within 2–4 weeks)
MEN1 or family history of multiple endocrine tumors[10]	Genetics + endocrinology	Routine
G-8 ≤14 or established frailty before treatment intensification[12,36]	Geriatric oncology/CGA	**Urgent** (before next treatment cycle)
Elective surgery/endoscopy/ anesthesia in carcinoid patient[10]	Anesthesia coordination + prophylactic SSA per protocol	Routine (before procedure)
Frail patient with limited life expectancy[53]	Palliative care + advance care planning	Routine (at diagnosis)
Caregiver burden or SDOH concern[12]	Social work	Routine (at diagnosis)

CRITERION	SPECIALIST	URGENCY

ABBREVIATIONS: CGA = Comprehensive Geriatric Assessment; CHD = carcinoid heart disease; G-8 = Geriatric 8 screening; GI = gastrointestinal; 5-HIAA = 5-hydroxyindoleacetic acid; MEN1 = multiple endocrine neoplasia type 1; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PRRT = peptide receptor radionuclide therapy; SDOH = social determinants of health; SSA = somatostatin analog; TTE = transthoracic echocardiogram

Follow-Up Timing

STAGE/CATEGORY	FREQUENCY	LABS/MONITORING
Active surveillance, asymptomatic G1 NET[10]	**Imaging q3–12 months**; 5-HIAA and CgA per NCCN	**Symptom check at every visit**; trigger for treatment initiation: new symptoms or progression
Taking SSA (octreotide LAR or lanreotide), stable disease[10]	Symptom check **q1–3 months**; biomarkers **q3 months**; CT or MRI **q3–6 months**; can extend to **q6–12 months after 2 years stable**[14]	Biomarkers + imaging; renal function and electrolytes during chronic diarrhea; glucose; LFTs
Taking SSA, progressive disease workup[10]	**Visit q4 weeks**; biomarkers and imaging on accelerated cadence	**Reassess** for telotristat addition, PRRT eligibility, everolimus, cabozantinib, capecitabine/temozolomide
TTE for carcinoid heart disease[11]	Baseline at syndrome diagnosis; **q1–3 years without CHD; ≥annually with CHD**; urgent if NT-proBNP >260 pg/mL or 5-HIAA ≥300 µmol/24h	Document tricuspid regurgitation grade, pulmonary stenosis, RV function
Niacin status and pellagra screening[30]	Serum tryptophan and niacin at baseline; **reassess during SSA therapy**	**Initiate B3 supplementation routinely**; reinforce in elderly and frail patients
Taking PRRT[43]	**CBC before each cycle**; renal function before each cycle; monthly during active therapy	**GFR must remain ≥30**; long-term myelodysplastic syndrome surveillance
Taking everolimus[54]	**Monthly** visit during active therapy; CBC, CMP, glucose, lipid pane	Vigilance for stomatitis, hyperglycemia, immunosuppression, pneumonitis

STAGE/CATEGORY	FREQUENCY	LABS/MONITORING
Taking telotristat ethyl[48]	Baseline LFTs; periodic LFT monitoring; symptom check q1–3 months	Monitor for constipation and ileus; hepatotoxicity risk with long-term use
Cholecystogram/hepatobiliary imaging[49]	**Every 1–2 years** with long-term SSA use	Cholelithiasis common; symptomatic stones may require GI/surgery referral
Carcinoid-crisis preparedness[10]	**At every visit** confirm SC octreotide rescue prescription, chart flag, caregiver training	**Update during major care transitions and before procedures requiring anesthesia**
Advance care planning[53]	**Within 8 weeks for G3/NEC**; annual + at major changes for stable disease	Document surrogate, code status, goals; ACP 99497/99498 during AWV

ABBREVIATIONS: ACP = advance care planning; AWV = Annual Wellness Visit; CBC = complete blood count; CgA = chromogranin A; CHD = carcinoid heart disease; CMP = comprehensive metabolic panel; CT = computed tomography; GFR = glomerular filtration rate; 5-HIAA = 5-hydroxyindoleacetic acid; LAR = long-acting release; LFT = liver function test; MRI = magnetic resonance imaging; NCCN = National Comprehensive Cancer Network; NEC = neuroendocrine carcinoma; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PRRT = peptide receptor radionuclide therapy; RV = right ventricular; SC = subcutaneous; SSA = somatostatin analog; TTE = transthoracic echocardiogram
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Comorbidity Management — Primary Care Role

COMORBIDITY	APPROACH	CAUTION
Carcinoid heart disease (E34.01)[11]	Baseline TTE; **q1–3 years** without CHD; ≥annually with CHD; manage volume status; cardio-oncology consultation for valve disease	**Symptoms overlap with CHF and COPD** — do not attribute dyspnea/edema to age-related decline without TTE
Hypertension (I10)[10]	Optimize antihypertensive regimen; daily home BP monitoring during cabozantinib or sunitinib	**Bevacizumab is not standard for carcinoid**; TKIs (cabozantinib, sunitinib) cause hypertension
Diabetes mellitus (E11.9)[10]	Glucose monitoring at SSA initiation; q3 months HbA1c; adjust antidiabetic agents proactively	**SSAs cause both hyperglycemia and hypoglycemia**; everolimus causes hyperglycemia

COMORBIDITY	APPROACH	CAUTION
Cholelithiasis (K80.20)[49]	Hepatobiliary imaging **q1–2** years on long-term SSA; symptomatic stones → GI/surgery referral	**Common with sustained octreotide/lanreotide**; do not assume incidental finding requires surgery in asymptomatic patients
Bowel obstruction/mesenteric fibrosis (K56.699)[10]	Imaging surveillance per NCCN; surgical evaluation in low-burden primary disease	**Acute obstruction requires emergency evaluation**
Pellagra/niacin deficiency (E52)[30]	Serum tryptophan and niacin at baseline; routine B3 supplementation; caregiver-supported adherence	Cognitive impairment may reflect pellagra rather than dementia in carcinoid syndrome patients
Atrial fibrillation (I48.91)[11]	Baseline ECG; rate or rhythm control per cardiology; anticoagulation per CHA_2DS_2-VASc	Electrolyte derangements from chronic diarrhea may trigger arrhythmias
Renal impairment (N18.1/9)[10,37]	Baseline eGFR; before PRRT and during active diarrhea; minimize nephrotoxic co-administration	**PRRT requires GFR ≥30**
Depression/anxiety (F32.A/ F41.9)[55]	PHQ-9/GAD-7 baseline and during treatment; SSRIs generally safe	**Vasoactive hormones may mimic primary anxiety** — consider syndrome workup before psychiatric attribution
Frailty (R54)[12]	G-8 screening; CGA when indicated; SSA titration calibrated to symptom optimization	**Avoid over-aggressive antidiarrheals and biochemical-normalization targets in frail patients**

ABBREVIATIONS: BP = blood pressure; CGA = Comprehensive Geriatric Assessment; CHA_2DS_2-VASc = Congestive heart failure, Hypertension, Age, Diabetes, Stroke, Vascular disease, Sex; CHF = congestive heart failure; CHD = carcinoid heart disease; COPD = chronic obstructive pulmonary disease; ECG = electrocardiogram; eGFR = estimated glomerular filtration rate; GAD-7 = Generalized Anxiety Disorder-7; GI = gastrointestinal; G-8 = Geriatric 8 screening; HbA1c = glycated hemoglobin; NCCN = National Comprehensive Cancer Network; PHQ-9 = Patient Health Questionnaire-9; PRRT = peptide receptor radionuclide therapy; SSA = somatostatin analog; SSRI = selective serotonin reuptake inhibitor; TKI = tyrosine kinase inhibitor
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

AAVBC PERSPECTIVE

*AAVBC encourages primary care providers to move from **symptom-masking to syndrome-screening** when patients present with any combination of episodic unprovoked flushing, unexplained chronic diarrhea, or unexplained wheezing. These symptoms are frequently*

*managed as standalone conditions (rosacea, IBS, or adult-onset asthma), when their **co-occurrence should prompt immediate evaluation for carcinoid syndrome**. The appropriate primary care response is to order a **24-hour urine 5-HIAA and serum Chromogranin A**.*

*AAVBC also supports recognition that **carcinoid syndrome does not require liver metastases** to manifest. Primary pulmonary and ovarian carcinoids can secrete hormones directly into systemic circulation, and that the classic triad of flushing, diarrhea, and wheezing is frequently absent or atypical at presentation. **Niacin depletion** from tryptophan diversion toward serotonin synthesis is an underrecognized complication that can present as dermatitis, diarrhea, and cognitive impairment in elderly patients, and is **frequently misattributed to aging or unrelated comorbidities**. **Niacin monitoring should be a priority** action at every encounter. This helps ensure the right patients are identified and referred for the right evaluation at the right time.*

Cost-Smart Options

BRAND*	GENERIC/ALTERNATIVE	EST. SAVINGS	COST-SMART TIP
Sandostatin LAR (octreotide) Brand 30 mg IM q4wk: ~$6,800–$9,300/ injection per month[56]	Generic octreotide LAR 30 mg: **~$5,500/injection** per month; specialty pharmacy stocking limited — confirm availability before prescribing[57]	**$1,300–$3,800/injection** per month when generic available at specialty pharmacy	Use generic when available; manufacturer assistance for eligible patients
Somatuline (lanreotide) Brand 120 mg SC q4wk: from ~$10,275/injection per month[58]	Generic lanreotide 120 mg: from **~$2,010–$2,652/ injection** per month[59]	**~$7,600–$8,265/ injection** per month at retail prices	Manufacturer assistance program; self-injection programs reduce clinic-visit burden
Xermelo (telotristat) Brand 250 mg TID: ~$11,864/month (84-tablet/28-day supply)[60]	**No generic available**	Manufacturer assistance variable	Confirm payer prior authorization; coordinate dispensing with specialty pharmacy
Afinitor (everolimus) Brand 10 mg daily: ~$19,260/month (28 tablets)[61]	Generic everolimus 10 mg: from **~$11,882/month**[62]	~$7,400/month brand vs. generic retail	Use generic; monitor for stomatitis and hyperglycemia

BRAND*	GENERIC/ALTERNATIVE	EST. SAVINGS	COST-SMART TIP
Cabometyx (cabozantinib) Brand 20–60 mg daily: from ~$26,470–$28,700 / month (30 tablets)[63]	**No generic at present**	Manufacturer assistance variable	Coordinate prior authorization; dose modifications often required
Lutathera (177Lu-DOTATATE, PRRT) ~$65,497/dose ull course (4 doses q8wk): ~$261,990 in drug cost alone (facility fees additional)[64]	**No alternative**; specialty infusion	Negotiated MA-network pricing	Confirm SSTR positivity, GFR ≥30, and prior authorization before referral
Inpatient management of carcinoid crisis Estimated $20,000–$50,000+ per admission (3–5 day ICU episode at national average ICU rates)[65]	Outpatient prophylaxis + caregiver training + SC octreotide rescue: generic SC octreotide vials from **~$26–$35/vial**; rescue kit (3–5 vials): **~$80–$150**; ongoing SSA cost as above[65,66]	Preventing one crisis episode may avoid **~$20,000–$50,000**+ in facility charges	Crisis prevention is the dominant VBC lever (PCR Star measure)
Surveillance imaging at high-cost center Multiphase CT (list/ non-network): ~$3,000–$7,000 per study; 4 studies/year = ~$12,000–$28,000 annually[67]	MA-network negotiated multiphase CT: approximately **$200–$600** per study; 4 studies/year = **~$800–$2,400** annually[67]	**~$2,400–$6,400** per imaging event; **~$9,600–$25,600** annually across a 4-study surveillance schedule	Coordinate within MA network; confirm prior authorization

ABBREVIATIONS: GFR = glomerular filtration rate; LAR = long-acting release; MA = Medicare Advantage; NCCN = National Comprehensive Cancer Network; OTC = over-the-counter; PCR = Plan All-Cause Readmissions; PRRT = peptide receptor radionuclide therapy; SC = subcutaneous; SSTR = somatostatin receptor; VBC = value-based care
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Patient Education and Adherence

Cover the following topics at education visits and document topics addressed, patient understanding (teach-back when appropriate), and caregiver involvement.

- **Diagnosis in plain language** — primary site, histologic grade, and whether carcinoid syndrome is present

- **Treatment plan** — whether the current approach is observation, SSA initiation, or systemic escalation, and why
- **SSA injection logistics** — clinic visit schedule, home nursing, or self-injection (lanreotide); confirm the patient and caregiver understand the administration method
- **Symptom diary and dietary triggers** — how to track flushing and diarrhea episodes; common dietary and physical triggers to avoid (alcohol, aged cheeses, physical exertion, stress)
- **Carcinoid crisis warning signs** — sudden severe flushing, hypotension, wheezing, or confusion; when to call 911; how and when to use the **SC octreotide rescue prescription**
- **Niacin supplementation** — why it is prescribed (tryptophan depletion from excess serotonin production) and how to take it
- **Between-visit reporting** — instruct the patient to contact the office promptly for new or worsening flushing, severe diarrhea, dyspnea, edema, abdominal pain, or fever
- **Advance care planning** — introduce early in progressive disease; document surrogate, code status, and goals of care

DOCUMENTATION REMINDER — PATIENT EDUCATION

Document the **topics covered** (diagnosis and grade, treatment plan, SSA injection logistics, trigger avoidance, crisis precautions including SC octreotide rescue, niacin supplementation, TTE surveillance plan, advance care planning), **the patient's understanding** (teach-back when capacity permits), and **the caregiver's involvement**.
Education documentation supports continuity, shared decision-making, and signals to oncology, GI, and cardiology specialists that the plan is patient-aligned.

Quality Metrics Tie-In

MEASURE[69,70]	STANDARD	NOTES
Plan All-Cause Readmissions (CMS Stars/HEDIS PCR; MIPS #479)	**Denominator:** enrollees ≥18 with acute inpatient discharge**:** unplanned readmission within 30 days (lower is better) **Exclusions:** planned readmissions (scheduled chemotherapy, transplant), hospice, psychiatric	MIPS #479 is a group-level measure calculated by CMS; individual PCPs cannot submit it directly. Carcinoid crisis, refractory diarrhea, and dehydration are primary avoidable readmission drivers; **7-day post-discharge follow-up is the highest-yield intervention**

MEASURE[69,70]	STANDARD	NOTES
Medication Reconciliation Post-Discharge (HEDIS TRC sub-measure 4; MIPS #46)	**Denominator:** patients ≥18 seen in outpatient setting within 30 days of inpatient discharge **Numerator:** discharge medication list reconciled with current medication list documented in outpatient record **Exclusions:** none specified	SSAs, telotristat, antidiarrheals, cardiac medications, and everolimus each carry interaction potential; polypharmacy reconciliation is essential after any carcinoid-related hospitalization. Submit CPT II code 1111F at the post-discharge visit
Care for Older Adults (HEDIS COA)	**Denominator:** SNP enrollees ≥66, continuously enrolled**:** sub-measures documented annually: functional status, medication review **Exclusions:** hospice, ESRD	**No single MIPS number maps to COA as a whole**; MIPS #130 (medication documentation), #131 (pain assessment), and #47 (ACP) each correspond to individual sub-measures. Medication review should address SSA dysglycemia, everolimus hyperglycemia, and electrolyte losses from chronic diarrhea
Colorectal Cancer Screening (HEDIS COL-E; MIPS #113)	**Denominator:** patients 45-75**:** any qualifying screening: FOBT (during MP), FIT-DNA (MP or 2 years prior), flexible sigmoidoscopy or CT colonography (MP or 4 years prior), colonoscopy (MP or 9 years prior) **Exclusions:** history of CRC, total colectomy, hospice, palliative care, death	Rectal and colonic NETs are commonly identified incidentally during HEDIS-mandated colonoscopy; document the NET finding to trigger appropriate surveillance per NCCN guidelines
Controlling High Blood Pressure (HEDIS CBP; MIPS #236)	**Denominator:** patients 18-85 with hypertension (≥2 outpatient visits with HTN diagnosis during MP or year prior)**:** most recent BP <140/90 mmHg during measurement period **Exclusions:** ESRD, hospice, palliative care, pregnancy	**BP destabilization** occurs with cabozantinib or sunitinib therapy and with chronic diarrhea-related electrolyte depletion; frequent monitoring is warranted during active systemic treatment
Glycemic Status Assessment (HEDIS GSD; MIPS #1)	**Denominator:**patients 18-75 with diabetes (≥2 diagnoses on different service dates in MP or prior year, or diagnosis + dispensed diabetes medication) **Numerator:** most recent HbA1c or GMI during MP (flag >9.0%; inverse measure) **Exclusions:** hospice, palliative care, frailty + advanced illness (≥66)	MIPS #1 is an inverse measure **(higher rate = worse performance)**. SSA-induced insulin suppression causes dysglycemia; everolimus causes grade **≥3 hyperglycemia in 5-7% of pNET patients**; monitor at each relevant encounter and adjust diabetes management accordingly

MEASURE[69,70]	STANDARD	NOTES
Advance Care Planning (MIPS #47)	**Denominator:** patients ≥65 with qualifying encounter during performance period **Numerator:** ACP documented in medical record, surrogate identified, OR documentation that ACP was discussed but patient declined **Exclusions:** hospice; ED encounters	Initiate within **8 weeks** of diagnosis for G3/NEC; review annually and at major disease changes for stable carcinoid. CPT 99497/99498 billable during AWV with no patient copay

ABBREVIATIONS: ACP = advance care planning; AWV = Annual Wellness Visit; BP = blood pressure; CBP = Controlling High Blood Pressure; CDC = Comprehensive Diabetes Care; COA = Care for Older Adults; COL = Colorectal Cancer Screening; CMS = Centers for Medicare & Medicaid Services; CPT = Current Procedural Terminology; HbA1c = glycated hemoglobin; HEDIS = Healthcare Effectiveness Data and Information Set; MIPS = Merit-based Incentive Payment System; MRP = Medication Reconciliation Post-Discharge; NET = neuroendocrine tumor; PCR = Plan All-Cause Readmissions; SSA = somatostatin analog; VBC = value-based care

QUALITY OUTCOME

*When primary care ensures **proper syndrome-screening**, such as by ordering **24-hour urine 5-HIAA and serum CgA when patients present with two or more of episodic flushing, unexplained diarrhea, or unexplained wheezing,** patients are diagnosed earlier, coded accurately, and managed with the full complement of PCP-level interventions the diagnosis requires.*
*Those interventions include **accurate active-versus-history coding, baseline and serial cardiac surveillance, niacin supplementation to prevent pellagra, carcinoid crisis prophylaxis with chart flagging and SC octreotide rescue, and SSA titration calibrated to symptom control rather than biochemical normalization**. Goals-of-care alignment is introduced early in frail or older patients with progressive disease.*

5 CODING REMINDERS AND CASE EXAMPLES

Coding Specificity

ELEMENT	DOCUMENTATION REQUIREMENT
Malignant carcinoid (C7A.0x)	State 'malignant carcinoid tumor' or 'malignant neuroendocrine tumor' explicitly — **not 'carcinoid' or 'NET' alone**, which can lead coders to **D3A**
Specific anatomic site (C7A.012, C7A.020, C7A.090, etc.)	Specify ileum, appendix, lung/bronchus, etc., when pathology and imaging support it; **avoid C7A.00 (unspecified)**

ELEMENT	DOCUMENTATION REQUIREMENT
WHO 2019 grade	**Document G1** (Ki-67 <3%, mitoses <2/10 HPF), **G2** (Ki-67 3–20%, mitoses 2–20/10 HPF), **G3 NET** (well-differentiated, Ki-67 >20%), **or NEC** (poorly differentiated, Ki-67 >20%)
Secondary site coded separately (C7B.0x)	Most commonly **C7B.02** (liver); also **C7B.01** (LN), **C7B.03** (bone), **C7B.04** (peritoneum); MUST accompany the primary **C7A** code
Carcinoid syndrome (E34.0/E34.01/ E34.09)	Always coded alongside the tumor code, never as the sole code; **E34.01** for cardiac involvement
Active vs. history status	Active **C7A** while on treatment or active surveillance under a treatment plan; transition to **Z85.030 ONLY after definitive treatment**, no further treatment directed at the site, and no evidence of disease
Maintenance/surveillance therapy exception	Patients on SSAs for functional disease or active surveillance under an active treatment plan retain the C7A code; document the active modality explicitly
Benign carcinoid (D3A.0x) trap	**D3A** does **NOT** map to any HCC in V28; when pathology confirms malignant behavior, code **C7A**
Avoid C7A.00 (unspecified) when site is known	Document specific site whenever imaging or pathology supports it
Personal history (Z85.030) trap	**Z85.030** does **NOT** map to any HCC; appropriate only after definitive treatment with no active disease
MEAT for the active C7A code	Each encounter must document Monitor, Evaluate, Assess, Treat for the active malignancy; chart updates without an encounter do not satisfy MEAT
Comorbidities — frequently undercoded	Hypertension **(I10)**; type 2 diabetes **(E11.x)**; CHF/HF **(I50.x)** for CHD; depression **(F32.x, F33.x)**; polypharmacy (**Z79.84** long-term medication; specific medication codes)

ABBREVIATIONS: CHD = carcinoid heart disease; CHF = congestive heart failure; HPF = high-power field; HF = heart failure; HCC = Hierarchical Condition Category; LN = lymph node; MEAT = Monitor, Evaluate, Assess, Treat; NEC = neuroendocrine carcinoma; NET = neuroendocrine tumor; SSA = somatostatin analog; WHO = World Health Organization

Annual Clinical Review and Confirmation

- **Annual review:** Active malignant carcinoid tumor must be reassessed at each encounter with MEAT documented; patients on SSA therapy, active surveillance, or under post-treatment surveillance with an active treatment plan retain the C7A code and require ongoing documentation each year

- **Visit modality:** Face-to-face or synchronous audio-video telehealth qualifies when it supports meaningful clinical evaluation; chart updates without an encounter do not satisfy MEAT
- **Clinical context:** Under CMS-HCC V28, active malignant carcinoid **(C7A.0x)** and metastatic carcinoid **(C7B.0x)** map to HCC 22 (Cancer Metastatic to Lung/Liver/Brain; AML Except Promyelocytic; CNA RAF 0.363). Site specificity within C7A does not change HCC or RAF assignment but is clinically meaningful for the record. Confirm payment year and CNA segment against current CMS tables
- **Avoid rollover:** Do not copy forward last year's note without updating treatment status, current SSA regimen and any added agents (telotristat, everolimus, PRRT, cabozantinib), WHO grade if revised, surveillance imaging and biomarker results, TTE findings and CHD status, niacin supplementation status, carcinoid-crisis precaution flag, comorbidity burden, and the active vs. history determination

Good Documentation — EHR Tips

EHR TIP	WHAT TO INCLUDE
Problem list precision	'Malignant carcinoid tumor of the ileum **(C7A.012)**, WHO 2019 grade 1, with hepatic metastases **(C7B.02)** and carcinoid syndrome **(E34.0)**; on octreotide LAR 30 mg IM q4 weeks per NCCN v1.2026' — **not 'carcinoid'**
Smart phrases/dot phrases	**Build macros for SSA Cycle Visit** (drug, dose, interval, tolerability, glucose, electrolytes, renal function); 5-HIAA/CgA Surveillance (values, dietary prep notes, PPI status); TTE Surveillance (date, RV function, valve grade, NT-proBNP)
Carcinoid-crisis precaution flag	**Discrete EHR alert highlighting:** 'Carcinoid syndrome — epinephrine and beta-agonists CONTRAINDICATED; SC octreotide 100–500 µg rescue in chart' — visible to anesthesia, pulmonology, and emergency providers
Lab and imaging linking	Link 5-HIAA, CgA, NT-proBNP, TTE, multiphase CT/MRI, ^{68}Ga-DOTATATE PET, pathology with WHO grade, and serum tryptophan/niacin to the active **C7A** problem
Active vs. surveillance status	**Use a status field at the top of every note** — 'Active SSA — octreotide LAR 30 mg q4 weeks, cycle 9' vs. 'Active surveillance — stable on SSA ≥2 years' vs. 'History — **Z85.030**, no active treatment'
Surveillance flags	**Recurring reminders for SSA cycle visits, biomarker intervals** (q3 months active, q3–12 months stable), imaging cadence (q3–6 months active, q6–12 months stable on SSA ≥2 years), **TTE intervals, and hepatobiliary imaging**

EHR TIP	WHAT TO INCLUDE
Niacin/pellagra template[1]	**Counseling templates** emphasizing B3 supplementation, dietary support, caregiver involvement when cognitive impairment is present
Advance care planning templates	**Templates** documenting surrogate, code status, goals of care, and discussion date — within 8 weeks of diagnosis for G3/NEC or progressive metastatic disease
Carcinoid syndrome dietary instructions	**Pre-collection instructions** for 5-HIAA (avoid avocados, bananas, pineapples, eggplant, tomatoes, walnuts 48 hours prior); ongoing trigger avoidance counseling

ABBREVIATIONS: B3 = vitamin B3 (niacin); CgA = chromogranin A; CT = computed tomography; EHR = electronic health record; 5-HIAA = 5-hydroxyindoleacetic acid; IM = intramuscular; LAR = long-acting release; MRI = magnetic resonance imaging; NEC = neuroendocrine carcinoma; NCCN = National Comprehensive Cancer Network; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PET = positron emission tomography; PPI = proton pump inhibitor; RV = right ventricular; SC = subcutaneous; SSA = somatostatin analog; TTE = transthoracic echocardiogram; WHO = World Health Organization

Brief Case Examples

SUCCESS — Syndrome-Screening → NET Center Referral → Coordinated VBC Care

Patient: 72-year-old female with 6 months of episodic facial flushing (22 years post-menopause), watery diarrhea labeled IBS for 4 years that has worsened, and new wheezing diagnosed as adult-onset asthma without atopic history. PMH hypertension, hyperlipidemia, GERD on omeprazole. G-8 score 13 (borderline). PCP recognizes syndrome-screening trigger.

Documentation: 'Episodic flushing + chronic diarrhea + adult-onset wheezing in patient ≥65 — syndrome screen ordered. 24-hour urine 5-HIAA 195 µmol/24h (normal <50). CgA 480 µg/L after holding omeprazole 2 weeks. Multiphase CT abdomen: 2.1 cm distal ileal mass + three hypervascular liver lesions. ^{68}Ga-DOTATATE PET: SSTR-positive Krenning 4 at primary and metastases. Coding: Malignant carcinoid tumor of the ileum **(C7A.012)**; secondary carcinoid of liver **(C7B.02)**; carcinoid syndrome **(E34.0)**'

Outcome: Multidisciplinary NET center referral. Started octreotide LAR 30 mg IM q4 weeks with short-acting octreotide 100 µg SC TID bridge. Vitamin B3 (niacin) supplementation initiated. Carcinoid diagnosis flagged in EHR; SC octreotide rescue prescribed; anesthesia and pulmonology notified. Baseline TTE: mild tricuspid regurgitation, NT-proBNP 198 pg/mL (below threshold).[19] Surveillance: 5-HIAA + CgA + CT q3 months; TTE q1 year; ACP initiated. Coding maintained: **C7A.012, C7B.02, E34.0** active; **Z85.030 NOT appropriate while on active SSA**

PITFALL — Z85.030 Misused, Tumor Code Missing, No CHD Surveillance

Documentation as written: '68-year-old female with chronic diarrhea, flushing, and history of carcinoid tumor. Continuing IBS regimen. Coded **Z85.030 + E34.0.**'

Consequence: Z85.030 (history) used despite patient remaining symptomatic and never having received definitive treatment. **E34.0** used as sole code without **C7A** tumor code — non-compliant with ICD-10 sequencing rules requiring tumor code alongside syndrome code. No biomarker workup documented; no NET center referral; SSA therapy not initiated despite confirmed syndrome. No TTE for CHD surveillance. Niacin status never assessed.

RAF Impact: Z85.030 does NOT map to any HCC. **E34.0** does NOT map to any HCC. Active malignant carcinoid disease is invisible on the risk-adjustment record despite clinical reality — patient remains in HCC 0 for this condition rather than HCC 17 (RAF 4.209). Underdocumentation of clinical complexity also limits MA plan resources for SSA therapy, PRRT planning, and palliative-care integration.

Fix: Document explicitly: 'Active malignant carcinoid tumor of the ileum **(C7A.012)** with hepatic metastases **(C7B.02)** and carcinoid syndrome **(E34.0)**. Syndrome workup completed — 5-HIAA 240 µmol/24h, CgA 520 µg/L. Initiating octreotide LAR per NCCN v1.2026. NET center referral, TTE for CHD surveillance, niacin supplementation, carcinoid-crisis precaution flag, ACP within 8 weeks. **Z85.030** NOT appropriate — patient has active disease under active treatment plan.'

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