



## Carcinoid Heart Disease

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The patient, C.P., is a 59-year-old woman who was diagnosed with metastatic carcinoid of the terminal ileum in May 2003. In June 2003, she underwent an extensive resection including hemicolectomy, cholecystectomy, distal pancreatectomy, and splenectomy with metastatic disease in her pancreas, mesentery, and liver. She had been treated with octreotide, everolimus, oxaliplatin, and multiple hepatic artery embolizations in the past eight years and, most recently, capecitabine and bevacizumab with monthly octreotide. She has had intermittent pleural effusions not requiring intervention and a trace pericardial effusion. Her tumor is functional, meaning it demonstrates hormonal hypersecretion which causes flushing, diarrhea, bronchospasm, and abdominal pain.

In April 2013, C.P. had acute onset of jaundice and a portal vein thrombus was detected on an abdominal computed tomography (CT) scan. She was started on low-molecular-weight heparin before being transitioned to warfarin. Her chronic diarrhea worsened and she was diagnosed with *Clostridium difficile* and capecitabine chemotherapy was held until it resolved, taking about four months. During this time, C.P.'s diarrhea was difficult to control and she lost about 20 pounds, causing decline in her performance status, weakness, fatigue, and dyspnea on exertion. Her disease as well as her symptoms, which were driven by carcinoid syndrome, progressed while off treatment. In September, C.P. was tachycardic at a follow-up visit to discuss reinitiating systemic therapy. She was transferred to the emergency department and diagnosed with atrial fibrillation and treated with pharmacologic intervention. An echocardiogram was performed as part of the cardiac

workup and revealed carcinoid heart disease (CHD) with tricuspid regurgitation, pulmonary insufficiency, and enlargement of the right atrium and right ventricle. Her clinical performance status and progressive metastatic carcinoid precluded surgical intervention with valve replacement and she was treated medically with diuretics and metoprolol.

Shortly after being diagnosed with CHD, C.P. developed a large transudative pleural effusion requiring thoracenteses and, ultimately, pleurodesis via video-assisted thoracoscopic surgery. Atrial fibrillation recurred and she was cardioverted. She was ultimately restarted on capecitabine and bevacizumab and her carcinoid stabilized during the next four months. Her CHD has caused several hospitalizations for management of congestive heart failure. Chronic symptoms include dyspnea, extreme weakness and fatigue, anorexia, diarrhea, and significant lower extremity edema. She is very frustrated by the limitations caused by her chronic heart failure.

### Overview of Gastrointestinal Carcinoids

Carcinoid tumors are rare neuroendocrine tumors that are well differentiated, low-to-intermediate grade, and primarily located in the gastrointestinal (GI) tract. Yearly estimates show about 2.5 cases per 100,000 in the Caucasian population and 4 per 100,000 in the African American population (Modlin, Lye, & Kidd, 2003). The incidence seems to be rising because of increased imaging and endoscopic evaluation. Carcinoid tumors are clearly distinct from pancreatic neuroendocrine tumors and poorly differentiated neuroendocrine tumors. Carcinoid tumors are divided into three major anatomic locations (see Figure 1).

The most common carcinoid tumors are located in the appendix and terminal ileum of the small intestine (midgut). Most carcinoids are found incidentally, and many are asymptomatic except small intestine carcinoid, which usually presents with abdominal pain, bowel obstruction, or mesenteric ischemia. More than 25% are multifocal with clusters of intraluminal tumors (Makridis et al., 1990). These patients often are misdiagnosed with irritable bowel syndrome (Kulke & Raut, 2008). About 58%–64% of patients with carcinoid of the small bowel will have metastatic disease to regional lymph nodes or the liver when they are diagnosed (Modlin et al., 2003).

Carcinoid tumors arise from enterochromaffin or neuroendocrine cells of the aerodigestive tract, and these cells stain positive with potassium chromate or chromaffin, indicating that the cells contain serotonin (Goldfinger & Strosberg, 2014). A hypersecretory or functioning carcinoid creates a cluster of symptoms that are caused by the presence of serotonin as well as other vasoactive amines and polypeptides. Facial flushing, secretory diarrhea, bronchospasm, abdominal pain, and hypo or hypertension are the most common symptoms of carcinoid syndrome. Small bowel carcinoids that have metastasized to the liver are most commonly associated with carcinoid syndrome since the liver normally filters these amines and polypeptides and the liver metastases secrete these substances directly into the systemic circulation, bypassing the portal circulation (Strosberg, 2012). Carcinoid syndrome will develop in about

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