A 57-year-old man presented with peripheral edema, fatigue, and dyspnea on exertion. He also had diarrhea and occasional flushing for 4 months. On physical examination he had elevated neck veins at 9 cm with a dominant v-wave, sternal lift with 2/6 systolic ejection murmur at the left upper sternal border and holosystolic murmur at the left lower sternal border, and a moderate lower extremity edema. His echocardiogram is shown (Figure 1). In a 24-hour urine collection, 5-hydroxyindolacetic acid was measured as 182 mg (normal, 2 to 8 mg). A computed tomography scan of the abdomen (Figure 2) showed multiple liver lesions consistent with metastatic carcinoid. Treatment with octreotide, a somatostatin analogue, was initiated. Both the tricuspid and pulmonary valves (Figure 3 and Figure 4) were replaced with bioprosthetic valves. The pathology of his native valves showed fibrous intimal thickenings on the inside surfaces of the valvular leaflets consistent with carcinoid heart disease (Figure 5 and Figure 6). The patient recovered well from surgery, and his symptoms of heart failure improved.

Figure 1. A continuous wave Doppler signal through the pulmonary valve shows increased systolic peak velocity consistent with stenosis and evidence of pulmonary regurgitation. There is rapid dampening of the regurgitant signal with late diastolic reversal of flow (arrow) consistent with pulmonary stenosis and elevated right ventricular pressure. See Movie I through Movie III.
Figure 2. Computed tomography scan of the abdomen showing multiple masses in the liver (arrows).

Figure 3. Surgically excised tricuspid valve appears white, with thickened leaflets and chordae. Courtesy of William D. Edwards, MD, Mayo Clinic, Rochester, Minn.

Figure 4. Excised pulmonary valve cusps. Courtesy of William D. Edwards, MD, Mayo Clinic, Rochester, Minn.

Figure 5. Microscopic examination of the tricuspid valve demonstrating typical fibrous tissue deposition (lower lighter layer) on the ventricular surface of the valve, with preservation of the native valve morphology (upper darker layer). Preserved chords are seen coursing through the carcinoid plaque in the right lower corner. Courtesy of William D. Edwards, MD, Mayo Clinic, Rochester, Minn.

Figure 6. Histopathological examination of the tricuspid valve. Note the clear demarcation between the upper normal valvular tissue and the lower fibrous carcinoid deposition. Courtesy of William D. Edwards, MD, Mayo Clinic, Rochester, Minn.
Carcinoid Heart Disease
Wissam A. Jaber and Kyle W. Klarich

Circulation. 2006;113:e160-e161
doi: 10.1161/CIRCULATIONAHA.105.509471

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/113/7/e160

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2006/02/15/113.7.e160.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/