

Right Ventricular Bypass for Palliation of Cardiac Sarcoma

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A 33-year-old woman with a nonresectable right ventricular sarcoma and pulmonary out-flow tract obstruction underwent a right ventricular bypass operation for symptomatic relief. The patient had an uneventful recovery and was asymptomatic on discharge, without jugular plethora or hepatomegaly. We consider this procedure to be an excellent palliative treatment of malignant right ventricular obstructive symptoms for improving the patient's quality of life. (*Tex Heart Inst J* 1996;23:178-9)

Nearly all primary cardiac malignancies are sarcomas; most of these are angiosarcomas.¹⁻⁴ Approximately 63% of reported angiosarcomas originate in the right atrium,⁵⁻⁷ 25% of which are intracavitary, causing valvular obstruction, right-heart failure, and hemorrhagic pericardial effusion.^{8,9} By the time they are diagnosed, these tumors have already metastasized in 26% of all angiosarcoma patients.¹⁰ From the time of presentation, the disease progresses rapidly, and the prognosis is very poor despite tumor excision and chemotherapy; however, early surgical intervention, combined with adjuvant therapy, may improve results.¹¹ For patients in whom the tumor has spread too extensively for radical surgery, palliative treatment of the obstructive symptoms is indicated.¹²⁻¹⁵

Angiosarcomas originating in the right ventricle are extremely rare among cardiac sarcomas¹⁶⁻¹⁸ and have a worse prognosis than those arising from the atrium because of the limited options for extensive surgical resection. In 1971, Fontan reported a corrective operation for tricuspid atresia¹⁹ that involved using the 1 available ventricle as a pump for the systemic circuit and connecting the right atrium directly to the pulmonary artery. This procedure takes advantage of the low pulmonary resistance in such patients; only a slight rise in systemic venous pressure is required to propel the systemic venous return through the pulmonary circuit into the left side of the heart.^{19,20} We describe a patient who had a non-resectable right ventricular sarcoma with consequent right-heart failure and hemodynamic compromise. By performing a right-heart bypass (a modified Fontan procedure), we successfully palliated the obstructive symptoms. To our knowledge, this is the 1st report of its kind in the world literature.

Case Report

Key words: Chemotherapy, adjuvant; heart neoplasms/surgery; heart ventricle; hemangiosarcoma; pericardial effusion; right ventricular bypass

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A 33-year-old woman was admitted to our hospital with right-heart failure. Her symptoms included shortness of breath, bilateral lower-extremity edema, marked jugular plethora, and abdominal discomfort due to a 2-cm enlargement of the liver. Transesophageal echocardiography revealed a 9.3- × 5.5-cm right ventricular intracavitary mass that involved the tricuspid valve and subvalvular apparatus. The tumor had an irregular surface, suggesting malignancy. Pericardial effusion was apparent. Right-heart catheterization revealed the tumor; the catheter was guided into the pulmonary artery, where pressure measurements were performed. No pulmonary hypertension was found. Endomyocardial biopsy provided the histologic basis for a diagnosis of primary cardiac angiosarcoma. Extensive computed tomographic scanning and abdominal ultrasound studies were performed in order to rule out the presence of distant metastases. The patient was taken to surgery, at which time we found 400 cc of hemorrhagic pericardial effusion and a large right ventricular mass, extending beyond the heart, fused to the pericardial sac. All tumor adhesions to the pericardium were dissected, and part of the extra-

cardiac mass was sent for intraoperative histopathologic analysis, which confirmed the previous diagnosis of angiosarcoma. The involvement of the apex and part of the left ventricle made it impossible to excise the tumor (or even to relieve the obstruction). Palliation of the symptoms with a right-heart bypass (modified Fontan procedure) was considered to be the only surgical option. The procedure was performed without the use of cardiopulmonary bypass. Under systemic heparinization, a 14-mm, preclotted, woven Dacron tube graft (Hemashield®, Meadox Medicals, Inc.; Oakland, NJ) was anastomosed proximally to the right atrial appendage and distally to the trunk of the pulmonary artery (Fig. 1). Heparinization was reversed, and the sternum was closed.

The patient's recovery was uneventful and her venous return improved, as manifested by the disappearance of jugular plethora and bilateral lower-extremity edema. She was discharged on the 8th postoperative day, and by the 2nd postoperative week the hepatomegaly was resolved. The patient was sent to the oncology department for adjuvant chemotherapy. Fifteen months after surgery, the patient was still asymptomatic. Echocardiography revealed a patent graft and slight growth of the intracavitary tumor, with a consequent increase in the right ventricular outflow tract obstruction.

Discussion

Treating malignant cardiac neoplasms is problematic. Most current operations for cardiac sarcomas can be described as macroscopically incomplete resections. Surgeons generally excise as much of the tumor as possible and administer adjuvant therapy in hopes of improving the prognosis and enhancing the patient's quality of life. In the case described above, the patient with a malignant cardiac angiosarcoma did not seek medical assistance until the tumor began obstructing the right ventricle. By then,

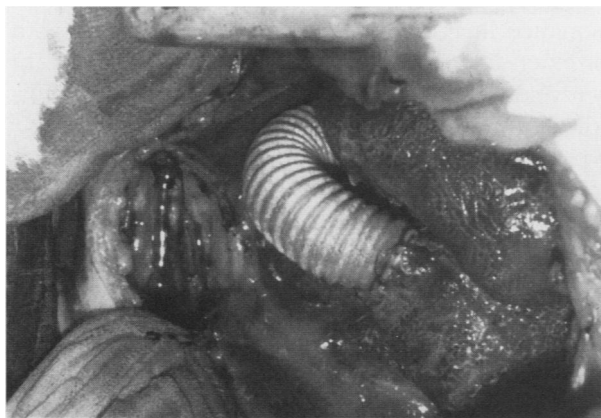


Fig. 1 Photograph of the completed modified Fontan procedure.

surgical resection was impossible because the lesion was so extensive, leaving palliative treatment as the sole alternative to diminish the obstructive symptoms. The right-heart bypass operation shunted the venous circulation toward the pulmonary artery, thus obviating the right ventricle. We believe that the right-heart bypass offers excellent alternative palliative possibilities for patients who have right ventricular obstructive lesions with preserved left cardiac function and normal pulmonary pressures.

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