Progressive Heart Failure
Secondary to a High Output State*

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A high cardiac output state has been an infrequent but previously reversible cause of heart failure in patients with hemodialysis fistulas. A patient with two proximal upper extremity fistulas was found to have progressive depression in his left ventricular function and a high output state. His symptoms did not respond to the removal of the most prominent fistula, suggesting that chronic volume overload from the high output state may irreversibly depress left ventricular function.

High cardiac output states have been ascribed to a number of diseases including systemic arterial-venous (A-V) fistulas, anemia, thyrotoxicosis, beriberi, Paget’s disease of the bone, and uremia.1 High output heart failure in the hemodialysis population with an AV fistula has been described infrequently.** In previous reports, heart failure has been reversible when the fistula was banded or removed. We report a patient with progressive cardiac failure with bilateral upper extremity A-V fistulas with no improvement after removal of the shunts.

CASE REPORT

The patient, a 26-year-old-white man, had end-stage renal disease secondary to postinfectious glomerulonephritis. Hemodialysis was begun in October 1984 after bilateral upper extremity A-V fistulas were placed. In January 1985, an echocardiogram and radionuclide ventriculogram were performed for symptoms of dyspnea on exertion. The echocardiogram (Table 1) was consistent with mild volume overload. The radionuclide ventriculogram demonstrated overall normal cardiac function (resting ejection fraction [EF] 56 percent, exercise EF 64 percent). The patient was maintained on hemodialysis until August 1985 when he was switched to continuous ambulatory peritoneal dialysis after placement of a Tenckhoff catheter. The A-V fistulas remained patent. The chest x-ray film and physical examination results were normal. The patient returned to the hospital in September 1985 with dyspnea and precordial chest pain that increased with inspiration. The physical examination at that time revealed a young man in moderate respiratory distress. The blood pressure was 120/60 mm Hg with a paradox of 10 mm Hg, the heart rate was 110 beats per minute, and the respiratory rate was 28. The neck veins were flat. Heart sounds were normal except for a three component friction rub at the left sternal border. The lungs had bilateral upper lung field rhonchi and scattered lower lung field rales. Both upper extremity fistulas were prominent with palpable thrills over the shunts. There was trace pedal edema. Laboratory findings included a hematocrit value of 28 percent, a blood urea nitrogen level of 67 mg/dl (23.9 mmol/L), and a creatinine value of 16.5 mg/dl (1450 μmol/L). These values were unchanged from six months earlier. Both thyroid function tests (T₄, 4.4 μg/ml; T₃, 125 ng/ml, and T₃RU, 46.9 percent) and thiamine level (66 ng/ml) were normal. Blood for two cultures was drawn and revealed no growth. The chest x-ray film revealed an enlarged cardiac silhouette. An echocardiogram revealed a small pericardial effusion and four-chamber enlargement (Table 1). The patient was treated with intensive peritoneal dialysis and analgesics with resolution of his pain and shortness of breath. A resting radionuclide ventriculogram revealed a marked decrease in the ejection fraction (21 percent) as compared to the previous study (56 percent). The heart rate was noted to fall from 114 to 110 beats per minute with compression of either A-V fistula or both fistulas (positive Branham-Nicoladoni sign). Right and left heart catheterization revealed the following: (1) RA pressure of 8, RV pressure of 36/12, PA pressure of 36/18, pulmonary capillary wedge pressure of 14 mm Hg, and cardiac output of 10 L/min by thermocoupling and 9 L/min by Fick (cardiac index of 5.89 liters/min/m²); (2) normal coronary arteries; (3) normal aortic and mitral valves; and (4) a diffusely hypokinetic left ventricle with an EF of 23 percent. The most prominent of the upper extremity fistulas was removed with resolution of his dyspnea; however, a repeat echocardiogram two months later failed to show any significant change in chamber size or function without change in other parameters (Table 1). Because the patient was extremely sensitive to afterload reduction agents, including nifedipine and captopril, treatment consisted of managing his fluid status with his peritoneal dialysis. The patient died of an unrelated cause in July 1986. On autopsy, the heart was enlarged, 750 g, with left ventricular dilation (20 mm). There was no evidence of valvular or congenital abnormalities. On microscopic examination, interstitial fibrosis was seen, but there was no inflammation or infarction.

DISCUSSION

This report documents a marked decline in cardiac function in a patient with a high output state from an A-V fistula. Contrary to previous reports indicating reversibility of the failure with correction of the fistula, our patient continued to demonstrate significant cardiac depression after removal of his fistula.

Heart failure in patients with chronic renal failure receiving hemodialysis is usually from fluid overload, ischemic heart disease, hypertensive heart disease, or pericardial disease. High output cardiac failure should be suspected when progressive dialysis-resistant failure persists while other factors remain stable. The diagnosis involves the following: (1) demonstration of an increased cardiac output, and if possible, an increased flow through the fistula; (2) demonstration of a positive Branham-Nicoladoni sign; and (3) elimination of other factors (eg, Paget’s, beriberi, anemia, and hyperthyroidism) which are associated with a high output state.

Frank persistent left ventricular failure from the high

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<td>Chamber dimensions, cm</td>
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<td>Left atrium</td>
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<td>Aortic root</td>
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<td>LVID (diastole)</td>
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output state had not been hemodynamically confirmed in the hemodialysis patient. Symptoms of heart failure from an A-V fistula are often attributed to volume overload rather than impaired systolic function. It has been documented, however, in animals who have been subjected to a large A-V fistula, that despite an initial increase in inotropic, Frank-Starling, and chronotropic reserves, there will eventually be severe, irreversible depression of left ventricular function.\1

Previous studies\2,3\ in patients with chronic aortic regurgitation have reported failure of reduction in postoperative heart size after aortic valve replacement when the left ventricular end-systolic dimension exceeds 5.5 cm. Such observations propose that compensatory dilation of the volume overloaded left ventricle has reversible limits. The present report further suggests that volume overload of the LV from a high output state with competent mitral and aortic valves has similar limits to compensatory dilation.

Intervention with banding or removal of the fistula, therefore, should be considered in the management of patients with dialysis-resistant heart failure. Echocardiographic left ventricular dimensions may be helpful in monitoring these patients and guiding the appropriate timing of therapy.

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Ring-Calcification of Coronary Artery Aneurysms in an Adolescent*

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A 17-year-old asymptomatic boy with a history of Kawasaki disease in infancy was found to have an abnormal ring-calcification as seen in the chest roentgenogram and the electrocardiographic evidence of anteroseptal myocardial infarction. Coronary angiogram revealed aneurysms associated with ring-calcifications in the right coronary artery and subtotal obstruction of the left anterior descending coronary artery. A plain chest roentgenogram may, at times, be useful for identifying coronary aneurysms in patients with a possible history of Kawasaki disease.

Kawasaki disease is an acute febrile illness of unknown etiology, which occurs predominantly in infancy and early childhood.\1\ Coronary artery aneurysms develop in approximately 15 to 20 percent of children during the acute stage of the disease.\2\ Although follow-up coronary angiography one to two years later has revealed regression of the aneurysms in more than one half the affected children, death may result from myocardial infarction secondary to thrombotic occlusion of the coronary artery, usually within one year after the onset of the disease.\3\4

Described herein is an unusual case, a 17-year-old asymptomatic boy with a history of Kawasaki disease in infancy, who first presented to us with an abnormal ring-calcification demonstrated on chest roentgenogram and through electrocardiographic evidence of anteroseptal myocardial infarction.

CASE REPORT

A 17-year-old school boy was referred to the hospital for evaluation of an abnormal ECG, although he was active and asymptomatic. He had a history of persistent fever for six weeks, unresponsive to the treatment of antibiotics, associated with rash and reddening of the lips and mucous membrane at the age of 15 months. On examination, the pulse rate was 84 beats per minute and the blood pressure 128/76 mm Hg. There were prominent third and fourth heart sounds. Chest x-ray film showed normal-sized heart and clear lungs. An oval calcification was noted at the base of the heart on lateral and oblique projections (Fig 1). The ECG showed abnormal Q waves associated with ST elevations in the anterior chest leads, consistent with anteroseptal myocardial infarction. The ST depressions in the inferolateral leads were also noted. Thallium myocardial scintigram revealed fixed defects in the anteroseptal segments.

Based on his history, the ECG, and the myocardial scintigram, a presumptive diagnosis of myocardial infarction secondary to Kawasaki disease was made. An oval calcification in the chest was then thought to represent calcification of a coronary aneurysm. Cardiac catheterization revealed the following: pulmonary artery pressure, 1118

Coronary Artery Aneurysms in an Adolescent (Doi et al)

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