Systemic Artery-to-Pulmonary Artery Fistula*

A New Therapeutic Approach

Douglas C. Wolf, M.D.; John J. Wurtzbacher, M.D.; and Joseph Horton, M.D.

A patient with systemic artery-to-pulmonary artery fistulae is described. Historically, all therapy of this type of lesion has been surgical. We describe a new mode of treatment, therapeutic embolization, which obviates surgical intervention and its attendant risks. Embolization is an alternative to surgery that should be considered in selected cases.

Since the initial case report in 1947,1 34 cases of systemic artery-to-pulmonary artery (SAPA) fistulae have been reported.2,3 Because of their adverse hemodynamic effects, risks of bacterial vegetations and rupture, surgical correction has been almost uniformly performed when this lesion is recognized. Therefore, it is important to confirm the presence of these fistulae when they are suspected. Because of their rarity, knowledge of their consequences and of the medical approaches to them is limited. We present a case of SAPA fistulae presenting as a continuous heart murmur. A successful alternative to surgical treatment will be described.

CASE REPORT

The patient is a 29-year-old white man with a history of refractory anemia, weight loss, and negative PPD. He had been started empirically on a one-year course of isoniazid in February, 1979, when a bone marrow biopsy done at another hospital revealed nonspecific granulomata. Cardiac examination at the time was reported to be unremarkable. He improved over the following year with weight gain and correction of his anemia.

He was readmitted to that hospital in May, 1980, with a fever of 38.5°C, chills, sinus congestion, and a heart murmur. The patient was transferred to West Virginia University Hospital. Physical examination revealed a pale, thin man in no acute distress. His blood pressure was 120/40 mm Hg, pulse rate 110 and bounding, respirations 18, and temperature 36.7°C. Continuous precordial (grade 2/6) and right supraventricular (grade 4/6) murmurs were present. The lungs were clear to percussion and auscultation. The spleen tip was palpable. Multiple "spider" angiomata were noted on the trunk.

Initial laboratory work-up revealed a hemoglobin of 11.0 g/dl with microcytic indices, normal electrolytes, low cholesterol, and elevated LDH and alkaline phosphatase. There was a reaction of 20 × 24 mm induration to intermediate-strength PPD skin testing. Bone marrow aspirate and biopsy revealed myeloid hyperplasia and epitheloid granulomata. Acid-fast stains of bone marrow aspirate and of multiple sputum and urine specimens were negative. Cultures of each were also negative. The electrocardiogram revealed normal sinus rhythm with prominent voltage and an axis of +60°. M-mode echocardiography revealed mild left ventricular dilatation. Phonocardiography confirmed the continuous murmur.

An aortic arch injection at the time of cardiac catheterization revealed multiple communications from the right subclavian, thyrocervical, and internal mammary arteries to right upper lobe pulmonary artery branches. Right heart and pulmonary artery pressures were normal. Oximetry revealed a marked oxygen saturation step-up in the right upper lobe pulmonary arteries.

The patient was discharged and readmitted several months later. Right subclavian angiography precisely localized the SAPA fistulae. Many of the communications were selectively catheterized, and five major ones embolized. For this, a mixture of polyvinyl alcohol foam (PVA) and gelfoam suspended in contrast material was used, and delivered by the Kerber flow-control technique.4 The three largest fistulous vessels were occluded at the first sitting. The supraventricular murmur disappeared and the precordial murmur decreased in intensity. At a second sitting approximately five days later, two other major feeders were occluded. At this time all murmurs had ceased. Figures 1, 2, and 3 show the fistulae before and after embolization.

FIGURE 1. Injection into the right subclavian artery opacifies the ascending cervical ( ), vertebral ( — ) and internal mammary ( ) arteries, among others. A laterally directed unnamed artery ( — ) arises from the internal mammary artery. Prompt filling of the right upper and lower pulmonary arteries ( ) and even the right main pulmonary artery ( ) follows. All highly tortuous vessels are part of the fistulae.

DISCUSSION

Vessels most frequently involved in SAPA fistulae are internal mammary and intercostal arteries, as well as anomalous branches of the subclavian artery and the...
No specific diagnosis was made regarding the etiology of the fistulae in this patient. Although it could have been due to an inflammatory or granulomatous lesion, the angiographic appearance was indistinguishable from that of a congenital racemose arteriovenous malformation.

There are several potential complications of SAPA fistulae, chiefly volume overload and resultant left ventricular failure. In this patient, left ventricular dilatation demonstrated by M-mode echocardiography was consistent with volume overload from the sizable fistulae. Although not reported to date in SAPA fistulae, the potential for bacterial seeding and enlargement with rupture exist as noted with patent ductus arteriosus. Neither of these complications was present in this case. Sizable systemic arterial fistulae, therefore, require correction for several reasons, foremost being the propensity for heart failure.

The rarity of these lesions, as well as their prompt surgical management once they are recognized, may explain the infrequency of complications noted. Surgical intervention may involve resection of the vascular anomaly from the lung parenchyma, lobectomy, or pneumonectomy. All but two of the reported cases were corrected surgically. In the treatment of SAPA fistulae, embolization is the least invasive method, sparing the patient the risks of thoracotomy and general anesthesia. The patient would be subjected to the normal risks of embolization, namely that of infarction of normal tissue. In treating subclavian branches, the possibility of emboli passing to the vertebral circulation would be present; however, using the “flow-control” technique, these risks are negligible.

Selection of patients for therapeutic embolization relies on identification of branches supplying the fistulae which do not supply normal tissue. If a branch supplying the fistulae cannot be selectively catheterized, then it cannot be safely embolized. This alone would preclude a patient from this mode of treatment.

In summary, SAPA fistulae are rare and can present as continuous murmurs over the chest. The origins of these malformations are often unclear, but may be congenital or secondary either to chronic inflammation or to trauma. Clinical suspicion of significant thoracic
vascular malformations must be confirmed angiographically and should be treated definitively. This is the first reported case in which a nonsurgical approach has been used in the treatment of SAPA fistulae. Embolization is an alternative to surgery and should be considered in most cases.

SELECTED REFERENCES
1 Burchell HB, Claggett OT. The clinical syndrome associated with pulmonary arteriovenous fistulas, including a case report of the surgical cure. Am Heart J 1947; 34:151

Rapid Attenuation of Response to Nifedipine in Primary Pulmonary Hypertension*

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In a 22-year-old woman with primary pulmonary hypertension resistant to all previous attempts to reduce the pulmonary vascular resistance, there was dramatic improvement after the first dose of nifedipine, 20 mg po, which was not sustained with subsequent doses. While there was a persistent reduction in systemic vascular resistance, the initial drug-related reduction in pulmonary vascular resistance was progressively attenuated with the subsequent four doses of nifedipine, 20 mg.

Primary pulmonary hypertension may be caused by pulmonary vasoconstriction. Attempts to alleviate vasoconstriction have included the use of oxygen, acetylcarnine, isoproterenol, phenolamine, diazoxide, tolazoline, prostacyclin, hydralazine, diltiazem, verapamil, and nifedipine.*• We report a patient with primary pulmonary hypertension in whom nifedipine caused a dramatic reduction in pulmonary hypertension initially. However, tachyphylaxis developed with subsequent doses.

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**Case Report**

A 22-year-old black woman was first admitted to this hospital in February, 1981 for progressive dyspnea on exertion and near syncopal episodes. These had begun approximately five months prior to her admission when she noted dyspnea on climbing stairs. Physical examination revealed the presence of a right ventricular heave and a grade 3/6 holosystolic heart murmur loudest at the upper left sternal border with radiation down the left sternal border toward the left mid-clavicular line. The lungs were clear to percussion and auscultation. There was moderate jugular venous distention without peripheral edema. There was RVH noted on the electrocardiogram, and cardiac enlargement noted on the chest x-ray film without any pulmonary parenchymal abnormalities. A ventilation/perfusion lung scan gave normal findings. Echocardiogram showed a small left ventricle with a hypokinetic septum and decreased systolic function, reduced EF slope and an enlarged right ventricle.

The patient underwent left and right heart catheterization. Right atrial (RA) pressures (mm Hg) were elevated to a = 25, v = 18 (mean RA-17); right ventricular pressures, 90/8; pulmonary artery pressures (PAP), 90/60 with a mean of 65. Aortic pressures were 140/80; left ventricular pressures (LVP), 140/80; mean pulmonary capillary wedge (PCW), pressure 8. Peripheral pulmonary artery angiogram with hand injection demonstrated the “pruned tree” effect found with primary pulmonary hypertension.

The patient failed to respond to a variety of therapeutic agents including 100 percent oxygen, 60 mg of tolazoline IV, 0.4 mg nitroglycerin SL, and prazosin 1 to 5 mg and hydralazine, 25 and 50 mg orally.

The patient was re-admitted to the hospital on September 30, 1981. Results of the physical examination had changed from the previous admission with the development of marked jugular venous distention to the angle of the mandible while sitting upright, hepatomegaly with a span of 18 cm in the right midclavicular line, a palpable splenic tip and 2+ pitting edema in the lower extremities extending to the mid thighs. The cardiac examination, electrocardiogram and chest x-ray film were unchanged. Radionuclide gated wall motion study showed a markedly enlarged

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Table 1—Summary of Hemodynamic Responses to Nifedipine 10 mg and 20 mg (1st, 2nd and 4th Doses)

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