confirmed significant amyloid deposition in this location. We feel that a more reasonable explanation for the echocardiographic findings is biventricular outflow tract obstruction secondary to massive myocardial amyloid deposition.

The lack of increased QRS voltage on the ECG has been noted to be a helpful distinguishing feature between amyloidosis and other forms of ventricular hypertrophy, and although in the patient presented here the diagnosis of systemic amyloidosis was established by the constellation of clinical, electrocardiographic, and echocardiographic findings, previous reports have documented that patients with extensive cardiac amyloidosis may not be recognized clinically or are mistaken to have hypertrophic cardiomyopathy by echocardiography. Because the echocardiographic manifestations of amyloid heart disease and hypertrophic cardiomyopathy can sometimes be very similar, endomyocardiobial biopsy may be necessary when the exact diagnosis is in doubt as the treatment of these two conditions is obviously dissimilar.

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Bilateral Pulmonary Artery Stenosis Associated with Pericarditis*

Results of Surgery and Follow-up by Magnetic Resonance Imaging

Mark Jay Friedman, M.D.; George E. Gabor, M.D.; Marcia C. Fishman, M.D.; and Randall B. Griepp, M.D.

Reported is a patient with bilateral pulmonary artery stenosis which developed concurrently with an episode of idiopathic pericarditis. This association has been unreported previously. The stenosis was corrected surgically. Postoperatively, magnetic resonance imaging was used to follow the status of the pulmonary arteries serially.

Aquired pulmonary artery stenosis has several etiologies. * There has been no previous report of this condition developing in association with acute idiopathic pericarditis. The patient reported herein had such an unique association. The diagnosis was confirmed by cardiac catheterization and pulmonary angiography. The lesions were surgically corrected and postoperatively the status of the pulmonary arteries has been effectively followed by serial magnetic resonance imaging (MRI) avoiding repeated invasive procedures.

Case Report

A 31-year-old white woman with no prior cardiovascular history was admitted to a community hospital in October, 1983 with a vague history of fever, malaise, and exertional dyspnea. An echocardiogram revealed pericardial effusion; a diagnosis of pleuropericarditis was entertained. The patient was treated with digoxin, furosemide, and prednisone with resolution of symptoms. Shortly after discharge, while the steroid therapy was being tapered, her symptoms returned. The patient was referred to The Brooklyn Medical Center for evaluation in January 1984.

On physical examination, her blood pressure was 120/90 mm Hg with no pulsus paradoxus. The pulse rate was 120 beats/min and irregular. She was afebrile. There was a grade 3/6 midystolic murmur confined to the second left intercostal space and a trace of pedal edema.

Echocardiographic examination of the chest revealed an enlarged cardiac silhouette consistent with a large pericardial effusion. The echocardiogram revealed sinus tachycardia and nonspecific ST and T wave abnormalities. The echocardiogram confirmed the presence of a large pericardial effusion, but there was no evidence of tamponade. Cardiac chamber sizes were normal.

Laboratory testing revealed a white blood cell count of 12,000/cu mm and hemoglobin, 15 g/dl. Blood urea nitrogen, glucose and electrolyte levels were normal. Test results of antinuclear antibody, anti-DNA, rheumatoid factor and cold agglutinins were negative. Complement levels as well as T3 and T4 levels were normal. The tuberculin skin test result was negative as were serologic tests for syphilis.

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The patient underwent a therapeutic and diagnostic pericardial window procedure with drainage of 2 L of straw-colored fluid. Cultures of pericardial fluid were negative for bacteria and fungus. Cytologic study was negative for malignant cells. Histopathology of the pericardium indicated mild perivascular nonspecific chronic inflammation and focal reactive mesothelial hyperplasia.

A repeat echocardiogram five days later showed no evidence of pericardial effusion. The pericardium was thickened and the right ventricle dilated. On follow-up study two months later (March, 1984), there was no pericardial effusion of significant pericardial thickening. The right ventricle was dilated with paradoxic septal motion. At that time, the patient still complained of exertional dyspnea. On physical examination the only change was fixed splitting of the second heart sound over the pulmonic area and a grade 3/6 systolic ejection murmur now heard over the entire precordium and radiating to both the shoulders and neck. The electrocardiogram showed a new right bundle branch block pattern with right axis deviation consistent with right ventricular hypertrophy. Cardiac catheterization was performed.

Right heart catheterization showed right atrial mean pressure of 6 mm Hg, right ventricular pressure was 70/10 mm Hg, and left main pulmonary artery pressure was 60/10 mm Hg with a mean of 36 mm Hg. The left distal pulmonary artery pressure was 20/10 mm Hg, right main pulmonary artery pressure was 70/10 mm Hg, and right distal pulmonary artery pressure was 40/7 mm Hg. There was no evidence of shunting in either direction.

Left ventriculography revealed a normally functioning left ventricle with an ejection fraction of 74 percent. Left ventricular end-diastolic pressure was 6 mm Hg.

Pulmonary angiogram revealed stenosis of both the right and left main pulmonary arteries (Fig 1).

Surgical repair was delayed ten months because of the recent pericarditis and was performed January 31, 1985. At operation, there were moderately dense adhesions binding the heart to the pericardium. The heart was slightly enlarged. There was a dense inflammatory reaction involving both pulmonary arteries as they neared the pericardial reflection. Both arteries were markedly narrowed by proliferation of tissue from within. The external diameter appeared to be relatively normal, but the surgeon described them as feeling "like wooden rods secondary to severe fibrosis of the vessel wall." The right pulmonary artery was narrowed to approximately 2 or 3 mm all the way out to its branching. The left artery was not as severely narrowed and had an estimated internal diameter of 6 mm. Unroofing and grafting of the walls of the pulmonary arteries were performed.

The pathology specimen taken from the right pulmonary artery showed fragments of fibroconnective tissue with severe chronic inflammation, rich in plasma cell infiltrate.

![Figure 1](image1.png)  
**Figure 1.** Angiogram. A (left panel), posterior-anterior view demonstrates narrowing of the right pulmonary artery (arrow). B (right panel), left anterior oblique view demonstrates narrowing of the left pulmonary artery (arrow).

![Figure 2](image2.png)  
**Figure 2.** Axial cardiac gated MRI, three months postoperation (TE 30ms and TR 560ms). A (upper panel), minimal narrowing of the distal right pulmonary artery (arrow). B (center panel), widely patent left pulmonary artery (arrow). C (lower panel), nine months postoperation, there is further narrowing of the right pulmonary artery (arrow).

Bilateral Pulmonary Artery Stenosis (Friedman et al)
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For Brief Summary, please see following page.

*While it is recognized that $\beta_2$-adrenergic receptors are the predominant receptors in bronchial smooth muscle, recent data indicate that 10 to 50% of the $\beta$ receptors in the human heart may be $\beta_2$ receptors. The precise function of these receptors, however, is not yet established.
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Fatalities have been reported in association with excessive use of inhaled sympathomimetic aerosols. The exact cause of death is unknown, but cardiac arrest was the unexpected development of a severe acute asthma crisis and subsequent hypoxia is suspected.

In the event of overdosage, symptoms include anginal pain, hypertension, hypokalemia, and tachycardia. Cardiac arrest and death may be associated with abuse of the aerosol inhaler. Deaths are believed to be due to arrhythmia in the setting of asthma rather than a direct sympathomimetic effect. Deaths have occurred in the presence of an ischemic coronary illness or during surgery for coronary artery disease. Deaths have occurred in patients who were taking monoamine oxidase inhibitors or tricyclic antidepressants. Deaths have occurred in patients who were taking sympathomimetic agents. Deaths have occurred in patients who were taking beta-blockers. Deaths have occurred in patients who were taking beta-blockers and sympathomimetic agents.

OVERDOSAGE: Manifestations include anginal pain, hypertension, hypokalemia, and exacerbation of the effects listed in Adverse Reactions. Cardiac arrest and death may be associated with abuse of the aerosol inhaler. Dialysis is not appropriate treatment for the judicious use of a cardioselective beta-receptor blocker, such as metoprolol tartrate, is suggested, bearing in mind the danger of inducing an asthmatic attack.

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The Congress will feature state-of-the-art lectures, clinical colloquia and oral and poster presentations. Topics will include new aspects in the treatment of ARDS; immunologic and interstitial lung disease; fungal disorders; heart/lung transplant; sleep apnea; weaning from mechanical ventilation; laser applications; primary prevention of coronary artery disease; systemic hypertension; arrhythmia detection and treatment; and advances in the pathophysiolo of chronic airflow limitations. Medical and technical exhibits will enhance the program.

For information contact:
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Immediately after surgery, the main pulmonary artery pressure was 30/10 mm Hg. A pulmonary angiogram prior to discharge revealed normal diameters of both pulmonary arteries.

Bilateral systolic murmurs remained audible but less intense than preoperatively. The MRI on March 1985, three months postoperation, showed the main pulmonary artery and the left pulmonary artery to be of good caliber while the distal right pulmonary artery was slightly narrowed (Fig 2A and B). Nine months after surgery, a repeat study showed progressive narrowing of the distal right pulmonary artery (Fig 2C). One year after surgery, the MRI was unchanged. The patient has remained asymptomatic and had an uncomplicated pregnancy a year and a half after surgery.

**Discussion**

Bilateral branch pulmonary artery stenosis in this patient occurred after an episode of pericarditis. The etiology of the pericarditis was not established by any of the diagnostic studies, including cytology and histology. Viral studies were not obtained; nevertheless, viral infection remains the most likely cause of the pericarditis, particularly as the patient has remained free of systemic disease for over two years.

The gamut of reported causes of branch pulmonary artery stenosis includes syphilitic aortic aneurysms, as well as anterior mediastinal tumors. These generally cause extrinsic compression, although invasion of the artery may occur. Sarcoidosis can cause stenosis by extrinsic node compression or by granulomatous infiltration of the pulmonary artery. Two years of observation have adequately excluded these conditions in our patient.

Takayasu's disease has been reported to be a cause of branch pulmonary artery stenosis, as well as pericarditis. This is usually in the context of more generalized disease involving the aortic branches, although cases of isolated pulmonary involvement may occur. Our patient has no finding suggestive of aortic involvement by MRI. In addition, the pathology of the pulmonary artery in our patient does not resemble Takayasu's arteritis. Congenital causes of pulmonary artery stenosis are easily eliminated here on the basis of the history and the pathologic findings. This patient represents a previously unreported cause of bilateral pulmonary artery stenosis.

Treatment of our patient involved unroofing and grafting the walls of the pulmonary arteries. Alternatively, a jump graft over the area of stenosis might have been used. Balloon angioplasty has been used in congenital varieties, but considering the rigidity of the vessels described at surgery, it is doubtful that it would have been successful in this case.

In the past, follow-up would have required repeat pulmonary angiography. This case shows the latest imaging technique (MRI) to be an effective, noninvasive method for following the progress of the lesions after surgery. Indeed, the initial diagnosis might have been established by MRI. Hemodynamic aspects of pulmonary hypertension caused by bilateral branch pulmonary artery stenosis can also be evaluated noninvasively by echocardiographic and Doppler techniques. This patient will be followed in this fashion to augment the anatomic information obtained from the MRI.

In conclusion, this patient had bilateral pulmonary artery stenosis associated with pericarditis and inflammatory reaction. Postoperatively, MRI has been used effectively to follow the patient's clinical course.

**References**


**Noninvasive Imaging of Left Coronary Arteriovenous Fistula**

Jean-Luc Vandenbossche, M.D.; Herbert Felice, M.D.; Andre Grivegnee, M.D.; and Marc Englert, M.D.

We investigated a patient with coronary arteriovenous fistula (CAVF) by two-dimensional echocardiography (2-D), cardiac Doppler, cardiac catheterization and nuclear magnetic resonance (NMR). These investigations clearly showed the abnormal vascular structures. NMR is another valuable noninvasive and safe method of confirming the presence of a CAVF.

Since the first description of CAVF in 1865, the methods used for their diagnosis have progressed from invasive coronary catheterization to 2-D, contrast 2-D, and more recently, Doppler echocardiography. This report is the first to describe the use of a further noninvasive method to confirm the diagnosis: NMR.

**Case Report**

A 21-year-old man was referred in 1985 for investigation of a heart murmur which had been diagnosed on routine examination. He was completely asymptomatic and gave no relevant past medical history. Examination revealed a continuous grade 2/6 murmur at the left fourth intercostal space. An electrocardiogram and a chest radiograph showed normal findings. Phonoangiography confirmed the auscultatory findings together with a continuous murmur of high frequency maximally in the suprasternal region. Two-dimensional echocardiography revealed a dilated vascular structure 1-1.5 cm in diameter along the left atrioventricular groove which corresponded to the circumflex artery (Fig 1). On the inferior surface there was a more marked dilatation of this vessel at the normal site of the coronary sinus. Color Doppler showed a continuous turbulent flow pattern of high intensity maximally in the region of the coronary sinus. The cardiac chambers and valvular structures appeared normal. A diagnosis of coronary arteriovenous fistula (CAVF) between the circumflex artery and the coronary sinus was made. Nuclear magnetic resonance (NMR) showed a dilated vascular structure running along the left atrioventricular sulcus. The myocardium appeared to be of uniform density and normal thickness (Fig 2). The images were T weighed and obtained with a cardiac...