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# Severe Takayasu's Arteritis of the Pulmonary Arteries: Report of a Case With Successful Surgical Treatment

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**Although involvement of the aorta and its large branches is more common, Takayasu's arteritis involving the pulmonary arteries is well recognized. This report describes an adolescent girl with an uncommonly severe form of Takayasu's arteritis involving the pulmonary arteries. A successful surgical treatment is presented.**

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Takayasu's arteritis is a nonspecific arteritis affecting portions of the aorta, its major branches or the pulmonary arteries, alone or in combination. It may cause stenosis, occlusion or aneurysm of any of these vessels. Its various synonyms include: aortic arch syndrome, pulseless disease, Martorell's syndrome, atypical coarctation, young female arteritis and reversed coarctation (1,2).

The disease was first described by Savory (3) in 1856. Takayasu (4) in 1908 reported the ocular changes in a 21 year old woman. The Discussion section of Takayasu's case report quotes Onishi's description of two similar cases and his correlation of the ocular findings with a lack of pulses in the upper limbs. The clinical features of the disease were further clarified by Shimigu and Sano (5) in 1951. The entity was named "Takayasu's arteritis" in 1954 (6). In 1967, Uneno et al. (7) described three types on the basis of the location and extent of the arteritis affecting the aorta. In 1977, Lupi-Herrera et al. (8) described an additional variant with pulmonary artery involvement, which they designated type IV (Fig. 1).

Pulmonary artery involvement in Takayasu's arteritis was not recognized until 1940 when Oota (9) at postmortem study demonstrated pulmonary artery lesions similar to those of involved systemic arteries. Subsequent reviews (8,10–12) suggested that about 50% of patients with Takayasu's arteritis have involvement of the pulmonary arteries. Pulmonary vasculitis has usually been mild, with only spotty involvement of lobar, segmental or subsegmental arteries. A few cases with significant pulmonary hypertension have been reported. These cases have been associated with obliteration of the pulmonary vascular bed distal to the right and left pulmonary arteries and, in some cases, with left atrial hypertension (8,12,13).

To our knowledge there are no reports of pulmonary artery surgery in the treatment of this disease. The patient reported in this study is important because she presented with an unusual kind of pulmonary vasculitis that made surgical palliation feasible.

## Case Report

A 15 year old white girl was referred to Texas Heart Institute and Texas Children's Hospital for evaluation and possible surgical treatment of pulmonary vasculitis related to Takayasu's arteritis. She reported a 1 year history of progressive shortness of breath and poor exercise tolerance. At the time of admission, she became severely dyspneic after climbing half a flight of stairs. She had a 3 year history of Takayasu's arteritis involving the descending aorta and the renal arteries. Two years before referral, she had right renal artery bypass surgery and left renal autotransplantation to the left iliac fossa because of hypertension and poor renal function. Subsequently, she was given corticosteroids and an immunosuppressive agent without remission of her disease. She had a 1 year history of a pulmonary murmur and progressive right ventricular hypertrophy on electrocardiography. Three months before referral, she underwent lim-

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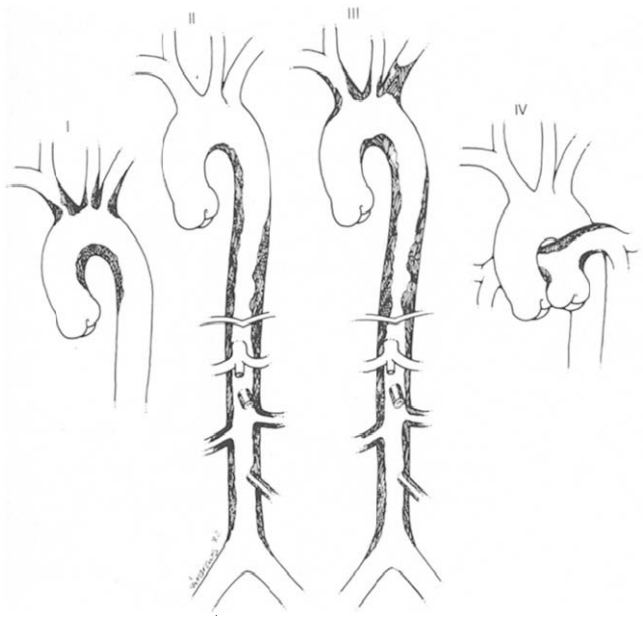



**Figure 1.** Classification of Takayasu's arteritis. Type I: involvement of the aortic arch and its brachiocephalic vessels. Type II: involvement of the thoracoabdominal aorta. Type III: involvement of both type I and type II. Type IV: pulmonary artery involvment (associated with involvement of systemic arteries).

ited cardiac catheterization which demonstrated severe pulmonary artery involvement and related right-sided heart failure.

**Clinical findings.** The blood pressure was 154/84 mm Hg in the right arm and 120/70 mm Hg in the right leg. The heart rate was 90 beats/min and the respiratory rate 20/min. The patient had mild peripheral cyanosis. There was a grade 2/6 systolic ejection murmur in the pulmonary area radiating into the lung fields, and there was a grade 3/6 lower-pitched systolic ejection murmur in the tricuspid area. The lungs were clear to auscultation and percussion. The liver edge was 3 cm below the right costal margin. The pulses were diminished in the lower limbs.

*The chest X-ray film* showed moderate cardiomegaly with prominent right atrial and main pulmonary artery contours. The distal pulmonary vascularity was diminished more on the right than on the left. The electrocardiogram revealed right ventricular hypertrophy and right atrial enlargement.

**Catheterization and angiographic findings.** Cardiac catheterization disclosed a mixed venous oxygen saturation of 54% and systemic saturation of 94%. The right ventricular and main pulmonary artery pressures were elevated with a systolic level of 100 mm Hg. There was a significant pressure gradient at the origin of the left pulmonary artery, with a left pulmonary pressure of 25/14 mm Hg. The right pulmonary artery was almost totally occluded. When the catheter was placed in the proximal right pulmonary artery, the mean pressure was 7 mm Hg with a wedge configuration (Fig. 2). Angiography demonstrated almost total occlusion

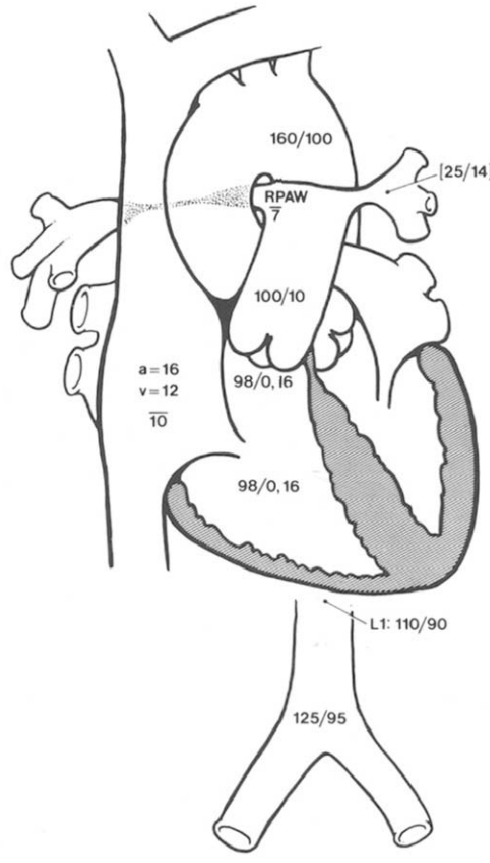
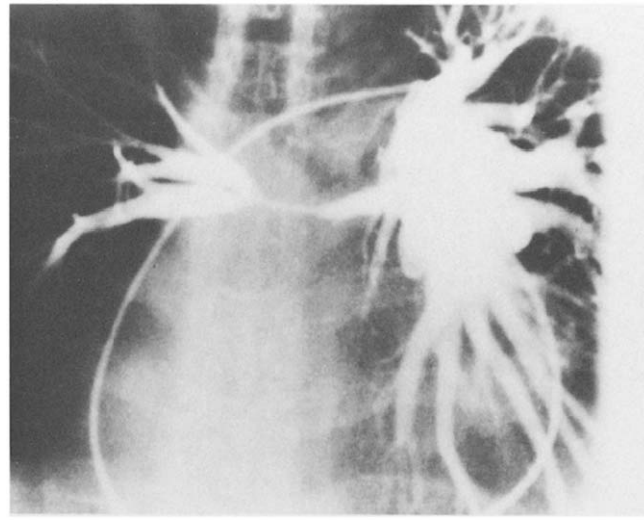



**Figure 2.** Cardiac catheterization findings with intracardiac and vascular pressures. L = lumbar; RPAW = right pulmonary artery wedge pressure. Values with overlying dash or after a comma are mean pressures.

of the right pulmonary artery (Fig. 3 and 5). The proximal left pulmonary artery was also severely stenotic (Fig. 4 and 5). The right ventricle was hypertrophied and dilated and

**Figure 3.** Pulmonary artery angiogram demonstrating almost total occlusion of the right pulmonary artery.



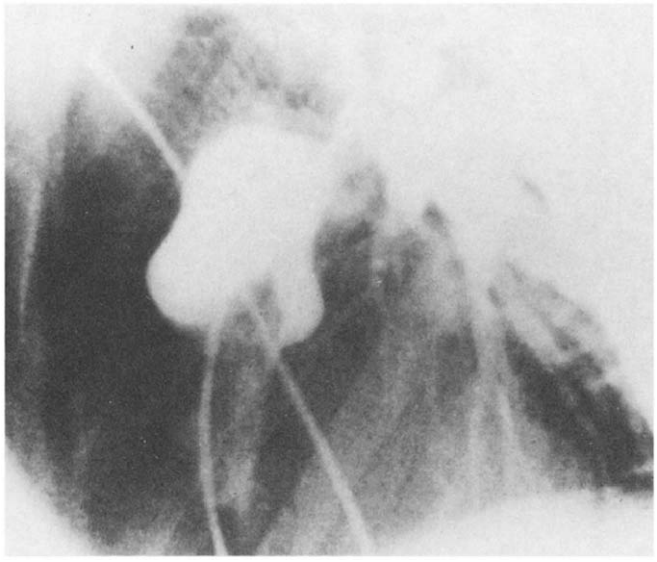


**Figure 4.** Pulmonary artery angiogram demonstrating severe proximal stenosis of the left pulmonary artery.

its contractility was poor. There was massive tricuspid insufficiency.

**Cardiac surgery and postoperative course.** Surgical treatment was recommended to relieve the pulmonary artery obstruction and was performed on October 19, 1983. Exposure was obtained through a midline incision and median sternotomy. Cannulation for cardiopulmonary bypass consisted of individual cannulation of the superior and inferior venae cavae through the right atrium and arterial cannulation of the distal ascending aorta. Bypass was instituted and caval snares were applied. The ascending aorta was cross-clamped and cold crystalloid injected into the aortic root to induce cardiac arrest. The main pulmonary artery and proximal right and left pulmonary arteries were found to be markedly thickened and fibrotic with surrounding inflammatory reaction and an enlarged lymph node. Biopsy of the pulmonary artery wall revealed adventitial fibrosis and chronic inflammation. Biopsy of a lymph node revealed chronic lymphadenitis.

The main pulmonary artery was opened transversely at the bifurcation and the incision extended into the left main pulmonary artery beyond the point of stenosis. This area was closed with a woven Dacron patch relieving the obstruction to the left main pulmonary artery. The right main pulmonary artery lateral to the superior vena cava and distal to the obstruction was isolated and opened longitudinally. A 14 mm woven Dacron tube graft was sutured end to side to the distal right pulmonary artery. The graft was placed anterior to the superior vena cava and the ascending aorta, and was sutured end to side to the left main pulmonary artery patch graft (Fig. 5). After discontinuation of cardiopulmonary bypass, the main pulmonary artery pressure was 35/21 mm Hg.

*The postoperative course was uncomplicated.* The patient was released from the hospital on the 10th postoperative day. At the time of discharge, her exercise tolerance had improved dramatically. She was able to climb three flights of stairs without dyspnea. On physical examination, she was no longer cyanotic, there was no hepatomegaly or murmur of tricuspid insufficiency and the murmur previously audible in the pulmonary area was diminished. Follow-up after 5 months revealed continuing improvement in exercise tolerance and general well-being.

## Discussion

**Previous reports on Takayasu's pulmonary arteritis.** There have been four small series of reports describing patients with various clinical and postmortem findings in Takayasu's arteritis involving the pulmonary arteries. Nasu (10) in 1962 reported 12 autopsy cases found during a review

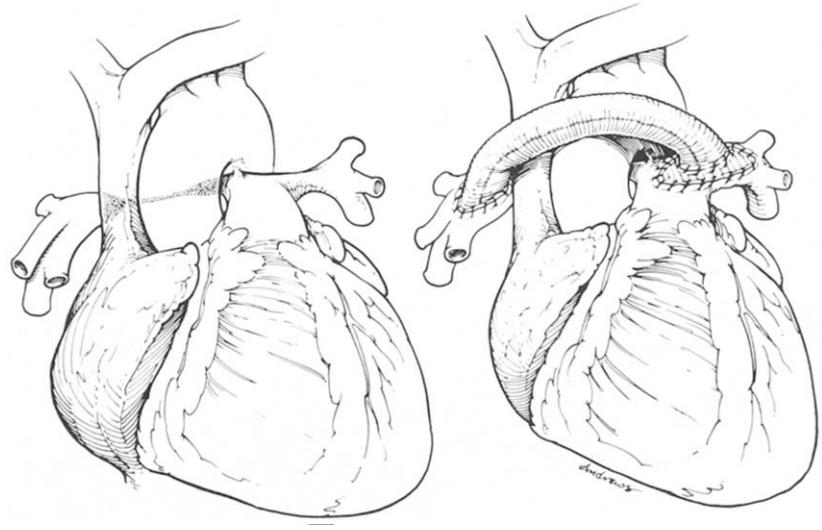


**Figure 5.** Artist's conception of the pathologic features demonstrated by angiography (**left**) and of the surgical repair (**right**).

of 21 autopsies of individuals who died with Takayasu's arteritis. That review indicated that the pulmonary trunk and the main pulmonary branches were involved with arteritis. There was no comment about degree of involvement or about involvement of smaller pulmonary arteries in the report. Moreover, no clinical information was provided about the patients. Kozuka et al. (11) reviewed 35 cases of Takayasu's arteritis and found 16 with pulmonary vasculitis. They described the findings on pulmonary angiography with injection of contrast medium into the right atrium or in large systemic veins. In one case, the right pulmonary artery failed to fill. In another case, there was constriction of the proximal part of the right pulmonary artery and no filling of the right upper lobe arteries. Four cases had various degrees of narrowing and rigidity of the main pulmonary artery; six cases showed stenosis of segmental arteries. The review provided no further clinical information about these patients.

Lupi-Herrera et al. (8) found 11 cases with pulmonary artery involvement among 22 patients with Takayasu's arteritis. These patients were all asymptomatic with respect to their pulmonary vasculitis. Although two patients had an increased pulmonary sound and four patients had a pulmonary murmur, no patient had signs of right heart failure or cyanosis. One patient had severe pulmonary hypertension with a pressure of 136 mm Hg in the right, left and main pulmonary arteries. Four other patients had pressures between 50 and 60 mm Hg in the right, left and main pulmonary arteries. On angiography, four patients had lesions of vascular wall irregularity of stenosis in the right, left or main pulmonary artery, but significant pressure gradients were not reported. Severe obstruction or occlusion of vessels was evident only among lobar, segmental or subsegmental arteries. Finally, Deutsch (12) reported four cases of occlusions or stenoses of segmental pulmonary arteries among 21 patients with Takayasu's arteritis. All of these patients were asymptomatic.

In addition, there have been several individual case reports of patients with Takayasu's pulmonary arteritis. Two of these are notable because the patients had documented pulmonary hypertension. Ishihama et al. (13) reported on a patient with a systolic pressure of 68 mm Hg in the main pulmonary artery. This patient's angiogram showed mild stenosis of the right pulmonary artery (no gradient was given) and complete obstruction of several segmental arteries. In addition, this patient had elevated left atrial pressures and left heart failure. Singh and Tan (14) reported on a patient with a systolic pressure of 90 mm Hg in the main pulmonary artery and complete occlusion of the right lower lobe artery with aneurysmal dilation of the right upper lobe vessels. The left pulmonary system had occlusion of several smaller arteries. This patient had recurrent hemoptysis and died with pulmonary hemorrhage and pneumonia.

*Our patient is unique among reported cases.* Her clinical presentation was dominated by dyspnea, cyanosis and right heart failure. The origin of these problems was obstruction to pulmonary blood flow caused by proximal pulmonary artery stenosis. There was preservation of smaller distal pulmonary arteries.

**Surgical treatment.** Specific treatment for pulmonary involvement from Takayasu's arteritis has not been reported. We can only speculate that in other situations treatment may have been unnecessary or unsuccessful. Furthermore, we are not aware of any reports describing surgical approaches to these lesions.

Because the patient's pulmonary vascular bed distal to these obstructions was relatively spared, these lesions were surgically approachable. Surgery was based on the techniques developed to treat systemic arterial involvement with Takayasu's arteritis (15–17). Endarterectomy was not acceptable because of the known high incidence of reocclusion. Resection of the diseased segments with graft replacement was not employed because of difficulty in approaching the right pulmonary lesion, together with the risk of misjudging the extent of the affected zone. The preferred surgical technique for discrete stenotic lesions has been patch angioplasty, and this was performed on our patient's proximal left pulmonary artery. The technique of choice for longer stenotic or occluded segments has been placement of a bypass graft. This was performed on the patient's right pulmonary artery (Fig. 5). Immediate hemodynamic improvement was confirmed by directed pressure measurements after the repair was completed. Immediate and 5 month follow-up examinations revealed marked improvement in exercise tolerance and no further evidence of right heart failure. We believe that other cases of pulmonary Takayasu's arteritis with right heart failure secondary to proximal pulmonary artery involvement should be considered for surgical palliation.

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