

# Massive Pulmonary Valve Insufficiency Associated with Double Pulmonary Artery and Ascending Aorta Aneurysm

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A case is reported of aneurysm of both the ascending aorta and pulmonary artery, associated with massive pulmonary valve insufficiency. Pulmonary artery aneurysm is a rare condition of unknown natural history; therapeutic management has not yet been established. Pulmonary valve insufficiency is also rare,

The case is reported of aneurysm of both the ascending aorta and pulmonary artery, associated with massive pulmonary valve insufficiency. Pulmonary artery aneurysm is a rare condition, the natural history of which is not well known. Neither has the therapeutic management of the condition yet been established. Pulmonary valve insufficiency is also a very rare condition, with reported etiologies consisting mainly of pulmonary valve anomalies. A comparative review of the literature relating to the diagnosis and therapeutic management of this condition is also provided.

## Case report

A 37-year-old male patient who underwent patent ductus arteriosus (PDA) ligation at the age of seven years also developed a pulmonary valve insufficiency (PVI), though this was fortuitously discovered at the age of 25 years. Ten years later, the patient presented with exertional dyspnea and was treated with digitalis and diuretics. An increasing dyspnea at moderate effort led to a complete cardiovascular check-up.

Cardiac examination revealed a severe systolodiastolic murmur at the pulmonary artery site. No sign of heart failure was observed, and the electrocardiogram demonstrated sinus rhythm. Transthoracic and transesophageal echocardiography revealed severe pulmonary disease associated with massive PVI. The

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mean transpulmonary gradient was 5.5 mmHg, with a peak of 10 mmHg. The right ventricle was significantly dilated (telediastolic diameter 43 mm). Both the pulmonary artery and ascending aorta had estimated 50-mm diameter dilatations. The aorta was normal at the level of the sinus of Valsalva, and the aortic and tricuspid valves were both normal. The left ventricular ejection fraction was 70%, and the mean pulmonary artery pressure 25 mmHg. Angiography demonstrated the presence of pulmonary artery and ascending aorta dilatation. Thoracic computed tomography (CT) scanning confirmed the echocardiographic findings (Fig. 1). There was no evidence of any systemic or inflammatory disease.

The decision was taken to perform surgery due to

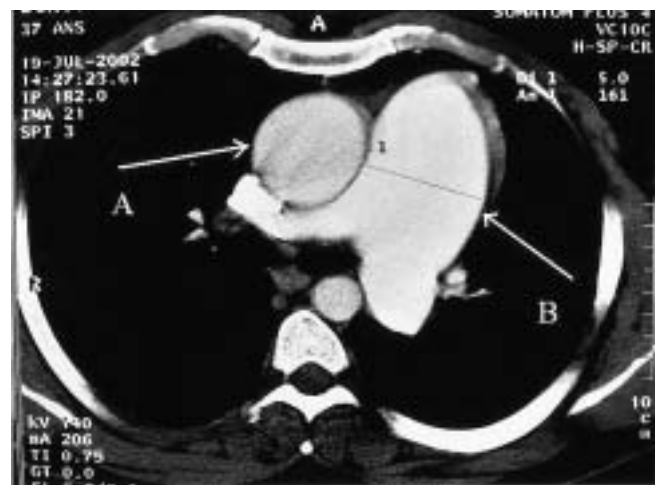


Figure 1: Preoperative computed tomography scan. A) Ascending aorta dilatation; B) pulmonary artery aneurysm.

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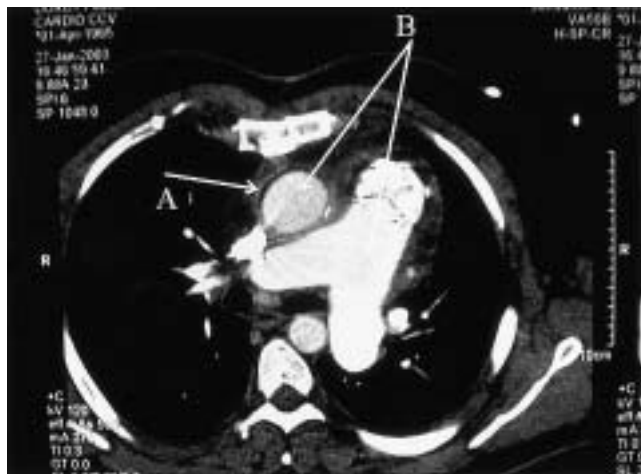


Figure 2: Postoperative computed tomography scan at the level of the pulmonary valve. A) Ascending aorta wrapping; B) Aorta and pulmonary artery replaced.

increasing clinical signs and their consequences on the right cavities, together with the presence of a double aneurysm dilatation. Following median sternotomy and opening of the pericardium, dilatation of the vessels was observed; the pulmonary artery aneurysm extended beyond the bifurcation. Cardiopulmonary bypass with moderate hypothermia was performed, using intermittent cold blood cardioplegia for myocardial protection. Subsequently, pulmonary valve replacement, pulmonary artery aneurysmorrhaphy and ascending aorta wrapping were carried out. The pulmonary artery aneurysmorrhaphy was performed by reducing its size through an orange segment-shaped resection. The pulmonary valve was bicuspid, there was posterior valve aplasia, and both other valves were prolapsed and thickened. The pulmonary valve was replaced using a Carpentier Edwards 29 bioprosthesis (Edwards Lifesciences, Irvine, CA, USA). The aortic dilatation extended proximal to the brachiocephalic artery, and the decision was taken to perform a prosthetic wrapping of the dilated segment.

The postoperative follow up was uneventful, and the patient's functional symptoms clearly improved. Postoperative echocardiography showed good functioning of the pulmonary prosthesis, without leakage. A decrease in right cavity dilatation was also noted.

Postoperative thoracic CT scanning demonstrated good outcome of the pulmonary artery and ascending aorta plasty (35 and 30 mm diameter, respectively) (Fig. 2) A pathological analysis of the pulmonary artery wall revealed slight wall thinning. The arterial intima did not show any structural anomalies, and there was no element in favor of any dystrophic disease.

## Discussion

Pulmonary artery aneurysm is a rare condition, with prevalence at autopsy having been reported as one in 13,695 cases (1). Congenital cardiac diseases represent the most frequent etiology (about half of the cases) of this condition, and include those with left-to-right shunt causing pulmonary artery hypertension. The first etiologies to be described are those of ventricular septal defect, atrial septal defect and PDA (2), though others have included infectious disease (syphilis, tuberculosis, bacterial endocarditis), traumatism and angitis. Certain dystrophic etiologies also generate pulmonary artery aneurysm; histological anomalies include fragmentation and reduction of media elastic fibers, a decrease in smooth muscle fibers, and an increase in collagen fibers. Cases of idiopathic pulmonary artery aneurysm have also been reported (3).

In the present patient, the congenital cardiac disease, although treated previously (by ligation of the PDA at the age of seven years), most likely contributed to the pulmonary dilatation. Histological analysis of the pulmonary artery wall did not reveal any structural anomalies, and there was no evidence of any Marfan-type dystrophic pathology or angitis.

Pulmonary valve insufficiency is also a rare condition, and in most cases it is associated with pulmonary valve anomaly (bicuspid or quadricuspid valve) or congenital cardiac disease. Pulmonary artery aneurysm represents one etiology of PVI, the mechanism consisting of a dilated pulmonary annulus (4). The association of pulmonary artery dilatation and aortic dilatation is very rare, and has been described by Dennison et al. (5) in cases of giant cell arteritis and Marfan disease. To the present authors' knowledge, no previous idiopathic case has been described.

Diagnosis is conducted using transthoracic and transesophageal echocardiography, catheterization with angiography, CT scanning and magnetic resonance imaging. The therapeutic strategy in case of pulmonary artery aneurysm is controversial, with some authors recommending a conservative approach in the absence of any significant shunt or pulmonary artery hypertension (6). Gradual dilatation and a risk of rupture, although rarely reported, have encouraged others to be more aggressive, with surgical repair performed in patients at low surgical risk (6). Surgical treatment has included prosthetic replacement or aneurysmorrhaphy, though the latter approach is performed more often due to its simplicity and short surgical time (1,6,7).

Surgical treatment is rare in cases of isolated PVI (2) and, when associated with pulmonary artery aneurysm, its treatment is indicated to prevent further dilatation. Treatment consists of valvular replacement

with a homograft or a bioprosthesis. In the present patient massive PVI was generated by valvular bicuspidy and pulmonary artery aneurysmal dilatation that severely affected the right-heart cavities, leading to clinical symptoms.

With regard to the ascending aorta dilatation, wrapping or external grafting is an interesting alternative to replacement when the sinuses of Valsalva are not dilated. Described initially by Robicsek et al. in 1971 (8), this technique is both simple and safe. In the present case, surgical treatment was motivated by massive symptomatic PVI, and limited access to a valve homograft led to a bioprosthesis being used to replace the pulmonary valve. Aneurysmorrhaphy of the pulmonary artery trunk was conducted for reasons previously described. Aortic wrapping is carried out by the authors when anatomic conditions permit, and both short- and mid-term results have been satisfactory.

#### References

1. Agarwal S, Chowdhury U, Saxena, Ray R, Sharma S, Airan B. Isolated idiopathic pulmonary artery aneurysm. *Asian Cardiovasc Thorac Ann* 2002;10:167-169
2. Metras D, Ouattrra K, Quezzin-Coulibaly A. Aneurysm of the pulmonary artery with cystic media necrosis and massive valvular insufficiency. Report of two successful surgical cases. *Eur J Cardiothorac Surg* 1987;1:119-124
3. Bartter T, Irwin RS, Nash G. Aneurysms of the pulmonary arteries - a review. *Chest* 1988;94:1065-1075
4. Holmes JC, Fowler NO, Kaplan S. Pulmonary valvular insufficiency. *Am J Med* 1968;44:851-862
5. Dennison AR, Watkins RM, Gunning AJ. Simultaneous aortic and pulmonary artery aneurysms due to giant cell arteritis. *Thorax* 1985;40:156-157
6. Kuwaki K, Morishita K, Sato H, Urita R, Abe T. Surgical repair of the pulmonary trunk aneurysm. *Eur J Cardiothorac Surg* 2000;18:535-539
7. Nair KK, Cobanoglu AM. Idiopathic main pulmonary artery aneurysm. *Ann Thorac Surg* 2001;71:1688-1690
8. Robicsek F, Daugherty HK, Mullen DC. External grafting of aortic aneurysms. *J Thorac Cardiovasc Surg* 1976;17:195-201