A 70-year-old woman was referred to cardiac evaluation because of the detection of dilation of the main pulmonary artery by chest computed tomography performed before the removal of a thoracic lipoma. The computed tomography scan with contrast also excluded arterial thromboembolism of the main and distal pulmonary branches.

The patient had a previous diagnosis of mild pulmonary stenosis with a right ventricle-pulmonary artery peak systolic pressure gradient of 24 mm Hg by right catheterization in 1978. Since then, the patient has been asymptomatic, and she has been conducting normal daily-life activities without undergoing further cardiovascular controls. In 2001, a previous computed tomography scan had already shown mild dilatation of the main pulmonary artery for which no investigation was later performed.

On the first evaluation, the patient was asymptomatic, her heart rate was 83 bpm, and her blood pressure was 130/70 mm Hg. Cardiac examination revealed a regular rhythm, with a 3/6 systolic murmur at the upper right and left sternal border. Lung fields were normal; neither jugular venous distention nor hepatomegaly and peripheral edema were detected. The ECG showed sinus rhythm and normal atrioventricular and intraventricular conduction, with slight ST-segment abnormalities in inferior leads.

Transthoracic echocardiography (Figure 1 and Movie I in the online-only Data Supplement) confirmed the presence of significant dilation of the main pulmonary artery (50 mm) involving the proximal segment of the left pulmonary artery. The pulmonary valve appeared dysplastic associated with an increase in transvalvular pulmonary flow velocity (derived mean systolic pressure gradient, 14 mm Hg; peak systolic pressure gradient, 22 mm Hg; and indexed valve area, 1.14 cm² calculated by continuity equation), suggesting a mild to moderate stenosis in the absence of significant regurgitation. The morphology of the pulmonary valve was not completely defined by transthoracic approach, but the systodiastolic doming of the thickened valve cusps was consistent with uni- or bicuspid valve.

Right ventricle morphology and systolic function were normal, and the prolapse of tricuspid valve with a mild regurgitation was detected. Left ventricular geometry and function were normal, and so was the morphology of the mitral and aortic valve which was trileaflet.

Cardiac and angiographic magnetic resonance were performed to clarify the morphology of the pulmonary valve and to study the extent of pulmonary artery dilatation. Cardiac MRI confirmed and magnified the main findings detected by the echocardiography (Figure 2 and Movie II in the online-only Data Supplement). The morphology of the pulmonary valve was bicuspid, tunnel-shaped, with anterior and posterior commissures, the semilunar cusp on the left side was larger than on the right side, and a mild restricted opening of the valve was shown (Figure 3 and Movie III in the online-only Data Supplement).

Contrast-enhanced cardiac magnetic resonance angiography showed significant saccular dilatation of the main pulmonary artery (48×62 mm) with involvement of the proximal left pulmonary artery, whereas the right pulmonary artery was preserved. In addition, the slight dilatation of the ascending aorta (36 mm, 24.6 mm/m²; normal value <21 mm/m²) and a more marked dilatation of innominate artery were detected (Figure 2 and Movie IV in the online-only Data Supplement).

Isolated bicuspid pulmonary valve without other congenital abnormalities is an extremely rare abnormality (≈0.1% incidence), unlike the more common bicuspid aortic valve (BAV). Since the description made by McKusick in 1972, the correlation between BAV and abnormalities of the ascending thoracic aorta has been largely described. Frequent occurrence of aortic involvement, regardless of valvular function but due to aortic media abnormalities of the ascending thoracic aorta, currently leads to the consideration of BAV as a disease involving both the aortic valve and the aortic wall, with a shared underlying developmental defect. Moreover, main pulmonary artery dilatation occurs in association with
BAV in the absence of pulmonary valve abnormalities, suggesting a common intrinsic disorder of the arterial wall connective tissue; thus, the use of the Ross operation is not recommended in patients with BAV disease. Nevertheless, mitral valve prolapse is frequently combined with BAV and aortic abnormalities in patients with Marfan syndrome and other connective diseases.

In the present case, all the same reported findings are present, but with an uncommon reversed location. The isolated bicuspid pulmonary valve is associated with progressive significant dilation of the ipsilateral arterial branch, the main pulmonary artery with the asymmetrical dilation of the left pulmonary artery rather than the right pulmonary artery, which cannot be explained by the hemodynamic mechanism of poststenotic expansion in the presence of a stable (at least since 1978) mild degree of the valve stenosis. In addition, the presence of tricuspid valve prolapse that is accompanied by other abnormalities of the right heart, and the involvement of the aortic segments, as well, complete the complex spectrum of widespread cardiovascular involvement that may be attributed to the connective tissue disorder.

Disclosures
None.

References

Figure 1. Transthoracic two-dimensional and color Doppler echocardiography. A, Upper parasternal short axis shows the dilatation of the MPA, and the dilatation of the origin of the LPA without the involvement of the right artery, as well. B, Parasternal short-axis view shows RV inflow/outflow with TV prolapse, dysplastic PV, and the dilatation of the main pulmonary artery with preserved size of the pulmonary valve annulus. Tricuspid AV is also documented. C, Continuous Doppler across pulmonary valve with peak velocity of 2.35 m/s. D, Double eccentric jets of mild tricuspid regurgitation due to valve prolapse. MPA indicates main pulmonary artery; LPA, left pulmonary artery; RV, right ventricle; TV, tricuspid valve; AV, aortic valve.
Figure 2. Cardiac MRI, fast cine steady-state free precession images. A and B, Saccular dilatation of the main pulmonary artery with involvement of the proximal left pulmonary artery, whereas the dimension of the right pulmonary artery was preserved. C, Right ventricular inflow/outflow view shows systolic doming of the pulmonary valve and the prolapse of the tricuspid valve (black arrow). D, 90 degrees left ventricular outflow tract view shows the slight dilatation of the ascending aorta and a more marked and localized dilatation of innominate artery (marked with the asterisk).

Figure 3. Cardiac MRI, fast cine steady-state free precession images. Short axis views (A, B) and right ventricular outflow tract (C, D) views showing the pulmonary valve in systolic (B, D) and diastolic (A, C) phase enhancing the bicuspid morphology with anterior and posterior commissures, the semilunar cusp on the left side was larger than on the right side and the mild narrowing of the valve.
An Inverted Location of the Bicuspid Valve Disease: A Variant of a Variant
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