

Congenital pulmonic stenosis in a 77-year-old woman successfully treated with percutaneous balloon valvuloplasty

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Congenital pulmonic stenosis (PS) rarely presents in patients over the age of 55 years. A stable asymptomatic course into late adulthood is the usual history of mild to moderate PS. Balloon valvuloplasty has become the procedure of choice for congenital PS, especially in children and young adults. There are rare reports of its use in older adults. Significant valve calcium is believed to limit its success. We present a case of a 77-year-old woman with symptomatic congenital PS and severe valve calcium who underwent successful balloon valvuloplasty.

CASE REPORT

A 77-year-old woman was referred for cardiac catheterization to evaluate exertional dyspnea of 3 months' duration. The dyspnea rapidly progressed to New York Heart Association class III limitation. Oral furosemide did not improve her symptoms.

She had systemic hypertension, which had been treated with carvedilol and losartan. She had three children, each vaginally delivered without complication, although she had been advised by a physician not to become pregnant after her first delivery. She never smoked, and there was no family history of heart or lung disease.

Oxygen saturation was 96% breathing room air. Her body mass index was 23 kg/m². Her neck veins were flat. There was a 3/6 systolic precordial murmur with a crescendo-decrescendo quality heard loudest at the left upper sternal border without radiation to the carotid arteries. There was no hepatosplenomegaly or subcutaneous edema. Electrocardiogram revealed right bundle branch block with tall biphasic R wave and right axis deviation consistent with right ventricular (RV) hypertrophy. Transthoracic echocardiogram findings included normal left ventricular ejection fraction, severe dilatation of the right atrium, and RV hypertrophy. No intraatrial communication was seen by color-flow Doppler. The pulmonic valve was not well visualized. Computed tomographic angiography of

the pulmonary vasculature revealed no pulmonary emboli. The pulmonary arteries distal to the pulmonic valve were dilated.

At cardiac catheterization, there was extreme difficulty advancing the wedge catheter across the RV outflow tract into the pulmonary trunk. The pulmonic valve was heavily calcified. The pulmonary arterial pressure was 23/16 (mean, 19); pulmonary capillary wedge mean, 4; right ventricle, 137/11; and peak-to-peak systolic pressure gradient from RV to pulmonary trunk, 114 mm Hg (*Figure 1*). A transesophageal echocardiogram showed the pulmonary trunk to be dilated (*Figure 2*). No infundibular or supra-ventricular stenosis was present. Color-flow Doppler showed highly turbulent flow through the stenotic pulmonic valve (*Figure 3*). The foramen ovale was patent.

Fluoroscopy confirmed a heavily calcified pulmonic valve (*Figure 4*). A right ventriculogram ruled out subvalvular outflow tract obstruction (*Figure 5*). An 18-mm balloon was successfully used for pulmonic valvuloplasty (*Figure 6*). After the procedure,

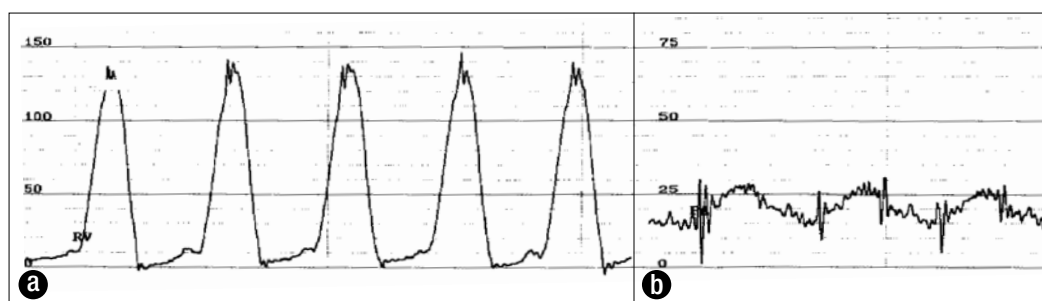


Figure 1. (a) Right ventricular pressure tracing and (b) pulmonary artery pressure tracing showing severe elevation in right ventricular pressure and a significant systolic gradient (114 mm Hg) between the right ventricle and pulmonary artery.

the RV pressure was 52/15 mm Hg, and the peak-to-peak gradient between the RV and pulmonary trunk was 25 mm Hg.

Two months after valvuloplasty, the dyspnea had vanished. Echocardiography confirmed the marked improvement in the pulmonic valve gradient after the procedure, as measured by continuous-wave Doppler (*Figure 7*).

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DISCUSSION

The etiology of pulmonic stenosis (PS) is usually congenital and rarely acquired. Our patient had no evidence of the carcinoid syndrome or rheumatic heart disease. To our knowledge, the oldest patient ever reported to undergo balloon valvuloplasty for congenital PS was 76 years old (1–5).

The stable asymptomatic course our patient had into such a late age is usual based on the natural history of mild to moderate congenital PS. The subsequent progressively symptomatic phase with a severe gradient is indeed rare. This could have largely been a consequence of advanced valve calcium, another feature not commonly seen in congenital PS (6). When calcium is seen in the pulmonic valve, there is severe valvular stenosis and with rare exception, age >30 years (7).

Congenital valvular lesions are often diagnosed during pregnancy, as possibly occurred in our patient. Hemodynamic changes can bring previously undiagnosed and asymptomatic or mildly symptomatic patients to a clinician's attention through incidental auscultation of a precordial murmur or workup of unexplained dyspnea and fatigue. Furthermore, this patient's uncomplicated pregnancies are typical of mild to moderate PS. Isolated PS is rarely a significant disorder in pregnancy and is associated with low maternal and fetal risk (8).

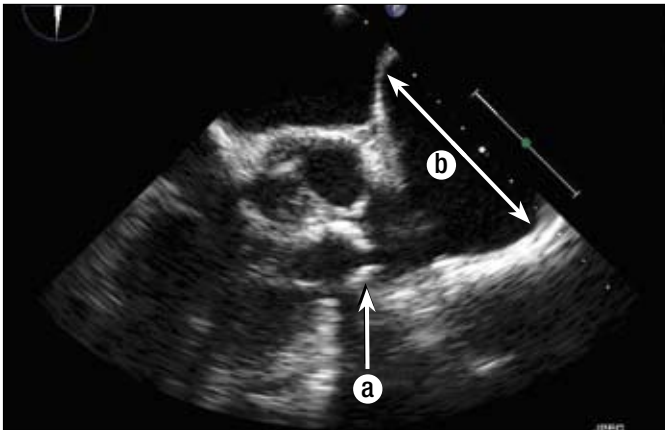


Figure 2. Transesophageal echocardiogram showing (a) a severely calcified and stenotic pulmonic valve and (b) a dramatic poststenotic pulmonary artery.

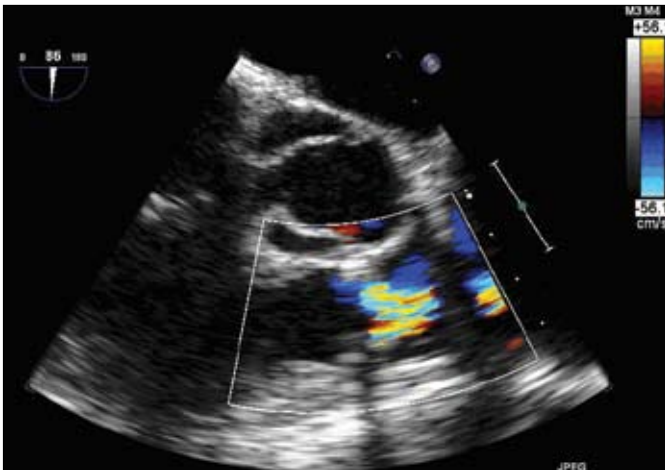


Figure 3. Color-flow Doppler across the pulmonic valve showing highly turbulent flow secondary to the stenotic orifice.

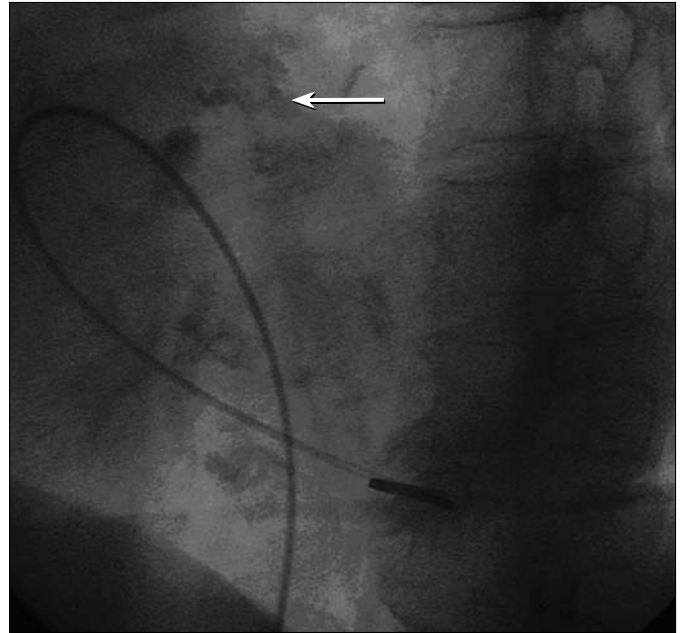


Figure 4. Fluoroscopy prior to right ventricular angiography showing heavy calcification of the pulmonic valve (arrow).

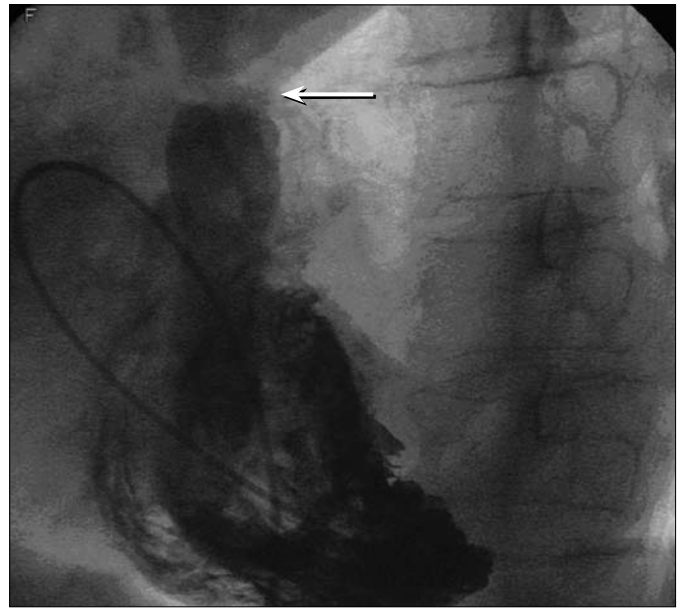


Figure 5. Right ventricular angiogram clearly showing pulmonic stenosis (arrow). Also note the absence of subvalvular hypertrophy or infundibular stenosis. Poststenotic pulmonary artery dilatation is clearly evident.

Our patient had no evidence of subvalvular RV outflow obstruction, which when isolated is a contraindication to balloon valvuloplasty. The lack of any subvalvular obstruction in our patient may in part have been due to being on carvedilol chronically. Al Kasab et al reported that the maximum post-procedure subvalvular RV outflow gradient did not exceed 15 mm Hg in patients with valvular PS on a beta-blocker (9). Also, Moulart et al used propranolol after surgical commissurotomy to determine if resection of infundibular hypertrophy was necessary (10). A decrease in RV pressure was noted with relaxation of the infundibulum. Whether beta-blockade significantly relieved possible infundibular spasm or prevented

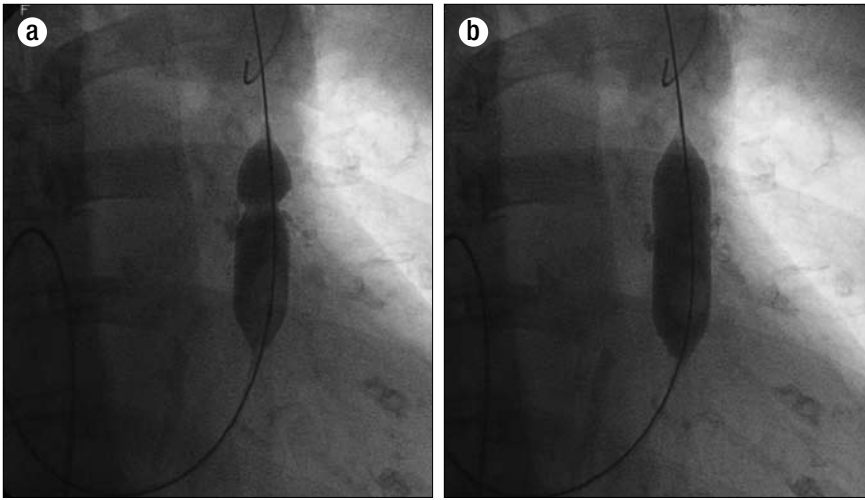


Figure 6. Dilation balloon (a) during inflation and (b) after valvuloplasty.

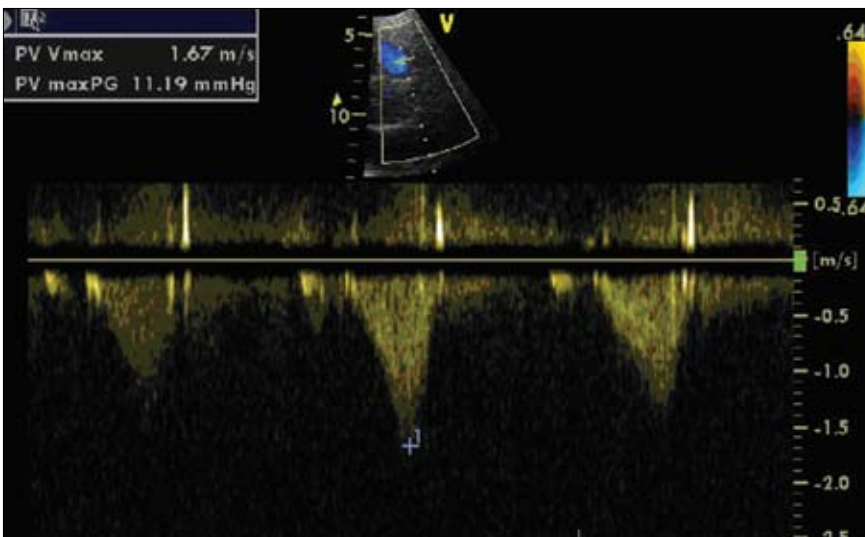


Figure 7. Continuous-wave Doppler flow through the pulmonic valve at 2-month postvalvuloplasty follow-up showing a significant reduction in gradient.

the development of subvalvular hypertrophy in our patient is plausible but uncertain.

Since its initial description by Kan et al (11) in 1982, percutaneous balloon valvuloplasty has been shown to be an effective treatment with durable results in children and adults with congenital PS (1–4, 12–14). None of their patients were older than 55 years. Patients with typical congenital PS who undergo balloon valvuloplasty have a low rate of restenosis. At follow-up of 10 years, results appear to be similar to those of surgical valvotomy (12). Hopefully, our patient, despite the heavy valve calcium, will have similar long-term results.

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