Percutaneous Balloon Dilation of the Pulmonary Valve in a 79-Year-Old With Congenital Pulmonary Valve Stenosis

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INTRODUCTION

Congenital pulmonary valve stenosis (PS) very rarely presents in the elderly population. Balloon valvuloplasty is used to treat valvar PS in children and adolescents, and limited reports exist describing its use in older adults. We describe a 79-year-old with congenital PS due to a unicommisural valve who underwent successful transcatheter valve dilation with an excellent hemodynamic and clinical response.

CASE PRESENTATION

A 79-year-old man presented with lower extremity edema and New York Heart Association class III heart failure symptoms and was diagnosed with severe valvar PS. The patient had not seen a physician in over 30 years. The physical exam showed symmetric 3+ pitting lower extremity edema. The cardiac exam showed normal precordial activity, an ejection click with s1, obscured splitting of s2, and a 3/6 medium frequency ejection murmur along the left sternal border. Transthoracic echocardiogram (TTE) showed mildly decreased right ventricular (RV) systolic function and a peak instantaneous gradient of 66 mm Hg across the pulmonary valve (PV) with thickened leaflets that dome in systole (Figures 1 and 2A, Video 1). Cardiac computed tomography (CCT) was performed using a 96-detector row dual-source system with image acquisition using a high-pitch prospective electrocardiogram-triggered systolic spiral technique. After the pre-contrast baseline scan demonstrated no PV calcifications, contrast was administered with timing optimized to visualize the right heart structures. This showed a unicommisural PV with thickened, noncalcified leaflets that domed in systole (Figure 3A-D). At catheterization, the patient had an RV pressure of 65/18 mm Hg and main pulmonary artery pressure of 22/18 mm Hg (Figure 4A and B). The PV annulus measured 20 mm, and balloon valvuloplasty was performed with a 22 mm compliant balloon (Figure 5A and B). Afterward, the RV pressure decreased to 36/14 mm Hg with a main pulmonary artery pressure of 34/16 mm Hg (Figure 4C and D). At 3 months' follow-up, the patient's lower extremity edema had resolved, exertional capacity had improved with New York Heart Association class II symptoms, and TTE showed mild pulmonic regurgitation and a peak instantaneous gradient of 20 mm Hg across the valve (Figure 2B).

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal basal short-axis view of the RV outflow tract with color-flow Doppler, demonstrates thickened PV leaflets with systolic doming, turbulent, accelerated flow pattern from stenosis, and mild pulmonic regurgitation.

View the video content online at www.cvcasejournal.com.

DISCUSSION

Congenital PS is a rare cause of right-sided heart failure in older adults. The normal PV is trileaflet with thin, mobile leaflets. In congenital cases of PS, the valve leaflets are typically fused and thickened.[1] In older adults, calcium deposition on the abnormal PV leaflets may further compromise leaflet mobility.[2] Typically, if mild to moderate PS is present in childhood and adolescence, it will not progress during adulthood. However, progression of severity can occur especially if calcium deposition occurs later in life.[2] Interestingly, our patient had a unicommisural PV without significant calcium deposition. Rare, acquired causes of PS exist, including carcinoid heart disease. However, we feel that our patient's multimodality imaging strongly supports the diagnosis of congenital PS given the unicommissural morphology of the valve.

The patient had not seen a physician in many years, but based on the clinical course, the degree of PS likely worsened over the past few years. Our patient presented with severe lower extremity edema and exertional fatigue. We hypothesize that the RV diastolic function gradually worsened and RV end-diastolic pressure increased above a critical point that led to severe systemic venous hypertension and lower extremity edema. Interestingly, the RV end-diastolic pressure decreased by 4 mm Hg immediately after dilation, and the patient's edema significantly improved following intervention.

Echocardiographic imaging is critical in making the diagnosis of congenital PS. The leaflet anatomy and annulus size can be characterized by TTE, and the amount of PV obstruction can also be estimated. Cardiac computed tomography is an emerging tool and can complement TTE, especially with characterization of PV anatomy. Our patient had a CCT that confirmed the unicommisural anatomy and lack of calcium. We felt that transcatheter balloon dilation would have a higher chance of success in the absence of a calcified valve. However, the degree of valve calcification has not been shown to

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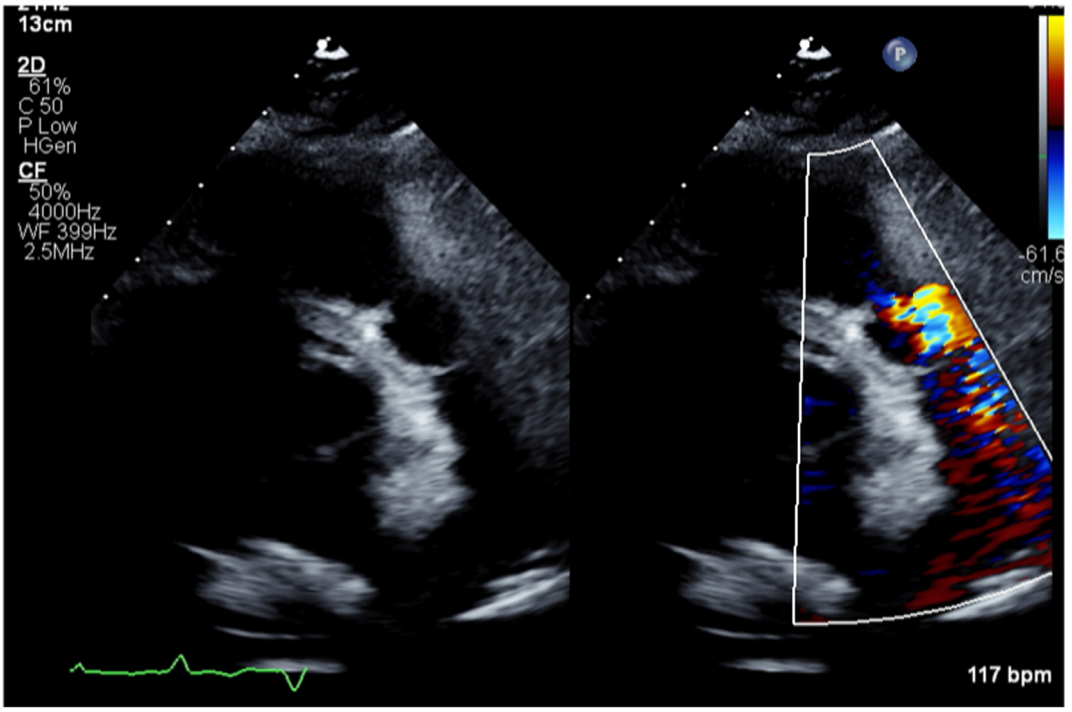

Figure 1 Two-dimensional TTE, parasternal basal short-axis view of the RV outflow tract, demonstrates the patient's thickened PV leaflets that dome in systole (*left*) and with color-flow Doppler demonstrates the turbulent, accelerated flow pattern from stenosis (*right*).

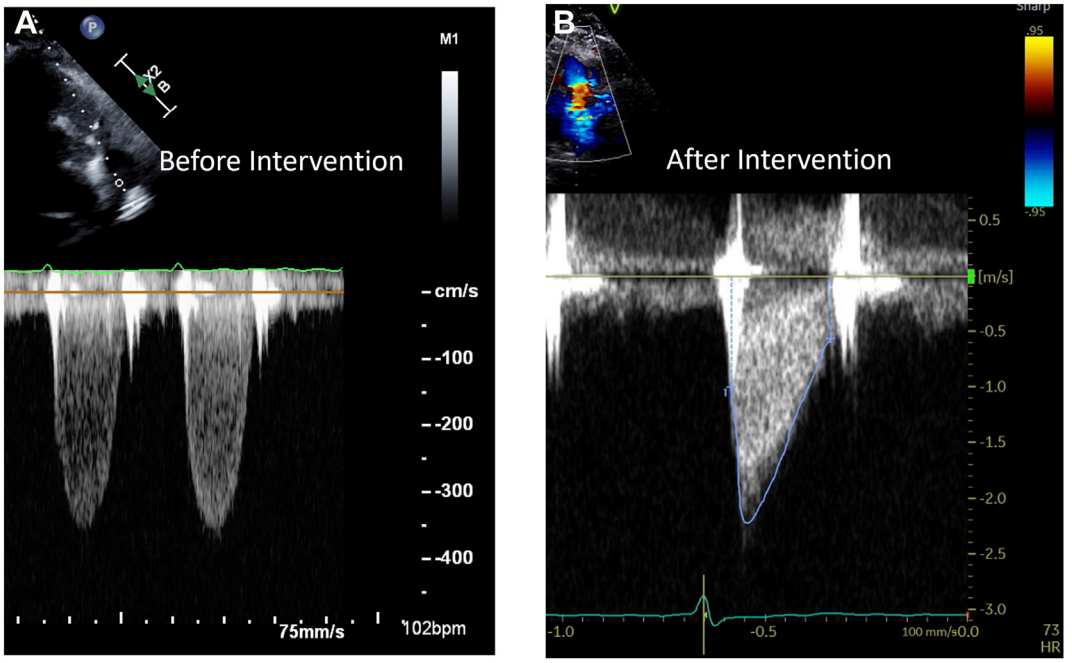

Figure 2 Two-dimensional TTE, parasternal basal short-axis view of the RV outflow tract, continuous-wave Doppler interrogation across the PV before **(A)** and 3 months after **(B)** transcatheter PV dilation, demonstrates the marked improvement in peak instantaneous gradient 3 months after intervention.

predict the effectiveness of transcatheter valvuloplasty, and valvuloplasty may still be successful in those with a congenital PS and calcified leaflets. The annulus was also clearly seen by CCT and was normal in size. The size of the valvuloplasty balloon is based on the annulus size, and the CCT provides very crisp, accurate images of the valve annulus, which assists with preprocedural planning. Furthermore,

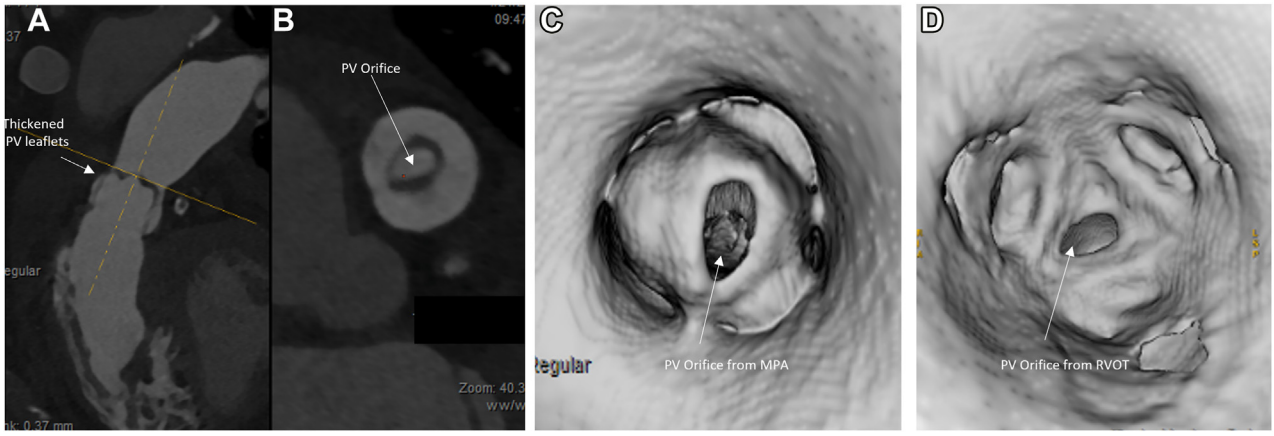

Figure 3 Cardiac computed tomography, multiplanar reconstruction images, oblique sagittal (long-axis) **(A)** and en face (short-axis) **(B)** displays, confirmed the congenitally malformed PV. Reformatted, volume-rendered, three-dimensional endoluminal displays of the PV from the perspective of the main pulmonary artery (MPA) **(C)** and from the RV outflow tract **(D)** demonstrate the unicuspid morphology with restricted, doming systolic excursion.

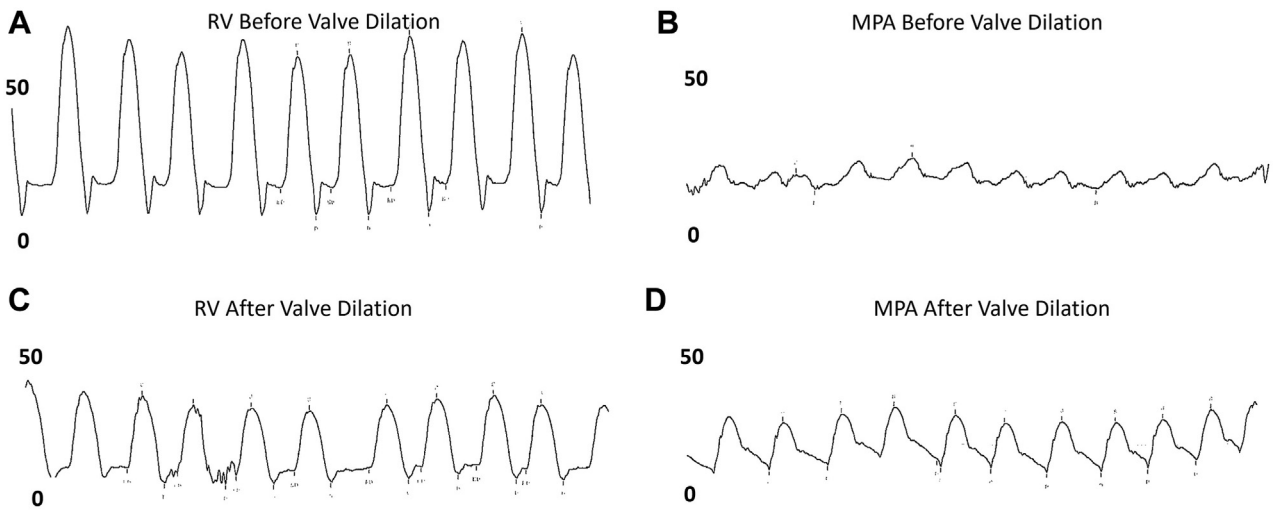

Figure 4 Hemodynamic tracings prior to PV dilation demonstrate an RV pressure of 65/18 mm Hg **(A)** and main pulmonary artery (MPA) pressure of 22/18 mm Hg **(B)**. Hemodynamic tracings after PV dilation demonstrate an RV pressure of 36/14 mm Hg **(C)** and MPA pressure of 34/16 mm Hg **(D)**.

CCT can clearly show the branch pulmonary artery anatomy and, thus, help exclude any branch pulmonary artery stenosis as an additional cause of RV hypertension.[1] Of note, the three-dimensional CCT views (Figure 3C and D) were used for a complete depiction of the PV structure. These images were not used for quantification since windowing of different contrast and tissue densities can significantly impact measurements.

Percutaneous PV dilation was first described in 1982 and has become the standard of care for the treatment of most cases of congenital PS in children and adolescents.[3,4] Several case series exist that support its efficacy in the treatment of adults with congenital PS.[5,6] Only rare case reports exist describing the use of percutaneous PV dilation in adults >70 years.[2] Our patient was 79 years old and, thus, is suspected to be one of the oldest reported patients to have had transcatheter balloon dilation of the PV. In children and adolescents, a balloon-to-annulus ratio of 1.25 is generally used, but in adults, a ratio of 1.0 to 1.1 can be effective with minimal residual regurgitation.[4,6]

CONCLUSION

We describe a 79-year-old man who presented with symptoms of right-sided heart failure due to congenital PS who was successfully treated with percutaneous balloon dilation.

ETHICS STATEMENT

The authors declare that the work described has been carried out by The Code of Ethics of the World Medical Association (Declaration of

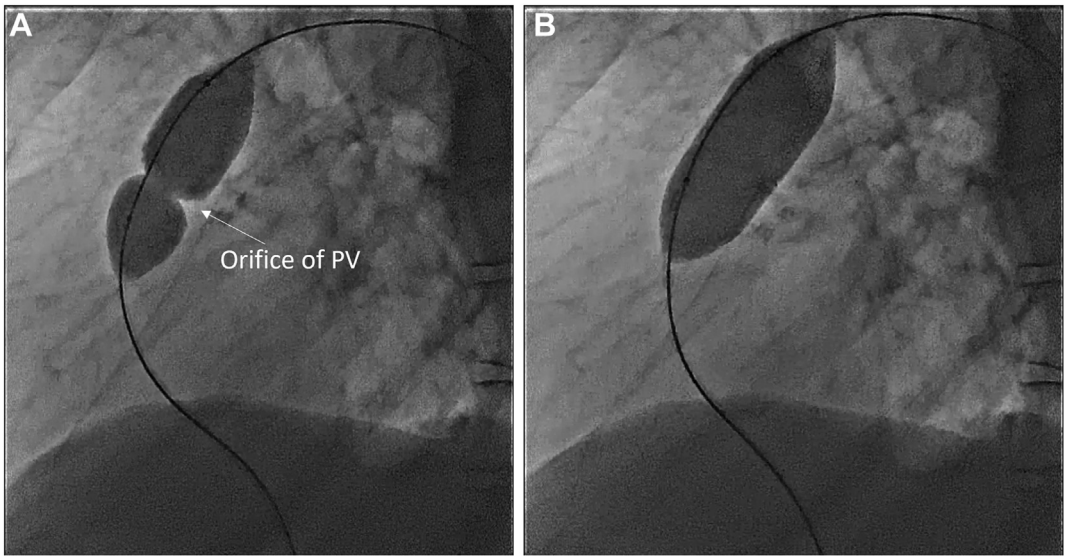

Figure 5 Lateral fluoroscopic projections of PV dilation with a 22 mm compliant balloon. During inflation of the balloon, a waist appears **(A)** at the level of the PV orifice and, at 1.5 atmospheres, the waist resolves **(B)**.

Helsinki) for experiments involving humans. Ethical approval was also obtained from the ethical review committee of the hospital.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.11.008.

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