# The Complementary Value of Proximal Isovelocity Surface Area in Large Left-to-Right Lesions With Pulmonary Vein Stenosis

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## INTRODUCTION

Pulmonary vein stenosis (PVS), a rare disease in the pediatric population, has higher mortality and morbidity despite medical and surgical interventions.[1] Many patients with PVS have associated congenital heart disease, and studies have shown that intrinsic baseline anatomy may contribute to the risk for postoperative PVS.[2] Congenital heart disease lesions with large left-to-right shunts by definition increase pulmonary blood flow, causing abnormal flow in the pulmonary veins.[3] Proximal isovelocity surface area (PISA), a flow convergence method, uses the principle of conservation of mass to estimate the size of valvular lesions and intracardiac shunts.[4]

Herein, we present a pediatric patient with multiple intracardiac lesions causing a large left-to-right shunt and concomitant PVS in whom PISA was used to calculate the orifice area of the narrowed pulmonary vein.

## CASE PRESENTATION

A preterm child born vaginally at 34 weeks of gestation had a history of tachypnea for the previous 3 months. The patient was evaluated elsewhere because of poor weight gain and tachypnea. At 5 months of age, the child underwent an echocardiographic examination, which revealed an intracardiac shunt, and was transferred to our hospital for surgery. At presentation, the child weighed 5 kg (<25th percentile) and had a height of 58 cm (<25th percentile). Transthoracic echocardiography showed right-sided heart enlargement with a 7-mm ostium secundum atrial septal defect, a 6-mm subaortic ventricular septal defect (VSD), an additional 4-mm midmuscular VSD, a 4-mm patent ductus arteriosus, and flow turbulence across the right upper pulmonary vein (RUPV). Imaging of the other pulmonary veins was not adequate with transthoracic echocardiography alone, and cardiac computed tomography (CCT) was performed, which showed

RUPV stenosis. The patient was referred for intracardiac repair and PVS repair.

Preoperative transthoracic echocardiographic findings were confirmed on intraoperative transesophageal echocardiography using a micro–transesophageal echocardiographic probe (Figure 1). Continuous-wave Doppler examination of the RUPV showed a continuous antegrade wave, with a peak velocity and gradient of 239 cm/sec and 23 mm Hg, respectively (Figure 2). Spectral Doppler examination of other pulmonary veins showed a normal flow pattern (Figure 3). Therefore, the decision was taken by the operating team to repair the RUPV stenosis. Color flow Doppler examination of the RUPV at a Nyquist limit of 38.5 cm/sec revealed color aliasing and formation of PISA at the entry point of the RUPV to the left atrium (Video 1). The junction of the RUPV and left atrium was zoomed in, and the upper baseline value of the Nyquist limit was adjusted to 28.9 cm/sec for acquisition of an optimal hemispherical PISA shell (Figure 4). The area of the RUPV at the opening of the left atrium derived from the PISA formula ([2 × $\pi$ × $r^2$] × [aliasing velocity/maximum velocity]) was 0.022 cm$^2$, with a PISA radius of 0.18 cm and pulmonary venous peak velocity of 239 cm/sec. Assuming the RUPV opening to be circular, its diameter estimated from the area obtained using the PISA method (circle area = 0.785 × diameter$^2$) was 0.18 cm (0.785 × 0.022). Angiographically, the estimated size of the pulmonary vein[5] in our patient was 0.60 cm (0.08 × height [cm] + 1.4), thereby confirming the RUPV stenosis. The diameter of the RUPV derived from CCT was 1.8 mm (Figure 5). Additionally, the operating surgeon failed to pass a 2-mm shunt but could pass a 1.5-mm shunt through the stenosed RUPV. Atrial septal defect and VSD closure was done with the pericardium, muscular VSD, and patent ductus arteriosus closed directly, and right pulmonary vein plasty was completed with bovine pericardium. Post–cardiopulmonary bypass examination of the heart revealed no residual intracardiac lesions and normal phasic laminar flow across the RUPV, with a peak gradient of 3 mm Hg (Figure 6). The patient had an uneventful postoperative period.

## DISCUSSION

Children with large left-to-right shunts will have increased flow across the pulmonary veins, thereby producing a characteristic continuous antegrade wave on pulmonary venous Doppler.[6] A continuous antegrade wave is also observed on pulmonary venous Doppler interrogation in the presence of pulmonary venous obstruction.[7] In our patient, both coexisted, making the diagnosis of PVS on transthoracic echocardiography very difficult. The symptomatology of patients with large left-to-right shunts often overlaps with that of patients with significant PVS. Our patient had no clear defining clinical feature suggestive of concomitant PVS. Although CCT revealed significant stenosis, the

final decision to address the PVS was decided after an intraoperative transesophageal echocardiographic assessment and direct visualization by the operating surgeon (Figure 7). Gadhinglajkar *et al.*[8] reported increased turbulence across the pulmonary veins due to a large left-to-right shunt. Color Doppler interrogation of the RUPV in the present patient showed turbulence, but it was not clear whether this turbulence was due to increased flow across the pulmonary veins due to a large left-to-right shunt or to pulmonary venous obstruction.

PISA, a flow convergence method, uses the principle of conservation of mass to estimate the area of an orifice. Its application has been confined to different intracardiac shunts and valvular lesions.[4] Although PISA was used for estimating the orifice area of the
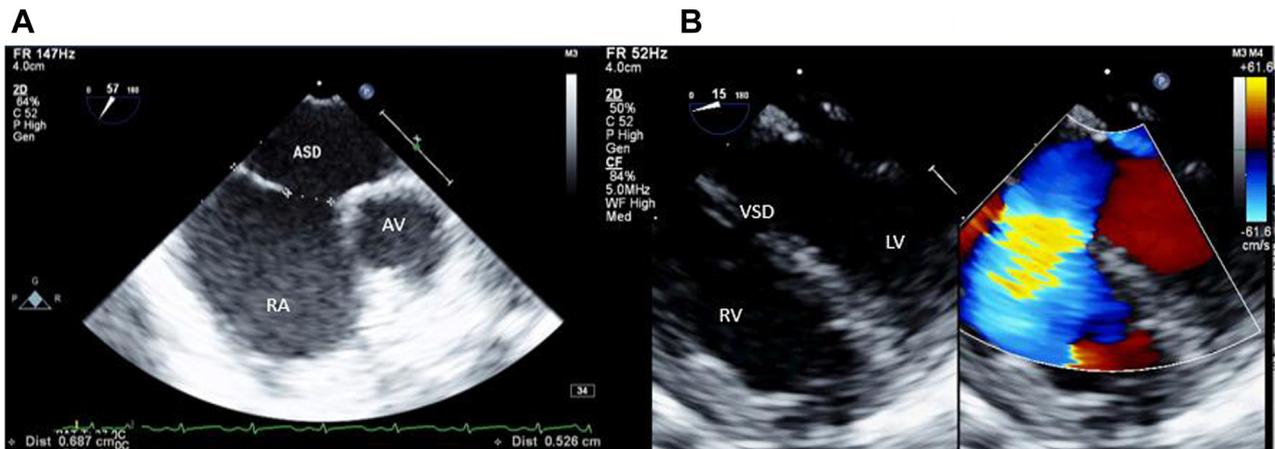
**Figure 1** Two-dimensional transesophageal echocardiography, midesophageal short-axis view (57°), demonstrating the atrial septal defect (ASD) with a deficient aortic superior rim **(A)** and the VSD in a long-axis four-chamber systolic view (**B**; 15°) without (*left*) and with (*right*) color flow Doppler. *AV*, Aortic valve; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.
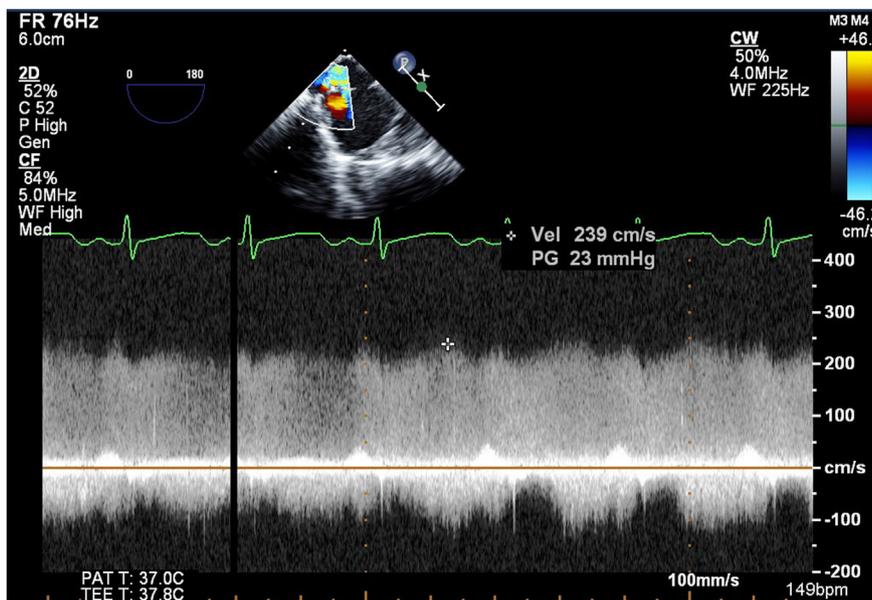


**Figure 2** Two-dimensional transesophageal echocardiography, midesophageal rightward-rotated short-axis view (0°) with color flow–guided, continuous-wave Doppler spectrum of the RUPV, demonstrating a continuous antegrade wave with a peak velocity of 239 cm/sec and a peak gradient of 23 mm Hg.
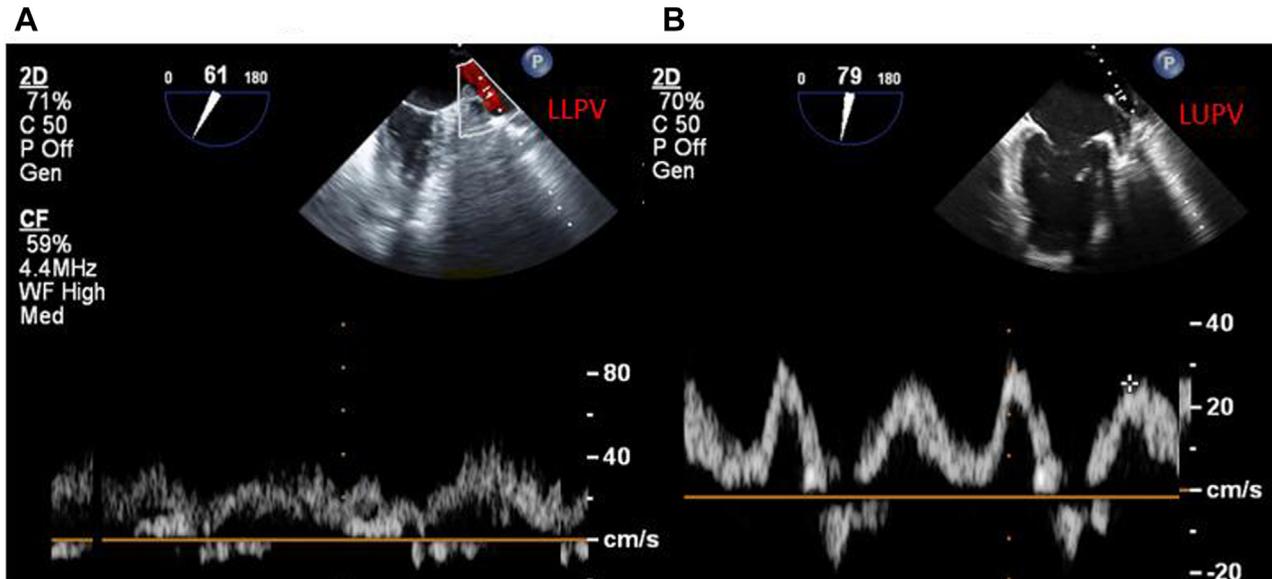
**Figure 3** Two-dimensional transesophageal echocardiography, midesophageal long-axis two-chamber views with color flow–guided, pulsed-wave Doppler examination of the **(A)** left lower pulmonary vein (LLPV; 61°) and **(B)** left upper pulmonary vein (LUPV; 79°). A normal phasic pulmonary venous blood flow pattern and velocities are demonstrated in both.
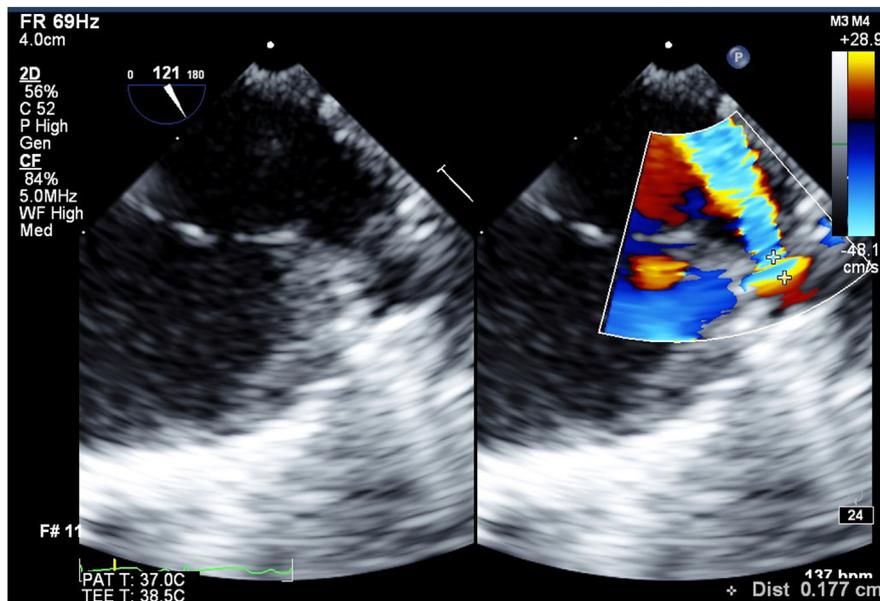


**Figure 4** Two-dimensional transesophageal echocardiography, midesophageal long-axis two-chamber view (121°), rightward rotated, without (*left*) and with (*right*) color flow Doppler, demonstrating the RUPV stenosis with a PISA radius of 0.177 cm at an aliasing velocity of 28.9 cm/sec.

pulmonary vein in a patient with concomitant PVS and atrioventricular septal defect,[9] our report further substantiates this method by correlating with the computed tomography–derived diameter of the RUPV opening. In addition, the operating surgeon could pass only a 1.5-mm shunt across the RUPV, confirming the PISA-derived values.

It is well known that the dimensions of cardiac chambers and vessels vary among the pediatric population depending on body surface area. Robida[5] described height-based formulas for comparing pulmonary vein diameters during angiographic studies. The calculated diameter of the RUPV in our patient was 0.60 cm on the basis of projected
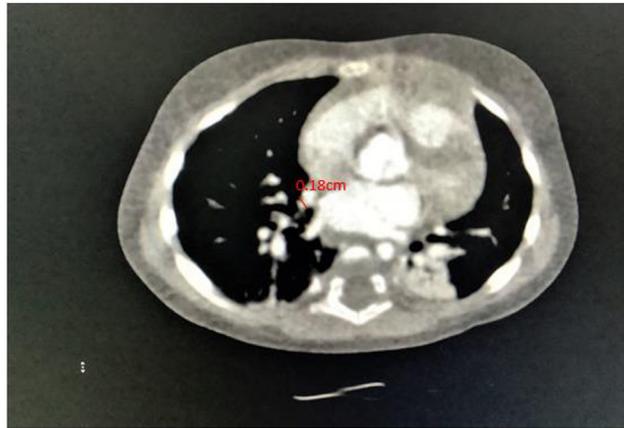
**Figure 5** Contrast-enhanced, low-resolution (neonatal) CCT, axial display, demonstrating the narrow RUPV measurement.
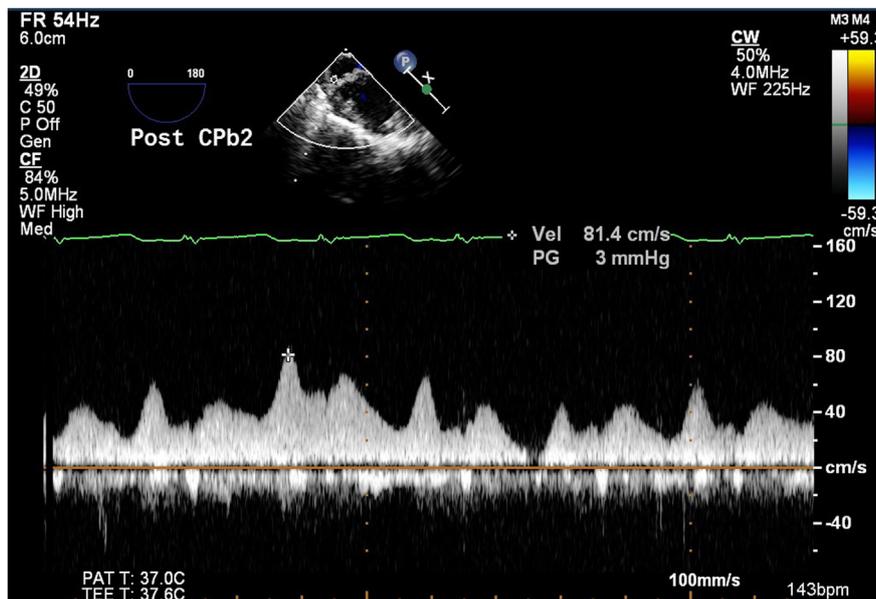


**Figure 6** Two-dimensional transesophageal echocardiography, midesophageal rightward-rotated short-axis view (0°) with color flow–guided, continuous-wave Doppler spectrum of the RUPV obtained postoperatively, demonstrating a normal phasic pulmonary venous flow pattern with a peak velocity and gradient of 81.4 cm/sec and 3.0 mm Hg, respectively.

angiographic nomograms, which was very high compared with the orifice area calculated using the PISA method, thus indicating the possibility of RUPV stenosis.

After cardiopulmonary bypass, the characteristic continuous antegrade wave in pulmonary venous Doppler reversed, with well-differentiated systolic, diastolic, and atrial contraction waves. With residual PVS, normalization of the pulmonary venous Doppler waveform may not take place, consequently increasing the possibility of an additional cardiopulmonary bypass run to address the stenosis. In the present case, surgical repair of both intracardiac shunts and PVS resulted in decreased peak pressure gradients across the RUPV to 3 mm Hg.

Aliasing observed on color Doppler examination can be an important clue in the diagnosis of PVS. In children with large left-to-right shunts, any turbulence across the pulmonary veins is often attributed to increased pulmonary blood flow and is often overlooked.

Additionally, CCT is not a routine investigation in patients referred for repair of atrial septal defects and VSDs. Hence, we recommend applying the PISA method for the calculation of pulmonary venous diameters in patients with large left-to-right shunts, especially if associated with color aliasing. Although the application of PISA is not a validated method for calculating pulmonary venous diameters, incorporating it into pulmonary venous examination protocols in patients with large left-to-right shunts with suspected PVS may be considered.

## CONCLUSION

PISA is a reliable semiquantitative method to determine the presence of PVS in children with large left-to-right shunts.
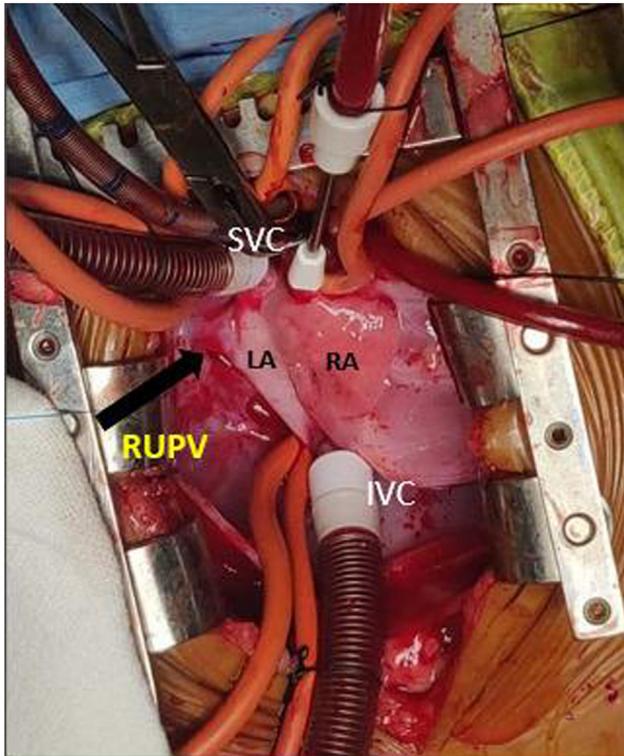
**Figure 7** Intraoperative image obtained during surgical repair demonstrates the narrowed RUPV (*black arrow*). *IVC*, Inferior vena cava; *LA*, left atrium; *RA*, right atrium; *SVC*, superior vena cava.

## ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with the following guidelines: This article adheres to the applicable EQUATOR guidelines.

## CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

## DISCLOSURE STATEMENT

The authors report no conflict of interest.

## SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.06.004.

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