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INTERMEDIATE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Perventricular Transcatheter Pulmonary Valve Implantation in a Symptomatic 3-Year-Old Child With Repaired Tetralogy of Fallot

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ABSTRACT

In select patients, transcatheter pulmonary valve replacement through a percutaneous approach can be challenging because of complicated anatomy or small patient size. In these patients, especially those weighing <20 kg, hybrid perventricular valve delivery may provide a preferred alternative approach. (**Level of Difficulty: Intermediate.**)
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HISTORY OF PRESENTATION

A 3-year-old syndromic boy with tetralogy of Fallot and severe branch pulmonary artery (PA) hypoplasia underwent right ventricular (RV) outflow tract (RVOT) stenting in the neonatal period and complete transannular patch repair at 8 months with fenestrated closure of an atrial septal defect. As a result of his worsening severe tricuspid regurgitation (TR), free pulmonary regurgitation (PR), and failure to thrive, he underwent early surgical placement of a 14-mm pulmonary homograft and tricuspid valve repair at 2.5 years of age. He experienced transient improvement in RV size, function, and clinical status, but the conduit quickly became incompetent, with worsening RV enlargement and TR. Clinically, he was dependent on high doses of diuretic agents, and he required several admissions for acute kidney injury secondary to low cardiac output. Given his worsening clinical status and the desire to avoid another bypass operation, he was referred for attempted transcatheter pulmonary valve replacement (TPVR) at a weight of 13 kg to establish pulmonary valve competency.

INVESTIGATIONS

Before TPVR, an echocardiogram showed severe PR and TR, a severely dilated right atrium and ventricle with moderate hypertrophy, and severely diminished RV systolic function (**Figures 1A and 1B**). Cardiac magnetic resonance demonstrated a PR fraction of 57% and a TR fraction of 69%. The RV end-diastolic volume measured 301 ml/m^2, with an RV ejection fraction of 24% (**Figure 1C**).

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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MANAGEMENT

TPVR was initially attempted through the right internal jugular vein, but it was unsuccessful because of an inability to advance the bifurcating flowering stent into the branch PAs (Figure 1D). The patient was brought back for hybrid perventricular intervention through a subxiphoid incision and sheath placement through the RV free wall. Following a negative result of a coronary compression test, a 3.4-cm covered CP stent (B. Braun Medical, Inc., Bethlehem, Pennsylvania) mounted onto a 14 mm × 4 cm balloon was implanted into the distal conduit, followed by a 26-mm Mega LD stent (Medtronic, Inc., Minneapolis, Minnesota) on 2 bifurcating

ABBREVIATIONS AND ACRONYMS

PA = pulmonary artery

PR = pulmonary regurgitation

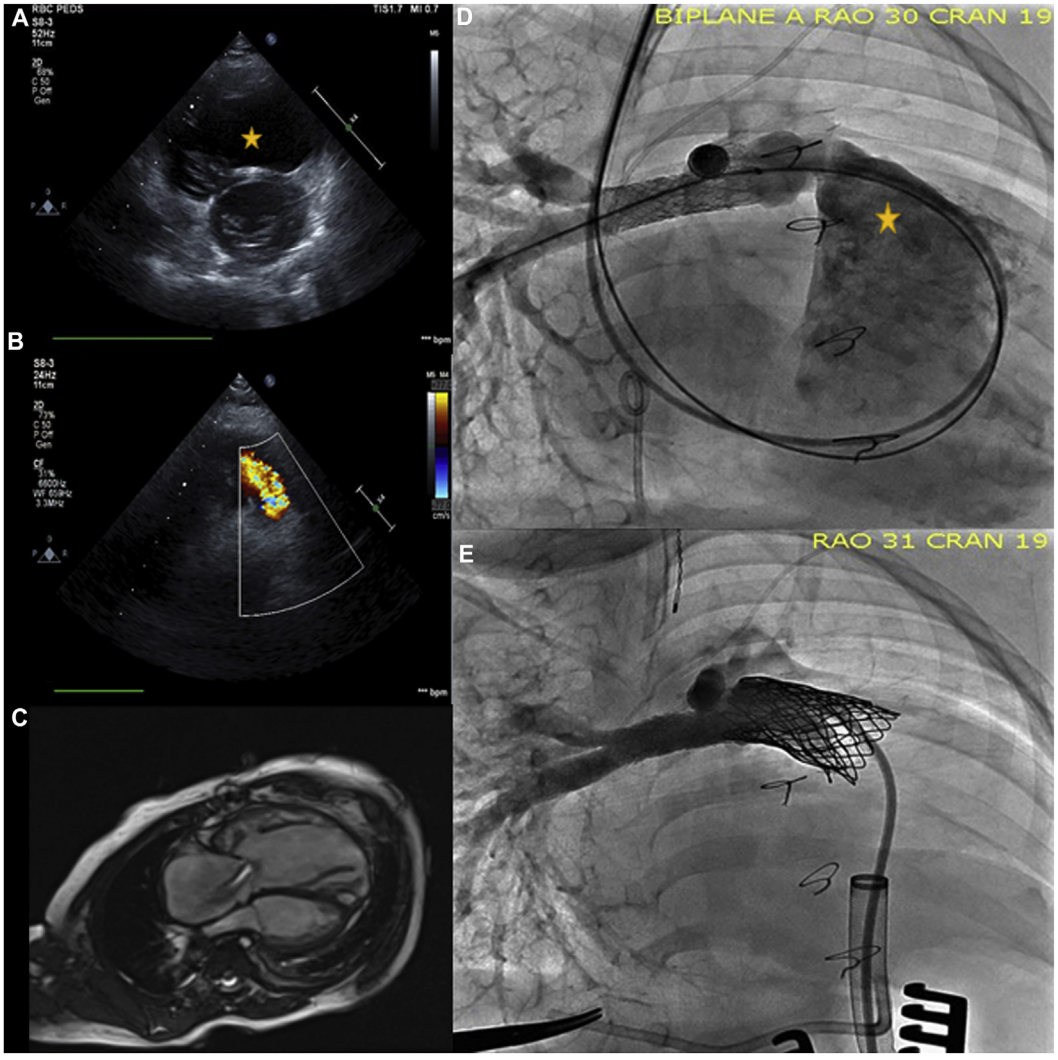
RV = right ventricular

RVOT = right ventricular outflow tract

TPVR = transcatheter pulmonary valve replacement

TR = tricuspid regurgitation

FIGURE 1 Pre- and Post-Procedural Imaging of Pulmonary Regurgitation



(A) Transthoracic echocardiogram from the parasternal short axis view demonstrates severe right ventricular dilation and moderate hypertrophy **(star)** secondary to free pulmonary regurgitation. **(B)** Free pulmonary regurgitation noted by the **red flow** through the pulmonary valve. **(C)** Cardiac magnetic resonance showing severe right ventricular dilation. The right ventricular end-diastolic volume measured 301 ml/m^2 (normal: <150 ml/m^2), with a right ventricular ejection fraction of 24% and a pulmonary regurgitation fraction of 57%. **(D)** Angiogram showing free pulmonary regurgitation, as seen by contrast filling in the right ventricle **(star). (E)** Angiogram showing that following conduit stenting and valve implantation, there is no regurgitation of contrast material noted in the right ventricle, a finding indicating a new competent pulmonary valve. CRAN = cranial; RAO = right anterior oblique.

balloons. Simultaneous bilateral branch PA and distal conduit balloon angioplasty was performed with a combination of noncompliant balloons to provide an expansion diameter of 18.5 mm. A 20-mm Melody valve (Medtronic, Inc.) was delivered on an 18-mm Ensemble delivery system (Medtronic, Inc.). The final angiogram showed excellent relief of obstruction and no valvar regurgitation (Video 1, **Figure 1E**).

FOLLOW-UP

Our patient experienced immediate improvement in kidney function and gradual clinical improvement in his cardiorespiratory status. The degree of generalized edema improved, and his B-type natriuretic protein trended down to reassuring levels over the subsequent 2 weeks (1,324 to 364 pg/ml). At 5 months post-intervention, he remains clinically well, with stable renal function, no interval admissions for acute kidney injury or volume overload, and an overall decrease in his diuretic medication requirement.

CONCLUSIONS

TPVR is now considered standard treatment for select patients with RV-PA conduit dysfunction. This procedure is most often performed through femoral vein access; however, the jugular venous approach is often effective in smaller children (1,2). In very small patients and those with complex anatomy, a perventricular hybrid approach provides direct access to the landing zone and allows for successful conduit preparation and implantation of TPVR when the transvenous route is unsuccessful.

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REFERENCES

1. Berman DP, McElhinney DB, Vincent JA, Hellenbrand WE, Zahn EM. Feasibility and short-term outcomes of percutaneous transcatheter pulmonary valve replacement in small (<30 kg) children with dysfunctional right ventricular outflow tract conduits. Circ Cardiovasc Interv 2014;7:142–8.

2. Zampi J, Berman D, Bocks M, et al. Factors associated with the internal jugular venous approach for Melody™ transcatheter pulmonary valve implantation. Cardiol Young 2015;26:948–56.

KEY WORDS chronic heart failure, stenosis, tetralogy of Fallot

APPENDIX For a supplemental video, please see the online version of this article.