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Percutaneous Closure of Paravalvular Leak After Tricuspid Valve Replacement for Ebstein's Anomaly

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Case Description

A 14-year-old girl was referred to the cardiology clinic for worsening dyspnea and lower-limb edema. She had a history of Ebstein's malformation associated with bicuspid aortic valve, coarctation of aorta, perimembranous ventricular septal defect (VSD), and subvalvular fibromuscular aortic stenosis. She had undergone coarctation of aorta repair, subaortic fibromuscular stenosis resection, and VSD patch closure during the first months of her life.

A Doppler color echocardiogram showed signs of Ebstein's disease (Carpentier type C) (Fig. 1) with severe tricuspid valve incompetence, mild left ventricular dysfunction (42%), bicuspid aortic valve with mild incompetence,

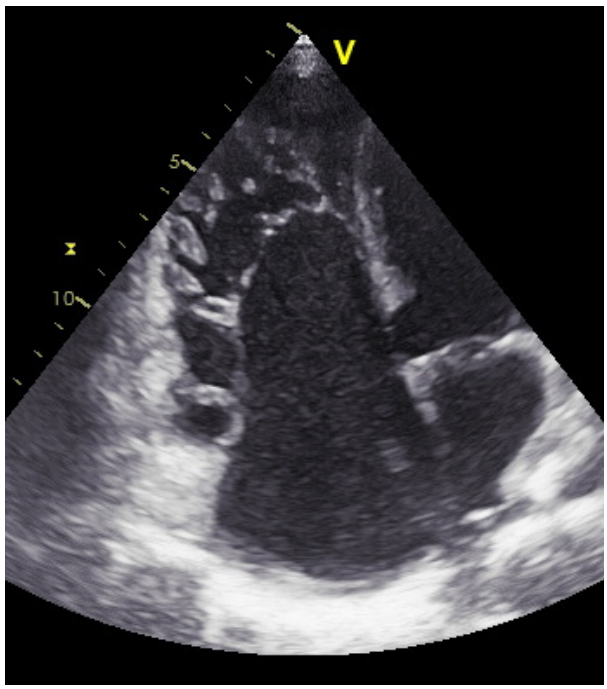

Fig. 1 Doppler color echocardiogram. Apical 4-chamber view shows the typical apical displacement of the tricuspid valve (severe Ebstein's malformation).

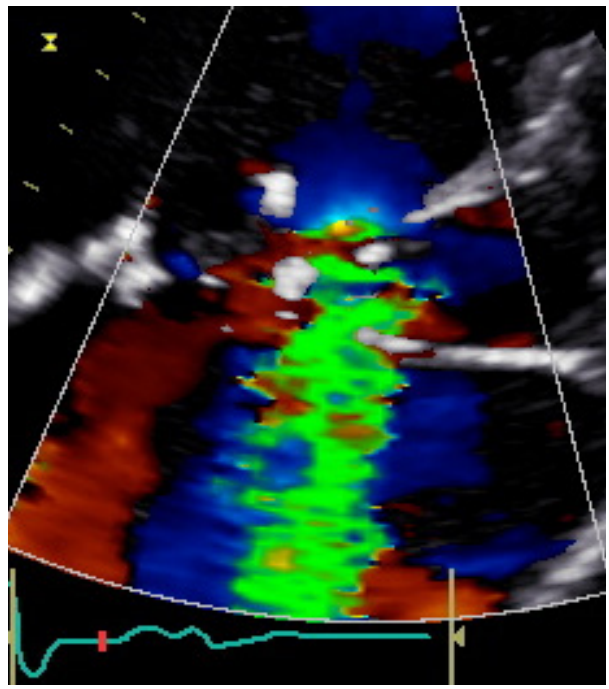

Fig. 2 Postsurgical Doppler color echocardiogram shows a severe tricuspid perivalvular leak.

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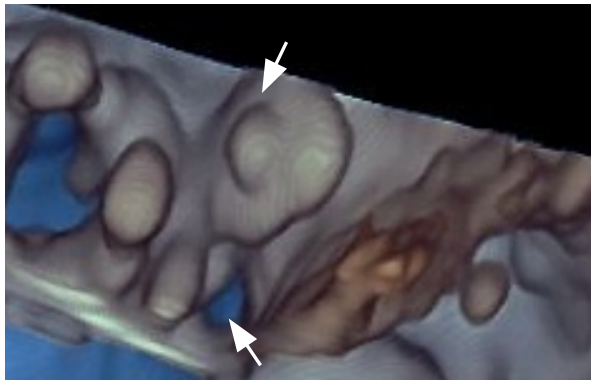

Fig. 3 Transthoracic echocardiography image after 3-dimensional reconstruction shows a right ventricular view. The PVL orifice is visualized located at 4 o'clock, close to the previously implanted VSD device.

PVL, perivalvular leak; VSD, ventricular septal defect.

Fig. 4 Transesophageal echocardiography image after 3-dimensional reconstruction shows the delivery sheath across the perivalvular leak from a right ventricular view.

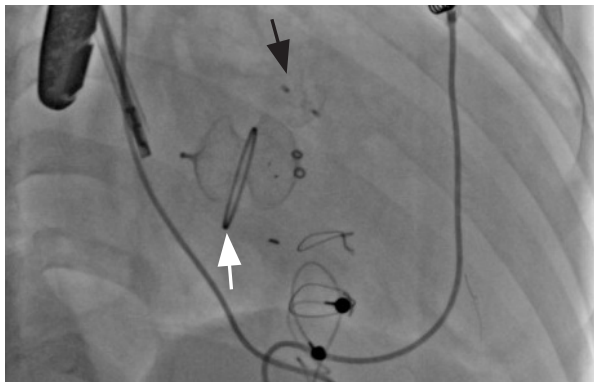

Fig. 5 Fluoroscopy image in a right anterior oblique projection shows the prosthetic biological valve ring (white arrow), both the VSD (black arrow) and the PVL devices, and the epicardial pacemaker leads.

PVL, perivalvular leak; VSD, ventricular septal defect.

Abbreviations and Acronyms

PVL	perivalvular leak
VSD	ventricular septal defect

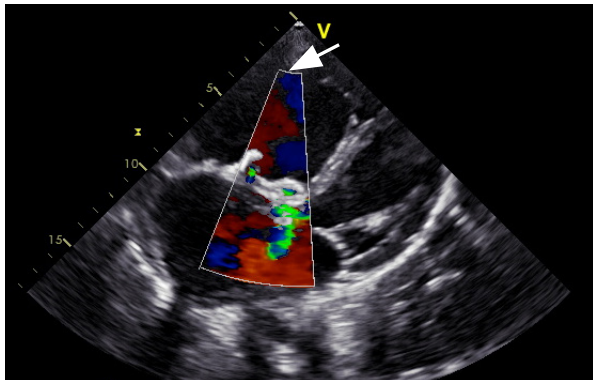

Fig. 6 Modified 4-chamber view showing a small residual shunt through the PVL device after intervention.

PVL, perivalvular leak.

and moderate residual VSD. Because of her clinical deterioration (New York Heart Association functional class III), she underwent surgical tricuspid valve replacement using a biological prosthesis (Hancock II 25 mm; Medtronic). Her postoperative course was complicated; volume/pressure overload through the residual VSD was initially considered and the defect percutaneously closed. Nevertheless, she continues to have pronounced symptoms (dyspnea, edema, and mild cyanosis) related to the large tricuspid paravalvular leak (PVL) measuring 12 mm × 10 mm (Fig. 2 and 3).

The PVL was percutaneously approached via the right internal jugular vein, and a 18-mm × 10-mm rectangular PVL device (Occlutech) was implanted with uneventful results with regard to closing the defect (Fig. 4 and 5). After 3 months, the patient's functional class had improved, and a trivial residual shunt was visualized (Fig. 6).

Comment

Ebstein's malformation is a rare and complex congenital lesion. Surgical treatment is recommended when progressive cardiomegaly, exercise intolerance, cyanosis, paradoxical embolism, or arrhythmias (atrial or ventricular) are observed. Tricuspid valve replacement is necessary when repair is not feasible or has failed. Frequently,

a biological prosthesis is preferred to a mechanical valve because of its lower thrombogenicity, mainly when right ventricular failure is present.[1]

The success of interventions for tricuspid PVL in congenital heart disease is anecdotal.[2,3] As an alternative to surgery, transcatheter occlusion has often been performed for mitral and aortic paravalvular leaks.[4] This case illustrates that a percutaneous approach for closure of a tricuspid PVL is feasible and may be a reasonable alternative to surgery in select patients.

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