

# Three-Dimensional Echocardiography Reveals Extensive Congenital Anterior Tricuspid Valve Prolapse



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## INTRODUCTION

Tricuspid valve (TV) surgery is usually performed secondary to left-sided valve surgery. Here, we report the case of a patient who underwent isolated TV repair because of severe tricuspid regurgitation (TR) caused by massive prolapse of the anterior leaflet and congenitally absent chordae tendineae.

Congenital TV morphologic abnormalities are difficult to evaluate using standard two-dimensional (2D) transthoracic echocardiography (TTE) because it is incredibly challenging to simultaneously visualize the three leaflets of the TV. In addition to conventional 2D TTE, three-dimensional (3D) transesophageal echocardiography (TEE) is useful in the comprehensive assessment of valve morphology. In our patient, using 3D TEE assisted in evaluating this complicated case of severe TR due to a congenital abnormality.

## CASE PRESENTATION

In 2017, a 44-year-old Japanese man presented to our hospital with cold sweats, dyspnea, and palpitations. He was diagnosed with an asymptomatic cardiac murmur at the age of 7 years, and he did not visit the hospital for follow-up. Five years ago, he was diagnosed with right bundle branch block, but once again, he did not visit the hospital for follow-up, because of the lack of symptoms. Upon presentation to our hospital, a physical examination revealed obesity, with an increased body mass index of 28.33 kg/m<sup>2</sup> (height 167 cm, weight 79 kg). Blood pressure was 106/78 mm Hg, and heart rate was 64 beats/min, both of which were unremarkable. Cardiac auscultation revealed an apical pansystolic murmur. The patient showed no jugular venous distention, and lung sounds were clear. Laboratory data did not reveal any significant findings. Electrocardiography revealed sinus rhythm with an incomplete right bundle branch block (Figure 1). Chest radiography did not reveal any congestion, and the cardiothoracic ratio was 54% (Figure 2). On performing TTE, we instantly recognized severe TR due to massive prolapse of the TV by color

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130

## VIDEO HIGHLIGHTS

**Video 1:** TTE in the modified apical four-chamber view showing a prolapse of the posterior TV leaflet.

**Video 2:** TTE in the modified apical four-chamber view with color Doppler showing severe TR due to prolapse of the posterior leaflet.

**Video 3:** TEE in the midesophageal four-chamber view showing prolapse of the posterior TV leaflet.

**Video 4:** TEE in the midesophageal four-chamber view with color Doppler showing severe TR due to prolapse of the posterior leaflet.

**Video 5:** TEE in the midesophageal short-axis view showing prolapse of the posterior TV leaflet.

**Video 6:** TEE in the midesophageal short-axis view with color Doppler showing severe TR due to prolapse of the posterior leaflet.

**Video 7:** Three-dimensional TEE in the right atrial view showing prolapse of the anterior TV leaflet.

**Video 8:** Three-dimensional TEE in the right atrial view with color Doppler showing severe TR due to prolapse of the anterior leaflet.

**Video 9:** Three-dimensional TEE by multiplanar reconstruction in the right atrial view showing prolapse of the anterior TV leaflet.

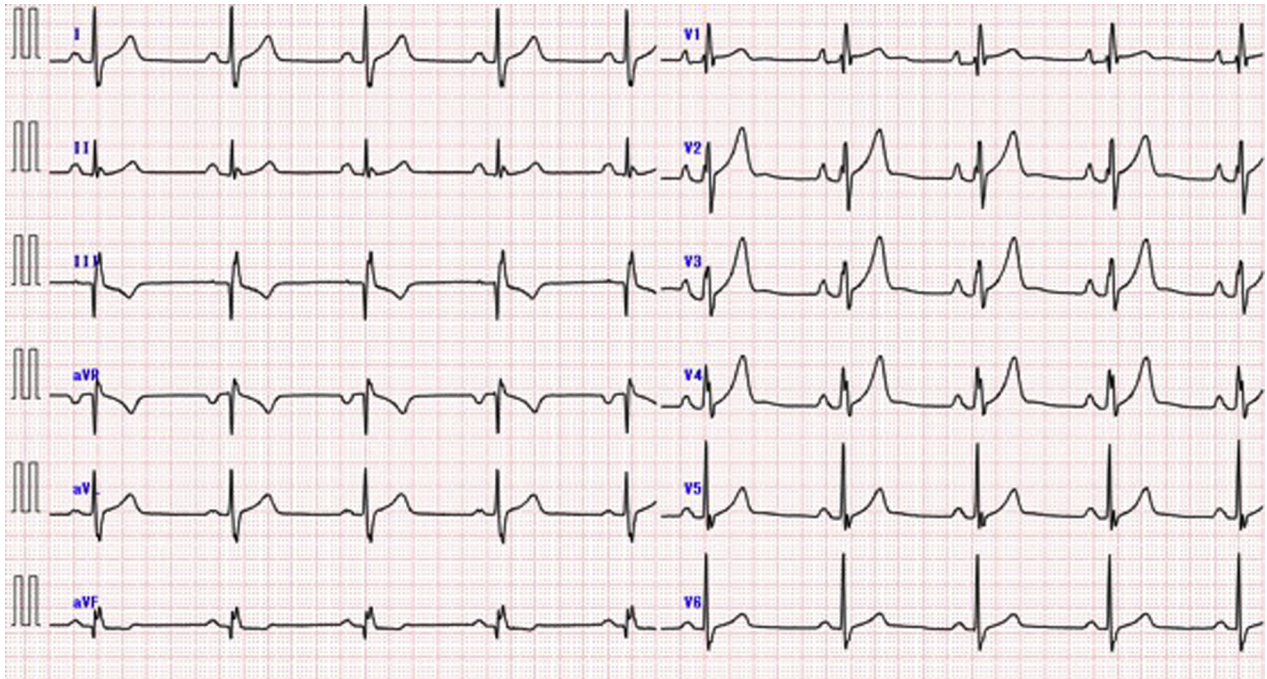
**Video 10:** Three-dimensional TEE by multiplanar reconstruction in the right atrial view with color Doppler showing severe TR due to prolapse of the anterior leaflet.

**Video 11:** Follow-up TTE in the modified apical four-chamber view, showing reduced RV and right atrial volume.

**Video 12:** Follow-up TTE in the modified apical four-chamber view with color Doppler, showing trivial TR.

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Doppler using a four-chamber view. The initial transthoracic echocardiographic examination suggested severe prolapse of the posterior TV leaflet (Figure 3, Videos 1 and 2) as well as a remarkably dilated right ventricle, a preserved right ventricular (RV) fractional area change of 50%, and a dilated right atrium and TV annulus. The estimated RV pressure with TR was 32 mm Hg at rest. Left ventricular function was normal, with intact aortic and mitral valves. Subsequently, the patient underwent 2D TEE, and again on initial 2D imaging, he appeared to show posterior TV leaflet prolapse (Figure 4, Videos 3-6).



**Figure 1** Electrocardiogram showing sinus rhythm with an incomplete right bundle branch block.



**Figure 2** Chest radiography showing no congestion; cardiothoracic ratio was 54%.

However, 3D TEE and multiplanar reconstruction images revealed a massive prolapsed huge anterior leaflet but not posterior TV leaflet (Figures 5 and 6, Videos 7-10). Ruptured chordae tendineae were

not detected. The TV annulus, measuring 56 mm, was remarkably dilated. Computed tomography revealed an intact coronary artery.

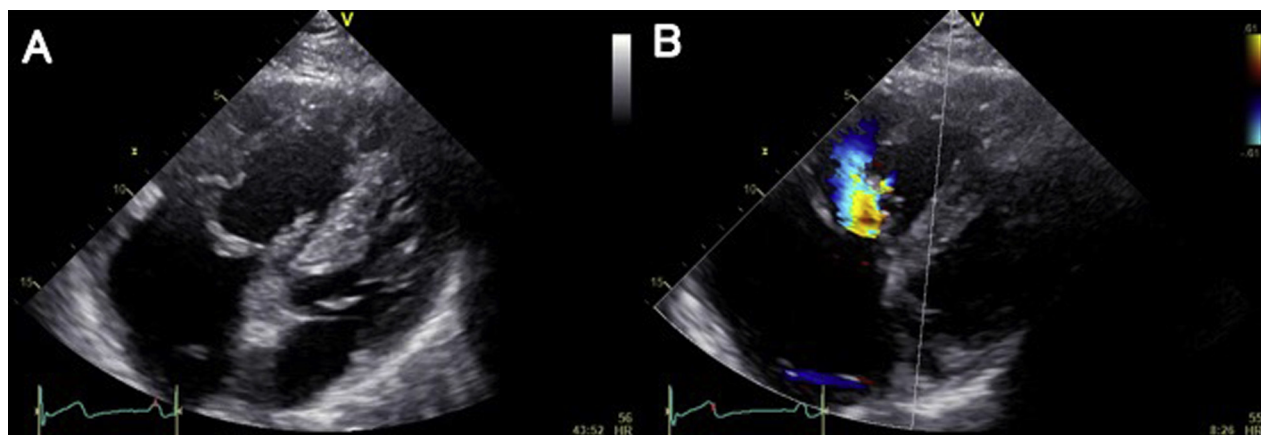
These examinations revealed that the clinical symptoms of our patient were caused by severe TR due to massive prolapse of the anterior leaflet. He was then referred for surgical TV repair. Surgical inspection revealed a massive prolapse of the anterior leaflet and absence of the anterior TV chordae (Figure 7). He underwent TV repair via triangular resection of the redundant anterior leaflet; three artificial chordae were added, and annuloplasty was performed with a 32-mm Edwards MC3 annuloplasty ring (Edwards Lifesciences, Irvine, CA). The postoperative course was uneventful, and the surgery resolved the patient's symptoms. Six months after discharge, follow-up TTE revealed trivial TR and reduced RV and right atrial volume (Figure 8, Videos 11 and 12).

## DISCUSSION

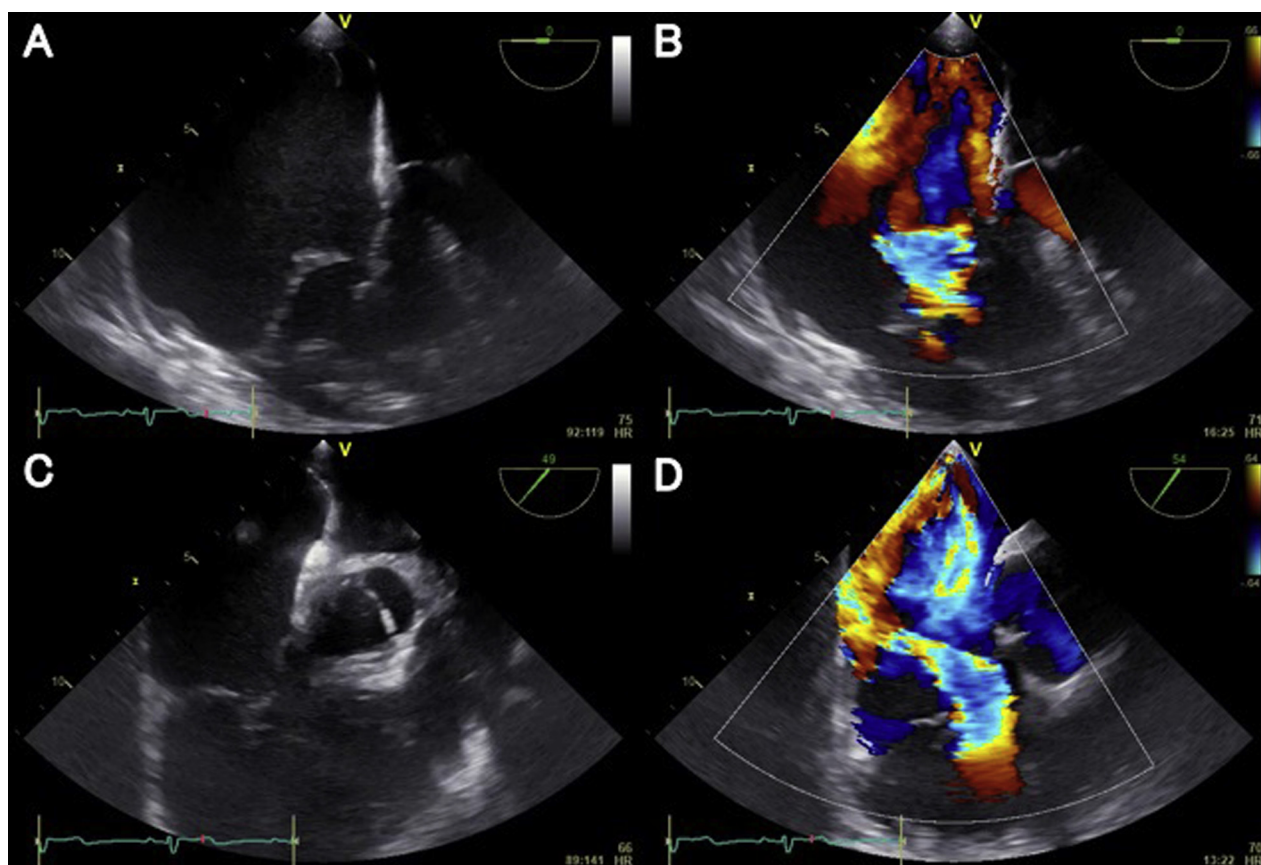
Isolated severe TR is associated with RV remodeling and can cause RV dysfunction. Severe TR has been reported to be associated with poor prognosis, independent of age and biventricular systolic function.<sup>1</sup> Therefore, the recommended optimal timing of surgical intervention for severe TR is before the onset of progressive RV dysfunction. In our case of congenital isolated TR, the patient underwent TV repair without RV dysfunction, resulting in postoperative reductions of RV and right atrial volume.

The American Heart Association/American College of Cardiology 2014 guidelines for the management of patients with valvular heart disease recommend isolated TV repair for symptomatic severe TR (class I) or when accompanied with progressive RV dysfunction (class IIb).<sup>2</sup> The 2017 European Society of Cardiology/European Association for Cardio-Thoracic Surgery guidelines for the management of valvular heart disease also recommend surgery for patients with primary symptomatic severe TR and asymptomatic patients with progressive RV dilatation or a decline in RV function.<sup>3</sup>





**Figure 3** TTE in the modified apical four-chamber view (A) and color Doppler (B), showing severe TR due to prolapse of the posterior leaflet.

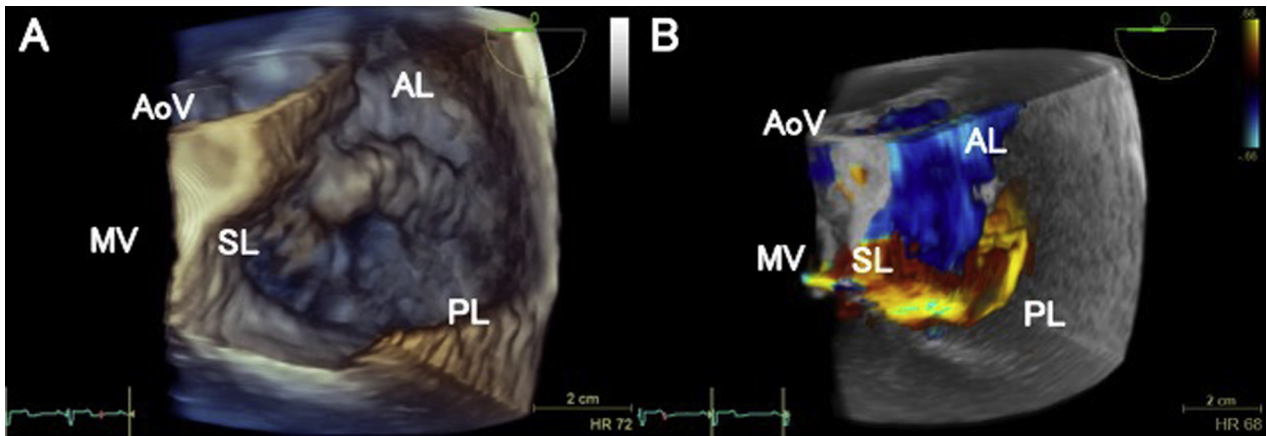


**Figure 4** TEE in the midesophageal four-chamber view (A) and color Doppler (B), the midesophageal short-axis view (C), and color Doppler (D), showing severe TR due to prolapse of the posterior leaflet.

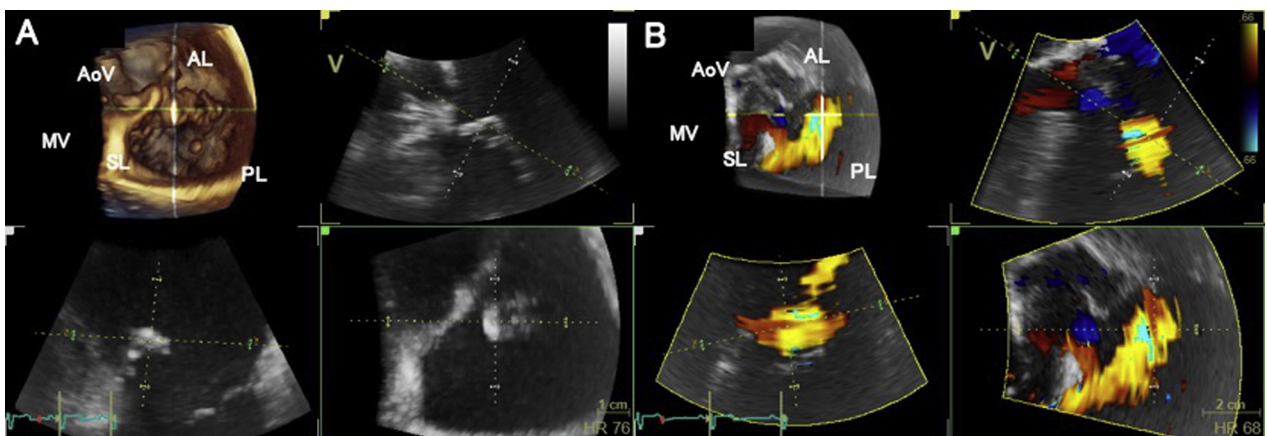
Although the prevalence of severe TV disease is on the rise,<sup>4</sup> surgery for isolated TV disease is rarely performed despite these recommendations, because of the relatively high in-hospital mortality rate.<sup>5,6</sup> Previous studies have reported that the 2-year event-free survival rate following TV surgery in patients with preoperative severe RV dysfunction is 57%.<sup>7</sup>

Cases of isolated TV surgery are limited in number in comparison with those of aortic or mitral valve repair. Consequently, there are

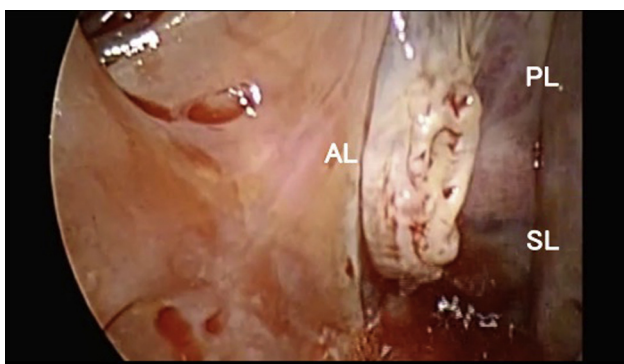
fewer opportunities for evaluating the morphology of the TV with TTE for surgical repair. In this case, the conventional four-chamber view and short-axis view on 2DTTE led to the mistaken interpretation of posterior TV leaflet prolapse. However, 3D TTE rapidly revealed a massive prolapse of the anterior leaflet, which was subsequently confirmed upon surgical inspection. This case highlights that standard 2D TTE is limited with regard to the accurate identification of TV leaflets. Three-dimensional multiplanar reconstruction should be added



**Figure 5** Three-dimensional TEE in the right atrial view (A) and color Doppler (B), showing severe TR due to prolapse of the anterior leaflet (AL). AoV, Aortic valve; MV, mitral valve; PL, posterior leaflet; SL, septal leaflet.



**Figure 6** Three-dimensional TEE by multiplanar reconstruction in the right atrial view (A) and color Doppler (B), showing severe TR due to prolapse of the anterior leaflet.



**Figure 7** Surgical inspection showing a massive prolapse of the anterior leaflet (AL). PL, Posterior leaflet; SL, septal leaflet.

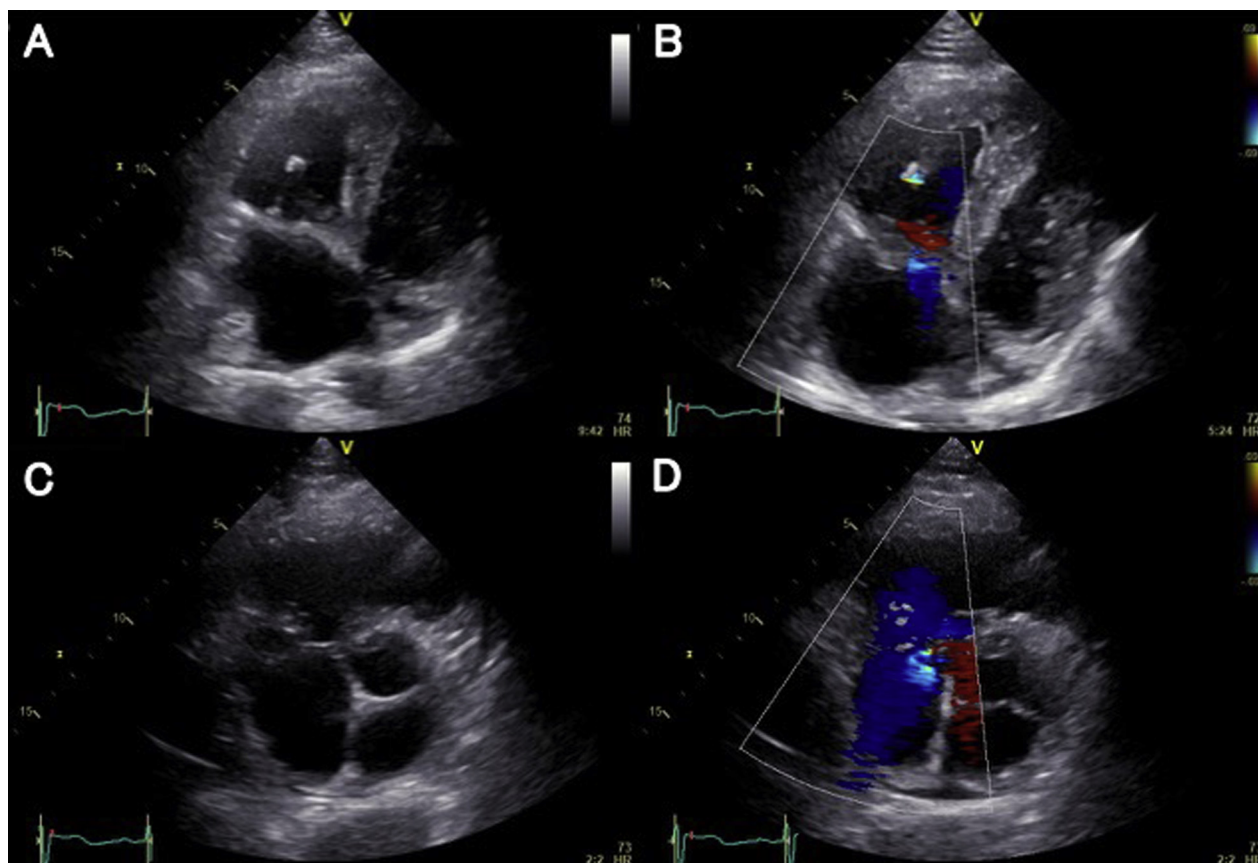
to the routine examination of abnormal TVs. By using 3D TEE, we could easily and instantly visualize this complex morphology to make an accurate diagnosis. Three-dimensional TEE visualization of the TV is more challenging than that of the mitral valve. In this case, we could visualize the prolapsed anterior TV leaflet itself but could not detect the absent chordae of the anterior TV in the RV using 3D TEE in advance of surgery. However, in cases with right atrial

and ventricular dilatation, it is usually possible to obtain diagnostic images. This has been demonstrated with the use of clip repair of the TV, for which transesophageal echocardiography guidance remains essential.<sup>8</sup>

Multiple percutaneous therapies have recently been developed for treating severe TR, including TV repair and replacement.<sup>8,9</sup> Echocardiography has become a critical tool for the initial assessment of the etiology and severity of TR before these interventions. Although cardiac magnetic resonance imaging may provide better and clearer morphologic information, there are only a few cases of magnetic resonance imaging examinations compared with those conducted with the widely used TEE. It is expected that not only 2D but also 3D TEE will become the gold standard in the evaluation of this forgotten valve.<sup>10</sup>

This case showed a structural abnormality of the prolapsed anterior tricuspid leaflet and absent chordae of the anterior tricuspid leaflet, which is classified as congenital TV dysplasia.<sup>11</sup> Regarding congenital TR, Ebstein anomaly is the most well known of the congenital TV diseases<sup>12</sup>; however, it is extremely rare, constituting only 0.5% of all congenital heart diseases.<sup>13</sup> TV dysplasia is a congenital structural abnormality of the leaflet, chordae, or papillary muscle associated with the TV and is rarer than Ebstein anomaly. This case provided us the opportunity to recognize the difficulty and importance of a comprehensive evaluation of a TV complex using echocardiography.





**Figure 8** Follow-up TTE in the modified apical four-chamber view (A) and color Doppler (B), the short-axis view (C), and color Doppler (D), showing trivial TR and reduced RV and right atrial volume.

Similar cases have been previously reported. The first case was of congenitally absent chordae in the anterior TV leaflet with hypoplastic papillary muscle.<sup>14</sup> The second case was of the congenital absence of anterior papillary muscle of the TV.<sup>15</sup> These case reports stated that 2D TTE was used for preoperative evaluation; however, a detailed evaluation using 3D TEE was not reported. The prolapsed leaflets in these two cases were smaller than those in our case and thus relatively easier to distinguish. Our patient had an extremely massive prolapse, and the prolapsed leaflet size made the evaluation more difficult. The use of 3D TEE was an effective solution in the detection of this extraordinary anatomical abnormality.

## CONCLUSION

Congenital leaflet prolapse with absent chordae of the TV is particularly rare among congenital heart diseases. Although 2D TTE is considered the gold standard to evaluate valve anatomy and function, we showed that 3D TEE can be a useful tool to augment 2D TTE for a comprehensive preoperative assessment of morphologic abnormalities of the TV.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.12.001>.

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