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Congenital tricuspid valve dysplasia in a septuagenarian

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Primary isolated tricuspid regurgitation (TR) can be due to congenital or acquired tricuspid valve (TV) pathology. Anatomic studies suggest great variability in number and morphology of TV cusps with only 62% having three cusps.[1] We describe the case of a 77-year-old woman presenting with her first episode of right heart failure secondary to severe TR. Transoesophageal echocardiography revealed a congenital bileaflet tricuspid valve. The patient underwent surgical TV replacement, visually confirming fusion of the anterior and posterior leaflets of her TV with no other congenital abnormalities.

A 77-year-old woman presented with exertional dyspnoea and symptoms of right heart failure (New York Heart Association III–IV). Her comorbidities include chronic atrial fibrillation, obstructive sleep apnoea, non-obstructive coronary artery disease, hypertension, and asthma. She was a lifelong non-smoker. Blood results including liver function tests were normal.

Transthoracic echocardiogram showed an apparently normal tricuspid valve (TV) with moderate-to-severe regurgitation, mildly dilated right ventricle (RV) with normal systolic function, mildly dilated right atrium (RA), borderline pulmonary pressure [right ventricular systolic pressure (RVSP 35 mmHg)], and normal left ventricular size and function.

Coronary angiogram showed minor coronary disease with normal left ventricular (LV) function and LV end-diastolic pressure (10 mmHg). Right heart catheterization showed normal pulmonary and

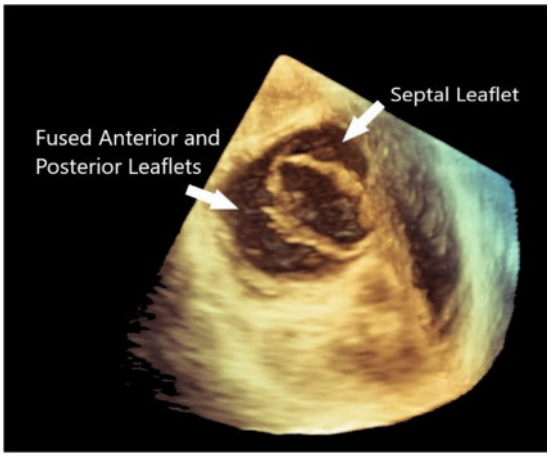

Figure 1 Three-dimensional transoesophageal echocardiography (TOE) showing atrial view of the tricuspid valve (TV) with two leaflets. AP, fused anterior and posterior leaflets; S, septal leaflet.

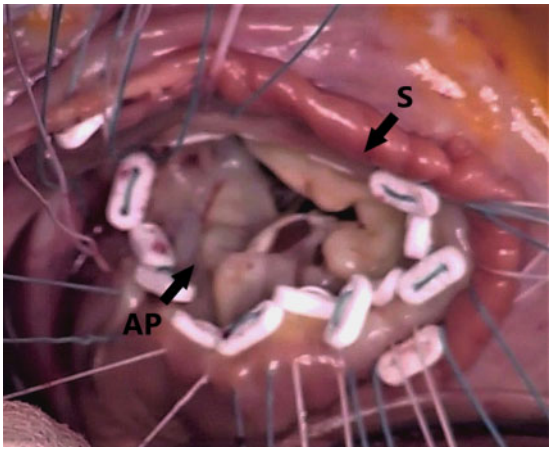

Figure 2 Intraoperative photograph of tricuspid valve with fusion of the anterior and posterior leaflets. AP, fused anterior and posterior leaflets; S, septal leaflet.

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RA pressures (6 mmHg), although large tricuspid regurgitation (TR) waves were evident, and no left to right shunt. Her cardiac output was reduced at 3.53 L/min (cardiac index 2.11 L/min/m^2) by thermodilution.

Transoesophageal echocardiogram showed bileaflet (fusion of anterior and posterior leaflets) TV, with no prolapse or flail leaflets (*Figure 1*). There was moderate-to-severe TR with moderately dilated RV and preserved systolic function.

As no other cardiac or respiratory cause for her dyspnoea was found, she proceeded to surgical TV replacement with a 29 mm Edwards Magna Ease prosthesis. Intraoperative findings confirmed the presence of a bileaflet TV (*Figure 2*). No other congenital abnormalities were identified.

This case highlights the importance of congenital TV abnormalities as a differential for primary isolated TR, resulting in RV and RA dilatation, with right heart failure, regardless of the age at presentation. We hypothesize that congenital TV pathology can present later in life due to the lower right-sided pressures. While functional TR remains the most prevalent aetiology, primary isolated TR (both congenital and acquired) still accounts for 10% of TR.[2] Tricuspid valve surgery remains the definitive therapy for right heart failure from primary isolated TV disease.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidelines.

Conflict of interest: none declared.

References

1. Gerola LR, Wafae N, Vieira MC, Juliano Y, Smith RBW, Prates J. Anatomic study of the tricuspid valve in children. *Surg Radiol Anat* 2001;**23**:149–154.
2. Taramasso M, Pozzoli A, Guidotti A, Nietlispach F, Inderbitzin DT, Benussi S *et al.* Percutaneous tricuspid valve therapies: the new frontier. *Eur Heart J* 2017;**38**: 639–647.