

Isolated double-chambered right ventricle in an adult patient: intraoperative imaging with three-dimensional transoesophageal echocardiography

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A 43-year-old female presented with dyspnoea on moderate activity of 1-year duration. Transthoracic echocardiography (TTE) showed double-chambered right ventricle (DCRV). This was confirmed by right ventricular (RV) angiography. The preoperative peak systolic gradient measured 140 mmHg. No other cardiac lesions were identified. She underwent intracardiac repair with resection of the hypertrophied bands. Intraoperative two- and three-dimensional transoesophageal echocardiography (TOE) showed the anomalous discrete hypertrophied muscle bands near the RV outflow tract (*Figure 1*; [Supplementary material online, Video S1](#)). Combined trans-pulmonary arteriotomy-trans-right atriotomy surgical approach was employed. Findings revealed a band like constriction dividing the RV into proximal and distal chambers (*Figure 2*). Post-resection peak systolic gradient measured 36 mmHg.

Double-chambered right ventricle is a rare congenital heart disease (CHD) where the RV is divided by anomalous muscle bundles into high pressure inlet and low pressure outlet chambers.¹ The incidence is 0.5–2% of all cases of CHD.¹ It usually results from conditions leading to increased flow and pressure within the RV like ventricular septal defects, pulmonary valve obstruction, or tetralogy of Fallot (TOF).¹ The other variant is acquired DCRV which results from an abnormally short distance between the moderator band and the pulmonary valve.¹ The anomalous muscles may be located near the pulmonary valve (high position) or the apex (low position).¹ The main differential diagnosis of DCRV is TOF. The absence of cyanosis and pulmonary valvular or subvalvular abnormalities are more in favour of DCRV.

Adult patients presenting with isolated DCRV are extremely rare.^{1,2} Although two-dimensional TOE is reported to be more

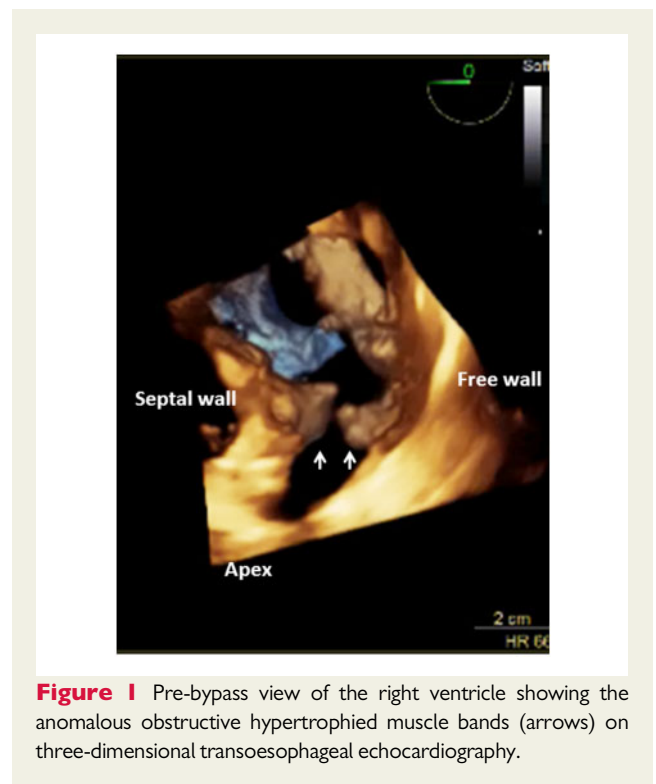


Figure 1 Pre-bypass view of the right ventricle showing the anomalous obstructive hypertrophied muscle bands (arrows) on three-dimensional transoesophageal echocardiography.

... sensitive than TTE in identifying this condition in adults,¹ it may still be of limited value because of an anterior location and increased trabeculations of the RV. Three-dimensional imaging allows free

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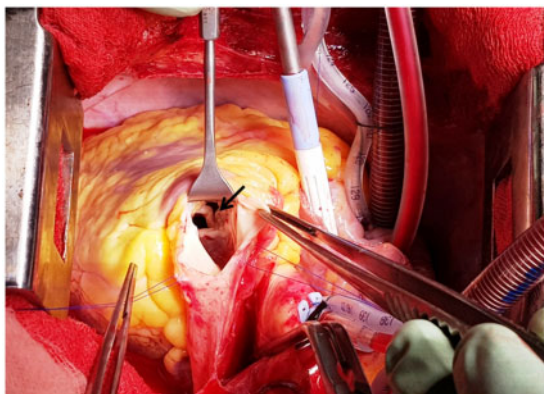


Figure 2 Surgical findings showing the obstructive band with a narrow orifice (black arrow).

rotation and cropping on to the area of interest and may result in better structural delineation.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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 guidance.

Conflict of interest: none declared.

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