

## CLINICAL IMAGE

# Gross right heart dilatation secondary to Ebstein's anomaly

Gaurav Singh Gulsin<sup>1,\*</sup>, Daniel Swarbrick<sup>2</sup>, and Gerry P. McCann<sup>2</sup>

<sup>1</sup>Department of Cardiovascular Sciences, University Hospitals of Leicester NHS Trust, Glenfield General Hospital, Leicester, UK, and <sup>2</sup>Department of Cardiovascular Sciences, The University of Leicester, Glenfield General Hospital, Leicester, UK

\*Correspondence address. Department of Cardiovascular Sciences, Glenfield General Hospital, University Hospitals of Leicester NHS Trust, Groby Road, Leicester LE3 9QP, UK. Tel: +44-300-303-1573; E-mail: gaurav.gulsin@nhs.net

Cardiac imaging is essential for the diagnosis and follow-up individuals with congenital heart disease (CHD). Currently echocardiography is the first-line investigation, although no single imaging technique is sufficient for complete evaluation of cardiac anatomy and physiology in these patients. Increasingly, adults with CHD are referred for cardiac magnetic resonance (CMR) imaging. The high spatial resolution, functional assessment capabilities and excellent reproducibility of CMR make it a desirable investigation for CHD [1].

Ebstein's anomaly is a rare (incidence  $\approx$ 1 in 200 000 live births) congenital heart disorder characterized by malformations of the tricuspid valve (TV) and right ventricle (RV). The hallmark features are clearly demonstrated in this CMR study. They are:

1. Malposition of the posterior and septal TV leaflets in the walls of the RV with enlargement, fenestration and/or tethering of the anterior leaflet.
2. Dilatation of the true atrio-ventricular junction and downward displacement of the functional TV annulus.
3. Atrialization of the RV [2].

In the four-chamber view (Fig. 1A), the grossly dilated right heart is clearly evident, with an atrialized RV and dilated tricuspid annulus. The enlarged anterior TV leaflet is well visualized (red arrow), as is the hypoplastic septal leaflet (yellow arrow). The LA and LV are small, owing to compression from the volume-overloaded right heart.

The massively dilated atrial and ventricular portions of right heart are visualized in the short-axis cine view (Fig. 1B). Again the tricuspid valve septal leaflet is seen (red arrow) and

the pale jet of tricuspid regurgitation is notable during systole. The intraventricular septum flattened during diastole consistent with right-sided volume overload and there is slow filling of the LV consistent with ventricular-ventricular interaction.

## SUPPLEMENTARY MATERIAL

Supplementary material is available at *Oxford Medical Case Reports* online.

## CONFLICT OF INTEREST STATEMENT

None declared.

## FUNDING

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## ETHICAL APPROVAL

None required.

## CONSENT

The patient presented in this report gave his full written informed consent for publication of this paper.

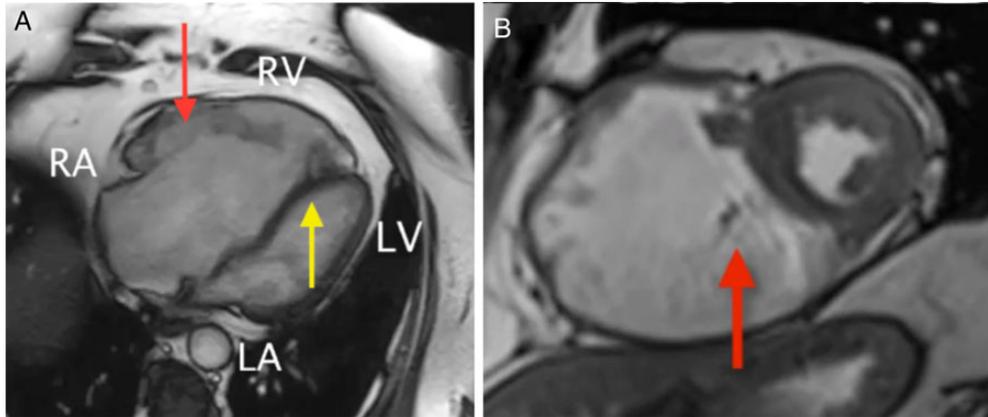
## GUARANTOR

Dr Gaurav S. Gulsin.

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**Figure 1:** (A) CMR four-chamber cine view. Seen here is the grossly dilated right heart, with an atrialized RV and dilated tricuspid annulus. The enlarged anterior TV leaflet is well visualized (red arrow), as is the mislaid and hypoplastic septal leaflet (yellow arrow). The LA and LV are small, owing to compression from the volume-overloaded right heart. LA, left atrium; LV, left ventricle; RA, right atrium. RV, right ventricle. (B) CMR short-axis cine view. The massively dilated atrial and ventricular portions of right heart are visualized. Again the tricuspid valve septal leaflet is seen (red arrow) and the pale jet of tricuspid regurgitation is notable here in systole.

## REFERENCES

1. Ntsinjana HN, Hughes ML, Taylor AM. The role of cardiovascular magnetic resonance in pediatric congenital heart disease. *JCMR* 2011;13:51.
2. Attenhofer CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Congenital heart disease for the adult cardiologist: Ebstein's anomaly. *Circulation* 2007;115:277–285.