

Images in  
Cardiovascular Disease



# Diagnosis of Ebstein Anomaly with Atrial Septal Defect and Persistent Left Superior Vena Cava Using Cardiac Magnetic Resonance Imaging

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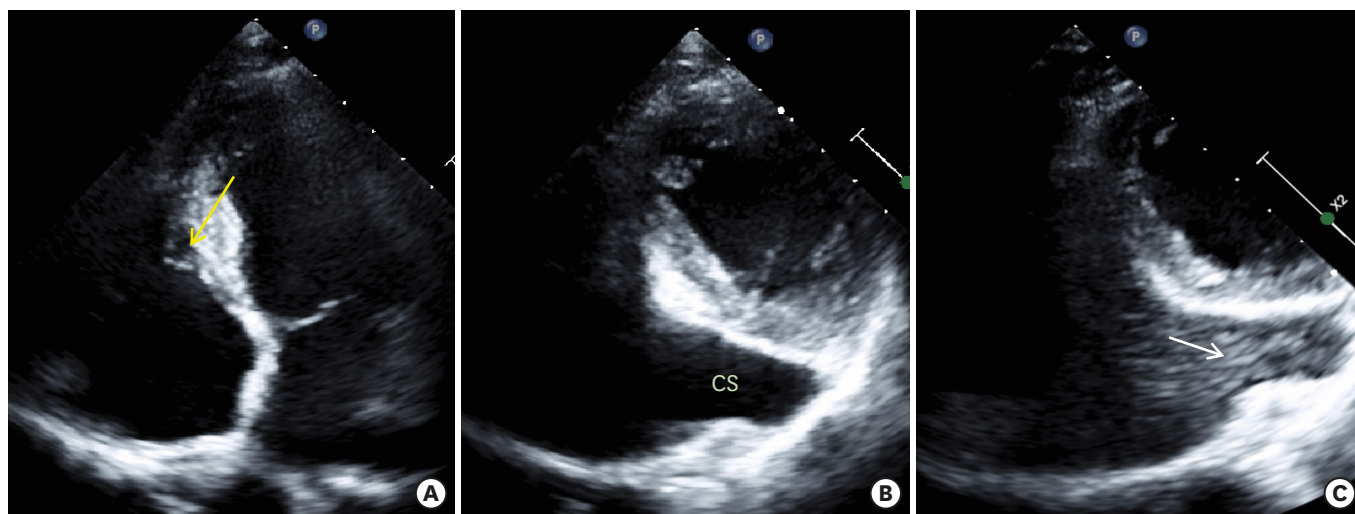
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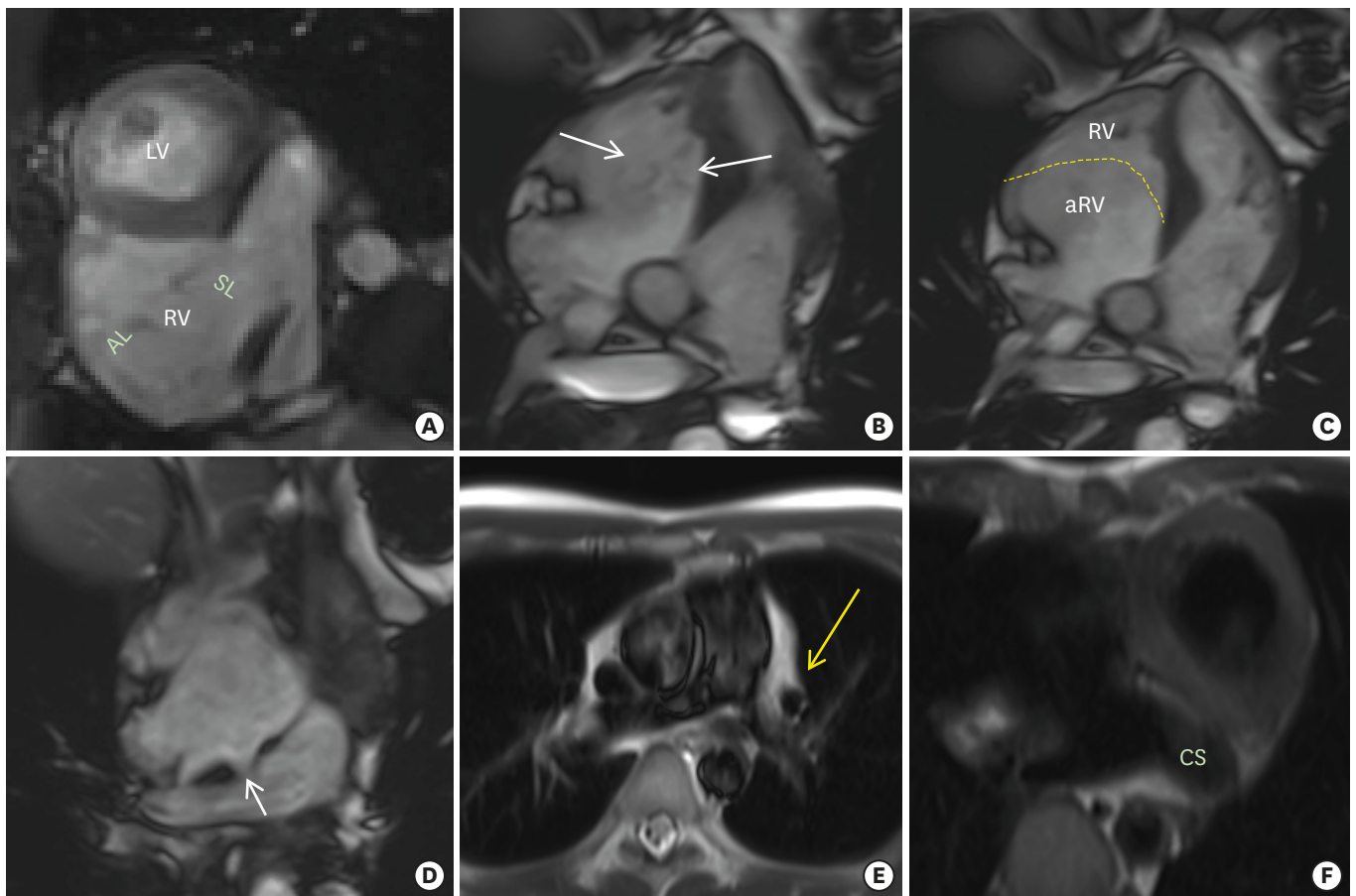
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A 26-year-old man was admitted to the hospital with acute onset of palpitation. His electrocardiogram revealed atrial fibrillation. Transthoracic echocardiography showed a huge coronary sinus and apical displacement of the tricuspid septal leaflets (**Figure 1A-B** and **Movie 1**). We suspected the Ebstein anomaly and tried to find atrial septal defect (ASD), which is often accompanied by such anomalies. But we could not find this defect because he had a very poor acoustic window, and the heart structure was hidden by a huge coronary sinus. To identify persistent left superior vena cava (PLSVC) that may be causing huge coronary sinus, we performed contrast echocardiogram with agitated saline. After infusing saline via the left antecubital vein, the coronary sinus is filled with agitated saline before right atrial enhancement (**Figure 1C** and **Movie 2**). This process suggested the existence of PLSVC. For detailed confirmation, the patient underwent cardiac magnetic resonance (CMR) imaging. Cine images showed apical displacement of the septal leaflet from the insertion of the anterior leaflet of the mitral valve by 1.35 mm/m<sup>2</sup> body surface area, consistent with Ebstein anomaly. In addition, 12-mm-sized secundum type ASD and presence of PLSVC were revealed (**Figure 2** and **Movie 3**).



**Figure 1.** Transthoracic echocardiogram: four-chamber view showing apical displacement of the tricuspid septal leaflets (A, yellow arrow) and huge CS (B). (C) CS is filled with agitated saline before right atrial enhancement (white arrow). CS: coronary sinus.



**Figure 2.** Cardiac magnetic resonance image. (A) Short-axis view showing severely dilated RV. Four-chamber views demonstrating apical displacement of the SL (B, white arrow), and aRV is shown between the atrioventricular junction (C, dotted line) and evidence of atrial septal defect (D). Panel (E) reveals a persistent left superior vena cava (yellow arrow), and panel (F) shows huge CS. AL: anterior tricuspid valve leaflet, aRV: atrialized part of the RV, CS: coronary sinus, LV: left ventricle, RV: right ventricle, SL: septal tricuspid valve leaflet.

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#### Conflict of Interest

The authors have no financial conflicts of interest.

Ebstein anomaly is a rare congenital heart disorder accounting for < 1% of all cases of congenital heart disease.<sup>1)</sup> Echocardiography is the diagnostic test of choice for this anomaly and is crucial for detecting the presence of associated cardiac malformations. However, echocardiography can be very limited, as in our case. This case highlights the usefulness of CMR imaging for detection of complex congenital heart disease.

## ACKNOWLEDGMENTS

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## SUPPLEMENTARY MATERIALS

### Movie 1

Transthoracic echocardiogram showing the apical displacement of the tricuspid septal leaflets.

[Click here to view](#)

**Movie 2**

Contrast transthoracic echocardiogram with agitated saline via left antecubital vein showing the coronary sinus is filled with agitated saline before right atrial enhancement.

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**Movie 3**

Cardiac MRI. Cine images showing the large atrialized right ventricle and 12mm sized secundum type ASD.

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## REFERENCES

1. Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. *Circulation* 2007;115:277-85.

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