Giant Right Atrium: A Rare Form of Congenital Heart Disease
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Clinical History
A 54-year-old male presented with a presumed diagnosis of Ebstein’s anomaly and atrial fibrillation. His past medical history was remarkable due to an exploratory cardiac surgery in his childhood for a suspected cardiac mass, which revealed an enlarged right atrium. A recent comprehensive echocardiogram revealed a severely enlarged right atrium with no obvious finding that would suggest Ebstein’s anomaly. He was referred for a cardiac magnetic resonance imaging (MRI) for further evaluation of cardiac anatomy and function.

Findings
The right ventricle and left heart chambers were displaced laterally and posteriorly by the enlarged right atrium (Figure 1A,B). The location and orientation of the tricuspid valve in relation to the mitral valve were normal with no evidence of apical displacement or tethering to suggest Ebstein’s anomaly. There was tricuspid annular dilatation and a resulting eccentric jet of tricuspid regurgitation seen on phase contrast images (Figure 2A,B).

Discussion
Idiopathic dilatation of the right atrium is a very rare condition of unknown origin and was first reported by Bailey. The partial loss of atrial muscle fibers with progressive atrial enlargement was proposed as the causative mechanism. The clinical presentation varies but is frequently an incidental finding detected on the chest radiography done for routine evaluation or during the evaluation of the atrial fibrillation. Approximately 50% of the patients are asymptomatic at the time of the diagnosis. The common presenting symptoms, when they occur, are shortness of breath (28%), palpitations (17%) and arrhythmia (12%).

Echocardiography, computed tomography (CT) and MRI can all provide important structural and functional information about this condition. MRI can provide high-resolution images of the right atrium and right ventricle as well as quantitative right ventricular volume and systolic function. In addition, alternative pathologies can be excluded. The differential diagnoses of this rare entity include Ebstein’s anomaly, Uhl’s anomaly, cardiac tumor, pericardial effusion or pericardial cyst. In our patient, a clinical diagnosis of Ebstein’s anomaly was first considered before the results of echocardiography and MRI were obtained. Surgical reduction of the right atrium can be considered in symptomatic patients although the optimal treatment of this rare condition is not well established.

REFERENCES