CASE REPORT

Cardiac Resynchronization Therapy in Patients with Ebstein’s Anomaly
A Case Report

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Summary
Ebstein’s anomaly is a rare congenital heart disease characterized by apical displacement of the septal and posterior tricuspid valve leaflets. It is often complicated with left ventricular (LV) dysfunction as well as right-sided abnormalities. On the other hand, in the presence of LV dysfunction, right ventricular pacing is likely to aggravate the diseased LV function, which is termed pacemaker-induced cardiomyopathy. Thus, deteriorating effects of RV pacing on cardiac function might be enhanced and result in pacemaker-induced cardiomyopathy in patients with Ebstein’s anomaly, even if they have preserved LVEF. Cardiac resynchronization therapy (CRT) is effective for the treatment of pacemaker-induced cardiomyopathy, and we present the first case of effect of CRT on pacemaker-induced cardiomyopathy associated with Ebstein’s anomaly.

Key words: Pacemaker-induced cardiomyopathy

Case Report
A 76-year-old woman was admitted to our hospital with exacerbated palpitation and shortness of breath. At the age of 36 years, she had been diagnosed as Ebstein’s anomaly by echocardiogram, but she had rejected intracardiac repair because of no symptoms. A physical examination revealed a diastolic heart murmur on the apical area, swelling of the lower legs, and cyanosis on her extremities. An electrocardiography revealed a first-degree atrioventricular block and an alternating right and left bundle-branch block. A transthoracic echocardiogram showed displacement of septal and posterior tricuspid leaflet, an “atrialized” right ventricle, a dilated right atrium, and moderate tricuspid valve regurgitation (Figure 1A). The patent foramen ovale was not identified and her left heart ejection fraction (EF) was preserved (Figure 1B). In laboratory test, brain natriuretic peptide (BNP) increased to 1330 pg/mL. From these findings, she was diagnosed with exacerbated heart failure due to Ebstein’s anomaly. After her hospitalization, the addition of diuretics attenuated her symptoms of heart failure, and the level of BNP decreased to 50 pg/mL. PM (DDD 70/120 beats per minute) was implanted for her alternating bundle-branch block and she was discharged.

One year later, her heart failure worsened again and her LV ejection fraction was markedly declined to 20% (Figure 1C). Atrial fibrillation (AF) was observed and her BNP increased to 2137 pg/mL. Because AF might contribute to the worsening of heart failure and her reduced EF, a pulmonary vein isolation (PVI) was performed. After PVI, her BNP decreased to 578 pg/mL, and she was discharged.

However, PVI was not sufficient to stabilize her condition and her heart failure re-exacerbate with low cardiac...
output syndrome. The level of BNP increased to extremely high with 3007 pg/mL. EF remained to be low (27%). Her dependency of pacing was 63.7% at that time and PM-induced cardiomyopathy was considered to be a principal cause for exacerbating heart failure, and CRT device was implanted.

After the intervention, QRS duration on ECG was shortened from 169 ms to 140 ms, and her symptoms improved markedly. One month after CRT, BNP decreased to 484 pg/mL (Figure 2). Surprisingly EF was improved to 43% and the left ventricular end-diastolic diameter was reduced from 59 mm to 46 mm (Figure 1D, 2). This remarkable reverse remodeling suggested CRT improved her PM-induced cardiomyopathy in a dramatic way.

**Discussion**

Ebstein’s anomaly commonly presents with right-sided heart failure, however it is often complicated with left ventricular dysfunction. Attenhofer et al. reported that left-sided heart failure was complicated during the natural course with Ebstein’s anomaly, and several investigators reported abnormal left ventricular shape, contractility, and reduced EF in these patients.

LV dysfunction in Ebstein’s anomaly is partly explained by its vulnerable myocardium including noncompaction, myocardial fibrosis, disruption of free wall myofibril continuity, fiber geometry, or mixed cardiac defect. Besides, the dilated right ventricle itself has been thought to contribute to LV dysfunction, which is mostly observed in the presence of a significant left to right shunt congenital heart disease. However, Goleski et al. demonstrated that LVEF was not correlated to their LV shape, which was distorted by right-sided dilatation, in patients with Ebstein’s anomaly. Therefore, in patients with Ebstein’s anomaly, there may be discordant mechanisms for LV shape and function abnormalities.

On the other hand, reduced LVEF and the dependency of the pacing (over 40%) are known to be the risk factors for the progression of PM-induced cardiomyopathy. It is evident by previous clinical trials (PACE, BLOCK HF) that CRT should be recommended in the presence of these two risk factors. However, there is not enough data to recommend CRT in the setting of LV abnormalities with preserved EF. Khurshid et al. reported that CRT was appropriate for the prevention of PM-induced cardiomyopathy in a wide native QRS duration (>115 ms) irrespective of reduced LVEF. However, even
in the lacking of wide QRS or reduced EF, RV pacing is presumed to alter geometry and function of the vulnerable LV through electrical and mechanical dysynchrony, and an increase LV end diastolic volume, leading to the LV systolic dysfunction. In patients with Ebstein’s anomaly, these deteriorating effects of RV pacing on cardiac function might be enhanced and result in marked depression of LVEF even if they have preserved LVEF.

In this patient, RV pacing is 63.7% of the time and her left ventricular myocardium was presumably vulnerable due to Ebstein’s anomaly, even if she had normal EF. She could be in a high-risk group of PM-induced cardiomyopathy, and CRT may be effective for preventing cardiomyopathy.

In some reports left ventricular reverse remodeling was observed in patients with CRT, and patients with PM-induced cardiomyopathy responded favorably to upgrading from RV pacing to CRT. In another randomized study for patients who needed permanent pacemaker with mild reduced EF (40%), the CRT group had significant improvements in exercise capacity, quality of life, and LVEF compared with the RV pacing group. However, currently there is not enough data to recommend CRT for all patients who require PM support. Especially, in patients with Ebstein’s anomaly, there may be a high probability that deteriorating effects of RV pacing on cardiac function result in marked depression of LVEF and we need careful observation of it. More studies are needed to reach the conclusion about the decision criteria of CRT and define the patients at risk of PM-induced cardiomyopathy.

Disclosures

Conflict of interest: The authors declare that they have no conflict of interest.

References
