Case Report

A case report of successful atrioventricular junction pacing in Ebstein's anomaly

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Abstract: We report a case of atrioventricular junction (AVJ) pacing in a patient with Ebstein's anomaly (EA). The patient was a 68-year-old man who suffered from pacemaker syndrome and complained of heart failure symptoms. He was initially diagnosed with EA in his thirties and received right ventricular (RV) apex pacing for safe during a surgery because of low heart rate atrial fibrillation (AF) 9 years ago. However, since the patient felt discomfort, the pacing rate was then programed down to 45–55 per/min. During recent years, he was often admitted for dyspnea, dizziness, or edema and was advised to undergo intracardiac repair, but he rejected this due to the high risk of the surgery. We believed that the patient's low heart rate and ventricular pacing burden (47.8%) might be important causes of the symptoms. Therefore, we suggested that the patient undergo an upgrade of the pacing mode. In consideration of possible abnormal cardiac coronary veins, we tried His bundle pacing (HBP) to upgrade pacing. However, the SelectSecure 3830 lead was fixed at the AVJ region and obtained steady pacing parameters. After the upgrading of the AVJ pacing mode. The patient's symptoms, exercise capacity and quality of life were all improved at the 2-year follow-up. Thus, we presented the first case of AVJ pacing in a patient with EA.

Keywords: Ebstein's anomaly (EA); atrioventricular junction pacing (AVJ pacing); upgrade; case report

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Introduction

Due to anatomical variations of the tricuspid valves and right ventricle, permanent pacing in patients with Ebstein's anomaly (EA) presents a unique challenge. Despite this, His bundle pacing (HBP) is a safe and feasible pacing strategy in patients requiring long-term pacing to prevent ventricular dyssynchrony and left ventricular dysfunction (1). Selective and non-selective HBP have been reported recently in patients with EA (2,3). Our patient in this case study suffered from pacemaker syndrome after receiving right ventricular (RV) apex pacing. We tried to upgrade the RV apex pacing to HBP but failed. On the other hand a stable a-V wave which was proved later as the atrioventricular junction (AVJ) potential was recorded during the operation. We then screwed in the SelectSecure 3830 lead at the AVJ and achieved stable permanent pacing. Thus, we present the first case of AVJ pacing in a patient with EA.

We present the following article in accordance with the CARE reporting checklist (available at https://dx.doi.org/10.21037/apm-21-1678).

Case presentation

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this study and any accompanying images.

A 68-year-old Chinese male was admitted to our hospital for dizziness, shortness of breath, and swollen lower legs on December 25, 2018. During his thirties, he was diagnosed with heart disease during a physical examination, without any

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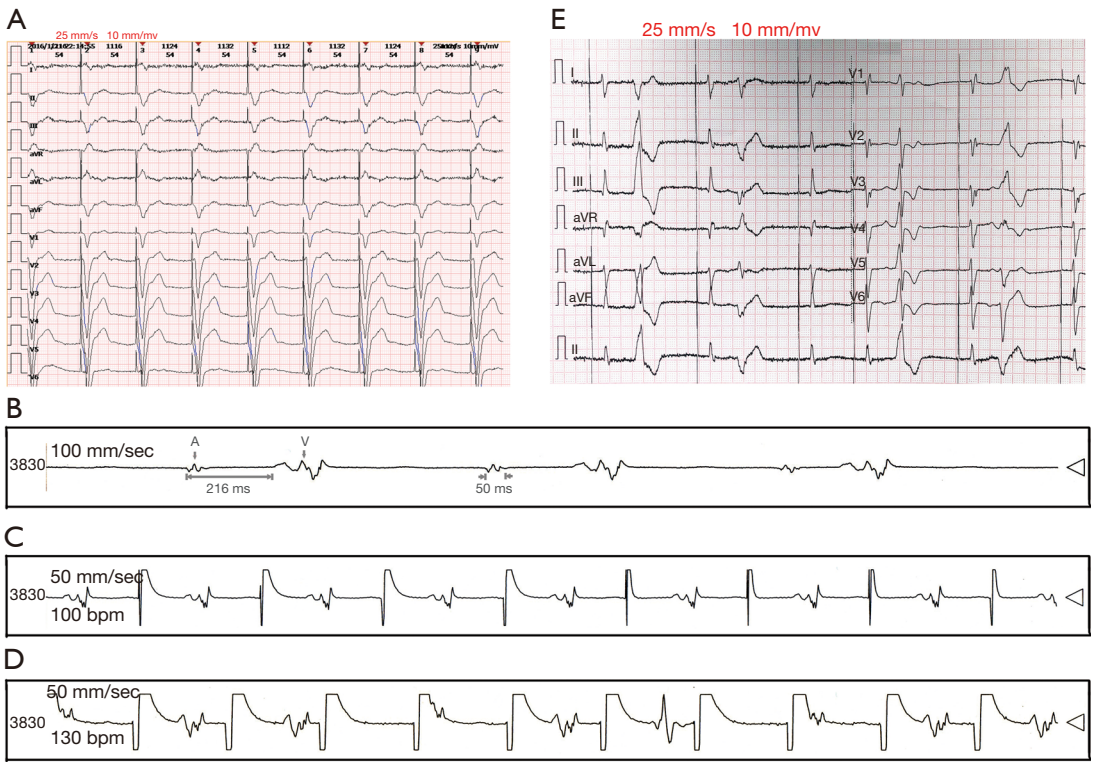

Figure 1 Electrophysiological record. (A) Ventricular apex pacing, QRS duration over 200 ms; (B) fixed rhythm, a-V interval 216 ms; (C) overdriving pacing at 100 per/min, signal conducted one by one; (D) overdriving pacing at 130 per/min, Wenckebach's phenomenon was found; (E) AVJ pacing, with QRS duration around 120 ms. AVJ, atrioventricular junction.

symptoms. Nine years ago, he was hospitalized in another hospital for a surgical operation and during that time he was diagnosed with EA with low heart rate atrial fibrillation (AF). Before the operation, a ventricular single chamber pacemaker was implanted for safety. The lead was implanted at the RV apex and heart rate was programed at 60 per/min. However, since the patient felt discomfort, the pacing rate was then programed down to 45–55 per/min. During recent years, he was often admitted for dyspnea, dizziness, or edema and was advised to undergo intracardiac repair, but he rejected this due to the high risk of the surgery. A physical examination revealed a systolic heart murmur in the apical area and swelling of the lower legs. Electrocardiogram (ECG) recorded a pacing rhythm with QRS duration over 200 ms (*Figure 1A*). Echocardiography once again demonstrated displacement of the septal and posterior tricuspid leaflet, an atrialized right ventricle, a dilated right atrium, severe tricuspid regurgitation, and preserved left ventricular ejection function (LVEF). Coronary angiography was performed to rule out coronary artery disease.

We believed that the patient's low heart rate and ventricular pacing burden (47.8%) might be important causes of the symptoms. Therefore, we suggested that the patient undergo an upgrade of the pacing mode. Cardiac resynchronization therapy (CRT) by HBP or a standard LV lead was considered. In consideration of possible abnormal cardiac coronary veins, we selected HBP as the first-line treatment. A SelectSecure 69 cm 3830 lead combined with a C315His sheath via the left subclavian was performed to detect H waves. We recorded clear "a" and V waves at the sites where H waves were assumed to be detected. We tried with great patience but found no clear H waves. On the hand, the "a" wave was quite stable and had a fixed rhythm of around 80 per/min. The a-V interval time was about 216 ms (*Figure 1B*). With pacing under 100 per/min, the signal could be conducted one by one to the ventricle, indicating acceptable conducting capacity (*Figure 1C*). The output rate was increased to 130 per/min, and Wenckebach's phenomenon was found (*Figure 1D*). The pacing threshold was excellent at 0.5 v/0.5 ms and the a-wave amplitude was

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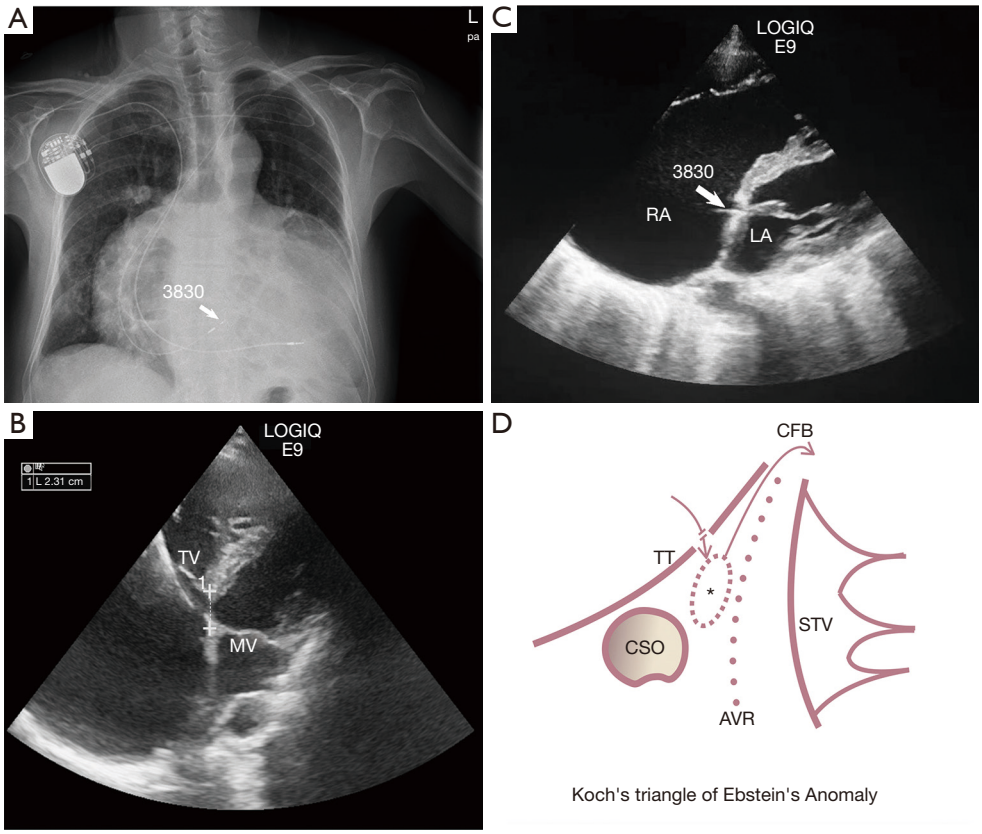

Figure 2 Lead location. (A) Chest radiograph recorded the leads and presented the large heart; (B) echocardiography showed the distance between the attachment points of MV and TV; (C) the tip of the 3830 lead was detected at the atrial side near the attachment point of MV; (D) imitate the Koch's triangle of the patient. Asterisk* means the pacing site. TT, Tendon of Todaro; STV, septal tricuspid valve; CSO, coronary sinus ostium; CFB, central fibrous body.

0.7 mV with a pacing impedance of 545 Ω, so we decided to choose it as our acceptable pacing site. A Medtronic dual chamber pacemaker REDr01 was connected with a 3830 lead at the atrial port and the old ventricular lead at the ventricular one. After implantation, we performed ECG again which showed similar AAI pacing at 60 per/min with formal complete right bundle branch block (CRBBB). ECG showed that the QRS duration was shortened from 200 to 120 ms (*Figure 1E*). A chest radiograph recorded the locations of the 2 leads and the large heart image (*Figure 2A*). Echocardiography before the operation showed the distance between the attachment points of MV and TV was 2.31 cm (*Figure 2B*). It detected the 3830 lead tip located at the atrial side near the septal mitral valve attachment point after the operation (*Figure 2C*).

The patient was followed up for 2 years and had stable AVJ pacing. Heart rate was programed at 70 per/min to maintain 100% pacing. The capture threshold increased slightly in the first month from 0.5 to 1.25 v/0.5 ms and remained steady. Impedance remained at around 450 Ω and amplitude remained low at 0.5–0.7 mv. After upgrading to AVJ pacing, the patient's symptoms, his exercise capacity and quality of life all improved markedly.

Discussion

Due to anatomical variations in the tricuspid valves and right heart chambers, permanent pacing can be quite difficult in EA (4). Studies have shown that stable long-term ventricular pacing can be achieved by positioning the lead either in the atrialized right ventricle, true right ventricle, or the cardiac venous system (5). Leads at the true right ventricle across the tricuspid valve can cause severe tricuspid regurgitation and may be the last option,

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causing pacing syndrome in this case. The cardiac venous system can be attempted with less stability because of the relatively high threshold, and sometimes the coronary sinus is inaccessible. The ventricular myocardium is present in the walls of the functional atrial chamber, thus, it can be used to obtain ventricular pacing with a screw-in lead without interfering with the tricuspid valve (6). HBP at a functional atrial chamber would hopefully achieve the best resynchronization, and was therefore our ideal option (7).

Studies on the bundle of His electrogram are rare. Kastor *et al.* reported the electrophysiological characteristics of 5 patients with EA (8). In the report, the bundle of His electrogram was recorded in its usual anatomical location. Infranodal conduction was prolonged in 4 cases (80%), with H-V intervals of 60, 65, 65, and 80 msec. In our case, a clear, stable a-V wave was recorded with intervals of 216 ms, which was much longer than the reported H-V intervals. The "a" wave morphology was also quite different from the typical H wave, and Wenckebach's phenomenon was recorded. Furthermore, late echocardiography detected the 3830 lead tip at the atrial side where no H wave could be recorded. Thus, we determined that the tip of the 3830 lead was located at the AVJ, as shown in *Figure 2D*. Overdrive pacing was used at 100 per/min to make sure it could be used. During the 2-year follow-up, the pacing parameters were kept stable, except for a slight increase in threshold.

Treatments for EA are quite complicated. Asymptomatic patients may be managed medically with observation for many years. Patients should be considered for operative intervention when they develop symptoms and/or worsening exercise capacity. Tricuspid valve repair is the goal of operative intervention; repair also typically includes RV plication, right atrial reduction, and atrial septal closure or subtotal closure. In this case due to the limitation of medical conditions the patient rejected the surgery. Around 3.7% patients with EA required permanent pacing for intrinsic conduction disease (4). Traditional ventricular apical pacing could aggravate heart function as recorded in this patient. HBP was advised to prevent ventricular dyssynchrony as reported (2,3). Through both HBP and AVJ pacing have the potential benefit of maintaining biventricular synchrony and preserving LV function, AVJ pacing is superior to HBP in consideration of long-term safety. We reported this case for ABJ pacing might be a supplement for HBP.

Conclusions

We presented a case of AVJ pacing in a patient with EA which achieved the best resynchronization and improved the patient's symptoms.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://dx.doi.org/10.21037/apm-21-1678

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://dx.doi.org/10.21037/apm-21-1678). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Written informed consent was obtained from the patient for publication of this study and any accompanying images. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013).

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