Ebstein Anomaly Associated With Unusual Conduction Delay and Sick Sinus Syndrome
—— A Case Report ——

Hiroki Mani, MD; Takeshi Shirayama, MD; Midori Yamamura, MD; Takashi Sakamoto, MD; Akiko Yoshida, MD; Masao Nakagawa, MD

A 64-year-old man was admitted to our hospital because of palpitation, dyspnea on effort, and facial edema. The echocardiographic diagnosis was Ebstein anomaly. Although the 12-lead electrocardiogram showed an atrial rate of 150 beats/min and no typical flutter wave, the electrophysiological study showed counterclockwise rotation of excitation along the tricuspid annulus. Because of sinus arrest and syncope, a permanent pacemaker was implanted, but the right atrium was not captured by electrical stimulation at 5V/0.4 ms, except for the orifice of coronary sinus, and the intracardiac P wave was only 0.2 mV or less. This is a rare case of Ebstein anomaly characterized by unusually prolonged conduction in the atrium, the basis of which was global myocardial damage, including the ventricles. (Circ J 2004; 68: 719–721)

Key Words: Common atrial flutter; Ebstein anomaly; Radiofrequency ablation; Sick sinus syndrome

Ebstein anomaly is characterized by apical displacement of the septal or posterior leaflet of the tricuspid valve and may be related to abnormal cell death. Furthermore, conduction abnormalities associated with this anomaly are often confined to the atrioventricular junction. We report a rare case of Ebstein anomaly associated with an unusual conduction delay in the whole heart, as well as systolic dysfunction of both ventricles, which indicates that the entire myocardium can be impaired globally in this anomaly.

Case Report

A 64-year-old man was admitted to our hospital because he had often felt palpitation and dyspnea on effort, and he had facial edema. On admission, the 12-lead electrocardiogram (ECG) revealed atrial tachycardia with 2:1 conduction and intraventricular conduction delay; his heart rate was 75 beats/min (atrial rate: 150 beats/min) (Fig 1). The cardiothoracic ratio was 61% on chest rentogenogram. Echocardiography showed severe enlargement of the right atrium and ventricle, decreased systolic function of both ventricles (end-diastolic and end-systolic left ventricular dimensions were 48 mm and 41 mm, respectively), apical displacement of the septal leaflet of the tricuspid valve by 16 mm (10.5 mm/m²), and mild tricuspid regurgitation with a peak velocity of 0.78 m/s and pressure gradient of 2.4 mmHg (Fig 2). The diagnosis was Ebstein anomaly.

An electrophysiological study revealed that the atrial rhythm was common atrial flutter. Atrial mapping showed a counterclockwise rotation of excitation along the tricuspid annulus; the atrial flutter cycle length was 400 ms (Fig 3). The post pacing interval after an extra-stimulation at the proximal coronary sinus and the low lateral right atrium was the same as the atrial flutter cycle length. Right atrial mapping showed that the slowest atrial conduction existed between RA3 and RA1 (Fig 3), located on the inferior vena cava–tricuspid annulus isthmus where low

Fig 1. 12-lead electrocardiogram on admission.
voltage and a fragmented atrial electrogram were recorded.

Linear catheter ablation was applied to the isthmus. When the atrial flutter was terminated after a 7-s application of radiofrequency current, sinus arrest occurred, so the catheter ablation procedure was continued under temporary VVI pacing. Bidirectional conduction block at the isthmus was established and atrial flutter was no longer inducible.

After catheter ablation, sinus rhythm appeared, but the heart rate was approximately 20–30 beats/min with His-ventricular (HV) prolongation (150 ms). On the 12-lead ECG, the P wave was wide (0.12 s) and biphasic in lead V1. On echocardiography, an atrial filling wave was detected (peak velocity: 0.64 m/s). Because the patient had a history of syncope and his maximal sinus node recovery time was 9.7 s, a permanent pacemaker was implanted. The best pacing site was carefully searched but the right atrium was not captured by electrical stimulation at 5 V/0.4 ms. The intracardiac P wave was only 0.2 mV or less. P wave sensing was not possible in any part of the right atrium other than the tricuspid annulus. Finally, an atrial pacing lead was anchored with a screw-in lead at the orifice of the coronary sinus, after which the atrial pacing threshold, intracardiac P wave, ventricular pacing threshold, and intracardiac R wave were 0.8 V/0.4 ms, 1.6 mV, 1.2 V/0.4 ms, and 3.6 mV, respectively.

**Discussion**

Ebstein anomaly is characterized by apical displacement of the septal or posterior leaflet of the tricuspid valve, resulting in 'atrialization' of the inflow tract of the right ventricle and consequently a variably small, but functional right ventricle. Varying degrees of tricuspid regurgitation result from this morphology with consequent right atrial enlargement. The diagnosis of this disease can often be made by echocardiography and apical displacement of the septal leaflet of the tricuspid valve by 8 mm/m² or more confirms the diagnosis. In the present case, the apical displacement was approximately 10.5 mm/m², which fulfilled the diagnostic criterion. Associated anomalies include atrial septal defect in approximately 50% of cases and accessory conduction pathways in 25%, but neither of these anomalies were observed in the present patient.

Atrial flutter has the appearance of a 'picket fence' wave with the rate ranging from 240 to 350 beats/min on the
12-lead ECG. The present case had an atrial rate of 150 beats/min and no typical flutter wave, so atrial tachycardia was suspected before the electrophysiological study. However, the study showed counterclockwise rotation of excitation along the tricuspid annulus within the right atrium, which was compatible with a diagnosis of common atrial flutter.

There are a few reports on Ebstein anomaly associated with atrial flutter. Okishige et al reported a case of Ebstein anomaly in which catheter ablation for common atrial flutter was performed; in that case, the atrial flutter cycle length was 312 ms and the flutter waves were wider and deeper on the 12-lead ECG. The unusually slow conduction in the right atrium of the present patient may have been caused by changes in the myocardium, such as fibrosis and dilatation, and the atypical flutter wave possibly resulted from the enlargement and distortion of the atria.

On the other hand, atrioventricular block is a common bradyarrhythmia in Ebstein anomaly, but sick sinus syndrome is rare. Noda et al reported a case of Ebstein anomaly with sick sinus syndrome and atrioventricular block in which the sinus node recovery time was 13 s and the right atrium was not captured by electrical stimulation at 10 V, except in the low septal right atrium and the coronary sinus. In that case a permanent pacemaker with a VVI mode was implanted. In the present case the right atrium was not captured at 5 V/0.4 ms, except at the orifice of the coronary sinus, and a screw-in-lead was necessary. The electrical activity of the right atrium was severely deteriorated and reduced to very low amplitude (<0.2 mV), except for the tricuspid annulus. There may have been partial atrial standstill because electrical activity hardly occurred in some parts of the atrium (such as RA2 in Fig 3). In addition, the P wave was wide and HV prolongation was apparent, which was indicative of intraatrial and His–Purkinje conduction delay.

Clark speculated that Ebstein anomaly may be related to abnormal cell death, and Celermajer et al demonstrated gross and microscopic abnormalities, with increased fibrous content of both ventricles, in neonatal hearts with Ebstein anomaly. These changes supported the concept of a genetic basis for abnormal cell death and a contribution to the development of arrhythmias. In the present case, not only sinus node function but conduction in the right atrium and His–Purkinje had deteriorated and moreover, the systolic function of both the right and left ventricles was decreased. Abnormal cell death may be the cause of this global cardiac abnormality.

In summary, we report a rare case of Ebstein anomaly characterized by unusually prolonged conduction in the atrium, caused by global myocardial damage, with associated common atrial flutter and sick sinus syndrome. The amplitude of the intraatrial electrogram was very low or undetectable, and the systolic function of both ventricles was impaired.

References