CASE REPORT

Atrioventricular nodal re-entrant tachycardia in a patient with Ebstein anomaly

G Mönnig, K Wasmer, L Eckardt

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The first case of successful slow pathway modulation is reported in a patient with Ebstein anomaly and recurrent atrioventricular nodal re-entrant tachycardia. Typical signals were recorded during electrophysiological study at the slow pathway region between the His bundle and the coronary sinus where ablation was performed successfully. Thus, slow pathway modulation seems to be a safe procedure even in selected patients with Ebstein anomaly.

A 31 year old woman with Ebstein anomaly was admitted for recurrent tachycardia associated with dyspnoea. She was referred because of an increasing frequency of

episodes, which were terminated by intravenous verapamil. She had documented episodes of supraventricular tachycardia with a heart rate of 180 beats/min without visible P waves. She presented with classic ECG characteristics of Ebstein anomaly such as right bundle branch block and right atrial hypertrophy and a normal PR interval. The tricuspid valve was greatly displaced to the ventricle (fig 1B, C). During the electrophysiological study, an accessory pathway was excluded and programmed atrial stimulation established the presence of dual atrioventricular nodal physiology. An atrioventricular nodal re-entrant tachycardia (cycle length 380 ms) was easily inducible by single atrial extrastimuli (fig 1A). Slow pathway modulation (4 mm tip catheter, 50 W, 60 s) was successfully performed. The patient remained free of symptoms during the follow up of 12 months.

Figure 1 Surface and intracardiac electrograms, and fluoroscopic view in a patient with Ebstein anomaly and atrioventricular nodal re-entrant tachycardia. (A) Recorded at 50 mm/s paper speed. Atrioventricular nodal re-entrant tachycardia was reproducibly induced by single atrial extrastimuli and identified as clinical tachycardia by the patient. Retrograde P waves can be seen in lead III (asterisks), which were missing during sinus rhythm (arrow). H, His bundle potential. Right ventricular angiogram is shown in (B) right anterior oblique 30° and (C) left anterior oblique 60° views. Of note is the small functional right ventricle, with displacement of the tricuspid leaflet attachments towards the right ventricular apex (arrow heads), as well as moderate degree of tricuspid regurgitation. The position of the ablation catheter is shown in (D) right anterior oblique 30° and (E) left anterior oblique 60° views. The inset shows the local signal at the successful site. Quadripolar catheters in the high right atrium and at the bundle of His, and a bipolar catheter in the right ventricular apex are also shown. Remarkably, typical signals were recorded at the slow pathway region where ablation was performed successfully.

DISCUSSION

The condition, which bears Ebstein's name, is characterised by an abnormality of the tricuspid valve in which the septal leaflet and often the mural (posterior) leaflet attachment are displaced from the atrioventricular ring into the right ventricular cavity towards the apex. The association of supraventricular tachycardia and Ebstein anomaly is common. Bidirectional accessory pathways and associated Wolf-Parkinson-White syndrome have been reported to be present in about 20–30% of these patients.¹ The associated atrioventricular reciprocating tachycardia can be life threatening. Rossi and colleagues² reported three cases of sudden cardiac death caused by supraventricular tachycardia in patients with Ebstein anomaly. Radiofrequency ablation is widely accepted as first line treatment for atrioventricular reciprocating tachycardia in these patients.³ To the best of our knowledge this is the first report of a successful slow pathway modulation in a patient with Ebstein anomaly and recurrent atrioventricular nodal re-entrant tachycardia.

Structural and anatomical abnormalities have been described in patients with Ebstein anomaly. Ho and colleagues⁴ examined the histology of five hearts with Ebstein anomaly. In three hearts, the compact atrioventricular node was closer to the coronary sinus ostium than in normal hearts. Therefore, the triangle of Koch may be smaller in Ebstein anomaly than in normal hearts.⁴ This may be why atrioventricular nodal re-entrant tachycardia has not been reported so far and is unlikely to occur in patients with Ebstein anomaly and a small triangle of Koch. In contrast to the anatomy described in the literature, we found a normal distance between the compact atrioventricular node and the coronary sinus ostium in the patient described here. Typical signals were recorded at the slow pathway region between

the His bundle and the coronary sinus (fig 1D, E) where ablation was performed successfully. Thus, slow pathway modulation seems to be a safe procedure even in selected patients with Ebstein anomaly.

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Authors' affiliations

G Mönnig, K Wasmer, Department of Cardiology and Angiology, Hospital of the University of Münster, Münster, Germany
L Eckardt, Institute for Arteriosclerosis Research, University of Münster, Münster, Germany

Correspondence to: Dr G Mönnig, Medizinische Klinik und Poliklinik C–Kardiologie und Angiologie, Universitätsklinikum Münster (UKM), D-48129 Münster, Germany; moennig@uni-muenster.de

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REFERENCES

- 1 **Smith WM**, Gallagher JJ, Kerr CR, *et al*. The electrophysiologic basis and management of symptomatic recurrent tachycardia in patients with Ebstein's anomaly of the tricuspid valve. *Am J Cardiol* 1982;**49**:1223–34.
- 2 **Rossi L**, Thiene G. Mild Ebstein's anomaly associated with supraventricular tachycardia and sudden death: clinicomorphologic features in three patients. *Am J Cardiol* 1984;**53**:332–4.
- 3 **Cappato R**, Schluter M, Weis C, *et al*. Radiofrequency current catheter ablation of accessory atrioventricular pathways in Ebstein's anomaly. *Circulation* 1996;**94**:376–83.
- 4 **Ho SY**, Goltz D, McCarthy K, *et al*. The atrioventricular junctions in Ebstein malformation. *Heart* 2000;**83**:444–9.