

Electrophysiological properties of dual atrioventricular nodes in patients with right atrial isomerism

Department of Paediatrics, Surgery and Internal Medicine, National Taiwan University, Taipei, Taiwan

M-H Wu
J-L Lin
J-K Wang
I-S Chiu
M-L Young

Mei-Hwan Wu, Jiunn-Lee Lin, Jou-Kou Wang, Ing-Sh Chiu, Ming-Lon Young

Abstract

There are reports that in right atrial isomerism the conduction system has paired sinus nodes and paired atrioventricular nodes. Electrophysiological studies were performed in two patients with right atrial isomerism. One patient had a delta wave on the surface electro-

cardiogram without tachycardia attacks. The other, who did not have manifest pre-excitation, had recurrent narrow QRS tachycardia. Electrophysiological studies suggested the presence of dual atrioventricular nodes. Only unidirectional atrioventricular or ventriculoatrial conduction was demonstrated for these dual atrioventricular nodes even after infusion of isoprenaline. It is suggested that unidirectional conduction may be a common property of the dual atrioventricular nodes in right atrial isomerism and that the absence of retrograde ventriculoatrial conduction protects the patients against tachycardia.

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Right atrial isomerism is usually associated with complex cyanotic congenital heart disease.¹ The conduction system had been reported to have paired sinus nodes and dual atrioventricular (AV) nodes.^{2,3} However, the electrophysiological properties, to the best of our knowledge, have not been reported. We report two patients with asplenia, right atrial isomerism, total anomalous pulmonary venous return, complete atrioventricular canal, double outlet right ventricle, and pulmonary stenosis. One had a delta wave on the surface electrocardiogram without tachycardia attacks. The other, who did not have manifest preexcitation, had recurrent narrow QRS tachycardia. Electrophysiological studies suggested the presence of dual AV nodes.

Case reports

CASE 1

In an 8 year old boy who had been cyanosed since early infancy echocardiography and cardiac catheterisation showed right atrial isomerism, total anomalous pulmonary venous return draining back to right superior vena cava without obstruction, complete atrioventricular canal, double outlet right ventricle, and pulmonary stenosis. Additionally, a delta wave with a short PR interval was found on one electrocardiogram taken during follow up. None the less, he had never had tachycardia attacks.

Electrophysiological study was performed before the total cavopulmonary anastomosis

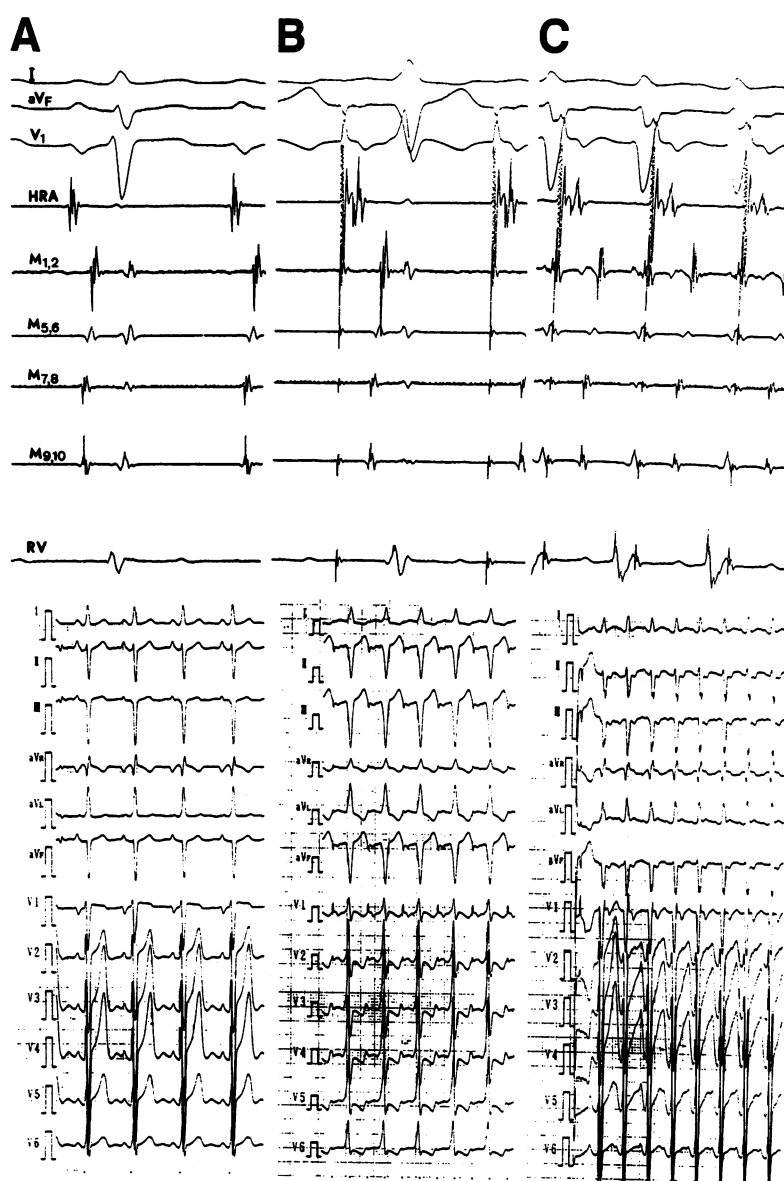


Figure 1 Electrocardiograms recorded at (A) rest: atrioventricular conduction through the posterior atrioventricular node with a narrow QRS complex and rS pattern in V1. Electrocardiograms recorded during right atrial pacing at 520 ms (B): pre-excitation was evident from the short PR interval and delta wave. The polarity of delta wave and QRS complex suggested a left posterior accessory pathway. Electrocardiograms recorded during right atrial pacing at 340 ms (C) showing atrioventricular conduction through an anterior node with a narrow QRS complex and a QS pattern in V1.

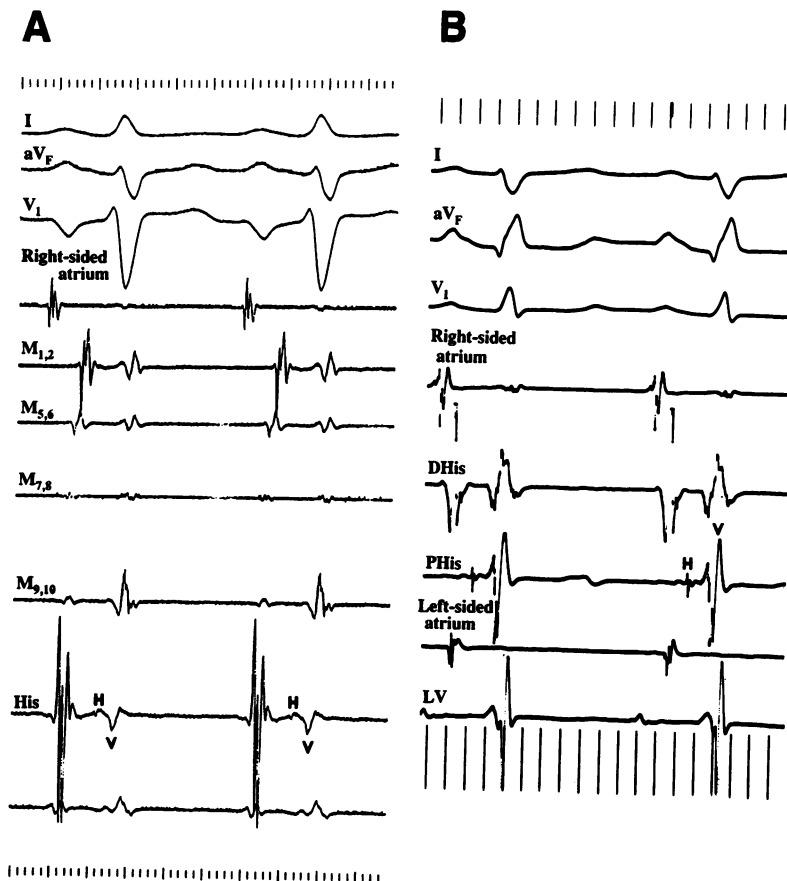


Figure 2 His potential recorded near the posterior atrioventricular node in case 1 (A) and case 2 (B).

operation. The intracardiac recording showed that resting cardiac rhythm originated from the right-sided sinus node and was conducted through a posterior AV node, which resulted in an rS pattern in V1 (fig 1A). When right atrium was paced at cycle lengths of 560–360 ms, the paced rhythm preexcited the ventricle through an accessory pathway. The earliest ventricular depolarisation was recorded from the left-sided recording electrodes, and the 12 lead electrocardiogram suggested an accessory pathway in a left posterior site (fig 1B). When the paced cycle length was reduced to 340 ms, the AV conduction shifted to the anterior AV node, as suggested by a QS pattern in V1 and normal PR (AV) interval (fig 1C). The Wenckebach cycle length of the anterior AV node was 290 ms. Right ventricular pacing produced only ventriculoatrial dissociation even after an intravenous infusion of isoprenaline had been given to raise the heart rate to 170/min. Atrial and ventricular extrastimuli did not induce any atrioventricular reentrant tachycardia. Short-run atrial flutter-fibrillation only was induced by using S_1S_1/S_1S_2 at 500/210 ms: the shortest RR interval was 440 ms. The effective refractory period of the anterior AV node was limited by an atrial refractory period of 200 ms. His potential during normal sinus rhythm was recorded near the posterior AV node (fig 2A). Because no tachycardia was induced and the risk of the accessory pathway was deemed to be low, no

further electrophysiological intervention was proposed. Later he underwent a total cavopulmonary anastomosis with an uneventful course.

CASE 2

A 4 year old cyanotic boy had had tachycardia for 3 years. Echocardiography and cardiac catheterisation showed right atrial isomerism, total anomalous pulmonary venous return to a right-sided atrium without obstruction, complete atrioventricular canal, double outlet right ventricle and pulmonary stenosis. An electrocardiogram recorded during an episode of tachycardia showed narrow QRS tachycardia (long PR, short RP) which could be cardioverted by ice bag immersion (fig 3). Alternating forms of the P wave were seen on resting electrocardiograms, and the QRS axis was not changed either during the normal sinus rhythm or rapid atrial pacing. During the electrophysiological study constant QRS configuration during rapid atrial pacing and a continuous $A_2H_2 v A_1A_2$ curve suggested that anterograde AV conduction occurred through the posterior AV node only and not the anterior AV node. His potential was recorded near the posterior AV node (fig 2B). After the earliest atrial depolarisation was mapped during tachycardia and ventricular pacing (by electrophysiological study and intracardiac mapping), the clinical tachycardia was attributed to a reentrant circuit conducting anterogradely through the posterior AV node and retrogradely through the anterior AV node. Ventricular pacing and retrograde ventriculoatrial conduction during tachycardia constantly depolarised the atria from the left anteroseptal area (anterior AV node horizon). Therefore, we concluded that the anterior AV node had the property of unidirectional retrograde ventriculoatrial conduction whereas the posterior AV node could conduct only unidirectionally, from the atria to the ventricles. Subsequently, the anterior AV node was surgically ablated during the total cavopulmonary anastomosis operation. The patient died of sepsis 2 weeks after the operation.

Discussion

Dickinson *et al* histologically studied the distribution of the conduction system in four patients with right atrial isomerism. There were dual AV nodes.² The posterior AV node was constant in position, lying close to the tendon of Todaro. In patients with two ventricles, the second AV node was anterior in position. A sling of specialised conducting tissue was found at the inferior rim of the ventricular septal defect and in most of the cases made contact with both AV nodes. Ho *et al* reported similar findings.³ However, the electrophysiological properties are still far from clear. The presence of a sling of specialised conducting tissue between the dual AV nodes clearly provides a possible reentrant circuit. We found unidirectional conduction (atrioventricular or ventriculoatrial) in both these dual AV nodes.

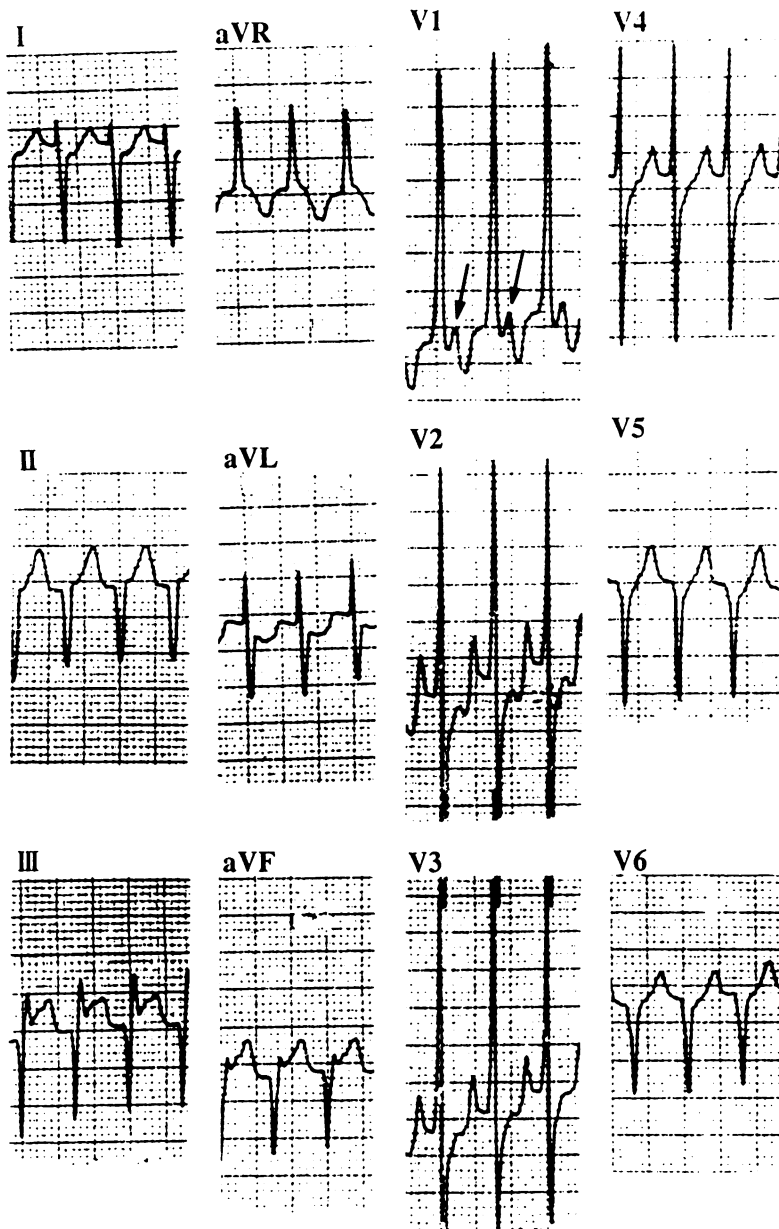


Figure 3 Narrow QRS tachycardia at a cycle of 280 ms with a long PR and short RP. Arrows indicate retrograde P wave.

The occurrence of AV reentrant tachycardia depends on the direction of unidirectional conduction in the anterior AV node. AV reentrant tachycardia is not common in patients with right atrial isomerism.⁴ Therefore, we speculate that unidirectional conduction may be a common property of the dual AV nodes in right atrial isomerism and that an absence of retrograde ventriculoatrial conduction protects the patients against tachycardia.

The conduction intervals and the refractory periods of the dual AV nodes were all normal for the age of the patient, nonetheless the properties of the anterior and posterior AV nodes were different. Such differences could be the result of different intrinsic properties or different innervation states in the two nodes. Autonomic blockade by atropine and propranolol may help to make the distinction.⁵ However, the possibility that pulmonary flow may increase after the relief of infundibular stenosis by propranolol and may unmask pulmonary venous obstruction should be considered.⁶

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