Successful demonstration of the detailed connection between the twin atrioventricular nodes and sling in a patient with asplenia syndrome

Hitoshi Mori, MD,* Naokata Sumitomo, MD, PhD,* Syota Muraji, MD,* Tomohiko Imamura, MD,* Toshiki Kobayashi, MD, PhD,* Ritsushi Kato, MD, PhD†

*From the *Department of Pediatric Cardiology, Saitama Medical University International Medical Center, Saitama, Japan, and †Department of Cardiology, Saitama Medical University International Medical Center, Saitama, Japan.*

Introduction

Two distinct twin atrioventricular (AV) nodes and a connecting sling could be confirmed in a number of patients with asplenia syndrome.[1–4] Atrioventricular reciprocating tachycardia (AVRT) via twin AV nodes can develop in some patients, and radiofrequency catheter ablation (RFCA) to interrupt or modify 1 of the 2 AV nodes that does not perform the main conduction has been attempted in tachycardia cases.[5,6] A pathologic analysis of autopsy specimens has demonstrated 2 separate AV nodes and a connecting sling.[3,4] There have been several hypotheses on the anatomic and electrophysiological characteristics of the AV nodes and connecting sling[4–6]; however, a detailed electrophysiological mechanism remains unknown. We report the case of a patient with AVRT via twin AV nodes after a Fontan procedure who underwent successful RFCA for 1 of the 2 separate AV nodes, and we reveal the precise connection between the AV nodes and connecting sling.

Case report

An 18-year-old man was referred to our hospital for RFCA of paroxysmal supraventricular tachycardia (PSVT). He had been diagnosed with asplenia, a complete endocardial cushion defect, hypoplastic right ventricle, partial anomalous pulmonary venous connection, and right aortic arch just after birth. An extracardiac total cavopulmonary connection with a tissue-engineered graft was performed at age 2 years 10 months. PSVT was first documented when he was 13 years old. RFCA for the PSVT was performed previously at another hospital. The tachycardia showed a centrifugal pattern near the 1 o'clock position on the valve annulus, and successful RFCA was performed at the origin of the tachycardia. Atrial stimulation showed 2 different QRS morphologies that may have suggested twin AV nodes. No tachycardia could be induced after RFCA, and no additional RFCA for the twin AV nodes was performed. However, the frequency of the palpitations recently increased, so he was referred to our hospital for RFCA.

The baseline electrocardiogram showed left-axis deviation. After written informed consent was obtained from the patient and his parents, an electrophysiological study was performed. A steerable 20-pole circular mapping catheter was positioned in the extracardiac conduit, and a 4F 4-pole electrode catheter was placed into the single ventricle. The ablation catheter was advanced into the single atrium via a retrograde transaortic approach under fluoroscopic guidance. Electrophysiological study with atrial pacing using a Lasso catheter revealed 2 different QRS morphologies that suggested twin AV nodes. Ventriculoatrial (VA) conduction was confirmed by ventricular burst pacing and was interrupted by a bolus injection of adenosine triphosphate, which may have proven that VA conduction was through the AV node. During atrial extrastimulus pacing, no AH jump-up phenomenon was observed. PSVT (cycle length 341 ms) was repeatedly induced by a triple atrial extrastimuli method (450-350-350-210 ms). The QRS morphology during PSVT had exactly the same pattern as that during sinus rhythm. During the tachycardia, the QRS morphology changed from a right bundle branch block pattern to a left bundle branch block pattern, without any change in tachycardia cycle length. This suggested that the ventricle was not included in the circuit of this tachycardia. AV conduction time shortened with the change in QRS morphology from a right bundle branch block pattern (conduction time 161 ms) to a left bundle branch block pattern (conduction time 127 ms) (Figure 1). However, AV conduction time did not shorten with the change from the complete left bundle branch block pattern to the incomplete left bundle branch block pattern; therefore, the left bundle branch did not contribute to AV conduction of this

KEYWORDS Asplenia syndrome; Catheter ablation; Congenital heart disease; Fontan operation; Supraventricular tachycardia; Twin atrioventricular node
(Heart Rhythm Case Reports 2018;4:480–483)

Address reprint requests and correspondence: Dr Naokata Sumitomo, Department of Pediatric Cardiology, Saitama Medical University International Medical Center, 1397-1 Yamane, Hidaka, Saitama 350-1298, Japan. E-mail address: sumitomo@saitama-med.ac.jp.

https://doi.org/10.1016/j.hrcr.2018.07.007

KEY TEACHING POINTS

- Macroreentrant atrioventricular (AV) tachycardia is sometimes observed in patients with asplenia syndrome, and the supraventricular tachycardia could be fatal in some patients after a Fontan operation.
- The detailed electrophysiological characteristics of the twin AV nodes and connecting sling in patients with asplenia syndrome are unknown.
- This report revealed the precise connection between the AV nodes and the connecting sling.

tachycardia (Figure 1). AV conduction time during sinus rhythm increased from 181 to 307 ms under atrial burst pacing, with a change in QRS morphology from a normal QRS to a right bundle branch block pattern. This suggested that the right bundle branch contributed to AV conduction in this patient. The earliest atrial activation site was at the 4 o'clock position on the valve annulus during the tachycardia (Figure 2A and B). The His potential was recorded during sinus rhythm at the earliest atrial activation site during the tachycardia, and the activation pattern on the Lasso catheter during ventricular pacing was the same as that during the tachycardia. Therefore, we confirmed retrograde conduction of this tachycardia was through the posterior AV node. QRS morphology changed from left-axis deviation to right-axis deviation during the atrial stimulation nadir of the posterior AV node. Therefore, the posterior AV node did not contribute to AV conduction during sinus rhythm. We did not confirm a dual AV nodal physiology of the posterior AV node. However, we believed that this tachycardia was not the AV nodal reentrant tachycardia of the posterior node because anterograde conduction of this tachycardia was through the anterior node. Unfortunately, the His potential of the anterior AV node could not be recorded. The earliest ventricular activation was at the 10 o'clock position on the valve annulus during atrial stimulation (Figure 2C and D). There was no difference in QRS morphology between sinus rhythm and atrial stimulation from the 10 o'clock position on the valve annulus. The anterior AV node should be located at the 10 o'clock position on the valve annulus, and the anterior AV node should provide anterograde conduction during sinus rhythm and the tachycardia. Therefore, we confirmed that anterograde conduction of this PSVT was via the anterior AV node and that retrograde conduction of this PSVT was via the posterior AV node. Radiofrequency current (30 W, 60 seconds) was delivered at the posterior AV node where the His potential was recorded (Figure 2E and F). VA conduction completely disappeared, and no additional tachycardia was induced after RFCA without any AV conduction disturbance. During 8-month follow-up, the patient did not experience any recurrence of PSVT.

Discussion

Macroreentrant atrial tachycardia is sometimes observed after a Fontan operation, and the atrial tachycardia can be fatal in some patients after a Fontan operation.[7] In addition, twin AV nodes have been confirmed in some patients with heterotaxy syndrome, and AVRT using twin AV nodes develops in some patients.[5,6] Severe interventricular dyssynchrony could be induced by a unilateral AV node modification after the Fontan operation,[8] so consideration of the conduction route to the ventricles in patients with twin AV nodes is important. There have been several hypotheses about the anatomic and electrophysiological characteristics of the 2 separate AV nodes and the connecting sling in heterotaxy syndrome. Although 2 separate AV nodes, a connecting sling, and the conduction tissue have been identified by pathologic analysis of autopsy specimens, previous reports have only demonstrated the presence of a rapid conducting bundle between the 2 AV nodes by electrophysiological analysis. No detailed electrophysiological studies on the relationship among the twin AV nodes, connecting sling, and conduction tissue have been reported. Our patient exhibited a change from a right bundle branch block pattern to a left bundle branch block pattern, without any change in tachycardia cycle length, which suggested that the ventricle was not involved in the tachycardia circuit. This finding indicated that twin AV nodes could be directly connected by a sling between each of them, without a ventricular connection. Each AV node was directly connected by the sling, but AV conduction was through the right bundle branch, which diverged from the nadir to the sling close to the anterior AV node (Figure 2G). During right bundle branch block, AV conduction was through the left bundle branch nadir to the posterior AV node via the connecting sling, and conduction time was prolonged (Figure 2H). However, AV conduction time did not change during the change from incomplete left bundle branch block to complete left bundle because the left bundle branch did not contribute to AV conduction.

Differentiation between this tachycardia and a supraventricular tachycardia due to an atriofascicular fiber might be difficult. Usually, anterograde conduction of a supraventricular tachycardia due to an atriofascicular fiber is via the atriofascicular fiber, and retrograde conduction is via the AV node. Anterograde conduction of this tachycardia was through the AV node. Furthermore, there was no preexcitation during anterograde conduction of the posterior AV node, and the tachycardia may be terminated when there is a proximal site of an ipsilateral bundle branch block of the atriofascicular connection. However, this tachycardia continued when there was a distal site of the bundle branch

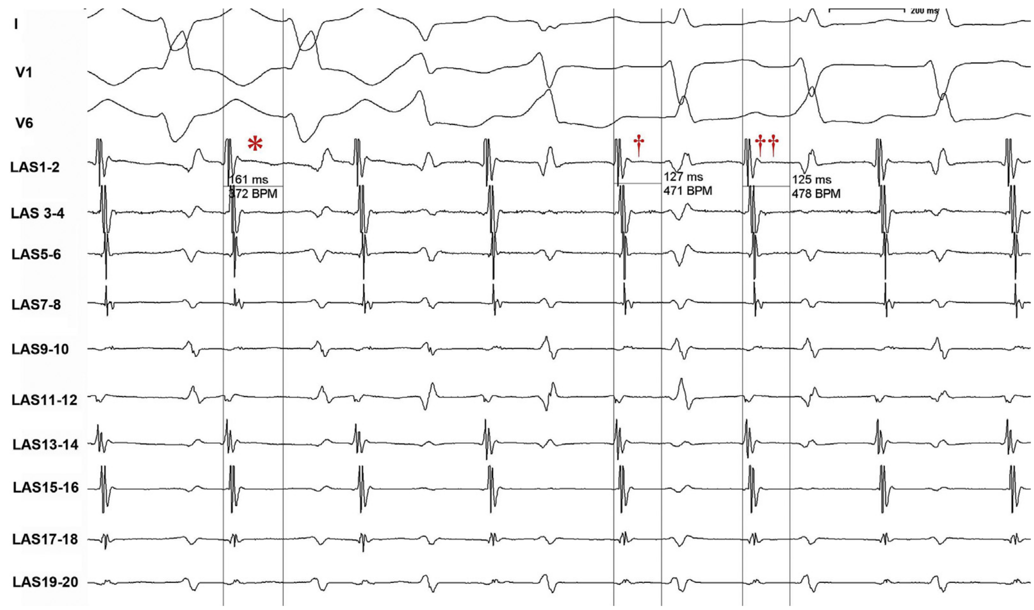

Figure 1 QRS morphology changed from a right bundle branch block pattern to a left bundle branch block pattern, without any change in tachycardia cycle length. The atrioventricular conduction time shortened with the change in QRS morphology from a right bundle branch block pattern (*) to a left bundle branch block pattern (†). However, the atrioventricular conduction time did not shorten with the change from the complete left bundle branch block pattern to the incomplete left bundle branch block pattern (††).

block of the atriofascicular connection. Therefore, we believe this tachycardia was an AV tachycardia using the 2 AV nodes. However, we cannot completely exclude the possibility of anterior conduction of this tachycardia via the anterior AV node and retrograde conduction via the atriofascicular fiber.

Conclusion

There are several hypotheses about the anatomic and electrophysiological characteristics of the twin AV nodes and the connecting sling. This report revealed the precise connection between the AV nodes and connecting sling.

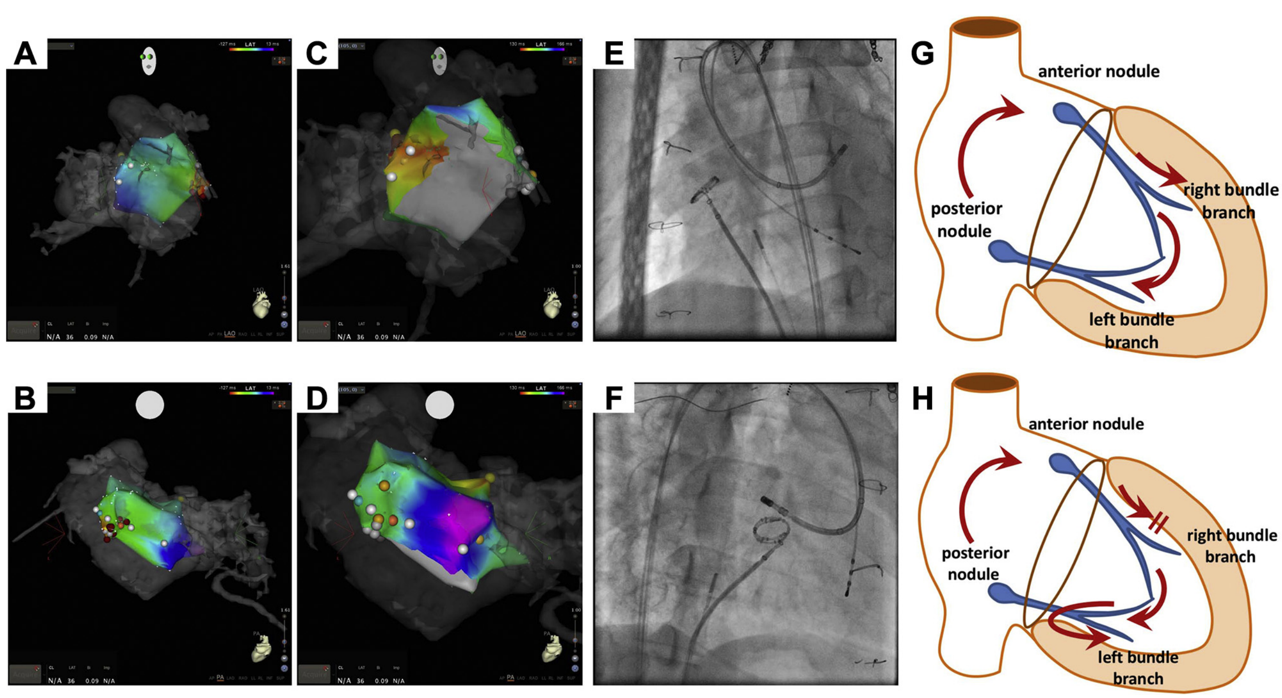

Figure 2 **A, B:** Earliest atrial activation site of the tachycardia was observed at the 4 o'clock position on the valve annulus. **C, D:** The Stim-QRS interval was recorded, and the interval was shortest at the 10 o'clock position on the valve annulus. **E, F:** Catheter ablation was performed at the posterior atrioventricular (AV) node. **G:** The twin AV nodes were directly connected, and AV conduction through the right bundle branch diverged from the sling nadir to the anterior AV node. **H:** During right bundle branch block, AV conduction was through the left bundle branch nadir to the posterior AV node via the connecting sling, and the conduction time prolonged.

References

1. Wu MH, Wang JK, Lin JL, Lai LP, Lue HC, Young ML, Hsieh FJ. Supraventricular tachycardia in patients with right atrial isomerism. J Am Coll Cardiol 1998; 32:773–779.
2. Wu MH, Lin JL, Wang JK, Chiu IS, Young ML. Electrophysiological properties of dual atrioventricular nodes in patients with right atrial isomerism. Br Heart J 1995; 74:553–555.
3. Dickinson DF, Wilkinson JL, Anderson KR, Smith A, Ho SY, Anderson RH. The cardiac conduction system in situs ambiguus. Circulation 1979;59:879–885.
4. Smith A, Ho SY, Anderson RH, Connell MG, Arnold R, Wilkinson JL, Cook AC. The diverse cardiac morphology seen in hearts with isomerism of the atrial appendages with reference to the disposition of the specialised conduction system. Cardiol Young 2006;16:437–454.
5. Bae EJ, Noh CI, Choi JY, Yun YS, Kim WH, Lee JR, Kim YJ. Twin AV node and induced supraventricular tachycardia in Fontan palliation patients. Pacing Clin Electrophysiol 2005;28:126–134.
6. Epstein MR, Saul JP, Weindling SN, Triedman JK, Walsh EP. Atrioventricular reciprocating tachycardia involving twin atrioventricular nodes in patients with complex congenital heart disease. J Cardiovasc Electrophysiol 2001; 12:671–679.
7. Nakagawa H, Shah N, Matsudaira K, et al. Characterization of reentrant circuit in macroreentrant right atrial tachycardia after surgical repair of congenital heart disease. Circulation 2001;103:699–709.
8. Sakaguchi H, Miyazaki A, Ohuchi H, Kagisaki K. Interventricular dyssynchrony due to unilateral atrioventricular conduction block in a patient with right atrial isomerism and twin atrioventricular nodes. Heart Rhythm 2011;8:1072–1075.