Wolff-Parkinson-White (WPW) Syndrome in Isolated Noncompaction of the Ventricular Myocardium (INVM)

— Three Cases —

Koichi Nihei, MD; Noriaki Shinomiya, MD; Hirohiko Kabayama, MD;
Chikako Ikeda, MD; Toshihiko Hosono, MD;
Tsugutoshi Aoki, MD; Norio Matsuo, MD

Isolated noncompaction of the ventricular myocardium (INVM) is a rare morphological abnormality caused by cessation of the compaction of the loose interwoven meshwork of myocardial fibers during intrauterine life. Three cases of INVM, including 2 siblings, were diagnosed from 2-dimensional echocardiographic findings of Wolff-Parkinson-White syndrome type B to which INVM can be attributed. (*Circ J* 2004; 68: 82–84)

Key Words: Cardiomyopathy; Isolated noncompaction of the ventricular myocardium; Right anteroseptal pathway; Wolff-Parkinson-White syndrome type B

Isolated noncompaction of the ventricular myocardium (INVM) is a rare disorder characterized by excessively prominent trabeculations and deep intertrabecular recesses, mainly in the left ventricle without other congenital cardiac malformations.¹ Most patients with INVM show various abnormalities on the electrocardiogram (ECG), including Wolff-Parkinson-White (WPW) syndrome,^{2,3} which is currently classified into several types based on the localization of the bypass tract. It has not previously been reported which type of WPW syndrome occurs in patients with INVM.^{3,4} We report 3 patients with INVM and WPW syndrome type B on ECG, which has not been previously reported.^{3,4}

Case Reports

Patient 1

A 9-month-old Japanese girl was admitted to hospital because of increasing dyspnea. She was the first child of healthy nonconsanguineous parents, and the pregnancy and delivery had been uneventful.

On admission, she was thin and malnourished: weight 6 kg and height 56 cm, both below the 10th percentile. She was pale and had cyanosis of the lips and nail bed. On auscultation, there was tachypnea and moderate retraction of the chest wall with moist rales over the entire lung field. A grade 3 pansystolic murmur was noted at the apex. Her abdomen was moderately distended, with the liver palpable 3 cm below the right costal margin. Chest X-ray showed marked cardiac enlargement, and pulmonary venous

congestion. She died suddenly at 11 months of age.

Patient 2

The 6-month-old younger sister of Patient 1 was referred for examination of her cardiac function. The pregnancy and delivery had been uneventful and she was currently alert and healthy. Her heart sounds were clear and the rhythm was regular without murmur.

Chest X-ray showed mild cardiac enlargement with a cardiothoracic ratio of 0.61. One year later, she still did not have symptoms, but her cardiac performance had deteriorated slightly, with a left ventricular fractional shortening (LVFS) of 24% and an ejection fraction (LVEF) of 57%. She was treated with oral angiotensin-converting enzyme inhibitor.

Patient 3

A 9-year-old girl was referred for an outpatient examination of an arrhythmia diagnosed previously during a school physical examination. She was alert and healthy with clear heart sounds and no murmur. No cardiac enlargement was noted on the chest roentgenogram.

Electrocardiography, Echocardiography and Magnetic Resonance Imaging

The ECG of Patient 1 revealed marked tachycardia of 150 beats/min and WPW syndrome type B, with localization of the accessory pathway in the right anteroseptal area (Fig 1A). Cardiac performance was markedly depressed. A 2-dimensional (D) echocardiogram (Fig 1B) and a T1-weighted magnetic resonance imaging (MRI) scan (Fig 1C) revealed numerous, excessively prominent ventricular trabeculations and deep intertrabecular recesses, but there was no evidence of endocardial thrombus over the left ventricular wall.

The ECG from Patient 2 revealed a normal sinus rhythm and WPW syndrome type B, with localization of the accessory pathway in the right anteroseptal area (Fig 2A). A 2-D

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Second Department of Pediatrics, Toho University School of Medicine, Tokyo, Japan

Mailing address: Koichi Nihei, MD, Second Department of Pediatrics, Toho University School of Medicine, Ohashi Hospital, 2-17-6 Ohashi, Meguro-ku, Tokyo 153-8515, Japan. E-mail: nihei@med.toho-u.ac.jp

Fig 1. Patient 1. (A) ECG showing sinus tachycardia and WPW syndrome type B. The accessory pathway is located in the right anteroseptal area. (B) Two-dimensional parasternal 4-chamber echocardiographic view showing inferoapical left ventricular trabeculations and deep intertrabecular recesses (arrowheads). LA, left atrium; LV, left ventricle. (C) T1-weighted transverse magnetic resonance image showing left ventricular trabeculations and deep intertrabecular recesses (white arrows). LV, left ventricle.

Fig 2. Patient 2. (A) ECG showing WPW syndrome type B. The accessory pathway is located in the right anteroseptal area. (B) Two-dimensional parasternal 4-chamber echocardiographic view showing inferoapical left ventricular trabeculations and deep intertrabecular recesses (white arrows). LV, left ventricle.

echocardiogram (Fig 2B) revealed prominent trabeculations and deep intertrabecular recesses over the entire endocardium of the left ventricle (Fig 2B). Left ventricular systolic function was normal with a LVFS of 44% and LVEF of 81%.

IN patient 3, the ECG revealed a normal sinus rhythm and WPW syndrome type B, with localization of the accessory pathway in the right anteroseptal area (Fig 3A). The 2-D echocardiogram revealed trabeculations and deep intertrabecular recesses over the inferoapical left ventricular area (Fig 3B), but left ventricular systolic function was normal.

Discussion

Two of the 3 patients with INVM and WPW syndrome type B were siblings. All 3 were diagnosed on the basis of the characteristic numerous, excessively prominent trabeculations and deep intertrabecular recesses on echocardiography.^{1,4} Congenital cardiac anomalies that have sometimes been reported in the myocardial pattern of persistent sinusoids, such as ventricular outflow tract obstruction,^{5,6} were not noted.

Patients with INVM can have abnormal ECG findings,

Fig 3. Patient 3. (A) ECG showing WPW syndrome type B. The accessory pathway is located in the right anteroseptal area. (B) Two-dimensional parasternal long-axis (a) and 4-chamber (b) echocardiographic views in showing inferoapical left ventricular trabeculations and deep intertrabecular recesses (white arrows). LA, left atrium; LV, left ventricle.

such as abnormal left frontal QRS axis deviation, varied degrees of atrioventricular block, T wave abnormality, premature ventricular contractions, left bundle branch block, WPW syndrome, atrial fibrillation and other arrhythmias!^{2,7} Ichida et al reported that the incidence of INVM with the abnormal ECG finding of WPW syndrome was higher in children than adults^{2,7} The subtype of WPW syndrome associated with INVM has not previously been reported and, based on the ECG findings, the 3 patients reported here all showed WPW syndrome type B.

Rosenbaum et al were the first to separate WPW syndrome according to the localization of the bypass tract into type A, a left bypass tract in which the QRS complex is dominantly upright in the right precordial leads, and type B, a right bypass tract in which the QRS complex is dominantly downward⁸ WPW syndrome has also been grouped into one of 4 anatomical locations of the accessory pathway: anteroseptal, left free wall, right free wall, or posteroseptal⁹ Type A pre-excitation is invariably associated with a left parietal or posteroseptal accessory pathway, but type B pre-excitation can be seen with all pathways. In all the present patients, the accessory pathway located in the right anteroseptal area⁹ was type B according to Rosenbaum's classification⁸

During early cardiogenesis, there is direct continuity between the atrial and ventricular myocardium and that continuity is lost when the annulus fibrosus forms!¹⁰ Defects in the annulus fibrosus are an explanation for the formation of accessory pathways on the right side of the heart around the tricuspid valve because these pathways traverse the annulus fibrosus and are usually subepicardial in location.¹¹ The final separation of the right atrium from the right ventricle is part of the normal postnatal development!¹² and it has been hypothesized that the basic morphogenetic abnormality of INVM is arrest of the normal process of compaction of the loose interwoven mesh of myocardial fibers in the embryo!¹ and that the normal process of development of the annulus fibrosus after birth is also arrested.

In conclusion, the ECG findings in INVM are related to

WPW syndrome type B because defects of the annulus fibrosus in cardiac embryogenesis explain the subendocardial position of the accessory atrioventricular connections around the tricuspid valve.

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