Familial Trifascicular Block with Autosomal Dominant Inheritance

Dae-Hee Shin, MD, PhD
Division of Cardiology, Gangneung Asan Hospital, University of Ulsan College of Medicine, Gangneung, Korea

ABSTRACT
A 29-year-old man presented to our hospital with dizziness and a history of recurrent syncopal episodes. His mother and 2 uncles had undergone permanent pacemaker implantation several years ago. At presentation, his heart rate was 49 bpm. Electrocardiography (ECG) indicated atrial flutter with right bundle branch block (RBBB) and left anterior fascicular block (LAFB). Twenty-four hour Holter monitoring showed ventricular pause up to 16 seconds during syncope. Radiofrequency catheter ablation was performed for atrial flutter. An additional ECG indicated a trifascicular block (RBBB, LAFB, and first-degree AV block). He then underwent permanent pacemaker implantation. ECG results of the patient’s brother and sister also indicated a trifascicular block, and the family pedigree showed autosomal dominant inheritance.

Key words: atrial flutter • familial heart block • trifascicular block

Introduction
The term “trifascicular block” is confusing because the involvement of the right bundle branch and both fascicles of the left bundle branch would generally manifest as a complete heart block. Moreover, the term trifascicular block is often inaccurately applied for cases with a bifascicular block and prolonged PR interval. Progressive familial heart block (PFHB) type I is an autosomal dominant cardiac conduction disorder that may progress to a complete atrioventricular (AV) block.

Type I PFHB is characterized by a right bundle branch block (RBBB), left anterior fascicular block (LAFB), prolonged PR interval, or complete AV block with broad QRS complexes.

Case
A 29-year-old man presented to our hospital with a complaint of dizziness. He had experienced 2 episodes of syncope 7 and 25 years ago. Examination of his family history indicated that his mother had undergone permanent pacemaker implantation at the age of 51 years (Figure 1). In addition, 2 of his uncles had also undergone permanent pacemaker implantation. On examination, the patient’s blood pressure was 120/80 mmHg, heart rate was 49 bpm, respiratory rate was 20 times per minute, and body
temperature was 36.8°C. An initial electrocardiography (ECG) indicated atrial flutter with variable ventricular response and a bifascicular block (RBBB and LAFB, Figure 2). On echocardiography, a mild left ventricular systolic dysfunction (ejection fraction = 50%) with enlargement of both atria was observed. At admission, he exhibited a seizure-like motion with syncope, and showed a ventricular pause up to 16 seconds on 24-hour Holter monitoring (Figure 3, 4). Subsequently, a temporary pacemaker was inserted. Radiofrequency catheter ablation (RFCA) with bidirectional cavotricuspid isthmus block was then performed for atrial flutter. Following RFCA, an additional ECG revealed a bifascicular block (RBBB and LAFB) and marked first-degree AV block (Figure 5), and 24-hour Holter monitoring indicated an intermittent second-degree AV block. Following frequent episodes of symptomatic high-degree AV block, he underwent permanent pacemaker implantation (DDD type). The ECG results of the patient's brother and sister also indicated a trifascicular block (Figure 6, 7). However, the ECG results of his sister's daughters were normal. A few months later, his brother underwent permanent pacemaker implantation for complete AV block at another hospital. The patient's family pedigree showed an autosomal dominant inheritance (Figure 8).

**Figure 1.** The patient’s mother underwent permanent pacemaker implantation at the age of 51.
Discussion

In 1968, Rosenbaum and his colleagues described the trifascicular nature of the intraventricular conduction system and the trifascicular block and hemiblock. The term “trifascicular block” is confusing, as the involvement of the 3 fascicles in the ventricle would generally manifest as a complete heart block. Therefore, the trifascicular block is often inaccurately applied to cases with alternating RBBB and LBBB or prolonged PR interval and bifascicular block.

Figure 2. Initial ECG showing atrial flutter with variable ventricular response and bifascicular block (RBBB and LAFB).

Figure 3. ECG monitor during syncope showing atrial flutter waves with two ventricular escape rhythms.
In 1977, Brink and Torrington described a new autosomal dominant familial heart disease (progressive familial heart block), which primarily affects the conduction tissue of the heart. The ECG features of type I PFHB are defined by the evidence of RBBB, LAFB, prolonged PR interval, or complete heart block with broad QRS complexes. These ECG features can help differentiate type I PFHB from progressive familial heart block type II (type II PFHB), wherein the onset of complete heart block is associated with narrow QRS complexes. Type I PFHB manifests symptomatically when complete heart block develops, and dyspnea, syncopal episodes, or sudden death are noted.

Figure 4. Twenty-four hour Holter monitor showing a ventricular pause up to 16 seconds during syncope.
Prompt implantation of a permanent pacemaker is vital for the successful management of patients with type I PFHB. The use of a prophylactic pacemaker in these conditions is controversial.\(^4\)

**Figure 5.** The ECG shows a bifascicular block (RBBB and LAFB) and marked first-degree AV block following RFCA (CTI block for atrial flutter).

**Figure 6.** The ECG result of the patient’s sister, 34-year-old shows a bifascicular block (RBBB and LAFB) and marked first-degree AV block. However, her daughters’ ECGs are still normal.
Follow-up visits and ECGs at 6-month intervals, at least, are recommended for patients with any degree of heart block, and an annual examination is recommended for the patient’s family members.
with normal ECGs. Although the global incidence of type I PFHB is not known, this disease may not only be confined to South Africa. A few reports have indicated a familial tendency of bradycardia in the Asian population as well.

References