Twenty-five-year-old woman with palpitations and hypertrophic cardiomyopathy

CLINICAL INTRODUCTION
A 25-year-old woman with a diagnosis of hypertrophic cardiomyopathy (HCM) and pre-excitation on ECG presented with unexplained syncope and daily palpitation. Genetic testing was positive for lysosome-associated membrane protein 2 (LAMP2) mutation which confirmed the diagnosis of Danon disease. Her younger sister was diagnosed with a similar condition and received a defibrillator implantation. Her 12-lead ECG (figure 1) and a long strip tracing (figure 2) are shown below.

QUESTION
Where is the location of the accessory pathway and what is the next appropriate management?

A. Anteroseptal pathway and catheter ablation
B. Mid-septal pathway and pacemaker/defibrillator implantation
C. Right lateral pathway and catheter ablation
D. Fasciculoventricular pathway and electrophysiological study
E. Left lateral pathway and electrophysiological study

Figure 1 12-lead ECG.
ANSWER: D

The correct answer is fasciculoventricular (FV) pathway and electrophysiological study should be the next step of management. Figure 1 demonstrates a 12-lead ECG showing sinus rhythm with a slightly short PR interval of 112 ms and evidence of pre-excitation, especially in precordial leads. The right lateral pathway would have a negative delta wave in lead V1 and left lateral pathway would have it in lead I and aVL. Figure 2 demonstrates a junctional rhythm on the first three beats and sinus rhythm on the last three beats of tracing. There is pre-excitation present as seen by slurring in upstroke of leads V2, V3, V4, and limb leads. Of note, the degree of pre-excitation is same in the sinus and junctional beats. In a typical atrioventricular accessory pathway, junctional beats will not show any pre-excitation since the depolarisation starts below the atrium and does not engage the accessory pathway. Hence, the finding of the similar degree of pre-excitation for junctional and sinus beat is diagnostic for FV pathway. Her electrophysiological study confirmed this diagnosis with a fixed HV interval. In addition, she had easily inducible atrioventricular nodal re-entry tachycardia which most likely caused her palpitation. Successful ablation of the slow pathway was performed. A single chamber defibrillator was implanted for prevention of sudden cardiac death from HCM. FV pathway had never been demonstrated to be the key component of re-entrant tachycardia which most likely caused her palpitation. Successful ablation of the slow pathway was performed. A single chamber defibrillator was implanted for prevention of sudden cardiac death from HCM. FV pathway had never been demonstrated to be the key component of re-entrant tachycardia which most likely caused her palpitation. Successful ablation of the slow pathway was performed. A single chamber defibrillator was implanted for prevention of sudden cardiac death from HCM.