Journal of the American College of Cardiology
© 2013 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Vol. 62, No. 9, 2013
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2013.02.099

IMAGES IN CARDIOLOGY

Cardiac Hypertrophy, Accessory Pathway, and Conduction System Disease in an Adolescent

The *PRKAG2* Cardiac Syndrome

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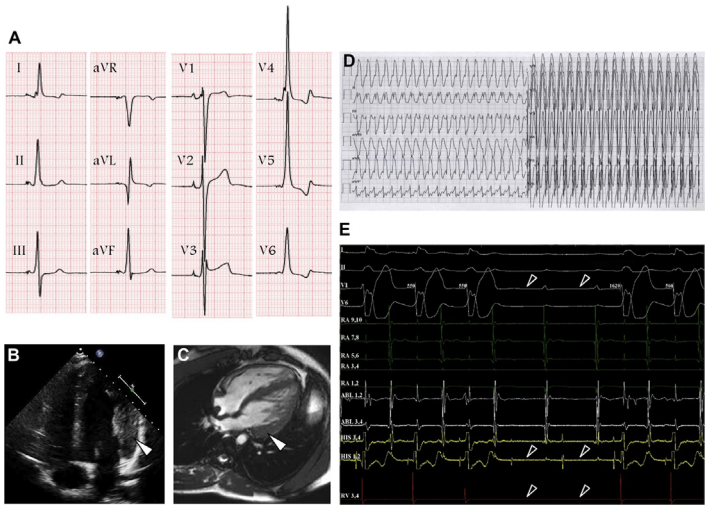

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Manuscript received January 16, 2013; revised manuscript received February 7, 2013, accepted February 12, 2013.

A 17-year-old asymptomatic boy was referred to our hospital for family screening because of his father's unexplained left ventricular hypertrophy (LVH). The father received a pacemaker at 35 years of age for sick sinus syndrome. The electrocardiogram showed LVH and a short PR interval and seemed to show a delta-wave **(A)**. Echocardiography showed mild asymmetrical LVH with posterolateral distribution (**B**, Online Video 1). Cardiac magnetic resonance imaging (**C**, Online Video 2) confirmed asymmetric LVH (maximal wall thickness of 13 mm). A few weeks later, he presented to the emergency department with sudden onset of palpitations. The electrocardiogram showed a supraventricular tachycardia with aberrant conduction **(D)**. An electrophysiological study showed a posteroseptal accessory pathway, which was successfully ablated. Before the procedure, an intermittent third-degree atrioventricular block was observed **(E)**, and a pacemaker was subsequently implanted. Genetic testing identified a missense mutation in the protein kinase, AMP-activated, noncatalytic, gamma-2 (*PRKAG2*) gene leading to an Arg302Glu substitution. As in this case, the *PRKAG2* autosomal dominant cardiac syndrome may be commonly characterized by LVH, an accessory pathway, and progression to conduction disease requiring implantation of a pacemaker (1).

REFERENCE

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