

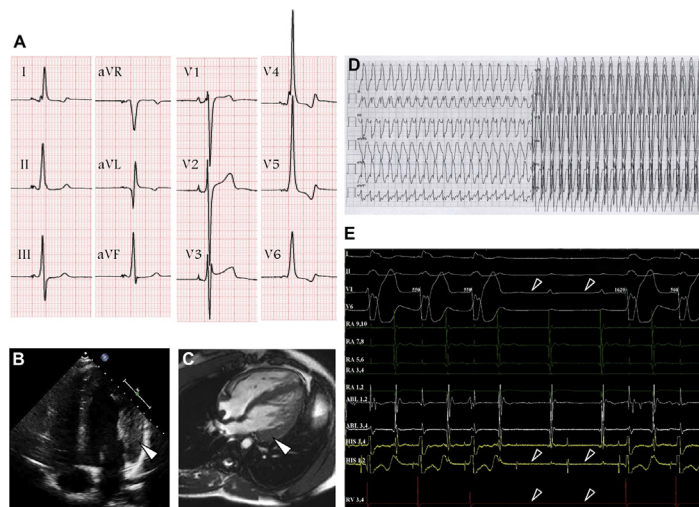
IMAGES IN CARDIOLOGY

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

### The *PRKAG2* Cardiac Syndrome

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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

1. Murphy RT, Mogensen J, McGarry K, et al. Adenosine monophosphate-activated protein kinase disease mimicks hypertrophic cardiomyopathy and Wolff-Parkinson-White syndrome. *J Am Coll Cardiol* 2005;45:922-30.