A 17-Year-Old Boy with Cardiac Arrest Occurring during Exercise

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The patient is a 17-year-old boy who was ostensibly well until admission to the cardiac intensive care unit (ICU). While he was in his physical education class and running “the mile” he collapsed and was noted to have what appeared to be agonal breaths. He subsequently began to have what appeared to be a seizure. It was recognized that he had a cardiac arrest, so cardiopulmonary resuscitation (CPR) was begun by one of his classmates and 911 was called immediately. An automated external defibrillator (AED) was located in the principal’s office and a student ran to retrieve that device while CPR was continually performed. After approximately 5 minutes of CPR, the AED was placed and it advised that a “shock” be delivered. After a single shock, the patient began to breathe spontaneously and regularly and became conscious. By this time (8 minutes after the collapse), the paramedics arrived on the scene. They noted that the boy was breathing spontaneously and was awake but very confused. They prepared him for transport to the hospital. The eventual interrogation of the AED documented ventricular fibrillation and documented a return to spontaneous circulation after the single shock delivered by the AED.

The patient was taken to the cardiac ICU where he was awake but confused, and he was noted to be hemodynamically stable. A careful review of his past medical history showed that he had been having intermittent chest pain over the past several months. The chest pain occurred twice per week, lasted about 10 minutes each time, and seemed to resolve spontaneously. It was mid-precordial in location and occurred at rest as well as with exercise. In addition, he had an episode of syncope last summer. It occurred on a particularly hot day, while he was in church, and he admits that he was not drinking enough water. He was evaluated by his family physician who thought that this episode was most likely from neurocardiogenic syncope and the patient was encouraged to stay better hydrated. Finally, about 2 months prior to this episode of collapse at school, the patient complained about feeling palpitations, then a racing heart. This episode resulted in dizziness but spontaneously resolved after about 90 seconds.

The patient’s family history is significant in that his father was evaluated for chest pain about 1 year prior to the patient’s presentation. The father presented to the emergency department and was then admitted to the hospital, where he received an echocardiogram and a cardiac catheterization study, at which time his coronary arteries appeared normal. A left ventricular angiogram was performed and he was started on a beta-blocker.

The patient’s examination included an electrocardiogram (ECG) (Figure 1), an echocardiogram, and laboratory studies.

Figure 1. An electrocardiogram from the patient’s examination.

For diagnosis, see page 532

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.
Diagnosis:
Wolf-Parkinson-White Syndrome

Although his initial ECG showed only nonspecific ST-T wave changes, several subsequent ECGs clearly showed the presence of Wolf-Parkinson-White syndrome (WPW). In addition, an echocardiogram showed a severely thickened left ventricle (LV). The thickening of his LV was asymmetric, with the ventricular septum being thicker than the free wall of the LV. There was no measurable gradient in the left ventricular outflow tract. A cardiac magnetic resonance study was performed and it was noted that he had moderate patchy delayed enhancement throughout the LV.

The patient was diagnosed with both WPW as well as hypertrophic cardiomyopathy (HCM). After review of his father’s studies from 1 year prior, it was presumed that his father also had HCM. The patient has two siblings, and an echocardiogram and ECG was performed on both. These studies were normal. Genetic testing was sent off for the patient. On day 4 of his hospitalization he was taken to the catheterization laboratory, where a successful ablation of his accessory pathway (WPW) was performed and where an implantable cardioverter-defibrillator (ICD) was placed. He was discharged from the hospital 2 days after this procedure.

DISCUSSION
Both WPW and HCM are potential etiologies of sudden cardiac death (SCD). WPW can be associated with SCD in the patient with WPW who develops atrial fibrillation, and it also has the ability to conduct very rapidly across the accessory pathway. WPW can be treated successfully with catheter ablation of the accessory pathway. HCM is also a known cause of SCD, and in many studies it is the most commonly associated cause of SCD in young athletes. The SCD associated with HCM can be minimized by the restriction of competitive sports and the employment of an ICD in select at-risk people. Risk stratification in patients with HCM that places patients with HCM into the higher-risk category includes a history and/or family history of SCD, an abnormal stress test response that includes hypotension or an attenuated blood pressure response, massive LV thickening on echocardiography (>30 mm thickness in adults), nonsustained ventricular tachycardia on 24-hour Holter monitoring, and an episode of unexplained syncope. There may be a role for cardiac magnetic resonance imaging (MRI) in the risk stratification of patients with HCM. It has been suggested that patients with HCM who have delayed enhancement on cardiac MRI should be in a higher-risk group.

Finally, the combination of WPW and HCM is an unusual one, but it has been described in Danon disease. This diagnosis has not been confirmed in our patient. Danon disease can also be associated with skeletal muscle weakness. The HCM in Danon disease often progresses to heart failure as a result of deterioration of HCM to a dilated cardiomyopathy. The cause of Danon disease is known but is not well understood. It is thought to be a result of damage to the LAMP2 gene; however, how this particular mutation results in the clinical finding of this disease is still unclear.

REFERENCES