Emergency Case

Harold Schubert, MD

Palpitations

Deadly hens teeth

QUESTIONS

What abnormalities does this ECG show? What syndrome might this represent, and what are its implications? What is the treatment for this syndrome?

A 23-year-old fit, athletic male student was sitting in class when he suddenly felt his heart racing. He perspired profusely and thought that he might faint. He left the class and ran to the nearby emergency department (ED). He felt better while running and, in the ED, felt back to his usual self.

On admission to the ED, his vital signs and physical examination were normal. **Figure 1** shows his electrocardiogram (ECG). Due to abnormalities on the ECG, hematology, basic chemistry, cardiac enzymes, and chest x-ray were done. Results were normal. Repeat cardiac enzyme tests after 4 hours continued to have normal results.

Palpitations are a common presenting complaint in EDs. For all types of paroxysmal supraventricular tachycardia (PSVT) alone, a recent United States study[1] has shown a prevalence of 2.25 per 1000 persons and an incidence of 35 per 100 000 person-years. Many more patients have premature atrial contractions or premature ventricular contractions with symptoms. For most patients, these arrhythmias are first diagnosed in an ED. Some of these patients have documented heart disease; many do not.

We are often reminded of the potential for fatal arrhythmias in people with ischemic heart disease. Healthy, young people, free of ischemic or structural heart disease, are also at risk of sudden death from ventricular arrhythmias. We would very much like to be able to identify these people.

More than 300 000 people die of ventricular fibrillation (VF) annually in the United States[2] (by extrapolation, more than 30 000 annually in Canada). Between 3% and 9% of these deaths occur out of hospital and

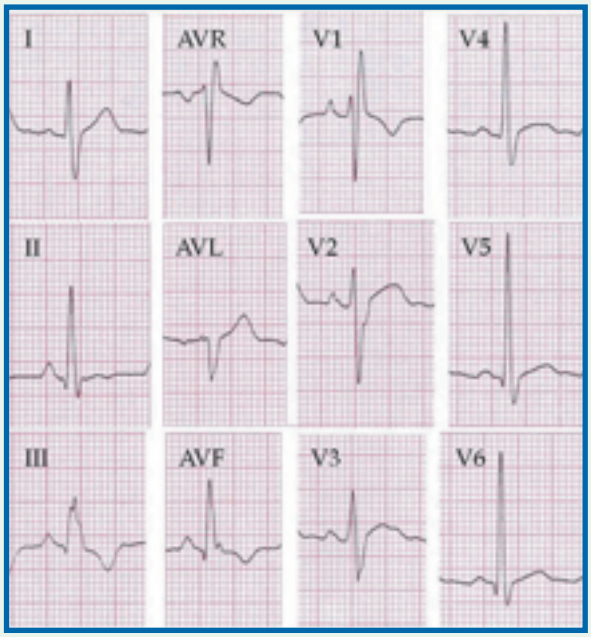

Figure 1. Electrocardiogram of a 23-year-old male student with a history of palpitations

are unrelated to myocardial infarction (>9000 to 27 000 in the United States; >900 to 2700 in Canada).

Three syndromes have thus far been identified to account for sudden death in otherwise healthy people: preexcitation syndrome (Wolff-Parkinson-White syndrome), long-QT syndrome,[3] and Brugada syndrome. The prevalence of VF associated with Brugada syndrome is estimated to be as high as 40% to 60% of all cases of idiopathic VF[2] in some countries.

Brugada syndrome

In 1992, Pedro and Josep Brugada described a new, distinct clinical and electrocardiographic syndrome based on studies of healthy patients with sudden (and aborted) cardiac death.[4] This was subsequently named Brugada syndrome.

Dr Schubert *practises emergency medicine at the University of British Columbia Hospital in Vancouver.*

Brugada syndrome is characterized by a peculiar ST-segment elevation in the right precordial leads, often accompanied by apparent conduction block in the right ventricle; a structurally normal heart; and a propensity for life-threatening ventricular tachyarrhythmias. With a high incidence of familial occurrence, Brugada syndrome appears to follow autosomal dominant inheritance with variable expression.

Brugada syndrome is believed to be a primary electrical disease caused by a defect in a sodium ion channel resulting in premature repolarization of some right ventricular epicardial sites. This results in "dispersion of repolarization," giving rise to closely coupled reentrant extrasystoles that can precipitate ventricular tachycardia (VT) and VF.[2]

In some patients with Brugada syndrome, ECG abnormalities transiently normalize, making diagnosis elusive. Sodium channel-blocking agents, such as procainamide and flecainide, can unmask the ST-segment abnormality. The ST-segment elevation is said to be more prominent before and after non-fatal ventricular arrhythmia episodes. Life-threatening arrhythmias occur most commonly at rest and during sleep. An ECG might actually return to normal with exercise.[2]

A diagnosis cannot be made on the basis of an ECG alone. The ST elevation must be idiopathic, ie, not due to ischemia, electrolyte or metabolic disorders, pulmonary or inflammatory diseases, or abnormalities of the central or autonomic nervous system.[2]

To further confuse the picture, features of Brugada syndrome overlap somewhat with those of other syndromes. Early repolarization syndrome (ERS) can have similar manifestation on ECG, but it is not associated with fatal arrhythmias.[2] A sudden, unexpected death syndrome affecting healthy young men is endemic to Japan and Southeast Asia and might be synonymous with Brugada syndrome.[2] The relatively high incidence of this syndrome among Asians prevails among immigrants to North America. Arrhythmogenic right ventricular dysplasia syndrome also shows similar features to Brugada syndrome on ECG and could represent a temporal progression of Brugada syndrome.[2]

For some people with Brugada syndrome, the first and only adverse cardiac event is sudden death. Those more fortunate come to an emergency department complaining of palpitations, dizziness, or syncope.

For emergency physicians, one point is simple and clear: the abnormality illustrated on the ECG of an otherwise healthy patient complaining of palpitations, dizziness, or syncope should raise suspicion for life-threatening ventricular arrhythmias. Once the suspicion is raised, early cardiology consultation is indicated.

A cardiologist will look for structural cardiac abnormalities using various imaging modalities. If Brugada syndrome is suspected (eg, unexplained syncope, family history of sudden death) but not apparent on ECG, the syndrome can be unmasked using sodium channel blockers. With electrophysiologic testing, VT or VF can be reproducibly induced in Brugada syndrome patients but not in those with ERS. Induction of VT or VF is considered diagnostic and an indication for treatment as Brugada syndrome.[2] Because of the high incidence of familial occurrence, testing family members is important.

Treatment

Treatment for Brugada syndrome is an implantable cardiac defibrillator.[2] The incidence of life-threatening arrhythmias can be reduced, but not eliminated, with β-blockers or amiodarone.[2] Highly selective pharmacologic blocking agents that can fully protect patients with Brugada syndrome might soon be available. Currently, however, the only form of treatment that can, with certainty, protect patients from sudden death is an implantable cardiac defibrillator.

ANSWERS

The ECG shows ST-segment elevation in leads V2 and V3, right bundle branch block (RBBB) and inverted T waves in II, III, AVF, and V1. In the absence of another cause of ST elevation, this pattern is characteristic for Brugada syndrome and is often accompanied by RBBB. Brugada syndrome is associated with sudden death from ventricular arrhythmias. Current treatment for Brugada syndrome is an implantable cardiac defibrillator.

References

1. Orejarena LA, Vidaillet H, DeStefano F, Nordstrom DL, Vierkant RA, Smith PN, et al. Paroxysmal supraventricular tachycardia in the general population. *J Am Coll Cardiol* 1998;31(1):150-7.
2. Gussak I, Antzelevitch C, Bjerregaard P, Towbin JA, Chaitman BR. The Brugada syndrome: clinical, electrophysiologic and genetic aspects. *J Am Coll Cardiol* 1999;33(1):5-15.
3. El-Sherif N, Caref EB, Yin H, Restivo M. The electrophysiological mechanisms of ventricular arrhythmias in the long QT syndrome. *Circ Res* 1996;79:474-92.
4. Brugada P, Brugada J. Right bundle-branch block, persistent ST segment elevation and sudden cardiac death: a distinct clinical and electrocardiographic syndrome. A multicenter report. *J Am Coll Cardiol* 1992;20:1391-6.