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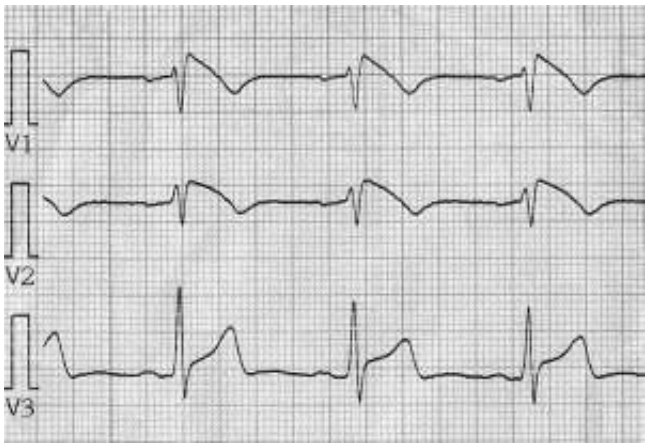
Brugada Syndrome

Patient Case:
24 y/o M recently immigrated from Vietnam for graduate school who presented to ED around 3am after syncopal event while studying for an exam. He had been using cocaine to stay awake while studying and now is completely asymptomatic. His father had died suddenly in his sleep at the age of 28. This is his EKG (or specifically V1-V3):



Only recently described, Brugada Syndrome (BS) is a **hereditary sodium channel disturbance that may result in sudden cardiac death**. It is diagnosed by typical EKG pattern plus one of the following: history of ventricular tachycardia or ventricular fibrillation, family history of sudden cardiac death (less than 45 years old), family history of coved type EKG (will describe later), syncope, nocturnal agonal respirations, and induction of VT during EP study. There are two different EKG patterns associated with Brugada Syndrome: **coved and saddle-backed**. The coved or Type I pattern, more common and more specific and sensitive for BS, is characterized as a convex upwards >2mm ST segment elevation that terminates in inverted t-wave in leads V1-V2. It is often associated with RBBB or incomplete RBBB pattern. The second pattern is called saddle-back or Type II and is characterized as concave upwards >2mm ST segment elevation in leads V1-V2 with RBBB/incomplete RBBB pattern. Interestingly, moving the right precordial leads superiorly to the 2nd or 3rd intercostal space may further unmask the Brugada pattern and increase sensitivity for detecting this EKG finding.

Although the typical patient is a young male from Southeast Asia, the average age for presentation of VF due to BS is **41 years old**, with ages ranging from 2-84 years old. The prevalence is higher in **Asia and Europe** for coved BS and the US prevalence is approximately 0.03%. **Males** are up to nine times more likely to have the Brugada EKG pattern than women. Patients with BS, in particular coved EKG pattern, have approximately 10% mortality per year if ICD is not placed. BS is associated with multiple arrhythmias including **VF, VT, Afib (11-14% of patients with BS), and rarely SVT**.

Several things have been shown to exacerbate BS including **fever, vagal tone, cocaine/alcohol use, drugs, ischemia, hypokalemia, and hypothermia. Medications include sodium channel blockers (class Ia and Ic), calcium channel blockers,**

nitrates, alpha agonists, and beta blockers. Interestingly, arrhythmias are more likely to happen from **midnight to 6am** which explains why in Thailand Brugada Syndrome is known as Lai Tai or "death during sleep."

Classically, medical literature has stated that Brugada Syndrome is associated with a structurally normal heart. Recent publications showed that this is likely not true. Brugada syndrome is often associated with **right ventricular structural abnormality**. In fact, in patients' death with BS, there has not been a single case submitted for a necropsy study without some form of structural heart abnormality.

When a patient with Brugada Syndrome presents to the ED, risk stratification has been ill-defined, and it is necessary to get cardiologists and/or electrophysiologists on board for assessment. Patients with Brugada Syndrome, especially with associated syncope, VF, or VT, will likely need **early ICD placement**. Even though Class Ia sodium channel blockers have been shown to unmask BS, the Class Ia sodium channel blocker **quinidine** is effective for recurrent ICD shocks or people who cannot have an ICD placed.

In summary, when any patient presents to the ED with syncope, it is absolutely necessary to rule out Brugada Syndrome because of the mortality associated with this condition if not treated by specialists. The typical EKG should be ingrained in every emergency physician's brain, in particular the more ominous Type I or coved pattern. Although there is no proven risk stratification model for these patients, it is vital to get cardiologists on board, even if asymptomatic, for possible ICD placement.

References:

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