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Visual Case Discussion

Distinguishing between Brugada and incomplete right bundle branch block on ECG

Greg Zahn, Julie L. Welch*, Steve Roumpf

Department of Emergency Medicine, Indiana University School of Medicine, Indianapolis, IN, USA

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26 year-old white male presented for syncope and fall from a ladder. He had been working in the hot sun without fluid intake. He had a history of bipolar disorder, was not on medications, and admitted to alcohol and marijuana use. He had no family history of sudden death. Physical exam showed normal vital signs, normal cardiac exam, and a 5 cm right posterior scalp laceration, which was stapled closed. ECG revealed an incomplete right bundle branch block (IRBBB) with Brugada pattern (Image 1). Trauma imaging and labs were negative for acute findings. He was admitted for syncope and closed head injury. A Cardiology consult concurred with the IRBBB with Brugada type 2 pattern. He underwent an antiarrhythmic drug challenge with procainamide that showed no arrhythmias and no evidence of right precordial J-point or ST-segment elevation, therefore negative for Brugada pattern/phenotype. It was concluded that he had IRBBB variant.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.visj.2019.100608.

Questions

1. What is thought to cause Brugada syndrome?
   a. Structural abnormality leading to accessory pathway
   b. Increased vagal tone leading to irritability of ventricular tissue
   c. Transient occlusion of proximal LAD or L main coronary artery
   d. Abnormality of sodium and potassium channels
   e. Progressive replacement of myocardium by fibro-fatty tissue
2. After Brugada pattern is identified on an ECG, what should be the next step?
   a. Immediate consult to cardiology
   b. Place defibrillator pads on the patient
   c. Give aspirin
   d. Obtain serial EKGs
   e. Obtain detailed history
3. Cardiologists have studied ECG criteria to discriminate between incomplete right bundle branch block and Brugada types 2 and 3 patterns.
   a. True or False
4. Brugada syndrome is an autosomal recessive inherited syndrome with variable penetrance.
   a. True or False

Answers

1. Abnormality of sodium and potassium channels. Explanation: “Brugada syndrome is an inherited heart disease without structural abnormalities that is thought to arise as a result of accelerated inactivation of Na channels and predominance of transient outward K current to generate a voltage gradient in the right ventricular layers.” It is thought this abnormality is the trigger for degradation into ventricular tachycardia/fibrillation. Reference: Bayes de Luna, A. et al. Current electrocardiographic criteria for diagnosis of Brugada pattern: a consensus report. *J Electrocardiol.* 2012;45:433–442.

* Corresponding author.
E-mail address: jlwelch@iupui.edu (J.L. Welch).

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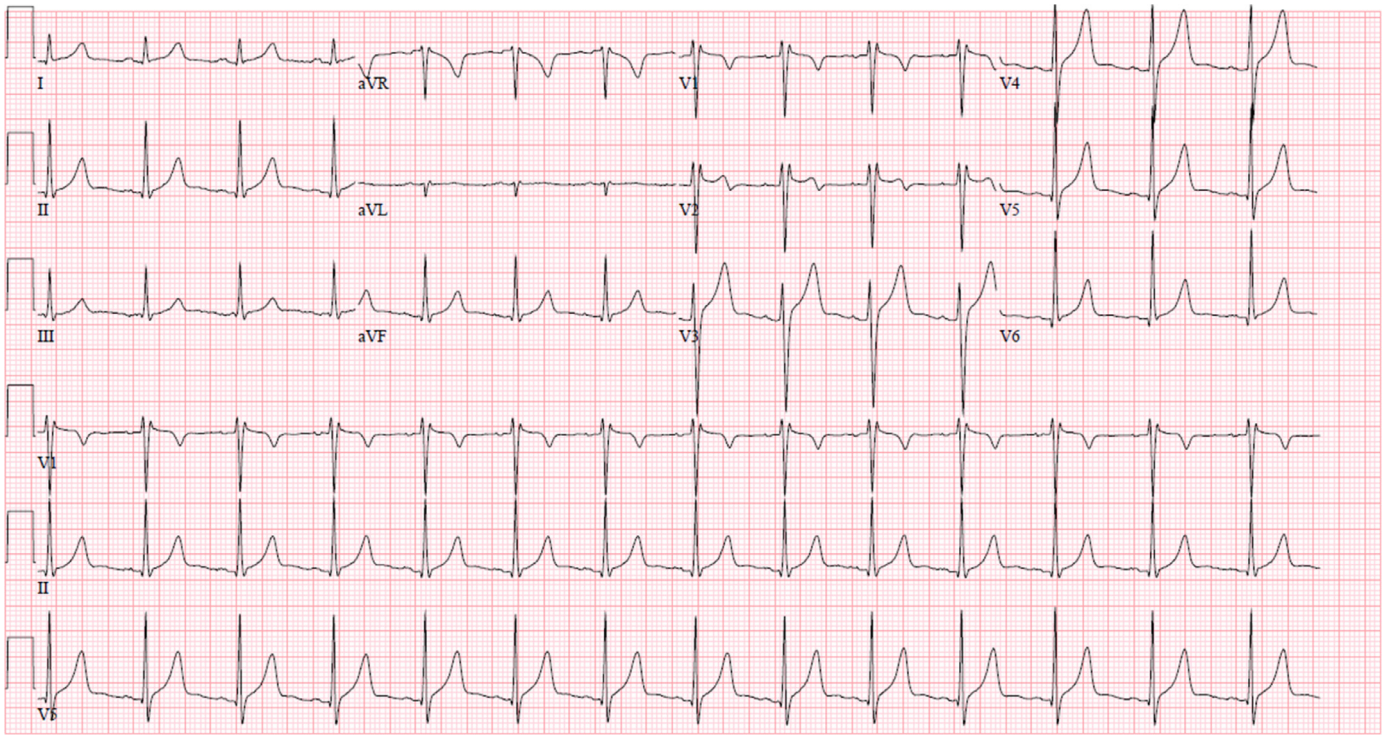

Image 1. ECG demonstrating incomplete right bundle branch block (QRS 96 ms with rsr') and Brugada pattern type 2 in V1, V2 defined as J-wave amplitude ≥2 mm, T wave biphasic or positive, ST-T saddle-back configuration, and ST segment terminal portion elevated ≥1 mm.

2. Obtain detailed history. Explanation: Brugada pattern on EKG does not diagnose Brugada syndrome. The pattern can exist with many other disease processes and is not necessarily specific to Brugada syndrome. History is essential when taking care of a patient with a concerning EKG. Pattern is only descriptive of the EKG, yet the history can lead one to suspect the syndrome. Type 1 (coved) is more diagnostic of the syndrome when it accompanies concerning history such as: history of VT/VF, family history of sudden cardiac death, coved type EKG in family members, agonal respirations during sleep, history of syncope or inducible arrhythmia with provocative testing. Ultimately, an EKG with the pattern accompanied by a concerning history requires specialist involvement. Reference: Priori SG, et al. HRS/EHRA/APHRS Expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. *Heart Rhythm*. 2013;10(12):1932–1963.

3. True. Explanation: Cardiologists originally described 3 types of Brugada syndrome. Type 1 is the "coved-pattern" and types 2 and 3 are defined as the "saddle-back pattern," which is defined as an incomplete right bundle branch block (IRBBB) with ST-segment elevation (type 2 with ≥0.1 mV and type 3 <0.1 mV.) A consensus report in 2012 simplified Brugada classification and combined types 2 and 3 together. Incomplete right bundle branch block (IRBBB) occurs in 3% of the population and can be confused with Brugada types 2 or 3. To diagnose Brugada types 2 or 3, patients undergo antiarrhythmic drug challenge (AAD) to unmasked the Na/K channel dysfunction and convert to type 1. If the patient does not convert to type 1, then it is presumed an IRBBB and not Brudaga pattern. Chevallier et al. studied ECG criteria for distinguishing between Brugada type 2 or 3 and IRBBB. They compared the measured the angle between the S-wave upslope and r' downslope in ECG leads V1 and/or V2. They found that patients with Brugada (i.e., converted with AAD) had baseline angle cutoff of >58° (36 ± 20° in IRBBB vs. 62 ± 20° in Brugada types 2 or 3, $p < 0.01$). This yielded a positive predictive value of 73% and negative predictive value of 87% for Brugada conversion on AAD. References: Chevallier S, Forclaz A, Tenkorang J, et al. New electrocardiographic criteria for discriminating between Brugada types 2 and 3 patterns and incomplete right bundle branch block. *J Am Coll Cardiol*. 2011;58(22):2290–2298; DOI: 10.1016/j.jacc.2011.08.039; Bayes de Luna A, et al. Current electrocardiographic criteria for diagnosis of Brugada pattern: a consensus report. *J Electrocardiol*. 2012;45:433–442.

4. False. Explanation: Brugada syndrome is a familial, genetically determined syndrome characterized by autosomal dominant inheritance in approximately 50% of cases with variable penetrance. Approximately 20% of cases are due to SCN5A mutations, but more than 70 Na channel mutations have been described. Reference: Bayes de Luna A, et al. Current electrocardiographic criteria for diagnosis of Brugada pattern: a consensus report. *J Electrocardiol*. 2012;45:433–442.