Fever Unmasked Brugada Syndrome in Pediatric Patient

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Introduction

Brugada syndrome is an important cause of sudden cardiac death and is prevalent in a young patient population, yet the classic EKG findings may not always be present. Of particular importance to the emergency physician, fever accentuates impaired sodium channel influx to not only unmask classic Brugada EKG changes, but may also induce potentially fatal ventricular arrythmias.[1]

Case Presentation

A 5-year-old African-American female presented to the emergency department with two days of subjective fevers at home. This was associated with congestion, a productive cough, sore throat, and injected conjunctiva. She also complained of nausea and her mother reported decreased oral intake. She was born full-term and has no past medical history. Her immunizations were up to date with the expectation of an annual influenza vaccine. In the emergency department, she was ill appearing, but well hydrated and non-toxic. Her vitals were as follows: Temp 101°F, HR 118, BP 105/70, RR 20, SpO2 98% on room air. HEENT exam was only notable for congestion and an erythematous pharynx. Her lungs were clear to auscultation bilaterally. Heart was tachycardic and irregular with no murmurs, rubs or gallops. Abdomen was soft, non-tender. Skin exam was unremarkable.

The patient's presentation was suspicious for a viral illness; however, her irregular heart rhythm raised concern for other underlying pathology. Differential diagnosis included myocarditis, pericarditis, endocarditis, rheumatic heart disease, premature ventricular beats, sinus arrythmia, atrial arrythmias, supraventricular tachycardia, cardiomyopathies, inherited channelopathies, hyperthyroidism, anemia, pheochromocytoma, toxin induced arrythmia, excessive caffeine intake, electrolyte derangements, among others.

Her CBC, CMP, CXR, magnesium, and troponin were within normal limits. She tested positive for Influenza A. Her EKG showed a sinus rhythm with frequent PVCs, right axis, and coved ST elevation in V1-V2. Her EKG was consistent with Brugada pattern. She had no history of unexplained syncope or family history of sudden cardiac death.

She was transferred to a children's hospital and evaluated by pediatric cardiology. An echocardiogram was negative for structural heart disease. An electrophysiology study was negative for inducible arrythmias and thus the decision was made to defer AICD placement. The patient and her family were referred for genetic testing. She is followed regularly with pediatric cardiology and has had no adverse cardiac events to date.

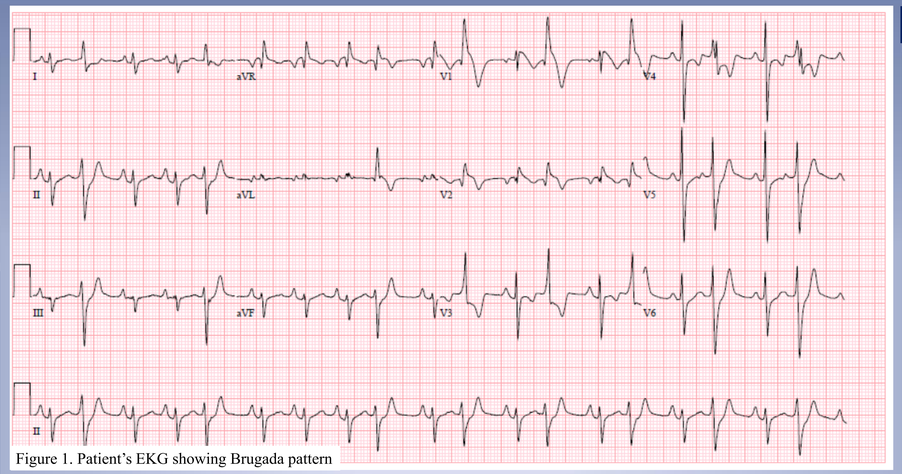


Figure 1. Patient's EKG showing Brugada pattern

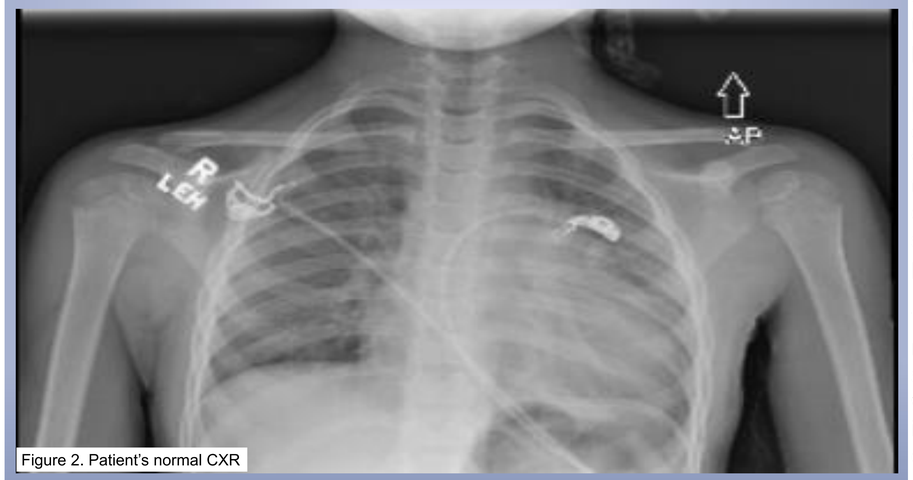
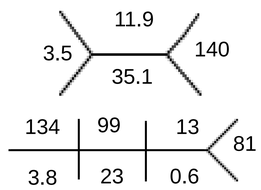


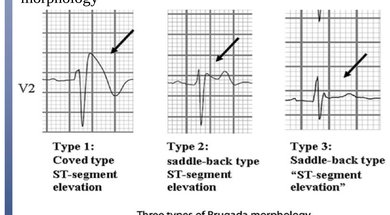
Figure 2. Patient's normal CXR

Figure 3. Patient's lab results



Magnesium: 2.1
POC Troponin I: <0.05

Figure 4. Examples of the three types of Brugada morphology



Three types of Brugada morphology
Image Source: http://circep.ahajournals.org/content/5/3/606/F3.large.jpg

Discussion

Brugada syndrome was first recognized in 1992 and has since been identified as an autosomal dominant inherited sodium channelopathy. Specifically, a loss of function mutation of the SCN5A gene causes an impairment of sodium influx during phase 0 of the cardiac cycle.[2] Classic EKG findings include three types of ST elevations in V1-V3: coved, saddle-back, or a combination.[3] The diagnostic criteria for Brugada syndrome is the presence of a type I pattern (coved ST elevation) on EKG with one of the following: documented ventricular arrythmia, family history of sudden cardiac death, type I pattern EKG in a family member, induced ventricular arrythmia with electrical stimulation, or history of unexplained syncope.[4]

Fever provokes these EKG changes in asymptomatic individuals with Brugada and the EKG may revert to normal when afebrile. While the exact mechanism is unknown, it is postulated that the sodium channels are temperature sensitive and thus fever exacerbates the impairment of sodium influx through the sodium channel.[4] This leads to nonhomogeneous repolarization of the right ventricle and thus a potential for re-entry arrythmias. 18% of cardiac arrests in Brugada patients have been associated with fever.[3]

In children, symptomatic Brugada is most frequently associated with fever. As clinicians, it is important to recognize that fever is the state most likely to induce ventricular arrythmia in children with Brugada.[5] Parents need to be educated to treat fevers aggressively and hospital admission for cardiac monitoring should be considered for patients without an AICD. Furthermore, emergency physicians should not be falsely reassured by the normalization of the EKG in this patient population.[6]

Treatment of asymptomatic patient's with Brugada is controversial in adults and not well studied in pediatrics. Symptomatic patients with a history of arrythmia, syncope, or family history of sudden cardiac death all receive an AICD. Those who are asymptomatic commonly undergo electrophysiology studies and if no inducible arrythmia is found, they are typically managed conservatively with close follow up.[3]

Conclusion

Brugada syndrome is primarily reported in adults with limited data on its presentation in pediatrics. In both adults and pediatrics, a febrile illness may unmask an underlying Brugada pattern EKG, even in individuals with normal previous EKGs. The impaired sodium channels are thought to be temperature sensitive and thus fever exaggerates the impaired sodium influx. This not only emphasizes the EKG changes, but also places the patient at risk for fatal arrythmia. Emergency physicians must be cognizant of the potentially life-threatening complications of fevers in this patient population and have a low threshold for admission.

References

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