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GAVIN PUBLISHERS

Advances in Pediatrics and Neonatal Care

Suzan N, et al. Adv Pediatr Neonatol care: APNC-116.

DOI:10.29011/APNC-116.100016

Case Report

Brugada Syndrome Diagnosed in a Pediatric Patient Presenting with Syncope

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Citation: Suzan N, Boshra N, Bar A, Yehia A, Saed AZ (2020) Brugada Syndrome Diagnosed in a Pediatric Patient Presenting with Syncope. Adv Pediatr Neonatol care: APNC-116. DOI:10.29011/APNC-116.100016

Received Date: 12 September, 2020; **Accepted Date:** 21 September, 2020; **Published Date:** 25 September, 2020

Abstract

Brugada syndrome is an inherited cardiovascular disorder. Approximately 25% of individuals affected with Brugada syndrome demonstrate a mutation in the SCN5a sodium channel. The main symptom is irregular heartbeats and, without treatment, may potentially result in sudden death. Frequently, sudden death can be the first manifestation of the disease. The condition affects between 1 and 30 per 10,000 people. It is more common in males than females and in those of Asian descent. It is characterized by a pattern of ST segment elevation in the precordial leads on an electrocardiogram (ECG) due to a sodium channelopathy.

This case report highlights the case of a 12-year-old female who presented to the emergency department due to syncope, and had an ECG consistent with Brugada syndrome.

Keywords: Brugada syndrome; Arrhythmia; ECG; Syncope; Channelopathy

Introduction

Brugada syndrome is a genetic disorder in which the electrical activity within the heart is abnormal, this disorder can lead to irregular heartbeats in the heart's lower chambers (ventricles), which is an abnormality called ventricular arrhythmia [1,2]. It is the major cause of sudden death in adults and most common reason of unexplained death without known underlying cardiac disease. Brugada syndrome usually becomes apparent in adulthood, although it can develop any time throughout life. The most frequent sign is a persistent ST elevation in the electrocardiographic leads V1-V3 with a right bundle branch block (RBBB) [3]. If untreated, the irregular heartbeats can cause fainting (syncope), seizures, difficulty breathing, or sudden death. These complications typically occur when an affected person is resting or asleep.

Some cases may be due to a genetic mutation or certain medications, About a 1/4 of those with Brugada syndrome have a family member who also has the condition due to that Testing people's family members may be recommended.

The most commonly involved gene is SCN5A which encodes the cardiac sodium channel [4]. Diagnosis is by ECG, however, the abnormalities may not be consistently present.

Medications such as ajmaline may be used to reveal the ECG changes. Similar ECG patterns may be seen in certain electrolyte disturbances or when the blood supply to the heart has been reduced [5]. There is no specific treatment for Brugada syndrome. Those at higher risk of sudden cardiac death may be treated using an implantable cardioverter defibrillator (ICD) [6]. Isoproterenol may be used in the short term for those who have frequent life-threatening abnormal heart rhythms, while quinidine may be used longer term.

Case Report

A healthy 12-year-old girl that was admitted to the Emergency Department due to syncope at midnight, Family history was negative. In the ambulance, ECG showed VT that responded to the treatment by Procor IV. In the Emergency Department she was ill appearing, but she was well-hydrated and non-toxic in appearance.

Physical examination showed stable vital signs. On physical On cardiac exam, she was tachycardic with an irregular rhythm exam. There were no murmurs, rubs, or gallops heard. Her lungs

were clear to auscultation bilaterally. Her abdomen was soft and non-tender.

The laboratory tests showed normal values of magnesium; Troponin and CPK were normal. The ECG showed a Picture of Brugada Syndrome, sinus rhythm with frequent premature ventricular contractions, a right axis, and coved ST elevation in V1–V2 (Image 1). A chest x-ray was normal, Echocardiogram without evidence of myocarditis or cardiomyopathy.

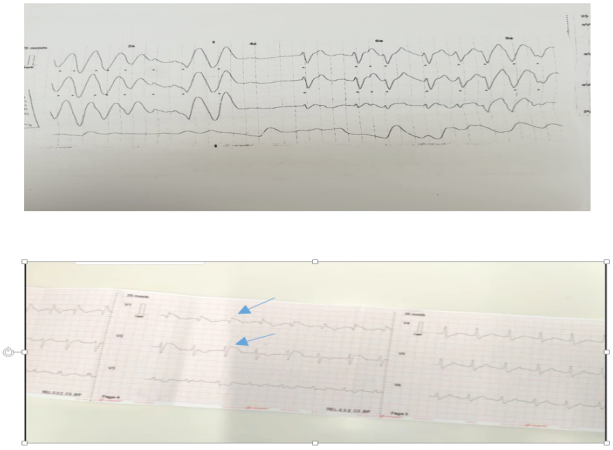

Image 1: ECG showing signs of Brugada Syndrome.

The child was sent to the ICU and treated with Procor and β-blockers. Followed by electrophysiological evaluation for ICD transplantation.

Discussion

Brugada syndrome is 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. Three different types of ECG patterns in Brugada syndrome are known; Type 1 has a coved type ST elevation with at least 2 mm (0.2 mV) J-point elevation and a gradually descending ST segment followed by a negative T-wave. Type 2 has a saddle-back pattern with a least 2 mm J-point elevation and at least 1 mm ST elevation with a positive or biphasic T-wave. Type 2 pattern can occasionally be seen in healthy subjects. Type 3 has either a coved (type 1 like) or a saddle-back (type 2 like) pattern, with less than 2 mm J-point elevation and less than 1 mm ST elevation. Type 3 pattern is not rare in healthy subjects [6-11].

According to current recommendations, only a Type 1 ECG pattern, occurring either spontaneously or in response to medication, can be used to confirm the diagnosis of Brugada syndrome as Type 2 and 3 patterns are not infrequently seen in persons without the disease.

Signs and symptoms, while many of those with Brugada syndrome do not have any symptoms, Brugada syndrome may cause syncope and/or cardiac arrest that occurs during sleep at night or at rest. Treatment may include application of an ICD. β-blockers and Quinidine are effective for patients with unstable ventricular arrhythmias.

Fever provokes ECG changes in asymptomatic individuals with Brugada and the ECG may revert to normal when the patient becomes afebrile [8]. 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 channel.

In children, symptomatic Brugada is most frequently associated with fever. Treatment of asymptomatic patients with Brugada is controversial in adults and not well studied in pediatrics.

Symptomatic patients with a history of arrhythmia, syncope, or family history of sudden cardiac death should all receive an ICD [7].

Those who are asymptomatic commonly undergo electrophysiology studies and if no inducible arrhythmia is found, they are typically managed conservatively with close follow-up [9-10].

Conclusion

Brugada syndrome is a very rare arrhythmia disorder. The physicians must think of it when repeated syncope and/or nightmare were found in healthy children and try to find the typical ECG findings.

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