

The Brugada Syndrome in children: presentation and outcome of proband patients.

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Background.

Brugada syndrome (BrS) is a potentially serious channelopathy that usually presents in adulthood and less frequently in children. We sought to examine the presentation and outcome of Brugada syndrome proband patients in childhood.

Methods and Results.

A review from two institutions, Brussels and Barcelona collected retrospectively from 1996 and prospectively from 2006. The criteria for inclusion were documentation of spontaneous or drug induced ≥ 2 mm coved (type 1) Brugada ECG pattern at age ≤ 16 years and no previous diagnoses of BrS in the family. 15 patients were identified. Presenting age ranged from 2 to 16 years (median age: 13 years). 10 patients were older than 12 years. Three patients (aged 12, 13 and 16 years) presented with aborted sudden cardiac death. In these 3, diagnosis was made retrospectively based on presentation and earlier ECG records. 5 patients presented with an aborted sudden death (age range 8 to 16 years). Syncope was the presentation in 6 other patients, aged from 2 to 16 years. 3 of these were previously diagnosed with Sick Sinus Syndrome or atrial fibrillation. 1 asymptomatic 16 year had coincidental Brugada ECG pattern. At presentation 12 patients had a type 1 ECG pattern. Ajmaline testing was performed in the 3 other patients and was positive. 1 patient required CPR following ajmaline infusion. An ICD was implanted in 7 patients. 4 patients received appropriate ICD shocks. 2 patients received inappropriate shocks. During the follow up of the remaining 12 patients, 1 patient with ICD died at the age of 18 years. 1 patient died 3 years after a first aborted sudden cardiac death because of misdiagnosis. One patient was lost from follow up after one year.

Conclusion .

In this series of proband children with BrS, onset of symptoms occurs from a very young age. The presenting symptom is often aborted sudden death or syncope. Most patients presented with a coved type ECG in rest. The other patients had positive ajmaline testing. Sinus node dysfunction and atrial conduction disorders are commonly associated and Brugada syndrome needs to be considered as an etiology.