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Right bundle branch block or Brugada syndrome?

Case report

A male patient, born 1961, from Sri Lanka was admitted to hospital with two generalised seizures during sleep. The electroencephalogram was not conclusive for epileptic potentials, the MRI of the cranium was normal. Two ECG showed atypical right bundle branch block at different times (fig. 1), which normalised later during hospital stay.

What is your diagnosis?

Do you suggest further tests, and if yes, which ones?

Explanatory answers

(1.) The history of seizures during sleep and the ECG on admission are highly suggestive for the Brugada syndrome, despite the fact that later ECG were within the normal range. Based on this evidence, an implantable cardioverter defibrillator was prophylactically inserted.

The Brugada syndrome has a high incidence in south east Asia and is known there to natives for many decades. In the Philippines for example it is known as "bangungut", which translates as "screams and dies", and in Thailand as "lai tai", which means "death during sleep". The ECG characteristics of the Brugada syndrome are a right bundle branch block pattern and descending ST segment elevation in leads V_1 to V_3 [1]. Clinically, the syndrome manifests by syncope and sudden cardiac death from fast polymorphic ventricular tachycardia and ventricular fibrillation typically in young male patients.

(2.) Family history is one of the cornerstone in diagnosing the Brugada syndrome, which is an autosomic dominant inherited disorder due to mutations in the *SCN5A* gene coding for the cardiac sodium channel (same as long QT syndrome type 3 and other congenital rhythm disorders). The diagnosis is based on the typical ECG findings that may be blunted and have to be unmasked by the administration of a class 1 anti-arrhythmic drug. Given the intermittent nature of the ECG findings, the diagnosis may be difficult in the interval. In this family, the syndrome was confirmed in the index patient and one of three siblings with

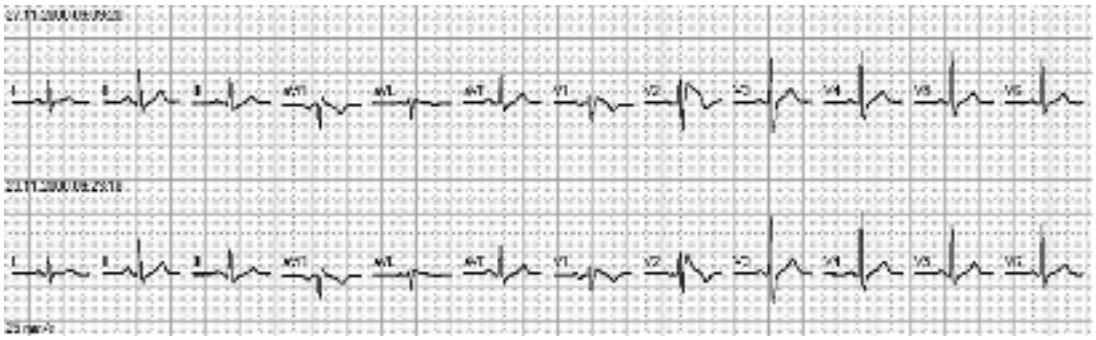

Figure 1
Baseline ECG of index patient I-2 at different times.

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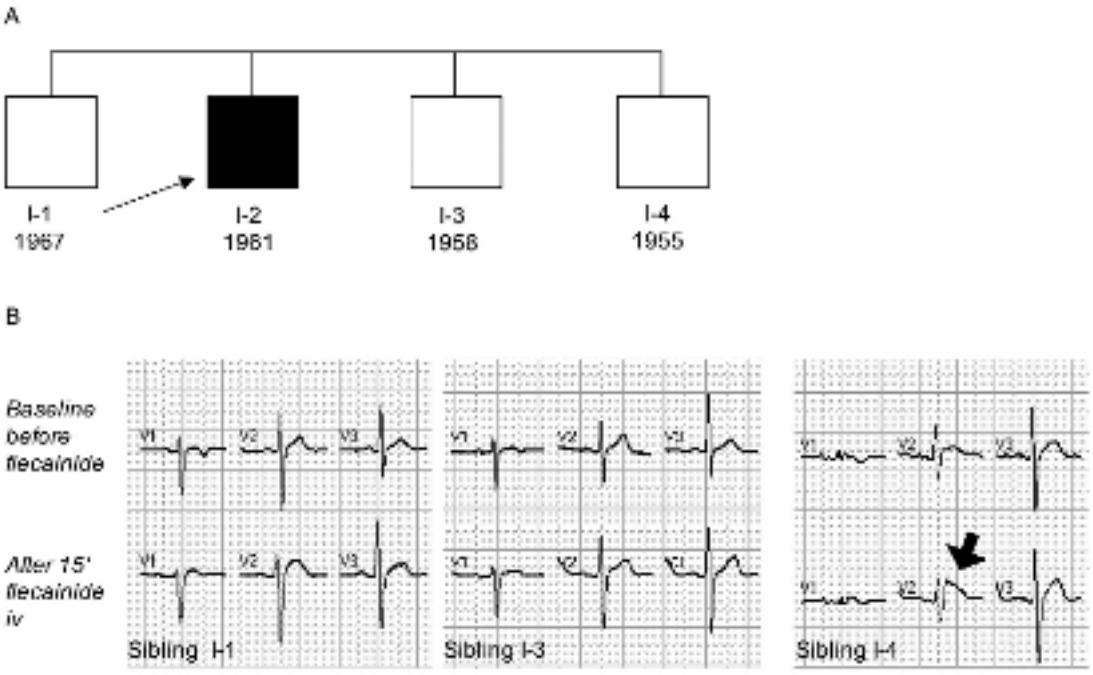

Figure 2
A Index patient I-2 and siblings.
B Flecainide testing in siblings.

normal ECG (sibling I-4) by the intravenous administration of flecainide (2 mg/kg body weight), which unmasked the typical ST-T configuration with a gradually descending ST-segment and a J wave amplitude ≥2 mm in lead V_2 [2] (fig. 2A and B). In addition to class 1 anti-arrhythmic drugs fever has also been shown to unmask or trigger the Brugada syndrome phenotype [3].

References

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