Brugada Syndrome and Sinus Node Dysfunction: Case Study

Beye SM¹, Tabane A², Sarr SA³, Mingou J⁴, Ndiaye MB⁵, Bodian M⁶, Diao M⁷, Kane Ad.⁸
Teaching Hospital Aristide Le Dantec / Department of cardiology

Abstract:
Introduction: Brugada syndrome is due to calcium channels dysfunction that can be responsible for serious ventricular disorders and conduction abnormalities.

Observation: It was a 37-year-old man, who had been seen for bradycardia. The interrogation found a notion of undocumented loss of consciousness at the age of 17 consecutive to an effort. Blood pressure was at 150/70 mmHg and the heart rate was 42 bpm. The physical examination was featureless apart from bradycardia. The electrocardiogram recorded a straight branch block aspect, an ST segment elevation in straight precordial leads (V1 to V3), and sinus pauses. The Doppler echocardiography was normal. The Holter monitor showed sinus pauses, the longest one lasting 6095 ms. No rhythm disorder was noted during the 24 hours. Electrophysiological exploration showed an extension of the AH intervals at 140 ms and HV at 70 ms. The corrected sinus recovery time was calculated at 3772 ms. Luciani Wenckebach's time was measured at 480 ms and the point 2/1 at 450 ms. The programmed ventricular pacing had not triggered ventricular arrhythmia. The patient benefited an implantable cardioverter defibrillator (ICD). The evolution was good at 3 months with control of the ICD parameters. Genetical analysis was not done due to lack of financial resources. A clinical family survey is in progress.

Conclusion: The association Brugada syndrome and sinus node dysfunction is not uncommon. Only the implantation of an ICD is an approved therapy.

Keywords - Brugada, Sinus node dysfunction, Electrophysiological exploration, Implantable defibrillator.

Introduction
Brugada syndrome is an autosomal dominant inheritance disorder with variable presentation, and is responsible for syncope or sudden death secondary to polymorphic ventricular tachycardia. The diagnosis is based on the characteristic aspect of the electrocardiogram which associates a right branch bundle block morphology and an ST segment elevation in the right precordial leads. In patients at risk of sudden death, only the implantable defibrillator provides effective protection against ventricular arrhythmias. We report a case of type I Brugada syndrome associated with sinus node dysfunction and infra-hisian block.

Observation
It was about a 37-year-old patient, a street vendor who had been seen for syncope. The interrogation had found a notion of loss of consciousness at the age of 17 after an effort. The blood pressure was 150/70 mmHg and the heart rate was 42 bpm. The physical examination was featureless apart from auscultatory bradycardia. The electrocardiogram recorded a straight branch bundle block aspect, an ST segment elevation in straight precordial leads (V1 to V3) and sinus pauses (Figure 1).

Figure 1: 12-lead surface ECG showing Brugada appearance and sinus dysfunction
The Doppler echocardiography was normal.
The Holter monitor showed sinus pauses, the longest lasting 6095ms. No rhythm disorder was noted during the 24 hours (Figure 2).

**Figure 2: sinus pauses at the ECG Holter**

Electrophysiological exploration showed an extension of the AH intervals at 141 ms and HV at 72 ms (Figure 3). The corrected sinus recovery time was extended and calculated at 3772 ms. Luciani Wenckebach’s time was measured at 480 ms and the point 2/1 at 450 ms (Figure 4).

**Figure 3: Measurment of intervals during electrophysical study**

The programmed ventricular pacing had not triggered ventricular arrhythmia. The patient had benefited from the placement of a branded dual chamber implantable cardioverter defibrillator (ICD), with a single therapy zone at 220bpm. The evolution was good at 3 months with control of the ICD parameters. No syncope was noted and no arrythmias recorded by the ICD.

A clinical family survey was proposed.

**Discussion**

Brugada syndrome was first described in 1992 by the Brugada brothers, clinically characterized by the occurrence of syncope or sudden death related to polymorphic ventricular tachycardias in patients with structurally normal heart, and electrocardiographically, by an ST segment offset and a straight branch bundle block aspect in precordial straight lines (V1 to V3) [1].

There is a clear predominance of the male sex: 13 women for 150 men in the series of Alings [8] taking up all the world cases published until 1999. The age of discovery is very variable, between 2 and 77 years [6,7]. This very broad range is explained by the great variability of diagnostic modalities, from symptomatic patients to systematic screening of familial forms.

The average age of onset of the first clinical event is 40 years [9]. Clinical manifestations vary from lipothymia, syncope to sudden death through misleading forms (memory loss). They most often occur at rest especially during sleep. Some patients are asymptomatic and the diagnosis is made when performing a systematic electrocardiogram or screening for familial forms [14].

Brugada syndrome is transmitted autosomal dominantly with variable expressivity and incomplete penetrance [14]. The SNC5A gene that codes for the alpha subunit of the sodium channel was identified for the first time in 1998 as responsible for Brugada syndrome [15].

Recent studies have shown that the association of sinus dysfunction and Brugada syndrome is not uncommon and is related to a genetic mutation involving cardiac sodium channels such as mutations of the SCN5A gene.

Mutations in the SCN5A gene lead to loss of function of the cardiac sodium channels [2]. Indeed, the sodium channels play a major role in the initiation and propagation of the action potential of the excitable cells. This sodium channel dysfunction can lead to disturbances of rhythm and conduction at different levels [10].
Esmita and Co [4] suggest that loss of function of the cardiac sodium channels may be responsible for ST segment elevation, rhythm disturbances, and conduction at different levels.

Morimata and Co [5] reported a case of autopsy of a subject with Brugada syndrome with significant lesions of the sinus node and the presence of adipose tissue and important fibrosis.

Sumiyoshi and Co [3] reported, in three patients with Brugada syndrome, a sinus pause of more than 3s. Electrophysiological exploration found an increase in recovery time corrected in two patients. Priori and Co [6] consider that the sensitivity and specificity of programmed ventricular pacing to identify patients at risk of sudden death are poor, 66% and 34% respectively, and do not include it in prognostic evaluation [7]. These results should be interpreted with caution, given the small proportion of patients who have had programmed ventricular pacing and the lack of protocol consistency.

Auricular vulnerability is increased in Brugada syndrome, as documented by electrophysiological studies.

Morita and Co [16] indicated that the effective atrial refractory period is not prolonged in Brugada syndrome, but that the intra-atrial conduction time is significant (168.4 ± 17.5 ms vs 131.8 ± 13.0 ms, p <0.001).

All 11 patients studied by Yamada and Co. [17] had atrial fibrillation induced by a protocol using up to two extrastimuli. Atrial fibrillation was induced only in 8 of the 14 patients (57%) of the Morita and Co serie [16] with an extrastimulus.

Conclusion

The association Brugada syndrome and sinus dysfunction is not uncommon. The low prevalence of Brugada syndrome can not be neglected because of its potential severity, which should lead to great vigilance including in asymptomatic patients. Only the implantation of an ICD is an approved therapy.

Bibliography


