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POSITION STATEMENT

Update on the Diagnosis and Management of Brugada Syndrome

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Brugada Syndrome (BrS) is an autosomal dominant channelopathy with variable penetrance affecting the sodium channel. It reduces the transport of sodium ions essential for proper generation of the cardiac action potential. The resulting inhomogeneous repolarisation in areas of the RV epicardium causes malignant ventricular arrhythmias.

BrS is diagnosed by typical cove shaped ST elevation of > 2 mm in ≥1 RV precordial lead V1, V2 occurring spontaneously or after provocative drug test with IV administration of Class 1 antiarrhythmic drug such as flecainide or ajmaline.

The incidence of BrS is variable being higher in South East Asians and is generally quoted as 1:2000. It is responsible for up to 20% of sudden arrhythmic deaths in those without structural heart disease. Typical presentation is syncope or resuscitated sudden death and symptoms usually occur at night or at rest especially after a large meal. Fever is a common trigger, particularly in children.

Genetic testing for BrS is a Class 2A indication and the yield has increased recently to nearly 40%. Genetic testing assists with family screening.

Keywords Brugada Syndrome • Atrial fibrillation • Genetic testing • Sudden death • Syncope • Heart disease

Management of Brugada Syndrome

Resuscitated cardiac arrest and cardiac syncope are Class 1 indications for implantation of an ICD. All family members of BrS patients should be screened and those with normal or non-diagnostic ECGs should be offered ajmaline or flecainide test. ICD implantation in BrS has a significant complication rate and should be avoided in asymptomatic patients. Family history of sudden death is not an indication for ICD implantation. Asymptomatic patients should be advised of lifestyle measures such as avoidance of 'Brugada drugs', prompt treatment of fever and avoiding excess of alcohol and big carbohydrate meals at night.

Risk Assessment of Asymptomatic Brugada Syndrome Patients

Event rate in those with spontaneous type 1 ECG has been reported as 0.24 to 1.7% per year. Drug induced BrS pattern ECG patients are at minimal risk. Several major trials have reported that programmed electrical stimulation is not helpful in risk stratification. Fragmentation of QRS on the ECG, RV ERP of ≤200msec, history of syncope, atrial fibrillation and spontaneous type 1 Brugada pattern on the ECG, put the patient in the higher risk category.

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Treatment of Arrhythmic Storms

Isoprenaline infusion is effective in acute situations and quinidine is the only effective drug in long-term treatment.

Clinical Characteristics

Definition and Prevalence

Brugada Syndrome (BrS) was described as a clinical entity in 1992 [1]. The diagnosis is made by electrocardiogram (ECG) and is defined by the presence of an atypical right bundle branch block (RBBB) pattern with a characteristic cove-shaped ST elevation in leads V_1 to V_3, in the absence of obvious structural heart disease, electrolyte disturbances or ischaemia. Inheritance can be autosomal dominant with incomplete penetrance, or be polygenic [2,3]. It can also appear as a consequence of structural changes in the right ventricular outflow tract from a variety of causes. BrS is reported to be responsible for 4% of all sudden deaths and 20% of sudden deaths in those without structural heart disease and is a leading cause of death in subjects under the age of 40 years. A family history is present in about 20 to 30% of patients. It is difficult to estimate the exact incidence of BrS in the general population but the prevalence is quoted as 1 in 2000 [4–6]. The condition is particularly common in South East Asia and amongst migrants of South Asian origin and in the Japanese population the prevalence is reported to be 0.5-1 per 1000. BrS has also been reported as Sudden Unexplained Death Syndrome (SUDS) or Sudden Unexplained Nocturnal Death Syndrome (SUNDS) [7]. The ECG changes of BrS are dynamic and can vary spontaneously.

BrS appears to be a channelopathy, and, besides electrophysiological changes, there have also been reports of subtle structural changes in the atria and the right ventricular outflow tract (RVOT) [4].

Clinical Presentation

The most typical presentation is syncope or resuscitated sudden death in the third or fourth decade of life due to polymorphic ventricular tachycardia (VT) or ventricular fibrillation (VF). Symptoms typically occur at night, or at rest during the day but also uncommonly during exercise [2]. Monomorphic VT is rare and is more prevalent in children and infants, among whom fever is the commonest trigger [8,9]. The diagnosis of BrS may also be made on family screening of patients with BrS or incidentally following a routine ECG. More than 80% of adult patients are males but in children there is an equal male:female ratio. Many subjects remain asymptomatic throughout life.

Clinical Diagnosis

Consensus statements regarding making the diagnosis of Brugada Syndrome

The first consensus report of 2002 proposed using ECG criteria alone. These are shown in Figure 1. Three subtypes of ECG features have been recognised [3].

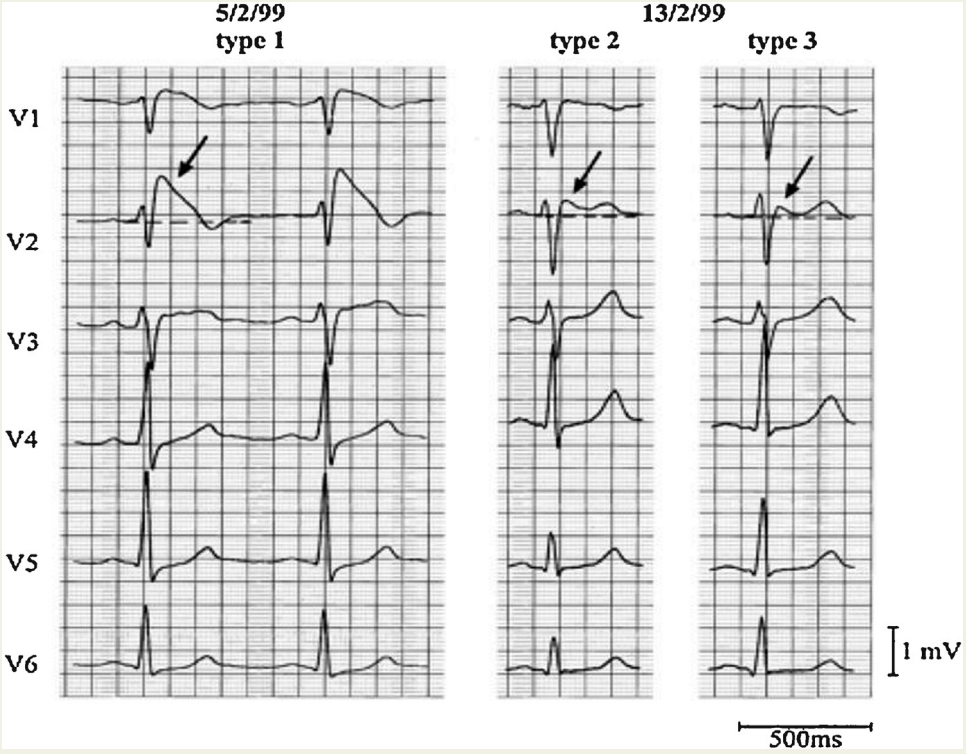

Figure 1 Precordial leads of a resuscitated patient with BrS showing all 3 ECG patterns and dynamic changes over an 8-day period. Arrows indicate J waves.(3) *(Reproduced with permission from: Wilde AA et al. Proposed diagnostic criteria for the Brugada syndrome. Circulation 2002; 106:2514-19).*