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Approach to Brugada Syndrome

Johnson Francis, MBBS, MD, DM

Baby Memorial Hospital, Kozhikode, Kerala, India. PIN: 673004

Address for Correspondence: Prof. Dr. Johnson Francis, MBBS, MD, DM, FCSI, FACC, FRCP Edin, FRCP London, Senior Consultant Interventional Cardiologist, Baby Memorial Hospital, Kozhikode, Kerala, India. PIN: 673004. E-mail: pulikkottil2002@hotmail.com

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Brugada Syndrome was initially described by Brugada brothers [1] in their seminal paper published in the *Journal of American College of Cardiology* in 1992. The syndrome was characterized by syncopal episodes and/or sudden cardiac death in association with right bundle branch block pattern with ST segment elevation in right precordial leads. Genetic basis involving mutations in sodium channel (SCN5A) was first described in 1998 [2]. But SCN5A mutations account for only about a fifth of the clinical cases of Brugada syndrome [3]. Mutations in several other genes have been documented in later studies (at least 16 of them), though SCN5A mutation is the commonest [4]. Life threatening ventricular arrhythmias with a structurally normal heart was the usual pattern in initial reports. But later on it was noted that several persons with similar ECG pattern never had any arrhythmias in their life time. There was significant variation in the severity of presentation between different regions of the world, with more malignant phenotypes noted in South East Asian countries.

Different Brugada ECG patterns have been described - Types 1-3 (**Figures 1, 2**). But it may be noted that only type 1 is diagnostic of Brugada syndrome. Types 2 and 3 have to be converted to type 1 either spontaneously or with drug challenge with a sodium channel blocker, typically flecainide, before a diagnosis of Brugada syndrome can be considered. In addition, there should be evidence of life threatening ventricular arrhythmias, positive family history or syncope of presumably cardiac origin to qualify for a diagnosis of Brugada syndrome. If these are lacking, it may be called as a Brugada ECG pattern and not Brugada syndrome. Brugada ECG pattern can fluctuate over time as is well illustrated in the electrocardiograms shown below.

Brugada ECG pattern is fairly common, especially types 2 and 3, though actual Brugada syndrome is rare. This often leads to a therapeutic dilemma due to the potential risk of life threatening arrhythmias in Brugada syndrome. Role of electrophysiological studies have been conflicting with some groups finding it very useful while the large majority of investigators have found it not very useful in prognostication. Other causes of ST segment elevation in anterior leads including ST segment elevation myocardial infarction should be ruled out before considering a diagnosis of Brugada syndrome.

Even though ventricular fibrillation and polymorphic ventricular tachycardia are the hall mark arrhythmias in Brugada syndrome, atrial arrhythmias are also well described. Incidence of atrial arrhythmias ranging from 6% to 38% have been documented, with atrial fibrillation being the most

common [5]. An association has been noted between inducibility of ventricular arrhythmias and the occurrence of atrial arrhythmias in Brugada syndrome [6]. Atrial arrhythmias are an important cause of inappropriate shocks in those implanted with an implantable cardioverter defibrillator (ICD) in Brugada syndrome.

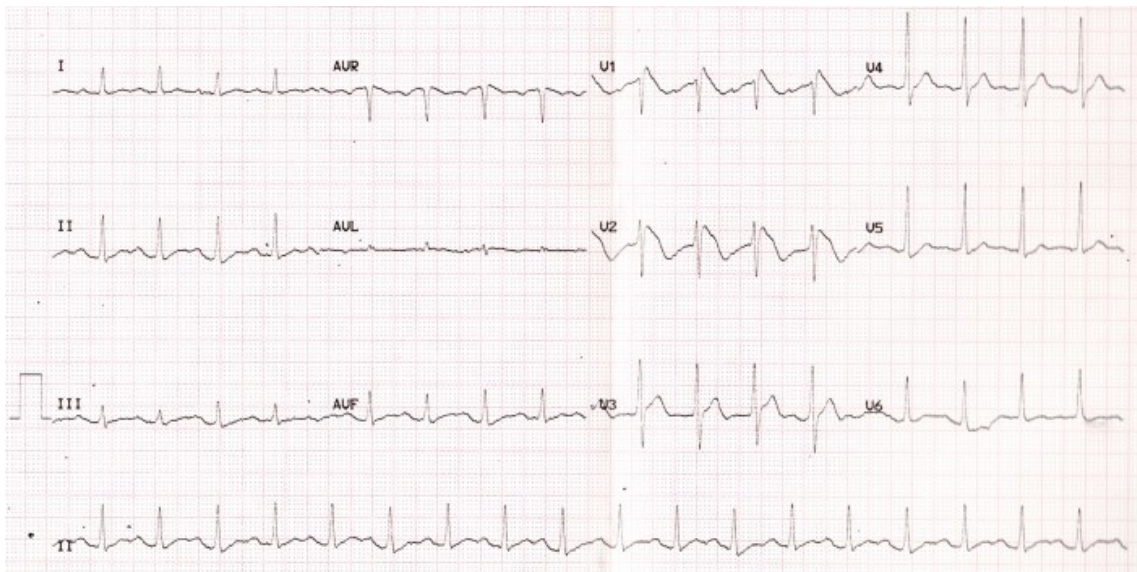


Figure 1: Brugada type 1 ECG. ST elevation with right bundle branch block pattern is seen in V1 and V2. ST elevation is maximal in V2.

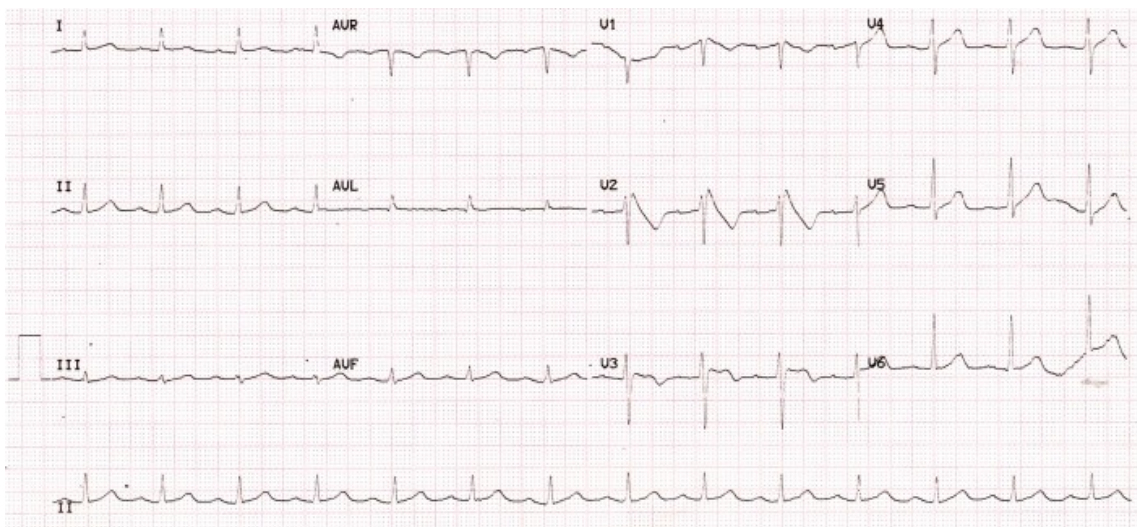


Figure 2: Saddle shaped ST segment with no significant ST elevation is seen in the first complex of V3 (type 3 pattern), while saddle shaped ST with elevation from baseline (type 2 pattern) is seen in the next two complexes. V2 shows type 1 pattern, thus illustrating the dynamic nature of ST segment in this case.

The only effective treatment for prevention sudden cardiac death in Brugada syndrome is implantation of an ICD. Recently totally subcutaneous ICDs have been used in Brugada syndrome as a less invasive option, though not devoid of disadvantages. The role of ICD in Brugada syndrome for secondary prevention after an episode of resuscitated sudden cardiac arrest or polymorphic ventricular tachycardia is unquestionable. But deciding who among the large number with a Brugada ECG pattern needs an ICD is very difficult. Prompt treatment of any febrile episode has been suggested as fever has been shown to precipitate ventricular arrhythmias in Brugada syndrome. Avoidance of drugs known to increase the chance of arrhythmia listed at brugadadrugs.org is of paramount importance. Holter monitoring and long term event monitoring is an option to look for clinically silent arrhythmias, the documentation of which may tilt the balance in favour of an ICD for secondary prevention.

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