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# Brugada Syndrome

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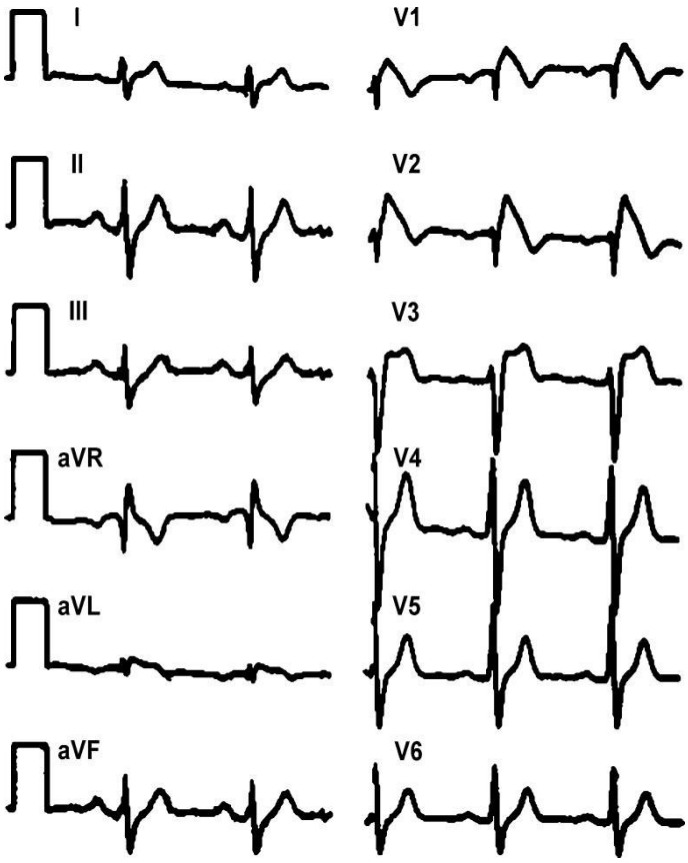
Initial description of Brugada syndrome in 1992 was that of syncopal episodes and/or sudden death in persons with structurally normal heart and a characteristic ECG pattern of right bundle branch block with ST segment elevation in leads V1 to V3 [1]. Sometimes individuals with a diagnostic ECG may be totally asymptomatic and may be having a family history of sudden death. Genetic nature of the disorder and mutation in sodium channel gene SCN5A was described in 1998 [2].




*With courtesy from [3] "Brugada J, Brugada P, Brugada R. Brugada syndrome: the syndrome of right bundle branch block, ST segment elevation in V1 to V3 and sudden death. Indian Pacing Electrophysiol J. 2001 Oct 1;1(1):6-11."*

I am always happy to see this ECG of Brugada syndrome as it was sent to me by Prof. Josep Brugada way back in 2001 for the inaugural issue of Indian Pacing and Electrophysiology Journal (IPEJ), along with his review article. Prof. Brugada's article was the first ever article which I received for IPEJ, when I started the journal. The article gave a great boost to the journal and later the journal was included in PubMed Central as the first online only Indian Journal to be included in the full text archive of US National Library of Medicine. It was an Indian Record, acknowledged by Limca Book of India Records in 2007.

Brugada syndrome manifests mostly in adulthood and has a definite male preponderance of almost eight-fold. Autosomal dominant inheritance pattern has been noted. Though mutations in several cardiac ion channels have been reported with Brugada syndrome, most important ones are in SCN5A gene. It may be noted that genotype positivity is not seen in many persons with diagnostic ECG pattern.

ECG in Brugada syndrome shows the characteristic coved ST elevation in anterior leads, well seen in V1 and V2 in the ECG shown above. Saddle shaped ST segment elevation seen in V3 was later described as type 2 pattern while the coved ST was considered as type 1 pattern. Saddle shaped ST with no significant ST elevation was called as type 3 pattern. But only type 1 pattern is diagnostic of Brugada syndrome. Other patterns may be converted to the diagnostic pattern by Flecainide or Ajmaline challenge test. Life threatening ventricular arrhythmias can occur during a drug challenge and should be undertaken only in a setup with facility to manage them quickly.

ECG pattern of Brugada syndrome can be mistaken for an ST elevation myocardial infarction (STEMI) if the person presents to the emergency department with chest pain for some reason. Clinical history and other investigations like cardiac troponin estimation and imaging studies are useful in ruling out ST elevation myocardial infarction in such cases. In a doubtful case coronary angiography may also be needed to exclude STEMI.

For some reason, occurrence of arrhythmias in Brugada syndrome is more likely when there is fever. Brugada syndrome, though world wide in occurrence, is more common in the Far Eastern region. Though most cases manifest in adulthood at an average age of around 45 years, it has also been considered as an important cause for Sudden Infant Death Syndrome (SIDS).

Online Mendelian Inheritance in Man (OMIM) has listed 9 genotypes for Brugada syndrome from BRGDA1 to BRGDA9. Number of genotypes may increase as still there are several cases which do not have an identified gene, but having features of Brugada syndrome.

Even though ventricular arrhythmias are the most dreaded arrhythmias in Brugada syndrome, atrial arrhythmias have also been described, with atrial fibrillation being commonest. It could be due to the enhanced duration of atrial action potential and increased intra-atrial conduction time [4]. Though some benefit has been noted with quinidine therapy in Brugada syndrome, the most important treatment is implantation of an Implantable Cardioverter Defibrillator (ICD). When an ICD is implanted for Brugada syndrome, atrial arrhythmias are a reason for the unwanted inappropriate ICD shocks, which can cause significant psychological distress and reduce battery life. Dual chamber ICDs with an extra atrial lead may be useful in discrimination of atrial from ventricular arrhythmias, though at an extra cost, reduced battery life and more problems likely due to the presence of an additional atrial lead. Careful programming of a single chamber ICD is another option to reduce inappropriate ICD shocks.

**References**

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