14. Management of Electrical Storm in a Patient With Brugada Syndrome

Rilong Hong, Chi Keong Ching, National Heart Centre Singapore, Singapore

Body

Background: The Brugada syndrome (BrS) is an inheritable arrhythmogenic disease that is associated with sudden cardiac death (SCD). Majority of BrS patients remain asymptomatic and fewer still present with electrical storm (ES).

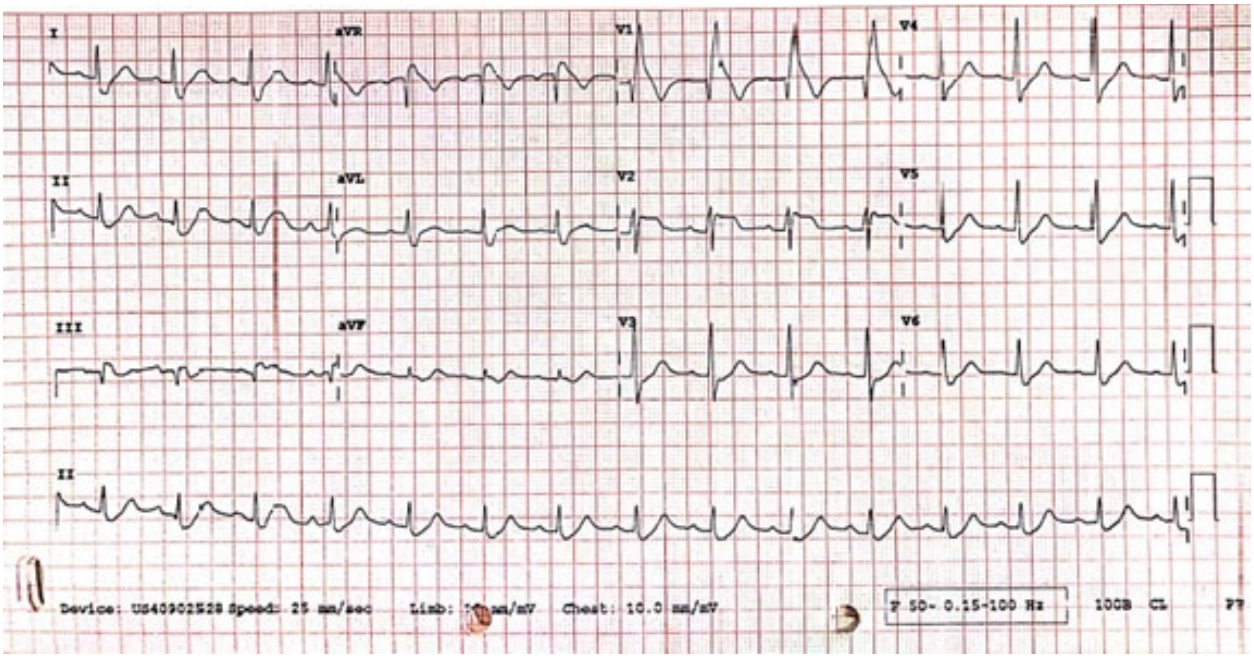
Case: A 48-year-old man was admitted for sudden onset syncope whilst in a recumbent position. This lasted for a few minutes, he woke up spontaneously complaining of palpitations and breathlessness.

The patient had a history of hyperlipidemia and non-anginal chest pain, with a normal treadmill echocardiogram performed a year prior. He had no family history of SCD or BrS.

He described episodes of waking at night with palpitations and shortness of breath for many years.

Clinical examination and initial investigations were unremarkable. His ECG revealed type 1 BrS pattern.

Image 1



During observation the patient developed multiple runs of unstable polymorphic ventricular tachycardia (VT), initiated by an R on T PVC; which required defibrillation.

He was started on isoprenaline at 40mcg per hour. Quinidine was subsequently started at 200mg Q8 hour. He had 3 further runs of polymorphic VT but by the following morning these had abated; he remained electrically quiescent for the rest of the admission.

Magnetic resonance imaging demonstrated a small non-transmural infarction at the basal inferior and inferoseptal wall with a focus of infarct/fibrosis at the basal inferior right ventricular wall. Invasive

coronary angiogram confirmed occluded distal RCA and 80% stenosis of the proximal LAD; percutaneous coronary intervention was performed to both vessels.

Discussion: ES was successfully suppressed by Quinidine and Isoprenaline. BrS is thought to occur due to mutations that decrease inward sodium current or increase transient outward potassium channel current (Ito). Both result in shifts in net transmembrane current at the end of phase 1 of the cardiac action potential (where Ito is most prominent). Quinidine with significant Ito blocking properties normalized action potentials on the surface ECG and prevented further ES. Isoprenaline helped to reduce vagal tone and increase intracellular calcium; thereby restoring the dome in phase 2 of the action potential.