



Ventricular fibrillation in a clinically normal heart

To the Editor: Sudden death as a result of ventricular fibrillation or fast ventricular tachycardia in the clinically normal heart is a preventable cause of death.

Prevention is only possible if the risk to an individual can be identified. Increasingly, repolarisation is being recognised as a time when various mechanisms or aberrations occur that could result in unstable ventricular rhythms.^{1,2} Repolarisation is the result of the sequential activation and deactivation of ion channels located in the sarcolemma of myocardial cells.³ One such disorder, the Brugada syndrome, has been recently described.¹ In this condition young men are prone to sudden death because of the occurrence of ventricular fibrillation due to a disorder of the sodium channel *SCN5A*.² Fibrillation usually occurs in the early hours of the morning.¹ The hallmark of the disease is partial right bundle branch block and ST segment changes in the anterior chest leads V1 to V3 on the surface ECG.¹ Electrophysiological studies are reportedly predictive in some studies² but not in others.⁴ A case is described here to draw attention to the value of the surface ECG in identifying individuals at risk for sudden death.

The patient, a 47-year-old man, had been well until August 1997 when he collapsed at a meeting. He was successfully resuscitated (including defibrillation) and transferred for further investigation and management. He had no added risks for ischaemic heart disease. On examination no abnormalities were detected and he suffered no neurological sequelae as a result of the cardiac arrest. The echocardiogram was entirely normal with no evidence of right ventricular dysfunction. The resting ECG was abnormal with ST segment elevation in V1 to V3 and a partial right bundle branch block (Fig. 1).

Angiographical examination revealed normal coronaries. Electrophysiological study showed easily inducible ventricular fibrillation with two extrastimuli at a cycle length of 500 msec.

Because of his history and the resting ECG changes the patient received an implantable defibrillator in 1997. There were no complications to the procedure. On 18 November 1998 he reported being woken from sleep with a shock. Interrogation of the device revealed an episode of spontaneous polymorphic ventricular tachycardia that rapidly degenerated into fibrillation which was successfully defibrillated (Fig. 2).

Discussion

To the best of our knowledge this is the first case of the Brugada syndrome to be described in South Africa.

The ECG features represent abnormalities in repolarisation.³ The action potential duration varies as a result of variability of cell types within the myocardium.⁴ This is attributable to differing expression of ion channels on cells in the different layers of the myocardium.⁴ This electrical heterogeneity causes

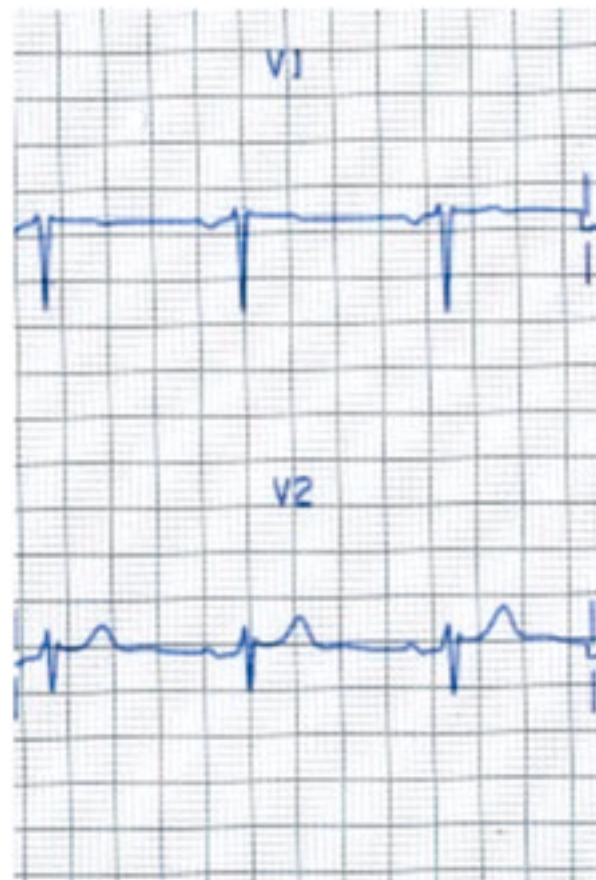


Fig. 1. Typical features of Brugada syndrome shown are ST elevation in V1 to V3 and in this case a partial right bundle branch block.

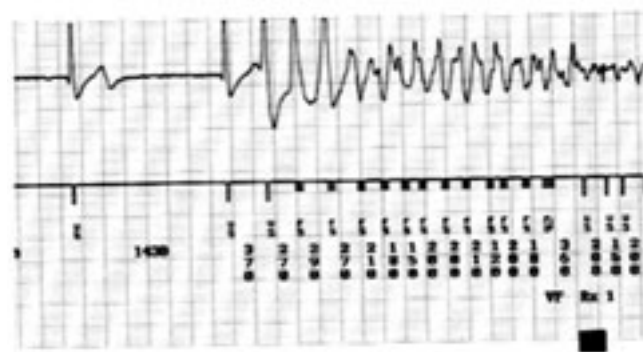


Fig. 2. Printout from the defibrillator showing the initiation of spontaneous ventricular fibrillation by a short run of ventricular tachycardia.

some cells within the wall of the heart to be repolarised while others are only partly repolarised.⁴ If the ion channels are abnormal and this alteration in function causes either



lengthening or shortening of the action potential then the electrical inhomogeneity will be exaggerated.⁴ This delay in repolarisation within the myocardium is reflected on the surface ECG.⁴ These after-depolarisations arise from already-repolarised cells reaching a state of depolarisation while other cells have only ended repolarisation.^{4,5} The occurrence of the after depolarisations in this critical period post repolarisation is sufficient to induce local re-entry resulting in polymorphic ventricular tachycardia which may degenerate into ventricular fibrillation.⁴

Successful treatment depends on identification of individuals at risk and the implantation of an automatic implantable cardioverter defibrillator.²

The importance of this syndrome is that the diagnosis is suggested by the surface ECG and that, once identified, appropriate steps can be taken to prevent sudden death. After the ECG pattern has been identified electrophysiological studies can be used to select high-risk patients; although the predictive value of positive electrophysiological study is debated, a negative study predicts a favourable outcome.^{2,5}

Currently the best predictors of mortality or further events are a family history of sudden death, the spontaneous occurrence of ST segment changes, and a history of syncope.¹

In summary, a case of Brugada syndrome is presented highlighting the usefulness of the ECG and the difficulties in treating these otherwise well patients, as well as interesting developments in the field of molecular electrophysiology.

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IN BRIEF

How accurate are 'guesstimates'?

If a doctor appearing as an expert witness or even an evidentiary witness in a trial is asked to describe an object or a lesion in terms of its size, it is assumed that that description is reasonably accurate. In order to assess the accuracy of 'guesstimation' by medical professionals, a team of researchers gave 11 forensic clinicians and 13 individuals from other professions ten test items and asked them to estimate their size.

The objects included a balloon (185 mm diameter), a red splotch (135 mm), a 50p UK coin (30 mm), a 10c US coin (15 mm), a scratch (18 mm), and a laceration (9 mm).

The sizes of the balloon, the scratch and the 10 cent coin were significantly overestimated, and both groups overestimated the size of the scratch. The forensic clinicians did not outperform the other participants, but they tended to be more accurate for small objects than for larger objects.

The taking of true measurements is advisable when the record of the injury may ultimately be used in court proceedings. When the use of a measuring device is deemed against the interests of the client and the examination, it is as well to be aware of one's limitations.

It has been shown in research conducted in 1992, that practice in estimating size can improve accuracy and precision. Repeated forensic examination of injuries, including recording of their dimensions, would provide such practice.

McLean I et al. *J R Soc Med* 2003; **96**: 497-498.