

Coronary Artery Bypass Surgery In A Patient With Asymptomatic Brugada Syndrome

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INTRODUCTION

Brugada syndrome (BS) is characterized by the diagnostic triad of a right bundle branch block (RBBB), ST segment elevation (ECG leads V1-V3) and sudden unexpected cardiac death (SUCD) due to polymorphic ventricular tachycardia (VT) or ventricular fibrillation (VF)¹. It is a rare inherited rhythm disorder with a familial predisposition (autosomal dominant) in 50% of cases. Mutations of the SCN5A gene (chromosome III) which encodes for the cardiac myocyte sodium channel are present in 20% of cases. BS usually occurs in structurally normal hearts but has been described in patients with right ventricular (RV) pathology and anomalous coronary anatomy. We report an asymptomatic BS patient who underwent successful bypass surgery (CABG) for concurrent coronary artery disease (CAD) and highlight some precautionary maneuvers that ensured a smooth perioperative course.

CASE REPORT

A 44-year-old hypertensive diabetic male non-smoker presented with unstable *Canadian Cardiovascular Society* (CCS) class 3-4 angina. He had a strong family history of SUCD when his brother suddenly collapsed and died atraumatically at age 25 years. Pre-operative electrocardiogram (ECG) demonstrated incomplete RBBB, ST segment elevation and a coving ST segment in V2, characteristic of a spontaneous Type 1 Brugada pattern (Figure 1).

Echocardiography demonstrated preserved LV function, no valve or regional wall motion abnormalities and a structurally normal RV. His coronary angiogram revealed severe complex triple vessel disease ; mid 95% Left anterior descending artery (LAD) bifurcation disease at the origin of the first diagonal artery, mid Circumflex artery 70% stenosis and mid 70% stenosis of an aneurysmal Right coronary artery (RCA) vessel which was technically not amenable for percutaneous intervention. Pre-surgery medication included acetylsalicylic acid, simvastatin and metformin. We decided to insert an implantable cardioverter defibrillator (ICD) following CABG as the patient was at intermediate risk of a future arrhythmia. He underwent an uneventful triple vessel CABG. His recovery was uneventful, arrhythmia-free and he was discharged home a week later. The patient was electively readmitted two months later for uneventful implantation of a single chamber (*Medtronic D284VRC Maximo II VR*) ICD following cessation of his anti-platelet therapy a week earlier.

DISCUSSION

Brugada syndrome was first described in 1992 as a leading cause of SUCD in young adult males particularly of South East Asian origin and may be responsible for 40-60% of cases of idiopathic VF¹. The prevalence of a Brugada ECG is 0.2-6% in the general population but whether it is a normal variant or a bad omen is debatable². Three distinct patterns are diagnostic; Type 1 is most characteristic with the coving elevated ST segment in V1-3 and has prognostic implications. Type 2 and 3 (saddleback) are subtle, can be a normal variant and generally carry a benign prognosis.

A mutational defect of the SCN5A gene results in a loss of the RV epicardium action potential dome resulting in transmural and epicardial dispersion of repolarization. The transmural dispersion underlies the characteristic ECG (ST segment elevation) and creates a vulnerable ventricular wall window. The epicardial dispersion of repolarization facilitates the development of a re-entry phenomenon in phase 2 of the cardiac myocyte action potential. This generates a re-entry extrasystole that captures the vulnerable window precipitating VT /VF.

The diagnosis in our Southeast Asian male patient was based on his pathognomonic spontaneous Brugada Type 1 resting ECG and strong family history of premature SUCD. A Brugada pattern can also be elicited with sodium channel blockers such as Ajmaline (Class 1A anti arrhythmic drug) or Flecainide (Class 1C) in susceptible individuals. This case was confounded by the presence of concurrent CAD however the patient had no previous myocardial infarction or regional wall motion abnormalities to suggest an ischaemic aetiology for the ECG findings.

Irrespective of his diagnosis of BS, our patient required surgery for symptomatic severe CAD. We modified our routine peri-operative care slightly with several simple but important maneuvers. The external defibrillator pads were applied to the patient's chest prior to the median sternotomy and once the heart was accessible, the internal defibrillator paddles were readily available to swiftly terminate any spontaneous VF/VT. We also commenced and maintained an isoprenaline infusion (class IIa indication) to attenuate any possible electrical storm precipitated by hypothermia or iatrogenic hyperkalaemic cardioplegic arrest³. The induction and maintenance of general anaesthesia was accomplished with the inhalational agent Sevoflurane and avoidance of

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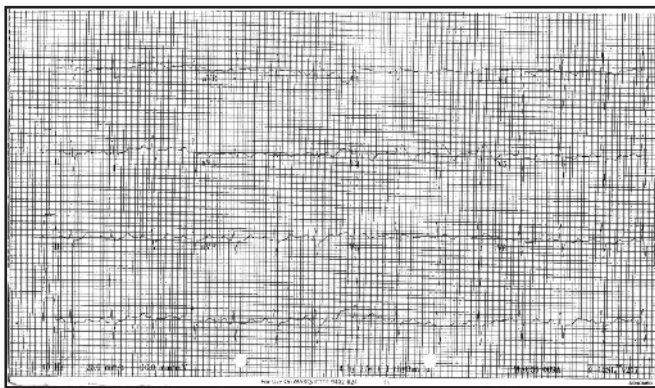


Fig. 1 : ECG demonstrating incomplete RBBB, ST segment elevation with coving in V2, pathognomonic of Type 1 spontaneous Brugada pattern.

sodium channel blockers. The patient's electrolytes were meticulously monitored and minor imbalances swiftly corrected. A magnesium sulphate infusion was commenced prophylactically in the early post-operative period to enhance myocardial membrane stability. Finally the external defibrillator pads were re-applied to the patient with a defibrillator machine strategically placed by his bedside and the epicardial atrio-ventricular pacing wires attached to a temporary dual chamber pacemaker on a "back-up" sensing mode. This arrangement was maintained till his discharge from hospital.

ICD implantation is currently the only proven therapy for symptomatic BS patients as the onset of VF/VT is unprovoked, unpredictable and invariably fatal if untreated (class 1 indication)³. For asymptomatic BS patients, the decision is less clear as the procedure is invasive, costly and not without risk. Brugada et al suggest inducible VF electrophysiology studies (EPS) are a valuable prognostic tool in evaluating asymptomatic individuals however their findings have controversially not been validated in subsequent studies^{4,5}. Differences in stimulation protocols and EPS-positive criteria may explain the conflicting data. Other suggested criteria to identify high risk individuals for ICD implantation include clinical profile, first degree relatives with a history of SUCD, ECG morphology, and exercise stress testing as ST segment elevation during recovery from exercise has been associated with a significantly higher risk of future arrhythmic events in Brugada patients.

Established *American College of Cardiology (ACC)/American Heart Association (AHA)* guideline recommendations for ICD implantation in BS patients who have a reasonable functional status and predicted life expectancy beyond a year includes individuals with a previous cardiac arrest despite optimal medical therapy (class I indication), spontaneous ST-segment elevation in V1, V2 or V3 with syncope (class IIa) and those with documented episodes of VT (class IIa)³. Despite the

absence of such criteria, our decision to insert an ICD in this asymptomatic patient was based on the knowledge that prognosis is worst in male patients with a spontaneous Type 1 ECG⁶. Prophylactic ICD implantation for the primary prevention of SUCD may be justified in such individuals.

Non-interventional therapies include administration of quinidine or isoproterenol to restore the epicardial action potential dome, normalize the ST segment and prevent a phase 2 reentry, a reasonable strategy for the low-risk asymptomatic patient.

CONCLUSION

We report to the best of our knowledge the first case of CABG surgery in a patient with asymptomatic BS and significant concurrent CAD. Although the two conditions are unrelated, this case highlights several simple but important precautionary peri-operative maneuvers that will help attenuate the risk of a potentially fatal arrhythmia at the time of coronary revascularization.

The diagnosis of BS can be elusive as some ECG features may be a normal variant.

The decision whether to insert an ICD in an asymptomatic patient like ours, is difficult and requires astute clinical risk assessment of various prognosticators such as a characteristic (type 1) ECG and/or significant family history. Currently, EPS appears to have a limited prognostic value due to conflicting data and hence may not necessarily assist with the decision making process.

REFERENCES

1. Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: a distinct clinical and electrocardiographic syndrome: a multi center report. *J Am Coll Cardiol* 1992; 20: 1391-6.
2. Oto A. Brugada sign: A normal variant or a bad omen? Insights for risk stratification and prognostication. *Eur Heart J* 2004; 25: 810-1.
3. Zipes DP, Camm AJ, Borggrefe M, Buxton AE, Chaitman B, Fromer M, et al. ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force and the European Society of Cardiology Committee for Practice Guidelines (Writing Committee to Develop Guidelines for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death). *J Am Coll Cardiol*. 2006 Sep 5; 48(5): e247-346.
4. Brugada P, Brugada R, Mont L, Rivero M, Geelen P, Brugada J. Natural history of Brugada syndrome: the prognostic value of programmed electrical stimulation of the heart. *J Cardiovasc Electrophysiol* 2003; 14: 455-7.
5. Priori SG, Gasparini M, Napolitano C, Bella AD, Ottonelli Ag, et al. Risk stratification in Brugada Syndrome; Results of the PRELUDE (Programmed Electrical stimulation predictive value) Registry. *J Am Coll Cardiol* 2012; 59(1): 37-45.
6. Gehi Ak, Duong TD, Metz LD, et al. Risk stratification of individuals with the Brugada electrocardiogram: A meta analysis. *J Cardiovasc Electrophysiol* 2006;17: 577-83.