

Case Report

Brugada Syndrome: A Review of Peri-operative Management for the Anesthesiologist

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Abstract

Brugada syndrome (BS) is a rare cardiac channelopathy that was first described in 1992. It accounts for four percent of all sudden deaths and up to twenty percent of sudden deaths in patients without structural cardiac defects. Since known and unknown BS patients can present for surgery at any time, it is important for the anesthesiologist to be familiar with this syndrome and how to safely manage these patients peri-operatively. A case study is described in which a 53 year old woman with BS required emergent embolization of her left facial artery and bilateral internal maxillary arteries due to severe epistaxis on two consecutive days. The etiology, mechanism and proper peri-operative management of BS are then reviewed, and a summary of appropriate medications is provided.

INTRODUCTION

It is important for the anesthesiologist to be familiar with Brugada syndrome (BS) since known and unknown BS patients may present for surgery. BS accounts for four percent of all sudden deaths and up to twenty percent of sudden deaths in patients without structural cardiac defects [1]. Proper identification of BS is important as the two year mortality approaches 30 percent [2]. Review of the literature reveals a series of case reports detailing a variety of methods of administering a successful anesthetic to BS patients including general, sedation, and regional anesthesia [3]. Nevertheless, there are certain guidelines to follow intraoperatively in order to minimize the risk of adverse cardiac events.

Brugada syndrome was first identified in 1992 and describes patients at risk of ventricular arrhythmias and sudden death who have a structurally normal heart [4]. Syncope and sudden death occur without warning and are caused by fast polymorphic ventricular tachycardia or fibrillation [2]. Of note, the characteristic electrocardiograph (ECG) findings can be transient with "pseudo-normalization" of the ECG [5]. Three ECG patterns have been identified; with type 1 being the only one diagnostic of BS [6] (Figure 1). In this type, a coved ST-segment elevation ≥ 2 mm is followed by a negative or flat T wave [5].

A right bundle branch block may be present but is not required for diagnosis [1]. Between 20 and 30% of patients have concomitant supraventricular tachycardias, most frequently atrial fibrillation, which may be the first manifestation of the disease [5]. The Brugada ECG pattern is more pronounced with

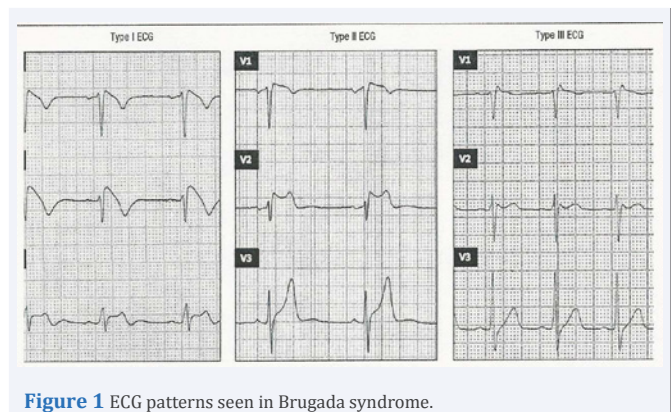


Figure 1 ECG patterns seen in Brugada syndrome.

sleep, rest, or after eating when vagal activity dominates and has been found to occur in sudden unexpected nocturnal death syndrome (SUNDS) and may be linked to sudden infant death syndrome (SIDS) [1]. The clinical presentation of BS varies from patients being asymptomatic, to having a history of syncope, seizures, palpitations, nocturnal agonal respiration, and aborted sudden death [5].

The prevalence of BS differs based on population and is more common in Southeast Asia (12/10,000) than in Europe (1-5/10,000) [1]. The average age of diagnosis is 40-45 years and 80% of patients are men [1]. BS has an autosomal dominant inheritance with variable penetrance [1], and the majority of patients have a family history of sudden death or malignant arrhythmias [2]. An implantable cardioverter-defibrillator (ICD)

is the only effective treatment and is recommended for patients who are symptomatic or have a spontaneous BS ECG type 1 pattern inducible with electrophysiological studies [1].

Mutations in ten genes have been linked to BS, the most common being a SCN5A mutation that affects cardiac sodium channel conduction [1]. The prevalence of SCN5A mutations is approximately 18-30% and causes reduced cardiac sodium current [5,6]. Other mutations lead to a decrease in L-type calcium current and decreased potassium transient outward current [7]. These electrolyte abnormalities lead to right ventricular conduction disturbances and the associated Brugada ECG pattern [7]. At this time, it is unclear whether the exact mechanism is due to an abnormal depolarization or repolarization pattern, or possibly both [1].

We report a case in which a patient with known BS received a successful general anesthetic on two consecutive days. A review of the literature is also presented along with a summary of perioperative care recommendations.

CASE REPORT

A 53 year old 63 kg woman presented for emergency embolization of her left facial artery and bilateral internal maxillary arteries due to severe epistaxis on two consecutive days. The patient had a history of BS that had been recently diagnosed at age 50 and she did not have an ICD. She was asymptomatic with a good functional status and otherwise healthy. Her pre-operative ECG showed a sinus rhythm with a heart rate of 86 beats per minute, a right bundle branch block, and a coved ST segment elevation >2mm in V1 and V2 followed by a negative T wave (Figure 2). This ECG pattern is consistent with the type I BS ECG.

Of note, the patient's brother and father also had BS. For both embolizations, defibrillator pads were placed pre-operatively as the patient did not have an ICD. Two peripheral intravenous lines and an arterial line were also placed. The patient was premedicated with midazolam for both procedures. The patient was normotensive for both procedures and a normal sinus rhythm between 60 and 100 beats per minute was maintained. Normothermia was maintained during both procedures using a forced air under body warming blanket. For the first

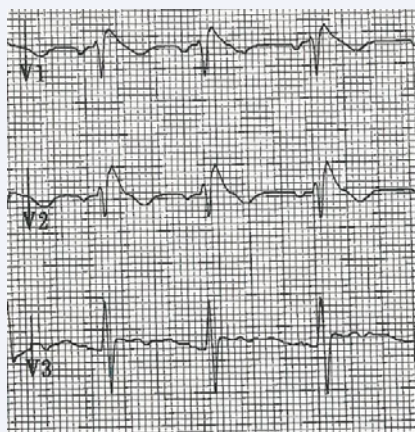


Figure 2 ECG of Patient in Case Report.

embolization, a rapid sequence induction was performed with fentanyl, propofol, and succinylcholine as the patient had severe epistaxis and had been swallowing a lot of blood. General anesthesia was maintained with sevoflurane and the patient was given rocuronium. The patient received dexamethasone and ondansetron for anti-emesis and paralysis was uneventfully reversed with glycopyrrolate and neostigmine. A second embolization had to be performed the next day due to renewed bleeding. General anesthesia was induced with lidocaine, fentanyl, propofol, and rocuronium, and was maintained with isoflurane. 20mg of esmolol was given in order to maintain a heart rate less than 100 beats per minute. The patient received metoclopramide and ondansetron for anti-emesis and paralysis was again reversed with glycopyrrolate and neostigmine. There were no complications during or after either procedure, including further ST segment changes or hemodynamic changes. The patient remained in a telemetry unit throughout her hospital stay and was discharged home two days after her second embolization.

Summary of recommended perioperative management

Pre-operatively, any patient with a type 1 ECG pattern, whether having a known diagnosis of BS or not, should be treated as having BS [8]. Patients with an ICD in place should have it turned off preoperatively as applicable [8,9]. All BS should have external defibrillator pads placed [8-14]. Electrolyte abnormalities should be corrected preoperatively as hyper- and hypokalemia and hypercalcemia can worsen BS ST segment elevation [8]. As autonomic changes can precipitate arrhythmias, adequate depth of anesthesia should be ensured and postural changes should be minimized [10]. Increased vagal tone has been implicated in the development of Brugada ECG changes [8]. The patient should be kept normothermic, as hyperthermia in particular can unmask BS due to premature inactivation of the sodium channel [6,8]. Some authors recommend the routine use of arterial lines in all cases in order to accurately monitor hemodynamic changes and the placement of central lines for major cases [8-11]. Patients with an ICD should have it reactivated postoperatively [8,9,11,12,15]. Patients should be observed via telemetry up to 36 hours after surgery as arrhythmias are more likely to occur postoperatively [10].

Benzodiazepines, narcotics, and ketorolac have not been associated with any adverse events [3,9-12,15,16]. Regional and neuraxial anesthesia have been successfully performed using a variety of local anesthetics including lidocaine, mepivacaine, bupivacaine and ropivacaine [3,9,10,16]. For induction of general anesthesia, propofol and various barbiturates have been successfully used [3,8-13,15]. Etomidate has been associated with one report of self-limited ST segment elevation [16]. High dose ketamine should be used with caution [8]. Although propofol drips have been successfully used in some reports, they are best avoided due to concern about the development of propofol infusion syndrome and potential alteration in ion channel function [16,17]. General anesthesia has been successfully maintained with nitrous oxide, desflurane, sevoflurane, and isoflurane [3,9-12,15,16], although it has been suggested that sevoflurane might be best since it has no effect on QT length [8]. Depolarizing and non-depolarizing paralytics are safe to use [3] as are reversal agents [9,12,13,16]. It is recommended to give reversal agents slowly as anticholinergics can cause tachycardia and neostigmine can cause ST elevation [8]. Atropine [15], glycopyrrolate, and dopamine have been used to treat

bradycardia [8,14]. Hypotension has been treated with ephedrine and phenylephrine [16]. Beta blockers and alpha agonists should be used with caution as they can increase ST elevation and unmask a Brugada ECG pattern [8,16]. Isoproterenol has been used to reduce ST segment elevation and suppress arrhythmias [8,16]. Commonly used anti-emetics such as dexamethasone, droperidol and ondansetron have been used without incident [3,16].

CONCLUSION

Current understanding is that BS is caused by various rare channelopathies resulting in aberrant cardiac conduction and a characteristic ECG in patients with a structurally normal heart. Inheritance is autosomal dominant and patients are at increased risk of sudden death, particularly due to ventricular fibrillation or tachycardia. As these patients may present for surgery with no warning as the patient in this case report did, it is important for the anesthesiologist to be familiar with this syndrome in order to provide a safe anesthetic and perioperative experience.

Summary of anesthetic drug recommendations in patients with bs¹⁷ (in a box on the side of the page)

Safe:

Benzodiazepines, narcotics, ketorolac

Regional and neuraxial anesthesia with a variety of local anesthetics

Propofol and barbiturates

Nitrous oxide, all volatile anesthetics

All depolarizing and non-depolarizing paralytics

Muscle relaxant reversals

Ephedrine, phenylephrine, dopamine

Isoproterenol

Dexamethasone, ondansetron, droperidol

Contraindicated (Class I):

Flecainide, procainamide

Preferably avoided (Class IIa and IIb):

Propofol infusion, alpha agonists, beta blockers, verapamil, amiodarone, cocaine, ergonavine, ketamine, tramadol, metoclopramide, diphenhydramine, dimenhydrinate, edrophonium, carbamazepine, phenytoin

No clear evidence of adverse effects (Class III):

Diltiazem, nifedipine, nitroglycerin

Class I- Strong evidence and general agreement that drug is potentially arrhythmic in BS patients

Class IIa- Conflicting evidence but weight/opinion is in favor of potentially arrhythmic effect in BS patients

Class IIb- Conflicting evidence but weight/opinion, potential arrhythmic effect less well established in BS patients

Class III- Little to no evidence of potential arrhythmic effect in BS patients

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