Original Article:

Anaesthetic considerations of a rare channelopathy- brugada syndrome"- a case report

Dr. Manjunatha R Kamath*, Dr Namitha C N**

*Additional Professor, Department of Anaesthesiology

** Junior Resident, Department of Anaesthesiology

K S Hegde Medical Academy, Mangalore

Corresponding author**

Abstract:

Brugada Syndrome (BS) is an autosomal dominant channelopathy with variable penetrance affecting the cardiac sodium channels which results in a characteristic electrocardiographic findings in precordial leads V_1 to V_3.[1,2] The majority of patients have uneventful course peri-operatively. However, there is a risk of worsening ST elevation and ventricular arrhythmias due to perioperative use of medications, surgical insult, electrolyte abnormalities & hyperthermia. Hence safe conduct of anaesthesia depends upon a detailed knowledge of these conditions.[3]

Massive bleeding can occur during resection of a retroperitoneal mass following an injury to adjacent major vessels or organ. Management includes the use of blood products, coagulation factor concentrates, pharmacological agents and vasopressors.

Key words: Brugada syndrome; Perioperative arrhythmia; Sudden cardiac death; Massive blood loss; Retroperitoneal liposarcoma

Introduction:

In 1992 Brugada and Brugada described a rare autosomal dominant channelopathy with variable penetrance affecting the sodium channels, caused by mutations in the gene SCN5A on chromosome 3, encoding the human cardiac sodium channel called 'Brugada syndrome'.[1,2] Its clinical presentation varies from being asymptomatic to having syncope, palpitation and even sudden cardiac death in patients with a structurally normal heart with an electrocardiogram (ECG) characteristic of right bundle branch block (RBBB) with ST segment elevation in leads V1 to V3 for its diagnosis.[3,4] These patients are at risk of developing malignant arrhythmias and sudden cardiac death on exposure to various triggers during the perioperative period, which may pose a significant risk to the patient.[5]

Case report:

A 51 year male patient, a case of retroperitoneal liposarcoma with BS underwent laparotomy under combined general and epidural anaesthesia. Incidental diagnosis of BS was made during routine pre-operative evaluation. No past history of syncope and no family history of sudden cardiac death. ECG showed RBBB with ST elevation in leads V1-V3 (Figure 1). Echocardiography was normal with ejection fraction of 60%. The patient was explained about the procedure & possible perioperative risks and consent was taken for the same.

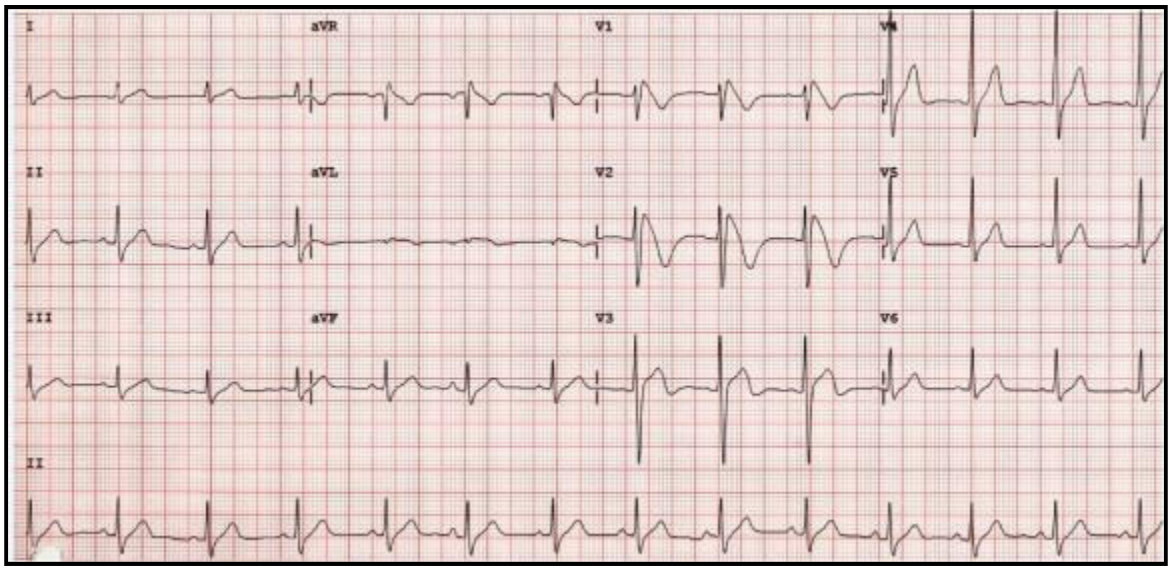


Figure 1: Preoperative electrocardiogram showing the right bundle branch block and coved type ST segment elevation in leads V1-V3

On the day of surgery, patient's identity was confirmed, consent form checked and availability of ICU bed and adequate blood products confirmed. The patient was shifted to operation theatre, monitors attached (5 lead ECG, pulse oximetry, non invasive blood pressure). Baseline vitals noted. The patient was positioned in the lateral position. Under all aseptic precautions epidural was placed at L1- L2 inter-space using 18G Tuohy needle and a test dose 3ml of 2% lignocaine with adrenaline was injected. In supine position preoxygenation done with 100% oxygen for 3min. Analgesia achieved with Morphine 5mg intravenously(IV). Induced with Propofol 100mg IV. After confirming adequate mask ventilation, Inj. Vecuronium 5mg IV given. Airway secured with cuffed endotracheal tube(8mmID) and connected to a ventilator. Anaesthesia maintained with oxygen and nitrous oxide (1:1) with isoflurane 1%. An arterial and a central venous (right subclavian vein) lines were secured. Intraoperatively patient developed life threatening acute blood loss of 4.5L. Monitors showed hypotension, tachycardia and non specific ST-T changes in ECG. It was managed successfully by control of surgical bleeding, supportive treatment with rapid fluid infusion, massive transfusion of blood products and administration of intravenous vasoactive agents along with prevention and treatment of complications such as hypothermia, acid base imbalance and electrolyte abnormalities. The patient received a total of 10units of Packed RBC, 8units of FFP, 4units of platelets and 2 hydroxy ethyl starch intraoperatively. Surgery was abandoned after packing the abdomen with mops. Postoperatively, he was transferred to ICU with mechanical ventilator support, where he received another 4 units of FFP, 2 units PRBC. A defibrillator was kept standby throughout. Continuous ECG monitoring was done. Serum electrolytes were monitored. Normothermia was maintained. Postoperative analgesia maintained with epidural infusion(ropivacaine 0.1%). He was extubated on post-op day1(POD1). His condition improved over next two days. He was taken up for surgery again with an uneventful intraoperative course. No untoward ECG changes (fatal arrhythmias) noted during the stay in the hospital. He was discharged from hospital on POD10.

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

Discussion:

Brugada syndrome is an autosomal dominant genetic disorder associated with an increased risk of sudden cardiac death (SCD) and ventricular tachyarrhythmia's.[1,2] Also referred to as sudden unexpected death syndrome or sudden unexpected nocturnal death syndrome.[5]

It has a prevalence of 1 in 2000.[5] It is more common in men than women and is responsible for 4% of all sudden deaths and 20% of sudden deaths in patients without any structural heart disease.[6]

There is no underlying structural heart disease by echocardiography but have distinctive electrocardiographic findings (Figure 2). Three types of BS has been described(Table 1)[1,2]

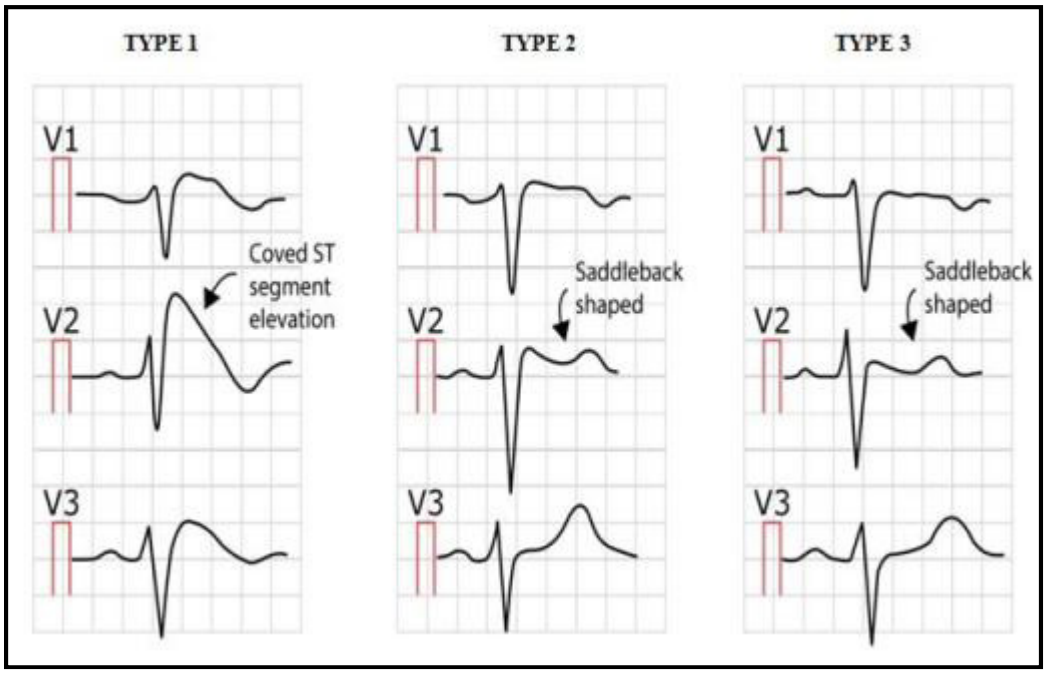


Figure 2: ECG patterns of Brugada syndrome

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

Drug challenge with sodium channel blockers such as Pilsicainide and Flecainide is a standard provocative test used to unmask BS. It can induce significant ST segment elevation>0.10 mV. [5]

Various perioperative triggers that can induce ECG changes in BS patients are shown in (Table 2) [1-4]

Table 1

	Type 1 BS	Type 2 BS	Type 3 BS
T wave	Negative	Positive or biphasic	Positive
ST-T configuration	Coved	Saddle back	Coved or saddle back
ST segment(terminal portion)	≥2 mm or 0.2 mV, gradually descending	>1mm, sloping toward the baseline before an upright T wave	Elevated , <1mm
J wave amplitude	>=2mm	>=2mm	>=2mm

Table 2: Triggers for ECG changes in BS perioperatively

Autonomic imbalance	Hyperthermia
Electrolyte abnormalities	Bradycardia
Myocardial ischemia	Sodium channel blockers
Beta blockers and Alpha agonists	Vagotonic drugs(Neostigmine
Cholinergics (Acetyl choline, Edrophonium)	Calcium channel blockers
Psychotropic drugs	

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

The implications of drugs used perioperatively (Table 3) [7]

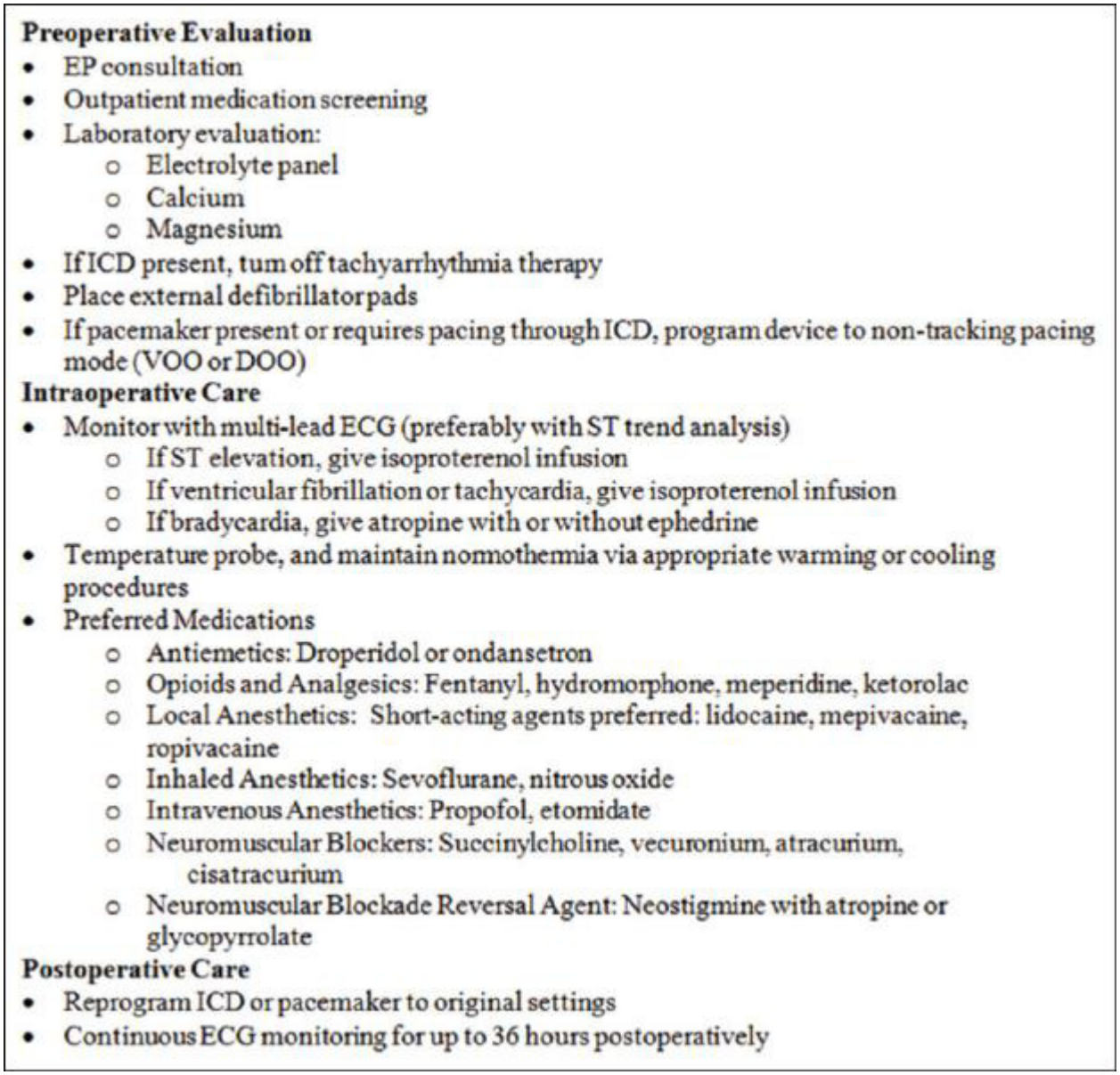
Drugs	Considerably safe	To be avoided
General anaesthetics	Fentanyl, Thiopentone, Midazolam, Isoflurane, Sevoflurane, N2O	Propofol infusion
Muscle relaxants	Vecuronium, Mivacurium, Rocuronium	
Local anaesthetics	Lignocaine	Bupivacaine (epidural)
Psychotropic drugs		Amitriptyline, Desipramine, Lithium, Loxapine, Trifluoperazine
Anti arrhythmics	Lignocaine, Mexiletine, Quinidine, Dysopyramide Amiodarone	Procainamide Flecainide Pilsicainide
Others	Atropine, Ephedrine, Propronolol, Isoproterenol	Noradrenaline Acetyl Choline, Cocaine Neostigmine

Table 3: Implications of drugs used perioperatively in BS patients

So to avoid perioperative arrhythmias and SCD it is mandatory to correct electrolyte abnormalities, maintain normothermia & continuous ECG monitoring perioperatively. Invasive arterial line and central venous cannulation are mandatory for all major cases associated with a significant fluid shift. External defibrillator pads should be attached to all patients routinely. Patients on implantable cardioverter defibrillator (ICD) should be turned off during the surgery. After surgery, patients with a pacemaker or ICD should have the settings restored to its preoperative mode. Due to the risk of postoperative arrhythmias, continuous ECG monitoring should continue for up to 36 h postoperatively. [3,5,6]

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

Checklist for perioperative management of BS (Figure 3). [5]



Preoperative Evaluation

- EP consultation
- Outpatient medication screening
- Laboratory evaluation:
  - Electrolyte panel
  - Calcium
  - Magnesium
- If ICD present, tum off tachyarrhythmia therapy
- Place external defibrillator pads
- If pacemaker present or requires pacing through ICD, program device to non-tracking pacing mode (VOO or DOO)

Intraoperative Care

- Monitor with multi-lead ECG (preferably with ST trend analysis)
  - If ST elevation, give isoproterenol infusion
  - If ventricular fibrillation or tachycardia, give isoproterenol infusion
  - If bradycardia, give atropine with or without ephedrine
- Temperature probe, and maintain normothermia via appropriate warming or cooling procedures
- Preferred Medications
  - Antiemetics: Droperidol or ondansetron
  - Opioids and Analgesics: Fentanyl, hydromorphone, meperidine, ketorolac
  - Local Anesthetics: Short-acting agents preferred: lidocaine, mepivacaine, ropivacaine
  - Inhaled Anesthetics: Sevoflurane, nitrous oxide
  - Intravenous Anesthetics: Propofol, etomidate
  - Neuromuscular Blockers: Succinylcholine, vecuronium, atracurium, cisatracurium
  - Neuromuscular Blockade Reversal Agent: Neostigmine with atropine or glycopyrrolate

Postoperative Care

- Reprogram ICD or pacemaker to original settings
- Continuous ECG monitoring for up to 36 hours postoperatively

Figure 3: Perioperative checklist for managing patients with Brugada syndrome [5]

It is imperative to recognize major blood loss early and institute effective action promptly if shock and its consequences are to be prevented. Massive blood transfusion and its related complications such as electrolyte abnormalities (hyperkalemia) or febrile reactions can trigger malignant arrhythmias in BS patients. Hence it is mandatory to keep a defibrillator ready if such acute events are anticipated.

Therapeutic goals during massive blood loss:[8,9]

- Maintenance of tissue perfusion and oxygenation by restoration of blood volume and hemoglobin
- Control of surgical bleeding
- Judicious use of blood component therapy
- Risks of massive blood transfusion(Table 4) - should be treated promptly if occurs

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

Acute complications	Delayed complications
Febrile non hemolytic transfusion reaction	Hemolysis
Transfusion related acute lung injury	Transfusion associated Graft versus host disease
Transfusion associated cardiac overload	Infections
Allergic reactions	Post-transfusion purpura
Hyperkalemia	
Dilutional coagulopathy	
Thrombocytopenia	
Electrolyte and acid-base imbalance	
Multi organ failure	

Table 4: List of possible complications following massive blood transfusion

Conclusion:

Acute surgical hemorrhage is a medical emergency with a high mortality rate hence requiring prompt and effective treatment including control of bleeding, transfusion of blood and blood products, maintaining normothermia, correcting electrolyte & acid-base abnormalities for maintaining adequate tissue perfusion and oxygenation.

BS has been recognized as an important cause of SCD in relatively young individuals. Many drugs have been reported to induce the characteristic Brugada pattern ECG abnormalities and/or fatal ventricular tachyarrhythmias. So adverse cardiac events in these patients can be prevented by avoiding drugs which trigger ECG changes, correct electrolyte abnormalities, maintain normothermia and most importantly vigilant monitoring of ECG perioperatively.

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 multicenter report. J Am Coll Cardiol 1992;20:1391-6.
2. Brugada R, Brugada J, Antzelevitch C, Kirsch GE, Potenza D, Towbin JA, *et al*. Sodium channel blockers identify risk for sudden death in patients with ST-segment elevation and right bundle branch block but structurally normal hearts. Circulation 2000;101:510-5.
3. Carey SM, Hocking G. Brugada syndrome – A review of the implications for the anaesthetist. Anaesth Intensive Care 2011;39:571-7.
4. Bayés de Luna A, Brugada J, Baranchuk A, Borggrefe M, Breithardt G, Goldwasser D, *et al*. Current electrocardiographic criteria for diagnosis of Brugada pattern: A consensus report. J Electrocardiol 2012;45:433-42.

5. Sorajja D, Ramakrishna H, Poterack AK, Shen WK, Mookadam F. Brugada syndrome and its relevance in the perioperative period. Ann Card Anaesth 2015;18:403.
6. Smith D, Martz DG. Brugada Syndrome: A Review of Peri-operative Management for the Anesthesiologist. Int J Clin Anesthesiol 2014;2:1019-21.
7. Postema PG, Wolpert C, Amin AS, Probst V, Borggrefe M, Roden DM, *et al*. Drugs and Brugada syndrome patients: Review of the literature, recommendations, and an up-to-date website (www. brugadadrugs. org). Heart Rhythm 2009;6:1335-41.
8. Pham HP, Shaz BH. Update on massive transfusion. Br J Anaesth 2013;111:71-82.
9. Stainsby D, MacLennan S, Thomas D, Isaac J, Hamilton PJ. Guidelines on the management of massive blood loss. Br J Haematol 2006;135:634-41.
10. Johansson PI, Ostrowski SR, Secher NH. Management of major blood loss: An update. Acta Anaesthesiol Scand 2010;54:1039-49.