

Anaesthetic Management of Rare Case of Arrhythmogenic Right Ventricular Dysplasia (ARVD) with Fracture Patella under Combined Sciatic Femoral Nerve Block

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Abstract

Arrhythmogenic right ventricular dysplasia (ARVD) is a rare but severe familial disease, with autosomal dominant inheritance and polymorphic phenotype expression. Its prevalence is estimated to be approximately 1 in 5000. Sporadic cases have been described in young adults and children with no clinically apparent cardiac disorders who experienced exercise-induced sudden deaths and complex ventricular arrhythmias. Antiarrhythmic drugs therapy is the first step of treatment, but the severity of ventricular tachycardia may necessitate surgical or 45yr old male patient with ARVD with fracture patella done under lower limb block successfully. The aim of this case presentation to describe clinical characteristics & anaesthetic specifics in the management of ARVD & how we managed case of ARVD with fracture patella & combined sciatic femoral nerve block was proven to be boon in such scenario.

Key Words: Arrhythmogenic right ventricular dysplasia (ARVD), Combined sciatic femoral

Introduction

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is a genetic cardiomyopathy characterised by fibro fatty replacement primary of right ventricular myocytes and electrical instability associated with ventricular tachycardia (VT). Genetically it is Autosomal dominant inheritance with prevalence 1:5000. Mostly affects young adult men approximately three times more frequently than women. ARVD as a disorder characterised by progressive loss of the myocardium & its replacement by fibro fatty tissue

Clinical spectrum can range from symptomless presentation to sudden cardiac death. Most common age of presentation is 10-50 years with mean age ~30 years. Patient generally present with Symptoms Like palpitations, syncope, atypical chest pain & Dyspnoea with Signs of RVF & Arrhythmias.

Four phases were proposed for ARVD in literature [1]. One is concealed phase which is characterised by subtle right ventricular structural abnormalities with or without VTs. Second is overt arrhythmia phase which contains symptomatic ventricular arrhythmias associated with overt structural and functional RV abnormalities.

Third is isolated right heart failure and fourth one is biventricular failure which contains significant LV involvement mimicking dilated cardiomyopathy.

Diffuse or segmental loss of right ventricular myocytes with replacement of right ventricular myocytes with replacement of fibro-fatty tissue and thinning of right ventricular wall. Most commonly affected area in the right ventricle are-RV outflow tract, Apex & Subtricuspid area [1-4]

ARVD is difficult to diagnose when asymptomatic. First presentation is cardiac arrest in up to 50% of the clinical cases. Post mortem examination confirms fibro-fatty histology. Screening of family members is important to identify and risk stratify genetically affected relatives to prevent SDC.

Diagnostic test specific for ARVD has not been invented. In 1994, the International Task Force proposed the first diagnostic criteria system and combined multiple diagnostic information (structural, histological, arrhythmic, echocardiographic, genetic and familial features. Electrocardiogram (ECG) abnormalities observed include T-wave inversion in anterior precordial leads, left axis deviation, wide QRS complexes and epsilon waves.

Ventricular tachy-arrhythmias may present in a wide spectrum on resting ECG from premature ventricular extrasystoles, non-sustained ventricular tachycardia and monomorphic ventricular tachyarrhythmia with left bundle branch block.

Figure 1: Complete or incomplete RBBB/QRS prolongation (in absence of RBBB)

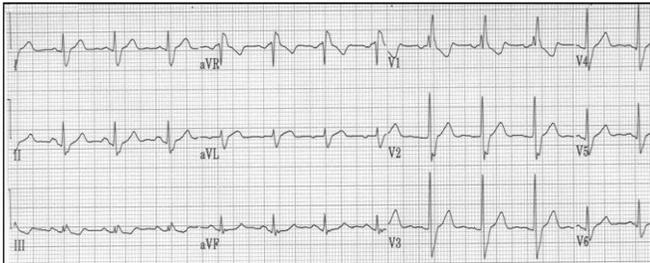


Figure 2: Twelve-lead electrocardiogram leads with regular sinus rhythm and an epsilon waves (arrow) in leads V1-V3.

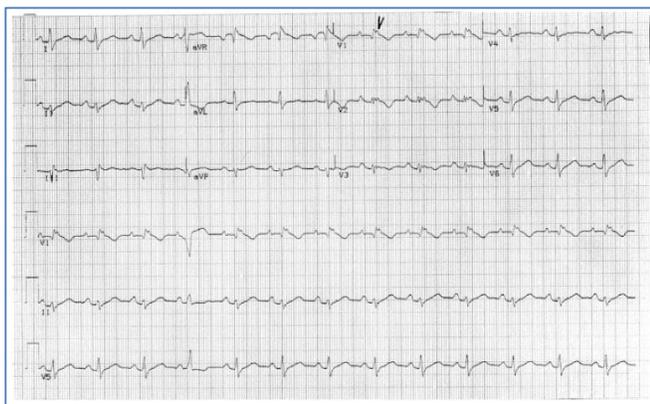
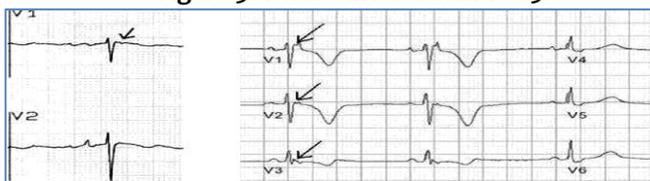


Figure 3: T wave inversion in V1-V3.



Echocardiography may detect regional or global dysfunction, predominantly affecting the RV. RV wall thinning and dilation with tricuspid regurgitation are the features of advanced right heart failure.

Cardiac magnetic resonance imaging (MRI) is a non-specific diagnostic modality, however allows reliable quantification of the right ventricle volume however may fail to detect the ventricular adipose tissue and wall thinning accurately.

Right ventricular angiogram is considered a reliable invasive method for evaluating regional right

ventricular wall motional abnormality. Cardiac MRI and 3D Echocardiography are non-invasive and preferred over right ventricular angiography for assessing morphology and functional abnormalities of the right ventricle [14].

Ventricular biopsy may confirm the suspected diagnosis of ARVD however negative biopsy should be interpreted with caution.

Immunohistochemical analysis: Reference diagnostic test highly sensitive and specific to ARVD would provide accurate early diagnosis in asymptomatic cases. Immunohistochemical analysis of plakoglobin in the intercalate discs found reduced signal level in patients with ARVD and may be of value in diagnosing ARVD [1-3]

Treatment

Current data suggest that asymptomatic patients with established ARVD do not benefit from prophylactic therapeutic interventions.

Once the clinical diagnosis of ARVD is established, treatment modalities target three main goals.

Life-style modification: Restriction from strenuous exercise and competitive sports is thought to be slow down myocardial cell death.

Prevention or effective termination of recurrent arrhythmias: Ventricular arrhythmias manifest in a wide spectrum. Sporadic premature ventricular extra systole may progress to non-sustained or sustained ventricular tachycardia and VF. VF leads to sudden cardiac arrest. Long term arrhythmia control may be achieved with implantable cardioverter defibrillator (ICD) alone or in combination with antiarrhythmic drugs.

The effect of β -blockers or Amiodarone in treatment of sustained VT has been proven. Sotalol provides the most effective arrhythmia control. Amiodarone is usually preserved for advance heart failure. In combination with B-blocker its efficacy is comparable to Sotalol

Catheter ablation is reserved for patients with recurrent ventricular arrhythmia not responsive to pharmacological treatment as it only offers short-term success, long-term efficacy remained debatable.

Cardiac failure: Symptomatic heart failure is rare and occurs predominantly in advanced stage. Cardiac failure should be treated in conjunction with conventional heart failure guidelines, most commonly with angiotensin-converting enzyme inhibitors, diuretics, digoxin and anticoagulants. In the rare occasions, when heart failure is

unresponsive to medical management, heart transplantation may be necessary [3-7].

Anaesthetic considerations

SDC during general anaesthesia is unresponsive to cardiopulmonary resuscitation. Disease awareness, understanding pathophysiology and hazardous conditions help improve safety in these patients. Perioperative management is divided in two groups: For patients with already diagnosed ARVD, and for patients who present with malignant arrhythmias and SDC.

Preoperative management

Preoperative evaluation of patients with ARVD should include collecting medical history, documenting physical findings and obtaining specialized diagnostic tests. All elective patients must be consulted with a cardiologist with experience with the disease. History of provoked arrhythmias and safe medication is essential [1-4]

Previous uneventful general or regional anaesthesia does not exclude ARVD. ECG is not used for routine assessment in young and healthy patients [1-3]

Patient with confirmed diagnosis of ARVD should undergo regular follow up to evaluate efficacy of arrhythmia control and cardiac function. Review of case notes and discussion with a cardiologist would enable the anaesthetist to gain information about the course of the disease. Physical examination carries limited significance in early stages, signs and symptoms of systemic or pulmonary congestion become apparent in advanced stage. Twenty-four hour holter-monitor is a reliable non-invasive method to discover sustained and non-sustained ventricular tachycardia. Echocardiography is available in most institutions to offer a quick insight to cardiac function. Preoperative optimisation carries high impact on perioperative mortality. Multidisciplinary team approach including cardiologist, anaesthetist and operating surgeon facilitate safe perioperative planning. Elective procedure should be postponed until ventricular arrhythmia controlled and heart failure is optimised. Regular anti-arrhythmic drugs should be continued until surgery. Patient with ICD should undergo device check to assess optimal function and number of appropriate ICD discharges. Electrical safety may warrant inactivation of the device in the immediate preoperative period [3-9].

Intraoperative management

Maintaining haemodynamic state near to the normal physiological conditions is likely to offer best

arrhythmia protection. Close haemodynamic monitoring is essential for early recognition of physiological instability and assessment cardiovascular response to intervention. Routine monitoring with ECG, oxygen saturation probe, non-invasive blood pressure cuff and capnography may be supplemented with invasive arterial blood pressure cannula even in case of minor surgical procedures. Invasive arterial pressure monitoring will allow for blood pressure monitoring even in the face of arrhythmia, when NIBP could be unreliable. Major interventions warrant more invasive monitoring. Central venous pressure provides information about right heart function and filling status. The role of pulmonary artery catheters may be controversial outside specialised cardiothoracic institutions due to induction of malignant ventricular arrhythmia and ventricular perforation during placement. However, intraoperative transoesophageal echocardiography offers superior diagnostic and avoids risks of arrhythmia. Less invasive forms of continuous cardiac output monitoring allow assessment of contractility, ventricular filling.

Malignant ventricular arrhythmia and SCD could occur at induction of general anaesthesia, during the surgical procedure or in the immediate postoperative stage. Therefore, appropriate post-operative monitoring and care locations must be chosen according to institutional organisation [1-4]

Safe anaesthetic management is facilitated by the following two principles

Understanding the pharmacokinetics and adverse effects of the anaesthetic agents: Hemodynamic variation during anaesthesia is related to specific effects of the anaesthetic agents on the sympathetic nervous system. Patient with extensive sympathetic blockade are prone to develop reduced systemic vascular resistance, reduced cardiac output and severe arterial hypotension specially in central neuraxial blockade [3,5,6]

Midazolam causes little direct myocardial depression and was reported to be safe for sedation in conjunction with opioid analgesics or for induction of general anaesthesia in combination with a different induction agent. Propofol is known to induce significant arterial hypotension and myocardial suppression. Judicious dosing or slow infusion for induction may help to overcome these unfavourable pharmacological properties making it one of the commonly used induction agent. Opioid analgesia should be selected carefully.

Fentanyl is considered to be safe. High dose of Fentanyl was used in a cardiothoracic patient population. Volatile anaesthetic agents should be administered cautiously to patients with ARVD as these are known to induce dose-dependent myocardial depression, tachycardia and arterial hypertension. Isoflurane was associated with intraoperative SCD but paradoxically, uneventful anaesthesia was also described.

Muscle relaxants have several unfavourable side effects on the circulation. Careful selection helps to avoid haemodynamic instability. Cisatracurium has minimal effect on the cardiovascular system and was associated with positive patient outcome. Anticipating and promptly responding physiological disturbance. These are the principles of balanced anaesthesia for all operations. However, there are no more important in any other group of patients other than ARVD.

Arterial perfusion pressure should be maintained. Hypertension may be treated with deepening the plane of anaesthesia. Intravenous Nicardipine has been described as a safe choice. Hypotension could diminish coronary perfusion and oxygen delivery. Although direct vasoactive drugs can provoke arrhythmias, these may need to be used to maintain haemodynamic parameters. Tachycardia increases the incidence of premature ventricular complexes and ventricular extra systole predisposing to malignant arrhythmias. During general anaesthesia tachycardia may be explained by for several factors. Pain control, adequate depth of anaesthesia and muscle relaxation should be maintained at all time. Agents with positive dromotropic and batmotropic effects have to be avoided where possible. Controlling electrolyte levels of potassium and magnesium can offer some protection. Persistent tachycardia may be treated with short acting intravenous β -blocker (Esmolol). When premature ventricular extrasystole increases in number, Amiodarone infusion should be commenced [3,6,8,9]

Preservation of Volume status, normoxia & normocapnia & normothermia is key of successful management.

Postoperative care

Maintaining physiological parameters near to the normal values decreases the incidence of malignant arrhythmias and SDC.

Adequate arterial perfusion pressure is essential to maintain coronary perfusion pressure, myocardial oxygen supply and end-organ perfusion. Perioperative

clinicians should have low threshold to treat bradycardia and tachycardia. Close monitoring of fluid balance and early recognition of significant aim to maintain haemodynamic stability.

Supplemental oxygen may help to avoid arrhythmias induced by hypoxia, however, importance of adequate carbon-dioxide removal must not be overlooked.

Adequate pain control reduces anxiety, maintains autonomic stability and prevents further catecholamine surge. Multimodal approach with non-opioid analgesic drugs may be appropriate choice after minor surgical interventions. Opioids analgesics have significant side effects on the cardiovascular and respiratory systems and their use is reserved to treat severe postoperative pain after. Regional and neuroaxial blockade may be useful after major surgical interventions.

Satisfactory treatment of postoperative nausea and vomiting eliminates vagotonic effect. Electrolyte balance maintains electrical stability of the diseased myocardium [6,11].

Case report

A 45 yrs old male patient weighing ,50 kgs was presented in PAC OPD with fracture patella to be posted for surgery .Preanaesthetic evaluation revealed h/o fall 10 days back and # right patella with no h/o head injury and no h/o loss of consciousness..Patient gave h/o chest pain 3months back for which patient was admitted for 5days and referred to SSH.

Patient was diagnosed as a case of pre-excitation syndrome with IHD with unstable angina 3months back and was started on tab. Amiodarone 200mg OD.Ptient also gave h/o syncopal attacks intermittently.

On preoperative work up his Blood investigations-were Hb-13.5, WBC-6000, Plt-1.76, RBS-103, Bld grp-O+, Na- 134, K- 3.9, Creat-0.9, T.bil- 0.1..ECG showed s/o RBBB with T wave inversion (V2-V3). 2D echo showing LVEF-60%, RA/RV dilated, RVOT dilated. Fibrofatty degeneration of RV. E/o mod TR, s/o ARVD. Cardiology opinion was taken Cardiac MRI was advised. MRI showed thinning of Rt. Ventricular free wall with fibrosis with mild dilatation of RVOT s/o ARVD. Patient Review with cardiologist opined that patient is very high risk case for surgical intervention and confirmed the diagnosis of ARVD. Patient was accepted under ASA IV explaining risk encountered during perioperative period

Anaesthetics Management of Patient

Patients with ARVD difficult to anaesthetised .Common Concerns for anaesthesia management are Minimum hemodynamic alterations, Adequate analgesia, Adequate muscle relaxation, Judicious fluid administration. To avoid proarrhythmic condition so we chose combined sciatic femoral block as sole anaesthetic technique [5].

Preoperative Period

On the day of surgery patient was accepted on high risk consent and PAC of the patient was noted & checked Monitors were attached NIBP, ECG and Pulse oximeter. Invasive arterial line was established. His PR-was 64/min, good volume, BP-128/68mmHg, SpO₂-99% on room air. Systemic examination-WNL. Plan of Anaesthesia was Combined Sciatic-femoral Nerve Block using most cardiostable local Anaesthetic Levobupivacaine using Fentanyl as Adjuvant. PNS guided lower limb blocks were administered after proper counselling and positioning was given

Sciatic nerve block: After proper positioning under all aseptic precautions PNS guided right sciatic nerve block was given with inj. Levobupivacaine 0.5% 15cc diluted to 20cc + inj. Fentanyl 50mcg.

Femoral nerve block: After proper positioning under all aseptic precautions, PNS guided right femoral nerve block was given with inj. Levobupivacaine 0.5% 10cc + inj. Fentanyl 50mcg., patient was handed over to surgery after confirming complete sensory and motor blockade. Throughout the intraoperative period patient was hemodynamically stable Overloading the patient with IV fluids was avoided. Judicious fluid administration was opted.500ml of crystalloids was administered in the intraoperative period with 100ml IV paracetamol towards the end of the surgery.

Postoperative period

Patient was conscious, oriented in the postoperative period. On examination: Pulse Rate was 68/min, BP-122/72mmHg, SpO₂-100%. Systemic examination: WNL Patient was shifted in Surgical Intensive Care Unit (SICU) for observation for the next 48hours. Recovery in SICU was uneventful. Patient was discharged after 8 days

Discussion

Patients with ARVD undergo variety of surgical procedures, the Physiological changes & medication in

perioperative period may have pro arrhythmogenic effect. Surgical insult activates stress response with catecholamine release while adverse anaesthetic effects induce significant cardiac instability. Early diagnosis could often control progress of disease and prevention of malignant arrhythmias and sudden cardiac arrest.

The effect of β -blockers or Amiodarone in treatment of sustained VT has been proven. Sotalol provides the most effective arrhythmia control. Amiodarone is usually preserved for advance heart failure. In combination with B-blocker its efficacy is comparable to Sotalol [3]. Our Patient was stabilised with 200mg Amiodarone OD [1-4].

In present case general anaesthesia was deliberately avoided to avoid polypharmacy, all the noxious stimuli causing catecholamine release during the process and causing arrhythmias. Pain control, adequate depth of anaesthesia and muscle relaxation should be maintained at all time.

There are cases reported of ARVD landing up in Sudden Cardiac Death on induction itself [3,8,9].

Even central Neuraxial block was overruled. Cardiovascular changes are the most important effects of central neural block. Keen watch on non-specific symptoms such as nausea, vomiting and syncope may be helpful to avoid cardiovascular collapse [3].

Epidural anaesthesia provides better controlled onset of the blockade with less marked effect on cardiovascular system. It also uses large doses of local anaesthetics, which can cause arrhythmias after their systemic absorption [3]. Anaesthesia protocol should aim at reducing adrenergic stimulation.

This was the major reason we opted for peripheral nerve block over neuraxial techniques and GA [3,5]. Sciatic nerve (L4,5,S1,2,3) is the largest nerve of lower extremity. Supplementation of sciatic with femoral block (post. Division of L2,3,4) provides anaesthesia for surgeries of thigh and knee. This avoids deleterious hemodynamic effects of neuraxial blocks and GA.

Avoidance of epinephrine as an adjunct to LA agent and large doses of bupivacaine is advocated in many studies to reduce risks of potential cardio toxicity. So we used dosages free of epinephrine and inj. Levobupivacaine (relatively cardiac stable than inj. Bupivacaine) for the nerve blocks.

Maintaining haemodynamic state near to the normal physiological conditions is likely to offer best

arrhythmia protection. PNS guided administration increases accuracy & high success rate Avoidance of epinephrine as an adjunct to LA agent and large doses of bupivacaine is advocated in many studies to reduce risks of potential cardio toxicity. So we used dosages free of epinephrine and inj. Levobupivacaine (relatively cardiac stable than inj. Bupivacaine) for the nerve blocks.

Maintaining haemodynamic state near to the normal physiological conditions is likely to offer best arrhythmia protection. We Maintained Normothermia. Hypothermia increases the risk of shivering and increased oxygen consumption and predispose to tachyarrhythmia. Volume status was strictly maintained. Excessive intravenous fluid administration is hazardous in case of ventricular dysfunction as it can distend the heart & increase overload [3,5,8,9].

Postoperatively Supplemental oxygen may help to avoid arrhythmias induced by hypoxia, however, importance of adequate carbon-dioxide removal must not be overlooked. Pain control by Multimodal approach with non-opioid analgesic drugs may be appropriate choice after minor surgical interventions.

Adequate pain control reduces anxiety, maintains autonomic stability and prevents further catecholamine surge. Malignant ventricular arrhythmia and SCD could occur at induction of general anaesthesia, during the surgical procedure or in the immediate postoperative stage. Appropriate post-operative monitoring and care locations must be chosen according to institutional organisation

Conclusion

Careful perioperative assessment and solid management plan focusing on the maintenance of physiological stability may help to avoid perioperative loss and provide the highest level of safety in this patient population. Chest pain in young adult male needs evaluation. Diagnosed cases of ARVD needs multidisciplinary approach Regular follow up of diagnosed cases of ARVD can minimise intra and postoperative complications. .Peripheral nerve blocks i.e Combined sciatic Femoral Nerve block proven to be a boon for patients with ARVD.

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