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Automatic Implantable Cardioverter Defibrillator Management in A Patient with Arrythmogenic Right Ventricular Dysplasia having Prone Thoracic Spine Surgery

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Abstract: Arrhythmogenic right ventricular dysplasia is an inherited condition causing right ventricular structural and functional changes that can manifest by ventricular arrhythmia, heart failure, and sudden death. Therapy is not well defined but avoidance of sudden death is a major goal in patients with the disease. Many patients at high risk for ventricular arrhythmia have undergone automatic implantable cardioverter defibrillator (ICD) placement. This case report discusses the intraoperative management of an ICD in a 50-year-old female with a history of arrhythmogenic right ventricular dysplasia who underwent posterior spinal arthrodesis of T7-L1.

Keywords: Implantable cardioverter defibrillator, arrythmogenic right ventricular dysplasia, intraoperative management, Prone spine surgery.

INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD) is an inherited condition affecting 1:1000 to 1:10000 [1,2,5]. It is manifested by ventricular arrhythmia, heart failure and sudden cardiac death. Right ventricular structural and functional changes including fibrous and fatty infiltration of the myocardium are seen on histopathology [3]. Therapy for ARVD is directed at preventing sudden death from ventricular arrhythmia. Several cases of perioperative death have been reported in patients with unrecognized ARVD [2,4]. We describe the management of a patient with ARVD prompting an automatic implantable cardioverter defibrillator (ICD) device. The patient underwent posterior spinal arthrodesis of T7-L1.

CASE REPORT

A 50-year-old female with a recently implanted ICD for ARVD presented for posterior instrumented spinal arthrodesis. She had suffered a burst fracture of T10 with resulting paraplegia following a syncopal episode. T7-L1 fusion with right posterior iliac crest bone graft was performed. She had a recent history of syncope and was diagnosed with ARVD 3 months prior to surgery. An MRI scan showed intramyocardial fatty infiltration. At the time of diagnosis of ARVD an ICD (St Jude V-168 Atlas II VR) was placed, and interrogation at 3 months from implantation showed some heart rates of 140-160, but she had not received any shock therapy for ventricular tachycardia.

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In addition to ARVD, the patient had medical history significant for left upper/ lower extremity numbness with electromyographic evidence of chronic C5-C7, and L5-S1 radiculopathy, polycystic ovarian syndrome, hypertension, asthma, depression, Barrett's esophagus, and migraine headache. Prior surgeries included a Nissen fundoplication, cholecystecctomy, and benign ovarian tumor removal. The patient's home medications were hydrochlorothiazide 25 mg daily, fluticasone/salmeterol 250/50 mg twice daily, alprazolam 50 mg twice daily, albuterol inhaler as needed, trazadone as needed for insomnia, paroxetine 20 mg daily, and aspirin 81 mg daily. At the time of presentation for surgery she was on intravenous hydrocortisone and phenylephrine infusions that were initiated in the neurosurgical ICU.

On exam, the patient was alert and oriented. Blood pressure was 109/54 mm Hg, heart rate was 96/min, and SpO2 was 100% on nasal oxygen at 2 L/min.

Intraoperative monitoring of somatosensory and motor evoked potentials of upper and lower extremities was performed. In order to avoid interference with this intraoperative monitoring, a total intravenous anesthetic was used with propofol and remifentanil infusions. It is common practice to disable anti-tachyarrythmic functions of ICD devices when electrocautery is planned. In our patient the ICD was left enabled during the procedure since the patient was positioned prone and it would be difficult to perform resuscitation for any hemodynamically significant arrhythmia. It would also be difficult to reactivate the device because of patient position. A magnet and an external defibrillator were immediately available throughout the case. The surgeons were made aware that the ICD would be left enabled during the procedure.

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Prior to induction the patient was connected to standard monitors including a blood pressure cuff, ECG, and pulse oximetry. In addition, the patient had a radial arterial line. She had femoral central access as well as peripheral venous access. Electrodes needed for somatosensory and motor evoked potentials were placed by a technician prior to induction of general anesthesia. A fiberoptic intubation was performed with the aid of remifentanil and propofol infusion. Remifentanil 0.15 mcg/kg/min and propofol 100-120 mcg/kg/min were infused as anesthesia maintenance. Mean arterial pressure was maintained at 65-75 mm Hg with phenlyephrine infusion. Methylprednisolone infusion of 486 mg/h was continued per protocol. The patient was positioned prone for the surgery, and care was taken that the ICD was accessible. Blood loss was replaced with 500 ml of hetastarch, 500 ml of normal saline, 4 units of packed red blood cells and 288 ml of salvaged cells. Adequate urine output occurred throughout the procedure. Continuous monitoring of ECG showed normal sinus rhythm throughout procedure without arrhythmia. Due to the significant volume shifts and being in the prone position for 7 h during surgery, the patient was left intubated for transport to the intensive care unit. The patient was later extubated without event. No inappropriate shock therapy occurred during the case. Interrogation of the ICD after the surgery did not reveal any adverse consequence to other device settings.

DISCUSSION

ARVD is an inherited condition associated with sudden cardiac death as a result of ventricular arrhythmia. Myocardial structure is replaced by fibrofatty tissue as a result of apoptosis, inflammatory disease (myocarditis), and myocardial dystrophy [5]. Ventricular arrhythmias are commonly brought on by exercise-induced stress in patients with ARVD [6]. A particular concern for anesthesiologists is the possibility of arrhythmias caused by surgical stress and anesthesia. Ventricular tachycardia has been described in relation to ARVD in the perioperative period after general anesthesia [7]. Case reports of unrecognized ARVD and perioperative death after general anesthesia have been reported. All three patients from these case reports had autopsies and examination of the hearts revealed typical features of ARVD [2,4]. Autopsies after unexplained perioperative deaths in France showed that 18 out of 47 patients with cardiac pathology had typical histologic features of patients with ARVD [8].

Treatment of ARVD is not well defined, but avoidance of sudden cardiac death is the major goal of therapy. Avoidance of competitive sports or any stressful activity that causes symptoms such as palpitations or syncope is recommended [9]. ICD implantation is common in those patients who have experienced sustained ventricular arrhythmia or those deemed at high risk of sudden cardiac death [10]. Radiofrequency ablation can be performed to treat arrhythmogenic foci associated with ARVD, but that will not eliminate the risk of sudden death due to the progression of disease in other areas [11]. Right ventricular exclusion surgery, in which the right heart is electrically disconnected from the left heart, has also been used for management of severe ARVD [12]. Antiarrhythmic drugs such as sotalol have been shown to reduce and prevent ventricular arrhythmias in patients with ARVD [13]. Heart transplantation is an option for patients with severe heart failure.

It is common practice to disable the antitachyarrhythmia functions of an ICD when a patient is undergoing surgery, especially when electrocautery is used. It is thought that interference caused by electrocautery could lead to inappropriate shocks or to alterations in device programming. Ventricular tachycardia and fibrillation have been reported during replacement of ICD devices [14]. In one report, three patients underwent a total of five surgical procedures in which electrocautery was used, and no sensing of electrocautery signals or other device malfunction occurred [15]. The ICD was left enabled in our case for several reasons. First, our patient had recent diagnosis of ARVD with multiple episodes of syncope that were likely due to hemodynamically significant arrhythmias. The patient was in the prone position, and resuscitation would have been difficult if a hemodynamically significant arrhythmia occurred. We chose not to have the device programmed off because it would have been more difficult to reactivate the device if necessary with the patient in the prone position. Proper placement of cutaneous defibrillation pads would have been difficult due to the prone position. It was also uncertain whether defibrillation pads would deliver an effective shock with the patient's body habitus. Should external defibrillation have been required, it may have adversely affected the ICD. Therefore, it was presumed that treatment of any intraoperative arrhythmia by the ICD would be safer than external defibrillation. Acute management of ventricular tachycardia with antiarrhythmic drugs in patients with ARVD has varying efficacy [13]. An external defibrillator and several antiarrhythmic medications were kept immediately available in case a life threatening arrhythmia occurred which was not managed appropriately by the ICD. Precautions were taken so that the electrocautery current return pad was placed on a lower extremity as far away from the device as possible. The surgeons used electrocautery in short intervals, which may have less effect on ICD devices.

Because of the possibilities of pacing suppression, inadvertent re-programming, and/or inappropriate delivery of shocks, pacemakers and cardioverter/defibrillators can be deliberated de-activated with a simple magnet or with a specific re-programming device.

Most ICDs (Medtronic, St. Jude, Biotronic) are inhibited from firing upon application of a simple magnet. Removal of the magnet permits defibrillation by the device. However, some Guidant ICDs do not so respond to magnets and require reprogramming for disabling of the defibrillator. Pacemaker suppression by electrocautery is a problem if the patient depends on the pacer to maintain perfusion. Simple magnets will keep pacers in the asynchronous mode, thus preventing suppression by cautery but carrying a risk of an R-on-T phenomenon. The Medtronic AT500 is a rare example of a pacemaker that cannot be so adjusted with a simple magnet.

Despite no apparent adverse effect of electrocautery in our case, it is probably best to suspend antitachyarrhythmic functions of an ICD in most cases. Follow up interrogation of such devices after electrocautery exposure should be used

Table 1. Summery of Considerations for ICD Management During Surgery

1. Though devices are increasingly protected from electromechanical interference, it is prudent to minimize exposure of the device.
2. Application of a magnet is rarely required with devices less than 4 years old. Because of malpositioning of the magnet or because of programming, a magnet does not prevent all ICDs from delivering shocks.
3. The skin electrode of a unipolar system should be placed so as to minimize current through the ICD system; direct contact of the ICD with an electrocautery electrode is likely to be damaging.
4. Electrocautery power should be at the lowest effective level. Short bursts are preferable to prolonged application of cautery.
5. Bipolar electrocautery is preferable.
6. The harmonic (ultrasonic) scalpel is virtually free of interference.
7. Continuous monitoring of the pulse (as by arterial cannulation, palpation, pulse oximetry, auscultation) is prudent.
8. Magnetic resonance imaging is contra-indicated.
9. An external defibrillator and pacer should be available. Adhesive skin pads are useful for defibrillation and for ventricular pacing. The possibility of transesophageal atrial pacing should be considered.

to ensure that device programming is intact. With increased indications for ICD implantation, anesthesiologists will have to manage these devices more frequently. Management of such devices should be tailored to the patient's specific device as well as the clinical situation, see Table **1**.

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