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Electrical storm in an adolescent with arrhythmogenic right ventricle cardiomyopathy treated with cardiac transplantation

To the Editor,

Arrhythmogenic right ventricle dysplasia/cardiomyopathy (ARVD/C) is an inherited cardiomyopathy characterized by fibro fatty replacement of the right and less frequently left ventricle (1, 2). Ventricular arrhythmias requiring implantable cardioverter defibrillator (ICD) are common in patients with ARVD/C and electrical storm (≥3 life-threatening ventricular arrhythmia within 24-hour period) resulting in ICD discharges is a major cause of morbidity and mortality (3). Radiofrequency ablation with three-dimensional (3-D) mapping and navigation systems has been recently advocated as a preferred treatment for recurrent ventricular arrhythmias (4). We had experience of a case of a15-year-old boy who presented with chest pain and decreased exercise capacity. He had undergone surgical closure for atrial septal defect and complicated with ventricular tachycardia/fibrillation postoperatively at 11 years-old. No history of congenital heart defect in family and syncope were found. Premature ventricular contractions were determined occasionally in 24-hour Holter monitoring. Echocardiography revealed a dilatation of the right ventricle and the right ventricle outflow tract. The dilated infundibulum and increased trabeculation in the right and left ventricle were shown in the cardiac angiography and magnetic resonance imaging. Non-sustained monomorphic ventricular tachycardia (VT) with the rate of 260 beats/minute was induced by programmed stimulation with a single extra-stimulus from the right ventricle. He was diagnosed as ARVD/C and ICD was implanted for primary prevention. Two years later, the episodes of ventricular tachycardia/fibrillation were repeated 35 times within one month. Amiodarone and sotalol administration was initiated and the ablation treatment was planned. A single 4 mm open-irrigation ablation catheter (Medtronic, MN, USA) was advanced to right ventricle via femoral vein by using the EnSite NavX 3D mapping and navigation system (St Jude Medical, MN, USA). The area with <0.5 mV during voltage mapping was considered as scar tissue. Radiofrequency ablation was applied to around the scar at the temperature 45°C with 30-35 Watt energy. Total procedure time was 280 minutes. Two months later, the electrical storm repeated again and the patient was arrested in a short time. He was immediately connected to the pump after cardiac resuscitation and underwent cardiac transplantation from an adult cadaver one day later. He has been on follow-up with no symptom for three years.

Stec et al. (4) reported a pregnant woman with an electrical storm due to ARVD/C of successful endocardial catheter ablation, by using 3-D mapping and navigation system. Although ventricular tachycardia frequency is reduced after catheter ablation, ventricular arrhythmia recurrence is still common in ARVD/C (2). It appears that ICD is currently an indispensable treatment option in ARVD/C.

Philips et al. (2) claimed that VT-free period after epicardial ablation was longer than those after endocardial ablation. They speculated that it was because of epicardial distribution of ARVD/C. In our case, recurrence of VT may be associated with endocardial ablation. Nevertheless, catheter ablation of ventricular tachycardia in ARVD/C can be considered as a beneficial method in terms of reducing the side effects of antiarrhythmic drugs and prolonging the life of ICD battery (2). The management of an electrical storm should be individualized for each patient and the treatment may indicate extracorporal membrane oxygenation and cardiac transplantation.

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Long term follow-up in a patient with acute type A aortic dissection complicated with cardiac tamponade without surgery

To the Editor,

Acute type A aortic dissection complicated by cardiac tamponade is a rare disease but frequently associated with poor outcomes. Urgent open surgical repair is required for this patient group. Here, we discussed long term follow-up in a patient who developed acute type A aortic dissection complicated by cardiac tamponade and did not undergo surgery.