The Right Ventricular Outflow Tract as an Unusual Location for an Implantable Defibrillator Electrode in a Patient With Arrhythmogenic Right Ventricular Dysplasia

Kiyotaka Matsuo, MD; Takashi Kurita, MD; Masamichi Eguchi, MD; Kojiro Nakao, MD; Norihiro Komiyama, MD; Hiroaki Kawano, MD; Shojiro Isomoto, MD; Genji Toda, MD; Motonobu Hayano, MD; Katsusuke Yano, MD

A 41-year-old woman with arrhythmogenic right ventricular dysplasia (ARVD) underwent the implantation of an implantable cardioverter-defibrillator (ICD), in which the defibrillator electrode was unusually located in the right ventricular (RV) outflow tract. Although fractionated electrograms were demonstrated in the RV apex, which is the usual site for ICD electrodes, normal electrograms were recorded in the RV outflow tract during an electrophysiologic study. An electrode with a screw-in tip was used to fix the implant in the RV outflow tract and obtain successful defibrillation. If normal electrograms are recorded in the RV outflow tract, the site may prove to be an alternative location for an ICD electrode even for ARVD patients. (Jpn Circ J 2001; 65: 994–996)

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Arrhythmogenic right ventricular dysplasia (ARVD) is characterized by local or diffuse wall motion abnormalities that exclusively or predominantly affect the right ventricle (RV), and is associated with recurrent ventricular arrhythmias originating from the RV.1,2 Implantable cardioverter-defibrillator (ICD) is indicated in patients with lethal arrhythmias to prevent sudden death.1 Because the RV apex, which is the usual site for an ICD electrode, is sometimes affected in ARVD, a defibrillator electrode located in the RV apex may be difficult to detect low amplitude electrograms.3 A patient with ARVD who had successful defibrillation in the RV outflow tract, which is an unusual site for an ICD electrode, is described.

Case Report

A 41-year-old woman, who had had an episode of successful resuscitation from ventricular fibrillation in November 1998, experienced a syncopal attack and was admitted to hospital in January 2000. Her father had also died suddenly at 58 years of age. A 12-lead electrocardiogram (Fig 1) demonstrated abnormal Q waves in leads II, III and aVF, and inverted T waves in leads II, III, aVF and V1–3. Epsilon waves (ie, post-excitation waves) were suspected in leads V2–4, which were confirmed by a signal averaging test. Computed tomography disclosed RV enlargement and a thrombus (2.0 cm×2.0 cm) in its apex (Fig 2), which disappeared after 4 weeks of treatment with warfarin. Left ventriculography showed hypokinesia of the inferior wall and a decrease of 39% in ejection fraction. Right ventriculography revealed an enlarged cavity and hypokinesia in the infero-apical wall. Coronary angiography found neither organic stenosis nor segmental spasm, with ST-segment elevation induced by the intracoronary injection of acetylcholine. Right ventricular endomyocardial biopsy specimen obtained from the septal site of the infero-apical wall indicated interstitial fibrosis. Accordingly, the patient was diagnosed as having ARVD with left ventricular involvement.

Fig 1. Twelve-lead electrocardiogram demonstrating abnormal Q waves in leads II, III and aVF, and inverted T waves in leads II, III, aVF and V1–3. Small epsilon waves (arrowheads) can be seen in leads V2–4, which were confirmed by a signal averaging test.
An electrophysiologic study demonstrated normal electrograms in the left ventricle during sinus rhythm. Conversely, fractionated electrograms, defined as those having a duration >100 ms and multiple rapid low amplitude (<1.0 mV) deflections, were recorded in the RV, which were located widely in the inferior wall and throughout the floor of the inflow tract to the apex. However, the other RV sites, including the outflow tract, were free from such abnormal electrograms and showed normal electrograms with spiky deflections (Fig 3). Ventricular tachycardia originating from multiple sites and ventricular fibrillation were induced easily by programmed pacing. Because sustained ventricular tachycardia recurred after 4 weeks of treatment with amiodarone, the patient was advised to receive an ICD in April 2000.

The patient was given amiodarone at the time of the ICD implantation. Under general anesthesia, a non-thoracotomy lead system was used. A 65-cm single-coil defibrillation lead with a screw-in tip (Medtronic 6943 SPRINT™; Medtronic Inc, Minneapolis, MN, USA) was inserted via the left subclavian vein and initially advanced to the RV apex. There was no R wave sensing at a value higher than 1.0 mV in the RV apex probably due to extensive endocardial fibrofatty replacement. Finally, we found an adequate site for sensing and pacing in the RV outflow tract, which had been detected earlier during the electrophysiologic study. R wave sensing was 9.6 mV, pacing threshold was 0.8 V at 0.4 ms, and pacing lead impedance was 500 Ω. The lead was finally fixed in the location using a screw-in tip against the high interventricular septum (Fig 4). Consequently, a coil-defibrillation electrode, measuring 5 cm in length, lay from the outflow to the inflow tract in the RV cavity. Ventricular fibrillation was induced through the electrode by shock on a T wave and we found the defibrillation threshold to be 15 J. A biphase ICD pulse generator (Medtronic 7223CX MICRO JEWEL™ II; Medtronic Inc) was implanted in the patient’s left infraclavicular pocket. The echocardiogram performed on the following day showed no signs of pericardial effusion but mild tricuspid regurgitation was evident. An ICD test 1 week after implantation indicated that ventricular fibrillation was sensed adequately and defibrillated successfully by the system. The position of the ICD electrode remained stable and unchanged on follow-up chest X-ray 8 months after her discharge.

**Discussion**

Results of the present study indicate that the RV outflow tract is a good alternative site for an ICD electrode and that mapping of RV endocardial electrograms during sinus rhythm in electrophysiologic studies is useful for finding a suitable location in an ARVD patient.

The RV outflow tract is an unusual location for an ICD electrode. Previous reports showed that the RV outflow tract is a good alternative location for defibrillator electrodes in patients with coronary artery disease or dilated or hypertrophic cardiomyopathy because the RV apex is inadequate for sensing and pacing as a result of extensive endocardial scarring. To our knowledge, however, there are few reports in which the RV outflow tract is used as an alternative location for patients with ARVD. Although the mechanism of defibrillation is unknown, passing defibrillating energy through the interventricular septum is important for successful defibrillation. Also, successful defibrillation occurs when a critical amount of myocardium becomes depolarized by an electrical discharge in the patient.
patient of the present study, the single coil-electrode lying from the RV outflow to the inflow tract in combination with the tip electrode fixed in the RV outflow tract against the high interventricular septum achieved successful defibrillation, suggesting that the location of the defibrillation electrode was sufficient for achieving a shock field across the heart. Nevertheless, it was necessary to take a follow-up ICD test after the patient’s discharge because the defibrillation threshold may increase, which may be associated with either the unusual site chosen for the defibrillation electrode or because ARVD was the underlying heart disease. The RV outflow tract proved to be an alternative site for a defibrillator electrode in the patient of the present study, whereas mild tricuspid regurgitation, which was documented in echocardiography after the ICD implantation, might be due to the single coil-electrode lying from the RV outflow to the inflow tract.

Marcus et al proposed that the most frequent sites of dysplasia obtained either from surgical treatment of ventricular tachycardia or from an autopsy are the RV outflow tract, the apex, and the inferior wall of the inflow tract, which is known as the ‘triangle of dysplasia’1. However, these 3 sites are not always affected in a patient with ARVD because it has also been reported that the most affected site is the inferior wall of the RV inflow tract and only one-third of patients have fractionated electrograms at the RV outflow tract during an electrophysiologic study12. We also found fractionated electrograms in the RV inferior wall throughout the floor of the inflow tract to the apex. The RV outflow tract was free from abnormal electrograms, which was expected considering the alternative location of the defibrillator electrode in the patient. Under these conditions, we used a defibrillation electrode with a screw-in tip in order to secure it in the RV outflow tract, resulting in stability and successful defibrillation. Therefore, evaluating electrograms in the entire RV before ICD implantation proved to be useful in searching for an adequate location for the ICD electrode.

Although the patient of the present study was diagnosed as having ARVD with left ventricular involvement, the RV endomyocardial biopsy specimen obtained from the septal site of the RV infero-apical wall did not indicate fatty replacement. A diagnosis based on RV endomyocardial biopsy is inherently difficult because the segmental nature of the disease causes a false negative and because the interventricular septum is rarely involved2.

In summary, the RV outflow tract may prove to be an alternative location for implanting an internal defibrillator electrode in ARVD patients with abnormal fractionated electrograms in the RV apex and without those in the RV outflow tract.

References