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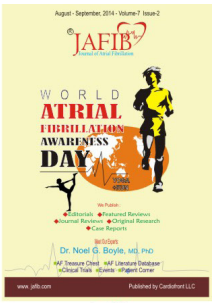

ICD Therapy In RVOT-VT And Early Stage ARVD/C Patients

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Abstract

Implantable cardioverter-defibrillators (ICDs) improve the survival of patients with ischemic or non-ischemic cardiomyopathy and a reduced ejection fraction. However, the efficacy of ICD therapy in patients with right ventricular outflow tract ventricular tachycardia (RVOT-VT) and early stage arrhythmogenic right ventricular dysplasia / cardiomyopathy (ARVD/C) has not been well clarified. Although the prognosis of RVOT-VT is generally good, malignant forms of RVOT-VT resulting in polymorphic VT have been reported by several investigators. Radiofrequency catheter ablation is still effective in such patients, and thus an ICD implantation is usually not required. On the other hand, according to the current guidelines in patients with ARVD/C, an ICD implantation is recommended for secondary prevention when the patients develop sustained VT or VF. An ICD implantation may also be considered for primary prevention in high-risk patients: extensive disease, family history of sudden cardiac death, or undiagnosed syncope. Since an ICD implantation in the early stage of ARVD/C is controversial, physicians should well consider its risks and benefits. Early intervention with ICD therapy in ARVD/C patients may reduce the arrhythmic death rate but increases the device related complications especially in younger patients.

Introduction

Implantable cardioverter-defibrillators (ICDs) have been proven to improve the survival of patients with a primary or secondary risk of sudden cardiac death.[1,2] However, the large clinical trials were mainly conducted in patients with ischemic or non-ischemic cardiomyopathy and a reduced cardiac function.[2,3]

In contrast, ventricular tachycardia (VT) originating from the right ventricular outflow tract (RVOT-VT) may occur in patients without any demonstrable structural heart disease or in patients with early stage arrhythmogenic right ventricular cardiomyopathy (ARVD/C). Since the patients demonstrate a normal cardiac function in the two situations, the indication for ICD therapy may be controversial. In this report, we reviewed the current perspectives on ICD therapy in those situations.

Right Ventricular Outflow Tract Ventricular Tachycardia (RVOT-VT)

VT occurring in the absence of structural heart disease is called idiopathic VT, and occupies approximately 10% of all VTs.[4] The most common idiopathic VT originates from the right ventricular outflow tract. RVOT-VT is diagnosed by its VT morphology; left bundle branch block and inferior axis occurring in the heart without any demonstrable disease. This VT rarely may be induced by rapid pacing during an electrophysiological study (EPS), but may easily be induced by exercise or may develop after the administration of catecholamines, usually by an infusion of isoproterenol. RVOT-VT can be well suppressed by adenosine or beta-blockers. The mechanism is considered to be related to cAMP mediated delayed after-depolarizations (DADs). It is important to exclude the existence of an early stage of ARCD/C or to diagnose its combination.

Manifestation Of RVOT-VT

Most RVOT-VT occurs as a repetitive non-sustained VT with a monomorphic morphology during exercise or emotional stress. Only rarely, RVOT-VT may sustain and result in hemodynamic deterioration. In addition, a malignant form of RVOT-VT has recently been reported by investigators. Viskin et al. reported three cases who originally presented with typical PVCs or VT of an RVOT origin, but developed polymorphic VT during the follow-up.[5] The short coupled PVCs trigger polymorphic VT, which degenerates into ventricular fibrillation (VF). Noda et al. also reported 16 cases of patients with polymorphic VT triggered by RVOT-PVCs from Japan.[6] In their cases, the coupling intervals of the polymorphic VT-triggering PVCs were not short. From the ECG findings, Kurosaki et al. reported that a positive QRS complex in lead I was reported to be a marker of malignant arrhythmias from the RVOT.[7]

Therefore, there may exist some cases with a malignant form of RVOT-VT. Their characteristics include a history of syncope, fast or polymorphic VT, and short-coupled PVCs with and without documented VF. An aggressive therapy would certainly be indicated in them. Of note, some patients develop ARVD/C during the follow-up.[8]

Key Words:

Ventricular Tachycardia, Implantable Cardioverter-Defibrillator, Arrhythmogenic Right Ventricular Cardiomyopathy.

Disclosures:
None.

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Therapeutic Strategy In Patients With RVOT-VT

Initiation Of Treatment

Treatment is initiated depending on the symptoms, heart rate and frequency of the VT episodes. Avoiding exercise or the administration of beta-blockers might be effective in most cases of RVOT-VT.

Elimination Of The PVCs

To cure RVOT-VT, radiofrequency catheter ablation (RFCA) is usually indicated since it is more effective (an approximate 90% success rate) than medical therapy. Malignant RVOT-VT may be suppressed by the elimination of the VF-triggering PVCs by RFCA.[9,10,11,12] In general, catheter ablation is recommended in patients with idiopathic VT who have multiple symptomatic VT episodes or very fast VT.

ICD Implantation For Preventing Sudden Cardiac Death

An ICD implantation may be indicated in patients who have a history of an aborted sudden cardiac death (SCD) or when RFCA fails or is contraindicated.

Arrhythmogenic Right Ventricular Dysplasia / Cardiomyopathy (ARVD/C)

ARVD/C is an inherited cardiomyopathy characterized by fibro-fatty replacement of the myocardium of the right ventricle (RV).[13] A wall motion abnormality or dilatation of the right ventricle is diagnostic. However, structural changes may be absent in the early stage of the disease. Progression of the disease involves not only the RV, but also the left ventricle (LV). Desmosomal gene mutations have been linked to ARVD/C and are inherited with an autosomal dominant pattern.[14] Recessive forms of ARVD/C, such as Naxos disease or Carvajal syndrome, have also been reported.

Clinical Manifestation Of ARVD/C

The clinical manifestation of ARVD/C is ventricular arrhythmias with an RV origin: mainly monomorphic sustained VT and rarely polymorphic VT and VF, heart failure and SCD, especially in young people. During the early stage of ARVD/C it would be difficult to differentiate an ARVD/C related VT from RVOT-VT. Rapid or polymorphic VT degenerating into VF would be accompanied by a hemodynamic collapse and could be a cause of the SCD.[15]

Antiarrhythmic drugs may decrease the incidence of VT or VF in ARVD/C patients, but they have not yet been shown to reduce the SCD in ARVD/C patients. Amiodarone or sotalol may be a drug of choice only when the patients are at risk for SCD but an ICD implantation is not feasible.[16] RFCA is challenging in patients with ARVD/C. It can be effective in eliminating or reducing the recurrence of monomorphic VT, but because of the progressive nature of this disease, the long-term efficacy of RFCA is limited and is considered as an adjunctive therapy in ARVD/C patients with recurrent VT.

Indications For An ICD In ARVD/C Patients

So far, there have been no prospective randomized trials on ICD therapy in patients with ARVD/C for primary and secondary prevention of sudden death. However, patients with ARVD/C that underwent ICD implantations were shown to have a high incidence of an appropriate ICD therapy and a low rate of arrhythmic death.[17]

The current guidelines recommend an ICD implantation for secondary prevention of SCD in patients with documented sustained VT or VF who are receiving optimal medical therapy.[16,18] RFCA is sometimes performed in patients with an ICD in order to reduce the incidence of frequent ICD shocks due to recurrent VT.

An ICD implantation may be considered for primary prevention if patients have any risk of SCD: a history of unexplained syncope, induction of VT during the EPS, non-sustained VT, male gender, extensive disease (severe dilation or extensive involvement of the RV and LV involvement), family history of SCD, and positive gene mutation related to ARVD/C. In asymptomatic family members, a less extensive RV lesion, or non-sustained VT, appears to constitute a low risk. The predictive value of the induction of VT during the EPS may be useful in predicting a future VT occurrence, and may be used in identifying patients at high risk for an overall cardiac mortality. The efficacy of ICDs for primary prevention has not established in the present guidelines.

Conclusion:

In conclusion, the ventricular tachyarrhythmias of RVOT-VT and ARVD/C look similar, and clinicians should be reminded of the need for a differential diagnosis between them. The risk for SCD in patients with RVOT-VT is generally low and RFCA is effective. Only a limited number of patients with aborted SCD would be indicated for an ICD. An ICD may prevent patients against SCD, but there are device-related complications such as infections, and inappropriate shocks especially in younger patients. There is no doubt that an ICD implantation is appropriate in patients with ARVD/C for secondary prevention, but the indications for primary prevention especially in patients during the early stage of ARVD/C may not be supported by the present guidelines.

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