Arrhythmogenic Right Ventricular Cardiomyopathy

Secondo ACC/AHA/ESC 2006 Guidelines for
Management of Patients With Ventricular
Arrhythmias and the Prevention of Sudden Cardiac Death

Recommendations

Class I

ICD implantation is recommended for the prevention of SCD in patients with ARVC with

- **documented sustained VT or VF**

who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 y. *(Level of Evidence: B)*

Class IIa

1. ICD implantation can be effective for the prevention of SCD in patients with ARVC with

- **Extensive disease, including those with LV involvement,**
- **1 or more affected family member with SCD,**
- **Undiagnosed syncope when VT or VF has not been excluded as the cause of syncope,**

who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 y. *(Level of Evidence: C)*

2. Amiodarone or sotalol can be effective for treatment of sustained VT or VF in patients with ARVC when ICD implantation is not feasible. *(Level of Evidence: C)*

3. Ablation can be useful as adjunctive therapy in management of patients with ARVC with recurrent VT, despite optimal antiarrhythmic drug therapy.
(Level of Evidence: C)

Class IIb

EP testing might be useful for risk assessment of SCD in patients with ARVC. *(Level of Evidence: C)*

	I	***IIA***
Prevenzione primaria		**1. ICD implantation can be effective for the prevention** of SCD in patients with ARVC with • **Extensive disease, including those with LV involvement,** • **1 or more affected family member with SCD,** • **Undiagnosed syncope when VT or VF has not been excluded as the cause of syncope,** who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 y. *(Level of Evidence: C)*
Prevenzione secondaraia	**ICD** implantation is recommended for the prevention of SCD in patients with ARVC with • **documented sustained VT or VF** who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 y. *(Level of Evidence: B)*	**1. Amiodarone or sotalol** can be effective for treatment of sustained **VT or VF** in patients with ARVC **when ICD implantation is not feasible.** *(Level of Evidence: C)* **2. Ablation** can be useful as adjunctive therapy in management of patients with ARVC with **recurrent VT, despite optimal antiarrhythmic drug therapy.** *(Level of Evidence: C)*

Risk Stratification

ARVC ("dysplasia") is suspected in patients, typically a young man, with RV arrhythmias or in relatives of individuals with known ARVC. Syncope, presyncope, and, less frequently, biventricular failure are also observed. The ventricular arrhythmias have LBBB morphology that spans the spectrum of simple ventricular ectopy, sustained and NSVT, or VF. ARVC needs to be considered along with idiopathic RV outflow VT in the individual with ventricular ectopy and VT coming from the RV outflow region. In contrast to ARVC, idiopathic RV outflow VT is usually not associated with the ECG abnormalities seen with ARVC, is more common in women, and is initiated by isoproterenol infusion instead of by EP testing. **The ECG in ARVC frequently shows precordial T-wave inversion, usually over V1 to V3, and QRS duration greater than 110 ms (721). Low voltage potentials following the QRS (epsilon waves) are characteristic but seen relatively infrequently, and late potentials are observed on the SAECG in greater than 50% of individuals** . Unfortunately, SCD is frequently the first manifestation of the disease . A standardized diagnostic scheme has been formulated to establish a clinical diagnosis on a point score basis . **The annual incidence of SCD has varied, ranging from 0.08% to 9%** . In an autopsy series, 24 of 27 patients were determined to have died suddenly and 3 to have died of congestive HF. SCD occurs relatively frequently during exercise or during stress, but SCD with no apparent provocation is not uncommon . In one Italian series, up to 25% of SCD in athletes was related to ARVC . Although SCD usually occurs in individuals with grossly visible RV abnormalities, it can occur in those with only microscopic abnormalities and no obvious RV enlargement. RV dilation, precordial repolarization abnormalities, and LV involvement have been associated with risk of sudden death. Certain genetic types may be associated with higher risk of SCD . SCD in **1 or more family members intuitively suggests a higher risk of SCD** in an affected individual, but this has not been well quantified

Electrophysiological Testing

The arrhythmic manifestations of the disease are variable **The prognostic role of EP testing in patients presenting with isolated PVCs or NSVT is not known.** The response to EP testing may be influenced by the severity of the disease. Progression of disease has to be considered. EP testing has been evaluated in a limited number of patients for risk stratification. Di Biase et al. used EP testing in 17 patients with "mild" dysplasia and induced VT only in patients with spontaneous sustained VT. VT was induced in 90% of 12 patients with spontaneous sustained VT. The positive predictive value for recurrent VT was only 55%. Sustained VT could not be induced in 20 patients presenting with NSVT . In this study, inducibility was 88% in 24 of 27 patients presenting with sustained VT. EP testing, in general, is used to reproduce clinical VT and to guide ablation .

Management

The treatment of ARVC is often based on individual patient presentation and local physician experience. **The ICD has been used in patients with unexplained syncope, sustained VT, or VF** with a high incidence of appropriate shocks. Although there are no specific large randomized trials in ARVC to support this, the situation is sufficiently "similar" to those disease states such as previous MI where these indications are well established .

ICD treatment in

- Individuals with a known family history of SCD
- Unexplained syncope is intuitively compelling but not rigidly proved.

The impact of medical therapy on mortality is not established**.**

RF ablation has been used in selected patients for VT in medically refractory patients . Elimination of 1 or more clinical tachycardias by RF ablation is useful for management of symptoms but may not be sufficient to prevent SCD.Operative therapy in the form of total electrical RV disconnection hasproved successful in medically refractory patients with normal LV function but does carry a risk of postoperative right HF. Heart transplantation and ventricular assist devices are an option in patients with biventricular failure.

Genetic Analysis

Genetic analysis is useful in families with RV cardiomyopathy, because whenever a pathogenetic mutation is identified, it becomes possible to establish a presymptomatic diagnosis of the disease among family members and to provide them with genetic counseling to monitor the development of the disease and to assess the risk of transmitting the disease to offspring. Based on current knowledge, genetic analysis does not contribute to risk stratification in RV cardiomyopathy.