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Characteristics of ventricular tachycardia and long-term treatment outcome in patients with dilated cardiomyopathy complicated by lamin A/C gene mutations.

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Abstract

BACKGROUND: Dilated cardiomyopathy caused by lamin A/C gene (LMNA) mutation is complicated with atrioventricular (AV) conduction disturbances, malignant ventricular arrhythmias, and progressive severe heart failure. Radiofrequency catheter ablation (RFCA) of ventricular tachycardia (VT) has been reported to be challenging due to the high recurrence rate in patients with LMNA-related cardiomyopathy. However, electrophysiological and histopathological characteristics of VT substrate remain to be fully elucidated.

METHODS AND RESULTS: We experienced 6 familial patients with LMNA-related cardiomyopathy in 3 pedigrees (6 males, 43.7±4.5 [SD] years). All patients had first VT attack at 50±6.6 [SD] years of age, and 4 underwent RFCA for incessant VT. Their electrocardiograms during VT showed similar QRS morphologies, characterized by an inferior axis, SR pattern in aVR, and QS pattern in aVL, suggesting the origin of the basal anterior ventricle. Indeed, the VTs had multiple exits around the basal anterior ventricular septum in all RFCA cases. Although we performed multiple RFCA procedures including epicardial ablation and surgical cryoablation, all cases experienced VT recurrences in 4.5±6.4 [SD] months after last procedure. All patients developed end-stage heart failure with frequent VT events, and died at 59.5±3.6 years of age (severe heart failure in 5 and lung disease in 1). In three autopsy cases with RFCA, fibrofatty degeneration was noted in the AV node. In addition, in the deep basal ventricular septum, inhomogenous fibrotic degenerated tissue was noted beyond the reach of RF lesions.

CONCLUSIONS: These results demonstrate that patients with LMNA-related cardiomyopathy are characterized by VTs refractory to RFCA probably because of the deep intramural focus at the basal ventricular septum, resulting in poor prognosis with progressive severe heart failure despite

all available optimized therapies. Thus, we should consider heart transplantation in their early 50s when several VT events begin to occur.

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KEYWORDS: Catheter ablation; Dilated cardiomyopathy; Lamin A/C; Ventricular tachycardia

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