Cardiac pacing in 21 patients with Emery-Dreifuss muscular dystrophy: a single-centre study with a 39-year follow-up.

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Abstract

BACKGROUND: Emery-Dreifuss muscular dystrophy (EDMD) is a genetic condition associated with cardiac arrhythmias. The patients typically develop early, asymptomatic bradyarrhythmia, which may lead to sudden death, preventable with a cardiac implantable electronic device (CIED). EDMD may be characterised by atrial electrical silence. Intra-operative electrophysiological evaluation of the myocardium helps ultimately determine the true nature of the disorder and select an appropriate CIED.

AIM: To analyse permanent electrotherapy procedures in EDMD patients: atrial pacing limitations that stem from the electrophysiological properties of the myocardium and long-term follow-up of implanted devices.

METHODS: A total of 21 EDMD patients (mean age 29 ± 9 years) with a CIED implanted (1976-2014) due to bradyarrhythmia were included in the study. The implantation procedures and factors determining the CIED type selection were analysed.

RESULTS: CIEDs were implanted in five women and in 16 men with EDMD types 1 and 2 (mean follow-up: 11 ± 8 years). Intra-operatively assessed atrial electrophysiology resulted in changing the planned CIED type during the procedure in three men with EDMD type 1. Eventually, we implanted: eight DDD, one VDD, 11 VVI, and one CD-DR device, with four of the patients' devices switched later from DDD to VVI mode in response to electrophysiological changes in the atria.

CONCLUSIONS: Intra-operative assessment of atrial electrophysiological properties resulted in changing the planned DDD mode for VVI in 19% of patients with EDMD type 1. Progression of the underlying disease over a 39-year follow-up resulted in a later change of the initially selected pacing mode from DDD to VVI in 40% of cases.

KEYWORDS: Emery-Dreifuss muscular dystrophy; cardiac arrhythmias; cardiac pacing; conduction abnormalities

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