

Prevalence and predictors of bradyarrhythmias requiring Permanent Pacing in patients with Anderson-Fabry disease.

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Abstract

Introduction: Bradyarrhythmias are an established red flag for storage cardiac conditions including Anderson-Fabry disease (AFD). The prevalence of bradyarrhythmias requiring a pacemaker (PM) and their timing in AFD is unresolved. We evaluated prevalence and predictors of PM requirement in a large AFD cohort, investigating the occurrence of bradyarrhythmias as initial versus late manifestation. **Methods:** we retrospectively evaluated 82 consecutive AFD patients referred to our multidisciplinary referral centre from 1994 to 2020 with a median follow up of 6.9 years, identifying those requiring pacing. Univariable analysis was performed to identify cardiac features associated with PM implantation. **Results:** Five of 82 (6%) AFD patients required PM implantation (5/39, i.e. 13% of those with cardiac involvement), always in the context of advanced cardiomyopathy. In none, bradyarrhythmias were the presenting feature. Indications included sick sinus syndrome in 3 patients, advanced atrio-ventricular block in 2 patients. QRS prolongation during follow up strongly correlated with the onset of bradyarrhythmias. **Conclusions:** Severe bradyarrhythmias are relatively frequent in patients with AFD cardiomyopathy, but do not represent a mode of presentation, occurring late in the disease course and always in the context of advanced cardiac involvement. Monitoring QRS variations over time may help to identify patients requiring pacing.

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