Electrocardiographic abnormalities and rhythmic events in a pediatric cohort of myotonic dystrophy I

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INTRODUCTION: Myotonic dystrophy (MD1) is an inherited muscular dystrophy responsible for myocardial degeneration of the cardiac conduction system, with a high risk of sudden deaths in adults. Long-term clinical course and ECG findings in a pediatric cohort have yet to be assessed.

METHODS: We retrospectively collected clinical and electrocardiographic data in patients with proven MD1 followed at our neuromuscular disease center. An arrhythmic event was defined as syncope or sudden cardiac death of presumed arrhythmic etiology. PR interval, QRS width, and axial deviation are compared to standard data described by Davignon et al. and defined as abnormal when values are superior to the 98th percentile for PR and QRS durations, and out of the 1st and the 98th percentiles for axial deviation.

RESULTS: A total of 56 patients were included with a mean age at diagnosis of 5.3 ± 4.9 years and a follow-up duration of 7.2 (range 1.9-15.6) years. Of these patients, 27 cases (48.2%) were congenital MD1 with a reported diagnosis in the first year of life. At baseline, 21.4 % (n=12) had PR values superior to the 98th percentile and 78.5 % (n=44) had a widening of QRS. By the end of a follow up, these conduction abnormalities do not regress with PR prolongation notified in 25.0 %, and widening of QRS in 87.5%. Only 12.5 % (n=7) in the cohort had normal ECG findings at the end of follow-up. Over the course of follow-up, 5 patients had syncope (mean age 15.1 ± 3.7) and 2 patients died suddenly (mean age 17.5 ± 1.9), corresponding to an event-free survival rate of 98 % at 7.2 years. All of these patients had abnormal ECG findings but, in univariate analyses, only the width of QRS is associated with arrhythmic events (110.3 ± 8.2 ms in the event group vs. 98.1 ± 1.7ms in the no-event group, p=0.03).

CONCLUSION: During a follow up nearly 16 years, more than 85% of pediatric patients who suffered from MD1 had ECG abnormalities, which increased over time. Rhythmic events in pediatric population are rare, and occur in patients with conduction abnormalities on basal ECG.