Diagnostic value of parental electrocardiographic screening in congenital and childhood, non-immune, isolated atrioventricular block.


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Introduction: The etiology of congenital or childhood non-immune, isolated AV block remains unknown. We hypothesized that this conduction abnormality in the young may be a heritable disease.

Methods: A multicenter retrospective study (13 French referral centers, from 1980 to 2009) allowed inclusion of 141 children with AV block diagnosed in utero, at birth or before 15 years of age, without structural heart abnormalities and without maternal antibodies. Parents and matched controls were investigated for family history and for ECG screening.

Results: In parents, family history of sudden death or of progressive cardiac conduction defect was found in 1.4% and 11.1% respectively. Screening ECGs from 130 parents (mean age 42.0 ± 6.8 years, 57 couples) were compared to 130 matched healthy controls. All parents were asymptomatic and in sinus rhythm, except one with unknown complete AV block. Conduction abnormalities were more frequent in parents than in controls, respectively found in 50.8% versus 4.6% (p < 0.001). Long PR interval was found in 18.5% parents but never in controls (p < 0.001). Complete or incomplete right bundle branch block was observed in 39.2% parents and 1.5% controls (p < 0.001). Complete or incomplete left bundle branch block was found in 15.4% parents and 3.1% controls (p < 0.001). Heritability estimate for isolated conduction disturbances was very high, calculated at 91% (standard error = 1.019, p = 2.10-16).

Conclusion: ECG screening in asymptomatic parents from children affected by idiopathic AV block revealed a high prevalence of conduction abnormalities with prolongation of intra-atrial, AV and/or intraventricular conduction delay. Heritability estimate confirmed a high contribution of genetic factors. These results support the hypothesis of an inheritable trait in congenital and childhood non-immune, isolated AV blocks.