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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 remainsunknown. Wehypothesizedthatthis conduction abnormality in the youngmaybe a heritabledisease.

Methods: A multicenterretrospectivestudy (13 French referralcenters, from 1980 to 2009) allowed inclusion of 141 childrenwith AV block diagnosedin utero, atbirth or before 15 years of age, without structural heartabnormalities and withoutmaternalantibodies. Parents and matchedcontrolswereinvestigated for familyhistory and for ECG screening.

Results: In parents, familyhistory of suddendeath or of progressive cardiac conduction defectwasfound in 1.4% and 11.1% respectively. Screening ECGsfrom 130 parents (meanage 42.0 ± 6.8 years, 57 couples) werecompared to 130 matchedhealthycontrols. All parents wereasymptomatic and in sinus rhythm, except one withunknowncomplete AV block. Conduction abnormalitieswere more frequent in parents than in controls, respectivelyfound in 50.8% versus 4.6% ($p<0.001$). Long PR intervalwasfound in 18.5% parents but never in controls ($p<0.001$). Complete or incomplete right bundle branch block wasobserved in 39.2% parents and 1.5% controls ($p<0.001$). Complete or incompleteleft bundle branch block wasfound in 15.4% parents and 3.1% controls ($p<0.001$). Heritabilityestimate for isolated conduction disturbanceswasveryhigh, calculatedat 91% (standard error=1.019, p=2.10-16).

Conclusion: ECG screening in asymptomatic parents fromchildrenaffected by idiopathic AV block revealed a highprevalence of conduction abnormalitieswith prolongation of intra-atrial, AV and/or intra-ventricular conduction delay. Heritabilityestimateconfirmed a high contribution of geneticfactors. Theseresults support the hypothesis of an inheritable trait in congenital and childhood non-immune, isolated AV blocks.