Results: AV block was asymptomatic in 119 (84.4%) and complete in 100 (70.9%) patients. Incomplete AV block progressed to complete in 29 (70.7%) patients with incomplete block over 2.8±3.4 years (1-155 months). Narrow QRS complex was present in 18 of 26 patients (69.2%) with congenital, and 106 of 115 (92.2%) with childhood AV block. Pacemakers were implanted in 112 children (79.4%), during the first year of life in 18 (16.1%) and before 10 years of age in 90 (80.4%). The mean delay between diagnosis of AVB and pacemaker implants was 2.6±3.9 years (0-300 months). The pacing indication was prophylactic in 70 children (62.5%). During a median follow-up of 11.6±6.7 years (1-32 years), no patient died or developed dilated cardiomyopathy. The long-term follow-up was uncomplicated in 127 children (90.1%).

Conclusions: In this large multicenter study, the long-term outcome of congenital or childhood, isolated, non-immune AV block was favorable, regardless of the patient’s age at the time of diagnosis. No patient died or developed dilated cardiomyopathy, and pacemaker-related complications were few. The progression of incomplete to complete AV block in nearly 70% of patients suggests a postsutural degeneration of the specialized conduction system.

Clinical outcome and therapeutic strategies in non proband children with genetically confirmed LQTS

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Hypothesis: Clinical outcome is favorable in genetically confirmed non proband LQT3 children, at least in their first years of life.

Methods: Our database was searched for all patients with genotype LQTS, identified by familial screening and aged 16 or less at diagnosis. We retrospectively recorded demographic and electrocardiographic data, personal and family histories and genetic diagnoses.

Results: 90 non proband children with genetically confirmed LQTS were included. Mean age at diagnosis was 7.3±5.2 years, and mean follow-up duration was 4.6±4.3 years. 7 (7.8%) were symptomatic before diagnosis, and 4 (4.4%) experienced LQTS-related symptoms during follow-up. No sudden cardiac death was reported. One LQT1 patient presented aborted cardiac arrest while swimming. Beta-blocker therapy was initiated in 51 patients (56.7%). Device therapy was infrequently used (one pacemaker, no implantable cardioverter defibrillator). Corrected QT interval (QTc) was the only factor correlated to the risk of LQTS-related symptom (p=0.02). Initiation of betablocker therapy was associated with mutations in KCNQ1 or KCNH2 (p=0.01), family history of LQTS-related cardiac event in a first-degree relative (p=0.02), diagnosis before four months of age (p=0.01) and a longer QTc (p=0.0016).

Conclusion: Clinical outcome is favorable in genetically confirmed non proband LQT3 children, and similar to what has been previously described despite less frequent use of beta blocker and device therapies. Thus, beta blocker therapy should probably not be initiated systematically in such patients, especially when QTc duration does not exceed 460 ms. ICD implantation does not seem mandatory in non proband LQT3 children, at least in their first years of life.

Optimal dosing of warfarin in a pediatric cohort: height, INR range, VKORC1 and CYP2C9 genotypes are the main contributors of the dose requirement

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Hypothesis: Optimal dose of warfarin is not calculated in the same way in pediatric population compared to the adult one. In order to take into account patient’s characteristics and genetics, we conducted a study to assess the impact of each one on the optimal dose of warfarin.

Methods: Our database was searched for all patients with genotyped warfarin dose, selected according to the following criteria: platelet count ≥100000/mm3, INR range 2-3, available personal and family histories and genetic diagnoses.

Results: 48 patients were included. Their mean age was 14.6±5.6 years, their mean height 150.5±18 cm. VKORC1 and CYP2C9 genotypes are the main contributors of the warfarin dose.

Conclusion: VKORC1 and CYP2C9 genotypes are the main contributors of the warfarin dose.