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QTc interval in patients with non-compaction of the myocardium

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Non-compaction of the myocardium is a genetically determined cardiomyopathy with wild range of outcomes. The aim of this study was to assess the association of non-compacted myocardium and prolonged QT interval in children.
Material and methods: Patients less than 18years of age with diagnosis of isolated non-compaction of the myocardium were reviewed for clinical, ECG and echocardiographic data and outcomes. QTc interval was measured according to the Bazett formula.
Results: From 1996 to 2010, 43 patients (23 males), aged 0 to 217months at diagnosis (median 9.3months), were followed-up for 0.5 to 99months (median 8.2months). ECG was abnormal in all cases, QTc interval >440ms in 23% (QTL= 10cases), <440ms in 77% (QTN: 33cases). Twenty-two presented with heart failure (51%), 2 with shock and cardiac arrest, 3 with arrhythmia, 2 with syncopes or chest pain and 14 had no symptom (32.5%), with no difference between QTL and QTN cases. Familial reccurrence was 21% (more frequent in QTL: 40% vs 15%) and parents cosanguinity 14% (50% if QTL vs 19% in QTN). N/C index was 2.5 ± 0.7 (median 2.3): mean 3.1 in QTL group and 2.2 in QTN group (p=0.01). LV apex was involved in 96.7%, with more than 3 locations in 25.5% (33.3% in QTL, 22% in QTN) and RV involvement in 9.3%. Shortening fraction was 25.4 ± 11.6% (median 25%), not different with QTc. Nine patients were diagnosed with previous small muscular VSD (none ranged in group QTL). Six patients died (13.9%) at median age 6.2months (3.6 to 43) and median follow-up 2.7months (0.5 to 12.5). Survival was not different between QTL and QTN patients. Five underwent heart transplantation (11.6%) at median follow-up 2.8months (1.6 to 27.4). At least one episode of heart failure occurred in 18 (42%), arrhythmic or thromboembolic events in respectively 5 (11.6%) and 3 (6.9%). Bad events were more frequent in QTL group (80%) than in QTN (57.5%, p= 0.1).
Conclusion: This study showed a specific association of non-compaction cardiomyopathy with prolonged QT interval in children, which is correlated with familial occurrence, N/C index and poor outcomes.