ABSTRACT

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 18 years 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 217 months at diagnosis (median 9.3 months), were followed-up for 0.5 to 99 months (median 8.9 months). ECG was abnormal in all cases. QTc interval >440ms in 25% (QTL: 10 cases), <440ms in 75% (QTN: 33 cases). Twenty-two patients presented with heart failure (51%), 2 with shock and cardiac arrest, 3 with arrhythmia, 2 with syncope or chest pain and 14 had no symptom (32.5%), with no difference between QTL and QTN cases. Familial occurrence was 21% (more frequent in QTL: 40% vs 15%) and parents consanguinity 14% (25% in QTL vs 19% in QTN). NC/C index was 2.5 ± 0.7 (median 2.3) vs mean 3.1 in QTL group and 2.2 in QTN group (p=0.01). LV apex was involved in 96.7%, with more involvement of QTL group (25.5%) (33.3% in QTL group, 28% in QTN) and RV involvement in 9.3%. Shortening fraction was 25.4% ± 11.6% (median 25%), not different with QTL. Nine patients were diagnosed with previous small muscular VSD (none ranged in group QTL). Six patients died (13.9%) at median age 6.2 months (3.6 to 43) and median follow-up 2.7 months (0.5 to 12.5). Survival was not different between QTL and QTN patients. Five underwent heart transplantation (11.6%) at median follow-up 2.8 months (1.6 to 27.4). At least one episode of heart failure occurred in 18 (44%), arrhythmia or thromboembolic events in respectively 5 (11.6%) and 3 (6.9%). Bad events were more frequent in QTL group (40%) than in QTN (28.6%, 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, NC/C index and poor outcomes.

BACKGROUND

Non-compaction of the myocardium is a genetically determined cardiomyopathy with wild range of outcomes and prognosis factors are to be defined. Diagnosis can be made by cardiac echography.

The aim of this study was to assess the association of non-compacted myocardium and prolonged QT interval in patients less than 18 years of age at diagnosis.

QTC interval in patients with Non-Compaction of the Myocardium


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MATERIAL and METHODS

Retrospective study

Criteria for inclusion on the study:

- Patients less than 18 years of age
- No associated heart disease
- Review of clinical, ECG and echocardiographic data and outcomes.

QTc interval measured with Bazett formula.

Comparison of QTN and QTL groups

RESULTS

Clinical Presentation

Echocardiography

LV apex location

RV location

>3 locations

Shortening fraction

Muscular VSD

Outcomes

Death: 6 (13.9%) (QTL: 5, QTN: 1)

Transplantation: 1 (QTL) vs 2 (QTN)

Med 2.8 months (1.6 to 27.4) vs med 2.1 months (1.6 to 27.4)

Bad event (arrhythmia and/or thromboembolia) and/or acute heart failure

QTL 80% vs QTN 57.6%; 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, NC/C index and poor outcomes.