Hôpitaux de Lyon

Université Claude Bernard Lyon 1

CARDIOLOGIE PEDIATRIQUE ET CONGENITALE DE L'ADULTE

QTc interval in patients with Non-Compaction of the Myocardium

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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 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).

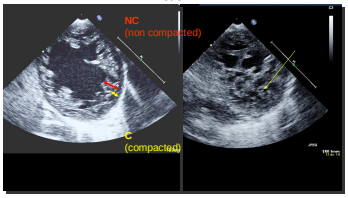
Conclusion: This study showed a specific association of non-compaction cardiomyopathy with prolonged QT interval in children, which is correlated with familial occurrence, N/C index and poor outcomes.

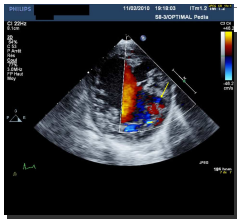
BACKGROUND

• 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 echoDoppler

• 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.

NC/C > 2





MATERIAL and METHODS

Retrospective study

Criteria for inclusion on the study :

- Patients less than 18years of age
- No associated heart disease

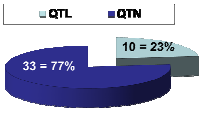
Review of clinical, ECG and echocardiographic data and outcomes.

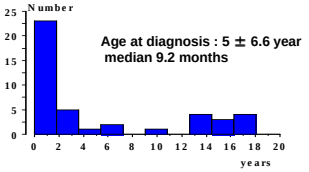
QTc interval measured with Bazett formula.

Two groups : normal QTc (QTN) / prolonged QT (QTL)

Comparison of QTN and QTL groups

N= 43
(23 males: 53.5%)

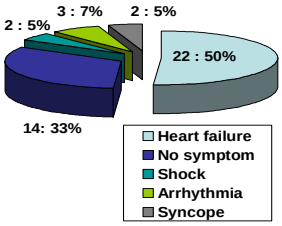




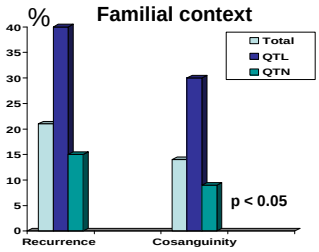
RESULTS

Clinical Presentation

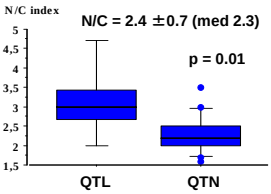
QTN *vs* QTL: NS



Familial context

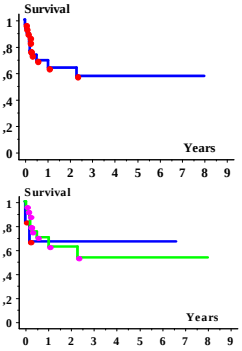


Echocardiography

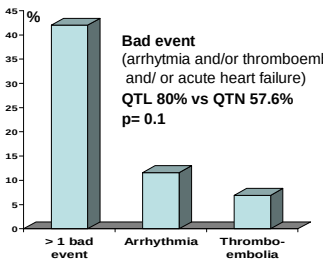


	Total	QTL	QTN	p
LV apex location	96.3%	100%	94%	NS
RV location	9.3%	0%	12%	NS
>3 locations	25.5%	33.3%	22%	NS
Shortening fraction	25.4±11.6% med 25%	23.5±14.6% med 21%	26±10.8% med 26%	NS
Muscular VSD	20.9%	0%	27.3%	NS

Outcomes



	QTL	QTN	p
Death: 6 (13.9%)	1	5	0.68
Med 2.7mths (0.5 to 12.5)	10%	15%	
Transplantation	1	4	0.85
Med 2.8mths (1.6 to 27.4)	10%	12%	



Bad event
(arrhytmia 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.

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