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Abstract

LONG QT SYNDROME: ELECTROCARDIOGRAPHY AND GENETIC CORRELATION IN MALAYSIA POPULATION

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Objective

Congenital Long-QT syndrome (LQTS) is a group of lethal cardiac arrhythmias characterized by prolonged QT interval and associated with syncope or cardiac arrest. It is potentially lethal and symptomatic patients without therapy have a high mortality rate of 21% within 1 year from the first syncope.

The importance of a correct diagnosis has assumed a new dimension in the molecular era. This is especially true in asymptomatic borderline long QT interval patient which can be observed in up to 50% of patients with LQTS because of the intermittent nature of QT prolongation. Yet they are still at risk of fatal cardiac events. We sought to profile the phenotype genotype correlation of the patients with LQTS in our institute. To our knowledge this is the largest cohort of LQTS patient analyzed in Malaysia.

Methods

We recruited all pediatric patients seen in our institute with suspected LQTS. Diagnosis of LQTS was made based on Schwartz criteria. Once consent was obtained, blood from index case and their 1st degree family members was sent to a reference laboratory in University Kebangsaan Malaysia. Clinical data were extracted from all the patients including their clinical presentation, presence of syncope and documented ventricular arrhythmias as well as the longest pre-treatment QTc.

Statistical analysis was performed by means of student *t*-test and ANOVA to determine if there were any correlation between presence of symptoms, genetically confirmed LQTS and the duration of QTc.

Result

A total of 30 patients were identified with suspected LQTS of which, 1 was excluded as the QTc pre-treatment was not available. There was only 1 death during follow up. The mean QTC was longer when patients were symptomatic as well longer in the group of patients with genetically confirmed subtypes of LQTS although statistically not significant.

No	Parameter	Yes/No	N	QTc (Mean + SD)	P value
1	All Clinical Events	Yes No	23 (79.31%) 6 (20.69%)	508.43± 61.38 476.33± 26.33	0.069 (NS)
2	Bradycardia	Yes No	14(48.28%) 15(51.72%)	500.36 ±57.53 503.13± 58.65	0.782 (NS)
3	Syncope	Yes No	9(31.03%) 20(68.97%)	538.44±51.20 485.30±52.69	0.980 (NS)
4	Heart Failure	Yes No	2(6.90%) 27(93.10%)	549.50 ±27.58 498.26 ±57.38	0.363 (NS)
5	VT/VF	Yes No	8(27.59%) 21(72.41%)	526.88± 70.64 492.24± 49.68	0.203 (NS)
6	Cardiomyopathy	Yes No	4(13.79%) 25(16.21%)	506.75 ±67.55 501.00 ±56.80	0.820(NS)
7.	Genetic result	Confirmed LQTS Others None	15(51.73%) 9 (31.03%) 5 (17.24%)	518.53± 63.07 473.44 ±43.01 502.60 ±48.25	0.175 (NS)

Table 1: Statistical analysis of the correlation between clinical symptoms and QTc duration.

Conclusion

This study demonstrates that; albeit a tendency towards longer QTc in symptomatic and genetically confirmed LQTS, no statistical significance was detected. Hence the importance of pre-symptomatic detection via family screening to enable earlier treatment. A larger cohort of patients would be required to analyze correlation of symptoms to the type of genetic abnormality in LQTS.