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Tissue Doppler Imaging in diagnosing fetal Long-QT syndrome: the development and validation of a novel diagnostic approach

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Introduction

A definite diagnosis of Long-QT syndrome (LQTS) in a fetus is impossible without invasive DNA analysis, due to the unavailability of a fetal ECG. We aimed to develop and validate a novel diagnostic approach in children using color Tissue Doppler Imaging (cTDI) in order to provide a potential marker for the prenatal diagnosis of LQTS.

Methods

Twenty-four healthy children were age- and gender-matched to LQTS patients with a confirmed pathogenic mutation. A 12-lead ECG and myocardial contraction duration (CD) on cTDI recordings were cross-sectionally obtained. Validation of the CD was done by comparing the parameter to a previously defined cTDI measurement in adult patients. Inter-method and intra-observer variability was presented in intraclass-correlation coefficients (ICC). Receiver-operating characteristic (ROC) analysis was done and the optimal cut-off value for CD was determined. Feasibility was tested in a pilot study among fetuses.

Results

LQTS children had a longer CD compared to controls ($p=0.008$), while there was no statistical difference in heart rate ($p=0.145$). CD had a high reproducibility ($ICC=0.94$), and reliably associates with the QT-interval ($ICC=0.75$) and the previously defined cTDI measure ($ICC=0.86$). The area under the curve for CD was 0.71, and an optimal cut-off value of 422 ms showed a 54% sensitivity and a 83% specificity in diagnosing LQTS. The measurement of the CD was feasible in fetuses.

Conclusion

Myocardial CD assessed by cTDI was prolonged in LQTS children, and correlated reliably with the QT-interval. Measuring the CD in fetuses seems feasible, and has the potential for diagnosing fetal LQTS. Examples of the CD measurement in fetal twins are shown in the figure.

FUTURE PERSPECTIVE

