



Analysis of myocardial fibrosis in children with hypertrophic cardiomyopathy: A report from the pediatric cardiomyopathy registry[☆]



Sonya Kirmani^{*}, Pamela Woodard, Charles E. Canter, Ling Shi, Paul Commean, Melanie Everitt, Elfriede Paul, Wendy K. Chung, Joseph Rossano, Kimberly Molina, Jason Czachor, James D. Wilkinson, Ashley Hill, Janine Wuebbles, Hiedy Razoky, Steven E. Lipshultz

Background

Myocardial fibrosis in patients with hypertrophic cardiomyopathy has been shown to have prognostic implications for sudden death. Present studies suggest using a threshold of 15% late gadolinium enhancement (LGE) on MRI as an indication for ICD placement for primary prevention in adults.

Methods

Using the Pediatric Cardiomyopathy Registry (PCMR), MRIs from 67 children with hypertrophic cardiomyopathy (median age 11.8 years, range 1–20 years) were analyzed in a core lab. Our hypothesis is that the pediatric population has less myocardial fibrosis than as reported in adults.

Results

We found the median left ventricular wall thickness was 17.8 mm (range 8.3–37 mm) and 54% of subjects had evidence for systolic anterior motion of the mitral valve. Nearly 80% of our subjects had evidence for myocardial fibrosis with median LGE of 27% (range 1–58%), predominantly in a patchy or diffuse subendocardial distribution throughout the left ventricle.

Conclusions

There are greater amounts of fibrosis in our pediatric subjects than as reported in adults. Given these findings, more investigation is warranted to correlate myocardial fibrosis with serum biomarkers, echocardiographic data, and risk factors for sudden death in children with hypertrophic cardiomyopathy.

[☆] Authors declare there is no conflict of interest.

^{*} Corresponding author.

E-mail address: kirmani@wustl.edu (S. Kirmani).