Title: Functional Status is Impaired and Correlated with Clinical Status in Pediatric Cardiomyopathy

Background. Little systematic information exists on the functional status of children with cardiomyopathy (CM) and its association with socioeconomic status (SES), growth, cardiac status, and risk of death/transplant.

Methods. The NHLBI Pediatric CM Registry measured parent-reported functional status using the Child Health Questionnaire and Functional Status IIR (FSIIR) in 249 children with cardiomyopathy.

Results. Subjects (mean age 7.9±5.9 yr) had lower functional status vs. historical controls (CHQ Physical summary score 43.2±13.1 vs. 53.0±8.8; Psychosocial 47.9±10.7 vs. 51.2±9.1), but 74% and 91% were in normal range, respectively. Poorer physical and psychosocial functioning was associated with impaired growth and parental non-completion of high school. In patients with dilated cardiomyopathy, younger age at dx (P=.004) and lower LV mass z-score (R= -0.32; slope -2.6; P=.01) independently correlated with better physical function. In patients with hypertrophic cardiomyopathy, better physical function was independently correlated with parental education (P<.001), lower LV posterior wall thickness (R= -0.29; P<.001) and closer to normal FS (R= -0.26; P=0.04). Clinical status was also associated with emotional impact of CM on parents. Across all pts, lower CHQ Physical summary score was a risk factor for death/transplant [hazard ratio 0.75 per 5-point increase in score, p=.013; hazard ratio 4.41 for score below vs. within normal range (logrank test p=.034)].

Conclusions. Functional status in children with CM is impaired relative to healthy children and is associated with SES, concurrent LV structure and function, and growth. Physical functioning
is associated with risk of death and transplant.

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