

IMPAIRED FUNCTIONAL STATUS IN CHILDREN WITH CARDIOMYOPATHY: A REPORT FROM THE PEDIATRIC CARDIOMYOPATHY REGISTRY

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Health status and functioning of children with heart disease has not been studied in a large cohort with standardized instruments. The NHLBI-sponsored Pediatric Cardiomyopathy Registry (PCMR) conducted a study of functional status in children diagnosed with cardiomyopathy (CM) at birth through age 18 years. CM was defined using strict echocardiographic criteria or biopsy. METHODS: The PCMR has over 2700 CM cases with demographic, echocardiographic, and clinical findings compiled by annual chart review. In this substudy, health status was assessed at 5 cardiac medical centers by the parents of 189 children using the Child Health Questionnaire (CHQ) for children age ≥ 5 yrs and the Functional Status II Revised (FSIIR) instrument for all ages. Both English and Spanish instruments were available. This report summarizes CHQ findings and examines the association of health status and CM severity, defined by fractional shortening (FS) Z-score for BSA. RESULTS: The CHQ sample (≥ 5 yrs old) was 11.7 ± 4.4 yr at assessment (2.9 ± 2.2 yr since CM diagnosis) and was 57% male. Pure dilated and hypertrophic CM comprised 45% and 42% of cases, and 72% were idiopathic. The echo closest to CHQ administration ($n=81$; FSz 0.45 ± 5.13 ; FSz < -2 in 25%) was between 2 and 14 months for most subjects. The CHQ Physical Summary Score was 43.1 ± 13.4 , one standard deviation below the U.S. norm for healthy children (53.0 ± 8.8 ; $p < .001$); 25% were $>2SD$ below the norm. The CHQ Psychosocial Summary Score was 48.2 ± 10.4 , also lower than the U.S. norm of 51.2 ± 9.1 ($p < .001$). CHQ domain scores indicated that greatest impairment was specifically in lower physical functioning and limitations in everyday physical activities, child self-esteem, impact on parental emotions, and general health perceptions. Although FS Z-score (< -2 vs. ≥ -2) was not associated with CHQ Physical Summary Score (43 ± 13 vs. 46 ± 12), there was a significant correlation with Impact on Parental Time domain ($R = .30$, $p = .007$), with lower CHQ score in children with FSZ < -2 (67 ± 35 vs. 88 ± 20 , $p = .010$). CONCLUSIONS: One-quarter of children with CM have severely impaired physical functioning $> 2SD$ below the norm, and psychosocial functioning is also affected.