

Clinicopathological investigation on pediatric cardiomyopathy with special reference to ongoing myocardial damages using pathophysiological parameters

Yonesaka S.(1), Takahashi T. (2), Otani K. (2), Jun S. (2), Konno Y. (2), Sato T. (2), Kitagawa Y. (2), Eto S. (2), Ueda T.(2), Sato A. (2), Ichinose K. (2), Kinjo M. (2)

Department of Nursing, Hirosaki University School of Health Sciences, Hirosaki, Japan(1);Department of Pediatrics, Hirosaki University School of Medicine, Hirosaki, Japan(2)

Purpose: Clinical significance of ongoing myocardial damage in pediatric cardiomyopathy has not been well defined. Furthermore, it is not so easy to estimate the severity of myocardial damages only from clinical symptoms and signs. To clarify the possible prognostic impact of ongoing myocardial damages, we investigated pathophysiological parameters, such as symptoms and signs related to HCM, dysrhythmias and ST-T and changes by Holter ECG and exercise ECG, electrophysiological study, biochemical markers and endomyocardial biopsy (EMB). Patients and method: They included 18 hypertrophic cardiomyopathy (HCM); with pressure gradient 7, without 11, aged from 2.5 to 18years, 7 patients with dilated cardiomyopathy(DCM); aged from 1 to 15 years. Male to female was 13 to 12. Selected biochemical markers in this study were high-sensitive C-reactive protein (hsCRP), myoglobin, Creatin Kinase MB (CK-MB), troponin T (TnT), heart-type fatty acid binding protein (H-FABP), ANP and BNP. Histopathology was evaluated with histomorphometric method. The fraction of myocardial volume occupied by fibrillar collagen (% fibrosis) and diameter of myocytes were detected by quantitative morphometry with an automated image analysis system. Results: Death or resuscitated sudden death occurred in 7 patients. Hypotensive response on rapid atrial pacing on EPS and exercise test was in 4. Pacemaker implantation was required in 2. Myocardial changes on EMB showed degeneration, interstitial fibrosis and disarray of myofibrils, inflammatory cell infiltration and microangiopathy. % fibrosis showed higher in cardiac death patients. Prevalence of abnormal biochemical markers was found in 35% of H-FABP, 35% of myoglobin, 32% of CK and 60% of ANP and BNP. Conclusions: Many cases with cardiomyopathy showed raised concentration of biochemical markers. Although the correlation between H-FABP concentration and heart failure severity was reported in a previous study, in this study clinical severity did not reveal statistic correlation with biochemical markers and histopathological severity obtained by endomyocardial biopsy. Nevertheless, H-FABP and other biochemical markers in cardiomyopathy might be one of the plausible predictors for the ongoing myocardial damage and ongoing ischemia. An aggressive diagnostic evaluation to detect ischemia and the use of a multifaceted treatment approach to prevent ischemia and sudden death may be beneficial.