

Clinicopathological investigation on dilated cardiomyopathy in infants and children: The role of endomyocardial biopsy and biochemical markers.

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Purpose: Early diagnosis of dilated cardiomyopathy (DCM) enables the start of effective treatment with the purpose of stopping the progress of the disease and delaying the development of symptomatic heart failure. Regarding this fact, routine and widely accessible diagnostic methods in assessing the state of advancement of the disease and the prognosis are requiring. Current applications of biomarkers may offer a means of identifying high-risk patients before they develop symptoms not only in adults but also in the pediatric population, however there are some differences from adults in the approach to the diagnosis and management of cardiomyopathy in children. In this aspect, we try to make clear the prognostic factors in pediatric cardiomyopathy comparing clinical symptoms with biomarkers and ultrastructural changes of pediatric patients with myocardial involvement by endomyocardial biopsy (EMB). Patients and method: They included 28 DCM; 7 patients with idiopathic DCM, 3 dilated phase hypertrophic cardiomyopathy(d-HCM), 2 noncompaction of left ventricle (NCVM), 7 myocarditis, 6 tachycardia induced cardiomyopathy(TIC), 2 hyperthyroidism and 36 Kawasaki disease(KD) as comparative subjects. Selected biochemical markers were high-sensitive CRP, myoglobin, Creatin Kinase MB, troponin T, heart-type fatty acid binding protein, ANP and BNP. Histopathology was evaluated with semiquantitative morphometry with an automated image analysis system. Results: Resuscitated sudden death occurred in 3 of idiopathic DCM and NCVM. Elimination of focus following EPS was 1 for TIC. ICD implantation was one for d-HCM. Myocardial changes on EMB showed various abnormalities such as inflammatory cell infiltration and higher % fibrosis in cardiac death patients. Prevalence of abnormal biochemical markers was found in severely symptomatic DCM rather than KD. Conclusions: Severely symptomatic DCM showed raised concentration of biochemical markers. Although clinical severity did not reveal statistic correlation with biochemical markers and histopathological severity, biochemical markers might be one of the plausible predictors for the severity of myocardial damage and ongoing ischemia. EMB may still be helpful to determine etiology in an otherwise undiagnose dilated cardiomyopathy in children. An aggressive diagnostic evaluation to detect severity of myocardial damage with biochemical markers and the use of a multifaceted treatment approach to prevent developing symptoms and sudden death should be required.