Clinicopathological investigation on cardiomyopathy with special reference to fatal cardiomyopathy in childhood using pathophysiological parameters: The impact of noncompaction cardiomyopathy on pediatric cardiomyopathy.

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Purpose: This study examines the demographic, clinical and histopathological features of fatal cardiomyopathy in childhood with special reference to its subtype and pathophysiologic parameters. Early diagnosis of cardiomyopathy enables the start of effective treatment with the purpose of better outcomes in this population. Noncompaction of the ventricular myocardium (NCVM) has gained increasing awareness and attention and become a widely recognized cardiomyopathy. The clinical spectrum of this complex pathology is highly varied. Accurate diagnosis of NCVM is clinically important as NCVM can present as sudden unexpected death. In this aspect, we try to make clear the prognostic factors in pediatric cardiomyopathy with special reference to fatal cardiomyopathy in childhood. Clinical symptoms were compared with baseline ECG, 2DE, MRI, biomarkers and endomyocardial biopsy (EMB). Patients and method: Between 1990 and 2011, a total of 46 cardiomyopathy patients were enrolled, including 21 hypertrophic cardiomyopathy (HCM) and 25 dilated cardiomyopathy (DCM); 7 idiopathic DCM, 4 NCVM, 8 myocarditis, 6 tachycardia induced cardiomyopathy (TIC). 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. Results: Resuscitated sudden death occurred in 2 out of 7 idiopathic DCM and in 3 out of 4 NCVM and 7 out of 21 HCM. Ablation was 1 for TIC. Pacemaker implantation was one for HCM, ICD implantation was one for d-HCM. Histopathology on EMB showed abnormalities of inflammatory cell infiltration, vacuolar degeneration, lysis of myofibrils and higher % fibrosis in cardiac death patients. Fatal cardiomyopathy was more frequent in NCVM with LQTc on baseline ECG. Conclusions: Although clinical severity did not reveal statistic correlation with biochemical markers and histopathological severity, symptomatic patients showed raised concentration of biochemical markers, so biochemical markers might be one of the plausible predictors for the severity of myocardial damage and EMB may still be helpful to determine etiology in undiagnose cardiomyopathy. High incidence of sudden cardiac death was found in NCVM with long QTc of ECG. Long QTc could be predictive of a poor prognosis. Therefore, early therapeutic implications for this condition may lead to better outcomes in this population.