

Characteristics of in paediatric patients with hypertrophic cardiomyopathy -Effect of school-based cardiovascular screening on the prognosis

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Background

A school-based screening program to uncover cardiovascular disease is effective for early diagnosis and prevention of symptoms of inherited arrhythmia syndrome, particularly long QT syndrome, in Japan. However, little is known whether this program improves the prognosis of hypertrophic cardiomyopathy (HCM).

Purpose

To determine whether the screening program is effective for early diagnosis, prevention of symptoms or improvement of prognosis of childhood HCM.

Methods

A nation-wide study group for pediatric cardiomyopathy obtained data of patients aged <20 years who visited hospitals between 2000 and 2017. Data included age at diagnosis, sex, diagnostic event (by the screening program, or not), symptoms before and after diagnosis, echocardiographic data, presence or absence of medication or non-pharmacological treatment (surgical intervention including myectomy, implantable cardioverter defibrillator (ICD) implantation, etc), and poor prognosis. Poor prognosis was defined as the presence of death/coma, heart transplantation, and out-of-hospital cardiac arrest (OHCA).

Results Among 375 patients with pediatric cardiomyopathy obtained, 134 had HCM. Of 96 and 38 patients with primary and secondary cases, respectively, 55 (57%) and seven (18%) cases were diagnosed with the screening program. Among the 96 primary cases, 10 patients had a poor prognosis; two patients died, one had transplantation, and nine had OHCA (overlapping was present). Fourteen patients received non-pharmacological treatment. Multivariate regression analysis using a poor prognosis as the dependent variable showed that the presence of symptoms before diagnosis (odds ratio, 9.88; 95% CI, 1.89–51.7; $P < 0.01$) and a higher left ventricular wall thickness (interventricular septal + posterior wall thickness) (1.07, 1.01–1.14, $P = 0.03$) were independent risk factors in primary HCM. No independent risk factors were found in secondary HCM. Screening with the program was not a predictive factor for lowering the prevalence of a poor prognosis.

Conclusion

Establishment of electrocardiographic and echocardiographic screening criteria for childhood HCM and early diagnosis and intervention before appearance of symptoms are essential for improving the prognosis of primary HCM in the pediatric population.