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Clinical and electrocardiographic features of restrictive cardiomyopathy in children

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On behalf of the Guidelines on the Electrocardiographic, echocardiographic, and genetic criteria for Cardiomyopathy in Children

Introduction: Restrictive cardiomyopathy (RCM) is a rare myocardial disease with an impaired diastolic function and poor prognosis. The mean survival duration after a diagnosis of RCM is reported to be around 2 years in children and most need heart transplantations.

Purpose: This study aimed to determine the 12-lead electrocardiogram (ECG) diagnostic criteria of RCM based on the initial diagnostic electrocardiogram.

Methods: ECGs in pediatric cardiomyopathy patients were collected from 15 institutes in Japan between 1979 and 2013. We compared the ECG findings, especially of the P wave, in RCM patients between the cardiomyopathy group and healthy children group separately for each gender and the age. The ECGs in the healthy group were obtained from school heart screening in Japan of first-graders, and seventh-graders. Statistical significance was determined as $p<0.001$.

Results: Among 376 registered cardiomyopathy patients, 63 had hypertrophic cardiomyopathy (HCM) (36%), 91 (24%) dilated cardiomyopathy (DCM), 106 (28%) a left ventricular myocardial noncompaction (LVNCs), 25 (7%) restrictive cardiomyopathy (RCM), 14 (4%) arrhythmogenic right ventricular cardiomyopathy (ARVC), and 5 (1%) other cardiomyopathies. Of the 25 RCM patients (9.9±3.4 years old, F:M=11:14), 36% were discovered during school heart screening. The first onset was an abnormal ECG in 9, symptoms of heart failure in 6, respiratory tract infections in 3, syncope in 1, and 6 with other. Of those patients, 2 (8%) had a family history of RCM, 24 (92%) no family history. A genetic diagnosis was performed in 5 of the 25 cases, and 3 had genetic abnormalities related to RCM. The mean follow-up period was 65±95 months (mean±standard deviation). During follow up, 19 patients (76%) survived, 6 (24%) died, 7 (28%) had heart transplantations, and 3 (12%) were waiting for heart transplantations with a left ventricular assist device.

The P wave was bimodal in lead I or biphasic in lead V1 in 15 patients (93%), and 13 (81%) patients had both variations. We evaluated the duration and amplitude of the first and second component of the P wave as P1 and P2. The number of control and RCM patients (control/RCM), duration of P1+P2, and sum total absolute value of the amplitude of P1+P2 in lead V1 were 8350/5, 90±9/116±10ms, and 72±28/528±278μV in first grade boys, 8423/3, 91±10/120±22ms, and 66±28/326±229μV in first grade girls, 8943/1, 97±1/100ms, and 71±31/328μV in seventh grade boys, and 9183/5, 98±11/112±10ms, and 55±27/315±56μV in seventh grade girls. Although the number of patients in the RCM group was small, sum total absolute value of the amplitude of P1+P2 in lead V1 showed a significant difference in any group.

Conclusion: The ECG in children with RCM exhibits P wave abnormalities in almost all patients. In particular, not the P wave interval but P wave shape in I and V1 and the sum total absolute value of the amplitude of P1+P2 in lead V1 were observed differences.