Clinical and electrocardiographic features of arrhythmogenic right ventricular cardiomyopathy in children

Authors:
T Imamura¹, N Sumitomo¹, S Muraji¹, K Yoshida², M Iwamoto³, E Nishihana⁴, S Tateno⁵, S Doi⁶, T Hata⁷, S Kogaki⁸, H Horigome⁹, S Ohno¹⁰, F Ichida¹¹, M Nagashima¹², M Yoshinaga¹³, ¹Saitama International Medical Center, Department of Pediatric Cardiology - Hidaka - Japan, ²Aichi Children’s Health and Medical Center, Department of Cardiology - Obu - Japan, ³Saiseikai Yokohama City Eastern Hospital, Department of Pediatrics - Yokohama - Japan, ⁴Ogaki Municipal Hospital - Ogaki - Japan, ⁵Chiba Cardiovascular Center, Department of Pediatrics - Ichihara - Japan, ⁶Tokyo Medical And Dental University, Department of Pediatrics - Tokyo - Japan, ⁷Fujita Health University, Department of Pediatrics - Toyoake - Japan, ⁸Osaka General Medical Center, Department of Pediatrics - Osaka - Japan, ⁹Ibaraki Children's Hospital, Department of Pediatric Cardiology - Mito - Japan, ¹⁰National Cerebral and Cardiovascular Center - Osaka - Japan, ¹¹University of Toyama, Faculty of medicine - Toyama - Japan, ¹²Aichi Saiseikai Rehabilitation Hospital - Nagoya - Japan, ¹³National Hospital Organization Kagoshima Medical Center, Department of Pediatrics - Kagoshima - Japan,

On behalf: On behalf of study group for Guidelines of Electrocardiographic, echocardiographic, and genetic criteria for Cardiomyopathy in Children

Topic(s):
Ventricular Arrhythmias and SCD - Pathophysiology and Mechanisms: Arrhythmogenic Right Ventricular Cardiomyopathy

Citation:

Funding Acknowledgements:
Japanese governmental grant

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a rare progressive myocardial disease characterized by fat and fibrotic degeneration of the right ventricle. Ventricular arrhythmias originating from fibrofatty tissue replaced by myocardium are often recognized between the ages of 20 to 40, and may cause sudden cardiac death. However ARVC is extremely rare under the age of 20.

The purpose of this study was to clarify the epidemiology, treatment, and electrocardiographic characteristics of pediatric patients with ARVC.

Pediatric cardiomyopathy patients between 1979 and 2013 were collected from 15 institutes in Japan.

Among 376 registered cardiomyopathy patients, 63 had hypertrophic cardiomyopathy (HCM) (36 %), 91 (24 %) dilated cardiomyopathy (DCM), 106 (28 %) left ventricular myocardial densification disorders (LVNCs), 25 (7 %) restrictive cardiomyopathy (RCM), 14 (4 %) ARVC, and 5 (1 %) with other cardiomyopathies. Of 14 ARVC patients (M:F = 7:7), 65 % were discovered during school heart screening. The first onset was cardiopulmonary resuscitation, chest pain, and fetal echocardiography in 1 patient. Most patients were asymptomatic at the first clinic visit. In those patients, 3 (22 %) had a family history of ARVC. The electrocardiogram showed no e waves in any cases, and no delayed S-wave upstroke was observed except for in several cases, but most patients had a T-wave inversion in the right precordial and inferior leads. A genetic diagnosis was performed in 9 out of 14 patients, and genetic abnormalities related to ARVC were found in 8 (89 %). Of 14 patients, 10 (72 %) received treatment, and 3 were untreated. Beta blockers were the most frequent medication, but diuretics, vasodilators, and anticoagulants were used depending on the cases.
pharmacological therapies (ICD, CRT-D, and catheter ablation) were performed in 3 patients respectively. Although 11 patients (79 %) survived, 2 (14 %) had an episode of out-of-hospital cardiac arrest.

Unlike adults, e-waves and a delayed S-wave upstroke were often negative in children with ARVC. Children with a definite clinical diagnosis had a high abnormality rate of ARVC genes, whereas the incidence of a family history was low, so a de novo mutation was considered to be the cause in most patients. It is necessary to propose new diagnostic criteria for children with ARVC.