Pediatrics and Neonatology (2018) xx, 1–9

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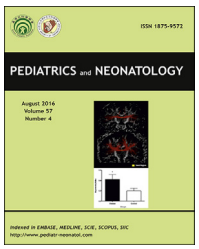

Review Article

Cardiomyopathy in the pediatric patients

Shi-Min Yuan*

Department of Cardiothoracic Surgery, The First Hospital of Putian, Teaching Hospital, Fujian Medical University, Putian, Fujian Province, People's Republic of China

Received Dec 16, 2015; received in revised form Apr 16, 2017; accepted May 9, 2017
Available online ■ ■ ■

Key Words
cardiomyopathy;
classification;
pediatrics

Pediatric cardiomyopathies are a group of myocardial diseases with complex taxonomies. Cardiomyopathy can occur in children at any age, and it is a common cause of heart failure and heart transplantation in children. The incidence of pediatric cardiomyopathy is increasing with time. They may be associated with variable comorbidities, which are most often arrhythmia, heart failure, and sudden death. Medical imaging technologies, including echocardiography, cardiac magnetic resonance, and nuclear cardiology, are helpful in reaching a diagnosis of cardiomyopathy. Nevertheless, endomyocardial biopsy is the final diagnostic method of diagnosis. Patients warrant surgical operations, such as palliative operations, bridging operations, ventricular septal maneuvers, and heart transplantation, if pharmaceutical therapies are ineffective. Individual therapeutic regimens due to pediatric characteristics, genetic factors, and pathogenesis may improve the effects of treatment and patients' survival.

1. Introduction

Pediatric cardiomyopathy is a rare but sometimes fatal myocardial disease affecting the pediatric population.[1] Cardiomyopathy can occur in children at any age,[2] and it is a common cause of heart failure and heart transplantation in children.[3] The incidence of pediatric cardiomyopathy is 4.8 per 100,000 infants and 1.3 per 100,000 children less than 10 years of age,[4] and an increasing incidence of this condition has been noted.[5] As reported by Cox et al.,[1] the patient population of pediatric cardiomyopathies was divided into 4 types: hypertrophic (34.2%), dilated (53.8%), restrictive (3.2%), and other or mixed (8.9%). Nevertheless, some pediatric cardiomyopathy cases were with an undetermined or idiopathic etiology, which accounted for 57–68%. Moreover, arrhythmogenic right ventricular dysplasia and the unclassified group including endomyocardial fibroelastosis are less common.[4] Genetic studies have revealed that cardiomyopathies, particularly the hypertrophic type, are caused by mutations of genes encoding the contractile sarcomeric proteins, such as *β-myosin heavy chain*, *myosin-binding protein C*, and *troponins T* and *I*, etc.[6] Recently, a novel pediatric

* The First Hospital of Putian, Teaching Hospital, Fujian Medical University, 389 Longdejing Street, Chengxiang District, Putian 351100, Fujian Province, People's Republic of China.
E-mail address: shiminyuan@126.com.

https://doi.org/10.1016/j.pedneo.2017.05.003

Please cite this article in press as: Yuan S-M, Cardiomyopathy in the pediatric patients, Pediatrics and Neonatology (2018), https://doi.org/10.1016/j.pedneo.2017.05.003

cardiomyopathy gene, *ALPK3*, was found to have potential implications on cardiomyopathy.[7] Nevertheless, the etiologies of cardiomyopathies are still uncertain. The prognosis of the pediatric patients with cardiomyopathy was poorer, especially in those with a cardiomyopathy of a known etiology, which showed a higher risk of death or heart transplantation.[8] Despite long-term observations, the complexity of this condition was not well understood in terms of etiologies, taxonomy, clinical manifestations, and treatment. The purpose of this article is to give an overview of pediatric cardiomyopathies.

2. Taxonomies

Cardiomyopathies can be classified as ischemic or non-ischemic. However, in pediatric patients, cardiomyopathies are all considered nonischemic, causing compromises of both structure and function of the heart.[9] In 1995, the World Health Organization and International Society and Federation of Cardiology modified the classification of cardiomyopathy into 5 types: dilated, hypertrophic, restrictive, arrhythmogenic, and inclassified.[10] In 2006, the American Heart Association updated the definition and classification by defining them as *a heterogeneous group of myocardial diseases with mechanical and (or) electrical dysfunction*,[11] which were categorized into primary and acquired types. In 2007, cardiomyopathy was classified into familial/genetic and non-familial/non-genetic[12] (Table 1). However, the classifications of cardiomyopathies are very complex, because cases can actually be classified as more than one type or change from one type to another.[13]

3. Clinical presentations

The symptoms of cardiomyopathy depend on the presence of heart failure. The exact symptoms depend on the type of

Table 1 Classifications of cardiomyopathy.

Year	Organization	Classification
1995	World Health Organization & International Society and Federation of Cardiology	HCM; DCM; ARVC; RCM; Unclassified
2006	American Heart Association	Genetic: HCM, ARVC/D, LVNC, Glycogen storage, Conduction disorder, Mt. myopathies, Ion channel disorder; • LQTS • Brugada • SQTS • CVPT • Asian SUNDS Mixed: DCM, RCM Acquired: Inflammatory (myocarditis), Stress (Takotsubo), Peripartum, Ischemic, Toxic (Alcoholic), Tachycardia, Infant of diabetic mother
2007	European Society of Cardiology	Familial/Genetic: Undetermined gene defect; Subtype disorder Non-familial/Non-genetic: Idiopathic; Subtype disorder

ARVC/D: arrhythmogenic right ventricular cardiomyopathy/dysplasia; CVPT: catecholaminergic polymorphic ventricular tachycardia; DCM: dilated cardiomyopathy; HCM: hypertrophic cardiomyopathy; LQTS: Long QT syndrome; LVNC: left ventricular non-compaction cardiomyopathy; Mt.: mitochondria; RCM: restrictive cardiomyopathy; SQTS: short QT syndrome; SUNDS: sudden unexpected nocturnal death syndrome.

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