DILATED CARDIOMYOPATHY AND CURRENT MANAGEMENT IN CHILDREN

Essay
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By

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List of Abbreviations

ACE	Angiotensin Converting Enzyme
AD	Autosomal Dominant
ALCAPA	Anomalous Left Coronary Artery arising from Pulmonary Artery
AOR	Aortic Root Diameter
ARVC	Arrhythmogenic Right Ventricular Cardiomyopathy
ATP	Adenosine Triphosphate
BiPAP	Bimodal Positive Airway Pressure
C-AMP	cyclic Adenosine Monophosphate
CAR	Coxsackievirus and Adenovirus Receptor
CHF	Congestive Heart Failure
CI	Cardiac Index
cMRI	Cardiac Magnetic Resonance
CPAP	Continuous positive airway pressure
CPT	Carnitine Palmitoyl Transferase
CRP	C Reactive Protein
CSD	Cardiac Support Device
CVB	Coxsackie-Virus B
DCM	Dilated Cardiomyopathy
DM	Diabetes mellitus
DNA	Deoxyribonucleic acid
DO2	Oxygen Delivery
ECG	Electrocardiogram
EDC	Emergency Department Care
ED	Emergency Department
EFE	Endocardial Fibroelastosis
ELISA	Enzyme Linked Immunosorbent Assay

EMS	Emergency Medical Services
GH	Growth Hormone
GM-CSF	Granulocyte Monocyte Colony Stimulating Factor
GM-CSFR	Granulocyte Monocyte Colony Stimulating Factor Receptor
HCM	Hypertrophic Cardiomyopathy
HIV	Human Immunodeficiency Virus
HLA	Human Leukocyte Antigen
ICD	Implantable Cardioveter-defibrilator
ICM	Ischemic Cardiomiopathy
ICU	Intensive Care Unit
IDC	Idiopathic Dilated Cardiomyopathy
IDCM	Idiopathic Dilated Cardiomyopathy
IgG	Immunogloulin G
IL	Interleukin
ISFC	International Society and Federation of Cardiology
IU	International Unit
IV	Intravenous
LAD	Left Atrial Diameter
LV	Left Ventricle
LVAS	Left Ventricular Assist System
LVEDD	Left Ventricular End Diastolic Diameter
LVNC	Left Ventricular Non Compaction
MIBI	Te 99 Msescamibi
MR	Mitral Regurgitati
MUGA	Multiple Gated Acquisition
MYH	Myosin Heavy Chain
NOS	Nitric Oxide Synthetase
NTHA	New York Heart Association

PCR	Polymerase Chain Reaction
PG	Prostaglandin
PLN	Phospholamban
RA	Rheumatoid Arthritis
RCM	Restrictive Cardiomyopathy
RNA	Ribonucleic acid
RV	Right ventricle
RVEF	Right Ventricular Ejection Fraction
SA	Sinoatrial node
SLE	Systemic Lupus Erythromatosis
SR	Sarcoplasmic Reticulum
SVR	Systemic Vascular Resistance
TD	Tissue doppler
TNF	Tumor Necrosis Factor
TNNT	Troponin T
TTE	Transthoracic Echocardiography
VO2	Oxygen consumption
WHO	Word Health Organization
XL	X-Linked

ABSTRACT

Dilated cardiomyopathy (DCM) refers to congestive cardiac failure secondary to dilatation and systolic (and/or diastolic) dysfunction of the ventricles (predominantly left).DCM is the most common type of heart muscle disease in children. Approximately 30-40%of all cases of DCM have a genetic or inherited basis. The major problems of DCM are; progressive heart failure, arrhythmias, thromboembolism and sudden death. Doppler echocardiography offers an excellent non invasive diagnostic method with ejection fraction below 45% is required for diagnosis. The treatment of dilated cardiomyopathy is essentially the treatment of heart failure**.** Vigorous treatment of heart failure may result in temporary remission, but relapses are common. When medical therapy fails, heart transplantation has been effective. several studies show that stem cell therapy may someday be used to reverse myocardial dysfunction.

Key word:

(Dilated cardiomyopathy, Myocarditis, Echocardiography, Carnitine, Stem Cell Transplantation)

INTRODUCTION

Cardiomyopathy is a serious disease with a poor prognosis and high mortality. The incidence varies widely but ranges between 2-17.2 per 100.000 with poorer survival were noted for old children ***(Ferencz and Neill, 1992).***

Cardiomyopathy is divided into 3 types (dilated, hypertrophic and restrictive). Of these, dilated cardiomyopathy is the most common and represents a large subset of congested heart failure cases. It is characterized by depressed systolic function, cardiomegaly and ventricular dilatation ***(Mobini et al., 2004).***

Dilated cardiomyopathy (DCM) is uncommon among children but constitutes the principal indication for cardiac transplantation in childhood **(*Boucek and Boucek, 2002)***.

According to ***O'connell et al. in 1993***, dilated cardiomyopathy is usually idiopathic however ***Winter and Buist in 2000*** described more than 75 specific diseases of heart muscle which can produce dilated cardiomyopathy. Approximately 30-40% of patients with dilated cardiomyopathy have a familial inheritance in the form of autosomal dominant transmission ***(Grunig et al., 1998)***.

Dilated cardiomyopathy is one of the most common causes of heart failure among children and is often progressive despite maximal medical therapy. Signs of congestive heart failure vary according to the age of child ***(Kay et al., 2001).***

Doppler echocardiography offers an excellent non invasive diagnostic method for detection of cardiomyopathy ***(Olson and Chan, 2001*)**. The classic picture is in the form of asymptomatic left ventricular systolic, diastolic dysfunction or both ***(Burger et al., 2002)***. ECG also shows variable arrhythmias in 50% of patients on initial evaluation ***(Colan and Newburger, 2001)***. Biopsy may be needed to identify the exact etiology ***(Olson and Chan, 2001).***

The treatment of dilated cardiomyopathy is essentially the treatment of heart failure ***(O'connell et al., 1993)***. The main drug therapies include inotropes (e.g., digoxin) to strengthen myocardial contraction; diuretics, such as furosemide, to control pulmonary congestion; and afterload reduction ***(Walter, 2001)***. ACE inhibitors are paramount in treatment of idiopathic dilated cardiomyopathy ***(Mohan et al., 2002).***

Vigorous treatment of heart failure may result in temporary remission, but relapses are common, and in time patients tend to become resistant to therapy, once this point is reached, the prognosis for survival beyond a year is poor. Patients with severely depressed myocardial function should be monitored closely for arrhythmias and, if present, treated aggressively with antiarrhythmic agents or an implantable cardioverter-defibrillator (ICD). They should also receive systemic anticoagulation ***(Dubin et al., 1999).***

β-adrenergic blocking agents, introduced gradually as part of a comprehensive heart failure treatment program, improve exercise tolerance, decrease hospitalizations, and reduce overall mortality. The chronic treatment of patients with heart failure should not be

administered when patients are still in the acute phase of heart failure ***(Bruns et al., 2001).***

In the cardiac tissue of the patients with dilated cardiomyopathy, a low level has been determined in the carnitine concentration, when compared with the normal ones. When L-carnitine is given in high doses, it has been discovered that the contractility of the left ventricle is increased ***(Donder et al., 1998).***

When medical therapy fails, heart transplantation has been effective in infants and children with dilated cardiomyopathy ***(Burch and Runciman, 1996).***

AIM OF THE WORK

The objective of this study is to review the topic of dilated cardiomyopathy and to throw light on the classic, current and recent trends in the management of dilated cardiomyopathy.

CARDIOMYOPATHIES

Cardiomyopathy constitutes a group of disorders in which the dominant feature is direct involvement of the heart muscle itself. They are distinctive because they are not a result of pericardial, hypertensive, congenital, or valvular diseases ***(Wynne and Braunwald, 2005).***

Classification of cardiomyopathy

A variety of schemes have been proposed for classifying the cardiomyopathies. The most widely recognized classification is that promulgated jointly by the World Health Organization (WHO) and the International Society and Federation of Cardiology (ISFC).

In WHO/ISFC classification, the cardiomyopathies are classified on the basis of their predominant pathophysiological features; other diseases that affect the myocardium but that are associated with a particular cardiac disorder or are part of a generalized systemic disorder are termed ***specific cardiomyopathies (Nakata and Koga,2000)***

Three basic types of functional impairment have been described:

1. **Dilated cardiomyopathy (DCM),** the most common type of heart muscle disease in children and refers to congestive cardiac failure secondary to dilatation and systolic (and/or diastolic) dysfunction of the ventricles (predominantly left) ***(Poothirikovil et al., 2007).***

2. **Hypertrophic cardiomyopathy (HCM),** recognized by inappropriate left ventricular hypertrophy, often with asymmetrical involvement of the inter ventricular septum, with preserved or enhanced contractile function until late in the course of the disease.

3. **Restrictive cardiomyopathy (RCM),** the least common form in Western countries, marked by impaired diastolic filling and in some cases with endocardial scarring of the ventricle ***(Artz and Wynne, 2000).***

Two less common form of cardiomyopathy are recognized ***arrhythmogenic right ventricular cardiomyopathy (ARVC)*** and ***unclassified;*** the latter includes fibroelastosis, systolic dysfunction with minimal dilation, and isolated ventricular non compaction, an unusual disease marked by prominent endocardial thickening with prominent trabeculations and deep recesses. Furthermore, ventricular dilation and systolic heart failure may occur late in the course of HCM and bear a resemblance to DCM ***(Oechslin et al., 2000)***.

This WHO/ISFC classification is shown in table(1).