MANAGEMENT OF CARDIOMYOPATHY

Essay

Submitted for partial fulfillment of master degree in General Intensive Care

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Abbreviations

ACE: Angiotensin-Converting Enzyme

AF: Atrial Fibrillation

AHA: American Heart Association

AII: Angiotensin II

AMP: Adenosine Monophosphate

ARB: Angiotensin-II Receptor Blocker

ARVC/D: Arrhythmogenic Right Ventricular

Cardiomyopathy / Dysplasia

ARVC: Arrhythmogenic Right Ventricular Cardiomyopathy

ASA: Alcohol Septal Ablation

B1ARs: **B**1-Adrenoceptors

BNP: Brain Natriuretic Peptide

CAD: Coronary Artery Disease

CMR: Cardiovascular Magnetic Resonance

CPVT: Catecholaminergic Polymorphic Ventricular

Tachycardia

CRT: Cardiac Resynchronisation Therapy

cTnI: Cardiac Troponin I

cTnT: Cardiac Troponin T

DCM: Dilated Cardiomyopathy

ECG: Electrocardiogram

EMF: Endomyocardial Fibrosis

ESC: European Society of Cardiology

FDC: Familial Dilated Cardiomyopathy

HCM: Hypertrophic Cardiomyopathy

HES: Hypereosinophilic Syndrome

HF: Heart Failure

H-FABP: Heart type Fatty Acid Binding protein

HFE: The official gene symbol for High Iron Fe

HFrEF: Heart Failure with Reduced Ejection Fraction

HH: Hereditary hemochromatosis

HJV: Hemojuvelin

ICD: Implantable Cardioverter Defibrillator

IFN-g: Interferon gamma

IL-6: Interlukin 6

IV: Intravenous

kDa: Kilodalton

LAD: Left-Axis Deviation

LAE: Left Atrial Enlargement

LAMP-2: Lysosome-Associated Membrane Protein-2

LBBB: Left Bundle Branch Block

LGE: Late Gadolinium Enhancement

LIM protein: Protein composed of LIM domains {Lin-

11, Isl-1, Mec -3}

LQTS: Long QT Syndrome

LV: Left Ventricle

LVEF: Left Ventricular Ejection Fraction

LVH: Left Ventricular Hypertrophy

LVNC: Left Ventricular Noncompaction

LVOT: Left Ventricular Outflow Tract

NE: Norepinephrine

NICMP: Nonischemic Cardiomyopathy

NYHA: New York Heart Association

PPCM: Peripartum Cardiomyopathy

PRKAG2: Activated Protein Kinase Gamma subunit 2

RBBB: Right Bundle Branch Block

RCM: Restrictive Cardiomyopathy

ROS: Reactive Oxygen Species

RV: Right Ventricle

Rx: Recipe (prescription)

SAM: Systolic Anterior Motion

SCD: Sudden Cardiac Death

SICM: Stress Induced Cardiomyopathy

SQTS: Short QT Syndrome

SUNDS: Sudden Unexplained Nocturnal Death Syndrome

SVT: Supraventricular Tachycardia

TFR2: Transferrin Receptor 2

TWI: T Wave Inversion

VAD: Ventricular Assist Device

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Introduction

Introduction

Progress of modern molecular biology and introduction into clinical cardiovascular medicine has considerably changed the approach to cardiomyopathies and has led to new classification schemes. While cardiomyopathies were initially defined as disorders that were idiopathic, expert panels classify cardiomyopathies now into: primary, acquired and mixed .This results from the data published by the latest American Heart Association (AHA) (2006) and European Society of Cardiology (ESC) (2008) classifications segregating cardiomyopathies into familial/genetic and nonfamilial/non-genetic. Both recommendations define a cardiomyopathy as a myocardial disorder in which the heart muscle is structurally and functionally abnormal in the absence of coronary heart disease, hypertension, valvular heart disease and congenital heart disease sufficient to cause the observed myocardial abnormality. Cardiomyopathies are therefore grouped

Introduction

into specific morphological and functional phenotypes and subclassified into familial and non-familial forms (*Parsai et al.*, 2012).

The most common clinical presentation in patients with cardiomyopathy is heart failure. The evaluation for underlying causes of heart failure includes a thorough and physical examination with baseline chemistries, including B-type natriuretic peptide (BNP) echocardiography and electrocardiography levels, (ECG). Echocardiography is key diagnostic modality for patients with suspected cardiomyopathy. The most important advance has been the use of cardiac MRI. prominent: Two aspects were the ability cardiomagnetic resonance (CMR) to detect myocardial segments 'invisible' to echocardiography (e.g., posterior septum and apex) and probably and more importantly, the ability to image myocardial scar using gadolinium enhancement (Bruder et al., 2010).