The relation between right and left ventricular functions in pediatric dilated cardiomyopathy assessed by tissue Doppler, A study in pediatric dilated cardiomyopathy patients at Cairo University Pediatric Hospital.

Thesis
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Abstract

Dilated cardiomyopathy is not purely a disease of the left ventricle. Right ventricular systolic and diastolic functions are impaired almost universally in children with dilated cardiomyopathy. Tissue Doppler imaging plays an essential role in evaluation of left and right ventricular functions. ***Aim of the work:*** we aimed to assess right ventricular systolic and diastolic functions in patients with dilated cardiomyopathy and to detect how much the right ventricular function impairment is correlated with the left ventricular function using tissue Doppler echocardiography.

Methods: cross sectional analytical study was conducted on 30 cases with dilated cardiomyopathy their ages ranging from 2 months to 12 years with median age of 2.2 years in addition to 30 age and sex matched controls. Both cases and controls were subjected to tissue Doppler echocardiography. LV dimensions were measured from M-mode and LV systolic function was calculated. Right ventricular function was assessed by Doppler tissue S', E' and A' waves; RVMPI; TAPSE and RVFAC.

Results: Right ventricular systolic and diastolic functions were significantly impaired in dilated cardiomyopathy patients. Tricuspid S', E' and TAPSE were significantly reduced ($p < 0.05$). Tricuspid S' and E' waves were decreased significantly with decreasing LVFS ($r=0.518$, $r=0.481$) respectively. ***Conclusion:*** right ventricular function is impaired in patients with dilated cardiomyopathy and correlated to the severity of left ventricular dysfunction.

Key words: dilated cardiomyopathy, Tissue Doppler echocardiography, right ventricular functions, left ventricular functions.

Abbreviations

2D	**2-Dimensional**
2DE	**2 Dimensional Echocardiography**
3D	**3-Dimensional**
3DE	**3 Dimensional Echocardiography**
A'	**Late Diastolic Wave**
ACE	**Angiotensin-Converting Enzyme**
AHA	**American Heart Association**
AMI	**Acute Myocardial Infarction**
ARVC	**Arrhythmogenic Right Ventricular Cardiomyopathy**
ARVC/D	**Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia**
AS	**Aortic Stenosis**
AV	**Atrioventricular**
BSA	**Body Surface Area**
CAD	**Coronary Artery Disease**
CHF	**Congestive Heart Failure**
CMP	**Cardiomyopathy**
CMR	**Cardiac Magnetic Resonance**
DCM	**Dilated Cardiomyopathy**
DS	**Disc Summation**
DTI	**Doppler Tissue Imaging**
E'	**Early Diastolic Wave**
E/E'	**E' Corrected For The TV Early Filling Velocity**
Ea	**Early Myocardial Relaxation Velocity**
ECG	**Electrocardiogram**
EDV	**End-Diastolic Volume**
EF	**Ejection Fraction**
Ei	**Eccentricity Index**

ESV	**End-Systolic Volume**
ET	**Ejection Time**
FS	**Fractional Shortening**
HCM	**Hypertrophic Cardiomyopathy**
ICT	**Isovolumic Contraction Time**
IRT	**Isovolumic Relaxation Time**
IVCV	**Isovolumic Myocardial Contraction Velocity**
IVRT-PW	**Isovolumetric Relaxation Time By Pulsed Wave**
IVRT-TD	**Isovolumetric Relaxation Time By Tissue Doppler**
IVRV	**Isovolumic Myocardial Relaxation Velocity**
LBBB	**Left Bundle Branch Block**
LV	**Left Ventricular**
LVEDD	**Left Ventricular End-Diastolic Dimension**
LVEDV	**Left Ventricular End-Diastolic Volume**
LVEF	**Left Ventricular Ejection Fraction**
LVESD	**Left Ventricular End-Systolic Dimension**
LVESV	**Left Ventricular End-Systolic Volume**
LVFP	**LV Filling Pressure**
LVH	**Left Ventricular Hypertrophy**
LVNC	**Left Ventricular Non-Compaction Cardiomyopathy**
MPI	**Myocardial Performance Index**
MRI	**Magnetic Resonance Imaging**
NYHA	**New York Heart Association**
PV	**Pulmonary Valve**
PVR	**Pulmonary Vascular Resistance**
PW	**Pulsed Wave**
RCM	**Restrictive Cardiomyopathy**
RV	**Right Ventricular**
RVD	**Right Ventricular Dysfunction**

RVD	**Right Ventricular Diastolic Area**
RVFAC	**RV Fractional Area Change**
RVMPI	**Right Ventricular Myocardial Performance Index**
RVOT	**RV Outflow Tract**
RVS	**Right Ventricular Systolic Area**
S'	**Peak Systolic Wave**
Sa	**Systolic Myocardial Velocity**
SD	**Standard Deviation**
SI	**Strain Imaging**
STE	**Speckle Tracking Echocardiography**
TAM	**Tricuspid Annular Motion**
TAPSE	**Tricuspid Annular Plane Systolic Excursion**
TDI	**Tissue Doppler Imaging**
TV	**Tricuspid Valve**
TVI	**Tissue Velocity Imaging**
US	**United States**
WHO	**World Health Organization**

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Introduction

Cardiomyopathy is a genetically and clinically heterogeneous disease of the myocardium that causes systolic and/or diastolic dysfunction. In affected individuals, pediatric cardiomyopathy has severe consequences with up to 40% of children progressing to death or transplant within five years of diagnosis ***(Colan et al., 2007)***.

Cardiomyopathy can be classified into five clinical phenotypes: hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), restrictive cardiomyopathy (RCM), left ventricular non-compaction cardiomyopathy (LVNC) and arrhythmogenic right ventricular cardiomyopathy (ARVC) ***(Kindel et al., 2012)***.

Dilated cardiomyopathy is the most common form of cardiomyopathy in children and its idiopathic type constitutes the majority of children with this disease. The incidence of dilated cardiomyopathy in children is 0.57 in 100,000 ***(Towbin et al., 2006)***.

In children, idiopathic myocarditis and neuromuscular diseases are the most common etiologies of dilated cardiomyopathy, and generally occur during the first year of life ***(Hulot et al., 2004)***.

The prevalence of right ventricular dysfunction in idiopathic dilated cardiomyopathy is incompletely studied in children. Furthermore, right ventricular function may signal worse outcomes ***(Groner et al., 2012)***.

Although cardiomyopathies may be asymptomatic in the early stages, most symptoms are typical of those seen in any type of heart failure, whether systolic (reduced ejection fraction) or diastolic (preserved ejection fraction) (***Wexler et al., 2009)***.

Echocardiography plays an essential role in evaluation of left and right ventricular functions. Over the past decade, the diagnostic armamentarium was fortified by tissue Doppler imaging (TDI) ***(Nagueh, 2008).***

Dilated cardiomyopathy is not purely a disease of the left ventricle. Right ventricular systolic and diastolic functions are impaired almost universally in children with idiopathic dilated cardiomyopathy. A number of studies have examined left ventricular systolic function and size in the setting of idiopathic dilated cardiomyopathy; nevertheless, the right ventricle remains difficult to interpret secondary to its unique morphology ***(Groner et al., 2012)***.

Recent published guidelines suggest an important role of tissue Doppler for right ventricular functional assessments in all patients ***(Jurcut et al., 2010)***.

The most important advantage of TDI in assessing right ventricular function is that measurement is independent of geometric assumptions and endocardial border tracing, it also minimizes the effect of preload and afterload on measurement ***(Wong et al., 2006)*** because this new modality provides quantitave measure of regional function, it may be more sensitive for detecting subclinical right ventricular abnormalities ***(Coghlan and Davar, 2008)***.

Aim of the work

The aim of our work is to assess right ventricular systolic and diastolic function in dilated cardiomyopathy using tissue Doppler echocardiography and to detect how much the right ventricular function impairment is correlated with the left ventricular function using tissue Doppler echocardiography.

Overview of Dilated Cardiomyopathy

Introduction:

Dilated cardiomyopathy (DCM) is a myocardial disorder characterized by a dilated left ventricular (LV) chamber and systolic dysfunction that commonly results in congestive heart failure (CHF) ***(Maron et al., 2006)***. In some cases, right ventricular dysfunction (RVD) is also noted and may add to the clinical severity of disease ***(Harmon et al., 2005)***.

Cardiomyopathy (CMP) is a common cause of heart failure in children and the most common indication for heart transplantation in children older than 1 year ***(Wilkinson et al., 2010)***.

According to the United States (US) Pediatric Cardiomyopathy Registry, the annual incidence of CMP was 1.13 per 100,000 children younger than 18 years ***(Lipshultz et al., 2003)*** with DCM as the most common (58%), followed by HCM (30%). There were relatively few cases of RCM (5%) and arrhythmogenic right ventricular cardiomyopathy (ARVC) (5%).

The incidence of CMP in children was 1.24 per 100,000 children younger than 10 years in Australia ***(Nugent et al., 2003)***.

The incidence of pure DCM in childhood in two regions in the Middle East area (Kuwait and Egypt) was 0.07 cases/ 100,000/year. The incidence was higher in males than in females (60 and 40% respectively) ***(Elkilany et al., 2008)***.

DCM is the most common form of CMP worldwide and has many causes. In 30% to 48% of patients, DCM is genetically inherited. Moreover, inflammatory disorders such as myocarditis, or toxic agents