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Original Article

Heart Stiffening in Pediatric Dilated Cardiomyopathy: Causes of Severity

Noor Mohammad Noori, MD[1], Alireza Teimouri, M. Phil, PhD[2*]

[1]*Department of Pediatrics, School of Medicine, Children and Adolescents Health Research Center, Research Institute of Cellular and Molecular Sciences in Infectious Diseases, Ali Ibne Abitaleb Hospital, Zahedan University of Medical Sciences, Zahedan, Iran.*
[2]*Children and Adolescents Health Research Center, Research Institute of Cellular and Molecular Sciences in Infectious Diseases, Ali Ibne Abitaleb Hospital, Zahedan University of Medical Sciences, Zahedan, Iran.*

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Abstract

Background: *Cardiomyopathy, characterized by heart stiffness, can lead to heart failure. This study aimed to investigate aortic stiffness in children with dilated cardiomyopathy (DCM) to better understand its contribution to disease severity.*

Methods: *This case-control study compared 48 children with DCM with 96 healthy children over a 10-year period starting in 2011. Aortic strain, aortic stiffness index, aortic distensibility, and pressure strain elastic modulus were measured. These parameters, along with several echocardiographic measures, were compared between the DCM and control groups. Statistical analyses were performed using SPSS 18, with a significance threshold set at a P value below 0.05.*

Results: *The participants included 57.6% boys, with 58.3% in the DCM group and 57.35% in the control group (χ^2=0.014, P=0.905). The age range was 2 to 18 years, with mean ages of 11.08±4.63 years for the DCM group and 10.77±2.82 years for the control group (P=0.691). Significant differences between groups were observed in aortic distensibility (P=0.004), aortic stiffness β index (P=0.001), and pressure strain elastic modulus (P=0.004). Post-treatment analyses based on ejection fraction and fractional shortening cutoffs indicated no changes in elasticity parameters except for the aortic stiffness β index, which varied according to the Ross classification.*

Conclusion: *Children with DCM exhibited reduced aortic strain and aortic distensibility, as well as elevated aortic stiffness β index and pressure strain elastic modulus.*

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Introduction

Dilated cardiomyopathy (DCM) is one of the most common types of cardiomyopathy (CM) and is associated with cardiac dysfunction, making it a frequent cause of end-stage heart failure (HF) in children.[1,2] Understanding CM is crucial as it is a leading cause of HF, described as a disorder of the heart muscle.[3] Recent findings reveal that DCM has a 9-year survival rate of 69.8%, hypertrophic CM has a survival rate of 93%, restrictive CM has a survival

[*]**Corresponding Author:** ***Alireza Teimouri,*** *Assistant Professor of Sociology, Children and Adolescents Health Research Center, Research Institute of Cellular and Molecular Sciences in Infectious Diseases, Ali Ibne Abitaleb Hospital, Khalij Fars Highway, Zahedan, Sistan and Baluchestan, Iran. 9816743111. Tel: +98 54 33295611. E-mail: alirezateimouri260@gmail.com.*

rate of 47.2%, and other types of CM have a survival rate of 42.0%.[4,5]

Research indicates that various pathological conditions, such as viral infections, drug abuse, toxins, and autoimmune disorders, can result in left ventricular (LV) dilation and dysfunction. Approximately half of the cases are idiopathic, with unknown causes. Notably, most patients with DCM do not present early clinical symptoms, and some are asymptomatic.[5] Nonetheless, the onset of DCM symptoms can be irregular, causing symptoms to vary among children depending on age, type, and severity of CM.[2,6]

The long-term prognosis of pediatric CM varies depending on the type and stage of the disease. Infants and children with DCM typically present with signs of congestive heart disorders, such as tachypnea, labored breathing, poor appetite, and slow weight gain.[6]

Two types of HF include systolic failure (muscle weakness) and diastolic failure (stiffness preventing normal heart relaxation).[7] The elastic properties of the aorta are essential for optimal ventricular-vascular coupling, as arterial stiffening in the peripheral vascular system can pose risks.[7] Echocardiography is the gold standard for diagnosing and classifying CM, as well as determining the extent of heart muscle dysfunction, with diagnostic criteria such as fraction shortening of less than 25% and ejection fraction of less than 55% in DCM.[8,9]

In DCM, the heart's main pumping chamber, the LV, becomes enlarged. As the chamber expands, its thick, muscular wall stretches, becoming thinner and weaker. This impairs the heart's ability to pump sufficient oxygen-rich blood to the rest of the body. DCM is the most common type of restrictive CM.[6–9]

This study aimed to investigate potential changes in heart stiffness associated with DCM and the factors contributing to its severity in pediatric patients, in light of the aforementioned literature and information.

Methods

This case-control study included 144 children, with 48 having DCM and 96 being healthy controls. The participants were aged 2 to 18 years old. The study was conducted at the cardiac centers of Zahedan University of Medical Sciences, Iran, over a 10-year period starting in 2011. Patients were diagnosed with DCM via echocardiography, while healthy children were randomly selected from those who had been referred to the cardiac center for annual checkups during the data collection period. All patients with complete profiles, considering exclusion criteria, were included in the study.

Various treatments for CM include lifestyle changes, medications, surgically implanted devices, ablation procedures (removing excess heart tissue to reduce thickening), and heart transplantation for severely damaged hearts. In this study, all patients received medical care. Ejection fraction (EF), fractional shortening (FS), and Ross factors were measured and recorded 1 year after discontinuing medication to evaluate the effectiveness of the treatment.

Patients and controls with endocrine and metabolic disorders, arrhythmias, various forms of CM, and complete HF were excluded from the study. Individuals with diseases that significantly affect cardiac stiffness, such as celiac disease, obesity, hypertension, diabetes, HF, arrhythmias, isolated valve disease, and congenital heart disease, were also excluded to determine the net effect of DCM on aortic stiffness.

The study protocol was approved by the university's ethics committee and assigned the code IR.ZAUMS.REC.1400.095. Informed consent was obtained from the parents or guardians of the patients who participated in the study.

The children underwent physical examination, medical history review, chest X-ray, and echocardiography using 3 and 8 MHz transducers (made in Italy) from the My Lab 60 system. Measurements were repeated for 3 cycles to enhance accuracy, and the average was calculated. Standard echocardiographic parameters used in this study consisted of aortic diastolic diameter (AOD), aortic systolic diameter (AOS), left ventricular end-diastolic dimension (LVDD), posterior diastolic wall dimension (PWD), posterior systolic wall dimension (PWS), systolic ventricular septal dimension (IVSS), relative wall thickness (RWT), LV diastolic diameter, EF, FS, and left ventricular mass (LVM), which were measured using standard left-sided echocardiography based on 3 cardiac cycles. The following formulas were utilized to calculate LVM and LVMI: LVM (g)=0.8 (1.04 (LVDD + PWD + IVSD)^3 - LVDD^3) + 0.6 and LVMI (g/m^2) = LVM / 2.7 (g/m^2).

The aortic diameter was calculated using the distance between the inner edges of the anterior and posterior walls of the aorta in systole and diastole. AOS was recorded when the aortic wall was fully open. The QRS peak on the ECG was simultaneously recorded with AOD (Figure 1). After at least 5 minutes of supine rest, blood pressure (BP) was measured from the brachial artery using a sphygmomanometer. Three measurements were taken at least 2 minutes apart, and the average was recorded. Korotkoff phases I and V were utilized for systolic and diastolic BP, respectively. The following parameters of aortic elasticity were calculated: aortic strain=100 *(AOS-AOD)/ AOD, aortic stiffness index=LN (SBP/DBP)/((AOS-AOD)/AOD), aortic distensibility (cm^2/dyn/10-6) =2× (AOS-AOD)/[(SBP–DBP) × diastolic diameter], pressure strain elastic modulus=(SBP–DBP)/((AOS–AOD)/AOD).

The modified Ross classification was used to stratify HF. The patients were divided into 4 categories based on this classification. Group I included patients without limitations

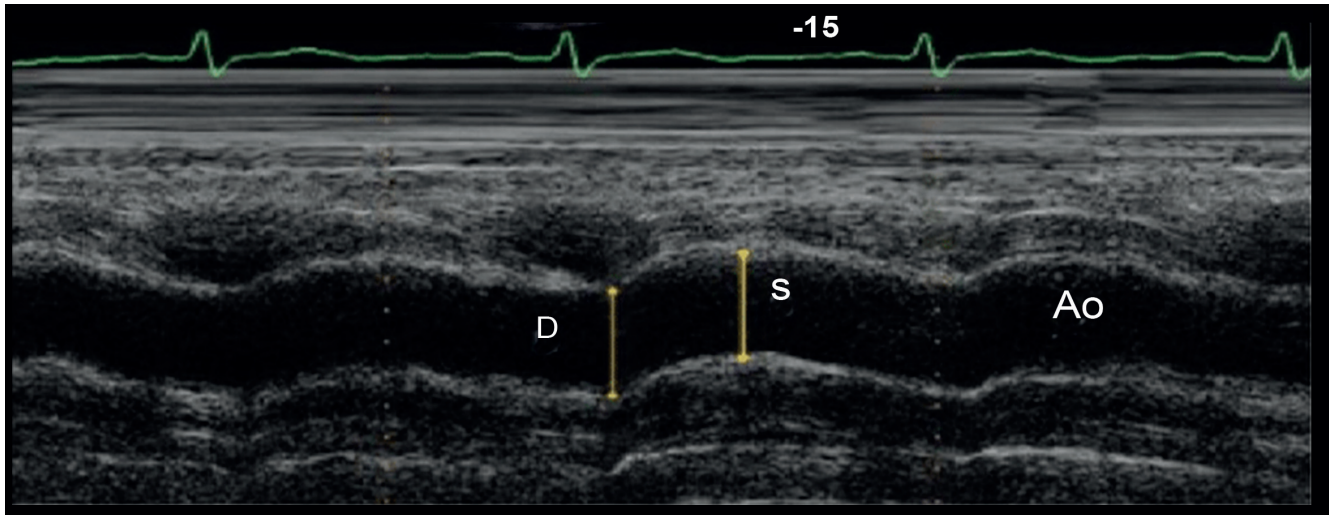

Figure 1. The image shows systolic (S) and diastolic (D) diameters of the ascending aorta in the M mode tracing at a level 3 cm above the aortic valve.

or symptoms. Group II consisted of patients with mild tachypnea or diaphoresis and dyspnea upon exertion in older children, with no growth failure. Group III comprised patients with marked tachypnea or diaphoresis during feedings or exertion and prolonged feeding times. Group IV consisted of patients who were symptomatic at rest, exhibiting tachypnea, retractions, grunting, or diaphoresis, and experienced growth failure due to congestive HF. An Iranian-made RASA Mark scale was utilized to measure weight in children, while height was assessed using a scale ruler in the standing position.

Data analysis was performed using the Statistical Package for the Social Sciences, version 18 (SPSS, Chicago, USA). The Kolmogorov–Smirnov test was employed to assess the normal distribution of data. Associations were examined using the independent samples t-test for normally distributed numerical data and the Mann-Whitney U-test for non-normally distributed data. For Ross classification, we employed ANOVA or a related nonparametric test, as appropriate. Statistical significance was set at a P value below 0.05.

Results

Forty-eight children with DCM and 96 healthy children participated in the study. The distribution of boys and girls was comparable between the groups, with boys accounting for 57.6% of all the participants: 58.3% in the DCM group and 57.35% in the control group (χ^2=0.014, *P*=0.905). Weight and AOS were normally distributed among all the participants (*P*>0.05). In children with DCM, AOD, IVSS, LVMI, and RWT showed similar trends (*P*>0.05). Table 1 shows that the groups of participants were matched by age (*P*>0.05) but significantly different in LVDD (*P*<0.001), LVDS (*P*<0.001), EF (*P*<0.001), FS (*P*<0.001), LVMI (*P*<0.001), AS (*P*=0.014), AD (*P*=0.004), ASBI (*P*=0.001), and pressure strain elastic modulus (*P*=0.004). From these variables, EF, FS, AS, and AD were lower in the DCM groups. Table 2 demonstrates an increasing trend in EF and FS after treatment compared with before treatment (*P*<0.001). The table also presents the changes in Ross classification after treatment compared with before treatment. Table 3 and Table 4 show that none of the stiffening parameters changed with the severity of FS and EF. Nevertheless, Table 5 shows that only ASBI (*P*=0.048) changed significantly concerning the Ross classification after treatment.

Discussion

Previous studies have indicated that aortic stiffness is higher in cases of diastolic HF and vascular failure.[10,11] DCM is a severe condition that weakens the heart muscle, impeding its ability to pump blood effectively. Aortic stiffening results in elevated SBP, heart rate, and myocardial oxygen consumption, as well as decreased DBP and coronary perfusion slope. Proximal rupture and periodic fluid volume overload lead to fibrosis, increasing cardiovascular responsiveness and wall pressure. Over time, damaged tissue is replaced by tendinous scar tissue, which hardens the heart and further accelerates the progression to cardiovascular collapse.[12]

The present review demonstrated that DBP, EF, FS, RWT, aortic stiffness, and AD were lower, while LVDD, LVDS, IVSD, PWD, PWS, LVMI, aortic stiffness β index, and PSEM were higher in children with DCM than in controls.

Patrianakos et al[10] conducted a study to investigate changes in cardiac functions and aortic stiffness in DCM patients compared with controls. They observed that LVDD, LVMI, and aortic stiffness increased in DCM patients after

Table 1. Comparisons of the variables in the study between the DCM (48 individuals) and healthy children (96 individuals) groups

Variables	Groups	Mean	SD	Critical Value	P	Variables	Mean	SD	Critical Value	P
Age (y)	Children with DCM	11.08	4.63	2210.50**	0.691	IVSD (cm)	0.74	0.13	1442.50**	<0.001
	Controls	10.77	2.82				0.66	0.11		
Weight (kg)	Children with DCM	32.71	18.18	-4.40*	<0.001	IVSS (cm)	0.85	0.17	2201.00**	0.662
	Controls	43.77	11.81				0.82	0.15		
Height (cm)	Children with DCM	132.17	23.42	1169.50**	<0.001	PWD (cm)	0.43	0.07	886.50**	<0.001
	Controls	153.24	12.78				0.35	0.05		
LVDD (cm)	Children with DCM	4.35	0.86	1525.00**	0.001	PWS (cm)	0.44	0.07	832.00**	<0.001
	Controls	3.81	0.59				0.36	0.05		
LVDS (cm)	Children with DCM	4.13	6.98	449.00**	<0.001	LVM (g)	78.6	36.34	1158.00**	<0.001
	Controls	2.08	0.31				51.44	17.65		
AOS (cm)	Children with DCM	2.07	0.39	-0.31*	0.754	RWT (-)	0.2	0.03	1691.50**	0.009
	Controls	2.09	0.3				0.2	0.12		
AOD (cm)	Children with DCM	1.88	0.39	2239.50**	0.784	AS (%)	10.7	6.94	1725.00**	0.014
	Controls	1.85	0.31				13.32	6.82		
SBP (mm Hg)	Children with DCM	98.58	13.26	2233.50**	0.762	AD ($cm^2dyn^{-1}\times10^{-6}$)	0.01	0.00	1616.00**	0.004
	Controls	98.4	10.17				0.01	0.00		
DBP (mm Hg)	Children with DCM	58.94	10.29	1818.00**	0.029	AsβI (-)	12.92	26.66	1494.50**	0.001
	Controls	61.84	9.28				4.32	2.05		
EF (%)	Children with DCM	51.54	19.67	208.00**	<0.001	PSEM (kPa)	10.91	24.72	1616.00**	0.004
	Controls	77.49	4.86				3.38	1.61		
FS (%)	Children with DCM	25.96	10.41	31.50*	<0.001					
	Controls	45.8	4.63							

*t-test
**Mann–Whitney U-test
LVDD, Left ventricular diastolic dimension; LVDS, Left ventricular systolic dimension; AOS, Aortic diameter in systole; AOD, Aortic diameter in diastole; SBP, Systolic blood pressure; DBP, Diastolic blood pressure; EF, Ejection fraction; FS, Fractional shortening; IVSD, Interventricular septal thickness in diastole; IVSS, Interventricular septal thickness in systole; PWD, Posterior diastolic wall dimension; PWS, Posterior systolic wall dimension; LWM, Left ventricular mass; RWT, Relative wall thickness; AS, Aortic strain; AD, Aortic distensibility; ASβI, Aortic stiffness β index Indication; PSEM, Pressure strain elastic modulus

Table 2. Ejection fraction, fractional shortening, and modified Ross classification changes after treatment

Variables Treatment	Mean	SD	Test Value	P
Fractional shortening (%)			-7.32	<0.001
Before	16.4	4.11		
After	25.96	10.41		
Ejection fraction (%)			-7.49	<0.001
Before	32.85	8.2		
After	51.54	19.67		
Ross classification			14.4	<0.001
Before	2.79	0.41		
After	1.85	1.15		

Table 3. Comparisons of the study variables in DCM children with normal (n=31) and abnormal (n=17) FS (normal status; FS>25)

Variables	FS Status	Mean	SD	Critical Value	P	Variables	Mean	SD	Critical Value	P
Age (y)	Abnormal	11.21	4.22	260.00**	0.940	IVSD (cm)	0.74	0.12	257.00**	0.888
	Normal	11.02	4.90				0.74	0.13		
Weight (kg)	Abnormal	26.94	11.69	206.50**	0.218	IVSS (cm)	0.83	0.14	-0.50*	0.617
	Normal	35.87	20.39				0.86	0.19		
Height (cm)	Abnormal	129.47	19.55	238.00**	0.582	PWD (cm)	0.44	0.05	209.50**	0.239
	Normal	133.65	25.47				0.42	0.07		
LVDD (cm)	Abnormal	5.14	0.85	57.50*	<0.001	PWS (cm)	0.46	0.06	203.00**	0.188
	Normal	3.91	0.48				0.43	0.07		
LVDS (cm)	Abnormal	6.97	11.38	22.00*	<0.001	LVM (g)	105.80	39.40	4.58*	<0.001
	Normal	2.58	0.34				63.69	24.33		
AOS (cm)	Abnormal	2.10	0.48	249.00**	0.754	RWT (-)	.18	0.03	-4.64*	<0.001
	Normal	2.06	0.35				.22	0.03		
AOD (cm)	Abnormal	1.87	0.41	-0.11*	0.916	AS (%)	12.20	7.49	206.50**	0.219
	Normal	1.89	0.39				9.87	6.61		
SBP (mm Hg)	Abnormal	92.06	8.67	181.50**	0.072	AD $cm^2dyn^{-1}\times10^{-6}$	.01	0.00	202.00**	0.185
	Normal	102.16	14.08				.01	0.00		
DBP (mm Hg)	Abnormal	54.35	9.23	163.50**	0.028	ASβI (-)	9.69	17.47	235.00**	0.539
	Normal	61.45	10.10				14.69	30.69		
EF (%)	Abnormal	26.47	8.62	17.00*	<0.001	PSEM (kPa)	7.20	13.77	202.00**	0.185
	Normal	64.32	7.39				12.94	29.06		

* t-test

** Mann–Whitney U-test

FS, Fractional shortening; LVDD, Left ventricular diastolic dimension; LVDS, Left ventricular systolic dimension; AOS, Aortic diameter in systole; AOD, Aortic diameter in diastole; SBP, Systolic blood pressure; DBP, Diastolic blood pressure; EF, Ejection fraction; IVSD, Interventricular septal thickness in diastole; IVSS, Interventricular septal thickness in systole; PWD, Posterior diastolic wall dimension; PWS, Posterior systolic wall dimension; LWM, Left ventricular mass; RWT, Relative wall thickness; AS, Aortic strain; AD, Aortic distensibility; ASβI, Aortic stiffness β index Indication; PSEM, Pressure strain elastic modulus

Table 4. Comparisons of the study variables in DCM children with normal (n=26) and abnormal (n=22) EF (normal status; EF>55)

Variables	EF Status	Mean	SD	Critical Value	P	Variables	Mean	SD	Critical Value	P
Age (y)	Abnormal	10.07	4.29	210.5**	0.116	IVSD (cm)	0.72	0.12	233.5**	0.276
	Normal	11.94	4.81				0.76	0.13		
Weight (kg)	Abnormal	24.50	11.23	153.5**	0.006	IVSS (cm)	0.80	0.14	-1.94*	0.059
	Normal	39.65	20.15				0.89	0.19		
Height (cm)	Abnormal	124.55	19.61	191.5**	0.050	PWD (cm)	0.43	0.05	273.5**	0.794
	Normal	138.62	24.77				0.43	0.08		
LVDD (cm)	Abnormal	4.80	0.99	146**	0.004	PWS (cm)	0.44	0.07	276**	0.835
	Normal	3.97	0.50				0.44	0.08		
LVDS (cm)	Abnormal	6.01	10.09	51.5*	<0.001	LVM (g)	92.17	43.02	2.51*	0.016
	Normal	2.54	0.36				67.12	25.07		
AOS (cm)	Abnormal	2.01	0.46	233**	0.272	RWT (-)	0.18	0.03	-4.285*	<0.001
	Normal	2.12	0.33				0.22	0.03		
AOD (cm)	Abnormal	1.80	0.39	-1.33*	0.189	AS (%)	11.63	6.76	228.5**	0.234
	Normal	1.95	0.38				9.91	7.13		
SBP (mm Hg)	Abnormal	91.36	7.74	143.5**	0.003	AD $cm^2dyn^{-1}\times10^{-6}$	0.01	0.00	221**	0.179
	Normal	104.69	13.99				0.01	0.00		
DBP (mm Hg)	Abnormal	53.36	8.42	114.5**	<0.001	ASβI (-)	8.98	15.35	265**	0.664
	Normal	63.65	9.44				16.25	33.37		
FS (%)	Abnormal	16.18	7.18	13.52*	<0.001	PSEM (kPa)	6.57	12.10	221**	0.179
	Normal	33.96	3.18				14.57	31.56		

*t-test,

**Mann–Whitney U-test

LVDD, Left ventricular diastolic dimension; LVDS, Left ventricular systolic dimension; AOS, Aortic diameter in systole; AOD, Aortic diameter in diastole; SBP, Systolic blood pressure; DBP, Diastolic blood pressure; EF, Ejection fraction; FS, Fractional shortening; IVSD, Interventricular septal thickness in diastole; IVSS, Interventricular septal thickness in systole; PWD, Posterior diastolic wall dimension; PWS, Posterior systolic wall dimension; LWM, Left ventricular mass; RWT, Relative wall thickness; AS, Aortic strain; AD, Aortic distensibility; ASβI, Aortic stiffness β index Indication; PSEM, Pressure strain elastic modulus

Table 5. Comparisons of the variables in DCM children classified based on the modified Ross classification using the Kruskal-Wallis test

Variables	ROSS Classification	Numbers of Patients	Mean	SD	Critical Value	P	Variables	Mean	SD	Critical Value	P
LVDD (cm)	First class	29	3.92	0.5	0.4	0.525	PWD (cm)	0.42	0.08	0.85	0.356
	Second class	3	3.97	0.21				0.4	0.05		
	Third class	10	5.11	0.94				0.42	0.04		
	Fourth class	6	5.35	0.71				0.48	0.05		
LVDS (cm)	First class	29	2.55	0.34	0.01	0.939	PWS (cm)	0.43	0.07	0.43	0.513
	Second class	3	3.03	0.23				0.4	0.05		
	Third class	10	8.86	14.83				0.44	0.07		
	Fourth class	6	4.43	0.75				0.48	0.06		
AOS (cm)	First class	29	2.09	0.33	0.07	0.789	LVM(g)	64.5	24.97	7.65	<0.001
	Second class	3	1.51	0.05				61.72	16.99		
	Third class	10	1.95	0.49				99.96	39.88		
	Fourth class	6	2.46	0.14				119.61	41.08		
AOD (cm)	First class	29	1.92	0.37	5.66	0.002	RWT (-)	0.22	0.03	8.42	<0.001
	Second class	3	1.37	0.05				0.2	0.01		
	Third class	10	1.74	0.42				0.17	0.03		
	Fourth class	6	2.17	0.2				0.18	0.02		
SBP (mm/ Hg)	First class	29	103.17	13.98	1.6	0.205	AS (%)	9.79	6.83	2.56	0.11
	Second class	3	90	5				9.96	1.87		
	Third class	10	92	10.85				11.66	7.4		
	Fourth class	6	91.67	5.16				13.84	8.56		
DBP (mm Hg)	First class	29	62.24	9.87	3.22	0.073	AD $cm^2dyn^{-1}\times10^{-6}$	0.01	0	2.17	0.141
	Second class	3	51.67	5.77				0.01	0		
	Third class	10	55.4	10.77				0.01	0.01		
	Fourth class	6	52.5	7.58				0.01	0		
EF (%)	First class	29	65.1	6.96	0.29	0.593	ASβI (-)	15.34	31.66	3.92	0.048
	Second class	3	52	2.65				5.77	1.71		
	Third class	10	31	11.25				12.57	22.71		
	Fourth class	6	20	5.48				5.34	2.76		
FS (%)	First class	29	32.97	4.56	0.21	0.647	PSEM (kPa)	13.59	29.97	2.17	0.141
	Second class	3	26.33	1.53				3.99	1.25		
	Third class	10	15.2	6.07				9.44	17.91		
	Fourth class	6	9.83	2.4				3.82	2.2		
IVSS (cm)	First class	29	0.87	0.19	0.7	0.557	IVSD (cm)	0.75	0.13	1.36	0.715
	Second class	3	0.76	0.12				0.73	0.13		
	Third class	10	0.8	0.14				0.71	0.13		
	Fourth class	6	0.88	0.15				0.77	0.12		

LVDD, Left ventricular diastolic dimension; LVDS, Left ventricular systolic dimension; AOS, Aortic diameter in systole; AOD, Aortic diameter in diastole; SBP, Systolic blood pressure; DBP, Diastolic blood pressure; EF, Ejection fraction; FS, Fractional shortening; IVSD, Interventricular septal thickness in diastole; IVSS, Interventricular septal thickness in systole; PWD, Posterior diastolic wall dimension; PWS, Posterior systolic wall dimension; LWM, Left ventricular mass; RWT, Relative wall thickness; AS, Aortic strain; AD, Aortic distensibility; ASβI, Aortic stiffness β index Indication; PSEM, Pressure strain elastic modulus

initial treatment, while FS and EF decreased compared to controls.

In another study by Patrianakos et al,[13] patients with DCM exhibited lower EF and higher pulse wave velocity (PWV). The authors concluded that there was a correlation between LV dysfunction and increased aortic stiffness in patients with HF caused by DCM.

Consistent with previous research, Puntmann et al[14] found a connection between aortic stiffness and myocardial remodeling, such as increased LVM. In contrast, Kaolawanich and Boonyasirinant[15] did not observe a significant difference in LVM and left atrial diameter. A recent systematic meta-analysis revealed a significant correlation between arterial stiffness and diastolic dysfunction in DCM patients, highlighting diastolic dysfunction as a primary cause of HF in patients suffering from heart failure with preserved ejection fraction (HFpEF).[16]

Seeland et al[17] and Alba et al[18] demonstrated a significant association between the prevalence of diastolic dysfunction and abnormal PWV, suggesting a stronger connection between asymptomatic diastolic dysfunction and arterial stiffness. Although dilated vessel stiffness is associated with asymptomatic diastolic dysfunction, the progression from asymptomatic diastolic dysfunction to HFpEF-induced cardiovascular collapse may be attributed to the increased stiffness of the dilated proximal aorta, as evidenced by the substantial difference in aortic dilatation between patients with DCM and those without HFpEF. In other words, the progression from diastolic dysfunction to HFpEF might be driven by the increased stiffness of the ascending aorta rather than the peripheral vasculature.[19]

In conclusion, patients with HFpEF exhibit arterial stiffening that exceeds age-related changes.[20] The present study showed that LVDD, LVDS, SBP, DBP, FS, LVMI, and RWT changed in patients based on their EF status. However, none of the stiffness parameters changed significantly in relation to EF or fractional shortening. Additionally, LVDD, LVDS, DBP, EF, LVMI, and RWT changed in patients based on their fractional shortening status.

AOD, LVMI, RWT, and aortic stiffness β index changed significantly according to the Ross classification, with the latter being the only stiffness parameter to exhibit a significant change based on the classification. Our findings can be explained by the following pathophysiologic model: As the aorta loses elasticity due to arteriosclerosis, aging, and hypertension, parallel structural changes such as hypertrophy and fibrosis occur in the heart.

In a study by Karagodin et al,[19] some heart stiffness parameters, such as LV end-diastolic volume index, diastolic wall strain, LV end-systolic volume, and RWT, were similar between the studied groups. The relative risk of cardiovascular death or HF hospitalization due to DCM is higher in patients with HFpEF than in those with reduced EF.[16,21] DCM has been identified as an independent predictor of morbidity and mortality because it impairs myocardial relaxation, leading to increased filling pressure and eventual congestive HF due to elevated afterload.[18]

In HFpEF, the interaction between ventricular and arterial stiffness significantly impacts cardiac function and determines the onset of symptoms. This is because coronary blood flow is more reliant on systolic pressure, and there is increased ischemia for a given drop in SBP.[22]

Measures of aortic stiffness, pressure, and flow pulsatility have been identified as potential contributors to cardiovascular disease, dementia, and kidney disease.[23] An excessively pulsatile load on the heart is associated with higher aortic stiffness and greater pressure and flow pulsatility, both of which increase LVM and reduce global longitudinal strain. Excessive stiffness and pulsatility are also linked to microvascular lesions in high-flow organs. Additionally, wave reflections originating in peripheral arteries and returning to the proximal aorta during mid-to-late systole significantly contribute to LV afterload in patients with HF.

Patrianakos et al[13] classified their patients based on HFpEF and found that the more severe group exhibited greater arterial stiffening. Traditional measures between the patient groups were comparable regarding disease severity, which is consistent with our findings that showed a slight difference. The development of symptomatic HF occurs when the LV is chronically exposed to elevated DBP over time.[18]

According to Chirinos et al,[24] an increased risk of cardiovascular events and the emergence of HF symptoms are linked to increased arterial wave reflection during mid-to-late systole, likely due to structural and functional alterations resulting from heightened wave activity and LV enlargement. Over time, this leads to elevated LV afterload, greater aortic stiffness, and chronically increased pressure in the ascending aorta, potentially resulting in HFpEF.

Considering the substantial impact of pulsatile loading on aortic stiffness, it is important to account for the proximal aortic stiffness when evaluating the aorta's connection to the LV myocardium. The observed improvements in cardiac parameters after treatment are promising, but the limitations of the study include the small sample size and insufficient follow-up time post-treatment.

Conclusion

This study revealed a decrease in aortic stiffness and aortic distensibility, alongside an increase in aortic stiffness β index and PSEM in children with DCM compared with controls. Moreover, stiffness parameters remained unaffected by DCM severity as indicated by

EF and fractional shortening. The Ross classification had a significant impact on aortic stiffness β index, the most critical stiffness index, suggesting that a stiffer proximal aorta with reduced shock absorption capacity may exacerbate LV afterload and compromise cardiovascular performance in patients with HF.

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