



Unveiling Cardiac Involvement in Juvenile Dermatomyositis Through Speckle-Tracking Echocardiography

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Abstract

Early detection of cardiac involvement in Juvenile Dermatomyositis (JDM) is difficult due to the absence of clinical signs and symptoms, with systolic dysfunction often emerging in late stages and associated with a poor prognosis. This study aimed to employ two-dimensional speckle-tracking echocardiography (STE) for subclinical assessment of left ventricular (LV) systolic failure in JDM and explore potential associations between impaired LV systolic function (LV-GLS) and disease activity. A prospective study enrolled 20 healthy volunteers and 26 JDM patients (< 18 years old) without cardiac symptoms. Clinical data were collected from medical records, and echocardiograms were conducted by a pediatric cardiologist. Our study cohort demonstrated similar age to controls ($13.5 \pm .6$ vs. 13.8 ± 4.7 ; $p=0.465$). Median illness duration at echocardiography was 5 (1.5–17.5) years, and conventional echocardiography indicated normal LV ejection fraction ($>55\%$) in all participants. However, STE revealed lowered LV GLS in JDM patients ($-22.2 \pm 4.1\%$ vs. $-26.5 \pm 5.3\%$ $p=0.022$). Pulse steroid users displayed lower GLS average values compared to non-users ($\beta=4.99$, 95% CI 1.34–8.64, $p=0.009$). Negative correlations existed between LV-GLS and age at diagnosis ($r=-0.499$; $p=0.011$), diastolic parameters (E/E' ratio) and age at diagnosis ($r=-0.469$; $p=0.018$), as well as RV global strain and age at diagnosis ($r=-0.443$; $p=0.024$). Employing STE in JDM patients facilitated the identification of preclinical cardiac dysfunction. Given JDM patients' younger age, early myocardial damage detection through STE may impact treatment decisions and long-term cardiovascular prognosis.

Keywords Juvenile dermatomyositis · Speckle tracking echocardiography · Global longitudinal strain · Cardiac dysfunction

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Introduction

Juvenile dermatomyositis (JDM) is characterized with a distinctive rash and symmetrical proximal muscle weakness. Because microvascular damage is classified as vasculopathy it may not be limited to the skin and muscle it may also comprise the cardiovascular system and gastrointestinal tract vasculature [1, 2]. Cardiovascular injury involves decreased cardiac conduction, and cutaneous inflammation predicts later cardiac systolic failure [3, 4]. Early detection of cardiac involvement in JDM is challenging due to the frequent absence of clinical signs and symptoms. Clinical manifestations of myocardial impairment usually occur in the late stages of the illness and are associated with a poor prognosis. Myocardial infarction and heart failure are the two main causes of the 5 to 17% range in cardiovascular mortality in adult patients [5]. Studying heart function increasingly involves measuring shape deformation or myocardial strain. Global longitudinal strain (GLS), global circumferential strain (GCS), and global radial strain (GRS) are measurements of the cardiac tissue shortening during systole in these three dimensions. Assessment of longitudinal systolic deformation, also known as GLS, is a more accurate method of systolic dysfunction early detection than ejection fraction (EF). Guerra et al. reported reduced GLS in the left ventricle (LV) in patients with polymyositis and dermatomyositis who still maintained unaffected LV EF (systolic function) in a recent paper [6]. However, neither disease activity nor the status of cumulative damage was shown to be correlated with LV strain in those authors' research. In a recently published another report LV GLS identified pediatric JDM patients with intact EF and early systolic myocardial impairment. While GLS impairment was connected to both disease activity and damage accumulation, GCS impairment was only connected to cumulative damage [7].

The current study used GLS by speckle tracking echocardiography to explore subclinical anomalies in LV systolic performance in JDM patients. Clinical parameters and potential relationships regarding LV GLS compromise were also investigated.

Materials and Methods

Twenty-six consecutive JDM patients were enrolled between September 2020 and December 2021 during their regular outpatient appointments to our University Hospital's Pediatric Rheumatology Department. All patients met the Bohan and Peter criteria for JMD [8]. Patients with significant acute or chronic comorbidities, such as

intercurrent infections leading to acute endothelial damage, or displaying signs of heart failure (New York Heart Association class I) [9], were excluded from the study.

Definition of Inactive Juvenile DM

Following a modified version of the Pediatric Rheumatology International Trials Organization (PRINTO) guidelines [10], clinically inactive juvenile DM was categorized based on meeting at least three out of four criteria: Creatine kinase (CK) levels below 150 units/liter, Childhood Myositis Assessment Scale (CMAS) score of 48/52 or higher, Manual Muscle Testing 8 (MMT-8) score, and Physician Global Assessment (PGA) score of 0.2 (out of a possible 10). Two senior doctors (OK and KB) independently reviewed the medical records of the patients to assess the severity of juvenile DM, resolving any discrepancies through discussion.

Healthy Controls

Healthy controls, consisting of 20 children of similar age and sex with no acute or chronic illnesses and not regularly taking any medications, were recruited from primary care clinics. All participants, both patients, and controls, had undergone prior echocardiographic examinations to rule out congenital heart problems. Our institution's ethical committee granted ethical permission for this study, and all participants and their legal representatives provided signed informed consent. The attending physician thoroughly reviewed clinical, laboratory, and treatment information from the patients' medical records. Active disease was defined as Childhood Myositis Assessment Score (CMAS) < 48 [11].

Demographic data collected included gender, age at diagnosis, current age, duration of the condition, and treatment duration. Disease onset was determined by the initial muscle or skin symptom, while the interval between disease onset and the last follow-up examination established the disease's duration.

Cardiac Imaging

Prior to the echocardiogram, each patient's weight and height were recorded to calculate their body surface area (BSA) using the Haycock formula [12]. A single pediatric cardiology specialist (RD) blinded to disease activity performed standard and 2D speckle tracking (STE) echocardiograms.

Echocardiography

The guidelines set forth by the American Society of Echocardiography for standard transthoracic echocardiography were adhered to, including M-mode, two-dimensional

imaging, and tissue Doppler examination [12]. The analyses were conducted using a Philips iE33 echocardiography machine (Philips Medical Systems) fitted with X5-1S MHz multifrequency transducer.

Conventional techniques were employed to assess both left ventricular (LV) systolic and diastolic functions at the last clinical visit. The parameters utilized for evaluating systolic function through conventional echocardiography are as follows:

Left Ventricular Systolic Function: Ejection Fraction % (EF).

Parameters for LV diastolic functions include:

Doppler Indices: E wave cm/sec, A wave cm/sec, E/A ratio

Tissue Doppler Indices: E' cm/sec, A' cm/sec

E/E' ratio (most sensitive for diastolic capillary wedge pressure)

Right ventricular systolic function was assessed using tricuspid annular plane systolic excursion (TAPSE), also referred to as tricuspid annular motion.

Traditionally, Doppler patterns of mitral inflow have been used to assess LV diastolic function during echocardiography. Trans-mitral velocities are independently and inversely related to ventricular relaxation and directly associated with left atrial pressure (preload), reflecting the pressure gradient between the left atrium and LV. Diastolic function assessment using tissue Doppler (TDI) is less dependent on load compared to traditional Doppler techniques. E' measures the speed of myocardial relaxation in the early stages of LV filling as the mitral annulus ascends. In addition to the Doppler measurements (E, A velocities, E/A ratio), assessment includes E' (early diastolic myocardial relaxation velocity) and E/E' ratio, where E' is the average value obtained through tissue Doppler at the lateral and septal annulus.

Speckle Tracking Echocardiography

Speckle tracking echocardiography (STE) is a commonly used non-invasive technology that complements conventional echocardiography by detecting subtle cardiac damage. STE operates on the principle that each segment of cardiac tissue exhibits a unique speckle pattern or pattern of grey values in the ultrasound image. Throughout the cardiac cycle, this speckle pattern is tracked to monitor myocardial mobility and quantify ventricular deformation. To assess overall longitudinal LV systolic deformation, the apical four-, three-, and two-chamber images were captured and digitally saved as two-dimensional harmonic imaging cine-loop recordings. The QLab™ software (Philips) automatically delineated myocardial wall borders, which were manually adjusted as needed to encompass the entire myocardium.

The resulting measurement, known as LV Global Longitudinal Strain (LV GLS), was calculated by summing the results from the three image windows (four-chamber, three-chamber, and two-chamber views) and presented as percentages (see Fig. 1). These deformation parameters were expressed as negative values, reflecting cardiac shortening [13, 14].

Results

Demographic and Clinical Parameters

JDM patients and controls exhibited similar ages (13.5 ± 3.6 years vs. 13.8 ± 4.7 years; $p=0.465$). The mean age at JDM diagnosis was 6.9 ± 3.7 years, and at the time of the echocardiogram, the median disease duration was 5 (1.5–17.5) years. According to medical records from outpatient clinics, neither patients nor controls displayed other heart disease risk factors such as arterial hypertension or lipidaemia. All patients had musculoskeletal and musculoscutaneous involvement and all patients were on remission. Specifics regarding the demographic and clinical parameters of the patients are outlined in Table 1.

Conventional Echocardiogram

All individuals exhibited intact LV EF (EF normal range $> 55\%$). However, patients had lower EF values compared to controls 62 ± 4.2 vs $67 \pm 3.4\%$; $p=0.001$) (Table 2). Diastolic function parameters: E/A and E/E' values were similar between patients and controls, suggesting normal LV end-diastolic pressures [13].

Speckle Tracking Echocardiography (STE)

All 26 JDM patients and controls yielded satisfactory images, and none were excluded from cardiac deformation analysis. In comparison to controls, patients demonstrated lower LV GLS, including LV GLS 4 chamber -22.7 ± 4 . vs. $-28.6\% \pm 4.51$, $p=0.001$), LV GLS 3 Chamber -20.9 ± 5.2 vs. $-27.0\% \pm 4.0$, $p=0.0001$), LV GLS 2 Chamber -23.1 ± 5.1 vs. $-25.5\% \pm 3.4$, $p=0.0001$), and LV GLS average (-22.2 ± 4.1 vs. -26.5 ± 5.3 , $p=0.02$) in patients compared to controls (Fig. 1).

Right Ventricular Functions

The right ventricular functions were within normal ranges and did not differ between patients and control subjects (TAPSE, RV frontal wall strain and RV Longitudinal strain 4 chamber).

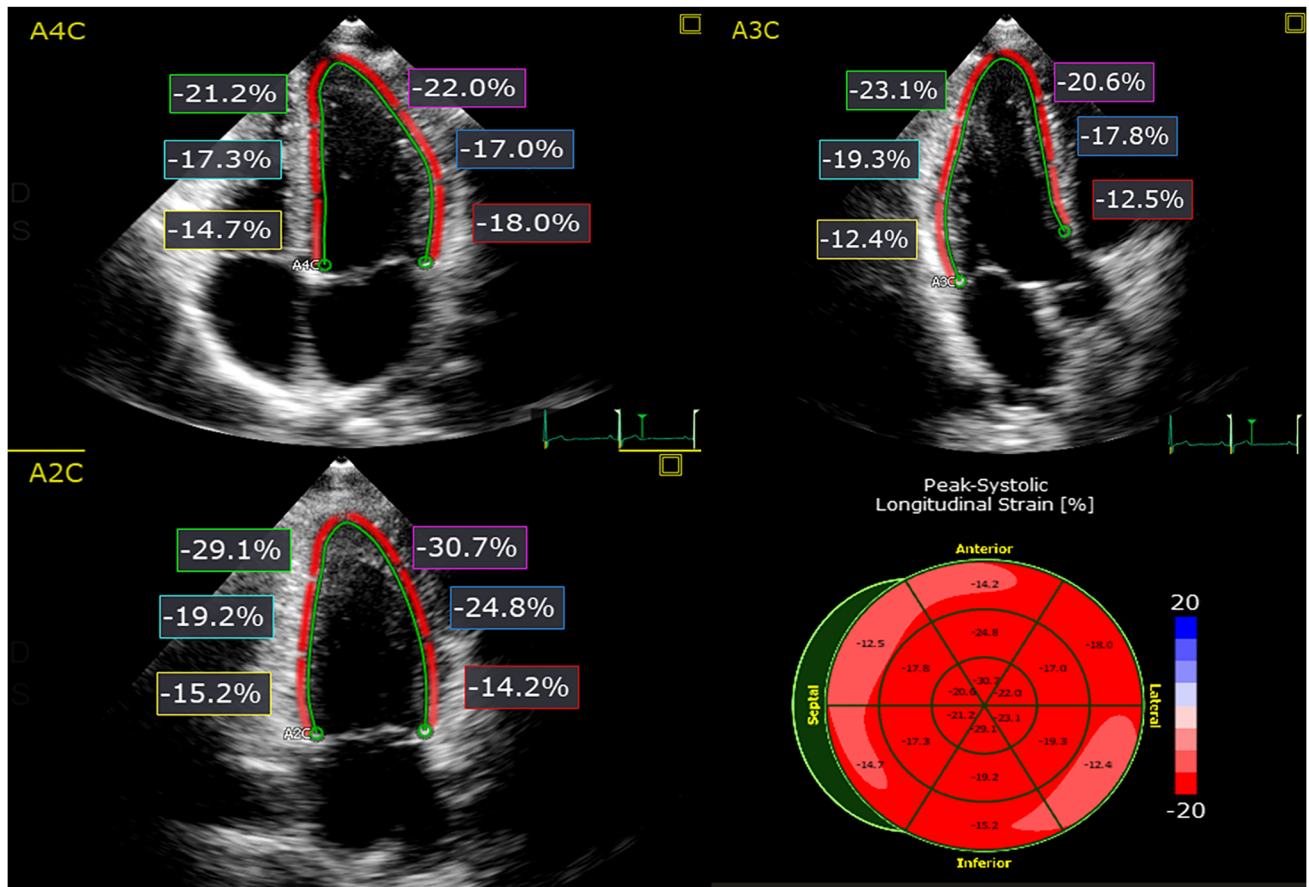


Fig. 1 Two-dimensional speckle tracking echocardiogram. Apical 4 chamber view, Apical 2 chamber view, Apical 3 chamber view, and bulls eye presentation of values LV left ventricle, LA left atrium; Ao aorta

Table 1 Demographic and clinical parameters of JDM patients and healthy controls

	Juvenile dermatomyositis patients (N:26) Mean \pm SD/Median (min–max) / %	Healthy controls (N:20)	P values
Female/Male	14 (57.7%) /12 (42.3%)	9 (45%) /11 (55%) (55%)	
Age (year)	13.5 \pm 3.6	13.8 \pm 4.76	0.465 ^t
BMI, kg/m ²	19.3 (15–26)	18.1 (14–24)	0.538 ^u
Systolic blood pressure mmHg	114 (90–120)	105 (90–115)	0.455 ^u
Diastolic blood pressure mmHg	65 (50–70)	62 (50–65)	0.168 ^u
Heart rate/minute	68 (60–92)	66 (63–75)	0.314 ^u
Age At Diagnose (year)	6.9 (\pm 3.7)		
Disease Duration (year)	5 (1.5–17.5)		
Gastrointestinal Involvement	6 (23%)		
Medications			
Pulse Steroid	7 (27%)		
Methotrexate	26 (100%)		
IVIG (intravenous gammaglobuline)	13 (50%)		
Hydroxychloroquine	13 (50%)		
Anti-TNF	9 (34%)		
Adalimumab	3 (11%)		
Etanercept	7 (26%)		

^tStudent t Test, ^uMann-Whitney U test

Table 2 Comparison of conventional and speckle tracking echocardiographic measurements of JDM patients and controls

	JDM patients (26) Mean \pm SD/median (min–max)	Healthy controls (20) Mean \pm SD/median (min– max)	p
E/A	1.48 \pm 0.36	1.50 \pm 0.18	0.217 ^t
E'	19.8 \pm 4.8	18.3 \pm 2	0.096 ^t
A'	8.2 \pm 2.3	7.9 \pm 1.9	0.075 ^t
S'	11.2 \pm 1.9	11.4 \pm 1.3	0.567 ^t
E/E'	4.04 \pm 1.1	4.22 \pm 1.2	0.223 ^t
EF (Teicholz) %	62 \pm 5.6	67 \pm 3.4	0.001 ^t
LV Longitudinal strain 4 chamber %	– 22.7 \pm 4.6	– 28.6 \pm 4.51	0.001 ^t
LV Longitudinal strain 3 chamber %	– 20.9 \pm 5.2	– 27.0 \pm 4.0	0.0001 ^t
LV Longitudinal strain 2 chamber %	– 23.1 \pm 5.1	– 25.5 \pm 3.4	0.0001 ^t
LV Global Longitudinal strain average %	– 22.2 \pm 4.1	– 26.5 \pm 5.3	0.02 ^t
Right Ventricle (RV)			
TAPSE (mm)	21.4 \pm 3.1	22.3 \pm 1.97	0.573 ^t
RV Frontal Wall strain %	– 26.5 \pm 4.2	– 28.9 \pm 3.6	0.456 ^t
RV Longitudinal strain 4 chamber %	– 23.9 \pm 4.1	– 24.0 \pm 1.6	0.764 ^t

A': late peak diastolic myocardial velocity, S': peak myocardial systolic velocity, E/ E': Ratio of early diastolic filling velocity to early diastolic myocardial relaxation velocity which is used to estimate LV filling pressures, EE': early diastolic myocardial velocity: early diastolic myocardial velocity

^tStudent t Test, ^uMann-Whitney U test,

^EEF ejection fraction, ^{E/A} Ratio of early diastolic filling velocity to accelerated diastolic filling velocity, Tissue Doppler parameters

Correlations of Cardiac Functions with Disease Severity and Treatment

A correlation was observed between LV GLS and age at diagnosis ($r = -0.499$; $p = 0.011$) (see Fig. 2). Similarly, a comparable correlation was noted between the E/E' ratio, a parameter considered prognostic for cardiovascular disease development, and age at diagnosis ($r = -0.469$; $p = 0.018$). There was a negative correlation between RV GLS and age at diagnosis ($r = -0.443$; $p = 0.024$).

When analyzing treatment strategies among children with dermatomyositis, it was found that patients using pulse steroids exhibited lower GLS average values compared to non-users ($\beta = 4.99$, 95% CI: 1.34–8.64, $p = 0.009$). No correlation was identified between medications other than pulse steroids and clinical parameters, as well as LV systolic or diastolic function parameters (GLS, E/E') ($p > 0.05$) Table 3.

Discussion

We present cardiac function parameters in children with Juvenile dermatomyositis based on a widely utilized new echocardiography platform. The distinctiveness of our study lies in the integration of STE with standard echocardiograms, which, despite being normal, enabled the identification of preclinical systolic function impairment in JDM. Furthermore, we established a significant correlation between

LV systolic function (GLS) and the age at diagnosis, while indicating that individuals using pulse steroids demonstrated decreased systolic function compared to non-users, it is essential to note that the number of patients utilizing pulse steroids was limited, making it challenging to draw precise conclusions.

Systolic Functions as Assessed by Speckle Tracking Echocardiography

Guerra et al. [6] affirmed that conventional parameters of systolic and diastolic function in controls versus dermatomyositis patients, through classical echocardiography, lack sensitivity for detecting cardiac involvement in asymptomatic individuals. Like Guerra et al., we detected subclinical LV systolic impairment (measured through GLS) in dermatomyositis patients using STE. However, contrary to their findings, we established an association between reduced GLS values and disease activity/damage indexes, illustrating the potential utility of GLS as an indicator of disease severity. Diniz et al. [7] previously demonstrated early systolic myocardial impairment in pediatric JDM patients with intact EF using STE, highlighting the connection between reduced GLS values and disease activity/damage. Our findings further contribute to this understanding by unveiling lowered GLS in JDM patients despite normal EF, indicating subclinical myocardial systolic dysfunction. Additionally, the correlation between age at diagnosis and GLS suggests a decline

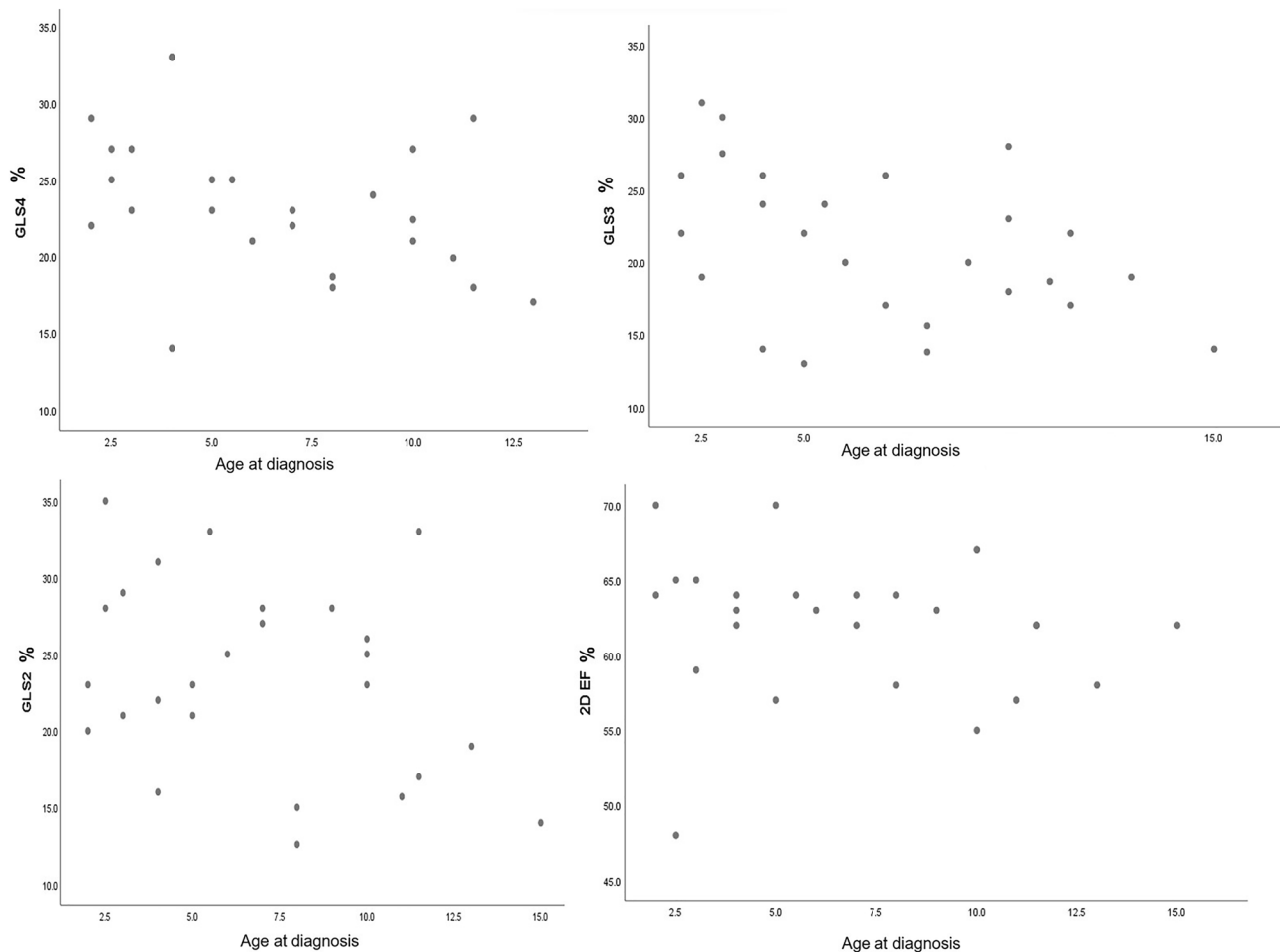


Fig. 2 Graphics showing scatter plots between cardiac functions and age at diagnosis of JDM patients

in systolic function with increasing age, possibly attributed to proinflammatory mediators causing cardiac remodeling and fibrosis [14]. The analogous correlation between RV GLS and age at diagnosis supports the notion of ongoing, low-grade inflammation affecting various organs in JDM.

Diastolic Functions

Schwartz et al. [5] previously detected LV diastolic dysfunction in JDM patients using conventional echocardiography, where 22% exhibited an abnormal E/E' ratio. While these authors reported diastolic dysfunction based on E' and E/E' ratios, our study, employing E/A and E/E' parameters along with established normal ranges for children [15], did not identify any difference in diastolic functions between patients and healthy controls. This discrepancy could potentially be attributed to the shorter disease duration and younger age of our cohort, suggesting that these factors may influence the observed differences.

In summary, our study contributes valuable insights into cardiac function assessment in JDM, particularly through the utilization of STE echocardiography. The identified correlations between LV systolic function, disease severity, and age at diagnosis, as well as the implications of pulse steroid use, provide important avenues for further research and clinical consideration.

Correlations of Cardiac Functions with Disease Severity and Treatment

Diniz et al. previously established a connection between reduced GLS and disease activity/damage indices in JDM patients, along with adverse correlations between GLS and cumulative doses of prednisone and methotrexate. These correlations likely reflect the severity and duration of the disease. Notably, the use of azathioprine was similarly associated with lower LV GLS values, indicating increased myocardial injury in more severe JDM cases. Given that all our patients were in remission and only a few cases exhibited

Table 3 Comparing conventional and speckle tracking echocardiographic measurements in juvenile dermatomyositis (JDM) patients who received pulse steroid therapy and those who did not

	JDM patients who received pulse steroid	Patients who did not received pulse steroid therapy	p
	Mean \pm SD/median (min–max)	Mean \pm SD/median (min–max)	
Systolic Blood pressure mmHg	116 (95–120)	108 (90–110)	0.567 ^u
Diastolic Blood pressure mmHg	69 (58–70)	63 (50–65)	0.305 ^u
BMI, kg/m ²	18.85 (17–26)	16.9 (14–24)	0.258 ^u
E/A	1.6 (1.25–1.80)	1.50 (1.2–2.1)	0.869 ^u
E/E'	4.1 (2.9–6)	4.4 (2.7–7.6)	0.921 ^u
2D Ejection fraction %	65.5 (62.5–78)	65 (57–74)	0.475 ^u
LV Global Longitudinal strain 4 chamber %	– 19.7 (14–27)	– 23.5 (14–33)	0.831 ^u
LV Global Longitudinal strain 3 chamber %	– 16.3 (13–27.5)	– 22.0 (14–31)	0.054 ^u
LV Global Longitudinal strain 2 chamber %	– 17.3 (12.6–29)	– 25(15–35)	0.0281 ^u
LV Global Longitudinal strain average %	– 18.2 (14–28)	– 23.7 (16.3–30.0)	0.023 ^u
Right Ventricle (RV)			
TAPSE (mm)	19.5 (17–25)	22.3 (18–28)	0.196 ^u
RV Frontal Wall strain %	– 23.5 (20–33)	– 28 (14.9–44)	0.123 ^u
RV Longitudinal strain 4 chamber %	– 21 (17–31)	– 25 (16.9–35)	0.831 ^u

E/A: Ratio of early diastolic filling velocity to accelerated diastolic filling velocity, E/E': Ratio of early diastolic filling velocity to early diastolic myocardial relaxation velocity which is used to estimate LV filling pressures

^uMann-Whitney U test

active disease during this study [16], we were unable to identify any links between GLS and disease activity. However, our findings indicated that patients undergoing pulse steroid treatment had lower GLS values than non-users. In cases of severe JDM attacks, pulse steroid treatment was administered, suggesting a potential detrimental impact of disease severity on LV deformation. The involvement of inflammation in heart disease has been widely recognized in numerous autoinflammatory disorders such as lupus and scleroderma, as well as in JDM. Our research, and others alike, utilized STE to reveal subtle, unexplained systolic dysfunctions not detectable through conventional echocardiography, suggesting the potential of STE for early dysfunction identification during follow-up [17–20]. Similarly, in JDM, an autoimmune condition similar to those listed, left ventricular strain, a measure of systolic function, was lower than in the control group.

Limitations

The limited size of our patient cohort restricts the generalizability of our findings to the broader population of JDM patients. Furthermore, since most of our patients were in remission, establishing a direct correlation between disease severity and cardiac function proved challenging. Although STE was administered to every patient, it was performed only once per patient. A more comprehensive understanding would emerge from longer follow-up periods with consecutive echocardiographic assessments.

Conclusion

GLS measurements have demonstrated associations with clinical disease indices, offering promise as diagnostic tools for assessing JDM patients throughout the disease's progression. STE even before a conventional echocardiogram, has the potential to identify preclinical cardiac dysfunction in JDM patients. Given the younger age of JDM patients, vigilant observation is necessary. Early detection of myocardial damage through STE may influence treatment decisions and long-term cardiovascular prognosis, highlighting its significance in the management of JDM patients.

Author Contributions RD, OK contributed to the design and data analysis and critically read the final manuscript. NU, AG, YC, SS, FO, AA, KB, SD, UG, EK, EA, MY contributed to data collection, analysis, read and approved the final manuscript.

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Declarations

Conflict of interest The authors declare no competing interests.

Ethical Approval The study was approved by the institution review board.

Informed Consent Informed consent was obtained from the parents or legal guardians of all individual participants included in the study.

References

- Papadopoulou C, McCann LJ (2018) The vasculopathy of juvenile dermatomyositis. *Front Pediatr* 6:284. <https://doi.org/10.3389/fped.2018.00284>
- Barut K, Aydin PO, Adrovic A, Sahin S, Kasapcopur O (2017) Juvenile dermatomyositis: a tertiary center experience. *Clin Rheumatol* 36(2):361–366. <https://doi.org/10.1007/s10067-016-3530-4>
- Witczak BN, Schwartz T, Barth Z et al (2022) Associations between cardiac and pulmonary involvement in patients with juvenile dermatomyositis—a cross-sectional study. *Rheumatol Int* 42(7):1213–1220. <https://doi.org/10.1007/s00296-021-05071-1>
- Ghosh R, Roy D, Dubey S, Abdelrahman K, Ray BK (2020) Juvenile dermatomyositis presenting as complete heart block in a 10-year-old girl. *Paediatr Int Child Health* 40(4):251–254. <https://doi.org/10.1080/20469047.2020.1765123>
- Schwartz T, Sanner H, Gjesdal O, Flato B, Sjaastad I (2014) In juvenile dermatomyositis, cardiac systolic dysfunction is present after long-term follow-up and is predicted by sustained early skin activity. *Ann Rheum Dis* 73:1805–1810. <https://doi.org/10.1136/annrheumdis-2013-203279>
- Guerra F, Gelardi C, Capucci A, Gabrielli A, Danieli MG (2017) Subclinical cardiac dysfunction in polymyositis and dermatomyositis: a speckle-tracking case-control study. *J Rheumatol* 44:815–821. <https://doi.org/10.3899/jrheum.161311>
- Diniz MFR, Kozu KT, Elias AM et al (2021) Echocardiographic study of juvenile dermatomyositis patients: new insights from speckle-tracking-derived strain. *Clin Rheumatol* 40(4):1497–1505. <https://doi.org/10.1007/s10067-020-05418-4>
- Bohan A, Peter JB (1975) Polymyositis and dermatomyositis. *N Engl J Med* 13:344–347. <https://doi.org/10.1056/NEJM197502132920706>
- Dulcan M (1994) Nomenclature and criteria for diagnosis of diseases of the heart and great vessels, 9th edn. Little, Brown & Co., Boston, pp 253–256
- Ruperto N, Martini A (2011) Networking in paediatrics: the example of the paediatric rheumatology international trials organisation (PRINTO). *Arch Dis Child* 96(6):596–601
- Huber AM, Feldman BM, Rennebohm RM et al (2004) Validation and clinical significance of the childhood myositis assessment scale for assessment of muscle function in the juvenile idiopathic inflammatory myopathies. *Arthritis Rheum* 50:1595–1603. <https://doi.org/10.1002/art.20179>
- Haycock GB, Schwartz GJ, Wisotsky DH (1978) Geometric method for measuring body surface area: a height-weight formula validated in infants, children, and adults. *J Pediatr* 93:62–66. [https://doi.org/10.1016/s0022-3476\(78\)80601-5](https://doi.org/10.1016/s0022-3476(78)80601-5)
- Mor-Avi V, Lang RM, Badano LP et al (2011) Current and evolving echocardiographic techniques for the quantitative evaluation of cardiac mechanics: ASE/EAE consensus statement on methodology and indications endorsed by the Japanese society of echocardiography. *J Am Soc Echocardiogr* 24:277–313. <https://doi.org/10.1016/j.echo.2011.01.015>
- Levy PT, Macheffsky A, Sanchez AA et al (2016) Reference ranges of left ventricular strain measures by two-dimensional speckle-tracking echocardiography in children: a systematic review and meta-analysis. *J Am Soc Echocardiogr* 29:209–225. <https://doi.org/10.1016/j.echo.2015.11.016>
- Eidem BW, McMahan CJ, Cohen RR et al (2004) Impact of cardiac growth on doppler tissue imaging velocities: a study in healthy children. *J Am Soc Echocardiogr* 17:212–221. <https://doi.org/10.1016/j.echo.2003.12.005>
- Lazarevic D, Pistorio A, Palmisani E et al (2013) The PRINTO criteria for clinically inactive disease in juvenile dermatomyositis. *Ann Rheum Dis* 72(5):686–693. <https://doi.org/10.1136/annrheumdis-2012-201483>
- Dedeoglu R, Şahin S, Koka A et al (2016) Evaluation of cardiac functions in juvenile systemic lupus erythematosus with two-dimensional speckle tracking echocardiography. *Clin Rheumatol* 35(8):1967–1975. <https://doi.org/10.1007/s10067-016-3289-7>
- Dedeoglu R, Adrovic A, Oztunç F et al (2017) New insights into cardiac involvement in juvenile scleroderma: a three-dimensional echocardiographic assessment unveils subclinical ventricle dysfunction. *Pediatr Cardiol* 38(8):1686–1695. <https://doi.org/10.1007/s00246-017-1714-6>
- Yildiz M, Dedeoglu R, Akdeniz B et al (2022) Systolic and diastolic cardiac functions in juvenile spondyloarthropathies. *J Clin Rheumatol* 28(1):e175–e179. <https://doi.org/10.1097/RHU.0000000000001674>
- Fairley JL, Wicks I, Peters S, Day J (2021) Defining cardiac involvement in idiopathic inflammatory myopathies: a systematic review. *Rheumatol (Oxford)* 61:103–120. <https://doi.org/10.1093/rheumatology/keab573>

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