Original Article

The value of quantitative muscle ultrasound in monitoring muscle and fascia inflammatory activity in juvenile dermatomyositis patients

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Background: Currently, the activity of juvenile dermatomyositis (JDM) is mainly assessed based on clinical manifestations, creatine kinase (CK) level, and magnetic resonance imaging (MRI), but certain limitations arise in these approaches for children. Thus, this cross-sectional study aimed to explore the value of ultrasound in evaluating muscle inflammation via the dynamic analysis of muscle ultrasound characteristics in children with active or stable JDM.

Methods: The data of a group of children who were diagnosed with JDM and admitted to the Rheumatology and Immunology Department of the Capital Pediatric Research Institute Children's Hospital between June 2022 and November 2023, and a normal control group were collected. The clinical, ultrasound, and laboratory data of the children with active and stable JDM were collected and compared with those of a normal control group. Muscle thickness (MT), fascia thickness (FT), muscle echo intensity (EI), muscle microvascular flow imaging (MVFI) distribution, and the blood flow resistance index (RI) were measured via ultrasound for the comparative analysis. A Spearman correlation analysis was conducted to assess the correlation between the ultrasound parameters in the JDM patients, and muscle scores and laboratory indicators of disease activity. Receiver operating characteristic (ROC) curves were generated for the ultrasound parameters. Five active JDM children were dynamically followed up.

Results: The data of 26 children with active JDM, 29 with stable JDM, and 31 normal control children were collected. The patients with active JDM had significantly greater EI (median 68.9 *vs.* 47.4, P<0.01), increased FT (median 0.25 *vs.* 0.15, P<0.01), and an increased MVFI distribution (P<0.01). The Spearman correlation analysis revealed a negative correlation between increased EI, FT, and MVFI distribution, and the Childhood Myositis Assessment Scale (CMAS) score (R=–0.662, –0.673, –0.667, all P<0.05). There was a statistically significant difference in EI between the stable JDM children and healthy children (median 47.4 *vs.* 39.0, P<0.05). During the follow-up period, two children with slow fasciitis resolution developed soft tissue calcification.

Conclusions: EI, FT, and MVFI distribution can be used to assess the activity status of individuals with JDM, and are correlated with clinical activity indices. EI may be abnormal even in children with a stable clinical condition, which suggests that ultrasound may more accurately reflect muscle status. Children with long-term unresolved fasciitis may be at risk of developing calcification.

Keywords: Juvenile dermatomyositis (JDM); quantitative muscle ultrasound; disease activity

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Introduction

Juvenile dermatomyositis (JDM) is a rare systemic autoimmune disease in children characterized by chronic muscle and skin inflammation leading to proximal muscle weakness and rash (1). The incidence of JDM is 2/1,000,000 to 4/1,000,000 (2), and it can affect multiple organs in the body, has a mortality rate of 1–5% (3-5), and poses a significant threat to children's health. JDM is the most common inflammatory muscle disease with an occurrence rate of up to 85% (6). Currently, there are many challenges in the diagnosis and treatment of JDM, such as how to monitor muscle inflammation in affected children.

Existing clinical and laboratory indicators include the Childhood Myositis Assessment Scale (CMAS) (7), muscle enzymes, muscle magnetic resonance imaging (MRI), and electromyography (8-10). It is difficult to obtain CMAS scores in younger patients because of their lack of cooperation. Additionally, the CMAS provides a subjective score, and the sensitivity and specificity of laboratory indicators, such as creatine kinase (CK) are relatively poor. MRI is sensitive to muscle edema and inflammation, but it is expensive, requires high cooperation from the child, may require anesthesia in younger patients, and is not suitable for serial investigations with dynamic monitoring. Conversely, ultrasound is convenient to perform and can provide real-time dynamic scanning to evaluate muscle structure and muscle perfusion (1,8,11). Currently, ultrasound has been used in the study of inflammatory muscle diseases (12,13), and the results showed that increased muscle echo intensity (EI), also known as echogenicity, can be used to determine the activity of JDM (14,15). However, the parameters are relatively simple, and no research has been conducted on important parameters such as fascia thickness (FT), muscle perfusion, and intramuscular blood flow dynamics, nor has an analysis of the correlation between ultrasound parameters, and clinical disease activity indicators of the JDM been conducted.

This study used the following five quantitative ultrasound parameters to conduct multidimensional and dynamic analyses of the quadriceps, anterior tibialis muscle, biceps brachii, and forearm flexors in children with active JDM, stable JDM, and healthy controls to explore the differences between the active phase, stable phase and no JDM: muscle thickness (MT) Z score; muscle EI; muscle FI; microvascular flow imaging (MVFI) distribution; and the flow resistance index (RI). To explore the diagnostic and monitoring value of the ultrasound parameters for disease activity, this study also examined the correlation between CMAS scores and laboratory indicators of ultrasound parameters in patients with this disease. We present this article in accordance with the STROBE reporting checklist (available at https://qims.amegroups.com/article/view/10.21037/qims-24-1035/rc).

Methods

Patients

Data from children who were diagnosed with JDM and admitted to the Rheumatology and Immunology Department of the Capital Pediatric Research Institute Children's Hospital between June 2022 and November 2023 were collected. In this study, a total of 55 JDM patients were enrolled, of whom 26 were in the active phase, and 29 were in the stable phase. To be eligible for inclusion in this study, the JDM patients had to meet the following inclusion criteria: have a diagnosis of JDM based on the diagnostic criteria proposed by Behan/Peter in 1975 (16); and meet the European League Against Rheumatism/American College of Rheumatology criteria (17). Biochemical, MRI, muscle biopsy, and electromyogram were performed at the beginning of diagnosis. The study included patients of any sex who were aged 0–16 years. The exclusion criteria for patients were concomitant infections, other autoimmune diseases, tumors, hereditary diseases, and other chronic conditions that might affect the assessment. The diagnostic criteria for JDM activity were patients who had not been treated or had experienced a JDM relapse, with clinical manifestations of decreased muscle strength, elevated CK, and abnormal signals on muscle MRI (on at least two indicators). The diagnostic criteria for a stable JDM state were normal clinical findings, laboratory indicators, and muscle MRI findings. There was no overlap between

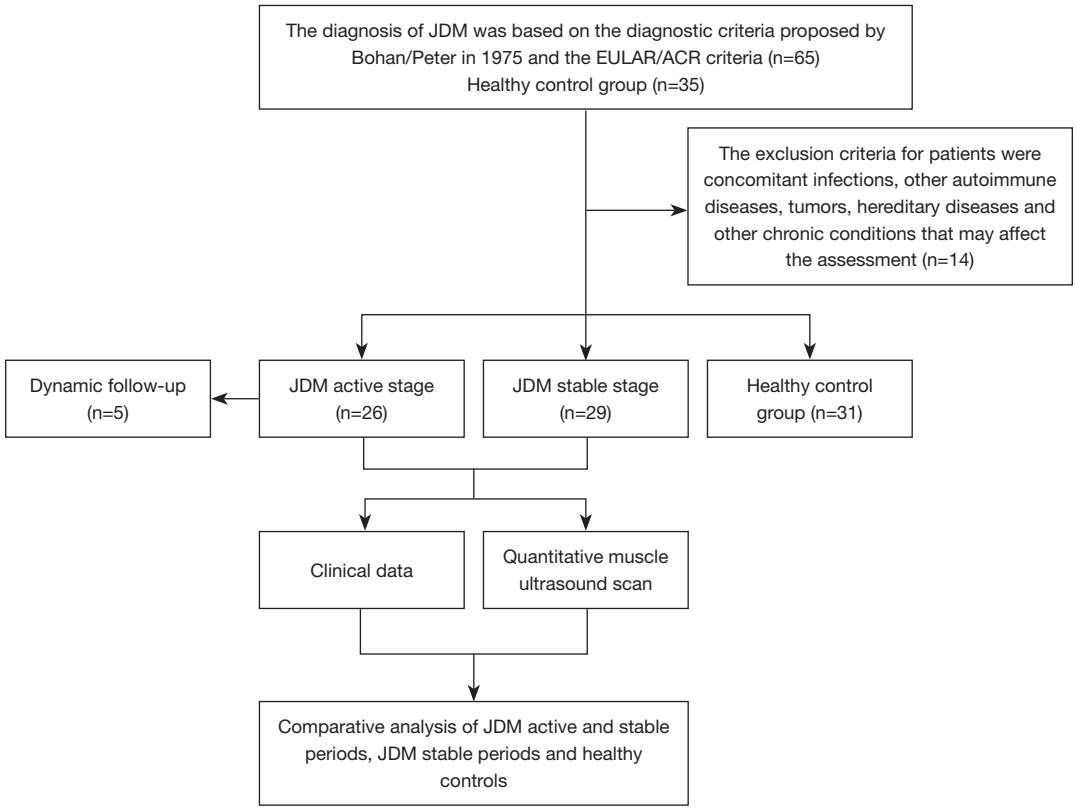

Figure 1 Flowchart of the overall study design. JDM, juvenile dermatomyositis; EULAR, European League Against Rheumatism; ACR, American College of Rheumatology.

children with active and stable JDM.

After the first examination, five children with JDM underwent dynamic follow-up by muscle ultrasound every 3 months for 12 months. Details of whether the child needed anesthesia during the ultrasound and MRI, the examination duration, and adverse reactions were recorded.

This study included 31 age- and sex-matched healthy controls. Healthy controls were matched with the stable JDM patients in terms of gender and age. Based on the gender and age distribution characteristics of the children with stable JDM, the children in the healthy control group were manually age- (1–14 years) and sex-matched (1:1).

The guardians of the healthy control group signed the informed consent form. The number of cases were treated at the hospital during the study period determined the sample size (*Figure 1*).

The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). This study was approved by the Ethics Committee of the Capital Pediatric Research Institute (No. SHERLL2023039), and informed consent was obtained from the patients' parents or legal guardians.

Ultrasonography examinations

Ultrasound was performed with a Logic E11 Color Doppler Ultrasound Diagnostic Instrument (GE HealthCare, Chicago, IL, USA) and a linear array probe, with a frequency of 6–15 MHz, using musculoskeletal condition with a gain compensation of 56 and a dynamic range of 63 dB. To avoid the influence of parameter changes on muscle EI during the examination process, all system setting parameters remained unchanged throughout the entire study (15). This study explored the biceps brachii, forearm flexors, quadriceps femoris, and tibialis anterior, and measurements were taken using transverse ultrasound images (*Figure 2*). The ultrasound examinations were performed by ultrasound physicians with more than 10 years of experience each, and the interpretation of the blood flow images was determined after consultation with

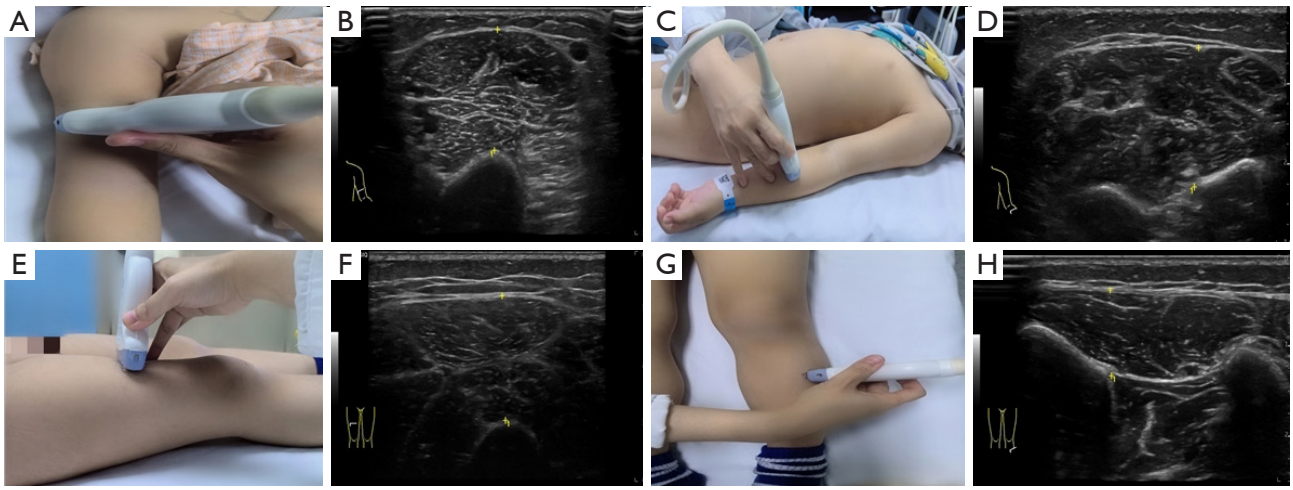

Figure 2 The measurement positions for the ultrasound examination of the limbs. (A,B) Biceps brachii: the biceps brachii is measured at two-thirds of the distance from the acromion to the anterior fold of the elbow. (C,D) Brachioradialis: the brachioradialis is measured at two-fifths of the distance from the anterior aspect of the olecranon to the distal end of the radius. (E,F) Quadriceps femoris: the quadriceps femoris is measured at half the distance from the anterior superior iliac spine to the upper border of the patella. (G,H) Tibialis anterior: the left tibialis anterior is measured at one-fourth of the distance from the inferior aspect of the patella to the lateral malleolus.

two physicians. The detailed examination parameters included the following:

- MT: the thickness of the biceps brachii, forearm flexors, quadriceps femoris, and tibialis anterior were measured. Considering the correlation between the MT at the measurement site and body weight, the following MT formulas were used: 0.77±0.028 W for the biceps brachii (SD: 0.16); 1.18±0.018 W for the forearm flexors (SD: 0.23); 1.63±0.042 W for the quadriceps femoris (SD: 0.34); and 0.81±0.019 W for the tibialis anterior (SD: 0.19), where W represents body weight, and SD represents the standard deviation. The MT was represented by the Z score, which was calculated as follow: Z score = measured thickness – corrected thickness/SD (18).
- FT: FT in the lateral thigh muscles was measured; three measurements were taken, and the thickest measurement was recorded.
- Muscle EI: muscle ultrasound images were collected for subsequent quantitative image analysis.
- Microvascular distribution in muscles: MVFI was used to clearly display muscle blood flow without significant noise. The blood flow criteria were as follows: Within a fixed sampling frame of 3 cm × 2 cm, 1–2 blood flow signals within the muscle indicate a small amount of blood flow, 3–4 blood flow signals indicate a moderate amount of blood flow, and ≥4 blood vessel signals indicate abundant blood flow within the muscle.
- Muscle blood flow RI: the blood flow spectrum of the intermuscular artery in the quadriceps femoris of the thigh was measured, and the RI was recorded.

Quantitative analysis of muscle ultrasound images via ImageJ software

For each group of patients, three muscle images were selected from four muscle groups. ImageJ software (National Institutes of Health, Bethesda, MD, USA) was used to select the region of interest and measure the muscle grayscale value. Images of reactive muscles with strong muscle inflammation were selected during the active period of JDM, and normal muscles were randomly selected during the stable period of the JDM and in the normal control group. The average value was taken to represent the muscle EI condition of the patient (*Figure 3*). The selection of the region of interest was determined through consultation and confirmation by two ultrasound physicians. The two physicians were blinded to the activity measures (CMAS score, CK, and MRI) of the children with JDM.

Clinical assessments

Clinical data, including sex, age at diagnosis, presenting symptoms, disease duration, weight of the patient during the

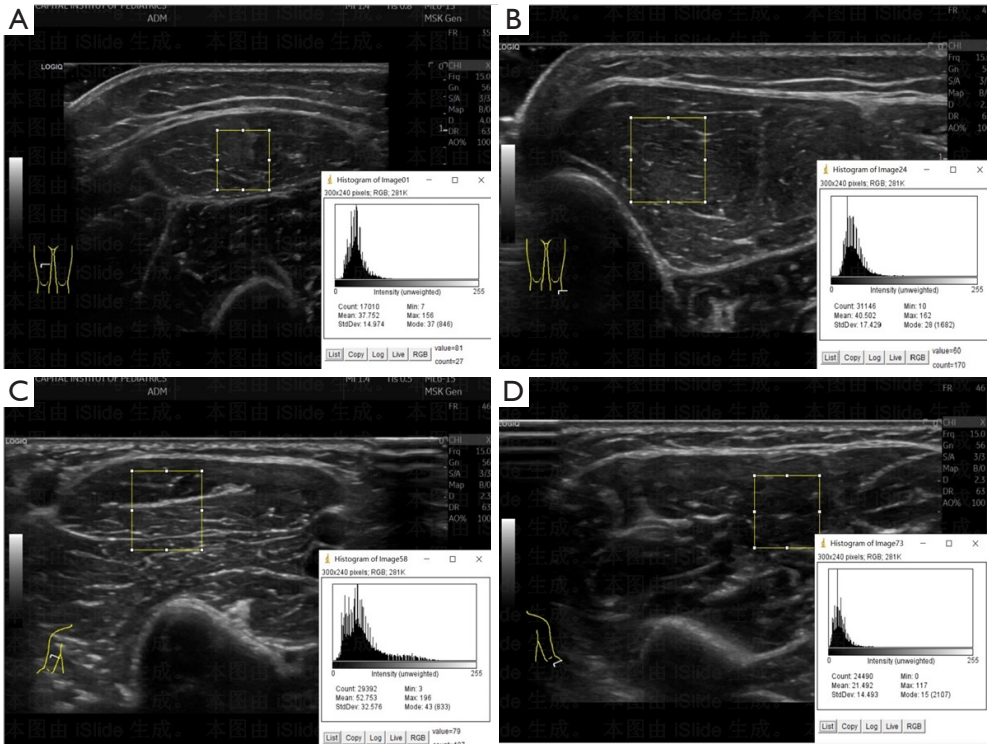

Figure 3 Quantifying muscle EI using image analysis software: (A) quadriceps femoris; (B) tibialis anterior; (C) biceps brachii; (D) brachioradialis. The regions of interest were selected, and ImageJ image analysis software was used to quantify the grayscale values. EI, echo intensity.

ultrasound examination, CMAS muscle strength score (19), laboratory indicators of CK, lactic dehydrogenase (LDH), α-hydroxybutyric acid (α-HBDH), the erythrocyte sedimentation rate (ESR), serum ferritin (SF), and myositis antibody, and MRI results, were collected.

Statistical analysis

All the data were analyzed using SPSS 25.0 (IBM Corp, Armonk, NY, USA). The continuous data are presented as the median (range), or the mean ± SD, while the categorical variables are presented as the count and percentage (%). A comparative analysis was conducted to examine the laboratory and ultrasound parameters in children with JDM, and to compare the ultrasound parameters between JDM and normal children. The Chi-square test was used to compare the categorical variables. The independent samples *t*-test was used to compare the continuous variables. The ultrasound findings and the MRI results were examined using the kappa consistency test. A Spearman correlation analysis was conducted to assess the correlation between the ultrasound parameters in the JDM patients, muscle scores, and laboratory indicators of disease activity. Receiver operating characteristic (ROC) curves were generated for the ultrasound parameters to determine JDM activity, and the area under the curve (AUC), sensitivity, and specificity were calculated. A two-sided P value <0.05 was considered statistically significant.

Results

Clinical features

In this study, a total of 55 JDM patients were enrolled, of whom 30 (54.5%) were male and 25 (45.5%) were female, and of whom 26 were in the active phase and 29 were in the stable phase. The age of onset ranged from 1 to 14 years with an average age of 5.43±2.59 years for patients in the active phase, and 6.03±3.52 years for patients in the stable phase. The duration of illness at the time of visit ranged from 1 to 48 months with an average duration of 7.54 months. Five of the 55 patients in the active phase were

Table 1 Population characteristics and disease activity indicators in the active and stable phases of JDM

Variables	Active (n=26)	Stable (n=29)	P
Age (years)	5.2 [1.3–10.2]	6.0 [1.1–14.2]	0.34
Sex (female)	11 (42.3)	14 (48.3)	0.66
Disease duration (years)	0.5 [0.1–3.0]	2.4 [0.3–7.7]	
Initial symptoms[†]			
Rash	13 (50.0)	11 (37.9)	0.57
Rash and fatigue	6 (23.1)	16 (55.2)	0.11
Fatigue	6 (23.1)	2 (6.9)	0.14
Joint swelling and pain	1 (3.8)	0	0.29
JDM disease activity assessment			
CMAS (normal range, 0–52)	15 [1–48][‡]	51 [35–52][§]	<0.01
CK (U/L) (normal range, 50–220)	233 [21–3,438]	81 [14–198]	<0.01
LDH (U/L) (normal range, 80–300)	424 [230–1,113]	224 [146–321]	<0.01
α-HBDH (U/L) (normal range, 120–260)	316 [163–682]	159 [102–251]	<0.01
ESR (mm/60 min) (normal range, 0–20)	13 [3–58]	9 [3–26]	0.21
SF (ng/mL) (normal range, 10–120)	190 [34–1,080]	91 [17–692]	0.06
Myositis antibody			
Negative	12 (46.2)	12 (41.4)	0.76
MDA5	4 (15.4)	6 (20.7)	0.71
NXP2	6 (23.1)	5 (17.2)	0.62
Jo-1	1 (3.8)	1 (3.4)	0.92
Mi-2β	1 (3.8)	1 (3.4)	0.92
PL12	1 (3.8)	1 (3.4)	0.92
TIF1γ	1 (3.8)	3 (10.3)	0.41

Data are presented as the median [range] or number (%). [†], all patients presented with a rash in the early stages of their illness; [‡], 12 patients; [§], 20 patients. JDM, juvenile dermatomyositis; CMAS, Childhood Myositis Assessment Scale; CK, creatine kinase; LDH, lactic dehydrogenase; α-HBDH, α-hydroxybutyric acid; ESR, erythrocyte sedimentation rate; SF, serum ferritin.

followed up regularly for dynamic monitoring. Because some of the children could not cooperate, 32 children (12 in the active phase and 20 in the stable phase) were assessed using the CMAS (20) (*Table 1*). The median CMAS score for JDM patients in the active phase was 15 (range, 1–48), whereas the median CMAS score for JDM patients in the stable phase was 51 (range, 35–52).

All 55 children with JDM underwent ultrasound and MRI. The ultrasound examination was performed without anesthesia, and the average examination time was 17 [15–20] minutes. No child experienced adverse reactions. Seventeen children under 5 years of age required anesthesia in MRI examination, and the average examination time was 27 [20–37] minutes, and no child experienced adverse reactions.

There were 31 participants in the healthy control group, of whom 16 were female and 15 were male. There were no statistically significant differences between the JDM stable phase group and the healthy control group in terms of sex or age.

Laboratory examinations

In this study, an independent samples *t*-test was used

to compare the laboratory indicators (i.e., CK, LDH, and α-HBDH) between the children in active and stable phases of JDM. The statistical results revealed that there were significant differences in CK, LDH, and α-HBDH between the two groups (P<0.05). However, there were no statistically significant differences in the ESR or SF between the two groups (P>0.05). ROC curves were plotted for the three laboratory indicators (i.e., CK, LDH, and α-HBDH). The AUC values were 0.82, 0.94, and 0.96, respectively. The sensitivity was 76.9%, 84.6%, and 88.5%, while the specificity was 82.8%, 93.1%, and 96.6% for CK, LDH, and α-HBDH, respectively (*Figure 4*).

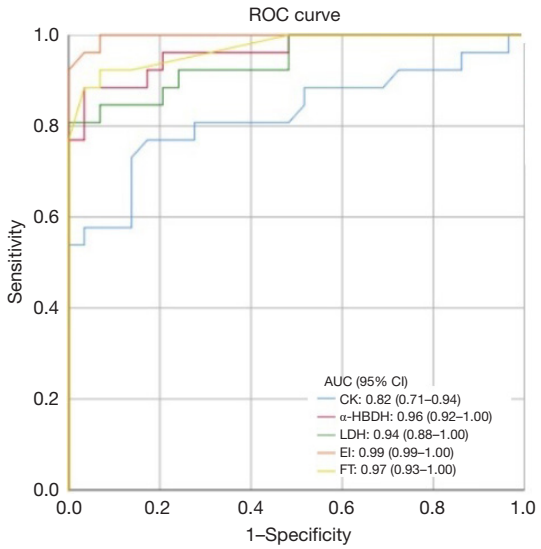

Figure 4 ROC curves of clinical and ultrasound parameters. ROC, receiver operating characteristic; AUC, area under the curve; CI, confidence interval; CK, creatine kinase; α-HBDH, α-hydroxybutyric acid; LDH, lactic dehydrogenase; EI, echo intensity; FT, fascia thickness.

Ultrasound findings

Ultrasonographic data during the active and stable phases of JDM

The statistical results revealed that the EI, FT, and MVFI distribution were significantly different between the groups with active and stable phases of JDM (P<0.01), while the MT and RI were not significantly different (P>0.05) (*Table 2*). Among the patients in the active phase of JDM, 15 of 26 had increased microvascular distribution, while 11 of 26 had normal microvascular distribution. Comparisons between the two groups revealed significant differences in the laboratory indicators (i.e., CK, LDH, and α-HBDH) (all P<0.05). The results of the ultrasound activity assessment were completely consistent with those of the MRI with a kappa value of 1.

A Spearman correlation analysis was performed to assess the correlation between the ultrasound parameters and CAMS muscle scores, as well as the laboratory markers of disease activity. The analysis revealed that increased muscle EI, increased FT, and increased microvascular signals were negatively correlated with the CAMS muscle scores (R=–0.662, –0.673, and –0.667, respectively; all P<0.05) (*Table 3*).

Table 2 Quantitative muscle ultrasonography parameters in the active and stable phases of JDM

Muscle ultrasound parameters	Active (n=26)	Stable (n=29)	P
MT			
Biceps brachii (z score)	–0.2 (–2.4 to 1.4)	–1.1 (–3.2 to 2.1)	0.13
Brachioradialis (z score)	–0.2 (–2.9 to 0.9)	–0.6 (–1.8 to 1.3)	0.22
Quadriceps femoris (z score)	–1.5 (–3.8 to –0.8)	–1.6 (–6.6 to 0)	0.11
Tibialis anterior (z score)	1.1 (–1.9 to 3.4)	0.9 (–1.0 to 2.5)	0.55
EI (GSL)	68.9 (58.5 to 78.2)	41.4 (27.4 to 51.8)	<0.01
FT (cm)	0.25 (0.16 to 0.41)	0.15 (0.09 to 0.22)	<0.01
MVFI (increased)	15 (57.7)	0	<0.01
RI	0.75 (0.62 to 1.37)	0.82 (0.70 to 1.20)	0.073

Data are presented as the median (range) or number (%). JDM, juvenile dermatomyositis; MT, muscle thickness; EI, echo intensity; GSL, grayscale level, 0–255 (unitless range); FT, fascia thickness; MVFI, microvascular flow imaging; RI, resistance index.

Table 3 Correlation analysis between JDM ultrasound parameters and disease activity indicators

JDM disease assessment	EI		FT		MVFI	
	Value	P	Value	P	Value	P
CMAS	–0.662	<0.01	–0.673	<0.01	–0.667	<0.01
CK	0.440	<0.01	0.472	<0.01	0.610	<0.01
LDH	0.691	<0.01	0.766	<0.01	0.693	<0.01
α-HDBH	0.707	<0.01	0.764	<0.01	0.661	<0.01

JDM, juvenile dermatomyositis; EI, echo intensity; FT, fascia thickness; MVFI, microvascular flow imaging; CMAS, Childhood Myositis Assessment Scale; CK, creatine kinase; LDH, lactic dehydrogenase; α-HBDH, α-hydroxybutyric acid.

Based on the independent samples *t*-test between the active and stable phases of JDM, the EI, FT, and MVFI distribution were identified as effective indicators of disease activity. A ROC curve analysis was performed of the EI, FT, and MVFI distribution. The results showed that EI and FT had high predictive value. The AUC, sensitivity, and specificity for muscle EI were 0.99, 96.2%, and 96.6%, respectively, while those for FT were 0.97, 92.3%, and 93.1%, respectively (*Figure 4*).

Dynamic monitoring of five children with JDM during the active phase

Among the 28 children with JDM, five underwent continuous dynamic monitoring. The monitoring period ranged from 12 to 19 months. The number of examinations for these patients ranged from 2 to 7. Among these five patients, three had normal muscle and fascia inflammation recovery, and two had normal muscle recovery but incomplete fascia inflammation recovery. The first ultrasound examinations of the five patients revealed EI values between 63.9 and 76.5, and FT values ranging from 0.17 to 0.35 cm. The last ultrasound examination revealed EI values between 30.4 and 59.4, and FT values ranging from 0.13 to 0.26 cm. Two patients experienced slower remission with muscle recovery times of 12 and 19 months, respectively. These two patients had significantly greater muscle EI values than the other patients with EI values of 57.2 and 59.4, respectively. Interestingly, among the five children who were followed up for more than 12 months, two with persistent fasciitis later developed subcutaneous soft tissue calcification (*Figure 5*, *Table 4*).

Discussion

JDM is a type of autoimmune microvascular disease that primarily affects the skin and muscles. Currently, the assessment of muscle damage in JDM patients is mainly based on muscle strength grading and muscle enzyme levels. However, assessing muscle weakness can be challenging, especially in children, and muscle enzymes may be more strongly associated with muscle inflammation in the early stages of the disease. As the condition progresses, muscle enzyme levels may gradually return to normal and may not accurately reflect muscle inflammation. Electromyography and muscle biopsy may yield false-negative results due to the location at which the test or biopsy is performed, and these invasive tests cannot provide dynamic monitoring (21-23). MRI examinations have high sensitivity but are limited in their application for disease assessment due to their high costs (24), the need for sedation in children, and the length of such procedures. Compared to MRI, ultrasound has many advantages, including that it is convenient, cost-effective, non-invasive, and allows for the real-time dynamic observation of muscle status. There have been reports of ultrasound EI being used to assist in the diagnosis and assessment of disease activity in individuals with JDM, but such studies have largely focused on muscle EI (25).

The study undertook a comprehensive evaluation of the value of ultrasound parameters in assessing disease activity in JDM. Data were collected from 55 children with JDM and multiple parameters, dimensions, and dynamic monitoring, including MT Z scores, muscle EI, muscle FT, MVFI distribution, and the vascular RI, were used. This study compared and analyzed the differences in ultrasound images between patients in the active and stable phases of JDM, as well as between patients in the stable phase of JDM and healthy controls. This study also explored the correlation between ultrasound parameters and CAMS scores and laboratory activity indicators.

In ultrasound images, muscle EI can be enhanced by increased inflammation, fibrous tissue, or fatty components (25-27). This study revealed that EI was significantly

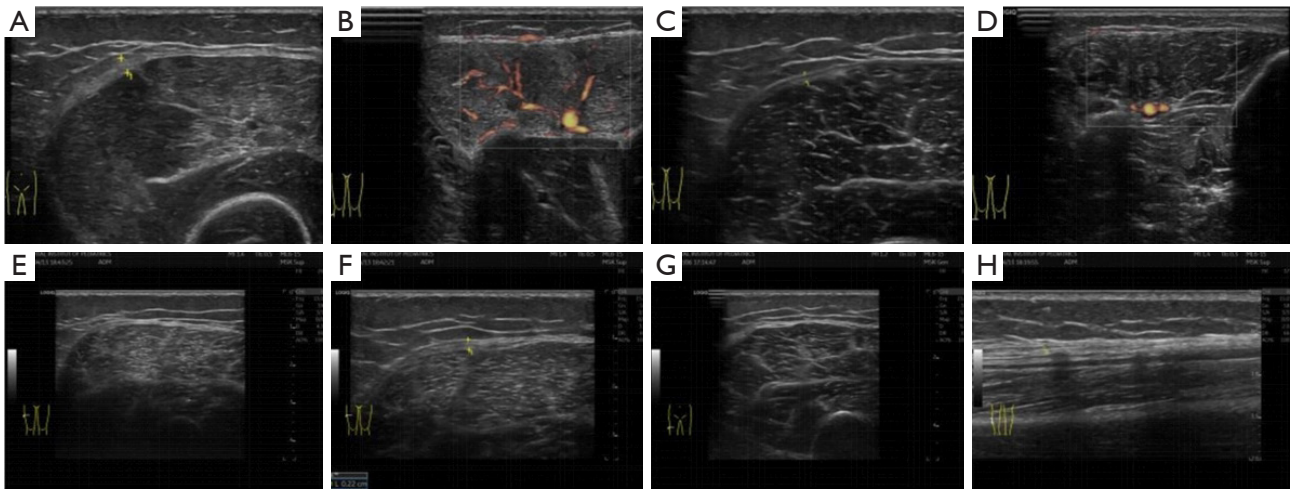

Figure 5 Muscle ultrasounds of active-phase JDM patients. (A) Increased muscle EI and thickened fascia. (B) Abundant blood flow signals. (C,D) Muscle ultrasounds of a stable-phase JDM patient. (C) Normal muscle EI and no thickening of the fascia. (D) No increased fine blood flow. (E-H) The follow-up MRI of patient 1. (E,F) The first examination in which the muscle EI was increased and the fascia was thickened. (G) Normal muscle EI after 8 months of re-examination. (H) Early fascial calcification found in one examination. JDM, juvenile dermatomyositis; EI, echo intensity; MRI, magnetic resonance imaging.

Table 4 Information on the five patients with active JDM who were followed-up

No.	Follow-up time (months)	Number of examinations	Recovery time (months)	First time			Last time			Complications
				EI	FT	RI	EI	FT	RI	
1	12	4	†	76.5	0.25	0.86	59.4	0.23	0.78	Calcification
2	12	5	10	63.9	0.17	0.83	30.4	0.14	1.02	‡
3	18	5	6	72.23	0.23	0.74	36.3	0.13	0.82	‡
4	19	7	†	72.9	0.35	0.77	57.2	0.26	0.93	Calcification
5	12	4	4	75.4	0.31	0.73	40.8	0.20	0.76	‡

†, muscle recovery, still with fasciitis; ‡, no complications. JDM, juvenile dermatomyositis; EI, echo intensity; FT, fascia thickness; RI, resistance index.

greater in children with active JDM than children with stable JDM (median 68.9 *vs.* 47.4, P<0.01), which is consistent with previous research findings. The study further analyzed the correlation between EI and clinical and laboratory indicators and performed a ROC curve analysis to determine sensitivity and specificity. The results showed that the EI was negatively correlated with the CAMS score, indicating that higher EI values were associated with muscle weakness. These findings suggest that EI not only reflects muscle inflammation but could also serve as an effective indicator for assessing the severity of muscle damage in children with JDM. The AUC, sensitivity, and specificity of muscle ultrasound EI were 0.99, 96.2%, and 96.6%, respectively, the sensitivity and specificity of EI were greater than those of laboratory markers of disease activity; thus, EI could be an important ultrasound parameter for diagnosing and evaluating JDM muscle damage. This study also found that during the stable phase, three children had high EI values (EI: 53–55); all three children were obese and had a high body mass index (BMI: 22, 27, 29 kg/m^2). This may be due to rapid weight gain caused by the use of glucocorticoids, leading to the infiltration of muscle by fat cells. Although the inflammation had disappeared, the muscles were still infiltrated by fat cells, which suggests that clinical stability may not represent complete recovery from muscle inflammation. Such children may require

longer follow-up periods to clarify the characteristics of the changes in muscle ultrasound EI.

This study revealed that, compared with those of the healthy controls, the clinical, laboratory, and muscle MRI findings of children with stable JDM returned to normal, but ultrasound EI only partially returned to normal in some children. Some children still had higher EI values than healthy children. The reason for this could be that ultrasound examination is more sensitive and can detect that the muscles in some children in the clinical remission phase have not fully recovered. Alternately, this could be due to muscle damage during treatment, leading to increased fat infiltration in the muscle interstitium, resulting in increased EI. Further studies are needed to confirm these findings, and long-term follow-up observations are still needed in the future.

JDM not only affects muscles but also muscle fascia (28-30). A previous study revealed that the FT of JDM patients was thicker than that of normal individuals (31). However, correlations between the JDM FT and disease activity, as well as the degree of FT change, have not been reported. The results of the present study revealed that the FT during the active period was significantly greater than that during the stable period (median 0.25 *vs.* 0.15, P<0.01). Thus, FT could be used to diagnose JDM and assess disease activity. The sensitivity and specificity of the FT were 92.3% and 93.1%, respectively, making it the second most important ultrasound evaluation index after EI. This study revealed that muscle FT was negatively correlated with the CMAS scores, which indicates that muscle FT can be used to assess the condition of JDM patients. In addition, it was observed during follow-up that even when EI returned to normal, the muscle FT did not fully return to normal. Further follow-up research is needed to explore whether fascial inflammation can fully recover, or if it resolves later than muscle inflammation.

Compared with MRI, ultrasound can be used to dynamically observe muscle blood perfusion in real time. Muscle blood perfusion is closely related to the pathological characteristics of inflamed muscle tissue and the degree of disease activity (32). Ultrasound MVFI technology has greater sensitivity in detecting blood flow, especially in slow microvessels (33). MVFI is highly consistent with contrast-enhanced ultrasound in showing small blood vessels with a diameter of 100 μm (34,35). JDM is a small vessel vasculitis disease (36), and MVFI can more sensitively detect increased blood perfusion in small vessels that are affected during the active period of JDM. The literature also indicates that microcapillary changes in nail fold changes are closely related to disease activity (37). In this study, the MVFI distribution was significantly increased in some children with active JDM, and the MVFI distribution was correlated with the children's muscle strength score and laboratory activity indicators. Comparisons between children with increased blood flow and those without increased blood flow revealed significant differences in CK and LDH, indicating that increased microvascular blood flow in children with JDM may indicate that the child is in an active disease state. However, the small sample size of this group of children might have introduced bias into the analysis.

This study also included corrected MT Z scores and muscle blood flow RI values. The results revealed that there was no significant difference in MT Z scores between patients in the active stage of JDM and those in the stable stage of JDM, which is consistent with the findings of Bansing (14). JDM is a small vessel vasculitis disease, and this study explored the differences in blood flow dynamics between muscle inflammation states and noninflammatory states. The results revealed that there was no significant difference in blood flow dynamics between the active and stable stages. The reason for this may be that children with JDM in the active period were at different stages of the disease at the time of diagnosis. Additionally, differences in MT Z scores and muscle blood perfusion were observed in children in the active period. Future longitudinal dynamic observations are needed to determine the common changes in MT and hemodynamics.

EI, FT, and MVFI distribution were all correlated with CMAS scores and laboratory indicators (i.e., CK, LDH, and α-HDBH) and thus can be used for JDM diagnosis. EI was the most sensitive indicator with an AUC of 0.99 and a sensitivity and specificity of 96.2% and 96.6%, respectively. Some stable JDM children still had greater EI values than healthy children, indicating that ultrasound may be more sensitive than clinical and laboratory indicators. FT can be used as an ultrasound index, and it may be related to JDM, and FT was second only to EI. Microvascular blood flow is more sensitive indicator in children with higher laboratory indicators associated with JDM, possibly MVFI is indicating a more active state of the disease.

In the follow-up of five children with active stage JDM, the results showed that ultrasound examination was suitable for the dynamic follow-up observation of JDM patients. FT recovered later than EI in these patients. Among the five children, two with persistent fasciitis later developed

subcutaneous soft tissue calcification. Calcification in JDM is a terrible problem, and it is not known when it will occur or how it can be predicted. More research needs to be conducted to determine if persistent fasciitis can predict the occurrence of calcification. Clinical attention should be given to such children, and further studies need to be conducted to improve the understanding of this disease.

The limitations of this study include that it was a single-center study with a relatively small sample size and short follow-up periods. Currently, the study is in the preliminary exploratory stage. After clarifying the diagnostic and follow-up value of ultrasound in JDM, collaborations with other centers will be explored to expand the sample size, conduct longer-term follow-up observations of cases, and further refine the clinical and laboratory indicators. This will enable a more in-depth exploration of JDM diagnosis and subsequent follow-up. In addition, the results of ultrasound examinations are closely related to each operator's experience, which may lead to poor reproducibility; thus, we hope to explore more quantitative indicators of JDM activity to avoid misdiagnosis caused by inexperience.

Conclusions

Ultrasound can serve as an effective diagnostic tool for the management of JDM in children. EI, FT, and MVFI distribution can be used as indicators of the activity of JDM. Additionally, EI and FT can reflect the severity of a child's condition to some extent, making them more accessible and cost-effective diagnostic and assessment tools for JDM.

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Footnote

Reporting Checklist: The authors have completed the STROBE reporting checklist. Available at https://qims.amegroups.com/article/view/10.21037/qims-24-1035/rc

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-24-1035/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. This study was approved by the Ethics Committee of the Capital Pediatric Research Institute (No. SHERLL2023039), and informed consent was obtained from the patients' parents or legal guardians. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013).

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