NEAR-INFRARED SPECTROSCOPY DURING EXERCISE AND RECOVERY IN CHILDREN WITH JUVENILE DERMATOMYOSITIS

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ABSTRACT: Introduction: We hypothesized that microvascular disturbances in muscle tissue play a role in the reduced exercise capacity in juvenile dermatomyositis (JDM). Methods: Children with JDM, children with juvenile idiopathic arthritis (clinical controls), and healthy children performed a maximal incremental cycloergometric test from which normalized concentration changes in oxygenated hemoglobin (ΔO2Hb) and total hemoglobin (ΔHb) as well as the half-recovery times of both signals were determined from the vastus medialis and vastus lateralis muscles using near-infrared spectroscopy. Results: Children with JDM had lower ΔHb values in the vastus medialis at work rates of 25%, 50%, 75%, and 100% of maximal compared with healthy children; the increase in ΔHb with increasing intensity seen in healthy children was absent in children with JDM. Other outcome measures did not differ by group. Conclusions: The results suggest that children with JDM may experience difficulties in increasing muscle blood volume with more strenuous exercise.


Juvneile dermatomyositis (JDM) is a pediatric autoimmune disease that affects approximately 3.2 per 1,000,000 children each year.1 Although the etiology of the disease remains largely unknown, environmental and genetic factors are thought to play a role in its development.2 Clinically, children with JDM commonly present with chronic inflammation of the microvasculature of the skin and skeletal muscles, which manifests as symmetrical proximal muscle weakness, muscle fatigue, and a characteristic rash (heliotrope rash over the eyelids and Gottron papules over the extensor joint surfaces).3

Recent reports have suggested that children with JDM may also suffer from severely diminished anaerobic and aerobic exercise capacity,4–8 a finding that may be linked to microvascular disturbances in muscle tissue.9 In fact, immunostaining for the membrane attack complex in muscle microvasculature was shown to be strong in JDM and weak in healthy controls.10–14 Furthermore, the microvascular endothelial cells and basal lamina appeared to be thickened in children with JDM.13,14 Taken together, these pathologic changes in the capillaries may induce hypoxia in the affected muscle tissue, which in turn would result in necrosis of the local muscle fibers.15 The active neovascularization found in JDM15 and the reported impairment in oxidative metabolism, as evidenced by a reduced rate of H+ efflux during recovery from exercise,16 further support the concept of reduced oxygen supply to the muscles.

Although microvascular disturbances are thought to play a central role in exercise intolerance in children with JDM, little evidence exists to support this hypothesis. Near-infrared spectroscopy (NIRS) may be a useful tool to assess this theory, because it allows for in vivo examination of oxygenation and hemodynamics in muscle tissue at the microvascular level during physical exercise. NIRS is a non-invasive and relatively inexpensive technology based on the relative transparency of biological tissue to light in the near-infrared region. More specifically, NIRS relies on the oxygen-dependent absorption of hemoglobin (Hb) in the near-infrared range, whereby deoxygenated Hb (HHb) absorbs more light compared with oxygenated hemoglobin (O2Hb) at wavelengths <800 nm, and less light compared with O2Hb at wavelengths >800 nm. According to the modified Lambert Beer law, the concentration changes in O2Hb and HHb can be calculated when the appropriate light wavelengths are chosen. Previous studies have shown that NIRS can detect abnormal muscle oxygenation and hemodynamics during exercise and recovery in adults and children with varying levels of exercise intolerance.17

Although NIRS has not yet been studied in children with JDM, its application is promising, as it may provide researchers and clinicians with a more profound physiological insight into the exercise intolerance commonly reported in this disease. This insight may subsequently be used to inform and improve treatment, medication use, and exercise therapy.
The objective of this study was to use NIRS to measure and examine muscle oxygenation and hemodynamics during exercise and recovery in children with JDM and compare them with children in a clinical control group and healthy children.

Concentration changes compared with rest of \( [\Delta \text{O}_2\text{Hb}] \) and total hemoglobin (\( [\text{Hb}] \); \( [\text{Hb}] = [\text{O}_2\text{Hb}] + [\text{HHb}] \)) of the vastus medialis (VM) and vastus lateralis (VL) muscle were determined with NIRS during incremental cycling exercise and recovery in the 3 groups of children.

METHODS

Subjects. A group of 11 children with JDM, a clinical control group of 10 children with juvenile idiopathic arthritis (JIA), and a control group of 13 healthy children participated in this cross-sectional pilot study. Children with JIA were included as a clinical control group because they tend to be physically inactive, much like children with JDM; however, unlike children with JDM, they do not commonly present with muscle disease.

Children with JDM and JIA between the ages of 8 and 19 years were recruited from the Pediatric Rheumatology Clinics at the Wilhelmina Children’s Hospital, University Medical Center Utrecht, The Netherlands. All patients were diagnosed to have JDM by a pediatric rheumatologist/immunologist according to the Bohan and Peter criteria\(^{18,19}\) or JIA according to the ILAR criteria.\(^{20}\) Within the JDM group, both patients with active disease (evidence of active myositis) and patients in remission (no evidence of active myositis) were included. Children were consecutively approached by the researcher to participate in the study. Of the initially eligible children, all of the children with JDM and 10 of 13 of the children with JIA agreed to participate. Healthy children were recruited by flyers posted on notice boards at the hospital. Children in the JDM and JIA groups were tested during a regular follow-up visit, whereas children in the healthy group were invited to attend 1 session at the Wilhelmina Children’s Hospital.

Exclusion criteria for all children included a medical status that contraindicated any form of exercise testing and/or an insufficient understanding of the Dutch language in the child and/or the parents/caregivers. Within the group of healthy children, additional exclusion criteria included a history of muscle disease, chronic medication use, and/or the presence of an underlying autoimmune disease. All parents/caregivers as well as children (when aged >12 years) provided informed consent prior to participating in the study. The study was approved by the medical ethics committee of the University Medical Center Utrecht.

Anthropometry. Height (cm) and body mass (kg) of the children were determined using a wall-mounted stadiometer and an electronic scale, respectively. Body mass index was calculated as weight/height\(^2\) (kg m\(^{-2}\)).

Cardiopulmonary Exercise Test. All children performed an incremental cardiopulmonary exercise test (CPET) on an electronically braked cycle ergometer (Lode Corival; Lode BV, Groningen, The Netherlands). The height of the saddle was adjusted to allow slight flexion in the knee joints when the feet were in the lowest pedal position. Before the start of exercise, the child rested on the cycle ergometer for 5 min or until a steady state was attained in the respiratory as well as the NIRS signals. During this time, the leg to which the NIRS device was secured was positioned at the lowest pedal position. Leg movement was restricted by instructing the child to remain as static as possible. After these resting measurements, the child was asked to start cycling with a 3-min unloaded warm-up, after which the work rate was increased each 12 s according to the Godfrey protocol by 10, 15, or 20 watts (W) min\(^{-1}\), depending on the child’s height.\(^{21}\) The child was instructed to maintain a cadence of between 60 and 80 revolutions min\(^{-1}\). The test was terminated when the child was no longer able to maintain the recommended cadence because of volitional exhaustion, despite strong verbal encouragement of the test leader. Immediately after test completion, a 5-min recovery period followed, during which the child returned to the identical seated position as during the resting measurements. This identical position particularly important, because pooling of the blood in the legs is significantly dependent on leg position.\(^{22}\)

During the CPET, the child wore a small face-mask (Hans Rudolph, Inc., Kansas City, Missouri) that was connected to a gas-analysis system (ZAN600; nSpire Health, Inc., Oberthulba, Germany). Volume calibration and gas-analysis calibration were done before each testing session. Breath-by-breath \( \text{VO}_2 \) and \( \text{VCO}_2 \) were calculated from expired gases and corrected for the dead space of the mask. The raw breath-by-breath data were averaged and stored over a 10-s interval.

Arterial oxygen saturation (%) was measured by pulse oximetry (Nellcor OxiMax, N-600x Pulse Oximeter; Covidien-Nellcor, Boulder, Colorado) at the index finger to ascertain that there was no clinically relevant arterial desaturation (arterial oxygen saturation <90%) during exercise. A blood pressure cuff (SuriTech Medical Instruments, Morrisville, North Carolina) was used to assess blood pressure (mm Hg) at rest, every 2 min during
exercise, at the end of exercise, and at the end of recovery. Heart rate (beats min\(^{-1}\)) was continuously measured using a 12-lead electrocardiogram (CardioPerfect; Welch Allyn, Delft, The Netherlands).

Outcome measures related to the CPET included respiratory exchange ratio (= \(\text{VCO}_2/\text{VO}_2\)), peak heart rate (beats min\(^{-1}\)), relative peak work rate (W kg\(^{-1}\)) as percentage of predicted, relative peak \(\text{O}_2\) uptake (ml kg\(^{-1}\) min\(^{-1}\)) as percentage of predicted, oxygen uptake (L min\(^{-1}\)) at ventilatory anaerobic threshold as percentage of peak oxygen uptake, relative oxygen uptake (ml kg\(^{-1}\) min\(^{-1}\)) at ventilatory anaerobic threshold as percentage of predicted relative peak oxygen uptake, and oxygen uptake to work rate slope (ml min\(^{-1}\) W\(^{-1}\)). Predicted values were obtained from ten Harkel et al.\(^{23}\)

**NIRS—Measurements.** The probes of a single-distance, continuous-wave photometer (OXYMON; Artinis, Zetten, The Netherlands) with 2 channels were fixed to the VM and VL muscles of the child’s leg. In children with JDM, the leg most affected by the disease was selected for the probes (in 6 subjects this was the right leg and in 5 subjects the left leg), whereas the dominant leg was chosen for the children in the JIA and healthy groups. The VL probe was placed at one third of the distance from the lateral epicondyle to the greater trochanter of the femur, whereas the VM probe was placed medially using the same transversal landmarks as the VL probe. The probes were fixed to the skin with tape to prevent probe shifting during movement. A black cloth was placed over the optode holders to reduce the intrusion of stray light and the loss of transmitted light from the field of examination. An additional elastic bandage was then wrapped around the leg to further minimize movement of the probes. No shifting of the probes occurred during the measurements.

Near-infrared light was emitted at 2 wavelengths (775 nm and 850 nm). The light source and detector were housed in the same holder, with a fixed source-detector distance of 3.0 cm, which means an average measurement depth of approximately 1.5 cm.\(^{24}\) To ascertain that the measurement depth was sufficient to reach the muscle tissue, skinfold plus adipose tissue thickness (SATT) at the location of the probes was measured with a skinfold caliper (Harpenden; Baty International, Burgess Hill, UK).

In this study, \(\Delta[\text{O}_2\text{Hb}]\) and \(\Delta[\text{tHb}]\) values were analyzed during cycling and recovery. \(\Delta[\text{O}_2\text{Hb}]\) reflects the change in balance between oxygen delivery (depending on blood velocity, blood volume, and arterial saturation) and oxygen consumption in the portion of tissue under consideration. \(\Delta[\text{tHb}]\) reflects the change in blood volume in the portion of tissue under consideration. Throughout the assessment, both \(\Delta[\text{O}_2\text{Hb}]\) and \(\Delta[\text{tHb}]\) were sampled and displayed in real time at a frequency of 50 Hz. The data were subsequently filtered by a Gaussian smoothing window (1-s width), down-sampled to 2 Hz, and exported to Excel for Windows (Microsoft, Inc., Redmond, Washington) for additional analysis.

**NIRS—Normalization.** NIRS outcome measures were normalized, as shown in Figure 1, to allow comparisons between children of the same group, as well as between groups. This normalization procedure involved averaging the values of \(\Delta[\text{O}_2\text{Hb}]\) and \(\Delta[\text{tHb}]\) over the last 30 s of the resting period before the exercise and assigning them a value of 0 arbitrary unit (AU). Next, \(\Delta[\text{O}_2\text{Hb}]\) and \(\Delta[\text{tHb}]\) during the recovery phase were averaged over a 10-s period, and the maximal values were assigned a value of 1 AU. All values between these 2 time-points were normalized to this scale. (B) \(\Delta[\text{O}_2\text{Hb}]\) and \(\Delta[\text{tHb}]\) during unloaded cycling, loaded cycling, and recovery after normalization. The normalized \(\Delta[\text{O}_2\text{Hb}]\) and \(\Delta[\text{tHb}]\) values averaged over 10 s were determined at the end of unloaded cycling and at work rates of 25%, 50%, 75%, and 100% of \(W_{\text{peak}}\).
Half-recovery times (T_{half} [s]) of Δ[O₂Hb] and Δ[tHb] were assessed during recovery.\(^{25-27}\) The T_{half} of both signals was defined as the time needed to reach half-recovery from the values at the end of exercise to 1 AU in recovery.

Statistical Analyses. Between-group differences were examined for the anthropometric, CPET, and NIRS outcome measures. Analyses for the NIRS outcome measures for the VM and VL muscles were done separately. Shapiro–Wilk tests of the residuals and Levene’s tests were used to assess normality and homogeneity of variance, respectively. All normally distributed variables with homogeneity of variance were analyzed by one-way between-group analysis of variance (ANOVA). If the residuals were normally distributed with unequal variance, Welch’s ANOVA was performed. Bonferroni’s post hoc tests were used when appropriate. Normally distributed data are presented as mean ± standard deviation (SD).

Any variables that failed the normality test were analyzed using the Kruskal–Wallis test. Post hoc Mann–Whitney U-tests with Bonferroni’s correction were used when appropriate. Non–normally distributed data are presented as median and interquartile range (IQR).

Eta-squared (η²) was used to determine the effect size. All statistical analyses were done using SPSS, version 15.0 (SPSS, Inc., Chicago, Illinois), with statistical significance set at \(p < 0.05\).

RESULTS
In total, 11 children with JDM, 10 children with JIA, and 13 healthy children completed the study. Characteristics of the children are depicted by group in Table 1.

CPET. All patients completed the CPET without complications or arterial desaturation. All but 1 (JDM) child achieved peak respiratory exchange ratio values \(>1.05\) (mean ± SD: 1.15 ± 0.09). Peak heart rate values were between 149 and 207 beats min\(^{-1}\) (188 ± 11 beats/min). The primary CPET parameters are presented by group in Table 2.

NIRS. Δ[tHb] values in the VM muscle were statistically significantly affected by group at work rates of 25\% [\(p < 0.05\); \(\chi^2(2) = 6.55\); \(\eta^2 = 0.20\)], 50\% [\(p = 0.01\); \(\chi^2(2) = 9.19\); \(\eta^2 = 0.28\)], 75\% [\(p < 0.01\); \(\chi^2(2) = 9.91\); \(\eta^2 = 0.30\)], and 100\% [\(p < 0.05\); \(\chi^2(2) = 6.15\); \(\eta^2 = 0.19\)] of W\(_{\text{peak}}\). The concerning Δ[tHb] values were significantly (\(p < 0.01\)) lower in children with JDM compared with healthy children at all these relative work rates (Table 3A).

In healthy children, the median Δ[tHb] value in the VM muscle was 0 AU at a work rate of 25\% of W\(_{\text{peak}}\), and this value increased with work rates.

### Table 1. Characteristics of the children presented by group.

<table>
<thead>
<tr>
<th>Outcome measure</th>
<th>JDM (n = 11)</th>
<th>JIA (n = 10)</th>
<th>Healthy (n = 13)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years), mean (minimum–maximum value)</td>
<td>14 (8–19)</td>
<td>14 (9–17)</td>
<td>13 (8–18)</td>
</tr>
<tr>
<td>Gender (boys/girls)</td>
<td>7/4</td>
<td>4/6</td>
<td>7/6</td>
</tr>
<tr>
<td>Height (cm), mean ± SD</td>
<td>161 ± 18</td>
<td>163 ± 11</td>
<td>158 ± 18</td>
</tr>
<tr>
<td>Body mass (kg), mean ± SD</td>
<td>53 ± 15</td>
<td>51 ± 13</td>
<td>45 ± 14</td>
</tr>
<tr>
<td>Body mass index (kg m(^{-2})), median (IQR)</td>
<td>19 (5)</td>
<td>18 (4)</td>
<td>17 (3)</td>
</tr>
<tr>
<td>SATT VM (mm), mean ± SD (range)</td>
<td>9 ± 5 (14)*</td>
<td>8 ± 4 (12)</td>
<td>6 ± 2 (5)</td>
</tr>
<tr>
<td>SATT VL (mm), mean ± SD (range)</td>
<td>11 ± 5 (15)*</td>
<td>8 ± 4 (15)</td>
<td>7 ± 2 (7)</td>
</tr>
</tbody>
</table>

None of the variables differed statistically significantly between the three groups.

*Three missing values.

### Table 2. The primary CPET parameters presented by group and relevant results of the statistical analyses.

<table>
<thead>
<tr>
<th>Outcome measure</th>
<th>JDM (n = 11)</th>
<th>JIA (n = 10)</th>
<th>Healthy (n = 13)</th>
<th>Statistic</th>
<th>η²</th>
</tr>
</thead>
<tbody>
<tr>
<td>%W_{\text{peak}}, kg(^{-1}), mean ± SD</td>
<td>80 ± 24(^{\text{JDM}})</td>
<td>110 ± 16(^{\text{JDM}})</td>
<td>127 ± 15(^{\text{JDM}})</td>
<td>(\chi^2(2) = 9.44)</td>
<td>0.56</td>
</tr>
<tr>
<td>%VO₂_{\text{peak}}, kg(^{-1}), mean ± SD</td>
<td>70 ± 24(^{\text{JDM}})</td>
<td>95 ± 14(^{\text{JDM}})</td>
<td>111 ± 12(^{\text{JDM}})</td>
<td>(\chi^2(2) = 7.62)</td>
<td>0.53</td>
</tr>
<tr>
<td>%VAT_{\text{peak}}, kg(^{-1}), mean ± SD</td>
<td>52 ± 13</td>
<td>62 ± 9</td>
<td>56 ± 10</td>
<td>(\chi^2(2) = 2.22)</td>
<td></td>
</tr>
<tr>
<td>%VAT_{\text{peak}}, median (IQR)</td>
<td>37 (19(^{\text{JIA}}))</td>
<td>58 (14(^{\text{JDM}}))</td>
<td>60 (17(^{\text{JDM}}))</td>
<td>(\chi^2(2) = 10.10)</td>
<td>0.34</td>
</tr>
<tr>
<td>ΔVO₂/ΔW (ml min(^{-1}) W(^{-1})), mean ± SD</td>
<td>9.8 ± 1.6</td>
<td>9.3 ± 1.3</td>
<td>9.9 ± 1.2</td>
<td>(\chi^2(2) = 2.05)</td>
<td></td>
</tr>
</tbody>
</table>

\(\%W_{\text{peak}}, kg\(^{-1}\):\) relative peak work rate as percentage of predicted; \(\%VO₂_{\text{peak}}, kg\(^{-1}\):\) relative peak oxygen uptake as percentage of predicted; \(\%VAT_{\text{peak}}, kg\(^{-1}\):\) oxygen uptake at ventilatory anaerobic threshold as percentage of predicted \(\%VAT_{\text{peak}}, kg\(^{-1}\):\) relative oxygen at ventilatory anaerobic threshold as percentage of predicted \(\chi^2(2) = 6.55; \eta^2 = 0.20\).

*Statistically significantly different from healthy.
*JDMStatistically significantly different from JIA.
*\(p < 0.05\); †\(p < 0.01\); ‡\(p < 0.001\).
at higher percentages of $W_{\text{peak}}$, indicating an increase in blood volume with increasing intensity. Conversely, the median $D[t\text{Hb}]$ value in the VM muscle in children with JDM was $<$0 AU at a work rate of 25% of $W_{\text{peak}}$, and this value remained $<$0 AU throughout exercise. Figure 2 shows a representative sample of $\Delta[t\text{Hb}]$ during exercise in a child with JDM and in a healthy child. Although children with JIA displayed a median $D[t\text{Hb}]$ of $>$0 AU at a work rate of 100% of $W_{\text{peak}}$ in the VM muscle, these children did not differ significantly from the children in the JDM group.

No statistically significant differences were found between the groups for either $D[t\text{Hb}]$ in the VL muscle at all time-points, or $D[O_2\text{Hb}]$ in both muscle groups at all time-points (Tables 3A and 3B). Similarly, no differences were seen between the groups for $T_{\text{half}}$ of $D[O_2\text{Hb}]$ and $T_{\text{half}}$ of $\Delta[t\text{Hb}]$ for both the VM and VL muscle (Table 4).

**DISCUSSION**

The objective of this study was to use NIRS to measure and examine muscle oxygenation and hemodynamics during exercise and recovery in children with JDM compared with children in a clinical control group and healthy children.

**NIRS during Cycling.** The findings suggest that children with JDM may experience difficulties in increasing muscle blood volume with more strenuous exercise. More specifically, the initial drop in median $\Delta[t\text{Hb}]$ in the VM muscle observed in all 3 groups was not followed by an increase to values $>$0 AU with increasing exercise intensity in children with JDM, in contrast to the children in the control group. This observation is in line with the hypothesis of the significant role that microvascular disturbances play in the exercise intolerance seen in children with JDM. However, the NIRS observations should be confirmed by complementary techniques such as magnetic resonance spectroscopy.28

Although the $\Delta[t\text{Hb}]$ values in the VM muscle of children with JDM at work rates of 25%, 50%, 75%, and 100% of $W_{\text{peak}}$ differed significantly from those of healthy children, it should be noted that the decreased $\Delta[t\text{Hb}]$ values were neither exclusive to children with JDM, nor were they observed in each of the children with JDM. In fact, some healthy children, as well as a few children with JIA, also demonstrated $D[t\text{Hb}]$ values $<$0 AU in the VM muscle throughout the whole exercise period. Therefore, at the individual level, the time course for $\Delta[t\text{Hb}]$ in the VM muscle did not fully discriminate between children with JDM and unaffected children.

**NIRS Outcome Measures.** Because the differential path length factor in the modified Lambert–Beer law equation is not known and may differ between children because of differences in optical properties under the optodes (i.e., differences in SATT), the recorded values of $\Delta[O_2\text{Hb}]$ and $\Delta[t\text{Hb}]$ could not be compared directly between children of the same group, nor between groups. We solved this problem by analyzing normalized data from the exercise period and by looking at time characteristics from the recovery period.

**Vastus Medialis vs. Vastus Lateralis Muscle.** Differences in $\Delta[t\text{Hb}]$ during cycling were found in the VM muscle but not in the VL muscle. This finding

![Figure 2](image-url)
NIRS in Juvenile Dermatomyositis

could presumably not be explained by differences in SATT between both muscles, because the SATT was not significantly different between the muscles, as demonstrated with the paired-samples t-test. Although the VM and VL muscle have the same time pattern of activation during cycling, the VM muscle is possibly recruited more during cycling. Another reason for the fact that differences in blood flow. However, this would be unexpected, because the patients with JDM did not show any active skin rash at the measurement sites during the measuring. Furthermore, blood flow through skin tissue is expected to not change during a short exercise period, which indicates that the increase in total hemoglobin concentration found in healthy subjects came exclusively from an increase in blood volume in muscle tissue. Whether skin blood flow, muscle blood flow, or both are impaired in JDM during exercise should be examined further with techniques like laser Doppler imaging.

The potential confounding effect of the use of prednisone and methotrexate on blood volume is probably limited in patients with JDM and JIA. Some of the patients with JDM had stabilized disease and were being treated with low-dose prednisone (0.10–0.25 mg kg\(^{-1}\) day\(^{-1}\)), without concomitant clinical signs, and with low-dose methotrexate (10 mg m\(^{-2}\) week\(^{-1}\) orally). The others were in prolonged remission with no medication for >6 months. The patients with JIA also had stabilized disease on maintenance therapy with non-steroidal anti-inflammatory drugs and without methotrexate.

**Skin and Adipose Tissue.** The fact the NIRS assesses not only oxygenation and hemodynamics in muscle tissue but also in overlying skin and adipose tissue may have significant implications for the calculated \(\Delta[O_2\text{Hb}]\) and \(\Delta[t\text{Hb}]\), because the different tissues have different optical characteristics as well as different metabolic demands during exercise.

van Beekvelt et al. examined the effects of SATT on oxygen consumption and blood flow measured with NIRS during arterial occlusion and venous occlusion, respectively, in healthy subjects. They found that SATT was negatively associated with muscle oxygen consumption, but it was positively correlated with blood flow. However, in our study, no significant relation was found between the SATT and the NIRS outcome measures, according to regression analyses. Furthermore, the SATT of the VM and VL muscle did not differ significantly between the 3 groups, as assessed by one-way between-group ANOVAs (see Table 1).

Because changes in muscle blood volume as well as changes in skin blood volume are attributable to the measured \(\Delta[t\text{Hb}]\) values, the altered blood volume changes we found in patients with JDM could be due simply to impairment of skin blood flow. However, this would be unexpected, because the patients with JDM did not show any active skin rash at the measurement sites during the measuring. Furthermore, blood flow through skin tissue is expected to not change during a short exercise period, which indicates that the increase in total hemoglobin concentration found in healthy subjects came exclusively from an increase in blood volume in muscle tissue. Whether skin blood flow, muscle blood flow, or both are impaired in JDM during exercise should be examined further with techniques like laser Doppler imaging.

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**NIRS Measurements during Exercise in other Myopathies.** The results of a recent study in adult patients with polymyositis correspond to the findings of this study. In that adult study, total Hb concentration during gripping exercise was decreased compared with rest in the patients, whereas no such decrease was seen in the healthy subjects. The similarity of those results with the current results is of interest, because microvascular disturbances in muscle tissue are believed to be present in patients with polymyositis, much like in children with JDM.

**Clinical Implications.** The findings indicate that children with JDM experience difficulty in increasing muscle blood volume during exercise.

<table>
<thead>
<tr>
<th>Work rate [% of (W_{peak})]</th>
<th>VM muscle</th>
<th>JDM (n = 11)</th>
<th>JIA (n = 10)</th>
<th>Healthy (n = 13)</th>
<th>VL muscle</th>
<th>JDM (n = 11)</th>
<th>JIA (n = 10)</th>
<th>Healthy (n = 13)</th>
</tr>
</thead>
<tbody>
<tr>
<td>End unloaded cycling</td>
<td>[-0.2 (0.3)]</td>
<td>[-0.3 (0.4)]</td>
<td>[-0.3 (0.2)]</td>
<td>[0.0 (0.6)]</td>
<td>[0.0 (0.4)]</td>
<td>[0.0 (0.2)]</td>
<td>[0.0 (0.4)]</td>
<td>[0.0 (0.2)]</td>
</tr>
<tr>
<td>25</td>
<td>[-0.2 (0.4)]</td>
<td>[-0.3 (0.5)]</td>
<td>[-0.1 (0.3)]</td>
<td>[0.0 (0.5)]</td>
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<td>[0.0 (0.4)]</td>
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<tr>
<td>50</td>
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<td>[-0.3 (0.5)]</td>
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<td>[-0.3 (0.3)]</td>
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<td>[-0.1 (0.3)]</td>
<td>[-0.4 (0.4)]</td>
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<td>75</td>
<td>[-0.5 (0.8)]</td>
<td>[-0.2 (0.7)]</td>
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<td>100</td>
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<td>[-0.5 (1.0)]</td>
<td>[-0.4 (0.6)]</td>
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<td>[-0.5 (0.7)]</td>
<td>[-0.3 (0.4)]</td>
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</tbody>
</table>

**Table 3B.** Median \(\Delta[O_2\text{Hb}]\) values [AU] in the VM and VL muscle at end unloaded cycling and at work rates of 25%, 50%, 75%, and 100% of \(W_{peak}\), presented by group as median (IQR).
Therefore, it may be advisable to focus treatment on improving the blood flow and hence oxygen delivery to the muscle tissue.

Earlier studies showed that NIRS is a useful tool for studying the effects of steroid therapy in patients with myositis. Among these studies, Okuma et al. examined patients with polymyositis and demonstrated a decrease in muscle blood flow during a gripping exercise before treatment, which was reversed after steroid therapy.34 Another study showed that, in untreated adult patients with dermatomyositis, oxygen consumption measured in muscle tissue during arterial occlusion was approximately 60% lower compared with controls, whereas recovery of \( \Delta[O_2\text{Hb}] \) was significantly slowed over a wide range of relative exercise intensities. After therapy, oxygen consumption was markedly increased, and \( \Delta[O_2\text{Hb}] \) recovery rates rose above the recovery rates measured in controls.35

The findings from the aforementioned studies highlight the potential for NIRS as a non-invasive and inexpensive clinical tool for longitudinal assessment of the effect of treatment on exercise intolerance in JDM. It is important to note, however, that only adult populations were assessed in previous works. Furthermore, in the study by van Beekvelt et al.,35 arterial occlusion was performed, which is considered an unethical procedure in children. Thus, further investigation into the role of NIRS as a longitudinal clinical tool in children with JDM is warranted.

In conclusion, children with JDM differed from healthy children with respect to change in total Hb concentration of the VM muscle from rest to incremental exercise. More specifically, healthy children demonstrated an increase in total Hb concentration above resting values throughout the exercise, whereas in JDM patients, this increase was absent, and total Hb concentration remained lower compared with rest throughout the entire exercise phase. This finding suggests that children with JDM may have difficulty with increasing muscle blood volume with more strenuous exercise. Future studies should confirm and specify these observations using other techniques, such as laser Doppler imaging or magnetic resonance imaging.

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