

Intramuscular dissociation of echogenicity in the triceps surae characterizes sporadic inclusion body myositis

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ABSTRACT

Background and purpose: Differential diagnosis of sporadic inclusion body myositis (s-IBM) and polymyositis (PM)/dermatomyositis (DM) is difficult and can affect proper disease management. Detection of heterogeneous muscular involvement in s-IBM by muscle sonography could be a unique diagnostic feature.

Methods: Sonography of the lower leg and forearm was performed in patients with s-IBM, PM/DM, and control subjects (n = 11 each). Echo-intensities (EIs) of the adjacent muscles (medial head of the gastrocnemius vs. soleus and the flexor digitorum profundus [FDP] vs. flexor carpi ulnaris [FCU]) were scored by three blinded raters. The mean EIs of these muscles were compared using computer-assisted histogram analysis.

Results: Both evaluation methods showed high echoic signals in the gastrocnemius of patients with s-IBM. EIs were significantly different between the gastrocnemius and soleus in patients with s-IBM, but not in those with DM/PM and the controls. In the forearm, although EI of the FDP was higher in the s-IBM group than in the other groups, the EI differences between the FDP and FCU did not differ significantly between disease groups. The difference in area under the curves (AUCs) to differentiate between s-IBM and DM/PM was greatest between the gastrocnemius-soleus EIs (0.843; $P = 0.006$).

Conclusions: High echoic signals in the medial gastrocnemius compared with those of the soleus are suggestive of s-IBM over PM/DM.

Key words: sonography, inclusion body myositis, triceps surae, echogenicity, histogram

Abbreviations:

AUC = area under the curve

DM = dermatomyositis

EI = echo-intensity

FCU = flexor carpi ulnaris

FDP = flexor digitorum profundus

GC = gastrocnemius

HS = Heckmatt rating scale

ICC = intraclass correlation coefficients

PM = polymyositis

ROC = receiver operating curve

ROI = region of interest

s-IBM = sporadic inclusion body myositis

Contributions

HN was involved in design of the study, analysis and interpretation of the data and drafting of the manuscript. NT was involved in design of the study and acquisition of the data. NM was involved in interpretation of the data. AM, YT, YS, YO, KM were involved in acquisition and analysis of the data. YI and RK were involved in design of the study and revision of the manuscript

Introduction

Sporadic inclusion body myositis (s-IBM) is a muscle disease that predominantly affects older individuals [1]. Diagnosis of typical s-IBM is straightforward with its characteristic preferential involvement of the deep finger flexors and knee extensors [2]. However, atypical presentations complicate its diagnosis and could lead to misdiagnosis as amyotrophic lateral sclerosis, polymyositis, or muscular dystrophy [3, 4]. Misdiagnosis can result in unnecessary therapies such as prolonged courses of steroids, as well as their adverse effects. Therefore, it would be useful to identify a measure to differentiate s-IBM from its mimics.

Muscular system imaging is useful for diagnosis and severity assessment of myopathic conditions [5]. Among modalities, MRI and sonography have been the most widely studied. Compared to muscle MRI, muscle sonography has the following advantages: (1) few contraindications, and (2) ability to test patients who are unable to be transported for MRI at their bedside.

Recently, Noto and colleagues observed heterogeneous involvement and reported dissociation of echoic signal intensities in patients with s-IBM, finding greater signal intensities in clinically more affected flexor digitorum profundus (FDP) than in less affected flexor carpi ulnaris (FCU) [6]. Cox and colleagues assessed muscle MRI findings in patients with s-IBM and reported the degrees of fatty infiltration of each limb muscle [7]. Intriguingly, the medial head of gastrocnemius muscle had the most extensive fatty infiltration, while the soleus, another component of the triceps surae, was only moderately affected. This finding suggests that dissociation of the head of triceps surae may be another diagnostic feature of s-IBM. However, detailed sonographic comparison of this muscle in s-IBM and other myopathies is lacking.

Therefore, the aim of the present study was to compare the sonographic features of the triceps surae and forearm in patients with s-IBM and other inflammatory myopathies.

Methods

Subjects

Three groups were recruited and prospectively assessed: (1) patients with s-IBM who met clinical-pathologically defined IBM or clinically defined IBM criteria based on guidelines from the European Neuromuscular Center (ENMC), with no family history of related conditions [2]; (2) patients diagnosed with either PM, DM, or myositis associated with a connective tissue disease according to established diagnostic criteria [8]; and (3) control subjects: asymptomatic persons with no neurological symptoms or signs. This study was approved by the Institutional Review Board of Vihara Hananosato Hospital and Tokushima University. The subjects provided written informed consent at the time of testing.

Sonography

A single technician (N.T.) blinded to patient diagnoses performed sonography using a LOGIQ7 (GE Healthcare Japan, Tokyo) with a fixed 11-MHz linear-array transducer. The subjects were tested in the supine position and the right upper and lower limbs were studied. This unilateral assessment was chosen in order to avoid potential selection bias of highly correlated bilateral data from the same individual. Similarly, we decided to study only on the right side to avoid potential side-dependency of the sonographic parameters. By applying the transducer to the medial aspect of the mid-calf, the medial head of gastrocnemius was identified where the soleus was visualized just under the gastrocnemius, with a clear margin by high-echoic fascicles (Figure

1). Visualization of the flexor digitorum profundus (FDP) and flexor carpi ulnaris (FCU) was performed as previously reported, by flexing the elbow and placing the probe 5 cm distal to the olecranon under a single view (Supplemental Figure 1) [6].

Visual assessment of muscle echo intensity (EI) was scored by three examiners who routinely performed muscle sonography, but who were blinded to the diagnoses and scores of the other raters. Four muscles (the medial head of gastrocnemius, soleus, FDP, and FCU) were rated based on the modified Heckmatt rating scale as follows: 1, normal; 2, slightly increased muscle EI with normal epimysium reflection; 3, moderately increased muscle EI with reduced epimysium reflection; and 4, severely increased muscle EI with no clearly identifiable epimysium [9]. Because a bone may not be visualized in the lower leg, epimysium instead of bone was used as reference. The differences of the rating scales of the adjacent muscles (i.e., gastrocnemius vs. soleus and FDP vs. FCU) were calculated by comparing the scores of the respective muscles.

For quantitative assessment of EI, the mean pixel intensity of the muscle was measured by gray scale analysis using the standard histogram function in ImageJ, version 1.48 (National Institutes of Health, USA). A region of interest (ROI) was set in a polygonal manner to include as much of the respective target muscle as possible without covering any bone or surrounding fascia. The EI value in the ROI ranged between 0 (black) and 255 (white). The mean EI of each muscle was obtained and averaged in each disease and control group. After standardizing the area under the histogram for each histogram, the grand-averaged histogram was obtained and the mean EI values were compared.

Data analysis

SPSS version 20.0J (Tokyo, Japan) was used for statistical analysis, using one-way ANOVA with Games-Howell post-hoc test, receiver operating curves with the area under the curve, intraclass correlation coefficients (ICC), Cronbach's alpha, and Spearman's correlation coefficient where applicable. A *P* value of 0.05 was set as the threshold for statistical significance. A statistically significant *P* value was set at 0.05.

Results

Clinical characteristics

The subject characteristics are summarized in Table 1. Of 11 patients with s-IBM, six were classified as having clinico-pathologically defined IBM, the remaining five had clinically defined IBM. The patients in the PM/DM group had the following diagnoses and related conditions: PM (seven patients [three as definite PM and four as probable PM: three as an isolated condition and one each with scleroderma, rheumatoid arthritis, mixed connective tissue disease, primary biliary sclerosis]) and DM (four patients with definite DM). All patients with PM or DM responded to oral steroids; their scores on manual muscle testing of the bilateral biceps brachii or quadriceps improved at least 1 degree in the respective muscle by the modified Medical Research Council scale (4-, 4, and 4+). One patient with PM was positive for anti-signal recognition particle antibody. There was no significant difference in gender, but the mean age of the PM/DM group was less than in the s-IBM group ($P = 0.04$) (see Discussion). Serum creatine kinase (CK) levels tended to be greater in the PM/DM subjects than in the s-IBM subjects ($P = 0.06$). The mean disease duration of the muscle weakness tended to be longer in patients with s-IBM than those with PM/DM ($P = 0.07$).

Sonography

Representative sonographic images are shown in Figure 1 and Supplemental Figure 1. In control subjects, the echoic signals were similar between the adjacent muscles, such as the gastrocnemius vs. soleus and the FDP vs. FCU. Patients with s-IBM showed selective hyperechogenicity in the gastrocnemius (Figure 1B) and FDP (Supplemental Figure 1B), with relatively sparse EI in the neighboring soleus and FCU. In contrast, this selectivity was not evident in patients with PM/DM and these muscles tended to be equally hyperechoic; thus, the separating perimysium was not clearly identified (Figure 1C and Supplemental Figure 1C).

Table 2 summarizes the modified Heckmatt rating scale results in these muscles. In the controls, the muscles were rated as normal (score = 1) in most of the subjects, particularly in the forearm. However, patients with s-IBM and polymyositis had higher scores. Patients with s-IBM had higher scores in the gastrocnemius than patients with polymyositis ($P < 0.001$), but the scores in the soleus, FDP, and FCU were similar among groups. This resulted in significantly greater scores for the difference between the gastrocnemius and the soleus in patients with s-IBM (i.e., selective hyperechogenicity in the gastrocnemius in these patients). This difference was not present between the FDP and FCU in the forearm. Supplemental Table 1, shows the high reliability of measurements among the three raters (i.e., ICC [2.1] > 0.7 ; Cronbach's alpha > 0.8) for the gastrocnemius and FDP, followed by the soleus, and the lowest reliability in the FCU. Difference between modified Heckmatt rating scale scores were compared between adjacent muscles. In the IBM group, 81.8% of patients had higher scores in the gastrocnemius than in the soleus, unlike the PM/DM and control groups ($P = 0.0002$: IBM vs. PM/DM) (Supplemental Table 2). In the forearm, the FDP had greater scores than the FCU in 54.5% of patients in the

IBM group, but this frequency was not statistically significant compared to that of the PM/DM group ($P = 0.4$).

Histogram analysis

The averaged mean EIs are shown in Table 3 and Figure 2. Similar to the modified Heckmatt score, the EIs tended to be significantly higher in the patient groups than in the control group. Of interest, the gastrocnemius in the s-IBM patients showed the greatest EI among all tested muscles, followed by the FDP in the same group. The difference in EIs between neighboring muscles (i.e., gastrocnemius vs. soleus and FDP vs. FCU) in the s-IBM group was significantly greater in the leg, but not in the forearm. To compare the discriminatory power between s-IBM and polymyositis, receiver operating curve (ROC) analysis was performed (Supplemental Table 3). The difference between the gastrocnemius and soleus that resulted in the greatest area under the curve (AUC) ($AUC = 0.843$, $P = 0.006$) combined with the best EI cut-off value (43.6) resulted in 72.7% sensitivity and 100% specificity (IBM vs. PM/DM). Of note, the modified Heckmatt scales and mean gray-scale values revealed significant positive correlations in all of the muscles (Figure 3), suggesting high intra-test agreement. However, after excluding control group data, only the gastrocnemius and soleus showed significant correlations ($P < 0.001$). There was no correlation between (1) patient age, disease duration, patient strength by manual muscle testing, or blood CK level; and (2) subjective (modified Heckmatt score) or gray-scale in any of the muscles in any of the groups.

Discussion

In this study, we showed sonographic dissociation of EIs in the neighboring muscles in both the triceps surae and forearm muscles in patients with s-IBM. This feature was more significant in the triceps surae and was able to discriminate s-IBM from DM/PM.

Sonographic-pathological association

Muscle sonography is considered a reliable and objective marker of muscle structure and underlying pathology in neuromuscular diseases [10]. Sonography has been reported to be correlated with clinical severity in diseases including spinal muscular atrophy, juvenile dermatomyositis, and muscular dystrophy, and to be correlated with findings of other diagnostic modalities such as MRI and muscle biopsy [11-14]. In many conditions, diseased muscles show abnormal echo signal intensity and size (e.g., thickness and volume). High EI indicates muscle atrophy, inflammation, fatty infiltration, or fibrotic changes. Studies analyzing a canine model for muscular dystrophy and a single case report of amyotrophic lateral sclerosis showed that interstitial fibrous tissue most significantly affected EI; however, this observation has not been reported in myositis [15, 16].

Heterogeneous muscular involvement in s-IBM

Selective and frequently asymmetric muscular involvement is a clinical hallmark of s-IBM, most notably in finger and wrist flexors and the quadriceps [17]. This clinical information has been confirmed by imaging modalities, such as MRI and sonography [6, 7, 18, 19]. In contrast to the classic clinical description of s-IBM [17], extensive assessment of the upper and lower limbs and the pelvis by MRI has shown the highest frequency of fat-infiltrated muscles in

the lower leg [7]. Among muscles in the lower leg, the medial head of the gastrocnemius had the highest frequency of severe fatty infiltration (approximately 85% of examined patients, the highest of the 42 upper and lower limb muscles), followed by the lateral head of the gastrocnemius (approximately 65%), both much greater than in the soleus (approximately 30%). This contrast was greater than the contrast between FDP (approximately 40%) and FCU (approximately 15%) [7]. The significantly different involvements between the medial head of the gastrocnemius and the soleus by MRI [7, 18] and sonography in the present study might not be clinically obvious because examiners may have difficulty overcoming ankle plantar-flexor force, even with significant strength deficit by objective methods of assessment [20].

In contrast, MRI assessment of PM/DM showed similar frequencies of abnormal signal intensities by T2-weighted images between the medial head of the gastrocnemius and soleus [18], similar to the present data. Thus, all data consistently suggest that dissociating imaging features between these two muscles could differentiate s-IBM from PM/DM.

Our data showed similar degrees of sonographic abnormalities in the FCU and FDP in both disease groups. This is contradictory to the notion that PM/DM involves proximal muscles. Reimers, et al. studied 18 patients with dermatomyositis by sonography and reported that the vastus lateralis, supinator, and pronator teres muscles had the highest median EIs among the 16 muscles of the upper and lower limbs and trunk [21]. Although they did not evaluate other forearm muscles, their data suggest the potential involvement of forearm muscles. Noto, et al. also reported similar sonographic abnormalities between s-IBM and PM/DM patients in the FCU, although the numbers were small (N = 6 each) [6].

Study significance

S-IBM can be difficult to diagnose, and misdiagnosis is possible, especially in patients with atypical presentations [3, 22, 23]. Although identification of the characteristic pathologic features by muscle biopsy is the diagnostic gold standard, its limited sensitivity may require multiple muscle biopsies to establish a diagnosis [24]. By contrast, a non-invasive imaging study could be utilized as a supportive diagnostic measure that can be tested repeatedly with little physical harm. From this standpoint, multiple imaging modalities could be used to determine if the predominant involvement is in the gastrocnemius rather than the soleus. As stated earlier, both MRI and sonography can fulfill this role. We are not aware of any study using computed tomography to compare densities between these muscles. Sonography has several advantages over MRI. Sonography generally involves lower cost for each session, it can be a useful alternative in patient for whom MRI scanning is contraindicated, such as those with pacemakers and other devices, and it can be performed at the bedside, and thus even claustrophobic patients can be assessed. Furthermore, the handiness of sonography enables testing where accessibility to MRI is limited (e.g., rural areas and in patients with limited mobility).

Muscle sonography has several potential technical difficulties that could affect EI determination. Because recording conditions such as thick subcutaneous fat can influence the absolute EI, comparison of multiple muscles under different views may not be reliable. In contrast, the method of comparing EIs of two muscles under a single view described in the current study is preferable due to its low variability. Interestingly, the visual scale and EI showed significant correlation in the triceps surae, but not in the forearm muscles. This could be due to the smaller sizes of the forearm muscles compared to those of the calf, potentially leading to evaluation bias influenced by neighboring muscles in the forearm.

Another benefit of sonographic examination of patients suspected of having s-IBM or PM/DM is that the severity of myopathy in a particular muscle can also be assessed. Because a minimum degree of involvement is key for correct pathological diagnosis (e.g., false negative findings due to minimal sampling of involved muscle and a lack of useful information by sampling severely involved muscle tissue with fatty replacement), sonography-guided muscle biopsy is recommended.

Study limitations

This study has several limitations. First, the number of subjects was relatively small, and the overall sonographic characteristics of the muscles in patients with s-IBM are largely unknown. For example, it is not clear in our study if the sonographic contrast between the gastrocnemius and soleus is present in the early stages of IBM; there were only two patients with disease durations less than 12 months. Additional studies with larger numbers of subjects at different stages of disease could elucidate the natural course of sonographic findings in s-IBM. The significant dissociation between the FDP and FCU in s-IBM reported by Noto and colleagues was not reproducible in the present study. There are multiple explanations that could account for this difference [6]. First, their study had fewer subjects than ours (six patients in each group, compared with 11 in our study), potentially resulting in increased influence from individual subjects. Second, the mean Heckmatt rating scales in the PM/DM group were higher in the present study, suggesting that the patients with PM/DM in our study were more severely affected. Third, we used the difference of Heckmatt scale scores between the FDP and FCU, while they Noto et al. used a relative ratio. Therefore, the ratio could change even when two muscles had identical difference [e.g., for FDP and FCU scales of 2 and 1 (case 1) and 4 and 3 (case 2), the

difference between the scale scores was the same (1), but the ratio differs (2 vs. 1.33 for cases 1 and 2, respectively).

Second, it is unknown whether the dissociated appearance of the medial-gastrocnemius and soleus is unique to s-IBM. For example, amyotrophic lateral sclerosis is frequently misdiagnosed and vice versa and has also been reported to show abnormally high signals in the limb muscles [25, 26]. Unlike s-IBM, however, the distribution is more diffuse in amyotrophic lateral sclerosis, such that sonography can likely also differentiate between these disorders. A large-scale sonographic evaluation including more diseases would clarify the significance of sonography for diagnosis of s-IBM and its mimics.

Third, we only used the mean EI for quantitative analysis. It is possible that other histogram parameters and complex parameters such as texture analysis may also have diagnostic power for differentiation.

Conclusion

In conclusion, sonographic detection of the preferential involvement of the gastrocnemius over the soleus was present in patients with s-IBM; this characteristic could be more sensitive than the contrast between FDP and FCU and thus allow differentiation of s-IBM from PM/DM.

Table 1: clinical characteristics of the subjects.

(CK = creatine kinase; PM/DM = polymyositis/dermatomyositis)

	s-IBM (A)	PM/DM (B)	Control (C)	P value (ANOVA/post-hoc)
Number	11	11	11	
Number of woman	4	6	5	0.7 (ANOVA)
Age [mean $\pm$ SD; (range)]	74.5 $\pm$ 6.8 (62 - 82)	63.2 $\pm$ 12.4 (39 - 80)	73.5 $\pm$ 9.9 (57 - 88)	0.04 (A vs. B)
Duration of weakness in months [mean $\pm$ SD; (range)]	48.0 $\pm$ 40.2 (12-156)	17.5 $\pm$ 34.7 (1-120)	N/A	0.07 (A vs. B)
Serum CK [mean $\pm$ SD; (range)] [reference: 40-200 U/L]	415.4 $\pm$ 268 (150-1053)	2037.3 $\pm$ 2531 (194-8003)	N/A	0.06 (A vs. B)

Table 2: Subjective evaluation of echoic intensities by the modified Heckmatt rating scale. Averaged intensities of three raters (range of mean scores of each rater)

Group	s-IBM (N = 11):(A)	PM/DM (N = 11):(B)	Controls (N = 11):(C)	<i>P</i> values (one-way ANOVA with Games-Howell post-hoc): *(< 0.05), # (< 0.01), § (< 0.001)
Gastrocnemius (GC)	3.12 (3.0:3.3)	1.91 (1.6:2.4)	1.12 (1.0:1.4)	§ (A:B, A:C), # (B:C)
Soleus	1.97 (1.9:2.0)	1.91 (1.7:2.0)	1.09 (1.0:1.2)	# (A:C, B:C)
Difference (GC: soleus)	1.15 (1.0:1.3)	0.00 (-0.5:0.4)	0.03 (-0.2:0.3)	# (A:B, A:C)
Flexor digitorum profundus (FDP)	2.48 (2.2:2.7)	2.30 (2.1:2.5)	1.00 (1.0:1.0)	§ (A:C), # (B:C)
Flexor carpi ulnaris (FCU)	1.63 (1.2:1.9)	1.69 (1.4:2.1)	1.00 (1.0:1.0)	§ (A:C), # (B:C)
Difference (FDP: FCU)	0.85 (0.7:1.0)	0.61 (0.3:0.8)	0.00 (0.0:0.0)	§ (A:C), * (B:C)

Table 3: Echo-intensity analysis by histogram showing the averaged mean echo-intensity

	s-IBM: (A)	PM/DM: (B)	Controls: (C)	<i>P</i> values (one-way ANOVA with Games-Howell post-hoc): *(< 0.05), # (< 0.01), § (< 0.001)
Gastrocnemius (GC)	103.2 ± 30.4	78.2 ± 18.1	51.0 ± 2.9	§(A:C), # (B:C), 0.08 (A:B)
Soleus	47.8 ± 24.5	60.0 ± 19.3	39.2 ± 3.4	*(B:C)
Difference (GC: soleus)	55.4 ± 38.0	18.2 ± 15.4	11.7 ± 14.3	#(A:C), *(A:B)
Flexor digitorum profundus (FDP)	86.1 ± 26.9	58.2 ± 21.1	38.7 ± 10.6	§(A:C), *(A:B, B:C)
Flexor carpi ulnaris (FCU)	66.5 ± 24.0	55.9 ± 28.9	38.7 ± 10.8	#(A:C)
Difference (FDP: FCU)	19.6 ± 22.0	2.2 ± 19.9	0.0 ± 8.5	*(A:C)

Supplemental Table 1: Intraclass correlation coefficients [ICC(2,1)] and Cronbach's alpha of subjective evaluating scales by three raters on all the subjects

	ICC(2,1) (95% confidence interval)	Cronbach's alpha
Gastrocnemius	0.76 (0.61 – 0.86)	0.91
Soleus	0.60 (0.41 – 0.76)	0.81
FDP	0.76 (0.62 – 0.86)	0.91
FCU	0.44 (0.21 – 0.64)	0.75

Supplemental Table 2: Comparison of the modified Heckmatt rating scale (the average of the three raters). The averaged difference of the score < 1 between the adjacent muscles was considered as similar.

	GC > soleus (1 or more)	GC $\div$ soleus	GC < soleus (1 or more)	frequency (GC > soleus)
IBM	9	1	1	81.8%
PM/DM	0	11	0	0%
Control	0	11	0	0%

$P = 0.0002$ (IBM vs. PM/DM (Fisher's exact test))

	FDP > FCU (1 or more)	FDP $\div$ FCU	FDP < FCU (1 or more)	Frequency (FDP > FCU)
IBM	6	5	0	54.5%
PM/DM	3	8	0	27.2%
Control	0	11	0	0%

$P = 0.4$ (IBM vs. PM/DM (Fisher's exact test))

Supplemental Table 3: Comparison of the area under the curve (AUC) by the receiver operating curve (ROC) analysis between s-IBM and polymyositis/dermatomyositis using the averaged mean echogenicity by histogram analysis

	AUC	<i>P</i> value (vs. true area = 0.5)
Gastrocnemius	0.727	0.071
Soleus	0.661	0.200
Difference: Gastrocnemius-Soleus	0.843	0.006
FDP	0.802	0.017
FCU	0.645	0.250
Difference: FDP-FCU	0.686	0.140

Figure legends

Figure 1: Representative sonographic axial images of the triceps surae. In the control subject (Panel A), the echo-intensities of the gastrocnemius and soleus muscle fibers are similarly low-echoic with high-echoic perimysium. Gastrocnemius and soleus are clearly separated by high-echoic epimysium (Heckmatt rating scale (HS) = 1). In the patient with inclusion body myositis (s-IBM) (Panel B), there was significant contrast of hyperechoic gastrocnemius (HS = 3) and relatively normoechoic soleus (HS = 1). In the patient with polymyositis (Panel C), both muscles were hyperechoic and the separating epineurium was not clearly identifiable (HS = 4 each).

Figure 2: Averaged gray-scale histograms of the sonographic images of four muscles in the three groups (N = 11 each). Pixel values range from 0 (black) to 255 (white) (inset, Panel A). Normal muscles have high peaks with narrow durations, reflecting homogenous echo-intensities. By contrast, the images of the myopathy patients tended to have higher (i.e., whiter) pixel values and broader durations than the control images, suggesting heterogeneous echo-intensities.

Figure 3: Correlations are shown between the modified Heckmatt scores (the averages of three raters) and the mean gray-scale values in all the subjects (N = 33). Significant positive correlations were present in all the muscles suggesting reliability of two evaluating methods. There were tendencies of stronger correlations in the triceps surae than in the forearm muscles.

Supplemental Figure 1: Representative sonographic axial images of the proximal forearm. In the control subject, the echo-signals were similar between the flexor digitorum profundus (FDP) and

flexor carpi ulnaris (FCU) (Heckmatt rating scale (HS) = 1 each). In the patient with s-IBM, the FDP was hyperechoic (HS = 2), whereas FCU was spared (HS = 1). Such contrast was lacking in the patient with PM/DM (Panel C), showing diffuse hyperechogenicity (HS = 3 each). FDS = flexor digitorum superficialis

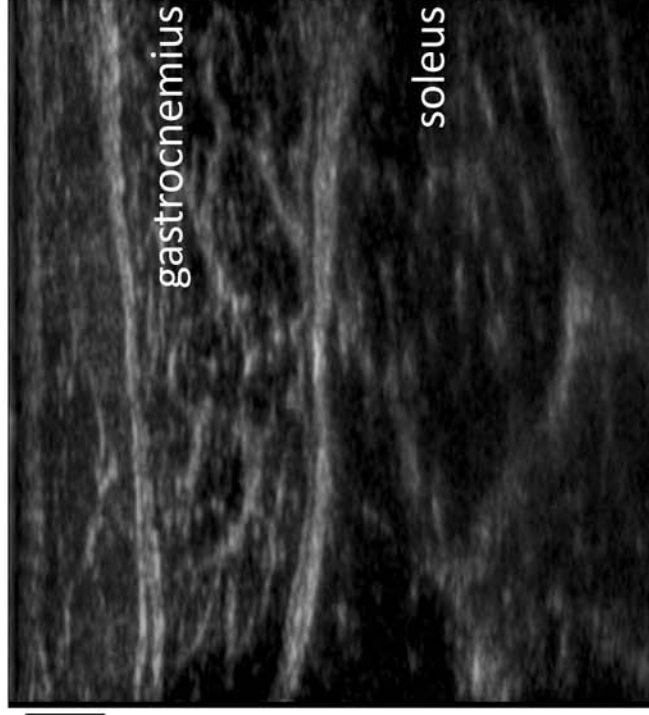
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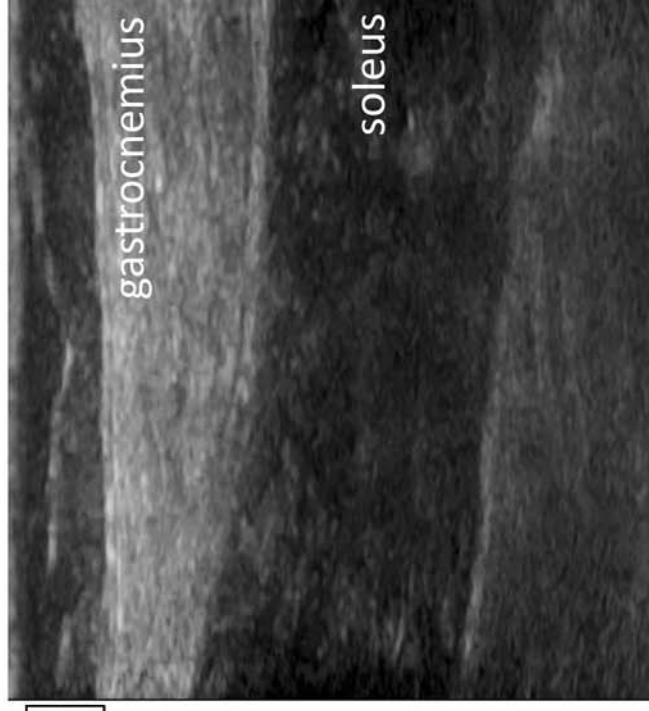
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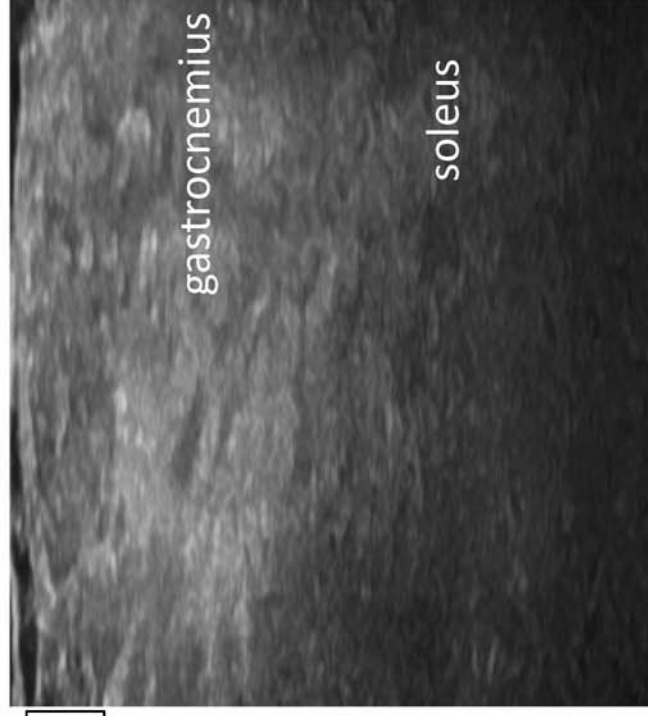


Figure 1

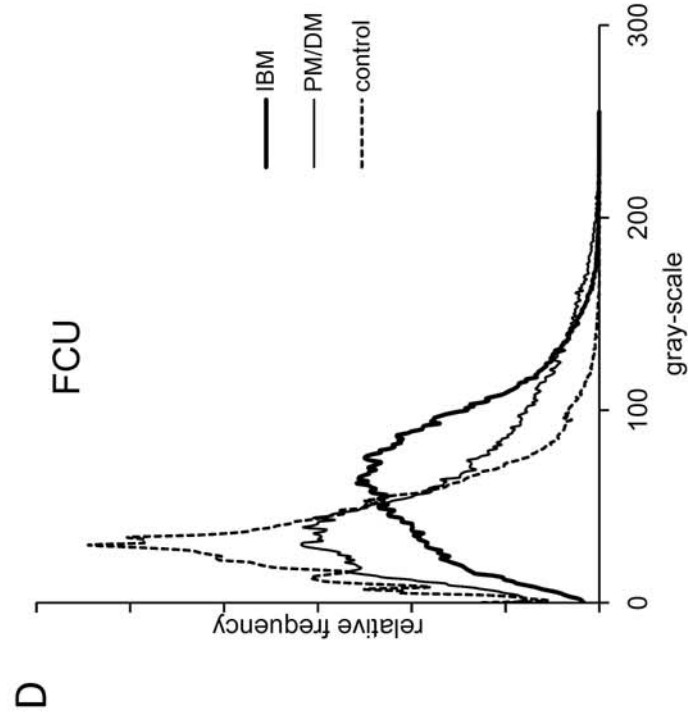
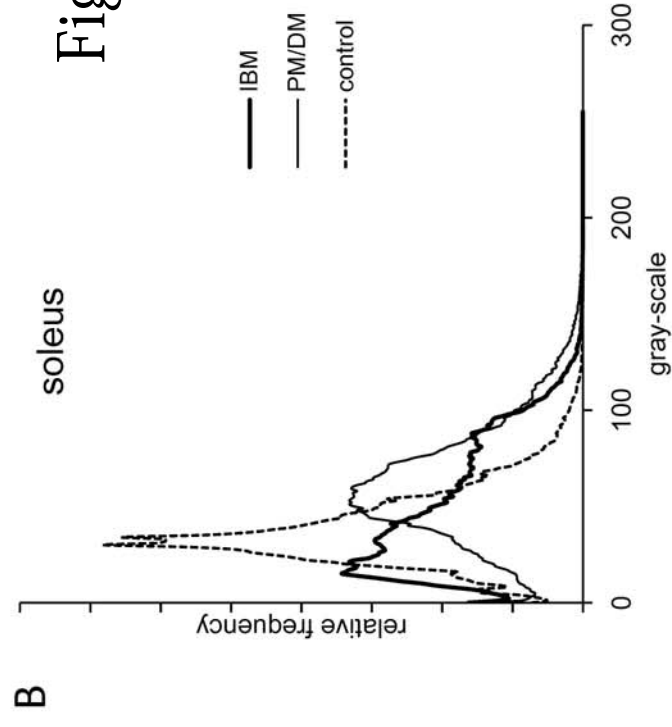
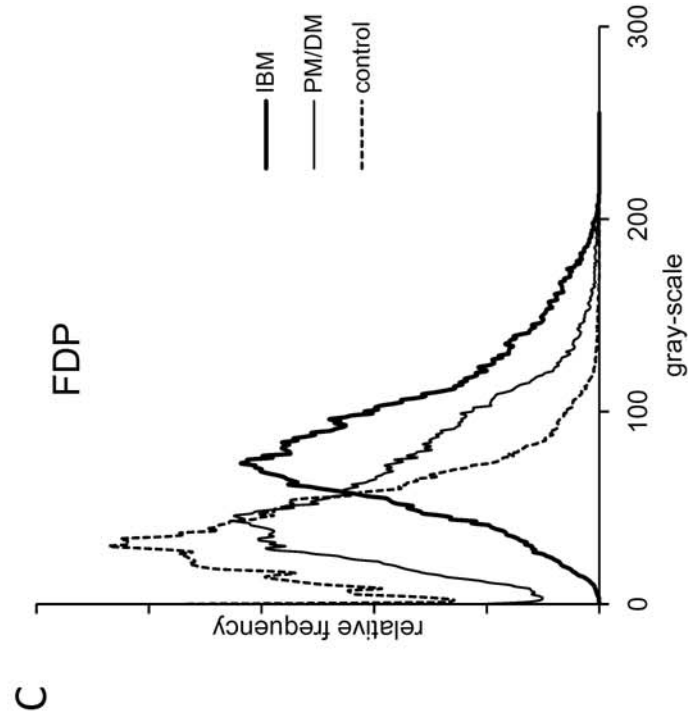
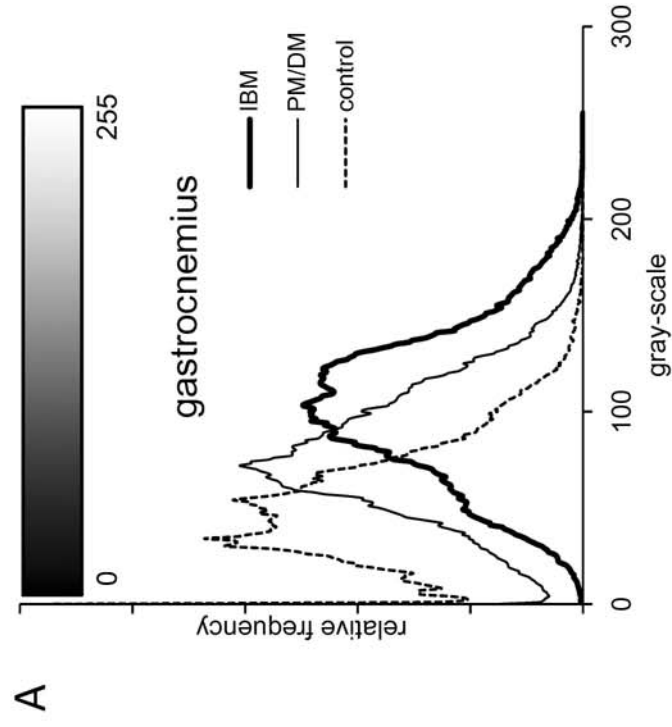


Figure 2

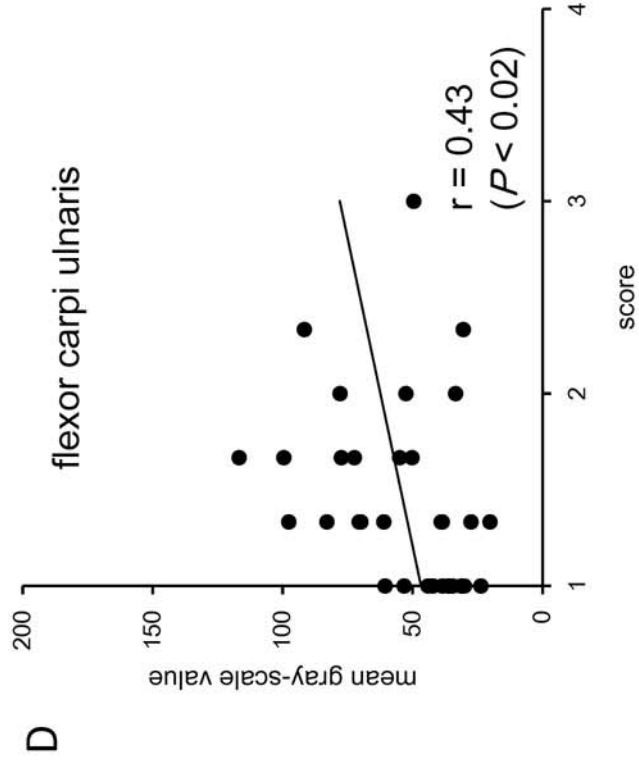
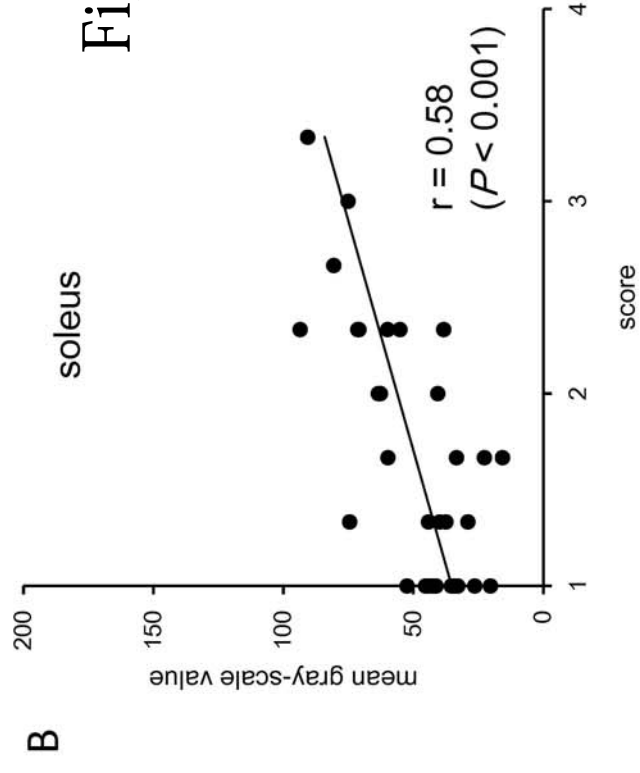
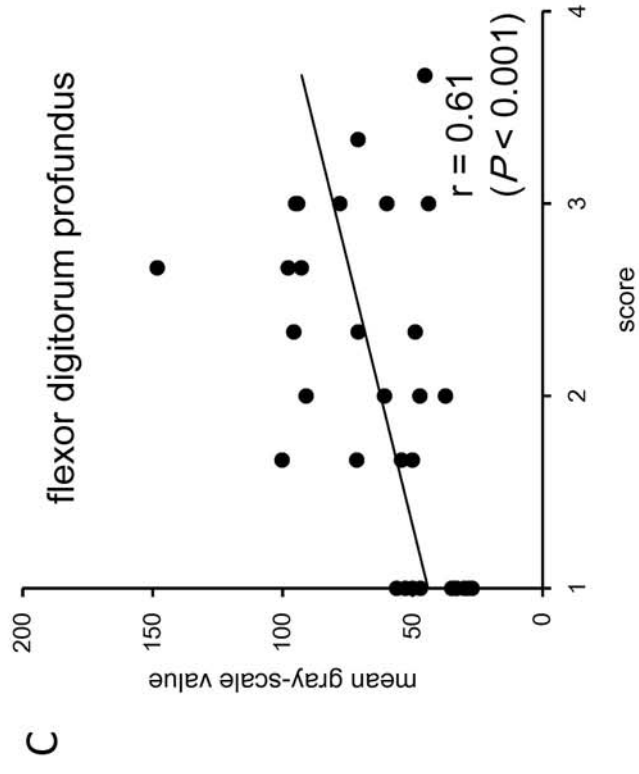
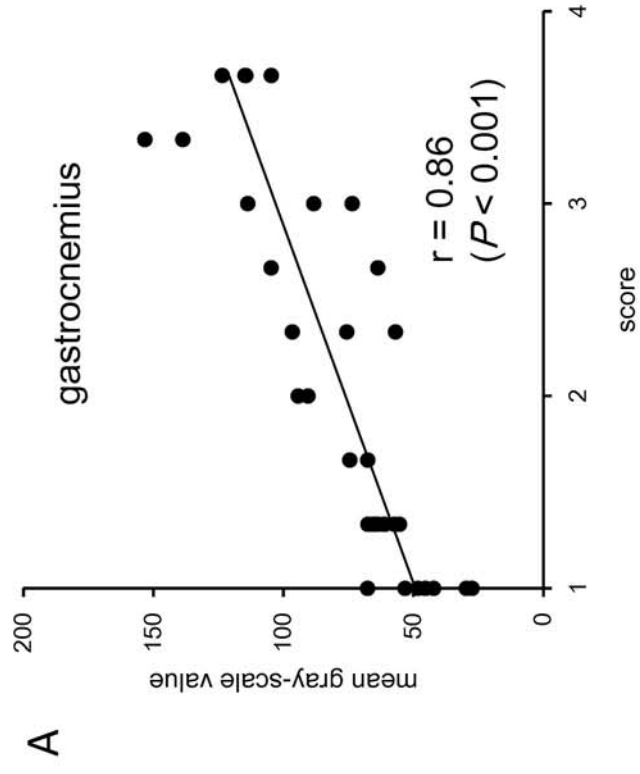
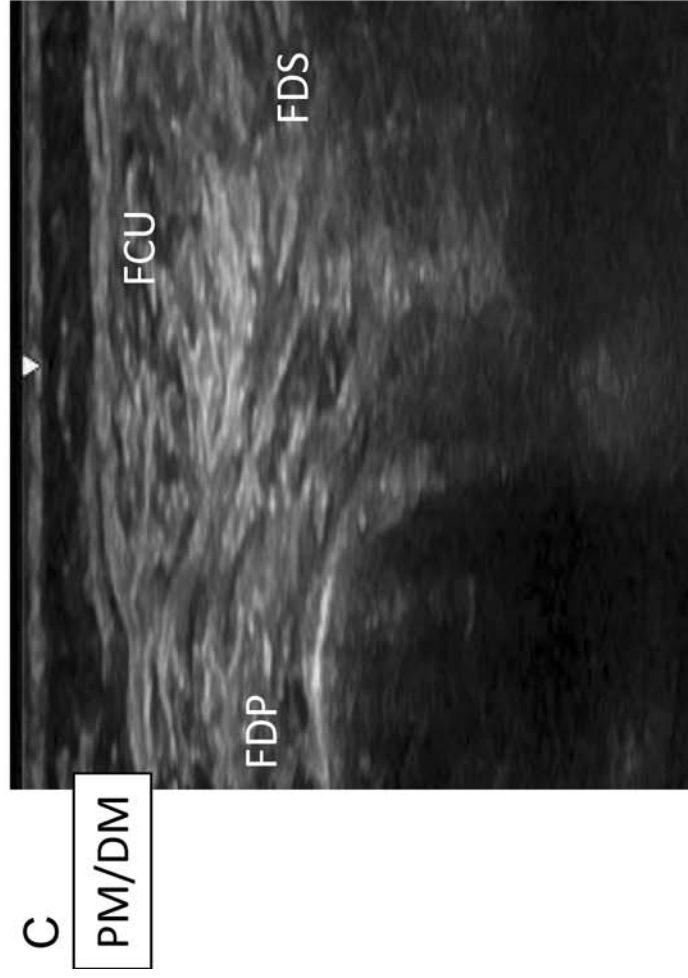
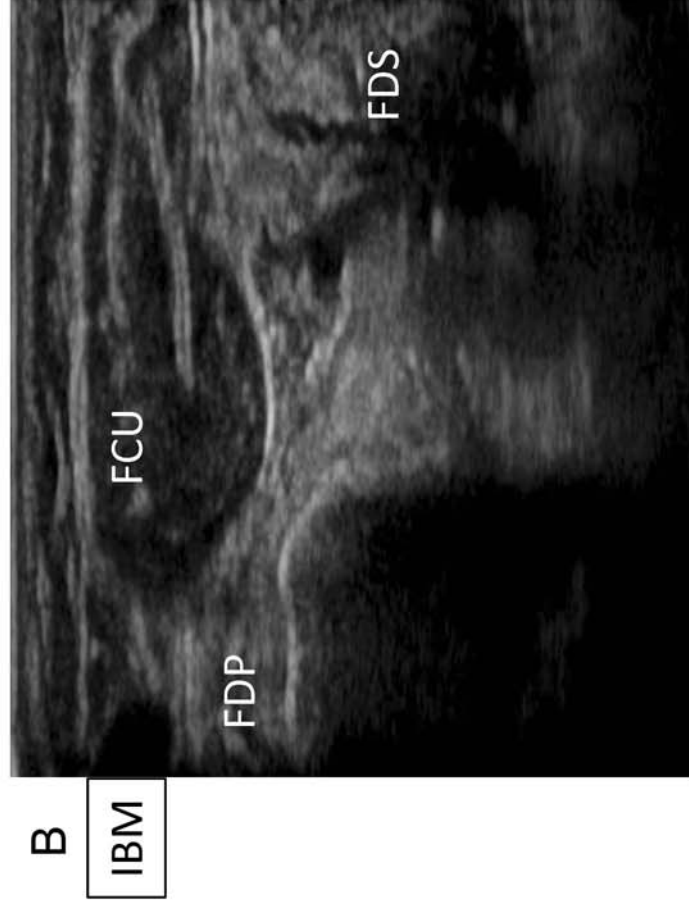
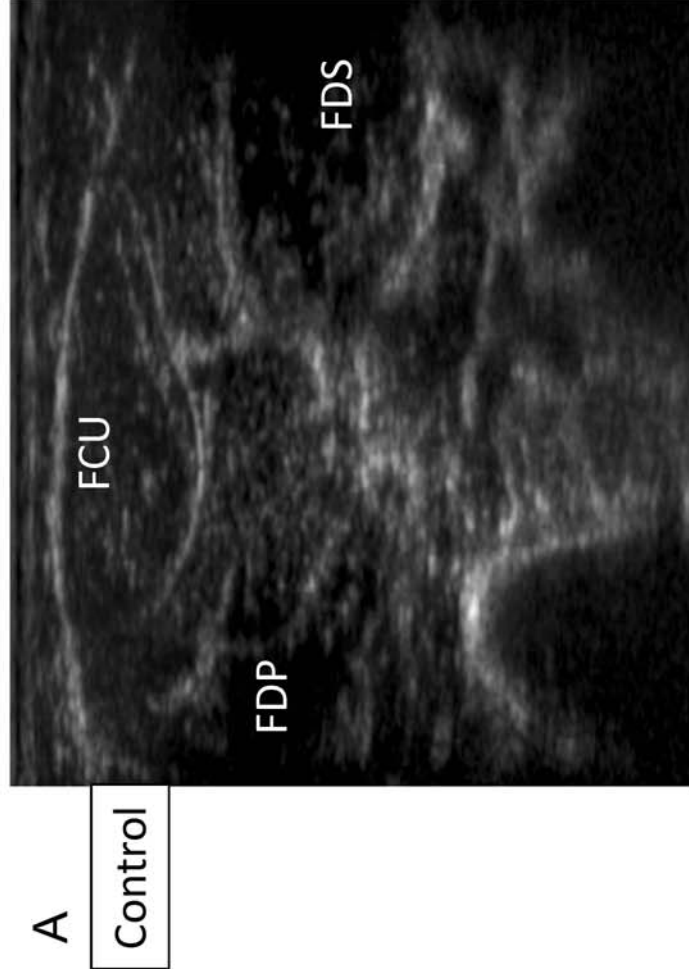


Figure 3



Supplemental figure 1