**Abstract**

**Objective.** To analyse whether MRI of upper and lower extremity muscles in a large patient group with sporadic IBM (sIBM) is of additional value in the diagnostic work-up of sIBM.

**Methods.** Thirty-two sIBM patients were included. Magnetic resonance (MR) parameters evaluated in 68 muscles of upper and lower extremity were muscle atrophy, fatty infiltration and inflammation. These findings were correlated with disease duration, weakness and serum creatine kinase (sCK) levels.

**Results.** Fatty infiltration was far more common than inflammation. Muscles most frequently infiltrated with fat were the flexor digitorum profundus (FDP), anterior muscles of the upper leg and all muscles of the lower leg, preferentially the medial part of the gastrocnemius. The rectus femoris was relatively spared compared with other quadriceps muscles as well as the adductors of the upper leg. Inflammation was common in general, but individually sparse, present in 78% of the patients with a median of two inflamed muscles per patient. A statistically significant correlation was found between the amount of fatty infiltration and disease severity, disease duration and sCK.

**Conclusion.** We provide a detailed description of the MRI in sIBM and show a distinct pattern of muscle involvement. Relatively severe affliction of the medial compartment of the gastrocnemius, combined with relative sparing of the rectus femoris or involvement of the FDP can be indicative of sIBM. MRI can contribute to the diagnosis in selected patients with clear clinical suspicion, but lacking the mandatory set of muscle biopsy features.

**Key words:** Inclusion body myositis, Magnetic resonance imaging, Muscles, Muscular diseases.

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**Introduction**

Sporadic IBM (sIBM) belongs to the idiopathic inflammatory myopathies and is characterized by painless multifocal muscle weakness and wasting, manifesting after the age of 40 years. Diagnosis is based upon the clinical features and characteristic muscle biopsy findings. Symptoms usually follow the onset of weakness in the quadriceps, finger flexor or pharyngeal muscles. The spread of muscle weakness is erratic and asymmetric, but in general ventral muscle groups are more affected than dorsal muscle groups in the extremities, and girdle muscle groups are least affected [1]. The characteristic features in the muscle biopsy are mononuclear inflammation with T-cell invasion of non-necrotic muscle fibres and rimmed vacuoles. In a subgroup of patients, histology does not show these characteristic biopsy features initially, leading to a delayed or wrong diagnosis [2, 3].

MRI has become a useful modality in the evaluation of inflammatory myopathies [4]. MRI provides detailed anatomical information and oedema-like signal intensity changes give information on the activity of the disease, an advantage compared with other imaging modalities. Five previous studies describe MRI in patients with sIBM. However, in contrast to our study, those studies comprise much smaller patient groups and were restricted with regard to the number of muscles studied [5-9]. It has been stated that MRI could distinguish PM from sIBM by the presence of isolated muscle inflammation in the absence of fatty infiltration or atrophy, as opposed to the findings in sIBM [6]. Another study focused on the forearm.
muscles, and showed explicit fatty changes in the flexor digitorum profundus (FDP) muscle, sometimes preceding clinically detectable weakness in this muscle [9].

To give an elaborate view of the severity and distribution of abnormalities in MRI, we investigated a large and well-defined group of sIBM patients. We investigated whether MRI could be of additional value to the clinical features during the diagnostic process.

Patients and methods

Patients
The study comprised 32 patients diagnosed with sIBM. All patients fulfilled the European Neuromuscular Centre (ENMC) criteria [10] for definite (n = 31) or probable (n = 1) sIBM. Patients were randomly selected from the Leiden University Medical Center (LUMC) registry of sIBM patients. The study was approved by the ethics committee of the LUMC and all patients gave informed consent.

History taking included functional grading scales [Barthel index, scale 0–20; Rivermead mobility index, scale 0–15; and Brooke’s grading system, scale 3–23 (with 23 being the worst score)]. Physical examination comprised manual muscle strength testing (MMT) of 34 muscle groups using the 6-point British Medical Research Council (MRC) scale. Serum creatine kinase (sCK) activity was measured (normal value ≤ 170 U/l in women and ≤ 200 U/l in men).

MRI

MRI examinations were performed at 1.5 T (Gyroscan NT15; Philips Medical System, Best, The Netherlands) by the following standard protocol. Each series was obtained with a body coil with the patient supine. The following sequences were used: (i) axial T1-weighted spin-echo series with 600/20 (repetition time ms/echo time ms), section thickness of 6.0 mm, field of view 250 mm, rectangular field of view of 59%, number of signal averages (NSA) = 2 and an acquisition matrix of 205 x 256; (ii) axial short TI inversion recovery (STIR) series with 1400/15 (repetition time ms/echo time ms), section thickness of 6.0 mm, field of view 250 mm, rectangular field of view of 59%, NSA = 4 and an acquisition matrix of 202 x 256. Imaging of the right shoulder, upper and lower right arm, the pelvis, and both upper and lower legs was performed. Two musculoskeletal radiologists, with, respectively, 4 and 14 years of clinical experience, without knowledge of the clinical findings, but aware of the diagnosis of sIBM, evaluated all magnetic resonance (MR) examinations in consensus. Muscles were scored for muscle atrophy, fatty infiltration and oedema-like changes. Muscle atrophy was defined as evident loss of muscle volume, scored as present or absent. T1-weighted MR images were used to estimate the degree of fatty infiltration. Abnormal signal intensity was consequently classified as mildly abnormal if only fatty streaks of increased signal intensity could be observed (< 30% of fat compared with muscle), moderately abnormal if 30–60% of the muscle showed increased signal intensity and severely abnormal if at least 60% of the muscle showed increased signal intensity [11].

On STIR images, any level of high signal intensity of the muscles was considered an abnormal finding: oedema-like changes. In this patient group, oedema-like changes are considered to reflect inflammation as previously used [4, 6, 8, 12, 13]. This was scored as present or absent. Asymmetry could be scored in the lower extremities only, as both sides were visualized. The examined muscles were: (i) shoulder region: deltoïd, infraspinatus, supraspinatus, subscapularis; (ii) upper arm: biceps, triceps; (iii) forearm: flexor carpi ulnaris, flexor carpi radialis, FDP and flexor digitorum superficialis (FDS), brachioradialis, extensor carpi ulnaris and radialis, extensor digitorum communis, supinator, pronator teres; (iv) pelvis: gluteus minimus, gluteus medius, gluteus maximus, iliopectineus, obturatorius internus and externus, pectineus; (v) upper leg: quadriceps femoris (rectus femoris, vastus lateralis, medialis and intermedius), semi-membranosus, semi-tendinosus, biceps femoris, sartorius, gracilis, adductor brevis, longus and magnus; and (vi) lower leg: gastrocnemius (lateral and medial part), soleus, tibialis posterior, tibialis anterior, peroneus longus and extensor digitorum longus. In total, 68 separate muscles were evaluated.

Statistics

Descriptive measures are presented as mean (±SD), unless otherwise stated. Correlation between MRI and different clinical parameters was calculated using the Spearman rank test. The Mann–Whitney U-test was performed to compare total strength of patients with and without a fatty infiltrated FDP. P < 0.05 was considered to be statistically significant.

Results

History, clinical examination and sCK

Mean age of the 32 included patients (19 men) was 68 (9) years, with a mean disease duration of 12 (5) years. Mean time to diagnosis was 8 (5) years. Median Barthel index score (range) was 19 (6–20), Rivermead mobility index score 12 (0–15) and for Brooke’s grading system 6 (3–16).

With MMT, the most frequently affected muscles were the ventrally located muscles in the arm, the upper and the lower leg. The most frequent and severely (MRC scale 0–3) affected muscles were the FDP (n = 15, 47%) and quadriceps (n = 11, 34%). Relatively spared muscles were the infraspinatus and adductors of the upper leg, which had normal strength in 19 (59%) and 21 (66%) patients, respectively. sCK was elevated in 28 patients (88%). Median sCK was 739 (121–3360) for men and 265 (44–802) for women.

MRI

General findings

In two patients, the quality of MRI of the forearm was not sufficient to be evaluated due to motion artefacts. In all patients, MR images showed abnormalities. All patients
had fatty infiltrated muscles. The median number of muscles infiltrated by fat per patient was 40 (range 5-68, interquartile range 29). With regard to the extent of involvement, the leg muscles were more frequently and severely affected as compared with the arm muscles. The lower legs were more frequently and severely affected than the upper legs. The shoulder and pelvic girdle muscles and adductors of the upper leg were relatively spared (Table 1).

Inflamed muscles were present in 78% of the patients. The median number of inflamed muscles was 2 (range 0-20, interquartile range 4). The most frequently inflamed muscles were the deltoid muscle (16%), the extensor carpi ulnaris (20%), the medial (22%) and lateral part of the gastrocnemius (16%) and the soleus muscle (16%). Isolated muscle inflammation (i.e. without fatty infiltration) was observed in 13 of all observed muscles. This phenomenon was mostly seen in muscles that were relatively spared fatty infiltration, such as the thigh adductors (n=5) and the extensor carpi ulnaris (n=2; Fig. 1C and D).

Atrophy was present in 94% of the patients. The median number of atrophic muscles was 22 (range 0-68, interquartile range 28). The most frequently atrophic muscles were the FDP (60%), the vastus muscles (69%) and the medial head of the gastrocnemius (81%). No hypertrophy was found.

Asymmetry of abnormalities (fatty infiltration as well as inflammation) was present in 14 patients (44%) in the lower extremities. Asymmetry of fatty infiltration was mostly seen in the adductor magnus and the tibialis anteriors.

**Table 1** Mean frequency of fatty infiltrated muscles and of those with severe fatty infiltration in different extremities

<table>
<thead>
<tr>
<th>Body region</th>
<th>Fatty infiltration, %</th>
<th>Severe fatty infiltration, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder</td>
<td>36</td>
<td>12</td>
</tr>
<tr>
<td>Arm</td>
<td>44</td>
<td>15</td>
</tr>
<tr>
<td>Upper</td>
<td>42</td>
<td>15</td>
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<tr>
<td>Lower</td>
<td>44</td>
<td>15</td>
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<tr>
<td>Pelvis</td>
<td>33</td>
<td>15</td>
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<tr>
<td>Leg</td>
<td>81</td>
<td>42</td>
</tr>
<tr>
<td>Upper</td>
<td>76</td>
<td>38</td>
</tr>
<tr>
<td>Anterior part</td>
<td>84</td>
<td>50</td>
</tr>
<tr>
<td>Posterior part</td>
<td>58</td>
<td>26</td>
</tr>
<tr>
<td>Lower</td>
<td>87</td>
<td>44</td>
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**Fig. 1** Images of inflammation (STIR) of the forearm and legs. Lower legs (A), showing inflammation in the medial part of the gastrocnemius muscle. Right forearm (B) showing inflammation in the extensor carpi ulnaris (ECU) muscle. Right forearm (C; T1-weighted) showing extensive fatty infiltration of all muscles, with sparing of the ECU. An STIR image (D) of the same patient shows exclusive inflammation of the ECU.
anterior muscle (13% in both), whereas asymmetry of inflammation was mostly seen in the tibialis posterior and soleus muscle (9% in both).

**MRI findings per body region**

**Shoulder.** Involvement through fatty infiltration of shoulder muscles as a group was less frequent and severe than in other upper extremity muscle groups (Table 1). Fatty infiltration in the shoulder region was most frequently observed in the subscapular muscle ($n=18$, 56%). Inflammation was most frequent in the deltoid muscle; five (16%) patients had inflammatory changes in the deltoid muscle compared with one (3%) patient in each of the other shoulder muscles (Figs 2–4).

**Upper arm.** Fatty infiltration was equally present in the biceps and triceps. No inflammation was seen in the biceps. Only two patients had inflammatory changes of the triceps (Figs 2–5).

**Forearm.** The most frequently and severely affected muscle of the upper extremity by fatty infiltration was the FDP. Twenty-two (73%) patients had an FDP infiltrated with fat, in 13 patients (43%) affliction was considered severe. Atrophy was also most frequently observed in the FDP ($n=18$, 60%). In eight patients (27%), the FDP was unaffected.

As a rule, if the FDP did not show fatty infiltration, then it was absent in all forearm muscles. The only exception was a patient with some fatty streaks in the supinator muscle. Preference for fatty infiltration was also seen in the FDS and the supinator muscle, but they were clearly less severely affected than the FDP. Patients with an unaffected FDP on MRI had a mean disease duration of 13 (5) years, demonstrating that the FDP can remain unaffected for a long time. Inflammatory changes were most frequently seen in the extensor carpi ulnaris ($n=6$, 20%) (Figs 1B and D, 2–5, 6A and B).

**Pelvis.** The muscles around the pelvis were less frequently and severely affected by fatty infiltration than other muscle groups of the lower extremity (Table 1). The gluteus maximus was the most frequently affected muscle by fatty infiltration ($n=19$, 59%) of the gluteal musculature, although not severely (9%) and without asymmetry. None of the gluteal muscles had inflammatory changes (Figs 2–4).

**Upper leg.** The anterior muscle group of the upper leg was frequently affected by fatty infiltration. The rectus femoris was relatively spared compared with other quadriceps muscles, but normal in only seven patients (22%). Complete sparing of the quadriceps muscles was seen in only two cases. In these patients, all other anterior

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**Fig. 2** Severity and frequency of fatty infiltration of muscle groups for the total group of patients.
and posterior thigh muscles were spared as well, but both patients had abnormalities in the lower legs. None of the patients had exclusive involvement of quadriceps.

The posterior muscle group of the upper leg was far less frequently and less severely affected by fatty infiltration as compared with the anterior compartment (Table 1). The hamstring muscles more often had a preference for fatty infiltration as compared with the relatively spared adductor muscles. Inflammation was rare, and the most frequently inflamed muscle was the adductor brevis \((n = 4, 13\%)\).

Atrophy was common in all muscles of the anterior compartment of the upper leg with exception of the gracilis muscle. This muscle was regarded as atrophic in only 28\% of the patient group (Figs 2–5, 6C and D).

**Lower leg.** All muscles of the lower leg were commonly affected by fatty infiltration, the medial part of the gastrocnemius muscle appearing as the most severely affected one. Inflammatory changes in the lower extremity were also more common in the lower leg, again with a preference for the medial part of the gastrocnemius, showing inflammation in seven patients (22\%). Atrophy was common in all lower leg muscle groups with the exception of the soleus and the tibialis posterior muscles; these muscles were atrophic in, respectively, 11 (34\%) and 10 (31\%) patients only (Figs 1A, 2–5, 6E and F).

**Correlation between MRI and other findings**

There was a statistically significant correlation between disease duration and number of muscles infiltrated by fat \((r = 0.6, P = 0.001)\). However, the pattern of muscles invaded with fat in patients with short and long disease duration was similar. No correlation was found between disease duration and the number of inflamed muscles \((r = 0.4, P = 0.8)\), implying that short disease duration was not a prerequisite for inflammation. In fact, the three patients with the highest frequency of inflamed muscles (>11) had a disease duration of 10, 11 and 16 years. No correlation was found between age and MRI abnormalities. The seriousness of functional incapacity, as measured by the grading scales, was associated with the number of muscles infiltrated with fat \((r = 0.6, P = 0.001)\). Functional grading scales did not correlate with the number of inflamed muscles.

Weakness was associated with more fatty infiltration as the sum score of all muscles tested with MMT had a negative correlation with the number of muscles infiltrated by fat \((r = −0.8, P < 0.005)\). The number of inflamed muscles showed no correlation with the MMT sum score \((r = −0.2, P = 0.4)\). Patients with an FDP infiltrated with fat scored significantly worse on MMT sum score (median = 245, \(n = 22\)) than patients with a normal FDP on imaging (median = 286, \(n = 8\), \(P = 0.002\)).
FDP muscles of normal strength with MMT, present in five patients, were normal on MRI, except for one case with some fatty streaks. sCK levels showed a moderate, although negative correlation with the number of fatty infiltrated muscles on MRI ($r = -0.417, P = 0.017$), but did not correlate with the number of inflamed muscles ($r = 0.252, P = 0.16$).

**Discussion**

MRI is an easy and excellent technique to visualize muscle pathology in sIBM, as demonstrated by the results of this large cohort of patients. All sIBM patients showed abnormalities on MRI of their muscles. Fatty infiltration was the most common abnormality in all patients. It was observed, in decreasing order of frequency, in the lower legs then the upper legs, followed by the forearm, pelvis, upper arm and the shoulder girdle. However, within these body parts large differences were seen. In the upper legs, the adductors were most frequently spared, the hamstrings intermediately affected and the quadriceps most affected. In the forearm, there proved to be preferential affliction of the FDP muscle, sometimes as single muscle involvement.

Overall, in MRI, ventrally located muscles were more frequently and severely affected than dorsally located muscles, and abductors and adductors of the pelvic and shoulder girdles were the most spared, confirming the clinical findings [1]. MRI also provides information on muscles that cannot be tested clinically. For example, in the lower leg the plantar flexors of the foot, the gastrocnemius and soleus muscles, are clinically relatively spared; however, MRI points out that especially the medial head of the gastrocnemius is abnormal, with relative sparing of the soleus muscle. In the upper leg, the rectus femoris is relatively spared compared with the other quadriceps muscles.

Involvement of the FDP appeared not to be obligatory for the disease. Although Sekul et al. [9] found a very high incidence of fatty infiltration in the FDP in 20/21 sIBM patients with an average disease duration of 6.5 years, we found unaffected FDP muscles in a quarter of sIBM patients. In one patient, MR abnormalities in the FDP preceded clinically detectable weakness, which has been described earlier [9].

Oedema-like changes are not specific for inflammatory myopathies, but can also be seen in other neuromuscular disorders (NMDs), e.g. in muscular dystrophy [11]. As in muscular dystrophy, sIBM patients also showed that muscles affected by oedema-like changes showed less fatty infiltration. As oedema-like changes were not associated with disease duration, in contrast to fatty infiltration,
these findings hint towards inflammation preceding fatty infiltration.

The pattern of fatty infiltration is the most informative when one wants to discriminate between different NMDs. The sIBM pattern described here is distinct from other myopathies that are to be considered in an elderly patient with insidious onset of muscle weakness, including dystrophinopathies, PM and DM.

In dystrophinopathies (Becker muscular dystrophy and limb-girdle muscle dystrophies), the quadriceps muscles are also commonly invaded with fat. The biceps femoris, the semi-membranosus and the adductors of the upper leg are also preferably affected in these disorders, in contrast to sIBM [14]. In PM, fatty infiltration is far less pronounced than in sIBM. Another difference is the pronounced localization of fat in the anterior muscle groups of the upper and the lower leg in sIBM contrary to PM [6].

In DM, inflammation is more pronounced than fatty infiltration and oedema is found along the fascia and subcutaneous fat [15]. PM and DM have more symmetrical abnormalities in contrast to sIBM. The marked
abnormalities of the FDP as observed in sIBM have never been described in another myopathy.

In clinical practice, selective MRI is preferred above imaging of six different body regions, mainly because imaging is time consuming. To differentiate between the myopathies mentioned above we suspect imaging of the upper legs and forearm to be the most informative. The sIBM pattern of the upper leg (relative sparing of the rectus femoris muscle and hamstrings) and the extent of fatty infiltration (87%), as well as the asymmetry, mostly seen in the adductor magnus, can effectively differentiate sIBM from other myopathies.

Although there is a high frequency of FDP abnormalities in sIBM on imaging, subclinical involvement appeared rare. Imaging of the forearm can be used to confirm weakness of the FDP, especially when the presence of weakness is subtle or uncertain. The absence of a control group limits interpretation of the study. Discrimination from other myopathies was therefore not based on direct comparison, rather on knowledge.

![Fig. 6 T1-weighted images with different levels of fatty infiltration in the forearm and legs. Right forearm showing moderate (A) and severe (B) fatty infiltration of the FDP. Upper legs (C and D) showing severe fatty infiltration of the vastus muscles, with relative sparing of the rectus femoris muscle and hamstrings. Lower legs showing moderate (E) and severe (F) fatty infiltration of the gastrocnemius muscles, especially the medial part. (F) Relative sparing of the extensor digitorum longus, tibialis posterior and soleus muscles.](image-url)
from published articles describing MR findings in other myopathies. The awareness of the diagnosis may also raise concern about bias by the radiologists. Although the patients in this study had a relatively long mean disease duration, subgroups of patients with shorter and longer disease duration had a similar pattern of fatty infiltration.

In summary, although muscle involvement on MRI in individual sIBM patients is diverse, it shows a pattern in sIBM different from other myopathies. The diagnosis of sIBM is often made after a mean disease duration of 7 years due to a combination of patient and doctor’s delay [16]. Not uncommonly the diagnosis is difficult to establish because of a not yet typical clinical picture or repeated muscle biopsies not showing the mandatory features. MRI of the skeletal muscles can, especially in these cases, contribute to a diagnosis of sIBM.

**Rheumatology key messages**

- Fatty infiltration is the most common MR abnormality in sIBM and correlates with weakness.
- The pattern of distribution of fatty infiltration in sIBM muscle is thought to be specific for the disease.
- MRI can contribute to diagnosing sIBM.

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**References**