

## **MRI Quantification of Abnormal Muscle Water Distribution in Chronic Neuromuscular Diseases: A Sensitive Biomarker**

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### **Abstract:**

**Background:** Quantitative muscle MRI in neuromuscular diseases can measure abnormal muscle water distribution through measurement of T2 relaxation times and the magnetisation transfer (MT) ratio, which may precede fatty infiltration and potentially provide a sensitive biomarker.

**Methods:** 20 patients with Charcot-Marie-Tooth disease type 1A (CMT1A), 20 patients with inclusion body myositis (IBM) and 27 matched controls underwent 3T MRI including short-tau inversion recovery (STIR), 3-point Dixon fat quantification, pseudo-T2-mapping and MT sequences. MRI analysis was performed blinded to diagnosis.

**Results:** There was qualitative STIR hyperintensity in more than half of lower limb muscles in IBM patients and in 31% of calf muscles in CMT1A patients. T2-times were significantly increased and MT ratio significantly reduced in thigh and calf muscles of IBM patients and calf muscles of CMT patients compared with controls. These differences remained significant ( $p < 0.001$ ) when only muscles with normal fat levels (<5%) on Dixon imaging were included, displaying T2 times of (mean  $\pm$  s.d. milliseconds): thigh – IBM  $50.2 \pm 8.1$ , CMT1A  $43.4 \pm 5.3$ , control  $43.1 \pm 5.0$ ; calf – IBM  $46.6 \pm 7.3$ , CMT1A  $42.6 \pm 5.0$ , control  $41.0 \pm 3.6$ ; and MT ratios of (mean  $\pm$  s.d. percentage units): thigh – IBM  $29.4 \pm 2.6$ , CMT1A  $31.7 \pm 2.0$ , control  $31.9 \pm 1.6$ ; calf – IBM  $30.4 \pm 2.4$ , CMT1A  $31.4 \pm 1.7$ , control  $32.1 \pm 1.3$ .

**Conclusions:** We demonstrated abnormal muscle water distribution in calf muscles in CMT1A and calf and thigh muscles in IBM using qualitative STIR and pseudo-quantitative T2 and MT sequences. The differences were greater in IBM patients than in CMT1A patients. The changes were evident in muscles with normal fat fraction, indicating these biomarkers are sensitive to early disease changes.

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