

**Supplementary Table 2 Clinical inclusion criteria**

Diagnostic category	Criteria
G-IBM	Patients fulfilling Griggs' definite criteria (rimmed vacuoles, inflammatory infiltrate with partial invasion of fibres and 15-18 nm tubulofilaments on EM) with prominent finger flexor and knee extensor weakness and CK <12 x ULN.
IBM+RV	Age at symptom onset >45 years, symptoms present for >12 months, finger flexion strength less than shoulder abduction strength and knee extension weakness greater than hip flexion weakness, CK ≤15 x ULN and a muscle biopsy revealing rimmed vacuoles on H&E or GT stained sections without features inconsistent with IBM on a standard diagnostic histological assessment for an inflammatory myopathy*.
IBM-RV	Clinical features and CK as detailed under IBM+RV. Rimmed vacuoles absent on H&E and GT stained sections and without features inconsistent with IBM on a standard diagnostic histological assessment for an inflammatory myopathy*.
PAM	Genetically or clinically and pathologically confirmed cases of PAM with typical rimmed vacuoles present on muscle biopsy and a genetically confirmed dystrophinopathy with typical rimmed vacuoles and protein aggregates present on muscle biopsy. Cases included myotilinopathy (n=2), hIBM with compound heterozygous mutations in GNE (n=1), IBMPFD with mutation in VCP (n=1), genetically unconfirmed cases of myofibrillar myopathy (n=2), and dystrophinopathy with deletion of exons 45-47 (n=1).
PM&DM	Subacute onset of limb girdle weakness, significantly raised CK, inflammatory cell infiltrate present on muscle biopsy and a sustained unequivocal clinical and biochemical response to steroid immunosuppression. DM cases also had to have cutaneous manifestations consistent with the diagnosis.
Normal controls	Patients investigated for cramps or fatigue, normal clinical examination performed by a muscle specialist, normal CK, normal neurophysiological assessment and normal muscle biopsy.

G-IBM = Griggs' pathologically-definite IBM; IBM+RV = Clinically-typical IBM with rimmed vacuoles; IBM-RV = Clinically typical IBM lacking rimmed vacuoles; PAM = Protein accumulation myopathies with rimmed vacuoles; PM&DM = Steroid-responsive inflammatory myopathies; hIBM = Hereditary inclusion body myopathy; IBMPFD = Inclusion body myopathy with Paget's disease and frontotemporal dementia; CK = Creatine kinase; GT = Gomori trichrome; ULN = Upper limit of normal. \* Standard histological assessment for inflammatory myopathy includes H&E, GT, Sudan black or oil red O, periodic acid Schiff, nicotinamide adenine dinucleotide dehydrogenase, succinate dehydrogenase, cytochrome c oxidase, combined cytochrome c oxidase and succinate dehydrogenase, phosphorylase, acid and alkaline phosphatase, adenylate deaminase, ATPases at pH 4.2/4.3/9.4 and immunohistochemical staining including neonatal myosin, utrophin, major histocompatibility complex class I, membrane attack complex and a combination of inflammatory cell markers.