

Table 1. Clinical and histopathological information. The percentage of fast, slow, developmental, and neonatal myosin-positive fibers was estimated using immunohistochemistry (see Methods)

Diagnosis (severity)	Tissue no.	Age/gender	Autopsy/surgical	% Fast fibers	% Slow fibers	% Dev fibers	% Neo fibers	Muscle type
NM (severe)	T15	3wk/M	A	65	35	38	49	diaphragm
NM (severe)	T24	3wk/M	A	62	38	0	33	psoas
NM (typical)	T26¹	17yr/F	S	0	100	0	0	limb
NM (typical)	T33	1yr/F	S	10	90	0	10	quadriceps
NM (severe)	T39²	1wk/M	A	85	15	80	65	diaphragm
NM (severe)	T41²	1wk/M	A	86	14	74	10	abdominal wall
NM (typical)	T43¹	19yr/M	A	0	100	0	0	skeletal
NM (typical)	T47¹	28yr/F	S	0.2	99.8	0	0.2	quadriceps
NM (severe)	T68	1m/F	A	89	11	8	0	quadriceps
NM (typical)	T80	2m/M	S	62	38	0	61	quadriceps
NM (typical)	T95	15 yr/M	S	10	90	0	0	deltoid
NM (typical)	T99	42yr/F	S	20	80	0	0	tibialis anterior
NM (typical)	T133	35yr/M	A	70	30	0	0	quadriceps
normal	T130	17yr/F	S	55	45	0	0	distal limb
normal	T136	15yr/F	S	60	40	0	0	distal limb
normal	T137	14yr/M	S	50	50	0	0	quadriceps
normal	T140	13yr/M	S	70	30	0	0	limb
normal	T141	2yr/M	S	55	45	0	0	skeletal
normal	T142	2yr/F	S	55	45	0	0	quadriceps
normal	T144	6yr/M	S	50	50	0	0	quadriceps
normal	T145	10yr/M	S	65	35	0	0	quadriceps
normal	T146	1yr/M	S	55	45	0	0	quadriceps
normal	T147	4yr/F	S	40	60	0	0	quadriceps
normal	T148	5yr/F	S	55	45	0	0	quadriceps
normal	T149	4wk/M	A	60	40	0	0	quadriceps
normal	T160	2yr/M	A	40	60	0	0	quadriceps
normal	T169	30yr/M	S	n/a ³	n/a ³	n/a ³	n/a ³	intercostal
normal	T170	65yr/M	A	n/a ³	n/a ³	n/a ³	n/a ³	quadriceps
normal	T172	60yr/M	S	n/a ³	n/a ³	n/a ³	n/a ³	quadriceps
normal	T177	67yr/M	A	35	65	0	0	pectoral
normal	T178	55yr/F	A	40	60	0	0	limb
normal	T179	72yr/M	A	65	35	0	0	limb
normal	T180	58yr/M	S	60	40	0	0	biceps
normal	T181	37yr/F	S	n/a ³	n/a ³	n/a ³	n/a ³	quadriceps

¹These biopsies met criteria for "non typing" cases with large rods in all fibers as described by Ryan *et al.* [(2003), *Neurology* **60**:665-673].

²T39 and T41 are two independent biopsies from the same patient.

³No tissue was available for immunohistochemistry.