

Table e-3: Proteins identified as over-represented in aggregates in MFM-filaminopathy

Accession	Protein	Mean proportion [%] ^a			
		Aggregates	Controls	Ratio	TTEST
Q14315	Filamin-C (FLNC) ^{*,b}	84.21	12.13	6.9	1.9E-05
P17661	Desmin (DES) ^{*,b}	49.95	13.71	3.6	6.7E-08
A4UGR9	Xin actin-binding repeat-containing protein 2 (XIRP2) ^{*,b}	24.42	0.00	n.a.	2.1E-05
Q86VF7	Nebulin-related-anchoring protein (NRAP) ^{*,b}	20.36	0.26	78.1	3.1E-07
P02511	Alpha crystallin B chain (CRYAB) ^{*,c}	13.93	1.15	12.1	3.1E-09
Q702N8	Xin actin-binding repeat-containing protein 1 (XIRP1) ^{*,b}	12.46	0.53	23.6	6.3E-09
Q5VST9	Obscurin (OBSCN) ^{*,b}	12.30	4.49	2.7	0.00016
P48681	Nestin (NES) ^{*,b}	10.85	0.14	77.2	5.7E-07
P12111	Collagen alpha-3(VI) chain (COL6A3) ^{*,d}	8.59	2.08	4.1	0.00081
Q13203	Myosin-binding protein H (MYBPH) ^{*,e}	6.61	0.42	15.9	0.00198
Q9UBF9	Myotilin (MYOT) ^{*,b}	6.60	3.19	2.1	0.00334
P04792	Heat-shock protein beta-1 (HSPB1) ^{*,c}	5.10	0.72	7.1	1.6E-05
O75923	Dysferlin (DYSF) ^{*,f}	4.82	0.58	8.3	0.00091
P62805	Histone H4 (H4C1)	3.99	1.95	2.0	0.03386
P11055	Myosin-3 (MYH3) [*]	3.68	1.24	3.0	0.02186
P02545	Lamin-A/C (LMNA) [*]	2.81	0.60	4.7	0.00655
Q13501	Sequestosome-1 (SQSTM1) ^{*,c}	2.71	0.00	n.a.	0.01148
P06396	Gelsolin (GSN) ^{*,e}	2.65	1.14	2.3	0.00921
P12109	Collagen alpha-1(VI) chain (COL6A1) ^{*,d}	2.21	0.41	5.3	0.02108
P11142	Heat shock cognate 71 kDa protein (HSPA8) ^{*,c}	2.14	0.64	3.3	0.00417
P12110	Collagen alpha-2(VI) chain precursor (COL6A2) ^{*,d}	1.60	0.40	4.0	0.00071
Q15124	Phosphoglucomutase-like protein 5 (PGM5) [*]	1.51	0.00	n.a.	0.01169
Q14764	Major vault protein (MVP) [*]	1.46	0.00	n.a.	0.01671
P08123	Collagen alpha-2(I) chain (COL1A2) ^d	1.43	0.36	4.0	0.02914
P28074	Proteasome subunit beta type-5 (PSMB5) ^{*,c}	1.33	0.05	28.0	0.0048
P54652	Heat shock-related 70 kDa protein 2 (HSPA2) ^{*,c}	1.22	0.48	2.6	0.01947
Q86TC9	Myopalladin (MYPN) ^{*,b}	1.22	0.00	n.a.	0.00088
Q14596	Next to BRCA1 gene 1 protein (NBR1) ^{*,c}	1.12	0.00	n.a.	0.00599
Q5BKX8	Caveolae-associated protein 4 (CAVIN4) ^e	1.10	0.00	n.a.	0.00083
Q9UBY9	Heat shock protein beta-7 (HSPB7) ^{*,c}	1.09	0.05	20.7	0.02386
Q0ZGT2	Nexilin (NEXN) ^{*,b}	1.06	0.16	6.6	0.00081

P13591	Neural cell adhesion molecule 1 (NCAM1)*,^f	1.05	0.00	n.a.	0.00128
Q9UDY4	DnaJ homolog subfamily B member 4 (DNAJB4) ^c	0.88	0.00	n.a.	0.02048
P55268	Laminin subunit beta-2 (LAMB2)*,^d	0.79	0.04	18.4	0.04305
P35580	Myosin-10 (MYH10)	0.76	0.05	15.9	0.01763
Q99623	Prohibitin-2 (PHB2)	0.76	0.40	1.9	0.04289
O95817	BAG family molecular chaperone regulator 3 (BAG3)*,^c	0.75	0.10	7.6	0.01147
Q92614	Unconventional myosin-XVIIIa (MYO18A)	0.70	0.04	16.3	0.01039
O75147	Obscurin-like protein 1 (OBSL1)*,^c	0.69	0.00	n.a.	0.02724
P11047	Laminin subunit gamma-1 (LAMC1)*,^d	0.66	0.00	n.a.	0.00549
Q14204	Cytoplasmic dynein 1 heavy chain 1 (DYNC1H1)*	0.65	0.00	n.a.	0.02304
P28070	Proteasome subunit beta type-4 (PSMB4)*,^c	0.65	0.00	n.a.	0.00154
Q6P5Q4	Leiomodin-2 (LMOD2)*,^e	0.63	0.05	13.3	0.00052
P54289	Voltage-dependent calcium channel subunit alpha-2/delta-1 (CACNA2D1)	0.63	0.09	7.0	0.04957
Q15773	Myeloid leukemia factor 2 (MLF2)	0.62	0.00	n.a.	0.00157
Q14BN4	Sarcolemmal membrane-associated protein (SLMAP) ^f	0.61	0.12	5.0	0.01531
Q9H7C4	Syncoilin (SYNC)*,^b	0.60	0.00	n.a.	0.00524
Q8TCD5	5'(3')-deoxyribonucleotidase, cytosolic type (NT5C)	0.58	0.00	n.a.	0.0004
P05023	Sodium/potassium-transporting ATPase subunit alpha-1 (ATP1A1) ^f	0.57	0.00	n.a.	0.00189
P11532	Dystrophin (DMD)*,^f	0.57	0.00	n.a.	0.03357
O14558	Heat-shock protein beta-6 (HSPB6) ^c	0.56	0.04	13.1	0.00457
O75369	Filamin-B (FLNB)	0.54	0.10	5.2	0.01172
P28289	Tropomodulin-1 (TMOD1)*,^e	0.52	0.05	10.4	0.00307
Q8TDC0	Myozenin-3 (MYOZ3) ^b	0.50	0.17	2.9	0.04084
Q96L58	Beta-1,3-galactosyltransferase 6 (B3GALT6)	0.49	0.00	n.a.	0.00502
Q8IYE0	Coiled-coil domain-containing protein 146 (CCDC146)	0.49	0.00	n.a.	0.00502
O43760	Synaptogyrin-2 (SYNGR2)*	0.48	0.00	n.a.	0.00133
P68371	Tubulin beta-4B chain (TUBB4B)*,^c	0.47	0.00	n.a.	0.00549
P55290	Cadherin-13 (CDH13)	0.47	0.04	10.9	0.0126
Q9NQW7	Xaa-Pro aminopeptidase 1 (XPNPEP1)	0.46	0.00	n.a.	0.0058
Q9Y3D6	Mitochondrial fission 1 protein (FIS1)	0.46	0.05	9.2	0.01633
Q9H992	E3 ubiquitin-protein ligase MARCHF7 (MARCHF7) ^c	0.46	0.00	n.a.	0.03732

Q0D2I5	Intermediate filament family orphan 1 (IFFO1)	0.44	0.00	n.a.	0.00086
P62140	Serine/threonine-protein phosphatase PP1-beta catalytic subunit (PPP1CB)*	0.42	0.00	n.a.	0.01723
P07585	Decorin (DCN)*^d	0.42	0.04	9.8	0.03579
P28072	Proteasome subunit beta type-6 (PSMB6)*^c	0.41	0.00	n.a.	0.00063
P35080	Profilin-2 (PFN2) ^e	0.40	0.00	n.a.	0.02249
Q6ZMU5	Tripartite motif-containing protein 72 (TRIM72)*^f	0.39	0.00	n.a.	0.04713
O75955	Flotillin-1 (FLOT1)*	0.35	0.00	n.a.	0.00956
P02786	Transferrin receptor protein 1 (TFRC) ^f	0.34	0.00	n.a.	0.00282
Q9UJW0	Dynactin subunit 4 (DCTN4)	0.33	0.00	n.a.	0.02568
O00487	26S proteasome non-ATPase regulatory subunit 14 (PSMD14)*^c	0.33	0.00	n.a.	0.00299
P09382	Galectin-1 (LGALS1)	0.31	0.00	n.a.	0.0329
P25788	Proteasome subunit alpha type-3 (PSMA3)*^c	0.29	0.00	n.a.	0.03593
Q9UKS6	Protein kinase C and casein kinase substrate in neurons protein 3 (PACSIN3)*	0.29	0.00	n.a.	0.00273
Q15286	Ras-related protein Rab-35 (RAB35)*^e	0.29	0.00	n.a.	0.00273
Q96CG8	Collagen triple helix repeat-containing protein 1 (CTHRC1) ^d	0.24	0.00	n.a.	0.01549
P16930	Fumarylacetoacetase (FAH)	0.23	0.00	n.a.	0.01725
Q12988	Heat shock protein beta-3 (HSPB3)*^c	0.23	0.00	n.a.	0.01725
Q9BSH3	Nicolin-1 (NICN)	0.23	0.00	n.a.	0.01725
P60900	Proteasome subunit alpha type-6 (PSMA6)*^c	0.23	0.00	n.a.	0.01725
Q53FT3	Protein Hikeshi (HIKESHI) ^c	0.23	0.00	n.a.	0.01725
Q92629	Delta-sarcoglycan (SGCD)*^f	0.22	0.00	n.a.	0.01708
P25686	DnaJ homolog subfamily B member 2 (DNAJB2)*^c	0.22	0.00	n.a.	0.01686
P11279	Lysosome-associated membrane glycoprotein 1 (LAMP1)*^c	0.22	0.00	n.a.	0.01686
Q7Z7L7	Protein zer-1 homolog (ZER1) ^c	0.22	0.00	n.a.	0.01686
Q08257	Quinone oxidoreductase (CRYZ)	0.22	0.00	n.a.	0.01686
Q00765	Receptor expression-enhancing protein 5 (REEP5)	0.22	0.00	n.a.	0.01686

n.a. not applicable. ^aper mill of total spectral counts. ^{*}validated by immunolocalization studies. ^{b-f}assigned to (see Fig. 4): ^bZ-disc (associated) proteins, ^cPQC and protein degradation, ^dECM and BL proteins, ^eRho//ROCK pathway / actin dynamics, ^fsarcolemmal proteins.