



Ref	ALS Model/Sample Type	Differentially Expressed Proteins	Proteins included in Comparison	Analysis Platform	Protein Database
[20]	Patient CSF	35	35	iTRAQ; API QSTAR XL (Applied Biosystems)	UniProt/Swiss-Prot
[31]	Patient CSF	123	110	Label-Free; linear ion trap (ThermoFisher Scientific)	UniProt/Swiss-Prot
[32]	Patient CSF	48	47	iTRAQ; LTQ-Orbitrap Velos (Thermo Electron)	Human Protein Database (NCBI)
[33]	Patient CSF	17 *	17	2D electrophoresis; Voyager-DE Pro MALDI-TOF (Applied Biosystems)	NCBI and UniProt/Swiss-Prot
[34]	Patient CSF	153	11	Peak recognition (statistical analysis); linear Microflex MALDI-TOF mass spectrometer (Bruker)	Entrez Protein Database
[35]	Patient CSF	33	7	SELDI-TOF-MS	Empirical Proteomics Ontology Knowledge Base
[36]	Patient CSF	6	5	2D-DIGE; Voyager DE-STR (Applied Biosystems)	NCBI database
[37]	Patient CSF	52 ⁺	3	SELDI-TOF-MS/MS; Ciphergen ProteinChip Reader (Ciphergen Biosystems)	UniProt/Swiss-Prot
[38]	Patient CSF	10 (PM-ALS) † 9 (L-ALS) †	2	SELDI-TOF-MS; Ciphergen ProteinChip Reader (Ciphergen Biosystems)	UniProt/Swiss-Prot
[39]	Patient CSF	3	2	SELDI-TOF-MS	Unknown
[40]	Patient CSF	3	3	UHPLC LC-MS/MS; Q Exactive HF tandem mass spectrometer (Thermo Fisher Scientific)	UniProt
[41]	Patient serum	7	7	2D-GE; SYNAPT-MS G1 (Waters Corporation)	UniProt
[42]	Rat neurons/TDP-43 Knockdown	63	63	Label-free; Orbitrap Q Exactive (Thermo Fisher Scientific)	UniProt/Swiss-Prot
[43]	Patient CSF injected into rat spinal cord /mitochondrial fraction	49	48	iTRAQ; LTQ-Orbitrap Velos (Thermo Electron)	RefSeq
[44]	SH-SY5Y cells/TDP-43 Loss	273	270	Label-free; LTQ Orbitrap XL (Thermo Fisher Scientific)	UniProt/Swiss-Prot
[45]	Patient muscle	5	5	2D-GE; API QStar PULSAR (AB-Sciex)	UniProt/Swiss-Prot
[46]	Patient muscle	11	11	Stable-isotope dimethyl labels; 7 T hybrid LTQ FT Ultra mass spectrometer (ThermoFisher Scientific)	UniProt/Swiss-Prot
[47]	Mouse astrocytes/SOD1 G93A	31	31	2D-GE: Reflex III MALDI-TOF (Bruker Daltonics)	UniProt/Swiss-Prot

Table S1. ALS proteomic studies used in the comparison.

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[48]	Patient blood mononuclear cells	44	44	2D DIGE; 4800 MALDI TOF/TOF (Applied Biosystems)	UniProt/Swiss-Prot
[49]	Mouse spinal cord/Wobbler Mouse	13	13	2D DIGE; Model 6430 Ion Trap (Agilent Technologies)	NCBI
[50]	Mouse ventral roots/SOD1 G93A	14 #	14	Label Free; LTQ (Thermo Finnigan)	UniProt/Swiss-Prot
[51]	Mouse embryonic motor neurons/SOD1 G85R	6	4	2D-GE; UltraFlex MALDI TOF/TOF (Bruker- Franzen Analytik)	UniProt/Swiss-Prot
[52]	Mouse spinal cord/SOD1 G127X	53	47	2D-DIGE; Voyager DE-STR MALDI-TOF (Applied Biosystems)	UniProt/Swiss-Prot
[53]	Mouse muscle/Wobbler Mouse	31	26	2D-GE; Reflex III MALDI-TOF (Bruker Daltonics)	UniProt/Swiss-Prot
[54]	Mouse spinal cord (insoluble fraction)/SOD1 G93A	32	32	2DE; Reflex III MALDI-TOF (Bruker Daltonics)	UniProt/Swiss-Prot
[55]	Mouse spinal cord (pre- symptomatic)/SOD1 G93A	15	15	2D-GE; ReflexIII MALDI mass spectrometer (Bruker Daltonics)	UniProt/Swiss-Prot
[56]	Mouse spinal cord/SOD1 G93A	7	7	2D-GE; VG 2E Tofspec laser desorption time of flight mass spectrometer (Waters)	NCBI
[57]	NSC34 cells (mitochondrial fraction)/SOD1 G93A	40	38	2D-GE; Qstar XL Q-TO, Applied Biosystems	NCBI and UniProt/Swiss-Prot
[58]	Rat spinal cord (mitochondrial fraction)/SOD1 G93A	40	40	2D-GE; MudPIT RCADiA platform	Saccharomyces Genome Database/ Ensembl
[59]	NSC34 cells/SOD1 G93A/G37R	7	7	2D-GE; Voyager-DE STR, Perspective Biosystems	UniProt/Swiss-Prot
[60]	Mouse hippocampus/Tg152 line overexpressing hSOD1	41	41	2D-GE; Ultraflex MALDI TOF/TOF (Bruker Daltonics)	NCBI and UniProt/Swiss-Prot
[61]	Patient anterior and posterior horn	32 (anterior horn) 3 (posterior horn)	18 (anterior horn) 3 (posterior horn)	2D-GE; LTQ linear ion trap mass spectrometer (ThermoFisher Scientific)	UniProt
[62]	Patient prefrontal cortex	103	101	Label-free; Fusion mass spectrometer (ThermoFisher Scientific)	UniProt

ALS studies included in the proteomic comparison are listed in table, together with ALS model, sample type, analysis platform and database used in each study. Number of differentially expressed proteins identified in each study and number of those proteins that were included in the comparison are shown in column three and four. * Proteins were considered significantly changed if they were identified in ALS samples, but not in CTR samples. ⁺ Significantly changed proteins in ALS were identified by applying second-pass peak selection with a signal to noise ratio of 1.5 and p < 0.01 [8,9]. [#] By dynamically adjusting the *p*-value cut-off, *p*-value of 0.05 was chosen for the identification of significantly changed proteins in ALS. PM-ALS—Samples from ALS patients taken post-mortem; L-ALS—Samples from living patients; CSF—Cerebrospinal Fluid.

Protein Name	No. of	Direction of	Pofe
(Official Gene Symbol)	Studies	Expression Change	Kels
Cystatin C (CST3)	4	Down	[35,37–39]
Alpha-1-Acid Glycoprotein 1 (ORM1)	2	Down	[20,32]
Apolipoprotein A-IV (APOA4)	2	Down	[34,41]
Haptoglobin (HP)	2	Down	[20,32]
Neurosecretory Protein VGF (VGF)	2	Down	[34,39]
Ribonuclease Pancreatic (RNASE1)	2	Down	[20,35]
Transferrin (TF)	2	Down	[32,36]
Chitinase 3-Like Protein 1 (CHI3L1)	3	Up	[32,34,41]
Chitinase 3-Like Protein 2 (CHI3L2)	2	Up	[31,32]
Chitotriosidase-1 (CHIT1)	2	Up	[32,41]
Hemoglobin Subunit Alpha 1 (HBA1)	2	Up	[20,35]

Table S2. Proteins differentially ex	xpressed in the same direct	ction in biofluids from A	ALS patients.
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Proteins with a consistent change in expression in biofluids from ALS patients across two or more proteomic studies are shown. The protein name is given, followed by the official gene symbol in brackets.

Table S3. Proteins that showed contradictory direction of differential expression in biofluids from ALS patients across at least two proteomic studies.

Protein Name (Official Gene Symbol)	No. of Studies	Direction of Differential Expression	Refs
Trees at hours the (TTD)	4	Down	[37,38,41]
Transtnyretin (TTR)	4	Up	[35] *
Homoglobin subunit bota (HBB)	2	Up	[20,35]
Tientogiobin subunit beta (TIBB)	3	Down	[32]
Protoin AMRD (AMRD)	2	Down	[20,34]
	3	Up	[41]
Applipaprotoin $A = I (A P O A 2)$	2	Down	[41]
Apolipoprotein A-II (AI OA2)	2	Up	[20]
Applipaprotain B 100 (APOR)	2	Down	[31]
Aponpoprotent b-100 (Ar Ob)	2	Up	[32]
Prothrombin (E2)	2	Down	[34]
r rounombin (F2)	2	Up	[32]
Tactican 2 (SPOCV2)	2	Down	[31]
Testican-2 (SPOCK2)	Z	Up	[20]
Zing Alpha 2 Chappendoin (AZCP1)	2	Down	[20]
Zinc-Aipna-2-Giycoprotein (AZGP1)	Z	Up	[36]

Proteins that showed contradictory change in expression in biofluids from ALS patients in two or more proteomic studies. Protein name is given, followed by official gene symbol in brackets. Direction of differential expression and number of studies that identified protein changes are shown (studies are listed in the reference column). * CysGly-transthyretin—Modified form of transthyretin generated by oxidative damage.



Figure S1. Bioinformatics analysis of eleven proteins that showed consistent change in expression across biofluids from ALS patients. Gene ontology analysis revealed enriched terms connected to (**A**) biological process, (**B**) cellular component. Terms are presented as bars, with the white numbers inside the bars indicating number of annotated proteins. (**C**) Search Tool for the Retrieval of Interacting Genes/Proteins (STRING) 10 analysis identified association network between four proteins: HBA1—Haemoglobin Subunit Alpha 1, HP—Haptoglobin, ORM1—Alpha-1-Acid Glycoprotein 1 and TF-transferrin. Type of the association between proteins is indicated by the colour (pink: experimentally determined interactors; light blue: interactors form curated databases; black: c890-expression; yellow: text-mining).

Protein (Official Gene Symbol)	No. of Studies	Refs	Sample Type	ALS Model
		Incre	eased expression	
		[57]	NSC34 cells (mitochondrial fraction)	SOD1 G93A
		[53]	Mouse muscle	Wobbler mouse
Aldolase A (ALDOA)	5	[49]	Mouse spinal cord	Wobbler mouse
		[48]	Patient blood mononuclear cells	NA
		[47]	Mouse astrocytes	SOD1 G93A
		[52]	Mouse spinal cord	SOD1 G127X
Superoxide Dismutase 2	4	[58]	Rat spinal cord (mitochondrial fraction)	SOD1 G93A
(SOD2)	_	[48]	Patient blood mononuclear cells	NA
		[44]	SH-SY5Y cells	TDP-43 Loss
		[56]	Mouse spinal cord	SOD1 G93A
Superoxide Dismutase 1 (SOD1)	3	[54]	Mouse spinal cord	SOD1 G93A
· · · ·		[53]	Mouse muscle	Wobbler mouse
Peroxiredoxin 2 (PRDX2)	3	[51]	Mouse embryonic motor neurones	SOD1 G85R
		[48]	Patient blood mononuclear cells	NA
		[44]	SH-SY5Y cells	TDP-43 Loss
	3	[54]	Mouse spinal cord	SOD1 G93A
14-3-3 Protein Gamma (YWHAG)		[52]	Mouse spinal cord	SOD1 G127X
		[44]	SH-SY5Y cells	TDP-43 Loss
	3	[56]	Mouse spinal cord	SOD1 G93A
Heat Shock Protein 1 (HSPB1)		[54]	Mouse spinal cord	SOD1 G93A
		[44]	SH-SY5Y cells	TDP-43 Loss
		[50]	Mouse ventral roots	SOD1 G93A
Calreticulin (CALR)	3	[48]	Patient blood mononuclear cells	NA
	-	[44]	SH-SY5Y cells	TDP-43 Loss
Heat Shock Protein		[54]	Mouse spinal cord	SOD1 G93A
Family A, Member 8	3	[48]	Patient blood mononuclear cells	NA
(H5ľA8)		[44]	SH-SY5Y cells	TDP-43 Loss
	2	[56]	Mouse spinal cord	SOD1 G93A
reroxiredoxin 6 (PKDX6)	3	[48]	Patient blood mononuclear cells	NA

Table S4. Proteins that showed consistent direction of differential expression in ALS cells and tissues across two or more proteomic studies.

		[49]	Mouse spinal cord	Wobbler mouse
		[54]	Mouse spinal cord	SOD1 G93A
Glial Fibrillary Acidic Protein (GFAP)	3	[47]	Mouse astrocytes	SOD1 G93A
· · · · · ·		[62]	Patient prefrontal cortex	NA
Glyceraldehyde-3-		[54]	Mouse spinal cord	SOD1 G93A
Phosphate Dehvdrogenase	3	[53]	Mouse muscle	Wobbler mouse
(GAPDH)		[49]	Mouse spinal cord	Wobbler mouse
Valosin Containing	0	[54]	Mouse spinal cord	SOD1 G93A
Protein (VCP)	2	[46]	Patient skeletal muscle	NA
	0	[50]	Mouse ventral roots	SOD1 G93A
Septin 9 (SEP19)	2	[44]	SH-SY5Y cells	TDP-43 Loss
Malate Dehydrogenase 1	0	[54]	Mouse spinal cord	SOD1 G93A
(MDH1)	2	[52]	Mouse spinal cord	SOD1 G127X
Actin, Alpha Cardiac	2	[53]	Mouse muscle	Wobbler mouse
Muscle 1 (ACTC1)	2	[44]	SH-SY5Y cells	TDP-43 Loss
NADH dehydrogenase		[57]	NSC34 cells (mitochondrial fraction)	SOD1 G93A
protein 8, mitochondrial (NDUFS8)	2	[61]	Patient spinal cord	NA
Columonin (CALU)	2	[50]	Mouse ventral roots	SOD1 G93A
Calumentin (CALO)		[44]	SH-SY5Y cells	TDP-43 Loss
Creatine Kinase,	C	[54]	Mouse spinal cord	SOD1 G93A
(CKMT1B)	Z	[49]	Mouse spinal cord	Wobbler mouse
Glutamate	C	[54]	Mouse spinal cord	SOD1 G93A
Dehydrogenase (GLUD1)	2	[49]	Mouse spinal cord	Wobbler mouse
Apolipoprotein E	C	[56]	Mouse spinal cord	SOD1 G93A
(APOE)	Z	[50]	Mouse ventral roots	SOD1 G93A
Enoyl Coenzyme A	2	[55]	Mouse spinal cord (pre- symptomatic)	SOD1 G93A
(ECHS1)	2	[47]	Mouse astrocytes	SOD1 G93A
Single Stranded DNA Binding Protein 1	2	[58]	Rat spinal cord (mitochondrial fraction)	SOD1 G93A
(SSBP1)	۷ -	[44]	SH-SY5Y cells	TDP-43 Loss
Glycerol Phosphate	2	[54]	Mouse spinal cord	SOD1 G93A
Dehydrogenase 2 (GPD2)	<u> </u>	[49]	Mouse spinal cord	Wobbler mouse

	2 -	[54]	Mouse spinal cord	SOD1 G93A
Aldolase C (ALDOC)		[47]	Mouse astrocytes	SOD1 G93A
Glutamine Ammonia		[54]	Mouse spinal cord	SOD1 G93A
Ligase (GLUL)	2	[49]	Mouse spinal cord	Wobbler mouse
Endoplasmic Reticulum	2	[52]	Mouse spinal cord	SOD1 G127X
Protein 29 (ERP29)	2	[48]	Patient blood mononuclear cells	NA
Isocitrate Dehydrogenase		[54]	Mouse spinal cord	SOD1 G93A
2 (IDH2)	2	[58]	Rat spinal cord (mitochondrial fraction)	SOD1 G93A
Lactate Dehydrogenase B	2	[54]	Mouse spinal cord	SOD1 G93A
(LDHB)	2	[48]	Patient blood mononuclear cells	NA
		Decr	eased expression	
Neurosecretory protein	2	[44]	SH-SY5Y cells	TDP-43 Loss
VGF (VGF)	Z	[62]	Patient prefrontal cortex	NA
Myosin Light Chain,	2	[50]	Mouse ventral roots	SOD1 G93A
Skeletal Muscle (MYLPF)	2	[45]	Patient skeletal muscle	NA
Nudix Motif 2 (NUDT2)	2	[52]	Mouse spinal cord	SOD1 G127X
		[43]	Rat spinal cord (mitochondrial fraction)	Patient CSF injected
Adenylate Kinase 2	2	[57]	NSC34 cells (mitochondrial fraction)	SOD1 G93A
(AK2)		[58]	Rat spinal cord (mitochondrial fraction)	SOD1 G93A
Far Upstream Element-	r	[48]	Patient blood mononuclear cells	NA
(FUBP1)	2	[44]	SH-SY5Y cells	TDP-43 Loss
		[48]	Patient blood mononuclear cells	NA
Fliamin A (FLINA)	2	[47]	Mouse astrocytes	SOD1 G93A
Calain P1 (COL CP1)	2	[48]	Patient blood mononuclear cells	NA
Golgin B1 (GOLGB1)	2	[44]	SH-SY5Y cells	TDP-43 Loss
Heat Shock Protein Family D. Member 1	2	[58]	Rat spinal cord (mitochondrial fraction)	SOD1 G93A
(HSPD1)	2	[44]	SH-SY5Y cells	TDP-43 Loss
Acyl- Coenzyme A	2	[57]	NSC34 cells (mitochondrial fraction)	SOD1 G93A
Chain (ACADM)	2 -	[46]	Patient skeletal muscle	NA
Nadh Dehydrogenase 1	2	[57]	NSC34 cells (mitochondrial fraction)	SOD1 G93A
Beta Subcomplex Subunit 10 (NDUFB10)	2	[44]	SH-SY5Y cells	TDP-43 Loss
Solute Carrier Family 25, Member 4 (SLC25A4)	2	[58]	Rat spinal cord (mitochondrial fraction)	SOD1 G93A

		[44]	SH-SY5Y cells	TDP-43 Loss
Ubiquinol-Cytochrome C	n	[47]	Mouse astrocytes	SOD1 G93A
(UQCRC1)	Z	[44]	SH-SY5Y cells	TDP-43 Loss
Chaperonin Containing	2	[57]	NSC34 cells (mitochondrial fraction)	SOD1 G93A
Tcp1 Subunit 6a (CCT6A)	Z	[44]	SH-SY5Y cells	TDP-43 Loss
2',3'-Cyclic Nucleotide 3	eotide 3	[52]	Mouse spinal cord	SOD1 G127X
Phosphodiesterase (CNP)	2	[44]	SH-SY5Y cells	TDP-43 Loss

Proteins that showed consistent direction of differential expression in cells and tissues in at least two proteomic studies of ALS. Proteins are listed according to direction of differential expression (proteins with increased expression are listed first, followed by proteins with decreased expression). Protein name is given, followed by official gene symbol in brackets. ALS model and sample type used in each study are listed, together with the number of studies that identified protein changes (studies are listed in the reference column).

	Increased	Decreased
Protein (Official Gene Symbol)	Expression	Expression
Prolyl 4-Hydroxylase Subunit Beta (P4HB)	[48,50,54,55]	[47]
Phosphoglycerate Mutase 1 (PGAM1)	[48,49,52]	[44,55,60]
Peptidylprolyl Isomerase A (PPIA)	[48,54,55]	[47]
Dihydropyrimidinase-Like 2 (DPYSL2)	[49,54]	[47,52,60]
Pyruvate Kinase, Muscle (PKM)	[49,54]	[47,60]
NADH Dehydrogenase Fe-S Protein 1 (NDUFS1)	[54]	[58,60]
Enolase 1 (ENO1)	[54,55]	[47,48]
ATP Synthase, H+ Transporting, Mitochondrial F1 Complex,	[= 4]	[47 57 (0]
Alpha Subunit 1 (ATP5A1)	[54]	[47,57,60]
Tubulin Alpha 1b (TUBA1B)	[44,60]	[57]
Voltage-Dependent Anion Channel 1 (VDAC1)	[47]	[52,57]
Transketolase (TKT)	[44,49]	[47]
Pyruvate Dehydrogenase E1 Alpha 1 (PDHA1)	[54,55]	[57]
4-Aminobutyrate Aminotransferase (ABAT)	[49,58]	[52]
Atp Synthase, H+ Transporting, Mitochondrial F1 Complex, Beta Polypeptide (ATP5B)	[48,60]	[44,48]
Aconitase 2 (ACO2)	[49.54.57]	[57]
Vimentin (VIM)	[44,47,54]	[47]
Heat Shock Protein Family A, Member 5 (HSPA5)	[61]	[47,48,57]
Actin Beta (ACTB)	[47,48]	[62]
Phosphoglycerate kinase 1 (PGK1)	[48,49,60]	[48,49]
Tubulin beta-2B chain (TUBB2B)	[44,60]	[62]
Glutathione S-transferase Mu 1 (GSTM1)	[62]	[59]
Dipeptidyl peptidase 3 (DPP3)	[61]	[62]
Prohibitin (PHB)	[47]	[58]
Tubulin, Beta 4b (TUBB4B)	[47]	[44]
Electron Transferring Flavoprotein, Alpha (ETFA)	[55]	[52]
Actin, Gamma, Cytoplasmic 1 (ACTG1)	[52]	[57]
Annexin A5 (ANXA5)	[54]	[52]
Creatine Kinase, Muscle (CKM)	[53]	[50,53]
Crystallin, Alpha B (CRYAB)	[54]	[47]
Growth Factor Receptor-Bound Protein 2 (GRB2)	[44]	[52]
Hexosaminidase Subunit Alpha (HEXA)	[50]	[43]
Heat Shock Protein 90 Alpha Family Class A Member 1 (HSP90A A1)	[54]	[44]
Isocitrate Dehydrogenase 3 (NAD+) Beta (IDH3B)	[58]	[57]
Heat Shock Protein 90, Beta (Grp94), Member 1 (HSP90B1)	[54]	[47]
Mitogen-Activated Protein Kinase 1 (MAPK1)	[54]	[47]
Lon Peptidase 1 (LONP1)	[44]	[57]
Dynamin 1 (DNM1)	[52,62]	[60]
2 4-Dienovl Coa Reductase 1 (DECR1)	[58]	[44]

Table S5. Proteins that showed contradictory direction of differential expression in cells and tissues across two or more proteomic studies of ALS.

Proteins that showed contradictory direction of differential expression in cells and tissues in at least two proteomic studies of ALS. Protein name is given, followed by official gene symbol in brackets. Studies that identified increased or decreased protein expression are listed in column two and three. **Table S6.** GO term analysis of proteins that were differentially expressed in tissues and cells in ALS proteomic studies.

Enriched Term	Count	Proteins
		Biological process
Programmed cell death	15	ACTC1, APOE, CALR, ERP29, FLNA, GAPDH, HSPB1, HSPD1, PRDX2, SOD1, SOD2, YWHAG, VCP, SLC25A4, NUDT2
Protein transport and localization	13	VGF, APOE, CALR, CCT6A, ERP29, FLNA, GLUD1, GLUL, HSPA8, HSPD1, SLC25A4, YWHAG, VCP
Nucleotide metabolism	13	CNP, NDUFS8, NDUFB10, AK2, ALDOA, ALDOC, APOE, GAPDH, HSPA8, LDHB, MDH1, UQCRC1, VCP
Carbohydrate derivative metabolism	11	NDUFS8, NDUFB10, AK2, ALDOA, ALDOC, APOE, GAPDH, GDP2, HSPA8, UQCRC1, VCP
Organic acid metabolism	10	ACADM, ALDOA, ALDOC, ECHS1, GLUD1, GLUL, GAPDH, IDH2, LDHB, MDH1
Protein complex assembly	10	NDUFS8, NDUFB10, ALDOA, CALR, GLUL, HSPA8, HSPD1, SEPT9, SOD2, VCP
Cytoskeleton organization	9	CNP, ACTC1, ALDOA, APOE, CALR, FLNA, GFAP, GAPDH, SOD1
Response to oxidative stress	9	NDUFS8, APOE, HSPB1, HSPD1, NUDT2, PRDX2, PRDX6, SOD1, SOD2
ATP metabolism	9	NDUFS8, NDUFB10, AK2, ALDOA, ALDOC, GAPDH, HSPA8, UQCRC1, VCP
Cell adhesion	9	ALDOA, CALR, FLNA, HSPA8, HSPB1, HSPD1, PRDX6, SEPT9, SOD1
Carbohydrate metabolism	9	ACADM, ALDOA, ALDOC, GAPDH, GPD2, IDH2, LDHB, MDH1, VCP
Mitochondrion organization	8	CNP, NDUFS8, NDUFB10, HSPD1, SSBP1, SLC25A4, SOD2, YWHAG
Cellular amide metabolic process	7	APOE, CALR, GAPDH, HSPB1, SLC25A4, SOD1, SOD2
Cellular respiration	7	NDUFS8, NDUFB10, IDH2, MDH1, SOD2, UQCRC1, VCP
Synaptic signaling	7	CNP, VGF, APOE, GFAP, GLUL, HSPA8, YWHAG
Protein stability	6	CALR, CCT6A, FLNA, GAPDH, HSPA8, HSPD1
Protein secretion	6	VGF, ERP29, GLUD1, GLUL, HSPD1, SLC25A4
NADH metabolism	6	ALDOA, ALDOC, GAPDH, LDHB, MDH1, VCP
Response to wounding	6	APOE, FLNA, GFAP, HSPB1, SOD1, SOD2
Response to unfolded protein	5	CALR, HSPA8, HSPB1, HSPD1, VCP
Gluconeogenesis	5	ALDOA, ALDOC, GAPDH, GPD2, MDH1
Regulation of protein kinase activity	5	APOE, ERP29, HSPB1, SOD1, YWHAG
Muscle contraction	5	ACTC1, ALDOA, FLNA, MYLPF, SOD1
Response to drug	5	ACTC1, CALR, HSPD1, SOD1, SOD2
Blood circulation	5	ACTC1, APOE, FLNA, SOD1, SOD2
Response to temperature stimulus	5	VGF, ACADM, HSPD1, SOD1, SOD2
Striated muscle tissue development	4	ACTC1, ACADM, CALR, MYLPF
Hormone secretion	4	VGF, GLUD1, GLUL, SLC25A4
Lipid catabolic process	4	ACADM, APOE, ECHS1, PRDX6
Aging	4	CNP, CALR, SOD1, SOD2
Glycolytic process	3	ALDOA, ALDOC, GAPDH
Cardiac muscle cell differentiation	3	ACTC1, ACADM, CALR

Response to starvation	3	ACADM, GLUL, HSPA8
		Cellular component
		CNP, NDUFB10, ACTC1, ACADM, AK2, ALDOA, ALDOC.
		APOE, CALR, CCT6A, ERP29, ECHS1, FLNA, GLUL, GAPDH,
Extracellular vesicle	28	HSPA8, HSPB1, HSPD1, IDH2, LDHB, MDH1, PRDX2, PRDX6,
		SSBP1 SOD1 SOD2 YWHAG VCP
		CNP NDUESS NDUEB10 ACADM AK2 ALDOC ECHS1
Mitochondrion	21	CLUD1 CLUL CDP2 HSPD1 JDH2 LDHB MDH1 NUDT2
witteenonarion	21	CEDDI, GEOL, GDI 2, HISEDI, IDH2, EDHD, MDH1, NOD12, CERDI SI CIEAA SODI SODI VIALA CHOCRCI
		ACTC1 ALDOA ALDOC CALD CCT(A FLNA CEAD
	01	ACICI, ALDOA, ALDOC, CALK, CCI6A, FLNA, GFAP,
Cytosol	21	GLUL, GAPDH, HSPA8, HSPB1, HSPD1, IDH2, LDHB, MDH1,
		MYLPF, PRDX2, PRDX6, SOD1, YWHAG, VCP
Myelin sheath	13	CNP, GFAP, GLUL, HSPA8, HSPD1, LDHB, MDH1, SLC25A4,
	15	SOD1, SOD2, YWHAG, UQCRC1, VCP
Cutoplasmia vosiala	10	CNP, VGF, ALDOA, APOE, CALR, CALU, ERP29, HSPA8,
Cytoplasmic vesicle	12	HSPD1, PRDX6, SOD1, YWHAG
		CNP, ACTC1, ALDOA, ALDOC, CCT6A, FLNA, GFAP,
Cytoskeleton	12	GAPDH, HSPB1, MDH1, MYLPF, SEPT9
		CNP VCF ACTC1 ALDOA APOF CALR HSPA8 HSPB1
Extracellular space	12	HSPD1 MDH1 PRDY6 SOD1
Cell projection	10	CNP, ACICI, ACADM, AK2, APOE, FLNA, GFAP, GLUL,
		SEP19, SODI
Cell junction	9	ACTC1, ALDOA, CALR, FLNA, HSPA8, HSPB1, PRDX6,
	-	SEPT9, YWHAG
Extra collular matrix	0	APOE, CALR, CCT6A, FLNA, GAPDH, HSPA8, HSPB1,
	9	HSPD1, SOD1
Perinuclear region of		
cytoplasm	6	CNP, CALR, FLNA, GAPDH, SEP19, VCP
Cell body	6	ACTC1, APOE, FLNA, GFAP, GLUL, SOD1
Contractile fiber	5	ACTC1 ALDOA FLNA HSPB1 MYLPF
Lysosome	5	CEAP HSPA8 MVI PE PRDY6 SOD1
Dereviseme	1	
Peroxisome	4	ACADM, HSPDI, IDH2, SODI
Oxidoreductase complex	4	NDUF58, NDUFBI0, GDP2, UQCRCI
Pigment granule	4	CNP, CALU, ERP29, HSPA8
Respiratory chain complex	3	NDUFS8, NDUFB10, UQCRC1
		Molecular function
		CNP, NDUFS8, NDUFB10, ACTC1, ACADM, AK2, ALODA,
	07	ALDOC, ERP29, ECHS1, GLUD1, GLUL, GAPDH, GPD2,
Catalytic activity	27	HSPA8, HSPD1, IDH2, LDHB, MDH1, NUDT2, PRDX2,
		PRDX6, SEPT9, SOD1, SOD2, UOCRC1, VCP
		CNP ACTC1 ACADM AK2 CCT6A GLUD1 GLUL
Nucleotide binding	16	CAPDH HSPA8 HSPD1 IDH2 I DHB MDH1 NUDT2
Nucleonae binding	10	CEDTO VCD
		ACTCL AV2 ADOL CALD CCT(A FLNA CEAD CLUD)
Carbonydrate derivative	14	ACICI, AK2, APOE, CALK, CCI6A, FLNA, GFAP, GLUDI,
binding		GLUL, HSPA8, HSPDI, NUD12, SEP19, VCP
Identical protein binding	14	ACADM, ALDOA, APOE, ERP29, FLNA, GFAP, GLUD1,
		GLUL, GAPDH, HSPB1, LDHB, SOD1, SOD2, VCP
Ovidoroductoro activity	14	NDUFS8, NDUFB10, ACADM, GLUD1, GAPDH, GDP2, IDH2,
	14	LDHB, MDH1, PRDX2, PRDX6, SOD1, SOD2, UQCRC1
	10	CNP, ALDOA, CALR, CCT6A, FUBP1, FLNA, GOLGB1,
KNA and DNA binding	13	HSPA8, HSPB1, HSPD1, SSBP1, YWHAG, VCP
		CALR, FLNA, GFAP, HSPA8, HSPB1, HSPD1, LDHB, PRDX6
Enzyme binding	12	SODI YWHAC LIOCRCI VCP
Coll adhesion malagula		
	7	ALDOA, CALR, FLNA, GFAP, HSPA8, PRDX6, SEPT9
binding		
Cytoskeletal protein	6	ACTC1, ALDOA, ALDOC, APOE. FLNA. GAPDH
binding	-	, - ,,,,

Coenzyme binding	6	ACADM, GLUD1, GAPDH, IDH2, LDHB, MDH1
Antioxidant activity	5	APOE, PRDX2, PRDX6, SOD1, SOD2
Unfolded protein binding	4	CALR, CCT6A, HSPA8, HSPD1
Chaperone binding	4	CALR, ERP29, HSPD1, SOD1
Glycoprotein binding	3	CALR, FLNA, GFAP

Enriched terms connected to "Biological process", "Cellular component" and "Molecular function" are presented together with the number of annotated proteins. Proteins are listed in column three.

Table S7. GO terr	n analysis	of	proteins	that	were	differentially	expressed	in	SMA	and	ALS
proteomic studies.											

Enriched term	Count	Proteins
	Biological	process
Drogeneration of call doubt	10	ANXA5, CALR, HSP90B1, HSPD1, PRDX2,
Programmed cell death	10	SOD1, VDAC1, GAPDH, HSP90AA1, YWHAG
Establishment of localization	10	CNP, ATP5A1, ALDOA, CALR, HSP90AA1,
Establishment of localization	10	HSP90B1, HSPD1, SOD1, YWHAG, VDAC1
Base and to show incl	10	ANXA5, CALR, HSP90B1, HSPD1, PRDX2,
Response to chemical	10	SOD1, CNP, GAPDH, HSP90AA1, YWHAG
Demilation of anotain matchelian	7	CALR, HSP90B1, HSP90AA1, YWHAG,
Regulation of protein metabolism	1	GAPDH, HSPD1, SOD1
	-	ALDOA, CALR, HSP90B1, SOD1, VIM, CNP,
Cytoskeleton organization	1	GAPDH
Nervous system development	6	CNP, CALR, SOD1, HSP90AA1, YWHAG, VIM
Bogulation of immuno system	6	CALR, HSP90B1, HSPD1, SOD1, GAPDH,
Regulation of Immune System	6	HSP90AA1
Vesicle-mediated transport	5	ALDOA, CALR, HSP90B1, SOD1, HSP90AA1
Metabolism of reactive oxygen species	5	HSPD1, PRDX2, SOD1, VDAC1, HSP90AA1
Oxidation-reduction process	5	ALDOA, PRDX2, PGK1, SOD1, GAPDH
Mitochondrion organization	5	CNP, HSP90AA1, HSPD1, YWHAG, VDAC1
Mitochondrial transport	5	CNP, ATP5A1, HSP90AA1, YWHAG, VDAC1
Cellular homeostasis	5	ALDOA, CALR, HSP90B1, PRDX2, SOD1
Nucleotide metabolism	5	ALDOA, GAPDH, PGK1, ATP5A1, CNP
Intracellular protein transport	4	CALR, HSP90B1, HSP90AA1, YWHAG
Epithelial cell differentiation	4	PGK1, SOD1, VIM, VDAC1
Response to unfolded protein	4	CALR, HSP90B1, HSPD1, HSP90AA1
ATP metabolism	4	ALDOA, GAPDH, PGK1, ATP5A1
Muscle contraction	4	ALDOA, SOD1, VIM, HSP90AA1
Glucose metabolism	3	ALDOA, GAPDH, PGK1
Response to ATP	3	HSP90B1, HSPD1, SOD1
NADH metabolism	3	ALDOA, GAPDH, PGK1
Response to temperature stimulus	3	HSPD1, HSP90AA1, SOD1
Aging	3	CALR, SOD1, CNP
Autophagy	3	VDAC1, GAPDH, HSP90AA1
• ~ × ·	Cellular co	mponent
		ALDOA, ANXA5, CALR, HSP90B1, HSPD1,
Extracellular vesicle	15	PRDX2, PGK1, SOD1, VIM, VDAC1, GAPDH,
		HSP90AA1, YWHAG, ATP5A1, CNP
		CNP, ATP5A1, ANXA5, CALR, GAPDH,
Plasma membrane	11	HSP90AA1, HSP90B1, HSPD1, SOD1, VIM,
		VDAC1
		ALDOA, CALR, HSP90B1, HSPD1, PRDX2,
Cytosol	11	PGK1, SOD1, VIM, GAPDH, HSP90AA1,
		YWHAG
Nuclous	10	CNP, ATP5A1, ALDOA, CALR, GAPDH,
Indeeds	10	HSP90AA1, HSP90B1, HSPD1, SOD1, VDAC1
Cytoplasmic vasicle	9	ALDOA, CALR, HSP90B1, HSPD1, SOD1,
Cytoplashic vesicle)	VDAC1, CNP, HSP90AA1, YWHAG
Extracellular matrix	8	CALR, HSP90B1, HSPD1, SOD1, VIM, ATP5A1,
	0	GAPDH, HSP90AA1
Protein complex	8	ATP5A1, CALR, GAPDH, HSP90AA1, HSP90B1,
	0	HSPD1, SOD1, VDAC1
Myelin sheath	7	HSPD1, VDAC1, HSP90AA1, YWHAG,
	,	ATP5A1, CNP, SOD1
Extracellular space	6	ALDOA, ANXA5, CALR, HSPD1, SOD1, CNP

Mitashandrian	(CNP, ATP5A1, HSPD1, SOD1, YWHAG,			
Mitochonarion	0	VDAC1			
Call impation	(ALDOA, ANXA5, CALR, HSP90B1, VIM,			
Cell junction	0	YWHAG			
Perinuclear region of cytoplasm	5	CNP, CALR, GAPDH, HSP90AA1, HSP90B1			
Peroxisome	3	HSPD1, SOD1, VIM			
Membrane microdomain	3	HSPD1, PGK1, VDAC1			
Melanosome	3	CNP, HSP90AA1, HSP90B1			
Molecular function					
DNA hinding	0	CNP, ATP5A1, ALDOA, CALR, HSP90AA1,			
KNA binding	9	HSP90B1, HSPD1, YWHAG, VIM			
En avera hinding	7	CALR, HSP90AA1, HSP90B1, HSPD1, SOD1,			
Enzyme binding		YWHAG, VDAC1			
Carbohudrata dariwatiya hinding	7	ATP5A1, CALR, HSP90AA1, HSP90B1, HSPD1,			
	1	PGK1, VIM			
Nucleotide hinding	7	CNP, ATP5A1, GAPDH, HSP90AA1, HSP90B1,			
Nucleofide binding	7	HSPD1, PGK1			
ATP binding	5	ATP5A1, HSP90AA1, HSP90B1, HSPD1, PGK1			
Recentor hinding	5	ATP5A1, CALR, HSP90AA1, HSP90B1,			
Receptor binding	5	YWHAG			
Unfolded protein binding	4	CALR, HSP90AA1, HSP90B1, HSPD1			
Protein complex binding	4	CALR, HSP90AA1, VIM, VDAC1			
Phosphatase binding	3	HSP90AA1, HSP90B1, SOD1			
Glycoprotein binding	3	CALR, HSP90AA1, VIM			
Chaperone binding	3	CALR, HSPD1, SOD1			
ATPase activity	3	ATP5A1, HSP90AA1, HSPD1			

Enriched terms connected to "Biological process", "Cellular component" and "Molecular function" are presented together with the number of annotated proteins. Proteins are listed in column three.

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