

## LIST OF ABBREVIATIONS FOR ALS/MND

Entries marked with an asterisk (\*) are known to cause familial ALS/MND when mutated. Mutated TAU can cause frontotemporal dementia (FTD), and approximately 40% of FTD patients show signs of motor neuron involvement, but it is still a moot point whether you can class mutated TAU as a cause of ALS/MND.

Every entry may not necessarily be fully related to the cause of ALS/MND, but the entries can be found in scientific research papers on ALS/MND.

If you amend or alter the stored copy of this list, make sure the researched relied upon is conducted in the pre-G1 phase of the cell cycle. Very little research is conducted in this condition.

AA	arachidonic acid	AMP	adenosine monophosphate. A compound containing adenine, ribose and one phosphate group
AADPR	O-acetyl-ADP ribose	AMPA	$\alpha$ -amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid receptor
ACETYL CoA	acetyl coenzyme A	AMPK	5'-AMP-activated protein kinase
ACh	acetylcholine	*ANG	angiogenin
AChR	acetylcholine receptor (nicotinic receptor)	APC	antigen-presenting cell
AD	Alzheimer's disease	ARF6	ADP-ribosylation factor 6
ADP	adenosine diphosphate	ATF	activating-transcription factor (4 or 6)
AGE	advanced glycation end-product	ATG	autophagy-related genes
AICAR	5-aminoimidazole-4 carboxamide ribonucleoside	ATM	ataxia telangiectasia mutated (ATM is a checkpoint kinase that helps activate the G1 check point, or restriction point-R)
AIF	apoptosis-inducing factor (in mitochondria)	ATP	adenosine triphosphate
aka	"also known as"	ATXN2	ataxin 2 (Interaction with TDP-43)
$\alpha$ KG	alpha ketoglutarate	BAD	Bcl-2 associated death protein (pro-apoptosis)
AKT1/2/3	aka PKB $\alpha$ / $\beta$ / $\gamma$ (the serine kinase S474 is activated in skeletal muscle during the G1 phase)	Bak	Bcl-2 antagonist/killer (pro-apoptosis)
ALDOC	aldolase	Bax	Bcl-2 associated X protein (pro-apoptosis)
ALS	amyotrophic lateral sclerosis/motor neurone disease	BBB	blood-brain barrier
ALS FTD	amyotrophic lateral sclerosis frontotemporal dementia	BCL-2	bcl-2 gene, aka B-cell lymphoma 2, (an anti-apoptosis regulator)
*ALS2	alsin (Predominant UMN phenotype)	BDNF	brain-derived neurotrophic factor
ALT	alternative lengthening of telomeres		

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BECN1	beclin 1	*CHCHD10	coiled-coil-helix-coiled-coil-helix domain containing 10
BH3	Bcl-2 homology domain 3	Chk1/Claspin	(DNA damage) checkpoint kinase 1
Bim	BIP binding immunoglobulin protein	*CHMP 2B	chromatin modifying protein 2B (Predominant LMN phenotype)
BIP	binding immunoglobulin protein	CHOP	CCAAT-enhancer-binding protein homologous protein
BMAA	beta-methalamino-1-alanine	CIP2A	cancerous inhibitor of PP2A (A protein encoded by the KIAA1524 gene)
BNB	blood-nerve barrier	CK	creatine kinase. (In brain, thyroid, skeletal and cardiac muscle)
Bnip3	Bcl2/adenovirus E1B 19 kDa protein-interacting protein 3	cMyc	cyclic myelocytomatosis oncogene
BRAF	baroreflex arc function	CNS	central nervous system
BULBAR	bulbar onset in neck area	COFILIN	An actin binding protein in skeletal muscle and a major actin depolymerisation factor in the CNS. Cofilin is inactivated after phosphorylation of serine residues.
C1-INH	complement C1 inhibitor	COX-1	cyclooxygenase-1. aka prostaglandin-endoperoxide synthase (PTGS). (Also, COX-2)
C1q	complement 1 subcomponent q	CPT-1&2	
C4BP	C4b-binding protein	CR1	complement receptor type 1
C5	complement C5	CREB	cyclic AMP-response element-binding protein
*C9orf72	chromosome 9 open reading frame 72 (Associates with LC3 and leads to ALS FTD)	CSF	cerebrospinal fluid
Ca2+	calcium	CSMN	corticospinal motor neuron
Calmodulin/Ca2+	intracellular calcium receptor	CYT C	cytochrome C
CALS	carer of ALS patient	DAF	decay-accelerating factor
cAMP	cyclic adenosine monophosphate	DAG	diacylglycerol
CBP	CREB binding protein. Assists acetylation	*DAO	d-amino acid oxidase
CCD	coiled-coil domain	DAPI	4,6-diamidino-2-phenylindole
*CCNF	G2-mitotic-specific cyclin-F	DC	dendritic cell
CCR2	CC chemokine receptor 2		
Cdc42	cell division cycle 42		
CDK4	cyclin-dependent kinase 4		
CDKN2A	cyclin-dependent kinase inhibitor 2A		

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\*DHFR (Associated with bulbar onset when mutated)

\*DCTN1 dynactin 1

DEPTOR an inhibitor of mTOR complex 2

DMF dimethyl fumarate (Activates Nrf2 and NQO1)

DNA deoxyribonucleic acid

dsDNA double-strand DNA

DSB double strand break

\*DYRK1A dual specificity tyrosine phosphorylation-regulated kinase 1A

EAAT2 excitatory amino acid transporter 2

eIF2 $\alpha$  eukaryotic initiation factor 2 $\alpha$

eIF4E eukaryotic initiation factor 4E

EMG electromyography

EMT epithelial-mesenchymal transition

ENO enolase

EPH A4 ephrin receptor A4 (Repels axons at NMJ)

ER endoplasmic reticulum

ERAD endoplasmic reticulum protein degradation

ERK extracellular-signal regulated kinase (aka MAPK)

ERV endogenous retrovirus

ESC embryonic stem cell

ETC electron transport chain

E2F1 E2F transcription factor 1, aka E2 promoter binding factor

FA fatty acid

FAD Flavin adenine dinucleotide

fALS familial ALS, (Usually caused before the age of 35 years; caused by a familial fault)

\*FIG4 SAC 1 lipid phosphatase domain containing (S cerevisiae)

FLD frontal lobe degeneration

FLCN folliculin

fMND familial MND (Same as fALS)

FOXO forkhead box O. There are 5 family members, Foxo1 (FKHR), Foxo2, Foxo3 FKHL1, Foxo4 (AFX), and Foxo6.

FOXP3 forkhead box P-3. aka scurfin

FTD frontotemporal dementia

FTLD frontotemporal lobar atrophy/disease

\*FUS fused in sarcoma (aka translocated in liposarcoma, or fused in osteosarcoma)

GABA gamma-aminobutyric acid. An inhibitory neurotransmitter. (Receptors A, A-p, and B)

GAPDH glyceraldehyde-3-phosphate dehydrogenase

G $\beta$ L co-activator of mTOR C2 (an ortholog of yeast LST8)

GEF guanine nucleotide exchange factor

GLU glucose

GLUT4 glucose transporter 4

GM-CSF granulocyte-macrophage colony-stimulating factor

GPCR G-protein coupled receptor

GPI glycosylphosphatidylinositol, (or glucose-6-phosphate isomerase)

GRK G-protein coupled receptor kinase

GSK3 glycogen synthase kinase 3 ( $\alpha$ ,  $\beta$ ,  $\gamma$ )

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GTP	guanosine triphosphate	ILK	beta 1-integrin-linked protein kinase
GWAS	genome wide association study	iNOS	inducible nitric oxide synthase
H3K9	histone H3K9	ILF3	interleukin factor 3
HCCS	human copper chaperone for SOD1	IP3	inositol 1,4,5-trisphosphate
HDAC	histone deacetylase	IR	insulin receptor
HDM2	human DM2, aka MDM2 for mouse	IRE-1	inositol-requiring kinase 1
HERV K	human endogenous retrovirus K	IRF 1-9	interferon regulatory factor 1 to 9
HIF-1 $\alpha$	hypoxia inducible transcription factor 1 $\alpha$	IRS 1,2	insulin receptor substrate (1 or 2)
HK	Hexokinase	ISREs	interferon-stimulated response elements
*hnRNPA1	heterogeneous ribonucleoprotein A1	iTreg	inducible regulatory T cells
*hnRNPA2B1	heterogeneous ribonucleoprotein A2B1	JAK	janus kinase. In ALS; only JAK1 (and TYK2) activated in muscles. JAK1 and JAK2 are activated in neurons, but not at the same time as TYK2 or G1 phase muscle regeneration. This is an important reason that a chronic muscular condition can cause the death of connected neurons in ALS.
hnRNP H	heterogeneous ribonucleoprotein H	JNK	c-Jun N-terminal kinase (aka SAPK)
HSP	heat-shock protein, with different kDa versions, (e.g. 60, 70, 90)	KD	kinase domain
IF	intermediate filaments	KLD	kinase-ligase Dpnl
IFN- $\alpha$ , $\beta$ or $\gamma$	interferon $\alpha/\beta/\gamma$ , a cytokine. There is also a Tau and Omega and other minor versions. (INF- $\alpha$ has 13 subtypes, IFN- $\beta$ has 2 subtypes, INF- $\gamma$ has 1 subtype). INF- $\alpha$ and $\beta$ are referred to as Type 1 Interferons. IFN- $\gamma$ is referred to as Type 2 interferon. In ALS; INF- $\beta$ will be activated.	LC3-1 or 2	microtubule-associated 1 light chain 3 $\beta$ (1 or 2), (associates with C9ORF72)
IGF-1	insulin-like growth factor-1	LDH/LD	lactate dehydrogenase
IGF-1R	insulin-like growth factor-1 receptor	LIR	LC3 interacting motive
IGF-2	insulin-like growth factor-2	LKB1	lyman-kutcher-burman 1 (activates 13 kinases of the AMPK family)
IGF-2R	insulin-like growth factor-2 receptor	LMN	lower motor neuron
IGIF	interferon gamma inducing factor. aka IL-18	LMND	lower motor neuron disease
I $\kappa$ B- $\alpha$	I kappa B-alpha	LPS	lipopolysaccharide
IKK	I kappa B kinase	MAC	membrane attack complex
IL-1 $\beta$	interleukin 1 beta (also IL-6, or IL-10)	MAPK	mitogen-activated protein kinase (aka ERK)

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MASP	MBL-associated serine protease	NAD <sup>+</sup>	nicotinamide adenine dinucleotide, (PARP consumes NAD <sup>+</sup> as a substrate). See NMN
*MATR3	matrin 3	NADH	the reduced form of NAD, (aka niacin or coenzyme 1)
MBL	mannose-binding lectin	NADP <sup>+</sup>	NAD phosphate
MCL-1	induced myeloid leukaemia cell differentiation protein 1, (enhances cell survival by inhibiting apoptosis)	NADPH	reduced nicotinamide adenine dinucleotide phosphate
MCP	membrane cofactor protein	NAM	nicotinamide; a form of vitamin B3. Helps increase sirtuin 1 in brain tissue, and decrease NFκB
MCT 1	monocarboxylate transporter 1	NAMPT	nicotinamide phosphoribosyltransferase, required to produce NAD <sup>+</sup>
MEK	MAPK/ERK kinase	NAP1	NAK-associated protein 1
MG	myasthenia gravis	NDP52	nuclear domain 10 protein 52
MHC	major histocompatibility complex. In ALS; activated MHC I in muscles. MHC II is inhibited via its receptor.	NEMO	NF-κB essential modulator
MIF	macrophage migration inhibitory factor	NF	neurofilament, or nuclear factor
MIP	macrophage inflammatory protein	NFκB	nuclear factor kappa-light-chain-enhancer of activated B cells. NFκB has two isoforms, p50 and p65.
mMCT	mitochondrial monocarboxylate transport inhibitor	NK	natural killer
MMP9	matrix metalloproteinase 9	NLS	nuclear localization signal
MND	motor neuron disease	NMJ	neuromuscular junction
MNLS	muscle-neuronal lactate shuttle	NMN	nicotinamide mononucleotide. A precursor of NAD <sup>+</sup>
MO25	calcium-binding protein 39, (aka CAB 39, CGI-66)	NO	nitric oxide
MPS	mononuclear phagocytic system	NOS	nitric oxide synthase
mRNA	messenger RNA	NOXA	a mediator of p53-induced apoptosis
mTOR	mammalian transducer of regulated CREB activity, (mammalian target of rapamycin). There are 2 complexes of TOR, a C1 and C2 complex. See "TORC1, TORC2" listing.	NQO1	NAD(P)H: Quinone oxidoreductase 1, (can be induced by dimethyl fumarate)
MW	molecular weight	Nrf2	nuclear factor erythroid 2-related factor 2, (aka NF kappa B-repressing factor 2). In C. elegans the equivalent of Nrf2 is called SKN-1.
MYC	myelocytomatosis oncogene		

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NT	non-transfected	PEP	phosphoenolpyruvate (a PPAR agonist)
nTreg	natural regulatory T cell	PEPck	phosphoenolpyruvate carboxykinase
*OPTN	optineurin (associates with TBK-1 and autophagy)	PERK	PKR-like endoplasmic reticulum kinase
OxPHOS	oxidative phosphorylation	PFK	phosphofructokinase
P16 <sup>INK4A</sup>	melanoma gene	*PFN1	profilin 1 (Mutations disrupt polymerization of monomeric G-actin to its filamentous F-actin form)
P21	a 21 kDa inhibitor of cyclin-dependent kinase	PGC-1 $\alpha$ /PGC-1 $\beta$	PPAR-gamma coactivator-1 alpha/beta
P27	a 27 kDa inhibitor of cyclin-dependent kinase	PGE2	prostaglandin E2
P53	protein 53	PGK	phosphoglycerate kinase
P70S6K1	PI(3,4,5)P3-dependent kinase (p70 or S6K1)	PGM	phosphoglycerate mutase
p300/HAT	histone acetyl transferase	PHGDH	phosphoglycerate dehydrogenase
PALS	patients with ALS	Pi	inorganic phosphate
PARP 1	poly (ADP-ribose) polymerase 1. (PARP 1 helps repair damaged single-stranded DNA and helps activate the G1 phase). PARP consumes NAD <sup>+</sup> as a substrate.	PI3K	phosphatidylinositol 3 kinase
PAS	phagophore assembly site	PK	pyruvate kinase
PAT1	paroxysmal atrial tachycardia	PKA	protein kinase A
PCD	programmed cell death	PKB $\alpha$ / $\beta$ / $\gamma$	protein kinase B $\alpha$ / $\beta$ / $\gamma$ . (aka Akt). The serine kinase PKB $\beta$ is activated in skeletal muscle during G1 phase
PCR	polymerase chain reaction	PKC	protein kinase C. (aka phosphokinase C)
PDH	pyruvate dehydrogenase	PKD	protein kinase D
*PDIA1/3	protein disulphide isomerase (1/3)	PKH	pyruvate kinase (heart)
PDK	pyruvate dehydrogenase kinase (1 to 4). Mutated PDK3 causes Charcot-Marie-Tooth neuropathy.	PKM	pyruvate kinase (muscle form)
PDP-1	pyruvate dehydrogenase phosphatase (muscle)	PKM1/2	are 2 different splicing products of PKM
PE	phosphatidylethanolamine	PLA2	phospholipase A2
PEA	palmitoylethanolamide (a PPAR agonist)	PLC	phospholipase C ( $\beta$ , $\delta$ , $\epsilon$ , or $\gamma$ )
		PLD	phospholipase D (1 or 2)
		PLS	primary lateral sclerosis

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PMA	progressive muscular atrophy	RIPK 1	receptor interacting protein kinase 1
PMA	phorbol myristate acetate (A phorbol ester that assists in inhibiting the vitamin D receptor)	RNA	ribonucleic acid
PMN	polymorphonuclear cell	RNAPII or RNPII	RNA polymerase II
PNS	peripheral nerve system	ROS	reactive oxygen species
PP2A	protein phosphatase-2A	S6K1	ribosomal protein S6 kinase 1
PP2A/Ca <sup>2+</sup>	protein phosphatase-2A or calcium	S6P	phosphorylation of ribosomal s6
PPAR- $\alpha$	peroxisome proliferator activated receptor- $\alpha$	sALS	sporadic Amyotrophic Lateral Sclerosis
PPAR- $\beta$	peroxisome proliferator activated receptor- $\beta$	SAPK	stress-activated protein kinase (aka JNK)
PRAS40	proline-rich Akt substrate of 40 kDa	SC	Schwann cell (or spinal cord)
pRb	retinoblastoma protein	SD	semantic dementia
PTEN (aka MMAC1)	phosphatase and tensin homologue deleted on chromosome 10 (aka MMAC1)	*SETX	senataxin
PTX	pertussis toxin	shRNA	small hairpin RNA
PUMA	p53 upregulated modulator of apoptosis	*SIGMA1	sigma nonopioid intracellular receptor 1
RA	retinoic acid	SIK2	salt-inducible kinase 2
RAF	repetitive atrial firing	SIN1/MIP1	stress activated protein kinase interacting protein 1. SIN1 assists with activating mTORC2.
RAG2	recombination activating gene 2	siRNA	small interfering RNA (aka. Silencing RNA)
Raptor	activator of mTOR Complex 1	SIRT	sirtuin (1 to 6)
Rb	retinoblastoma	sMND	sporadic Motor Neuron Disease
REDD-1	regulated in development and DNA damage response 1	SOCS	suppressor of cytokine signalling. In ALS; SOCS1 is activated and SOCS3 will be inhibited.
Rheb1-GTP	Rheb aka Ras homolog enriched in brain, is a GTP-binding protein involved in the mTOR C1 pathway	*SOD1	superoxide dismutase 1 (there is already 170 different SOD1 mutations reported in fALS with different virulence)
Rictor	activator of mTOR Complex 2	SOL	soleus
RIP1	receptor interacting protein 1	*SPG11	spatacsin 11
		*SQSTM1	sequestosome 1
		Src	non-receptor tyrosine kinase

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ssDNA	single-stranded DNA	Treg	regulatory T cell
STAT	signal transducers and activators of transcription. In ALS; stats 1 and 2 will be activated, and stat3 inhibited.	TSC1	tuberous sclerosis complex 1 (aka hamartin)
STRAD	straightened	TSC 2	tuberous sclerosis complex 2 (aka tuberin)
TA	tibialis anterior	*TUBA4A	tubulin $\alpha$ 1
*TANK	TRAF family member-associated NF $\kappa$ B activator	TYK2	tyrosine kinase 2
TARDBP	transactivation response element (TAR) DNA-binding protein of 43 kDa (The gene encoding TDP-43)	*UBQLN2	ubiquilin2
TAU	a protein	UCP-2/3	uncoupling protein (2 or 3)
*TBK-1	TANK binding kinase 1	ULD	ubiquitin-like domain
TBST	tris-buffered saline tween	ULK	Unc 51-like autophagy activating kinase
TCA	tricarboxylic acid cycle	UMND	upper motor neuron disease
TCF-3	transcription factor 3	UNC13A	unc-13 homolog A. (Regulates neurite outgrowth and synaptic neurotransmission)
TCR	T cell receptor	UPR	unfolded protein response
*TDP-43	transactivation response element (TAR) DNA-binding protein of 43 kDa	*VAPB	vesicle-associated membrane associated-protein B. (Mutations disrupt endocytosis and transport of vesicles from the golgi and ER)
TERT	telomerase reverse transcriptase	VB1	vitamin B1 (thiamine). Benfotiamine is a lipid-soluble analogue (synthetic) VB1
TGF	transforming growth factor	VCAM-1	vascular cell adhesion molecule
TK	tyrosine kinase	*VCP	valosin-containing protein
TKTL1	transketolase-like enzyme 1	VDR	vitamin D receptor, (the receptor is inhibited in ALS/MND)
TLR	toll like receptor	VEGF	vascular epithelial growth factor
TNF	tumour necrosis factor	WASP	Wiskot-Aldrich syndrome protein
TORC1, TORC2	TOR complex 1 or 2. Complex 1 (G $\beta$ L, mTOR, Raptor) is inhibited, and Complex 2 (G $\beta$ L, mTOR, Rictor) is activated in ALS/MND skeletal muscle. Also see "mTOR" listing.	WT	wild-type
TPI	triose phosphate isomerase	XBP1	x-box binding protein 1
		ZO-1	zonular occludens 1, (aka tight junction protein-1)
		4E-BP1	eukaryotic initiation factor 4E-binding protein 1



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14-3-3 binding motive