

ABBREVIATIONS

5' UTR	5'-Untranslated region	Mito-miRs	Mitochondrial miRNAs
AGO2	Argonaute2	MRI	magnetic resonance imaging
AIF	Apoptosis inducing factor	mRNA	messenger RNA
ALS	Amyotrophic lateral sclerosis	mSCs	Mitochondrial super complexes
ATM	ataxia telangiectasia mutated	mtDNA	mitochondrial DNA
ATP	Adenosine triphosphate	mtDNA	mitochondrial DNA
BLAST	Basic local alignment search tool	NADH	nicotinamide adenine dinucleotide + hydrogen (H)
BN-PAGE	Blue native polyacrylamide gel Electrophoresis	ncDNA	Nuclear DNA
CNS	Central nervous system	ncRNA	non-coding RNA
CSF	Cerebrospinal fluid	NDUFB7	NADH dehydrogenase 1 beta subcomplex subunit 7
CUGBP1	CUG triplet repeat, RNA binding protein 1	NDUFS2	NADH dehydrogenase [ubiquinone] iron-sulfur protein 2
DAVID -	Database for annotation, visualization and Integrated Discovery	NEMP	Nuclear encoded mitochondrial Protein
ddPCR	Droplet digital PCR	NES	Nuclear export signal
DGCR8	DiGeorge syndrome critical region 8	NGS	Next generation sequencing
DMPK	Dystrophia myotonica protein kinase	NLS	Nuclear localization sequence
EARS2	Glutamyl-TRNA Synthetase 2	OMIM	Online Mendelian Inheritance in Man
ER	Endoplasmic reticulum	OXPHOS	Oxidative phosphorylation
ETC	Electron transport chain	P bodies	Processing bodies
FASTKD5	FAST kinase domains 5	PAGE	Polyacrylamide gel electrophoresis
FLAIR	Fluid-attenuated inversion recovery	PARP	Poly-ADP ribose polymerase
FM	Full mutation	PBMCs	Peripheral blood mononuclear cells
FMR1	Fragile X mental retardation 1	PBS	Phosphate buffer saline
FMRP	Fragile associated mental retardation Protein	PI	propidium iodide
FXAND	Fragile X-associated Neuropsychiatric Disorders	PINK	PTEN-induced putative kinase 1
FXPOI	Fragile X associated primary ovarian insufficiency	PM	Premutation
FXS	Fragile X syndrome	pre-miRNA	precursor miRNA
FXTAS	Fragile X-associated tremor/ataxia syndrome	Pri-miRNA	Primary miRNA
GAPDH	Glyceraldehyde 3-phosphate Dehydrogenase	qPCR	quantitative polymerase chain reaction
GO	Gene ontology	RISC	RNA induced silencing complex
GPX1	Glutathione peroxidase-1	RNA-IP	RNA immunoprecipitation
hnRNP	Heterogeneous nuclear ribonucleoproteins	RNP	ribonucleoprotein
IGA	In Gel Activity	ROS	reactive oxygen species
IMS	Intermembrane space	rRNA	ribosomal RNA
iPSCs	Induced pluripotent stem cells	SDH	Succinate dehydrogenase
LAP2β	Lamina-associated polypeptide 2 beta	SDS	Sodium dodecyl sulfate
LDH	Lactate dehydrogenase	snRNA	Small nuclear RNA
MBNL1	Muscleblind Like Splicing Regulator 1	SOD	Superoxide dismutase
MDRS	mitochondrial disulfide relay system	TMRM	Tetramethylrhodamine, methyl ester
miRNA	microRNA	tRNA	transfer RNA
MIM	Mitochondrial inner membrane	WCL	Whole cell lysate