S1 | **A list of characterized genes encoding pentatricopeptide repeat-containing proteins. The genes are arranged according to the kingdom to which the organism belongs and then listed alphabetically.**

PPR protein-encoding gene	Organism	Localization (Predicted, Confirmed)/ target RNA (Predicted, *in vivo* association, *in vitro* confirmation)	Possible function or involvement	Loss of function phenotype	Reference	Notes
Plants						
CRP1	Maize	Chloroplast stroma (C) /*psaC* 5' UTR (*in vivo*), *petA* 5' UTR (*in vivo*), *petD* (P)	Translation of *petA, psaC* mRNA, processing of *petD* mRNA	Reduced cytochrome *f* (*petA*) and photosystem I subunit (*psaC*) protein, unprocessed polycistronic *petD* mRNA, loss of cytochrome b_6f complex, decrease of photosystem I activity	1-3	Associated with multisubunit protein complexes
CRR2 (*At3g46790*)	*Arabidopsis*	Chlorplast (P) /*rps7-ndhB* pre-mRNA (P)	Controls *ndhB* expression by regulating intragenic processing of *rps7-ndhB* transcript	Decreased NdhB protein levels, reduced chloroplast NA(P)DH dehydrogenase complex activity	4	
CRR4 (*At2g45350*)	*Arabidopsis*	Chloroplast (C) /*ndhH* (*in vitro*)	Binds *ndhD* mRNA to recruit RNA editing machinery	Reduced NdhH protein, reduced chloroplast NA(P)DH dehydrogenase complex activity	5,6	
EMP4	Maize	Mitochondria (C) /unknown	Mitochondria development, seed development	Lethal loss of endosperm development in seeds, reduced levels of mitochondrial *rps2A*/*rp2B*, *rps3*/*rpl13*, *mttb* RNA's	7	
Grp23 (*At1g10270*)	*Arabidopsis*	Nucleus (C) /RNA polymerase II, subunit III (*in vivo*)	Transcriptionally regulates early embryo development through interactions with RNA polymerase II	Loss of proper embryonic development	8	
GUN1 (*At2g31400*)	*Arabidopsis*	Chloroplast (C) /unknown	Chloroplast development, Retrograde signaling	*gun* mutant, does not repress photosynthesis-related nuclear genes after chloroplast photo-oxidative damage or translation inhibition	9	
HCF152 (*At3g09650*)	*Arabidopsis*	Chloroplast stroma (C) /*petB* pre-mRNA(*in vitro*)	Controls processing of *psbB-psbT-psbH-petB-petD* polycicstronic RNA	Impaired *petB* intron splicing/stabilization, impaired endonucleolytic cleavage of *psbH-petB* mRNA transcript, reduced cytochrome b_6f complex activity	10-12	
LOI1 (*At4g14850*)	*Arabidopsis*	Mitochondria (P) /unknown	Post-transcriptionally regulates 3-hydroxy-3-methylglutaryl coenzyme A reductase and isoprenoid biosynthesis	Insensitive to isoprenoid biosynthesis inhibitors lovastatin and clomazone	13	Binds single stranded nucleic acids *in vitro*
MCA1	*Chlamydomonas reinhardtii*, (eukaryotic algae)	Chloroplast (C) /*petA* (P)	Regulates *petA* mRNA transcript stability	Reduced levels of *petA* mRNA transcript, reduced levels of cytochrome f (*petA*) protein, reduced activity of cytochrome b_6f complex	14,15	MCA1 is a short-lived protein, abundance fluctuates rapidly under different growth conditions
OTP3 (*At1g74910*)	*Arabidopsis*	Mitochondria (P) /*nad1* (*in vivo*)	Required for *trans*-splicing of intron 1 of *nad1* mRNA	Reduced levels of Complex I activity. Defects in seed development and germination	16	
PGR3 (*At4g31850*)	*Arabidopsis*	Chloroplast (P) /*petL* (P), *ndhD* (P)	Regulates stabliziation/translation of *petL* mRNA and translation of *ndhD* mRNA	Reduced *petL*, *petG* mRNA, reduced NdhD protein, reduced level of cytochrome b_6f and NA(P)DH dehydrogenase complex activities	17	

PPR2	Maize	Chloroplast stroma (C)/ unknown	Required for the synthesis or assembly of chloroplast translation machinery	Chloroplasts lack ribosomes, albino seedlings	18	
PPR4	Maize	Chloroplast stroma (C) /1st intron of *rps12* pre-mRNA (*in vivo*)	Regulates *rps12 trans*-splicing and chloroplast ribosome biogenesis	Defective in *rsp12 trans*-splicing, albino seedlings lacking chloroplast ribosomes	19	
PPR531-11	*Physcomitrella patens* (moss)	Chloroplast (P) /*clpP* (P)	Regulates intergenic cleavage between *clpP* and 5'-*rps12* mRNA and splicing of *clpP* mRNA	Reduced levels of photosystem II reaction center protein D1 (*clpP*), abnormal chloroplast morphology	20	
PTAC2 (*At1g74850*)	*Arabidopsis*	Chloroplast (C) /unknown	Required for normal chloroplast encoded RNA polymerase-dependent transcription	Reduction of chloroplast transcription, albino seedlings	21	Associated with chloroplast transcriptionally active complexes *in vivo*
Tbc2	*Chlamydomonas reinhardtii*, (eukaryotic algae)	Chloroplast stroma (C) /*psbC* 5' UTR (P)	Regulates translation of *psbC* mRNA as part of a protein complex	Reduced levels of chlorophyll-binding photosystem II reaction center subunit P6 protein (*psbC*), reduced photosystem II activity	22,23	
Plant CMS restorers						
Rf1a, PPR791, PPR8-1	Rice	Mitochondria (P) /B-*atp6*/*orf79* (*urf-rmc*) (P)	Dominant restorer of BT-type CMS, reduces levels of aberrant CMS-associated protein, promotes endonucleolytic cleavage of *urf-rmc* RNA, role in *atp6* mRNA editing	Male sterility, accumulation of aberrant *urf-rmc* protein product	24-27	
Rf1b	Rice	Mitochondria (P) /B-*atp6*/*orf79* (*urf-rmc*) (P)	Dominant restorer of BT-type CMS, reduces levels of aberrant CMS-associated protein, destabilizes *urf-rmc* mRNA	Male sterility, accumulation of aberrant *urf-rmc* protein product	24,25	
Rfk1, *ORF687*	Radish	Mitochondria (P) /*orf125* (P)	Dominant Kosena-CMS restorer of fertility, decreases accumulation of CMS-associated protein ORF125 but does not decrease transcript levels	Male sterility, accumulation of ORF125 protein product	28	
Rfo	Radish	Mitochondria (P) /*orf138* (P)	Dominant Ogura (*ogu*)-CMS restorer of fertility, decreases accumulation of CMS-associated protein ORF138 but does not decrease transcript levels	Male sterility, accumulation of ORF138 protein product	29,30	
Rf-PPR592	Petunia	Mitochondria (C) /*pcf* (*in vivo*)	Dominant CMS restorer of fertility, decreases accumulation of CMS-associated protein PCF	Male sterility (CMS), accumulation of CMS associated protein PCF	31,32	
Animals						
BSF	*Drosophila melanogaster*	Cytoplasm (C) /*IV*/*V* RNA (3' UTR of bicoid (*bcd*) mRNA) (*in vitro*)	Regulates stability of *IV*/*V* RNA during oogenesis	Reduced levels of IV/V RNA	33	

LRPPRC, LRP130	Human	Mitochondria (C), nucleus (C) /*coxI*, *coxIII* (P)	Regulates the stability/ translation of *coxI*, *coxIII* mRNA's	Leigh syndrome French Canadian, cytochrome *c* oxidase deficiency	34-37	Associated with nuclear and mitochondrial RNA *in vivo*, has nucleotide-binding activity *in vitro*
Protists						
TbPPR1-TbPPR8	*Trypanosoma brucei* (protist causes African Sleeping Sickness)	TBPPR1-7: Mitochondria (C) TBbPPR8: Cytosol (C) /TbPPR5: 12S rRNA (*in vivo*)	TbPPR2-7 involved in mitochondrial ribosome assembly/function	Defects in oxidative phosphorylation and growth in glucose medium, *TbPPR2-7* mutants have reduced mitochondrial rRNA levels	38	
Fungi						
Cya-5	*Neurospora crassa*	Mitochondria (P) /*coxI* (P)	Required for the stabilization/translation *cox1* mRNA	Reduced cytochrome *c* oxidase activity	39	
PET309	*Saccharomyces cerevisiae*	Mitochondrial inner membrane (C) /*coxI* 5' UTR(P)	Required for the normal processing/ stabilization of *coxI* pre mRNA and its translation	Reduced cytochrome *c* oxidase activity, reduced levels of mature *coxI* mRNA and COXI protein	40,41	

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