

Huntington Disease

Species		Tissue	PGC-1α Expression	Protein/Transcript		Downstream Genes	Expression	Citation	PMID
Human	Presymptomatic, postmortem caudate		Decreased	Transcript	Not measured		N/A	[112]	17018277
Human	Presymptomatic, postmortem cerebellum, hippocampus		No Change	Transcript	Not measured		N/A	[112]	17018277
Human	Presymptomatic, postmortem caudate (PMID16467349)		Decreased	Transcript		ACADM, COX7C, CYCS, LDHB, NDUFS3, NDUFB5	Decreased	[113]	17055784
Human	Brain lysates		Decreased; clinical grade dependent	Protein	TFAM		Decreased	[94]	20660112
Human	Symptomatic, putamen		FL-PGC-1α inconsistent	Protein	Not measured		N/A	[118]	21757867
			NT-PGC-1α decr. grade 2, incr. grade 3&4	Protein	Not measured		N/A	[118]	21757867
Human	Myoblasts		FL-PGC-1α inconsistent	Protein	Not measured		N/A	[118]	21757867
Human	Myoblasts		NT-PGC-1α decreased	Protein	NRF1, TFAM		Decreased	[118]	21757867
Human	Symptomatic, muscle biopsy		Decreased	Transcript	NRF1, TFAM		No change	[115]	19460884
Mouse	STHdhQ111 (HD KI: PMID11092756)		Decreased	Protein/Transcript	Coxiv, Cycs		Decreased	[112]	17018277
Mouse	STHdhQ111		NT-PGC-1α decreased	Protein	Not measured		N/A	[118]	21757867
Mouse	STHdhQ111		Decreased	Transcript	Cycs		Decreased	[115]	20665636
Mouse	STHdhQ111		Decreased	Transcript	Not measured		N/A	[128]	30941017
Mouse	STHdhQ111		Decreased	Transcript	Not measured		N/A	[114]	22589249
Mouse	STHdhQ111		FL-PGC-1α inconsistent	Protein	Not measured		N/A	[118]	21757867
Mouse	R6/2; brain		FL-PGC-1α, NT-PGC-1α decreased	Transcript	Tfam		Decreased	[314]	22095692
Mouse	R6/2; muscle		FL-PGC-1α, NT-PGC-1α decreased	Transcript	Tfam		Decreased	[314]	22095692
Mouse	R6/2; brown adipose tissue		FL-PGC-1α, NT-PGC-1α decreased	Transcript	Tfam		Decreased	[314]	22095692
Mouse	R6/2, N171-82Q striatum (younger)		NT-PGC-1α decreased	Protein	Not measured		N/A	[118]	21757867
Mouse	R6/2, N171-82Q striatum (symptomatic)		NT-PGC-1α increased	Protein	Not measured		N/A	[118]	21757867
Mouse	R6/2, N171-82Q striatum		FL-PGC-1α inconsistent	Protein	Not measured		N/A	[118]	21757867
Mouse	R6/2; striatum		No Change	Transcript	Pvalb		Decreased; p>0.05	[45]	22916173
Mouse	LCM-captured medium spiny neurons (HD KI: PMID12926013)		Decreased	Transcript	Not measured		N/A	[112]	17018277
Mouse	LCM-captured nNOS interneurons (HD KI: PMID12926013)		Increased	Transcript	Not measured		N/A	[112]	17018277
Mouse	D9-N171-98Q striatum		Increased	Transcript	Pvalb		Increased	[105]	21177255
Mouse	N171-82Q striatum		Decreased	Transcript	Cox6a, Cycs, Ldhb, Ndufs3, Tfam		Decreased	[113]	17055784
Mouse	NLS-N171-82Q; soleus muscle		Decreased	Protein/Transcript	Tfam		Decreased	[115]	19460884
Mouse	NLS-N171-82Q; extensor digitorum longus		Decreased	Protein/Transcript	Ppargc1b, Tfam		Decreased	[115]	19460884
Mouse	NLS-N171-82Q;gastrocnemius		No Change	Protein/Transcript	Tfam		Decreased	[115]	19460884
Mouse	NLS-N171-82Q; striatum		Decreased	Protein/Transcript	Cox2, Creb, Esrra, Nr1f1, Nr1f2, Ppard, Tfam		Decreased	[116]	20529956
Mouse	NLS-N171-82Q; cortex		No Change	Protein/Transcript	Coxiv, Esrra, Ppard		Decreased	[116]	20529956
Mouse	NLS-N171-82Q; liver		Decreased	Transcript	Hnf4a, Tfam		Decreased	[116]	20529956
Mouse	NLS-N171-82Q; brown adipose tissue		Decreased;p >0.05	Transcript	Ucp1		Decreased; p>0.05	[116]	20529956
Mouse	HD knock-in series (PMID26908599); striatum		No Change	Transcript	Ak1, Idh3a, Nceh1, Phyh, Wdr77		Increased	[61]	29491012
Mouse	HD knock-in series (PMID26908599); cortex		No Change	Transcript	Syt2, Nefh, Nceh1, etc.		No change	[62]	32222555
Mouse	HD knock-in series (PMID26908599); hippocampus		No Change	Transcript	Syt2, Nefh, Nceh1, etc		No change	[62]	32222555
Mouse	PVcre-mhtt		No Change	Transcript	Pvalb		No change	[69]	24121117
Rat	Primary oligodendrocytes with lenti-mutant Htt exon1 (72Q)		Decreased	Transcript	Hmgcs1, Hmgcr, Mbp		Decreased	[116]	21715619
Rat	Primary neurons; full-length Htt (wtHtt with 15Q or mtHtt with 138Q)		No Change	Transcript	Not measured		N/A	[117]	19915593

Parkinson Disease

Species		Tissue	PGC-1α Expression	Protein/Transcript		Downstream Genes	Direction	Citation	PMID
Human	iPSC human derived		Decreased	Transcript	Not measured		N/A	[187]	24290359

Human	Substantia nigra from human post mortem PD patients Braak stage 5 or 6	Decreased	Transcript	Not measured	N/A	[188]	25363075
Human	Human H4 neuroglioma cells	Decreased	Transcript	Not measured	N/A	[188]	25363075
Human	Human derived iPSC dopaminergic neurons, LRRK2 mutation carriers	Decreased	Protein	Not measured	N/A	[197]	29129681
Human	Human substantia nigra samples from PD patients	Decreased	Protein/Transcript	Soluble Parkin decreased, Insoluble Parkin increased, NRF-1 decreased, PARIS increased	Decreased	[177]	21376232
Human	Human substantia nigra samples from PD patients	Decreased	Protein/Transcript	Insoluble Parkin, PARIS	Increased	[177]	21376232
Human	PARKIN deficient human dopaminergic neurons	Decreased	Protein/Transcript	PARIS/ZNF746	Increased	[181]	32795422
Human	PARKIN deficient human dopaminergic neurons	Decreased	Protein/Transcript	<i>PARK2</i>	Decreased	[181]	32795422
Human	Laser captured dopaminergic neurons, PD patients with Lewy pathology	No change	Protein/Transcript	Nuclear encoded mitochondrial genes	Increased/Decreased	[182]	20926834
Human	Human substantia nigra samples from PD patients	Decreased	Protein/Transcript	Not measured	N/A	[205]	30236862
Human	Human substantia nigra samples from PD patients	Decreased	Protein	<i>SDHA, TOMM20</i>	Decreased	[74]	27622213
Human	Neural progenitor Striatum cells	Increased	Transcript	<i>CAT1, CYCS, SOD1, SOD2, GPX1, UCP2</i>	Increased	[52]	17055439
Human	SHSY5's MPTP treated with Necdin lentiviral expression	Increased	Protein	Not measured	N/A	[207]	26971449
Mouse	A30P (Thy-1 alpha-syn mice) Brainstem and Cortex	Decreased	Transcript	Not measured	N/A	[188]	25363075
Mouse	Conditional Paris knockout mice injected with LentiGFP-Cre	Decreased	Protein/Transcript	<i>Paris/Zfp746</i>	Increased	[177]	21376232
Mouse	Conditional Paris knockout mice injected with LentiGFP-Cre	Decreased	Protein/Transcript	<i>Nrf1, Park2</i>	Decreased	[177]	21376232
Mouse	Conditional Pgc-1alpha knockout injected with AAV-CRE-GFP	Decreased	Protein/Transcript	<i>Sdha, Tomm20</i>	Decreased	[74]	27622213
Mouse	PGC-1alpha null mice	Decreased	Transcript	<i>Cytc, Ucp2, Ant1, Sod2, Gpx1, Cat1</i>	Decreased	[52]	17055439
Mouse	PGC-1alpha null mice	Decreased	Transcript	<i>Ucp3 , Sod1</i>	No change	[52]	17055439
Mouse	10T1/2 cells treated with hydrogen peroxide	Increased	Transcript	<i>Cytc, Ucp2, Ucp3, Ant1, Sod1, Sod2, Gpx1, Cat1</i>	No change	[52]	17055439
Mouse	PGC-1alpha null mice injected with MPTP	Decreased	Protein/Transcript	Not measured	N/A	[52]	17055439
Mouse	MPTP treated non-transgenic mice	Decreased	Protein/Transcript	<i>Esrra, Sirt3</i>	Decreased	[206]	26421366
Mouse	MPTP treated non-transgenic mice	Decreased	Protein/Transcript	<i>Atp5b, Sod2</i>	Increased	[206]	26421366
Mouse	MPTP treated non-transgenic mice with Sirt3 deletion in dopaminergic neurons	Decreased	Protein	<i>Esrra, Sirt3</i>	Decreased	[206]	26421366
Mouse	MPTP treated non-transgenic mice with Sirt3 deletion in dopaminergic neurons	Decreased	Protein	<i>Atp5b, Sod2</i>	Increased	[206]	26421366
Mouse	MPTP injected striatum, cortex,and cerebellum 90 minutes after injection	Increased	Transcript	Not measured	N/A	[209]	28161458
Mouse	MPTP injected striatum, cortex,and cerebellum 7 days after injection	No change	Transcript	Not measured	N/A	[209]	28161458
Mouse	MPTP injected striatum, cortex,and cerebellum 12 days after injection	No change	Transcript	Not measured	N/A	[209]	28161458
Mouse	MPTP-induced models of 6h-PSS, 24h-PSS, AdvPSS andESS of PD	Decreased	Transcript	<i>Kif1b, Mybb1a, Nrf1, Paris/Zfp746, Park2, Mybb1a</i>	Decreased	[210]	32280590
Drosophila	Drosophila S2 cell line or brain tissue with paris mutation	Decreased	Protein/Transcript	<i>Park2, Pink1</i>	Decreased	[179]	32138754
Rat	6-hydroxydopamine lesioned rats treated with ferulic acid	Increased	Protein/Transcript	<i>Drp1, Mfn2</i>	Increased	[219]	31657074

Neurodevelopmental Disorders

Species	Tissue	Direction	Protein/Transcript	Downstream Genes	Direction	Citation	PMID
Human	Human genomic DNA from major depressive disorder and controls	Increased	Transcript	Not measured	N/A	[236]	30381832
Human	Postmortem cingulate cortex tissue from patients with schizophrenia	No change	Transcript	<i>Syt2, Cplx1, Nefh, Parv, Nrf1</i>	Decreased	[248]	26683626
Mouse	Ppp1r2-Cre/fGluN1 knockout mice	Decreased	Protein	<i>Cat1, Gpx1, Sod1, Sod2</i>	Decreased	[53]	23348010

Supplemental Table S1. Summary of studies mentioned in the main text which reported the expression of PGC-1α transcript or protein in disease states.