

Appendix Table of Contents

• Appendix Table S1; QPCR primer sequences	p1
• Appendix Table S2; P values	p2
• Appendix Figure S1	p4
• Appendix Figure S2	p5
• Appendix Figure S3	p6
• Appendix Figure S4	p7
• Appendix Figure S5	p8
• Appendix Figure S6	p9

QPCR Primers

Species	Target	Forward	Reverse
Human	<i>18s</i>	AGTCCCTGCCCTTTGTACACA	GATCCGAGGGCCTCACTAAAC
Human	<i>COASY</i>	GCTGAAGATACTCACGGACATT	CAATCACACACACACGCTTTC
Human	<i>Mt-ATP6</i>	ThermoFisher Hs02596862_g1	
Human	<i>Mt-CO1</i>	ThermoFisher Hs02596864_g1	
Human	<i>Mt-CYB</i>	ThermoFisher Hs02596867_s1	
Human	<i>Mt-ND1</i>	ThermoFisher Hs02596873_s1	
Human	<i>PPCDC</i>	CAACTGAGAGAGCCAAACATTTTC	GGCTCTTCCATATCTCCCATTCT
Human	<i>PPCS</i>	CCTGTCTCTGAAATGCCTGAA	TGGGAGCCCAATCTTTAACC
Human	<i>TFRC</i>	GACAATGCTGCTTTCCCTTTC	TCCATGGTGGTACCCAAATAAG
Mouse	<i>Calb1</i>	AAGGCTGGATTGGAGCTATC	GACGTGAGCCAACTCTACAA
Mouse	<i>Coasy</i>	GGAGGCCTTTGGAACAGATATT	GAGGATCTTCATCTGCTTCTTGT
Mouse	<i>Cpt1c</i>	GTTTCCTCTGGAGGTGGATT	GCTGGAGATATGGAAGGTGATT
Mouse	<i>Drd1</i>	TCCCAGATCGGGCATTG	CTCTTCCTGGTCAATCTCAGTC
Mouse	<i>Drd2</i>	GCCATTGTTCTTGGTGTGTTT	TAGAGGACTGGTGGGATGTT
Mouse	<i>Gabra3</i>	ATCCGGTCTAGTACAGGAGAAT	GATACATGGCAAGTAGGTCTGG
Mouse	<i>Gabra6</i>	GAGTCAGTCCCAGCAAGAAC	GCATAGGACACTTTGGGTAGAG
Mouse	<i>Gapdh</i>	TGCACCACCAACTGCTTAGC	GGCATGGACTGTGGTCATGAG
Mouse	<i>Hepc</i>	TTGCGATACCAATGCAGAAG	TGCAACAGATACCACACTGG
Mouse	<i>Ireb2</i>	GACAAGCACTGGAAAAGAGTTC	ACGAGTCATGAAAGATACCACG
Mouse	<i>Pank1α</i>	GTTCGCCCAGCATGATTCTC	CTTAACCAGGGTTCCACCGAT
Mouse	<i>Pank1β</i>	CTGAGCCTAACTCCATTCAACT	TCCACCGATATCCATACCAAAC
Mouse	<i>Pan-Pank1</i>	GGACATTTACGGAGGAGACTATG	CTTTGCTGATGGACTCTCTCTT
Mouse	<i>Pank3</i>	TCACTGGGAACCAAAGGATAAA	CAGTGAGGGATACTTCCCATTATAG
Mouse	<i>Ppcdc</i>	TAACAACAGAGAGAGCCAAACA	GCTTCCACATCTCCCATTCA
Mouse	<i>Ppcs</i>	CTCTCAGTCCATTAGGCTCTTC	GGATCTTGTGTTCAAGGCATTTC
Mouse	<i>Tfric</i>	GAGTATCACTTCCTGTCGCCCTATG	GCTGAGAGAGTGTGAGAGCCAGAGC
Mouse	<i>Th</i>	CCCTACCAAGATCAAACCTACC	CTGGATACGAGAGGCATAGTTC

Appendix Table S1 QPCR primers are listed for each target gene

Exact P Values and Types of ANOVA Analyses

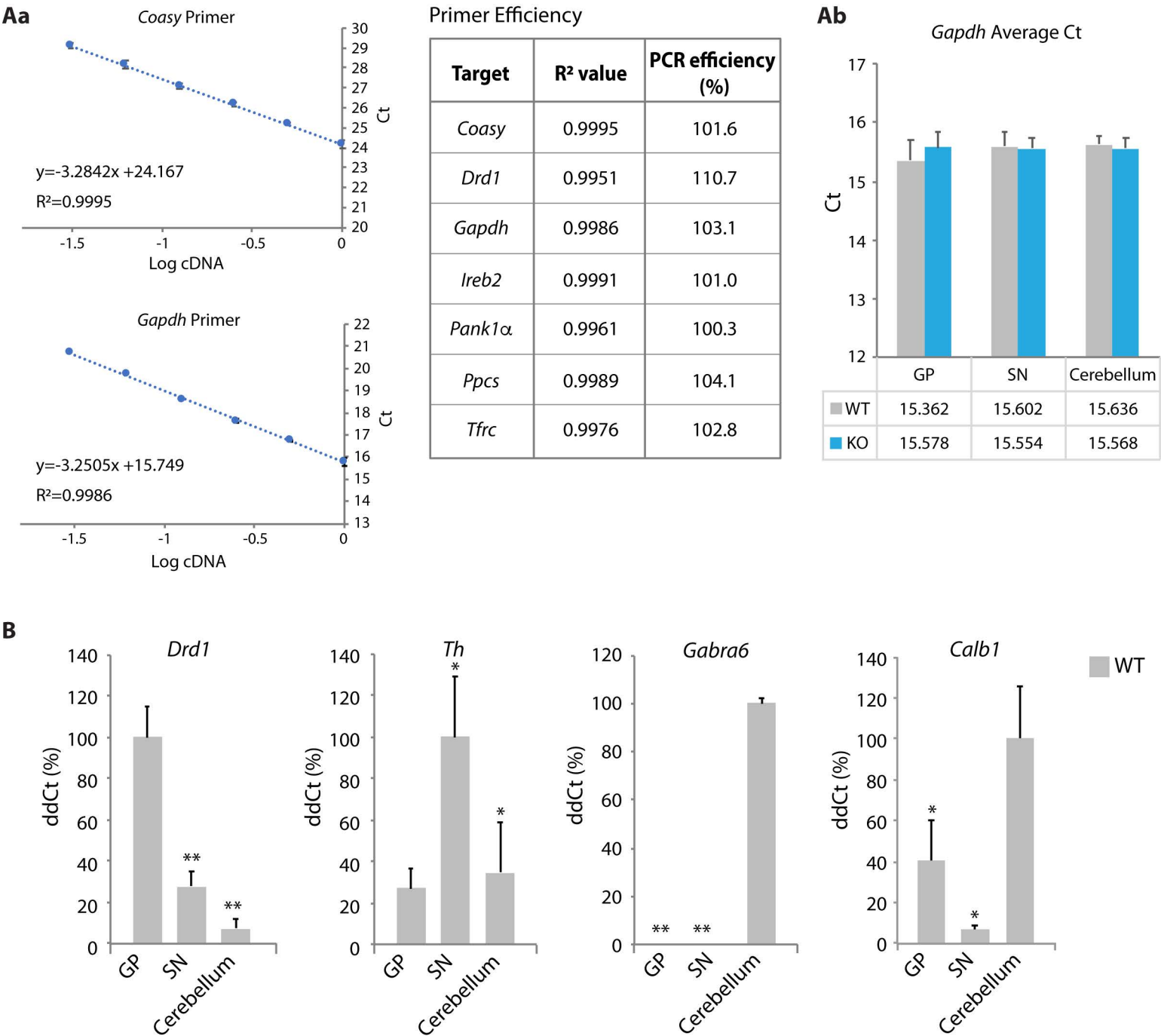
Figure		P				ANOVA
Fig 1C		<i>Ppcs</i>	<i>Ppcdc</i>	<i>Coasy</i>		
	GP	3.2000E-03	8.4463E-01	9.3703E-07		1w
	SN	1.5323E-01	1.5681E-01	5.0979E-02		1w
	Cerebellum	2.6651E-01	3.5626E-01	8.6756E-01		1w
Fig 1D	Within vehicle	7.6512E-03				2w
	Within KO	9.9097E-05				
Fig 2A		<i>Tfrc</i>	<i>Ireb2</i>	<i>Hamp</i>		
	GP	2.2904E-05	2.7871E-02	6.8690E-03		1w
	SN	5.8679E-01	3.7452E-01	1.4969E-01		1w
	Cerebellum	4.1119E-02	2.9212E-01	5.1926E-02		1w
Fig 2B	Cytosol	3.4071E-02				1w
	Mitochondria	3.5259E-02				1w
Fig 2C		Mitochondrial	Cytosol			
	Brain	1.4553E-04	3.8716E-01			1w
	Liver	4.6571E-01	5.0120E-01			1w
Fig 2D	Within vehicle	2.8158E-03				2w
	Within KO	2.1842E-03				
Fig 2E	Within vehicle	1.8000E-02				2w
	Within KO	2.0000E-02				
Fig 2F		<i>Tfr1</i>	<i>PDHE1a1</i>			
	Within vehicle	3.1626E-06	1.3863E-01			2w
	Within KO	7.8443E-04	5.1421E-02			
Fig 3A		<i>Drd1</i>				
	GP	5.3339E-06				1w
	SN	4.3633E-01				1w
	Cerebellum	7.3619E-01				1w
		<i>Drd1</i>				
	Within vehicle	1.3818E-04				2w
	Within KO	1.2694E-05				
Fig 3B-C		<i>Drd2</i>	<i>Gabra3</i>			
	GP	2.7109E-02	9.2253E-01			1w
	SN	9.9616E-01	5.6443E-01			1w
	Cerebellum	9.3350E-01	2.1192E-01			1w
Fig 3D	WT vs. KO	1.4058E-02				1w
Fig 4A		COASY				
	Fibroblasts	8.6730E-05				1w
	Lymphoblasts	4.5552E-03				1w
Fig 4B		COASY/Coasy				
	Human	2.8232E-08				1w
	Mouse	4.7685E-03				1w
Fig 4C	OCR	1.7873E-03				1w
	ECAR	3.0106E-03				1w

Exact P Values and Types of ANOVA Analyses

Figure		P				ANOVA
Fig 4D						
	Within vehicle	1.3744E-02				2w
	Within KO	1.5759E-02				
Fig 4E						
	<i>MT-ND1</i>	4.8595E-02				1w
	<i>MT-CYB</i>	4.8953E-02				1w
	<i>MT-CO1</i>	4.7521E-02				1w
	<i>MT-ATP6</i>	4.9782E-02				1w
Fig 4F	Ctl vs. PKAN	3.6231E-02				1w
Fig 5A		<i>Pank1a</i>				
	GP	5.0287E-03				1w
	SN	8.8661E-01				1w
	Cerebellum	5.3904E-01				1w
	<i>Pank1a</i>	5.0287E-03				1w
	<i>Pank1b</i>	6.9087E-01				1w
	Total	1.7742E-02				1w
Fig 5B		<i>Pank3</i>				
	GP	7.1749E-01				1w
	SN	1.8733E-01				1w
	Cerebellum	2.7469E-01				1w
Fig 5C		<i>Pank1a</i>	<i>Coasy</i>			
	WT-veh vs. KO-veh	2.8964E-03	9.5749E-05			1w
	WT-veh vs. KO-Beza	8.7788E-04	2.1066E-03			1w
	WT-veh vs. KO-Gem	2.0877E-02	1.3012E-03			1w
Fig 6A						
	WT-veh vs. KO-veh	2.7497E-05				1w
	KO-veh vs. KO-pPanSH	1.6837E-03				1w
Fig 6B		<i>Coasy</i>	<i>Tfrc</i>	<i>Ireb2</i>	<i>Drd1</i>	
	WT-veh vs. KO-veh	2.7497E-05	2.2904E-05	3.1837E-02	1.4354E-04	1w
	WT-veh vs. WT-20	1.3324E-03				1w
	WT-veh vs. KO-20	2.0024E-03				1w
	WT-veh vs. KO-8.2				6.1119E-03	1w
Fig 6C						
	CTL-0 vs. PKAN-0	4.3592E-02				1w
	CTL-0 vs. PKAN-400	3.6906E-02				1w
Fig 6D						
	within vehicle	1.1068E-05				2w
	within PKAN	1.1001E-03				
Fig 7A		<i>Coasy</i>	<i>Tfrc</i>	<i>Drd1</i>		
	WT-veh-D0 vs. KO-veh-D0	2.1540E-07	1.4228E-06	2.0254E-04		1w
	WT-veh-D0 vs. KO-pPanSH-D7	2.4302E-03	8.9396E-03	1.5690E-02		1w

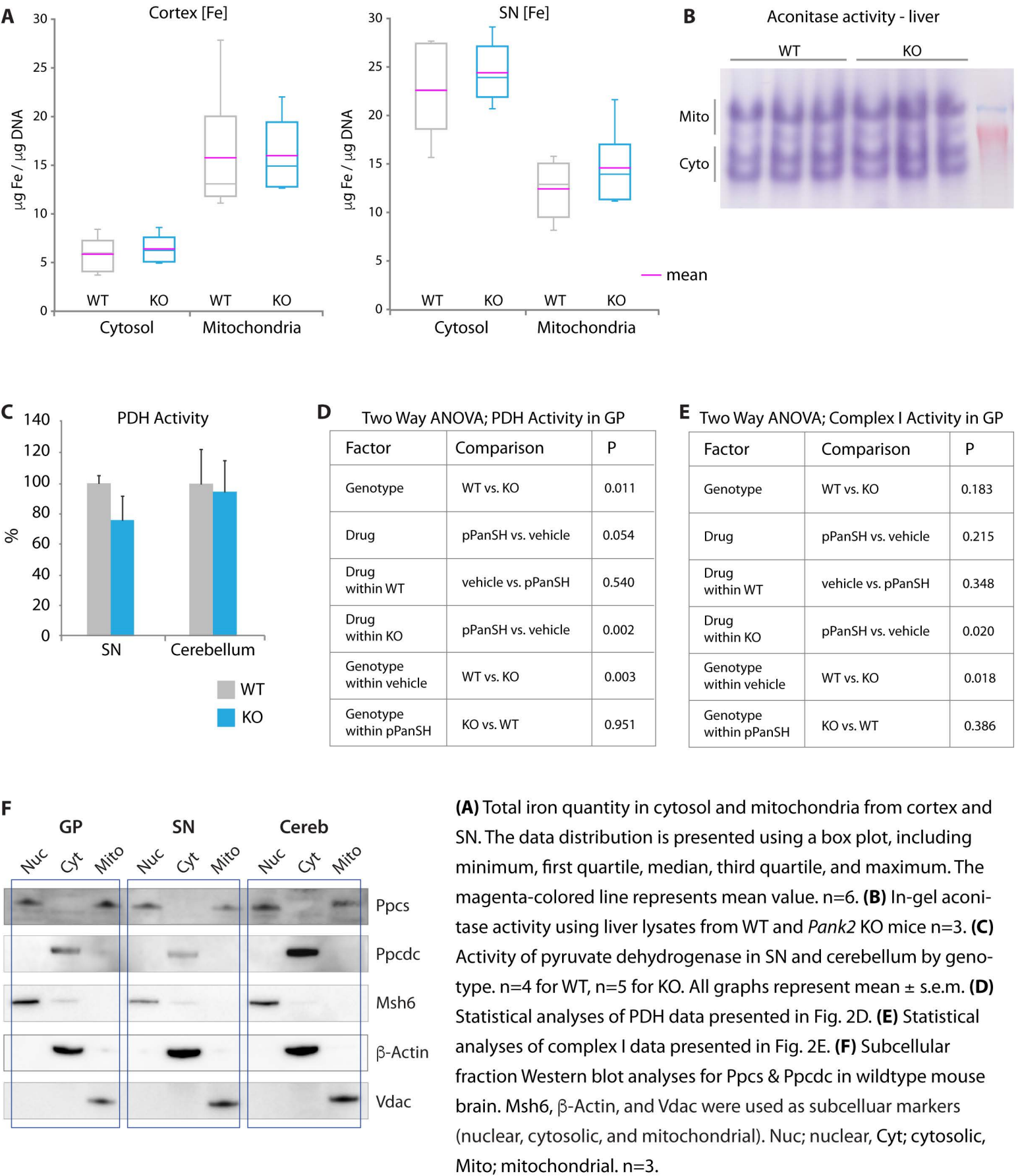
1w; One-way ANOVA, 2w; Two-way ANOVA analysis

Appendix Figure S1

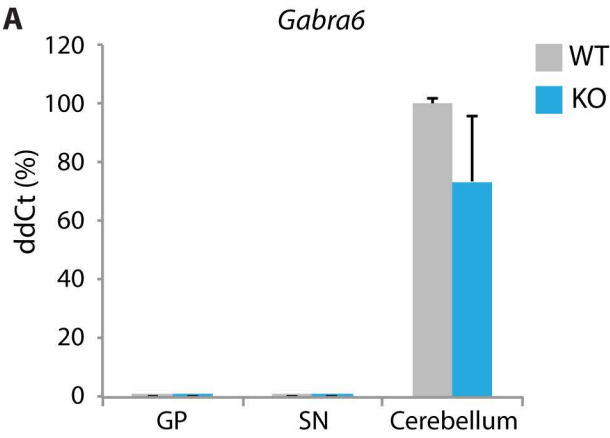


(Aa) Efficiency of primers per target gene showed all primers fit between 90~110% range. QPCR reaction was performed using serially diluted duplicated samples and PCR efficiency was calculated using $\text{Efficiency (\%)} = \{-1 + 10^{(-1/\text{slope})}\} \times 100$. **(Ab)** Average *Gapdh* Ct showed no difference between genotype nor brain area. **(B)** Genes that are regionally expressed based on public data were selected and quantified by region. Data were evaluated by one-way ANOVA. * $P < 0.05$; ** $P < 0.01$. $n = 3$. All graphs represent mean $\pm$ s.e.m.

Appendix Figure S2

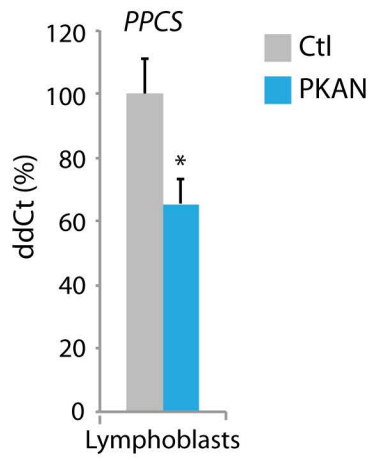


Appendix Figure S3



(A) Relative expression of a GABA receptor gene, *Gabra6* in GP, SN, and cerebellum. n=11,4,4 for WT and n=8,4,4 for KO (GP, SN, Cerebellum, respectively). All graphs represent mean $\pm$ s.e.m.

A



(A) Relative quantification of *PPCS* mRNA from cultured human lymphoblasts by genotype. n=5 for both genotypes. Data were evaluated by one-way ANOVA.

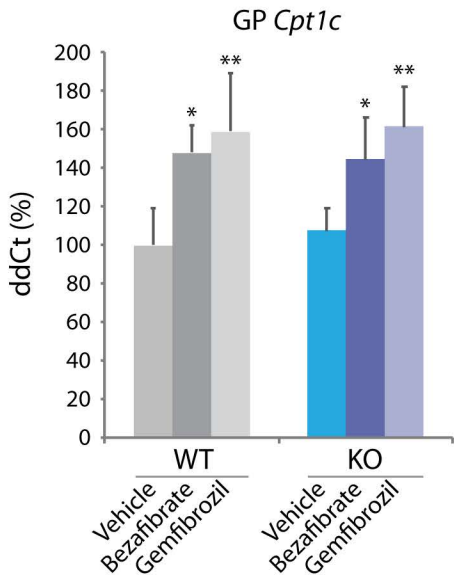
*P<0.05. All graphs represent mean $\pm$ s.e.m.

A

One Way ANOVA; <i>Pank1α</i>	
Comparison	P
WT veh vs. KO veh	0.003
WT veh vs. KO Beza	0.0009
WT veh vs. KO Gem	0.021
KO veh vs. KO Beza	0.346
KO veh vs. KO Gem	0.662
KO Gem vs. KO Beza	0.320

One Way ANOVA; <i>Coasy</i>	
Comparison	P
WT veh vs. KO veh	<0.0001
WT veh vs. KO Beza	0.002
WT veh vs. KO Gem	0.001
KO veh vs. KO Beza	0.730
KO veh vs. KO Gem	0.607
KO Gem vs. KO Beza	0.684

B



Two Way ANOVA; <i>Cpt1c</i>		
Factor	Comparison	P
Genotype within vehicle	WT vs. KO	0.522
Drug within WT	vehicle vs. bezafibrate	0.001
Drug within WT	vehicle vs. gemfibrozil	<0.001
Drug within WT	beza vs. gem	0.695
Drug within KO	vehicle vs. bezafibrate	0.021
Drug within KO	vehicle vs. gemfibrozil	<0.001
Drug within KO	beza vs. gem	0.38

(A) Tables of two-way ANOVA results are presented for *Pank1α* and *Coasy* are shown for data presented in Figure 5C. **(B)** Relative quantification of *Cpt1c* mRNA in GP by genotype and treatment status. n=8,8,4 for both genotypes (GP, SN, Cerebellum, respectively). A table of two-way ANOVA results for *Cpt1c* is shown. *P<0.05; **P<0.01. All graphs represent mean ± s.e.m.

Appendix Figure S6

