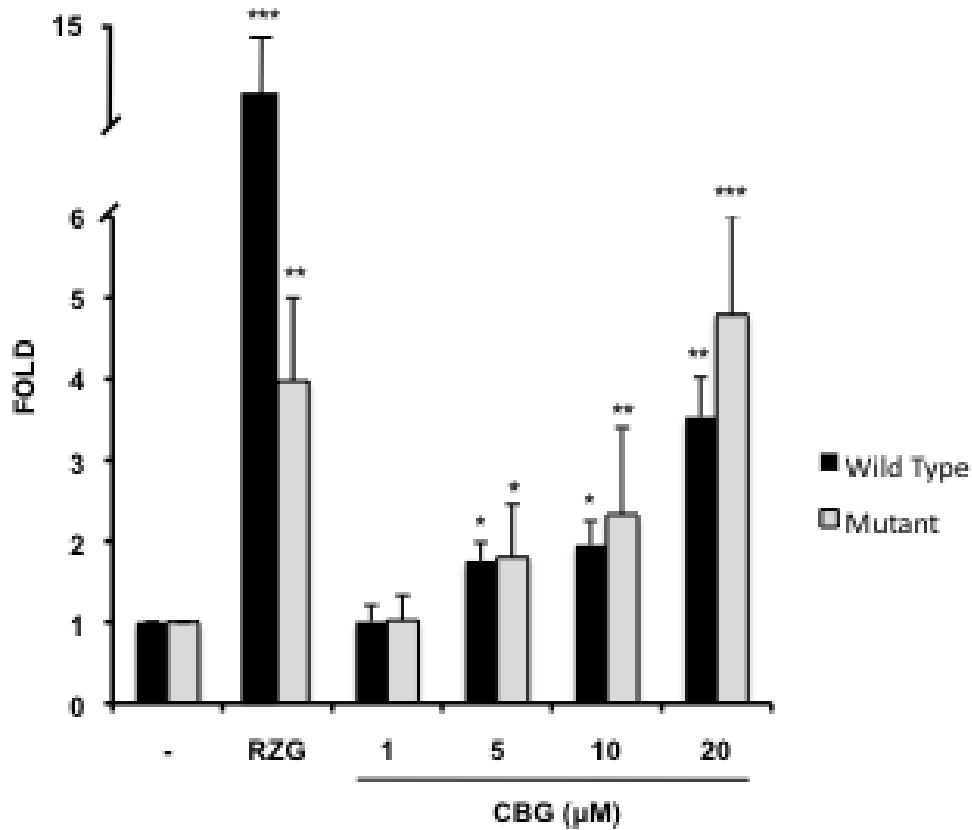


Supplementary Figure 1



Effect of CBG on PPAR γ transcriptional activity. STHdh^{Q7/Q7} (wild-type) and STHdh^{Q111/Q111} (mutant) cells were co-transfected with GAL4-PPAR γ and GAL4-luc. Cells were treated with of CBG at the indicated concentrations for 12 h and luciferase activity measured in the cell lysates. Fold activation level was calculated, taking the control sample as reference. Data are expressed as mean \pm S.D. of at least three independent experiments. * $p < 0.05$, ** $p < 0.01$ and *** $p < 0.005$ in an unpaired two-tailed Student's *t* test.

Table 1S. HD array analysis (84 genes) showing their up- or down-regulatory responses in the striatum of control and 3NP-lesioned mice treated with CBG or vehicle. Values correspond to number of folds that a specific gene is up- (positive value) or down-regulated (negative value).

3NP vs control

3NP+CBG vs control

Gene	Fold Up- or Down-Regulation	Gene	Fold Up- or Down-Regulation
Akt1	-1,0396	Kcnc3	1,0098
Apoe	-1,4804	Lpl	1,0454
Aqp1	1,0898	Map3k10	-1,0989
Arfp2	1,2008	Ncor1	1,1204
Atp2b2	-1,2025	Nefl	-1,5326
Bax	-1,0253	Ngef	-1,0468
Bbox1	-1,0762	Ntrk2	1,2431
Bdnf	1,5845	Pacsin1	-1,0324
C3	1,1761	Pgk1	1,0454
Calb1	-1,1376	Plcb4	1,0238
Casp3	1,1925	Plod2	-1,0837
Casp8	1,3890	Ppargc1a	1,3794
Cd44	3,0189	Ppp3ca	-1,0253
Cltc	1,1519	Prkcb	1,1050
Cnr1	-1,0042	Prpf40a	1,2518
Creb1	-1,1615	Ptpn11	1,4083
Crebbp	1,1680	Rab6	1,0673
Dctn1	1,1127	Rcor1	1,0168
Dlg4	-1,2449	Rest	1,5845
Eef1a2	-1,0112	Rgs4	-1,2449
Egfr	-1,2277	Rilp	1,9106
Elmo1	1,2781	Rph3a	-1,3717
Fgf12	1,2260	Rxrb	-1,0042
Gabrd	-1,1696	Sgk1	2,7207
Gja1	-1,6088	Sin3a	1,0028
Gjb6	1,2260	Slc14a1	1,8583
Gnaq	1,0526	Slc25a4	1,0822
Gpx1	1,2092	Snap25	1,0168
Grb2	1,1599	Sod1	1,1127
Grin2a	1,3794	Sox2	-1,1142
Grin2b	1,2431	Sp1	-1,1535
Grm5	-1,0614	Sympk	-1,0989
Hap1	1,0098	Syn1	1,0098
Hdac1	1,2176	Tac1	1,1439
Hdac2	-1,1142	Tbp	1,1761
Hip1	1,1599	Tgm2	2,0907
Homer1	-1,1455	Tollip	1,3324
Hpca	1,1127	Trp53	1,1599
Htt	1,1050	Tubb5	-1,1297
Ift57	1,0526	Zbtb16	1,6178
Igf1	1,3416	Actb	1,1439
Itpr1	-1,0541	B2m	1,0310
Kcnab1	-1,1142	Gapdh	-1,1065
Kcnab2	1,0238	Gusb	-1,0253
		Hsp90ab1	-1,0396

Gene	Fold Up- or Down-Regulation	Gene	Fold Up- or Down-Regulation
Akt1	-1,1810	Kcnc3	1,0943
Apoe	-1,1892	Lpl	-1,0140
Aqp1	1,1096	Map3k10	1,0210
Arfp2	-1,0570	Ncor1	1,0718
Atp2b2	-1,0792	Nefl	-1,2142
Bax	-1,0425	Ngef	-1,1096
Bbox1	-1,1647	Ntrk2	1,0570
Bdnf	1,0644	Pacsin1	-1,0425
C3	1,0792	Pgk1	-1,1408
Calb1	-1,2226	Plcb4	1,1408
Casp3	-1,3195	Plod2	-1,1647
Casp8	1,3566	Ppargc1a	1,1728
Cd44	-1,3104	Ppp3ca	1,0353
Cltc	-1,1096	Prkcb	1,0644
Cnr1	-1,1173	Prpf40a	1,0281
Creb1	1,1567	Ptpn11	1,0792
Crebbp	1,0570	Rab6	1,1647
Dctn1	1,1487	Rcor1	-1,0281
Dlg4	-1,1408	Rest	1,0210
Eef1a2	1,0070	Rgs4	-1,1647
Egfr	-1,1647	Rilp	-1,0570
Elmo1	1,1810	Rph3a	-1,0792
Fgf12	1,0353	Rxrb	1,1567
Gabrd	-1,0570	Sgk1	-1,4142
Gja1	-1,0353	Sin3a	1,1019
Gjb6	1,1567	Slc14a1	1,2311
Gnaq	-1,0353	Slc25a4	-1,1096
Gpx1	1,2746	Snap25	-1,0140
Grb2	1,0353	Sod1	1,0210
Grin2a	1,1892	Sox2	-1,1019
Grin2b	1,2058	Sp1	-1,0867
Grm5	1,0070	Sympk	1,0425
Hap1	-1,0497	Syn1	1,0570
Hdac1	1,0353	Tac1	1,3472
Hdac2	-1,1567	Tbp	1,1975
Hip1	1,1408	Tgm2	1,5911
Homer1	1,0281	Tollip	1,0718
Hpca	1,0210	Trp53	1,3851
Htt	-1,0000	Tubb5	-1,0718
Ift57	1,1329	Zbtb16	1,0943
Igf1	1,1251	Actb	-1,0644
Itpr1	1,1892	B2m	1,1728
Kcnab1	-1,1647	Gapdh	-1,2483
Kcnab2	1,0353	Gusb	1,0867
		Hsp90ab1	1,0425

Table 2S. HD array analysis (84 genes) showing their up- or down-regulatory responses in the striatum of wild-type and R6/2 mice treated with CBG or vehicle. Values correspond to number of folds that a specific gene is up- (positive value) or down-regulated (negative value).

R6/2 vs wild-type

R6/2+CBG vs wild-type

Gene	Fold Up- or Down-Regulation	Gene	Fold Up- or Down-Regulation
Akt1	1,0733	Kcnc3	-1,1313
ApoE	-2,3424	Lpl	-1,4722
Aqp1	-1,4025	Map3k10	1,0807
Arfp2	-1,0629	Ncor1	1,3967
Atp2b2	-2,5633	Nefl	1,4459
Bax	1,1503	Ngef	-1,8251
Bbox1	2,5883	Ntrk2	1,0958
Bdnf	-1,2209	Pacsin1	-1,6795
C3	-2,6537	Pgk1	1,1423
Calb1	1,4969	Plcb4	-1,5562
Casp3	1,3491	Plod2	1,7435
Casp8	1,3398	Ppargc1a	1,0014
Cd44	-3,3589	Ppp3ca	-3,0063
Cltc	1,0084	Prkcb	-1,1631
Cnr1	-3,9945	Prpf40a	1,3491
Creb1	1,1345	Ptpn11	1,1663
Crebbp	1,6609	Rab6	1,3122
Dctn1	-1,8125	Rcor1	2,8521
Dlg4	-2,4083	Rest	-1,4221
Eef1a2	-1,8895	Rgs4	-3,2898
Egfr	1,3122	Rilp	-1,2816
Elmo1	-1,0852	Rph3a	-1,5347
Fgf12	-1,1157	Rxb1	-3,8053
Gabrd	-7,0031	Sgk1	-1,5889
Gja1	-3,3823	Sin3a	-2,3751
Gjb6	-1,2041	Slc14a1	-1,3268
Gnaq	1,6044	Slc25a4	1,5497
Gpx1	1,5178	Snap25	-1,0777
Grb2	1,8050	Sod1	1,9480
Grin2a	-1,1958	Sox2	-1,1080
Grin2b	-2,1555	Sp1	1,6609
Grm5	1,0367	Sympk	-5,4945
Hap1	2,2847	Syn1	1,0882
Hdac1	1,7195	Tac1	1,2159
Hdac2	2,2532	Tbp	1,5284
Hip1	-1,0338	Tgm2	1,6609
Homer1	-1,9026	Tollip	1,4459
Hpca	-3,0908	Trp53	1,2159
Htt	-1,6223	Tubb5	-1,1004
Ift57	1,0733	Zbtb16	-1,4722
Igf1	-1,8506	Actb	-1,9697
Itpr1	-2,7856	B2m	1,5497
Kcnab1	-1,5670	Gapdh	1,0367
Kcnab2	1,1503	Gusb	1,1583
		Hsp90ab1	1,0585

Gene	Fold Up- or Down-Regulation	Gene	Fold Up- or Down-Regulation
Akt1	-1,0483	Kcnc3	-1,1313
ApoE	-2,5105	Lpl	-2,9856
Aqp1	-1,5999	Map3k10	-1,0852
Arfp2	1,0439	Ncor1	1,0154
Atp2b2	-3,0063	Nefl	1,2414
Bax	-1,0556	Ngef	-2,1856
Bbox1	-1,2995	Ntrk2	-1,1958
Bdnf	1,2414	Pacsin1	-1,4025
C3	-3,7529	Pgk1	1,0295
Calb1	-1,5562	Plcb4	-1,3268
Casp3	1,2675	Plod2	-1,1712
Casp8	-1,4722	Ppargc1a	-1,2041
Cd44	-1,2728	Ppp3ca	-2,5456
Cltc	-1,1080	Prkcb	-2,0820
Cnr1	-4,0784	Prpf40a	1,2243
Creb1	-1,1392	Ptpn11	-1,1712
Crebbp	1,0585	Rab6	1,3305
Dctn1	-1,7630	Rcor1	1,2329
Dlg4	-2,4419	Rest	-2,0392
Eef1a2	-1,6223	Rgs4	-3,2898
Egfr	-1,2294	Rilp	-1,4928
Elmo1	-1,8000	Rph3a	-1,4520
Fgf12	-1,1157	Rxb1	-1,8506
Gabrd	-4,3409	Sgk1	-1,4722
Gja1	-3,3357	Sin3a	-1,8125
Gjb6	-1,1794	Slc14a1	-1,7876
Gnaq	1,1266	Slc25a4	1,3775
Gpx1	1,0439	Snap25	1,0733
Grb2	1,2414	Sod1	1,1034
Grin2a	-1,3454	Sox2	-1,2816
Grin2b	-1,8895	Sp1	1,0439
Grm5	-1,8251	Sympk	-3,3823
Hap1	-1,0556	Syn1	1,0882
Hdac1	1,2763	Tac1	-1,3177
Hdac2	1,2852	Tbp	1,1423
Hip1	-1,8378	Tgm2	1,2763
Homer1	-2,2315	Tollip	1,1663
Hpca	-4,1068	Trp53	1,1423
Htt	-1,9159	Tubb5	-1,0703
Ift57	-1,1471	Zbtb16	1,2588
Igf1	-1,2294	Actb	-1,5889
Itpr1	-2,1705	B2m	1,2329
Kcnab1	-2,5991	Gapdh	1,1111
Kcnab2	-1,1004	Gusb	1,1991
		Hsp90ab1	-1,0338