Supplemental Tables

Supplemental Table S1 Combined ranks created from Illumina and Affymetrix genome-wide expression analysis on YAC128 and control mice at 24 months of age.

Rank	Gene name	Gene symbol	Illumina probe	Affymetrix probe	Illu rank	Affy rank	Combined rank
1	Wilms tumor 1 homolog	Wt1	scl022431.11_253-S	1425995_s_at	1	1	1
2	netrin G1	Ntng1	scl0080883.1_122-S	1449286_at	2	27	14
3	transducer of ERBB2, 2	Tob2	scl0057259.2_60-S	1448667_x_at	24	55	40
4	protocadherin 20	Pcdh20	scl00219257.2_313-S	1424701_at	62	20	41
5	Scm-like with four mbt domains 2	Sfmbt2	scl21295.26.802_230-S	1434353_at	12	155	84
6	pogo transposable element with KRAB domain	Pogk	ri|C230043G09|PX00174N19|AK048752|3011-S	1459896_at	104	144	124
7	aspartoacylase (aminoacylase) 3	Acy3	scl52796.6.1_4-S	1448539_a_at	71	203	137
8	ArfGAP with FG repeats 2	Hrbl (Agfg2)	scl25869.12_138-S	1441347_at	245	32	137
9	polymerase (RNA) II (DNA directed) polypeptide A	Polr2a	scl40020.28_128-S	1426242_at	204	83	138
10	von Willebrand factor C domain-containing protein 2-like	Vwc21	scl00320460.2_251-S	1440413_at	182	109	146
11	DNA-damage-inducible transcript 4-like	Ddit4l	scl22560.5_561-S	1439332_at	291	2	146
12	OTU domain containing 7B	Otud7b	scl0229603.11_13-S	1429139_at	196	150	156
13	CD82 antigen	Cd82	scl012521.1_26-S	1416401_at	89	305	173
14	ATP-binding cassette, sub-family A (ABC1), member 6	Abca6	scl0001569.1_1-S	1453817_at	309	167	174
15	protein kinase C, theta	Prkcq	scl0003216.1_934-S	1426044_a_at	410	68	197
16	RAB GTPase activating protein 1-like	Rabgap1l	scl0029809.2_171-S	1429196_at	432	56	206
17	zinc finger protein 157	Zfp157	scl0072154.1_145-S	1452945_at	138	356	238
18	phosphoserine aminotransferase 1	Psat1	scl52691.13_82-S	1451064_a_at	451	67	238
19	contactin 2	Cntn2	scl021367.1_30-S	1456962_at	584	52	239
20	protocadherin alpha 4	Pcdha4	scl52062.1.1_324-S	1420429_at	322	438	244
21	sulfotransferase family 2A, DHEA-preferring, member 1	Sult2a1	GI_38086373-S	1419528_at	338	262	247
22	ATP-binding cassette, sub-family C (CFTR/MRP), member 4	Abcc4	ri|A830027H05|PX00154E24|AK043744|1727-S	1454408_at	583	105	259
23	family with sequence similarity 3, member C	Fam3c	scl29247.11.6_29-S	1448904_at	508	195	318
24	CD59a antigen	Cd59a	scl00333883.1_4-S	1418710_at	178	659	352
25	CD59b antigen	Cd59b	scl00333883.1_4-S	1418710_at	178	659	418
26	sulfotransferase family 2A, DHEA-preferring, member 2	Sult2a2	scl075142.2_189-S	1419528_at	481	413	418
27	zinc finger protein 446	Zfp446	scl0269870.3_40-S	1439025_at	533	364	447
28	LIM domain containing preferred translocation partner in lipoma	Lpp	GI_38081184-S	1438597_x_at	770	138	448
29	mortality factor 4 like 1	Morf4l1	GI_38081184-S	1438597_x_at	770	138	454
30	RAD52 motif 1	Rdm1	scl40876.7.1_108-S	1417661_at	528	379	454
31	family with sequence similarity 5, member C	Fam5c	scl075142.2_189-S	1446809_at	481	439	460
32	RIKEN cDNA 0610010F05 gene	0610010F05RIK	scl0001343.1_24-S	1456660_a_at	33	899	460
33	spermatid perinuclear RNA binding protein	Strbp	ri|D330022O09|PX00093P17|AK021286|1503-S	1421125_at	510	450	466
34	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, theta polypeptide	Ywhaq	scl022630.1_91-S	1460621_x_at	874	90	482
35	zinc finger protein 644	Zfp644	scl0003996.1_126-S	1439769_at	284	703	494
36	ring finger protein 32	Rnf32	GI_38081184-S	1455742_x_at	770	228	499
37	tetraspanin 2	Tspan2	scl0070747.2_23-S	1424567_at	803	211	507
38	tribbles homolog 3	Trib3	scl18503.5_29-S	1456225_x_at	387	644	511
39	NPC1-like 1	Npc1l1	scl40543.19.1_28-S	1438514_at	267	775	516
40	leucine rich repeat containing 50	Lrrc50	ri|4833436O22|PX00028P19|AK029441|2683-S	1421974_at	197	851	521

Genes were ranked based on p-values and assigned to probes using Gemma (22) and Illumina and Affymetrix provided annotations. In the case multiple probes were targeting a specific gene we only included the probe with the lowest p-value and highest rank for that specific gene. We calculated the combined rank for each gene by averaging the individual ranks obtained from the Affymetrix and the Illumina platform, respectively.

Supplemental Table S2 Combined ranks created from Illumina and Affymetrix genome-wide expression analysis on YAC128 and control mice at 12 months of age.

Rank	Gene name	Gene symbol	Illumina probe	Affymetrix probe	Illu rank	Affy rank	Combined rank
1	myeloid/lymphoid leukemia; translocated to, 10	Mllt10	scl0017354.1_147-S	1444194_at	51	90	70
2	surfeit gene 4	Surf4	scl19517.6_242-S	1436797_a_at	104	81	92
3	actinin alpha 2	Actn2	scl44289.22.3_135-S	1448327_at	4	231	118
4	forkhead box J3	Foxj3	scl0002837.1_1-S	1459055_at	154	181	168
5	transportin 1	Tnpo1	scl0320826.1_66-S	1459341_at	103	316	210
6	dimethylarginine dimethylaminohydrolase 1	Ddah1	GI_38081067-S	1429299_at	48	425	236
7	NEDD4 binding protein 1	N4bp1	scl34522.1.1_225-S	1455606_at	366	218	292
8	WAP four-disulfide core domain 1	Wfdc1	ri|9030625A11|PX00025F06|AK018575|782-S	1431335_a_at	117	562	340
9	zinc finger and SCAN domain containing 5B	Zfp371 (Zscan5b)	scl0170734.1_214-S	1449543_at	391	302	346
10	protein phosphatase 1, regulatory subunit 9A	Ppp1r9a	scl30419.24_421-S	1432037_at	242	469	356
11	RIKEN cDNA A530023O14 gene	A530023O14Rik	scl00244183.1_117-S	1444179_at	635	95	365
12	mitogen-activated protein kinase kinase kinase 12	Map3k12	scl0026404.1_188-S	1438908_at	300	444	372
13	cAMP responsive element modulator	Crem	scl0002242.1_3-S	1418322_at	281	523	402
14	calcium channel, voltage-dependent, α2/Δ subunit 1	Cacna2d1	scl012293.44_74-S	1441608_at	379	487	433
15	LOC628308	LOC628308	GI_38077065-S	1427174_at	432	440	436
16	zinc finger protein 260	Zfp260	scl0026466.2_245-S	1427831_s_at	827	60	444
17	LOC432823	LOC432823	GI_38090966-S	1420500_at	428	498	463
18	RIKEN cDNA 1300010F03 gene	1300010F03Rik	ri|F730029G24|PL00003B03|AK089438|3826-S	1431800_at	472	478	475
19	peptidoglycan recognition protein 1	Pglyrp1	ri|A730072G01|PX00151H04|AK043227|1009-S	1449184_at	179	773	476
20	tripartite motif-containing 14	Trim14	scl0074735.2_104-S	1436199_at	590	427	508
21	prolylcarboxypeptidase	Prcp	ri|2610104A14|ZX00061A19|AK011816|791-S	1444419_at	580	501	540
22	NIPA-like domain containing 3	Nipal3	ri|E230003H01|PX00208F16|AK053937|2624-S	1434315_at	1009	84	546
23	sigma non-opioid intracellular receptor 1	Sigmar1	scl0002696.1_5-S	1451956_a_at	706	385	546
24	regulatory factor X, 4	Rfx4	scl0071137.2_208-S	1436931_at	890	216	553
25	protocadherin alpha 4	Pcdha4	scl093711.1_0-S	1446332_at	547	565	556
26	protocadherin γ subfamily A, 1	Pcdhga1	scl093711.1_0-S	1446332_at	547	565	556
27	protocadherin γ subfamily A, 2	Pcdhga2	scl093711.1_0-S	1446332_at	547	565	556
28	protocadherin γ subfamily A, 3	Pcdhga3	scl093711.1_0-S	1446332_at	547	565	556
29	runt-related transcription factor 1	Runx1t1	GI_38078080-S	1444615_x_at	498	657	578
30	neurofascin	Nfasc	scl000893.1_11-S	1459357_at	1217	14	616
31	phosphodiesterase 4A, cAMP specific	Pde4a	ri|6530402D11|PX00649G05|AK078339|1262-S	1457408_at	238	1013	626
32	roundabout homolog 1	Robo1	ri|A530006L21|PX00139F22|AK040651|2419-S	1454565_at	897	364	630
33	Tp53rk binding protein	Tprkb	scl0001238.1_35-S	1425410_at	1097	204	650
34	zinc finger protein 458	Zfp458	GI_38074875-S	1445824_at	1039	260	650
35	predicted gene 2026	Gm2026	scl076958.5_244-S	1427174_at	873	440	656
36	RIKEN cDNA 2210418O10 gene	2210418O10Rik	scl076958.5_244-S	1427174_at	873	440	656
37	zinc finger, CCHC domain containing 6	Zcchc6	scl0003616.1_4-S	1444421_at	1213	103	658
38	protein phosphatase 1, regulatory (inhibitor) subunit 2	Ppp1r2	ri|4833447I12|PX00313B20|AK076545|2034-S	1447706_at	1289	46	668
39	zinc finger protein 644	Zfp644	scl0003996.1_126-S	1439769_at	736	600	668
40	regulator of G-protein signaling 9	Rgs9	ri|6030456M03|PX00314P15|AK077945|3443-S	1426033_at	1067	299	683

Genes were ranked based on p-values and assigned to probes using Gemma (22) and Illumina and Affymetrix provided annotations. In the case multiple probes were targeting a specific gene we only included the probe with the lowest p-value and highest rank for that specific gene. We calculated the combined rank for each gene by averaging the individual ranks obtained from the Affymetrix and the Illumina platform, respectively.

Supplemental Table S3

Primers used in qPCR for quantification of striatal mRNA from YAC128 and controls

Gene symbol	**Forward primer**	**Reverse primer**
Pcdh20	GAGCCAACTCATAGGAAGGTAGAATCT	GATGGAGCCCAAGCTTATGACT
Tob2	GTTCTTGGAGAAATCGCCCTTT	GGAAGGGCTGGTTGGGATA
Wt1	AAGGACACGACTGTGGATCTACATC	TTCCGGCAAACCTGATAGGA
Ddit4l	GCAGCGCCTAGGGAAAGAT	CTCAAACTGCCCGTTGCAA
Hrbl	CTGAGCCTGAAGTCCTGTTCCT	GGTATCAACGATGTCCGAGCAT
Polr2a	CAGGACACTGGACCGCTCAT	GCATAATATTCTCAGAGACTCCCTTCA
Sfmbt2	AACTCATCCTCCTCTTTGGAAAAG	CAGGTTGGTATCTTCCTCTGAGTAAA
Pogk	GAGACTGTCCTGTCCCTGGAATT	CTGGAGTCCTAAAGTCAGATTCTTCA
Acy3	CCTCACCTTCCTTGGTTCCA	GCAGGTCTAGGGTAAAGTCAAAAGC
DMPK	TGCATGGCCTTCAGAGACAA	GCCACTAGGTCAGCCTCTTCA
Dleu7	TCCCTGTCCACCTGAAGGATA	TGGTGAGCGGCATTCAAGT
Gsg1l	GGAGGACTGGAGACCACACTCTT	CCATGCAGCAGGTGAAGGA
Ntng1	CACAACACTAGAGGGCAGCA	TTTAGGCCCTGTTGTTCCAG
Dzip1l	TGCAGGCATGGCAGATGT	TCCCCTTGGGTCCACTTGA
Dok3	GCACGTAAAGTTTGGCAAGAAAT	TCACGCACGTCCCAGCTT
Rab21	TGTGGGAGCCAAGCATTACC	CCTGGGCTGTCTCTATCATTCTTTTA
Spata5	TCCAGGTCCAGCCTCTTTTG	TTGTCACTCAGCGCCAAGTC
Entpd7	AAAAGGAAACGGAGGTGACAGA	ACGGAGAAACGGGCTCACT
Grasp	GCAGTACCTTAAGCAAACCCTGTAC	TTTCACCACCAGGCCATGTA
Slc45a3	GACACTATGATGAAGGCATTCGAA	TCTGTACCAGCCTGTCCATGAC
Rtel1	CTTTCTCGAAACATGAAACAGCAA	TGCGCCTTACGACTCTTAGCA
Rgag4	GCCCATGTGCACACACTTG	CATCACCATGAGCTCTCCATGT
Ddc	TCCCCAGAGTTCACACAAGCT	ATGCGCCTGATCAGATGTGTAA
Usmg5	GGGTTCGGACGAAGATTGAC	TGAACTGGAATTGGCCATCA
Mllt10	CCAGAAGGTCACAAGACTTAGTGATAA	GGCGAGTTCTGGCCATTTT
Surf4	GAAGAAGAAAGAGTGGTAACACACAGA	CACAAACGCCGGCAGTTT
Foxj3	CCAAGGATGATCCCGGAAA	ATGGAGTTGAGGCCCGTTCT
Actn2	GCGCAGGAGCTCAATGAACT	CTATCCCACTGGTCGCAAATTT
Tnpo1	TTTATGATTGTGGCTCTTGACTTACTC	TTTATCCTGCATGCACTGGTACA
Ddah1	CGCAAAGGTCTATGAGAAACTCAA	AGCCGTCCACCTTTTCCAT
Wfdc1	GCTCTGAGTTTCCTGTTGCTACTG	AACCTCTTCAGCTCGGGACTT
Ppp1r9a	CGGATGAGAGCCTGGATATGAT	GCCCGATTCTGACTCTGAGTGT
N4bp1	CGAGAACCTCCCCAGTAATCAG	TGTCTGCGTGGGCTTCAAC
Rplo	TGGGCATCACCACGAAAAT	ATCAGCTGCACATCACTCAGAATT
Hprt1	CGTCGTGATTAGCGATGATGA	TCCAAATCCTCGGCATAATGA
Rn18s	AGAAACGGCTACCACATCCAA	GGGTCGGGAGTGGGTAATTT
B2m	TCTCACTGACCGGCCTGTATG	TGGGTGGCGTGAGTATACTTGA
Actb	CCAGCCTTCCTTCTTGGGTAT	TGTGTTGGCATAGAGGTCTTTACG
Gapdh	AACTTTGGCATTGTGGAAGG	ACACATTGGGGGTAGGAACA

Supplemental Table S4

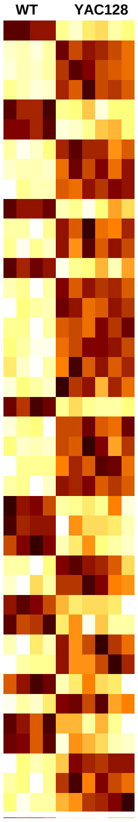
Primers used in qPCR for quantification of caudate mRNA from HD cases and controls.

Gene symbol	**Forward primer**	**Reverse primer**
WT1	CGCCCTACAGCAGTGACAATT	TTAAGGTGGCTCCTAAGTTCATCTG
DDIT4L	AACCCGGCCAGCATTTC	TCCCAGTAGTCAAAATCACTTAGCA
GSG1L	ACTTCCGGGAGAGGATGGA	TGGTGTCGGGCAGGGTAT
SLC45A3	TCGGCCAGGATCTGAGTGAT	CCGCCAACTGCCTAGGAAT
DZIL1L	GCAAGAGTGGACCAAACTTTATGG	TGCCCGCAGTTTCTCTTCTAG
PCDH20	CACAAATCTCAATAGAACCGACTCA	AGAGCTACTAAGGTGGGCATACAAG
SFMBT2	TCGGCTCAGCTAATGGAAATG	AAGCCAGTTTCCTCCAAGCTT
ACY3	AGCTGCAGGACCGAGACTTC	CCGTGGACTCTCCCTCATAGAG
POLR2A	TCTGCACAAAATGTCCATGATG	CGGAGTTGTCACACTAAGATTCAAG
ACTN2	CGATGGAGCACATTCGTGTT	TTCGCATCTCTCGTCAGGATCT
DDAH1	TTTAAGGACTATGCAGTCTCCACAGT	AGCCATGCTGCAGAAACTCTTC
PPP1R9A	CCATCAGAACTGGACACAAGCA	GCCTGCAGCTTGGTCTTCA
DMPK	CATCGTGGTGAGGCTTAAGGA	CACTACCGCTACCTCGCTGAA
GSG1L	ACTTCCGGGAGAGGATGGA	TGGTGTCGGGCAGGGTAT
RGAG4	GGACAGCGCCCAACATTG	CTGGCTACCCTTTAGGCAACA
HPRT1	TTATGGACAGGACTGAACGTCTTG	GCACACAGAGGGCTACAATGTG
PGK-1	CAAATGGAACACGGAGGATAAAG	CTTTACCTTCCAGGAGCTCCAA
ACTB	AGTACTCCGTGTGGATCGGC	GCTGATCCACATCTGCTGGA
GAPDH	GAAGGTGAAGGTCGGAGTC	GAAGATGGTGATGGGATTTC
B2M	AATATAAGTGGAGGCGTCGCGC	TAAGGCCACGGAGCGAGACATC
18S RNA	CGCCGCTAGAGGTGAAATTC	TTGGCAAATGCTTTCGCTC

Supplemental Figures and Figure legends

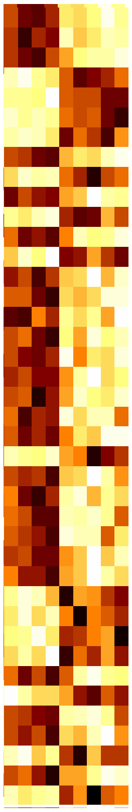
Supplemental Figure S1

Affymetrix



Name	Symbol	Probe	P-value	Fold change
Wilms tumor homolog	*Wt1*	1425995_s_at	2.10E-06	0.76
DNA-damage-inducible transcript 4-like	*Ddit4l*	1439332_at	2.89E-06	-1.16
dystrophia myotonica-protein kinase	*Dmpk*	1434944_at	1.51E-05	-0.35
GSG1-like	*Gsg1l*	1436013_at	1.54E-05	-0.53
deleted in lymphocytic leukemia, 7	*Dleu7*	1442180_at	2.15E-05	0.62
spermatogenesis associated 5	*Spata5*	1438844_x_at	4.14E-05	0.48
deiodinase, iodothyronine type III, opposite strand	*Dio3os*	1435121_at	5.64E-05	-0.24
ectonucleoside triphosphate diphosphohydrolase 7	*Entpd7*	1435625_at	5.72E-05	-0.43
GRP1 (general receptor for phosphoinositides 1)-associated scaffold protein	*Grasp*	1441894_s_at	9.11E-05	-0.39
solute carrier family 45, member 3	*Slc45a3*	1426663_s_at	1.02E-04	1.06
regulator of telomere elongation helicase 1	*Rtel1*	1435516_x_at	1.02E-04	-0.14
cleft lip and palate associated transmembrane protein 1	*Clptm1*	1441083_at	1.32E-04	-0.43
G protein-coupled receptor 101	*Gpr101*	1441382_at	1.35E-04	1.54
tyrosine kinase, non-receptor, 2	*Tnk2*	1448298_at	1.79E-04	-0.43
Leucine rich repeat containing 49	*Lrrc49*	1445486_at	2.00E-04	-0.20
G protein-coupled receptor 153	*Gpr153*	1426973_at	2.45E-04	-0.68
RIKEN cDNA BB336256 gene	*BB336256Rik*	1436092_at	2.51E-04	-1.65
MAS-related GPR, member B1	*Mrgprb1*	1437529_at	2.71E-04	-0.18
RIKEN cDNA BB667665 gene	*BB667665Rik*	1444415_at	2.84E-04	-0.26
protocadherin 20	*Pcdh20*	1424701_at	2.96E-04	1.11
reticulon 3	*Rtn3*	1443220_at	3.03E-04	-0.35
PITPNM family member 3	*Pitpnm3*	1455140_at	3.03E-04	-040
phospholipase C-like 4	*Plcl4*	1441587_at	3.26E-04	-0.20
heparan sulfate (glucosamine) 3-O-sulfotransferase 3A1	*Hs3st3a1*	1435622_at	3.28E-04	-0.28
programmed cell death 6 interacting protein	*Pdcd6ip*	1426184_a_at	3.93E-04	0.19
ankyrin repeat domain 15	*Ankrd15*	1433742_at	4.06E-04	0.31
netrin G1	*Ntng1*	1449286_at	4.64E-04	0.80
G-protein signalling modulator 1 (AGS3-like, C. elegans)	*Gpsm1*	1423689_a_at	5.25E-04	-0.37
hypothetical LOC676575	*LOC676575*	1454312_at	5.59E-04	-0.12
G-protein coupled receptor 85	*Gpr85*	1424897_at	5.66E-04	0.19
ubiquitin specific peptidase 25	*Usp25*	1448939_at	5.67E-04	0.29
HIV-1 Rev binding protein-like	*Hrbl*	1441347_at	6.04E-04	-0.20
ribonuclease, RNase A family 4	*Rnase4*	1422603_at	6.13E-04	-0.67
cholecystokinin A receptor	*Cckar*	1421195_at	6.36E-04	0.26
RIKEN cDNA D630029P09 gene	*D630029P09Rik*	1455468_at	6.57E-04	-0.48
TBCC domain containing 1	*Tbccd1*	1436752_at	6.82E-04	0.34
beta-1,4-N-acetyl-galactosaminyl transferase 2	*B4galnt2*	1421522_at	6.83E-04	0.26
Usher syndrome 2A (autosomal recessive, mild) homolog (human)	*Ush2a*	1445972_at	7.14E-04	-0.18
RIKEN cDNA 2410089E03 gene	*2410089E03Rik*	1443422_at	7.33E-04	-0.42
regulatory factor X, 3 (influences HLA class II expression)	*Rfx3*	1425413_at	7.58E-04	-0.20

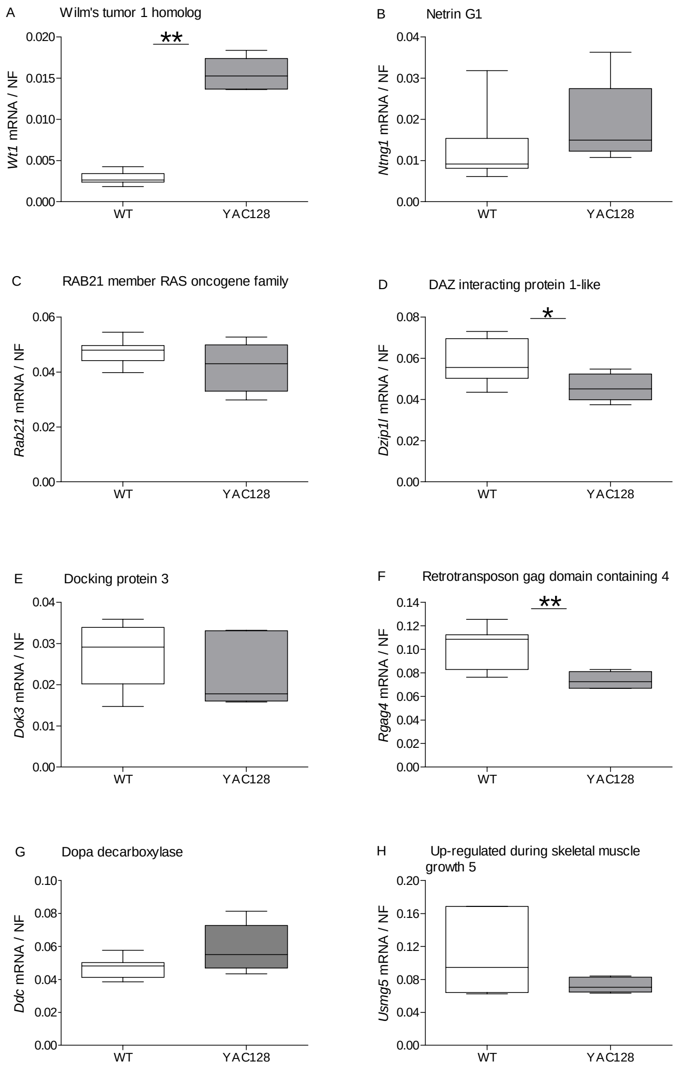
Illumina



Name	Symbol	Probe	P-value	Fold change
Wilms tumor homolog	*Wt1*	scl022431.11_253-S	1.46E-04	0.20
netrin G1	*Ntng1*	scl0080883.1_122-S	3.05E-04	0.06
Rab21; member RAS oncogene family	*Rab21*	scl37488.7_270-S	4.60E-04	0.11
DAZ interacting protein 1-like	*Dzip1l*	scl072507.17_285-S	5.35E-04	-0.05
docking protein 3	*Dok3*	scl43920.5.585_30-S	8.48E-04	-0.07
Olfactory receptor 1502	*Olfr1502*	scl0258793.1_202-S	9.84E-04	-0.03
fibronectin type III domain containing 8	*Fndc8*	scl41151.4.1_158-S	1.04E-03	-0.02
weakly similar to STEROL C5-desaturase	*D830041I17Rik*	ri|D830041I17	1.05E-03	0.02
retrotransposon gag domain containing 4	*Rgag4*	scl53982.1_257-S	1.10E-03	-0.16
dopa decarboxylase	*Ddc*	scl40512.21.1_20-S	1.22E-03	0.13
upregulated during skeletal muscle growth 5	*Usmg5*	scl0066477.1_316-S	1.31E-03	-0.02
Scm-like with four mbt domains 2	*Sfmbt2*	scl21295.26.802_230-S	1.42E-03	0.06
RIKEN cDNA 5530601H04 gene	*5530601H04Rik*	scl0071445.1_39-S	2.08E-03	-0.02
annexin A5	*Anxa5*	scl011747.1_83-S	2.20E-03	0.24
transmembrane protein 22	*Tmem22*	GI_38090024-S	2.47E-03	0.17
solute carrier family 47, member 1	*Slc47a1*	scl0001298.1_25-S	2.59E-03	0.03
RIKEN cDNA E330009J07 gene	*E330009J07Rik*	scl29095.10_195-S	3.02E-03	0.20
inhibitor of growth family, member 3	*Ing3*	scl30327.11_168-S	3.49E-03	0.05
leucine rich repeat protein 1, neuronal	*Lrrn1*	scl29678.2_366-S	3.58E-03	0.25
folylpolyglutamyl synthetase	*Fpgs*	scl0003272.1_5-S	3.74E-03	0.03
protein kinase, AMP-activated, alpha 2 catalytic subunit	*Prkaa2*	GI_28545636-S	3.76E-03	0.02
leucine-rich repeats and calponin homology (CH) domain containing 3	*Lrch3*	scl070144.7_24-S	3.81E-03	0.04
molybdenum cofactor synthesis 1	*Mocs1*	ri|3110045D15	4.14E-03	-0.03
transducer of ERBB2, 2	*Tob2*	scl0057259.2_60-S	4.23E-03	0.14
solute carrier family 30 (zinc transporter), member 8	*Slc30a8*	scl47916.2.1_25-S	4.37E-03	0.02
RIKEN cDNA 1110007C09 gene	*1110007C09Rik*	scl0003702.1_48-S	4.42E-03	0.02
aldo-keto reducase family 1 member B3 (aldose reductase)	*Akr1b3*	scl17956.1.1_169-S	4.45E-03	0.02
sema domain, transmembrane domain (TM), and cytoplasmic domain, (semaphori	*Sema6a*	scl51424.2_347-S	4.89E-03	0.03
RIKEN cDNA 1190002N15 gene	*1190002N15Rik*	GI_38089999-S	5.01E-03	0.19
tubulin gamma complex associated protein 6	*Tkbgcp6*	scl00328580.1_176-S	5.06E-03	-0.02
Zinc finger FYVE domain containing protein 26	*Zfyve26*	scl0211978.1_134-S	5.33E-03	-0.06
RIKEN cDNA C030037F17 gene	*C030037F17Rik*	scl35066.3.1_104-S	5.42E-03	-0.03
RIKEN cDNA 0610010F05 gene	*0610010F05Rik*	scl0001343.1_24-S	5.49E-03	-0.02
proline rich 5 like	*Prr5l*	scl18958.13.1_24-S	5.64E-03	0.13
predicted gene 7461	*Gm7461*	ri|4930580J09	5.80E-03	-0.03
solute carrier family 35, member F4	*Slc35f4*	GI_38076112-S	5.92E-03	0.03
preferentially expressed antigen in melanoma	*Prame*	scl53822.10.1_312-S	5.93E-03	0.03
cancer antigen 1	*Cage1*	scl0003725.1_2-S	5.99E-03	-0.03
dynein cytoplasmic 1 intermediate chain 2	*Dync1i2*	scl20807.21.1_18-S	6.02E-03	0.10
RIKEN cDNA 1700020D12 gene	*1700020D12Rik*	scl42585.2.1_143-S	6.12E-03	-0.01

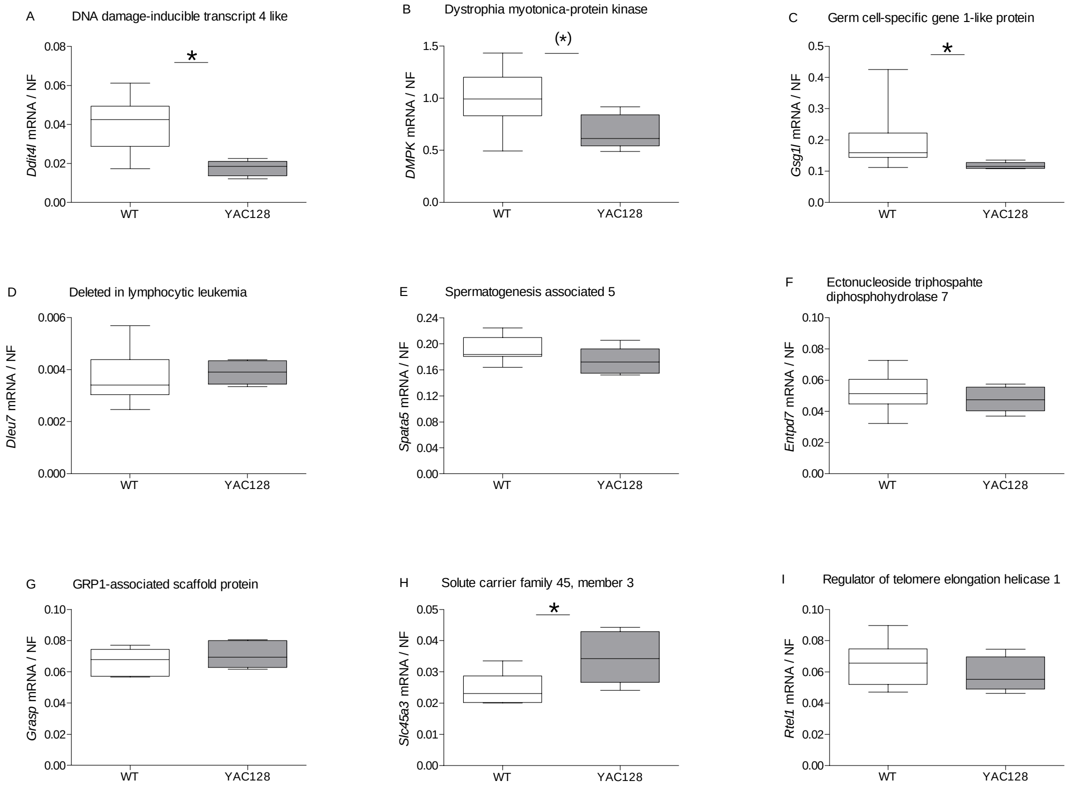
Supplemental Figure S1 Genome-wide expression profiling of striatal mRNA from 24 month old YAC128 mice and controls using Affymetrix and Illumina beadarray platforms. Higher transcriptional expression levels are indicated by a light yellow color, while a darker brown color is corresponding to lower transcriptional expression in the heat map. Ranking is here based on p-values prior to correction for multiple comparisons. Fold change is shown as $\log_2$ transformed values. Positive values indicate up-regulation in YAC128 compared to controls; Negative values indicate down-regulation in YAC128 compared to controls. YAC128 (n=6); wildtype littermate controls (n=4).

Supplemental Figure S2



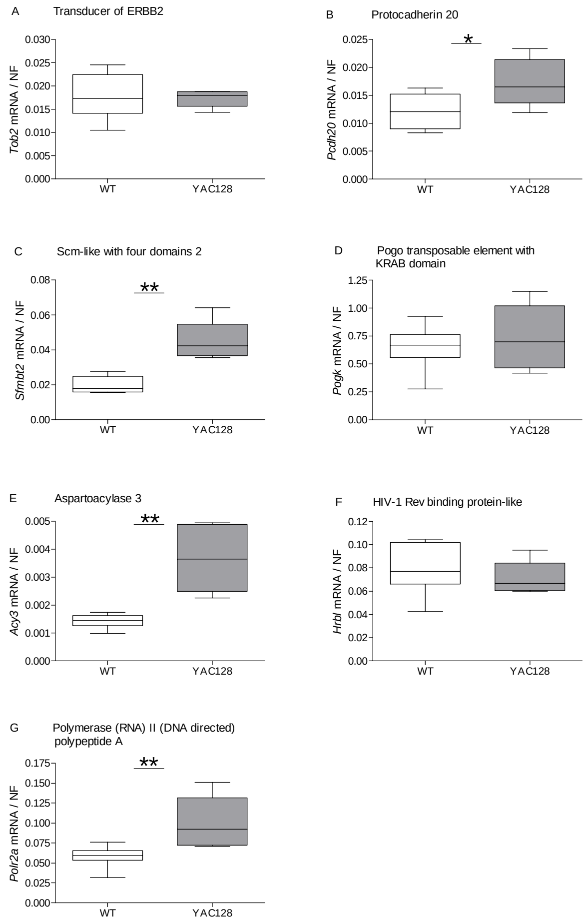
Supplemental Figure S2 Quantification of the mRNA expression assessed by qPCR of genes identified by Illumina genome-wide expression analysis. The genes that were ranked as top ten on the Illumina platform for 24 month old YAC128 and wildtype controls (WT) were analyzed. Three of the genes were validated to be differentially expressed between YAC128 and WT. *Wt1* showed up-regulation in YAC128 compared to controls (p=0.0012) (**A**). *Ntng1* (**B**) showed a trend for up-regulation that was not statistically significant. *Dzip1l* (p=0.022) (**D**) and *Rgag4* (p=0.0082) (**F**) were both down-regulated in YAC128. *Rab21* (**C**), *Dok3* (**E**), *Ddc* (**G**) and *Usmg5* (**H**) were not differentially expressed. Three different primer sets were tested for *Olfr1502* and *Fndc8*, but we were unable to amplify and quantify the expression of these genes from mouse striatal RNA. The middle line of the box and whiskers plot shows the median, the top and bottom lines show the 75th and 25th percentile, respectively. The top and bottom whiskers indicate the largest and smallest values. Statistical analysis was performed using Mann-Whitney 2-tailed U test; * $p<0.05$; ** $p< 0.01$; *** $p<0.001$.
YAC128 (n= 7); WT (n=6)

Supplemental Figure S3



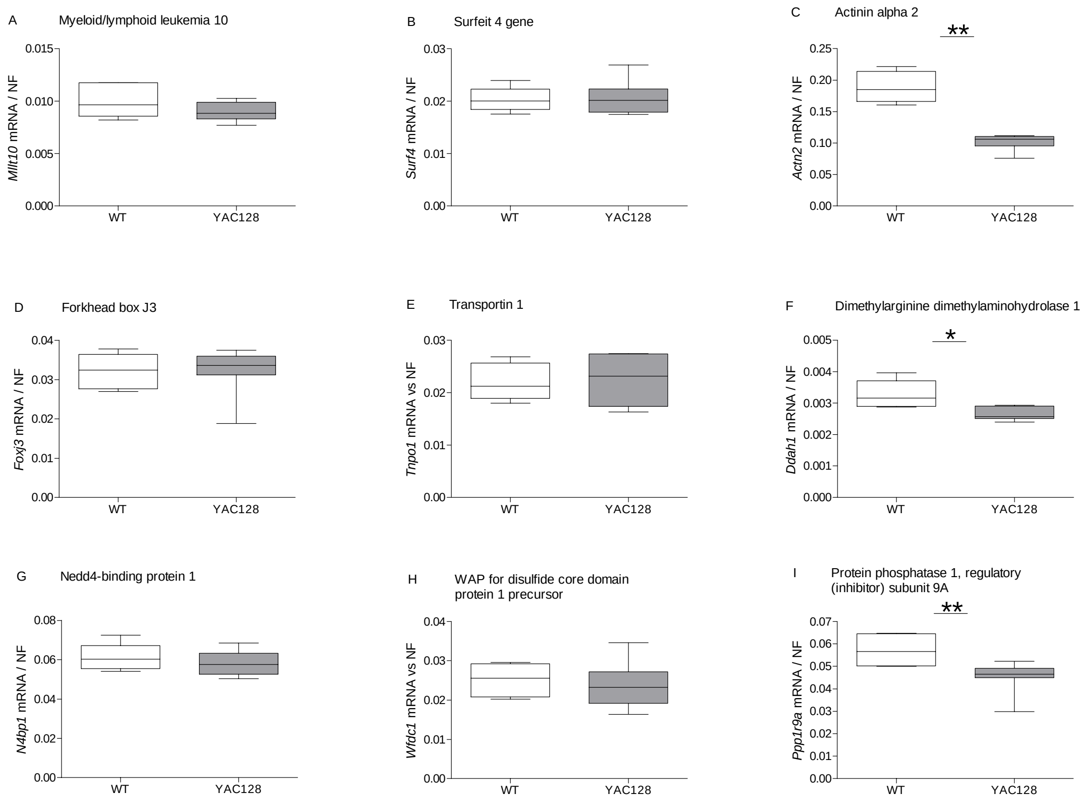
Supplemental Figure S3 Quantification of the mRNA expression assessed by qPCR of genes identified by Affymetrix genome-wide expression analysis. The genes that were ranked as top ten on the Affymetrix platform for 24 month old YAC128 and wildtype controls (WT) were analyzed. Four of the Affymetrix genes were validated to be differentially expressed between YAC128 and WT. *Ddit4l*, *Gsg1l* and *Slc45a3* were differentially expressed, in addition to *Wt1*. *Ddit4l* (p=0.014) (**A**) and *Gsg1l* (p=0.018) (**C**) were down-regulated in YAC128. *Dmpk* showed a non-significant trend for down-regulation in YAC128 (p=0.051) (**B**). *Dleu7*, *Spata5*, *Entpd7*, *Grasp* and *Rtel1* did not show differential expression between YAC128 and WT (**D-G, I**). *Slc45a3* (p=0.015) (**H**) was up-regulated in YAC128. The middle line of the box and whiskers plot shows the median, the top and bottom lines show the 75th and 25th percentile, respectively. The top and bottom whiskers indicate the largest and smallest values. Statistical analysis was performed using Mann-Whitney 2-tailed U test; * $p<0.05$;
** $p< 0.01$; *** $p<0.001$. YAC128 (n= 7); WT (n=6)

Supplemental Figure S4



Supplemental Figure S4 Quantification of the mRNA expression assessed by qPCR of genes from the combined rank list for the 24 month old YAC128 and controls. We created the combined rank by averaging the individual ranks on the Affymetrix and Illumina platforms. We analyzed the top ten genes from the combined rank list. In addition to *Wt1* and *Ddit4l*, four additional genes were validated to be differentially expressed in YAC128 compared to controls at 24 months of age: *Pcdh20* (p=0.035) (**B**), *Sfmbt2* (p=0.0012) (**C**), *Acy3* (p=0.0012) (**E**) and *Polr2a* (p=0.0047) (**G**)**.** All four genes were significantly up-regulated in YAC128 compared to controls. *Tob2* (**A**), *Pogk* (**D**) and *Hrbl* (**F**) were not differentially expressed in YAC128 and controls at 24 months of age. The middle line of the box and whiskers plot shows the median, the top and bottom lines show the 75th and 25th percentile, respectively. The top and bottom whiskers indicate the largest and smallest values. Statistical analysis was performed using Mann-Whitney 2-tailed U test; * $p<0.05$; ** $p< 0.01$; *** $p<0.001$. YAC128 (n= 6); WT (n=7)

Supplemental Figure S5



Supplemental Figure S5 Quantification of the mRNA expression assessed by qPCR of genes from the combined rank list for the 12 month old YAC128 and controls. We created the combined rank by averaging the individual ranks on the Affymetrix and Illumina platforms. We analyzed the top ten genes from the combined rank list. Three out of ten genes were validated to be differentially expressed in YAC128 compared to controls at 12 months of age: *Actn2* (p=0.0012) (**C**), *Ddah1* (p=0.0221) (**F**) and *Ppp1r9a* (p=0.0047) (**I**). *Mllt10* (**A**), *Surf4* (**B**), *Foxj3* (**D**), *Tnpo1* (**E**), *N4bp1* (**G**) and *Wfdc1* (**H**) were not differentially expressed. Three different primer sets were tested for *Zfp371*, but we were unable to amplify and quantify the expression of this gene from mouse striatal RNA. The middle line of the box and whiskers plot shows the median, the top and bottom lines show the 75th and 25th percentile, respectively. The top and bottom whiskers indicate the largest and smallest values. Statistical analysis was performed using Mann-Whitney 2-tailed U test; * p<0.05; ** p< 0.01; *** p<0.001.
YAC128 (n= 7), WT (n=6)