

On-line Table 1: CLN2 mutations with corresponding proteins

Subject Identifier	Allele 1			Allele 2		
	gDNA	cDNA	Protein	gDNA	cDNA	Protein
BDrh-01	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-02	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-03	G3556C	509-IG>C	Intron	G3556C	509-IG>C	Intron
BDrh-04	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-05	C3670T	622C>T	Arg208X	G4655A	1094G>A	Cys365Tyr
BDrh-06	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-07	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-09	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-10	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-11	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-12	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-13	G3556C	509-IG>C	Intron	G4655A	1094G>A	Cys365Tyr
BDrh-14	G3556C	509-IG>C	Intron	G3556C	509-IG>C	Intron
BDrh-15	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-16	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X
BDrh-17	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-18	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-19	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-20	G3556C	509-IG>C	Intron	G3556C	509-IG>C	Intron
BDrh-21	G3556C	509-IG>C	Intron	Other	Other	Other
BDrh-22	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X
BDrh-23	C3670T	622C>T	Arg208X	Other	Other	Other
BDrh-24	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X
BDrh-25	Other	Other	Other	Other	Other	Other
BDrh-26	C3670T	622C>T	Arg208X	Other	Other	Other
BDrh-27	C3670T	622C>T	Arg208X	Other	Other	Other
BDrh-28	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X
BDrh-29	G3556C	509-IG>C	Intron	G3556C	509-IG>C	Intron
BDrh-30	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-31	C3670T	622C>T	Arg208X	Other	Other	Other
BDrh-32	G4655A	1094G>A	Cys365Tyr	Other	Other	Other
BDrh-33	C3670T	622C>T	Arg208X	Other	Other	Other
BDrh-34	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-35	C3670T	622C>T	Arg208X	G3556C	509-IG>C	Intron
BDrh-36	G3556C	509-IG>C	Intron	G3556C	509-IG>C	Intron
BDrh-37	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X
BDrh-38	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X
BDrh-39	C3670T	622C>T	Arg208X	C3670T	622C>T	Arg208X

Note:—gDNA indicates genomic DNA; cDNA, complementary DNA.

On-line Table 2: The rate of decline (millimeters/month) and mean cortical thickness in the left hemisphere between CLN2 and healthy control populations

Left Hemisphere	CLN2 Slope	Control Slope	P Value	CLN2 Mean	Control Mean	P Value
Bankssts	−0.035(9)	−0.003(3)	<.001	2.28(39)	2.90(22)	<.001
Caudal anterior cingulate	−0.008(14)	−0.001(4)	1.000	2.68(50)	2.83(34)	.571
Caudal middle frontal	0.000(5)	−0.002(3)	1.000	2.27(19)	2.90(21)	<.001
Cuneus	−0.034(12)	−0.004(3)	<.001	1.65(33)	2.14(22)	<.001
Entorhinal	−0.018(13)	−0.005(6)	.600	2.64(46)	3.14(53)	<.001
Frontal pole	0.005(24)	−0.003(4)	1.000	2.35(47)	2.92(35)	<.001
Fusiform	−0.029(8)	−0.001(2)	<.001	2.43(37)	2.81(18)	<.001
Inferior parietal	−0.023(6)	−0.002(2)	<.001	2.24(31)	2.92(18)	<.001
Inferior temporal	−0.029(10)	−0.002(2)	<.001	2.39(35)	2.91(19)	<.001
Insula	−0.015(7)	−0.004(3)	.067	2.90(26)	3.30(19)	<.001
Isthmus cingulate	−0.036(10)	−0.002(3)	<.001	2.50(41)	2.84(20)	<.001
Lateral occipital	−0.028(9)	−0.003(3)	<.001	1.88(30)	2.37(17)	<.001
Lateral orbitofrontal	−0.019(9)	−0.002(3)	.012	2.57(28)	2.88(19)	<.001
Lingual	−0.025(9)	−0.003(2)	<.001	1.73(27)	2.24(16)	<.001
Medial orbitofrontal	−0.016(8)	−0.002(3)	.028	2.50(30)	2.78(25)	<.001
Middle temporal	−0.014(7)	−0.001(2)	.007	2.58(34)	3.16(18)	<.001
Paracentral	−0.020(11)	−0.003(3)	.039	2.09(29)	2.71(19)	<.001
Parahippocampal	−0.029(8)	−0.002(5)	<.001	2.55(43)	2.74(42)	.003
Pars opercularis	−0.005(7)	−0.001(2)	1.000	2.40(19)	2.92(14)	<.001
Pars orbitalis	0.011(17)	0.000(5)	1.000	2.55(33)	2.98(31)	.005
Pars triangularis	0.000(8)	−0.001(2)	1.000	2.23(23)	2.87(22)	<.001
Pericalcarine	−0.016(12)	−0.004(3)	.514	1.37(19)	1.86(29)	<.001
Postcentral	−0.011(5)	−0.001(2)	<.001	1.80(19)	2.29(14)	<.001
Posterior cingulate	−0.025(12)	−0.003(3)	.006	2.52(38)	2.88(19)	<.001
Precentral	−0.012(6)	0.000(2)	.002	2.12(20)	2.73(15)	<.001
Precuneus	−0.031(10)	−0.004(2)	<.001	2.23(36)	2.81(16)	<.001
Rostral anterior cingulate	−0.001(12)	−0.006(5)	1.000	3.04(43)	3.15(42)	.075
Rostral middle frontal	0.008(5)	−0.002(2)	.012	2.13(20)	2.78(16)	<.001
Superior frontal	−0.004(4)	−0.002(2)	1.000	2.46(21)	3.14(14)	<.001
Superior parietal	−0.021(6)	−0.002(2)	<.001	1.98(25)	2.56(17)	<.001
Superior temporal	−0.021(6)	−0.001(2)	<.001	2.37(30)	3.03(15)	<.001
Supramarginal	−0.017(7)	−0.002(2)	<.001	2.26(30)	2.96(15)	<.001
Temporal pole	0.000(12)	0.000(8)	1.000	3.28(47)	3.48(60)	.231
Transverse temporal	−0.029(8)	−0.003(3)	<.001	2.08(34)	2.71(22)	<.001

Note:—Bankssts indicates banks of the superior temporal sulcus.

On-line Table 3: The rate of decline (millimeters/month) and mean cortical thickness in the right hemisphere between CLN2 and healthy control populations

Right Hemisphere	CLN2 Slope	Control Slope	P Value	CLN2 Mean	Control Mean	P Value
Bankssts	−0.027(11)	−0.003(4)	<.001	2.41(34)	2.93(22)	<.001
Caudal anterior cingulate	−0.015(7)	−0.003(3)	.148	2.61(52)	2.74(26)	.008
Caudal middle frontal	−0.018(8)	−0.001(3)	<.001	2.37(24)	2.91(22)	<.001
Cuneus	−0.028(11)	−0.004(3)	<.001	1.65(30)	2.19(24)	<.001
Entorhinal	−0.016(9)	−0.005(7)	.321	2.91(41)	3.40(63)	<.001
Frontal pole	0.014(17)	−0.004(5)	.272	2.36(36)	2.92(34)	<.001
Fusiform	−0.029(8)	−0.002(3)	<.001	2.47(39)	2.85(19)	<.001
Inferior parietal	−0.020(6)	−0.002(2)	<.001	2.38(27)	2.95(17)	<.001
Inferior temporal	−0.026(6)	−0.001(3)	<.001	2.45(33)	2.97(18)	<.001
Insula	−0.022(8)	−0.005(3)	.002	2.93(28)	3.32(24)	<.001
Isthmus cingulate	−0.029(12)	−0.004(4)	<.001	2.46(44)	2.80(24)	<.001
Lateral occipital	−0.030(7)	−0.003(2)	<.001	1.94(31)	2.48(16)	<.001
Lateral orbitofrontal	−0.024(9)	−0.002(3)	<.001	2.65(32)	2.85(26)	<.001
Lingual	−0.024(9)	−0.005(2)	<.001	1.74(28)	2.31(21)	<.001
Medial orbitofrontal	−0.026(10)	−0.002(4)	<.001	2.54(33)	2.82(31)	<.001
Middle temporal	−0.013(6)	−0.002(3)	.006	2.74(27)	3.21(20)	<.001
Paracentral	−0.020(9)	−0.002(2)	.003	2.07(28)	2.67(16)	<.001
Parahippocampal	−0.021(12)	−0.001(6)	.029	2.53(45)	2.72(39)	.013
Pars opercularis	−0.021(8)	−0.001(3)	<.001	2.49(27)	2.93(19)	<.001
Pars orbitalis	0.001(16)	−0.003(4)	1.000	2.70(30)	3.02(28)	<.001
Pars triangularis	−0.027(5)	−0.001(3)	<.001	2.42(32)	2.92(25)	<.001
Pericalcarine	−0.012(7)	−0.005(3)	.321	1.36(15)	1.88(28)	<.001
Postcentral	−0.020(8)	−0.002(2)	<.001	1.83(24)	2.31(13)	<.001
Posterior cingulate	−0.018(8)	−0.001(2)	<.001	2.48(36)	2.82(18)	<.001
Precentral	−0.018(7)	0.000(2)	<.001	2.16(24)	2.74(14)	<.001
Precuneus	−0.033(11)	−0.003(3)	<.001	2.21(37)	2.76(17)	<.001
Rostral anterior cingulate	−0.023(15)	−0.003(5)	.119	3.21(51)	3.06(39)	.277
Rostral middle frontal	−0.006(5)	−0.002(2)	.505	2.22(22)	2.74(21)	<.001
Superior frontal	−0.013(6)	−0.001(2)	.002	2.49(23)	3.09(20)	<.001
Superior parietal	−0.025(9)	−0.001(2)	<.001	2.03(29)	2.58(15)	<.001
Superior temporal	−0.019(8)	−0.002(2)	<.001	2.49(29)	3.08(14)	<.001
Supramarginal	−0.019(7)	0.000(3)	<.001	2.37(27)	2.95(18)	<.001
Temporal pole	−0.002(15)	−0.001(7)	1.000	3.37(54)	3.61(54)	.080
Transverse temporal	−0.028(10)	−0.002(3)	<.001	2.16(34)	2.65(25)	<.001

Note:—Bankssts indicates banks of the superior temporal sulcus.

On-line Table 4: The rate of decline (millimeters/month) and mean cortical thickness for Brodmann areas in the left and right hemispheres between control and CLN2 populations

Hemisphere	CLN2 Slope	Control Slope	P Value	CLN2 Mean	Control Mean	P Value
Left						
BA1	−0.010(6)	0.000(3)	.033	1.91(24)	2.33(23)	<.001
BA2	−0.011(6)	−0.001(3)	.008	1.90(23)	2.54(19)	<.001
BA3a	−0.008(4)	−0.003(2)	.053	1.41(18)	2.01(17)	<.001
BA3b	−0.010(8)	−0.001(2)	.124	1.65(18)	2.02(15)	<.001
BA4a	−0.022(11)	−0.001(2)	.003	2.24(32)	2.78(20)	<.001
BA4p	−0.010(7)	0.000(3)	.049	1.93(22)	2.47(21)	<.001
BA6	−0.007(6)	−0.001(2)	.124	2.38(20)	3.04(15)	<.001
BA44	−0.008(6)	−0.001(2)	.043	2.40(19)	2.87(13)	<.001
BA45	0.002(8)	−0.001(3)	.468	2.21(22)	2.84(21)	<.001
V1	−0.014(14)	−0.004(3)	.329	1.43(20)	1.89(27)	<.001
V2	−0.031(12)	−0.003(2)	<.001	1.76(32)	2.23(17)	<.001
V5/MT	−0.026(9)	−0.002(3)	<.001	2.00(33)	2.65(19)	<.001
Right						
BA1	−0.020(8)	−0.002(3)	<.001	2.06(29)	2.52(24)	<.001
BA2	−0.026(11)	−0.001(3)	<.001	1.86(32)	2.45(20)	<.001
BA3a	−0.010(4)	−0.003(3)	.003	1.39(18)	2.00(20)	<.001
BA3b	−0.012(7)	−0.002(3)	.014	1.54(19)	1.86(21)	<.001
BA4a	−0.021(10)	0.001(2)	<.001	2.21(30)	2.68(19)	<.001
BA4p	−0.013(7)	−0.001(2)	<.001	1.95(23)	2.43(19)	<.001
BA6	−0.014(5)	0.000(3)	<.001	2.41(22)	3.03(17)	<.001
BA44	−0.018(10)	−0.001(2)	<.001	2.37(27)	2.85(12)	<.001
BA45	−0.022(5)	−0.002(3)	<.001	2.46(28)	2.91(20)	<.001
V1	−0.013(6)	−0.004(3)	.018	1.43(16)	1.97(26)	<.001
V2	−0.031(9)	−0.004(2)	<.001	1.78(32)	2.32(18)	<.001
V5/MT	−0.030(8)	−0.002(2)	<.001	2.10(35)	2.69(19)	<.001

Note:—BA1/2/3a/3b indicates the somatosensory areas; BA4a, primary motor area (anterior); BA4p, primary motor area (posterior); BA6, premotor area; BA44, Broca area (pars opercularis); BA45, Broca area (pars triangularis); V1 (BA 17), primary visual area; V2 (BA 18), secondary visual area; V5/MT, visual area, middle temporal.