

On-line Table 1: Distribution of signal changes stratified according to the additional involvement of deep gray matter (cortex only, cortex and striatum, or cortex, striatum, and thalamus; χ^2 test, 2 degrees of freedom)

Cortical Area	Cortex Only (N = 3)	Cortex and Striatum (N = 16)	Cortex, Striatum, and Thalamus (N = 20)	χ^2	P*
	n (%)	n (%)	n (%)		
Insula	3 (100)	15 (93.75)	19 (95)	0.2042	.902
Cingulate gyrus	3 (100)	13 (81.25)	15 (75)	1.0520	.5911
Superior frontal gyrus	3 (100)	15 (93.75)	19 (95)	0.2042	.902
Paracentral lobe	3 (100)	9 (56.25)	18 (90)	6.6790	.0355*
Precuneus	3 (100)	15 (93.75)	16 (80)	1.9820	.3713
Occipital gyrus	3 (100)	13 (81.25)	15 (75)	1.0520	.5911
Parietal superior lobe	1 (33)	5 (31.25)	10 (50)	1.3710	.5038
Angular/supramarginal lobe	3 (100)	9 (56.25)	13 (65)	2.1160	.3472
Medial frontal gyrus	3 (100)	13 (81.25)	16 (80)	0.7204	.6975
Cuneus	3 (100)	14 (87.5)	14 (70)	2.5080	.2853
Inferior frontal gyrus	1 (33)	10 (62.5)	9 (45)	1.5090	.4703
Medial temporal gyrus	2 (67)	6 (37.5)	7 (35)	1.1160	.5724
Precentral gyrus	1 (33)	5 (31.25)	10 (50)	1.1370	.5038
Postcentral gyrus	1 (33)	4 (25)	6 (30)	0.1519	.9268
Superior temporal gyrus	2 (67)	5 (31.25)	5 (25)	2.1290	.3449

* P < .05. Correction for multiple analyses: results are not significant.

On-line Table 2: Distribution of signal abnormalities on FLAIR and DW images (n = 18 patients with both DW and FLAIR sequences)

Cortical Area	DW Only (N = 8)	DW > FLAIR (N = 6)	DW = FLAIR (N = 3)	FLAIR > DW (N = 1)	χ^2	P
	n (%)	n (%)	n (%)	n (%)		
Insula	8 (100)	6 (100)	3 (100)	1	NA	NA
Cingulate gyrus	7 (88)	6 (100)	3 (100)	1	NA	NA
Superior frontal gyrus	8 (100)	6 (100)	3 (100)	1	NA	NA
Paracentral lobe	6 (75)	6 (100)	3 (100)	1	2.813	.4214
Precuneus	6 (75)	6 (100)	3 (100)	1	2.813	.4214
Occipital gyrus	6 (75)	6 (100)	3 (100)	1	2.813	.4214
Parietal superior lobe	3 (38)	5 (83)	3 (100)	0	6.604	.0857
Angular/supramarginal lobe	2 (25)	4 (67)	3 (100)	0	6.667	.0833
Medial frontal gyrus	7 (88)	5 (83)	3 (100)	1	0.7031	.8725
Cuneus	6 (75)	6 (100)	3 (100)	0	7.200	.0658
Inferior frontal gyrus	4 (50)	3 (50)	3 (100)	0	3.825	.281
Medial temporal gyrus	3 (38)	4 (67)	2 (67)	1	2.306	.5113
Precentral gyrus	3 (38)	2 (33)	3 (100)	0	5.006	.1713
Postcentral gyrus	2 (25)	1 (17)	3 (100)	1	8.182	.0424*
Superior temporal gyrus	1 (13)	2 (33)	2 (67)	0	3.669	.2995

Note:—DW indicates diffusion-weighted; FLAIR, fluid-attenuated inversion recovery; NA, not applicable. The cortical changes were either only detectable on DW images (column 1, n = 8 patients); or DW images showed the cortical changes more effectively than FLAIR (column 2, n = 8 patients); or DW and FLAIR showed the cortical changes equally (column 3, n = 3 patients); or FLAIR showed the pathologic changes more effectively than DW (column 4, n = 1 patient). The comparison of the sequences did not show significant differences (χ^2 test, 3 degrees of freedom).

*P < 0.05. Correction for multiple analyses: results are not significant.

On-line Table 3: Stratification of patients according to Codon 129 (χ^2 test, 2 degrees of freedom)

Cortical Area	MM (N = 25)	MV (N = 4)	VV (N = 5)	χ^2	P*
	n (%)	n (%)	n (%)		
Insula	24 (96)	4 (100)	5 (100)	0.3709	.8307
Cingulate gyrus	24 (96)	4 (100)	5 (100)	0.3709	.8307
Superior frontal gyrus	25 (100)	4 (100)	4 (80)	5.976	.0504
Paracentral lobe	21 (84)	3 (75)	4 (80)	0.2145	.8983
Precuneus	21 (84)	4 (100)	5 (100)	1.632	.4422
Occipital gyrus	24 (96)	2 (50)	3 (60)	8.807	.0122*
Parietal superior lobe	19 (76)	2 (50)	1 (20)	6.151	.0462*
Angular/supramarginal lobe	16 (64)	3 (75)	3 (60)	7.595	.0224*
Medial frontal gyrus	23 (92)	3 (75)	3 (60)	3.785	.1507
Cuneus	23 (92)	3 (75)	2 (40)	7.921	.0191*
Inferior frontal gyrus	16 (64)	3 (75)	0 (0)	7.595	.0224*
Medial temporal gyrus	11 (44)	2 (50)	0 (0)	3.681	.1587
Precentral gyrus	18 (72)	2 (50)	1 (20)	5.036	.0806
Postcentral gyrus	8 (32)	2 (50)	1 (20)	0.9192	.6315
Superior temporal gyrus	8 (32)	2 (50)	1 (20)	0.9192	.6315

Note:—MM indicates homozygous methionine; VV, homozygous valine; MV heterozygous methionine and valine.

* P < .05. Correction for multiple analyses: results are not significant.