

Supplementary Tables

A. Logistic regression for variables affecting presence of psychotic features in all types of prion disease (in 279 patients)

Independent variables		Total n	With psychotic features		Logistic Regression		
			n	%	Odds Ratio	95% CI	p value
Diagnosis	sCJD	170	70	41.18%	1	(ref)	0.001
	vCJD	25	12	48.00%	0.654	0.175 - 2.443	0.528
	iCJD	7	1	14.29%	0.1	0.01 - 0.955	0.045
	IPD OPRI	29	7	24.14%	0.239	0.075 - 0.767	0.016
	IPD Point	48	9	18.75%	0.189	0.075 - 0.476	<0.001
Age at Onset	Per year				0.979	0.953 - 1.006	0.125
PRNP Codon 129	MM	159	54	33.96%	1	(ref)	0.008
	MV	83	37	44.58%	1.959	1.064 - 3.607	0.031
	VV	37	8	21.62%	0.488	0.201 - 1.181	0.112
Gender	Male	134	50	37.31%	1	(ref)	
	Female	145	49	33.79%	0.944	0.556 - 1.601	0.831
Constant					2.646		0.303

B. Logistic regression for variables affecting presence of mood disorder in all types of prion disease (in 279 patients)

Independent variables		Total n	With mood disorder		Logistic Regression		
			n	%	Odds Ratio	95% CI	p value
Diagnosis	sCJD	170	57	33.53%	1	ref	0.372
	vCJD	25	17	68.00%	3.044	0.838 - 11.054	0.091
	iCJD	7	4	57.14%	1.689	0.326 - 8.741	0.532
	IPD OPRI	29	15	51.72%	1.555	0.559 - 4.327	0.397
	IPD Point	48	28	58.33%	1.96	0.917 - 4.192	0.083
Age at Onset	Per year				0.984	0.96 - 1.009	0.204
PRNP Codon 129	MM	159	67	42.14%	1	ref	0.11
	MV	83	43	51.81%	1.684	0.933 - 3.037	0.083
	VV	37	11	29.73%	0.76	0.331 - 1.745	0.517
Gender	Male	134	53	39.55%	1	(ref)	
	Female	145	68	46.90%	1.68	1.002 - 2.818	0.049
Constant					0.955		0.958

C. Logistic regression for variables affecting presence of agitated features in all types of prion disease (in 279 patients)

Independent variables		Total	With agitated features		Logistic Regression		
		n	n	%	Odds Ratio	95% CI	p value
Diagnosis	sCJD	170	28	16.47%	1	ref	0.657
	vCJD	25	8	32.00%	0.615	0.135 - 2.81	0.531
	iCJD	7	0	0.00%	<0.001	0 - ???	0.999
	IPD OPRI	29	10	34.48%	1.067	0.322 - 3.531	0.916
	IPD Point	48	7	14.58%	0.511	0.18 - 1.45	0.207
Age at Onset	Per year				0.966	0.936 - 0.997	0.03
PRNP Codon 129	MM	159	35	22.01%	1	ref	0.699
	MV	83	13	15.66%	0.763	0.359 - 1.618	0.48
	VV	37	5	13.51%	0.711	0.245 - 2.068	0.531
Gender	Male	134	27	20.15%			
	Female	145	26	17.93%	0.961	0.511 - 1.809	0.902
Constant					2.172		0.476

D. Logistic regression for variables affecting presence of BPS at onset in all types of prion disease (in 279 patients)

Independent variables		Total	With BPS at onset		Logistic Regression		
		n	n	%	Odds Ratio	95% CI	p value
Diagnosis	sCJD	170	74	43.53%	1	(ref)	<0.001
	vCJD	25	23	92.00%	9.682	1.687 - 55.584	0.011
	iCJD	7	2	28.57%	0.359	0.06 - 2.159	0.263
	IPD OPRI	29	16	55.17%	1.105	0.396 - 3.081	0.848
	IPD Point	48	11	22.92%	0.297	0.128 - 0.692	0.005
Age at Onset	Per year				0.991	0.966 - 1.017	0.493
PRNP Codon 129	MM	159	80	50.31%	1	(ref)	0.071
	MV	83	37	44.58%	1.205	0.667 - 2.176	0.537
	VV	37	9	24.32%	0.425	0.182 - 0.990	0.047
Gender	Male	134	66	49.25%	1	(ref)	
	Female	145	60	41.38%	0.907	0.54 - 1.525	0.714
Constant					1.614		0.601

E. Logistic regression for variables affecting presence of BPS at onset in sCJD (in 103 patients)

Independent variables		Total		With BPS at onset		Logistic Regression		
		n		n	%	Odds Ratio	95% CI	p value
Age at Onset	Per year					0.935	0.889 - 0.984	0.01
PRNP Codon 129	MM	46		19	41.30%	1	(ref)	0.333
	MV	33		17	51.52%	1.197	0.447 - 3.210	0.72
	VV	24		6	25.00%	0.464	0.135 - 1.594	0.223
Gender	Male	46		20	43.48%	1	(ref)	
	Female	57		22	38.60%	0.945	0.387 - 2.311	0.902
MRI	Striatum	Abnormal	79	32	40.51%	0.826	0.204 - 3.352	0.789
		Normal	24	10	41.67%			
	Thalamus	Abnormal	60	24	40.00%	0.813	0.266 - 2.484	0.717
		Normal	43	18	41.86%			
	Cortex	Abnormal	83	36	43.37%	1.205	0.342 - 4.242	0.771
		Normal	20	6	30.00%			
Constant						65.816		0.033

F. Logistic regression for variables affecting presence of psychotic features in sCJD (in 103 patients)

Independent variables		Total		With psychotic features		Logistic Regression		
		n		n	%	Odds Ratio	95% CI	p value
Age at Onset	Per year					0.962	0.917 - 1.009	0.113
PRNP Codon 129	MM	46		19	41.30%	1	(ref)	0.048
	MV	33		20	60.61%	2.031	0.759 - 5.436	0.158
	VV	24		6	25.00%	0.411	0.121 - 1.387	0.152
Gender	Male	46		19	41.30%	1	(ref)	
	Female	57		26	45.61%	1.632	0.665 - 4.003	0.285
MRI	Striatum	Abnormal	79	35	44.30%	1.257	0.318 - 4.974	0.744
		Normal	24	10	41.67%			
	Thalamus	Abnormal	60	27	45.00%	0.816	0.271 - 2.456	0.717
		Normal	43	18	41.86%			
	Cortex	Abnormal	83	38	45.78%	0.961	0.281 - 3.284	0.95
		Normal	20	7	35.00%			
Constant								

G. Logistic regression for variables affecting presence of mood disorder in sCJD (in 103 patients)

Independent variables		Total		With BPS at onset		Logistic Regression		
		n		n	%	Odds Ratio	95% CI	p value
Age at Onset	Per year					0.961	0.914 - 1.01	0.116
PRNP Codon 129	MM	46		14	30.43%	1	(ref)	0.599
	MV	33		12	36.36%	1.522	0.538 - 4.304	0.428
	VV	24		7	29.17%	0.824	0.239 - 2.845	0.76
Gender	Male	46		12	26.09%	1	(ref)	
	Female	57		21	0.36842105	2.024	0.784 - 5.226	0.145
MRI	Striatum	Abnormal	79	23	29.11%	1.169	0.292 - 4.674	0.826
		Normal	24	10	41.67%			
	Thalamus	Abnormal	60	16	26.67%	0.41	0.128 - 1.309	0.132
		Normal	43	17	39.53%			
Cortex	Abnormal	83	25	30.12%	0.547	0.159 - 1.874	0.337	
	Normal	20	8	0.4				
Constant						8.89		0.257

H. Logistic regression for variables affecting presence of agitated features in sCJD (in 103 patients)

Independent variables		Total		With BPS at onset		Logistic Regression		
		n		n	%	Odds Ratio	95% CI	p value
Age at Onset	Per year					0.969	0.913 - 1.029	0.304
PRNP Codon 129	MM	46		5	10.87%	1	(ref)	0.179
	MV	33		9	27.27%	3.327	0.92 - 12.038	0.067
	VV	24		4	16.67%	1.585	0.34 - 7.386	0.558
Gender	Male	46		7	15.22%	1	(ref)	
	Female	57		11	0.19298246	1.812	0.575 - 5.709	0.31
MRI	Striatum	Abnormal	79	14	17.72%	1.452	0.259 - 8.14	0.671
		Normal	24	4	16.67%			
	Thalamus	Abnormal	60	10	16.67%	0.5	0.124 - 2.023	0.331
		Normal	43	8	18.60%			
Cortex	Abnormal	83	15	18.07%	1.049	0.224 - 4.921	0.951	
	Normal	20	3	0.15				
Constant						0.675		0.863

I. Logistic regression of MRC-PDS score and Disease Type vs Hallucinations

		Logistic Regression		
		Odds Ratio	95% CI	p value
MRC Score	Per point	0.935	0.909 - 0.963	<0.001
Diagnosis	sCJD	1	(ref)	<0.001
	vCJD	0.327	0.122 - 0.874	0.026
	iCJD	1.334	0.461 - 3.864	0.595
	IPD OPRI	0.083	0.025 - 0.279	<0.001
	IPD Point	0.254	0.121 - 0.531	<0.001
	Constant	0.783		0.171

J. Logistic regression of MRC-PDS score and Disease Type vs Depressive Symptoms

		Logistic Regression		
		Odds Ratio	95% CI	p value
MRC Score	Per point	0.981	0.961 - 1.002	0.078
Diagnosis	sCJD	1	(ref)	0.328
	vCJD	1.31	0.661 - 2.597	0.439
	iCJD	0.601	0.167 - 2.16	0.435
	IPD OPRI	0.644	0.37 - 1.12	0.119
	IPD Point	0.858	0.497 - 1.482	0.583
	Constant	0.412		<0.001