Table 1. **Clinical and demographic features of human prionopathies in the published literature. A sensitivity analysis after removing missing data from cognitive behavioral disorder**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
	N(%)	N(%)	N(%)	N(%)	N(%)	N(%)	
N	**133**	**22**	**12**	**43**	**28**	**45**	
Age at onset (y), median (IQR)	62 (53, 69)	36 (26, 41)	45.5 (37, 52.5)	61 (46, 69)	45 (30, 53.5)	45 (37, 56)	<0.001
Disease duration (m), median (IQR)	8 (3, 17)	16.5 (10, 18)	9.5 (6.5, 12)	5 (4, 16)	11 (8.5, 13)	60 (15, 96)	<0.001
Cognitive/Behavioral							0.002
Onset	95 (71.4%)	18 (81.8%)	8 (66.7%)	34 (79.1%)	10 (35.7%)	29 (64.4%)	
Later in course	38 (28.6%)	4 (18.2%)	4 (33.3%)	9 (20.9%)	18 (64.3%)	16 (35.6%)	
Not reported	0 (0%)	0	0 (0%)	0 (0%)	0 (0%)	0 (0%)	
Movement Disorders							0.03
Onset	60 (45.1%)	5 (22.7%)	8 (66.7%)	12 (27.9%)	8 (28.6%)	25 (55.6%)	
Later in course	71 (53.4%)	17 (77.3%)	4 (33.3%)	30 (69.8%)	20 (71.4%)	18 (40%)	
Not reported	2 (1.5%)	0 (0%)	0 (0%)	1 (2.3%)	0 (0%)	2 (4.4%)	
Dysautonomia							<0.001
Onset	0 (0%)	0 (0%)	0 (0%)	0 (0%)	5 (17.9%)	0 (0%)	
Later in course	1 (0.8%)	0 (0%)	1 (8.3%)	7 (16.3%)	15 (53.6%)	0 (0%)	
Not reported	132 (99.2%)	22 (100%)	11 (91.7%)	36 (83.7%)	8 (28.6%)	45 (100%)	
Sleep disorder							<0.001
Onset	2 (1.5%)	0 (0%)	1 (8.3%)	7 (16.3%)	19 (67.9%)	0 (0%)	
Later in course	0 (0%)	0 (0%)	0 (0%)	5 (11.6%)	7 (25%)	0 (0%)	
Not reported	131 (98.5%)	22 (100%)	11 (91.7%)	31 (72.1%)	2 (7.1%)	45 (100%)	

IQR: Interquartile range; *sCJD: Sporadic Creutzfeldt-Jakob disease; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease;* p-value: Wilcoxon rank sum test for continuous data and Chi square test for categorical data

Table 2. **Prevalence of movement disorders in human prionopathies. A sensitivity analysis after removing missing data from cognitive behavioral disorder**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
	N(%)	N(%)	N(%)	N(%)	N(%)	N(%)	
N	**133**	**22**	**12**	**43**	**28**	**45**	
Gait ataxia	82 (61.7%)	20 (90.9%)	9 (75%)	28 (65.1%)	15 (53.6%)	32 (71.1%)	0.058
Limb ataxia	50 (37.6%)	9 (40.9%)	7 (58.3%)	22 (51.2%)	7 (25%)	9 (20%)	0.017
Myoclonus	79 (59.4%)	13 (59.1%)	8 (66.7%)	32 (74.4%)	19 (67.9%)	12 (26.7%)	<0.001
Tremor	6 (4.5%)	1 (4.5%)	1 (8.3%)	3 (7%)	4 (14.3%)	4 (8.9%)	0.41
Parkinsonism	9 (6.8%)	2 (9.1%)	1 (8.3%)	7 (16.3%)	4 (14.3%)	9 (20%)	0.14
Rigidity	57 (42.9%)	7 (31.8%)	3 (25%)	25 (58.1%)	6 (21.4%)	19 (42.2%)	0.037
Chorea	5 (3.8%)	7 (31.8%)	0 (0%)	0 (0%)	0 (0%)	2 (4.4%)	<0.001
Dystonia	10 (7.5%)	2 (9.1%)	0 (0%)	1 (2.3%)	0 (0%)	2 (4.4%)	0.52
Gaze palsy	13 (9.8%)	3 (13.6%)	1 (8.3%)	1 (2.3%)	0 (0%)	0 (0%)	0.032

sCJD: Sporadic Creutzfeldt-Jakob disease; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease; p-value: Fisher's exact test

Table 3. **Time (months) from movement disorder onset until death. A sensitivity analysis after removing missing data from cognitive behavioral disorder**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
Gait ataxia, median (IQR)	6 (2, 12)	8 (5.5, 17.5)	7 (6, 8)	4 (3.5, 9)	7 (4, 12)	52.5 (10.5,78)	<0.001
Limb ataxia, median (IQR)	4 (2, 12)	7 (6, 18)	6 (4, 13)	4.5 (3, 9)	5 (3, 17)	60 (48, 72)	0.01
Myoclonus, median (IQR)	2 (1, 5)	6 (3, 12)	4.5 (2, 9)	4 (2, 10)	5 (3, 8)	5.5 (2.5, 71)	0.003
Tremor, median (IQR)	3 (1, 8)	35 (35, 35)	10 (10, 10)	4 (4, 60)	9.5 (5, 13)	31.5 (2, 84)	0.59
Parkinsonism, median (IQR)	3 (2, 4)	4.5 (1, 8)	10 (10, 10)	18 (16, 90)	8.5 (5.5,18)	36 (24, 65)	0.007
Rigidity, median (IQR)	5 (3, 14)	17 (7, 18)	13 (8, 13)	5 (4, 16)	7.5 (5, 11)	36 (7, 96)	<0.001
Chorea, median (IQR)	10 (4, 18)	3 (2, 8)	-	-	-	2.5 (2, 3)	0.22
Dystonia, median (IQR)	4 (2, 14)	14.5 (11, 18)	-	3 (3, 3)	-	60.5 (1, 120)	0.66
Gaze palsy, median (IQR)	11 (7, 14)	8 (7, 14)	13 (13, 13)	16 (16, 16)	-	-	0.69

IQR: Interquartile range; *sCJD: Sporadic CJD; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease;* p-value: Wilcoxon rank sum test

Table 4. Clinical and demographic features of human prionopathies in the published literature. A restricted analysis of studies published in neurological journals

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
	N(%)	N(%)	N(%)	N(%)	N(%)	N(%)	
N	116	12	11	38	22	44	
Age at onset (y), median (IQR)	62 (54, 69)	26.5 (21, 37.5)	44 (35, 53)	62 (54, 69)	38.5 (25, 53)	48 (36.5, 57)	<0.001
Disease duration (m), median (IQR)	7 (4, 18)	15.5 (7.5, 19.5)	10 (6, 13)	5 (4, 12)	12 (9, 14)	58.5 (22.5, 102)	<0.001
Cognitive/Behavioral							0.016
Onset	65 (56%)	10 (83.3%)	7 (63.6%)	29 (76.3%)	7 (31.8%)	27 (61.4%)	
Later in course	32 (27.6%)	2 (16.7%)	4 (36.4%)	7 (18.4%)	13 (59.1%)	12 (27.3%)	
Not reported	19 (16.4%)	0 (0%)	0 (0%)	2 (5.3%)	2 (9.1%)	5 (11.4%)	
Movement Disorders							0.047
Onset	66 (56.9%)	3 (25%)	7 (63.6%)	12 (31.6%)	8 (36.4%)	27 (61.4%)	
Later in course	48 (41.4%)	9 (75%)	4 (36.4%)	25 (65.8%)	14 (63.6%)	15 (34.1%)	
Not reported	2 (1.7%)	0 (0%)	0 (0%)	1 (2.6%)	0 (0%)	2 (4.5%)	
Dysautonomia							<0.001
Onset	0 (0%)	0 (0%)	0 (0%)	0 (0%)	2 (9.1%)	0 (0%)	
Later in course	1 (0.9%)	0 (0%)	1 (9.1%)	5 (13.2%)	13 (59.1%)	0 (0%)	
Not reported	115 (99.1%)	12 (100%)	10 (90.9%)	33 (86.8%)	7 (31.8%)	44 (100%)	
Sleep disorder							<0.001
Onset	1 (0.9%)	0 (0%)	1 (9.1%)	6 (15.8%)	13 (59.1%)	0 (0%)	
Later in course	1 (0.9%)	0 (0%)	0 (0%)	1 (2.6%)	7 (31.8%)	0 (0%)	
Not reported	114 (98.3%)	12 (100%)	10 (90.9%)	31 (81.6%)	2 (9.1%)	44 (100%)	

IQR: Interquartile range; *sCJD: Sporadic Creutzfeldt-Jakob disease; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease;* p-value: Wilcoxon rank sum test for continuous data and Chi square test for categorical data

Table 5. Prevalence of movement disorders in human prionopathies. A restricted analysis of studies published in neurological journals

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
	N(%)	N(%)	N(%)	N(%)	N(%)	N(%)	
N	116	12	11	38	22	44	
Gait ataxia	76 (65.5%)	10 (83.3%)	8 (72.7%)	27 (71.1%)	14 (63.6%)	31 (70.5%)	0.85
Limb ataxia	48 (41.4%)	2 (16.7%)	7 (63.6%)	21 (55.3%)	6 (27.3%)	10 (22.7%)	0.006
Myoclonus	67 (57.8%)	6 (50%)	8 (72.7%)	27 (71.1%)	15 (68.2%)	11 (25%)	<0.001
Tremor	5 (4.3%)	1 (8.3%)	1 (9.1%)	3 (7.9%)	4 (18.2%)	4 (9.1%)	0.22
Parkinsonism	10 (8.6%)	2 (16.7%)	1 (9.1%)	5 (13.2%)	1 (4.5%)	9 (20.5%)	0.29
Rigidity	48 (41.4%)	5 (41.7%)	3 (27.3%)	21 (55.3%)	7 (31.8%)	18 (40.9%)	0.47
Chorea	3 (2.6%)	3 (25%)	0 (0%)	0 (0%)	0 (0%)	2 (4.5%)	0.024
Dystonia	12 (10.3%)	2 (16.7%)	0 (0%)	1 (2.6%)	0 (0%)	2 (4.5%)	0.22
Gaze palsy	16 (13.8%)	2 (16.7%)	1 (9.1%)	1 (2.6%)	0 (0%)	0 (0%)	0.009

sCJD: Sporadic Creutzfeldt-Jakob disease; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease; p-value: Fisher's exact test

Table 6. **Time (months) from movement disorder onset until death. A restricted analysis of studies published in neurological journals**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
Gait ataxia, median (IQR)	5.5 (2, 12.5)	8 (7, 17)	7 (5, 10.5)	4 (3, 9)	10 (5, 12)	56 (9, 84)	<0.001
Limb ataxia, median (IQR)	4 (2, 12)	19 (7, 31)	6 (4, 13)	5 (3, 9)	11.5 (5, 20)	58.5 (48, 72)	0.002
Myoclonus, median (IQR)	2 (2, 6)	3.5 (2, 11)	4.5 (2, 9)	4 (2, 8)	5 (2, 9)	5 (2, 88)	0.054
Tremor, median (IQR)	3 (1, 3)	35 (35,35)	10 (10, 10)	4 (4, 60)	9.5 (5, 13)	31.5 (2, 84)	0.51
Parkinsonism, median (IQR)	3.5 (3, 20)	4.5 (1, 8)	10 (10, 10)	60 (16, 90)	27 (27, 27)	36 (24, 65)	0.024
Rigidity, median (IQR)	5.5 (3, 16.5)	17 (14, 18)	13 (8, 13)	5 (3, 12)	9 (5, 17)	42 (7, 108)	<0.001
Chorea, median (IQR)	4 (2, 48)	2 (2, 8)	-	-	-	2.5 (2, 3)	0.63
Dystonia, median (IQR)	4 (2, 10)	14.5 (11, 18)	-	3 (3, 3)	-	60.5 (1, 120)	0.58
Gaze palsy, median (IQR)	10 (3.5, 14)	11 (8, 14)	13 (13, 13)	16 (16, 16)	-	-	0.64

IQR: Interquartile range; *sCJD: Sporadic CJD; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease;* p-value: Wilcoxon rank sum test

Table 7. **Clinical and demographic features of human prionopathies in the published literature. A sensitivity analysis of studies published in neurological journals after removing missing data from cognitive behavioral disorder**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
	N(%)	N(%)	N(%)	N(%)	N(%)	N(%)	
N	**97**	**12**	**11**	**36**	**20**	**39**	
Age at onset (y), median (IQR)	62 (54, 68)	26.5 (21, 37.5)	44 (35, 53)	62 (54, 69.5)	38.5 (25, 53.5)	45 (36, 56)	<0.001
Disease duration (m), median (IQR)	8 (4, 18)	15.5 (7.5, 19.5)	10 (6, 13)	5 (3.5, 12)	12 (8.5, 13.5)	60 (15, 108)	<0.001
Cognitive/Behavioral							0.016
Onset	65 (67%)	10 (83%)	7 (64%)	29 (81%)	7 (35%)	27 (69%)	
Later in course	32 (33%)	2 (17%)	4 (36%)	7 (19%)	13 (65%)	12 (31%)	
Not reported	0 (0%)	0	0 (0%)	0 (0%)	0 (0%)	0 (0%)	
Movement Disorders							0.11
Onset	48 (49%)	3 (25%)	7 (64%)	10 (28%)	6 (30%)	22 (56%)	
Later in course	47 (48%)	9 (75%)	4 (36%)	25 (69%)	14 (70%)	15 (38%)	
Not reported	2 (2%)	0 (0%)	0 (0%)	1 (3%)	0 (0%)	2 (5%)	
Dysautonomia							<0.001
Onset	0 (0%)	0 (0%)	0 (0%)	0 (0%)	2 (10%)	0 (0%)	
Later in course	1 (1%)	0 (0%)	1 (9%)	4 (11%)	12 (60%)	0 (0%)	
Not reported	96 (99%)	12 (100%)	10 (91%)	32 (89%)	6 (30%)	39 (100%)	
Sleep disorder							<0.001
Onset	1 (1%)	0 (0%)	1 (9%)	6 (16%)	13 (65%)	0 (0%)	
Later in course	0 (0%)	0 (0%)	0 (0%)	1 (3%)	6 (30%)	0 (0%)	
Not reported	96 (99%)	12 (100%)	10 (91%)	29 (81%)	1 (5%)	39 (100%)	

IQR: Interquartile range; *sCJD: Sporadic Creutzfeldt-Jakob disease; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease;* p-value: Wilcoxon rank sum test for continuous data and Chi square test for categorical data

Table 8. **Prevalence of movement disorders in human prionopathies. A sensitivity analysis of studies published in neurological journals after removing missing data from cognitive behavioral disorder**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
	N(%)	N(%)	N(%)	N(%)	N(%)	N(%)	
N	**97**	**12**	**11**	**36**	**20**	**39**	
Gait ataxia	62 (64%)	10 (83%)	8 (73%)	25 (69%)	12 (60%)	26 (67%)	0.81
Limb ataxia	35 (36%)	2 (17%)	7 (64%)	19 (53%)	5 (25%)	9 (23%)	0.018
Myoclonus	58 (60%)	6 (50%)	8 (73%)	27 (75%)	14 (70%)	11 (28%)	<0.001
Tremor	5 (5%)	1 (8%)	1 (9%)	3 (8%)	4 (20%)	4 (10%)	0.3
Parkinsonism	7 (7%)	2 (17%)	1 (9%)	5 (14%)	1 (5%)	9 (23%)	0.24
Rigidity	40 (41%)	5 (42%)	3 (27%)	20 (56%)	6 (30%)	17 (44%)	0.44
Chorea	2 (2%)	3 (25%)	0 (0%)	0 (0%)	0 (0%)	2 (5%)	0.02
Dystonia	9 (9%)	2 (17%)	0 (0%)	1 (3%)	0 (0%)	2 (5%)	0.34
Gaze palsy	12 (12%)	2 (17%)	1 (9%)	1 (3%)	0 (0%)	0 (0%)	0.032

sCJD: Sporadic Creutzfeldt-Jakob disease; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease; p-value: Fisher's exact test

Table 9. **Time (months) from movement disorder onset until death. A sensitivity analysis of studies published in neurological journals after removing missing data from cognitive behavioral disorder**

	sCJD	vCJD	iCJD	gCJD	FFI	GSS	p-value
Gait ataxia, median (IQR)	6.5 (2, 13)	8 (7, 17)	7 (5,10.5)	4 (3, 8)	9 (5, 12)	52.5 (6, 72)	<0.001
Limb ataxia, median (IQR)	4 (2, 12)	19 (7, 31)	6 (4, 13)	4 (3, 7)	6 (5, 17)	60 (48, 72)	0.011
Myoclonus, median (IQR)	2 (2, 6)	3.5 (2, 11)	4.5 (2, 9)	4 (2, 8)	5 (2, 9)	5 (2, 88)	0.13
Tremor, median (IQR)	3 (1, 3)	35 (35, 35)	10 (10, 10)	4 (4, 60)	9.5 (5, 13)	31.5 (2, 84)	0.51
Parkinsonism, median (IQR)	3 (3, 20)	4.5 (1, 8)	10 (10,10)	60 (16,90)	27 (27, 27)	36 (24, 65)	0.028
Rigidity, median (IQR)	6 (3, 20)	17 (14, 18)	13 (8, 13)	4.5 (2.5,14)	7.5 (5, 11)	36 (7, 96)	0.004
Chorea, median (IQR)	10 (4, 18)	3 (2, 8)	-	-	-	2.5 (2, 3)	0.22
Dystonia, median (IQR)	4 (2,14)	14.5 (11, 18)	-	3 (3, 3)	-	60.5 (1, 120)	0.66
Gaze palsy, median (IQR)	11 (7,14)	8 (7, 14)	13 (13,13)	16 (16, 16)	-	-	0.69

IQR: Interquartile range; *sCJD: Sporadic CJD; vCJD: Variant Creutzfeldt-Jakob disease; iCJD: Iatrogenic Creutzfeldt-Jakob disease; gCJD: Genetic Creutzfeldt-Jakob disease; FFI: Fatal familial insomnia; GSS: Gerstmann-Sträussler-Scheinker disease;* p-value: Wilcoxon rank sum test