

TABLE E-1

	Stage 1	Stage 2	Stage 3	Stage 4
Name	Vague symptomatic state	Possible CJD state	Probable CJD state	Chronic vegetative state
Clinical characteristics	Unrelated vague symptoms only	Specific findings that do not satisfy the clinical criteria for probable CJD	Satisfy clinical criteria for probable CJD	Full dementia; no spontaneous purposeful movement; no verbal output
Patients included in each stage	31	32	34	33
Median duration (IQR)	1 month (4 m)	1 month (1.2 m)	1 month (1.5 m)	Not available (>2 weeks)*
DWI, % (n)	100 (6/6)	100 (18/18)	100 (25/25)	100 (13/13)
EEG, % (n)	0 (0/1)	25 (2/8)	53.8 (14/26)	73.7 (14/19)
CSF 14-3-3 protein, % (n)	0 (0/1)	84.6 (11/13)	90.9 (20/22)	75 (3/4)

IQR = Interquartile range.

* The median duration could not be determined because there were several patients still living.

Schema of sCJD clinical stages, with the duration of each stage and the results of diagnostic tests according to the clinical stage