

Table 2  
Characteristics of the included studies for meta-analysis

Study	Ethnicity	Country	Age at onset (years)	Case age/Control age (years)	Genotyping method	Cases(familial /sporadic)	Controls(familial /sporadic)	Cases carry C9 repeat expansions(familial /sporadic)	Cases carry C9 intermediate repeats(familial /sporadic)	Controls carry C9 repeat expansions(familial /sporadic)	Controls carry C9 intermediate repeats(familia l/sporadic)	Diagnostic criteria	NOS score
Susan et al. (2012)	Caucasian	Ireland			rpPCR, Southern blot	435 (49/386)	188	39 (20/19)		0		El Escorial	7
Beck et al. (2013)					rpPCR, Southern blot	360	7579	29		11			4
Chio et al. (2012)	Caucasian	Italy			rpPCR	475 (36/439)	245	32 (24/8)		0		El Escorial	7
Debray et al. (2013)	Caucasian	Belgium			rpPCR	590 (119/471)	384	77 (32/45)		0		revised El Escorial	5
Dombroski et al. (2013)	others	Guam			rpPCR	24	43	1		0		El Escorial	7
Garcia-Redondo et al. (2013)	Caucasian	Spain			rpPCR	936 (155/781)	248	67 (42/25)	0	0	0	El Escorial	8
Gijselinck et al. (2012)	Caucasian	Flanders		59.9±11.6/65.3± 14.8	rpPCR	137 (15/122)	856	13 (7/6)		0		revised El Escorial	8
Harms et al. (2013)	Caucasian	US			rpPCR, Southern	51/797	526	77 (22/55)		2		El Escorial	8

Table 1. Characteristics of the studies included in the meta-analysis									
Study	Population	Country	Age (years)	Gender	Method	Number of cases	Number of controls	Number of deaths	Number of survivors
Jiao et al. (2014)	Asian	China	41.1±11.5/39.7±8.9	blot	rpPCR	110 (10/100)	150	1 (1/0)	0
Kenna et al. (2013)	Caucasian	Irish	61.7±12.0	/60.3±11.8	high throughput sequencing	444 (50/394)	311	39 (15/24)	0
Konno et al. (2013)	Asian	Japanese			rpPCR	168 (58/110)	180	2 (2/0)	0
Millecamps et al. (2012)	Caucasian	French			rpPCR	950 (225/725)	580	162 (104/58)	0
Ogaki et al. (2012)	Asian	Japanese	60.4±11.7	/60.6±10.3	rpPCR	563 (11/552)	197	2 (0/2)	0
Ratti et al. (2012)	Caucasian	Italy	57.1±12.7		rpPCR	1534 (259/1275)	862	128 (62/66)	2
Sabatelli et al. (2012)	Caucasian	Italy			rpPCR	/1757	619	69 (0/69)	0
Tsai et al. (2012)	Asian	China			rpPCR	124 (22/102)	300	6 (4/2)	0
van Rheenen et al. (2012)	Caucasian	Netherlands	/63.1±11.8		rpPCR	1500 (78/1422)	768	120 (33/87)	0
Xi et al. (2012)	Caucasian	or North American countries	57.6±12.3	/70.2±9.5	rpPCR	389 (47/342)	602	36 (18/18)	0

Borghero et al. (2014)	Caucasian	Italy	61.1±12.1/	rpPCR	375 (100/275)	700	51 (33/18)	0	revised El Escorial	8
Brettschneider et al. (2012)	Caucasian	America	55–69/60–78	rpPCR	75 (13/62)	11	14 (6/8)	0	revised El Escorial	7
Cooper–Knock et al. (2012)	Caucasian	Northern England		rpPCR	563 (63/500)	361	62 (27/35)	0	El Escorial	8
Laere et al. (2014)	Caucasian	Belgium	60.4±12.6	rpPCR	70	20	11	0	revised El Escorial	8
Rutherford et al. (2012)	Caucasian	America, British		rpPCR	995	1444	211	0	El Escorial	8
Zou et al. (2013)	Asian	China		rpPCR	344 (20/324)	245	0	0	El Escorial	8