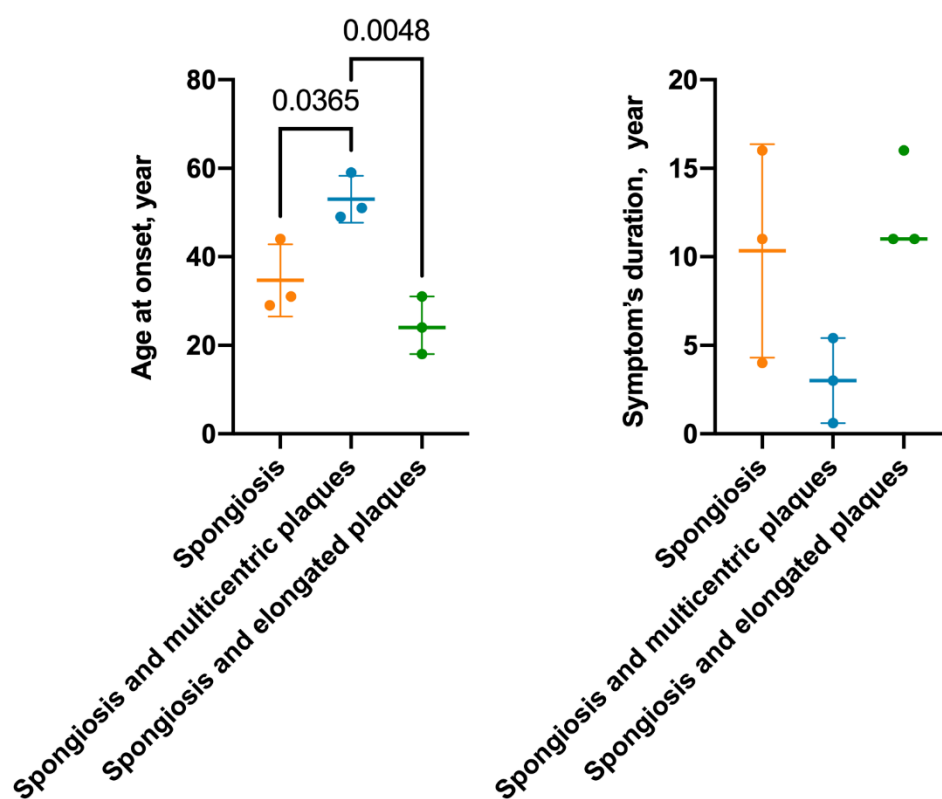


## Supplementary Materials



**Figure S1.** Comparison of age at onset and duration of symptoms in patient groups demarcated by pathological changes.

**Table S1.** Clinical and auxiliary characteristics of all known 17 patients carrying 7-OPRI.

Number	Year	Country	Gender	Family History	Age at Onset (Years)	Duration (Years)	First Symptom of Disease	Cognitive Dysfunction	Psychiatric Disturbances	Parkinsonism	Cerebellar Signs	Myoclonus	Pyramidal Signs	Speech Disorders	References
1	1991	USA	F	+	31	11	Psychiatric disturbances	+	+ (Mood change, indifference, confusion)	+	+		+	+	[8,10]
2			F	+	23	10	Psychiatric disturbances	+	+ (Abnormal behavior)	+	+	+	+	+	
3			F	+	28	13	Ataxia (abnormal gait)	+	+ (Euphoria)	+	+	+	+	+	
4	1992	Japan	NA	+	NA	7	Cognitive dysfunction	+		+					[9]
5	2000	Belgium	M	+	32	7	Cognitive dysfunction	+		+					[11]
6			M	+	24	11	Cognitive dysfunction	+	+ (Psychosis)	+	+	+			
7			F	+	31	11	Ataxia	+	+ (Depression/ mood change)		+		+		
8	2003	Australia	NA	+	29	16	Psychiatric disturbance and	+	+ (Abnormal behavior)	+					[12]

							cognitive dysfunctio n									
9	200 7	Italy	M	-	18	16	Cognitive dysfunctio n and psychiatric disturbance	+	+	(Depression, psychosis, bipolar disorder)		+			[13,14,17]	
10	200 8	China	F	+	44	4.2	Cognitive dysfunctio n	+				+	+	+	[15,16]	
11	201 1	Netherland s	F	+	50	0.7	Cognitive dysfunctio n	+		+	(Emotional lability, anxiety attacks)			+	+	[7]
12			M	+	49	3	Cognitive dysfunctio n	+		+	(Emotional lability, apathetic)	+				
13			M	+	59	0.6	Ataxia	+		+	(Apathetic)	+	+	+		
14			M	+	51	5.4	Ataxia, cognitive dysfunctio n and psychiatric disturbance	+		+	(Depression, panic attacks)	+	+	+		
15	202 2	China	NA	+	40s	>0.6	Cognitive dysfunctio n	+		+	(Mood change, abnormal behavior)	+	+	+	+	Our case

16	NA	+	60s	5.5	Cognitive dysfunction	+	+ (Mood change, abnormal behavior)	+	+	+	+	Our case
17	NA	+	50s	1.5	Cognitive dysfunction	+			+	+		Our case

**Table S1.** Continued.

Number	Visual Signs	Mutism	Seizure	PSWCs on EEG	CSF 14-3-3 Protein	CSF Tau Protein	CSF RT-QuIC	Hyperintensity on MRI	Condon 129	Neuropathology	Immunoblot	Mutation	Misdiagnose	References
1			+	- (Diffuse slowing)					Cis-M	Spongiosis, gliosis, neuronal loss		R2c-R3-R2-R3-R2-R3g-R2-R3g	Schizophrenia	[8,10]
2		+		- (Diffuse slowing)					Cis-M	Mild gliosis and neuronal loss (no spongiosis)				
3		+		- (Paroxysmal episodes of slow wave spikes)					Cis-M					
4									NA	Kuru-like plaques in the cerebellum		R3-R2-R2-R2-R3g-R2-R2		[9]
5									Cis-M			NA	Multiple sclerosis	[11]
6										Spongiosis, gliosis, neuronal loss, elongated				

									cerebellar plaques cerebellar cortex										
7		+							Cis- M(MM)	Spongiosis, gliosis, neuronal loss, elongated cerebellar plaques cerebellar cortex									
8		+	- (Diffuse slowing)						Cis- M(MV)	Spongiosis, gliosis, neuronal loss, non-linear granular deposits cerebellar cortex	Type 1	R3-R2-R3- R2-R2-R2- R2	Huntington's disease	[12]					
9	+		- (Diffuse slowing)	-	-	- (MRI revealed widespread cortical brain atrophy)			Cis- M(MM)	spongiform degeneration, cortical PrP plaques, and elongated PrP formations in the cerebellum.	type 1 in the cerebral cortex and mixed types 1 and 2 in the cerebellum	R3-R2-R2- R3g-R2-R2- R2	Psychosis and bipolar disorder	[13,14,17]					
10	+		+			- (no remarkable changes)			Cis- M(MM)	Spongiosis, gliosis, neuronal loss	Type 1	R2-R2-R2- R3g-R2-R3g- R2a		[15,16]					
11									NA			NA		[7]					
12			- (Non- specific changes)						NA	Spongiosis +++, gliosis, neuronal loss, multicentric plaques	Type 1	NA							

