

**Supplemental Table:** Clinical, genetic and imaging findings of 30 FXTAS cases with functional neuroimaging reported in the literature.

	Clinical findings		CGG repeats	MCP sign	SPECT	
	Symptoms	L-Dopa response			DaTSCAN	IBZM
<b>Zuhlke et al;</b> <b>1 female</b> <b>(J Neurol 2004)</b>	T, A, N, P	-	84	N	Abnormal	Abnormal
<b>Ceravolo et al;</b> <b>4 males</b> <b>(Neurology 2005)</b>	T, A, P	-	105	Y	Normal	-
	T, P	-	73	N	Normal	-
	A, C, AutD, P	N	85	N	Normal	-
	T, P	N	97	-	Normal	-
<b>Kamm et al;</b> <b>1 male</b> <b>(Brain 2005)</b>	T, P, A, AutD	N	61	N	-	Abnormal
<b>Scaglione et al;</b> <b>5 Males</b> <b>(J Neurol 2008)</b>	T, P, G, A	-	103	Y	Normal	-
	T, P, G, A	-	80	Y	-	-
	T, P, A	N	91	Y	Abnormal	Abnormal
	T, P, G, A	N	97	Y	Abnormal	-
	T, P, A	-	95	Y	Abnormal	-

<b>Apartis et al;</b> <b>4 females, 18 males*</b> <b>(Neurology 2012)</b>	A (91%) T (86%) P (64%) C (55%)	N (3/3)	102	64% (14/22)	47% (7/15) abnormal	-
<b>Hall et al;</b> <b>2 females, 1 male</b> <b>(Parkinsonism Relat</b> <b>Disord 2010)</b>	T, P  T, P, C  T, P	Y  Y  Y**	41  51  60	N  N  N	Normal  Normal  Normal	-  -  -
<b>Healy et al (case 1)</b> <b>(Mov Disord 2009)</b>	T, P, N, A	N	72	Y	Abnormal	Abnormal
<b>&amp; De Pablo-</b> <b>Fernandez et al</b> <b>(case 2)</b> <b>2 males</b>	T, P, A, G	N	100	N	Abnormal	-
<b>Total:</b>		Y 23% (3/13)		Y 57% (21/37)	Abnormal 45% (13/29)	Abnormal 100% (4/4)

\* Only 15 patients with functional neuroimaging available

\*\* responded to pramipexole

Abbreviations: A, ataxia; AutD, autonomic dysfunction; C, cognitive decline; DaTSCAN, striatal dopamine transporter SPECT; G, vertical gaze palsy; IBZM, striatal postsynaptic D2 receptor SPECT; MCP, middle cerebellar peduncle; N, no; P, parkinsonism; T, tremor; Y, yes.