

CORRIGENDUM

Novel UCHL1 mutations reveal new insights into ubiquitin processing

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In Table 1 of the article, the clinical state of patients III-4, III-5 and III-6 had unfortunately been reported incorrectly in four rows. The correct clinical state in rows 'Brisk deep tendon reflexes (UL/LL)', 'Impaired superficial sensation (UL/LL)' and 'Impaired vibration sense (UL/LL)' are '±' in all, i.e. unaffected upper limbs and affected lower limbs. The correct clinical state for 'Impaired positional

sense' is '+', i.e. affected in all three patients. An updated version of Table 1 is provided below. The table has also been corrected in the original article. The authors apologize for the errors.

Also, post publication the disorder described in the article has been assigned the disease symbol SPG79 (Autosomal recessive spastic paraplegia-79, OMIM 615491).

[†]The authors wish it to be known that, in their opinion, the first two authors should be regarded as joint First Authors.

[‡]The authors wish it to be known that, in their opinion, the last two authors should be regarded as joint Last Authors.

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Table 1. Clinical findings and results of supplementary investigations in the patients in this study (patients III-4, III-5 and III-6), and in the patients described by Bilguvar *et al.* (NG 1024-1, NG 1024-2, NG 1024-3) (14)

	Patient III-4	Patient III-5	Patient III-6	NG 1024-1 ^a	NG 1024-2 ^a	NG1024-3 ^a
Age at examination	65	62	62	28	33	34
Age at onset	10	10	10	5	7	5
First symptom	Visual loss	Visual loss	Visual loss	Visual loss	Visual loss	Balance problems
Vision	0,1 o.u.	Light perception	Light perception	-	-	-
Optical atrophy	+	+	+	+	+	+
SPRS	13	44	45	na	na	na
SARA	18	27	26	na	na	na
Standing without assistance	-	-	-	-	-	-
Gaze evoked nystagmus	+	+	+	+	+	+
Head titubation	+	-	-	+	+	+
Postural tremor	+	-	-	na	na	na
Intention tremor UL	+	+	+	na	na	na
Brisk deep tendon reflexes (UL/LL)	-/+	-/+	-/+	+/+	+/+	+/+
Inverted plantar responses	-	+	+	+	+	+
Hoffman responses	-	-	-	+	-	-
Grade of reduced muscle strength (UL/LL)	Mild/mild	Mild/severe	Mild/moderate	Normal/mild	Normal/mild	Normal/mild
Impaired superficial sensation (UL/LL)	-/+	-/+	-/+	+/+	+/+	+/+
Impaired positional sense	+	+	+	+	+	+
Impaired vibration sense (UL/LL)	-/+	-/+	-/+	+	+	+
Spasticity LL	-	+	+	+	+	+
Abnormal cerebellar tests, dysmetria (UL/LL)	+/nt	+/nt	+/nt	+	+	+
Ophthalmoplegia	-	Paresis upward gaze	-	na	na	na
Hand myotonia	-	-	+	+	-	-
Myokymia	+	-	-	na	na	na
Cognitive impairment	-	-	-	na	IQ 74	IQ 71
Fasciculations	-	+	+	na	na	na
Pectus carinatum	-	+	+	na	na	na
Pes cavus	+	+	+	na	na	na
EMG/neurography (age ^b)	Chronic neurogenic axonal changes (65)	Axonal sensorimotor neuropathy (62)	Axonal sensorimotor neuropathy (62)	Normal nerve conduction velocity, myokymic activity (in all)		
Brain MRI (age ^b)	Optic atrophy (63)	Optic atrophy. Mild cbl.atr. (63)	Optic atrophy (62)	Bilateral optic nerve and chiasm atrophy, Wallerian degeneration of the optic radiations, cbl. and mild cerebral atr. (in all)		
Visual evoked potentials (VEP) flash	na	Inc. lat., red. amp. ^c	Inc. lat., red. amp. ^c	Nearly absent response o.u. (in all)		
VEP electroretinogram	na	Normal	Normal	Normal	Normal	Normal
Muscle biopsy (age ^b)	Normal (56)	na	Chronic denervation (48)	na	na	na

^aExamination findings in patients NG 1024-1, NG 1024-2 and NG 1024-3 are adapted from Bilguvar *et al.*, including Supporting Information.

^bAge at examination, in years.

^cIncreased latency time and reduced amplitude both eyes. LL = lower limbs; UL = upper limbs; - = absent; + = present; o.u. = both eyes; cbl.atr. = cerebellar atrophy; nt = not testable; na = not available.