Supplement Table 1 : Clinical and paraclinical features of copper related neurological disorders

Copper related neurological disorders	***Gene*/protein implicated**	**Clinical features**	**Radiological findings**
Aceruloplasminemia[1,2]	Ceruloplasmin[3]	Dystonia/abnormal gait, dysarthria, dementia[3]	T2* high signal due to progressive iron accumulation involving the retina and basal ganglia[3]
Amyotrophic lateral sclerosis (familial form)[1]	SOD1[4]	Adult-onset progressive muscle weakness[4]	MRI : T2 pyramidal tracts hyperintensity[4], T1 tongue hyperintensity in bulbar form[5]
Deficiency of the Cytochrome C Oxidase Assembly Protein SCO1[1]	Heterozygosity for *SCO1* gene mutations[6]	Neonatal-onset hepatic failure and ketoacidotic comas[6]	No specific MRI pattern
Deficiency of the Cytochrome C Oxidase Assembly Protein SCO2[1]	*SCO2*[7]	Hypertrophic cardio-myopathy and encephalopathy[7]	+/- Brainstem or cerebellar abnormalities seen on MRI[7,8]
Menkes disease[1]	*ATP7A*[9,10]	Feeding difficulties, hypotonia progressing to hypertonia and	MRI : cerebral atrophy, ventriculomegaly, and cerebellar atrophy and

		seizures (including infantile spasms with hypsarrhythmia)[9]	focal lesions in the basal ganglia[10]
MEDNIK syndrom[1]	*AP1S1*[11,12]	Mental retardation, Enteropathy, Deafness, peripheral Neuropathy, Ichthyosis, and Keratoderme[11,12]	MRI : brain atrophy, T2 hyperintensity of the basal ganglia, especially caudate and putamen[11,12]
Occipital horn syndrome[1]	*ATP7A*[13]	Mild allelic variant of Menkes[13]	No specific MRI pattern
Wilson disease[1,2]	*ATP7B*[14]	Movement and behavior/personality disorders[14]	MRI T2 images of high signal intensity in the tegmentum except for the red nucleus, with preservation of the signal intensity of the lateral portion of the pars reticulata of the substantia nigra and hypointensity of the superior colliculus (Panda Sign). Cerebral atrophy[15]
X-Linked Distal Hereditary Motor Neuropathy[1]	*ATP7A*[13]	Distal muscular atrophy and weakness in older children or adults[13]	No specific MRI pattern

Abbreviations :

MRI – Magnetic resonance imaging ; SOD1 – Super Oxyde Dismutase 1 ; SCO1,2 - Synthesis Of Cytochrome C Oxidase 1,2; *ATP7A/B* – ATPase copper-transporting ATPase ½ ; MEDNIK - Mental retardation, Enteropathy, Deafness, peripheral Neuropathy,Ichthyosis, and Keratoderma ; *AP1S1*- Adaptor Related Protein Complex 1 Subunit Sigma 1 ;

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