SUPPLEMENTARY MATERIAL

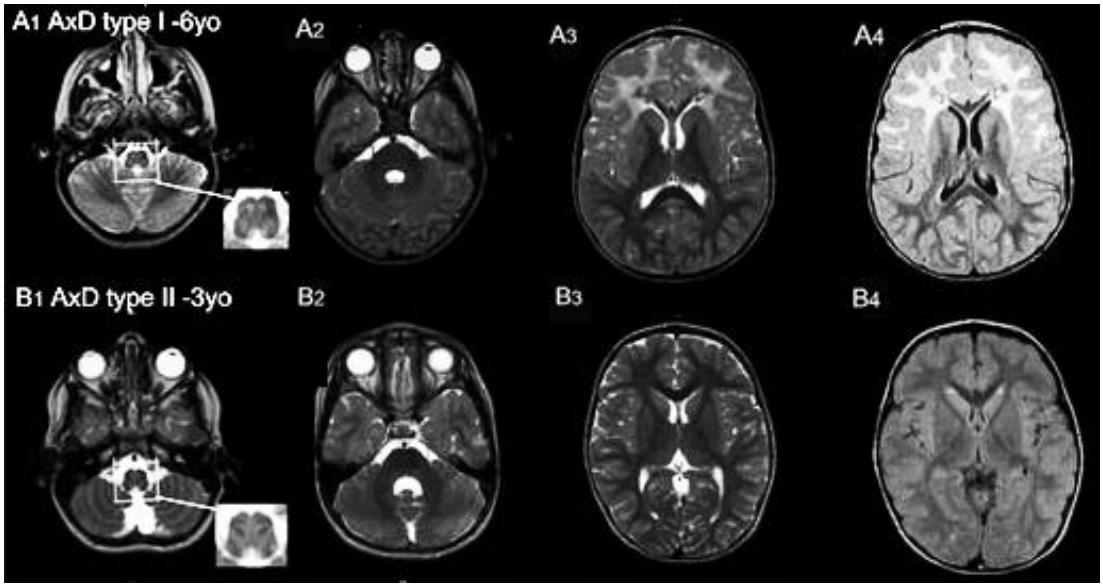

Figure 1: Additional brain MRI features in subjects with AxD presenting the *Chipmunk sign*.

A: Type I AxD, 6-year-old patient; B: type II AxD, 3-year-old patient. Brain axial T2WI (1,2,3). Brain axial T2 FLAIR sequences for all subjects (4). *Chipmunk sign* in axial T2WI focused on the medulla oblongata (boxes in A_1, B_1). Typical frontal white matter involvement predominance in AxD type I (A_{3-4}), in contrast with the AxD type II, that does not show supratentorial white matter involvement (B_{3-4}).

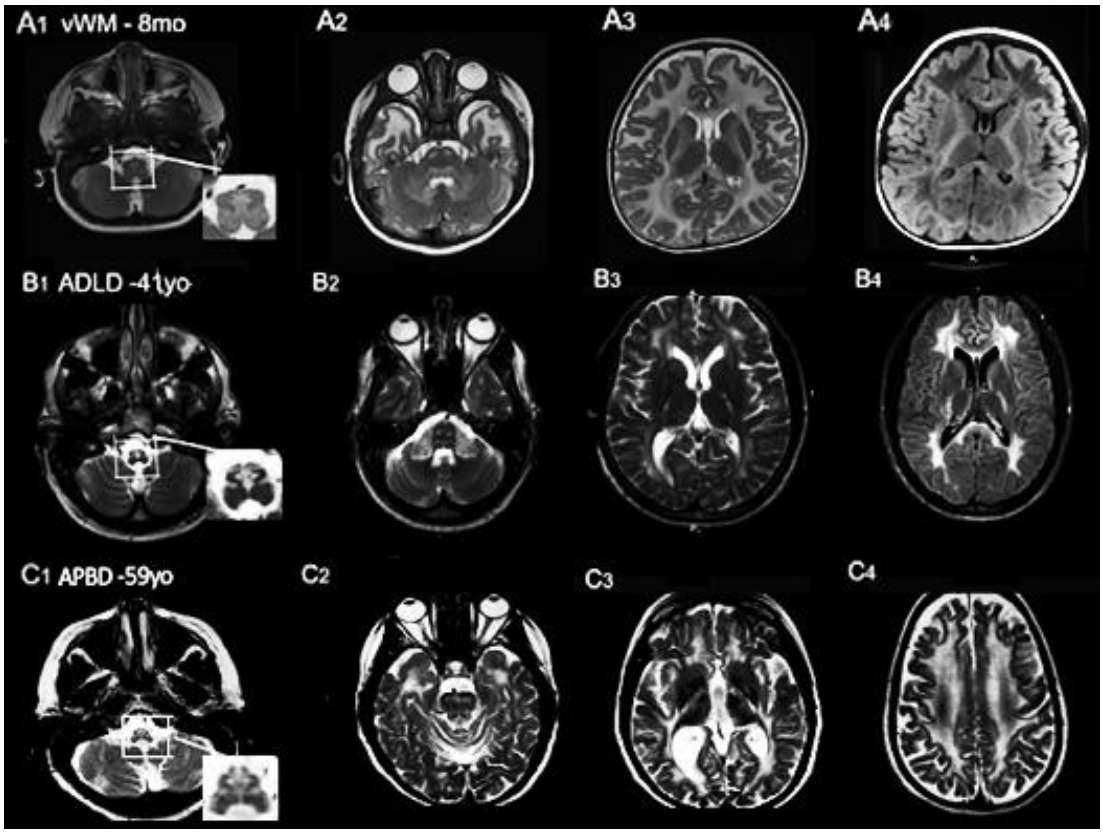

Figure 2: Additional brain MRI features in subjects with other leukodystrophies presenting the *Chipmunk sign*.

A: Vanishing White Matter (VWM), 8-month-old patient; B: Adult-onset autosomal dominant leukodystrophy (ADLD), 41-year-old patient; C: Adult Polyglucosan Body Disease (APBD), 59-year-old patient. *Chipmunk sign* in axial T2WI focused on the medulla oblongata (boxes in A_1, B_1, C_1). Diffuse cerebral white matter involvement in VWM, ADLD and APBD (A_{3-4}, B_{3-4}, C_{3-4}), with no evidence of disease gradient, unlike AxD. VWM shows white matter rarefaction (T2 FLAIR sequences A_4), absent in other subjects (T2 FLAIR sequences B_4, T2WI C_4).

Table 1: Presence of the *Chipmunk Sign* in individuals diagnosed with non-AxD leukodystrophies and genetic leukoencephalopathies.

Diagnosis	*Chipmunk sign*		Total
	Absent	Present	
4H Leukodystrophy	4	1	5
AARS	2	0	2
AARS2	1	0	1
ADLD	2	1	3
AGS	11	0	11
APBD	**0**	**4**	**4**
AxD	**7**	**48**	**55**
Cockayne	3	0	3
DARS	5	0	5
EARS2	2	0	2
GM1	2	0	2
HABC	4	0	4
HDLS	1	0	1
Krabbe infantile	2	0	2
Krabbe juvenile	1	0	1
MCT8 deficiency	1	0	1
Mcomplex I	1	0	1
MLD	2	0	2
NDUFA2	1	0	1
NDUFS7	1	0	1
PEX5	1	0	1
PMD	**2**	**1**	**3**
POLG1	2	0	2
RMND1	2	0	2
SDH	4	0	4
SURF1	1	0	1
VWM	**3**	**3**	**6**
XALD	2	0	2
XALD_ao	1	0	1
Total	71	58	129

4H: 4H Leukodystrophy; AARS: *AARS*-related leukoencephalopathy; AARS2: *AARS2*-related leukoencephalopathy; ADLD: Autosomal dominant leukodystrophy with autonomic disease, AGS: Aicardi-Goutieres Syndrome; APBD: Adult Polyglucosan Body Disease; AxD: Alexander Disease; Cockayne Syndrome; DARS: *DARS*-associated leukoencephalopathy; EARS2: Leukoencephalopathy with thalamus and brainstem involvement and high lactate; GM1: Gangliosidosis GM1; HABC: Hypomyelination with atrophy of Basal ganglia and Cerebellum; HDLS: Hereditary diffuse leukoencephalopathy with neuroaxonal spheroids; MCT8: MCT8 Deficiency; MComplex I: Mitochondrial Complex I deficiency; MLD: Metachromatic Leukodystrophy; NDUFA2: *NDUFA2*- related disorder; NDUFS7: *NDUFS7*- related leukoencephalopathy; PEX5: Peroxisome Biogenesis Factor 5 deficiency; PMD: Pelizaeus-Merzbacher Disease; POLG1: *POLG1*-related leukoencephalopathy; RMND1: *RMND1*-related leukoencephalopathy; SDH: Succinate Dehydrogenase deficiency; SURF1: *SURF1*-related leukoencephalopathy; VWM: Vanishing White Matter; XALD: X-linked Adrenoleukodystrophy.