## Scoring severity of brain MRI findings<sup>8</sup>

T2-weighted imaging hyperintense areas  0 = No hyperintense lesions P1 = Hyperintense lesions confined to periventricular WM P2 = Hyperintense lesions atperiventricular and subcortical WM  D = Hyperintense diffuse changes in WM signal perivascular spaces enlargement at the WM, BG+T, and CC  2 = PVS number < 10 and PVS size < 3 mm 3 = PVS number ≥ 10 and PVS size < 3 mm 3 = PVS number ≥ 10 and PVS size ≥ 3 mm  Cortical atrophy  0 = None 1 = Mild widening of the sylvian fissure and/or the interhemispheric fissure > 3 to >5 mm 3 = Severe widening of the sylvian fissure and/or the interhemispheric fissure > 3 to >5 mm 3 = Severe widening of the sylvian fissure and/or the interhemispheric fissure ≥ 5 mm  Supratentorial ventricles enlargement  0 = None 1 = Width of III ventricle < 5 mm, without temporal horn dilatation 2 = Width of the III ventricle between 5 and 10 mm and temporal horn dilatation 3 = Width of the III ventricle > 10 mm  Sella turcica enlargement  Not present Present  Optic nerve sheath enlargement  0 = Nerve sheath diameter ≤ 5.7 mm 1 = Unilateral nerve sheath diameter > 5.7 mm 2 = Bilateral nerve sheath diameter > 5.7 mm	Cervical MRI	Findings
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1 = Unilateral nerve sheath diameter > 5.7 mm		Present
	Optic nerve sheath enlargement	0 = Nerve sheath diameter ≤5.7 mm
2 = Bilateral nerve sheath diameter >5.7 mm		1 = Unilateral nerve sheath diameter >5.7 mm
		2 = Bilateral nerve sheath diameter >5.7 mm

Adapted from Lachman, J Pediatr Rehabil Med, 2010.

 $Abbreviations: BG+T, basal\ ganglia/thalamus; CC, corpus\ callosum; WM, white\ matter.$