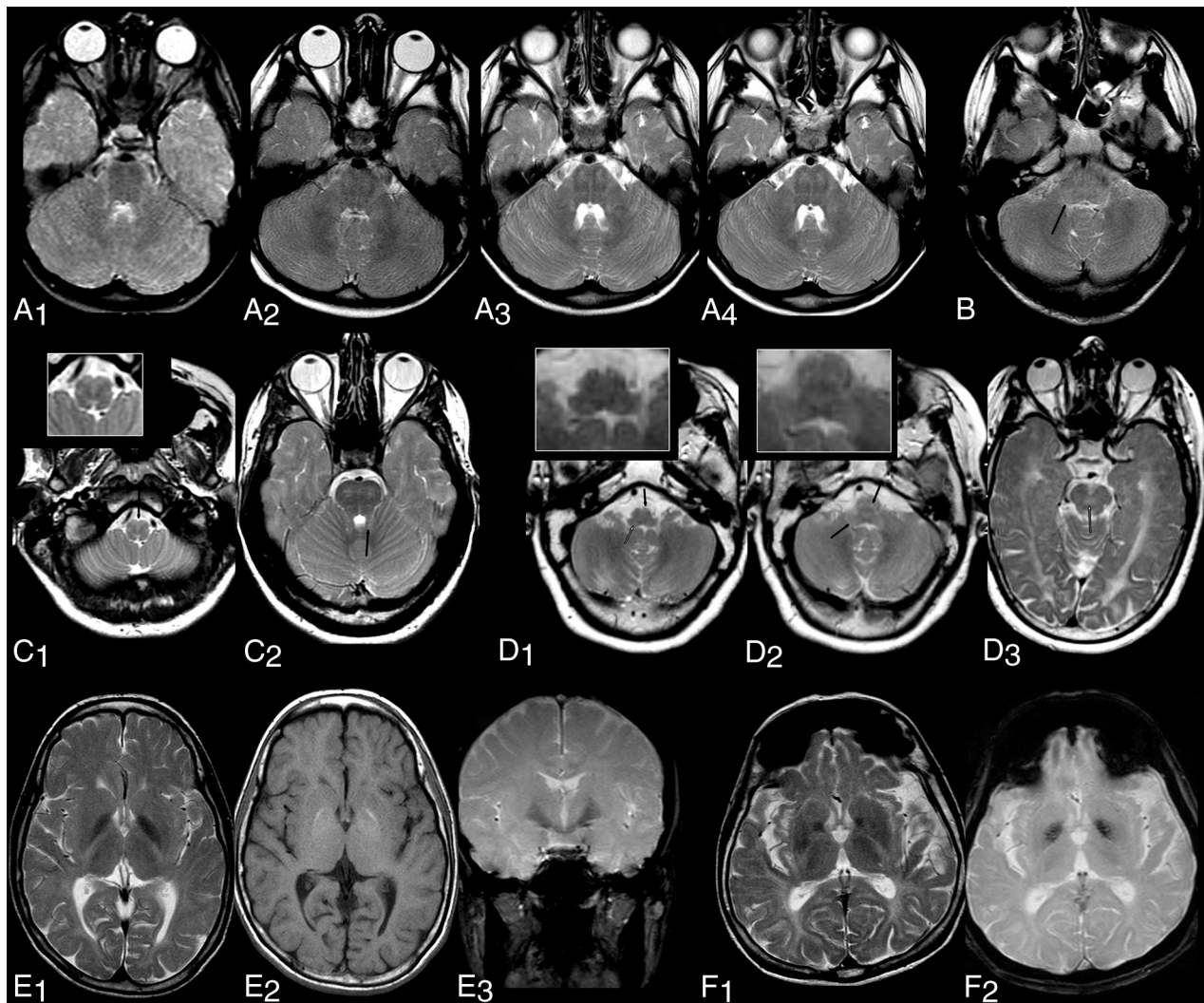


ON-LINE FIG 1. Overview of MR imaging changes of all patients. Representative T2WI at the level of the pons and middle cerebellar peduncles (A), pallidum (B), corpus callosum (C), Rolandic area (D), and midsagittal T1 images (E). Each row shows from left to right patients 1–6, with corresponding annotations, eg, A_{1,2,3}. Field strength is 3T in patient 3 (A₃–E₃); otherwise, it is 1.5T. Images are from the last scan from patients with multiple scans.



ON-LINE FIG 2. Additional brain stem tract (A–D) and pallidal changes (E and F). A, Initially normal signal at 5 years of age (A₁) and evolving T2 hyperintensity of the pontine pyramidal tract in patient 1 on follow-up at 5.8, 7.5, and 8.9 years of age (A_{2–4}). B, Involvement of the inferior cerebellar peduncle in patient 2 (*black arrow*). C, Involvement of the pyramidal tract and medial lemniscus at the level of the medulla oblongata (*black arrow* in C₁) and the superior cerebellar peduncle (*black arrow* in C₂) in patient 3. D, T2 hyperintensity of the pyramidal tract and inferior cerebellar peduncle at the level of the medulla (D₁) and pontomedullary junction (D₂) as well as of crossing of the superior cerebellar peduncle at the level of the mesencephalon (D₃) in patient 4 (*black arrows*). E, Signal changes of the pallidum in patient 2 (E) and patient 6 (F) with pronounced hypointensity on T2-weighted and gradient-echo T2*-weighted images in patient 2 with corresponding abnormal T1 hyperintensity. (T2WI: A–D, E₁, F₁; T1WI: E₂; T2*WI: E₃, F₂).

On-line Table 2: Metabolite changes in white matter ¹H-MRS

Patient (Sex)	Age (yr)	NAA	Cho	Cr	mIns	NAA/Cr	Cho/Cr	mIns/Cr	Lac
1 (M)	8.9	~	~	(↑)	~	(↓)	(↓)	(↓)	ND
2 (F)	18.2	~	~	↑	(↑)	↓	↓	~	ND
3 (F)	19.2	~	~	~	~	~	~	~	ND
4 (F)	40.8	(↓)	(↓)	↑	~	↓	↓	↓	ND
5 (M)	43.7	~	~	↑	↑	(↓)	↓	~	ND
6 (F)	51.9	~	↓	↑	↑	(↓)	↓	(↑)	ND

Note:—ND indicates not detected; Lac, lactate; ~, metabolite values within the mean \pm 1 SD; (↑)/(↓), values differing between 1 and 2 SDs from the mean; ↑/↓, values differing >2 SDs from the mean.