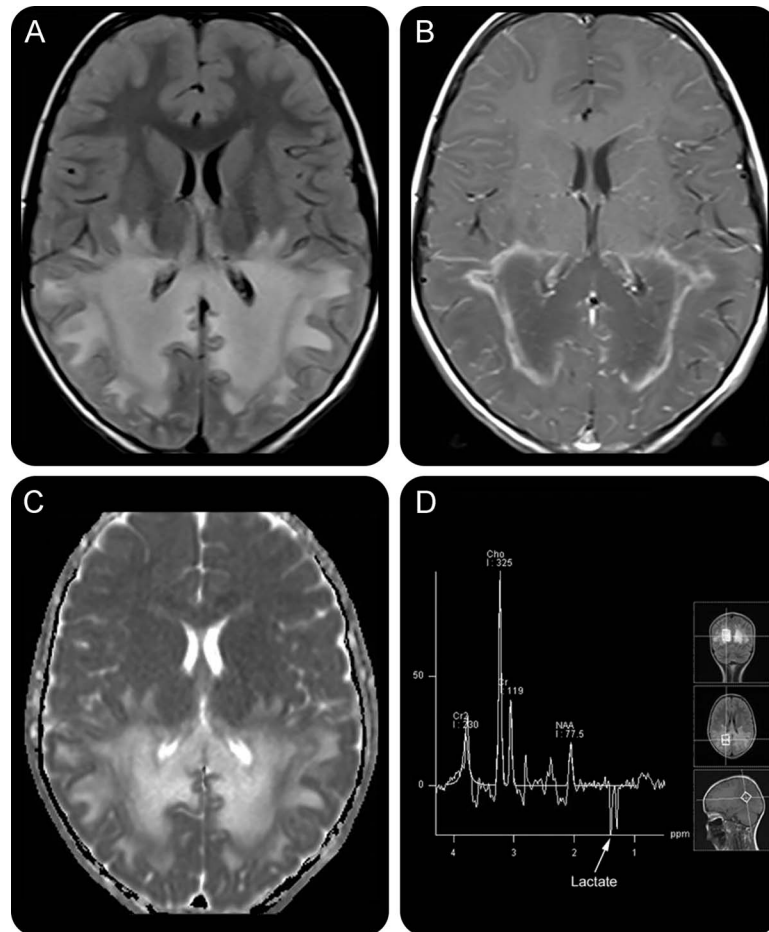


# Teaching NeuroImages: MRI findings in X-linked adrenoleukodystrophy

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**Figure** MRI brain with spectroscopy at the time of diagnosis



(A) Fluid-attenuated inversion recovery (FLAIR) image shows symmetric confluent hyperintense white matter lesion in posterior cerebral hemispheres. (B) Postcontrast T1-weighted image shows symmetric linear enhancement, known as intermediate inflammatory zone, which is much less hyperintense on FLAIR and apparent diffusion coefficient map (C). (D) Magnetic resonance spectroscopy shows decreased N-acetylaspartate (NAA) and elevated choline (Cho) and lactate peaks. Cr = creatine.

A 6-year-old boy presented with 4 months progressive verbal comprehension difficulties, declining school performance, strabismus, and visual impairment. General examination noted moderate hypotension and hyperpigmented skin. Pertinent neurologic examination abnormalities included difficulty with comprehension, decreased visual acuity (20/50 bilaterally), disconjugate gaze, bilateral dysmetria, and unsteady gait. CSF protein

was elevated (1.82 g/L). Brain MRI showed posterior cerebrum and brainstem abnormalities consistent with adrenoleukodystrophy<sup>1</sup> (figure). Morning cortisol was decreased (21 nM) and adrenocorticotrophic hormone stimulation test confirmed adrenal insufficiency. Diagnosis was confirmed by very long chain fatty acid analysis (elevated C26:0/C22:0 and C24:0/C22:0 ratios) and identification of pathogenic *ABCD1* mutation.

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## AUTHOR CONTRIBUTIONS

Kenneth A. Myers: drafted the manuscript, prepared the figures, and approved the final manuscript as submitted. Jong M. Rho: reviewed and revised the manuscript and approved the final manuscript as submitted. Dr. Rho ensured that the patient history was accurately characterized. Xing-Chang Wei: reviewed and revised the manuscript and approved the final manuscript as submitted. Dr. Wei ensured that the patient's MRI findings were accurately characterized.

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## DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org](http://Neurology.org) for full disclosures.

## REFERENCE

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