

Alexander Disease

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History

2 year old male with macrocephaly and regressive development.

Diagnosis

Alexander Disease

Discussion

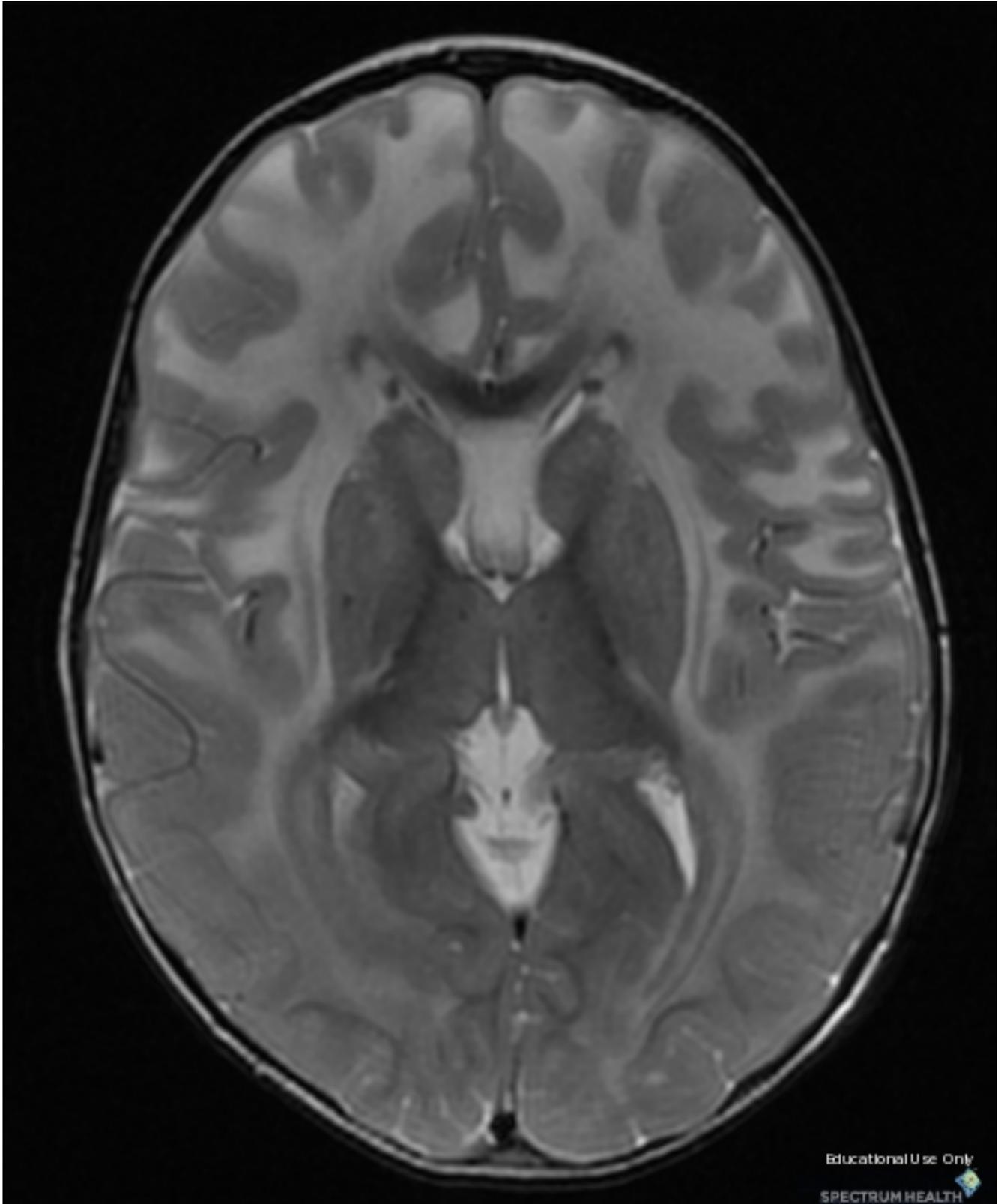
Alexander disease, or fibrinoid leukodystrophy, is characterized by massive deposition of Rosenthal fibers (astrocytic cytoplasmic inclusion bodies) in the subependymal, subpial, and perivascular regions. Three clinical forms exist. The infantile form is characterized by early onset of macrocephaly, psychomotor retardation, and seizure with death occurring within 2–3 years. In the juvenile form, progressive bulbar symptoms and spasticity manifest between 7 and 14 years of age. In adults, symptoms may be indistinguishable from multiple sclerosis. Alexander disease has a predilection for the frontal subcortical white matter early which becomes more confluent and eventually progresses posteriorly to the parietal white matter and internal and external capsules. White matter involvement is hypodense on CT and T2 hyperintense on MR; enhancement is often seen near the tips of the frontal horns early in the disease course. In the late stages of the disease, cysts may develop.

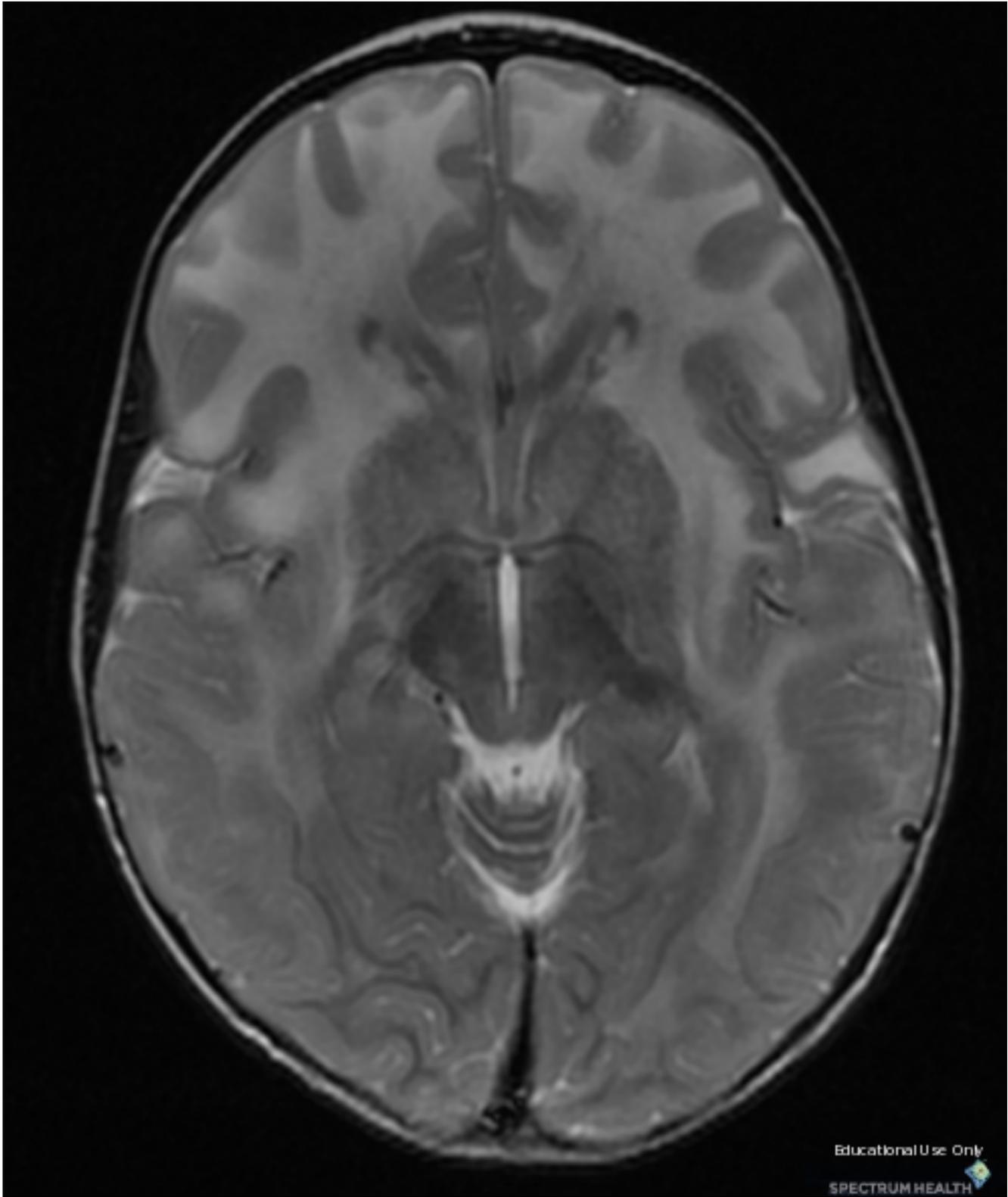
Findings

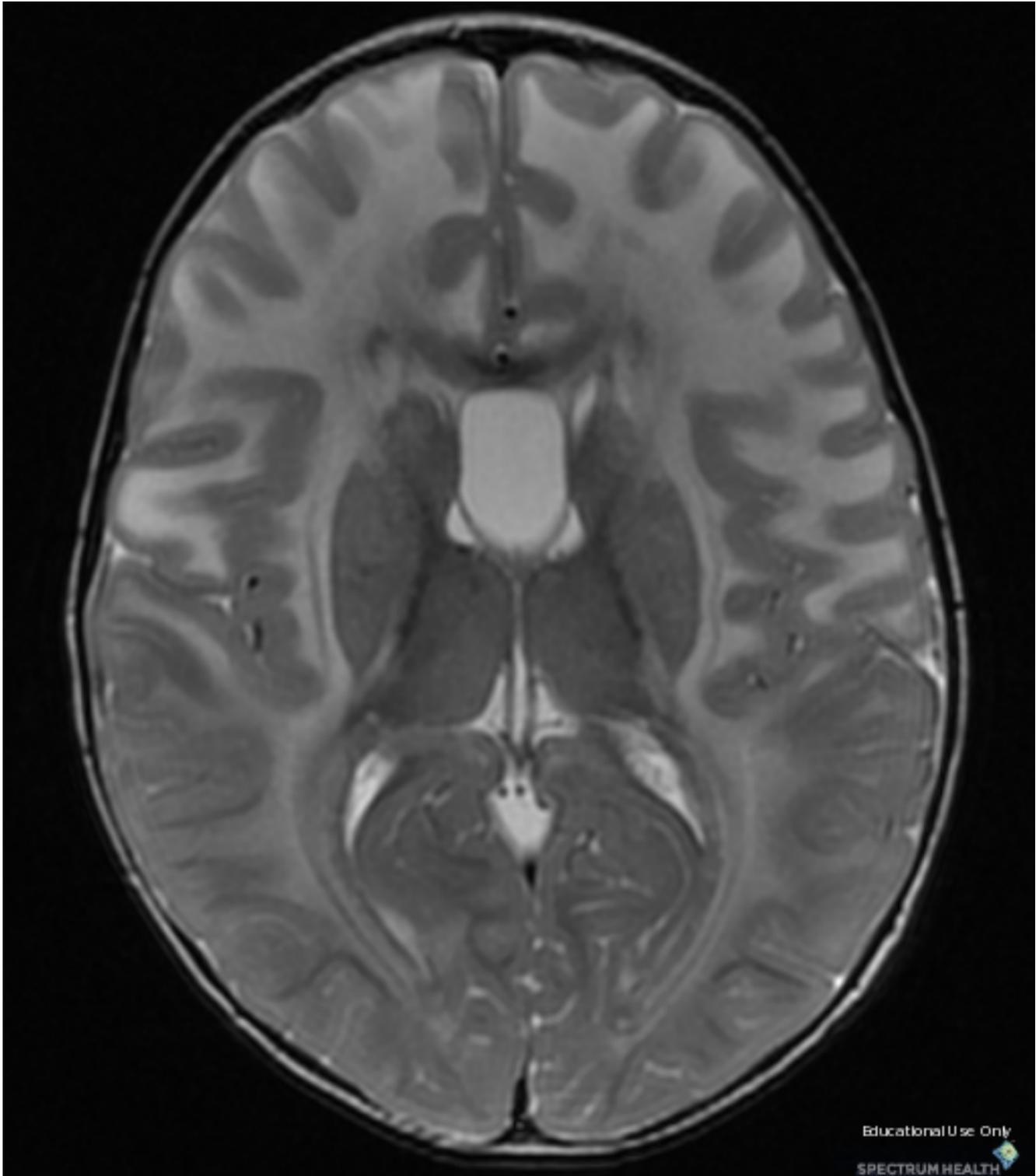
MR-Extensive symmetric T2 hyperintensity of the cerebral white matter mainly anteriorly but also involving the external capsules.

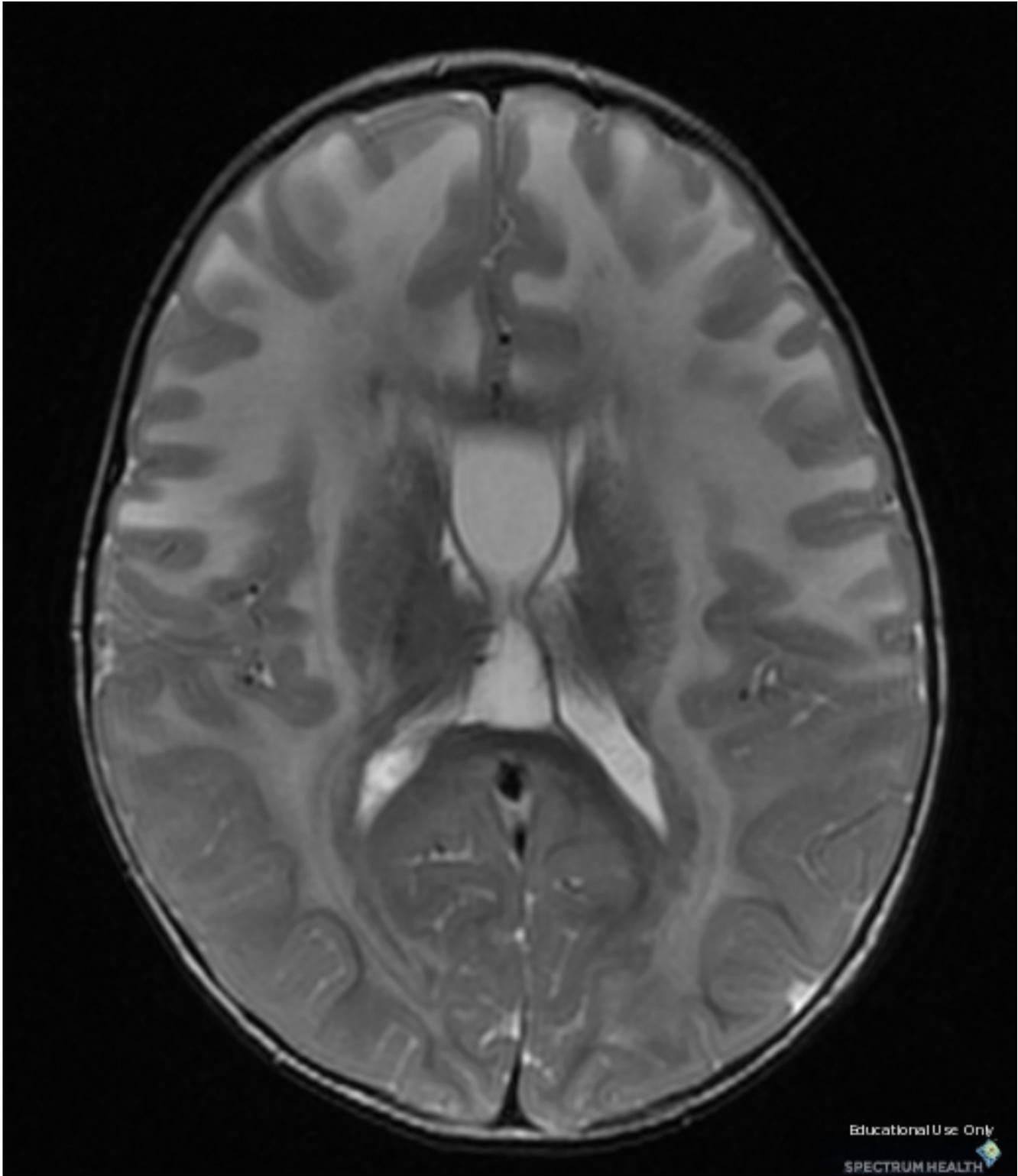
Reference

Cheon JE, Kim IO, Hwang YS, et al. Leukodystrophy in Children: A Pictorial Review of MR Imaging Features. *Radiographics* (2002); 22:461-476.









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