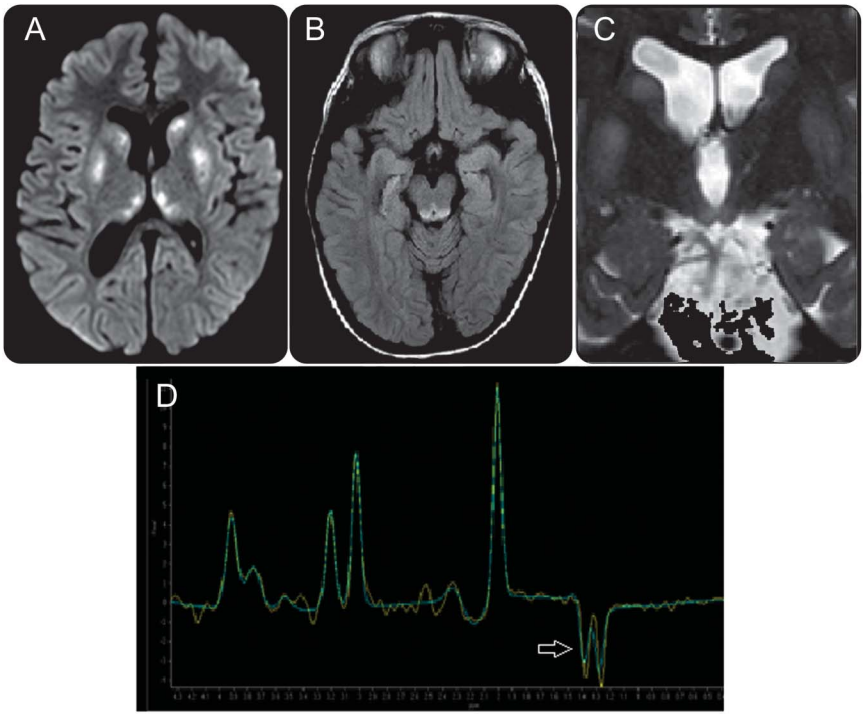
MR spectroscopy in pediatric Wernicke encephalopathy

Figure Brain magnetic resonance with spectroscopy



(A) Diffusion restriction in bilateral striata and medial thalami. (B) Increased T2 signal in periaqueductal gray on axial fluid-attenuated inversion recovery. (C) T2 hyperintensities in mamillary bodies on coronal T2. (D) Magnetic resonance spectroscopy demonstrating a large lactate doublet (arrow) at 1.33 ppm (voxels taken from left basal ganglia and left parietal white matter).

The patient is a 7-year-old girl with a history of repaired gastroschisis and short gut syndrome. She presented with a 4-day history of progressive encephalopathy, truncal ataxia, and omnidirectional gaze-evoked nystagmus. Peripheral lactate was 7.1 mmol/L. Serum thiamine level was decreased (<5 nmol/L). Brain magnetic resonance with spectroscopy is shown in the figure. A clinical diagnosis of Wernicke encephalopathy was made. The patient was started on thiamine and demonstrated a rapid clinical recovery. Peripheral lactate normalized within 1 day. Increased lactate has been reported in thiamine deficiency, presumed secondary to the role of thiamine as a cofactor for the pyruvate dehydrogenase complex and α-ketoglutarate dehydrogenase.[1]

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Author contributions: Lance H. Rodan: preparation of manuscript. Navin Mishra: editing of manuscript. Ingrid Tein: editing of manuscript, supervisor.

Study funding: No targeted funding reported.

Disclosure: The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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1. Desjardins P, Butterworth R. Role of mitochondrial dysfunction and oxidative stress in the pathogenesis of selective neuronal loss in Wernicke's encephalopathy. Mol Neurobiol 2005;31:17–25.

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Neurology 2013;80;969
DOI 10.1212/WNL.0b013e3182840cd9

This information is current as of March 4, 2013

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