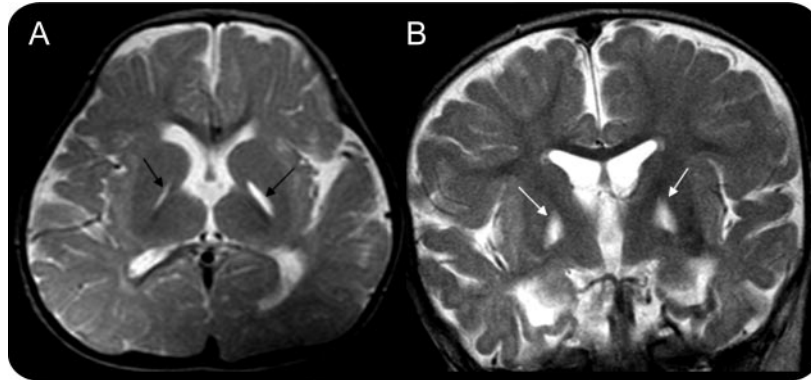


Teaching NeuroImages: MRI in methylmalonic acidemia

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Figure MRI: Axial (A, black arrows) and coronal (B, white arrows) T2W MRI demonstrating bilateral symmetric hyperintensity involving globus pallidi



An 18-month-old girl presented with recurrent episodes of encephalopathy, starting from the third postnatal day, and delayed development. Her parents were nonconsanguineous. She had microcephaly, generalized hypotonia, brisk stretch reflexes, extensor plantar response, choreiform movements, and dystonia of hands and feet. Evaluation showed metabolic acidosis and hyperammonemia. Tandem mass spectroscopy at age 18 months revealed elevated methylmalonyl carnitine ($2.68 \mu\text{mol/L}$; normal $0.00\text{--}1.02$) and propionyl carnitine ($16.73 \mu\text{mol/L}$; normal $0.08\text{--}6.50$), indicating methylmalonic acidemia (MMA). MRI at the same time showed bilateral symmetric T2 hyperintensities involving globus pallidi

(figure, A and B). Selective necrosis of the globus pallidus is an important MRI finding in MMA.^{1,2} Differential diagnoses include propionic acidemia, pyruvate dehydrogenase deficiency, kernicterus, and carbon monoxide poisoning. Management consists of protein restricted diet, carnitine, and parenteral vitamin B12.

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Disclosure: The authors report no disclosures.

Neurology[®]

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Neurology 2010;74;e14
DOI 10.1212/WNL.0b013e3181cc0b7b

This information is current as of January 25, 2010

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