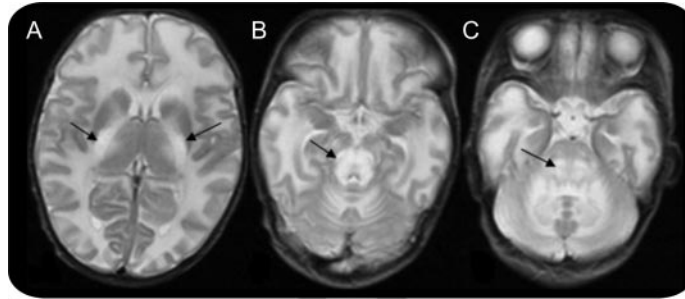


Teaching NeuroImages: MRI in maple syrup urine disease

Parayil Sankaran Bindu,
MD
Jerry M.E. Kovoov, MD
Rita Christopher, MD

Address correspondence and
reprint requests to Dr. Parayil
Sankaran Bindu, Department
of Neurology, NIMHANS,
Bangalore, India
drpsbindu@yahoo.co.in

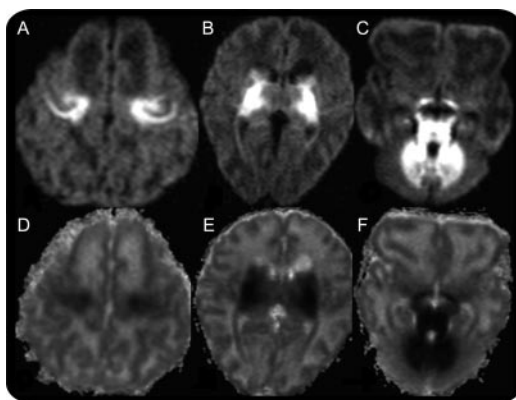
Figure 1 T2-weighted MRI demonstrates elevated signal intensity of white matter in the thalamus, globus pallidus, and posterior limb of the internal capsule (A); midbrain (B); and pons and cerebellar white matter (C)



A baby girl, born normally to consanguineous parents, presented on the fifth postnatal day with poor feeding, lethargy, and seizures. Examination

on the 10th day showed hypotonia and poor neonatal reflexes. Tandem mass spectroscopy showed elevated branched chain amino acids suggesting maple syrup urine disease (MSUD). MRI on day 14 revealed findings typical of MSUD (figures 1 and 2).^{1,2} The characteristic pattern of restricted diffusion, attributed to intramyelinic edema, corresponds to areas that are myelinating at birth. Unmyelinated areas show vasogenic edema. A similar pattern of restricted diffusion is seen in nonketotic hyperglycinemia and Canavan disease.

Figure 2 Diffusion-weighted images with corresponding apparent diffusion coefficient maps demonstrate restricted diffusion in the central part of the centrum semiovale (A, D); posterior limbs of the internal capsules and thalami (B, E); and pons and cerebellar white matter (C, F)



REFERENCES

1. Sakai M, Inoue Y, Oba H, et al. Age dependence of diffusion-weighted magnetic resonance imaging findings in maple syrup urine disease encephalopathy. *J Comput Assist Tomogr* 2005;29:524–527.
2. Jan W, Zimmerman RA, Wang ZJ, Berry GT, Kaplan PB, Kaye EM. MR diffusion imaging and MR spectroscopy of maple syrup urine disease during acute metabolic decompensation. *Neuroradiol* 2003;45:393–399.

From the Departments of Neurology (P.S.B.), Neuroimaging & Interventional Neuroradiology (J.M.E.K.), and Neurochemistry (R.C.), National Institute of Mental Health And Neurosciences (NIMHANS), Bangalore, India.

Disclosure: The authors report no disclosures.

Neurology[®]

Teaching NeuroImages: MRI in maple syrup urine disease
Parayil Sankaran Bindu, Jerry M.E. Kovoor and Rita Christopher
Neurology 2010;74:e12
DOI 10.1212/WNL.0b013e3181ca0101

This information is current as of January 18, 2010

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/74/3/e12.full
References	This article cites 2 articles, 0 of which you can access for free at: http://n.neurology.org/content/74/3/e12.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Pediatric http://n.neurology.org/cgi/collection/all_pediatric Amino acid http://n.neurology.org/cgi/collection/amino_acid DWI http://n.neurology.org/cgi/collection/dwi Metabolic disease (inherited) http://n.neurology.org/cgi/collection/metabolic_disease_inherited MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

