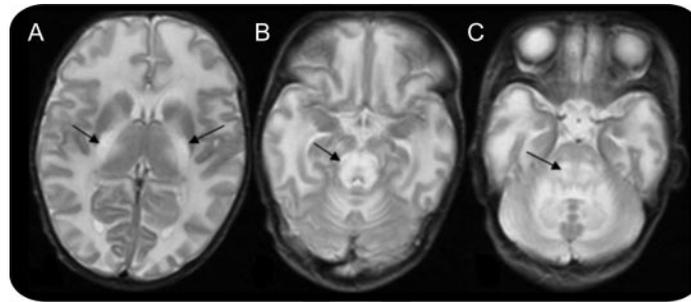


# Teaching NeuroImages: MRI in maple syrup urine disease

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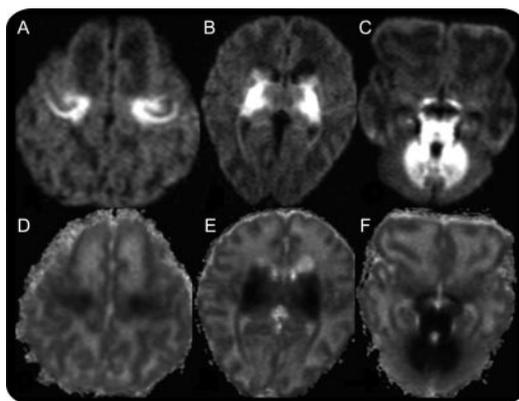
**Figure 1** T2-weighted MRI demonstrates elevated signal intensity of white matter in the thalamus, globus pallidus, and posterior limb of the internal capsule (A); midbrain (B); and pons and cerebellar white matter (C)



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).<sup>1,2</sup> 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)



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