Brain MRI normalization after liver transplantation

Before and at 1 and 12 months after successful liver transplantation

Rovira et al. demonstrate on fast-FLAIR MR images the presence of asymptomatic high-signal intensity in the hemispheric white matter in patients with chronic liver disease. Normalization of this signal abnormality with liver transplantation suggests that it reflects mild edema.

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The value of MRI in hepatic encephalopathy

Commentary by Alan H. Lockwood, MD

Hepatic encephalopathy refers to the spectrum of neuropsychiatric abnormalities attributable to liver failure.¹ At present, the criteria that define the syndrome are usually based on clinical and neuropsychological evaluations. During the past several years, increasingly sophisticated imaging techniques have taught us more about the pathophysiology of this syndrome and suggested the possibility of an image-based diagnosis.²

The Rovira et al. report uses MR techniques that suggest that patients with severe liver disease have asymptomatic edema of the white matter that resolves after successful

transplantation.¹ Earlier studies have shown that T1-weighted scans of these patients are likely to reveal high-signal abnormalities in the globus pallidus, an abnormality linked to the presence of excess manganese. More careful quantitative studies have shown that T1 signal abnormalities extend throughout the brain. Others have shown shortening of the T2 values in many brain areas, an abnormality that is not usually evident on a visual examination of scans. Finally, spectroscopic studies have shown abnormalities in the concentration of myoinositol, choline, and glutamine.

Hepatic encephalopathy is still a clinical diagnosis. However, it seems likely that imaging techniques will become increasingly important in evaluating and monitoring these patients.

References

1. Ferenci P, Lockwood AH, Mullen KD, Tarter RE, Weissenborn K, Blei AT et al. Hepatic encephalopathy—definition, nomenclature, diagnosis and quantification: final report of the working party at the 11th World Congresses of Gastroenterology, Vienna, 1998. *Hepatology* 2002;35:716–721.
2. Lockwood AH, Weissenborn K, Butterworth R F. An image of the brain in patients with liver disease. *Curr Opin Neurobiol* 1997;10:525–533.

Is low potassium a risk factor for stroke?

The Green et al. study of older adults found an increased risk of stroke among diuretic users with lower serum K⁺ and among nonusers of diuretics with low dietary K intake. Diuretic users with both lower serum K⁺ and atrial fibrillation had a 10-fold greater risk of stroke compared to those with higher serum K⁺ and normal sinus rhythm.

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The accompanying editorial by Levine and Coull notes the fact that the modern Western diet has a very low K content when compared with prehistoric humans and with lower stroke risk populations. They review animal data supporting a salutary effect of dietary K and consider possible mechanisms of K benefit. The impressive epidemiologic and animal data and the low cost of K supplementation argue for a clinical trial to test the hypothesis.

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Bioterrorism and CNS infections: Hemorrhagic CSF with high mortality: Anthrax meningoencephalitis

The Lanska review of anthrax meningoencephalitis considers the clinical findings, diagnostic test results, treatment, and outcome of reported cases over the last 50 years. CSF demonstrated hemorrhagic meningitis with positive Gram's stains and CSF cultures. Anthrax meningoencephalitis has a high case-fatality rate, even with aggressive antibiotic treatment and supportive therapy.

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The accompanying editorial by Roos considers the various agents that can be "weaponized," focusing on the frequently lethal airborne B. anthracis. She details current treatment recommendations and notes that in some settings even a suspected traumatic LP may demand urgent treatment for possible anthrax infection.

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Behavior disorders associated with Tourette's syndrome

To overcome the potential confound of ascertainment bias in determining the behavior spectrum of Tourette's syndrome, Kurlan et al. used standard psychiatric interviews and rating scales to study 1,596 community school children. The following conditions were linked to the presence of tics: obsessive-compulsive disorders, anxiety disorders, mood disorder, attention-deficit disorder, and disruptive behavior.

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Improved diagnosis of ulnar neuropathy at the wrist

*Wrist and palmar
sites of stimulation*

Cowdery et al. prospectively evaluated 20 patients with clinically defined ulnar neuropathy at the wrist (UNW) vs normal and ulnar neuropathy at the elbow controls. Conduction block and conduction slowing across the wrist are more sensitive and specific for UNW than traditional electrodiagnostic tests.

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Migraine and cervical artery dissection

Tzourio et al. found that a history of migraine was more frequent in patients with a cervical artery dissection than in controls, suggesting that an underlying arterial anomaly could predispose to migraine.

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Congenital disorders of eye movements: Looking through a window at neurodevelopment

The fourth ventricle is enlarged; the pons and medulla have reduced volume

Pieh et al. correlated MRI and clinical findings in two siblings with the syndrome of congenital horizontal gaze palsy with kyphoscoliosis (HGPS). MRI revealed a hypoplastic pons and dysplastic medulla oblongata.

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Jen et al. mapped horizontal gaze palsy with progressive scoliosis, an autosomal recessive disorder with congenital gaze palsy and delayed scoliosis, to chromosome 11q. Genes involved in neuronal development are likely candidates.

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The accompanying editorial by Engle and Leigh notes how MRI localization and genetic linkage are combining to make rapid progress in identifying genes responsible for neurodevelopment of brain stem nuclei and the disorders they can cause: congenital fibrosis of the extraocular muscles, congenital ptosis, Duane and Möbius syndromes, in addition to HGPS.

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Distal myopathy with rimmed vacuoles vs hereditary IBM: Mutations in the GNE gene

V572L in exon 10

In distal myopathy with rimmed vacuoles (DMRV) patients, Tomimitsu et al. found three novel mutations in the *N*-acetyl mannosamine kinase gene, the causative gene of one form of hereditary inclusion body myopathies. DMRV and the h-IBM appear to be a single disease—clinically similar and sharing similarities in the sites of mutation in the *GNE* gene.

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Recovery from aphasia: A poem and personal history

Kamal is a physicist with aspirations to be a writer. He suffered a stroke with aphasia 2 years ago. In “Nisus: Neurology and the Humanities,” he gives poetic expression to his experience of having “gambled away all my precious words/in on prodigal night.”

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