## Striatal transplantation in Huntington's disease

*" . . . are fetal transplantation approaches really leading to something better?"*

Hauser et al. report results of a seven-patient open-label study of bilateral transplantation of human fetal striatal cells in Huntington's disease (HD). In a subject who died 18 months after transplantation, autopsy demonstrated surviving grafts of typical developing striatal morphology. Three subjects developed subdural hemorrhages and two required surgical drainage.

**see page 687**

*"If we replace only 8 to 10% of the striatum, which represents only 20% of the degeneration in HD, are we likely to have a major impact?"*

*The accompanying editorial by Greenamyre and Shoulson emphasizes the compelling need for treatment for desperate patients with HD. However, striatal degeneration, while characteristic of HD, in only part of the disease and is often the least disabling. While it is logical to attempt to replace neurons, there are no controlled data to support the transplantation of fetal striatal cells. Moreover, Hauser et al. showed no change in the primary outcome variable—motor function—and the procedure had major complications.*

**see page 675**

## Regional and progressive thinning of the cortical ribbon in Huntington's disease

Rosas et al. describe regional cortical thinning in a cohort of patients with Huntington's disease using a novel surface reconstruction technique. This cortical involvement is often selective and may account for some of the variation in the clinical picture in HD. These results also emphasize the point that Greenamyre and Shoulson make concerning the major deficits of HD unrelated to striatal degeneration.

**see page 695**

## Antibody heterogeneity in patients with Rasmussen encephalitis (RE)

Baranzini et al. studied antibody diversity in the RE brain. They found evidence of expansions of selected clones of B cells, implying the presence of an antigen-driven immune response. Analysis of patients' sera reveals a wide spectrum of reactivity against neural antigens.

**see page 709**

*The accompanying editorial by McNamara traces the history of RE, from its original description by Rasmussen as intractable focal epilepsy in children, to its current understanding as a treatable autoimmune disorder resulting from antibodies to glutamate receptors—and additional neural antigens as suggested by the results of Baranzini et al.*

**see page 677**

## Normal metabolism by PET in children with focal epilepsy

Gaillard et al. studied 40 children with recent-onset, localization-related epilepsy with FDG-PET. They found regional hypometabolism in only 20% of children. Metabolic abnormalities are less common and profound in children with new-onset epilepsy than in adults with chronic partial epilepsy.

**see page 717**

## Pediatric frontal and temporal lobe epilepsy

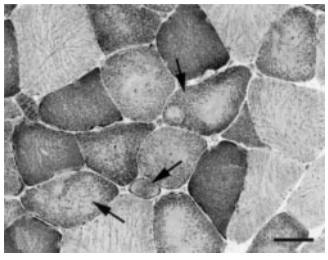
Lawson et al. compared 56 children with intractable frontal (FLE) or temporal lobe epilepsy (TLE) using video-EEG telemetry and quantitative MRI. In contradistinction to TLE, nonlateralized EEG findings and bilateral frontal cortical volume loss suggested that FLE is frequently a bilateral disease. Thus, a minority of FLE children have a good surgical outcome.

**see page 723**

*continued on page 674*

## Progressive myosin myopathy with rimmed vacuoles



Tajsharghi et al. described an autosomal dominant congenital myopathy associated with a missense mutation in the myosin heavy chain IIa gene. The expression of the aberrant copy of the gene increased progressively with age and was associated with dystrophic muscle changes and rimmed vacuoles.

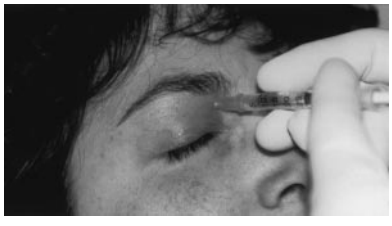
**see page 780**

## Adaptation in the motor cortex following cervical spinal cord injury

Mikulis et al., studying cortical activation in the primary sensorimotor cortex following chronic cervical spinal cord injury, found that the de-efferented/de-afferented ("disconnected") cortex represents an available motor network that becomes active during movement of muscles unaffected by injury.

**see page 794**

## Trocheitis: A treatable trigger of migraine



Yangüela et al. describe trochleitis, causing orbital pain, associated with migraine headache. Local infiltration of the trochlea region improved both disorders. Trochleitis may represent a new and treatable trigger point in migraineurs.

**see page 802**

## Accuracy of muscle localization in limb dystonia

Botulinum toxin is an established treatment for dystonia; however, the role of EMG is unclear. Molloy et al. demonstrate that EMG guidance is needed for correct localization of desired muscles.

**see page 805**

## Spontaneous CSF leaks: Connective tissue disorder?

Mokri et al. identified features of a connective tissue disorder in nine of 58 consecutive patients with spontaneous CSF leaks. A dural weakness likely preexists in some spontaneous CSF leaks.

**see page 814**

## Effects of L-arginine on the acute phase of stroke in three patients with MELAS

The cause of stroke in MELAS is not known. Koga et al. administered L-arginine as a vasodilator to three MELAS patients in the acute phase of stroke evaluating its clinical, biochemical, and hemodynamic effects. L-Arginine improved clinical findings and lowered serum lactate.

**see page 827**