

nerve pain, without rash, with a mass in the trigeminal ganglion and chronic ganglionitis (Hevner R et al. **Lancet Neurol** 2003;2:567-571).

Ocular manifestations of the congenital varicella syndrome were described in 3 children seen at Great Ormond Street Hospital, London, UK (Lambert SR et al. **Arch Ophthalmol** 1989;107:52-56) and reviewed in **Progress in Pediatric Neurology I**, PNB Publ, 1991;424-5). Ocular abnormalities followed a maternal varicella infection in the second trimester of pregnancy, and included chorioretinitis, atrophy of optic discs, cataract, and Horner's syndrome. Neurologic complications included bulbar palsy, hemiparesis, learning disorder, and psychomotor retardation.

Varicella with delayed hemiplegia is described in 4 children reported from Japan. (Ichiyama T et al. **Pediatr Neurol** 1990;6:279-281; and **PPN I**, PNB 1991;425). Cerebral angiitis with occlusion of the middle cerebral artery was cited as the cause. Neurologic complications of varicella are due to viremia with encephalitis, post-exanthematous encephalitis or cerebral angiitis. Cerebellar ataxia is the most frequent neurologic complication of varicella and hemiplegia is unusual.

IMMUNOTHERAPIES IN RASMUSSEN'S ENCEPHALITIS

Corticosteroids, IV immunoglobulins (IVIg), cyclophosphamide, therapeutic plasma exchange (TPE), and protein A IgG immunoabsorption (PAI) were evaluated in 15 patients with confirmed Rasmussen's encephalitis (RE) and reported from various centers in Italy. *Steroids* were used for status epilepticus or worsening seizures. In 6/11 patients steroid treatment temporarily reduced seizure frequency. All attempts to withdraw prednisone resulted in seizure exacerbation, and treatment was maintained until surgery. *IVIg* in 11 patients was partially effective in 6. *Cyclophosphamide* was ineffective in 4 patients treated, 1 developing severe leukopenia. *TPE* was effective in the control of status epilepticus in 1 of 5 patients treated. *PAI* given chronically initially controlled status epilepticus and improved the neurologic condition in 3 patients treated, but the effect was not sustained. *Surgery* (hemispherotomy or hemispherectomy) in 13 patients at 5.2 +/- 3.9 years after disease onset controlled seizures initially, and antiepileptic drugs were withdrawn in 4 and reduced in 7. After surgery, the neurologic condition improved in all patients. Surgery is the only treatment that halts disease progression. Immunomodulatory therapy is considered when surgery is not feasible, in late-onset patients with slow progression, and in the infrequent case of bilateral disease. (Granata T, Fusco L, Gobbi G et al. Experience with immunomodulatory treatments in Rasmussen's encephalitis. **Neurology** December (2 of 2) 2003;61:1807-1810). (Reprints: Dr Tiziana Granata, Division of Child Neurology, Istituto Nazionale Neurologico C. Besta, Via Celoria 11, 20133 Milan, Italy).

COMMENT. The authors propose immunotherapy to delay surgery in patients with 1) late onset (adolescent or adult) RE with slower and milder course than the typical childhood-onset form; 2) dominant hemisphere involvement and slow progression, when surgery is contraindicated; 3) suspected cases of RE without deterioration and atrophy; 4) bilateral RE, and 5) severe cases requiring stabilization before surgery. Surgery is the preferred and optimal treatment to control seizures and halt neurologic deterioration. Bolus corticosteroids and PAI block status epilepticus. IVIg may have a role in adult patients but is ineffective in childhood.