

## CONGENITAL MALFORMATIONS

### **PERINEAL SENSATION PREDICTIVE OF SPINA BIFIDA OUTCOME**

Neurologic examination, including perineal sensation, was conducted in a prospective cohort study of 117 consecutive patients with open spina bifida at St George's, University of London, and Addenbrooke's Hospital, Cambridge, UK. Backs were closed non-selectively at birth between 1963 and 1971. Data were recorded within 48 hrs of birth and during six reviews between 1972 and 2002. Thirty three (28%) had perineal sensation, defined as intact sensation to pinprick in at least one dermatome on one side in the saddle area (S2-4). At follow-up using medical records and the Office of National Statistics, by December 2005, 57% (67/117) of the cohort had died. The mean age of the 50 survivors (43% of the cohort) was 38 years (range 35-41). Of 33 with intact perineal sensation, 23 (70%) survived, whereas only 27 (32%) of 84 without perineal sensation were living at follow-up ( $p<0.001$ ). Renal-related disease accounted for 19 of 57 deaths in those without perineal sensation; no renal-related deaths occurred in those with perineal sensation (19/84 vs 0/33,  $p=0.003$ ). Among survivors, those with perineal sensation were more likely to be continent of urine and feces, able to walk at least 50 m, and never to have had pressure sores. (Oakeshott P, Hunt GM, Whitaker RH, Kerry S. Perineal sensation: an important predictor of long-term outcome in open spina bifida. *Arch Dis Child* Jan 2007;92:67-70). (Respond: Dr P Oakeshott, Community Health Sciences, St George's, University of London, London SW17 0RE, UK; E-mail: oakeshot@sgul.ac.uk).

COMMENT. The neurologic examination, specifically perineal sensation, in infants born with open spina bifida is a simple and practical method for prediction of long-term outcome. Infants with intact sensation in the saddle area were more likely to survive, to have normal renal function, to be continent of urine and feces, ambulant, and not have pressure sores. It is refreshing to read of studies based on the clinical acumen of the neurologist rather than invasive machines.

## INFECTIOUS DISORDERS

### **FRONTAL LOBE MRI ABNORMALITIES IN HHV-6 ENCEPHALOPATHY**

Magnetic resonance imaging and SPECT findings in 10 children with a diagnosis of human herpesvirus 6 (HHV-6) encephalopathy are reported from Jikei University School of Medicine, Tokyo; and Saitama Children's Medical Center, Japan. T1 and T2-weighted MR imaging showed no abnormalities, but diffusion-weighted imaging showed abnormal hyperintensity in the subcortical white matter of the frontal lobes in 6 patients tested during the acute phase. Cerebral blood flow single-photon emission CT revealed decreased perfusion in the frontal lobes of all 9 tested. These findings with localization to the frontal lobes may be specific for HHV-6 encephalopathy. (Yoshinari S, Hamano S, Minamitani M, Tanaka M, Eto Y. Human herpesvirus 6 encephalopathy predominantly affecting frontal

lobes. **Pediatr Neurol** Jan 2007;36:13-16). (Respond: Satoshi Yoshinari MD, Division of Neurology, Saitama Children's Medical Center, 2100 Magome, Iwatsuki-ku, Saitama-city, Saitama 339-8551, Japan).

COMMENT. HHV-6 infection is most commonly associated with febrile seizures; it accounts for one-third of all first-time cases in children up to 2 years of age (Hall CB et al. **N Engl J Med** 1994;331:432-438). These authors detected HHV-6 DNA by PCR in 2 of 7 CSF samples of HHV-6 associated febrile seizure patients. In 9 patients with acute infantile encephalopathy predominantly affecting the frontal lobes, the associated infection was influenza type A in 3, HHV-6 in 2, measles in 1, and upper respiratory viral illness unspecified in 3. (Yamanouchi H et al. **Pediatr Neurol** 2006;34:93-100; **Ped Neur Briefs** March 2006;20:20-21). Serial MRIs showed atrophic changes in both frontal lobes, and PET studies revealed decreased perfusion in frontal lobes, with normalization at 7<sup>th</sup> to 38<sup>th</sup> month after onset. Prolonged impairment of consciousness after convulsions, and behavioral sequelae were consistent with an encephalopathy. These cases are unique in the predilection for the frontal lobes, but they are not HHV-6 specific.

## HERPES SIMPLEX VIRUS INFECTIONS IN PRETERM INFANTS

A retrospective review of herpes simplex virus (HSV) infections within the first 30 days after birth in infants born at <37 weeks was conducted at Johns Hopkins University School of Medicine, Baltimore, MD. Ten preterm singletons and a set of twins were infected with HSV-2 and presented with either disseminated disease (n=9) or encephalitis (n=3). All infants with disseminated disease died, whereas the 3 with encephalitis survived. All developed respiratory distress, and viral cultures were positive. Ten were treated with acyclovir within 48 hours of onset; 2 of 3 treated with high-dose acyclovir (60 mg/kg/day) survived. (O'Riordan DP, Golden WC, Aucott SW. Herpes simplex virus infections in preterm infants. **Pediatrics** Dec 2006;118:e1612-e1620).

COMMENT. HSV infections in preterm infants present with respiratory distress and a high incidence of disseminated disease. In this population, response to acyclovir is poor and mortality high. The AAP Red Book (2006;p365) states that most neonates treated for HHV encephalitis survive, but most suffer substantial neurologic sequelae. Approx 25% of neonates with disseminated disease die despite antiviral therapy.

## NEUROMUSCULAR DISORDERS

### CSF MARKERS IN GUILLAIN-BARRE SYNDROME

A positive 14-3-3 protein assay of CSF was observed in 29 of 38 patients with GBS and in 4 with motor neuron disease and other neuropathies studied at Universities of Milan and Verona, Italy. The protein is detected as early as 12 to 48 hours after disease onset and represents a useful biological marker in GBS. (Bersano A, Fiorini M, Allaria S et al. Detection of CSF 14-3-3 protein in Guillain-Barre syndrome. **Neurology** Dec 2006;67:2211-2216). (Respond: Dr S Monaco; e-mail: salvatore.monaco@univr.it).