MUSCLE DISORDERS

ACUTE RECTUS PALSY AND MYOSITIS

Orbital myositis as the cause of palsy of the extraocular rectus muscle is reported in 7 children presenting with acute ocular pain at the Scottish Rite Children's Hospital, Atlanta, GA. All had chemosis and erythema of the conjunctiva restricted to the quadrant overlying the involved muscle. All had ocular pain and some had redness and swelling of the lids with ptosis. All were afebrile. The diagnosis was confirmed by CT demonstration of an enlarged lateral rectus muscle. All had a benign course and were immediately responsive to corticosteroids. A recurrence in 2 patients was attributed to abrupt withdrawal of steroids. (Pollard ZF. Acute rectus muscle palsy in children as a result of orbital myositis. J Pediatr February 1996;128:230-3). (Reprints: Zane F Pollard MD, 5455 Meridian Mark Road, Suite 220, Atlanta, GA 30342).

COMMENT. The differential diagnosis includes orbital cellulitis which is distinguished by fever and response to antibiotics. Reported isolated causes of orbital myositis include Lyme disease, cysticercosis, and a paraneoplastic syndrome.

IV IMMUNOGLOBIN THERAPY IN DERMATOMYOSITIS

Improved strength and functional abilities following IV immunoglobulin treatment for chronic dermatomyositis is reported in two children from the University of Mississippi Medical Center, Jackson, MS. Both patients had developed side effects during prior treatment with prednisone and immunosuppressive agents. The response to IVIG was slow and occurred in a stepwise fashion after repeated monthly courses (2 g/kg). The rash on the face and hands also resolved. (Vedanarayanan V et al. Treatment of childhood dermatomyositis with high dose intravenous immunoglobulin. Pediatr Neurol 1995;13:336-339). (Respond: Dr Vedanarayanan, Division of Pediatric Neurology, University of Mississippi Medical Center, Jackson, MS 39216).

COMMENT. The authors consider IVIG a useful adjuvant therapy for dermatomyositis, permitting reduction in steroid dosage and lessening of treatment morbidity.

SUBARACHNOID AND VENTRICULAR CSF DISORDERS

INFANTILE MACROCRANIA AND SUBARACHNOID FLUID

Macrocrania caused by subarachnoid fluid collections (SFC) in 12 very low birth weight (VLBW) infants is reported from the Department of Pediatrics, University of Manitoba, and the Newborn Follow-up Program, Health Sciences Centre, Winnipeg, Manitoba, Canada. Ultrasound had shown grade II and III intraventricular hemorrhages in 7 infants in the neonatal period. The prevalence of SFC in VLBW infant survivors attending this clinic was 2.6%. SFC accounted for 30% of cases of macrocrania in VLBW infants. The incidence of SFC was 3.3 per 1000 VLBW survivors annually. The occipitofrontal circumference was at 5 to 50th percentile at birth and >95th percentile at age of diagnosis (mean, 7.7 months). A frontal subarachnoid space 6 mm or more, an interhemisperic fissure 8mm or more, and normal ventricles on ultrasound were required for diagnosis of SFC. Head growth

stabilized along a curve above and parallel to the 95th percentile by age 15 to 18 months. Transient neurodevelopmental abnormalities (hypertonia and hyperreflexia) found in 5 infants at 6 to 8 months had disappeared by 18 months, and all had normal findings at 25 month follow-up. No infant had cerebral palsy or retardation <70, and none required neurosurgery. (Al-Saedi SA et al. Subarachnoid fluid collections: a cause of macrocrania in preterm infants. I Pediatr February 1996;128:234-6). (Reprints: Oscar G Casiro MD, FRCPC, Director, Newborn Follow-up Program, Children's Hospital, 840 Sherbrook Street, Winnipeg, Manitoba R3A 1S1, Canada).

COMMENT. Various names used to describe benign SFC include "external hydrocephalus," and "benign subdural collection of infancy." If sonograms are normal except for SFC the infant may be followed with periodic head circumference measurements and clinical evaluation. An abnormal ultrasound may require follow-up with CT. The prognosis is usually favorable without intervention. Fukuyama Y et al have provided norms for age-specific CT measurements of subarachnoid spaces up to 1 year. (Dev Med Neurol 1979;21:425). The abnormal measurements by ultrasound used for diagnosis of SFC in the above study exceeded the Fukuyama upper limits of CT norms: >5.7mm sa spaces and >7.6 mm ih fissures.

Cerebral ventricular enlargement in female adolescents with anorexia nervosa corelated with the degree of malnutrition and returned to normal after refeeding and weight gain in an MRI quantitative study at the Schneider Children's Hospital, Albert Einstein College of Medicine, New Hyde Park, NY. (Golden NH et al. I Pediatr February 1996;128:296-301). Body mass and ventricular volume were inversely correlated.

SEIZURE DISORDERS

PREDICTORS OF INTRACTABLE EPILEPSY

Risk factors for intractable epilepsy at the time of initial diagnosis were determined in a case-control study at Yale University, New Haven, CT. Children with an average of one seizure or more per month over a 2-year period and refractory to at least 3 different AEDs were compared to controls who had been seizure-free for >2 years and had never had intractable epilepsy. Independent predictors of intractability were infantile spasms, early age of onset, remote symptomatic epilepsy, and status epilepticus. (Berg AT et al. Predictors of intractable epilepsy in childhood: a case-control study. Epilepsia 1996;37:24-30). (Reprints: Dr AT Berg, School of Allied Health Professions, Program in Community Health, DeKalb, IL 60115).

COMMENT. Age at onset was the predominant predictor of seizure intractability in this study, even after controlling for infantile spasms as a cause. Prognosis was progressively better with increasing age at onset during childhood and adolescence. For reports of neuropathology associated with intractable epilepsy, see Progress in Pediatric Neurology I, 1991, pp 131-138; and II, 1994, pp 129-141.