

NEUROMUSCULAR DISORDERS

MANAGEMENT OF BRACHIAL PLEXUS INJURIES

The results of early neurosurgical treatment of 58 infants with various types of brachial plexus birth injury have been compared with non-surgical intervention in 91 patients followed by a multidisciplinary team at the Brachial Plexus Program, Miami Children's Hospital, FL. In the non-surgical group, three subsets of patients' long-term outcomes were described: Type I (n=12) patients with global palsy (C5/C6/C7) had extensive limitations with minimal functional use; type II (n=63) infants with typical Erb's palsy (C5/C6) showed no significant limitations in hand and wrist function and mild limitations in shoulder range of motion; and type III (n=16) infants with typical Erb's palsy at birth had a persistent deformity and functional loss at long-term follow-up. Types I and III with a poor outcome following non-surgical therapy were compared with a similar cohort of patients treated surgically between 3 and 5 months of age. In 22 C5/C6 cases available for a minimum 3-year follow-up, the results were graded as fair in 4.5%, satisfactory in 13.6%, good 26.6%, and excellent 45.4%. For 30 C5/C6/C7 cases, ratings were fair in 3.3%, satisfactory in 30%, good 56.6%, and excellent in 10%. Late nerve reconstruction between 12 and 32 months of age in more than 40 patients presenting after 1 year of age with persistent shoulder sequelae was also beneficial in selected infants. None had surgical complications. Secondary musculoskeletal procedures to maximize extremity function were necessary in 25% of children undergoing nerve reconstruction. Rehabilitation therapy, orthotic and bracing techniques, neuromuscular electrical stimulation, and botulinum toxin A are considered important ancillary modalities. (Grossman JAI, DiTaranto P, Price AE et al. Multidisciplinary management of brachial plexus birth injuries: the Miami experience. **Seminars in Plastic Surgery** 2004;18(4):319-326). (Reprints: Dr John AI Grossman, 8940 N Kendall Drive, Miami, FL 33176).

COMMENT. In a multidisciplinary specialized center, surgical intervention between 3 and 5 months of age can improve functional outcome in selected patients with brachial plexus birth injury.

A review of 186 patients with obstetrical brachial palsy at Children's National Medical Center, Washington, DC, found that in 46 with typical Erb's palsy graded as moderate in severity at <3 months, 28 (61%) had persistent functional limitations at long-term follow-up. Complete recovery was exceptional and mild sequelae were the rule. (Eng GD et al. **Muscle Nerve** 1996;19:884-891; **Ped Neur Briefs** July 1996).

PREDNISONE THERAPY FOR DUCHENNE DYSTROPHY

The effects of prednisone on muscle function and the extent of steroid-related adverse effects were studied in 17 ambulant children with Duchenne muscular dystrophy (DMD) at University Hospital, Groningen; Rehabilitation Centre, Utrecht; and Leiden University Medical Centre, the Netherlands. In a randomized, placebo-controlled, crossover trial with 6 months of treatment, the time needed to run 9 m and to climb 4 stairs was significantly lower during prednisone treatment, and the quality of life was not affected. Short-term prednisone

treatment is recommended to preserve motor function in ambulant children with DMD. (Beenakker EAC, Fock JM, Van Tol MJ et al. Intermittent prednisone therapy in Duchenne muscular dystrophy. **Arch Neurol** Jan 2005;62:128-132). (Respond: Ernesto AC Beenakker MD, Department of Neurology, University Hospital Groningen, Hanzeplein 1, 9700 RB Groningen, the Netherlands).

COMMENT. In previous studies of the effects of prednisone in DMD, a significant increase in muscle strength, pulmonary function, and functional ability has been demonstrated in a randomized controlled trial in 99 boys treated at the University of Rochester, NY (Griggs RC et al. **Arch Neurol** 1991;48:383-388; **Ped Neur Briefs** April 1991). Improvement was rapid, occurring in 10 days and was maximal after 2 months. A dose of 0.75 mg/kg/day was recommended in patients who experience functional decline. In another report (Fenichel GM et al. **Neurology** 1991;41(Suppl 1):166), the beneficial effects of prednisone, 0.65 mg/kg/day, extending over a two year observation period were demonstrated in 89 boys with DMD. See **Ped Neur Briefs** June 1991 for further articles on prednisone in DMD and its effect on immunological mechanisms.

CEREBRAL MALFORMATIONS

SPECTRUM OF CORPUS CALLOSUM AGENESIS

Magnetic resonance imaging and clinical features of 16 children with agenesis of the corpus callosum (ACC) are reviewed at UAE University, United Arab Emirates. Three groups of patients were recognized: Group 1). 8 patients without involvement of other brain areas, 3 having lipomas, or interhemispheric cyst, 3 with partial ACC, 5 with Probst bundles (aberrant longitudinal fiber bundles on medial hemispherical walls), and 4 with epilepsy; Group 2). 4 patients with ACC complicated by severe telencephalic dysgenesis (periventricular heterotopia, microcephaly with cortical dysplasia, microlissencephaly and cerebellar hypoplasia, diffuse agyria-pachygyria and pontocerebellar dysplasia), absent Probst bundles, and developmental delay and epilepsy in 3; Group 3). 4 children with ACC as part of a syndrome: 1 with Aicardi syndrome, 2 with L1 disease and mutations in the L1CAM gene, and 1 with Mowat-Wilson syndrome with mutation in the zinc finger homeo box 1B (ZFX1B) gene; all 4 had severe developmental delay and mental retardation and 1 had infantile spasms. Severe handicaps, developmental delay, mental retardation, and neurologic deficit occurred in patients of groups 2 and 3. Two of the 8 patients in group 1 were developmentally delayed, mentally retarded, and had neurologic deficits. Parental consanguinity was present in 7 of the 16 patients, and genetic factors are involved in ACC cases associated with various syndromes. (Sztirha L. Spectrum of corpus callosum agenesis. **Pediatr Neurol** February 2005;32:94-101). (Respond: Dr Sztirha, Department of Pediatrics, Faculty of Medicine and Health Sciences, UAE University, POB 17666, Al Ain, United Arab Emirates).

COMMENT. MRI is important in the evaluation of developmental delay or epilepsy. ACC is also recognized on prenatal ultrasound after the 20th week. Dobyns describes true types and secondary types of ACC (**Am J Hum Genet** 1996;58:7-16). Those associated with major malformations or degeneration are considered secondary.