

PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW

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Vol. 14, No. 4

April 2000

NEUROMUSCULAR DISORDERS

OBSTETRIC BRACHIAL PALSY RECOVERY MECHANISM

A group of 162 infants (aged 4-14 months) with obstetric brachial palsy (OBP) and a group of 184 child and adult patients (aged 6-74 years) with a traumatic brachial plexus lesion were compared, using an EMG and nerve-conduction-study protocol, developed since 1985 at the Department of Clinical Neurophysiology, Atrium Medisch Centrum, Heerlen, The Netherlands. Subjects selected for comparison had complete avulsion of both roots C5 and C6 and/or complete rupture of the upper trunk verified during operation. Fourteen infants (aged 4 months) with OBP, and 19 adults (aged 16-30 years) met selection criteria. The infants had a nearly normal recruitment pattern of motor units in the biceps brachii and deltoid muscles and little or no denervation, while the same lesion in adults caused complete denervation of both muscles. In three infants with OBP who had a more extensive lesion involving also C7 or rupture of the middle trunk, almost complete denervation of both muscles was observed. At birth, C7 contributes to the biceps and deltoid innervation, while in adults this innervation is lost by apoptosis. Spontaneous recovery of OBP in infants results from an intact C7 innervation and remodelling of central pathways. (Vredeveld JW, Blaauw G, Sloof BACJ, Richards R, Rozeman SCAM. The findings in paediatric obstetric brachial palsy differ from those in older patients: a suggested explanation. Dev Med Child Neurol March 2000;42:158-161). (Respond: Dr Jan W Vredeveld, Department of Clinical Neurophysiology, Atrium Medisch Centrum, Heerlen, Postbus 4446, 6401 CX Heerlen, The Netherlands).

COMMENT. This study involved only patients with surgically verified complete avulsion of C5 and C6 and/or rupture of the upper trunk of the brachial plexus. In adults, the biceps and deltoid muscles are innervated by C5 and C6 roots, whereas in infants their innervation also relies on an intact C7 root. Recovery from Erb's palsy due to obstetric injury is explained by an intact C7 innervation and central reinnervation of the biceps and deltoid muscles. Loss of this supplementary innervation by apoptosis may explain the persistence of paralysis found in adult-acquired traumatic brachial plexus injury. The authors report a

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poor correlation between EMG and nerve conduction studies and outcome in infants with obstetric brachial palsy. Surgery was considered in infants with OBP when no spontaneous motor recovery was clinically evident after 3 to 4 months. See Progress in Pediatric Neurology III, PNB Publ, 1997;pp357-359, for further comment on outcome of OBP following conservative management, and interpretation of EMG fibrillation potentials in newborns.

Outcome in OBP with and without surgery. Investigators at the Department of Neuropediatrics, Karolinska Hospital, Stockholm, examined the functional outcome at age 5 years of 247 children with OBP, with and without microsurgical reconstruction. In the operated group, improved shoulder range of movement in C5-6 palsies was the only benefit, compared to non-operated patients, and outcome was not correlated with the timing of operation, before or after age 6 months. A decrease in grip and bimanual function in C5-6 palsies was unexpected. Those with widespread C5-T1 lesions had the most root avulsions and the poorest hand function. Nerve reconstruction in the majority of infants with upper OBP lesions should be delayed until after 6 to 9 months. (Strombeck C, Krumlinde-Sundholm L, Forrsberg H. Dev Med Child Neurol March 2000;42:148-157).

CONGENITAL MYOPATHY WITH APOPTOTIC CHANGES

A case of congenital myopathy with myonuclear changes consistent with apoptotic degeneration in a 4-year-old girl is reported from the National Institute of Neuroscience, Tokyo, Japan. She was hypotonic at birth and psychomotor development was markedly delayed. Brain MRI was normal. DNA fragmentation in myonuclei was demonstrated by the TUNEL method and supported by ultrastructural characteristics and immunochemistry. (Ikezoe K, Yan C, Momoi T et al. A novel congenital myopathy with apoptotic changes. Ann Neurol April 2000;47:531-536). (Respond: Dr Ikezoe, Department of Ultrastructural Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Tokyo 187-8502, Japan).

COMMENT. The authors add a new type of structural abnormality associated with congenital myopathy. Other types include central core disease, nemaline myopathy, myotubular myopathy, and congenital fiber-type disproportion.

HIGH-DOSE IV IMMUNOGLOBULIN IN JUVENILE MYASTHENIA

Ten children (median age, 13 years; range, 3-18 yrs) with juvenile myasthenia gravis were treated with high-dose intravenous immunoglobulin (2 gm/kg body wt) and prospectively evaluated for 5 years at the Department of Neurology, Wayne State University, Children's Hospital of Michigan, Detroit, MI. Improvement in functional strength occurred initially in 8 patients, but the response decreased after multiple monthly treatments. No correlation was noted between clinical response and a decrease in anti-AChR antibody levels measured in 3 patients. One child developed hypotension during the infusion and treatment was discontinued. Six patients complained of headache. (Selcen D, Dabrowski ER, Michon AM, Nigro MA. High-dose intravenous immunoglobulin therapy in juvenile myasthenia gravis. Pediatr Neurol 2000;22:40-43). (Respond: Dr Nigro, 28595 Orchard Lake Rd, Farmington Hills, MI 48334).

COMMENT. High-dose IV immunoglobulin may be of benefit in juvenile myasthenia gravis as a short-term therapy, at times of myasthenic crisis, and in preparation for surgery.