Acta Paediatr 2002;91:617-625). (Respond: C Hager-Ross, Department of Community Medicine and Rehabilitation, Section for Physiotherapy, Umea University, SE-901 87 Umea, Sweden).

COMMENT. The tables and data of grip strength provided in this article permit comparisons of a patient's score with those of normally developed children according to age, gender, handedness and body weight, height and hand length. Right-handed children may be expected to be 10% stronger with the right hand while left-handers are equally strong in right or left. Boys are stronger than girls, but only over 10 years of age. Grip strength is directly correlated with hand length and body weight.

CORTICAL MALFORMATIONS

GENETICS AND PRENATAL INJURY IN CORTICAL MALFORMATIONS

The interrelationship of genetics and prenatal injury in the genesis of malformations of cortical development (MCD) was studied at the University of Campinas, SP, Brazil. In a series of 76 consecutive patients with MCD, 21 (28%) had focal cortical dysplasia, 19 (25%) had heterotopias or agyria-pachygyria, and 36 (47%) had polymicrogyria or schizencephaly. In the group with heterotopias, 6 (32%) had a family history of MCD, mental retardation, or miscarriages, suggesting a genetic factor in etiology. In the group with polymicrogyria, 5 (14%) had a family history of MCD. Prenatal events had occurred in 28 (37%) of the total series and only 2 of controls (5%); they were significantly more frequent in the patients with heterotopias and polymicrogyria (P<.001). Epilepsy occurred in all patients with focal cortical dysplasia, in 89% of the heterotopia group, and less frequently (P<.001) in patients with polymicrogyria (47%). Epilepsy associated with polymicrogyria was more easily controlled than in other forms of MCD. (Montenegro MA, Guerreiro MM, Lopes-Cendes I, Guerreiro CAM, Cendes F. Interrelationship of genetics and prenatal injury in the genesis of malformations of cortical development. Arch Neurol July 2002;59:1147-1153). (Reprints: Marilisa M Guerreiro MD PhD, Department of Neurology, University of Campinas, PO Box 6111, 13083-970 Campinas, Sao Paulo, Brazil).

COMMENT. The variable clinical manifestations encountered with different forms of MCD are determined by a combination of genetic and prenatal factors. The more frequent and severe epilepsy associated with focal cortical dysplasia is less frequently related to genetic and prenatal factors, whereas the less frequent and milder epilepsy common to the polymicrogyria group has a stronger association with genetic and prenatal events. Heterotopias are frequently linked to genetic predisposition.

MUSCLE DISORDERS

INHERITANCE OF CONGENITAL MYASTHENIC SYNDROMES

Two novel slow-channel congenital myasthenic syndromes (SCCMS) with mutations in the AChR e subunit are reported from the John Radcliffe Hospital, Oxford, UK. In two of three kinships, the syndrome showed an atypical recessive inheritance pattern. Typically SCCMS has a dominant inheritance. In Pedigree 1, the index patient presented at 29 years of age with failure to breathe after a general anesthetic. Her parents were consanguineous. Examination revealed

bilateral ptosis, weakness of eye closure, facial muscles, shoulder, hand and hips. In Pedigree 2, the index patient developed ptosis at 17 years, and weakness progressed to other muscle groups over the next 7 years. Pedigree 3 index patient noted transient weakness of finger extensors in her twenties, which later progressed to mild involvement of wrist, neck and other muscles. Antibodies to AChR were absent in all three patients and there was no response to anticholinesterase treatment. EMG showed decrement of the CMAP and a repetitive response to a single nerve stimulus. Only Pedigree 3 showed a typical dominant inheritance pattern. (Croxen R, Hatton C, Shelley C et al. Recessive inheritance and variable penetrance of slow-channel congenital myasthenic syndromes. Neurology July (2 of 2) 2002;59:162-168). (Reprints: Dr David Beeson, Neurosciences Group, Weatherall Institute of Molecular Medicine, The John Radcliffe, Headington, Oxford OX3 9DS, UK).

COMMENT. Slow-channel congenital myasthenic syndromes are typically of dominant inheritance and caused by missense mutations in the muscle nicotinic acetylcholine receptor (AChR). Symptoms are present at birth or may be delayed until adulthood. Fatigable muscle weakness, selectively involving neck, shoulder and finger extensors, is mild or severe and tends to be slowly progressive. Response to anticholinesterase treatment is absent, and EMG shows a double response to a single nerve stimulus. Adding to the 11 previously described mutations underlying the SCCMS, the Oxford team describes two new mutations in the e subunit, with symptoms present only in the index patient, and the first reported examples of recessively inherited SCCMS.

ANOXIC DISORDERS

OUTCOME FACTORS IN HYPOXIC ISCHEMIC ENCEPHALOPATHY

The predictive value of history, examination, Glasgow Coma Scale (GCS) scores, EEG and sensory evoked potentials (SEP) in the prognosis of children with acute hypoxic-ischemic encephalopathy (HIE) was evaluated at the University Hospital of Lille, France. Of 53 consecutive children who were mechanically ventilated for HIE, 12 had died at 24 hours after admission, 3 were awake, and 42 showed impaired consciousness or were in coma (GCS <8).

In the 42 with uncertain prognosis, outcome was good in 12 and mild or moderate disability in 4 patients (a favorable outcome in 38%), and severe disability in 7 patients; 19 ultimately died. Predictors of an unfavorable outcome included: 1) an initial cardiopulmonary resuscitation duration longer than 10 minutes; 2) a GCS <5 at 24 hrs after admission; 3) EEG showing a discontinuous pattern and spikes or epileptiform pattern; and 4) bilateral absence of the N20 wave on SEPs. (Mandel R, Martinot A, Delepoulle F et al. Prediction of outcome after hypoxic-ischemic encephalopathy: a prospective clinical and electrophysiologic study. J Pediatr July 2002;141:45-50). (Reprints: Pr F Leclerc MD, Pediatric Intensive Care Unit, Hopital Jeanne de Flandre, CHRU, 2 place O, Lambret, 59037 Lilli9 Cedex, France).

COMMENT. At 24 hours after birth, clinical signs, the GCS, EEG and SEPs permit early prediction of prognosis of children with HIE.

Microcephaly after HIE may be predicted by serial head circumference measurements between birth and 4 months of age. A decrease in HC ratios of >3.1% by 4 months correlates with development of microcephaly and neurologic sequelae before 18 months. (see Progress in Pediatric Neurology III, 1997;p396).