

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  $\epsilon$  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