The Wada test and its possible replacement with fMRI are discussed in an editorial. (Abou-Khalil B, Schlagger BL. Is it time to replace the Wada test? Neurology July (2 of 2) 2002;59:160-161). Only "when a paradigm or battery of fMRI paradigms can be validated for identification of language regions that should be excluded from excision, and for postoperative memory function, We are almost there."

DEVELOPMENTAL DISORDERS

DEVELOPMENTAL LANGUAGE DISORDER AND POLYMICROGYRIA

Neuroimaging studies were conducted in 15 children (11 boys), ages 4 to 14 years, with primary complaint of language delay, in a study at the Department of Neurology, University of Campinas, Brazil. Six with severe developmental language disorder (DLD) had diffuse perisylvian polymicrogyria (PMG): they did not speak or had mixed phonologic-syntactic deficit syndrome. Six with PMG restricted to posterior parietal regions had milder DLD, a phonologic programming deficit syndrome (excessive use of jargons). The clinical manifestations of DLD associated with polymicrogyria vary in relation to the extent of the cortical abnormality. (Guerreiro MM, Hage SRV, Guimaraes CA et al. Developmental language disorder associated with polymicrogyria. Neurology July (2 of 2) 2002;59:245-250). (Reprints: Dr Marilisa M Guerreiro, Department of Neurology, PO Box 6111, 13083-970 Campinas, SP, Brazil).

COMMENT. Polymicrogyria involving perisylvian or temporoparietal regions may be associated with developmental language disorder (DLD) or developmental dyslexia. The complete perisylvian syndrome is manifested by pseudobulbar palsy, cognitive deficits, epilepsy, and MRI evidence of cortical abnormalities. In the above study, patients diagnosed with DLD and polymicrogyria had normal or borderline cognitive function and no history of epilepsy. A subtle form of posterior parietal polymicrogyria can present with DLD and represents a mild form of perisylvian syndrome.

MOTOR DEVELOPMENT AND HANDEDNESS

NORMAL GRIP STRENGTH IN YOUNG CHILDREN

Norms for grip strength of children aged 4 to 16 years were determined in a study of 530 Swedish children at the Department of Rehabilitation, Umea University, Sweden. Peak grip strength over a 10 sec period and sustained grip strength averaged across the 10 secs were measured using an instrument Grippit dynamometer, with adjustable handles and digital display. A standardized position for the child was followed as recommended by the American Society of Hand Therapists. Up to 10 years of age, boys and girls showed increases in strength with age that were parallel; after 10 years, boys were significantly stronger than girls (55% higher at 16 years). Grip strength was strongly correlated with weight, height, and especially, hand length. Right-handed children were significantly stronger (up to 10%) in the dominant hand, while left-handers showed no difference between hands. Sustained grip strength was 80-85% of peak grip strength; it was lower in younger children. These norms for peak grip strength were slightly lower than the USA and Australia 1980s data, probably related to differences in instruments used and age groupings of children in the studies. (Hager-Ross C, Rosblad B. Norms for grip strength in children aged 4-16 years.

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