

LLI children have been found to have a temporal processing deficit, expressed by limited identification of brief phonetic elements of speech and impaired sequencing of short-duration acoustic stimuli presented in rapid succession. When presented in slower forms and rates, the stimuli are correctly perceived and receptive language is improved. Temporal processing deficits of 11 LLI children studied at the Center for Integrative Neurosciences and Coleman Laboratory, University of California, San Francisco, were corrected by adaptive training exercises and computer games designed to modify temporal processing skills. (Merzenich MM, Tallal P et al. Temporal processing deficits of language-learning impaired children ameliorated by training. Science Jan 5, 1996;271:77-81).

COGNITIVE AND SPEECH DEFICITS WITH OPSOCLONUS-MYOCLONUS

Thirteen patients, aged 1.7 to 16 years, with opsoclonus-myoclonus syndrome were evaluated for neuropsychological, psychosocial and adaptive function at the Children's National Medical Center, George Washington University, Washington, DC. IQs of six older children ranged from 50 to 72 on the Wechsler scales. One infant had a Mental Index of 71 on the Bayley, and a 46-month-old child tested at the 20-month level. Severe problems related to motor output, involving ambulation, fine motor coordination and speech, while some age-appropriate cognitive skills were retained. Verbal and visual reasoning approached the borderline to normal range. On the Achenbach Child Behavior Checklist, mild to moderately severe behavioral irritability and emotional lability were reported in 8 of 12 non-medicated children. On Vineland Adaptive Behavior Scales, severe adaptive limitations were noted; self-care was significantly delayed in areas related to feeding, dressing and toileting. Motor problems contributed to low scores in daily living and communication scales. One child, aged 8 years examined while treated with ACTH, tested in the normal range. (Papero PH, Pranzatelli MR et al. Neurobehavioral and psychosocial functioning of children with opsoclonus-myoclonus syndrome. Dev Med Child Neurol 1995;37:915-932). (Respond: Dr Patricia H Papero, Department of Psychiatry, Children's National Medical Center, 111 Michigan Ave NW, Washington DC 20010).

COMMENT. In 27 patients with opsoclonus-myoclonus syndrome reported previously from the same center (Pranzatelli et al. 1995, see Ped Neur Briefs March 1995;9:19), the etiology was paraneoplastic in 46% and infectious in the remainder. Hammer, Larsen, and Stack, at Children's Memorial Hospital, Chicago, have reported on the developmental outcome of 11 children with opsoclonus-myoclonus syndrome, the majority having an associated neuroblastoma. (See Ped Neur Briefs August 1995;9:61). Delayed development with motor incoordination and speech delay occurred in 8 patients and 3 had behavioral problems. Development was normal in 2 of 3 patients without neuroblastoma and in only 1 of 8 whose opsoclonus was associated with neuroblastoma. While the majority of patients in this study had significant developmental delay, others have reported a 50% incidence of intellectual deficit. The poor outcome might possibly be related to etiology, but symptoms of ataxia and opsoclonus were not improved by removal of a neuroblastoma.

CORTICAL MOTOR OUTPUT OF BRAILLE READING HAND

Focal transcranial magnetic stimulation (TMS) was used to map the

motor cortical outputs to the right and left first dorsal interosseous (FDI) and right abductor digiti minimi (ADM) muscles of the reading hand in 6 blind proficient Braille readers studied at the National Institute of Neurological Disorders and Stroke, NIH, Bethesda, MD. All subjects had learned Braille before age 13, using the right index finger for character recognition and the left index for line keeping. Comparison of cortical output maps obtained on working days and off days showed that the maps for the FDI of the reading hand were significantly larger in the evening after the working shift than in the morning after having not worked for 2 days. The map shrunk following vacation days and enlarged following a return to work. On control days, the motor threshold for the muscles stayed constant, whereas on the work day, the motor thresholds for the right FDI were significantly lower in the evening study session after practice than in the morning test before practice. The results illustrate the rapid modulation in motor cortical outputs effected by Braille reading. (Pascual-Leone A, Hallett M et al. The role of reading activity on the modulation of motor cortical outputs to the reading hand in Braille readers. Ann Neurol December 1995;38:910-915). (Respond: Dr Pascual-Leone, Unidad de Neurobiología, Departemento Fisiología, Universidad Valencia, Avda Blasco Ibanez 17, Valencia 46010, Spain: or Dr Hallett, NINDS, NIH, Bldg 10, Rm 5N226, 10 Center DR MSC 1428, Bethesda, MD 20892).

COMMENT. The authors emphasize the critical importance of timing when looking for changes in neural networks associated with learning skills. Learning a new skill requires plastic changes and rapid modulation of intracortical connections that result in temporary enlargement of the cortical motor output. These neurophysiological changes are supported by PET scanning studies in the same laboratory, showing that Braille reading is associated with an increased activation of the sensorimotor and striate cortex contralateral to the reading hand. (Sadato N et al. 1995).

INTELLIGENCE AND MRI CHANGES IN NEUROFIBROMATOSIS

Brain MRI changes were studied in relation to intelligence in a group of 28 children, aged 4 to 16 years, with neurofibromatosis 1. The mean FS IQ on the Wechsler scales (WPPSI, WISC-R) was 89 (range 54-148), mean VS was 95 and PS 84. Eight children who had neurological disease (epilepsy, hydrocephalus, tumor, or post-irradiation) scored significantly lower on IQ tests than the 20 without neurological disease. Eighteen children with hyperintense T2 weighted foci had a mean FS IQ comparable with the 10 without spots. In the group without neurologic disease, the 10 showing hyperintense spots had a higher mean FS IQ than 10 without. There was no significant relation between the number or location of T2 weighted foci and FS IQ. (Legius E et al. Neurofibromatosis type 1 in childhood: correlation of MRI findings with intelligence. J Neurol Neurosurg Psychiatry December 1995;59:635-640). (Respond: Dr Eric Legius, Center for Human Genetics, University Hospital Gasthuisberg, Herestraat 49, 3000 Leuven, Belgium).

COMMENT. Previous studies have shown that the mean full scale IQ in children with neurofibromatosis 1 is shifted to the left, between 88 and 94. T2 weighted hyperintense foci in the brain MRI were not correlated with intelligence of children with neurofibromatosis 1 in the present study, and similar results have been reported from other centers. (see Progress in Pediatric Neurology 1, PNB Publishers, 1991, pp376-77). However, the authors cite two recent reports showing a significant correlation between a lowered IQ