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LANGUAGE AND LEARNING DISORDERS

DEVELOPMENTAL DYSPHASIA

The neuropathological findings in a seven year old girl with developmental dysphasia who died of complications of infectious mononucleosis are reported from the Departments of Neurology and Pediatrics and the Department of Pathology, Medical College of Georgia, Augusta, GA. The child had been followed for developmental dysphasia and attention disorder with hyperactivity and had been treated with methylphenidate and behavior modification. Her birth was normal, she walked at 17 months and language milestones were significantly delayed with first words at two years and short phrases at four years of age. Her six year old brother had developmental dyslexia of the dysphonetic subtype. Speech and language evaluations at 2 yrs 10 mo showed no expressive language and wishes were communicated through pointing or gesturing. Receptive language function was at an 18 month level and play audiometry revealed normal hearing. The neurological examination showed inconstant asymmetry of deep tendon reflexes and a questionable Babinski on the left side. The head circumference was at the 20th percentile. Her intelligence level by the "WPPSI" was 70, expressive vocabulary 70 and receptive vocabulary 72. In addition to the language disorder she had a dysfunction in short term auditory memory and short term visual memory. In contrast her visual spatial perception and construction were relatively strong. At six years of age she did not have number or letter recognition and was unable to write her name. Arithmetically, she could count to four by rote and demonstrated number concepts to two. Academically a global learning disability was present. The neuropathological studies revealed atypical symmetry of the plana temporale and a dysplastic gyrus on the inferior surface of the left frontal cortex along the inferior surface of the Sylvian fissure. The authors proposed that these anomalies are likely related to midgestation, the period of neuronal migration from the germinal

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matrix to the cerebral cortex and are consistent with a neurodevelopmental cause of developmental dysphasia. (Cohen M et al. Neuropathological abnormalities in developmental dysphasia. Ann Neurol June 1989; 25: 567-570).

COMMENT. The Child Neurology Society Task Force on the nosology of disorders of higher cerebral function in children (1981) defined developmental dysphasia as a delayed, and usually aberrant, acquisition of language for communicative use, provided the delay is not accounted for by deafness or by severe mental retardation. It must be distinguished from acquired aphasia as a result of focal pathological lesions usually affecting the left hemisphere. In the present case report there was no evidence of an acquired insult or disease process although the patient did have a delay in walking. The language disorder was associated with oromotor apraxia, anomia and the use of gesture. The assumed developmental basis for the dysphasia was correlated with subtle developmental brain anomalies characterized by symmetry of the plana temporale similar to that reported in cases of dyslexia. (Galaburda AM et al. Ann Neurol 1985; 18:222).

PSEUDOBULBAR PALSY AND MACROGYRIA

Four patients with medically intractable epilepsy, pseudobulbar palsy and mental retardation and found to have bilateral central macrogyria on CT and MRI are reported from the Department of Neurology and Neurosurgery, McGill University, and the Montreal Neurological Institute and Hospital, Montreal, Canada; and the Department of Neurology, University of Minnesota, St. Paul Ramsey Hospital, St. Paul, MN. The pseudobulbar palsy was associated with oromotor incoordination, developmental delay and mild retardation. Minor seizures developed between the ages of eight and nine years and one patient had infantile spasms at three months of age. Electroencephalographic epileptogenic abnormalities were secondary generalized or multifocal. CT scans revealed symmetrical bilateral sylvian and rolandic macrogyria extending into the parietal regions. The cortex appeared thick and smooth with the underlying white matter diminished. The MRI confirmed the CT findings and showed that the abnormal cortex had a lower signal as compared to normal gray matter of frontal and occipital regions. The thick cortical structures surrounded a large central sulcus reminiscent of a fetal sylvian fissure. Two patients tried on multiple anticonvulsants continued to have frequent seizures whereas two treated by callosotomy had no subsequent drop attacks and improved behavior. The authors suggest that the clinical and imaging features of these patients indicate a distinct and specific syndrome and the malformations appear to result from specific derangement of neuronal migration. (Kuzniecky R, Andermann F et al. Bilateral central macrogyria: Epilepsy, pseudobulbar palsy, and mental retardation -- A recognizable neuronal migration disorder. Ann Neurol June 1989; 25:547-554).

COMMENT. Neuronal migration disorders, including agyria (or lissencephaly), macrogyria (or pachygyria), polymicrogyria,