PEDIATRIC NEUROLOGY BRIEFS
A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 25, No. 6 June 2011

PERINATAL DISORDERS

MOTOR PATHWAY INJURY WITH PERIVENTRICULAR LEUKOMALACIA AND SPASTIC DIPLEGIA

The relationship between motor pathway injury and motor impairment was investigated by voxelwise correlation analysis in 43 patients (median age 12 years) with periventricular leucomalacia and spastic diplegia at Yonsei University College of Medicine, Seoul, Korea. Functional connectivity of motor cortical and thalamocortical pathways was also evaluated in 11 patients using functional MRI, and results in patients with spastic diplegia CP were compared with normal controls. White matter volume reduction was not significantly correlated with motor dysfunction. Fractional anisotropy within the white matter tracts of patients was lower than controls, and that within the corticospinal tracts and corpus callosum was significantly correlated with motor dysfunction ($P<0.03$). A lesser correlation occurred with that in thalamocortical pathways ($P<0.05$). Cortical volume of pre- and post-central gyri and paracentral lobule tended to be negatively correlated with motor function. Motor cortical connectivity was diminished in the somatosensory cortex, paracentral lobule, cinglate motor area and visual cortex, whereas thalamovisual connectivity was spared, despite optic radiation injury. Neuronal g-aminobutyric acid receptor binding potential, measured with positron emission tomography scans (n=27) and compared with controls (n=20), was focally increased in the lower extremity homunculus, cingulate cortex, visual cortex and cerebellum ($P<0.05$), a compensatory plasticity response to injury. The mechanism of motor dysfunction in patients with periventricular leucomalacia involves injury to descending motor tracts and reduction in cortical volume and functional connectivity. (Lee JD, Park H-J, Park ES, et al. Motor pathway injury in patients with periventricular leucomalacia and spastic diplegia. **Brain** April 2011;134:1199-1210). (Response: Jong Doo Lee MD,

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 2011 covers selected articles from the world literature and is published monthly. Send subscription requests ($68 US; $72 Canada; $75 airmail outside N America) to **Pediatric Neurology Briefs - J. Gordon Millichap, M.D., F.R.C.P.-Editor,** P.O. Box 11391, Chicago, Illinois, 60611, USA. The editor is Pediatric Neurologist at Children's Memorial Hospital and Professor Emeritus, Northwestern University Medical School, Chicago, Illinois.
PNB is a continuing education service designed to expedite and facilitate review of current scientific information for physicians and other health professionals.

Diagnostic Radiology and Nuclear Medicine, Yonsei University College of Medicine, Seoul, 120-752, Korea. E-mail: jdlee@yuhs.ac).

COMMENT. Pathophysiological mechanisms of motor impairment in patients with periventricular leukomalacia and spastic cerebral palsy are controversial (Bax et al, **JAMA** 2006; Woodward et al, **N Engl J Med** 2006) The above study demonstrates that descending motor pathway injury and reduction of overlying cortical volume are leading mechanisms.

Neuronal cell death in neonatal hypoxia-ischemia, reviewed by researchers at Johns Hopkins University School of Medicine (Northington FJ et al. **Ann Neurol** May 2011;69:743-758), is manifested as a continuum of programmed cell death (PCD) from apoptosis (dismantling of cells, Type I PCD) to necrosis (lytic destruction of cells) and autophagy (degradation of cells by lysosomal system, Type II PCD), and not as distinct categories of neurocellular degeneration. Hypothermia in infants with HIE protects against necrosis and apoptosis.

NEURODEVELOPMENTAL DISORDERS

NEUROANATOMICAL ABNORMALITIES IN BOYS WITH IDIOPATHIC AUTISM COMPARED TO FRAGILE X SYNDROME

Researchers at Stanford University, CA, and University of North Carolina, Chapel Hill, used voxel-based morphometric analyses, and multivariate pattern and clustering analyses to compare brain MRI studies in 165 young boys, aged 1.57- 4.15 years, diagnosed with idiopathic autism (iAUT) or fragile X syndrome (FXS) and idiopathic developmentally delayed and normal controls. Frontal and temporal gray and white matter regions of brains of boys with iAUT and FXS were developmentally aberrant as compared with controls. The differences were in opposite directions in iAUT and FXS relative to controls; iAUT brains had greater volume compared to controls, and FXS brains had smaller volume than control brains. Pattern of brain structure in iAUT generally resembled that of controls more than FXS, with or without autism. (Hoeft F, Walter E, Lightbody AA et al. Neuroanatomical differences in toddler boys with fragile X syndrome and idiopathic autism. **Arch Gen Psychiatry** March 2011;68(3):295-305). (Response: Allan L Reiss MD, Dept Psychiatry and Behavioral Sciences, Stanford University School of Medicine, Stanford, CA 94305. E-mail: reiss@stanford.edu).

COMMENT. The authors conclude that idiopathic autism and fragile X syndrome have distinct neuroanatomical patterns, a finding that supports the neurobiological heterogeneity of idiopathic autism. They suggest that significant differences in aberrant brain morphometry may be found in other ASD-associated genetic disorders, eg Angelman syndrome, Rett syndrome. ASD has been diagnosed in more than 20 genetically determined syndromes, most prominently, FXS. (Harris JC. Editorial 2011; Moss J et al. 2009).