

# PEDIATRIC NEUROLOGY BRIEFS

## A MONTHLY JOURNAL REVIEW

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

---

Vol. 24, No. 1

January 2010

---

### MOVEMENT DISORDERS

#### DYSKINETIC ENCEPHALITIS LETHARGICA

Twenty children (12 female; age range, 1.3-15 years) with encephalitis lethargica were studied over 9 years by researchers at University of Sydney, Westmead, Australia; and John Radcliffe Hospital, Oxford, UK. Encephalitis lethargica is characterized by psychiatric features (catatonia, agitation, compulsive behaviors), extrapyramidal movement disorders (*hyperkinetic*, chorea, oculogyric crises, dystonia, hemiballism, or *akinetik*, Parkinsonism), and sleep disturbance (insomnia or hypersomnolence). Seizures, autonomic dysfunction, encephalopathy, and cognitive dysfunction are less common. Ten sera (from 2 males and 8 females) and 6 cerebrospinal fluid samples, taken during the first 3 weeks of the illness before immunotherapy, were positive for N-methyl-d-aspartate receptor autoantibodies. Of 10 *NMDAR-Ab-positive* patients, 4 were <5 years of age, and only 2 had preceding infection. Presentation was often dramatic with agitation, catatonia, dyskinesias and seizures, and Parkinsonism and somnolence were prominent only in 4 patients. Unusual dyskinesias included cycling, stereotyped flailing, and orolingual "rabbit" movements. At follow-up, 4 patients were completely recovered, and 6 were impaired. None had identified ovarian tumor. Of 10 *NMDAR-Ab-negative* patients, none was <5 years of age; 7 were postinfectious. Parkinsonism was the dominant movement disorder. Seven had persistent symptoms at follow-up. Immunotherapies included IV methyl-prednisolone followed by oral prednisolone (n=13) or IV immunoglobulin (n=2). (Dale RC, Irani SR, Brilot F, et al. N-methyl-d-aspartate receptor antibodies in pediatric dyskinetic encephalitis lethargica. *Ann Neurol* Nov 2009;66:704-709). (Respond: Dr Angela Vincent, Neuroimmunology Group, West Wing and Weatherall Institute of Molecular Medicine, John Radcliffe Hospital, Oxford OX3 9DS, UK. E-mail: [angela.vincent@imm.ox.ac.uk](mailto:angela.vincent@imm.ox.ac.uk)).

---

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 2010 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.

Fax: 312-943-0123.

COMMENT. NMDAR-Ab encephalitis is a dyskinetic form of encephalitis lethargica that may affect very young children and is rarely preceded by infection. Detectable ovarian tumors are uncommon in female patients under the age of 20 years. Outcome is poor, and early immunotherapy is considered warranted. The detection of specific antibodies is important in children with symptoms of encephalitis. The authors previously reported 20 cases of encephalitis lethargica with evidence of basal ganglia autoimmunity, mainly in children, and the condition is not rare. (**Brain** 2004;127:21-33; **Ped Neur Briefs** Jan 2004;18:1-2).

## INCREASED PREVALENCE OF DYSKINETIC CEREBRAL PALSY

The prevalence and severity of dyskinetic cerebral palsy (DCP) in European children born 1976-1996 were analyzed in a multicenter study in Göteborg, Sweden; Cork, Ireland; Tübingen, Germany; and Grenoble, France. Of 578 children with DCP, 70% were born at term. The prevalence per 1000 live births increased from 0.08 in the 1970s to 0.14 in the 1990s. In 386 children (70%) with a birth weight >2500 g the increase of DCP was significant (0.05 to 0.12), whereas neonatal mortality in this birth weight group decreased. Compared to children with bilateral spastic cerebral palsy (BSCP), children with DCP had more severe motor and cognitive impairments: 59% needed a wheelchair, 24% walked with aids, and 16% without aids; and 52% had severe learning disability. Epilepsy developed in 51%, and severe visual and hearing impairment in 19% and 6%, respectively. Cognitive impairments increased concurrently with severity of motor deficits. In children born 1991-1996, perinatal adverse events (Apgar score <5 at 5 min and convulsions before 72 h) had occurred more frequently in DCP children of those with BSCP. (Himmelmann K, McManus V, Hagberg G, Uvebrant P, Krageloh-Mann I, Cans C, on behalf of the Surveillance of Cerebral Palsy in Europe. Dyskinetic cerebral palsy in Europe: trends in prevalence and severity. **Arch Dis Child** Dec 2009;94:921-926). (Respond: Kate Himmelmann, Queen Silvia Children's Hospital, SE-416 85 Göteborg, Sweden. E-mail: [kate.himmelmann@vgregion.se](mailto:kate.himmelmann@vgregion.se)).

COMMENT. Professor PO Pharoah, Department of Public Health, Liverpool, UK, in an editorial, comments as follows: Prevalence is determined by a function of incidence and duration of a disease; an increase in duration or survival results in increased prevalence without change in incidence. The authors consider the increase in prevalence of dyskinetic CP in term infants is significant because there was no concomitant increase among preterm infants. However, preterm infants contributed more to the decrease in neonatal mortality of term infants, and therefore to the improved duration due to greater survival. The increasing trend in prevalence of dyskinetic CP may be related to the higher proportion of small for gestational age infants involved and a prepartum etiological factor. Epidemiological data and caveats are difficult to interpret, (Pharoah PO. **Arch Dis Child** 2009;94:917-918).

Professor TTS Ingram of Edinburgh University, in his classic studies of "Paediatric Aspects of Cerebral Palsy." (E & S Livingstone, 1964;pp360-1), compares the prevalence of various types of CP in children under age 15 years in Edinburgh in 1951. Of total cases including all social classes, dyskinetic CP occurred in 0.17 and diplegia in 0.70 per 1000. Dyskinesia and diplegia are markedly more prevalent in Social