

Neuropediatr June 1994;1:59-64). Chorea resolved after 15 days haloperidol therapy, and no further ischemic episodes had occurred in 16 months during treatment with nicardipine. The authors cite two previous reports of moyamoya and chorea.

## MYASTHENIA GRAVIS

### JUVENILE MYASTHENIA AND PUBERTY

The influence of race, sex, and puberty on incidence, severity, and outcome of juvenile myasthenia gravis beginning before age 20 years was evaluated in 115 patients seen at the University of Virginia, Duke University, and University of North Carolina at Chapel Hill. White patients with prepubertal disease onset had an equal sex ratio, and female predominance increased during and after puberty. Males had less severe disease than females. Black patients showed a constant F:M ratio of 2:1 in all pubertal-onset groups. Spontaneous remissions only occurred in white patients with prepubertal onset; and persistent symptoms for more than 10 years were least frequent in this group. Early thymectomy in white patients was followed by more remissions and milder symptoms than late thymectomy. Black patients had infrequent remissions, and similar disease severity after early or late thymectomy. (Andrews PI et al. Race, sex, and puberty influence onset, severity, and outcome in juvenile myasthenia gravis. Neurology July 1994;44:1208-1214). (Reprints: Dr P Ian Andrews, Division of Pediatric Neurology, Box 3533, Duke University Medical Center, Durham, NC 27710).

COMMENT. This study documents the importance of race, sex, and puberty on the incidence, severity, response to thymectomy, and outcome in juvenile myasthenia gravis. Thymectomy was most effective in white patients when performed within 1 year of peripubertal disease onset. See Progress in Pediatric Neurology II, Chicago, PNB Publ, August 1994, for further reports of juvenile myasthenia gravis from the University of Iowa, a multicenter study in Italy, and from the Mass General Hospital, Boston.

## TOXIC DISORDERS

### LEAD EXPOSURE: INAPPROPRIATE SCREENING PRACTICES?

Physician screening practices at a hospital-based, university-affiliated pediatric primary care center serving an urban high-risk population in Rochester, NY were evaluated to determine the feasibility of the 1991 Centers for Disease Control guidelines. Among 632 children aged 9 to 25 months who attended the center between 1989 and 1991, screening was deficient in 55%, 34%, and 29% at ages 9-13 months, 14-19 months, and 20-25 months, respectively. Many high-risk children living in houses built before 1950, including those making well-child visits, were not appropriately screened for lead toxic effects, and opportunities for testing were frequently missed. (Campbell JR, McConnochie KM, Weitzman M. Lead screening among high-risk urban children. Are the 1991 Centers for Disease Control and Prevention Guidelines feasible? Arch Pediatr Adolesc Med July 1994;148:688-693). (Reprints: Dr Campbell, Department of Pediatrics, Rochester General Hospital, 1425 Portland Ave, Rochester, NY 14621).

COMMENT. The authors believe that the new CDC guidelines for biannual screening and retesting can be achieved with a modest increase in tests, if sick visits are also utilized for retesting. Proper cleansing of the fingertip or alternatively, venipuncture should decrease the proportion of false-positive results and reduce the need for additional visits. At this urban PC Center, 30% of the screened high-risk children had lead levels of 10 mcg/dL or higher, now considered to be toxic. In contrast, only 4.7% of children screened from a suburban, middle class private practice in Cherry Hill, NJ, were found to have elevated blood lead levels, and no child had a level of 25 mcg/dL or higher requiring treatment. (Taubman B et al. Arch Pediatr Adolesc Med July 1994;148:757-760).

**Who Bears the Burden?** Wical BS, of the Departments of Neurology and Pediatrics, University of New Mexico, Albuquerque, addresses the dilemmas and burdens of blood lead testing of 22 million children in the US from 6 to 72 months of age. (Arch Pediatr Adolesc Med July 1994;148:760-761). In Milwaukee, private physicians play a major role in lead poisoning screening and case identification. In 1992, the number of cases of poisoning identified (BPb 25 mcg/dL or higher) in the private sector increased by over 600% in a 2 year period, a higher number than in public centers. Physicians voluntarily changed their practice patterns in accordance with 1991 CDC recommendations, partly as a result of physician education by the Milwaukee Health Department and the Children's Hospital of Wisconsin. (Schlenker TL et al. Arch Pediatr Adolesc Med July 1994;148:761-764).

**The Child Neurologist's Role.** Pediatric neurologists may need to take some responsibility in the blood lead screening of their patients, particularly those in high-risk categories. No child is exempt from risk, however; one infant, a pediatric surgeon's child, living in a high-rise co-op apartment, the first to be built in Chicago in 1924, and another, a house master's child at a renowned boarding school in Massachusetts, were exposed to lead containing paint in the homes. Blood lead levels, in both cases prompted by the mothers and found to be elevated, responded to a temporary change of residence and lead abatement measures.

**Declining Lead Levels.** Despite the dramatic overall decrease in blood lead levels in the US population in recent years to 2.8 mcg/dL, national estimates for children 1 to 5 years of age show that 8.9%, or 1.7 million children, have BPb levels of 10 mcg/dL or greater, high enough to be of concern. (Brody DJ, Pirkle JL et al. Blood lead levels in the US population. Phase 1 of the third National Health and Nutrition Examination Survey (NHANES III, 1988 to 1991). JAMA July 27, 1994;272:277-283). Pirkle JL, Brody DJ et al, commenting on the decline in blood lead levels in the US, attribute the fall to the removal of lead from gasoline and soldered cans. Lead in paint, dust and soil needs to be addressed before the decline can continue, especially in children in low income, urban areas. (JAMA July 27, 1994;272:284-291). Goldman LR and Carra J, from the Office of Prevention, Pesticides, and Toxic Substances, US Environmental Protection Agency, Washington, DC, emphasize the need for both targeted screening efforts and improvements in screening methods in high-risk children. (Childhood lead poisoning in 1994. JAMA July 27, 1994;272:315-316).

### **Public Lead Awareness and Responsibility.**

Since current compliance with CDC recommended guidelines for blood lead screening is not universally appropriate, an increased public and parent awareness of the hazards and the symptoms and signs of lead poisoning, especially in children, should be encouraged. (Millichap JG. Environmental Poisons in Our Food. Chicago, PNB Publ, 1993).

### **DIAZINON EXPOSURE AND INFANTILE HYPERTONIA**

A 12-week-old infant girl who developed persistent hypertonicity 5 weeks following exposure to the organophosphate insecticide diazinon (Knox-Out 2FM) in the home is reported from Oregon State University, Corvallis, OR. The infant's urine contained alkylphosphate metabolites of diazinon (60 ppb diethylphosphate and 20 ppb diethylthiophosphate). Serum cholinesterase was normal. Diazinon levels in the home (floor, vacuum cleaner dust, and air) were excessive even at 6 months after application. Six weeks after evacuating the home, the infant's muscle tone returned to normal, ankle clonus had resolved, and subsequent development was normal. (Wagner SL, Orwick DL. Chronic organophosphate exposure associated with transient hypertonia in an infant. Pediatrics July 1994;94:94-97). (Reprints: Dr Sheldon L. Wagner, Agricultural Chemistry, Oregon State University, Agricultural & Life Sciences 1007, Corvallis, OR 97331).

COMMENT. None of the typical muscarinic or nicotinic symptoms of organophosphate intoxication was present in this infant. Organophosphates can cause a delayed neurobehavioral toxicity, characterized by neuritis, paralysis, and psychological changes, the result of degeneration of myelin and nerve axons and effects on neurotransmitters. A relationship between Parkinson's disease and exposure to pesticide chemicals has been demonstrated in agricultural workers. Children presenting with unexplained neurobehavioral symptoms should be investigated for possible exposure to insecticide environmental toxins.

### **MOVEMENT DISORDERS**

#### **HEMIFACIAL SPASMS AND CEREBELLAR ANGLE TUMORS**

Two children, aged 3 years, with hemisomatic spasms caused by tumors in the ipsilateral cerebellopontine angle are reported from the Division of Pediatric Neurology and Department of Neurology, University of Texas Southwestern Medical Center, Dallas, TX. Patient 1 had persistent hemifacial spasms with onset soon after birth; some were complicated by flexion of the arm and extension of the leg. An initial diagnosis of partial seizures was not confirmed by video-EEG monitor, and anticonvulsants were of no benefit. CT and ultrasound were normal, but MRI revealed a C-P angle tumor. Following partial resection of a low-grade ganglioneuroma, the spasms were less severe. Patient 2 developed left sided jerks at 2 years of age, turning of the head to the right, and flexion of left elbow and hip, without loss of consciousness. Movements were worse while speaking or watching television. The EEG was normal and carbamazepine without benefit. MRI uncovered a left sided angle tumor, and spasms ceased after partial resection of a ganglioneuroma. (Al-Shahwan SA, Roach ES et al. Hemisomatic spasms in children. Neurology July