

methylmercury in the diet. For previous reports of neuropsychological effects of methylmercury and PCP exposures see Progress in Pediatric Neurology III, PNB Publ, 1997;pp278-280.

**Iatrogenic exposure to mercury after hepatitis B vaccination in preterm infants** is reported by Stajich GV et al. (J Pediatr May 2000;136:679-681). Thimerosal, a mercury-derived preservative, has been used in some vaccines since the 1930s. Thimerosal is composed of 49.6% ethylmercury, which behaves like methylmercury. According to a review editorial by Pless R and Risher JF (J Pediatr May 2000;136:571-573), infants may be exposed to cumulative doses of mercury from vaccines in the first 6 months, exceeding the EPA limit of 0.1 mcg/kg/d for chronic daily exposure to methylmercury. The Advisory Committee on Immunization Practices has recommended that hepatitis B vaccine used in infants at birth should not contain thimerosal.

## PRE- AND PERI-NATAL DISORDERS

### **EARLY PREDICTORS OF HIE ADVERSE OUTCOME**

A retrospective study of 35 term infants with post-asphyxial hypoxic-ischemic encephalopathy (HIE) was conducted at the KK Women's and Children's Hospital, Singapore, to determine early predictors of mortality or major motor morbidity at 18 months of age. A severe adverse outcome occurred in 23; thirteen died and ten had major neurological sequelae. Risk factors included a low 5 min Apgar score (<4), the use of adrenaline, low arterial pH (<7.1) and high base deficit (>20 mEq/L). The high base deficit and low Apgar score combined had a positive predictive value of 100%. (Toh VC. Early predictors of adverse outcome in term infants with post-asphyxial hypoxic ischaemic encephalopathy. Acta Paediatr March 2000;89:343-347). (Respond: Dr Veronica C Toh, Department of Neonatology, KK Women's and Children's Hospital, 100 Bukit Timah Rd, Singapore).

COMMENT. The combination of a low Apgar score and high base deficit in term infants with post-asphyxial HIE is an early predictor of mortality or major neurological sequelae.

**Serum CPK and outcome of HIE** An elevated serum CPK measured within 4 hours after birth is a sensitive indicator of brain damage in asphyxiated term infants but is of limited prognostic value in assessment of neurological outcome, according to one previous report, whereas another study showed that CPK measured in cord blood correlates with outcome after asphyxia and compares favorably with imaging studies. (Progress in Pediatric Neurology I, 1991;pp 332).

Cranial ultrasonography and spectroscopy are of value in the prediction of neurodevelopmental outcome of HIE (see Progress in Pediatric Neurology II, 1994; 313-331).

### **MATERNAL THYROID FUNCTION AND INFANT DEVELOPMENT**

The effect of maternal thyroid function in the first half of pregnancy on the neurologic development of 20 infants in the first two years of life was studied at the University of Amsterdam and Emma Children's Hospital, The Netherlands. At the age of 6 and 12 months, the mean mental developmental index (MDI) score was 16 points lower for 7 infants born to mothers with subclinical hypothyroidism compared to 6 with euthyroid mothers (P=.03 and .02, respectively). At 24 months, a mean 6 point lower MDI score was not statistically significant. One infant out of

7 born to mothers with hyperthyroidism developed transient hyperthyroidism, but none showed changes in the MDI. Motor nerve conduction, somatosensory evoked potentials, and Bayley scales showed no differences among groups. (Smit BJ, Kok JH, Vulsma T et al. Neurologic development of the newborn and young child in relation to maternal thyroid function. Acta Paediatr March 2000;89:291-295). (Respond: Dr BJ Smit, Department of Neonatology, H3N-148, Academic Medical Center, University of Amsterdam, Emma Children's Hospital AMC, PO Box 22700, 1100 DE Amsterdam, The Netherlands).

COMMENT. Maternal subclinical hypothyroidism in the first half of pregnancy is associated with a lower mental development score in the infants during the first year of life. Optimal treatment of maternal thyroid disease throughout pregnancy is emphasized.

Low triiodothyronine levels in preterm infants are correlated with a 6.6 point deficit in Full Scale IQ scores on the WISC Scales at 8 year follow-up, and an increased risk of developing cerebral palsy and cognitive deficits is reported in premature infants with low thyroxine levels in the first week of life. Some authorities caution that thyroxine therapy for hypothyroid mothers is unlikely to benefit premature babies, and triiodothyronine replacement in premature infants may be dangerous. Neonatologists generally withhold therapy pending a retest after the infant reaches term. (See Progress in Pediatric Neurology III, 1997;pp 281-2).

## HEREDITARY NEUROPATHIES

### GENETICS OF MOTOR AND SENSORY NEUROPATHY TYPE 2C

Thirty-three subjects in a large family with hereditary motor and sensory neuropathy (HMSN) type 2C were examined by linkage analysis at the Mayo Clinic, Rochester, MN. The HMSN 2C phenotype, including 12 affected and 11 at risk, were not linked to either the HMSN 2B or 2D loci. HMSN 2C is genetically distinct from HMSN 2A, 2B, and 2D. (Nagamatsu M, Jenkins RB, Schaid DJ, Klein DM, Dyck PJ. Hereditary motor and sensory neuropathy type 2C is genetically distinct from types 2B and 2D. Arch Neurol May 2000;57:669-672). ((Reprints: Peter James Dyck MD, Peripheral Neuropathy Research Center, Mayo Clinic, 200 First St SW, Rochester, MN 55905).

COMMENT. HMSN type 2C is a clinical variety of HMSN 2 manifested by motor and sensory neuropathy of the limbs and progressive weakness of the vocal cords, diaphragm, and intercostal muscles. HMSN 2 shows genetic heterogeneity, and at least 4 genetic varieties have been confirmed: HMSN 2A (1p35-p36), HMSN 2B (3q13-q22), HMSN 2D (7p14), and HMSN 2C (not linked to any of these loci).

The many faces of Charcot-Marie-Tooth disease (HMSN) are reviewed in an editorial (Vance JM. Arch Neurol May 2000;57:638-640). Phenotype classifications in CMT include CMT1 (autosomal dominant, demyelinating), CMT2 (autosomal dominant, axonal presentation), CMT3 (autosomal recessive, axonal presentation), CMT4 (autosomal recessive, demyelinating presentation), and CMTX (X-linked, axonal, or demyelinating presentation). Both clinical and molecular classifications are discussed.