

Denmark. Phenobarbital imposed no risk of cerebrovascular damage in newborns with fetal distress. (Andersen K et al. The effect of phenobarbital on cerebral blood flow in newborn infants with foetal distress. Eur J Pediatr Aug 1994;153:584-587).

COMMENT. Phenobarbital is the most commonly used anticonvulsant in neonates and has been advocated in the prevention of periventricular hemorrhage in preterm infants. This demonstration of the safety of phenobarbital in neonates with HIE is encouraging and should offset in part the poor rating the drug has received in some febrile seizure studies.

RETT SYNDROME

POLYSOMNOGRAPHY IN RETT SYNDROME

Respiratory patterns, awake and asleep, were investigated by polysomnography in 30 female patients with Rett syndrome and compared with 30 controls at the Eudowood Division of Pediatric Respiratory Sciences, Johns Hopkins University, and the Department of Neurology, Kennedy Krieger Institute, Baltimore, MD. The median age was 7 years (range, 1 to 32 years). During wakefulness, 67% of RS patients had a characteristic pattern of disordered breathing (hyperventilation followed by central apnea and desaturation). Breathing was normal during sleep. Arterial oxygen saturation during REM sleep was slightly lower in RS cf controls but within normal range. The authors postulate a normal brain-stem control of ventilation in RS and an abnormality or loss of the normal cortical influence on ventilation during wakefulness. The precise cause of the cortical dysfunction is unknown. (Marcus CL, Naidu S et al. Polysomnographic characteristics of patients with Rett syndrome. J Pediatr Aug 1994;125:218-24). (Reprints: Dr Marcus, Johns Hopkins Hospital, Div Pediatric Pulmonology, Park 316, 600 North Wolfe St, Baltimore, MD 21287).

COMMENT. Patients with Rett syndrome have normal respiration during non-REM sleep, slightly abnormal breathing in REM sleep, and markedly disturbed breathing during wakefulness. The hyperventilation is attributed to a cortical dysfunction.

RETT VARIANTS: A WIDENING SYMPTOM COMPLEX

In a collaborative Swedish-Norwegian project, at Goteborg Sweden and Oslo, Norway, a model for the clinical delineation of atypical cases of Rett syndrome was applied to a pilot series of 16 mentally retarded females, aged 11 to 47 years (median: 23). The atypical variants included forme fruste cases (8), late regression (6), and congenital variants (2). The model was based on age >10 years, 3 of 6 primary inclusion criteria for RS (eg. hand stereotypes, decelerated head growth, stages of regression and recovery of contact), and 5 of 11 supportive characteristics (eg. breathing irregularities, air swallowing, teeth grinding, gait dyspraxia, scoliosis). The model identified RS variants and distinguished them from other disorders, eg. Angelman's syndrome that fulfilled 3 supportive criteria. (Hagberg BA, Skjeldal OH. Rett variants: a suggested model for inclusion criteria. Pediatr Neurol July 1994;11:5-11). (Respond: Dr Hagberg, Dept of Pediatrics, Ostra Sjukhuset, S-416 85 Goteborg, Sweden).

COMMENT. For a review of recent international research on RS, see "Rett syndrome: from gene to gesture." J R Soc Med Sept 1994;87:562-566).