COMMENT: ACTHis effective in the control of infantile spasms and hypsarrhythmia in 50% of cases. The response rate is higher in infants treated early and under one year of age than in those diagnosed later. Hypertension, cushingnoid obesity, congestive heart failure, infection, and cerebral atrophy are some of the more serious side-effects of ACTH therapy. The significant response of infantile spasms to TRH without serious toxicity offers a promising alternative therapy to ACTH. The anticonvulsant action of TRH appears to be central and unrelated to its endocrine action through the pituitary-thyroid axis.

NEOPLASMS AND RELATED CONDITIONS

NEUROFIBROMATOSIS

LINK (Let's Increase Neurofibromatosis Knowledge), the British Neurofibromatosis Association, organised a major European Symposium at Egham, Surrey, Feb 5-7, 1987, and clarified the distinguishing features of two syndromes with separate genetic markers: 1) von Recklinghausen's neurofibromatosis (VRNF), the so-called peripheral type, and 2) bilateral acoustic neurofibromatosis (BANF), the central variety.

VRNF with a prevalence of 1 in 3000 is inherited as an autosomal dominantcondition with 100% penetrance and a high mutation rate. Serious complications, occurring in about 20% include large plexiform neurofibromas, kyphoscoliosis, and optic nerve or chiasmal gliomas. Children should be examined twice a year to check for complications.

The gene responsible for VRNF, although not identified, was narroweddown to a few chromosomes by data that provided an 'exclusion map' at this conference. The genetic analysis of BANF patients has shown deletions on chromosome 22. a step closer to the identification of the defective gene responsible for acoustic neuromas. (Lancet 1987; i:663-664)

COMMENT: A similar conference on neurofibromatosis is scheduled for July 13 - 15, 1987 in the U.S. to be sponsored by the National Institute of Health, Bethesda, Md. and chaired by Dr. David A. Stumpf of Northwestern University Medical School. It is perhaps unfortunate that the European and US sponsors could not have pooled their resources to make this an International Symposium.

DEGENERATIVE DISORDERS

RETT'S SYNDROME

The sleep and respiratory patterns associated with this disorder have been studied in 11 females aged 2 through 15 years at the Methodist Hospital, Houston, Tx. Polygraphic recordings showed a pattern of disorganised breathing and compensatory hyperpnea during wakefulness with regular, continuous breathing during sleep. The findings suggest an altered or impaired voluntary/behavioural respiratory control system in patients with Rett's Syndrome. (Glaze DG, Frost JD Jr, Zoghbi HY, Percy AK. Rett's Syndrome: characterisation of respiratory patterns and sleep. Ann Neurol 1987;21:377-382)

COMMENT: In 1966, Rett described a progressive dementia in girls with onset in early childhood and associated with autistic behaviour, apraxia of gait, and stereotyped use of the hands. The cause of Rett's syndrome is unknown. I have seen several atypical cases that fit the description except for the absence of so-called pathognomonic hand wringing movements and hyperventilation, and some were boys. Is Rett's syndrome a specific disorder or nonspecific, with more than one etiology? For a review of Rett syndrome, refer to Ann J Med Genet 1986 (suppl).

DYSLEXIA AND LEARNING DISABILITIES

GENETICS AND READING DISABILITIES

Psychologists and psychiatrists at the University of Surrey, Guildford, Surrey, and the Hospital for Sick Children, Great Ormond Street, London, UK studied the reading skills of 285 pairs of 13 year-old twins using standardized measures of intelligence, reading and spelling ability and correlations in monozygotic and same-sex dizygotic twins. Genetic factors played only a moderate role in general reading backwardness and specific reading retardationwhereas strong genetic influences for spelling disability were found. (Stevenson J, Graham P, Fredman G, McLoughlin V. A twin study of genetic influences on reading and spelling ability and disability. J Child Psychol Psychiat. 1987; 28:229-247)

COMMENT: Of a total of 96 twin pairs reported in the literature, 36 (88%) monozygotic twins and only 16 (29%) dizygotic twins were concordant for dyslexia (Dyslexia: as the Neurologist and Educator read it. Charles C Thomas, Springfield, Illinois, USA, 1986). Between 25 and 50% of children with reading disability demonstrate transmission within families. Hallgren (1950) concluded that his data best fitted an autosomal dominant genetic mechanism and others have proposed alternative genetic models: autosomal dominant with reduced penetrance in females, and sex-linked recessive. These studies are at variance with the present authors' conclusions that emphasise the complexity of genetic influences on literacy skills and the importance of changes that occur with development in our understanding of the causation of reading difficulties.

ADDITIVES AND HYPERKINETIC BEHAVIOUR

The authors studied 39 children with hyperkinetic and learning disorders in a summer camp setting. The behaviour was monitored by videotape for 4-minute intervals at mealtimes. The Feingold diet was administered for 1 week followed by a diet containing additives and preservatives for 1 week. Three observers who were blind to the respective diet periods rated the behaviour for motor restlessness, disorganised behaviour, and misbehaviour. No significant differences were found in behaviour during weeks 1 and 2. The authors conclude that the Feingold Diet has no beneficial effect on most children with learning and hyperkinetic diisorders. (Gross MD, Tofanelli RA, Butzirus SM, Snodgrass EW. J Amer Acad Child Adol Psychiat. 1987; 26:53-55)