syndromes.

DEMYELINATING AND DEGENERATIVE DISEASES

NATURAL HISTORY OF EARLY ONSET MULTIPLE SCLEROSIS

The clinical course of multiple sclerosis (MS) in 116 patients with onset before age 16 (prevalence 3.6% in total MS clinic attendees) was evaluated by longitudinal study (mean duration 19.76 +/- 0.9 years) at the University of British Columbia MS Clinic. Mean age of onset of MS was 12.73 +/- 0.25 years; 23 (19.8%) had onset at age 10 or younger, and 6 (5.2%) at age 7 or younger. The female to male ratio was 2.87:1. Female preponderance was highest for patients with onset at ages 13 - 14 (pubertal age). Most frequent initial symptoms were sensory disturbances (25.9%), optic neuritis (21.6%), brainstem dysfunction (12.9%), gait disorders (9.4%), and cerebellar incoordination (6.9%). Sensory and brainstem disturbances were more frequent in girls, whereas boys were affected more by motor and gait disturbances. The MS course was primary progressive (PP) in 3 cases (2.6%), and secondary progressive (SP) in 60 (53.1%),with 50% probability of SP by 23 years after onset. For relapsing remitting (RR) or SPMS cases, the mean duration from onset to an Expanded Disability Status Scale (EDSS) 3 was 16.03 +/- 1.17 years, at a mean age of 28.47 +/- 1.14 years. EDSS 6 was reached by 19.39 +/- 1.43 years, at a mean age of 32.32 +/- 1.44 years. Annual relapse rate was 0.54 +/- 0.05 per year. The course of MS was significantly correlated with the number of relapses in the first year after onset. The majority of cases had a RR course, and early age at permanent disability. Disease-modifying therapy should be considered early in the course of early onset MS. (Boiko A, Vorobeychik G, Paty D et al. Early onset multiple sclerosis. A longitudinal study. Neurology October (1 of 2) 2002;59:1006-1010). (Reprints: Dr DW Paty, Room S195, 2211 Wesbrook Mall, UBCH, Vancouver, BC V6T 2B5, Canada).

COMMENT. Patients with early onset MS (EOMS) tend to recover from the initial manifestations and have a relatively long first and second remission. However, EOMS cases include patients with frequent relapses, early age at permanent disability, and occurrence of malignant cases. In those with duration more than 10 years, 65% are disabled. The frequency of relapses in the first 5 years after onset, and the duration of first and second remissions correlate with the risk of permanent disability. Initial brainstem symptoms are predictive of a poor prognosis. Therapy should be considered sooner rather than later in EOMS.

PATTERNS OF REGRESSION IN RETT SYNDROME

Patterns and features of regression in a case series of 53 girls and women with Rett syndrome were studied at the Institute of Child Health and Great Ormond Street Children's Hospital, London, UK. Diagnostic criteria for classical Rett syndrome were met in 46 cases, and 7 had an atypical/variant form. The most common period for regression was 12 - 18 months, reported by parents or in casenotes in 49% of patients. Mean age of regression was 16 months. Skills lost in order of frequency were hand use (85%), non-verbal vocalizations and simple gestures (59%), non-verbal play (51%), motor skills (49%), and words (45%). Pre-regression developmental delays were noted in more than two-thirds of cases (85% in youngest cases when parental reports were most reliable). Age at regression was not an index of neurological severity (epilepsy, breathing abnormalities, mobility, joint contractures, and oral-motor dysfunction). (Charman T, Cass H, Owen L et al. Regression in individuals with Rett syndrome.