

## CEREBRAL TUMORS

### BRAIN TUMORS IN INFANTS

The clinical manifestations, histological typing, location and results of surgical and oncological treatment in 76 children with intracranial tumors presenting during the first 2 years of life are reported from the Hopital Neurologique et Neurochirurgicale Pierre Wertheimer, Service de Neurochirurgie Infantile, BP Lyon Montchat, Lyon, France. Patients were analyzed in groups according to age (29 under 1 yr and 47 between 1-2 yrs) and malignancy. A slight male preponderance was observed in children up to 1 yr, while an equal sex distribution was found in the older group. Supratentorial tumors were most prevalent during the 1st year, and the posterior fossa location was most frequent after 1 year. Increased intracranial pressure and hydrocephalus were the chief clinical manifestations; medulloblastoma and ependymoma were most frequent in the highly malignant group and astocytoma in the low-malignant group.

The overall survival rate was 46% following operation; 22% recovered completely and have survived 8 mos to 14 yrs, and 13% have a mild neurological deficit. In those with medulloblastoma or ependymoma requiring radiotherapy, only 20% had favorable neuropsychological results. No patient with a highly malignant tumor operated on during the first year of life survived later. Brain irradiation in this age group leads to a severely handicapped child and chemotherapy is preferred for highly malignant tumors, especially when surgical excision is incomplete. Radical surgery is proposed as the ideal treatment. (Lapras C et al. Brain tumors in infants: a study of 76 patients operated upon. Child's Nerv Syst April 1988;4:100-103).

**COMMENT.** In 17 children with intracranial neoplasms presenting within the first 2 months of life, alterations in behavior, anorexia, vomiting, irritability, or unusual quietness were the most common symptoms and 10 had macrocrania and hydrocephalus (Jooma R et al. Surg Neurol 1984;21:165). The operative mortality was 40% and the total case mortality was 80%. These authors from the Hosp for Sick Children, Great Ormond St, London, felt that an aggressive approach to most of the cases in this age group was not warranted at present, and the radiation dose is limited by the increased sensitivity of the immature brain.

A first report of medulloblastoma in an 8-yr-old patient with Coffin-Siris syndrome (Rogers L et al. Child's Nerv Syst 1988;4:41), was diagnosed during an evaluation for neurogenic causes of apnea and feeding difficulties. This syndrome is a rare congenital disorder characterized by mental retardation, deficient postnatal growth, joint laxity, and brachydactyly of the 5th digit with absence of the nail bed. Several cases of Dandy Walker cysts and a case of brain-stem heterotopia have been described previously as complications of the syndrome.

## NUTRITIONAL DISORDERS

### FOOD ADDITIVES AND HYPERACTIVITY

Of 220 children referred to the Dept of Paediatrics, Royal Children's Hospital, Parkville, Victoria, Australia, because of suspected

hyperactivity, 55 were included in a 6-week open trial of the Feingold diet, 26 (47%) showed a placebo response, and 14 were identified as likely reactors. Of 8 who subsequently completed a double-blind crossover study (utilizing each child as his own control), 2 demonstrated a significant dependent relationship between the challenge and ingestion of azo dye colorings (tartrazine and carmoisine 50 mg) and behavioral change. Extreme irritability, restlessness and sleep disturbance rather than attention deficit were the common behavioral patterns associated with the ingestion of food colorings, as described by the parents in this study. The authors conclude that the inclusion of children in trials on the basis of attention deficit alone may miss some reactors, and there is little place for use of a coloring-free diet in children with ADD unless the other behavioral features of irritability, restlessness and sleep disturbance are present. (Rowe KS. Synthetic food colourings and 'hyperactivity': A double-blind crossover study. Aust Pediatr J. April 1988;24:143-147).

**COMMENT.** The phoenix of the Feingold diet rises again with the suggestion that the treatment has been erroneously discarded because of inappropriate behavioral rating instruments and failure to identify specific reactors to food additives. In England, where the avoidance of all foods containing additives is widespread, the major problem is the level of public misinformation, occasionally leading to handicapping dietary restriction. (David TJ. Arch Dis Child 1988;63:582).

#### **BIOTINIDASE DEFICIENCY AND SEIZURES**

Preliminary experiences with screening of 24,300 newborns detected 1 infant with biotinidase deficiency at the Depts of Paediatrics, Univ of Verona, Policlinic Borgo Roma, Verona, Italy, and the Hosp for Sick Children, Toronto, Canada. The patient was a full-term baby girl with uncomplicated delivery and a positive family history for seizures in an aunt who had died at 8 months of age. At 2 months of life, the infant developed dermatitis and sparse scalp hair followed by multifocal motor seizures resistant to anticonvulsant drugs. Neurological exam showed hypertonia and hyperreflexia, the EEG revealed increased slow wave activity, and the CT finding was a mild cortical atrophy. Large amounts of 2-oxoglutarate and small amounts of 3-hydroxyisovalerate were found on chromatographic examination of the urine. Treatment with 10 mg/biotin daily resulted in complete recovery within 2-3 days. (Burlina AB et al. Neonatal screening for biotinidase deficiency in north eastern Italy. Eur J Pediatr April 1988;147:317-318).

**COMMENT.** The authors consider that biotinidase deficiency is as common as other well-known metabolic disorders and satisfies all criteria for inclusion into neonatal mass screening programs for inborn errors of metabolism. The absence of the expected organic acidopathy noted in the present case-report confirms the need for biotinidase enzyme estimations in diagnosis.