

COMMENT: This syndrome was first described by Segawa M et al in Japan (Therapy 1971;24:667) and should correctly be referred to as "Segawa Syndrome". Diurnal fluctuation of the dystonia is not invariably present and a trial of levodopa is worthwhile in possible variants of this dystonic syndrome. Emotional disturbance is a feature in some cases and may lead to a diagnosis of psychogenic etiology. In fact, in all cases of dystonia musculorum deformans (torsion dystonia) that I have treated, a diagnosis of conversion hysteria had previously been entertained and psychotherapy prescribed.

CONGENITAL CNS DEFECTS

HYDROCEPHALUS AND SHUNT INFECTIONS

In the 10-year period, 1973-82, 431 children underwent cerebrospinal fluid shunt insertion for hydrocephalus at Children's Memorial Hospital, Chicago. The authors, now in Verona, Italy (Casella Postale 401.1-37100), have studied the relationship between the etiology of hydrocephalus, age at the time of shunt placement, and infection etc. Meningomyelocele was present in 40%, congenital communicating or obstructive hydrocephalus in 34%, and tumors in 18%. Intraventricular hemorrhage and meningitis were the causes in 5% and 3%, respectively. The age at surgery was less than 1 year in 83% and 1 week or younger in 18%. Each patient had an average of 3 procedures. Infections occurred as a complication of the shunt in 96 patients at rates of 22% per patient and 6% per procedure. Younger patients and those with meningomyelocele were most susceptible to infection. In the meningomyelocele group, infection occurred less often when shunted at 2 weeks of age or later, compared to 1 week or earlier, when the rate was 48%. (Ammirati M, Raimondi AJ. Cerebrospinal fluid shunt infections in children. Child's Nerv Syst 1987;4:106-109).

COMMENT: The rate of operative shunt infection reported in this study is high, and the authors are able to cite similar statistics from two other centers. Attempts to reduce the incidence of infection by perioperative antibiotics or a surgical isolator had not been successful. If a rate of infection of 20% or more per patient is the rule with the operative treatment of hydrocephalus, a reappraisal of techniques and indications for surgery would seem to be a necessity.

Recent experience at Children's Memorial Hospital indicates a rate of infection lower than that reported here, and Dr. Luis Yarsagaray at Loyola Stritch Medical Center, Chicago, recalls only 3 cases of shunt infection in a total of 2000 patients of all ages, both children and adults, that he has himself treated by surgery over a 17 year period (personal communication).

ARNOLD-CHIARI WITH MYELOMENINGOCELE

The outcome of 19 infants with complications of Arnold-Chiari malformation and meningomyelocele was reviewed at the Depts. of Pediatrics, Pathology, and Neurosurgery, University of Pennsylvania School of Medicine and the Children's Hospital of Philadelphia. Vocal cord paralysis and inspiratory stridor alone occurred in 10 (grade I), apnea was an additional symptom in 4 (grade II), and cyanotic spells and dysphagia were associated in 5 (grade III).

Ventricular shunt was performed in 14 infants, with resolution of symptoms in 7 (in 5 of 8 with grade I, 2 of 4 with grade II, and none of 2 with grade III symptoms). Of 10 with posterior fossa decompressions, symptoms resolved in only 2 (in 1 of 4 with grade I, one of 2 with grade II, and none of 4 with grade III symptoms). Within 6 months after symptoms began, one infant with grade II and 3 with grade III died. No deaths occurred with the grade I group. Infants with grade II or III symptoms have more extensive brain stem damage, such as hemorrhage, infarction and necrosis, and carry a poor prognosis whereas those with grade I symptoms often improve after neurosurgical procedures. (Charney EB et al. Management of Chiari II complications in infants with myelomeningocele. J. Pediatr 1987;111:364-71).

COMMENT: The grading of cases according to complications is useful in investigation, treatment and prognosis. In a previous study from the University of Toronto (Park TS et al. Neurosurgery 1983;13:147), decompression was recommended before rapid neurologic deterioration takes place, even if a functioning shunt is present. Of 45 infants with surgical decompression of the Chiari malformation, 28 survived and showed improved neurologic function and in 24 of these, recovery was complete. About 71% died of those patients who developed cardiorespiratory arrest, vocal cord paralysis, or arm weakness within 2 weeks before decompression, compared with 22% of those with more gradual neurologic deterioration.

BRAIN TUMORS

SUBARACHNOID HEMORRHAGE FROM BRAIN TUMORS

Six children with subarachnoid hemorrhage as the initial symptom of brain tumor are reported from the Depts of Neurosurgery, Univ. of Occupational and Environmental Health, Kitakyushu, and Kumamoto Univ Med Sch, Kumamoto. They represented a 3.6% of 167 new pediatric cases of brain tumor seen in 7-17 years at 2 centers in Japan. Two neonates presented with irritability, vomiting, cyanotic spells, and unilateral facial paresis. Four children, ages 4 to 15 years, developed sudden headache and vomiting with or without alteration of consciousness. The tumor locations were posterior fossa (2 medulloblastomas, one ependymoma, one hemangioma) and hypothalamus (one astrocytoma and one unverified). All were located close to the III or IV ventricles. The ultimate prognosis was poor. (Yokota A et al. Child's Nerv Syst 1987;3:65-69).

COMMENT: Medulloblastoma is more apt to bleed than other neuroectodermal tumors in pediatric patients. Compared to brain tumors in adults, those in children bleed more frequently and are more commonly located in the posterior fossa. Brain tumor should be considered as a possible etiology of subarachnoid hemorrhage in the neonate and child.

MUSCLE DISEASE

CONGENITAL MYOTONIC DYSTROPHY

Ten infants with congenital myotonic dystrophy admitted to the Dept