DEVELOPMENTAL DISORDERS

CHANGING INCIDENCE, OUTCOME AND MANAGEMENT OF MYELOMENINGOCELE

Pediatric neurosurgeons at Children's Memorial Hospital, Chicago, review their long-term experience and the evolution of the etiology, diagnosis and management of patients born with myelomeningocele (MM) in 1975-1979 and followed for 25 years in a multi-disciplinary spina bifida clinic. Genetic factors and folic acid deficiency are implicated in the etiology of neural tube defects. Supplementation with folic acid has significantly reduced but not eliminated the risk of MM. The prevalence of spina bifida worldwide ranges from 0.17 to 6.39/1000 live births. Incidence varies with gender (girls > boys), ethnicity (increased in Hispanic and Northern Chinese), age (< 20 and >30 years more susceptible), geographic areas (in N America, higher in east and south cf west), and nutritional factors. Incidence was declining before use of folic acid, but following periconceptual multi-vitamin supplementation, a significant decrease occurred (MRC, 1991). Since 1992, US center for disease control recommends 400 mcg folic acid daily for women of reproductive age, and by 1998, fortification of all grain products. Incidence in US decreased 22.9%, when comparing period 1995-6 to 1998-9. Elective termination of pregnancy following prenatal diagnosis by serum alpha-fetoprotein screening and ultrasonography is also responsible for a declining incidence. The "lemon" sign and "banana" sign of Chiari II malformation are present on ultrasound after the 12th postmenstrual week. Fetal surgery for MM may arrest leakage of spinal fluid and prevent or reverse Chiari II malformation and hydrocephalus. Selective treatment for MM, as advocated by some authorities in the 1970, was generally not followed in the US, and today most centers treat all viable newborns aggressively without selection. The outcome of in utero closure awaits evaluation by the management of MM (MOM) study group.

Long-term outcome of non-selective treatment of 118 infants with MM born 1975-79, and followed at Children's Memorial Hospital, compared to a cohort born in 2000-2005, found a decline in the number of live births with MM: 16-32/year in the older cohort vs 1-13/year in the younger cohort of 40 children. The overall mortality in the older cohort at 20-25 year follow-up was 24%, the majority of deaths (18/28) occurring in infancy and pre-school years, secondary to hindbrain dysfunction or shunt malfunction. In the younger cohort, none have died during infancy and early childhood. Shunt placement has decreased from 86% in the older 1975-79 group to 65% in children born 2000-2005. (In a 2008 report from Great Ormond Street Hospital, London, UK, the rate of shunt placement is now 51%). The increased survival rate presents a challenge for pediatric and adult healthcare providers. (Bowman RM, Boshnjaku V, McLone DG. The changing incidence of myelomeningocele and its impact on pediatric neurosurgery: a review from the Children's Memorial Hospital. **Childs Nerv Syst** July 2006;25:801-806). (Respond: Dr Robin M Bowman, Division of Neurosurgery, Children's Memorial Hospital, 2300 Children's Plaza, PO Box 28, Chicago, IL 60614. E-mail: RBowman@childrensmemorial.org).

COMMENT. As with many congenital or early childhood chronic nervous system diseases, the transition period from pediatric to adult care poses problems. Pediatric subspecialists are frequently unfamiliar with adult care. Pediatric clinics are often

geographically separated from clinics for adults. Patients have difficulty separating from a team that has cared for them for two decades. The authors comment that shunt malfunction is a primary consideration in long-term care of MM, and adult colleagues need to become familiar with this problem. Learning and attention deficit disorders are additional troublesome complications of MM that require specialized treatment.

BRAIN TUMORS

CHANGING EPIDEMIOLOGY OF PEDIATRIC BRAIN TUMORS

Neurosurgeons at the Hospital for Sick Children, Toronto, Canada, analyzed and classified 1, 866 surgical pathology cases of brain tumors in children under age 19 years, treated 1980-2008. Astrocytomas accounted for 39.4%, (low-grade I/II 32.3%, grades III/IV 7.1%), medulloblastoma (10.6%), ependymoma (7.0%), craniopharyngioma (6.8%), meningioma (1.7%), and hypothalamic hamartoma (1.6%). Male preponderance (56.8%) occurred in all age groups, and particularly with medulloblastoma (M/F, 3/2). Classified by age group, ependymomas peaked at 0-2 years, and medulloblastoma at 3-5 years; astrocytomas increased in prevalence up to 9-11 years and then decreased. Distribution of tumors over the 3 decades showed little variation, except for medulloblastoma that showed a decreased percentage in 1990-99, and pilocytic astrocytoma (grade I) that increased steadily from 0.36% in 1980-89, to 7.32% in 2000-2008. The findings were consistent with published series from other countries, and changes in epidemiology may be attributed to changing classification systems, improved imaging and developments in epilepsy surgery. Under-reporting in the older groups (>15 years) may occur due to referrals to adult centers. (Kaderali Z, Lamberti-Pasculli M, Rutka JT. The changing epidemiology of paediatric brain tumours: a review from the Hospital for Sick Children. **Childs Nerv Syst** July 2009;25:787-793). (Respond: Dr JT Rutka, Division of Neurosurgery, The Hospital for Sick Children, The University of Toronto, Suite 1503, 555 University Ave, Toronto, Ontario, Canada M5G 1X8. E-mail: james.rutka@sickkids.ca).

COMMENT. The National Cancer Institute registry lists 488 CNS tumors reported in children in the state of Connecticut over a 39-year period; 467 were intracranial and 21 were spinal. Astrocytomas accounted for 28%, medulloblastomas 25%, ependymomas 9%, craniopharyngiomas 9%, and glioblastoma multiforme 9%. (Farwell JR et al. **Cancer** 1977;40:3123-3132). The incidence of medulloblastoma in the Connecticut series is double that found in the Toronto patients, whereas the percentages of other CNS tumors are similar. Of 6 series of childhood brain tumors (1949-1980) tabulated by Cohen ME and Duffner PK (**Brain Tumors in Children: Principles of Diagnosis and Treatment**. New York, Raven Press, 1984), one series reports medulloblastoma in 10.6% of 425 cases, similar to the Toronto series, whereas 5 series show percentages varying from 16.3% to 25% (mean, 20%). Over a 30-year period the incidence of specific types of childhood brain tumors had not changed appreciably. With an emerging microRNA brain tumor classification, the epidemiology of childhood brain tumors becomes more complicated. The influence of environmental risk factors (ionizing radiation, chemotherapeutic agents) is thought to be small (Baldwin RT et al. **Toxicol Appl Pharmacol** 2004;199:118-131).