Management of Craniopharyngioma in Childhood*

DONALD D. MATSON, M.D., AND JOHN F. CRIGLER, JR., M.D.
Departments of Neurosurgery and Medicine (Endocrinology), The Children's Hospital Medical Center, and Departments of Surgery and Pediatrics, Harvard Medical School, Boston, Massachusetts

The purpose of this paper is to present briefly a summary of recent clinical experience at the Children's Hospital Medical Center in Boston with radical surgery for craniopharyngioma and then to discuss in more detail the complex fluid and steroid problems involved in management during the early postoperative period. In addition, mention will be made of the more significant neuroendocrine studies involved in conscientious long-term follow-up of these patients.

Material

In all, 74 patients with surgically verified craniopharyngiomas have been treated by this neurosurgical clinic.

Of these, 10 were treated prior to 1950 when adrenocorticotropic (ACTH) and the glucocorticoid, cortisone, became available for clinical use. In none of these patients was the tumor completely and successfully removed, and all subsequently died because of the tumor or attempts to irradiate or remove it. These patients will not be referred to further.

An additional seven patients were adults varying in age from 35 to 65 years when operated on. These constitute a separate group with different surgical indications, usually much simpler operative problems, and special postoperative plans for endocrine management. They, also, will be excluded from further discussion.

This leaves a group of 57 children treated since 1950, 37, or two-thirds, of whom have remained under long-term care or have been regularly followed by the endocrinological and neurosurgical staffs.

The age and sex distribution of this group of pediatric craniopharyngioma patients since 1950 are shown in Fig. 1. Why do these children come for medical care? Their clinical symptoms and signs are seen in Fig. 2; it should be emphasized these are all initial findings, that is, prior to any medical or surgical treatment. The absence of diabetes insipidus as a preoperative complaint should be noted.

Radiological investigation confirms the diagnosis in this age group with great precision (Fig. 3). All but one of these 57 children showed suprasellar calcification. Approximately half also showed some abnormality of the sella turcica itself on plain X-ray films, and many showed separation of the cranial sutures. Contrast studies have proved of limited value and are being done less and less. Angiography has virtually been given up whenever preoperative calcification is present, and air studies are done only if there is clinical or electroencephalographic evidence of a large extension into the frontal or middle fossa. Under these conditions, air studies may help determine the best side for a surgical approach.9

Surgical Management

Since replacement steroid therapy has been available, it has been the policy in this clinic to attempt total removal of every craniopharyngioma occurring in a child.8 It has been our impression that the younger the child the easier the operation technically and the less complicated the postoperative course. Recurrence of symptoms has been so relentless and rapid in such a high percentage of cases seen in early childhood if the tumor is not radically removed, that our conviction becomes steadily stronger that optimum treatment consists in making every
effort to effect total excision at the first operation. As will be emphasized subsequently, almost all of our major intraoperative and postoperative problems, and all of our operative fatalities, have come with secondary and tertiary attempts to remove tumors that had again become symptomatic after a previous subtotal surgical procedure. All of these tumors, no matter where they go (into the sella, into the frontal, middle, or posterior fossa, or up into the hypothalamus and third ventricle), arise in the region of the pi-

Fig. 1. Sex and age of 57 children with craniopharyngioma.

FIG. 2. Clinical symptoms and signs in 57 children with craniopharyngioma.

FIG. 3. X-ray findings in 57 children with craniopharyngioma.