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Surgical Removal of Craniopharyngiomas by a Transcranial Approach through the Lamina Terminalis and Sphenoid Sinus

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We describe a method of attempting total removal of a craniopharyngioma using a subfrontal approach; dissection of the tumor from the brain stem, hypothalamus, and optic chiasm through the lamina terminalis; and then piecemeal removal of the tumor through the sphenoid sinus. Eleven patients treated by this approach are reported. In these patients, prior subfrontal operation did not affect outcome, but a prior transventricular operation or prior high dose radiation therapy made complete tumor removal less likely. We believe that this operation is one way to remove a craniopharyngioma that affords satisfactory exposure and reduces the chances of injury to the frontal lobes, hypothalamus, and optic pathways. (*Neurosurgery*, 7: 111–117, 1980)

Key words: Craniopharyngioma, Lamina terminalis, Radiation, Sphenoid sinus, Subfrontal approach, Transventricular approach, Tumor

INTRODUCTION

The management of patients with craniopharyngioma continues to challenge neurosurgeons despite recent advances in surgical technique, radiation therapy, and endocrine replacement therapy. Although the tumor seems indolent in some patients, this is not the rule, and in the majority of patients the tumor will ultimately recur after the conventional management of partial removal followed by radiation therapy (2, 4, 8, 11).

Matson, more than anyone else, drew attention to the difficulties associated with surgical treatment of craniopharyngioma. He showed that the removal of all visible tumor was possible with low risk in patients who had received no previous therapy (7). However, review of his series revealed that his radical operation, although salutory in many respects, had at least three disadvantages: the mortality rate was substantial for secondary operations, the tumor recurred in 26% of cases, and postoperative fluid and electrolyte imbalance often occurred and could be life-threatening (5). Consequently, interest remains high in alternative modalities of treatment, such as partial removal followed by improved forms of radiation therapy.

Since Matson's report the advent of the operating microscope has refined the surgical management of craniopharyngioma, and the results are easier to assess now that computed tomography is available (12). This has stimulated neurosurgeons to re-examine the possibilities of achieving a cure through operation alone, thereby eliminating the need for radiation therapy and its attendant risks.

The location and configuration of the tumor influences the choice of surgical approach. Tumors that are wholly intrasellar can be removed comfortably by the trans-sphenoidal route, whereas tumors that extend far laterally under the temporal lobe are perhaps best handled by a temporal approach. The majority of craniopharyngiomas, however, are located beneath the chiasm and extend up into the hypothalamus and 3rd ventricle, down between the brain stem and clivus, and anteroinferiorly into the sella. In these circumstances some surgeons recommend an approach along the sphenoidal ridge, whereas others favor a subfrontal approach, either obliquely over the roof of the orbit or along the falx. The purpose of this report is to describe a technique of removing these tumors that incorporates two maneuvers that are familiar to neurosurgeons but that, we believe, have not been used together systematically. In our experience this procedure has increased the likelihood of total removal and decreased the chances of damage to the hypothalamus and optic apparatus.

SURGICAL TECHNIQUE

This report is based on our experience with 11 patients who underwent the removal of a suprasellar craniopharyngioma by an approach through the lamina terminalis and sphenoid sinus.

The operative approach is much like that described by Ray for pituitary surgery, namely, through a small, low, unilateral bone flap on the right side above the orbit that spans the distance between the falx and the temporal line just above the supraorbital ridge (10). The right side is convenient for a right-handed surgeon, and a unilateral approach along the falx affords approximately equal exposure of both sides of the optic chiasm. If all that were required was exposure of the optic nerves and chiasm, the use of osmotic diuretics and the lumbar drainage of cerebrospinal fluid would provide ample room without the need for much retraction. However, because we favor approaching the tumor through the lamina terminalis behind the optic chiasm, it is wise in most cases to remove a small strip, perhaps 1 to 2 cm wide and 1 to 2 cm deep, of the undersurface of the frontal lobe from the frontal pole to the chiasm along the falx. In any case, retractor pressure on the frontal lobe must be minimized because much of the potential morbidity and mortality of the operation is a consequence of edema and hemorrhage in the frontal lobe due to excessive mechanical pressure.

After the optic nerves and chiasm are exposed, the first step is to remove the tuberculum sellae, whether the optic nerves are long or short. Removal of the tuberculum is accomplished easily by turning back a flap of dura from the planum sphenoidale, breaking into the sphenoid sinus with a chisel or a high speed drill, and then removing the tuberculum and anterior wall of the sella turcica with fine punches (9). Dividing the dura in front of the sella after coagulating the circular sinus exposes the sellar contents. Next, the frontal lobe and anterior cerebral arteries are retracted from the optic chiasm and lamina terminalis, and the latter is opened between the optic tracts. This maneuver, coupled with the previous removal of the tuberculum sellae, widely exposes the tumor on both sides of the optic chiasm (Fig. 1).

Most tumors are particularly adherent to the hypothalamus, almost as if they arose there, and between the tumor and the hypothalamus a true plane of separation rarely exists. In contrast, the tumor outside the hypothalamus is usually surrounded by a film of arachnoid and separates readily from the various arteries and the brain stem. The attachment to the optic chiasm is variable; sometimes the tumor separates easily, and at other times the tumor is adherent, although rarely so much as is the case with the hypothalamus.

The tumor is removed by pushing pieces down and away from the optic chiasm and hypothalamus toward the sphenoid sinus, from which the pieces can be extracted without fear of hurting the optic apparatus. The tumor in the hypothalamus is removed by working under direct vision between the optic tracts, although at times the plane of separation may be poorly defined. In most cases a complete removal can be achieved with a good margin of safety. At the end of the procedure the sphenoid and frontal sinuses are repaired by being packed with fat held in place by some bridging sutures.

CASE REPORTS

Case 1

J.S. is a 12-year-old boy who presented with a 1-year history of visual loss, headaches, nausea, and vomiting. His height was in the 3rd percentile and his weight was in the 25th percentile. Visual acuity was 20/200 in one eye and 20/100 in the other, and bitemporal hemianopia was present. A computed tomographic (CT) scan, radionuclide scan, pneumoencephalogram (PEG), and angiogram revealed a large suprasellar tumor almost completely filling the 3rd ventricle (Fig. 2, *A* and *B*). Partial removal of the craniopharyngioma was achieved by a transcallosal approach. Postoperatively a shunting procedure was necessary. Eleven months later the patient returned, blind in the left eye and with vision reduced to finger counting on the right. A presumed total removal was accomplished in 1977 by the approach described above. Postoperatively the patient lost the remainder of his vision, and he remained somewhat apathetic. He requires complete endocrine replacement therapy and tends to have a high serum sodium level, which is managed satisfactorily with arginine vasopressin. CT scans reveal residual tumor that has shown no signs of growth to date (Fig. 2*C*).

Case 2

W.D., a 12-year-old girl, underwent the partial removal of a craniopharyngioma followed by postoperative radiation to a tumor dose of 3000 rads. Her postoperative vision was 20/70 in one eye and 20/20 in the other, with a right homonymous hemianopia. A ventriculoatrial shunt was performed at age 22. At age 26, her vision deteriorated to light perception in the left eye and 20/30 with useful vision restricted to the inferior nasal quadrant of the right eye. A total removal of the tumor was accomplished as described above. Postoperatively her vision remained 20/30, but panhypopituitarism coupled with diabetes insipidus and a defective thirst mechanism resulted in periodic hypernatremia. This latter symptom was controlled satisfactorily with arginine vasopressin, and the patient returned to work. Postoperative CT scans showed no residual tumor. At age 29 she died of a pulmonary embolus, and no residual tumor was found at postmortem examination.

Case 3

J.N., a 46-year-old woman, had a 3-month history of visual loss. She had been hypothyroid and mildly diabetic for about 5 years. Her visual acuity was 20/100 in both eyes, with an incomplete bitemporal hemianopia. A CT scan, PEG, and

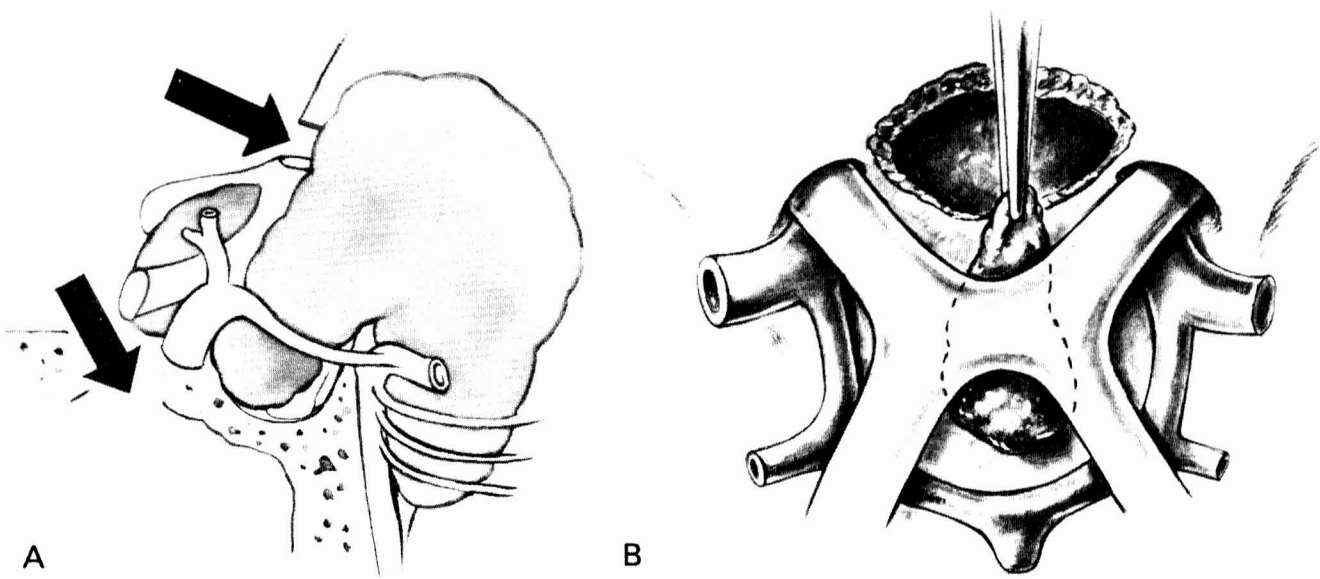

FIG. 1. *A*, approach to the tumor between the optic tracts through the lamina terminalis; *A* and *B*, removal of tumor by way of the sphenoid sinus.

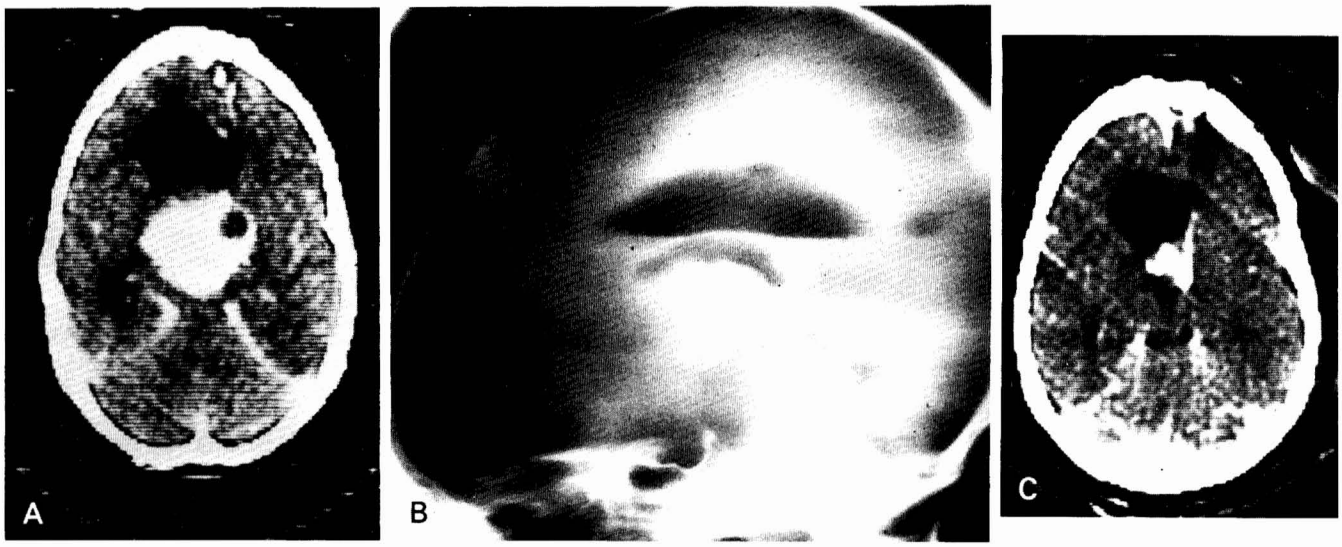

FIG. 2. Case 1. CT scan (*A*) and pneumoencephalogram (*B*) before operation. *C*, residual tumor after operation. This is the only patient in this series in whom residual tumor was identified.

angiogram revealed a suprasellar tumor, which was removed completely. Postoperative acuity was 20/25 in the left eye and 20/20 in the right eye. Her visual fields returned to nearly normal. Three years later a possible slight change in the visual fields was noted, but a CT scan, a PEG, and the cerebrospinal protein content were normal. The patient is maintained on thyroxine, prednisone, insulin, and vasopressin, and she leads a normal life.

Case 4

F.S., a 14-year-old boy, had undergone the subtotal resection of a craniopharyngioma the previous year because of impaired visual acuity and bitemporal hemianopia. Postoperatively his visual acuity became normal, but a temporal defect persisted in one eye. He required thyroid, adrenal, and gonadal hormone replacement therapy and also arginine vasopressin. Because a CT scan revealed a substantial amount of residual tumor (Fig. 3*A*) and the endocrine deficit seemed to be increasing, he underwent reoperation in June 1977, and a total removal was achieved. Postoperatively his vision did not change, and the CT scan showed no residual tumor (Fig. 3*B*). He now functions normally.

Case 5

R.N., a 52-year-old woman, presented with a 2-month history of visual loss and a bitemporal hemianopia. Skull films and a CT scan were normal. Sixteen months later she returned with an increase in the visual field defect and a reduction in acuity to 20/30 in one eye. Acuity in the other eye could not be evaluated because of a congenital defect. A repeat CT scan was normal, but a PEG revealed a suprasellar mass, which was removed totally. The postoperative course was stormy due to difficulties in the regulation of fluid and electrolyte balance. The patient requires long term maintenance on adrenal and thyroid hormone replacement and vasopressin. Her visual acuity and fields were unchanged postoperatively, and she now functions normally.

Case 6

H.S., a 32-year-old man, developed visual impairment and diminished libido. His visual acuity was reduced to 20/60 in both eyes, and a partially calcified suprasellar mass was identified. The following year the tumor, which was partly cystic, was removed subtotally and radiation therapy was started but then abandoned because of confusion and headaches. The patient required cortisone, thyroid, and gonadal hormone supplements. Two years later a temporal defect developed in the right eye, and a second operation was performed, during which a recurrent cyst was evacuated. The patient was then well until age 39 when his visual acuity fell to 3/100 in the left eye and a right homonymous hemianopia appeared. He underwent reoperation, and total removal of the tumor was achieved by the approach through the lamina terminalis and sphenoid sinus. A mild diabetes insipidus developed, but otherwise his postoperative course was unremarkable, and he was discharged on the 15th postoperative day. Visual fields and acuity returned to normal in the right eye, but vision was reduced to counting fingers at 10 ft in the left eye. The diabetes insipidus subsided and required no treatment. He returned to work as a television producer; follow-up CT scans revealed no tumor (Fig. 4).

Case 7

J.L., a 40-year-old man, developed decreased libido and other signs of hypopituitarism at age 30; he was treated with thyroid and gonadal hormones. Destruction around the sella that was evident on skull films had led to the diagnosis of pituitary adenoma. A CT scan revealed a partially calcified intrasellar mass with a large suprasellar extension. A total removal was performed, preserving the pituitary stalk. The postoperative course was benign, and the patient was given prednisone, thyroxine, and testosterone; diabetes insipidus did not develop. A repeat CT scan performed 1 year later was normal.

Case 8

At age 43, D.H. presented with a 2-year history of weight gain, amenorrhea, and galactorrhea. Examination revealed a visual acuity of 10/30 in one eye and 10/20 in the other, with an incomplete bitemporal hemianopia. The patient was started on radiation therapy, but this was stopped because of severe headaches. She underwent operation and a partial removal of the tumor was achieved. Her vision returned to normal, but at age 50 her vision deteriorated to 20/50 in one eye and 20/30 in the other, with an incomplete bitemporal hemianopia. The

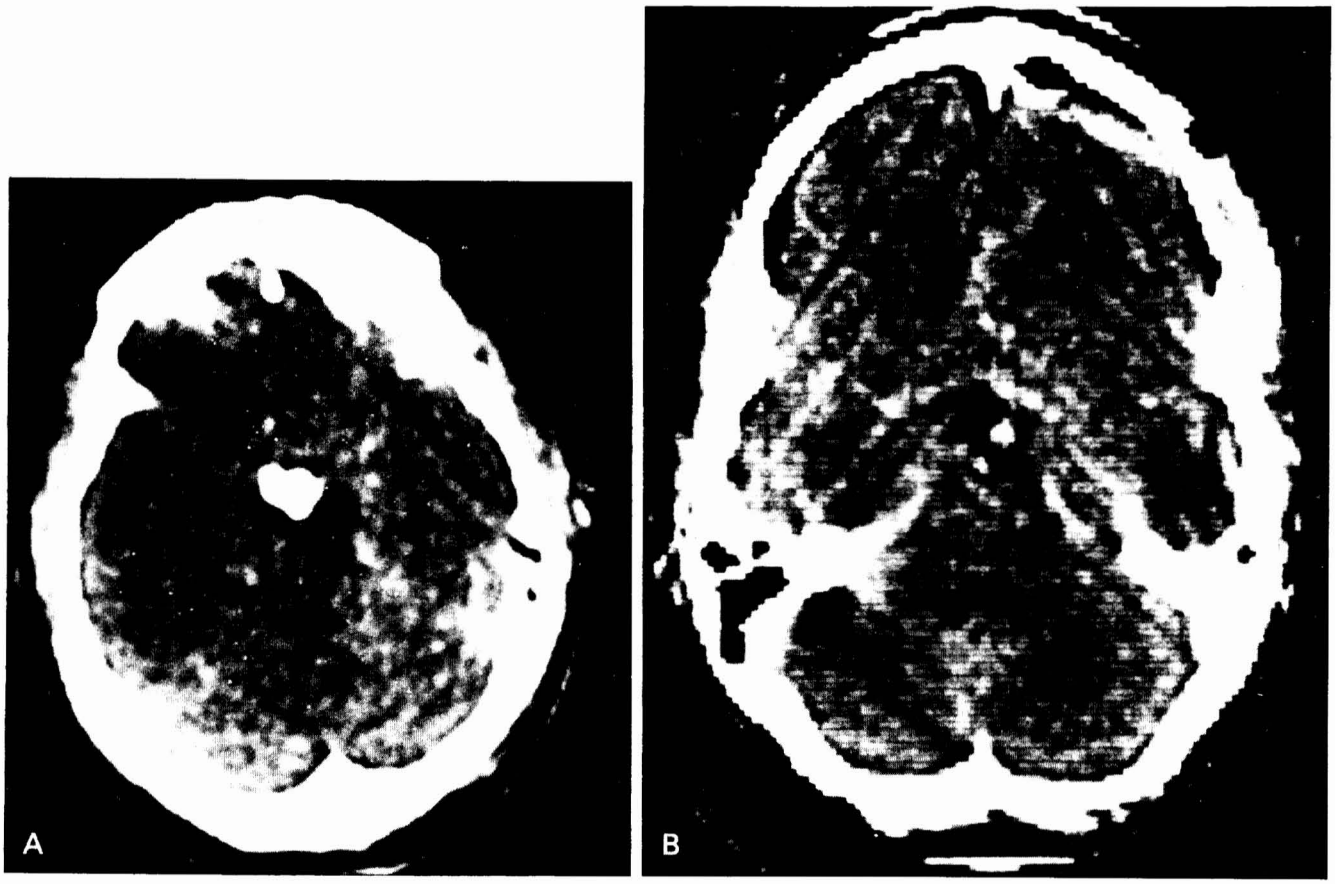

FIG. 3. Case 4. *A*, enhanced CT scan after partial removal of the tumor and before the second operation. *B*, enhanced CT scan after removal of persistent tumor through the lamina terminalis at a second operation.

patient underwent reoperation and a total removal was accomplished by the methods described above. Her visual acuity improved to 20/25 and 20/20. Her visual fields returned to normal in one eye, but a quadrantanopia remains in the other. She requires prednisone, thyroxine, and lypressin spray, but otherwise lives a normal life.

Case 9

A.S., a 48-year-old woman, had undergone the partial excision of a craniopharyngioma in another institution 14 months earlier after a 4-year history of panhypopituitarism and visual loss. Postoperatively she had had temporary difficulty with fluid and electrolyte balance and had been treated with 5750 rads of radiation therapy. Further deterioration of vision to counting fingers with one eye and seeing hand motions with the other occurred 6 weeks before her admission to The New York Hospital. A second operation was performed as described above, and the tumor, optic nerves, and hypothalamus were found to be densely adherent and discolored yellow, suggesting radiation changes. Only a partial removal could be achieved. Postoperatively, difficulties with fluid and electrolyte balance required treatment with arginine vasopressin. The patient's vision did not improve, and she requires custodial care.

Case 10

E.S., a 50-year-old woman, presented with a 2-month history of visual loss. Examination revealed an incomplete left homonymous hemianopia with normal visual acuity. Computed tomography and pneumoencephalography confirmed the presence of a suprasellar mass. At operation, the entire tumor was removed as described above except for a small scrap that was densely adherent to the hypothalamus. This scrap was left in place for fear of inducing serious difficulties with water balance. Postoperatively the visual fields returned to normal, and the patient was treated with 5000 rads of radiation therapy. The CT appearance is now normal. The patient takes thyroxine, steroid hormone, and arginine vasopressin and works full-time as a lawyer.

Case 11

L.L., a 7-year-old girl, complained of headaches and watery eyes, and her mother reported lethargy and increased thirst since 3 weeks before admission. Examination revealed that the patient was at the 50th percentile in height and weight. Her visual acuity was 20/20 and 20/40, diplopia was present on left lateral gaze, and her visual fields were normal. Computed tomography and angiography demonstrated a large suprasellar mass. At operation, a total removal of the tumor was achieved without difficulty. The postoperative course was smooth, and the patient was treated with thyroid and steroid hormones and arginine vasopressin. Postoperative computed tomography revealed no residual tumor.

DISCUSSION

Because of the risk of operation, particularly radical surgery, some physicians interested in the management of craniopharyngioma advocate that patients be treated with either radiation

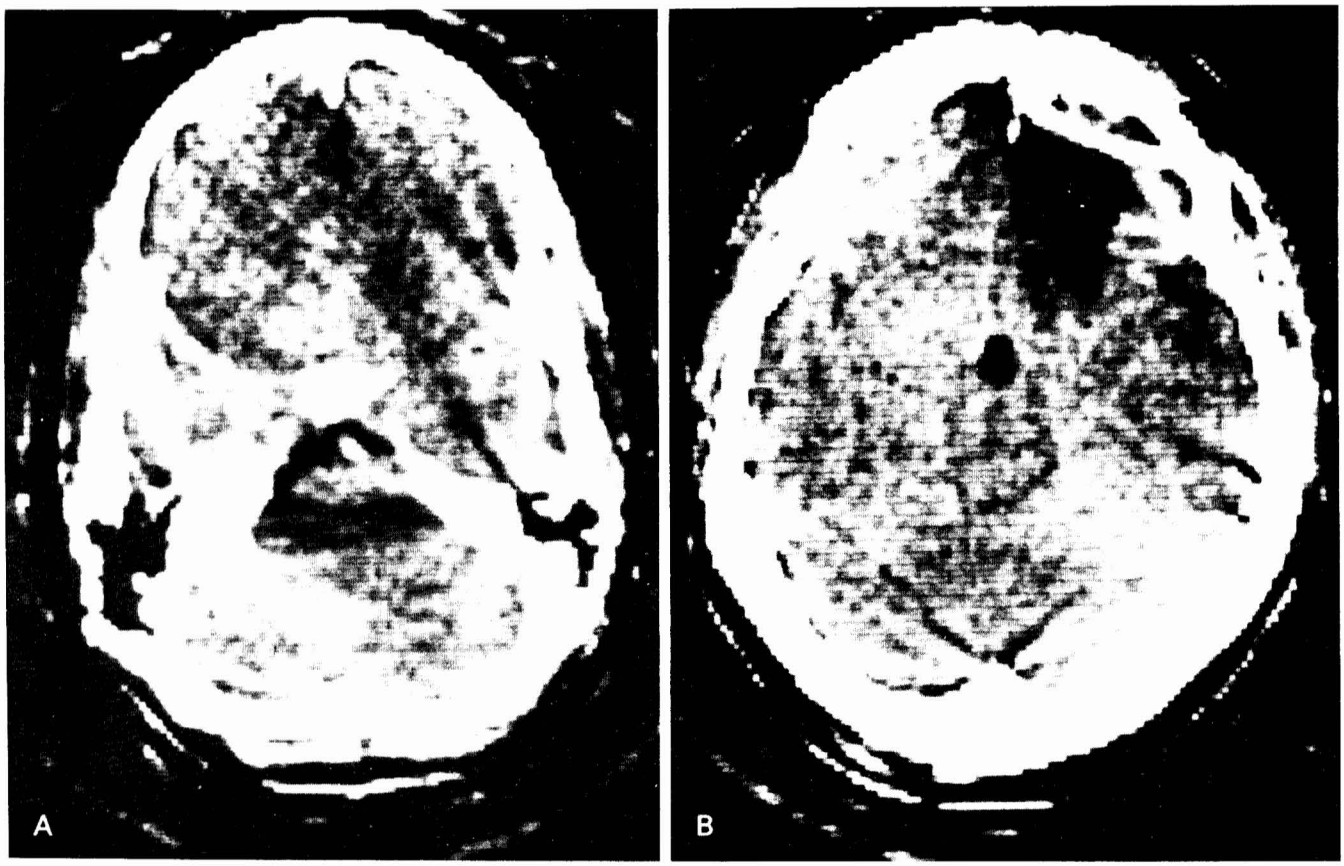

FIG. 4. Case 6. *A*, enhanced CT scan after two previous operations; *B*, enhanced CT scan after removal of recurrent tumor through the lamina terminalis at a third operation.

therapy alone or a conservative operation followed by radiation therapy. Bloom reported 5- and 10-year survival rates of 76 and 61%, respectively, for patients treated with operation and conventional radiation therapy at the Royal Marsden Hospital. The 5-year survival rate was increased to 96% of 29 patients treated with a conservative operation and radiation from a 6-MeV linear accelerator (1). Kramer has reported a similar series with 30 long term survivors and 13 patients who died. Some of the deaths were due to recurrent tumor and others were associated with radiation necrosis (6). Such important information as the functional status of the survivors and whether the tumor recurred is not detailed in either of these series.

The New York Hospital experience with partial removal and radiation therapy of craniopharyngiomas was reported by Hoff and Patterson in 1972 (2). The tumor recurred in 72% of the patients within 5 years after partial removal alone, and radiation therapy in the range of 3000 to 4500 rads reduced this figure to 41% (Table 1). These unsatisfactory results prompted us to attempt more aggressive surgery in managing patients with craniopharyngioma. Although some earlier surgical series reported a substantial mortality rate for even partial removal of a craniopharyngioma and the mortality rate in most series has been high for operations on recurrent tumor, our experience indicates that these tumors can be operated upon safely. The only death in 85 craniopharyngioma operations at The New York Hospital since 1950 occurred in a newborn child who survived the operation for craniopharyngioma but succumbed in the 3rd week of life to gastrointestinal hemorrhage from esophageal varices brought about by an unsuspected malignant

TABLE 1
*Results of Partial Removal and Radiation Therapy for Craniopharyngioma**

Follow-up Period (yr)	Operation Alone		Operation and Radiation	
	No. patients	Recurrence rate	No. patients	Recurrence rate
2	18	66%	23	30%
5	18	72%	22	41%
10	18	89%	19	63%

* From the data of J. T. Hoff and R. H. Patterson: Craniopharyngiomas in children and adults. J. Neurosurg., 36: 299–302, 1972.

tumor of the liver. Gentle handling of the frontal lobe, we believe, is the key to a low operative mortality rate.

Tumor recurrence looms next in importance to surgical mortality as a major problem. As outlined above, failure to remove all of the tumor usually occurs because pieces are left in the hypothalamus. Matson described teasing the tumor out between the optic nerves, but the fact that tumor persisted in 26% of 34 patients operated upon once and in 50% of patients operated upon for recurrence underscores the inadequacy of this technique (6, 7). Some have favored an approach along the sphenoid ridge, and this does allow a degree of access behind the tumor, but the optic pathways and carotid artery on the side of the operation are in the way, and the exposure of nerves and arteries on the contralateral side is poor. For this reason, we favor the approach along the falx. Hoffman and

coworkers have recommended that the lamina terminalis be incised to gain exposure if the optic nerves are short (3). This maneuver deserves extension to all cases because it affords good access to the tumor in the hypothalamus and thereby greatly increases the likelihood of achieving surgical cure.

Regardless of the approach to the tumor, a substantial risk to the optic nerves, optic chiasm, and optic tract exists when hard portions of the craniopharyngioma are removed from between either the optic nerves or optic tracts. Each time a fragment of tumor is extracted it is likely to brush against the visual apparatus. This can be prevented if the tuberculum sella is removed in all cases; then the solid portion of the tumor can be pulled down and away from the optic nerves and chiasm and out through the sinus without the risk of harming vision.

Of our 11 patients, 6 had undergone one or more prior operations, and only 5 patients had received no prior treatment. All 11 patients resumed full activity postoperatively except 2 who were virtually blind before operation and who continue to be handicapped. Complete tumor removal was apparently achieved in all but 3 patients, as judged by postoperative roentgenographic contrast studies and by autopsy in the patient who died of a pulmonary embolus several years after operation. Incomplete removal in the first of these patients was a consequence of the dense scarring that followed a prior transventricular approach to a giant tumor in the 3rd ventricle. In the second patient a small scrap was left knowingly in the hypothalamus for fear of inducing an intolerable deficit, and in the third patient prior radiation therapy in a dose of 5750 rads had obliterated the usual planes and caused dense adhesions between the brain and the tumor, as well as gliotic changes in the optic nerves. This radiated patient, as well as Patient 2, who had been treated previously with 3000 rads, had difficulty with fluid balance after operation; no difficulty was experienced in the 2 patients who had received an incomplete course of radiotherapy. We conclude that prior radiation therapy in high doses or a prior transventricular operation reduces the chances of obtaining a complete removal and increases the chances of inducing diabetes insipidus coupled with adypsia. On the other hand, prior operation by a subfrontal approach does not preclude total removal at a subsequent operation.

Although it seems feasible to remove a craniopharyngioma in most cases, one can question the wisdom of attempting removal in a growing child whose growth hormone secretion is normal. Such circumstances are not common, but if they occur and the tumor is small, perhaps partial removal and radiation therapy might be a better choice in hope that the child will achieve greater height. However, if the tumor is large, the secretion of growth hormone is impaired, the child is a suitable height, or the tumor separates easily from the hypothalamus, then we would favor an attempt at complete removal.

With respect to postoperative radiation therapy, we recognize that, despite a normal CT scan, residual tumor might still be present but be either too small to show or lost in artifacts caused by the posterior clinoid processes. However, radiation therapy is not always innocuous, and we have preferred to withhold treatment in patients in whom gross total removal has been accomplished if the postoperative scan shows no residual tumor.

In conclusion, we believe that the total removal of craniopharyngioma is possible and can be accomplished with an acceptable morbidity rate if measures are taken to deal with the tumor in the hypothalamus and to protect the frontal lobe and optic pathways. We have outlined an operative approach that meets these goals.

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COMMENTS

I was pleased to read about the authors' success in achieving total resection of craniopharyngiomas even of large bulk using their unique approach. I have long thought that the subfrontal approach is the most efficient and successful means of removing craniopharyngiomas and was accordingly gratified to learn of their excellent results.

In cases where the tumor is small and the optic nerves are not prefixed, I question the necessity for strip resection of the undersurface of the frontal lobe (with the consequent risk of producing epilepsy), opening of the sphenoid sinus (with the risk of bacterial contamination), or opening of the lamina terminalis (with the risk of impairment of the vascular supply of the optic chiasm). With large tumors that protrude high into the 3rd ventricle and displace the chiasm forward, thus shortening the optic nerves, I have found that removal of the bulk of the tumor may necessitate opening of the lamina terminalis. I have not had occasion to use the authors' approach, but I think that it would be truly worthwhile in dealing with large solid and calcified tumors that require large ports of exit which cannot be found between optic nerves, optic tracts, or carotid artery and optic nerve.

As the authors state, craniopharyngiomas are adherent to the hypothalamus in the region of the tuber cinereum and, in my own experience, I have been unable to remove a craniopharyngioma totally with complete preservation of the pituitary stalk and with the patient being free of diabetes insipidus.

My initial impression of craniopharyngiomas was that total resection was possible only at the first operation. However, like the authors, I have found that secondary surgery may be successful if a different operative approach than that of the initial procedure is used.

The authors' success with secondary resection of large recurrent tumors is very impressive. Of the 11 cases that they describe, 9 had total removal of the tumor and 5 of these were

achieved at the secondary operation. These excellent results must, in part, be due to the unique operative approach.

I take issue with the reluctance of the authors to remove a craniopharyngioma in a growing child with normal growth hormone secretion. They suggest that partial removal and radiotherapy might be a better choice in such a patient, but in our institution we have found that low dose radiotherapy given supratentorially to children with medulloblastoma will cause hypothalamic damage in a majority of patients and lead to impaired growth hormone secretion. Further, in 25% of our patients whose craniopharyngiomas were removed totally, normal growth continued despite the lack of growth hormone secretion or growth hormone therapy, indicating that growth arrest should not be considered an inevitable or universal postoperative problem. In cases where growth does become retarded, the administration of human growth hormone is effective and seems far preferable to the dangers of radiotherapy.

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This seems to me to be an important communication. It makes the point (for the first time of which I am aware) that, in a significant number of patients with craniopharyngioma in all age groups, removal can be facilitated by removing the roof and anterior wall of the sphenoid sinus. This gives added room for a subfrontal extraction piecemeal from anteroinferior to the optic chiasm. I am convinced that the advocated removal of these boundaries of the sphenoid sinus should be a standard procedure if there is any intrasellar extension of tumor, the anteroinferior limits of which cannot be seen via the transfrontal route. However, in 23 of my first 37 patients I thought that the tumor lay mainly within the 3rd ventricle. One can usually identify and remove anteroinferior parts of the capsule in these patients as one removes the tumor through an opening in the lamina terminalis above the chiasm. In such cases I do not see how opening the sphenoid sinus would aid the operation. On the other hand, in at least 3 of the recurrences after my attempted total removal I think that the regrowth was from an anterior intrasellar remnant not seen directly during the original operation. Therefore, in the past few years I have been following the course recommended by Patterson and Danylevich if there is an anterior intrasellar component to the tumor. Infection consequent upon opening the sphenoid sinus has not materialized. I have sought to keep the sphenoid sinus mucosa intact and to displace it downward and forward against the ostium of the sinus as an additional barrier against infection provided by the overlying fat graft.

In some craniopharyngiomas the tumor grows extensively beneath the temporal lobe and/or back and down into the posterior fossa, around the brain stem along a posterior cerebral artery, or laterally into a sylvian fissure. Reasonably obvious modifications in the surgical approach are required—perhaps even including a second stage if the awkward extension comes as a surprise. The visualization of small recesses into which the tumor extends may be aided by a tiny mirror mounted on a flexible silver shaft so that its angle can be varied as desired.

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