Craniopharyngioma in adults and children: a study of 122 surgical cases

RÉMY VAN EFFENTERRE, M.D., AND ANNE-LAURE BOCH, M.D.

Department of Neurosurgery, Groupe Hospitalier Pitié-Salpêtrière, Paris, France

Object. This work is devoted to a 25-year retrospective study of 122 cases of craniopharyngiomas in adults and children treated and followed by the same neurosurgeon (R.V.E.). In this homogeneous series, the aim was total microsurgical removal of the tumor, without postoperative radiotherapy.

Methods. The operation was performed via a frontopterional approach in 112 cases and a transsphenoidal approach in 10 cases. The tumor removal was considered total in 59%, subtotal in 29%, and partial in 12%. The surgical mortality rate was 2.5%. Even when tumor removal was not complete, radiotherapy was not systematically administered; it was reserved for cases of recurrence. The authors have studied clinical signs, operative characteristics, and ophthalmological, endocrinological, and functional outcomes, as well as recurrence risk and long-term patient survival. The mean follow-up period was 7 years. The functional results in these patients were excellent in 85%, good in 9%, fair in 5% (usually because of ophthalmological sequelae), and poor in 1%. Tumors recurred in 29 patients, but the salvage treatment, by operation or radiotherapy, was successful in 83%. The actuarial patient survival rate was 92% after 5 years and 85% after 10 years.

Conclusions. These results compared favorably with the data reported in the literature, suggesting that radical surgery of craniopharyngiomas allows good outcome in terms of survival, full recovery, and quality of life for both adults and children.

Signs and Symptoms

Visual deficits were the most common complaints on admission (75%). At ophthalmological examination, impairment of vision was found in 105 cases (86%), consisting of unilaterally or bilaterally decreased visual acuity (80%) and visual field defects (79%). Ocular motor deficiency was uncommon, occurring in only 5% of patients (paresis of cranial nerves three and six). Visual impairment was severe (visual acuity less than 1/20): unilaterally in 36 cases (30%) and bilaterally in 36 cases (30%) and bilaterally in 14 cases (11%). Seven of these 14 patients were children younger than the age of 10 years. Funduscopic examination revealed that papilledema was present in 16%, and pale optic disc was noted in 40%.

Neurological signs were mainly headache (53%), probably due to distension of sella turcica in most cases. Signs and symptoms of raised intracranial pressure were present in 20%, with drowsiness in 8%. Impairment of mentation (confusion, impaired memory, slow mentation) was present in 16 patients and complex psychiatric symptoms were noted in five. Six patients had a motor disorder on admission and four had cranial nerve impairment (cranial nerves five, seven, and 12).

Symptoms and signs of endocrinological dysfunction were present in 77 cases (63%). Nineteen patients (16%) were of short stature (13 of 29 children), 37 (30%) experienced marked asthenia, 49 (40%) sexual dysfunction, and 22 (18%) polyuria-polydipsia. The endocrinological tests were performed preoperatively in 90 cases, revealing a completely normal hormone status in only 18 patients.
The mean delay between the initial sign and diagnosis was 27 months. The duration of delay was because of the patients’ willingness to tolerate endocrinological symptoms (maximum delay 15 years after growth arrest).

Neuroimaging Studies

Neuroimaging studies included CT scanning in all but 14 patients and MR imaging in 70. Computerized tomography scanning demonstrated the presence of calcifications in 57%; all but one child had calcified tumors. The tumor was predominantly cystic in 13%, solid in 16%, and mixed in 71%. It was considered giant (frontal, temporal, or posterior fossa extension) in 13 cases (11%). Hydrocephalus was present in 25 cases (20%; 12 children and 13 adults). The diagnosis of craniopharyngioma was made preoperatively in 106 cases. In the other 16 cases, differential diagnosis with pituitary adenoma or hypothalamic glioma was not possible before the intervention.

Surgical Treatment

All patients were treated with the same goal: total tumor removal, without neurological, visual, or vital risk. The decision to perform a subtotal or partial removal was made by the surgeon during the operation, based on the anatomical findings. Because of hydrocephalus, a CSF shunt was inserted preoperatively in nine cases (7%). The approach was right frontopterional in 112 cases, transsphenoidal in 10 cases.

Conventional Radiotherapy

Radiotherapy was never prescribed as a primary treatment immediately after surgery. It was reserved for cases of partial removal (seven patients, of whom four were difficult to follow) and for recurrences (first recurrence, 16 patients; second recurrence, three patients).

Results

Surgical Findings

Anatomical localization and tumor development allowed a precise classification of the tumor, depending on its anatomical development.

Sella Turcica and Diaphragma Sellae. The tumor location was purely intrasellar in 4%; intra- and suprasellar infradiencephalic in 26%; intra- and suprasellar transdiencephalic in 7%; intra- and suprasellar supradiencephalic in 7%; and purely suprasellar in 56%. Purely suprasellar tumors were always supradiaphragmatic and purely endosellar ones were infradiaphragmatic. When growing, a tumor may compress or expand the diaphragma but usually does not invade it. Only transdiencephalic craniopharyngiomas involved both sides of the dural leaf. These tumors probably began in the sella turcica and expanded up through the diaphragma along the pituitary stalk.

Optic Chiasm. The tumor was purely prechiasmatic in 3%; purely infrachiasmatic in 10%; purely retrochiasmatic in 20%; pre- and infrachiasmatic in 6%; infra- and retrochiasmatic in 47%; pre-, infra-, and retrochiasmatic in 14%. Note that 61% of tumors were located partially behind the chiasm, and 20% were entirely behind it (that means not accessible between the optic nerves at the beginning of the surgical procedure).

Third Ventricle. Tumors were extraventricular in 92%; purely intraventricular in 3%; and partially intraventricular in 5%. Partially intraventricular craniopharyngiomas were tumors located below the floor of the third ventricle and penetrating the ventricle after invasion of the nervous parenchyma (usually invasion of the floor of the third ventricle). Purely intraventricular craniopharyngiomas were located entirely in the third ventricle, arising above the floor of the third ventricle in the region of tuber cinereum. Whether they were invading the nervous parenchyma or not, they were totally enclosed in the ventricle walls. Even purely intraventricular craniopharyngiomas were approached and removed via the frontopterional approach. Opening the lamina terminalis allowed good exposure of the anterior part of the third ventricle.

Invasiveness. The tumor was invasive (pituitary stalk, infundibulum, floor or walls of the third ventricle, optic chiasm, brainstem, and carotid adventice) in 67%. This means that a clear cutting plane was impossible to find, which necessitated the sacrifice of the parenchyma to complete the removal. Sacrifice was made only when the pituitary stalk and the infundibulum were involved. The other invaded structures were respected not sacrificed, thus resulting in incomplete removal. The removal was considered total (no tumor left under maximum optic magnification, clear cut plane with the brain, and resection of invaded parenchyma if necessary) in 72 cases (59%), subtotal (no macroscopic fragment left under maximum optic magnification, but invaded parenchyma not resected) in 35 cases (29%), partial (nervous tissue debulking but persistence of macroscopic fragments) in 15 cases (12%). To allow the maximum possible removal, the pituitary stalk or infundibulum was resected in 58 patients (48%). It was preserved in 64 (52%), when the dissection plane between the parenchyma and the tumor was adequate, or when total removal was impossible for another reason. The stalk was always preserved in transsphenoidal surgery.

Postoperative Imaging

All patients underwent CT scanning during the first 24 hours postoperatively. This examination was performed to evaluate the postoperative cerebral collapse and guide the fluid balance; however, it may also be useful to assess the quality of removal. The results of CT scanning indicated no residual tumor detectable in 103 patients (84%) and residual tumor in 19 patients (16%).

One hundred sixteen patients underwent imaging follow up between the 2nd and 4th months. At the beginning of the series, CT scanning was performed in 48 patients; however, since 1984, MR imaging has been the preferred modality (74 patients; six patients underwent both CT and MR examinations). Imaging studies demonstrated no residual tumor in 92 patients (79%), residual solid tumor in 11 patients (10%), residual cystic tumor in four patients (3.5%), residual solid and cystic tumor in five patients (4%), and residual isolated calcifications in four patients (3.5%).

Postoperative Course

The postoperative course was uncomplicated in 99
(81%) of the 122 patients, who were discharged from the hospital after a 1- to 3-week stay. The duration of stay was due to postoperative examination of endocrinological functions and equilibration of substitutive treatment. Perfect equilibration of diabetes insipidus with vasopressin sometimes required 1 to 2 weeks. Twenty-three patients (19%) experienced a medical or surgical complication. A second intervention was necessary in six patients (evacuation of an epidural hematoma, CSF shunt placement, evacuation of a subdural hematoma, two removals of infected bone, and exclusion of an opened frontal sinus responsible for a CSF leakage). Perioperative mortality was 2.5% (three patients) due to the following problems: ischemia in the area of perforating vessels after removal of a calcified tumor in a 65-year-old patient (Day 12), dehydration and cerebral collapse due to hypothalamic failure (Day 19), septicemia after epidural infection due to opening of frontal sinus (Day 38).

Long-Term Follow-Up Study

All but two surviving patients have been followed for a minimum of 2 months. One hundred ten patients have been followed for at least 1 year (mean 7.5 years). Sixty-three patients have been followed for more than 5 years, and 31 patients for more than 10 years. The maximum follow-up duration was 21 years.

The overall tumor-related mortality rate in our series is 11% (13 of 122 patients). There were three postoperative deaths; one death was attributable to rapid tumor regrowth after partial removal before radiotherapy could be administered; one death from an unexplained left parietal hematoma 4 months postoperatively; five recurrences; one radiation-induced dementia; one shunt dysfunction; and one radiation-induced tumor (ethmoid esthesioneurocytoma).

During the follow-up period, there were four unrelated deaths (colic cancer, infectious pneumonia after measles, breast cancer, and myocardial infarction).

The probability of survival was analyzed using the Kaplan–Meier algorithm and was 95% at 2 years, 91% at 5 years, 83% at 10 years. No death was observed after postoperative Year 8. The overall recurrence-free probability was 84% at 2 years, 78% at 5 years, and 60% at 10 years. Figures 1 and 2 show detailed survival curves.

Recurrence Rates

During the follow-up period, 29 patients (24%) experienced one (26 patients) or more (three patients) recurrences. The delay to recurrence was 1 to 180 months (mean 42 months, median 12 months). The risk of recurrence depends on the quality of surgical removal. Nine (13%) of 71 surviving patients in whom a total removal was achieved experienced recurrence of their tumor; 11 (33%) of 33 with subtotal removal experienced recurrence; and nine (69%) of 13 with partial removal suffered a recurrence of tumor. A new attempt to remove the tumor was made in 20 patients. These interventions were more difficult than the first ones and resulted in total removal in only five cases (25%), subtotal in eight cases (40%), and partial in six cases (30%). In one case, removal was considered impossible. One 81-year-old woman was treated conservatively by a puncture of a cyst; CSF shunt placement was necessary in three cases; and radiotherapy was administered in 16 cases (55%), alone (10 cases) or after subtotal or partial removal (six cases). Three patients died after their first tumor recurrence; two were stabilized with neurological impairment; the others were able to return to their regular activities. Three of the 26 surviving patients experienced a second recurrence, and two ultimately died of uncontrolled disease. The remaining 24 patients have been followed for 1 to 19 years (mean 6.5 years), with no other recurrences.

Functional Outcome

Visual Outcome. In the patients whose visual acuity was normal preoperatively (67 eyes of which 51 were followed 2 months or more), none was impaired postoperatively. When visual acuity was decreased preoperatively (177 eyes of which 152 were followed), the postoperative results at 2 months were improved in 70% (in which half increased to 20/20), identical in 15%, and worsened in 15%. At 1 year postoperatively, visual acuity was normal in 60% of eyes, impaired but superior to 1/20 in 27%, and inferior to 1/20 in 13%. Thirty-seven patients (34% of those patients who could be followed) had 20/20 vision bilaterally, and five (four children and one adult) had less than 1/20 vision bilaterally. All of the severely impaired patients had visual atrophy preoperatively. The improvement of vision was marked...
cranial nerve three postoperatively. All but one cranial nerve deficit resolved before patients were discharged. The last one, which was preexisting, resolved entirely after 6 months. During the frontopterional approach, brain collapse resulted in the lesioning of one (67%) or both (3%) olfactory tracts. Ten patients (9%) complained of a persistent anosmia.

**Neuropsychological Outcome.** The preoperative impairment of cognitive functions resolved in a few weeks in all but two patients. There were two new memory deficits, but both patients restored normal status in fewer than 6 months. During follow-up, four patients experienced cognitive deterioration due to multiple recurrences that were treated with surgery and radiotherapy. A change in personality was noticed in 11 patients. In three cases this change (aggressiveness, intolerance to frustration, emotional fluctuations) was associated with the surgical procedure, and in two of the three patients it resolved in a few months. The other eight patients had a long medical history, with multiple explicative factors (recurrences, new operations, and radiotherapy). All of these disorders were usually present in the same patients, so the rate of normal neuropsychological condition was 91%, as assessed by the patient and family (no memory deficit, normal speech and comprehension, normal concentration, no change in personality, character, and mood, return to his/her preillness status in terms of employment in adults, return to school no more than 2 years behind their grade level in children).

Behavioral problems were not uncommon in children, especially during adolescence. One girl, who underwent surgery three times at 11, 17, and 19 years of age, and then underwent radiotherapy, became a psychopath and drug addict. When she was finally lost to follow-up 14 years postoperatively, she was entirely unsocialized. Another young patient, who underwent surgery at age 15 years, developed anorexia postoperatively. Psychotherapy was initiated and resulted in complete cure within 2 years. The girl was able to graduate from high school and then entered medical university, with excellent academic results.

**Social Outcome and Autonomy.** One hundred (91%) of 110 patients with long-term follow-up were able to integrate successfully into society. In 9%, social integration was disturbed because of the disease. Eighty-three percent of adults achieved or returned to their premorbid occupation/profession (full-time employment, with the same level of responsibility; part-time employment was considered to be in this category only when it was also the case before the disease). In 17%, professional occupation was impaired or impossible, because of vision disturbance, multiple recurrences, or neuropsychological disorder. Thirty-one children or teenagers of school age were available for long-term follow up. School progress was normal for 23 cases (74%), with nine teenagers attending university. School performance was impaired in eight cases (26%); two children were more than 2 years behind their grade level, six required special education because of blindness (three cases) or neuropsychological impairment (three cases). Two of these children had a previous intellectual deficit, which was unrelated to their tumor (one had autism and one had an unexplained mental deficiency). Assessment of autonomy was made in a global manner by using an autonomy scale at 1, 2, 5, and 10 years. The results were maintained over time: normal life in

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**Neurological Outcome.** Preoperative motor deficiency resolved in all patients postoperatively. There were no new motor deficits. Nine patients experienced a paresis of
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85%; normal social life with light handicap in 9%; partial dependence in 5%; and total dependence in 1%.

Discussion

Indications for Surgery

Surgery of craniopharyngioma has three goals. First, it should confirm the diagnosis, which is probable but not certain even with modern imaging tools; 26 second, it should decompress the nervous structures, cure raised intracranial pressure, and improve function, especially vision; third, it should prevent recurrences. As do most authors, 8,40,50,63,82,85,92 we think that surgery is indicated in symptomatic craniopharyngioma entirely or partially situated above the sella turcica. Because of fluctuation in the size of cysts, the neurological or visual aggravation may be rapid, and surgical debulking should not be delayed after discovery of a cystic tumor. The only craniopharyngiomas suitable for medical follow up are those that are purely intrasellar. Many are therefore progressive and they will need surgery. In case of hydrocephalus, we think, as do Goel 32 and Tomita and McLone, 85 that shunt insertion should be avoided. External shunt placement predisposes to infections; internal shunt insertion is not necessary after tumor removal and poses the risk of shunt dysfunction. 93 Both make the radical surgery more difficult because the tumor is more confined in the brain. 52 Active hydrocephalus (present in 20% of our patients) is an indication for emergency surgical debulking.

Surgical Technique

We chose the frontopterional approach in 112 of 122 cases as the best approach when attempting a total resection, as did many other authors. 8,31,40,50,63,85,71,72,73 It allows for different approaches around the tumor axes: opening of sylvian fissure by using a lateral approach (useful in retrochiasmatic tumors); working in the interoptic space and in the sella by using a median approach (intrasellar and subchiasmatic tumors); and opening of lamina terminalis (intraventricular and retrochiasmatic tumors). We usually perform a rightsided approach, which is less dangerous for the patient (minor hemisphere), more comfortable for a right-handed surgeon, and suitable for each of these median tumors. The left part of the tumor is removed at the end of the procedure, without any special difficulty even in asymmetrical lesions.

We agree with Rougerie 10 and Garcia-Uria 31 that the transsphenoidal approach is not suitable for radical removal of craniopharyngioma. In contrast to adenomas that are usually soft, craniopharyngiomas are firm, calcified, and adhere to adjacent structures. Dissection to the optic pathway and the floor of the third ventricle is difficult and dangerous by using a transsphenoidal approach. The resection of diaphragma sellae is often necessary to complete tumor removal and may cause CSF leakage. Most of our patients who underwent surgery by using this approach did so because the tumor was misdiagnosed as being a pituitary adenoma. In a few cases of small, purely intrasellar or sphenoidal tumors, the transsphenoidal approach allows a good removal with excellent results in terms of visual recovery and endocrinological function. 1,4,9,31,52,54,56 In our experience, such cases represent less than 10% of all craniopharyngiomas, which are more often entirely or partially suprasellar.

Extent of Tumor Resection

The extent of tumor resection possible depends on three factors: invasion of the brain, localization of tumor, and size and consistency of the tumor. Invasion of the brain is the main limiting factor for a complete removal, because it is usually impossible to dissect the invaded parenchyma. Removal in such conditions obliges the sacrifice of the parenchyma, a decision acceptable for the pituitary stalk and the infundibulum but not for the walls of the third ventricle, optic tract, or brainstem. In agreement with Till 10 and Ammirati, et al., 5 we think that invasion cannot be predicted preoperatively, even with excellent-quality MR studies. During surgical exploration, the surgeon, on discovering the relationships between brain and tumor, must decide whether to resect a structure to complete the resection. In our 82 cases of invasive craniopharyngiomas, the resection was total in only 40 cases (49%). In these cases, invasion concerned only the stalk and infundibulum, which were removed. When the walls of the third ventricle, optic tract, or brainstem were invaded, the resection was never considered total. It was subtotal in 28 cases (34%) or partial in 14 (17%). In contrast, in noninvasive tumors, resection was total in 85%. Removal was considered subtotal in 15%, because of a doubt about the quality of dissection in a blind area.

Localization of tumor in relation to the optic chiasm is another limiting factor in removal. When the tumor is purely retrochiasmatic, it is not visible in the interoptic space. We found this factor limiting in 26% of our cases, close to the 30% in the series by Tomita. 85 The surgery of purely retrochiasmatic craniopharyngiomas (25 cases in our series) is more difficult than other types. In these cases, some authors recommend drilling the jugum. 62,81,85 This technique opens the sphenoid sinus, causing a risk of CSF leakage. Like Ammirati, et al., 5 we think it is possible to remove retrochiasmatic craniopharyngiomas with a prefixed chiasm by beginning in the interoptocarotid or laterocarotid spaces. Opening of the lamina terminalis is useful, not only in intraventricular tumors 47,48,50,55 but also in tumors that develop under the floor of the third ventricle. In these cases, the surgeon may use a cottonoid to push the tumor to the axis of vision. 10 Nevertheless, total removal is less often achieved in retrochiasmatic craniopharyngiomas: only 31% in our series, compared with 80% in other locations. This may be the consequence of a frequent invasion of nervous parenchyma in retrochiasmatic tumors (85% in our series).

In agreement with many authors, 5,8,31,40 we think the size of tumor does not interfere with removal. It is usually possible to debulk progressively the solid masses. Cysts are also frequent, helping the dissection. In our 13 giant craniopharyngiomas, removal was total in six (46%), subtotal in four (31%), and partial in three (23%).

Evaluation of the quality of removal is a very important point postoperatively. It is important for the patient, whose risk of tumor recurrence depends greatly on this factor, and for the surgeon, who wants to compare his or her work with the literature data. It has been proposed 40 that obtaining a CT scan in the early postoperative period would be the best indicator of the presence or absence of residual tumor. For many authors, 7,8,13,34,40,85 the classification of total or subtotal
removal is based entirely on the results of postoperative CT scanning. We think that this examination underestimates the microscopic tumor residues left by the surgeon in cases of nonresected invaded parenchyma. This is particularly true in the floor of the third ventricle. Even when performing postoperative MR imaging it is possible to miss such a remnant. In our series, the 3-month postoperative contrast-enhanced MR image was normal in 60 patients, but in 14 cases, the surgeon knew that he had left a microscopic remnant of the parenchyma. All removals thought to be total by the surgeon had normal postoperative CT or MR imaging; similarly all removals thought to be partial had residual tumor on the control imaging. In the subtotal group (32 patients), the MR image or CT scan was abnormal in only 10 patients, which is less than one third of the real residual tumors. We believe that the estimation of the surgeon is the best evaluation of the quality of removal. Compared with the recurrence rate, which is actually the only criterion for true total removal, the error is approximately 15% (25% on imaging-based assessment).

Surgical Mortality

The postoperative mortality in our series is 2.5%, which compares favorably with those reported in the literature.\(^{11,27,40,46,71,76,82,92}\) Lower mortality rates may be obtained in transsphenoidal surgery series, where it can drop to 1%.\(^{27,51,54,56}\) It is questionable, however, if the tumors treated using this approach (small intrasellar tumors) can be compared with large suprasellar craniopharyngiomas that invade nervous structures and are far more difficult to resect. Of note is that minimal surgery (cyst puncture, biopsy, very partial removal) does not always produce a lower rate of postoperative complications than radical surgery. In the series by Shapiro, et al.,\(^{73}\) four of 22 patients who received conservative treatment (18%) died after cyst puncture before the treatment was completed. In the same series, the postoperative mortality rate was only 8% (three of 38 patients) after radical surgery. The surgical mortality associated with these minimal procedures is underestimated in publications by authors at radiotherapy centers, where only the patients who survived the surgery are included.\(^{66,67}\)

### Tumor Recurrences

Recurrence is the major risk in the evolution of craniopharyngiomas. Table 1 gives a comparison among major literature series. A good evaluation of the risk in terms of recurrence demands a long follow-up because this is a slow-growing tumor. Five years of follow-up study should be the minimum; 10 years would be even better to reassure the patient, although most recurrences occur in the first 3 years after surgery.\(^{29,35,39,48,91}\) As a matter of fact, it is not a true recurrence but an evolution of tumor fragments, left by surgery because of microscopic invasion of nervous or vascular structures.\(^{4,16,21,74,81,87,88}\) In radical surgery, the recurrence rate depends on the quality of removal, from 15% (mean value found in literature data) in total removal to 75% in partial removal. Radiotherapy after partial removal produces a risk of recurrence close to that of total removal surgery.\(^{28,66,67}\) however, radiotherapy has its own complications (radionecrosis, dementia, and tumors), and recurrence is not constant even in partial removal.\(^{36,59,81,90}\) Mori, et al.,\(^{39}\) report on three patients who survived without recurrences after a 30-year follow up. Wen, et al.,\(^{39}\) studying 19 series, concluded that 74% of incompletely removed tumors will recur. Otherwise, radiotherapy is efficient when administered at recurrence: 16 of our patients underwent radiotherapy for a recurrence, and only one experienced a second recurrence. Because of these data, our position is to reserve radiotherapy for patients in whom a very partial removal has been obtained and for those in whom tumors recur. Our

### Table 1

Data from large surgical series reported in the literature on craniopharyngioma removal and recurrence*  

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Total Removal (%)</th>
<th>Postop Mortality (%)</th>
<th>Mean Follow Up (yrs)</th>
<th>Total Removal</th>
<th>Subtotal Removal</th>
<th>Partial Removal</th>
<th>Removal &amp; RT</th>
<th>Percentage of Recurrence</th>
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<tr>
<td>Shapiro, et al., 1979</td>
<td>60C</td>
<td>33</td>
<td>11.6</td>
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<td>23</td>
<td>78</td>
<td>78</td>
<td>38</td>
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<tr>
<td>Thomsett, et al., 1980</td>
<td>42C</td>
<td>24</td>
<td>0</td>
<td>6</td>
<td>29</td>
<td>91</td>
<td>91</td>
<td>18</td>
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<tr>
<td>Cabezudo, et al., 1981</td>
<td>45AC</td>
<td>36</td>
<td>8.8</td>
<td>?</td>
<td>25</td>
<td>63</td>
<td>6</td>
<td>6</td>
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<tr>
<td>Sung, et al., 1981</td>
<td>109AC</td>
<td>34</td>
<td>4.6</td>
<td>0-27</td>
<td>49</td>
<td>81</td>
<td>25</td>
<td></td>
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<td>43C</td>
<td>33</td>
<td>2.3</td>
<td>?</td>
<td>50</td>
<td>93</td>
<td>93</td>
<td>21</td>
<td></td>
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<td>König, et al., 1986</td>
<td>31AC</td>
<td>6</td>
<td>3.2</td>
<td>4</td>
<td>37</td>
<td>82</td>
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<td>50C</td>
<td>44</td>
<td>16.9</td>
<td>6.4</td>
<td>10</td>
<td>43</td>
<td>24</td>
<td></td>
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<tr>
<td>Weiss, et al., 1989</td>
<td>31C</td>
<td>61</td>
<td>3.2</td>
<td>5.2</td>
<td>33</td>
<td>100</td>
<td>100</td>
<td>20</td>
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<td>Yaşargil, et al., 1990</td>
<td>144AC</td>
<td>90</td>
<td>16.7</td>
<td>0-22</td>
<td>7</td>
<td>67</td>
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<td>90</td>
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<td>Symon, 1994</td>
<td>62AC</td>
<td>48</td>
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<td>Maira, et al., 1995</td>
<td>57AC</td>
<td>75</td>
<td>1.8</td>
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<td>0</td>
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<td>DeVile, et al., 1996(^{24})</td>
<td>75C</td>
<td>52</td>
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<td>48C</td>
<td>27</td>
<td>12.5</td>
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<td>53</td>
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<td>Fahlbusch, et al., 1999</td>
<td>168AC</td>
<td>49</td>
<td>1.2</td>
<td>5.4</td>
<td>11</td>
<td>51</td>
<td>69</td>
<td></td>
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<td>Duff, et al., 2000</td>
<td>121AC</td>
<td>43</td>
<td>?</td>
<td>10</td>
<td>19 (prob)</td>
<td>59</td>
<td>8 (prob)</td>
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<tr>
<td>present study</td>
<td>122AC</td>
<td>59</td>
<td>2.5</td>
<td>7.5</td>
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<td>67</td>
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* A = adults; C = children; prob = cumulative probability; RT = radiotherapy.
† Mortality rate in primary surgery = 9.8%; mortality rate in surgery for recurrence = 40.6%.
‡ Mortality rate in primary transcranial surgery = 1.1%; mortality rate in transcranial surgery for recurrence = 10.5%.
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Position concerning treatment of recurrences has changed since the beginning of the series. Considering the difficulty of a new operation in recurrences and its morbidity and mortality rates, we think at the present time that radiotherapy is the elective treatment at this stage, with a good result in the majority of cases and fewer risks than radical surgery. Thanks to close radiological follow up, made possible by modern imaging tools, most recurrences can be discovered when the tumor volume is still small, which does not necessitate new surgical debulking. Radiosurgery may be a promising treatment but needs further evaluation.

Functional Outcome

Quality of life is a main concern in patients treated for a benign tumor such as craniopharyngioma. Duff, et al., propose that good outcome should be evaluated by eight criteria: 1) still alive at the follow-up examination; 2) no major motor deficit related to treatment or tumor progression; 3) functional vision; 4) a Katz grade of A; 5) a Karnofsky Performance Scale score of at least 80; 6) school status of no more than 1 year behind the expected grade for children and young adults; 7) employability for adults of working age; and 8) absence of debilitating psychological or emotional problems. Evaluated with these criteria, 60.3% of their series of 121 surgically treated patients achieved a good outcome. We assessed the outcome in our series with the global criterion of dependence or independence in social and professional life in adults and school status in children. Our study demonstrates a high rate of independent living with social integration and normal professional occupation (84% in adults, 74% in children); most patients returned to their preillness status (employment at same level of responsibility in adults, no more than 2 years behind their grade level at school). These good results are associated with low rates of ophthalmological and neuropsychological sequelae. Ophthalmological condition is an important limitation of autonomy in the literature. Reviewing the available series, it seems that this point has not received the attention it deserves. The authors who focused on it, 2,12,29,33,56,60,65,68 agreed on four conclusions: 1) the probability of recuperation depends on the severity of preoperative impairment; optic atrophy (seen at fundoscopy) carries the worst prognosis; 2) children are often more affected than adults, but at the same impairment level, the prognosis after optic decompression is the same at every age; 3) surgery offers better results than radiotherapy; and 4) recurrences are often responsible for definitive visual aggravation. In conclusion, ophthalmological impairment is a frequent after effect in patients with craniopharyngiomas. The handicap is severe in 10 to 30% of cases documented in the literature. Our series demonstrates that with close attention to this problem, these results can be improved to less than 5% permanent blindness.

Anterior pituitary failure is common after treatment of a craniopharyngioma. It is constant and complete when the pituitary stalk has been divided for tumor removal. In 50% of cases, the stalk is not invaded by the tumor, and it is possible, by close dissection, to preserve it anatomically. This does not translate into intact endocrinological functions postoperatively in every case but we think, like Honegger, et al., and Lapras, et al., that it is worth preserving the pituitary stalk when possible. A partial endocrinological deficit allows better quality of life than complete hypopituitarism, and some patients experience even complete preservation of all hormone functions. This was the case in 20 of our patients, 16 in whom there was total tumor removal. These patients have the best results in the series, enjoying a normal life without any substitutive therapy. Interestingly, they all have excellent neuropsychological outcome and absence of weight gain, all facts supporting the absence of hypothalamic injury.15 This result was achieved by using a transspheonoidal approach in two cases and, which is remarkable, via a transcranial approach in 14 cases of large, supradiaphragmatic tumors. It demonstrates that, in selected cases of suprasellar craniopharyngiomas, endocrinological prognosis may be better than what is usually reported. If any doubt remains on the quality of dissection between the stalk and the tumor, we prefer to resect the parenchyma to complete the removal—hypopituitarism being far less serious than recurrence.

The neuropsychological prognosis of patients with craniopharyngioma is diversely assessed in literature. Authors of older publications have reported severe psychological disorders in patients after treatment, especially in children.17,19,20,30,40,45,57,83 These patients were described as immature, impulsive, and intolerant to frustration, with frequent impairment of cognitive functions. Social and professional integration were compromised. The role of radiotherapy compared with surgery in such dysfunctions has been a great subject of debate.8,28,29,35,63,67,69,79,83 In recent series, 26,27,29,71,82,85,91 it seems that intellectual and psychological results have been improved. In a prospective study of 13 adults, all but one of whom underwent surgery, Honegger, et al., demonstrated that neuropsychological performances and quality of life are not impaired. Donnet, et al., came to the same conclusion in a study of 22 patients: all but four of them were able to return to their preillness activities. Abrams and Repka reported that these results can be achieved in children. Our results support these conclusions; most of our patients were able to return to normal active life after treatment. In children, the question of social prognosis remains; some authors have described severe impairment in school performance and later autonomy. More than neurological deficits, these patients commonly experience behavioral disorders associated with short stature, obesity, headache, and emotional and sexual disturbances, which may cause a genuine handicap.

Conclusions

We recommend a frontopterional approach to surgery in all symptomatic craniopharyngiomas in adults and children. Radiotherapy should not be administered postoperatively unless the removal of tumor is impossible, which is unusual. Recurrences are a problem in 25% of patients, but they can be treated with radiotherapy, achieving a good result in most cases. Using this protocol, the operative mortality rate is low, the functional results are good, and the probability of long-term patient survival is high.

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