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Cranial Surgery for Adult Craniopharyngiomas: Techniques and Indications

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Abbreviations

AC V	Anterior choroidal vein
ACA1	First segment of the cerebral artery
AChA	Anterior choroidal artery
ACoA	Anterior communicating artery
AO-CP	Adult onset craniopharyngiomas
CCPs	Cystic craniopharyngiomas
Ch	Chiasm
ChP	Choroid plexus
FL	Frontal lobe
GTR	Gross total removal
hS V	Thalamostriate vein
HW	Hypothalamic walls
IC V	Internal cerebral vein
ICA	Internal carotid
III V	Third ventricle

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E. Jouanneau, G. Raverot (eds.), *Adult Craniopharyngiomas*, https://doi.org/10.1007/978-3-030-41176-3_6

LT	Lamina terminalis
MBA	Mammillary body angle
MF	Foramen of Monro
ON	Optic nerve
PG	Pituitary gland
PS	Pituitary stalk
RXT	Radiotherapy
SF	Sylvian fissure
STR	Subtotal removal
TC	Trans-callosal approach
TF	Trans-frontal approach
Th	Thalamus
TL	Temporal lobe
TLT	Trans-lamina terminalis approach

6.1 Indications and Principles of the Surgery

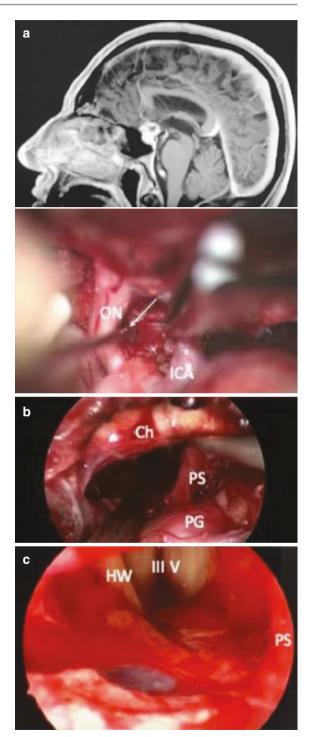
Surgical excision remains the accepted first-line treatment for the large majority of AO-CP and should be systematically considered in the absence of major co-morbidities.

However, the optimal surgical modalities used for AO-CPs remain uncertain. The extension of resection remains controversial and it is still unclear if GTR leads to a better prognosis than STR associated with RXT in AO-CPs even though the rates of recurrence are clearly lower for the first of these options [1]. AO-CPs cohorts are too small for a definitive conclusion to be drawn. Pituitary stalk conservation, whenever possible, can be recommended but not at the expense of a GTR. Indeed, hormonal substitution is quite easy to manage (see chapter by G Raverot in this book). The challenge is at the level of the infundibulum and the hypothalamus. Recent data has shown that the hypothalamic part of the tumor can be removed whenever possible with an acceptable morbidity [2]. However, the authors would not recommend a GTR for all cases and a universal approach. Surgeons need to study all pre-operative parameters, especially the MRI results (outlined in the chapter by R Prieto). Preoperative hypothalamic syndrome indicates a lesion from the tumor but does not mean that a pseudo-gliosis cleavage plane cannot be found. It is therefore of paramount importance to have the best vision of such a plane.

The surgical approach is therefore designed to expose the tumor, in the best possible way, with minimal morbidity.

Endoscopic transnasal techniques have benefited from an impressive development over the last two decades [3–5] and have become the best option, especially in AO-CP. Indeed, sellar-suprasellar spaces are in the axis of the endonasal route and the endoscopic approach offers a great benefit for controlling the retrochiasmatic part of the tumor and the third ventricle floor exposure (Fig. 6.1) [2, 6]. Moreover, a posterior extension toward the posterior perforating space or behind the clivus can be managed through an extended trans-clival approach [7].

Fig. 6.1 Better exposure of the third ventricle floor using the extended endonasal endoscopic technique. (a) Operative view of a suprasellar craniopharyngioma operated on using a sub-fronto-pterional approach. The sub- and retrochiasmatic spaces are difficult or impossible to expose without pushing on the optic nerve. The surgeon is gently pushing away the optic nerve to work in between the internal carotid and the optic nerve (arrow). The third ventricle floor, behind and below the chiasm, cannot be directly exposed and the tumor will be removed by a piece-meal technique without being able to see the hypothalamus floor. (**b**, **c**) Operative views of the same type of craniopharyngioma. Using an extended transtubercular route, the surgeon has good exposure of third ventricle floor after tumoral debulking when approaching the endoscope. The dissection, under direct visualization, can be softer and safer to preserve, as much as possible, the hypothalamus while doing radical removal of the tumor. ON optic nerve, ICA internal carotid artery, Ch Chiasm, PS pituitary stalk, PG pituitary gland, HW hypothalamic walls, III V third ventricle



6.2 Trans-Cranial Approaches

Cranial routes are therefore less frequently used in adults. This approach is considered when the tumor extends towards the anterior skull base or the Sylvian fissure (Fig. 6.2) [6].

When using pterional subfrontal approaches, microsurgical techniques must be used without any specificity for the cisternal part of the tumor: debulking, and after extracapsular dissection, working through the usual inter-optic, inter-opticocarotid, and retro-carotid spaces, and paying close attention to the small perforating arteries [8]. The surgical steps are described in Fig. 6.3.

The specificity comes from the management of the ventricular extension of the tumor, where this is present, especially in case of a pre-fixed chiasma. Conversely to a nasal approach (see Fig. 6.1), the exposure of the third ventricle floor from a transcranial approach is far more complicated and can lead to unsatisfactory control of the intraventricular portion of the tumor (and thus to an incomplete removal).

A trans-lamina terminalis route can be used in two ways: the first is to use a cottonoid to push down the third ventricle and the tumor and thus expose it in the infrachiasmatic space; the second is to actually remove the tumor through the small window. More detail of this approach is given below.

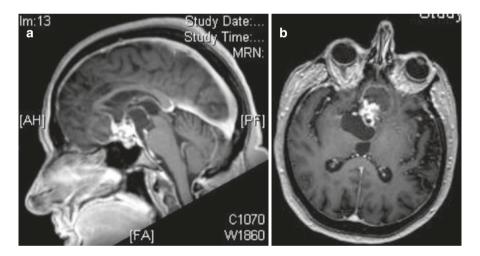


Fig. 6.2 A typical indication for cranial surgery in an AO-CP because of multiples cysts that extend anteriorly and laterally

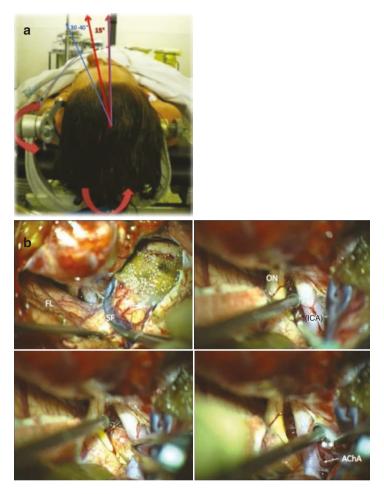


Fig. 6.3 Surgical steps for a pterional sub-frontal approach. (a) The patient is supine, the head slightly extended and turned, depending on the tumor extension. To work in the inter-optic space, 15° lateral tilt is required but for retro-carotid space, the head should be tilted around $30-40^{\circ}$. (b) The suprasellar cistern and Sylvian fissure are opened up largely for CSF drainage and to expose all of the crucial landmarks, the optic Nerve (ON), internal carotid (ICA), and the inter-optico-carotid space (*). Working behind the ICA is dangerous because of the perforating arteries (anterior choroidian artery, AChA, and other small hypothalamic vessels coming from the posterior communicating artery).

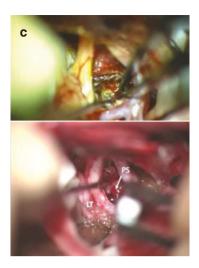


Fig. 6.3 (continued) (c) The surgical technique is similar to that used for other extra-cerebral tumors, piece-meal first followed by dissection. The particularity in this case is the hypothalamus dissection. The lamina terminalis (LT) can be opened up to expose the ventricular part of the tumor or left alone in case of a pure intraventricular CP. In that case, after an intratumoral debulking, the pituitary stalk (PS) has been located and kept intact. Unfortunately, the patient's condition worsened, with a pan-hypopituitarism post-surgery. *FL* frontal lobe, *TL* temporal lobe, *SF* sylvian fissure, *LT* lamina terminalis, *ICA* internal carotid, *AChA* anterior choroidal artery, *PS* pituitary stalk

6.3 Specific Situations

6.3.1 Combined Approaches for Giant CPs

Combined approaches form part of the history of surgery for CPs. From a sub-frontal route, in the 1930s, Dott proposed to intentionally cut and sacrifice the blind optic nerve to control the retrochiasmatic extension, with a second surgical step using a trans-frontal trans-ventricular approach for the ventricular extension (see the chapter on Epidemiology in this book [9]). Using this ventricular approach, he was the first to recognize the role of the hypothalamus, observing that in the case of a sharp dissection, the patient's state worsened with an increased hypothalamic syndrome.

In our era, combined approaches can be considered for management of huge CPs. The transcranial route allows the management of anterior and lateral extensions and the transnasal route allows the management of the retrochiasmatic and ventricular extensions. In these cases of giant tumors, achieving a GTR without major neurological damage is illusory. Thus, the goal of this strategy must be to achieve STR combined with RXT. The goal is to decompress the brain and optic pathway, reducing the size of the tumor so that it can be efficiently treated by RXT. The optimal sequence—transcranial approach followed by transnasal versus transnasal followed by transcranial—is dictated as for giant pituitary adenomas by the question: which route is best for decompression of the optic apparatus? This

strategy is easier to use in CPs. Indeed, contrary to giant pituitary adenomas, the risk of apoplexia of the residual tumoral portion that is left is far lower in CPs.

This strategy was used in the case illustrated in Fig. 6.7. This patient was referred to our center after sub-frontal surgery where a simple biopsy was done. The patient was almost blind and had total pituitary insufficiency. Surprisingly, no hypothalamic syndrome was present. The tumor was then approached using a trans-frontal transventricular approach to remove the upper part. We left in place a piece of tumor that was too strongly adhered to the optic chiasm and the ventricle. The proposal was thereafter for upfront radiotherapy. Unfortunately, the patient postponed treatment and returned 9 months later with a recurrence, which was treated using a nasal approach. The small piece of recurrence after 4 years. The clinical status of the patient was unchanged with an optic atrophy, as was observed pre-operatively, and with no hypothalamic syndrome.

Lastly, considering the issue of CSF leak when two surgical steps (nasal and cranial) are scheduled, the two approaches should be performed as separate surgeries.

6.3.2 Management of Intraventricular Tumor

It is sometimes very difficult to determine if the CP is located inside the 3rd ventricle or if it is a suprasellar tumor that has secondarily extended to the ventricle. This is a key point in selecting the best approach. Using the wrong route, in particular reaching the tumor through the third ventricle floor by a transnasal approach may lead to dramatic damage to the hypothalamus.

In our experience, MRI T2 sequences, especially the mid-sagittal view, are the best option to determine this anatomical situation. The position of the mammillary bodies and the mammillary body angle (MBA) must be carefully studied. An MBA inferior to 50° indicates an intraventricular CP (Fig. 6.4). This rule is, however, not absolute as can be seen in Fig. 6.5, where despite an MA inferior to 50°, the tumor appeared to pass through the third ventricle, therefore, being accessible using an endonasal approach. Readers can also refer to the chapter by Prieto in this book for more details.

The 3rd ventricle can be accessed by a number of routes: a trans-lamina terminalis approach (TLT), a trans-frontal approach (TF), or a trans-callosal (TC) approach.

The choice should be made by considering the following parameters:

- the size of the tumor and its height: small- or medium-sized tumors that do not extend too greatly superiorly or inferiorly are good candidates for a TLT approach;
- The presence of hydrocephalus: For a large tumor with ventricle dilatation, a TF route may be adequate; while for a large tumor without ventricle dilatation, a TC route may be more appropriate including its variants (inter-forniceal; inter thalamo-trigonal);
- The surgeon's own previous experience.

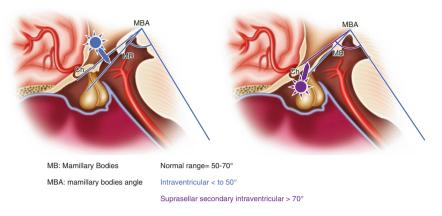


Fig. 6.4 Mamillary bodies angle (MBA) for intra- or extra-ventricular CPs (modified from R Prieto)

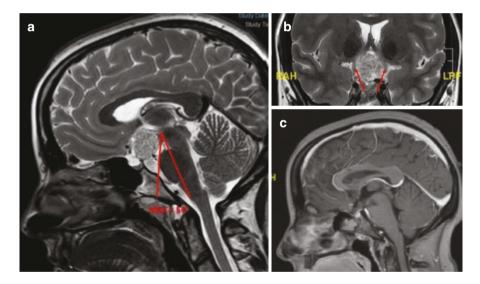


Fig. 6.5 One example of CP with an MA < 50° that was, however, approached using an endonasal route. The CP passing through the 3rd ventricle floor can be approached from below with few additional morbidity than the tumor already creates. (a) MA < 50° suggesting an intraventricular CP and a transventricular approach. (b) In the coronal view the tumor passes through the ventricle floor to point into the suprasellar space (ventricle walls with the optic pathway on both sides [arrowed]). (c) An endonasal endoscopic extended approach has been employed therefore to completely remove the tumor. The visual outcome was good, but the patient's endocrine state worsened, which was expected as the tumor originated from the infundibulum. Pre-operatively, the patient was overweight, and this increased slightly after surgery and was controlled by diet and exercise

6.3.2.1 Trans-Lamina Terminalis Route (Fig. 6.6)

Considering the role of the extended endonasal approach, the trans-lamina terminalis window may be considered useful in two different situations:

- As a complement during a sub-frontal approach to reach the retrochiasmatic or intraventricular part of a CPs
- For a pure intraventricular CPs, as in the case presented in Fig. 6.6

This technique, which has been used for many years, is outlined in numerous publications [10–14].

For a pure TLT approach, an interhemispheric approach may be preferable to a pterional sub-frontal in order to obtain a perfect exposure of the ventricle walls on both sides. Though, as shown in Fig. 6.6, with a sub-frontal approach, the hypothalamic wall on the same side as the approach can be difficult to expose. A trans-sinus approach (or a midline bi-frontal craniotomy in case of small sinuses) can therefore be performed with the patient placed supine, the head neutral and strictly midline in a three-pin head holder.

Once the lamina terminalis has been exposed, it can be opened up to directly access the tumor. This strategy is therefore a piece-meal technique and as usual, the critical point is the dissection of the tumor from the hypothalamic walls where the

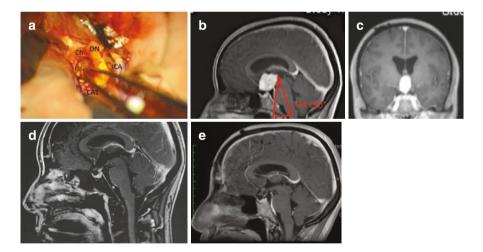


Fig. 6.6 A case of a pure intraventricular CPs removed through a pterional sub-frontal TLT approach. (a) Operative view showing a sub-frontal TLT approach. The ipsilateral hypothalamic wall may be difficult to expose without pushing on the optic nerve. (b, c) The mammillary body angle (MBA) is inferior to 50° indicating an intraventricular tumor. (d) Post-operative MRI at 3 months. The result was classified as a GTR without endocrine and visual symptoms. (e) At 6 months, a small cystic recurrence at the level of the pituitary stalk appeared, which was treated by conformational radiotherapy. The patient is disease free at 4 years. *ON* optic nerve, *ICA* internal carotid, *Ch* chiasm, *CA1* anterior cerebral artery first segment

surgeon must decide between a pseudo-gliosis plan, where a GTR can be attempted, and an invasion where a STR would be advised. The same formula can be used whichever approach is used: when the tumor can be easily and gently mobilized (soft dissection), it can be removed. In other situations, in case of a sharp dissection, the tumor should be kept in place.

Pros:

- Direct ventricle vision is obtained without crossing any neurological structures, allowing tumor removal without causing endocrine deficit, as in the case presented in Fig. 6.6 and as has been reported in the literature [14]
- Such an approach is independent of the length of the optic nerves

Cons:

- Provides a narrow window, especially in case of short ACA1 or fenestrated ACoA, and sometimes difficult to mobilize. Splicing the ACoA can be used when the artery length is sufficient with, however, the sacrifice of an anastomosis that may be useful in where there are stroke issues with age
- Vascular risks for the ACoA and the perforating arteries of the anterior complex
- Limited exposure of the upper part and the third ventricle floor that can lead to STR, as illustrated in the Fig. 6.6. An endoscopic-assisted technique can be used, however, this requires caution as the window is quite narrow and there can be issues with warming if approaching too close to the endoscope tip.

6.3.2.2 Trans-Frontal Trans-ventricular Route (Fig. 6.7)

When the tumor is located in the 3rd ventricle and results in an hydrocephalus, the surgeon can choose a transcortical trans-frontal approach.

The craniotomy is performed one-third behind the coronal suture, two-thirds in front, without crossing the midline. A 3 cm antero-posterior cortical incision is made and the frontal horn of the ventricle reached at a depth of about 4 cm. Usually, the tumor is approached through the dilated Monro foramen which can be enlarged posteriorly using a interthalamic forniceal approach through the choroidal fissure and the superior tela after coagulation and section of the septal vein. Great care should be taken for the fornix and of the other veins.

Pros:

 An easy approach with less complicated structures involved than the transcallosal approach

Cons

- Only possible when there is hydrocephalus
- Transcortical approach can result in sequelae seizure

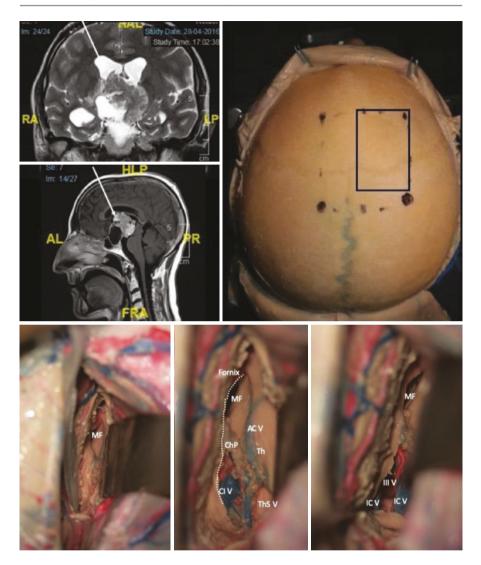


Fig. 6.7 A case of a huge CP which was first operated in another center using a sub-frontal approach without satisfactory removal. A second step using a trans-frontal approach was possible to performed because of the hydrocephalus. After a linear incision, a frontal craniotomy is carried out one-third behind the coronal suture—two-thirds in front. *Cadaveric study*: The foramen of Monro (MF) is exposed after entering the frontal horn. Tumor exposure can be improved using a transchoroidal approach, separating the fornix (dotted line) and the thalamus medially to the choroid plexus (ChP) to get inside the third ventricle. *MF* foramen of Monro, *AC V* anterior choroidal vein, *ThS V* thalamostriate vein, *Th* thalamus, *ChP* choroid plexus, *IC V* internal cerebral vein, *III V* third ventricle. *The different surgical steps for the patient*:

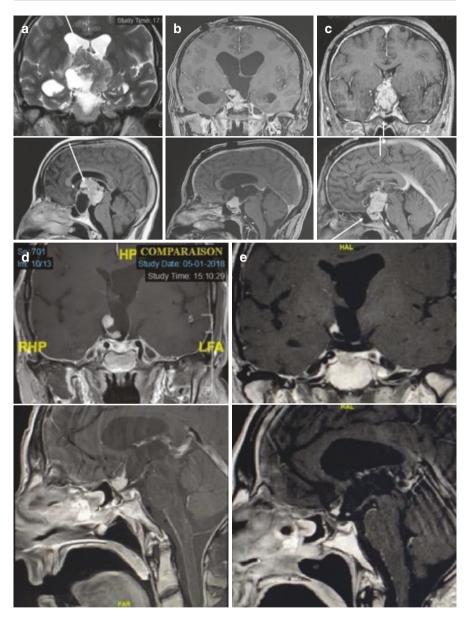


Fig. 6.7 (continued) (**a**) Pre-operative MRI; (**b**) After the trans-frontal approach with a deep tumor remnant at the level of the ventricle floor. (**c**) Radiotherapy was delayed due to the patient and 9 months later a recurrence in the ventricle was observed. The patient was operated on using an endonasal approach. (**d**) Post-operative result with a piece of tumor left in place because of strong adhesions. Radiotherapy was performed at 4 months. (**e**) At last follow-up, after 3 years, there was still perfect control of the tumor

- Not a midline approach
- Deepest part of the tumor is difficult to expose at the level of the posterior perforating space

6.3.2.3 Trans-Callosal Route (Fig. 6.8)

In the absence of hydrocephalus (or when a shunt has been placed before), an interhemispheric trans-callosal approach can be used, including its three variants: interforniceal (between the fornix bodies), and the inter thalamo-trigonal approaches with its trans- and subchoroidal variants (opening the choroidal fissure on the fornix and on the thalamic side, respectively) [15, 16].

The patient positioning is the same as that used for a transcortical trans-frontal approach, with a craniotomy crossing the midline in order to achieve an interhemispheric approach.

The dura mater is opened in a U shape and great care is taken of the afferent veins to the sagittal longitudinal sinus.

Once the corpus callosum is reached, a short 2 cm antero-posterior incision is made in between the pericallosal arteries.

A strictly midline approach can also be used, separating the fornix bodies to reach the third ventricle through the superior tela in between the internal cerebral veins.

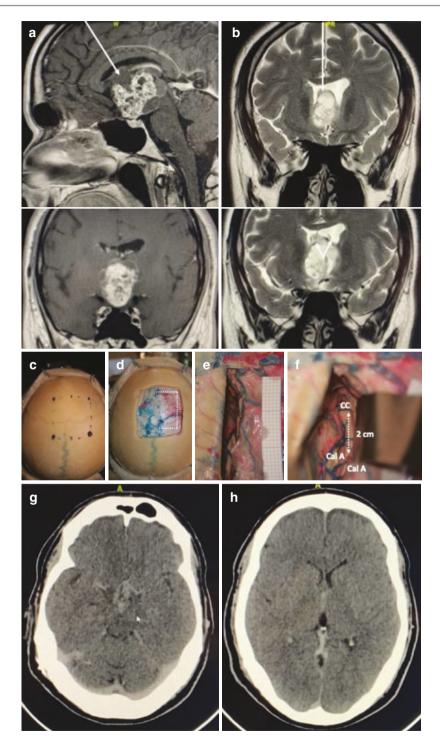
Another option is to enter the frontal horn of the lateral ventricle. The third ventricle is therefore reached using the same approach as using the trans-frontal one, using a transchoroidal or a subchoroidal variant. The difference between the two latter approaches is minor, with perhaps a more direct and larger view obtained with the transchoroidal approach. To increase the antero-posterior exposure, the septal vein can be cut at the level of the Monro foramen. From that point, exactly the same principles as with the trans-frontal approach are followed.

Pros:

- Avoids the transcortical route and any brain lesion

Cons:

- Difficult management of veins
- Involves manipulation of the Fornix with possible sequelae memory impairments
- The deepest part of the tumor is difficult to expose at the level of the posterior perforating space



6.3.3 Management of Cystic Craniopharyngiomas (CCPs)

There is a considerable number of publications regarding pediatric-onset Cystic Craniopharyngiomas (cCP). In huge cCP in children, where total excision may be impossible and when radiotherapy may have to be postponed for developmental reasons, drainage and local therapies may be the best options. Intracystic therapies are based on the inflammatory theory of the tumoral walls at the origin of the cyst, recently validated by molecular studies (see chapter by Martinez-Barbera in this book [17]).

To date, Bleomycin and IFN- α have been the most tried therapies, either at initial diagnosis or after recurrence.

The first step is to insert a catheter inside the cyst, connected to a sub-cutaneous Ommaya reservoir. A few days after confirming that the system is "watertight" (especially for Bleomycin), the treatment can be started.

Bleomycin has been generally used three times per week with a median dose of 0.43 mg/kg/week for a total dose of between 8 and 75 mg (mean 36 mg). The treatment duration is several weeks [18].

For IFN- α , 3 MIU/day is injected inside the cyst, after withdrawing as much of the liquid as possible, on occasion up to a total of 36 MIU over several courses in cases where the cyst did not respond well [19].

Bleomycin treatment appeared to be effective (with close to 100% of cyst response and a shrinkage superior to 90% in one quarter of cases), but the severe side effects observed have led to a loss of interest in Bleomycin. In addition, the shrinkage was also found to be transitory: less than 1 year for half of the population and with sustained benefit seen in the other half of the population, with a mean follow-up of 34 months [20].

Since the pioneering work of Cavalheiro in 2005, IFN- α has proven to be effective with less side effects than Bleomycin. A recent multicenter assessment concluded that IFN- α therapy was useful in delaying more invasive treatment in children [21, 22].

For adult onset cCP, data are scarce due to pure or predominantly cystic CP being uncommon. We recently reviewed our own data (11 cases) focusing on a single point: is drainage sufficient to manage cystic CP? Interestingly, with a mean followup of 40 months, only 27% of patients had recurrence, 2 in a cystic manner, one

Fig. 6.8 Trans-callosal approach. Huge intraventricular CPs without hydrocephalus. A transcallosal approach was chosen. (**a**) A trans-callosal approach was chosen to remove this intraventricular CP, revealed by an ICP, with hydrocephalus. The patient has been firstly operated on to insert a shunt and was then referred to our center. (**b**) The variant used here was an inter-forniceal route because the CP had itself created the route. On the T2 coronal view, we can clearly see a cyst pointing in between the two fornix (arrows) and the internal cerebral veins. (**c**–**f**) Anatomical studies: the different steps for the interhemispheric trans-callosal approach are shown. A craniotomy crossing the midline is performed to prepare the interhemispheric approach (**c**). A U-shaped opening is made in the Dura-mater (**d**). A 2 cm incision of the corpus callosum (CC) is made between the peri-callosal arteries (Cal A) (**f**). (**g** and **h**) Post-operative contrast CT showing a sub-total removal. The inferior infiltration at the level of the third ventricle floor has been left in place. The outcome was uneventful with a total recovery of the patient's mnesic issues, without any endocrine or visual deficits

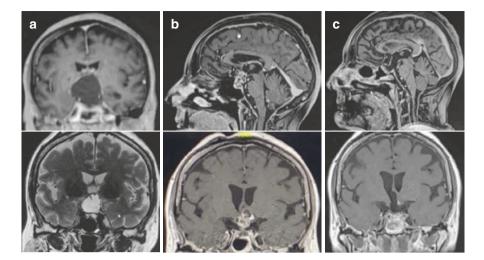


Fig. 6.9 Unpredictable evolution of a cystic CP after drainage. (a) Cystic CP diagnosed in a 68-year-old woman with pseudo-dementia syndrome which was completely resolved after drainage. (b) The cyst disappeared after 1 year and the solid part of the tumor started growing 3 years later. An endonasal approach has been performed. (c) Post-operative MRI after a STR via the nose

from a solid part of the tumor (Fig. 6.9). Only one patient required secondary drainage (using the Ommaya) and one patient in whom the catheter had been removed due to infection showed no recurrence after 5 years (Fig. 6.10). The high rate of tumor control produced by a simple drainage has been confirmed by other authors, as well as the low rate of repeated aspiration [23]. This observation can be explained by the continuous drainage of the cyst following the insertion of the catheter in the ventricle or CSF spaces. Thus, some authors advocate a large opening of the cyst in the ventricle, rather than a simple insertion of a catheter using the endoscopic technique [24, 25].

The role of IFN- α instillation in adult CCP remains unclear, and perhaps it may preferably be used in recurrence.

There is also very little data available on immediate post-drainage management, meaning a wait and see approach or upfront radiotherapy both need to be considered.

Considering the low recurrence rate after a simple drainage the unpredictable evolution of the tumor (recurrence may arise from the solid part rather than the cystic part of the tumor (Fig. 6.9) or the cyst disappearing after a single drainage (Fig. 6.10)), the possible enlargement of the cyst after radiotherapy, and similar recurrence after upfront radiotherapy versus a wait and see approach, palliative management of cCP only by drainage, may represent an interesting option ([23, 24], personal data). This allows not only good tumor control but also good clinical outcomes, including preservation of pituitary function, and preservation of good visual and cognitive function.

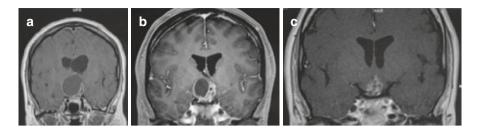


Fig. 6.10 One example of spontaneous regression of a cyst after a single drainage. (a) At diagnosis a predominant cystic CP was observed. A catheter was inserted using a stereotactic technique which was then removed at 3 months because of an infection. (b) The cyst remained stable at 1 year and the patient was free of symptoms. (c) Two years later, the cyst disappeared and the solid tumoral part remains stable 4 years after the initial diagnosis

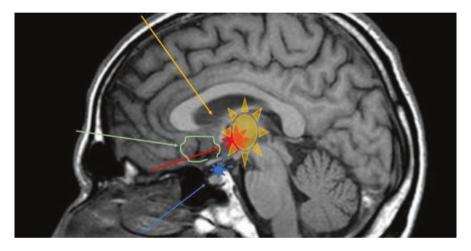


Fig. 6.11 Surgical choices according to CP anatomy. — Sellar-suprasellar, retrochiasmatic CPs: nasal route; — Anterior and lateral extensions: cranial route; — Intraventricular small CPS: trans-lamina terminalis route; — Intraventricular huge CPS: trans-frontal (if hydrocephalus present) or trans-callosal (without hydrocephalus) route

6.3.4 Key Points for Beginners: Fig. 6.11

- There are two routes to operate AO-CPs: the nasal route through the skull base and the historical cranial approach.
- There is no randomized study comparing the outcome of both routes, but all experts currently consider that the third ventricle floor is better exposed in the endoscopic endonasal approach with an expected improvement of the GTR.
- Therefore, AO-CPs which are mainly sellar-suprasellar tumors are operated on using the endoscopic endonasal techniques despite the issue of a higher rate of CSF leak not being completely solved.

Surgical appr	roach	Indications	Pros	Cons
Transnasal		Sellar-Suprasellar- retrochiasmatic- retroclival tumors	 Direct access to the tumor Better exposure of the hypothalamus and pituitary stalk Better GTR rate Avoids optic nerve and chiasm mobilization 	 Cannot control lateral extensions Not adequate for pure intraventricular tumor CSF Leaks Meningitis Rhino-sinusitis
Transcranial		Tumors with anterior and lateral extensions	Less issue of CSF leaks	 Worse exposure of the ventricle, the third ventricle floor and retrochiasmatic space Mobilization of nerves and vessels Issue of seizures Worse visual outcome
Combined nasal and cranial		Huge tumors with intraventricular extensions		
Specific 3rd ventricle approaches	TLT	Medium- and small-sized intraventricular tumors	Easy access without nerve mobilization	 Narrow window Vascular risks Poor exposure of the third ventricle floor
	TF	Intraventricular tumor with hydrocephalus		 Vein and fornix risks Seizure issues Poor exposure of the deepest part of the tumo
	TC	Intraventricular tumor without hydrocephalus		 Vein and fornix risks Poor exposure of the deepest part of the tumo

Table 6.1 Respective indications and drawbacks

TC trans-callosal, TF trans-frontal approach, TLT trans-lamina terminalis approach

- Rare indications for cranial approaches still remain for AO-CPs, in particular when the CPs has developed away from the midline with lateral or anterior extension(s). When the tumors are located inside the third ventricle, the surgeon has three choices: reaching the tumor through the *lamina terminalis* just above the optic chiasm (small or middle size tumor), through the frontal cortex (possible when there is hydrocephalus), or through the corpus callosum (in the absence of a ventricle dilatation) (Fig. 6.11).
- Evolution of Cystic CPs is unpredictable and minimally invasive management with a simple drainage through a catheter and then a wait and see policy may be advisable. Around two-thirds of cystic CPs are controlled by a simple drainage after more than 3 years of follow-up. Continuous drainage of the cyst in the ventricle may contribute to better control. Thus, an endoscopic fenestration should be preferred rather than a simple catheter stereotactic insertion.

- The role of intracystic therapies, mainly using IFN- α , remains unclear for AO-CPs.
- The surgical treatment of these rare tumors is difficult and impacts directly on the outcomes, in particular the management of the ventricular portion of the tumor, requiring highly specific skills. Thus, these patients should preferably be managed in dedicated referral centers (Table 6.1).

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