Draining Techniques for Cystic Craniopharyngiomas

9

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9.1 Introduction

Microsurgical total removal still remains the gold standard in the treatment of primary craniopharyngiomas with the best long-term prognosis. Excellent results have been achieved in particularly skilled and experienced hands [1, 2]. However, the intimate relationship with delicate neurovascular structures, the frequent absence of clear-cut cleavage, and the biologically benign nature of the tumor often suggest a less aggressive attitude [3]. This is particularly true in recurrent cases and in very young or very old patients, who are more prone to devastating intra- and postoperative complications. Only 10 % of craniopharyngiomas are totally solid, while more than half are purely or predominantly cystic [1, 4, 5]. In such cases, control of mass effect, often caused by enlargement of the cystic component, may represent a suitable alternative to resection and several techniques have been proposed [4–10]. In this chapter we are going to describe two techniques for the treatment of cystic craniopharyngiomas:

1. Cysto-ventriculo-cisternostomy with neuroendoscopic approach: drainage and wide marsupialization into CSF spaces (cysto-ventriculo-cisternostomy) are achieved to ensure mass effect control, continuous dilution, and reabsorption of the cyst's fluid. We can see that this is a more modern sump drainage [11, 12] in respect to the one performed in the past where the catheter was linked to a reservoir permanently lodged in the subgaleal space.
2. Cystosphenoidostomy (either by microsurgical or endoscopic approach): this technique allows to create a permanent communication between the cyst of the tumor and the sphenoid sinus through a silastic catheter, in order to prevent growth of the cyst due to liquid accumulation and mass effect. The catheter is easily placed through a standard transsphenoidal approach [4, 13, 14].

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9.2 Technique of the Cysto-Ventriculo-Cisternostomy

Between December 1996 and December 2010, 15 patients (6 males, 9 females; age range 9–73 years; average 52 years) harboring predominantly cystic craniopharyngiomas underwent neuroendoscopic exploration at our institution. The seven recurrent tumors had previously undergone: microsurgery and sump drainage (1), microsurgery and fractionated radiation therapy (3), and microsurgery alone (3). All tumors had close relationships with the ventricular system and could be classified as type c (9), type d (2), and type f (4) according to Yasargil et al. [2]. In 12 cases the cyst represented >90 % of total neoplasm's volume, while in the remaining patient, it was approximately 60–70 %. Indications for neuroendoscopy can be summarized as follows: age and poor general conditions (6 patients), recurrent tumors (7 patients), and refusal of open surgery (2 patients). All procedures were carried out by means of a 4-mm fiberoptic steerable, flexible endoscope (Codman & Shurtleff) with a single working channel. A freehand technique was used: surgical maneuvers were carried out by means of monopolar cautery, Fogarty catheter (3 French), alligator, and cup microforceps. Draining of the cyst and creation of a cysto-ventriculo-cisternostomy were obtained (patient in the supine position) as follows (Video 9.1):

1. Standard pre-coronal parasagittal burr hole and ventricular puncture by means of a "peel-away" trocar. This catheter is useful for protecting the cortex around the endoscopic tract during endoscopic procedures.
2. Identification and puncture of the cyst's dome and complete drainage of content by aspiration and washing with Ringer's solution (Fig. 9.1a).
3. Cavity exploration, biopsy, coagulation, and resection of the cyst's dome as extensively as possible.

The last steps of the procedure are perforation of the cyst's fundus into the basal cisterns (Fig. 9.1b) and positioning of the transcystic multiholed catheter with ends reaching into the cistern (caudally) and lateral ventricle (rostrally) (Fig. 9.1c). This stenting was devised in an attempt to promote continuous CSF rinsing (circulation) and prevent the cyst's reclosure. The burr hole was sealed with bone wax and the skin closed with separate sutures.

9.2.1 Results and Complications

Complete drainage was achieved in all but one patient, due to an unaccessible pouch separated from the main cavity. The mean surgical time was 126 min. There were no intraoperative complications and even the postoperative course was uneventful: surprisingly no chemical meningitis was observed and transient hyperpyrexia in three cases rapidly subsided. The mean hospital stay was 7 days. During the follow-up period (range 12–72 months; average 40 months), we observed two recurrences of the cystic portion. Both patients underwent successful repeat neuroendoscopy procedure. One died postoperatively due to unrelated causes (pulmonary embolism) while neuroradiologic examinations had shown complete drainage of the cyst. In the remaining patients, follow-up examinations have demonstrated persistent drainage and resolution of symptoms (Fig. 9.2). One patient (20-year-old man) underwent microsurgical removal of an enlarging solid nodule 9 months after endoscopy (Fig. 9.3).

9.3 Technique of Cystosphenoidostomy

Between January 1985 and June 2012, 21 patients (5 pediatric patients, age range 6–17 years, mean 11 years; 16 adults, age range 35–74 years, mean 59 years) underwent cystosphenoidostomy as surgical treatment for mainly cystic sellar craniopharyngioma. Six patients had preoperative diabetes insipidus requiring medical therapy, 16 patients had some degree of endocrine disturbance requiring hormonal substitution, and all patients had visual field defects that ranged from minimum peripheral deficits to

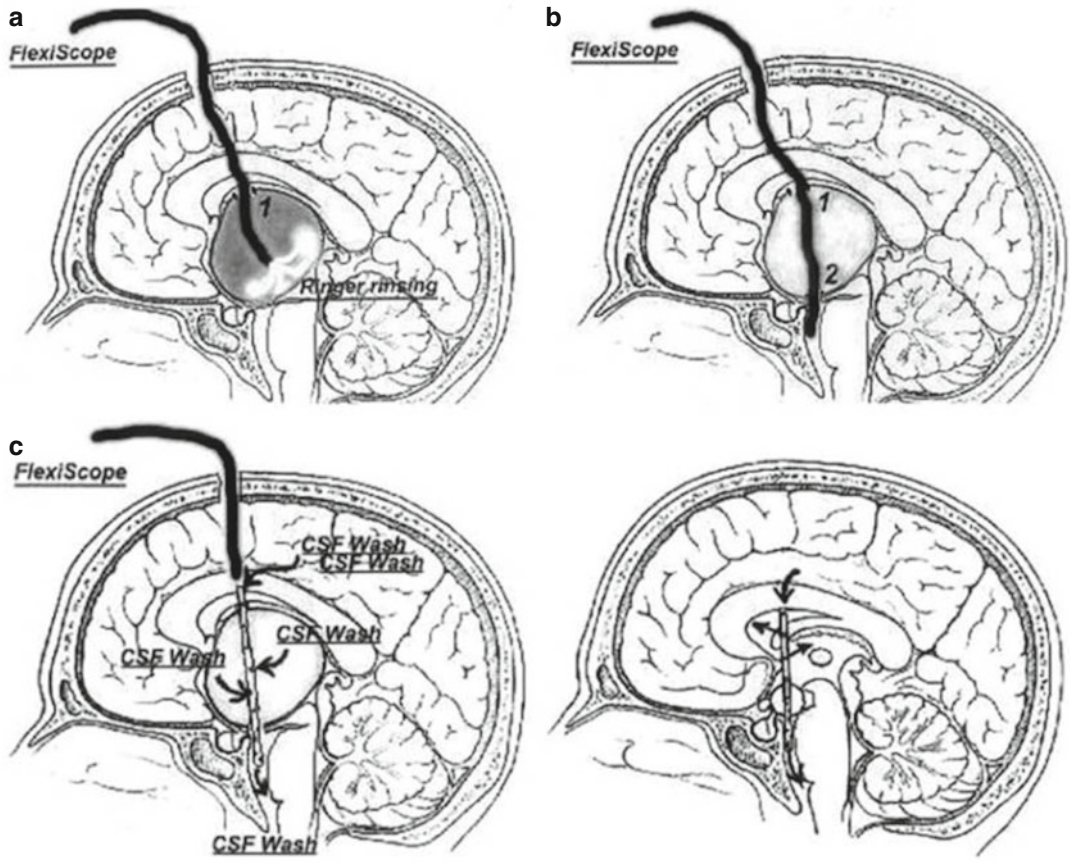

Fig. 9.1 (**a**–**c**) Purely neuroendoscopic, transventricular technique. (**a**) Puncture of the cyst's dome and complete drainage of content by aspiration and washing with Ringer's solution. (**b**) Perforation of the cyst's fundus into the basal cisterns. (**c**) "Stenting" of the cavity and its progressive shrinkage with time (Reproduced with permission from Delitala et al. [15])

monocular blindness in one case. All tumors had an intrasellar component, with sellar enlargement and supradiaphragmatic extension. In all cases the cyst was at least partially within the pituitary fossa. Indications for cystosphenoidostomy were recurrent tumor (6 cases), open surgery refusal (3 cases), unlikeliness of total removal basing on neuroradiological appearance (7 cases), and high anesthesiological risk in case of craniotomy (5 cases). No patients had preoperative hydrocephalus. Recurrent tumors had previously been treated with microsurgical technique either by a transcranial approach (2 cases) or a transsphenoidal approach (4 cases); 3 patients had undergone to radiotherapy as an adjuvant treatment.

Surgical technique consisted in a microsurgical or purely endoscopic transsphenoidal approach. A wide opening in the sellar floor was performed in order to expose the cyst wall. The cyst wall was incised, the contents were suctioned, and cyst walls were dissected as far as possible, taking care in avoiding any lesion to the arachnoid and cerebrospinal fluid leakage. The permanent communication with the sphenoid sinus was established through a self-securing X- or H-shaped silastic catheter, which was positioned over the sellar floor so that two arms lied within the pituitary fossa and the other two in the sphenoid sinus (Fig.9.4).

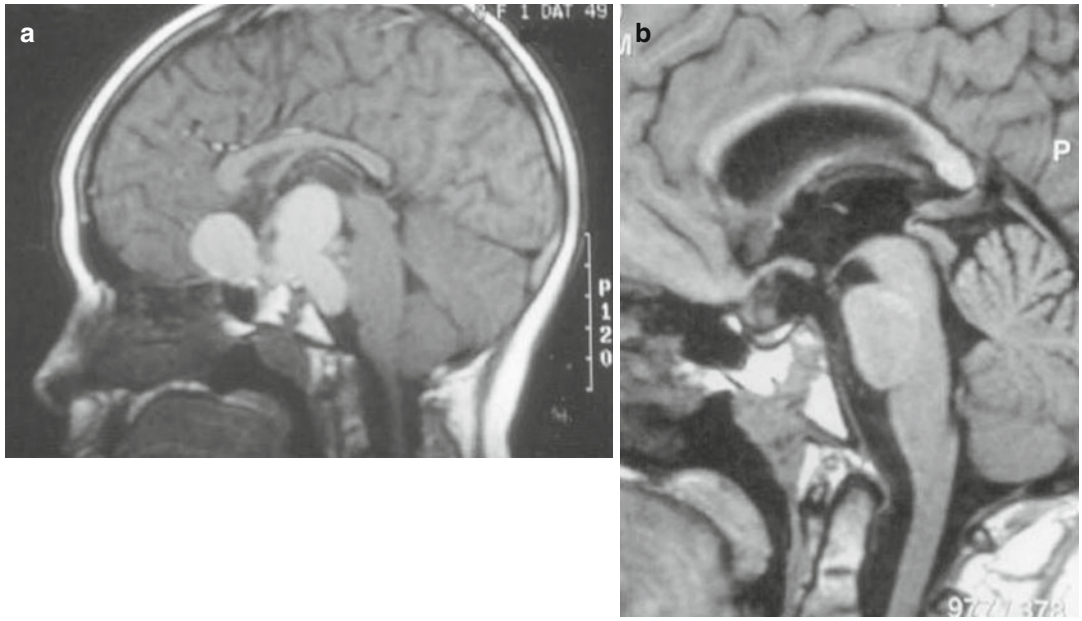

Fig. 9.2 (**a**, **b**) Patient 1: 9-year-old girl with a giant cyst recurrent after microsurgery, external radiotherapy, and sump drainage. (**a**) Pre-neuroendoscopy and (**b**) post-neuroendoscopy MRI scans at 6-year follow-up. The patient has normal visual and endocrine function (Reproduced with permission from Delitala et al. [15])

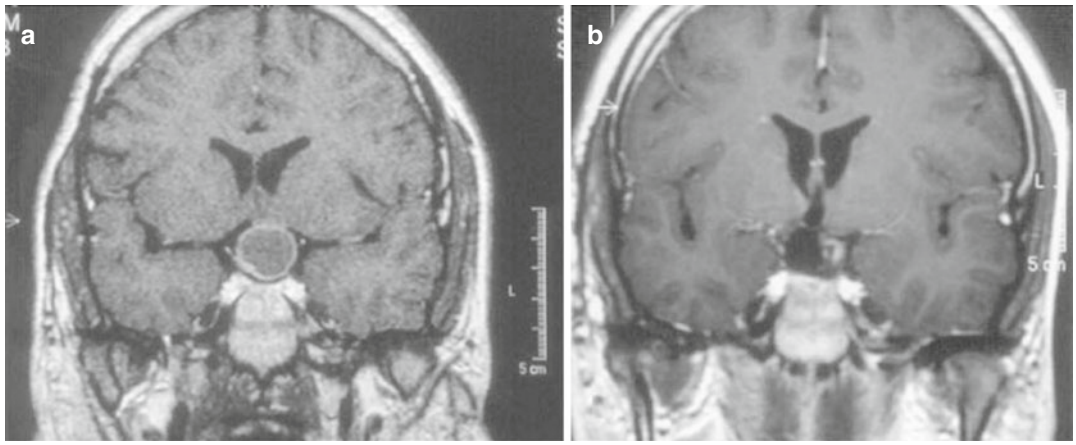

Fig. 9.3 (**a**, **b**) Patient 2: 20-year-old man with a cyst recurring after microsurgery. (**a**) Pre-neuroendoscopy and (**b**) post-neuroendoscopy MRI scans (at 22 months). The cyst did not recur and an enlarging solid subchiasmatic nodule was successfully removed by microsurgery 9 months late (Reproduced with permission from Delitala et al. [15])

9.3.1 Results and Complications

Significant cyst volume decrease was achieved in all patients and visual defects invariably improved in all cases. In one case, cystosphenoistomy was planned but intraoperatively aborted after CSF leakage was suspected. The mean surgical time was 103 min. The mean hospital stay was 5 days. One early major complication, consisting in postoperative severe hyponatremia in a patient with preoperative diabetes insipidus, was observed. We had to remove

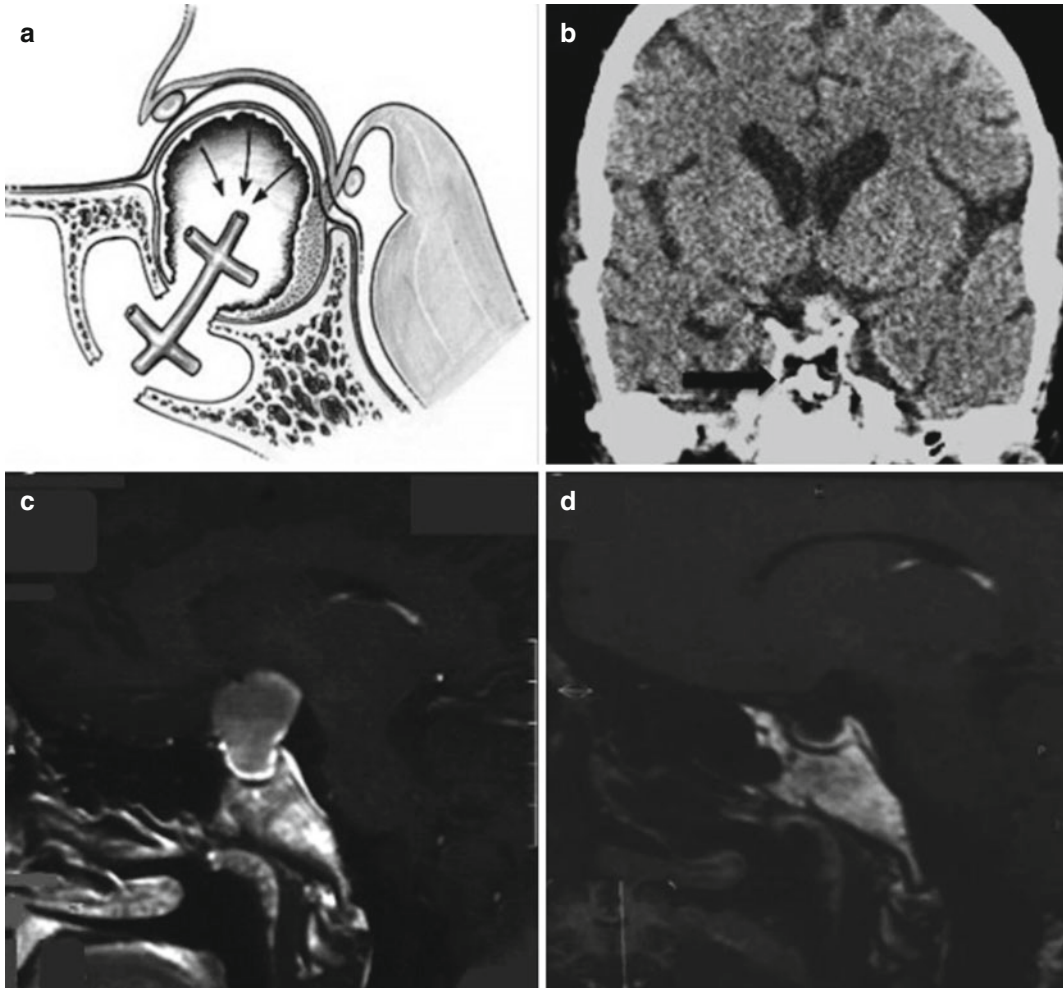

Fig. 9.4 Cystosphenoidostomy: (**a**) scheme of the technique; the homemade H-shaped silastic catheter is placed over the sellar floor, with two arms within the pituitary fossa and two arms in sphenoid sinus. (**b**) Postoperative CT scan showing catheter positioning (*arrow*). (**c**) Preoperative sagittal contrast-enhanced T1w MRI showing a large cystic intra- and suprasellar craniopharyngioma. (**d**) 3-year postoperative sagittal contrast-enhanced T1w MRI showing the cyst disappearance. No visual deficits; mild hypopituitarism under hormone replacement

the catheter in two cases, one CSF leakage that required surgical revision and one case for postoperative recurrent meningitis. Sixteen patients underwent postoperative conformational radiotherapy. In five cases we observed postoperative worsening of a previously existing endocrine disturbance. Follow-up ranged from 8 months to 20 years, and only four patients were lost at follow-up. In four cases we observed a postoperative slow growth of the solid nodule of the tumor, but in no cases a surgical excision was necessary so far.

Conclusions

The results of direct surgical attacks on craniopharyngiomas have enormously improved over the last half century. Surgical mortality has dropped from 41 % [16] before replacement therapy became available to 2 % or less in recent series [1, 2, 17]. However, a closer insight raises several issues. First of all, radicality can be achieved in only 45.7 % of transcranial procedures [1] and an overall long-term good clinical outcome in 60.3 % of patients [17]. Endocrinological sequelae are

the rule and 80 % of Yasargil's patients required substitution therapy [2]. Postoperative diabetes insipidus must be accepted as a common sequela since its incidence increases 3.7 times after surgery [2, 18] and anterior pituitary function is only slightly but constantly affected [18]. More subtle neuropsychological deficits have been observed in surgically treated children in the long term [19]. Focusing on the subgroup of recurrent tumors, total removal is possible in only 21.1 % of craniotomies, and all procedures (craniotomies and transsphenoidal approach) are burdened by a 10.5 % operative mortality [1]. Finally, it must also be emphasized that these figures are quoted from some of the most successful series in the literature: craniopharyngioma is a rare tumor (1.3/million/year) [20] and it is reasonable to assume that less experienced hands would perform less brilliantly. A predominantly cystic tumor is observed in 60 % of craniopharyngioma patients [4]. Since mass effect rather than infiltration is responsible for the clinical syndrome, permanent/repeated cyst drainage seems a suitable compromise in most such cases. Fox has to be credited for the first implant of a sump drainage system in a cystic suprasellar tumor [7]. Similar experiences followed for Miles and Gutin et al. [8, 9]. In all their cases, a catheter connected to the subgaleal reservoir was positioned at craniotomy. Cysto-ventriculostomy, as proposed by Spaziante and de Divitiis, represents an evolution of these early experiences: the authors introduced the concept of marsupialization of the cyst into the ventricles, aiming at continuous dilution of the cyst's fluid and resorption through the CSF pathways [4]. They reported clinical improvement in all patients and a good outcome in the six patients available for follow-up [4]. Periodic aspiration has also been implemented with intracavitary administration of antineoplastic agents in the attempt to overcome the palliative nature of sump drainage. Although β-emitting radiocolloids and bleomycin have proven effective under certain conditions, there have also been some major drawbacks related to isotope handling and radiation damage [10, 21] and the absence of unitary protocols and fatal toxicity. In both cases treatment failures are not uncommon [4, 6, 22–24]. Fluid-filled spaces represent an ideal battlefield for neuroendoscopists: since most craniopharyngioma cysts impinge or grow into the ventricular systems (types c–f according to Yasargil's system), an endoscopic approach seems a suitable option. Minimum invasiveness, a radical cyst drainage, and wide marsupialization into CSF fluid spaces under direct visual control represent clear-cut advantages over other draining techniques. Indeed in the last decade, different institutes have published endoscopic treatment of cystic craniopharyngioma [3, 5, 12, 25] On the other hand, we never attempted nor in the light of our experience recommend an endoscopic resection of solid components due to the risk of uncontrollable damage to neurovascular structures. We prefer to remove as much of the relatively avascular dome and leave behind basal tumor remnants that can later be considered for radiation therapy. In our hands, the neuroendoscopic management of cystic craniopharyngiomas has proven to be safe, repeatable, and effective in the midterm, and it should be especially considered in recurrent tumors and in very young or very old patients. No intraoperative or postoperative complications related to surgery were observed, including chemical meningitis, probably due to careful rinsing of all debris. On the other hand, it must be emphasized that neuroendoscopy is oncologically palliative and not compatible with further intracavitary treatments, which require a sealed cavity. The recurrence rate is undetermined, but seems unrelated to surgical aggressiveness. In only two of our cases, reclosure and filling of the cyst led to clinical symptoms several months after the first procedure and was probably due to insufficient dome stripping. Systematic cisternostomy and stenting of the internal cyst create an additional outflow route to the intracystic CSF circulation and can limit the need for further procedures. Remote recurrence of craniopharyngiomas has been related to intra-

operative transplant or CSF spreading [26], and therefore cysto-ventriculo-cisternostomy should in theory be at risk. However, seeding represents an exceptional event, with only eight cases reported to date in the world literature. All of these cases occurred after open surgery with the manipulation of solid, more cellular tumors. The surgical treatment of craniopharyngiomas is extremely challenging and often success requires a multimodal approach. Dealing with mostly cystic tumors can be deceiving since the microsurgical approach ensures drainage, but only seldom radicality, since stripping of the capsule away from the hypothalamus and major vessels is highly risky. Simple drainage has therefore been achieved by puncture and aspiration. The neuroendoscopic approach represents a major advance compared with classic draining techniques, since emptying is obtained under direct vision and operativity of the instrument allows a partial resection of the capsule for diagnostic purposes and especially wide marsupialization. Perforation into the basal cisterns and stenting are measures to prevent reclosure and recurrence, and other therapeutic modalities (microsurgery, radiosurgery, fractionated external radiation therapy) can later be scheduled to eradicate solid remnants, if indicated. A purely endoscopic approach as a first choice is an alternative to microsurgery, in selected cases. Case selection must be based on anatomo-surgical (cystic/solid rate, Yasargil's type) and clinical (primary/recurrent, age, conditions) considerations: in such cases, if tumor control and satisfactory reintegration into the appropriate social setting are the goals, endoscopy cannot be overlooked.

Cystosphenoidostomy has been proved to be a safe and effective palliative treatment for cystic craniopharyngiomas. In particular, it allows significant reduction of mass effect and resolution of symptoms caused by compressive mechanisms, even if it does not affect tumor growth, and it can be combined to other treatment, such as radiotherapy or radiosurgery, aimed to control tumor progression. This technique can be easily proposed to patients whose tumor has a major cystic component that can be reached through a transsphenoidal approach, without a cisternal layer interposing between the tumor and the sellar floor. Even tumors whose cysts have a voluminous suprasellar development may be approached, as the cyst content is usually fluid and can be simply suctioned [4]. The only absolute contraindication to cystosphenoidostomy is intraoperative CSF leakage detection, as the connection may sustain a permanent CSF fistula, with subsequent intracranial hypotension and alarming infectious sequelae. However, it is mainly a low-risk procedure, as it avoids manipulation of neurovascular structures, provides indirect decompression of optic nerves and chiasm, and has a low incidence of pituitary stalk damage and subsequent diabetes insipidus [13, 14]. Furthermore, cystosphenoidostomy has been proven to be effective and safe even in children, in which hypothalamic damage due to tumor compression or surgical manipulation can induce deplorable long-term sequelae [13].

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