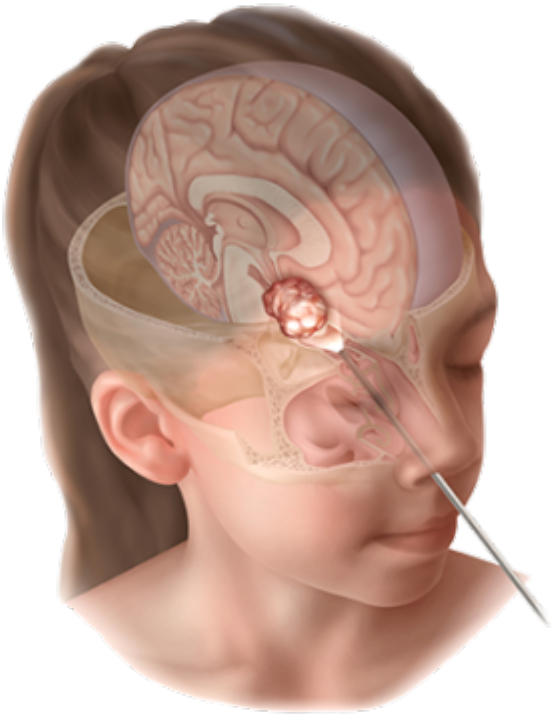
Craniopharyngioma surgery



Historically, aggressive surgical resection was the treatment goal to minimize the risk of tumor recurrence via open transcranial midline, anterolateral, and lateral approaches, but could lead to clinical sequela of visual, endocrine, and hypothalamic dysfunction. However, recent advances in the endoscopic endonasal approach over the last decade have mostly supplanted transcranial surgery as the optimal surgical approach for these tumors. With viable options for adjuvant radiation therapy, targeted medical treatment, and alternative minimally invasive surgical approaches, the management paradigm for craniopharyngiomas has shifted from aggressive open resection to more minimally invasive but maximally safe resection, emphasizing quality of life issues, particularly in regards to visual, endocrine, and hypothalamic function. [1].

Multimodal management of craniopharyngiomas seems to provide a better rate of survival and greater long-term disease control. It is suggested that GKS combined with adjuvant neuroendoscopy should be used as an alternative in treating recurrent or residual craniopharyngiomas if additional microsurgical removal cannot guarantee a cure [2].

Gross total resection of this tumor is the gold standard surgical treatment, but the preservation of neurological and hypothalamopituitary functions is also an important facet in the management of this complex tumor [3].

Microscopic surgical resection and endoscopic cyst fenestration along with intracystic catheter placement of the Ommaya reservoir are the commonly performed surgical techniques

Craniopharyngioma treatment has evolved over time, as our treatment options have expanded and our understanding of the long-term consequences of radical resection has grown. Originally, aggressive resection was the only hope of controlling this tumor; in spite of the often severe morbidity of such an approach, it remained the mainstay of treatment. But as the Craniopharyngioma natural history of this tumor is a recurrence, it may be considered a chronic disease, with the goal of maximizing control but minimizing patient morbidity.

To date, no class I recommendations exist for the best treatment of these tumors. Management should be by a multidisciplinary team (neurosurgery, endocrinology, ophthalmology, psychology, oncology, and radiation oncology) and be individualized for each patient. There are some practical surgical considerations to keep in mind. In cases where total resection can be obtained without significant morbidity (i.e., cases where the tumor is not invading or adherenct to the hypothalamus), GTR remains the treatment of choice. In cases where the tumor is small and the solid portions are primary intrasellar, without significant extension laterally in the suprasellar space or without encasement of vessels, an endoscopic approach may provide good outcomes. In cases where the tumor is densely involved in critical structures and has a significant cystic component causing mass effect, an Ommaya placement with or without chemotherapy infusion may represent a less invasive way to decompress neural structures and control tumor progression.

Finally, in cases of subtotal resection (STR) or recurrent disease, particularly with a favorable margin between the tumor and the optic chiasm, adjuvant RT or SRS is likely to improve progression-free survival. Promising treatments from PBTC trials may offer hope for future therapies with lower side effect profiles. Ultimately, this tumor remains one of the most difficult pediatric neurosurgical problems, and recommendations will continue to evolve [4].

Microsurgical resection has been considered the gold standard of craniopharyngioma treatment, but lately, it has found less favor due to its morbidity and is being replaced by minimally invasive cyst drainage procedures.

The highest priority of craniopharyngioma treatment with surgery is to maximize tumor removal without compromising the patients' long-term functional outcome.

The challenges are due to their proximity to the hypothalamus, circle of Willis, optic chiasm, optic nerve, pituitary gland and pituitary stalk. The hypothalamus and pituitary stalk are often challenging dissection planes, and attempts to preserve these structures are important in mitigating risk associated with resection. Craniopharyngiomas tend to push the surrounding neurovasculature upward and outward from a central location. Open skull base approaches have traditionally been limited by the need for significant brain retraction and for working through small surgical windows around major vasculature. Whereas gross total resection (GTR) provides the patient with the highest rate of progression free survival (PFS), intentional subtotal resection performed to preserve hypothalamic and/or pituitary function by both endoscopic skull base surgery (ESBS) and transcranial surgery (TCS) has become more common, particularly in the pediatric population [5].

Schwartz et al., from the Weill Cornell Brain and Spine Center, compared surgical results for Endoscopic skull base surgery (ESBS) with transcranial surgery (TCS) for several different pathologies

over two different time periods (prior to 2012 and 2012-2017) to see how results have evolved over time. Pathologies examined were craniopharyngioma, anterior skull base meningioma, esthesioneuroblastoma, chordoma, and chondrosarcoma.

ESBS offers clear advantages over TCS for most craniopharyngiomas and chordomas. For well-selected cases of planum sphenoidale and tuberculum sellae meningiomas, ESBS has similar rates of resection with higher rates of visual improvement, and more recent results with lower CSF leaks make the complication rates similar between the two approaches. TCS offers a higher rate of resection with fewer complications for olfactory groove meningiomas. ESBS is preferred for lower-grade esthesioneuroblastomas, but higher-grade tumors often still require a craniofacial approach. There are few data on chondrosarcomas, but early results show that ESBS appears to offer clear advantages for minimizing morbidity with similar rates of resection, as long as surgeons are familiar with more complex inferolateral approaches.

ESBS is maturing into a well-established approach that is clearly in the patients' best interest when applied by experienced surgeons for appropriate pathology. Ongoing critical reevaluation of outcomes is essential for ensuring optimal results [6].

One criticism of comparisons between ESBS and TCS is that smaller tumors may be chosen for ESBS, whereas larger and more complex tumors are chosen for TCS. This would bias the results in favor of ESBS. To address this criticism, Moussazadeh et al. published a comparison study in which all tumors were viewed by 3 surgeons blinded to the approach chosen, to ensure that all tumors could be removed equally well with either approach.

If one approach was favored, then the case was eliminated. In this series, ESBS had a higher rate of GTR (90.0% vs 40.0%; $p = 0.009$), a higher rate of visual improvement (63.0% vs 0.0%; $p < 0.05$), less postoperative FLAIR signal in the frontal lobe (0.16 ± 4.6 cm3 vs 14.4 ± 14.0 cm3; $p < 0.001$), and fewer complications (20.0% vs 80.0%; $p < 0.001$) compared with TCS [7].

In children, for whom minimally invasive approaches are particularly attractive and large tumors are often found, recent series have shown utility and efficacy with ESBS, with high rates of visual improvement and infrequent major complications as well as lower rates of hypothalamic obesity in some, but not all, series [8] [9] [10].

Today the majority of children with craniopharyngioma are treated with a transcranial approach, but this trend may be changing as pediatric specialists partner with skull base colleagues with endoscopic experience and it becomes clear that this approach is at least comparable if not more effective and safer [11] [12]

Qiao et al., in 2018 conducted a systematic review and meta-analysis. They conducted a comprehensive search of PubMed to identify relevant studies. Pituitary, hypothalamus functions and recurrence were used as outcome measures. A total of 39 cohort studies involving 3079 adult patients were included in the comparison. Among these studies, 752 patients across 17 studies underwent endoscopic transsphenoidal resection, and 2327 patients across 23 studies underwent transcranial resection. More patients in the endoscopic group (75.7%) had visual symptoms and endocrine symptoms (60.2%) than did patients in the transcranial group (67.0%, $p = 0.038$ and 42.0%, $p = 0.016$). There was no significant difference in hypopituitarism and panhypopituitarism after surgery between the two groups: 72.2% and 43.7% of the patients in endoscopic group compared to 80.7% and 48.3% in the transcranial group ($p = 0.140$ and $p = 0.713$). They observed same proportions of transient and permanent diabetes insipidus in both groups. Similar recurrence was

observed in both groups (p = 0.131). Pooled analysis showed that neither weight gain (p = 0.406) nor memory impairment (p = 0.995) differed between the two groups. Meta-regression analysis revealed that gross total resection contributed to the heterogeneity of recurrence proportion (p < 0.001). They observed similar proportions of endocrine outcomes and recurrence in both endoscopic and transcranial groups. More recurrences were observed in studies with lower proportions of gross total resection [13].

In 2016 Wannemuehler et al., from the Indiana University School of Medicine, Indianapolis performed a retrospective review of all patients who underwent resection of their histopathologically confirmed craniopharyngiomas between 2005 and 2015. Pediatric patients, revision cases, and patients with tumors greater than 2 standard deviations above the mean volume were excluded. The patients were divided into 2 groups: those undergoing primary TCM and those undergoing a primary EEEA. Preoperative patient demographics, presenting symptoms, and preoperative tumor volumes were determined. Extent of resection, tumor histological subtype, postoperative complications, and additional outcome data were obtained. Statistical significance between variables was determined utilizing Student t-tests, chi-square tests, and Fisher exact tests when applicable.

After exclusions, 21 patients satisfied the aforementioned inclusion criteria; 12 underwent TCM for resection while 9 benefitted from the EEEA. There were no significant differences in patient demographics, presenting symptoms, tumor subtype, or preoperative tumor volumes; no tumors had significant lateral or prechiasmatic extension. The extent of resection was similar between these 2 groups, as was the necessity for additional surgery or adjuvant therapy. CSF leakage was encountered only in the EEEA group (2 patients). Importantly, the rate of postoperative visual improvement was significantly higher in the EEEA group than in the TCM group (88.9% vs 25.0%; p = 0.0075). Postoperative visual deterioration only occurred in the TCM group (3 patients). Recurrence was uncommon, with similar rates between the groups. Other complication rates, overall complication risk, and additional outcome measures were similar between these groups as well.

Based on this study, most outcome variables appear to be similar between TCM and EEEA routes for similarly sized tumors in adults. The multidisciplinary EEEA to craniopharyngioma resection represents a safe and compelling alternative to TCM. The authors' data demonstrate that postoperative visual improvement is statistically more likely in the EEEA despite the increased risk of Cerebrospinal fluid fistula. These results add to the growing evidence that the EEEA may be considered the approach of choice for resection of select confined primary craniopharyngiomas without significant lateral extension in centers with experienced surgeons. Further prospective, multiinstitutional collaboration is needed to power studies capable of fully evaluating indications and appropriate approaches for craniopharyngiomas [14].

In 2012 Komotar et al., performed a MEDLINE search of the literature (1995-2010) to identify open and endoscopic surgical series for pediatric and adult craniopharyngiomas. Comparisons were made for patient and tumor characteristics as well as extent of resection, morbidity, and visual outcome. Statistical analyses of categorical variables were undertaken by the use of $\chi(2)$ and Fisher exact tests with post-hoc Bonferroni analysis to compare endoscopic, microsurgical transsphenoidal, and transcranial approaches.

Eighty eight studies, involving 3470 patients, were included. The endoscopic cohort had a significantly greater rate of gross total resection (66.9% vs. 48.3%; P < 0.003) and improved visual outcome

(56.2% vs. 33.1%; P < 0.003) compared with the open cohort. The transsphenoidal cohort had similar outcomes to the endoscopic group. The rate of Cerebrospinal fluid fistula was greater in the endoscopic (18.4%) and transsphenoidal (9.0%) than in the transcranial group (2.6%; P < 0.003), but the transcranial group had a greater rate of seizure (8.5%), which did not occur in the endonasal or transsphenoidal groups (P < 0.003).

The endoscopic endonasal approach is a safe and effective alternative for the treatment of certain craniopharyngiomas. Larger lesions with more lateral extension may be more suitable for an open approach, and further follow-up is needed to assess the long-term efficacy of this minimal access approach [15].

Craniopharyngioma transcranial surgery

see Craniopharyngioma transcranial surgery.

Craniopharyngioma endoscopic endonasal approach

see Craniopharyngioma endoscopic endonasal approach.

Adherences

Craniopharyngioma (CP) adherence represents a most baffling problem for the neurosurgeon.

A article of Prieto et al., from the Department of Neurosurgery, Puerta de Hierro University Hospital, Madrid, Spain. La Princesa University Hospital, Madrid, Spain. Federal Pathologic-Anatomical Museum in the Narrenturm, Vienna, Austria. Sureste University Hospital, Madrid, Spain. Ramón y Cajal University Hospital, Madrid, Spain, presents a comprehensive review of the pathological, surgical, and radiological sources of evidence supporting that CP adherence, despite being heterogenous, is characterized by repeating patterns. The key underlying factors of CP adherence are also discussed. Three components define the type of adherence for each case: (i) the intracranial structures attached to the tumor, (ii) the adherence morphology, and (iii) the adhesion strength. Combination of these three components gives rise to five hierarchical levels of increased risk of hypothalamic injury during tumor removal. Tumor topography has been identified as the major predictor of the type of CP adherence. The most extensive and strongest adhesions to the hypothalamus occur in CPs originated in the suprasellar cistern that secondarily invade the third ventricle (secondary intraventricular CPs) and in those originated within the third ventricle floor itself (not-strictly intraventricular CPs). Three findings observed on preoperative conventional MRI scans have proven to be reliable predictors of adherence severity. A position of the hypothalamus around the middle portion of the tumor, an amputated pituitary stalk, and an elliptical tumor shape points to the severe and critical risk levels, and in those cases, a safer limited removal is strongly recommended [16].

Endoscopic cyst fenestration along with intracystic catheter

placement of the Ommaya reservoir

Nakamizo A, Inamura T, Nishio S, Inoha S, Ishibashi H, Fukui M. Neuroendoscopic treatment of cystic craniopharyngioma in the third ventricle. Minim Invasive Neurosurg. 2001 Jun;44(2):85-7. doi: 10.1055/s-2001-16003. PMID: 11487790.

Joki T, Oi S, Babapour B, Kaito N, Ohashi K, Ebara M, Kato M, Abe T. Neuroendoscopic placement of Ommaya reservoir into a cystic craniopharyngioma. Childs Nerv Syst. 2002 Nov;18(11):629-33. doi: 10.1007/s00381-002-0638-4. Epub 2002 Sep 28. PMID: 12420123.

Jaiswal S, Jaiswal M, Jaiswal P, Bajaj A, Srivastava C, Chandra A, Ojha BK, Vikas J, Yadav A. Endoscopic Transcortical Transventricular Management of Cystic Craniopharyngioma: Outcome Analysis of 32 Cases at a Tertiary Care Center. Asian J Neurosurg. 2020 Dec 21;15(4):846-855. doi: 10.4103/ajns.AJNS_252_20. PMID: 33708653; PMCID: PMC7869278.

Kuramoto T, Uchikado H, Tajima Y, Tokutomi T, Shigemori M. [Neuroendoscopic placement of the reservoir in an elderly patient with recurrenced craniopharyngioma: case report]. No Shinkei Geka. 2005 Dec;33(12):1207-12. Japanese. PMID: 16359032.

Nakahara Y, Koga H, Maeda K, Takagi M, Tabuchi K. Neuroendoscopic transventricular surgery for suprasellar cystic mass lesions such as cystic craniopharyngioma and Rathke cleft cyst. Neurol Med Chir (Tokyo). 2004 Aug;44(8):408-13; discussion 414-5. doi: 10.2176/nmc.44.408. PMID: 15508348.

Clinical and radiological data of all cystic craniopharyngioma patients treated by transventricular endoscopic cyst drainage and Ommaya placement were retrieved and analyzed.

Results: Thirty-two patients underwent endoscopic cyst drainage during the study period. All patients had immediate clinical and radiological improvement. No significant complications were seen. All patients underwent adjuvant radiotherapy and six patients (18.7%) showed recurrence. Three patients died in the follow-up period.

Conclusions: Endoscopic transcortical transventricular cyst drainage with Ommaya reservoir along with adjuvant radiotherapy is a simple, safe, and effective treatment modality [17)].

Complications

Craniopharyngioma surgery complications

References

1)
Hong CS, Omay SB. The Role of Surgical Approaches in the Multi-Modal Management of Adult Craniopharyngiomas. Curr Oncol. 2022 Feb 24;29(3):1408-1421. doi: 10.3390/curroncol29030118. PMID: 35323318; PMCID: PMC8947636.

2)
Barajas MA, Ramírez-Guzmán G, Rodríguez-Vázquez C, Toledo-Buenrostro V, Velásquez-Santana H, del Robles RV, Cuevas-Solórzano A, Rodríguez-Hernández G. Multimodal management of craniopharyngiomas: neuroendoscopy, microsurgery, and radiosurgery. J Neurosurg. 2002 Dec;97(5 Suppl):607-9. doi: 10.3171/jns.2002.97.supplement. PMID: 12507105.

3)
Yaşargil MG, Curcic M, Kis M, Siegenthaler G, Teddy PJ, Roth P. Total removal of craniopharyngiomas. Approaches and long-term results in 144 patients. J Neurosurg. 1990 Jul;73(1):3-11. doi: 10.3171/jns.1990.73.1.0003. PMID: 2352020.

4)
Reddy GD, Hansen D, Patel A, Lin Y, Jea A, Lam S. Treatment options for pediatric craniopharyngioma. Surg Neurol Int. 2016 Mar 11;7(Suppl 6):S174-8. doi: 10.4103/2152-7806.178570. eCollection 2016. PubMed PMID: 27057397; PubMed Central PMCID: PMC4804398.

5)
Cheng J, Fan Y, Cen B: Effect of preserving the pituitary stalk during resection of craniopharyngioma in children on the diabetes insipidus and relapse rates and long-term outcomes. J Craniofac Surg 28:e591–e595, 2017

6)
Schwartz TH, Morgenstern PF, Anand VK. Lessons learned in the evolution of endoscopic skull base surgery. J Neurosurg. 2019 Feb 1;130(2):337-346. doi: 10.3171/2018.10.JNS182154. Review. PubMed PMID: 30717035.

7)
Moussazadeh N, Prabhu V, Bander ED, Cusic RC, Tsiouris AJ, Anand VK, et al: Endoscopic endonasal versus open transcranial resection of craniopharyngiomas: a casematched single-institution analysis. Neurosurg Focus 41(6):E7, 2016

8) , 11)
Alalade AF, Ogando-Rivas E, Boatey J, Souweidane MM, Anand VK, Greenfield JP, et al: Suprasellar and recurrent pediatric craniopharyngiomas: expanding indications for the extended endoscopic transsphenoidal approach. J Neurosurg Pediatr 21:72–80, 2018

9)
Ali ZS, Lang SS, Kamat AR, Adappa ND, Palmer JN, Storm PB, et al: Suprasellar pediatric craniopharyngioma resection via endonasal endoscopic approach. Childs Nerv Syst 29:2065–2070, 2013

10)
Lin Y, Hansen D, Sayama CM, Pan IW, Lam S: Transfrontal and transsphenoidal approaches to pediatric craniopharyngioma: a national perspective. Pediatr Neurosurg 52:155–160, 2017

12)
Bakhsheshian J, Jin DL, Chang KE, Strickland BA, Donoho DA, Cen S, et al: Risk factors associated with the surgical management of craniopharyngiomas in pediatric patients: analysis of 1961 patients from a national registry database. Neurosurg Focus 41(6):E8, 2016

13)
Qiao N. Endocrine outcomes of endoscopic versus transcranial resection of craniopharyngiomas: A system review and meta-analysis. Clin Neurol Neurosurg. 2018 Apr 7;169:107-115. doi: 10.1016/j.clineuro.2018.04.009. [Epub ahead of print] Review. PubMed PMID: 29655011.

14)
Wannemuehler TJ, Rubel KE, Hendricks BK, Ting JY, Payner TD, Shah MV, Cohen-Gadol AA. Outcomes in transcranial microsurgery versus extended endoscopic endonasal approach for primary resection of

adult craniopharyngiomas. Neurosurg Focus. 2016 Dec;41(6):E6. PubMed PMID: 27903125.
15)

Komotar RJ, Starke RM, Raper DM, Anand VK, Schwartz TH. Endoscopic endonasal compared with microscopic transsphenoidal and open transcranial resection of craniopharyngiomas. World Neurosurg. 2012 Feb;77(2):329-41. doi: 10.1016/j.wneu.2011.07.011. Epub 2011 Nov 1. Review. PubMed PMID: 22501020.
16)

Prieto R, Pascual JM, Hofecker V, Winter E, Castro-Dufourny I, Carrasco R, Barrios L. Craniopharyngioma adherence: a reappraisal of the evidence. Neurosurg Rev. 2018 Jul 24. doi: 10.1007/s10143-018-1010-9. [Epub ahead of print] Review. PubMed PMID: 30043262.
17)

Jaiswal S, Jaiswal M, Jaiswal P, Bajaj A, Srivastava C, Chandra A, Ojha BK, Vikas J, Yadav A. Endoscopic Transcortical Transventricular Management of Cystic Craniopharyngioma: Outcome Analysis of 32 Cases at a Tertiary Care Center. Asian J Neurosurg. 2020 Dec 21;15(4):846-855. doi: 10.4103/ajns.AJNS_252_20. PMID: 33708653; PMCID: PMC7869278.