Ventriculoscopic Surgery for Cystic Retrochiasmatic Craniopharyngiomas: Indications, Surgical Technique, and Short-Term Patient Outcomes

Todd C. Hollon, MD
Luis E. Savastano, MD
David Altshuler, MD, MS
Ariel L. Barkan, MD
Stephen E. Sullivan, MD

Department of Neurosurgery, University of Michigan, Ann Arbor, Michigan

Correspondence:
Stephen E. Sullivan, MD,
Department of Neurosurgery,
University of Michigan,
1500 E. Medical Center Drive, Room 3552,
Taubman Center,
Ann Arbor, MI 48109-5338.
E-mail: ssulliva@med.umich.edu

Received, January 25, 2017.
Accepted, September 19, 2017.

BACKGROUND: Attempted gross-total resection for the management of cystic retrochiasmatic craniopharyngiomas can cause severe hypothalamic dysfunction and decrease overall survival. Ventriculoscopic surgery is a minimally invasive alternative; however, potential indications and technique have not been well defined.

OBJECTIVE: To present our indications and technique for the ventriculoscopic treatment of cystic retrochiasmatic craniopharyngiomas.

METHODS: We evaluated all patients with retrochiasmatic craniopharyngiomas for ventriculoscopic surgery. Indications and operative technique were developed to minimize operative morbidity, relieve mass effect, and optimize functional outcome. Cyst size and functional outcomes were statistically evaluated to determine radiographic and short-term clinical outcome.

RESULTS: Indications for ventriculoscopic surgery included (1) radiographic evidence of hypothalamic involvement and (2) major cystic component. Ten patients met indications, and mean follow-up was 2.5 $\pm$ 1.6 yr. The surgical technique included wide cyst fenestration at the foramen of Monro, and fenestration of inferior cyst wall/third ventriculostomy ("through-and-through" technique). Preoperative Karnofsky performance status was 70 $\pm$ 15 and was inversely correlated with preoperative cyst size (13 $\pm$ 13 cm^3). A statistically significant reduction in cyst size was found on early postoperative imaging (2.1 $\pm$ 4.3 cm^3). Seven patients received postoperative radiotherapy. Postoperative performance scores (81 $\pm$ 8.3) had improved; no patient suffered functional decline. Pre- and postoperative body mass indices were similar. No patient had short-term hypothalamic obesity.

CONCLUSION: Ventriculoscopic surgery, with or without adjuvant treatments, can reduce early postoperative tumor volume and improve short-term functional status in cystic retrochiasmatic craniopharyngiomas with hypothalamic involvement; it should be considered a minimally invasive option in the multimodal treatment of craniopharyngiomas. Further studies are needed to determine long-term efficacy.

KEY WORDS: Cystic tumors, Hypothalamic dysfunction, Ventriculoscopic surgery, Retrochiasmatic craniopharyngioma, Neuroendoscopic surgery

Operative Neurosurgery 0:1–11, 2017 DOI: 10.1093/ons/opx220

Retrochiasmatic craniopharyngiomas are challenging neurosurgical lesions due to their deep anatomic location, hypothalamic involvement, and high morbidity. Craniopharyngiomas in this location represent a tumor subgroup with the highest surgical morbidity following radical resection. Aggressive attempts at gross-total resection are associated with significant morbidity and mortality and have highly variable success rates (18%-90% gross-total resection).[1-8] Postoperative visual decline, worsened endocrinopathy, and hypothalamic dysfunction (eg, hypothalamic obesity and neurocognitive deficits) are common following attempted gross-total resection for retrochiasmatic craniopharyngiomas.[9,10] Therefore, the management of retrochiasmatic

ABBREVIATIONS: BMI, body mass indices; **CSF,** cerebrospinal fluid; **CT,** computed tomography; **FLAIR,** fluid-attenuated inversion recovery; **KPS,** Karnofsky performance status, MRI, magnetic resonance imaging

craniopharyngiomas requires a multidisciplinary treatment strategy to achieve optimal surgical results and patient outcome.

Minimally invasive techniques with the surgical goal of tumor debulking have been developed to reduce postoperative morbidity and alleviate symptoms related to tumor burden and mass effect. Craniotomy for subtotal resection, transsphenoidal or endoscopic endonasal approach, stereotactic cyst aspiration with or without reservoir placement, cyst chemotherapy, and stereotactic radiosurgery have all been used as alternative, less-invasive surgical strategies.[11]

Over the last decade, ventriculoscopic surgery has emerged as a minimally invasive technique for cystic retrochiasmatic craniopharyngiomas and is a useful tool in the neurosurgical armamentarium to ameliorate the risk of intraoperative hypothalamic injury.[12-17] Although several small series have been published, a systematic investigation of ventriculoscopic surgery as a treatment strategy has not been completed, and the indications are poorly defined. Here, we present our indications and surgical technique on ventriculoscopic surgery for cystic retrochiasmatic craniopharyngiomas. We provide short-term clinical outcome information on early postoperative cyst decompression and functional status. We aim to better define the role of ventriculoscopic surgery for craniopharyngioma with the hope that future larger studies will elucidate surgical efficacy.

METHODS

Patient Selection

All patients included in the study consented to the surgery at our institution by the senior author (S.E.S.). Among the cohort of patients diagnosed with craniopharyngiomas, all patients with retrochiasmatic craniopharyngioma since 2009 were prospectively considered for ventriculoscopic surgery with or without postoperative radiation after Institutional Review Board approval. A detailed description of the inclusion criteria and clinical indications can be found below in Operative Technique subsection. Twelve patients with cystic retrochiasmatic lesions were identified as surgical candidates. One patient was diagnosed with a colloid cyst, and another was found to have a large cystic pituitary adenoma. Both were excluded from the study. For the 10 patients diagnosed with retrochiasmatic craniopharyngiomas treated with ventriculoscopic surgery, chart review was completed to determine clinical history, radiographic features, surgical management, postoperative radiation, and functional status. Hypopituitarism was defined as need for anterior pituitary hormone replacement. Panhypopituitarism was defined as need for both anterior and posterior hormone pituitary replacement. Preoperative and postoperative follow-up body mass indices (BMIs) were recorded. Karnofsky performance status (KPS) was determined based on clinical documentation both preoperatively and at the time of most recent follow-up visit. A statistically significant increase in BMI was interpreted as evidence for the development of postoperative hypothalamic obesity. As defined in previous investigations, hypothalamic involvement was defined as increased T2/fluid-attenuated inversion recovery (FLAIR) signal in unilateral or bilateral hypothalami.[8,18,19]

Statistical Analysis

Statistical analysis was completed to determine cyst size reduction immediately postoperatively and at final follow-up. Cyst size was measured using the greatest dimension in either the axial, sagittal, or coronal planes on gadolinium-enhanced T1-weighted magnetic resonance imaging (MRI). Cyst volume was measured by summing the axial surface area multiplied by the number of slices and MRI slice thickness. For both cyst greatest dimension and volume, the T1-weighted hypoattenuated cystic center was measured excluding the enhancing cyst capsule. Preoperative and postoperative KPS scores were analyzed to determine the effect of ventriculoscopic surgery on functional status and clinical outcome. Age, BMI, KPS score, largest cyst dimension, and cyst volume were analyzed as continuous variables. Sex was analyzed as a categorical value. *P*-values were calculated using a 2-tailed paired Wilcoxon signed-ranked test (nonparametric paired difference test) to avoid assumption of normal distribution. Statistical correlation was determined using Pearson's product-moment correlation. Statistical significance was defined as *P*-value less than .05. Statistical analyses were completed using software from the R Project of Statistical Computing (https://www.r-project.org/).

Operative Technique

Preoperative Evaluation

Preoperative imaging should include MRI with and without gadolinium to evaluate cyst morphology and hypothalamic involvement. A right-sided approach is preferred when planning the surgical approach, unless cyst asymmetry is such that a left-sided approach is deemed more effective. Figure 1 shows an example of a predominately right-sided third ventricular cystic craniopharyngioma treated ventriculoscopically via a right-sided approach. Figure 2 shows neuronavigation is critical given the small ventricular size in many of these patients. We recommend thin-slice (≤2 mm) contrasted computed tomography (CT) or CT angiography to examine the bony anatomy of the sella and the location of basilar artery apex in the interpeduncular cistern. In our experience, standard endoscopic landmarks such as the mammillary bodies and the infundibular recess are distorted due to tumor mass effect, making bony structures (eg, dorsum sella) more reliable to guide endoscopic third ventriculostomy.

Indications for Ventriculoscopic Surgery

Retrochiasmatic craniopharyngiomas extending into the third ventricle are considered for ventriculoscopic surgery (Figure 1). The strongest indication for ventriculoscopic surgery is radiographic evidence of hypothalamic involvement (Figure 2). Favorable radiographic characteristics include (1) high cystic/solid component ratio (>50% cystic), (2) ventriculomegaly, and (3) cystic portion within the anterior third ventricle. A high clinical suspicion for craniopharyngioma is essential to avoid inappropriate ventriculoscopic surgery for other cystic lesion better treated via an endonasal approach (eg, cystic pituitary adenoma). Patients without a significant cystic proportion and optic chiasm compression underwent open tumor debulking via either a translaminar terminalis or endonasal approach based on anatomic factors. Noncystic tumor without optic compression would undergo endoscopic biopsy and postoperative radiation. Risks associated with ventriculoscopic surgery include forniceal injury, uncontrolled bleeding, and seizures and must be weighed against the benefits of other treatment. Using hypothalamic involvement as the major decision point, our treatment algorithm for retrochiasmatic craniopharyngiomas can be found in Figure 3.

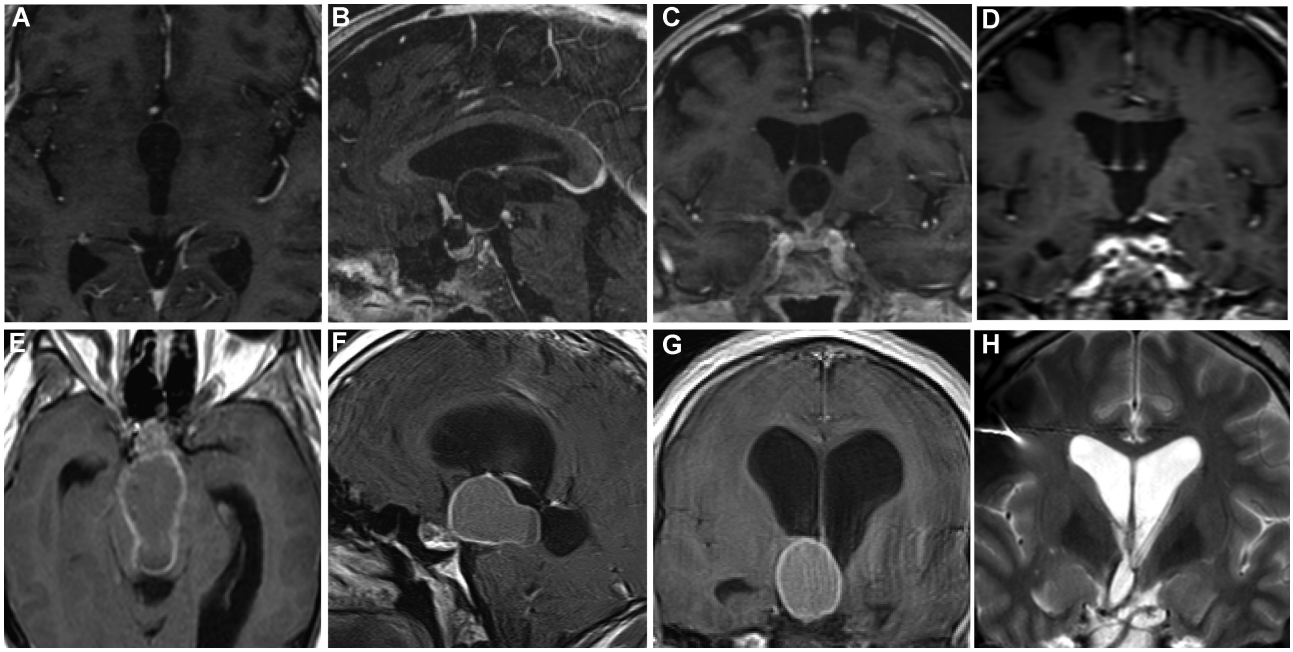

FIGURE 1. *Representative examples of primary and recurrent cystic retrochiasmatic craniopharyngiomas. Top panel (**A**-**D**) shows a primary cystic tumor (patient 5) and bottom panel (**E**-**H**) shows a cystic recurrence (patient 7). **A** and **E**, Preoperative axial gadolinium-enhanced T1-weighted MRIs show enhancing cyst wall with fluid layer within the cyst. **B** and **F**, Sagittal image shows extension from suprasellar region into the third ventricle. **C** and **G**, Coronal reconstructions aid surgical approach and operative planning. **D** and **H**, Immediate postoperative coronal images show early cyst decompression.*

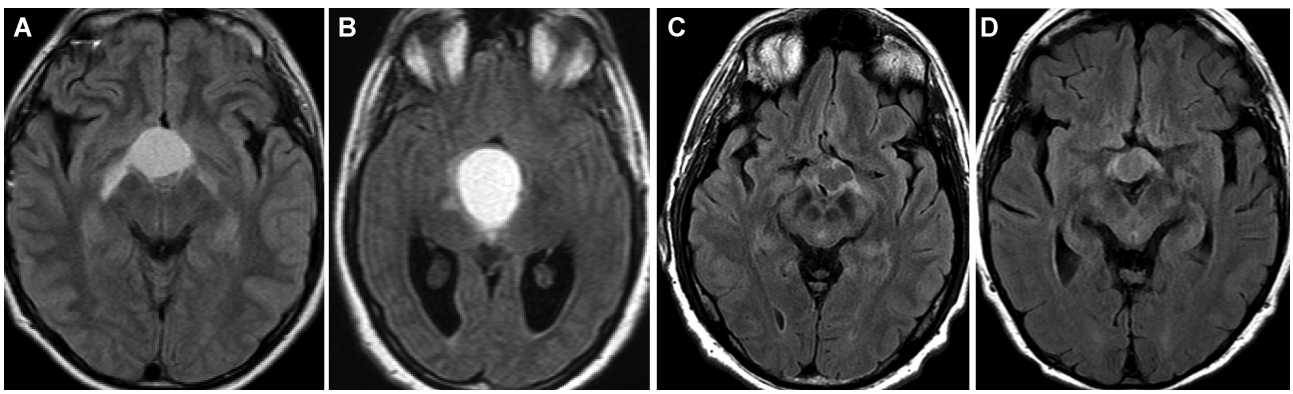

FIGURE 2. *Hypothalamic involvement of cystic retrochiasmatic craniopharyngioma. Axial T2-weighted fluid-attenuated inversion recovery (FLAIR) sequences of patients with hyperintensity in the hypothalamus and evidence of hypothalamic involvement. Images displayed are from (**A**) patient 6, (**B**) patient 7, (**C**) patient 8, and (**D**) patient 5. Patients in **A** and **B** are young patients with recurrent tumors. Patient in **C** is an older patient with recurrence, and patient in **D** is a primary cystic tumor. Bilateral hypothalamic involvement was found in patients with both large (**A** and **B**) and comparatively smaller (**C** and **D**) cystic retrochiasmatic craniopharyngiomas.*

Surgical Technique

The patient is positioned supine in rigid head fixation using a Mayfield® (Schaerer Mayfield USA Inc, Cincinnati, Ohio) head holder. The head is positioned in neutral position with approximately 15° neck flexion. Planning of incision is done using both anatomic landmarks and neuronavigation (Figure 4). An ideal trajectory starts with a burr hole at the coronal suture passing through the ipsilateral foramen of Monro into the cyst contents, and then ending at the tuber cinereum over the interpeduncular cistern just anterior to the basilar artery apex ("through-and-through" technique). The burr hole entry point is planned based on

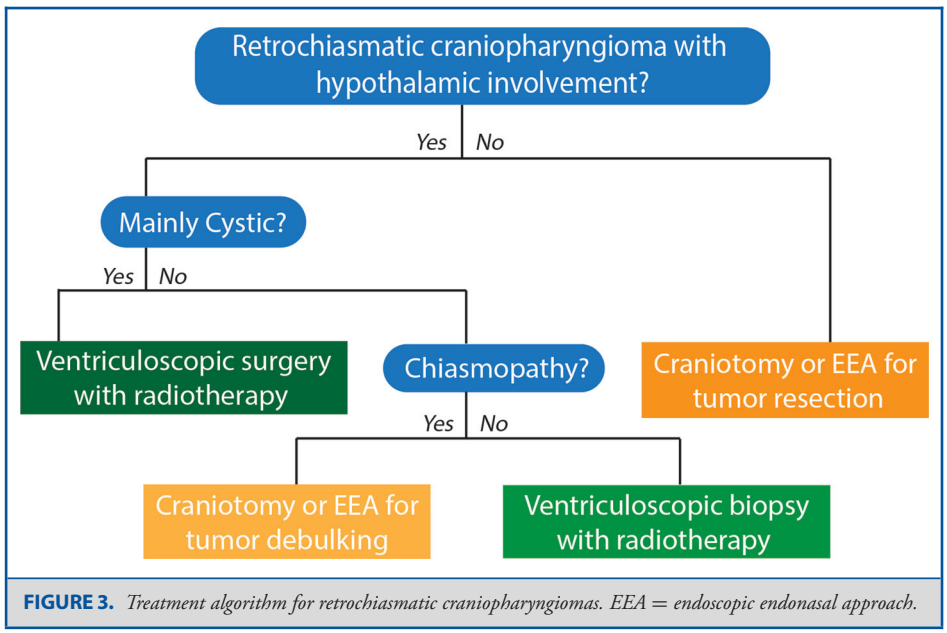

FIGURE 3. *Treatment algorithm for retrochiasmatic craniopharyngiomas. EEA = endoscopic endonasal approach.*

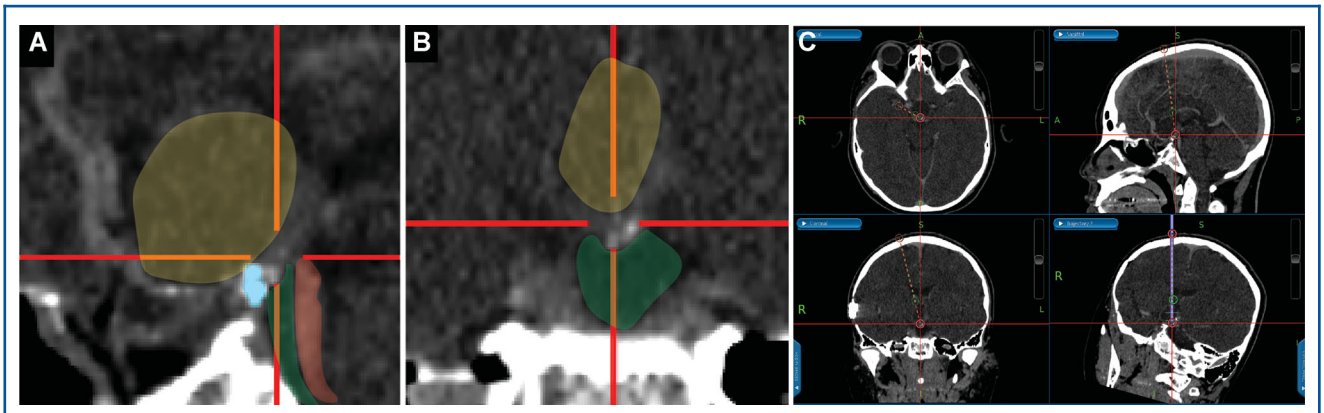

FIGURE 4. *Stereotactic navigation planning for ventriculoscopic surgery in patient 1.* **A** *and* **B**, *Target (cross hairs) is set on the tuber cinereum/inferior cyst wall just posterior to the dorsum sella (light blue) and anterior to the basilar artery apex (red) in the interpeduncular cistern (green).* **C**, *Trajectory is set by connecting the target (red circle) with a second target at the foramen of Monro (green circle) and extending the line to the outer table of the skull. A burr hole is planned over this navigated entry point.*

a single trajectory through the foramen of Monro and ending at a target point at the tuber cinereum.

After placement of the burr hole, a ventricular catheter is passed along the planned trajectory using stereotactic guidance. This is done to confirm accurate stereotactic registration and for continuous irrigation during ventriculoscopic surgery. A peel-away introducer sheath is then passed adjacent to the ventricular catheter under stereotactic guidance to the lateral ventricle just distal to the ependymal surface. A 0° rigid fiberoptic endoscope is then passed through the introducer sheath into the lateral ventricle (Figure 5A). Orientation of the choroid plexus, anterior septal vein, and thalamostriate vein is used to confirm location within the ipsilateral ventricle. The cyst is easily visualized through the foramen of Monro (Figure 5B). The cyst wall is cauterized using monopolar cautery (Figure 5C) and fenestrated using endoscopic scissors (Figure 5D). Cyst fluid is then allowed to egress (Figure 5E) and is irrigated away until cerebrospinal fluid (CSF) is clear (Figure 5F). Continuous high-volume irrigation through the ventricular catheter reduces the risk of postoperative inflammatory meningitis. Cyst fenestration is aggressively expanded using ventriculoscopic side-biting microdebrider and NeuroBalloon catheter (Integra LifeSciences Corp, Plainsboro, New Jersey). Ventricular irrigation is changed from the ventricular catheter to the endoscopic irrigation port. The cyst is entered through the fenestration (Figure 6A) and irrigated for several minutes until cyst contents are completely cleared (Figure 6B). Exophytic tumor

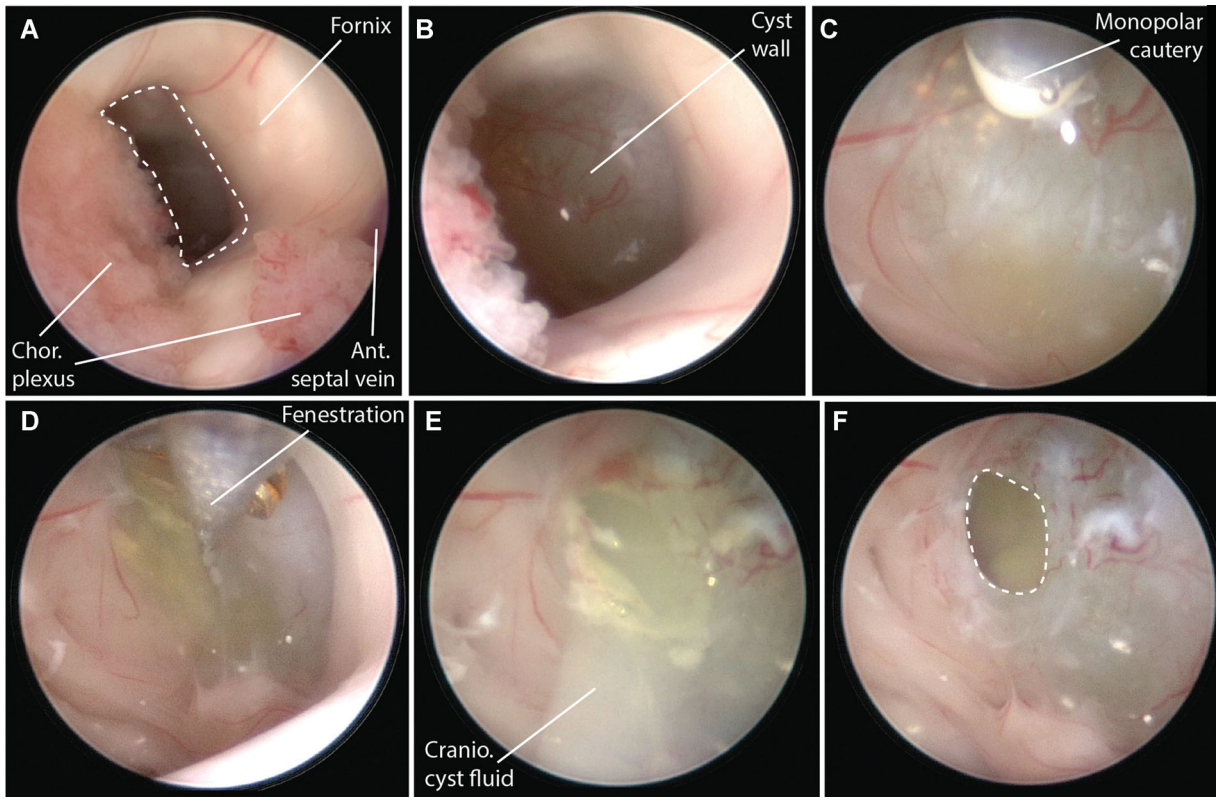

FIGURE 5. *Intraoperative ventriculoscopic images of cyst fenestration from patient 5.* **A**, *Choroid plexus, anterior septal vein, and fornix are visualized to confirm entrance into right lateral ventricle with visualization of foramen of Monro (dashed white line).* **B**, *Craniopharyngioma cyst wall is well visualized through the foramen.* **C**, *Monopolar cautery is used to coagulate small vessels running in the cyst wall portion selected for fenestration. Care is taken to avoid thermal injury to the ipsilateral fornix located anterosuperiorly.* **D**, *Endoscopic scissors are used to puncture and fenestrate the cyst wall.* **E**, *Egress of craniopharyngioma cyst fluid is routinely encountered and is irrigated away until ventricular CSF is clear and fenestration easily visualized (dashed white line in* **F**). *The cyst wall is further resected using microdebrider to widen fenestration (not shown).*

bulk within the cyst is biopsied and resected using tumor forceps and microdebrider. Only that portion which is nonadherent to the third ventricular walls is resected.

Attention is turned toward completing the inferior cyst wall fenestration and third ventriculostomy if deemed to be safe by the surgeon. Standard anatomic landmarks, such as the mammillary bodies, infundibular recess, and optic chiasm, are usually distorted due to tumor mass effect. Bony landmarks have been more reliable in our operative experience. The tip of the NeuroBalloon (Integra Lifesciences Corp) is used to gently palpate in the region of the dorsum sella (Figure 6B). Palpation proceeds posteriorly until no bony prominence is felt. The tip of the NeuroBalloon is then used to perforate through the interior cyst wall into the interpeduncular cistern. We recommend against using monopolar or bipolar cautery to avoid vascular injury. The perforation is then explored with the endoscope looking for basilar artery within the interpeduncular cistern (Figure 6C). Once identified, the perforation is dilated using the NeuroBalloon. Fenestration is explored again to confirm wide fenestration of tuber cinereum and Liliequist membrane with visual confirmation of basilar artery and no evidence of vascular injury (Figure 6D). To prevent possible vascular complications, we recommend against this technique if the surgeon cannot confidently identify the dorsum sella and interpeduncular cistern prior to making the inferior cyst wall fenestration.

RESULTS

Patient Characteristics and Preoperative Radiographic Features

Ten patients underwent ventriculoscopic surgery for cystic retrochiasmatic craniopharyngioma at our institution since 2009 (Table 1), with patients divided into primary cystic and cystic recurrence groups. Five patients had at least 1 previous surgical resection and presented with cystic recurrence. Clinical presentation fell into 3 categories: visual deficit, hydrocephalus, and endocrinopathy. Five patients presented with a visual deficit, 4 with hydrocephalus, and 2 with endocrinopathy. For all included patients, mean preoperative BMI was 33 ± 5.8, and mean KPS score was 70 ± 15 (range 50-90). Patients presenting with hydrocephalus tended to have worse KPS scores (n = 4; mean KPS

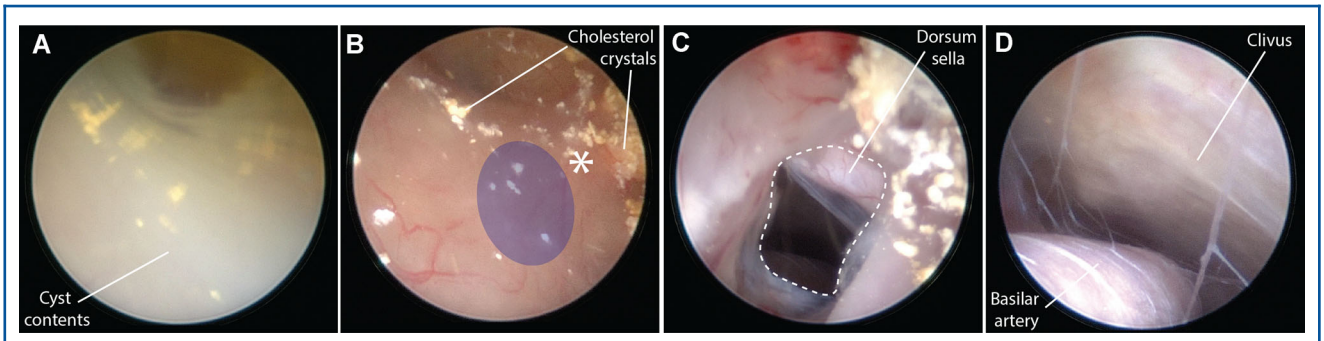

FIGURE 6. *Intraoperative ventriculoscopic images of cyst irrigation, exploration, and third ventriculostomy in patient 5.* **A**, *After entering the cyst, irrigation is used to clear cyst contents and improve visualization.* **B**, *Endoscopic exploration of cyst anatomy and palpation for bony prominences at inferior cyst wall is completed to identify the dorsum sella (white asterisk) and suspected location of the interpeduncular cistern (blue oval).* **C**, *Third ventriculostomy reveals the dorsum sella and interpeduncular cistern.* **D**, *Basilar artery and clivus is visualized to confirm inferior cyst wall fenestration. Neuroballoon dilation is used to ensure wide fenestration of cyst wall and Liliquest's membrane.*

score 60, range 50-70) compared to nonhydrocephalic patients (n = 6; mean KPS score 78, range 50-90) due to progressive cognitive decline and failure to thrive. Prolactin levels were elevated in patients 1 and 10 (37 and 41.2 ng/mL, respectively), who presented with endocrinopathy. Preoperative cyst size for each patient can be found in Table 1. Mean preoperative cyst volume was larger in the cystic recurrence group but did not reach statistical significance ($P = .22$).

Preoperative functional status was inversely correlated with cyst size. Both cyst greatest dimension ($P = .01$) and cyst volume ($P = .04$) were inversely correlated with lower preoperative KPS scores.

Perioperative Clinical Course and Cyst Size Reduction

No perioperative complications occurred during ventriculoscopic surgery in our series. No patient required conversion to an open procedure. Three patients did not undergo an endoscopic third ventriculostomy due to complicated cyst structure and anatomic distortion of the suprasellar region. Two patients who presented with hydrocephalus had ventriculoperitoneal shunts placed at the time of surgery due to large cyst size (patient 7) and larger solid component (patient 4). No patient had postoperative chemical meningitis or refractory headaches from spillage of cyst contents. No patient suffered worsened endocrinopathy related to hypothalamic injury. No patient had rapid visual decline after acute cyst decompression of optic apparatus. All postoperative cyst size measurements were done on MRIs completed within 48 h of surgery. Mean postoperative cyst greatest dimension was 13 ± 5.2 mm in the primary cystic group and 16 ± 4.0 mm in the cystic recurrence group (Table 2). A statistically significant reduction in both cyst greatest dimension and volume was found with *P*-values of .002 and .002, respectively (Figure 7).

Clinical and Radiographic Short-Term Outcome

A summary of each patient's postoperative clinical course can be found in Table 2.

Of the 5 patients who presented with visual symptoms, 4 patients had stable or improved visual deficits after ventriculoscopic surgery. One patient (patient 8) had repeat ventriculoscopic surgery after worsening of visual symptoms, which then improved after a second fenestration. No patient suffered delayed hydrocephalus after cyst fenestration. A statistically significant improvement ($P = .034$) in KPS scores at early postoperative follow-up (<2 mo) was found compared to preoperative functional status (82 ± 8.4 for primary cystic group; 80 ± 10 for cystic recurrence group). No patient suffered a functional decline after ventriculoscopic surgery.

Mean follow-up time was 2.5 (±1.6) yr for the cohort. Seven patients received conformal radiation therapy after ventriculoscopic surgery, with an average radiation dose of 54 ± 3.5 Gy. Panhypopituitarism occurred in 5/7 patients who received postoperative radiation therapy. No patient who received ventriculoscopic fenestration followed by radiation required craniotomy or endoscopic surgery for tumor recurrence. Of the 3 patients who did not receive radiation, 1 required a craniotomy and an endoscopic endonasal approach for resection of solid tumor recurrence. Mean postoperative BMI was 33.9 ± 6.1, which did not significantly increase from preoperative BMI ($P = .50$). One patient (patient 9) did transition from class I obesity (BMI 30-34.9) to class II obesity (BMI 35-39.9). There was no clinical or radiographic evidence of central nervous system dissemination of craniopharyngioma or sequela from cyst fluid in the ventricular system. Cyst greatest dimension had decreased from 14.6 ± 4.6 mm postoperatively to 7.4 ± 7.3 ($P = 0.048$) at final follow-up. Figure 7 provides a summary of cyst size reduction and clinical outcomes at final follow-up. An illustrative case (patient 9) of radiographic response is presented in Figure 7.

DISCUSSION

We present a prospective case series of cystic retrochiasmatic craniopharyngiomas treated with ventriculoscopic surgery with

TABLE 1. Patient Characteristics

Patient	Sex	Age (yr)	Prior treatment	Preoperative endocrine function, visual, and neurological symptoms	Preop body-mass index	Preop KPS	Preop cyst size, largest dimension (mm)	Preop cyst size, volume (cm^3)
Primary cystic group								
1	F	34	None	Amenorrhea	27.4	90	12.8	1.3
2	F	57	None	Dense right temporal hemianopsia, left superior quadrant anopsia	44.2	70	15	1.8
3	M	59	None	Junctional scotoma right > left, hydrocephalus	30.7	60	30	15.8
4	M	59	None	Gait instability, hydrocephalus	35.5	60	29.2	13.6
5	F	78	None	Memory decline, gait instability, hydrocephalus	38.1	70	19.3	2.57
Means		57 ± 16			35 ± 6.5	70 ± 12	21 ± 8.0	7.0 ± 7.1
Cystic recurrence group								
6	M	15	Pterional craniotomy, EEA	Panhypopit; progressive visual decline	24.5	80	28	11.4
7	M	23	Transphenoidal approach	Hypopit; gait instability, hydrocephalus	33.4	50	41.5	36
8	M	54	Bifrontal craniotomy	Bitemporal hemianopsia, and progressive visual decline; normal endocrine	32.3	50	27.1	10.3
9	F	60	Pterional craniotomy	Progressive right temporal hemianopsia	33.3	90	18.4	3.3
10	M	66	Transphenoidal, pterional craniotomy at recurrence	Hypopit, cyst expansion on repeat imaging	26.6	80	40	33.5
Means		43 ± 23			30 ± 4.2	70 ± 18	31 ± 9.6	19 ± 15

EEA, endoscopic endonasal approach; hypopit, hypopituitarism; KPS, Karnofsky performance status; panhypopit, panhypopituitarism; preop, preoperative

a detailed description of the indications, surgical technique, and short-term clinical outcomes. Retrochiasmatic craniopharyngiomas with (1) evidence of hypothalamic involvement and (2) major cystic component are ideal candidates for ventriculoscopic surgery in our experience. We favor this technique for cystic retrochiasmatic craniopharyngiomas to avoid exacerbating hypothalamic dysfunction, the primary risk factor for worse functional status and overall survival. A surgical technique that includes wide cyst fenestration at the foramen of Monro, copious irrigation of cyst contents, resection of exophytic tumor, and inferior cyst wall fenestration ("through-and-through" technique) provides a statistically significant reduction in cyst size and mass effect in short-term follow-up. No complications occurred perioperatively and no sequelae resulted from the communication between the cyst interior and the ventricular system. No patients developed postoperative hypothalamic obesity. A statistically significant improvement of functional status was found at early postoperative follow-up compared to preoperative KPS scores.

Hypothalamic involvement was recently identified as the strongest predictor of worse quality of life, hypothalamic obesity, and decreased overall survival in a large cohort of childhood-onset craniopharyngiomas.[9,10,20] Complete tumor resection did not prolong progression-free survival or provide a survival benefit over patients with subtotal resections. Because mortality is most often the result of hypothalamic dysfunction and coexisting medical illnesses, gross-total resection did not confer a survival benefit in a large multicenter prospective cohort.[10] Due to limited treatment options for hypothalamic obesity, prevention of hypothalamic injury should be the preferred treatment strategy when managing retrochiasmatic craniopharyngiomas.[21] Recent evidence indicates that endoscopic endonasal approaches may reduce the amount of hypothalamic injury during resection of retrochiasmatic craniopharngiomas.[22-24] Similarly, ventriculoscopic surgery in our series did not result in increased BMI/hypothalamic obesity, and functional status improved over the short term. We developed the treatment algorithm shown in Figure 3, first to prevent exacerbation of hypothalamic dysfunction, and second, to provide optimal surgical management given the solid/cystic ratio and optic apparatus compression. The long-term radiographic and functional outcomes will be needed to determine if ventriculoscopic surgery is effective in managing cystic retrochiasmatic craniopharyngiomas.

Several series have been published evaluating the use of ventriculoscopic surgery in combination with postoperative

TABLE 2. Postoperative Clinical Course and Radiographic Outcome After Ventriculoscopic Surgery

Patient	ETV? completed?	Postop cyst size, largest dimension (mm)	Postop cyst size, volume (cm^3)	Postop KPS	Postop body-mass index	Postop radiation?	Short-term clinical status	Follow-up (yr)
Primary cystic group								
1	No	4.5	0.044	90	28.1	Yes	Endocrine function/resolution of amenorrhea; no visual deficit; small residual	2.7
2	Yes	13.1	1.34	80	47.6	No	Normal endocrine function; dramatic improvement in visual fields with mild right hemianopsia; craniotomy 2 yr after fenestration for solid recur.	6
3	Yes	17.3	2.6	70	32.7	Yes	Postradiation panhypopit, mild inferior temporal hemianopisa, no residual	2.3
4	No	17.1	2.3	90	35.4	Yes	Normal endocrine function; no visual deficits; No residual	1.5
5	Yes	14.7	1.87	80	35.5	No	Normal endocrine function; no visual deficits; small residual, no RT due to age	0.3
Mean		13 ± 5.2	1.6 ± 1.0	82 ± 8.4	35.9 ± 7.21			2.6 ± 2.1
Cystic recurrence group								
6	Yes	18	3.1	80	27.4	Yes	Stable panhypopit; visual fields normalized; residual with no recur.	.5
7	No	12.5	1.02	90	33.5	Yes	Stable hypopit; no visual deficits; resolved hydrocephalus, no residual	3
8	Yes	17.2	2.5	70	28.1	Yes	Postradiation hypopit; vision stable; repeat fenestration at 4 mo after visual decline with stable cyst size and visual function	2.3
9	Yes	11.3	0.77	90	37.8	Yes	Postradiation hypopit, small residual	4
10	Yes	20.9	4.2	70	26.8	No	Panhypopit; bitemporal hemianopsia; refused radiation, cystic/solid recur. at 10 mo and 18 mo, EEA for recurrence X 2	2
Mean		16 ± 4.0	2.3 ± 1.4	80 ± 1.8	30.7 ± 4.8			2.4 ± 1.3

EEA, endoscopic endonasal approach; hypopit, hypopituitarism; KPS, Karnofsky performance status; panhypopit, panhypopituitarism; postop, postoperative; RT, radiation therapy; recur, recurrence

radiation therapy. Park and colleagues[13] compared either subtotal surgical resection (14 patients) or ventriculoscopic surgery (13 patients) combined with Gamma knife radiosurgery. Over a mean follow-up time of 32 mo, they found a nonstatistically significant trend toward higher rates of recurrence with ventriculoscopic surgery compared to subtotal resection (53% vs 14%; $P = .183$). However, endocrine function was significantly better in the ventriculoscopic group and visual outcomes were similar. These results are in contrast to another investigation evaluating the use of ventriculoscopic surgery with fractionated stereotactic radiotherapy.[15] With a follow-up time of approximately 6 yr, tumor control was achieved in approximately 90% of patients with a statistically significant reduction in tumor volumes.

No previous large case series have investigated the use of combining superior and inferior cyst fenestration with third ventriculostomy. We adopted this technique to promote CSF circulation through the cyst interior both to maintain patency of fenestrations and promote continued egress of cyst contents.

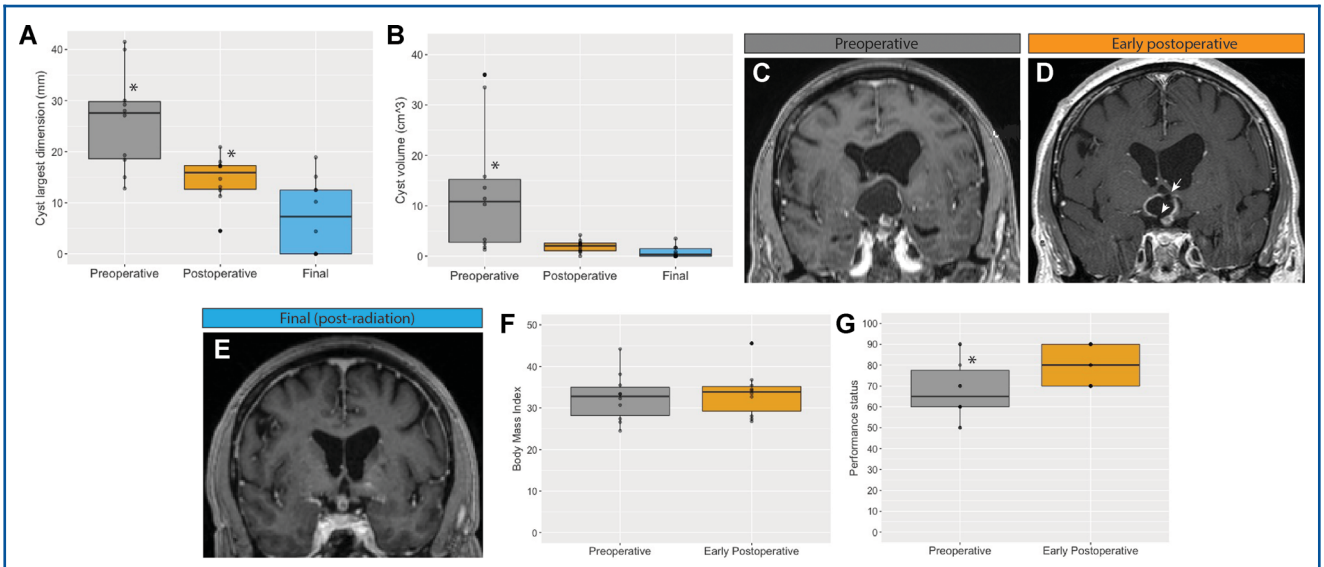

FIGURE 7. *Cyst size reduction and short-term clinical outcomes following ventriculoscopic surgery.* **A**, *Cyst largest dimension and* **B**, *cyst volume at preoperative, early postoperative, and final follow-up (including post-radiation) are shown in boxplots.* **C**-**E**, *Representative MRIs from patient 9 are provided to show radiographic cyst size reduction. The superior (long arrow) and inferior (short arrow) fenestrations are well visualized in postoperative imaging (***D***).* **F**, *BMI did not significantly increase following ventriculoscopic surgery.* **G**, *A statistically significant increase in KPS score was found at follow-up. Statistical significance is indicated by (*), and compares the mean value to the right-adjacent value.*

Initial concerns regarding potential chemical meningitis or CSF dissemination were not realized in our series and patients tolerated the procedure well. Larger studies will be needed to confirm whether the addition of inferior cyst fenestration/third ventriculostomy provides better tumor control than wide superior cyst fenestration at the foramen of Monro alone.

One major advantage of ventriculoscopic surgery over open subtotal resection techniques is the ability to perform multiple repeated fenestrations in the event of cyst reaccumulation. Patient 8 in our series had visual decline due to cyst recurrence that improved after repeat cyst fenestration. The use of repeated ventriculoscopic surgery in elderly patients with cystic third ventricular craniopharyngiomas for palliation has been described.[25] Our institution has adopted a multimodal management strategy that treats recurrent craniopharyngiomas as a chronic neurosurgical disease. A management strategy that embraces repeat ventriculoscopic surgery for recurrent cystic retrochiasmatic craniopharyngiomas may provide long-term tumor control and symptom management with minimal morbidity.

Limitations

The major limitation of this study is the small sample size due to the low incidence of cystic retrochiasmatic craniopharyngiomas amenable to ventriculoscopic surgery; therefore, efficacy of ventriculoscopic surgery cannot be judged against other treatment options such as minimally invasive transcranial approaches or endoscopic endonasal approaches. Craniopharyngiomas account for 1% to 3% of intracranial tumors, and retrochiasmatic tumors represent less than half of all tumors.[26] A matched cohort of cystic retrochiasmatic craniopharyngiomas treated with open resections for treatment comparison could not be identified at our institution. Our small sample size necessitates appropriate interpretation of our statistical analyses. We used nonparametric statistical tests to make the fewest assumptions about the study sample and to increase the threshold for identifying statistical significance.

CONCLUSION

Ventriculoscopic surgery, with or without postoperative radiation, is a minimally invasive surgical technique that can provide local control of retrochiasmatic craniopharyngiomas in the short term. Wide cyst fenestration at the foramen of Monro with inferior cyst fenestration/third ventriculostomy provided an immediate reduction in cyst size and relief of mass effect. We favor this technique for cystic retrochiasmatic craniopharyngiomas to avoid exacerbating hypothalamic dysfunction, the primary risk factor for worse functional status. A reduction in cyst size and improvement in functional status was found at early postoperative evaluation in patients who underwent ventriculoscopic surgery with or without postoperative radiotherapy. Larger investigations are needed to determine long-term efficacy and treatment durability.

Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

REFERENCES

1. Karavitaki N, Brufani C, Warner JT, et al. Craniopharyngiomas in children and adults: systematic analysis of 121 cases with long-term follow-up. *Clin Endocrinol*. 2005;62(4):397-409.
2. Kawamata T, Kubo O, Kamikawa S, Hori T. Ectopic clival craniopharyngioma. *Acta Neurochir*. 2002;144(11):1221-1224.
3. Hoffman HJ, Silva MD, Humphreys RP, Drake JM, Smith ML, Blaser SI. Aggressive surgical management of craniopharyngiomas in children. *J Neurosurg*. 1992;76(1):47-52.
4. Sweet WH. Radical surgical treatment of craniopharyngioma. *Clin Neurosurg*. 1976;23:52-79.
5. Yaşargil MG, Curcic M, Kis M, Siegenthaler G, Teddy PJ, Roth P. Total removal of craniopharyngiomas. *J Neurosurg*. 1990;73(1):3-11.
6. Van Effenterre R, Boch A. Craniopharyngioma in adults and children: a study of 122 surgical cases. *J. Neurosurg*. 2002;97(1):3-11.
7. Weiner HL, Wisoff JH, Rosenberg ME, et al. Craniopharyngiomas. *Neurosurgery*. 1994;35(6):1001-1011.
8. Puget S, Garnett M, Wray A, et al. Pediatric craniopharyngiomas: classification and treatment according to the degree of hypothalamic involvement. *J Neurosurg*. 2007;106(1 suppl):3-12.
9. Muller HL, Gebhardt U, Teske C, et al. Post-operative hypothalamic lesions and obesity in childhood craniopharyngioma: results of the multinational prospective trial KRANIOPHARYNGEOM 2000 after 3-year follow-up *Eur J Endocrinol*. 2011;165(1):17-24.
10. Sterkenburg AS, Hoffmann A, Gebhardt U, Warmuth-Metz M, Daubenbüchel AMM, Müller HL. Survival, hypothalamic obesity, and neuropsychological/psychosocial status after childhood-onset craniopharyngioma: newly reported long-term outcomes. *Neuro Oncol*. 2015;17(7):1029-1038.
11. Youmans JR, Winn HR. *Youmans Neurological Surgery*. 6th ed. Philadelphia, PA: Saunders/Elsevier; 2011; https://www.clinicalkey.com/dura/browse/bookChapter/3-s2.0-C20091594336.
12. Barajas MA, Ramirez-Guzman G, Rodriguez-Vazquez C. Multimodal management of craniopharyngiomas: neuroendoscopy, microsurgery, and radiosurgery. *J Neurosurg*. 2002;97(5 suppl):607-609.
13. Park YS, Chang JH, Park YG, Kim D. Recurrence rates after neuroendoscopic fenestration and Gamma Knife surgery in comparison with subtotal resection and Gamma Knife surgery for the treatment of cystic craniopharyngiomas. *J Neurosurg*. 2011;114(5):1360-1368.
14. Joki T, Oi S, Babapour B, et al. Neuroendoscopic placement of Ommaya reservoir into a cystic craniopharyngioma. *Child's Nerv Syst*. 2002;18(11):629-633.
15. Takano S, Akutsu H, Mizumoto M, Yamamoto T, Tsuboi K, Matsumura A. Neuroendoscopy followed by radiotherapy in cystic craniopharyngiomas—a long-term follow-up. *World Neurosurg*. 2015;84(5):1305-1315.e2.
16. Mori R, Joki T, Nonaka Y, Ikeuchi S, Abe T. Parallel insertion endoscopic technique for precise catheter placement in cystic craniopharyngiomas. *J Neurol Surg A Cent Eur Neurosurg*. 2014;75(6):442-446.
17. Gaab MR, Schroeder HWS. Neuroendoscopic approach to intraventricular lesions. *J Neurosurg*. 1998;88(3):496-505.
18. Mortini P, Gagliardi F, Bailo M, et al. Magnetic resonance imaging as predictor of functional outcome in craniopharyngiomas. *Endocrine*. 2016;51(1):148-162.
19. Van Gompel JJ, Nippoldt TB, Higgins DM, Meyer FB. Magnetic resonance imaging–graded hypothalamic compression in surgically treated adult craniopharyngiomas determining postoperative obesity. *Neurosurg Focus*. 2010;28(4):E3.
20. Muller HL, Faldum A, Etavard-Gorris N Functional capacity, obesity and hypothalamic involvement: cross-sectional study on 212 patients with childhood craniopharyngioma. *Klin Padiatr*. 2003;215(6):310-314.
21. Müller HL. Craniopharyngioma and hypothalamic injury. *Curr Opin Endocrinol Diabetes Obes*. 2016;23(1):81-89.
22. Yano S, Hide T, Shinojima N. Surgical outcomes of endoscopic endonasal skull base surgery of craniopharyngiomas evaluated according to the degree of hypothalamic extension. *World Neurosurg*. 2017;100:288-296.
23. Leng LZ, Greenfield JP, Souweidane MM, Anand VK, Schwartz TH. Endoscopic, Endonasal Resection of Craniopharyngiomas. *Neurosurgery*. 2012;70(1):110-124; discussion 123-114.
24. Cavallo LM, Frank G, Cappabianca P, et al. The endoscopic endonasal approach for the management of craniopharyngiomas: a series of 103 patients. *J Neurosurg*. 2014;121(1):100-113.
25. Sato K, Oka H, Utsuki S, Fujii K. Repeated neuroendoscopic palliative surgery in elderly patients with predominantly cystic craniopharyngioma in the third ventricle: three case reports. *Innov Neurosurg*. 2013;1(1):67-72.
26. Bunin GR, Surawicz TS, Witman PA, Preston-Martin S, Davis F, Bruner JM. The descriptive epidemiology of craniopharyngioma. *J Neurosurg*. 1998;89(4):547-551.

Acknowledgment

The authors thank Holly Wagner for her contribution in editing the manuscript.

COMMENTS

This small case series provides results of transventricular 'through and through' fenestration of 10 patients with retrochiasmatic craniopharyngioma cysts with hypothalamic involvement. This is used as part of multimodality treatment of craniopharyngiomas which are thought to be higher risk for hypothalamic injury.

This technique should be considered in the treatment of these challenging tumors but does have some aspects that need to be considered before widespread application.

Using this technique for primary tumors is based on the assumption of correct pathological diagnosis based on imaging alone, which can be difficult with craniopharyngioma. In addition, long-term outcomes are unknown.

The true test of the authors' proposed multimodality algorithm (cyst fenestration plus fractionated radiation) will come with time. Complete resection remains the gold standard for tumor control, but requires a balance between long-term tumor-free survival and morbidity. The technique they describe certainly appears to limit short-term morbidity, but this must be weighed against long-term morbidity that occurs with tumor recurrence and subsequent attempts at treatment which will be significantly complicated by prior treatments. Indeed, every patient requires additional treatment (only 1 did not thus far, but only has 4 months of follow-up and certainly will require it), making this technique only an adjuvant, temporary treatment for the cystic portion of these tumors.

Finally, radiosurgery should be considered for small tumor residuals to further decrease the risk to the hypothalamus.

Paul A. Gardner
Pittsburgh, Pennsylvania

The authors provide a nice description of short-term results for treating cystic craniopharyngiomas with intraventricular fenestration and radiation therapy. Indeed, cyst fenestration is certainly effective for decreasing mass effect on the chiasm and hypothalamus in predominantly cystic craniopharyngiomas and we have used this technique successfully in certain of our patients. However, it is important to bear in mind that the patients in this series who went on to receive radiation therapy mostly became hypopituitary and those that did not receive radiation therapy generally recurred and required a craniotomy or endonasal surgery. It has been our experience that a significant portion

of patients treated in this manner often recur after several years with progressive cyst enlargement and require additional surgery to remove the cystic and solid portions. The fact that they have already had a prior fenestration makes the tumor more difficult to remove and scarring occurs between the top portion of the tumor and the floor of the third ventricle and chiasm. For this reason, we prefer to offer adult patients complete resection and utilize radiation therapy for subtotally resected or recurrent tumors that are not amenable to curative re-operation. We encourage the authors to establish the long-term results of their treatment algorithm for further comparison to determine the true rate of long term recurrence.

Theodore H. Schwartz MD
New York, New York