



Outcomes After Endoscopic Endonasal Resection of Craniopharyngiomas in the Pediatric Population

Vishal S. Patel¹, Andrew Thamboo¹, Jennifer Quon², Jayakar V. Nayak¹, Peter H. Hwang¹, Michael Edwards², Zara M. Patel¹

■ **BACKGROUND:** Craniopharyngiomas have traditionally been treated via open transcranial approaches. More recently, endoscopic transsphenoidal approaches have been increasingly used; however, few case series exist in the pediatric population.

■ **METHODS:** A retrospective review of patients (aged <18 years) undergoing endoscopic transsphenoidal resection of craniopharyngiomas between 1995 and 2016 was performed. Preoperative data included presenting symptoms, tumor size, location, and components. Postoperative outcomes included symptom resolution, visual outcomes, endocrine outcomes, disease recurrence, and major complications.

■ **RESULTS:** Sixteen pediatric patients with mean age of 11.0 years (range, 5–15 years) were included. The median follow-up time was 56.2 months. Mean maximal tumor diameter was 3.98 cm. Most of the tumors had suprasellar (93.8%) and intrasellar (68.8%) components. The gross total resection rate was 93.8%. The most common presenting symptoms were vision changes (81.3%) and increased intracranial pressure (56.3%). Most patients (66.7%) had their presenting symptoms resolved by their first postoperative visit. Vision improved or remained normal in 69.2% of patients. Postoperatively, new incidence of panhypopituitarism or diabetes insipidus developed in 63.6% and 46.7% of patients, respectively. New hypothalamic obesity developed in 28.6% of patients. The postoperative cerebrospinal fluid leak rate was 18.8%. One patient died

of intraventricular hemorrhage postoperatively. The major complication rate was 12.5%. Disease recurrence occurred in 1 patient with gross total resection (6.3%).

■ **CONCLUSIONS:** Endoscopic transsphenoidal resection for craniopharyngiomas can achieve high rates of total resection with low rates of disease recurrence in larger tumors than previously described. However, hypothalamic-pituitary dysfunction and cerebrospinal fluid leak remain significant postoperative morbidities.

INTRODUCTION

Craniopharyngiomas are benign, often calcified, cystic tumors that originate from embryonic epithelial cells of the craniopharyngeal duct or from metaplasia of the pituitary stalk, with a reported incidence of 300–400 new cases per year in the United States.¹ More than a third of these tumors are within the pediatric population and account for roughly 10% of all pediatric intracranial tumors.^{1,2} Craniopharyngiomas can lead to intracranial compression, visual compromise, and hypothalamic-pituitary dysfunction, making prompt surgical intervention necessary. However, the proximity of these tumors to critical neural structures, including the pituitary gland, optic chiasm, third ventricle, hypothalamus, and major intracranial vessels, makes safe surgical resection inherently challenging, regardless of approach.

Key words

- Craniopharyngioma
- Endonasal
- Endoscopic
- Pediatric neurosurgery
- Skull base surgery

Abbreviations and Acronyms

- BMI:** Body mass index
- CSF:** Cerebrospinal fluid
- DI:** Diabetes insipidus
- eTSA:** Endoscopic transsphenoidal approaches
- GTR:** Gross total resection
- mTSA:** Microscopic transsphenoidal approaches
- OT:** Open transcranial

RT: Radiation therapy

STR: Subtotal resection

From the ¹Division of Rhinology, Department of Otolaryngology—Head and Neck Surgery and ²Division of Pediatric Neurosurgery, Department of Neurosurgery, Stanford University School of Medicine, Palo Alto, California, USA

To whom correspondence should be addressed: Zara M. Patel, M.D.

[E-mail: zmpatel@stanford.edu]

Citation: *World Neurosurg.* (2017) 108:6–14.

<http://dx.doi.org/10.1016/j.wneu.2017.08.058>

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

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There has been great debate over the surgical philosophy of craniopharyngiomas, with arguments supporting gross total resection (GTR) versus subtotal resection (STR) followed by adjuvant radiotherapy.³⁻¹³ Despite this disagreement, there has been an overall consensus that balance must be made between maximal tumor removal and limiting damage to surrounding structures.³ With this new paradigm of tumor management, there have been parallel advancements in surgical approaches that aim to maximize tumor resection and limit surgical morbidity. Traditionally, various open transcranial (OT) approaches have been used with variable surgical results and postoperative outcomes.¹⁴⁻²⁰ In addition, because of the location of these tumors, transcranial approaches generally require some degree of brain retraction, as well as manipulation of neurovascular structures, which has increased risk for perioperative morbidity. Over the last decade, endoscopic endonasal transsphenoidal routes have been established for the resection of craniopharyngiomas, which has allowed direct midline exposure, an increased field of visualization, and safe microsurgical dissection of the tumor from the undersurface of the optic chiasm and hypothalamus. Despite these benefits, some concerns regarding endoscopic approaches remain, including higher rates of postoperative cerebrospinal fluid (CSF) leak, comparatively poorer intraoperative hemorrhage control, and increased difficulty in physically reaching the tumor as the dissection proceeds more laterally. Because the benefits of endoscopic surgery have been weighed against these concerns, it has historically been saved for smaller tumors whereas larger tumors that are more difficult to access have been treated with an open approach.²¹

Although there have been many recent studies evaluating the outcomes of endoscopic, endonasal resection of craniopharyngiomas, most of these reports have looked at combined adult and pediatric populations.²²⁻³² Few studies have been reported solely on the endoscopic, endonasal resection of childhood craniopharyngiomas.³³⁻³⁸ Pediatric patients have smaller nostrils, narrower nasal cavities, and immature pneumatization of the sphenoid sinus, suggesting that they may have distinct surgical outcomes.³⁹

Here, we report a series of 16 pediatric patients with craniopharyngiomas who underwent resection via a pure endoscopic endonasal approach. Symptom resolution, visual improvement, endocrine dysfunction, rates of hypothalamic obesity, operative complications, and disease recurrence are presented.

METHODS

Approval was first obtained from the Stanford University institutional review board. STRIDE (Stanford Translational Research Integrated Database Environment), a standards-based informatics platform developed at Stanford that supports clinical and translational research, was used to collect a list of eligible patients.⁴⁰ We retrospectively reviewed the STRIDE database of all pediatric patients (<18 years of age) with craniopharyngiomas confirmed by pathology resected by an endoscopic, endonasal approach at Stanford Medical Center from 1995 to 2016.

Indication for Endoscopic Transsphenoidal Approach

Deciding between a transsphenoidal versus a transcranial approach is typically based on tumor location, size, shape,

invasion of nearby structures, and the degree of calcification, along with the presenting neurologic status of the patient. At our institution, transsphenoidal approaches are typically chosen for tumors that have an intrasellar or suprasellar but infrachiasmatic origin, whereas suprachiasmatic tumors extending into the third ventricle are more often approached transcranially. However, cases with predominantly suprasellar origins that extend into the third ventricle can be, and have been, approached transsphenoidally. For tumor sizes that require further lateral access or have predominant suprachiasmatic, intraventricular components, transcranial approaches are preferred. Specifically for pediatric patients, transsphenoidal surgery is easier in children 3 years and older because pneumatization of the sphenoid sinus has begun; however, conchal or presellar-type sphenoid sinus configurations are not considered contraindications for a transsphenoidal approach. The sizes of skull base and piriform aperture are also assessed for feasibility of transsphenoidal access in each individual case.

Inclusion and Exclusion Criteria

Inclusion criteria required 1) patient age <18 years at time of surgery, 2) diagnosis of craniopharyngioma based on pathology, 3) resection via a pure endoscopic endonasal transsphenoidal approach, 4) follow-up time of 3 months (an exception was applied for 1 patient who died), and 5) preoperative magnetic resonance imaging. Any patient who did not meet these criteria was excluded.

Preoperative Evaluation

Preoperative patient evaluation included 1) age, 2) gender, 3) total follow-up time, 4) previous neurologic surgeries, 5) presenting symptoms, 6) preoperative body mass index (BMI) for age—gender, 6) tumor size defined by maximum tumor diameter, 7) tumor components, and 8) tumor location.

Postoperative Evaluation

Postoperative evaluation included 1) extent of resection, confirmed by both intraoperative visual inspection and postoperative magnetic resonance imaging; 2) resolution of presenting symptoms, determined at the first postoperative office visit; 3) improvements in visual deficits (visual acuity or a visual field loss) measured by subjective symptoms, objective physical examination, or formal ophthalmologic testing within the first 3 postoperative months; 4) endocrine outcomes including hypopituitarism or diabetes insipidus (DI) based on hormone levels documented by laboratory results or from formal endocrine evaluation; 5) hypothalamic obesity based on postoperative BMI (calculated as weight in kilograms divided by the square of height in meters) for age—gender at last follow-up; 6) major complications, defined by visual compromise, neurovascular injury, or death; and 7) disease recurrence.

Surgical Procedure

At the initiation of the case, a lumbar drain was placed. Patients were then placed in pin fixation to allow for more precise neuronavigation with preoperative magnetic resonance imaging or computed tomography. The otolaryngology team began with initial exposure. The inferior, middle, and superior turbinates were first outfractured and a large nasoseptal flap was then

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