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FOCUS SESSION

Endoscopic endonasal skull base surgery for pediatric brain tumors

Yong Hwy Kim[1] · Ji Yeoun Lee[1,2,3] · Ji Hoon Phi[1,2] · Kyu-Chang Wang[1,2] · Seung-Ki Kim[1,2]

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Abstract

Purpose The utility of endoscopic endonasal skull base surgery (EES) in various pathologic entities in adults has been published in the literature. However, the role of EES in children has not been clearly elucidated. We evaluated the feasibility of EES in children with brain tumors.

Methods We retrospectively reviewed clinical features, surgical outcomes, and complications in children who underwent EES for intracranial and skull base tumors at a single institution from July 2010 to October 2018.

Results A total of 82 patients underwent EESs for 77 intracranial and 5 skull base bony tumors. The mean age at diagnosis was 11.4 years (range 4–18 years), and the mean follow-up period was 46.8 months. The most common tumors were craniopharyngioma in the intracranial tumor and chordoma in the skull base. Gross total resection was the goal of surgery in 55 patients and achieved in 90.9%. The vision was improved in 76.1% of patients with visual impairments. Preoperatively, various endocrinological deficiencies were revealed in 73.7% of 76 patients with hypothalamus-pituitary lesions, and the hyposomatotropism was most common. Endocrinological status was improved only in 10. Aseptic or bacterial meningitis (7.3%) was the most common surgical complication, and the cerebrospinal fluid leakage rate was 2.4%.

Conclusions EES provides favorable neurological outcomes with acceptable risk for children with brain tumors. The high incidence of endocrinological deficits in cases with hypothalamus-pituitary lesions emphasizes the importance of judicious pre- and postoperative evaluation.

Keywords Children · Brain tumor · Endoscopic endonasal surgery · Outcome · Complication

Introduction

Endoscopic endonasal skull base surgery (EES) has been regarded as an alternative surgical technique for the indicated intracranial and skull base pathologies in the adult population. A vast number of recent anatomical studies have permitted surgical access not only to midline structures such as cribriform, planum, tuberculum, sella, and clivus but also to parasellar and petrous bone through the endonasal route and have resulted in the expansion of the spectrum of EES to paramedian areas [1].

✉ Seung-Ki Kim
nsthomas@snu.ac.kr

[1] Department of Neurosurgery, Seoul National University Hospital, Seoul, Republic of Korea

[2] Division of Pediatric Neurosurgery, Department of Neurosurgery, Seoul National University Children's Hospital, College of Medicine, Seoul National University, Seoul, Republic of Korea

[3] Department of Anatomy, College of Medicine, Seoul National University, Seoul, Republic of Korea

EES has a unique advantage, providing a straightforward surgical corridor to the midline and paramedian skull base without extensive bony works and manipulations of critical neurovascular structures. In contrast to the vast literature providing results in adults, reports on EES in children are limited, potentially due to the difference in the incidence of sellar and parasellar tumors in children [2–6]. In addition, anatomical differences, such as a small skull base, narrow nasal cavity, and poorly pneumatized sphenoid sinus, are also challenging features for EES. However, the wide panoramic view under the diverse angled endoscopes could provide great value even in children. In this study, we retrospectively reviewed the indicated pathological entities, surgical outcomes, and complications and evaluated the feasibility of EES in children.

Patients and methods

This study was approved by the institutional review board of the author's institution (No. 1903-086-1017) and included patients who underwent EES for intracranial or skull base bony

tumors from July 2010 to October 2018. The clinical and pathological results were collected by review of the medical records. Our policy recommended EES to be performed in patients 6 years of age or older, considering the width of the nasal cavity and postoperative nasal care in poorly cooperative age groups, except younger cases with the comparable width of pyriform apertures to in 6 years old. A preoperative computed tomography scan was acquired in all patients to identify the individual anatomical variations of the nasal cavity and paranasal sinus. The degree of sphenoid sinus development did not affect the surgical approach decision: EES or the transcranial approach.

The goals of the surgeries were identified by the preoperative informed consents, and the degree of tumor removal was decided by the postoperative magnetic resonance (MR) images with gadolinium contrast acquired within 48 h after surgery. We defined gross total resection (GTR) as no evidence of any residual tumor capsule in the surgical field or enhancement on postoperative MR images. The follow-up schedule of MR images depended on the pathologic result. Indolent tumors such as pituitary adenomas were followed annually, and a 6-month interval follow-up was recommended until 2 years and then annually checked in tumors with a high propensity of recurrences such as craniopharyngiomas or chordomas.

Patients with lesions around optic nerves underwent neuro-ophthalmological evaluations including corrected and uncorrected visual acuity and Goldmann perimetry testing, before surgery and within 6 months afterward. The degree of visual disturbance was expressed by the visual impairment scale (VIS) [7].

The endocrinological evaluation was performed in patients with sellar and parasellar lesions. It included growth hormone, insulin-like growth factor-1, luteinizing hormone, follicle-stimulating hormone, estradiol or total testosterone, prolactin, free T4, thyroid-stimulating hormone, adrenocorticotropic hormone (ACTH), and cortisol levels from serum sampled in the early morning. The preoperative combined pituitary function test (CPFT) was not routinely performed. Instead, the rapid ACTH stimulation test was performed only in cases with clinical suspicion or low cortisol levels. The water deprivation test was also performed only in cases without definite abnormalities in serum electrolytes, urine electrolytes, or urine specific gravity, despite the presence of polyuria. Endocrinological deficiencies were corrected before surgery with appropriate hormone replacement, beginning following the basal hormone study in the first postoperative month. The CPFT or rapid ACTH stimulation test with basal hormone study was performed within 6 months after surgery, and the water deprivation test was recommended within 3 months in all cases with a history of diabetes insipidus before or after surgery. We analyzed the endocrinological results for the cohort compared with the final results, and any impairment in one of the normal pituitary axes was regarded as deteriorated pituitary function.

The surgical procedures were similar to our previous reports [8]. Exposure of the dura was dependent on the required corridors for each tumor. In tumors involving the pituitary stalk, we made a midline incision on the pituitary stalk after identification of normal structures. The remnant pituitary stalk or gland was preserved after tumor removal. The low-flow CSF leakage in the transsellar approach was reconstructed with multilayer fibrin sealant patch techniques. We used the nasoseptal flap with a multilayer technique in high-flow CSF leakage. We inserted the lumbar drainage only in those in whom the autologous fascia and fat were used for high-flow CSF leakage and removed within 7 days.

We examined the nasal cavity first on the third postoperative day to remove the packed material preventing epistasis and to evaluate the reconstructed skull base. Nasal examinations were scheduled weekly for 1 month and then in 3 months. Patients with any minor fever or meningeal irritation symptoms immediately underwent an endoscopic examination to identify the CSF leakage, and then a CSF examination was performed by lumbar puncture. We made a diagnosis of meningitis based on the CSF cell index with a cut-off value of 4, representing the ratio of white blood cells to red blood cells in the CSF divided by the ratio of white blood cells to red blood cells in the blood, to avoid bias from surgical procedures [9].

The clinical features of patients, tumor characteristics, surgical approach, and complications were descriptively analyzed. We used Fisher's exact test to compare the GTR and visual improvement. Wilcoxon's Rank-sum test was used to discriminate differences in changes in the VIS. Data were collected using Excel 2010 (Microsoft Corp.), and SPSS software (version 23.0, IBM) was used in the statistical analyses.

Results

Patients

Eighty-two patients underwent EESs for variant pathological entities. The mean age at the time of surgery was 11.4 years (range 4–18 years), and there was no significant difference in sexual distribution (male:female = 43:39). The mean follow-up period was 46.8 months (range 1–102 months). Twenty-one patients underwent 26 transcranial and 8 transsphenoidal surgeries before EES for 19 craniopharyngiomas and 2 pituitary adenomas. Nine of nineteen patients with recurrent craniopharyngiomas had undergone eleven stereotactic radiosurgeries before EES.

Presentations by pathologic entities (Table 1)

Eighty-two tumors were composed of 78 intracranial tumors and 4 skull base bony tumors. The visual disturbance was the

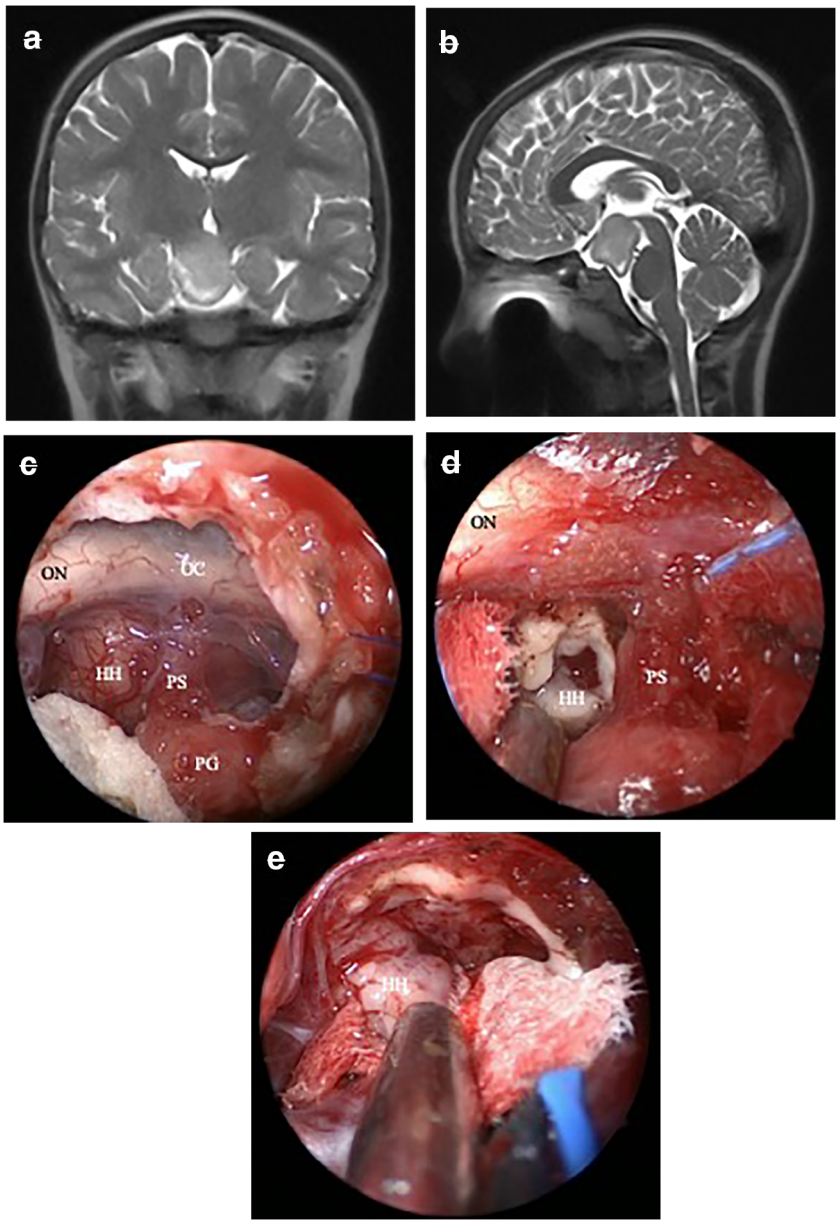

Fig. 1 Hypothalamic hamartoma. **a, b** The coronal (**a**) and sagittal (**b**) T2-weighted images showed a sessile mass of right hypothalamus with mixed high signal intensity. **c** Hypothalamic hamartoma (HH) with abundant cortical vessels was exposed, and the surgical corridor was defined by the optic nerves (ONs), chiasm (OC), pituitary stalk (PS), and pituitary gland (PG). **d, e** Partial removal was performed through a small cortisectomy

most common symptom in craniopharyngiomas, and all except one were retrochiasmatic. Thirteen pituitary adenomas were composed of eight nonfunctioning pituitary adenomas and five functioning tumors (3 Cushing's diseases and 2 acromegalies). All recurrent tumors were detected by routine follow-up. Rathke's cleft cysts presented with visual disturbance in all. Half of the germinomas presented with a mass effect such as visual disturbance or hydrocephalus and diabetes insipidus was the initial symptom in 20%. Amenorrhea and growth retardation were the most common symptomatic endocrinological deficiencies, and diabetes insipidus was presented only in germinoma and Rathke's cleft cyst. Hypothalamic hamartoma was presented with intractable gelastic seizure (Fig. 1). Optic pathway glioma showed rapid impaired bitemporal hemianopsia and growth retardation for several years (Fig. 2). All chordomas presented with diplopia and two Langerhans cell histiocytosis located at the right optic canal and left petrous apex, respectively.

Hydrocephalus was identified in 40% of 30 patients with headache and in 50% of 20 primary craniopharyngiomas.

Surgical corridors

Nasal turbinates were preserved in all cases, and posterior ethmoidectomy was performed in one to expose the lateral optic canal. Trans-tuberculum/planum approach was performed in 46 patients following trans-sella in 30, trans-clivus in 4 and a combined approach of trans-tuberculum/planum and trans-clivus with extradural resection of dorsum sellae in 2.

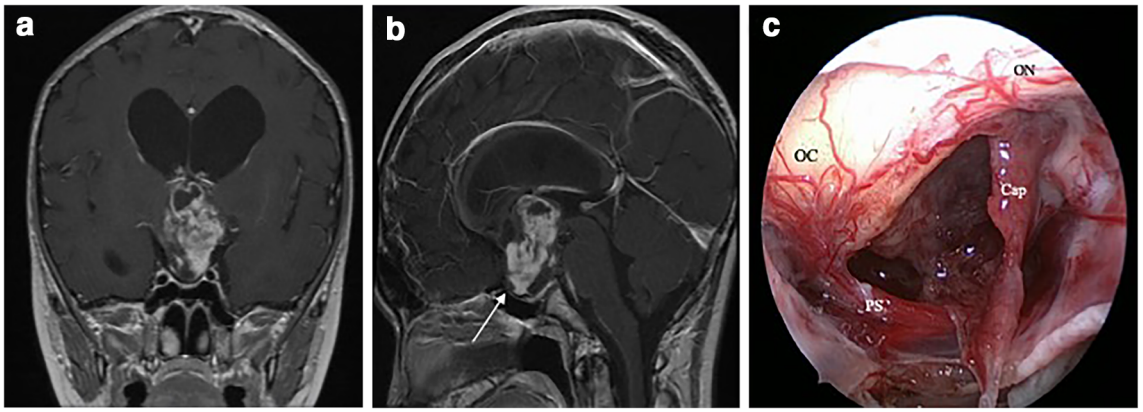

Fig. 2 Optic pathway glioma. **a, b** MR images from a 16-year-old boy who presented with panhypopituitarism and recent visual impairment. The pituitary gland was atrophic, and the optic chiasm (white arrow) was displaced anteriorly. **c** A photograph obtained during biopsy showed the displaced optic nerves, chiasm, and pituitary stalk by the tumor. The residual tumor capsule (Cap) was clearly demarcated from the surrounding neural tissues

Goal of surgery and tumor control (Table 2)

GTR was the primary goal of surgery in craniopharyngiomas, pituitary adenomas, and chordoma, and it was achieved in 92.3%, 84.6%, and 100%, respectively. The GTR rate of recurrent craniopharyngiomas was not significantly different from the primary ones (84.2% vs 100.0%, $p = 0.106$). The causes of subtotal resection (STR) were adhesion to the optic nerves or hypothalamus in craniopharyngiomas and invasion into the cavernous sinus in pituitary adenomas. Adjuvant radiosurgery was performed in 1 craniopharyngioma and 2 pituitary adenomas.

A hypothalamic hamartoma was resected partially for seizure control by disconnection. Biopsy with extradural decompression was performed in all cases of Langerhans cell histiocytosis in the bony skull base. The diagnosis of optic pathway glioma was made by open biopsy of the suprasellar mass with atypical features on preoperative imaging work-up.

Tumors recurred in 6 (15.4%) of 39 craniopharyngiomas and 1 each in Cushing's disease, Rathke's cleft cyst, and chordoma. The median time to recurrence of craniopharyngiomas was 19.0 months (range, 9.0–52.0 months). Four patients with recurrence underwent revision surgeries with 2 craniopharyngiomas, 1 Cushing's disease, and 1 Rathke's cleft cyst. The pathologic examination of a recurrent Rathke's cleft cyst revised from metaplasia of Rathke's cleft cyst to craniopharyngioma after the second surgery. Adjuvant radiosurgery was recommended in the other 4 recurrent craniopharyngiomas. One patient with recurrent chordoma refused the surgery and was lost in follow-up.

Neurological outcomes

The preoperative ophthalmic evaluation revealed visual impairment in 46 (58.2%) of 79 patients with parasellar tumors. The vision was improved in 35 (76.1%) and deteriorated in 1 (2.2%) after surgery. The mean VIS before and after surgery were 49.6. and 23.7, respectively. The vision improvement rates were similar between patients with the primary disease and recurrent ones (79.4% vs 66.7%, $p = 0.441$); however, the degree of VIS improvement in cases experiencing visual improvement was higher in primary rather than recurrent patients (mean 37.0 vs 19.2, $p = 0.034$). The visual field deterioration without visual acuity change occurred in a recurrent craniopharyngioma. The visual impairment by the Langerhans's histiocytosis recovered to normal vision.

Hydrocephalus by obstruction of the Foramen of Monro was resolved in all 12 cases without additional CSF diversion procedures. Diplopia in 2 craniopharyngiomas and 3 chordomas was recovered completely, and a patient with hypothalamic hamartoma who showed a seizure-free state from postoperative 8 months with monotherapy. No additional neurological impairments have occurred in the patients who underwent EES for biopsy. All patients except one with delayed intraventricular hemorrhage returned to their normal social life without assistance.

Endocrinological outcomes (Table 3)

Preoperative endocrinological evaluations in 76 patients with hypothalamus-pituitary tumors, including craniopharyngiomas, pituitary adenomas, Rathke's cleft cysts, germinoma, and hypothalamic hamartoma, revealed normal pituitary function in 22 (28.9%), partial deficiency in 23 (30.3%), and panhypopituitarism in 31 (40.8%) patients. The pituitary function was improved in 7 (9.2%) patients, including two with panhypopituitarism and one isolated ACTH deficiency. Hypocortisolism was the most common new-onset endocrinopathy. Endocrinological status at the last follow-up showed normal pituitary function in 13 (17.1%) and partial deficiency in 8 (10.5%) patients. Diabetes insipidus was highly prevalent in craniopharyngiomas, Rathke's cleft cyst, and germinoma.

Table 1 Pathologic entities and clinical presentations

Location	Intracranial					Skull base		Total ($n = 82$, %)
Pathologies	Craniopharyngioma ($n = 39$, %)	Pituitary adenoma ($n = 13$, %)	Rathke's cleft cyst ($n = 15$, %)	Germinoma ($n = 8$, %)	Others ($n = 2$, %)	Chordoma ($n = 3$, %)	Langerhans cell histiocytosis ($n = 2$, %)	
Sex (M:F)	23:16	6:7	7:8	2:6	2:0	2:1	1:1	43:39
Age (range)	11.0 (4–18)	14.0 (10–17)	10.9 (6–15)	11.9 (9–15)	11.0 (6–16)	7.7 (6–10)	10.5 (10–11)	11.4 (4–18)
History of recurrence	19 (48.7)	2 (15.4)	0	0	0	0	0	21 (25.6)
Neurologic symptoms								
Visual disturbance	25 (64.1)	6 (46.2)	9 (60.0)	3 (37.5)	1 (50.0)	1 (33.3)	1 (50.0)	46 (56.1)
Headache	14 (35.9)	3 (23.1)	8 (53.3)	2 (25.0)	0	1 (33.3)	2 (100.0)	30 (36.6)
Hydrocephalus	11 (28.2)	0	0	1 (12.5)	0	0	0	12 (14.6)
Nausea/Vomiting	7 (17.9)	1 (7.7)	3 (20.0)	0	0	0	0	11 (13.4)
Altered consciousness	3 (7.7)	0	0	0	0	0	0	3 (3.7)
Seizure	1 (2.6)	0	0	0	1 (50.0)	0	0	2 (2.4)
Diplopia	2 (5.1)	0	0	0	0	3 (100.0)	0	5 (6.1)
Syncope	1 (2.6)	0	0	1 (12.5)	0	0	0	2 (2.4)
Endocrinological symptoms[†]								
Amenorrhea	0	4 (30.8)	0	1 (12.5)	0	0	0	5 (6.1)
Growth retardation	2 (5.1)	2 (15.4)	3 (20.0)	3 (37.5)	1 (50.0)	0	0	11 (13.4)
Fatigue	1 (2.6)	0	2 (13.3)	2 (25.0)	0	0	0	5 (6.1)
Morphologic change	0	4 (30.8)	0	0	0	0	0	4 (4.9)
Body weight change	1 (2.6)	1 (7.7)	1 (6.7)	1 (12.5)	0	0	0	4 (4.9)
Polyuria/polydypsia	0	0	2 (13.3)	7 (87.5)	0	0	0	9 (11.0)
Galactorrhea	0	2 (15.4)	0	0	0	0	0	2 (2.4)
Incidental	0	0	1 (6.7)	1 (12.5)	0	0	0	2 (2.4)
Found during follow-up	19 (48.7)	2 (15.4)	0	0	0	0	0	21 (25.6)

[†] Only clinical symptoms at presentation excluding laboratory abnormalities and the alleged deficiencies controlled by medications

Table 2 Surgical outcomes by the goals of surgery

	Goal of surgery									Total ($n = 82$, %)
	Gross total resection					Partial resection with biopsy				
	Craniopharyngioma		Pituitary adenoma		Chordoma ($n = 3$, %)	Rathke's cleft cyst ($n = 15$, %)	Germinoma ($n = 8$, %)	Langerhans cell histiocytosis ($n = 2$, %)	Others ($n = 2$, %)	
	Primary ($n = 20$, %)	Recurrent ($n = 19$, %)	Primary ($n = 11$, %)	Recurrent ($n = 2$, %)						
Extent of tumor resection										
GTR	20 (100.0)	16 (84.2)	10 (90.9)	1 (50.0)	3 (100.0)	0	0	0	0	50 (61.0)
STR	0	3 (15.8)	1 (9.1)	1 (50.0)	0	0	0	0	1 (50.0)	6 (7.3)
Biopsy	0	0	0	0	0	0	8 (100.0)	2 (100.0)	1 (50.0)	11 (13.4)
Fenestration	0	0	0	0	0	15 (100.0)	0	0	0	15 (18.3)
Improved/pre-existing impairments										
Vision	11/13 (84.6)	8/12 (66.7)	3/6 (50.0)	0	1/1 (100.0)	9/9 (100.0)	2/3 (66.7)	1/1 (100.0)	0/1(0.0)	35/46 (76.1)
Diplopia	2/2 (100.0)	0	0	0	3/3 (100.0)	0	0	0	0	5/5 (100.0)
Hydrocephalus	10/10 (100.0)	1/1 (100.0)	0	0	0	0	1/1 (100.0)	0	0	12/12 (100.0)
Complications										
Meningitis	5 (25.0)	0	0	0	0	1 (0.0)	0	0	0	6 (7.3)
CSF leakage	0	0	0	0	0	2 (13.3)	0	0	0	2 (2.4)
Hemorrhage	1 (0.5)	0	0	0	0	0	0	0	0	1 (1.2)
Recurrence	4 (20.0)	2 (10.5)	1 (9.1)	0	1 (33.3)	1 (6.7)	0	0	0	9 (11.0)

Table 3 Endocrinological status in the hypothalamus-pituitary tumors before and after the surgery

		Craniopharyngioma		Pituitary adenoma		Rathke's cleft cyst ($n = 15$, %)	Germinoma ($n = 8$, %)	Hypothalamic hamartoma ($n = 1$, %)	Total ($n = 76$, %)
		Primary ($n = 20$, %)	Recurrent ($n = 19$, %)	Primary ($n = 11$, %)	Recurrent ($n = 2$, %)				
	Anterior hormones								
At presentation	Normal	3 (15.0)	2 (10.5)	7 (63.6)	1 (50.0)	6 (40.0)	3 (37.5)	0	22 (28.9)
	Partial deficiency	13 (65.0)	1 (5.3)	0	0	5 (33.3)	3 (37.5)	1 (100.0)	23 (30.2)
	Hypopituitarism	4 (20.0)	16 (84.2)	4 (36.4)	1 (50.0)	4 (26.7)	2 (25.0)	0	31 (40.8)
	Hypothyroidism	4 (20.0)	16 (84.2)	4 (36.4)	1 (50.0)	4 (26.7)	3 (37.5)	0	31 (40.8)
	Hypocortisolism	7 (35.0)	16 (84.2)	4 (36.4)	1 (50.0)	5 (33.3)	2 (25.0)	0	35 (46.1)
	Hyposomatotropism	17 (85.0)	17 (89.5)	4 (36.4)	1 (50.0)	8 (53.3)	5 (62.5)	1 (100.0)	53 (69.7)
	Hypogonadism	5 (25.0)	16 (84.2)	4 (36.4)	1 (50.0)	6 (40.0)	2 (25.0)	0	34 (44.7)
	Diabetes insipidus	4 (20.0)	16 (84.2)	0	0	2 (13.3)	7 (87.5)	0	29 (38.2)
Changes by operation	Improved	1 (5.0)	1 (5.3)	2 (18.2)	2 (100.0)	1 (6.7)	0	0	7 (9.2)
	Same	4 (20.9)	15 (78.9)	8 (72.2)	0	8 (53.3)	3 (37.5)	0	38 (50.0)
	Deteriorated	15 (75.0)	3 (15.8)	1 (9.1)	0	6 (40.0)	5 (62.5)	1 (100.0)	31 (40.8)
	Anterior hormones								
Final status	Normal	1 (5.0)	1 (5.3)	5 (45.5)	2 (100.0)	4 (26.7)	0	0	13 (17.1)
	Partial deficiency	1 (5.0)	0	1 (9.1)	0	4 (26.7)	2 (25.0)	0	8 (10.5)
	Hypopituitarism	18 (90.0)	18 (94.7)	5 (45.5)	0	7 (46.7)	7 (75.0)	0 (100.0)	55 (72.4)
	Hypothyroidism	18 (90.0)	18 (94.7)	5 (45.5)	0	7 (46.7)	6 (75.0)	1 (100.0)	55 (72.3)
	Hypocortisolism	18 (90.0)	18 (94.7)	6 (54.5)	0	10 (66.7)	7 (87.5)	1 (100.0)	60 (78.9)
	Hyposomatotropism	19 (95.0)	18 (94.7)	6 (54.5)	0	10 (66.7)	7 (87.5)	1 (100.0)	61 (80.3)
	Hypogonadism	18 (90.0)	18 (94.7)	6 (54.5)	0	11 (73.3)	8 (100.0)	1 (100.0)	62 (81.6)
	Diabetes insipidus	18 (90.0)	18 (94.7)	0	0	2 (13.3)	6 (75.0)	0	44 (57.9)

Complications

Two (2.4%) patients with Rathke's cleft cyst experienced CSF leakage, in whom the skull base was reconstructed with only a fat graft followed by the lumbar drainage despite the large arachnoid defect in the early period in our experience. The defects were successfully reconstructed with the pedicled nasoseptal flap. Postoperative meningitis occurred in 6 patients (7.3%) and was managed without sequelae. One delayed intraventricular hemorrhage occurred in postoperative 12 h after the successful removal of craniopharyngioma, causing the sudden hydrocephalus and severe sequelae despite emergent craniotomy.

Discussion

EES in children

EES is a less frequently used surgical method in children compared with adults, mainly because of the disease incidence [2, 4, 6, 10–14]. Our cohort of 39 craniopharyngiomas and 13 pituitary adenomas over 9 years reflected the epidemiological features of pediatric brain tumors and our strict surgical management protocol for incidental pituitary adenomas [15–17]. We usually considered EES as a surgical option in patients aged 6 years or older because the width of the pyriform apertures and cavernous internal carotid arteries are increased to 80% of the adult at 6 to 7 years of age [6, 18, 19]. Additionally, manipulation of the nasal septum and skull base after 5 or 6 years of age causes no developmental abnormalities on nasal and midface growth [20, 21].

Goals of surgery and tumor control

The goals of surgery depend on the preoperative diagnosis of each case and could be debatable. The GTR rate of EES in pediatric craniopharyngiomas ranges from 40 to 94% and is largely dependent not on the limitations of the surgical approach but on the surgeon's philosophy [22, 23]. Studies have advocated STR with adjuvant irradiation, based on the risks of neuro-endocrinological deficits, high recurrence rate despite GTR, and a comparable long-term control rate [22, 24–26]. The constant recurrence rate of approximately 20% after GTR or STR followed by irradiation, which is similar to our results, indicated to us that the treatment strategy for primary

craniopharyngioma should include the feasible management options for the recurrence in the future. A surgical resection is a unique option for most recurrent tumors after radiation therapy, especially in the adamantinomatous type, and the comparable outcomes of EES for recurrent craniopharyngiomas in this study and the literature support its value [27, 28]. The heavy burden on life evidenced by the history of the 33 surgical procedures and 11 radiosurgeries in 19 patients with recurrent tumors in this study, increased complication rate with repeated surgeries, surgical difficulties and risks caused by scarring, and deterioration of pituitary function despite the preservation of the stalk convinced us that GTR should remain the goal of craniopharyngioma surgery in cases with acceptable risk [8, 29].

Our strategy of Rathke's cleft cyst was similar to those in studies showing the reliable neuro-endocrinological improvement with acceptable recurrence rates by partial excision with content drainage [30–35]. We experienced a case of an asymptomatic small recurrence, but the regular follow-up was sufficient for the indolent natural course.

Our policy for surgical manipulation of optic pathway gliomas is strictly limited because of the indolent clinical course and the risk of visual function deterioration by surgery [36–38]. Open biopsy of the suprasellar mass was decided based on the atypical clinical and imaging features, and we limited the extent of the operation with the intraoperative pathologic confirmation. Partial resection of the hypothalamic hamartoma, type III by Regis' classification, was determined based on the mass size and adjacent optic nerves, although the benefits of radiosurgery in this type were documented [39].

Clinical outcomes

Restoration of vision after surgery for parasellar tumor depended on the several factors, including the duration of visual impairment, preoperative status, characteristics of the tumor, and degree of tumor removal [22, 23, 40, 41]. We speculated that the difference in the degree of visual improvement between primary and recurrent craniopharyngiomas was related to the different recovery abilities and pre-existing permanent damage before the aggravation by recurrent tumors [40]. There might be a bias in the objectivity and reliability of vision evaluation in poorly cooperative children. Therefore, the visual outcomes in this study, improvement in 76.1% and maintenance in 21.7% have worth not as absolute values but as indicators of the beneficial role of EES for parasellar tumors in children.

The preservation or improvement of pituitary function in craniopharyngioma ranged from 0 to 45%, and endocrinological deficits after surgery occurred in approximately 10–30% of the sellar lesion in children [2, 4, 22, 23, 28, 42–44]. Preoperative hypopituitarism was found in 26.7% and 25.9% of germinomas and Rathke's cleft cysts in this study, as compared with 86.5% and 50% in the large cohorts, respectively [42, 45]. And a higher deterioration rate was noticed in Rathke's cleft cyst and germinomas, although the final results for each pathologic entity were similar to the literature. We speculated that the limited indications for the preoperative CPFT in prepuberty could underestimate the partial deficiencies of hormones, in contrast to the in-depth postoperative evaluations in all susceptible patients to avoid the long-term insults caused by partial deficiencies [46, 47]. Additionally, we analyzed the final endocrinological status, which might be affected by adjuvant therapies, and pre-existing gonadotropin deficiency could be diagnosed with aging up to puberty during follow-up [45].

Complications

Two cases of CSF leakage in the early period in our experience changed the reconstruction strategy from tolerable risk to maximal safety and supported the freedom to use the pedicled nasoseptal flap. The most common complication was postoperative meningitis (7.3%), which seemed to be related to the stricter criteria for diagnosis than the other report, with a cut-off value of 4, for avoiding the risk of delayed diagnosis [48, 49]. Delayed intraventricular hemorrhage after the severe temper tantrum changed our protocol to lightly sedate the uncooperative patients.

Limitations

This retrospective study analyzed only 82 patients for 9 years at a single institution. A large proportion of recurrent diseases, especially craniopharyngiomas, reflected the features of the referral hospital, and bias due to the nature of the retrospective study was inevitable. Additionally, the long-time required to collect the cohort likely influenced the details of the surgical techniques and perioperative care, although the key strategies remained unchanged as the backbone. These limitations might be ineluctable without a prospective multicenter study, considering the low incidence of the diseases.

Conclusions

EES revealed favorable tumor control and neurological recovery with acceptable complications in various pediatric brain and skull base tumors. A judicious and careful postoperative management protocol is mandatory considering the different features in children. Meticulous evaluations and proper hormone replacement therapy are required for the high prevalence of endocrinological deficiencies.

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Compliance with ethical standards

This study was approved by the institutional review board of the author's institution (No. 1903-086-1017) and included patients who underwent EES for intracranial or skull base bony tumors from July 2010 to October 2018.

Conflict of interest All authors have no conflict of interest to declare.

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