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# Visual Outcome after Surgical Removal of Craniopharyngiomas

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**Objective :** In the present study, we evaluate visual outcomes after surgical removal of craniopharyngiomas and analyze the factors that are considered to affect visual outcomes.

**Methods :** This study includes 30 patients with craniopharyngioma, who underwent surgery in this clinic during the last 10 years. The changes of visual function (visual acuity and field) of the patients were assessed preoperatively and postoperatively, and paired data of this change were compared. Also, the factors that influence on this change were analysed.

**Results :** Among the 21 patients complaining of decreased vision before the operation, 8 patients were improved (38.0%), and 7 patients were worse (33.3%). However, Four out of 9 patients without any preoperative visual symptoms developed worse vision after the operation (44.4%). The average duration of symptom was 17.2 months in the improved group and 23.6 months in the aggravated group. The tumors recurred in only 5 patients after the gross total resection : Four of them showed the aggravation of visual function after surgical removal and 1 had improvement.

**Conclusion :** After removal of craniopharyngiomas, the overall rate of vision improvement, no change and aggravation are 26.6%, 36.7% and 36.7%, respectively. Aggravation of postoperative visual function is higher in males, children and patients with a longer duration of symptom and the tumor recurres more frequently in patients who complained of worsened vision after surgical removal.

**KEY WORDS :** Craniopharyngioma · Visual outcome · Preoperative · Surgical removal.

## Introduction

Craniopharyngioma, which makes up 3% of the primary brain tumors, occurs in the sellar and suprasellar area. This tumor is considered histologically benign, but clinically malignant[5]. It compresses the pituitary stalk, hypothalamus and optic chiasm, and the disease is usually presented with the characteristic features such as endocrinologic symptoms and visual dysfunction. The cranial nerves and major vessels are known to especially adhere to these tumors, so it is difficult to remove them completely and moreover, in cases where there are tumor remnants after surgery, recurrence is the rule. In recent studies, aggravation of the visual function reached nearly 50% postoperatively[13,16]. The major symptom is general headache associated with increased intracranial pressure in children, and decreased visual acuity or visual field defect and hypopituitarism in adults. Children usually do not complain visual symptoms until significant loss of visual acuity has occurred, so these tumors tend to be ignored, but adults usually complain of visual field defect and decreased visual acuity due to compression of the optic chiasm. The purposes of this study are to evaluate visual outcomes after surgical removal of craniopharyngiomas and also to analyse the relationship between postoperative visual function and the patient-related factors such as age, sex, symptom duration, the tumor-related factors such as tumor size, consistency, calcification, and the surgery-related factors such as surgical approach and extent of tumor resection.

## Materials and Methods

The subjects of this study were thirty patients who underwent surgery from March, 1988 to June, 2003 to remove pathologically confirmed craniopharyngiomas. Radiologic

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findings including the site, size, calcification and the consistency of the tumors and their clinicopathologic features such as sex, age, major symptom, visual acuity/field, and symptom duration were checked preoperatively. We also postoperatively assessed the surgical approach, the extent of removal, visual acuity and field, the presence of recurrence, and the pre- and postoperative data of visual function. Visual function was evaluated by visual acuity and visual field examination results. Visual acuity was measured numerically for both eyes. Visual field examination was done by ophthalmological consultation and cross-sectional areas of visual defect were compared before and after the surgery. In most cases, postoperative visual data were obtained at discharge and longterm follow-up of visual function was not available. The subjects were divided into two groups: adults over 18 and children under 18; they were also divided according to the improved group showing better postoperative visual acuity and field, and the worsened group showing worse postoperative visual acuity and field.

Statistical analyses were performed using SPSS version 11.0 for Windows, the cross tabulation report and Chi-square tests.

## Results

The 30 patients ranged in age from 4 to 62 years (mean age : 35.1), the sex ratio was 1.5:1 (male 18, female 12) and there were 23 adults and 7 children. The most common symptom was the decreased visual function (21 patients), and there was decreased visual function in 17 adult patients (74.0%) and headache in 6 children patients (86.0%). The average duration of symptom was about 17.3 months (from 10 days to 7 years). Twenty four cases of tumor were located in suprasellar area, 3 tumors were in the endosellar area with suprasellar extension, and 3 tumors were in the ventricles.

Gross morphologic findings of the tumors were as follows; 19 cystic, 5 solid, and 6 mixed types and 16 cases showed calcification. The average tumor size was 3.14cm (from 1.5 to 4.5cm). For the surgical approaches, the pterional approach was made in 13 cases, subfrontal in 8, interhemispheric in 4, transcallosal in 2, combined (transcallosal & pterional) in 2, and transsphenoidal in 1. For the degree of tumor removal, GTR (gross total resection) was made in 21 cases, NTR (near total resection) in 4, STR (subtotal resection) in 4, and partial removal in 1.

Among the 21 patients complaining of decreased vision before the operation, 8 patients had improved vision (38%), and 7 patients had worsened vision (33.3%). Four out of 9 patients without any preoperative visual symptoms developed worsened vision after the operation (44.4%). The mean rate of visual function aggravation was 36.7% (Table 1). By gender group, among the 18 male patients, 4 patients (22.2%) ended up with improved vision, but 8 (44.4%) of them had worsened vision. In contrast, 6 of 12 female patients had improved vision (50.0%), but 3 of them had worsened vision (25.0%). By age group, 8 of 23 adult patients had improved vision (34.8%) and other 8 of them had worsened vision (34.8%). Among the 7 children patients, 2 of them had improved vision (28.6%), and 3 of them had worsened vision (42.8%)(Table 2). The average duration of symptom was 7.3 months in the improved group, 13.3 months in no-change group, and 16.2 months in the worsened group. No significances were found on the basis of the pathologic features, the tumor size, the amount of removal, and the changes of vision(Table 3, 4, 5, 7). As to the operative approaches, the pterional approach was performed

**Table 1.** Visual outcome after surgical removal of craniopharyngioma

| Preoperative evaluation | Postoperative evaluation | no |
|---|---|---|
| Decreased visual function (n=21) | Improved | 8 |
| | No change | 6 |
| | Aggravated | 7 |
| Intact visual function (n=9) | No change | 5 |
| | Aggravated | 4 |

Overall Aggravation Rate : 36.7%

**Table 2.** Visual outcome according to patient-related factors

| | Improved | No change | Aggravated |
|---|---|---|---|
| Male (n=18) | 4 (22.2%) | 6 (33.3%) | 8 (44.5%) |
| Female (n=12) | 6 (50.0%) | 3 (25.0%) | 3 (25.0%) |
| Adults (n=23) | 8 (34.8%) | 7 (30.4%) | 8 (34.8%) |
| Children (n= 7) | 2 (28.6%) | 2 (28.6%) | 3 (42.8%) |

**Table 3.** Visual outcome according to tumor consistency

| | Consistency | Improved | No change | Aggravated |
|---|---|---|---|---|
| Decreased VF (n=21) | Cystic (15) | 6 (40.0%) | 3 (20.0%) | 6 (40.0%) |
| | Solid (3) | 1 | 1 | 1 |
| | Mixed (3) | 1 | 2 | – |
| Intact VF (n=9) | Cystic (4) | – | 2 | 2 |
| | Solid (2) | – | 2 | – |
| | Mixed (3) | – | 1 | 2 |

VF : Visual function

**Table 4.** Visual outcome according to calcification

| | Calcification | Improved | No change | Aggravated |
|---|---|---|---|---|
| Decreased VF (n=21) | (+) (10) | 4 (40.0%) | 2 (20.0%) | 4 (40.0%) |
| | (–) (11) | 4 (36.4%) | 4 (36.4%) | 3 (27.2%) |
| Intact VF (n=9) | (+) ( 6) | – | 3 | 3 |
| | (–) ( 3) | – | 2 | 1 |

VF : Visual function

**Table 5.** Visual outcome according to tumor size

| | Tumor size | Improved | No change | Aggravated |
|---|---|---|---|---|
| Decreased VF (n=21) | < 3cm ( 7) | 3 (42.8%) | 2 (28.6%) | 2 (28.6%) |
| | ≥ 3cm (14) | 5 (35.7%) | 4 (28.6%) | 5 (35.7%) |
| Intact VF (n=9) | < 3cm ( 5) | – | 3 | 2 |
| | ≥ 3cm ( 4) | – | 2 | 2 |

VF : Visual function

**Table 6.** Visual outcome according to the extent of surgical removal

| | Extent of removal | Improved | No change | Aggravated |
|---|---|---|---|---|
| Decreased VF (n=21) | GTR (14) | 5 | 5 | 4 |
| | NTR (2) | 1 | – | 1 |
| | STR (4) | 1 | 1 | 2 |
| | PR (1) | 1 | – | – |
| Intact VF (n=9) | GTR (7) | – | 3 | 4 |
| | NTR (2) | – | 2 | – |

VF : Visual function, GTR : gross total resection, NTR : near total resection, STR : subtotal resection, PR : partial resection

**Table 7.** Visual outcome according to surgical approach

| | Surgical approach | Improved | No change | Aggravated |
|---|---|---|---|---|
| Decreased VF (n=21) | Pterional (11) | 5 | 3 | 3 |
| | Subfrontal (4) | 1 | 1 | 2 |
| | Interhemispheric (4) | 2 | 1 | 1 |
| | Transcallosal (1) | – | 1 | – |
| | Combined (1) | – | – | 1 |
| No change (n=9) | Pterional (2) | – | 1 | 1 |
| | Subfrontal (4) | – | 1 | 3 |
| | Transcallosal (1) | – | 1 | – |
| | Combined (1) | – | 1 | – |
| | TSA (1) | – | 1 | – |

TSA : Transsphenoidal approach, VF : Visual function

in 13 patients and 4 showed aggravation of visual function postoperatively. On the other hand, the subfrontal approach was performed in 8 patients and 5 showed aggravation of postoperative visual function. Lacking of statistical significance, the pterional approach provided more favorable visual outcome than the subfrontal approach (Table 6). Among the 21 gross total resection group, only 5 patients showed recurrences : Four of them belonged to the worsened group (11 patients) for postoperative visual function and only one patient belonged to the improved group (10).

## Discussion

Craniopharyngioma was initially named by Frazier and Alpers in 1931, and by Cushing in 1932[9,21]. Craniopharyngioma is noted as a non-gliomatous brain tumor making up 3% of the primary brain tumors and it is a benign tumor histologically[5,11,12]. However, this tumor is usually located near the hypothalamus, optic pathway and pituitary gland, and it tends to invade locally into the surrounding tissue. Moreover, after removal, its recurrence rate is so high that it is hard to cure clinically. The origin cell of craniopharyngioma is thought to be a non-disappearing remnant cell of the hypophyseal-pharyngeal duct that exists temporarily in embryonal period. About half of the patients are children and craniopharyngioma is the third most common intracranial non-glial, non-neural cell tumor in children[5].

The first sign of craniopharyngioma is usually progressive visual field defect and unless the tumor is removed, it will result in permanent visual loss. The prognosis for the visual function is very important after removal. However, most of the studies were not focused on visual function; but rather, they were focused on the neurologic and endocrinologic prognosis that can occur due to the difficulties of tumor removal.

Konig et al. compared the transsphenoidal approach with the transcranial approach for the postoperative visual function[14]. The study showed a much greater permanent visual loss after a transcranial operation, but Konig did not evaluate preoperative visual function. Baskin and Wilson reported on tumors of 74 patients that were removed by the transcranial approach (61%) and by the transsphenoidal approach (39%) : in that study, 73% of patients complained of preoperative visual dysfunctions and 93% (48 of 53) of them showed postoperative improvement[3].

Bruce et al. analysed the visual function for 25 children patients with craniopharyngioma. Five of them were bilaterally blind in preoperative period and twenty of them showed various degrees of preoperative visual acuity decrements. Visual function was improved in 10 out of 20 children, and the others did not show significant change and none had aggravated postoperative vision. Bruce reported that visual function was improved or stabilized in the patients who had operations for craniopharyngiomas[4].

Repka et al. analyzed the change of the preoperative and postoperative visual function in thirty patients (12 children, 18 adults) confirmed as craniopharyngiomas[16]. The pre- and postoperative visual function was normal in two children and one adult, and the postoperative visual function was improved in five adults.

This study showed the visual function was worse in children than in adults, and this might be due to delayed recognition of the children's visual dysfunctions. Moreover, postoperative visual function was scarcely improved and Repka concluded by stating that the children had more severe visual dysfunctions and Chen et al. reported the visual function disorder might result from the compression on the visual apparatus and the prognosis was worse in children than in adults because of delayed diagnosis[6]. Fisher et al. categorized the cases with visual dysfunctions under 6 years as a poor prognostic factor[8]. Bang et al. analysed long-term results after surgical treatment of craniopharyngiomas in 100 adult patients and they concluded that visual disturbance could be improved by early diagnosis and surgical decompression[2]. In this study children revealed more worsened vision postoperatively than adults probably due to delayed diagnosis associated with children's inability to express their visual symptoms.

Since craniopharyngioma compresses the optic nerve fibers from below at the chiasm, the patients usually present with

bitemporal hemianopsia or more commonly inferior temporal quadrantanopsia[4,10,20,22]. John et al. analyzed the visual function in 121 patients and the results were as follows : 22.5% had homonymous hemianopsia : 21.6% had bitemporal hemianopsia : 8.4% had blindness in one eye : 7.5% had quadrantanopsia : and 2.5% had blindness in both eyes[13]. On the visual acuity test, 24.3% were aggravated, 52.5% showed no change and 23.4% were improved, so the results were similar to the results of this study. In the study of Effenterre and Laure, the pre- and postoperative visual function was analysed, and 70% of 122 patients were improved, 15% were aggravated and 15% showed no change[7].

In the adamantinomatous type of craniopharyngioma, common in children, the squamous cells are packed into the endothelial cells and they form themselves into the keratin nodules. When the outer surface of the basal cells with abundant keratin makes contact with the pial membrane, then calcification occurs[1]. Calcification around the tumor makes complete surgical removal impossible due to the difficult separation of the tumor from the surrounding tissue containing the optic nerve and chiasm. In this study, over half of the children patients (57.1%) showed calcifications around their tumors and 71.4% of the tumors had cystic components. Pierre et al. reported that, in the cases of craniopharyngiomas with large cysts in children, aspiration of the cyst prior to removal made the prognosis for visual function better and also that the worse the preoperative visual function, the higher the severity of the visual function aggravation[16]. Our current study showed more aggravation of visual symptoms in the group having no change of their preoperative visual function.

Craniopharyngioma shows various visual dysfunctions that are in contrast to pituitary adenoma and suprasellar meningioma[15,17,19,22]. The prognosis for the postoperative visual function of craniopharyngioma is not good, and immediate and steady improvement in visual acuity and field is not usually noted. Because craniopharyngioma adheres to the optic pathway and located in the posterior of the optic chiasm, complete removal is difficult to achieve. Although meningioma is an adhesional lesion generally, it can be removed safely through the transcranial approach due to its superior location on the optic pathway. Also, adhesion is not severe for pituitary adenoma and it can be removed adequately by the transsphenoidal approach. Therefore, Repka et al. recommended to give warning to the patients with preoperative severe visual dysfunction in craniopharyngiomas about the possibility of permanent visual loss[18].

In current study, tumor recurrences were observed more often in patients with worsened vision postoperatively. This presumed that the craniopharyngioma with more invasive nature to the optic apparatus could not be resected completely.

## Conclusion

After removal of craniopharyngiomas, the overall rates of vision improvement, no change and aggravation were 26.6%, 36.7% and 36.7%, respectively. Worsened vision was observed more in males and children but there was no statistical significance. Aggravation of vision was higher with a longer duration of symptom, and tumor recurred more frequently in patients who complained of worsened vision after surgical removal.

The tumor size, consistency, calcification, surgical approach and extent of removal did not have significant influence on the postoperative visual function. Therefore, It seemed that the aggravation of visual function was not associated with tumor-related factors or surgery-related factors but might be associated with patient-related factors although it was difficult to find statistical significances due to small numbers of total subject patients.

Finally, the long-term follow up of postoperative visual function is necessary for more exact defining the factors influencing on the visual outcome after surgical removal of craniopharyngiomas.

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## Commentary

The authors analyzed in this article the visual outcome after the surgery of craniopharyngioma. It has been generally accepted that the worse the preoperative visual function, the higher the severity of the visual function aggravation after the surgery. So to many neurosurgeons, it is worrisome the possibility of the visual loss after the surgery in case of craniopharyngioma with preoperative poor visual function. The authors experienced that about one third of craniopharyngioma patients had postoperative aggravation of visual function and they also suggested that the aggravation of postoperative visual function was higher in males, children and patients with a longer duration of symptom. These conclusions are quite understandable. Because the radical surgery to the craniopharyngioma with above-mentioned circumstances could give the chance of visual loss, flexible and individualized surgical decision should be applied to improve the quality of treatment for craniopharyngioma.

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