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## POSTOPERATIVE VISUAL RECOVERY FOLLOWING SURGICAL TREATMENT OF CRANIOPHARYGIOMAS

*VIDNA OŠTRINA NAKON OPERATIVNOG LEČENJA KRANIOFARINGEOMA*

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### Summary

**Introduction.** Craniopharyngiomas are rare tumors which are typically focused in the sellar and suprasellar region. Secondary to mass effect, these tumors commonly mediate neurologic, endocrinologic or visual functions. The purpose of this study was to investigate the pre and postoperative visual acuity in patients with a craniopharyngioma in the area of the optic chiasm. **Material and Methods.** This retrospective study included 42 patients with a craniopharyngioma demonstrated by computerized tomography or magnetic resonance imaging. The visual status was analyzed both before and after surgery (10 days, one month and six months after surgery).

**Results.** Progressive loss of visual acuity was a typical initial eye symptom. Postoperatively, improvement in visual acuity was seen in 47.2% of eyes. Normal vision was attained in the majority of eyes (from 27% preoperatively to 40% six months after surgery). The percentage of eyes with heavily reduced visual acuity decreased as well (from 38% preoperatively to 11% six months after surgery). Visual acuity improved at least in one eye in 36.58% of patients, and 28% of patients achieved normal visual acuity in both eyes, six months after surgery. The improvement of 0.5 and better at least in one eye was observed in 33% of patients. The majority of eyes showed immediate improvement after surgical decompression, during first ten postoperative days. **Conclusions.** The majority of patients with craniopharyngioma show a significant improvement of visual function, particularly in the first ten postoperative days.

**Key words:** Craniopharyngioma; Recovery of Function; Visual Acuity; Vision Disorders; Treatment Outcome; Signs and Symptoms; Magnetic Resonance Imaging; Tomography, X-Ray Computed

### Introduction

Craniopharyngiomas are histologically benign, dysembryogenic tumors, originating from embryonic epithelial remnants of invagination of primary oral cavity – craniopharyngeal canal and Rathke's pouch. They represent 2.5 – 4% of all intracranial tumors, accounting for one third of all supratentorial and one half of all suprasellar tumors in children [1, 2].

Up to a half of these tumors occur in childhood and during adolescence by the age of 20. However, in cases with slow growth, their discovery is often postponed up to the fourth and fifth decade of life, making craniopharyngiomas part of adult pathology.

### Sažetak

**Uvod.** Kraniofaringeomi su retki tumori prvenstveno lokalizovani u selarnom i supraselarnom prostoru. Zbog svog mas efekta ovi tumori dovode do neuroloških i endokrinoloških poremećaja, kao i oštećenja vidne funkcije. Cilj ovog rada je da ispita stanje vidne oštine pre i posle operacije kraniofaringeoma lokalizovanih u zoni optičke hijazme. **Materijal i metode.** Retrospektivno je analizirano 42 pacijenta sa kraniofaringeomom koji je utvrđen pregledom kompjuterizovanom tomografijom ili magnetnom rezonancijom. Pacijenti su podvrgnuti detaljnom oftalmološkom pregledu preoperativno i postoperativno (pri otpustu – 10 dana posle operacije, posle mesec dana i posle šest meseci). **Rezultati.** Progresivni gubitak vidne oštine bio je tipični inicijalni simptom. Pобољшanje vidne oštine uočeno je u nešto manje od polovine ispitivanih očiju – 47,2%, sa najizraženijim promenama u smislu povećanja broja očiju sa normalizovanom vidnom oštrinom – od 27% preoperativno do 40% posle 6 meseci i smanjenjem broja očiju sa teško redukovanom vidnom oštrinom od 38% očiju preoperativno do 11% 6 meseci postoperativno. U odnosu na broj pacijenata – 36,58% pacijenata pokazuje poboljšanje vidne oštine (bar na jednom oku), 28% postiže za 6 meseci normalnu vidnu oštrinu na oba oka, dok 33% pacijenata postiže bar na jednom oku vidnu oštrinu od 0,5 i veću. Većina očiju pokazuje brz oporavak vidne oštine, nakon hirurške dekompresije tokom prvih deset dana posle operacije. **Zaključak.** Većina pacijenata sa kraniofaringeomom pokazuje signifikantno poboljšanje vidne oštine, naročito u prvih 10 dana nakon operacije.

**Ključne reči:** Kraniofaringeom; Oporavak funkcije; Vidna oštrina; Poremećaji vida; Ishod lečenja; Znaci i simptomi; MRI; CT

Craniopharyngiomas are often characterized by extensive growth, reaching gigantic proportions, and predisposition to recur. Their localization can be intrasellar, suprasellar or parasellar and basically anywhere alongside the craniopharyngeal canal [1, 2]. Macroscopically, they are usually presented by big solitary mass composed of solid and cystic parts. Walls of cysts commonly contain calcifications.

The first detailed description of cystic suprasellar craniopharyngioma was made in 1857 by Zenker. More than 40 years later, in 1899, Mott and Barrett hypothesized that tumor originated from the cells of embryonic epithelial remnants of craniopharyngeal

**Abbreviations**

CT	– computed tomography
MRI	– magnetic resonance imaging
VA	– visual acuity
NLP	– no light perception
CF	– counting fingers
LP	– light perception
HM	– hand movement

canal, while Lewis in 1910 published the first attempt of its surgical removal.

Hypopituitarism, visual function impairment, vegetative and psychological disturbances (thermoregulation, hunger, sleep, affect and memory disorders) and increase of intracranial pressure occur as the consequence of the compression of pituitary gland and its stalk, optical pathways, hypothalamus and third ventricle.

Clinical presentation, tendency to grow, predisposition to recur, possibility of its complete removal, and the prognosis differ significantly in various age groups. In childhood, craniopharyngiomas are more prone to show expansive, aggressive growth, recidivate more often, and cause more severe endocrine disorders [1–4]. In adults, as a consequence of a long term irritation due to the leakage from cystic parts of tumor, numerous fibrous adhesions with surrounding structures are formed, making radical surgical removal much harder [1, 2, 4–8].

The decrease of mortality rate itself is not the only measure of the successful treatment. Today, the aim is to preserve and recover any neurological, psycho-emotional and hormonal, as well as visual functions, leading to longer lifetime expectancy and improved quality of life of patients.

The goal of this research was to determine the degree of recovery of visual acuity after surgical treatment of craniopharyngioma by decompression of optochiasmal region.

**Material and Methods**

Forty two patients with optic chiasmal craniopharyngioma, confirmed by computerized tomography (CT) and/or magnetic resonance imaging (MRI)

scan, were included in the study. All of them were operated between 1999 and 2004 at the Institute for Neurosurgery, Clinical Centre of Serbia, Belgrade and the Department of Neurosurgery, Clinical Centre of Vojvodina, Novi Sad. Based on patient's history and neuro-ophthalmological examination, 37 patients (88%) with positive ophthalmological findings and with no prior disease of the eye or optic nerve were selected. Of those, 31 patients (83.7%) were operated by trans-sphenoidal surgical approach, and 6 (16.2%) by transcranial surgical approach. Complete tumor resection was achieved in 29 patients (78.4%), while surgical therapy was incomplete in 8 (21.6%).

Detailed ophthalmologic examination included visual acuity, color vision, visual field, pupillary reaction to light, oculomotor nerves functions, as well as measurement of eye bulbs protrusion and fundus inspection. The examination was done preoperatively and postoperatively, on the day of patient's discharge from hospital (10<sup>th</sup> day postoperatively), after one month and after six months.

Visual acuity (VA) was determined using Snellen optotype, from the distance of 6 meters, in the light of constant brightness and with best refractive correction. Based on VA, the patients were classified into five groups:

1. with no visual acuity decrease BCVA=1.0;
2. with mild decrease BCVA=0.9 – 0.5;
3. with moderate decrease BCVA=0.4 – 0.1;
4. with severe decrease BCVA= counting fingers (CF), hand movement (HM), light perception (LP);
5. blind patients BCVA = no light perception (NLP).

Null hypothesis that there is no change in visual acuity before and after surgery was tested using Student t-test.

**Results**

Out of 42 patients with diagnosed craniopharyngioma, 37 had positive ophthalmologic findings. The study sample consisted of 18 males (48.64%) and 19 females (51.35%) according to demographic data, whose average age was 30.4 years (12 – 67 yrs.). There were three children up to the age of 14 (8.1%),

**Table 1.** Proportion of neuroophthalmological disturbances  
**Tabela 1.** Zastupljenost neurooftalmoloških poremećaja

Disturbance/Poremećaj	No of patients/Broj pacijenata	%
Decreased visual acuity/Pad vidne oštrine	28	75.6%
Impaired color vision/Pad kolornog vida	28	75.6%
Visual field defects/Ispadi u vidnom polju	31	83.7%
PNO atrophy/Atrofija PNO	25	67.5%
RAPD	8	21%
Papilloedema/Papiledem	6	16%
Oculomotor palsies/Okulomotorne pareze	4	10.8%
Proptosis/Proptoza	0	0

*PNO - papilla nervi optici, RAPD - relativni aferentni pupilarni defekt*

**Table 2.** Visual acuity at different time intervals - eyes**Tabela 2.** Vidna oštrina u različitim vremenskim intervalima - oči

	10 days after surgery <i>10 dana posle operacije</i>		1 month after surgery <i>1 mesec posle operacije</i>		6 months after surgery <i>6 meseci posle operacije</i>	
Unchanged/ <i>Nepromenjena</i>	43	59.7%	34	47.2%	28	38.9%
Improved up to 2 lines <i>Poboljšana do 2 linije</i>	15	20.8%	13	18%	11	15.3%
Improved more than 2 lines <i>Poboljšana preko 2</i>	5	6.9%	15	20.8%	23	31.9%
Decreased/ <i>Pogoršana</i>	9	12.5%	10	13.9%	10	13.9%
Total/ <i>Ukupno</i>	74	100%	72	100%	72	100%

26 patients were between 15 and 40 years of age (70.27%), and 8 patients were over 40 years of age (21.62%). More than half of the patients (56.7%) were in their first three decades of life. Three patients (8.1%) were over 60 years of age.

Out of all signs and symptoms in the clinical presentation of craniopharyngioma, a decrease in visual acuity (31 patients/83.7%) and endocrine disorders (13 patients/35.1%) were the most common ones. Headache was present in 12 patients (32.4%) as an isolated finding and in combination with nausea and vomiting, as a symptom of raised intracranial pressure, it was observed in 6 patients (16.2%). Neurological disorders leading to motor and mental deficit (muscular weakness, memory loss, disorientation, depression), as well as paresis of external eye muscles were observed in 4 patients (10.8%).

There was an interval from 6 weeks to 9 years (24.4 months on average) from the onset of symptoms to the confirmation of diagnosis. In 16 cases (43.2%) the diagnose was established within 6 months from the beginning of symptoms, in 8 cases (21.6%) during the interval from 7 months to 2 years, and in 13 cases (35.1%) it took more than 2 years to confirm the true diagnosis.

All the patients with pronounced neuro-ophthalmologic manifestations had tumor of suprasellar localization. Intraseellar localization with suprasellar propagation was found in 12 patients (32.4%). Suprasellar tumors of extraventricular localization were present in 19 patients (51.3%), four of which with parasellar and five with retrosellar spreading. Intraventricular location of tumor was observed in 6 patients (16.2%).

The size of tumor, determined by its greatest diameter, was measured using MRI or CT scanning techniques.

Twenty-six patients had complete data on the size of tumor. Most of them were tumors of medium size, between 20 and 40 mm, found in 15 patients (57.6%), followed by small-sized tumors up to the 20 mm in diameter, found in 7 patients (26.9%), while large tumors, bigger than 40 mm were present in 4 patients (15.3%).

The most common neuro-ophthalmological disorders were visual field scotoma, decrease in VA, optic nerve head atrophy, color vision disturbances and relative afferent pupillary defect (RAPD), while

papilledema and oculomotor paresis were seen much less frequently (**Table 1**).

Out of the total number of 74 eyes involved, the normal preoperative VA was found in 20 eyes (27.02%); mild and moderate reduction was found in 24 eyes (32.4%), while 28 eyes (37.8%) had severely decreased VA, including CF, HM and LP. Two eyes (2.7%) were totally blind, with NLP.

Normal binocular VA was found in 8 patients (21.6%). Monocular decrease of VA was observed in only 4 patients (10.8%) and binocular in the remaining 25 patients (67.5%). Severe reduction of VA, at least in one eye, was found in more than half of the patients (23 patients, 62.1%).

Thirty-six patients have completed the study, since one patient, aged 52, with tumor size of 45 mm passed away in the first few days postoperatively. He had had symptoms for 6 years prior to surgery, as well as the total loss of monocular visual function.

The check-ups after 10 days, one month and 6 months revealed postoperative normal VA in 22 (30.5%), 23 (31.94%) and 29 eyes (40.28%); mild and moderate loss in 28 (38.8%), 30 (41.66%) and 24 eyes (33.33%); severe reduction in 18 (25%), 8 (11.11%) and 8 eyes (11.11%); while NLP in 4 (5.5%), 4 (5.5%) and 11 eyes (15.28%), respectively.

Visual acuity remained the same in 43 eyes (59%) 10 days after surgery, including 20 eyes with normal preoperative visual acuity (VA=1.0). It remained unchanged in 34 (47%) and 28 (39%) one month and 6 months after surgery, respectively. Improvement in VA was found in 20 eyes (28%) after 10 days with the tendency of rising at 1 month and 6 month check-ups (28 eyes, i.e. 39% and 34 eyes, i.e. 47.22%, respectively). More than 2 lines of Snellen were gained in 5 (7%), 15 (21%) and 23 eyes (32%), 10 days, 1 month and 6 months after surgery. Vision aggravated in 9 eyes (13%) 10 days postoperatively and in 10 eyes (14%) one and six months after surgery (**Table 2, Graph 1**).

At first, binocular normal VA was present in 9 patients (25%) 10 days after surgery and then 10 patients (27.78%) were found to have it at the last check-up 6 months later. Initially, VA in the better eye was normal in 4 patients (11%) on the 10<sup>th</sup> day postoperatively and then in 5 patients (13.89%) and finally in 9 patients (25%) at two following check-

**Table 3.** Visual acuity of both eyes in relation to patients  
**Tabela 3.** Vidna oštrina oba oka u odnosu na pacijente

	Preoperative <i>Pre operacije</i>		10 days after surgery <i>10 dana posle operacije</i>		1 month after surgery <i>1 mesec posle operacije</i>		6 months after surgery <i>6 meseci posle operacije</i>	
Bilateral normal/ <i>Oba normalna</i>	8	21.6%	9	25%	9	25%	10	27.8%
Unilateral normal/ <i>Jedno normalno</i>	4	10.8%	4	11.1%	5	13.9%	9	25%
Unilateral 0,5-0,9/ <i>Jedno 0,5-0,9</i>	7	18.92	9	25%	13	36.1%	12	33.3%
Bilateral under 0,5/ <i>Oba ispod 0,5</i>	18	48.6%	14	38.9%	9	25%	5	13.9%
Total/ <i>Ukupno</i>	37	100%	36	100%	36	100%	36	100%

ups. Visual acuity lower than 0.5 in the better eye was found in 14 patients (39%) at first, but that number decreased with time, ending up with 5 patients (13.89%) 6 months after surgery (**Table 3**).

Null hypothesis that postoperative VA does not change with time after surgery was established and tested with Student t-test. The following values of t and p were calculated: t = -3.23881 (p=0.0012) 10 days after surgery, t = -4.53335 (p=0.0002315) one month after surgery and t = -5.79627 (p = 3.8 x10<sup>-7</sup>) 6 months after surgery. The negative values of t and p<0.01 indicate that null hypothesis was wrong, showing that there was a statistically significant increase in postoperative VA over the time, with the level of confidence of 99%.

Improvement of VA was observed in 47.22% of eyes, with the most prominent change in the eyes with normal VA from 27% preoperatively to 40% 6 months after surgery. There was also a significant decrease in the number of eyes with severely impaired VA: from 38% preoperatively to 11.6% after 6 months. Visual acuity remained unchanged in 39% and deterioration of vision was noted in 14% of eyes.

Regarding the number of patients, 36.58% of them had improvement of VA (at least in one eye), normal binocular VA was reached in 28% of them 6 months postoperatively, while 33% of patients achieved monocular VA of 0.5 and higher. There was no change in 28% of patients and a decrease in vision was noted in 14% of patients.

## Discussion

Craniopharyngiomas have always been controversial and “mysterious problem” for neurosurgeons, according to one of the pioneers of modern neurosurgery Harvey Cushing. This view is supported by their close relationship and adherence to vital structures at the base of the brain, their size and composition, as well as high mortality and postoperative morbidity in the form of motor and VA impairment, psycho-emotional and vegetative disturbances as the direct consequence of intraoperative injury of hypothalamus.

A number of new microsurgical techniques were developed in the last couple of decades which allowed for better visualization of tumor capsule, its relationship with surrounding tissues and better surgical removal of tumor itself. These advancements led to a significant decrease in mortality and

postoperative morbidity of patients and to an increase in survival rates with suitable quality of life.

The issue of quality of life is of great importance for patients operated on for benign tumors such as craniopharyngioma. Successful treatment involves normal motor, cognitive and intellectual development, and emotional stability of the patients, their ability to integrate into social and professional life and to provide for themselves. One of the most important factors for this is the reduction of neuropsychological sequels, one of them being visual impairment.

Craniopharyngiomas, tumors of dysembryogenic type, are diagnosed in young people, mostly in the second decade of life, and in middle age in the fourth and fifth decade. The patients included in our study were between 13 and 68 years of age, the average age being 30.5 years, which corresponds well with the scope of 27 to 42 years found in available literature [4, 9–11]. There were 92% of people over 16 years of age. The peak incidence of craniopharyngioma was in the second (37.83%) and fourth and fifth decade of life (32.43%), that being similar with findings of other authors [9–11]. Gender distribution was basically equal, 19:18 in favor of women.

It is well known that the patient’s age is an important factor for clinical presentation, tendency for growth, relapse affinity, as well as for the treatment prognosis of this tumor.

Our study showed no statistically significant difference in VA improvement among various age groups. However, several authors have correlated a poorer postoperative recovery of vision with young age, speculating that the reason for that is delayed tumour diagnosis in this age group, leading to more advanced visual impairment preoperatively [4, 10–12].

Two of the most common features in clinical picture of craniopharyngioma in our patients were a decrease in VA (83.7%) and endocrine disorders (35.1%), mostly in the form of gonadal insufficiency and diabetes insipidus. Headache, as an isolated symptom, was present in 12 patients (32.4%), and combined with nausea and vomiting, it was as a part of raised intracranial pressure in 6 patients (16.2%). Neurological disorders, with predominant motor or mental deficit (motor weakness, obliviousness, disorientation or depression) and paresis of oculomotor nerves, were observed in 4 patients (10.8%).

The majority of authors agree that monocular or binocular loss of vision is a prevalent finding in young

and adult craniopharyngioma patients [4, 7, 9, 11, 13–16]. Van Effenterre and Boch [4] have found it in 80% of cases, while other studies have shown it to be in the range from 42% to 72% [7, 11, 13, 15]. In our patient series there were 75.6% of patients with a decrease in VA, most of them binocular (67.5%). Severely reduced vision, at least in one eye was found in 65.7% of patients, and 83.7% had scotomata in visual field, regardless of their effect on the vision itself.

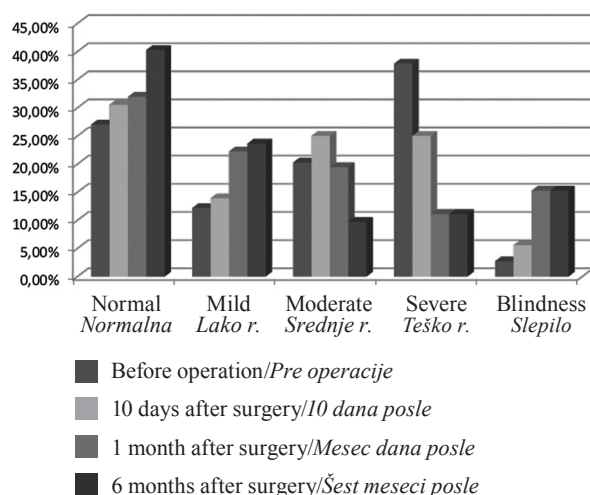
Such a high proportion of craniopharyngioma patients with affected vision can be explained with its retrochiasmal location, which primary leads to the destruction of crossed fibres of the optic nerve serving central parts of visual field.

Six months after surgery, 58.33% of our patients showed an improvement in VA, at least in one eye. In comparison with the situation before surgery when 21.62% of patients had normal VA in both eyes, it was evident that 6 months after surgical treatment there were 27.78% of patients with normal values of binocular VA. Furthermore, there were additional 25% of patients with normal monocular vision. Monocular visual acuity of 0.5 and higher was observed in 33.33% of patients, in contrast to 13.89% of patients who had binocular VA lower than 0.5. No change in vision after 6 months was recorded in 27.77% of our patients, most of them preserving normal preoperative values of VA (96.2%). As for the number of eyes involved, 39% of them maintained preoperative VA, 47% showed improvement, and only 14% suffered from a drop in vision 6 months after surgery.

Van Effenterre and Boch [4] reported an improvement in VA in 70% of eyes at the 2-month follow-up, half of them having achieved normal VA, and 15% of them showing no change, whereas VA deteriorated in 15% of eyes. Yamada et al. [14] studied a group of 61 patients and found that an improvement was observed in 90.2% of cases after surgery. Numerous recent papers report various degrees of postoperative improvement of VA, ranging from 52% to 93% [9, 14, 16–21].

Examples of significant improvement of post-surgical VA in the presence of severe or total preoperative loss of vision have also been known. Stark et al. [22] have described the case of a nine-year old child with NLP who had experienced a full visual recovery within a year after surgery and remained stable throughout the five year follow-up. The pathophysiological explanation for such an astonishing recovery includes gradual remyelination of nerve fibers and reorganization of neural connections in the lateral geniculate nucleus, leading to the improvement in synaptic transmission.

Usual intraoperative findings in patients with severely reduced preoperative VA include compression of large portions of optic nerve and optic chiasm with tumor tissue, leading to ischemia, stretching of the nerve or penetration of tumor cells into the nerve itself, making any surgical manipulation potentially hazardous. Compression on optic nerve fibers itself causes venous stasis and disruption of fast and slow phase of axoplasmatic transport. Edema and anoxia



**Graph 1.** Eyes (%) and visual acuity in different time intervals

**Grafikon 1.** Vidna oštrina u odnosu na broj očiju u različitim vremenskim intervalima

of nerve fibers ensue and later degeneration and demyelination of nerves rendering nerve impulse transmission defective or even impossible.

It was perceived that after surgical treatment of craniopharyngioma there was a quick improvement of VA in the first 10 to 14 days, followed by a slower recovery during the period of several months or even years [4, 14–16]. Our data show that there was a statistically significant improvement of VA in 36% of patients and 28% of eyes, in the first 10 days after surgery.

Elimination of this so called physical block of impulse transmission is at the core of early, quick recovery of visual function. Sometimes, prompt desorption of local edema of reversibly damaged nerve fibers happens. A later phase of the recovery is believed to be caused by remyelination and axoplasmic transport restoration along nerves [23, 24].

## Conclusion

It has been concluded that for the period of 6 months there is a statistically significant improvement of visual acuity regarding both the number of eyes and the number of patients reaching normal vision, at least in a better eye, which enables the patients to perform everyday vision related tasks.

Visual acuity in most of craniopharyngioma patients improves significantly in postoperative period, thus improving the quality of their life.

Craniopharyngioma and its treatment pose several challenges for patients suffering from it. One of the most common among them is an impairment of visual function. Our research shows that there is a constant and statistically significant improvement of visual acuity during the postoperative period of 6 months. Further studies are needed in order to make a proper assessment of the scope of its impact on the patients' quality of life.

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