

9

Original Article

How Effective is Superolateral Orbital **Decompression in Cases of Drug-Resistant** Graves' Ophthalmopathy

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Received: 11 Dec 2019 **Accepted:** 13 Mar 2020 **Published:** 30 Sep 2020

Citation: Shamov T, Eftimov T, Krasimirov G. How effective is superolateral orbital decompression in cases of drug-resistant Graves' ophthalmopathy. Folia Med (Plovdiv) 2020;62(3):462-7. doi: 10.3897/folmed.62.e49313.

Abstract

Introduction: Graves' ophthalmopathy (GO) is the most common and difficult-to-treat extrathyroidal symptom of Graves' disease. Though retraction of the upper eyelid is the most common clinical feature of GO, it can have a much more severe clinical manifestation with symptoms such as conjunctival chemosis, keratopathy, extraocular muscle dysfunction, proptosis of the bulb and dysthyroid optic neuropathy. Treatment methods include control of the thyroid function, corticosteroid and immunosuppressive therapy as well as radiotherapy. These approaches are ineffective in one-third of cases, with patients being refractory to all aforementioned therapeutic modalities. In these cases, surgical decompression of the orbit is in order.

The spectrum of surgical techniques is wide and varies from decompression of the lateral wall of the orbit to decompression via removal of all four orbital walls. The aim of the current study was to evaluate the results of superolateral orbital decompression.

Patients and methods: The study is retrospective and covers the period from January 2009 to January 2019. During that period eight patients with Graves' ophthalmopathy underwent surgery and were followed up in the Department of Neurosurgery in the Military Medical Academy, Sofia. The mean age of the patients was 57 years, with the youngest being 30 years old and the oldest - 74 years old. The gender distribution was 1.6/1 with predominance in females (5 women and 3 men). The surgical approach we used is a combination of lateral and upper orbitotomy and was described in detail by Al-Mefty. All of patients underwent ophthalmic examinations in the pre- and postoperative period, with special attention to their visual acuity, the condition of the eyelid and the width of the ocular slit. Exophthalmometry was obtained via Hertel's method. The participants in this study are followed for a period of six months after the operation.

Results: All eight patients underwent superolateral orbitotomy. There were a total of ten orbital decompressions. Improvement of visual acuity and reduction of the proptosis were reported in all other surgically treated patients. The mean reported improvement of visual acuity (measured via Snellen's method) was 0.27±0.17. The mean reported a reduction of proptosis was 7.53±2.58 mm.

Conclusions: Although the surgical techniques for orbital decompression we used have significant disadvantages, they remain the only alternative in order to avoid the complication of severe GO.

Keywords

exophthalmos, Graves' disease, neurosurgery, ophthalmopathy, thyroid diseases.

INTRODUCTION

Graves' ophthalmopathy (GO) is the most common and difficult-to-treat extrathyroidal symptom of Graves' disease.¹ The incidence of GO in Graves' disease is about 25%, yet with detailed neurological and radiological evaluations the incidence could reach up to 80%.²

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Although retraction of the upper eyelid is the most common clinical feature of GO, it can have a much more severe clinical manifestation with symptoms such as conjunctival chemosis, keratopathy, extraocular muscle dysfunction, proptosis of the bulb, and dysthyroid optic neuropathy.³ Most of the clinical features are attributed to an ongoing inflammatory process, leading to an increase of the intraorbital volume and of the retrobulbar pressure. The underlying reason for this process is the diffuse infiltration of the retrobulbar tissues by activated T-lymphocytes. The activation of fibroblasts is involved in the process as well. The production of glycosaminoglycans causes interstitial edema and fibrosis.⁴

Treatment methods include control of the thyroid function, corticosteroid, immunosuppressive therapy, and radiotherapy. In one-third of the cases these approaches are ineffective, with patients being refractory to all aforementioned therapeutic modalities. In these cases, a surgical decompression of the orbit is in order.

The spectrum of surgical techniques is wide and varies from decompression of the lateral wall of the orbit to decompression via removal of all four orbital walls.⁵ The approaches can be divided into anterior, mainly utilized by ophthalmologists and ENT specialists, and superolateral – subject of interest for neurosurgeons.

At the beginning of the 20th century, Kronlein introduced the lateral approach to the orbit in excision of orbital tumors. Later on, in the 1950s, Naffziger achieved transcranial decompression of the orbit by removing the orbital roof.⁶ In the 1970s, McCarty introduced the superolateral orbital decompression and reported very good results in 46 patients.⁷ Marron and Kennerdell modified this approach by transposing the skin incision in the facial region – the eyebrow, the lateral canthus, and the orbital crease achieving by this technique decompression of all four walls.^{8,9} Eliseevich also had input on the approach. He suggested a variant of orbital decompression with anterior displacement of the superolateral part of the orbital rim.¹⁰

AIM

The aim of the current study was to evaluate the results of superolateral orbital decompression.

PATIENTS AND METHODS

The study is retrospective and covers the period from January 2009 to January 2019. During that period, eight patients with Graves' ophthalmopathy underwent surgery and were followed up in the Department of Neurosurgery of the Military Medical Academy. The mean age of the patients was 57 years, with the youngest being 30 years old and the oldest – 74 years old. The gender distribution was 1.6/1 with predominance of females (5 women and 3 men). All operated patients had had a history of hyperthyroidism

symptoms with severe thyroid-associated ophthalmopathy. The mean duration of the disease from the moment it was diagnosed to the surgical intervention was 5 years, with variation of the period from 2 to 18 years. All patients had been administered L-thyroxin substitution therapy. One of the patients had been treated with combined L-thyroxin and methinazole. All patients had been treated with high dose corticosteroid therapy without satisfactory outcome. Four of the patients received immunosuppressive therapy and five underwent partial resection of the thyroid gland. Three of the patients underwent radiotherapy and another four were administered retrobulbar corticosteroids.

All of the patients underwent ophthalmic examinations in the pre- and postoperative period, with special attention to their visual acuity, the condition of the eyelid and the width of the ocular slit. Exophthalmometry was obtained via Hertel's method. Regular examination of eye-motility took place as well. The intraorbital pressure was measured and fundoscopy was performed. In two of the patients, there was an increase of intraorbital pressure. All patients were examined and followed up for 3-6 months postoperatively.

The data were statistically analyzed by using SPSS v.26. The main used methods are elements from descriptive statistics as mean value and standard deviation. T-test was used to compare paired samples and calculate the level of significance.

Description of the surgical technique

The surgical approach we used is a combination between lateral and upper orbitotomy and was described in detail by Al-Mefty.¹¹ The patient is placed on the operating table

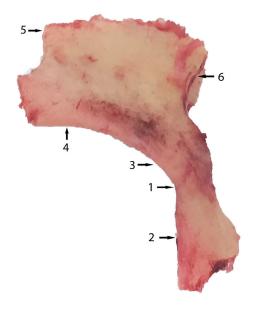


Figure 1. General appearance of the removed fronto-orbital flap. 1) Fronto-zygomatic suture; 2) Frontal procesus of the zygomatic bone; 3) Zygomatic procesus of the frontal bone; 4) Superior orbital edge; 5) Frontal bone; 6) Superior temporal line. in supine position, with the head fixated with three-point fixator in 30° rotation contralateral to the side of incision and in 20° deflexion. Antibiotic and antiedemic therapy is administered perioperatively. The skin is incised in the eyebrow region, 3-4 cm in length, twisting over the lateral orbital rim and the zygomatic arch, up to a maximum of 2 cm from the orbital rim (in order to prevent eventual lesion of the frontal branch of the facial nerve. Over the frontal flap, a periosteal flap is created, which can later be used for frontal sinuplasty. The supraorbital nerve is dissected and displaced medially. The periorbit is separated and a single frontal trepanation hole is made, which perforates the anterior and the posterior wall of the frontal sinus. The eyebrow rim is cut with a reciprocating oscillating saw blade. The temporal muscle is separated from the initial part of linea temporalis and the fronto-zygomatic edge. A second trepanation aperture is made in McCarty's point.⁷ Using an introductor, the dura is detached from the frontal bone, which is cut in arcuate mode with its convexity upward. With a Gigli saw, an oblique incision is made, cutting downwards the lateral orbital margin, after initial detachment of the periorbit. The orbital roof is carefully broken up, creating a small bone flap. After cranialization of the frontal sinus, the periostal flap is approximated to the ostium and fixated with fibrin glue.

prevent adhesions to the temporal muscle. We place drainage for 24-36 hours. Finally, we reapproximate and fixate the bone flap.

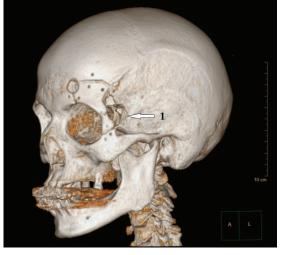
The general appearance of the removed fronto-orbital bone flap and 3-D CT reconstruction of the skull after supero-lateral orbital decompression are given in **Figs 1** and **2**.

RESULTS

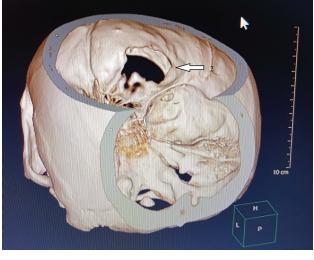
All eight patients underwent superolateral orbitotomy. The overall number of orbit decompressions was ten. The results of the surgical treatment are shown in **Table 1**.

Two of the patients underwent bilateral orbitotomy, completed in two stages with interval of one month (patients Nos 2 and 4). One of the patients was followed up for 3 months and due to unsatisfactory results was transferred to an ETN specialist for endoscopic decompression of the floor and the medial wall of the orbit. After that the patient was lost to the follow-up – patient No. 5.

Improvement of visual acuity and reduction of the proptosis were reported in all other surgically treated patients. The mean reported improvement of visual acuity (measured via Snellen's method) was 0.27±0.17. The mean repor-



2a



2b

Figure 2. Lateral (2a) and superior (2b) view of skulls with orbital decompression-3-D CT. 1) The lateral orbital wall is removed; 2) View of the removed superior orbital wall.

Afterwards, using fine bone instruments and a highspeed borer to sculpt the bone, the lateral wall of the orbit is thinned down to the inferior orbital fissure in a classical, clastic way. The upper wall is removed as well and with that, the orbit is revealed in its upper-lateral aspect.

A longitudinal cut is made and in case there is prolapsed adipose tissue, it is removed. Next, meticulous hemostasis is achieved and in some cases, we have used Lyoplant to ted reduction of proptosis was 7.53±2.58 mm. It is worth noting that all of the results were reported at least three months after the procedures, after full recovery of all soft tissue postoperative alterations. All patients who underwent surgery had improvement of the soft tissue status and disappearance of the diplopia. Complications were observed in three patients – one with partially damaged frontal branch of the facial nerve, and the other two – with dysfunction of

Table 1. Results of surgical treatment (reported between month 3 and month 6), in patients with severe form of GO, using superolateral	
orbitotomy	

Case No.	Sex	Age yrs	L/R	Visual acuity Assessed using Snelen method N=10 (eyes)		Improvement of visual acuity	Exophthalmos In millimetres using Hertel method N=10 (eyes)		Orbital recession					
				Preop	Postop		Preop	Postop						
1	F	46	D	0.5	0.8	0.3	23	17	6					
h	м	м	м	м	м	м	4.4	L	0.3	0.6	0.3	26	17	9
2	М	44	D	0.5	0.8	0.3	24	18	6					
3	М	30	D	0.8	1.0	0.2	27	17	10					
4	F	F	Г	E 4	L	0.4	0.6	0.2	29	18	11			
4			54	D	0.6	0.8	0.2	26	19	7				
5*	F	57	L	0.7	0.9	0.2	27	24	3					
6	М	70	D	0.8	0.9	0.1	25	14	10					
7	F	74	D	0.9	1.0	0.1	28	22	6					
8	F	68	L	0.5	0.8	0.3	23	18	5					
				0.6±0.2	0.82±0.1	0.27±0.2	25.8±2	18.4±2	7.3±3					
					p<0.001			p<0.001						

*This patient was followed up for 3 months and then she dropped out of observation.

the supraorbital nerve.

Good cosmetic results were achieved in all patients. The applied statistical methods, as noted in **Table 1**, showed significant difference in the visual acuity and the grade of the exophthalmos before and after the intervention.

DISCUSSION

The surgical technique we applied is effective in severe cases of Grave's disease and the study we held showed statistically significant improvement of the visual acuity and reduction of the proptosis. Surgical treatment in such cases is the only alternative. It is only recently that the biological treatment of GO became a part of the clinical practice. To date there has been too little experience and data on the effects of rituximab, monoclonal antibody against CD20 receptor of the B-lymphocytes, which decreases the production of antibodies against the TSH receptor of fibrocytes. There are two studies which fail to provide strong evidence of the effectiveness of the drug.^{12,13} There are high hopes for a future therapy with tocilizumab and teprotumumab, yet to this moment the scientific data for their effectiveness are insufficient.¹

Superolateral techniques for decompression, mainly practiced by neurosurgeons, allow reduction of the proptosis around 0.6 to 1 cm to be achieved.^{14,15} Our study concluded mean values around 7.53±2.58, which does not differ significantly from the results of similar studies. Infero-medial decompressions lead to reduction of mean 6.9 mm, yet these decompressions have a high rate of complications (up to 20%), such as CSF leakage and orbital cellulitis. Neurosurgical approaches allow direct control of intraoperative

complications, such as dural lacerations and opening of the frontal sinus. Lateral decompression shows reduction of the proptosis around 3-4 mm. The effects are optimal with decompression of all four walls – around 1-1.5 cm.^{9,14} Vision acuity is improved in almost all orbital decompression techniques, with insignificant variation of the measured values in the different studies. Most authors conclude that orbital decompressions should be recommended in cases with vision acuity measured lower than 0.6 and proptosis over 20-23 mm.¹⁴

The main limitation of our study is the low number of patients included, which calls into question the statistical significance of the results. The cardinal disadvantage of the applied technique is the incision in the facial region and the risk of stretching and lesions to the nerves in the area, due to the chartering. In order to avoid these problems, it is reasonable to apply the coronary approaches – with incision in the capillitium. Even though the operative wound is wider, these approaches allow maneuvers to protect the facial and supraorbital nerves.

Last but not least in importance, it is essential to acknowledge the fact that measuring the exophthalmos using the Hertel's method in the postoperative period could alter the results and lead to mistakes due to the edema of the soft tissues and the displacement of bone structures. This fact itself puts into question the verity of the reported results in different studies.

As recommendations for future studies on the matter, we suggest measurement of the results in surgical approaches that include anterior displacement of superolateral margin of the orbit, which leads to enlargement of the orbital space. The technique, proposed by Eliseevich L et al., sounds promising in this respect.¹⁰

CONCLUSION

In conclusion, though there are significant disadvantages of the applied surgical techniques for orbital decompression, they remain the only alternative to avoid the complication of severe GO.

Acknowledgements

The authors declare no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Насколько эффективна суперолатеральная орбитальная декомпрессия в случаях лекарственноустойчивой офтальмопатии Грейвса

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Дата получения: 11 декабря 2019 **• Дата приемки:** 13 марта 2020 **• Дата публикации:** 30 сентября 2020

Образец цитирования: Shamov T, Eftimov T, Krasimirov G. How effective is superolateral orbital decompression in cases of drug-resistant Graves' ophthalmopathy. Folia Med (Plovdiv) 2020;62(3):462-7. doi: 10.3897/folmed.62.e49313.

Резюме

Введение: Офтальмопатия Грейвса (ОГ) является наиболее распространённым и трудным для лечения симптомом экстратиреоидной болезни Грейвса. Хотя ретракция верхнего века является наиболее распространённой клинической особенностью ОГ, она может иметь гораздо более тяжёлые клинические проявления с такими симптомами, как конъюнктивальнный хемоз, кератопатия, дисфункция экстраокулярных мышц, проптоз глазного яблока и дистироидная зрительная нейропатия. Терапевтические методы включают контроль функции щитовидной железы, кортикостероидную и иммуносупрессивную терапию и лучевую терапию. Эти подходы неэффективны в одной трети случаев, и пациенты не реагируют на вышеупомянутые методы лечения. В этих случаях проводится хирургическая декомпрессия орбиты.

Диапазон хирургических методов широк и варьируется от декомпрессии боковой стенки орбиты до декомпрессии путём удаления всех четырёх стенок орбиты. Целью настоящего исследования была оценка результатов суперолатеральной орбитальной декомпрессии.

Пациенты и методы: Исследование было ретроспективным и охватывало период с января 2009 года по январь 2019 года. За этот период восемь пациентов с офтальмопатией Грейвса перенесли операцию и были обследованы в отделении нейрохирургии Военно-медицинской академии, София. Средний возраст пациентов составлял 57 лет, самому молодому – 30 лет, а самому старшему – 74 года. Распределение по полу составило 1.6 / 1, причём преобладали женщины (5 женщин и 3 мужчины). Хирургический подход, который мы использовали, представляет собой комбинацию боковой и верхней орбитотомии и подробно описан Аль-Мефти. Все пациенты проходили обследование глаз в пред– и послеоперационный период, особое внимание было уделено остроте зрения, состоянию века и ширине глазной щели. Экзофтальмометрия проводилась методом Гертеля. Участники этого исследования наблюдались в течение шести месяцев после операции.

Результаты: Все восемь пациентов прошли суперолатеральную орбитотомию. Всего было выполнено десять орбитальных декомпрессий. У всех других пациентов, получавших хирургическое лечение, отмечалось улучшение остроты зрения и уменьшение проптоза. Сообщённое среднее улучшение остроты зрения (измеренное методом Снелена) составило 0.27 ± 0.17. Среднее измеренное снижение проптоза составило 7.53 ± 2.58 мм.

Заключение: Хотя применяемые нами хирургические методы декомпрессии орбиты имеют существенные недостатки, они остаются единственной альтернативой, позволяющей избежать осложнений тяжёлой ОГ.

Ключевые слова

экзофтальм, болезнь Грейвса, нейрохирургия, офтальмопатия, заболевания щитовидной железы