Chapter 18

Malignant Glaucoma

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Additional information is available at the end of the chapter

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1. Introduction

Malignant glaucoma was described for the first time and named so by Albrecht von Graefe in 1869 [1]. It is characterized by normal or increased IOP *(intraocular pressure)* associated with axial shallowing of the entire anterior chamber in the presence of a patent peripheral iridotomy [2,3]. The pathology is based on the existence of a block for normal flow of aqueous humour, which results in the accumulation of aqueous at an improper location in the eyeball [4]. The proposed mechanism involves a misdirection of aqueous humour passing posteriorly into or behind the vitreous gel [5]. This is a dynamic process, and if untreated, causes loss of vision. Local hypotensive treatment does not cause normalization of IOP, and conventional glaucoma surgery proves to be ineffective [3].

1.1. Classification

Classification includes phakic, aphakic, and pseudophakic malignant glaucoma. Aphakic malignant glaucoma is the onset of symptoms after a cataract surgery or the persistence of symptoms after treatment of phakic malignant glaucoma through the cataract extraction [6]. "Non-phakic malignant glaucoma" is a general term used for both types: aphakic and pseudophakic malignant glaucoma [6]. The term *malignant-like glaucoma* was proposed for cases with a known cause of forward displacement of the lens along with the frontal surface of the vitreous body other than the "trapping" of humour inside of the vitreous body [7]. There also exists a classification of malignant glaucoma into that occurring after surgical intervention and without such intervention [8].

The not fully known etiology of the process creates difficulties in the standardization of nomenclature. Certain authors suggest that the malignant glaucoma group should exclude cases in which e.g. pupillary block or choroidal detachment has been stated [9]. Others believe that using this term to encompass a broader spectrum of eye diseases will create a better un-

derstanding of the pathophysiology and the relationship between pathologies with similar clinical pictures [4].

1.2. Occurrence

According to literature, malignant glaucoma develops in 2% to 4% of patients with a history of acute or chronic angle-closure glaucoma that have undergone filtration surgery [3]. In own material, consisting of a total of 1689 penetrating and non-penetrating operations, performed as glaucoma surgery alone or combined with cataracts, malignant glaucoma occurred in 1.3% of all eyes after surgery. After penetrating surgery this complication was noted in 2.3% of eyes. It was also observed after laser iridotomy [10], phacoemulsification of cataract [11], posterior capsulotomy using a Nd-YAG laser *(Neodymium-yttrium-aluminum-garnet laser)* [12], cyclophotocoagulation [13], after implantation of large-sized IOLs *(intraocular lens)* [14], after local application of miotics [15], after suturolysis [16], and even in eyes that did not undergo surgical procedures [17]. Cases of malignant glaucoma have also been described in eyes in which glaucoma had not been established earlier [11].

Malignant glaucoma occurs significantly more frequently after penetrating surgery than in the case of non-penetrating surgery, after just the glaucoma surgery than after treatment combined with phacoemulsification, as well as in eyes with narrow angle glaucoma. It was stated with greater frequency among women, which may be related to the lesser dimensions of the anterior segment of the eyeball in this group of patients [18]. This complication can take place at various times after the operation, sometimes immediately, and sometimes after one year has passed or even after a longer period of time [3].

2. Anatomical basis

It is considered that incorrect anatomical relationships lead to disruptions in the direction of aqueous humour flow [4,19]. The place of increased resistance may be located at the level of the iris-lens, ciliary-lens, iris-hyaloid, and ciliary-hyaloid block [4,20]. Structures that are particularly related to the development of malignant glaucoma and its clinical picture:

Sclera – a thick sclera may lead to partial stenosis of the vortex veins, impairing normal venous outflow and causing overfilling of the choroid [21], as stated in eyes with malignant glaucoma [22]. Opening of the anterior chamber during surgery, which causes lowering of IOP, together with possible movements of the irido-lenticular diaphragm can trigger a malignant glaucoma mechanism in such eyes.

Lens – the exciting cause for malignant glaucoma in many cases is a lens that is too large for the eye [23]. Disproportions between its volume and the volume of the entire eyeball can occur; furthermore, particular anatomical relationships between the anterior vitreous, ciliary processes, and the lens foster the occurrence of malignant glaucoma [4,19].

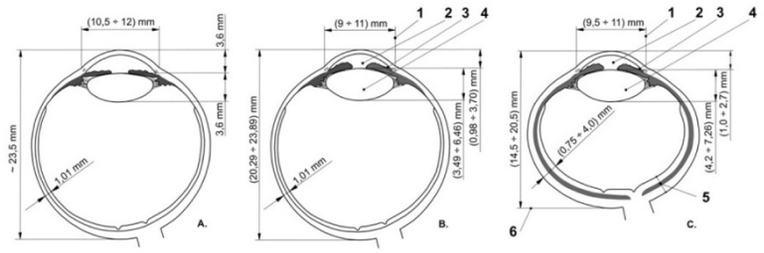
Choroid – the choroid has a lobular structure with a tendency for accumulation of blood and thickening when outflow is impaired. Secondary, ciliary body and iris rotate to the front in patients with malignant glaucoma [24], closing access to the filtration angle from the back.

Vitreous body – Slit-lamp examination of the vitreous may reveal optically clear areas within the vitreous body – reservoirs of aqueous humour trapped in its gel structure [3], which may be confirmed on ultrasound [25]. In aphakic eyes, the anterior surface of the vitreous body may directly adhere to the ciliary processes [3].

The anterior and posterior chambers and their relationship – total obliteration of the posterior chamber by the vitreous and a highly resistant hyaloid membrane may be observed in aphakic and pseudophakic eyes [26].

3. Predisposing factors

The anatomic and functional differences of predisposed eyes seem to be a significant factor for determining the occurrence of malignant glaucoma. The following predisposing factors have been described, among others: axial hyperopia [27], nanophthalmos [28], disorders of anatomical proportions in the anterior chamber [18].



NORMAL EYE	RELATIVE ANTERIOR MICROPHTHALMOS (RAM)	NANOPHTHALMOS
	1 – Decreased corneal diameter	1 – Decreased corneal diameter
	2- Shallow anterior chamber	2- Shallow anterior chamber
	3 – Risk of the filtration angle closure	3 – Risk of the filtration angle closure
	4 – The lens takes up a disproportionately large percetnage of the volume of the eye	4 – The lens takes up a disproportionately large percetnage of the volume of the eye
		5 - Thickening of the choroido-scleral layer
		6 - Decreased axial length

Figure 1. Normal eye, relative anterior microphthalmos and nanophthalmos.

It is considered that malignant glaucoma is related to a special eye anatomy (small eye phenotype). Lynch et al. stated that it occurs more frequently in small eyes with an anatomically narrow iridocorneal angle [11]. Many nanophthalmic and RAM eyes have narrow angles with crowded structures in the anterior chambers. Typically, the lens is of normal or increased thickness, leading to a high lens:eye ratio and this crowding results in a shallow anterior segment that predisposes to angle-closure glaucoma [21]. In microphthalmos, due to small eye size, the increase in the size of the lens with age is critical,

and a relative pupillary block forms with progressive shallowing of the central and circumferential anterior chamber, narrowing, and gradual angle closing [29]. However this is not the only angle closing mechanism in this pathology. Peripheral iridectomy, which eliminates pupillary block, does not prevent progressive overfilling of the choroidal bed, which may cause further angle closure. If the aqueous humour is directed to the vitreous cavity instead of the posterior chamber, symptoms of malignant glaucoma will occur [30]. Thus, in a genetically conditioned microphthalmos, glaucoma with a complex iris and ciliary block may be expected [20].

The occurrence of malignant glaucoma in the pathology that is the microphthalmos may not only be connected to abnormal anatomical relationships but also to incorrect histological structure of the sclera. The sclera in a microphthalmos is thicker relative to physiological conditions and its collagen fibers are more disorganized [31]. Trelstad et al. stated, that in a microphthalmos, collagen fibers of the intercellular substance in the connective tissue of the sclera have a normal thickness, but the collagen fibers are longer, less organized, and more interwoven [32]. Yue et al. stated that a greater heightened level of fibronectin, and speculated that a change in the glycosaminoglycan metabolism may influence the contraction of collagen fibers and lead to thickening of the sclera. The authors believe, that an incorrect glycosaminoglycan metabolism may cause a decrease in the elasticity of the sclera, which hampers normal development of the eye [33]. Based on known measurements of the thickness of the sclera, increased thickness of the tissues, including the retina, choroid, and sclera in echographic measurements was considered to be a value above 1.7 mm [34]. The increased thickness of the sclera in hyperopic eyes and its simultaneously lower surface area decrease transscleral protein transport, what, in consequence, causes choroidal expansion [22]. According to Quigley et al., a similar situation occurs in many eyes that do not achieve such small sizes, and malignant glaucoma can occur in eyes of correct sizes as well as in small eyes, but all cases would have dramatic choroidal expansion or vitreous flow abnormality [22]. In the case of a nanophthalmos, a tendency toward spontaneous or postoperative uveal effusion was also observed [21]. Quigley et al. observed that in eyes with extremely small sizes, displacement of the lens to the front occurs, caused by choroidal expansion [22]. Furthermore, the increased pressure in the vortex veins occurring in a microphthalmos as well as disrupted transscleral protein transport and increased oncotic pressure of the vitreous body may be linked to an increased risk of development of malignant glaucoma [21,22].

One of the more important factors predisposing the occurrence of malignant glaucoma is also partial or total closing of the filtration angle at the time of the surgery, especially if the malignant glaucoma occurred in the second eye [3]. However, IOP has no direct correlation to the risk of occurrence of malignant glaucoma. In Simmons's studies, the IOP level during the operation was not correlated with the probability of development of malignant glaucoma after surgery [3]. Moreover, it should be pointed out, that in the case of malignant glaucoma in one eye, the fellow eye exhibits a predisposition for occurrence of a malignant process [6].

4. Pathomechanism

The causes of malignant glaucoma are complex and there are several theories on the subject of factors that may have an influence on its development. As of now, the pathophysiological mechanism of malignant glaucoma is not yet fully understood. There is no certainty as to what structures or biochemical processes lead to the development of malignant glaucoma, and its cause seems to be conditioned by many factors.

An anterior rotation of the ciliary body processes, leading to ciliolenticular touch and ciliary block, has been suggested [25]. Forward displacement of a relatively large lens, which then blocks communication between the posterior and anterior chamber, as well as outlets from the eye, is the essential anatomical feature of malignant glaucoma [35]. Congestion of the uveal tract may play a part in pushing the lens into its forward position and holding it there [35]. In addition, in certain cases, the lens capsule and zonules may constitute a place of resistance for the flow of aqueous humour to the front [36]. The aqueous humour produced to the posterior chamber is directed to the back instead of to the anterior chamber [5], causing anterior displacement of the lens-iris diaphragm. Furthermore, swelling of the ciliary processes caused by inflammation or miotics can cause critical narrowing of an already anatomically narrow space between the lens equator and the ciliary body and relative block of forward aqueous flow [18]. Abnormal choroidal circulation may also lead to accumulation of blood and swelling of the ciliary processes. Moreover, Epstein and coauthors hypothesized that there is decreased permeability of the vitreous body or the anterior hyaloid to anterior flow of aqueous humour into the anterior chamber in malignant glaucoma [37].

Probably, there are eyes with predispositions for malignant glaucoma, in which there is a pathology of connective tissue related to a predominance of intercellular substance, mainly comprised of glycosaminoglycans. Glycosaminoglycans produced by fibroblasts of pathological connective tissue accumulate in the vitreous of such eyes with malignant glaucoma. Glycosaminoglycans, together with proteins gathered in the vitreous body because of impaired transscleral outflow, are responsible for the increase of oncotic pressure and accumulation of water. Moreover, high viscosity caused by mucopolysaccharides content makes the flow from the posterior to the anterior chamber more difficult. Glycosaminoglycans may also be a cause of iridocorneal angle damage.

The coexistence of anatomical and physiological predispositions and changes in IOP in the anterior chamber during surgery, activates a specific pump mechanism caused by movements of the lens-iris diaphragm, which may have an influence on the development of malignant glaucoma. The malignant process can have various dynamics with clinical manifestation occurring directly after surgery, when exciting factors cannot be compensated in the closed system of the eyeball. On the other hand, the occurrence of malignant glaucoma symptoms may be delayed if a relative equilibrium between the volume of the produced fluid and the outflow from the eyeball is reached.

5. Objective symptoms

Myopic shift in refraction related to the anterior dislocation of the iris-lens diaphragm with secondary improvement of near vision [38].

Narrowing or shallowing of the circumferential and central part of the anterior chamber even if patent iridotomy or iridectomy is present. Shallowing of the anterior chamber is related to anterior dislocation of the iris-lens diaphragm [39,40] and iris-hyaloid diaphragm with coexistence of increased IOP [40]. Persistent symptoms of malignant glaucoma lead to the formation of intensified anterior adhesions due to the long-lasting shallowing of the anterior chamber [41].

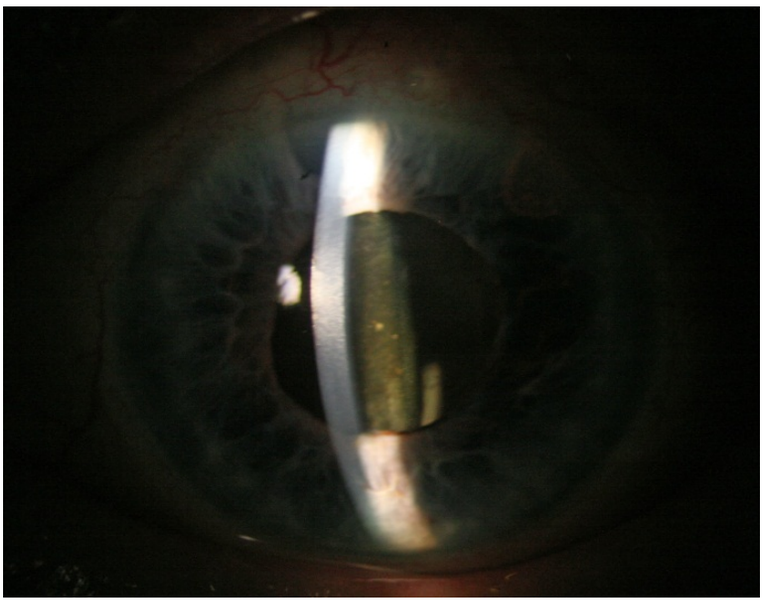


Figure 2. Axial shallowing of the anterior chamber in an eye with malignant glaucoma.

Increased IOP – intraocular pressure may increase slowly with simultaneously intensifying shallowing of the anterior chamber [42]. It is characteristic that in the presence of an active, well functioning filtering bleb, the increase in intraocular pressure can be moderate [43].

No decrease of IOP in response to conventional antiglaucoma treatment [4].

In many cases, a decrease of IOP or curing as a result of mydriatic-cycloplegic therapy [44].

Reaction to surgical treatment of the vitreous body [6].

6. Differential diagnosis

Glaucoma with pupillary block – pupillary block angle closure occurs when the posterior surface of the iris, in the pupillary margin, comes in contact with the lens. The increased pupillary block obstructs the flow of the aqueous humour from the posterior chamber to the

anterior chamber, resulting in increased pressure in the posterior chamber and forward bowing of the peripheral iris. This closes the anterior chamber angle, obstructing the trabecular meshwork and the outflow channels with subsequent elevation of the IOP. Laser peripheral iridotomy is the treatment of choice [45] and should be performed in all cases of pupillary block glaucoma. In pupillary block, there should not exist axial shallowing of the anterior chamber (movement of the IOL toward the cornea). The anterior chamber usually remains deeper in the center than on its circumference, in contrast to malignant glaucoma, where axial chamber shallowing also occurs. If there is axial shallowing, then fluid has somehow moved posteriorly and the vitreous is acting to shallow the chamber [46].

Angle closure glaucoma – shallowing of the anterior chamber occurs symmetrically in both eyes. In the affected eye, the filtration angle is closed, there is a sudden increase in IOP, and microcystic edema of the cornea. Conjunctival injection and a medium size pupil may accompany these symptoms [47]. It occurs regardless of surgery and is caused by anatomical predisposition.

Choroidal effusion - a static condition which is observed independently of operation and has inflammatory (trauma and intraocular surgery, scleritis, following cryocoagulation and photocoagulation, chronic uveitis, Vogt-Koyanagi-Harada disease) or hydrostatic causes (hypotony and wound leak, dural arteriovenous fistula, abnormally thick sclera in nanophthalmos, possibly in emmetropic or myopic eyes or associated with Hunter's syndrome). Uveal effusion should not be considered to be a distinct clinical entity but rather a state characterized by abnormal amounts of fluid in the choroid resulting in thickening of the choroid, accumulation of fluid in the suprachoroidal space resulting in choroidal detachment, and in some cases, accumulation of fluid in the subretinal space, resulting in nonrhegmatogenous retinal detachment. IOP may be normal but is often reduced in uveal effusion secondary to inflammatory factors. An exception occurs in nanophthalmic uveal effusion wherein IOP is normal or frequently elevated and chronic angle closure glaucoma may develop [48].

Suprachoroidal hemorrhage – shallowing of the anterior chamber coexists with increased IOP, sudden pain, and the presence of a haemorrhagic, non-serous detachment of the choroid in biomicroscopic and ultrasonographic examination. It occurs most often within 1 week after surgery, rarely later [6]. Suprachoroidal hemorrhage may be caused by bleeding diathesis, anti-coagulants, paranasal sinusitis, or may occur spontaneously. Small suprachoroidal hemorrhages occurring during surgery are usually absorbed extemporaneously. Suprachoroidal hemorrhage may be also related to postoperative hypotony, and in the late postoperative period, may be connected to increased venous pressure or increased tension of the abdominal press.

7. Testing

Medical history – determination of predisposing factors and early statement of symptoms accompanying the occurrence of malignant glaucoma

Slit lamp examination – assessment of the depth of the anterior chamber shows that there is axial (central and peripheral) shallowing of the anterior chamber and, unlike in pupil block, the iris is not typically bowed forwards, and anterior lens movement is noted. Patency of the iridotomy, if such exists, should be evaluated – if there is no iridotomy or the patency is in doubt, laser iridotomy can be performed or repeated to rule out pupil block, but it does not cause resolution of the condition. Seidel test should be performed to exclude filtering bleb leaking after filtration surgery. Biomicroscopy assessment of the posterior segment is necessary for the purpose of ruling out choroidal detachment or suprachoroidal hemorrhage

Tonometry – usually reveals increased IOP

Ultrasonography – conducted for the purpose of determining the axial length of the eyeball (which tends to be shorter than normal) and to determine the position and size of the ciliary body and its processes [25]. Moreover, information on the thickness of the choroid may be obtained through ultrasonographic examination

Ultrabiomicroscopy (UBM) – this test gives images of the iris, the intraocular lens and ciliary body as well as their relative positions before and after the occurrence of malignant glaucoma. The rotation of the ciliary body to the front and shallowing of the anterior chamber may be subject to normalization after tearing of the anterior hyaloid [24]. This test enables visualization of the structures of the anterior segment, although the capability of conducting tests in the early postoperative period is limited due to the immersion technique

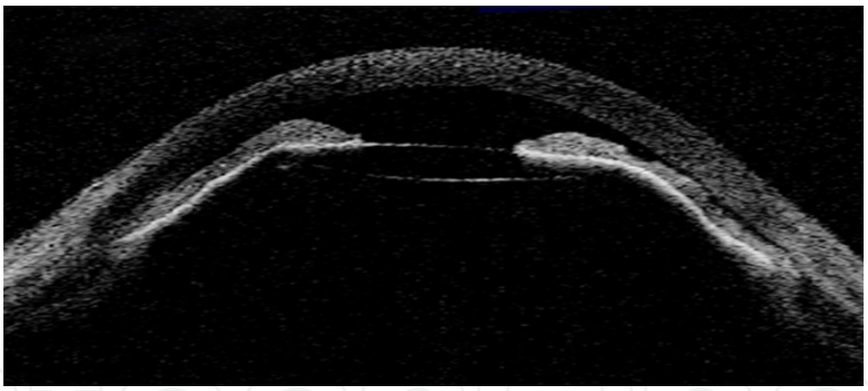


Figure 3. OCT of the anterior segment in malignant glaucoma – shallowing of the anterior chamber, peripheral irido-corneal touch, forward shift of the IOL.

Anterior segment OCT *(optical coherence tomography)* – a non-invasive high resolution technique that can be used for the purpose of objective imaging of the iridocorneal angle structure as well as for qualitiative and quantitative assessment. Parameters such as: AOD – *anterior chamber opening distance,* ACA – *anterior chamber angle* have been adapted from ultrasound biomicroscopy for the OCT method. Measurements of scleral thickness, CCT – *central corneal thickness*, and central depth of the anterior chamber during an episode of malignant glaucoma can also be conducted. Marked displacement of the structures of the anterior segment, peripheral irido-corneal touch, and forward shift of the lens may be noted Examination may reveal a decreased anterior chamber angle with extreme shallowing of the anterior chamber

depth during the acute malignant glaucoma phase and an increase of ACA and AOD quantitative values after effective treatment of this condition. It is helpful to objectively evaluate the structures of the anterior chamber or to monitor changes in the anterior segment after surgery. Since the presence of corneal oedema is an indication of prompt surgical intervention it can be used to assess this parameter in a non-contact fashion [40].

8. Treatment

8.1. Conservative treatment

The goal of conservative treatment is to decrease the production of aqueous humour and shrink the vitreous while simultaneously decreasing resistance in the path of aqueous humour flow to the anterior chamber through applied cycloplegia.

The active mechanism of the drugs used in the treatment of malignant glaucoma is as follows:

Mydriatics – cycloplegics – paralysis of the ciliary muscle, widening of the ciliary processes ring, tightening of the zonule apparatus, backwards movement of the lens.

Osmotically active agents – increase of blood osmolality causing movement of water from the eyeball in the direction of hyperosmotic plasma, which results in a decrease of the hydration of the vitreous body and makes it possible to retract the iris-lens diaphragm and deepen the anterior chamber.

β-blockers – suppression of aqueous humour production, as a result of which the volume of humour directed towards the vitreous is reduced.

Carbonic anhydrase inhibitors – reduction of secretion of aqueous humour by inhibiting carbonic anhydrase activity in the epithelium of the ciliary body.

Corticosteroids – by limiting inflammation, they reduce edema in the area of the ciliary body and help to minimize inflammatory adhesions of the lens or vitreous body with the ciliary body [20].

According to data from the literature, approximately 50% of patients react to medical therapy [3]. In the work of Debrouwere et al., however, the percentage of recurrences after conservative treatment of patients with malignant glaucoma was equal to 100%, despite an initially good response to such therapy [49]. Also, in own experience, a lack of success in reversing the pathogenic mechanism by means of conservative treatment in malignant glaucoma concerns the great majority of cases. In own material, reactions to conservative treatment were observed in 5 eyes with malignant glaucoma out of 22 of those tested [22.7%), however, ultimately, a surgical procedure was necessary in three of them due to the recurrence of typical symptoms and no control over IOP. Permanent improvement after pharmacological treatment was achieved in only 2 eyes [9.1%). The observations of other authors also confirm transient effectiveness of medical therapy during the initial period [11,42]. Even if IOP con-

trol is achieved as a result of such treatment, long-term cycloplegia is necessary to maintain this effect in many eyes [25]. In some cases, when medications are discontinued or changed, tendencies of recurrence of malignant glaucoma symptoms are observed [50]. Therefore, medical treatment is thought to be of temporary effect and is used until definite treatment with laser iridotomy, posterior capsulotomy and hyaloidotomy is performed. The currently valid regimen for conservative treatment includes locally applied: atropine, phenylephrine, β-blockers, acetazolamide, and generally administered 50% glicerol solution in oral doses and intravenously administered mannitol. Locally applied corticosteroids play the role of limiting the accompanying inflammatory process. If improvement has been achieved, the dosage of hyperosmotic agents can be decreased, followed by carbonic anhydrase inhibitors, however treatment with mydriatic-cycloplegic agents should be continued [3]. The following treatment schedule can also be applied: mannitol 2 g per kg intravenously once or twice a day, acetazolamide 250 mg tid, and locally: 1% Tropicamide qid, Cosopt (dorzolamide hydrochloride-timolol maleate ophthalmic solution) bid, 0.1 % Dexamethasone phosphate tid. This regimen is usually successful until laser treatment is performed.

8.2. Surgical treatment

8.2.1. Laser treatment

Laser therapy is usually used together with conservative treatment and should be performed as early as possible because postponement of this therapy may lead to increased IOP with injury to the optic nerve and loss of visual field as a consequence, flattening of the anterior chamber, corneal-lens touch, and corneal decompensation. This method of management can also be used after malignant glaucoma surgery, and then can serve to sustain or restore the effects of the operation. The main limitation of laser techniques – excluding transscleral cyclophotocoagulation with a diode laser – is their dependency on corneal transparency. Topical glycerol may lead to temporary clearance of corneal edema and make the procedure viable.

In cases of suspected malignant glaucoma, pupillary block should be eliminated as a possible contributory element to the shallow anterior chamber by assessing the size and patency of iridotomy, when present, or by the creation of a patent iridotomy, if necessary [51]. Surgeons may prefer to use the Nd-YAG laser alone or argon laser pre-treatment followed by the Nd-YAG laser. With an Nd-YAG laser energy of 2-5 mJ, 1-3 pulses per burst are usually used.

Currently, as the treatment of choice in aphakic and pseudophakic eyes, several laser effects are used in combination: laser iridotomy with anterior hyaloidotomy and posterior capsulotomy, all through the same location. In this case, a positive effect of laser therapy is the creation of direct communication between the vitreous, the posterior chamber, and the anterior chamber, and such a procedure can restore normal dynamics of aqueous humour flow in malignant glaucoma [52]. If needed, it may be applied in more than one location.

Capsulotomy is usually performed using an energy of 1 to 4 mJ per pulse. The energy and pulses may be increased gradually according to the thickness of the capsule until an open-

ing is achieved. 5-15 bursts with an energy of 1-3 mJ through iridotomy or iridectomy are usually effective in achieving communication. An immediate effect of such a procedure is often observed in the form of deepening of the anterior chamber. If there is no access to the iridectomy, communication can be achieved through the lens capsule in a pseudophakic eye using an energy of 1 mJ near the edge of the IOL. Such a procedure may be preceded by decompressing the vitreous chamber by puncturing it with a 25 gauge needle through pars plana. The above scheme may be repeated. The magnitude and patency of communication between the anterior and posterior segments of the eye are decisive to the distribution of pressure between the anterior and posterior segment of the eyeball. Recurrences may occur even when communication is present but is not effective enough to decrease the force shifting the iridolenticular diaphragm forward. In the case of difficult access to the circumferential part of the lens capsule in the area of iridectomy, the effect of deepening of the anterior chamber is to be achieved by creating a capsulotomy within the pupil or outside the edges of the artificial lens, after which a capsulotomy in the area of the iridectomy that is as large as possible should be created.

The goal of Nd-YAG laser hyaloidotomy, in turn, is to tear the anterior hyaloid face, as a result of which the depth of the anterior chamber is normalized [24]. Epstein and others treated aphakic and pseudophakic eyes with an energy of 3 to 11 mJ delivered to the anterior hyaloid face [53]. This treatment can be conducted through surgical iridectomies or laser iridotomies, often in many places. It is carried out centrally, to the back of the lens capsule, or in combination with capsulotomy in pseudophakic patients [46].

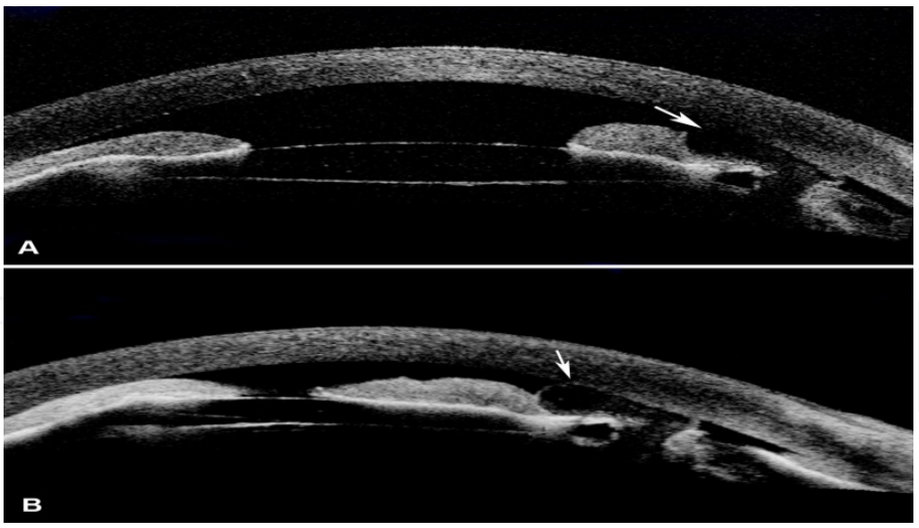


Figure 4. Anterior segment OCT in eye with malignant glaucoma – complication after Nd-Yag laser capsulotomy with hyaloidotomy - hyaloid gets across the iridotomy into the anterior chamber (white arrow); note shallow anterior chamber, forward movement of the IOL and iridocorneal touch at considerable area.

Transscleral cyclophotocoagulation is a procedure with different applications. The laser beam causes ablation of the ciliary body, which causes reduction of aqueous humour secretion. Energy absorption by melanine leads to thermal coagulation and destruction of

the pigment epithelium and accompanying vessels. Deep coagulative necrosis of the pigment epithelium, pathological reconstruction of collagen fibers in the stroma, and intravascular coagulation in the blood vessels of the ciliary body take place [43]. Significant complications include postoperative inflammation, pain, cystoid macular edema, and phthisis. Thus, indications for cyclophotocoagulation are generally limited to patients whose glaucoma has been resistant to medical and surgical therapies, with no potential for improvement in visual acuity.

8.2.2. Surgical treatment

The indication for surgical intervention is a lack of effectiveness of conservative and laser treatment [11,36]. An operative procedure should not be conducted too late due to the development of complications resulting from the persistence of the malignant process.

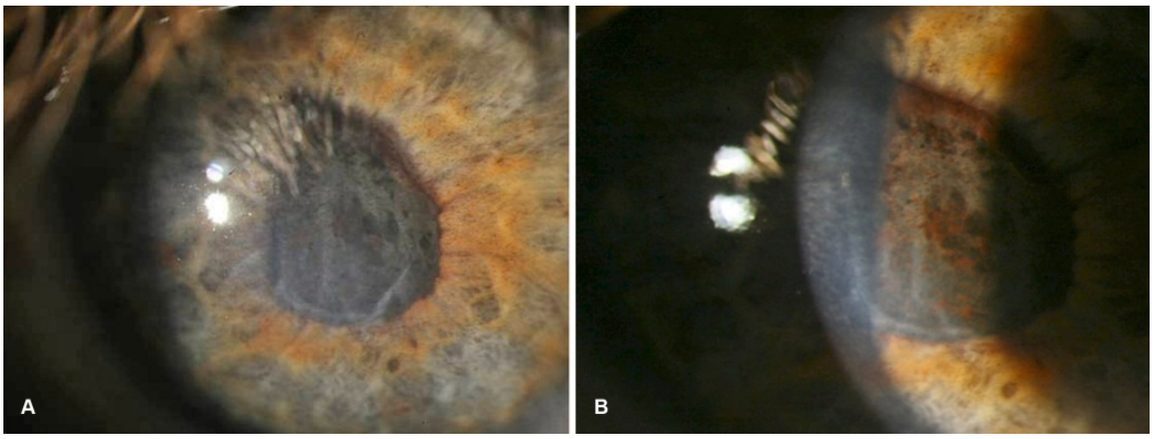


Figure 5. A,B: Advanced stage of malignant glaucoma - shallow anterior chamber, corneal oedema and posterior synechiae in pseudophakic eye.

Currently used methods of surgical treatment were introduced when the role of the pathology of the anterior segment and the vitreous body in the pathogenesis of the malignant process were discovered. As of now, surgical intervention in malignant glaucoma is directed towards lowering IOP, achieving correct anatomical relationships between the vitreous body, lens, and ciliary body, and additionally enabling correct flow of aqueous humour from the posterior segment to the anterior chamber of the eye. Achievement of communication seems to be necessary, because the disruption of aqueous humour flow in malignant glaucoma can last even after PPV [54]. The concept of such a procedure is based on the observation of regression of the symptoms of malignant glaucoma in the case of direct communication between the vitreous cavity and anterior chamber being ensured [25]. The iridectomy may be performed using Vannas scissors or a vitrectomy tip, whereas the posterior capsulotomy and hyaloidotomy may be done with a vitrectomy tip. The anterior chamber may be reformed with air. All of these procedures should be performed in one setting through the same location. Additionally synechiolysis may be performed if the iridocorneal angle is completely closed using a spatula or a viscoelastic agent. The performance of all

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three steps will usually result in complete resolution of the condition. Pars plana vitrectomy is reserved for cases that did not respond to the procedure above, and in any case, it should be combined with opening of the anterior hyaloid face. Thus, in refractory malignant glaucoma, partial PPV should be performed and supplemented by procedures making it possible to achieve communication between the anterior chamber and the vitreous cavity. Achievement of correct flow and equalization of pressure between the posterior and anterior segment of the eyeball is decisive for the effectiveness of the surgery. Partial PPV should be conducted conservatively, preferably using trocars and a 25 gauge vitrectome. Communication between the anterior chamber and vitreous cavity may be achieved by cutting out the lens capsule using a vitrectome or puncturing it through the cornea with a needle, alternatively by cutting the anterior and posterior capsules with cystotome from the side of the anterior chamber within iridectomy.

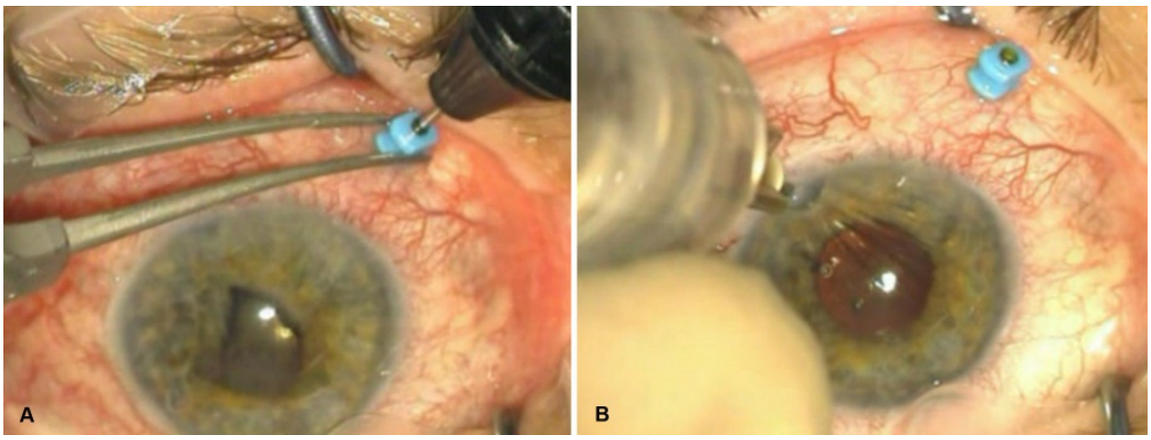

Figure 6. Combined partial pars plana vitrectomy with capsulotomy communicating anterior chamber and vitreous cavity in surgical treatment of malignant glaucoma: A. The trocar is inserted through pars plana 3.5 mm posteriorly to the corneal limbus before PPV. B. Achieving communication between the anterior chamber and vitreous cavity using a vitrectome.

9. Prognosis

Malignant glaucoma remains a difficult clinical problem that results in irreversible blindness if treatment is delayed and not adequate. The surgeon should be aware preoperatively of eyes at risk and observe them closely during follow-up visits. Early recognition is the most important step to prevent irreversible loss of vision. The prognosis depends on the duration and the severity of the malignant glaucoma attack. In patients with glaucoma in its early stage, the prognosis can be good if the attack is discontinued and IOP is well controlled. The problem is that malignant glaucoma is often resistant to conservative treatment, and laser procedures are not always effective as well. Partial pars plana vitrectomy combined with capsulotomy communicating the anterior chamber and vitreous cavity in such cases is an efficacious method of intervention when it comes to IOP control, postoperative BCVA, and reduction of the number of antiglaucoma medications. The prognosis after laser and surgical

treatment depends on the occurence of complications after performed procedures. Complications after malignant glaucoma surgery observed in own material included: increased IOP during the early post-operative period (above 21 mmHg) [5%), inflammatory effusion [5%), hyphema [10%), occurrence of posterior adhesions [5%), no effectiveness of filtration surgery preceding the occurrence of malignant glaucoma [55%), macular edema [10%), and retinal detachment [5%). Recurrence of malignant glaucoma with the full range of symptoms was observed in 15% of eyes subjected to surgery. In the case of post-operative shallowing of the anterior chamber, it is possible to conduct a capsulotomy through iridectomy, and the use of an Nd-YAG laser for this purpose is a safe and effective method in most cases.

10. Summary and conclusions

The condition continues to be one of the most difficult types of secondary glaucoma to manage. The diagnosis, definition, pathomechanism, and procedure in the case of malignant glaucoma still give rise to controversy. The currently applied treatment has the goal of unburdening the anterior chamber during the early period of the malignant process and to create communication between the anterior chamber and the vitreous. This is the result of the assumption, that in the case of a lack of communication through the iridotomy, recurrence of the malignant process can be expected after vitrectomy. New modifications of surgical procedure may increase operative effectiveness and improve the long-term results of applied procedures.

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