Received: 19 November 2023 | Accepted: 1 December 2023

DOI: 10.1111/aos.16599

Acta Ophthalmologica

PERSPECTIVE

Guidelines for the management of open-angle glaucoma

National Program Area Eye Diseases, National Working Group Glaucoma

Gauti Jóhannesson[1,2,3] | **Ulf Stille**[4] | **Amelie Botling Taube**[5] | **Markus Karlsson**[6] | **Lada Kalaboukhova**[7] | **Anders Bergström**[8] | **Dorothea Peters**[9] | **Christina Lindén**[1]

[1]Department of Clinical Sciences, Ophthalmology, Umeå University, Umea, Sweden
[2]Wallenberg Center for Molecular Medicine, Umeå University, Umeå, Sweden
[3]Department of Ophthalmology, University of Iceland, Reykjavik, Iceland
[4]Department of Ophthalmology, Falu Lasarett, Falun, Sweden
[5]Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden
[6]Department of Biomedical and Clinical Sciences, Linköping University, Linkoping, Sweden
[7]Department of Clinical Neuroscience, Institute of Neuroscience and Physiology, Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden
[8]Department of Clinical Sciences, Ophthalmology, Lund University, Lund, Sweden
[9]Department of Clinical Sciences, Ophthalmology, Lund University, Malmo, Sweden

Correspondence
Gauti Jóhannesson, Department of Clinical Sciences, Ophthalmology, Umeå University, Umea, Sweden.
Email: gauti.johannesson@umu.se

KEYWORDS
glaucoma, guidelines, management, Sweden

1 | PURPOSE

These guidelines provide a brief guide on how primary open-angle glaucoma, exfoliation glaucoma and pigment glaucoma can be diagnosed, treated and monitored. The aim is to nationally spread knowledge-based high-quality care, to stimulate the use of scientifically evaluated and effective measures, to even out differences in care and to provide support in setting priorities. Angle-closure glaucoma is not dealt with in this publication but is mentioned where relevant.

2 | METHOD DESCRIPTION—HOW THE GUIDELINES HAVE BEEN DEVELOPED

The National Working Group Glaucoma has made an inventory of applicable current international, Nordic and Swedish guidelines, care programmes and guidelines for glaucoma. Through the regional representatives, we have reviewed a majority of regional and local documents. Answers to specific questions have been sought in PubMed and via Cochrane reports. In compiling these guidelines, we have primarily based our work on the European glaucoma guidelines (European Glaucoma Society, 2021) (which are evidence-graded according to the AGREE system regarding so-called key questions 2020). For evidence grading (A–D), the Finnish guidelines from 2014 were also used (Tuulonen et al., 2014). Furthermore, the current document is based on the previous Swedish guidelines, Guidelines for Glaucoma Care from 2010 (Heijl et al., 2012). On the issues where there is insufficient scientific evidence, the group has discussed its way to a consensus on best clinical practice.

3 | BACKGROUND

The goal of all glaucoma treatment is to preserve the patient's visual function, well-being and related quality of life with a long-term sustainable use of resources. Treatment costs in the form of discomfort and side effects, as well as economic costs for the individual patient and society, should be taken into account. Quality of life is strongly linked to visual function and is affected differently depending on life situation. In general, patients with mild glaucoma damage have good visual function and a largely preserved quality of life, while quality of life is severely affected if both eyes have advanced vision loss that, e.g., affects driving.

Glaucoma treatment must be individualized and adapted to the patient, situation and available resources.

The increasing availability of randomized controlled clinical trials (RCTs) makes it possible to base clinical recommendations to a greater extent on scientific evidence. There are a number of RCTs that clearly show that intraocular pressure lowering treatment of glaucoma is effective. Glaucoma damage can be slowed down and, in the best case, stopped if patients are properly diagnosed, treated and followed up.

The degree of visual field damage and how it develops over time determines the individual treatment strategy (Figure 1). The goal is to maintain good quality of life without advanced loss of visual function throughout life.

3.1 | Definition and characteristics

Glaucoma is a progressive disease that causes typical damage to the optic nerve head, the retinal nerve fibre layer and the visual field. Intraocular pressure is not included in the definition.

In primary open-angle glaucoma, the chamber angles should be open and specific causes of glaucoma should not be identifiable. Subjects with primary open-angle glaucoma may have high (high tension glaucoma) or normal (normal tension glaucoma) intraocular pressure.

In exfoliation glaucoma, there are also protein precipitates and exfoliations. This type of glaucoma is also known as pseudoexfoliation glaucoma (PEX glaucoma) or capsular glaucoma. The exfoliation material is most easily visible on the front surface of the lens. In the Nordic countries, we often include exfoliation glaucoma in the group of primary glaucomas, while in other parts of the world, it is considered a secondary glaucoma.

Pigmentary glaucoma is a secondary glaucoma characterized by the Krukenberg spindle (pigment on the corneal endothelium), slits in the mid-periphery of the iris (transillumination defects) and a smooth, dark pigmented trabecular meshwork in the chamber angle. It is a rare form of glaucoma that typically affects younger people (20–50 years), often myopic and more common in men (5:1).

3.2 | Epidemiology

Glaucoma is one of the most common age-related eye diseases and among the leading causes of blindness in the world. In a Swedish study, 42% of patients with open-angle glaucoma became blind in one eye and 16% in both eyes during their lifetime.

The disease is uncommon before the age of 40 and increases with age. It is estimated that the global prevalence of open-angle glaucoma is 3.5% in the 40–80-year-old group and 0.5% for angle-closure glaucoma. In 2014, the number of subjects with glaucoma was estimated to reach 76 million by 2020 and is expected to increase to 112 million by 2040.

The large Swedish screening study, Malmö Eye Survey, showed a glaucoma prevalence of just over 5% in 75-year-olds or about 2% in the age group 57–79 years. Smaller Swedish studies have shown both higher and lower prevalence. Varying prevalence of exfoliation syndrome in the population could help explain differences in the prevalence of glaucoma. Exfoliation syndrome is common in Sweden, especially in women, and its prevalence increases considerably with age.

> In the context of glaucoma, the terms exfoliation, pseudo-exfoliation (PEX) and exfoliation syndrome are often used interchangeably.

3.3 | Resource requirements

A Swedish study published in 2008 showed that about 25% of all visits to Swedish ophthalmic care providers were glaucoma-related.

The number of patients in Sweden with diagnosed glaucoma is not known but has previously been estimated at around 100 000. However, this figure is very uncertain. Based on prescriptions in Sweden in 2008 and 2017, the number of unique individuals treated with intraocular pressure-lowering drops was found to be 144 000 and 172 000, respectively, an increase of almost 20% in 9 years. This increase is expected to continue.

In addition to patients with diagnosed glaucoma and treated ocular hypertension, many patients with risk factors for developing glaucoma are also checked at Swedish eye clinics, such as untreated ocular hypertension, exfoliations, pigment syndrome and more. This means that significantly more patients than the 172 000 are covered by the guidelines and undergo examination at Swedish eye clinics.

Life expectancy in Sweden has increased significantly in recent decades and with it the proportion of older people in the population. According to Statistics Sweden's population projection for 2019, the number of inhabitants over the age of 80 will increase by over 50% between 2019 and 2030. Several studies predict a similar percentage increase in the number of patients monitored and/or treated in glaucoma care by 2040. In order to maintain a decent level of glaucoma care, significant resources must therefore be added.

3.4 | Symptoms

Open-angle glaucoma usually does not cause symptoms until at late stages. The elevated eye pressure is usually painless. The loss of visual field usually affects the paracentral visual field first and is difficult to detect for the patient.

In connection with pigment release in people with pigmentary glaucoma, acute pressure increase may occur. At the same time, corneal oedema can cause blurred vision and halo phenomena.

3.4.1 | Normal and elevated intraocular pressure

Traditionally, the upper limit of normal intraocular pressure has been considered to be ≤21 mm Hg. It is based on several population studies where the average pressure in

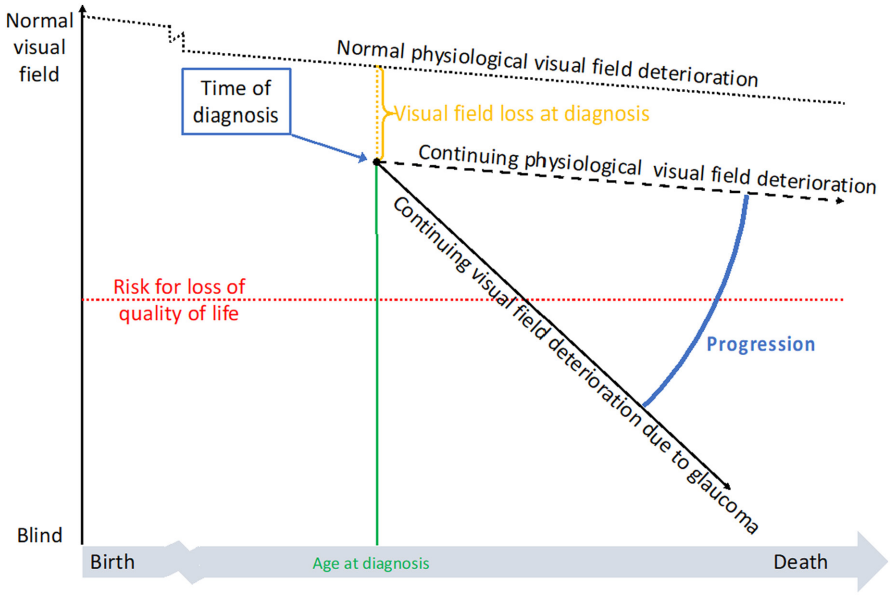

FIGURE 1 Schematic diagram of visual field deterioration in support of individualized treatment and follow-up. Note that the data in many visual field devices are age-corrected, which means that the curve regarding physiological visual field deterioration is straight. The figure is inspired conceptually by a similar figure in the EGS Guidelines page 32, 2020, www.eugs.org (European Glaucoma Society, 2021).

the adult population is about 16 mm Hg and has a standard deviation of about 2.5 mm Hg. However, this pressure level does not say anything about what eye pressure is harmful at the individual level, since some people suffer injuries at significantly lower levels, and some can tolerate higher levels. Thus, from a functional point of view, all pressure levels that do not give rise to glaucoma damage are 'normal' in the individual.

3.5 | Risk factors

There are a number of factors that increase the risk of developing and/or worsening glaucoma. Knowledge of risk factors is of great importance in order to identify persons who may require (more frequent) monitoring or (more vigorous) treatment, at least until the individual rate of progression is known. Several risk factors in the same person further increase the risk.

I. Risk factors for the development of open-angle glaucoma

1. Individual
   a. Older age—open-angle glaucoma is very rare before the age of 40 and uncommon before the age of 50. Glaucoma prevalence: >40 years—1%, >75 years—5%.
   b. A positive family history—glaucoma in first-degree relatives (siblings/parents).
   c. Descent—people of non-Caucasian, mainly African, origin.
2. Eyes
   a. Elevated intraocular pressure is the most important risk factor for the development of glaucoma. The higher the eye pressure, the greater the risk.
   b. Exfoliation syndrome in combination with increased intraocular pressure.
   c. Moderate to high myopia (-3D or more).
   d. Optic disc haemorrhages.
   e. Pigment dispersion syndrome.
   f. Thin central corneal thickness (CCT) in eyes with increased intraocular pressure. The thinner the cornea, the higher the risk. CCT is particularly useful in patients with ocular hypertension when treatment and/or follow-up visits are considered.
3. General diseases
   a. Low diastolic blood pressure, approximately 60 mm Hg or lower.

For other potential risk factors, such as cardiovascular diseases, diabetes mellitus, migraines, Raynaud's phenomenon and sleep apnea, the evidence regarding association with glaucoma is more questionable.

> Do not correct intraocular pressure with CCT algorithms.

> Rule of thumb: thin CCT < 500 µm, thick CCT > 600 µm.

II. Risk factors for glaucoma progression

1. Individual
   a. Older age.
2. Eyes
   a. Elevated intraocular pressure is the most important risk factor for glaucoma progression—the higher the intraocular pressure, the greater the risk of progression.

b. Exfoliation syndrome—independent of intraocular pressure, the risk of glaucoma progression increases.
c. Optic disc haemorrhages.
d. The thinner the CCT, the higher the risk in eyes with increased intraocular pressure. CCT may be useful when normal tension glaucoma is progressing.
e. Advanced visual field damage at diagnosis.

It is unclear whether cardiovascular diseases also affect the risk of glaucoma progression.

4 | EXAMINATIONS/DIAGNOSIS

The following examination and patient history data should be included in the primary assessment of patients with manifest as well as suspected glaucoma, or with risk factors for developing glaucoma. These parameters then form the basis for decisions on how any continued follow-up and treatment should be designed.

4.1 | History

- Family history of glaucoma (first-degree relatives – siblings/parents).
- Medications, such as blood pressure medications and steroids.
- Previous eye trauma or inflammation.
- Prior refractive surgery (affecting CCT).
- Other diseases, such as asthma/chronic obstructive pulmonary disease and cardiovascular disease.
- Hypersensitivity to drugs.

4.2 | Clinical examinations

- Visual acuity and refraction.
- Intraocular pressure.
- CCT.
- Gonioscopy.
- Slit lamp examination focusing on chamber depth, exfoliation syndrome, pigment dispersion syndrome, iris configuration, iris slits, rubeosis of the iris, inflammation and cataract.
- Visual field status (monocular with computerized perimetry).
- Optic disc examination with dilated pupil. Avoid dilation with narrow chamber angles.
- Optic disc photograph as a reference image for future comparisons.
- Optical Coherence Tomography (OCT) of the optic disc, peripapillary nerve fibre layer and/or ganglion cell layer in the macula is of limited value and cannot replace perimetry. OCT does not have to be performed but can be of value as a complement in the diagnostic workup.
- Pupil reflexes with the swinging flash test. A relative afferent pupillary defect (RAPD) can indicate glaucoma.

> OCT abnormalities alone are not sufficient to make a diagnosis of glaucoma.

The above initial examination package is extensive, but of value in order to be able to perform an adequate assessment and decide on further checks and treatment. Patients with suspected or manifest glaucoma are checked regularly over a long period of time and a proper investigation helps to avoid both over- and under-diagnosis.

All examinations do not necessarily have to be carried out at the same time. Rather, it may be valuable to divide these, for example into a visit to the doctor and one to a nurse, optician or optometrist. The time interval between these initial assessments is determined by the level of eye pressure and other risk factors. To get a good idea of the untreated pressure before starting treatment, at least two, but preferably three, separate pressure measurements are recommended. These are often done at different times of the day. At very high-pressure levels, treatment can be started at the first visit, but even in these cases, it should be preceded by at least two separate pressure measurements, preferably performed by different examiners and with different Goldmann tonometers.

4.3 | Methods of diagnosis and examination

4.3.1 | Measurement of intraocular pressure

Goldmann applanation tonometry (GAT) is the standard method for measuring intraocular pressure in glaucoma care. The calibration of the GAT tonometer shall be checked regularly in accordance with the manufacturer's recommendations, which in the case of Haag-Streit tonometers, for example, means at least once a month. Other measurement methods such as rebound tonometry (Icare) can be used in patients who have difficulty undergoing GAT tonometry (e.g. young children) or in screening situations. Non-contact tonometry (NCT) should be avoided if other alternatives are available, as the measurement method is subject to large variations of measurement results.

> Follow each patient using the same pressure measurement method – GAT is standard.

> Intraocular pressure should be checked on **at least** 2 separate occasions (preferably at different times during the day) before any pressure-lowering treatment is initiated and further follow-up is planned.

4.3.2 | Visual field examination

Glaucoma causes damage to the visual field, primarily within the central 20°. The standard measurement method for the diagnosis and follow-up of glaucoma is Standard Automated Perimetry (SAP). SAP is a computerized static visual field examination (perimetry) based on the principles of Goldmann perimetry, with white stimuli on a white background. Static (stimuli in predetermined test locations) threshold measuring perimetry is more sensitive and detects glaucomatous visual field damage earlier compared to manual kinetic (with moving stimuli) perimetry (Goldmann perimetry), which should therefore not be used. In addition, computerized perimetry is less subjective, the results are numerical, and there are software programmes that assist in the assessment of the results. The most common perimeter in Sweden is the Humphrey perimeter, but Octopus and other perimeters are also used.

If there is only a low suspicion of glaucoma, for example when checking patients with a positive history of glaucoma, screening tests, suprathreshold tests that use bright stimuli that are well above the threshold level, should be used. These are easier to perform for patients with no prior experience with computer perimetry and generate fewer false positives. In the Humphrey perimeter, suprathreshold tests with the C-40 pattern and the 2-zone strategy are primarily chosen as screening tests. Another method called Frequency Doubling Technology (FDT) has a couple of screening programmes. C20-1 has the best combination of sensitivity and specificity and is therefore more suitable for screening.

In case of greater suspicion of glaucoma, threshold programmes are used. These quantify the field of view. It is desirable to use the same program and the same test point pattern in the diagnosis and follow-up of glaucoma patients. Suitable applications include, for example, the Humphrey perimeter's SITA Standard, SITA Fast or SITA Faster or the Octopus perimeter's Normal Strategy or Dynamic Strategy.

There are various indices that summarize the results of the visual field examination. The Humphrey perimeter calculates mean deviation (MD), which is the average deviation in dB from the age-adjusted normal field of view, and the visual field index (VFI), which is expressed as a percentage of a normal field of view. VFI is most often used nowadays. It is significantly less sensitive to cataract development than MD and weights central parts of the visual field higher. The mean defect (also abbreviated MD) calculated in the Octopus perimeter is similar to mean deviation, but note that the signs are different in the two instruments. Damage has a negative designation in the Humphrey perimeter (e.g. −5 dB) and positive in the Octopus perimeter (e.g. +5 dB). Also note that the results from the Humphrey and Octopus perimeters are not directly comparable as the basis of the decibel scale differs between the instruments, 10 dB in Humphrey corresponds to 6 dB in Octopus.

In the Humphrey perimeter, VFI can be plotted over time and a coefficient describing the average rate of deterioration per year is calculated to illustrate the Rate of Progression (RoP). Other perimeters have similar indexes (for example, MD slope in the Octopus) that can be used to calculate the rate of progression.

In the Humphrey perimeter, the SITA Faster program is the first choice for the vast majority of glaucoma patients when choosing a testing strategy. Thorough instructions and information are needed for everyone undergoing visual field examinations, especially if SITA Faster is being used for the first time. Please note that SITA Faster uses stimuli that are very close to the presumed threshold for detection and are thus more difficult to perceive compared to SITA Fast and SITA Standard. SITA Fast may therefore be a more suitable option for patients who have difficulty completing SITA Faster.

In the case of severely reduced visual acuity, such as in macular degeneration, it is sometimes necessary to use another fixation light, called a large diamond (in Octopus called a ring). In the case of advanced visual field damage, it may be useful to focus more on the central part of the remaining visual field (e.g. visual field programme 10–2 in the Humphrey perimeter). There are no follow-up programmes for these test strategies.

4.3.3 | OCT in glaucoma

Optical Coherence Tomography (OCT) is a non-invasive method whose technique is based on the analysis of light reflected from the retina. In this way, the different layers of the retina can be visualized. In glaucoma diagnostics, it is primarily the retinal nerve fibre layer and the ganglion cell layer that are of interest. The OCT examination is easy to perform and is used frequently but cannot replace clinical assessment and/or visual field examination.

Digital imaging and automatic image analysis with OCT can be helpful in diagnosing glaucoma provided that the examination is of good quality. However, the risk of artefacts that could lead to misinterpretation of the OCT results is significant. The results of the investigation should be reviewed for any sources of error before assessment. Repeated OCT examinations may show false positive progression, which means that the benefit of OCT in assessing glaucoma progression is limited. Also, the so-called floor effect, which can occur already in patients with moderate visual field damage, complicates progression assessment with OCT.

In the case of optic discs that are difficult to assess, for example in cases of severe myopia or pronounced peripapillary atrophy, analysis of the ganglion cell layer in the macula may be preferable to measurements of the peripapillary nerve fibre layer.

4.3.4 | Optic disc photograph/optic disc assessment

A clinical examination of the optic disc is necessary for the diagnosis and follow-up of glaucoma. When diagnosing, it is advisable to take a good-quality optic disc photograph for future comparison. Optic disc size must be taken into account in particular when assessing the

cupping. Both small and large discs can be difficult to assess from a glaucoma point of view.

Measuring the vertical optic disc diameter through a lens in the slit lamp can provide guidance in assessing disc size. For example, if a 60 D Volk lens is used, <1.65 mm is considered a small disc and >2.2 mm is a large one. Another simple method is to compare the disc diameter (DD) with the disc-fovea distance (DM), measured from the centre of the disc to the fovea. The mean DM/DD ratio is 2.5. With a ratio < 2.0, the disc is large.

The ISNT rule can be used to assess whether the thickness of the nerve fibres follows the normal distribution of nerve fibre bundles. Usually, the widest rim is found inferiorly, followed by superiorly, nasally and temporally in descending order.

4.4 | Diagnostic criteria and codes

4.4.1 | Ocular hypertension

Elevated intraocular pressure without damage to the visual field, optic disc or nerve fibre layer is referred to as ocular hypertension (OH). This is usually defined as intraocular pressure > 21 mm Hg.

4.4.2 | Glaucoma

A confirmed diagnosis of glaucoma requires an eye disease with characteristic progressive optic nerve damage with a corresponding visual field defect.

Establishing the diagnosis is often easy but can sometimes be very difficult. Corresponding optic nerve and visual field damage, often in combination with elevated intraocular pressure, is a classic combination. For a completely reliable diagnosis, one should also require progression of the damage. In everyday clinical practice, one rarely waits for this. If strict glaucoma criteria are used in diagnosis, it is also not necessary. Abnormal optic disc appearance, other eye diseases, sensitivity of methods, lack of patient compliance, etc. are other factors that often complicate and/or affect making a diagnosis.

The perimetric rate of progression at the group level varies, depending on the type of glaucoma, among other things. For example, exfoliation glaucoma usually has a more aggressive course compared to normal tension glaucoma. Even without treatment, visual field deterioration is often slow in subjects with normal tension glaucoma. Pigmentary glaucoma differs from others in that it often involves higher and more fluctuating intraocular pressure levels in the initial stages. Therefore, correct diagnosis is crucial for planning the continued care.

If there is uncertainty as to whether glaucoma is present, one can wait and follow the development in most patients, with the possible exception of young patients or patients with very high intraocular pressures. If the intraocular pressure is normal or only slightly elevated, there is even more reason to postpone treatment.

A diagnosis affects both the patient and the healthcare system. A diagnosis on dubious grounds often causes unnecessary anxiety as well as incorrect use of resources and should therefore be avoided. In case of doubt, the most appropriate diagnosis is used, which is then re-evaluated when more information is obtained. A uniform approach to diagnosis is preferred, see Tables 1 and 2 for diagnosis codes for all glaucoma types with comments.

- Avoid using non-specific diagnoses such as H40.0X Suspected glaucoma, unspecified, H40.1 Chronic open-angle glaucoma, H40.1X Primary open-angle glaucoma, unspecified and H40.9 Glaucoma, unspecified.
- If a patient with a primary open-angle glaucoma (H40.1A and H40.1D) develops exfoliation syndrome, the diagnosis is changed to exfoliation glaucoma regardless of the pressure level.

5 | DIFFERENTIAL DIAGNOSES

With typical glaucomatous optic nerve injury with corresponding visual field defect, no further investigation with radiology is usually needed, even if the intraocular pressure is normal. However, it is not uncommon for neurological conditions that cause optic nerve or chiasmal compression to be misdiagnosed as glaucoma. Thus, visual field defects without typical glaucomatous optic nerve damage should prompt radiological investigation.

There can be several reasons for the discrepancy between visual field and optic disc appearance. The following may be helpful:

5.1 | Visual field damage with a normal optic nerve

Although glaucoma causes typical visual field damage that often starts nasally and forms an arcuate defect in the upper or lower half of the visual field, a glaucomatous visual field defect can have different appearances. If the visual field damage appears to be glaucomatous but the optic nerve appears normal, the following should be taken into account:

- Is the optic disc small? In such a case, even a small cup can be glaucomatous and have a clear visual field defect.
- Are there optic disc drusen?
- Is there a retinal disease that can cause visual field damage?
- Is there severe myopia? Atrophic areas in these eyes can cause visual field defects.
- Is there a nerve fibre layer defect that can explain the visual field defect?
- Has the patient understood the visual field examination? Can the visual field defect be verified at a follow-up examination? Was the lens correction correct? Incorrect lens correction can cause a general reduction in the field of vision, but not localized defects as in glaucoma.

TABLE 1 Suspected glaucoma (including risk factors).

Diagnosis code	Diagnosis	Comment
H40.0	Suspected glaucoma	Only used in case of suspected glaucoma under investigation. To be replaced by a defined diagnosis as soon as possible
H40.0A	Ocular hypertension	Pressure > 21 mm Hg without treatment. Normal visual field, optic disc and nerve fibre layer
H40.0B	Narrow chamber angle	
H40.0C	Suspected optic disc cupping	
H40.0D	Exfoliation syndrome	
H40.0E	Pigment dispersion syndrome	Iris slits, Krukenberg spindle, pigmentation of the trabecular meshwork
H40.0 W	Other type of suspected glaucoma	For example, pressure difference between the eyes, optic disc haemorrhage and more
Z 83.5	Positive family history for glaucoma	In sibling or parents

TABLE 2 Manifest glaucoma.

Diagnosis code	Diagnosis	Comment
H40.1A	Primary open-angle glaucoma	Untreated pressure > 21 mm Hg with manifest damage. Also referred to as high-tension glaucoma or simple glaucoma
H40.1B	Pigmentary glaucoma	
H40.1C	Exfoliation glaucoma	Sometimes referred to as pseudoexfoliation glaucoma, formerly also capsular glaucoma.
H40.1D	Normal tension glaucoma	Untreated pressure ≤ 21 mm Hg (occasional pressure readings up to 24 mm Hg)
H40.1 W	Other specified form of open-angle primary glaucoma	
H40.2	Angle closure glaucoma	
H40.3	Glaucoma secondary to ocular trauma	
H40.4	Glaucoma secondary to ocular inflammation	
H40.5	Glaucoma secondary to other eye diseases	For example, glaucoma after congenital cataract surgery, aniridia
H40.6	Glaucoma secondary to drugs	
H40.8	Other forms of specified glaucoma	For example, glaucoma in Nail-Patella syndrome, Sturge–Weber syndrome
H42.0	Glaucoma secondary to endocrine and metabolic diseases	For example, glaucoma in amyloidosis
H42.8	Glaucoma secondary to other diseases classified elsewhere	For example, glaucoma in neurofibromatosis
Q15.0	Congenital glaucoma	
H44.5	Absolute glaucoma	Blind eye

- If a relative afferent pupillary defect (RAPD) with swinging flashlight test is present, this supports a neuronal injury. RAPD is seen in asymmetric optic nerve involvement. Since the glaucoma damage is often asymmetric, RAPD can often be detected even in bilateral disease.

5.2 | Optic nerve damage with normal visual field

When the disc looks glaucomatous, but the field of vision is normal, the following should be taken into account:

- Is the disc large? Large normal discs have a large cup. Look for findings that support glaucoma diagnosis such as a disc haemorrhage, notch formation and cupping reaching the disc margin.
- Has the patient understood and was able to comply with the visual field-testing instructions? Abnormally high dB numbers or high numbers of false positives can mask a visual field defect. Can the visual field defect be verified at a follow-up examination?
- Could it be a precursor to glaucoma where visual field damage has not yet developed?

6 | TREATMENT

The goal of all treatment for glaucoma is to preserve the patient's visual function, well-being and quality of life with a long-term sustainable use of resources. The most important factor to consider when choosing a treatment strategy is to evaluate the patient's risk of developing visual impairment and vision-related quality of life reduction

during their remaining lifetime. Factors that increase the risk are long life expectancy, major damage at diagnosis and disease in both eyes. Thus, younger persons with mild bilateral visual field impairment have significantly higher risks of developing quality-of-life-affecting visual impairment during the remainder of their life compared to older persons with moderate visual field damage in one eye. In addition, the rate at which the disease worsens (RoP) varies from person to person. It is therefore important to individualize treatment. When choosing therapy, other factors should also be taken into account such as side effects, quality of life, compliance and cost.

All available treatments, whether pharmacological or laser and surgery, are aimed at lowering intraocular pressure, either by reducing the production of aqueous humour or by increasing the outflow via the trabecular meshwork or the uveoscleral pathway.

6.1 | Target pressure

A target pressure should be set. Target pressure is the upper pressure limit for each eye that is currently accepted and at which the rate of deterioration is deemed acceptable and consistent with a satisfactory quality of life of the patient.

At diagnosis, the rate of deterioration is unknown. A rule of thumb (non-evidence-based) that is sometimes used before the rate of progress is known is based on:

- In case of early damage (≤6 dB) – 20% pressure reduction, but at least to 18–20 mm Hg.
- In case of moderate damage (>6–12 dB) – 30% pressure reduction, preferably to 15–17 mm Hg.
- Advanced damage (>12 dB) may require even lower target pressures, such as 10–12 mm Hg.

The principle is that the greater the damage, the longer the life expectancy and the lower the untreated eye pressure, the lower the target pressure.

More damage and high eye pressures require adequate target pressure to be achieved without delay. The presence of exfoliation syndrome warrants extra vigilance.

> Target pressure is continuously evaluated and adjusted depending on the development of the visual field damage.

If the rate of deterioration (RoP) is too high, the target pressure is lowered. In the case of slow visual field progression but not achieved target pressure, the target pressure may be adjusted upwards, especially if the treatment is not well tolerated.

6.2 | General management principles

- Start with monotherapy (or with laser trabeculoplasty).
  - Prostaglandin analogues have a good pressure-lowering effect, are generally well tolerated and should be used as first-line drugs.
  - Beta-blockers have an almost equally good pressure-lowering effect but more systemic side effects and are also a possible first choice.
- If there is no or little effect from the prescribed drug, the treatment should primarily change to a different monotherapy.
- If the effect of the drug is good but insufficient, an additional substance is added. This substance should be from a different class of drugs and preferably with a different mechanism of action. For good adherence, a fixed combination is usually chosen.
- If the pressure reduction is still not sufficient, a third substance from a third group of preparations is added.
- Pressure reduction of additional drugs is uncertain and surgical alternatives should be considered instead. If treatment is initiated with a fourth or fifth drop or additional laser trabeculoplasty (LTP), a clear follow-up plan should be in place to ensure that the effect is achieved and that any necessary surgery is not unduly delayed.
- Laser trabeculoplasty can be considered at any level in the treatment ladder, even as a first-line treatment.
- Please note that preservatives can be a cause of eye irritation, ocular surface disease (OSD) and allergies or otherwise cause problems. In this case, use alternatives without preservatives.
- Due to frequent side effects, oral carbonic anhydrase inhibitors (e.g. acetazolamide) are generally only used to lower intraocular pressure temporarily while awaiting surgery. Acetazolamide is also used chronically by a few patients where other treatment alternatives are missing.

> Identify a rapid rate of progression early on and consider surgery in these cases. Many patients undergo surgery too late.

A schematic overview of the step-wise increase of treatment is shown in Figure 2.

6.3 | Pharmacological treatment

There are six different classes of intraocular pressure-lowering agents, see Table 3 below. It also shows which substances belong to each group, mechanism of action, approximate pressure reduction and some significant and/or common side effects. In the event of a change in pharmacological treatment, intraocular pressure is usually checked within 1 month. In high-risk patients or patients with very high intraocular pressures, the interval may need to be shortened. For advice on the practical approach to initiating and changing treatment, please refer to the section on general treatment principles above.

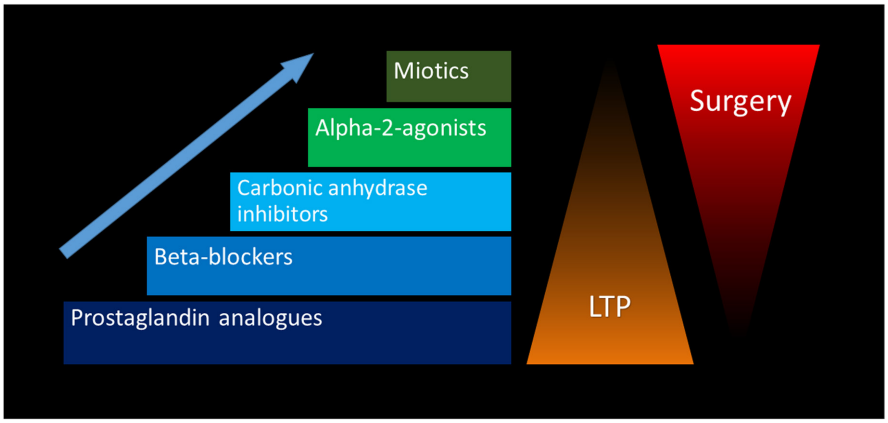

FIGURE 2 Treatment ladder. Schematic picture of successive increases in pharmacological treatment with eye drops, but rarely/never all types and not always in the order of the image. LTP and surgery can be performed at any stage. LTP is usually performed early in the treatment arsenal, even as first-line treatment. Surgery is usually done only after visual field progression, despite previous treatment with laser and drops.

6.4 | Laser trabeculoplasty

In the late 70s, when laser treatment of the chamber angle, laser trabeculoplasty (LTP), was introduced, argon lasers were used and the treatment was therefore referred to as ALT. Although other lasers came into use, the name was kept or the treatment was simply called LTP. Nowadays, a Q-switched, frequency-doubled YAG laser is most often used and selective laser trabeculoplasty (SLT) is performed. It is a gentler treatment that has fewer side effects, is easier to use and has increased repeatability. Both treatments have comparable efficacy and lower intraocular pressure approximately as much as an eye drop. At higher eye pressures, a greater effect is achieved. The effect wears off over time. At low-pressure levels (<15 mm Hg), LTP often has no or poor effect. ALT has less effect in sparsely pigmented eyes, while SLT appears independent of pigmentation.

LTP can be used either as a primary treatment or as adjunct therapy.

Contraindications include neovascular and traumatic glaucomas, as well as glaucoma due to uveitis, narrow chamber angle or malformations of the chamber angle.

6.4.1 | Procedure and follow-up

The laser treatment is done under topical anaesthesia and with a special gonioscopy lens. At SLT, one aims at the trabecular meshwork, and at ALT the aim is to place the effects in the anterior part of the pigmented trabecular meshwork (Table 4).

If the chamber angle is heavily pigmented, as in pigmentary glaucoma, lower energy is often used and a smaller area is treated, such as a quadrant of the chamber angle. For patients with very severe glaucoma damage or otherwise when it is considered important to avoid a possible pressure peak, an α-agonist drop or other pressure-lowering treatment can be given in connection with the laser procedure.

There are a few complications to LTP. Sometimes iritis occurs, in most cases low-grade. This usually responds well to steroids, such as dexamethasone eye drops three times daily for a week. In most cases, such treatment is not given prophylactically.

Inform the patient to continue with existing eye drops if such are used, and to seek medical attention in case of eye pain the next day. Follow-up pressure control and evaluation of effect takes place after approximately 6 weeks. High-risk patients are managed individually.

If the previous SLT treatment has been effective, additional laser treatment of the same area may be considered a second time. There is a lack of evidence for more re-treatments. In the case of ALT, re-treatment of the same area is often discouraged, as it is considered to cause more complications.

6.5 | Surgical treatment

6.5.1 | General information about surgical treatment

If pharmacological and/or laser treatment does not provide sufficient pressure-lowering effect and the rate of progression is unacceptable, there is an indication for surgery. This is the main indication for pressure-lowering surgery. Since surgery is not dependent on adherence to medical treatment, non-adherence may in some cases be a relative indication. Pressure-lowering surgery is not the first choice for glaucoma treatment because there is a risk of serious sight-threatening complications during and after the procedure. Surgery provides more effective pressure reduction than medical and laser treatment. A relatively early decision on pressure-lowering surgery is therefore important and should be considered whenever medical and/or laser treatment is deemed incapable of maintaining visual function and not as a last resort, especially in cases with initially rapid progression or high intraocular pressure. Surgery should also be considered if adequate pressure reduction is not achieved with three pharmacological substances. The ophthalmologist should always make an individual assessment and discuss the benefits and risks of surgery with the patient.

The aim of surgery is to reduce intraocular pressure so that the patient has good pressure control without medical

TABLE 3 Overview of drug classes and substances.

Class	Substance	Mechanism of action and dosage	Pressure reduction	Some common/important side effects, possible comments
Prostaglandin analogues	Latanoprost 0.005% Travoprost 0.003% and 0.004% Bimatoprost 0.01% and 0.03% Tafluprost 0.0015%	Uveoscleleral outflow ↑ Dosed × 1	25%–35%	Hyperaemia, stinging, increased pigmentation (periorbital, lashes, iris), eyelash changes, orbitopathy, uveitis, macular oedema
	Latanoprostenbunod 0.024%	Uveoscleleral and trabecular outflow ↑ Dosed × 1		Latanoprostenbunod is not yet approved in Sweden
β-receptor blockers	Non-selective: Timolol 0.1% and 0.5%	Aqueous humour production Dosed × 1–2	Just over 25% Approx. 20%	Local: hyperemia, punctata Systemic: bradycardia, mental disorders such as depression and nightmares, erectile dysfunction and more
	β1-Selective: Betaxolol 0.25% and 0.5%			NB! Cave asthma, AV block and more Systemic side effects may be less pronounced for betaxolol
Carbonic anhydrase inhibitors Topical	Dorzolamide 2%	Aqueous humour production ↓	Approx. 20%	Stinging, punctate cornea, bitter taste
	Brinzolamide 1%	Dosed × 2(−3)		
Carbonic anhydrase inhibitors Systemic	Acetazolamide, tablets and injection	Aqueous humour production ↓ Dose: individual 250–500 mg × 2–3	Approx. 30%–40%	Paraesthesia, fatigue, nausea, loss of appetite is common. Electrolyte imbalances, kidney stones, aplastic anaemia NB! Cave sickle cell anaemia!
α_2-selective adrenergic agonists	Apraclonidine 0.5% and 1%	Aqueous production↓ Dosed × 3 (0.5%). For more info and for 1% see Swedish Medical Product Agency	25%–35%	Local: allergy common (apraclonidine > brimonidine), hyperemia, conjunctival paleness, eyelid retraction, mild mydriasis (apraclonidine) Systemic: dry mouth, somnolence
	Brimonidine 0.2%	Aqueous humour production ↓ (+ uveoscleral outflow ↑?) Dosed × 2	18%–25%	NB! Contraindicated in children or people on MAO inhibitors
Cholinergics	Pilocarpine 2%–4%	Trabecular outflow ↑ Dosed × 3	20%–25%	Local: miosis, myopia (accommodation paralysis). Systemic: headaches
Rhokinase inhibitors	Netarsudil 0.02%	Trabecular outflow ↑ and episcleral venous pressure ↓ (+ aqueous humour production↓?) Dosed × 1	20%	Hyperemia common, cornea verticillata, subconjunctival haemorrhages
	Netarsudil + latanoprost 0.005%	As above + uveoscleral outflow ↑ Dosed ×1	Better pressure reduction than the individual components	

treatment. However, supplementation with pressure-lowering drugs may be necessary to obtain optimal intraocular pressure. Indications for different pressure-lowering surgical techniques vary and depend on several factors. Factors influencing the choice of surgical technique include:

- Type of glaucoma.
- Target pressure.
- Previous medical history: amount of visual field damage, medication, previous eye surgery.
- Risk profile: single eye, refractive error.
- Patient preferences, expectations and ability for postoperative adherence.
- The surgeon's experience and preferences.

6.5.2 | Glaucoma in children

Primary congenital glaucoma and other secondary glaucomas in children require highly specialized care.

TABLE 4 Laser trabeculoplasty settings.

	ALT	SLT
Laser	Argon lasers, diode lasers or equivalent	Q-switched, frequency-doubled YAG laser
Spot size	50 μm	400 μm
Duration	0.1 s	3 ns (fixed)
Power/energy	500–1200 mW (depending on instrument and pigmentation)	0.4–1.2 mJ; start at 0.8 mJ, titrate to the highest energy that does not produce gas bubbles, so-called champagne bubbles
Optimal response	Single Gas Blisters	See above
Number of effects/ treatment	50 in 180° or 100 in 360° evenly distributed effects	50 in 180° or 100 in 360° non-overlapping effects

In primary congenital glaucoma, trabeculotomy is usually performed.

6.5.3 | Surgical techniques

The scientific evidence regarding glaucoma surgery is inadequate as controlled randomized trials are often lacking. Surgical techniques can be grouped as follows:

1. Trabeculectomy

Trabeculectomy is the most common surgical technique in glaucoma and is considered the gold standard in glaucoma surgery. The pressure-lowering effect is achieved by creating a new aqueous humour pathway from the anterior chamber via an opening in the iridocorneal angle and passage under a scleral flap to the subconjunctival space. The long-term pressure-lowering effect of trabeculectomy on previously non-operated eyes is good, but criteria for defining satisfactory outcome (success rate) vary in different studies.

2. Non-Penetrating Surgical Techniques

Deep sclerectomy, canaloplasty and viscocanalostomy are techniques that are considered to have smaller risks of postoperative complications. However, the pressure-lowering effect is not as effective as after trabeculectomy.

3. Glaucoma Shunt Surgery with Long Tube

Surgery with glaucoma drainage devices such as Molteno, Baerveldt or Ahmed are used in glaucoma with a high risk of failure with conventional filtering surgery, see Table 5. In some cases, glaucoma drainage devices may be used as a first-line surgical procedure.

4. MIGS

Several surgical techniques have been developed to reduce complication risks and provide faster recovery compared to conventional filtering surgery. These techniques are collectively referred to as minimally invasive glaucoma surgery (MIGS). The MIGS techniques are classified according to whether subconjunctival/transscleral filtration occurs, whether Schlemm's canal is treated by drainage/bypass/expansion, or whether a suprachoroidal drainage is constructed, with or without implants. Some of the methods can be combined with cataract surgery and the pressure-lowering effect of MIGS cannot be separated in the individual patient from the pressure reduction that the cataract surgery itself provides. MIGS tend to have a moderate pressure-lowering effect and may reduce medical treatment, according to some studies. MIGS may therefore be suitable in early or moderate glaucoma and be indicated at an earlier stage of glaucoma disease than other filtering procedures. Thus, the goal of MIGS is not the same as that of other filtering glaucoma surgery. There is currently no evidence that any of the techniques are better than traditional filtering surgery. The available data do not provide sufficient information on long-term effects and safety.

TABLE 5 Conditions with a high risk of poorer outcome of filtering surgery.

Conditions with a high risk of poorer outcome of filtering surgery
• Child.
• African descent.
• Inflammatory eye disease.
• Neovascular glaucoma.
• Long-term use of topical medical treatment.
• Previous complicated cataract surgery.
• Recent intraocular surgery (<3 months).
• Previous interventions in the conjunctiva.
• Aphakia.
• Previously failed filtering surgery.

5. Cyclo-destructive procedures

Cyclo-destructive procedures are indicated when filtering surgery has not worked, is not considered effective enough, or is not feasible. Cyclo-destructive procedures may also be indicated on blind eyes to reduce pain caused by high intraocular pressure. The ciliary body is partially destroyed by these treatment methods, which can be performed by laser, cryo or ultrasound technology. Transscleral cyclodiode laser technology is commonly used in Sweden. In endoscopic laser treatment, the ciliary processes are treated directly via the limbus or pars plana. The effect of cyclocryo therapy is variable and may cause pain, inflammation and swelling postoperatively, which is why the method is not common nowadays. The main complications of cyclo-destructive therapy are uveitis, anterior chamber haemorrhage, corneal decompensation, visual impairment, hypotension and phthisis.

6.5.4 | Antimetabolites

Wound healing at the surgical site affects the long-term pressure-lowering effect after glaucoma surgery, and the pressure-lowering effect of the procedure is improved

when antimetabolites are used. Chemotherapy drugs such as mitomycin-C (MMC) and 5-flurouracil (5FU) are routinely used in glaucoma surgery to reduce scarring of the conjunctiva. Risk factors for increased scarring are described in Table 5. As there are several complications of antimetabolites, an individual assessment should be made when considering the concentration and timing of any adjuvant antimetabolite therapy.

6.5.5 | Postoperative care

The care after glaucoma surgery requires frequent checkups and a high level of continuity to monitor the healing process. Topical steroid therapy is given at a high dose postoperatively to reduce inflammation at the surgical site and ensure effective filtration. The treatment is individual and guided by the appearance of the surgical site, anterior chamber depth, degree of inflammation, intraocular pressure and the presence of choroidal effusion. If high pressure occurs early postoperatively after trabeculectomy, the sutures of the scleral flap can be adjusted. If high pressure occurs after a deep sclerectomy, a puncture of the scleral window, known as goniopuncture, can provide adequate pressure reduction. High pressure associated with vigorous healing/scarring with the onset of a tenon cyst is treated with needling. The procedure is combined with injection of a steroid and/or an antimetabolite into the conjunctiva. If the pressure is low postoperatively, a cycloplegic can be added and sometimes it is necessary to reform the anterior chamber with injection of a viscoelastic intracamerally.

6.5.6 | Complications after filtering surgery

Development of cataract is common after trabeculectomy. Over-filtration with low intraocular pressure may occur early or late after glaucoma surgery. Treatment with antimetabolites may induce thin and leaky filtration blebs associated with blebitis and endophthalmitis. Patients undergoing filtering surgery should therefore be advised to consult an ophthalmologist if pain, redness, secretion or visual impairment occurs in the operated eye. Hypotension due to choroidal detachment or maculopathy can lead to visual impairment postoperatively.

6.5.7 | Cataract surgery and glaucoma surgery

When glaucoma surgery is indicated, and the patient also has a cataract affecting vision, pressure-lowering surgery and cataract surgery can either be combined or performed separately. Combined cataract surgery and filtering surgery have a better pressure-lowering effect than cataract surgery alone but have a lower success rate than filtering surgery alone.

Cataract surgery performed after trabeculectomy may affect pressure control postoperatively. Even uncomplicated cataract surgery causes inflammation and scarring and can negatively affect the function of a previous trabeculectomy. Cataract surgery can also affect the outcome of later glaucoma surgery if the glaucoma procedure is performed shortly after the cataract surgery due to the inflammation that occurs due to the cataract surgery. It is therefore beneficial to perform cataract surgery early in glaucoma patients when the pressure is well controlled.

> Consider early cataract surgery in patients with glaucoma.

Cataract surgery may have some pressure-lowering effect in open-angle glaucoma, but intraocular pressure may also rise postoperatively. Cataract surgery is therefore not recommended as the only pressure-lowering procedure. However, in the case of primary angle-closure (PAC), cataract surgery is a treatment option, both for primary angle-closure glaucoma (PACG) and high-pressure PAC.

7 | LEVEL OF CARE—WHO CAN/SHOULD DO WHAT?

7.1 | Opticians, optometrists and primary care

Many glaucomas are diagnosed in connection with an eye examination by an optometrist, or through contact with primary care providers. Thus, opticians, optometrists and general practitioners play an important role in identifying people with undetected glaucoma. It is becoming increasingly common for optical shops to be equipped with advanced diagnostic instruments such as a visual field device, a fundus camera and an OCT. Since the specificity of several devices is limited and the methods are subject to sources of error that may indicate false pathology, there is a risk of overdiagnosis of glaucoma with increasing access to and use of diagnostic equipment.

In order to avoid unnecessary referrals to health care with the associated resource consumption, it is therefore important that the indication for examination is well justified, and that examinations are repeated on a later occasion in case of uncertain results. Similarly, offices with diagnostic equipment as described above should ensure that sufficient medical competence is associated with the office so that a qualified assessment of the examination results is made.

7.1.1 | Who can be checked by an optometrist/optometrist outside of ophthalmic care?

A prerequisite for a long-term sustainable healthcare system is that priorities and resource considerations are made. Therefore, it cannot be considered justified that persons with a low risk of developing glaucomatous

disease are monitored in the context of ophthalmology clinics. Instead, these persons can be checked outside of ophthalmic health care system, for example by opticians and optometrists.

- Positive family history of glaucoma (siblings or parents). These individuals can be followed at an office with the ability to perform visual field examination (screening program) and/or optic disc assessment in addition to measuring intraocular pressure. See follow-up intervals on page 26.
- Borderline pressures of 22–24 mm Hg without damage or other risk factors in persons over 40 years of age can be checked every 1–2 years. If the pressure normalizes, checks can be terminated.

7.1.2 | At what pressure level is referral to ophthalmic care based solely on elevated intraocular pressure indicated?

- At pressure levels ≥25–30 mm Hg (measurement result verified on separate occasions).
- At pressure levels >30 mm Hg (one-time measurement sufficient).
- In case of verified pressure difference ≥5 mm Hg between the eyes regardless of pressure level. Observe! Keep in mind that a pressure difference can also be caused by previous cataract surgery.
- Observe! If exfoliation syndrome has been noted, referral may be sent at pressure levels >21 mm Hg.

> Adults under 40 years of age and persons with an intraocular pressure ≤ of 21 mm Hg should not be regularly monitored for glaucoma.

7.2 | Ophthalmic care

As resources are limited, it is important to prioritize how they are used. Ophthalmologists are responsible for glaucoma treatment. Assessment of examination results can, if appropriate, be delegated to a nurse/optometrist in ophthalmic care.

7.2.1 | Who is checked in ophthalmic care?

- Patients with manifest glaucoma.
- Persons with multiple risk factors.
- Persons with intraocular pressure ≥ 25 mm Hg.
- Persons with a pressure difference between the eyes ≥5 mm Hg.
- Persons with suspected glaucoma (abnormal optic disc and/or visual field appearance) where a diagnosis of glaucoma could not be made.

Evaluation and decision on continued controls and inspection intervals must be made continuously.

> It is likely that there is a significant overdiagnosis of glaucoma, which causes unnecessary check-ups, risk of overtreatment and patient anxiety. Therefore, for the correct use of resources and the benefit of patients, diagnosis, treatment needs, monitoring intervals and whether the patient needs to continue to be monitored should be regularly re-evaluated.

8 | FOLLOW-UP

The evidence for specific follow-up schedules is insufficient for patients with ocular hypertension as well as for suspected or manifest glaucoma. All glaucoma management and follow-up should be based on the individual's needs. In situations of limited resources, these should be focused according to the order of the Swedish medical priority list. As with all chronic medical conditions, a high level of continuity should be sought in the follow-up, for example via the patient-responsible doctor, nurse or equivalent (Figure 3).

The risk of developing glaucoma and glaucoma progression increases with the number of risk factors a patient exhibits. Patients with multiple risk factors should therefore be monitored at more frequent intervals. Patients with ocular hypertension or stable glaucoma are usually followed with less frequent follow-up visits compared to others. Although the definition of ocular hypertension is usually given as >21 mm Hg, the risk of developing glaucoma damage with slightly elevated pressure is small. Therefore, if other risk factors are missing, a higher pressure level is often chosen as the limit for follow-up in eye care.

8.1 | Follow-up intervals

At the initiation of treatment, a confirmatory eye pressure check should be performed within one month, but if the intraocular pressure is very high (≥35 mm Hg), this should be done within one week.

> Intraocular pressure should be checked on at least 2 separate occasions (preferably at different times during the day) before any pressure-lowering treatment is initiated and further follow-up is planned.

8.1.1 | Ocular hypertension

Patients with ocular hypertension should initially be followed up at intervals of 6–12 months, extended to check-ups every 1–2 years if the intraocular pressure remains stable and no glaucoma damage is detected. Follow-up visits are aimed at identifying possible glaucoma damage which is why the visual field and/or the optic disc

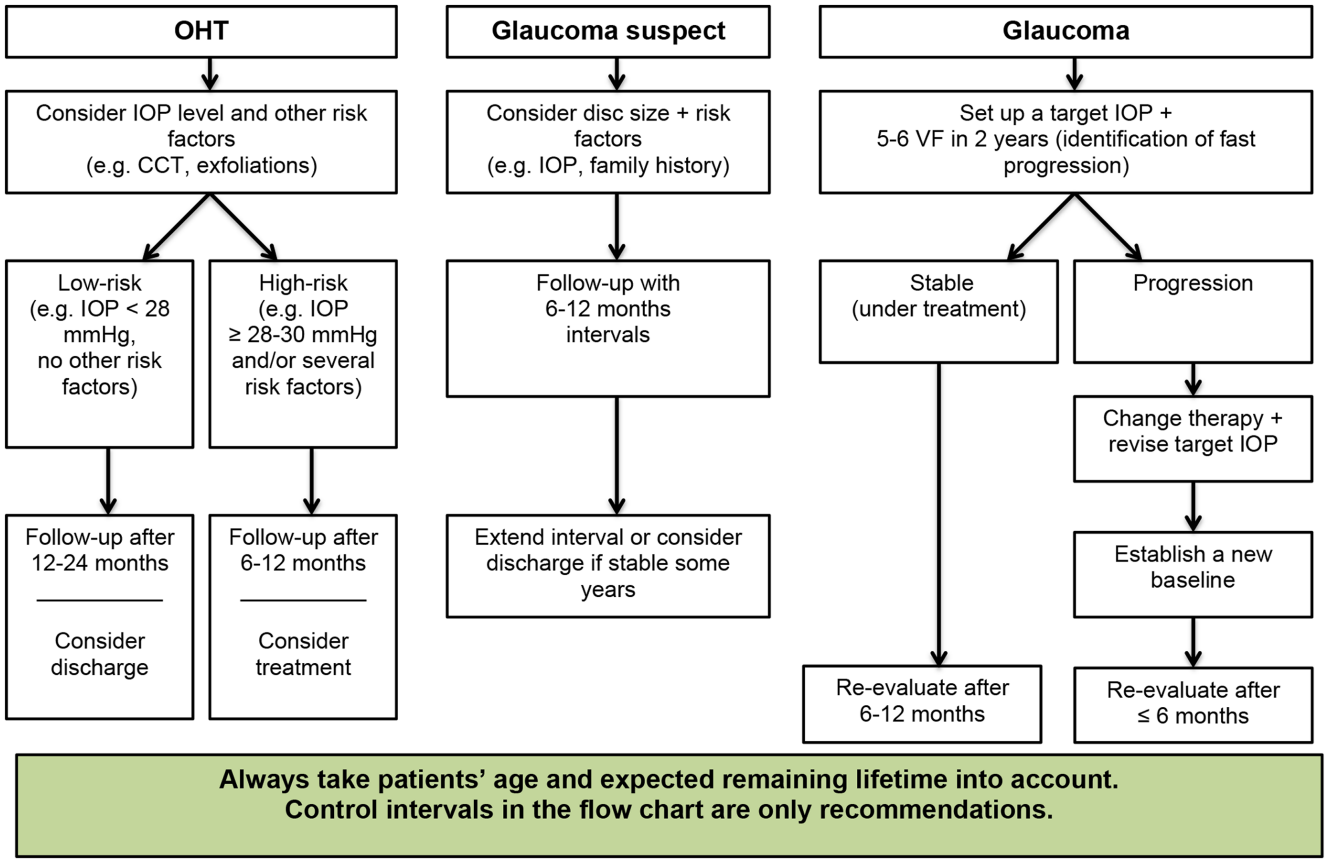

FIGURE 3 Flowchart inspired conceptually by a similar figure in the EGS Guidelines page 98, 2020, www.eugs.org (European Glaucoma Society, 2021).

must be examined. Consider terminating follow-up visits at repeated normal pressures.

- If IOP ≥25 mm Hg, check every 1–2 years.
- Consider treatment at repeated pressures of ≥28–30 mm Hg or 25 mm Hg if exfoliation syndrome is present.

Intraocular pressure-lowering therapy reduces the risk of developing glaucoma in patients with ocular hypertension.

8.1.2 | Suspected glaucoma

Patients with suspected glaucoma (suspected optic disc configuration and/or unclear visual field defects) are initially followed at intervals of 6–12 months. Extend the interval or consider terminating patient follow-up if pressure, optic nerve appearance and visual field remain unchanged after a few years of follow-up.

8.1.3 | Newly diagnosed glaucoma

Patients with newly diagnosed glaucoma should undergo visual field examination 5–6 times during the first two years in order to identify those with a rapid rate of deterioration as early as possible. In patients with a lower risk of developing visual field damage affecting quality of life, two visual fields per year for the first three years may be sufficient. Special consideration must also be given to the patient's expected remaining lifetime, ability to perform visual field examination, etc. With more visual field tests, progression can be detected more rapidly.

> 5–6 visual fields during the first 2 years do NOT apply to patients with newly discovered ocular hypertension!

8.1.4 | Glaucoma with known rate of progression

Once the rate of progression is determined after 5–6 visual field tests, further follow-up should be adjusted according to this and the patient's individual risk profile.

In case of exfoliation or pigmentary glaucoma, more frequent follow-up visits with pressure measurement may be indicated (compared to primary open-angle glaucoma or normal tension glaucoma).

> Re-assessment after the first two years: High rate of progression → more frequent checks. Low rate of progression → less frequent follow-up visits.

8.1.5 | After a change in target pressure

After identification of rapid progression when additional pressure-lowering treatment such as surgery has been necessary, a new evaluation of the rate of progression should be made (2–3 visual field tests per year) to ensure that the new lower target pressure has had a sufficient effect.

8.2 | Follow-up intervals for different risk factors

8.2.1 | Positive family history (glaucoma in siblings/parents)

- First check-up at the age of 50 at an office with the ability to perform visual field examinations (screening program) and/or optic disc assessment in addition to measuring intraocular pressure.
- Follow-up is then recommended every 5 years between 50 and 60 years and every 3 years between the ages of 60–75 years. After the age of 75, checks only linked to family history do not need to be performed.
- In the case of massively positive family history (≥2 first-degree relatives), the checks should be more frequent and start earlier if the relatives have been diagnosed at a younger age.
- Subjects with more distant relatives with glaucoma do not need to be checked because of a positive family history.

> In the case of a positive family history of glaucoma, checking intraocular pressure only is insufficient!

8.2.2 | Exfoliation syndrome

- Without elevated eye pressure, no follow-up.
- With elevated intraocular pressure—consider visual field examination every year, pressure measurements more often.

8.2.3 | Optic disc haemorrhages

- Infrequent examinations, (1-) 2 year intervals.

8.2.4 | Pigment dispersion syndrome

- Usually younger patients, more often men.
- Greater pressure variations.
- Higher risk of pressure peaks, for example in connection with physical activity.
- Without elevated intraocular pressure – infrequent check-ups (1-) 2 years intervals.
- With increased eye pressure > 21 mm Hg – more frequent checks.
- The majority of those who develop glaucoma do so within 10–15 years.

> The presence of several risk factors necessitates shorter intervals between check-up visits.

9 | COMPASSIONATE PATIENT MANAGEMENT

As a diagnosis of glaucoma is often associated with great anxiety and sometimes also entails significant limitations in the patient's life situation, accurate information about the nature, treatment and prognosis of the disease is of the utmost importance. At the time of diagnosis, sufficient time should be set aside for this. Particular attention should be paid to the importance of good adherence to treatment and regular check-ups, as well as information on patient associations and appropriate information material. A quick follow-up visit or telephone contact can be of value in answering further questions and ensuring that the patient understands the meaning of their illness. The establishment of glaucoma schools can promote the above through further information and by creating affinity and exchange of experience among newly diagnosed patients and individuals with practical experience of living with a diagnosis of glaucoma.

It is important that the patient is involved in decisions concerning their disease and that there is continuity in the follow-up of the disease. In the case of glaucoma-related visual impairment, rehabilitation can be crucial for a patient to maintain quality of life. It should be taken into account that loss of visual field can cause extensive functional impairment and referral to visual rehabilitation should be offered, even if central visual acuity is maintained.

Many factors affect a person's ability to access and cope with glaucoma treatment. An open approach in patient contact where healthcare professionals do not hesitate to raise difficult issues is very important.

10 | WISE ADVICE

Don't…

- Recalculate the intraocular pressure based on the CCT.
  - There is a lack of validated correction algorithms.
- Base glaucoma diagnosis and progression assessment solely on OCT measurements.
  - Abnormal OCT results are statistical deviations from a reference material and cannot be equated with clinical diagnosis.
- Use C/D ratio for glaucoma diagnosis and progression assessment.

- Normal cupping varies depending on optic disc size.
- Replace gonioscopy with different imaging methods.
  - Imaging methods are not accurate enough.
- Replace clinical assessments with AI (artificial intelligence).
  - Technology can support but not replace clinical assessment.
- Use stress tests to diagnose narrow chamber angles.
  - A negative test does not rule out the risk of acute narrow-angle attacks.
- Treat blind, symptom-free eyes with high intraocular pressure.
  - In the absence of visual function, treatment is only warranted in the case of pain.
- Use carbonic anhydrase inhibitors and hyperosmotic drugs in sickle cell disease.
  - These drugs can induce a hemolytic crisis in this disease.
- Use 21 mm Hg as target pressure in advanced glaucoma.
  - The intraocular pressure needs to be lowered significantly.

11 | QUALITY FOLLOW-UP

A reasonable quality follow-up of glaucoma care in Sweden requires a national quality register. These are available for several groups of eye diseases, including cataracts, macular degeneration and corneal diseases, but not yet for glaucoma. The surveys/cross-sectional studies on glaucoma that have been conducted so far in the country, have required large and irregular efforts and are not considered to lead to a sustainable flow of data. For this, a quality register is crucial.

A register can provide information on whether the proportion of severely visually impaired people varies across the country, but also on differences in incidence/prevalence, diagnostic criteria, treatment criteria and waiting times. Even if only a few parameters (e.g. only for a fraction of patients) such as social security number, diagnosis, visual acuity, intraocular pressure, VFI and any waiting time are registered annually, it would provide clear help to ensure patients' equal treatment and follow-up throughout the country.

ORCID

Gauti Jóhannesson https://orcid.org/0000-0003-0218-4563
Lada Kalaboukhova https://orcid.org/0000-0002-1125-0609
Dorothea Peters https://orcid.org/0000-0001-8062-2026
Christina Lindén https://orcid.org/0000-0002-3597-4740

REFERENCES

Heijl, A., Alm, A., Bengtsson, B., Bergström, A., Calissendorff, B., Lindblom, B. et al. (2012) The glaucoma guidelines of the Swedish ophthalmological society. *Acta Ophthalmologica. Supplement*, 2012(251), 1–40.

European Glaucoma Society Terminology and Guidelines for Glaucoma, *5th ed.* (2021). *The British Journal of Ophthalmology*, 105(Suppl 1), 1–169. PMID:34675001.

Tuulonen, A., Forsman, E., Hagman, J., Harju, M., Kari, O., Lumme, P. et al. (2014) How to interpret evidence in everyday practice: The Finnish Current Care Guidelines for open-angle glaucoma. Current Care Guidelines: Finnish Guidelines for Glaucoma Care. https://www.kaypahoito.fi/wp-content/uploads/sites/15/2019/01/Glaucoma-Current-Care-Guideline-2014-1.pdf

How to cite this article: Jóhannesson, G., Stille, U., Taube, A.B., Karlsson, M., Kalaboukhova, L., Bergström, A. et al. (2024) Guidelines for the management of open-angle glaucoma. *Acta Ophthalmologica*, 102, 135–150. Available from: https://doi.org/10.1111/aos.16599