The Glaucomas – a Review

DONALD MILLS, MD

SUMMARY The author reviews the group of disease entities called glaucoma. He delineates current thinking about the various forms of glaucoma in black-and-white. In actuality, some of the stated views still remain in the grey zone in ophthalmological circles. The author discusses the four major types of glaucoma, and what the family physician should know about them.

Dr. Mills is clinical assistant professor of ophthalmology, University of Western Ontario Medical School, and director of the Glaucoma Clinic at Victoria Hospital, London, Ont.

THE GLAUCOMAS are a complex of disease entities all characterized by an increased intraocular pressure which produces, in time, rather specific defects in the field of vision and excavation of the optic nerve head. The term glaucoma is no more precise than the designation 'high blood pressure' applied to another group of diseases. When one uses the unqualified term 'glaucoma' he usually is referring to the most common type – chronic open-angle glaucoma.

Incidence

Large surveys have shown that in the general population glaucoma affects about two percent of the people. If the subjects 40 years of age and over are studied separately, the incidence rises to about four percent. This means that glaucoma is as common as diabetes mellitus. Just as diabetes makes up a significant proportion of the family physician's practice, so glaucoma accounts for a large share of the ophthalmologist's practice.

Of the people registered with the Canadian National Institute for the Blind, about 12 percent have been found to be legally blind from glaucoma. Certain other causes of blindness (e.g. cataract) are fortunately remedial. However, visual loss in glaucoma is permanent.

Ocular Physiology

To understand the classification of the glaucomas, and the basis of therapy, it is essential to know a few basic points of ocular physiology.

The eyeball is well-named. It is essentially a ball with a constant internal pressure which runs within normal limits (11 to 22 mm. of mercury). Of course, the concept of a constant pressure is fallacious since the intraocular pressure varies with every beat of the heart and with every breath just as the blood pressure does. However, these refinements need not concern us here.

The structures of the eye (cornea, sclera, iris, choroid, retina, lens, vitreous humor, etc.) are fixed and unchanging, with one exception: the aqueous humor is constantly formed within the eye and constantly makes its egress from the eye. The aqueous therefore is the primary regulator of intraocular pressure.

Aqueous is actively and continuously secreted by the ciliary body into the posterior chamber of the eye (the region between the posterior surface of the iris and the anterior surface of the lens). From the posterior chamber the aqueous passes through the pupil into the anterior chamber and thence to the angle of the anterior chamber (the area where the iris root meets the cornea). In the angle of the anterior chamber the aqueous percolates through a special porous connective tissue called the trabecular meshwork and gains entrance to Schlemm's canal. From Schlemm's canal the aqueous is returned to the venous side of the circulation. Normally the amount of aqueous secreted is just balanced by the volume leaving the eye so that for all intents and purposes the intraocular pressure remains constant and within a normal range. The circulation of aqueous maintains the nutrition of the crystalline lens and keeps the eye in its nice spherical shape to function as an optical instrument.

Effects of Increased Intraocular Pressure

The structure in the eye which is most susceptible to a prolonged increase in the intraocular pressure is the optic nerve head. It was originally believed that the basic effect is a mechanical one – that the nerve fibers, as they leave the retina, are simply pinched against the edge of the optic nerve foramen in the sclera. In time, the nerve fibers become atrophic and there is loss of substance of the optic disc to give the characteristic cupping that one sees in advanced glaucoma.

Current studies, however, indicate that the primary effect of the increased pressure inside the eye is to occlude small capillaries which nourish the optic nerve head. Thus the damage to the nerve fibers is on an ischemic basis. This damage to the nerve fibers is not something that occurs quickly. It is probably a question of months (perhaps years) of raised intraocular pressure before permanent injury

occurs at the optic disc. The time required for production of atrophy of the nerve head is variable from one glaucoma patient to another. The inferior nerve fibers seem to be most susceptible to the raised pressure and the first visual field defect that one can plot on perimetry is usually an arc-shaped blind area near the upper pole of the physiologic blind-spot. In time, an inferior arc-shaped blind region develops; these arcuate scotomas break through to join, and in the end-stages the glaucoma patient may only have a small central island of vision.

If the intraocular pressure is elevated quickly and to very high levels (60-80 mm. of mercury) which can occur in acute angle-closure glaucoma, then the patient can rapidly go blind in the affected eye from occlusion of the central retinal vein, or, less likely, occlusion of the central retinal artery.

Measurement of Intraocular Pressure

The intraocular pressure can be assessed in the following ways:

1. Finger (tactile) tension – this method is listed simply to be condemned. Feeling the eyes to try to determine the intraocular pressure is akin to trying to guess the blood pressure by feeling the pulse.

2. Indentation tonometry – this is the time-honored technique of determining how far a weight of given magnitude sinks into the anesthetized cornea. The distance of indentation is converted to units by means of a long stylus and the units are converted to millimeters of mercury by reference to a table. The classical instrument for indentation tonometry is the Schiotz tonometer. In the average patient this form of tonometry is quite reliable and accurate. However, it can give falsely low readings in patients with low ocular rigidity, such as highly myopic individuals.

Most ophthalmologists are in favor of the family practitioner doing routine Schiotz tonometry as part of the complete physical examination, providing he is willing to take the time to learn to use the instrument reliably first. The Schiotz tonometer can be made to read anything the operator wishes. It requires several weeks of practice to obtain dependable readings. If the family physician is not willing to learn to use the instrument correctly, (glaucoma clinics are a good place to learn tonometry, just as diabetic clinics are a good place to learn to take blood), then he is better off without a tonometer. False readings lead to false referrals, or a false sense of security on the patient's part.

3. Applanation tonometry – this technique employs an instrument attached to the ophthalmologist's bio-microscope (or slit-lamp, as it is sometimes called). The force required to flatten a given area of the anesthetized cornea (and which is directly proportional to the intra-ocular pressure) is recorded and is read on the instrument in millimeters of mercury. This is the most accurate clinical assessment of the intraocular pressure and is independent of factors such as ocular rigidity which affect the Schiotz tonometer. Applanation tonometry requires equipment peculiar to the ophthalmologist and is not applicable to family practice use.

4. Cannulation of the anterior chamber – this direct method is undoubtedly the most accurate of all techniques. It is of value in the experimental animal but of course has no role in clinical practice.

Classification of the Glaucomas

In the 1950s, ophthalmologists interested in glaucoma agreed on a classification which is now universally accepted. This classification has been an important step forward because it has made it possible for all to know which type of disease other workers are discussing.

The basis of the classification is the angle of the anterior chamber of the eye (the iridocorneal angle).

The following is a simplified classification of the glaucomas, giving the approximate percentage of cases which fall into each category:

1. Chronic open-angle glaucoma (also called chronic simple glaucoma) – comprises about 85 percent of cases.

2. Acute angle-closure glaucoma – less than 10 percent of cases.

3. Glaucoma secondary to other eye diseases (iritis, dislocated lens, intraocular tumors, etc.) – less than five percent of cases.

4. Congenital glaucoma – present at birth – less than one percent of cases.

Chronic Open-Angle Glaucoma

1. *Mechanism.* This is by far the most common form of glaucoma and when one uses the word 'glaucoma' loosely, he means this type.

In chronic open-angle glaucoma, as the term indicates, the angle of the anterior chamber is wide open, and aqueous has no difficulty in reaching the trabecular meshwork. However, in this disease, the trabecular tissue is abnormal and a great deal of obstruction to outflow of aqueous is encountered in the pore tissue of the trabecular meshwork. It is easily understood that if the aqueous is produced at a constant rate and has difficulty making its normal exit through the trabecular channels, then the pressure inside the anterior chamber, and hence inside the globe, will rise. This, it is believed, is the primary mechanism obtaining in chronic open-angle glaucoma. Production of aqueous is normal but its egress from the eye through the trabecular meshwork is impaired so that there is a rise in base-pressure inside the eye. The base-pressure in chronic open-angle glaucoma is usually in the range of 30 to 40 millimeters of mercury.

2. *Signs and Symptoms.* It should be stressed that this, the common type of glaucoma, does not produce pain. The patient doesn't know he has chronic open-angle glaucoma, even when he has almost gone blind in one eye from it.

The family physician who sees a patient with intermittent vague pain around both eyes and headaches should dismiss from his mind the thought that this may represent glaucoma. It almost surely doesn't. In addition, the author speaks for many ophthalmologists when he says that headaches alone are very seldom found to be of ocular origin.

Early recognition of chronic open-angle glaucoma depends on routine careful measurement of the intraocular pressure at every ocular examination. Measurement of the intraocular pressure is a manoeuver that is never omitted by an ophthalmologist. It used to be common practice to check only patients over 40 years of age for glaucoma. We now do tonometry on everyone old enough to permit it. Adequate patient cooperation starts in the upper teens. Early recognition of glaucoma comes when the ophthalmologist examines each patient for it. There is little merit in making the diagnosis when the individual with a white cane is led into the office.

3. *Treatment.* Knowing the mechanism of chronic open-angle glaucoma, it is easy to understand the basis of therapy. This form of glaucoma is primarily treated medically. It is never cured but it can be controlled (again similar to diabetes mellitus). The medications available are preferably given topically in the form of drops. They either improve the outflow of aqueous by opening the pore tissue of the trabecular meshwork (e.g. pilocarpine or carbachol drops) or they decrease the production of aqueous by the ciliary body (epinephrine drops). If topical therapy is not adequate, one can treat the patient systemically with acetazolamide (or its derivatives). This compound inhibits carbonic anhydrase in the ciliary body and will decrease aqueous production by up to 60 percent. However, systemic side effects are common and one prefers topical therapy if it will achieve control.

Once the diagnosis of chronic open-angle glaucoma is established, it means the patient will be under treatment for the rest of his life. His status of control is judged by measuring the intraocular pressure, by determining the facility of aqueous outflow through a procedure called tonography, by plotting the visual fields and by assessing the appearance of the optic nerve heads. Generally the glaucoma patient is seen by his ophthalmologist serially at intervals of three to six months.

The family physician should be aware that there is little in the way of medications he may prescribe that will interfere with the patient's glaucoma control. The wide range of drugs which carry warnings that they should not be used in glaucoma have no effect on chronic open-angle glaucoma. These drugs usually have some atropine-like action and could conceivably induce an acute attack of angle-closure glaucoma in a susceptible subject by dilating the pupil. However, the person with acute angle-closure glaucoma doesn't know he has it until he gets it and so it seems asinine to put out a warning concerning the use of these medications in anyone with glaucoma. The family physician can safely disregard such contraindications.

If medical therapy does not control the glaucoma adequately and the patient continues to lose his field of vision in the face of full medical treatment, it is necessary to operate to lower the intraocular pressure.

The usual procedure is designed to create a permanent fistula between the anterior chamber and the subconjunctival space so that the globe is essentially no longer a closed system. This will effectively lower the pressure without ancillary medical therapy in about 80 percent of cases. Unfortunately, surgery often leads to undesirable sequelae such as cataracts of rather rapid onset. Thus, one may have the glaucoma well controlled in an eye that is reduced in visual efficiency because of a cataract.

Acute Angle-Closure Glaucoma

1. *Mechanism.* This is the second most common type of glaucoma but occurs much less frequently than chronic open-angle glaucoma. It is a different disease altogether. Acute angle-closure glaucoma occurs in certain predisposed individuals. It can occur only in a small, far-sighted eye in which the anterior chamber is quite shallow and there is little distance separating the peripheral iris and the back of the cornea. Should the iris be pushed forward to touch the posterior cornea, the approach to the trabecular meshwork (and Schlemm's canal) is completely occluded, so there is no chance of aqueous leaving the eye; the pressure rises precipitously. It seems that the initiating factor in acute angle-closure glaucoma is a block of movement of aqueous from the posterior chamber into the anterior chamber by the pupil. At a certain stage of pupillary dilatation (probably mid-dilatation) the peripheral iris is lax enough in predisposed eyes to move forward easily and the aqueous meets enough obstruction at the pupillary margin in its attempt to enter the anterior chamber that the root of the iris is pushed forward until the iris touches the posterior cornea and the angle of the anterior chamber is occluded. Under such circumstances the pressure in the globe rises quickly to extremely high levels. Intraocular pressures of 70 or 80 millimeters of mercury are not uncommon in acute angle-closure glaucoma.

2. *Signs and symptoms.* Acute angle-closure glaucoma is the disease about which every medical student learns. It is the type that can be induced by pupillary dilatation. The sudden rise in intraocular pressure to extremely high levels produces a very red, painful eye. Usually the pain is so severe that there are autonomic effects – nausea and vomiting – and the problem may superficially resemble an abdominal crisis.

The patient presents with a very painful eye with the vision usually reduced to the level of hand movements or light perception. The pupil is semi-dilated, ovoid and fixed, because of paralysis of the iris sphincter in the face of the high intraocular pressure. The eye is markedly injected in the perilimbal region. The cornea is quite steamy (edematous) because the sudden high pressure forces aqueous into the corneal substance and the cornea loses its state of dehydration which is largely responsible for its clarity.

The picture of acute angle-closure glaucoma is usually so characteristic that the diagnosis easily falls into the province of every family physician.

3. *Treatment.* Acute angle-closure glaucoma is an ocular emergency. If the intraocular pressure is not lowered promptly it is not difficult to understand that the central retinal vein or artery can be occluded and all useful vision lost.

The patient with acute angle-closure glaucoma is admitted to hospital at once and medical measures are employed to lower the intraocular pressure in the affected eye. Pilocarpine drops used intensively may be partially successful in improving the outflow of aqueous but they are not uniformly effective because the angle of the anterior chamber remains occluded. Systemic medications which suppress aqueous production (acetazolamide used intramuscularly or intravenously initially) are usually effective in aborting the acute attack. If these measures fail, one can almost invariably break the acute attack by means of hyperosmotic agents – glycerol by mouth or urea, or mannitol by vein. Osmotherapy quite effectively dehydrates the globe and brings the acute attack to an end.

Once the pressure in acute angle-closure glaucoma is lowered to the normal range, the final management is surgical. One creates a permanent opening between posterior and anterior chambers so that pupillary block of aqueous can never again initiate an acute attack of angle-closure glaucoma. In this procedure (called peripheral iridectomy) the ophthalmologist makes a surgical coloboma in the iris at its root.

It should be pointed out that the eyes are symmetrical structures as a rule, and thus the patient who has had an acute attack of angle-closure glaucoma in one eye has a great

risk of having an attack in the other eye. One large study has shown that acute attacks of glaucoma of the angle-closure type occurred in the fellow eyes in at least 50 percent of cases within five years following the attack in the first eye. Accordingly, it is sound ophthalmological practice to carry out a peripheral iridectomy prophylactically in the unaffected eye prior to the patient's discharge from hospital.

Although acute angle-closure glaucoma comes on quickly with severe pain and is a frightening experience for the patient, if it is recognized early and treated promptly, it is a type of glaucoma which can be cured surgically.

Secondary Glaucoma

1. *Mechanism.* There are many mechanisms leading to secondary glaucoma. However, all secondary glaucomas have in common the fact that some initial ocular disease antedates the onset of the glaucoma. The glaucoma, when it occurs, can be of the open-angle or angle-closure type. Ocular diseases which can produce secondary glaucoma range all the way from entities like iritis, hypermature cataract and dislocated lens, to life-threatening problems such as intraocular malignant tumors. Most of the primary eye diseases leading to secondary glaucoma lie beyond the diagnostic range of the family physician.

There is, however, one type of secondary glaucoma which the family practitioner may actually produce and about which he should be informed. It is known now that one-third or more of normal individuals when put on potent corticosteroid-containing eye drops will develop, in the space of four to six weeks, a condition completely indistinguishable from chronic open-angle glaucoma. This is a steroid-induced problem which can produce the sequelae of chronic open-angle glaucoma. Fortunately, the glaucomatous state is totally reversible once the corticosteroids are withdrawn.

The family physician who prescribes corticosteroid-containing eye drops (or ointment) for his patient for some trivial external eye problem, and permits the prescription to be repeatedly renewed, may unknowingly allow the patient to develop a secondary glaucoma from the medication. In actual fact, ocular conditions requiring corticosteroid therapy usually lie outside the field of competence of the family doctor. There is virtually no reason why he should prescribe a steroid ophthalmic preparation. It is a wise physician who takes the time to see what is in the ophthalmic drops and ointments he prescribes. The number containing potent steroids is legion. If the family doctor deems it imperative to prescribe a steroid ocular preparation, he should see to it that the prescription is non-repeatable. It would be better still for him to learn the names of several antibacterial eye drops or ointments (the most frequently prescribed agents by family doctors) which don't contain corticosteroids – and to stick to these. There is virtually no place for the antibacterial-steroid combination ophthalmic product in family medicine.

2. *Treatment.* The treatment of secondary glaucoma, of course, is directed towards the initiating cause. Treatment may be sublimely simple, as when one has the patient stop using the corticosteroid eye drops he has been using for some time. On the other hand, treatment may require such a formidable procedure as enucleation of the eye for an intraocular malignant melanoma.

Congenital Glaucoma

1. *Mechanism.* The mechanism operating in this fortunately rare type of glaucoma is not completely elucidated. However, it seems certain that there is a failure of normal embryological development of the angle of the anterior chamber. The angle is wide open but the trabecular meshwork is imperfectly developed or is covered by an impermeable membrane. Schlemm's canal appears to be normally present in most of these children.

The response of the adult eye to an elevated intraocular pressure is excavation of the nerve head. In the infant (up to the age of two years or so) the response of the young, elastic eye to the elevated intraocular pressure is to enlarge to a considerable size. Hence congenital glaucoma is also called buphthalmos ('ox-eye') because of the enlarged globe. Excavation of the optic nerve head does occur in these children as well, however.

2. *Signs and symptoms.* The earlier congenital glaucoma is diagnosed, the better the prognosis.

Early findings may include an irritable infant with tearing, photophobia and blepharospasm.

The occurrence of corneal enlargement and edema produces a big, steamy cornea which is strongly suggestive of congenital glaucoma.

Congenital defects in one system are frequently accompanied by congenital defects in other systems. Thus patients with infantile glaucoma may also have congenital pyloric stenosis, deafness, cardiac problems, mental retardation, etc.

Although the condition is rare (the average ophthalmologist sees one new case of congenital glaucoma every five years) it is not unusual for the family physician to be the first to suspect the nature of the ocular disease.

3. *Treatment.* Congenital glaucoma is a surgical problem. One attempts to open a route for aqueous flow into Schlemm's canal by means of an incision into the imperfectly formed trabecular meshwork. The procedure is called a goniotomy.

If the pressure is elevated and the cornea is hazy at birth, there is much less chance of effecting a cure than when the signs and symptoms appear after the second month. In those infants with signs present at birth the salvage rate in terms of visual function is about 50 percent. Over 80 percent can be arrested when the disease becomes obvious after the second month.

All in all, congenital glaucoma still offers a rather discouraging prognosis. ◀

CORRECTION

Dr. John Z. Garson, co-author of an article on electrocardiograms which appeared in our October issue, was inadvertently described as 'formerly of Saskatoon Community Clinic'. Dr. Garson is still very much a member of the Saskatoon Community Clinic to which he will be returning after he completes his present one year leave of absence, doing a course in epidemiology at the University of Toronto. We regret any embarrassment our statement may have caused Dr. Garson or his colleagues in Saskatoon.