

THE CAMBRIDGE GLAUCOMA LETTER

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UNANSWERED QUESTIONS

Glaucoma is a disease, or rather a group of diseases, of such interest and challenge that no apology is required for a newsletter devoted exclusively to questions about glaucoma diagnosis and treatment.

The simple fact of increased intraocular pressure is a common denominator for all the glaucomas. Comprehensible to the least sophisticated of patients and a puzzle to the most astute investigators, this unifying concept persuasively illustrates both the force and futility of logical reduction. The pathophysiology of glaucoma has been more succinctly identified than that of many other diseases, but precisely at the point where we presume to grasp the cause, the cause eludes us, and what we know begins to appear insignificant in comparison with what we have yet to learn. The cause that we pursue proves to be the shadow of our reason, which reason cannot catch.

By definition, the study of glaucoma centers around the phenomenon of destructive intraocular pressure. This pressure, in turn, is understood as a function of the flow of the aqueous humor, secreted or filtered from the capillary circulation of the ciliary body and perhaps the retina. It passes then around or through the vitreous into the posterior chamber, around the equator of the lens, between lens and iris, through the pupil, into the anterior chamber, to its angle and then through the trabecular meshwork into the canal of Schlemm, through the aqueous veins, to the episcleral venous circulation and thence again into the mainstream of the blood. It is a schema familiar to every clinician, and if the catechetical recitation of its course could give control over the flow of aqueous, the glaucoma problem would already be solved. But this is not the case. The description that we give serves partly to conceal all that we do not understand.

Each discovery concerning the mechanism of glaucoma becomes then the source of new questions and problems. Both the se-

cretion of the aqueous from the capillaries and its resorption into the episcleral veins raise unanswered questions as does virtually every intermediate stage. The entrance of aqueous into and its passage through the vitreous body determine in part the depth of the anterior chamber, but the mechanism by which this occurs is not yet understood. For the present, the etiology of malignant glaucoma eludes us. The circulation of the aqueous through the pupil and the attendant differential pressure enveloping the iris account for at least some of the anterior displacement of the iris that occurs in angle closure. The other forces that control the position of the lens and of the iris root are, however, undefined. The anatomy of the trabecular meshwork has been the subject of much study, but we have only hypotheses to suggest the way in which the size of its pores is altered in glaucoma, and we can only guess at the mechanism by which its outflow resistance is altered when the ciliary body is subjected to the various anti-glaucoma medications. It remains a mystery by what forces the episcleral venous pressure is maintained and how it is possible for the intraocular pressure at times to be less than the pressure of the venous circulation into which the aqueous drains. And finally, the control system responsible for maintaining intraocular pressure relatively constant, at least in the healthy eye, remains totally undefined.

This catalogue of uncertainties is far from complete but it suffices to illustrate the fact that the reduction of a disease process to mechanical relationships does not, at least in the case of glaucoma, give anything but tentative answers. It suggests also that the function of scientific theory is perhaps not to provide conclusions but to generate questions. These questions then serve as a framework for observation and reflection. Questions are valid not to the degree that they admit of answers but to the extent that they constitute an accurate conceptual reflection of experience. Advances in our understanding of glaucoma are likely to come as by-products, inadvertently, sometimes unexpectedly in the

pursuit of questions that remain unanswered and that in the final perspective may be unanswerable. Discovery cannot be forced. When the climate is favorable, it arises spontaneously.

It is characteristic of the scientific literature that authors are expected to provide answers, to contribute some item of novelty to the ever-swelling sum of human knowledge. What distinguishes this publication is its concern with questions, even if these questions cannot be answered, or perhaps for that very reason.

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GLAUCOMA SCREENING

Glaucoma is remarkable among the common causes of loss of vision in that its deleterious consequences are, by and large, preventable. It is fashionable nowadays to insist that, if at all possible, medicine should be preventive rather than curative. The prophylaxis of glaucomatous blindness therefore appears both to the public and to the professions as a major challenge and obligation. In the eagerness to "prevent" glaucoma, the various inherent difficulties of this challenge are sometimes overlooked.

The term "glaucoma screening" is customarily applied to abbreviated examinations, usually of an entire group of individuals, for the express purpose of detecting glaucoma. Such projects suggest that glaucoma may be readily diagnosed with a minimum of instrumentation and time. This assumption is doubtlessly correct for patients with advanced glaucoma. In the earliest stages of the disease, however, a short examination may be inadequate to establish - or to exclude - the diagnosis of glaucoma with reasonable certainty.

In order to understand the problems revealed and created by group screening projects for glaucoma, it is helpful to consider such efforts as bringing to a focus the obligation of all ophthalmologists and optometrists to utilize every encounter with their patients to try to exclude the presence of incipient glaucoma. This perspective is particularly useful, because it provides a yardstick by which the glaucoma screening clinic may be evaluated.

The group screening procedures are designed to circumvent the economic barriers to frequent examination for glaucoma

for large segments of the population. By making the screening procedure a well-advertised public event, they tend also to dispel the fears and inhibitions that might keep unknowing victims of the disease from seeking diagnostic help. The physical examination of the patient is removed to a public place such as a high school auditorium or the lobby of a hospital. One of the screening clinics in which I participated was conducted in a large house trailer which unfortunately was parked on a grade, so that I had the unbalancing experience of examining patients on an inclined plane. Nurses, optometrists, ophthalmologists and secretaries are usually asked to donate their services. Their status as volunteers tends to make them independent of administrative control. As a result the procedures of the screening clinic are often improvised, and any deficiencies of the project are compensated by the public-spirited dedication of the participants.

A number of important facts are commonly overlooked by those who organize glaucoma screening clinics and by those who attend them. The term glaucoma is applied to a wide spectrum of eye pathology. It is a diagnosis which has different implications for different patients. The inference unavoidably arises that glaucoma is a single disease entity, where in fact the word is a name for a group of diseases that share a common pathophysiologic mechanism but have different etiologies. The prognosis of glaucoma varies with the particular circumstances. Furthermore, even within a well-defined category, the disease may behave differently. For example some patients with chronic angle closure have a remarkably benign course, while in others the same condition leads to catastrophic attacks with very high pressure, synechia formation, and early blindness. Finally, and not least in importance, all pathophysiologic considerations aside, the disease affects each person in a manner peculiar to his situation in life. Any given disease will have different significance for different individuals. It is, after all, only a heuristic abstraction to consider a disease as distinct from the human being who is affected by it. Many a well-meaning examiner who volunteered to help with a glaucoma screening clinic has been troubled by patients' misapprehensions to which he inadvertently contributed.

Often the screening examination will not suffice to diagnose glaucoma. At best, it can serve only to select those patients who are most likely to benefit from more extensive examination. Even when no disease is found, the examination should be repeated at intervals of one to

two years. Patients should be informed accordingly. Still, no matter how explicit such cautions are made, patients invariably demand more information than the screening examination is designed to furnish, and the examiner regularly finds himself in the position where he must plead ignorance for lack of opportunity to perform a more thorough evaluation.

Patients who are "screened" for possible glaucoma fall naturally into three groups. There are those, in the first place, who have no suggestive signs of glaucoma whatsoever. Then there are others, whose intraocular pressure is perhaps 30 mm Hg or higher where the presence of glaucoma is a foregone conclusion. Finally there are those who have various signs suggestive of glaucoma but none so conclusive as to warrant an affirmative diagnosis.

The only truly satisfactory outcome for the examiner is to be able to announce to the patient that there are no signs of glaucoma and that nothing more than another routine screening examination after an appropriate interval seems indicated. If, on the other hand, the findings of the examination are unequivocally positive, i.e. if unmistakable signs of glaucoma are found, if the screening examination has truly fulfilled its purpose, the examiner will be frustrated in his inclination to follow a potentially interesting and important problem, and this all the more so, if the brief examination has sufficed to convey to the patient the doctor's concern and the patient responds with a plea for help.

Patients in the second group, those who have obvious glaucoma, present the lesser problem to the examiner. According to the rules, they are to be referred to an ophthalmologist of their choice for further treatment. It is not at all certain that they will obtain the treatment that they require. This may be of particular concern if the glaucoma is very acute, when the pressure is very high or the disc is badly cupped, and procrastination may cause irreparable harm. The screening clinic will of necessity be so organized as to preclude any lengthy or detailed explanation to the patient or any effective support for the anxiety that has been evoked by the unfavorable report.

Most perplexing are those situations where one or more signs suggest the possibility of glaucoma but none are sufficiently conclusive to warrant an affirmative diagnosis. The examiner is then placed in the position of having to declare his uncertainty, of admitting that he does not know, while at the same time

assuring the patient that another examiner in a more favorable situation would probably be able to give him the definitive answer which he desires. No wonder that it is not always easy to persuade examiners to volunteer for glaucoma screening clinics.

The most common cause of uncertainty is probably the borderline tension. When the Schiøtz tonometer is used, a value of 4 scale readings with the 5.5 Gm. weight, with the more accurate Goldmann or Perkins applanation tonometers, values above 21 mm. Hg. would be considered abnormal. The probability that a given patient has glaucoma is, mathematically speaking, a function of the value of the isolated tonometric reading. I know of no empirical studies to this effect, but clinical experience suggests that the probability that any given patient has glaucoma approximates an exponential function of the value of an isolated tonometric measurement. The uncertainty that arises from borderline values of the pressure is usually not too difficult for patients to accept. Although most patients have no appreciation of measurement errors, they understand that a given range of values might have indeterminate diagnostic significance. Actually the accuracy of tonometry in the setting of the screening clinic is probably overrated, since the novelty of the experience is likely to make patients - and their extraocular muscles - transiently tense, and while such tension almost certainly does not explain permanently elevated pressures or cause glaucoma, it does account for what in retrospect may appear as an inexplicably elevated tonometry reading.

The examination and evaluation of the disc present some of the most difficult problems of glaucoma screening. Assuming, as is not always the case, that an accurately aligned, transformer-powered ophthalmoscope is available, and that the examination is performed in a place where illumination can be controlled, the combination of hazy lenses, small pupils, and roving eyes can still prove to be a trial for even the keenest and most patient of observers. Screening clinics are designed like assembly lines. The examiner is under pressure to commit himself to decisions which can sometimes be very difficult to make. Indecision is likely to be misconstrued as incompetence rather than as conscientiousness. The announced purpose of the screening clinic will belie all explanations about the uncertainties inherent in glaucoma diagnoses. To be sure, many a flat, pink disc can be dismissed off-hand as being non-glaucomatous, and the pale, atrophic disc with the deep, broad cup that reaches

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everywhere to the disc margin must be assumed to have a glaucomatous cause. But between these extremes there is that large area of diagnostic uncertainty which we refer to as physiologic cupping or saucerization, about which no expert can be certain whether or not it is evidence of glaucoma. Nor do we know whether such excavation makes the disc more sensitive to glaucomatous damage if and when elevation of pressure supervenes. One evaluates such discs in conjunction with the tonometry value and perhaps, less satisfactorily, with the anterior chamber depth. In one's own mind one then assigns to them some position on a scale of relative risk and advises the patient accordingly.

What proportion of patients with minimal signs of glaucoma one refers for further examination must reflect the expectations of the community. If none of

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them are referred, one will almost surely miss some cases of early glaucoma. If one refers them all, there will be many patients put to expense and anxiety which in retrospect might have been avoided. What is usually forgotten in these circumstances is that the examiner has been asked to work without the aid of some of his most valuable tools. He has no descriptions of the discs or comparison photographs from prior examinations, and most important, he lacks the opportunity for repeated examination over a period of months or years, which may be the only reliable and conclusive way to establish the diagnosis. Finally, those of us accustomed to gonioscope all patients who do not have obviously deep chambers are embarrassed not to have a chance to perform this sometimes most revealing of examinations, and all the more so, since angle closure glaucoma is the only form of glaucoma which can truly be cured. Guessing at the width or narrowness of the angle from the depth of the anterior chamber and from the convexity of the iris plane is obviously better than ignoring the hazards of angle closure altogether, but it seems a shame not to take a look at the angle through the gonioscopy lens where such valuable information is so close at hand.

The screening procedure presupposes an accepted standard of caution upon which the examiner should base his advice concerning the need for follow-up examination. But custom and personal preference are variable and the hypothetical "standard of care" is a figment of the legal imagination. What seems sensible to one examiner may seem either unduly risky or extravagantly cautious to another. And who is to decide?

Notwithstanding its imperfections, glaucoma screening remains an important tool in the early diagnosis of the disease. It will be an even more effective one, if those involved in glaucoma screening understand its limitations.