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Angle Closure Glaucoma - 3

The discovery that angle closure glaucoma is usually precipitated by pupillary block makes this disease uniquely amenable to treatment and is responsible for the circumstance that when diagnosis is timely, angle closure, alone among all the glaucomas, is susceptible to definitive cure. It is the clarity with which pupillary block is defined and the conclusiveness with which it can be treated, which lends to angle closure glaucoma that aura of nosologic perfection to which we have alluded in previous issues of the Glaucoma Letter. In contrast, however, to the elegance of pathophysiological explanations, the practical implementation of our theoretical knowledge is beset with difficulties, incongruities which have no place in the textbook's catalogue of factual knowledge, and which the lecturer neglects to mention lest his inability to resolve them reflect unfavorably on his expertise. The difficulties spurned by academic theory do not go away. Day after day they confront the ophthalmologist in practice, who must improvise solutions as intuition suggests and custom seems to demand, compromises acceptable to the patient and consistent with the physician's view of his task. Until problems have been conceptually articulated, they cannot be scientifically resolved, and techniques for dealing with them will be drawn from the art rather than the science of medicine. Nor is subspecialization always the answer. The subspecialist also, in many an instance, will be unable to solve the problem, and his task then becomes one of making the limitations of medicine psychologically and socially acceptable, more plainly, of muddling through in style.

As is the case with the other glaucomas, the damage caused by angle closure glaucoma, if sufficiently protracted and severe, cannot be reversed, and the therapeutic aim, therefore is to prevent such damage from occurring. As with all glaucomas, one wishes to protect the optic disc from excavation and the optic nerve fibers the atrophy that is caused by prolonged tension elevation. In addition there are two other sorts of preventable injury which do not occur in open angle glaucoma. In the first place, the very high intraocular pressure generated by acute angle closure may, in a matter of hours, precipitate blindness from atrophy of the optic nerve head. Secondly, the apposition of the iris to the trabecular meshwork may lead to the formation of permanent peripheral anterior synechia, which once they are established, are often responsible for subsequent intractable elevation of the intraocular pressure. In the presence of inflammation and elevated pressure, synechia may form in a matter of days, if not indeed of hours, while if the pressure is low and the eye is uninflamed, iris apposition to the meshwork may be tolerated for months without the formation of synechia.

Since timely iridectomy will, in most instances, prevent both the acute attack and the gradual development of permanent synechial closure, thus eliminating the tension elevation that leads to blindness, the prerequisite to effective therapy is accurate prediction: to identify those eyes that will, in the proximate future, sustain irreversible damage from angle closure. One seeks to avert three distinct pathological processes: 1) the catastrophe of acute angle closure, 2) the development of synechia in a chronically closed angle, and 3) as in open angle glaucoma, the progression of nerve damage in consequence of chronically elevated intraocular pressure. Unless we are able to predict the onset of the disease process accurately, we will either treat our patient prematurely, or we will treat him too late. However, to predict the onset of angle closure glaucoma is to foretell the future. That is the prerogative of the prophet, and turns out to be far more difficult than the textbooks lead one to expect.

In assessing the risk of potential angle closure glaucoma, the classical description of the acute attack is beside the point. When a patient comes to us complaining of sudden onset in a previously healthy eye of pain, blurring, haloes, and when tonometry then demonstrates a very high intraocular pressure, and gonioscopy shows the angle to be closed, then diagnosis poses no problem and there is no doubt about the need for emergency treatment, but no doubt also that such treatment is in many instances too late. The prevention of angle closure requires

attention to far more subtle signs. The factors that signal an increased risk of angle closure glaucoma include a history of a sibling or a parent who has angle closure, as distinct from other glaucoma, a history of recurrent ocular pain or discomfort with or without diffraction haloes in the field of vision, an apparently shallow anterior chamber on preliminary examination with flashlight or slit lamp, tension elevation in the absence of a manifestly deep anterior chamber. When one or more of these risk factors is recognized, the ophthalmologist must make additional effort to assess the possible development of angle closure.

The threshold issue in the diagnosis of impending angle closure glaucoma is, "When and whom do we gonioscope?" In recent years, a good deal of effort and ink have been expended to promote the thesis that angle closure can be predicted without gonioscopy, by means of flashlight examination of the iris convexity, for example, or by calibration of the depth of the peripheral anterior chamber at the slit lamp. Such efforts would make sense if gonioscopy were indeed a difficult or dangerous or expensive examination. But given that gonioscopy is so quick and simple and safe, and that the instrumentation required is relatively inexpensive, it seems inappropriate to rely on these cruder and less accurate techniques for anything more than the selection of patients for whom gonioscopy is worthwhile. It is surely better to err on the side of performing gonioscopy too often than not often enough. The rule is that when in doubt, one should gonioscope. If one follows this precept, one will not only overlook few if any eyes with potential closure, but one will be surprised how variable the relationship between peripheral chamber depth and angle width in fact turns out to be. Another benefit of not making of gonioscopy a rare procedure is that frequent gonioscopy increases ones understanding of the wide spectrum of angle anatomy that is compatible with a normal intraocular pressure, and reminds one how many extremely narrow angles there are which do not close.

Although we dismiss the effort to diagnose angle closure without gonioscopy, it is worth pausing a moment to identify it as an example of a recurring tendency to substitute intuition and guesswork for diagnostic precision. Ultimately all selection of diagnostic procedures is subject to criteria of cost-benefit evaluation similar to those used to assess the value of therapeutic efforts. When we consider on the one hand the substantial value of gonioscopy, enabling us as it does, to look at the anterior chamber angle, and on the other hand, the minimal effort and risk it entails, particularly in the cooperative patient, our analysis most likely will lead us to perform the procedure in every eye whose iris is convex and whose anterior chamber is not patently deep.

Undeniably, in contrast to the ease of gonioscopy, there is a sportsmanlike challenge to the precarious enterprise of attempting to predict the width of the angle with flashlight or slit lamp, much like the challenge of estimating the intraocular pressure by palpation rather than by tonometry. At the time of scleral buckling surgery, one sometimes presumes to assess the height of the intraocular pressure by peering at the disc through the hazy cornea with the indirect ophthalmoscope instead of picking up the Schiotz tonometer from the instrument table to ascertain what is really going on. In this same vein, I recently heard a lecturer advise his audience to dispense with fluorescein angiography in making diagnoses of retinal vacular disease, but to rely on stereo fundus photos alone. When we were children, we would coast down the street on our bicycles in search of admirers shouting to anyone in sight, "Look, look, no hands!" But when it really matters, when the patient's eyesight is at stake, it seems better to forego the acrobatics and the display of virtuosity, and to grip the handlebars tightly and take a look through the gonioscope to make sure the angle is safely open.

As diagnostic examinations go, gonioscopy has a high benefit ratio. It costs little and tells much. A quick look through the Koeppe lens immediately eliminates a large proportion of suspects whose angles are so wide that they could not possibly have closed in the recent past, and will surely not do so in the forseeable future. But while it is easy to identify many patients as being at no risk whatever for angle closure, the converse is not true. With the gonioscope alone the identification of individuals who will develop acute angle closure is subject to large statistical uncertainties. Many patients have angles that are slit-like in some meridians, and optically closed in others. If such patients have normal intraocular pressures, we infer that they do not yet have, but are at risk to develop angle closure. If their intra-ocular pressure is elevated, and if less than one third of the angle circumference is closed, we make a diagnosis of open angle glaucoma with narrow angles, while if more than one third of the angle circumference is closed, we make a diagnosis of chronic angle closure glaucoma. In this last instance, we cannot be certain of our diagnosis until after iridectomy. Then, if when pupillary block is relieved, the angle opens and proves to be free of synechia, but the pressure remains elevated nonetheless, we infer that we were dealing with open angle glaucoma all along and that the angle closure which we observed pre-operatively was optical rather than hydrodynamic. Yet even so, we do not regret the iridectomy. For one thing,

it permits the use of both epinephrine and miotics without fear of thereby precipitating angle closure. It also makes the angle accessible to laser trabeculoplasty, should that treatment be required.

Since pharmacological mydriasis induces angle closure glaucoma which is clinically indistinguishable from the naturally occurring disease, the theory is sometimes entertained that mydriatic and spontaneous angle closure are expressions of one and the same disease, and that therefore mydriasis is a valid diagnostic maneuvre in patients known to have narrow angles. Moreover, it has been argued that the patient whose angle is inadvertently closed by mydriasis has in fact received the benefit of early diagnosis of a potentially blinding disease. However, more critically examined, there appear to be many patients for whom there is an unacceptable risk of mydriatic induced angle closure whose angles are yet unlikely to close spontaneously. If one is careful one can practice ophthalmology for decades without ever precitipitating pharmacologic angle closure, and that, indeed, should be our aim. Gonioscopy enables us to distinguish degrees of risk of mydriatic closure. In determining whether one should dilate the pupil get a better look at the fundus, one makes a clinical judgment which must reflect the particularities of the situation, and the patient fares much better if his physician does not follow a routine. At least implicitly, one must, in each instance, answer the questions: What is dilation of the pupil to accomplish? For what disease is one looking, and what would one do about it if one found it? The routine dilation of patients with narrow angles as a ritual of thoroughness is difficult to justify. In the absence of systemic disease and the absence of visual symptoms and in the presence of normal visual fields, the likelihood of discovering disease amenable to treatment is very low and probably does not warrant the risk of even an occasional case of angle closure. On the other hand, where there is a clear suggestion of treatable posterior pole pathology, and the fundus is not sufficiently visible through the small pupil, one has little choice but to recommend to the patient that he accept the risks of possible mydriatic closure for the putative benefits of the additional diagnostic information to be obtained. This in particular is an area, where one should obtain the patient's informed consent before proceeding on a course that may lead to the most serious of ocular complications.

The diagnosis of potential spontaneous angle closure is a very different matter. The ophthalmologist who undertakes the preventive treatment of angle closure glaucoma is confronted with a large group of patients who have very narrow angles and normal or mildly elevated tensions. What explanations do we give, and what treatment, or what further studies do we recommend? The response of an eye to potent doses of mydriatic is, as we have pointed out, so different from its normal behavior, that it is probably meaningless to instill mydriatic drops for the purpose of ascertaining the risk of spontaneous angle closure, for there is many an angle which will close with mydriatics which in the absence of such provocation would never have given the patient any problems. The rational provocative test for angle closure is exposure to darkness. Darkness dilates the pupil and in susceptible eyes, it causes the angle to close and the pressure to rise. The darkroom test simulates a situation in which the patient is likely to find himself virtually every day, and it is not difficult to persuade him that if merely spending an hour in the dark should be sufficient to provoke an attack of glaucoma, one ought seriously consider prophylactic surgery.

The darkroom test is simple in theory and simple in practice. One measures the applanation tension, asks the patient to stay in the dark for one hour, and one then repeats the measurement. Sometimes it is awkward to make the room absolutely dark. Therefore a mask is applied to exclude the remaining light, carefully so as not to abrade the eyes still numb from topical anesthesia. A few precautions are appropriate. The topical anesthetic required for tonometry sometimes inhibits the blinking reflex, with subsequent drying out of the corneal epithelium and epithelial breakdown. Therefore the patient must be instructed to blink frequently. Another possible source of difficulty is movement of of the mask during the test. If it rides up, its edges may abrade the anesthetized cornea. For this reason also no cotton or other padding should be placed under the mask. Since patients who require darkroom testing are often of advanced age, there is some risk of their becoming disoriented while confined for an hour in the dark. They may fall asleep and forget where they are; they may unwisely try to get up and walk blindfolded. We deal with this problem by asking a member of the patient's family to keep him company during the test. We also give the patient a small bell to hold, so that if he becomes uncomfortable in the course of the test, he can readily summon help. Finally it is thoughtful if an assistant looks in on the patient every few minutes to make certain that all is well.

It is not so much the absolute value of the intraocular pressure at the end of the test, but any rise that occurred in its course which is of diagnostic significance. In general, if the pressure rises by 8 mm. or more in one or both eyes, the test is considered positive, and the patient is assumed to have angle closure glaucoma. If the pressure rise is smaller, the test is considered negative, and angle closure in the immediate future is

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relatively unlikely. A pressure rise greater than 8 mm. should not be taken uncritically to infer the presence of the disease. One always performs gonioscopy at this juncture to confirm that closure of the angle does indeed accompany the rise in intraocular pressure. Sometimes one is surprised to find the angle open. Normally, if the darkroom test is positive, exposure to light will produce a prompt reduction of pressure. If the pressure stays elevated, the elevation may not have been due to angle closure after all, but to some other unspecified influence. However, an immediate return of the pressure to normal in the course of a series of consecutive applanation tonometries raises the possibility that it may not have been angle closure but the transient tensing of the extraocular muscles that was responsible for the pressure rise. Elevation in the tension bilaterally might well result from a coincident diurnal variation of pressure so common in open angle glaucoma. A marked unilateral rise, therefore, is more suggestive of angle closure.

When one elicits a positive darkroom test, it is wise not to proceed uncritically with iridectomy, but to select a course of action in view of all available clinical data. Occasionally, when pain and blurring of vision confirm the abnormal tonometry values, there is indeed no question that treatment is imperative, Sometimes, however, the absence of symptoms and the patient's reluctance to submit to surgery make it difficult to know what to do. There is then an obvious, but rarely utilized alternative. It is to repeat the darkroom test after a few days. If the pathologic problem is genuine, the surge in pressure should be reproducible. But if, of a series of darkroom tests, only the first was positive, the patient's conservative instinct probably served him well, for under these conditions, I have never observed angle closure to supervene.

Ernst J. Meyer, M.D.

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On July 1, 1983, the Cambridge Glaucoma Foundation was accorded tax exempt status as a public charity under Section 501(c)(3) of the U.S. Internal Revenue Code, for a period of 30 months. At the end of this period we must provide evidence that we have public as distinct from private support. Readers who would like to help us demonstrate to the Internal Revenue Service that the Foundation has public support may do so by mailing us a contribution of one dollar. Larger sums, though of course welcome, are probably unnecessary for this purpose.

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