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LOOKING AT THE DISC
I

Looking at the disc is one of the most perplexing, and again one of the most interesting, and, when all is said and done, one of the most satisfying activities of my daily practice. If I had to name a single task that prevented the examination of even the normal eye from becoming tiresome, I would nominate the inspection of the optic disc. I have found it an unending source of fascination and that in more perspectives than one.

The optic disc is the intraocular vestige of the optic stalk from which the eye developed in the embryo. It is the anatomic and functional link between the retina and the brain. It is the boundary between the low pressure tissue environment and the sphere of elevated intraocular pressure within the globe. In some instances it reveals a record in physiological archeology as it were, of the pressure history of the eye. Description of the optic disc puts to the test ones powers of observation, ones judgment, ones good sense, and ones intellectual integrity. It gives occasion for examples of astute clinical judgment no less than for naive unwitting parodies of reason. It provides the setting alike for thoughtful, conscientious observation and for rash, overly zealous pretentions to knowledge. To interpret the changes which the disc appears to undergo is a challenge and has on occasion proved a trap for the aspiring ophthalmologic statistician or biophysicist. The obvious fallacies of reason that it has inspired invite comparison with those other disciplines, phrenology and iridology, which systematically require of the observer that he intoxicate himself with fantasy. And there are few structures in the human body that have been more lavishly bedecked with emperors' new clothes.

Given the ubiquity of ophthalmoscopes and the ever increasing numbers of examiners, the profusion of medical periodicals and meetings to communicate new discoveries, one would think that the last word about the optic disc had long ago been confided to the microphone at some national meeting, or appeared on the glossy pages of a refereed scientific publication. But I do not think this has been or will be the case, if only because, at least for the forseeable future, the results of disc examination will not lend themselves to codification or to other mathematical description, any more than will mathematical description disclose the identity of an acquaintance whom we recognize intuitively by looking at the features of his face, an achievement of which the infant is capable long before its grandfather has given up on the research project for parametric description of the disc. The interpretation of the disc of any given patient, therefore, remains a subjective enterprise, a skill that is acquired with practice, that cannot be forced, a power of observation and judgment that increases with the years, until it becomes attenuated and ultimately extinguished with the growth of ones own lens opacities or the deterioration of his macula. These essays, therefore, by no means purport to give the last word on the interpretation of the disc, or to foreclose the possibility of computer-aided image analysis or photogrammetry adding significantly to our knowledge. All I want to do is to report some of the ideas that come to my mind as I look at discs day after day. It also helps, if one wishes to entertain new ideas, to do a certain amount of preliminary intellectual housecleaning, and to try to dispose of various misleading suppositions that make life difficult for the beginner.

As a first step in thinking about the disc, nothing has proved so valuable as simple considerations concerning its geometry and structure, facts that are so elementary as to be almost beneath repetition, but facts that each observer can confirm by looking through his ophthalmoscope and for which he is not dependent on specialized but inaccessible instrumentation. Think of the nerve fibers that converge from all quadrants of the retina onto the optic disc, there to turn backward through a defect in the sclera into the sheaths of the optic nerve. The nerve fibers are transparent, the very phenomenon of vision depends on that fact, and therefore one can often, but not always, see the rim of scleral tissue that is the edge of the foramen. Because such

a large proportion of the nerve fibers come from the macula, there is a heaping up of the fibers at both the upper and lower poles of the disc. The disc appears to be covered with a glistening transparent membrane, which seems continuous with the internal limiting membrane of the retina. It is not uncommon to see hemorrhages beneath the internal limiting membrane of the retina extend onto the optic disc. In the center of the nerve head, at what was once the origin of the vascular supply to the primary hyaloid, there is said to be a defect in the internal limiting membrane. The varying degrees of excavation that are visible in healthy eyes have developed incident to the atrophy of the primary hyaloid artery. Inasmuch as this vessel atrophies to the point at which it branches from the central retinal artery, the depth of the excavation in healthy eyes is most likely determined by the location at which vessels of the primary hyaloid originally branched from those of the retina. The configuration of the cup is in part the expression of genetic traits. Unusually deep or broad physiologic cupping often runs in the family. It is helpful, therefore, when confronted with a youthful patient who has a large central excavation, to find that one of his brothers or sisters or one of his parents has a disc of similar appearance. However, the circumstance that an excavation in an otherwise healthy eye is not corroborated in other members of the family has only very limited diagnostic import.

Two observations suggest that there may be much variation in the depth of the lamina cribrosa, and much variation, consequently, in the orientation of the nerve fibers that constitute the substance of the optic disc. On occasion one sees a patient with an unusually deep and broad optic cup at the bottom of which one gets a glimpse of the lamina cribrosa. It is also not uncommon to see patients with discs excavated and atrophic from glaucoma, where the cup is shallow and the lamina cribrosa is relatively anterior. Thus where the lamina cribrosa is anterior, the substance of the optic disc is thin and the orientation of the nerve fibers is predominantly parallel to the plane of the disc. On the other hand, where the lamina cribrosa is posterior, the substance of the disc is thick and the orientation of the nerve fibers is predominantly perpendicular to the plane of the disc.

Finally, but not least in interest, there are the central retinal vessels which divide on the surface of the disc into superior and inferior branches. The vein is usually collapsed as it descends into the nerve. Between the ribbon-like vein that disappears from view and the distended cylindrical vein that crosses the margin of the optic disc, there is often visible a segment of vein with spontaneous pulsation, a nice demonstration of nature's automatic pressure-reducing valve which maintains pressure in the retinal veins at a level greater than intraocular pressure, and thereby prevents the intraocular pressure from emptying the retinal vascular bed. It is worth noting that the circulation at the upper and lower poles of the disc drains into a more peripheral segment of vein which contains blood under higher than intraocular pressure, while the nasal and temporal poles of the disc drain into a more proximal section of central vein of which one can say with certainty, since it is collapsed, that it is under lower than intraocular pressure. When the intraocular pressure rises, the upper and lower poles of the disc, those areas which show the earliest evidence of atrophy, drain into segments of retinal vein whose pressure is maintained, by the mechanism just described, at higher than intraocular pressure, while the temporal and nasal poles of the disc, those areas which are most resistant to pressure induced atrophy, largely drain into more central segments of vein, which are collapsed and contain blood whose reduced pressure is unaffected by the rise in intraocular tension.

The potential effects of increased intraocular pressure on the optic disc are multiple, and it is important to distinguish between them. In the first place we must remind ourselves that even in the healthy eye, with normal intraocular pressure and without any excavation or atrophy of the disc, there is a pressure gradient through the tissues of the disc. If we could place a transducer on the internal limiting membrane at the surface of the disc and another transducer between the sclera and the nerve fibers as they turn to enter the disc, we should find a pressure gradient, albeit small, which would have the effect of compressing the nerve fibers. It is this difference in pressure which molds the disc and determines the contour of its surface, a fact which becomes strikingly apparent when, with increased intracranial pressure, the gradient is reversed, the excavation disappears, and papilledema ensues.

Another effect of increased intraocular pressure which concerns us is that which brings about the atrophy of the individual nerve fibers. The most widely held theory concerning the mechanism by which increased intraocular pressure causes destruction of the nerve is the hypothesis that increased intraocular pressure deprives the optic nerve head of its circulation, which is derived from the short posterior ciliary arteries and the Circle of Zinn, a supply source of lower pressure than the retinal artery and therefore more susceptible to impairment by pressure elevation. A second hy-

pothesis is that damage to the nerve is simply mechanical in origin. It is thought that with deepening of the cup there is increasing stretch of individual nerve fibers which atrophy as a result of the induced strain. It has also been suggested that nerve fibers might be strangulated in the constricted interstices of a lamina cribrosa that is ballooned posteriorly by chronically elevated tension. We quickly dispose of this notion by pointing out that where intersecting fibrous strands of lamina cribrosa are not free to slide one upon the other, axial distention of these fibrous strands necessarily entails distention, not constriction, of the interstices that they enclose. More to the point is the possibility that it is not impairment of the arterial but of the venous circulation, by a mechanism suggested above, which produces atrophy of the nerve. This possibility would seem to be corroborated by the circumstance that those areas where the venous pressure is demonstrably highest are the ones where cupping begins and where the initial field defects in glaucoma are customarily observed.

Glaucomatous excavation necessarily entails displacement of tissue. The nature of this displacement is a fundamental issue of physics that requires consideration. In the first place, it appears possible that as a result of prolonged tension elevation, the lamina cribrosa might itself become atrophic and that the resultant loss of fibrous tissue might contribute to the volume of the cup. Secondly, it is conceivable that the lamina cribrosa is stretched posteriorly, and that this posterior displacement adds to the volume of the excavation. Attenuation of the lamina cribrosa from either cause would go far to explain the apparently exponential relationship between intraocular pressure and the rate of excavation that was commented upon in previous issues of the Glaucoma Letter. How consistently destruction of fibrous tissue and/or its posterior displacement contribute to cup formation is a question that we must leave to future observation and reflection. There is no doubt about the atrophy of nerve fibers. So far as the loss of nerve fiber substance is not compensated by glial ingrowth, the configuration of the resulting cavity will be affected by the orientation of the nerve fibers destroyed. Where the lamina cribrosa is relatively far posterior and the orientation of the nerve fibers is predominantly perpendicular to the surface of the disc, atrophy of the nerve fibers may be expected to bring about primarily the peripheral enlargement of a preexisting central excavation. As individual nerve fibers atrophy, the intraocular pressure compacts the remaining fibers, the sleeve of nerve tissue lining the posterior scleral foramen becomes thinner, and the central excavation enlarges until it finally occupies the entirety of the opening. In eyes, however, where the orientation of the nerve fibers is predominantly parallel to the surface of the disc, atrophy of individual nerve fibers will lead to a diffuse depression of the surface of the disc, causing it to assume somewhat the shape of a saucer. In this circumstance the glaucomatous excavation develops not by enlargement of a pre-existing "physiological" cup, but by the progression of concavity over the entire surface of the disc. Between these two extremes there is, of course, a spectrum of intermediate cases which combines some of the characteristics of each.

In the light of these considerations, we may now reflect once more on the task of inspecting the disc for glaucomatous damage. The presence of a membrane of undefined stiffness covering large areas of the nerve fibers on the disc suggests at least the possibility that some early and perhaps some not so early nerve fiber loss might remain obscured under a uniform contour. The occasional disc that has an overhanging rim of tissue, projecting like a cornice over a bulbous central excavation, may possibly sustain undetectable loss of nerve substance for long periods of time. Similarly, when glaucoma develops in an eye with a perfectly flat disc, one would also expect considerable loss of nerve fibers to occur before the resulting volume deficit became visible as saucerization. On the other hand, if, as in infantile glaucoma, the elevated intraocular pressure produced a posterior displacement of the lamina, saucerization and even frank excavation might become apparent before there had been any significant loss of nerve tissue. It is important to understand that in the flat disc the first evidence of rim formation may be at the margin of the disc itself.

The identification of disc pathology is not always easy. At each hour of the clock around the periphery of the disc, one assesses its color, and one looks for excavation, of which the apparent discontinuity of a vessel coursing over the margin of the disc is perhaps the single most reliable index. For the examiner who is not an accomplished artist, the best way to record the appearance of the disc, short of photography, is to describe what one sees in simple English prose, as if one were explaining to an inexperienced associate the significant findings for which he should look. The evaluation of the disc in terms of a "cup/disc ratio" is in general unsatisfactory. The only instance to which it is applicable is that of an excavation with a circular margin, everywhere clearly discernable, which is centered upon the disc. Otherwise, a single estimate of the cup disc ratio, derived from but a single meridian, as proposed by some authors, clearly fails to

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do justice to the complexity of the alterations that are visible on the surface of the glaucomatous disc. In its early stages, the glaucomatous excavation is seldom circular and is usually eccentric. It almost always encroaches upon the vertical poles of the disc before it encroaches upon the horizontal ones. More likely than not, the excavation of advancing glaucoma will damage the lower pole of the disc some months or years before it reaches the upper pole, and will reach the nasal margin only much later. Inferotemporally there is often a gradual declivity, a saucer-shaped deformation of the disc which produces a rim only when it is far advanced. In this situation, the notion of a cup/disc ratio is meaningless, and the requirement that it be estimated is an invitation to fabrication.

Of particular interest is the description of incipient glaucomatous cupping in a disc that was previously entirely flat, where cupping progresses as a saucer-shaped deformity which depresses the entire surface of the disc without fashioning any rim of nerve tissue whose diameter one might estimate. When finally an edge becomes apparent, it is a rim of sclera, not of nerve fibers, and then the damage that one would have liked to prevent with timely treatment of glaucoma has been done. In this situation, the attempt to assess the development of glaucomatous atrophy by calculation of the cup/disc ratio will be of no help. It may indeed make for a false sense of security and the examiner who dismisses his patient as having "ocular hypertension" with a pressure of 30 and a C/D ration equal to 0, may be unpleasantly surprised when he encounters his patient some time later to find a C/D ratio equal to .9 and a substantial field defect.

Without disparaging the contributions which many investigators have made to our understanding of disc pathology, it seems fair to note that few authors who write on the subject acknowledge with sufficient candor that in many situations the condition of the disc cannot be unambiguously interpreted. Then the examiner's duty is, above all, to refrain from claims of precision which cannot be sustained. In that situation, the experienced examiner accepts the appearance of the disc as an unknown, and deals with this unknown factor as he would with a dependent variable in an algebraic equation. Under these circumstances extraneous information, for example, the presence or absence of a field defect, a history of elevated intraocular pressure, the appearance of the disc in the fellow eye, comparison with photographs on prior occasions, and comparison with the appearance of the disc in siblings or parents will all contribute to the elucidation of the unknown.

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