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THE CAMBRIDGE GLAUCOMA LETTER

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GLAUCOMA MODELS

On the trip through medical school one picks up, in addition to other items of truth, various truisms which come in handy when one practices medicine. One of the more memorable of these platitudes urges you to "listen to the patient, and the patient will tell you the answer."

In the late autumn of 1978 there came to my office an old man, who told me he was born in 1892, had good vision all his life, and asked me to check his eyes. The vision was satisfactory for his age: 20/20 in the right eye, and 20/30 in the left. The left lens had some cortical changes which I thought might account for the decreased acuity. When I looked at the fundi with the direct ophthalmoscope, I saw that the discs were asymmetrical. The right disc was pink and flat. The left disc was somewhat pale and had a shallow concavity which extended toward the lower pole. The visual field was full to 3W/1000 on the tangent screen. The intraocular pressure was 26 in the right eye, and 32 in the left.

"Pretty good eyes for my age, wouldn't you say?" he began. I evaded the question. "I think you may have glaucoma," I began somewhat tentatively, because I had a feeling he would not take well to the news. "Glaucomia," he said, "So you say I have glaucomia." I was encouraged. He seemed more prepared than I had anticipated to accept the diagnosis. "I am afraid so," I said gravely. I thought he needed to be impressed with the potential seriousness of the problem.

He remained silent for a moment while my typewriter clattered away as I composed my notes. Then he said, as if awakening from a dream: "I never knew I had glaucomia." "Most of the time, patients don't know when they have the disease," I explained. I thought he was really beginning to understand. He lapsed into silence again as I continued making my notes. Then he straightened up in the examining chair, lifted his chin and said with deliberation: "I don't want it. Take it out!" I was embarrassed, but the old man was now too excited to notice. "Take it out?" I asked tentatively, "I don't understand what you mean." "I want you to take it out," he said, now almost shouting. "I want you to take the glaucomia out of my eye." "I can't take it out," I said emphatically. "Well," he demanded, "where is the glaucomia?" "In your eyes," I said. "The right eye or the left eye?" "I think in both eyes." "Take it out," he said. "I wish I could," I replied, "But it can't be done." "Why not?" he demanded. "Because it isn't the kind of thing one can take out." "But you say it's in my eye." "Yes." "And you say you can't take it out. Some doctor you are!"

He fell silent for a few moments, and I was at a loss how to resume the consultation. The old man was thinking. What would he think of next? "What is glaucomia," he began again," Tell me, doctor, just exactly what is glaucomia?" I felt relief. It was a question to which I had the answer. It had been asked of me, and I had answered it, many times before. "Glaucoma, " I began, "is a disease where the pressure in the eye is elevated over long periods of time, and this elevated pressure causes a wasting of the optic nerve that passes from the eye to the brain, and the wasting of the optic nerve causes a gradual loss of the field of vision." "How do you know that?" he asked searchingly. "It's what I have been taught." "Have you seen it yourself?" "Seen what myself?" "Seen the pressure in the eye make people blind?" Before I had a chance to answer, my patient sat upright in the examining chair again. "They told my nephew he had glaucomia, and they said it was from diabetes. A woman in my apartment house had a terrible pain in her eye. She went blind, and they said that was glaucomia. I know another fellow was in a car accident and cut his eye and he got glaucomia."

I looked at my patient as I tried to find an exit from the verbal maze that had us both trapped. But just at that moment, he fastened onto my gaze and said firmly: "You are just talking. Glaucomia is just a word. None of you doctors knows what glaucomia really is."

We do and we don't. Our minds are filled with formulas and phrases which we take for granted, and usually we act upon

them, as in a trance, without thinking about what they mean. The more complex the concepts which we memorize, the more valuable it becomes to challenge them with our own experience. Concepts, if they are to remain meaningful, require continuing reinterpretation in the light of the experiences to which they refer. Consider how, in moments of reverie, concepts may become transparent, letting us glimpse, if only transiently, the diaphanous memories that they summarize. At such instances the most familiar of symbols, sometimes even the letters that represent ones own name, can dissolve before ones eyes, and one reflects, not without some embarrassment, on the incongruity that an arbitrary assemblage of symbols could have come to mean so much.

From such reverie, one awakens with an uncanny awareness that the name which we give to a disease, if not indeed the very concept of disease itself, however useful it may be as a pointer to our experiences, tends also to function as a screen that conceals from us what is really going on. There is no more telling evidence of how little meaning inheres in our nomenclature than the promiscuousness with which we attach to newly identified diseases as eponyms the names of their discoverers, names that tell us nothing, scrawled like graffiti on the walls of the labyrinth of our not-knowing, a labyrinth from which we seek to escape in vain. A name is but a label by which we manage our affairs, and sometimes when we are unable to manage them as well as we might like, it pays to check on the label, and to make sure that the words which we use serve to some purpose better than to confirm ourselves in our old prejudices.

It helps to look back and to reflect on the many different meanings that the term glaucoma has assumed through the ages. The Greeks invented it, but we know what they meant by the term "glaucoma" no more than we know how they pronounced it. For over two thousand years, doctors have written about glaucoma. Few authors have used the word to mean the same thing, and prior to the past twenty-five years none has meant by it what we mean by it now. We speak with such confidence as if we had succeeded in correcting all the errors of the past. Implicit in our self-assurance is the assumption that although the past was wrong, we are right. The past appears to us as an ever lengthening chain of error which, as we grasp its end, changes to truth in our hands.

Our professions commit us to a stance of confidence in our knowledge. Rare indeed is the patient willing to entrust his eyesight to a doctor who admits his ignorance. But if we succeed in divesting ourselves of the magicians' and soothsayers' robes in which our patients insist on seeing us, we discern from history that only a fraction of what we think we know endures. The rest is seasonal. Most of the trees of knowledge are deciduous. Their buds turn into leaves, their blossoms become fruit. The leaves wither and the fruit decays. In due time both fall to the ground to nourish the next cycle of learning. As we review what we "know" about glaucoma, we should try, if we can, to identify those elements which will remain after this season's fruits of knowledge have fallen.

It is difficult to dispense with the notion that there is a set of distinguishable diseases which it is the investigator's task to discover, and the clinician's task to diagnose and treat. Yet if we succeeded in doing so, we might save ourselves much trouble. We would no longer need to consider, for instance, whether glaucoma is one or many, or whether certain phases of glaucoma should be denominated as ocular hypertension, whether there is a plateau iris configuration or only a plateau iris syndrome, whether there is ciliary block glaucoma or aqueous diversion syndrome or malignant glaucoma, or all three, whether there is anterior ischemic neuropathy or low tension glaucoma.

Instead of busying ourselves with scholastic distinctions concerning the definition of glaucoma, we would do well to borrow from the mathematicians and from the engineers and from the memories of our own childhood, the notion of making models, not in this instance, physical models of boats or airplanes or trains, but logical models, maps as it were, on which we define the boundaries that we have surveyed and the terra incognita that remain to be explored.

Conceptual models do not add to our knowledge directly, but they help us to cope with our ignorance. Like maps of the ground, they are the crudest of approximations, yet often indispensable in enabling us to find our way through an unfamiliar landscape. The important characteristic of such models is that they are not expected to coincide with experience, but only to anticipate it. Implicit in the modeling process is the awareness that we are not in possession of the truth, the admission of ignorance which is the hallmark of all valid knowledge. A model is never more than a tentative expression of our understanding. It is only an instrument by means of which we try to come to terms with reality. No sooner is it devised than we begin to test it, and we

modify it as experience dictates. A model of glaucoma was, however, not helpful in satisfying my 86 year old patient who wanted me to remove the glaucomia from his eye.

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A MODEL OF THE OPTIC NERVE IN GLAUCOMA

The conventional nosology of glaucoma purports to provide invariant categories to account for all components of the broad spectrum that we recognize as the pathology of glaucoma. The categories are artifacts. They serve at best to generate vacuous arguments about the nomenclature of the disease. At worst they confuse the diagnostic issues and confound the goals of therapy. Instead we propose a set of simple pathophysiologic models with statistical parameters, some of which must remain conjectural, and some of which are susceptible to empirical confirmation.

We begin by reflecting on the diversity of what we observe. There are patients with normal pressures, whose discs remain healthy and whose fields remain intact. There are patients with elevated pressures whose discs show gradually increasing excavation and progressive field loss. But there are also patients who have elevated tensions, who, at least over a limited period of observation, develop neither excavation or field loss. There are patients with apparently glaucomatous field loss who have neither elevated tensions nor disc excavation. There are patients whose discs seem extraordinarily resistant to increased pressure, where excavation supervenes only after years of tension elevation, and there are those whose discs seem to be abnormally sensitive, where progressive cupping of the disc occurs after only brief periods of slightly elevated intraocular pressure or none at all. There are those where the process of excavation ceases once the pressure has been lowered, and there are those where field loss continues inexorably even though the pressure has been reduced to normal levels.

Given this diversity of phenomena, it is no wonder that there is no uniformity of views on glaucoma. The traditional means of coping with this logical complexity has been to distinguish on the one hand between patients who are normal and who have no disease, and on the other hand, patients who are ill and are afflicted with any one of a number of separable disease entities. Patients are labeled with the names of diseases, notwithstanding the admitted fact that the expression of the hypothetical disease in different individuals may not be at all comparable. Disease names are dealt out to patients like cards to players in a poker game; some patients end up with a very bad hand. Thus is invented the set of all patients who "have" open angle glaucoma, another set of patients who "have" angle closure glaucoma, yet a third set who "have" pigmentary glaucoma, pseudoexfoliative glaucoma, inflammatory glaucoma, and so forth, until all our diagnoses have been exhausted and all patients are accounted for.

The nominal description of glaucoma has the following defects: 1. It fails to reflect variations in the severity of the disease. It offers no explanation for the fact that one patient with glaucoma may go blind in a few months, while another patient, presumably with the same disease, may keep his vision for many years. 2. It fails to describe the coincidence of pathophysiologic mechanisms, as for example, when a patient with elevated intracranial pressure as from chronic emphysema tolerates without disc excavation elevated intraocular pressures which would leave the discs of most other patients badly cupped. 3. It postulates an absolute qualitative difference between the healthy and the diseased eye, and is incapable of explaining "borderline" conditions. Inasmuch as patients are assumed either to "have" or not to "have" glaucoma, the nominal description cannot account for incipient, latent, or preclinical forms of the disease.

The modeling process begins when instead of labeling them with the names of diseases, we observe our patients and attempt to discern patterns in what we see. Such patterns reflect not only the characteristics of what is seen but the characteristics also of the mind that does the seeing. We look for entities, "something to hang your hat on." We look for objects to which we can give names. We look for temporal sequences that appear quasi purposeful to construe them as cause and effect. We try to understand the processes which we observe in terms of elementary physical relationships. Indeed, glaucoma fascinates because it appears amenable at least on the surface to simple causal explanations in terms of fluid dynamics.

It is common to observe in a single eye the coincidence of elevated tension, optic nerve cupping and field loss. We see this triad so often, that we infer its members to be causally related. Our understanding of the anatomy and physiology of the optic nerve suggests to us that the field loss follows the excavation of the optic disc and that this excavation in turn is a result of elevated pressure.

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That is our model. The model states that intraocular pressure of a given value acting over a given period of time brings about a certain degree of excavation of the optic nerve head; and when this excavation involves the disc up to its margin, there develops a nerve fiber bundle defect involving those fibers that are adjacent to the area of excavation. This is not the description of a disease. It is the model of a pathologic process. It sheds light on the problems of many patients. It gives a sufficient explanation for none.

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This model provides us with a framework for thinking about one of the central problems of glaucoma. It offers tentative answers to some questions, but it raises even more. The questions which it raises are most readily stated in mathematical terminology. We hypothesize that field loss is a function of disc excavation, and that disc excavation in turn is a function of the destructive effect of the intraocular pressure. This destructive effect of the intraocular pressure is almost surely a function of the height of the pressure on the one hand and of its duration on the other. It is probably also a function of the radius of the globe and of the thickness of the disc. The resistance of the disc to pressure-induced excavation and the resistance of the nerve fibers to atrophy are two seemingly independent variables that are potentially susceptible to calibration even with presently available instruments.

The immediate consequence of this manner of thinking is to make explicit what we do not know about the glaucomatous eye. Specifically, we do not know the statistical distribution of values for time and pressure required to bring about a given amount of excavation. Is the relationship between the pressure and the rate of excavation linear or does it resemble an exponential or a power function? Even more basic is the question, whether over the range of time and pressure that we observe, the function relating these to the excavation of the disc is discontinuous or continuous, whether, in other words, there are critical turning points either in the value of the time or of the pressure at which one would expect to see a drastic change in the behavior of the eye. The model does not answer these questions, but at least it makes it possible to ask them.

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