

## **The Patient's Journey: Glaucoma [as supplied by authors]**

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### **Glaucoma: An overview**

Glaucoma is an eye disease characterized by the death of the nerve that connects the eye and the brain. In contrast to some other diseases of the eye, such as macular degeneration or cataract, the eye continues to receive input from the environment. The problem is that the transmission pathway between the retina and the brain slowly dies, causing areas of nothingness within an individual's visual field which slowly expand. Although glaucoma is a well studied phenomenon, its etiology is still a mystery, and a cure remains elusive. All conventional treatments, which include medications, laser treatments, and surgeries, are designed to lower the pressure inside the eye (otherwise known as "intraocular pressure" or "IOP"—there is a separate pressure in each eye). In the United States, most people with glaucoma have high IOPs, although elevated IOP alone does not necessarily indicate that someone has glaucoma. Approximately one third of glaucoma patients develop the disease at IOPs that are within the statistically average range – a condition known as low tension glaucoma. I, Christine (Tine), have this type. I am continually frustrated that although all experts agree that I have this disease, they cannot explain why or offer any satisfactory treatment.

### **My good news and bad news**

"You have glaucoma," an ophthalmologist informed me when I was 36. Given the uncanny ability of the human brain to compensate for missing visual information, I don't know when I would have noticed my blindness had I not had some unexplained pain in my right eye one morning. As I lay in bed with my hand over my right eye, I looked around the room using only my left eye. I noticed that when I gazed straight ahead at the dresser, I couldn't see part of the closet door. And when I moved my gaze to the closet door, part of the room door disappeared. For the first day, I tried to put it out of my mind, but when the symptoms persisted, I began to get very nervous.

A week later, I sat in the dark examining room, stunned. Tears came to my eyes despite my desperate attempt to appear in control. “You’re young to have this,” explained the doctor. “But the good news is that there are lots of medications to treat it.” Without looking at me he added, “It’s not something to be too concerned about.” He glanced instead pointedly at his watch. Was I going to become emotional and make it difficult for him to leave the room? Ignoring his impatience, I persisted and asked whether there was anything else that might be causing my vision loss, anything we might want to rule out. He shrugged, “Well, it could be a brain tumor. We could do an MRI, but...” His entire body language finished the sentence: “why bother?”

Why bother, indeed? They were not his eyes, and he was sure of himself and his diagnosis. But they *were* my eyes. When I got home that afternoon I immediately began calling around and soon had an appointment with the best specialist I could find. After a thorough examination, I sat before the ophthalmologist whose specialty was vision loss from brain tumors. He told me, “Well, we’re going to do an MRI, but I’m sure you have a brain tumor. All your symptoms suggest that.” The MRI was duly performed and two weeks later was pronounced to show no abnormality. I happily returned to being a glaucoma patient. It sounded good after the words “brain tumor.” What a relief. It’s *just* glaucoma. There are lots of drugs to treat it.

Since then, I have been lucky to find a physician who is able to look into my eyes and still see the human being, not just the diagnosis. Doug respects me both as a patient and as a colleague (my department and his were affiliated with the same university). In the beginning, we both approached my situation with optimism. There *were* a lot of drugs to try. When the first one had no effect on lowering my IOPs, we moved on, unconcerned. His glaucoma clinic was only a block from where I worked, and I came in for frequent appointments. We tried more and more drugs, but with no effect and with an increasing sense of desperation on both our parts. I began coming in every ten to fourteen days and learned the names of all the clinic staff members, who were kind about squeezing me into the schedule when I had to run back to a meeting. One day after a few months of unsuccessful treatment I stood in the hall outside an examining room and one of the staff members handed me another patient’s chart. “Could you take this down to surgery for me?” she asked. I looked at her. “Where?” “Surgery. I just don’t have the time right now.” “You want *me* to take the chart?” Luckily someone else overheard this conversation and rescued me. Embarrassed apologies followed. Apparently, since I was so much younger than the average patient (the mean age of onset for glaucoma is 65) and the woman was so used to seeing me around, she had thought I was one of the trainees who frequented the clinic.

Four years later, we still haven’t come up with an effective treatment for my condition. The disease is affecting both eyes, though my left eye is much worse than the right. I don’t take any medications, because none of them work. I underwent a laser trabeculoplasty in my left eye, with no effect. I have my eyes examined every three months, making the trip to Boston from Philadelphia, because Doug has switched universities but I have chosen to continue to see him. At this point we’re just gathering data points to discover what the slope of the line of deterioration might be. I have had a number of optic nerve haemorrhages, which are highly correlated with progression, but we’re not sure how rapidly the disease is going to progress or how much sight loss I will tolerate before I avail myself of surgical options. Up to now I have been reluctant to consider surgery because of the potential for complications and the risks involved because my eye pressure is already low.

### **How I travel alone towards an uncertain future and sometimes lose the path**

When I try to explain what it’s like not to see with glaucoma, I tell people to imagine a digital TV screen with pixels, and to imagine that some of the pixels in an area have ceased to

function. The entire picture still exists and moves, but there are areas which simply disappear from the screen. There is no shadow, no light, no color—just nothing. Unusually and unfortunately, in my case the malfunctioning pixels are near the middle of the screen.

When I try to explain what it's like to *live* with glaucoma, I don't have such a simple analogy. Part of learning to live with any chronic, debilitating, progressive condition has to be about learning to live with fear of the unknown. In the case of glaucoma, deterioration takes time, and even if my journey results in complete blindness, it will be years down the road. And yet neither the loss I have now nor the possible loss in the future is ever completely out of my thoughts. The fear climaxes every time I have to take a visual field test. Sitting in the darkened room, most often with someone else on the machine next to me, I listen to the speed of the other patient's clicking. The more points of light you can see, the more you click, a competition of noise. When my left eye is being tested, I have to remember to breathe. Was that a light? Should I click? Surely *that* was a light? Am I clicking less than last time? Because my brain perceives less light through my left eye, after a few minutes even keeping it open becomes a struggle. They tell me they can tape the eye open during the test if it becomes too much of a problem.

Because my own future is uncertain, so is that of my closest relatives and friends. Who will take care of me if I become unable to live without help? I told my fiancé on our second date about my condition, not because I was so certain at that point that our relationship would be life-long, but because I could not ethically justify to myself revealing such a potential impediment at any later point. Would he walk away? Would he accept me regardless? I had to give him the choice, although I have none.

At work I have started paying for the most comprehensive long-term disability insurance. I am lucky that my career is flexible enough to accommodate me thus far. The other problems are still surmountable. At the moment, I can't see the top of the steering wheel when I am driving. When I bought a new house last year, I picked one near a train station and in a town where we can walk to everything, because one of the first things I will do if my vision loss progresses much further is to take myself off the road. I can't see my own feet when I walk, so I always hold on to a railing or someone else when I descend. I can't see people's mouths move when I stand a comfortable distance away from them during a conversation, so I sometimes have trouble hearing. I often can't see gestures people make with their hands. I can't see my fingers as I type this. But what's most distressing is that I will never again be able to see, up close, the entirety of someone's face. Nevertheless, at this point I still have many more active pixels in the screen of my vision than dead ones.

### **My companion on the journey (Doug Rhee's perspective)**

I am Tine's ophthalmologist. When I first met Tine, she had seen a few colleagues who had correctly diagnosed her low tension glaucoma and excluded other potential causes. She already had significant damage in both eyes with moderate (by conventional standards) visual field loss. I thought that it was unusual for someone as young as she to have developed low tension glaucoma; however, the diagnosis was clear.

I started out quite optimistic, as we have numerous treatments. I became more despondent as each medication we tried and added failed to have any effect in lowering her IOPs. In my experience, low tension glaucoma does not respond to medications and lasers as well as glaucomas that are associated with high IOPs. And despite the low likelihood of complications from surgery—our next option—the risks are real. Low tension glaucoma can be more difficult to treat exactly because the IOP is already within the average range. Very low IOPs achieved through surgery (generally less than 6mmHg) are often not well tolerated and have the potential to harm the eye more quickly than the glaucoma itself.

Studies estimate that approximately 25% of people with glaucoma receiving the state-of-the-art care will still go blind in one eye and 10% will go blind in both eyes (i.e. functionally blind) despite all our technology. I choose to specialize in glaucoma, in part, because of those daunting statistics: I want to help to stop the harm from the disease, or from the treatments themselves. However, doctors are susceptible to marginalizing patients who “fail” our treatments, because there is the potential to view the situation as personal failure. Avoidance of uncomfortable feelings is a natural human tendency and doctors are people, too. Nevertheless, even after treatments fail or someone goes blind, there is still the potential to offer healing. The patient’s life still goes on, and perhaps that is when they need us the most. The patient needs a physician (or healer, rather than a simple medical technician) to help them deal with the grief and ultimately move on to the next phases of life. Tine’s description pointedly shows that the harm from glaucoma is not limited to functional alterations necessitated by visual field changes but also includes the social and psychological aspects of having a medical condition.

I have spent my career trying to convey my genuine compassion and be a good healer. However, Tine’s story made me understand how limited I and others physicians are, in some cases because of the time pressures induced by a managed care system, and in most cases because we often don’t fully understand what it is to have glaucoma. Few people can describe the symptoms of a visual field defect – i.e. the nothingness – except in the very end stages. Even if we have heard the descriptions of “fog” or “misty” (or worse yet “dark”), we never truly know about the actual impact of these visual deficits. Tine’s detailed and emotionally open descriptions made the condition more real for me. I have continually striven to understand from patients the impact of glaucoma, and I even lecture to physicians on what it is that patients “see” and the physiologic basis for compensatory mechanisms, yet I did not have a complete picture until I heard Tine’s testimony.

Tine and I have walked hand in hand, trying to pursue treatments that make medical sense and that also work with Tine’s overall life-plan. I am relieved that her rate of decline has been very slow, giving us time to wait for new discoveries rather than forcing our hand towards a surgical method to lower her IOPs. I stand ready to take the next step of conventional glaucoma surgery if we both decide that this has become necessary. Until that day, I continue to endeavor in the laboratory to help develop some of those new treatments to someday help Tine and others like her. Although we save the sight of most, we still need to do a better job.

### **A patient’s ways of coping**

I do question why I was not diagnosed earlier. I have myopia and have been to see an ophthalmologist almost yearly since I was ten. During the regular eye exams I was sometimes even “tested for glaucoma”—that is, tested for elevated IOPs. But I can only remember receiving one dilated eye exam in my life, when I was in my late teens. And no health practitioner ever asked me simply to close one eye, look at a fixed spot in the room, and notice whether anything was missing.

Fortunately, up to now I have been able to live without much aid. I have considered joining a glaucoma support group, but given the age of the typical glaucoma patient, I am hesitant. To calm my own fears, I empower myself by learning as much as possible about the disease’s theories, treatments, and outcomes. I work at a medical college, so I have ready access to and read all the glaucoma literature. I have not been reticent to seek second opinions, for which I have flown to Florida and have telephoned an expert in Switzerland. I have sought out complementary and alternative treatments. For instance, I have been to see a national expert in ways of treating the eye with traditional Chinese techniques and have been taking the Chinese herbs he has prescribed for a number of years. But there is no way to judge whether unconventional therapies are having any beneficial effect, and there are

distinct limits to what I will try. Nevertheless, all of these things I do to remain somewhat in control of a situation which is almost completely out of my control. Over the long run, I believe the only thing I can do is to remain the expert about my own eyes.

### **What I need along the way**

I have used the Internet to gather basic information about glaucoma. Here are some of the websites I have found most useful:

<http://www.nlm.nih.gov/medlineplus/glaucoma.html>: provides links to many other sites on specific glaucoma-related topics

<http://www.glaucomafoundation.org/index.php>: gives an overview of the disease, provides answers to common questions, and provides links to other sites

[http://www.nei.nih.gov/health/glaucoma/glaucoma\\_facts.asp](http://www.nei.nih.gov/health/glaucoma/glaucoma_facts.asp): defines the disease and its causes, symptoms, and treatment and contains links to publications

<http://www.glaucoma.org>: provides information about glaucoma, living with the disease, and treatments, as well as an overview of some current research

### **My journey's end**

At any age increasing vision loss is incredibly frightening and debilitating, but facing this prospect so early in life adds a layer of complexity. Nevertheless, I have hope for the future: hope that understanding of glaucoma will progress to the point that treatments will no longer be shots in the dark, hope that the medical system will increase its flexibility and support of those of us who have to visit clinics frequently for chronic conditions, and hope that education of medical personnel will increasingly emphasize a more holistic perspective. The uncertainties of the present I intend to confront with confidence in my own abilities and the help of the companions on my journey. After all, glaucoma patient or not, I still have a life to live.

Competing interests: None declared.