The wayward eye

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Professor Power, sixty-three years old, was of slightly less than median height and more than slightly over his ideal weight. Indeed, the margin by which he escaped the dread category of "obese" of his body mass index was uncomfortably narrow, even for him, and he was never one to be much preoccupied with his physical appearance. Like his father before him, he was fond of lamely joking that he was "too short for his weight." That was not the only characteristic he had inherited—the male pattern baldness gene had long ago left him with a monkish tonsure, and the hair pattern, coupled with oversize glasses for his myopia, gave him an owlish appearance, which he imagined made him seem wiser than he really was. He therefore cultivated it. He reasoned that for someone in his chosen profession it could do little harm and might sometimes work to his advantage. His students, however, were not fooled.

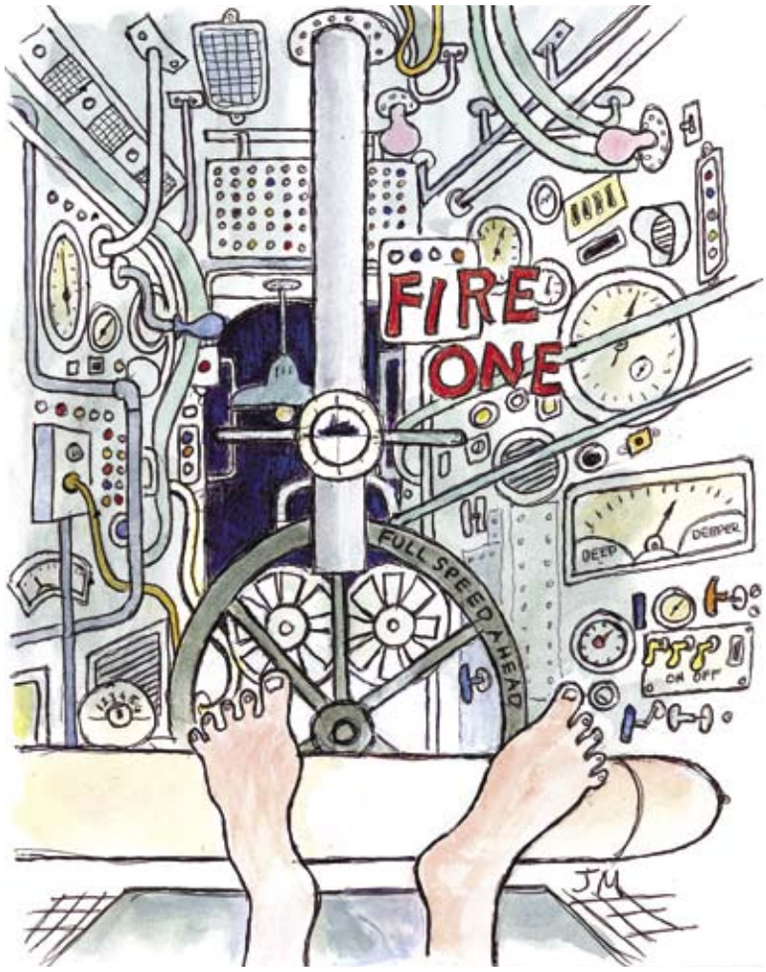
For someone approaching retirement, Power was in reasonably good shape. In the relatively near future, he would need a cardiac pacemaker. His pulmonary function would become the physiological equivalent of getting by on roughly one lung, the result of thirty years of a pack-a-day cigarette habit. When he finally quit, he had promptly developed asthma, which gradually deteriorated over the years into chronic obstructive pulmonary disease. He had had a very mild stroke, necessitating daily warfarin. Then there were the occasional bouts of gout, eczema, and gastro-esophageal reflux disease. He had recently undergone a prostatectomy, and his internist was starting the long search for an appropriate and effective antihypertensive.

Among a handful of other physicians, Power had for several years been under the care of ophthalmologist Dr. Hasselback. Dr. Hasselback was a brilliant, if somewhat remote, caregiver who managed to maintain a productive and high visibility research program along with his busy practice. That suited Power just fine. He always claimed that he wanted "a physician who knew something, and the heck with the bedside manner." Hasselback was said to be particularly adept in the surgical suite, where his skill and facility at implanting artificial lenses was generally admired. Dr. Hasselback had been keeping a careful watch on two small cataracts in Professor Power's eyes. The one in the left eye was growing, whereas the one in the right eye seemed to be stable. Hasselback was very careful to explain that Power was the sole decider about the course of action to pursue. Hasselback recommended doing nothing so long as Power's vision was correctable to about 20/20, or until Power himself began to notice some impairment. Perhaps it was the mere knowledge that the cataracts existed, and the vague feeling that something ought to be done about the one that was growing that led Power to decide finally to have the lens in his left eye replaced. The surgery was scheduled for late January of 1995.

It was not an easy decision. The very thought of a surgeon slicing directly through one's eyeball is enough to make even the most stoic of patients a little queasy, especially since general anesthesia is not usually used. The eye itself, of course, is rendered insensitive to pain with a local anesthetic, but the idea that the patient in theory could look directly at the scalpel as it descended to do its work was more than a little unnerving. Not to mention the needles and sutures that would be used to sew the incisions back together. In brief, Hasselback would cut through the cornea to the capsule that contained the natural lens. That lens would be extracted through another incision and a synthetic one inserted in its place. Both incisions would then be sutured.

The doctor says, "Good surgery."
The patient isn't so sure.

The operation went far better than Power could ever have imagined. Although it was true that he was not fully anesthetized, he had been given enough "chemical happiness" in the form of anti-anxiety medication to turn even the meekest of mice into a lion-hearted daredevil. It was completely painless, and the worry about watching the operation through the very eye that was being abused proved groundless. Recovery was a little tedious because of the several types of eye drops administered at frequent intervals. Perhaps the worst part was looking



at the recovering eye in the mirror. It was stomach-churningly bloody and somewhat black and blue, but that was easily concealed from others by dark glasses. The final result, however, as soon as Dr. Hasselback felt that recovery was complete was a little disappointing. Power could tell no difference at all as a result of his new lens. But Hasselback declared the operation an unqualified success, and was extremely pleased with the quantitative examination results. As pointed out by Hasselback from the beginning, the operation would not obviate Power's need for glasses because of the myopic and astigmatic condition of the other eye.

A touch of ptosis, then decreased vision, then diplopia

About two years later, Power woke up one morning with his right upper eyelid drooping halfway over the eyeball. He was pleased that he knew the name for that condition—ptosis. It can be an early sign of myasthenia gravis, but in that case it is usually symmetrical, involving both eyes. Perhaps, he thought, it is some sort of local infection or inflammation, and he treated it himself with warm compresses. It gradually resolved. At odd moments Power would remove his glasses and focus first one eye and then the other on some photographs at the far end of his office. He still could not discern any improvement in the eye with the artificial lens, but his overall vision seemed completely satisfactory. Certainly, there was no deterioration in his vision that he could tell, until one day about a month later, when he noticed that the vision in his right eye—the one that still had its original lens—seemed to be a little worse than it had been. The next week it was even worse, and a few days later still more deterioration had occurred. Power seemed to be losing vision in his right eye at an alarming rate. He could barely make out the faces in the photographs. He was also experiencing diplopia, at first episodic, and then nearly constant.

The ophthalmology service in his local hospital had been overextended for years. A new patient could anticipate a wait of six to nine months for an appointment. When Power called for one to have his eye evaluated, an appointment six months in the future was suggested. After several more days in which he had further loss of vision, he called and asked for an emergency examination. After some negotiations between the appointments secretary and Dr. Hasselback, the ophthalmologist agreed to see Power over his lunch hour. At the appointed time he entered the examining room carrying a sandwich and soft drink as if to indicate, "This had better be good." Perhaps he even felt that Power was questioning his professional competence by imagining that the operation on the left eye had somehow caused a problem in its untouched counterpart. Power had reason to believe, however, that the examination itself was every bit as thorough and meticulous as he was accustomed to receiving. The intraocular pressure was checked, and Hasselback spent some time examining the retina. At the end of the exam, Hasselback pronounced judgment: "There is absolutely nothing wrong with your eye." Power was totally crushed. Hearing that there is nothing wrong with you when you are convinced that you are at death's door is almost as bad as hearing that you have an unsuspected fatal disease. He slunk out of Hasselback's office mouthing abject apologies.

Back in his own office, however, Power found to his horror that the vision in his right eye was now almost totally gone. How can that be? And, more importantly, what can he now do about it? There was no use calling Hasselback again. Even if he could get a timely appointment with another ophthalmologist, it was unlikely that anyone in that department would want to put him- or herself in the position of second-guessing a colleague. Then he thought of his genial optometrist, and Dr. Blain promptly made special arrangements to see him. Power

told him the whole story. Blain had him read an ordinary eye chart, and the results were clearcut. There had been a very significant deterioration in the vision in Power's right eye since his last examination. Now it was up to Blaine, a humble optometrist, to convince Hasselback, a somewhat imperious opthalmologist, that something was definitely wrong with Power's eye. That must have taken courage, but he did it. Hasselback referred Power to the neurology service that very day, and Power began to rethink his assessment of the value of more cordial and intimate doctor-patient relationships.

MRI of pseudotumor of the orbit.
Courtesy of Nancy J. Fischbein, MD, associate professor of Radiology, Stanford University School of Medicine.

Why neurology?

Although he was relieved not to have been referred to psychiatry, Power was somewhat puzzled by why he was handed over to the neurologists. Within hours, however, he found himself being examined by Dr. Hatfield, the neurology chief resident. Many of the tests performed by Dr. Hatfield seemed to have no relevance to his vision, but Power was so pleased to be receiving some attention that he complied without question. Indeed, Dr. Hatfield's fascination with his case was a little flattering. For a chief resident, he seemed to have little else to do except to take extraordinary personal care of Power. He brought in a constant stream of fellow residents to examine him. By now it was getting rather late in the day on a Friday afternoon, and many hospital personnel were leaving for the weekend. It was then that Dr. Hatfield casually remarked that Power should be admitted at least overnight, and that Hatfield had made emergency arrangements for an MRI image of Power's head.

As the possible significance of this suggestion slowly dawned on Power, his blood turned to ice water. My God! I've got a brain tumor! I'm a goner for sure. All those years of smoking must have resulted in unsuspected lung cancer, and it has now metastasized to my brain. It must be malignant. I wonder how long I have? How are my wife and kids going to function? With those thoughts continuously circling in his brain like an electric train on a track, he forced himself to lie as still as possible inside the claustrophobic chamber, so like a torpedo tube, into which he had been inserted for the scan. The machine made loud clanking noises as if it was indeed a submarine. Power giddily chanted, Let's all go on the yellow submarine. Fire one! Fire two! Damn the torpedoes—full speed ahead. All that without benefit of medication! After what seemed an eternity, the clanking stopped and Power resurfaced. In the meantime, Hatfield had made arrangements for the radiologist on call to come in and look at the images after they were developed. "MRIs can be a little tricky to interpret," he said.

In the dead of night Hatfield was back. "Actually, it's good news," he said.

Wonderful, thought Power. That must mean it's benign or operable, or even benign and operable.

"What you have is an unusual condition called a pseudotumor of the orbit.[1,2] Not only that, but you have a rare variant of that unusual condition. Yours is painless. Usually, the loss of vision is accompanied by a sudden onset of pain. In addition to the confirmatory MRI results, one of the diagnostic clues was that you are prone to atopic diseases such as your asthma and eczema. The etiology of pseudotumor of the orbit is obscure, but it is believed to have an immune basis. The orbit of the eye is a bony cavity that contains the eyeball. In a pseudotumor, an inflammatory infiltrate accumulates in the orbit to occupy space and compress tissues, including the optic nerve and muscles controlling movement and axis of vision of the eye. Compression of the optic nerve, while usually self-limiting and benign, can in some cases lead to serious ocular damage and possible loss of vision. The mass-lesion effect is similar to a true tumor. Since the condition is external to the eyeball, in the early stages at least there are no visible pathological lesions on the retina.

"But it gets even better. This condition is almost invariably reversed quickly and permanently with a single course of treatment with an anti-inflammatory steroid like prednisone."

Prednisone, of course, thought Power, beautiful, wonderful, lovely, magical prednisone. The drug that has cured my asthma flare-ups so many times. What better remedy for a disease of unknown etiology than a drug with an incompletely understood mechanism of action? And only thirty-six hours later he was sitting up in bed trying out his newly cured right eye and thinking, Wow, that red-headed nurse across the hall is really hot!

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