

Little Men with Big Teeth

The patient, Evelyn Fuller, a frail, elderly woman, the widow of a farmer and mother of three who still lived alone in the family home, was brought in for neurological consultation because of visual hallucinations after telling her daughter there was “something in her house.” The daughter related her mom describing a man sitting on her bedroom ceiling fan looking down on her at night when she was lying in bed. A man who just looked at her. She had also described seeing other men in the house, men of different sizes who had big teeth. This had been going on for about a year.

The patient had undergone bilateral cataract surgery in the past. The surgery went well, with good preservation of vision afterwards. She had no history of auditory hallucinations and no history of psychiatric disease. The daughter did describe her mom growing forgetful recently with a tendency to lose things, such as her purse. There was also a history of anxiety, misunderstanding things and becoming upset over trivialities. The family’s concerns about dementia led her primary care practitioner to start a medication to help her memory. She was on no medications likely to cause hallucinations.

I said, “Ms. Fuller, tell me about these things you are seeing. Tell me what it’s like.”

“Well,” she said. “It’s men. With old big teeth. And, uh, when I go to bed, I got a fan up there. And this one, he comes every night and he just sits up there on top of that fan. Do you believe that? Do you believe I see that? I see that. Do you believe it?”

“I believe whatever you tell me you see.”

“Well, this is what I’m seeing, but other people, they look at me and say, you know. You know what they say. But I tell them, I say, well I know what I’m seeing in my house. I’m seeing these things.”

“Is it just that one man?”

“Naw, it’s a whole lot of ’em. Like around my walls at night. And sometimes they’ll be messing around the window. I’ll go and maybe push my blinds together, but they push ’em back. I wake up at night. I go to bed at nine o’clock, thinking maybe I’ll sleep for an hour and then wake up and see what’s going on. I keep a flashlight and I lay it in the bed. It’s so hot in there. Some people say keep a lamp on in there, a light on in my room, but sometimes it’s too hot with that light on, so I cut it off. It’ll be cool when I’m there in the dark but then I don’t know what’s going on.”

“Tell me more about the men with big teeth,” I said. “Is it all men? Are there any women?”

“All I can see is men.”

“Do they all have big teeth?”

“Looks like it to me.”

“Do you see them during the day or just at night?”

“I see ’em in the daytime, too. Everywhere in my house.”

“Can you see any in here?” I asked.

“Oh, yeah, uh-huh, ya’ll can’t see it, but look,” she said, pointing. “There’s one right there. When I ask somebody, they say they can’t see it. But I see those things. Is it my eyes?”

“Do they ever talk to you? Do they ever make any noise?”

“I can see ’em working their mouths but no sound comes out. What they say, I don’t know. They smile sometimes. They mostly just sit there and watch me. I get sick of ’em.”

“Are you afraid of them?”

“I try not to be. I ask the Lord to help me be strong.”

“Do you think they’re going to hurt you?”

“Oh, yeah.”

“What do you think they’re going to do?”

“What do you think men do?” she asked. “What do you think some men do when they come around? What do you think they want? What do men want? I’m gonna tell it like it is.”

“You think they’re trying to get in the bed with you?”

“Oh, yeah, I know that’s what they’re trying to do. Sometimes they bump my mattress.”

The patient was carrying a heavy, varnished stick, about four feet long, more hiking pole than walking stick, resting it against her knee throughout the visit.

“Do you use that stick to keep them away?”

“I’ve got two or three of these. I always keep two or three around my bed. They try to hurt me and I’m gonna hurt them. Between my stick and my flashlight, they better watch out. I’m not crazy now. You know I’m not crazy.”

“Yes, Ma’am, I know you’re not crazy.”

The patient’s neurologic examination was normal. Her visual acuity was close to normal with her glasses and there were no visual field abnormalities. There was no evidence of Parkinson disease, but her mental status examination showed she had dementia. MRI and EEG were normal, and the results of all her lab tests were normal.

What would cause an elderly woman to suddenly develop isolated visual hallucinations? She was reminiscent of a patient I had seen several years before, also an older lady, but not as frail and not demented. In that case, the patient saw a squad of Lilliputian men who would come

into her house through the windows at dusk. They would climb up onto the dresser and chest-of-drawers in her bedroom and just sit there watching her. At dawn, they would leave.

The most common cause of visual hallucinations is psychosis, and the most common cause of psychosis is schizophrenia. But schizophrenia is a disease seen in the young, and in the usual types of psychosis the most prominent hallucinations are auditory—the patient hears voices. Visual hallucinations in psychosis are usually much less prominent than the auditory ones, and isolated visual hallucinations are rare.

Patients in alcohol withdrawal often have visual hallucinations—the infamous pink elephants. The effects of other substances, particularly those taken recreationally, may include visual hallucinations. Indeed, that is frequently the point. In modern societies, hallucinations occur with such drugs as amphetamines, cocaine, LSD and Ecstasy.

The hallucinations are not an unwelcome side effect of the mind-altering properties of these illicit agents. I once saw a patient who loved to smoke marijuana and huff leaded gasoline so that he could hallucinate in color. Unleaded gasoline didn't work. With substance abuse, hallucinations can occur during acute intoxication or as part of withdrawal, and the hallucinations may occur on their own or as part of a drug-induced psychosis.

Always innovating, drug seekers discovered the hallucinogenic properties of the active ingredient, bufotenine, in certain glands of the Colorado River toad. Bufotenine is a controlled substance, but owning the toad is not illegal and by pressing on parts of the toad and then licking it, a determined, committed individual can get high and hallucinate. Such people are called toad lickers. The psychedelic properties of bufotenine come from a naturally occurring hallucinogen akin to LSD.

Long before LSD and Ecstasy, men devoted a lot of time and energy, and took risks, in pursuit of hallucinogens and other mind-altering agents. Some of their discoveries live on. Hallucinogens, primarily plant alkaloids, were often worshipped or held in an exalted status in Indigenous cultures, particularly in the Americas and specifically the Amazon, where there was a rich diversity of flora to exploit. The visual hallucinations these substances could induce were frequently rich, kaleidoscopic colors that seemed unreal and otherworldly to early man. The hallucinogens were smoked, swallowed, snuffed, drunk or applied topically. Some of the compounds used include peyote, mescaline, belladonna, ayahuasca, henbane and cannabis. In Siberia, it was found that the active ingredient of a psychoactive mushroom appeared in the urine unaltered. The psychoactive urine of intoxicated individuals was then consumed by others—an early form of recycling.

Certain hallucinogens commonly used in Old Europe, particularly belladonna, henbane, mandrake and jimsonweed are topically absorbed. Some of the rituals and ceremonies associated with witches are attributable to the use of these topical hallucinogens. Medieval witches would rub their bodies with hallucinogenic salves and ointments. The vaginal mucosa was particularly efficient at absorbing the compounds, and the original witch's broomstick was in fact an applicator for hallucinogens. They were tripping, but not across the sky.

Psilocybin and psilocin are the active compounds in hallucinogenic mushrooms, so-called magic mushrooms or just "shrooms." Magic mushrooms are psychedelic fungi that have been used for thousands of years, perhaps even by prehistoric hominids—McKenna's "stoned ape" theory of evolution. The effects are similar to LSD. Visually, colors brighten, halos and auras

appear around objects, things appear larger or smaller than they are, objects shrink or stretch, and colorful patterns may appear. Synesthesia can occur: the user sees sounds and hears colors.

The experience a particular individual has on ingesting a hallucinogen depends on their expectations and the environment, both physical and social, in which the drug is taken. Those who go into the rainforests of Oregon deliberating seeking to ingest hallucinogenic mushrooms usually have a pleasant experience; those who mistakenly ingest psilocybin while looking for edible mushrooms think they have consumed a poisonous variety.

There was no reason to suspect this nice, elderly lady might be abusing drugs, eating ‘shrooms or licking toads.

Visual hallucinations can occur as a side effect of many medications, or because of medication interactions. Simple hallucinations appear as unformed lights, colors or vague shapes, or often as halos or shimmering around objects. Complex hallucinations may appear as images of actual objects, people or animals. Responsible medications include antihypertensives, vasoconstrictors, vasodilators, erectile dysfunction medications, anti-parkinsonian agents, psychotropic medications and some antibiotics. Physicians have speculated for decades that the yellow halos van Gogh painted around objects, such as the stars in *The Starry Night* and the lamps in *The Night Cafe*, were due to the effects of digitalis intoxication.

Ms. Fuller was taking no medications likely to cause visual hallucinations.

Patients with severe brain dysfunction due to systemic illness may hallucinate, especially in the face of an underlying dementia. Once, while I was seeing a patient in the ER, the lady in the adjoining bed began doing excited color commentary on an imaginary baseball game across the room. She was in acute renal failure and about to start dialysis. Her brain, addled by the

circulating toxins her failing kidneys could not remove, was causing her to see men on base. As I recall, she was very upset that the guy on first was about to steal with a man on second. She was yelling at him to get back as the pitcher was spinning to throw a pick-off. She seemed to see all this vividly.

On another occasion, I was asked to see an elderly man with confusion. As is often the case, it was a patient with dementia who had developed an infection, in this case pneumonia. He was apparently hallucinating Jesus hovering over his bed, because he was looking toward the ceiling and calling out to whatever he was seeing. He was agitated and spoke fervently: “OK, Jesus, I see you, I see you. Hold on now. A little to the left. A little to the right. OK, Jesus, hold it right there, that’s close enough.”

So, if Ms. Fuller’s hallucinations were not the effect of some substance or medication, and there was no intercurrent medical illness, could it be her eyes, as she had asked? She did have that history of cataracts and cataract surgery, but her vision was now fine. Was that possibly related?

Sometime in the mid-eighteenth century, an 89-year-old Swiss man underwent bilateral cataract surgery. Although he did well initially, his vision soon deteriorated, and, as his sight failed, he began to experience complex visual hallucinations that included vivid figures of people and animals that he knew were not real. In 1760, his grandson, the Swiss scientist Charles Bonnet, described his grandfather’s visual hallucinations in an essay. In 1967, another Swiss scientist, Georges de Morsier, first used the term Charles Bonnet syndrome to refer to the visual hallucinations that sometimes accompany profound visual loss. Affected patients know the hallucinations are not real and do not suffer from any neurologic or psychiatric disorder. Charles

Bonnet syndrome is well documented in the neurologic, ophthalmologic, geriatric and psychiatric literature. There is some uncertainty about the prevalence, as many suffer in silence lest they be thought mentally unstable.

Patients with Charles Bonnet syndrome usually have visual field loss and/or moderate to severe bilateral visual impairment, typically related to cataracts, severe glaucoma or age-related macular degeneration. The most common underlying eye disease is macular degeneration.

The absence of visual impairment or visual field loss and the presence of dementia made it very unlikely that Ms. Fuller's hallucinations were due to Charles Bonnet syndrome.

Having considered and eliminated all the non-neurologic possibilities, I was now faced, as I suspected I would be from the beginning, with the neurologic possibilities.

One neurologic condition that could cause such dramatic, formed visual hallucinations is a disorder known as peduncular hallucinosis. A peduncle in neuroanatomy is a large, dense white matter bundle. The cerebral peduncles are two huge, paired pillars at the base of the brain, located where the thalamus, a brain structure, transitions into the midbrain, a brainstem structure. Jacques Jean Lhermitte (1877-1979) was a famous French neurologist, best known for describing Lhermitte's sign, an electric shock sensation radiating down the back brought on by bending the neck, most often seen in multiple sclerosis. In 1922, he described an elderly woman with evidence of midbrain disease who had bizarre visual hallucinations. The condition is occasionally referred to as Lhermitte's hallucinosis. Two years later, van Bogaert described a similar case and proposed the term peduncular hallucinosis, which is the name that stuck.

If peduncular hallucinosis had been the cause of Ms. Fuller's visual hallucinations, her MRI would have shown abnormalities in the region of the midbrain, thalamus or cerebral peduncles. But the study was normal, excluding that possibility.

Patients with migraine may have all manner of visual disturbances. A migraine aura, which usually precedes a headache, may cause flashing lights, wavy lines, shimmering, jagged edges, and blank spots in the visual field. But these are transient unformed visual disturbances, lasting typically 20 to 30 minutes and usually seen in young women. Migraine patients may occasionally report more unusual episodes that include formed hallucinations, the illusion of objects becoming too large (macropsia) or too small (micropsia) or too close or too far away. These episodes are referred to as Alice in Wonderland syndrome. This syndrome is associated most often with migraine, although there are other causes, and tends to occur in children, with many growing out of it by the teenage years. Some patients feel as if their body, or body parts, are growing or shrinking. Perceptions of accelerations and decelerations of time can also occur.

Authorities have debated for years whether Lewis Carroll had migraine and whether suffering these sorts of episodic migraine related visual distortions and misperceptions led to his writing of *Alice*. Others counter that he was just taking hallucinogens. The best judges would be those who have experienced this syndrome and also read Carroll's book. I cannot help but think most of them would come down on the side of the migraine hypothesis.

Patients with seizures, particularly those originating in the temporal lobe of the brain, often experience strange visual distortions, especially macropsias and micropsias, as an aura to the seizure or as part of the ictus. Formed hallucinations such as Ms. Fuller's would be unusual.

She had been experiencing hallucinations on a near constant basis for a year. These considerations made seizures as an explanation for the hallucinations most unlikely.

Visual hallucinations occur commonly in patients with Parkinson disease, but only in the context of long-standing disease and exposure to anti-parkinsonian drugs, especially dopamine agonists. Visual hallucinations are not a feature of early Parkinson disease. Ms. Fuller had no evidence of Parkinson disease on examination and had never taken any anti-parkinsonian medications.

So, I was rapidly coming face to face with the diagnosis that seemed most likely from the moment I saw “visual hallucinations” written on the referral form and noted her age. I knew then she would most likely turn out to have dementia with Lewy bodies (DLB).

Most people know about dementia and know about Alzheimer’s disease, but get a little fuzzy on whether or not they are the same thing. They are not. Dementia is a broad disease classification and refers generally to loss of cognitive abilities, but also to conditions that can begin as behavioral aberrations, visuospatial dysfunction and other kinds of impairment before cognition is affected.

After excluding treatable conditions such as an under-active thyroid gland or vitamin B12 deficiency, four diseases account for 90% of all cases of dementia: Alzheimer disease, the most common cause; vascular dementia, which results from multiple strokes and impaired blood supply to the brain; DLB; and frontotemporal dementia. The only one of these that causes early and prominent visual hallucinations is DLB.

A Lewy body is a round, reddish inclusion found in the cytoplasm of neurons, named for its discoverer, Dr. Friedrich Lewy. These inclusions are composed of a protein, alpha-synuclein,

and are seen most commonly in Parkinson disease. Lewy bodies in Parkinson disease are found primarily in the substantia nigra, a structure in the midbrain that contains a heavy concentration of dopamine-containing neurons. It is the loss of these dopamine-containing neurons that produces the motor manifestations of Parkinson disease. Lewy bodies can occur in other locations. They may be found in the cerebral cortex of some patients with Parkinson disease, but are seen in the cortex of all patients with DLB.

DLB is the second most common cause of dementia after Alzheimer disease. As with most neurodegenerative diseases, the etiology remains unknown. The clinical hallmarks are dementia, visual hallucinations and parkinsonism, but many other features can occur in some patients. The term parkinsonism refers to a combination of clinical signs and symptoms, including tremor, rigidity, paucity of movement, impaired balance and a host of other motor and nonmotor manifestations. Parkinson disease is the most common cause of parkinsonism, but it is far from the only cause. Another common cause is DLB.

A characteristic clinical feature of DLB is extreme sensitivity to neuroleptics—potent tranquilizers and anti-psychotics such as Thorazine and related drugs—which are the very drugs we might ordinarily give to a patient to control hallucinations. Neuroleptics in DLB patients may exacerbate any parkinsonian features and cause severe, even life-threatening, side effects at relatively low doses.

Parkinson disease is far more common than DLB. The typical patient with early Parkinson disease has primarily motor manifestations. The disease causes tremor and rigidity, often unilateral, and progresses slowly over years, responding well to treatment initially. Some patients with advanced Parkinson disease develop dementia, referred to as Parkinson disease

dementia (PDD). The typical patient with early Parkinson disease has prominent parkinsonian signs (tremor and rigidity) and no dementia. The typical patient with early DLB has dementia with no parkinsonism, or minimal parkinsonism not enough to warrant a diagnosis of Parkinson disease.

Ms. Fuller had no evidence of parkinsonism and there was nothing to suggest PDD as a consideration. Having excluded all other reasonable possibilities, a diagnosis of DLB appeared most likely.

The most famous victim of DLB was Robin Williams. He suffered with an array of symptoms: insomnia, constipation, gut discomfort, urinary difficulty, memory loss, anxiety, paranoia, loss of smell and an intermittent tremor in his left hand. He was thought to have Parkinson disease. His wife described the illness in an editorial titled, “The terrorist inside my husband's brain.”¹ As she put it, “He died from suicide in 2014, at the end of an intense, confusing, and relatively swift persecution at the hand of this disease's symptoms and pathology.” He kept saying, “I just want to reboot my brain.” Mr. Williams never complained of visual hallucinations, but his wife speculated that he probably had them but never talked about it.

Ted Turner, founder of CNN, revealed in 2018 that he too suffers from DLB. Fame and fortune offer no protection from neurodegenerative diseases—did not protect Ronald Reagan or Glen Campbell from Alzheimer disease, Michael J. Fox or Neil Diamond from Parkinson disease, Lou Gehrig from ALS or Dudley Moore from progressive supranuclear palsy.

There was not much to do. There were no pressing safety concerns and, given the potential sensitivity to neuroleptics, trying to control the hallucinations might pose more risk

than benefit. And Ms. Fuller seemed to be dealing with the situation—dealing with the little men with big teeth.

As long as she had her stick.

1. Williams SS. The terrorist inside my husband's brain. *Neurology*. 2016;87:1308-1311.