Rip Van Winkle Disease

As if under a spell, adolescents sleep for weeks, even months

The disease sounds as if it came straight out of Grimms' fairy tales. Teens fall into a slumber, dozing weeks or even months at a stretch. Marathon sleeping spells come and go, cropping up intermittently for roughly a decade. Then, symptoms vanish as mysteriously as they first appeared.

Called Kleine-Levin Syndrome, the condition is so rare that only a few cases have been reported in the world medical literature. Though the disorder is little known or understood, a flurry of recent research could change that. A few centuries ago, KLS might have been attributed to a witch's curse. Now the hunt for its cause focuses on genes and infectious agents.

Stephen Maier is among the unlucky few who know the groggy misery of KLS firsthand. He developed the disorder at age 13, after what seemed like a severe case of the flu. "My parents couldn't wake me up, and even when they succeeded, I was incoherent," says Maier. "So they took me to an emergency room."

Maier was subjected to a battery of tests, but they all came back negative. Two weeks later, he finally woke up and returned to normal life. But the bad dream wasn't behind him: Extended sleeping episodes of about 10 to 20 days' duration recurred about every six months throughout his teens. When they hit, he would spend about 22 hours a day in a haze of sleep. He slept through his basketball team's tournament and his senior prom. Like Rip Van Winkle, Maier emerged from these long spells in bed plagued by a sense of time warp. "Friends and family have to fill me in on what's been going on in the world," he says.

By his early twenties, the episodes gradually became less frequent, with his sleep time reduced to about 16 hours a day. During these relapses, though, even when he was awake, he felt like a zombie. "You could drag me to the doctor, but I could barely string together a sentence," Maier recalls. "Sometimes I'd speak in baby talk."

Now a 33-year-old accountant in San Jose, California, Maier has not had a recurrence in five years. He hopes that's the end of it – and he could be right. KLS typically comes on with a vengeance at puberty, gradually abates by the mid-twenties, and vanishes altogether by about 30.

"Owing to its rarity, few sleep specialists have seen a single case of the disorder," says Emmanuel Mignot, an authority on KLS at Stanford University. "For that reason there is skepticism about it in the medical profession, but KLS is without question a distinct disorder, with symptoms that are unique and very consistently manifested by those who fit the diagnosis."

Like Maier, sufferers often report that the onset of the illness coincides with a flu-like infection, leading many doctors initially to mistake the disorder for mononucleosis or viral encephalitis. At regular intervals after that the patient is afflicted by hypersomnia – a sleeping bout that typically lasts around 10 days. In one of the longest known episodes, one young woman slept nearly a year.

During these periods of hypersomnia, patients usually get up only to shovel food into their mouths, bathe, and take care of bodily functions. Even then, they're in a hazy, confused state. Light and sound may be irritating, and they have tremendous difficulty focusing. Reading or even holding a conversation is a challenge. A common complaint is that the world seems unreal or strangely off-kilter. "About the only thing I can do during episodes, apart from sleep, is watch mindless videos that I've seen a dozen times," says 17-year-old Eric Haller of Placentia, California, who has suffered bouts of KLS since age 12. "Anything new is too taxing on my brain."

Heredity may play a role. A questionnaire sent to 100 patients and their parents revealed that KLS disproportionately affects Ashkenazi Jews. Given that the disease is frequently preceded by fever and other flu-like symptoms, it seems that a combination of factors precipitates the illness. "My leading hypothesis," says Mignot, "is that KLS is caused by a virus or bacterium that some individuals are more genetically susceptible to."

Exploiting methods that he developed in his hunt for the gene responsible for narcolepsy, Mignot is now in hot pursuit of a KLS gene. He's identified several families with a strong hereditary pattern of the disease and has begun searching for common genetic markers. At the same time, the KLS Foundation is working with laboratories to collect blood samples from KLS patients and their families for a genetic study, and nasal swabs from patients during active episodes with the aim of isolating an infectious trigger. "We're very encouraged that we will soon have some answers to an illness that has heretofore been a complete mystery," says Neal Farber, father of a son and a daughter with KLS and co-president of the foundation.

For now, medicine has little to offer sufferers. "Stimulants can make KLS patients less sleepy, but they're still in this weird fog and can become agitated or aggressive," says Mignot. "That can be worse than simply sleeping." Other treatments that have been tried with marginal or no benefit are lithium, antidepressants, anticonvulsants, and mood stabilizers. "Many doctors just want to act," says Mignot, "but the best course of action may be to do nothing."

No magic pill or even a kiss from a handsome prince (or princess) will awaken sufferers from their long slumber. But in keeping with its fairy-tale symptoms, KLS does have a happy ending: Virtually everyone outgrows it.

Source: *Sleep Medicine,* the official journal of the World Association of Sleep Medicine and International Pediatric Sleep Association. July 2007.

Questions for Non-Fiction Article

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Directions: On a separate sheet of paper, answer the questions below. To receive credit, you must write complete, thoughtful sentences.

1. What symptoms might lead a doctor to suspect a patient has Kleine-Levin Syndrome?

2. Based on the material in the article, do you view Emmanuel Mignot as a credible source of information? Why or why not?

3. Emmanuel Mignot's work focuses primarily on understanding the causes of narcolepsy. What is narcolepsy? Feel free to use a dictionary to look up the word and write down its definition. How is KLS similar to narcolepsy?

4. Looking at the case of Stephen Maier, how could he be considered a success story?

5. What, do you suppose, are some of the consequences of missing out on significant chunks of time during one's teen years?

6. Think back over the past 30 days of your life. If you had been asleep for the last month, what important or valuable experiences would you have missed? List and explain at least three things.

7. What additional information do you want to know about KLS that is not included in this article?

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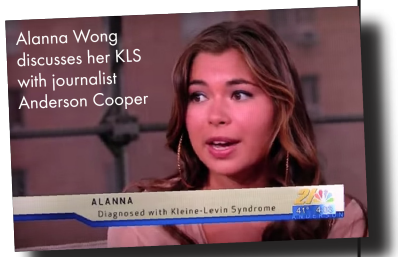

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