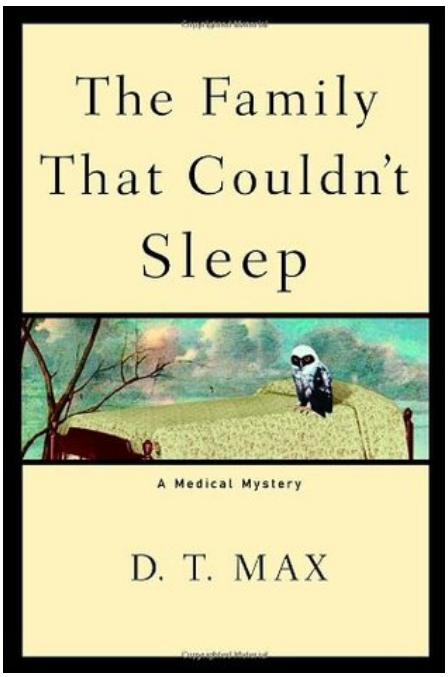

The Family That Couldn't Sleep: A Medical Mystery

D.T. Max

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For two hundred years a noble Venetian family has suffered from an inherited disease that strikes their members in middle age, stealing their sleep, eating holes in their brains, and ending their lives in a matter of months. In Papua New Guinea, a primitive tribe is nearly obliterated by a sickness whose chief symptom is uncontrollable laughter. Across Europe, millions of sheep rub their fleeces raw before collapsing. In England, cows attack their owners in the milking parlors, while in the American West, thousands of deer starve to death in fields full of grass.

What these strange conditions–including fatal familial insomnia, kuru, scrapie, and mad cow disease–share is their cause: prions. Prions are ordinary proteins that sometimes go wrong, resulting in neurological illnesses that are always fatal. Even more mysterious and frightening, prions are almost impossible to destroy because they are not alive and have no DNA–and the diseases they bring are now spreading around the world.

In *The Family That Couldn't Sleep*, essayist and journalist D. T. Max tells the spellbinding story of the prion's hidden past and deadly future

The Family That Couldn't Sleep: A Medical Mystery Details

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From Reader Review The Family That Couldn't Sleep: A Medical Mystery for online ebook

Richard says

?
Every time I donate blood (and I've donated well over ten gallons) I'm asked whether I've spent at least three months in the U.K. prior to 1996 (c.f.). This is because of what we all called "Mad Cow Disease" and what the medical folks now call *Variant Creutzfeld-Jacob Disease* (vCJD).

This book is poorly titled. Certainly, the horrible fate of an Italian family brings immediate human pathos to the story of prion diseases, but the more pressing story for many of us will be the atrocious practices in our food supply that permits—or even encourages—diseases like vCJD. But the real story lies elsewhere.

(It doesn't impress me that the Italian family is described as "noble", according to the blurbs. What on earth does that have to do with anything? Is the disease be more terrible because it is inflicted on "modern, cultured Italians, with stylish hair and eyewear"?)

There are several parallel stories told here. The chapters dealing with the Italian family's curse are interesting, as are those describing the investigation into *Kuru* ("a transmissible spongiform encephalopathy associated with the cannibalistic funeral practices of the Fore people of New Guinea"), although the chapters discussing the petty squabbles and large egos of the leading researchers in the field were less than riveting. But the heart of the book is the drama of how foolish cultures (British and American, mostly) willingly tolerate a socioeconomic system that eviscerates governmental regulation and industrial oversight, and how that played out in the Mad Cow disease crisis of the late 80s to mid-90s. (America mostly sitting smug on the sidelines, yet meanwhile tolerating incredibly disgusting practices in meat production, as well as incubating the *Chronic Wasting Disease* that is still spreading).

A European Union committee estimated that the English ate as many as 640 billion "doses" of Bovine Spongiform Encephalopathy during the mad-cow crisis. Luckily, it turns out the disease jumps the species barrier to humans very, very poorly—although there was a time when we didn't know that. British agricultural *regulators* are also responsible for the *promotion* of British agriculture (as so with the USDA), so they buried the story as long as they could. By the mid-90s, when the crisis reached its peak, one expert admitted that they could not "rule out 500,000 cases", and newspapers were wondering whether Great Britain would soon end up quarantined with half a million dying a year.

The problem? A fairly rare disease was spread through milk herds of Britain through the practice of protein supplements fed to milk cows. Among the sources of that protein are the carcasses of cows too diseased to permit into the human food supply. (Another source is chicken feces, which is still quite legal in the United States, by the way). Almost all diseases are eliminated through the rendering process, but "prions" are incredibly durable proteins.

But early on, British authorities were astonishingly stupid in their response to this disease. Americans had developed an antibody test that would almost instantly confirm the presence of a prion-based disease, but the British regulators choose to use a laborious and lengthy process of microscopic visual inspection rather than admit to trading partners that their food supply might have yet another problem. In fact, England's chief epidemiologist "wasn't a believer in prions". This, despite the fact that by this time two different Nobel Prizes had been awarded (to Americans!) for the discovery of this new disease vector.

Britain—and the world—got lucky because BSE/vCJD isn't easily transmissible from cows to humans or between humans. Unlike scrapie in sheep, for instance, or perhaps Kuru (which might, perhaps, be infectious by merely handling the corpse of an infected person, instead of through cannibalism). The United States has inadvertently pushed scrapie into the wild population of deer, elk, etc., in the American wild, where it spreads with apparent ease.

After finishing this book, it is a relief to ponder that all-in-all the number of people that die annually due to prion diseases is probably barely one hundred, and so this is less of a realistic threat to the reader than, say, bee stings or choking on a chicken bone. But prions are the worst instances of proteins-gone-bad, and misfolding proteins are also suspect in many other diseases (e.g., Huntington's and Alzheimer's), and so we'll probably be hearing more about prion-like diseases in the future.

This was an engaging and well-written book; I highly recommend it for anyone that enjoys science and medical non-fiction.

Petra Eggs says

This stunning book is about a very rare inherited neurological disease which strikes in middle age and one of whose main symptoms is the inability to sleep which quite quickly leads to death. The book is written in a very readable way and follows one family, the main family who suffer frm this terrible disease.

Cindy says

Prion diseases are freaky! That little bits of proteins could mis-fold, and that topological change could decimate a brain is just bizarre. One of the facts I was most surprised by is that prion diseases have three methods of infection: genetic, direct contact (i.e. eating or touching infected tissue), and spontaneous (i.e. a protein accidentally misfolds in the body). No other disease vector can spread via all three methods like prions. They are freaky disease superstars!

The Italian family in the title is beset with FFI, fatal familial insomnia, an inherited/genetic prion disease. It's sufferers tend to develop symptoms in middle-age (usually), and die fairly quickly. It's grim: their pupils turn into pin-pricks, they start sweating profusely, and they become unable to achieve any type of restful sleep. Eventually they lose all control, go into mad fits, finally fall into a coma and die. All the while their mind is intact.

The family's biography is only a frame for the rest of the book. In order to explain FFI and how difficult it was to diagnose as a prion disease, you have to understand the history of prion diseases, and the history of the field. Max delves into scrapie, kuru, GSS, CJD, BSE, and other known prion diseases. In some ways, the story of the researchers trying to pin-down this new class of illness was more fascinating than the family that couldn't sleep. There are huge egos, government cover-ups, and other non-science dramas that affect the lives of many people.

I really, really enjoyed this book - Max sets up the story in an extremely engaging way. It reads like a

medical thriller - like something out of that TV show *Mystery Diagnosis*, but on steroids. There are twists and turns to the diagnosis, and a whole lotta shock factor.

And yet I had to dock it a star. I thought there were two questions not just left unanswered, but totally unaddressed. 1) How does a genetic version of a prion disease, like FFI, not cause symptoms until middle-age? Is the disease building up slowly over time, or does something later set it off? 2) How does a simple mis-folding of a protein lead to a swiss-cheese brain? How do you connect the dots from misfolding to erosion of brain tissue and development of plaques.

There's a good chance that there aren't satisfactory answers to either of those questions, but I was hoping Max would at least acknowledge or address them. He certainly didn't shy away from other more technical discussions.

I had just finished the chapters on Mad Cow/BSE/CJD and Max goes into detail about the state of affairs today. Spoiler: it's not good at all, particularly the government's reluctance in the US. British beef is safer than US beef. Scary. My husband and I had already planned on eating beef for dinner - we had some leftover steaks that needed finishing. It certainly gave me pause. I don't eat a whole lot of beef as it is, but I might try to cut back a little more. Prions are just that freaky.

Trena says

This book ranks with A Short History of Nearly Everything and Animals in Translation as one of the best pieces of science writing I have ever read and I highly recommend it to everyone.

The book covers all aspects of prion diseases, the most famous of which is Mad Cow Disease (aka Bovine Spongiform Encepholopathy). I have a sort of superiority complex fascination/horror with BSE; as a vegetarian for the past 17 years I feel relatively safe from it. Prions are especially terrifying foodborne illnesses because there is no such thing as safe handling and nothing kills them--heat has absolutely no effect on them, for instance, nor do chemicals such as bleach.

But BSE is only one of the prion diseases, and the book covers the rest of the known family, such as scrapie in sheep, chronic wasting disease in deer (don't eat wild-caught venison in Wisconsin people, for real), and all the human varieties such as Cruetzfeldt-Jakob, kuru, and Fatal Familial Insomnia. The latter (FFI) is a large part of the book as the "medical mystery" refers not only to prions in general but also to a horrible, terminal disease that has afflicted a family in Italy for many generations and has only recently been identified as a prion disease. It is arranged in roughly chronological order and follows the scientific community as it seeks to uncover the mystery, but goes as far back in the humanoid medico-anthropological record as 500,000 BCE in a particularly interesting episode.

The writing is just amazing. Everything is put with crystal clarity without feeling dumbed down or watered down. As with Bryson's Short History, you feel really smart while you're reading the book because you get it so effortlessly. Every paragraph is interesting. There is no chaff in this book, but it's not written too lean either, as there is plenty of detail. It is completely engrossing and difficult to put down when it's time to go to bed.

If you only plan to read one non-fiction book this year, this should be it. I am serious.

AdiTurbo says

Excellent non-fiction that reads like a thriller and gives you a good review of the history of prion research, the development and spreading of prion diseases in animals and humans and the scientific aspects of the discovery and treatment of prions. It's fascinating, super-scary and even moving. Daniel Max the writer is emotionally invested in the issue and it shows. The writing is far from dry, and is full of compassion towards the people who have suffered from these diseases and their relatives who have to live with the grief and the fear of developing the disease themselves. A great read.

Gina says

I'll try not to give 5-star ratings willy-nilly, but this was a pretty amazing family biography, spanning centuries' worth of generations and shedding light on a variety of subjects through the lens of this bizarre and incredibly rare genetic disease. Agriculture, attitudes toward disease, the relationship between science and doctors, the mysterious biological function of sleep, international politics, economic competition and, of course, personal profiles of the afflicted. The drama of the current generation, who have a 50/50 chance of one day suddenly losing the capacity for sleep, ties it all together, and gives you a significant reason to keep reading and caring. I learned so much! Also, I gained an appreciation for the medical significance of the anomoly. A successful and engaging blend of family biography and the wide-reaching social history.

Note: seems like every other word in this book is "sleep," so read it when you're not too tired, or you won't make it very far. Furthermore, you will feel like a jerk when this family's exhaustion somehow functions as your bedtime story/lullaby.

Kate says

I learned a lot about prion diseases from this book, but it suffers from some major issues:

1. It is poorly organized. The chapters alternate between telling the story of the "family that couldn't sleep"--an Italian family suffering from Fatal Familial Insomnia, or FFI--and covering the history of prion diseases & research. That would be fine on its own if there was still some kind of timeline holding everything together, but there isn't: one chapter will discuss prion research from 1970-2004, and then the next chapter will return to research in 1980 (and then to a disease not mentioned for 5 chapters), and so forth. It was impossible to get my bearings in this book. Furthermore, at no point does DT Max set aside time to go through a simple explanation of prions, prion diseases, and some of their basic differences: all these facts appear willy-nilly, buried in the text, without a glossary for assistance. Some terms aren't made clear until near the end.

Also, the Italian family with FFI is discussed at length, and my copy of the book does not have a family tree. With this many relatives and a heritable disease involved, a family tree is a necessity.

2. Lack of scientific rigor. The last few chapters inexplicably step away from science and turn to online message boards (no, REALLY, message boards) and amateur speculations. The book is subtitled "A Medical

Mystery," not "A Medical Mystery plus stuff I read from these people online oh and I wonder about vaccines sometimes."

3. One of the key researchers of prion disease--and a Nobel winner, to boot--is a pedophile. An unrepentant one, at that. Max, to his credit, does not sweep this under the rug, but his attempts to be serious-but-not-TOO-serious fall flat. It's gross.

In sum: I did, as I noted, learn from this book, but it has serious issues (WHITHER THE EDITOR? WHITHER?) and that prevents me from giving it any more than the "it was okay" two stars. If you're not highly science-literate AND very interested in the topic, just skip this one.

Anita Dalton says

The family that could not sleep is a family in Italy that suffers from a disease called Fatal Familial Insomnia. There are several other families in the world affected by the condition, so it is extremely rare. It is a condition that strikes family members generally in late middle age and causes them to begin to lose physical control of their bodies as they stop sleeping. They sweat, they develop a very distinct pinprick appearance to the pupils in their eyes, they stop sleeping, and in end stages, have virtually no control over their bodies. In many cases, those who suffered from it were assumed to be either crazy or chronic alcoholics, and there is not a thing that can be done to help them. So few people suffer from the condition, and a cure would be so expensive to find that there is little incentive for drug companies even to research the condition.

Read the rest of the review here.

Paquita Maria Sanchez says

Seriously terrifying.

Talulah Mankiller says

Max’s “hook” is the family history of an Italian clan that suffers from a rare prion disease called fatal familial insomnia: basically, it eats away part of your brain, burns out your adrenal gland, and eventually kills you because you can’t sleep. Several victims were actually observed by a world-renowned sleep clinic before their deaths, and even though the patients went into REM and everything, their sleep wasn’t…normal. REM is supposed to momentarily paralyze the sleeper, but the people with FFI were up, walking around, and occasionally bowing to the Queen of England (no, I didn’t make that last part up). Unsurprisingly, when the poor bastards finally “awakened,” they didn’t feel like they’d slept at all. Because they hadn’t.

Say it with me: THAT SHIT BLOWS.

Anyway, that’s the hook, and it’s a good one, but the truth is that Max actually wrote what is essentially a pop history of prion diseases in general: he covers mad cow, scrapie, and kuru, among others. Prions, in case you didn’t know (and I didn’t) are defective proteins that replicate themselves ad infinitum—introduce a prion into your system, and it’ll turn your good proteins bad. Which, in turn, will gunk up your brain, drive

you crazy, and then kill you. Fun!

Prions are actually a source of much contention and debate, because they go against our current understanding of infection: basically, we believe that in order for something to infect us, it has to be alive. Viruses technically aren't, but even they have RNA/DNA, so they pass on a technicality. Prions, though, are just protein. They aren't alive, not even on a technicality. There are still scientists who believe that the protein theory is wrong and that if we can just get the purification process right, we'll find out that prions are nothing more than teensy, weensy little viruses.

We live in hope.

ANYWAY. Not surprisingly, given the fact that we still don't agree on what even causes them, prion diseases are incurable; the ones that have reached epidemic levels, like scrapie and kuru, have gone away only because they burned through their original hosts and the practices that led to them were discontinued, so there are no fresh victims. This made The Family that Couldn't Sleep a downer, because Max goes through all the horrible, HORRIBLE symptoms these poor people have to deal with before their early deaths, and then…yeah, there's no cure. Not even the hint of a cure. Not even the whiff of a cure. And although there's now a test that can predict who's a carrier (that is, an eventual victim), and although most of the family has taken that test, everyone has refused to learn the results. Which I can totally understand, because that's not something I could live with knowing, either, but at the same time? The chances of passing this on are 50/50, which means that the younger members of the family who choose to have children will give it to at least some of their babies.*

BUMMER doesn't even begin to describe it.

The Family that Couldn't Sleep is fascinating, albeit depressing, but it is hugely, HUGELY problematic. To begin with, Kuru is an illness that devastated certain tribes in Papua New Guinea in the decades immediately following WWII. The people trying to cure it? Were either colonial Australians (they got the territory as a "gift" from the defeated Germans) or Americans. In fact, the chief American investigator on the scene was always going on and on about how great and unspoiled and untouched the "wild" places were and blah blah blah, racist bullshit. Max makes some efforts to show that these attitudes were BAD WRONG BAD WRONG, but usually he's just very quietly ironic, or he's silent on the subject altogether.

Horrifyingly enough, however, the racism isn't the worst part. Guys, I know that we disagree on a lot of things as a society (or societies). I know that we have conversations about what constitutes racism and sexual violence and sexism and whatnot, but I thought we were ALL AGREED that having sex with under-aged boys was just plain wrong. There's some debate about under-aged girls because we suck at being human beings, but we were ALL AGREED that LITTLE BOYS WERE OFF-LIMITS. And yet! One of the big early researchers in the field of prions was a man named Daniel Carleton Gajdusek. Daniel Carleton Gajdusek? Was a self-admitted pedophile who was later convicted of molesting one of the multiple boys he "adopted"; oh, and he just loved spending time in Papua New Guinea studying Kuru, because some of the tribes there had cultural traditions of pedophilia, and that shit was RIGHT UP HIS ALLEY.

And Max doesn't condemn him. In fact, the way that he talks about Gajdusek's "sexuality" is just…ugh. It's very noncommittal, and at times he almost treats it like it's a joke. Hahahahaha! He touched a little boy's weiner! How amusing! Max treats Gajdusek as kind of a loveable, sad little joke, and frankly? The man was a predator. He may have done great things,** but he's still a goddamn predator. Be honest about that.

Recommended for: Anyone who really, really enjoys reading about diseases and can stand all the FAIL.

*Individual members of the family have taken steps to avoid passing the gene on; one woman waited to have children until her own mother was past the age where the disease was a possibility (it strikes by or before middle age). Once her mother had made it through the dangerous age without showing symptoms, the woman had children. Another woman, whose father had already died of the disease, chose to adopt; that was probably a good decision, because she later succumbed as well.

**And whether he accomplished very much at all is EXTREMELY debatable. Another thing I got from The Family that Couldn't Sleep? It's easier to get a Nobel Prize than you'd think. Gajdusek got one, and I still can't really figure out why. He wasn't even the one who established that what was killing kuru patients wasn't a virus; as far as I know, he just ran around collecting brains and bodily fluids and shooting up chimpanzees and waiting to see if they got sick. I don't get how that is so stupendous.

Lynne King says

I reread this review today (1st March 2014) that I wrote last year because a friend, of a friend of mine, has died from Prion's disease and has lost two siblings in the past year. How dreadful...I must reread this book.

* * * * * * * * *

I have a problem and it concerns books. If I see a title that sparks my curiosity, I must have it. I can normally keep this under control but then an enemy was unleashed in the form of my Kindle Paperwhite in February 2013. As a consequence, one click on Amazon and yet another book is added.

This book is a typical example of my "addiction" (which I hope is temporary), but the title cried out "buy me", "buy me" and that's what I did there and then. The three magical key words "sleep", "medical" and "mystery" gave the seal of approval. I could feel that sense of anticipation and I knew that this was going to be an excellent read, which it proved to be.

The author immediately whetted my appetite when I read that Stanley Prusiner received the Nobel Prize in Physiology or Medicine in October 1997 for his twenty-five years' of study on prions. This dreadful "infectious agent responsible for bovine spongiform encephalopathy, or mad cow disease, Creutzfeldt-Jakob disease, and a disease in sheep called scrapie."

The main reason for my intrigue was that prions are neither "a virus or a bacterium but a protein, a non-living thing" and also strangely enough, there's no DNA. Prions cannot even be "killed", even though they are not alive. A bizarre statement.

How could a non-living thing cause the havoc that it did in patients' brains and still continue to do so? Well I soon found out when D.T. Max skillfully sets the scene for this incredible book with the first of various medical mysteries, that of a family in Veneto who in 2001 were thinking about what Prusiner had said. This Italian family has been suffering from a very rare disease called "Fatal familial insomnia (FFI)", that is, not being able to sleep and finally dying from it. I initially thought that must be nonsense, because everyone sleeps for roughly a third of a day (that's part of our makeup). However, FFI symptoms are grim, normally arrive in family members over fifty, sweating is profuse, troubles with sleeping, and finally they cannot sleep at all. The brain is in constant overdrive and they fall into a state of exhaustion that resembles a coma. Regrettably it's downhill all the way until the eventual death, normally after about fifteen months.

This family could trace the disease back to the mid-eighteenth century to a Venetian doctor as a document was found describing his horrifying, but very similar symptoms, resulting in his death.

With further reading, I found out that when brains were examined after death, dreadful discoveries were made:

“The brains were badly damaged – full of holes, astrocytes, and plaques, piles of tangled dead protein.”

Comments from individuals like Pierluigi Gambetti, a director at the national prion surveillance center in Cleveland, one of the discoverers of the disease stated:

“I used to think that Alzheimer’s was the worst diseases you could get? But to see a loved one disintegrate in front of your eyes – and for that person to know it is happening? Somehow, the fact that it is so rare makes it even worse, it seems to me. I think now even a car accident would be less cruel.”

But to me, the most remarkable section of this book is when the author takes us to Papua New Guinea:

“Young man, be careful! I ate your grandfather” – Elderly Fore to his nurse at an Okapa hospital in 1947.

I had read about the practice of shrunken heads in Papua New Guinea, but it was rather unnerving to read that tribes out there were still cannibals in the 20th century.

Here, in the late 1950’s, Western doctors had found that the Fore tribe (an extremely primitive tribe; hunter-gatherers but also great fighters – they just fought everyone and for the most minor reasons) were suffering from a rather strange ailment called “kuru”. This was another case where a non-living molecule could reproduce itself and cause an infection as if it were actually alive.

How is that possible? It’s an absolute contradiction. But here again we have Carleton Gajdusek, another Nobel Prize winner (1976) for his work and what an interesting character he turns out to be; even spending time in jail in the States for his rather interesting “pleasures”. He liked young boys, “took an intense sexualized interest in the children he studied” but he said that this was only for medical research. The authorities in the States thought differently as they knew what a pedophile was.

Nevertheless, Carleton Gajdusek did excellent work in Papua New Guinea and he deserved to be a Nobel Prize winner. However, he still couldn’t “crack kuro on his own”. Help was needed from epidemiologists and anthropologists, and in fact it was the latter who found out what the problem was there. All I can say here is that cannibalism is connected but in all fairness to the Fore, “they never ate their own children or grandchildren, though: they saw that as incest which was taboo”.

I kept on wondering why the author had written this book and it transpires that he suffers from an inherited neurological illness. Not life-threatening such as FFI, but his muscles are slowly wasting away and he has to wear leg braces.

After finishing this book, all I could think about was what a fantastic social document it is; plus poignant, moving, amusing but also uplifting in that it’s an excellent medical mystery, so do try it! I’m sure that you won’t be disappointed.

Grumpus says

This is based upon the audio download from [www.audible.com:].

Narrated by: Grover Gardner

Like a smart consumer in the market for electronics, appliances or cars, I research my purchases by looking up recommendations on Consumer Reports. When I am looking for a good book to read, I turn to my trusted source for reviews—Goodreads. Based on member recommendations, I know going in that the book I choose will more than likely receive a higher than average rating from me. You guys have never let me down. . . well, until now. Don't worry, I won't hold it against you. :-)

The title of this book and the medical mystery it implied intrigued me and I was excited to read it. It wasn't long before I felt that the title of the book was a misnomer. The audio version on which I base this opinion, was just over eight hours long and I would be surprised if there was one hour of cumulative time on the discussion of the Italian family who "couldn't sleep". A more apt title would be something like, Prion Disease: A History of Discovery in Animals and Humans. But then, who's going to read that?

The book dealt more with the study of prions—the smallest known infectious agent which is a naturally occurring protein molecule that lacks nucleic acid. It is these prions that are at the root of this family's illness called Fatal Family Insomnia (FFI). It starts with sweating and constricted pupils the size of pin points and ultimately prevents family members with FFI from sleeping, leading to death. Members of this family have a 50/50 chance of passing this hereditary illness on to their offspring.

The majority of the book discusses the history and pathology of such prion-related diseases such as mad cow disease in cows, scrapies in sheep, kuru in humans (cannibal-related), Creutzfeldt-Jakob syndrome and Alzheimer's and NOT the subject of the book as titled.

Everyone knows how horrible Alzheimer's is with the loss of mind that accompanies it. Just imagine how awful it would be to have Alzheimer's but you still know what is happening to you. That is what it is like for this family. FFI is horrendous and rare (only 40 families in the world have it) and it is this rarity that prevents the needed money being allocated to it for researching its cure.

It's a sad family story, but again, and disappointingly so, the "family" is minor player in the saga. It was an okay book if you're looking for a medical mystery but it was not what I had signed on for. The author had me at the title but I still felt mislead on this one from the beginning.

Rose says

I enjoyed this book, but there's one part of it I don't understand. If you can point out what I'm missing please do so - it's been a while since I studied genetics.

The book says that the gene encoding the "prion gene" involved in CJD/kuru/FFI has two alleles. One codes for a methionine at a particular site and the other codes for a valine in the same position. If you have two

copies of the same allele, one maternal and one paternal, you are homozygous. If you have one of each kind, you are heterozygous.

Apparently, "homozygotes were at higher risk of prion disease than heterozygotes [this appears to go for both val/val and met/met] and there were more heterozygotes in the British population than chance would dictate." This lead the researchers involved to suggest that "some evolutionary force might have favoured heterozygotes over homozygotes".

Backing up this suggestion is the fact that "heterozygotes were overrepresented worldwide, in every race and every ethnicity". (Though Max also states at a different point in the book that "Japan is terrified of BSE because its population's genetic makeup is heavily homozygous," which doesn't seem to agree with the previous statement.) This lead to the conclusion that "their common ancestors must have faced a situation that winnowed out the homozygotes".

If both parents are heterozygous (both val/met), according to classical genetics, 25% of the offspring will be val/val, 25% met/met and 50% val/met - since it appears that both variants of heterozygosity (val/val and met/met) result in vulnerability, that means half of the offspring will be homozygous and therefore vulnerable, and half heterozygous - exactly equal proportions.

So how can you winnow out the homozygotes and leave your population full of heterozygotes? Whenever you have two heterozygous parents, half of the offspring will be homozygous. You might see a pattern in which homozygotes for a particular allele are underrepresented in a population because they have an increased risk of foetal nonviability, but there was no suggestion in the book that this might apply in the case of the prion gene. It seems like the homozygotes are generally healthy unless they are exposed to a prion disease or also inherit a prion-creating mutation, as in FFI.

Like I said, it's been several years since I studied this, so maybe I'm off base completely. I'd appreciate any comments that could clear things up.

Also, Max states that "In World War I, Italy had entered the wrong war on the wrong side at the wrong time." OK, so Italy didn't do well out of the Great War...but it was still on the winning Allied side, right?

Eve says

Prions. Before reading The Family That Couldn't Sleep, I had no idea what those were. Since finishing this book, I've developed an equal sense of respect and fear of them. "Prions are ordinary proteins that sometimes go wrong, resulting in neurological illnesses that are always fatal. Even more mysterious and frightening, prions are almost impossible to destroy because they are not alive and have no DNA.?" How's that for a mouthful?

At the center of this book is a Venetian family with a deadly legacy of Fatal Familial Insomnia dating back to the 1700s. FFI is a disease that strikes its victims in middle age, and causes complete insomnia, exhaustion, and eventual death within a matter of months. Max, himself a victim of a degenerative neurological disorder, expounds on the history of prions, theories on their origins, and the culminative affects on peoples and lands throughout the world. Cast your mind back to the Mad Cow Disease scare in Europe, or even the first cases of scrapie among sheep in Europe in the 18th century; these can be linked back to very bad little prions.

I really enjoyed the break down of scientific terms and I especially loved the history part. I find that I almost always enjoy the style and flow of books that are written by journalists, which is probably why it put me in mind of Brain on Fire by Susannah Cahalan and Lost in Shangri-la by Mitchell Zuckoff. A great read whether you're scientifically inclined, or just along for the adventure ride! Another plus: I now kinda understand the scientific references Amy Farrah Fowler, a fictional neurobiologist on the show The Big Bang Theory, periodically makes to her research work. Winning!

Lsexton says

You, too, will have trouble sleeping after you read this book.

This is a true-medicine thriller that leads the reader through the investigation of prion diseases, with surprising revelations along the way. The mysteries include a wasting brain disease that plagues an Italian family for generations, an epidemic of a similar disease called "kuru" found in a remote tribe in New Guinea; and the origins of prion diseases in sheep and cattle.

The book also introduces us to the scientists who are researching prions. We laypeople would like to think the field of brain science is captained by the most rational scientists in the world; alas, it appears to be headed by egoists, charismatics and at least one pedophile.

And we hope the USDA is doing its utmost protecting us from mad cow disease, guided by science rather than the beef industry. Sorry, more bad news on that topic awaits you in this book.

My advice after reading this book: Put ... the hamburger ... down ...
