

Erin Watt
Book Review
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“The Family That Couldn’t Sleep”: Insight into the Prion Mind

A middle-aged man during a frigid morning has to change his shirt because of the excessive perspiration on the collar; his pupils have turned to pinpricks and upon looking in the mirror his head is cocked in an unfamiliar way. The next few nights, the man cannot sleep and notices his pulse has shot sky high. In the following months this man is beyond exhausted and eventually is unable to walk. Sadly, the victim is aware of his body deteriorating but physically unable to halt the tragic developments. Approximately 15 months later, the man enters a comatose state and dies. The latter case study is of an Italian man suffering from a neurodegenerative disease, adversely this case is hereditary and relatives of the victim have a 50% chance of dying under similar circumstances. Prior to the late 1990s many physicians, family members and friends of the victim and future victims would be perplexed by the cause of these horrific, sudden deaths. However, in 1997 Stanley Prusiner, professor at the University of California-San Francisco was awarded the Nobel Prize for founding Fatal Familial Insomnia (FFI). This disease not only provoked the above tragic cases but also D. T. Max to author a book called, “The Family That Couldn’t Sleep” describing various prion diseases.

Max started his writing career being a book publisher in the early 1990’s and went onto become a writer and essayist for *The New Yorker*, *Los Angeles Times*, *The Wall Street Journal*, *San Francisco Chronicle* and *Chicago Tribune*. Aside from his passion for writing, he too like the victims of FFI, suffers from a neuromuscular disease caused by an unknown mutation. His syndrome is similar to spinal muscular atrophy (SMA) and Charcot-Marie-Tooth (CMT). The disease is formed from a mutation on a gene which negatively changes the structure or quantity

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of the proteins necessary for his nerves to successfully send electric impulses to the appropriate muscles. Therefore, he is unable to walk or balance without using braces. Due to Max's personal struggle, the reader has the impression that he was in part motivated to research neurodegenerative diseases and write this book.

In the beginning of his research on FFI, Max has the opportunity to attend the long, anticipated reunion of the Italian family. During this event the Italians discuss the different strategies needed to capture the media's attention, in order to increase the chances of finding a cure. The family members have different viewpoints regarding media attention; many of them afraid of the town people harassing or calling them freaks due to their family curse. According to Max, "Their neighbors, aware that many members of the family had died strange deaths before reaching sixty, began to think twice about marrying into the family." However, eventually the family gives Max permission to write a book of their family disease in hopes that some day it strikes the interest of a neurologist itching to find a cure. Throughout the book Max gives the reader countless anecdotes, some centuries old, describing many of the Italian family members suffering with FFI, all of them doomed with the same outcome death. This insight into the family lineage intrigues the reader's curiosity and commits their urge to learn more about this sickly, mysterious disease.

In my opinion Max conveys the process of prion formation in an exceptional, concise way allowing the general, non-scientific audience to understand the concept. Prions are made up of ordinary proteins from ordinary genes; however, prions strangely become tangled causing them to behave like a virus or bacteria. Normal proteins manufacture as ribbon, folding into a 3-

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dimensional shape, but prion proteins have two different shapes, one normal and the second being infectious. The initial abnormal protein begins the process of “conformational influence” meaning the way it is folded affects the bond with neighboring healthy proteins causing them to become malignant. This chain reaction causes cell deterioration, leaving gaps in the brain tissue. According to a victim’s wife from Max’s book, “My husband seen his brain x-ray and said it looked like someone shot him with .22 shotgun.”

Aside from the Italian family anecdotes and the struggle to find a cure, Max also includes other prion diseases such as kuru, mad cow disease and scrapie. Each chapter focuses on one of the prion diseases; this variety keeps the reader engaged and eager to continue turning the pages. Interestingly all of the prion diseases described by Max have similar, if not, identical symptoms. Carleton Gajdusek, researcher and pediatrician, investigated kuru, a brain disease, in Papua New Guinea. In a matter of 3-6 months kuru caused neurological degeneration and death. After many genetic tests, Gajdusek postulated Kuru was spread through the Fore’s tribal rituals of cannibalism. Throughout this chapter Marx informs the reader of the different tests and procedures needed to conclude the cannibalism theory. I thought this detail was insightful; the average reader may not be aware of the rigorous efforts involved in postulating a theory.

Scrapie, a prion disease discovered in Europe originating in sheep, causes the animal to scratch its back and top of tail by “scraping” against objects until bleeding occurs. This need to itch for some sense of relief is due to alternations in the sheep’s brain caused by prions. In later stages of scrapie the sheep tend to lose their balance and fall over to their death. The epidemic

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finally ended upon the collapse of the European sheep breeding industry, no longer did an economic pay off exist.

Mad cow disease caused cows to “tremble, walk clumsily and fall down”, after experiencing these symptoms many were unable to stand and would fall down and die. Unfortunately, the government officials ignored these growing cow fatalities, despite health warnings from specialists. Therefore, many Europeans consumed unsafe, infected meat and human fatality began to occur. It was extremely interesting to draw parallels and contrasts between the different prion diseases.

Although, Max does an excellent job of creating awareness to the greater public regarding variations of prion disease, I do not like the amount of history included. Frankly, the extensive amount of background details regarding the various prion diseases bored me and seemed insignificant toward the aim of the book. I couldn't help but feel a little lost when Max traced every minute detail such as laboratory locations, irrelevant tangents regarding the victims and countless names of all the people involved. Being a reader who is not extremely familiar with prions, I was more interested in the molecular and behavioral causes and the anecdotes from victims. The history lessons were neither necessary nor meaningful to me personally; I understand it is important to accredit the researchers and people involved, however, I felt this could have been performed in a more abbreviated fashion.

Overall, I felt this book was a worthwhile, informative and interesting read yet a little too history orientated. Max captivated my attention in the first ten pages and continually kept me interested throughout the entire book with the variety of victims and places around the world

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afflicted with the different prion diseases. Max's style of writing did not require the reader to have an extensive background in the sciences nor did it bore an undergraduate with a neuroscience minor. I believe Max met the expectations of the Italian family, which was to inform the general public of this rare disease and use this information to motivate researchers worldwide in finding a cure. Personally I thought the best part of the book was the personal stories Max acquired from the victims of FFI; being a psychology major I enjoyed learning the behavioral impact the disease brought onto the victim. The worst part of the book was the elongated history lessons, mentioned previously in the review.

I would recommend this book to anyone who is interested in neurodegenerative diseases or medical mysteries. The reader's academic background, age and occupation are irrelevant. An audience of particular interest would be anyone who has done research in the field of neuroscience simply because the topic is neurodegenerative diseases. Max not only sheds light on this rare, tragic disease but also bewilders the reader, leaving him or her with an urgency to know more information regarding FFI. Hopefully in the future case studies of middle age people sweating excessively, with pinprick pupils and their head cocked a certain way will be merely history.