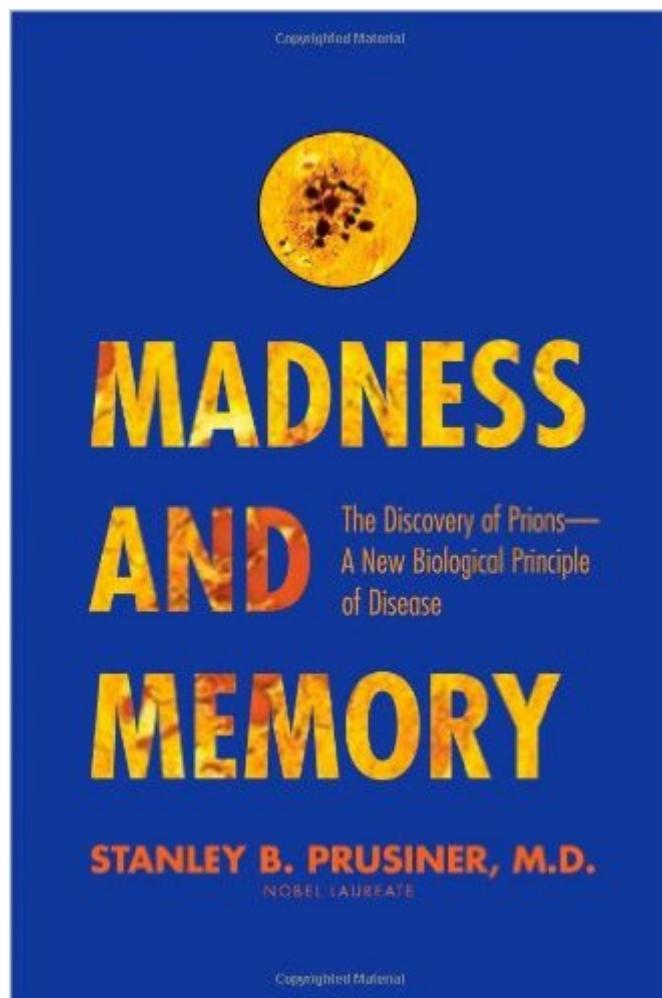


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# Madness And Memory: The Discovery Of Prions--A New Biological Principle Of Disease



## Synopsis

A first-person account of a revolutionary scientific discovery that is now helping to unravel the mysteries of brain diseases. In 1997, Stanley B. Prusiner received a Nobel Prize, the world's most prestigious award for achievement in physiology or medicine. That he was the sole recipient of the award for the year was entirely appropriate. His struggle to identify the agent responsible for ravaging the brains of animals suffering from scrapie and mad cow disease, and of humans with Creutzfeldt-Jakob disease, had been waged largely alone and in some cases in the face of strenuous disagreement. In this book, Prusiner tells the remarkable story of his discovery of prions—“infectious proteins that replicate and cause disease but surprisingly contain no genetic material”—and reveals how superb and meticulous science is actually practiced with talented teams of researchers who persevere. He recounts the frustrations and rewards of years of research and offers fascinating portraits of his peers as they raced to discover the causes of fatal brain diseases. Prusiner’s hypothesis, once considered heresy, now stands as accepted science and the basis for developing diagnoses and eventual cures. He closes with a meditation on the legacy of his discovery: What will it take to cure Alzheimer’s, Parkinson’s, Lou Gehrig’s and other devastating diseases of the brain?

## Book Information

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## Customer Reviews

The prion story changed our understanding of neurological disease, not only the infectious type , but also the common Alzheimer’s dementia. The key to this revolution was realization of the importance

of the folding of amino-acid chains for the correct function of proteins and how this could change in disease. Looking back, it seems curious how resistant the scientific community was to this idea. The fact that proteins had four levels of structure was basic: I learned this in medical school 50 years ago. But the dogma insisted that if something pathologically significant was wrong with a protein, it could only come from the gene, the DNA coding for the amino-acid sequence. And dogma is very hard to displace. Anyone advancing a radically new idea will not merely have their data and interpretations challenged: they will be subject to vilification and personal attack. For the lay reader, this book will give telling insight into the pettiness, and downright viciousness, of scientists competing in a high-profile area. Those of us who are practising scientists ourselves will by-and-large be aware of the sabotage and dirty dealing which is commonplace in scientific politics, so the book will not shock us. Instead, it will give an inspiring insight into how a person with dogged persistence can eventually succeed by sticking to his convictions and answering every criticism with solid data from rigorous experiments. The writing of the book has attracted comment from some reviewers. It is not up to the standard of literary fiction, but fully adequate to the task of conveying a tale about science. The level of detail is just right. The lay reader with only a smattering of biology will be able to follow, while those of us who are working biologists will be able to fill in the details for ourselves. All in all, an enthralling read on an important topic.

Prusiner, a great scientist, is rightly honored for the first and very difficult purification of prions (he coined the term) and with insisting and proving, against the tide, that they were infectious proteins with no discernible amount of genetic material, an idea that was prescient for its time. In this book he tells the story of the excruciating efforts it took to obtain enough material for study, including funding struggles to house the huge number of animals needed for the assays. The prion skeptics challenged his ideas for years (some have never given up), leading him to refine his experimental proofs. It's definitely worthwhile reading for this inside look at battles within science. For my taste, too much of the book was taken up with personalities and grudges. I would have preferred more on the science and mechanisms of prion replication (particularly the discovery and experimental manipulation of prions in yeast) and more of a look at the future of prion research. OK for the general reader, though the writing is a bit pedestrian and the experimental detail and departmental politics may deter some.

I chose this rating of five stars since it met my high expectations. I chose to order the book because I heard the author speak about his book, the trials and tribulations and his view that many chronic

neurological illnesses such as Parkinson's, which my husband had, could be linked to his field, that of prions. I liked that he spoke about his difficulties and successes which only came after decades and how scientists depend on their aides, money supply and his determination to see things through. I would recommend this book to anyone interested in the brain and its disorders, our unknown part of our body, Alzheimer, CJD, frontal temporal lobe and dementia.

Stan Prusiner is a prominent scientist working in the field of neurodegenerative disorders. He won the Nobel Prize in 1997 for his groundbreaking discovery of prions (misfolded proteins that cause diseases like mad cow disease). Much of the work on prions was done in late 1970's/early 1980's and it was an immensely controversial idea. Established dogma held that diseases are transmitted by DNA and/or RNA species (like bacteria or viruses). Prusiner's discoveries (aided by work done by many others in the field as well) rightfully shifted the paradigm but he faced an uphill task convincing the community. In this memoir, he describes the laborious task of isolating and identifying the agent that causes scrapie (mad cow's version in sheep). He also describes the hostility he faced both at his own institution (UCSF, where he still works) and at other places. Initially even he was in disbelief when he discovered that misfolded proteins can transmit disease without the aid of any genetic material, but as the evidence increased, he mounted a vigorous campaign to widely publish his work and convince others. A lot of science memoirs are sterilized i.e. the writer scrubs the past clean and all too often portray him/herself as a lone idealistic crusader interested only in unearthing the lofty truth. Prusiner's memoir is not one of those. Here you will find all the dirty laundry aired in its full glory. Every feud, petty or otherwise, gets a detailed treatment. Every person who slighted or supported him gets a mention. Prusiner doesn't hold back on his opinions and this book is refreshing for that. I gave this book three stars (instead of four) because I believe it flounders terribly towards the end. After spending pages and pages painstakingly outlining his research and displaying immense maturity in refusing to overstate his claims, Prusiner gets careless and starts speculating wildly about prions in other neurodegenerative diseases, like Alzheimer's. I work in the same field of research and there is this idea that has gained traction recently that a whole host of diseases like AD, Parkinson's etc could be triggered and developed in a person's brain through a 'prion-like' manner. There is limited evidence showing this might be the case, but a lot of other issues have to be worked out. For Prusiner to rashly call amyloid-beta peptide (the main component of plaques seen in Alzheimer's) a prion is gross overexaggeration and is misleading. His speculation mars an otherwise good book.

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