

BOOK REVIEW

Prion disease research: ethical aspects

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Max DT. *The family that couldn't sleep. A medical mystery.* New York: Random House; 2006 Hard cover. ISBN: 1-4000-6245-4. 301 pages. US\$25.95

"This is a book about prion diseases - what they are, what causes them, who they afflict, how we might cure them and how we found out what we know about them." This bland statement on page xxv of the introduction gives no clue to the unique manner in which Mr D T Max narrates an account that had me spellbound. Prions have fascinated researchers as they cause diseases in animals and man but cannot be shown to replicate like viruses or bacteria and cannot be destroyed by any of the means used to kill micro-organisms. (Stanley Prusiner – of whom more later – was searching for a name as catchy as "quark" for the factor responsible for these diseases and came up with the term 'prion'. Nature commented that it was "a rather tortured acronym for proteinaceous infectious particle.")

I needed to know the contents of the book as prion diseases cause catastrophic havoc within the brain and I study ailments of the nervous system. "Writing of her father, who had died of Creutzfeldt-Jakob disease (one of the human forms of prion disease), one woman posted on an e-mail list: 'My husband seen his brain X-ray and said it looked like someone shot him with a .22 shotgun.'" (She was referring the many holes created in the patient's brain by the disease.)

Even so, the width and depth of Mr Max's study, the fluency with which he weaves accounts of afflicted men and women with those of the purely technical and scientific aspects of prion diseases and of the scientists who have spent decades unravelling the mysteries of prions proved irresistible. There is also the poignancy of the fact that Mr Max, himself, suffers from a progressive muscle disease that may leave him crippled.

Readers of this journal will find sections of this book relevant. The factors that drive researchers, the pressures that lead to one-upmanship, the need to prove supremacy again and again, the manner in which economics triumphs over the welfare of humans, the pride and territorial imperatives that overrule reason and result in the wanton loss of lives are illustrated again and again by the author. The detailed accounts of the measures used by Carleton Gajdusek and Stanley Prusiner are especially interesting.

Let us consider some of the breaches in medical ethics discussed in the book.

As a young research fellow, Gajdusek had worked with Sir Macfarlane Burnet. In order to study child growth, development and disease patterns in primitive and isolated cultures, Gajdusek decided to work in Papua New Guinea in 1957. There he learnt about kuru - a disease that rendered its human victims progressively unsteady on their feet, cross-eyed, aggressive and emotionally unstable till their death. As he tried to gain access to victims of this disease, Gajdusek came across a file that showed that Burnet was long aware of the disease and even had a brain from a victim for study. He had, however, kept all this hidden from his own protégé, Gajdusek, despite knowing that the latter could use this information fruitfully. Learning of Burnet's deviousness, Gajdusek wrote in his diary: "Bitter? No? Just disappointed at my fallen idol... for my, how he falls!!!" When Burnet learnt that Gajdusek was tracking kuru and realised that a chance for a major discovery was slipping away from him, he even tried, unsuccessfully, to get Gajdusek to leave.

Discussing Prusiner's shortcomings, Max provides examples of his stinginess in acknowledging the source of his ideas. As his colleague at the National Institutes of Health in Bethesda, Maryland, put it, Prusiner believed in his dictum, "Repeat and expand (observations and statements made earlier by others) and the original will never be cited again. That's Stan's trick."

Max noted that protein and virology labs are full of Prusiner's former graduate students who, mysteriously, left the prion field once they left Prusiner's lab. Another colleague, Richard Johnson, provided the reason: "I've had people tell me that he told them they'd better not go into competition with him or he'd ruin them."

Prusiner was also caught red-handed misusing the peer review process. In the mid-1980s Prusiner was asked to judge a paper submitted by Paul Brown to the *New England Journal of Medicine (NEJM)*. Prusiner recommended its rejection and then submitted a similar paper based on his own lab work. The NEJM ultimately published Brown's paper and rejected that by Prusiner.

In closing, let me give you one of the many anecdotes that make the book interesting. Max introduces to us Tikvah Alper, born in South Africa and later a radiologist in Britain, who helped her husband develop an improved method for the analysis of bacteria. In a footnote, Max tells us that Alper refused a Royal sixtieth wedding anniversary greeting from the queen

because the queen issued it to her and her husband under his last name. Buckingham Palace relented and the greeting was re-sent to Dr Max Stern and Dr Tikvah Alper. Alper's comment? "I suppose the other way around would have been too much to hope for."

What about the title of the book? An Italian family – whose

members we are privileged to meet through the courtesy of the author – has several individuals across generations who were afflicted by a prion disease that destroyed large collections of nerve cells within the depths of their brains (in the thalamus and neighbouring structures) and made sleep progressively and frighteningly impossible till death afforded them relief.

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