



## The art of medicine

### Stigma, history, and Huntington's disease

Until my mother was diagnosed with Huntington's disease—a hereditary degenerative neurological and psychiatric disorder characterised by chorea, cognitive impairment, and emotional disturbance—she never mentioned that her father and three brothers had all died with this disease. Perhaps that missing family history was why I became a historian. I wanted to understand my mother's shame, and the origins of her devastating silence.

My first insight came at one of the public hearings organised in 1977 by the USA Congressional Commission for the Control of Huntington's Disease, which was headed by my sister Nancy Wexler. A woman from an affected family offered testimony that still resonates: "If I had one wish that this Commission could accomplish, it would be to take away the stigma of Huntington's disease and take it out of the closet". Another witness explained, "there is a shame associated with the disease, and they [family members] are so embarrassed by the whole thing that they just want to forget it". A third person recalled that "one of the recommendations our first neurologist made was that we all ought to be sterilized. That was the first thing he said after he announced the diagnosis."

Stigmatisation is often attributed to ignorance. Yet history suggests that while knowledge in the context of democratic values can help overcome prejudice, scientific and medical knowledge apart from such values can coexist with, or even contribute to, increased stigmatisation and rejection. For instance, after the rediscovery of Gregor Mendel's theory in 1900, the British geneticist William Bateson established definitively (in 1909) that Huntington's chorea, as it was called, was inherited as a Mendelian autosomal dominant disease. At the same time with the emergence of eugenics, Huntington's chorea became subject to demands for surveillance and control. A leading figure in legitimising such demands was the North American biologist Charles B Davenport who was director of the prestigious Biological Laboratory at Cold Spring Harbor, New York, USA, founder of the Eugenics Record Office, and an early proponent of Mendelism. In 1911 Davenport hired a physician, Elizabeth B Muncey, to undertake the first large-scale pedigree study of families with Huntington's disease in New York and New England, going back nearly 12 generations. He then used her data as the basis for a 1916 paper, in which he claimed that from just a few progenitors came a vast number of victims and called for immigration restrictions, surveillance of families, and compulsory sterilisation. Published in the *American Journal of Insanity*, "Huntington's Chorea in Relation to Heredity and Eugenics", was routinely cited in the biomedical literature on Huntington's as one of the foundational texts on the disease.

In an era when eugenics attracted scientists, physicians, and intellectuals of all political stripes, Davenport spoke with the authority of science. So too did a Connecticut psychiatrist named Percy R Vessie, whose 1932 paper became part of the Huntington's disease canon. Building on Muncey's pedigrees, Vessie traced the genealogy of one of his own patients back to her 17th-century New England immigrant ancestors, identifying three married couples from the English village of Bures in Suffolk as the most likely progenitors of Huntington's disease in the USA. For Vessie, witchcraft accusations against one woman and her relatives indicated the likely presence of the disease. The supposed "misconduct" of the men also pointed to the possibility of affliction, and indicated their disreputable character. Indeed, Vessie portrayed all these individuals more as villains than as victims. The men were "illiterate and arrogant, and none attained recognition or respectability". The women were the "lamentable" cause of introducing Huntington's to the USA. The "true story" of Huntington's disease in the USA, according to Vessie, was "revealed in the witchcraft trials of women in the Bures group". Because they showed no inclination to remain childless, it was imperative "to warn all such choreics and their children against propagation". Later he would call for "rigid sterilization".

Soon after it appeared in *The Journal of Nervous and Mental Disease*, Vessie's portrait gained traction in other prestigious medical and popular publications. In 1933, *The Lancet* abstracted Vessie's paper, boasting that "we [Britons] may congratulate ourselves on their loss, for...there can be no doubt that Wilkie, Nichols, and Jeffers [Vessie's pseudonyms for the three men] and their progeny were undesirable characters, and would nowadays be classified as belonging to the social problem group". *The Literary Digest*, a popular US magazine, in turn summarised *The Lancet's* version under the prejudicial title "The Witchcraft Disease". Upping the ante, the young British neurologist MacDonald Critchley claimed, in 1934, that all members of families affected by Huntington's disease were "liable to bear the marks of a grossly psychopathic taint, and the story of feeble-mindedness, insanity, suicide, criminality, alcoholism and drug addiction becomes unfolded over and over again". No wonder families like that of my mother tried to keep this illness secret, and were haunted by a sense of shame.

These unsavoury portraits of families with Huntington's appeared at a time of widespread compulsory sterilisations of institutionalised people with disabilities in such countries as the USA and Sweden, and especially in Germany where psychiatric patients and those with other disabilities were later subject to euthanasia. And despite postwar revulsion against Nazi eugenic policies, the distinguished American

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Alice and Nancy Wexler

Alice Wexler with her mother, Leonore, and her younger sister, Nancy, in 1947

geneticist, James V Neel, argued in the first (1949) issue of *The American Journal of Human Genetics*, that “even the most hesitant of us to venture into the field of negative eugenics would probably agree upon the desirability of the detection and exclusion of such immigrants on the same grounds that the United States Public Health Service now excludes those infected with certain contagious diseases”. As late as 1972, Critchley, who was by then President of the World Federation of Neurology, repeated Vessie’s claims that the earliest cases of Huntington’s disease in North America were identifiable “by reason of their sociopathic traits and their criminality”.

These narratives undoubtedly played a part in strengthening hostile perceptions of families with Huntington’s disease, within medicine as well as outside it. They helped legitimise the notion that certain classes of people were undesirable as citizens. They bolstered stereotypes of individuals with disabilities as aggressive, violent, criminal, and dangerous, while adding to the stigmas associated with alcoholism, mental illness, and cognitive impairment. They gave a rationale for doctors to endorse sterilisation or even celibacy for people at risk of Huntington’s, and encouraged medical indifference toward the care of those with the disease. As one neurologist testified at the 1977 Commission hearings, “It’s unbelievable how few doctors know about Huntington’s Chorea, and even worse than that” were those “who know about it, but will drop the patient like a hotcake. They don’t want to have anything to do with unpleasant, nasty diseases like that.”

Prejudicial representations of Huntington’s disease also encouraged flawed biomedical research, such as the 1951 study published in *Science* claiming (falsely) that those with the mutant Huntington’s gene had vastly more children

than their siblings without it. Only in the civil rights era of the 1960s and 1970s, with the repudiation of eugenics and the advent of the first lay associations of families affected by Huntington’s disease in North America and in Europe, did research priorities and representations begin to change. The 1977 Commission, with its extensive input from family members, was another critical step in reframing the way that knowledge about Huntington’s was produced.

As it happened, Vessie’s witchcraft theory turned out to rest on a case of mistaken identity, derived in part from Muncey’s flawed data, which no one had bothered to check. No one, that is, until Thomas H Gilmore and Mary B Hans showed, in 1969, that Vessie had confused Elinor Knapp, the immigrant ancestor of Huntington’s families in Connecticut who was never accused as a witch, with Goodwife Knapp, executed as a witch, but unrelated to families associated with the disease. The other accused women were also unrelated. In fact, I have found no credible evidence linking any of the well-documented New England women accused of witchcraft or their near descendants with behaviours resembling Huntington’s disease. Furthermore, my own research revealed that the “misconduct” of Vessie’s three men (who were not all from Bures) and their near descendants consisted mainly of minor infractions, such as profanity and drunkenness, or even political protest. It did not suggest Huntington’s or social pathology. Although one or more individuals among Vessie’s three couples could have brought the disease from England to New England, they were fairly typical settlers rather than Vessie’s “bad characters from Bures”.

Ironically, Vessie’s discredited witch thesis continues to circulate today while Hans and Gilmore’s critique is largely ignored. Moreover, when I tell members of families affected by Huntington’s disease that the witch story has no historical foundation, some are disappointed, as if it served as a metaphor for their feelings of being isolated and misunderstood. I have come to realise that this story may be historically inaccurate yet read from the perspective of the families, it seems to express a subjective, emotional truth.

In short, medical histories matter. Whatever the truths of the 17th century, it is clear that some 20th-century scientists and clinicians created historical narratives that deepened the stigmatisation of people with Huntington’s disease in their own time—and of other psychiatric and neurological conditions as well. Confronting the eugenic origins and harmful psychological and social legacies of these narratives cannot undo the past, but I believe it can help us change the conditions that perpetuate stigma and shame in the present.

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**Further reading**

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