

## EDITORIAL

### The history of Huntington's chorea<sup>1</sup>

Probably the first reference in medical literature to the condition known as Huntington's chorea or Huntington's disease appeared in Dunglison's *Practice of Medicine*, which was first published in 1842. The author's information derived from a letter from the Rev. C. O. Waters referring to a group of residents in the south-east corner of New York State who showed a rare hereditary disorder. The syndrome comprised the onset in adulthood of involuntary movements, unclear speech, advancing over the years to a state of dementia. In Dunglison's third edition (1848) reference was also made to similar cases in and around Philadelphia, recorded by Charles Gorman, and known locally as the 'magrums'. John Christian Lund reported the same disorder affecting members of two families in Saetersdalen, in southern Norway, in one instance involving four generations. The vernacular terms were the 'rykka' (or 'jerks') or else the 'arvesygen' (or 'hereditary disease').

Turning back the pages of medical history we find that in 1633 a certain Simon Huntington, a grocer from Norwich, decided to settle in the New England colonies, patriotically naming his new habitat Norwich, Connecticut. In 1777 Simon Huntington's great-great-great grandson Abel was born. After qualifying as a doctor he moved to East Hampton, Long Island. His son, George Lee, was born in 1811 and subsequently joined his father's practice. In 1850 his grandson George was born, qualifying at New York in 1872 and he, too, joined his father and grandfather in Long Island. He chose the topic of Sydenham's chorea for his M.D. thesis. The final paragraph called attention to the occurrence in East Hampton of a number of families who showed the identical syndrome of jerking movements and mental disorder. All three of the doctors were familiar with these cases and, indeed, the manuscript of the thesis contains marginal notes pencilled in by the father and grandfather. Obviously, they were ignorant of what had previously been published on this disorder. Locally, it was referred to with awe as 'that disorder' for it carried with it a stigma, being popularly attributed to a curse which had befallen a remote ancestor who had mocked the suffering of Christ upon the cross.

With the later discovery of other affected families elsewhere in the world, and particularly North America, considerable interest was raised in the genealogy of this disorder which by now had rightly received the eponymous designation of Huntington's chorea. Thus it was possible to determine that the Long Island cases were the offspring of a man named Mulfoot or Mulford, who had been born and brought up in the Massachusetts–Connecticut area. The Mulfoot ancestry was then laboriously traced back to three young men and their wives who, in 1630, had left the village of Bures St Mary on the border between Suffolk and Essex, to settle in the American colonies. The Lord of the Manor at Bures was the wealthy, puritanical Herbert Pelham. Finding himself increasingly dissatisfied with the High Church proclivities of the government he decided to emigrate. He offered to give passage to any villagers who wished to join him. In 1630 Pelham and the three families embarked in the James Winthrop Fleet, landing three months later in Salem, Mass. Pelham was a man of probity but his companions were very different. Even on the voyage they were in constant trouble on account of their reprehensible behaviour. Once established in their new home, they and their descendants proved to be delinquents. Intermarriage, illegitimacy and incest were common: the males were constantly in and out of goal. The fate of the female members was worse. No fewer than seven of them were arraigned, punished and even burnt at the stake by reason of witchcraft. Indeed, the notorious Groton witch was a member of this family.

A study of the transcripts of these early trials specifically refers to the wild, uninhibited grimacings

<sup>1</sup> Address for correspondence: Dr Macdonald Critchley, The National Hospital, Queen Square, London WC1N 3BG.

and contortions, thus indicating that the alleged demoniacal possession was, in reality, the expression of a neurological disorder.

Who were the original three families from Bures and what was the real reason for their escaping to America? We have every reason to suspect that the three males were blood-relations and possibly their wives too. A study of the Church records at Bures makes it clear that a certain Mary Haste was the parent of at least two of these men and almost certainly of all three.

It is now realized that the descendants of Mary Haste were responsible for the largest community of cases of Huntington's chorea in America. Some of these cases are known to have later moved so as to settle in the mid West, some indeed as far away as Oregon, California, and even Hawaii.

Some time ago a hardheaded economist estimated that the cost to the United States of each member of this clan has been at least 400 dollars a year, and deplored that the rigid rules for the entry of cattle did not apply to humans. After investigating 4600 descendants of this group, Davenport & Muncey (1916) regretted that there had been no embargo on entry, or even such prophylactic measures as compulsory sterilization. Had it been known, they said, that one parent of the three brothers who came in the seventeenth century from England was choreic and had been excluded on that account, 'the United States would have been spared 900 cases of one of the most dreadful diseases that man is liable to'. But, of course, not every member of a family which is at risk ever contracts the disease and, if the original family-group had been rejected, the community of the United States would also have been bereft of the services of two leading educators, a surgeon or two, a couple of State senators, two or three Congressmen, and several ministers of religion. *Cui bono fuerit?*

The Bures group, though the largest, is not the sole component of the Huntingtonian cases in New England. Another assemblage is made up of the Welles family. Whence and when did they appear upon the American scene? Apparently, their ancestry derives from one Nathaniel Welles, a respected and prosperous tradesman who migrated in that same sinister year – 1630. Wonder of wonders, he was a citizen of Colchester, a town located a mere ten miles from Bures – that is to say, within easy walking distance of Mary Haste.

Did Huntington's chorea exist among Mary Haste's forebears? It seems likely, though it cannot be proved. Essex and Suffolk were blackspots of alleged witchcraft in the sixteenth and seventeenth centuries. A most careful search of the lists of those accused of sorcery in East Anglia has uncovered two pairs of victims sharing names which appear in such far-removed areas as East Anglia and New England.

Similar, though smaller, clusters of cases of Huntington's chorea have come to light in many parts of the world. In Great Britain family-groups have been identified in Avoch, Ross-shire, Cornwall and Northamptonshire. The majority of the cases encountered in Australia are known to stem from a Miss Cundick, a Huguenot who left her native Somerset in 1848 to take up residence in Tasmania. She was twice widowed and had had children by both husbands. The same fate befell the descendants of both of the male progenitors. Eventually, some moved and settled in New South Wales.

In the Province of Ontario there are one or two sizeable groups of cases. There was a notorious mass-murderer in Toronto who had eleven close relatives with Huntington's chorea. Their ancestry was traced back to a migrant who left England early in the eighteenth century.

In 1685 a Huguenot family left Monbéliard on the French-Swiss border to take refuge in South Carolina. Later they went north to settle in Nova Scotia in the village of Tatamagouche. Many were victims of Huntington's chorea.

From the literature it appears that this is a disease which has been encountered in almost every country, although it would appear that there is a conspicuous immunity to this disease among members of the Jewish race.

Such are some of the highlights of the history of what has indeed been dubbed the true 'American tragedy'. By reason of its genetic qualities it is a disease which is naturally liable to increase in numbers rather than decrease. Within the last twenty years the medical interest in Huntington's chorea has grown considerably but the end is not yet in sight. Huntington's chorea is not a

compulsorily notifiable disorder. It does not respond to so-called genetic counselling, largely because there is still no measure – clinical, anthropomorphic, electroencephalographic, biochemical or pharmacological – that will identify those members among a family at risk who are destined to become victims.

MACDONALD CRITCHLEY

## REFERENCES

- Davenport, C. B. & Muncy, E. B. (1916). Huntington's chorea in relation to heredity and eugenics. *American Journal of Insanity* 73, 195–222.
- Dunglison, R. (1848). *The Practice of Medicine* (3rd edn), p. 218. Lee & Blanchard: Philadelphia.