ON CHOREA

By George Huntington, MD, Of Pomeroy, Ohio

Essay read before the Meigs and Mason Academy of Medicine at Middlepor, Ohio, February 14, 1872.

And now I wish to draw your attention more particularly to a form of the disease which exists, so far as I know, almost exclusively on the east end of Long Island. It is peculiar in itself and seems to obey certain fixed laws. In the first place, let me remark that chorea, as it is commonly known to the profession, is a description of which I have already given, and more especially when these manifestations have been of a serious nature, one or more of the offspring almost invariably suffer from the disease, if they live to adult age. But if any chance these children go through life without it, the thread is broken and the grandchildren and great-grandchildren of the original sufferers may rest assured that they are free from the disease. This you will perceive differs from the general laws of so-called hereditary diseases, as for instance in phthisis, or syphilis, when one generation may enjoy entire immunity from their dread savages, and yet in another you find them cropping out in all their hideousness. Unravel and whetstale as the disease may be in other respects, in this it is firm, it never skips a generation to again manifest itself in another, once having yielded its claim, it never again regains them in all the families, or nearly all in which the choreic taint exists, the nervous temperament greatly preponderates, and in my grandmother’s father’s and father’s experience, which conjoinly cover a period of 70 years, nervous excitement in a marked degree almost invariably attends upon every disease these people may suffer from, although they may not when in health be over nervous.

2. The tendency to insanity, I sometimes find that form of insanity which leads to suicide, is marked. I know of several instances of suicide from this form of chorea, or who will have an experience, and I have in my mind and body both gradually fail until death relieves them of their sufferings. At present I know of two married men, whose wives are living, and who are constantly talking love to some young lady, not seeming to be aware that there is any impropriety in it. They are suffering from chorea to such an extent that they can hardly walk, and would be thought, by a stranger, to be intoxicated. They are men of about 50 years of age, but never let an opportunity to flirt with a girl go past unimproved. The effect is ridiculous in the extreme.

3. Its third peculiarity is its coming on, at least as a grave disease, only in adult life. I do not know of a single case that has shown any marked signs of chorea before the age of thirty or forty years, while those who pass the fortieth year without symptoms of the disease, are seldom looked upon as an ordinary chorea might begin, by the irregular and spasmodic action of certain muscles, as of the face, arms, etc. These movements gradually increase, when muscles hitherto unaffected take on the spasmodic action, until every muscle in the body becomes affected (except the involuntary ones), and the poor patient presents a spectacle which is anything but pleasing to witness. I have never known a recovery or even a amelioration of symptoms in this form of chorea; when once it begins it clings to the bitter end. No treatment seems to be of any avail, and indeed nowadays its end is so well-known to the sufferer and his friends, that medical advice is seldom sought. It seems at least to be one of the incurables.

Dr. Wood, in his work on the practice of medicine, mentions the case of a man, in the Pennsylvania Hospital, suffering from chorea, which resisted all treatment. He finally left the hospital unscarred. I strongly suspect that this man belonged to one of the families in which chorea existed. I know nothing of its pathology. I have drawn your attention to this form of chorea, gentlemen, not that I considered it of any great practical importance to you, but merely as a medical curiosity, and as such it may have some interest.

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Huntington's Chorea

Advances in neurology have not been made only by the laboriously documented treatises of academics. George Huntington was a general practitioner from East Hampton, Long Island, who was 22 years old when he described hereditary chorea. The description was made possible by the fact that Huntington, his father, and his grandfather had practiced medicine in the same town and had observed several generations of afflicted families. Huntington's report is written in a vivid, personal style and is eminently readable. William Osler said of this paper: "In the history of medicine there are few instances in which a disease has been more accurately, more graphically, or more briefly described." The unpretentiousness of Huntington's article is matched by the modesty shown by later years when he referred to its acclaim as "unsought, unlooked for honor."

Despite its brevity, little has been added to Huntington's original description. The trait is attributable to an autosomal dominant gene with complete penetrance. Although Huntington stated that the onset is always in adulthood, rare cases have since been described in children. Variations of the classical clinical picture have been observed, and the rigid and akinetic forms are particularly noteworthy. Pathologic changes in the brain, consisting primarily of degeneration in the basal ganglia and the frontal cortex, were identified by Marie and Doyne. The social history of the disease has also been investigated, and it is evident that many persons burned as witches in England and America were victims of Huntington's chorea. The ancestry of Huntington's cases in East Hampton has been traced to "a young man of Bures [England], by the name of Jeffer, and not of chronic strain, who fell in love with the daughter of a choreatic and wished to marry her. His family stoutly objected, and so to obtain his end, he, in 1590, had to marry her and take her to America."

Huntington begins his article with a review of the well-known Sydenham's chorea. He concludes with the description that made him famous.

References