MD REMEMBERS: A Classic Contribution

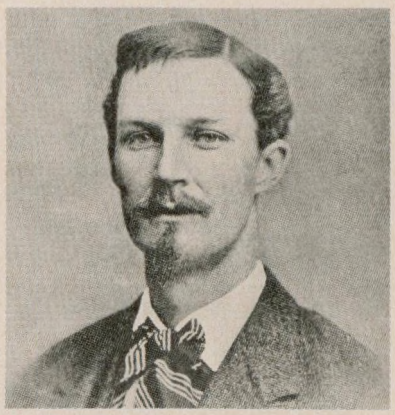
On February 15, 1872, Dr. George Huntington, a 21-year-old graduate of Columbia University's College of Physicians and Surgeons, reported to the Meigs and Mason Academy of Medicine in Middleport, Ohio, on a grave form of chorea with a tendency to insanity that afflicted only adults in a few families living in eastern Long Island. He also reported that the disorder never skipped a generation and if the victims' children escaped it "the thread is broken and the grandchildren and great-grandchildren of the original shakers may rest assured that they are free from the disease."

Dr. Huntington, whose physician father and grandfather had observed the disorder in certain East Hampton families over a period of 78 years, saw his first cases as a boy while riding on rounds with his father. He later recalled that he stared in wonderment, almost in fear, when they came upon a mother and daughter, "both tall, thin, almost cadaverous, both bowing, twisting, grimacing."

Soon after Dr. Huntington's paper was published° it attracted international attention and by the 1890s a number of neurologists were investigating "Huntington's chorea." The author subsequently noted that the disorder which he reported "as a matter of curiosity" had been mentioned previously by other physicians.

The first known reference to chronic, progressive hereditary chorea was in an 1841 letter from Dr. Charles Oscar Waters of Franklin, N. Y. to Dr. Robley Dunglison of Jefferson Medical College. Dr. Waters wrote to his former professor to ask if a hereditary convulsive disorder, commonly called "the magrums," could be a peculiar modification of chorea. Another Jefferson graduate, Dr. Charles Rollins Gorman of Lucerne County, Pa., mentioned that the disease prevailed in other parts of the country in 1848 and Dr. Irving Whitall Lyon of Bellevue Hospital reported three cases of chronic hereditary chorea in 1863. Dr. Lyon wrote that people among whom the disease occurred "have repeatedly been known to interdict marriage alliances between their children and those believed to be tainted with the migrim diathesis."

While Huntington was not the



first to refer to the disorder, Sir William Osler said of the young doctor's paper: "In the history of medicine there are few instances in which a disease has been more accurately, more graphically or more briefly described."

Demonic Disorder. Huntington's Disease, called the most demonic of all human afflictions, is caused by a dominant gene. In the early 1930s Dr. P. R. Vessie traced the disease through 12 generations to three women who came from choreic families in Suffolk, England, to Boston in 1630. Some investigators believe that a number of women accused of being witches in 17th century New England may have been victims of chorea. Cases have been recorded in many areas and geneticists at the University of Adelaide, Australia, reported a relatively high incidence in a community of aborigine descent. It is believed that the disease was introduced into the community by a European man a little over a year ago and in the present (fifth) generation there are 47 individuals who are known to have had a choreic parent. The investigators noted that "the prechoreic individual often shows an unstable temperament."

The incidence of the disease in the United States was thought to be 10,000 to 14,000 but the Committee to Combat Huntington's Disease has received letters from families indicating that the number of afflicted individuals is closer to 50,000. The committee founded by the widow of folk-singer Woody Guthrie who died of the disease in 1967 after a 13-year illness, now has 18 chapters in the United States. One of the projects that the organization is supporting is a complete bibliography on chorea that is being compiled by five scientists and will be crossindexed and microfilmed by the Library of Louvain in Belgium.

An official registry of patients has been established at Creedmoor Institute where Dr. John R. Whittier heads the research and treatment program. Dr. Whittier has reported that fluphenazine hydrochloride (tradenamed Proxilin by Squibb) is the drug of choice, that depression which is frequently severe in these patients responds to antidepressant medication and that pharmacologic support may begin as early as the latent phase of the disease if anxiety is a problem.

Drs. Giuseppe Scotti of Montreal and Hans Spinnler of Milan recently reported that amantadine administered to five patients had no effect on involuntary movements of three but slightly improved their mood and alertness. An improved gait and reduction of involuntary movements was observed in the fourth patient and in the fifth who also received haloperidol and reserpine.

In most cases symptoms of the disease appear in the third decade but two percent of the victims develop the disorder in childhood and five percent after the age of 60. Researchers are trying to develop a biochemical test to identify the carriers of the gene before chorea develops and the committee is hopeful that a specific therapeutic agent will be discovered.

Next month° in Columbus, Ohio the World Federation of Neurology Research Commission on HD, headed by Dr. André Barbeau, professor of neurology at the University of Montreal, is holding a centennial symposium on the disease.

° In *The Medical and Surgical Reporter* of April 13, 1872.

° March 26 to 29.