



This genetic brain disorder turned Woody Guthrie's life from songs to suffering

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From the wreckage of the 1935 "Great Dust Storm" — which decimated the Oklahoma panhandle, parts of Texas, Arizona and other farmlands in the Great Plains, and set in slow-motion a mass exodus to the more fertile soil of California — emerged some of America's major artists.

One was the Nobel Prize-winning novelist John Steinbeck, who was born in Salinas, California, in 1902, and whose brilliant portrayal of migration in "The Grapes of Wrath" continues to inform our understanding of poverty and desperation to this day.

Another artist, whose birthday we celebrate on July 14, was Woodrow Wilson Guthrie. Better known as Woody Guthrie, he was born in Okemah, Oklahoma, in 1912. The folk singer, activist and songwriter recorded album after album of beloved and socially important songs, including "This Land is Your Land," (which originally had each verse end with the line, "God Blessed America for You and Me," in response to the hyper-patriotic Irving Berlin tune, "God Bless America," belted out by Kate Smith).

Guthrie also wrote a wonderful series of songs he called "The Dust Bowl Ballads," which included "Do Re Mi," (a song about experiences of the Dust Bowl migrants, often known as "Okies," with the famous refrain, "Oh if you ain't got the do re mi, folks, you ain't got the do re mi..."), "Tom Joad" (inspired by the main character in Steinbeck's "The Grapes of Wrath"), and "Pretty Boy Floyd," (a song celebrating the famous gangster and some of his criminal exploits in Oklahoma).

Steinbeck and Guthrie knew each other well; Guthrie named one of his sons Joady, after the novelist's protagonist. In 1943, Guthrie even penned a partially fictionalized, "semi-autobiography" called "Bound for Glory," which many have compared in power to Steinbeck's "The Grapes of Wrath." (It was later made into a film in 1976 starring Keith Carradine.)

By almost everyone's measure, including his own, Guthrie suffered from active alcoholism for most of his adult life. But he was dealt an extra blow — inheriting Huntington's disease (HD), from his mother, Nora Belle Guthrie.

HD is primarily an autosomal dominant genetic disorder, meaning a child has a 50-percent chance of inheriting the dominant trait and, hence, the disease from the affected parent. In rare cases, HD may be due to a new mutation. Most Huntington's patients do not develop obvious symptoms until between the ages of 30 and 50. HD sets in motion the production of an abnormal protein that destroys brain cells, leading to serious mood disorders, followed by uncoordinated and involuntary body movements (known as chorea), balance problems, psychotic breaks, dementia and death, roughly 15 to 20 years after the diagnosis is first made.

This terrible malady was first described in a paper entitled "On Chorea," which appeared in the April 13, 1872, issue of the Philadelphia Medical and Surgical Reporter. It was written by George Huntington, an American physician who was practicing in Ohio at the time he presented his paper to the Meigs and Mason Academy of Medicine located in Middleport, Ohio. A brief quote from his remarkable essay is merited:

"The hereditary chorea, as I shall call it, is confined to certain and fortunately a few families, and has been transmitted to them, an heirloom from generations away back in the dim past. It is spoken of by those in whose veins the seeds of the disease are known to exist, with a kind of horror, and not at all alluded to except through dire necessity, when it is mentioned as "that disorder." It is attended generally by all the symptoms of common chorea, only in an aggravated degree, hardly ever manifesting itself until adult or middle life, and then coming on gradually but surely, increasing by degrees, and often occupying years in its development, until the hapless sufferer is but a quivering wreck of his former self....There are three marked peculiarities in this disease: 1) Its hereditary

nature. 2) A tendency to insanity and suicide. 3) Its manifesting itself as a grave disease only in adult life."

In 1908, the great Johns Hopkins physician William Osler applauded Huntington's paper with a glowing review: "In the history of medicine, there are few instances in which a disease has been more accurately, more graphically or more briefly described."

Chronic alcoholism, itself, can cause a host of neurologic and cognition problems and can add much fuel to the fire that is HD. As a result, when Guthrie was beginning to show classic symptoms of HD in the late 1940s, he and his doctors blamed his health issues on booze. Adding to the confusion was that fact that back in the mid-20th century HD was a rare and difficult-to-diagnose disease.

In 1952, Woody was committed to the Brooklyn State Hospital and his doctors told his wife Marjorie to divorce him and take custody of his children because of Woody's raging paranoia and occasional violent acts against family members. Despite the divorce, she remained close to Woody for the rest of his life and supervised all of his complex health needs. When he was discharged in September of 1952, Woody wrote his friend Pete Seeger that he was suffering from "the mental disease my mother had, Huntington's Chorea" and that the "chorea keeps me just as dizzy [as whiskey] and [was] a good bit cheaper. I feel a thousand million times better now that I'm a[n] old dry drunk AA [Alcoholic's Anonymous] man."

The following year, in 1953, J. Edgar Hoover's FBI began interviewing his physicians. They had been keeping tabs on Guthrie for years because of his fabled connections to the Communist Party of America. The FBI agents demanded that Guthrie's doctors break patient confidentiality so that the federal government could more closely follow the folk singer's health status, thus adding one more "reason" to classify him as a potential security risk.

By 1965, Guthrie was unable to talk and could only communicate to Marjorie by flailing his arm at flash cards she made saying "No" and "Yes." All too soon, he could not even do that, but appeared to blink his eyes purposefully when Marjorie entered the room.

Woody died at age 55 on Oct. 3, 1967 at the Creedmoor State Hospital in Queens, New York. That same year, Marjorie founded the Committee to Combat Huntington Disease (CCHD), which offered care and information to afflicted families and raised funds for discovering research and medical treatments. For years, she lobbied state legislatures and the U.S. Congress to allocate more money for research on this terrible disease. Only a few months after Marjorie's death, in 1983, scientists discovered the gene that causes HD, known as HTT, on the short arm of chromosome 4. HD caused by the abnormal and multiple (36 or more) repeats of an unstable Cytosine-Adenosine-Guanine sequence in the HTT gene coding for a cytoplasmic protein known as huntingtin.

Thanks to his remarkable body of work, the splendid songs and searching prose, Woody Guthrie's legacy remains vibrant and continues to grow. His musical influence inspired numerous songsmiths of our era, including Bruce Springsteen and, perhaps most famously, Nobel laureate Bob Dylan.

Long after he sang his anthems for the everyman, Woody's legacy is also alive in the organization founded in his honor, now called **Huntington's Disease Society of America**, that continues its important work in this land, and by similar groups around the world.

By – Dr. Howard Markel

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