Harry Lee Parker and paroxysmal dysarthria and ataxia

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ABSTRACT

Objective: To review descriptions of paroxysmal dysarthria and ataxia in multiple sclerosis (MS), with special attention given to Parker and his 1946 case series.

Methods: Evaluation of original publications describing paroxysmal dysarthria and ataxia, bibliographic information, writings, and unpublished letters from the Mayo Clinic Historical Unit.

Results: In 1940, Störring described a patient with MS with paroxysmal symptoms that included dizziness and trouble speaking, but also unilateral extremity weakness. In 1946, Parker published a series of 11 patients with paroxysmal dysarthria and ataxia. Six of these patients had MS, and he recognized this phenomenon as a manifestation of the disease. The term "paroxysmal dysarthria and ataxia" was first used in 1959 by Andermann and colleagues. Since that time, paroxysmal dysarthria and ataxia has become a well-recognized phenomenon in MS. More recent reports have suggested that the responsible lesion is located in the midbrain, near or involving the red nucleus.

Conclusions: Parker was the first to accurately describe paroxysmal dysarthria and ataxia in patients with MS. *Neurology*® 2013;80:311–314

GLOSSARY

MS = multiple sclerosis.

Paroxysmal motor and sensory phenomena are well-described complications of multiple sclerosis (MS). These stereotyped events are of sudden onset and brief duration, and can occur multiple times an hour. Paroxysmal symptoms that have been reported include diplopia, trigeminal neuralgia, facial paresthesia, pain, itching, tonic seizures, akinesia, and dysarthria and ataxia.[1–3] There is disagreement about who first described paroxysmal dysarthria and ataxia in MS, with some authors giving priority to Gustav Störring and others to Harry Lee Parker.[2,4–7] Parker (figure) described patients with attacks of dysarthria and ataxia in 1946. Since then, paroxysmal dysarthria and ataxia has become a well-known phenomenon in MS, appearing in numerous neurology and MS textbooks.[1,2]

OBJECTIVE The objective of the study was to review the earliest descriptions of paroxysmal dysarthria and ataxia and its association with MS. Particular attention was given to Parker and his 1946 case series. The authors compared these early descriptions to subsequent case reports.

METHODS The authors evaluated original publications describing paroxysmal dysarthria and ataxia in patients with MS, as well as bibliographic information, writings, and unpublished letters from the Mayo Clinic Historical Unit pertaining to Harry Lee Parker. The Harry Lee Parker Collection in the Mayo Clinic Historical Unit contains a quarter foot of material. PubMed was searched using the search term "paroxysmal dysarthria and ataxia" to identify more recent case reports and case series.

RESULTS AND DISCUSSION **Biography of Harry Lee Parker.** Harry Lee Parker was born in Limerick, Ireland, in 1894. He received his medical degree from Trinity College, Dublin, Ireland, in 1918, and served in France during World War I as a volunteer in the Royal Army Medical Corps of England, achieving the rank of second lieutenant. In 1919, he came to Rochester, Minnesota, as a fellow in surgery of the Mayo Foundation, but

From the Mayo Clinic, Rochester, MN.

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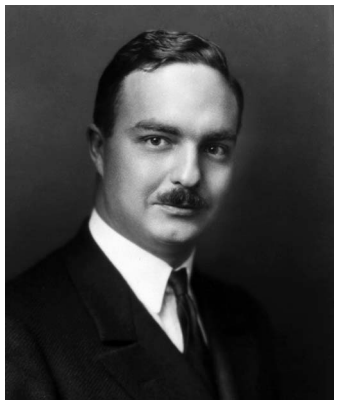
Figure Harry Lee Parker



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subsequently transferred to neurology. Parker joined the Mayo Clinic staff in 1925. Other than the years 1934–1945, when he practiced neurology in Ireland, he worked in neurology at the Mayo Clinic until his death in 1959 at age 65.[8]

Among his colleagues, Parker was known as the "wild Irishman," a tribute to his abundance of energy, hard work, and dramatic lecturing style. Dr. Frederick Moersch, Parker's colleague at the Mayo Clinic, wrote that "he was an untiring worker…in the course of a day's work he was able to consult on more patients than any of us."[9] Parker was also known for his love of life and sense of humor. For example, in 1945, on his way from Ireland back to the Mayo Clinic, he sent a telegram to Henry Woltman, chair of the Mayo Clinic neurology section, that read, "on my way full of beans seeing you soon."[10]

Academically, Parker was best known for *Clinical Studies in Neurology*, published in 1956. The book's structure was influenced by his love of Sir Arthur Conan Doyle's *The Adventures of Sherlock Holmes*. Each chapter consisted of a mysterious clinical case with the diagnosis revealed only at the end. One of the chapters featured "Soapy Mouth Burke," a patient who chewed soap until his mouth frothed, faked a convulsion to draw a crowd, and then exclaimed that the only cure for him would be some brandy.[11] In addition to this work and his description of paroxysmal dysarthria and ataxia, Parker studied trigeminal neuralgia in MS, and is referenced several times in Kinnier Wilson's *Neurology*, originally published in 1940.[12]

Original description of paroxysmal dysarthria and ataxia. Parker described the combination of paroxysmal dysarthria and ataxia and its association with MS in 1946. In that year, he reported a series of 11 patients with "periodic ataxia" in the *Collected Papers of the Mayo Clinic*.[13] The patients were evaluated at the Mayo Clinic between 1924 and 1943 and had a "clear-cut but bizarre" symptom complex consisting of "acute transient general cerebellar dyssynergia affecting all the muscles of the body, including those of articulation."[13] He commented that the attacks lasted no longer than a few seconds to a few minutes, and that they would disappear as suddenly as they came on. They could occur every few minutes to every hour, and the period during which the attacks took place varied from days to weeks. Nystagmus was noted before, during, and after an attack. Parker did not mention overbreathing as an attack trigger.

He provided the following clinical description:

> The attacks…interfere, while they are on, with all bodily activities. Speech may be reduced during the attack to a mumble, the gait is staggering to the point of falling and performance of all finer movements of the fingers and hands becomes impossible. There is marked contrast between the patient's helplessness during the attack and the relative normality between attacks. There is no loss of consciousness or even mental acuity…strength is preserved and the patient does not complain of vertigo . . . there are no emotional reactions.[13]

Parker noted that 6 of the 11 patients also had MS and recognized that "periodic ataxia" was a manifestation of this disease. As to the other 5 patients, 4 were believed to have familial cerebellar ataxia, and one patient's symptoms were of unknown etiology. As to this last patient, Parker commented that the patient's father had an atypical disease of the CNS, which could have represented late-onset MS or degenerative cerebellar spinal ataxia.

Parker's 1946 article on "periodic ataxia" did not receive much attention initially, and no additional case descriptions appeared in the literature until 13 years after his article was published.[2] In 1959, Andermann et al. [14] wrote an article for *Neurology*® entitled "Paroxysmal dysarthria and ataxia in multiple sclerosis: a report of 2 unusual cases." This article contained the first use of the term "paroxysmal dysarthria and ataxia," instead of Parker's term "periodic ataxia." Neither Parker nor his paper was mentioned in the article by Andermann et al. However, because of the attention the article by Andermann et al. received and the new interest in paroxysmal attacks in MS that the article by Andermann et al. generated, Parker's 1946 article became more widely known and cited.[2,4–7,15]

In an article by Ostermann and Westerberg[7] published in 1975 on paroxysmal attacks in MS, Parker and Dr. Gustav Störring were both credited with the earliest descriptions of paroxysmal dysarthria and ataxia. In 1940, Störring published "Epilepsie und multiple Sklerose."[16] In Störring's article (translated by author W.S.), one of the cases (case 7) is Else B., a 30-year-old woman with MS, tonic seizures, and dizzy spells. The dizzy spells she experienced usually lasted less than a minute. Dizziness was her most prominent complaint, but she also had "a hard time speaking" and she became "clumsy with her hands" during the spells. Else B.'s symptoms could represent paroxysmal dysarthria and ataxia, but she also had a

"feeling of paralysis" and being "very weak" in her right arm when examined during a spell. Paralysis is not characteristic of paroxysmal dysarthria and ataxia. It is unclear whether Else B.'s clumsiness was due to ataxia, weakness, or a combination of both. Additionally, even though her dizzy spells continued for many years, the neurologic symptoms she experienced during the spells were not stereotyped and actually varied over the years.

The clinical symptoms and signs presented by Parker in 1946 and Andermann et al. in 1959 are more consistent with the phenomenon we recognize today as paroxysmal dysarthria and ataxia associated with MS. Störring's 1940 article predated Parker's and contributed to our understanding of the paroxysmal symptoms of MS, but it cannot be considered a predecessor. Parker's 1946 "periodic ataxia" article is the first well-defined description of paroxysmal dysarthria and ataxia in the medical literature.[2,4–6]

Current studies of paroxysmal dysarthria and ataxia. By 1980, paroxysmal dysarthria and ataxia was a well-recognized phenomenon in MS and there were a total of 47 cases described in the literature.[15] However, there have been relatively few recent articles. With the advent of MRI, many of the modern articles have focused on identifying the location of the lesion responsible for causing paroxysmal dysarthria and ataxia. In a review of the available English medical literature, we identified 6 reports since 1980, representing a total of 10 cases.[4,5,17–20] Of these patients, 8 had lesions in the midbrain, 1 had a lesion in a cerebellar hemisphere, and 1 had multiple lesions involving the midbrain, pons, and cerebellar hemispheres. In addition to the reports of paroxysmal dysarthria and ataxia in patients with MS, there is one reported case occurring in a patient with neuro-Behçet disease.[21] This patient had many lesions in the periventricular white matter and brainstem. No definitive conclusions can be drawn on such a small number of cases, but the midbrain must be considered one possible site for generating the paroxysms of dysarthria and ataxia, since most reported patients have lesions in that region of the brainstem, near or involving the red nucleus.

The attacks of paroxysmal dysarthria and ataxia usually abate after weeks or months, and recurrence is unusual.[1] Carbamazepine is often used to lessen the frequency and severity of symptoms. Doses of carbamazepine ranging from 200 to 800 mg/day have been reported to be effective.[4,5,15,17–19,21] Alternative medications of benefit include phenytoin, lamotrigine, and acetazolamide.[15,19,20] Ibuprofen and bromocriptine have been used to treat other paroxysmal symptoms in MS, but not specifically paroxysmal dysarthria and ataxia, to our knowledge.[22,23]

Paroxysmal dysarthria and ataxia is now a well-recognized phenomenon in patients with MS. Harry Lee Parker first described it in 1946 as "periodic ataxia." Further research is needed before the work Parker began can be considered complete, and before the clinical phenomenon that he first described can be fully and finally understood.

AUTHOR CONTRIBUTIONS

James P. Klaas: conceptualization of the study, data analysis and interpretation, and drafting the manuscript. David B. Burkholder: data analysis and interpretation and manuscript revision. Wolfgang Singer: conceptualization of the study, translations, and manuscript revision. Christopher J. Boes: conceptualization of the study, data analysis and interpretation, and manuscript revision.

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DISCLOSURE

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