Early Controversies over Athetosis: I. Clinical Features, Differentiation from other Movement Disorders, Associated Conditions, and Pathology

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Abstract

Background: Since the description of athetosis in 1871 by American neurologist William Alexander Hammond (1828–1900) the disorder has been a source of controversy, as were many aspects of Hammond’s career.

Methods: Primary sources have been used to review controversies in the 50-year period since the initial description of athetosis, in particular those concerning clinical features, differentiation from other movement disorders, associated conditions, and pathology. Controversies concerning treatment will be addressed in a subsequent article.

Results: Hammond struggled to establish athetosis as a distinct clinical–pathological entity, and had successfully predicted the striatal pathology in his initial case (albeit somewhat serendipitously). Athetosis was, nevertheless, considered by many neurologists to be a form of post-hemiplegic chorea or part of a continuum between chorea and dystonia. European neurologists, and particularly the French, initially ignored or discounted the concept. Additional controversies arose over whether the movements persisted during sleep, whether athetosis was, or could be, associated with imbecility or insanity, and how it should be treated.

Discussion: Some controversies concerning athetosis served to identify areas where knowledge was insufficient to make accurate statements, despite prior self-assured or even dogmatic statements to the contrary. Other controversies illustrated established prejudices, even if these biases were often only apparent with the greater detachment of hindsight.

Keywords: History of neurology, nineteenth century, history of neurology, twentieth century, athetosis, dystonia, chorea, diagnosis, pathology, sleep-related movement disorders, mental retardation, dementia

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Athetosis is an involuntary movement disorder characterized by slow, smooth, sinuous, writhing movements, particularly involving the hands.1,2 Since its description in 1871 by American neurologist William Alexander Hammond (1828–1900) the disorder has been a source of controversy, as were so many aspects of Hammond’s career.3–8

Hammond was a seminal figure in the development of neurology in the United States. He was recognized as an enterprising and brilliant organizer, and during the US Civil War was appointed Surgeon General of the Union Army with a rank of brigadier-general, bypassing many more senior medical officers (Figure 1). In this capacity, he authorized the founding of the first neurological specialty hospital in the United States, the U.S.A. Hospitals for Injuries and Diseases of the Nervous System in Philadelphia, where his friend Silas Weir Mitchell (1829–1914) conducted pioneering studies of peripheral nerve injuries with colleagues George Reed Morehouse (1829–1905) and William Williams Keen (1837–1932). However, Hammond soon alienated prominent officers with his arrogance and pomposity, and was ultimately court-marshaled on flimsy and trumped-up testimony, found guilty of “irregularities” in the purchase of medical supplies, convicted of “con duct unbecoming an officer,” and given a dishonorable discharge – the only US Army Surgeon General to suffer such an indignity (although this was eventually annulled by Congress in 1879 after a re-review of the original evidence). After the
humiliation of his court martial, Hammond's friends helped him get re-established in New York City. He then began an increasingly lucrative clinical practice focusing on neurology and psychiatry, and soon became one of the highest-paid physicians in the country (Figure 2). Hammond soon became a seminal figure in the development of neurology in the United States, and his innovative organizational leadership produced new civilian specialty hospitals, a post-graduate medical school, and various other institutions and professional medical organizations. In particular, in 1875, Hammond was the principal organizing leader in founding the American Neurological Association, the first professional neurological organization. Nevertheless, Hammond's flamboyance, arrogance, and obstinacy proved to be counterproductive to fostering acceptance of neurology as a specialty, and some general practitioners and psychiatrists considered his fees (which were well above prevailing rates) not only exorbitant, but criminal. Later controversies in Hammond's roller-coaster career included his vicious public conflicts with "alienists" (psychiatrists) over the management of psychiatric illness, his attempts to undermine the authority of asylum superintendents and take control of the asylums, his "bogus" testimony in high-profile legal cases as an expert witness, his later financially and professionally disastrous entrepreneurial ventures with both his private sanatorium and various animal extracts, multiple personal legal battles, and his description and characterization of athetosis.

Although Hammond struggled to establish athetosis as a distinct clinico-pathological entity, and indeed had successfully predicted the striatal pathology in his initial case (albeit somewhat serendipitously), athetosis was nevertheless considered by many late nineteenth- and twentieth-century neurologists as a form of post-hemiplegic chorea or part of a continuum between chorea and dystonia. European neurologists, and the French in particular, initially ignored or discounted the concept. Additional controversies arose over whether the movements persisted during sleep, whether it was, or could be, associated with imbecility or insanity, and how it should be treated. The purpose of the present article is to review some of the controversies concerning athetosis in the 50-year period following its initial description. Treatment controversies will be addressed in a subsequent paper.

**Design/methods**

Reports about athetosis in the 50-year period after the description of athetosis in 1871 were identified through IndexCat (the National Library of Medicine’s online version of the 61-volume Index-Catalogue of the Library of the Surgeon General's Office, U.S. Army, Series 1–5, spanning 1880–1961), as well as through a search of
other electronic databases and search engines (Google, Google Scholar, Google Books, Internet Archive, HighWire, and PubMed), and a serial review of reference lists in articles/monographs. Images were identified from primary source documents on athetosis and through a search of various archival image sources (U.S. National Library of Medicine Images from the History of Medicine, U.S. National Archives Archival Research Catalog, the Library of Congress Prints and Photographs Online Catalog, Google Image, and Wikimedia Commons).

**Results**

**Hammond's description of athetosis**

In 1871, Hammond published the first comprehensive American textbook of neurology, *Treatise on Diseases of the Nervous System*, which ultimately went through nine editions, and which remains the most highly cited nineteenth-century American neurology textbook.\(^2\)\(^,\)\(^14\)\(^,\)\(^15\) In his textbook\(^2\) and in separate articles\(^1\)\(^,\)\(^3\) Hammond described a condition that he called “athetosis” (from the Greek term for “without fixed position”), “characterized by an inability to retain the fingers and toes in any position in which they may be placed, and by their continual motion,” and associated with “pains in the spasmodically-affected muscles, and especially complex movements of the fingers and toes, with a tendency to distortion.”\(^1\)\(^,\)\(^2\) Hammond emphasized that athetosis has a slower, more sinuous quality than chorea, and occurred in his cases without associated weakness.\(^2\)

Hammond's initial and archetypal case was a 33-year-old book-binder with a history of alcohol abuse, recurrent seizures that began around 24 years of age, and an episode of protracted delirium tremens around 29 years of age. Upon recovery from delirium tremens, he noted right-sided numbness, pain affecting the right arm and the toes of the right foot, as well as complex involuntary movements of the fingers and toes on that side that greatly diminished his ability to do fine manual tasks (Figure 3).

At first the movements of the fingers were to some extent under the control of his will, especially when this was strongly exerted, and assisted by his eyesight, and he could by placing his hand behind him, restrain them to a still greater degree… The right forearm[,] from the continual action of the muscles, was much larger than the other; and the muscles were hard and developed, like those of a gymnast. When told to close his hand, he held it out at arm’s length, clasped the wrist with the other hand, and, then exerting all his power, succeeded, after at least half a minute, in flexing the fingers, but instantaneously they opened again and resumed their movements.\(^2\)

Thaddeus Marshall Brooks (“T.M.B.”) Cross (1839–1922), then a resident physician working with Hammond at the New York State Hospital for Diseases of the Nervous System (and soon to be, under Hammond’s principal leadership, one of the seven-member organizing committee responsible for founding the American Neurological Association), noted in January 1871:

There are … involuntary grotesque muscular movements of the fingers and toes of the right side, and these are not those of simple flexion and extension, but of more complicated form. They occur, not only when he is awake, but also when he is asleep, and are only restrained by certain positions, and by extraordinary efforts of the will. Thus those of the fingers are arrested when the wrist is firmly grasped by a strong hand, or when it is less forcibly held in a vertical position. But, if the arm be extended horizontally, the fingers at once begin their movements. During their continuance the arm is hard and rigid, and the calf of the leg is also in the same state of tonic spasm while the toes are in motion. The movements are somewhat paroxysmal, being worse at times than at others… The involuntary contractions of the fingers and toes do not take place quickly, but slowly, apparently as if with deliberation and great force… The toes are not involved to the same degree as the fingers. Position does not, however, afford the same relief to them as to the fingers, and the spasms are more tonic in character. The muscular development is greater in the right arm and leg, from the almost continuous muscular action. The toes are kept restrained to some extent by the boot, but as soon as it is removed they become flexed and take on their peculiar movements.\(^2\)
Hammond, and later his son Graeme Monroe Hammond (1858–1944), published and spoke widely on athetosis, and this became recognized as the senior Hammond’s major contribution to neurological phenomenology.1–6,24–29 In 1875, Hammond presented his initial case of athetosis at the inaugural annual meeting of the American Neurological Association in New York City, and his son later presented reports of the response of this patient to nerve stretching at the eighth annual meeting held in New York City in 1882, and ultimately of the neuropathology from that case at the sixteenth annual meeting held in Philadelphia in 1890. Athetosis was subsequently often referred to as Hammond’s disease, particularly in American reviews, medical dictionaries, and monographs, although athetosis was actually a sign of a specific form of abnormal movement, rather than a syndrome or disease per se.30–35 Some European authors, while acknowledging Hammond’s description, nevertheless correctly pointed out that athetosis could not correctly be termed a disease.36 When Hammond died, athetosis was mentioned in many of his obituaries: “Hammond subsequently enriched Neurology by the discovery of Athetosis and other newly described nervous states … and his great book on Diseases of the Nervous System [sic].”37

Is athetosis a form of post-hemiplectic chorea? Or vice versa?

Several years after Hammond’s description of athetosis, his friend and colleague Silas Weir Mitchell described similar cases under the term “post-hemipletic chorea.” Mitchell noted that, “as there is a post-choreal paralysis, so, also, is there a post-hemipletic chorea… [In] adults who have had hemiplegia and have entirely recovered power, there is often to be found a choreal disorder, sometimes of the leg and the arm, usually of the hand alone” (Figure 4).23 Nevertheless, in a later monograph Mitchell was ready to consider “athetosis” or “Hammond’s disease” (of which he felt Hammond had given an “admirable account”) in a discussion of “unusual forms of spasmodic affections in women,” if only to wonder whether his patient had a hysterical component to her presentation, or, as Mitchell put it, whether the particular patient experienced “an athetosis proper, or an hysterical imitation of athetosis, or merely athetosis grown, as one might say, on an hysterical soil, and modified by its place of growth.”38 Hammond responded that “Dr. Mitchell addsuces several interesting cases in support of these propositions [concerning post-hemiplectic chorea], and quite a large number have come under my own observation. But the condition in question is an entirely different affection from athetosis, with which it has been frequently confounded.”66

Shortly thereafter, British neurologist Sir William Gowers (1845–1915) felt there was considerable clinical overlap between Hammond’s athetosis and “post-hemiplectic disorders of movement.” Gowers described similar patients in whom the movement disorder followed a sudden hemiplegia with some degree of recovery (Figures 5 and 6).21,22 Others including German neurologist Adolph von Strumpell (1833–1925) and Russian-Swiss neuropathologist Constantin von Monakow (1853–1930) described similar cases (Figures 6 and 7).30,39 Gowers considered athetosis as only one of a range of abnormal movements within a spectrum of “post-hemiplectic disorders of movement,” between the irregular “quick, clonic spasm” of chorea and the “slow, cramp-like incoordination” and tonic spasms associated with “spastic contracture.”44 As a result, Gowers was willing to accept athetosis as a recognizable state of abnormal movement within a broader continuum, with the proviso that hemiparesis could be associated, depending on the extent of the lesion. Boston neurologist James Jackson Putnam (1846–1918) in a discussion of “post-hemiplectic and allied forms of mobile spasm (athetosis)” concurred largely with Gowers, and argued that athetosis “has so many features in common with other spasmodic affections that, from the physiological standpoint at least, they must be studied in conjunction with each other.”39 Still, Putnam accepted the phenomenological distinction between the irregular quick movements of chorea and the continuous slow movements of athetosis.40

In contrast, French neurologist Jean-Martin Charcot (1825–1893) refused to consider Hammond’s athetosis as a novel disorder, and instead brushed it aside as “simply choreiform movements” or as “only a variety of post-hemiplectic hemichorea” (Figure 8).20 Charcot’s dismissal of athetosis was influential in France, as indicated in a review of the work on athetosis by Paul Oulmont (1849–1917), who had been a resident physician under Charcot at the Salpêtrière Hospital in Paris: “athetosis, an affection little known as yet, and of which the name even was until lately almost ignored in France … [Since] the labour of [Hammond],
several other observers have spoken of athetosis, chiefly in America and in England." However, even the English authors were sometimes dismissive. English neurologist Henry Charlton Bastian (1837–1915) agreed with Charcot, concluding that, “It may be pretty confidently affirmed that post-hemiplegic hemi-athetosis, is only a variety of post-hemiplegic hemi-chorea,” although he acknowledged that there was insufficient data to verify his supposition that the two disorders involved a similar locus within the brain. To these authors, and to Charcot particularly, Hammond retorted:

I have only to say that the distinction between the two conditions is as well marked as between chorea and disseminated cerebrospinal sclerosis. In athetosis the movements are slow, apparently determinate, systematic, and uniform; in post-hemiplegic chorea they are irregular, jerking, variable, and quick. Moreover, athetosis is not by any means necessarily post-hemiplegic.

In 1885 Professor Alfred George (A.G.) Barrs (1853–1934) at the Leeds General Infirmary in England, and later one of the founders of the Association of Physicians of Great Britain and Ireland, noted considerable confusion concerning the categorization of athetosis cases in the absence of prior paralysis, given the prior pronouncements of Charcot and Gowers:

We have the authority of Charcot, and with some qualification of Dr. Gowers, for regarding such cases as those published by Hammond and all such, as examples of one of the rather numerous classes of post-hemiplegic motor disturbances – as example, in short, of what has for long been known as post-hemiplegic chorea. … [The] absence of any strictly paralytic condition in the present and in many other published cases of athetosis points to its being an unusual sequence of ordinary hemiplegia. The case here published, together with those of [Paul] Oulmont and others, goes far to establish a considerable class of cases in which slow, irregular, involuntary, contortions of certain members, especially of the hands, are altogether independent of any anterior paralyzing lesion or lesion of the sensory tract. … From a clinical point of view, though easily distinguished from, they have a close resemblance, in kind at any rate, to ordinary chorea.
Hammond and his supporters, though, sought to separate athetosis as a distinct clinical entity (and ultimately a distinct clinicopathological entity) fully distinguishable from chorea and other movement disorders, rather than lumped with it. Such a true dichotomy could not be maintained, however, because some cases had features of both athetosis and chorea, either simultaneously or serially, and because many cases with clearly athetotic features did have an associated hemiparesis, despite Hammond’s initial emphasis that his original cases never had a hemiparesis. Thus, Hammond and some of his colleagues tried to assimilate post-hemiplegic chorea into a broader concept of athetosis, rather than accepting athetosis as part of the spectrum of post-hemiplegic choreiform disorders. Indeed, New York neurologist Landon Carter Gray (1850–1900), in his textbook, *A Treatise on Nervous and Mental Diseases, for Students and Practitioners of Medicine* (1892), remarked: “The so-called cases of post-hemiplegic chorea are really, as Dr. Hammond agrees with me in believing, cases of post-hemiplegic athetosis” (Figure 9).44 Some modern authors also erred in considering Hammond’s original cases as examples of a post-hemiplegic movement disorder,45,46 but, as emphasized by Hammond, “In the original case there had never been hemiplegia, nor was there such a state in the second case, on which [Hammond’s] description of the disease was based.”6 Although Hammond later accepted that hemiplegia could be an antecedent in some cases, he emphasized that this was only a superimposed or superadded feature: “Where the motor tract is implicated there will be hemiplegia, spastic spasm, and exaggerated reflexes in addition to the athetosis” [emphasis added].6 Thus, Hammond considered that the pyramidal findings were due to the extension of pathology into neural structures beyond those involved in the pathogenesis of athetosis.

In 1885, Philadelphia neurologist Wharton Sinkler (1845–1910), a protégé of S. Weir Mitchell who would later become president of the American Neurological Association in 1891, attempted to synthesize...
the confused literature by distinguishing primary and secondary forms of athetosis—an approach consistent with Hammond’s views around this time:

As originally described by Hammond, athetosis was not associated with or preceded by paralysis of the affected parts … In the great majority of cases, however, there has been hemiplegia preceding the athetoid movements; and it is, therefore, best to divide the affection into a primary and secondary form. Primary athetosis occurs without premonitory symptoms. In many instances no direct cause can be traced. … In the secondary form hemiplegia or diplegia precedes the irregular movements, and there is always some gross cerebral lesion …

Nevertheless, many preferred to incorporate athetosis into a broader conceptualization of chorea, noting that some cases included features of both types of abnormal movement, and that both could occur after hemiparesis.

In 1950, neuroanatmist Malcolm Carpenter (c. 1922–99) reviewed the literature and concluded that athetosis and chorea were separate entities, a conclusion supported by later expert reviews.

Athetosis is a pattern of involuntary dyskinesia which can be distinguished from chorea and is characterized by increases and decreases of tone in irregular sequence in antagonistic muscle groups and slow involuntary movements involving chiefly, but not exclusively, the distal appendicular musculature such that vermicular activity results … Hemiathetosis usually develops after hemiparesis, or in association with it, as a consequence of necrotizing cerebrovascular lesions which destroy part of the internal capsule and striatum on the side opposite that of the activity.

Recent definitions of athetosis have emphasized not only the relative slowness and continuity of the movements with repeated and preferential involvement of the same regions of the body (particularly the hands and feet), in contrast to chorea and myoclonus, but also the absence of identifiable submovements or movement fragments seen in chorea, the sustained postures seen in dystonia, or the rapid shock-like movements seen in myoclonus.

Can athetosis be distinguished from other movement disorders?

Hammond’s American colleagues, particularly other New York neurologists, typically had little apparent difficulty recognizing athetosis and separating it from other movement disorders. For example, New York neurologist Landon Carter Gray noted in 1893 that, “The differential diagnosis of athetosis is exceedingly easy, because the movements are not similar to that of any other form of nervous disturbance. Even in the athetoid chorea the waviness and gradual worm-like character of the movements is readily distinguished from the quickly beginning and quickly ending fibrillary jerk of the true Sydenham chorea.” Similarly, New York neurologist Moses Allen Starr (1854–1932) dismissed any likeness of athetosis to chorea, tics, myoclonus, or the “so-called rotary movements of the feeble-minded, described by Weir Mitchell, which are rhythmic and which follow one another in definite order.” As a result, Starr concluded that “it is evident that [athetosis] cannot be mistaken for anything else.”

However, the diagnostic boundaries of athetosis were apparently unclear to many other physicians, particularly it seemed to those in Europe, many of whom expressed either puzzlement or disdain for the concept. Cases of athetosis, to which the respective authors sometimes acknowledged similarities to Hammond’s cases, were nevertheless given other labels, often on the basis of flimsy distinctions. In addition, various disorders that would now be labeled as something else were lumped under the umbrella of athetosis, including cases of both chorea and dystonia. As noted by Hammond in 1893:

It is no matter for surprise that many of the cases regarded as being athetosis are not instances of that affection … A similar event took place when aphasia was first prominently brought to the notice of the medical profession. Every case of loss or impairment of the faculty of speech, whether from paralysis of
“dystonia” was similarly indiscriminately applied in many patients with various extrapyramidal disorders. Dystonia is now defined as “a syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements, or abnormal postures” or as “a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both.” Unfortunately, though, some degree of phenomenological overlap persists between athetosis and dystonia in the “twisting and repetitive movements” and in the development of secondary muscle hypertrophy, and in any case they frequently occur together.

In addition, the designation of an abnormal movement such as athetosis or post-hemiplegic chorea often depended on factors extraneous to the abnormal movement per se, including even the subsequent pathology at autopsy. For example, Fletcher Beach, the Medical Superintendent of the Darenth Asylum for Imbeciles in London, reported three cases of athetosis in 1880, but after identifying cortical pathology on autopsy of one of the cases he had been “induced to alter his opinion regarding the diagnosis of his case, and thinks it was an example of what is termed ‘post-hemiplegic choreiform movements.’” The athetosis in this case had in fact been preceded by secondarily generalized seizures, post-ictal hemiparesis, which gradually recovered over an unspecified time period (possibly a Todd’s paresis), and later a static hemiparesis. The movements, though, affected particularly the left arm and to a minor extent the left toes, and the left arm was noted to be “continually in action, and the hand is undergoing quick spasmodic movements.” Further, Beach had confidently noted with his initial report: “The movements of athetosis are very different from those of chorea. They are less jerky and less incontrollable than in the latter disease.” What changed as far as the diagnosis was concerned was not the character of the movements, but the finding of cortical pathology, which was different from the expectation for athetosis at the time.

In the long run, a greater clinical difficulty in separating athetosis from other movement disorders, particularly chorea and dystonia, resulted from an awareness that some cases included features of more than one type of abnormal movement, either simultaneously or sequentially, and that more than one type of abnormal movement could develop after hemiparesis. For example, in a review in 1902, New York psychiatrist Leon Pierce Clark described a case of athetosis, although he labeled it “athetosis.” Similarly Philadelphia neurologist William Gibson Spiller (1863–1940) described a case of generalized dystonia, and very likely an early case of dystonia musculorum deformans or torsion dystonia, in 1908, but lumped it with “athetosis” (Figure 10). Perhaps Spiller’s lumping of dystonia with athetosis explains his later statement that, “Athetosis is essentially a form of spasticity, differing from ordinary spasticity chiefly in the varying degrees of tonicity in the different muscles.” Dystonia was only separated as a clinical entity during the first half of the twentieth century, particularly by German neurologist Hermann Oppenheim (1858–1919) and later by German-American neurologist Ernst Herz (1900–65). Later a diagnosis of dystonia was proposed for such mixed cases when he noted:13

**Figure 10. Generalized Dystonia Diagnosed as Athetosis.** In 1908, Philadelphia neurologist William Spiller described a 12-year-old boy as having progressive spasticity and generalized “athetosis,” but provided clinical data that contradicted both of these findings. This was likely a generalized dystonia, and probably a case of dystonia musculorum deformans or torsion dystonia (three years before the disorder was described by German neurologist Hermann Oppenheim).
Sometimes there are similar conditions which one observer regards as athetosis, and another, perhaps, as chorea. Literature furnishes examples of cases in which there were choreic movements as well as athetosis, also of cases in which athetosis passed into chorea, or vice versa. The transitions are ... gradual ... [and] almost all authors who have written upon this subject fail to regard the necropsy findings of these conditions [as] distinctive. 36

**Does athetosis continue during sleep?**

In his original observations in 1871, Hammond had reported that athetosis persists in sleep,13 and Graeme Hammond's report in 1882 of the senior Hammond's original case suggested that the movements had persisted continuously for over a decade: “Up to the time of the operation [of peripheral nerve stretching] the movements continued uninterruptedly day and night, and thus interfered with sleep and impaired the general health of the patient.”72 In reviews or discussion on the subject, many subsequent authors simply reiterated Hammond's statement that athetosis persists during sleep, generally without evidence of direct observation during sleep. Some authors admitted that their patients could not be certain of whether the movements continued during sleep.69 Others gave a range of opinions but often hedged their statements. Sinkler, for example, stated that, “As a rule, the movements [of athetosis] do not cease during sleep.”38 Similarly Paul Oulmont stated that “they persist during rest, often even during sleep, at least to the degree of fixing the limb in an abnormal position.”42 Adolph von Strumpell stated that, “during sleep they generally cease, although in certain instances they have persisted even then, only being diminished.”70 German-Swiss internist Hermann Eichhorst (1849–1921) noted that the movements of athetosis “do not always cease during sleep, but become less marked.”71 William Spiller stated in regard to double athetosis that, “The irritative motor phenomena may persist when the patient is quiet, even in sleep.” Others reported observing that the movements stopped during sleep, sometimes based on direct observation.52,66,72–75

Anecdotal information indicated that in at least some cases the movements interfered with sleep by inducing sleep-onset insomnia (although this did not exclude other mechanisms by which they might disrupt sleep). Hammond originally stated in regard to his index case that athetosis interfered with sleep onset (as well as persisting during sleep itself): “On account of the severe pain in the whole arm, caused by the spasms in the muscles, the patient is at times unable to go to sleep until quite exhausted.”12 Similarly, in 1884, homeopathic neurologist Charles Porter Hart (1827–?) reported that, “For half an hour or so after sleep there is usually a period of comparative repose, the movements then being somewhat less severe; but sometimes the patient has great difficulty in getting to sleep, in consequence of the severity of the pain cause by the tonic contractions.”176

In 1873, on the basis of no new supportive evidence, Hammond stated emphatically and unequivocally that persistent motor activity during sleep was a discriminating feature of athetosis: “The movements in chorea cease during sleep, while those of athetosis continue.”3 However, in a clinical lecture, published in 1874, and in the sixth edition of his monograph, published in 1876, Hammond abstracted two cases—those of Thomas Clifford Allbutt (1836–1925) at the Leeds General Infirmary (already recognized as the inventor of the clinical thermometer in 1866, and later knighted in 1907) and William Tennant Gairdner (1824–1907), professor of medicine at the University of Glasgow—where the authors reported that the movements ceased during sleep.4,5,77,78 The abstracted cases were meant to show that others had recognized and reported cases of the disorder, and Hammond made no comment at the time on the discordant reports of the movements stopping when the patients were asleep. Others, though, were not reticent to comment. When Hammond recognized these reports as legitimate cases of athetosis, Gowers noted, somewhat acerbically, that this eliminated continuation of the movements during sleep as a discriminating factor for diagnosis.21

In one case [reported by Hammond] it is noted that the movements continued during sleep, and it is implied that this was the case in all [such cases]. ... But Dr. Hammond claims as instances of athetosis cases in which, as in that of Dr. Clifford Allbutt, the movements ceased during sleep, and much weight, therefore, cannot be attached to the distinction.21

In fairness to Hammond, however, Allbutt was not in fact as clear as Gowers had made out: According to Allbutt's report, “Unlike Dr. Hammond's patients, however, Mrs. D.'s spasmodic movements ceased, or almost ceased, during sleep.”77 Nevertheless, Hammond clearly understood the increasing evidence that at least in many otherwise acceptable cases of athetosis the abnormal movements were reported to stop during sleep. In subsequent editions of his textbook, he made no general claim that athetosis persists during sleep, or that this feature can be used to distinguish athetosis from chorea.

Limited later anecdotal information suggested that athetosis may persist only during certain stages of sleep. In a posthumously published review in 1906, Denver neurologist Jeremiah Eskridge (1848–1902) reported “four cases of hemi-athetosis in which the movements ceased during profound sleep, but persisted to a moderate degree during light sleep.”79 Presumably this observation was based on that made earlier by German neurologist Albert Eulenberg (1840–1917).80

The confused literature on this issue was never clarified with a detailed observational study of the movements during sleep in a representative sample of cases. Although subsequent reviews have tended to indicate that athetosis may persist during sleep, no systematic observational study has apparently been done in a representative sample of cases (for example, including unilateral post-hemiplegic cases, unilateral symptomatic cases sans hemiparesis, and bilateral cases).

**Is athetosis associated with imbecility or insanity?**

The relationship between abnormal movements and neuropsychiatric disorders was considered by many late nineteenth- and early
twentieth-century authors, and indeed occasional cases of hemi-
athetosis were reported in association with either cognitive impairment
or insanity, with cognitive impairment most commonly noted in
congenital cases of double athetosis.42,66,67,69,70,71,83 Strümpell stated
that, “The intelligence of the patient [with double athetosis] is
sometimes, but not always, diminished.”70 Bastian stated that,
“Sometimes athetosis occurs as a bilateral affection, and in this form
it is most common in imbecile children.”42 With regard to double
athetosis, Spiller stated that, “Imbecility is almost constant, but many
authors have observed that intelligence is intact.”82

Influential Scottish psychiatrist J. Batty Tuke (1835–1913) at a
meeting of the Medico-Psychological Association in Glasgow on June
10, 1873, which he chaired, affirmed that athetosis was an appropriate
topic for consideration by psychiatrists: “for if it is not insanity of the
mind, it is insanity of the muscles.”78 The quaint concept of “insanity
of the muscles” had in fact been applied decades before in regard to
chorea, as noted as early as 1841 by British physician Sir Thomas
Watson (1792–1882) (Figure 11),84,85 and subsequently applied later
by others.52 In 1871, Watson explained the “ingenious theory out of
which this expression has arisen”:85

A certain portion of the encephalon ministers to the intellectual
functions; certain altered states of that portion lead to mental
aberration; the persons thus affected form false judgments;
cannot associate their ideas aright. So also a certain portion of
the encephalon presides over the locomotive functions; and there
are altered states of that portion, which lead to a loss of the due
association of the muscular contractions; to insanity of the
muscles.” [Original emphasis]85

In 1873, London physician Thomas Claye Shaw (1841–1927)
described cases of congenital bilateral athetosis under the rubrics of
“imbecility with ataxia” and “ataxic imbecility,” leaving as he said the
appellation of athetosis “to designate those where the muscular
movements come on as a disease subsequent to a previous state of
health,” i.e., only the acquired forms (Figure 12).52 More importantly,
Shaw distinguished the progressive dementia of what he called
“choreic insanity” (Huntington’s disease) from the static mental
retardation in his patients.52

Chorea, if long continued or existing to any great extent, leads to
a condition of dementia more or less complete; but here, though
there is congenital feebleness of intellect, this feebleness does not
increase; on the contrary, when educated, the subjects of it show

![Figure 11. British Physician Sir Thomas Watson, c. 1873. Watson
applied the concept of “insanity of the muscles” in regard to chorea as early as
1841. This concept was later applied to athetosis. Photograph by Barraud &
Jerrard, Photographers, London. Courtesy of the U.S. National Library of
Medicine.](http://www.tremorjournal.org)

![Figure 12. Cognitive Impairment and Athetosis. Patient with cognitive
impairment and athetosis reported by British physician Thomas Claye Shaw in
1873. Occasional cases of hemi-athetosis were reported in association with either
cognitive impairment or insanity, with non-progressive cognitive impairment most
commonly noted in congenital cases of double athetosis. Unlike the situation with
Huntington’s disease, adult-onset hemi-athetosis was not associated with either
progressive dementia or psychiatric dysfunction.](http://www.tremorjournal.org)
Early Controversies over Athetosis: I

Lanska DJ

John Hughlings Jackson (1835–1911) had applied Willis’ concepts 22 years. There was a lesion involving the posterior thalamus, Hammond, in 1890 after the patient had suffered with athetosis for 6,28,29 autopsy of the original case that was reported by his son, Graeme Hammond, in 1890, confirmed Hammond’s prediction. Furthermore, “the direct motor tract in the internal capsule was not involved to any extent.” Hammond’s prediction and the subsequent confirmation are often regarded as a landmark in the clinicopathologic correlation of movement disorders, and specifically in the linkage of abnormal movements to pathology of the basal ganglia.

Those cases in which athetosis exists and those in which it does not occur are about equal in ability to undertake trades. Possibly the non-athetotic are slightly duller students, but this defect offsets the physical agitation of the affected parts in the athetotic class.47

Moses Starr dismissed any associated cognitive impairment as incidental: “The association with weak-mindedness, in some cases, is accidental, for in the majority of cases recorded the mental state of the individual has been perfect” as it was even in the two childhood cases Starr had observed.20

In retrospect, many cases with congenital bilateral athetosis had varying degrees of non-progressive mental retardation, while adult-onset unilateral cases typically had normal intellects, and, unlike the case with Huntington’s disease, were not associated with development of dementia or psychiatric dysfunction.

Does athetosis have a specific and unique pathology?

Hammond speculated in his initial report in 1871 that “one probable seat of the morbid process is the corpus striatum,” based largely on Thomas Willis’ (1621–75) archaic concept that the corpus striatum is the seat of motor power. Hammond was in good company in making such a localization: only several years earlier British physician William Broadbent (1835–1907) and British neurologist John Hughlings Jackson (1835–1911)²⁷ had applied Willis’ concepts and implicated the striatum in chorea. All of these localizations were essentially correct, but somewhat serendipitous, based as they were on a misunderstanding of the anatomy of the motor pathways.

Hammond’s proposed localization was ultimately supported by the autopsy of the original case that was reported by his son, Graeme Hammond, in 1890 after the patient had suffered with athetosis for 22 years.6,28,29 There was a lesion involving the posterior thalamus, part of the internal capsule, and the lenticular nucleus (Figure 13). Graeme Hammond “called attention to the fact that the motor tract was not implicated in the lesion, and claimed that this case was further evidence of his theory that athetosis was caused by irritation of the thalamus, the striatum, or the cortex, and not by a lesion of the motor tract.”²⁶

By the ninth edition of his textbook in 1891 (revised and corrected in 1893), updated with the collaboration of his son Graeme, the senior Hammond was able to collect 13 cases of athetosis from the literature since 1871 in which autopsies were obtained.⁶ Pathology was recognized in “either the cortex, the thalamus, or the striatum” in the reported cases, but in no case was the lesion confined to the corticospinal fibers in the subcortical white matter, causing Hammond to conclude that “athetosis is a distinct pathological entity.” Hugo Summa (1859–1917), Professor of Pathology and Clinical Medicine at Marion-Sims College of Medicine in St. Louis, in a very negative and sarcastic review of the ninth edition of Hammond’s textbook, nevertheless argued cogently that athetosis should not be considered as a distinct clinicopathologic entity given that it did not have a unique nervous system localization.⁶⁶

Cases of double athetosis generally supported pathological localization to the corpus striatum and particularly the putamen.⁶²,⁹⁴ In Étude clinique sur l’athéose (Paris, 1878), Paul Oulmont had described athéose double ou athéose générale (i.e., bilateral athetosis) as a severe form of cerebral palsy with bilateral athetosis and developmental delay.⁴¹,⁹⁰,⁹¹ In L’athéose double, Jean Audry (1858–1950) reviewed 93 cases with chorea and athetosis and observed corresponding basal ganglia and cerebral atrophy.⁹² In 1896 Austrian neuropsychiatrist Gabriel Anton (1858–1933) found bilateral putaminal lesions with patches of “hypermyelinated” nerve fibers in congenital cases of double athetosis (Figure 14).⁹³,⁹⁴ In a series of papers beginning in 1911 French-German neuropathologist Cécile Munger Vogt (1873–1962) along with several collaborators defined the “corpus striatum syndrome”
(also called Vogt’s disease and Vogt’s syndrome), usually associated with athetoid cerebral palsy, and characterized pathologically by what she called \( \text{\`etat marbre\`} \) (i.e., marble state), due to abnormal myelinated fibers in the corpus striatum. Later, Spiller also found that the putamen was severely affected in such cases (Figure 15): Each lenticular nucleus was about one-half the normal size. The globus pallidus on each side was firm but the putamen on each side had a worm eaten appearance and contained numerous small holes. … The globus pallidus was atrophic, as was also the nucleus caudatus [caudate nucleus], but neither showed the peculiar tissue seen in the putamen.

Although Cécile Vogt had delineated status marmoratus as a specific pathological entity of presumed prenatal onset, in 1921 Boston child neurologist Bronson Crothers (1884–1959) and later authors instead stressed the likelihood that perinatal factors, including birth injury, were responsible for cases of double athetosis. The association of athetosis with neonatal jaundice in low birthweight babies was established in the 1960s, subsequently, aggressive treatment of perinatal hyperbilirubinemia produced a decline in chronic bilirubin encephalopathy so that kernicterus is now a rare cause of dyskinetic cerebral palsy.

**Discussion**

Although British physician and anatomist Thomas Willis (1621–75) had suggested a role for the corpus striatum in controlling movement as early as the late 1600s, the functions of the extrapyramidal system were poorly understood in the latter half of the nineteenth century at the time of Hammond’s description of athetosis. Prior to 1870, the cerebrum was often considered in somewhat vague terms to subserve the functions of the mind, including the capacities that underlie consciousness, intelligence, and “the will.” The cerebral cortex was not thought then to be directly responsible for motor activity, in part because 1) stimulation of the cerebral cortex to that point had failed to produce muscular contractions; and 2) anatomists had failed to...
recognize the continuity of white matter tracts from the cortex passing through the corpora striata en route to the spinal cord, and instead held to the presumption that the tracts originated in the corpora striata. Even the term extrapyramidal wasn’t introduced into neurology until British neurologist Samuel Alexander Kimner Wilson (1878–1937) introduced it in his paper on “Progressive lenticular degeneration: a familial nervous disease associated with cirrhosis of the liver” (which he later insisted must be called Wilson’s disease, even though it had been reported earlier by others).103

The role of the motor cortex itself was first really appreciated in the 1870s, following the electrophysiologic stimulation studies of German physiologists Gustav Fritsch (1838–1927) and Eduard Hitzig (1838–1907) on the cerebral cortex of dogs,104 the stimulation and ablation experiments on various animals by British physiologist David Ferrier (1843–1928),105 and the careful clinical and clinical-pathologic studies in people by John Hughlings Jackson (1835–1911). By 1876 Jackson considered the “motor centers in Hitzig and Ferrier’s region … higher in degree of evolution than the corpus striatum,”106 Particularly with Kinnear Wilson’s subsequent electrical stimulation and ablation studies of different parts of the corpus striatum in 1914 (with the assistance of Sir Victor Horsley), many of the previously assigned functions of the corpus striatum were recognized as properties of neighboring corticospinal pathways, and consequently the corpus striatum “seemed to fall from its high estate and depreciate in physiological significance.”106

Whatever functions the corpus striatum once possessed there is no experimental evidence in apes to show that it exercises any motor function comparable to that of the motor cortex. There is no evidence to suggest that it is a centre for so-called automatic movements … and comparatively large unilateral lesions do not give rise to any unmistakable motor phenomena.106

Nevertheless, some of the early clinicopathological studies of athetosis sans hemiparesis,20,25 and later pathological studies of double athetosis by Vogt,26 indicated that lesions of the basal ganglia did result in disorders of motor function in humans, findings which were reinforced by Wilson’s studies of hepatolenticular degeneration,103 and studies of other basal ganglia diseases by New York neurologist James Ramsay Hunt (1874–1937) among others.107,108 Later experimental work in primates by Margaret Kennard (1899–1975) at Yale in the 1930s and 1940s also demonstrated that tremors and other motor disturbances did develop after large bilateral lesions of the striatum.109–111 Thus, the corpus striatum came to be viewed as one of the major components of the “extrapyramidal motor system,” whose normal function was to help regulate movement rather than to initiate movement per se.45,107,108,112

Development of clinicopathological correlations for athetosis and other extrapyramidal disorders in the late nineteenth century, and into the early twentieth century, was hampered by clinical confusion and consequent diagnostic imprecision and error regarding the distinctions between different forms of abnormal movements, failure to establish groups of clinically similar cases for study (e.g., with similar circumstances and clinical findings, thus increasing the likelihood of similar underlying neuropathology), reliance on infrequent autopsies of well-studied clinical cases, limited available neuropathological stains and techniques, limited neuroanatomical information regarding the connections of the basal ganglia, lack of understanding of the role or function of these structures, and lack of an animal model.

In the 50-year period since the initial description of athetosis, there were no careful systematic observational studies of the continuation of abnormal movements during sleep in a representative sample of patients with athetosis, and in large measure this has still only been superficially addressed. Certainly a modern understanding of sleep stages had to await the development of human electroencephalography by German physiologist and psychiatrist Hans Berger (1873–1941) around 1924, and subsequent development of commercial electroencephalogram (EEG) machines and further clinical studies in several countries in the 1930s, recognition of the various sleep stages from the late 1930s through the 1950s, and the development of video-EEG monitoring in the 1960s.113,114 In the 1960s some studies began to look at small numbers of patients with choreoathetosis attributed to Huntington’s disease or Sydenham’s chorea, but these cases could undoubtedly be classified more as cases of different forms of chorea than of athetosis per se.115,116 Nevertheless, it was recognized that among the selected small sample of cases with “choreo-athetotic movements,” the abnormal movements often lessen or disappear with falling asleep or in light sleep, but can reappear transiently, especially with lightening of sleep, or with global or gestural body movements in the absence of either awakening or lightening of sleep.115,116 Later polysomnographic studies of Huntington’s chorea or drug-induced choreoathetosis reported that “abnormal EMG discharge groupings corresponding to clinical choreo-athetosis decreased considerably during sleep; but they could appear in any sleep stage except stage 4 with the same EMG characteristics as in wakefulness.”117

In some cases the early controversies concerning athetosis served to identify areas where knowledge was as yet insufficient to make accurate statements, despite prior self-assured or even dogmatic statements to the contrary. Such questions could potentially be addressed by a systematic review of existing cases, as was repeatedly attempted at least at a superficial level, but often a more thorough prospective analysis of additional case material was necessary to resolve disagreements, often in conjunction with more refined case definitions, attention to collection of representative case material, stratification or selection into uniform clinical types, and comparison to appropriately selected controls.

In other cases, the controversies concerning athetosis served to illustrate established prejudices, even if these biases were often only apparent with the greater detachment of hindsight. Seldom were polarized parties converted to opposing viewpoints, despite the escalation of claims and counterclaims. Instead, the open debates served to sway others without such vested interests to adopt one view or the other, depending often on the established authority of the respective parties, rather than on the basis of the arguments presented.
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