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# Cervical spondylotic myelopathy in the young adult: A review of the literature and clinical diagnostic criteria in an uncommon demographic

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**Key Words:** cervical spondylotic myelopathy; diagnostic criteria; gait disturbance; spasticity

**Running Head:** “Cervical spondylotic myelopathy: A review of the literature”

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## **Abstract**

**Background** Cervical spondylotic myelopathy (CSM) is typically encountered in the elderly population. Significant inconsistencies currently exist regarding the definition of the disorder, the true incidence of CSM in younger populations, and the established diagnostic criteria.

**Objective** To highlight the lack of standardization in the definition and diagnosis of CSM.

**Methods** A PubMed literature search was conducted spanning the years 2001 to 2011. The search was limited by the following terms: 1) English language, 2) Adults (19-44 years old), and 3) “cervical spondylotic myelopathy.” Each article was reviewed to determine if the presence of the definition of CSM existed in the article. The clinical characteristics used to make the diagnosis of CSM were recorded for each article. Cochran’s Q statistic was used to determine whether some clinical characteristics were more frequently used than others.

**Results** 93 papers were reviewed in detail and 16 case reports, reviews, and articles concerning less than three patients were excluded, resulting in 77 articles in the final analysis. The most common clinical definitions were gait disturbance (22/77 articles (28.6%)), upper limb paresthesias or sensory disturbance (21/77 (27.3%)), and clumsy hands (15/77 (19.5%)). Hyperreflexia, spasticity, and pathologically increased reflexes were identified as diagnostic criteria in a minority of patients.

**Conclusion** The literature employs a wide range of neurologic signs and symptoms to make the diagnosis of CSM, with a majority of studies failing to rely on strict diagnostic criteria. The clinician should not discount CSM as an explanation for the aforementioned findings, as it is well-reported in the literature among the ages 18-44.

**Key Words:** cervical spondylotic myelopathy; diagnostic criteria; gait disturbance; spasticity

## **Introduction**

Cervical spondylotic myelopathy (CSM), the most common disorder of the spinal cord in persons older than 55 years of age, remains a challenging pathology to manage in modern neurosurgical practice.[1] As the mean age of the population continues to rise, it can be expected that an increasingly frequent number of patients will present with the signs and symptoms of CSM. However, among patients with CSM, there is considerable variability in both the clinical presentation and imaging findings(Figure 1-4).[2] The temporal evolution of symptoms covers a wide range, spanning from acute to chronic progression and neurologic deterioration.[3-5], the optimal management strategy must be tailored to each individual patient and multiple surgical approaches are routinely utilized.[6-9] Despite the high frequency with which CSM is encountered in clinical practice, the diagnostic criteria continues to suffer from a significant lack of uniformity.

The diagnosis of CSM is primarily based on the clinical signs found on physical examination and is supported by imaging findings of cervical spondylosis with cord compression.[2] Numerous authors have addressed the defining clinical characteristics and diagnostic criteria, however, a careful analysis of the literature reveals multiple inconsistencies. The underlying discrepancies are multifactorial in nature, stemming from the complexity of the disease and the vast number of treatment options currently available. Additionally, the existing literature emanates from diverse specialties, including, neurology, neurosurgery, internal medicine, and rehabilitation medicine. Practitioners from different specialties encounter patients of varying acuity of symptom onset, thereby contributing to the multiple approaches to

treatment. As a result, the comparison of data across studies, such as, indications for surgery, complications, and treatment efficacy is of limited utility.

Utilizing the PubMed search engine, the literature over the last ten years (2001 – 2011) pertaining to the clinical presentation, diagnosis, and surgical management of CSM were reviewed. Specifically, articles were reviewed to determine if CSM was explicitly defined, to pin point the diagnostic criteria used, and to identify discrepancies and inconsistencies between manuscripts. Additionally, we also present a review of the presenting findings in the literature of younger patient populations affected with CSM (ages 19-44).

## **Methods**

Using the PubMed search engine, a search was conducted spanning the years 2001 to 2011. The search specifically focused on the following limiting terms: 1) English language, 2) Adults (19-44 years old), and 3) “cervical spondylotic myelopathy.” Each article was individually reviewed and analyzed to determine if the authors explicitly defined CSM within the manuscript. The clinical characteristics used to make the diagnosis of CSM were recorded for each article as follows: gait disturbance, lack of coordination, clumsy hands, paresis, spasticity, hyperreflexia, hyporeflexia, sensory disturbance, pattern of weakness, Lhermitte’s sign, Romberg’s sign, Hoffman’s sign, Babinski’s sign, and bowel or bladder dysfunction.

Cochran’s Q statistic was used to determine whether some clinical characteristics were more frequently used than others. Cochran’s test allows analysis of data where the same item occurs in more than one category. A critical range was calculated to indicate which characteristics were selected at significant rates.[10]

## **Results**

93 articles meeting the search criteria were identified. 16 case reports, reviews, and articles concerning less than three patients were excluded, resulting in the inclusion of 77 articles in the present study. The breakdown of the signs and symptoms are presented in Table 1 (**Table 1**). Cochran's Q statistic indicated that the rates were significantly different, with a critical range of 16.37%.

Gait disturbance, cited in 22 (28.57%) of the papers reviewed, was the most common sign or symptom found in patients diagnosed with CSM. Reported in 21 (27.27%) of the papers, loss of sensation in the upper extremities was the second most common finding. The critical range of the Cochran Q test was 16.37%. Gait disturbances were therefore significantly more frequently used in CSM diagnoses than lower limb weakness ( $28.57\% - 11.69\% > 16.37\%$ ) or any of the less frequently used criteria. Similarly, upper limb paresthesia or sensory disturbance was significantly different from lower-limb hyperreflexia or any less common characteristic ( $27.27\% - 10.39\% > 16.37\%$ ).

Upper extremity spasticity and hyperreflexia were identified as diagnostic criteria in 15.58% and 14.29% of papers, respectively. Lower extremity spasticity and hyperreflexia, which appeared in 8 (10.39%) and 7 (9.09%) papers, respectively, were not described as frequently as identical findings in the upper extremities. Pathologically increased reflexes were noted in a minority of studies: Hoffman's sign (6.49%) and Babinski's sign (5.19%). Eleven different signs or symptoms were found to be reported as diagnostic criteria with a frequency between 1.30% and 9.09%.

## **Discussion**

CSM remains a poorly defined clinical entity due, in large part, to the widely varying diagnostic criteria employed by practitioners. As a result, it is difficult to make meaningful

comparisons between data sets, particularly when attempting to define indications for surgery, outcomes, and improvement or decline in functional status. As demonstrated in this review, the diagnosis of CSM should not be regarded as solely a disease of the elderly. Numerous studies are cited in the literature, as in our search which found 7510 patients across 77 papers. The present study also highlights the inconsistencies within the existing literature and identifies signs and symptoms commonly attributed to CSM.

### *Long-tract Signs*

Long tract signs, such as, upper and lower extremity spasticity and hyperreflexia, serve as evidence of a disease process arising in the upper motor neurons of the corticospinal, spinothalamic, and posterior column tracts. In patients with CSM, degenerative changes in the cervical spine result in compression of the spinal cord and resultant upper motor neuron dysfunction. As a result, long-tract signs are generally regarded as hallmarks of CSM.[11-14] In the present study, however, only a relatively small number of articles cited these findings. Signs consistent with pathologically increased reflexes, such as Hoffman's (5.38%) and Babinski's (4.30%) signs, were also noted in only a small percentage of studies.

The relatively low frequency with which these findings were included among the diagnostic criteria was perhaps the most striking evidence of the poorly defined parameters. Most often, evidence of CSM on MRI was the major inclusion criteria, in place of long tract findings on examination. Still, findings on MRI were not standardized across studies, with broadly generalized spondylosis and cord compression being the most routinely found description. Other potential explanations for the disparities, include, improper diagnosis of a subset of patients or the inclusion of patients with varying degrees of disease progression under an all-encompassing term of CSM. However, we chose a younger age group to limit the

variability in disease progression than occurs in later adulthood. In either case, the comparison of data between studies is rendered inadequate.

The differential diagnosis for a patient presenting with myelopathy is broad and includes, inflammatory, infectious, vascular, traumatic, degenerative, toxic/metabolic, and neoplastic processes.[15] Few papers in our series attempted to pursue diagnostic testing beyond imaging to establish a diagnosis of CSM, thereby raising the question of misdiagnosis. When evaluated in the context of the widely varying diagnostic criteria described above, the diagnostic accuracy is considered even more suspect. Patients with true CSM presenting without hyperreflexia or spasticity on exam are most likely presenting earlier in their disease course. Inclusion of these patients with those more profoundly affected skews data interpretation, as the natural history of CSM is variable and difficult to predict.[16-18]

### *Symptoms*

Gait disturbance (28.57%) and loss of sensation in the upper extremities (27.27%) were the most commonly identified presenting symptoms. However, they were noted in less than a third of the articles reviewed and there was no significant difference between the frequency of their reporting and several other commonly cited neurologic findings. Additionally, eleven different signs or symptoms were found to be reported as diagnostic criteria with a frequency between 1.30% and 9.09%. These results highlight the variability in diagnostic criteria between studies and the broad range of findings utilized by various authors in making the diagnosis of CSM.

Multiple scores, including, the Nurick-score, modified Japanese orthopaedic association score (mJOA score), Cooper myelopathy scale (CMS), Prolo-score, and European myelopathy score (EMS) have been developed in an attempt to standardize the diagnosis of CSM.[19-24]

These scores are commonly referenced in the literature and are most extensively used in the research setting. They are rarely employed in clinical practice and their utility is limited by the fact that they are largely based on the ability of patients to participate in activities of daily living.[2] As a result, they do not reflect objective criteria on neurologic examination, results are inherently “user” dependent, and meaningful interpretation across studies is limited. In our analysis, we found the majority of papers to report the JOA score, which is calculated from answers provided by patients on a questionnaire pertaining to their neurologic function. Obviously, this scoring system relies heavily on the interpretation of symptoms by an individual patient and lacks stringent criteria when attempting to define diagnostic criteria or outcomes. Nikaido et al. highlighted these inconsistencies in a prospective analysis of 87 patients who had undergone surgical treatment of CSM.[21] The authors reported a lack of improvement in upper extremity function on JOA scores despite the fact that improved function was clearly observed on objective testing.

The present study is limited by its retrospective design and the broad spectrum of study types from which data was pooled. As mentioned above, the majority of studies did not pursue advanced diagnostic testing to establish a true diagnosis of CSM. Thus, there is significant suspicion of inclusion of patients without CSM. Also, due to the very nature and goals of the study, the inconsistencies inherent in the papers limit analysis of the data. Nevertheless, we have effectively demonstrated the need for specific standards for reporting in CSM, given its high prevalence as a surgical disease. Furthermore, the data emphasizes the need for prospective analysis of patients presenting with symptomatology consistent with CSM and highlights the shortcomings of existing patient-centered clinical scales. Recent prospective studies in the past five years have been published that specify strict radiographic criteria for defining spondylotic

disease. However, standards should be adopted to make for accurate comparison, as these radiographic criteria vary between studies, as well as the clinical criteria.

Surgical decompression has been shown in previous studies to limit disease-progression and improve myelopathy [25-32]. However, a specific subgroup analysis comparing earlier onset CSM as defined by our age range 18-44 and their respective surgical outcomes is lacking. The authors were able to identify the frequency of this disease where possible in the literature, however, due to the lack of a common myelopathy grading scheme (JOA, mJOA, Nurick, Oswestry) and unifying age group, conclusions on surgical outcome are restricted to that of prior studies.

## **Conclusion**

The literature demonstrates a wide range of neurologic signs and symptoms that are employed to make the diagnosis of CSM, with the majority of studies failing to employ stringent diagnostic criteria. The clinician should not discount CSM as an explanation for the aforementioned findings, as it is well-reported in the literature among the ages 18-44.

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## Table Legend

**Table 1. Incidence of individual signs/ symptoms found in reviewed papers.** Gait disturbance, which was identified in 28.57% of papers, was the most commonly cited sign or symptom. Eleven different signs and symptoms were reported with a frequency between 1.30 and 9.09%. Data was analyzed by Cochran's Q test and the critical range was 16.37%.

Sign/ Symptom	Upper/ Lower Extremity Involvement	Number of Papers	Percentage of Papers
Gait disturbance		22	28.57%
Pattern of loss of sensation (paresthesia/sensory disturbance)	UL	21	27.27%
Clumsy hands		15	19.48%
Spasticity	UL	12	15.58%
Pattern of loss of sensation (paresthesia/sensory disturbance)	LL	12	15.58%
Hyperreflexia	UL	11	14.29%
Pattern of weakness	UL	11	14.29%
Pattern of weakness	LL	9	11.69%
Hyperreflexia	LL	8	10.39%
Spasticity	LL	7	9.09%
Bowel/bladder dysfunction		7	9.09%
Lack of coordination		6	7.79%

Paresis	UL	6	7.79%
Paresis	LL	5	6.49%
Hoffman's Sign		5	6.49%
Babinski's Sign		4	5.19%
Lhermitte's Sign		2	2.60%
Romberg's Sign		2	2.60%
Hyporeflexia	UL	1	1.30%
Hyporeflexia	LL	1	1.30%

### Figure Legend

**Figure 1.** MRI, sagittal T2-weighted imaging of the cervical spine demonstrating pronounced focal cervical stenosis between C3 and C6. Hyperintensity of the cervical spinal cord is seen, most pronounced at C4.



Figure 2. MRI, axial T2-weighted imaging of the cervical spine at the level of C4-C5 showing a circumferential absence of cerebrospinal fluid signal.

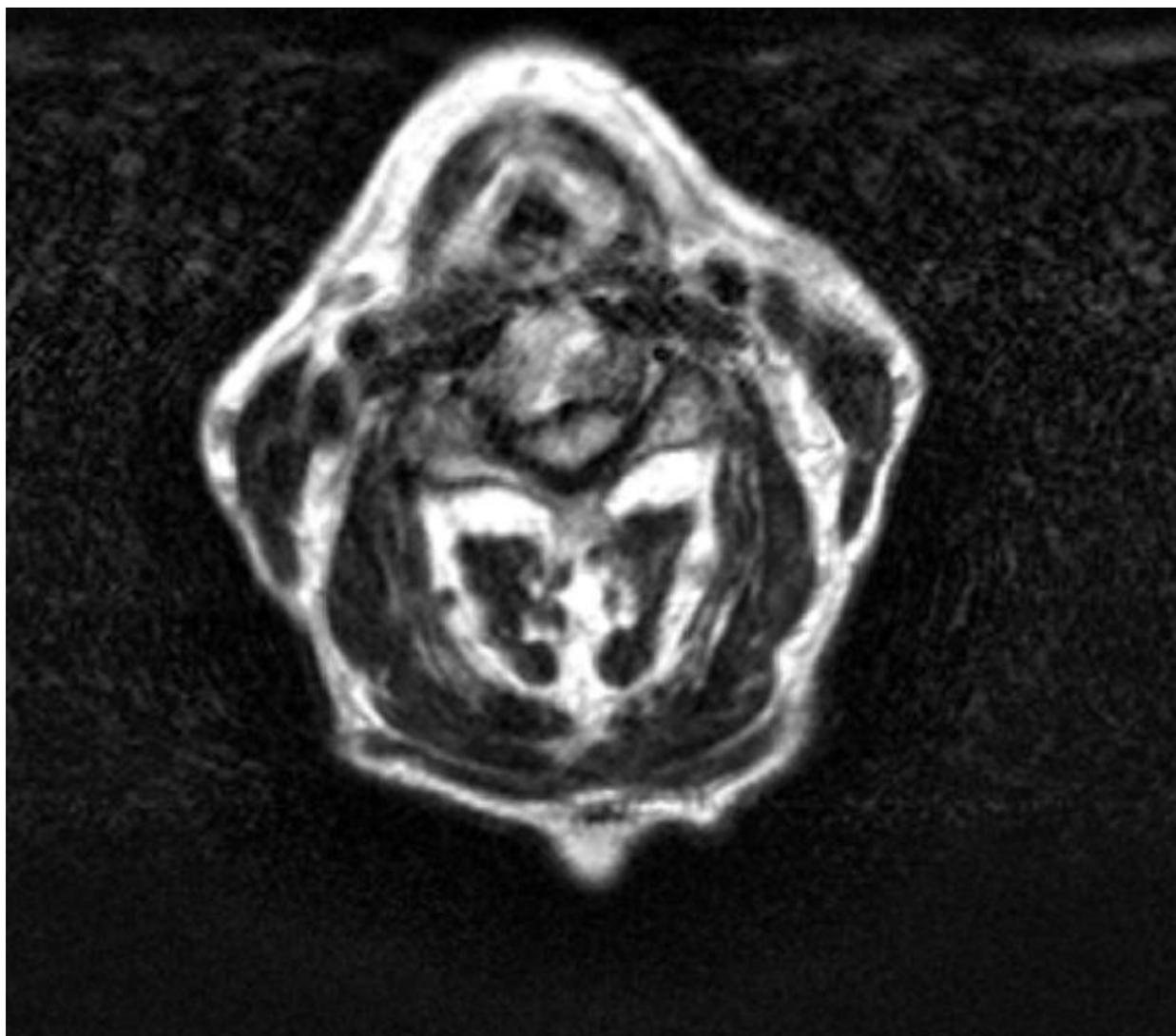


Figure 3-4. CT, sagittal and corresponding C4-C5 axial views of the cervical spine showing minimal to moderate degenerative spondylosis of the neck.



