The DOWN Questionnaire: A Novel Screening Tool for Cervical Spondylotic Myelopathy

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Abstract

Study Design: Case-control study.

Objectives: Cervical spondylotic myelopathy (CSM) is the most common cause of spinal cord injury in adults aged over 55 years. However, since the onset is typically insidious, accurately diagnosing CSM can be challenging, often requiring referral to a subspecialist and advanced imaging. To help identify patients at risk for CSM, this case-control study compared responses to a series of 4 questions (DOWN questionnaire) in myelopathic and non-myelopathic patients.

Methods: Ninety-two patients, 46 with and 46 without myelopathy, were recruited for the study. Each patient answered 4 questions encompassing common symptoms associated with CSM. Responses between patient groups were compared, and Cohen’s $\kappa$ was used to assess for agreement between responses and the diagnosis of myelopathy.

Results: We found a sensitivity of 91% and a $\kappa$ of 0.54 to 3 positive responses and a sensitivity of 72% and a $\kappa$ of 0.61 to 4 positive responses.

Conclusions: Positive responses to 3 or more DOWN questions has high sensitivity and moderate agreement with the diagnosis of myelopathy based on history, physical exam, and review of advanced imaging by an orthopedic or neurological surgeon. The DOWN questionnaire is a potentially useful screening tool to identify patients at risk for CSM.

Keywords
cervical, degenerative disc disease, disc herniation, MRI, spondylitis, myelopathy, spondylosis

Introduction

Cervical spondylotic myelopathy (CSM) is a compression of the cervical spinal cord due to degenerative changes that leads to neuronal damage and dysfunction. The development and progression of degenerative changes in the cervical spine is associated with aging.¹ While some studies have estimated the prevalence of CSM to be 1.6 per 100,000, the exact incidence and prevalence remains unknown.²,³ Nonetheless, CSM is the leading cause of spinal cord dysfunction worldwide and the most common cause of spinal cord dysfunction in adults age 55 or older.³,⁴,⁵,⁶

The natural history of CSM was classically described by Clarke and Robinson. Their work revealed that patients with CSM have progressive neurological decline that culminates in paralysis and potentially death.⁷ Surgical intervention is typically indicated to alter the natural progression of the disease and prevent further neurological decline.⁵ While CSM is a well-known entity in the neurology and spine surgery communities, it is less appreciated in the general medical community. One study reported a mean delay in diagnosis of 2.2 years in patients with CSM, with 69% of patients initially presenting to

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a family practitioner. This is significant because early diagnosis and intervention results in better patient outcomes. In particular, Ebersold et al found that the duration of disease before surgical intervention was the only significant variable predictive of outcome. Since CSM decreases patients’ quality of life in all health domains, including emotional and mental health, early identification and intervention has a profound impact on patients’ overall well-being.

There are several instruments that quantify the severity of CSM such as the Nurick score, the Japanese Orthopaedic Association score (JOA) and its modified version (mJOA), and the Cooper Myelopathy Scale. Due to their complexity, these instruments are not frequently used in the clinical setting or for screening and instead are typically used in research settings. To our knowledge, there is no validated screening tool for CSM. Consequently, a simple, effective screening tool that can be utilized to diagnose CSM would be extremely valuable. To address this, we created the DOWN questionnaire to identify patients with CSM that should be further evaluated both clinically and with advanced imaging.

Methods

Study

After institutional review board approval, patients were prospectively recruited to participate in the study at the University of Southern California (USC) spine center clinics. Patients were recruited from November 1, 2016, to June 30, 2017. Any adult patient (age >18) that presented with advanced cervical spine imaging (ie, magnetic resonance imaging or computed tomography myelogram) was invited to participate in the study. Patients that were <18 years old, had previous cervical spine surgery, tumors or neoplasms, infection, history of cerebrovascular accident, history of neuromuscular disease, worker’s compensation, or imaging studies greater than 12 months old were excluded. The diagnosis of CSM was determined by 1 of 5 fellowship trained spine surgeons based on advanced imaging and objective clinical exam findings of spinal cord compression, with or without subjective symptomatology. Participants were divided into 2 groups: those with a diagnosis of CSM (study group) and those without a diagnosis of CSM (control group). After obtaining informed consent, patients in both groups completed the DOWN questionnaire as well as the mJOA. The mJOA consists of 4 categories: motor dysfunction in upper extremities, motor dysfunction in lower extremities, sensory dysfunction, and sphincter dysfunction. Patients selected the statement in each category that best describes their symptoms and level of dysfunction. Points are allocated based on the severity of dysfunction in each category. The point values from each category were summed, resulting in a score ranging from 0 (worst) to 18 (best). mJOA responses were classified as mild (≥15), moderate (12-14), or severe (<12) based on the criteria of Fehlings et al for both groups. Demo- graphic data, smoking history, clinical diagnosis (other than CSM), and length of symptoms were also obtained.

Questionnaire

CSM may present with a constellation of symptoms including difficulty walking, unsteady gait, upper extremity weakness or numbness, diminished dexterity, and changes in bladder control. The mJOA score succinctly captures these symptoms by asking questions about upper and lower extremity function, sensory changes, and bladder function. To further streamline the process, we focus on gait instability, dexterity, and motor/sensory function in the upper extremities. In our practice, we found that these symptoms are frequently associated with CSM. For instance, one study of patients with CSM reported that gait disturbances occurred in 80% of patients, clumsiness in 67%, weakness of arms in 47%, and numbness of arms in 84%.

The DOWN questionnaire is composed of 4 questions that encompass common symptoms associated with CSM in both the literature and practice experience at USC. DOWN is an acronym representing the following questions:

1. Have you noticed that you are Dropping things or that your hands feel clumsy?
2. Have you felt more Off-balance or unsteady on your feet?
3. Do you feel Weakness in one or both of your arms or hands?
4. Do you feel Numbness or tingling in one or both of your arms or hands?

Responses to each question were recorded in a binary fashion, yes or no. Responses were then converted to numerical data, with 1 point for every “yes” response and 0 points for a “no” response. The sum of responses to the 4 questions was recorded allowing for a maximum of 4. We hypothesized that this questionnaire will adequately screen for CSM with a 3-item positive response criterion (≥3).

Statistics

Cohen’s κ coefficient was calculated to determine the agreement between the physician’s diagnosis of CSM and the questionnaire diagnosis of CSM. A priori power analysis determined that a sample size of 91 patients would be required for 85% power in a 1-sided test of H1: \( \kappa > 0.5 \) versus H0: \( \kappa \leq 0.5 \) computed at \( \kappa |H_1 = 0.7 \) and \( \alpha = 0.05 \). Pearson \( \chi^2 \) analyses were run to evaluate the correlation between a 3-item positive response and the diagnosis of CSM, as well as a 4-item positive response and the diagnosis of CSM. Fisher exact tests were performed to determine the association between the physician’s diagnosis of myelopathy and mJOA severity classification, the questionnaire 3-item positive diagnosis of CSM and mJOA severity classification, and the questionnaire 4-item positive diagnosis of CSM and mJOA severity. Sensitivity and specificity were also determined for the 3-item and 4-item positive responses. Statistical analyses were performed using SAS software version 9.4 (SAS Institute, Inc, Cary, NC) and STATA version 14.2 (StataCorp LP, College Station, TX).
Receiver operating characteristic curve analysis was performed using MATLAB R2013B (Natick, MA).

**Results**

Ninety-two patients, 46 patients in both the control and study groups, from the outpatient clinic at the USC spine center were recruited according to the inclusion and exclusion criteria described in the methods. Basic demographic statistics of the sampled population is shown in Table 1. Overall, the study population had a mean age of 59.7 years, a mean duration of symptoms of 27.6 months, with the majority being Caucasian and male. The only statistical difference in demographics between groups was mean age (66.0 myelopathic vs 53.4 non-myelopathic, \( P < .001 \), by 2-sample t test). The age range for patients with myelopathy was 36 to 93 years, while the age range for patients without myelopathy was 19 to 77 years.

Shown in Figure 1 is the distribution of DOWN responses of myelopathic and non-myelopathic patients in our study. As can be seen, patients with myelopathy tended to have higher DOWN scores. The average DOWN score for patients with myelopathy was 3.6 (standard deviation of 0.8), while the average DOWN score for patients without myelopathy was 1.9 (standard deviation of 1.3). This difference was statistically significant under a 2-sample t test (\( P < .001 \)).

To evaluate the accuracy of the DOWN questionnaire under different cutoff scores, we constructed a receiver operating characteristic (ROC) curve (Figure 2). We found an area under the curve (AUC) of 0.89 (95\% confidence interval = 0.82-0.96), indicating that the DOWN questionnaire has very good accuracy.\(^{13} \) Although a DOWN score of 2 had a sensitivity of 96\%, it had a specificity of only 37\%. We therefore restricted further analyses to scores of 3 to 4.

Shown in Table 2 are the sensitivity, specificity, positive likelihood ratios, negative likelihood ratios, agreement, and Cohen’s \( \kappa \) under 3 and 4 positive responses to the DOWN questionnaire. We found that the highest accuracy (80\%) between the diagnosis of myelopathy and the DOWN questionnaire occurs with a cutoff of 4 positive responses. At this cutoff, the DOWN questionnaire had a sensitivity of 72\%, a specificity of 89\%, and a Cohen’s \( \kappa \) of 0.61—indicating a substantial agreement beyond chance.\(^{14,15} \) For 3 positive responses, the accuracy was 77\%, with a sensitivity of 91\%, a specificity of 63\%, and a Cohen’s \( \kappa \) of 0.54—indicating moderate agreement beyond chance. Given that the diagnosis of myelopathy requires a detailed history, a full neurological exam and confirmation with advanced imaging, it is surprising that the DOWN questionnaire can achieve this level of accuracy.

**Table 1. Basic Demographics of Study Population.**

| Patients, n (%) | Myelopathic (n = 46) | Non-Myelopathic (n = 46) |
|-----------------|----------------------|--------------------------|
| Age, mean (SD), years | 66.0 (14.6) | 53.4 (14.4) |
| Men | 32 (70) | 27 (59) |
| Race/ethnicity | | |
| African American | 3 (7) | 3 (7) |
| Asian | 6 (13) | 6 (13) |
| Caucasian | 27 (59) | 27 (59) |
| Hispanic | 9 (20) | 7 (15) |
| Other | 1 (2) | 3 (7) |
| Current smoker | 4 (9) | 6 (13) |
| Duration of symptoms, mean (SD), months | 26.8 (43.9) | 28.3 (66.9) |

**Figure 1.** Responses to DOWN questions in myelopathy and non-myelopathy patients.

**Figure 2.** Receiver operating characteristic (ROC) curve of DOWN score.

**Table 2. Performance of DOWN Score Under Different Cutoffs.**

| Score | Sensitivity | Specificity | PLR | NLR | Accuracy | \( \kappa \) |
|-------|-------------|-------------|-----|-----|----------|---------|
| 3     | 0.913       | 0.6304      | 2.4706 | 0.1379 | 0.7717 | 0.54 |
| 4     | 0.7174      | 0.8913      | 6.6 | 0.3171 | 0.8043 | 0.61 |

Abbreviations: DOWN, Dropping, Off-balance, Weakness, Numbness; PLR, positive likelihood ratio; NLR, negative likelihood ratio.
The design of the DOWN questionnaire was motivated by the simplicity and success of the CAGE questionnaire as a screening tool to identify patients at risk for alcohol abuse or dependence. The CAGE questions have been shown to have a significant impact on the ability of physicians to screen for alcohol abuse. ROC analyses of the CAGE questions have an AUC of 0.89 to 0.91 which is identical to the AUC of the DOWN questions that we observed.

While the incidence of CSM is unknown, it is the most common cause of spinal cord dysfunction in adults age 55 or older. Moore and Blumhardt evaluated 585 patients with nontraumatic spastic paraparesis or tetraparesis and found CSM was the most common diagnosis (23.6%). In our study, the mean age of patients diagnosed with CSM was 66. This is consistent with other literature which found a mean age of 64. In the United States, this population demographic will continue to grow in the coming years. According to the US Census Bureau, by 2030 there will be 72 million people (approximately 1 in 5) over the age of 65.

Table 3. Sensitivity and Specificity of DOWN Questions.

|       | D | O | W | N |
|-------|---|---|---|---|
| Sensitivity | 0.91 | 0.83 | 0.93 | 0.91 |
| Specificity  | 0.67 | 0.70 | 0.43 | 0.26 |

Abbreviations: D, Dropping; O, Off-balance; W, Weakness; N, Numbness.

Table 4. Distribution of Symptom Severity in Patients with and without Myelopathy.

|                | mJOA Category |
|----------------|---------------|
|                | Mild | Moderate | Severe | Total |
| (-) Myelopathy | 40   | 6       | 0      | 46    |
| (+) Myelopathy | 9    | 10      | 27     | 46    |
| Total          | 49   | 16      | 27     | 92    |

Abbreviation: mJOA, modified Japanese Orthopaedic Association.

Table 5. Distribution of Symptom Severity in Patients with DOWN Score of 4 and < 4.

| DOWN Score | mJOA Category |
|------------|---------------|
|            | Mild | Moderate | Severe | Total |
| <4         | 44   | 8       | 2      | 54    |
| 4          | 5    | 8       | 25     | 38    |
| Total      | 49   | 16      | 27     | 92    |

Abbreviations: DOWN, Dropping, Off-balance, Weakness, Numbness; mJOA, modified Japanese Orthopaedic Association.

To determine how individual questions in the DOWN questionnaire contributed to its performance, we calculated the sensitivity and specificity for each question (Table 3). We found that the weakness question had the highest sensitivity while the balance question had the highest specificity.

To understand the severity of impairment due to myelopathy in our study sample, we used the mJOA score. As shown in Table 4, 37/46 patients (80%) had a moderate-severe degree of impairment as assessed by the mJOA score. Of patients identified as having a high likelihood of having myelopathy (DOWN score 4), 33/38 (86%) had a moderate-severe degree of impairment as assessed by the mJOA score (Table 5).

Discussion

In this study, we validated the DOWN questionnaire, a novel screening tool for CSM. Using a simple, 4-question screen we found that patients answering yes to 3 or 4 questions were statistically significantly more likely to be myelopathic. In our study, positive responses to 3 questions had a sensitivity of 91% for detecting myelopathy. Given that high sensitivity is desirable in a screening test since such a test would rarely miss patients with the disease, we propose that the DOWN questionnaire can be used as a simple and effective tool to screen for CSM using a 3-affirmative response threshold.
can be routinely administered by primary care physicians and gerontologists, to screen at-risk populations, helping diagnose patients earlier and avoiding the morbidity associated with diagnostic delays. Since patients with CSM are frequently misdiagnosed with carpal tunnel syndrome and cervical radiculopathy, we propose that the DOWN questions be used in any patient presenting to primary care physician who complains of neck, arm or hand pain.

There are some limitations to this study. First, the sample size of 92 patients is relatively small. We believe this reflects the challenges of prospectively recruiting patients with the intent to develop a new screening tool. While the sample size is small, given the strength of our findings, it is unlikely that increasing the sample size will significantly alter the findings of this study so as to invalidate it. In addition, this sample size is commensurate with the literature and many of the cited work in this article that address CSM. Furthermore, the a priori power analysis determined that we reached 85% power with this sample size. Second, patients were not recruited in an unbiased fashion—all patients had advanced imaging prior to referral to the USC spine center. As such, there was already a clinical suspicion for some form of cervical spine pathology that warranted imaging and referral. This “referral filter” has implications for study the generalizability of our findings. In our control group, the most common diagnosis was cervical radiculopathy (63%). In these patients, numbness, tingling, pain, and weakness in the arm or hand are common symptoms particularly when the radiculopathy involves the lower cervical levels. As a result, in our control group, many patients with cervical radiculopathy have a DOWN score of 2 (positive responses to the weakness and numbness questions), leading to a narrow difference between patients with CSM and patients with simple radiculopathy. This is reflected in the relatively low specificity for DOWN scores of 3 and 4, in our sample population, of 63% and 89%, respectively. However, in a more broad-based population with a smaller percentage of radiculopathy patients, the specificity of the DOWN questions will likely be higher. Similarly, other disease states (eg, multiple sclerosis or stroke) could confound the accuracy of the DOWN questionnaire, and as such, future studies to validate our findings in a different patient population would be helpful. A third limitation of our study is that the DOWN questions were chosen a priori. Ideally, a large number of questions would have been given to both patient groups and then the small subset of questions that resulted in the highest diagnostic accuracy could have been identified. This approach was not pursued in this study because of logistical difficulty and poor patient compliance—it was already challenging to simply enroll patients in this study in a busy clinic setting. Despite having to choose questions a priori, our finding of a sensitivity of 91% for a DOWN score of 3 argues that the DOWN questionnaire is useful as a screening test for CSM (in this patient population). Nevertheless, given that the DOWN questionnaire was tested in a subspecialty setting, we encourage further study and recommend validation with a large group of patients in a primary care setting.

Conclusions
The DOWN questionnaire adequately screens for CSM using a 3-affirmative response threshold. Patients reaching this threshold should be treated with a high index of suspicion for CSM and obtain advanced imaging.

Declarati on of Conflicting Interests
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