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REVIEW ARTICLE

# Cervical spondylotic amyotrophy: a systematic review

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## Abstract

**Purpose** Cervical spondylotic amyotrophy (CSA) is characterized by upper limb muscle weakness and atrophy, without sensory deficits. The pathophysiology of CSA has been attributed to selective injury to the ventral nerve root and/or anterior horn of the spinal cord. This review aimed to delineate the history of CSA and to describe the epidemiology, etiology, pathophysiology, classification, clinical features, radiological and electrophysiological assessment, diagnosis, differential diagnosis, natural history and treatment of CSA.

**Methods** A comprehensive search of PubMed, EMBASE, Cochrane library and Web of Science databases was conducted, from their inception to April 3, 2018.

**Results** Clinically, CSA is classified into three types: a proximal-type (involving the scapular muscles, deltoid and biceps), a distal-type (involving the triceps and muscles of the forearm and hand) and a diffuse-type (involving features of both the distal- and proximal-type). Diagnosis requires documentation of muscle atrophy, without significant sensory deficits, supported by careful neurological, radiological and neurophysiological assessments, with amyotrophic lateral sclerosis, Parsonage–Turner syndrome, rotator cuff tear and Hirayama disease being the principle differential diagnoses. Conservative management of CSA includes cervical traction, neck immobilization and physical therapy, with vitamin B12 or E administration being useful in some patients. Surgical treatment, including anterior decompression and fusion or laminoplasty, with or without foraminotomy, is indicated after conservative treatment failure. Factors associated with a poor outcome include the distal-type CSA, long symptom duration, older age and greater preoperative muscle weakness.

**Conclusion** Although the disease process of CSA is self-limited, treatment remains challenging, leaving scope for future studies.

## Graphical abstract

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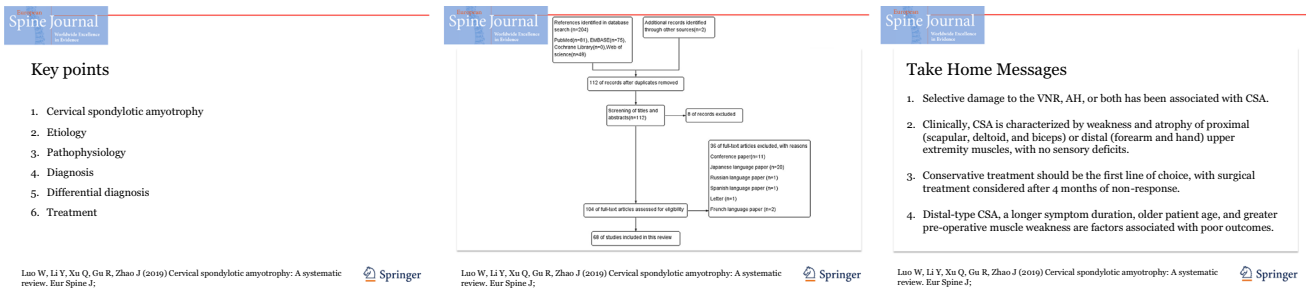


**Keywords** Cervical spondylotic amyotrophy · Etiology · Pathophysiology · Diagnosis · Differential diagnosis · Treatment

## Introduction

Cervical myelopathy and radiculopathy secondary to cervical spondylosis are well-known cervical spine disorders. Occasionally, patients with cervical spondylosis present

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with weakness and wasting of upper limb muscles, without significant sensory deficits, a condition known as cervical spondylotic amyotrophy (CSA) [1–3]. Because of its rarity, the pathophysiology, clinical features and surgical outcomes of CSA are not well understood, and consequently, treatment remains controversial. Therefore, the purpose of our review was to delineate the history of CSA and to describe its epidemiology, etiology, pathophysiology, classification, clinical features, radiological and electrophysiological assessment, diagnosis, differential diagnosis, natural history and treatment.

## Materials and methods

We conducted a structured search of PubMed, EMBASE, Cochrane library and Web of Science using "cervical spondylotic amyotrophy" as the main search term. Considering the historical perspective of our review, publication dates from the inception of each database to April 3, 2018, were included. The flow diagram of our search is shown in Fig. 1. A total of 206 relevant publications were identified: PubMed ($n=81$), EMBASE ($n=75$), Cochrane ($n=0$), Web of Science ($n=48$) and additional sources ($n=2$). After exclusion of duplicate titles, 112 publications were retained. Review of the titles and abstracts reduced the number of articles to 104, with a further detailed review retaining 68 articles for final inclusion in the review. The characteristics of these articles are summarized in Table 1.

### Historical perspectives

Brain et al. [4] first described cases of cervical spondylosis with upper limb muscle atrophy, without sensory disturbance or pyramidal signs, in 1952. In 1965, Keegan described these cases as a "dissociated motor loss of the upper extremities with cervical spondylosis" [5], with Sobue et al. [6] confirming segmental myelopathy as the root cause of this selective muscle atrophy and recommending use of the term CSA in 1975. Since then, CSA has been defined as a clinical syndrome of cervical spondylosis resulting in severe muscular atrophy of the upper extremities, without sensory deficit or pyramidal signs [2, 7–10]. Of note, however, Iizuka et al. [11] did include the presence of pyramidal tract signs in their description of CSA.

### Epidemiology

The exact incidence of CSA has yet to be established. In their case series of 653 patients who underwent cervical

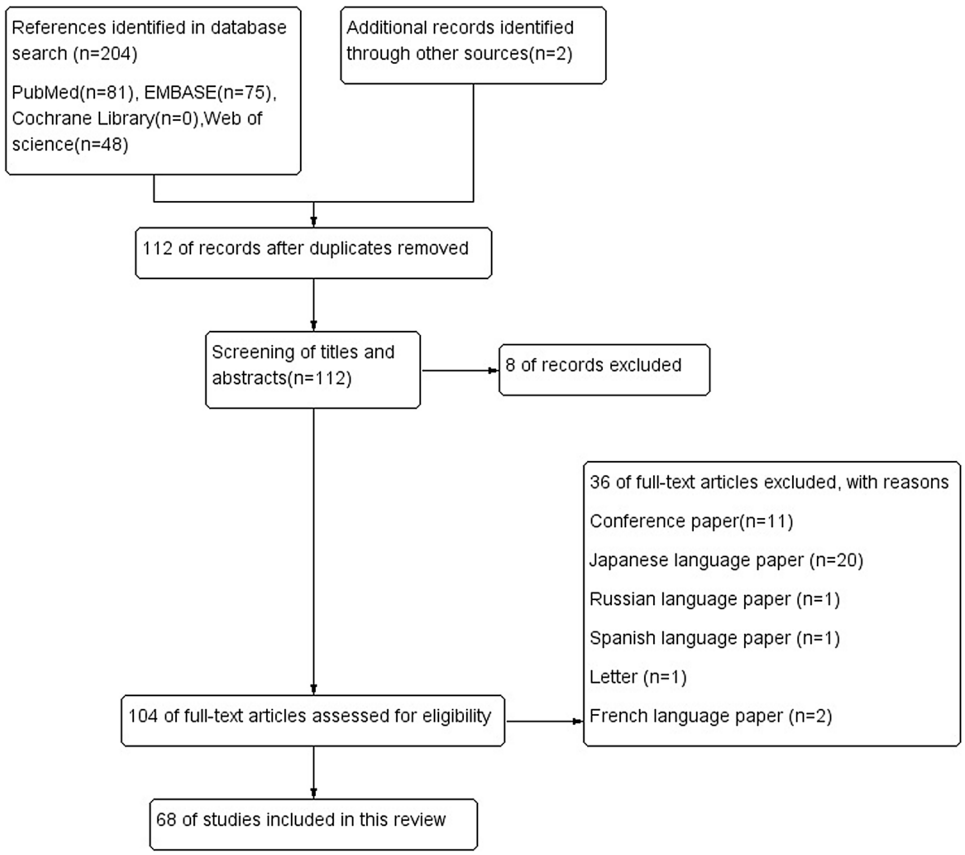


**Fig. 1** Flowchart of article identification and selection

**Table 1** Characteristics of the studies included in the review

| References | Country of origin | References | Country of origin |
|---|---|---|---|
| Kaneko et al. [1] | Japan | Yanagi et al. [2] | Japan |
| Tofuku et al. [3] | Japan | Brain et al. [4] | United Kingdom |
| Keegan [5] | United States | Sobue et al. [6] | Japan |
| Imajo et al. [7] | Japan | Sasai et al. [8] | Japan |
| Funaba et al. [9] | Japan | Fujiwara [10] | Japan |
| Iizuka et al. [11] | Japan | Zhang et al. [12] | China |
| Hashiguchi et al. [13] | Japan | Kameyama et al. [14] | Japan |
| Asaka et al. [15] | Japan | Srinivasa Rao and Rajshekhar [16] | India |
| Zhang et al. [17] | China | Mori et al. [18] | Japan |
| Shinomiya et al. [19] | Japan | Takebayashi et al. [20] | Japan |
| Tauchi et al. [21] | Japan | Fujiwara et al. [22] | Japan |
| Wang et al. [23] | China | Tauchi et al. [24] | Japan |
| Jin et al. [25] | China | Shibuya et al. [26] | Japan |
| Imajo et al. [27] | Japan | Ahdab et al. [28] | France |
| Hatanaka et al. [29] | Japan | Nouri et al. [30] | Canada |
| Mizuno et al. [31] | Japan | Gebere-Michael et al. [32] | Japan |
| Kumar et al. [33] | Ethiopia | Iwata et al. [34] | Japan |
| Sasaki and Iwata [35] | Japan | Koda et al. [36] | Japan |
| Badar et al. [37] | India | Zhang and Wang [38] | China |
| Inui et al. [39] | Japan | Jiang et al. [40] | China |
| Uchida et al. [41] | Japan | Shibuya et al. [42] | Japan |
| Park and Park [43] | South Korea | Stark et al. [44] | United Kingdom |
| Fang et al. [45] | China | Iwata et al. [46] | Japan |
| Miller et al. [47] | United States | Rowland [48] | United States |
| Yamada et al. [49] | Japan | Ghasemi [50] | Iran |
| Kuwabara et al. [51] | Japan | Kuncl et al. [52] | United States |
| Ellenberg et al. [53] | United States | Tauchi et al. [54] | Japan |
| Sonoo et al. [55] | Japan | Shindo et al. [56] | Japan |
| Jiang et al. [57] | China | Hatanaka et al. [58] | Japan |
| Zheng et al. [59] | China | Hirayama et al. [60] | Japan |
| Okumura and Homma [61] | Japan | Baxter et al. [62] | Canada |
| Meng et al. [63] | China | Kong et al. [64] | China |
| Liu et al. [65] | China | Terai et al. [66] | Japan |
| Dorsen and Ehni [67] | United Kingdom | Ebara et al. [68] | Japan |

spine surgery, Zhang et al. [12] identified seven patients with a distal-type CSA (incidence rate of 1.1%).

## Etiology and pathophysiology

Different opinions regarding the etiology and pathophysiology of CSA have been put forward. Imajo et al. [7] attributed CSA to a selective intradural compression of the ventral nerve root (VNR) by posterolateral osteophytes, while author researchers have suggested that CSA develops from an intrinsic injury to the anterior horn (AH) of the spinal cord [13–16]. Some researchers have included injury to the VNR and/or AH as the cause of CSA, including injury to the intradural anterior rootlets [17–24]. Injury to the AH could result from insufficient blood supply, secondary to stenosis of the spinal canal of the cervical spine or from compression/stretching of the intra- and extramedullary vessels with cervical spine movement [1, 9, 25], which can result in multi-segmental injury and loss of AH cells [2, 14, 16, 22, 26–30]. Tofuku et al. [3] reported good-to-excellent clinical outcomes in the treatment of CSA using hyperbaric oxygen therapy in ten patients, which supports the hypothesis of circulatory insufficiency to the AH as being a possible cause of CSA. Confirmation of this specific pathophysiological pathway, however, is difficult due to the inherent difficulty in measuring blood flow in the anterior spinal artery at the level of the cervical spine. Using spinal cord-evoked potentials, Takebayashi et al. [20] confirmed AH involvement in distal-type CSA, with involvement of a greater number of intervertebral levels

for distal- compared to proximal-type CSA [12]. Mizuno et al. [31] reported an association between hypertrophy of the posterior longitudinal ligament and CSA. Imajo et al. [7] used the amplitude of the compound muscle action potential (CMAP) to differentiate between AH and VNR involvement, with the following criteria proposed: a > 50% amplitude of the CMAP, compared to the unaffected side (CMAP of 5–10 mV), being indicative of AH involvement, with a decrease ≤ 50% in CMAP, compared to the unaffected side, being indicative of involvement of both the AH and VNR. In patients with a normal central motor conduction time (CMCT), Imajo et al. [27] proposed that an increase > 10% in CMAP amplitude, relative to the unaffected side, was indicative of involvement of the VNR. In their case report of three patients with CSA, Kameyama et al. [14] identified symmetrical high intensity signals in the intramedullary region, with a "snake eye"-like appearance (SEA), indicative of intrinsic AH pathology.

## Classification and clinical presentation

The muscle atrophy of the upper limbs associated with CSA is generally classified into two subgroups, according to the predominant muscles affected: proximal-type CSA (with atrophy of the scapular muscles, deltoid and biceps) and distal-type CSA (with atrophy of the triceps and muscles of the forearm and intrinsic muscles of the hand) [7, 17, 18, 23, 25, 27]. However, some authors have classified CSA into three types: proximal-type (with atrophy of the deltoid, biceps and triceps), distal-type (with atrophy of the muscles of the forearm and intrinsic muscles of the hand) and diffuse-type (with atrophy of both proximal and distal muscles) [1, 3, 16]. According to Srinivasa Rao and Rajshekhar [16], distal CSA rarely progresses to include the proximal musculature, although Gebere-Michael et al. [32] did report a case of proximal-type CSA, with a bilateral symmetrical presentation, that progressed to include distal muscle groups. Generally, CSA affects patients between the ages of 40 and 70 years, with a unilateral presentation [13, 33]. However, a bilateral presentation is possible [28]. and Kaneyama et al. [14] did report a case of bilateral proximal-type CSA with an asymmetric presentation. In the same way, while CSA is generally associated with drooping shoulders (due to involvement of the scapular muscles with proximal-type CSA) [2, 9, 14, 16, 25, 34], Ahdab et al. [28] did report occurrence of a drooped head in a patient with CSA due to atrophy of the neck extensor muscles, in addition to the shoulder girdle muscles.

With regard to involvement of the pyramidal tracts, a decrease in deep tendon reflexes of the upper extremities has been reported for both proximal- and distal-type CSA [18, 35], including the fingers in some patients [9, 25, 36].

## Radiological examination

A high intensity zone, on T2-weighted magnetic resonance (MR) imaging, indicative of spinal cord compression and spinal canal stenosis, has been reported for both proximal- and distal-type CSA [1, 23], in addition to the unique SEA neuroimaging findings in the spinal parenchyma at the level of muscle atrophy [32, 37, 38]. Of note, however, in their detailed MR imaging for three patients with CSA, Mori et al. [18] did not observe any abnormality in signal intensity in the intermedullary area. Querin et al. [69] reported that CSA was associated with gray matter atrophy between C2 and C6 vertebral levels, with a specific increase in gray matter density in the motor and extra-motor regions. The increase in cortical volumes detected in some patients with CSA may reflect a compensatory mechanism to the progressive gray matter degeneration of the spinal cord, resulting from central nervous system plasticity [69]. With regard to the spinal levels involved, proximal-type CSA normally involves C3/4 or C4/5 intervertebral levels, corresponding to a compression of the C5 or C6 spinal nerve roots, respectively, while the distal-type CSA is usually associated with spondylosis of the spinal canal and the foramen of C5–C6, C6–C7 or C7–T1 intervertebral levels, with involvement of the C7–T1 nerve roots [1, 20, 39]. Overall, involvement of the C5–C6 spinal segment is most common for distal-type CSA and C4–C5 for proximal-type CSA [12, 16, 40, 41].

## Electrophysiological examination

No consistent electrophysiological findings have been reported. Shibuya et al. [42] identified lower muscle activity amplitudes of the abductor digiti minimi (ADM), compared to the abductor pollicis brevis (APB), while Jin et al. [25] reported a normal ADM-to-APB ratio. Jin et al. [25] reported a lower CMAP ulnar-to-median nerve ratio among patients with CSA ($1.21 \pm 0.53\%$), compared to healthy controls ($1.15 \pm 0.23\%$; $P > 0.05$). Park et al. [43] identified the F-wave latency of the ulnar nerve as the only parameter of statistical significance between patients with CSA and healthy controls ($P < 0.001$). Stark et al. [44] reported a gradation of abnormalities in electromyographic (EMG) signals of patients with distal-type CSA, with definite changes in C7 myotome and more severe changes in C8 and T1 myotomes. Clinical examination, in combination with whole muscle and single-fiber EMG studies, has confirmed a greater loss of AH cells at the levels of C8–T1, with a moderate loss at the level of C7 [44], with no abnormal findings at the level of thoracic paraspinals and lower limb muscles [1, 32]. With regard to differential diagnosis, the ADM-to-APB CMAP amplitude ratio was significantly higher among patients with ALS compared to controls ($P < 0.001$), with this ratio being

lower among patients with distal-type CSA compared to controls ($P < 0.001$) [45].

## Diagnosis and differential diagnoses

The diagnosis of CSA is based on a comprehensive examination of the clinical presentation and disease course, supplemented by neurological examination, imaging and electrophysiological studies [9, 18], and exclusion of other possible disorders which might account for the findings [7, 25, 26, 28, 46]. Exclusion of sensory deficit is a consistent (and often considered to be a mandatory) finding of CSA [5, 11, 21, 23, 34, 39, 45, 46]. Unilateral or bilateral weakness or muscles atrophy of the upper limbs is additional important clues of possible CSA, with the possibility of pyramidal signs among patients with a cervical spondylosis, but without gait disturbances [11, 23, 24, 39, 45, 46]. As previously stated, a high intensity SEA area on axial T2-weighted MR images may be of benefit for CSA diagnosis [3, 14, 16]. Clinically, the symptoms of CSA are similar to those of motor neuron disease and other neurological conditions, underlining the importance of differential diagnosis, with the principal differential diagnoses being amyotrophic lateral sclerosis (ALS), rotator cuff tears, Hirayama disease (HD), Parsonage–Turner syndrome [47] and posterior interosseous nerve (PIN) palsy [32, 34].

### Amyotrophic lateral sclerosis

Both ALS and CSA share common clinical profiles, including a typical presentation in middle age and elderly individuals, making it difficult to distinguish between these two conditions in the early stages of the disease [3, 21, 48]. However, there are key differences between ALS and CSA. ALS is characterized by a progressive degeneration of motor neurons in the primary motor cortex, brainstem and spinal cord that leads to death [43, 49], with a peak onset between the ages of 50 and 75 years [50]. Of note, severe atrophy of the thenar muscles is a rare occurrence in patients with ALS [43, 51], despite early involvement of the hand in 41–53% of cases of ALS [51]. Drooping of the head and shoulders, with paraspinal wasting, is additional important localizing signs of ALS [52]. Careful electrophysiological examination of the tongue, facial muscles and the sternocleidomastoid and/or trapezius muscles, to detect bulbar disease, may be the key to making the proper diagnosis of ALS [53]. Specifically, atrophy or fasciculation of the tongue, involvement of the bulbar muscles, dysarthria and dysphagia, atrophy of the neck musculature and diffuse atrophy of the muscles of the upper extremities, while common in ALS [21, 24] or uncommon in CSA [54]. With regard to electrophysiological studies, which play an important diagnostic role in both ALS and CSA [21, 55], Park et al. [43] confirmed a lower ulnar-to-median nerve latency ratio and the terminal latency of the median nerve among patients with ALS ($P < 0.006$). Shindo et al. [56] also reported an increase in muscle sympathetic nerve activity (MSNA) in a patient with ALS, with a decrease in a patient with CSA. Sonoo et al. [55] reported that diffuse fasciculation potentials in the trapezius and paraspinal muscles were suggestive of ALS but not CSA. As previously stated, the ADM-to-APB ratio of the CMAP may also be a differentiating feature of ALS and CSA [51, 57], with a higher ulnar-to-median nerve ratio for ALS than CSA ($P < 0.0001$) [25]. Hatanaka et al. [58] reported that the repetitive nerve stimulation (RNS) test for the trapezius muscle had a specificity of 100% and sensitivity of 78% for ALS, but not for CSA. Zheng et al. [59] reported that decreases in the CMAP with RNS of the trapezius muscles were useful to differentiate CSA from ALS. In the same way, abnormal spontaneous electromyographic activity of paraspinal muscles, associated with three or more thoracic segments, from T7 to T10, was useful in differentiating ALS from CSA [52].

### Rotator cuff tear

Proximal-type CSA, which includes impairment in shoulder elevation, is likely to be misdiagnosed as a rotator cuff tear. Rotator cuff tears are typically diagnosed based on physical findings, disease course and imaging [34]. Assessment of weakness of the biceps and supinator muscles (which is typical in addition to the deltoid in proximal-type CSA) can help to differentiate a rotator cuff tear from CSA [21, 46]. The shoulder provocation test can also be useful to differentiate a rotator cuff tear from CSA [34].

### Hirayama disease

HD, also known as juvenile muscular atrophy of the distal upper extremity, was first reported by Hirayama et al. [60], as a benign motor neuron disease characterized by unilateral atrophy of the muscles of the forearm innervated by the ulnar nerve, as well as atrophy of the intrinsic muscles of the hand, including the dorsal and palmar interossei, thenar and hypothenar muscles and the extensor and flexor digitorum communis muscles [61]. HD primarily affects males, with a male-to-female ratio of 20:1, between the ages of 15 and 25 years, compared to a typical onset of CSA in middle age and elderly individuals [25]. The clinical features of HD are similar to those of distal-type CSA [25]. In fact, Hashiguchi et al. [13] did report one case of HD who was diagnosed with CSA 20 years later. MR imaging in neck flexion is useful to differentiate between CSA and HD [25]. The ratio of the ulnar-to-median CMAP amplitude can further assist in differentiating HD from CSA, with the ratio being lower among patients with HD than CSA ($0.55 \pm 0.41\%$; $P < 0.001$) [25]. As well, the CMAP amplitude of the APB is lower

among patients with distal-type CSA than those with HD ($P = 0.004$), while the ADM-to-APB ratio is significantly lower among patients with HD ($P < 0.001$) [45].

## Natural history

CSA is characterized by unilateral (and sometimes bilateral) severe motor weakness and wasting of muscles of the upper extremities that does not progress beyond a few myotomes. Sensory loss and pyramidal signs are typically absent or clinically insignificant. Of note, CSA follows a self-limited course, with the symptoms typically stabilizing for years after an initial progressive course.

## Conservative treatment

It is vital that conservative treatment be the first line of intervention for CSA, particularly in the early stage of the disease and include cervical traction, immobilization of the neck, using a collar, and physical therapy, with administration of vitamin B12 or E also having been shown to be effective for some patients [14, 18, 32, 39, 62]. Shibuya et al. [26] also reported a recovery in motor function in three patients with CSA after administration of prostaglandin E1. Tofuku et al. [3] reported marked improvement in muscle strength (measured by manual muscle testing) with hyperbaric oxygen (HBO) therapy, with a mean increase in strength from 1.9/5 before treatment to 4.4/5 after HBO treatment. Starting with conservative treatment for a few months allows for monitoring of progression in symptoms, which is more likely for a progressive neurological disorder, such as ALS, and less likely for CSA.

## Surgical treatment

Once the diagnosis of CSA is confirmed, surgical treatment can be useful to halt disease progression [9]. However, there is currently no consensus as to the best surgical approach for the treatment of CSA, with both anterior cervical decompression and fusion or posterior decompression (laminoplasty, with or without foraminotomy) having been used [1, 18, 19, 23, 39, 40, 62]. In addition, complications of anterior or posterior decompression for CSA are similar to those experienced with surgical management of cervical degenerative disease and include instrument failure, graft extrusion, pseudoarthrosis, subsidence, C5 nerve palsy, axial pain, fusion failure and a high rate of adjacent segment disease [20, 64, 70–72]. In order to reduce complications, it is important to choose an appropriate individualized treatment as to maximize the chance of optimal neurological recovery, while minimizing the risk of complications [70]. Anterior decompression is effective for the treatment of most patients with CSA [18, 19, 23, 41, 64, 65], with an improvement rate of 66.7% having been reported at a mean follow-up period of 46.5 months [16]. Although anterior decompression provides optimal neurological recovery, this surgical approach is often being used in patients with one or two compressive lesions [16, 17, 22]. Recently, it has been reported that posterior decompression is also effective, particularly for patients with three or more compressive lesions [20, 39, 41]. Interestingly, most Japanese studies have identified laminoplasty as the treatment of choice for patients with distal-type CSA [16]. Terai et al. [66] reported resolution of the muscle weakness at 6 months post-surgery, in three patients with CSA treated using a Tandem keyhole foraminotomy. Dorsen et al. [67] reported on the effectiveness of decompressive laminectomy and foraminotomy in improving muscle strength and bulk among patients with CSA, 4–12 years after surgery. Fujiwara et al. [22] reported on the effectiveness of laminoplasty and foraminotomy in most patients with CSA, with good-to-excellent results achieved in 78% of cases. Sasai et al. [8] reported that microsurgical posterior foraminotomy, combined with laminoplasty, is a beneficial treatment option, improving deltoid muscle strength in approximately 80% of cases. In fact, one study reported better outcomes with foraminotomy and laminoplasty than anterior decompression among patients with multi-segmental stenosis [8]. In their case series of 28 patients with CSA, Takebayashi et al. [20] reported excellent outcomes in 18 cases using selective laminoplasty combined with foraminotomy, with good outcomes achieved in nine cases. Baxter et al. [62] reported on the use of nerve transfers as an alternative surgical treatment to restore shoulder function in patients with proximal-type CSA. Kumar et al. [32] reported that multi-muscle nerve transfer improved overall shoulder function, achieving a mean abduction range of motion of 91°, with 111° of shoulder flexion, 23° of external rotation and 110° of elbow flexion.

## Timing of surgery

Tauchi et al. [21] proposed that surgical treatment should be considered after 4 months from the onset CSA symptoms, in the absence of benefits of conservative treatment. This was supported by Jiang et al. [40] who recommended waiting at least 2–3 months after symptom onset, considering that a large number of patients show recovery of muscular weakness with conservative treatment over time. Tauchi et al. [54] recommended early surgical treatment for patients with CSA who do not respond to conservative treatment, as well as for those with severe preoperative muscle weakness and those with distal-type CSA. A decrease of 30–50% in the CMAP amplitude of the deltoid and biceps muscles has also been suggested as an indication for surgical treatment [27]. Of note, Meng et al. [63] recommended surgery as the first line

of treatment for patients with proximal-type CSA, especially those with serious spinal cord compression.

### Prognosis and factors associated with a poor outcome

Outcomes of surgical treatment have been reported to be worse for older than younger patients [18, 39], as well as for patients with lower manual muscle grades before surgery [21, 54, 62], multi-segmental compression [7, 39] and a longer duration of symptoms [17, 18, 21, 39, 54, 62, 64]. Treatment outcomes are usually better for proximal- than distal-type CSA [12, 17, 22, 23, 40, 41, 54, 63]. There are several possible reasons why outcomes are better for proximal- than distal-type CSA. Foremost, the distal-type CSA is usually associated with AH injury, with the AH having a lower capacity for regeneration than the VNR, the latter being more commonly implicated in proximal-type CSA [12, 17, 22]. Secondly, the average duration of symptoms tends to be longer among patients with distal- than proximal-type CSA, with a longer duration of symptoms being associated with worse outcomes [17, 22]. Lastly, AH involvement, with or without VNR compression, is a risk factor for poor surgical outcomes [7, 17, 19, 22, 39, 64]. Although outcomes are better for proximal- than distal-type CSA, Ebara et al. [68] reported outcomes for distal- than diffuse-type CSA. Moreover, a CMAP amplitude > 10 mV on the normal side and a CMAP amplitude > 50% on the affected side are typically associated with full recovery of deltoid and biceps function, despite severe preoperative weakness [7], with fair outcomes reported for patients with a CMAP amplitude < 30% of the normal side [27, 62].

The presence of a high signal intensity change on T2-weighted MR imaging can be used as prognostic factors for patients with CSA [11, 18], although the findings of Tauchi et al. [54] do not support this conclusion. Iizuka et al. [11] proposed that pyramidal tract signs could be used as a prognostic factor, although Inui et al. [39] reported that pyramidal tract signs did not have any prognostic value, either among patients treated conservatively or those treated surgically.

## Conclusion

Selective damage to the VNR, AH or both has been associated with CSA. Clinically, CSA is characterized by weakness and atrophy of proximal (scapular, deltoid and biceps) or distal (forearm and hand) upper extremity muscles, with no sensory deficits. Conservative treatment should be the first line of choice, with surgical treatment considered after 4 months of non-response. Distal-type CSA, a longer symptom duration, older patient age and greater preoperative muscle weakness are factors associated with poor outcomes. Although the disease process of CSA is normally self-limited, the treatment of CSA remains a challenge for spinal surgeons. Future studies should focus on clarifying the pathogenesis of CSA and providing strong evidence of diagnostic methods.

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### Compliance with ethical standards

**Ethical approval** This article does not contain any studies with human participants or animals performed by any of the authors.

**Conflict of interest** The authors had no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Informed consent** Informed consent was obtained from all individual participants included in the study.

## References

1. Kaneko K, Taguchi T, Toyoda K, Kato Y, Azuma Y, Kawai S (2004) Distal-type cervical spondylotic amyotrophy: assessment of pathophysiology from radiological findings on magnetic resonance imaging and epidurally recorded spinal cord responses. Spine (Phila Pa 1976) 29(9):E185–E188
2. Yanagi T, Kato H, Sobue I (1976) Clinical characteristics of cervical spondylotic amyotrophy. Rinsho shinkeigaku = Clinical neurology 16(7):520–528
3. Tofuku K, Koga H, Yone K, Komiya S (2011) Conservative treatment with hyperbaric oxygen therapy for cervical spondylotic amyotrophy. Spinal Cord 49(6):749–753
4. Brain WR, Northfield D, Wilkinson M (1952) The neurological manifestations of cervical spondylosis. Brain 75(2):187–225
5. Keegan JJ (1965) The cause of dissociated motor loss in the upper extremity with cervical spondylosis. J Neurosurg 23(5):528
6. Sobue I, Kato H, Yanagi T (1975) Clinical characteristics and classification of cervical spondylotic myelopathy. Rinsho Seikeigaku 10:999–1006 **(in Japanese)**
7. Imajo Y, Kato Y, Kanchiku T, Suzuki H, Taguchi T (2011) Pathology and prognosis of proximal-type cervical spondylotic amyotrophy: new assessment using compound muscle action potentials of deltoid and biceps brachii muscles. Spine (Phila Pa 1976) 36(7):E476–E481
8. Sasai K, Umeda M, Saito T, Ohnari H, Wakabayashi E, Iida H (2006) Microsurgical posterior foraminotomy with laminoplasty for cervical spondylotic radiculomyelopathy including cervical spondylotic amyotrophy. J Neurosurg Spine 5(2):126–132
9. Funaba M, Kanchiku T, Imajo Y et al (2017) A novel scoring system associated with surgical outcome of distal-type cervical spondylotic amyotrophy. Clin Spine Surg 30(9):E1182–E1189
10. Fujiwara K (2001) Cervical spondylotic amyotrophy with intramedullary cavity formation. Spine (Phila Pa 1976) 26(10):E220–E222
11. Iizuka Y, Iizuka H, Mieda T et al (2014) Prognostic factors for cervical spondylotic amyotrophy: are signs of spinal cord

involvement associated with the neurological prognosis? Spinal Cord 52(5):364–367
12. Zhang JT, Yang DL, Shen Y, Zhang YZ, Wang LF, Ding WY (2012) Anterior decompression in the management of unilateral cervical spondylotic amyotrophy. Orthopedics 35(12):e1792–e1797
13. Hashiguchi S, Ogasawara N, Watanabe A, Kawachi Y, Miki N (1997) Cervical spondylotic amyotrophy associated with Hirayama's disease. Intern Med (Tokyo, Japan) 36(9):647–650
14. Kameyama T, Ando T, Yanagi T, Yasui K, Sobue G (1998) Cervical spondylotic amyotrophy. Magnetic resonance imaging demonstration of intrinsic cord pathology. Spine (Phila Pa 1976) 23(4):448–452
15. Asaka T, Satake R, Takamori M, Matsushima A (1995) Dissociated motor loss syndrome with cavities in the anterior horns. Intern Med 34(10):1027–1029
16. Srinivasa Rao NV, Rajshekhar V (2009) Distal-type cervical spondylotic amyotrophy: incidence and outcome after central corpectomy. J Neurosurg Spine 10(4):374–379
17. Zhang J, Cui C, Liu Z, Tong T, Niu R, Shen Y (2016) Predisposing factors for poor outcome of surgery for cervical spondylotic amyotrophy: a multivariate analysis. Sci Rep 6:39512
18. Mori K, Yamamoto T, Nakao Y, Maeda M (2006) Cervical spondylotic amyotrophy treated by anterior decompression. Three case reports. Neurol Med Chir (Tokyo) 46(7):366–370
19. Shinomiya K, Komori H, Matsuoka T, Mutoh N, Furuya K (1994) Neuroradiologic and electrophysiologic assessment of cervical spondylotic amyotrophy. Spine (Phila Pa 1976) 19(1):21–25
20. Takebayashi T, Yoshimoto M, Ida K, Tsuda H, Terashima Y, Yamashita T (2013) Minimum invasive posterior decompression for cervical spondylotic amyotrophy. J Orthop Sci 18(2):205–207
21. Tauchi R, Imagama S, Inoh H et al (2015) Appropriate timing of surgical intervention for the proximal type of cervical spondylotic amyotrophy. Eur J Orthop Surg Traumatol 25(Suppl 1):S107–S113
22. Fujiwara Y, Tanaka N, Fujimoto Y, Nakanishi K, Kamei N, Ochi M (2006) Surgical outcome of posterior decompression for cervical spondylosis with unilateral upper extremity amyotrophy. Spine (Phila Pa 1976) 31(20):E728–E732
23. Wang HL, Li HC, Jiang JY, Lu FZ, Chen WJ, Ma XS (2014) Evaluation of characteristics and surgical outcomes in cervical spondylotic amyotrophy. Indian J Orthop 48(5):511–517
24. Tauchi R, Imagama S, Inoh H et al (2014) Characteristics and surgical results of the distal type of cervical spondylotic amyotrophy. J Neurosurg Spine 21(3):411–416
25. Jin X, Jiang JY, Lu FZ, Xia XL, Wang LX, Zheng CJ (2014) Electrophysiological differences between Hirayama disease, amyotrophic lateral sclerosis and cervical spondylotic amyotrophy. BMC Musculoskelet Disord 15:349
26. Shibuya R, Yonenobu K, Yamamoto K et al (2005) Acute arm paresis with cervical spondylosis: three case reports. Surg Neurol 63(3):220–228
27. Imajo Y, Kato Y, Kanchiku T et al (2012) Prediction of surgical outcome for proximal-type cervical spondylotic amyotrophy novel mode of assessment using compound action potentials of deltoid and biceps brachii and central motor conduction time. Spine (Phila Pa 1976) 37(23):E1444–E1449
28. Ahdab R, Creange A, Benaderette S, Lefaucheur JP (2009) Cervical spondylotic amyotrophy presenting as dropped head syndrome. Clin Neurol Neurosurg 111(10):874–876
29. Hatanaka Y, Chiba T, Okuma H, Kanbayashi T, Kuwabara M, Higashihara M (2017) Diagnostic utility of repetitive nerve stimulation test of the trapezius muscle in amyotrophic lateral sclerosis. Clin Neurophysiol 128(5):823–829
30. Nouri A, Tetreault L, Singh A, Karadimas SK, Fehlings MG (2015) Degenerative cervical myelopathy: epidemiology, genetics, and pathogenesis. Spine (Phila Pa 1976) 40(12):E675–E693
31. Mizuno J, Nakagawa H, Hashizume Y (2002) Cervical amyotrophy caused by hypertrophy of the posterior longitudinal ligament. Spinal Cord 40(9):484–488
32. Gebere-Michael SG, Johnston JC, Metaferia GZ, Wuhib MZ, Fernandez HH (2010) Bilaterally symmetric cervical spondylotic amyotrophy: a novel presentation and review of the literature. J Neurol Sci 290(1–2):142–145
33. Kumar KK, Doi K, Hattori Y, Sakamoto S, Yonemura H, Montales T (2014) Reconstruction of shoulder and elbow function using multiple muscle transfers for cervical spondylotic amyotrophy. Spine (Phila Pa 1976) 39(21):E1269–E1275
34. Iwata E, Shigematsu H, Inoue K, Egawa T, Sakamoto Y, Tanaka Y (2017) Muscle weakness in the empty and full can tests cannot differentiate rotator cuff tear from cervical spondylotic amyotrophy: pain provocation is a useful finding. Open Orthop J 11:1081–1086
35. Sasaki S, Iwata M (1999) Atypical form of amyotrophic lateral sclerosis. J Neurol Neurosurg Psychiatry 66(5):581–585
36. Koda M, Furuya T, Rokkaku T et al (2017) Drop finger caused by 8th cervical nerve root impairment: a clinical case series. Eur Spine J 26(4):1096–1100
37. Badar F, Azfar SF, Ahmad I, Kirmai S (2011) MRI feature of cervical spondylotic amyotrophy: snake eye appearance. JK Science 14(2):107
38. Zhang ZF, Wang HG (2014) Is the "snake-eye" MRI sign correlated to anterior spinal artery occlusion on CT angiography in cervical spondylotic myelopathy and amyotrophy? Eur Spine J 23(7):1541–1547
39. Inui Y, Miyamoto H, Sumi M, Uno K (2011) Clinical outcomes and predictive factors relating to prognosis of conservative and surgical treatments for cervical spondylotic amyotrophy. Spine (Phila Pa 1976) 36(10):794–799
40. Jiang SD, Jiang LS, Dai LY (2011) Cervical spondylotic amyotrophy. Eur Spine J 20(3):351–357
41. Uchida K, Nakajima H, Yayama T et al (2009) Anterior and posterior decompressive surgery for progressive amyotrophy associated with cervical spondylosis: a retrospective study of 51 patients. J Neurosurg Spine 11(3):330–337
42. Shibuya K, Misawa S, Nasu S et al (2013) Split hand syndrome in amyotrophic lateral sclerosis: different excitability changes in the thenar and hypothenar motor axons. J Neurol Neurosurg Psychiatry 84(9):969–972
43. Park D, Park JS (2017) Terminal latency abnormality in amyotrophic lateral sclerosis without split hand syndrome. Neurol Sci 38(5):775–781
44. Stark RJ, Kennard C, Swash M (1981) Hand wasting in spondylotic high cord compression: an electromyographic study. Ann Neurol 9(1):58–62
45. Fang J, Liu MS, Guan YZ et al (2016) Pattern differences of small hand muscle atrophy in amyotrophic lateral sclerosis and mimic disorders. Chin Med J (Engl) 129(7):792–798
46. Iwata E, Shigematsu H, Inoue K et al (2018) Biceps-related physical findings are useful to prevent misdiagnosis of cervical spondylotic amyotrophy as a rotator cuff tear. Asian Spine J12(1):69–73
47. Miller JD, Pruitt S, McDonald TJ (2000) Acute brachial plexus neuritis: an uncommon cause of shoulder pain. Am Fam Physician 62(9):2067–2072
48. Rowland LP (1998) Diagnosis of amyotrophic lateral sclerosis. J Neurol Sci 160(Suppl 1):S6–S24
49. Yamada M, Furukawa Y, Hirohata M (2003) Amyotrophic lateral sclerosis: frequent complications by cervical spondylosis. J Orthop Sci 8(6):878–881
50. Ghasemi M (2016) Amyotrophic lateral sclerosis mimic syndromes. Iran J Neurol 15(2):85–91

51. Kuwabara S, Mizobuchi K, Ogawara K, Hattori T (1999) Dissociated small hand muscle involvement in amyotrophic lateral sclerosis detected by motor unit number estimates. Muscle Nerve 22(7):870–873
52. Kuncl RW, Cornblath DR, Griffin JW (1988) Assessment of thoracic paraspinal muscles in the diagnosis of ALS. Muscle Nerve 11(5):484–492
53. Ellenberg MR, Honet JC, Treanor WJ (1994) Cervical radiculopathy. Arch Phys Med Rehabil 75(3):342–352
54. Tauchi R, Imagama S, Inoh H et al (2013) Risk factors for a poor outcome following surgical treatment of cervical spondylotic amyotrophy: a multicenter study. Eur Spine J 22(1):156–161
55. Sonoo M, Kuwabara S, Shimizu T et al (2009) Utility of trapezius EMG gor diagnosis of amyotrophic lateral sclerosis. Muscle Nerve 39(1):63–70
56. Shindo K, Watanabe H, Tanaka H, Nagasaka T, Tsunoda S, Shiozawa Z (2002) A comparison of sympathetic outflow to muscles between cervical spondylotic amyotrophy and ALS. Amyotroph Lateral Scler Other Motor Neuron Disord 3(4):233–238
57. Jiang M, Yan X, Yan LR, Zhan YB, Hu HT (2017) Value of split hand in the differential diagnosis of amyotrophic lateral sclerosis and cervical spondylotic amyotrophy. Zhonghua Yi Xue Za Zhi 97(47):3729–3732
58. Hatanaka Y, Higashihara M, Chiba T, Miyaji Y, Kawamura Y, Sonoo M (2017) Utility of repetitive nerve stimulation test for ALS diagnosis. Clin Neurophysiol 128(5):823–829
59. Zheng C, Jin X, Zhu Y, Lu F, Jiang J, Xia X (2017) Repetitive nerve stimulation as a diagnostic aid for distinguishing cervical spondylotic amyotrophy from amyotrophic lateral sclerosis. Eur Spine J 26(7):1929–1936
60. Hirayama K, Tsubaki T, Toyokura Y, Okinaka S (1965) Juvenile muscular atrophy of unilateral upper extremity. Wakayama Med Rep 9(3):161–167
61. Okumura H, Homma TT (1994) Juvenile compression myelopathy in the cervical spine. Spine (Phila Pa 1976) 19(1):72–76
62. Baxter C, Miller TA, Ross DC, Doherty C (2018) Treatment of cervical spondylotic amyotrophy with nerve transfers. J Hand Surg 43(7):e1–e4
63. Meng YK, Zhang J, Yang Y et al (2017) Clinical diagnosis and management of cervical spondylotic amyotrophy. Zhonghua Yi Xue Za Zhi 97(17):1320–1323
64. Kong LD, Wang LF, Zhang JT, Zhang YZ, Ding WY, Shen Y (2015) Predictive factors relating to prognosis of anterior decompressive surgery for proximal-type cervical spondylotic amyotrophy. J Back Musculoskelet Rehabil 28(2):261–266
65. Liu XY, Li CD, Yi XD, Li H, Yu ZR (2007) Surgical treatment of cervical spondylotic amyotrophy. Zhonghua Yi Xue Za Zhi 87(47):3339–3342
66. Terai H, Suzuki A, Toyoda H et al (2014) Tandem keyhole foraminotomy in the treatment of cervical radiculopathy: retrospective review of 35 cases. J Orthop Surg Res 16(9):38
67. Dorsen M, Ehni G (1979) Cervical spondylotic radiculopathy producing motor manifestations mimicking primary muscular atrophy. Neurosurgery 5(4):427–431
68. Ebara S, Yonenobu K, Fujiwara K, Yamashita K, Ono K (1988) Myelopathy hand characterized by muscle wasting. A different type of myelopathy hand in patients with cervical spondylosis. Spine (Phila Pa 1976) 13(7):785–791
69. Querin G, Mendili MM, Lenglet T et al (2018) The spinal and cerebral profile of adult spinal-muscular atrophy: a multimodal imaging study. Neuroimage Clin. https://doi.org/10.1016/j.nicl.2018.101618
70. Visocchi M, Conforti G, Roselli R et al (2015) From less to maximally invasiveness in cervical spine surgery. Int J Surg Case Rep 9:85–88
71. Spallone A, Marchione P, Li Voti P et al (2014) Anterior cervical discectomy and fusion with "mini-invasive" harvesting of iliac crest graft versus polyetheretherketone (PEEK) cages: a retrospective outcome analysis. Int J Surg 12(12):1328–1332
72. Barbagallo GM, Certo F, Visocchi M et al (2014) Double-level cervical total disc replacement for adjacent segment disease: is it a useful treatment? Description of late onset heterotopic ossification and review of the literature. Eur Rev Med Pharmacol Sci 18(1 Suppl):15–23




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