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PERSPECTIVE

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Is it time to redefine or rename the term "Central Cord Syndrome"?

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As a Specialist in Rehabilitation Medicine, I often have patients referred by surgical colleagues with "central cord syndrome" (CCS). As clinicians working in the field of spinal cord injury (SCI), we all believe that we have the same understanding of CCS. Most or all of us have been trained in the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) and American Spinal Injuries Association (ASIA) impairment scale (AIS). So, why do we sometimes choose to use the term CCS over the neurological level of injury/AIS? And what is meaning of CCS to those who use it?

Over the past couple of decades, a general understanding developed, that patients with CCS had favourable neurological and functional outcomes. This may have influenced decisions regarding rehabilitation, such as in-patient versus out-patient programmes, length of stay for in-patients and out-patient therapy follow-up. This consensus on better outcomes was based on studies which reported that substantial proportions of patients with CCS had improvements in their neurological status (upper and lower limb motor scores and/or AIS), gains in their independence and/or mobility scores and sphincter function [1–3]. However, all used an imprecise definition of CCS e.g. spinal cord trauma with greater upper versus lower extremity weakness with sacral sparing, and none had a comparison group.

It was no surprise therefore, when 2 publications in 2010 confirmed a lack of consensus in the literature and amongst spine surgeons as to the definition of CCS [4, 5]. Arising from this, it was suggested that a difference of 10 points or more between total upper extremity and total lower extremity motor scores might be an appropriate diagnostic criterion for traumatic CCS (TCCS) [4, 5]. It was mooted that this could lead to a reliable diagnosis of traumatic CCS, vital for research purposes, given that this group of SCI patients was thought to have a more positive outcome compared with patients with other incomplete tetraplegia.

In the first paper published using this proposed new diagnostic criterion [4, 5], the cases of 142 patients with incomplete cervical SCI up to 12 months post onset, were examined [6]. These cases were categorised as either non-traumatic central cord syndrome (non-TCCS)—upper extremity motor score (UEMS) greater than or equal to lower extremity motor score (LEMS); intermediate TCCS—LEMS 1-9 points greater than UEMS; or TCCS—LEMS 10 or more points greater than UEMS. Cases were then also stratified by AIS, C or D [6]. The authors concluded that use of the AIS grade was more useful in prognostication of outcomes and recruitment into research studies than classification of an injury as CCS [6]. This was based on a number of observations including that (i) although those with TCCS had lower UEMS at onset of injury, there were no difference in UEMS by either 6 or 12 months, whether in the non-TCCS, intermediate TCCS or TCCS group (ii) there was no difference in independence scores, measured by Spinal Cord Independence Measure (SCIM) II, across the 3 incomplete cervical level injuries even though AIS D patients had higher UEMS by approximately 10 points, than AIS C patients at 6 or 12 months (iii) TCCS had higher LEMS at onset of injury than other groups and appeared to have better mobility outcomes (only the independent indoor mobility aspect of SCIM II was measured) but not when stratified into AIS C or D [6].

So, given these findings, the obvious question is whether this new definition, which does not currently appear on the ISNCSCI worksheet, would be accepted by clinicians working with affected patients, and if so, would this be reflected in subsequent research practice?

In my experience, this new definition was used by some specialist clinicians but not by all, and in general, patients continued to be referred with the traditional label of CCS. This label was communicated encompassing many of the descriptors outlined in the 2 aforementioned publications, including some/all of "neurological deficit", "upper extremities more affected than lower extremities", "cervical or lower thoracic level of injury", "bladder dysfunction", "sacral sparing: incomplete SCI", "neuroanatomy of affected structures", "injury mechanism", "pre-existent stenotic, spondylotic spinal canal" [4, 5]. The assumption regarding positive outcomes in this patient cohort also seemed to persist.

Adoption of the proposed new definition into clinical SCI research [4, 5] appeared challenging. As an example, in a 2016 publication examining differences in outcomes in patients with CCS, managed surgically or non-surgically, CCS was defined as cervical SCI producing disproportionately greater weakness in the upper limbs than lower limbs with varying degrees of sensory loss and bladder dysfunction [7]. The authors also reported that neurological examination and imaging confirmed the diagnosis [7], although there were no imaging criteria in the proposed new definition from 2010.

Another publication in 2018 may have offered some promise that the new definition was accepted into use, when it was described that patients with CCS (based on the diagnostic criterion of a difference of at least 10 points between UEMS and LEMS) had significantly better walking ability than those with Brown Séquard Plus Syndrome, and a higher rate of recovery of

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voluntary bladder control than those with other incomplete cervical level syndromes [8]. Although numbers were small (42 patients with a syndrome), perhaps there was some merit after all in the assumption of a better outcome for patients with CCS [8].

Recently, the largest study thus far (566 patients) comparing outcomes in patients with CCS and other incomplete cervical level SCI has been published, with a different conclusion to what has gone before [9]. The CCS definition of a 10 point difference between upper and lower motor extremity scores in favour of the lower extremities was used. It was found that UEMS were consistently lower and LEMS consistently higher in CCS compared with incomplete cervical SCI at onset, 6 months and 12 months [9]. However, the only notable functional difference was that patients with CCS had lower SCIM self-care scores than incomplete cervical SCI at onset, 6 & 12 months, even though CCS had a significantly greater improvement in UEMS by 1 year post injury [9]. At 6 & 12 months, there was no difference in overall independence or mobility scores based on SCIM, Walking Index for Spinal Cord Injury (WISCI), 10 metre walk test (10MWT) and 6 minute walk test (6MWT), even though incomplete cervical SCI have significantly greater improvement in LEMS at 1 year [9]. Similarly, there was no difference between the 2 groups with regard to bladder and bowel control at 1 year [9]. These findings are in contrast to previous reports of superior outcomes in patients with CCS.

So, why continue to use this term CCS when the use of the ISNCSCI/neurological level of injury and AIS are much more meaningful descriptors of injury than CCS and much more informative in the exchange of information, describing clearly a SCI as AIS C or D?

The case has been made previously for "Revisiting Central Cord Syndrome" [10]. This report focused on the approach to surgical management of CCS, while considering that there are various mechanisms of injury resulting in CCS [10]. It called into question the origins of CCS with regard to its pathophysiology, raising another issue of uncertainty around this terminology, CCS [10]. The authors raised concerns that these patients may be less likely to be considered for surgical decompression, due to long-held beliefs that there will be substantial neurological recovery and that surgical intervention may impact negatively on this. Similar concern could be raised, that patients labelled with CCS might be less likely to be referred for specialist rehabilitation at a SCI centre, due to a presumption that they will have spontaneous neurological recovery.

If a term, about which we are questioning the pathophysiology and clinical definition, is playing such a role in influencing decision-making in surgery and rehabilitation, then surely it is time to re-think its use. Perhaps, we cannot discontinue use of the term CCS, given that it is engrained in our practice vocabulary; but maybe in our clinical practice, we could accompany the term CCS with information on the neurological level of injury/AIS, while we await a more definitive plan. As for that plan, a review of the definition of CCS followed by clear dissemination of the conclusion of that review, is required so that there is a mutual understanding of the term amongst surgeons, rehabilitation physicians, therapists and researchers.

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COMPETING INTERESTS

The author declares no competing interests.

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