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PERSPECTIVE

The case for revisiting central cord syndrome

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The epidemiology of acute traumatic spinal cord injury (SCI) is changing, and so too are trends in injury phenotypes. The prototypical case of SCI was once a young adult with a spinal fracture and/or dislocation and severe resultant neurological injury from a motor vehicle collision or other high-impact trauma. However, demographic shifts in our population have dawned a new era for spinal cord injury that is marked by an ever-increasing proportion of cases comprised of older patients with cervical incomplete cord injury, often without disruption of the spinal column, due to low-energy mechanisms (e.g., falls) [1]. With the continued aging of the global population, and therefore the continued expected growth of this injury subtype, there is a critical unmet need to further understand the pathophysiology and clinical course of these injuries, and to identify treatment strategies that may mitigate disability and improve functional status in this vulnerable patient population.

Central cord syndrome (CCS) is the most common incomplete SCI subtype or "spinal cord syndrome". The first detailed account of CCS came from Schneider et al. [2] in 1954; they described a type of cervical SCI involving "disproportionately more motor impairment of the upper than of the lower extremities, bladder dysfunction, usually urinary retention, and varying degrees of sensory loss below the level of the lesion." The contemporary definition of CCS retains the core elements of this original description, with the pathognomonic feature being disproportionate weakness of the upper compared with the lower limbs [3]. More recent attempts have been made at operationalizing objective diagnostic criteria for CCS. One proposed criterion has been at least a 10-point difference between American Spinal Injury Association (ASIA) lower extremity motor score (LEMS) and upper extremity motor score (UEMS), in favor of the lower limbs (LEMS–UEMS ≥ 10) [3–5]. This definition has been helpful in allowing authors to uniformly and objectively define a patient cohort of interest for clinical research, whereas previously, studies of patients with "central cord syndrome" relied upon subjective inclusion criteria; nonetheless, this definition has yet to achieve universal acceptance. Moreover, it is recognized that CCS encompasses a spectrum of clinical presentations of incomplete cervical SCI which are not fully captured by the above criterion. CCS is hypothesized to arise from a hyperextension mechanism, particularly in the context of a spondylotic or congenitally-narrow spinal canal. Schneider et al. [6] proposed that sudden compression of the spinal cord between a hypertrophic spondylotic disc-osteophyte complex anteriorly and a buckled ligamentum flavum posteriorly causes a hematomyelic cavity to form within the central gray matter of the spinal cord; the result is disruption of the medially-placed corticospinal tract (CST) fibers that control hand and upper limb function, but relative sparing of the more laterally-placed CST fibers supplying the lower limbs [6]. Dating back to Schneider et al.'s original report, the clinical course of CCS has historically been considered favorable, regardless of intervention [2].

Nonetheless, our conceptualization of the pathophysiology and natural history of CCS has, and continues to, evolve. The original account of the etiology of disproportionate upper limb weakness in CCS was predicated upon the work of Foerster [7], who posited that the lateral CST, like the fasciculus cuneatus and gracilis and the lateral spinothalamic tract, had a somatotopic organization. Several landmark experiments have since advanced our understanding of spinal cord functional anatomy and the etiopathogenesis of CCS. First, neuroanatomic tracer [8] and Marchi degeneration [9] studies in monkeys have indicated lack of laminar (i.e., somatotopic) organization to the CST caudal to the pons, at the level of the medullary pyramids

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and the posterolateral funiculus of the spinal cord. Second, autopsy and MRI investigations have found central hemorrhage to occur rarely in CCS; instead, these have revealed diffuse white matter injury to the lateral columns [10]. Third, monkey transection experiments have demonstrated non-recovering hand dysfunction, but a striking paucity of gross locomotor deficits, following complete pyramidotomy [11]. Indeed, it is now thought that the lateral CST is critical for hand function and dexterity, but less so for locomotion, and accordingly, any injury to the CST, however diffuse, may produce a syndrome of disproportionately greater arm and hand dysfunction [11, 12]. The current understanding is therefore that the pattern of predominantly arm and hand weakness seen in CCS has little to do with selective injury to the centrally-located regions of the CST, and more to do with relative preservation of the extrapyramidal fiber tracts [12]. This brings into question the fundamental pathoanatomic basis of central cord syndrome and indeed, the origins of this very term. Also, despite previous reports of a comparatively favorable natural history, there is mounting evidence that the degree of recovery seen in patients with CCS is not substantially different from other forms of incomplete SCI [5]. From a treatment perspective, surgical intervention for CCS has historically been discouraged out of fear of derailing potential for natural recovery. However, more recent evidence would suggest that early operative management of patients with CCS, undertaken within 24 h of injury, may be safe and effective at improving long-term neurological and functional recovery [13–15].

There is a strong and well-founded impetus to critically revisit the clinical management and outcomes of central cord syndrome, and the validity and utility of its distinction as a unique clinicopathological entity.

First, the term, "central cord syndrome", itself is misleading, because it invokes a pathophysiology—namely, selective injury to the "central" areas of the spinal cord—that is incorrect. There is compelling evidence that the lateral CST does not have a somatotopic organization and CCS is marked by diffuse white matter injury to the lateral columns, not central hematomyelia within the spinal cord gray matter, as once thought.

Second, the term, "central cord syndrome", engages less proactive management strategies from many clinicians, owing to a widely-held belief that the natural history of this condition is favorable, and operative intervention might actually effectuate poorer neurological recovery. However, these views are predicated upon outdated historical reports. Indeed, the 1954 publication by Schneider et al. [2] engendered CCS with a pervasive "stigma" of sorts that has persisted over decades. It is important to recognize, however, that surgical outcomes for SCI have improved with advances in surgical techniques (e.g., microsurgery), operative adjuncts (e.g., intraoperative neurophysiological monitoring, operative microscope), imaging technology (e.g., conventional and quantitative microstructural magnetic resonance imaging [MRI]), perioperative and critical care, systems infrastructure (e.g., specialized centers of care for SCI, transport services), and our understanding of the pathophysiology of spinal cord injury. Accordingly, many of the original concepts relating to the surgical treatment of CCS have been rendered antiquated and obsolete in the modern era. In addition to standard posterior decompression via laminectomy, Schneider et al. [2] performed a dural opening, dentate ligament resection, and transdural discectomy; this invasive operation bears little resemblance to surgery for SCI today. Moreover, natural recovery is not the rule in CCS. Baseline neurological status has been shown to be an important predictor of neurological and functional outcomes after CCS, highlighting that recovery is heterogeneous and dependent by and large on the initial severity of injury [16]. Even if the recovery profile of CCS is generally favorable, surgical decompression may be expected to work in synergy with, rather than in opposition to, this natural recovery potential, resulting in superior neurological outcomes. Certainly, it could be argued that CCS may in fact be the quintessential target for neuroprotective strategies aimed at mitigating secondary injury, such as early surgical decompression, given the low-energy mechanisms and less severe primary insult typically seen in these injuries, and the presumed ongoing secondary injury to an edematous spinal cord from compression from a spondylotic or congenitally-narrow spinal canal.

Third, the distinction of "central cord syndrome" attempts to fit injuries of disparate etiology, kinetic energy, morphology, biomechanical properties, and demographic characteristics under the same umbrella. There are at least three dissociable phenotypes of CCS: (1) cervical hyperextension injury (e.g., secondary to a fall) without spinal column disruption in the setting of preexisting central canal stenosis, usually in older patients; or (2) cervical spinal column fracture and/or dislocation, or (3) acute cervical disc herniation, both typically from high-energy trauma among younger age groups. Although the clinical phenotype of these injuries may be similar, being characterized by disproportionate upper limb weakness, these clearly represent very different patient populations. Given that CCS encompasses heterogeneous injury patterns, it is perhaps not surprising that the extent of recovery, too, is heterogeneous, and dependent upon a number of patient, injury, and imaging factors [16]. Indeed, the diagnosis of CCS does not indicate anything about patient characteristics, the morphology, severity, or mechanism of injury, or potential disruption of the spinal column, all of which directly impact clinical management and long-term prognosis.

Based on the evolution in standards of acute SCI care, we would argue in favor of the recent guidelines issued by AOSpine, which advocate for early surgical decompression for any case of traumatic SCI, including CCS, when medically feasible [14, 15]. However, it is recognized that additional high-quality clinical studies investigating CCS, and more broadly, cervical incomplete SCI, are needed. This area of research should be a key public health priority and we urge the international clinical community to embrace the opportunity to address the entity of incomplete cervical SCI, including CCS.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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