Central Cord Syndrome

New Definitions For an Old Syndrome

Einat Haber, MD

Steven Kirshblum, MD

Brittany Snider, DO

Amanda Botticello, PhD





DISCLOSURES

The Devivo Mentored Research Award 2022 provided support for some of the work showcased in this presentation.



Changing demographics

Huge variability in incidence

Changing pathophysiology

Clinical decision making - timing of surgery

_

 $\langle \langle \langle \rangle \rangle \langle \rangle$

CCS ROADMAP

EM-SCI 10-pt definition

Other quantifiable criteria

Considerations in CCS

EXISTING CCS DEFINITIONS NEW CCS DEFINITIONS

Full CCS

Unilateral CCS

Borderline CCS

Overlap with EM-SCI

Outcomes

CCS vs motorincomplete tetraplegia

Limitations

Future directions

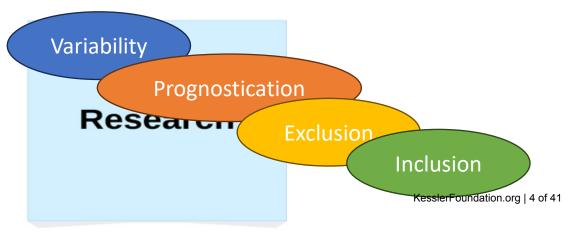
SUMMARY

CENTRAL CORD SYNDROME (CCS)

"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"

Schneider, 1954





IMPACT

Most common syndrome

Changing demographics

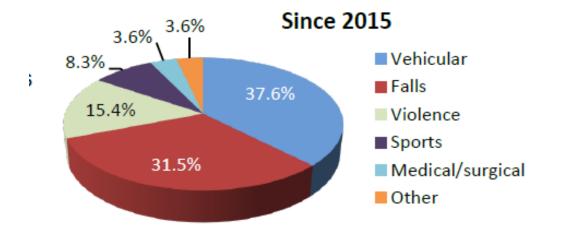
Huge variability in incidence

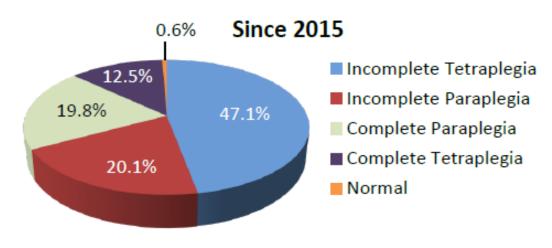
Changing pathophysiology

Clinical decision making - timing of surgery

CHANGING DEMOGRAPHICS

	Age at Injury			
Year of Injury	N	Mean	Standard Deviation	Minim
1972-1979	4,562	28.7	14.1	
1980-1984	4,949	30.5	14.7	
1985-1989	3,843	32.3	15.8	
1990-1994	3,295	33.7	16.0	
1995-1999	3,623	36.4	17.0	
2000-2004	3,443	37.6	16.7	
2005-2009	3,606	40.5	18.0	
2010-2014	2,947	42.3	18.3	
2015-2020	4,465	43.2	18.5	
Total	34,733	35.8	17.3	





AUTHORS, YEAR	PARTICIPANTS	CCS	BSS	ACS
Shrosbree, 1977	955	99 (10%)		
Bracken, 1978	133		4 (3%)	
Pickett, 2006 ²	151	49 (32%)	6 (4%)	17 (11%)
McKinley, 2007	839	77 (9%)	30 (4%)	8 (1%)
Lenehan, 2009	807	50 (6%)		
Pouw, 2011	916	97 (11%)*		
Furusawa, 2012	2,413	186 (8%)*		
Kepler, 2015	426	80 (19%)		
Thompson, 2015	831	241(29%)		
Engel-Haber, 2022 ding Central Cord Syndrome	3,639	499 (14%)*	71 (2%)	236 (6.5%)

Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist Ting and Caroline Reynolds Center for Spinal Stimulation, Kessler Foundation

AUTHORS, YEAR	PARTICIPANTS	CCS	BSS	ACS
Shrosbree, 1977	955	99 (10%)		
Bracken, 1978	133		4 (3%)	
Pickett, 2006 ²	151	49 (32%)	6 (4%)	17 (11%)
McKinley, 2007	839	77 (9%)	30 (4%)	8 (1%)
Lenehan, 2009	807	50 (6%)		
Pouw, 2011	916	97 (11%)*		
Furusawa, 2012	2,413	186 (8%)*		
Kepler, 2015	426	80 (19%)		
Thompson, 2015	831	241(29%)		
Engel-Haber, 2022	3,639	499 (14%)*	1 (2%)	236 (6.5%)

Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist Timand Caroline Reynolds Center for Spinal Stimulation, Kessler Foundation

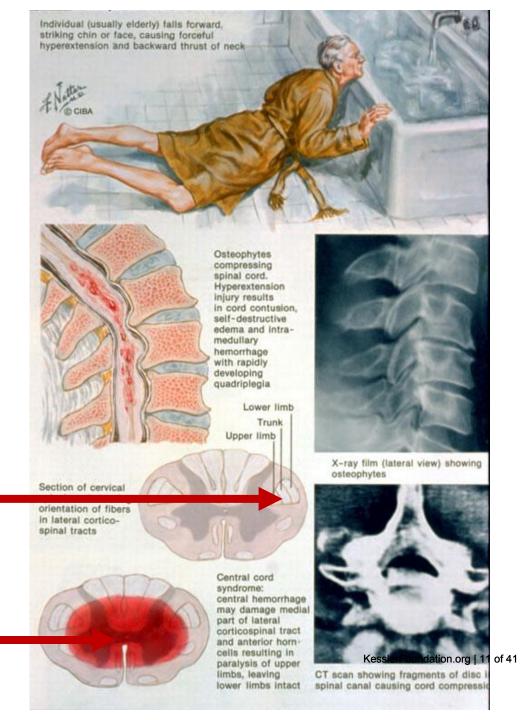
AUTHORS, YEAR	PARTICIPANTS	CCS	BSS	ACS
Shrosbree, 1977	955	99 (10%)		
Bracken, 1978	133		4 (3%)	
Pickett, 2006 ²	151	49 (32%)	6 (4%)	17 (11%)
McKinley, 2007	839	77 (9%)	30 (4%)	8 (1%)
Lenehan, 2009	807	50 (6%)		
Pouw, 2011	916	97 (11%)*		
Furusawa, 2012	2,413	186 (8%)*		
Kepler, 2015	426	80 (19%)		
Thompson, 2015	831	241(29%)		
Engel-Haber, 2022	3,639	499 (14%)*	71 (2%)	236 (6.5%)

Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist Timand Caroline Reynolds Center for Spinal Stimulation, Kessler Foundation

INCIDENCE - INCOMPLETE TETRAPLEGIA^a

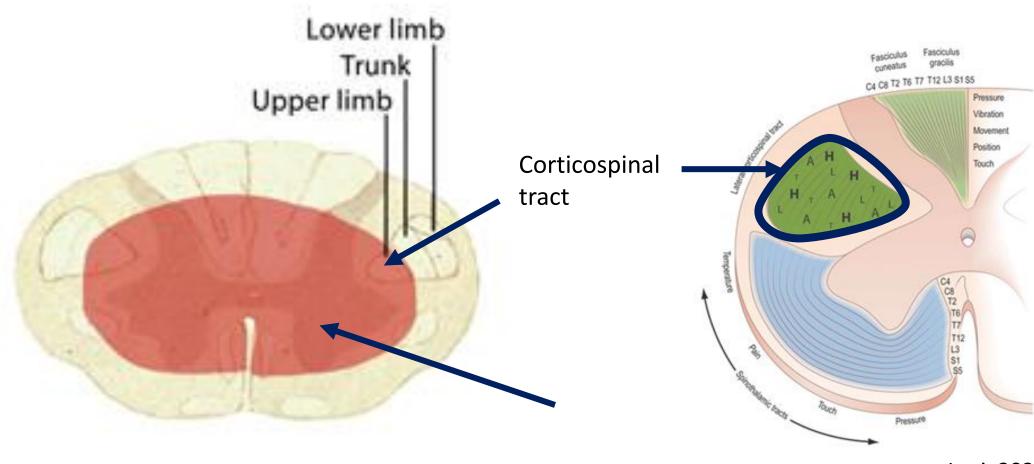
AUTHORS, YEAR	PARTICIPANTS	CCS	BSS	ACS
Bosch, 1971	60	42 (70%)	5 (8%)	12 (20%)
Bohlman, 1979	85	15 (18%)	8 (9%)	51 (60%)
Pollard, 2003	412	97 (24%)	66 (16%)	190 (46%)
Pouw, 2010	228		52 (23%) [‡]	
Pouw, 2011	248	97 (39%)*		
Badhiwala, 2020	801	185 (23%)*		
Engel-Haber, 2022	1,649	499 (30%)*	55 (3%)	169 (10%)

CHANGING PATHOPHYSIOLOGY



Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist Tim and Caroline Reynolds Center for Spinal Stimulation, Kessler Foundation

CHANGING PATHOPHYSIOLOGY



Levi, 2022

TIMING OF SURGERY - DEBATE

"Natural history is favorable. Operative intervention leads to poorer neurological recovery." Schneider, 1954

"Early operative management (<24 h) Improves neurological and functional recovery." Fehlings, 2012

"Early surgical decompression (<12 h) does not result in statistically significant or clinically meaningful neurological improvement." Hosman, 2023



Changing demographics

Huge variability in incidence

Changing pathophysiology

Clinical decision making - timing of surgery

_

 \Diamond

CCS ROADMAP

EM-SCI 10-pt definition

Other quantifiable criteria

Considerations in CCS

EXISTING CCS DEFINITIONS NEW CCS DEFINITIONS

Full CCS

Unilateral CCS

Borderline CCS

Overlap with EM-SCI

Outcomes

CCS vs motorincomplete tetraplegia

Limitations

Future directions

SUMMARY

EXISTING CCS DEFINITIONS^a

EM-SCI:

LEMS – UEMS ≥ 10^b

^aEngel-Haber, 2023 ^bvan Middendorp, 2010;

AUTHORS, YEAR	PARTICIPANTS	CCS	BSS	ACS
Shrosbree, 1977	955	99 (10%)		
Bracken, 1978	133		4 (3%)	
Pickett, 2006 ²	151	49 (32%)	6 (4%)	17 (11%)
McKinley, 2007	839	77 (9%)	30 (4%)	8 (1%)
Lenehan, 2009	807	50 (6%)		
Pouw, 2011	916	97 (11%)*		
Furusawa, 2012	2,413	186 (8%)*		
Kepler, 2015	426	80 (19%)		
Thompson, 2015	831	241(29%)		
Engel-Haber, 2022	3,639	499 (14%)*	71 (2%)	236 (6.5%) Kess

Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist Timand Caroline Reynolds Center for Spinal Stimulation, Kessler Foundation

Engel-Haber, 2022

KesslerFoundation.org | 16 of 41

EXISTING CCS DEFINITIONS a

EM-SCI:

LEMS – UEMS ≥ 10^b

also: LEMS – UEMS $\geq 1^{c}$, 5^{d} or 19^{e}

^aEngel-Haber, 2023 ^bvan Middendorp, 2010;

^cWaters, 1996; ^dBadhiwala, 2022; ^eWirz, 2010;

EM-SCI: LEMS - UEMS ≥ 10

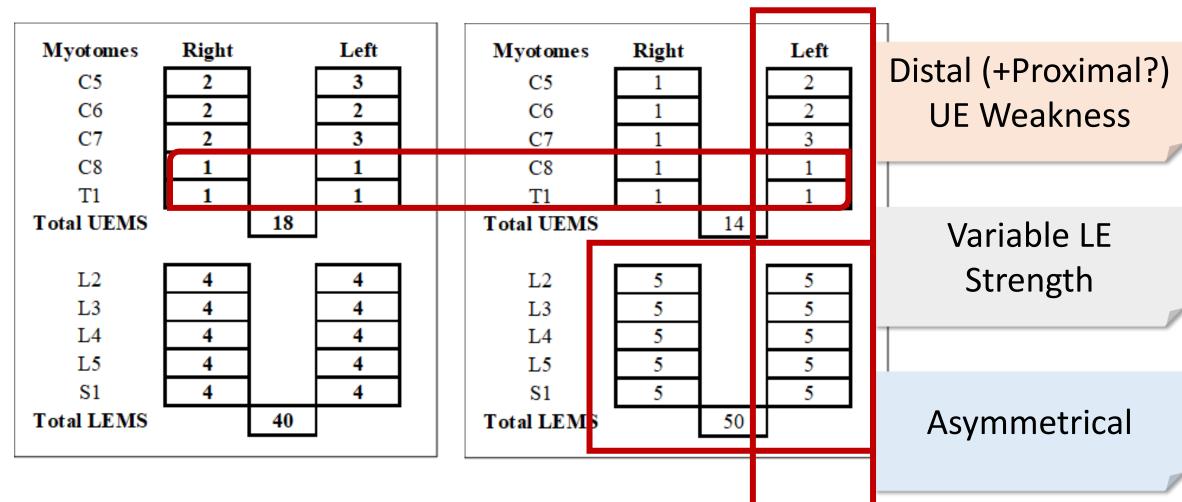
Myotomes	Right		Left
C5	2		3
C6	2		2
C7	2		3
C8	1		1
T1	1		1
Total UEMS		18	
L2	4		4
L3	4		4
L4	4		4
L5	4		4
S1	4		4
Total LEMS	_	40	
			•

Right		Left
1		2
1		2
1		3
1		1
1		1
	14	
5		5
5		5
5		5
5		5
5		5
	50	
	5 5 5	5 5 5 5 5

$$40 - 18 > 10$$

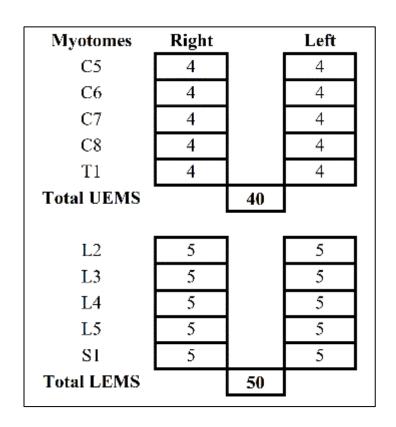
$$50 - 14 > 10$$

EM-SCI: LEMS - UEMS ≥ 10



EM-SCI: LEMS - UEMS ≥ 10

Myotomes	Right		Left
C5	0		0
C6	0		0
C7	0		0
C8	0		0
T1	0		0
Total UEMS		0	
		_	
L2	0		3
L3	0		3
L4	0		3
L5	0		3
S1	0		3
Total LEMS		15	
			•



Significal Asymmet

CCS?!

d CCS Case

Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist

KesslerFoundation.org | 20 of 41

ADDITIONAL CCS DEFINITIONS

- "Clinical impression of CCS"a,b
- CCS described on a continuum^c:

$$\left(1 - \frac{aUEMS\ below\ NLI}{aLEMS} \times 100 > 10\%\right)$$

'Central myelopathy index (CMI)' describes the same ratio^d.

^aSchroeder, 2015; ^bGuest, 2002; ^cHayes, 2000 ^dBlasetti, 2020

Consistency in Research

'CCSness' 0%-100%

ADDITIONAL CCS DEFINITIONS



Myotomes	Right		Left
C5	5		5
C6	5		5
C7	5		5
C8	0		0
T1	0		0
Total UEMS		30	
L2	2		2
L3	4		4
L4	2		2
L5	2		2
S1	2		2
Total LEMS		24	

CONSIDERATIONS IN CCSa

Distal vs Proximal UE Weakness

Asymmetrical Weakness

Disproportionate Weakness

AIS Grades Used in Definition

Imaging / Injury
Mechanism Used
in Definition

^aEngel-Haber, 2023

CONSIDERATIONS IN CCSa

Distal vs Proximal UE Weakness

Asymmetrical Weakness

"Any type of acute sensory or motor deficit localized to the cervical spinal cord from a traumatic event in the **absence of fracture or dislocation**". Avila, 2021

AIS Grades Used in Definition

Imaging / Injury
Mechanism Used
in Definition

Engel-Haber, 2023

"Radiographic and/or clinical presence of a cervical SCI without ongoing compression, which most often occurred after acute trauma in the setting of pre-existing spondylosis and a narrowed canal". Lessing, 2020 KesslerFoundation.org [24 of 41]

SUMMARY THUS FAR...

- The clinical diagnosis is deeply embedded in the clinical field
- Significant variation observed
- We support previous calls to revise the definition of CCS^{a,b}

^aSmith, 2021; ^bBadhiwala, 2020



Changing demographics

Huge variability in incidence

Changing pathophysiology

Clinical decision making - timing of surgery

_

 \Diamond

CCS ROADMAP

EM-SCI 10-pt definition

Other quantifiable criteria

Considerations in CCS

EXISTING CCS DEFINITIONS NEW CCS DEFINITIONS

Full CCS

Unilateral CCS

Borderline CCS

Overlap with EM-SCI

Outcomes

CCS vs motorincomplete tetraplegia

Limitations

Future directions

SUMMARY

OBJECTIVES & METHODS

"Defining New, Quantifiable Criteria for Central Cord Syndrome"

- Objectives:
 - 1. Define different clinical variations of CCS
 - 2. Assess frequency, overlap with existing criteria
 - 3. Compare outcomes

OBJECTIVES & METHODS

"Defining New, Quantifiable Criteria for Central Cord Syndrome"

- Objectives:
 - 1. Define different clinical variations of CCS
 - 2. Assess frequency, overlap with existing criteria
 - 3. Compare outcomes
- SCIMS database (2010-2020)
- N=1,490 individuals with motor incomplete tetraplegia (=Cervical with AIS C or D)
- Statistical analysis: descriptive, comparative (Chi-square, ANOVA)

PROPOSED CCS SUBSETS

- Distal UE weakness
- Extent of symmetry

Туре	Definition
	(avg right LE — avg right distal UE) ≥ 2
Full (bilateral) CCS	AND
•	(avg left LE — avg left distal UE) ≥ 2
	(avg right LE — avg right distal UE) ≥ 2
Unilateral CCS	OR
	(avg left LE — avg left distal UE) ≥ 2
	1 ≤ (avg right LE — avg right distal UE) < 2
Borderline CCS	AND
	1 ≤ (avg left LE — avg left distal UE) < 2

PROPOSED CCS SUBSETS

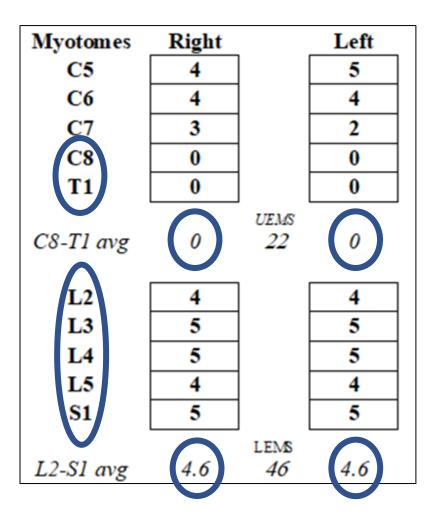
- Distal UE weakness
- Extent of symmetry

Туре	Definition
	(avg right LE — avg right distal UE) ≥ 2
Full (bilateral) CCS	AND
,	(avg left LE — avg left distal UE) ≥ 2
	(avg right LE — avg right distal UE) ≥ 2
Unilateral CCS	OR
	(avg left LE — avg left distal UE) ≥ 2
	1 ≤ (avg right LE — avg right distal UE) < 2
Borderline CCS	AND
	1 ≤ (avg left LE – avg left distal UE) < 2

FULL CCS

(avg right LE – avg right distal UE) ≥ 2 AND

(avg left LE – avg left distal UE) ≥ 2



UNILATERAL CCS

(avg right LE – avg right distal UE) ≥ 2 OR

(avg left LE – avg left distal UE) ≥ 2

Right		Left
0] [5
0] [5
0] [5
1	ĺ	5
0	ĺ	5
0.5	UEMS	
0.5	26	5
4] [5
	1 1	5
4] [5
4		5
4] [5
	LEMS	
4.2	46	5
	0 0 0 1 0 0.5	0 0 0 1 0 0.5 UEMS 26

Myotomes	Right		Left
C5	2] [0
C6	2		0
C 7	2] [0
C8	2	l	0
T1	2	ĺ	0
•		<i>UEM</i> S	
C8-T1 avg	2	10	0
L2	4		1
L3	5		0
L4	5		0
L5	5]	0
S1	5] [0
		LEMS	
L2-S1 avg	4.8	25	0.2

CCS?!

B O R D E R L I N E C C S

1 ≤ (avg right LE – avg right distal UE) < 2 AND

1 ≤ (avg left LE – avg left distal UE) < 2

Myotomes	Right		Left
C5	4		4
C6	4		4
C7	4] [4
C8	4		4
T1	4		4
C8-T1 avg	4	<i>UEM</i> S 40	4
L2	5] [5
L3	5]	5
L4	5]	5
L5	5] [5
S1	5]	5
L2-S1 avg	5	LEMS 50	5

Myotomes	Right		Left
C5	0		1
C6	0		0
C 7	1		0
C8	0		0
T1	0		0
		UEMS	
C8-T1 avg	0.2	2	0.2/
		1	
L2	1		1
L2 L3	1 2		1 2
			\vdash
L3	2		2
L3 L4	2		2
L3 L4 L5	2 1 1	LEMS	2 1 1

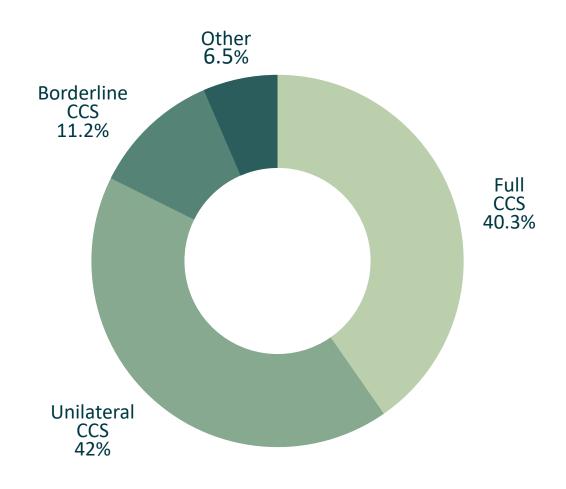
RESULTS

- N=1490 with motor incomplete tetraplegia
- 52% had some variant of CCS:
 - 17.5% with full CCS
 - 25.6% with unilateral CCS
 - 9% with borderline CCS

OVERLAP WITH EM-SCI

• N=582 (39%) fulfilled the EM-SCI criteria (LEMS – UEMS ≥ 10)

- Incomplete coverage. Not captured by the EM-SCI criteria:
 - 10% of full CCS
 - 36% of unilateral CCS
 - 52% of borderline CCS



FULL CCS

NON-CCS

Characteristics Older More falls

AIS D

80% on admission 98.5% on 1-y

AIS D

52% on admission 85% on 1-y

UEMS

Lower on admission Recovered by 1-y

LEMS

Lower on admission Still low on 1-y

FULL CCS

NON-CCS

Characteristics
Older
More falls

AIS D

80% on admission 98.5% on 1-y

AIS D

52% on admission 85% on 1-y Functional Differences?

Defining and Decoding Central Cord Syndrome Einat Haber, MD, Associate Research scientist UEMS

Lower on admission Recovered by 1-y

LEMS

Lower on admission Still low on 1-y

Tim and Caroline Reynolds Center for Spinal Stimulation, Kessler Foundation Einat Haber, MD, Associate Research scientist

Tim and Caroline Reynolds Center for Spinal Stimulation

KesslerFoundation.org | 37 of 41



Changing demographics

Huge variability in incidence

Changing pathophysiology

Clinical decision making - timing of surgery

_

 \Diamond

CCS ROADMAP

EM-SCI 10-pt definition

Other quantifiable criteria

Considerations in CCS

EXISTING CCS DEFINITIONS NEW CCS DEFINITIONS

Full CCS

Unilateral CCS

Borderline CCS

Overlap with EM-SCI

Outcomes

CCS vs motorincomplete tetraplegia

Limitations

Future directions

SUMMARY

DISCUSSION & CONCLUSIONS

- Is it really a syndrome?

In motor incomplete tetraplegia:

- 52% with any CCS (17.5% full CCS)
- 66% with UEMS
 1!)

DISCUSSION & CONCLUSIONS

- Limitations:
 - 1-year functional data not available on SCIMS
 - Still significant variation
- No clear reason to exclude CCS from studies
- Clinical & research benefits
- Needs review and validation



THANK YOU