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Introduction

- In the past two decades, interest in CP has increased remarkably particularly for spastic CP
- Assessment and treatment of patients with dystonic CP is still underreported compared with spastic CP
- To gain insight into severity and distribution of dystonia and to guide and delineate therapeutic interventions, reliable and valide assessments are indispensable
- Currently, three rating scales are used: the Barry-Albright Dystonia Scale (BADS), the Burke-Fahn-Marsden Scale (BFMS) and the Unified Dystonia Rating Scale (UDRS)

Aims

- To investigate the reliability of the BADS, BFMS and UDRS in patients with bilateral dystonic CP
- To assess the validity of the BADS, BFMS and UDRS in patients with bilateral dystonic CP

Methods

Participants

- 10 dystonic CP patients (5-22years)
- One patient each was classified as level I-IV in the Gross Motor Function Classification System and six were classified as level V

Procedure

- All patients were videotaped based on the videotaping protocol of the Dystonia Study Group (1997)
- Two child neurologist and one physical therapist independently scored the videotapes in a randomized order
- All raters had several years of clinical experience and were trained in scoring the three scales.
- Raters applied definitions of dystonia as described, according to each of the 3 investigated scales

Statistics

Reliability analysis

- Interrater reliability: intra class correlation coefficient (ICC)
- Standard error of measurement (SEM) and smallest detectable difference (SDD)
- Internal consistency: Cronbach's α

Validity Analysis

- Concurrent validity: Pearson's correlation coefficient
- Content analysis

Results

Barry-Albright Dystonia Scale (BADS)

Interrater reliability BADS		
Body region	ICC	95% CI
Eyes	0.39	0.00 - 0.77
Mouth	0.47	0.08 - 0.81
Neck	0.57	0.19 - 0.85
Trunk	0.65	0.29 - 0.89
Arm Right	0.63	0.26 - 0.88
Arm Left	0.61	0.24 - 0.87
Leg Right	0.76	0.47 - 0.93
Leg left	0.67	0.32 - 0.89
Total	0.87	0.67 - 0.96

Burke-Fahn-Marshden Scale (BFMS)

Interrater reliability BFMS						
Body region	Provoking Factor		Severity Factor		PF x SF	
	ICC	95% CI	ICC	95%CI	ICC	95%
Eyes	0.68	0.34 - 0.90	0.32	-0.07 - 0.72	0.62	0.25 - 0.87
Mouth	0.77	0.47 - 0.93	0.70	0.36 - 0.90	0.79	0.52 - 0.94
Speech & swal	0.41	0.02 - 0.78	0.93	0.81 - 0.98	0.81	0.55 - 0.94
Neck	0.56	0.17 - 0.85	0.68	0.34 - 0.90	0.71	0.39 - 0.91
Trunk	0.56	0.18 - 0.85	0.40	0.01 - 0.77	0.45	0.06 - 0.80
Arm R	0.34	-0.05 - 0.73	0.90	0.74 - 0.97	0.67	0.67 - 0.32
Arm L	0.35	0.04 - 0.74	0.87	0.66 - 0.96	0.72	0.40 - 0.91
Leg R	0.37	-0.02 - 0.76	0.87	0.67 - 0.96	0.71	0.37 - 0.91
Leg L	0.46	0.06 - 0.80	0.91	0.77 - 0.98	0.86	0.65 - 0.96
Total	0.64	0.28 - 0.88	0.89	0.72 - 0.97	0.86	0.66 - 0.96

Unified Dystonia Rating Scale (UDRS)

Interrater reliability UDRS						
Body region	Duration Factor		Motor Severity Factor		\sum DF + MSF	
	ICC	95% CI	ICC	95%CI	ICC	95%
Eyes	0.42	0.02 - 0.78	0.44	0.04 - 0.79	0.51	0.11 - 0.82
Lower face	0.71	0.38 - 0.91	0.63	0.26 - 0.88	0.78	0.50 - 0.93
Jaw/tongue	0.64	0.28 - 0.88	0.81	0.56 - 0.94	0.62	0.25 - 0.88
Larynx	0.17	-0.17 - 0.63	0.83	0.83 - 0.59	0.63	0.26 - 0.88
Neck	0.76	0.47 - 0.93	0.74	0.43 - 0.92	0.81	0.57 - 0.94
Trunk	0.41	0.01 - 0.78	0.54	0.15 - 0.84	0.49	0.09 - 0.81
Arm R	0.38	0.02 - 0.76	0.41	0.02 - 0.78	0.51	0.12 - 0.83
Arm R	0.46	0.06 - 0.80	0.58	0.20 - 0.86	0.59	0.22 - 0.86
Arm L	0.32	-0.06 - 0.73	0.50	0.11 - 0.82	0.57	0.19 - 0.85
Arm L	0.30	-0.08 - 0.72	0.49	0.09 - 0.81	0.47	0.07 - 0.81
Leg R	0.44	0.05 - 0.79	0.35	-0.04 - 0.74	0.42	0.03 - 0.78
Leg R	0.31	-0.08 - 0.72	0.48	0.09 - 0.81	0.39	0.00 - 0.77
Leg L	0.41	0.02 - 0.78	0.26	-0.11 - 0.69	0.37	-0.02 - 0.75
Leg L	0.29	-0.09 - 0.71	0.60	0.22 - 0.87	0.44	0.05 - 0.79
Total	0.74	0.43 - 0.92	0.79	0.51 - 0.94	0.79	0.52 - 0.94

Concurrent validity

- Pearson's correlation revealed high associations between the total scores of the BADS, BFMS and the UDRS: range 0.86 - 0.95 ($p < 0.001$)

Content analysis

- Limitations in sensitivity: less or no distinction between (1) rest/activity, (2) severity in time/amplitude, (3) prox/distal limbs
- Dystonia and choreoathetosis are simultaneously present in most of the dyskinetic CP patients

- High SEM (BADS) = 6.36%
- High SDD (BADS) = 17.72%
- Good Internal consistency: Cronbach's ranged from 0.87 to 0.91

- High SEM (BFMS) = 9.88%
- High SDD (BFMS) = 27.39%
- Good Internal consistency: Cronbach's ranged from 0.92 to 0.94

- High SEM (UDRS) = 8.89%
- High SDD (UDRS) = 24.63%
- Good Internal consistency: Cronbach's ranged from 0.93 to 0.95

References

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- Barry MJ et al. *Dev Med Child Neurol* 1999; 41:404-11
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Conclusions

- High SDD % makes the scales insufficiently useful for intervention studies and longitudinal follow-up
- Content analysis revealed limitations in sensitivity of the scale construction
- This study suggests to develop a new scale that evaluates dystonia and choreoathetosis separately in CP