

Egen Klassifikation (EK) revisited in spinal muscular atrophy

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Background

The Egen Klassifikation (EK) is an assessment instrument to determine functional ability in non-ambulatory people with Duchenne muscular dystrophy (DMD) or spinal muscular atrophy (SMA).

EK¹⁻¹⁰ is a composite scale consisting of 10 items that are clinically relevant to non-ambulatory persons with DMD or SMA.

Various studies on reliability and validity have been published 1995-2002. EK¹⁻¹⁰ is not as sensitive to change over time in individuals with SMA as in individuals with DMD.

A collaboration between physiotherapists in the United Kingdom, Italy and Denmark supported by Treat NMD identified ten, later reduced to 7, new items with special clinical relevance for SMA and characteristic for the non-ambulatory stages.

Purpose

to examine...

- Content validity
- Reliability
- Discriminatory power

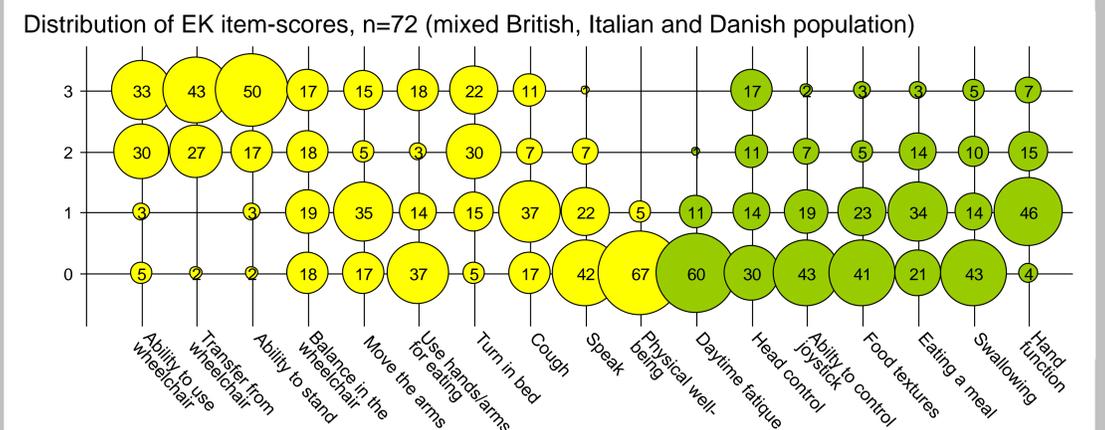
...of the 17 item version of EK (EK¹⁻¹⁷) in a population of persons with SMA.



Content validity

Methods: The 7 new items were tested for discriminatory and content validity in conjunction with the original EK¹⁻¹⁰ scale in 72 non-ambulatory persons with SMA, aged 2-70 years.

Results:



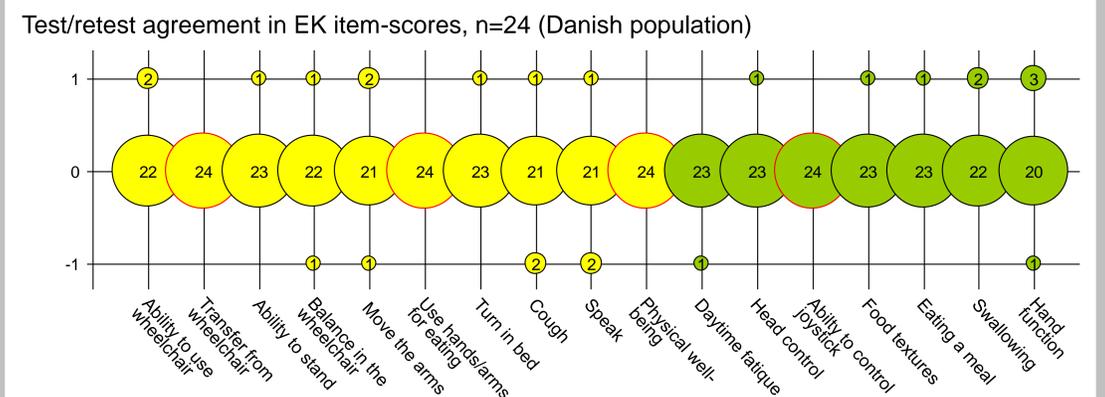
The weakest in the population had an EK¹⁻¹⁷-score of 42 (picture A), the strongest had an EK¹⁻¹⁷ score of 3 (picture B).

Conclusion: There might be ceiling or floor issues on individual items but not on the EK scores in a population with SMA. The content of the EK¹⁻¹⁷ scale is relevant for the discrimination of individuals with SMA.

Reliability

Methods: 24 persons with SMA were assessed 2 times by different experienced physiotherapists

Results:



Conclusion: Agreement between assessors in scoring EK¹⁻¹⁷ was consistent with earlier tests (+/- 1.5 EK sum units when performed by different assessors).

Discriminatory power

Methods: A regression model was composed to test to which degree variation in Muscle strength (MRC%), Forced Vital Capacity (FVC%), Brooke upper limb classification (Brooke) and Hammersmith Functional Measure Scale (HFMS) were explained by: the EK¹⁻¹⁰ and the EK¹⁻¹⁷ respectively.

Results:

	MRC% (n=50)	FVC% (n=49)	Brooke (n=58)	HFMS (n=56)
EK ¹⁻¹⁰	90%	75%	88%	82%
EK ¹⁻¹⁷	92%	81%	91%	84%

The percentages shown in the table is EK's degree of explanatory power (r^2) of the variance of the response variable

Conclusion: The categories of EK¹⁻¹⁷ have stronger explanatory power than the categories of EK¹⁻¹⁰