Appendix A

Procedure

The expert group formulated criteria for clinical suitability, based on their technical knowledge as well their practical experience with current instruments for the evaluation of motor abilities. In a first focus group meeting, each expert individually listed a series of requirements. This list of identified criteria was discussed and a clear definition of each criterion was drafted. In a second meeting, the criteria were ranked according to their perceived relevance and a final, limited set of criteria was agreed upon by consensus of the whole group.

Criteria for clinical suitability

The expert group formulated the following suitability requirements:

1) Low Level of motor abilities (GMFCS V). Motor functioning of children with SMD is very low, meaning these children are often unable to change body position, to take up and maintain an upright body position and moving around such as turning around, rolling and crawling. As a result, these children often stay in a supine position or sit in a fully supported wheelchair in daily living. Examples of motor abilities of a low level are for example raising the head, reaching with the arms, maintaining a sitting position or roll over. Motor abilities of a high level such as walking, running, jumping, catching, swimming and also tinkering, scissoring or washing are too complex for these children. Therefore an instrument for these children is suitable if the motor abilities are of a low level and not too difficult or complex. Motor abilities in lying or upright body positions will be an essential part of the test.

2) The grading or scoring should be sensitive to subtle changes in motor abilities. Due to the low level of motor abilities in children with SMD, in many instruments steps between changes in motor abilities are too large. Test items focused on changes in motor abilities are often related to the normal development such as the development from lying to standing in which intermediary steps are rolling over, sitting, crawling and running. Children with SMD often show subtle but relevant changes in motor abilities related to their care, eating or playing. Therefore an instrument is needed in which subtle changes have to be captured by small intermediary steps, which are relevant for these children. An example is the motor ability ‘rolling over’. The desired result is coming from lying on the back to supine lying. In children with SMD a step in between can be rolling over into lying on the side or the child’s cooperation during changing position in contrast to being passive.

3) Capability versus capacity and performance. An important part of instruments in general that assess motor ability is assessing the performance of motor abilities (what does the child do), which influences the score. In children with SMD the precise performance of a motor ability is of secondary importance. Of primary importance for these children is the capability in performing the desired motor ability no matter how that movement is executed (what can a child do). For example rolling over; a qualitative well-performed execution is to roll over by rotating the spine (performance). For motor functioning of these children the result and active participation is most important. Whether or not the child used rotation of the spine in the execution of rolling over is of lesser importance in these children (capability). Based on the difficulty to test the capacity (what the child can do in standardized situations) of motor abilities in a specific test situation, the capability of motor abilities of children with SMD have to be studied in a more naturalistic setting.

4) Non-verbal instruction; in children with SMD the level of understanding is very basic, which makes them barely instruction-oriented. In many instruments the execution of items is based on verbal assignments or

instructions. Due to the low level of cognitive development, which is comparable to that of children with a maximum age of one year, responding to commands is not possible.

5) Manual support and/or support by devices. Children with SMD need support maintaining a position due to their severe cognitive and motor disabilities. Support can be given by devices such as a wheelchair and can be given by manual support. More specifically, a child can be supported by sitting upright in a wheelchair but can also be supported to be active in sitting upright or to maintain this position by stimulation by the hands of another person. Another example is that, on the one hand the child can be pushed from supine to prone position during diaper change and, alternatively, the child can be activated and supported by the performance of rolling over. Instruments for evaluating motor abilities are often based on observation of spontaneous or provoked motor functioning. Children are provoked to comply with specific conditions by verbal instructions or by providing triggering materials without touching the child. Due to the severe motor, cognitive and sensory disabilities, these children are often unable to understand spoken language, perform motor abilities against gravity and comply with instructions without caretakers present to stimulate attention of the child. In the performance of daily activities these children often need to be supported by their surroundings.
The experts decided that a suitable instrument should satisfy all of these criteria.

Evaluating the instruments

Instruments identified as potentially suitable through the systematic review [3] were; 1) The Chailey Levels of Ability (CLA) [18,19] 2) Gross Motor Function Measure (GMFM-88) [20,21], 3) Modified Hammersmith Functional Motor Scale (MHFMS) [22,23], 4) Lower extremity physical functioning and mobility skills (LE85) [24], 5) Motor function measure scale (MFM) [25], 6) Top Down Motor Milestone Test (TDMMT) [12], 7) Vulpe Assessment Battery (VAB) [26] and 8) Functional Independence Measure for Children (WeeFIM) [27]. One of these instruments has been specifically developed for children with SMD (TDMMT), and the others for children with cerebral palsy (CLA, GMFM-88, LE85), spinal muscular atrophy (MHFMS), neuromuscular diseases (MFM), and motor disabilities in general (VAB, WeeFIM).

The experts agreed that all of the criteria had to be met for an instrument to be suitable for children with SMD. They considered and discussed the possibility to adapt an existing instrument. The instrument's clinical suitability is shown in the table below.

Table: Scoring of the clinical suitability of the instruments

	Criteria of clinical suitability*					
Instrument	**1)**	**2)**	**3)**	**4)**	**5)**	**Total of positive scored criteria**
CLA	+	+	-	+	-	3
GMFM	+	-	-	-	+	2
MHFMS	-	-	-	-	+	1
LE85	-	+	-	-	+	2
MFM	-	-	-	-	-	0

TDMMT	-	-	-	+	+	2
VAB	-	+	+	-	-	2
WeeFIM	-	-	-	+	+	2

*Criteria of clinical suitability

1) Low Level of motor abilities

2) The grading or scoring

3) Capability versus capacity and performance

4) Non-verbal instruction

5) Manual support and/or support by devices

+ = positive scoring

- = negative scoring

CLA = The Chailey Levels of Ability

GMFM-88 = Gross Motor Function Measure (88 items)

MHFMS = Modified Hammersmith Function Motor Scale

LE 85 = Lower Extremity physical functioning and mobility skills

MFM = Motor Function Measure scale

TDMMT = Top Down Motor Milestone Test

VAB = Vulpe Assessment Battery

WeeFim = Functional Independence Measure for Children

Conclusion

None of the instruments was found to be completely suitable and therefore the focus group decided that development of a new instrument is needed. This instrument will need to adhere to all suitability requirements and should have adequate psychometric properties.

Appendix B Modified version of Movakic

Structure of the instrument

Positions	Lying	Sitting	Standing
Situations (13) →	1 Supine 2 Supine with device 3 Prone 4 Prone with device 5 Side 6 Side with device	7 Flat surface 8 Dangling legs 9 Chair/ sitting device 10 feet on subsurface	11Without device 12 With device
Grouping of motor abilities ↓	13 Care situation		
Maintaining position	Items with questions on 1 Extent of manual or support by device 2 Activity of the child 3 Extent of manual facilitation/ stimulation		
Activities			
Changing body position			
Moving around			

Items of Movakic

		Situations (see table above)												
	Items	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**	**13**
1	Maintaining position	x		x		x		x	x	x	x	x	x	x
2	Duration maintaining position					x		x	x	x	x	x	x	x
3	Turning head	x	x	x	x	x	x	x	x	x	x	x	x	
4	Upright head	x	x	x	x	x	x	x	x	x	x	x	x	
5	Maintaining upright head position	x	x	x	x			x	x	x	x	x	x	
6	Reaching with the arms	x	x	x	x	x	x	x	x	x	x	x	x	
7	Take support (fore)arms			x	x									
8	Take support hands			x	x			x	x			x		
9	Grasping with the hands	x	x	x	x	x	x	x	x	x	x	x	x	
10	Roll over to the left	x		x										x
11	Roll over to the right	x		x										x
12	Roll over to prone	x				x								x
13	Roll over to supine			x		x								x
14	Transfer from lying to sitting	x	x	x		x								x
15	Transfer from sitting to lying							x	x					x
16	Transfer from sitting to standing							x		x	x		x	x
17	Transfer from standing to sitting									x	x	x	x	x
18	Pivoting							x						x

19	Minor voluntary postural changes					x	x	x			x
20	Move on	x	x		x		x		x	x	
21	Distance	x	x		x		x		x	x	
22	Move on and change direction						x		x	x	
23	Moving backwards						x		x	x	

Questions and answer categories

What is the intensity of manual support you give to the child?	o	o	o	o	o
	Full				None
What is the intensity of activity of the child?	o	o	o	o	o
	Full Passive				Full Active
What is your the intensity of facilitation to stimulate the motor ability?	o	o	o	o	o
	Full				None