

## Příloha č. 1: CHOP INTEND – 1. část (Glanzman, 2010)

The Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders							
CHOP INTEND							
Name: _____		Diagnosis: _____		Gestational age: _____ weeks			
Medical Record Number: _____		Date of Birth: _____		Age _____			
Date of Evaluation: ____/____/____		Time of evaluation: _____		Time since last feeding: _____			
Current health: Good health <input type="checkbox"/>		URI <input type="checkbox"/>		Other illness <input type="checkbox"/> : _____		Recent surgery <input type="checkbox"/> : _____	
Feeding support: None <input type="checkbox"/>		Nasogastric Tube <input type="checkbox"/>		Gastrostomy Tube <input type="checkbox"/>			
Respiratory support: None <input type="checkbox"/>		BiPAP <input type="checkbox"/>		for _____ hours/day _____ Time off BiPAP at testing _____			
	Position	Test Procedure	Graded Response	Score			
<b>1</b> Spontaneous movement (Upper extremity)	Supine	<i>Observe throughout testing</i>  May unweight limb or stimulate infant to facilitate response	Antigravity shoulder movement (achieves elbow off surface)	4	L	Best side:  Beazleton State:	
			Antigravity elbow movement (achieves hand and forearm off surface)	3			
			Wrist movement	2			R
			Finger movement	1			
			No movement of limbs	0			
<b>2</b> Spontaneous movement (Lower extremity)	Supine	<i>Observe throughout testing</i>  May unweight limb or stimulate infant to facilitate response	Antigravity hip movement (achieves feet and knees off surface)	4	L	Best side:  Beazleton State:	
			Antigravity hip adduction/internal rotation (knees off surface)	3			
			Active gravity eliminated knee movement	2			R
			Ankle movement	1			
			No movement of limbs	0			
<b>3</b> Hand grip	Supine	Grip strength: place finger in palm and lift until shoulder comes off surface observe when infant loses grasp  May use toy of similar diameter for older children	Maintains hand grip with shoulder off bed	4	L	Best side:  Beazleton State:	
			Maintains grip with elbow off surface (shoulders on surface)	3			
			Maintains grip with forearm off surface (elbow supported on surface)	2			R
			Maintains grip only with no traction	1			
			No attempt to maintain grasp	0			
<b>4</b> Head in midline with visual stimulation*	Supine head midline	Visual stimulation is given with toy.  <i>If head is maintained in midline for 5 seconds:</i> Place head in maximum available rotation and provide visual stimulation to encourage midline	Rotates from maximum rotation to midline	4	L>R	Best side:  Beazleton State:	
			Turns head part way back to midline	3			
			Maintains midline for 5 or more seconds	2			R>L
			Maintains midline, less than 5 seconds	1			
			Head falls to side, no attempts to regain midline	0			
<b>5</b> Hip adductors	Supine, no diaper	Hips flexed and adducted  Feet hip width apart and thighs parallel, knees slightly apart	Keeps knee off surface of bed > 5 sec or lifts foot off surface	4	L	Best side: Beazleton State:	
			Keeps knees off surface of bed 1-5 sec	2			
			No attempt to maintain knees off surface	0			R


## Příloha č. 2: CHOP INTEND – 2. část (Glanzman, 2010)

<b>6</b> Rolling: elicited from legs*	Supine (arms at side) Keep side tested up roll away from the  Side tested	1. Holding infant's lower thigh, flex hip and knee and adduct across midline bringing pelvis vertical  maintain traction and <i>pause in this position.</i> 2. If infant rolls to side apply traction at a 45° diagonal to body and <i>pause</i> to allow infant to attempt to derotate body	When traction is applied at the end of the maneuver, rolls to prone with lateral head righting	4	To R	Best side:  Brazelton State:
			Rolls through side lying into prone without lateral head righting, clears weight-bearing arm to complete roll	3	To L	
			Pelvis, trunk and arm lift from support surface, head turns and rolls onto side, arm comes thru to front of body	2		
			Pelvis and trunk lift from support surface and head turns to side. Arm remains behind trunk	1		
			Pelvis lifted passively off support surface.	0		
<b>7</b> Rolling: elicited from arms*	Supine (arms at side) Keep side tested up roll away from the  Side tested	1. Hold infant at the elbow move toward opposite shoulder maintain traction on limb and <i>pause with the shoulders vertical</i> allow infant to derotate 2. if the pelvis achieves vertical continue to provide traction	Rolls to prone with lateral head righting	4	To R	Best side:  Brazelton State:
			Rolls into prone without lateral head righting; must clear weight-bearing arm completely to finish roll	3	To L	
			Rolls onto side, leg comes thru and adducts, bringing the pelvis vertical	2		
			Head turns to side and shoulder and trunk lift from surface	1		
			Head turns to side; body remains limp or shoulder lifts passively	0		
<b>8</b> Shoulder and elbow flexion And horizontal abduction	Side-lying with upper arm at 30° of shoulder extension and elbow flexion and supported on body (restrain lower arm if needed)	Prompt reach for a toy presented at arms length at shoulder level (may provide stimulation and <i>observe spontaneous movement</i> )	Clears hand from surface with antigravity arm movement	4	L  R	Best side:  Brazelton State:
			Able to flex shoulder to 45 degrees, without antigravity arm movement	3		
			Flexes elbow after arm comes off body	2		
			Able to get arm off body	1		
			No attempt	0		
<b>9</b> Shoulder flexion & Elbow flexion	Sitting in lap or on mat with head and trunk support (20° recline)	Present stimulus at midline and at shoulder level at arms length (may provide stimulation and <i>observe spontaneous movement</i> )	Abducts or flexes shoulder to 60 degrees	4	L  R	Best side:  Brazelton State:
			Abducts or flexes shoulder to 30 degrees	3		
			Any shoulder flexion or abduction	2		
			Flexes elbow only	1		
			No attempt to lift arm	0		
<b>10</b>	Sitting in lap or over edge of mat with head and trunk support (20° recline) thigh horizontal to ground	Tickle plantar surface of foot Or gently pinch toe	Extends knee to > 45 degrees	4	L  R	Best side:  Brazelton State:
			Extends knee 15 to 45 degrees	2		
			Any visible knee extension	1		
			No visible knee extension	0		



## Příloha č. 3: CHOP INTEND – 3. část (Glanzman, 2010)

<b>11</b> Hip flexion and foot dorsiflexion	Hold infant against your body with legs flex, facing outward. Support at the abdomen with the child's head resting between your arm and thumb.	Stroke the foot or pinch the toe	Hip flexion or knee flexion > 30°	4	L R	Best side: Brazelton State:
			Any hip flexion or knee flexion	3		
			Ankle dorsiflexion only	2		
			No active hip, knee or ankle motion	0		
<b>12</b> Head control*	Sitting with support at the shoulders and trunk erect	Place the infant in ring sit with head erect and assistance given at the shoulders (front and back). <i>(may delay scoring a grade of 1 and 4 until end of test)</i>	Attains head upright from flexion and turns head side to side	4		Score: Brazelton State:
			Maintains head upright for >15 sec (for bobbing head control score a 2)	3		
			Maintains head in midline for >5 sec. with the head tipped in up to 30° of forward flexion or extension	2		
			Actively lifts or rotates head twice from flexion within 15 seconds (do not credit if movement is in time with breathing)	1		
			No response, head hangs	0		
<b>13</b> Elbow flexion Score with item 14	Supine	Traction response: pull to sit extend arms at 45 degree angle, to point of nearly lifting head off surface	Flexes elbow	4	L R	Best side: Brazelton State:
			Visible biceps contraction without elbow flexion	2		
			No visible contraction	0		
<b>14</b> Neck Flexion Score with item 13	Supine	Traction response: hold in neutral proximal to wrist and shoulder at 45°, to point of nearly lifting head off surface	Lifts head off bed	4		Score: Brazelton State:
			Visible muscle contraction of SCM	2		
			No muscle contraction	0		
<b>15</b> Head/Neck Extension (Landau)	Ventral suspension: Prone, held in one hand upper abdomen	Stroke along the spine from neck to sacrum. The coronal axis of the head when parallel to the bed surface = 0 degrees (horizontal)	Extends head to horizontal plane or above	4		Score: Brazelton State:
			Extends head partially, but not to horizontal	2		
			No head extension	0		
<b>16</b> Spinal Incurvation (Galant)	Ventral suspension: Prone, held in one hand upper abdomen	Stroke Right then Left thoracolumbar paraspinals or tickle abdomen or foot or tih in infants with integrated Galant For infant over 10 kg knees and head may touch	Twists pelvis towards stimulus off axis	4	L R	Best side: Brazelton State:
			Visible paraspinal muscle contraction	2		
			No response	0		
Total score, best score on each side for each item (maximum 64 points):						

## Příloha č. 4: HFMSE – 1. část (Ramsey, 2017)

Revised Hammersmith Scale for SMA (RHS)							17.03.2015			
Name		SMA Type		DOB		Assessor				
Date of assessment		Albuterol/Salbutamol		Y / N		Date of spinal surgery				
Please circle <u>highest</u> current level of independent mobility:										
None		Rolls		Bottom shuffles		Crawls		Walks with KAFO's / AFO's	Walks with crutches / frame	Independent walking
Comment.....										
LBC = Limited by contracture <span style="float: right;">*WHO separate scoring at end of RHS</span>										
Test	Instruction	2	1	0	L B C	Comments	RHS Score			
SITTING	1* <b>Sitting</b> Using plinth / floor. (Back / feet unsupported)	Sitting unsupported: Ring or 90/90	Maintains seated position via propping with hand/s: Ring or 90/90	Unable to sit		WHO 1* <input type="checkbox"/> Long sitting				
	2 <b>Hands to head in sitting</b>  (hands touch head above level of ears/eyebrows)	Able to bring both hands to head at same time – arms free from side without flexing head or trunk	Able to bring one hand to head – arms free from side without flexing head or trunk <input type="checkbox"/> R <input type="checkbox"/> L	<input type="checkbox"/> Using compensations – flexing head & trunk or "crawling" hand/s <input type="checkbox"/> Unable to bring hand to head		Sitting position described: <input type="checkbox"/> 90/90 <input type="checkbox"/> Ring <input type="checkbox"/> Long				
	3 <b>Sitting to lying</b>	Able to lie down through side lying or using clothes in a controlled/safe way	Able to lie down by going forwards and rolling sideways, or through prone in a controlled/safe way	Unable or completes in uncontrolled/unsafe way						
SUPINE	4 <b>Adduction from crook</b> (hook) lying – supine hips @ 45°, knees @ 90°, feet hip width apart. Passively abduct leg	Able to adduct to bring 1 leg back to neutral	Holds crook lying position for a count of 3	Unable to maintain/achieve starting position.		<input type="checkbox"/> R <input type="checkbox"/> L				
	5 <b>Right hip flexion in supine</b>	Full hip flexion achieved	Initiates right hip and knee flexion (>10% of available range of motion)	Unable		Flexion contracture > 15° present? Hip Y <input type="checkbox"/> N <input type="checkbox"/> Knee Y <input type="checkbox"/> N <input type="checkbox"/>				
	6 <b>Left hip flexion in supine</b>	Full hip flexion achieved	Initiates left hip and knee flexion (>10% of available range of motion)	Unable		Flexion contracture > 15° present? Hip Y <input type="checkbox"/> N <input type="checkbox"/> Knee Y <input type="checkbox"/> N <input type="checkbox"/>				
ROLLING	7 <b>Lifts head from supine</b>	Can lift head up through neck flexion and holds for a count of 3	<input type="checkbox"/> Can lift head with compensatory movements for a count of 3 <input type="checkbox"/> Can lift head up momentarily through neck flexion (< a count of 3)	Unable						
	8 <b>Supine to side-lying</b> (end position leading leg ends up on top of other)		Able to roll onto side	Unable to roll onto side in any direction		<input type="checkbox"/> R <input type="checkbox"/> L				
	9 <b>Rolls supine to prone</b>	Rolls fully into prone without pulling/pushing on arms	Rolls fully into prone by pulling/pushing on arms	Unable to roll into prone in any direction. Does not initiate or complete		<input type="checkbox"/> R <input type="checkbox"/> L				
PRONE	10 <b>Lifting head from prone</b>	Lifts straight up, arms down by sides for a count of 3	Lifts straight up arms forward for a count of 3 	Unable or lifts less than a count of 3						
	11 <b>Prone; prop on forearms</b>	Achieves position and holds head up independently for a count of 3	Maintains position with head supported on hands for a count of 3	Unable or holds for less than a count of 3						
ROLLING	12* <b>Four-point kneeling/crawling</b>	Crawls moving all 4 limbs at least 2 times in a row	Achieves four-point kneeling	Unable		WHO 2*				
	13 <b>Rolls prone to supine</b>	Rolls fully into supine without pulling/pushing on arms	Rolls fully into supine by pulling/pushing on arms	Unable to roll into supine in any direction. Does not initiate or complete		<input type="checkbox"/> R <input type="checkbox"/> L				

## Příloha č. 5: HFMSE – 2. část (Ramsey, 2017)

Test	Instrucción	2	1	0	L B C	Comments	RHS Score
SITTING	14 Lying to sitting Can you get from lying (supine) to sitting?	Able via supine / side lying using: <input type="checkbox"/> 1 hand <input type="checkbox"/> 2 hands	Able by using strategies: <input type="checkbox"/> Turns into prone or towards floor <input type="checkbox"/> Utilises momentum/ pulls on both legs	Unable			
	15 Sit to stand (Starting in a 90/90 position using a Plinth / Bench / Chair) Can you stand up from the chair / bench keeping your arms folded if you can?	Able to stand up without using arms (keeping arms folded, feet do not move)	Stands by using strategies: (circle relevant) Uses hands on legs/chair Turns Prone / Widens Base	Unable			
STANDING	18* Cruising/ supported stand Can you walk holding on around furniture?	Cruise at least 5 steps around furniture <input type="checkbox"/> R <input type="checkbox"/> L	Stands supported for a count of 3	Unable to stand supported		WHO 3 & 4*	
	17* Standing Can you stand without holding onto anything for a count of 3?	Stands upright and symmetrically, without compensation for a count of 3	Stands but with some degree of compensation for a count of 3	Cannot stand independently, needs support		WHO 6*	
RUN / WALK	18* Walking Can you walk without using any help or aids? Show me	Takes at least 5 steps unaided	Able to take < 5 steps unaided	Unable		WHO 8*	
	19 Runs 10m Can you go as quickly and safely as you can to.....(give 10 m marker) *Ready steady GO?	Can run - both feet off the ground	Speeds up walk but always maintains one foot on the floor	<input type="checkbox"/> Walks with no extra speed <input type="checkbox"/> Only with aids <input type="checkbox"/> Unable		TIME:.....seconds	
STANDING	20 Squat down and up Can you squat down and stand up again?  Pretend you are going to sit on a very low seat - only go as far down as you are able to get up from on your own	Full squat down (without using hands) and up* (with/without using hands)  At least 90° achieved at knees/ hips  *must be able to get up from final range	Initiates squat in both knees (10° to <90°) with controlled movement +/- using hands on self  <input type="checkbox"/> Using hands	Unable			
	21 Stand to sit on floor Can you sit on the floor in a controlled/ safe way from standing?  Try not to use your arms	Able to sit down arms free in a controlled manner	Sits on floor using hands on floor/body	<input type="checkbox"/> Uses furniture <input type="checkbox"/> Unable			
MAT	22 High kneeling Can you kneel like this (high kneeling) for a count of 10?	Maintains high kneeling without holding on for a count of 10	Maintains high kneel with one arm support for a count of 10	Unable			
	23 High Kneeling to Right half kneel  (Left foot forward)	Able to achieve and maintain position without arm support for a count of 10	Maintains half kneel with one arm support for a count of 10  With: <input type="checkbox"/> support <input type="checkbox"/> no support	Unable			
	24 High Kneeling to Left half kneel  (Right foot forward)	Able to achieve and maintain position without arm support for a count of 10	Maintains half kneel with one arm support for a count of 10  With: <input type="checkbox"/> support <input type="checkbox"/> no support	Unable			
MAT / FLOOR	25 Rise from floor Can you get up from the floor using as little support as possible and as fast as you can?  Stand up as quickly as possible when the instruction *GO* is given	Achieves rise from floor without hands (any unsupported method)	Able to get up from floor using hands on floor and/or body	<input type="checkbox"/> Able to get up from floor using furniture/ assistance  <input type="checkbox"/> Unable to get up from floor using any method		If uses 1/2 kneel: <input type="checkbox"/> R (L foot forwards) <input type="checkbox"/> L (R foot forwards) Further detail: TIME:.....seconds	
SINGLE LEG STAND	26 Stand on one leg – Right Can you stand on your right leg for a count of 3?	Able to stand in a relaxed manner (no fixation) for a count of 3	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable			
	27 Stand on one leg – Left Can you stand on your left leg for a count of 3?	Able to stand in a relaxed manner (no fixation) for a count of 3	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable			
	28 Hops – Right Can you hop on your right leg?		Distinct hop: clears forefoot and heel off floor	Unable			
	29 Hops – Left Can you hop on your left leg?		Distinct hop: clears forefoot and heel off floor	Unable			

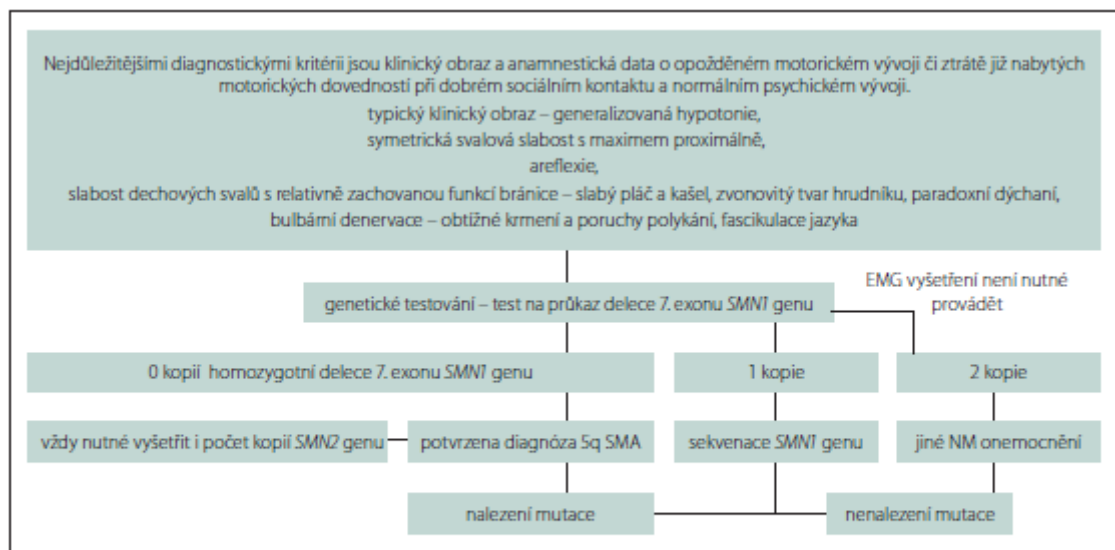
## Příloha č. 6: HFMSE – 3. část (Ramsey, 2017)

Test	Instruction	2	1	0	L B C	Comments	RHS Score
STAIRS	30 Ascend stairs 4 standard stairs	Can you walk up the steps? <i>If support is needed you may only use one rail (can use two hands to one rail)</i>	Independently (without support/rail) ascends using alternating pattern	<input type="checkbox"/> Alternating pattern with one support (one rail/hand on body) <input type="checkbox"/> Marking time with/without support	Unable		
	31 Descend stairs 4 standard stairs	Can you walk down the steps? <i>If support is needed you may only use one rail (can use two hands to one rail)</i>	Independently (without support/rail) descends using alternating pattern	<input type="checkbox"/> Alternating pattern with one support (one rail/hand on body) <input type="checkbox"/> Marking time with/without support	Unable		
STEP	32 Climbs box step – Right	Can you step onto the top of the box using your right leg first?	Faces forwards, climbs up – no support needed	Goes up sideways or needs support	Unable		
	33 Descends box step – Right	Can you step down from the box using your right leg first?	Faces forward, climbs down controlling weight bearing leg. No support needed	Goes down sideways or skips down or needs support	Unable		
	34 Climbs box step – Left	Can you step onto the top of the box using your left leg first?	Faces forwards, climbs up – no support needed	Goes up sideways or needs support	Unable		
JUMP	35 Descends box step – Left	Can you step down from the box using your left leg first?	Faces forward, climbs down controlling weight bearing leg. No support needed	Goes down sideways or skips down or needs support	Unable		
	38 Jump forwards 30cm (12")	Can you jump as far as you can, with both feet, from this line all of the way to the other line?	Jumps forward and lands with two feet simultaneously moving forward at least 30cm (12")	<input type="checkbox"/> Jumps with two feet simultaneously but lands one at a time moving forward/up travelling < 30cm (12") <input type="checkbox"/> Jumps and lands with two feet simultaneously moving forward < 30cm (12")	Unable to initiate jump		
		2's =	1's =	0's =		Total =	/69

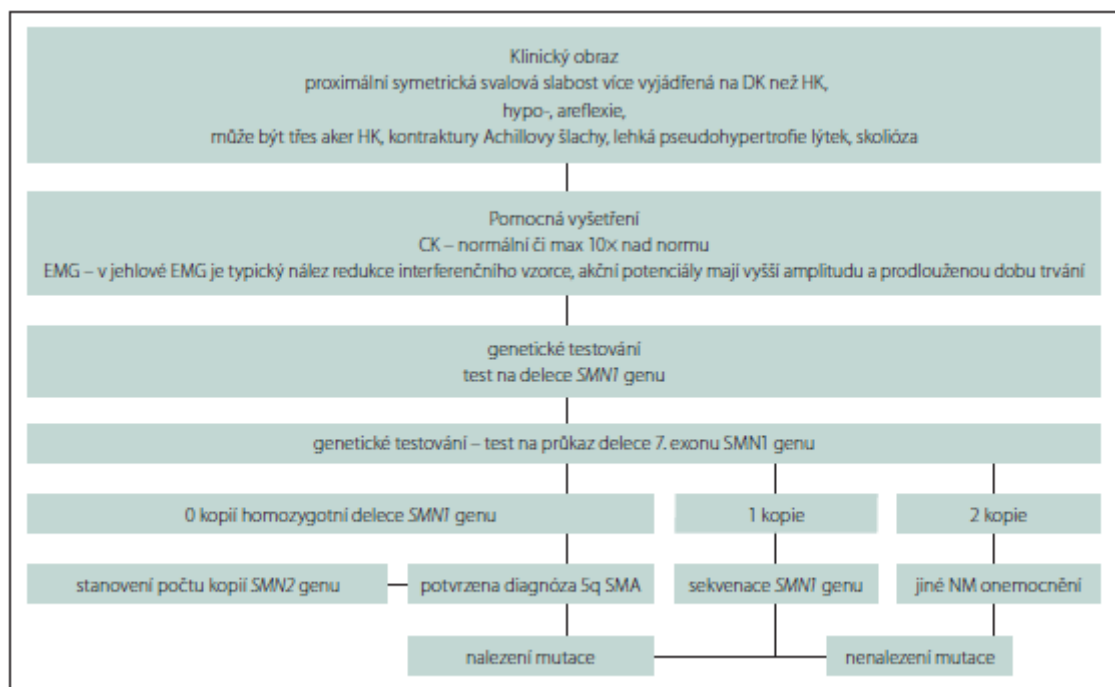
*WHO DEVELOPMENTAL MILESTONES	Qualifier met:
W1. Sitting without support: Child sits upright with head erect for at least 10 seconds. Child does not use arms or hands to balance body or support position.	Y <input type="checkbox"/> N <input type="checkbox"/>
W2. Hands and knees crawling: Child alternately moves forward or backward on hands and knees. The stomach does not touch the supporting surface. There are continuous and consecutive movement, at least 3 in a row.	Y <input type="checkbox"/> N <input type="checkbox"/>
W3. Standing with Assistance: Child stands in upright position on both feet, holding onto a stable object (e.g. furniture) with both hands without leaning on it. The body does not touch the stable object, and the legs support most of the body weight. Child thus stands with assistance for at least 10 seconds.	Y <input type="checkbox"/> N <input type="checkbox"/>
W4. Walking with Assistance: Child is in upright position with the back straight. Child makes sideways or forward steps by holding onto a stable object (e.g. furniture) with one or both hands. One leg moves forward while the other supports part of the body weight. Child takes at least 5 steps in this manner.	Y <input type="checkbox"/> N <input type="checkbox"/>
W5. Standing Alone: Child stands in upright position on both feet (not the toes) with the back straight. The legs support 100% of the child's weight. There is no contact with a person or object. Child stands alone for at least 10 seconds.	Y <input type="checkbox"/> N <input type="checkbox"/>
W6. Walking Alone: Child takes at least 5 steps independently in upright position with the back straight. One leg moves forward while the other supports most of the body weight. There is no contact with the person or object.	Y <input type="checkbox"/> N <input type="checkbox"/>
<b>Total WHO Milestones Achieved:</b>	
	/ 6

Revised Hammersmith Scale for SMA (RHS)	17.03.2015
Is this test a true reflection of the subjects ability Y <input type="checkbox"/> N <input type="checkbox"/> If no please detail why	
Comments on Assessment	

## Příloha č. 7: Diagnostický algoritmus spinální muskulární atrofie (Ruchsová, 2020)



Kojenci a malé děti do 3 let



Větší děti od 3 let věku, adolescenti a dospělí