## **CORE DOMAINS**

#### 1. C Communication

Engage the child or young person in conversation. Score from the best spontaneous speech up to the end of the assessment; encourage speech as appropriate; if child is unwilling return to the task later, e.g. at the end of the assessment, or mark item(s) as "Not Assessed" (NA).

C.a. Max Phonation Time    Ca
0= Significantly impaired 0= Significantly impaired 1= Mildly impaired 1= Mildly impaired 2= Normal 2= Normal NA NA NA  C.c. Articulation C.g. Response time 0= Significantly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 2= Normal NA NA  C.d. Prosody C.h. Rate 0= Significantly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 2= Normal NA NA  C.d. Prosody C.h. Rate 0= Significantly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 1= Normal NA NA  C.i. Speech Intelligibility 0= Speech is an inadequate means of communication (even with known context) 2= Only words, short or rote phrases (adequate when predictable or in known context) 4= Meaning understood, some phrases missed (adequate when unpredictable even when context) 6= Meaning understood, some phrases missed (adequate when unpredictable even when context is not known)
0= Significantly impaired 1= Mildly impaired 3= Normal NA  C.d. Prosody C.h. Rate 0= Significantly impaired 1= Mildly impaired NA  C.d. Prosody C.h. Rate 0= Significantly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 1= Mildly impaired 2= Normal NA  NA  C.i. Speech Intelligibility 0= Speech is an inadequate means of communication (even with known context) 2= Only words, short or rote phrases (adequate when predictable or in known context) 4= Meaning understood, some phrases missed (adequate when unpredictable even when context) 6= Meaning understood, some phrases missed (adequate when unpredictable even when context is not known)
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NA
Speech by Observation /26

If part of the assessment is not possible ("NA"), adjust the score up as if it was out of 26; if no speech is demonstrated, but the patient appears to be selectively mute (from the parent / caregiver history) score as "NA". The reported speech can be scored by the A-T Functional Scale.

# C.j. Best handwriting / drawing

Ask a pre-school child to copy developmentally appropriate shapes you have just drawn, in the box below, e.g. dots and lines under 3 years, circle age 3, perpendicular cross age 4, perpendicular rectangle age 4.5, diagonal cross age 5, triangle age 5.5 etc.

Ask young primary school child or older child or adult to write their name and a simple phrase.

Sampl 0= 1= 2= 3= 4= NA or	le of short phrase for size and position (school age and older; 3-10 words None Single character More than half words with character variation (size or position) Less than half words with character variation (size or position) All words with consistent character size and variation	in line)		
Sampl 0= 1= 2= 3= 4= NA or	None Scribble Some legible but less than half the letters More than half the letters legible All letters legible			
Copy 6 0= 1= 3= 4=	of primary shapes (pre-school or younger than school age)  None severely impaired, very slow mildly impaired and or slow normal for age			
Prima	ary shapes			
NA	F	Raw C		/30
	То	tal C	$\dashv$	/15

# 2. E Eye movements video if possible

### E.a. Conjugate gaze

0= Persistent dysconjugate in primary position: eso exo 1= Transient dysconjugate (i.e. tropia): eso exo

2= Conjugate

NA

#### E.b. Horizontal nystagmus in primary gaze

0= Sustained
1= Unsustained

2= Absent

NA

#### E.c. Nystagmus on lateral gaze

0= Sustained 1= Unsustained 2= Absent

NA

#### E.d. Convergence / Accommodation (if dysconjugate examine with each eye covered in turn)

0= Absent

1= Impaired but either transient or limited

2= Normal

NA

#### E.e. Off-foveal gaze tendency (vertical or horizontal)

0= Frequent and persistent

1= Present only with certain activities

2= Not present (normal)

NA

## E.f. Pursuit (away from primary gaze)

0= No pursuit velocity movement generated

1= Fragmentary

2= Normal

NA

#### **E.g. Saccade amplitude** (head fixed, horizontal gaze-shift of 30 degrees)

0= No saccades

1= Multiple small hypometric or staircase saccades

2= Perseveration of hypometric saccades with repeated gaze shifts to defined target

3= Normal amplitude with experience to defined target

## E.h. Saccade latency

All or most delayed

1= Less than 50% delayed

2= Normal latency

NA

# E.i. Oculo-Motor Apraxia (large amplitude lateral gaze-shifts)

Present on all gaze shifts

Present on most (> 50%) gaze shifts 1=

2= Occasionally present

Not present 3=

NA

# E.j. Up Gaze restriction

0= No vertical gaze

1= Limited vertical gaze

Normal vertical gaze 2=

NA

#### E.k. Up gaze persistence or vertical nystagmus

Persistent

1= Present under certain conditions

2= Not present (normal)

NA

#### E.I. VOR gain

0 =Abnormal

1= Normal

NA

### E.m. VOR cancellation

0= Absent

1= Diminished

2= Normal

NA

### E.n. Post-rotatory nystagmus (after slow rotation, 6 full rotation, 6 sec/rotation)

Persists more than 10 secs

1= Present, but persists no more than 10 secs

2= Not present

NA

### E.o. Periodic Alternating Nystagmus (after slow rotation, 6 full rotation, 6 sec/rotation)(score only if nystagmus present in primary gaze)

0= Present

1= Not present

NA

Raw E

Total E /15 Running total C E

#### 3. A Ataxia

A.a. Finger to nose (target horizontal distal phalanx; 10 if possible, right to right, then left to left)

A.a.R A.a.L Right Left

0= 0= On target or mid phalanx 2/10 (or 25%) times or less, or mean 5 sec or longer for each

1= On target or mid phalanx 3/10 (25%) or more, and less than 5 sec mean 1=

2= 2= On target or mid phalanx 8/10 (75%) or more

3= 3= On target 8/10 (75%) or more

On target 10/10 (100%) but with some in-coordination 4= 4=

5= 5= Normal, no in-coordination

NA NA

**A.b. Sitting** (observe 30 sec. on firm examination couch, or the edge of seat, arms crossed with hands on contra-lateral shoulders, feet un-supported)

0= Requires total support

Requires only flat posterior support 1=

2= Sits with self support of arms in lap or laterally

3= No support but sways markedly (raises body part for balance)

No support but sways slightly /occasionally 4=

5= Normal /age appropriate / no sway

NA

A.c. Standing (30 sec. once feeling confident. Score "NA" if foot deformity prevents passive ranging to a stable weight bearing position. Can use braces and shoes if required to keep feet plantigrade)

0= unable to stand without vertical support

- 1= consistently leans into or away from supporting hand (i.e., supporting against fall in only one direction); can stand leaning against a wall, 30 seconds (≥ 75% of trials)
- 2= consistently able to stand upright with one hand held securely at chest level, elbow partially flexed, but without consistently leaning into on or away from support (hence must guard against potential falls in either lateral, forward/backward, or twisting directions), 30 seconds (≥ 75% of trials)
- stands independently without support but requires corrective steps, 30 seconds (≥ 75% of trials) 3=
- 4= feet separated no more than shoulder width, no corrective steps, 30 seconds (≥ 75% of trials)
- 5= feet together, minor excessive corrective movement but no corrective steps, 30 seconds (≥ 75% of trials)

feet together, no corrective movements, 30 seconds 6=

A.d. Walking (10 meters)  0= Requires vertical support  1= Massive lateral support (above elbow or ward)  2= Some lateral support (below elbow or hand)  3= Only touch support needed  4= No support, wide base or takes corrective some support, normal speed with mild path defined by Normal path width without corrective steps  NA	held, elbow relaxed) tagger steps
	Total A / 27
	Running total C E A / 57
4. M Movement disorder	
Bradykinesia M.a. Hand Tapping (score best side after demo) (taps hand on paper on table or wheelchair tray at elbow height as fast as possible, with wrist held down, for 10 secs, ensure optimal support)	<ul><li>M.b. Body hypokinesia</li><li>0= Slowness and poverty of movements</li><li>1= Normal</li></ul>
Number of taps in 10 secs  0= < 5 taps  1= 5 < 10 taps  2= 10 < 20 taps  3= 20 < 30 tap  4= 30 < 40 taps  5= 40 and more	

Bradykinesia

	Present with both social / cognitive and motor activation  Present with either social / cognitive or motor activation  Not present
M.d. 0= 1= 2= 3= 4= NA	Retro-Colic Spasms / Jerks  Present with all postures at rest  Present sitting posture with hand support  Present with both social /cognitive and motor and posture activation / removal of head rest  Present with either social /cognitive or motor or posture activation / removal of head rest  Not present
M.e. 0 0= 1= 2= 3= NA	Other proximal adventitious movements – not rhythmic / tremor (face, head, trunk)  Present at rest  Present with both social / cognitive and motor activation  Present with either social / cognitive or motor activation  Not present
M.f. T 0= 1= 2= 3= NA	Present at rest / without activation Present with both social /cognitive and motor activation Present with either social /cognitive or motor activation Not present
M.g 0= 1= 2= 3= NA	Present with full support Present with removal of head rest / support Present with removal of trunk support Present with removal of trunk support Not present  Hyperkinesia /16
<b>M.h. I</b> 0=Lim	onia / posture distortions / slow movements  Facial expression / timing and duration of large expressions  nited or absent emotion expression (mask-like or expressionless face)

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2=Normal

M.i. T 0= 1= 2= 3= NA	No lateral movements (on command with mouth open) No lateral movements (moves only with mandible) Lateral movements depend on some mandible movement Lateral movements independent of mandible but impaired Normal lateral movements	
<b>M.j. I</b> 0= 1= 2= 3= NA	Present at rest / without activation Present with both social /cognitive and motor activation Present with either social /cognitive or motor activation Not present	
M.k. I 0= 1= 2= 3= NA	Head / trunk posturing / dystonia  Present at rest / without activation  Present with both social /cognitive and motor activation  Present with either social /cognitive or motor activation  Not present	Dystonia //11 Total M // 33
		Running total C E A M / 90

5. P Power (document an P.a. Ankle Dorsiflexion	y muscle 0= 1= 2= NA	weakness in box, so Severe weakness Mild weakness Normal for age	ore weakest side) (MRC 0, 1, or 2) (MRC 3, or 4) (MRC 5)	MRC #:
P.b. Knee extension	0= 1= 2= NA	Severe weakness Mild weakness Normal for age	(MRC 0, 1, or 2) (MRC 3, or 4) (MRC 5)	MRC #:
P.c. Hip flexion	0= 1= 2= NA	Severe weakness Mild weakness Normal for age	(MRC 0, 1, or 2) (MRC 3, or 4) (MRC 5)	MRC #:
P.d. Arm abduction	0= 1= 2= NA	Severe weakness Mild weakness Normal for age	(MRC 0, 1, or 2) (MRC 3, or 4) (MRC 5)	MRC #:
P.e. Elbow flexion	0= 1= 2= NA	Severe weakness Mild weakness Normal for age	(MRC 0, 1, or 2) (MRC 3, or 4) (MRC 5)	MRC #:
Document muscle weaknes	ss:			
P.f. Hand muscles	0= 1= 2= NA	Claw hand (extension Thenar / hypothenal Normal	on of all 4 MCP joints at r wasting	rest)
P.g. Document foot deform	ity:			
P.h. Document spinal defor	mity:			
				Raw P/12
				Total P /4
			Running total C E	AMP / 94

Neuropathy (tendon reflexes presenting) N.a. Ankle tendon reflexes  0= Absent  1= Present  NA	N.b. Knee tendon reflexes  0= Absent  1= Present  NA
N.c. Biceps tendon reflexes 0= Absent 1= Present NA	
N.d. Proprioception in toes (if unvibration sense with 128 Hz tuning 0= Absent movement sensibility intacts 1= Movement sensibility intacts 2= Rare position errors 3= No position errors / normation NA OR	lity ct, position errors >25%
	(only if proprioception not reliable) ct, errors >25%
N.e. Plantar responses normal	Right Left
equivocal	
absent striatal	
Babinski	
N.f. Tone description  Normal symmetrica	al Abnormal
Document abnormal tone:	
	Total N / 6
	Total Pure Neuro (C E A M P N)

6.

# **GROWTH / NUTRITION, SWALLOWING AND FEEDING SUPPLEMENTAL DOMAINS**

7. Gr ( Height Weigh	::	n (4 points) cm (centile: kg (centile:	) OR )	Head: Arm s	pan:	cm (ce	entile:	)		
1: 0.4t 2: 2th	4th cen h < 2nd < 9th ce	tile centile	0: < 2 <sup>n</sup> 1: 2 <sup>n</sup> 2: 9 <sup>th</sup>	eight fo centile < 9 <sup>th</sup> c < 25 <sup>th</sup> c	entile centile	:	OR BMI SDS 0: > -2.67 SD 1: -2.67 > -2.0 2: -2.0 > -1.3 4: -1.33 SDS	S O SDS 33 SDS	Gr/ 4	
		on (score by observat	tion or re	eport, 2	0 points	)				
Feedin	ig tube	Y L NL								
<b>F.a. Dr</b> 0= 1=	Droolin	(NA <4 years of age) g all the time only when tired <b>and</b> of	concent	rating	1=	Cough	with all liquid i when drinking w		thick liquids	3
2= 3= 4= NA		only when tired <b>or</b> colaccumulates at sides			2= 3=	Cough Cough	when drinking when drinking I for age			
F.b. Fe 0= 1= 2= 3= 4= 5= NA	No fund Needs Needs Needs Indepe	ctional self-feeding help with some feedir help with some feedir help with some feedir ndent feeding only wit I for age	ng, even ng, even	with all with sp	l utensils					
F.d. Sv 0= 1= 2= 3= 4= 5= 6= 7= NA	Nothing Tube d Tube d Partial with sp Partial without Total o Total o	ng / Diet Status g by mouth ependent with minima ependent with consist oral feeding with tube ecific liquid/food limita oral feeding with tube specific liquid/food lir ral feedings with spec ral feedings with care ral feedings without ar	ent oral suppled ations or suppled mitations ific liquingiver us	intake mentation special mentations or special different	on for liq I prepara on for liq cial prep imitation ecific fee	quids or ations quids or paration as or sp eding ro	nutrition, and nutrition, and nutrition, and nutrition, and nutrition, and nutricial preparation nutrines	oral feed ons Raw Nu Total N	dings	9,
							MPNGrNu)			

# **LEARNING ABILITY / MENTAL STATE SUPPLEMENTAL DOMAINS**

<ol><li>LA Learning Ability / Cognitive (score by Self Report or Carer Report observation, 3 points)</li></ol>	t, and confirm if possible b
0= Profound or severe cognitive impairment likely 1= Moderate cognitive impairment likely 2= Mild global cognitive impairment likely 3= Normal or superior cognition, or just specific learning difficulties NA	Total LA / 3
10. MS Mental State (score by clinical impression, 8 points) a. Appearance 0= Impaired 1= Normal NA	
<ul> <li>b. Mood</li> <li>0= Severe abnormality; severely manic or depressed</li> <li>1= Moderately depressed, or flat affect, or moderately manic or hypomanic</li> <li>2= No abnormality detected, a range of emotions was expressed</li> <li>NA</li> </ul>	
c. Thoughts or speech content  0= Inaccessible, apparently psychotic or deluded  1= Suspicion of abnormality only  2= Normal	
d. Abnormal experiences / hallucinations 0= Present 1= Not present / no evidence present 2= Normal NA	
e. Rapport 0= Very poor 1= Normal	
List any drugs being taken for mood or mental disorder(s):	
	MS Total /8