## A-T Neuro Examination Scale Toolkit (A-TNEST)

## 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 $\quad$ secs
$0=<1 / 4$ lower limit of normal (LLN)
$1=\quad>1 / 4$ LLN, $<2 / 3$ LLN
2= $\quad>2 / 3$ LLN, $<$ LLN
3= $\geq$ LLN
NA
C.b. Loudness in conversation
$0=\quad$ Significantly impaired
1= Mildly impaired
2= Normal
NA

## C.c. Articulation

$0=\quad$ Significantly impaired
1= Mildly impaired
3= Normal
NA
C.d. Prosody
$0=\quad$ Significantly impaired
1= Mildly impaired
2= Normal
NA

## C.e. Vocal Pitch

$0=\quad$ Significantly impaired
1= Mildly impaired
2= Normal

## C.f. Phonation breaks in conversation

$0=\quad$ Significantly impaired
1= Mildly impaired
2= Normal
NA

## C.g. Response time

$0=\quad$ Significantly impaired
1= Mildly impaired
2= Normal
NA

## C.h. Rate

$0=\quad$ Significantly impaired
1= Mildly impaired
2= Normal
NA

## C.i. Speech Intelligibility

$0=\quad$ 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)
$4=\quad$ Meaning understood, some phrases missed (adequate when predictable or in known context)
6= Meaning understood, some phrases missed (adequate when unpredictable even when context is not known)
8= $\quad$ Normal (adequate for age even when unpredictable and context is not known)
NA
Speech by Observation
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.

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## 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.
Sample of short phrase for size and position (school age and older; 3-10 words in line)
0= None
1= Single character
2= More than half words with character variation (size or position)
3= Less than half words with character variation (size or position)
4= All words with consistent character size and variation
NA
or
Sample of name (younger primary school age)
0= None
1= Scribble
2= Some legible but less than half the letters
3= More than half the letters legible
4= All letters legible
NA
or
Copy of primary shapes (pre-school or younger than school age)
$0=\quad$ None
$1=$ severely impaired, very slow
3= mildly impaired and or slow
4= normal for age

## Primary shapes

NA
Raw C $\square$ /30

Total C $\square$ /15

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## 2. E Eye movements video if possible

## E.a. Conjugate gaze

$0=\quad$ 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 I Accommodation (if dysconjugate examine with each eye covered in turn)
$0=$ Absent
1= Impaired but either transient or limited
$2=\quad$ Normal
NA
E.e. Off-foveal gaze tendency (vertical or horizontal)
$0=\quad$ Frequent and persistent
1= Present only with certain activities
$2=\quad$ Not present (normal)
NA
E.f. Pursuit (away from primary gaze)
$0=\quad$ No pursuit velocity movement generated
1= Fragmentary
2= Normal
NA
E.g. Saccade amplitude (head fixed, horizontal gaze-shift of 30 degrees)
$0=\quad$ 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

$0=\quad$ All or most delayed
1= Less than 50\% delayed
2= Normal latency
NA
E.i. Oculo-Motor Apraxia (large amplitude lateral gaze-shifts)

0= Present on all gaze shifts
$1=\quad$ Present on most (>50\%) gaze shifts
2= Occasionally present
3= Not present
NA
E.j. Up Gaze restriction
$0=\quad$ No vertical gaze
1= Limited vertical gaze
$2=\quad$ Normal vertical gaze
NA

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

0= Persistent
$1=\quad$ 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
$2=\quad$ Normal
NA
E.n. Post-rotatory nystagmus (after slow rotation, 6 full rotation, $6 \mathrm{sec} /$ rotation)
$0=$ Persists more than 10 secs
$1=\quad$ Present, but persists no more than 10 secs
$2=\quad$ Not present
NA
E.o. Periodic Alternating Nystagmus (after slow rotation, 6 full rotation, $6 \mathrm{sec} /$ rotation)(score only if nystagmus present in primary gaze)
0= Present
1= Not present
NA
Raw E $\square$


## 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
$0=\quad 0=\quad$ On target or mid phalanx $2 / 10$ (or $25 \%$ ) times or less, or mean 5 sec or longer for each
$1=\quad 1=\quad$ On target or mid phalanx $3 / 10(25 \%)$ or more, and less than 5 sec mean
$2=\quad 2=\quad$ On target or mid phalanx $8 / 10(75 \%)$ or more
$3=\quad 3=\quad$ On target $8 / 10(75 \%)$ or more
$4=\quad 4=\quad$ On target 10/10 (100\%) but with some in-coordination
$5=\quad 5=\quad$ 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)
$0=\quad$ Requires total support
$1=\quad$ Requires only flat posterior support
$2=\quad$ Sits with self support of arms in lap or laterally
$3=\quad$ No support but sways markedly (raises body part for balance)
4= No support but sways slightly /occasionally
$5=\quad$ 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)
$0=\quad$ unable to stand without vertical support
$1=\quad$ 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 ( $\geq 75 \%$ of trials)
$2=\quad$ 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 ( $\geq 75 \%$ of trials)
$3=\quad$ stands independently without support but requires corrective steps, 30 seconds ( $\geq 75 \%$ of trials)

4=
$5=$ feet together, minor excessive corrective movement but no corrective steps, 30 seconds ( $\geq 75 \%$ of trials)
6= feet together, no corrective movements, 30 seconds
NA

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A.d. Walking ( 10 meters)
$0=\quad$ Requires vertical support
1= Massive lateral support (above elbow or waist held)
2= Some lateral support (below elbow or hand held, elbow relaxed)
3= Only touch support needed
4= No support, wide base or takes corrective stagger steps
5= No support, normal speed with mild path deviations or corrective steps
$6=\quad$ Normal path width without corrective steps
NA

## 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)

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
NA

## M.b. Body hypokinesia

$0=\quad$ Slowness and poverty of movements 1= Normal
Total A $\square$| 27
Running total C E A $\square$ / 57

Hyperkinetic movements (tics, chorea, myoclonus, ballismus, arrhythmic adventitious movements) M.c. Distal / hand adventitious movements - not rhythmic / tremor (limbs) (score worse side)
$0=\quad$ Present at rest
$1=\quad$ Present with both social / cognitive and motor activation
2= Present with either social / cognitive or motor activation
$3=\quad$ Not present
NA

## M.d. Retro-Colic Spasms / Jerks

$0=\quad$ Present with all postures at rest
$1=\quad$ Present sitting posture with hand support
2= Present with both social /cognitive and motor and posture activation / removal of head rest
3= Present with either social /cognitive or motor or posture activation / removal of head rest
4= Not present
NA
M.e. Other proximal adventitious movements - not rhythmic / tremor (face, head, trunk)
$0=\quad$ Present at rest
1= Present with both social / cognitive and motor activation
$2=\quad$ Present with either social / cognitive or motor activation
3= Not present
NA
M.f. Tremor / rhythmic movements (while sitting with usual support)
$0=\quad$ Present at rest / without activation
1= Present with both social /cognitive and motor activation
2= Present with either social /cognitive or motor activation
3= Not present
NA
M.g. Tremor / rhythmic movements (while sitting with respect to support)
$0=\quad$ Present with full support
1= Present with removal of head rest / support
2= Present with removal of trunk support
3= Not present

## Dystonia / posture distortions / slow movements

M.h. Facial expression / timing and duration of large expressions
$0=$ Limited or absent emotion expression (mask-like or expressionless face)
1=Hypomimea / abnormal persistence (dystonic facial expression / smile too sustained / mouth open)
2=Normal
NA

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M.i. Tongue lateral movements (on command with mouth open)
$0=\quad$ No lateral movements (moves only with mandible)
$1=\quad$ Lateral movements depend on some mandible movement
2= Lateral movements independent of mandible but impaired
3= Normal lateral movements
NA
M.j. Limb posturing / dystonia (score worse side)
$0=\quad$ Present at rest / without activation
1= Present with both social /cognitive and motor activation
2= Present with either social /cognitive or motor activation
3= Not present
NA
M.k. Head / trunk posturing / dystonia
$0=\quad$ Present at rest / without activation
1= Present with both social /cognitive and motor activation
$2=\quad$ Present with either social /cognitive or motor activation
3= Not present
NA

Dystonia $\square / 11$
Total M $\quad$ / 33

Running total C E A M $\square$ 190

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5. P Power (document any muscle weakness in box, score weakest side)
$\left.\begin{array}{lllll}\text { P.a. Ankle Dorsiflexion } & 0= & \begin{array}{l}\text { Severe weakness } \\ 1= \\ \text { Mild weakness }\end{array} & \begin{array}{l}\text { (MRC 0, 1, or 2) } \\ \text { (MRC 3, or 4) } \\ (\text { MRC 5) }\end{array} \\ 2= & \text { Normal for age }\end{array}\right)$

Document muscle weakness:

| P.f. Hand muscles | $0=$ | Claw hand (extension of all 4 MCP joints at rest) |
| :--- | :--- | :--- |
| $1=$ | Thenar $/$ hypothenar wasting |  |
| $2=$ | Normal |  |
|  | NA |  |

P.g. Document foot deformity:
P.h. Document spinal deformity:



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6. N Neuropathy (tendon reflexes present on either side)

| N.a. | Ankle tendon reflexes |
| :--- | :--- |
| $0=$ | Absent |
| $1=$ | Present |
| NA |  |

N.a. Ankle tendon reflexes

0= Absent
NA
N.c. Biceps tendon reflexes
$0=\quad$ Absent
1= Present
NA
N.d. Proprioception in toes (if unable to answer position questions in proximal joints, attempt vibration sense with 128 Hz tuning fork or score as null "NA")
$0=\quad$ Absent movement sensibility
$1=\quad$ Movement sensibility intact, position errors $\mathbf{>} \mathbf{2 5 \%}$
$2=\quad$ Rare position errors
$3=\quad$ No position errors / normal for age
NA
OR
N.dv. Vibration sense at ankles (only if proprioception not reliable)
$0=\quad$ Absent vibration sense
$1=\quad$ Some vibration sense intact, errors $>25 \%$
2= Rare errors
$3=\quad$ Normal for age
NA


## GROWTH / NUTRITION, SWALLOWING AND FEEDING SUPPLEMENTAL DOMAINS

7. Gr Growth (4 points)

Height: cm (centile:
Weight: $\quad \mathrm{kg}$ (centile:
BMI : $\quad \mathrm{kg} / \mathrm{m}^{2}$ (centile:
0 : < 0.4 th centile
1: 0.4 th $<2$ nd centile
2: 2 th $<9$ th centile
4: 9th centile and heavier

Head: cm (centile: )
) OR Arm span: cm )
) OR Weight for Height:
0 : $<2^{\text {nd }}$ centile
1: $2^{\text {nd }}<9^{\text {th }}$ centile
2: $9^{\text {th }}<25^{\text {th }}$ centile
4: $25^{\text {th }}$ centile +

OR BMI SDS
$0:>-2.67$ SDS
1: $-2.67>-2.0$ SDS
2: $-2.0>-1.33$ SDS
4: -1.33 SDS +
Total Gr

8. Nu Nutrition (score by observation or report, 20 points)

Feeding tube $\mathrm{Y} \bigsqcup \mathrm{N} \square$
F.a. Drooling (NA <4 years of age)
$0=\quad$ Drooling all the time
1= Drools only when tired and concentrating
2= Drools only when tired or concentrating
3= Saliva accumulates at sides of mouth
4= None
NA

## F.c. Drinking

$0=\quad$ Cough with all liquid intake
1= Cough when drinking thin and thick liquids by straw
2= Cough when drinking thin liquids by straw
3= Cough when drinking liquids by open cup
4= Normal for age

## F.b. Feeding

$0=\quad$ No functional self-feeding
$1=\quad$ Needs help with some feeding, even with finger feeding
2= Needs help with some feeding, even with all utensils
$3=\quad$ Needs help with some feeding, even with spoon
4= Independent feeding only with adaptations
5= Normal for age
NA

## F.d. Swallowing / Diet Status

$0=\quad$ Nothing by mouth
1= Tube dependent with minimal oral tastes
2= Tube dependent with consistent oral intake of specific liquids or foods
$3=\quad$ Partial oral feeding with tube supplementation for liquids or nutrition, and oral feedings with specific liquid/food limitations or special preparations
4= Partial oral feeding with tube supplementation for liquids or nutrition, and oral feedings without specific liquid/food limitations or special preparations
$5=\quad$ Total oral feedings with specific liquid/food limitations or special preparations
$6=\quad$ Total oral feedings with caregiver use of specific feeding routines
7= Total oral feedings without any restrictions


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## LEARNING ABILITY / MENTAL STATE SUPPLEMENTAL DOMAINS

9. LA Learning Ability / Cognitive (score by Self Report or Carer Report, and confirm if possible by observation, 3 points)

0= Profound or severe cognitive impairment likely
Observed

$1=$ Moderate cognitive impairment likely
2= Mild global cognitive impairment likely
$3=$ Normal or superior cognition, or just specific learning difficulties NA

Total LA $\square$ 13
10. MS Mental State (score by clinical impression, 8 points)
a. Appearance
$0=$ Impaired
$1=$ Normal
NA
b. Mood

0= Severe abnormality; severely manic or depressed
1= Moderately depressed, or flat affect, or moderately manic or hypomanic
$2=$ No abnormality detected, a range of emotions was expressed
NA
c. Thoughts or speech content
$0=$ Inaccessible, apparently psychotic or deluded
1= Suspicion of abnormality only
2= Normal
d. Abnormal experiences I 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):
$\square$

