

Neurologic Outcome Assessment in Childhood Acquired Brain Injury

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Primary Neurologic Outcomes

- Mortality due to neurologic injury
 - Brain death
 - Somatic death following termination of life support due to profound brain injury (usually global HIE or massive hemorrhage)
- Neurologic events during acute hospital course
 - Seizures
 - Ischemic stroke
 - Intracranial hemorrhage
 - Hypoxic-ischemic encephalopathy
 - CNS infection
 - Hydrocephalus
- Post-acute neurologic sequelae of acute illness
 - Recurrent stroke
 - Disability/Impairment
 - Epilepsy
 - ADD/ADHD
 - Movement disorder

Psychosocial Outcomes Secondary to Neurologic Morbidity

- Child:
 - Impaired educational attainment, need for education support
 - Impaired socialization
 - Impaired recreation participation
- Family:
 - Altered child-parent and sib-sib relationships and home life
 - Financial and employment distress
 - Learning to negotiate medical and rehabilitation systems
 - Near term educational advocacy
 - Long-term planning for altered dependency
- Society:
 - Health care costs (length of stay, rehabilitation)
 - Educational costs – need for special services and placement
 - Opportunity costs – lost income production by family & patient, social comorbidities

Outcome: domains & measurement

OUTCOME

MEASUREMENT PROPERTIES

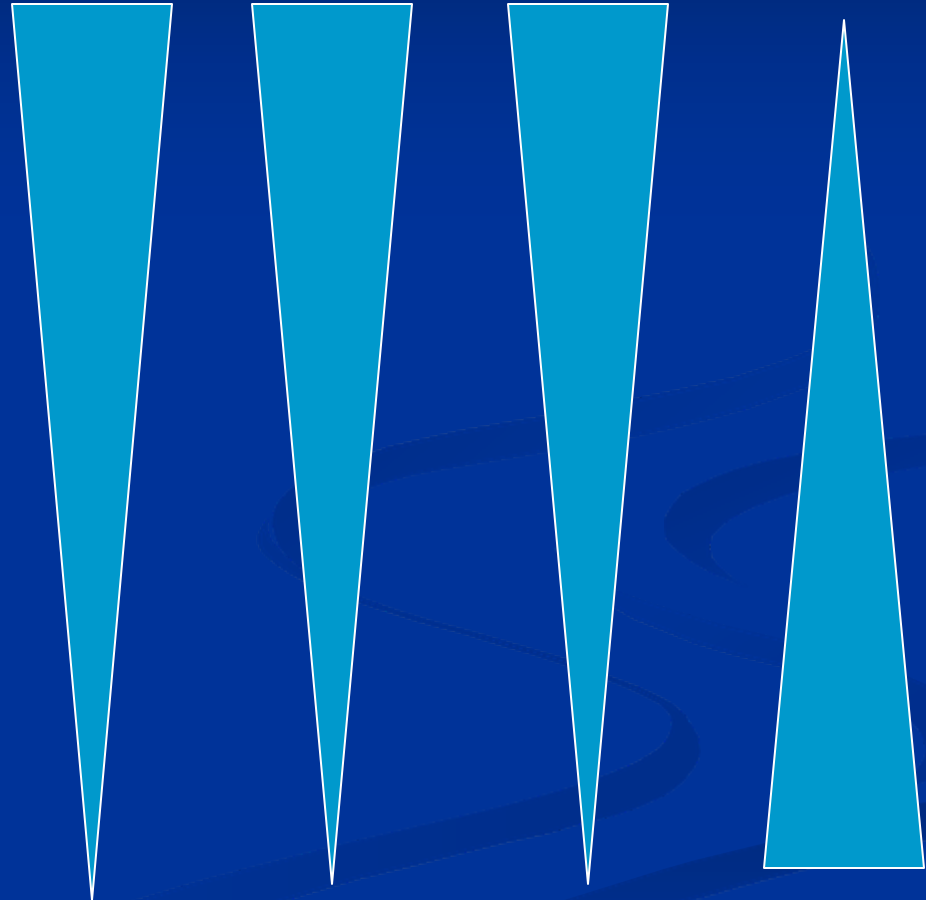
Mechanistic Accuracy Objectivity Relevance

Pathology: % cell death, infarct volume, altered brain development – **example MRI**

Impaired function: Neurologic deficit, developmental deficits – **example NIHSS**

Limited performance: Loss of mobility, ADL, communication, learning – **example PEDI**

Restricted participation: Altered participation in school, family & social functions – **Example PedsQL**



Primary Neurologic Outcomes: How to Measure?

- Mortality due to neurologic injury
 - Brain death
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“Hard Endpoints” -

Assure event reporting:

1. Clear definitions
2. Prospective ascertainment
3. Adjudication?

Primary Neurologic Outcomes: How to Measure?

- Mortality due to neurologic injury
 - Brain death
 - Somatic death following termination of life support due

Assure event assessment & reporting:

1. Clear definitions – criteria including clinical, radiologic, EEG
2. Mandate neurologic assessment & event characterization
3. Prospective ascertainment & contemporaneous with clinical event
4. Other issues:
 1. Event adjudication?
 2. Measure event severity?
 3. Characterize & adjudicate causal factors?

Neurologic events during acute course: How to Detect?

Mandate surveillance vs. await symptoms/signs?

Event	Typical presentation
Seizures	Observation of paroxysmal activity by bedside caregivers, confirm dx by <u>neurological consultation + EEG</u>
Ischemic stroke & Intracranial hemorrhage	Sudden appearance of new focal deficit, focal seizures or altered LOC, confirmed by <u>neurological consultation and imaging.</u> May not be evident until cessation of sedating drugs. Parents often the most sensitive reporters of change.
Hypoxic-ischemic encephalopathy	Failure to recover baseline LOC, usually after cardioresp decompensation/arrest, \pm seizures. Confirm by <u>neurological consultation + imaging + EEG + lab testing to r/o other causes of symptoms.</u>
CNS infection	Fever + change in baseline neurologic function. Confirm by <u>imaging + lab testing.</u>
Hydrocephalus	Signs/symptoms of high ICP, confirm by <u>neurosurgical consultation + imaging.</u>

Neurologic events during acute course: How to Measure & Classify?

- Instruments to capture/characterize **acute** neurologic events
 - Seizures – no adequate method exists to classify
 - Ischemic stroke & intracranial hemorrhage – diagnosis by neurological consult, then classify severity & type by standard instruments
 - GCS – standardized method of describing decreased LOC
 - PedNIHSS – semiquantitative measure of deficit severity, not useful < age 1 yr
 - Neuroimaging (grading scheme of subtype and size, e.g. “small, medium, large”?)
 - Risk factor assessment & causal attribution?
 - Hypoxic-ischemic encephalopathy
 - Duration & depth (GCS) of post-anoxic coma?
 - Neurologic exam at specified interval?
 - Global functional status at specified interval and at hospital discharge?
 - CNS infection – qualitative reporting
 - Hydrocephalus – qualitative reporting

ALL TYPES OF EVENTS REQUIRE STANDARD CLINICAL NEUROLOGICAL CONSULTATION WITH STANDARDIZED REPORTING OF FINDINGS IN FORMAT THAT ALLOWS ASCERTAINMENT OF TYPE OF EVENT, AND - DEPENDING ON STUDY REQUIREMENT - SEVERITY OF EVENT.

Neurologic events during acute course: How to Measure & Classify?

- Standardized recording of neurologic exam & interpretation and rating the findings
 - GCS – for gross measure of coma severity, and level of depressed consciousness
 - PedNIHSS – to characterize severity of acute focal neurologic deficit due to infarct or parenchymal hemorrhage (validation in progress)
 - SITT Neurological consultation report –
 - Detailed recording of neurologic exam
 - Could be used in acute or post-acute phase
 - Includes a summary/impression from PSOM to rate the severity of deficits
 - Includes an interpretation as to nature of event
 - THAPCA Neurological exam report – similar to SITT exam, adapted to capture bilateral & diffuse deficits on exam in the post-acute phase
 - PSOM – detailed recording of neurologic exam in the post-acute phase & a final summary of impressions providing a rating on scale 0-10 of severity of impairment

Neurologic status in post-acute stage: How to Measure & Classify?

- Standardized recording of neurologic exam & interpretation and rating the findings
 - SITT Neurological consultation report – as described
 - THAPCA Neurological exam report – as described
 - PSOM – as described
 - Global functional rating scales –
 - KOSCHI – five-point scale, accuracy dependent on qualifications & procedure used by examiner
 - PCPC – similar to KOSCHI
 - Pediatric Stroke Activity Limitation Measure

Pediatric Stroke Outcome Measure (PSOM)

- Design: Items derived from STOP trial (stroke in Sickle Cell anemia) for and tested in over 600 children with arterial ischemic stroke or sinovenous thrombosis in the Toronto Children's Stroke Clinic
- Reliability: inter-rater reliability on small subset $R=0.94$ for composite summary score
- Validity: significant correlations with Vineland & COPT scales, & WISC-FS IQ

Score each modality by severity as:

0 = no deficit

0.5 = minimal deficit without functional impairment

1 = moderate deficit with functional impairment

2 = severe deficit with absent function

Item
Sensorimotor deficit, right
Sensorimotor deficit, left
Language deficit, expressive
Language deficit, receptive
Cognitive or behavioral deficit
TOTAL (range 0-10)

Pediatric Stroke Activity Limitation Measure

Gordon et al “Functional outcome following stroke in children”

J Ch Neurol 2002;17:429 (London group)

- Derived from items on the PEDI and Vineland
- Six domains: gross motor, fine motor, self care, communication, social/emotional, education
- Each item scored 0-3, 0 performed without difficulty, 1 some difficulty, 2 great difficulty, 3 can't perform
- Scores for all domains were averaged to give overall score of 0-3 for all domains
- Reliability data: two examiners independently assessed each pt; Cohen's kappa = 0.89 for overall score in sample of 17 pts

Pediatric Stroke Activity Limitation Measure

Item	Score (0-3)
Gross Motor: Stand up from chair and sit down	
Walking across a room	
Lifting & carrying large box with 2 hands	
Lifting & carrying beaker with 1 hand	
Fine Motor: Holding/moving a pencil	
Picking up "chocolate button"	
Taking screwtop lid off jar using 2 hands	
Self-Care: Bathing	
Toileting	
Feeding	
Washing	
Dressing	
Communication: Follows spoken or written first language	
Expresses complex ideas verbally or nonverbally	
Social/emotional: Interacts appropriately with others	
Able to use prior experience to solve new problems	
Remembers familiar people & routines	
Education: Keeps up with peers at school academically	
Keeps up with peers at school physically	

KOSCHI* – King's Outcome Scale for Childhood Head Injury

- Proposed brief functional outcome classification system for childhood acute brain injury, adapted from Glasgow Outcome Scale - stroke, intracranial hemorrhage, TBI.
- Intended to resemble the grading system used in the modified Rankin scale used in adult stroke.
- used easily and successfully in pediatric intracranial hemorrhage studies (Lori Jordan at Johns Hopkins, Lauren Beslow at CHOP), and compliments very nicely the other outcome measures we use to stratify functional outcome in a global way.
- Currently being used in PedNIHSS validation study (Ichord et al) to compliment PSOM, based on findings of a complete pediatric neurological consultation & exam.

*Crouchman M, Rossiter L, Colaco T, Forsyth R: A practical outcome scale for paediatric head injury. Arch Dis Child 2001; 84(2): 120-124.

Score	Definition KOSCHI
1	DEATH
2	VEGETATIVE The child is breathing spontaneously and may have sleep/wake cycles. He may have non-purposeful or reflex movements of limbs or eyes. There is no evidence of ability to communicate verbally or non-verbally or to respond to commands.
3	<p>SEVERE DISABILITY</p> <p>(3a) The child is at least intermittently able to move part of the body/eyes to command or make purposeful spontaneous movements; for example, confused child pulling at nasogastric tube, lashing out at carers, rolling over in bed. May be fully conscious and able to communicate but not yet able to carry out any self care activities such as feeding.</p> <p>(3b) Implies a continuing high level of dependency, but the child can assist in daily activities; for example, can feed self or walk with assistance or help to place items of clothing. Such a child is fully conscious but may still have a degree of post-traumatic amnesia.</p>
4	<p>MODERATE DISABILITY</p> <p>(4a) The child is mostly independent but needs a degree of supervision/actual help for physical or behavioural problems.</p> <p>(4b) The child is age appropriately independent but has residual problems with learning/behaviour or neurological sequelae affecting function.</p>
5	<p>GOOD RECOVERY</p> <p>(5a) Minor abnormalities having minimal impact on function, e.g. headaches, well controlled epilepsy, minor exam abnormalities.</p> <p>(5b) Implies that the information available is that the child has made a complete recovery with no detectable sequelae from the head injury.</p>

Neurological Outcome Scoring for Pediatric Resuscitation after Cardiac Arrest (PRCA score)

THAPCA (Therapeutic Hypothermia after
Pediatric Cardiac Arrest)

Neurology investigators:

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Approach

- The basic approach is for a pediatric neurologist to perform a conventional, detailed age-appropriate neurological exam at 12 months after cardiac arrest
- The examiner records the exam systematically and assigns a Likert-style score to each element of the exam
- A quantifiable representation of the exam is captured in two ways in the recording form:
 - Global assessment score: assigned as a global clinical impression by the examiner, with 6 subscores, each capturing abnormalities in major domains of function, range 0 -21.
 - Total cumulative normalized score: sum of all item scores normalized on a 0-100 scale

Scoring: Global assessment score (0-21)

- This reflects overall assessment of function in several major domains (sensorimotor, language, cognition, behavior)
- It reflects global clinical judgment of the examiner, rather than a mathematically sum of item scores
- This score is weighted heavily to the assessment of language and cognition
- This score reflects assumptions about domains that are most likely to be adversely affected by hypoxic-ischemic brain injury (i.e. higher cortical function)

Scoring: Total normalized cumulative abnormality score (0-100)

- The examiner scores each exam element
 - Normal - 0
 - Abnormal mild - 1
 - Abnormal moderate - 2
 - Abnormal severe - 3
 - Not done

- Normalized cumulative score =

$$\frac{\text{Total abnormal score values}}{3 \times \text{total number of items scored}} \times 100$$

- In our scoring plan, we will only analyze elements that were scored (“not done – doesn’t count”)

Forms

- Infant version – for up to age 3 (up to third birthday) at time of evaluation
- Child version – 3 to 18 years old at time of evaluation

Using the forms 1.

- The forms will have an introduction page with instructions on scoring
- The forms will be easy to fill in
- The forms will have sufficient room for adding narrative comments
- The site co-ordinator will print an age-appropriate form, and provide it to the designated neurologist

Using the forms 2.

- We expect neurologists to fill in the forms and global assessment scores, and return them to co-ordinators
- Co-ordinators will submit these data to the DCC
- The neurologist will be expected to generate a clinical report per local institutional standards to provide information from the exam to the PCP with a copy to the family (we will not collect these letters)

Next

- Let's look at the forms.



PRCA NEUROLOGICAL OUTCOME MEASURE - INFANT VERSION (Up to 3 years old)

STANDARDIZED NEUROLOGICAL EXAM modified from PSOM-SNE (Revised 5.03.07)

Revised 4.13.09

IDENTIFYING DATA

ID# _____ Site: _____ Date of assessment (yyyy-mm-dd): _____

Date of birth: (yyyy-mm-dd): _____ Location of Assessment: In-patient Out-patient Clinic

SCORING INSTRUCTIONS:

Each item can be scored as Normal, Abnormal, or Not Done (includes items that are not age-appropriate or that were not assessed). For abnormal items, assess and score severity of abnormality (1- mild; 2- moderate; 3-severe), based on your best clinical judgment. Guidelines for Scoring are given as suggestions, and need not be viewed as absolute criteria for scoring.

LEVEL OF CONSCIOUSNESS

TEST ITEM	Normal	Abnormal	Notes
Level Of Consciousness			

BEHAVIOR, MENTAL STATUS

TEST ITEMS	Normal	Abnormal	Not Done	Guidelines for Scoring
Activity Level				Excessively quiet, hyperresponsive, fidgety – age dependent
Interpersonal Interaction				With parents and examiner
Cooperation				Age dependent
Attention				Age dependent
Affect				Extremely shy, withdrawn totally flat, gaze avoidance
Object Permanence				

LANGUAGE

TEST ITEMS	Normal	Abnormal	Not Done	Guidelines for Scoring
Receptive language				6 mo orients to sound 10 mo inhibits to "no" 12 mo one-step command with gesture 16 mo one-step command without gesture, points to body parts 24 mo two-step commands, points to named pictures
Expressive language development				By 12 mos. - 1-2 words 12-18 mos. - 10-20 single words 2 years. - 50 words, 2 word phrase 3 years - 200 words, 3+ word sentences
Comments				

CRANIAL NERVES

TEST ITEMS	Normal	Abnormal	Not Done	Guidelines for Scoring
Visual Fields / Vision	Right			Facing patient at 2-3 ft encourage to stare at your eyes and tell when they see object come into view from side (or note gaze shifting toward object)
	Left			
Pupillary Light Reflex	Right			Direct and Consensual
	Left			
Funduscopy	Right			Note Abnormalities:
	Left			
Ocular Motility	Right			Move pen or red object or light smoothly from right to left and back testing full range. Watch for nystagmus or dysconjugate eye movements
	Left			
Facial Movements	Right			Observe smile, observe mouth symmetry during vocalization. Listen to speech quality, observe eye closure for symmetry
	Left			
Hearing	Right			Finger-rub for infants or whisper at 2 - 3 feet away.
	Left			
Swallow				Based on history or observation
Palate and gag	Right			Observe during open mouth crying or demonstrate with tongue protruded . 'Say 'ahhhhh.' Listen to voice quality
	Left			
Head/neck control				
Handling secretions				

GROSS MOTOR Testing

Developmental Gross Motor Use this table only for children who cannot walk independently

TEST ITEMS – Posture & Mobility Skills	Normal	Abnormal	Not Done	Comments
Central Tone: Head lag on 'pull-to-sit'				
Central Tone: Slip Thru On Vertical Suspension				
Central Tone: Ventral Suspension				
Rolls Over (Front To Back)				
Rolls Over (Back To Front)				
Sits Alone				
Moves From Laying To Sitting Unassisted				
Weight-Bearing, Supported				
Walks Holding On				
Walks Independently				

	POWER AND TONE				INVOLUNTARY MOVEMENTS*	
	Normal	Abnormal	Not Tested	Comments	Normal (None)	Abnormal (Present)
Neck/Trunk Muscles						
Right Arm						
Proximal						
Distal						
Left Arm						
Proximal						
Distal						
Right Leg						
Proximal						
Distal						
Left Leg						
Proximal						
Distal						

***Type(s) of Involuntary Movements Seen:** Check all that are present

TYPE	COMMENT
Head/neck tremor	
Limb Tremor	
Choreoathetosis	
Dystonic Posturing	
Tics	
Myoclonus	

TENDON REFLEXES

TEST ITEMS		Normal	Abnormal	Not Done	Comments
Biceps	Right				
	Left				
Triceps	Right				
	Left				
Quadriceps	Right				
	Left				
Ankle Jerk	Right				
	Left				
Babinski	Right				
	Left				
Elicited ankle clonus	Right				
	Left				

FINE MOTOR / COORDINATION

TEST ITEMS		Normal	Abnormal	Not Done	Guidelines for Scoring
Pincer Grasp	Right				Encourage to pick up small 2–3 mm ball of rolled up paper
	Left				
Reaching for object	Right				Observe for unusual or asymmetric tremor on reaching for object
	Left				
Sitting Balance					
Standing Balance					
Comments:					

SENSORY

TEST ITEMS		Normal	Abnormal	Not Done	Comments
Light Touch	Right				
	Left				
Pin Prick or Cold	Right				
	Left				

GAIT

Test Only if walking without support

TEST ITEMS	Normal	Abnormal	Not Done *	Comments
Gait - Walking				
Gait - Running				
Climbing up 5 stairs				

*Not Done includes skills that are not developmentally appropriate for the child's current age.

Other comments:

GLOBAL Assessment Score.

Summarize and grade your impressions in the following 7 categories. Circle the score in each category that best represents your final impression of your exam findings:

A. Sensorimotor Deficit - score each side separately

	<u>A1.Left side</u>	<u>A2.Right side</u>
None	0	0
Mild but no impact on function	1	1
Moderate with some functional limitations	2	2
Severe or Profound with missing function	3	3

For items scored in Category A1 and A2, identify all the of the types of Sensorimotor Deficits that you observed:

- Abnormality of tone
- Quadriplegia
- Hemiparesis
- Sensory deficit
- Global delay in gross motor skill attainment
- Global delay in fine motor skill attainment

B. Other motor or sensory deficit (includes cranial nerve deficits)

None	0
Mild but no impact on function	1
Moderate with some functional limitations	2
Severe or Profound with missing function	3

For items scored in Category B, identify all the of the types of Other Sensorimotor Deficits that you observed:

- Vision impairment
- Difficulty with drinking, chewing or swallowing
- Ataxia
- Movement disorder
- Other, describe:

C. Language Deficit – Production (include dysarthria)

None	0
Mild but no impact on function	1
Moderate with some functional limitations	2
Severe or Profound with missing function	3

D. Language Deficit - Comprehension

None	0
Mild but no impact on function	1
Moderate with some functional limitations	2
Severe or Profound with missing function	3

E. Cognitive Deficit

None	0
Mild (little impact on daily function)	1
Moderate with some functional limitations	2
Severe or Profound with missing function	3

F. Behavioural Deficit

None	0
Mild (little impact on daily function)	1
Moderate with some functional limitations	2
Severe or Profound with missing function	3

For Categories E and F, describe the Cognitive or Behavioral Deficits that you observed:

Other comments regarding scoring:

TOTAL SCORING: _____ / 21

PEDI

Pediatric Evaluation of Disability Inventory

- Measure of functional capability in three domains:
 - Self care
 - Mobility
 - Social function
- Measured in three ways:
 - Direct: Functional Skill Scale
 - Indirect: Level of Caregiver Assistance
 - Assistive Technology Needs: Modifications Scale
- Age level measured: 0.5 – 7.5 years (or equivalent developmental level)

PEDI

Validation & Standardization

- Normative data obtained on sample of 412 healthy children.
- Tested on 60 disabled children to establish reliability ($R = 0.74-.96$).
- Validated in three groups of disabled children for ability to distinguish abnormal from normal children.
- Concurrent validity tested against other standardized scales, Batelle & WeeFIM ($R=0.67-.97$).
- Responsiveness to change was established in groups of disabled children measured at baseline and 6-8 months later. Expected direction and amplitude of change was detected.

PEDI

Administration & Scoring

- Who: Ideally, the parent, or someone familiar with the child's typical performance across all areas and in multiple settings.
- How:
 - Parental structured interview:
 - Part I: Functional Skills – most parents can complete on their own, and answers reviewed & checked by interviewer
 - Part II: Caregiver Assistance – requires structured interview
 - Part III: Modifications – requires structured interview
 - Professional team administration: completion by team members in a rehab or special ed program
- Training: : review manual & understand item criteria; complete sample cases in manual; observation of two “live” interviews by a another examiner