

# Disorders of Consciousness

## Part 1: Terminology, Assessment, and Outcome

Tuesday, April 20<sup>th</sup>, 2021

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Kennedy Krieger Institute

Johns Hopkins University, School of Medicine



**IPRC**  
International  
Pediatric Rehabilitation  
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# About our presenters

Stacy Suskauer, MD

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# Objectives

At the end of the session:

- The learner will be able to identify key features of vegetative state/unresponsive wakefulness syndrome and minimally conscious state,
- The learner will be able to describe at least two assessment measures for evaluating children with DOC,
- The learner will be able to discuss patterns of outcome among children with DOC.



# Disclosures

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## Dr. Suskauer:

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Myomo

Will discuss off label use of medications (Part II)



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# Plan

## Part 1 -- today

- Terminology and Pathology
- Assessment
- Prognosis and Outcome

## Part 2 – next week

- Interventions
- Case study



# Additional Program Description

Current Physical Medicine and Rehabilitation Reports  
<https://doi.org/10.1007/s40141-019-0214-4>

PEDIATRIC REHABILITATION MEDICINE (A HOUTROW AND M FUENTES, SECTION EDITORS)

## Rehabilitation in Children with Disorder of Consciousness

Nancy Yeh<sup>1,2</sup> • Beth S. Slomine<sup>1,2,3</sup> • Valerie Paasch<sup>1,3</sup> • Heather B. McLean<sup>1</sup> • Stacy J. Suskauer<sup>1,2,4</sup>



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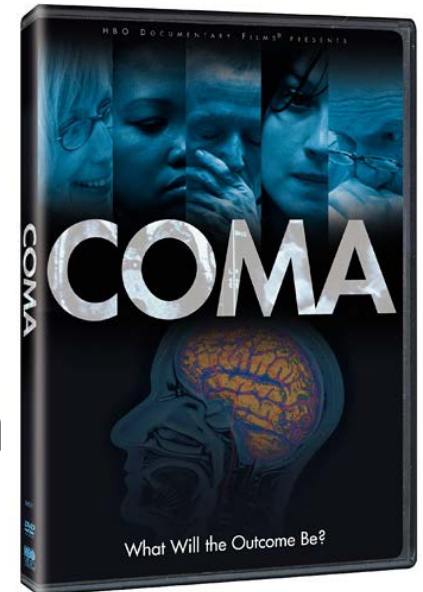
# Terminology and Pathology



# Severe Disorders of Consciousness

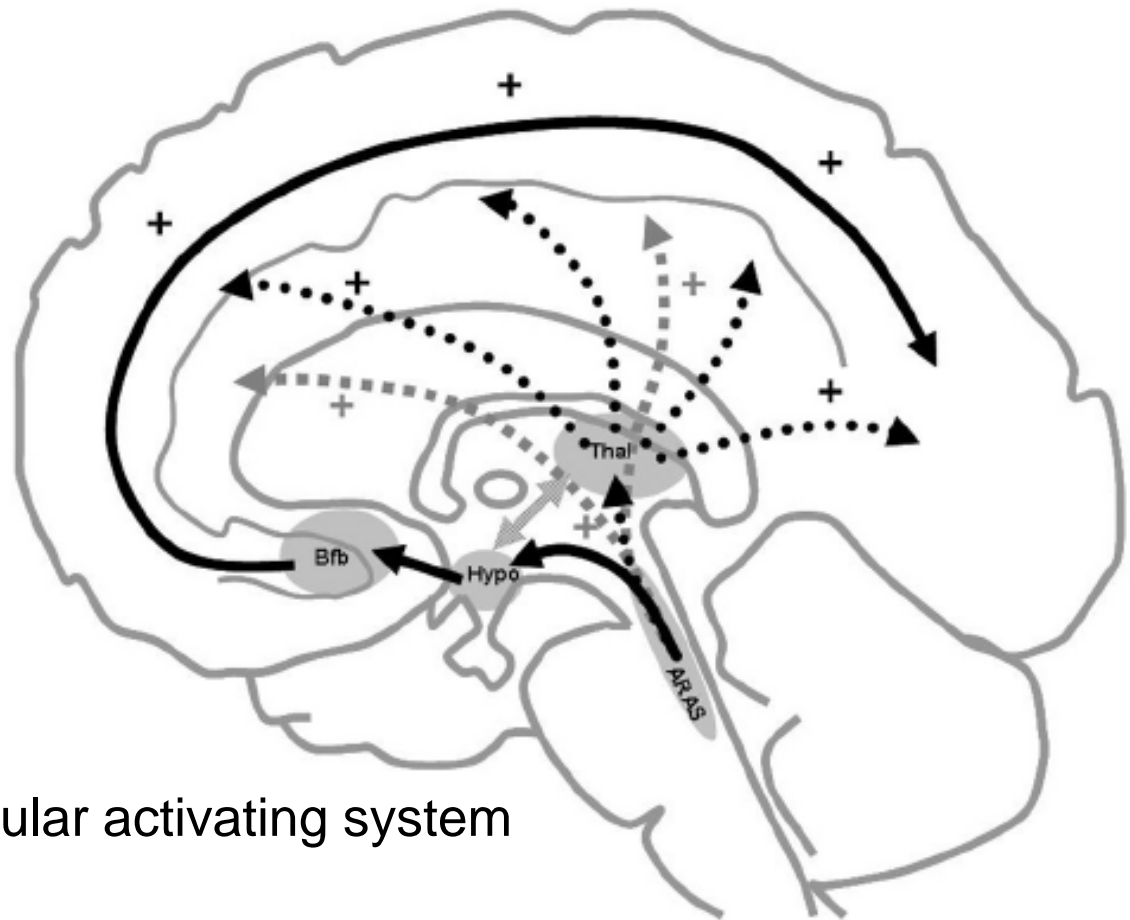
- Severely altered arousal and/or awareness of self and the environment
  - Coma
  - Vegetative State
  - Minimally Conscious State

Consensus definitions from Aspen  
Neurobehavioral Workgroup





# Anatomic structures subserving awareness and arousal



Bfb: Basal forebrain

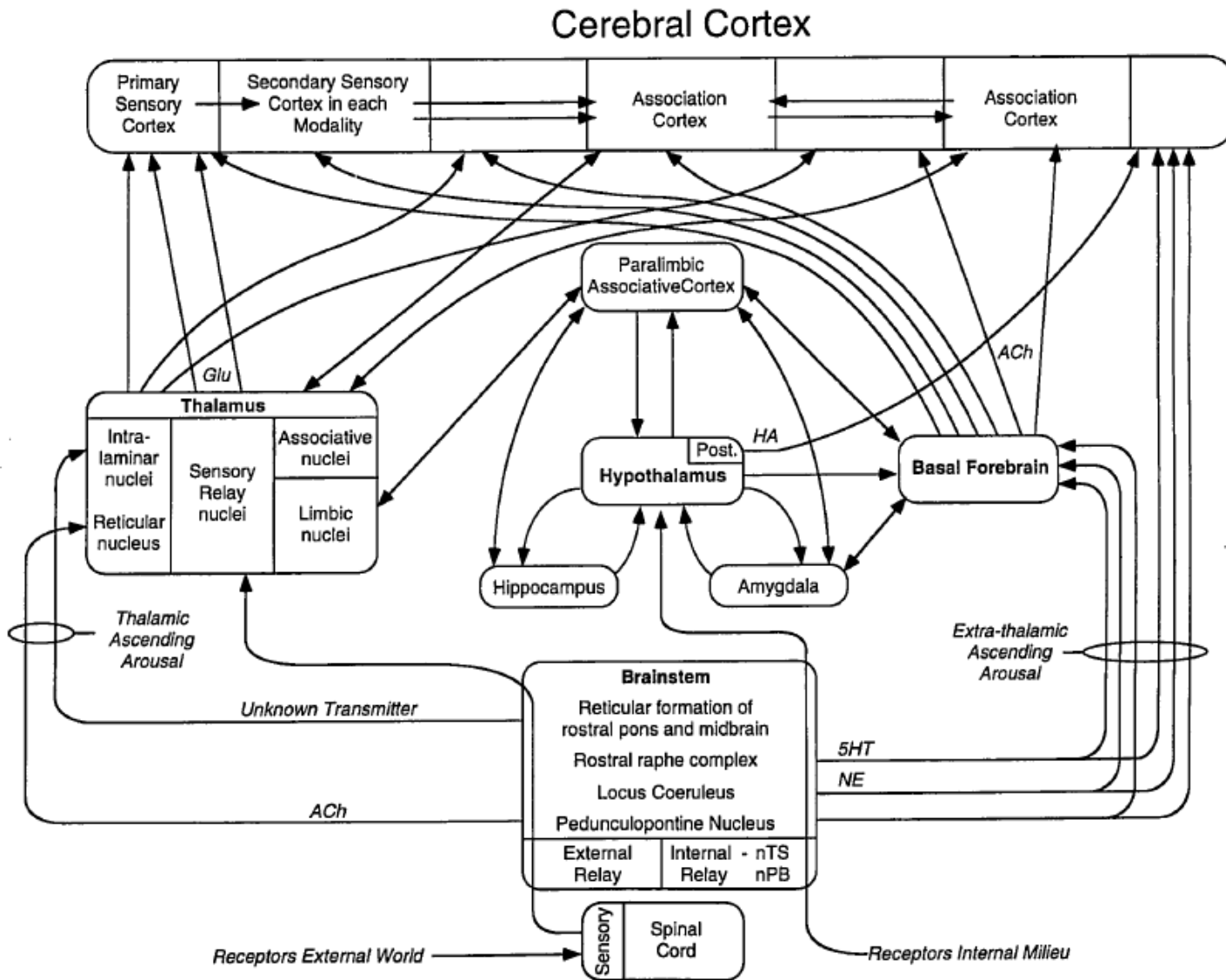
Hypo: Hypothalamus

Thal: Thalamus

ARAS: Ascending reticular activating system

Weiss et al., Critical Care, 2007

# A "simplified" view of consciousness



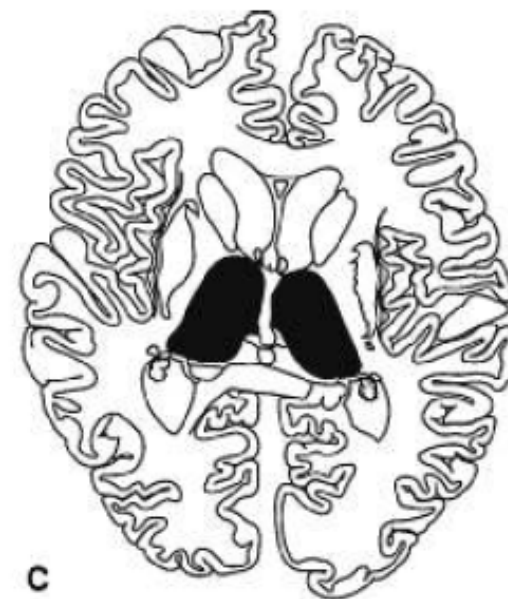
# Neuropathology of Vegetative State



A  
Diffuse Cortical Injury



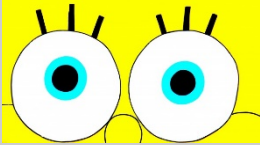




B  
Diffuse Subcortical +/-  
Brainstem Injury



C  
Thalamic Injury

figure from Kinney and Samuels, J Neuropath and Exp Neuro 1994

# Diagnostic Criteria

State	Coma	Vegetative/ Unresponsive Wakefulness	Minimally Conscious	Conscious
Sleep/wake cycles 	No	Yes	Yes	Yes
Purposeful/voluntary behavioral responses 	No	No	Yes	Yes
Consistent yes/no OR Functional object use   	No	No	No	Yes

# Newer terminology

Vegetative State =  
“Unresponsive Wakefulness Syndrome”

Prolonged VS/UWS:

Describe as:

Vegetative State +  
Etiology +  
Duration

Eliminate use of:

“Persistent Vegetative State”  
“Permanent Vegetative State”



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# More newer terminology

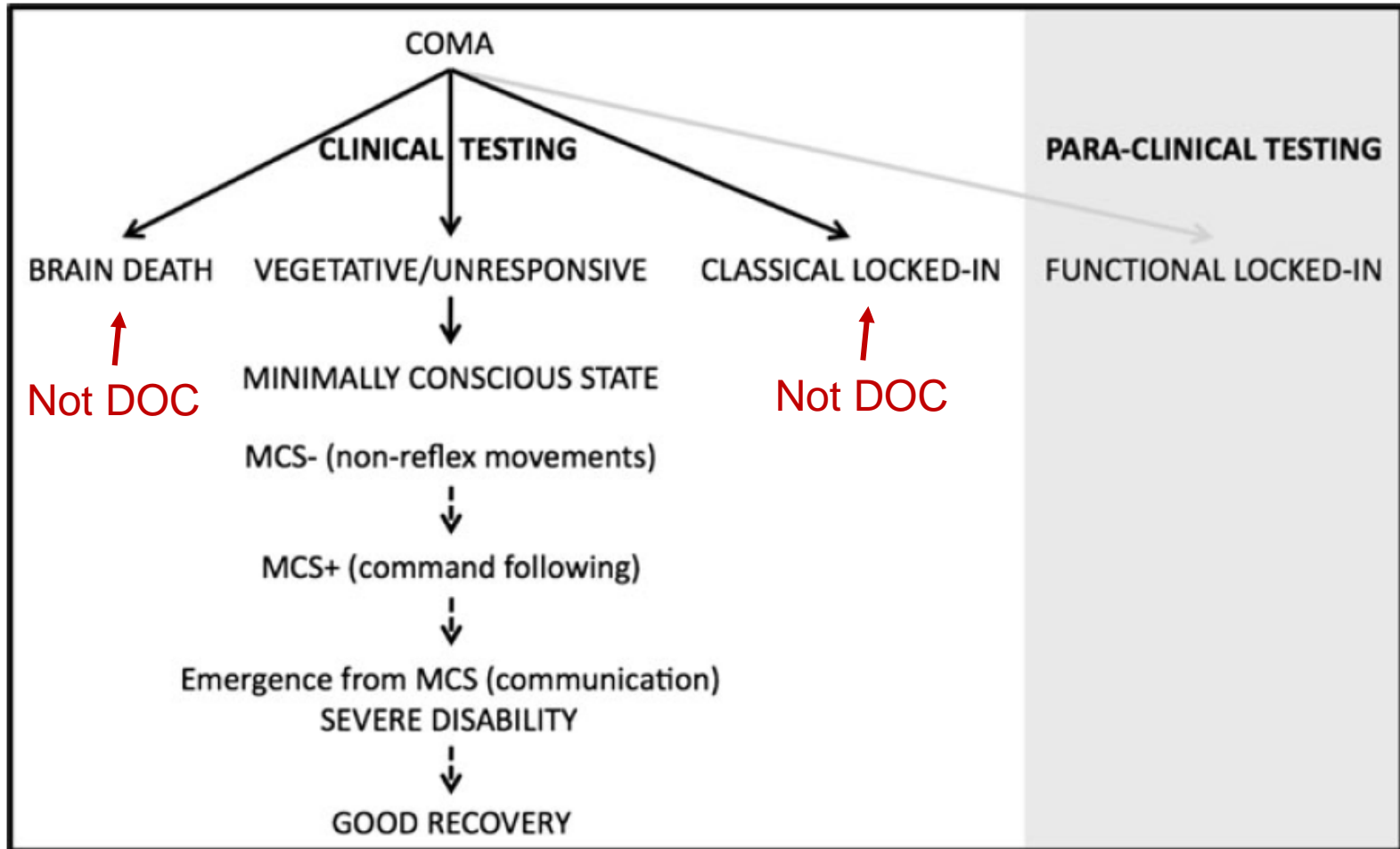
## MCS (-)

Minimal levels of behavioral interaction characterized by the presence of non-reflex movements such as: (i) orientation of noxious stimuli, (ii) pursuit eye movements that occur appropriately in relation to relevant environmental stimuli.

## MCS (+)

Presence of (i) command following, (ii) intelligible verbalization or (iii) gestural or verbal yes/no responses.

# Pediatric DOC

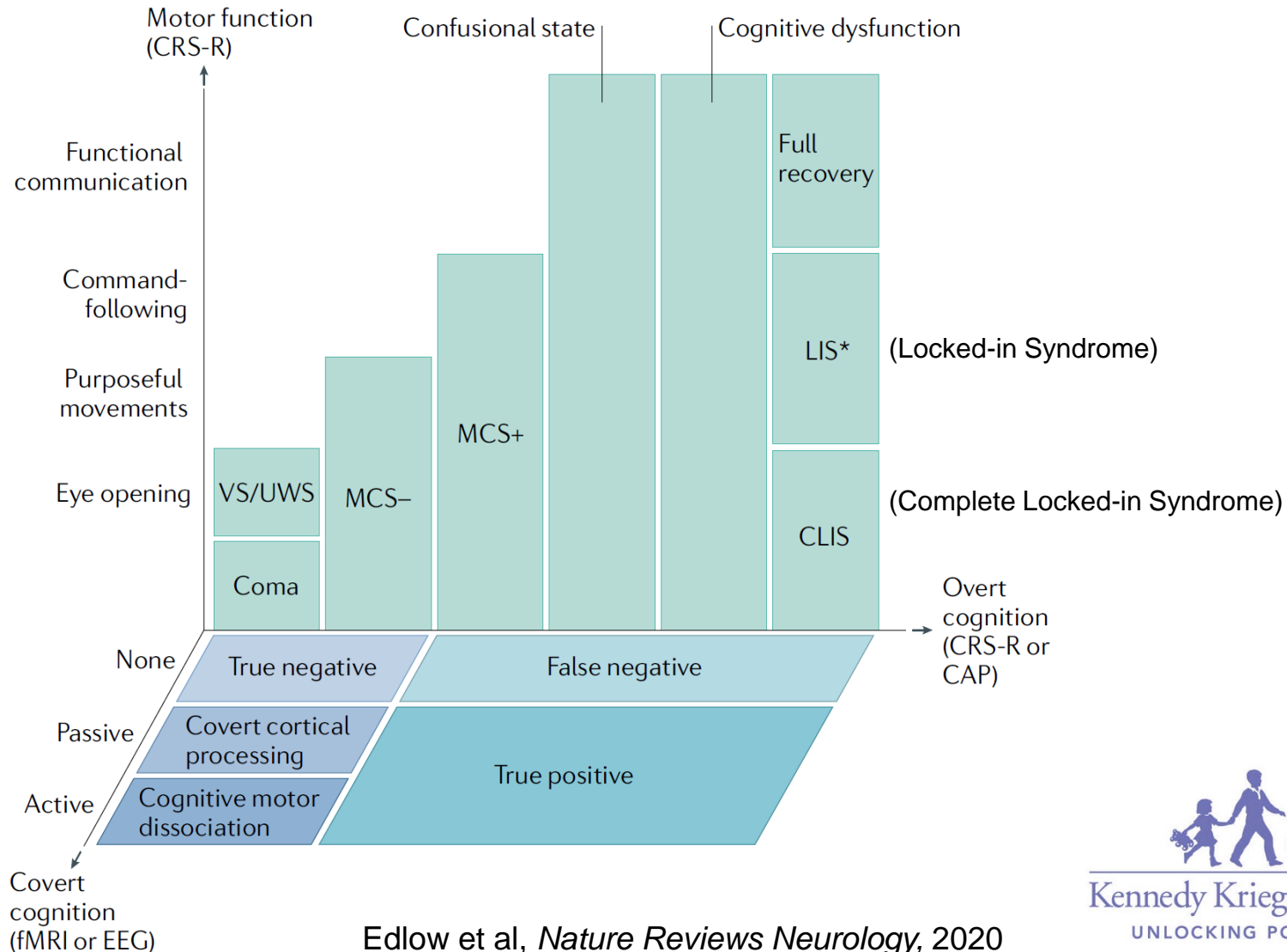


Bruno et al., J Neurol, 2011



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# Multidimensional Assessment of Consciousness





# Assessment



## Pediatric DoC – Systematic Review (2018)

15 Recommendations for Adults, 3 for Children

- **Rec 16** – Treat confounding conditions, increase arousal, use standardized behavioral assessment targeted for children, conduct serial evaluations
- Rec 17 – Counsel families that natural history/prognosis is not well defined and there are no evaluations to improve prognostic accuracy
- Rec 18 – Counsel families that there are no established therapies for prolonged DOC

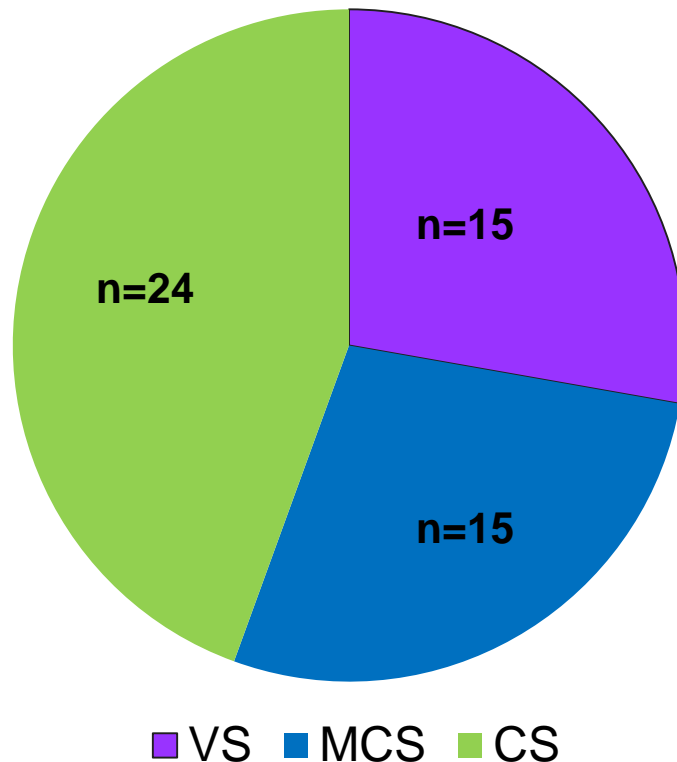
# Standardized Evaluation Tools

- **JFK Coma Recovery Scale – Revised (CRS-R)**
  - Coma Recovery Scale – Pediatrics (CRS-P)
- **Rappaport Coma/Near Coma Scale (CNCS)**
- Western Neuro Sensory Stimulation Profile (WNSSP)
- Disorders of Consciousness Scale (DOCS)
- Sensory Modality Assessment and Rehabilitation Technique (SMART)
- Wessex Head Injury Matrix (WHIM)
- Sensory Stimulation Assessment Measure (SSAM)

# JFK Coma Recovery Scale - Revised

- Auditory Function
- Visual Function
- Motor Function
  - Functional object use\*
- Oromotor/Verbal Function
- Communication
  - Functional communication\*
- Arousal

## Young Children with ABI (n = 54), 31% TBI

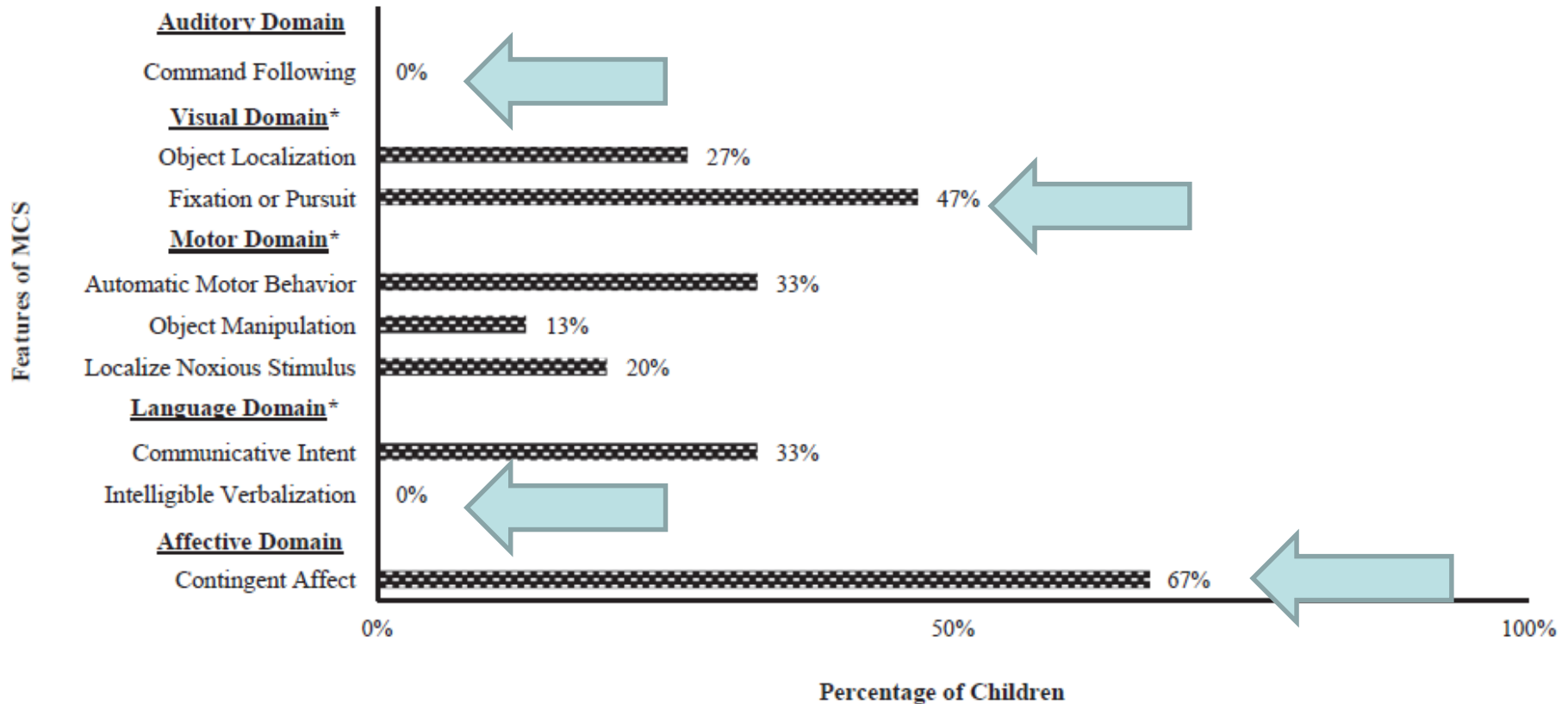


- 9/15 children in VS emerged to MCS
- No child emerged from VS to CS
- 5/15 children emerged from MCS to CS



# Features of MCS at Admission

**B** Children in a Minimally Conscious State ( $N = 15$ ).



\*Note. Ratings reflect the best demonstrated skill within the domain.

# Features of CS

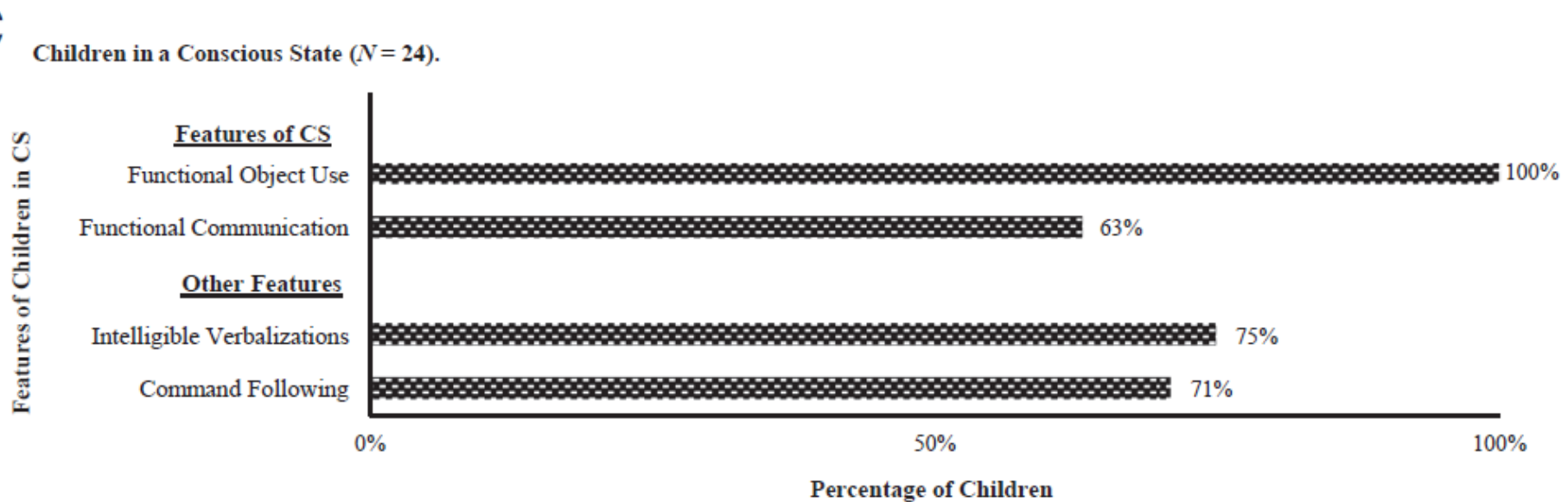


Fig 1 Features of states of consciousness at admission.

## At admission

- Functional Object Use not observed <12 months
- Functional Communication not observed <20 months

## By discharge

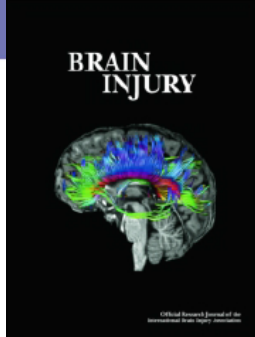
- 5 more emerged to CS, 2/5 had Functional Object Use Only

## Clinical Features of DOC in very young children

- Visual and motor skills may be most applicable
- Language-based skills may be least applicable
- Accurate classification may have important prognostic implications







## Brain Injury

ISSN: 0269-9052 (Print) 1362-301X (Online) Journal homepage: <https://www.tandfonline.com/loi/ibij20>

### Preliminary validation of the coma recovery scale for pediatrics in typically developing young children

Beth S. Slomine, Stacy J. Suskauer, Rachel Nicholson & Joseph T. Giacino

**33 typically developing children (8 months to 59 months)**

#### Modifications

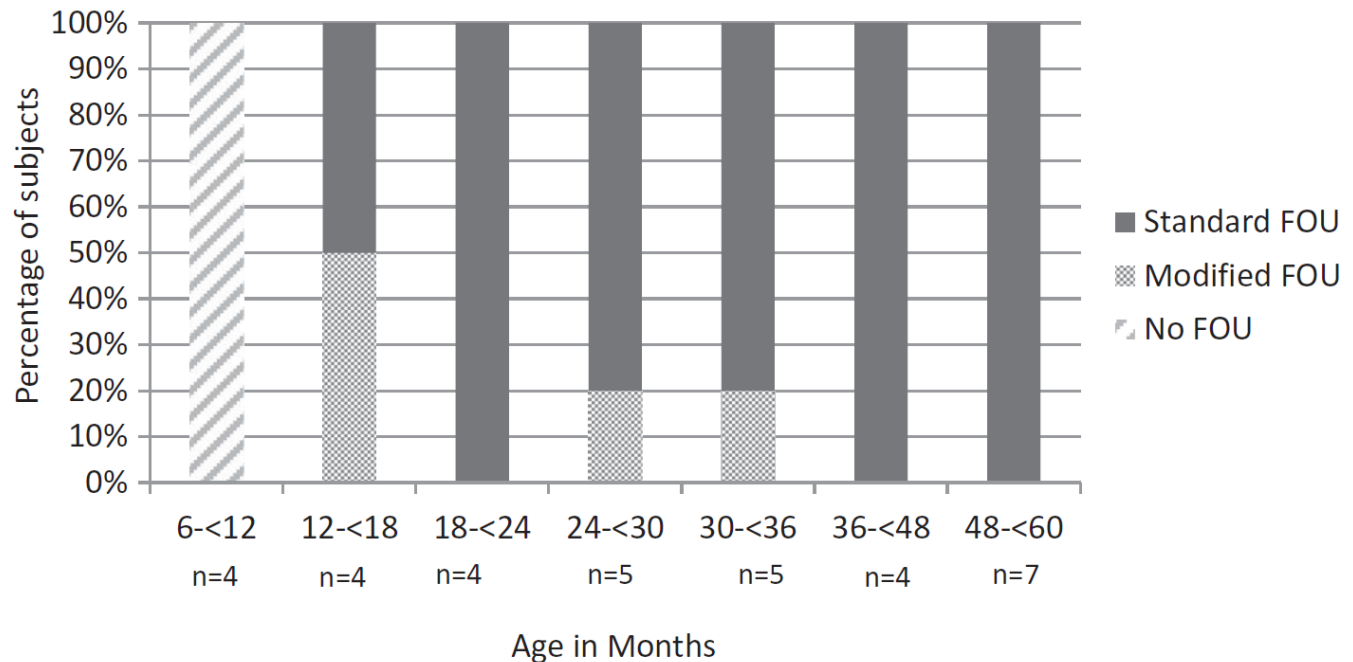
- toys/language
- Functional Object Use – spontaneous play
- Functional Communication – questions from a picture book
- Intelligible Verbalizations – What is this...this is a...?
- Arousal – definition modification
- Automatic motor responses with play
- Affect scale



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# Functional Object Use

Uninjured Children:  
At what age is functional object use established?

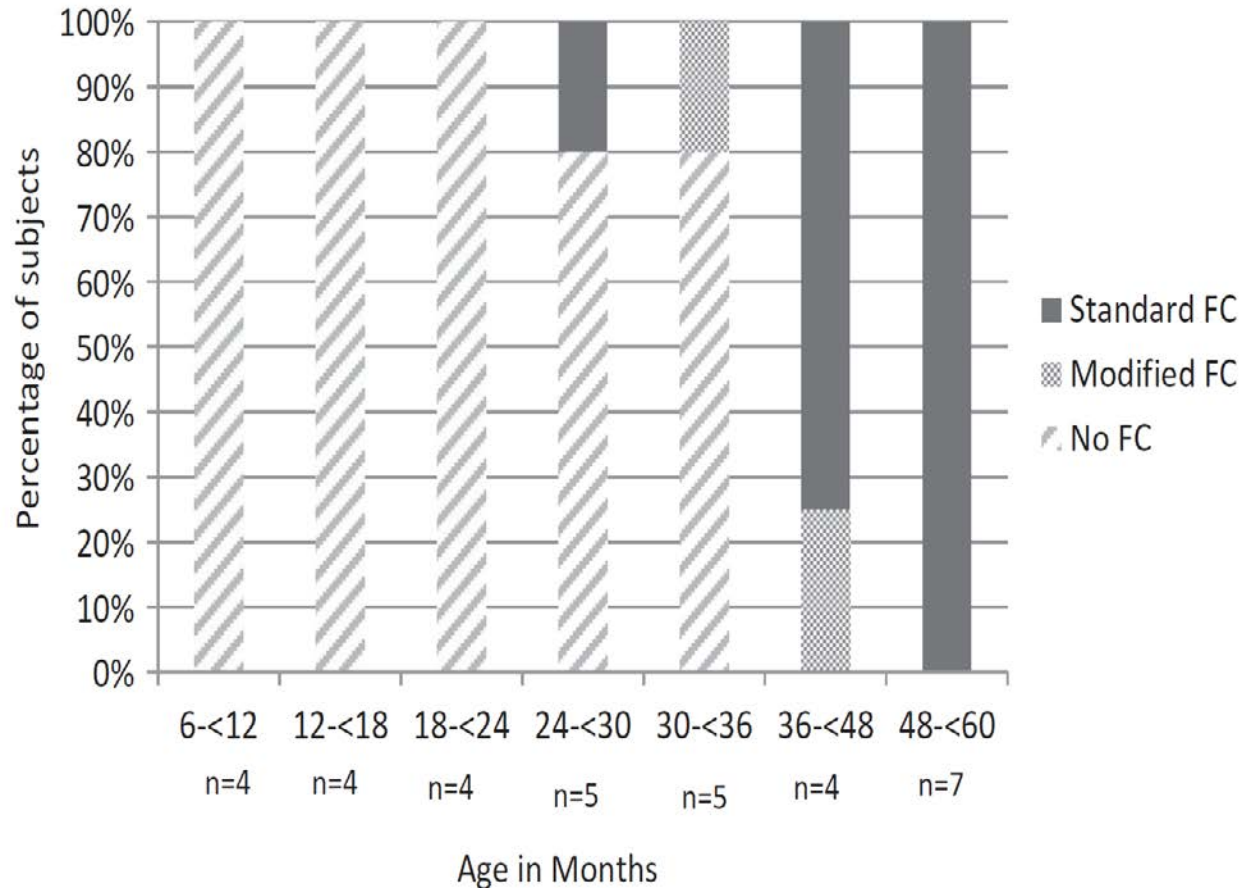
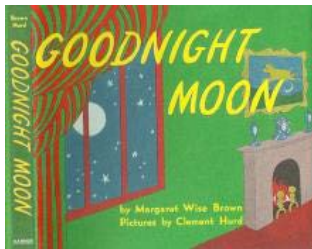




### Preliminary validation of the coma recovery scale for pediatrics in typically developing young children

Beth S. Slomine, Stacy J. Suskauer, Rachel Nicholson & Joseph T. Giacino

# Uninjured Children: at what age is Functional Communication established?



## Summary from CRS-Pediatrics validation study (in typically functioning children)

- Visual and motor skills develop earliest
- Language-based skills develop later
- Some younger children showed FOU and FC only with modifications
- Overall – caution when assessing DOC in young children



# Individualized Assessments

- Target a few behaviors of particular interest
  - Short assessments
  - Can be repeated throughout day by varying staff and family members
- Examples:
  - Arousal: eye opening, response to stimulus
  - Command following versus automatic movements
  - Vision/Hearing: preferential attention to salient stimuli



# Individualized Protocols

Explanation, Positioning, Directions, Observations

<b>Trials (15 seconds each)</b>	<b>Target response</b>	<b>Other arm movement (define)</b>	<b>Other Response (smile, posturing)</b>	<b>No Response</b>
<i>Touch the iPad</i>				
Observation				
<i>Touch the iPad</i>				
<i>Touch the iPad</i>				
Observation''				
<i>Touch the iPad</i>				
Observation				
Observation				



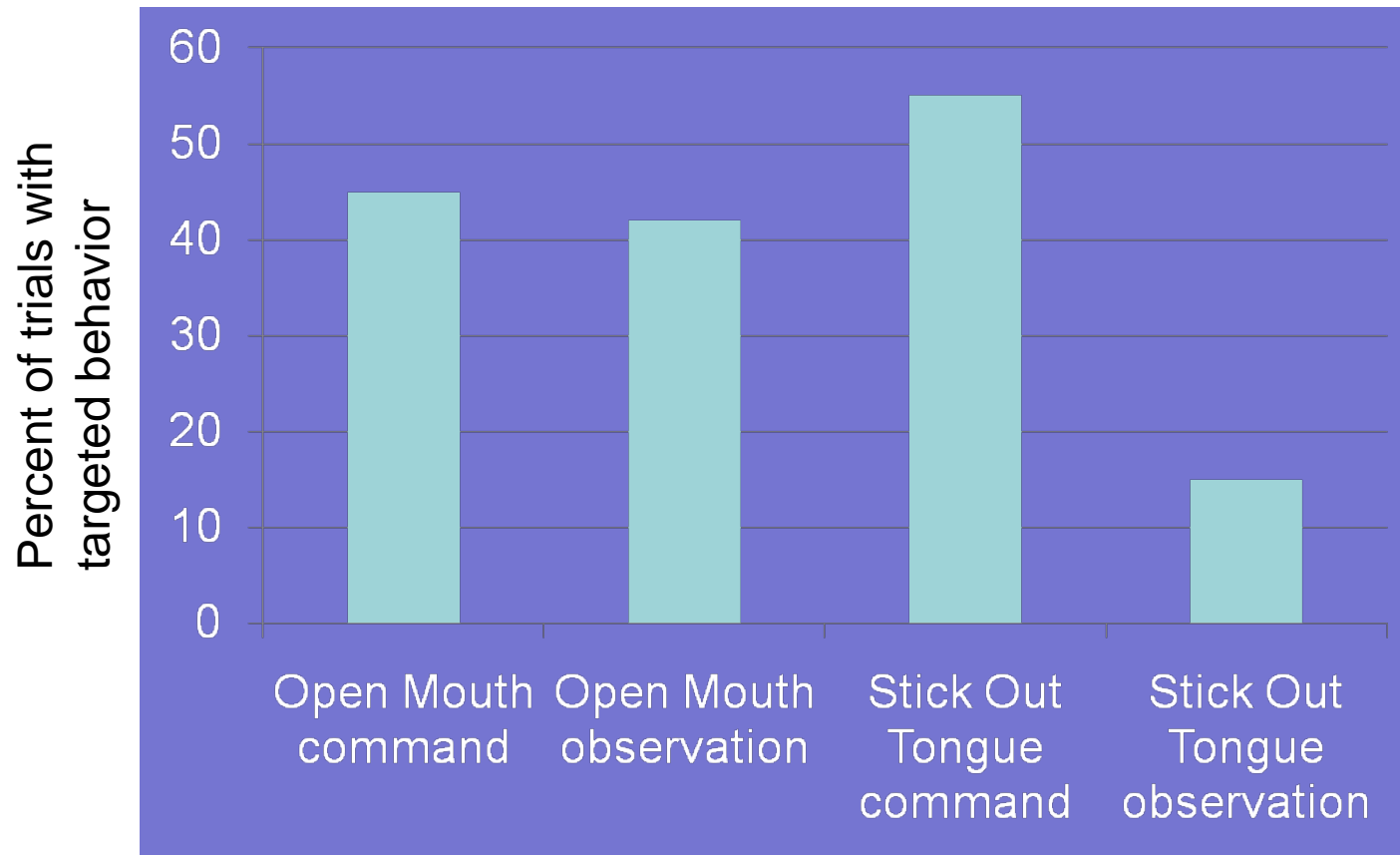
# Command following or not?

# Individualized Protocol- Command Following

	Opens Mouth	Sticks Out Tongue	No Response
Stick out your tongue			
(No Command)			
Open your mouth			
Stick out your tongue			
Open your mouth			
(No command)			
(No command)			
Open your mouth			
Stick out your tongue			

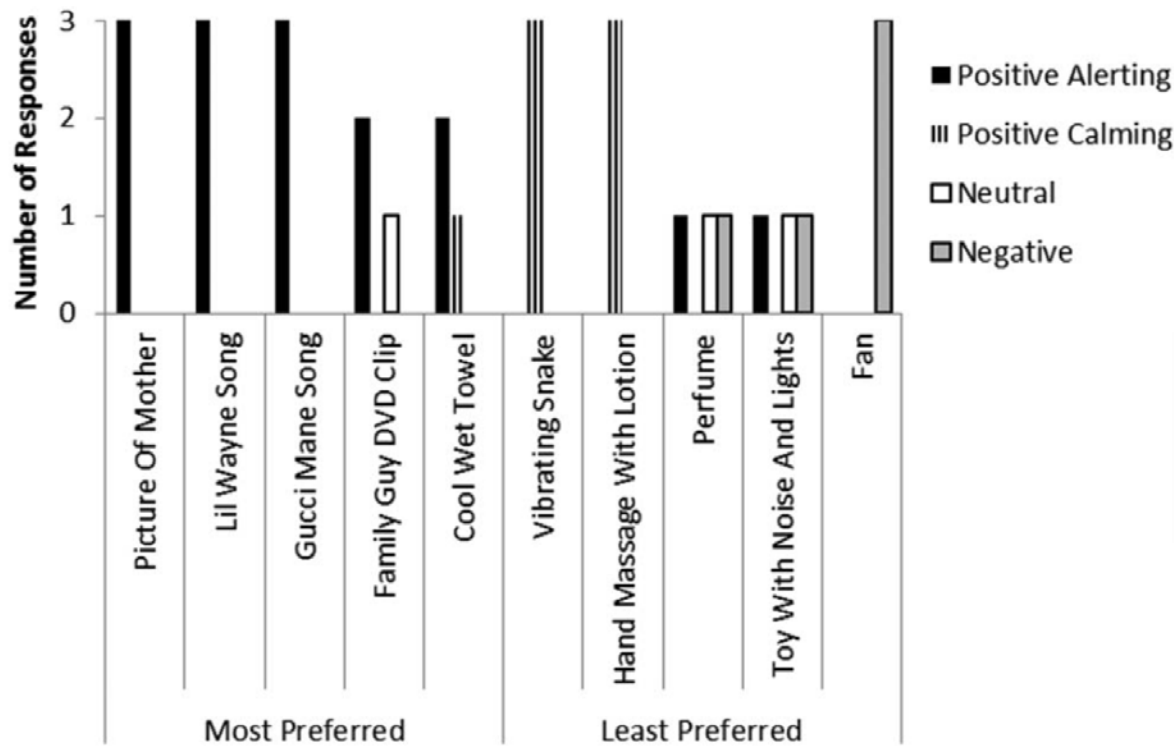


# Individualized Protocol Results



# Conducting Preference Assessments for Youth With Disorders of Consciousness During Rehabilitation

Adrianna Amari, Stacy J. Suskauer, Valerie Paasch, Lauren K. Grodin, and Beth S. Slomine  
 Kennedy Krieger Institute, Baltimore, Maryland, and Johns Hopkins University School of Medicine



Amari et al., *Rehab Psych*, 2017



# Cognitive and Linguistic Scale (CAL S)

Arousal  
Responsivity  
Emotional Regulation  
Inhibition  
Focusing and Resisting  
Response Time  
Orientation  
Memory for New Information  
Simple Receptive Language  
Complex Receptive Language

Simple Expressive Language  
Complex Expressive Language  
Initiation  
Pragmatics  
Simple Problem Solving  
Complex Problem Solving  
Visuoperceptual Ability  
Visuospatial ability  
Self-Monitoring/Evaluation  
Cognitive Safety



# Physical Abilities and Mobility Scale (PAMS)

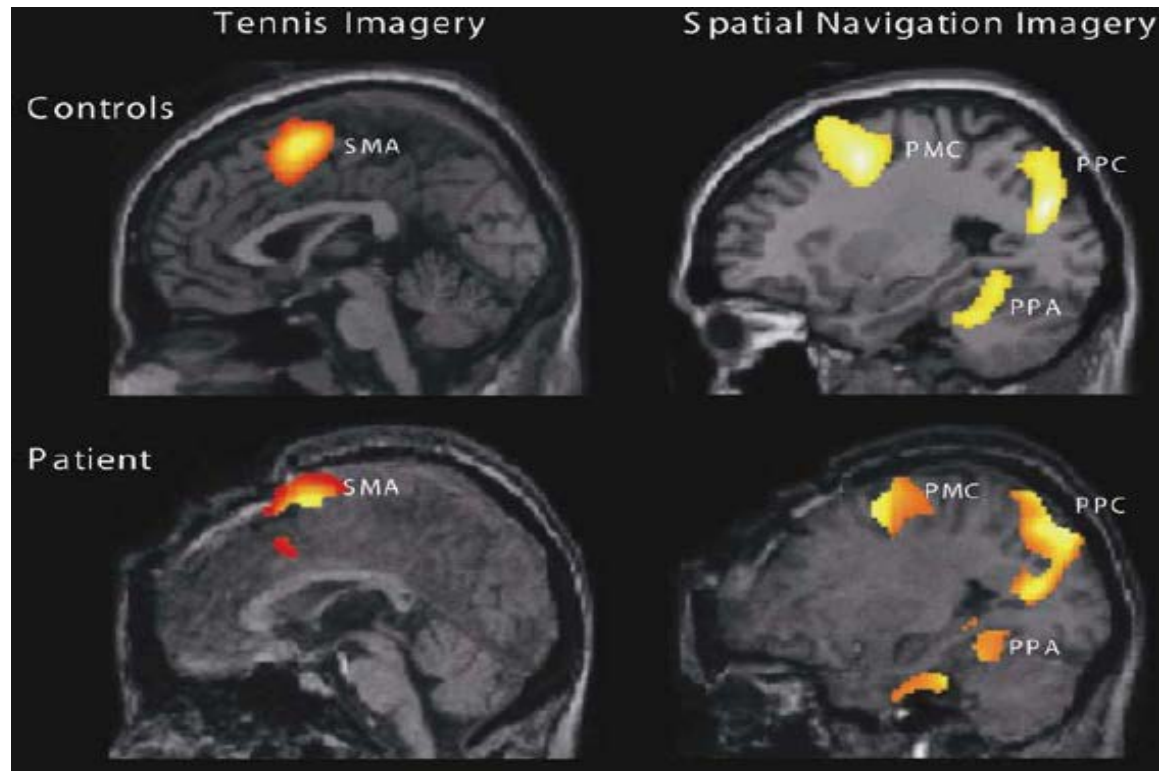
Tolerance to positioning  
Tolerance to sitting in a wheelchair  
Tolerance to orthosis or splint  
Support for seating system  
Head control  
Trunk control  
Rolling supine to/from prone  
Transitioning from supine to sit  
Transitioning from sit to stand  
Standing  
Transitioning from floor to stand

Environmental transfers  
Transfers into and out of a car  
Walking on level ground:  
    assistive device  
Walking on level ground: distance  
Walking on level ground: level of  
    assistance  
Community skills  
Wheelchair mobility  
Standing balance  
Stairs



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# Assessment of Covert Cognition



Owen et al., Science, 2006



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# Prognosis and Outcomes



## Pediatric DoC – Systematic Review (2018)

15 Recommendations for Adults, 3 for Children

- Rec 16 – Treat confounding conditions, increase arousal, use standardized behavioral assessment targeted for children, conduct serial evaluations
- **Rec 17 – Counsel families that natural history/prognosis is not well defined and there are no evaluations to improve prognostic accuracy**
- Rec 18 – Counsel families that there are no established therapies for prolonged DOC

## Short-term prognosis differs for patients with TBI in VS/MCS- vs MCS+ at admission to inpatient rehab

	<b>Emerged (n = 10), mean (SD)</b>	<b>Did not emerge (n = 4), mean (SD)</b>	<b>P</b>	<b>Effect size</b>
Age at injury (years)	14.9 (5)	8.7 (5)	.08	0.49
Initial CNCS score <sup>a</sup>	1.8 (0.4)	2.8 (0.4)	.025	0.62
Initial CALS responsiveness item score <sup>b</sup>	3.8 (0.8)	1.3 (0.6)	.007	0.71
In MCS+ at admission <sup>c</sup>	80%	0%	.015	0.73

Abbreviations: CALS, Cognitive and Linguistic Scale; CNCS, Coma/Near-Coma Scale; CS, conscious state; MCS, minimally conscious state; VS, vegetative state.

<sup>a</sup>CNCS was not administered to 1 patient (who emerged to CS).

<sup>b</sup>CALS was not administered to 1 patient younger than 2 years at injury (who did not emerge to CS).

<sup>c</sup>Versus VS or MCS-.

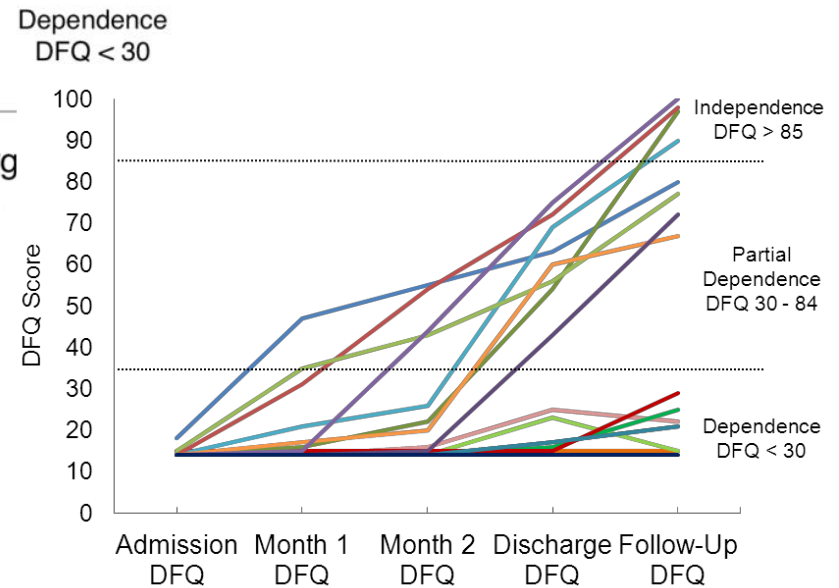
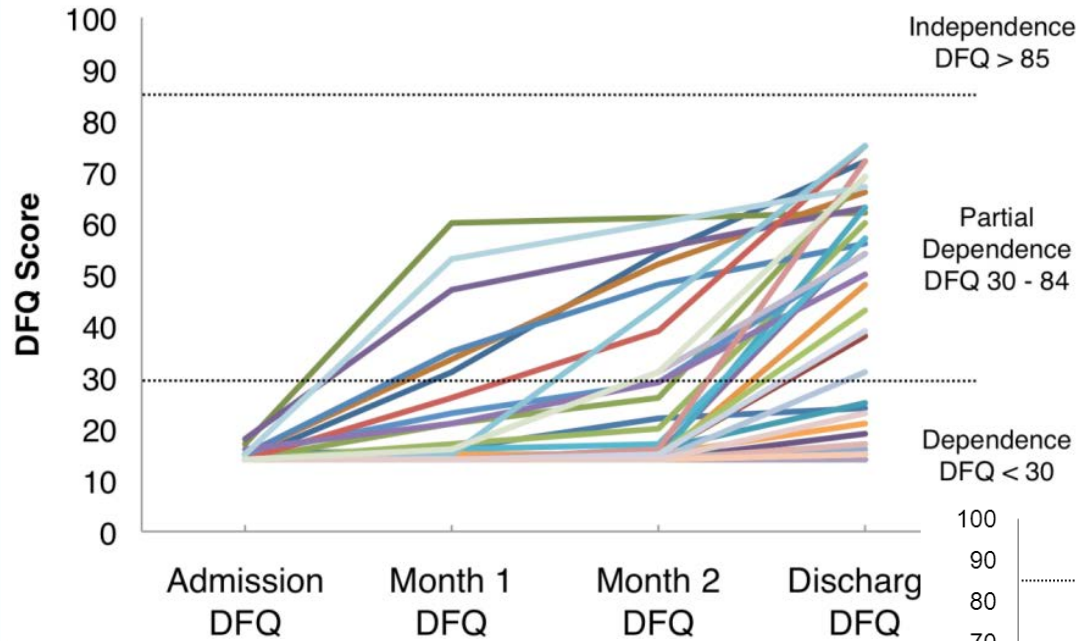
Pham et al., JHTR, 2014



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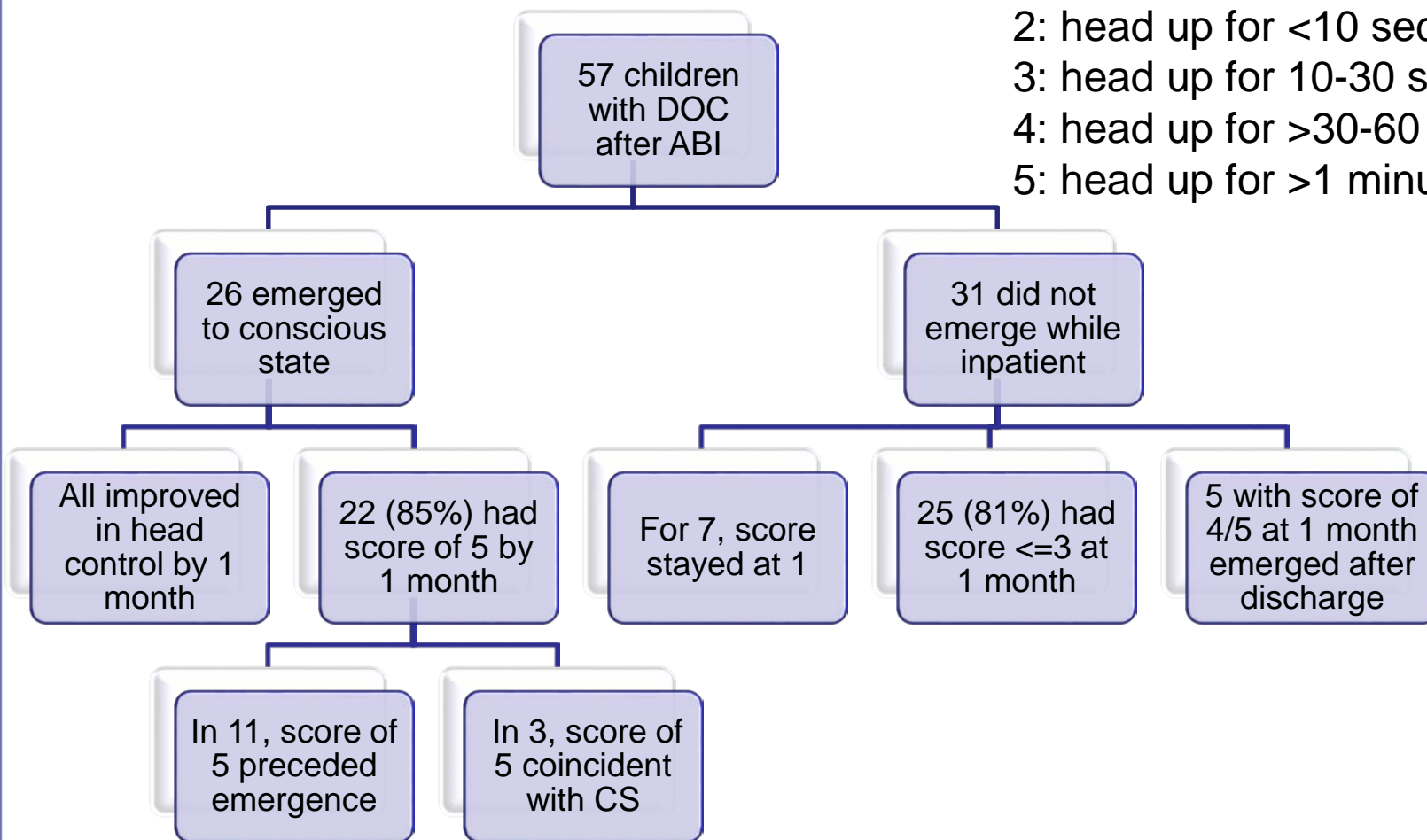


# Early changes predicts further recovery



# PAMS head control scores within 1 month of admission to inpatient rehabilitation

Head Control scoring:  
 1: unable to maintain head up  
 2: head up for <10 seconds  
 3: head up for 10-30 seconds  
 4: head up for >30-60 second  
 5: head up for >1 minute



# Outcomes



Archives of Physical Medicine and Rehabilitation

journal homepage: [www.archives-pmr.org](http://www.archives-pmr.org)

Archives of Physical Medicine and Rehabilitation 2021; ■: ■■■■-■■■



## ORIGINAL RESEARCH

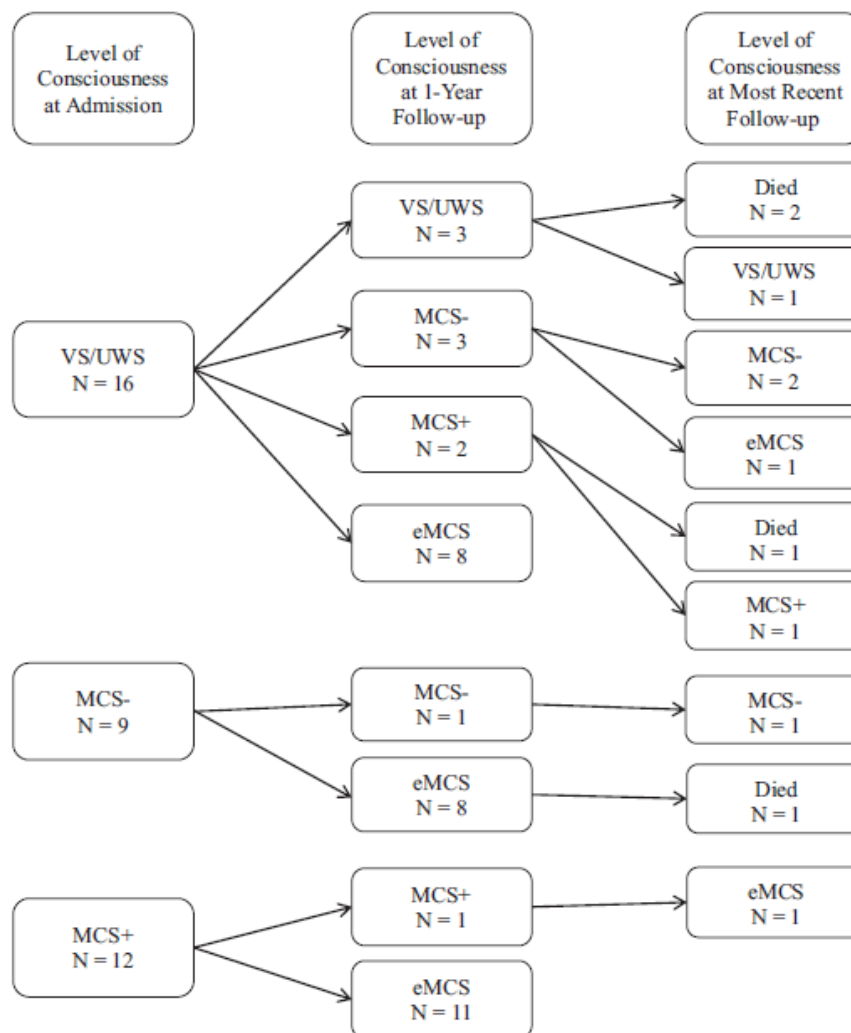
### Very Long-Term Outcomes in Children Admitted in a Disorder of Consciousness After Severe Traumatic Brain Injury

Sandra Rodgin, PhD,<sup>a,b</sup> Stacy J. Suskauer, MD,<sup>c,d</sup> Julia Chen, PhD,<sup>e</sup> Elana Katz, MD,<sup>f</sup> Kimberly C. Davis, PhD,<sup>g,h</sup> Beth S. Slomine, PhD<sup>a,b,c</sup>

37 children, ages 2-18 year, admitted to inpatient rehabilitation

Admission CALS scores <30

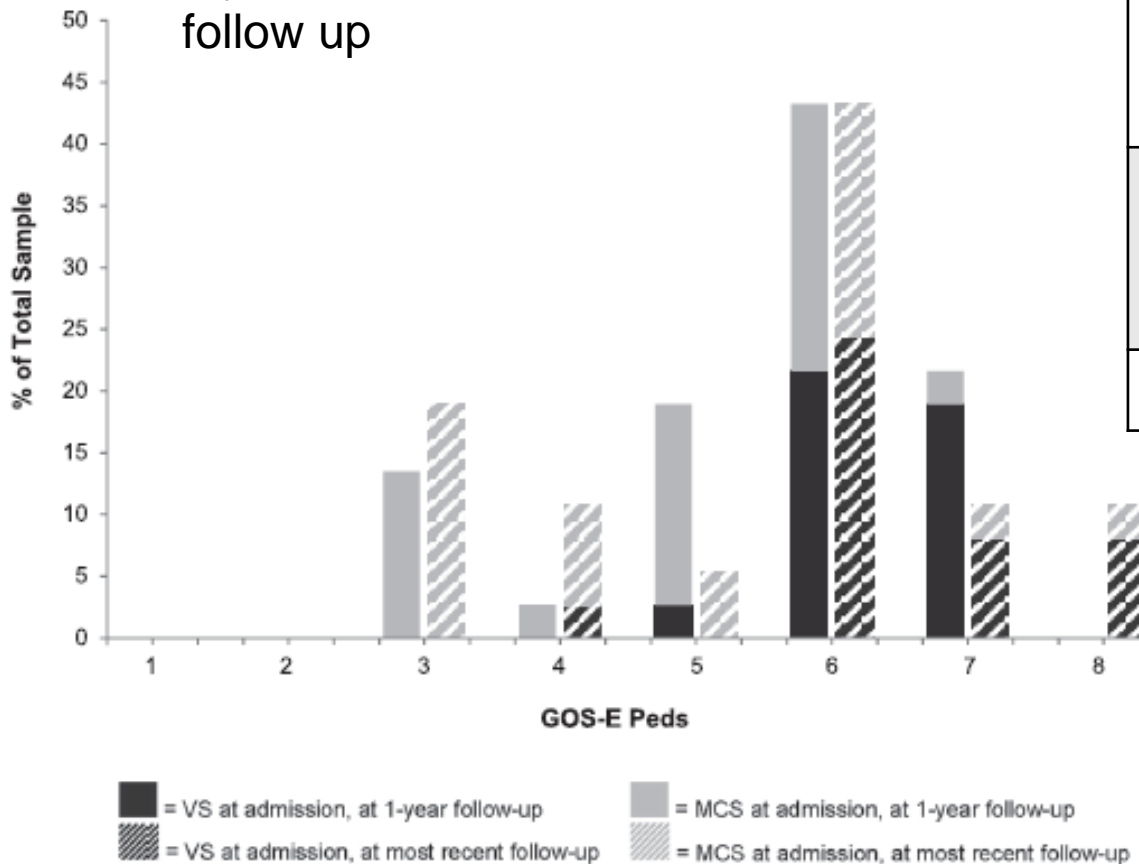
# Very Long-term outcome



**Fig 1** Flowchart of levels of consciousness at admission, 1-year follow-up, and the most recent follow-up. Abbreviation: UWS, unresponsive wakefulness syndrome.

# Very Long-Term Outcome

6/33 (18%) improved between 1 year and most recent follow up



**Fig 2** Distribution of GOS-E Peds scores at 1-year follow-up and the most recent follow-up.

Category	GOS-E Peds score and description of level
Death	8
Vegetative State	7 = Child is unable to communicate or follow commands
Severe Disability	6 = Child requires a caretaker at home most times.
	5 = Child requires some caregiver support at home or requires caregiver support outside the home.
Moderate Disability	4 = Child is in a self-contained school or sheltered job or experiences daily and intolerable psychological problems.
	3 = Child has a reduced work or school capacity.

## Take home points

**Assessment** - Serial neurobehavioral assessment is feasible and useful, including measures such as CRS-R, CRS-P, CALS, PAMS

**Prognosis** – early responsiveness/recovery associated with later outcome

**Outcomes** – range of outcomes with continued recovery over many years in a subset of patients

# Collaborators and Funding Sources

## KKI/JHSOM Faculty

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Adrianna Amari, Ph.D.  
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## Brooks Rehabilitation

Sarah Lahey, Ph.D.



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# Questions?

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