

JOHNS HOPKINS ALL CHILDREN'S HOSPITAL

Childhood Arterial Ischemic Stroke Clinical Pathway

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Owners: Dr. John Deveikis; Dr. Neil Goldenberg; Katie Mills, APRN

This pathway is intended as a guide for physicians, physician assistants, nurse practitioners and other healthcare providers. It should be adapted to the care of specific patient based on the patient's individualized circumstances and the practitioner's professional judgment.

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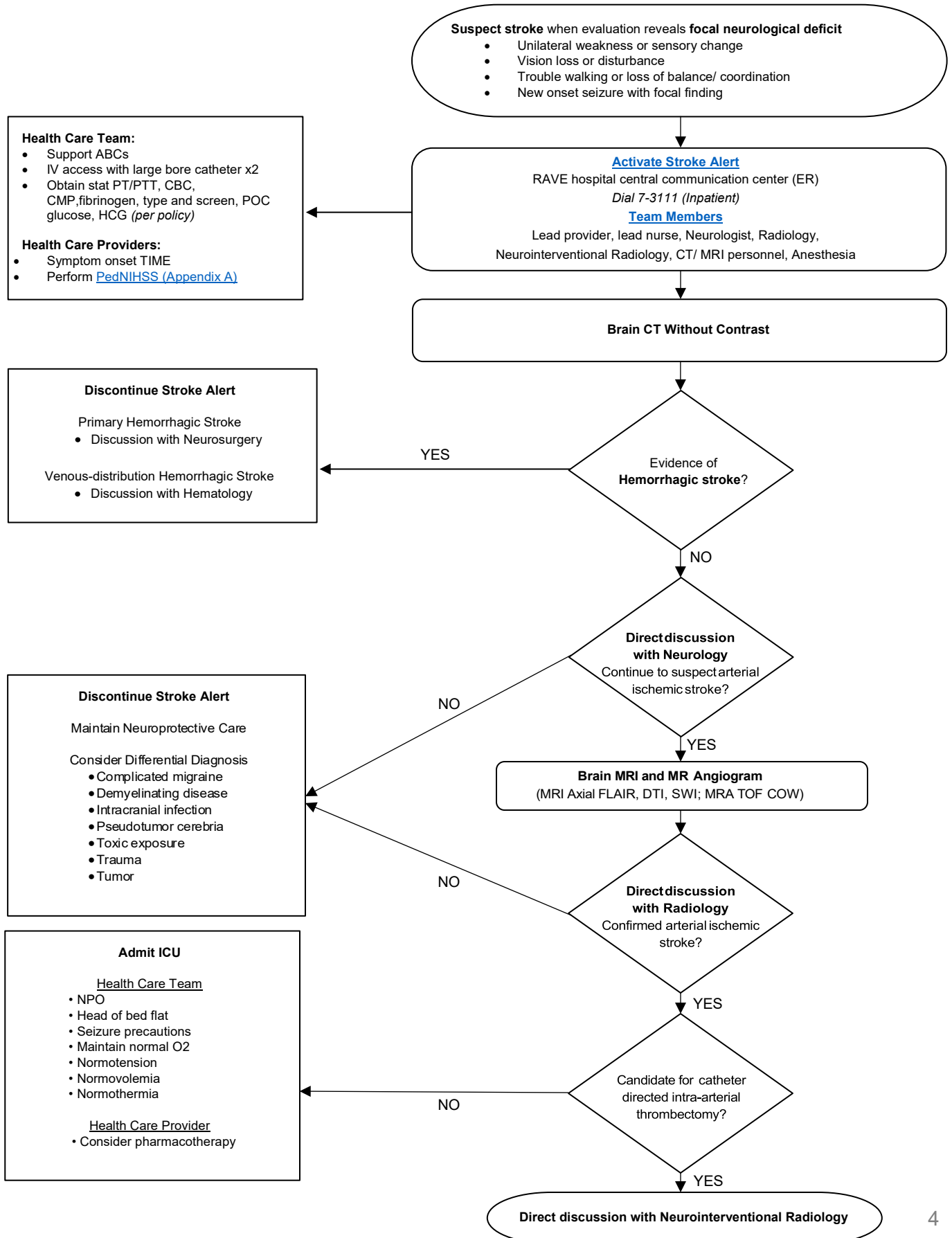
Childhood Arterial Ischemic Stroke Clinical Pathway

Rationale

This pathway was developed by a consensus group of JHACH Pediatric Emergency Medicine Physicians, Critical Care Physicians, Hematologists, Neurologists, Neurosurgeons, Pediatric Interventional Radiologists, and key health care professionals directly involved in the management of pediatric stroke. This pathway was developed to standardize the management of children with suspected or confirmed arterial ischemic stroke (AIS). This pathway addresses the following clinical questions and problems:

1. Evaluation of pediatric stroke
2. Appropriate diagnostic testing
3. Management of childhood AIS
4. When to consider intravenous tPA
5. When to consider catheter directed intra arterial thrombectomy
6. Pharmacotherapy for childhood AIS

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Background

Cerebral ischemia occurs within minutes to hours of arterial occlusion. Due to the lack of recognition of acute neurological deficits and a failure to include stroke as a differential diagnosis, children encounter considerable diagnostic delay of stroke when compared to adults. Many patients present within 6 hours of symptom onset, but median time to radiological confirmation ranges from 15 to 21 hours.

The incidence of childhood arterial ischemic stroke is 1-2/100,000 per year in Western developed countries.

While the mortality rate is approximately 1/3 that of adults, stroke in children carries lifelong morbidity. Of those children greater than one month affected by stroke nearly 65% sustain lasting motor deficits, and 50% are found to have cognitive deficits.

Definitions

Acute perinatal arterial ischemic stroke occurs in the neonatal period, often presenting with symptomatic seizures. The injury likely occurred immediately prior to, during, or immediately following birth.

Presumed perinatal arterial ischemic stroke is retrospectively diagnosed by the presence of a chronic ischemic injury on neuroimaging in an infant presenting in the first year of life with emerging hemiparesis.

Childhood arterial ischemic stroke diagnosed by the presence of acute ischemic injury on neuroimaging beyond first month of life.

Hemorrhagic stroke, also referred to as intracranial hemorrhage, occurs as a result of bleeding from an artery in the brain directly into brain tissue.

Sub-acute stroke defined by our facility for purposes of neurointervention and acute management includes ischemic injury > 24 hours and ischemic injury > 48 hours for posterior circulation stroke.

Suspected Stroke

Diagnosis

Clinical presentation varies according to age, setting, and stroke subtype.

Individuals ≥ 18 years of age exhibiting symptoms of stroke should be considered for transfer to a Primary Stroke Center (Adult). The use of intravenous tissue-type plasminogen activator (tPA) may significantly benefit an adult with arterial ischemic stroke, a treatment option with limited guidelines in pediatrics.

Evaluation

1. Focal neurological deficit (one or more of the following)
 - a. Unilateral weakness or sensory change
 - b. Vision loss or disturbance
 - c. Speech or language disturbance
 - d. Trouble walking or loss of balance/ coordination
 - e. New onset seizures with focal findings

OR

2. Focal neurological deficit occurring suddenly or a sudden worsening of a pre-existing deficit.

AND

1. Time of symptoms onset
2. When was the individual last seen symptom free/ "normal"?
3. When did the individual last eat or drink?

Examples of a focal neurological deficit include, but are not limited to, abrupt onset hemiparesis, monoparesis, monocular or binocular vision loss, diplopia, aphasia, dysarthria, ataxia, and sudden decrease in level of consciousness.

Stroke Alert

Activated by ED Provider, via hospital central communication center (RAVE), when an acute arterial ischemic stroke is suspected in a patient presenting to the Emergency Department.

If suspected stroke occurs while inpatient the health care provider activates Stroke Alert (dial 7-3111), and prepares for transfer to higher level of care (ICU). Responding members will include the Medical Emergency Team (Rapid Response) and those health care personnel will assume care of the patient until the Stroke Alert is discontinued.

Please note, only health care providers should activate Stroke Alert. If other health care personnel suspect stroke a Rapid Response should be activated for urgent patient evaluation by members of the Medical Emergency Team.

Standard text should include "Stroke Alert: patient location, patient age, requesting provider, call back number"

Team members

Active role

- a. Lead Provider
- b. Nurse Leader
- c. Neurologist (All Children's Specialty Physician Group)
- d. Neurointerventional Radiologist
- e. CT and MRI personnel
- f. Anesthesia

On deck

- g. Neurosurgery
- h. Intensive Care Provider (*if alert called in ED*)
- i. Intensive Care Nurse Leader (*if alert called in ED*)
- j. Nursing Supervisor
- k. Hospitalist

Diagnostic Imaging

1. Brain CT without contrast
 - a. Evidence of arterial ischemic stroke proceed with MRI
 - b. Evidence of primary hemorrhagic stroke
 - i. Discontinue Stroke Alert via hospital central communication center (RAVE). Standard text should include "Stroke Alert: Discontinued"
 - ii. Direct discussion with Neurosurgery
 - iii. Admit to appropriate inpatient unit
 - iv. Consider brain CT angiogram evaluate for arteriovenous malformation or aneurysm
 - c. Evidence of venous-distribution hemorrhagic stroke
 - i. Discontinue Stroke Alert via hospital central communication center (RAVE).

- Standard text should include “Stroke Alert: Discontinued”
 - ii. Direct discussion with Hematology
 - iii. Admit to appropriate inpatient unit
 - iv. Consider brain CT venogram or MR venogram evaluate for cerebral sinovenous thrombosis (CSVT)
2. Brain MRI and brain MR angiogram
 - a. Obtain on 3T or stronger
 - b. Stroke neuroimaging protocol MRI Fluid-attenuated inversion recovery, diffusion tensor imaging, susceptibility weighted imaging; MRA time-of-flight, Circle of Willis (MRI Axial FLAIR, DTI, SWI; MRA, TOF, COW)
 - c. Evidence of *acute* arterial ischemic stroke on MRI send confirmation via hospital central communication center (RAVE). Standard text should include “Stroke Alert: Confirmed AIS, patient location, patient name, patient age, primary provider, call back number”
 - d. Evidence of *sub-acute* arterial ischemic stroke on MRI. Discontinue Stroke Alert via hospital central communication center (RAVE). Standard text should include “Stroke Alert: Discontinued”

Roles and Responsibilities

1. Lead Attending Physician or Designee (APP/ Resident)
 - a. Report to bedside to perform and document Pediatric National Institutes of Health Stroke Score ([PedsNIHSS Appendix A](#))
 - b. Assume responsibility as primary provider and contact until patient transferred to appropriate unit
 - c. Provide emergent and urgent support treatment (refer to ED nursing)
2. Nursing
 - a. Initiate emergent support treatment
 - i. Support ABCs (refer to PALS)
 - ii. Obtain intravenous access with large bore catheter (2 sites)
 - iii. Obtain *stat* PTT, CBC, fibrinogen, POC glucose, and electrolytes (BMP), type and screen, HCG (per policy)
 - b. Initiate urgent support treatment
 - i. NPO
 - ii. Head of bed flat
 - iii. Maintain seizure precautions
 - iv. Maintain normal O₂
 - v. Normotension
 - vi. Normovolemia
 - vii. Normoglycemia
 - viii. Normothermia
3. Neurologist (All Children’s Specialty Physician Group)
 - a. Report to the bedside if within the hospital, or call ED Attending Physician
 - i. Confirm suspected acute childhood arterial ischemic stroke based on report and proceed with diagnostic imaging
 - ii. *If* suspected *sub-acute* childhood arterial ischemic stroke discontinue Stroke Alert and continue to management

- b. Seizure control by treating any suspected seizure activity as indicated
- 4. Neurointerventional Radiologist
 - a. Prepare and mobilize staff for diagnostic imaging and/or neurointervention
- 5. CT and MRI Personnel
 - a. Prepare and mobilize for diagnostic imaging
- 6. Anesthesia
 - a. Prepare and mobilize for sedation needs for diagnostic imaging and/or neurointervention
- 7. Neurosurgery
 - a. Prepare for intervention in the event of hemorrhagic stroke

Catheter Directed Intra-arterial Thrombectomy Considerations

Amount of ischemic injury may be reduced when recanalization and reperfusion is accomplished within the first hours of symptom onset.

Individuals ≥ 18 years of age exhibiting symptoms of stroke should be considered for transfer to a Primary Stroke Center (Adult). The use of intravenous (systemic) tissue-type plasminogen activator (tPA) should be considered as it may significantly benefit an adult with arterial ischemic stroke. Systemic tPA in pediatric patients is not well established and is not routinely practiced at this institution.

Catheter directed intra-arterial thrombectomy can be performed in the Johns Hopkins All Children's Interventional Radiology Suite by Neurointerventional Radiologist.

Considerations

1. Radiographically confirmed cerebral large artery occlusion
2. [PedNIHSS](#) ≥ 6
3. Symptom onset ≤ 24 hours (≤ 48 hours for posterior circulation stroke)
4. Ability to transfer patient to PICU immediately following the procedure
5. Neurointerventionalist to determine technical feasibility based on weight and age of patient on a case-by-case basis
6. Absolute contraindication
 - a. Intracerebral hemorrhage

If patient meets considerations #1-4 without contraindications a direct discussion between Attending Physician and Neurointerventional Radiologist is strongly recommended.

Management of Confirmed Stroke

Supportive Treatment

1. Nothing by mouth (minimum 24 hours)
2. Head of bed flat
 - a. If intracerebral hemorrhage suspected or confirmed HOB 30 degrees
3. Maintain seizure precautions
4. Maintain normal O₂
5. Maintain normal CO₂
6. Maintain normal pH
7. Normotension
 - a. Target SBP between 50th and 90th percentile for age
 - b. For hypertension, decrease BP by approximately 15% over 24 hours
 - c. Patients with an intracranial arteriopathy may be sensitive to rapid decreases in blood pressure
 - d. Monitored hourly or more frequently as clinically indicated
8. Normovolemia
 - a. Isotonic fluids at maintenance requirements with fluid bolus as indicated
9. Normoglycemia
 - a. Closely monitor during first 24 hours after stroke
 - b. Closely monitor to prevent hypoglycemia and hyperglycemia
 - c. Blood glucose goal 60 - 180 mg/dL
10. Normothermia
 - a. Identify and treat sources of hyperthermia
 - b. Treat temperature >38° with acetaminophen
11. Neurological Check
 - a. Performed every 15 minutes for the first 2 hours, then every 30 minutes for the next 4 hours, then every hour for a total of 24 hours.

Consultations

1. Neurology to assist in seizure management and treatment decisions
2. Neurosurgery to assist in managing suspected arteriopathy or stroke involving ½ MCA territory (concern for malignant edema requiring hemicraniectomy)
3. Hematology to assist in laboratory evaluation and pharmacotherapy
4. Cardiology if cardioembolic source suspected
5. Genetics if connective tissue disorder suspected (e.g. Ehlers-Danlos, Loeys-Dietz)
6. Rehabilitation/ Physiatry to assist in rehabilitation needs and potential for an inpatient rehabilitation facility
7. Educational Services for school aged children to assist in Individualized Education Plan (IEP) and/or 504 plan

Pharmacotherapy

The absence of antithrombotic therapy is associated with a 1.5-2- fold increased risk of recurrent stroke.

1. Non-cardioembolic arterial ischemic stroke without severe thrombophilia, and/or most cases of dissection
 - a. Aspirin 3-5mg/kg/day for 3 months
 - b. Following 3 months of therapy reduce aspirin dose to 1-2mg/kg/day for a minimum of an additional 21 months
 - c. Decision on whether to continue antiplatelet therapy after two years should be made by primary stroke team
2. Cardioembolic arterial ischemic stroke, known severe thrombophilia , recurrent AIS while on antiplatelet therapy, and/or consider for select cases of extracranial dissection
 - a. Low molecular weight heparin (dosing per policy) for 3 months
 - b. Following 3 months of therapy transition to aspirin 1-2mg/kg/day for an additional 21 months
 - c. Decision on whether to increase length of anticoagulation therapy should be made by primary stroke team
3. Arterial ischemic stroke > ½ MCA territory
 - a. No antithrombotic therapy for the initial 96 hours
 - b. Consider frequent clinical assessment and serial imaging within first 72 hours to monitor cerebral edema or hemorrhagic transformation as directed by neurosurgery
 - c. Consider decompressive surgery to prevent herniation and brainstem compression
 - d. Initial aspirin per above recommendations after 96 hours and when patient free from cerebral edema or hemorrhagic transformation
4. Unfractionated heparin maybe used in lieu of low molecular weight heparin, per policy, if there is concern for bleeding complications

Diagnostic Imaging

1. Transthoracic echocardiogram with bubble study to assess for a right-to-left shunt
2. EKG to assess for arrhythmia
3. Electrocephalography (EEG) to assess for subclinical seizures
4. If not already obtained, Brain MR Angiogram to assess for arteriopathy
5. Consider neck MR Angiogram to assess for cervical arterial dissection
6. Consider four extremity Doppler ultrasound to assess for deep vein thrombosis in embolic stroke with right-to-left intracardiac shunt
7. Brain CT 24 hours following catheter directed intra-arterial thrombectomy

Laboratory Evaluation

1. Thrombophilia evaluation to evaluate for an inherited or acquired condition that may lead to a hypercoagulable state
2. Inflammatory markers (ESR, CRP, ANA)
3. Infectious disease evaluation as childhood infections have been identified as a trigger for stroke including herpesvirus (EBV, CMV, VZV) and parvovirus B19

Discharge Planning

Much of the recovery happens within the first months after a stroke, but improvement can still be seen even long after a stroke occurs. The cornerstone of helping the repair and recovery process is rehabilitation. Rehabilitation involves different therapies including physical, occupational, and speech therapy.

Discharge Considerations

1. Patient may benefit from transfer to an inpatient rehabilitation facility
2. Medications available for pick up at patient preferred pharmacy
3. Ensure caregivers understand instructions for medication administration and when to seek immediate medical attention

Follow up

1. Primary Care Physician within 1-2 weeks
2. Neurology within 4 weeks of discharge unless otherwise indicated
3. Stroke Clinic (Neurology, Hematology, and Rehabilitation) 3 – 4 months after initial diagnosis.
Stroke Clinic is currently held one day per month on the 5th floor OCC

Outcome Measures

1. Pediatric National Institutes of Health Stroke Scale on presentation
2. Time: Presentation (“door”) to brain CT
3. Time: Presentation (“door”) to brain MRI
4. Time: Presentation (“door”) to site puncture for thrombectomy
5. Post thrombectomy Thrombolysis in Cerebral Infraction (TICI) score
6. Pediatric National Institutes of Health Stroke Scale on discharge
7. Pediatric Stroke Outcome Measure (PSOM) recorded throughout follow up

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Clinical Pathway Team
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Johns Hopkins All Children's Hospital

Owner(s): Dr. John Deveikis; Dr. Neil Goldenberg; Katie Mills, APRN

Also Reviewed by:

Intensive Care: Dr. Ladonna Bingham; Dr. Elliot Melendez
Emergency Center: Dr. Wassam Rahman, Dr. Lisa Odendal, MD; Courtney Titus, PA-C;
Hematology/Oncology: Dr. Marisol Betensky
Neurology: Dr. Thomas Geller
Neurosurgery: Dr. George Jallo
Radiology: Dr. Javier Quintana
Nursing: Elizabeth Halterman, RN;

Clinical Pathway Management Team: Joseph Perno, MD; Courtney Titus, PA-C

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Disclaimer

Clinical Pathways are intended to assist physicians, physician assistants, nurse practitioners and other health care providers in clinical decision-making by describing a range of generally acceptable approaches for the diagnosis, management, or prevention of specific diseases or conditions. The ultimate judgment regarding care of a particular patient must be made by the physician in light of the individual circumstances presented by the patient.

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Appendix A

Pediatric National Institutes of Health Stroke Scale *(adaptation)*

| | |
|---|--|
| Level of Consciousness Choose score, even if full evaluation is hindered by obstacles such as intubation, language barrier | 0 = Alert, responsive 1 = Arousable by minor stimulation 2 = Requires repeated or painful stimulation 3 = Responds with reflex motor or unresponsive |
| Level of Consciousness Questions “How old are you?” Credit given if child states number or shows correct number of fingers. “Where is <i>family member</i> ?” <i>Family member</i> must be present. Credit given if child points to or gazes purposefully in the direct of family member. | 0 = Two answers correct 1 = One answer correct 2 = Zero answers correct |
| Level of Consciousness Commands 1. Open and close eyes 2. Touch/ show me your nose. Substitute another one step command if the hands cannot be used. | 0 = Two tasks correct 1 = One task correct 2 = Zero tasks correct |
| Best Gaze Only test horizontal eye movements. A score of 1 is given when gaze is abnormal in one or both eye, but forced deviation or total gaze paresis is not present. | 0 = Normal 1 = Partial gaze palsy 2 = Forced deviation or total gaze paresis not overcome by oculocephalic maneuver. |
| Visual For children >6 years, visual fields are tested by confrontation using finger counting. For children age 2 – 6 years visual fields are tested by visual threat. | 0 = No visual loss 1 = Partial hemianopia 2 = Complete hemianopia 3 = Bilateral hemianopia (including cortical blindness) |
| Facial Palsy Score symmetry of grimace, close eyes, raise eyebrows, and show teeth. | 0 = Normal symmetrical movement 1 = Minor paralysis 2 = Partial paralysis (total of near total paralysis of lower face) 3 = Complete paralysis of one or both sides |
| Motor Arm (Left and Right) Arms are extended with palms down at 90 degree if sitting, or 45 degree if supine. Drift is scored if the arm falls before 10 seconds. For children too young to follow directions, power in limb should be graded by observation, excluding time limits. | 0 = No drift, limb holds for full 10 seconds 1 = Drift but has some effort against gravity 2 = Some effort against gravity but limb cannot get to or maintain position 3 = No effort, limb falls 4 = No movement 9 = Amputation, joint fusion (explain) |
| Motor Leg (Left and Right) Legs are extended 30 degrees while supine. Drift is scored if the leg falls before 5 seconds. For children too young to follow directions, power in limb should be graded by observation, excluding time limits. | 0 = No drift, limb holds for full 5 seconds 1 = Drift but has some effort against gravity 2 = Some effort against gravity but limb cannot get to or maintain position 3 = No effort, limb falls 4 = No movement 9 = Amputation, joint fusion (explain) |
| Limb Ataxia Finger-nose-finger and heel-shin test bilateral. For children <5 years substitute task with reaching for toy for arms, and kicking a toy or examiner’s hand for legs. | 0 = Absent 1 = Present in one limb 2 = Present in two limbs |
| Sensory Sensation to pin prick. For children too young to report sensory loss, observe for behavioral response to pin prick. Examiner should test as many body areas as needed to check for hemi sensory loss (not hands). | 0 = Normal, no sensory loss 1 = Mild/ moderate sensory loss 2 = Severe/ total sensory loss, patient not aware of being touched |
| Best Language For children > 6 years and normal language development prior to stroke, patient should perform table 1 , and figures 1, 2, and 3 . For children ages 2–6 years score based on observation of language comprehension and speech during exam | 0 = Normal, no aphasia 1 = Mild/ moderate, some obvious loss of fluency 2 = Severe, all communication through fragmented expression 3 = Global aphasia, usable speech or auditory comprehension |
| Dysarthria If patient thought to have adequate sample of speech, aphasia can be rated by the clarity of articulation. | 0 = Normal 1 = Mild/ moderate, slurs at least some words 2 = Severe, unintelligible or mute 9 = Intubated or other barrier (explain) |
| Extinction and Inattention (formerly Neglect) Sufficient information to identify neglect may be obtain throughout testing. Since the abnormality is score only if present, the item is never untestable. | 0 = No abnormality 1 = Inattention to simultaneous stimulation in one of the sensory modalities 2 = Profound hemi-inattention or orients to only one side of space |

This tool was adapted from the Pediatric NIHSS. Words have been eliminated and descriptions have been condensed to provide single sheet reference text. Items of evaluation and scoring remain the same.

| | |
|--------------------------|--|
| Repetition | Repetition task for the following words: “Stop,” “stop and go,” “If it rains we play inside,” “The President lives in Washington.” |
| Reading | Read items in <i>Figure 3</i> |
| Naming | Name items in <i>Figure 1</i> |
| Fluency and word finding | Describe what is happening in <i>Figure 2</i> |

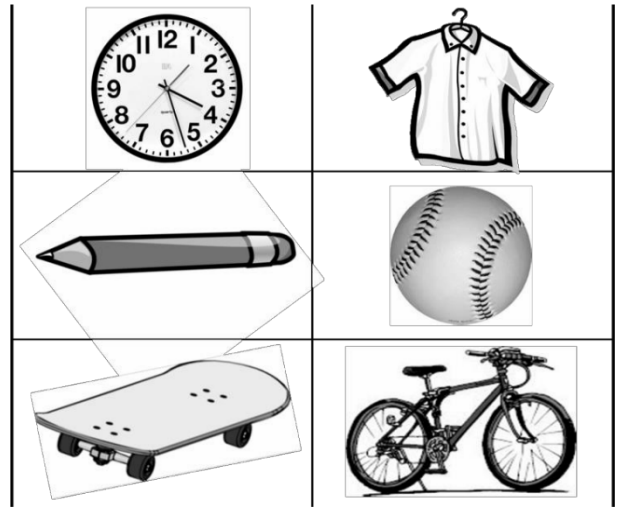


Figure 2

Table 1

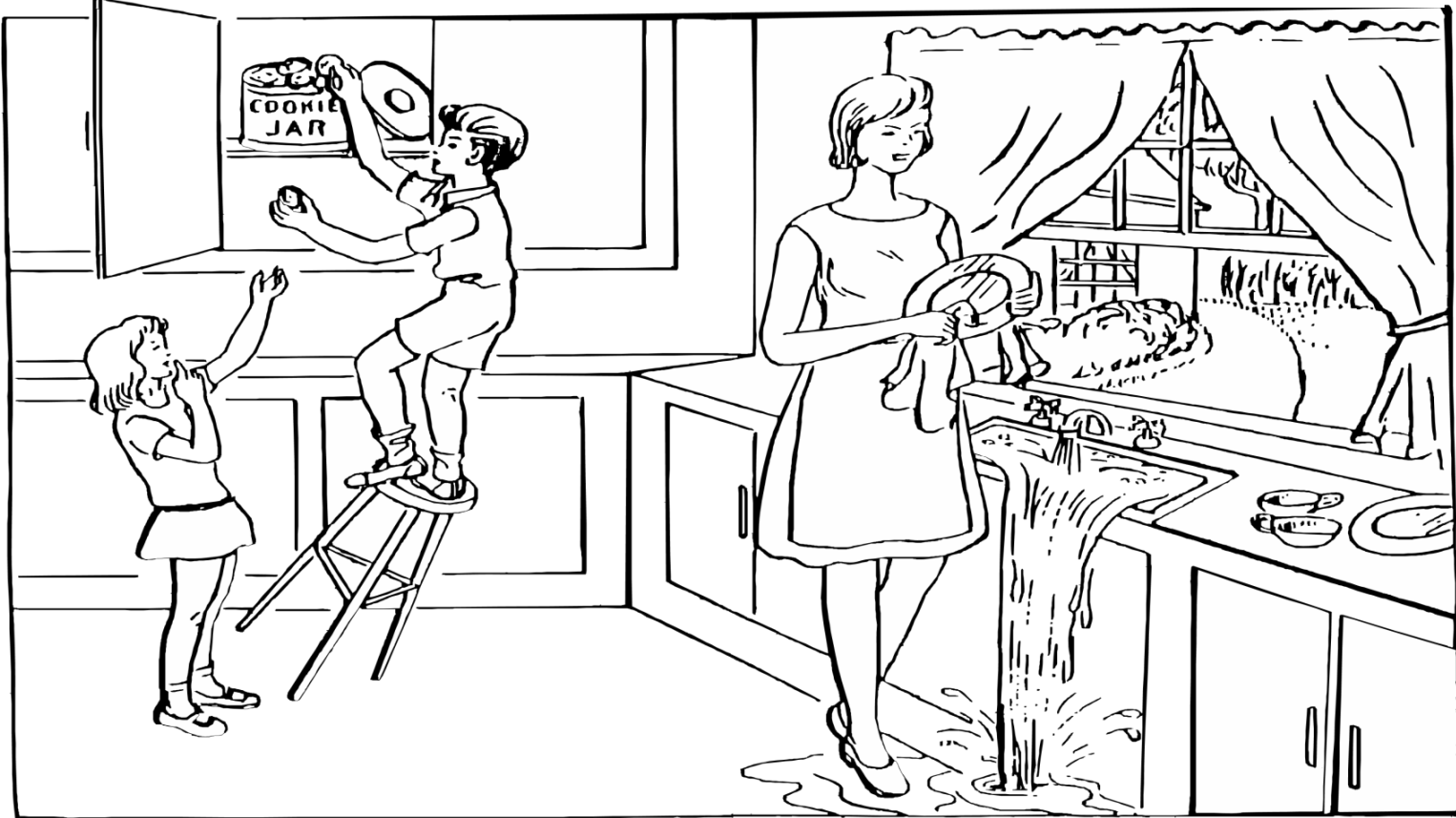


Figure 2

Stop
See the dog run
Little children like to play outdoors

Figure 3