Welcome to peds neuro consults! This is probably not going to be an overly fun 6wks, but you'll get through it like everyone before you. SLCH is a very different place than Barnes and child neurology is a very different beast than adult neurology, but you will be expected to be able to answer mommy calls and Children’s Direct (DAL) calls on your third day… so… yeah.

Schedule:
- Monday through Friday - get in ~7am and leave ~5-6pm. You will have either COH or movement clinic EVERY Tuesday afternoon. Journal club is every Tuesday at noon—you will present in one of these during your rotation. NSGY/neuro/neurorads rounds are every Thursday at 7am
  - Wednesday: home call. This means you take pagers (neuro floor, consult and NICU) from 5pm (or earlier if peds neuro resident is in clinic that afternoon) until 7AM next day. Further details on this below.
- Saturday - get in at ~7AM and leave once you’ve rounded and finished notes (varies a lot depending on the day). You start covering NICU and consult pagers at 7AM until noon. You will have a “helper” attending on until noon.
- Sunday - typically off unless there are two adult

Call Days:
- Every Wednesday starting at 5pm and ending Thursday at 7am
- Every Saturday starting at 7am and ending ~12pm
- Sometimes Sunday starting at 7am and ending at 5pm

The Consult Team: Will always be a “fellow” (mostly one of your BFF PGY-4s, but sometimes a PGY-5), you, and some med students. Sometimes you’ll be lucky and have one of the child neuro PGY-3s or another adult rotator. Attendings rotate every 2wks, fellow rotates every 1-2wks.

Locations:
- The Peds Neuro office is located in Northwest Tower (the building on top of the SLCH parking garage; you run directly into it when you’re walking down the link hallway to get to SLCH.) Go to 12th floor then walk down the only hallway available until almost the end, when you see the Peds Neuro sign/door on the left. You have to ring the doorbell to get in, then you’ll need to ask Laura Graves to submit your name/last 4 for your own actual access. This is where you’ll hang out most of the time.
- The ED is on the 1st floor near the pharmacy (opposite the cafeteria). It’s not quite as much of a maze as ours, but I still mostly got lost when I tried to find things. MRI is near there as well.
- The NICU is on the 5th floor. It is a huge place (99 beds!!) split into fanciful sections called “gardens” that make you want to punch everyone in the face.
- The PICU/CICU (most consults) are on the 7th floor.
The neuro floor is the 12th floor, and is where the EMU is. Everything between 8 and 11 doesn’t matter.

**How to Forward the Pagers: Because They’re Not Actually Real**

- Dial 314-454-7777 from any phone then press * over the initial message.
- Enter the “messaging ID” when asked followed by #.
  - Consult pager ID: 314-407-6848
  - NICU pager ID: 314-490-0085
  - Floor pager ID: 106111
- Press 1, then press 1 again after the prompts.
- Enter your messaging ID (your Spok username, which may or may not be your old pager number - sometimes it’s random numbers; check by looking yourself up in Spok) and press #.
- Listen to confirm it worked then hang up (you’ll immediately get a Spok text telling you that you’re covering the pager).

**What Happens During the Weekdays:** The consult team is responsible for non-12th floor (neuro floor) consult calls and ED calls as well as Children’s Direct calls from OSHs. The floor/PICU consults will be split between you and the fellow (and the PGY-3 if on). Follow-ups will be assigned by the fellow the day before the same way we do it.

- There will be a “PICU huddle” first thing (since we are consulted on a lot of the PICU kids, we schedule a time every morning to meet with the PICU fellow/attending of the 2 teams before they round to discuss all of the shared kids). This will be 7:45am most days, or 7:15am if there’s grand rounds.
- At 9am, you’ll meet with the EEG attending in the PICU or EMU to discuss all of the kids that you have on cEEG
- Rounds will begin after EEG rounds. Most of the attendings don’t care if you pre-round. These rounds will include olds, news from the prior afternoon, and news from overnight that the on-call person took.
- If you get consults during the day to see, some attendings will want to meet in the afternoon to get through those. Depends on attending and day.
- After rounds, it’s the usual note-writing, seeing new pts, etc.
- During weekday days, there is a fellow on the NICU who takes NICU consults and a fellow on the neuro floor who deals with floor stuff.

**What Happens on Saturdays/Sundays:** The consult and the NICU fellows are off and only you are on with the weekend “helper” attending. You will hold the consult pager and NICU pager from 7am to ~12pm on Saturdays and 7am to 5pm on Sundays. You and him/her will meet at a mutually agreed upon time in the AM to round on only the especially ill olds and news (also talk to the weekend EEG attending if you have kids hooked up). You will also be responsible for old and new NICU consults.

**What Happens on Call:** #1 - You will hate all previous decisions that led you to this particular point in your life. #2 - Weds at 5pm (earlier if fellow has clinic), you will pick up the consult
pager, the NICU pager, and the floor pager. Then will be barraged with calls until the wee hours of the morning. This is technically home call, but you will probably stay late and have to come in ~⅓ of your call days before the morning. The pagers go away at 7am.

- Contrary to Barnes, not all ED “consults” need to be seen overnight or at all. In fact, most of them don’t. You can discharge them home without seeing them or staffing with an attending, or you can admit them to neuro the same way. The only ones that NEED to be seen are stroke pages, kids in status that they can’t control, or any other emergent/urgent issue that they call about and you get the feel that a neurologist should lay eyes on them sooner rather than later. Non-urgent floor and ICU consults can be put off until morning. However, if you are coming in to see a patient, most attendings want you to call them to discuss the patient afterwards. NO ONE will fault you for calling them.

- Your goal on peds consults is NOT to be the defender of the neurology service like at Barnes. The neuro people here are very protective of their patients and almost never mind them getting admitted to the floor, even if it’s for something stupid like “mom doesn’t feel comfortable taking Timmy home because his baseline 20-30 seizures/day is up to 50/day in the context of a URI” or “these are the most obvious pseudoseizures I’ve ever seen in my life”.

- Types of calls you will receive:
  - 1. ED calls - these can usually be managed over the phone by discharging home or admitting to neuro, or if they’re getting admitted elsewhere, saying that you’ll see them in the morning. The only time you have to staff an ED consult with an attending is if you came in to see them but are still sending them home. If you admit to neuro, call the 12 senior to let them know about the kid and provide a basic plan for overnight. You can contact 12 senior by Spok. 12 workroom number: 314-454-3793
  - 2. Mommy calls - The. Worst. Getting after-hours clinic calls from (almost always) moms asking inane questions like “how do you dose Tylenol?” and “Will you refill his Onfi?” You cover all the attending & resident clinics so some of these kids are ridiculously complicated. Note: unlike Children’s Direct calls, you do not have to answer these calls within five minutes, generally you have about 1 hour until they page again. Some mommy’s are aggressive though and will page after 15-30 minutes so I generally try to return these calls within this amount of time. Also, if you are in an acute situation you can always call the number back and give them a heads up that it will be one hour and that if it is an emergency they should call 911. The general rule of thumb: answer calls, put a call note in Epic, and route the resident/attending.
  - 3. OSH calls - Through “Children’s Direct” (CD), typically EDs but sometimes from the floor. You need to return this call within 5 minutes else they will page page again- it is okay to call back if in an acute situation (stroke/status, etc) and tell them an estimated time you can return the OSH call. The reason for the call can vary from “can I discharge this simple febrile seizure home?” (yes) to “Ohgodthiskidisinstatuswhatdolovee” (Benzo, benzo, fPHT). If they want to transfer, the answer is typically yes, and the kid can be gotten to SLCH via: one-way ambulance from the OSH (for the very stable kids), having the SLCH
transport team get them via ambulance (the mildly unstable kids that you don’t want to the EMS people to break), or via air (the very sick/urgent kids). If you think you need to fly the kid, you should get ED physician on the phone to help with medical transport -they are very helpful with triaging the situation. They can either be sent to the ED (they are very sick or you have no idea what’s going on) or be direct admitted to 12 (very stable kids). Default should be to send the kids through the ED. If you send to the ED, call them (through Children’s Direct) and either talk to someone to tell them they’re coming, or leave a message with the person who picks up the phone saying “this kid’s coming, please do X,Y,Z then call me”. If you’re direct admitting to 12, the CD person will get the admitting hospitalist on the line and you have to run the kid by them to make sure they’re cool with it, then call 12 and let the senior know.

4. Floor/PICU/NICU consults - Unless the person calling the consult says it’s urgent and you need to come in to see them, you can either put this off until morning, or provide basic instructions (ie load LEV for a seizure) and put it off until morning. If you come in to see the kid, you need to write a consult note. You’ll also get calls for kids that you’re already on board for if they need urgent help with something.

5. EMU calls - The EEG tech telling you they saw a seizure on a kid you have hooked up so you can do something about it. They may or may not have run this by the EEG attending. If the seizure is subtle or tech is not sure tell them to run it by the EEG attending. You do not have access to EEG software at home.

6. Urgent calls from the 12 resident/intern about one of the kids on the neuro service - Just act like you’re the floor chief and do what sounds right. These are peds residents who know nothing about neurology, however, so be careful about trusting them.

FYI: You can always call the consult attending (for consult or floor questions), the NICU attending (on weekdays), or the EEG attending if you have questions about anything going on. They are mostly much…. kinder than our attendings when called in the middle of the night, and don’t get mad about stupid questions.

Make an overnight update email about the overnight events and send out to all the fellows on service and all the attendings on service.

Which Attending Should I Be Calling: A Fanciful Game
This sometimes changes and so always ask on signout from the fellows, who is on call at night—here is a generally rule of thumb:

During the week
- Consult questions: consult attending
- cEEG questions: the ICU EEG attending (if there is one) or the EMU attending if not. If it is after 12pm on Friday, then it’s the weekend EEG attending
- Floor questions (at night): consult attending
NICU questions (at night): NICU attending
Acute stroke pages: “stroke attending”, only the Children’s Direct person will know who that is so just ask them (same on the weekend)

On Saturday
Consult questions: the “weekend helper” attending but only until you’re done rounding. After that, the floor attending.
cEEG questions: the weekend EEG attending
NICU questions: the “weekend helper” attending until you’re done rounding, then the floor attending

Diagnostics: EEGs are way easier to get but MRIs are harder. Rules:

- **cEEG:** If you decide you want to hook up a kid in the middle of the night, congrats! There’s actually a tech there 24/7 who won’t bitch you out for wanting an EEG! All you have to do is call the EEG attending to get permission (give the quick background and why you want it), then text the EEG tech (via Spok, phone name is “SLCH EEG tech on call”). They have MRI compatible leads (gasp), which you should have them use if you think the kid’s gonna need an MRI while still on EEG.
  - The overnight EEG tech actually reads the EEGs too, then if they see a seizure they’ll either call the EEG attending to verify then text you, or they’ll just text you if it’s an obvious seizure. During the day, however, the EEGs are not being watched continuously so a kid can be in NCSE for an hour and you won’t know (literally). Also we don’t have access to the EEG program so don’t even bother trying.
  - “Automatic” cEEG hookups (aka you have to hook them up no matter what) include post-cardiac arrests, new ECMO patients, cooling babies, and severe TBIs. You do not need to ask an attending before calling EEG techs. Would send an email with one-liner to the EEG attending on-call. However, you should still come in and see the patient, unless the patient is pharmacologically paralyzed, then can ask the attending if you still need to come in. Should be a conversation with attending either way.

- **MRI:** These are not done 24/7 and they actually call in a tech to get urgent scans overnight, who are just as pissed about it as our EEG techs are. So you generally won’t get these overnight unless it’s for a stroke page or whatever. If you need an MRI overnight, you will have to touch base with the attending on call as they will get the call via Children’s Direct to okay this scan—typically the attendings will back us up on a scan we think is necessary. They will almost always get done on the day they get ordered, however, so there’s no waiting around for 3 days like at Barnes. That’s nice.

- **hCT:** You can get these 24/7, but beware that everyone thinks that CTs are the cancer-bringing devil so you have to have a really good reason to want one (ex. concern for acute hydrocephalus or bleed), otherwise you’ll just get an MRI in the morning.

- **LP:** Just like Barnes, consult service doesn’t do them unless there’s a damn good reason for it. Also the ED is much happier to do LPs than ours, so there’s less pushback that way. Also you don’t have to fight with lab medicine to get weird tests run, which is magical.
Stroke Pages: And You Thought Barnes Was Bad

This is the one area where we shine because we actually know what NIHSS stands for and how to do one. However the people around you have absolutely no idea what’s going on so it’s up to you to force things to happen, even more so than at BJH. Every stroke page (that you are concerned has a stroke-not the ones paged for a headache on a sickle cell patient that has no focal findings) gets a hyperacute MRI, no matter what, and the MRI techs don’t believe that hyperacute is a thing so it’s always a clusterf***. Just fight the man for the good of the kids (who can actually have strokes).

- The highest concern for stroke is in the sickle cell kids, and it’s very important to know that tPA and thrombectomy are not done in these kids -- if a sickler has an acute stroke, all they get is emergent exchange transfusion, which requires PICU admission and heme consult. Don’t try to tPA them.
- See the SLCH stroke guidelines, attached to your orientation email

Journal Club: No one will tell you, but you have to present at 1 journal club during your rotation. Works pretty much the same as our except the attendings come. Schedule:

7/31 Uzo-Okereke
8/21 Yechoor
10/9 Perelstein
11/20 Laws
1/8 Levasseur
2/12 Garret
3/19 Hwang
4/16 Coman
5/7 Butt
6/4 Chou

Run your article past Amanda Rogers or Alyssa Smith (ask any of the fellows for suggestions if you don’t know what to do), then post article here:

https://docs.google.com/spreadsheets/d/1kx7zuoES_QJT6JrxNYvhJgwly7SSFDYjqaDOf7PT-K8/edit?usp=sharing

Managing Kid Seizures: Most of Your Consults, and Also The Worst

Damn, s*** just got serious. Everyone uses phenobarb all the time. Also they use AEDs you’ve never heard of before. Also, weight-based dosing is a scourge upon society.

- New onset seizures:
  - If kid is <6wks, they need to be treated as sepsis rule-out with LP.
  - If kid is >6wks but have fever and focal neurologic signs (weakness, focal seizure), they should also have an LP.
- If kid is <6mos, they should probably be admitted for non-emergent evaluation (EEG/MRI) and observation. Also consider if <24mos.
- If kid had a seizure but is not back to baseline (post-ictal is much shorter in kids), they should get admitted and probably get immediate evaluation.
- If kid had multiple seizures in one day, admit.
- If ED says parents are not comfortable taking home, admit.
- If these are obvious pseudoseizures but kid has been to 3 EDs in 2 days and parents are sure something real is wrong… just admit. Psychology. Psychiatry, and PT consults actually exist and are helpful in this world.
- On the off chance that you can actually are able to send a new-onset seizure home… there exists a “new onset seizure (NOS) clinic” that will get kids seen relatively quickly. The kid has to be >6mos, developmentally normal, no other serious neurologic issues (mild ADHD OK), and not already seen by any other neurologist. Task Rachel Hillen to get this scheduled. Any sort of spell type can go in this clinic, even the syncopal sounding ones. This is the NOS clinic scheduling number: 314-454-4355

**Febrile seizures:** Mostly the ED manages these without calling, but not always and OSHs may call too. Defined as: a seizure that happens in a child >6mos and <5yrs, when temp is >38C, without meningitis/encephalitis, and without another cause for seizure (metabolic abnormality).
- *Simple febrile seizure:* This is a generalized seizure that lasts <15min and does not recur. These can be discharged home without neuro follow-up.
- *Complex febrile seizure:* Becomes complex if the seizure has a focal onset, lasts >15min, or occurs >1x in 24hrs. If the kid is >18mos, they generally do not need an urgent LP/neuroimaging, but maybe should get admitted and if not go home with Diastat.

**Seizures in a kid who has epilepsy:** Just do whatever is best for the kid, and start a klonopin bridge (dosed 0.01-0.03mg/kg div BID) if they’re sick or have another obvious reason to seize. Feel free to increase their AED if there’s an obvious change that can be made (ie monotherapy with lots of room to go).
- Certain attendings hate klonopin bridges more than others...only dose klonopin if its a bridge to something (illness, missed meds). If patient having a flurry of seizures, okay to give one dose at home while either increasing maintenance med or waiting until the morning to increase a maintenance med
- Good rule of thumb is to not change any AEDs if patient is on 3 AEDs, unless plan is already laid out in call note or clinic note

**Status epilepticus/seizures in ICU patients:** general treatment strategies + concept that you can load an AED without actually starting it scheduled [ie something we don’t do]
- Do not give Phenytoin, CBZ for primary generalized epilepsy syndromes such as Absence status or JME seizures
- Do not give Phenytoin, CBZ, lamotrigine or vigabatrin for SCN1A related seizures disorders (such as Dravet, Doose, GEFS+)
- Do not load with Depakote if mitochondrial d/o is suspected or be careful to use Depakote in under 2 years old
IV phenobarbital is preferred for Status Epilepticus in infancy (<3 months) compared to IV phenytoin.

IV Keppra is ok to use in any age, but it is not a first line med for status.

It is always better to ask your senior/attending if you are unsure what to do.

Particularly for the serious and chronically ill patients, it is best to listen to their primary caretaker. They tend to know when they should be worried and what problems/reactions to treatments their child has had in a similar situation before.

**Turning Babies Into Popsicles: AKA Therapeutic Hypothermia:** See the baby overnight with the NICU team and score them (NEAT score) with the NICU team. Then call the NICU neuro attending and talk to them. Don’t think about it too hard, it’s not worth the brain space.

- See Hypothermia Guidelines as part of your orientation materials
- Website for all things baby exam: fun fact-Dr. Larsen’s dad made this video when he was a child neurologist: [http://library.med.utah.edu/pedineurologicexam/html/home_exam.html](http://library.med.utah.edu/pedineurologicexam/html/home_exam.html)
- Try to get in to see this in action during the day sometime, you can just tag along with the NICU team

Miscellaneous things:

- Vomiting up of AEDs: redose full dose if <30 min or see the medicine in the vomit.
- Task Erika Ramirez for NF clinic follow up
- Parking: On Saturdays can park in Tier 1 at the BJH garage; then coming in from home on home call, pull a ticket as enter garage and then stop at the second floor info desk on the way out to ask for a parking voucher to exit. You will be asked to sign your name and department.
- If a patient does not have a chart yet (because they are at an outside hospital etc), then you can write the information in an email and email it to Laura Graves requesting that she make an Epic chart for the patient. She will then scan your email into the patient’s new chart as a call note.
- NEVER tell families that someone will call them (unless you plan to call them yourself). Just tell them to call in the morning.
- Any procedure that needs to be done that night before admission should be done in the ER (placing IVs, drawing blood/urine, basic or advanced radiology, and LPs).
- There is no real urgent consult clinic, there are urgent slots, likely 6 per week. However the priority of these are given to PCP’s calling NICU attending during business hours. This process if still in flux. The best thing to do if a child needs to see a neurologist urgently would be to admit the child to neurology, or to medicine with neurology consult.
Comprehensive Stroke Guidelines for St. Louis Children’s Hospital (v.1)
May 2018

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I. St. Louis Children’s Hospital Guidelines for Evaluation of Acute Stroke

Scope: The purpose of these guidelines is to provide guidance and ensure consistency in the management of suspected and/or confirmed acute arterial ischemic stroke (AIS) in children at St. Louis Children’s Hospital. They are not intended for use in children in the neonatal ICU (please see NICU guidelines). Children with sickle cell disease have disease-specific treatment for suspected stroke. Please see the Sickle Cell Acute Cerebrovascular Event Management Guidelines for management recommendations. With all children, treatment should be tailored to the medical needs of individual patients.

Background: The incidence of pediatric AIS is approximately 3 per 100,000 children. Mortality rate ranges from 3-6% \(^{1,2}\) and approximately 60% of patients will have long-term neurologic deficits as a result of AIS \(^{3}\). Unlike adults, many acute focal neurologic symptoms in children do not actually represent stroke, therefore, correct diagnosis is of utmost importance to guide management\(^4\).

Initial Assessment: Stroke is a medical emergency. Early suspicion and timely recognition of possible stroke is critical for accurate diagnosis and appropriate management.

1. Signs and symptoms of stroke include (but are not limited to):
   a. New focal weakness, often unilateral
   b. Sensory changes, often unilateral
   c. Visual changes
   d. Loss of balance, vertigo
   e. Aphasia
   f. Dysarthria or slurred speech
   g. “Worst headache of my life”
   h. Encephalopathy or altered level of consciousness

2. If stroke is suspected, call Children’s Direct (314-474-7001) to activate Stroke Alert
   a. A page will be sent to the following members
      i. Pediatric Neurology Fellow
      ii. Pediatric Neurology Stroke Alert Attending
      iii. MR technologist
      iv. Pediatric Emergency Medicine attending
      v. Pediatric Emergency Medicine fellow
      vi. Pediatric Emergency Medicine charge nurse
      vii. SLCH administrative supervisor

3. Document:
   a. Neurological findings at time of initial presentation and time of examination
      i. Pediatric Neurology Fellow will repeat exam and document PedNIHSS [appendix 1]
      ii. Repeat neurologic examination q15 minutes to monitor for progression
b. Vital signs q15 minutes
c. Last time the child was seen well (at baseline) - “Time Last Seen Well” (TLSW)
d. Last time the child ate or drank anything

4. Place IV and obtain the following STAT labs:
   a. Blood glucose
   b. CBC
   c. BMP
   d. Type and Screen
   e. PT/PTT
   f. Fibrinogen
   g. 12-lead ECG
   h. Urine comprehensive drug screen
   i. Blood gas, if clinically indicated
   j. Urine beta-HCG, if female 13 years or older
   k. If child is on any anticoagulation, please also send anti-Xa and platelet mapping TEG

Initial Management: Goal is to minimize further injury and maintain cerebral perfusion.
1. Assess ABC’s (Airway, Breathing, Circulation), maintain SaO2 >/= 92%
2. Assess for trauma, based on history
3. Keep child flat to maintain cerebral perfusion. If concern for possible increased intracranial pressure (obtunded +/- hypertension +/- bradycardia), keep the head of bed at 30 degrees and notify neurosurgery
4. Strict NPO
   a. Treat hypovolemia with normal saline (NS); do not give hypotonic fluids.
6. Temperature Regulation: If temperature >38.0, remove any extra blankets or coverings to allow passive cooling.
   a. Consider acetaminophen 15mg/kg per rectum; do not give anything PO.
7. Glucose Control: Maintain glucose between 80 – 200 g/dL.
   a. Check glucose q8 hours until stable for 24 hours. Check more frequently if actively treating hypo- or hyperglycemia.
8. If not already in a pediatric ICU, admit to the pediatric ICU for close neuromonitoring (this may be deferred until after hyperacute treatment, if patient is eligible)

Neuro-Imaging: Time sensitive interventions may be available to those patients presenting within 4.5 hours of symptoms onset, or, for select patients, within 16-24 hours. For this reason, STAT neuroimaging for diagnosis confirmation is critical.
1. A non-contrast head CT should be obtained. CT has low sensitivity for detecting early ischemia, but is excellent for identifying skull fractures or intracranial bleeds.
a. If skull fracture or intracranial hemorrhage is identified, please consult neurosurgery immediately for further recommendations.

2. A **brain MRI hyperacute protocol with perfusion + Magnetic Resonance Angiogram (MRA)** [appendix 3] can confirm ischemic stroke, or help identify alternative diagnoses including hemorrhagic stroke, or posterior reversible encephalopathy syndrome (PRES). MRA must be ordered as a separate order in addition to the hyperacute brain MRI.
   a. MRA of head and neck can detect possible dissection, vasculopathy, or large vessel arterial occlusion.
   b. MRV can diagnose cerebral venous sinus thrombosis.
   c. For either vascular study, non-contrast time-of-flight (TOF) imaging may be obtained initially, but may need to be repeated with contrast.

3. CT-angiogram of the head and neck is a more definitive examination for vascular abnormalities than MRA, but carries additional radiation exposure and may require anesthesia. **If a patient cannot safely obtain an MRI, and clinically suspected large vessel occlusion, obtain CTA.**

**TpA Criteria:** Patients may be eligible for hyperacute tPA (alteplase) for possible treatment at SLCH if:

a) 14 years of age or older (post-pubescent),

b) *Previously neurologically high functioning*

c) Present within 4 hours of “last known normal” time
   
   i. If basilar artery stroke or stroke with CTA positive large vessel occlusion (ie. Middle Cerebral Artery), presentation within 24 hours of “last known normal”, the patient may be considered for mechanical thrombectomy by interventional neuroradiology.

d) Clinical diagnosis consistent with ischemic stroke causing measureable neurologic deficit. PedNIHSS >2 or debilitating symptoms (i.e. aphasia), but not >25 if last known well time is 3-4 hours.
   
   a. If hyperacute MRI can be obtained within 4-hour window from time last known well, it is preferable to confirm diagnosis of ischemia, given high rate of stroke mimics in pediatrics. However, if obtaining MRI would not be possible within tPA window, and patient is eligible based upon other inclusion/exclusion criteria, may give tPA without imaging confirmation.

e) Exclusion criteria:
   
   a. Intracranial hemorrhage on head CT
   b. Serious head trauma in prior 3 months
   c. Active bleeding or suspected underlying abnormality, including but not limited to:
      
      i. INR >1.7
      ii. Platelet count <100,000
      iii. Patient has received *heparin* within 24 hours AND has elevated PTT >40
      iv. Patient has received treatment (not prophylactic) doses of injectable anticoagulants (*enoxaparin, dalteparin, tinzaparin, fondaparinux*) in past 48 hours.
      v. Patient takes *apixaban* and last dose administered within 24 hours (36 hours if moderate-severe renal impairment, i.e. Cr Cl <30 ml/min).
      vi. Patient takes *dabigatran* and thrombin time >30.
vii. Patient takes *rivaroxaban*, and PT >13 and anti-Xa>0.1

d. On repeated measurement, SBP > 160 and DBP >100 at time of alteplase bolus administration.
e. High suspicion for subarachnoid hemorrhage, despite no hemorrhage on CT
   (e.g. “thunderclap headache”, family history of aneurysm)
f. Pregnant females

f) Relative exclusion criteria:
   a. Major surgery or serious trauma within last 14 days
   b. Intracranial or intraspinal surgery within previous 3 months
   c. Serum glucose <50 mg/dL or >400 mg/dL
   d. Stroke within previous 3 months
   e. History of intracranial hemorrhage (does not include hemorrhage associated with
      prematurity)
f. GI or GU hemorrhage within 21 days or known structural GI malignancy at risk
   of bleeding
g. Intracranial or intraspinal neoplasm
h. Arterial puncture at non-compressible site (e.g. subclavian artery)
i. Intracerebral aneurysm
j. Arteriovenous malformation. May consider alteplase in patients with known
   AVM having moderate to severe ischemic stroke with morbidity and mortality
   felt to outweigh the risk of ICH.
k. Rapidly improving symptoms
l. Frank hypodensity on head CT. Frank hypodensity suggests irreversible ischemic
   injury and risks of alteplase likely exceed benefit.

g) Other considerations:
   a. Alteplase may be considered for patients presenting with acute ischemic stroke,
      even if they have undergone lumbar dural puncture in preceding 7 days.
b. Menstruation: Because potential benefits of alteplase in an acute ischemic stroke
   likely outweigh risks of serious bleeding in patients with recent or active
   menorrhagia without clinically significant anemia or hypotension, alteplase
   administration may be considered. For patients with active vaginal bleeding who
   receive alteplase, degree of vaginal bleeding should be monitored for 24 hours
   after alteplase.

If Pediatric Neurology Stroke Attending and Fellow agree that patient meets above
criteria, team member will CALL CHILDREN’S DIRECT at 314-747-7001 to send TPA
GO ALERT, providing patient name and location.

See SLCH tPA guidelines (p. 6) for further details of administration and management

**Thrombectomy Criteria**

A patient with a large vessel occlusion (MRA/CTA) and evidence of penumbra (MRI perfusion/
diffusion mismatch) may be eligible for thrombectomy up to 24 hours of time last seen well.

Please see thrombectomy guidelines (p 11) for further details of evaluation and procedure.

**After stroke evaluation complete (+/- imaging), call Children’s Direct (314-747-7001) to
send notification of Stroke Alert complete.**
II. Tissue Plasminogen Activator (tPA/Alteplase) Evaluation and Administration

**Background:** Intravenous tissue plasminogen activator (tPA, aka Alteplase) is standard of care for adults ages 18 years and older with signs and symptoms of acute ischemic stroke who meet selection criteria. Although at present there is insufficient evidence to support treating children with tPA under the 18 years of age, Washington University pediatric and adult stroke physician experts agree that it is reasonable to consider intravenous tPA treatment for pediatric stroke patients, as no alternative treatments are available to reduce long-term disability in this patient population. Pediatric stroke patients as young as 14 years of age might be considered for treatment with tPA on a case-by-case basis. Therefore, all children between 14-18 years of age should be evaluated for possible brain-saving treatment and offered tPA if deemed a good candidate for treatment.

**Evaluation**

If suspect patient may be a tPA candidate, immediately place 2 large bore IVs, and confirm the following STAT labs have been obtained:

- CBC, PT/PTT, fibrinogen, whole blood electrolytes, BUN, Creatinine, type and screen.

**TPA Criteria:** Patients may be eligible for hyperacute tPA (alteplase) at SLCH if:

- h) 14 years of age or older (post-pubescent),
- i) Previously neurologically high functioning
- j) Present within 4 hours of “last known normal” time
  - ii. If basilar artery stroke or stroke with CTA positive large vessel occlusion (ie. Middle Cerebral Artery), presentation within 24 hours of “last known normal”, the patient may be considered for mechanical thrombectomy by interventional neuroradiology.
- k) Clinical diagnosis consistent with ischemic stroke causing measureable neurologic deficit. NIHSS or PedNIHSS >2 or debilitating symptoms (i.e. aphasia), but not >25 if last known well time is 3 -4 hours.
  - a. If hyperacute MRI can be obtained within 4-hour window of time last known well, it is preferable to confirm diagnosis of ischemia, given high rate of stroke mimics in pediatrics. However, if obtaining MRI would not be possible within tPA window, and patient is eligible based upon other inclusion/exclusion criteria, may give tPA without imaging confirmation.
- l) Exclusion criteria:
  - a. Intracranial hemorrhage on head CT
  - b. Serious head trauma in prior 3 months
  - c. Active bleeding or suspected underlying abnormality, including but not limited to:
    - i. INR >1.7
    - ii. Platelet count <100,000
    - iii. Patient has received *heparin* within 24 hours AND has elevated PTT >40
iv. Patient has received treatment (not prophylactic) doses of injectable anticoagulants (*enoxaparin, dalteparin, tinzaparin, fondaparinux*) in past 48 hours.

v. Patient takes *apixaban* and last dose administered within 24 hours (36 hours if moderate-severe renal impairment, i.e. Cr Cl <30 ml/min).

vi. Patient takes *dabigatran* and thrombin time >30.

vii. Patient takes *rivaroxaban*, and PT >13 and anti-Xa>0.1

d. On repeated measurement, SBP > 160 and DBP >100 at time of tPA bolus administration.

e. High suspicion for subarachnoid hemorrhage, despite no hemorrhage on CT (e.g. “thunderclap headache”, family history of aneurysm)

f. Pregnant females

m) Relative exclusion criteria:

a. Major surgery or serious trauma within last 14 days

b. Intracranial or intraspinal surgery within previous 3 months

c. Serum glucose <50 mg/dL or >400 mg/dL

d. Stroke within previous 3 months

e. History of intracranial hemorrhage (does not include hemorrhage associated with prematurity)

f. GI or GU hemorrhage within 21 days or known structural GI malignancy at risk of bleeding

g. Intracranial or intraspinal neoplasm

h. Arterial puncture at non-compressible site (e.g. subclavian artery)

i. Intracerebral aneurysm

j. Arteriovenous malformation. May consider tPA in patients with known AVM having moderate to severe ischemic stroke with morbidity and mortality felt to outweigh the risk of ICH.

k. Rapidly improving symptoms

l. Frank hypodensity on head CT. Frank hypodensity suggests irreversible ischemic injury and risks of tPA likely exceed benefit.

n) Other considerations:

a. tPA may be considered for patients presenting with acute ischemic stroke, even if they have undergone lumbar dural puncture in preceding 7 days.

b. Menstruation: Because potential benefits of tPA in an acute ischemic stroke likely outweigh risks of serious bleeding in patients with recent or active menorrhagia without clinically significant anemia or hypotension, tPA administration may be considered. For patients with active vaginal bleeding who receive tPA, degree of vaginal bleeding should be monitored for 24 hours after tPA.

If Pediatric Neurology Stroke Attending and Fellow agree that patient meets above criteria, team member will CALL CHILDREN’S DIRECT at 314-747-7001 to send TPA GO ALERT, providing patient name and location.
“TPA GO” alert is sent to the following individuals:

1. Pediatric Neurology Fellow
2. Pediatric Neurology Stroke Attending
3. Inpatient Pharmacist
4. Pediatric Emergency Attending
5. Pediatric Emergency Fellow
6. Pediatric Emergency Charge Nurses
7. Pediatric Intensive Care Charge Nurse
8. Administrative Supervisor

TPA Administration

1. Pharmacy will come to bedside to confirm and provide tPA dose as follows:
   a. For <100 kg, give 0.9 mg/kg, with 10% (0.09 mg/kg) given immediately as an IV push, and the remaining 0.81 mg/kg over 1 hour.
   b. For 100 kg or greater, give 90 mg total, 9 mg as IV push, followed by 81 mg over 1 hour.

   If pharmacy is not at the bedside within 10 minutes of TPA GO activation, call (314) 319-1957.

2. Obtain vital signs, including heart rate, blood pressure, pulse oximetry, and respiratory rate immediately PRIOR to giving tPA IV push. Confirm BP <160/100.
3. If patient develops severe headache, acute hypertension, nausea, vomiting, or has worsening neurologic examination, discontinue the infusion and obtain a STAT Head CT.
4. Once tPA bolus given and infusion started, blood pressure (BP) should be monitored closely according to the following guidelines:
   a. Hours 0-4 BP recorded at least every 15 minutes
   b. Hours 4-8: BP recorded at least every 30 minutes
   c. Hours 8-24: BP recorded at least every 1 hour
   d. Hours 24-48: BP recorded at least every 2 hours
   e. Hours 48-72: BP recorded at least every 4 hours

Blood pressure management

1. Goal Systolic BP is at least 50th percentile for age (110 mmHg for 14-17-year-old), and not greater than 15% above 95th percentile for age (>150 mmHg for 14-year-old, >160 mmHg for 17-year-old, using 95th percentile SBP of 130 mmHg for 14-year-old, and 140 mmHg for 17-year-old).
2. If SBP is greater than 20% above 95th percentile for age (>156 mmHg for 14-year-old, or >168 mmHg for 17-year-old), treat upon confirmation of BP measurement accordingly:

   N.B. Do not drop SBP more than 25% of original value. If hypotension occurs, consider IV NS bolus, discontinuing nicardipine infusion and/or reversing anti-hypertensive effects
For patients 50 kg or greater:

Hydralazine 10 mg IV. May repeat after 10 minutes 1-2 times as needed. May also consider Labetalol 10 mg over 1-2 minutes, if heart rate >70 bpm.

OR

Nicardipine infusion : 5 mg/h, titrate by 2.5 mg/h at 5-15 minute intervals, maximum dose 15 mg/h; when desired blood pressure is reached, immediately reduce infusion to 3 mg/h.

For patients less than 50 kg:

Hydralazine 0.1 mg/kg IV. May repeat after 10 minutes 1-2 times as needed. May also consider Labetalol 0.2 mg/kg over 1-2 minutes, if heart rate >70 bpm.

OR

Nicardipine infusion: 0.5 mcg/kg/min, titrate by 0.5 mcg/kg/min at 5-15 minute intervals, maximum dose 5 mcg/kg/min; when desired blood pressure is reached, immediately reduce infusion to 0.3 mcg/kg/min.

After infusion started and blood pressure stable and within goal range, patient should be admitted to PICU or CICU for further monitoring and care

Post-TPA care to include:

1. Vital signs q15 minutes x 4 hours, then q30 minutes x 4 hours, then q60 minutes x 16 hours post-alteplase administration. Notify provider if blood pressure >95th, and treat as detailed above.
2. Neurological Checks q 15 minutes x 2 hours, then 30 minutes x 6 hours, then q60 minutes x 16 hours post-alteplase administration. Notify provider for any change in patient’s neurologic parameters.
3. Pulse oximetry.
4. During infusion and 8 hours post-tPA: Closely monitor potential bleeding sites; avoid IM/SQ injections; no rectal temperature/suppositories; no lumbar punctures; no urinary catheterization; carefully perform venipunctures only if necessary; avoid arterial punctures; no indwelling central catheters to be pulled or placed, avoid nasogastric tube placement.
5. Avoid physical therapy/chest physiotherapy and nonessential handling of patient during first 8 hours post-tPA administration.
6. Post Sign at Bedside: During infusion and 8 hours post-tPA, post sign at head of bed indicating the use of systemic thrombolytics to notify all providers about patient’s high bleeding risk.
Standard post-stroke care including:

1. Head of bed flat unless significant concern for increased ICP.
2. Intravenous fluids: 0.9% Normal Saline (NS) at 1.5 L/m². Add dextrose if needed to maintain glucose >80 mg/dL. Fluid resuscitate with NS as needed for dehydration or hypotension.
3. PedNIHSS documented by nurse on admission, qshift, and with concerns for significant change in neurologic status. Notify MD if change in score by 2 or more points (better or worse). Neurology fellow to confirm PedNIHSS on admission and repeat with consultations.
4. PT/OT/ST consult (after 8 hours post-alteplase administration).
5. NPO until cleared by speech therapy if ANY facial or speech deficits during acute stroke symptoms.

Please refer to “Pediatric Ischemic Stroke Inpatient Care” (p.22) for further workup and management recommendations.
III. Acute Ischemic Stroke Endovascular Treatment (Thrombectomy)

**Background:** Mechanical thrombectomy for patients with acute ischemic stroke and evidence of large vessel occlusion (LVO) on imaging who present within 6 hours of symptom onset became standard of care for adults in 2015 after multiple large, randomized clinical trials were published demonstrating efficacy.\(^1\) Although the trials were limited to patients 18 years and older, the pathophysiology of ischemic stroke with LVO in younger teenagers ages 14-17 is not meaningfully different from the young adult (18-30 year old) patients who were included in the trials. Additionally, case series and reports of pediatric thrombectomy demonstrate potential benefit.\(^2,3\) As such, this guideline offers procedural details for the rapid evaluation and management of teenage patients with acute ischemic stroke due to LVO.

This guideline is part of the comprehensive pediatric stroke guidelines. Please see Comprehensive Pediatric Stroke Guidelines for further details of stroke alert and management prior to imaging and for patients who do not meet inclusion criteria listed below.

-----------------------------------------------------------------------------------------------------------------------------

**Inclusion criteria:**

1. Patient age 14 years or older*  
2. Clinical symptoms consistent with acute ischemic stroke.  
3. Time last seen well (TLSW) 24 hours or less from activation  
4. Radiographic evidence of LVO (see below for radiology details)

*Younger patients may be considered on a case by case basis after discussion with neuro-interventional radiology (NIR) attending.

**Radiology Confirmation:**

Large Vessel Occlusion may be demonstrated by either CT Angiography (CTA) or Magnetic Resonance Angiography (MRA). Imaging choice is at discretion of evaluating neurologist, based on patient factors, immediacy of imaging availability, and time window.

1. CT or MRI  
   a. Absence of intracranial hemorrhage, mass, or mass effect as a cause of presenting symptoms  
   b. DWI confirmation of ischemia (MRI)  
   c. ASPECTS ≥ 6 (if head CT is available)

2. CTA or MRA  
   a. **Large vessel occlusion** involving M1, ICA, vertebral artery, or basilar artery  
   b. Absence of vascular anatomy or concomitant vascular lesion that would substantially impede safe endovascular treatment

3. CTP or MRI (based on TLSW, (Ped)NIHSS)  
   a. If treatment expected **within 16 hours** of TLSW and **PedNIHSS or NIHSS ≥ 6**,  
      core infarct volume ≤ 70 mL, perfusion Mismatch Ratio > 1.8  
   b. If treatment expected **within 16-24 hours** of TLSW and **PedNIHSS or NIHSS ≥ 10**, core infarct volume ≤ 30 mL

CT/CTA should be pushed to PICOM as each series loads for NIR to review (instead of batching at end). MRI/MRA results should be sent via RAPiD software. Pediatric neurology fellow to call neuroradiology
fellow (314-508-3767) to notify of STAT read. If unable to reach after hours, call Barnes neuro reading room at 314-362-2562.

Exclusion criteria:

1. Pre-morbid disability (mRS ≥ 2) or comorbidities that will affect recovery potential (see Appendix for modified Rankin Scale)
2. Intracranial hemorrhage, mass, or mass effect as cause of stroke symptoms
3. Current severe uncontrolled hypertension (e.g. SBP > 185 mmHg or DBP > 110 mmHg) despite reasonable efforts to treat
4. Bleeding diathesis**
   a. Use of warfarin (Coumadin, Jantoven, Marevan): INR > 3.0
   b. Use of dabigatran (Pradaxa, Pradax, Praza-ka): TT > 30 sec and PTT > 70 sec (wait for manual TT correction to rule out heparin contamination)
   c. Use of rivaroxaban (Xarelto): PT > 20 sec or αFXa > 0.5 U of heparin activity (check only if PT 15-20 sec; takes 30 minutes to run)
   d. Use of apixiban (Eliquis): αFXa > 0.1 U of heparin activity
   e. Use of edoxaban (Savaysa): αFXa > 0.1 U of heparin activity
   f. Thrombocytopenia: platelet count < 30,000/microliter

** Correction of bleeding diathesis may be considered on a case-by-case basis following discussion between neurology and INR teams.

Request for thrombectomy:

Pediatric Neurology Fellow should call Children’s Direct (314-747-7001) to reach NIR fellow and Pediatric Neurology Stroke attending to discuss case once imaging demonstrates arterial occlusion.

Children’s Direct can reach NIR fellow:

Mon-Fri 8 AM-5 PM @ reading room (314-362-2584) or reception desk (314-362-7113).
If no answer, page NIR fellow at 314-766-4482.
Mon-Fri 5PM-8AM and weekends/holidays: page 314-766-4482 and leave call back number on voicemail page.

Peds neuro fellow to communicate details to NIR as follows:

“I am calling about a potential thrombectomy.

This is a [#]-year-old [boy/girl]; imaging is available on [PICOM/ClinDesk] under [patient name].

Basic stroke details:
Imaging shows occlusion of [artery].
Last known normal was [#] hours ago.
NIH stroke scale is [#] due to [symptoms].
rt-PA [was/was not] given. [If not, why?]

Basic medical history:
Baseline mRS is [#].
Pertinent past medical history includes [...].
Relevant medications include [...].
Basic contact info

The stroke attending is [name]. My name is [name] and I can be reached at [phone #].
Consent can be obtained from [patient/family/two physicians] at [phone #].

Who will be the neuroIR attending for the case?"

Case Decision and Team Activation:

Once decision made, NIR fellow to call Children’s Direct (314-747-7001)

NO: request to connect with pediatric neurology fellow and attending to notify of decision

YES: request Children’s Direct to activate “Thrombectomy” (see Figure 1).

“Thrombectomy” text page of Name, DOB, and patient current location will be sent to following individuals:

1. NIR fellow (314-766-4482) and attending to confirm page sent. If NIR fellow does not receive page within 2 minutes after activation, call Children’s Direct to resend page.
2. Pediatric Anesthesia First Call Attending (SmartWeb on call) to head to NIR suite (Mallinkrodt 326, map appendix A) for set-up. Attending should also call BJH Trauma Anesthesia Attending 314-747-4363 to discuss coordination of anesthesia resources.
3. Pediatric 3rd call Anesthesia Attending (SmartWeb on call) for back-up if First Call Attending not available for case.
4. Pediatric Anesthesia On-call resident/fellow (314-491-1111) goes immediately to patient for pre-anesthesia assessment and consent.
5. BJH Trauma Anesthesiology 314-747-4363 for awareness of case and to assist pediatric anesthesia team as needed.
6. OR Nurse (314-574-7324) to book off-site case.
7. Pediatric Critical Care attending to escort patient to NIR suite (Mallinkrodt 326, map Appendix A). The PICU (314-319-9878) and CICU (314-471-4212) attending to have brief, direct conversation about who will accompany patient and which unit will admit patient post-thrombectomy.
8. Pediatric CCM patient care tech to bring transport and airway equipment and stretcher to patient, if patient is not in SLCH Emergency Room. Zoom stretcher is located in APC room 10.
9. Pediatric Neurology fellow (314-407-6848) to confirm case acceptance and communicate to family and current providers. Pediatric Neurology Fellow will also accompany parents to NIR suite.
10. Pediatric Stroke Attending to confirm case acceptance, and call BJH admitting @ 314-747-9635 to provide patient name and DOB notify them to meet patient in 3rd floor Neurointerventional Radiology Suite.

To prepare for transfer:
1. If patient is in ED, ED to print chart to take with the patient to NIR. Also bring extra set of monitoring leads.
2. If patient is not in ED, the PICU PCT will bring transport stretcher, monitor, and airway equipment.
3. If occurs during night shift, ICU attending may call ICU Backup attending to come in to assist with unit management while ICU attending off-site, if needed.

MAP TO NIR SUITE IS AVAILABLE AS APPENDIX ___ AND ALSO AVAILABLE IN iBOOK APP ON ICU ATTENDING PHONES

Patient Arrival to NIR Suite:

IF BJH admitting not present on arrival—call 314-747-9635 for quick registration.

ICU attending hand-off to peds anesthesia according to following script:

This is [Name], [DOB], who was last seen well at [LKW time]. S/He had [presenting stroke symptoms] at [stroke symptom onset time]. His/her initial NIHSS at SLCH was [PedNIHSS score]. The [CTA/MRA] demonstrated occlusion of [vessel occluded]. His/her past medical history is significant for [pertinent PMH]. Home medications include [home medications]. Allergies are [Allergies]. S/He last ate [food/drink] at [time]. Consenting parent(s) is/are [parent name] who are in [parent location] with [person staying with parents]. Since thrombectomy activation, [no change/notable events/change in status/meds provided].

ICU attending to provide number for anesthesia to call when case completed to make sure room is ready and notify whether or not patient will be intubated upon arrival.

Anesthesia to transport patient back to SLCH ICU (PICU or CICU, depending on patient past medical history).

Post-thrombectomy care:

1. Vital signs q 15 minutes x 4, q30 minutes x 2, then q1 hour.
   a. Blood pressure should be maintained > 5th percentile for age (see Appendix)
   b. O₂ saturation should be maintained > 92%.
   c. PedNIHSS on arrival to ICU.
   d. Maintain temperature <= 38° C. Acetaminophen should be ordered for prn temperature >38° C. (Reminder: strict NPO until cleared by speech therapy, may give rectally or per nasogastric tube).
2. Neurovascular checks: check puncture site, pedal pulses, extremity perfusion q 15 minutes x 4, q30 minutes x 2, q1 hour x 2, then minimum of q4 hours. If any bleeding observed at puncture site, hold pressure for 20-30 minutes and notify MD.
3. Strict bed rest, including flat time with straight extremity until time recommended by NIR. If necessary, may use dexmedetomidine infusion to protect puncture site during flat time.
4. Neurological checks: q 15 minutes x 4, q30 minutes x 2, q1 hour x 9, then frequency determined by clinical team. Recommend a minimum of q2 hours for first 24 hours.

5. If patient did not receive tissue plasminogen activator (tPA), and is not receiving anticoagulation, should receive 325 mg aspirin daily.

6. Strongly consider EEG monitoring, particularly if patient remains intubated on arrival to ICU.

7. Echocardiogram to evaluate for thrombus.

Standard post-stroke care including:

8. PedNIHSS should be done on admission, qshift, and with any concern for significant change in neurologic status. Routine neuro checks should repeat abnormal sections of stroke scale in order to closely follow neurologic deficits, but do not need to include entire assessment.
   a. Until EPIC implementation, document score under “Neuro Assessment comment section”.
   b. Notify MD if change in score of 2 or more points (better or worse).

9. IVF of NS + 20 KCl @ 1.5 L/m²

10. PT/OT/speech consult within 24 hours of admission.

11. NPO until cleared by speech therapy.

Please refer to “Pediatric Ischemic Stroke Inpatient Care” (p. 22) for further workup and management recommendations.

LVO= Large Vessel Occlusion
PedNIHSS= Pediatric National Institute of Health Stroke Scale
NIR= Neuro Interventional Radiology
IV. Sickle Cell Acute Cerebrovascular Event Management Guidelines

**Background**

Children with sickle cell disease (SCD) are at risk for acute cerebrovascular events, most commonly ischemic stroke. Up to 10-25% of children with SCD may experience a stroke between childhood and young adulthood. Studies have shown that children who are treated with an exchange transfusion at the time of their first stroke have decreased risk for subsequent strokes compared to children who are treated with either a simple transfusion or no transfusion. Not all neurologic changes represent a stroke, but a rapid evaluation is needed to determine whether or not the presenting symptoms are consistent with stroke and require further urgent intervention.

The main goal of treatment is to avoid progression of irreversible brain injury. Therefore, exchange transfusion (preferred therapy) or simple transfusion (if exchange transfusion is contraindicated) constitutes the main priority of management in patients experiencing acute stroke. However, neuroimaging (MRI) should be obtained as soon as reasonably possible as diagnoses such as central venous thrombosis, reversible posterior leukoencephalopathy (RPLE) and other cerebrovascular abnormalities (Moya-Moya, vascular dissection) can significantly influence management in the acute and sub-acute phase of care.

**Indications** The purpose of this guideline is to apply best clinical practice to the acute management of children with known SCD who present with an acute change in neurologic function, suggestive of stroke.

**Inclusion criteria:**

1. Known sickle hemoglobinopathy.
   a. Evidence is strongest for providing an exchange transfusion to treat stroke in children with Hb SS, Hb S-beta zero thalassemia, or rarely Hb S-beta plus thalassemia, or other types of SCD with low baseline hemoglobin, as these are the SCD patients who are most likely to experience an ischemic stroke.
   b. Patients with Hb SC or Hb S-beta-thalassemia with high baseline hemoglobin warrant rapid evaluation, but have a much lower overt stroke risk. Furthermore, appropriate short- and long-term treatment strategies have not been defined for this patient population. Therefore, they are less likely to benefit from emergent exchange transfusions. Nonetheless, careful consideration must be given to diagnostic and treatment options.
2. Acute (<72 hours*) change in neurologic function (including but not limited to unilateral motor/sensory deficits, change in speech, change in mental status, seizure)

* Please see below for guidelines of management of symptoms which have been present greater than 72 hours.
Assessment

Both the hematology fellow and neurology fellow should be notified/consulted immediately. This notification is most efficiently achieved by calling Children’s Direct (314-747-7001) and asking to activate a Pediatric Stroke Alert for a patient with sickle cell disease. This will include Pediatric Stroke Alert Team members, as detailed in Section I, as well as hematology fellow and attending.

A clear, complete neurologic exam must be documented with time and date immediately. The neurology fellow will repeat and confirm physical exam findings (within 30 minutes of consult), however, this step should not delay care.

1. Key components of the neurological exam, which should be documented before exchange transfusion, within 4-12 hours post transfusion, at discharge from the ICU and at discharge from the hospital:
   a. Mental status (Awake/alert, sleepy but arousable, lethargic)
   b. Speech (clear and comprehensible, dysarthric?)
   c. Visual fields, pupils, facial strength and symmetry
   d. Motor function in all four limbs
   e. Sensation in all four limbs (clearly document location and extent of any abnormalities)
   f. Pediatric NIH Stroke Scale Score (to be documented by the Neurology Fellow) (Appendix 3)

2. Complete vital signs, including blood pressure, should be documented every 15 minutes for at least the first hour of neurologic change.
3. If the patient is admitted to the floor, consider calling the Rapid Response Team if vital signs are abnormal or concerns about clinical status.
4. The patient should be made NPO immediately.
5. Provide supplemental oxygen to keep oxygen saturations greater than 95%.

6. STAT Laboratory work:*  
   a. Fingerstick glucose  
   b. CBC  
   c. Type and cross  
   d. Hb S level**  
   e. Stat Arterial Blood Gas if any signs of respiratory distress or hyperventilation  
   f. Reticulocyte count  
   g. PT/PTT/INR  
   h. BMP  
   i. Ionized Calcium  
   j. Ammonia  
     *If patient has port for chronic transfusions, please see Appendix 1  
     **Hb S will not be resulted STAT, and result is not necessary to proceed with protocol

7. Imaging, if deemed necessary by evaluating teams:
a. Hyperacute, non-sedated Brain MRI, MRA sequence (ordered as “hyperacute stroke protocol” in additional comments of order, Appendix 2) should be obtained within 3 hours of guideline initiation. It is preferred to not delay the exchange transfusion to obtain an MRI; if an urgent MRI cannot be obtained within 3 hours of guideline initiation, a Head CT should be done prior to pheresis to rule out intracranial hemorrhage before proceeding with exchange transfusion.

b. *It is preferred to obtain imaging without sedation,* but if the patient is unable to tolerate MRI scan without sedation, MRI should be coordinated by the admitting team immediately so that the MRI can be performed with sedation by Anesthesiology as soon as possible, *which may or may not be prior to exchange transfusion.* If the MRI is obtained after pheresis, then it may be scheduled during regular business hours.

i. Monday-Friday 0700-1630: MRI should be scheduled thru the APC by calling the APC charge nurse @ 314-454-8787.

ii. All other times: the sedated scan is arranged through Children’s Direct.

If the attending neurologist and hematologist feel it is imperative that a sedated MRI be performed as a medical emergency before pheresis because the findings will significantly change the acute management, the neurology attending should discuss their concerns with the attending anesthesiologist and ICU attending to weigh the risks associated with an emergent MRI under anesthesia and the potential imaging benefits in this acute clinical setting.

If the patient is being sedated prior to going to PICU, the anesthesiologist should discuss with the PICU attending whether continued or coordinated sedation may be needed in ICU for urgent line placement.

**To summarize: Preferred order of brain imaging:**

1. Unsedated MRI prior to erythrocytapheresis
2. Unsedated low radiation Head CT prior to erythrocytapheresis with a sedated MRI after erythrocytapheresis
3. Sedated MRI before erythrocytapheresis should be performed only if imaging results will significantly alter acute management

**Treatment**

If the hematology and neurology fellows and attendings agree that the patient is likely experiencing a stroke, the following is a recommended treatment plan:

1. If the patient is not already receiving chronic transfusions, an emergent exchange transfusion to reduce level of Hb S <30% and raise total Hb level, not to exceed 10 g/dL (Kirkham 2007)
2. NPO
3. Hydration: give 20 ml/kg NS bolus if indicated and then NS at maintenance. Adjust IV fluids to assure adequate hydration and minimize risk of hypotension.

4. Heme/Onc Fellow to contact the SLCH pheresis team and nephrology attending after discussed with neurology fellow (contact Dialysis Unit 454-6065 between 8:00 am-4:30pm and Pheresis on call 314-848-8477 after hours. If the team decides to not exchange transfuse the patient after the pheresis team has been contacted, it is the Heme/Onc fellow’s responsibility to notify the pheresis team of the change in plans.

5. Admit to the PICU (most patients need procedural sedation for pheresis catheter placement; consider placement of arterial line)

6. It is recommended to provide 100% oxygen during procedural sedation. Sedation drugs per ICU attending routine clinical practice.

7. PICU team to confirm with radiologist absence of hemorrhage on imaging prior to pheresis initiation.

8. PICU team to complete pheresis checklist prior to pheresis initiation.

9. If hemorrhage or midline shift is found on imaging, person receiving results (ER or PICU) should consult neurosurgery immediately.

10. Consider emergent simple transfusion of 10 mL/kg red blood cells (maximum 2 units) prior to erythrocytapheresis if:
    a. Evidence of splenic sequestration, acute chest syndrome, or aplastic crisis, or Hb < 7 g/dL for any reason
    b. If known that erythrocytapheresis cannot be initiated within 4 hours (Royal College of Physicians 2004).
    c. N.B. With above scenarios, simple transfusion should primarily be considered if Hb < 7 g/dL. A simple transfusion may result in high hemoglobin concentration (Hb > 10 g/dL) leading to high blood viscosity and increased risk of tissue ischemia, and therefore a careful risk/benefit assessment must be documented if Hb is above 7 g/dL, and effort should be made to keep Hb less than 10 g/dL post-transfusion.

Children receiving chronic blood transfusion therapy most often have <50% Hb S at any time, so the primary benefit of erythrocytapheresis (rapid lowering of Hb S from >80% to <30%) is less applicable to these patients. There is currently no evidence supporting or refuting additional erythrocytapheresis at the time of acute neurological events improving acute or long-term neurologic outcomes. Therefore, if the patient is already receiving chronic exchange transfusion, discussion amongst clinical teams should weigh risks and benefits of an additional, unscheduled manual exchange transfusion versus a simple transfusion.

Points to consider:

1. Duration of time from previous transfusion and next scheduled transfusion, as a child who has had a recent exchange transfusion is unlikely to gain substantial benefit from an additional exchange transfusion.
2. If Hgb < 8 g/dL, consider giving 15 ml/kg red blood cell simple transfusion, maximum 2 units.
**Monitoring/Management**

The goals of monitoring include detection of complications of stroke, including increased intracranial pressure, intraparenchymal hemorrhage and hypertensive or hypotensive crisis.

   (http://www.nhlbi.nih.gov/guidelines/hypertension/child_tbl.pdf)  
   a. Note: Children with sickle cell disease tend to have lower blood pressures at baseline. Therefore, a “normal” blood pressure may represent mild hypertension. Age-based parameters should still be used to avoid hypotension and maintain adequate cerebral perfusion pressure.

2. Place on continuous pulse oximeter monitor; Keep O2 saturation >95%. Notify MD of increasing O2 requirement.

3. Normal saline +/- KCL IV fluids at maintenance. Add dextrose if needed to maintain serum glucose > 80 mg/dL. Fluid resuscitation with NS to correct dehydration or hypotension

4. Maintain strict normothermia (temperature less than 38 degrees C)

5. Hourly neuro assessments

6. PT/OT/ST evaluation within 24 hours of admission

7. If *any* facial weakness, NPO until cleared by speech therapy (even if facial weakness appears to have resolved)

8. Consider starting aspirin, PO (or PR if facial weakness).
   a. \(<=15 \text{ kg give } 40.5 \text{ mg daily}
   b. \(>15 \text{ mg give } 81 \text{ mg daily}

9. Schedule full MRI as soon as possible, within less than 4 days of symptom onset; otherwise defer until 3 weeks out.

**SUBACUTE SYMPTOMS**

If symptom onset occurred greater than 72 hours before presentation to either clinic or Emergency Room, the child will still likely benefit from urgent lowering of the proportion of Hb S. In this time period, imaging may not be helpful, as certain sequences may be in a “pseudo-normalization” period as the injury transitions from acute to chronic.

*Inclusion criteria:*

1. Known hemoglobinopathy: Hb SS, Hb SC, Hb S-beta-thalassemia

2. Subacute (\(\geq 72 \text{ hours}\)) change in neurologic function (including but not limited to unilateral motor/sensory deficits, change in speech, change in mental status, seizure)
**Assessment:**

Similarly as above, both the hematology fellow and neurology fellow should be notified/consulted through a Pediatric Stroke Alert by calling Children’s Direct at 314-747-7001.

1. A complete neurologic exam should be documented with date and time, including, but not limited to:
   a. Mental status (Awake/alert, sleepy but arousable, lethargic)
   b. Speech (clear and comprehensible, dysarthric)
   c. Visual fields, pupils, facial strength and symmetry
   d. Motor function in all four limbs
   e. Sensation in all four limbs (clearly document location and extent of any abnormalities)
   f. Pediatric NIH Stroke Scale Score (to be documented by the Neurology Fellow)

2. Lab work should include:
   a. Fingerstick glucose
   b. CBC
   c. Type and cross
   d. Hb S level
   e. Arterial Blood Gas if any signs of respiratory distress or hyperventilation
   f. Reticulocyte count
   g. PT/PTT/INR
   h. BMP
   i. Ionized Calcium

3. Imaging: MRI and MRA are likely to still provide helpful information. Hematology, Neurology, and Radiology should all agree on optimal timing. The MRI may be ordered as “Sickle Cell Stroke Protocol,” and will require separate orders to add MRA.

**Treatment/Management:**

1. If systolic blood pressure is less than 5th percentile of normal range for age, administer 20 mL/kg bolus NS. Goal systolic blood pressure 50-99th percentile for age.
2. Hematology fellow should arrange for a manual exchange transfusion as an inpatient on the floor.
3. PT/OT/ST consults
4. If any facial weakness, NPO until cleared by speech therapy (even if facial weakness appears to have resolved)
5. Consider starting aspirin, PO (or PR if facial weakness).
   a. \(\leq15 \text{ kg} \) give 40.5 mg daily
   b. \(>15 \text{ mg} \) give 81 mg daily
6. Repeat neurologic examination and Pediatric NIHSS should be documented after transfusion and daily during hospital admission.
Pediatric Ischemic Stroke Inpatient Care

These guidelines are general principles and recommendations for the care of children with acute ischemic stroke. Patients with stroke and other underlying co-morbidities may require individualization and modification at discretion of clinical team.

If symptoms < 72 hours, admit to PICU. If symptoms > 72 hours and no progression of symptoms, may consider admission to 12th floor Neurology Service.

ADMISSION TO ICU (0-24 hours)
Monitoring/Orders

1. If patient received tissue plasminogen activator (tPA) please see tPA guidelines for specific post-tPA care parameters for in first 24 hours. If patient had thrombectomy, please see thrombectomy guidelines for specific post-thrombectomy care in first 24 hours.

2. Head of bed flat unless significant concern for increased ICP

3. Q1 hour vital signs: Temp, BP, HR, RR, O2 saturation. Maintain temperature <= 38° C. Acetaminophen should be ordered for prn temperature > 38° C. Consider external cooling (Arctic Sun or Cincinnati Sub-Zero) to maintain normothermia if patient intubated and able to tolerate.
   a. Blood pressure should be maintained > 5th percentile for age (see Appendix)
   b. O2 saturation should be maintained > 92%, unless patient’s baseline saturations not expected to be >92% (i.e. single ventricle physiology).

4. Intravenous fluids
   a. 0.9 % NS at 1.5 L/m². Add dextrose if needed to maintain blood glucose > 80 mg/dL. Fluid resuscitation with NS as needed for dehydration or hypotension.

5. PedNIHSS documented by nurse on admission, qshift, and with concerns for significant change in neurologic status. Neurology fellow to confirm PedNIHSS on admission and repeat with consultations.
   a. Until EPIC implementation, nurses will chart score and reason for points under “Neuro Assessment comment section”
   b. Notify MD if change in score by 2 or more points (better or worse)
   c. Neurology fellow will document score in consult/progress notes.

6. Q1 hour Neuro checks

7. Routine neuro checks should repeat abnormal sections PedNIHSS in order to closely follow neurologic deficits, but do not need full assessment unless significant clinical change.

8. PT/OT/speech consult within 24 hours of admission. (This may be delayed for up to 8 hours after tPA administration).

9. Activity level determined by neurologic deficits and fall risk, as assessed by clinical team.

10. NPO until cleared by speech therapy if ANY facial or speech deficits during acute stroke.

11. Apply sequential compression devices (SCD) to prevent deep vein thromboses. (Defer if patient received tPA).

Treatment

1. Pharmacotherapy:
a. Antiplatelet therapy for confirmed AIS, except in patients with significant hemorrhagic transformation or other significant bleeding. **Aspirin should be administered in the first 24 hours of symptom onset, unless patient received tPA. If received tPA, give aspirin 24 hours after tPA completed.**
   
   i. Aspirin dose (Note: If patient NPO, can give aspirin PR or via NG tube)
   1. <=15 kg give 40.5 mg daily
   2. > 15 kg give 81 mg daily

2. Surgical intervention:
   a. If deteriorating mental status or neurologic exam concerning for increased ICP, consult Neurosurgery and obtain STAT CT head.

**Investigation for etiology of stroke** (to be completed during the course of admission)

1. Cardiac: EKG, ECHO
   a. protein C/ protein S deficiency
   b. activated protein C resistance
   c. lipoprotein (a)
   d. plasma homocysteine
   e. factor V Leiden
   f. prothrombin G20210A mutation
   g. cardiolipin antibodies
   h. β glycoprotein antibodies.
3. Infection: If fever present, consider LP
4. Vascular imaging: MRA, consider CTA/cerebral angiogram if MRA demonstrates abnormalities or is unable to be obtained

**24- 48 HOURS AFTER PRESENTATION**

**Assessment**

1. Has etiology of stroke been identified?
2. Is patient stable for transfer to the 12th floor Neurology service?

**Labs**

At the discretion of the Primary Attending in consultation with Neurology.

**Imaging**

If urgent imaging was not performed or incomplete, schedule MRI brain and MRA (“Stroke Protocol”) within 4 days of symptoms. Discuss with Neurology and ICU teams if patient warrants CT angiogram or 4 vessel cerebral angiogram.

**Monitoring/Orders**

1. Q1 hour vital signs: Temp, BP, HR, RR, O2 saturation; Maintain temperature <=38.0. Acetaminophen should be ordered for prn temperature >38.0. Consider external cooling (Arctic Sun or Cincinnati Sub-Zero) to maintain normothermia if patient intubated and able to tolerate.
   a. Blood pressure should be maintained > 5th percentile for age (see Appendix)
   b. O2 saturation should be maintained > 92%, unless single ventricle patient
2. Q2 hour neuro checks at discretion of ICU Attending. May consider spacing from Q1 hour if symptom resolution or no new focal deficits.
3. Daily PedNIHSS to be performed by Neurology Fellow and at beginning of each nursing shift
4. Intravenous fluids
   a. NS at 1.5 L/m2 +/- KCl based on absence of renal disease and BMP. Add dextrose if needed to maintain blood glucose > 80 mg/dL, or if unable to start enteral feeds. Fluid resuscitation with NS as needed for dehydration or hypotension (i.e, blood pressure < 5th percentile for age and height (see Appendix for table).
5. Head of Bed: With Neurology or ICU team member at bedside, raise head of bed to 30 degrees and assess for symptomatic worsening
6. Activity: After Head of Bed evaluated, advance activity as tolerated to allow participation in therapy evaluation
7. NPO until cleared by speech therapy if ANY facial or speech deficits during acute stroke.

Treatment
1. Pharmacotherapy:
   a. Antiplatelet therapy for confirmed AIS, except in patients with ICH
2. Surgical intervention:
   a. If deteriorating mental status or neurologic exam concerning for increased ICP, consult Neurosurgery and arrange for STAT CT head.
3. Continue therapy services (PT/OT/speech as appropriate)

ADMISSION TO FLOOR

Assessment
1. Has etiology of stroke been identified?
2. What are rehabilitation needs?

Labs
At the discretion of the primary attending, in consultation with Neurology if Neurology not primary service.

Imaging
If urgent imaging was not performed or incomplete, schedule MRI brain and MRA (“Stroke Protocol”) within 4 days of symptoms. Discuss with Neurology and ICU teams if patient warrants CT angiogram or 4-vessel cerebral angiogram.

Monitoring
1. Q4 hour vital signs: Temp, BP, HR, RR, O2 saturation.
   a. Maintain temperature <= 38.0. Acetaminophen should be ordered for prn temperature >38.0.
   b. Blood pressure should be maintained > 5th percentile for age (see Appendix)
   c. O2 saturation should be maintained > 92%, unless single ventricle patient.
2. Daily PedNIHSS
3. Diet assessment
   a. If patient not safe for PO intake, maintain NG tube for enteral feeding.
4. Intravenous fluids if patient not ready for enteral feeding.
   a. NS at 1.5 L/m2 +/- KCl based on absence of renal disease and BMP. Add dextrose if needed to maintain blood glucose > 80 and patient unable to tolerate enteral feeds. Fluid resuscitation with NS as needed for dehydration or hypotension (i.e., blood pressure < 5th percentile for age and height (see Appendix for lookup table).

5. Q4 hour Neuro checks
6. NPO until cleared by speech therapy

**Treatment**

1. Pharmacotherapy:
   a. Continue antiplatelet therapy (aspirin) for confirmed AIS, except in patients with significant symptomatic hemorrhagic transformation
   b. Treatment targeting etiology of stroke as applicable.

2. Surgical intervention:
   a. If deteriorating mental status or neurologic exam concerning for increased ICP, consult Neurosurgery and arrange for STAT CT head.

**DISPOSITION PLANNING**

1. Consider Neurorehabilitation consult if patient with significant deficit after 24 hours and able to tolerate 3 hours of therapy daily.

2. Patients with ischemic stroke should be discharged on aspirin, unless clear contraindication

3. Follow up with Neurology in 3 months; may also participate in Neurocritical Care Follow Up clinic with assessment 1 month out from discharge.
Pediatric Stroke Admission to ICU 0-24 Hours

**Confirmed AIS**

- Antiplatelet therapy w/in 24 hrs (EXCEPT pts w/significant hemorrhage)
- Vascular imaging shows extracranial internal carotid artery dissection or ECHO w/thrombus AND stroke does not involve ≥2/3 of hemisphere

**High-suspicion for AIS**

- Neuroimaging: Hyperacute MRI of the brain if stable, or CT

**Q 1hr Neuro check**

- Stable?

**Deteriorating mental status/concern for ↑ ICP**

- Investigate etiology of stroke (during admission):
  - Cardiac: EKG, ECHO
  - Infection: If fever present, LP
  - Vascular Imaging: MRA, consider CTA/cerebral angiogram
  - HSV: IgG/IgM PCR, consider starting acyclovir
  - Hypercoagulability: protein C/protein S deficiency, activate protein C resistance, increased lipoprotein (a), increased plasma, homocysteine, factor V Leiden, prothrombin G20210A and MTHFRTT677 mutations and antiphospholipid antibodies

**Confirmed AIS**

**No**

- No Stroke

**No**

- Treat for likely diagnosis

**Evaluate other etiology**

**Goals of Care:**

- NPO until cleared by speech therapy
- HOB flat unless significant concern for ↑ ICP
- Bedrest
- Q1 hour vital signs
  - Temp ≤ 38 → acetaminophen PRN for temp > 38
  - Consider Cooling Blanket to maintain normothermia
  - BP maintained > 5th %ile for age (see Appendix)
  - O2 sats > 92%
- IVF 0.9 % NS at 1.5 L/m2
  - Add dextrose if needed to maintain blood glucose > 80
  - Fluid resuscitation w/NS as needed for dehydration or hypotension
- Q1 Neuro checks
- PT/OT/speech consult orders w/in 24 hrs of admission
- Q shift PedNIHSS to be performed by bedside nurse

**Aspirin Therapy:**

- ≤ 15 kg give 40.5 mg daily
- > 15 kg give 81 mg daily
Pediatric Stroke ICU Care 24-48 Hours

**Confirmed AIS?**
- No
  - **Consider Neuroimaging**
    - To confirm AIS
  - **MRI confirms AIS?**
    - Yes
      - Evaluate for other etiology
    - No
      - **Stable?**
        - Yes
          - Continue antiplatelet therapy (Except in pts w/significant hemorrhage) & Q 1hr neuro checks, space to Q2 if stable
        - No:
          - ↓ Mental Status, Worsening deficits/Concern for ↑ ICP
            - **Consult Neurosurgery and STAT CT head, Continue Q 1hr neuro checks**

- **HOB test: HOB to 30 degrees and assess for worsening symptoms With ICU Fellow/Attending or Neurology at bedside**

- **HOB test fail**
  - **HOB Test pass**
  - Discuss transfer to 12th floor w/Neurology

**Goals of Care:**
- NPO until cleared by speech therapy
- After HOB evaluated, advance activity as tolerated to allow participation in therapy
- Q1 hour vital signs
  - ~Temp ≤ 38 → acetaminophen PRN for temp > 38
  - ~Consider Cooling Blanket to maintain normothermia
  - ~BP maintained > 5th %ile for age (see Appendix)
  - ~O2 sats >92%
- IVF 0.9 % NS at 1.5 L/m2
  - ~Add dextrose if needed to maintain blood glucose > 80 or if unable to start enteral feeds
  - ~Fluid resuscitation w/NS as needed for dehydration or hypotension (i.e, BP < 5th %ile for age and ht, see Appendix)
- Neuro checks may be spaced to Q2 or more as appropriate
- Continue therapy services as appropriate (PT/OT/Speech)
- Q shift PedNIHSS to be performed by bedside nurse

**Aspirin Therapy:**
- ≤ 15 kg give 40.5 mg daily
- > 15 kg give 81 mg daily
References:

## Appendix 1

**PedNIHSS from Ichord et al. “Interrater Reliability of the Pediatric National Institutes of Health Stroke Scale (PedNIHSS) in a Multicenter Study.” Stroke March 2011; 42: 613-617, supplemental data**

PedNIHSS INSTRUCTIONS: Administer stroke scale items in the order listed. Follow directions provided for each exam item. Scores should reflect what the patient does, not what the clinician thinks the patient can do. **MODIFICATIONS FOR CHILDREN:** Modifications to testing instructions from the adult version for use in children are shown in bold italic with each item where appropriate. Items with no modifications should be administered and scored with children in the same manner as for adults.

<table>
<thead>
<tr>
<th>Item# and Instructions</th>
<th>Scale Definition and Scoring Guide</th>
</tr>
</thead>
</table>
| **1a. Level of Consciousness:** the investigator must choose a response, even if a full evaluation is prevented by such obstacles as an endotracheal tube, language barrier, orotracheal trauma/bandages. A 3 is scored only if the patient makes no movement (other than reflexive posturing) in response to noxious stimulation. | 0 = Alert; keenly responsive.  
1 = Not alert, but arousable by minor stimulation to obey, answer, or respond.  
2 = Not alert, requires repeated stimulation to attend, or is obtunded and requires strong or painful stimulation to make movements (not stereotyped).  
3 = Responds only with reflex motor or autonomic effects or totally unresponsive, flaccid, areflexic. |
| **1b. LOC Questions:** The patient is asked the month and his/her age. The answer must be correct - there is no partial credit for being close. Aphasics and stuporous patients who do not comprehend the questions will score 2. Patients unable to speak because of endotracheal intubation, orotracheal trauma, severe dysarthria from any cause, language barrier or any other problem not secondary to aphasia are given a 1. It is important that only the initial answer be graded and that the examiner not "help" the patient with verbal or non-verbal cues. **Modified for children, age 2 years and up. A familiar Family Member must be present for this item:** Ask the child "how old are you?" Or "How many years old are you?" for question number one. Give credit if the child states the correct age, or shows the correct number of fingers for his/her age. For the second question, ask the child "where is XX?" XX referring to the name of the parent or other familiar family member present. Use the name for that person which the child typically uses, e.g. "mommy". Give credit if the child correctly points to or gazes purposefully in the direction of the family member. | 0 = Answers both questions correctly.  
1 = Answers one question correctly.  
2 = Answers neither question correctly. |
| **1c. LOC Commands:** The patient is asked to open and close the eyes and then to grip and release the non-paraetic hand. **For children one may substitute the command to grip the hand with the command "show me your nose" or "touch your nose".** Substitute another one step command if the hands cannot be used. Credit is given if an unequivocal attempt is made but not completed due to weakness. If the patient does not respond to command, the task should be demonstrated to them (pantomime) and score the result (i.e., follows none, one or two commands). Patients with trauma, amputation, or other physical impediments should be given suitable one-step commands. Only the first attempt is scored. | 0 = Performs both tasks correctly  
1 = Performs one task correctly  
2 = Performs neither task correctly |
| **2. Best Gaze:** Only horizontal eye movements will be tested. Voluntary or reflexive (oculocephalic) eye movements will be scored but caloric testing is not done. If the patient has a conjugate deviation of the eyes that can be overcome by voluntary or reflexive activity, the score will be 1. If a patient has an isolated peripheral nerve paresis (CN III, IV or VI) score a 1. Gaze is testable in all aphasic patients. Patients with ocular trauma, bandages, pre-existing blindness or other disorder of visual acuity or fields should be tested with reflexive movements and a choice made by the investigator. **Establishing eye contact and then moving about the patient from side to side will occasionally clarify the presence of a partial gaze palsy.** | 0 = Normal  
1 = Partial gaze palsy. This score is given when gaze is abnormal in one or both eyes, but where forced deviation or total gaze paresis are not present.  
2 = Forced deviation, or total gaze paresis not overcome by the oculocephalic maneuver. |
| **3. Visual:** Visual fields (upper and lower quadrants) are tested by confrontation, using finger counting (for children > 6 years) or visual field (for children age 2 to 6 years) as appropriate. Patient must be encouraged, but if they look at the side of the moving fingers appropriately, this can be scored as normal. If there is unilateral blindness or enucleation, visual fields in the remaining eye are scored. Score 1 only if a clear-cut asymmetry, including quadrantanopia is found. If patient is blind from any cause score 3. Double simultaneous stimulation is performed at this point. If there is extinction patient receives a 1 and the results are used to answer question 11. | 0 = No visual loss  
1 = Partial hemianopia  
2 = Complete hemianopia  
3 = Bilateral hemianopia (blind including cortical blindness) |
<table>
<thead>
<tr>
<th>4. Facial Palsy: Ask, or use pantomime to encourage the patient to show teeth or raise eyebrows and close eyes. Score symmetry of grimace in response to noxious stimuli in the poorly responsive or non-comprehending patient. If facial trauma, bandages, orotracheal tube, tape or other physical barrier obscures the face, these should be removed to the extent possible.</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 = Normal symmetrical movement</td>
</tr>
<tr>
<td>1 = Minor paralysis (flattened nasolabial fold, asymmetry on smiling)</td>
</tr>
<tr>
<td>2 = Partial paralysis (total or near total paralysis of lower face)</td>
</tr>
<tr>
<td>3 = Complete paralysis of one or both sides (absence of facial movement in the upper and lower face)</td>
</tr>
</tbody>
</table>

| 5 & 6. Motor Arm and Leg: The limb is placed in the appropriate position: extend the arms (palms down) 90 degrees (if sitting) or 45 degrees (if supine) and the leg 30 degrees (always tested supine). Drift is scored if the arm falls before 10 seconds or the leg before 5 seconds. For children too immature to follow precise directions or uncooperative for any reason, power in each limb should be graded by observation of spontaneous or elicited movement according to the same grading scheme, excluding the time limits. The aphasic patient is encouraged using urgency in the voice and pantomime but not noxious stimulation. Each limb is tested in turn, beginning with the non-paretic arm. Only in the case of amputation or joint fusion at the shoulder or hip, or immobilization by an IV board, may the score be "9" and the examiner must clearly write the explanation for scoring as a "9." Score each limb separately. |
| 5a. Left Arm |
| 5b. Right Arm |
| 0 = No drift, limb holds 90 (or 45) degrees for full 10 seconds. |
| 1 = Drift, Limb holds 90 (or 45) degrees, but drifts down before full 10 seconds; does not hit bed or other support. |
| 2 = Some effort against gravity, limb cannot get to or maintain (if cued) 90 (or 45) degrees, drifts down to bed, but has some effort against gravity. |
| 3 = No effort against gravity, limb falls. |
| 4 = No movement |
| 9 = Amputation, joint fusion explain: |

| 6a. Left Leg |
| 6b. Right Leg |
| 0 = No drift, leg holds 30 degrees position for full 5 seconds. |
| 1 = Drift, leg falls by the end of the 5 second period but does not hit bed. |
| 2 = Some effort against gravity, leg falls to bed by 5 seconds, but has some effort against gravity. |
| 3 = No effort against gravity, leg falls to bed immediately. |
| 4 = No movement |
| 9 = Amputation, joint fusion explain: |

| 7. Limb Ataxia: This item is aimed at finding evidence of a unilateral cerebellar lesion. Test with eyes open. In case of visual defect, insure testing is done in intact visual field. The finger-nose and heel-knee tests are performed on both sides, and ataxia is scored only if present out of proportion to weakness. In children, substitute this task with reaching for a toy for the upper extremity, and kicking a toy or the examiner's hand, in children too young (< 5 years) or otherwise uncooperative for the standard exam item. Ataxia is absent in the patient who cannot understand or is paralyzed. Only in the case of amputation or joint fusion may the item be scored "9," and the examiner must clearly write the explanation for not scoring. In case of blindness test by touching nose from extended arm position. |
| 0 = Absent |
| 1 = Present in one limb |
| 2 = Present in two limbs |

| 8. Sensory: Sensation or grimace to pin prick when tested, or withdrawal from noxious stimulus in the obtunded or aphasic patient. For children too young or otherwise uncooperative for reporting gradations of sensory loss, observe for any behavioral response to pin prick, and score it according to the same scoring scheme as a "normal" response, "mildly diminished" or "severely diminished" response. Only sensory loss attributed to stroke is scored as abnormal and the examiner should test as many body areas (arms not hands), legs, trunk, face) as needed to accurately check for hemisensory loss. A score of 2, "severe or total," should only be given when a severe or total loss of sensation can be clearly demonstrated. Bilateral and aphasic patients will therefore probably score 1 or 0. |
| 0 = Normal; no sensory loss. |
| 1 = Mild to moderate sensory loss; patient feels pinprick is less sharp or is dull on the affected side; or there is a loss of superficial pain with pinprick but patient is aware he/she is being touched. |
| 2 = Severe to total sensory loss; patient is not aware of being touched in the face, arm, and leg. |
9. **Best Language:** A great deal of information about comprehension will be obtained during the preceding sections of the examination. For children age 6 years and up with normal language development before onset of stroke: The patient is asked to describe what is happening in the attached picture, to name the items on the attached naming sheet, to repeat words from the attached list, and to read from the attached list of sentences (Table 51, Fig 51, 52, 53). Comprehension is judged from responses here as well as to all of the commands in the preceding general neurological exam. If visual loss interferes with the tests, ask the patient to identify objects placed in the hand, repeat, and produce speech. The intubated patient should be asked to write. The patient in coma (question 1a-3) will arbitrarily score 3 on this item. The examiner must choose a score in the patient with stupor or limited cooperation but a score of 3 should be used only if the patient is mute and follows no one step commands. For children age 2 yrs to 6 yrs (or older children with premorbid language skills < 6 yr level), score this item based on observations of language comprehension and speech during the examination.

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No aphasia, normal</td>
</tr>
<tr>
<td>1</td>
<td>Mild to moderate aphasia; some obvious loss of fluency or facility of comprehension, without significant limitation on ideas expressed or form of expression. Reduction of speech and/or comprehension, however, makes conversation about provided material difficult or impossible. For example in conversation about provided materials examiner can identify picture or naming card from patient’s response.</td>
</tr>
<tr>
<td>2</td>
<td>Severe aphasia; all communication is through fragmentary expression; great need for inference, questioning, and guessing by the listener. Range of information that can be exchanged is limited; listener carries burden of communication. Examiner cannot identify materials provided from patient response.</td>
</tr>
<tr>
<td>3</td>
<td>Mute, global aphasia; no usable speech or auditory</td>
</tr>
</tbody>
</table>

The patient with brain stem stroke who has bilateral loss of sensation is scored 2. If the patient does not respond and is quadriplegic score 2. Patients in coma (item 1a-3) are arbitrarily given a 2 on this item.
### 10. Dysarthria
If patient is thought to be normal an adequate sample of speech must be obtained by asking patient to read or repeat words from the attached list. If the patient has severe aphasia, the clarity of articulation of spontaneous speech can be rated. Only if the patient is intubated or has other physical barriers to producing speech, may the item be scored "9", and the examiner must clearly write an explanation for not scoring. Do not tell the patient why he/she is being tested.

<table>
<thead>
<tr>
<th>0</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Mild to moderate; patient slurs at least some words and, at worst, can be understood with some difficulty.</td>
</tr>
<tr>
<td>2</td>
<td>Severe; patient’s speech is so slurred as to be unintelligible in the absence of or out of proportion to any dysphasia, or is muttering.</td>
</tr>
<tr>
<td>9</td>
<td>Intubated or other physical barrier, explain:</td>
</tr>
</tbody>
</table>

### 11. Extinction and Inattention (formerly Neglect)
Sufficient information to identify neglect may be obtained during the prior testing. If the patient has a severe visual loss preventing visual double simultaneous stimulation, and the cutaneous stimuli are normal, the score is normal. If the patient has aphasia but does appear to attend to both sides, the score is normal. The presence of visual spatial neglect or anosagnosia may also be taken as evidence of abnormality. Since the abnormality is scored only if present, the item is never unintestable.

<table>
<thead>
<tr>
<th>0</th>
<th>No abnormality.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Visual, tactile, auditory, spatial, or personal inattention or extinction to bilateral simultaneous stimulation in one of the sensory modalities.</td>
</tr>
<tr>
<td>2</td>
<td>Profound hemi-inattention or hemi-inattention to more than one modality. Does not recognize own hand or orients to only one side of space.</td>
</tr>
</tbody>
</table>
Table S1. Language testing items for PedNIHSS:

<table>
<thead>
<tr>
<th>Repetition</th>
<th>Each of 4 word-repetition tasks is presented:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>a. Stop</td>
</tr>
<tr>
<td></td>
<td>b. Stop and go</td>
</tr>
<tr>
<td></td>
<td>c. If it rains we play inside</td>
</tr>
<tr>
<td></td>
<td>d. The President lives in Washington</td>
</tr>
<tr>
<td>Reading</td>
<td>Each of 3 items is presented for the child to read in Fig 1. Adjust expectations according to child’s age/school level</td>
</tr>
<tr>
<td>Naming</td>
<td>Pictures are presented and of a clock, pencil, skateboard, shirt, baseball, bicycle (Fig 2).</td>
</tr>
<tr>
<td>Fluency and word finding</td>
<td>The picture (Fig 3) is presented and the child is asked to describe what he/she sees.</td>
</tr>
</tbody>
</table>

Stop

See the dog run

Little children like to play outdoors

Fig S1. Reading items for PedNIHSS
Fig. S2 Pictures to test naming for item 9 Best Language of PedNIHSS
Fig. S3 Picture to test story-telling for Item 9 Best Language of PedNIHSS
Table which may be used to track scoring, if helpful

<table>
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Appendix 2. Pediatric Blood Pressure Charts

National Institutes of Health – National Heart, Lung and Blood Institute (NIH-NHLBI)
From the Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents
## Blood Pressure Levels for Boys by Age and Height Percentile

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BP, blood pressure

* The 90th percentile is 1.28 SD, 50th percentile is 1.645 SD, and the 99th percentile is 2.326 SD over the mean.

For research purposes, the standard deviations in Appendix Table B-1 allow one to compute BP Z-scores and percentiles for boys with height percentiles given in Table 3 (i.e., the 5th, 10th, 25th, 50th, 75th, 90th, and 99th percentiles). These height percentiles must be converted to height Z-scores given by (5% = -1.645; 10% = -1.28; 25% = -0.68; 50% = 0; 75% = 0.68; 90% = 1.28; 95% = 1.645) and then computed according to the methodology in steps 2-4 described in Appendix B. For children with height percentiles other than these, follow steps 1-4 as described in Appendix B.
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BP, blood pressure

* The 90th percentile is 1.23 SD, 95th percentile is 1.645 SD, and the 99th percentile is 2.326 SD over the mean.

For research purposes, the standard deviations in Appendix Table B-1 allow one to compute BP Z-scores and percentiles for girls with height percentiles given in Table 4 (i.e., the 5th, 10th, 25th, 50th, 75th, 90th, and 95th percentiles). These height percentiles must be converted to height Z-scores given by (5% = -1.645; 10% = -1.26; 25% = -0.68; 50% = 0; 75% = 0.68; 90% = 1.28; 95% = 1.645) and then computed according to the methodology in steps 2-4 described in Appendix B. For children with height percentiles other than these, follow steps 1-4 as described in Appendix B.
Appendix 3: Hyperacute Stroke MRI Protocol

1. Localizer (0:15)
2. Axial DWI/ADC Trace (3 directions, 2 averages) (1:30)
3. Axial FLAIR (4:30)
4. Axial T1W (2:30)
5. Axial T2-star gradient echo(4:30)
6. 3D Time-of-flight MRA of Circle of Willis (5:30)

(20 minutes of time on scanner)
Appendix 4: Modified Rankin Scale

0 - No symptoms.
1 - No significant disability. Able to carry out all usual activities, despite some symptoms.
2 - Slight disability. Able to look after own affairs as expected for age without assistance, but unable to carry out all age-appropriate activities.
3 - Moderate disability. Requires significantly more help than peers, but able to walk unassisted.
4 - Moderately severe disability. Unable to attend to own bodily needs without assistance, and unable to walk unassisted.
5 - Severe disability. Requires constant nursing care and attention, bedridden, incontinent.
6 - Dead.
Imaging confirms LVO

Neuro fellow discusses w NIR fellow/attg; confirms thrombectomy candidacy

NIR fellow calls CD to activate “Thrombectomy GO” alert

CD sends alert w name, DOB, current location

- Neuro IR fellow/attending
- Ped Anesth 1st call attending
- Ped Anesth 3rd call attending
- Ped Anesth in-house res/fellow
- OR nurse
- BJH Trauma Anesthesia Attending
- ICU attendings
- ICU fellows & charge nurses
- Ped Neuro Stroke Attg & fellow

- Prepare for case; aware page sent
- Head to NIR to prepare for case
- Backup 1st call anesthesia attending
- Go to pt; assess and consent for anesthesia
- Book off-site case
- Aware, assist w anesth resources
- Go to pt location to transport to NIR
- Aware of possible admission
- Notify providers & BJH; escort family
Take the Level 2 link south to the Mallinckrodt elevators (or West elevators if need more space). Neuro-IR suite is on Level 3 of MIR. Look for small black sign: Radiology/Neuroradiology/Musculoskeletal.

In case of emergency, call 314.362.5000
Appendix 7. Guidance for children with pre-existing double port-a-cath (typically for patients with sickle cell disease receiving chronic exchange transfusions)

A. Labs should be drawn in ER or clinic through pre-existing port-a-cath, and then patient may be de-accessed.
   a. An additional IV access site should be obtained for hydration and medications.
   b. LMX placed on port-a-cath site.

B. Upon arrival to PICU, Alteplase (tPA) and LMX should be ordered to bedside.
   a. Alteplase 2mg diluted into 5mls into EACH lumen of portacath (2 lumens). Pheresis nurse will administer.
   b. LMX-4: Verify LMX is in place. (Pheresis requires special needles. Standard Huber needles are not adequate. Pheresis will access immediately prior to Pheresis procedure.)

C. Note that children who already are receiving transfusions as part of their sickle cell management will very likely require more to time to find appropriately matched blood.
   a. Hem/Onc Fellow should get estimate of how many units will be needed from pheresis team and notify blood bank.
The main goals of early management are:

1) Monitor the neurological status of “at risk” infants closely (at least hourly) in the first hours after birth using the neonatal encephalopathy assessment tool (NEAT).

2) Avoid hyperthermia (core temperature > 37.5 °C) during this observational period.

3) Initiate hypothermia in eligible infants in the NICU as soon as meets eligibility criteria, between 60 min and 6 hours of life.

4) Ensure early communication with the delivering Obstetrician via Children’s Direct once the decision is made to initiate hypothermia.

5) Communication with the family.

1. Eligibility criteria to be considered for cooling – must fulfill all three criteria (A+B+C):

   A. Infants ≥ 35 weeks gestational age

   B. Any one of the following perinatal factors:
      1) Low Apgar scores \( \rightarrow \) \( \leq 5 \) at 10 minutes of life
      2) Prolonged resuscitation at birth \( \rightarrow \) chest compressions and/or intubation or mask ventilation at 10 minutes
      3) Severe acidosis \( \rightarrow \) pH \( \leq 7.1 \) from cord or patient arterial blood gas within 60 minutes of birth
      4) Base deficit \( \geq 12 \) mmol/L from cord or patient arterial blood gas within 60 minutes of birth

   C. Any one of the following:
      1) Seizure or any clinical event concerning for seizure
      2) Moderate-Severe Neonatal encephalopathy (in at least 3 of the 6 clinical criteria) on the Neonatal Encephalopathy Assessment Tool (NEAT)

2. Identification of Infants at BJH (Parkview Tower):

   A. Initial evaluation by the fellow/attending in the delivery room can inform post-delivery room placement of the infant. Infants can be monitored in the delivery room on the warming table with the thermostat at 36.5°C until stabilized (up to 60 minutes, through the first exam).

   B. A neonatal encephalopathy examination is to be completed and documented by the Neonatology team at 60 minutes of life if the patient satisfies the first two eligibility criteria (gestational age and perinatal factor) using the NEAT tool.

   C. Once the baby is admitted to the NICU (or Neonatal Assessment Center (NAC) under select circumstances*), a concurrent encephalopathy evaluation is to be conducted by two providers (fellow/attending/NPP level) from the Neonatology and Pediatric Neurology teams.

   D. If patient has mild or rapidly improving moderate neonatal encephalopathy on initial exam in
the NICU, repeat examination with documentation on the NEAT tool at least hourly as indicated; continue to repeat examinations based upon results and course until 6 hours of age or a decision to initiate therapeutic hypothermia has been made.

E. Eligible infants who meet cooling criteria (moderate-severe encephalopathy in 3/6 features on the NEAT at 60 minutes) should be transported to the NICU for cooling following activation of the encephalopathy/hypothermia protocol via Children's Direct (CD).

3. Exclusion Criteria for Therapeutic Hypothermia (consistent with existing evidence):

1) Gestational age less than 35 weeks or greater than 6 hours of life (see exceptions under Special Considerations section below)
2) Severe congenital anomalies/syndromes/known metabolic disorders
3) ECMO (at discretion of attending physician)
4) Severe PPHN (at discretion of attending physician)
5) Active bleeding (at discretion of attending physician)
6) Severe hemodynamic compromise/perfusion sensitive states (e.g., sepsis)
7) IUGR (birth weight < 1800 g)

**Staffing capacity for serial neurological evaluation, rapid availability of Pediatric Neurology team members and the ability to perform a stat EEG makes the NICU the ideal location to manage infants with encephalopathy. However, infants who are mildly encephalopathic or moderately encephalopathic initially but improving rapidly in the delivery room may be admitted to the NAC for close observation under the following conditions:

1) The infant can be observed on a warming table with thermostat at 36.5°C with a target axillary temperature of 36.5°C documented every 30 minutes
2) If the encephalopathy does not improve or worsens by the second hourly examination, the infant should be transferred to the NICU for further management and timely initiation of therapeutic hypothermia.
3) Infants can be observed in the NAC for up to 3 hours. If there are still concerns about potential for meeting cooling criteria, the infant should be transferred to the NICU for either continued close observation and monitoring or cooling, if the decision has been made.
Management of In-Born (BJH-PVT) Infants Eligible for Therapeutic Hypothermia (i.e., those with moderate-severe encephalopathy)

1. Therapeutic hypothermia to be initiated as soon as the patient meets eligibility criteria (also see Special Consideration section).

2. Activate the Neonatal Encephalopathy Team upon admission/arrival to the NICU (through call to Children's Direct).

3. Procedure to be followed after admission to the NICU:
   
   A. Place infant on the Curewrap/Criticool system and follow guidelines for connecting and starting the device (flip chart attached to each machine).
   
   B. Connect both surface (in exposed area) and rectal temperature probes into the Criticool device.
   
   C. Scenarios for setting target temperature (always keep bubble wrap pillow between infant’s head and wrap)
      1) If infant has core temperature of 31-34°C, set target temperature to 33.5°C.
      2) If core temperature is <31°C, set target temperature at 1°C above actual core temperature. Target temperature should be reached in ~30 min. Keep increasing target temperature in increments of ~1°C every 30 minutes until core temperature is >32°C, and then set target temperature to 33.5°C.
      3) The blanket remains warm to touch and this is normal as it tries to maintain the infant’s temperature.

4. Medical Management Guidelines by Systems:
   
   A. Fluid and Electrolytes
      1) NPO for the first 48 hours. Consider trophic feeds (20 ml/kg/d) during cooling once evidence of return of bowel function (as defined by presence of clinical improvement and normal bowel sounds).
      2) Start IV maintenance fluids: 50-60 ml/kg/day of D15W (or D10W if hyperglycemic).
      3) Consider TPN on DOL# 2 with 3 gm/kg/d protein and 1gm/kg/d Intralipid if electrolytes and urine output are stable and severity of encephalopathy predicts poor feeding tolerance after therapeutic hypothermia (restrictive criteria are due to ongoing drug shortages). TPN duration determined by enteral feeding tolerance at 100ml/kg/d.
      4) Monitor and maintain electrolytes within normal limits
         • Calcium (ionized or total calcium)
         • Potassium
         • Glucose
         • Magnesium
      5) Management of acidosis – avoid base replacement therapy if circulation is re-established and patient can self-correct over time
      6) Treat hypovolemia with volume administration as needed
         • Normal Saline – 10 ml/kg IV
         • Packed Red Blood Cells (± plasma) – if blood loss is etiology
      7) If worsening acidosis or cardiovascular collapse with BE ≥ -10 consider:
• Normal Saline – 10 ml/kg IV
• Sodium Bicarbonate – 1 mEq/kg over 30 minutes

8) Access: Place a UVC +/- UAC for access

B. Respiratory

1) Ventilator Support – provide any respiratory support as needed
   • Avoid hypocapnea (<40 mm Hg)
     o Cooling can ↓ pCO2
     o Maintain blood gas pCO2 goal: 45-50 mmHg
   • Avoid hyperoxia
     o Maintain Oxygen saturations: 90-98%.
     o PaO2 should be < 100mmHg
2) Maintain air humidifier in normothermic range (37°C)

C. Cardiovascular

1) Blood Pressure Management – continuous arterial line monitoring preferred
   • Maintain blood pressure in normal range, despite bradycardia
   • Treat hypovolemia with volume administration as needed
   • Support blood pressure with fluids or inotropes only if indicated
     o Normal Saline – 10 ml/kg IV fluid bolus
     o Continuous IV inotrope infusions:
       • Dopamine (1st choice agent)
       • Dobutamine (2nd choice agent)
2) Heart Rate
   • Expect bradycardia (<100bpm) when temperature < 34°C
   • Monitor with 3-lead EKG per routine
   • For deep bradycardia (< 80bpm)
     o May be tolerated, if blood pressure is maintained adequately
     o Raising rectal temperature to 34°C alone may be adequate
   • Monitor for arrhythmias and consider an EKG and checking electrolytes, calcium and magnesium

D. Infectious Disease

1) The majority of neonates with encephalopathy do not have an infectious etiology. Review need to start antibiotics based on perinatal risk factors.
   If a decision is made to start antibiotics, draw blood cultures and perform a lumbar puncture prior to starting antibiotics (if clinical condition hemodynamically stable)
   • Ampicillin: 200 mg/kg/day IV divided q12 hours
   • Gentamicin: 5 mg/kg/dose IV q24 hours
   OR
   • Cefotaxime: 50 mg/kg/dose IV q8 hours only if high suspicion of meningitis
2) Duration of antibiotics to be based on risk factors/labs concerning for sepsis/clinical status

E. Neurological

1) Neurology Encephalopathy Rapid Response Team activated (via Children’s
Direct

2) Sedation: maintain adequate sedation with morphine (Load with 50 mcg/kg followed by 10 mcg/kg/hour, reduced to 5 mcg/kg/hour after 12 hours to avoid accumulation and toxicity) or dexmedetomidine*** (0.2mcg/kg/hour for 1 hour, then 0.3 mcg/kg/hour for 3 hours and then, if needed, 0.4 mcg/kg/hour) (goal: no shivering, irritability, facial grimacing or tachycardia attributable to sedation needs [HR > 120 bpm])

3) Seizure control
   - Obtain clinical one hour EEG on admission (to be ordered STAT by Pediatric Neurology resident)
   - Continue video EEG recording for minimum of 24 hours on all encephalopathic cooled infants
   - Treat seizures – clinical or per Neurology recommendations based upon EEG results (i.e., subclinical seizures)
   - Phenobarbital (1st choice agent)
     - Load: 20 mg/kg IV; repeat if seizures persist 20 minutes after load complete
     - Consider checking serum levels 2-12 hours after load based upon course
   - If 2nd agent required: Fosphenytoin 20 mg/kg load
   - If 3rd agent required: Midazolam – load with 0.05 mg/kg IV and then infusion of 0.15 mg/kg/hour for 12 hours, taper over another 12-24 hours

4) MR imaging
   - If severely encephalopathic with severe EEG depression and signs of brainstem injury, consider a MRI at 24-48 hours (if withdrawal of support is being considered)
   - Routine MRI with MRS – on DOL #4-5 (after rewarming)
   - Follow-up MRI on/after DOL #10 (no MRS)

5) Complete and document a neurological exam once daily during hypothermia, rewarming and at discharge.

*** Dosing adapted from guideline used at Seattle Children’s Hospital

5. Rewarming

   A. Rewarming the infant begins after 72 hours of cooling and is completed over a 24 hour period
   B. Initial rewarming is accomplished by increasing core temperature setting by 0.2°C every 2 hours for 12 hours and then by 0.3°C every 2 hours for the next 12 hours
   C. Once the infant’s core temperature reaches 36.5°C, the Curewrap is removed and the Criticool machine is turned off
   D. The rectal probe may be removed at this time
   E. Monitor neurological status closely (every 2-4 hours) during rewarming
   F. Consider obtaining a 1-hour EEG during the rewarming period if the infant has had seizures during hypothermia, if the infant remains encephalopathic or if the infant experiences a significant deterioration in neurological status
   G. Early Re-warming: Once a decision is made to cool a baby based on moderate-severe encephalopathy staging at any of our system NICUs (MBMC/PW/BMH/Shiloh) by a faculty neonatologist, the baby should be actively or passively cooled during transport and cooling continued for a full 72 hours. Premature cessation of cooling therapy in babies even with mild neonatal encephalopathy does not exclude residual brain injury and adverse long-term neurodevelopmental outcomes.
Special Considerations:

1. **Management of mildly encephalopathic infants** in the delivery room at BJH-PVT or infants who meet the first 2 criteria after delivery:

   A. Mild encephalopathy is manifest by a hyperalert state, jitteriness and irritability, with mild distal flexion and a weak/intermittently absent suck.

   B. These infants should be kept 36.5°C during transit to the NICU and while in the NAC. Core temperature can be maintained using blankets with a radiant warmer with the thermostat (skin probe) set at 36.5°C

   C. Encephalopathy stage should be documented using the neonatal encephalopathy assessment tool (NEAT) jointly by two medical care providers (ideally fellow/attending from Neonatology and Pediatric Neurology) hourly until the decision is made to either cool the infant or the infant reaches 6 hours of life, whichever comes first.

2. **Moderate-Severe encephalopathy between 6-24 hours**

   Recent research into delayed cooling shows that cooling between 6-24 hours of life may be beneficial, but there is uncertainty in its effectiveness (Laptook et al. 2017. JAMA 318(16):1550-1560). Based upon these data, we recommend that the eligibility window be extended to 24 hours of life unless the infant demonstrates seizures (after which cooling does not offer benefit).

3. **Cord gas alert:** When a fellow gets a call from lab regarding an abnormal cord gas, the initial neonatal encephalopathy exam can be conducted in the mother’s room.

4. **Therapeutic Hypothermia and ECMO:**

   Infants with severe HIE often meet ECMO support criteria secondary to pulmonary hypertension with or without meconium aspiration and/or hypoxic respiratory failure. Recent literature has examined the practice of continuing hypothermia therapy while undergoing ECMO with good results. One practice management that may be initiated as ECMO becomes likely is nitric oxide (NO). The use of NO, even at low dosing (5 ppm), can decrease the use of ECMO by as much as 35% when the oxygen index is > 25. However, despite the use of NO, ECMO is often still the required therapy in this population. A recent survey to the directors of ECMO programs showed that there is significant variability in how neonates are selected for ECMO once severity of illness criteria has been met. In this survey, 48% of the responders stated they would not offer ECMO to infant’s with severe HIE.

   Recommendations:

   1. Hypothermia may be continued safely during ECMO.
   2. Early discussion of the ECMO criteria for infants with severe HIE should be established. The discussion should include multiple disciplines (neonatology, pediatric surgery, pediatric neurology and the family).

   Ref:


Documentation

1) All encephalopathic infants admitted for observation to the NAC or for observation or hypothermia to NICU should have an admit note clearly indicating the screening criteria met and the encephalopathy stage at admission and indication for hypothermia, if initiated, using language from the NEAT tool.

2) Carefully review all the information from the obstetrical and nursing records in order to ensure accuracy of information in the admit note.

3) If there any deviations from the guideline e.g. early rewarming, the details about why that decision was made and the discussions with sub-specialists and the family should be documented in detail.

Summary Statement

1) For all infants who clearly meet criteria for moderate or severe encephalopathy after birth and a decision has been made to initiate hypothermia, the infant should be rapidly transferred to the NICU for active cooling. During transit, the thermostat (skin probe) should be set at 36.5°C (lower limit of normal).

2) Hyperthermia (core temperature >37.5 °C) is deleterious to all infants who have hypoxic-ischemic encephalopathy and should be rigorously avoided.

3) Mildly encephalopathic infants in the delivery room should be transferred to the NICU (or NAC in select circumstances) for further monitoring. Their encephalopathy stage should be documented hourly until a decision is made to cool the infant or until the infant reaches 6 hours of age. During this time, the thermostat (skin probe) should be set at 36.5°C.

4) Infants who require significant resuscitation in the delivery room but seem to recover rapidly can be observed in the NICU (or NAC in select circumstances) for further monitoring. They should be transferred to the NICU for continued evaluation and possible cooling.

5) In-house delivery guidelines do not address passive cooling due to the inability to monitor core rectal temperatures at BJH at this time and the short transit time to the NICU.
Neonatal Encephalopathy Assessment Tool (NEAT) Definitions

- **Hyperalert** – Full wakefulness with eyes open/staring but decreased frequency of blinking/tracking. Spontaneous motor activity normal or decreased with lowered threshold to all stimulus types.
- **Irritability** – Lowered threshold with excessive response to all stimulus types. Can be seen with varied states including hyperalert, lethargy, and obtundation.
- **Lethargy** – Slightly delayed but complete response to stimuli with slightly increased threshold for eliciting responses and decreased spontaneous movements.
- **Obtundation** – Delayed and incomplete responses with markedly increased threshold to all sensory stimuli and little or no motor activity.
- **Stupor** – No spontaneous eye opening and tactile stimulation elicits poorly sustained eye opening. Responds only to strong, noxious stimuli. Absent gag, corneal reflex.
- **Coma** – No eye opening with vigorous tactile stimulation.
- **Decreased spontaneous activity** – Decreased frequency or amplitude of spontaneous facial and extremity movements.
- **Absent spontaneous activity** – Movements absent in frequency and amplitude.
- **Hypotonia** – Focal or generalized decreased resistance to passive movement. Associated with greater extension of the extremities than normal.
- **Flaccid** – “Flat on the mat” appearance. May be associated with frog-leg posturing with arm and hips/legs lying in abduction.
- **Distal flexion** – Fingers, toes in strong flexion; incomplete extension of fingers when stroked on dorsal surfaces. Thumbs flexed, adducted, opposed across palms (i.e. “cortical thumbs”).
- **Decerebrate posturing** – Head, neck, and back are arched in extension (opisthotonos), elbows are extended, wrists are pronated, and hips are abducted.
- **Weak suck** – Some sucking noted, but it is not as vigorous or sustained as it should be. The pacifier can be easily pulled from the mouth.
- **Absent suck** – No sucking or root reflex elicited.
- **Incomplete Moro** – The Moro reflex is elicited by holding the baby’s head and shoulders off the mat with arms held in flexion on chest. The examiner suddenly lets the head and shoulders drop a few inches while releasing the arms. The arms should fully abduct and extend, then return towards the midline with the hand open and the thumb and the index finger forming a “C” shape. An incomplete Moro is marked by absence of any component or any asymmetry in movements. Incomplete Moro reflex often extends irregularly but typically does not return to midline.
- **Absent Moro** – Absence of any reflexive activity (see above for method of eliciting Moro reflex).
- **Dilated pupils (Mydriasis)** – Normal pupil size for term newborns are 3.9 mm +/- 0.8 mm. Dilated pupils are larger than this even in bright light.
- **Constricted pupils (Miosis)** – Normal pupil size for term newborns are 3.9 mm +/- 0.8 mm. Constricted or pinprick pupils are smaller than this even in dim light.
- **Unequal; Fixed; Dilated; Poor light reflex pupils** – Pupils that are not normally symmetrically aligned or symmetrically dilated, are fixed in position, or that do not accommodate (constrict) in the presence of light.
- **Tachycardia** – Resting heart rate of 160-180 beats per minute. Only occasional decreases to 120 beats per minute are noted.
• **Bradycardia** – Resting heart rate of 80-90 beats per minute. Only occasional increases to 120 beats per minute are noted.

• **Variable HR** – Resting heart rate varies considerably without a consistent baseline.

• **Periodic breathing** – Three or more respiratory pauses of three seconds or longer separated by normal breathing for less than 20 seconds. Often associated with shallow breathing.

• **Apnea** – Absence of airflow and respiratory effort lasting 20 seconds or longer. Apnea may also be present if a respiratory pause is shorter than 20 seconds but associated with heart-rate change or oxygen desaturation.
A. Infant ≥ 35 weeks
B. Perinatal Eligibility (any one)
   1. pH ≤ 7.1
   2. BE ≥ -12
   3. Respiratory support at 10 min
   4. Apgar score ≤ 5 at 5

Mild/Improving
Moderate - may go to NAC for observation

Target temperature 36.5°C
Avoid hyperthermia >37.5°C

Activate Encephalopathy Team while under observation in NAC

Infant needs (at least) hourly encephalopathy staging (NEAT)
Target axillary temp 36.5°C

Transfer to NICU directly for cooling by 3 hours if encephalopathy not improving

Moderate-severe infants go directly to the NICU if not improving in the DR

Target temperature 36.5°C
Avoid hyperthermia >37.5°C

Activate Encephalopathy Team once in the NICU

Admit using TH order set Initiate Hypothermia
Midazolam infusion titration for status epilepticus

Initial bolus 0.1 mg/kg (max bolus 10 mg)
Start infusion at 0.2 mg/kg/hr

Seizure continues within 10 minutes

Seizure stops

Maintain current infusion

Bolus 0.2 mg/kg (max bolus 10 mg)

Seizure continues within 10 minutes

Seizure stops

Maintain current infusion

Bolus 0.2 mg/kg (max bolus 10 mg)
Increase infusion by 0.2 mg/kg/hr

Seizure continues within 10 minutes

Seizure stops

Maintain current infusion

No

Is infusion up to 1.6 mg/kg/hr?

Yes

Consider adding alternative agent

Clinical endpoint of seizure control or burst suppression to be determined by care team.
Pentobarbital infusion titration for status epilepticus

Initial bolus 3-5 mg/kg (over 30 min)
Start infusion at 1 mg/kg/hr

Seizure continues after bolus completed
Seizure stops
Maintain current infusion

Bolus 3-5 mg/kg (over 30 min)

Seizure continues after bolus completed
Seizure stops
Maintain current infusion

Bolus 3-5 mg/kg (over 30 min)
Increase infusion by 1 mg/kg/hr

Seizure continues after bolus completed
Seizure stops
Maintain current infusion

Clinical endpoint of seizure control or burst suppression to be determined by care team.
<table>
<thead>
<tr>
<th>Benzodiazepine Name</th>
<th>Time to peak (onset)</th>
<th>Duration of action (t¹/₂)</th>
<th>Major Active Metabolite?</th>
<th>Approximately Equivalent Oral Dosages (mg)</th>
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<tbody>
<tr>
<td>Alprazolam</td>
<td>1-2 hours</td>
<td>Intermediate/Short (5-20 hrs)</td>
<td>NO</td>
<td>0.5 mg</td>
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<tr>
<td>Chlordiazepoxide</td>
<td>1.5-2.5 hours</td>
<td>Long (5-30 [200] hrs)</td>
<td>YES</td>
<td>15-20 mg</td>
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<tr>
<td>Clobazam</td>
<td>0.5-4 hours</td>
<td>Intermediate (12-60 hrs)</td>
<td>YES</td>
<td>10-20 mg</td>
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<tr>
<td>Clonazepam</td>
<td>1-2 hours</td>
<td>Long (18-50 hrs)</td>
<td>NO</td>
<td>0.25-0.5 mg</td>
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<tr>
<td>Clorazepate</td>
<td>0.5-2 hours</td>
<td>Long ([30-100] hrs)</td>
<td>YES</td>
<td>7.5-15 mg</td>
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<tr>
<td>Diazepam</td>
<td>0.5-2 hours</td>
<td>Long (15-80 [200] hrs)</td>
<td>YES</td>
<td>5-10 mg</td>
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<tr>
<td>Flurazepam</td>
<td>0.5-2 hours</td>
<td>Intermediate ([50-114] hrs)</td>
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<td>10-20 mg</td>
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<tr>
<td>Lorazepam</td>
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<td>Intermediate (6-40 hrs)</td>
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<td>0.5-1 mg</td>
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<tr>
<td>Nitrazepam</td>
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<td>Long (20-40 hrs)</td>
<td>NO</td>
<td>5 mg</td>
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<tr>
<td>Oxazepam</td>
<td>2-4 hours</td>
<td>Short (3-20 hrs)</td>
<td>NO</td>
<td>15 mg</td>
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<tr>
<td>Temazepam</td>
<td>2-3 hours</td>
<td>Short (8-20 hrs)</td>
<td>NO</td>
<td>10-15 mg</td>
</tr>
<tr>
<td>Triazolam</td>
<td>1-2 hours</td>
<td>Very short (2-6 hrs)</td>
<td>NO</td>
<td>0.25 mg</td>
</tr>
</tbody>
</table>

Duration of action:
- Very Short < 6 hours
- Short < 12 hours
- Intermediate < 24 hours
- Long > 24 hours

Ativan 0.05mg/kg q8h for all conversion
Journal Club Schedule 2018-2019

7/3 - Amanda
7/10 - Regina
7/17 - Kim
7/31 - Uzo-Okereke (Adult)
8/7 - Town Hall
8/14 - Ben
8/21 - Yechoor (Adult)
9/4 - Ben
9/11 - Jasia
9/18 - Regina
10/2 - Town Hall
10/9 - Perelstein (Adult)
10/16 - CNS - No journal club
10/30 - Kim
11/6 - Regina
11/13 - Oleg
11/20 - Laws (Adult)
12/4 - Liz
12/11 - Jasia
12/18 - Kim
12/25 - Merry Christmas!
1/1 - Happy New Year!
1/8 - Levasseur (Adult)
1/15 - Town Hall
1/29 - Ben
2/5 - Amanda
2/12 - Garret (Adult)
2/19 - Jasia
3/5 - Kim
3/19 - Hwang (Adult)
4/2 - Alyssa
4/9 - Liz
4/16 - Coman (Adult)
4/30 - Oleg
5/7 - Butt (Adult)
5/14 - Regina
5/21 - Jasia
6/4 - Chou (Adult)
6/11 - Ben
6/18 - Alyssa