Child Neurology Handbook

Children's National Medical Center

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House Staff Rule #1

Every Patient is Your Patient.

Take Ownership of Each Patient You Are Caring for Every Time You Are in the Hospital.

Section 1 –Structure and Expectations for Medical Team on Neurology Wards

Neurology Ward Team Structure

- 4 interns and 1 senior on service each month
  - 2 interns on wards each day from 6am-5pm Monday - Friday
  - 1 intern in Neurology or Endocrine clinic outpatient per week
  - 1 intern on night float from Monday night through Saturday morning
  - 1 senior (PGY3) on wards each month
    - Each ward intern will have one half day of continuity clinic each week and will be covered by another team member
    - Every afternoon will have at least 1 intern and 1 senior only covering neurology on weekdays
    - Seniors will be responsible for all general medical issues on the floor
- The service includes neurology and endocrinology
- Endocrine will round before neurology from 8-830 or after neurology rounds at 11am
- Interns "cap" at 10 patients to preround on in the mornings, ie they cannot write notes on more than 20 patients per day
  - When the ward census is over 20 patients, fellows are responsible for writing notes on patients. This should still be a team led effort and interns, seniors, and fellows should devise a plan to be efficient and improve work flow to facilitate discharge and reduce the census as soon as possible.

Neurology Rounding Rules

- Rounds should occur from 9am to 11am on weekdays, and on 8am to 10am Saturday and Sunday
- Rounding will be presented in the following order: Med Student – Intern – Senior - Nursing, Ancillary Services – Fellow – Attending – Family – Nursing (clarification and order read back)
- Fellows will examine all patients every day before rounds and obtain their own history
- Pre-rounding plans may occur to allow for pediatric interns to be more ready and have viable plans, but this must occur before 9am (timing determined by fellow)
- Family centered rounds to cap at 12 patients, will "table" round on the remainder with attending, fellow, or NP seeing families after rounds and then updating interns on any changes immediately. Stable long term patients and vEEG patients are lowest priority when census exceeds 12 patients.
- Rounds will occur outside of the room, analogous to PICU rounds. Family to have brief time for questions, but rounds will be work focused with majority of counseling and questions being answered by fellow in afternoon. Fellows responsible for rounding on patients independently in the afternoons to develop rapport with patients and families, answer additional questions
- Graduated responsibility for fellows: First half of the year fellows will participate in rounds by stating their assessment and plan but attending will then explain and counsel family during rounds. By January fellows will have observed attending counseling and will start to provide brief explanation and counseling to families
- NP will improve efficiency on rounds by staying with NOS patients or other common conditions if family has a lot of questions

Expectations for Medical Students on Neurology

1. Perform a complete neurologic exam on every patient every day before rounds.
2. Follow patients throughout their entire admission.
    a. If possible, see patients in the ED and complete the initial H+P.
    b. Complete daily progress notes, including daily neurologic exam to ensure consistency in exam, or confirm changes.
    c. Present patients on morning rounds each day **in a concise format**, including pertinent changes or events in the prior 24hrs, pertinent exam findings, test results, assessment, and plan for the day.
        i. When presenting, make the story interesting by explaining the reason for admission with all pertinent details in order to convince the team of what you think actually occurred.
            a. Typically, your HPI should take up 75% of presentation
            b. Be specific about the details of the event
                i. what led up to the event (aura)
                ii. what happened during the event (ictus), including eye position, arms, and legs
                iii. what occurred after the event (postictal)
        ii. Please state all home medications (mg/kg/day) and allergies
        iii. You may decide to only tell pertinent birth, developmental, past medical, family, and social history
        iv. Present weight and **head circumference** on all pts, and a very focused general exam, if necessary, and pertinent neurologic exam findings
        v. Create a reasonable differential diagnosis (top 3) and have a plan.
    d. Complete a discharge summary for the patient, summarizing their hospital course in as few as one and as many as four sentences. Be sure to record pending lab results, pertinent test results (including MRI, EEG, and labs including LP, but not to include routine lab results), home emergency plans, and follow up appointments.
    e. Note: You must forward all notes you write to the intern assigned to the patient every day. Please try to do this before rounds.
3. Attend scheduled conferences, including daily lectures in the neurology team room on Monday and Tuesday at 830am, and Sunrise Lectures on Thursdays at 7am in the neurology library on the 4th floor West Wing.
4. Attend weekly lectures on Wednesday afternoons.
5. If you are going to be absent for illness or interviews, plan accordingly to include coverage for your patients while away, and be sure the residents and fellows are aware beforehand of your absence when possible.

Expectations for PGY-1 Pediatric Residents on Neurology

1. **As an intern, you are the primary physician responsible for each patient. Take ownership of your patients, regardless of if you are night-float, daytime, or covering patients. Their well being is completely dependent on you and you should take this responsibility and privilege seriously.**
2. Perform a complete neurologic exam on every patient you are responsible for every day before rounds, regardless of medical students that may be following your patient.
    a. If possible, see patients in the ED and complete the initial H+P.
    b. Complete daily progress notes, including daily neurologic exam to ensure consistency in exam, or confirm changes.
        i. For billing purposes, all history and physicals notes must include the following:
            a. Chief complaint
            b. History of present illness with at least 4 factors
            c. Birth, Developmental, Past Medical, Medications, Allergies, Family History, and Social History
            d. 10 point review of systems
            e. 18 point exam
            f. Assessment and planning
        ii. For billing purposes, notes must include the following on a daily basis:
            a. Overnight issues with at least 4 components
            b. 2+ point review of systems
            c. 12 point exam
            d. Assessment and planning
    c. If a medical student is following your patient, you must confirm their exam findings every day, listen to their presentation on rounds and add any pertinent details once they are done on rounds speaking, ensure accuracy of all notes they write and correct them as necessary, give feedback to students.
        i. Be sure to add your name and pager number to medical students notes.
    d. If there is no medical student following your patient, you are expected to present patients on morning rounds each day in a concise format, including pertinent changes or events in the prior 24hrs, pertinent exam findings, test results, assessment, and plan for the day.
        i. When presenting make the story interesting by explaining the reason for admission with all pertinent details in order to convince the team of what you think actually occurred.
            a. Typically, your HPI should take up 75% of presentation
            b. Be specific about the details of the event
                i. what led up to the event (aura)
                ii. what happened during the event (ictus), including eye position, arms, and legs
                iii. what occurred after the event (postictal)
        ii. Please state all home medications (mg/kg/day) and allergies
        iii. You may decide to only tell pertinent birth, developmental, past medical, family, and social history
        iv. Present weight and **head circumference** on all pts, and a very focused general exam, if necessary, and pertinent neurologic exam findings
        v. Create a reasonable differential diagnosis (top 3) and have a plan.

vi. Before leaving every patient room, discuss an emergency plan for acute events, primarily seizure, along with the criteria for administering such (ie, ativan 1mg, which is 0.1mg/kg, for seizure longer than 5 mins)
    i. Ativan maximum dose 2mg, regardless of weight

e. Complete a discharge summary for the patient, summarizing their hospital course in as few as one and as many as four sentences. Be sure to record pending lab results, pertinent test results (including MRI, EEG, and labs including LP, but not to include routine lab results).
    i. Discharge summary should include patient's weight, medication and form, concentration, and instructions for families in ml rather than mg. Any titration schedule should be explicitly written out.
    ii. Prescriptions should be given to families prior to discharge date when possible to ensure their ability to fill it and answer any possible questions.
f. Schedule Neurology follow-up (typically at 4-6 weeks but can confirm on rounds) prior to discharge. Appointments should be made ahead of time for patients expected to be discharged over the weekend.
g. Note: You must forward all H+P's and discharge notes to the fellow for review. Please review all prescriptions with the PGY-3 as well before giving to nursing or families.
h. Before rounds arrange for language interpretation services for any non English speaking families.
i. Perform accurate and complete sign out to each team member that will be taking over for your patient each day, and receive similar sign out when back in the hospital.
j. Complete entry of orders discussed on rounds in a timely manner, prioritizing urgent care issues and discharges.
k. Follow up on all consults, test results, and other patient issues and discuss results with senior resident, fellow, or attending concerning plan of action.
    i. Follow up any testing that is pending upon discharge through use of a personal log book. Be sure all tests that are pending on hospital discharge are clearly listed on the discharge summary to allow for outpatient physicians to be aware of them and follow.
l. Inform primary care physician of patient's status and diagnosis on admission and discharge, along with ensuring office follow up at an appropriate time interval.
m. You are responsible for performing lumbar punctures on your patient when necessary. You should coordinate anesthesia when necessary, and expect the PGY-3 resident to guide you through the procedure. Fellows are available for additional help when both PGY-1 and PGY-3 were unable to obtain CSF despite reasonable attempts.

3. Attend your regularly scheduled conferences along with required neurology didactics, including daily lectures in the neurology team room on Monday and Tuesday at 830am, and Sunrise Lectures on Thursdays at 7am in the neurology library on the 4th floor West Wing.
4. Familiarize yourself with resident goals and objectives for neurology, distributed prior to the rotation.
5. Plan to be the primary contact for your patients from nursing, social work, diagnostic services, etc. If you can answer concerns yourself please do so, as a primary goal of residency is to become an independent physician. If you have questions about a

particular issue, please follow the appropriate chain of command, starting with PGY-3 residents or neurology fellows, based on the nature of the question. If you are dissatisfied with the response, please escalate to the attending.

6. Don't accept any admissions an attending or fellow has not told you about.
7. Pick 2 goals for month and achieve them – lumbar puncture, fundoscopic exam, DTR's, interpretation of MRI/CT.
8. ASCOM phones are the enemy to education and safety during rounds. During rounds, if your phone rings while rounding on your patient, hand the phone off. Similarly, do not make calls during rounds in the patient room. Disrupting rounds with constant calls disrupts your education and patient safety significantly.

<u>Expectations for PGY-3 Pediatric Residents on Neurology</u>

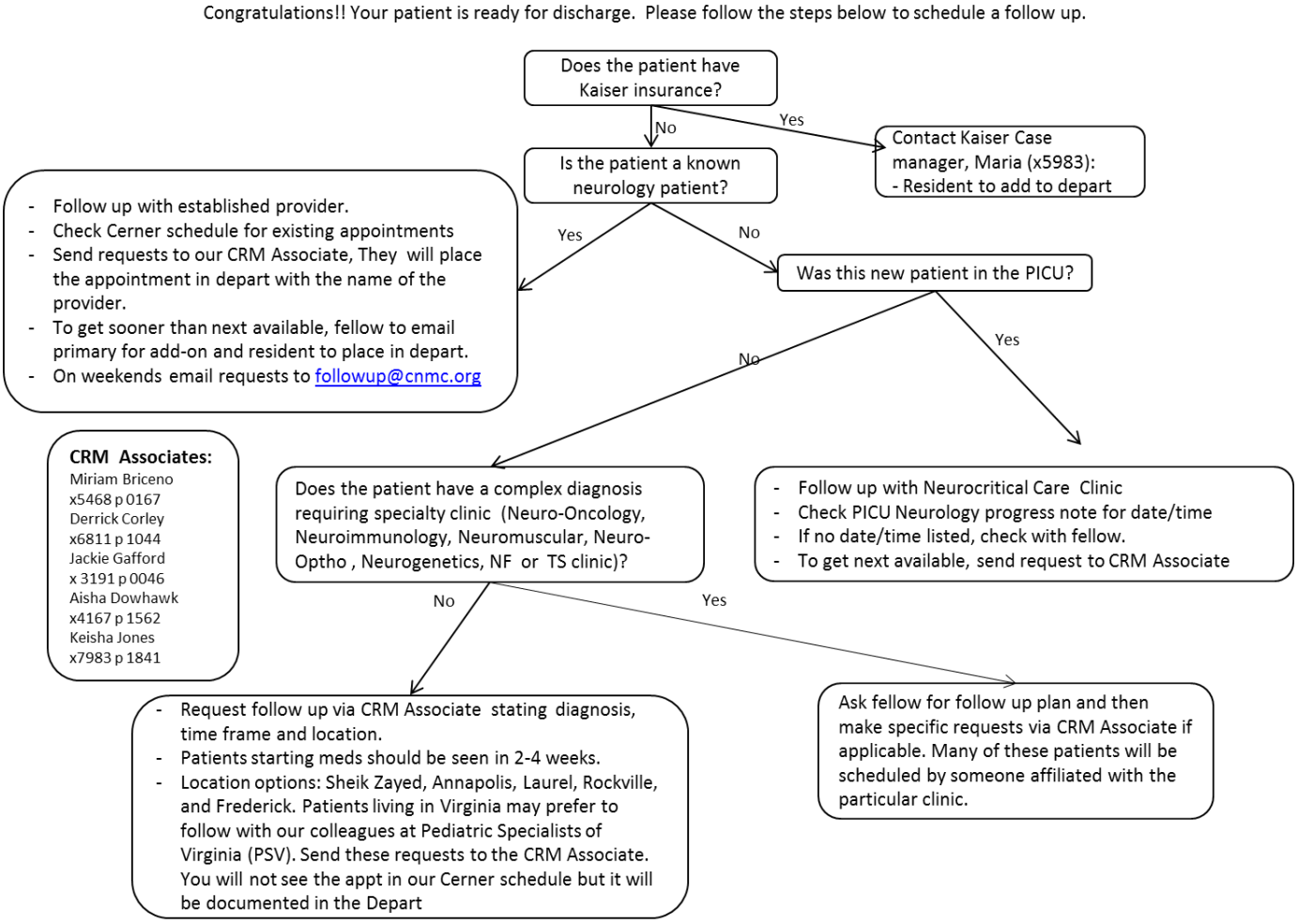
1. Pre-round daily on neurology patients in coordination with medical students and interns as follows:
    a. Confirm key neurologic exam findings on all critical patients on the service, and ensure medical students and interns are examining all patients each day.
    b. **Personally examine all new admissions and sick patients** in the morning before rounds.
    c. Address urgent intern and medical student issues.
    d. Review medication orders and lab results each morning and after rounds.
    e. Complete transfers and admissions that occur between 5am and rounds and hand off to interns at completion of rounds.
2. Identify patients that are acutely ill or decompensating, and triage appropriately with the fellow, attending, nursing staff, and involve the ICU or CAT team when necessary.
3. Ensure PGY-1 residents are writing accurate and complete notes (H+P, progress, discharge, procedure notes), entering orders accurately, that all tests are received in the lab, and when they are not, arrange for repeat collection, and aid in arranging tests ordered (including EEG, MRI, EMG, etc).
4. Once the PGY-1 has finished presenting patients, you should present any additional information you feel is necessary. **You should summarize the plan clarifying the following information on each patient every day if not done by the intern:**
    a. Indications for continued admission, goals of the admission, and discharge criteria should be reviewed daily on each patient and documented in your note.
    b. Tests to be completed, along with if they will occur as inpatient or outpatient (ie, MRI will often be scheduled as outpatient if non-urgent)
    c. Read back all active medication orders before leaving room, including total dose (ie, 10mg BID) followed by weight based dosing in mg/kg/day (5mg/kg/day)
5. You are responsible for observing and teaching PGY-1 residents in the performance of lumbar punctures on their patients when necessary. You should help coordinate anesthesia when necessary, and are expected to guide the PGY-1 through the procedure and perform the procedure when they are unable to. Fellows are available

for additional help when both PGY-1 and PGY-3 were unable to obtain CSF despite reasonable attempts.

6. **All general medical issues should be discussed with PGY-1 residents and appropriately managed and considered your responsibility.** Common pediatric issues such as infections, feeding and nutrition, respiratory issues, etc, will be diagnosed and managed by PGY-3 with questions directed to fellows or attending when necessary.
7. **In the morning, review all labs and active orders daily to ensure accuracy.**
8. Ask the attending on service to round on post-call resident's patients first. If this is not happening discuss with the fellow or ask your chief to help.
9. In conjunction with the neurology fellow and charge nurse, be sure an orientation to 5 East Neuroscience Floor occurs during the first month of service.
10. Daily safety rounds occur at 3:20 PM and afternoon rounds should start at 3:30 PM. This is required and should include on call residents, fellows, charge nurse, and case management.
11. Check anesthesia orders that are entered by the PGY-1 for procedures such as LP. If the PGY-1 is unable to do so, you should ensure the tray and needles are ready and also ensure that the CSF reaches the lab and is processed correctly. Inform fellow when this is completed.

INSTRUCTIONS FOR HOW TO SCHEDULE NEUROLOGY FOLLOW-UP VISITS

Start the discharge appointment process during the admission, not the discharge, because it can take several days to finalize this.



Expectation for Pediatric Neurology Fellows on Ward Service

1. As the pediatric neurology fellow on service, you serve as the primary neurology contact for each patient.
    a. You must ensure that you have updated information available on pertinent history and exam findings, current medications, lab results and pending labs, and EEG and imaging results.
    b. Before rounds, personally obtain a history for each new patient and an interim history for previously admitted patients, examine every patient, and review diagnostic results and notes on all current service patients.
    c. Draft a list of patients that need imaging and lists of patients that need EEGs, those that need to stay on EEG, and those that can be taken off EEG monitoring to review with the EEG techs.
2. Perform a complete neurologic exam on all patients admitted to the floor. This must be performed prior to rounds. In cases where examination will alter management, this is always done urgently. If there are abnomalities, please be sure residents are aware of them. Any abnormal findings should be confirmed on a daily basis to ensure its time course and observe for additional abnormalties.
3. During rounds, listen to PGY-1 and PGY-3 presentations, and confirm or refute any details after they are complete. You should have a reasonable differential diagnosis on all patients, and create a plan for work up and treatment, including specific medications, forms (liquid, sprinkle tablets, IV, etc), specific dosing and titration schedule, and long term management plan (duration of treatment, etc). Do not provide counseling to the family or an explanation during work rounds; please go back in the afternoons to further explain diagnoses, testing, and management to families.
4. Rounds should begin at 9AM and end by 11AM. If the attending is unavailable, the fellow may begin rounds at the discretion of the fellow and attending.
    a. Daily afternoon rounds are required with on call residents, fellows, nursing, and case management, and they should start at 3:20 for Safety Rounds and 3:30 PM for Discharge Rounds and be completed as soon as possible.
5. Review all H+P's, discharge summaries, orders, and prescriptions on each patient to ensure accuracy and make corrections as needed. A summary statement should be added including at least one sentence summarizing the history, along with plans for management. Once completed, forward to neurology attending for signature. Be sure all patients have appropriate follow up appointments.
    a. For billing purposes, confirm that all history and physicals notes must include the
    following:
        i. Chief complaint
        ii. History of present illness with at least 4 factors
        iii. Birth, Developmental, Past Medical, Medications, Allergies, Family History, and Social History
        iv. 10 point review of systems
        v. 18 point exam
        vi. Assessment and planning
        vii. For billing purposes, notes must include the following on a daily basis:
        viii. Overnight issues with at least 4 components
        ix. 2+ point review of systems
        x. 12 point exam

xi. Assessment and planning

b. All notes on patients seen during rounds should be completed before 12pm each day. All H+Ps and discharge summaries should be signed and forwarded to attendings within 24 hours of resident completion.

6. Respond to nursing, resident, and family issues on the floor in a timely and professional manner.
7. Discuss test results and management decisions with families every afternoon to ensure their understanding, answer questions, and create an appropriate discharge plan. This usually requires you to go back to patient rooms after rounds and sit with families.
8. Be sure all tests that are pending on hospital discharge are clearly listed on the discharge summary to allow for outpatient physicians to be aware of them and follow.
9. Personally review EEGs, lab results, and neuroimaging on all patients to ensure accuracy and create appropriate management plans.
    a. Review EEG results with the neurophysiology team after service rounds are complete and update the neurophysiology team about critical new EEGs (those whose results may immediately change management or effect discharge planning).
    b. If possible the neurology service team should review imaging with Neuroradiology.
10. Supervise and manage adult neurology residents, rotating residents, and medical students to ensure efficient use of time and appropriate triage of patients and issues.
    a. You must utilize your available resources, which requires management of all members of the medical team. You will not have time to perform a complete H+P on every consult in every unit, but should be able to prioritize them, have students and residents perform a complete evaluation, be presented the information for confirmation of findings and management decisions, and oversee presentations to attendings.
    b. On days where a team member is unavailable due to clinic/post-call/other responsibilities, plan ahead by rounding with the attending early or receiving sign out on patients that need to be rounded on later in the day.
    c. Contact teams that have requested neurology consults to inform them of our impression and recommendations, as well as best way to follow-up with neurology.
11. Ensure education of residents and students occurs as follows:
    a. Resident and student teaching should occur at 8:30 AM on Mondays and Tuesdays
    b. On Wednesdays at 8 AM, Pediatric Grand Rounds attendance is required for pediatric residents and students
    c. Sunrise Lecture on Thursdays at 7 AM is optional for pediatric residents and students.
12. Contact nurse managers (Audrey Scully) and scheduling (John Schultz) to arrange urgent appointments requested by outside physicians or by the rotator on call the night before. This is best achieved by emailing Neurology_Scheduling@childrensnational.org
13. Assist Neurology attendings who refer patients for urgent direct admission (attending is responsible for obtaining prior-authorization when needed and calling admissions)
    a. Coordinate direct admissions from outside hospital by discussing with attending.
    b. Personally examine any patients in the ER or hospital with a concern for acute worsening of neurologic status. Patients with paroxysmal events may be

triaged for a later time, but patients not waking up, actively seizing, or degenerating require urgent evaluation. Additionally, if an attending physician asks you to see a patient coming to any hospital area you should do so once you are made aware of their presence in the hospital. If you are unable to see them because of other patients that require higher urgency, notify the attending on service or the one that asked you to see the patient.

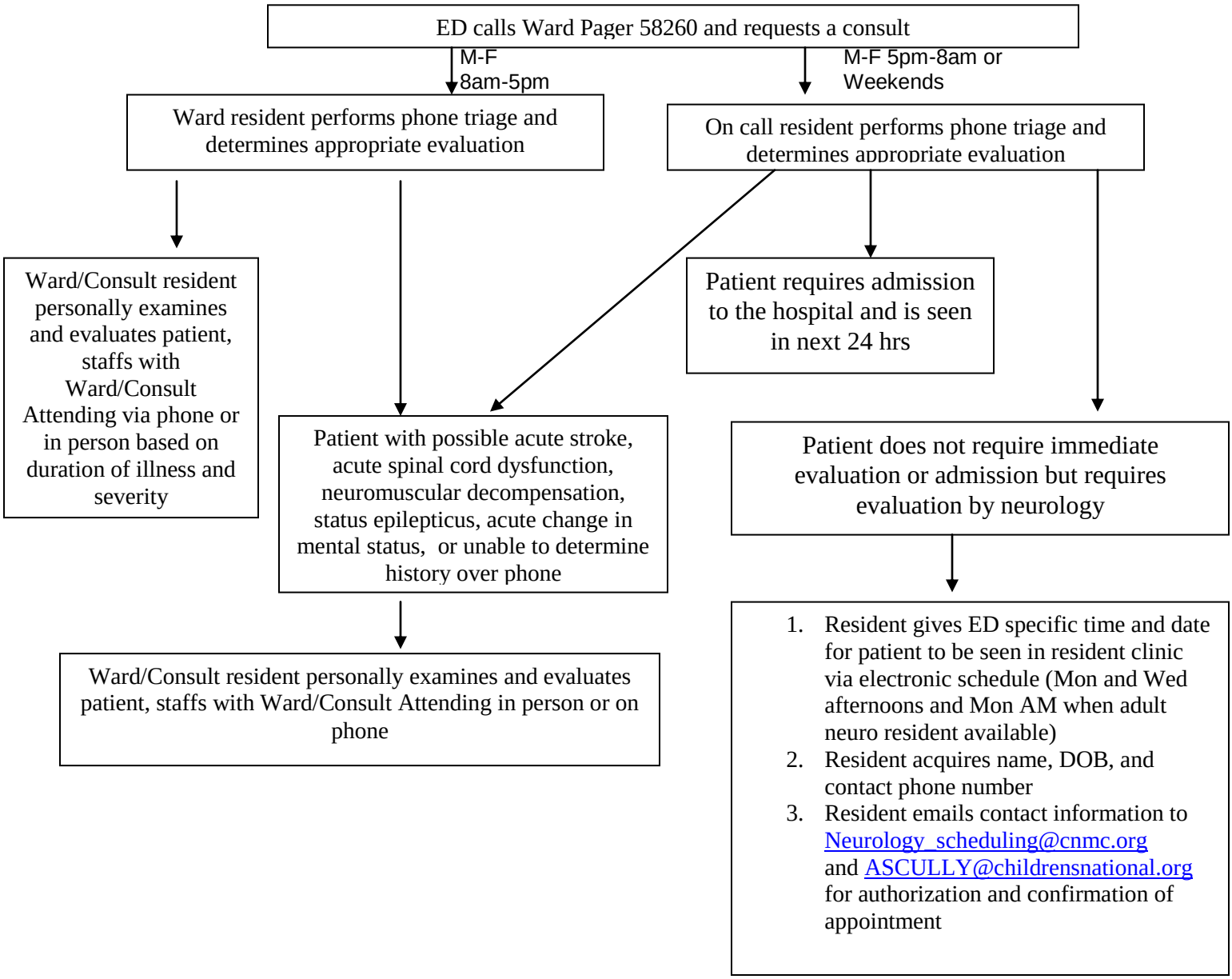
i. Communicate back to the referring neurology attending about the outcome of the ER visit and/or admission
    1. at time of discharge
    2. if findings are different from expected
    3. if family meetings are planned

14. Fellows are personally responsible for 23 hour EEG patients in cases where the nurse practioner is unavailable (Tuesdays and vacations), including performing H&Ps, rounding with attending on day of admission, completing discharge summaries (patients should always be priority discharge), entering home medications and orders, and signing-out to fellow on call who will primary contact for nursing.
15. When the ward census is over 20 patients, fellows are responsible for writing notes on patients. This should still be a team led effort and interns, seniors, and fellows should devise a plan to be efficient and improve work flow to facilitate discharge and reduce the census as soon as possible.

Child Neurology Call Coverage:

- PGY-5s should take a minimum of three weekday calls per month with one call free month, but may cover weekends on circumstances such as unexpected emergencies, vacation coverage
- PGY-4s take 5-8 calls per month with the following rules:
    - PGY-4 and 5's should round post-call when they are followed by a PGY-3 to avoid them being alone on that day.
    - PGY-4 and 5's do not need to round post-call when they are followed by another PGY-4 or 5, but may need to do so at the request of the attending or following day resident in circumstances of a busy hospital patient census.
- PGY-3s should take weekday buddy call only for the first 3 calls with PGY-5 (chief) and then can cover weekends thereafter with following rules:
    - PGY-3s should round when post-call except when there is an adult rotator assigned to that day in addition to the child neurology fellow.
    - PGY-3s should not round on weekends alone. The other person can be a PGY-4 or 5, or an adult or Walter Reed rotator.

Neurology ED Consultation



Expectation for Child Neurology Attendings

1. Rounds should be conducted from 9-11am daily, and you should round on post-call resident patients first to avoid violation of ACGME duty hours requirements.
   a. Rounds are family centered. This means that the family is invited to witness and participate in rounds. The family should participate in and understand the goals of the day.
   b. The attendings should limit their activities on rounds to medically necessary activities. Bonding with patients and their families can be done in the afternoon after formal rounds. If a family needs more time to discuss the clinical situation this should be done outside of rounds, with or without the fellows and residents.
   c. Rounds are multidisciplinary and must include nursing, along with other specialties including PT/OT/ST, nutrition, social work, etc. Nursing observations from overnight should be expressed and nursing must understand the goals of the day.
2. The period of time between 8:30 and 9:00 AM on Monday and Tuesday will be used for lectures to the pediatric residents. You are responsible for giving one of these lectures each week and should be coordinated with the fellows.
3. Consult rounds should be done immediately after wards starting between 11AM-1PM when possible, however based on fellow availability, may be moved as necessary. All consults that are urgent should be seen by fellows and staffed immediately, and those for more chronic issues may be seen by fellows and rounded on the following day, time permitting.
4. The neurology fellows and residents have to be able to attend electrophysiology rounds on Tuesday at 11:30 (this is mandatory). They also need to be at the Thursday noon time teaching conferences from 12 - 2 PM (this is mandatory).
5. All attendings should have a formal sign out with each other when covering for weekends, end or service, etc. The format is to the discretion of the attending receiving the sign out, and may be in person, via telephone, or email. This needs to occur anytime there is a change in patient coverage, and trainees need to be made aware of the attending that is responsible.
6. A time should be set where the inpatient ward attendings interact with the neurophysiology attending and neuroradiologists to review actual EEG tracings and radiolographic data. Fellows and any other interested team members should also be present for this.
7. Daily afternoon rounds are required with on call residents, fellows, nursing, and social work, and they should start at 3:20 PM for Safety Rounds and 3:30 PM for Discharge Rounds and be completed as soon as possible.
8. Notes need to be co-signed daily in order to bill for patient care. The attending cannot co-sign a note unless the attending physically interacts with the patient.
9. Interpreters must be used for non-English speaking families. This can be coordinated through the hospital translation services and is done best if arranged before rounds. These families must receive the same care and have the same degree of understanding as English speaking families.
10. You are responsible to answer urgent issues for other attendings' patients while they are unavailable due to conferences or vacation. This should not be referred back to the fellow.
11. Billing needs to reflect the level of care. Remember that times spent reviewing lab data, films and EEGs count. Reading notes from TVES and EPRS, reviewing medical literature all are a part of medical care and should appropriately noted. Document discussions with radiologists, neurophysiologists, and consulting physicians.

Expectations for Nursing

1. All patient issues should be communicated to the resident responsible for the patient. This is likely the PGY-1 or PGY-3 residents. Fellows should not be directly contacted with patient issues as this will take away valuable educational experiences from trainees, and disrupt the flow of information and patient care.
2. All non-urgent phone calls and pages to trainees should to be held from 9-11am during rounds to avoid disruption and provide safe care. Urgent issues may include acute events such as seizures, change in patient status, and need for urgent medications, or other events seen as urgent should be brought to the attention of the trainees immediately as necessary.
3. Verify medications given while in the hospital with family and the medical team for dosage and frequency. If a medication or dose seems incorrect, please call the trainee or pharmacy to avoid a mistake.
4. Verify medications for discharge with family and the medical team for dosage and frequency. If a medication or dose seems incorrect, please call the trainee or pharmacy to avoid a mistake.

Pediatric Housestaff Goals and Objectives

Pediatric Resident Goals and Objectives

N. Neurology

GOAL 7.73 Normal Versus Abnormal (Neurology). Understand how to differentiate between normal and abnormal states of the neurologic system.

OBJECTIVES:

a. Recognize normal infant and child developmental milestones including gross and fine motor, social, and language development
b. Demonstrate a skillful neurologic exam including primitive reflexes, mental status, cranial nerves, motor , sensory, cerebellar function and gait, and deep tendon reflexes
c. Understand basic neuroanatomy in order to be able to localize historical and exam components to specific structures within the nervous system.

GOAL 7.74 Common Conditions Not Referred (Neurology). Understand how to diagnose and manage common conditions which generally do not require referral.

OBJECTIVES:

a. Recognize and institute an appropriate basic diagnostic and management plan of these conditions:

    1. Simple febrile seizure
    2. Absence epilepsy
    3. Primary headache disorders
    4. Speech delay
    5. Positional plagiocephaly
    6. Attention Deficit Hyperactivity Disorder
    7. Behavioral Disorders
    8. Transient motor or vocal tic disorders
    9. Familial macrocephaly
    10. Mild closed head trauma and simple linear skull fractures

GOAL 7.75: Conditions Generally Referred (Neurology). Understand how to recognize, initiate management of, and refer conditions which generally require referral.

OBJECTIVES:

a. Recognize, provide appropriate interim care, and appropriately refer these conditions:

    1. Unprovoked seizures, complex febrile seizures, epilepsy, infantile spasms
    2. Headaches not responding to basic treatment or worrisome for secondary processes
    3. Disorders of increased intracranial pressure, including idiopathic intracranial hypertension, hydrocephalus, hemorrhage, tumors
    4. Global developmental delay and autism
    5. Cerebral palsy and other abnormalities of tone
    6. Chronic motor or vocal tic disorders, Tourette Syndrome
    7. Movement disorders including chorea, hemiballismus, athetosis, myoclonus
    8. Spinal cord pathology
    9. Gait dysfunction and ataxia
    10. Neuromuscular disorders and muscle weakness
    11. Disorders of myelination, including multiple sclerosis, leukodystrophies
    12. Neurocutaneous syndromes
    13. Stroke
    14. Infections of the nervous system including encephalitis, meningitis, and Lyme disease
    15. Acute or recurrent encephalopathy

GOAL 7.76: Seizures (Neurology). Understand how to evaluate, manage, and refer patients with seizures and spells concerning for seizure.

OBJECTIVES:

a. Recognize and provide emergency and interim treatment for seizures and spells concerning for seizure, and provide basic inpatient care for patients with epilepsy:

    1. Recognize basic features of seizures including those events leading up to the seizure, the appearance of seizures themselves, and the postictal period
    2. Discriminate between epileptic and non-epileptic spells by clinical history and recommend appropriate testing or referral
    3. Understand the basic classification of seizures as simple partial, complex partial, and generalized seizures
    4. Provide families with a basic seizure emergency plan including

appropriate use of rectal diazepam
5. Have a basic knowledge of the routine diagnostics performed for seizures including EEG and neuroimaging studies
6. Understand the basic indications for initiation of treatment for seizures
7. Understand the basic indications for different antiepileptic drugs and their major side effects and interactions
8. Provide families with realistic expectations and precautions for patients with recurrent seizures

GOAL 7.77: Headache (Neurology). Understand how to evaluate and manage headaches.

OBJECTIVES:

a. Recognize and provide basic diagnostic and treatment plans for children presenting with headache in both the inpatient and outpatient settings:

    1. Elicit a thorough headache history including headache location, quality, intensity, duration, frequency, associated symptoms, and family history of headaches
    2. Recognize symptoms and signs of secondary headache disorders
    3. Discern qualities of different primary headache disorders including migraine with and without aura and tension headache
    4. Perform a reliable and focused neurologic exam on headache patients to help rule out secondary headache disorders
    5. Understand indications for further testing for headaches including neuroimaging, lumbar puncture, and ophthalmology examination
    6. Provide basic outpatient treatment for headaches, including lifestyle modification, avoidance of medication overuse, and basic first line therapies
    7. Provide basic inpatient abortive treatment for chronic migraines along with understanding drug pharmacology, side effects, and efficacy
    8. Understand basic indications for preventative medications, evidence for their efficacy, and side effects

GOAL 7.78: Complex neurological testing (Neurology). Understand the types of complex neurologic testing available, the indications for ordering such tests, and basics of interpretation.

OBJECTIVES:

a. Understand indications for, limitations of, basic interpretation of, risks, and

cost of the following procedures and tests:

a. Lumbar puncture
b. Electroencephalogram (EEG)
c. Head computerized tomography scan (CT)
d. Head magnetic resonance imaging scan (MRI)
e. Electromyography (EMG) and nerve conduction velocity (NCV)

GOAL 7.79: Neurological Pharmacology (Neurology). Demonstrate a basic understanding of neurological medications, mechanisms of action, indications, monitoring, and side effects.

OBJECTIVES:

a. Understand basic mechanisms of action and indications for the most common neurologic drugs, including anti-epileptics and migraine agents
b. Provide realistic expectations to families concerning side effects of these agents and monitor with appropriate lab tests
c. Understand contraindications and interactions of common neurologic drugs

GOAL 7.80: Prevention (Neurology). Understand the pediatrician's role in the prevention and management of neurological disorders.

OBJECTIVES:

a. Avoidance of injury due to seizure or loss of consciousness including appropriate driving restrictions
b. Counsel parents/patients about prevention of head and spinal cord trauma (seat belts, car seats, helmets, firearm safety, diving injuries)
c. Counsel parents about prevention related to environmental toxins (e.g., lead) and household poisonings.
d. Early intervention for developmental delay
e. Avoidance of medication discontinuation due to lack of patient education, intolerable side effects, and inappropriate use
f. Avoidance of medication overuse in patients with neurologic conditions

Pediatric Resident Neurology Suggested Lecture Schedule

The team and fellows may decide to make changes in lectures based on educational needs and patient exposure.

	Monday	Tuesday	Wednesday	Thursday	Friday
Week 1					
	830-9am Neurologic Exam (required)	830-9am Localization (required)		7-8am Sunrise Conf (optional) 1-2pm Intake Conf (optional)	
Week 2					
	830-9am Cerebral Palsy (required)	830-9am Acute Encephalopathy (required)		7-8am Sunrise Conf (optional) 1-2pm Intake Conf (optional)	
Week 3					
	830-9am Increased Intracranial Pressure (required)	830-9am Tics and Tourette Syndrome (required)		7-8am Sunrise Conf (optional) 1-2pm Intake Conf (optional)	
Week 4					
	830-9am Neurocutaneous Syndromes (required)	830-9am Neuromuscular Disorders (required)		7-8am Sunrise Conf (optional) 1-2pm Intake Conf (optional)	

Resident Lecture Series:

Neurologic Examination
Resident Lecture Series

1. Mental Status
    a. Orientation to person, place, time, and situation
        i. Use familiar references like cartoons
    b. Attention and concentration
    c. Memory
    d. Language
        i. Body parts, numbers, colors, alphabet, reading, writing
2. Cranial Nerves
    a. Olfactory – rarely tested
    b. Optic – visual fields, Snellen chart, fundoscopic exam
    c. Oculomotor, Trochlear, Abducens – eye movements
    d. Trigeminal – facial sensation, corneal reflex
    e. Facial – facial movements
    f. Vestibulocochlear – hearing, nystagmus
    g. Glossopharyngeal, Vagus – phonation and palate movement
    h. Accessory – SCM and shoulder shrug
    i. Hypoglossal – tongue movement
3. Muscle strength, tone, and bulk
    a. Graded 0-5 as follows
        i. 0 – no movement
        ii. 1 – trace movement
        iii. 2 – movement with gravity eliminated
        iv. 3 – movement against gravity with no resistance
        v. 4 – movement against slight resistence
        vi. 5- normal strong movement
    b. Abnormalities and cause:
        i. proximal weakness – myopathy
        ii. distal weakness – neuropathy
        iii. unilateral weakness - UMN lesion inlcuding stroke
        iv. spastic hypertonia (clasp knife)- CP, old stroke
        v. rigid hypertonia (lead pipe or cogwheeling) – Parkinsons
        vi. paratonia (gegenhalten) - frontal lobe dementia
        vii. hypotonia - myopathy
4. Sensation
    a. Light touch, vibration, temperature, pain, proprioception
5. Coordination
    a. Finger nose finger, heel shin heel
    b. Rapid alternating movements
6. Reflexes
    a. Deep tendon reflexes graded 0-4 as follows
        i. 0 – no reflexes
        ii. 1 – trace reflex
        iii. 2 – normal reflex
        iv. 3 – increased reflex crossing two joints

v. 4 – increased with clonus

b. Primitive reflexes
    i. Moro – normal 0-6mos – mediated by vestibular nuclei brainstem
    ii. Tonic neck – normal 0-6mos - mediated by vestibular nuclei brainstem
    iii. Grasp – normal 0-6mos - mediated by vestibular nuclei brainstem
    iv. Rooting – normal 0-6mos - mediated by trigeminal nuclei
    v. Trunk incurvation – normal 0-9mos – mediated by spinal cord level
    vi. Parachute – normal after 6-8mos and never disappears – mediated by vestibular nuclei brainstem

c. Babinski defined as upgoing toe against noxious stimuli to plantar surface of foot indicating UMN process

7. Gait
    a. Normal gait
    b. Tandem gait

Neurologic Localization for the Pediatric Resident
Resident lecture series

Localization Teaching Points:

1. Upper motor neuron lesions are characterized by:
Hypertonia (Spasticity for corticospinal tracts; rigidity for extrapyramidal tracts)
Hyperreflexia
Clonus
Extensor plantar response

2. Lower motor neuron lesions are characterized by:
Hypotonia
Weakness
Hyporeflexia
Fasciculations

3. Cortical lesions are associated with cortical type deficits:
Dysphasia for language cortex
Visual disturbances for occipital cortex
Hemiparesis for motor cortex

4. Brainstem lesions are associated with cranial nerve signs in combination with long tract and crossed signs, i.e. weakness of ipsilateral face and contralateral body

5. Spinal cord lesions are characterized by:
    a. Sensory level
    b. Change in bowel or bladder function

EXAMPLE: Gait Disturbances

Cortical: dyspractic (frontal lobe), circumductive
Basal ganglia: shuffling/festinate, choreic/dancing
Cerebellar: ataxic
Myelopathy: scissoring
Radiculopathy: antalgic
Neuropathy: steppage
Neuromuscular junction: fatiguable
Myelopathy: waddling

Cerebral Palsy

Resident lecture series

Background

- Cerebral palsy (CP) can be defined as a disorder of motor control and posture secondary to a static CNS lesion that is not the result of a recognized progressive or degenerative brain disease.
- The brain abnormality may occur pre-, peri-, or postnatally.
- Children with CP do not have progressive loss of function as would be seen with a metabolic d/o

Epidemiology

- CP is a common problem, occurring in about 2 to 2.5 per 1,000 live births.
- CP occurs more commonly in children who are born very prematurely or at term.
    - Data from Sweden on 241 children with CP indicate that 36% were born at a gestational age (GA) of less than 28 weeks; 25% at 28 to 32 weeks GA; 2.5% at 32 to 38 weeks GA; and 37% at term.

Risk Factors

- Prematurity - 50-60% of all CP
- Hypoxic Ischemic Injury
- Asphyxia – Low APGAR, low pH, multiorgan failure, early seizure
- Other: trauma, multiple gestation, TORCH, stroke, chorioamnionitis

Classification

- Spastic - 75% of cases including:
    - Diplegic- 44% of all CP types with UE affected less than LE
        - Periventricular leukomalacia
        - Low seizure risk
    - Quadriplegia - 6% of all CP types
        - hypoxic damage in full term
        - intrauterine disease(TORCH)
        - cerebral malformations
        - most severe form with worst cognitive and seizure prognosis prognosis; often swallowing difficulty with supranuclear bulbar palsy, aspiration pneumonia common; may have athetosis in mixed form
    - Hemiparetic - 33% of all CP types due to vascular insult or cerebral malformations; may have thrombophilic disorder
    - Paraparetic-involves lower half of body only, rare
- Dyskinetic - 10% of cases including:
    - choreoathetotic- kernicterus, genetic, metabolic, sudden hypoxia as with placental abruption; full term often; poor head control, poor feeding, and development of increased tone and dystonia over many years
- Rigid
- Atonic
- Ataxic
- Mixed

Diagnosis

AAN Practice Parameter: *Neurology* 2004;62;851-863:
1. In order to establish that a brain abnormality exists in children with CP that may, in turn, suggest an etiology and prognosis, neuroimaging is recommended with MRI preferred to CT (Level A).
-MRI abnormal in 90%; CT abnormal in 77%

2. Metabolic and genetic studies should not be routinely obtained in the evaluation of the child with CP (Level B).

3. If the clinical history or findings on neuroimaging do not determine a specific structural abnormality or if there are additional and atypical features in the history or clinical examination, metabolic and genetic testing should be considered (Level C). Detection of a brain malformation in a child with CP warrants consideration of an underlying genetic or metabolic etiology.
-Rate of finding etiology in this study was very low at 0-4%

4. Because the incidence of cerebral infarction is high in children with hemiplegic CP, diagnostic testing for coagulation disorders should be considered (Level B). However, there is insufficient evidence at present to be precise as to what studies should be ordered.
- Hypercoagubility work up is positive in 13-37% of hemiplegic CP

5. An EEG is not recommended unless there are features suggestive of epilepsy or a specific epileptic syndrome (Level A).
-45% of all CP will develop seizures

6. Because children with CP may have associated deficits of mental retardation, ophthalmologic and hearing impairments, speech and language disorders, and oral-motor dysfunction, screening for these conditions should be part of the initial assessment (Level A).

Treatment

- Multidisciplinary approach to avoid complications and manage disabilities
- PT/OT/ST - 50% of CP will require assistive devices, 70% with other disabilities (MR common)
- PM+R, psychiatry, surgery, nutrition, ophthalmology
- Surgery- tendon lengthening, dorsal rhizotomy
- Botulism toxin injections
- Baclofen

Acute Encephalopathy

Resident Lecture Series

Definitions

Coma = total or near-total unresponsiveness. Not arousable to any stimuli

Stupor = state of severely impaired arousal. Some responsiveness to vigorous stimuli.

Obtundation = some responsiveness to touch or voice

Lethargy/Somnolence = state in which pt has diminished arousal but is able to maintain arousal spontaneously or with repeated light stimulation.

Acute Confusional State = inadequate arousal to perform a coherent thought or action (inattention, disorientation)

Delirium = state of confusion w/periods of agitation, irritability, hallucinations. Typically alternates with periods of decreased arousal.

Localization

Results from damage to components of bilateral cerebral cortex or reticular activating system (midbrain and rostral pons to thalamus to cerebral cortex)

Etiology

Trauma (bleeding, concussion), intoxication (medications or illicit drug), infection (meningitis, encephalitis), metabolic disturbance (lytes, thyroid, mitochondrial disorders, inborn error of metabolism), vascular (acute ischemic stroke, venous thrombosis, CNS vasculitis), seizure/post-ictal, hypoxia, paraneoplastic process, ADEM, diffuse motor weakness mimicking encephalopathy (e.g. botulism), migraine (acute confusional), psychogenic

History considerations: Time course? Medications? History of trauma? Headache, Hemiparesis, Ataxia, Vomiting? Medications at home? History of psych illness? History of seizure? Occupational, environmental exposures?

Exam: Vitals (especially fever), rash, bruising/petechiae, ulcerations (including genital), neurocutaneous stigmata, hepatosplenomegaly, lymphadenopathy, murmurs, ticks, insect bites, nuchal rigidity, language function since sometimes "mental status changes" is really aphasia

Management

Head CT

UTox, EtOH, GCMS, chemistry with glucose, ammonia, coagulation studies, blood and urine culture

LP after CT including routine studies, HSV PCR, gram stain, enterovirus and arbovirus PCR

Antibiotics if suspect infectious etiology to include acyclovir, ceftriaxone/cefotaxime

Neonates: ampicillin and gentamicin

Consider vancomycin if possible resistant bacteria

Consider MRI if etiology is unclear on CT, suspect ADEM, tumor, etc

EEG and antiepileptics is suspect seizure/nonconvulsive status epilepticus

Steroids, IVIG in suspicious cases for inflammatory conditions

NPO until awake, compression stockings, frequent neuro checks

Seizures and Epilepsy
Resident lecture series

Tips for history taking: Goal is to answer two questions: 1. Seizure or not? 2. If seizure, was it focal or generalized?

1. Were they Conscious → did they respond to questions, tactile stimulation or look around the room? Do they remember people talking to them during the event?
2. Were they Distractible → could the event be stopped by noise / talking to the patient or by external stimuli including touching, tickling, or pinching the child?
3. What was the Movement and Progression→
   Limbs: jerking, stiff, limp, bilateral or unilateral, rhythmic, synchronous, and/or stereotyped
   Face: was head deviated to one side, lips smacking, chewing, drooping, and/ or twitching
4. What did the Eyes do→ open or closed, rolled back, forward or stuck to one side
5. Other questions
   - Loss of bladder or bowel
   - Color change
   - Unusual noises / cry
   - Tongue biting
   - Length of event
   - What was patient doing before the event: was it associated with sleep?
   - What did patient do after the event: sleepy or confused and for what length of time

Definition of Epilepsy: More than one unprovoked seizure separated by 24hours.

Epidemiology: The incidence of epilepsy (adult and childhood) in the USA is approximately 1% of the population but 4-10% of children will have a seizure by 16yrs of age.

Generalized Seizure:

1. The first clinical and EEG changes indicate initial involvement of both hemispheres

2. Etiology
   a. Genetic
   b. Metabolic
   c. Electrolytes
   d. Glucose
   e. Unknown

3. Clinical characteristics:
   a. All generalized seizures will have loss of consciousness
   b. Eyes will be open. Sometimes described as "rolled back or staring" but should not have eye deviation to one side as this is a localizing characteristic.
   c. Types of movements include tonic, clonic, myoclonic, atonic and staring
   d. Movements will be symmetric and rhythmic

Focal Seizure:

1. The first clinical and EEG changes show initial activation of one part of one cerebral hemisphere.

a. In some cases there is more than one network (multifocal), and more than one seizure type, but each individual seizure type has a consistent site of onset

2. These are the most common type of seizures during childhood.
3. Etiology
    a. Cortical malformations
    b. Mesial temporal sclerosis (hippocampal sclerosis)
    c. Trauma
    d. Strokes
    e. Tumors
    f. Infection
    g. Electroyltes
    h. Glucose
    i. Genetic (multifocal)
    j. Unknown
4. Clinical characteristics :
    a. May have associated aura including sensory, gustatory, olfactory, emotional
        i. Unusual smells, fear, laughter
    b. Pure motor or sensory symptoms in one specific area or one side of the body
        i. Unilateral hand or limb jerking, head and/or eye deviation, facial twitching
        ii. Tingling or unusual sensation in one limb
        iii. Visual symptoms; ie. lights flashing
    c. Automatisms: staring, lips smacking, chewing, drooling, picking
    d. Subclasses
        i. Focal without impairment of consciousness (Simple Partial)
        ii. Focal with impairment of consciousness (Complex Partial): behavioral arrest and unresponsiveness

Focal Seizure with secondary generalization:

1. The first clinical and EEG changes show initial activation of one part of one cerebral hemisphere and then spreads to include both hemispheres.
2. Manifest clinically as GTC seizures but history gives clues to a focal onset.

Unclassified: Epileptic spasms

Epilepsy Syndromes

1. Absence Epilepsy
    a. Onset 4-10yrs; Peak 6-7yrs
    b. Clinical characteristics :
        i. Behavioral arrest with a motionless stare
        ii. Lasts 10 to 15 seconds
        iii. Eyelids may droop or flutter.
        iv. Resume their full activity after the seizure or may be very briefly confused (<30 seconds).
    c. EEG: bilateral generalized 3Hz spike and wave discharges
    d. Can be provoked by Hyperventilation in 30%

e. Treatment: Ethosuximide is first line; evidence supports use of valproic acid and lamotrigine also
f. Many children grow out of these by 12-16yrs of age
g. About 15% evolve into JME

2. Juvenile Myoclonic Epilepsy
    a. Onset at 5-15yrs
    b. Clinical characteristics
        i. Bilateral Myoclonic jerks on awakening/ early morning. ("Clumsy in the morning" "Spills juice")
            1. Not associated with loss of consciousness
        ii. GTC seizures (90%)
        iii. Development of absence seizures (~1/3rd of patients)
    c. EEG: generalized 4-6Hz polysplike and slow wave discharge
    d. Can be provoked by Photic Stimulation, sleep deprivation and etoh.
    e. Treatment: Valproic Acid, Keppra, Lamictal

Note: The term myoclonus refers to quick, involuntary muscle jerks that involve any part of the neuroaxis. They do not always originate from the cortex and thus are not always epileptic. Examples include post anoxic myoclonus and sleep myoclonus.

3. Benign Childhood Epilepsy with centrotemporal spikes (BECTS; Benign Rolandic Epilepsy)
    a. Onset at 2-13yrs (peak 9-10yrs)
    b. Clinical characteristics
        i. Unilateral facial sensory-motor symptoms
        ii. Oropharyngogutteral symptoms: hypersalivation and speech arrest
        iii. Examples:
            1. Child is drowsy but has difficulty speaking and is drooling with unilateral facial twitching
            2. Unilateral Facial twitching and a "funny sensation"
        iv. Majority occur during sleep
        v. Many present with GTC seizures due to secondary spread
        vi. Development is not impaired
    c. EEG: biphasic,independent focal centrotemporal spikes and slow waves with horizontal dipole.
    d. Many children grow out of these by age 16 and they often do not occur more than 5 times total
    e. Can be provoked by sleep deprivation
    f. Treatment: Decision to treat is based on frequency, parental anxiety and overall degree to which they are disturbing the patient's life. Goal is also to prevent GTC seizures. Oxcarbazepine is first line.
4. Mesial Temporal Lobe Epilepsy +/- mesial temporal sclerosis
    a. Mesial Temporal Sclerosis: hippocampal sclerosis with decreased hippocampal volume and abnormal increased signal intensity on MRI T2 and flair images. Can be unilateral or bilateral.

b. Three main features
    i. history of febrile status in the first year of life (40-50% of patients with MTS)
    ii. partial onset seizures without fever occurring later in adolescence or young adulthood
    iii. inadequate response to AEDs
c. Clinical Characteristics
    i. Brief auras of fear, smell, rising epigastic feeling, or déjà vu followed by behavioral arrest.
    ii. Often have autonomic symptoms and oral or upper extremity automatisms.
    iii. Can have head deviation and upper extremity dystonic posturing.
d. These patients are good surgical candidates if MRI shows MTS and they have failed 2 or more medications as defined by the presence of 1 or more seizures per month.
    i. Surgical evaluation includes
        1. fMRI and neuropsychilogical testing to determine if language or memory would be impaired with surgery.
        2. Video EEG monitoring, usually with coming off medications, to show a clear and consistent seizure focus. This can be difficult because mesial structures are very deep.

Epileptic encephalopathy : Describes when epileptic activity itself is thought to contribute to severe cognitive and behavioral impairments above and beyond what might be expected from the underlying pathology alone and that these can worsen over time. These impairments may be global or more selective and they may occur along a spectrum of severity.

5. Infantile Spasms
    a. Considered an epileptic encephalopathy
    b. Onset in first year of life (most 3-7months)
    c. West syndrome is the Triad of infantile spasms, hypsarrhythmia and psychomotor retardation with or without developmental regression in a previously normal infant who has normal structural brain imaging and absence of genetic or metabolic identified etiologies.
    d. Clinical characteristics
        i. Sudden but briefly sustained flexion, extension or mixed flexion-extension of the arms, legs and trunk
        ii. Each spasms lasts up to 10 seconds but usually 1-2 seconds
        iii. 90% occur in clusters.
        iv. Most occur when aroundsleep/wake cycles
    e. EEG: hypsarrhythmia – interictal high amplitude waves and a background of irregular spikes
    f. Etiology
        i. Genetic: Downs syndrome, Aicardi syndrome, Tuberous Sclerosis

ii. Structural/metabolic: hypoxic ischemic encephalopathy; brain malformations; infections, including TORCH infections and encephalitis with herpes simplex virus, metabolic disorders
iii. Unknown cause: it may have a fundamental genetic defect at its core or it may be the consequence of a separate as yet unrecognized disorder.

g. Prognosis
   i. Best for unknown cases that are treated early
   ii. Spasms almost always resolve over time but development of other seizure types is common as the child grows older (15-20% develop LGS)
   iii. Very high rate of mental retardation (reported up to 80%) and/or epilepsy (reported up to 60%)

h. Treatment:
   i. ACTH: high dose for 2weeks followed by taper. Remains gold standard
   ii. Prednisone: insufficient evidence to be considered 1st line at this time
   iii. Vigabatrin: treatment of choice for children with tuberous sclerosis, requires very strict optho monitoring for retinol toxicity leading to constriction of peripheral visual fields. Has been reported in 16-44% of testable patients. Most likely dose and treatment length dependent.

6. Lennox-Gastaut Syndrome (LGS)
   a. Onset 3-10yr; peak 3-5yrs
   b. Clinical characteristics
      a. multiple types of generalized seizures including tonic, atypical absence, and drop attacks.
      b. developmental delay and intellectual disability
   c. EEG: 2.5Hz diffuse slow spike and slow waves
   d. Treatment: Depakote, lamotrigine, topirimate, clobazam, rufinamide, felbamate

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Anti-Epileptic Medications
Resident lecture series

Risk of seizure recurrence:

- After the first unprovoked seizure: 30-50% by 2years if idiopathic
  - Higher rates of recurrence (up to 65%) in patients with:
    - abnormal EEGs: spike and sharp waves are more predictive of recurrence then focal or generalized slowing.
    - prior static brain injury: cerebral malformation, trauma, prior CNS infection, prior stroke
    - developmental delays
- After 2 seizures: 70-80%

Any danger in waiting for the 2nd seizure?

- AEDs reduces risk of seizure recurrence by 30-60%
- Likelihood of seizure freedom at 3-5 yrs similar whether AED is started after the first or second seizure

►It is reasonable to wait until the 2nd seizure in most instances. The decision to treat should be based on the number of seizures the patient has had and not on the hope of preventing the development of "chronic epilepsy" by treating early

Focal or secondarily generalized epilepsy : (Carbamazpine), oxcarbazepine, phenytoin
2nd line: lacoasmide, topiramate, Valproate, leviteracetam

Generalized epilepsy

1. **Tonic-clonic** Valproate, lamotrigine, topiramate
2. **Absence:** Ethosuximide, valproate, lamotrigine
3. **Juvenile Myoclonic Epilepsy:** Valproate, lamotrigine, topiramate
4. **Lennox-Gestaut syndrome :** Valproate, lamotrigine, topiramate
5. **Infantile spasm**: ACTH, prednisone, vigabatrin (*TS)

2nd line: zonisamide, banzel, felbamate, keppra

Valproate: Depakote

- Very broad spectrum
- SE:
  - Bad for teenage girls: weight gain, alopecia, teratogenic (1-2% spina bifida)
  - tremor (dose related)
  - hepatotoxic (1:500 in children <2yrs)
  - pancreatitis
  - thrombocytopenia, leukopenia (low ANC)
- Enzyme INHIBITOR: Increases levels of other drugs
- Other uses: Mood stabilizer, Headaches
- IV and PO (sprinkles are better than liquid)

Lamotrigine: lamictal

- Very broad spectrum
- SE: Severe Rash (STEVENS-JOHNSON), increased risk if given with enzyme inhibitors including valproic acid, drowsiness

- To decrease risk of rash need to start low and titrate up slowly (takes about 6 weeks to get to affective starting dose)

Topiramate: Topomax

- Broad spectrum; useful in generalized and multifocal epilepsy
- Also used for headache prophylaxis
- SE:
    - cognitive slowing ("dopomax")
    - decreased appetite (weight loss)
    - 1-2% can get kidney stones because renal excreted
    - Metabolic acidosis : carbonic anhydrase inhibitor (caution with Keto diet and some patients experience decreased sweating)

Levetiracetam: Keppra

- Broad spectrum
- SE:
    - behavior changes and agitation; little evidence but can use B6 supplementation to help alleviate these effects
- Renal excretion
- No drug interactions
- PO and IV

Oxcarbazepine: Trileptal

- Focal seizures
- Less CNS effects then carbamazepine and fewer drug interactions
- SE:
    - Hyponatremia
    - SIADH (not dose dependent and very rare)
    - dizziness, somnolence, and ataxia: these are dose dependent and most common when dose is increased too fast

Phenytoin: Dilantin

- Best for Partial seizures but used for STATUS EPILEPTICUS
- SE:
    - Also bad for teenage girls when used long term: Hirsutism, gum hypertrophy, Teratogenic (craniofacial anomalies),
    - Ataxia
    - nystagmus
    - stevens-johnson
    - Anxiety and aggression
    - Soft tissue injury if IV form infiltrates **
    - cardiac arrhythmias**

      **why we use fosphenytoin
- Enzyme INDUCER: decreases the levels of other drugs
- Lots of drug to drug interactions
- A small increase in dose may lead to a large increase in drug due to zero order kinetics

Phenobarbitol:

- Broad spectrum but used for STATUS EPILEPTICUS
- First line for neonatal seizure
- SE:
    - Hyperactivity
    - Irritability
    - Lethargy
    - Hypotension
    - Long term effects on learning and cognition
- Enzyme INDUCER: decreases the levels of other drugs
- Lots of drug to drug interactions
- Very long t1/2

Ethosuximide

- Use ONLY for Absence
- Nausea, headache

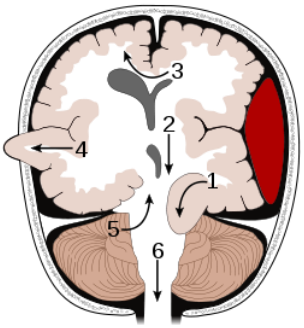
Increased Intracranial Pressure (ICP)

Resident lecture series

- **Etiologies of raised ICP**
  - The skull contains: brain, blood and CSF
  - Additional brain volume: cerebral edema, tumor
  - Disturbance of intracranial blood flow: intracranial hemorrhage, CSVT
  - Impairment of CSF production or absorption: obstructive/communicating hydrocephalus, CSF producing tumor

- **Signs and symptoms of increased ICP**
  - Headache (esp. if worse in am or worse with lying down)
  - Behavioral change/irritability
  - Depressed mental status
  - Nausea/vomiting
  - Diplopia/ visual changes
  - Papilledema (ICP must be elevated for > 1 day before papilledema develops)
  - Cushing triad (bradycardia, hypertension, irregular respirations)-does not occur without depressed mental status and is a late finding
  - Initial signs may be focal or diffuse depending etiology (e.g. CSVT vs mass)

- **Herniation syndromes** (Final common pathway)
  - Uncal herniation (1) (lateral transtentorial)
    - Mechanism: herniation of medial temporal lobe past edge of tentorium and down, causes compression of CN III and contralateral cerebral peduncle
    - Signs/symptoms: ipsilateral weakness, ipsilateral CN III palsy ("blown" pupil)
  - Tonsillar herniation (6)
    - Mechanism: mass effect from above leads to downward shift of pontomedullary junction through foramen magnum causing compression of cerebellar tonsils and medulla
    - Signs/symptoms: dysregulation of respiratory and cardiovascular systems
  - Subfalcine herniation (3)
    - Mechanism: mass effect causing the cingulated gyrus against or under the falx
    - Signs/symptoms: personality changes, contralateral leg weakness
  - Upward transtentorial herniation (5)
    - Mechanism: mass effect in the posterior fossa displacing the midbrain upward causing compression of the cerebral aqueduct/4^{th} ventricle
    - Signs/symptoms: diffuse signs of obstructive hydrocephalus, homonymous hemianopsia (PCA infarct)



- **Management of increased ICP**
  - Impending herniation
    - Call code and neurosurgery
    - Raise head of the bed to 30 degrees
    - Hyperventilate patient (temporizing measure only, causes vasoconstriction and therefore decreased cerebral blood flow)
    - Send patient emergently to head CT
  - Acute increased ICP (including slowly progressive elevations of ICP)
    - No LP – may cause herniation leading to death
    - Cerebral edema- hyperosmolar therapies
    - Mass lesion – surgery, corticosteroids
    - Hydrocephalus - EVD/VP shunt, treat obstructive lesion
    - ICH – identify and treat source of hemorrhage (e.g. AVM)
    - Arterial infarcts and venous thromboses – identify and treat vascular compromise (e.g. arterial dissection, CSVT)
    - Decompressive craniectomy – removal of a portion of the skull to accommodate increased pressure

Tics and Tourette Syndrome

Resident Lecture Series

Epidemiology

- Tics are a very common movement disorder seen in 2 – 15% of school aged children, with a higher incidence in males.
- Tourette Syndrome is seen in approximately 4 per 10,000

Clinical Aspects

- The three defining characteristics of tics include their being repetitive, stereotyped, and intermittent.
- Tics may be motor, such as eye blinking, or vocal, such as grunting. Tics may are classified as simple if they involve one movement in one part of the body, such as eye blinking or grunting, or complex if they involved multiple body parts and have a sequence of events, such as slapping oneself or making complex vocal noises.
- Having both motor and vocal tics for a duration of one year, with no more than 3 months without tics during that time, defines Tourette Syndrome.
- Most tics are not rhythmic, involve the head and upper body, and are temporarily suppressible by the patient, however "build up" inside and may result in a rebound immediately after.
- Tics are usually variable over time, with periods of diminished and more frequent occurrences. Emotional or cognitive stress often exacerbates tics, and concentration may either worsen or improve tics, depending on the patient.
- Most tics have their onset by 6 years of age and peak around 11 years, all have onset prior to 18yo
- Approximately 3% of children in the population have chronic tics, defined as lasting more than one year, and about half of those with chronic tics will have resolution by 18 years of age
- Often more disabling than tics are the common comorbities seen in 50-75% of these patients, including obsessive-compulsive disorder (OCD), anxiety, and attention deficit hyperactivity syndrome (ADHD).

Pathogenesis

- The pathophysiologic basis for tics is unknown, but thought to involve the basal ganglia and movement circuits of the brain, as related to dopamine transmission. This is further supported by the therapeutic response to dopamine altering agents, such as the neuroleptics.

Evaluation

- Most children with tics and Tourette Syndrome do not require extensive workups.
- A child presenting with a typical clinical scenario, a normal neurologic exam, and the ability to temporarily suppress the tics often will not require any further testing.
- Tics may become apparent due to medications including stimulants and neuroleptics, although these likely unmask a predisposition to these disorders and are not thought to cause them.
    - NIH Treatment Trial ADHD and Tics Neurology - Volume 58, Issue 4 (February 2002)
        - The proportion of individual subjects reporting a worsening of tics as an adverse effect was no higher in those treated with methylphenidate

(20%) than those being administered clonidine alone (26%) or placebo (22%).

- Other more rare disorders that may results in tics and other neurologic abnormalities, such as dystonia or encephalopathy, include pantothenate kinase associated neuodegeneration (PKAN), Wilson disease, neuroacanthocytosis, Huntington disease, and stroke.

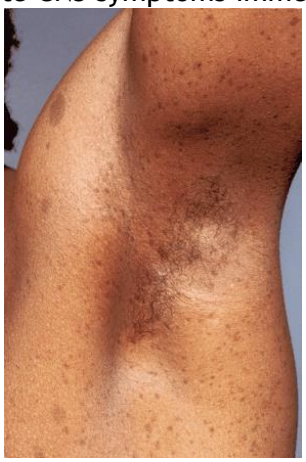
Treatment

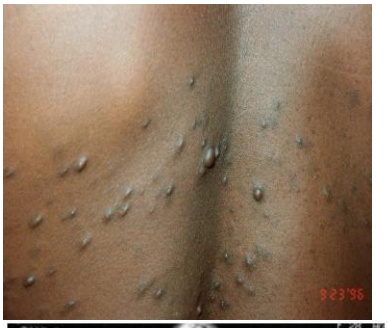
- Treatment should be tailored to the most disabling symptom, whether it be the tic, OCD, or ADHD.
- Tics do not require treatment unless they are interfering with the patient's ability to learn at school, perform activities of daily living, are disruptive to others in the classroom setting, or cause pain.
- Common medications used to treat tics include alpha-2 adrenergic agonists, with guanfacine and clonidine as first line. Other agents used include antiepileptics and neuroleptics.
- www.wemove.org

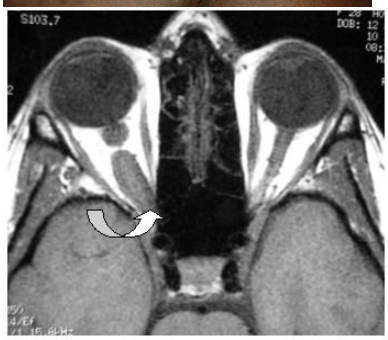
Neurocutaneous Syndromes
Resident Lecture Series

1. Neurofibromatosis Type 1 and 2
   1 NF Type 1 is autosomal dominant, progressive disease characterized by 2 of following:
      - six of more cafe au lait >5mm if prepubertal, 15mm if postpubertal,
      - axillary or inguinal freckling,
      - 2 or more iris lisch nodules (hamartomas seen with slit lamp, rare in children <3yo but in 100% of >21yo),
      - 2 or more neurofibromas or 1 plexiform neurofibroma typically in skin with small rubbery purple color,
      - distinctive osseous lesion such as sphenoid dysplasia (scoliosis common but does not count),
      - optic glioma (in 15% of pts),
      - 1st degree relative with NF
   2 NF 2 is classified as follows:
      - Confirmed (definite) NF2: Bilateral vestibular schwannomas
      - Presumptive (probable) NF2: Family history of NF2 (first degree family relative) plus Unilateral vestibular schwannoma or any two of the following: meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity, juvenile cortical cataract
   3 Symptoms
      - Typically present with learning problems and neurocutaneous stigmata
      - susceptible to neuro complications including moyamoya with TIA/CVA, generalized tonic clonic or complex partial seizure low grade gliomas and hamartoma's; malignancy including pheochromocytoma, myeloid leukemia, wilms, rhabdomyosarcoma's; neurocognitive deficiencies including learning disabilities (30-45% with specific learning disability), speech deficits, ADHD
      - HTN, precocious puberty
   4 Pathogenesis
      - due to defect in NF1 gene which codes for neurofibromin protein, a tumor suppressor gene
      - also in that intron is oligodendrocyte myelin a gene which results in abnormal myelination of the cortex seen as white matter abnormalities on MRI
      - NF 2 is typically due to mutation at center of long arm of 22q11
   5 Management
      - Multidisciplinary approach including neurology, PM+R, ophthalmology, nephrology, oncology
      - Should have yearly history and physical exam and ophtho exam

- Investigation of any new or acute CNS symptoms immediately with MRI/CT



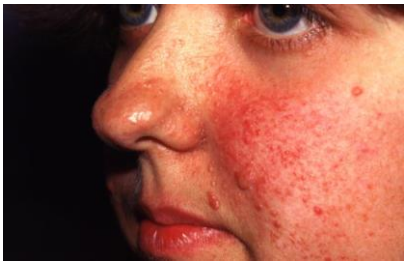
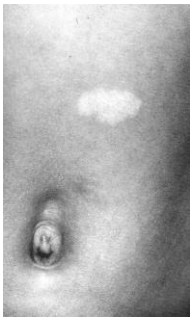
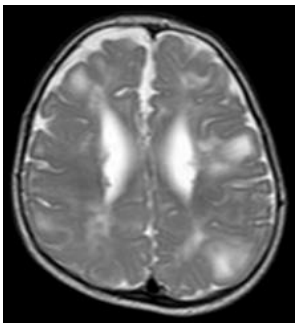




2. Tuberous Sclerosis

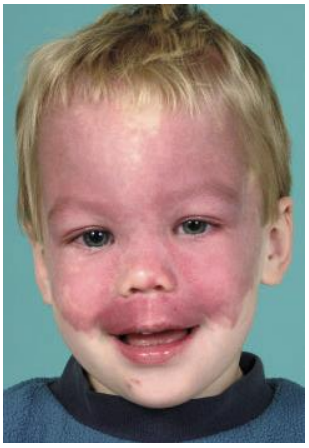
- Autosomal dominant multisystem neurocutaneous disorder resulting in hamartomas in skin, CNS, kidneys, eyes, heart, and lung affecting 1 in 10,000
- auto dominant with 70% due to sporadic mutations
- 2 genes: TS1 on 9q34 with hamartin protein affected, TS2 on 16p13 with tuberin affected
- The classic TSC central nervous system findings of cortical tubers, subependymal nodules (SENs), subependymal giant-cell astrocytomas (SEGAs), and white-matter abnormalities
- cortical tubers, so called for their gross pathologic "potato-like" appearance in 95% of patients
- Subependymal nodules are also seen in the majority of TSC patients. They can be identified early in infancy and increase in number through the first decade of life. SENs are most commonly found around the area of the foramen of Monro.
- SEGAs are benign tumors that occur in approximately 10% of patients with TSC.
- They may develop relatively quickly in patients without evidence of SENs on prior imaging but are unlikely to enlarge in patients over 21 years of age.
- Seizures are the presenting symptom in 92% of individuals with TSC, making epilepsy a significant cause of morbidity associated with this disease. Seizure onset usually occurs during infancy or early childhood but may happen at any age.

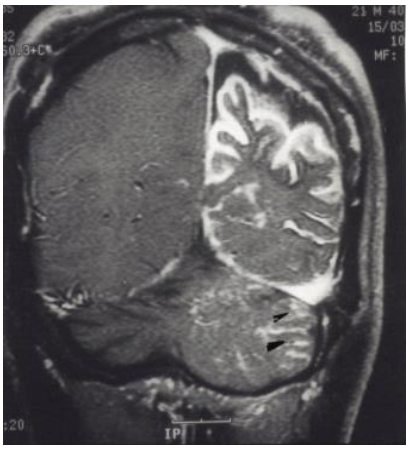
- Infantile spasms develop in approximately one third of TSC patients and may be the initial symptom in almost 70% of affected infants that come to medical attention.
- vigabatrin appears to be more effective than adrenocorticotropic hormone (ACTH) in managing infantile spasms in children with TSC.
- Early population-based studies found cognitive abnormalities in 50% to 65% of patients
- Hypomelanotic macules or ash leaf spots occur in 61% to 97.2% of individuals with TSC. They are often present at birth, and they may become more numerous with time
- Facial angiofibromas (adenoma sebaceum) are pathognomonic for TSC and are seen in over 70% of patients. They first appear during the preschool years and with time develop into discrete, papular, pink, or erythematous lesions in a symmetrical malar distribution on the face.
- The incidence of shagreen patches varies depending on the age of the patient population studied but occur in approximately 50% of patients. They may be noted in early childhood.
- The renal manifestations of TSC are found in a majority of patients. They include angiomyolipomas (AMLs), simple cysts, polycystic kidney disease, and renal-cell carcinoma. These lesions likely arise in infancy or early childhood, increasing in size and number with age.
- Cardiac rhabdomyomas are the most common intracardiac tumor of infancy and childhood and are highly associated with TSC. They are found in 30% to 50% of affected individuals. Rhabdomyomas are an early manifestation of the disease, appearing at 22 to 28 weeks of gestation. 80% pts with rhabdomyosarcomas haveTS.
- LAM occurs almost exclusively in young women, typically presenting between 30 to 35 years of age.
- Hamartomas are the most common retinal manifestation of TSC and are identified in approximately 40 to 50% of individuals.

3. Sturge Weber
    - Characterized by the combination of a facial port wine nevus, contralateral hemiparesis, hemiatrophy of the brain, mental retardation, and homonymous hemianopsia
    - caused by a sporadic mutation with resultant leptomeningeal angiomatosis and resultant localized ischemia
    - Port wine stain on face at birth (V1 distribution most common) with possible glaucoma ipsilaterally, brain atrophy ipsilaterally
    - Intracranial Ca on CT used for diagnosis (tram track calcifications)
    - 80-95% develop seizures on contralateral side to nevus, intractable in many, resulting in hemispherectomy
    - 50% with MR

- Diagnosed by skull radiographs with Ca, CT with unilat atrophy and ipsilat ventricle dilation, MRI with letomeningeal angiomas





Hypotonia

Resident Lecture Series

Background

- Typically manifest as: (1) unusual resting posture; (2) diminished resistance of the joints to passive movement; (3) increase range of joint movement
- typically categorized by etiology as central (UMN) or peripheral (LMN)
- usually evident from birth although acquired causes are possible and more concerning

History

- Important to obtain accurate gestational history asking about fetal movements, polyhydramnios, fetal position, exposure to teratogens, maternal disease, history of NM disorders
- Birth history itself critical to look for suggestions of fetal distress and possible hypoxic injury
- Hospital course after delivery, including need for ventilator, feeding difficulties, seizure, apnea, contractures on exam

Causes

- Localize to one of following based on exam: CNS->spinal cord-> anterior horn cells-> Nerve-> NM junction-> muscle
- Hypoxic ischemic encephalopathy most common cause overall
- Other causes:
    - CNS/spinal cord: stroke, bleed, spinal cord transaction, genetic causes (Prader Willi, Angelman, Downs), metabolic (fatty acid oxidation, organic acidurias)
    - Anterior Horn Cell: SMA, Poliomyelitis
    - Nerve: AIDP, MLD
    - NM Junction: Botulism, Myasthenia Gravis
    - Muscle: Storage disorders (Lipid and Glycogen Storage Disorders), Myotonic Dystrophy, Mitochondrial Disorders, Muscular Dystrophy (Congenital, Inflammatory), Connective Tissue Disorders

Exam Findings

- Most hypotonic infants demonstrate a characteristic posture of full abduction and external rotation of the legs as well as a flaccid extension of the arms.
- When traction is delivered to the arms, there is a prominent head lag.
- The presence of profound weakness as well as hypotonia suggests a disorder of LMN.
- Weakness can be easily detected in the presence of a low-pitched cry/progressively weaker cry, readily distinguished from the vigorous cry of a normal infant.
- There is a paucity of antigravity movements in the weak and hypotonic infant.
- Infants with neuromuscular disease are visually quite alert in comparison to those with central nervous system involvement where obtundation and depressed level of consciousness is often present.
- The relative preservation of muscle power with hypotonia and hyperreflexia favors a central origin to the hypotonia, while the combination of weakness in the antigravity limb muscles and hypo/areflexia together favor a neuromuscular disorder.

Localization

- UMN: spastic, hyperreflexia, normal sensation
- Anterior Horn Cell: weakness, areflexia/hyporeflexia, normal sensation
- Nerve: weakness, atrophy, hyporeflexia, sensation may be normal or abnormal
- NM junction: weakness, DTR +/-, normal sensation
- Muscle: weakness, DTR +/-, sensation normal, CPK normal or high, possible arthrogryposis

Diagnosis

- If suspect central hypotonia: karyotype/DNA microarray and neuroimaging; specific genetic testing based on clinical suspicion
- If suspect peripheral hypotonia: EMG/NCS, CK; muscle biopsy and specific genetic testing based on clinical suspicion
- Other tests may include metabolic testing (lactate, pyruvate, ammonia, acylcarnitine, free fatty acids, serum amino acids, urine organic acids); stool botulinum toxin; transferrin isoelectric focusing (CGD); VLCFA (peroxisomal disorders); SMN testing (SMA)

Resources

- http://neuromuscular.wustl.edu/index.html
- www.genetests.org

DUCHENNE AND BECKER MUSCULAR DYSTROPHY

Resident Lecture Series

Background

- Muscular dystrophies are a group of inherited disorders that result in the progressive degeneration of muscle fibers.
- Duchenne Muscular Dystrophy (DMD) is the most common form of muscular dystrophy, affecting 1 in 3300 live male births. It is inherited in an X-linked recessive manner resulting in destabilization of the muscle membranes due to lack of dystrophin.
- Becker Muscular Dystrophy is inherited in a similar fashion but is less severe and less common, affecting 1-18,000 live male births.

Clinical Aspects

- DMD typically present to a physician between the ages of 2 and 5 years due to an inability to climb stairs, run, and jump.
- At presentation, the patient's gait is often described as waddling, and calf pseudo-hypertrophy may be prominent, due to fibrofatty deposition within these muscles.
- The pattern of weakness is proximal and symmetric.
- When asked to get up from a seated position on the floor, patients with DMD perform a Gower maneuver, which is described as the need to use the arms to support oneself into the upright position, often times into a tripod position on the ground or legs.
- Cardiac muscles are similarly affected, often resulting in a cardiomyopathy.
- IQ is below normal in most with DMD, but cognitive decline is not seen.
- Patients will become wheel chair dependent by 13 years in DMD.
- Patients with DMD typically survive until a mean age of 20 years, with causes of death including cardiac failure and respiratory insufficiency.
- BMD is similar to DMD but is less severe, with later age at onset of clinical symptoms, typically around 12 years, although onset as late as the fourth decade has been described.
- Patients with BMD are not wheelchair bound until after 16 years, and the age at death is typically for reasons similar to DMD, and between 30 and 60 years.

Pathogenesis

- DMD and BMD are due to deficient muscle membrane protein, dystrophin. Dystrophin resides in the plasma membrane of muscle tissue and acts to stabilize the glycoprotein complex. Without dystrophin to stabilize the muscle membrane, the glycoprotein complexes degrade and result in muscle destruction, eventually replaced by connective tissue and fat, i.e. fibrofatty replacement.

Evaluation

- The diagnosis of DMD and BMD is primarily one based on clinical grounds.
- The primary marker of muscle destruction is creatine kinase, or CK, which is elevated in the serum over ten times normal in DMD, typically in the range of 5,000 – 40,000 IU/L, and at least five time normal in BMD.
- EMG may be used to identify the myopathic process that is occurring, with early signs of damage evidenced by positive waves on insertion during the needle exam, the development of fibrillations, and rapid low amplitude recruitment to compensate for loss of muscle fibers. As fibrofatty replacement occurs, insertional activity will

become reduced, as may compound muscle action potentials on nerve conduction. Sensory nerve action potentials are typically normal

- Muscle biopsy is typically deferred due to its invasiveness and the availability of accurate genetic testing. In DMD, muscle fibers display absent dystrophin staining, and in BMD, staining is typically normal or reduced. Fibrofatty replacement is obvious within hypertrophied muscle fibers, and central nuclei are also present.
- When clinically suspected in a patient with elevated CK, genetic testing can be sent for analysis by PCR, sequencing, or Southern blot assay. In patients with a family history, genetic testing may not be necessary in the scenario of a typical presentation and existing family member with genetic confirmation.

Treatment

- Treatment for DMD and BMD include a multidisciplinary approach, with neurology, physical medicine and rehabilitation, pulmonary, psychiatry, and cardiology involved in the coordination of care. Medications, assistive devices, and physical and psychological therapies are all utilized
- Steroids are the primary drug used to slow the progression of disease in DMD. AAN Practice Parameter *Neurology* 2005;64;13-20
    - Prednisone has been demonstrated to have a beneficial effect on muscle strength and functionin boys with DD and should be offered (at a dose of 0.75 mg/kg/day) as treatment (Level A).
    - Benefits and side effects of corticosteroid therapy need to be monitored. Timed function tests, pulmonary function tests, and age at loss of independent ambulation are useful to assess benefits.
    - Deflazacort (0.9 mg/kg/day) can also be used for the treatment of DD in countries in which it is available (Level A). Patients should be monitored for asymptomatic cataracts as well as weight gain during treatment with deflazacort.
    - Current evidence suggests that therapy with steroids should begin around the age of 5 years and patients be closely monitored for side effects.
- Around the age of 10 years, annual surveillance for cardiomyopathy, pulmonary function testing, and monitoring for orthopedic complications, such as scoliosis, should be performed by the appropriate specialists.
- Avoidance of bony mineral loss should be addressed with vitamin D therapy.

Neurology Outpatient Clinic Schedule

Clinic Schedule:

The Neurology clinics are located at 1300 Main Hospital (1st Floor between the main lobby welcome desk and ER). Clinics generally start at 830am and end by 5pm except as noted below. The following schedule is based on calendar month and should be verified prior to attending due to day to day variability.

Weeks 1, 3, 5									
MONDAY		TUESDAY		WEDNESDAY		THURSDAY		FRIDAY	
AM	PM	AM	PM	AM	PM	AM	PM	AM	PM
Epilepsy	Resident/ General Child Neuro Clinic	Multidiscipl NF Clinic 7:45-1230 genetics dept EMG	NF Clinic 1-3 NM Clinic ICU Neuro-Ophth	Sickle Cell/CVA Headache	Sickle Cell/CVA Resident/ General Child Neuro Clinic	Movement disorders Neuro-oncology 4 East Neuro-Sleep MDA clinic	Movement disorders MDA clinic Neonatal Injury	Myelin disorders Neuro-Ophth	Myelin disorders
Weeks 2 and 4									
AM	PM	AM	PM	AM	PM	AM	PM	AM	PM
Epilepsy	Resident/ General Child Neuro Clinic Perinatal brain injury	EMG	NF Clinic 1-3 NM Clinic ICU Neuro-Ophth	Headache Stroke	Resident/ General Child Neuro Clinic	Movement disorders Neuro-oncology 4 East Neuro-Sleep	Movement disorders Ketogenic Diet	Neuro-genetics Neuro-Ophth	Neuro-genetics

PEDIATRIC NEUROLOGY: DEPARTMENTAL CONFERENCES
REQUIRED CONFERENCES IN BOLD UNDERLINE (>70% ATTENDANCE)

Monday	Tuesday	Wednesday	Thursday	Friday
830am Pediatric Resident Lecture	830am Pediatric Resident Lecture 12pm EEG Lecture	8am Grand Rounds	7AM Sunrise Lecture 8AM Epilepsy Surgery 12PM Fellow Meeting 1PM Ward Intake 330 PM Tumor Board	12PM Localization/Board Review
830am Pediatric Resident Lecture	830am Pediatric Resident Lecture 12pm EEG Lecture	8am Grand Rounds	7AM Sunrise Lecture 8AM Epilepsy Surgery 12PM Clinical NS Lecture 1PM Formal Intake 2PM Journal Club 3PM Staff Meeting	12PM Localization/Board Review
830am Pediatric Resident Lecture 12 pm PICU/NCC Conference (BearMedia)	830am Pediatric Resident Lecture 12pm EEG Lecture	8am Grand Rounds 4pm Stroke Conference (Rads Purple)	7AM Sunrise Lecture 8AM Epilepsy Surgery 12PM NS Lecture 1PM PICU Intake 330 PM Tumor Board	12PM Localization/Board Review
830am Pediatric Resident Lecture	830am Pediatric Resident Lecture 12pm EEG Lecture	8am Grand Rounds	7AM Sunrise Lecture 8AM Epilepsy Surgery 12PM IDDRC Lecture 1PM NICU Intake	12PM Localization/Board Review
830am Pediatric Resident Lecture	830am Pediatric Resident Lecture 12pm EEG Lecture	8am Grand Rounds	7AM Sunrise Lecture 8AM Epilepsy Surgery 12PM IDDRC Lecture 1PM NICU Intake	No lecture

Mandatory Lectures

- **70% attendance for all fellows via MedHub evaluation form**
- **RED = Mandatory on all services and electives**
- **BLUE = Mandatory on PICU rotation**

Pathways and Protocols

Children's National Medical Center
Division of Neurosciences
Pathways and Protocols

General Information

Seizure management plan

Dr Gaillard – May 2014

If new onset seizure or epilepsy:

- Admit and observe.
- PRN ativan (0.025-0.05 mg/kg/dose) that could be repeated in 8-12 hours if subsequent or prolonged seizures.
- If seizures after 0.05 mg/kg of Ativan, load with fosphenytoin and start fosphenytoin maintenance until complete evaluation and arrive at long term Rx.
- If wish to avoid sedation with ativan, load with fosphenytoin (there are alternatives— LVT, LCM).

If exacerbation in seizures:

- If PO meds available in IV form then IV load and adjust PO meds
- If not… PO adjustment and use of ativan (0.025-0.05 mg/kg/dose) that could be repeated in 8-12 hours if continued seizures more than baseline.
- If continued seizures more than baseline after 0.05 mg/kg of Ativan, IV load fosphenytoin/other seizure medications (as appropriate for epilepsy syndrome, e.g. VPA if an only if primary generalized seizures and labs fine)

If ill and vomiting:

- ativan (0.025-0.05 mg/kg/dose) q 8-12 hours until taking PO (max 2 days).
- If persistent seizures following 0.05 mg/kg of ativan, then load with IV meds eg. fosphenytoin (or others as appropriate for epilepsy syndrome)

Criteria for direct admission to neurology floor from outside ED

New onset afebrile seizure

Children to be directly admitted to the floor

- Child is > 6 months and
- Seizure was < 10 min and
- Child has returned to baseline function

Children who should be evaluated in the ED

- Seizure was ≥ 10 min and/or pt has had multiple seizures in the last 24 hours or
- Child's MS is not at baseline or
- Child is ≤ 6 months or
- Any child who has a seizure during transport

Atypical febrile seizure:
Evaluation for meningitis should be considered either by clinical exam or by LP at outside hospital

Children to be directly admitted to the floor

- Child is > 12 months and
- If LP was done, CSF shows < 5 WBC and normal protein and
- Child's immune function is presumed to be normal and
- Child has returned to baseline function

Children who should be evaluated in the ED

- Child is ≤ 12 months or
- Child's MS is not at baseline or
- CSF results are unknown or
- LP is indicated but was unsuccessful at the outside ED or
- Any child who has a seizure during transport

Seizure with fever in patient <6months (by definition not a febrile seizure)

- Under all circumstances must be evaluated in the CNMC ED before admission to the Neurology floor

Recurrent or escalating seizures in known seizure patient

Children to be directly admitted to the floor

- Child has returned to baseline function
- Child has not returned to baseline b/c they received sedating medication but opens eyes to voice and there is no evidence of ongoing subclinical status (e.g. nystagmus, fixed gaze, etc)
- Patient has been seizure free for > 1 hour

Children who should be evaluated in the ED

- Child's MS is not at baseline
- Any child who has a seizure during transport

- **Transfer is not dependent on there being a HCT or MRI but if imaging is indicated on a urgent basis transfer should not occur until imaging is obtained**
    - **Example 1: Child w/ a focal seizure who has returned to baseline needs imaging but not necessarily within hours of admission**
    - **Example 2: Child w/ a focal seizure and a residual Todd's paralysis needs imaging to exclude other causes of the paralysis (ICH, AIS, etc). Transfer would occur only after imaging is obtained and reviewed.**
- **Patients in need of an LP will not be directly admitted until LP is performed**
- **Children < 4 years of age who have had > 1 seizure in the last 24 hours must have a working IV**
- **Direct admission will NOT occur if admitting neurologist is uncomfortable or unclear about the information our staff received from the referring facility. The outside facility may be called directly to clarify.**

MELAS (acute management—Please discuss if long QT or cardiomyopathy with Dr. Gropman or genetic doctor on call

Stroke-like episode

1. MRI to look for lesion (MRA and MRS at clinician discretion).
2. L-Arginine 0.5 g/kg bolus prepare in 10% solution and run at 50 mL/hour until completed
    a. (Max is 20 grams; so if >40kg give 20 gram bolus)
    b. Central line is preferred, if peripheral is used in emergency, must be observed closely since significantly caustic if infiltrates
    c. Can cause hypotension, so fluid bolus may be necessary
3. Start D10 NS +20 mEq/L potassium chloride (Or 20mEq/L potassium acetate) at 1.5 maintenance
4. Vital signs should be at least every 15 minutes while Arginine is infused or according to unit protocol if more frequent.
5. Laboratory: blood glucose every 30 minutes during arginine infusion.
6. Levocarnitine at home dose, but given IV (eg. Home 990 mg every three time daily, becomes 990 mg IV every 8 hours)—(-no IV carnitine if known cardiomyopathy or long QT
7. B50 Complex (banana bag if not tolerating enteral):
    a. To order in SMS: B complex: one soft gel (daily if <20 kg), or (BID if >20 kg) (Nonformulary pharmacy order contains pyridoxine, pantothenate, riboflavin, niacin, thiamin)

MELAS related Seizures

Use the Children's National acute seizure pathway --- usually very responsive (most on Keppra + 2nd medication for their baseline seizures

Maintenance Management for MELAS

1. Resume home medications at those doses (otherwise use below) if possible
2. L-arginine by mouth at 0.3-0.4 grams/kg/day (divided 2-3 times): Goal is plasma amino acid of arginine about 150 mcmol/L as soon as possible.
3. If cannot give oral, give 0.5 grams/kg arginine infused over 24 hours.
4. L-carnitine 50-100 mg/kg/day
    a. B50 Complex (to order in SMS: B complex: one soft gel (daily if <20 kg), or (BID if >20 kg) (Nonformulary pharmacy order contains pyridoxine, pantothenate, riboflavin, niacin, thiamin)
5. Vital signs: blood pressure at least every 4 hours.
6. Vitamin C: 25 mg/kg/day

7. Ubiquinone or CoQ10 (often home med): 5 mg/kg/day (<20 kg), 10 mg/kg/day (>20 kg)
8. Vitamin E 22.5 IU/kg/day

Updated: 6-22-2016

Ketogenic Diet Information

The Ketogenic Diet

Important Details for surgical procedures/other procedures that require anesthesia

What is it?

A high-fat, low-protein, and very-low-carbohydrate diet used to treat epilepsy which has not responded to antiseizure medications. Its benefit is tied to the production of ketones. It is also used as primary treatment for some inborn errors of metabolism.

General Precautions

- The amount of carbohydrate intake per day has been carefully calculated and measured, to reach a specific ratio of fat to [carbs plus protein], e.g. 2:1, 3:1, 3.5:1, 4:1. An increase in carbohydrates may eliminate ketosis, putting the child at risk for seizures/status epilepticus.
- Approach the ketogenic diet therapy as you would a **medication**!
- Prepare IV medications in **normal saline** (not D5W) if safe and compatible
- If medications MUST be in D5W, our nutritionists should adjust the carbohydrate content of the diet to compensate
- Do not change the formulation of home meds. In general, tablet forms have less carbohydrate than syrups or chewables; carb content of meds has been factored into calculations.
- There are reports of increased incidence of propofol infusion syndrome in ketogenic diet patients--**Avoid propofol**

NPO procedures

- For patients on a clear liquid diet, sugar-free and calorie-free clear liquids may be given. Up to 8-10 ounces of UNFLAVORED pedialyte may be given in a 24 hour period. Blood glucose should be monitored every 4 hours while on clear liquid diet.
  - If glucose is <40 and symptomatic
    - Give 15 ml apple juice with ice chips OR 3 oz. unflavored Pedialyte
    - Wait 15 mins and recheck until >40 mg/dL
    - Dextrose-free IVFs if needed
  - If glucose<25
    - Give 30 ml apple juice with ice chips OR 5 oz. unflavored Pedialyte
    - Wait 15 mins and recheck until >40 mg/dL
    - Dextrose-free IVFs if needed

- For patients that are NPO, only IV solutions that are DEXTROSE-FREE may be given. Monitor blood glucose every 2 hours while NPO.
  - If glucose is <40 and symptomatic
    - Give 40 ml D5% solution
    - Wait 15 mins and recheck until >40 mg/dL
  - If glucose<25
    - Give 75 ml D5% solution

- Wait 15 mins and recheck until >40 mg/dL

Laboratory norms

- **Glucose** is normally 55 to 75 mg%
- Goal **CO2** is >= 16 mmol/L
- Goal urine **ketones** is 80-160 mg/dL (3+-4+ on urine ketone sticks), or serum beta hydroxybutyrate 4-6 mmol/L or 40 to 60 mg/dL (but takes a week to come back)

Possible complications and what to do

- Constipation
  - Rectal suppositories, Fleet enema, Miralax okay to use (see www.charliefoundation.org for a list of no/low-carbohydrate meds)
- Vomiting
  - If related to KD, usually occurs early after initiation of the diet
  - Check for hypoglycemia, dehydration, excess ketosis, acidosis, constipation (in addition to general medical causes)
  - Parents should have "sick-days" instruction sheet
  - Sugar-free clear fluids as much needed or Pedialyte 8 to 10 oz/day max
  - IVF's without dextrose if needed
  - Resume meals at full fluid, half strength food
- Dehydration
  - Diet causes diuresis
  - Goal spec grav is <1.025
- Encephalopathy
  - Check for hypoglycemia, dehydration, excess ketosis, acidosis
  - Concern for inborn error of metabolism? Check lactate, NH3, urine organic acids, carnitine, acylcarnitine
- Hypoglycemia
  - If glucose is <40 and symptomatic
    - Give 15 ml apple juice with ice chips OR 3 oz. unflavored Pedialyte
    - Wait 15 mins and recheck until >40 mg/dL
    - Dextrose-free IVFs if needed
  - If glucose<25
    - Give 30 ml apple juice with ice chips OR 5 oz. unflavored Pedialyte
    - Wait 15 mins and recheck until >40 mg/dL
    - Dextrose-free IVFs if needed
- Excess ketosis
  - Symptoms include rapid panting, irritability, increased heart rate, facial flushing, unusual fatigue, and vomiting
  - Give 2 tablespoons of juice or 1 ounce of unflavored Pedialyte
  - If symptoms persist after 15-20 mins, give 2nd dose of juice/ Pedialyte
  - *Dextrose-free IVFs if needed ?*
- Metabolic acidosis

- If CO2 is 12-16
  - Assess hydration and hydrate if needed
  - If well-hydrated and symptomatic, give CytraK or decrease ratio
  - Check CO2 q6 to 8 hrs until stable or symptoms resolve
- If CO2 is <12
  - Hydrate if needed
  - CytraK 1-2 meq/kg/day
  - Eval for etiology (e.g. Inborn error of metabolism or renal tubular acidosis)

Less Common or Long-term Complications

- Hyperlipidemia
- Nutritional deficiencies (cardiomyopathy)
- Kidney stones
  - Risk factors include family history, carbonic anhydrase inhibitors, dehydration
  - IVFs, cytraK
- Osteoporosis
- Bruising (related to abnormalities in platelet function)

Resources

1) Dana Casendino, Dietitian 202 476 5985, pager 0831. On call dietitian pager 50335
2) On-call neurologist at CNMC (ask operator 202-476-5000 to page)
3) www.charliefoundation.org for list of carbohydrate-low or carb-free meds

Ketogenic Diet Patient Management Tips (also see 'Sick Days' Patient handout)

- **Vomiting**
  - Usually D 2-3
  - Check for hypoglycemia, dehydration, excess ketosis, acidosis, constipation

- **Encephalopathy**
  - Check for hypoglycemia, dehydration, excess ketosis, acidosis
  - If concern inborn error of metabolism-lactate, NH3, UOA, carnitine, acylcarnitine

- **Hypoglycemia**
  - **Glu will be low normal (55-75mg%)**
  - **If glu<40 and symptomatic**
    - give 15 ml apple juice with ice chips OR 3 oz unflavored Pedialyte
    - wait 15 minutes and recheck until >40 mg/dL
  - **If glu<25**
    - give 30 mL apple juice with ice chips OR 5 oz unflavored Pedialyte
    - wait 15 minutes and recheck until >40 mg/dL

- **Metabolic acidosis**
  - **Higher risk if IEM, Zonisamide, Topamax, Diamox, <3 yr**
  - **Goal CO2 >=16mmol/L**
  - **If 12-16**
  - Assess hydration

- If well-hydrated and symptomatic, start Bicitra (500 mg sodium citrate/334 mg citric acid=1 meq/ml bicarb) 1-2 meq/kg/day or Decrease ratio
- Check CO2 Q6-8 hrs until stable or symptoms resolve
- **If <12**
- If well-hydrated, start Bicitra 1-2 meq/kg/day
- Evaluate for etiology
    - If AG acidosis
        - Check betahydroxybutyrate (BHB), lactate
        - If increased lactate- r/o energy metabolism d/o (carnitine, acylcarnitine, nh3, urine oa)
    - If Non AG acidosis
        - Check urine pH (alkalotic if RTA)
- Check CO2 q6 hours until stable

- **Dehydration**
    - Diet causes some diuresis
    - Goal SG<1.025

- **Ketogenic Diet Patients Who Are Admitted to Hospital**
    - Be alert for food and medication with sugar/carbohydrate
    - Alert Nutrition if has to have food or medication with sugar/carb
    - Consult Nutrition for all inpatients on Ketogenic Diet
    - Enter dextrose and propofol "allergies" into Cerner
    - With all med orders, enter Comment: "Ketogenic Diet"

Bicarbonate supplementation for Keto patients

1) Baking soda ¼ teaspoon = 1 packet Cytra-K; mix in 8 oz water to dilute taste, follow labs weekly to determine need to increase
2) Cytra-K packet = 30 meq bicarb and 30 meq K (one packet has 0.063 grams carbohydrate)
3) Bicitra liquid: 1 ml = 1 meq bicarb (1 ml has 0.230 grams carb)

Worse options (limit is typically 1 gram or 1000 mg of carb per day):

1) Cytra-K liquid: 1 ml = 2 meq bicarb and 2 meq K; each 5 ml has 0.5 grams carb (white bottle; the brown bottle has 1 gram per 5 ml)
2) Cytra-2 liquid: 1 ml = 1 meq bicarb; each 5 ml has 0.8 grams carb (brown bottle; the white bottle has 1.5 grams per 5 ml)

Protocol for Bone Health Monitoring of Ketogenic Diet Patients
Based on discussion with Dr. Alison Boyce, Bone Health Clinic, Endocrinology 5/22/14
Plus Endocrine Society guidelines 2011

All patients: Screen with 25-hydroxyvitamin D
Patients with chronic conditions (including anticonvulsants, glucocorticoids, antifungals, obese, AIDS):

Consider deficiency as <32 ng/ml (PTH stabilizes above 32)
Consider sufficient as 40-60 ng/ml
Thus insufficient: 32-39 ng/ml

<u>Treatment of deficiency (D3 or cholecalciferol is better-absorbed):</u>

0-1 year—2000 IU/day for 6 weeks, then maintenance
1-7 years—5000 IU/day
>7 years—5000 IU/day (10,000 IU/day if 25[OH]vit D <10)
Re-check 25[OH]vit D level in 6 weeks, with PTH

For maintenance:
0-1 year—400 IU/day

>1 year—1000 IU/day

Tolerable upper limits:
0-6 months—1000 IU/day
6-12 months—1500 IU/day
1-3 years—2500 IU/day
4-8 years—3000 IU/day
>8 years—4000 IU/day

Criteria for Bone Density Scans and/or Referral to Bone Health Clinic

Bone density scans:
1) Baseline at 3 months if continuing the Diet (would not expect a significant change in bones by 3 mos)
2) >2 years on Ketogenic Diet
(If unable to get bone density scan due to movement, consider lateral spine Xray if concerned, to look for subjective osteopenia, measure vertebral heights over time).

Refer to Bone Health clinic:
1) Persistently low vitamin D levels
2) Fractures or symptoms of bone disease i.e. pain
3) Abnormal bone density scan (especially concerning in ambulating children)
   If low bone density, will check lateral spine Xray to screen for compression fractures (would suggest need for bisphosphonates)
4) < age 1 (risk of toxicity on typical doses of vitamin D)

Consider lab testing:
PTH—if high, body is working hard to keep the calcium high, thus diet is deficient
Random urine calcium—if low, diet is deficient; if high, overdose of supplements (decrease supplements if >60ng/ml; if >80 ng/ml, check urine and serum calcium)
Logistical details:
Bone Health Clinic: 202-476-2121
They are able to do bone density scans or Xrays during the bone health clinic appt
Dexa scans are X-ray absorptiometry—X-rays are passed through the bone to measure the density. Gives much less radiation than a standard X-ray, probably equivalent to flying on an airplane. It involves lying still on a table for up to 10 minutes. Locations analyzed: lateral femur (easier to obtain, decent normative data), total body less head and spine are most validated. It is challenging for some kids with developmental delays or movement disorders, however the

technologists are quite good, and are able to adapt the sites to get data on challenging kids. They do not sedate, but can offer child life and distracters.

Other bone-related data:

Standing is good for lungs, balance, but walking (even with gait trainer) is what helps bone health

Bisphosphonates are not given until fractures are seen, due to lack of data; can be very helpful for pain

DIASTAT DOSING CHART

How to calculate the appropriate individualized dose

The following table provides acceptable weight ranges for each dose and age category, such that patients will receive the maximum beneficial effect—between 90% and 180% of the calculated recommended dose. The safety of this strategy has been established in clinical trials.[1]

DIASTAT® AcuDial™ (diazepam rectal gel)
Dosing Recommendations (by age and weight)

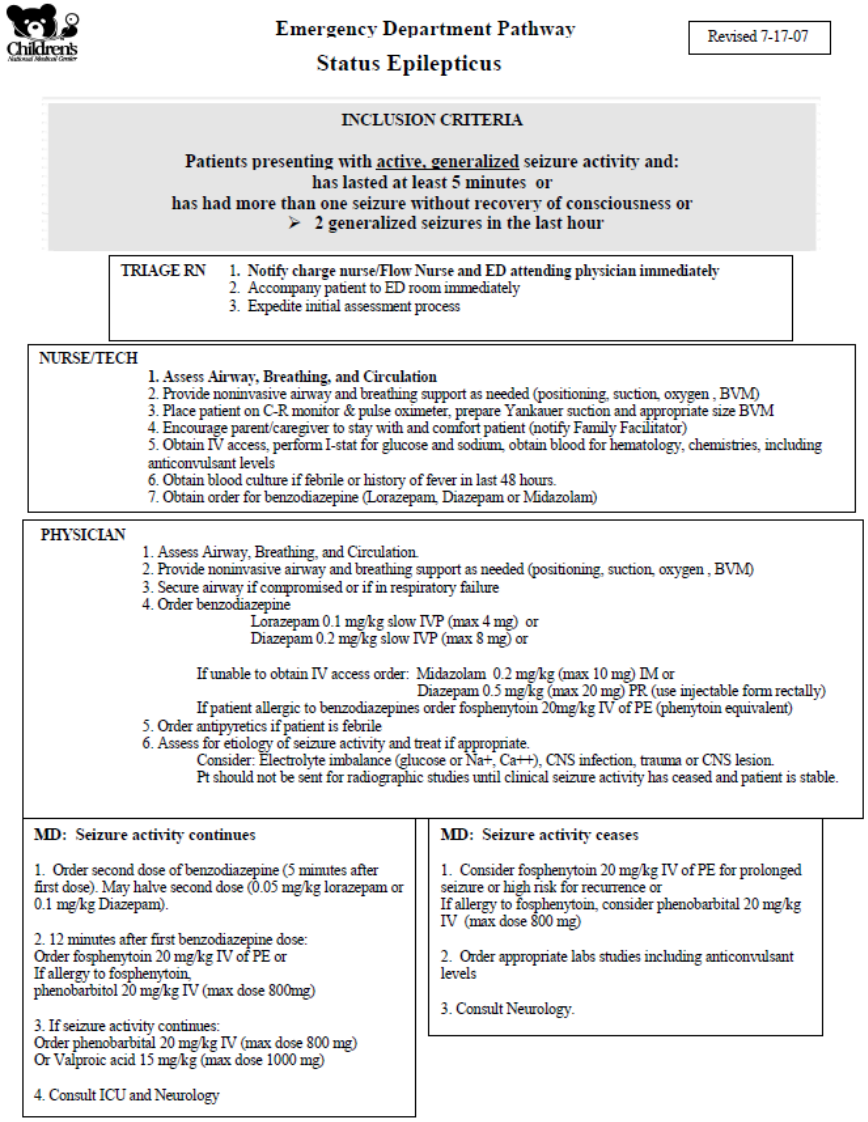
2 to 5 years 0.5 mg/kg		6 to 11 years 0.3 mg/kg		12+ years 0.2 mg/kg	
Weight (kg)	Dose (mg)	Weight (kg)	Dose (mg)	Weight (kg)	Dose (mg)
6 to 10	5	10 to 16	5	14 to 25	5
11 to 15	7.5	17 to 25	7.5	26 to 37	7.5
16 to 20	10	26 to 33	10	38 to 50	10
21 to 25	12.5	34 to 41	12.5	51 to 62	12.5
26 to 30	15	42 to 50	15	63 to 75	15
31 to 35	17.5	51 to 58	17.5	76 to 87	17.5
36 to 44	20	59 to 74	20	88 to 111	20

Please see accompanying full Prescribing Information or visit **www.diastat.com.**

Reference: 1. Diastat AcuDial [package insert]. Aliso Viejo, CA: Valeant Pharmaceuticals North America; 2007.

ANTICONVULSANTS	Supplied	Initial/ Maintenance Dose: Children	Initial and Maintenance Dose: Adults	Maximum Daily Dose	Level mcg/ml*	Adverse Effects	Half-Life	Metab	Ptn Binding
Carbamazepine PO	100 chewable ,200 mg tab	5mg/kg div. BID increase 5mg/kg q3-5d	100mg bid x 3 days; 200 mg bid x 3	35mg/kg (children)	4-12	Diplopia, ataxia, rash, SIADH	12 hours	Hepatic	60-70%
(Tegretol)	100mg/5ml	to 15-20mg/d div BID/TID/QID	days; 300 mg bid x 3 days or 200mg	2400mg/day (adult)		hepatitis, neutropenia,	autoinduction	CYP3A4 85%	
Sustained Release			tid x 1 wk; check level and titrate			aplastic anemia		UDPGT 15%	
(Carbatrol, Equetro)	100,200, 300 mg tab		dose as BID					Renal < 1%	
(Tegretol XR)	100, 200, 400 mg tab							Renal Dosing+	
Clobazam PO	10 mg tab	0.25mg/kg/d div BID, inc Qwk to effect	10 mg QHS inc 10 mg/wk to 30mg/d	1mg/kg/d(child)	N/A	sedation,increased secretions	18hours	Hepatic	83%
				30mg/d(adult)					
Clonazepam PO	0.5,1.0, 2mg tab	0.01-0.03 mg/kg div. QD/BID/TID	0.5-1 mg BID; advance as necessary	0.1-0.2 mg/kg/d (children)	N/A	Rebound insomnia, tremor,	72 hours	Hepatic	80-90%
(Klonopin)	**0.1mg/ml susp***		every 1-2 weeks	20 mg/d (adults)		disinhibition, increased secretions			
(Klonopin wafer)	0.125, 0.25.0.5, 1, 2 ODT					psychosis, sedation			
Diazepam	**2,5,10 mg tab**	0.2 - 0.5 mg/kg/dose IV SE	5-10 mg IV SE		N/A	Sedation, disinhibition	20 - 45 hrs	Hepatic	98%
(Valium)	**5 mg/5ml susp**		2-10 mg BID - QID adjunct ORAL				met. 50-100h		
	5 mg/ml injection		0.3 mg/kg/dose RECTAL						
Diastat	2.5,5,10,15,20 mg supp	0.3-0.5 mg/kg/dose (rectal)							
Ethosuximide PO	250mg tab,	10mg/kg/d div QD/BID,inc 5-10mg/kg/d	250mg; increase 250mg/week to	30-40 mg/kg (children)	40-120	Rash, lupus, psychosis, nausea	50 - 60hrs (a	CYP3A4 65%	<10%
(Zarontin)	250mg/5ml	Q2wk to15-40mg/kg divided QD/BID	500- 1,500 mg/day div QD/BID	2000 mg (adult)			30 - 40hrs (c	Renal 20-25%	
Felbamate PO	400,600mg tab	15mg/kg/day div TID, inc Q 3-7 days to	1200mg/day div TID/QID advance	75-100 mg/kg (children)	40-100	Aplastic anemia, hepatic failure	14 - 23 hrs	CYP3A4 <20%	25%
	600mg/5ml	45mg/kg/day	Q 3-7 days to 2,400-3,600 mg/day	1800-4800mg/day (adult)		anorexia, insomnia, rash		UDPGT 10%	
								Hydrolysis 25%	
								Renal 45-55%	
(Felbatol)								Renal Dosing+	
Gabapentin PO	100,300,400, 600, 800mg tab	5 mg/kg/d div TID/QID, increase by 5 mg/	300 mg initial increase by 300mg/d	45-60 mg/kg (children)	<5	Irritability, sedation, headache	6 hours	Renal > 95%	none
(Neurontin)	250mg/5ml	kg/d for 3 days to 15-20mg/kg/d;	to 900 mg/d div TID/QID;	3600 mg (adult)					
		30-40 mg/kg/d	1,800-2,400 mg/d divided TID					Renal Dosing+	
Lacosamide**	**50**,100,150,200 mg tab	1.5-2mg/kg/day divided BID	50 mg BID; increase weekly 100mg/day	400 mg/day (adult)	NA	dizziness, diplopia, fatigue	13h	demethylation	<15%
(Vimpat)	**10mg/ml inj., solution**		up to 200-400mg/day	8mg/kg/d					
								Renal Dosing+	
Lamotrigine PO	25,100,150,200 mg tab	0.3mg/kg/d x2wks, 0.6mg/kg/d x2wks	25mg QD x 2 wks; 50mg QD\ x 2 wks	15-20mg/kg (children)	4-15	Rash (Stevens-Johnson),	19 - 25 hrs	UDPGT 70-80%	<55%
(Lamictal)	2,5, 25 chew	1.2 mg/kg/d x1wk, 1.8mg/kg/dx 1wk	50 mg bid x 1 wk, 100mg bid x 1 week	(600 mg adult)		hepatitis, sedation, insomnia	(altered	(1A4)	
	25,50,100,200 XR tab	3mg/kg/d x1 wk then inc as nec by	maintenance 5-8 mg/kg/day divided BID	On VPA max 5 mg/kg			by AED	Renal 10%	
	1 mg/ml susp*	1-1.2mg/kg/d Qwk to 5-10mg/kg/d div BID					interactions)		
		On VPA, start 0.1-0.15 mg/kg/d; inc to					varies w/age		
		0.15 mg/kg/d x2wks inc 0.3mg/kg/dx 2wks							
		inc 0.5mg/kg/d x 1wk inc 0.5mg/kg/d Qwk							
		to maint 2 mg/kg/d							
Levetiracetam PO, IV	250,500,750,1000mg tab	5-10 mg/kg/day div BID	500 mg/day increased every 1-2 weeks	100mg/kg/day (children)	trough 7-34	Dizziness, somnolence,	6-8hrs	Hydrolysis 24%	none
(Keppra)	100mg/ml	advance 5-10/mg/d Qweek,	in 500 mg/day increments to	3-4,000mg/day (adult)	peak 36-70	irritability, psychosis		Renal 66%	
	500, 750 mg XR tab	to 20-30 mg/kg/day	2000 - 3000mg/day						
					for max dosing			Renal Dosing+	
Lorazepam PO,IV,IM	0.5,1,2 mg tab	0.025-0.10 mg/kg Q8hrs	1-4 mg/dose q 6-12 hours	10mg/day	N/A	Sedation, respiratory suppression	12 hrs	Hepatic	85%
(Ativan)	2mg/ml	usually start 0.05 mg/kg/dose b/c resp supr.	(usually max 1-2 mg/dose)			disinhibition, dizziness			
Oxcarbazepine PO	150,300,600mg tab;	Start 5 mg/kg/day divided BID, advance	Initial 300mg BID, increase	50-60mg/kg/day (child)	8-35	Fatigue, headache, dizziness,	2 hrs (9 hrs	CYP 4%	40%
		by 5 mg/kg/day q 3-7 days to 20-35 mg/kg/day	by 300 mg/day q 3-7 days to			ataxia, hyponatremia, rash		UDPGT 49%	
								Renal 27%	
(Trileptal)	300mg/5ml		to 1200-2400 mg/day	4400mg/day (adult)			metabolite)	Renal Dosing+	
Phenobarbital	15,30,60,100 mg tab,	4-6mg/kg/d div BID (neonate)	30-60 mg/d, increase every 1-2 weeks	12 mg/kg/day (child<5)	15-40	Rash, Stevens-Johnson, serum	96 hrs	CYP 20-35%	45%-55%
PO,IM,IV	20mg/5ml	3-5 mg/kg/d div BID (child)	to 60-180 mg	6 mg/kg/d (chld>5)		sickness, depression, rickets,	(varies w/	(2C9/19)	
								UDPGT 15-30%	
								Renal 20-25%	
				(1-3mg/kg adult; 240 mg/d)		hyperactivity	age)		
Phenytoin PO,IV	30,100 tab, 50 chewable	8-12mg/kg/d div Q8hr (neonates, infants)	200-300 mg/d (QD or BID)	12 mg/kg/day (neonates)	10-20	Rash, hepatitis,rickets, gingival	7 - 42 hrs	CYP 90%	85%-90%
(Dilantin)	125mg/5ml (bad form)	5 mg/kg/d div BID (child, adol)	(oral load 20 mg/kg over 24-48 hours	8 mg/kg/day (children)	free:0.1-2.0	hypertrophy, coarsening of	varies w/age	(2C9 major/	
								2C19 minor)	
Fosphenytoin - IV	200,300ER		divided q 8-12 h)	400-500 mg/day (adult)		facial features (kids), hirsutism		Renal < 5%	
Pregabalin PO	25, 50, 75, 100, 150, 200,	2mg/kg/d div TID inc by 2mg/kg/d	50mg TID inc q wk by 50mg TID	6-8 mg/kg/d (child)	N/A	myalgia, dec plt,sedation	6hrs	none	minimal
(Lyrica)	300mg tab	to 4-6mg/kg/d	to 300mg/d	600mg/day(adult)		rash, CHF exacerbation,wt gain			
Primidone PO	50,250 mg tabs	Start 50mg QHS for 3 days; 50 mg	Start 50mg/d, advance	25 mg/kg/day (children)	5-13	Rash, Stevens-Johnson, serum	8-12 hrs	Hepatic > 40%	<30%
(Mysoline)		BID for 3 days;100mg BID for 3 days.	125mg/Q week	1500mg/day (adult)		sickness, depression, nausea,		Renal 40-60%	
		maintenance: 125-250mg TID (10-25 mg/kg)				hyperactivity			
Rufinamide PO	200,400 mg tabs	10 mg/kg/d div BID, inc by 10 mg/kg/d	>30 kg; 400 mg daily, increase by 400 mg	1800 mg/day (child- 60mg/kg/d)	NA	Headaches, dizziness, somnolence	6-10 hrs	Renal	34%
(Banzel)		Q5-7days to goal 30-45 mg/kg/day	q2d to max of 1.8 gm daily div BID	3200 mg daily (adult > 70 kg)		nausea, vomiting, anorexia, rash			
		if VPA 1/2 dosing , so start 5 mg/kg/d etc	> 50 kg; up to 2.7gm daily div BID			shortened QT interval			
				on VPA max 30 mg/kg/d					
Topiramate PO	25,50, 100,200mg tab	Start 0.5 - 1.0 mg/kg/day div QD/BID	50mg/day for 2 weeks; increase	15-18 mg/kg/day (children)	(10-35)	Anorexia, encephalopathy	18-23 hrs	CYP 20-40%	15%
(Topamax)	15, 25 mg sprinkles	Increase 0.5-1mg/kg q 2 weeks x 2	by by 50mg every week	(25 mg/kg infantile spasms)		somnolence, nausea, ataxia		Renal 60-70%	
	6 mg/ml suspension*	then q week to 5-8mg/kg QD/BID	to 400mg/day total dose	600 mg/d (adult)		confusion,dysarthia, renal stones			
		Infants start 1-2mg/kg/d				anhidrosis, glaucoma, metabolic acidosis		Renal Dosing+	
Tiagabine PO	2,4,12,16,20 mg tab	0.1mg/kg/d. Increase 0.1 mg/kg/weekly	4mg QD. Increase by 4-8mg	32mg/day (children)	N/A	Dizziness, nausea, somnolence,	4-8 hours	CYP3A4 90%	96%
(Gabitril)		to 0.4mg/kg/d div TID	at weekly intervals up to 32-56mg/day	56mg/day (adult)		rash, non-convulsve status		Renal < 2%	
Valproate PO	125,250,500mg tab,	Start 5 - 10 mg/kg/day po div QD/BID	125 BID for 3-5 days then	60-70mg/kg/day (children)	50-110	Hepatitis, pancreatitis,	10 hrs (oral)	CYP < 20%	70%-90%
(Depakote)	125 mg sprinkle	increase by 5 - 10 mg/kg/day div bid q3-5 days	250 BID for 3-5 days; then 500 mg BID	4000-5000 mg/d (adult)		thrombocytopenia (dose dep)		UDPGT 30-50%	
(Depakene)	250 cap; 250mg/5ml	to 15 - 25 mg/kg/day div BID/TID				alopecia, weight gain, nausea,		(1A3 & 2B7)	
(Depakote ER)	250, 500ER	IV load 20mg/kg/dose (max < 20 mg/min)				polycystic ovarian dysfunction	4-6 hrs (IV)	Oxidation 40%	
		IV maintenance is total daily dose div q6hrs				Somnolence, headache, nausea		Renal < 3%	
(Depacon) IV									
Vigabitrin PO	500mg tab	25mg/kg/d div QD/BID then	500 mg BID	100 mg/kg/day	N/A	Visual field loss, drowsiness,	5-7 hours	Renal	none
(Sabril)		Inc by 25 mg/kg/d Qwk	increase in 500 mg increments	200 mg/kg/d infantile spasms		weight gain, viral illness			
		to 50-100mg/kg/d div QD/BID	no addtion'l benefit with >4g/day	4.0mg/day (adult)		dizziness,agitation, cough		Renal Dosing+	
Zonisamide PO	25, 50, 100 mg capsules	2mg/kg/day inc 2 mg/kg/d Q2wks	100-200mg for adults, adjusted every	12 mg/kg/day (children)	max 40	Somnolence, anorexia, anhidrosis	50-68 hours	CYP3A4 50%	40%
(Zonegran)		to 8- 10mg/kg/d div QD/BID	2 weeks by 100-200 mg	400mg/day (adult)		confusion, depression, agitation,		Acetylation 15%	
						hyperthermia,		Renal 35%	
						sulfa-allergies (Stevens-Johnson)		Renal Dosing+	

Emergency Department Protocols



Children's National Medical Center

Emergency Department Pathway

Status Epilepticus

Revised 7-17-07

INCLUSION CRITERIA

Patients presenting with active, generalized seizure activity and:
has lasted at least 5 minutes or
has had more than one seizure without recovery of consciousness or
- **2 generalized seizures in the last hour**

TRIAGE RN
1. **Notify charge nurse/Flow Nurse and ED attending physician immediately**
2. Accompany patient to ED room immediately
3. Expedite initial assessment process

NURSE/TECH
1. **Assess Airway, Breathing, and Circulation**
2. Provide noninvasive airway and breathing support as needed (positioning, suction, oxygen , BVM)
3. Place patient on C-R monitor & pulse oximeter, prepare Yankauer suction and appropriate size BVM
4. Encourage parent/caregiver to stay with and comfort patient (notify Family Facilitator)
5. Obtain IV access, perform I-stat for glucose and sodium, obtain blood for hematology, chemistries, including anticonvulsant levels
6. Obtain blood culture if febrile or history of fever in last 48 hours.
7. Obtain order for benzodiazepine (Lorazepam, Diazepam or Midazolam)

PHYSICIAN
1. Assess Airway, Breathing, and Circulation.
2. Provide noninvasive airway and breathing support as needed (positioning, suction, oxygen , BVM)
3. Secure airway if compromised or if in respiratory failure
4. Order benzodiazepine
   Lorazepam 0.1 mg/kg slow IVP (max 4 mg) or
   Diazepam 0.2 mg/kg slow IVP (max 8 mg) or

   If unable to obtain IV access order: Midazolam 0.2 mg/kg (max 10 mg) IM or
   Diazepam 0.5 mg/kg (max 20 mg) PR (use injectable form rectally)
   If patient allergic to benzodiazepines order fosphenytoin 20mg/kg IV of PE (phenytoin equivalent)
5. Order antipyretics if patient is febrile
6. Assess for etiology of seizure activity and treat if appropriate.
   Consider: Electrolyte imbalance (glucose or Na+, Ca++), CNS infection, trauma or CNS lesion.
   Pt should not be sent for radiographic studies until clinical seizure activity has ceased and patient is stable.

MD: Seizure activity continues

1. Order second dose of benzodiazepine (5 minutes after first dose). May halve second dose (0.05 mg/kg lorazepam or 0.1 mg/kg Diazepam).

2. 12 minutes after first benzodiazepine dose:
Order fosphenytoin 20 mg/kg IV of PE or
If allergy to fosphenytoin,
phenobarbitol 20 mg/kg IV (max dose 800mg)

3. If seizure activity continues:
Order phenobarbital 20 mg/kg IV (max dose 800 mg)
Or Valproic acid 15 mg/kg (max dose 1000 mg)

4. Consult ICU and Neurology

MD: Seizure activity ceases

1. Consider fosphenytoin 20 mg/kg IV of PE for prolonged seizure or high risk for recurrence or
If allergy to fosphenytoin, consider phenobarbital 20 mg/kg IV (max dose 800 mg)

2. Order appropriate labs studies including anticonvulsant levels

3. Consult Neurology.

Emergency Department Pathway

Revised 2-1-06

NEW ONSET AFEBRILE SEIZURE

INCLUSION CRITERIA

First time seizure activity in a patient without a fever

NOTE: This pathway excludes patients in status epilepticus, previous seizure history, known seizure disorder, with fever, ventricular shunt, known head trauma or known ingestion

TRIAGE RN

1. **If actively seizing,** "Scoop & run" to ED, call for help, then expedite Triage process once ED physician and nurse assume care
2. **If post-ictal with altered mental status,** place in ED room, then expedite Triage process once ED nurse assumes care
3. **If awake and alert,** expedite Triage process and facilitate placement in ED room

NURSE/TECH

1. **Place on cardiac-respiratory and pulse oxymetry monitor *(POx)* if**
   - Actively seizing
   - Post-ictal with altered mental status
   - Concern for meningitis
2. **Document vital signs, POx and condition assessment**
   - Actively seizing: Q 5 minutes
   - Post-ictal with altered mental status: Q 30 minutes
   - Concern for meningitis Q 1 hour
   - Awake/alert, no concern for meningitis Q 2 hours
3. **Document temperature Q 4 hrs**
4. **Perform STAT, if RN assessment identifies concern for meningitis**
   - Notify Staff Physician *(not resident)* immediately
   - Establish IV access *(saline lock)*
   - Draw CBC, blood culture, BMP
5. **Ensure patient supervision** by adult family member or ED staff when out of bed

PHYSICIAN

1. **Evaluate patient.**
2. **Identify predisposing conditions for seizure activity** *(e.g. signs/symptoms of meningoencephalitis, toxic ingestion, recent closed head injury, cerebral vascular accident, coagulopathies, sickle cell disease, hydrocephalous, travel to cystcercosis endemic areas, hypertension, hypoglycemia, etc.)*
3. **Consider and order the following when appropriate:**
   - **"D Stick" Glucose:** If suspected hypoglycemia.
   - **Blood Work:** Order BMP. (Order CBC only if starting an anticonvulsant that suppresses bone marrow, or if needed for fever evaluation.)
   - **Urine tox screen** and/or **serum ethanol level:** If suspected ingestion.
   - **CT Scan:** If focality to history or to exam, persistent altered mental status/no return to baseline, if MRI not available within 24 hours of admission, or if not being admitted. Otherwise, CT or MRI will be performed as inpatient.
   - **LP:** If suspected meningoencephalitis. Consider need for CT prior to LP.
   - **EKG:** If concern for arrhythmia or syncope.
4. **Order inpatient bed on Neurology Service** as soon as emergent condition excluded.
5. **Discuss with Neurology/Admit.**

RN

1. **Accompany to CT if ordered** and monitor during procedure
2. **Provide parent/caregiver teaching about what to do if a seizure occurs again**
   - Refrain from putting fingers or objects in child's mouth
   - Position to facilitate breathing during seizure *(Side-lying or on back with modified jaw thrust)*
   - Call for help and seek timely medical evaluation

Revised 2-1-06

Emergency Department Pathway
SEIZURE WITH FEVER
IN CHILDREN 6 MONTHS TO 6 YEARS OF AGE

INCLUSION CRITERIA

Seizure activity accompanied by fever in child age 6 months to 6 years
(Temperature elevations: Rectal ≥38.0 °C *or* Oral ≥37.6°C)

NOTE: This pathway excludes patients with status epilepticus, known seizure disorder, ventricular shunt, known head trauma or ingestion

TRIAGE RN

1. **If actively seizing,** "Scoop & run", call for help, then expedite Triage process when ED physician and nurse assume care
2. **If post-ictal with altered mental status,** place in ED room, then expedite Triage process when ED nurse assumes care
3. **If awake and alert,** expedite Triage process
4. **Administer antipyretics** per EMTC policy # D8m: "Treatment of Fever"

NURSE/TECH

1. **Place on cardiac-respiratory and pulse oxymetry monitors *(POx)* if**
   - Actively seizing
   - Post-ictal with altered mental status
   - Concern for meningitis
2. **Document vital signs, POx and condition assessment**
   - Actively seizing: Q 5 minutes
   - Post-ictal with altered mental status: Q 30 minutes
   - Concern for meningitis Q 1 hour
   - Awake/alert, no concern for meningitis Q 2 hours
3. **Document temperature Q 2 hrs**
4. **Perform STAT, if RN assessment identifies concern for meningitis**
   - Notify Staff Physician *(not resident)* immediately
   - Establish IV access *(saline lock)*
   - Draw and send CBC, blood culture, BMP
5. **Ensure patient supervision** by adult family member or ED staff when out of bed

PHYSICIAN

1. **Evaluate patient**
2. **If simple febrile seizure,** perform fever evaluation as needed
   - Strongly consider LP if age <6 months
3. **If meningitis suspected**
   - Follow-up on bloodwork (CBC, blood culture, BMP)
   - Perform LP, after considering need for CT scan
   - Consider ordering *STAT* antibiotics
4. **If CNS lesion/focality suspected,** order emergent CT
5. **If complex febrile seizure** *(focal seizure activity, seizure lasting >15 minutes, multiple seizures within 24 hours, persistent altered mental status),* perform fever evaluation and strongly consider:
   - BMP *(Not usually indicated for simple febrile seizure)*
   - CT
   - LP
   - Discussion with Neurology

RN

1. **Provide parent/caregiver teaching about fever control and what to do if seizure occurs again**
   - Review correct dosing of antipyretics, as well as other methods of fever control (remove clothing, washcloths, bathing, etc.)
   - Refrain from putting fingers or objects in child's mouth
   - Position to improve breathing during seizure *(side-lying or on back with modified jaw thrust)*
   - Call for help and seek timely medical evaluation

PHYSICIAN

1. **Reassess patient**
2. **Give follow-up instructions** for PMD evaluation within 24 hrs

Never abbreviate medication names. Other prohibited abbreviations:				
U IU µg	Leading Decimal (.2 mg) Trailing Zero (5.0 mg)	QD QOD	MS MSO4 MgSO4	HCT HCTZ

ED Migraine Plan or ED Migraine Plan UMC

Pt wt: kg Allergies:

Addressograph

☒	Service	Emergency Medical Service at CNMC
☒	Service	Emergency Medical Service at UMC
INCLUSIONS		
•	Headache lasting 1-72 hours	
•	Headache has ≥ 2 of the following characteristics: 1. Unilateral location, may be bilateral, front to temporal (not occipital) 2. Pulsing quality 3. Moderate or severe pain intensity 4. Aggravated by or causing avoidance of routine physical activity.	
•	During headache at least 1 of the following occur: 1. Nausea / vomiting or both 2. Photophobia or phonophobia, which may be inferred from behavior	
•	Headache not attributed to another disorder (see below)	
•	Normal neurologic exam including lack of papilledema, cranial nerve palsies, nuchal rigidity, ataxia, clonus, or abnormal/asymmetric reflexes	
EXCLUSIONS		
•	Trauma, focal neurological deficit, abnormal head imaging, ataxia, abnormal reflexes or eye movements, hypertension, papilledema, fever, or pregnancy	
ED INITIATION ORDERS		
☒	Communication Order	Confirm patient meets inclusion criteria of the ED Migraine Headache Pathway
☒	Assessment Pain with Pain Scale	STAT
☒	Communication Order	Inform MD stat if pain >5/10 and obtain orders for pain medication per pathway
☒	Communication Order	Urine pregnancy test in female patients post menarchal STAT
LABS		
☐	Urine Pregnancy Test perform POC	STAT Once

Never abbreviate medication names. Other prohibited abbreviations:				
U IU µg	Leading Decimal (.2 mg) Trailing Zero (5.0 mg)	QD QOD	MS MSO4 MgSO4	HCT HCTZ

Neurology Headache Plan

Pt wt: kg Allergies:

Addressograph

☒	**ADMIT TO Plan**					
Indicate diagnosis as status migranosus or as headache refractory to first line medications						
•	Service	Neurology	Attending		Diagnosis	

REFERENCES

- Reference (CNMC INTRAnet)
- Reference (INTERnet)

INCLUSIONS

- Headache refractory to >= 2 therapeutic medications provided in outpatient setting or ED
- Requires admission for escalation of treatment of headache
- Intractable debilitating headache

EXCLUSIONS

- Pregnant patients
- History of metabolic liver disorder

RELATIVE CONTRAINDICATIONS

Consider neuroimaging or further testing if any of the following criteria are met:

- Neurologic exam abnormalities including papilledema, abnormalities in eye movements, stiff neck, weakness, dysmetria, ataxia, hyperreflexia
- Presence of fever with headache
- Migraine in child under 5 years of age
- Headache that awakens a child from sleep repeatedly
- Sudden onset of severe headache (worst headache of life) or subacute progressive headache
- Headache associated with excessive vomiting

- Headache associated with substantial period of confusion
- Brainstem aura including headache with new focal weakness, speech changes (aphasia or dysarthria), tinnitus, vertigo, diplopia, ataxia
- A change in headache quality compared to prior headaches
- Absent family history of headache
- Family history of CNS tumors or multiple cerebral aneurysm
- Posterior location of headache
- Sudden severe headache associated with bursting or popping sensation followed by light sensitivity and nausea
- Patient with other complex medical disease including but not limited to sickle cell, oncologic process, stroke, clotting disorder, congenital or acquired cardiac disease, autoimmune disorders

CONDITION		
☒	Condition	☐ Good ☐ Fair ☐ Guarded ☐ Critical
VITAL SIGNS		
☒	VS Vital Signs	Every 4 hours
☒	Pain with Pain Scale	With vital signs and 30 minutes after each headache medicine administered
☒	PEWS (Standard Escalation)	
☒	Notify Provider For	Vital signs outside of age-specific ranges or alarm parameters
ACTIVITY		
☒	Activity ad lib	
NURSING		
☒	Measure Weight	Once on admit

☒	Measure Height/Length	Once on admit
☒	Measure Head Circumference	Once on admit
☒	Measure Strict I&O	
☒	CR monitor – Acute Care	Specify Monitor Type Pulse Oximetry, Instructions for Use with Vital Signs
DIET		
☒	Regular diet	
CONTINUOUS INFUSIONS		
☒	Sodium Chloride 0.9% Bolus	_____mL (20 mL/kg/dose; max 1000 mL/dose) IV STAT once over 30 minutes. *Give prior to starting maintenance IV fluids.*
☒	Dextrose 5% with 0.9% NaCl	_____mL/hr IV
LABS		
☒	CBC with diff	Lab collect, once
☒	CMP	Lab collect, once
☒	HCG Urine	
DIAGNOSTIC TESTS		
If anticipate ordering dihydroergotamine (DHE)		
☒	EKG - Tracing	Rule out coronary artery disease prior to starting dihydroergotamine (DHE)
RADIOLOGY		
☐	MRI Brain without contrast	Evaluate for space occupying lesion, cerebral edema and/or hemorrhage
☐	CT Head or Brain without contrast	Evaluate for space occupying lesion, cerebral edema and/or hemorrhage
CONSULTS		
☐	Consult Pain Medicine	
☒	Child Life Assessment	Coping with illness/injury, Medium priority
☐	Consult Social Work	Coping with chronic illness, Medium priority
MEDICATIONS		
☒	LMX Lidocaine Cream	1 app cream TOP every 1 hour PRN painful procedure – anticipated, routine
If not previously ordered in ED or prior to admission		
☐	**Neurology Headache Phase I Subplan**	
For ordering only by Neurology Service OR with approval by Neurology Consult		
☐	**Neurology Headache Phase II (Dihydroergotamine (DHE)) Subplan**	

Never abbreviate medication names. Other prohibited abbreviations:

U IU µg	Leading Decimal (.2 mg) Trailing Zero (5.0 mg)	QD QOD	MS MSO4 MgSO4	HCT HCTZ

Neurology Headache Phase I Plan

Pt wt: kg Allergies:

Addressograph

For use only with Neurology Headache Plan

MEDICATIONS

☒	Ketorolac (Toradol)	_____mg (0.5 mg/kg/dose; max 30 mg/dose) IV every 6 hours
☒	Prochlorperazine (Compazine)	_____mg (0.15 mg/kg/dose; max 10 mg/dose) IV every 8 hours PRN nausea/vomiting
If Prochlorperazine (Compazine) not available		
☐	Metoclopramide (Reglan)	_____mg (0.15 mg/kg/dose; max 10 mg/dose) IV every 8 hours PRN nausea/vomiting
☒	Valproic acid	_____mg (20 mg/kg/dose; max 1000 mg/dose) IV once
☒	Diphenhydramine	_____mg (1 mg/kg/dose; max 50 mg/dose) IV once STAT PRN acute dystonic reaction

Never abbreviate medication names. Other prohibited abbreviations:

U IU µg	Leading Decimal (.2 mg) Trailing Zero (5.0 mg)	QD QOD	MS MSO4 MgSO4	HCT HCTZ

Neurology Headache Phase II (Dihydroergotamine (DHE)) Plan

Pt wt: kg Allergies:

Addressograph

For use only with Neurology Headache Plan		
RESTRICTIONS		
•	For ordering ONLY by the Neurology Service or with Neurology Consult approval	
REFERENCES		
•	Reference (CNMC INTRAnet)	Children's National Formulary: Dihydroergotamine Mesylate (DHE) Repetitive Intravenous Injection Guidelines *http://online.lexi.com/lco/action/doc/retrieve/docid/chinat_f/1923943*
EXCLUSIONS		
•	Triptan use within past 24 hours, e.g. Imitrex (sumatriptan), Zomig (zolmitriptan), Maxalt (rizatriptan)	
•	Potent 3A4 inhibitors , e.g. macrolides, protease inhibitors (e.g. ritonavir, nelfavir, indinavir)	
•	Azole antifungals, e.g. ketoconazole, itraconazole	
•	MAO Inhibitor (e.g. selegiline, phenelzine) use within last 2 weeks	
•	Ischemic heart disease, uncontrolled hypertension, sickle cell anemia, or coronary artery vasospasm	
•	Hemiplegic migraine or basilar migraine	
•	Severe renal and/or hepatic impairment	
•	Pregnancy	
•	Blood pressure elevated >= 2 standard deviations of normal for age	
•	History of stroke	
•	Abnormal EKG - need Cardiology clearance before proceeding with dose	
VITAL SIGNS		
☒	Vital Signs	Prior to initiating DHE infusion and every 15 minutes for 1st hour after DHE infusion complete
☒	Notify Provider For	*DHE adverse effects, including:* *Nausea/ vomiting, abdominal discomfort, chest discomfort, limb pain, diarrhea,*

		paresthesias, dystonia *Cardiovascular effects: vasospasms, tachycardia, bradycardia, hypertensior* *Coldness of skin and/or numbness and tingling of extremities (may indicate ergotism, which can lead to gangrene)*
NURSING		
☒	Cardiorespiratory/CR Monitor – Acute Care	☒ Cardiac ☒ Respiratory ☒ Pulse Oximetry ☒ Continuous Rationale: During DHE Infusion and for 1 hour post-infusion
☒	Communication Order	If female patient, must have a negative pregnancy test prior to receiving DH
☒	Notify Provider	EKG complete
MEDICATIONS		
DIHYDROERGOTAMINE (DHE)		
INITIAL DOSE: Test Dose 1		
☒	Dihydroergotamine	_____mg (0.007 mg/kg/dose, max 0.4 mg/dose) IV once *(Test Dose 1. Star minutes after pre-medications.)* *Notify provider for vomiting, worsening of headache, itching, flushing, chest paresthesias or abdominal pain*
INITIAL DOSE: Test Dose 2: If no vomiting, worsening of headache, itching, flushing, chest pain, paresthesias or abd pain following Dihydroergotamine (DHE) Test Dose 1, proceed with Test Dose 2		
☒	Dihydroergotamine	_____mg (0.007 mg/kg/dose, max 0.4 mg/dose) IV once *(Test Dose 2. Star minutes after Day 1 Test Dose 1 completed.)* *DO NOT ADMINISTER if vomiting, worsening of headache, itching, flushing pain, paresthesias or abdominal pain with Test Dose 1 and notify provider.*
MAINTENANCE DOSE:		
☒	Dihydroergotamine	_____mg (0.014 mg/kg/dose, max 1 mg/dose) every 8 hours x 10 doses. *St hours after 1st DHE test dose.*
PRE-MEDICATIONS		

Children's National Medical Center

Ventricular Shunt Malfunction Diagnostic Algorithm

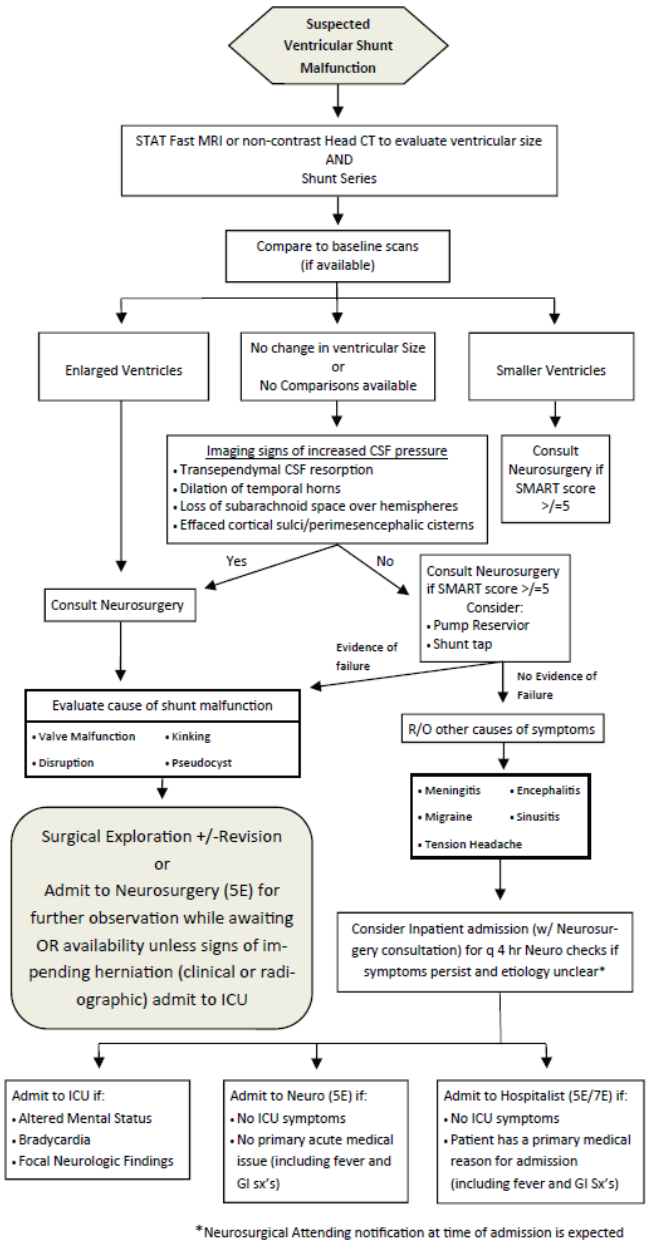
June 2013

Acute Symptoms		
• Nausea	• Headache	• Irritability
• Vomiting	• Positional Headache	• Lethargy
• Hypertension	• Double Vision	• Stupor
• Bradycardia	• Sundown Sign	• Coma
• Seizures	• Transient visual obscurations (e.g. visual blackouts)	

Subacute/Chronic Symptoms	
• Change in behavior (e.g. agitation)	• Developmental regression
• Altered gate	• Change in cognitive function (e.g. attention span)
• Change in feeding patterns	• Daily headaches
• Change in school performance	• Increased head size

History to be Obtained Prior to Neurosurgical Consult
• Prior history of shunt failure
• Size of ventricles at last shunt failure
• Prior history of shunt failure without change in ventricular size
• Presence or absence of fevers
• Presence or absence of above acute and chronic symptoms

Fast MRI vs Head CT	
• Available weekdays 8 am– 10 pm and weekends 8 am – 4 pm	
• Patient must be able to lie still/cooperate for 10 minutes without sedation (roughly age >/=5 yrs)	
Contraindications to Fast MRI	
• History of trauma	• Altered mental status
• r/o hemorrhage or pneumocephalus	• If shunt catheter needs to be visualized
• Programmable VP shunts	• Patients with other MRI contraindication (ex– pacemaker)
• Unable to obtain within 1 hour of presentation	



SMART SCORE

(SHUNT MALFUNCTION ASSESSMENT AND REASSESSMENT TOOL)

Inclusion Criteria: Patient with VA or VP shunt and age > 1 month
Exclusion Criteria: Patients in the NICU

Major Criteria: (10 points each)

* Clinical signs of herniation (including Cushing's Triad)
* Imaging with signs of herniation/impending herniation
* Papilledema

Minor Criteria:

Tier 1 (5 points each)

* Positional headache (include awakening from sleep due to headache pain)
* New diplopia/CN VI palsy
* Altered MS (e.g. irritability, lethargy) in a patient w/ no (or mild) prior neurological deficit
* Neuroimaging with increased size of ventricles and/or decrease in sulci and/or cisterns (more sensitive if baseline large ventricles)
* History of VP shunt failure without prior change in ventricular size
* Head circumference increasing across percentiles (e.g. > 10%)

Tier 2 (2 points each)

* Nausea/vomiting
* Headache
* Bradycardia (not baseline)
* Increased seizure frequency from baseline
* Recent manipulation of VP shunt (e.g. change of setting of valve, revision in the last 30 days)
* Change in baseline in a patient with moderate to severe baseline deficits
* Neuroimaging with inc size of ventricles and no change in sulci and/or cisterns
* Radiographic evidence of shunt tubing disconnection

Recommended response based on assigned score

10 = immediate response from neurosurgery attending

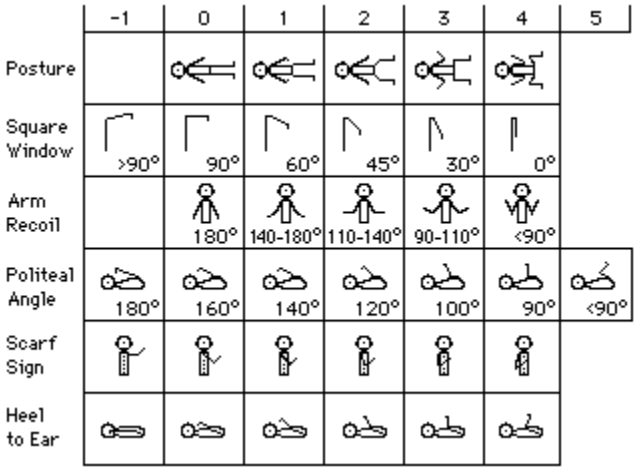
5 -9 = high suspicion for shunt malfunction, neurosurgical consult. If no intervention prescribed, consider escalation to neurosurgical attending.

4 = moderate suspicion for shunt malfunction. Observation recommended. If symptoms not easily explained by other medical condition, consult neurosurgery.

** If score remains ≥ 4 consider repeat imaging Q2-3 days and/or ophthalmology evaluation for papilledema, even if other explanations plausible for observed symptoms.

Dubowitz/Ballard Exam for Gestational Age

Neuromuscular Maturity



Posture: With the infant supine and quiet, score as follows:

- Arms and legs extended = 0
- Slight or moderate flexion of hips and knees = 1
- Moderate to strong flexion of hips and knees = 2
- Legs flexed and abducted, arms slightly flexed = 3
- Full flexion of arms and legs = 4

Square Window: Flex the hand at the wrist. Exert pressure sufficient to get as much flexion as possible. The angle between the hypothenar eminence and the anterior aspect of the forearm is measured and scored:

- >90 degrees = -1
- 90 degrees = 0
- 60 degrees = 1
- 45 degrees = 2
- 30 degrees = 3
- 0 degrees = 4

Arm Recoil: With the infant supine, fully flex the forearm for 5 seconds, then fully extend by pulling the hands and release. Score the reaction:

- Remains extended 180 degrees, or random movements = 0
- Minimal flexion, 140-180 degrees = 1
- Small amount of flexion, 110-140 degrees = 2
- Moderate flexion, 90-100 degrees = 3
- Brisk return to full flexion, <90 degrees = 4

Popliteal Angle: With the infant supine and the pelvis flat on the examining surface, the leg is flexed on the thigh and the thigh fully flexed with the use of one hand. With the other hand the leg is then extended and the angled scored:

- 180 degrees = -1
- 160 degrees = 0
- 140 degrees = 1
- 120 degrees = 2
- 100 degrees = 3
- 90 degrees = 4
- <90 degrees = 5

Scarf Sign: With the infant supine, take the infant's hand and draw it across the neck and as far across the opposite shoulder as possible. Assistance to the elbow is permissible by lifting it across the body. Score according to the location of the elbow:

- Elbow reaches or nears level of opposite shoulder = -1
- Elbow crosses opposite anterior axillary line = 0
- Elbow reaches opposite anterior axillary line = 1
- Elbow at midline = 2
- Elbow does not reach midline = 3
- Elbow does not cross proximate axillary line = 4

Heel to Ear: With the infant supine, hold the infant's foot with one hand and move it as near to the head as possible without forcing it. Keep the pelvis flat on the examining surface. Score as shown in the diagram above.

Physical Maturity

Sign	-1	0	1	2	3	4	5
Skin	Sticky, friable, transparent	Gelatinous red, translucent	Smooth pink, visible veins	Superficial peeling and/or rash, few veins	Cracking, pale areas, rare veins	Parchment, deep cracking, no vessels	Leathery, cracked, wrinkled
Lanugo	None	Sparse	Abundant	Thinning	Bald areas	Mostly bald	
Plantar Creases	Heel-toe 40-50 mm = -1, <40 mm = -2	Heel-toe >50 mm, no creases	Faint red marks	Anterior transverse crease only	Creases over anterior 2/3	Creases over entire sole	
Breast	Imperceptible	Barely perceptible	Flat areola, no bud	Stippled areola, 1-2 mm bud	Raised areola, 3-4 mm bud	Full areola, 5-10 mm bud	

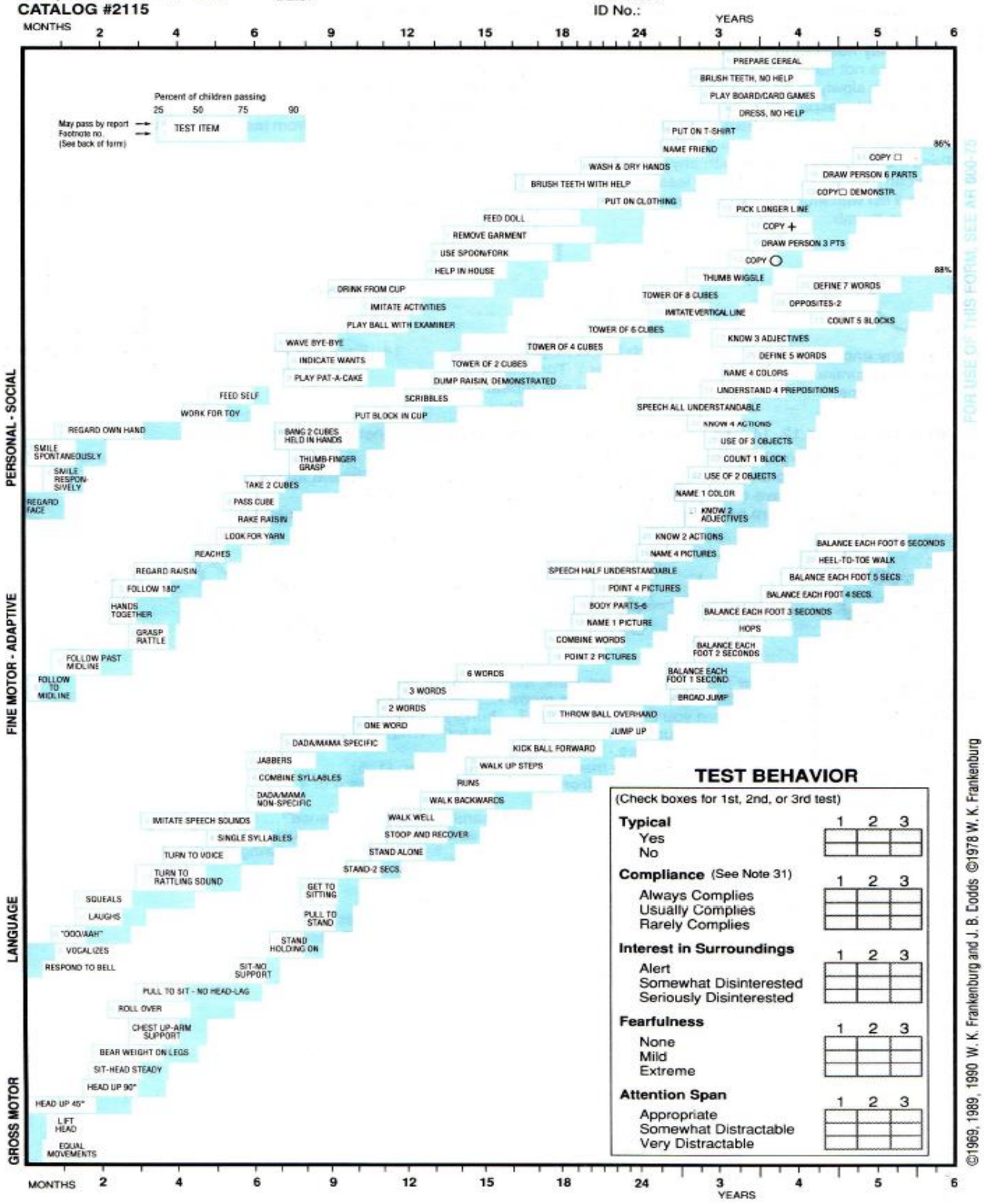
Eye & Ear	Lids fused, loosely = -1, tightly = -2	Lids open, pinna flat, stays folded	Slightly curved pinna, soft with slow recoil	Well-curved pinna, soft but ready recoil	Formed and firm, with instant recoil	Thick cartilage, ear stiff
Genitals, male	Scrotum flat, smooth	Scrotum empty, faint rugae	Testes in upper cannal, rare rugae	Testes descending, few rugae	Testes down, good rugae	Testes pendulous, deep rugae
Genitals, female	Clitoris prominent, labia flat	Prominent clitoris, small labia minora	Prominent clitoris, enlarging minora	Majora and minora equally prominent	Majora large, minora small	Majora cover clitoris and minora

Maturity Rating

Add up the individual Neuromuscular and Physical Maturity scores for the twelve categories, then obtain the estimated gestational age from the table below.

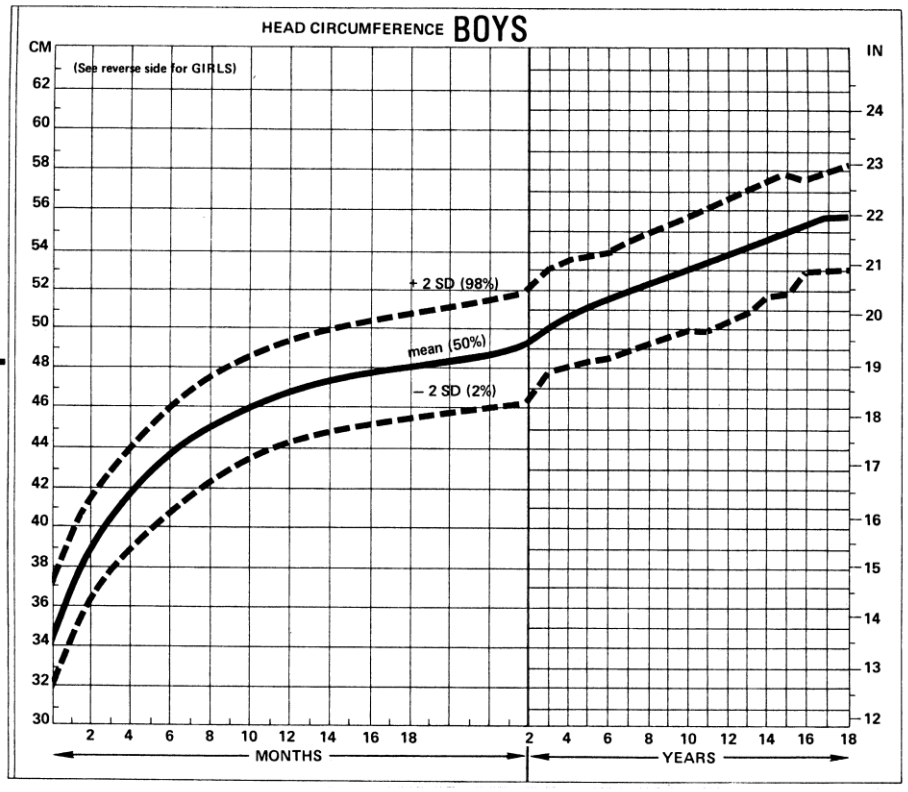
Total Score	Gestational Age, Weeks
-10	20
-5	22
0	24
5	26
10	28
15	30
20	32
25	34
30	36
35	38
40	40
45	42

50	44


Denver II
DDM, INC. 1-800-419-4729
CATALOG #2115
Examiner:
Date:
Name:
Birthdate:
ID No.:
MONTHS
YEARS
Percent of children passing
25 50 75 90
May pass by report
Footnote no.
(See back of form)
TEST ITEM
PERSONAL - SOCIAL
FINE MOTOR - ADAPTIVE
LANGUAGE
GROSS MOTOR
PREPARE CEREAL
BRUSH TEETH, NO HELP
PLAY BOARD/CARD GAMES
DRESS, NO HELP
PUT ON T-SHIRT
NAME FRIEND
WASH & DRY HANDS
BRUSH TEETH WITH HELP
PUT ON CLOTHING
FEED DOLL
REMOVE GARMENT
USE SPOON/FORK
HELP IN HOUSE
DRINK FROM CUP
IMITATE ACTIVITIES
PLAY BALL WITH EXAMINER
WAVE BYE-BYE
INDICATE WANTS
PLAY PAT-A-CAKE
FEED SELF
WORK FOR TOY
REGARD OWN HAND
SMILE SPONTANEOUSLY
SMILE RESPONSIVELY
REGARD FACE
BANG 2 CUBES HELD IN HANDS
THUMB-FINGER GRASP
TAKE 2 CUBES
PASS CUBE
RAKE RAISIN
LOOK FOR YARN
REACHES
REGARD RAISIN
FOLLOW 180°
HANDS TOGETHER
GRASP RATTLE
FOLLOW PAST MIDLINE
FOLLOW TO MIDLINE
TOWER OF 8 CUBES
IMITATE VERTICAL LINE
TOWER OF 6 CUBES
TOWER OF 4 CUBES
TOWER OF 2 CUBES
DUMP RAISIN, DEMONSTRATED
SCRIBBLES
PUT BLOCK IN CUP
6 WORDS
3 WORDS
2 WORDS
ONE WORD
DADA/MAMA SPECIFIC
JABBERS
COMBINE SYLLABLES
DADA/MAMA NON-SPECIFIC
IMITATE SPEECH SOUNDS
SINGLE SYLLABLES
TURN TO VOICE
TURN TO RATTLING SOUND
SQUEALS
LAUGHS
"OOO/AAH"
VOCALIZES
RESPOND TO BELL
SIT-NO SUPPORT
PULL TO SIT - NO HEAD-LAG
ROLL OVER
CHEST UP-ARM SUPPORT
BEAR WEIGHT ON LEGS
SIT-HEAD STEADY
HEAD UP 90°
HEAD UP 45°
LIFT HEAD
EQUAL MOVEMENTS
GET TO SITTING
PULL TO STAND
STAND HOLDING ON
STAND-2 SECS.
WALK WELL
STOOP AND RECOVER
STAND ALONE
WALK BACKWARDS
RUNS
WALK UP STEPS
KICK BALL FORWARD
JUMP UP
THROW BALL OVERHAND
BROAD JUMP
BALANCE EACH FOOT 1 SECOND
HOPS
BALANCE EACH FOOT 2 SECONDS
COMBINE WORDS
POINT 2 PICTURES
BODY PARTS-6
NAME 1 PICTURE
POINT 4 PICTURES
SPEECH HALF UNDERSTANDABLE
BALANCE EACH FOOT 3 SECONDS
BALANCE EACH FOOT 4 SECS
BALANCE EACH FOOT 5 SECS
HEEL-TO-TOE WALK
BALANCE EACH FOOT 6 SECONDS
NAME 4 PICTURES
KNOW 2 ACTIONS
KNOW 2 ADJECTIVES
NAME 1 COLOR
USE OF 2 OBJECTS
COUNT 1 BLOCK
USE OF 3 OBJECTS
KNOW 4 ACTIONS
SPEECH ALL UNDERSTANDABLE
UNDERSTAND 4 PREPOSITIONS
NAME 4 COLORS
DEFINE 5 WORDS
KNOW 3 ADJECTIVES
COUNT 5 BLOCKS
OPPOSITES-2
DEFINE 7 WORDS
THUMB WIGGLE
COPY ○
DRAW PERSON 3 PTS
COPY +
PICK LONGER LINE
COPY□ DEMONSTR.
DRAW PERSON 6 PARTS
COPY □
86%
88%
TEST BEHAVIOR
(Check boxes for 1st, 2nd, or 3rd test)
Typical
Yes
No
Compliance (See Note 31)
Always Complies
Usually Complies
Rarely Complies
Interest in Surroundings
Alert
Somewhat Disinterested
Seriously Disinterested
Fearfulness
None
Mild
Extreme
Attention Span
Appropriate
Somewhat Distractable
Very Distractable
1 2 3
©1969, 1989, 1990 W. K. Frankenburg and J. B. Dodds ©1978 W. K. Frankenburg
FOR USE OF THIS FORM, SEE AR 800-75

PATIENT INFORMATION:

Name _____
Birth Date _____
Notes:

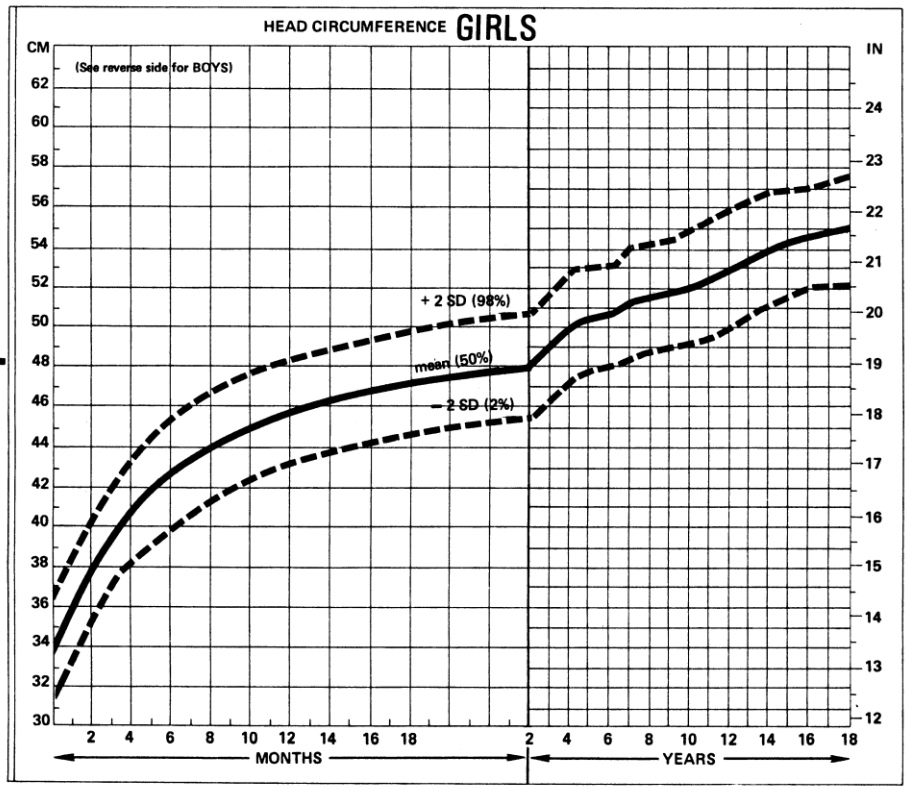


Ref: NELLHAUS, G., Composite International & Interracial Graphs, Pediatrics 41:106, 1968

BOYS

PATIENT INFORMATION:

Name _____
Birth Date _____
Notes:



GIRLS

Important Phone Numbers

Fellow Phones:
Ward x8260
PICU x8261
NICU x

2014-2015 Role Based Pagers

58260 Neurology Ward Fellow

58261 Neurology PICU Fellow

58259 Neurology Inpatient Consult Fellow

58262 Neurology NICU/ED Consult Fellow

To Page: 202-259-XXXX
Or in house: 6600 --- XXXX # --- call back #
ASCOM phones: 202-476-XXXX

Hospital Operator: 202-476-5000
- physician access to operator: 202-476-4880

Inpatient Service Impt #s
- Neurology Resident p1363
- Resident phone x8265
- Neurology Floor x5150
- Charge Nurse x8276
- Derrick(mri/f/up appts) p1044
- Emily G (case management) x8231, p4678

ER: x5203
Lifeline (transport team) x5433 (L-I-F-E)

PICU (3 east):
- Main number x 2010
- Fellow On Call x8038
- PICU residents – x8040, x8042, x8045, x8048

NICU (6 east):
- Main number x5040

- Fellow On Call x8744

Admissions: x4068 (call in any direct admissions)

EEG:
- Downstairs (techs): x5651
- Downstairs Reading Room: x5455
- 5 east Reading Room: x7105
- Tech ASCOM phone: x8263
- Nakeeta (Video EEG scheduling) x5645

MRI:
- Nurses (for scheduling, sedation ?s) x 2927
- Reading Room: x2988, 2989
- Outpatient Scheduling: x3666, x4700

Clinic: x2673
- Audrey (urgent appts) x2666
- Keisha (appointments) x2828
- Fairfax – Nancy Elling 571-226-8343 or 571-226-8316

Appointment Line (for parents) x2610

Office Staff: x2120, x2165
- Jessica x2265
- Fax Number: x2864

Long Distance Code -- 9-1-xxx-xxxx then 7880 #

IT Help Desk: x4357 (H-E-L-P)

Main Lab x 5355
Pathology Lab: x2051 on call pathologist p0784
Radiology: x8643

Other important pagers:
Laura (Dietician) p0335
Language Services: p0370 (x5444)

Expectations for Notes for General Neurology clinic:

1. Notes to be completed within 48 hours (or by the end of their month-long rotation, whichever is sooner).

2. Records should be completed with appropriate level of documentation:
- HPI should have at least 4 points
- Allergies and medications documented (including vitamins/OTC meds)
- Medication reconciliation form reviewed with family
- Birth history, past medical history, family history, social history, and developmental history reviewed
- Review of systems with at least 10 points completed
- Complete exam meeting requirements)

3. Current medications should include the following information:
- Name of medication (brand or generic)
- Formulation of medication (liquid vs. pill, concentration of liquid/strength of pill)
- Frequency and timing of when the medication is taken per day
- Calculation of mg/kg/d
- Compliance (average number of missed doses per week)

4. Plan should include mention of any prescriptions written, including:
- Name of medication (and if generic specified)
- Formulation of medication (liquid vs. pill, concentration of liquid/strength of pill)
- Frequency and timing of when the medication is taken per day
- How much med given (# pills and/or month supply, how many refills)

5. Notes for children with epilepsy should include (as per http://www.aan.com/globals/axon/assets/8092.pdf):
- Seizure type(s) and current seizure frequency (of each)
- Documentation of etiology of epilepsy or epilepsy syndrome
- EEG results reviewed, requested, or test ordered
- MRI/CT scan reviewed, requested, or scan ordered
- Querying and counseling about antiepileptic drug side effects
- Surgical therapy referral consideration for intractable epilepsy
- Counseling about epilepsy specific safety issues
- Counseling for women of childbearing potential with epilepsy
- Prior antiepileptic medications tried (and reason stopped)

6. Notes for children with headaches should include:
- Headache type(s) and current frequency (of each)
- Average length of headaches

- Impact on quality of life
- Etiology of headache (if known)
- Formal diagnosis
- Review of previous imaging done (if any)
- Triggers (if known)
- Current preventive mediation(s), compliance, side effects
- Current abortive medication(s), dosage, frequency, side effects
- Prior preventive medication tried and reason stopped
- Prior abortive medications tried and reason stopped

Neurology Call Room

The Neurology call room is located on the 4th Floor, Main Building, just outside of the neurology floor. It is labeled "Neurology" and is shared with the neurology intern.

We have two extra call rooms that are unisex with bunk beds for on call staff needing a bed on an overnight call.

Main Hospital, 4th Floor - PHAST Unit
Room 4280 (female) and Room 4278 (male)
Door Code 9375