

# Child Neurology Handbook For Pediatric Residents

Children's National Medical Center

Vice President of the Center for Neurosciences: Roger Packer, MD  
Chief of Neurophysiology and General Child Neurology: William Gaillard, MD  
Medical Unit Director, Inpatient Neurology: Elizabeth Wells, MD  
Medical Unit Director, Outpatient Services: Marc DiFazio, MD  
Neurointensive Care Director: Jessica Carpenter, MD  
Neonatal Intensive Care Director: Tae Chang, MD

Medical Director Education and Child Neurology Program Director: Marc DiSabella, DO

[mdisabel@childrensnational.org](mailto:mdisabel@childrensnational.org)

Pager: 202-259-0855

Cell: 215-262-0186

Child Neurology Program Coordinator: Laura Falcon

[lfalcon@childrensnational.org](mailto:lfalcon@childrensnational.org)

Phone: 202-476-2652

## **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.**

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### **Helpful Links:**

**MedHub:** <http://cnmc.medhub.com>

Login: neurocl

Password: Drbear111

- Includes the neurology handbook, curriculum, conference schedule, duty hours log, journal club articles, and hospital policies.
- Go to Articles/Must Read Article Database to review a compilation of important articles in each neurologic subspecialty.

**Hospital Pathways:** <http://intranet.cnmc.org/healthcare-professionals/physicians/Pages/publications.aspx#18>

**Child Neurology Society:** <http://www.childneurologysociety.org/>

**American Academy of Neurology:** <http://www.aan.com/>

# 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 running monthly mock codes and giving an evidence based medicine lecture one Friday per month
    - Seniors will be responsible for all general medical issues on the floor (pneumonia, UTI, etc)
- 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
  - This does not include 23hr vEEG, priority DC’s
  - If more than 20 patients are on neurology, fellows are to write additional notes until “cap” is lifted
- Due to 16hour maximum shift for interns and increasing supervision needed, weekends are structured as follows:
  - One of the ward interns covers the day shift on Saturday and Sunday (6AM - 6PM and 6AM - 7PM, respectively)
  - One of the ward interns covers the night shift on Saturday and Sunday (6PM - 930 AM and 7PM - 6AM, respectively)
  - This requires the neurology attending and fellow to round on wards at 8am on weekend and complete them by 930 am Saturday and Sunday
  - Neurology will have a dedicated senior during weekend rounds and the day shift on Saturday and Sunday. Fellows and Residents should check in with the PL2 (x8299) or PL3 (x8300) on weekend nights for necessary updates on sick patients.

## Neurology Rounding Format

- Rounds should occur from 9am to 11am on weekdays, and on 8am to 10am Saturday and Sunday
- Rounds should be family centered when educational as time permits. This allows for family questions to be dealt with during working rounds in a time efficient manner. Please plan time accordingly to round when service becomes large.

Rounding will be presented in the following order: Med Student - Intern - Senior - Nursing, Ancillary Services - Fellow - Attending - Family - Nursing (clarification and order read back)

## 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 4<sup>th</sup> 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 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 4<sup>th</sup> 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. 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.
9. You are responsible for performing lumbar punctures on your patient when necessary. You should coordinate anesthesia when necessary, print out lab slips and obtain tray and needles, expect the PGY-3 resident to guide you through the procedure, and then you should ensure the CSF is delivered to the lab (inform PGY-3 and fellow when it has been delivered).

#### Expectations for PGY-3 Pediatric Residents on Neurology

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. **After the attending and family have completed discussing the final details of the case, you should summarize the plan clarifying the following information on each patient every day:**
  - 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 afternoon rounds are required with on call residents, fellows, nursing, and case management, and they should start at 3:30 PM.
- 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

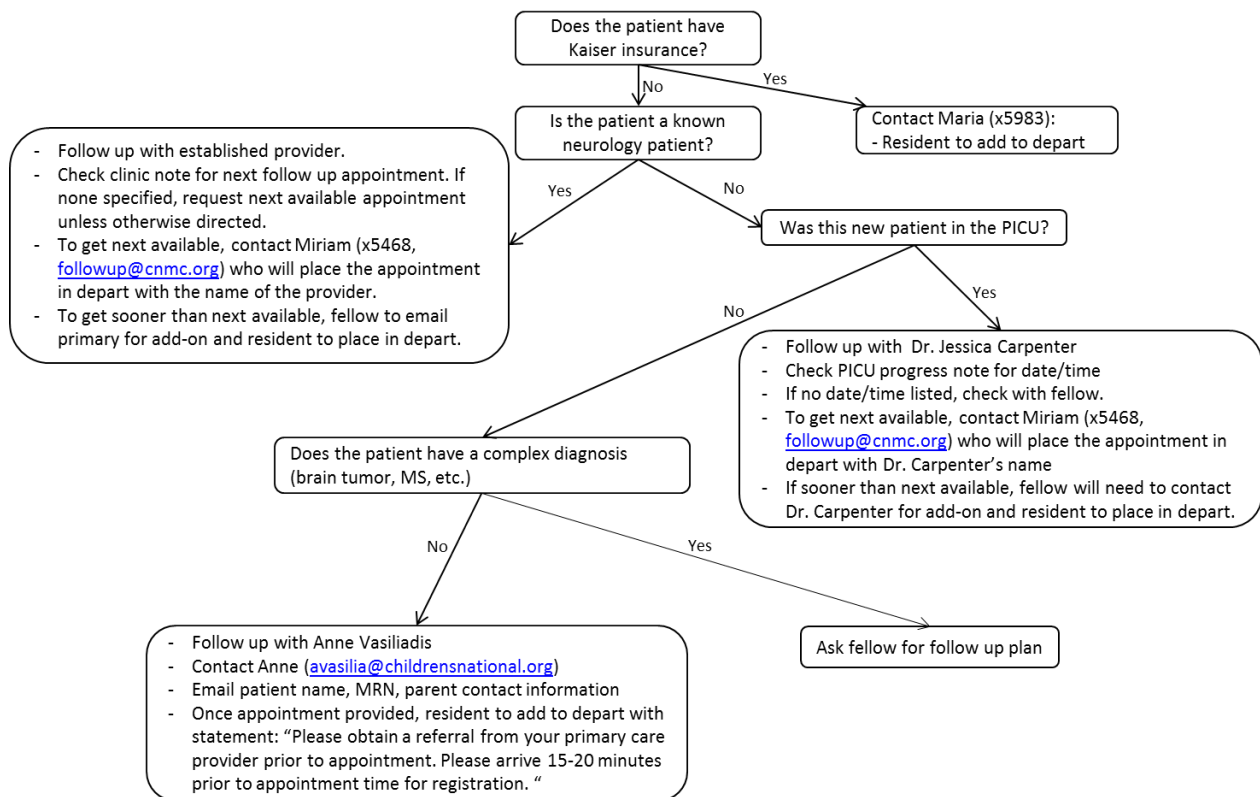
- 1) **Discharges during the week (Monday-Friday):**
  - a. Arrange follow-up with NP Anne in Neurology Clinic if patient fulfills below criteria: [AVasilia@childrensnational.org](mailto:AVasilia@childrensnational.org)
    - i. Requirements: "Complex" patients seen within 7 days, others seen within 14 days of discharge
      1. Clinical Criteria:
        - a. Spells/non-epileptic events
        - b. Complex febrile seizures
        - c. First time seizures
        - d. New onset epilepsy\*
          - i. \*Also make appointment with epileptologist (2-3 months) at time of discharge
        - e. Uncomplicated meningitis/encephalitis
        - f. Headaches \*\*

- i. **\*\*Also make appointment in headache clinic (2-3 months) at time of discharge**
  - g. Post infectious cerebellar ataxia
  - h. Anyone who only needs a one-time follow up close to discharge
- b. **Myriam Briceno, CRM Associate is your scheduling resource for making other appointments.** She is available Mon-Fri and will enter the information in Depart, so your patients have the information when they leave. Check the outpatient record for the name of the primary CNHS neurologist if the patient has one. Note in the H&P who the primary neurologist is and make the appointment request on day one of admission.

**2) Discharges during weekends/holidays:**

- a. Please email patient information including (patient name, MR, DOB, family phone, requested follow-up, reason for follow-up, and when you would like follow-up) to: [followup@cnmc.org](mailto:followup@cnmc.org) with the request and relevant information

Congratulations!! Your patient is ready for discharge. Please follow the steps below to schedule a follow up.



- **On weekends/holidays/after hours**, please still email Miriam or Anne for follow up. Anne will get back to you in real time so that you can include the appointment in the depart and Miriam will schedule the appointment and contact the family during the week.
- If family is not contacted for appointment, instruct them to call the appointment line **202-476-3611** NOT the neurology clinic.

## 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, review information on new neurology service admissions and updated clinical information on current service patients.
  - c. Review new imaging and lab results.
  - d. 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 can be performed prior to rounds or during rounds, except in cases where examination will alter management in which case this is always done urgently. If there are abnormalities, 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 abnormalities.
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).
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 3:30 PM 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 should be completed before the end of the day sign out for attendings, which is at 5pm. It is strongly encouraged to have notes signed and forwarded to attendings immediately after rounds.
- 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 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. In conjunction with residents, follow up any testing that is pending upon discharge through use of a 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.
- 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 at least twice per week.
- 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 should not plan to have time to perform a complete H+P on every patient called, 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. It is best to have adult neurology trainees be primarily responsible for consults in the ER and floors. You should act to oversee this and help answer questions, but the child neurology fellow's primary responsibility is with patients on the neurology service.
  - c. 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.
  - d. 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,
  - b. Tuesdays and Fridays, following the neurology lecture schedule for the pediatric residents' neurology rotation.
  - c. residents' neurology rotation.
  - d. On Wednesdays at 8 AM, Pediatric Grand Rounds attendance is required for pediatric residents and students, and they are also required to attend Neurology
  - e. Sunrise Lecture on Thursdays at 7 AM.
- 12. Contact nurse managers (Audrey Scully) or schedulers to arrange urgent appointments requested by outside physicians or by the rotator on call the night before

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 practitioner is unavailable, 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.

#### 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 (max 28hrs)
  - a. Rounds are family centered. This means that the family is invited to witness and participate in rounds. The family should 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, Tuesday, and Friday 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 fellow on service. You should attempt to be present for all three of these weekly lectures when time permits.
3. Consult rounds should be done immediately after wards at 11AM when possible, however based on fellow availability, may be moved to the afternoon when 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 of 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 radiographic 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 3:30 PM 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.

## Section 2 -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
<b>Week 1</b>					
	830-9am Neurologic Exam (required)	Morn Report  830-9am Localization (required)		7-8am Sunrise Conf (Neuro conf room - required)  1-2pm Intake Conf (optional)	830-9am Endocrinology Lecture
<b>Week 2</b>					
	830-9am Cerebral Palsy (required)	Morn Report  830-9am Acute Encephalopathy (required)		7-8am Sunrise Conf (Neuro conf room - required)  1-2pm Intake Conf (optional)	830-9am Endocrinology Lecture
<b>Week 3</b>					
	830-9am Increased Intracranial Pressure (required)	Morn Report  830-9am Tics and Tourette Syndrome (required)		7-8am Sunrise Conf (Neuro conf room - required)  1-2pm Intake Conf (optional)	830-9am Endocrinology Lecture
<b>Week 4</b>					
	830-9am Neurocutaneous Syndromes (required)	Morn Report  830-9am Neuromuscular Disorders (required)		7-8am Sunrise Conf (Neuro conf room - required)  1-2pm Intake Conf (optional)	830-9am Endocrinology Lecture

## 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 resistance
    - vi. 5- normal strong movement
  - b. Abnormalities and cause:
    - i. proximal weakness - myopathy
    - ii. distal weakness - neuropathy
    - iii. unilateral weakness - UMN lesion including 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.

- Hypercoagulability 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 if 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. Electrolytes
  - 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. Automatism: 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 (~1/3<sup>rd</sup> of patients)
  - c. EEG: generalized 4-6Hz polyspike 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. Oropharyngoguttural 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 epigastric 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 neuropsychological 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 1<sup>st</sup> 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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John M. Pellock et al. "Infantile spasms: A U.S. consensus report" Epilepsia, 51(10):2175-2189, 2010

Christian M. Korff and Douglas R. Nordli, Jr. Paroxysmal Events in Infants: Persistent Eye Closure Makes Seizures Unlikely Pediatrics 2005;116:e48

## 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 than 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 2<sup>nd</sup> 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 2<sup>nd</sup> 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 :** (Carbamazepine), oxcarbazepine, phenytoin  
2<sup>nd</sup> line: lacosamide, topiramate, Valproate, levetiracetam

**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)  
2<sup>nd</sup> 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:
  - o cognitive slowing (“dopomax”)
  - o decreased appetite (weight loss)
  - o 1-2% can get kidney stones because renal excreted
  - o Metabolic acidosis : carbonic anhydrase inhibitor (caution with Keto diet and some patients experience decreased sweating)

#### Levetiracetam: Keppra

- Broad spectrum
- SE:
  - o 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 than carbamazepine and fewer drug interactions
- SE:
  - o Hyponatremia
  - o SIADH (not dose dependent and very rare)
  - o 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:
  - o Also bad for teenage girls when used long term: Hirsutism, gum hypertrophy, Teratogenic (craniofacial anomalies),
  - o Ataxia
  - o nystagmus
  - o stevens-johnson
  - o Anxiety and aggression
  - o Soft tissue injury if IV form infiltrates \*\*
  - o 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:
  - o Hyperactivity
  - o Irritability
  - o Lethargy
  - o Hypotension
  - o Long term effects on learning and cognition
- Enzyme INDUCER: decreases the levels of other drugs
- Lots of drug to drug interactions
- Very long t<sub>1/2</sub>

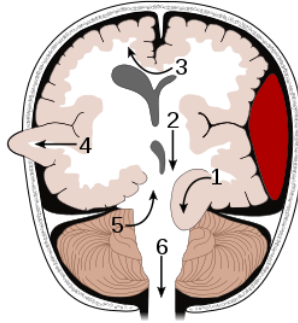
| Ethosuximide

- 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 cingulate 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<sup>th</sup> 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 be 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 comorbidities 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 result in tics and other neurologic abnormalities, such as dystonia or encephalopathy, include pantothenate kinase associated neurodegeneration (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](http://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:
  - o six of more cafe au lait >5mm if prepubertal, 15mm if postpubertal,
  - o axillary or inguinal freckling,
  - o 2 or more iris lisch nodules (hamartomas seen with slit lamp, rare in children <3yo but in 100% of >21yo),
  - o 2 or more neurofibromas or 1 plexiform neurofibroma typically in skin with small rubbery purple color,
  - o distinctive osseous lesion such as sphenoid dysplasia (scoliosis common but does not count),
  - o optic glioma (in 15% of pts),
  - o 1st degree relative with NF
- 2 NF 2 is classified as follows:
  - o Confirmed (definite) NF2: Bilateral vestibular schwannomas
  - o 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
  - o Typically present with learning problems and neurocutaneous stigmata
  - o 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
  - o HTN, precocious puberty
- 4 Pathogenesis
  - o due to defect in NF1 gene which codes for neurofibromin protein, a tumor suppressor gene
  - o also in that intron is oligodendrocyte myelin a gene which results in abnormal myelination of the cortex seen as white matter abnormalities on MRI
  - o NF 2 is typically due to mutation at center of long arm of 22q11
- 5 Management
  - o Multidisciplinary approach including neurology, PM+R, ophthalmology, nephrology, oncology
  - o Should have yearly history and physical exam and ophtho exam

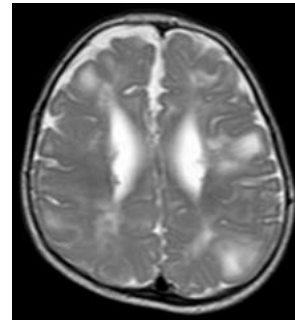
- o Investigation of any new or acute CNS symptoms immediately with MRI/CT



## 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 (SEs), 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. SEs 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 SEs 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 adrenocorticotrophic 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.

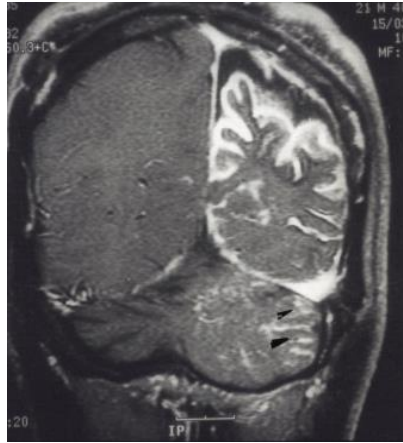


### 3. Sturge Weber

- 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 leptomenigeal angiomas 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 letomenigeal 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 arthrogyposis

### 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](http://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 function in 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 (1<sup>st</sup> Floor between the main lobby welcome desk and ER). Clinics generally start at 9am and end by 5pm except as noted below. The following schedule is based on calendar month.*

Weeks 1, 3, 5		TUESDAY		WEDNESDAY		THURSDAY		FRIDAY	
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		TUESDAY		WEDNESDAY		THURSDAY		FRIDAY	
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

# Section 5: Appendix



## Emergency Department Pathway

Revised 7-17-07

### Status Epilepticus

#### 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<sup>+</sup>, Ca<sup>++</sup>), 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,  
phenobarbital 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.

# 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.<sup>1</sup>

<b>DIASTAT® AcuDial™ (diazepam rectal gel)</b> <b>Dosing Recommendations (by age and weight)</b>					
<b>2 to 5 years</b> <b>0.5 mg/kg</b>		<b>6 to 11 years</b> <b>0.3 mg/kg</b>		<b>12+ years</b> <b>0.2 mg/kg</b>	
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.diaostat.com](http://www.diaostat.com).

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



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D961-0508 Printed in USA

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



## 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 cystercosis 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

## 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  $\geq$  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  $\leq$  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  $\leq$  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.



## Emergency Department Pathway SEIZURE WITH FEVER

Revised 2-1-06

### 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  $\geq 38.0^{\circ}\text{C}$  *or* Oral  $\geq 37.6^{\circ}\text{C}$ )

*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



## Emergency Department Pathway Migraine Headache

Revised 7/20/07

### INCLUSION CRITERIA

- A. Headache lasting 1-72 hours
- B. Headache has  $\geq 2$  of the following characteristics –
  1. Unilateral location, may be bilateral, frontotemporal (not occipital)
  2. Pulsing quality
  3. Moderate or severe pain intensity
  4. Aggravated by or causing avoidance of routine physical activity.
- C. During headache at least 1 of the following occur-
  1. Nausea/Vomiting or both
  2. Photophobia or phonophobia, which may be inferred from behavior
- D. HA not attributed to another disorder (see below)

### Exclusion Criteria:

Trauma, Pregnancy, Focal neurological deficit, Abnormal head CT, Complicated migraine Hypertension, Papilledema

### TRIAGE RN

1. Assign ESI score
2. Expedite placement of patient in treatment room if score 1 or 2
3. Make Patient NPO

### INITIAL NURSING CARE

1. Document vital signs and pain assessment
3. Urine hcg in menarchal female
4. Provide comfort measures
5. Reassess Pain after therapy

### Physician

1. Examine patient and determine eligibility for pathway
2. Order Medications as below:
  - a. Patients 6-11 years old:
    - Ibuprofen 10 mg/kg/dose PO (max dose = 600 mg) or
    - If failed NSAIDS at home: Ketorolac (Toradol) 0.5 mg/kg IV (max dose =30 mg)
    - If nausea vomiting present: Metoclopramide 0.15 mg/kg/dose IV (max=10 mg/dose)
3. Patients  $> 11$  years old:
  - a. Sumatriptan (Imitrex) 0.06 mg/kg SQ (max 6 mg) **If no risk factors-see below)**
  - or
  - b. **If patient has risk factors for Triptans:**  
(Has already tried a Triptan/Ergot derivative at home, history, signs or symptoms of ischemic, cardiac, cerebrovascular or peripheral vascular disease syndromes, use of an MAOI agent within 2 weeks, Hemiplegic/basilar migraine, severe hepatic impairment, Sickle Cell Disease, ischemic bowel disease)  
Metoclopramide 10mg IV Q30mins up to 4 times  
Diphenhydramine 1 mg/kg IV (max = 25 mg) prior to dose 1 and 3
  - c. If fails a and b, or a and b contraindicated:  
Valproic acid 10 mg/kg/dose PO / IV (max 1000mg/dose), may repeat in 1 hour



# Pediatric Acute Encephalopathy/Encephalitis Diagnostic Pathway

April 2013, Page 1

### Nursing Care

- Seizure precautions
- Compression stockings and/or medical DVT prophylaxis if post puberty
- NPO until emergent diagnostic studies complete
- Aspiration precautions
- Neuro checks Q1 hour for at least first 24 hrs
- Bed rest (Fall precautions)
- Total Care Sports Specialty bed

### Supportive Care

- PT/OT consult
- Encephalopathy > 48 hrs, consult PM&R
- Ophthalmology consult (no drops for 1st 48hrs)

### Additional Medications

- Seizures (see AEDs for ICU):**
- Lorazepam 0.1 mg/kg (max 2 mg/ dose) PRN seizure
  - Fosphenytoin 20 mg PE/kg x1 with maintenance 5mg/kg/d div BID
  - Phenobarbital 20mg/kg x1
  - Versed gtt 0.1mg/kg/hr titrate to seizure suppression
- Fever:**
- Acetaminophen 15mg/kg/dose (max 4g/24h) PRN T>37.5
  - Cooling blanket
- ABX/antivirals (CNS dosing):**
- Ceftriaxone 50 mg/kg IV q12 hrs
  - Vancomycin 10 mg/kg IV q6 hrs
  - Acyclovir 10mg/kg/dose IV q8 hrs
- Immunotherapy (Recommend discussion w/ ID and Neuro before giving):**
- Steroids
  - IVIG 2g/kg over 2 days (obtain all titers prior to 1st dose)

### Laboratory Studies

- Call lab to prioritize studies, freeze extra CSF

### Diagnostic Guidelines

- Neuroimaging:**
- HCT- initial study without contrast
  - MRI brain with and without gad
  - MRS if suspicion for metabolic process
- Electrophysiology:**
- Continuous vEEG for at least 48 hrs or until awake

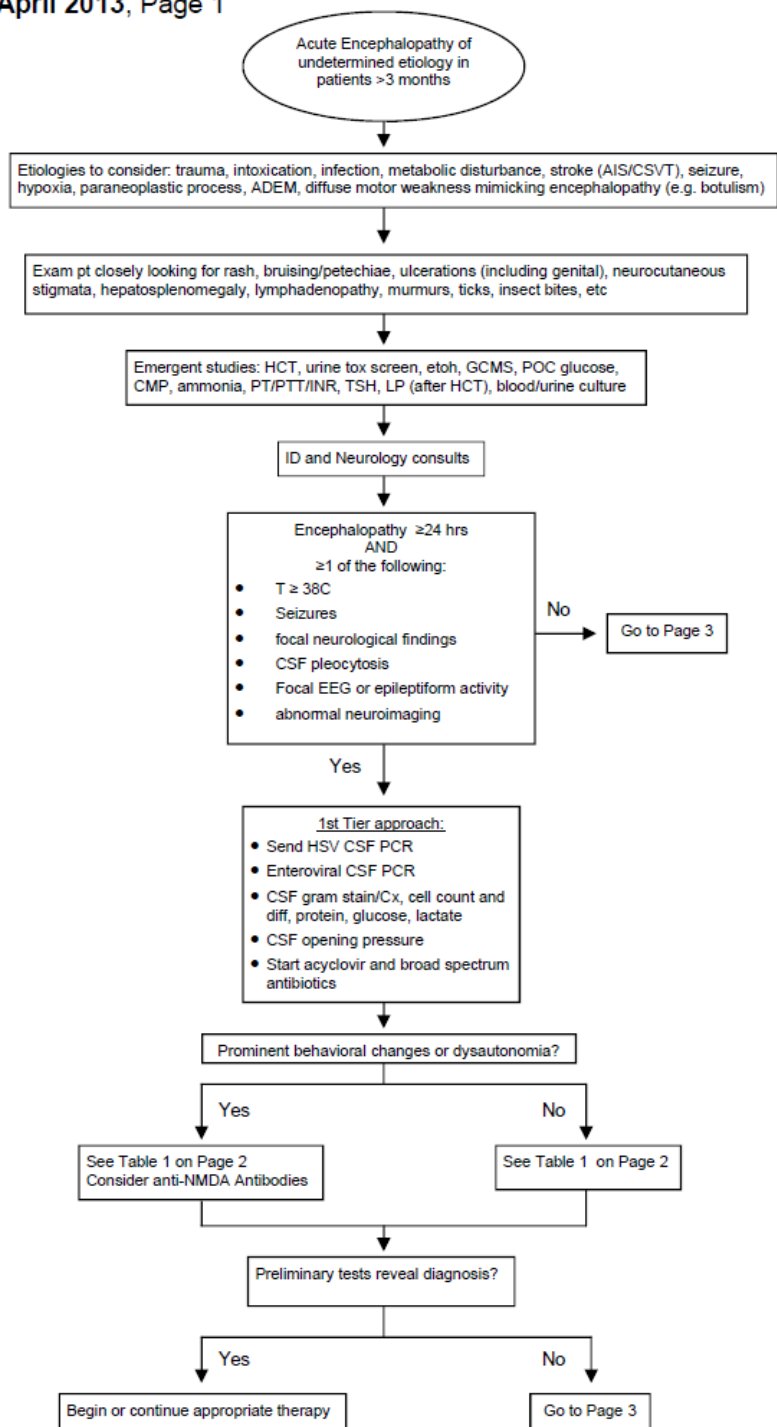




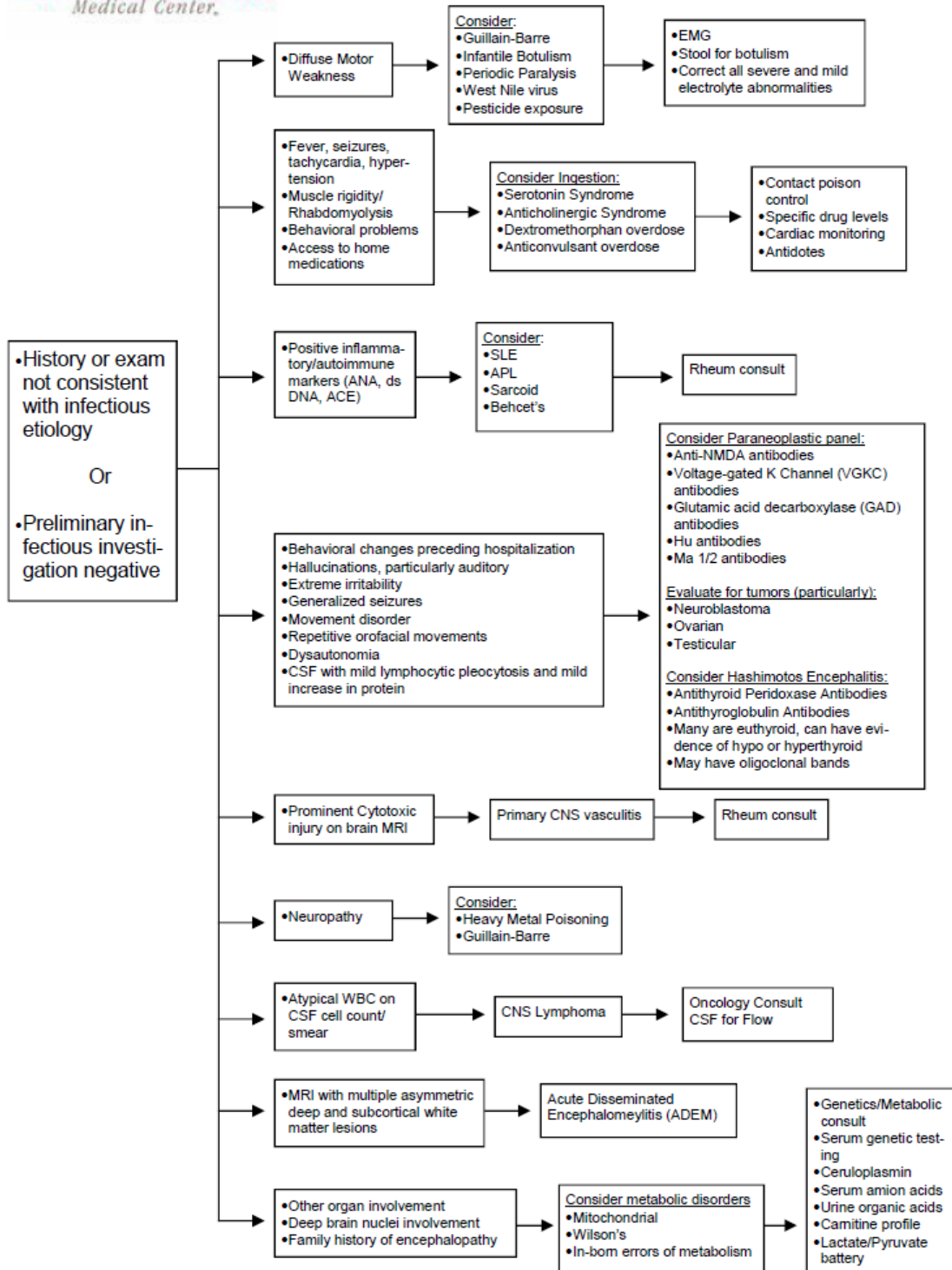
Table 1– Diagnostic Tests for Infectious Causes of Encephalitis

Organism	Blood	CSF	Other
<b>1st Priority*</b>			
Herpes Group (HSV, VZV, EBV, CMV)	EBV, CMV serology, VZV serology	HSV, VZV, EBV, CMV PCR	HSV PCR of skin lesions, VZV DFA and PCR of skin lesions Ophtho exam if suspected CMV <b>CMV PCR (CSF), EBV PCR (CSF) must be ordered misc</b>
Enterovirus	PCR	PCR	NP and rectal PCR
Arbovirus (West Nile, La Crosse, EEE, WEE, SLE)**	WNV serology***, arboviral serology,	WNV serology*** Arboviral serology	<b>Arboviral IgM (CSF) must be ordered misc</b>
Respiratory viruses			Nasal Respiratory Viral PCR, CXR
Mycoplasma	serology	PCR	NP PCR, CXR
<b>2nd Priority*</b>			
Bartonella	serology		Ophtho exam if suspected
HIV	serology and DNA PCR	DNA PCR	<b>HIV DNA PCR (CSF) must be ordered misc</b>
Lyme	EIA screen serology with reflex WB	EIA screen serology with reflex WB	Ophtho exam if suspected
Mycobacterium TB		PCR, AFB smear and culture	PPD, CXR, PCR and culture of respiratory secretions <b>PCR (CSF) must be ordered misc</b>
Rickettsia**– RMSF, Ehrlichia, Anaplasma	RMSF serology, Ehrlichia serology, anaplasma serology	CSF PCR's all low yield	Review blood smear for morula if suspected anaplasma
Parvovirus		PCR	
<b>3rd Priority*</b>			
Malaria	Thin and Thick Smears		
Rotavirus			Stool/rectal rotavirus antigen
Parvovirus	serology and PCR	PCR	
Rabies			Conjunctiva and Saliva PCR
Syphilis	VDRL or RPR w/ FTA-ABS if +	CSF VDRL	
Endemic Fungi- Histoplasma, Coccidioides		Histo antigen and serology	Urine Histo antigen
Amoeba– Naegleria, Balamuthia	Balamuthia serology (IFA) via CDC	Naegleria wet prep	<b>Balamuthia, Naegleria (Cx), Amoeba Wet Prep must be ordered misc</b>
<b>Immunocompromised Host</b>			
HHV-6		PCR	
West Nile Virus		PCR	
Cryptococcus	Cryptococcal Antigen	Cryptococcal Antigen	CXR
JC-Virus		JCV PCR	
Toxoplasma	Toxoplasma serology	Toxo PCR	Ophtho exam
Amoeba– Acanthamoeba		Wet prep	<b>Must be ordered as misc test</b>

\* Priority may change based on history, exposures and clinical exam. Please confirm priority with ID and Neurology consultants prior to ordering

\*\* Repeat serologic testing recommended 10-21 days after initial presentation

\*\*\* WNV is not included in the arboviral panel (although it is an arbovirus)





## Ventricular Shunt Malfunction Diagnostic Algorithm

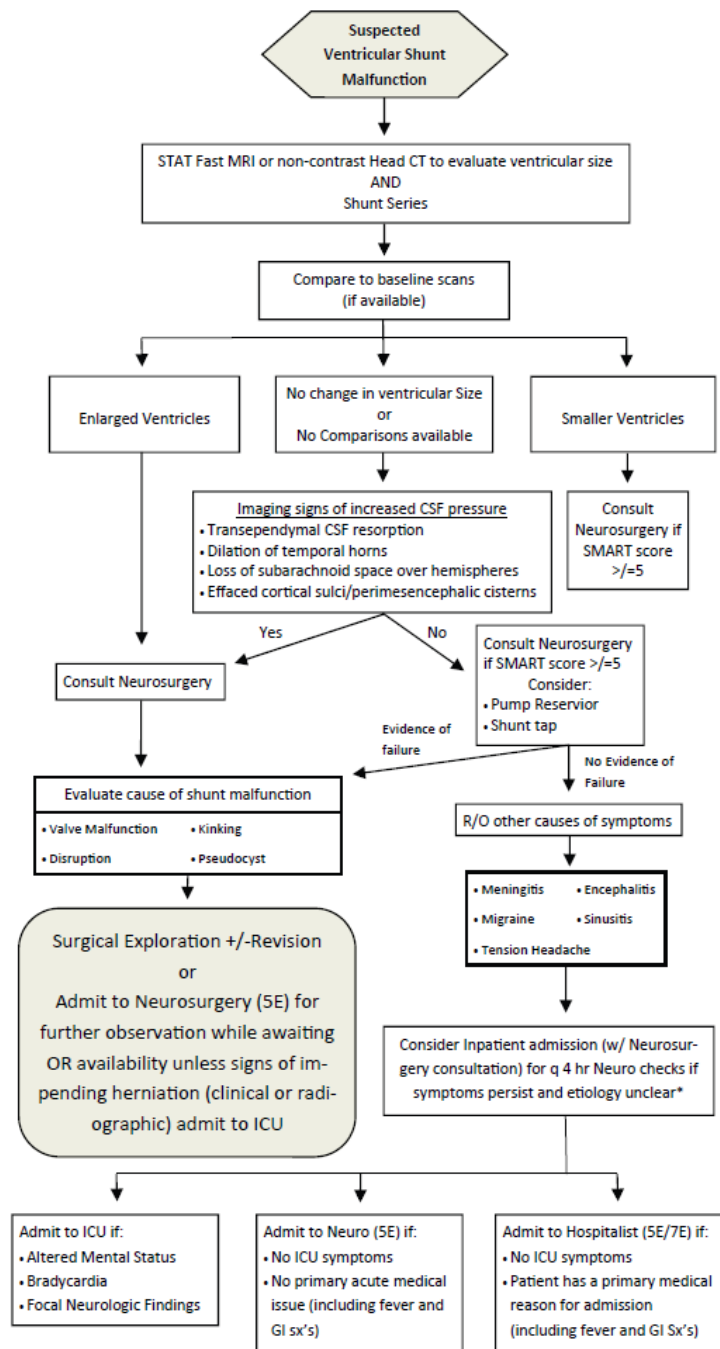
June 2013

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	



\*Neurosurgical Attending notification at time of admission is expected



**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  $\geq 4$  consider repeat imaging Q2-3 days and/or ophthalmology evaluation for papilledema, even if other explanations plausible for observed symptoms.

**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 D5 NS +20 mEq/L potassium chloride (Or 20mEq/L potassium acetate) at 1.5 maintenance
4. 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
5. 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 + 2<sup>nd</sup> medication for their baseline seizures

### Maintenance Management for MELAS

1. Resume home medications at those doses (otherwise use below)
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
3. 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)
4. Vitamin C: 25 mg/kg/day
5. Ubiquinone or CoQ10 (often home med): 5 mg/kg/day (<20 kg), 10 mg/kg/day (>20 kg)
6. Vitamin E 22.5 IU/kg/day

**Dubowitz/Ballard Exam for Gestational Age**  
**Neuromuscular Maturity**

	-1	0	1	2	3	4	5
Posture							
Square Window							
Arm Recoil							
Popliteal Angle							
Scarf Sign							
Heel to Ear							

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	Well-curved pinna, soft but ready	Formed and firm, with	Thick cartilage, ear stiff	

			slow recoil	recoil	instant recoil	
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

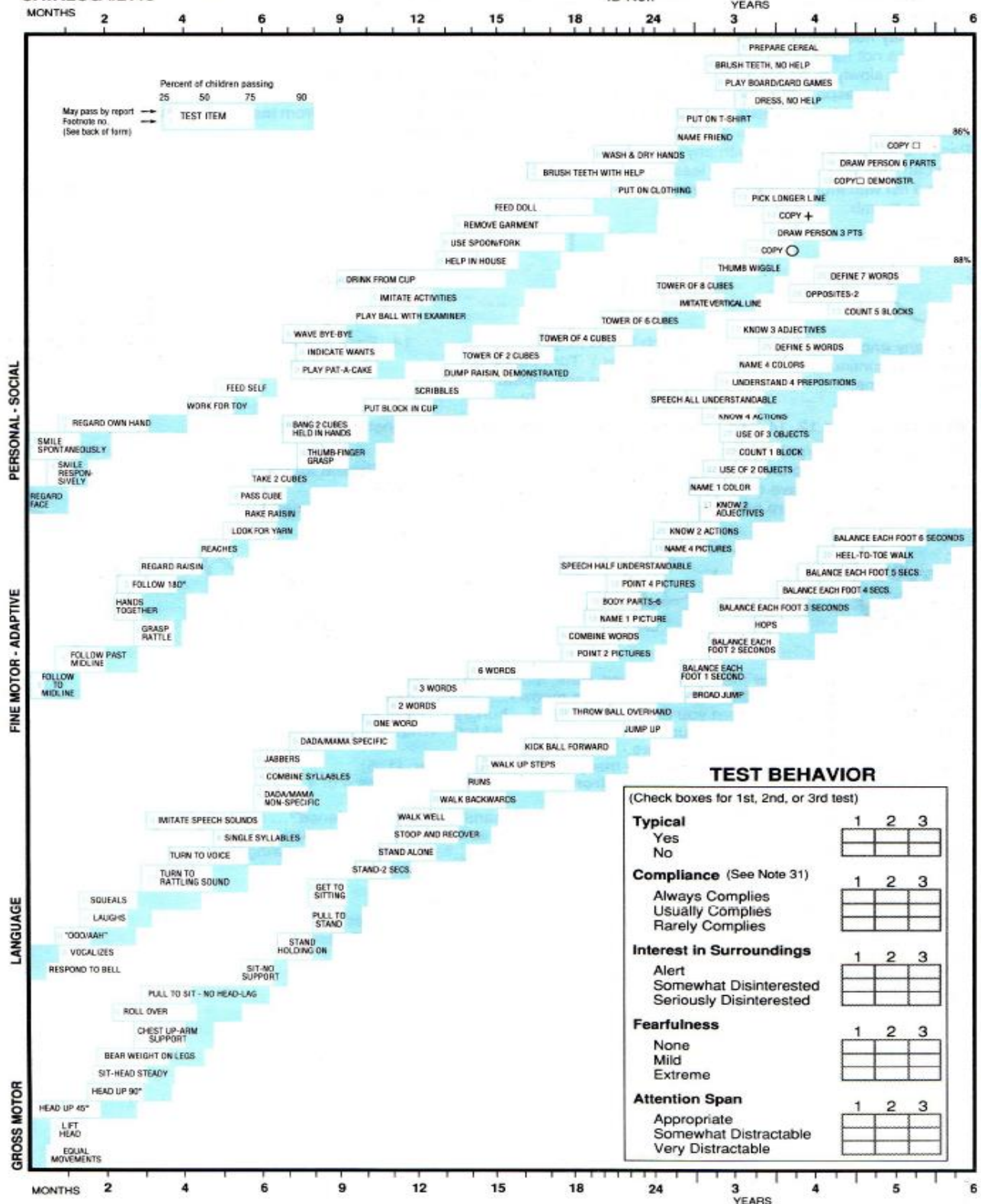


# Denver II

DDM, INC. 1-800-419-4729  
CATALOG #2115

Examiner: \_\_\_\_\_  
Date: \_\_\_\_\_

Name: \_\_\_\_\_  
Birthdate: \_\_\_\_\_  
ID No.: \_\_\_\_\_

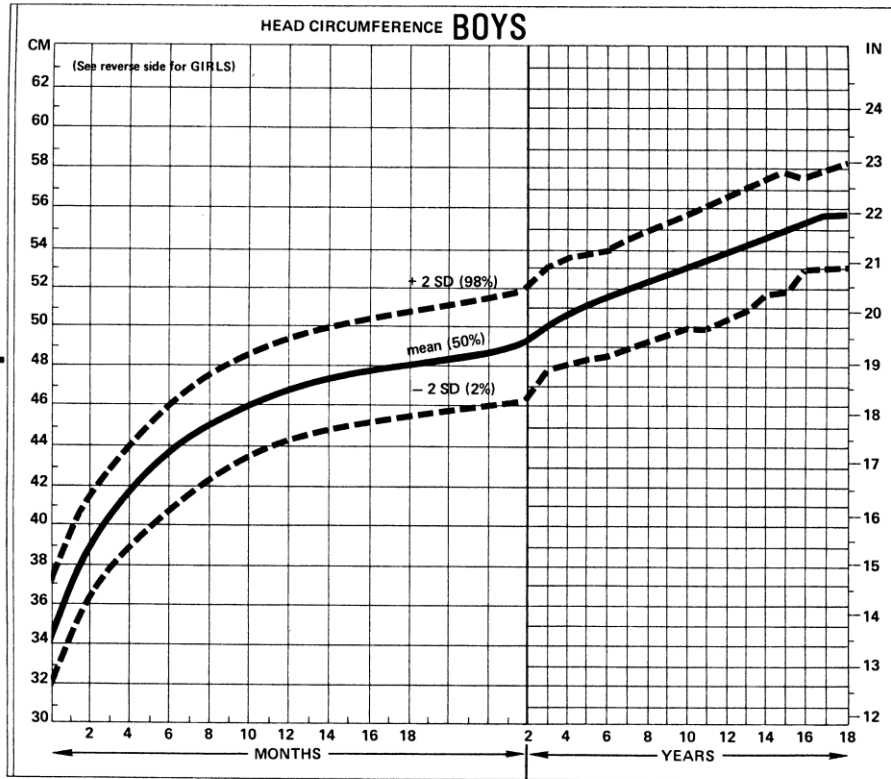


FOR USE OF THIS FORM, SEE AIR 600-75

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PATIENT INFORMATION:

Name \_\_\_\_\_  
Birth Date \_\_\_\_\_  
Notes \_\_\_\_\_

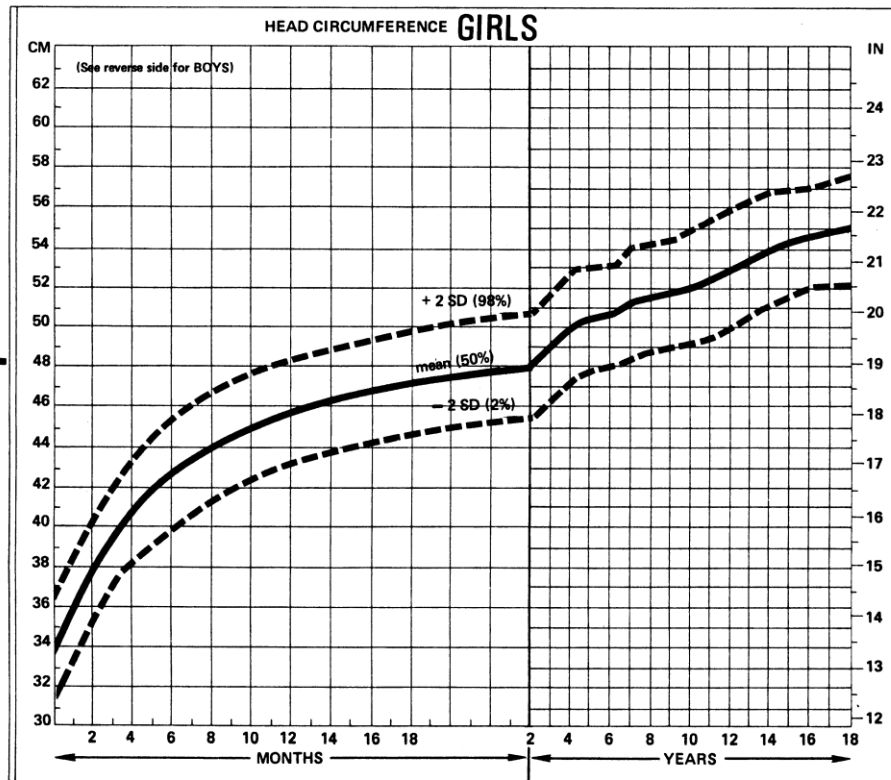


Ref: NELLHAUS, G., Composite International & Interracial Graphs, Pediatrics 41:106, 1968

BOYS

PATIENT INFORMATION:

Name \_\_\_\_\_  
Birth Date \_\_\_\_\_  
Notes \_\_\_\_\_



GIRLS

## Important Phone Numbers

### Fellow Phones:

Ward x8260  
PICU x8261  
NICU x8259

### Role Based Pagers - 202-476-6600

58260 Neurology Ward Fellow

58261 Neurology PICU Fellow

58259 Neurology Inpatient Consult Fellow

58262 Neurology NICU/ED Consult Fellow

To Page: 202-259-XXXX

Role based pagers: 202-476-6600

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:

Michiko p4696

Laura (Dietician) p0335

Language Services: p0370 (x5444)

<b>NEUROLOGY DEPARTMENT PHONE LIST</b>			
<b>Phone: 202-476-2120</b>		<b>Fax: 202-476-2864</b>	

PHYSICIANS	EXTENSION	PAGER	MANAGERS
Acosta, Maria	5513	0458	Villongco, Joe
Avery, Rob	5686	2875	Favor, Celeste
Carpenter, Jessica	4657	7017	Saunders, Steve
Chang, Taeun	6195 (DC)	0170	Wyatt, Sarah
Civitello, Lucy	2962	8268	Tracey Church
Conry, Joan	3205	8269	<b>EXECUTIVE A</b>
Depositario-Cabacar, Dewi	6739	0249	Ralph, Abby (
Disabella, Marc	3826	0855	<b>ADMINISTRAT</b>
Difazio, Marc	301-765-5633 / 240-389-4056 C	0095	Begazo, Grace
Freilich, Emily	4830	1233	Ceja, Jessica
Gaillard, William	5224	8272	Brailsford, Jo
Gropman, Andrea	3511	1612	Perlman, Linc
Nguyen, Thanh	3099	0.016	DaCosta, Nata
Kao, Amy	5932	2188	Nahabedian, S
Lateef, Tarranum	4656	1354	<b>SOCIAL WORK</b>
Lavenstein, Bennett	6929	8417	Cushner-Wein
McClintock, William	6928	8418	Mehlman, Sus
Nguyen, Thanh	4830	0016	<b>RESEARCH AS</b>
Packer, Roger	5997	8274	Julian, Percy
Reese, James	4010	2258	Stephens, Car
Scafidi, Joseph	6144	1217	Belaineh, Tek
Thangarajh, Mathula	2771	5943	Kebede, Tewo
Tsuchida, Tammy	5949	0136	Barrios, Emily
Weinstein, Steve	3514	0533	Gable, Chris (
Vanderver, Adeline	4959	8878	Stephans, Cha
Wells, Beth	4836	1588	Malik, Sabrina
Zelleke, Tesfaye	4309	1328	Pamu, Swapn
			Hostetter, Sa
<b>FELLOWS/RESIDENTS</b>	8260, 8261		Helman, Guy
			Chatterjee, S
Kurz, Jon	3512	4480	Harris, Kari (C
Fellows Room	6591		<b>ADMISSIONS</b>
Bernier, Raquel, MD	6591	2514	In-patient -L
Kahn, Ilana	6569	2519	Surgical Posti
Demarest, Scott	2658	1631	<b>APPOINTMEN</b>
<b>NURSES &amp; PA</b>			FFX
Albert, Julie (Packer)	2359	3737	MMC/Rockvill
Dodge, Leslie	8315	N/A	LLC
Dome, Lauren (Difazio)	301-765-5465	1945	<b>Concussion C</b>

Elling, Nancy	8316	8051	Keto Diet- La
Gelabert, Ana	8373	0239	Neuropsychol
Havens, Katie (Gaillard)	3514	1159	Sleep Lab- Da
Kolodgie, Marian	2649	8780	<b>MRI</b>
Lendenmann, Michiko	3987	4696	Prior Auth
Leach, Meganne (Neuromuscular)	6392	2240	Pior Auth
(Rockville)	301-765-5469		<b>VEEG/Sedate</b>
Murphy, Jennifer (Vanderver, Gropman)	5243	3084	Accounts Paya
Scully, Audrey	2666/3175 f	7932	BearTime/Pay
Vasiliadis, Anne	4010/6274 (Ascom)	1393	BILLING & Cus
Shibuya, Peter (Acosta)	4598	2017	Billing Office
Sullivan, Meg	8377	7311	Check-in desk
Fairfax Dr. Offices	571-226-8372	8346f	Concierge Ser
Fairfax Emergency Number	571-226-8336	8346f	<b>EEG Reading</b>
<b>Coordinators: Neuro, NF, Genetics</b>			EEG/EMG Tec
Pappa, Belen (Gropman)	6249	1118	Engineering
Pizzino, Amy (Vanderver)	4975	2876	<b>Environment</b>
Trimboli - Heidler, Carm (Avery)	3633	4781	Human Resou
Smart, Kathleen (NeuroMuscular)	6193	3975	International
<b>Miscellaneous</b>			<b>IR (IT)- Infor</b>
Pager System (CNMC)- 6600	NIH Pager- 301-496-1211	AV:19027	<b>Lab Medicine</b>
Welcome Desk_Operator - 3000_5000	QuestAcct#: 19662; 1-800-336-3718		Lab:SendOut
ParkingOffice_ SecurityOffice - 5457_2065	LabCorpPhone#: 703-742-3100, 5, 1		Language Ser
IR: Cedric_Bruce: 6441_6108	LabCorpAcct#: 08555320		Mailroom_Rec
<b>Willie Scruggs- x4571 pager: 0178 CANON Copier</b>	<b>Craig (HIPPA Bin): 202-213-0381</b>		<b>MEDICAL REC</b>
Courier Services: 301-683-9009	Capital Cab: 202-398-0500		Medical Staff
<b>Neurology Team Room- 7104 (5-East)</b>	Hoyt Wright (Locksmith/Keys) - 3233		<b>MRI Library_</b>
<b>FLOORS</b>	<b>EXTENSION</b>		Occupational
<b>NICU</b>	<b>5040</b>		Physician Acc
<b>PICU</b>	<b>2010</b>		<b>RADIOLOGY -</b>
<b>Tax ID: 530-196-580</b>			<b>Referrals</b>
Updated: 6.5.14 J.Brailsford			Volunteer Ser


Neurology Attending List with emails:

Name	Email	Pager	Extension
Acosta, Maria	<a href="mailto:macosta@childrensnational.org">macosta@childrensnational.org</a>	0458	5513
Avery, Rob	<a href="mailto:ravery@childrensnational.org">ravery@childrensnational.org</a>	2875	5686
Cabacar, Dewi	<a href="mailto:dcabacar@childrensnational.org">dcabacar@childrensnational.org</a>	0249	6739
Carpenter, Jessica	<a href="mailto:jcarpent@childrensnational.org">jcarpent@childrensnational.org</a>	7017	4657
Chang, Taeun	<a href="mailto:tchang@childrensnational.org">tchang@childrensnational.org</a>	0170	6195 (DC)
Civitello, Lucy	<a href="mailto:lcivitel@childrensnational.org">lcivitel@childrensnational.org</a>	8268	2962
Conry, Joan	<a href="mailto:jconry@childrensnational.org">jconry@childrensnational.org</a>	8269	3205
Disabella, Marc	<a href="mailto:mdisabel@childrensnational.org">mdisabel@childrensnational.org</a>	0855	3826
Freilich, Emily	<a href="mailto:efreilich@childrensnational.org">efreilich@childrensnational.org</a>	1233	4830
Gaillard, William	<a href="mailto:wgaillar@childrensnational.org">wgaillar@childrensnational.org</a>	8272	5224
Bharucha-Goebel, Diana	<a href="mailto:dbharuch@childrensnational.org">dbharuch@childrensnational.org</a>	5483	none
Gropman, Andrea	<a href="mailto:agropman@childrensnational.org">agropman@childrensnational.org</a>	1612	3511
Kao, Amy	<a href="mailto:akao@childrensnational.org">akao@childrensnational.org</a>	2188	5932
Lavenstein, Bennett	<a href="mailto:blavenst@childrensnational.org">blavenst@childrensnational.org</a>	8417	6929
McClintock, William	<a href="mailto:wmcclint@childrensnational.org">wmcclint@childrensnational.org</a>	8418	6928
Nguyen, Thanh	<a href="mailto:tnguyen@childrensnational.org">tnguyen@childrensnational.org</a>	0016	3099
Packer, Roger	<a href="mailto:rpacker@childrensnational.org">rpacker@childrensnational.org</a>	8274	5997
Reese, James	<a href="mailto:jreese@childrensnational.org">jreese@childrensnational.org</a>	2258	4010
Scafidi, Joseph	<a href="mailto:jscafidi@childrensnational.org">jscafidi@childrensnational.org</a>	1217	6144
Schreiber, John	<a href="mailto:jschreib@childrensnational.org">jschreib@childrensnational.org</a>	5837	none
Thangarajh, Mathula	<a href="mailto:mthangar@childrensnational.org">mthangar@childrensnational.org</a>	5943	2771
Turnacioglu, Sinan	<a href="mailto:sturnaci@childrensnational.org">sturnaci@childrensnational.org</a>	0233	none
Tsuchida, Tammy	<a href="mailto:ttsuchid@childrensnational.org">ttsuchid@childrensnational.org</a>	0136	5949
Weinstein, Steve	<a href="mailto:sweinste@childrensnational.org">sweinste@childrensnational.org</a>	0533	3514
Vanderver, Adeline	<a href="mailto:avanderv@childrensnational.org">avanderv@childrensnational.org</a>	8878	4959
Wells, Beth	<a href="mailto:ewells@childrensnational.org">ewells@childrensnational.org</a>	1588	4836
Zelleke, Tesfaye	<a href="mailto:tzelleke@childrensnational.org">tzelleke@childrensnational.org</a>	1328	4309

MRI orders should have attending name listed, ICD-9 code (348.3 encephalopathy, 742.9 dysgenesis, 345.90 epilepsy), and accurate phone number for family. Email order to neuroscienceauth@cnmc.org



Today's Date:

Department of Diagnostic Imaging and Radiology

Name:

DOB:

MRN:

PATIENT INFORMATION

REQUESTING PHYSICIAN	PATIENT INFORMATION
PRINT NAME:	MOTHER'S NAME:
SIGNATURE:	FATHER'S NAME:
CLINIC: Neurology	ARE THERE PRECAUTIONS?
OFFICE PHONE NUMBER: 202-476-2120	DOES PATIENT HAVE ALLERGY? NKDA
E-MAIL:	IS PATIENT PREGNANT?
TELEPHONE REPORT WANTED?:	
SCHEDULING RADIOLOGIST:	
SPECIAL INSTRUCTIONS: [ ]	

EXAM REQUESTED (no abbreviations)	PRELIMINARY INTERPRETATION
<input type="text"/>	<input type="text"/>
<input type="text"/>	
<input type="text"/>	
DATE & TIME STUDY DESIRED:	RESIDENT/FELLOW:
	REPORTED TO:

CLINICAL HISTORY (INCLUDE DATE OF ONSET OF INJURY)

[ ]

[ ]

INDICATION/SUSPECTED DIAGNOSIS

[ ]:

[ ] Other:

TECHNOLOGIST'S COMMENTS: \_\_\_\_\_

BILLING INFORMATION							
TECHNOLOGIST	INIT.			CODERS	INIT.		RADIOLOGIST INIT: #
Exam s Done	Exam Number	Access. Number	# Films Taken	CPT-4 Code	CFA	ICD-9 Code	Indication/Diagnosis



## 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 CO<sub>2</sub> is  $\geq$  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.charlifoundation.org](http://www.charlifoundation.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, NH<sub>3</sub>, 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 CO<sub>2</sub> is 12-16
  - Assess hydration and hydrate if needed
  - If well-hydrated and symptomatic, give CytraK or decrease ratio
  - Check CO<sub>2</sub> q6 to 8 hrs until stable or symptoms resolve
- If CO<sub>2</sub> 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](http://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, NH<sub>3</sub>, 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 CO<sub>2</sub> >=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

Treatment of deficiency (D3 or cholecalciferol is better-absorbed):

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

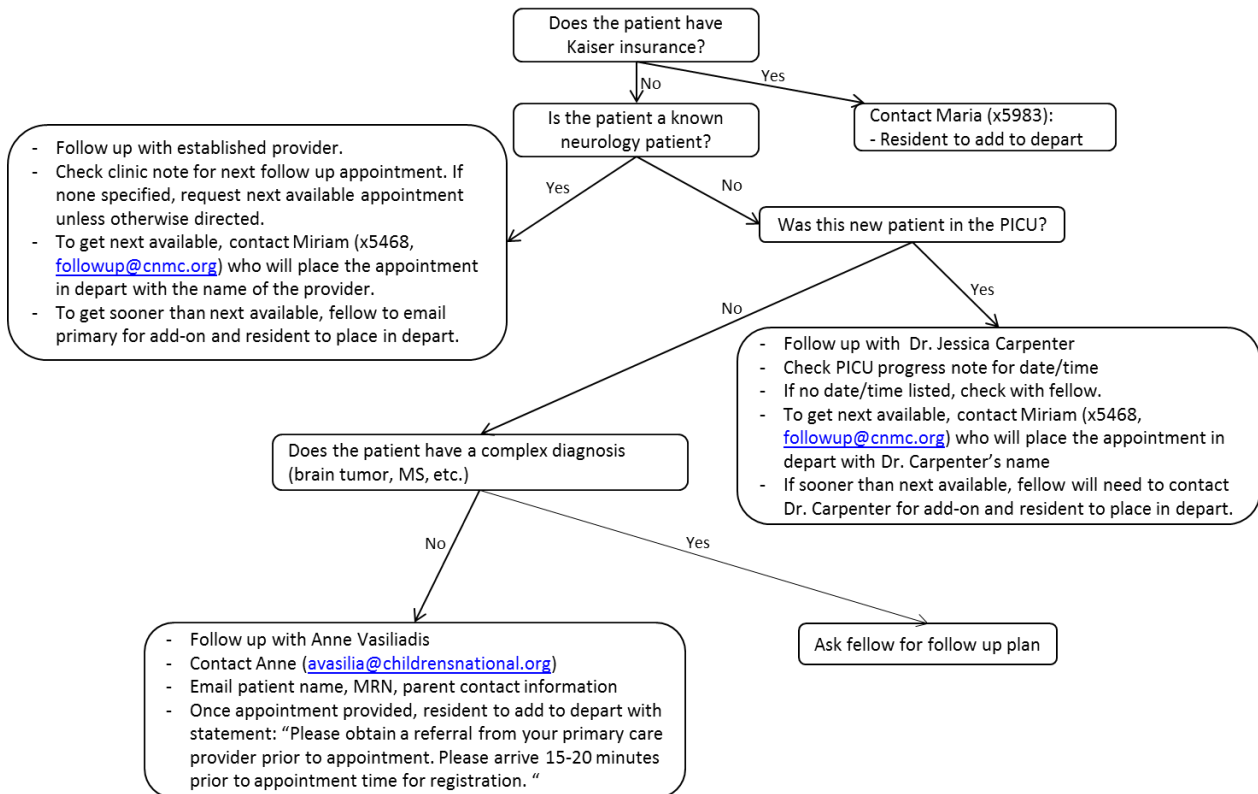
technologists are quite good, and are able to adapt the sites to get data on challenging kids. They do not sedate, but can offer child life and distracters.

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

Congratulations!! Your patient is ready for discharge. Please follow the steps below to schedule a follow up.



- On weekends/holidays/after hours, please still email Miriam or Anne for follow up. Anne will get back to you in real time so that you can include the appointment in the depart and Miriam will schedule the appointment and contact the family during the week.
- If family is not contacted for appointment, instruct them to call the appointment line **202-476-3611** NOT the neurology clinic.