

Comer Children's Hospital

AT THE FOREFRONT OF KIDS MEDICINE"



# Pediatric Neurology Vade Mecum

Peds Neuro Fellow 7678	Comer 5 Fax 2-4361
Attendings:	Comer 5 Charge RN 5-7870
Dr. Tonsgard 3133, (c)312-718-2024	Lab 2-1316
Dr. Henry 6179, (c)407-341-9026	Pharmacy 5-0093
Dr. MacMillan 7141, (c)312-479-3448	Bed Access 4-9130
Dr. Phitsanuwong 7978, (c)312-523-7325	Transfer Center 4-4782
Dr. Nordli 9375, (c)630-659-6976	Neuroradiology Reading Room 5-8044
Dr. Doll 9268, (c)630-544-4295	Peds MRI 2-5185
Dr. David 9281 (c)661-373-1924	PET scan 2-0346
Dr. Hur 2396, (c)312-561-0584	Sedation (MRI scheduling) 4-8585
Fellows:	Nurses:
Kaylene Fiala 4670 (c)608-772-5293	<b>Debbie Ruckman</b> 3977, 4-1927
Shawn Kacker 4674 (c)205-370-7346	Jackie Miller 4721, 4-8965
Pitchamol Vilaisaktipakorn 2925 (c)617-763-5485	Joy Johnican 7562, 4-3456
Rebecca Spasari 2922 (c)224-545-0710	Kymberli Lynom 6969, 2-7529
Carol Park 4296 (c)847-420-4193	<b>EEG Room:</b> 43665 Fax 40830
Jeong-a Kim 4240, (c)586-212-5009	EEG tech on call: pager 7374
Nurse practitioners:	Lead EEG tech phones 68698
Audrey Oetomo 3508, (c)415-318-6074	Inpatient tech phone 68697
Danielle Bender 8329, (c)630-204-8388	Outpt EEG tech phone 68696
Peds Neuro Office (Janice) 4-8966	EMU group tech pager 11500
Richard Cashman 4-2422	<u>Silver team</u> 7587/5-7878
Peds Neuro Fax 2-4786	Maroon team (night team) 7337/5-9619
Comer Clinic 2-6169 fax 2-3267	PICU Fellow 7428/5-7949
Peds Neuro SW Maria Perez 3950	PICU Residents 7430/5-7935
Ketogenic RD Stephanie Schimpf 5622	NICU resident 6361/5-6372
On-call SW 6807	NICU APN 6360/2-6185
Translator 133 ext 2233	LaRabida"ROC" 773-256-2585
Extensions: (773) 70-2, 75-3, 83-4, 79-5, 92-6	Comer ER: 2-6249

## Antiepileptic Medications in Pediatric Neurology

Phenobarbital: 20mg/5mL tabs: 15mg, 30mg, 60mg, 100mg; Loading 20mg/kg (not exceed 300mg/dose);

Maintenance 4-7 mg/kg/day; Level 14-40 (4 hours after load)

Phenytoin/Fosphenytoin (Dilantin): 125mg/5mL, Caps 50mg chewable, 30mg, 100mg, 200mg, 300mg;

Loading 15-20mg/kg singe or divided; Maintenance 5-10mg/kg/day (no exceed 300mg/day;

Level total 10-20, Free 1-2 (use Fospheny IV)

Carbamazepine (Tegretol, Carbatrol): 100mg/5mL, Caps 100mg chewable, 100mg, 200mg, 300mg, 400mg;

Maintenance 10-40mg/kg/day; Level 8-12

Oxcarbazepine (Trileptal): 300mg/5mL, Tabs: 150mg, 300mg, 600mg; Maintenance 20-60mg/kg/day Valproate (Depakote, Depakene, Depakon): 250mg/5mL, sprinkles 125mg, Tabs 125mg, 250mg, 500mg;

Loading 10mg/kg; Maintenance 20-60mg/kg/day; Level 50-200 (VPA Loading: (Goal level-current level) X Wt (kg) X Volume of distribution. Volume of distribution is ~0.2 in adults and ~ 0.4 in children)

Lamotrigine (Lamictal): Tabs 5mg, 25mg chewable, 100mg, 150mg, 200mg; Maintenance 5-15mg/kg/day

(start at 0.3mg/kg/day if not on VPA, 0.15mg/kg/day of on VPA); Level 2.5-15

Levetiracetam (Keppra): 100mg/mL, Tabs 250mg, 500mg, 1000mg; Maintenance 20-60mg/kg/day

Topiramate (Topamax): 6mg/mL, sprinkles 15mg, 25mg, Tabs 25mg, 50mg, 100mg, 200mg

Start 1-2mg/kg/day Maintenance 5-10mg/kg/day Level 2-25

Ethosuximide (Zarontin): 250mg/5mL caps 250mg; Maintenance 15-40mg/kg/day; Level 40-100

Lacosamide (Vimpat): 10mg/mL, Tabs 50mg, 100mg, 150mg, 200mg;

Start 1 mg/kg/day, Max 5-10 mg/kg (max 200mg BID)

Clobazam (Onfi): Tabs 10mg, 2.5mg/mL Start 5mg QHS; Maintenance 1-2mg/kg/day, Max 40mg/day

Clonazepam (Klonapin): Tabs 0.5mg, 1mg, 2mg; Maintenance 0.1-0.2mg/kg/day

Rufinamide (Banzel): 40mg/mL, Tabs 200mg, 400mg; Start 10mg/kg/day, Maintenance 45mg/kg/day Max dose 3,200mg/day

Vigabatrin (Sabril): 50mg/mL, Tabs 500mg, Start 50mg/kg/day, Max dose 150mg/kg/day

# **Serum Drug Levels**

Oxcarbazepine	8-35 mcg/mL
Carbamazepine	4-12 mcg/mL
Ethosuximide	40-100 mcg/mL
Felbamate	40-65 mcg/mL
Lamotrigine	1.5-15 mcg/mL
Phenytoin	10-20 mcg/mL (Free 1-2)
Phenobarbital	15-40 mcg/mL
Topiramate	2-25 mcg/mL
Valproate	50-100 mcg/mL
Zonisamide	10-20 mcg/mL
Rufinamide	3-30 mcg/mL

Adverse Effects of Anti-seizure Medications in Children

Medications	Common side effects	Serious side effects
Carbamazepine (Tegretol)	Dizziness, vertigo, ataxia Diplopia Hyponatremia (tend to be asymptomatic) Worsen myoclonus and absence	Hepatotoxicity Steven-Johnson syndrome (beware HLA- B1502 in Han Chinese population)
Clobazam (Onfi)	Drowsiness, somnolence, short attention span Hypersalivation (increased drooling) Weight gain	CVS and RS suppression (dose dependent)
Ethosuximide (Zorontin)	Nausea, vomiting Abdominal pain (better to give with meal)	Leukopenia Depression, memory problems
Felbamate (Felbatol)	Headache Insomnia Weight loss (from decreased appetite)	Fatal hepatotoxicity Aplastic anemia
Fosphenytoin/ Phenytoin (Dilantin)	Nystagmus Ataxia, incoordination Worsen myoclonus and absence Coarse faces, Hirsutism (long-term use) Gingival hypertrophy (long-term use) Altered vitamin D metabolism	Hepatotoxicity Cardiac arrhythmia (can be life-threatening) Paradoxical seizure (from supratherapeutic level) Cerebellar atrophy (long-term use) Drug-induced SLE Fetal hydantoin syndrome
Lacosamide	Dizziness, headache	Ataxia, impaired balance
(Vimpat)	Diplopia	Cardiac arrhythmia

# Adverse Effects of Anti-seizure Medications in Children

Medications	Common side effects	Serious side effects
Lamotrigine (Lamictal)	Fatigue, confusion May worsen myoclonus (~7%)	Steven-Johnson syndrome, TEN (titration dependent)
Levetiracetam (Keppra)	Aggression, irritability, nervousness	Psychosis
Oxcarbazepine (Trileptal)	Hyponatremia (tend to be asymptomatic) Dizziness, vertigo, ataxia (less frequent than CBZ) Diplopia (less frequent than CBZ) Worsen myoclonus and absence	Hepatotoxicity Steven-Johnson syndrome (beware HLA-B1502 in Han Chinese population)
Parampanel (Fycompa)	Agression	Suicidality Psychosis
Phenobarbital	Drowsiness, sedation Effect on cognition (long-term use) Hyperactivity and irritability (long-term use) Altered vitamin D and K metabolism (causing low vit D and K)	Hepatotoxicity Withdrawal seizure (from abrupt discontinuation) Connective tissue changes (contracture, fibroma) Respiratory suppression (dose dependent)
Rufinamide (Banzel)	Somnolence Nausea, vomiting	QT prolongation

Pyrexia

# Adverse Effects of Anti-seizure Medications in Children

Medications	Common side effects	Serious side effects
Topiramate (Topamax)	Impaired concentration Somnolence Word-finding difficulty Cognitive dysfunction Paresthesia Weight loss (from decreased appetite)	Kidney calculi Increased intra-ocular pressure (beware in glaucoma) Oligohydrosis (lead to hyperthermia)
Valproate (Depakote)	Weight gain (from increased appetite) Nausea, vomiting, abdominal pain Tremor Hair loss (temporary and may be prevented with zinc supplement) Thrombocytopenia (dose depenent)	Acute encephalopathy Hyperammonemia Hepatotoxicity (can be fatal in POLG1 mutation) Pancreatitis Mitochondrial dysfunction Secondary carnitine deficiency Fetal major congenital malformation
Vigabatrin (Sabril)	Drowsiness, somnolence Change of muscle tone Non-specific white matter signal change	Irreversible visual field restriction (particularly in the nasal sides)
Zonisamide (Zonogran)	Fatigue Altered thinking	Oligohydrosis (leads to hyperthermia) Kidney calculi

<u>Seizure</u>: A transient occurrence of signs and/or symptoms due to Abnormal excessive or synchronous neuronal activity in the brain.

# ILAE 2017 Classification of Seizure Types Expanded Version <sup>1</sup>

## **Focal Onset**

Aware Impaired Awareness

### **Motor Onset**

automatisms atonic <sup>2</sup> clonic epileptic spasms <sup>2</sup> hyperkinetic myoclonic tonic

### **Nonmotor Onset**

autonomic behavior arrest cognitive emotional sensory

## **Generalized Onset**

### Motor tonic-clonic

clonic

tonic myoclonic myoclonic-tonic-clonic myoclonic-atonic atonic

# Nonmotor (absence)

typical atypical myoclonic eyelid myoclonia

epileptic spasms

## **Unknown Onset**

Motor tonic-clonic epileptic spasms Nonmotor behavior arrest

Unclassified <sup>3</sup>

focal to bilateral tonic-clonic

# DIASTAT (Diazepam rectal gel) Dosing recommendations by age and weight

<b>2 - 5 Y</b> ears 0.5 mg/kg	
Weight (kg)	Dose (mg)
6 to10	5
11 to 15	7.5
16 to 20	10
21 to 25	12.5
26 to 30	15
31 to 35	17.5
36 to 44	20

<b>6 - 11 Years</b> 0.3 mg/kg		
Weight (kg)	Dose (mg)	
10 to 16	5	
17 to 25	7.5	
26 to 33	10	
34 to 41	12.5	
42 to 50	15	
51 to 58	17.5	
59 to 74	20	

12+ Years			
0.2 ו	0.2 mg/kg		
Weight	Dose		
(kg)	(mg)		
14 to 25	5		
26 to 37	7.5		
38 to 50	10		
51 to 62	12.5		
63 to 75	15		
76 to 87	17.5		
88 to 111	20		

# **Antiepileptic Medications of Choice**

Depends on seizure type, syndrome, medication side effect profile, administration, dosing, and parental choice.

- 1. Neonatal seizure: Phenobarbital, Phenytoin, Levetiracetam
- Genetic Generalized Epilepsies: Valproate, Topiramate, Lamotrigine, Zonisamide, Levetiracetam, Clobazam
- Narrow spectrum: Carbamazepine, Oxcarbazepine, Lacosamide, Phenytoin, Phenobarbital, Vigabatrin
- 4. Status epilepticus: Benzodiazepine, Phenytoin, Phenobarbital, Valproate, Levetiracetam
- 5. Absence: Ethosuximide, Valproate, Lamotrigine (avoid Sodium channel blockers)
- **6. Juvenile myoclonic epilepsy:** Valproate, Lamotrigine, Levetiracetam (avoid Sodium channel blockers)
- 7. Rolandic epilepsy (if treating): Carbamazepine, Oxcarbazepine, Levetiracetam
- 8. ESES/CSWS: Diazepam, corticosteroids, Valproate, Clobazam, Topiramate, Ethosuximide
- 9. Lennox- Gastaut syndrome: Valproate, Felbamate, Topiramate, Lamotrigine, Clobazam, Rufinamide
- 10. Dravet Syndrome: Valproate, Clobazam, Stiripentol (avoid Sodium channel blockers)
- 11. Infantile Spams: ACTH, Vigabatrin, Corticosteroids

### **Drug Facts:**

Enzyme inducers: PHT, PB, CBZ, OXC

Enzyme inhibitor: VPA

HLA-B1502 increased risk of SJS/TEN in Asian population with CBZ,OXC,PHT

Status Epilepticus = seizure > 5 min or frequent seizures without return to baseline in between **Supportive Care Antiseizure Meds (ASM) Evaluation** Time Confirm any meds give in the last Hx: fever, trauma, ingestion, prior hx Airway, breathing, circulation 12 hours! of seizure/epilepsy Cardiac Monitor If hypoglycemic give dextrose 10% Exam: temperature, blood pressure, Pulse Ox 2.5 mL/kg (unless pt on ketogenic diet. signs of trauma, mental status, ask 0 - 5 min see keto protocol) patient questions and give Start timing the seizure commands Page 1st contact Labs: POC alucose, electrolytes, Ca. Mg, CBC, tox screen, ASM levels2 provider Page Ped Neuro 7678 1st Line Assess mental status 1 If patient has received a rescue dose of No IV Access: IntraNasal Midazolam Screen for seizure risk factors4 benzodiazepine within the past 2 hours, then only give If cardiorespiratory 0.2 ma/ka (max 10ma), give 1/2 dose up 5 - 10 min compromise, call PET3 each nostril one dose of 1 dose benzodiazepine before moving on to 2<sup>nd</sup> line ASM IV Access; Lorazepam, 0.1 mg/kg, (max 4ma/dose), IV push 2 Labs: It is not necessary to repeat CBC, tox screen

10 - 15 min IN Midazolam or IV Lorazepam Monitor closely for 2nd Line - Levetiracetam 60 mg/kg IV respiratory failure and hypotension2 15 - 25 min Call PET and prepare for transfer to PICU

Place IV

Alternate - Fosphenytoin 20 PE/kg IV 3rd Line -Fosphenytoin 20 PE/kg IV Alternate -Valproic acid 30 mg/kg IV (max 3g)

Give 2nd dose of 1st Line1

Convulsive Status Epilepticus Protocol for Pediatric Inpatients

Assess mental status If cause of seizure remains unknown: order CT head, and consider lumbar puncture Place 24hr video EEG order, and contact EMU 43665

Assess mental status

**EEG** monitoring 4 Seizure risk factors: prematurity, developmental delay/regression, family history, hx of meningitis, encephalitis, head trauma or sepsis Assess mental status

and ASM levels if obtained within last 24h

3 If patient has respiratory failure, activate PET,

intubate and skip ahead to Continuous IV Infusion with

25 - 35 min 35+ min

PICU Transfer patient to PICU Continuous cardiopulmonary monitoring

Prepare for transfer to

 Lacosamide 8 ma/ka IV PHenobarbital 20 mg/kg IV (max 300ma) Continuous IV Infusion - titrate to seizure cessation Midazolam - bolus 0.2 mg/kg (max 10ma), infusion 0.1 to 2 ma/kg/hr

OR

minutes, re-bolus with each increase)

over 1-2 hrs, infusion 0.5 to 5 ma/ka/hr

PENTobarbital - bolus 5 to 10 mg/kg

(increase by 0.5-3 mg/kg/hr)

EEG monitoring - depth of EEG coma should be determined on a case-by-case basis (increase by 0.1 mg/kg/hr every 10-15 Blood gasses and metabolic testing as appropriate for monitoring for

complications of prolonged induced

coma. PenTobarbital suppresses

fever and causes ileus.

Please refer to the Pediatric Status Epilepticus Protocol online for further details

# Febrile Seizures (3mo to 5yrs)

Incidence 2-5%, 25-40% have family history: genetic epilepsy with febrile seizures plus (GEFS+) involving SCN1A, 2A, 1B and GABRG2 mutations

Spectrum: febrile seizures → GEFS+ → Dravet Syndrome

Simple	Complex (20-30%)	Febrile status
< 15 min	> 15 min	>30 min
Non-focal	Focal	
1 episode during illness	> 1 event during 24hr/illness	
No Todd's paralysis	Todd's paralysis	

### Management:

- Treat infection and fever
- LP guidelines per AAP 2012:
- > Should be performed in all children with signs, symptoms, or history concerning for CNS infection
- > Optional in 6-12mo olds with unknown vaccination history or deficient in Hib/strep pneumo
- > Optional in children who have been pre-treated with antibiotics as this may mask meningeal signs
- Imaging not warranted unless clinical suspicion for an acute neurological condition or focal hemi-convulsions suggesting possible structural abnormality
- EEG: up to 1/3 will show transient EEG abnormalities in complex febrile seizures and febrile status epilepticus
- Treat febrile status epilepticus like non-febrile status; be cautious in children with sodium channelopathies (such as Dravet) where sodium channel blockers such as fosphenytoin are contraindicated

# **Neurologic Emergencies**

## **Refractory Status Epilepticus**

(Titrate to cessation of electrographic seizures or burst suppression – depth of EEG coma should be determined on a case-by-case basis)

**Midazolam**: Loading dose 0.2 mg/kg (max 10 mg), Continuous infusion 0.1 mg/kg/hr, increase by 0.1 mg/kg/hr every 10-15 minutes (re-bolus with each rate increase), max rate of 2 mg/kg/hr as hemodynamic stability permits

**Pentobarbital**: Loading dose 5-10 mg/kg over 1-2 hours, Continuous infusion 1 mg/kg/hr, increase by 0.5-3 mg/kg/hr, max rate of 5 mg/kg/hr (when tapering, decrease by 0.5 mg/kg every 12 hours) **Ketamine**: Loading dose 0.5-2 mg/kg, Continuous infusion 0.3-1.2 mg/kg/hr (5-20 mcg/kg/min)

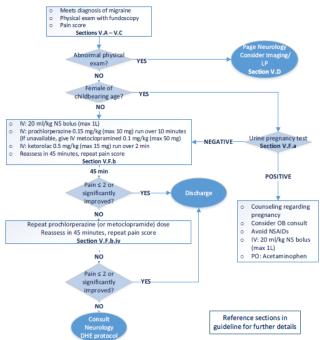
### Increased ICP

Elevate HOB 30 degrees
Mannitol 0.25-1 g/kg/dose IV
3% NaCl 6 cc/kg run at 1 cc/min
Sedation and analgesia

### **Cerebral Edema**

**Dexamethasone:** IV/PO Loading dose 1-2 mg/kg/dose, Maintenance dose 1-1.5 mg/kg/day divided every 4-6 hours (max 16 mg/day)

# **Migraine Management**



Patients who present within 4.5 hrs → activate Stroke Code (Dial 144) If patient has sickle cell disease, page Hematology fellow on call **Initial Management:** 

Pediatric Acute Stroke Guideline

- ABCs, place on cardiac monitor
- Complete Peds NIHSS, full neuro exam
- Staff patient with Dr. Henry David (661-373-1924)
- Place 2 PIVs 0.9NS bolus 10 ml/kg (max 1000 ml)
- STAT labs: CBC, CMP, coags, T/S (+/- Hgb electrophoresis, retic)
- Stat head CT wo\* → consider tPA (assess inclusion/exclusion criteria)
- EKG, CXR, UA (+/- Utox, ETOH, AED levels)
- **Additional Imaging:**

- Head of bed flat unless hemorrhagic stroke, NPO, vital signs and neurocheck q2h for at least 24hrs then q4h

- \*Consider CTA head/neck after CTH obtained based on degree of suspicion for embolic stroke/clot - MRI brain wo and MRA brain/neck wwo if CTA not obtained

## **BP Management:**

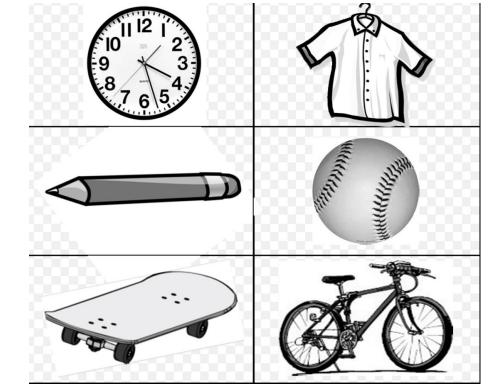
Notify and treat if systolic or diastolic BP >95%ile on >3 repeated measurements

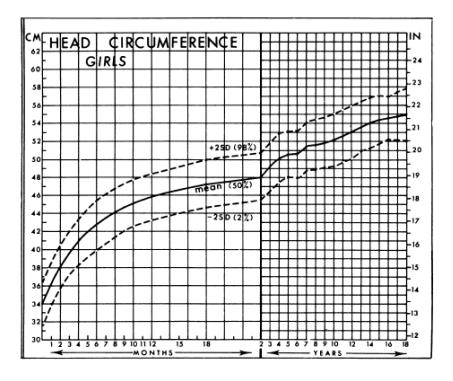
In setting of hemorrhage, maintain BP >50%ile and <95%ile

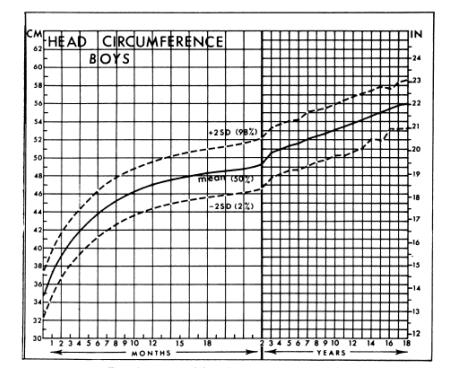
- Labetalol: 0.2-1 mg/kg/dose q4-6 hrs (max 40 mg/dose)
- Hydralazine: 0.1-0.2 mg/kg/dose q4-6hrs (max 20 mg/dose)
- Nicardipine infusion if requiring multiple PRNs: 0.5-1 mcg/kg/min, nurse to titrate to goal BP

# **Further management:**

- If new onset ischemic stroke → LDL, HbA1C, TSH, ESR/CRP, TTE, start ASA 3-5 mg/kg/day, DVT prophylaxis
- SCD and/or Hep 5000 units SQ BID, consult PT/OT/ST - Consider hypercoagulable workup: Antithrombin III, Protein C/S, Factor V Leiden, Prothrombin Mutation,
- Homocysteine level, Anticardiolipin IgM/IgG, Thrombin Time - Consider hematologic conditions: PT20210 and MTHFR gene mutation, Lipoprotein A, Factors II, VIII, XI level
- Consider metabolic syndrome: lactate, pyruvate, ammonia, CK, plasma amino acid, urine organic acid







Age	Gross Motor	Fine Motor	Language and Cognition
1 mo	Head up while prone	Hands fisted	Fixed and follow
2 mo	Chest up in prone	Hands unfisted 50%	Social smile, regards speaker
4 mo	Roll front to back, no head lag	Hands to midline, reach for objects	Social laugh
6 mo	Roll back to front, sit w/o support	Transfer objects, hold bottle	Babbling
9 mo	Crawling, pull to stand	Bangs 2 toys, starts pincer grasp	Orient to name, "peek-a-boo", understands "NO", finds hidden object
12-15 mo	Walking +/- 3mo	Imitate scribble, tower of 2 blocks	Knows 1-2 body parts, says "NO", follows simple commands, few words
18 mo	Walk up stairs, throw ball, jump	3 cube tower	20 words, 3 body parts
24 mo	Run, jump, kick ball, walk up and down stairs	Draw vertical line	Follow 2-step command, 2 word sentences, refers to self by name
3 yr	Rides a tricycle	Turns door know, copy a circle, independent eating	Gives full name, knows age and gender, toilet training
4 yr	Hop on 1 foot, alternate feet descending stairs	Copy a cross, dress, buttons	Colors, counting, fantasy play, tells stories
5 -6 yr	Walk on tiptoes, skips, catches ball	Copy a triangle	Knows L and R, age and birthday, days of week, Bday, reading
7-8 yr	Tandem walk	Ties shoes, combs hair, print name	Days of the week, add/subtract, tells time