

Ketogenic and Modified Ketogenic Diets

Jennifer Madan-Cohen, MD

Beth Chatfield, RDN

Jamie Cubanski, RN



What is a Clinical Pathway?



An evidence-based guideline that decreases unnecessary variation and helps promote safe, effective, and consistent patient care.

Objectives of Pathway



- Establish medical and safety standards of care for the initiation of the ketogenic and modified ketogenic diet
 - Standardize the diet education for the caregivers
 - Promote improved tolerance to the ketogenic diet, with fewer side effects
 - Educate medical staff on ketogenic diets as treatment for seizures
-

Why is this pathway necessary?



- Many patients at CCMC require initiation of ketogenic diets for seizure control.
 - This pathway will help standardize care of patients who are starting, or maintaining a ketogenic diet.
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Ketogenic and Modified Ketogenic Diets



- The Ketogenic and Modified Ketogenic diets are high-fat, low-carbohydrate, evidence-based diets, used to help control seizures.
 - At Connecticut Children's, both of these are medically supervised treatments for seizures, **not** "diets" for weight loss.
 - They are Medical Nutrition Treatment options for epilepsy
-

Ketogenic and Modified Ketogenic Diets



- The goal: alter the body's fuel source from glucose to fat and mimic the effects of fasting.
- Glucose → Fat (as the primary fuel source)
- Ketone bodies : the acidic products formed from excessive breakdown of fat. The brain uses the ketones as fuel.
- Underlying mechanisms of the benefits to seizure disorder remain unclear and are most likely parallel and potentially synergistic, including: changes in neurotransmitter systems, metabolic coupling, glycolytic restriction, enhancement of TCA cycle, inhibitory effects of fatty acids, improved cellular bioenergetics and mitochondrial function along with recent research on inflammatory agents.

What makes Modified Keto Diet Different?



- Carbs counted by family
 - Added fat per meal usually 1-3 Tablespoons
 - Can have more protein than recommended daily intake as long as extra fat is used
 - Calories are not restricted
 - First follow up blood work done at 1 month instead of 2 weeks (takes longer to get into ketosis for some)
-

Why Choose Modified?



- More flexibility
 - Efficacy for seizure control is the same as the KETO Diet at the 1 month mark for ages 2 and older
 - No need for a scale
 - Can eat more protein
 - More commercially available products that can be used due to allotted carb amount higher
-

Ketogenic and Modified Ketogenic Diets



Every child that is actively on the Ketogenic or Modified Ketogenic diet will have a “keto alert” listed in EPIC.

The screenshot displays the EPIC patient summary interface. At the top, a patient information bar includes fields for MRN, CSN, Weight (11.8 kg), Allergies (Dilantin [Phe...]), Code (Full), Isolation (Droplet), and other clinical data. A blue arrow points to a 'Keto alert' in the top bar, which is circled in pink. Below the top bar, the 'Summary' section is visible, featuring a 'Handoff' section with an 'Edit Handoff' link. A yellow bar highlights the 'Patient on Ketogenic Diet' alert, which is also circled in pink. Below this, a 'BestPractice Advisories' section is shown, with a link to view active advisories. A large blue text box is overlaid on the summary section, stating: 'Patients will have alerts in EPIC in both the top bar as well as with the BestPractice Advisories'.

2018 Updates in Pathway



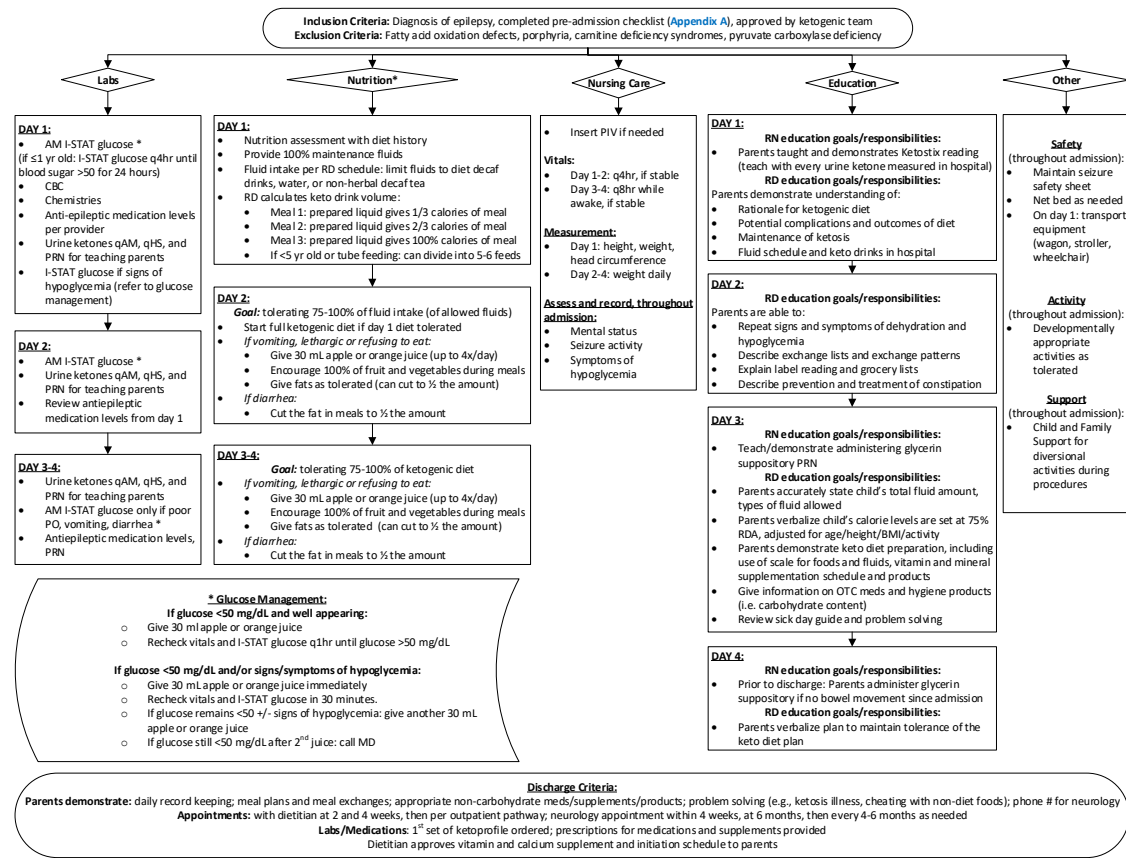
- New guidelines for blood glucose monitoring – will discuss in following slides.

Ketogenic Diet Pathway



This is the Ketogenic Diet Clinical Pathway.

We will be reviewing each component in the following slides.



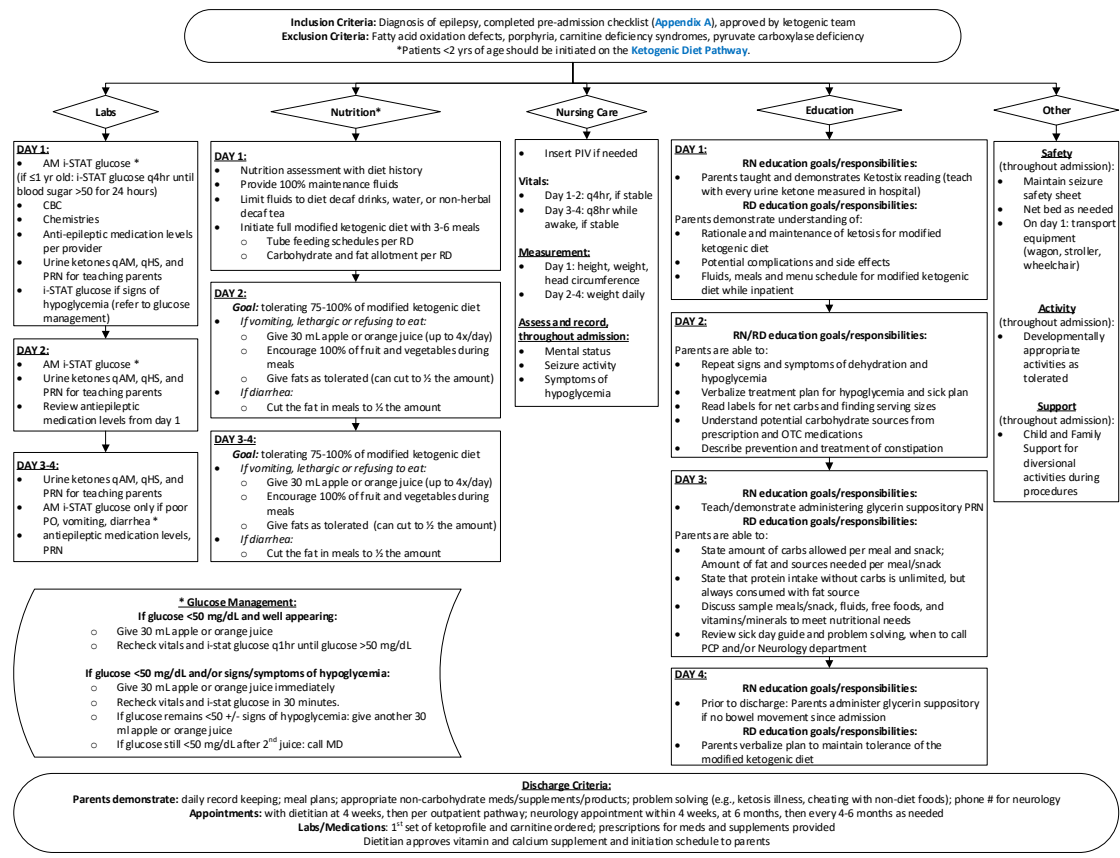
Modified Ketogenic Diet Pathway



This is the Modified Ketogenic Diet Clinical Pathway

What are the differences between the two pathways?

Many things are exactly the same on both pathways. We will review differences on the Modified Ketogenic Diet Pathway as we go along.



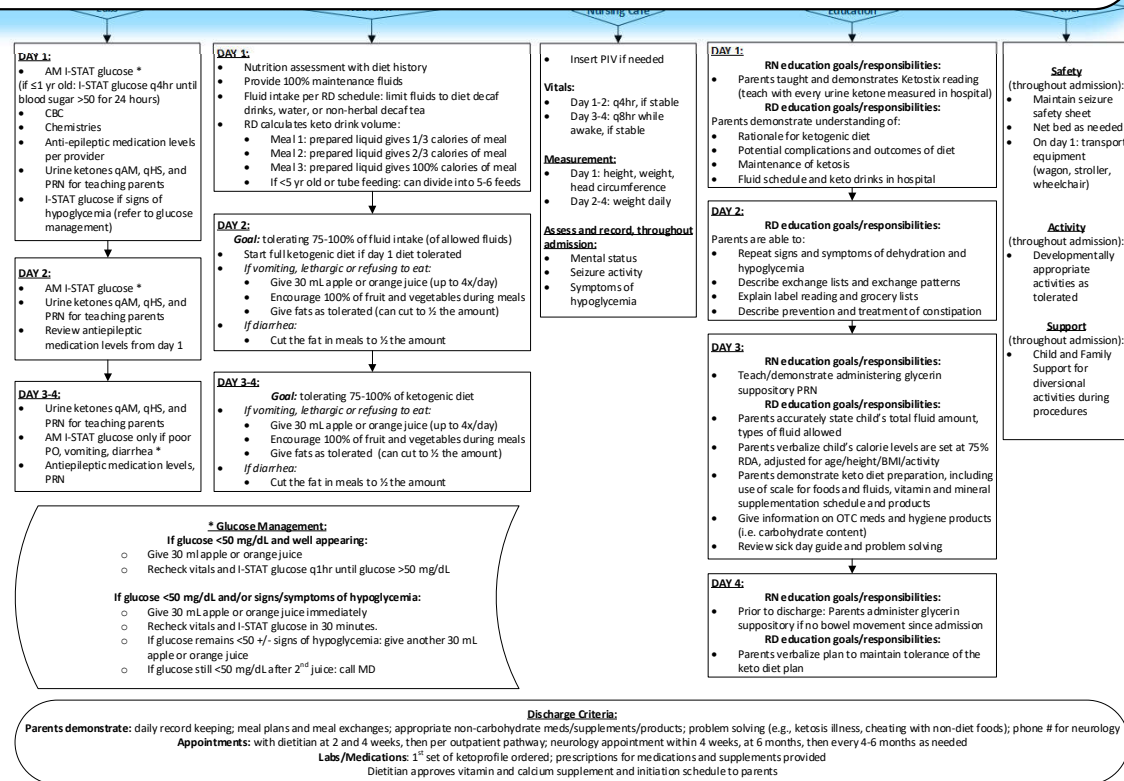
Keto



Inclusion Criteria: Diagnosis of epilepsy, completed pre-admission checklist (**Appendix A**), approved by ketogenic team
Exclusion Criteria: Fatty acid oxidation defects, porphyria, carnitine deficiency syndromes, pyruvate carboxylase deficiency

Both pathways are for patients with the diagnosis of epilepsy that have already completed the pre-admission check list and are approved by ketogenic team.

Appendix A is the pre-admission checklist. See next slides.

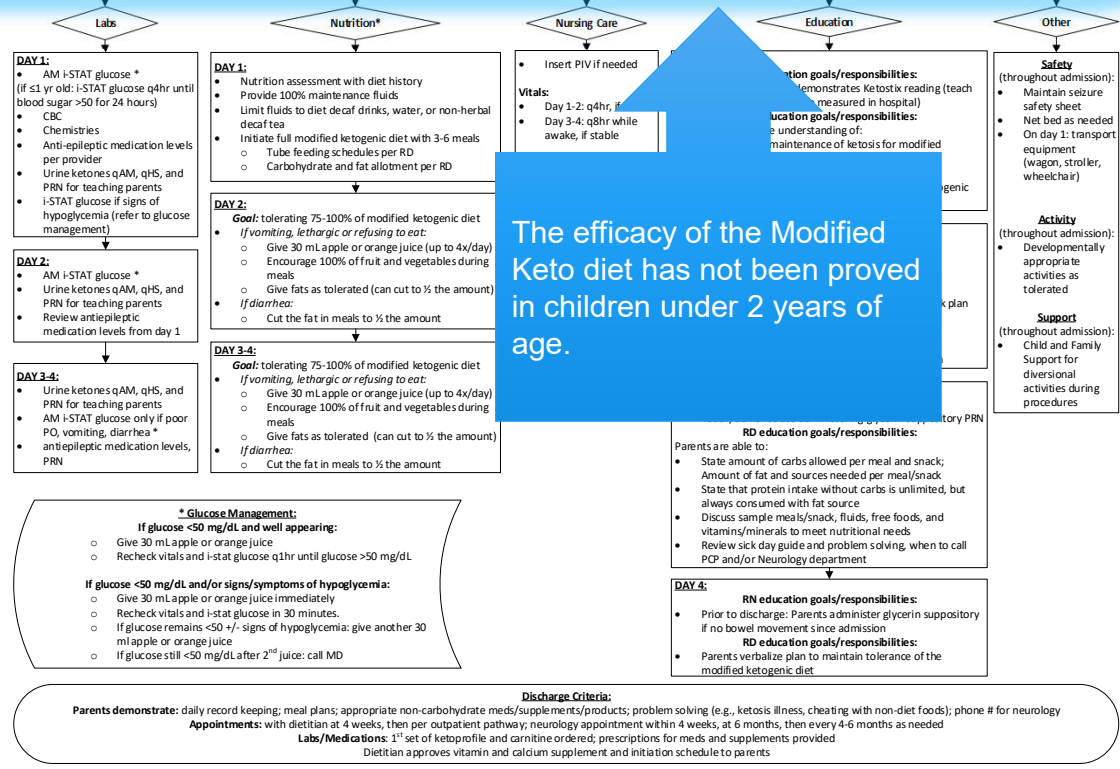


Modified Keto



Inclusion Criteria: Diagnosis of epilepsy, completed pre-admission checklist ([Appendix A](#)), approved by ketogenic team
Exclusion Criteria: Fatty acid oxidation defects, porphyria, carnitine deficiency syndromes, pyruvate carboxylase deficiency
 *Patients <2 yrs of age should be initiated on the [Ketogenic Diet Pathway](#).

One major thing to note is that patients younger than 2 years of age will only ever be initiated on the Ketogenic Pathway, NOT the Modified Ketogenic Pathway



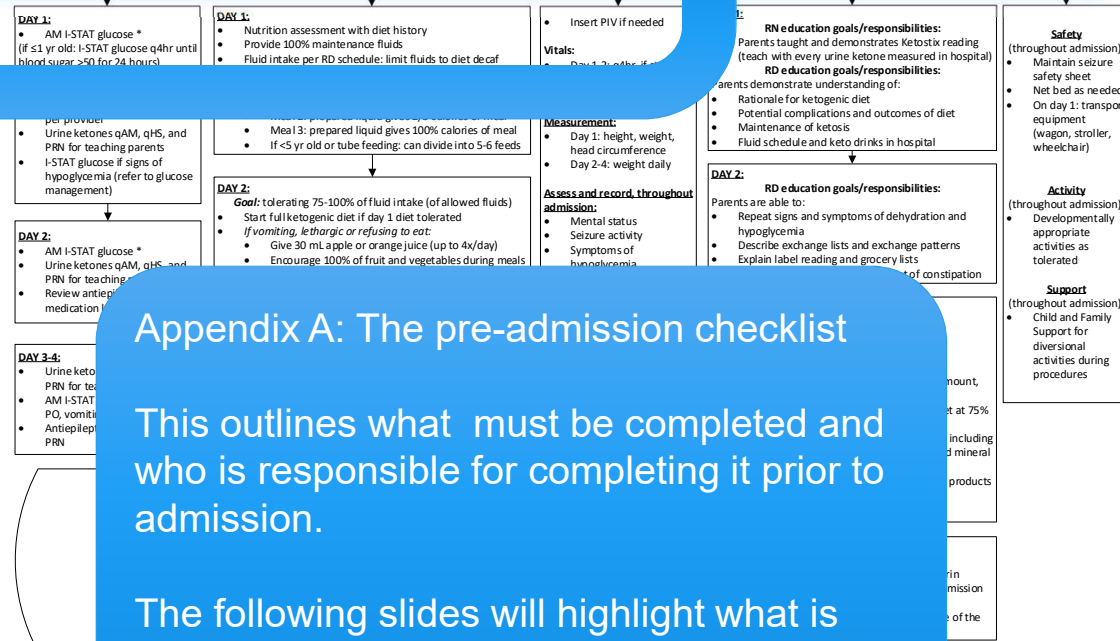
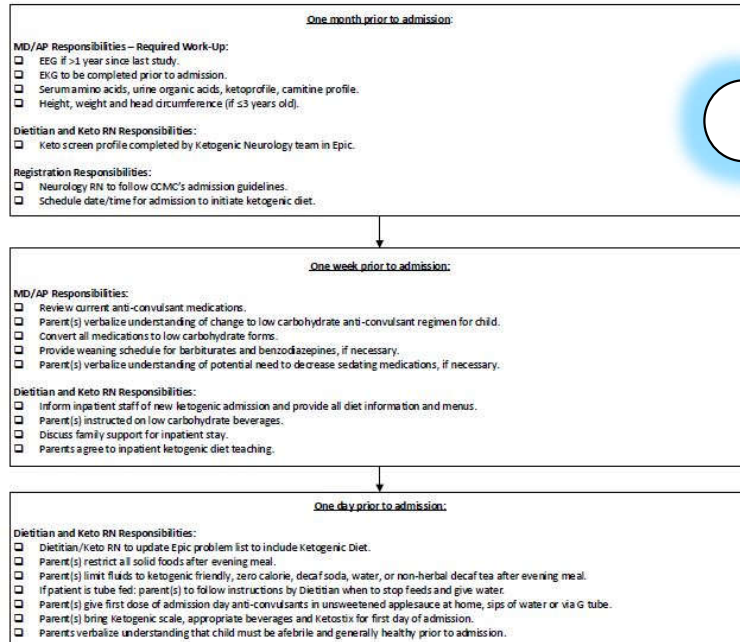
The efficacy of the Modified Keto diet has not been proved in children under 2 years of age.

CLINICAL PATHWAY:
Ketogenic Diet
Appendix A: Pre-admission Checklist

THIS PATHWAY
SERVES AS A GUIDE
AND DOES NOT
REPLACE CLINICAL
JUDGMENT.



Inclusion Criteria: Diagnosis of epilepsy, completed pre-admission checklist (Appendix A), approved by ketogenic team
Exclusion Criteria: Fatty acid oxidation defects, porphyria, carnitine deficiency syndromes, pyruvate carboxylase deficiency



Appendix A: The pre-admission checklist

This outlines what must be completed and who is responsible for completing it prior to admission.

The following slides will highlight what is required

CONTACTS: JENNIFER MADAN COHEN, MD | BETH CHATFIELD, RDN | JAMIE CUBANSKI, RN



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CLINICAL PATHWAY:
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One month prior to admission:

MD/AP Responsibilities – Required Work-Up:

- EEG if >1 year since last study.
- EKG to be completed prior to admission.
- Serum amino acids, urine organic acids, ketoprofile, carnitine profile.
- Height, weight and head circumference (if ≤3 years old).

Dietitian and Keto RN Responsibilities:

- Keto screen profile completed by Ketogenic Neurology team in Epic.

Registration Responsibilities:

- Neurology RN to follow CCMC's admission guidelines.
- Schedule date/time for admission to initiate ketogenic diet.

- Dietitian and Keto RN Responsibilities:**
- Inform inpatient staff of new ketogenic admission and provide all diet information and menus.
 - Parent(s) instructed on low carbohydrate beverages.
 - Discuss family support for inpatient stay.
 - Parents agree to inpatient ketogenic diet teaching.

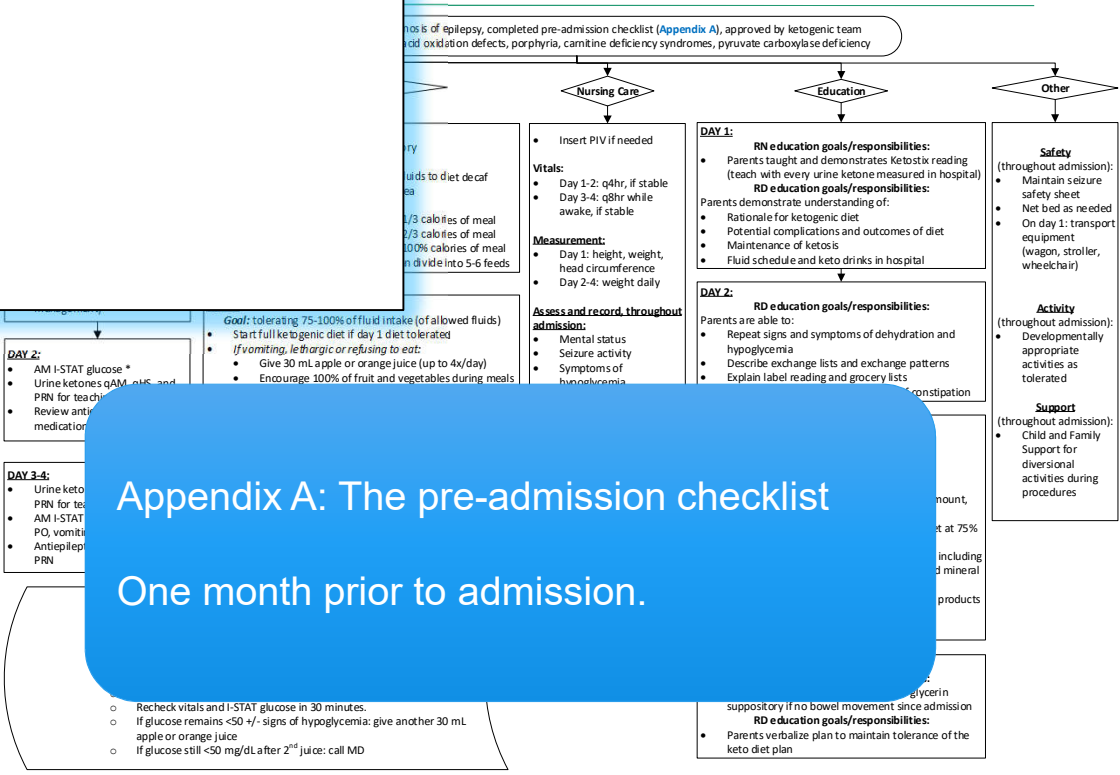
One day prior to admission:

- Dietitian and Keto RN Responsibilities:**
- Dietitian/Keto RN to update Epic problem list to include Ketogenic Diet.
 - Parent(s) restrict all solid foods after evening meal.
 - Parent(s) limit fluids to ketogenic friendly, zero calorie, decaf soda, water, or non-herbal decaf tea after evening meal.
 - If patient is tube fed: parent(s) to follow instructions by Dietitian when to stop feeds and give water.
 - Parent(s) give first dose of admission day anti-convulsants in unsweetened applesauce at home, sips of water or via G tube.
 - Parent(s) bring ketogenic scale, appropriate beverages and ketotrix for first day of admission.
 - Parents verbalize understanding that child must be afebrile and generally healthy prior to admission.

- DAY 2:**
- AM I-STAT glucose *
 - Urine ketones qAM, pH, and PRN for teaching
 - Review anti-convulsant medications
- DAY 3-4:**
- Urine keto
 - PRN for teaching
 - AM I-STAT
 - PO, vomiting
 - Antiepileptic PRN

- Goal: tolerating 75-100% of fluid intake (of allowed fluids)**
- Start full ketogenic diet if day 1 diet tolerated
 - *If vomiting, lethargic or refusing to eat:*
 - Give 30 mL apple or orange juice (up to 4x/day)
 - Encourage 100% of fruit and vegetables during meals

Appendix A: The pre-admission checklist
One month prior to admission.



Discharge Criteria:
Parents demonstrate: daily record keeping; meal plans and meal exchanges; appropriate non-carbohydrate meds/supplements/products; problem solving (e.g., ketosis illness, cheating with non-diet foods); phone # for neurology

Appointments: with dietitian at 2 and 4 weeks, then per outpatient pathway; neurology appointment within 4 weeks, at 6 months, then every 4-6 months as needed

Labs/Medications: 1st set of ketoprofile ordered; prescriptions for medications and supplements provided
Dietitian approves vitamin and calcium supplement and initiation schedule to parents

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- Height, weight and head circumference (if <3 years old).

Dietitian and Keto RN Responsibilities:

- Keto screen profile completed by Ketogenic Neurology team in Epic.

Registration Responsibilities:

- Neurology RN to follow CCMC's admission guidelines.
- Schedule date/time for admission to initiate ketogenic diet.

One week prior to admission:

One week prior to admission:

MD/AP Responsibilities:

- Review current anti-convulsant medications.
- Parent(s) verbalize understanding of change to low carbohydrate anti-convulsant regimen for child.
- Convert all medications to low carbohydrate forms.
- Provide weaning schedule for barbiturates and benzodiazepines, if necessary.
- Parent(s) verbalize understanding of potential need to decrease sedating medications, if necessary.

Dietitian and Keto RN Responsibilities:

- Inform inpatient staff of new ketogenic admission and provide all diet information and menus.
- Parent(s) instructed on low carbohydrate beverages.
- Discuss family support for inpatient stay.
- Parents agree to inpatient ketogenic diet teaching.

DAY 1:

- AM I-STAT glucose (if <1 yr old: I-STAT glucose)
- CBC
- Chemistries

- Fluid intake per RD schedule: limit fluids to diet decaf drinks, water, or non-herbal decaf tea
- RD calculates keto drink volume:

- 2/3 calories of meal
- 100% calories of meal
- divide into 5-6 feeds

- (of allowed fluids) tolerated
- ice (up to 4x/day)
- vegetables during meals to 1/2 the amount)

- ketogenic diet
- ice (up to 4x/day)
- vegetables during meals to 1/2 the amount)

- If glucose <50 mg/dL and well appearing:
 - Give 30 mL apple or orange juice
 - Recheck vitals and I-STAT glucose q1hr until glucose >50 mg/dL

- If glucose <50 mg/dL and/or signs/symptoms of hypoglycemia:
 - Give 30 mL apple or orange juice immediately
 - Recheck vitals and I-STAT glucose in 30 minutes.
 - If glucose remains <50 +/- signs of hypoglycemia: give another 30 mL apple or orange juice
 - If glucose still <50 mg/dL after 2nd juice: call MD

- Day 1-2: q4hr, if stable
 - Day 3-4: q8hr while awake, if stable
- Measurement:**
- Day 1: height, weight, head circumference
 - Day 2-4: weight daily

- Assess and record throughout admission:**
- Mental status
 - Seizure activity
 - Symptoms of hypoglycemia

- (teach with every urine ketone measured in hospital)
- RD education goals/responsibilities:**
- Parents demonstrate understanding of:
 - Rationale for ketogenic diet
 - Potential complications and outcomes of diet
 - Maintenance of ketosis
 - Fluid schedule and keto drinks in hospital

- DAY 2:**
- RD education goals/responsibilities:**
- Parents are able to:
 - Repeat signs and symptoms of dehydration and hypoglycemia
 - Describe exchange lists and exchange patterns
 - Explain label reading and grocery lists
 - Describe prevention and treatment of constipation

- DAY 3:**
- RN education goals/responsibilities:**
- Teach/demonstrate administering glycerin suppository PRN
- RD education goals/responsibilities:**
- Parents accurately state child's total fluid amount, types of fluid allowed
 - Parents verbalize child's calorie levels are set at 75% RDA, adjusted for age/height/BMI/activity
 - Parents demonstrate keto diet preparation, including use of scale for foods and fluids, vitamin and mineral supplementation schedule and products
 - Give information on OTC meds and hygiene products (i.e. carbohydrate content)
 - Review sick day guide and problem solving

- DAY 4:**
- RN education goals/responsibilities:**
- Prior to discharge: Parents administer glycerin suppository if no bowel movement since admission
- RD education goals/responsibilities:**
- Parents verbalize plan to maintain tolerance of the keto diet plan

Other

- Safety** (throughout admission):
- Maintain seizure safety sheet
 - Net bed as needed
 - On day 1: transport equipment (wagon, stroller, wheelchair)

- Activity** (throughout admission):
- Developmentally appropriate activities as tolerated

- Support** (throughout admission):
- Child and Family Support for diversional activities during procedures

Discharge Criteria:
Parents demonstrate: daily record keeping; meal plans and meal exchanges; appropriate non-carbohydrate meds/supplements/products; problem solving (e.g., ketosis illness, cheating with non-diet foods); phone # for neurology

Appointments: with dietitian at 2 and 4 weeks, then per outpatient pathway; neurology appointment within 4 weeks, at 6 months, then every 4-6 months as needed

Labs/Medications: 1st set of ketoprofile ordered; prescriptions for medications and supplements provided

Dietitian approves vitamin and calcium supplement and initiation schedule to parents

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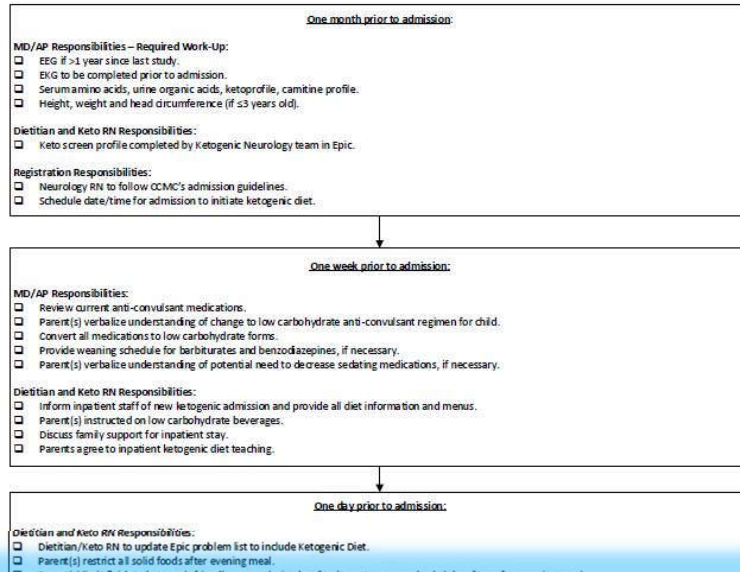
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Appendix A: The pre-admission checklist
One week prior to admission.

CLINICAL PATHWAY:
Ketogenic Diet
Appendix A: Pre-admission Checklist

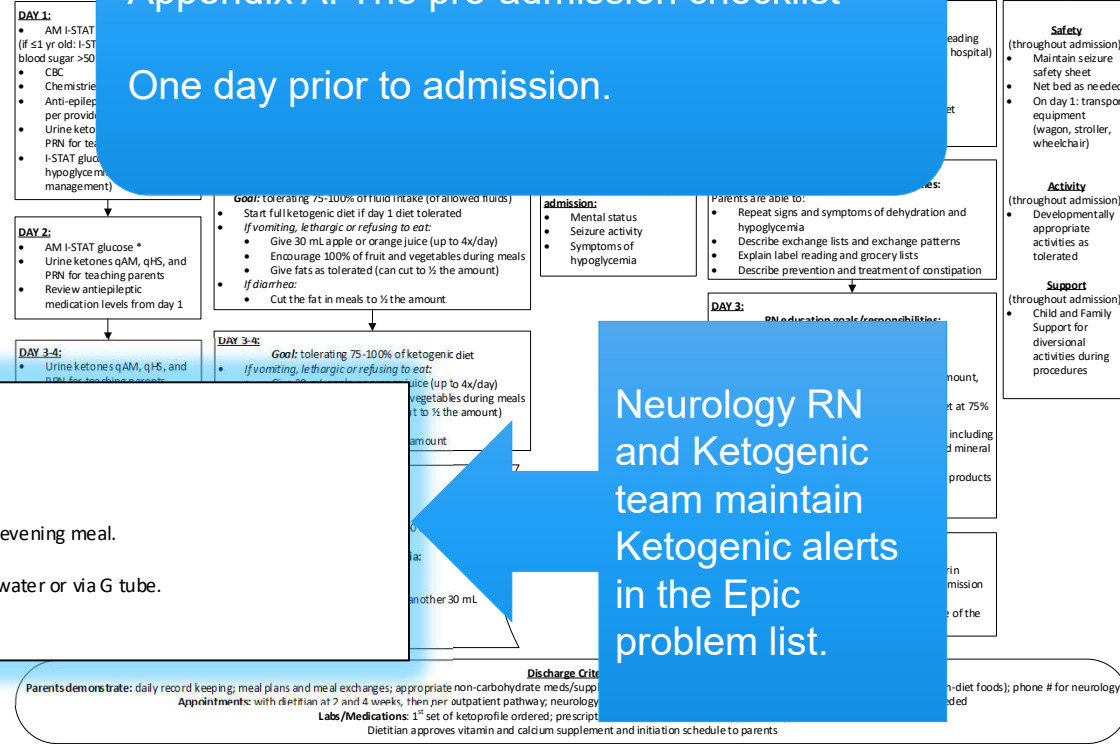
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 - Parent(s) restrict all solid foods after evening meal.
 - Parent(s) limit fluids to ketogenic friendly, zero calorie, decaf soda, water, or non-herbal decaf tea after evening meal.
 - If patient is tube fed: parent(s) to follow instructions by Dietitian when to stop feeds and give water.
 - Parent(s) give first dose of admission day anti-convulsants in unsweetened applesauce at home, sips of water or via G tube.
 - Parent(s) bring Ketogenic scale, appropriate beverages and Ketostix for first day of admission.
 - Parents verbalize understanding that child must be afebrile and generally healthy prior to admission.

Neurology RN and Ketogenic team maintain Ketogenic alerts in the Epic problem list.



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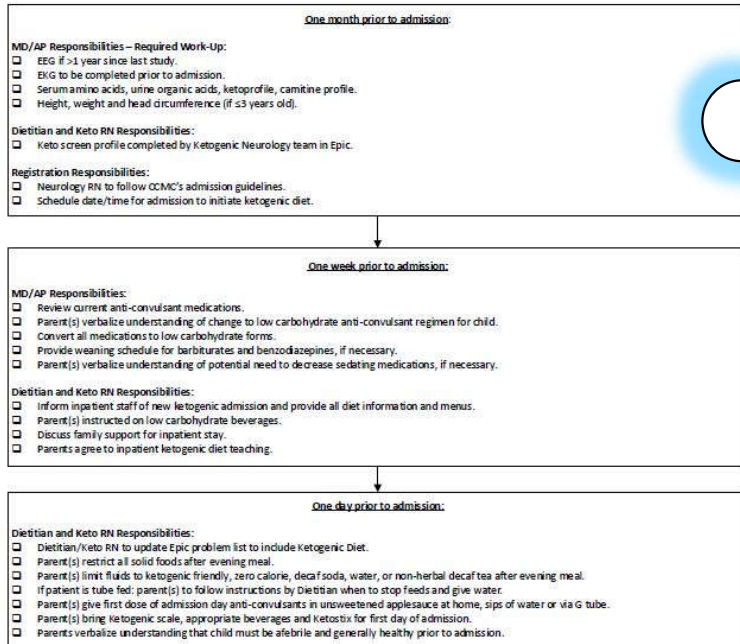
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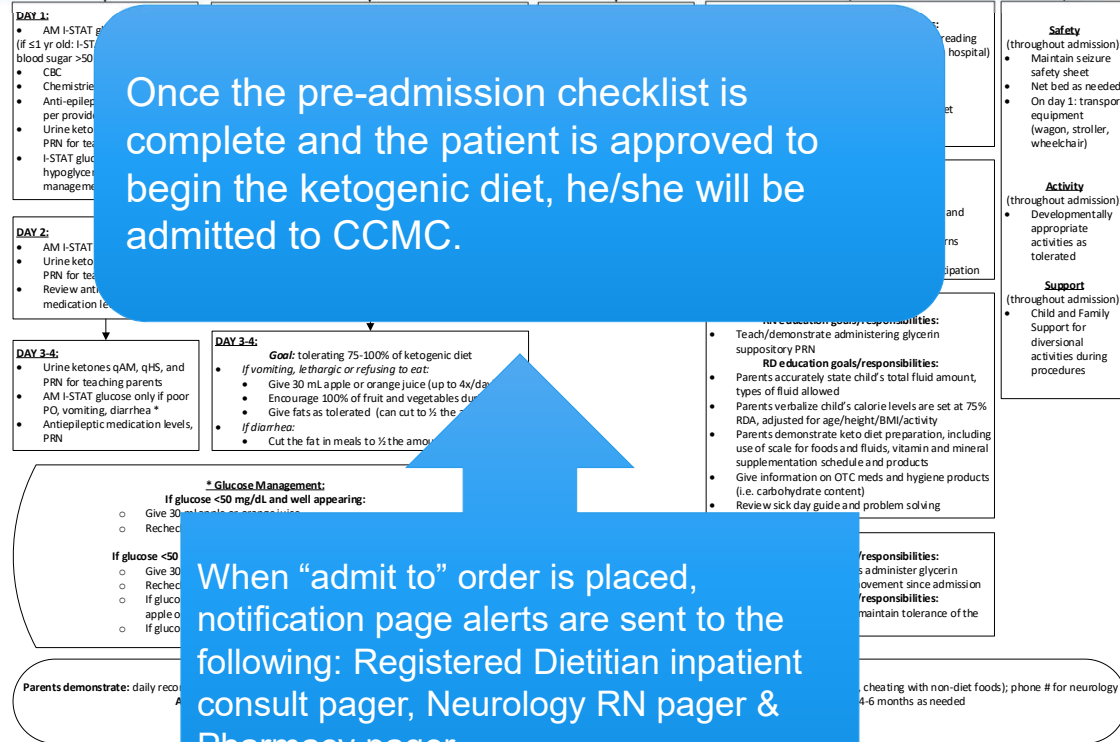
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Inclusion Criteria: Diagnosis of epilepsy, completed pre-admission checklist (Appendix A), approved by ketogenic team
Exclusion Criteria: Fatty acid oxidation defects, porphyria, carnitine deficiency syndromes, pyruvate carboxylase deficiency



Once the pre-admission checklist is complete and the patient is approved to begin the ketogenic diet, he/she will be admitted to CCMC.



When "admit to" order is placed, notification page alerts are sent to the following: Registered Dietitian inpatient consult pager, Neurology RN pager & Pharmacy pager

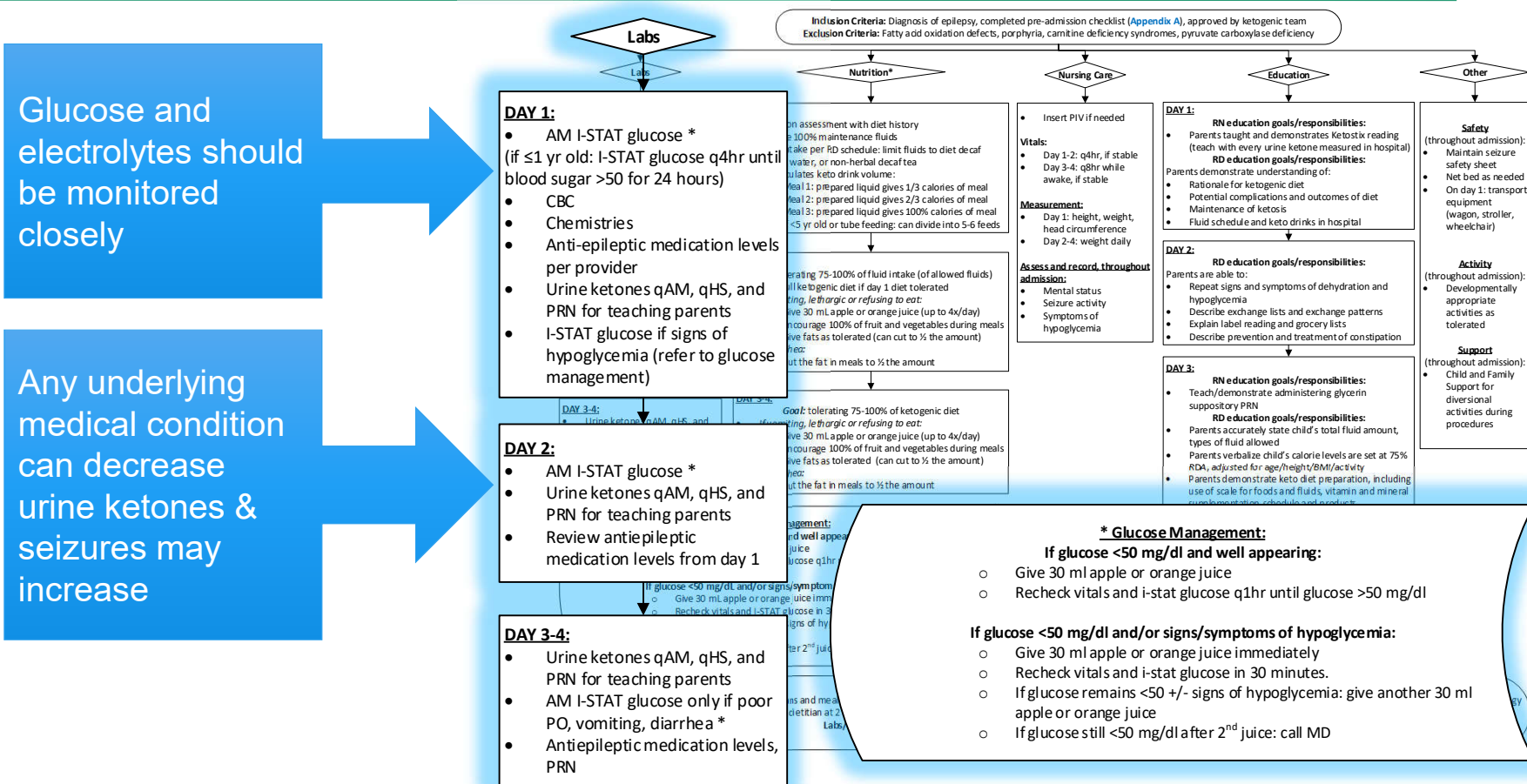
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Labs: Keto and Modified Keto



Glucose and electrolytes should be monitored closely

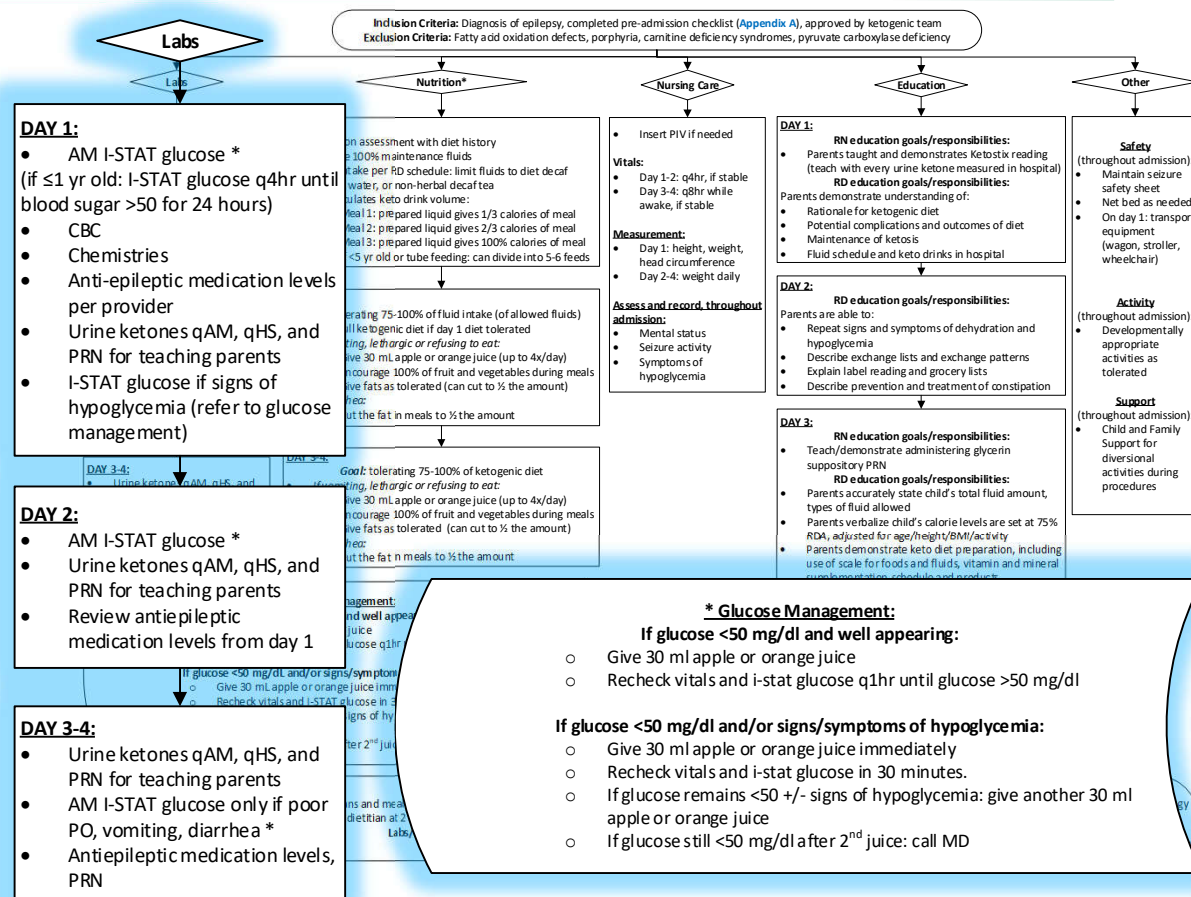
Any underlying medical condition can decrease urine ketones & seizures may increase

Labs: Keto and Modified Keto



There are special blood glucose guidelines for infants (patients 1 year old or younger):

- If glucose is less than 50mg/dl
 - Check an istat glucose every 4 hours until blood sugar is greater than 50 for 24 hours
- Follow protocol for glucose management



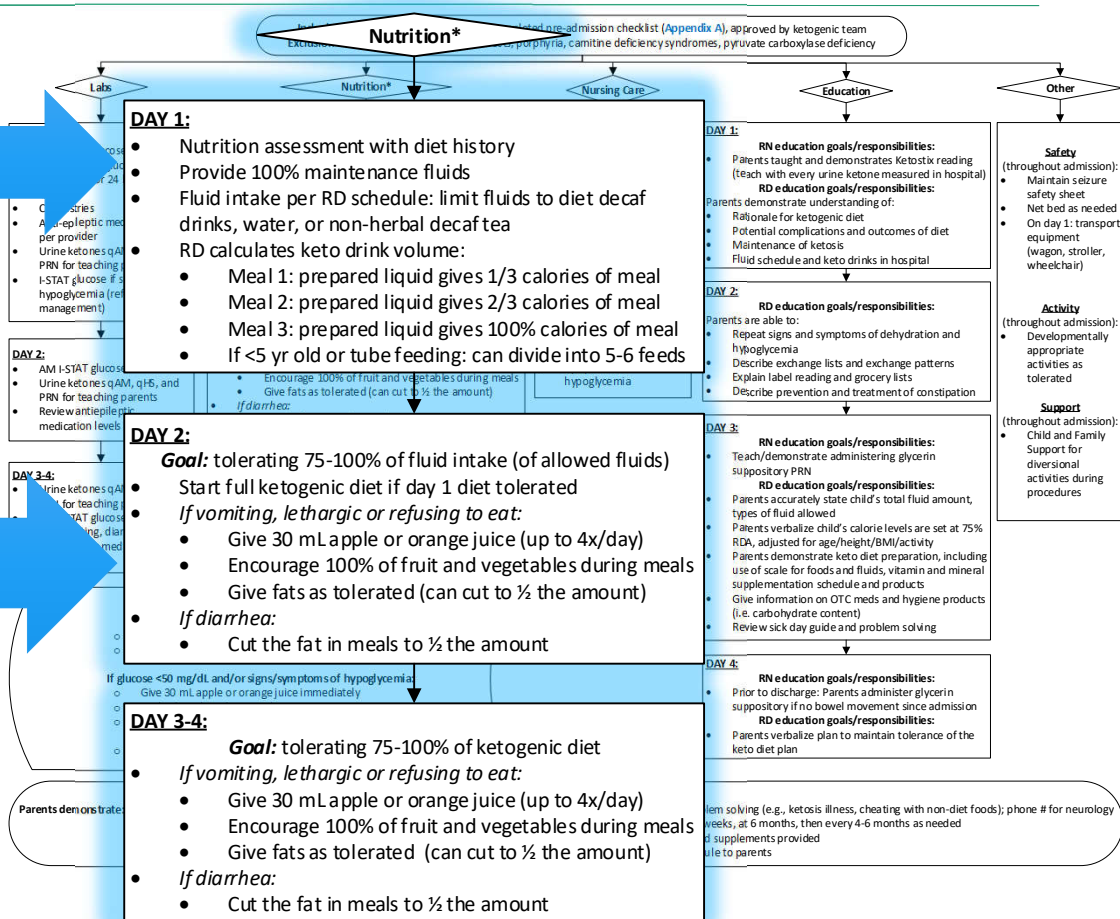
Nutrition: Keto



Ketogenic or Modified Ketogenic diets can be chosen from diet orders in EPIC.

Patients are at higher risk for hypoglycemia, dehydration and acidosis.

Small amounts of juice 1 oz. 3-4 times per day maybe required if not tolerating regular meals



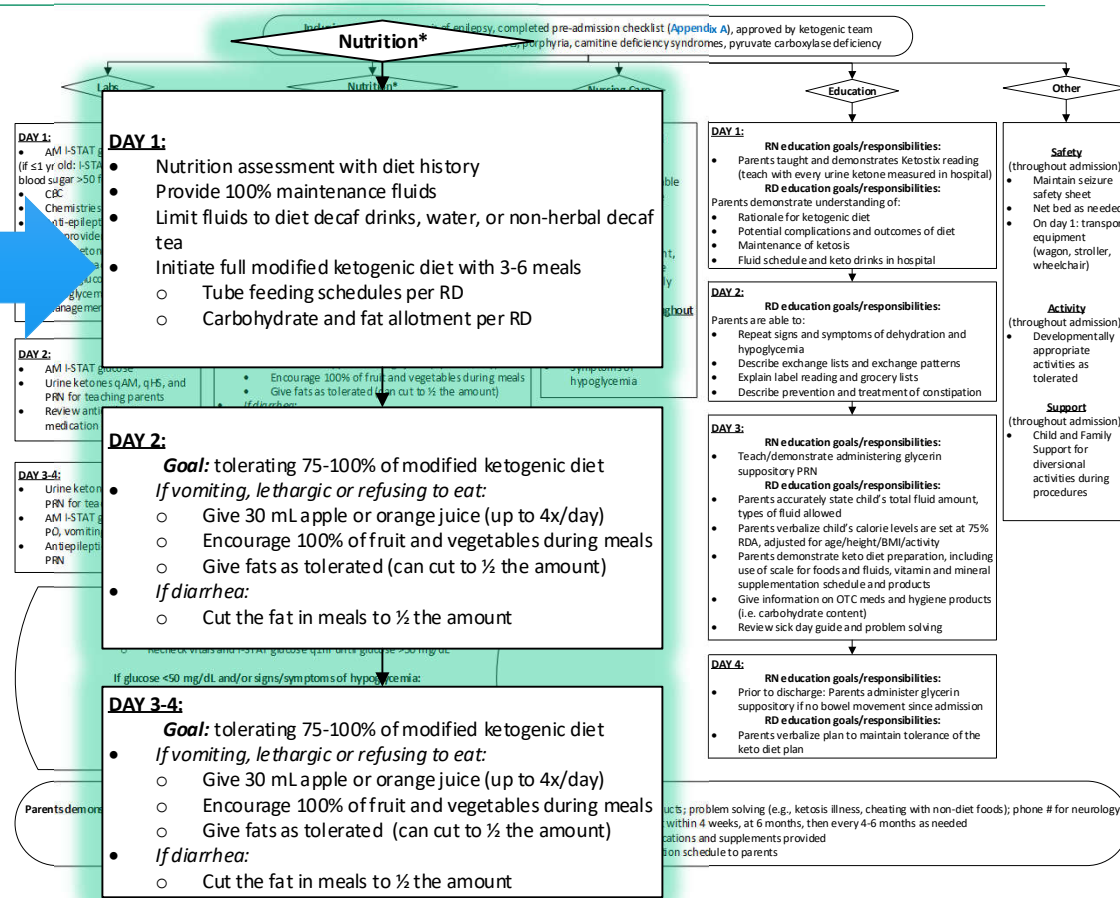
Nutrition: Modified Keto



Ketogenic or Modified Ketogenic diets can be chosen from diet orders in EPIC.

The Modified Keto Diet is initiated fully over 3-6 meals, instead of as a liquid diet

Goals for monitoring tolerance and glucose management are the same between the two pathways

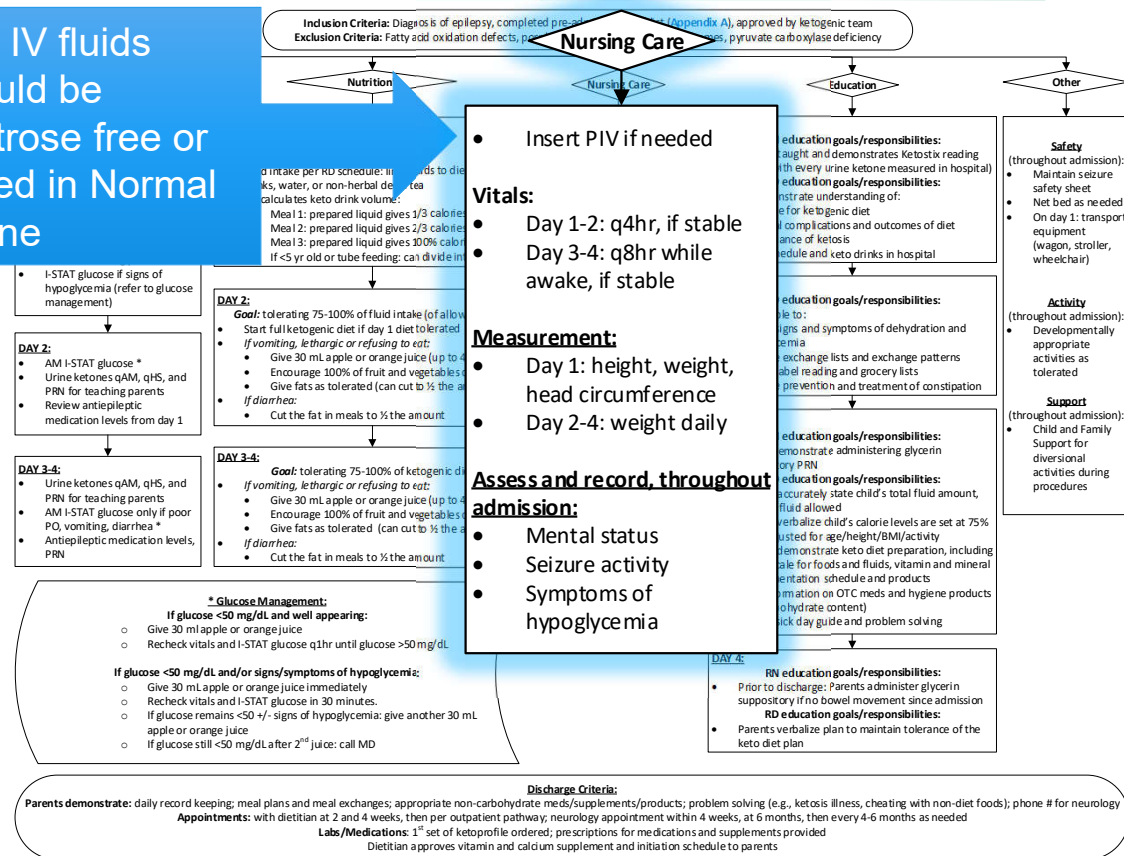


Nursing Care: Keto and Modified Keto



- Nursing should double check all medications given to ensure they are in sugar free solutions.
- Neurologic exams should be monitored throughout admission.

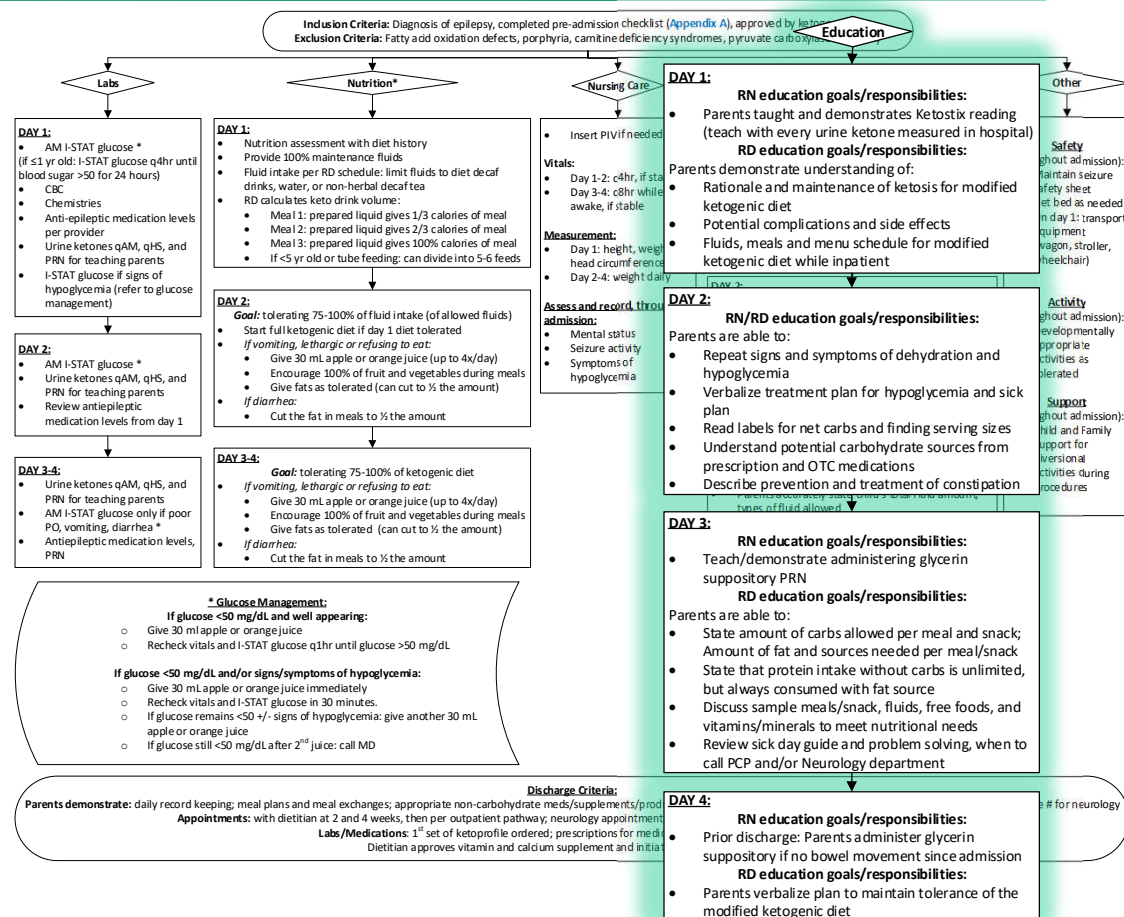
Any IV fluids should be dextrose free or mixed in Normal Saline



Education: Modified Keto



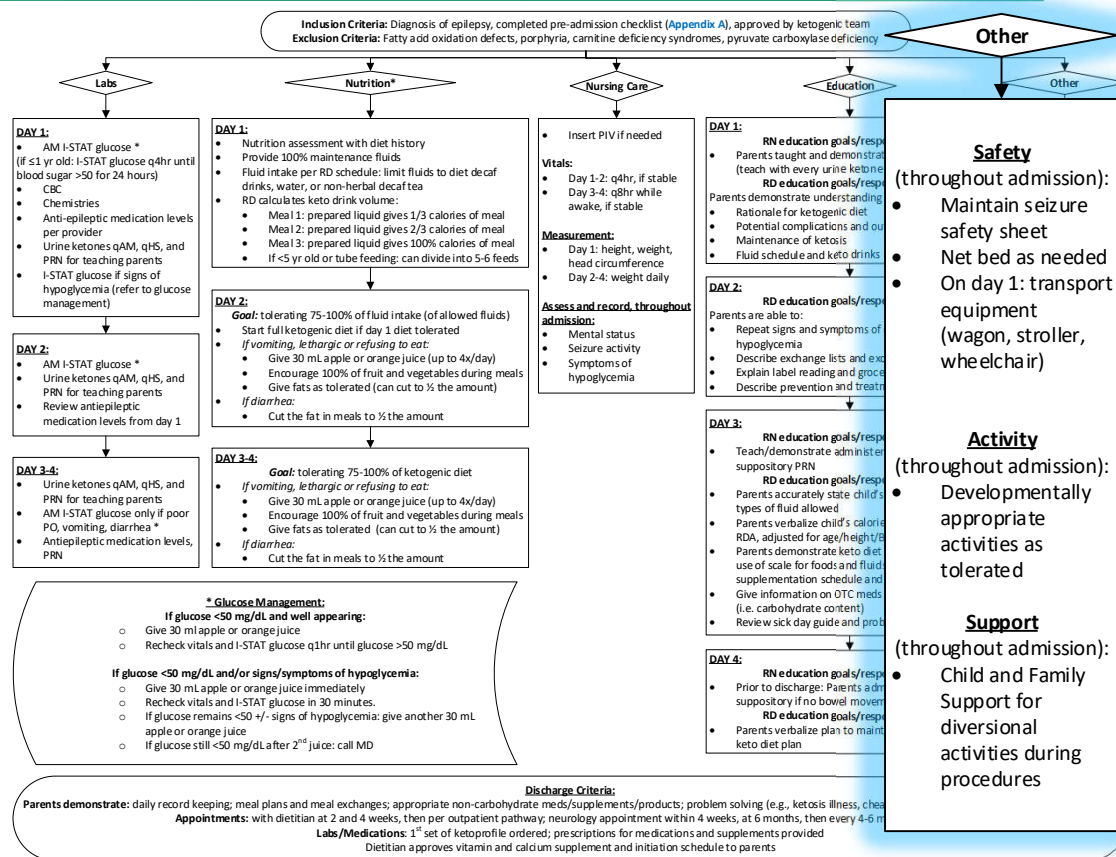
Daily Education is very important in both pathways. Most of the education goals and responsibilities are similar between the two, but will vary based on each child's specific dietary requirements.



Other: Keto and Modified Keto



All patients should have the appropriate safety, activity and support standards.

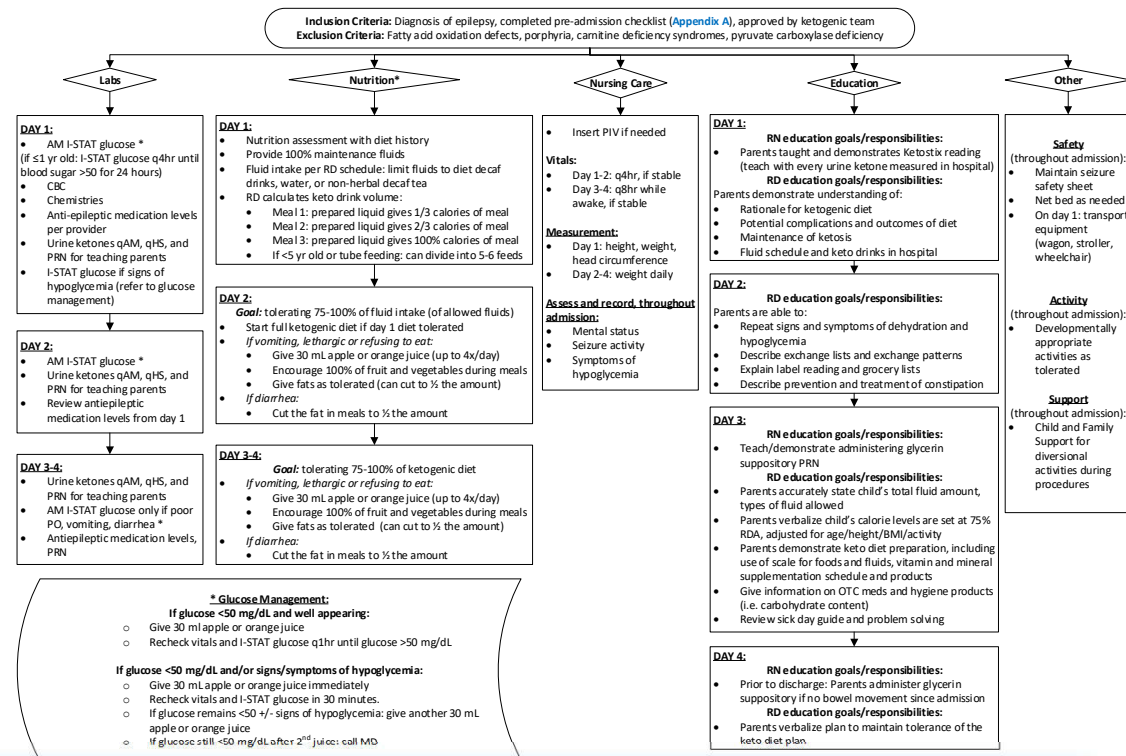


Discharge Criteria: Keto and Modified Keto



Also need:

- Appointments scheduled with:
 - Dietitian at 2 and 4 weeks AND
 - Neurology at 4 weeks and 6 months
- 1st set of ketoprofile labs ordered
- Prescriptions for meds and supplements
- Dietitian approved vitamin and calcium supplements and initiation schedule



Discharge Criteria:

Parents demonstrate: daily record keeping; meal plans and meal exchanges; appropriate non-carbohydrate meds/supplements/products; problem solving (e.g., ketosis illness, cheating with non-diet foods); phone # for neurology

Appointments: with dietitian at 2 and 4 weeks, then per outpatient pathway; neurology appointment within 4 weeks, at 6 months, then every 4-6 months as needed

Labs/Medications: 1st set of ketoprofile ordered; prescriptions for medications and supplements provided
 Dietitian approves vitamin and calcium supplement and initiation schedule to parents

Using Medications on a Ketogenic Diet



- Did you know ketogenic patients have a 6th vital sign?? “Medication Carbohydrate Content”
- Carbohydrates are hidden in many medications, typically liquids and chewable tablets.
- Sugar-free does not mean carbohydrate-free.
- Suppositories, caplets, and adult tablets are **less likely** to contain high amounts of carbohydrates.
- Lexicomp contains carbohydrate content of medications and pharmacy can double check.

Tips and Tricks for Appropriate Medications



- NO liquids.
 - NO chewable medications.
 - NO Dextrose, Glucose, or Lactate Ringers Solutions.
 - When in doubt, check with Pharmacy.
 - Discharge options: If the tablet form cannot be crushed, some specialty pharmacies can make a carbohydrate-free compounded form of most medications (using ORA PLUS or ALMOND OIL ONLY).
 - Suppositories are available for fever and pain relief in infant and pediatric versions.
-

Ketogenic Diet Illness Guidelines



The two most important items for families:

(1) recognizing dehydration and hypoglycemia

(2) when to contact pediatrician and Keto team

- If signs and symptoms of hypoglycemia are seen:
 - provide 1oz of juice 3-4 times a day if child is refusing or not tolerating meals.
- Monitor child for signs of dehydration.
 - Three wet diapers in a 24 hour period and pale colored urine (like lemonade) are positive signs of hydration. Drink small amounts of liquids every hour.
- It is common for urine ketones to fluctuate when a child is sick, ranging from their usual to negative.
 - No treatment to improve ketones is required during illnesses.

Ketogenic Diet Illness Guidelines



- **Fever:**

- Give only sugar-free fever reducing medication. It needs to be in capsule or tablet form, not liquid.
- All suppositories are also available over the counter.

- Offer plenty of sugar free fluids (G-2 Gatorade, unflavored Pedialyte, water, fruit 2-O, flavored waters, sugar free Jell-O, broth).

- Unflavored Pedialyte is okay for up to 24 hours, but meals should then be started at $\frac{1}{4}$ or $\frac{1}{2}$ strength. Tolerance should be evaluated at every meal. No child should stay on Pedialyte alone for more than 24 hrs.

- If vomiting or diarrhea:

- The fat content of meals can be titrated at every meal: $\frac{1}{4}$, $\frac{1}{2}$, $\frac{3}{4}$ to full-strength fat, to assess tolerance.

Review of Key Points



- Infants 1 year of age and less with hypoglycemia need to have blood sugar tested every 4 hours until it is normal for 24 hours.
- Parental education and medication checks are important to ensure no carbohydrates are given while on the ketogenic diet.
- Parents should know:
 - how to treat dehydration and hypoglycemia
 - when to call the PCP or keto team.

Quality Metrics



Ketogenic Diet

- Percentage of patients with pathway order set
- Percentage of patients who tolerate the diet by day 3
- Percentage of patients with education completed by day 3
- Percentage of patients discharged by day 3
- ALOS

Modified Ketogenic:

- Percentage of patients with glucose >50 for all 3 days
 - Percentage of patients with development of diarrhea
 - Percentage of patients with development of vomiting
 - Percentage of patients with education completed by day 3
-

Pathway Contacts



- Jennifer Madan Cohen, MD
 - Pediatric Neurology
- Jamie Cubanski, RN
 - Pediatric Neurology
- Beth Chatfield, RD
 - Pediatric Neurology

References



- <https://charlifoundation.org>
- Kossoff EH, Zupec-Kania BA, Auvin S, et al. [Optimal clinical management of children receiving dietary therapies for epilepsy: Updated recommendations of the International Ketogenic Diet Study Group](#). *Epilepsia Open*. 2018 May;3(2):175-192.

Thank You!



About Connecticut Children's Clinical Pathways Program

Clinical pathways guide the management of patients to optimize consistent use of evidence-based practice. Clinical pathways have been shown to improve guideline adherence and quality outcomes, while decreasing length of stay and cost. Here at Connecticut Children's, our Clinical Pathways Program aims to deliver evidence-based, high value care to the greatest number of children in a diversity of patient settings.

These pathways serve as a guide for providers and do not replace clinical judgment

This Educational Module was edited by:

Abby Theriaque, APRN

Educational Module Specialist
