1.0 Introduction

The Ketogenic Diet therapy is a nonpharmacological treatment used to treat children with drug resistant epilepsy when traditional pharmacological treatments do not adequately control seizures. The ketogenic diet therapy is a low carbohydrate, adequate protein, and high fat diet that puts the child in a state of ketosis which helps control seizures.

Most children who have epilepsy will respond to pharmacological treatment. However, 20-30% will develop drug resistant epilepsy. It is this “non-responder” population that may be considered as candidates for the ketogenic diet therapy.

At Sick Kids, the Ketogenic Diet Team (Keto Team) manages all children who are being considered for, started on and/or managed by the ketogenic diet therapy to control seizures. This clinical practice guideline (CPG) has been developed by the Keto Team to facilitate safe and effective management of these children and families.

Purpose:

1. Ensure safe and consistent care for children on a ketogenic diet therapy;
2. Streamline the referral process to the SickKids Keto Team to ensure optimal and timely care for the patient; and
3. Provide recommendations for health care providers at SickKids and in the community for the care and requirements of patients on the ketogenic diet therapy.

There are five main sections of this guideline that focus on patient care at SickKids and recommendations for care when SickKids’ patients are at home and being cared for in the community. These include:

1. Determining eligibility for the ketogenic diet therapy;
2. Ketogenic diet therapy initiation;
3. Inpatient and Emergency Department Care;
4. Caring for the ketogenic diet patient in the community; and
5. Information and education for patients and families.
Target Users

- All clinical staff at SickKids
- Referring Neurologists in the community or in hospital and Pediatricians in areas (where no neurologist is available)
- Community health care professionals who care for patients on the ketogenic diet.

*Please Note: Notify the Ketogenic Diet Team for all inpatients admitted to Sick Kids who are already on the ketogenic diet. Contact the Ketogenic Diet team at x 201785.*
Section 1: Patient Eligibility & Pre-diet Evaluation

1.1 Patient Eligibility
- Does your patient meet the inclusion criteria for the ketogenic diet?
- Does your patient have any other medical conditions that would exclude them as a candidate for the ketogenic diet?

1.2 Referral Process
- SickKids Inpatient: Ketogenic Team NP Consult X 207722 /page 416-242-0253
- NEW Referrals via ARMs* (From community and other SickKids’ clinics)

1.3 First Ketogenic Diet Clinic Visit
- SickKids Ketogenic Team First Visit Assessment
- Laboratory and diagnostic workup

Section 2: Diet Initiation

2.1 Basic Principles for Diet Initiation

2.2 Inpatient Initiation
- Pre-admit prep for IP start
- IP diet initiation

2.3 Outpatient Initiation
- Pre-admit prep for OP start
- OP diet initiation

*ARMs: Ambulatory Referral Management system is the SickKids online referral database where all referrals must be processed
Section 3: Ongoing monitoring of the Ketogenic Diet

3.1 Initial Post-Initiation Monitoring
- SickKids Ketogenic Diet Clinic at months 1 and 3 and 6 post-initiation
- Team follow-up phone calls/e-mail as required by patient

3.2 Routine Monitoring at 3-6 Month Intervals
- Nutritional, Medical and Social re-assessments

3.3 Laboratory Investigations
- Blood and Urine investigations

3.4 Other Diagnostic and Imaging Interventions
- Other monitoring including Nephrology, Cardiac, Bone Health and when to refer for a consultation

3.5 Discontinuing the Ketogenic Diet

Section 4: Caring for the Ketogenic Diet Patient

4.1 General Principles - Caring for your Ketogenic Diet Patient
- Ask for the patient’s “Discharge Letter” and “Nursing/Physician Information” sheets outlining their Ketogenic Diet plan
- Medications to ALWAYS avoid

4.2 Inpatient & ED Care
- Urine and blood monitoring
- NPO guidelines
- Ketogenic diet patients and General Anesthesia
- Vomiting Guidelines

4.3 Care by the Community MD
- Family education and resources
- Medications to avoid
- Utilizing pharmacy expertise
- Contacting the Ketogenic Team

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Ketogenic Diet Overview

The ketogenic diet is a non-pharmacologic nutrition medical treatment used to treat drug-resistant epilepsy.\(^1,6\)

There are three types of diet therapies used to treat epilepsy at SickKids:

1. The classic ketogenic diet
2. The medium chain triglyceride (MCT) ketogenic diet
3. The modified Atkins diet (MAD)

For a detailed explanation of these diets, click here.

Section 1: Patient Eligibility & Pre-diet Evaluation

1.1 Patient Eligibility

1.1.1 Children with drug-resistant epilepsy who have been tried on two adequately dosed antiepileptic medications and have no contraindications to diet therapy are eligible to be treated with the ketogenic diet. This includes inpatients in status epilepticus or who cannot be discharged home due to unstable seizures.

1.1.2 Any clinician considering a referral for a patient for the ketogenic diet should review the indications and absolute contraindications listed in Table 1. Please also refer to the SickKids algorithm determining eligibility of a patient for the Ketogenic Diet.

1.1.3 Patients who meet the eligibility criteria can be referred to the Ketogenic Diet Clinic. Refer to section 1.2 for referral processes.

1.1.4 The ketogenic diet requires a significant economic and time commitment by the family. For diet therapy to be successful, families must be able to commit the time and effort required to prepare the diet. Additionally, consideration to the ongoing added expenses for families that are not covered by OHIP or extended health care plans should be reviewed and in place before starting the diet.
**Table 1. Indications and Contraindications for the Ketogenic Diet**

### Indications for Ketogenic Diet
- Children with seizures that are not controlled by drugs should be evaluated to determine if they are a candidate for epilepsy surgery or the ketogenic diet.
- The ketogenic diet may be particularly helpful for certain metabolic conditions, such as GLUT-1 deficiency syndrome and Pyruvate dehydrogenase deficiency.

### Contraindications to Ketogenic Diet
- Certain metabolic conditions may be a contraindication to the ketogenic diet:
  - B-oxydation defects
  - Porphyrias
  - Carnitine deficiency (primary)
  - Carnitine palmitoyltransferase (CPT) I or II deficiency
  - Carnitine translocase deficiency
  - Medium-chain acyl dehydrogenase deficiency (MCAD)
  - Long-chain acyl dehydrogenase deficiency (LCAD)
  - Short-chain acyl dehydrogenase deficiency (SCAD)
    - Long-chain 3-hydroxyacyl-CoA deficiency
    - Medium-chain 3-hydroxyacyl-CoA deficiency
    - Pyruvate carboxylase deficiency
- Children with any disorder that requires them to have a diet high in carbohydrates.
- Certain drugs may destabilize ketosis, including steroids, which may require further adjustment of the ketogenic diet.
- Valproic acid should not be used in combination with MCT oil as this may cause hepatotoxicity
- Patient or caregiver noncompliance
- Inability to maintain adequate nutrition
- Children with frequent vomiting/feeding intolerance or history of known aspiration will require further investigations
1.2 Referral Process

SickKids Inpatient Referrals

1.2.1 All inpatients under consideration for an urgent ketogenic diet initiation should have a consultation with the Ketogenic Diet Team. To initiate this consult, phone extension 201785 Monday to Friday 8:30-4:30.

New Outpatient Referrals

1.2.2 All patients must be referred to the Ketogenic Diet Clinic for assessment prior to initiation of diet treatment via ARMs by their Neurologist or Community Paediatrician when there is no local neurologist.

1.2.3 ARMs criteria are detailed below. Full completion of all items is required to be able to promptly process this referral.

<table>
<thead>
<tr>
<th>Table 2: ARMs Referral Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Referrals will be accepted for the following patients (ARMs):</td>
</tr>
<tr>
<td>□ Epilepsy unresponsive to standard antiepileptic drug treatment</td>
</tr>
<tr>
<td>□ Metabolic conditions for which the ketogenic diet is the primary treatment.</td>
</tr>
</tbody>
</table>

Pre-visit Requirements

□ A detailed referral letter describing the patient’s seizures (type and frequency), current and past antiepileptic drugs, investigations and any medical history

□ EEG

□ Feeding assessment if child has known feeding problem

□ Lab results – lactate, AST, ALT, alk phos, urine organic acids, urine and serum amino acids, carnitine, CBC

□ MRI of brain

**In some cases there will be exceptions to these requirements. Please discuss with the ketogenic diet team.

Referrals Not Accepted

□ Patients with recurrent aspiration pneumonia or a fatty acid oxidation defect

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1.3 Ketogenic Pre-Diet Clinic Visit

1.3.1 All patients who will be seen in clinic to be assessed for the ketogenic diet should receive the pre-consultation assessment prior to the appointment day.

- Pre-diet Questionnaire
- Parent information booklet
- Canada Food Guide

1.3.2 The ketogenic diet can have a significant impact both socially and financially on the patient and family. Early recognition of potential social and financial barriers is important to ensure timely access to support. Through use of the pre-diet questionnaire potential social and/or financial barriers will be flagged. A proactive approach to uncovering these issues will set the stage for greater success with diet initiation and ongoing management. Families who require specific assistance in these areas can schedule an appointment with a Social Worker by contacting the Social Work Neurology Single Session Clinic. The contact information is:
  phone: 416-813-7654 ext. 201190 or email: neurology.singlesessionclinic@sickkids.ca.

Outline of costs
- Table of funding opportunities

1.3.3 An Information Session is offered four times a year for parents to attend. This provides detailed information about the diets and the necessary commitment to maintain child on the ketogenic diet.

1.3.4 Patients under consideration for the ketogenic diet will attend the SickKids Ketogenic Diet clinic for an interdisciplinary assessment. This appointment includes:
  - Medical History and Physical Examination
  - Nutrition Assessment
  - Social / Financial Assessment

2.1 General Principles

2.1.1 The majority of patients initiating the classic and MCT ketogenic diet are started with an inpatient admission. The length of stay will vary from 2-4 days depending on the patient's tolerance and safety with their new diet. Outpatient initiations are an options and done on a case-by-case basis.

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2.1.2 The goal during the initiation of the ketogenic diet is to establish an adequate state of ketosis. The purpose of the inpatient admission is to:

1. Monitor for potential acute complications such as hypoglycemia, excessive ketosis, emesis, diarrhea and/or dehydration;
2. Educate Parents/Caregivers around how to manage the diet at home and how to treat potential complications;
3. Ensure Parents/Caregivers can safely manage the diet at home;

2.1.3 Patients who have been on the classic or MCT diet before and are restarting the diet may be started as an outpatient. These diets will be started at a lower level of dietary restriction (lower diet ratios and/or percentage of MCT) to reduce the risk of potential complications. It will be gradually advanced based on patient's tolerance and needs. The goal is to achieve ketosis in less than one week. Prior to starting the diet, a teaching session will be booked with the dietitian and the family/care giver to review the diet and how to manage it at home.

2.1.4 The MAD is started on an outpatient basis. The family/care givers and the patient, when appropriate attend a 3-hour teaching session with the Ketogenic Team dietitian and nurse practitioner. The family initiates the diet at home.

2.1.5 All patients who are starting the ketogenic diet should have a baseline EKG, abdominal ultrasound and fasting blood work panel completed prior to diet initiation.

Laboratory Investigations are as follows: The following complete biochemical index should be assessed prior to initiation of the diet and every 6 months at follow up visits when the patient is on the KD 

- blood urea nitrogen (BUN)
- creatinine
- uric acid
- alkaline phosphatase
- aspartate transaminase (AST)
- alanine transaminase (ALT)
- complete blood count (CBC) with differential
- fasting blood glucose
- calcium
- phosphorous
- magnesium
- fasting lipid profile
- electrolytes
- trough antiepileptic drug levels
- total protein, albumin
- antiepileptic drug (AED) levels,
- serum iron, ferritin, transferrin
- red blood cells (RBC) folate
- vitamin E
- 25-hydroxy vitamin D
- zinc
- selenium
- total/free carnitine
- PTH
- venous gases
- urine: creatinine, osmolality, calcium, citrate

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2.2 Inpatient Initiation of the Ketogenic Diet

Pre-admission preparation for an inpatient start

2.2.1 A patient will be pre-booked to come onto 5C to initiate the ketogenic diet. This informs the admitting epilepsy fellow or nurse practitioner, nursing and neurology staff that a patient is coming to start the ketogenic diet.

2.2.2 Prior to the inpatient admission, a phone call is made to the family/caregivers from both the Keto Team dietitian and nurse practitioner. The following will be completed:

1. Assess patient's most up-to-date feeding pattern and anthropometric data
2. Answer last minute questions for the family
3. Ensure the family has all the necessary feeding and teaching supplies/equipment to bring to hospital. Required supplies include a decimal gram scale (weighs to 1/10th), urine ketostix, blood glucose monitoring equipment.
4. Adjust the current feeding schedule so the 12 hour fasting baseline blood work can be completed

2.2.3 Patients are instructed to stop all sweets one week prior to initiating the diet

Inpatient Diet Initiation

2.2.4 At admission, the dietitian will complete the nutrition assessment to determine the most appropriate KD for each individual. The diet is calculated the morning of the admission date so the food can be weighed and sent by 1200h on admission day. Throughout the admission, the dietitian will then adjust the diet if there are any complications such as vomiting, diarrhea, dehydration, excessive ketosis and hypoglycemia.

2.2.5 The dietitian assesses the blood work and the diet to determine each individual's micronutrients supplements and makes sure the patient/family/caregivers have the necessary formulas/diet supplies to manage the diet once at home.

2.2.6 During diet initiation, dietitians provide the following teaching to patients/parents/caregivers:

- identifying foods and products that are appropriate for the diet;

An alternate method of testing ketosis is to use a serum ketone monitor.
Ketogenic Diet

- how to calculate the diet and meal planning;
- how the diet prescription is made;
- how to weigh foods/formula;
- label reading, adequate vitamins/minerals supplements needed for the patient;
- how to deal with complications and potential problems;
- how to manage the Ketogenic diet during illness
- how to adjust the diet overnight if there are complications

2.2.7 Nurses teach patients/parents/caregivers how to measure blood glucose, urine ketone and specific gravity. In addition, nurses support the family to ensure the child is drinking adequately while monitoring ins and outs and advance the diet as per diet prescription. Care-by-parent is encouraged so family is comfortable managing the diet at home.

2.2.8 Patients are sent home with 3 follow-up appointments booked with the Keto Team at months 1 and 3 months and 6 months of being on the ketogenic diet.

2.2.9 Parents/caregivers are provided with the following at time of discharge:
- MCT or Classic Ketogenic diet booklet
- Discharge letter documenting patient’s ketogenic diet (Link to new template)
- Nursing/physician information sheet (Link to template)
- Copy of diet prescription
- Prescriptions for necessary supplies and some patients may need formula or MCT oil
- List of necessary micronutrient supplements
- Prescriptions for ketostix, glucometer, glucose strips and lancets

2.3 Outpatient Diet Initiation

Preparation for an outpatient start

2.3.1 A teaching session is booked with parents/caregivers and patients (if appropriate) for initial teaching of the MAD diet and when restarting the Classic or MCT Ketogenic diet.

2.3.2 Complete fasting blood requisition and prescription for Ketostix to give to patient/family.

2.3.4 For the MAD, this is started on an outpatient basis. The family/caregivers and sometimes the patient will attend a 3 hour teaching session with the dietitian and nurse practitioner.
Outpatient Diet Initiation

2.3.5 During the MAD teaching session the following teaching is provided to the parents/caregivers/patients, such as:

- Identifying foods that are appropriate for the diet;
- How to count net carbohydrate content of food;
- Label reading, adequate vitamins/minerals supplements needed for the patient;
- How to deal with complications and potential problems;
- Knowing what to do if child becomes sick;
- Meal planning

2.3.6 After the MAD teaching session, the parents/caregivers/patients are asked to demonstrate accurate carbohydrate counting and meal planning. Once this is completed, and the family is ready to start, they will initiate the diet at home and notify our team once it has started.

Parents are provided with the following at the end of the session:
- Ketogenic diet teaching booklet
- List of necessary micronutrients

2.3.7 Patients will be booked for 3 follow-up appointments with the ketogenic diet team at 1 month, 3 months and 6 months of being on the MAD.
Section 3: Ongoing Monitoring at SickKids KD Clinic

3.1 Initial post-diet initiation monitoring (Both inpatient and outpatient starts)

3.1.1 Once patient is discharged they will be followed through phone calls or email by a Ketogenic Diet Team provider as required, plus clinic visits at months 1 and 3. Follow up clinic visits will continue to be scheduled every 3-6 months depending on patient’s needs.

3.1.2 Prophylactic treatment to prevent kidney stones should be considered at 1-2 mmol/kg/d of potassium citrate.

3.2 Routine monitoring

3.2.1 Regular follow-up visits should be scheduled every 3-6 months. These visits should include:

- Nutritional Assessment
- Medical Assessment
- Social Assessment

3.3 Laboratory Investigations

3.3.1 See section 2.1.5 for routine blood work, which is completed at least every 6 months.

3.3.2 All lab value norms may vary from each individual lab. Abnormal biochemical indexes need to be repeated in a time frame of by the next appointment or earlier at the discretion of the clinical team. The diet’s calorie, protein, vitamins and minerals supplementation are readjusted according to the patient’s laboratory value in combination of the full interdisciplinary clinic assessment, while the patient is on the KD.

3.3.3 Anyone on potassium citrate, blood work should be done at 1 and 3 months to check potassium and TCO2 to ensure tolerance. Urine levels of citrate, calcium, osmolality and creatinine to be tested at 3 months and dosage of potassium citrate adjusted accordingly.

3.4 Other Diagnostic and Imaging interventions & Subsequent Referrals

3.4.1 ECG, Kidney Ultrasound, DEXA Scans should be performed at least annually or more frequently if clinically indicated.

3.5 Discontinuing the Ketogenic Diet

3.5.1 Patients discontinue the Ketogenic diet at the discretion of the Ketogenic diet team and in collaboration with the family. Possible reasons to discontinue the ketogenic diet include:

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**Ketogenic Diet**

- Persistent or worsening complications from the KD Therapy (i.e. fatty liver, gastroesophageal reflux, arrhythmia, osteopenia, neutropenia)
- Parents and/or Patient no longer wish to continue with the ketogenic diet
- Patient is not receiving adequate seizure control from the ketogenic diet

3.5.2 There is additional consideration prior to weaning Patients with Glut 1 and PDH, given this is the preferred treatment for these conditions. In the event that weaning is being considered, the metabolic team should be consulted.

3.5.3 The length of time that a patient is sustained on the ketogenic diet varies greatly. Patients will remain on the ketogenic diet for a minimum of 3 months to ensure an adequate trial with the Ketogenic diet.

3.5.4 If patients have a satisfactory response to the ketogenic diet, they may stay on the diet for as long as they do not have adverse events and the parents/patients are managing the ketogenic diet safely at home.

3.5.5 Once a decision to wean the ketogenic diet is made, the duration of weaning is recommended to be at least 2-3 months by gradually lowering the Ketogenic diet ratio, but is often done on a case by case basis.

3.5.6 In some cases, a patient may be rapidly weaned. This is to be done in consultation with the treating medical team and ketogenic diet team.

### 4.0 Care of the Patient on a Ketogenic Diet

#### 4.1 General Principles when caring for your KD Patient

4.1.1 Ask the patient and/or caregiver for the “Discharge Letter” or “Nursing/Physician Information” sheet and about patient’s KD regimen. Families have been instructed to bring a few meals of foods/formula, diet orders and menus to the hospital. Diet regimens include required daily fluid intake. Menus can be submitted to the diet office. Refer to the Ketogenic Diet order set for admission orders.

4.1.2 **All oral medications must be in tablet form.** Suspensions, syrups and chewable tables have excess carbohydrates. Sugar free liquids are not permitted as they may contain carbohydrates. When in doubt, discuss with hospital pharmacist.

4.1.3 **No IV Dextrose solutions**, including premixed I.V medications, unless absolutely necessary

4.1.4 When starting any form of steroid therapies, including topical treatments, or blood products, such as IVIG and albumin that contain a high protein load, consult the Keto team. WARNING: **These may negatively impact** ketosis and may result in seizures.

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4.1.4 Many topical creams are high in carbohydrates and may interfere with therapy. Check with Keto Team member or hospital pharmacist prior to starting any new topical preparation.

PATIENT SAFETY ALERT:
- Suspensions, syrups and chewable tablets have excess carbohydrates. Sugar free liquids are not permitted as they may contain carbohydrates.
- Steroid therapy or blood products, such as albumin and IViG may negatively impact ketosis and may result in seizures. The diet may be adjusted in an effort to compensate for this. Please notify the treating team if this treatment is to be started.
- When in doubt, discuss with a Keto Team member or the hospital pharmacist.

4.2 Inpatient and Emergency Department Orders

4.2.1 Check urine ketones with every void, at least twice per day.

<table>
<thead>
<tr>
<th>If the urine ketone level is...</th>
<th>Intervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 16 mmol/L (&lt;160 mg/dl)</td>
<td>No action required</td>
</tr>
<tr>
<td>&gt;16 mmol/L (&gt;160 mg/dl),</td>
<td>Treat immediately with 15 ml of orange juice or 10 ml of apple juice.</td>
</tr>
<tr>
<td>(turns to dark purple before 40 sec on multistix OR 15 sec on ketostix)</td>
<td>If NPO treat with 25mL D5W</td>
</tr>
<tr>
<td>at 16 mmol/L (≥160 mg/dl),</td>
<td>Treat immediately with 7 ml of orange juice or 5 ml of apple juice unless specific instruction from ketogenic diet team or parent letter then treat on a case by case basis</td>
</tr>
<tr>
<td>(turns to dark purple at 40 sec on multistix OR 15 sec on ketostix)</td>
<td>If NPO treat with 12.5mL D5W</td>
</tr>
</tbody>
</table>

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Target ketone range varies with each Pt.  
Ask families for the Patient’s usual ketone range. Notify the ketogenic diet team if Patient’s ketones are outside of this range.

**Please note, volume of juice given for treatment is independent of body weight.**

If patient is testing serum ketone levels, please refer to patient’s individual treatment plan for how to treat elevated serum ketones.

4.2.2 Measure urine specific gravity with each void. Urine specific gravity should be less than 1.030.
- If urine gravity is greater than 1.030 increase fluid intake to ensure hydration. Ask Parents for the patient’s maximum daily fluid intake.

4.2.3 Check blood glucose at least every 6 hours for ill patients. Check twice daily for stable patients; prior to breakfast and dinner or first and last G-tube feed.

<table>
<thead>
<tr>
<th>If the blood glucose is...</th>
<th>Intervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 2.5 mmol/L or if symptomatic for hypoglycemia</td>
<td></td>
</tr>
</tbody>
</table>
- Treat immediately with 30 mL of orange juice or 20 mL of apple juice, and recheck blood glucose within 20 minutes. If blood glucose is still < 2.5 mmol/L, treat immediately with 15 mL of orange juice or 10 mL of apple juice.  
- If NPO treat with 50 mL D5W, and recheck blood glucose within 1 hour. |

| 2.5-3.0 mmol/L |  
- Treat immediately with 15 mL of orange juice or 10 mL of apple juice. Recheck blood sugar in 1 hour.  
- If NPO treat with 25 mL D5W |
**Please note that volume of juice given for treatment is independent of body weight.**

### 4.2.4 Ketogenic Diet NPO Guidelines (used for illness, minor procedures)
- Use IV normal saline for maintenance fluids.
- No IV Dextrose solutions unless absolutely necessary. If necessary, use ½ or ¼ strength. (to be discussed and approved by neurologist or keto team).
- Check blood sugar at least every 6 hours.
- Check urine ketones with each void.

### 4.2.5 Patient Undergoing General Anaesthesia who follows a ketogenic diet (when admitted to hospital and unable to achieve full feeds within 24 hrs)

**Please notify the ketogenic diet team at ext 201785 if you have a patient admitted for who this applies**

**Pre-op**
- Finish last formula the night before by midnight
- Provide ½ strength Pedialyte up to 3 hours before procedure and then NPO.

**During Procedure**
- Maintenance fluid during the OR, preferably normal saline (unless otherwise indicated).
- (For G-tube insertion) When glucagon is used, extra seizures may occur. Extra monitoring of blood glucose (BG) is required. With the use of glucagon, BG levels are checked every 30 mins for at least 4 hours post-procedure and then every 1 hour as per IGT.
- Maintain normoglycemia and monitor for acidosis during the procedure (see details for treatment below).
- No liquid medications (suspensions containing sugar and/or sweeteners)

**Recovery**
- Run I.V. normal saline for maintenance fluids,
- No liquid medications (suspensions containing sugar and/or sweeteners)
Maintain normoglycemia and monitor for acidosis during the procedure (see details for treatment below).

**Unit**

- The ketogenic diet team will provide guidance and/or order the necessary I.V. fluids to manage prolonged NPO periods post procedure.
- Refer to RD order for clear fluids via G-tube. Maintain TFI.
- Refer to RD order for diet advancement post-procedure. Do not exceed TFI.
- No liquid medications (suspensions containing sugar and/or sweeteners)
- Maintain normoglycemia and monitor for acidosis during the procedure (see details for treatment below).

**Treatment of Hypoglycemia and Excessive Ketosis**

Monitor urine ketones every void.

Please refer to above parameters for treatment.

Monitor blood glucose every 3 hours (see above note for gastrostomy insertion with the use of glucagon for the schedule of more frequent BG checks).

Please review to above parameters for treatment.

### 4.2.6 Ketogenic Diet Vomiting Guidelines

- Treat vomiting with dimenhydrinate suppositories. Use after 1st vomit.
- Discontinue ketogenic diet foods/formula
- If oral rehydration is required, use ½ strength pedialyte or 30% strength enfalyte
- Measure urine ketones, urine specific gravity and blood sugar as indicated in general guidelines.
- Re-introduce ketogenic diet foods/formula gradually when vomiting stopped
5.0 Implementation of CPG

- *Daily News* story to alert the SickKids community that there is a Ketogenic Diet CPG
- Target the health care professionals and teams who collaborate in the care of children on the ketogenic diet via e-mail to make them aware of the new Ketogenic Diet CPG (e.g. PICU, General Paediatrics, Complex Care Team, Gastroenterology, Emerg, Neurology, Image Guided Therapy, Anesthesia, 5C Neuroscience Unit Nurses, Pharmacists).
- Send out an e-mail blast to the members of the Division of Neurology regarding the Ketogenic diet CPG with a link to the CPG.
- Offer in services to various subspecialties to review the CPG.

6.0 Evaluation of CPG

The Ketogenic Diet Team will evaluate compliance with the new Ketogenic Diet CPG by tracking the following:

- # of safety reports that are filed regarding a child on the ketogenic diet as it pertains to adherence to the diet parameters.
- # of consultations with the member of the Ketogenic Diet Team to guide patient management in the event of unplanned admissions.
- # of medical orders that are entered into KIDCARE by admitting subspecialty teams for a child on the ketogenic diet who had an unplanned admission to ensure that the appropriate level of monitoring was done and that treatment parameters were followed in the event of low blood glucose levels, decrease in level of ketosis, hyperketosis, blood product, or medication administration.
- Non-adherence to the CPG will be identified and a plan to re-educate who require remediation will be put forward.
- Feedback received by end-users as they utilize the CPG. Feedback will be shared among users and will be, if appropriated used in future revisions of the Ketogenic Diet CPG.

7.0 References

8.0 Guideline Group and Reviewers

Guideline Group Membership:

1. Maria Zak, NP-Paediatrics, Division of Neurology
2. Yeoumei Christiana Liu, Clinical Dietician, Ketogenic Diet, Clinical Dietetics
3. Helen Lowe, Clinical Dietician, Ketogenic Diet, Clinical Dietetics
4. Valerie Chan RN, Neurology Clinic
5. Janice Mulligan, Social Work, Neurology
6. Elizabeth Donner, MD, Director of Comprehensive Epilepsy Program, Staff Neurologist
7. Jeff Kobayashi, MD Director of Ketogenic Diet Program, Staff Neurologist

Internal Reviewers:

1. Dr. Elizabeth Harvey
2. Dr. Michael Weinstein
3. Diana Kalnins, RD
4. Dr. Anne-Marie Guerguerian
5. Dr. Moharir and John Oldenhof (parent).

Appendix: Grades of Recommendation

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Level of Evidence</th>
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</thead>
<tbody>
<tr>
<td>Section 1</td>
<td>C (Recommendation supported by expert opinion or experience of a consensus panel.)</td>
</tr>
<tr>
<td>Section 1.1 Table Indications</td>
<td>B (Recommendation supported by at least one cohort comparison, case study or other experimental study)</td>
</tr>
<tr>
<td>Section 1.1 Table Contraindications</td>
<td>C</td>
</tr>
<tr>
<td>Section 2</td>
<td>C</td>
</tr>
<tr>
<td>Section 3.1, 3.2, 3.3, 3.4</td>
<td>C</td>
</tr>
<tr>
<td>Section 3.5</td>
<td>B</td>
</tr>
<tr>
<td>Section 4</td>
<td>C</td>
</tr>
</tbody>
</table>

Attachments:

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Ketogenic Diet

Pediatric Nutrition Care Manual®, Academic of Nutrition and Dietetics (American Dietetic Association).doc
Keto Diet Funding Opportunities.docx
nurse physician information.doc
SSC Poster w Colour FINAL jmREV.pdf
SickKids Ketogenic Diet Guidelines 2014.doc
final Info session handout June 2010.doc
Table 1. Determining which Ketogenic Diet to initiate with patients.pptx
SK Letter Head Classic Discharge Letter.doc
Printable Keto Diet Indications and Contraindications.pptx
Knowledge pathway, nervous system paediatric society.pdf
Pre diet Information Package updated 2014.docx
Ketogenic Diet Dietary Reference Intakes and Calculations.docx
Figure 1. Determining eligibility of a patient for the Ketogenic Diet (KD).pptx
pre diet questionairre.doc
Sickkids clinical guideline Overview of Ketogenic Diet Types.docx