

Ketogenic Diet Initiation Care Guideline

Inclusion Criteria: a patient who is deemed a candidate by a child neurologist and the multidisciplinary team in the Ketogenic Diet Clinic and who have met the following:

- Failed 2 or more appropriately chosen antiepileptic medications
- Compliant with antiepileptic drug regimen
- Completed screening labs: serum amino acids, urine organic acids, acylcarnitine profile, lactate, pyruvate, and ammonia
- Parental consent and interest/motivation

Exclusion Criteria: malnourished, non-compliance with antiepileptic drug regimen, defect in fatty acid oxidation

Assessment

- Vital signs q 4 hours until tolerating diet (without emesis or hypoglycemia), then q shift
- Daily weights

Interventions

- Seizure precautions
- Continue prescribed antiepileptic drugs
- Pharmacy to change all medications to lowest CHO form.
- Lab: CMP + Phos on admit, BMP + CA ⁺⁺ daily, serum Ketones (KET) daily beginning on day 2
- Accuchecks q 2 hrs if < 1 yr, after 24 hours q 4 hrs if no hypoglycemia
- Accuchecks q 4 hrs if > 1 yr
- If blood glucose < 40 mg/dL or patient symptomatic, give 15 mL juice and recheck in 30 minutes (repeat as necessary until > 50 mg/dL). If NPO, give 0.25 gm/kg D10W. Notify provider
- If intractable hypoglycemia (3 episodes of BG < 40 mg/dL within 24 hrs), consider D2.5W-D5W continuous infusion to maintain blood glucose 50-80mg/dL
- Monitor for acidosis; treat if symptomatic and/or CO₂ < 20 mmol/L X2 with oral sodium bicarbonate, 1 meq/kg BID
- Urine ketones, specific gravity, and pH q void; if specific gravity > 1.030 consider IV fluid bolus (no dextrose)
- Ketogenic diet – PO (Modified diet) or Enteral(ketogenic recipe) (see p. 2)
- Fluids – maintenance divided throughout the day (caffeine & calorie free). If NPO, provide maintenance IVF (no dextrose)
- Consults: Nutrition, Social Service, and Child Life; Psychology PRN

Goals

- Urine ketones: 80-160 mg/dL (mod to large)
- Ketones (Beta-Hydroxybutyrate): 40 -80 100 mg/dL (4-8 mmol/L)
- Urine specific gravity (USG): 1.010-1.020-5
- Blood glucose (BG) (non-fasting): 50-80 mg/dL
- Urine pH: 6-8
- CO₂ > 20 mmol/L

Discharge Criteria

- Consumed and tolerated 3 full strength keto meals or feedings at goal ratio
- Normoglycemic (> 50 mg/dL) for previous 12 hrs
- Ketones in urine are moderate to large
- Parents have all necessary supplies (gram scale, formula, medications, urine dipsticks)
- Parental education complete; successful return demonstration
- Order ketostix, measure urine ketones BID.
- Order all medications through CHOC OP pharmacy;; CHOC outpatient pharmacy (pharmacy to ensure keto friendly brands)
- Order NanoVM &/or Nature's Bounty calcium plus D3 (see RD note)

Recommendations/ Considerations/Information

- The ketogenic diet is a high fat, low carbohydrate diet that has been employed as a treatment for medically refractory epilepsy since the 1920's
- The ketogenic diet reduces seizures in up to two-thirds of children refractory to anticonvulsant drugs
- The diet mimics the biochemical changes associated with starvation and induces, among other changes, production of ketone bodies (mainly beta hydroxybutyrate, and to lesser extent, acetoacetate and acetone), which has been implicated in the mechanisms of seizure control
- The ketogenic diet is strictly calculated requiring family to weigh all food consumed. The family and social structure of the patient is critical to its success. If the family cannot help maintain complete compliance, ketosis cannot be achieved
- Patients are scheduled for a 4 – 5 day admission for ketogenic diet initiation

Patient/Family Education

Education by RN

Day 1

- Urine ketone testing
- Urine specific gravity testing

Education by RD

Day 1

- Ketogenic Diet: Parents' Guide
- Meal plan, vitamins and minerals
- Fluids

Day 2

- Ketogenic food prep
- Reading labels

Day 3

- Monitoring and sick day
- Refer to CharlieFoundation.org

Ketogenic Diet Initiation Care Guideline

Ketogenic Oral (PO) Diet

Day 1
Breakfast

- No concentrated sweets (NCS) diet at home + 240 mL of fluid

Remaining Meals

- Provide ketogenic meals at 1:1 ratio, goal calories

Day 2

- Ketogenic meals 2:1 ratio, goal calories

Day 3

- Ketogenic meals 3:1 ratio, goal calories

Day 4

- Advance to ketogenic 4:1 ratio or goal ratio, at goal calories

Ketogenic Enteral Diet

Day 1
Breakfast

- Usual formula feeding at home

Remaining meals:

- Full strength ketogenic formula at 1:1 ratio

Day 2

- Full strength ketogenic formula at 2:1 ratio

Day 3

- Full strength Ketogenic formula at 3:1 ratio

Day 4

- Advance to full strength ketogenic 4:1 ratio or goal ratio

References

Ketogenic Diet Initiation Care Guideline

- Bergqvist C, Schall JI, Gallagher PR, et al. Fasting verses Gradual Initiation of the Ketogenic Diet: Prospective, Randomized Clinical Trail of Efficacy. *Epilepsia*. 205; 46(11): 1810-1819
- Bough KJ, Rho JM. Anticonvulsant Mechanisms of the Ketogenic Diet. *Epilepsia* 2007; 48(1): 43-58.
<http://www3.interscience.wiley.com/journal/117957479/abstract>
- Hartman AL, Vining EPG. Clinical Aspects of the Ketogenic Diet. *Epilepsia* 2007; 48(1): 31-42.
<http://www3.interscience.wiley.com/journal/117957476/abstract>
- Freeman JM, Kossoff EH, Hartman AL. The Ketogenic Diet: One Decade Later. *Pediatrics* 2007; 119(3): 535-543. <http://pediatrics.aappublications.org/cgi/content/abstract/119/3/535>
- Henderson CB, Filloux FM, et al. Efficacy of the Ketogenic Diet as a Treatment Option for Epilepsy: Meta-analysis. *Journal of Child Neurology* 2006; 21: 193-198.
<http://jcn.sagepub.com/cgi/content/abstract/21/3/193>
- Kang HC, Chung DE, et al. Early and Late-onset Complications of the Ketogenic Diet for Intractable Epilepsy. *Epilepsia* 2004; 45(9): 1116-1123.
<http://www3.interscience.wiley.com/journal/118751048/abstract>
- Kossoff EH. International Consensus Stament on Clinical Implementation of the Ketogenic Diet: Agreement, Flexibility, and Controversy. *Epilepsia* 2008; 49(Suppl. 8): 11-13.
<http://onlinelibrary.wiley.com/doi/10.1111/j.1528-1167.2008.01823.x/abstract>
- Kossoff EH, Zupec-Kania BA, et al. Optimal Clinical Management of Children Receiving the Ketogenic Diet: Recommendations of the International Ketogenic Diet Study Group. *Epilepsia* 2009; 50(2): 304-317. <http://www3.interscience.wiley.com/journal/121419559/abstract>
- Kossoff EH, Zupec-Kania BA, et al. Ketogenic Diets: An Update for Child Neurologists. *Journal of Child Neurology* 2009; 24 (8): 979-88.
<http://jcn.sagepub.com/content/early/2009/06/17/0883073809337162.abstract>
- Kossoff EH, Laux LC, et al. When do Seizures Usually Improve with the Ketogenic Diet? *Epilepsia* 2008; 49(2): 329-333. <http://www3.interscience.wiley.com/journal/119390118/abstract>
- Lefevre F, Aronson N. Ketogenic Diet for the Treatment of Refractory Epilepsy in Children: A Systematic Review of Efficacy. *Pediatrics* 2000; 105(4): 1-7.
<http://pediatrics.aappublications.org/cgi/content/abstract/105/4/e46>
- Mady MA, Lossoff EH, et al. The Ketogenic Diet: Adolescents Can Do It, Too. *Epilepsia* 2003; 44(6): 847-851. <http://www3.interscience.wiley.com/journal/118867304/abstract>
- The Charlie Foundation. <http://charliefoundation.org>
- Zupec-Kania B, Zupanc ML. Long-term Management of the Ketogenic Diet: Seizure Monitoring, Nutrition, and Supplementation. *Epilepsia* 2008; 49(Suppl. 8): 23-26.
<http://onlinelibrary.wiley.com/doi/10.1111/j.1528-1167.2008.01827.x/abstract>