

Disclaimer: This clinical pathway is provided as a general guideline for use by Licensed Independent Provider's (LIP) in planning care and treatment of patients. It is not intended to be and does not establish a standard of care. Each patient's care is individualized according to specific needs.

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

A patient who is malnourished, non-compliance with antiepileptic drug regimen, defect in fatty acid oxidation

Assessment

- Vital signs per unit standard of care
- Weights on Monday and Thursday

Interventions

- 24 hour VTM upon admission for baseline seizure quantification
- Seizure precautions
- Continue prescribed antiepileptic drugs
- Pharmacy to change all medications to lowest CHO form.
- Lab: CMP + Phos on admit, BMP + CA ++ daily, beta-hydroxybutyrate (B-OH) daily beginning on day 2
- Accuchecks q 4 hrs (starting on Day 2)
- 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
- Ensure the patient has a documented allergy of "Sugars, Metabolically Active"
- 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 mg/dL (4-8 mmol/L)
- Urine specific gravity: 1.010-1.020
- Blood glucose: 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 hours
- 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 pharmacy; Ensure keto-friendly dosage forms (eg. Tablet/capsule or carb-free liquid)
- Order vitamin/mineral supplements

Recommendations and Considerations

- 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 and Family Education

Education by RN

- Urine ketone testing
- Urine specific gravity testing

Education by RD

- Ketogenic Diet: Parents' Guide
- Meal plan, vitamins and minerals
- Fluids
- Ketogenic food prep
- Reading labels
- Monitoring and sick day
- Review common drug information

Ketogenic Diet Initiation Care Guideline

Ketogenic Oral (PO) Diet

Ketogenic Enteral 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 1

Breakfast

- Usual formula feeding at home

Remaining meals:

- Full strength ketogenic formula a 1:1 ratio

Day 2

- Ketogenic meals 2:1 ratio, goal calories

Day 2

- Full strength ketogenic formula at 2:1 ratio

Day 3

- Ketogenic meals 3:1 ratio, goal calories

Day 3

- Full strength ketogenic formula at 3:1 ratio

Day 4

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

Day 4

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