



Special Circumstances on the Ketogenic Diet

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About our Keto Ambassadors



Lindsey Thompson, MS RD CSP LD BASED IN KANSAS CITY, MO

Lindsey Thompson is a registered dietitian in the Comprehensive Epilepsy Center at Children's Mercy Hospital in Kansas City, Missouri, and is currently in pursuit of her PhD in Nutrition from the University of Kansas Medical Center. Lindsey is passionate about the ketogenic diet because of the remarkable difference it makes in the lives of patients, families and caregivers affected by epilepsy and other neurological disorders. Many of her patients seek the diet as a "last resort" and it brings Lindsey great pleasure to deliver optimal outcomes for a family in need. While the concepts

of the ketogenic diet date back to the early 20th century, Lindsey believes we are just now paving the way for modern medicine to seek and employ diet and nutrition as a first-line treatment for both chronic and debilitating disease states.



Zahava Turner, RD CSP LDN BASED IN BALTIMORE, MD

Zahava Turner is a Senior Clinical Pediatric Dietitian, Board Certified in pediatric nutrition working at the Johns Hopkins Hospital in Baltimore, Maryland since 2005. She received her BS and completed her dietetic internship at Queens College CUNY in New York with an emphasis in clinical nutrition and worked for 2 years as a pediatric dietitian at Schneider's Children's Hospital. Zahava specializes in using the Ketogenic diet for infants and children with epilepsy and has spoken both nationally and internationally at several conferences on the ketogenic and Modified Atkins diet

for epilepsy. She is a co-author of 18 publications and the widely-referenced book Ketogenic Diets 5th edition. Zahava lives in Baltimore with her husband and three children.



STATUS EPILEPTICUS

Disclosures



Consultant - Nutricia North America as Keto Ambassador

Objectives



- Define status epilepticus
- Identify ways to initiate the ketogenic diet for a patient in status epilepticus

Status Epilepticus



- Ongoing convulsive or non convulsive seizures following administration of benzodiazepine and non-benzodiazepine antiepileptic medicine
- Affects 5-41/100,000 annually
 - 38% mortality rate and over 60% occurring after prolonged seizure duration
- Refractory status occurs in 30% of patients with status epilepticus- to first and second line of treatment
- Causes: low anti seizure drug levels, drug withdrawal, stroke,
- Other than medications – VNS, surgery, transcranial magnetic stimulation and the ketogenic diet

FIRES



- Febrile Infection Related Epilepsy Syndrome
- Rare epileptic syndrome
 - Previously healthy individual during or close to febrile episode
- Seizures rapidly increase and result in status epilepticus
- Emergent placement on the diet
 - Patient is usually intubated, sedated in ICU
- 4:1 ratio used to induce ketosis quickly
- 7-10 day usual response time

Ketogenic Diet in Status



- Multiple case reports on using the ketogenic diet for refractory status
- Kossoff et al: 2013 - combines literature up till 2008 with 32 children and adults placed on the diet with 25 (78%) seizure free
 - Found similarities on those that responded well to dietary treatment
- Encephalitis and Febrile Infection Related Epilepsy Syndrome (FIRES) noted to respond well
 - 13/32 (41%) had FIRES

Table 1. Studies Examining the Ketogenic Diet for Status Epilepticus.

Author (year)	Age range (years)	Number of subjects	Diet tried	Seizure-free rate (%)	Time to response	Etiology
Bodenant (2008) ⁵	54	1	Ketogenic	1 (100%)	7 days	Partial
Villeneuve (2009) ⁶	1-10	5	Ketogenic	4 (80%)	1-10 days	Cryptogenic (2), Sturge-Weber, encephalitis, hypomelanosis of Ito
Kumada (2009) ⁷	5	2	Modified Atkins	2 (100%)	5-10 days	Frontal lobe epilepsy, heterotopias
Wusthoff (2010) ⁸	29-34	2	Ketogenic	2 (100%)	4-8 days	Rasmussen syndrome, head trauma
Nabbout (2010) ⁹	5-8	9	Ketogenic	7 (78%)	4-6 days	Febrile-illness related epilepsy syndrome
Ismail (2011) ¹⁰	14	1	Ketogenic	1 (100%)	10 days	Febrile-illness related epilepsy syndrome
Cervenka (2011) ¹¹	49	1	Ketogenic then modified Atkins	1 (100%)	7 days	Idiopathic, possibly autoimmune
Nam (2011) ¹²	4-40	5	Ketogenic	2 (40%)	7-19 days	Viral encephalitis
Vaccarezza (2012) ¹³	1-14	5	Ketogenic	4 (80%)	2-3 days	Febrile-illness related epilepsy syndrome, partial
Martikainen (2012) ¹⁴	26	1	Low-glycemic index treatment	1 (100%)	4 days	Mitochondrial polymerase gamma
Total	1-54	32	Various	25 (78%)	1-19 days	Various

- 9 children from Paris and Argentina
 - 5-8 years old
- Failed 3-6 anti convulsants – up to 55 days in status
- Ketogenic diet started via NG tube
 - Ketosis achieved within 2.8 days
 - Seizure control 4-6 days in 7/9 children

Case Study - PW



- 14 year old female
 - No significant past medical history
 - Outside hospital for 4 months presented with seizures
 - Admitted to Johns Hopkins Hospital with altered mental status, status epilepticus and fever
 - Started ketogenic diet 6 days after admission

Case Study



Admit

- Wt: 71.9kg (94th%ile) z score: + 1.59
 - Ht: NA (in PICU)
- Diet Rx: Jevity[®] 1 Cal @ 50mL/hr = 18 kcals/kg
 - Estimated needs using WHO = 18-20kcals/kg

Initiation



- Stop previous formula and run normal saline at maintenance for 12- 24 hours
- Start ketogenic diet at half strength rate for 12 hours and advance to goal rate
- For PW: Changed formula to KetoCal[®] 4:1 1200 calories with a goal of 50mL/hr = 18 kcals/kg and 0.34g pro/kg

Case Study



- 1 week on diet
 - PW waking up, more movement
- Albumin trending down - 3.0
 - Increased calories, added Beneprotein® and reduced the ratio to 3:1 to provide more calories & protein
 - Diet Rx: 1325 kcals 3:1@ 55mL/hr
- Transferred out of the ICU to Adolescent unit
 - Wt decreased to 66.6kg (84th%ile)
 - Awake, alert, moving around more
 - Increased to 1600 kcals, 3:1 ratio
 - Overnight @ 85mL/hr x 8 hours
 - 3 bolus of 220mL + 125mL water

Case Study



- Transferred to local rehab facility
 - Increased mobility
 - Increased calories to 1800 kcals, 3:1 ratio
 - Transitioned to 3 soft meals and 3 snacks
- 4 months on diet: Ratio decreased to 2:1
- After 6 months on diet, PW found 2:1 ketogenic diet to be very difficult
 - Stopped the diet with no wean
 - Continues to be seizure free

Practical Issues



- Requires established ketogenic diet team
 - Neurologist, Ketogenic Dietitian
 - To assist and follow with acute issues i.e hypoglycemia, ketosis
 - ICU team needs to be on board
 - Need team to be able to follow once discharged
- Have found it very difficult to maintain the ketogenic diet once patient is awake and alert

Future Research



Prospective studies

- More patients
 - To help define what etiologies respond better
- Help to determine best way to administer the diet
 - Fasting vs non fasting
 - Ratio?
 - Time frame to give diet a chance?
 - How long should patient stay on the diet once it worked?



KETOGENIC DIET IN THE NICU (NEONATAL ICU)

Disclosures



Consultant - Nutricia North America as Keto Ambassador

Objectives



- Review the ketogenic diet induction process in the NICU
- Recite potential complications and troubleshooting related to diet initiation in the NICU

Ketogenic Diet Baseline Screening



- Labs

- Urine organic acids*
- Serum amino acids*
- Acetylcarnitine*
- Ammonia*
- Lactic acid*
- Complete blood count with differential
- Basal metabolic panel
- Hepatic function test
- Magnesium
- Zinc
- Selenium
- Lipid profile
- Beta-hydroxybutyrate
- Vitamin D
- PTH

- EKG

- Nutrition

- Ability to safely eat/swallow

Ketogenic Diet Induction



- Slow induction into ketosis over a number of days
 - Typical progression: 1:1 → 2:1 → 3:1 → 4:1
- Maintain baseline feeding regimen in terms of calories, protein, rates
- Formula selection
 - Ketocal[®] 4:1 LQ
 - RCF[®]

Ketogenic Diet Induction (cont)



- Modulars
 - MCT Oil
 - Liquid protein fortifier
 - Infant formula/breast milk
- Monitor for tolerability and side effects
- Parent Education

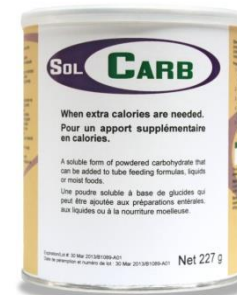
Ketogenic Diet



grams of fat

→ **4:1** ←

grams of
carbohydrate +
protein



Ketogenic Diet- Medications



- All medications changed to tablets (or lowest carbohydrate form)
 - Any suspension or syrup likely contains carb
 - Looking for "carbohydrate-free" not just "sugar-free"
- No IV solutions with dextrose added
 - If IV fluid are needed use Normal Saline

Micronutrient Supplementation



Individualized based on age, formula intake, nutrient needs

- Ketocal[®] 4:1 LQ
 - 0.5 mL Poly-Vi-Sol[®] (contains 460 mg carbohydrate)
 - Iodine – typically needs to be supplemented
 - Monitor potassium levels as Ketocal[®] provides more than the AI
- RCF[®]
- Phlexyvits or FruitiVits[™] (powdered)

NOTE: KetoCal[®] is for over one year of age and used at HCP's discretion.

Normal Labs for the KD Patient in the NICU



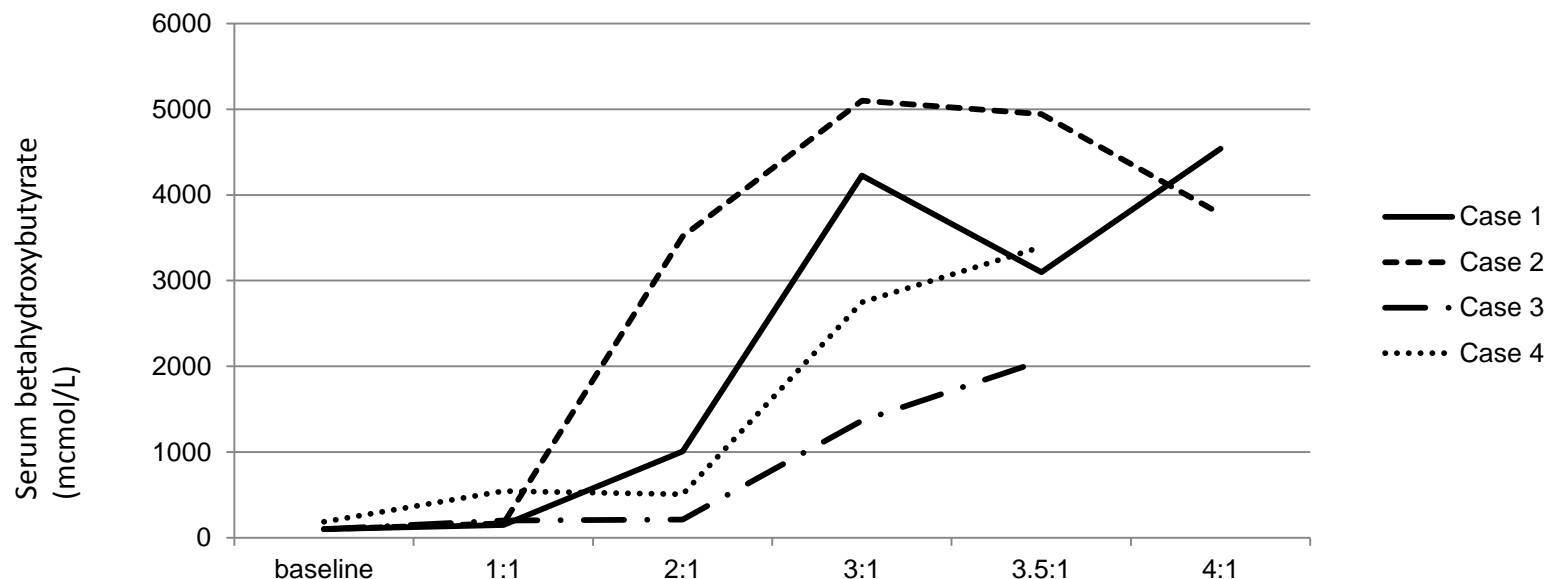
- Blood glucose: 40-100 mg/dL
 - Check q 6 hours upon diet initiation, then q 12-24 hrs
 - If <40 mg/dL, treat with dextrose bolus or apple juice (2 mL/kg)
 - NOTE: 10 mL D10 and 10 mL apple juice contain 1 gram carbohydrate
- CO₂: >15 mmol/L
 - Checked QOD during initiation
 - If below 15 mmol/L, treat with IV Bicarbonate

NICU Labs (cont)



- β -OHB : >2,000 $\mu\text{mol/L}$ (>2 mmol/L)
 - Checked QOD during initiation
 - Satisfactory level very individualized based on seizure control and other parameters
- Urinary ketones: moderate-large
 - Checked q void during initiation
 - NOTE: Urine ketones are very weak in infants and may not be as accurate as serum ketones

Serum and urinary ketone levels in infants initiating the ketogenic diet in the NICU



	Baseline	1:1	2:1	3:1	3.5:1	4:1
Case 1	Negative	Negative	Negative	Negative	Trace	Moderate
Case 2	Negative	Negative	Small	Small	Small	Moderate
Case 3	Negative	Negative	Negative	Trace	Moderate	
Case 4	Negative	Negative	Negative	Trace	Small	

Urinary
ketones

Constipation



- Glycerin suppositories
- MCT Oil
 - Added to formula by RD
 - Start at 5% of kcals, can increase up to 40% of kcals as tolerated
- Miralax® – work with your physician to determine the appropriate amount for the child's age and weight

Special Circumstances



- Surgery/Procedures that require NPO status
 - Provide non-dextrose containing fluids (normal saline or ½ normal saline)
 - Check blood sugars frequently q 2-6 hours

Potential Complications

Acute – During Initiation

- Hypoglycemia (glucose <40 mg/dl)
- Acidosis (CO_2 <15)
- Weight loss
- Lethargy
- Irritability – “waking up” / change in sensory experience
- Constipation

Long Term

- Osteopenia
- Renal stones

Thickeners



Be aware! Most thickeners are cornstarch based!

- Simply Thick[®] – can be used with the ketogenic diet
 - FDA warning re: Simply Thick[®]
 - Not to be used in patients <1 year of age or with history of necrotizing enterocolitis (NEC)
- May have to consider non-PO feeding for patients requiring thickeners

Frequency of Follow Up Evaluations



- Infants < 1 year of age
 - Monthly
 - Can alternate seeing epilepsy provider
- Children > 1 year of age
 - 1 month after diet initiation
 - Every 3 months thereafter
 - Can consider every 6 months for children who have been on the diet long-term

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Thank you!