Navigating Through Obstacles with the Medical Ketogenic Diet for Epilepsy

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Disclosures

- Consultant & Keto Ambassador – Nutricia North America
- Advisory Board – Trumacro™

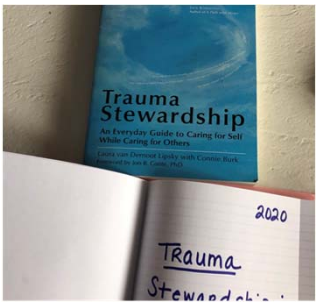
Objectives

- Identify common and rare side effects of the medical ketogenic diet
- Assess various ways to resolve side effects of the medical ketogenic diet
- Determine ways to proactively avoid harmful side effects for patients on medical ketogenic diet

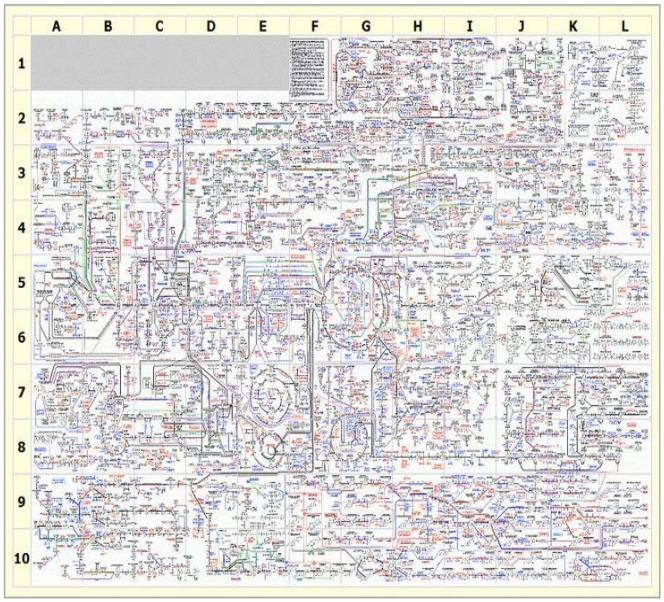
Agenda

1. Common obstacles
2. Resolving & preventing side effects
3. Story - Danny
4. Weaning the diet – when & how

What has Robyn been up to?



The Metabolic Pathways in Humans

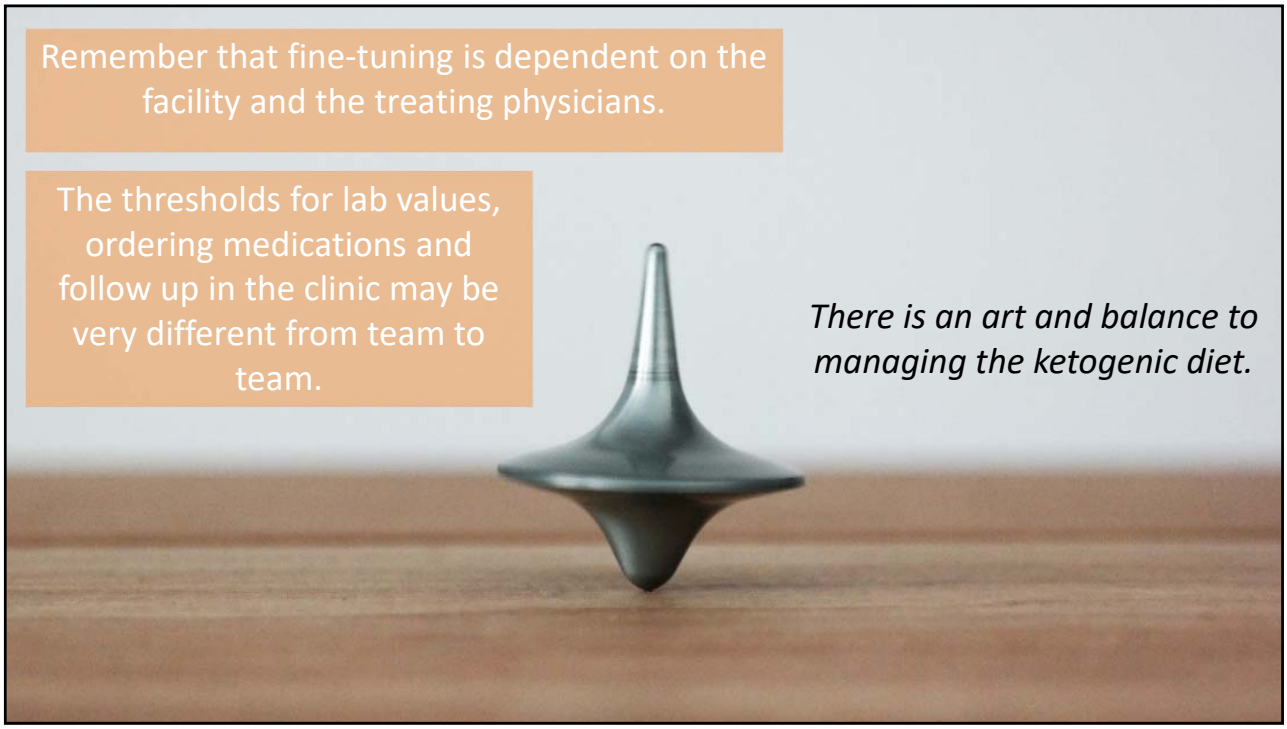


Multiple mechanisms for **reducing seizures**

Multiple mechanisms for **side effects**

Biochemical Pathways,
G. Michal, editor,
Third Ed, Part 1
Roche Applied Science

Poster now available digitally: http://biochemical-pathways.com/#/map/1



The consensus paper 2018 tells us …

Should be considered:

- Angelman syndrome
- Complex 1 mitochondrial disorders
- Dravet syndrome
- Epilepsy with myoclonic–atonic seizures (Doose syndrome)
- Glucose transporter protein 1 deficiency syndrome (GLUT-1)
- Febrile infection–related epilepsy syndrome (FIRES)
- Formula-fed (solely) children or infants
- Infantile spasms
- Ohtahara syndrome
- Pyruvate dehydrogenase deficiency
- Super-refractory status epilepticus
- Tuberous sclerosis complex

Could be considered:

- Adenylosuccinate lyase deficiency
- CDKL5 encephalopathy
- Childhood absence epilepsy
- Cortical malformations
- Epilepsy of infancy with migrating focal seizures
- Epileptic encephalopathy with continuous spike-and-wave during sleep
- Glycogenosis type V
- Juvenile myoclonic epilepsy
- Lafora body disease
- Landau-Kleffner syndrome
- Lennox-Gastaut syndrome
- Phosphofructokinase deficiency
- Rett syndrome
- Subacute sclerosing panencephalitis

Kossoff EH, Zupec-Kania BA, et al. Optimal clinical management of children receiving dietary therapies for epilepsy: Updated recommendations of the International Ketogenic Diet Study Group. *Epilepsia Open*, 3(2): 175-192, 2018.

Common obstacle: explaining and choosing which diet

Source: Ketogenic Therapy for Neurological Disorders – Charlie Foundation

Age Group	Classic (4:1, 3:1) or Modified (2:1 or 1:1) KD	MCT Oil Diet	LGIT	MAD
Infant	X	X	Not recommended	Not recommended
Toddler	X	X	Not recommended	Not recommended
Child	X	X	X	X
Adult	X - Modified	X	X	X

Table based on table 3.7 from pocket guide

All proceeds benefit the Charlie Foundation

Understanding the options

Dietary Element	LGIT	MAD	MKD	CKD
Carbohydrate (excluding fiber)	10% of kcals GI < 50	10-20 g/day	15-30 g/day or 5% of kcals	4% of kcals
Fat	60% of kcals	Ad lib (LCT)	60-80% of kcals	90% of kcals
Protein	30% of kcals	Ad lib	Ad lib	6% of kcals
Food Measurements	Weighed; Household	Visual	Weighed; Household	Weighed
Ketone Testing	Urinary	Urinary	Urinary and blood	Blood
Prescribed ketogenic nutritional products	No	Initiation Only (LCT)	Yes (LCT/MCT)	Yes (LCT)

Based on Table 1 from article

Martin-McGill KJ, et al. Understanding the core principles of a 'modified ketogenic diet': a UK and Ireland perspective. J Hum Nutr Diet. 2019;32:385-90.

Common obstacle: ketone fluctuation

Dietary Indiscretion	Teachable moments
Medication or Supplement change	Change back if possible or help to find a better option
Level of hydration	Regulate fluid intake over the course of the day
Illness and outside stressors	Maybe do nothing, if seizures are well controlled or illness/stress is short term

Common obstacle: constipation

Manage with fiber containing foods, extra water, coconut oil, MCT oil, group A vegetables, avocado

Other options

- Laxatives & stool softeners
  - MiraLax®, Senna, suppository
- Smooth Move® Tea
- Heather's Tummy Care™ Organic Fennel Tummy Tea
- Simethicone / Mylicon® drops
- Chia Seeds

Common obstacle: nausea, vomiting, reflux

Possibly from:

- hypoglycemia
- excess ketosis
- metabolic acidosis

Corrected with 15-30 mL juice

May need a longer term supplement of potassium citrate or bicarbonate

Common obstacle: NPO

Inpatient:
- Normal Saline IV
- Clears by PO/NG
- KetoCal® by GT
- May lead to TPN

Outpatient:
- Watch for low urine output
- Illness vs Strike

Either:
- ½ Pedialyte® + ½ Water
- Stay hydrated, carb free beverages
- Oral Electrolyte Solution

(3/4 tsp Morton® Lite Salt + 1/5 tsp Baking Soda + 1 liter Water)

Common obstacles: growth

Hormone changes associated with KD can affect growth over time

Increasing protein rarely helps a keto kid grow in height/length

Make adjustments in kcals as needed for BMI goals

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Navigating a common obstacle

Danny - 5 year old boy

Note: this patient is not actually "Danny"

Diagnosis

History of lissencephaly-pachygyria associated with a pathogenic PAFAH1B1 mutation

(global developmental delays, subsequent epilepsy with infantile spasms and focal seizures with onset at 5 months of age)

- Spasms were initially controlled with vigabatrin
- Age 4 started having seizures again and needing escalating doses of medications

Navigating a common obstacle

Danny - 5 year old boy

Social

Lives at home with his parents and sisters (11 and 15yo)

Parents trade off work shifts to be home to care for him

Mother: Spanish speaking
Father: bilingual

Navigating a common obstacle

Danny - 5 year old boy

Nutrition

- PO ad lib 3 meals per day, fed by parent on spoon
- Not a very good drinker, but mixing fluid with food
- BMI= 12.42 (z-score: -3.91)

Speech

- Consumes Level 3 (honey thick liquids) and purees
- Mom typically blends all meals for him
- Typically takes Daniel 30-60 minutes to consume a meal

Navigating a common obstacle

Danny - 5 year old boy

Keto diet initiation

- Tolerating meals very well, low fluid intake
- Passed swallow study, but needs thickened liquids or NGT
- Guar gum mixed with water was started

Food Daniel loved before keto

Navigating a common obstacle

Guar Gum + Water

Navigating a common obstacle

Danny - 5 year old boy

4 month follow-up

- Tolerating 3 meals by mouth very well
- All 32 ounces (960 mL) of water by GT (placed 2 months after KD start)
- Urine ketones – small/moderate
- Seizures – much improved
- Cognition – improved
- Continue to fine-tune: ratio, calories, protein, hydration

Common obstacle: Seizure Control

- Breakthrough seizures = seizures that happen after a period of seizure free time
- We may not make *any* adjustments due to a breakthrough seizure.

Change in diet if:

Seizures continue and ketones are not at maximum levels

No Change in diet if:

Illness or some factor can be blamed (not finishing fat portion of meal, etc.)

WHEN to Wean

Reasons to discontinue the KD:

- seizure freedom (23%)
- prolonged KD use w/ limited continued benefit (23%)
- no efficacy (20%)

Slower wean	Had achieved significant seizure reduction (>50%)	Longer duration of KD	Were having fewer monthly seizures
Faster wean	Completely unsuccessful sz control	Shorter duration of KD	Were having same/more seizures

Worden LT, et al. *Epilepsy Research.* 2011;Apr 30.

HOW to Wean

Most patients whose seizures worsened during weaning were able to regain some degree of control when increased again.

Consider stopping citrate supplement on the day the diet ends, but continue MVI

Diet effective	Consider weaning by 0.25-0.5 in ratio every 2-4 weeks, pending patient tolerance
Diet ineffective	Consider faster wean (off diet within a few days)

Worden LT, et al. *Epilepsy Research.* 2011;Apr 30.

Diet responses to when medications are weaned

Weaning ACTH/steroid treatment	• Expect appetite, glucose, ketones and heart rate to return to baseline • Watch for weight loss or weight stabilization
Weaning carbonic anhydrase inhibitors (Topamax®, Zonegran®, Diamox®)	• Expect ↑appetite, ketone change, normal C02 • Watch for weight and ketone changes
Weaning Depakote®	• Expect ↓appetite, normal carnitine levels • Watch for weight changes and ↓need for Carnitor®

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- Worden LT, et al. Is there an ideal way to discontinue the ketogenic diet? Epilepsy Res. 2011;95:232-236.

Questions and Discussion

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