

Ketogenic Diet

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Ketogenic diet (KD)

- High fat
 - Low carbohydrate
 - Calories control
 - Adequate protein
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- Therapeutic diet for epilepsy
 - As effective as an AED or VNS

International guideline

Epilepsia, 50(2):304-317, 2009
doi: 10.1111/j.1528-1167.2008.01765.x

SPECIAL REPORT

Optimal clinical management of children receiving the ketogenic diet: Recommendations of the International Ketogenic Diet Study Group

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- International Ketogenic Diet Study Group
- 26 ped epileptologists & dietitian (9 countries)
- standardized protocol

Practical approach*

- Case selection
- Pre-KD assessment
- Ketosis induction
- Evaluation
- Maintenance
- KD discontinuation

Indications

- Intractable epilepsy (any age, Sz type)

Specific for

- Glucose transporter 1 (GLUT1) deficiency
- Pyruvate dehydrogenase deficiency
 - Essential energy for brain
 - Treat
 - seizures
 - cognitive function

GLUT1 deficiency

- GLUT1 protein
- transfers glucose from blood to CSF
- Low CSF glucose, normal plasma glucose
- No other cause (CNS infection/ SAH)
- Intractable Sz, MR, movement disorder
- Ketone → main energy source

PDHD deficiency

- Mitochondrial dysfunction
- Lactic acidosis
- “Pyruvate-to-Acetyl CoA” defect
- Intractable Sz
- Ketone → bypass to TCA cycle

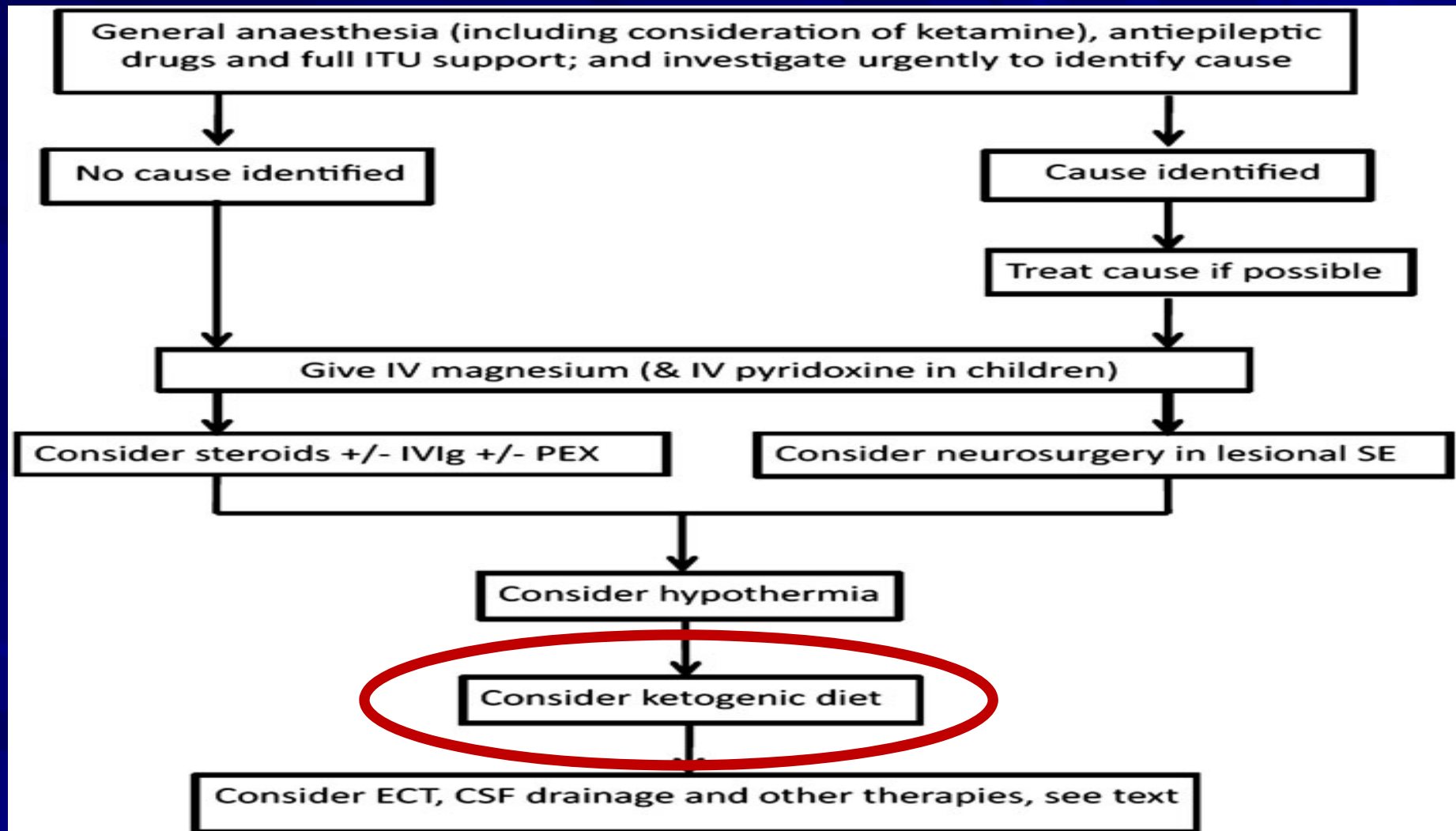
Particular benefit in*

- Tuberous sclerosis complex
- Myoclonic-astatic epilepsy
- Rett syndrome
- Dravet syndrome
- Infantile spasms
- infants or enterally fed patients

Recent indication

- Super refractory status epilepticus
 - status epilepticus
 - continues or recurs
 - despite general anesthesia Rx for 24 h
- Several case series – favorable outcome
- Enteral & parenteral induction

Super refractory SE



S. Shorvon and M. Ferlisi, The treatment of super-refractory status epilepticus: a critical review of available therapies and a clinical treatment protocol. Brain 2011: p1-17

Absolute Contraindication

- Primary carnitine def
- Carnitine palmitoyl transferase (CPT) def
- Carnitine translocase def
- β -oxidation defects
- MCAD/ LCAD/ SCAD
- Long-chain 3-hydroxyacyl-CoA def
- Medium-chain 3-hydroxyacyl-CoA def
- Pyruvate carboxylase def
- Porphyria

**Fatty acid transport
& oxidation defect**

Pre-KD evaluation*

- Counseling
- Sz assessment
- Nutritional evaluation
- Lab evaluation

Available formulas*

- Classical formula (LCT)
- MCT formula
- Modified Atkins
- Low glycemic index (LGI)

Diet route

- Bottle feed / normal food / tube feed

Classical KD

- Widely used
- 4: 1 ratio of fat: protein - carbohydrate
- Main fat source = LCT
- Adequate protein > 1 g/kg
- Low carb - just to prevent hypoglycemia
- Calorie control = 75 - 100% requirement
- Fluid restriction - not necessary

MCT KD

- Increasingly used → better ketosis
- 30%-60% fat: total energy
- More carbohydrate allowance
- Less restrictive, bigger meal
- Similar efficacy to LCT
- MCT can't be cooked → not palatable

Ketosis induction

■ Rapid induction

- fasting (12 h - whenever ketosis)
- admission required
- risk of dehydration, glucose, acidosis
- diet titrating up to the target ratio
- caregiver training during admission

Ketosis induction

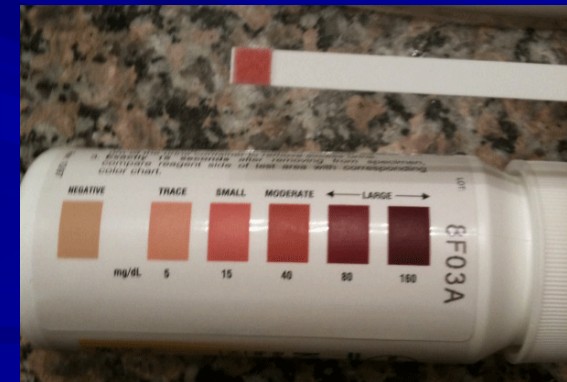
■ Gradual initiation

- without fasting
- admission = optional
- slower but comparable Sz control at 3 m
- lower initial side effect

Bergqvist, A.G., et al., *Fasting versus gradual initiation of the ketogenic diet: a prospective, randomized clinical trial of efficacy*. Epilepsia, 2005. **46**(11): p. 1810-9.

Maintenance phase

- Efficacy evaluation after 3 month
- Neuro
 - seizure control
 - cognitive improvement
- urine ketone - compliance
- serum ketone - Sz control



Maintenance*

- GI & nutritional assessment
- Blood tests
- Supplements
- Oral citrate
- Adverse effects
- Sick rules

Sick rules

ข้อแนะนำเมื่อมีอาการป่วย

1. แจ้งแพทย์ และแสดงบันทึกนี้ทุกครั้ง
2. งดการให้น้ำเชื่อมทุกชนิด
3. หลีกเลี่ยงยาเม็ดและยาฉีดที่มีส่วนผสมของน้ำตาล แอลกอฮอล์ และ แป้ง ในจำนวนสูง
4. หลีกเลี่ยงการให้น้ำเกลือ ถ้าจำเป็นต้องให้ ห้ามให้น้ำเกลือชนิดที่มีน้ำตาลผสมอยู่
5. จำกัดปริมาณน้ำตามที่กำหนดไว้ในแต่ละวัน
6. ถ้ามีการเสียน้ำ เช่น อาเจียน ท้องเสีย เพิ่มปริมาณน้ำได้ชั่วคราวตามเหมาะสม
7. ถ้าป่วยหนัก จำเป็นต้องนอนโรงพยาบาล ควรตรวจน้ำตาลในเลือดตามเหมาะสม

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บันทึกการซัก

ผู้ป่วย ketogenic diet

ภาควิชากุมาร รพ. จุฬาลงกรณ์

ชื่อ

HN

สูตรอาหาร

วันที่เริ่ม

Diagnosis

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side effects

Early

- Dehydration
- N/V, diarrhea
- Hyperlipidaemia
- Hyperuricaemia
- HypoCa, HypoMg
- Metabolic acidosis

Late

- Osteopenia
- Renal stones
- Low carnitine
- Fe def anemia
- Cardiomyopathy(rare)

*** GI & metabolic effect**

***Mostly transient**

Discontinuation

- Diet maintenance - 2 years if effective
- longer as necessary for GLUT-1, PDHD
- Sudden glucose intake / diet cessation → Sz
- Slow weaning over 2-3 months
- overall recurrence risk - 20%
- Higher in TSC, abnormal EEG, MRI

Martinez, C.C., P.L. Pyzik, and E.H. Kossoff, *Discontinuing the ketogenic diet in seizure-free children: recurrence and risk factors*. Epilepsia, 2007. 48(1): p. 187-90.

Draw back*

- Family - Difficult recipe
- Patient - Limited meal

Options

- MAD
- LGIT

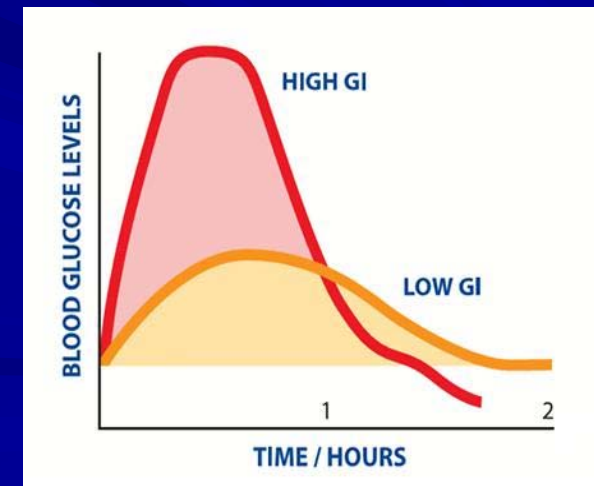
Modified Atkins

- Similar composition to classical KD
- 1: 1 ketogenic ratio
- Restrict carbohydrate (10-20 g/d)
- No limit on protein, fluids, and calories
- Easier meal planning
- Preliminary effective

Kossoff et al. Epilepsy Behav 2007:432–436.

Low GI

- Less fat than KD
 - More carbohydrate 40–60 g/day
 - CHO type → low glycemic index <50
 - e.g.lentils, grapefruit, whole grain bread
-
- Less ketone level than KD
 - Still preliminary effective



Pfeifer and Thiele. Neurology 2005;65:1810–1812.

Conclusion

- proven option for epilepsy
- good efficacy
- need good compliance
- also recommended in SRSE
- Enteral or parenteral route

A large white cruise ship with multiple decks is docked at a wooden pier. The pier is lined with several wooden buildings, likely a restaurant or hotel. The background features steep, green mountains and a snow-capped peak in the distance. The water is calm and reflects the ship and the surrounding landscape. The sky is clear and blue.

Thank you for
your attention