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Denmark

Non-diabetic ketotic hypoglycemia





Literature review provided by professor Henrik Thybo Christesen

Hans Christian Andersen Children's Hospital, Odense, Denmark



Hypoglycemia is seen in 1-10% of newborns in Denmark.

Lower after the introduction of a national prevention program





The birth of a Shaking baby

Noah



Noah

- Born shaking
- 0.8 mmol/l (14 mg/dl)
- An i.v glucose need of 12 mg/kg/min indicate of hyperinsulinism
- Diazoxide for 5 days, good effect
- Spontaneous resolution → discharged
- Nursed night and day until 12 months
- Seizure at 17 months old
- One year later, diagnosed with Idiopathic
 Ketotic Hypoglycemia



A small group of children with neonatal hypoglycemia has prolonged or persistent hypoglycemia as consequence of congenital metabolic diseases or congenital hormone disorder



The birth of a hyper-nursing baby

Savannah



Savannah

- BG dropped to 2.2 mmol/l (39 mg/dL) but stabilized with nursing. Milk was running from day 1.
- Nursed 24/7. Nursed all night, with short breaks in the daytime.

17 month old: BG 3.7 (66 mg/dl), ketones 3.3, admitted.

- Doctors believed it was due to malnutrition
- 2nd opinion from professor Christesen:

Gave her a Dexcom CGM, cut down the nursing at night: **Admitted and diagnosed with severe ketotic hypoglycemia.**





ADHD, bulimia and a bad memory

Danielle





Danielle

ADHD

Extreme bad memory

Bulimia

- Low blood glucose after glucose tolerance test
- · Ice cream while giving birth
- Suspected low blood glucose during both pregnancies
- Diagnosed with non-diabetic ketotic hypoglycemia at 26-year-old
- Dexcom and cornstarch
- "Cured" from ADHD and bulimia

Non-diabetic ketotic hypoglycemia can be seen in children without diabetes because of growth hormone deficiency, adrenocortical deficiency or fatty acid metabolism, and glycogen storage diseases



When these diagnoses are ruled out, non-diabetic ketotic hypoglycemia can be categorised as unexplained or idiopathic (IKH), otherwise known as accelerated starvation.



Genetics and treatment

Genetics

- GSD panel: for all known GSD types normal
- Exome Sequency: No clear genetic cause.

However: A novel disturbance in the metabolism of glycogen is suspected

Treatment

- Octreotide and Diazoxide tried with no success.
- Current treatment: Continuous feeding with maltose infusions in g-tubes during sleep.
- Emergency regime: Cornstarch and juice











Clowns and ambulances



Ambulance trips



Hospital visits

100 - 110

Very strong bond to the hospital clowns

IKH is the most frequent state with low blood sugar in preschool kids. However the incidence is unknown. Most commonly it debuts around **18 months** with spontaneous normalization around 6-7 years of age, but this does not account for all.

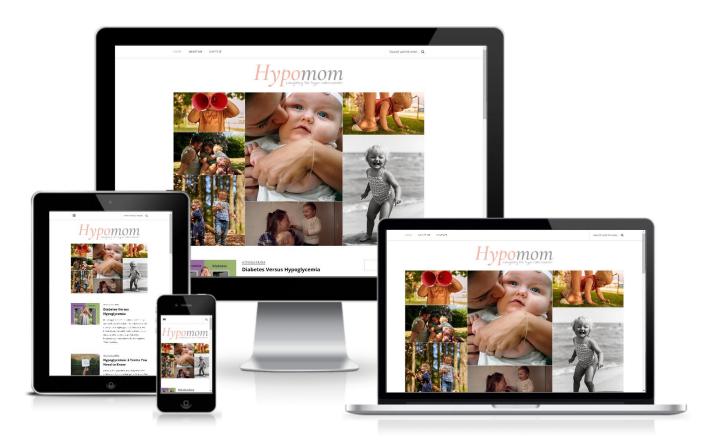


Our development

- **Normal** psychomotor development
- Ketone bodies as **alternative** fuel
- Blood glucose levels not as extremely low as some CHI patients
- EEG normal (Savannah and Danielle)
- Normal daycare with medical support staff









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DIABETES

KETOTIC HYPOGLYCEMIA

CONGENITAL HYPERINSULINISM

Microsoft Word Document

KETOTIC HYPOGLYCEMIA **BLOOD SUGAR GUIDE**

Ketones

3.0 - 10.0 Exfremely high: Call 311, give juice (if he is not responsive use his g-lube), check blood glucose, contact more, be ready for selecte

1.5 - 2.9

0.6 - 1.4

0.0 - 0.5

10.0 and above

4.5 - 9.9

4.0 - 4.4 Olive Nim Rood

3.5 - 3.9

3.0 - 3.4 One juice and comstants through g-tube, cell more, wetch him closely until more gets there, check betones

LO - 2.4
Call 911, call more, be ready for seizure, give juice through grace, check fustones



Symptoms of hypoglycemia

 Poor feeding or vanishing. · Problems keeping the body warm

Parents Info
Mon, Device 50 40 30 08
Ded, Server, 50 49 30 08

Personal info

Nosh is known with idepathic ketotic hypoglycensia:

Nosh reacts to Olivoagon. De siert on severe hypos 30 min after Olivoagon is

Noch has a g-tube



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