Feeding problems in CHI
- mechanisms, facts and treatment -

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Magensonde wegen Diazoxideinnahme

...aber uns kurz in Who is Who vorgestellt). Seitdem ist unsere Leben total verändert, chaotisch und von den Blutzuckerswerten bestimmt. Wir wurden aus dem Krankenhaus in Heidelberg mit einer Magensonde entlassen, wodurch Timmy einen Grossteil seiner Nahrung und das Diazoxid erhalten hat. Das Essen ist um...
Feeding is a scientific "white spot" ....

- Very few scientific articles about "feeding and CHI" in the literature.
- Only short mentioned in reviews, guidelines and recommendations...
- Feeding is the natural and physiologic counterpart to increased insulin secretion...

International, 1999 - 2009

<table>
<thead>
<tr>
<th>Affected functionality</th>
<th>reported</th>
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</thead>
<tbody>
<tr>
<td>Feeding problems</td>
<td>67%</td>
</tr>
<tr>
<td>Dietary treatment</td>
<td>53%</td>
</tr>
<tr>
<td>Ongoing tube feeding</td>
<td>41%</td>
</tr>
<tr>
<td>Developmental delay (surgery)</td>
<td>47%</td>
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<tr>
<td>Brain damage</td>
<td>36%</td>
</tr>
<tr>
<td>Developmental delay (no surgery)</td>
<td>33%</td>
</tr>
<tr>
<td>Ongoing medication</td>
<td>28%</td>
</tr>
<tr>
<td>Speech delay</td>
<td>24%</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>14%</td>
</tr>
</tbody>
</table>

Data from 136 children reported to Congenital Hyperinsulinism International. (M Hopkins, J Raskin)
Food aversion among patients with persistent hyperinsulinemic hypoglycemia of infancy

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Nutrition basics - 01

• Recommendations for food composition in children

  – 50% Carbphydrate
  – 30% Protein
  – 20% Fat
Carbohydrates

- Physiologic targets of insulin action
  → Increase of blood sugar level
- Different time and duration of blood-sugar effects

Glucose

Saccharose (normal sugar)

Starch, glycogen
Who's using carbohydrates...?
Some words about fat and protein…

- **Protein**
  - Necessary for growth, body functions
  - *Induces hypoglycaemia in protein-sensitive HI* (e.g. HIHA-syndrom)

- **Fat**
  - Highest energy content of all types of food
  - Reduces gastric emptying time and thereby slows resorption of carbohydrates

- **Fibers**
  - Nearly not resorbed
  - Delayed resorption of carbohydrates.
Special issues in CHI?

• Feeding is a main pillar of treatment
• Feeding is in the responsibility of the family (nurses)
• Feeding can antagonize insulin action (in theory)
• Intensified feeding is a way to treat CHI...
• BUT....
Feeding problems in CHI…

• Reported in 50 – 60% of patients, in combination with:
  – Overweight
  – Refusal to eat
  – Vomiting
  – ...it's a daily fight about feeding …
Overweight

• To counterbalance the insulin action patients get more food than needed...

⇒ Inappropriate weight gain
  – Delayed motoric development
Feeding problems in CHI - 01

- Most frequent symptom in patients
- Usually "not-eating" is the main problem.
- Sometimes aversion to very specific feeding situations develop (e.g. bottle aversion)
- Very variable in severity and frequency
- Most frequent in infants, but reported until school age.
Understanding the feeding problem in CHI...

• Correlated to the duration of initial stay in hospital
  – correlation to the timepoint of transfer to a highly specialized center has been reported (deLonlay, p.c.)

• Possible mechanisms behind
  – Insulin (CNS) is a strong regulator of appetite
    high CNS insulin $\rightarrow$ lower appetite
  – Psycho-interactive development of food aversion
Classical conditioning

• Pavlov's dog – experiment:
Do we condition our babies "not to eat"?

Hypo → Baby crying

Hypo → Baby crying

Combination

Feeding-Problem
Feeding problems in older children

1. "not-eating" → fear in parents

2. Not-eating → strong emotional reaction in parents
   → additional parental attention

→ Not-eating = more attention (from child's perspective)

- Disease-specific factors promote the development of feeding disorders in CHI-patients.
- Background could be psycho-interactive

→ Psycho-social interaction must be considered in specialized clinics. Parents need support to avoid typical mechanisms.
Tubing - blessing or curse...?

• Oral or nasal tubing
  – Promotes regurgitation and vomiting.
  – Obstacle in swallowing
  – Visual sign that the child is ill.
  → Only for limited periods adequate (day – weeks)

• PEG (Gastrostomy)
  – Invasive but invisible
  – Permanent
  – Parents might get dependent...?
Some practical aspects...
Basics

- Feeding-analysis and nutritional advising is part of every HI-treatment.
- Parent's problems and suggestions are important in understanding and treatment.
- Medication used in CHI has effects on feeding:
  - Diazoxid → less appetite, queasiness
  - Octreotid → abdominal pain after injection
  - Glucagone → nausea
Newborns / Infants

- prefer medication over feeding in treatment
- Awareness about mechanisms of conditioning.
- Enhance/establish feeding as a positive situation (mum and child)
- Try to keep input within age-appropriate caloric range.
- Try to feed the child according its own rhythm (chance to be hungry sometimes)
Toddler / school-children

• Don't let feeding become a power game
• Do not reward not-eating with giving better food (sweets)..
• Take meals together with other children and family
• Meals should be positive situations (for everyone)
Summary

• Nutrition is an important element in CHI-treatment

• Feeding should be age-appropriate (frequency, caloric intake, food)
  – Medical options should be tried first

• Feeding problems can have a psycho-interactive background – be aware (psychologist)!
  – Treatment is easier when earlier

• Most feeding difficulties resolve until school-age but are a main problem for many CHI families in earlier years.
Don't give up...
Unusual association of diseases/symptoms

Neonatal hyperinsulinism secondary to maternal intake of high-sugar drinks

Nicol Jon West,¹ Matthew Thorpe²

¹Department of Paediatrics, Bristol Children’s Hospital, Bristol, UK;

Krankheitsbild des schweren HI über 7 Tage, dann spontan gebessert....

Mutter hat 2 Liter Enegry-Drink pro Tag in der SS getrunken.
(entspricht 174g Glukose)
Long-Term Lanreotide Treatment in Six Patients with Congenital Hyperinsulinism

Peter Kühnen\textsuperscript{a} Jan Marquard\textsuperscript{b} Andrea Ernert\textsuperscript{a} Thomas Meissner\textsuperscript{b} Klemens Raile\textsuperscript{a} Gertrud Wannenmacher\textsuperscript{c} Oliver Blankenstein\textsuperscript{a}

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so much left to do…