MANAGING CONGENITAL HYPERINSULINISM AT SCHOOL

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Large portion of a child’s day spent in school.
SCHOOL

HEALTH CARE TEAM

FAMILY

CHI AT SCHOOL
CHI AT SCHOOL

- FAMILY
- SCHOOL
- HEALTH CARE TEAM

OPTIMAL MANAGEMENT
SAFETY
MAXIMAL ACADEMIC OPPORTUNITIES
1. Understanding of disease.

2. Trained in its management (specially emergency situations).
Basic HI overview:

- My child’s pancreas produces too much insulin.
- That can make him prone to hypoglycemia.
- What is hypoglycemia?
  - Blood glucose < 70 mg/dL.
Who?:
Personnel who interact with the child:
- Day care providers.
- Teachers.
- School administrators.
- School nurses.
- Coaches.
- Health aides.
- Bus drivers.
- Etc.
1. **Make a plan:**
   a. Specific needs.
   b. Specific instructions.

2. **Responsibilities:**
   ✓ Parent/Guardian.
   ✓ School personnel.
   ✓ Student.

3. **Expectations.**
1. **PLAN:**
   
a. **Blood glucose monitoring:**
   - Frequency.
   - Circumstances.

b. **Treatment administration:**
   - Doses.
   - Times.
   - Storage.

c. **Meals and snacks:**
   - Food content.
   - Amounts.
   - Timing.
d. Hypoglycemia:
   - Symptoms.
   - Treatment.

e. Physical activity.

e. Emergency evacuation/School lock-down instructions.
g. **Hyperglycemia:**
   - Symptoms.
   - Treatment.

h. **Checking for ketones:**
   - Appropriate actions.
Diabetes Medical Management Plan (DMMP)

This plan should be completed by the student’s personal diabetes health care team, including the parents/guardian. It should be reviewed with relevant school staff and copies should be kept in a place that can be accessed easily by the school nurse, trained diabetes personnel, and other authorized personnel.

Date of Plan: ___________ This plan is valid for the current school year: ____ - ____
Student’s Name: __________________________ Date of Birth: ___________________
Date of Diabetes Diagnosis: ___________ ☐ type 1 ☐ type 2 ☐ Other ___________

CONTACT INFORMATION

FAMILY

SCHOOL

HEALTH CARE TEAM

Pancrectectomy related
CHECKING BLOOD GLUCOSE

Target range of blood glucose: ☐ 70–130 mg/dL  ☐ 70–180 mg/dL
☐ Other: ____________________________

Check blood glucose level: ☐ Before lunch  ☐ ____ Hours after lunch
☐ 2 hours after a correction dose  ☐ Mid-morning  ☐ Before PE  ☐ After PE
☐ Before dismissal  ☐ Other: ____________________________

☐ As needed for signs/symptoms of low or high blood glucose
☐ As needed for signs/symptoms of illness

Preferred site of testing: ☐ Fingertip  ☐ Forearm  ☐ Thigh  ☐ Other: ______
Brand/Model of blood glucose meter: ____________________________

Note: The fingertip should always be used to check blood glucose level if hypoglycemia is suspected.

Continuous Glucose Monitor (CGM): ☐ Yes  ☐ No
Brand/Model: ____________________________ Alarms set for: ☐ (low) and ☐ (high)

Note: Confirm CGM results with blood glucose meter check before taking action on sensor blood glucose level. If student has symptoms or signs of hypoglycemia, check fingertip blood glucose level regardless of CGM.
HYPOGLYCEMIA TREATMENT

Student's usual symptoms of hypoglycemia (list below):

If exhibiting symptoms of hypoglycemia, OR if blood glucose level is less than ______ mg/dL, give a quick-acting glucose product equal to ______ grams of carbohydrate.

Recheck blood glucose in 10–15 minutes and repeat treatment if blood glucose level is less than ______ mg/dL.

Additional treatment:

15 grams of CH

15 min

< 70 mg/dL

< 70 mg/dL

AT ANY AGE: SUPERVISION!!!!!!
Explain your child’s most frequent precipitants

Table 1. Hypoglycemia signs and symptoms

<table>
<thead>
<tr>
<th>Autonomic signs and symptoms</th>
<th>Neuralglycopenic signs and symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shakiness</td>
<td>Poor concentration</td>
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<tr>
<td>Sweatiness</td>
<td>Blurred or double vision</td>
</tr>
<tr>
<td>Trembling</td>
<td>Disturbed color vision</td>
</tr>
<tr>
<td>Palpitations</td>
<td>Difficulty hearing</td>
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<tr>
<td>Pallor</td>
<td>Slurred speech</td>
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<td></td>
<td>Poor judgment and confusion</td>
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<td></td>
<td>Problems with short-term memory</td>
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<td></td>
<td>Dizziness and unsteady gait</td>
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<td></td>
<td>Loss of consciousness</td>
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<td></td>
<td>Seizure</td>
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<tr>
<td></td>
<td>Death</td>
</tr>
<tr>
<td>Behavioral signs and symptoms</td>
<td>Irritability</td>
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<tr>
<td></td>
<td>Erratic behavior</td>
</tr>
<tr>
<td></td>
<td>Agitation</td>
</tr>
<tr>
<td></td>
<td>Nightmares</td>
</tr>
<tr>
<td></td>
<td>Inconsolable crying</td>
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<tr>
<td>Non-specific symptoms</td>
<td>Hunger</td>
</tr>
<tr>
<td></td>
<td>Headache</td>
</tr>
<tr>
<td></td>
<td>Nausea</td>
</tr>
<tr>
<td></td>
<td>Tiredness</td>
</tr>
</tbody>
</table>

Explain your child’s precipitants

Table 2. Clinical factors associated with hypoglycemia

<table>
<thead>
<tr>
<th>Precipitants</th>
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</thead>
<tbody>
<tr>
<td>Excess insulin</td>
</tr>
<tr>
<td>Less food consumption</td>
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<tr>
<td>Exercise</td>
</tr>
<tr>
<td>Sleep</td>
</tr>
<tr>
<td>Alcohol ingestion</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Risk factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Younger age, &lt;6 yr</td>
</tr>
<tr>
<td>Lower A1C levels</td>
</tr>
<tr>
<td>Hypoglycemia unawareness</td>
</tr>
<tr>
<td>Previous severe hypoglycemia</td>
</tr>
<tr>
<td>Longer duration of diabetes</td>
</tr>
</tbody>
</table>
Glucagon Injection:
For severe lows
Sick; vomiting; too weak or having a seizure.
Inject into a muscle
Recheck in 20-30 minutes and go to Hospital.
HYPERGLYCEMIA TREATMENT

Student’s usual symptoms of hyperglycemia (list below):

________________________________________________________________________

Check  □ Urine  □ Blood for ketones every _____ hours when blood glucose levels are above _____ mg/dL.

For blood glucose greater than _____ mg/dL AND at least _____ hours since last insulin dose, give correction dose of insulin (see orders below).

For insulin pump users: see additional information for student with insulin pump.

Give extra water and/or non-sugar-containing drinks (not fruit juices): _____ ounces per hour.

Additional treatment for ketones: ______________________________________

Follow physical activity and sports orders (see page 7).

• Notify parents/guardian of onset of hyperglycemia.

• If the student has symptoms of a hyperglycemia emergency, including dry mouth, extreme thirst, nausea and vomiting, severe abdominal pain, heavy breathing or shortness of breath, chest pain, increasing sleepiness or lethargy, or depressed level of consciousness: Call 911 (Emergency Medical Services) and the student’s parents/guardian.

• Contact student’s health care provider.
Diabetes Medical Management Plan (DMMP) – page 4

INSULIN THERAPY

Insulin delivery device:  □ syringe  □ insulin pen  □ insulin pump

Type of insulin therapy at school:

□ Adjustable Insulin Therapy
□ Fixed Insulin Therapy
□ No insulin

Adjustable Insulin Therapy

• Carbohydrate Coverage/Correction Dose:
  Name of insulin:

• Carbohydrate Coverage:
  Insulin-to-Carbohydrate Ratio:
  Lunch: 1 unit of insulin per _____ grams of carbohydrate
  Snack: 1 unit of insulin per _____ grams of carbohydrate

Carbohydrate Dose Calculation Example

\[
\text{Grams of carbohydrate in meal} \quad \text{Insulin-to-carbohydrate ratio} = _____ \text{ units of insulin}
\]

• Correction Dose:
  Blood Glucose Correction Factor/Insulin Sensitivity Factor = _____
  Target blood glucose = _____ mg/dL

Correction Dose Calculation Example

\[
\text{Actual Blood Glucose–Target Blood Glucose} \quad \text{Blood Glucose Correction Factor/Insulin Sensitivity Factor} = _____ \text{ units of insulin}
\]
Diabetes Medical Management Plan (DMMP) – page 5

INSULIN THERAPY (Continued)

When to give insulin:
Lunch
☑ Carbohydrate coverage only
☑ Carbohydrate coverage plus correction dose when blood glucose is greater than _____mg/dL and ____ hours since last insulin dose.
☐ Other: __________________________________________________________________________

Snack
☐ No coverage for snack
☑ Carbohydrate coverage only
☑ Carbohydrate coverage plus correction dose when blood glucose is greater than _____mg/dL and ____ hours since last insulin dose.
☐ Other: __________________________________________________________________________

☑ Correction dose only: For blood glucose greater than _____mg/dL AND at least ____ hours since last insulin dose.
☐ Other: __________________________________________________________________________

Fixed Insulin Therapy
Name of insulin: __________________________________________________________________
☐ ____ Units of insulin given pre-lunch daily
☐ ____ Units of insulin given pre-snack daily
☐ Other: __________________________________________________________________________

Parental Authorization to Adjust Insulin Dose:
☐ Yes  ☐ No Parents/guardian authorization should be obtained before administering a correction dose.
☐ Yes  ☐ No Parents/guardian are authorized to increase or decrease correction dose scale within the following range: +/- ____ units of insulin.
☐ Yes  ☐ No Parents/guardian are authorized to increase or decrease insulin-to-carbohydrate ratio within the following range: ____ units per prescribed grams of carbohydrate, +/- ____ grams of carbohydrate.
☐ Yes  ☐ No Parents/guardian are authorized to increase or decrease fixed insulin dose within the following range: +/- ____ units of insulin.
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Diabetes Medical Management Plan (DMMP) – page 6

INSULIN THERAPY (Continued)

ADDITIONAL INFORMATION FOR STUDENT WITH INSULIN PUMP

<table>
<thead>
<tr>
<th>Brand/Model of pump:</th>
<th>Type of insulin in pump:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basal rates during school:</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Type of infusion set:</th>
</tr>
</thead>
</table>

- □ For blood glucose greater than _____ mg/dL that has not decreased within ______ hours after correction, consider pump failure or infusion site failure. Notify parents/guardian.
- □ For infusion site failure: Insert new infusion set and/or replace reservoir.
- □ For suspected pump failure: suspend or remove pump and give insulin by syringe or pen.

Physical Activity

May disconnect from pump for sports activities  □ Yes  □ No
Set a temporary basal rate  □ Yes  □ No  % temporary basal for ____ hours
Suspend pump use  □ Yes  □ No

Student’s self-care pump skills:

<table>
<thead>
<tr>
<th>Count carbohydrates</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Bolus correct amount for carbohydrates consumed</th>
<th>Independent?:</th>
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</table>

<table>
<thead>
<tr>
<th>Calculate and administer correction bolus</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Calculate and set basal profiles</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Calculate and set temporary basal rate</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Change batteries</th>
<th>Independent?:</th>
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</thead>
</table>

<table>
<thead>
<tr>
<th>Disconnect pump</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Reconnect pump to infusion set</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Prepare reservoir and tubing</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Insert infusion set</th>
<th>Independent?:</th>
</tr>
</thead>
</table>

| Troubleshoot alarms and malfunctions | Independent?: |

104  www.YourDiabetesInfo.org
Diabetes Medical Management Plan (DMPM) – page 7

OTHER DIABETES MEDICATIONS
Name: ____________________ Dose: __________ Route: ________ Times given: ________
Name: ____________________ Dose: __________ Route: ________ Times given: ________

MEAL PLAN
Meal/Snack Time Carbohydrate Content (grams)
Breakfast ____________________ ________ to ________
Mid-morning snack ____________________ ________ to ________
Lunch ____________________ ________ to ________
Mid-afternoon snack ____________________ ________ to ________

Other times to give snacks and content/amount:

Instructions for when food is provided to the class (e.g., as part of a class party or food sampling event):

Special event/party food permitted: □ Parents/guardian discretion
□ Student discretion

Student’s self-care nutrition skills:
□ Yes □ No Independently counts carbohydrates
□ Yes □ No May count carbohydrates with supervision
□ Yes □ No Requires school nurse/trained diabetes personnel to count carbohydrates

PHYSICAL ACTIVITY AND SPORTS
A quick-acting source of glucose such as □ glucose tabs and/or □ sugar-containing juice must be available at the site of physical education activities and sports.

Student should eat □ 15 grams □ 30 grams of carbohydrate □ other __________
□ before □ every 30 minutes during □ after vigorous physical activity
□ other __________

If most recent blood glucose is less than __________ mg/dL, student can participate in physical activity when blood glucose is corrected and above __________ mg/dL.

Avoid physical activity when blood glucose is greater than __________ mg/dL or if urine/blood ketones are moderate to large.

(Additional information for student on insulin pump is in the insulin section on page 6.)
Diabetes Medical Management Plan (DMMP) – page 8

**DISASTER PLAN**
To prepare for an unplanned disaster or emergency (72 HOURS), obtain emergency supply kit from parent/guardian.

- Continue to follow orders contained in this DMMP.
- Additional insulin orders as follows: __________________________________________
- Other: __________________________________________

**SIGNATURES**
This Diabetes Medical Management Plan has been approved by:

<table>
<thead>
<tr>
<th>Student’s Physician/Health Care Provider</th>
<th>Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>I, (parent/guardian:____________________ give permission to the school nurse or another qualified health care professional or trained diabetes personnel of (school:____________________ to perform and carry out the diabetes care tasks as outlined in (student:____________________’s Diabetes Medical Management Plan. I also consent to the release of the information contained in this Diabetes Medical Management Plan to all school staff members and other adults who have responsibility for my child and who may need to know this information to maintain my child’s health and safety. I also give permission to the school nurse or another qualified health care professional to contact my child’s physician/health care provider.</td>
<td></td>
</tr>
<tr>
<td>Acknowledged and received by:</td>
<td>Date</td>
</tr>
<tr>
<td>Student’s Parent/Guardian</td>
<td>Date</td>
</tr>
<tr>
<td>Student’s Parent/Guardian</td>
<td>Date</td>
</tr>
<tr>
<td>School Nurse/Other Qualified Health Care Personnel</td>
<td>Date</td>
</tr>
</tbody>
</table>
HI INFORMATION FOR NURSERY/PRESCHOOL

NAME: Lucas. 3 yo.
DIAGNOSIS: Congenital Hyperinsulinism.
QUICK INFORMATION:
   Lucas has a genetic disease with inappropriate insulin secretion that can cause hypoglycemia (BG <70).
   That’s why he requires measurement of blood sugars frequently and assessment of medical and nutrition and exercise.
MEDICINE: Diazoxide BID (reduces insulin secretion).
TIMES TO CHECK BLOOD SUGARS: He can’t do it by himself (completely dependant).
   - Before snack (11:00 am).
   - Before lunch (1:00 pm).
   - 2 hours after lunch (3:00 – 3:30 pm).
   - When suspicion of hypoglycemia: Lucas will usually say that he feels hunger or headache. See other possible symptoms. See attached hypoglycemia plan action.
FASTING TOLERANCE: 8 hours (4 when he’s sick).
   - Medications: Diazoxide BID (given at home).
   - Snack (11:00 am): In his bag (4 Maria cookies or chocolate granola bar)
   - Lunch (1:00 am): Days 09, 17 and 28 will have to add to the school menu an extra 20 g bread slice (in his bag).
CONTACT INFORMATION:
   - Luis (Father): 685663328. María (mother): 673209645
   - Dr. L. Salamanca Fresno (La Paz Hospital): 917277210

HYPOGLYCEMIA (BG <70 mg/dL):
1. Give Lucas 100 cc orange juice (it’s in his bag) + 1 cookie.
2. Recheck BG in 10 minutes.
3. If Lucas is still < 70 mg/dL give to Lucas 100 cc orange juice (it’s in his bag) + 1 cookie again.
4. Recheck BG in 10-15 minutes again.
5. If Lucas is still < 70 mg/dL inject Lucas in his thigh 0.5 mg of Glucagon (nurse’s fridge) and call parents (María and Luis) and Hospital Reference (Hospital La Paz, Madrid).
6. If at any time Lucas is unconcious or unable to PO eating start hypo treatment in point 5. (Glucagon)

IMPORTANT: Lucas must never be left alone until hypoglycemia has resolved!!!!
2. RESPONSIBILITIES:

a. Parent/Guardian:
   - **Materials:**
     - Equipment/Insulin/Medications.
     - Maintenance of blood glucose monitoring equipment.
     - Ensure proper disposal of materials.
     - Separate record logbook kept at school.
   - **Plan** completed/signed by diabetes health care team.
- Supplies to treat hypoglycemia:
  - Source of glucose.
  - Glucagon emergency kit.

- Information about HI.

- Emergency phone numbers:
  - Parent/Guardian.
  - Diabetes health care team.
- Student’s meal/snack schedule.
  - Closer as possible with other students’
  - Including: Parties, activities etc.

- Signed release of confidentiality from legal guardian.
b. **School:**

- **School nurse:** Opportunities for training and education.
- **Training for school personnel:**
  - Basic HI overview.
  - Typical needs.
  - Recognition of hypo or hyperglycemia.
  - Who to contact for help?
- Immediate accessibility of hypoglycemia's treatment.
  ALWAYS Supervision!!!

- Accessibility to scheduled treatments (Plan).

- School location providing privacy or classroom or anywhere:
  - Blood glucose monitoring.
  - Treatment administration.
- Know student meal/snack schedule:
  - Remind snack times.
  - Notify parent/guardian expected changes:
    - Meal times.
    - Exercise routine.

- **Permission** for capable students:
  - Carry equipment, supplies, medication, snacks.
  - Cell phone access.
- Permission to see **school nurse/personnel** if requested.
- Permission to eat a **snack** anywhere.
- Permission to use the **restroom** and access to fluids.
- Permission to **miss school** without consequences.
a. Toddlers and preschool-aged children:
  - Unable to perform tasks independently: Adult required.
  - Many: Difficulty in recognizing hypoglycemia symptoms.

b. Elementary school-aged children:
  - Many: Own blood glucose checks: Adult supervising.
  - Older: Beginning to self-administer treatment: Adult supervising.
  - Understand effect of physical activity and nutrition.
  - Some: Hypoglycemia unawareness.

c. Middle school- and high school-aged children:
  - Usually able to provide self care.
  - Need help when severe hypoglycemia.
Student’s competence and capability for HI tasks set out in Plan.

- At all ages: Help for blood glucose check when hypoglycemia.
  - Reminder for eat/drink.
  - Supervised until treatment has taken place and blood glucose value has returned to normal range.

- Emergency situations: Always help.
1. Feeding issues.
2. Blood glucose monitoring.
3. Neurobehavioral deficits.
4. Medications/Insulin.
5. Sports.
1. **FEEDING ISSUES:**

   - Feeding: major role in HI management:
     - Frequent feeding.

   - Management:
     - Oral feeding.
     - Gastrostomy feedings.
     - Oral and nasogastric tube feeds.

   - Food aversion: significantly prevalent.
   - Independent of development delay.
Food aversion among patients with persistent hyperinsulinemic hypoglycemia of infancy

Saud Al-Shanafey*, Hussain AlKhudhur

King Faisal Specialist Hospital and Research Center, PO Box 3354, MBC 40, Riyadh 11211, Saudi Arabia
Protein-induced hypoglycemia is a feature of KATP-HI.

Despite the absence of leucine sensitivity (GDH-HI).

Aa can stimulate insulin secretion.

OPTT (Oral Protein Tolerance Tests): 1,0-1,5 g/Kg of Protein (eggs, protein and RESOURCE Instant Protein Powder).
Responses to oral protein tolerance test in patients with recessive $K_{ATP}$ hyperinsulinism

<table>
<thead>
<tr>
<th>Patient</th>
<th>Baseline BG (mg/dL)</th>
<th>Nadir BG (mg/dL)</th>
<th>Delta BG (mg/dL)</th>
<th>Time to nadir (min)</th>
<th>Delta insulin (μU/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>97</td>
<td>48</td>
<td>-49</td>
<td>120</td>
<td>49</td>
</tr>
<tr>
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<td>109</td>
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<td>10</td>
<td>76</td>
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<tr>
<td>11</td>
<td>71</td>
<td>47</td>
<td>-24</td>
<td>180</td>
<td>ND</td>
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</table>

Mean ± SD
- Baseline BG: 94 ± 17
- Nadir BG: 58 ± 10‡
- Delta BG: -36 ± 18‡
- Time to nadir: 80 ± 50
- Delta insulin: 44 ± 44‡

GDH-HI‡ (n = 12)
- Baseline BG: 74 ± 14 (60–106)
- Nadir BG: 50 ± 9‡ (35–72)
- Delta BG: -23 ± 16* (-9 to -62)
- Time to nadir: 108 ± 66 (28–180)
- Delta insulin: 14 ± 11 (1–30)

Control subjects (n = 12)
- Baseline BG: 85 ± 7 (74–94)
- Nadir BG: 79 ± 7 (71–90)
- Delta BG: -6 ± 6 (-13 to 2)
- Time to nadir: 70 ± 60 (15–180)
- Delta insulin: 10 ± 8 (0–27)

*P < .05 versus control subjects.
‡P < .0001 versus control subjects.
Reference 21.
Leucine

- Red meat, sausage, viscera, fish, cheese, yoghurt, eggs.
- Whole wheat and cereals, legume, corn, potatoes, sesame, soy.
- Dried fruits (pistachios, peanuts).

Glutamine:

- Dairy products.
- Raw meats.
- Salmon.
- Eggs.
- Miso, soy.
- Cabbage.
REGULAR PATTERN:
Eat 3 meals and snacks a day--every day.

Same amount at the same time every day
Avoid skipping meals.
Diet:

- **Frequent high-carbohydrate feedings**: formula supplemented with glucose polymer.
- **Continuous feedings through nasogastric or gastric tube**.
- **Cornstarch**: slow-release carbohydrate.
- **Avoidance of protein-rich meals**.
2. **BLOOD GLUCOSE MONITORING:**

- **CBG:**
  - When?
  - How?
  - Who?

- **CGMS:**

  Glucose *excursions*:
  - Correct medication titration and administration.
  - Exercise effect.
  - Meals/Snacks.

Useful information about **trends**.

*ALWAYS confirm with CBG*
CGM could be useful in HI

- Detects if the blood sugar is already decreasing before exercise or activity.
- Indicates if you are responding to low blood sugar treatment.
- Shows the stability or instability of blood sugars at school before walking home or getting on the bus.
**Abstract**

**Congenital hyperinsulinism** (CHI) is the commonest cause of persistent hypoglycemia in neonates. Diazoxide is the first-line drug in its treatment, but the more severe cases are usually diazoxide-resistant. Recessive ABCC8 and KCNJ11 mutations are responsible for most (82%) of the severe diazoxide-unresponsive CHI. Oral nifedipine has been effective in isolated cases of CHI. Successful treatment of diazoxide-unresponsive CHI with a combination of octreotide and nifedipine has been reported in a single isolated case so far. We report here a case of diazoxide-resistant CHI due to homozygous ABCC8 nonsense mutation. In this case, hypoglycaemia uncontrolled by pancreatectomy and octreotide alone showed a good response to a combination of nifedipine and octreotide. Octreotide was tapered off by one year age and thereafter the child is euglycaemic on oral nifedipine alone. Continuous glucose monitoring sensor was used as an aid to monitor glycaemic control and was found to be a safe and reliable option reducing the number of needle-pricks in small children.
3. **NEUROBEHAVIORAL DEFICITS:**

- **Recurrent hypoglycemia:** Risk of neurocognitive dysfunction.
- **Surgical HI:** Higher risk.
  - 1980’s studies: 50% HI neurologic dysfunction.
  - Other studies:
    - Neurodevelopmental abnormalities 26-46%.
    - Epilepsy: 25-43%.
Abnormal Neurodevelopmental Outcomes are Common in Children with Transient Congenital Hyperinsulinism.


39% (Avatapalle et al., 2013).

- Speech, language, motor and vision.
- TCHI and PCHI: Similar incidence.
- Risk factors:
  - Early presentation (neonatal).
  - Severe CHI.
**Table 3.** Prevalence of Reported Neurobehavioral Abnormalities

<table>
<thead>
<tr>
<th>Psychiatric/Behavioral</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech delay</td>
<td>22 (18%)</td>
</tr>
<tr>
<td>Learning disability</td>
<td>19 (16%)</td>
</tr>
<tr>
<td>Seizures</td>
<td>16 (13%)</td>
</tr>
<tr>
<td>Physical disability</td>
<td>13 (11%)</td>
</tr>
<tr>
<td>ADHD</td>
<td>12 (10%)</td>
</tr>
<tr>
<td>Autism</td>
<td>2 (2%)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>58 (48%)</strong></td>
</tr>
</tbody>
</table>

**Table 4.** Neurobehavioral Measures

<table>
<thead>
<tr>
<th>Adaptive Behavior Assessment System – II* (n = 69)</th>
<th>Mean ± SD</th>
<th>% &lt; 1 SD</th>
<th>% &lt; 2 SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>General adaptive composite score</td>
<td>96 ± 25</td>
<td>27.5³</td>
<td>18.8³</td>
</tr>
<tr>
<td>Conceptual composite score</td>
<td>98 ± 22</td>
<td>21.2</td>
<td>11.8³</td>
</tr>
<tr>
<td>Social composite score</td>
<td>100 ± 21</td>
<td>22.1</td>
<td>14.7³</td>
</tr>
<tr>
<td>Practical composite score</td>
<td>92 ± 25</td>
<td>30.9³</td>
<td>16.2³</td>
</tr>
<tr>
<td>Child Behavior Checklist* (n = 62)</td>
<td>Mean ± SD</td>
<td>% &gt; 1 SD</td>
<td>% &gt; 2 SD</td>
</tr>
<tr>
<td>Total problems</td>
<td>49 ± 16</td>
<td>16.1</td>
<td>8.1³</td>
</tr>
<tr>
<td>Internalizing problems</td>
<td>49 ± 13</td>
<td>16.1</td>
<td>9.7³</td>
</tr>
<tr>
<td>Externalizing problems</td>
<td>47 ± 11</td>
<td>11.5</td>
<td>6.5³</td>
</tr>
</tbody>
</table>

* Normal population mean is 100 with SD of 15; higher scores are more favorable
* Normal population mean is 50 with SD of 10; lower scores are more favorable
³P ≤ 0.02; ¹P ≤ 0.01; ⁵P ≤ 0.001 compared to normal population.
Developmental assessment is essential in HI!!!!:

- Only a quarter reported receiving formal assessment.
- Many only identified after struggling academically in school.

Why get a developmental assessment?

Early identification = early treatment

- Developmental therapies
- Special education services/supports
- Special insurance benefits
- Increased access to behavioral support services
- Increased understanding among family, teachers, etc.
When to get a developmental assessment?
- After discharge from diagnostic admission (**baseline**).
- Any time milestones seem to be lagging.
- Probably helpful at certain ages:
  - Kindergarten readiness
  - Between 1st and 3rd grades (higher chance of finding a learning disability)
4. **MEDICATIONS/INSULIN:**

Pancreatectomy increases the risk for diabetes:

- 36% CHOP series.
  - Median age 7.7 years (first 2 decades of life).
  - Treatment:
    - Insulin: 86%.
    - Antidiabetic medications (BMI >25 Kg/m2).
    - Diet modifications.
  - Other studies: 42% < 8 years.

*J Clin Endocrinol Metab.* 2015 Sep 1;jc20152539. [Epub ahead of print]

**High risk of diabetes and neurobehavioral deficits in individuals with surgically treated hyperinsulinism.**

**Lord K**, Radcliffe J, Gallagher PR, Adzick NS, Stanley CA, De León DD.
Explain your child’s HI treatments:

**Diazoxide:**
- **Dose:** 5-15 mg/kg/day by mouth
- **Side effects:**
  - Excessive body hair
  - Suppression of appetite
6. **SPORTS:**
   
a. No HI, no diabetes.

b. HI without diabetes.

c. HI with subsequent diabetes.
a. **No HI, no diabetes:**

- Organism detects lowering in glucose and therefore reduces insulin production.
- Not enough?: Very large duration. Liver (glyconeogenesis and glycogenolysis).
- Consequence: Not probably hypoglycemic.
a. **HI without diabetes:**
- Insulin is not inhibited (medications).
- Insulin modulates glyconeogenesis.
- Consequence: *More prone to hypoglycemia.*

b. **HI with subsequent diabetes:**
2 options:
- Exercise + insulin excess: *Hypo possible.*
- Exercise + insulin deficit: *Liver effect. Hyper.*
a. **HI without diabetes:**
   - More prone to hypoglycemia.
   - Strict control/medications/meals & snacks.

b. **HI with subsequent diabetes:**
   - Lower insulin dose but not avoid it.
   - BG (instructions in DMMT):
     - <100: 5-15 g HC and delay exercise 10-15 minutes.
     - 100-250: Start exercise normally.
     - >250: Check ketones: >1: AAR+ HC.
# Meals & Snacks

<table>
<thead>
<tr>
<th></th>
<th>Low Intensity</th>
<th>Medium Intensity</th>
<th>High Intensity</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;30 min</td>
<td>No</td>
<td>No</td>
<td>20 g</td>
</tr>
<tr>
<td>30-60 min</td>
<td>10-20 g</td>
<td>30 g</td>
<td>50 g</td>
</tr>
<tr>
<td>&gt; 60 min</td>
<td>15-25 g / h</td>
<td>20-70 g / h</td>
<td>30-100 g / h</td>
</tr>
</tbody>
</table>

**Moderate or low** glycemic index CH.

**High** glycemic index:
- During exercise.
- Prior if BG <100-130.
- After if < 100.
Exercise type:

✓ **Cardiovascular resistance:** Walking, running, swimming, bicycle riding, rowing. **High hypo effect.** During exercise and even 12-24 hours post.

✓ Muscle effect exercise: weight lifting, push-ups and sit-ups. Less hypo effect, even hyper.

Intensity:

✓ Low Intensity: <60% maximum heart rate. Walking, swimming. Low hypoglycemic effect.
✓ Medium Intensity: 60-75%
✓ **High Intensity:** > 75%. **High hypo effect** but high amounts of glucose produced by contrarregullar hormones.
TAKE HOME

- The closest communication: Family/School/Health care team = BEST OUTCOME.
- Make a Plan for your child at school. SIMPLE AND CLEAR.
- When hypoglycemia is detected, children must be supervised (AT ALL AGES).
- Auto-capability must be detailed in plan.
- Meal & snack schedule must be similar or close to other student´s.
- Avoide protein-rich meals.
- Developmental assessment is essential in HI.
- Sports and exercise are healthy habits and must not be prohibited but hypos must be prevented.
SPECIAL THANKS

- Dr. Diva D. De Leon Crutchlow
THANK YOU!