# MANAGING CONGENITAL HYPERINSULINISM AT SCHOOL


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# CHI AT SCHOOL

- Large portion of a child´s day spent in school.

# CHI AT SCHOOL



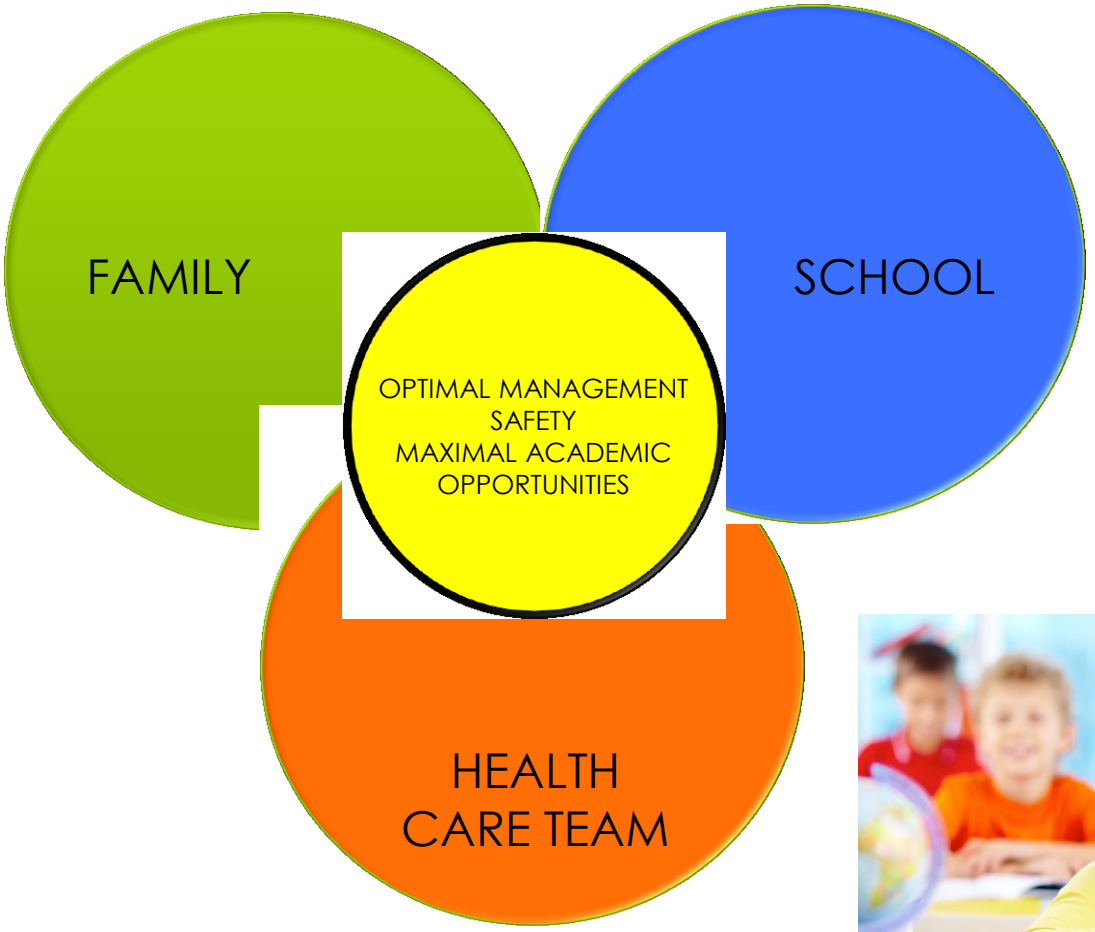
# CHI AT SCHOOL

FAMILY

SCHOOL

HEALTH
CARE TEAM

# CHI AT SCHOOL




# CHI AT SCHOOL


1. Understanding of disease.

2. Trained in its management (specially emergency situations).

# CHI AT SCHOOL

- **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.

# CHI AT SCHOOL

- **Who?:**

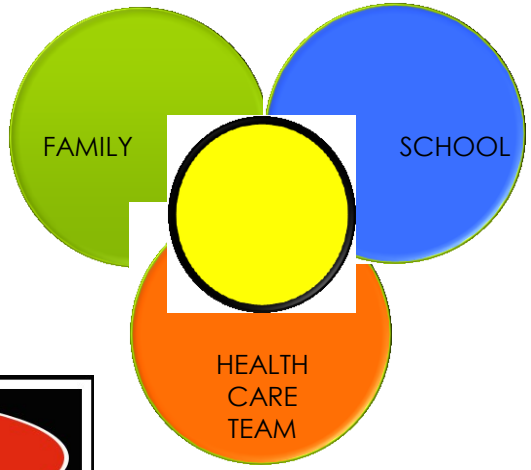
  Personnel who interact with the child:

  - Day care providers.
  - Teachers.
  - School administrators.
  - School nurses.
  - Coaches.
  - Health aides.
  - Bus drivers.
  - Etc.

# CHI AT SCHOOL



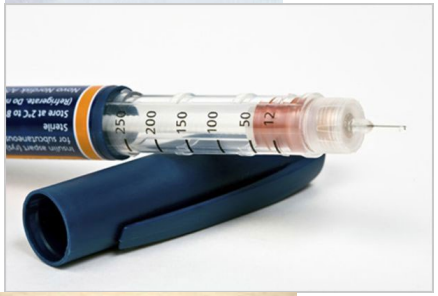
1. **Make a plan:**
   a. Specific needs.
   b. Specific instructions.
2. **Responsibilities:**
   - ✓ Parent/Guardian.
   - ✓ School personnel.
   - ✓ Student.
3. **Expectations.**



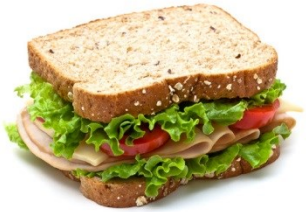

# CHI AT SCHOOL


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

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

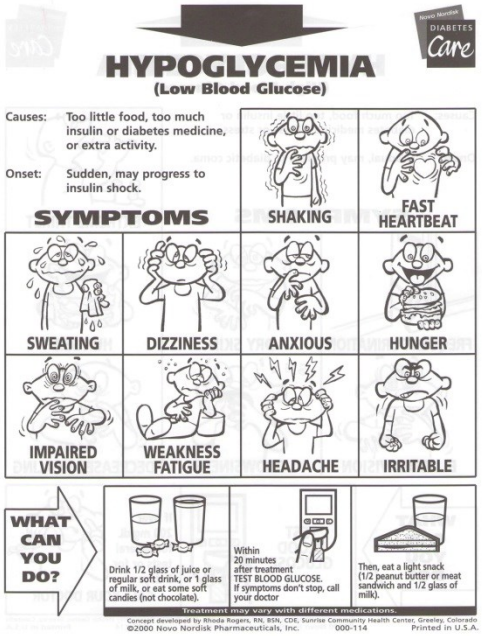



   c. **Meals and snacks:**
      - Food content.
      - Amounts.
      - Timing.



# CHI AT SCHOOL


d. **Hypoglycemia:**
   - Symptoms.
   - Treatment.



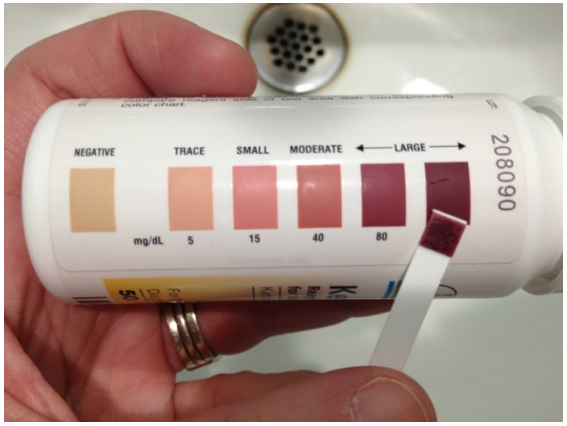

e. **Physical activity.**

e. **Emergency evacuation/School lock-down instructions.**

# CHI AT SCHOOL


g. **Hyperglycemia:**
   g. Symptoms.
   h. Treatment.

h. **Checking for ketones:**
   g. Appropriate actions.

# CHI AT SCHOOL


Pancretectomy related

## Diabetes Medical Management Plan (DMMP)

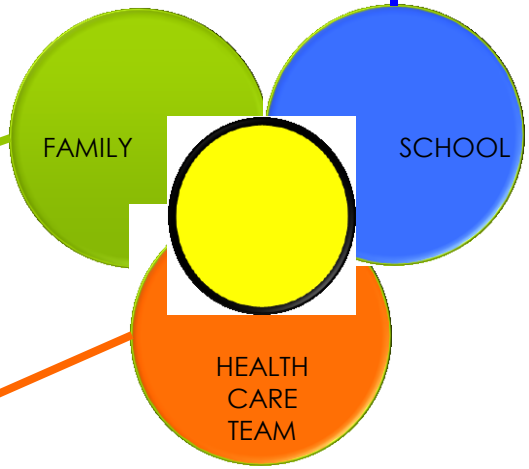
This plan should be completed by the student's personal diabetes health care team, including the parents/guardian. It should be reviewed with relevant school staff and copies should be kept in a place that can be accessed easily by the school nurse, trained diabetes personnel, and other authorized personnel.

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**



# CHI AT SCHOOL


**Diabetes Medical Management Plan (DMMP) – Page 2**

## 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.*


# CHI AT SCHOOL


**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: _____



MEDICAL PROGRESS

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## Recommendations from the Pediatric Endocrine Society for Evaluation and Management of Persistent Hypoglycemia in Neonates, Infants, and Children

Paul S. Thornton, MB, BCh[1], Charles A. Stanley, MD[2], Diva D. De Leon, MD, MSCE[2], Deborah Harris, PhD[3], Morey W. Haymond, MD[4], Khalid Hussain, MD, MPH[5], Lynne L. Levitsky, MD[6], Mohammad H. Murad, MD, MPH[7], Paul J. Rozance, MD[8], Rebecca A. Simmons, MD[9], Mark A. Sperling, MBBS[10], David A. Weinstein, MD, MMSc[11], Neil H. White, MD[12], and Joseph I. Wolfsdorf, MB, BCh[13]

< 70 mg/dL

15 grams of CH

15 min

< 70 mg/dL

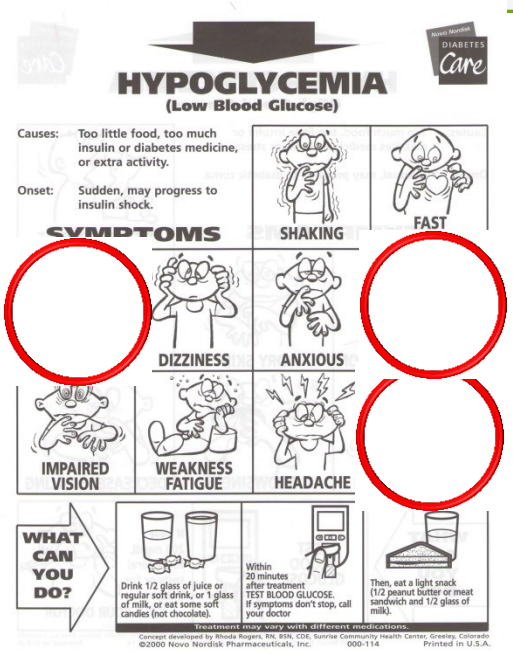
AT ANY AGE: SUPERVISION!!!!!!

## Explain your child´s most frequent

**Ly et al.**

Table 1. Hypoglycemia signs and symptoms

| |
|---|
| *Autonomic signs and symptoms* |
| Shakiness |
| Sweatiness |
| Trembling |
| Palpitations |
| Pallor |
| *Neuroglycopenic signs and symptoms* |
| Poor concentration |
| Blurred or double vision |
| Disturbed color vision |
| Difficulty hearing |
| Slurred speech |
| Poor judgment and confusion |
| Problems with short-term memory |
| Dizziness and unsteady gait |
| Loss of consciousness |
| Seizure |
| Death |
| *Behavioral signs and symptoms* |
| Irritability |
| Erratic behavior |
| Agitation |
| Nightmares |
| Inconsolable crying |
| *Non-specific symptoms* |
| Hunger |
| Headache |
| Nausea |
| Tiredness |




## Explain your child´s precipitants

Table 2. Clinical factors associated with hypoglycemia

| |
|---|
| *Precipitants* |
| Excess insulin |
| Less food consumption |
| Exercise |
| Sleep |
| Alcohol ingestion |
| *Risk factors* |
| Younger age, <6 yr |
| Lower A1C levels |
| Hypoglycemia unawareness |
| Previous severe hypoglycemia |
| Longer duration of diabetes |

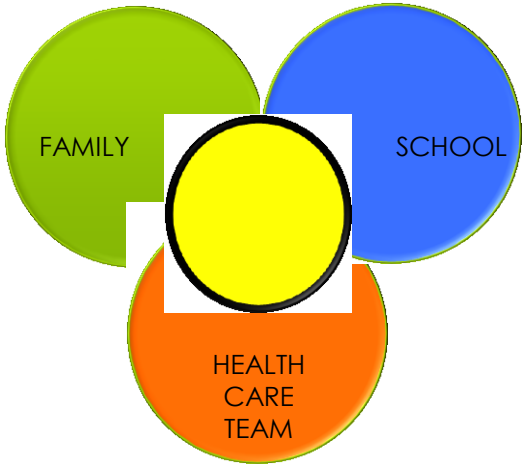
# CHI AT SCHOOL


**Diabetes Medical Management Plan (DMMP) – Page 3**

**HYPOGLYCEMIA TREATMENT** (Continued)

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

- If the student is unable to eat or drink, is unconscious or unresponsive, or is having seizure activity or convulsions (jerking movements), give:
- Glucagon: ❑ 1 mg ❑ 1/2 mg Route: ❑ SC ❑ IM
- Site for glucagon injection: ❑ arm ❑ thigh ❑ Other:_____
- Call 911 (Emergency Medical Services) and the student's parents/guardian.
- Contact student's health care provider.





## 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.

# CHI AT SCHOOL


## 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).

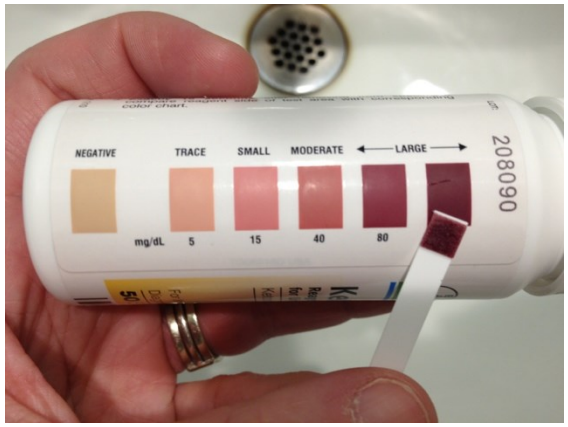
For insulin pump users: see additional information for student with insulin pump.

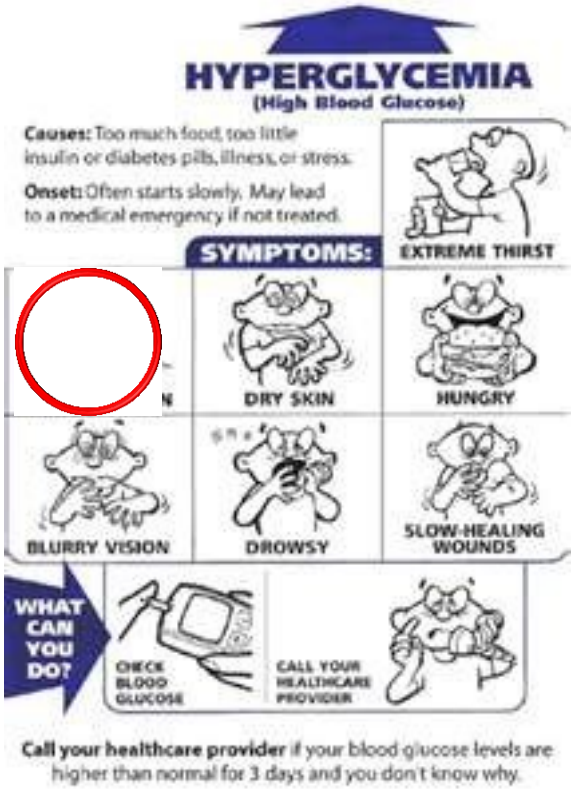
Give extra water and/or non-sugar-containing drinks (not fruit juices): _____ounces per hour.

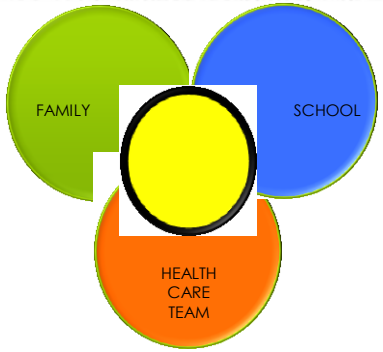
Additional treatment for ketones: _____

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.



# CHI AT SCHOOL


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**

$$\frac{\textit{Grams of carbohydrate in meal}}{\textit{Insulin-to-carbohydrate ratio}} = \_\_\_\_\_ \text{ units of insulin}$$

- **Correction Dose:**

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

**Correction Dose Calculation Example**

$$\frac{\textit{Actual Blood Glucose–Target Blood Glucose}}{\textit{Blood Glucose Correction Factor/Insulin Sensitivity Factor}} = \_\_\_\_\_ \text{ units of insulin}$$

# CHI AT SCHOOL


**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.


# CHI AT SCHOOL




**INSULIN THERAPY** (Continued)

## ADDITIONAL INFORMATION FOR STUDENT WITH INSULIN PUMP

Brand/Model of pump: _____ Type of insulin in pump: _____

Basal rates during school: _____

Type of infusion set: _____

❑ 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:** | **Independent?** |
|---|---|
| Count carbohydrates | ❑ Yes ❑ No |
| Bolus correct amount for carbohydrates consumed | ❑ Yes ❑ No |
| Calculate and administer correction bolus | ❑ Yes ❑ No |
| Calculate and set basal profiles | ❑ Yes ❑ No |
| Calculate and set temporary basal rate | ❑ Yes ❑ No |
| Change batteries | ❑ Yes ❑ No |
| Disconnect pump | ❑ Yes ❑ No |
| Reconnect pump to infusion set | ❑ Yes ❑ No |
| Prepare reservoir and tubing | ❑ Yes ❑ No |
| Insert infusion set | ❑ Yes ❑ No |
| Troubleshoot alarms and malfunctions | ❑ Yes ❑ No |



# CHI AT SCHOOL




## 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.

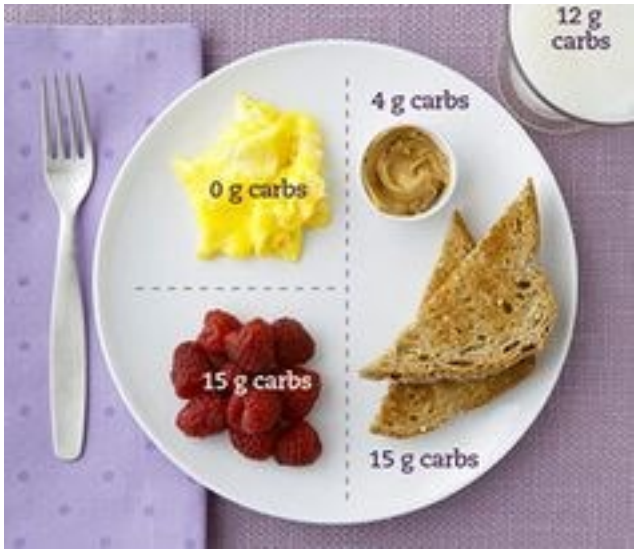
Student should eat ❑ 15 grams ❑ 30 grams of carbohydrate ❑ other_____
❑ before ❑ every 30 minutes during ❑ after vigorous physical activity
❑ other _____

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.)






## FASTING TOLERANCE

# CHI AT SCHOOL


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:

_____

Student's Physician/Health Care Provider Date

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.

_____

Acknowledged and received by:

_____

Student's Parent/Guardian Date

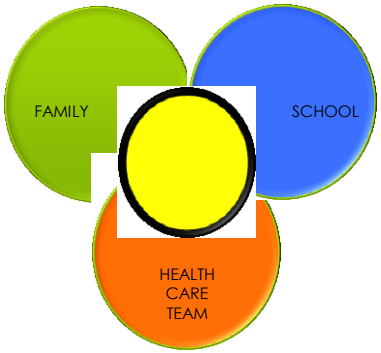
_____

Student's Parent/Guardian Date

_____

School Nurse/Other Qualified Health Care Personnel Date





# CHI AT SCHOOL

MAKE YOUR OWN PLAN!!!

## HI INFORMATION FOR NURSERY/PRESCHOOL

**NAME:** Lucas. 3 yo.

**DIAGNOSIS:** Congenital Hyperinsulinism.

**QUICK INFORMATION:**

Lucas has a genetic disease with inapropriate 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):**

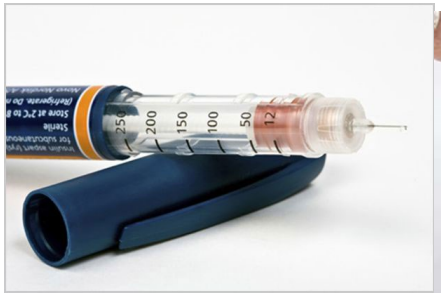
1.Give Lucas 100 cc orange juice (it´s in his bag) + 1 cookie.
2.Recheck BG in 10 minutes.
3.If Lucas is still < 70 mg/dL give to Lucas 100 cc orange juice (it´s in his bag) + 1 cookie again.
4.Recheck BG in 10-15 minutes again.
5.If Lucas is still < 70 mg/dL inject Lucas in his thigh 0,5 mg of Glucagon (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)

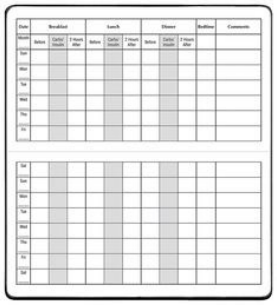
IMPORTANT: Lucas must never be left alone until hypoglycemia has resolved!!!!

## 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.



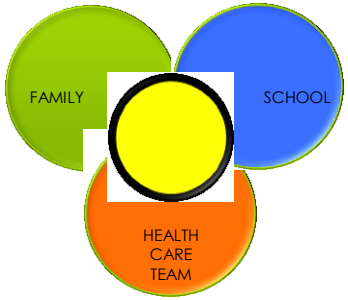
- Plan completed/signed by diabetes health care team.

# CHI AT SCHOOL

- Supplies to treat hypoglycemia
  - Source of glucose.
  - Glucagon emergency kit.

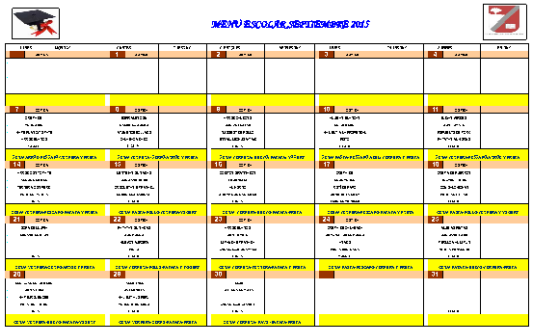

- Information about HI.

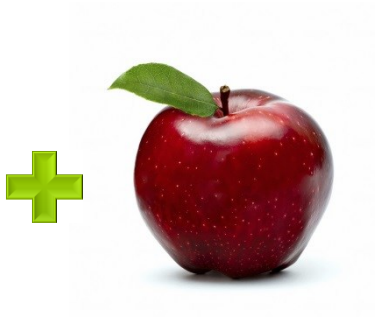

- Emergency phone numbers:
  - Parent/Guardian.
  - Diabetes health care team.



# CHI AT SCHOOL

- Student´s meal/snack schedule.
  - Closer as possible with other students´
  - Including: Parties, activities etc.



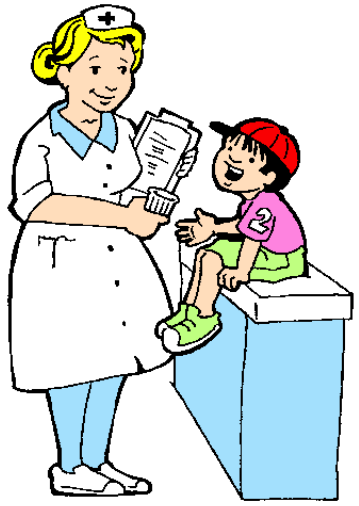


- Signed release of confidentiality from legal guardian.

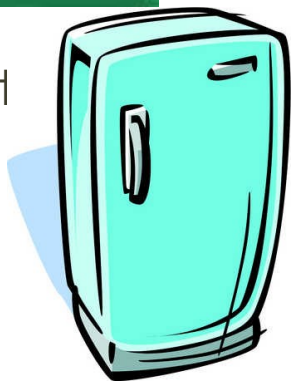
# CHI AT SCHOOL


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?.



# CHI AT SCHOOL

- Immediate accessibility of hypoglycemia´s treatment

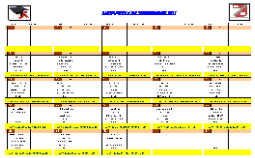
  **ALWAYS Supervision!!!!**

- Accessibility to scheduled treatments (Plan).

- School location providing privacy or classroom or anywhere:
  - Blood glucose monitoring.
  - Treatment administration.

# CHI AT SCHOOL


- 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.

# CHI AT SCHOOL


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.

# CHI AT SCHOOL


- 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.**

# CHI AT SCHOOL

## DO NOT FORGET TO EXPLAIN SCHOOL!

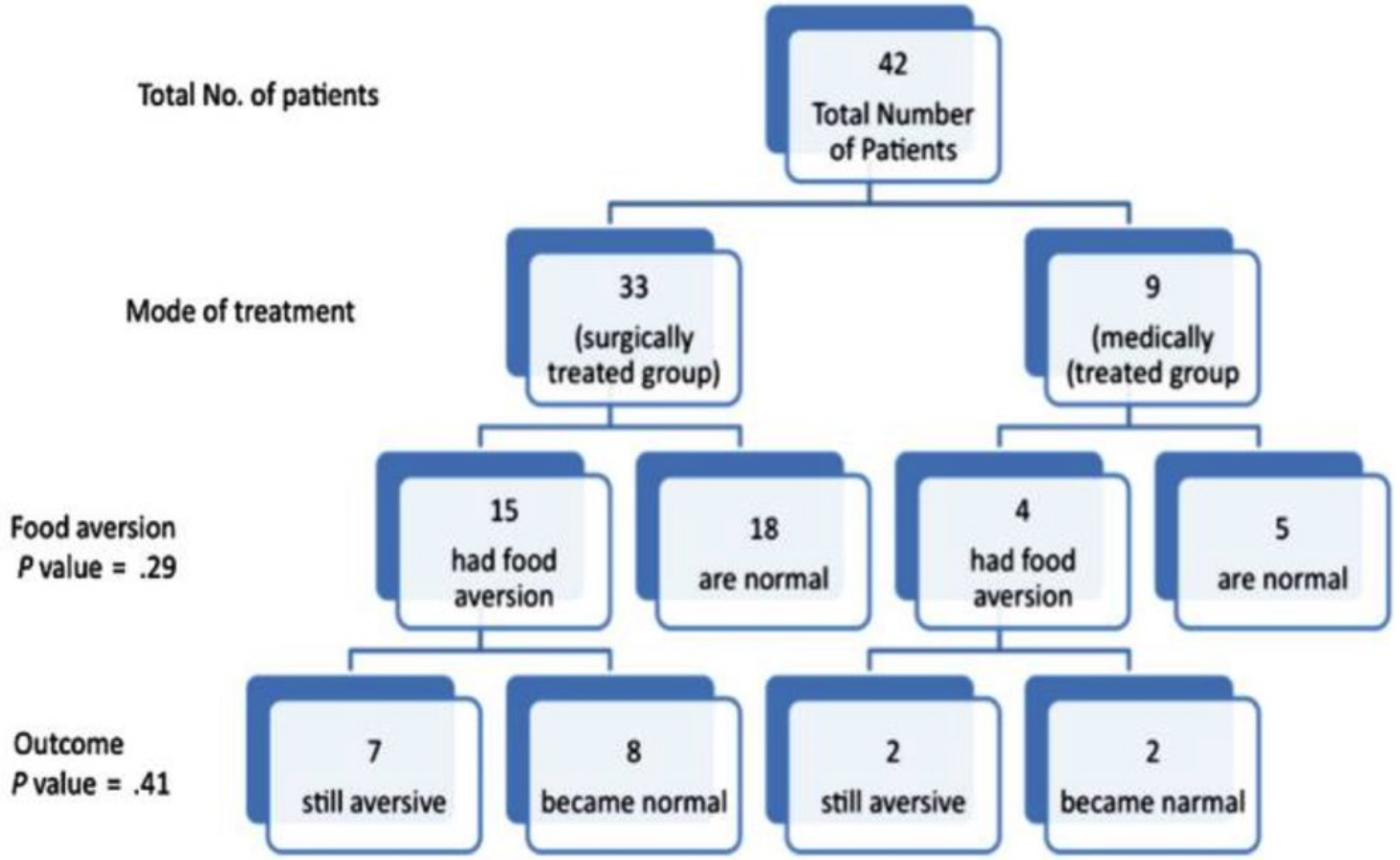
1. Feeding issues.
2. Blood glucose monitoring.
3. Neurobehavioral deficits.
4. Medications/Insulin.
5. Sports.

# CHI AT SCHOOL

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.

# CHI AT SCHOOL




## 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*

# CHI AT SCHOOL

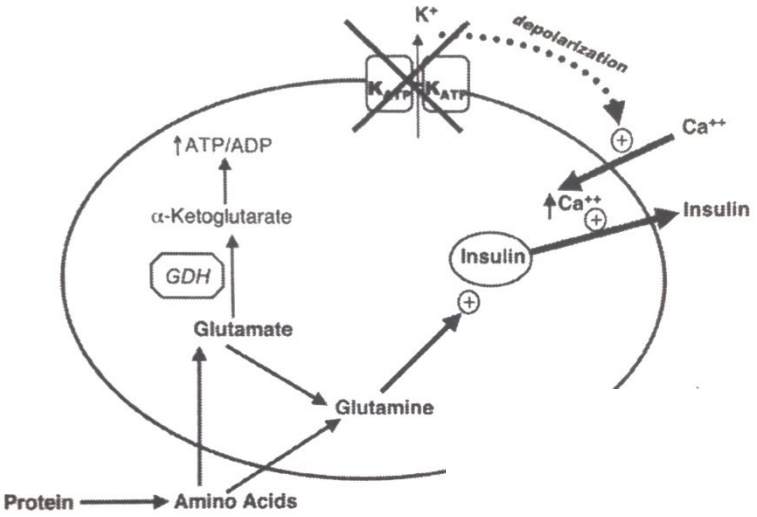
J Pediatr. 2006 Jul;149(1):47-52.

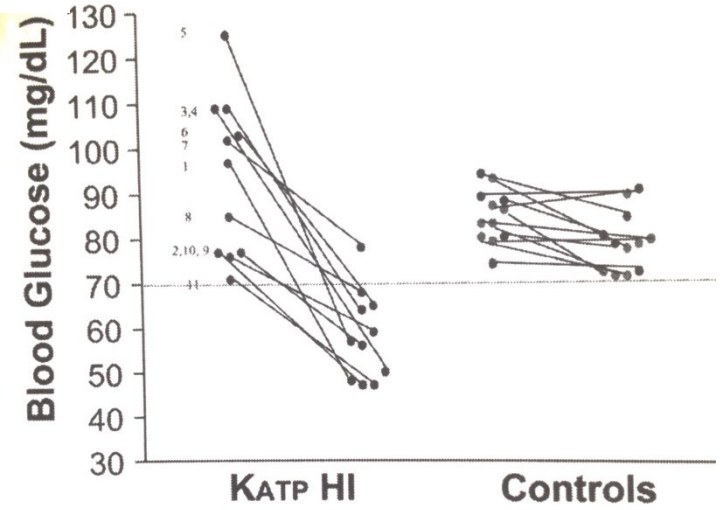
**Protein-sensitive hypoglycemia without leucine sensitivity in hyperinsulinism caused by K(ATP) channel mutations.**

Fourtner SH[1], Stanley CA, Kelly A.

- 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).

# CHI AT SCHOOL



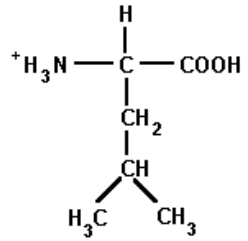

**Responses to oral protein tolerance test in patients with recessive $K_{ATP}$ hyperinsulinism**

| Patient | Baseline BG (mg/dL) | Nadir BG (mg/dL) | Delta BG (mg/dL) | Time to nadir (min) | Delta insulin (μU/ml) |
|---|---|---|---|---|---|
| 1 | 97 | 48 | −49 | 120 | 49 |
| 2 | 109 | 65 | −44 | 60 | 26 |
| 3 | 77 | 57 | −20 | 60 | 82 |
| 4 | 109 | 50 | −59 | 60 | 72 |
| 5 | 125 | 56 | −69 | 45 | 140 |
| 6 | 103 | 64 | −39 | 45 | 0 |
| 7 | 102 | 78 | −24 | 45 | 37 |
| 8 | 85 | 68 | −17 | 150 | 0 |
| 9 | 77 | 47 | −30 | 30 | 29 |
| 10 | 76 | 59 | −17 | 180 | 6 |
| 11 | 71 | 47 | −24 | 180 | ND |
| Mean ± SD | 94 ± 17 | 58 ± 10† | −36 ± 18† | 80 ± 50 | 44 ± 44* |
| GDH-HI‡ (n = 12) | 74 ± 14 (60–106) | 50 ± 9† (35–72) | −23 ± 16* (−9 to −62) | 108 ± 66 (28–180) | 14 ± 11 (1–30) |
| Control subjects (n = 12) | 85 ± 7 (74–94) | 79 ± 7 (71–90) | −6 ± 6 (−13 to 2) | 70 ± 60 (15–180) | 10 ± 8 (0–27) |

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

# CHI AT SCHOOL

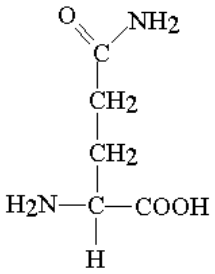
- Leucine



Leucine

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

- Glutamine:



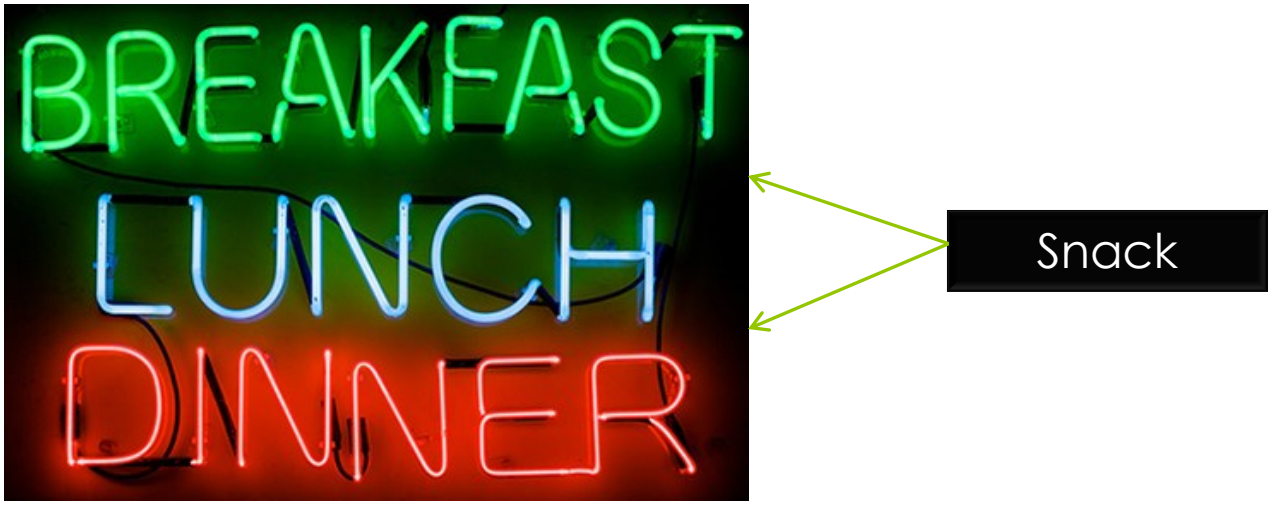
glutamine

  - Dairy products.
  - Raw meats.
  - Salmon.
  - Eggs.
  - Miso, soy.
  - Cabbage.

# CHI AT SCHOOL

- **REGULAR PATTERN**:

  Eat 3 meals and snacks a day--every day.




Same ammount at the same time every day

Avoid skipping meals.

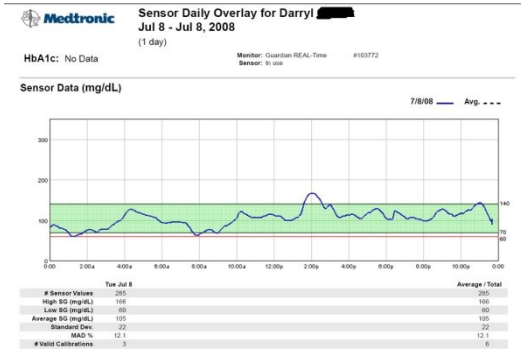
# CHI AT SCHOOL

**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?

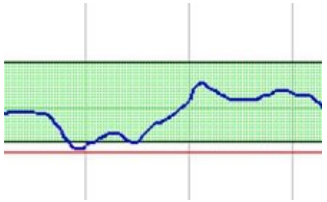

- CGMS:

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

  Useful information about **trends.**

ALWAYS confirm with CBG

# CHI AT SCHOOL



## 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.

# CHI AT SCHOOL

J Clin Res Pediatr Endocrinol. 2015 Jun 5;7(2):151-4. doi: 10.4274/jcrpe.1978.

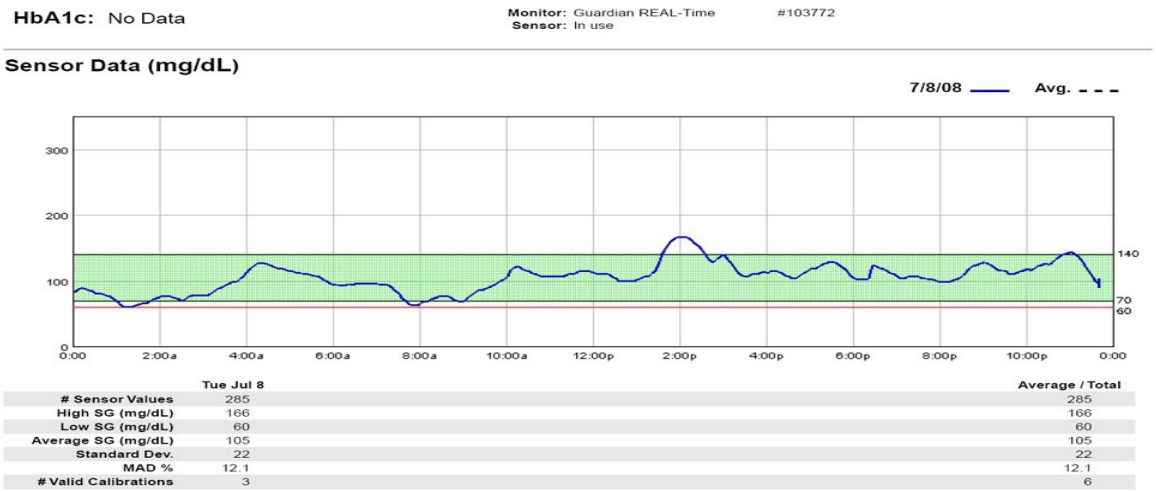
## Nifedipine in Congenital Hyperinsulinism-A Case Report.

Khawash P[1], Hussain K, Flanagan SE, Chatterjee S, Basak D.

Author information

**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.




| | Tue Jul 8 | Average / Total |
|---|---|---|
| # Sensor Values | 285 | 285 |
| High SG (mg/dL) | 166 | 166 |
| Low SG (mg/dL) | 60 | 60 |
| Average SG (mg/dL) | 105 | 105 |
| Standard Dev. | 22 | 22 |
| MAD % | 12.1 | 12.1 |
| # Valid Calibrations | 3 | 6 |

# CHI AT SCHOOL

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%.

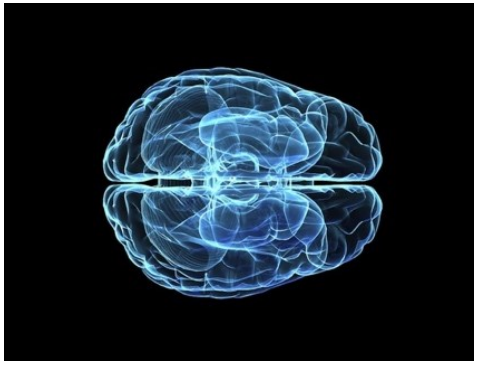
# CHI AT SCHOOL

Front Endocrinol (Lausanne). 2013 May 20;4:60. doi: 10.3389/fendo.2013.00060. eCollection 2013.

**Abnormal Neurodevelopmental Outcomes are Common in Children with Transient Congenital Hyperinsulinism.**

**Avatapalle** HB[1], Banerjee I, Shah S, Pryce M, Nicholson J, Rigby L, Caine L, Didi M, Skae M, Ehtisham S, Patel L, Padidela R, Cosgrove KE, Dunne MJ, Clayton PE.

- 39 % (Avatapalle et al., 2013).
  - Speech, language, motor and vision.
  - TCHI and PCHI: Similar incidence.
  - Risk factors:
    - Early presentation (neonatal).
    - Severe CHI.



# CHI AT SCHOOL

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**[1,2], Radcliffe J[2], Gallagher PR[3], Adzick NS[4], Stanley CA[1,2], De León DD[1,2].

**Table 3.** Prevalence of Reported Neurobehavioral Abnormalities

| Psychiatric/Behavioral | 25 (21%) |
|---|---|
| Speech delay | 22 (18%) |
| Learning disability | 19 (16%) |
| Seizures | 16 (13%) |
| Physical disability | 13 (11%) |
| ADHD | 12 (10%) |
| Autism | 2 (2%) |
| **Total** | **58 (48%)** |


**Table 4.** Neurobehavioral Measures

| Adaptive Behavior Assessment System – II* ($n$ = 69) | Mean ± SD | % < 1 SD | % < 2 SD |
|---|---|---|---|
| General adaptive composite score | 96 ± 25 | 27.5[γ] | 18.8[β] |
| Conceptual composite score | 98 ± 22 | 21.2 | 11.8[β] |
| Social composite score | 100 ± 21 | 22.1 | 14.7[β] |
| Practical composite score | 92 ± 25 | 30.9[β] | 16.2[β] |
| **Child Behavior Checklist# ($n$ = 62)** | Mean ± SD | % > 1 SD | % > 2 SD |
| Total problems | 49 ± 16 | 16.1 | 8.1[γ] |
| Internalizing problems | 49 ± 13 | 16.1 | 9.7[β] |
| Externalizing problems | 47 ± 11 | 11.5 | 6.5[α] |

* 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

[α]$P \leq 0.02$; [γ]$P \leq 0.01$; [β]$P \leq 0.001$ compared to normal population.

# CHI AT SCHOOL

**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.**

# CHI AT SCHOOL

**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[1,2], Radcliffe J[2], Gallagher PR[3], Adzick NS[4], Stanley CA[1,2], De León DD[1,2].

# CHI AT SCHOOL


## Explain your child´s HI treatments:

### Diazoxide:

- Dose: 5-15 mg/kg/day by mouth
- Side effects:
  - Excessive body hair
  - Suppression of appetite

**Timetable**

Calendarpedia
Your source for calendars

Name:

| Time / period | Monday | Tuesday | Wednesday | Thursday | Friday |
|---|---|---|---|---|---|
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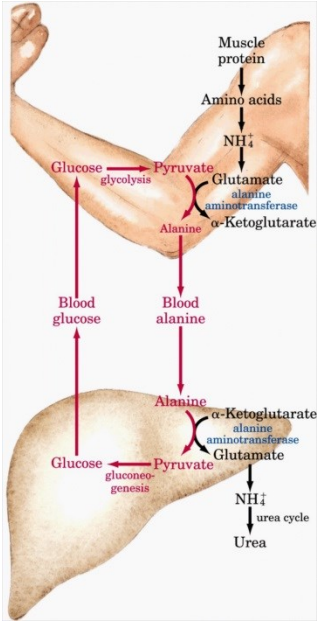
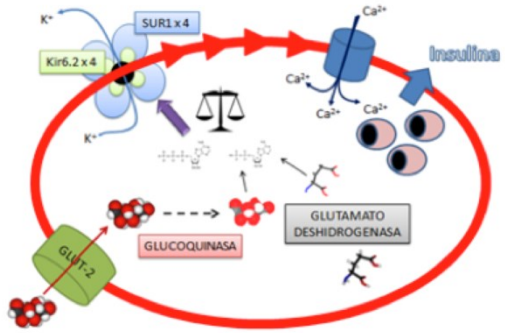
6. **SPORTS:**
    a. No HI, no diabetes.
    b. HI without diabetes.
    c. HI with subsequent diabetes.

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.



# CHI AT SCHOOL

a. **HI without diabetes:**
   - Insulin is not inhibited (medications).
   - Insulin modulates glyconeogenesis.
   - Consequence: **More prone to hypoglycemia.**



CÉLULA β

b. **HI with subsequent diabetes:**

2 options:

   - Exercise + insulin excess: **Hypo possible.**
   - Exercise + insulin deficit: **Liver effect. Hyper.**

# CHI AT SCHOOL

a. **HI without diabetes:**
   - **More prone to hypoglycemia.**
   - Strict control/medications/meals & snacks.

Timetable

Name:

| Time / period | Monday | Tuesday | Wednesday | Thursday | Friday |
|---|---|---|---|---|---|
| | | | | | |
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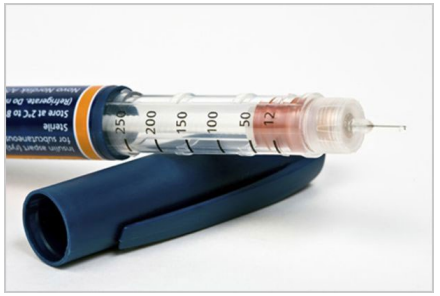

b. **HI with subsequent diabetes:**
   - Lower insulin dose but not avoid it.
   - BG (instructions in DMMT):
     - <100: 5-15 g HC and delay exercise 10-15 minutes.
     - 100-250: Start exercise normally.
     - >250: Check quetones: >1: AAR+ HC.

# Meals & Snacks

| | Low Intensity | Medium Intensity | High Intensity |
|---|---|---|---|
| <30 min | No | No | 20 g |
| 30-60 min | 10-20 g | 30 g | 50 g |
| > 60 min | 15-25 g / h | 20-70 g / h | 30-100 g / h |

**Moderate or low** glycemic index CH.

**High** glycemic index:

- During exercise.
- Prior if BG <100-130.
- After if < 100.

# CHI AT SCHOOL

## Exercise type:

- ✓ **Cardiovascullar 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% maximmum heart rate. Walking, swimming. Low hypoglycemic effect.
- ✓ Medium Intensity: 60-75%
- ✓ **High Intensity:** > 75%. **High hypo effect** but high ammounts 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!