


The HI Global Registry:

Shared Experiences for Improved Care & Treatments for Congenital Hyperinsulinism



Lauren Lopez, Ph.D.
Registry Director
Congenital Hyperinsulinism International



**HI GLOBAL
REGISTRY**

A Future Without Lows

The HI community needs:

Improved understanding of HI

Better treatment options

Clear clinical care guidelines

Improved recognition & faster diagnosis

How can we get there?



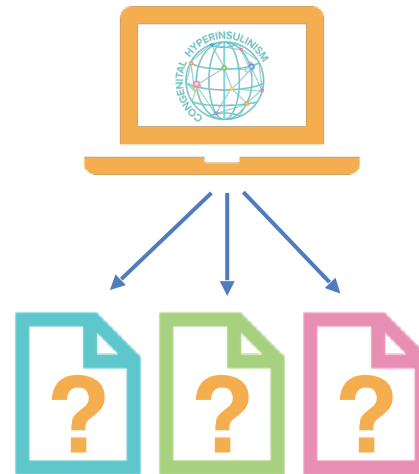
Participate in research!



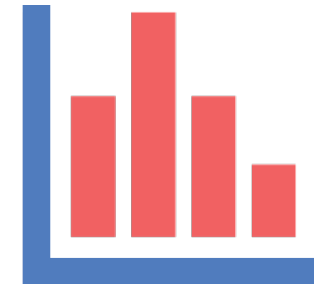
What is the HI Global Registry?



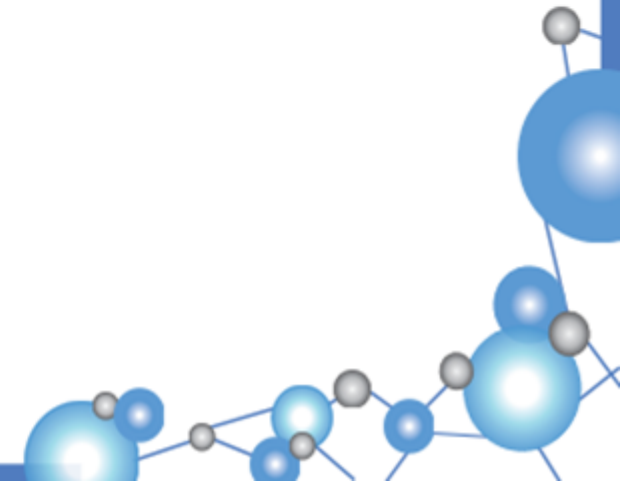
HIGR is the only patient-powered registry for hyperinsulinism research



Surveys that people with HI or their caregivers complete online from home



Qualified researchers can request the data for studies to learn more about HI, develop new treatments, and improve care



Who can join HIGR?

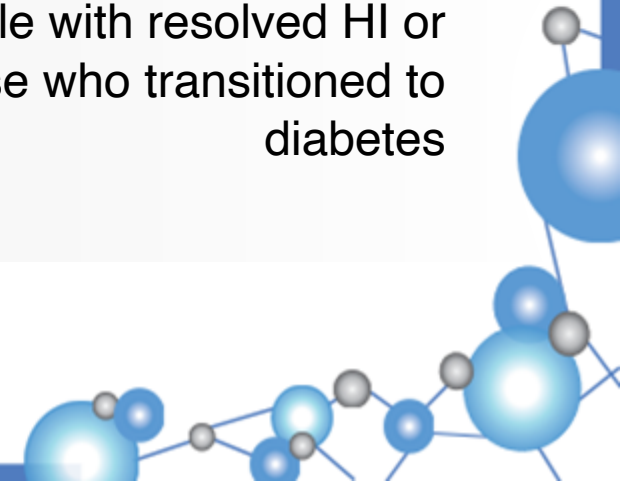
People with HI who are over
the age of 18



Caregivers of people with HI
who are under 18 or do not
have the capacity to consent to
research

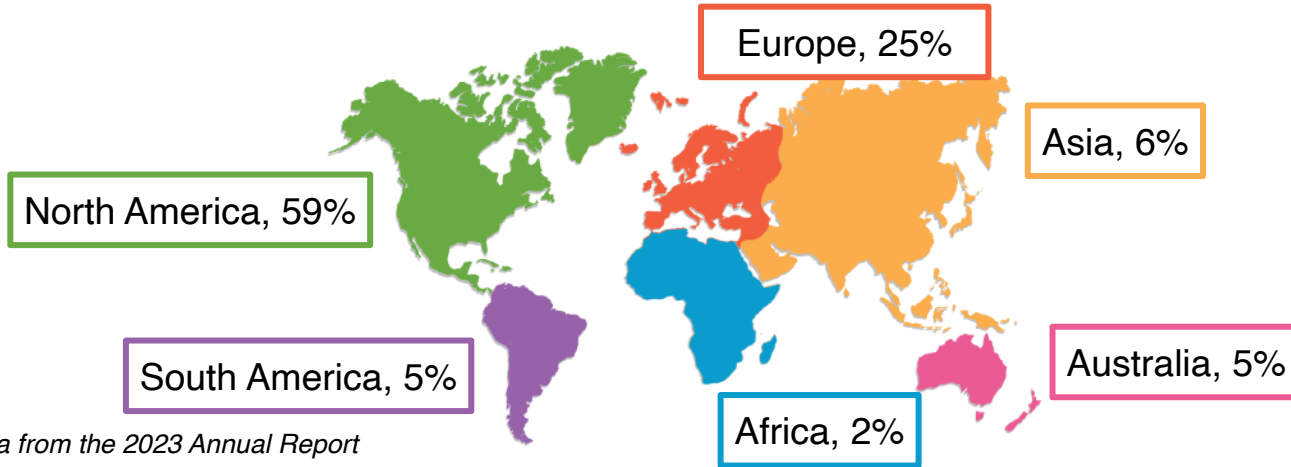
Additional caregivers of HIGR
participants
** Caregiver surveys only*

People with resolved HI or
those who transitioned to
diabetes



A truly global community

Over 500 participants across 57 countries enrolled in the HI Global Registry



Data from the 2023 Annual Report



Welcome!
환영합니다! Bienvenue!
Bienvenue!
Bem-vindo!
Benvenuti!
Willkommen!
¡Bienvenido!

higlobalregistry.com



Have you ever asked....

Is the use of emergency/rescue glucagon routine for other HI families?



At what age are children typically diagnosed with HI?



How often do babies with HI need to feed?



How often do most parents check their child's blood sugar levels?



Are seizures common with HI?



Do families receive support when kids with HI start school?

How many people become diabetic after having subtotal pancreatectomy for diffuse HI?



Do kids with HI usually have G-tubes?



HIGR Surveys



Contact & Demographics



MaxHIGR



Pregnancy



Birth



Diagnosis



Other Medical Conditions



Development



Medical Management



Surgical Management



Diet & Feeding



Glucose Monitoring



Quality of Life - Caregiver



Quality of Life - Participant

NEW surveys coming soon!



Provides permission for CHI to contact physician



CHI asks physician to complete MaxHIGR Physician Form

Physician-reported data as a complement to patient-reported data

Verify patient-reported information

Fill in any gaps in information

Provides opportunity for conversation

Survey Completion

Missing Questions

Original: Questions could be skipped

New: All questions require a response

Unknown

Prefer not to answer



ACTION: Log in to HIGR & fill in the blanks!

Survey Updates

Complex clinical management means things change

New / stop medication

Change in dosage

New medical diagnosis

Resolved feeding

problem



ACTION: Log in to HIGR & update surveys!

100% Survey Completion

Complete surveys at your own pace

Don't forget to go back to finish all surveys!

Maximize your contribution to HI

research



ACTION: Log in to HIGR & complete all surveys!

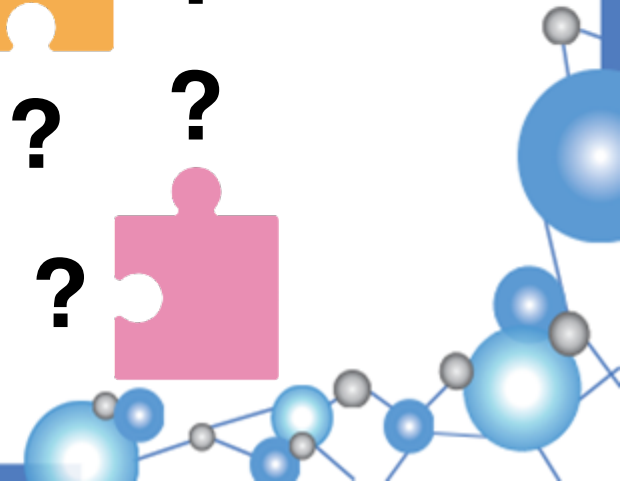
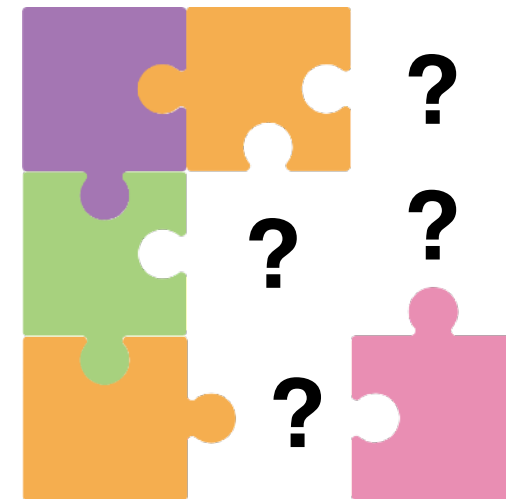
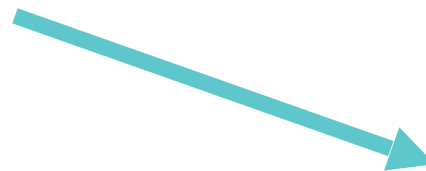
Surveys = Puzzle Pieces



All surveys completed



Missing / Incomplete surveys



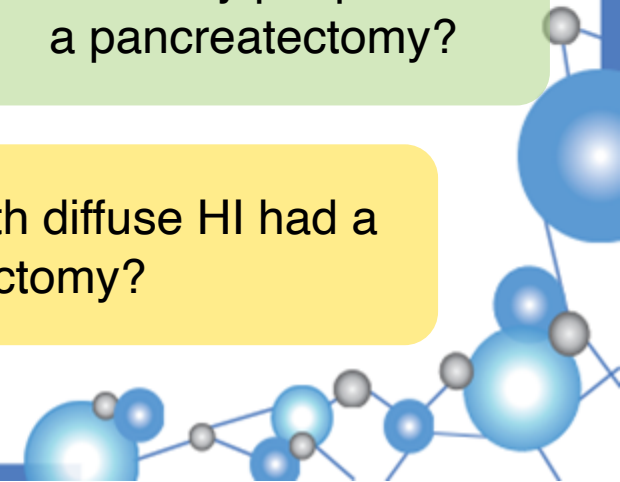
Surveys = Puzzle Pieces



How many people have diffuse HI?

How many people had a pancreatectomy?

How many people with diffuse HI had a pancreatectomy?



HIGR Data for Research Studies

HIGR Annual Report

CHI-led Research Studies

Physician-led Research Studies

Industry Data Requests



Qualified Researchers



Deidentified



Summary Data



Data Use Agreement

73%

of people living with hyperinsulinism report having feeding issues

HIGR Annual Report 2023



22%

of people with HI report having hypoglycemia more than once a day

HIGR Annual Report 2023



98%

of people taking diazoxide* report at least one side effect

HIGR Annual Report 2023

*the most common medicine used to treat HI



41%

of families are struggling to pay for the costs associated with hyperinsulinism

HIGR Annual Report 2023



HIGR Contributions to Research



International Guidelines

Research Papers

Conference Posters

Hormone Research in Paediatrics
Consensus Statement
Horm Res Paediatr
DOI: 10.1159/000531766

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International Guidelines for the Diagnosis and Management of Hyperinsulinism

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Ignacio Bergada^d Tricia Bhatti^e Louise S. Conwell^f Junfen Fu^g
Sarah E. Flanagan^h David Gillisⁱ Thomas Meissner^j Klaus Mohnike^k
Tai L.S. Pasquini^l Pratik Shah^m Charles A. Stanleyⁿ Adrian Vellaⁿ
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Keywords: Hyperinsulinism · Guidelines · Hypoglycemia · Insulin

Abstract: Background: Hyperinsulinism (HI) due to dysregulation of pancreatic beta-cell insulin secretion is the most common and most severe cause of persistent hypoglycemia in infants

Keywords: Hyperinsulinism · Guidelines · Hypoglycemia · Insulin

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Banerjee et al.
Orphanet Journal of Rare Diseases
DOI: 10.1186/s13023-022-02174-y

REVIEW
Open Access

Congenital and childhood hypoglycemia and the pediatric population

Indraneel Banerjee¹, Merete Hammer², David ...

Abstract: Background: Congenital hypoglycemia is a rare condition in children, and carries a complex and often delayed diagnosis. Despite not significantly improving psychosocial and financial monitoring throughout the life cycle. **Results:** In this review, we explore the current diagnostic and management of patients with congenital hypoglycemia and the impact of specialist centers. **Conclusions:** To ensure early diagnosis and appropriate management, a network of specialist centers of excellence for children is essential. Awareness of the condition and its management by pediatricians and other healthcare professionals is essential to ensure the best outcomes for these children, even those who are not yet diagnosed.

Keywords: Congenital hypoglycemia, childhood hypoglycemia, pediatric hypoglycemia

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Rastkin et al.
Orphanet Journal of Rare Diseases
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REVIEW
Open Access

Congenital Hyperinsulinism International: A Community Focused on Improving the Lives of People Living With Congenital Hyperinsulinism

Jake Rastkin¹, Tai L.S. Pasquini², Sheila Bose, Dina Talis and Jennifer Schmitt

Abstract: Congenital hyperinsulinism (HI) is a rare disease affecting newborns. HI causes severe hypoglycemia due to the overproduction of insulin. The signs and symptoms of hyperinsulinism in HI babies is often not discovered until brain damage has already occurred. Prolonged hypoglycemia from HI can even lead to death. Disease management is often complex with a high burden on caregivers. Treatment options are extremely limited and often require long hospital stays to devise. Cascading from suboptimal treatments and diagnostic practices are a host of other problems and challenges that many with HI and their families experience including continued fear of hypoglycemia and feeding problems. The aim of this paper is (1) to describe the current challenges of living with HI including diagnosis and disease management, (2) from the perspective of people who live with the condition (3), to provide family stories of living with HI, and (4) to share how a rare disease patient organization, Congenital Hyperinsulinism International (CHI) is working to improve the lives of HI patients and their families. CHI is a United States based nonprofit organization with a global focus. The paper communicates the progress the patient advocacy organization has put into place to support HI families through its virtual and in-person gatherings. The organization also helps individuals access diagnosis, medical experts, and treatments. CHI also raises awareness of HI to improve patient outcomes with information about HI and prolonged hypoglycemia in twenty three languages. CHI also shows innovation for new and better treatments by funding research pilot grants, conducting research through the HI Global Registry, and providing patient experience expertise to researchers developing new treatments. The organization is also the sponsor of the CHI Collaborative Research Network which brings medical and scientific experts together for the development of a patient-focused prioritized research agenda.

Keywords: congenital hyperinsulinism, hypoglycemia, rare disease, burden of disease, caregivers, patient organization

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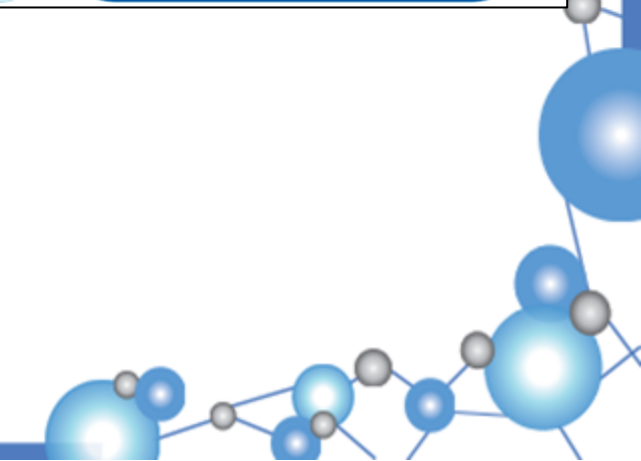
Neurologic Outcomes of HI Global Registry Participants

Tai L.S. Pasquini¹, Paul S. Thornton², Divya D. De Leon³, Merete Hammer⁴, Junfen Fu⁵, Sarah E. Flanagan⁶, David Gillis⁷, Thomas Meissner⁸, Klaus Mohnike⁹, Pratik Shah¹⁰, Charles A. Stanley¹¹, Adrian Vella¹², Tohru Yorifuji¹³, Paul S. Thornton¹⁴

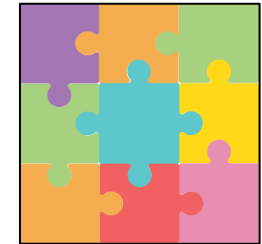
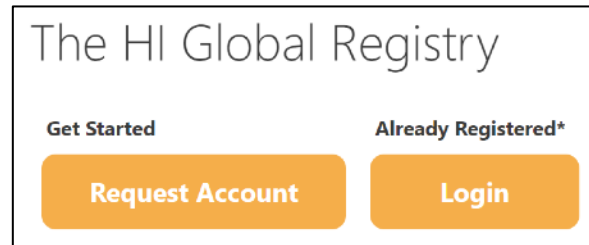
Abstract: The aim of this study was to determine the patient journey to an HI diagnosis for infants who were also diagnosed with a neurologic condition. **Methods:** Retrospective review of the patient and caregiver response data from surveys in HI Global Registry from October 2018 to March 2023. We evaluated the time to an HI diagnosis and actions based on the survey data for individuals who were later also diagnosed with a neurologic condition. All families were categorized as HI+ or HI- based on survey data. **Results:** Of 105 participants in HI Global Registry, 17.2% (n=18) reported that they had received one or more neurologic diagnoses. Of 95 HI+ participants, 22.0% (n=21) reported that they had received one or more neurologic diagnoses. Of 87 HI- participants, 0% (n=0) reported that they had received one or more neurologic diagnoses. **Conclusions:** The early identification of HI and timely appropriate care is critical to prevent brain damage and subsequent neurologic sequelae. The administration of glucose or diazoxide following an initial abnormal blood glucose value did not prevent subsequent neurologic damage in a large subset of HI Global Registry participants. Management must be proactive and not reactive to symptoms that have been established. This highlights the importance of following HI guidelines for care and management and the need for better treatments. Limitations include a risk of selection and recall bias and incomplete information, as individuals can choose which questions to answer.

Keywords: congenital hyperinsulinism, hypoglycemia, rare disease, burden of disease, caregivers, patient organization

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How to Join the HIGR Community

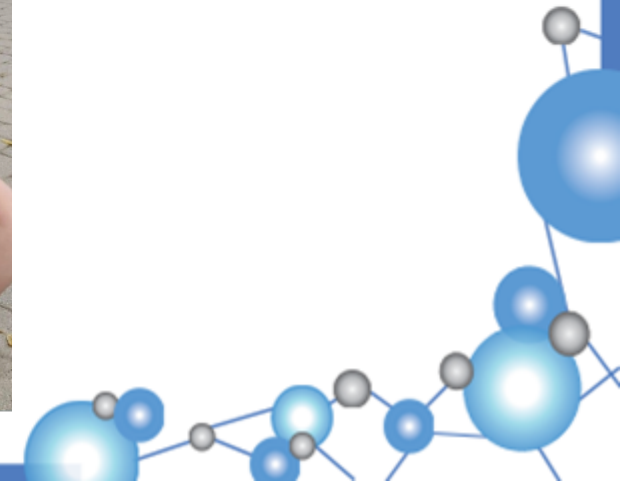
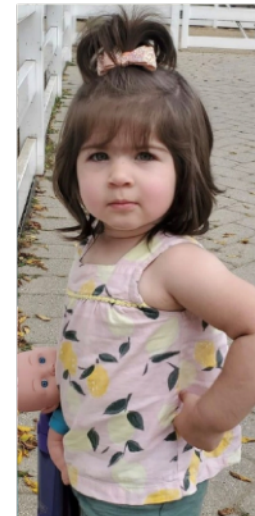


1. Visit higlobalregistry.org


2. Click “Request Account”

3. Complete the registration forms

4. Complete all surveys!



Helpful Resources



DONATE TODAY!

CHI Congenital Hyperinsulinism International
When too little sugar hurts, we help.™

About Us ▾ Awareness & Resources ▾ Research ▾

Support & Clinical Care ▾ Events Donate Blog Contact ▾

Search ... 🔍

The HI Global Registry

Get Started **Already Registered***

[Request Account](#) [Login](#)

*New password required: If you have previously registered to participate in HIGR but have not logged in since before November 10, 2023, please click the Login button and click on the link to "Reset your password"

What is the HI Global Registry?

The HI Global Registry (HIGR) is the only patient-powered hyperinsulinism (HI) registry. A patient registry is an organized way of collecting information about a group of individuals with the same or related diseases. The HI Global Registry contains a series of surveys designed to capture information about various aspects of being diagnosed with and living with hyperinsulinism.

HIGR launched in 2018 and consists of thirteen surveys that record various aspects of the patient and caregiver experience with HI over their lifetime. Additionally, MaxHIGR provides complementary data from the physician perspective. These insights are critical to the entire HI community and become even more meaningful as more individuals participate. Survey responses are made anonymous/deidentified and shared with researchers to improve the overall understanding of HI to help advance new treatments, to improve patient care, and ultimately, to bring the lived experience of HI to the forefront of research.

Since its launch in 2018, data from the HI Global Registry has been quoted in or informed:

- The International Hyperinsulinism Clinical Care Guidelines
- Clinical Trial protocol design and inclusion/ exclusion criteria for studies
- Four peer-reviewed journal articles
- Five medical conference poster presentations



QUICK LINKS

- [Frequently Asked Questions](#)
- [Guides & Tutorials](#)
- [Annual Reports & Publications](#)
- [Steering Committee](#)
- [Registry Objectives](#)
- [Community](#)

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Frequently Asked Questions



Video Tutorials



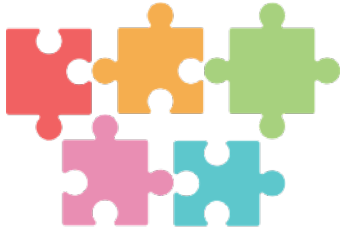
PDF Help Guides with screenshots

What's Next?



HI GLOBAL
REGISTRY

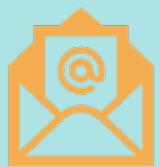
Join the HI Global Registry!



Complete / Update all Surveys



Dexcom Volunteers?



info@higlobalregistry.org

**Your
experience
with
hyperinsulinism
matters.**



Join HIGR today and complete surveys in English, Spanish, Italian, French, Portuguese, German, or Korean.

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