

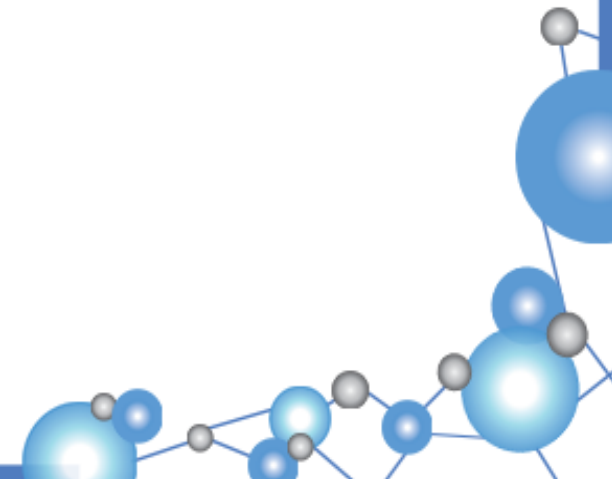
The HI Global Registry



HI GLOBAL
REGISTRY

The HI Global Registry

- The HI Global Registry (HIGR) launched in 2018
- Provides a baseline or an overall understanding of the reality of the day-to-day experience of living with HI
- Helps identify the most pressing issues and on-going challenges
- HIGR invites people worldwide to participate, increasing the pool of knowledge about the condition



Who can join the HI Global Registry?

- People with HI or their caregiver can share their experiences by answering surveys through an online platform
- All information collected is kept on a secure system
- Questions were developed by HI patients, clinicians, and researchers

Parent of a child
living with HI



Respondent

Participant

Adult living with HI



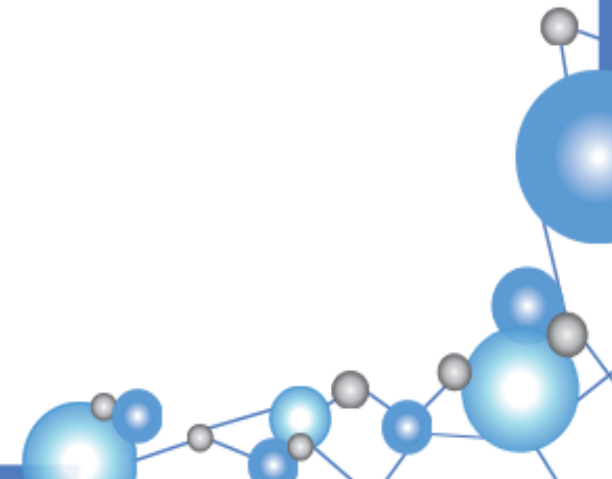
Participant and respondent



Experiences of Interest

- ALL!
 - positive/negative genetics,
 - diffuse/focal,
 - on treatment/ no longer on treatment,
 - surgery/no surgery,
 - daily lows/ no longer experiencing lows, etc.
- EVERY story is an important piece of the constellation!

53 countries represented
in HIGR



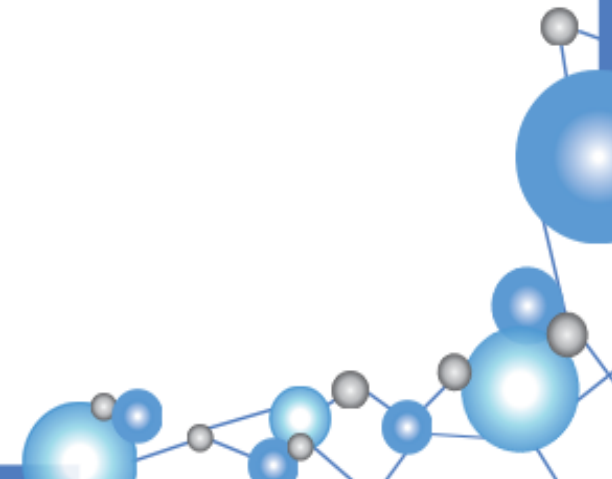
Benefits of Participating

- Research increases our knowledge of HI and potentially leads to:
 - More timely diagnosis
 - New and better treatments
 - Fewer undesirable effects
- Receive information about medical advances and other news
- Personal satisfaction of being a member of the research team and process



Risks of Participating (Minimal)

- We follow the best practices of data privacy, security, and all applicable laws (HIPPA, GDPR)
- Questions can be sensitive, and you may feel uncomfortable
 - You do not have to share information that you do not want to
- Risk that the participant's information could be misused, but the chance of this happening is very small
 - We have protections in place to lower this risk!
- Respondents may withdraw consent at any time



Surveys

Submit once

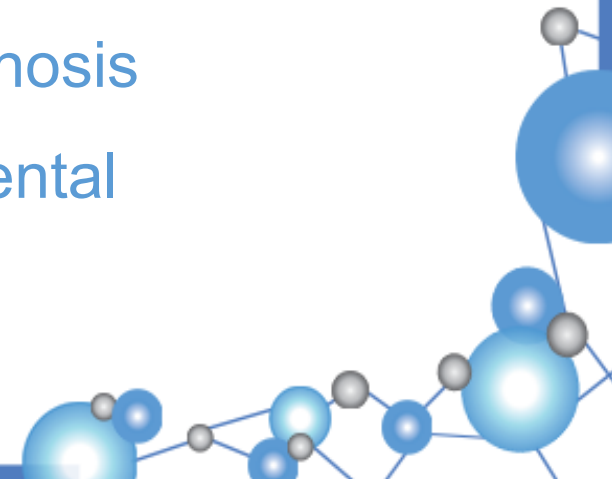
- Pregnancy
- Birth

Longitudinal

- Glucose monitoring - 6 months
- Quality of life (Parent/ LAR) - Annual
- Quality of life (Participant) - Annual
-

Updatable

- Contact information
- Demographics
- Diagnosis
- Medication management
- Diet & feeding management
- Surgical management
- Other diagnosis
- Developmental



MaxHIGR- Physician Contributed Information

- 1st Step Survey in your portal

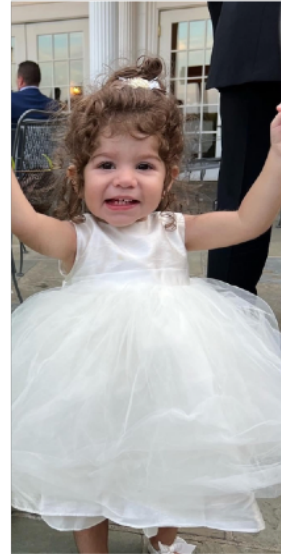
Patient Initials: _____ Max HIGR Form Today's Date: _____

Please complete the form to the best of your ability with applicable patient information:

Country	<input type="checkbox"/> Australia <input type="checkbox"/> Denmark <input type="checkbox"/> Germany <input type="checkbox"/> Kazakhstan <input type="checkbox"/> UK <input type="checkbox"/> US <input type="checkbox"/> Other (please list): _____
HI center	<input type="checkbox"/> CHOP <input type="checkbox"/> COOK <input type="checkbox"/> GOSH <input type="checkbox"/> NORCHI <input type="checkbox"/> NUSOM <input type="checkbox"/> Other
Other HI center	_____
Current age of participant	Months _____ Years _____
Diagnosis type	<input type="checkbox"/> Focal <input type="checkbox"/> Diffuse <input type="checkbox"/> Atypical <input type="checkbox"/> Other
Other diagnosis type	_____
Gestation at birth	Days _____ Weeks _____
Method of delivery	<input type="checkbox"/> Normal <input type="checkbox"/> C-section <input type="checkbox"/> Forceps
5 min Apgar score	<input type="checkbox"/> Yes <input type="checkbox"/> No Apgar score 1-10: _____
Birth weight	lbs _____ oz _____ or gm _____
History of neonatal hypoglycemia	<input type="checkbox"/> Yes <input type="checkbox"/> No
Symptoms of neonatal hypoglycemia	<input type="checkbox"/> No obvious signs <input type="checkbox"/> Excess jittery <input type="checkbox"/> Seizures <input type="checkbox"/> Unresponsive <input type="checkbox"/> Other
Other symptoms	_____
Age at presentation of hypoglycemia	Days _____ Weeks _____ Months _____
Age at diagnosis of hyperinsulinism	Days _____ Weeks _____ Months _____
Please provide the basis for diagnosis	Plasma Glucose: mg/dL _____ or mmol/L _____
Please provide the basis for diagnosis	Insulin levels: pmol/L _____ or mU/L _____
Low Betahydroxybutyrate	<input type="checkbox"/> Yes <input type="checkbox"/> No Betahydroxybutyrate Levels (mmol/L): _____
Confirmatory glucagon test	<input type="checkbox"/> Yes <input type="checkbox"/> No
Syndromes	<input type="checkbox"/> Beckwith-Wiedemann <input type="checkbox"/> Rubinstein Taybi <input type="checkbox"/> Kabuki <input type="checkbox"/> Turner <input type="checkbox"/> Sotos <input type="checkbox"/> Costello <input type="checkbox"/> Fanconi <input type="checkbox"/> Other
Other syndromes	_____
Other co-morbidities at presentation	<input type="checkbox"/> Yes <input type="checkbox"/> No
Describe co-morbidities	_____
Diabetes	<input type="checkbox"/> Yes <input type="checkbox"/> No
Describe Diabetes	_____

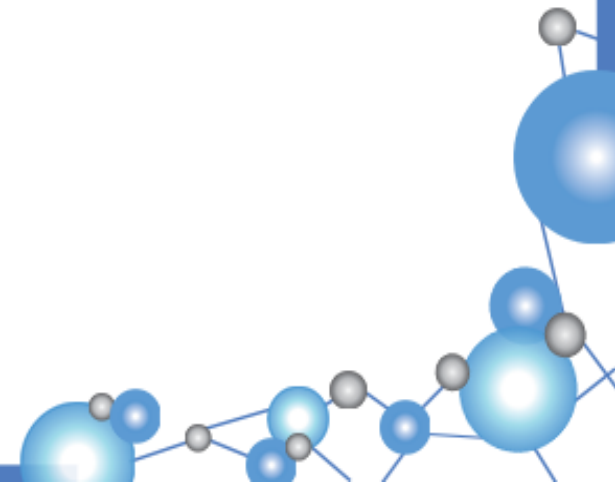
Importance of completing all surveys

- Complete data!
- Even if an experience doesn't apply, we can't make assumptions



Exciting new features!

- Multiple languages (Coming Soon) :
 - English
 - German
 - Spanish
 - Italian
 - Portugese
 - French
 - Korean
 - Japanese
 - Hebrew
- CGM and Glucometer data
- New validated surveys, including QOL
- Ability for multiple people to contribute data



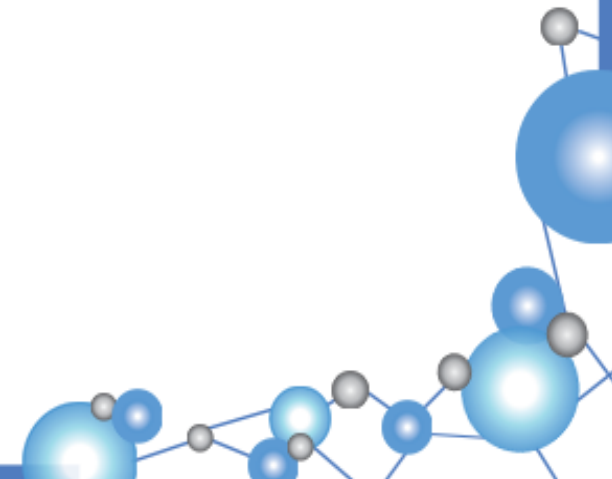
Case Study: Increasing Accuracy

Has the participant experienced any feeding issues on a regular basis (check all that apply)?

	Feeding Issues	No Feeding Issues
Responses	0 (0%)	2 (100%)

#1:
No feeding
issues

#2:
No feeding
issues



Case Study: Feeding Issues

	Feeding Issues	No Feeding Issues
Responses	2 (40%)	3 (60%)

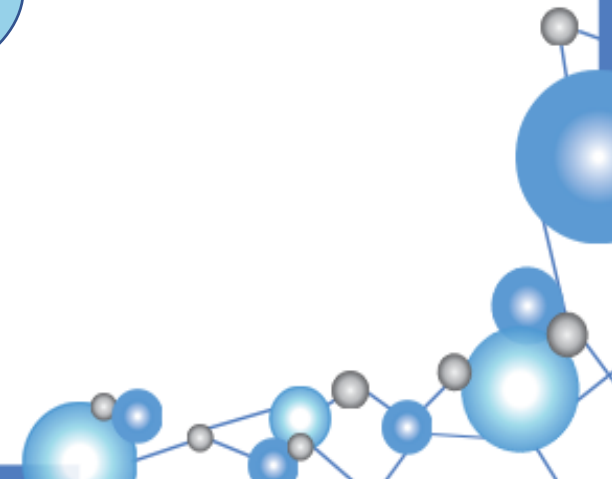
#1:
No feeding
issues

#2:
No feeding
issues

#3:
Poor
appetite

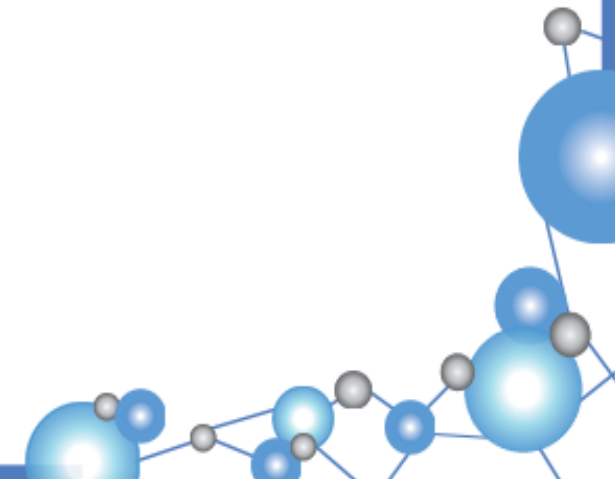
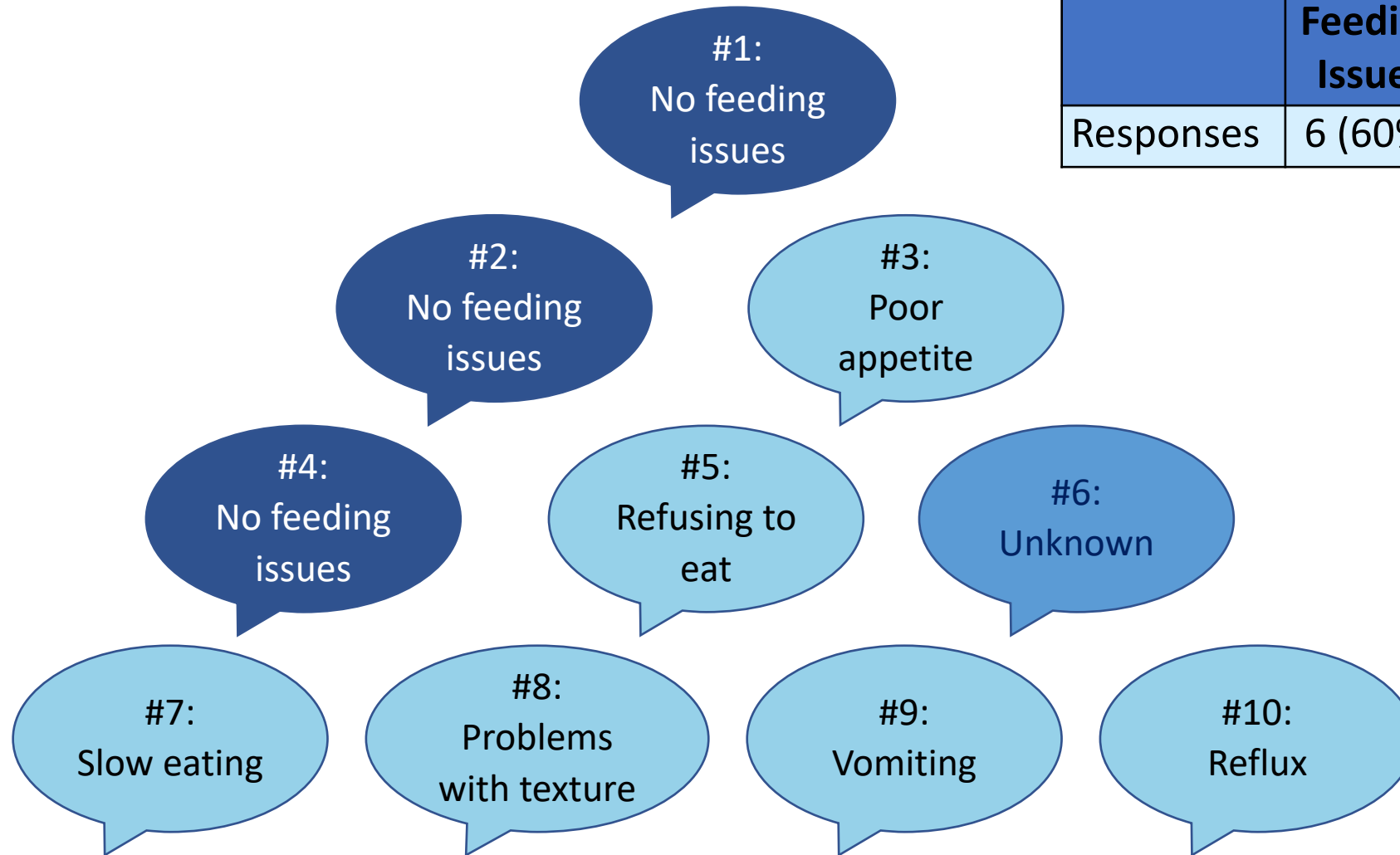
#4:
No feeding
issues

#5:
Refusing to
eat



Case Study: Feeding Issues

	Feeding Issues	No Feeding Issues	Unknown
Responses	6 (60%)	3 (30%)	1 (10%)



Feeding Issues (n=211)

