

Patient Initials:

**MaxHIGR Form**

Today's Date:

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

Patient country	Australia	Denmark	Germany	Kazhakstan	United Kingdom
	United States	Other (please list):			
HI center					
Person completing this form					
Current age of participant	Months	Years			
Diagnosis/type	Focal	Diffuse	Atypical	Other	
Other diagnosis/type					
Gestation at birth	Days	Weeks			
Method of delivery	Normal	C-section	Forceps		
5 min Apgar score	Yes	No	Apgar score 1-10:		
Birth weight	lbs	oz	or	gm	
History of neonatal hypoglycemia	Yes	No			
Symptoms of neonatal hypoglycemia	No obvious signs	Excess jittery	Seizures	Unresponsive	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	Yes	No	Betahydroxybutyrate levels (mmol/L):		
Confirmatory glucagon test	Yes	No			
Syndromes	Beckwith-Wiedemann	Rubinstein Taybi	Kabuki	Turner	
	Sotos	Costello	Fanconi	Other	
Other syndromes					
Other co-morbidities at presentation	Yes	No			
Describe co-morbidities					
Diabetes	Yes	No			
Describe diabetes					

Was genetic testing performed	Yes	No	If yes, was a positive mutation found:		Yes	No
Identified gene mutations	Unknown	ABCC8 (SUR1)	KCNJ11 (KIR6.2)			
	GLUD1 (Glutamate dehydrogenase)		GCK (Glucokinase)		HNF4A	
Other gene mutations	HNF1A	HADH	Other (please list)			
Gene inheritance	Heterozygous		Homozygous		Compound heterozygous	
Gene inheritance	Recessive		Dominant		Unknown	
Gene inheritance	Paternal		Maternal		Unknown	
Gene inheritance	Paternal		Maternal		Unknown	
Gene inheritance	Paternal		Maternal		Unknown	
Initial treatment profile	High concentration dextrose/glucose:		Yes	No	Glucagon treatment:	
Other initial treatments					Used	Not used
Treatments tried	None		Diazoxide (Proglycem, Eudamine)		Octreotide (Sandostatin)	
	Long-acting octreotide (Sandostatin LAR)		Sirolimus (Rapamune)		Lanreotide (Somatuline)	
Other treatments	Nutritional Support		Glucocorticoids		Glucagon	
			High concentration dextrose/glucose		Other	
Age at introduction of diazoxide	Days	Weeks	Months			
Diazoxide brand/formulation	Proglycem		Special formulations		Eudamine	
Other brand/formulation					Other	
Diazoxide delivery	Tablet/Capsule		Liquid/Oral suspension			
Starting dose (mg/kg/day)	5	10	15	Other		Other dose:
Discharged home on diazoxide	Yes	No		Currently using diazoxide:		Yes
Discharged home on diazoxide	Yes	No		Currently using diazoxide:		Yes
Age when stopped using diazoxide	Days	Weeks	Months			
Max dose of diazoxide ever taken (mg/kg/day)						
Side effects of diazoxide	Congestive heart failure		Fluid overload		Pulmonary hypertension	
	Abnormal blood count		Excess hair		Face changes	
Other side effects of diazoxide	Other					

Echocardiogram	Pre-diazoxide	Post-diazoxide	Both	None
Currently using octreotide	Yes	No	Age at introduction of octreotide: Weeks	Months
Dose of octreotide (mcg/kg/day)		Age when stopped using octreotide: Weeks	Months	
Max dose of octreotide ever taken (mcg/kg/day)				
Response to octreotide	Responsive	Unresponsive	Partially responsive	
Side effects while taking octreotide	Gallstones/sludge	Hepatitis	Necrotizing enterocolitis	
Other side effects of octreotide	Growth failure	Injection site problem	Hypothyroidism	Other
Currently using long acting somatostatin analogue	Yes	No	Type of long acting somatostatin analogue: Sandostatin LAR	Lanreotide
Age at introduction of long acting somatostatin analogue	Weeks	Months	Years	
Age when stopped using somatostatin	Weeks	Months	Years	
Max dose of long acting somatostatin analogue ever taken (mg)				
Interval (weeks) of long acting somatostatin analogue	3	4	5	6
				Other
Side effects while taking long acting somatostatin analogue	Gallstones/sludge	Hepatitis	Necrotizing enterocolitis	
Other side effects of long acting somatostatin analogue	Growth failure	Injection site problem	Hypothyroidism	Other
Other trial medications, delivery and dose				
Stable glucose on glucose monitoring at discharge	Yes	No		
Safe age-appropriate fasting	Yes	No	Tolerance fast (hours):	6
			12	18
Other fast period (hours)	Other			
Additional carbohydrate supplements	Tube feeding: Nasogastric (NG)/Orogastric (OG)			
	Tube feeding: Gastrostomy/G-tube/button			
	TPN (long-term intravenous [IV] feeds)			
Experiences blood glucose below 70 mg/dL (3.9 mmol/L) more than 3 times a week	Yes	No	Unknown	

18F DOPA PET scan performed	Yes	No						
18F DOPA PET scan results	Identified focal HI		Didn't identify focal		Other			
Other scan results								
Pancreatectomy	Not required		Recommended, not performed		Partial pancreatectomy			
	Subtotal pancreatectomy >90%		Focal lesionectomy		Whipple's procedure			
Age at first pancreatectomy	Days	Weeks	Months					
Additional details								
>1 pancreatic surgery	Yes	No						
Describe >1 pancreatic surgery								
Post-pancreatectomy diabetes	Yes	No	Age at development of diabetes:		Weeks	Months	Years	
Post-surgery hypoglycemia	Yes	No	N/A		Post-surgery hyperglycemia	Yes	No	N/A
Post-surgery exocrine insufficiency requiring enzyme supplements	Yes	No	N/A		Age at permanent use of enzyme supplements:	Weeks	Months	Years
Pancreas histology	Normal pancreas		Diffuse	Focal	Atypical	Insulinoma		
	Unknown		Other					
Other histology								
Feeding difficulties	Yes	No						
Describe feeding difficulties								
Neurodevelopmental abnormalities	None	Anxiety	Visual problems		Autism spectrum disorder			
	Motor delay		Speech delay		Global development delay			
	Epilepsy		Cerebral palsy		Not performing at grade level			
	Movement disorder		Specific learning difficulties		Dystonia	Other		
Other abnormalities								
Brain scan	Yes	No	Abnormality identified in brain scan:		Yes	No		
Describe brain scan abnormality								
Additional information you would like to share								