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Preventing Severe Hypoglycemia with a Continuous Glucose Monitor Device in an Infant with Glycogen Storage Disease 1a undergoing a Gastrostomy Tube Placement: Case Report

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Abstract

We describe a case in which a continuous glucose monitor (CGM) was used during hospitalization to safely prevent severe hypoglycemia in a 10-month-old male with glycogen storage disease 1a undergoing placement of a gastrostomy tube. The child was admitted to the hospital for placement of a gastrostomy tube given increased severe hypoglycemic episodes at home secondary to worsening feeding aversion. During hospitalization, the CGM was monitored before, during and after surgery. Severe hypoglycemic episodes were avoided while hospitalized. The CGM device assisted with the management and titration of continuous dextrose infusion while the child was fasting. The CGM device improved patient and family centered outcomes by reducing the need for frequent finger stick point-of-care glucose checks and allowed the family and anesthesiologist to carefully monitor the child's blood glucose continuously during the procedure.

Keywords: Hypoglycemia; Glycogen Storage Disease; Continuous Glucose Monitoring Device; Gastrostomy Tube

Introduction

Continuous glucose monitoring (CGM) systems have been shown to be safe and effective in improving glycemic control and reducing hypoglycemia in patients with type 1 and type 2 diabetes **[1].** In patients at high risk for severe hypoglycemia, such as those with a glycogen storage disease (GSD), CGMs have demonstrated good correlation between capillary blood glucose values and reducing the duration of hypoglycemia, lactic acidosis, liver size and hyperlipidemia **[2,3].** CGM devices allow for remote monitoring and reduce the need for frequent point-of-care glucose checks **[4].** Patients with GSD can develop severe hypoglycemia and worsening lactic acidosis while undergoing anesthesia **[5].** There is a paucity of information on using CGM devices in young children with GSD undergoing surgical procedures. We present a case of a child with GSD Ia whose glycemic control was successfully managed using a CGM during a high-risk prolonged fasting period for surgical gastrotomy tube placement.

Case Presentation

A 10-month-old male with glycogen storage disease 1a (GSD Ia) was admitted to the hospital for gastrostomy tube placement (G-tube) given worsening feeding aversion at home which resulted in increasing severe hypoglycemic episodes. The child had no recent illness or fevers and had otherwise been healthy leading up to the surgery. The family had been feeding him a soy-based Enfamil Prosobee formula (27kcal/oz) 120 ml every 2 hours during the day to maintain his blood glucose levels (**Figure 1**). For the past 2 weeks, the child had been refusing multiple feeds requiring glucose gel to be used as a rescue 3-4 times per day. The glucose gel (5-10 grams of dextrose) was challenging to administer as the child would refuse and most times required two caregivers to administer. Glucose gel only maintained his blood glucose levels for 30 minutes to 1 hour and required repeated dosing if the child refused to feed again. The family reported significant sleep disturbances for the child along with severe family distress.

The child was born at 38 weeks' gestation and on day 2 of life had a seizure during circumcision secondary to severe hypoglycemia. He was hospitalized for 2 weeks in the neonatal intensive care unit for management of ongoing hypoglycemia. Given the persistent hypoglycemia episodes in the NICU, a continuous glucose monitor (Free Style Libre 2) was used to monitor his glycemic trends (this was later changed to Dexcom G6 at 3 weeks of age to allow for remote monitoring). He required feedings every 2 hours to maintain adequate glucose levels. At 3-weeks of age, genetic testing revealed the diagnosis of autosomal recessive liver glycogen storage disease Ia (gene G6PC, variant c.247C>T (p. Arg83Cys)). In this disease, glycogenolysis is ineffective, and this results in severe hypoglycemia while fasting and is associated with the excessive accumulation of glycogen within the liver, kidneys, and intestinal mucosa **[6].**

The child had been developing appropriately at 10-month-old, and his growth parameters included weight 97% (11.2 kg), length 95% (76.3 cm), and head circumference 40% (45.1 cm). He was provided speech therapy weekly for his severe feeding aversion.

The child was admitted the day before the procedure for continuous dextrose infusion while fasting. Vital signs on admission included a temperature 97.3 F, pulse 128, respiratory rate 34-minute, blood pressure 115/57 mmHg, SpO2 100%. The physical exam was unremarkable as there were no oral mucosal ulcers to explain his feeding aversion and he had age-appropriate dentition. There was no abdominal distention, pain, or hepatosplenomegaly. His CGM was located on his right outer thigh. Laboratory values were unremarkable (**Table 1**), and an upper gastrointestinal tract radiograph showed no evidence of small bowel malrotation. An infusion of Dextrose 10% with ½ Normal Saline was started at 42 ml/hr (Glucose infusion rate (GIR) of 6.4 mg/kg/min = maintenance rate with Holiday-Segar calculation) and once the child started fasting on the morning of the procedure, the rate was increased to 84 ml/hr (GIR 12.8 mg/kg/min) **(Table 2)**. Point-of-care glucose checks showed good correlation with CGM values with less than a 15-point deviation. While receiving a GIR of 12.8 mg/kg/min, the CGM readings did not decrease below 80. In comparison, when he is well at home, he demonstrates rapid drops in blood glucose levels every 2 hours typical for his disease process **(Figure 1).** The CGM device was used during the procedure and checked every 5 minutes. His family was also able to monitor his CGM readings in the waiting room. When the CGM reported 146 mg/dL, the continuous dextrose infusion was decreased from 84 ml/hr to 70 ml/hr (GIR 10.7 mg/kg/min). A sharp decline was observed in his CGM reading by 54 mg/dL within 15 minutes to 89 mg/dL (**Figure 2**). The family also noticed this rapid decrease in the CGM reading and communicated their concerns to the anesthesiologist, who increased his dextrose fluids back to 84 ml/hr. This increase in CGM readings occurred around the time the incisions for the gastrostomy tube were made and may have been related to cate-

cholamine stress response during surgery. The child tolerated the procedure well without any complications. The fasting time was 14 hours. Once enteral feedings were tolerated post-operatively at 4 hours, the dextrose infusion was decreased by 10% every hour over a 12-hour period while the child was fed every 2-3 hours. When the GIR decreased below 2.2 mg/kg/min or 14 ml/hr, the glycemic pattern started to match the patient's usual rise and fall pattern (**Figure 3**). The GCM alarmed with glucose < 70 mg/dL and prompted feedings. The child did well overnight and was fed through the gastrostomy tube allowing him to sleep longer. The following day, he maintained adequate glucose levels with enteral feedings and was discharged home. The family reported improvement in the child's sleep duration at night as they were able to feed him through the G-tube and administer glucose gel more efficiently in hypoglycemic emergencies.

In following up with the family, we discovered that the child was rushed to an outlying emergency department 1 month after discharge for refractory vomiting and diarrhea. They described the illness developing over a two-hour period from a suspected viral gastrointestinal illness from which the father was ill as well. The family administered 5 doses of glucose gel through the G-tube en route to the hospital over a 35-minute period to treat his hypoglycemia. He presented to the emergency department with blood glucose of 22 mg/dL and lactic acidosis of 11 mmol/L. He was treated with intravenous rehydration with continuous dextrose infusion improving blood glucose levels. The family reported that they coordinated with the pediatric hospital staff during the hospitalization in using the CGM device to assist with titrating the continuous dextrose infusion, reduce the need for frequent POC glucose checks, and provided important glycemic trends in response to intravenous dextrose therapy. His total hospital stay was 7 days as he recovered from the gastrointestinal illness and improvement in his lactic acidosis. The family stated that the use of the CGM during this hospitalization also helped to improve their child's care and reduced uncertainty with glucose monitoring in the hospital.

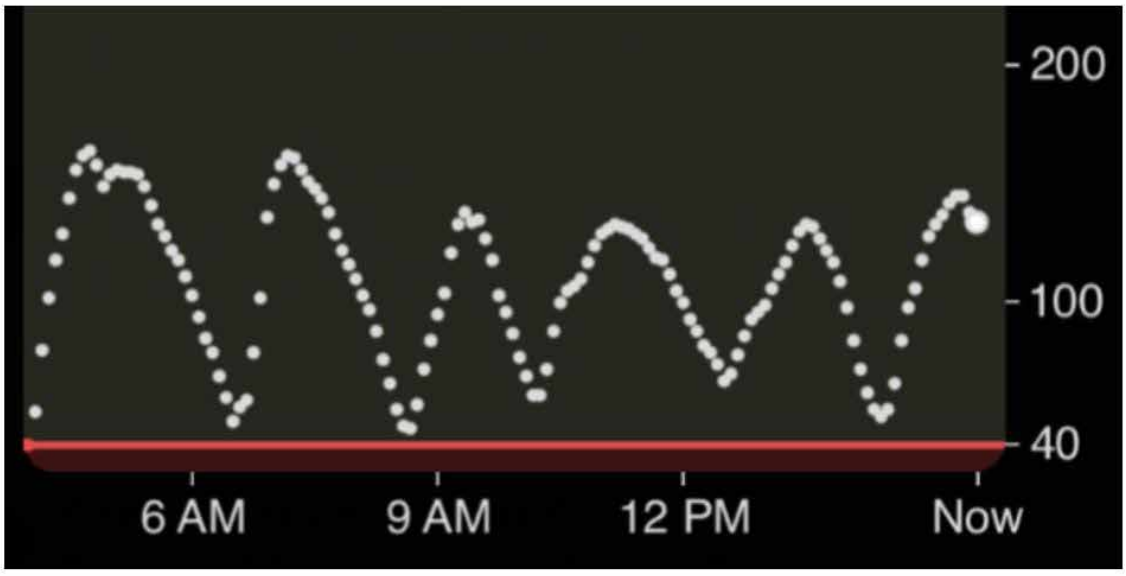

Figure 1: CGM display of glucose values (mg/dL) for typical glycemic trends when the child is well and tolerating feeds every 2 hours at home. Time progression is from the left to the right of the image. Glucose values fall rapidly within 2 hours requiring frequent monitoring of CGM device to coordinate feeds

Table 1: Basic Metabolic Panel on admission

Basic Metabolic Panel	
Sodium	136 MMOL/L
Potassium	4.2 MMOL/L
Chloride	98 MMOL/L
CO2	21 MMOL/L
Urea Nitrogen	8 MG/DL
Creatinine	< 0.38 MG/DL
Glucose	129 MG/DL
Calcium	10.5 MG/DL
Anion Gap	17 MMOL/L

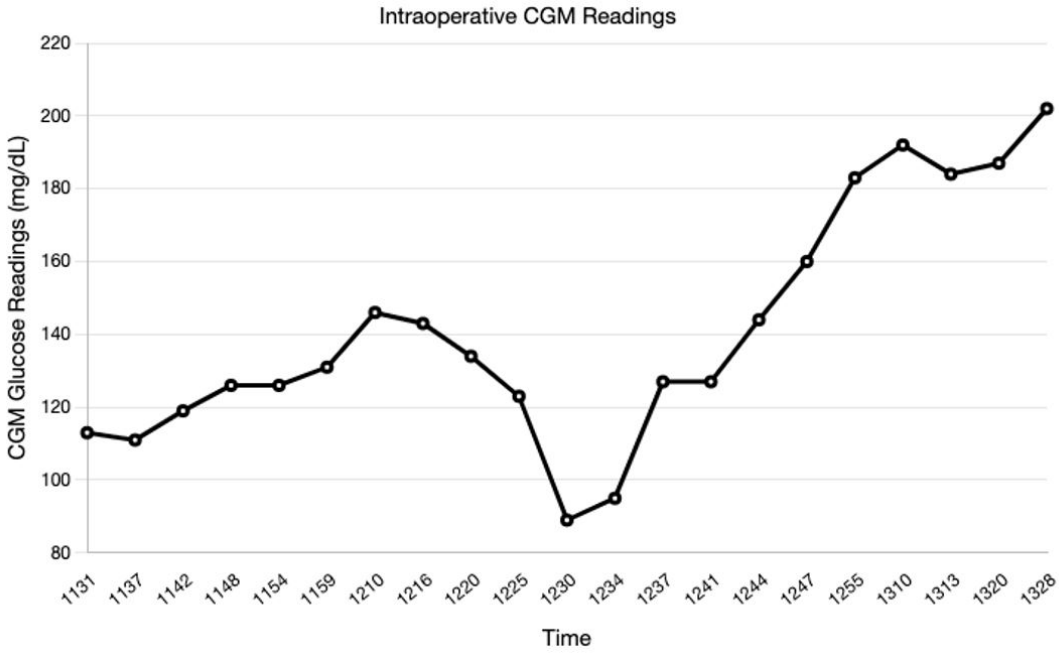

Figure 2: Intraoperative CGM recordings obtained by anesthesia during the procedure

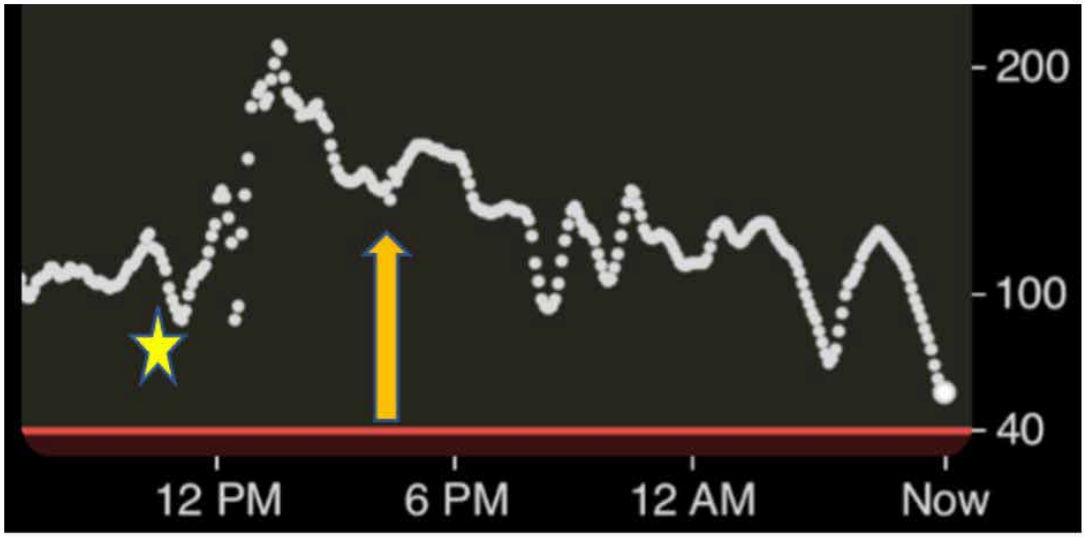

Figure 3: CGM display of glucose values (mg/dL) while the child was undergoing continuous dextrose infusion during fasting state (time is measured from the left of the screen to the right). Star indicates drop in glucose while undergoing surgical procedure from 13.1 mg/kg/min to 10.9 mg/kg/min. The up-arrow indicates the first wean of the dextrose infusion following the first enteral feeding post-procedure

Table 2: Table showing the inpatient feeding and glucose log for each hospital day. Dextrose infusion rate along with feeding volumes included. CGM measurements were similar in value to point-of-care glucose measurements

Hospital Day	Time	Dextrose 10% Infusion Rate (ml/hr)	CGM Glucose Reading (mg/dL)	Point-of-Care Glucose (mg/dL)	Feeding Amount (mL)
1	12:00	-	94	113	80
	13:10	-	70 (ALARM)	69	80
	15:30	42	92		120
	19:00	42	98		120
	20:16	42	124		120
2	00:02	42	81		150
	06:45	84	110	109	
	08:20	84	111	123	
	10:15	84	110		
Surgery					
	16:30	74	163		120
	19:00	54	136	147	
	20:20	54	98		
3	00:03	29	114		120 (G-tube)
	03:00	14	95		120
	06:00	-	70 (ALARM)		120

Discussion

Children with the rare inheritable metabolic condition glycogen storage disease 1a require strict frequent feedings every 2-3 hours to prevent severe hypoglycemia which can result in seizures, lactic acidosis, brain damage, or even death **[6].** These children are unable to regulate glucose homeostasis given the deficiency of the glucose-6-phophatase (G6Pase) enzyme preventing effective breakdown their glycogen stores into glucose **[7].** With the challenges of uninterrupted dietary treatments, a considerable number of children with GSD develop feeding difficulties, orofacial myofunctional disorders and fear of feedings **[8].** For infants with GSD Ia, maintaining blood glucose > 70 mg/dL is recommended to prevent life-threatening hypoglycemia, it is also recommended that parents be trained in inserting a nasogastric (NG) tube or that a G-tube be placed surgically to ensure reliable access for treatment of hypoglycemia **[6].** Inserting an NG tube incorrectly can result in severe complications such as aspiration pneumonia **[9].** The addition of cornstarch to a child's diet with GSD Ia has been associated with prolonging the fasting period and slowing the rapid drop in serum glucose levels **[10].** Our patient developed diarrhea following the initiation of cornstarch to his diet at 6 months of age and was discontinued. This may have been related to immature amylase enzyme activity for his age **[11].** Because of the challenges associated with feeding aversion and increasing hypoglycemic episodes, the decision to place a gastrostomy tube was made to provide more consistent feedings and an effective route to administering rescue dextrose gel during periods of severe hypoglycemia.

Frequent monitoring of blood glucose levels in children with GSD can be difficult but is critical to prevent hypoglycemia and lactic acidosis which can affect growth and brain development. Our patient was provided a CGM within the second week of life in the NICU. The family gained several months of experience with the device and understanding their child's typical glycemic trend. At home, when the CGM was not functioning properly, the family would rely on alarm clocks set every 1-2 hours to remind them to obtain point-of-care checks using a glucometer. Continuous glucose monitoring devices are commonly used in children with diabetes but are not approved for children less than two years of age **[12].** Devices such as the Dexcom G5 have been approved to replace fingerstick testing for children with diabetes over two years of age **[13].** For patients with GSD, the use of a CGM may allow detection of asymptomatic hypoglycemia and document glycemic trends **[6].** CGMs provide valuable insight when changes to GSD dietary regimens are made such as when the child has rapid growth and/or participates in various activities to optimize therapies **[14, 15].**

In 2017, the Diabetes Technology Society Panel consensus for CGM usage for inpatient care suggested that CGM in the hospital has the potential to improve patient clinical outcomes and reduce hypoglycemia **[16].** There are only a few studies that have evaluated CGM device usage for hospitalized children and those undergoing surgical procedures. Galderisis found a 20% improvement in euglycemia range for very preterm infants (<1500 grams) in the first 48 hours of life when CGM was used to assist with dextrose titration in this high-risk population **[17].** Piper and colleagues demonstrated that CGM can be used safely in infants and children less than 3 years' old who underwent cardiac bypass even when hypothermia was present and inotrope medications were administered **[18].**

The continuous glucose monitor used in our patient (Dexcom G6) is a minimally invasive device which uses subcutaneous sensors to determine glucose concentration in interstitial fluid **[19].** Glucose measurements are sent to the transmitter every 5 minutes. Potential issues and limitations related to our patient's CGM include a risk of skin irritation, infection at the insertion site, a 2-hour sensor warm up period after placement which requires point-of-care testing and calibration before CGM readings are available, the need for the transmitter to be within 6 meters of the child, and the risk for falsely elevated values following administration of acetaminophen **[20].** The sensor can last up to 10 days. The sensor used for our patient was on day 4 when he was admitted to the hospital and started the continuous dextrose infusion. The family had been placing the CGM device in the child's upper outer thigh to prevent the child from laying on it which may result in sensor error (**Figure 4**). The CGM readings, as seen in **Figure 2**, provided accurate monitoring of the child's glucose levels while fasting before, during, and following surgery. The CGM device was useful for titrating the continuous dextrose infusion during surgery along with weaning off the dextrose fluids in the post-operative phase (**Figure 3**) This child was at significant risk for hypoglycemia during the fasting period given the drastic drop with his typical glycemic trends (**Figure 1**). These typically occur 2 hours after a feeding. The utilization of the child's CGM proved to be quite valuable in titrating the continuous dextrose infusion while the child was in a prolonged fasting state while hospitalized and in preventing severe hypoglycemic episodes.

To our knowledge this is the first case in which a CGM device was successfully used for the management and prevention of hypoglycemia along with the titration of continuous dextrose infusion in a young child with GSD Ia undergoing prolonged fasting state for placement of a gastrostomy tube. Our experience with this case suggests that CGM devices in children with GSD undergoing prolonged fasting for surgical procedures can provide important glycemic trends, improve identification of hypoglycemic episodes, reduce point-of-care testing, and provide better patient centered care.

Conclusion

We describe the case of 10-month-old male with GSD Ia undergoing a prolonged fasting state while on continuous dextrose infusion admitted for G-tube placement. To our knowledge this is the first case to describe home CGM device usage in the hospital in such a young child with GSD for the titration of continuous dextrose infusion, prevention of severe perioperative hypoglycemia, and reduction of point-of-care testing in the prolonged fasting state.

Figure 4: Continuous glucose monitor shown in the child while standing. Placement of the CGM in the upper outer posterior thigh as seen in the figure. There is a skin mark just lateral to the CGM where an older sensor had been placed. The family switches legs after placing a new sensor and rotate insertion sites

Consent: Written informed consent was obtained from the patient's parents. Our signed consent form can be made available to the journal or ethics committee on request.

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