

Continuous glucose monitoring–guided optimization of peritoneal dialysis prescription in glycogen storage disease type 1a: a case report

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Case Report

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Abstract

Background

Glycogen storage disease type 1a (GSD-1a) is a rare inherited metabolic disorder characterized by impaired glucose homeostasis and progressive renal involvement. As life expectancy has improved, an increasing number of patients develop end-stage kidney disease (ESKD); however, optimal renal replacement therapy (RRT) strategies remain unclear.

Case presentation

We report a 48-year-old man with GSD-1a who progressed to ESKD and underwent initiation of peritoneal dialysis (PD). Despite dietary management, he experienced recurrent nocturnal hypoglycemia. Continuous glucose monitoring (CGM) was used to evaluate glucose profiles under different PD modalities. Intermittent PD without nocturnal dialysate dwelling and continuous ambulatory PD were insufficient to prevent nocturnal hypoglycemia. Switching to nightly intermittent PD enabled continuous nocturnal glucose absorption from dialysate exchanges, resulting in the resolution of nocturnal hypoglycemia without the need for late-night snacks.

Conclusion

This case demonstrates that CGM can be a useful tool not only for detecting asymptomatic hypoglycemia but also for individualizing PD prescription in patients with GSD-1a. Individualized RRT strategies based on metabolic characteristics may improve safety and quality of life in this rare population.

Background

Glycogen storage disease type 1a (GSD-1a) is a rare inherited disorder of glycogen metabolism caused by deficiency of glucose-6-phosphatase, resulting in impaired glucose homeostasis during fasting [1, 2]. Hypoglycemia is the most critical clinical feature and requires strict dietary management with frequent carbohydrate intake throughout life [3].

With advances in early diagnosis and supportive care, the life expectancy of patients with GSD-1a has improved substantially. Consequently, an increasing number of patients develop chronic kidney disease and eventually progress to end-stage kidney disease (ESKD), requiring renal replacement therapy (RRT). Although renal involvement is well recognized in GSD-1a, evidence guiding the optimal selection of RRT modality in this population is scarce, particularly regarding the management of recurrent hypoglycemia.

Here, we present a rare case of a patient with GSD-1a and ESKD in whom peritoneal dialysis (PD) was successfully introduced. Continuous glucose monitoring (CGM) was used to evaluate glucose profiles

under different PD modalities, enabling optimization of PD prescription to prevent nocturnal hypoglycemia.

Case Presentation

We report the case of a 48-year-old man with GSD-1a who developed ESKD. He presented with hepatomegaly in infancy and was diagnosed with GSD-1a. His medical history included diabetes mellitus requiring oral hypoglycemic agents, nephrolithiasis, hyperuricemia, hypertriglyceridemia, and osteoporosis, all considered complications of GSD-1a. He was referred to a nephrologist at 34 years of age because of persistent proteinuria. Despite continued management, his renal function progressively deteriorated, and he eventually reached ESKD. He chose PD as RRT and was admitted to the hospital for initiating PD.

On admission, physical examination revealed the following: height 165.5 cm, body weight 54.6 kg, body mass index 19.9 kg/m², BP 139/85 mmHg, HR 80/min, SpO₂ 99% (room air). He had characteristic facial features associated with GSD-1a and short stature. Laboratory tests showed renal dysfunction and metabolic acidosis. Type 2 diabetes mellitus was well controlled with an oral α -glucosidase inhibitor, with an HbA1c level of 5.5%. Nocturnal hypoglycemia had been prevented by a late-night snack. Dyslipidemia (hypertriglyceridemia) and hyperuricemia were managed with medication (Table 1).

Table 1
Laboratory findings at admission.

Parameter	Value	Unit	Reference range
WBC	7.5	$\times 10^9/\text{L}$	3.9–9.7
Hemoglobin	10.7	g/dL	13.4–17.1
Platelet count	251	$\times 10^9/\text{L}$	153–346
Serum creatinine	8.15	mg/dL	0.6–1.0
Blood urea nitrogen	35	mg/dL	9–21
eGFR	6.4	mL/min/1.73 m ²	> 60
AST	19	U/L	5–37
ALT	16	U/L	6–43
ALP	65	U/L	38–113
Albumin	3.6	g/dL	4.0–5.2
Total cholesterol	177	mg/dL	150–219
Triglycerides	120	mg/dL	30–149
HbA1c	5.5	%	4.6–6.2
Uric acid	4.0	mg/dL	3.5–6.9
Bicarbonate	16.5	mmol/L	22–26
Abbreviations: WBC, white blood cell; eGFR, estimated glomerular filtration rate; AST, aspartate aminotransferase; ALT, alanine aminotransferase; ALP, alkaline phosphatase.			

During hospitalization, diet therapy was continued with controlled total energy and carbohydrate intake, excluding fructose, galactose, sucrose, and lactose, in accordance with standard management for GSD-1a. Supplementary feeding was provided between meals, typically at mid-morning and mid-afternoon. Late-night snacks were also necessary to prevent asymptomatic nocturnal hypoglycemia.

He underwent implantation of a straight PD catheter (Long Shoot Catheter 650 with reinforcement JL-1(A)S®; Hayashidera Medinol, Kanazawa, Japan) using the short-term PD induction and education technique. The 4-h dialysate/plasma creatinine ratio was 0.66 by the peritoneal equilibration test, and the patient's small-molecule transport status was categorized as high average at PD initiation.

An intermittently scanned CGM device (FreeStyle Libre®; Abbott Diabetes Care, Alameda, CA, USA) was used to assess glucose profiles under different PD prescriptions. The device is an intermittently scanned CGM system that records interstitial glucose levels at 15-minute intervals. CGM data were collected for several consecutive days (typically 3–5 days) under each PD modality. Recurrent nocturnal

hypoglycemia was observed during intermittent PD without nocturnal dialysate dwell (Fig. 1a). Continuous ambulatory PD (CAPD) with four daily exchanges using 1.5% or 2.5% glucose dialysate also failed to prevent nocturnal hypoglycemia (Fig. 1b, c). Therefore, nightly intermittent PD (NIPD) with three nocturnal exchanges using 1.5% glucose dialysate was introduced to ensure continuous nocturnal glucose absorption. This strategy successfully prevented hypoglycemia without the need for late-night snacks (Fig. 1d). The final NIPD prescription consisted of three nocturnal exchanges using 1.5% glucose dialysate (1,500 mL each exchange) during 8 hours (2.5 hours of dwell per cycle). The total daily glucose load was approximately 61.2 g/day, weekly total Kt/V was 1.87, and daily ultrafiltration volume was approximately 0 to 100 mL. Time below range (TBR; <70 mg/dL) decreased from 25.0-37.5% during CAPD to 3.1% after switching to NIPD.

Discussion

GSD-1a is a rare autosomal recessive metabolic disorder characterized by impaired glucose homeostasis due to deficiency of glucose-6-phosphatase. Because endogenous glucose production during fasting is markedly impaired, patients require continuous exogenous carbohydrate supply, particularly during prolonged overnight fasting. In addition to metabolic abnormalities, GSD-1a is also associated with progressive renal involvement [4, 5]. Renal manifestations typically begin with glomerular hyperfiltration during childhood and may gradually progress to proteinuria and ESKD over time [6, 7]. With advances in early diagnosis and long-term dietary management, the life expectancy of patients with GSD-1a has improved substantially. Consequently, an increasing number of patients are now reaching adulthood and developing ESKD requiring RRT [8, 9].

However, evidence guiding the optimal selection of RRT modality in patients with GSD-1a remains extremely limited. Kidney transplantation or combined liver-kidney transplantation has been reported in selected cases, although long-term outcomes and the risk of recurrent nephropathy remain uncertain [10, 11]. Hemodialysis (HD) has also been successfully performed in some patients with GSD-1a [12]; however, frequent carbohydrate intake required to prevent hypoglycemia may lead to excessive interdialytic weight gain and hemodynamic instability during HD sessions [13]. Furthermore, intermittent HD provides only limited glucose exposure during treatment sessions, which may predispose patients with GSD-1a to fluctuations in blood glucose levels and increase the risk of hypoglycemia during the interdialytic period. In contrast, PD provides continuous exposure to glucose-containing dialysate and may therefore help stabilize glucose levels in patients with impaired endogenous glucose production [2]. Previous reports have suggested that PD may improve metabolic disturbances in patients with GSD-1a complicated by chronic kidney disease. However, the optimal PD prescription for preventing hypoglycemia has not been well described.

In the present case, nocturnal hypoglycemia persisted despite intermittent PD without nocturnal dialysate dwelling and continuous ambulatory PD (CAPD). The patient was categorized as a high-average transporter according to the peritoneal equilibration test. During prolonged nocturnal dwell, the glucose concentration gradient between dialysate and plasma gradually decreases, resulting in

diminished glucose availability over time. In patients with relatively high peritoneal transport characteristics, glucose absorption occurs rapidly and the osmotic gradient may dissipate earlier during the dwell period, potentially limiting sustained glucose delivery overnight.

Switching to nightly intermittent peritoneal dialysis (NIPD) enabled repeated nocturnal exchanges and continuous glucose exposure throughout the night. This strategy successfully prevented nocturnal hypoglycemia and eliminated the need for late-night snacks, thereby improving the patient's quality of life. Importantly, no symptomatic hypoglycemic episodes occurred after the PD prescription was modified. Although additional glucose exposure from dialysate may theoretically exacerbate hypertriglyceridemia in patients with GSD-1a, no clinically significant worsening was observed in this patient during the observation period.

To our knowledge, this is the first report describing the use of CGM to guide PD prescription in a patient with GSD-1a. CGM enabled detailed evaluation of glucose dynamics under different dialysis modalities and facilitated identification of nocturnal hypoglycemia that might not have been detected by intermittent capillary glucose measurements. These findings suggest that CGM may serve not only as a diagnostic tool for detecting asymptomatic hypoglycemia but also as a practical method for optimizing dialysis prescriptions in metabolically vulnerable patients.

Nevertheless, this report has several limitations. First, this is a single-case observation, and the optimal dialysis prescription may differ depending on peritoneal transport characteristics, residual kidney function, and dietary management. Second, long-term metabolic outcomes, including lipid metabolism, require further evaluation. Accumulation of additional cases will be necessary to establish optimal dialysis strategies for patients with GSD-1a complicated by ESKD.

Taken together, this case highlights the importance of individualized dialysis prescriptions that consider not only solute clearance but also the metabolic characteristics of the underlying disease. CGM-guided PD prescription may represent a practical strategy for improving metabolic stability and quality of life in patients with GSD-1a undergoing dialysis.

Conclusion

We report a rare and clinically informative case of GSD-1a complicated by ESKD in which PD prescription was successfully optimized using CGM. CGM enabled detailed assessment of glucose dynamics under different dialysis modalities and facilitated identification of nocturnal hypoglycemia that was not detected by routine monitoring. These findings suggest that CGM-guided PD prescription may represent a practical strategy for individualized dialysis management in patients with metabolic disorders associated with impaired glucose homeostasis.

Abbreviations

GSD-1a; glycogen storage disease type 1a

ESKD; end-stage kidney disease

RRT; renal replacement therapy

PD; peritoneal dialysis

CGM; continuous glucose monitoring

Declarations

Ethics approval and consent to participate

This report describes a single case encountered in routine clinical practice. According to the institutional policy of Juntendo University, ethical approval from an Institutional Review Board was not required for the publication of an individual case report. The study was conducted in accordance with the Declaration of Helsinki, and written informed consent for publication was obtained from the patient.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Availability of data and materials

The data supporting the conclusions of this article are included within the article. Further inquiries can be directed to the corresponding author.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

Haruna-Fukuzaki-Ishii conceived the article and drafted the manuscript. All authors read and approved the final manuscript.

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Figures

Figure 1

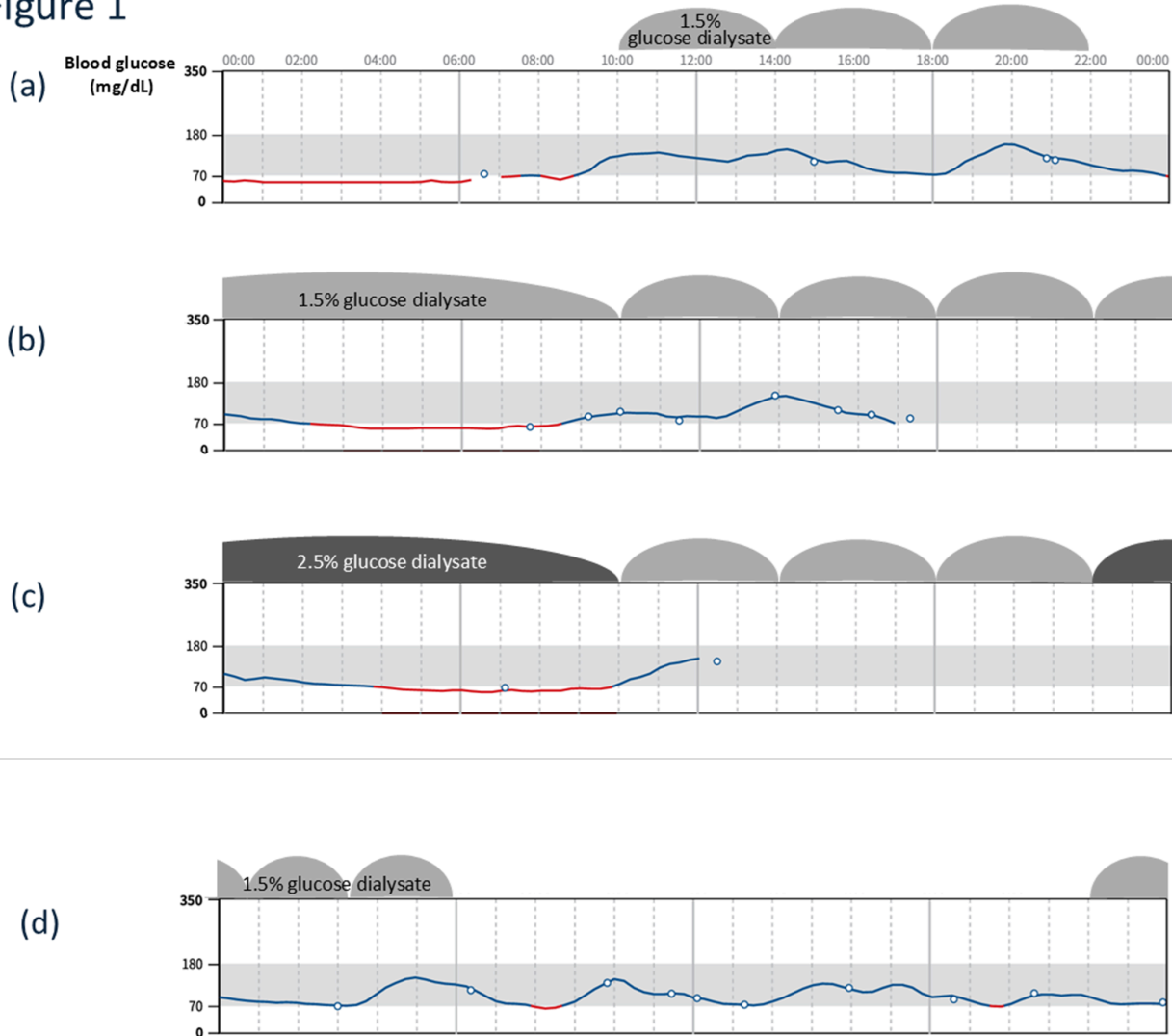


Figure 1

Continuous glucose monitoring profiles under different peritoneal dialysis modalities. All exchanges were performed with a dialysate fill volume of 1,500 mL. The shaded area indicates the target glucose range (70-180 mg/dL). CGM data were collected for several consecutive days under each dialysis modality.

(a) Intermittent peritoneal dialysis without nocturnal dialysate dwelling demonstrated recurrent nocturnal hypoglycemia.

(b, c) Continuous ambulatory peritoneal dialysis with four daily exchanges using 1.5% or 2.5% glucose dialysate did not sufficiently prevent nocturnal hypoglycemia.

(d) Nightly intermittent peritoneal dialysis with three nocturnal exchanges using 1.5% glucose dialysate markedly reduced nocturnal hypoglycemia without the need for late-night snacks.

