Case Report

Successful perioperative management of a pediatric patient with medium-chain acyl-CoA dehydrogenase deficiency using a continuous tissue glucose monitoring device: A case report

ABSTRACT

Medium-chain acyl-CoA dehydrogenase deficiency can cause symptoms resulting from β-oxidation disorder during preoperative fasting. Tight perioperative glucose monitoring is needed to avoid these symptoms. We report the first pediatric case using continuous tissue glucose monitoring devices. The patient was a 9-year-old boy with medium-chain acyl-CoA dehydrogenase deficiency for whom femoral osteotomy and selective muscle release of the hip and knee was planned to treat hip dislocation and joint contracture. To monitor tissue glucose levels continuously during preoperative fasting, a percutaneous sensor was attached to the right upper extremity, 2 days before the operation. Anesthetic management using Ringer's acetate containing 5% glucose, an inhalational agent, and epidural anesthesia without a muscle relaxant or propofol was performed without complications. The device achieved tight perioperative glucose monitoring. Continuous tissue glucose monitoring devices helped perioperative glucose monitoring of the pediatric patient with medium-chain acyl-CoA dehydrogenase deficiency.

Key words: Acylcarnitines; continuous tissue glucose monitoring; fasting; hypoglycemia; medium-chain acyl-CoA dehydrogenase deficiency

Introduction

Medium-chain acyl-CoA dehydrogenase deficiency (MCADD) is a common disorder of mitochondrial fatty acid β-oxidation,[1] resulting in hypoglycemia during preoperative fasting. The administration of glucose-containing solutions and frequent monitoring of blood glucose levels are crucial in the perioperative management of MCADD patients.[2]

The FreeStyle Libre (Libre®; Abbott Laboratories, Chicago, USA), continuous tissue glucose monitoring device (CTGMD), consists of a pair of reader and sensor. CTGMD can monitor tissue glucose levels continuously with a painless 1-s scan.[3] We herein report the first case in which a CTGMD was used in perioperative glucose monitoring for the management of a pediatric MCADD patient. In this case report, written informed consent was obtained from parents.

Case History

A 9-year-old boy (height 127 cm, weight 18.6 kg) was bedridden due to the sequelae of neonatal-onset MCADD.

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The patient usually had breakfast, lunch, and dinner around 7:00 am, 0:00 pm, and 7:00 pm, respectively. There were no symptoms of β-oxidation disorder, despite not having a midnight snack to prevent starvation. Femoral osteotomy and the selective muscle release of the hip and the knee were planned as treatments for hip dislocation and joint contracture. Preoperative blood tests, chest X-rays, and electrocardiography showed no abnormalities. A CTGMD sensor was attached to the right upper extremity 2 days before the operation. The perioperative tissue glucose levels are shown in [Figure 1]. While the minimum tissue glucose level before and during preoperative fasting was 40 and 54 mg/dL, respectively, the patient remained well until the day of the operation, even though he did not receive an intravenous infusion of glucose-containing solution.

The patient was taken into the operating theatre at 9:15 am without symptoms resulting from β-oxidation disorder after 14 h of preoperative fasting. General anesthesia was induced using oxygen, nitrous oxide, and sevoflurane, and then intravenous access was established on the right upper extremity. As the intraoperative infusion, Ringer's acetate containing 5% glucose was administered at a maintenance rate, according to Holliday and Segar's formula.[4] Tracheal intubation was performed without muscle relaxants. An arterial line was established in the left radial artery, and the epidural catheter was inserted from L4/5. The levels of blood glucose, tissue glucose, and serum-free/acylcarnitine at the same time points are shown in [Table 1]. The operation was finished without symptoms of β-oxidation disorder, and the patient returned to the general ward after extubation. The total anesthetic time was 484 min.

Intravenous infusion of Ringer's acetate containing 5% glucose was continued until 7 days after the operation because of the patient's poor appetite. His appetite gradually improved 6 days after the operation. The infusion rate decreased and the infusion was interrupted as his appetite improved. The CTGMD sensor was removed 8 days after the operation. We could confirm that the patient did not show low tissue glucose levels for 24 h after the discontinuation of intravenous infusion. He was discharged 2 months after the operation without complications.

Discussion

Anesthetic management of MCADD patients is performed to avoid hepatic glycogen depletion, which may be induced by fasting, surgical stress, and infection in the perioperative period. The guidelines published by the British Inherited Metabolic Disease Group (BIMDG) recommend that 10% glucose/0.45% saline infusion be started at maintenance rates, according to the Holliday and Seger formula, on the morning of the operation (from lunchtime) if the patient is on a morning (afternoon) list.[2] As a result, a previously published review showed some episodes of perioperative hyperglycemia that resulted from intravenous infusion (containing 10% glucose) and surgical stress, in which it was necessary to adjust the fluid regimen.[5] We, therefore, decided to manage with Ringer's acetate (containing 5% glucose), with tight perioperative glucose management using CTGMD.

The use of CTGMDs has facilitated the perioperative management of MCADD patients. In the preoperative period, with the use of the CTGMD, the patient was able to avoid unnecessary intravenous infusion before the operation. The CTGMD sensor was attached 2 days before the operation to monitor tissue glucose levels before and during preoperative fasting. While low tissue glucose levels of approximately 40–54 mg/dL were observed, there were no symptoms of β-oxidation disorder. It is possible to start intravenous infusion containing glucose during the preoperative fasting period if symptoms of β-oxidation disorder are observed or if the tissue glucose levels deteriorate. In the intraoperative period, the use of the CTGMD reduced the number of blood samples required for blood glucose measurements. Although hyperglycemia (approximately 150 mg/dL) occurred due to the administration of an intravenous infusion (containing 5% glucose) and surgical stress, the tissue glucose levels recovered to within the normal range without adjusting

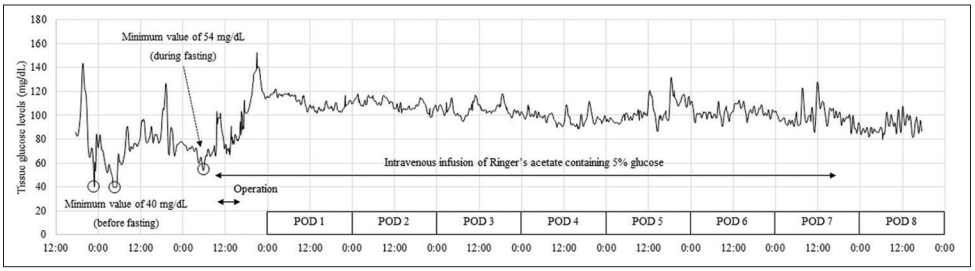

Figure 1: Perioperative tissue glucose levels. The tissue glucose levels were monitored using a CTGMD (FreeStyle Libre®; Abbott Laboratories, Chicago, USA).(CTGMD, continuous tissue glucose monitoring devices; POD, postoperative day)

Table 1: The levels of blood glucose, tissue glucose, and serum-free/acyl-carnitine during the operation

Events (Time)	Induction (9:20 am)	(10:42 am)	(0:25 pm)	(2:06 pm)	(3:31 pm)	Before emergence (4:52 pm)
Blood glucose (mg/dL)	64	122	93	84	84	115
Tissue glucose (mg/dL)	67	102	68	76	79	88
Free carnitines (μmol/L)	34.9	-	-	-	-	23.3
Acylcarnitine (μmol/L)	28.9	-	-	-	-	9.0
Acyl/Free carnitine ratio	0.83	-	-	-	-	0.39

Reference values: Free carnitine (36-74 μmol/L), Acylcarnitines (6-23 μmol/L), Acyl/Free carnitine ratio (0.12-0.30)[7]

the fluid regimen. Even if hyperglycemia had persisted, the CTGMD could have been used to repeatedly monitor tissue glucose levels after adjusting the fluid regimen. In the postoperative period, CTGMD made it easy to monitor the glucose levels of non-appetite patients over several days. Since a loss of appetite could cause symptoms resulting from β-oxidation disorder, frequent fingerstick measurements are required to monitor blood glucose levels in patients without a CTGMD. The sensors of the Libre®, work for up to 14 consecutive days and are suitable for perioperative glucose monitoring.

On the other hand, the appropriate range of tissue glucose levels for MCADD patients is unclear. While previous reports have shown a good correlation between blood glucose levels and tissue glucose levels measured with the FreeStyle Libre®,[6] our results [Table 1] show some discrepancies. Besides, since hypoglycemia is a late symptom resulting from β-oxidation disorder, the elevation of free fatty acid or acylcarnitine, which represents a biochemical disturbance, might be more appropriate as an early indicator. In this case, the accumulation of acylcarnitines was observed after 14 h of preoperative fasting. This was improved at the end of the operation when the glucose-containing solution was administered for 7.5 h. While the patient showed no symptoms of β-oxidation disorder during preoperative fasting, potential β-oxidation disorder might have been present.

In conclusion, while the CTGMD facilitated the perioperative management of pediatric MCADD patients, further studies should be undertaken to determine the appropriate range of tissue glucose levels for avoiding biochemical disturbances in MCADD patients.

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Conflicts of interest

There are no conflicts of interest.

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