

Preserving Therapeutic Ketosis During Perioperative Management of Adult-Onset Glucose Transporter Type 1 Deficiency Syndrome: A Case Report and Practical Anesthetic Considerations

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Case Report

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Abstract

Background

Glucose transporter type 1 deficiency syndrome (GLUT1DS) is a rare metabolic encephalopathy in which ketone bodies serve as an essential cerebral energy substrate. In the perioperative setting, conventional management strategies may inadvertently suppress ketogenesis and compromise cerebral energy metabolism.

Case presentation

We report the perioperative management of a woman in her 40s with adult-onset GLUT1DS undergoing hip osteotomy under total intravenous anesthesia (TIVA). The patient had been maintained on a well-controlled ketogenic diet prior to surgery. Anesthesia was induced and maintained using propofol, remifentanyl, and rocuronium, while glucose-containing solutions were avoided. Lipid emulsion was administered during fasting as an alternative energy substrate to support ketogenesis. Depth of anesthesia was continuously monitored using the bispectral index (BIS), which remained within the normal range throughout the procedure. Perioperative laboratory findings demonstrated stable blood glucose levels without hypoglycemia or hyperglycemia, suppressed insulin levels, and elevated ketone bodies with preserved acid–base balance. The patient recovered uneventfully and resumed a ketogenic diet 4 hours after emergence without neurological complications.

Conclusions

This case highlights that perioperative management focused on preserving therapeutic ketosis may be feasible in adult-onset GLUT1DS. Careful metabolic and neurological monitoring, avoidance of unnecessary glucose administration, and predefined contingency plans are essential for safe anesthetic management.

Background

Glucose transporter type 1 deficiency syndrome (GLUT1DS) is a rare neurological disorder caused by impaired glucose transport across the blood–brain barrier due to dysfunction of the GLUT1 transporter [1]. This defect results in chronic cerebral energy deficiency, as glucose is the primary energy substrate for the brain under normal physiological conditions.

The estimated prevalence of GLUT1DS is approximately 1 in 90,000 individuals, with most cases diagnosed in infancy or early childhood [2,3]. Adult-onset presentations are considered exceptionally rare, and detailed epidemiological data, particularly in adult populations, remain limited [4].

In patients with GLUT1DS, ketone bodies function as an essential alternative cerebral energy substrate, compensating for impaired glucose transport. Consequently, ketogenic diet therapy is the standard treatment and has been shown to improve neurological symptoms and quality of life [5]. Importantly, in

this population, elevated circulating ketone body levels represent a therapeutic metabolic state rather than a pathological condition [6].

For anesthesiologists unfamiliar with GLUT1DS, perioperative management presents a unique challenge. Standard anesthetic practices—such as perioperative glucose administration or suppression of ketogenesis—may inadvertently compromise cerebral energy supply and precipitate neurological deterioration. While perioperative management strategies have been reported in pediatric or adolescent cases [7,8], evidence regarding anesthetic management in adult-onset GLUT1DS remains extremely limited, with only isolated case reports currently available. Moreover, practical perioperative strategies—including metabolic monitoring and contingency planning to maintain therapeutic ketosis, defined by preserved acid–base balance without metabolic complications—remain insufficiently described, particularly in adult-onset cases.

Here, we report the perioperative management of an adult-onset GLUT1DS patient undergoing orthopedic surgery under total intravenous anesthesia (TIVA). This case highlights key considerations for maintaining therapeutic ketosis during anesthesia and illustrates how perioperative metabolic monitoring can help distinguish physiological ketosis from pathological ketoacidosis.

Case Presentation

A woman in her 40s (height 168 cm, weight 50 kg) was diagnosed with adult-onset GLUT1DS. Prior to diagnosis, she had experienced persistent fatigue and generalized malaise of unknown cause. These symptoms improved markedly after initiation of a ketogenic diet, which prompted further investigation and confirmation of GLUT1DS. Since starting dietary therapy, she had remained clinically stable and physically active. She had no history of epilepsy, seizures, loss of consciousness, or ketoacidosis.

The patient was scheduled to undergo transposition osteotomy of the acetabulum of the hip. This was her first surgical procedure. She had no history of diabetes mellitus, other metabolic disorders, or previous exposure to general anesthesia, and her cardiopulmonary function was normal.

The patient had been on a well-controlled ketogenic diet prior to surgery, with good adherence confirmed by sustained clinical improvement and elevated ketone body levels. Preoperative fasting was well tolerated without adverse neurological or metabolic events. Anesthesia was induced and maintained using total intravenous anesthesia with propofol (target-controlled infusion; 2.0–3.0 µg/mL), remifentanyl (0.1–0.3 µg/kg/min), and rocuronium (intermittently administered; total dose 50 mg). Depth of anesthesia was continuously monitored using the bispectral index (BIS), which remained within the normal target range throughout the procedure. The duration of anesthesia was 147 minutes, and the surgical time was 104 minutes. Estimated blood loss was 120 mL, the intraoperative fluid balance was +446 mL, and no blood transfusion was required.

Glucose-containing solutions were deliberately avoided throughout the perioperative period to prevent suppression of ketogenesis. During fasting, 50 mL of 20% lipid emulsion was administered slowly to

provide an alternative energy substrate.

Perioperative laboratory data, including blood glucose, serum insulin, ketone body fractions, and venous blood gas parameters, are summarized in Table 1. Despite fasting and surgical stress, blood glucose levels remained within the normal range throughout the perioperative period, with no episodes of hypoglycemia or hyperglycemia observed.

The patient emerged smoothly from anesthesia without neurological symptoms. Oral intake was resumed approximately 4 hours after emergence with a ketogenic diet, carefully planned and supervised by a multidisciplinary nutrition team. Her postoperative course was uneventful, and she was discharged without complications.

Discussion

Glucose transporter type 1 deficiency syndrome (GLUT1DS) presents a unique perioperative challenge because cerebral energy metabolism depends primarily on ketone bodies rather than glucose [1,5]. Consequently, metabolic states typically considered abnormal, such as hyperketonemia, represent a therapeutic condition in this population [5,6]. In contrast, conventional perioperative practices—including glucose administration or suppression of ketogenesis—may paradoxically impair cerebral energy supply and lead to neurological deterioration [2,5].

In the present case, therapeutic ketosis was successfully preserved throughout the perioperative period. Despite prolonged fasting and surgical stress, blood glucose levels remained stable without hypoglycemia or hyperglycemia, while ketone body levels remained elevated and acid–base balance was maintained. These findings support the interpretation that the patient remained in a state of controlled therapeutic ketosis rather than pathological ketoacidosis. Importantly, this distinction should be based on comprehensive metabolic assessment, including pH, bicarbonate, and lactate levels, rather than ketone concentrations alone [5].

A key aspect of perioperative management in this case was the implementation of continuous neurological monitoring using the bispectral index (BIS), which remained within the normal range throughout the procedure. Although BIS is not a direct measure of cerebral metabolism, stable intraoperative values may provide supportive but indirect information regarding anesthetic depth. Objective monitoring of neurological status is particularly important in GLUT1DS, where clinical signs may be masked during general anesthesia.

The patient had been maintained on a well-controlled ketogenic diet prior to surgery, which likely contributed to metabolic stability during the perioperative period [5]. Preoperative optimization of ketogenic therapy should therefore be considered an essential component of perioperative planning in patients with GLUT1DS.

Total intravenous anesthesia (TIVA) was selected to minimize potential metabolic variability during anesthesia. In this case, TIVA allowed stable metabolic control without fluctuations in blood glucose or ketone levels, which may be advantageous when preserving therapeutic ketosis is a primary goal. While inhalational anesthesia has also been reported to be feasible in adult patients with GLUT1DS [9], the choice of anesthetic technique should be guided by the ability to maintain metabolic stability rather than by a specific modality alone. Experimental studies have suggested interactions between anesthetic agents and ketone metabolism [10], although the clinical relevance in humans remains unclear.

Another important consideration is the establishment of a contingency plan for potential metabolic or neurological deterioration. In the event of hypoglycemia, cautious glucose administration would be required while avoiding abrupt suppression of ketosis [5]. Conversely, if metabolic acidosis or excessive ketosis had developed, adjustments in fluid and lipid management would have been necessary. Lipid administration may support ketone body production by providing fatty acid substrates, although evidence in the perioperative setting remains limited. In addition, any signs of neurological deterioration would have prompted immediate reassessment of metabolic parameters and consideration of alternative energy supplementation strategies. Although such events did not occur in this case, proactive planning is essential for safe perioperative management.

Early postoperative resumption of a ketogenic diet was also prioritized. Resumption of enteral ketogenic intake is critical to maintain cerebral energy supply, and close collaboration with a multidisciplinary team is required to ensure continuity of care [5,11].

This report has several limitations. It describes a single adult-onset case undergoing a relatively short surgical procedure, and the findings may not be generalizable to more complex or prolonged surgeries. In addition, optimal perioperative ketone target ranges in adults with GLUT1DS remain poorly defined [11]. Further accumulation of cases is needed to establish standardized perioperative management strategies.

This case report was prepared in accordance with the CARE reporting guideline for case reports.

Abbreviations

GLUT1DS: Glucose transporter type 1 deficiency syndrome

TIVA: Total intravenous anesthesia

BIS: Bispectral index

Declarations

Ethics approval

Not applicable.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

HT managed the perioperative care of the patient, conceptualized the report, and drafted the manuscript.

AK contributed to patient management and collected the clinical data.

KT assisted in perioperative management and contributed to data interpretation.

SI critically revised the manuscript for important intellectual content.

All authors read and approved the final manuscript.

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Table

Tables

Table 1. Perioperative laboratory data.

Ketone bodies represent the sum of acetoacetate and β-hydroxybutyrate.

Laboratory data	Normal range	Pre-op	Before surgery	After surgery	POD 1
Blood glucose (mg/dL)	70~109	84	80	80	101
Serum insulin (μU/mL)	1.7~10.4	2.9	0.8	1.3	4.2
Total ketone bodies (μmol/L)	22~122	613	1670	1280	964
Acetoacetate (μmol/L)	13~69	190	330	290	236
β-hydroxybutyrate (μmol/L)	0~76	423	1340	989	728
Venous blood gas data					
pH	7.35~7.45	7.421	7.437	7.337	7.347
HCO ³⁻ (mmol/L)	20~28	20.58	21.47	24.0	20.0
Lactate (mmol/L)	0.5~2.2	1.2	1.0	1.9	1.7