Case Report

Managing Anaesthesia in GLUT1 Deficiency Syndrome: Case Report

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Abstract

Glucose Transporter Type 1 Deficiency Syndrome (GLUT1-DS) is a rare genetic disorder caused by mutations in the SLC2A1 gene, leading to impaired glucose transport across the blood–brain barrier. Those patients are susceptible to neurologic decompensation, particularly during periods of stress such as surgery. The ketogenic diet, which provides ketones as an alternative energy source, is the cornerstone of treatment. Anaesthetic management in these patients requires careful planning to maintain metabolic stability and preserve neurologic function. Unfortunately, no formal guidelines exist for anaesthetic management.

We present the case of a 26-year-old female with confirmed GLUT1-DS undergoing elective septoplasty and inferior turbinate reduction under general anaesthesia. The patient was on a strict ketogenic diet and had a history of dystonia, epilepsy, and dyskinesia. Perioperative strategies focused on maintaining ketosis, preventing hypoglycaemia, and avoiding agents that could lower the seizure threshold. Fasting was limited to 8 hours, fluids were restricted to 0.9% NaCl and blood glucose was closely monitored (range 4.6–4.8 mmol/L). Anaesthesia was induced with propofol and sufentanil, maintained with sevoflurane and remifentanil, and supported by multimodal non-opioid analgesia. The procedure was uneventful, with no signs of neurologic deterioration postoperatively. The ketogenic diet was resumed the same day.

This case highlights the importance of individualised perioperative management in GLUT1-DS. Elaborate planning and multidisciplinary collaboration that includes maintenance of ketosis, avoidance of glucose-containing solutions, and careful selection of anaesthetic agents is essential to prevent neurologic decompensation. Further studies are warranted to establish evidence-based guidelines.

Keywords: GLUT1 deficiency syndrome; ketogenic diet; anaesthesia; perioperative management

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1 Introduction

Glucose transporter type 1 deficiency syndrome (GLUT1-DS) is a rare neurometabolic disorder caused by mutations in the SLC2A1 gene, which encodes the GLUT1 protein responsible for glucose transport across the blood-brain barrier (1, 2). Impaired glucose transport results in chronic cerebral energy deficiency, leading to a characteristic clinical triad of drug-resistant epilepsy, developmental delay, and complex movement disorders (3). To date, approximately 105,000 cases of GLUT1-DS have been diagnosed worldwide, although recent data suggest a gradual increase in recognised cases due to improved genetic testing (4, 5). Owing to its complex and pleiotropic phenotype, GLUT1-DS is still likely underdiagnosed (6). Early diagnosis and initiation of a ketogenic diet are crucial for improving neurological outcomes by supporting brain development during this vulnerable period (7). Maintenance of ketosis is therefore essential, especially in the perioperative period, where fasting and stress may exacerbate neurological symptoms or precipitate seizures (3). In patients with GLUT1-DS, standard anaesthetic practices, such as prolonged preoperative fasting, use of glucose-containing intravenous fluids, or interruption of dietary therapy can pose serious risks (6). Despite these challenges, there are no formal guidelines for anaesthetic management in GLUT1-DS. This case report presents a patient with genetically confirmed GLUT1 deficiency undergoing general anaesthesia, illustrating key perioperative considerations and strategies to minimise neurologic decompensation.

2 Case Report

A 26-year-old female with genetically confirmed GLUT1 deficiency, diagnosed in childhood after presenting with dystonia, epilepsy, and dyskinesia, was scheduled for elective septoplasty and inferior turbinate reduction. She had been on a 2.5:1 ratio (fats : carbohydrates and proteins) ketogenic diet since 2015 with good seizure control. She also had longstanding nasal obstruction, previously treated with adenoidectomy, but with persistent septal deviation and turbinate hypertrophy. Preoperative preparation included maintaining normoglycaemia and continuing the ketogenic diet. The last meal was 8 hours prior to the surgery, and the last recorded blood glucose level was 4.8 mmol/L.

Upon arrival in the operating room, standard anaesthetic monitoring was established, and one large-bore peripheral intravenous line was inserted. Clinical assessment did not suggest a difficult airway, and anaesthesia was induced using 20 mcg of sufentanil, 140 mg of propofol, and 50 mg of rocuronium. Endotracheal intubation was performed without complications or signs of aspiration. Anaesthesia was maintained with sevoflurane in an oxygen and air mixture, and continuous remifentanil infusion at 100 mcg/h. As additional analgesic therapy, the patient received 1 g of paracetamol and 300 mg of ibuprofen. For antiemetic prophylaxis, 8 mg of dexamethasone and 4 mg of ondansetron were administered. Fluid replacement consisted of 0.9% NaCl. More intense glucose monitoring was used intraoperatively, since the maintenance of a

normoglycaemic status was considered as especially important to avoid brain seizures, with an average measured glucose of 4.6 mmol/L.

Throughout the procedure, the patient remained haemodynamically and respiratory stable, with no need for additional interventions. At the end of the surgery, the patient was awake, breathing spontaneously, and transferred to the recovery room, where no additional analgesia or oxygen therapy was required. The patient was then transferred back to the ward with a recommendation for regular monitoring of blood glucose, electrolytes, and urinary ketones. No postoperative nausea or other adverse effects of general anaesthesia were reported. Diet was introduced at the ward the same day, after the patient was fully awake and without nausea.

3 Discussion

This case highlights the perioperative challenges and management strategies in a patient with Glucose Transporter Type 1 Deficiency Syndrome. Although the syndrome is rare, the pleiotropic manifestations and risk of neurologic decompensation necessitate individualised anaesthetic approaches (3, 8). The key pathophysiologic feature is impaired glucose transport across the blood-brain barrier, resulting in chronic cerebral energy deficiency (1). Maintenance of ketosis via ketogenic dietary therapy is the mainstay of treatment and has been shown to significantly reduce seizure burden and improve neurodevelopmental outcomes (9).

The perioperative period poses specific risks to patients with GLUT1-DS. Prolonged fasting, perioperative stress, and use of glucose-containing intravenous fluids can disrupt ketosis, precipitate seizures, and worsen movement disorders (2, 8). For this reason, strategies must focus on minimising fasting, avoiding dextrose-containing fluids, and maintaining normoglycaemia while preserving ketosis. In the present case, these principles were successfully implemented: fasting was limited to 8 hours, intravenous fluids were restricted to isotonic saline, and normoglycaemia was maintained throughout the procedure. Importantly, urinary ketone monitoring was recommended postoperatively to ensure ongoing adherence to the ketogenic regimen.

Anaesthetic agents must also be selected with caution. Comparable perioperative approaches have been described in the literature. Kloka et al. (2019) described the anaesthetic management of a GLUT1-DS patient using total intravenous anaesthesia with propofol and remifentanil, emphasising the importance of avoiding glucose-containing solutions and maintaining normoglycaemia throughout the procedure. Their successful outcome supports our approach and further illustrates that both inhalational and intravenous techniques can be safely employed when metabolic stability is closely monitored (10).

While no single agent has been shown to directly exacerbate GLUT1-DS, agents that significantly alter cerebral metabolism or glucose homeostasis should be avoided (2). In this case, induction with propofol and sufentanil, maintenance with sevoflurane and remifentanil, and multimodal analgesia with paracetamol and ibuprofen were well tolerated. The avoidance of long-acting opioids and benzodiazepines likely contributed to the smooth emergence and stable recovery.

Reports in the literature emphasise that there are no formal anaesthetic guidelines for GLUT1-DS (8). Nevertheless, a common theme in published case descriptions is the emphasis on perioperative maintenance of ketosis and glucose homeostasis. One review noted that "maintenance of dietary therapy is essential to prevent perioperative seizures and neurologic deterioration" (2). This case supports such observations and demonstrates the feasibility of safely performing elective surgery under general anaesthesia in these patients when meticulous perioperative planning is employed.

4 Conclusion

This case underscores the importance of tailored perioperative management in patients with GLUT1-DS. Key considerations include minimising fasting, avoiding glucose-containing solutions, ensuring normoglycaemia, and maintaining ketogenic dietary therapy. Anaesthetic agents that preserve haemodynamic stability without interfering with cerebral glucose metabolism can be safely used. Although no standardised guidelines currently exist, careful preoperative planning and multidisciplinary collaboration can mitigate risks and optimise outcomes in this vulnerable patient population. Future studies and case series are warranted to establish evidence-based recommendations for anaesthetic management in GLUT1 deficiency syndrome.

Ethics Statement

For every elective and urgent procedure in our Hospital, it is required to obtain an informed consent form. The patient had signed the informed consent form and therefore gave the Hospital permission to perform procedures as well as use the data for scientific purposes with strong protection of personal information.

Conflict of Interest

The authors declare that they have no conflicts of interest.

Author Contributions

A.J. and R.C.R. managed the patient, supervised the anaesthetic procedure and conceptualised the report. A.J. and N.M. reviewed the literature and drafted the manuscript. R.C.R. revised the manuscript and approved the final version.

References

1. De Vivo DC, et al. Defective glucose transport across the blood-brain barrier as a cause of persistent hypoglycorrhachia, seizures, and developmental delay. N Engl J Med. 1991;325(10):703–709.

2. Leen WG, et al. Glucose transporter-1 deficiency syndrome: the expanding clinical and genetic spectrum of a treatable disorder. Brain. 2010;133(3):655–670.

3. Klepper J, Leiendecker B. GLUT1 deficiency syndrome—2007 update. Dev Med Child Neurol. 2007;49(9):707–716.

4. Tang M, Monani UR. Glut1 deficiency syndrome: New and emerging insights into a prototypical brain energy failure disorder. Neurosci Insights. 2021 Apr 28;16:26331055211011507. doi: 10.1177/26331055211011507.

5. Klepper J. Glut1 Deficiency Syndrome: Novel Pathomechanisms, Current Concepts, and Challenges. J Inherit Metab Dis. 2025 May;48(3):e70044. doi: 10.1002/jimd.70044.

6. Mochel F, et al. Ketogenic diet in Glut1 deficiency syndrome: a 10-year follow-up. J Inherit Metab Dis. 2010;33(6):803–810.

7. Neal EG, et al. The ketogenic diet for the treatment of childhood epilepsy: a randomized controlled trial. Lancet Neurol. 2008;7(6):500–506.

8. Gras D, Roze E, Caillet S, et al. GLUT1 deficiency syndrome: An update. Rev Neurol (Paris). 2014;170(2):91–99. doi: 10.1016/j.neurol.2013.09.005.

9. Alter AS, Engelstad K, Hinton VJ, et al. Long-term clinical course of Glut1 deficiency syndrome. J Child Neurol. 2015;30(2):160–169. doi: 10.1177/0883073814531822.

10. Kloka J, Kranepuhl S, Zacharowski K, Raimann FJ. Total Intravenous Anesthesia in GLUT1 Deficiency Syndrome Patient: A Case Report. Am J Case Rep. 2019 May 5;20:647–650. doi: 10.12659/AJCR.914865.