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Successful Anesthetic Management of a Liver Transplant Recipient With Glycogen Storage Disease Type 1a Undergoing Dental Surgery: A Case Report

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Abstract

Glycogen storage disease type 1a (GSD 1a) is a rare metabolic disorder characterized by impaired glucose regulation, often requiring liver transplantation (LT) to manage complications. Anesthetic management in these patients can be challenging, particularly during surgical interventions, due to their metabolic instability and increased surgical risks.

This report discusses the anesthetic management of a LT recipient with GSD 1a undergoing elective dental surgery. Comprehensive perioperative planning ensured metabolic stability and minimized complications.

A multidisciplinary approach, meticulous monitoring, and individualized anesthetic planning are essential for successful outcomes in patients with complex metabolic and transplant histories.

Keywords

Glycogen Storage Disease Type 1a (GSD 1a); Liver Transplantation; Dental Surgery; Anesthetic Management; Perioperative Care; Multidisciplinary Approach.

Introduction

Glycogen storage disease type 1a (GSD 1a), also known as von Gierke disease, results from a deficiency in glucose-6-phosphatase. This enzyme is vital in the final stage of glycogenolysis, and gluconeogenesis is essential for maintaining blood glucose levels during fasting. The absence of this enzyme results in the accumulation of glycogen in the liver and kidneys, leading to hepatomegaly, renal dysfunction, and severe fasting hypoglycemia. Patients with GSD 1a often exhibit additional metabolic derangements, including hyperuricemia, hyperlipidemia, and lactic acidosis [1]. The primary treatment for GSD 1a involves meticulous dietary management, including frequent meals rich in complex carbohydrates and avoiding fasting to prevent hypoglycemia. However, some patients experience progressive complications, such as hepatic adenomas, cirrhosis, or portal hypertension, necessitating LT. Liver transplantation not only corrects the enzymatic defect in the liver but also alleviates many systemic complications, significantly improving the patient's quality of life [1,2].

Despite the success of liver transplantation, these patients remain metabolically fragile and are prone to complications, particularly in the perioperative period. Surgery and anesthesia introduce significant metabolic stress, which can exacerbate the residual risks of hypoglycemia, electrolyte imbalances, and acidosis. Additionally, immunosuppressive therapy required for transplant recipients adds complexity to perioperative management [2].

Dental procedures requiring general anesthesia pose unique challenges for GSD 1a patients with LT. Dental infections or poor oral health can impact overall health, particularly in immunosuppressed individuals. The decision to proceed with general anesthesia must account for the patient's metabolic vulnerabilities, the need for precise glucose control, and the prevention of drug interactions that could affect immunosuppressant levels. This case report highlights the careful planning and multidisciplinary coordination necessary for the successful anesthetic management of an LT recipient with GSD 1a undergoing elective dental surgery.

Case Presentation

Preoperative situation

A 4-year-old male child with a history of GSD 1a underwent LT in 2022 due to complications associated with the disease. At the time of the dental procedure, the patient weighed 18 kg and regularly took sirolimus for immunosuppression. Additionally, he had received intravenous immunoglobulin (IVIg) treatment three months prior to surgery. Due to a tooth abscess, amoxicillin was administered one week before the procedure. In order to publish this case, the patient's verbal and written consent was obtained before the operation.

Anesthetic management

The patient's American Society of Anesthesiologists

(ASA) physical classification was III, reflecting significant systemic disease. Preoperative laboratory evaluations revealed hemoglobin of 12.1 g/dL, leukocyte count of 8.06 × 10^3/mm^3, and platelet count of 272 × 10^3/mm^3. Coagulation parameters showed an INR of 1.06 and aPTT of 30.1 seconds. Liver function tests indicated normal bilirubin levels (T. Bil: 0.60 mg/dL, D. Bil: 0.19 mg/dL) and transaminase levels (ALT: 17 U/L, AST: 45 U/L). The albumin level was 4.4 g/dL. Given the risk of hypoglycemia during the preoperative fasting period, a continuous infusion of 40 mL/h of 1/3 saline with 10% dextrose was initiated and kept throughout the intraoperative phase. Standard monitoring was implemented, involving electrocardiography, noninvasive blood pressure and pulse oximetry. After preoxygenation was achieved with 80% oxygen for three minutes, anesthesia was induced with lidocaine (1 mg/kg), propofol (2 mg/kg), and rocuronium (0.6 mg/kg). Intubation was performed using a 5 mm cuffed endotracheal tube. Although the Cormack–Lehane score was 2 during laryngoscopy, the procedure was completed without difficulty or needing adjuncts such as bougies or guides. After intubation, anesthesia was maintained with a 50% oxygen+air mixture, inhaler sevoflurane 2.2%. Post-intubation arterial blood gas analysis showed a pH of 7.44, PaO_2 of 109 mmHg, $PaCO_2$ of 32 mmHg, lactate level of 1.1 mmol/L, and blood glucose of 135 mg/dL, with electrolytes within normal limits. The patient was ventilated in volume control mode with a 6 mL/kg tidal volume, respiratory rate of 20 breaths/min, PEEP of 3 cmH_2O, and FiO_2 of 0.4. The dental procedure, which lasted approximately 45 minutes, included a compomer filling for teeth 53,63,83,72, extractions of teeth 55,54,5 2,51,61,62,65,75,74,73,83,84 and 85, and fissure sealant treatments for tooth 64. Vital signs remained stable throughout, with heart rate between 80-110/min, blood pressure ranging from 100–80/55–50 mmHg, SpO_2 at 98–100%, and $EtCO_2$ levels of 32–40 mmHg. Analgesia was provided with 25 mg of tramadol, and no additional doses of rocuronium were required. Spontaneous ventilation was recovered without the use of neostigmine. Extubation was performed after spontaneous breathing returned, tidal volume exceeded 4 mL/kg, and laryngeal secretions were completely cleared. There were no signs of laryngospasm or bronchospasm after extubation, and the process was completed without any problems.

Post-surgical course

The patient was transferred to the recovery room and the pediatric in-patient service. He was discharged in good condition with stable vital signs two days after surgery. This case underscores the importance of meticulous perioperative planning and glucose management in a pediatric patient with complex medical conditions, such as GSD 1a and a history of LT, to ensure a successful anesthetic and surgical outcome.

Discussion

The successful anesthetic management of a pediatric liver transplant recipient with GSD 1a undergoing dental surgery presents several unique challenges. This case emphasizes critical considerations in the perioperative management of such patients, focusing on metabolic control, immunosuppression, and airway management, all of which are pivotal for the successful outcome of such procedures. Glycogen Storage Disease Type 1a (GSD 1a) is characterized by impaired glycogen breakdown and gluconeogenesis, making patients highly susceptible to hypoglycemia during fasting periods. Hypoglycemia, if left unaddressed, can result in serious complications such as seizures, brain damage, or even death. In this case, the risk of hypoglycemia was mitigated by administering an intravenous infusion of 1/3 physiological saline prepared with 10% dextrose, ensuring stable blood glucose levels throughout the procedure. This approach is consistent with current recommendations for managing patients with GSD 1a during the perioperative period, highlighting the importance of glucose infusion to prevent hypoglycemia and associated complications (Derks et al., 2021) [3]. Monitoring blood glucose, lactate, and electrolytes is essential for optimizing metabolic control and preventing metabolic crises in these patients (Zavala et al., 2016) [4]. Liver transplant recipients often exhibit altered hepatic function and may have ongoing immune suppression as a result of immunosuppressive medications. The patient had normal liver enzymes (AST, ALT) and bilirubin in this case. However, in the case of elevated liver function test results, extremely careful anesthetic management should be planned for LT recipients, as the liver plays a crucial role in drug metabolism and clearance. Sevoflurane and propofol were chosen due to their minimal hepatotoxic effects and favorable pharmacokinetic properties in patients with compromised liver function (Mangus RS et al., 2018) [5]. Due to immunosuppressive therapy makes LT patients more vulnerable to infections, making careful perioperative infection control paramount (Chelala et al., 2015; Wu DX et al., 2024) [6,7]. For pain management, tramadol was selected for its mild impact on hemodynamics and its suitability in pediatric patients for short procedures (Thigpen JC et al., 2019) [8]. However, The use of these weak opioids is not recommended in patients with hypersensitivity to codeine and tramadol (e.g., ultrarapid metabolizers). Understanding variations in CYP450 enzymes is crucial for evaluating drug metabolism and anticipating potential risks [9]. The absence of neostigmine, a commonly used agent for the reversal of muscle relaxants, reduced the risk of potential complications such as bradycardia and nausea in this patient. The patient's smooth recovery with spontaneous ventilation and successful extubation without additional neuromuscular blockade antagonism

aligns with best practices for muscle relaxant management in pediatric anesthesia (Yang L et al., 2023) [10]. Patients with immunosuppressive therapy, such as those who have undergone LT, are at heightened risk for infections, particularly oral infections. Dental procedures in these patients require careful planning to avoid complications that could result in systemic infections. This patient underwent an extensive dental procedure, including multiple extractions and a fissure sealant treatment, under general anesthesia. The procedure was completed without incident, illustrating the importance of a multidisciplinary approach that includes collaboration between anesthesiologists, surgeons, and dentists. Preventive dental care is crucial for transplant patients at increased risk for local and systemic infections due to compromised immune system (Ziebolz et al., 2011) [11].

Conclusion

This case highlights several key principles in the anesthetic management of pediatric patients with complex medical histories, especially those with GSD 1a and LT. First, managing hypoglycemia through glucose infusion and continuous monitoring of metabolic parameters is essential to prevent potentially life-threatening complications. Second, anesthetic agents must be carefully selected for liver dysfunction or immunosuppression patients. Finally, multidisciplinary collaboration is necessary to ensure the safe and effective management of complex cases, as demonstrated by the coordination between the anesthesia and dental teams in this case.

Further research is needed to develop standardized protocols for anesthetic management in pediatric liver transplant recipients, particularly those with co-existing metabolic disorders like GSD 1a. This case demonstrates that with comprehensive preoperative evaluation, careful planning, and close monitoring, even complex pediatric patients can safely undergo elective procedures with favorable outcomes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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References

1. Rake JP, Visser G, Labrune P, Leonard JV, Ullrich K, Smit GP. Glycogen storage disease type I: diagnosis, management, clinical course and outcome. Results of the European Study on Glycogen Storage Disease Type I (ESGSD I). Eur J Pediatr 2002;161: 20-34.
2. Williams R. Sherlock's disease of the liver and biliary systems. Clin Med (Lond) 2011;11(5):506.
3. Derks TGJ, Rodriguez-Buritica DF, Ahmad A, de Boer F, Couce ML, Grünert SC, et al. Glycogen Storage Disease Type Ia: Current Management Options, Burden and Unmet Needs. Nutrients 2021;13(11):3828.
4. Owusu-Agyemang P, Zavala AM, Van Meter AK, Williams UU, Ifeanyi IC, Hagan KB, et al. Perioperative management of a child with Von Gierke disease. Open J Clin Med Case Rep 2016; 1140.
5. Mangus RS, Kinsella SB, Farar DT, Fridell JA, Woolf LT, Kubal CA. Impact of Volatile Anesthetic Agents on Early Clinical Outcomes in Liver Transplantation. Transplant Proc 2018;50(5):1372-1377.
6. Chelala L, Kovacs CS, Taege AJ, Hanouneh IA. Common infectious complications of liver transplant. Cleve Clin J Med 2015;82(11):773-84.
7. Wu DX, Hu JX, Wu XL, Han JN, Chang KY, Quan XL, et al. Preoperative Evidence-Based Practice for Prevention of Early Postoperative Infections in Patients Receiving a Liver Transplant. Ann Transplant. 2024;29:e943610.
8. Thigpen JC, Odle BL, Harirforoosh S. Opioids: A Review of Pharmacokinetics and Pharmacodynamics in Neonates, Infants, and Children. Eur J Drug Metab Pharmacokinet 2019;44(5):591-609.
9. Fortenberry M, Crowder J, So TY. The Use of Codeine and Tramadol in the Pediatric Population-What is the Verdict Now? J Pediatr Health Care. 2019 Jan;33(1):117-123. doi: 10.1016/j.pedhc.2018.04.016. PMID: 30545525.
10. Yang L, Hu N, Chang H, Yang D, Zuo Y. Routine administration of neostigmine after recovery of spontaneous breathing versus neuromuscular monitor-guided administration of neostigmine in pediatric patients: a parallel, randomized, controlled study Trials 2023;24(1):19.
11. Ziebolz D, Hraský V, Goralczyk A, Hornecker E, Obed A, Mausberg RF. Dental care and oral health in solid organ transplant recipients: a single center cross-sectional study and survey of German transplant centers. Transpl Int 2011;24(12):1179-88.

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