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Total Intravenous Anesthesia in GLUT1 Deficiency Syndrome Patient: A Case Report

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 2
Final Diagnosis: GLUT1 deficiency syndrome
Symptoms: Mastoiditis
Medication: —
Clinical Procedure: General anesthesia
Specialty: Anesthesiology

Objective: Congenital defects/diseases

Background: GLUT1-deficiency-syndrome (G1DS) is an autosomal dominant genetic disorder based on a mutation of the SLC2A1 gene. This mutation can lead to an encephalopathy due to abnormal glucose transport in the brain. G1DS is a rare disease, with an estimated incidence of 1: 90 000.

Case Report: We report a case of a 10-year-old female who presented with recurrent fever, headaches, and vertigo for more than 3 days within 2 weeks following pneumonia. A bilateral mastoiditis was proven by a cerebral magnetic resonance imaging and a cranial computed tomography scan. The patient had to undergo mastoidectomy and thus, her first general anesthesia. Half a year previously she was diagnosed with G1DS. According to the standard of care, a ketogenic diet had been administered since the patient's diagnosis 6 months earlier. Our patient received a total intravenous anesthesia (TIVA) using propofol, fentanyl, and rocuronium administered without any incidents.

Conclusions: We recommend normoglycemia during the perioperative phase and avoidance of glucose-based medication to keep a patient's ketotic state. Our case highlights that TIVA, with the outlined medication used in this case, was safe when the patient's ketotic state and periprocedural blood glucose was monitored continuously. Nevertheless, we would suggest using remifentanil instead of fentanyl for future TIVAs due to a reduced increase in blood glucose level in our patient.

MeSH Keywords: Anesthesia, Intravenous • Glucose Transporter Type 1 • Ketogenic Diet • Metabolism, Inborn Errors • Monosaccharide Transport Proteins • Rare Diseases

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Background

GLUT1 deficiency syndrome (G1DS) is an autosomal dominant genetic disorder based on a mutation of the SLC2A1 gene. This mutation can lead to an encephalopathy due to abnormal glucose transport in the brain. G1DS is considered as a rare disease, with an estimated birth incidence of 1 in 90 000 [1]. Until 2007, fewer than 100 cases were reported worldwide [2]. It is very likely that G1DS is underdiagnosed because of its complex and pleiotropic phenotype [3]. An early diagnosis followed by a ketogenic diet is crucial to improve neurologic outcomes by nourishing the immature brain during this important period of development [4].

In 1991, De Vivo et al. first described 2 patients with infantile seizures, delayed development, and acquired microcephaly with the biochemical finding of low glucose concentration in the cerebrospinal fluid and a normal level of glucose in the blood, which was caused by a GLUT1-deficiency [5].

In 2005, Wang et al. found that 13 out of 16 patients (81%) with G1DS suffered from the most common classic phenotype. This phenotype presents with symptoms like delayed neurologic development, dysarthria, acquired microcephaly, and complex movement disorders including ataxia, dystonia, and chorea. Seizures were unresponsive to typical anticonvulsant medication, but responded rapidly to a ketogenic diet, while 2 patients suffered from a milder non-classic G1DS (~10% of affected individuals) [6]. The non-classic G1DS type is characterized with no clinical seizures, frequent paroxysmal dyskinesia including intermittent ataxia, choreoathetosis, dystonia, and alternating hemiplegia [7].

To the best of our knowledge, there is no information available on performing total intravenous anesthesia (TIVA) in this population. Therefore, we want to share and discuss our findings of a patient with G1DS who was admitted to our hospital for an emergency mastoidectomy undergoing this operation using TIVA.

Case Report

A 10-year-old female weighing 32 kg presented with repeated fever attacks, headaches, and vertigo for more than 3 days within 2 weeks following pneumonia. When admitted to the hospital, she presented a new acute ear infection, lethargy, and fever with a high C-reactive protein (CRP) level of 46 mg/dL.

The antibiotics were changed from oral cefaclor to intravenous cefuroxime adapted to her bodyweight. After consulting the Ear-Nose-Throat Department of our University Hospital, additional radiological diagnostics were performed. A bilateral mastoiditis was proven by cerebral magnetic resonance imaging (MRI) and cranial computed tomography (CT) scan. The patient had to undergo mastoidectomy and thus, her first general anesthesia. The history of our patient includes seizures and impaired global development.

Lamotrigine, levetiracetam, and Orfiril (valproate) were used for her long-term antiepileptic medication. Half a year previously, the diagnosis of G1DS was made. According to the standard of care, a ketogenic diet was administered since her diagnosis 6 months previous. This modified Atkins diet allows for 15 grams of medium-chain triglyceride [1] fats, and 15 grams of carbohydrates per day.

At first contact, the physical examination showed signs of dehydration. The patient's pupils were medium sized and equal while showing a slightly belated but consensual reaction to light. Our patient was somnolent and hardly spoke. The parents reported signs of photophobia and reported that the headache had gotten worse over the last 2 days. The patient preferred to lie with knees bent. Stretching her legs increased discomfort. The parents also reported that the child had not properly eaten in at least 2 days and had been drinking very little. However, the girl had approximately 100 mL water to drink within the last 2 hours of admission.

We did not detect any indicators of a difficult airway and performed a rapid sequence induction according to our standard operating procedure using 0.1 mg fentanyl, 200 mg of 1% propofol, and 25 mg rocuronium.

The tracheal tube was placed without any difficulties or signs of aspiration. We continued the anesthesia totally intravenously with a continuous infusion of 2% propofol (6–9 mg/kg per hour) and repeated doses of fentanyl (cumulative dose 0.5 mg per 4 hours). The girl required rather high doses of both hypnotics and fentanyl to maintain an adequate level of anesthesia. High doses of hypnotics and fentanyl could be due to enzyme induction by long-term administration of anticonvulsants. As an additional pain medication, she received 330 mg of metamizole and 7.5 mg of piritramid. We chose 1 g of meropenem as an antibiotic perioperative single shot. Ventilation and oxygenation presented no difficulties throughout the whole procedure. Neither did circulation, heart rhythm, or heart rate.

First, we aimed for rehydration using balanced crystalloid intravenous solutions. A bolus of 350 mL was started before induction and we continued the administration of 330 mL per hour for the first hour of the procedure. Moreover, in this case with infection and too little calorie intake over the last days, a pediatric balanced solution containing 1% glucose was used to stabilize metabolism. The patient received 8 g of glucose within 4.5 hours.

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During induction and over the nearly 4 hours of surgery, the patient was kept warm using an inflatable hot-air blanket. Shortly after surgical completion of the mastoidectomy, the patient awoke promptly, free from any respiratory or circulatory problems. No additional pain medication was needed in the recovery room. Our patient was sleepy but easily woke when touched. For the first 15 minutes, we supplemented oxygen by nasal cannula with a flow of 4 L per minute.

Blood glucose tests and venous blood gas analyses were performed hourly during the procedure and once more in the recovery room. At first, glucose was low (83 mg/dL). After cautious supplementation, we detected 94 mg/dL before transporting the patient to the pediatric ward. Potassium remained within the normal range. On the pediatric ward, urine testing for ketone bodies was carried out repeatedly. There were no reports of nausea or any other negative side effects after general anesthesia.

Discussion

G1DS is a rare disease with less than 100 reported cases until to 2007 [2]. The literature on this disease is very limited. To the best of our knowledge, there is no data available on performing TIVA on a GLUT1-deficit patient. G1DS patients require a ketotic diet which should be started directly after diagnosis to reduce neurological complications. Therefore, we should consider all parts of TIVA and its influence on a patient's blood glucose level.

Opioids

In this patient's case, for TIVA we used repetitive doses of fentanyl. Draskovic et al. tested whether fentanyl, alfentanil, or remifentanil in combination with rocuronium and propofol could lead to cortisol and/or blood glucose increases during herniectomy or orchidopexy. Therefore, 150 ASA I–II male children age 2 to 5 years old were divided into 3 groups. All operations were performed in the morning to avoid the influence of circadian rhythm on hormone levels. The highest increase in cortisone was recorded in the fentanyl group. Corresponding to this finding, the blood glucose level increased the most in the fentanyl group from induction until awakening. The lowest increase of blood glucose was recorded in the remifentanil group [8].

Besides the fact that anesthesia with fentanyl was tolerated well by our patient, we recommend using remifentanil in other cases of anesthesia in G1DS patients to avoid an increase in blood glucose during the procedure. In our patient's case, the patient suffered from reduced caloric intake over a couple of days. Therefore, in this special situation, we decided to supplement our medication with glucose until normal blood glucose level was restored.

Hypnotics

On the one hand, there is evidence suggesting that barbiturates inhibit glucose transport in patients, and might harm them [9], while on the other hand, there is evidence that propofol-based anesthesia is more effective at stabilizing blood glucose level. Cok et al. [10] compared propofol-remifentanil with sevoflurane-remifentanil for craniotomy with respect to blood glucose levels. In both groups, due to stress response, blood glucose increased while levels were kept more stable in the propofol group [10]. In addition, volatile anesthetics solely has been shown to have a suppressing effect on insulin secretion [11,12].

Muscle relaxants

We used rocuronium as a representative of steroid muscle relaxants. There is no evidence, that rocuronium influences blood glucose level.

Conclusions

In summary, we present the case of a 10-year non-fasting patient who received TIVA using propofol, fentanyl, and rocuronium performed without any incidents. We recommend normoglycemia during the perioperative phase and avoidance of glucose-based medication to keep the patient's ketotic state. Our case highlights that TIVA, with the outlined medication used in this case, was safe when the patient's ketotic state and periprocedural blood glucose was monitored continuously. Nevertheless, we would suggest using remifentanil instead of fentanyl for future TIVAs due to a reduced increase of blood glucose level.

Conflict of interests

None.

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