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CASE REPORT

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Type 1 citrullinemia patient with Brugada pattern undergoing general anesthesia for dental extractions: A case report

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Key Clinical Message

The perioperative control of ammonia, reduction of stress, and administration of drugs tolerated in type 1 citrullinemia and Brugada pattern allowed the successful and uneventful management of general anesthesia in the study patient.

Abstract

The aim of this study was to report the targeted perioperative management of general anesthesia (GA) adopted for dental extractions in a rare patient with type 1 citrullinemia and Brugada pattern. A male, Caucasian, adult type 1 citrullinemia patient needed dental extractions under GA. The medical history showed neurodevelopmental impairment, growth retardation, epilepsy, and a Type 2 Brugada electrocardiographic pattern in the second precordial lead. The authors focused the anesthesiologic protocol on the prevention of hyperammonemia and fatal arrhythmias. Changes in diet and 10% glucose solution administration prevented protein catabolism due to the fasting period (ammonia was 44 μmol/L preoperatively and 46 μmol/L postoperatively; glycemia was 120 g/dL preoperatively and 153 g/dL postoperatively). The patient received a continuous electrocardiogram, noninvasive blood pressure, pulse oximeter, entropy monitoring, train-of-four monitoring, and external biphasic defibrillator pads. Midazolam, remifentanil, and dexamethasone were administered for pre-anesthesia; thiopental and rocuronium for induction; remifentanil and desflurane for maintenance; sugammadex for decurarization. After the intraligamentary injection of lidocaine 2% with epinephrine 1:100,000 for local anesthesia, the patient developed a transient Type 1 Brugada pattern that lasted a few minutes. The whole procedure lasted 30 min. The patient's discharge to ward occurred 3 h after the end of GA. The perioperative management of ammonia, reduction of stress, and administration of drugs tolerated in Type 1 citrullinemia and Brugada pattern allowed the successful and uneventful administration of GA in the study patient.

KEYWORDS
Brugada syndrome, citrullinemia, dental care, general anesthesia, lidocaine

1 | INTRODUCTION

Patients affected by urea cycle disorders (UCDs), such as type 1 citrullinemia, show various degrees of neurodevelopmental impairment because of the neurological damage induced by hyperammonemia during growth. Those patients may be poor or not cooperative with dental practitioners because of their cognitive deficits. Thus, general anesthesia (GA) is the only way to receive dental treatments for those special needs patients. Anesthesiologic management requires perioperative measures for patients with UCDs to prevent the development of hyperammonemia as a life-threatening complication.[1,2] In addition, other comorbidities may require more safety precautions, such as a Brugada pattern exposing the patient to the risk of developing a lethal ventricular arrhythmia in response to the administration of local and general anesthetic agents.[3] The purpose of this study is to show the targeted perioperative management of GA that the authors adopted for dental extractions in a rare patient with Type 1 citrullinemia and Brugada pattern. The authors reported the current case in line with Surgical Case Report (SCARE) guidelines 2020.

2 | CASE PRESENTATION

In May 2021, a male, Italian, Caucasian, 23-year-old patient came to the authors' attention because of recurrent bilateral mandibular odontogenic abscesses. The patient was accompanied by his mother as the caregiver who gave informed consent to treatment and publication since he suffered from a cognitive deficit. The family history was unremarkable and without cases of sudden cardiac death (SCD). The patient was born pre-term by Cesarean section in the 35th week. In childhood, the patient showed neurodevelopmental impairment, growth retardation, and epilepsy that led to the Type 1 citrullinemia diagnosis. Daily the patient received 0.25 mg of risperidone preventing psychotic episodes, two administrations of glycerol phenylbutyrate (3.5 mL/m^2) chelating ammonia, and up to two doses of an osmotic laxative agent (macrogol 4000). The patient was on a low-protein diet that allowed the intake of 16 g of proteins per day. Four times per day, the patient was supplemented with 2.49 g of L-arginine and 2 g of urea cycle disorders 2 (UCD2) synthetic essential amino acids to compensate for the deficit of argininosuccinate synthase. The patient was overweight (BMI of 26.83, 62 kg, and 1.52 m in height). In addition, the patient was also supplemented with minerals, multivitamins, 5 mg of vitamin B9 weekly, and 1000 µg of vitamin B12 twice a week. In 2020, a routine electrocardiogram (ECG) showed a Type 2 Brugada pattern, a "saddleback" ST segment elevated ≥1 mm in V2 (Figure 1).[4] Anyway, the following cardiologic consult excluded the hypothesis of Brugada Syndrome (BrS) because of his medical history. Moreover, the patient did not undergo a head-up tilt test, electrophysiological study, genetic testing for BrS, or even the ajmaline or flecainide challenge test.[5,6] The patient needed GA for the extraction of the lower left second premolar and lower right first molar. Preliminary standard blood tests showed normal values for glycemia (88 mg/

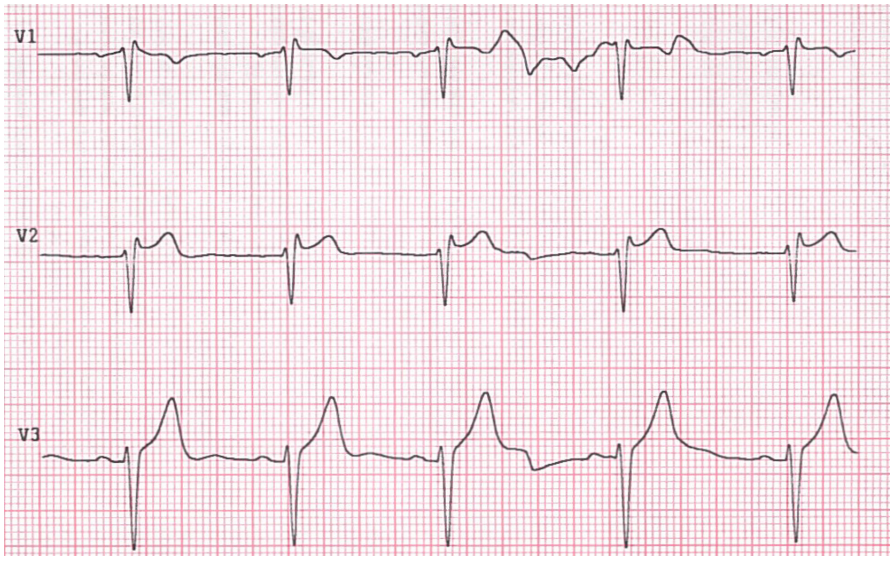

FIGURE 1 Preoperative ECG showing Type 2 Brugada pattern. The current picture shows five consecutive type 2 Brugada patterns in the second precordial leads as "saddle-back" ST elevations ≥1 mm. BrS, Brugada syndrome.

dL), bilirubin (total, 0.58 mg/dL; direct, 0.18 mg/dL; indirect, 0.40 mg/dL), aspartate aminotransferase (24 U/L), alanine aminotransferase (36 U/L), serum gamma-glutamyl transferase (43 U/L), prothrombin time (11.7 s, INR = 1.12), activated partial thromboplastin time (27.1 s), albumin (4.1 g/dL), and cholinesterase (15,323 U/L; dibucaine number, 84%). On the day of surgery, the patient received risperidone, glycerol phenylbutyrate, L-arginine, and 10 g of dietary instead of the usual 16 g; in addition, the patient avoided all other medications and supplementations. After obtaining peripheral venous access, the authors administered 500 mL of normal saline solution and 100 mL of 10% glucose solution per hour. In addition, the authors placed continuous ECG, noninvasive blood pressure, pulse oximeter, external biphasic defibrillator pads, and both entropy and train-of-four (TOF) monitoring. Preoperative ammonia was 44 µmol/L, whereas glycemia was 120 g/dL. The patient received 2 g of cefazolin as an antimicrobial prophylaxis protocol. The anesthetist administered 1 mg of midazolam, 35 µg of remifentanil, and 4 mg of dexamethasone as pre-anesthesia. Then, the anesthetist induced GA by using 300 mg of thiopental and 60 mg of rocuronium. Nasotracheal intubation (diameter 5 mm, armed cuffed tube) was chosen to avoid the invasion of the surgical field. The anesthetist set the ventilation on pressure-regulated volume control (PRVC) mode, the positive end-expiratory pressure (PEEP) at 5 cmH_2O, and the additional inspiratory oxygen at 40%. The patient's end-tidal CO_2 values ranged between 35 and 40 mmHg. GA was maintained by administering 0.1 µg/kg/min of remifentanil and 4.9% of desflurane. The oral surgeon administered local anesthesia by intraligamentary injecting two 1.8 mL vials of lidocaine 2% with epinephrine 1:100,000. After the administration of local anesthetic, the ECG showed the Type 1 Brugada pattern, a "coved-type" ST elevation ≥2 mm with a negative T wave (Figure 2).[4] The maximum elevation reached by the ST segment was 4.4 mm and reduced gradually to the range of 2.2–2.6 mm. The ECG turned to a physiologic pattern within a few minutes. 200 mg of Sugammadex was administered for decurarization. The whole procedure lasted 30 min. Postoperative ammonia was 46 µmol/L, while glycemia was 153 g/dL. Once the patient awakened, the ECG and pulse oximeter monitoring lasted for a further 3 h. Postoperative analgesic therapy comprised 1000 mg of acetaminophen and 30 mg of ketorolac. Glucose and saline solution administration was disrupted 2 h after the end of surgery. The ECG did not show alterations during the observation period. Then, the patient was discharged to the ward with a glycemia of 122 g/dL. The authors discharged the patient to home after overnight hospital observation. The patient completed the 30-day follow-up period without complications.

3 | DISCUSSION

The current case report showed a Type 1 citrullinemia patient carrying a Brugada pattern. This is an unusual finding in such a rare disease, and there are no previous reports of other patients affected by both conditions to the best of our knowledge. Citrullinemia is a rare recessive autosomal disease (incidence = 1:57,000 people) and is one of the most common UCDs.[1,2,7] The urea cycle is the enzymatic pathway that allows the renal clearance of the waste nitrogen by converting it to ammonia and then to urea.[7,8]

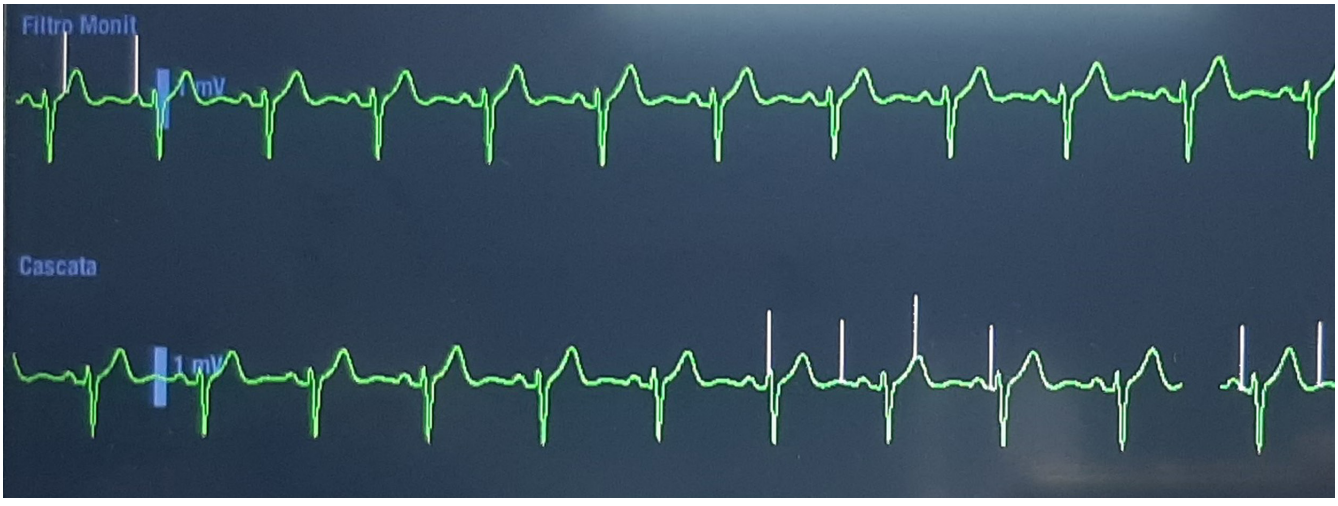

FIGURE 2 Intraoperative ECG showing induced Type 1 Brugada pattern. This picture shows the Type 1 Brugada pattern, as a "coved-type" ST elevation ≥2 mm with a negative T wave, which occurred under GA after the injection of local anesthesia by lidocaine 2% with epinephrine 1:100,000. The maximum ST elevation was 4.4 mm whereas the minimum was 2.2 mm. The pattern resolved spontaneously and uneventfully before the end of surgery. BrS, Brugada syndrome; GA, general anesthesia.

UCDs interfere with the synthesis of urea thus causing the accumulation of ammonia (hyperammonemia), which damages the central nervous system inducing lethargy, seizures, ataxia, severe neurodevelopmental impairment, and eventually death.[2,7,8] Type 1 or "classic" citrullinemia has neonatal onset and is caused by the deficit of argininosuccinate synthase which condensates citrulline and aspartate in L-argininosuccinate, whereas Type 2 citrullinemia has adult-onset (range 11–79 years) and is caused by the deficit of the citrin (aspartate glutamate carrier 2, AGC2).[1,2,7,8] BrS is a rare inherited autosomal dominant disease of the myocardial ionic channels (commonly Na^+), with incomplete penetrance and variable expressivity characterized by an increased risk of developing fatal arrhythmias and SCD in a normally structured heart.[3,5] Early diagnosis of BrS can be achieved by recording the Type 1 ECG pattern (a "coved-type" ST elevation ≥2 mm with a negative T-wave in a right precordial lead) whether spontaneous or induced by sodium channel blockers from Type 2 (a "saddleback" ST elevation ≥1 mm) or Type 3 pattern ("coved-type" or a "saddleback" ST elevation of maximum 1 mm). The patients showing the Type 1 ECG pattern and at least one symptom or sign receive the diagnosis of the syndrome, whereas those without symptoms or signs receive the diagnosis of the Brugada pattern.[3] Syncope, nocturnal agonal respiration, previous ventricular fibrillation or polymorphic tachycardia, susceptibility to ventricular tachycardia in programmed electrical stimulation tests, and family history positive for SCD are manifestations of the BrS. Only syndromic patients carry a high risk of SCD and thus receive an implantable cardioverter-defibrillator (ICD) whereas those with the Brugada pattern do not; both types of patients follow behavioral preventive measures and avoid proarrhythmic drugs (www.brugadadrugs.org).[3] Patients with UCDs, as well as those with BrS, have special needs and should be treated under GA only in specialized hospitals because the evidence about the anesthesiologic management and risk for those patients is limited and not definitive due to the low prevalence of such diseases.[1–3,8,9] Therefore, the patient described in the current report carried an undefined anesthesiologic risk associated with the risk of developing hyperammonemia and/or a fatal arrhythmia.[3,10] Hyperammonemia occurs in UCDs because of the rise in protein catabolism caused by the stress response to dehydration, peripartum, blood loss, chemotherapy, steroids, trauma, infections, fever, fasting, surgery, and/or GA.[2,7,8,10] The precipitating factors of fatal arrhythmia in BrS may be stress, electrolytic anomalies, fever, and autonomous nervous system imbalances in favor of parasympathetic tone (e.g., fear, abundant meals).[3] Therefore, the authors focused on managing ammonia, limiting the perioperative stress, and administering GA by using drugs in line with recommendations for both citrullinemia and BrS (Table 1). During the perioperative period, ammonia can be managed by maintaining a low-protein diet with a high content of carbohydrates, arginine and other amino acid supplementation, and administration of scavengers of ammonia (e.g., sodium benzoate, sodium phenylacetate, sodium phenylbutyrate) in compliance with the recommendations about fasting before GA and avoidance of postoperative large meals due to the BrS.[7,8,10] Moreover, the measurement of ammonia is mandatory in citrullinemia patients before and after surgery, and hemodialysis should be considered for those who carry ammonia values between 250 and 500 μmol/L, whereas it is mandatory for those who have concentrations above 500 μmol/L.[1,10] Perioperatively, the supplementation of dextrose 5% in normal saline or 10% in water can limit protein catabolism.[1,2] Anyway, multiple intraoperative measurements of glucose levels are warmly suggested to avoid hyperglycemia, osmotic diuresis, and fluid loss.[1,8] Limitation of perioperative stress was crucial to limit the rise in catecholamine and cortisol concentrations and then prevent both hyperammonemia and fatal arrhythmias.[2,3,8] The authors administered dexamethasone to avoid the vomit-induced parasympathetic overstimulation, which could trigger arrhythmias.[3] Before the induction of GA, stress due to anxiety can be managed by the administration of intravenous benzodiazepines, such as midazolam.[3] Several authors used dexmedetomidine, an α2 adrenergic agonist, to reduce the stress response.[2,10] Choi et al. successfully used dexmedetomidine for sedation (with regional anesthesia) in a 48-year-old drug abuser with Type 2 citrullinemia who underwent two consecutive urologic procedures within 20 days.[10] Patel et al. described the use of dexmedetomidine for the maintenance of GA in a 16-year-old girl with Type 1 citrullinemia, spastic quadriparesis, and refractory epilepsy. Patel et al. induced GA by using 3 mg/kg of propofol and 1 mg/kg rocuronium.[2] During the first half of the surgery, they maintained GA by using sevoflurane (2.2%). During the second half of the surgery, they titrated down sevoflurane and maintained GA by using dexmedetomidine (0.5 μg/kg/h). At the end of the surgery, the anesthetist reduced dexmedetomidine to 0.1 μg/kg/h and reversed GA by administering 0.03 mg/kg of neostigmine and 0.01 mg/kg of glycopyrrolate. The patient was discharged uneventfully on the same day.[2] Dexmedetomidine, as well as the other α2 agonists (e.g., clonidine), intensifies the vagal stimulation of the heart and is avoided in BrS even if there is no definitive evidence of complications.[2,3] Adequate analgesia by local anesthesia and postoperative administration of analgesics is also important for the reduction of stress caused by pain.[3] Local anesthetic agents are antiarrhythmic drugs of the Ib class and can trigger arrhythmias in BrS patients.

TABLE 1 Main perioperative interventions for safe administration of general anesthesia in a patient with citrullinemia and Brugada pattern.

Perioperative Management of Ammonia **Measurement of ammonia and glycemia is mandatory before and after surgery**		
Preoperative and postoperative diet	**Prevention of protein catabolism**	**Essential drugs and supplementations**
Preoperative fasting	Intraoperative intravenous glucose and hydration supplementation	Arginine and amino acid supplementation
Low-protein and high-carbohydrate diet	Preoperative and/or postoperative hemodialysis to keep ammonia below 250 μmol/L	Administration of scavengers of ammonia (e.g., sodium benzoate, sodium phenylacetate, sodium phenylbutyrate)
Avoidance of large meals	Arginine and amino acid supplementation	
Perioperative limitation of stress		
Prevention of parasympathetic over-stimulation	**Adequate analgesia**	**Prevention of infections and immediate treatment of fever**
Anti-emetic prophylaxis (e.g., dexamethasone)	Local anesthesia by lidocaine with epinephrine	Acetaminophen to treat fever
Anxiolytic prophylaxis (e.g., midazolam)	Postoperative analgesia (e.g., morphine, ketorolac, diclofenac, and acetaminophen)	Antibiotic prophylaxis
GA Administration in Line with UCDs and BrS Recommendations		
Intraoperative monitoring and early defibrillation	**Recommended drugs**	**Postoperative monitoring**
Continuous ECG monitoring	Volatile anesthetics	Keep the ECG monitoring for 24 h after GA
Apply external defibrillator pads	Intravenous hypnotics (midazolam, thiopental, single bolus of propofol) Nondepolarizing neuromuscular blockers OR Depolarizing neuromuscular blockers (avoiding rapid multiple bolus administration)	Repeat measurement of ammonia
Monitoring of the depth of anesthesia (e.g., BIS, entropy)	Sugammadex	Overnight observation

Note: The perioperative management of ammonia, limitation of stress, and administration of drugs tolerated in Type 1 citrullinemia and Brugada pattern are the aims of the protocol for GA in a patient with those diseases, to prevent hyperammonemia and fatal arrhythmias as complications.

Abbreviations: BIS, bispectral index; BrS, Brugada syndrome; ECG, electrocardiogram; GA, general anesthesia; UCDs, urea cycle disorders.

Therefore, the use and dosages should be limited in those patients. Lidocaine formulations with vasoconstrictors are the local anesthetics preferable in BrS patients because of the fast onset, the fast recovery, and the rapid metabolization that concur to have low plasma levels thus avoiding cardiac effects. The individual response of BrS patients is unpredictable. Many authors described transient ST-tract elevations after the administration of lidocaine.[3,9] In addition, the Type 1 Brugada pattern or the worsening of the ST-tract elevation may occur because of the drugs administered during anesthesia, surgery, or invasive procedures. After the discontinuation of the trigger drugs, the electrocardiographic abnormalities resolve in most cases, as in the current.[3,9] Morphine, ketorolac, diclofenac, and acetaminophen have been used postoperatively as analgesic agents in both citrullinemia and BrS without adverse events.[2,3,9] Moreover, acetaminophen is a life-saving drug in case of fever in both patients with BrS and/or UCDs since hyperthermia could trigger both hyperammonemia and fatal arrhythmias.[2,3,5,10] Acetaminophen should be used with caution in citrullinemia patients who also carry liver damage.[2] The administration of GA to the current patient was mostly influenced by guidelines regarding BrS because those who carry the pattern need to be managed as those with the syndrome.[3,5] Therefore, the authors started the ECG continuous recording before anesthesia and applied the external defibrillator pads to be ready for early defibrillation of fatal arrhythmias that could occur during the perioperative period.[3,5] Monitoring the depth of anesthesia is also recommended in BrS to provide adequate analgesia and the authors successfully used the entropy measurement as a valid alternative to the bispectral

index (BIS) which was suggested for this kind of patient.[3] Volatile anesthetics, such as isoflurane and sevoflurane, can be used in both BrS and UCDs but should be avoided in patients with concomitant long QT syndrome because those drugs prolong the QT-segment.[3] Among intravenous hypnotics, the use of a single bolus of propofol (e.g., 1.5 mg/kg) over a few seconds is currently recommended to induce GA in BrS patients even if may induce transient type 1 pattern with low ST-segment elevation, whereas high dosages (>5 mg/kg/h) in continuous infusion are still to avoid because of the risk of inducing the Propofol Infusion Syndrome (PRIS).[3,5,6,9] Many authors documented the use of intravenous hypnotics such as midazolam and thiopental without complications.[3,9] Ketamine is a drug to be avoided in BrS because of documented toxicity.[3] Depolarizing neuromuscular blockers such as succinylcholine can be used in BrS, but rapid multiple bolus administrations should be avoided whereas nondepolarizing neuromuscular blockers (e.g., atracurium, vecuronium) showed no adverse effects.[3,5] Among reversal agents of neuromuscular blockers, sugammadex is safe because has low cardiovascular effects.[3] During the postoperative period, ECG should be monitored because BrS-related arrhythmias occur commonly during such a period; in addition, an overnight observation with frequent measurements of ammonia is recommended in patients with UCDs.[3,8] Anyway, the authors discharged the current patient a few hours after the surgery because of the low risk of SCD, the brief lasting of GA, and the patient regained consciousness without problems. Gharavifard et al. described the delayed awakening of a 3.5-year-old patient with Type 1 citrullinemia after 4 h under GA performed for dental procedures.[1] They used sevoflurane (8%) and N_2O/O_2 (60%/40%) via a face mask to set up venous access and then induced GA by using propofol (2 mg/kg), fentanyl (3 μg/kg), and cisatracurium (0.1 mg/kg), whereas postoperative pain was treated by using morphine (100 μg/kg). The unexpected delay in regaining consciousness lasted 4 h along with ataxia and difficulty in supporting head posture, then the child was admitted to the pediatric intensive care unit and discharged 10 days later.[1] Gharavifard et al. did not find a clear explanation for such complications, which could be the result of multiple contributions by factors such as the rise of both glycemia and ammonia, the effect of morphine, and the side effects of anesthetic agents.[1] Delay in awakening can be also avoided in citrullinemia patients by using a lower dosage of anesthetic agents in association with renal and hepatic blood flow reduction.[2] Generalizability is the main limitation of the current study because the patient carries a concomitant Brugada pattern and Type 1 citrullinemia, which is a rare occurrence. Further studies should focus on the update of the prevalence of the Brugada pattern in UCDs, the research of genetic mutations shared by those diseases, and the application of the study protocol in other patients.

4 | CONCLUSION

In the authors' experience, GA was successfully administered in the study patient because of the perioperative management of ammonia, limitation of the stress, and administration of drugs tolerated in Type 1 citrullinemia and Brugada pattern. In addition, such measures also prevented hyperammonemia and fatal arrhythmias as complications.

AUTHOR CONTRIBUTIONS

Fabio Dell'Olio: Writing – original draft. **Pantaleo Lorusso:** Writing – original draft. **Rosaria Arianna Siciliani:** Writing – review and editing. **Maria Massaro:** Conceptualization. **Giuseppe Barile:** Writing – review and editing. **Angela Tempesta:** Supervision. **Salvatore Grasso:** Project administration. **Gianfranco Favia:** Conceptualization. **Luisa Limongelli:** Project administration.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no conflict of interest.

DATA AVAILABILITY STATEMENT

Data available on request due to privacy/ethical restrictions

CONSENT

The mother of the study patient provided written informed consent for the surgical procedure and the publication.

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REFERENCES

1. Gharavifard M, Sabzevari A, Eslami R. Anesthetic management in a child with citrullinemia: a case report. *Anesthesiol Pain Med*. 2014;4(3):e21791. doi:10.5812/aapm.21791
2. Patel H, Kim J, Huncke TK. General anesthesia in a patient with citrullinemia using Precedex as an adjunct to prevent delayed emergence. *J Clin Anesth*. 2016;33:403-405. doi:10.1016/j.jclinane.2016.04.045
3. Espinosa Á, Ripollés-Melchor J, Brugada R, et al. Brugada syndrome: anesthetic considerations and management algorithm. *Minerva Anestesiol*. 2019;85(2):173-188. doi:10.23736/S0375-9393.18.13170-1
4. Dell'Olio F, Lorusso P, Barile G, Favia G. Brugada syndrome updated risk assessment and perioperative Management in Oral Surgery: a case series. *J Oral Maxillofac Surg*. 2021;79(11):2269.e1-2269.e11. doi:10.1016/j.joms.2021.07.012
5. Ciconte G, Santinelli V, Brugada J, et al. General anesthesia attenuates Brugada syndrome phenotype expression: Clinical implications from a prospective Clinical trial. *JACC Clin Electrophysiol*. 2018;4(4):518-530. doi:10.1016/j.jacep.2017.11.013
6. Goraksha S, Bidaye S, Gajendragadkar S, Bapat J, Butani M. General anaesthesia for insertion of an automated implantable cardioverter defibrillator in a child with Brugada and autism. *Indian J Anaesth*. 2010;54(6):562-564. doi:10.4103/0019-5049.72648
7. Häberle J, Burlina A, Chakrapani A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders: first revision. *J Inherit Metab Dis*. 2019;42(6):1192-1230. doi:10.1002/jimd.12100
8. Yeoh C, Teng H, Jackson J, et al. Metabolic disorders and anesthesia. *Curr Anesthesiol Rep*. 2019;9(3):340-359. doi:10.1007/s40140-019-00345-w
9. Kloesel B, Ackerman MJ, Sprung J, Narr BJ, Weingarten TN. Anesthetic management of patients with Brugada syndrome: a case series and literature review. *Can J Anaesth*. 2011;58(9):824-836. doi:10.1007/s12630-011-9546-y
10. Choi JJ, Kim HS, Lee KC, Shin Y, Jo YY. Anesthetic experience of an adult male with citrullinemia type II: a case report. *BMC Anesthesiol*. 2016;16(1):92. doi:10.1186/s12871-016-0253-7

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