

Peri-operative Management of a Child with Maple Syrup Urine Disease in a Non-tertiary Paediatric Hospital

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Abstract

Introduction: Maple Syrup Urine Disease (MSUD) is a rare, autosomal recessive aminoacidopathy. We report the peri-operative management of an eleven year old girl, with well controlled MSUD who presented to a non-paediatric, tertiary hospital for a non-emergency procedure, under general anaesthetic.

Case Presentation: An 11 year old girl presented to the Emergency Department with a history of nasal bone trauma. She was admitted for a manipulation of nasal bones (MNB) under general anaesthetic (GA). She had a history of MSUD diagnosed on day three of life. The patient was neurologically intact and had leucine levels within the accepted range prior to her surgery. In times of illness her leucine levels were generally well controlled.

The patient was admitted the night prior to surgery following her normal evening meal. Her urine was checked for ketones on admission and this was negative. Overnight carbohydrate management was instituted. Her surgery was performed first on the theatre list. The procedure lasted 14 min and was uneventful. Oral intake was recommenced at 10.00 am. Her urine was again checked for ketones post-operatively and was negative. The patient was well over the weekend and her leucine level post-surgery were within normal limits.

Discussion: MSUD is a rare disease. There is limited experience of the conduct of anaesthesia in these patients outside of specialist paediatric centers. Most reports advised transfer to a tertiary paediatric center. We demonstrated that low risk surgery can be carried out safely in these patients.

Keywords: Peri-operative management; Maple syrup urine disease; Anaesthesia in maple syrup urine disease

Introduction

Maple Syrup Urine Disease (MSUD) is a rare, autosomal recessive aminoacidopathy resulting from an enzyme defect in the catabolic pathway of leucine, isoleucine and valine [1,3]. Accumulation of these branched-chain amino acids (BCAA) and their metabolites leads to progressive neurodegeneration. Even in patients with good dietary control, inter-current stressful situations such as illness, trauma or surgery, can lead to high leucine levels with neurological sequelae. We report the peri-operative management of an eleven year old girl, with well controlled MSUD who presented to a non-paediatric, tertiary hospital for a non-emergency procedure, under general anaesthetic.

Case Presentation

An 11 year old girl presented to the Emergency Department with a history of nasal bone trauma one week previously with subsequent deviation of the nasal bridge. She was admitted for a manipulation of nasal bones (MNB) under general anaesthetic (GA). She had a history of MSUD, diagnosed on day three of life when she presented with poor feeding and lethargy. Early detection of the disease with implementation of an appropriate low protein diet and regular leucine monitoring had resulted in the patient avoiding the progressive neurodegeneration associated with inadequately controlled disease. An

in-depth history from the patient and her father revealed that the patient was neurologically intact and had leucine levels within the accepted range for the previous five weeks. In the week of surgery the patients leucine level was 268. In times of illness her leucine levels were generally well controlled. She adhered to a strict diet avoiding prolonged fasting by having specific alarm set times for her daily meals with carbohydrate loading in stressful situations and closely monitored protein intake. We researched the condition extensively and liaised with a paediatric specialist in metabolic disorders as well as the patients' parents in order to determine the best plan for her anaesthetic management.

The patient was admitted the night prior to surgery following her normal evening meal. Her urine was checked for ketones on admission and this was negative. To avoid prolonged fasting she was given a carbohydrate load of 110 ml of Duocal[®] (Nutricia, North America) at 02.30 and was woken again at 06.30 to drink 200 ml of apple juice concentrate. Her urine was again checked for ketones at this point and again this was negative. Her surgery was performed first on the theatre list at 08.30. Intravenous (IV) access was established, a venous blood sample was sent and 300 ml of NaCl 0.9% IV was commenced. She underwent a gas induction with sevoflurane and air following pre-oxygenation. Her airway was secured with a size 3 LMA following 30 mcgs of fentanyl IV and she was allowed to breath spontaneously. Intra-operatively she received 1 g of paracetamol and 25 mg of diclofenac as PR analgesia as well as IV ondansetron 3 mg and IV dexamethasone 4.5 mg as anti-emetics. The procedure lasted 14 min

and her SpO₂, EtCO₂, EtSevoflurane, HR and BP were maintained within normal limits throughout. Her LMA was removed in the recovery room where she was alert and orientated with no complaints of pain or nausea prior to returning to the paediatric ward. Oral intake was recommenced at 10.00 with a glass of apple juice. Her urine was again checked for ketones when she passed urine on the ward post-operatively and was negative. She restarted normal diet at lunch time and on review by the consultant anaesthetist, prior to discharge that afternoon, the patient was well. She was sent home with urine dipstick test strips and both the patient and her father were instructed in how to use them. Instructions were also given to further carbohydrate load that day as the patient normally would if she were ill and to return to the hospital immediately if her urine tested positive for ketoacids or she became unable to tolerate oral diet. The patient was well over the weekend and her leucine level when checked three days post-surgery were within normal limits.

Discussion

As MSUD is a rare disease there is limited experience of the conduct of anaesthesia in these patients outside of specialist paediatric centres [1]. On review of the literature we found a paucity of information, much of which was dated [1-3]. Most reports advised transfer to a tertiary paediatric centre, routine arterial blood gas monitoring and administration of IV fluids throughout the fasting period [1,4].

In this case these measures seemed inappropriate and excessive. The nearest paediatric tertiary referral centre was a 280 km journey away and our patient's disease was well controlled with consistently normal leucine levels on her current diet, which included a 9 h fast each night.

In view of this, and the nature of her procedure, we undertook to perform her procedure in our hospital leading to the minimum disruption to her normal routine. As her disease was well controlled we determined to use urine dipstick monitoring, for ketoacids as a surrogate marker for acidosis, in place of frequent arterial blood gas (ABG) sampling. As she was habituated to a 9 h fast overnight, we did not administer preoperative IV fluids but instead allowed her a carbohydrate rich drink 2 h prior to her procedure. We ensured that at all times there was the facility to perform ABG sampling were her urine dipstick to prove positive for ketoacids or the patient unable to pass urine for testing.

The key points in the management of this case were ensuring 1) a well patient with normal pre-operative leucine levels, 2) adequate carbohydrate loading for the extra stress associated with a surgical procedure, 3) short procedure with avoidance of fluid shifts, 4) minimal fasting time and 5) a pain free patient with no nausea who could resume her normal dietary intake as soon as possible post-operatively.

References

1. Fuentes-Garcia D, Falcon-Arana L (2009) Perioperative management of a patient with maple syrup urine disease. *BJA* 102: 144-145.
2. Kahraman S, Ercan M, Akkuş O, Erçelen O, Erdem K, et al. (1996) Anaesthetic management in maple syrup urine disease. *Anaesthesia* 51: 575-578.
3. Strauss KA (2018) Maple syrup Urine Disease.
4. Haberstick P, Kindler CH, Schürch M (2010) Anaesthesia in patients with maple syrup urine disease. *Anaesthesist*. 59: 914-917.