Clinical Case Reports

Open Access

CASE REPORT

Anesthetic management of a patient with Edwards syndrome

Stephanie Mifsud, Maureen Bezzina & Simon Paris

Department of Anaesthesia, Mater Dei Hospital, Msida, Malta

Correspondence
Stephanie Mifsud, Department of Anaesthesia, Mater Dei Hospital, Msida, MSD 2090, Malta. Tel: +35625457251;
E-mail: mifsteph@gmail.com

Funding Information
The study was funded by departmental resources.

Received: 19 August 2015; Revised: 11 April 2016; Accepted: 6 May 2016

***Clinical Case Reports* 2016; 4(8): 740–742**

doi: 10.1002/ccr3.595

Key Clinical Message

The use of suxamethonium in our case was uneventful and despite craniofacial anomalies, airway management was straightforward. This case illustrates that pediatric patients with trisomy 18, presenting with potentially acute life-threatening conditions and requiring emergency major surgery can be managed successfully with a multidisciplinary approach.

Keywords

Airway management, anesthesia, Edwards syndrome, intestinal obstruction, pediatrics, trisomy 18.

Introduction

Edwards syndrome (trisomy 18), first described by Edwards et al. [1] in 1960, is the second most common autosomal trisomy, after trisomy 21. It consists of craniofacial anomalies, visceral defects, and delayed mental and motor development. The survival rate beyond the first year of life is 5–10%, with a live-birth prevalence estimated at 1:3,600–10,500 [2]. In Malta, the estimated prevalence is 3.26 per 10,000 births [3]. We report the anesthetic management in a patient with Edwards syndrome undergoing emergency laparotomy for acute intestinal obstruction.

Case Report

A 3-year, 6-month-old, 8.5 kg girl, diagnosed with trisomy 18 at birth, was admitted with brown-blood streaked vomiting and diarrhea. Previous admissions were attributed to gastro-oesophageal reflux disease (GORD) and treated medically. During this admission, the vomiting and abdominal distension did not resolve despite medical treatment and nasogastric tube. She also became febrile. Plain abdominal X-rays were highly suggestive of intestinal malrotation and she was booked for an emergency laparotomy.

A multidisciplinary meeting was held with both parents. The risks of her having general anesthesia and surgery, and the expectations of outcome were discussed prior to obtaining parental consent. She was classified ASA IV and had never had anesthesia. Of note preoperatively, her serum potassium was 2.46 mmol/L (3.5–5. 1 mmol/L). Thus, 10 mL (13 mmols) of 20% potassium chloride (KCL) was added to 500 mL of 5% dextrose in 0.45% normal saline at a rate of 35 mL/h (4 mL/kg/h) intravenously (IV). Subsequently, the potassium supplementation was increased to 20 mL (26 mmols) of 20% KCL in 500 mL of maintenance fluid. However, after an hour of this IV potassium replacement, it was decided to proceed with surgery to minimize potentially extensive bowel necrosis. Her echocardiogram showed partial ventricular septal defect (VSD), patent foramen ovale (PFO), and large patent ductus arteriosus (PDA) shunting bidirectionally, with secondary pulmonary hypertension and right ventricular hypertrophy and dilation.

Standard noninvasive monitoring was set up. The team was briefed with regards to the airway management plan and in case of a difficult intubation, the pediatric difficult airway guidelines by the difficult airway society (DAS) would be followed [4]. It was ensured that the pediatric intubation trolley contained all the necessary equipment. Following preoxygenation, rapid sequence induction

(RSI) was performed using 5 mcg (1–2 mcg/kg) fentanyl, 30 mg thiopentone (3.5 mg/kg), and 20 mg suxamethonium (1–2 mg/kg) IV, via an already sited 22G cannula. Laryngoscopy was performed using a Mackintosh blade 1 and it was a Cormack-Lehane grade I. Intubation was relatively easy and a size 5 uncuffed endotracheal tube was inserted over a gum elastic bougie. Anesthesia was maintained with sevoflurane (mean end-tidal of 2.1) and 50:50% oxygen in air using pressure-controlled ventilation with Pinsp 12 cm H_20, PEEP 5 cm H_2O, respiratory rate of 16 bpm and Pmax was 17 cm H_2O. After induction of anesthesia, a right radial arterial cannula and right femoral vein 13 cm double lumen central line were inserted under strict aseptic technique. An epidural catheter was sited at L2/L3 using an 18G Tuohy needle with a depth to space of 1.5 cm and mark at skin of 5 cm.

Antibiotic cover at induction was 60 mg metronidazole (7.5 mg/kg), 250 mg amoxicillin (30 mg/kg), and 40 mg gentamicin (7 mg/kg loading dose) IV. The surgery lasted 3 h. Ileocecal intussusception with a Meckel's diverticulum as lead point was found and reduced. An appendicectomy was also done. An episode of bradycardia (75 bpm) intraoperatively was corrected with 200 mcg of atropine (20 mcg/kg) IV. Analgesia intraoperatively was maintained with 10 mcg of fentanyl IV and a total of 10 mL of 0.2% bupivacaine and 2 mcg/mL fentanyl via epidural catheter. Muscle relaxation was maintained with 10 mg of atracurium (0.3 mg/kg) IV, at 1 and 2 h into the operation, respectively. Urine output intraoperatively was 12 mL/h and total drainage from the NGT was 200 mL. Fluid replacement intraoperatively was with Hartmann's solution at 40 mL/h and 5% dextrose in 0.45% normal saline with 20 mmol of 20% KCL at 35 mL/h. Potassium was additionally supplemented with 4 mmol (1.5 mL) KCL in 50 mL normal saline over 1 h in view of persistent low potassium levels (2.8 mmol/L and 2.58 mmol/L) on arterial blood gases (ABGs) intraoperatively. Fluids were administered through an IV enFlow fluid warmer system. A temperature of 37.5°C was maintained throughout the whole operation.

Postoperatively, the child was transferred, intubated and ventilated, to NPICU for monitoring of biochemical and hemodynamic stability. An epidural infusion consisting of 0.1% bupivacaine and 2 mcg/mL fentanyl was started at 3 mL/h for postoperative pain relief. In NPICU, an IV midazolam infusion (24 mg in 50 mL of 0.9% NaCl) was started at 2 mL/h for sedation and maintenance fluids were 5% dextrose in 0.45% saline with 10 mmol (10 mL) of 20% KCL at 31 mL/h. She was extubated the following day and transferred to a surgical ward 4 days later. She had no complications and was discharged home 9 days postoperatively.

Discussion

Airway management considerations include dolichocephaly, micrognathia, small mouth and neck which may make mask ventilation and intubation difficult. Thus, it is essential to prepare for a difficult intubation. Alternatively, Bailey and Ghung [5] successfully used a laryngeal mask airway in a 3-year-old girl undergoing bilateral myringotomies and grommet insertion. In our case, risk of aspiration was minimized with an IV RSI with suxamethonium. Although, rocuronium and sugammadex are available in our institution, experience with their use in critically ill children is still limited.

Matsuda et al. [6] reported a boy with trisomy 18, undergoing testicular fixation, who developed muscular rigidity following suxamethonium administration, making endotracheal intubation impossible. Intraoperatively, he developed a temperature of 38.4°C and elevated serum creatinine phosphokinase. However, this is the only documented case, and thus, there is no clear link between trisomy 18 and malignant hyperthermia. In our case, the use of suxamethonium was uneventful.

Structural heart defects occur in over 90% of infants with Edwards syndrome. The most common cardiac lesions are atrial and ventricular septal defects (ASD), VSD, PDA, and polyvalvular disease [2]. Courreges, et al. [7] in their management of a 7-year-old girl with Edwards syndrome undergoing a Cohen procedure, suggest that these patients can be considered as cardiac patients. Thus, it is imperative that hypercapnia and hypoxia are prevented in pulmonary hypertension to avoid potential reversal of the bidirectional shunt. Other anesthetic considerations are maintenance of hemodynamic stability and homeostasis between systemic and pulmonary circulations [2]. The use of volatile inhalational agents with the least cardiac depressant effects, such as sevoflurane, is the sensible option. Fentanyl, utilized as IV and epidural analgesia, is considered as the first drug of choice in children with pulmonary hypertension [8].

Courreges et al. [7] performed an inhalational induction with sevoflurane which took longer (8 minutes) than usual but this was attributed to a change in ventilation/perfusion ratio caused by the cardiac disease. Arun et al. [2], report using an inhalational induction due to problems with securing venous access in their management of a 13-day-old girl with trisomy 18 undergoing closure of a PDA and pulmonary artery banding. However, they reported an uneventful and regular duration of induction with sevoflurane. In our case, an IV RSI was performed utilizing peripheral venous access established preoperatively.

In conclusion, anesthetic perioperative management of patients with Edwards syndrome is an uncommon occurrence and presents a number of challenges for the

anesthetist. There are only seven reported cases in the literature on the anesthetic management of patients with this syndrome and as such no definitive anesthetic protocol exists. Thus, the publication of additional case reports will contribute to augmenting further our knowledge in the anesthetic management of this condition. This case describes the management of an emergency surgery as opposed to the other reports of elective surgeries, which needed to be undertaken outside normal operating theater time, on a weekend.

Ethics Approval

None necessary.

Conflict of Interest

No conflicts of interest declared.

References

1. Edwards, J. H., D. G. Harnden, A. Cameron, V. M. Crosse, O. H. Wolff, et al. 1960. A new trisomic syndrome. Lancet 1:787–790.
2. Arun, O., B. Oc, M. Oc, and A. Duman. 2014. Anesthetic management of newborn with trisomy 18 undergoing closure of patent ductus arteriosus and pulmonary artery banding. Cardiovasc. J. Afr. 25:e10–e12.
3. Detailed breakdown of anomalies registered and prevalence 1993-2012. [Online]. Available at: https://health.gov.mt/en/dhir/Pages/Registries/birthdefects.aspx. [Accessed 22nd May 2015].
4. Difficult airway society. *Paediatric difficult airway guidelines*. Available at: http://www.das.uk.com/guidelines/paediatric-difficult-airway-guidelines [Accessed 23rd March 2016].
5. Bailey, C., and R. Ghung. 1992. Use of the laryngeal mask airway in a patient with Edwards' syndrome. Anaesthesia 47:713.
6. Matsuda, H., S. Kaseno, Y. Gotoh, K. Furukawa, and K. Imanaka. 1983. Muscle rigidity caused by succinylcholine in Edwards' syndrome. Masui 32:125–128.
7. Courreges, P., R. Nieuviarts, and D. Lecoutre. 2003. Anaesthestic management for edward's syndrome. Paediatr Anaesth. 13:267–269.
8. Shew, S. B., T. H. Keshen, N. L. Glass, F. Jahoor, and T. Jaksic. 2000. Ligation of a patent ductus arteriosus under fentanyl anesthesia improves protein metabolism in premature neonates. J. Pediatr. Surg. 35:1277–1281.