Anesthetic management of a patient with Edwards syndrome

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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.

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.
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.

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