ANESTHETIC MANAGEMENT FOR TETHERED CORD SYNDROME IN A CHILD WITH EMANUEL SYNDROME

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ABSTRACT Background: Emanuel syndrome (ES) is a rare hereditary disorder characterized by growth and mental retardation, congenital malformations, and craniofacial anomalies. There is limited data on the anaesthetic management of this syndrome, all of which come from case reports. **Report:** A 7-year-old girl presented with urinary incontinence caused by tethered cord syndrome. Laminoplasty plus transection of the tight filum terminale to release the tethered conus medullaris under total intravenous anaesthesia was performed. **Discussion:** Anesthesia management of patients with ES requires special attention due to the airway difficulties associated with micrognathia and palate anomalies and accompanying congenital malformations that primarily affect cardiovascular and neurologic systems. Monitoring the cardiac parameters and neurological signs during the surgery is of great importance for patient safety. **Conclusion:** This case report is the first demonstrating total intravenous anaesthesia in patients with ES, and may help to guide anesthesiologists in the anaesthetic management of these patients.

KEYWORDS Airway management, total intravenous anaesthesia, difficult intubation, Emanuel syndrome

Introduction

Emanuel syndrome (ES) or supernumerary der(22) syndrome is a recessive hereditary disorder caused by chromosome 11;22 translocation [1]. It is an uncommon entity with a frequency of 1:110.000 live births. It is characterized by developmental retardation, seizure, congenital heart disease, genitourinary malformations, tracheomalacia, and various craniofacial anomalies including microcephaly, preauricular tags, cleft or high-arched palate, and micrognathia [2,3].

There is limited data on the anaesthetic management of these patients, all of which come from case reports [4-6]. However, these patients are at high anaesthesia risk due to coexisting disorders and anatomical anomalies. Here, anaesthetic management for tethered cord syndrome in a child with ES was presented.

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Case presentation

A 7-year-old girl, 13 kg in weight, presented with lumbar pain and urinary incontinence. She was diagnosed with ES based on chromosomal study within the first months of birth. She had growth retardation, aphasia, mild pulmonary and tricuspid insufficiencies, unilateral renal agenesis, and paresthesia. She had undergone multiple operations including the correction of hip dislocation, balloon valvuloplasty for pulmonary stenosis, and palatoplasty for cleft palate. Lumbar five vertebrae laminoplasty plus transection of the tight filum terminale under general anaesthesia was scheduled for the treatment of tethered cord syndrome.

Her routine preoperative tests were normal. Airway assessment was consistent with mallampatti class 2. She was classified as American Society of Anesthesiologists score 4.

On the operation day, monitoring included electrocardiography, noninvasive blood pressure and pulse oximetry. After preoxygenation, anesthesia was induced by remifentanil (1 μg/kg), lidocaine (0.5 mg/kg), and propofol (4 mg/kg) with air:oxygen (50%:50%) at 4 L/min total gas flows, with rocuronium (0.6 mg/kg). Intubation was then performed using two no video laryngoscope blade, without any difficulty. Radial artery and internal jugular vein catheters were inserted to closely monitor the

hemodynamic parameters. For maintenance, total intravenous anaesthesia (TIVA), remifentanil (0.2 μg/kg/h) and propofol (4 mg/kg/h), and air: oxygen (50%:50%) at 4 L/min total gas flows, was preferred since the surgeons used neuromonitorization to prevent neural damage during dissection of the surgical area. The operation was performed in a prone position and lasted two hours without any significant complication. She was uneventfully discharged on the fifth postoperative day.

Discussion

The patients with ES usually need surgical intervention at some time of their life, because of the coexisting congenital anomalies. Similar to our patient, all patients in a large series had at least one congenital anomaly, most of which were cleft palate and heart malformations.[7] Anesthetic management carries several risks in these patients. First of all, these patients are generally in the pediatric population whose anaesthesia management has its challenges. Mental and developmental retardations also affect all steps of anaesthesia, particularly choice and doses of medication.

Airway management is a significant concern for anesthesiologists because of the craniofacial anomalies. Micrognathia and cleft or high-arched palate are common and may lead to difficult intubation. Therefore, an emergency tracheotomy set should be available during the intubation. Nishinarita et al. reported two cases of ES in their work and experienced difficult intubation in one who had micrognathia [5]. In another study, the patient had a tracheotomy due to micrognathia with breathing difficulty [6]. In our case, no difficulty in the intubation was observed because the patient had no significant facial anomaly. She had also had surgery previously for cleft palate, without any intubation difficulty.

The other risk factor for anaesthesia management in patients with ES is the presence of system malformations. Congenital heart disease is among the most frequent anomalies in these patients [7,8]. Therefore, detailed preoperative evaluation is an important step of anesthesia management. Hemodynamic parameters should also be closely followed up during the perioperative period since hemodynamic responses to the anaesthetic agents may cause significant circulatory disturbances [6]. Depending on the complexity of the surgical procedure, central venous catheterization may also be required.

In the present case, propofol was the anaesthetic agent chosen due to the induction of anaesthesia. Tsukamoto reported an increased hemodynamic response during the first minutes of induction, which might be associated with agitation by sevoflurane, administration of atropine, or movement of the head [6]. Of note, no acute hemodynamic disturbance occurred in our patient. However, it should be kept in mind that sevoflurane may cause seizure-like activity, especially in combination with hyperventilation [2,6]. Therefore, use of this inhalation agent should be avoided in patients with seizure or other related neurological disorders. Maintenance of end-tidal carbon dioxide within the normal limits may help to prevent such adverse effects of sevoflurane. Propofol and isoflurane can also be used to suppress seizure-like effects [7]. Although our patient had no seizure or other neurological dysfunction, we did not use sevoflurane to avoid seizures and seizure-like activity.

Conclusion

This case report is the first demonstrating total intravenous anaesthesia in patients with ES and may help to guide anesthesiologists in the anaesthetic management of these patients. High clinical attention should be paid to preoperative patient preparation, risk of difficult intubation mainly associated with micrognathia and cleft or high-arched palate, and closed monitorization of cardiac parameters and neurological signs in such patients.

Disclosure Statement

There were no financial support or relationships between the authors and any organization or professional bodies that could pose any conflict of interests.

Conflict of Interest

Written informed consent obtained from the patient for publication of this case report and any accompanying images.

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