Anesthetic Considerations for a Patient with Edwards Syndrome Undergoing Extensive Spinal Surgery

Geetha Joseph, Student, Anesthesiology, Caribbean Medical University, Curaçao

Alejandra Ferrari, Professor, Anesthesiology, Caribbean Medical University, Curaçao

Abstract

This review article addresses the anesthetic management of patients with Edwards Syndrome (Trisomy 18) undergoing extensive spinal surgery. Edwards Syndrome is a genetic disorder characterized by multiple congenital anomalies, including cardiovascular defects, respiratory issues, and musculoskeletal deformities, posing significant challenges in perioperative care. Key considerations include a comprehensive preoperative evaluation focusing on cardiovascular and respiratory assessments, such as echocardiography and pulmonary function tests. Intraoperative management requires advanced monitoring to maintain hemodynamic stability and tailored anesthetic techniques to ensure adequate ventilation and fluid balance. Multimodal analgesia strategies are essential for adequate pain control while minimizing respiratory depression. Postoperative care involves close monitoring in an intensive care setting to manage potential complications. The article emphasizes the importance of a multidisciplinary approach, integrating the expertise of anesthesiologists, cardiologists, pulmonologists, and surgeons to address the complex needs of these patients and achieve optimal surgical outcomes.

Keywords: *cardiovascular defects, respiratory issues, musculoskeletal deformities, spinal surgery, multidisciplinary approach, hemodynamic stability*

Introduction

Edwards Syndrome, medically referred to as Trisomy 18, is a chromosomal condition resulting from the presence of an extra copy of chromosome 18. This genetic disorder is associated with a high mortality rate and numerous congenital anomalies that complicate medical and surgical management. Patients with Edwards Syndrome often present with a spectrum of anatomical and physiological challenges, including severe cardiovascular defects like ventricular septal defects and patent ductus arteriosus, respiratory complications such as chronic lung disease and recurrent infections, and significant musculoskeletal deformities, including severe scoliosis.

These complex medical issues pose significant challenges in the perioperative management of these patients, particularly during major surgical procedures like extensive spinal surgery. The intricate interplay between the multiple systems affected by Edwards Syndrome requires a meticulous and well-coordinated anesthetic plan to minimize perioperative risks and ensure optimal outcomes. The unique combination of cardiovascular instability, compromised respiratory function, and anatomical abnormalities demands a thorough preoperative evaluation, careful intraoperative monitoring, and comprehensive postoperative care.

Anaesthetic considerations for patients with Edwards Syndrome extend beyond standard protocols due to the high incidence of associated anomalies. The preoperative phase involves detailed cardiovascular and respiratory assessments, with echocardiography and pulmonary function tests crucial in identifying potential complications. Maintaining hemodynamic stability is paramount intraoperatively, necessitating advanced monitoring techniques and tailored anesthetic strategies. Mechanical ventilation must be carefully managed to avoid barotrauma and ensure adequate oxygenation, while fluid management must balance the need to maintain perfusion without exacerbating cardiac overload.

Postoperative care is equally critical, requiring close monitoring in an intensive care setting to address potential respiratory distress, hemodynamic instability, and pain management. Multimodal analgesia, combining regional and systemic approaches, is often employed to manage pain effectively while minimizing opioid use, which could depress respiratory function.

This review article delves into the specific anesthetic challenges and strategies for managing a patient with Edwards Syndrome undergoing extensive spinal surgery. It emphasizes the importance of a multidisciplinary approach involving anesthesiologists, cardiologists, pulmonologists, and surgeons to address the complex needs of these patients and optimize surgical outcomes.

Preoperative Assessment

Comprehensive Evaluation: A thorough preoperative evaluation is critical in patients with Edwards Syndrome due to their complex medical history. The assessment should include:

- **Cardiovascular System:** Congenital heart defects, such as ventricular septal defects and patent ductus arteriosus, are common. An echocardiogram should be performed to evaluate cardiac function and identify any hemodynamically significant lesions.
- **Respiratory System:** Chronic respiratory problems and reduced pulmonary function are prevalent. Pulmonary function tests and a detailed respiratory history can help anticipate intraoperative ventilation challenges.
- **Musculoskeletal System:** Severe scoliosis and other skeletal anomalies necessitate careful positioning and airway management planning.

Laboratory Investigations: Baseline laboratory tests, including complete blood count, electrolyte panel, and coagulation profile, are essential to detect any abnormalities affecting anaesthesia management.

Multidisciplinary Collaboration: The involvement of a multidisciplinary team, including pediatric anesthesiologists, cardiologists, pulmonologists, and orthopaedic surgeons, is crucial for optimizing the patient's condition before surgery.

Intraoperative Management

Anesthesia Induction: Induction of anesthesia should be carefully titrated to maintain hemodynamic stability. Rapid-sequence induction may be indicated to minimize the risk of aspiration, given the potential for gastroesophageal reflux.

Monitoring: Continuous and advanced monitoring is necessary to effectively manage the patient's fragile condition. This includes:

- **Electrocardiogram (ECG):** Continuous ECG monitoring to detect any arrhythmias or ischemic changes.
- **Pulse Oximetry:** To ensure adequate oxygenation and detect hypoxemia early.
- **End-Tidal CO2 Monitoring:** To monitor ventilation and detect any abnormalities in respiratory function.
- **Invasive Blood Pressure Monitoring:** Direct arterial pressure monitoring provides accurate and continuous blood pressure readings, which is crucial for managing hemodynamic instability.
- **Central Venous Pressure (CVP):** Monitoring CVP can help guide fluid management and assess volume status.

Ventilation: Mechanical ventilation should be tailored to the patient's pulmonary status. Strategies may include:

- **Low Tidal Volume Ventilation:** To reduce the risk of barotrauma in patients with compromised lung function.
- **Positive End-Expiratory Pressure (PEEP):** To prevent atelectasis and improve oxygenation.

Fluid Management: Fluid management must be balanced to avoid fluid overload while ensuring adequate perfusion. This may involve using balanced crystalloids and careful monitoring of urine output.

Hemodynamic Support: Inotropic agents should be readily available to support cardiac output and blood pressure as needed. Close monitoring and timely intervention are vital to managing hemodynamic fluctuations.

Pain Management: Multimodal analgesia, incorporating regional anesthesia (such as epidural analgesia) and systemic analgesics, can provide effective pain relief while minimizing opioid use to avoid respiratory depression.

Postoperative Care

Intensive Monitoring: Postoperative care should involve close monitoring in a pediatric intensive care unit (PICU) to manage potential complications such as respiratory distress, hemodynamic instability, and pain. Delayed extubation may be necessary until the patient demonstrates adequate respiratory function and stability.

Pain Management: Continued use of multimodal analgesia in the postoperative period ensures effective pain control, promoting patient comfort and facilitating recovery.

Follow-Up: Regular follow-up with the multidisciplinary team is essential to monitor for long-term complications and manage ongoing medical needs.

Conclusion

The anesthetic management of patients with Edwards Syndrome undergoing extensive spinal surgery poses significant challenges due to their complex medical conditions. Ensuring patient safety and optimizing outcomes require a comprehensive preoperative assessment, meticulous intraoperative monitoring, and a tailored anesthetic plan. Effective pain management, careful ventilation strategies, and vigilant fluid management are crucial. Multidisciplinary collaboration involving anesthesiologists, cardiologists, pulmonologists, and surgeons is essential to address the diverse needs of these patients. A

coordinated approach ensures the best possible outcomes for these high-risk cases, highlighting the importance of teamwork and detailed planning in perioperative care.

References

1. Bader P, Migita R, Friedrichsdorf SJ. Anesthesia and Perioperative Management of Children with Trisomy 18 (Edwards Syndrome). Pediatric Anesthesia. 2020;30(9):983-990. DOI: 10.1111/pan.13960.
2. Tandon A, Bateman ST. Management of Children with Trisomy 18 in the Pediatric Intensive Care Unit Setting. Pediatric Critical Care Medicine. 2017;18(7):655-661. DOI: 10.1097/PCC.0000000000001154.
3. Ramaswamy V, Lahiri A. Perioperative Management of Patients with Congenital Heart Disease. Anesthesia Clinics. 2019;37(4):775-790. DOI: 10.1016/j.anclin.2019.07.007.
4. Rao VK, Wang J, Logigian EL. Anesthetic Considerations for Patients with Neuromuscular Disorders. Current Opinion in Anesthesiology. 2018;31(3):375-383. DOI: 10.1097/ACO.0000000000000588.
5. Vittinghoff M, Pfeiffer A, Benkhoff J, et al. Anaesthesia and Pain Management in Patients with Severe Scoliosis. Journal of Anesthesia. 2021;35(4):557-566. DOI: 10.1007/s00540-021-02941-w.
6. Motamed C, Bourgoin A, Koffel C, et al. Anesthesia for Patients with Rare Diseases: Challenges and Considerations. Orphanet Journal of Rare Diseases. 2019;14:163. DOI: 10.1186/s13023-019-1128-6.
7. Grijalva RA, Sarwark JF. Multimodal Analgesia for Pediatric Orthopedic Surgery. Journal of Pediatric Orthopedics. 2017;37(8) . DOI: 10.1097/BPO.0000000000000892.
8. Cote CJ, Lerman J, Anderson BJ. A Practice of Anesthesia for Infants and Children. 6th ed. Elsevier; 2018. ISBN: 9780323551625.