Poland Syndrome And Pediatric Anesthesia - Case Report And Literature Review

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Citation

D Malhotraa, A Gerdak. *Poland Syndrome And Pediatric Anesthesia - Case Report And Literature Review*. The Internet Journal of Anesthesiology. 2023 Volume 42 Number 1.

DOI: 10.5580/IJA.56800

Abstract

Introduction

Poland syndrome is a rare condition characterized by pectoralis major muscle hypoplasia. There is usually a difference between thoracic deformities and syndactyly deformities, which requires chest reconstruction or syndactyly release. Anesthetic management of pediatric Poland syndrome has not yet been described.

Case Report

Herein, we present our anesthesia management of syndactly released fingers with a wrist split-thickness graft in an 11-month-old child with left-sided Poland Syndrome by total intravenous anesthesia with invasive ventilation, Perioperative procedures to ensure safe surgery.

Conclusion

The PS imposes specific anesthetic management and adequate monetarization to avoid complications related to the chest deformity.Preoperative evaluation, intraoperative care, anesthetic selection, and postoperative pain management should be considered. The anesthetic technique was total intravenous anesthesia to reduce the doses used and also it was performed to avoid Malignant Hyperthermia

INTRODUCTION

Poland Syndrome (PS) is a rare congenital condition. The estimated incidence of this problem is 1:30,000 to 1: 80,000 live births. [1] This pain is more common in men (female-to male ratio of 1:2 to 1:3) and usually occurs on the right side of the chest rather than the left side. kidneys and lungs. The most common feature of this syndrome is chest deformity. Ipsilateral hand abnormalities are the most important and may present as syndactyly or exodus. [2,3] This condition is also known as ipsilateral chest syndrome, limb/body wall defect, thoracic fissure, pectoral hypoplastic syndrome, or unilateral thoracic hand deformity, depending on the affected body. Pulmonary complications may develop in patients owing to the use of muscle relaxants for the induction and maintenance of anesthesia. In addition, these patients have a high risk of malignant hyperthermia during anesthesia. General anesthesia should be administered with caution because of the risk of malignant hyperthermia. In this report, we will discuss an 11-month-old female child with left Poland syndrome who did not have the pectoralis major muscle and absent 3rd-5th ribs, and planned syndactly release of the 2nd and 3rd fingers, with partial thickness graft from the wrist with paradoxical chest movement.

We also searched PubMed to review reports on anesthesia management in Poland syndrome and gathered perioperative standards to guarantee successful surgery.

CASE REPORT

An 11 month-old baby weighing 9.7 kg presented with left Poland syndrome, defective pectoralis major, missing 3-5 ribs, chest anomalies, and planned interphalangeal 2nd and 3rd finger release with split thickness graft from the wrist. On physical examination, there was a left-sided chest wall defect; absence of the pectoralis major and pectoralis minor; absent 3rd,4th,5th rib and syndactyly fingers of the left hand.

Paradoxical respiratory movements were observed upon deep inhalation and pulsations of the heart were visible through the defect. Echocardiography revealed levocardia and a small atrial septal defect. On the day of surgery, the patient was premedicated with buccal midazolam 0.5 mg/kg. The preoperative blood oxygen saturation was 98% and the heart rate was 102 beats/min. All precautions were taken to prevent malignant hyperthermia. The cold blanket was wrapped around the pillow, and the machine remained in standby. A designated anesthesia machine without inhalational agents and a new circuit was used, 26G IV cannula was inserted on the patient's dorsum hand.

Because of the decision to continue Total Intravenous Anesthesia, propofol (2 mg/kg) and fentanyl (2 mcg/kg) were administered for anesthesia induction. Intubation was performed using a 3.5 mm cuff ETT on the C-MAC. Anesthesia was maintained with propofol (3-4 mg/kg/hour) and remifentanil (0.3-0.5 mcg/kg/min). Intubation with C-MAC was graded as view 2. The patients' body temperatures and end-tidal carbon dioxide levels were monitored. After intubation, controlled ventilation (P 15 cmH2O, R 16 bpm, I:E 1:2) was administered, and end-tidal carbon dioxide (EtCO2) was maintained at approximately 37 mmHg. The nasopharyngeal temperature was monitored at 37 ºC. Warm the patient at 3 °C with a patient warmer (Warm Touch™ 5200) and warm the fluid from line IV. The total anesthesia time was 4 h. The patient was extubated in the operating room and was transferred to the recovery room. No complications occurred during this procedure. Intraoperative paracetamol, ketorolac, and morphine were administered.

The pathogenesis of PS remains unknown and several hypotheses have been proposed. The etiology of PS is hypoplasia of the ipsilateral subclavian artery or one of its branches in the seventh week of pregnancy, failure of blood supply to the embryo due to prenatal exposure to possible teratogens (cocaine, misoprostol or cigarettes), and vascular development. [4] It has also been reported that there is less than a 50% reduction in the size and velocity of the subclavian artery in PS patients. [5] This condition can cause abnormalities depending on the location of the blood flow. Although most reports have described sporadic cases of PS, the identification of families with inherited patterns suggests that PS may have a genetic component. [6,7]

DISCUSSION

Here, we present the anesthesia management of an 11-month-old patient with PS who underwent syndactic release using a split-thickness wrist graft. Patients with Poland syndrome show a variety of physical impairments that have multiple effects on pre-anesthesia management, particularly ventilation management, and the possibility of malignant hyperthermia. Before surgery, various tests are required to diagnose related diseases. In addition to basic hematology, chest radiography, arterial blood gas analysis, and echocardiography should be performed to evaluate cardiorespiratory abnormalities. A detailed airway examination is necessary to identify any difficulties. The presence of chest wall defects in these patients leads to drawing of the chest wall in that area during inspiration, and the opposite occurs when the patient expires, thereby causing paradoxical respiration. Under stress, spontaneous breathing can result in respiratory muscle fatigue and reduced respiratory muscle strength, leading to cardiopulmonary collapse. Additionally, the lung on the side of the thoracic deformity may be small or hypoplastic. Therefore, to avoid mismatches with spontaneous respiration, controlled positive-pressure ventilation is mandatory for these patients during the intraoperative period. To prevent worsening of respiratory embarrassment due to pain in the postoperative period, proper pain management using a multimodal technique is necessary in these patients. Hence, positive pressure ventilation was performed via an endotracheal tube during surgery. Multimodal analgesia with opioids and NSAIDS was administered intraoperatively along with a wrist block by the surgeon at the end of the procedure.

Second, owing to the congenital nature of the disease, which is characterized by defects in muscular development, difficulties are anticipated on laryngoscopy, and an optimal view of the glottis may not be obtained while securing the airway with an endotracheal device. [9] Airway management requires a primary plan and multiple backup plans. We were well equipped with our difficult airway cart prior to induction and successfully intubated the trachea of our patient using an oral styletted endotracheal tube with the help of the C-MAC. Malignant hyperthermia may have appeared in patients with Poland syndrome due to involvement of the musculoskeletal system. Therefore, an anesthesiologist is required to take preventive measures. Hence, during the management of this case, we did not expose the patient to halogenated inhalational agents; instead, we performed a non-triggering anesthetic technique with fentanyl, remifentanil, and propofol. Throughout the intraoperative period, continuous monitoring of body

temperature and EtCO2 monitoring was performed for the early detection of malignant hyperthermia.

CONCLUSION

Musculo skeletal abnormalities as in Poland syndrome are important in anaesthesia practice. [8] The possible complications such as malignant hyperthermia and hypoxia should be considered. Also we should avoid succinylcholine and inhalational agents to prevent malignant hyperthermia, should use controlled respiration to prevent hypoxia. There is no doubt that anesthesia management in patients with Poland syndrome is difficult. However, preoperative evaluation, operation planning and proper preparation of anesthesia equipment are essential for successful anesthesia in these patients. Anesthesiologists should be aware of the complications that occur especially in patients with Poland syndrome and should be careful.

Statements Acknowledgement.

We thank the OT Anesthesia Technicians, especially Mr. AS for his assistance with case management.

Statement of Ethics.

Written informed consent from parents (mother) was taken for publishing case report but refused to print any pictures. No ethics approval required as per institutional ethics committee, Research Medical affairs.

Disclosure Statement.

The authors have no potential conflicts of interest to disclose.

Funding Sources: None.

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