CASE REPORT | PEER REVIEWED | OPEN ACCESS

Total intravenous anesthesia in a patient with Poland syndrome submitted to video-laparoscopic inguinal hernioplasty: A case report

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ABSTRACT

Introduction: Poland syndrome (PS) is a rare disease characterized by hypoplasia/agenesis of the pectoralis major muscle, associated ipsilateral upper limb abnormalities and/or thoracic abnormalities. Few cases of patients with PS have been reported in the literature, and a detailed description of the anesthetic management of these patients is scarce. Poland syndrome imposes specific anesthetic conducts to avoid complications related to the degree of thoracic wall deformity and the possibility of malignant hyperthermia (MH) due to the use of succinylcholine and some inhaled anesthetics.

Case Report: We report the anesthetic management of a patient diagnosed with PS submitted to video-laparoscopic hernioplasty by total intravenous anesthesia with invasive ventilation. The patient presented absence of the left pectoral muscles, besides ipsilateral syndactyly. We performed monitorization with non-invasive blood pressure, pulse oximetry, esophageal thermometer, capnography, and electrocardiogram. The anesthetic technique was total intravenous anesthesia. Induction with propofol, fentanyl, cisatracurium, followed by maintenance with propofol target-controlled infusion guided by bispectral index (BIS). At the end of the procedure, tenoxicam, dipyrone, dexamethasone, and ondansetron were administered, besides atropine and neostigmine for reversal of neuromuscular blockade.

Conclusion: The PS imposes specific anesthetic management and adequate monitorization to avoid complications related to the chest deformity. The anesthetic technique was total intravenous anesthesia to reduce the doses used and also it was performed in a context to avoid MH.

Keywords: Anesthesia, Case reports, Perioperative care, Poland syndrome

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INTRODUCTION

Poland syndrome (PS) is a rare disease, with an estimated incidence of 1:30,000 to 1:80,000 live births [1], characterized by hypoplasia/agenesis of the pectoralis major muscle, associated ipsilateral upper limb abnormalities and/or thoracic abnormalities [2].

Poland syndrome is more common in males (ratio 3:1) [1]. The absence of the pectoralis major muscle is the most common component. Other manifestations of the syndrome include rib defects, lack of subcutaneous tissue, nipple anomalies, absence of axillary hair, and hand anomalies [2].

The syndrome commonly manifests sporadically, and the percentage of patients with cases in the family is less than 1% [2]. Severe rib cage abnormalities, including abnormalities in more than one rib, pectus carinatum, pectus excavatum, can lead to paradoxical rib cage movements. Dextrocardia and pulmonary hernia are also described among those with PS.

Poland syndrome imposes specific anesthetic conducts to avoid complications related to the degree of thoracic wall deformity and the possibility of MH due to the use of succinylcholine and some inhaled anesthetics, especially halothane [3]. For this reason, most of the literature recommends not to use these agents in patients with PS, although there are no described cases of PS patients suffering from MH [3].

Few cases of patients with PS have been reported in the literature, and a detailed description of the anesthetic management of these patients is scarce [4]. Most surgeries and anesthetics described in patients with PS are aesthetic surgeries, such as thoracic or mammary reconstruction, or diagnostic procedures, such as computed tomography [4].

The present study aims at reporting the anesthetic conduct in a patient with PS submitted to video-laparoscopic hernioplasty, detailing the perioperative management and the anesthetic strategy used.

CASE REPORT

A 56-year-old black male patient was admitted to the surgery service of the University Hospital of the Federal University of Maranhão with a complaint of bulging in the right inguinal region and was referred to undergo video-laparoscopic inguinal hernioplasty. His weight was 60 kg.

The patient worked as a bricklayer’s assistant, being retired for 15 years due to the left-hand deformity. He was married and father of five daughters. No comorbidities were found.

During the physical examination, absence of the left pectoral muscle (Figure 1) and ipsilateral syndactyly were identified. He reported no similar cases in the family or his circle of coexistence.

Computed tomography was performed (Figure 2) to identify other possible alterations, where absence of the major and minor pectoral muscles to the left was found, without other anatomical alterations of the chest.

He was submitted to video-laparoscopic hernioplasty using the totally extraperitoneal laparoscopic technique (TEP) without any intercurrence.

The patient was placed in Trendelenburg position and monitored with oximetry, electrocardiogram, esophageal thermometer, non-invasive blood pressure and capnography. The anesthetic technique was total intravenous anesthesia. Induction with propofol 2 mg/kg, fentanyl 5 mcg/kg, cisatracurium 0.15 mg/kg, followed by maintenance with propofol target-controlled infusion (TCI) guided by bispectral index (BIS) (40–60).

Figure 1: Front view of the chest, with evident deformity due to the absence of the major and minor pectoral muscles.

Figure 2: Computerized tomography. At the left sagittal section, showing absence of the major pectoralis muscle. On the right, a 3D reconstruction, clearly showing the absence of the major and minor pectoral muscles.
At the end of the procedure, tenoxicam 40 mg, dipyrone 1 g, dexamethasone 40 mg, and ondansetron 8 mg were administered, besides atropine 0.5 mg and neostigmine 2 mg for reversal of neuromuscular blockade. The surgery time was 60 minutes. During postoperative outpatient follow-up, the presence of other components of the syndrome was investigated.

DISCUSSION

Inguinal hernia is a very common disease worldwide. The 2018 [5] “International guidelines for groin hernia management” indicate that the chance of hernia in the groin (inguinal or femoral) is 27–43% in men and 3–6% in women.

Thus, it is feasible that patients with rare diseases will develop abdominal wall hernia. It is important that the surgeon knows how to recognize the patient's associated comorbidities and the particularities in the surgical management of their patients.

The chest wall deformity, without bone or underlying muscle makes this condition similar to an open thorax, which can generate a paradoxical breathing picture when the PS patient is submitted to stress, such as surgery. Uncontrolled spontaneous breathing under general anesthesia can worsen paradoxical breathing and lead to cardiopulmonary collapse, requiring the use of controlled positive pressure ventilation even for short procedures. Another important aspect is that the ipsilateral lung to the deformity is usually hypoplastic or smaller, leading to the possibility of deep postoperative respiratory depression and requiring adequate monitorization to avoid complications early [3].

In this study, the anesthetic technique used was the total intravenous technique, following recommendations to reduce risk of complications related to the chest wall deformity and the use of inhaled anesthetics. Due to the positioning in Trendelenburg, no regional anesthesia was performed to avoid discomfort to the patient.

There is no standardization in the literature regarding preoperative exams to be requested for a patient with PS. Considering the theoretical risks, together with routine laboratory exams, it is justified to obtain: (1) Chest imaging exams (radiography and tomography) to assess the degree of thoracic cavity deformity; (2) Respiratory function test and gasometry analysis of arterial blood to assess the consequences of possible ventilatory restriction or paradoxical breathing; (3) Color Doppler of subclavian arteries, given possible interruption of arterial supply. If the subclavian artery is significantly reduced in size or has a reduced blood flow on the affected side, the contralateral subclavian artery should be used in case of need to measure non-invasive (NIBP) and arterial blood pressure (ABP); (4) Two-dimensional Doppler echocardiography, due to the risk of cardiac malformation, such as dextrocardia [5]. In the present case, contrast-enhanced chest tomography helped in the evaluation of the mentioned data. Spirometry was not requested because the patient was asymptomatic from the respiratory point of view throughout his life and had good functional capacity (>10 METs).

In 2018, Gui et al. [4] described anesthesia for breast reconstruction surgery in PS, using rocuronium with neuromuscular blocker, as well as monitorization of body temperature and capnography, assuming the risk of malignant hyperthermia.

CONCLUSION

Poland’s syndrome is a rare condition, which may eventually be present in patients undergoing routine surgery, so that the general surgeon, as well as the anesthesiologist and his teams, must be prepared for particularities in the perioperative management of these patients. A broader preoperative evaluation is prudent, investigating and documenting possible anomalies associated with the syndrome that may have repercussions during and after the surgical/anesthetic procedure. It is justified to request imaging examinations to analyze the degree of thoracic deformity, color Doppler of subclavian arteries, respiratory function tests, and a two-dimensional Doppler echocardiogram. Because of the presumed risk of malignant hyperthermia, it is considered prudent to avoid succinylcholine as a neuromuscular blocker and inhalatory anesthesia, and careful monitorization of ETCO$_2$ and body temperature.

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Author Contributions

Rayssa Fiterman Rodrigues – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be
accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Rodríguez Castro de Lima – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor of Submission
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Ethical Approval Statement
This study was approved by the Ethics and Research Committee of Hospital São Domingos CAAE 30842220.6.0000.5085; Number 4052483. All procedures performed in the study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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