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Case Report

Anaesthesia Section

Anaesthetic Management of a Child with Poland Syndrome undergoing Left Inguinal Herniotomy: A Case Report

POONGANI NAGARAJAN[1], ARUNA PARAMESWARI[2], SOMA GANESH RAJA NEETHIRAJAN[3]

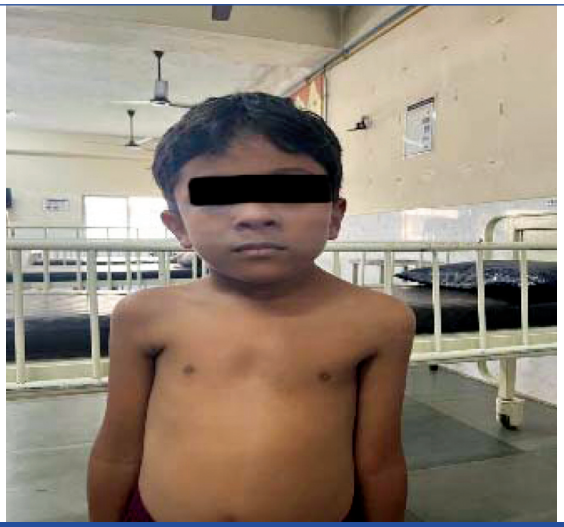
ABSTRACT

Poland syndrome is a rare congenital disorder characterised by unilateral absence or hypoplasia of the pectoralis major muscle, often associated with ipsilateral chest wall deformities, rib anomalies, and upper limb abnormalities. The severity of presentation varies widely, ranging from mild cosmetic deformities to significant thoracic asymmetry and respiratory compromise. Anaesthetic management in such patients can be challenging because of altered respiratory mechanics, associated musculoskeletal anomalies, difficult airway considerations, and the possible association with Malignant Hyperthermia (MH). Careful perioperative evaluation and individualised anaesthetic planning are therefore essential to ensure patient safety. Authors report the anaesthetic management of a six-year-old child with Poland syndrome posted for elective left inguinal herniotomy. The child had left-sided chest wall hypoplasia, thoracic hemivertebrae, mild kyphosis, and right thumb dysplasia, with preserved cardiopulmonary function. Considering the possible risk of MH, a trigger-free anaesthetic technique was planned. The anaesthesia workstation was prepared by removal of vaporisers and high-flow oxygen flushing before induction. Total Intravenous Anaesthesia (TIVA) using propofol with atracurium was administered, and airway management was successfully achieved using an i-gel supraglottic airway device. An ultrasound-guided Quadratus Lumborum (QL) block provided effective perioperative analgesia and minimised opioid requirement. Intraoperative haemodynamics and ventilation remained stable, and recovery was uneventful. This case highlights the importance of meticulous preoperative assessment, avoidance of triggering agents, and the role of multimodal anaesthetic techniques in ensuring safe perioperative management of children with Poland syndrome.

Keywords: General anaesthesia, Malignant hyperthermia, Paediatric surgery, Quadratus lumborum block, Total intravenous anaesthesia

CASE REPORT

A six-year-old male child weighing 18 kg, presented to the Department of Paediatric surgery with complaints of swelling over left inguinal region for past three weeks, a known case of Poland syndrome diagnosed during infancy, with features of left-sided chest wall hypoplasia, thoracic hemivertebrae, right thumb dysplasia and mild kyphosis. He was born preterm weighing 1.88 kg, had respiratory distress syndrome at birth requiring Continuous Positive Airway Pressure support, treated for neonatal sepsis. Neonatal imaging documented thoracic hemivertebrae and a small restrictive VSD (≈2 mm) which required no intervention. Ophthalmologic evaluations were normal. He achieved developmental milestones appropriate for corrected age. He had no other medical co-morbid illnesses, immunised to date with a surgical history of right thumb amputation under local anaesthesia at two years of age. Clinical examination revealed unilateral anterior chest wall hypoplasia with flattening of the affected hemithorax [Table/Fig-1] and posterior thoracic asymmetry with mild kyphosis [Table/Fig-2]. He was accepted under American Society of Anaesthesiologists (ASA) II for elective left inguinal herniotomy.



[Table/Fig-1]: Clinical photograph showing unilateral anterior chest wall hypoplasia with underdeveloped pectoralis major muscle and flattening of the affected hemithorax in a patient with Poland syndrome.

Preoperative Evaluation

On clinical examination, he had a heart rate of 93 beats per minute, blood pressure- 90/60 mmHg, respiration rate- 20 breaths per minute, and room air saturation of 98%. The child was neuro-developmentally normal with unremarkable airway assessment. A respiratory examination revealed slight thoracic asymmetry and obvious bilateral air entry, but no functional compromise. The cardiovascular examination was normal. Laboratory investigations were within normal limits. Echocardiography showed a 65% ejection fraction, no pulmonary hypertension, no shunts, and normal biventricular function. He was accepted for surgery under general anaesthesia with QL block because of his steady cardiorespiratory state and structural abnormalities.

Anaesthetic Management

After written informed consent from parents and adequate fasting as per institutional protocols, a trigger-free anaesthetic technique was administered with propofol (2 mg/kg) and atracurium (0.3 mg/kg). The anaesthesia workstation was prepared by removing vaporisers, flushing the fresh breathing circuit and fresh CO_2 absorbent with 100% oxygen at 10 L/min for 40 minutes. During the washout, the adjustable pressure limiting valve was kept open to ensure maximal throughput of oxygen through the system. At the end of 40 minutes,

[Table/Fig-2]: Posterior view of a child demonstrating unilateral chest wall asymmetry with mild kyphosis.

the volatile agent concentration on the anaesthesia workstation monitor registered nil inhaled (MAC=0) and end tidal levels of inhaled anaesthetic agent (sevoflurane) confirming successful washout. The cleaned circuit enabled safe initiation of TIVA without risk of unintended volatile exposure. The patient was then shifted onto the prepared workstation for induction.

After shifting the child to the operating theatre, standard ASA monitoring- ECG, non invasive blood pressure, pulse oximetry, capnography, and temperature- were applied. A 22 G intravenous cannula was secured in left metacarpal vein under aseptic precautions. The child was preoxygenated with 100% oxygen for 3-5 minutes to ensure a tight mask seal and achieve end-tidal oxygen concentration >90%. Anaesthesia was initiated using a Target-Controlled Infusion (TCI) propofol system, with effect-site concentrations titrated slowly to achieve smooth induction without haemodynamic fluctuation. Once adequate depth of anaesthesia was reached, fentanyl 25 µg was administered for analgesia, followed by atracurium 5 mg to facilitate airway instrumentation. Mask ventilation remained easy throughout, with good chest rise and stable oxygenation. After ensuring good neuromuscular relaxation, the airway was secured using an i-gel supraglottic airway device, size 2. An i-gel was preferred over endotracheal intubation as it provides adequate airway control for short-duration lower abdominal procedures while avoiding laryngoscopy and tracheal instrumentation. In addition, it allows effective ventilation with lower airway pressures, which is beneficial in patients with altered chest wall mechanics such as Poland syndrome. Insertion was atraumatic, and correct placement was confirmed by adequate chest expansion and square-wave capnograph tracing.

The child was placed in the lateral position for the ultrasound-guided QL block after anaesthesia was induced. A 22 G jelco needle was aimed at the anterolateral border of the QL muscle using an in-plane technique. A total volume of 12 mL of 0.25% bupivacaine combined with 10 µg dexmedetomidine was deposited in the QL plane with appropriate spread confirmed.

The TCI propofol and additional fentanyl infusion were used to maintain anaesthesia. The TCI propofol effect-site concentration was titrated to maintain adequate anaesthetic depth, and intermittent bolus of atracurium (1 mg) was administered as required to maintain muscle relaxation. Pressure-controlled ventilation ensured age-appropriate tidal volumes and maintained normocapnia. Throughout, haemodynamics remained stable and oxygen saturation was constantly higher than 98%. Excellent intraoperative analgesia was guaranteed by the QL block, which decreased the need for further opioids.

All infusions were tapered off and ceased at the end of the procedure. Neostigmine (0.85 mg) and glycopyrrolate (0.2 mg) injections were used to reverse neuromuscular blockade guided by clinical assessment as Train of four was not available. Adequacy of reversal was assessed clinically by the presence of regular respiratory pattern, oxygen saturation >98%, adequate spontaneous ventilation with tidal volume >5 mL/kg, sustained purposeful limb movements (sustained head lift and leg lift for 5 secs) and appropriate response to verbal commands. The i-gel was removed and the child was shifted to recovery in stable condition with adequate analgesia.

DISCUSSION

Poland syndrome is a rare congenital anomaly characterised by unilateral absence or hypoplasia of the pectoralis major muscle associated with ipsilateral chest wall deformities and upper limb abnormalities. The clinical presentation varies widely from mild cosmetic deformities to severe thoracic asymmetry with respiratory compromise [1,2]. Anaesthetic management in these patients can therefore be challenging because of altered respiratory mechanics, musculoskeletal abnormalities, difficult airway concerns, and possible susceptibility to MH [3-7].

Küpper first emphasised the anaesthetic implications of Poland syndrome and highlighted that thoracic deformities may reduce pulmonary compliance and predispose patients to perioperative hypoventilation and hypoxia, thereby necessitating meticulous respiratory evaluation and careful intraoperative monitoring [3]. Kim YH et al., later reported successful anaesthetic management during chest wall reconstruction surgery in a patient with Poland syndrome and stressed the importance of cautious ventilatory strategies in patients with severe chest wall deformities [4].

İnce I et al., described successful administration of general anaesthesia in a patient with Poland syndrome and discussed the possible association between Poland syndrome and MH susceptibility [5]. Rosenberg H et al., described MH as a potentially fatal pharmacogenetic disorder triggered by volatile anaesthetic agents and succinylcholine and emphasised the importance of early recognition and trigger avoidance [6]. Riazi S et al., further recommended trigger-free anaesthesia techniques along with proper anaesthesia workstation preparation in susceptible patients [7]. Hopkins reviewed advances in MH diagnosis and perioperative management and highlighted the role of TIVA in preventing MH episodes [8]. Litman RS et al., discussed MH susceptibility and related myopathies, emphasising vigilant perioperative monitoring and preparedness in at-risk patients [9]. Cieniewicz A and Trzebicki J also reinforced the importance of awareness and preventive strategies for MH during anaesthetic management [10]. Kumar NG et al., specifically discussed the anaesthetic implications in Poland syndrome and recommended individualised perioperative planning with avoidance of triggering agents whenever MH susceptibility is suspected [11].

Regional anaesthesia techniques form an important component of multimodal perioperative analgesia because they reduce perioperative opioid consumption and facilitate enhanced recovery. Blanco R et al., demonstrated that QL block provides effective analgesia for lower abdominal surgeries with reduced postoperative opioid requirement [12]. Hebbard P et al., further highlighted the usefulness of ultrasound-guided fascial plane blocks in improving perioperative analgesia and reducing systemic analgesic requirement [13].

Gui L et al., described successful anaesthetic management for chest wall reconstruction in a child with Poland syndrome and emphasised the importance of detailed respiratory evaluation and vigilant intraoperative monitoring [14]. Rodrigues RF et al., reported successful use of TIVA in a patient with Poland syndrome undergoing laparoscopic inguinal hernioplasty and highlighted the benefits of avoiding volatile anaesthetic agents [15]. Sharma S

and Sharma S discussed anaesthetic challenges including airway management in children with Poland syndrome and stressed the importance of individualised airway planning [16]. Jun YE et al., also demonstrated successful perioperative management using careful airway assessment and tailored anaesthetic techniques in a patient with Poland syndrome [17].

In the present case, meticulous preoperative evaluation, avoidance of triggering agents, appropriate anaesthesia workstation preparation, and use of propofol-based TIVA ensured safe perioperative management. The i-gel supraglottic airway device provided adequate ventilation with minimal airway stimulation, while ultrasound-guided QL block ensured effective perioperative analgesia with stable haemodynamics and minimal postoperative opioid requirement.

Overall, the present case supports existing literature that individualised perioperative planning, trigger-free anaesthetic techniques, vigilant monitoring, and multimodal analgesia can ensure safe perioperative outcomes in children with Poland syndrome.

CONCLUSION(S)

Poland syndrome presents unique anaesthetic challenges due to associated chest wall deformities and the potential risk of MH. This case highlights the importance of thorough preoperative evaluation and careful planning of a trigger-free anaesthetic technique. TIVA combined with QL block provides effective analgesia while minimising opioid requirements. The use of a supraglottic airway device can be a safe and appropriate alternative to endotracheal intubation in selected cases. With vigilant monitoring and a tailored perioperative approach, children with Poland syndrome can undergo surgical procedures safely with favourable outcomes.

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PARTICULARS OF CONTRIBUTORS:

1. Postgraduate Student (Final Year), Department of Anaesthesiology, Sri Ramachandra Institute of Higher Education and Research, Chennai, Tamil Nadu, India.
2. Professor and Head, Department of Anaesthesiology, Sri Ramachandra Institute of Higher Education and Research, Chennai, Tamil Nadu, India.
3. Associate Professor, Department of Anaesthesiology, Sri Ramachandra Institute of Higher Education and Research, Chennai, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Poongani Nagarajan,
Postgraduate Student (Final Year), Department of Anaesthesiology, Sri Ramachandra Institute of Higher Education and Research, Ramachandra Nagar, Porur, Chennai-600116, Tamil Nadu, India.
E-mail: poonganinagarajan@gmail.com

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