Content available at: https://www.ipinnovative.com/open-access-journals

IP Journal of Surgery and Allied Sciences

Journal homepage: https://www.jsas.co.in/

Case Report

Perioperative management of a patient with tetra-phocomelia undergoing anterior cervical decompression and fixation: A case report

Prajakta N Patil[1]*, Vaibhavi Baxi[1], Yasaswini Jami[1], Naba Mateen Madoo[1]

[1]Dept. of Anaesthesia, Lilavati Hospital and Research Centre, Mumbai, Maharashtra, India.

Abstract

Phocomelia is a rare congenital condition characterized by severe limb malformation, where the limbs are either partly or completely underdeveloped. Phocomelia can occur as a syndrome or as a limb-specific abnormality. It is characterized by high rates of stillbirth, intrauterine demise, and early neonatal mortality. Administering anaesthesia to these patients is challenging because of anatomical and physiological variabilities. Limited experience exists regarding the management of this condition in surviving adults, and no standardized perioperative or anaesthetic guidelines are available. We present the perioperative management of a 22-year-old female with isolated non-syndromic phocomelia, with no history of intrauterine teratogenic exposure, who underwent anterior cervical decompression and fixation at the C3–C5 level. Key anaesthetic challenges included difficult intravenous access, complex arterial and venous catheterization due to anatomical variation, and positioning difficulties.

Keywords: Phocomelia, Tetra-phocomelia, Anaesthetic Management, Cervical Decompression

Received: 29-04-2025; **Accepted:** 28-05-2025; **Available Online**: 25-07-2025

1. Introduction

Phocomelia is a congenital disorder characterized by the partial or complete absence of long bones, resulting in abnormal limb development. The proximal portion of the limb is either missing or noticeably hypoplastic, while the hands and feet are usually normal or nearly normal. Phocomelia affects approximately 0.6 to 4.2 per 100,000 live births worldwide, with a higher incidence in the left-sided upper limbs.[1] Unilateral limb involvement is more prevalent than bilateral involvement.[2]

The primary etiological factors of phocomelia include genetic inheritance and teratogenic exposure, notably thalidomide during pregnancy.[3] Based on the absent portion of the limb relative to the trunk, phocomelia is classified into three categories:

1. **Type 1** (Complete Phocomelia): Complete absence of proximal and distal limb segments, with hands or digits directly attached to the trunk.
2. **Type 2** (Proximal Phocomelia): Absence of the proximal limb, where the humerus is missing, and the forearm with hands and digits is attached to the trunk.
3. **Type 3** (Distal Phocomelia): Absence of the distal limb, where the forearm is absent, and the hand or digits are directly attached to the humerus.[4]

This report presents a case of a young adult with tetra-phocomelia (Type 3) requiring surgical intervention for cervical spine pathology and discusses its anaesthetic implications.

2. Case Report

A 22-year-old female with Type 3 phocomelia was scheduled for anterior cervical decompression and fixation at the C3–C5 level. The patient had no history of teratogenic drug exposure or radiation during intrauterine life. Preoperative assessment revealed severe upper and lower limb deformities, posing significant challenges for vascular access, intraoperative monitoring, and patient positioning.

*Corresponding author: Prajakta N Patil
Email: prajaktanpatil@yahoo.co.in

https://doi.org/10.18231/j.jsas.2025.018

The patient, the fourth child of a non-consanguineous marriage, was delivered at home via normal vaginal delivery. She had a family history of phocomelia, with one elder sibling affected and two others who did not survive, though their cause of death was not specified. Her youngest sibling was unaffected. She reported progressive neck pain, tingling, numbness, and bilateral upper-limb radiculopathy. Neurological examination revealed motor power of 4/5 on the right and 3/5 on the left, with intact sensory function. A CT scan showed reduced intervertebral disc space at C3–C5, loss of cervical lordosis, and mild degenerative changes.

Figure 1: Preoperative image of patient.

Figure 2: Intra operative patient positioning

She weighed 50 kg, had no craniofacial abnormalities, a Mallampati score of 2, and normal cardiopulmonary function. She had tetra-phocomelia with abscence of distal part of all four limbs and partially fused digits attached to the proximal limbs, consistent with Type 3 phocomelia. Anaesthetic challenges included vascular access, airway management, and positioning. Although a difficult airway was anticipated due to a short neck, she had adequate mouth opening, and tracheal intubation was successfully achieved without difficulty using a C-MAC video laryngoscope. A multidisciplinary anaesthetic plan was devised, and informed consent was obtained.

Standard ASA monitoring, including pulse oximetry (SpO_2), electrocardiography (ECG), invasive arterial line, and bispectral index (BIS), was initiated. Non-invasive blood pressure (NIBP) monitoring was not feasible due to inadequate limb length, which made standard cuff application impractical. Therefore, invasive arterial blood pressure monitoring was mandated, necessitating cannulation of a proximal artery for continuous beat-to-beat monitoring.

Right femoral arterial cannulation was initially attempted; although arterial puncture was successful, guidewire advancement was unsuccessful, likely due to anatomical variation. Subsequently, an attempt was made at left femoral artery cannulation, but similar difficulties were encountered. Ultimately, arterial access was successfully secured, ensuring reliable invasive blood pressure monitoring throughout the surgery. Consequently, right internal jugular vein cannulation was performed under ultrasound guidance, ensuring secure central venous access.

General anaesthesia was induced intravenously with midazolam 2 mg, fentanyl 100 µg, propofol 90 mg, and cisatracurium 10 mg. Tracheal intubation was successfully performed using C-MAC video laryngoscopy with a flexo-metallic 6.5 cuffed tube, fixed at 20 cm. Anaesthesia depth was guided by BIS (40–50) and age-adjusted MAC (0.8–1). The patient remained hemodynamically stable. Temperature was monitored using an oesophageal probe and maintained with a forced air-warming device. Post-induction, a nasogastric tube and urinary catheter were inserted. Prophylactic cefaperazone-sulbactam 1.5 g IV was administered for infection prevention, and tranexamic acid 500 mg IV was given to minimize intraoperative bleeding. The patient was positioned supine with neck extension and adequate padding. Positioning posed challenges due to her short stature and short neck, which limited head and neck mobility. The short neck could have complicated placement on a horseshoe headrest had the procedure required a prone position. Fortunately, the anterior cervical approach required supine positioning, which simplified intraoperative management. Anaesthesia was maintained with sevoflurane, remifentanil infusion, and cisatracurium.

Surgery was performed via the Smith-Robinson approach, which is commonly used for anterior cervical spine procedures. A C3-C4 and C4-C5 discectomy was performed, followed by anterior cervical plating from C3 to C5 using a 14 × 4 self-drilling screw. The patient remained hemodynamically stable with minimal blood loss (100–125 mL), managed with crystalloids and colloids. Reversal was

achieved with IV myopyrrolate, and after confirming recovery, she was extubated. The patient tolerated the procedure well and was transferred to the ward after observation in the recovery room.

2.1. Postoperative management

After extubation, the patient was transferred to the post-anesthesia care unit (PACU) for monitoring. Invasive arterial blood pressure monitoring continued, and oxygen was administered to maintain SpO_2 above 96%. Postoperative analgesia was managed with intravenous paracetamol and low-dose fentanyl for breakthrough pain. Neurological function was assessed regularly, with no new deficits observed.

The patient was monitored for respiratory complications, with early chest physiotherapy and regular arterial blood gas analyses. Central venous was removed after 24 hours once hemodynamic stability was confirmed. She was gradually mobilized, wearing a cervical collar for stabilization. Oral intake was resumed 6 hours post-extubation. The patient had an uneventful recovery, with no complications, and was discharged on postoperative day 5 with instructions for follow-up care and physiotherapy.

Table 1: Postoperative care table

Postoperative Care Aspect	**Management Details**
Monitoring	Continuous monitoring of vitals, SpO_2, ECG, and invasive arterial blood pressure.
Respiratory Care	Oxygen therapy to maintain SpO_2 above 96%, chest physiotherapy to prevent atelectasis.
Analgesia	IV paracetamol for pain relief; low-dose fentanyl for breakthrough pain.
Neurological Assessment	Regular neurological checks for any new deficits or changes in motor/sensory function.
Vascular Access	Central venous and arterial lines removed 24 hours postoperatively after hemodynamic stability is confirmed.
Mobilization	Gradual mobilization with a cervical collar for support.
Diet and Fluid Intake	Oral intake resumed 6 hours post-extubation, with gradual advancement.
Postoperative Complications	No complications observed, with regular monitoring for signs of infection, bleeding, or neurological deficits.
Discharge Planning	Discharged on postoperative day 5 with follow-up instructions for physiotherapy and wound care.

3. Discussion

Phocomelia is a rare congenital anomaly characterized by the complete or partial absence of limbs, classified into different types based on the extent of limb development.[4] Our patient belonged to Type 3 phocomelia, wherein there was complete absence of the proximal and distal portions of all four limbs. Such anatomical abnormalities present unique challenges for anesthetic management, especially in terms of monitoring, vascular access, and patient positioning.

One of the primary anesthetic considerations in this patient was the inability to apply a non-invasive blood pressure (NIBP) cuff, which is a standard monitoring modality.

A major anesthetic consideration was the inability to use non-invasive blood pressure (NIBP) monitoring because of inadequate limb length, preventing proper cuff placement. Therefore, continuous invasive blood pressure monitoring was essential. Given the absence of adequate distal vessels, cannulation of a proximal artery was required to obtain reliable arterial access for real-time hemodynamic monitoring and arterial blood sampling during the procedure.[5] An arterial line was placed under local anesthesia prior to induction, providing continuous beat-to-beat blood pressure monitoring and facilitating blood sampling when required. This ensured close hemodynamic monitoring, essential for patient safety, particularly in the context of intraoperative fluid shifts and anesthetic depth adjustments.

Airway management was also a critical consideration. Although patients with phocomelia may have associated craniofacial anomalies[6] our patient had only short neck as difficult airway predictors, and mask ventilation and intubation were successfully achieved with standard techniques using CMAC. However, it is imperative to always anticipate airway difficulties and have a difficult airway cart readily available.

Venous access can also be challenging in such patients due to altered anatomy and restricted vascular sites. In our case, peripheral venous access was not feasible; central venous cannulation was therefore necessary[7]

Positioning for surgery in patients with phocomelia presents unique challenges due to altered body proportions and anatomical constraints. In our case, the patient's short stature and short neck made head and neck positioning technically demanding. The short neck would have made placement in a horseshoe headrest particularly difficult if a prone position were required. However, since the anterior cervical decompression was performed in the supine position,

optimal positioning was achieved using standard supports with meticulous padding to prevent pressure injuries and nerve compression.[8]

Given the rarity of phocomelia, it is also important to consider associated anomalies, particularly cardiac, renal, and skeletal defects.[9] Our patient had no significant cardiac involvement; however, preoperative evaluation should always include a thorough cardiac assessment to rule out congenital heart disease, which may further impact anesthetic planning.

Postoperative care for patients with complex congenital conditions like phocomelia requires close monitoring and a tailored approach. The patient was closely observed in the recovery room for any signs of complications. A detailed postoperative care plan was formulated, and the key aspects of care were summarized in the table above.

4. Conclusion

Phocomelia poses significant anesthetic challenges due to the absence of limbs, making standard monitoring and vascular access difficult. Invasive arterial blood pressure monitoring becomes essential in such cases to ensure patient safety. Careful preoperative planning, anticipation of airway and positioning difficulties, and a vigilant intraoperative strategy can lead to successful management. Awareness of associated congenital anomalies is crucial. A tailored, patient-centric approach ensures safe anesthetic care in patients with this rare congenital disorder.

5. Conflict of Interest

None.

6. Source of Interest

None.

References

1. Hecht F, Gorlin RJ. Phocomelia: an analysis of sixty-two cases and delineation of the syndrome. *Am J Hum Genet*. 1970;22(3):298-305.
2. Stevenson RE, Hall JG, Goodman RM. Human Malformations and Related Anomalies. Oxford: Oxford University Press; 1993.
3. Vargesson N. Thalidomide-induced limb defects: resolving a 50-year-old puzzle. Bioessays. 2009;31(12):1327-36.
4. Aase JM. Diagnostic Dysmorphology. New York: Springer; 2005.
5. Ahmed S, Shankar H. Invasive Blood Pressure Monitoring. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2023.
6. Sousa SB, Abdul-Rahman OA, Bottani A, Graham JM, Manouvrier-Hanu S, Fryns JP. et al. Phenotypic and genotypic variability in Roberts syndrome. *Am J Med Genet A.* 2010;152A(6):1470-9.
7. Brown L, Morgan CJ. Central venous access: techniques and complications. *Anesthesiol Clin.* 2017;35(4):735-52.
8. Langeron O, Birenbaum A, Francon D. Handling the patient with a difficult airway. *Minerva Anestesiol*. 2009;75(7-8):447-52.
9. Tosi LL, Warman ML. Primary limb abnormalities. *J Am Acad Orthop Surg.* 2004;12(6):409-17.

Cite this article: Patil PN, Baxi V, Jami Y, Madoo NM. Perioperative management of a patient with tetra phocomelia undergoing anterior cervical decompression and fixation: A case report. *IP J Surg Allied Sci.* 2025;7(2):85-88.