European Journal of Clinical Pharmacy
Print ISSN:2385-409X **Online ISSN**:3105-0409
Website: https://farmclin.com/

Neuroprotective Anesthesia in a Ruptured Meningomyelocele: Managing a High-Risk Infant with Hydrocephalus

Case Report

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Received: 03-10-2025
Revised: 28-10-2025
Accepted: 09-11-2025
Published: 24-12-2025

Abstract:

Background: Meningomyelocele (MMC) is a severe neural tube defect often associated with hydrocephalus, neurological impairment, and risk of infection when ruptured. Anesthetic management in such infants is complex due to difficult positioning, raised intracranial pressure (ICP), fragile neural placode, risk of sepsis, and associated congenital cardiac anomalies. A multidisciplinary perioperative approach is crucial to ensure a safe surgical outcome.

Case Presentation: A 4-month-old male infant weighing 3.48 kg presented with fever, CSF leakage from a lumbar swelling, progressive increase in head size, and bilateral clubfoot deformity. Clinical evaluation and imaging confirmed ruptured lumbar MMC with obstructive hydrocephalus, along with congenital heart disease including ASD, VSD, and a small PDA. General anesthesia was administered with emphasis on neuroprotection, maintaining stable hemodynamics, and preventing pressure on the exposed neural tissue. Modified positioning was required during induction and intubation to avoid sac compression. Smooth induction, maintenance of normocapnia, cautious fluid therapy, and avoidance of succinylcholine were practiced to minimize ICP surges. A right ventriculo-peritoneal shunt was inserted followed by MMC repair in prone position, with vigilant monitoring of cardiorespiratory parameters and pressure points throughout positional changes. Post-operative care included continued neurological monitoring, infection prophylaxis, shunt surveillance, and appropriate analgesia. The infant tolerated the procedure well and was shifted to the recovery room in stable condition. In infants with ruptured MMC and hydrocephalus, tailored anesthetic management focusing on airway safety, ICP control, infection prevention, and meticulous positioning is essential for favorable outcomes.

Keywords: Ruptured meningomyelocele, Hydrocephalus, Pediatric anesthesia, Ventriculo- peritoneal shunt, Neuroprotection

INTRODUCTION

Meningomyelocele (MMC) is the most severe and common form of open spinal dysraphism, resulting from failure of neural tube closure between the 3rd and 4th weeks of gestation. It is associated with exposed neural elements, varying degrees of lower limb paralysis, sensory loss, bowel and bladder dysfunction, and significant long-term neurodevelopmental disability (1). Hydrocephalus coexists in nearly 70–90% of MMC cases due to Arnold–Chiari II malformation and progressive obstruction of cerebrospinal fluid pathways (2). Early surgical intervention remains the cornerstone of management to reduce the risks of infection, protect exposed neural tissue, and enhance the potential for improved neurological outcomes. However, in low-resource settings, delayed presentation continues to be a major challenge due to limited access to specialized neurosurgical services (3).

The perioperative management of infants with MMC and hydrocephalus is particularly complex for anesthesiologists. Multiple concerns must be simultaneously addressed including raised intracranial pressure (ICP), difficult airway access, risk of autonomic instability, and frequent presence of congenital anomalies such as cardiac defects, renal abnormalities, and orthopedic deformities (4). Any rise in ICP during induction, laryngoscopy or intraoperative positioning may compromise cerebral perfusion and worsen neurological function. Additionally, hydrocephalus can lead to increased head size, complicating airway alignment and mask sealing, demanding an experienced pediatric anesthesia team (5).

Rupture of MMC introduces further urgency and risk. Loss of cerebrospinal fluid integrity predisposes to meningitis, septicemia, electrolyte imbalance, and prolonged hospitalization. A ruptured sac also increases perioperative vulnerability due to direct exposure of neural tissue during transport and handling (6). Anesthetic planning must emphasize strict aseptic protocols, careful positioning to avoid compression of the defect, and cautious airway establishment without exacerbating neural damage or ICP fluctuations (7).

Positioning poses a unique anesthetic challenge in MMC surgery, particularly in large lumbosacral lesions where prone positioning is required. Induction frequently requires lateral positioning to prevent pressure on the fragile sac until definitive repair (8). Furthermore, infants with neural tube defects often exhibit latex allergy due to repeated exposures during early life, requiring strict avoidance of all latex-containing materials in the operating theatre to prevent life-threatening hypersensitivity reactions (9).

Coexisting congenital heart disease, as often seen in MMC cases, significantly influences anesthetic decisions. Maintenance of stable systemic and pulmonary circulations is essential to prevent shunt imbalance and hypoxia (10). Detailed preoperative cardiac evaluation and vigilant intraoperative monitoring are required to mitigate perioperative risks, especially during fluid management and ventilatory adjustments (11).

Modern anesthetic strategies prioritize neuroprotection through smooth induction, controlled ventilation targeting normocapnia, opioid-sparing analgesia, prevention of hypothermia, and meticulous fluid therapy to maintain euvolemia without worsening cerebral edema (12). Postoperative planning includes extubation readiness evaluation, continued ICP monitoring, ventilatory support if needed, and infection prevention to ensure shunt function and neurological preservation (13).

The case report emphasises on detailed perioperative anesthetic management of a high-risk infant presenting with ruptured lumbar MMC, hydrocephalus, and congenital cardiac anomalies. The report highlights the need for individualized anesthetic strategies, multidisciplinary coordination, and vigilant monitoring throughout perioperative stages to achieve a favorable surgical outcome in such critically vulnerable patients.

CASE PRESENTATION

Patient Profile: A 4-month-old male infant, weighing 3.48 kg, was brought to the hospital with complaints of fever for four days and persistent discharge from a lumbosacral swelling since birth. Parents also reported progressive enlargement of the head over two months and reduced lower limb movement. There was no history of seizures. The baby was delivered at term by lower segment cesarean section, and a soft swelling was noted over the lower back immediately after birth, however, surgical intervention was delayed due to socioeconomic limitations.

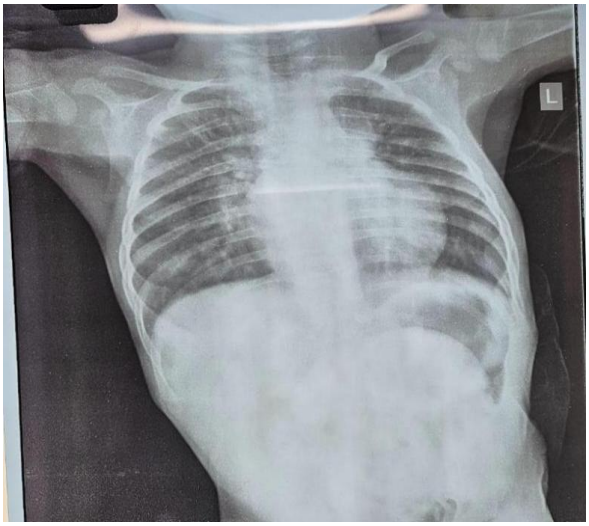
The mother gave a history of antiepileptic drug intake during pregnancy, specifically valproate, which is a well-established teratogen. Antenatal exposure to valproate is known to significantly increase the risk of neural tube defects, hydrocephalus, congenital cardiac anomalies, and musculoskeletal deformities. This exposure likely contributed to the multisystem involvement observed in the neonate, including meningomyelocele with hydrocephalus, associated cardiac defects, and congenital talipes equinovarus. The presence of multiple valproate-related congenital anomalies placed the child in a high-risk category for perioperative anesthetic management.

Clinical Examination: On admission, the infant was alert but irritable, with a bulging anterior fontanelle and a positive sunset sign indicating increased intracranial pressure (ICP). The lumbosacral lesion was ruptured with continuous CSF leakage and exposed neural tissue. Cardiovascular examination revealed a systolic murmur. Neurological evaluation showed bilateral lower limb weakness and bilateral clubfoot deformities. Temperature and infection markers suggested ongoing local infection risk.

Figure 1: Clinical presentation of a 4-month-old infant positioned in the lateral posture, demonstrating macrocephaly and modified positioning to avoid pressure on the lumbosacral meningomyelocele during perioperative care.

Preoperative Investigations and Diagnosis: MRI of the brain and spine confirmed a ruptured lumbar meningomyelocele with obstructive hydrocephalus, low-lying conus, and evidence of Arnold–Chiari II malformation. Two-dimensional echocardiography revealed atrial septal defect (ASD), ventricular septal defect (VSD), and a small patent ductus arteriosus (PDA), all demonstrating a left-to-right shunt pattern. Routine blood tests were within acceptable limits for surgery. A diagnosis of ruptured MMC with hydrocephalus and associated congenital heart disease was established, necessitating urgent neurosurgical repair and CSF diversion.

Figure 2: Chest Radiograph (Anteroposterior View) of a 4-Month-Old Infant



Anesthetic Management: The anesthetic management was carefully planned to address the combination of ruptured meningomyelocele, obstructive hydrocephalus, and underlying congenital heart disease in a small infant. The primary goals were to protect the exposed neural tissue, avoid further increases in intracranial pressure, maintain stable hemodynamics in the presence of left-to-right shunts, and ensure safe airway management despite difficult positioning constraints.

Preparation, Monitoring, and Positioning Strategy: Given the well-known association of MMC with latex sensitivity, all latex-containing materials were strictly excluded from the operating room. Standard monitors (ECG, non-invasive blood pressure, pulse oximetry, temperature, andcapnography) were applied along with precordial stethoscope and appropriate warming devices to prevent hypothermia. Large occiput and hydrocephalus posed challenges to neutral positioning of the

head and alignment of the airway. Because supine positioning would compress the ruptured sac,a plan for induction and intubation in the lateral position was formulated. Adequate padding of the sac and pressure points was ensured, and all tubes and lines were secured with extra care anticipating multiple position changes (lateral, supine briefly, and prone).

Figure 3: Intraoperative photograph demonstrating lateral-position endotracheal intubation in a 4-month-old infant with macrocephaly due to hydrocephalus. The lateral decubitus position was used to avoid compression of the lumbosacral meningomyelocele during airway management, with full standard monitoring and pediatric anesthesia support.

Induction and Airway Management: After instituting standard monitoring, anesthesia was induced with the patient positioned laterally. Given the anticipated difficult airway and the need for rapid and controlled airway securing, succinylcholine was administered after careful risk– benefit evaluation. The decision was made in view of the requirement for prompt airway control in a neonate with poor airway reserve, distorted anatomy, and a ruptured meningomyelocele, where prolonged airway manipulation could have resulted in hypoxia and hemodynamic instability. Endotracheal intubation was successfully achieved in the lateral position using a C-MAC video laryngoscope to minimize airway manipulation and optimize glottic visualization. A pediatric stylet was required due to difficulty in aligning the oral, pharyngeal, and laryngeal axes secondary to the disproportionately enlarged head.

Attenuation of Intubation Response: Immediately following intubation, the sympathetic response was attenuated using sevoflurane and a short-acting opioid, as documented in the peri- anesthetic evaluation sheet. This approach was adopted to prevent abrupt increases in heart rate and blood pressure, minimize sympathetic stimulation, and avoid sudden rises in intracranial pressure, especially in the presence of underlying congenital heart disease and raised intracranial pressure.

Intraoperative Maintenance and Neuroprotection: Intraoperatively, meticulous attention was directed toward maintaining hemodynamic stability in view of the underlying congenital cardiac defects with left-to-right shunt physiology. Normoxia, normocapnia, and normothermia were strictly maintained throughout the procedure to prevent increases in pulmonary vascular resistance. Anesthetic depth was carefully titrated to avoid sudden reductions in systemic vascular resistance, which could otherwise exacerbate left-to-right shunt flow and compromise systemic perfusion. Intravenous fluids were administered judiciously to maintain adequate preload while avoiding volume overload. Continuous monitoring of heart rate, noninvasive blood pressure, oxygen saturation, and end-tidal carbon dioxide was ensured throughout the intraoperative period to promptly detect and correct any hemodynamic fluctuations.

Hemodynamic Management and Cardiac Considerations: In view of ASD, VSD, and PDA, hemodynamic strategy focused on avoiding increases in pulmonary vascular resistance and large fluctuations in systemic vascular resistance. Hypoxia, hypercarbia, acidosis, and hypothermia all of which could increase PVR were meticulously prevented. Sudden decreases in SVR (for example, from deep volatile anesthesia or large boluses of vasodilating drugs) were avoided to prevent worsening left-to-right shunt and potential heart failure. Any tachycardia, hypotension, or desaturation was promptly investigated and managed with incremental fluid boluses, titrated anesthetic depth adjustments, and vasoactive support if needed.

Analgesia and Emergence: Multimodal analgesia was provided using intravenous

paracetamol and opioids in carefully titrated doses appropriate for age and weight, aiming for smooth emergence without agitation, which could raise ICP and strain the surgical repair. Regional techniques were avoided in view of the open spinal defect and recent neural repair. At the end of surgery, residual neuromuscular blockade was reversed and the child was extubated only after fulfilling strict criteria: adequate spontaneous respiratory effort, acceptable gas exchange, normothermia, hemodynamic stability, and absence of excessive airway secretions. Extubation was performed in the lateral position to maintain airway patency and protect the surgical site. Controlled mechanical ventilation was employed using a lung- protective strategy. Ventilation parameters were adjusted to maintain normocapnia, with end- tidal carbon dioxide maintained within normal limits to avoid hypercapnia-induced increases in intracranial pressure and pulmonary vascular resistance. Excessive airway pressures were avoided to prevent barotrauma and hemodynamic compromise. Ventilatory settings were reassessed and optimized following positioning to ensure stable cerebral and cardiovascular physiology.

Postoperative Care and Follow-Up: Postoperatively, the infant was transferred to a monitored setting for close observation of respiratory status, shunt function, wound integrity, and neurological signs. Pain control was continued with intravenous paracetamol and small opioid doses as required. Fluid balance, temperature, and infection parameters were closely monitored. Parents were counselled about warning signs of shunt malfunction, infection, and the need for long-term follow-up with neurosurgery, cardiology, pediatrics, and rehabilitation services. The overall perioperative course remained uneventful, reflecting the effectiveness of a carefully planned and individualized anesthetic strategy in this high-risk infant.

DISCUSSION

The anesthetic management of infants with meningomyelocele (MMC), particularly when complicated by both hydrocephalus and rupture, poses significant challenges requiring multidisciplinary coordination. MMC frequently coexists with Arnold–Chiari II malformation, resulting in obstructive hydrocephalus and elevated intracranial pressure (ICP), which exacerbate neurological vulnerability and guide many anesthetic decisions (Ntimbani et al., 2020; Takoutsing et al., 2023) (1,2). Raised ICP necessitates careful perioperative control of ventilation, maintenance of cerebral perfusion, and avoidance of precipitous hemodynamic changes that may compromise brain function (Rajesh et al., 2017; Tripathy & Ahmad, 2019) (4,5).

Airway management is further complicated in MMC by macrocephaly, limited neck motion, and the inability to use the standard supine position due to the exposed neural sac (Huang et al., 2023) (14). Literature supports lateral intubation as a best-practice approach in these difficult-airway scenarios, minimizing the risk of sac compression and neurological deterioration (Vagyannavar et al., 2017) (15). Our case closely follows these recommendations, with successful lateral-position intubation ensuring airway security while protecting the ruptured lesion. Succinylcholine avoidance in our infant reflected concerns regarding increased ICP response and potential neuromuscular abnormalities in dysraphism (Singh et al., 2010) (16).

Rupture of MMC, as in our infant, demands urgent neurosurgical closure to prevent meningitis, CSF contamination, and progressive cord injury (Suryaningrat et al., 2024) (17). Meticulous asepsis and perioperative antibiotics remain essential, paralleling evidence showing reduced infectious complications in CSF leak scenarios when early antimicrobial coverage is maintained (Wang et al., 2023; Chowdhury et al., 2014) (6,18).

Prone positioning used during MMC closure significantly alters respiratory mechanics, venous return, and risks endotracheal tube displacement (Kwee et al., 2015) (8). Our approach aligned with the literature by performing ventriculo-peritoneal (VP) shunt placement first, thereby decompressing the ventricles before prone positioning. This staged strategy is supported by Kahle et al. (2024), who demonstrate improved compliance and reduced intraoperative ICP spikes following CSF diversion (19). Additionally, continuous reassessment of airway and hemodynamics during repositioning reduced complications consistent with perioperative recommendations (Vincent Bargnes et al., 2025) (20).

Congenital heart defects, including ASD, VSD, and PDA as seen in our patient, remain

common comorbidities in MMC and significantly influence anesthetic decisions (Junghare & Desurkar, 2017) (10). Guidelines advise preventing hypoxia, hypercarbia, acidosis, and excessive fluid administration that may increase pulmonary vascular resistance and shunt burden (Farooq et al., 2025; Malbrain et al., 2020) (11,21). Hemodynamically stable conduct of our case highlights the success of a balanced approach, optimized ventilation, and vigilant monitoring.

Latex allergy is another major concern in spina bifida patients, often under-recognized until anaphylaxis occurs (Meneses et al., 2020) (9). Consistent with preventive guidance, a latex- free environment was strictly enforced in our case to mitigate sensitization risk (Arasi et al., 2023) (22).

Post-extubation respiratory compromise remains a known risk due to brainstem compression from hydrocephalus and prolonged anesthesia duration (Bruder & Ravussin, 1999; Zhou et al., 2025) (13,23). In line with postoperative recommendations, extubation in the lateral position ensured minimal strain on the repair and adequate airway patency. Multimodal, opioid-sparing analgesia optimized recovery while preserving ventilatory drive, supporting current strategies for safe neurosurgical anesthesia (Hosseinzadeh & Nourazarian, 2025; Saraswat, 2015) (12,24).

This case underscores the complex perioperative anesthetic challenges associated with valproate-related multisystem congenital anomalies. Severe hydrocephalus with gross macrocephaly significantly altered airway anatomy, rendering conventional supine intubation impractical. Planned lateral position intubation using video laryngoscopy, conscious use of succinylcholine after careful risk–benefit assessment, and prompt attenuation of the intubation response were pivotal in securing the airway safely. Additionally, tailored intraoperative hemodynamic and ventilatory strategies were essential to balance the competing demands of raised intracranial pressure and congenital heart disease.

Comparison with Published Evidence

Our case differed significantly from typical reported scenarios that emphasize early neonatal MMC repair (Suryaningrat et al., 2024) (17). Instead, delayed presentation at 4 months with sac rupture, active CSF leak, and preexisting cardiac anomalies represented a higher-risk and more complex perioperative challenge. Nonetheless, adherence to literature-supported strategies led to a stable and favorable outcome, reinforcing the critical role of early planning and structured neuroprotective anesthesia in resource-limited environments (Shah et al., 2024) (25).

CONCLUSION

Infants with ruptured meningomyelocele and hydrocephalus represent a high-risk population requiring specialized anesthetic management to reduce morbidity and preserve neurological function. The presence of raised intracranial pressure, difficult airway positioning constraints, risk of sepsis, and coexisting congenital cardiac defects demands a balanced approach that ensures cerebral protection, stable hemodynamics, and secure ventilation throughout surgery. In this case, a carefully planned induction in the lateral position, strict latex-free precautions, controlled ventilation, cautious fluid therapy, and staged surgical strategy with initial VP shunt placement enabled a smooth intraoperative course and favorable postoperative outcome. Continuous postoperative monitoring and multidisciplinary follow-up remain essential for long-term neurological and systemic well-being. This case reinforces that individualized perioperative strategies and coordinated team involvement are fundamental to achieving successful outcomes in infants undergoing complex neurosurgical surgery for ruptured MMC with hydrocephalus.

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