Case Report

DOI: 10.7860/JCDR/2026/84546.22336

Perioperative Anaesthetic Challenges in a Case of Lumbosacral Meningomyelocele: A Case Report

Anaesthesia Section

POORNIMA DHAKSHINAMURTHY[1], SHYAMOLIMA BHUYAN[2]

ABSTRACT

Meningomyelocele is a congenital neural tube defect caused due to incomplete closure of the neural tube during foetal development. In meningomyelocele, a sac is formed due to herniation of the neural tissues and cerebrospinal fluid through defective vertebral arches and is covered by a thin membrane. It produces paraplegia, hydrocephalus, urinary incontinence and higher mental function impairment. It is almost always associated with Arnold Chiari type 2 malformations, wherein there is cerebellar herniation. It is prevalent in developing countries due to non-compliant prenatal care. Proper folic acid supplementation throughout pregnancy helps prevent these neural tube defects. But owing to various socio-economic concerns, that is usually not the case. Paediatric anaesthesia can be a challenging task as they have various anatomical and physiological differences from adults. Proper counselling and explanation play a crucial role in meningomyelocele repair surgeries, especially in the paediatric age group. So many patients end up having post-neurological sequelae if not intervened at the right time. Sometimes, meningomyelocele repair surgeries can be done in the prenatal period as either an open or a foetoscopy approach. This case report is about a 15-day-old neonate with lumbosacral meningomyelocele associated with Arnold Chiari type 2 malformation, a soft palate cleft and a small atrial septal defect posted for meningomyelocele repair surgery. The presence of a cleft palate made it a case of an anticipated difficult airway and with Atrial Septal Defect (ASD), strict hemodynamic control had to be maintained throughout the surgery.

Keywords: Arnold-Chiari malformation, Neonate, Neuroanaesthesia, Neural tube defects

CASE REPORT

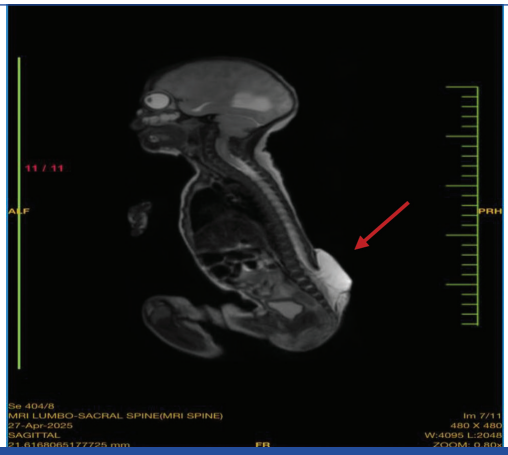
A 15-day-old neonate weighing 2.43 kg born to a gravida 2 para one mother at full-term through caesarean section due to breech presentation was posted for meningomyelocele repair surgery under American Society of Anaesthesiologists (ASA) class III. The Appearance Pulse Grimace Activity Respiration (APGAR) scores were eight at one minute and nine at five minutes after birth. The newborn was immunised to date. Antenatal history was found to be insignificant.

On examination, the head circumference was 32 cm, with normal limb movements, and no neurodeficit was noted. A cystic swelling was noted in the lumbosacral area measuring 5 cm with skin intact. Airway examination revealed a soft palate cleft with adequate mouth opening. All routine investigations were done [Table/Fig-1].

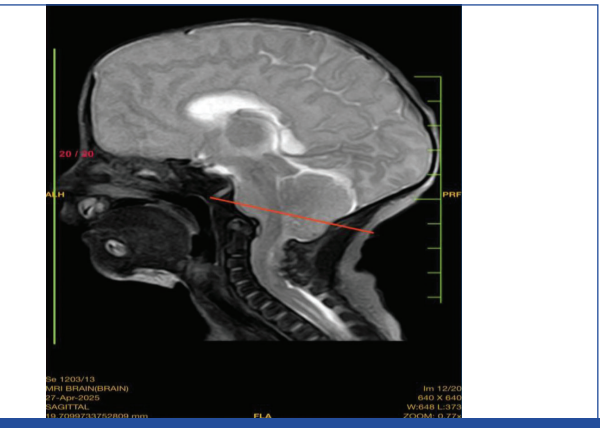
Chest X-ray and electrocardiogram were found to be normal. Two-dimensional echocardiography showed a small patent foramen ovale. Magnetic Resonance Imaging (MRI) of the lumbosacral spine suggested a Lumbosacral (LS) meningomyelocele with syrinx [Table/Fig-2]. MRI brain confirmed the presence of Chiari malformation type II with a herniation of 6 mm caudally [Table/Fig-3].

Laboratory parameters	Value	Normal values
Haemoglobin (mg/dL)	13.1	12-16
Total leucocytes (cells/mm^3)	12000	4500-11000
Platelets (lac/microlitre)	3.4	1.5-4
C-reactive protein (mg/dL)	0.797	< 1
Urea (mg/dL)	19	9-20
Creatinine (mg/dL)	0.6	0.6-1.2
Sodium (meq/L)	139	137-145
Potassium (meq/L)	4.8	3.5-5.1
Total protein (g/dL)	5.9	5.7-8
Albumin (g/dL)	3.5	3.5- 5
Total bilirubin (mg/dL)	9.2	0.2-1.3
Unconjugated bilirubin (mg/dL)	8.8	0-1.1

[Table/Fig-1]: Laboratory investigations.

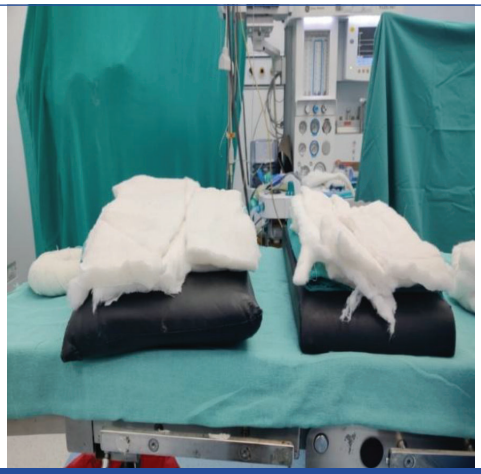


[Table/Fig-2]: MRI LS Spine with arrow showing lumbosacral meningomyelocele with syrinx.



[Table/Fig-3]: MRI brain showing herniation of cerebellum.

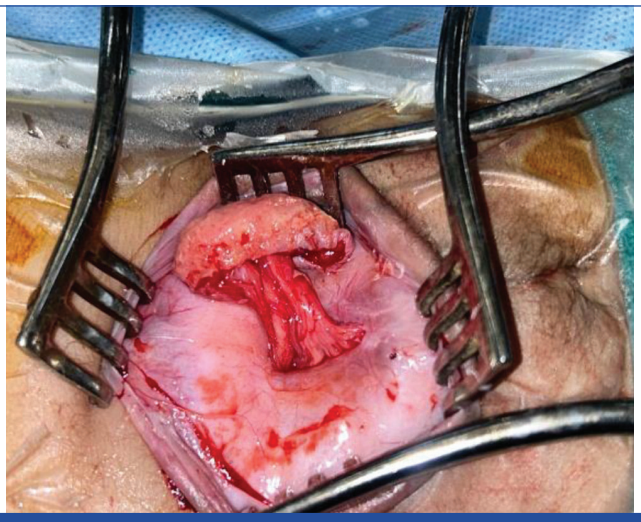
[Table/Fig-2]: MRI LS Spine with arrow showing LS meningomyelocele with syrinx. The neonate was kept nil by mouth for four hours for breast milk and one hour for clear fluids (six hours for formula milk, four hours for breast milk, one hour for clear fluids). A well-written and verbally explained consent was taken one day prior and on the day of the surgery. A 24-gauge intravenous cannula was secured and after ensuring adequate warming of the table up to 40°C for 30 minutes, arranging a difficult airway trolley, the neonate was shifted for surgery. The child was positioned in supine with soft pillow wedges so that there was no pressure on the sac during intubation [Table/Fig-4,5]. All multipara monitors were attached and premedication with intravenous glycopyrrolate 0.004 mg/kg, intravenous midazolam 0.05 mg/kg was given. Induction with sevoflurane at 2% volume concentration, 100% oxygen and intravenous fentanyl 2 µg/kg gave ideal intubating conditions. Gentle laryngoscopy was done with a size 0 curved blade after intravenous atracurium 0.5 mg/kg was given. Airway was secured with a 3.5 mm uncuffed endotracheal tube and fixed at 9 cm after the 5-point auscultation technique and ensuring equal air entry on both sides. Controlled ventilation was set to deliver a tidal volume of 25 mL respiratory rate of 24 breaths/minute, FiO2 60%, post-end expiratory pressure of 5 cmH_2O and peak pressure of 10 cmH_2O. Anaesthesia was maintained by sevoflurane at 2% volume concentration with a minimum alveolar concentration of 2, and oxygen and air with flow rates of 1.5 mL/min each. Intermittent atracurium intravenous boluses at 0.1 mg/kg and fentanyl at 0.5 micrograms/kg/hour were given. Strict fluid management using the Holliday-Segar formula with a deficit of 38 mL corrected over 3 hours with Ringer's lactate (4 mL/kg/hour for the first 10 kg weight, +2 mL/kg/hr for the next 10 kg, +1 mL/kg/hr beyond 20 kg). Care was taken not to inject air bubbles into venous lines. After intubation, the child was placed in the prone position and the tube position was confirmed by auscultation.



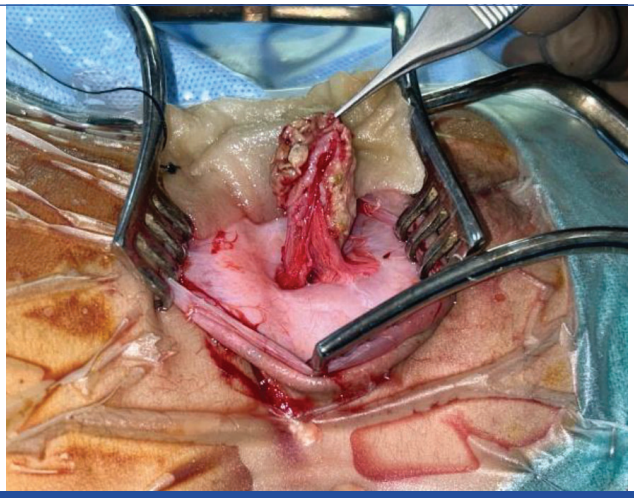
[Table/Fig-4]: Operating table arrangement to accommodate the sac during intubation in the supine position.

[Table/Fig-5]: Intubation in supine position with arrow pointing to the sac being free of pressure.

The surgical repair included sac repair and detethering of the cord [Table/Fig-6,7]. Our institution did not have neuromonitoring facilities. There was a delayed emergence from anaesthesia for at least one hour and intravenous neostigmine (0.05 mg/kg) with glycopyrrolate (0.01 mg/kg) was given. The neonate was extubated after all criteria were met and shifted to the neonatal intensive care unit.



[Table/Fig-6]: Swelling with spinal cord.



[Table/Fig-7]: Detethering of cord.

DISCUSSION

Meningomyelocele is one of the common types of neural tube defects in developing countries, with an incidence of 3.9 per 1000 live births [1]. It can lead to weakness, paralysis, hydrocephalus, bowel and bladder dysfunction, Chiari malformation, etc. Arnold Chiari type 2 consists of structural defects like herniation of cerebellar tonsils. As a result, there can be obstructive hydrocephalus [2]. The intraoperative anaesthetic challenges can be summarised as proper positioning to avoid sac rupture, airway management, eye padding, calculated fluid and drug dosing, and temperature monitoring [3]. In this case, the neonate was prone to hypothermia because of loss of autonomic control below the level of defect and because of less subcutaneous fat, a larger body surface area than adults [2,3]. The surgical steps involve opening the dural sac, detethering the neural tissues, repairing the sac and closure [3].

Differences in airway anatomy include the presence of a big head, large tongue, floppy epiglottis, subglottic narrowing and anteriorly placed larynx. Care must be taken while positioning for intubation. Placement of folded clothes and a donut ring around the lesion is done to avoid rupture and gentle laryngoscopy with minimal neck extension is used to avoid worsening of Chiari malformation [4]. Meningomyelocele is associated with many congenital abnormalities and one such condition is an Atrial Septal Defect (ASD). The two parameters to be considered while providing anaesthesia are

Systemic and Pulmonary Vascular Resistance (SVR, PVR). The goal is to avoid hypoxic shunt, which happens when SVR is less than PVR. This can happen due to bronchospasm, laryngospasm, or hypovolaemia [5].

Three different scenarios can happen:

1) High PVR, Low SVR: hypoxic shunt worsens;
2) Normal PVR, High SVR: increased magnitude of left-to-right shunt;
3) Normal PVR, Low SVR: avoids hypoxic shunt and decreases the magnitude of shunt [5].

Some precautions, like avoiding air bubbles in intravenous lines, less nitrous oxide use, avoiding sympathomimetics and higher fraction of inspired oxygen for longer durations, can help [5].

However, complications like bradycardia, cardiac arrest, brainstem compression, respiratory compromise, and prolonged ventilation can only be anticipated but cannot be prevented [3] Vig S et al., mentioned a case where a two-year-old having sacral meningomyelocele after three hours of surgery had non-awakening from anaesthesia. An urgent Computed Tomography (CT) confirmed an acute exacerbation of Chiari malformation known as foramen magnum syndrome. Foramen magnum syndrome is a very rare but fatal complication of Chiari malformation, which occurs due to excessive Cerebrospinal Fluid (CSF) leak. This can be prevented by the Trendelenburg position during sac repair [6]. Other surgical complications include wound dehiscence and infection, postoperative ileus, CSF leak, and worsening Central Nervous System (CNS) function [7]. Some of the surgical complications can be prevented by using intraoperative nerve monitoring with Electromyography (EMG), Sensory and Motor evoked potentials. The EMG responses help in safe detethering of the cord without nerve damage. Krause M et al., did a study about neurological function outcomes using neuromonitoring intraoperatively and postoperative functional assessment [8]. Apart from ASD, other anomalies like patent foramen ovale, scoliosis, syringomyelia, absent ribs, and cleft palate can also co-exist. Meningomyelocele management should involve a perioperative approach. Attention during the postoperative period involves infection control due to prolonged intubation and ventilation, central venous catheters and synthetic dural grafts [9]. Thaware P et al., discussed a 23-year-old patient with post-neurological sequelae after meningomyelocele repair being posted for femur fracture reduction surgery. He had developed scoliosis with an anticipated difficult airway. There was an awake fibreoptic checkscopy to have an idea of airway anatomy. The procedure was done under dexmedetomidine infusion [10]. Suryaningrat FR et al., did a case series with four cases of ruptured meningomyelocele management during the perioperative period in neonates. It had lumbosacral and thoracic meningomyelocele were observed and surgical complications were confirmed. One of the cases had a few episodes of seizures in the postoperative period due to electrolyte imbalance. All the patients were managed in prone positions [11].

CONCLUSION(S)

In-depth knowledge about paediatric physiology and anticipation of perioperative events, along with proper planning and execution, help provide safe anaesthesia in such high-risk surgeries. Special care during intubation in the supine position to not compress the sac and during ventilation in the prone position must be considered. The plan of anaesthesia must be properly explained to the relatives and routine use of neuromonitoring during spinal cord tethering must be considered. Also, prenatal surgical repairs must be considered as a viable option.

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PARTICULARS OF CONTRIBUTORS:
1. Resident, Department of Anaesthesia, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
2. Resident, Department of Anaesthesia, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Poornima Dhakshinamurthy,
104, Subhadra Towers, Sawangi Meghe, Wardha-442001, Maharashtra, India.
E-mail: poornima.dakshinamurthi@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Oct 09, 2025
- Manual Googling: Nov 22, 2025
- iThenticate Software: Nov 24, 2025 (3%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Oct 08, 2025**
Date of Peer Review: **Oct 17, 2025**
Date of Acceptance: **Nov 26, 2025**
Date of Publishing: **Feb 01, 2026**