TPM Vol. 32, No. S3, 2025
ISSN: 1972-6325
https://www.tpmap.org/

MULTIDISCIPLINARY MANAGEMENT AND SURGICAL OUTCOMES IN NEONATES WITH MYELOMENINGOCELE AND CHIARI II MALFORMATION: A CASE SERIES

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Abstract

Background: Neonatal neurosurgical conditions, including myelomeningocele, Chiari II malformation, hydrocephalus, and occipital encephalocele, pose significant challenges requiring multidisciplinary management. Early diagnosis, surgical intervention, and postoperative care play a crucial role in improving outcomes.

Objective: This case series aims to present the clinical course, surgical management, and outcomes of four neonates diagnosed with congenital neurosurgical conditions, emphasizing key aspects of neonatal intensive care, infection control, and long-term fol`low-up.

Methods: The study included neonates with neurosurgical conditions admitted to the Newborn Care Unit (NICU) at Saveetha Hospital Chennai between Decemeber 2022 and January 2025 , with a follow-up period of six months**.**

Four neonates with congenital anomalies, including cervical and lumbar myelomeningocele, hydrocephalus, Chiari II malformation, neonatal jaundice, and atretic occipital cephalocele, were admitted and managed with neurosurgical intervention, intensive neonatal care, and post-discharge follow-up. Clinical findings, imaging studies, surgical procedures, postoperative care, and outcomes were analyzed.

Results: All four neonates underwent surgical intervention, including myelomeningocele repair, VP shunt placement, and intensive care ,phototherapy for neonatal jaundice. Postoperative complications included sepsis, VP shunt malfunction, and CSF infections, requiring targeted antibiotic therapy and additional neurosurgical interventions. Despite initial complications, all neonates showed clinical improvement, stable growth, and appropriate neurological development at discharge. Long-term follow-up was planned for neurodevelopmental assessment, immunization, and surgical reassessment.

Conclusion: Early identification, prompt surgical management, and meticulous neonatal care are essential for optimizing outcomes in neonates with congenital neurosurgical conditions. A multidisciplinary approach involving neonatologists, neurosurgeons, pediatricians, and infection specialists is critical for reducing morbidity and improving long-term prognosis. This case series highlights the importance of comprehensive neonatal care and structured follow-up in managing complex neurosurgical conditions in infancy.

Keywords: Neonatal neurosurgery, Myelomeningocele, Chiari II malformation, Hydrocephalus, Occipital cephalocele, Neural tube defects, Cerebrospinal fluid leak, Ventriculoperitoneal shunt, Neonatal intensive care, Neurosurgical intervention, Congenital anomalies, Neonatal sepsis, Phototherapy, Tethered cord syndrome, Pediatric neurosurgery.

INTRODUCTION

Neonatal neurosurgical conditions, including neural tube defects, hydrocephalus, and congenital anomalies, require early diagnosis and multidisciplinary management to optimize outcomes. This case series presents four neonates with varying congenital conditions—cervical and lumbar myelomeningocele, Chiari II malformation, hydrocephalus, neonatal jaundice with occipital cephalocele, and associated complications. Each case highlights the importance of prenatal diagnosis, surgical intervention, neonatal intensive care, and long-term follow-up. The series underscores the challenges of managing these conditions, emphasizing the role of timely neurosurgical procedures, infection control, respiratory support, and nutritional care in improving survival and developmental outcomes. Through detailed case analysis, this series provides insights into current management strategies, complications, and prognostic considerations in neonatal neurosurgical care.

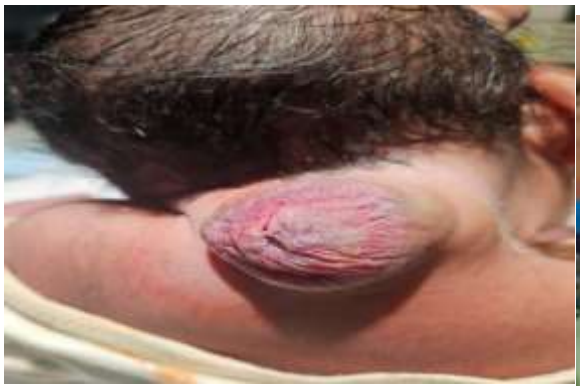
CASE PRESENTION

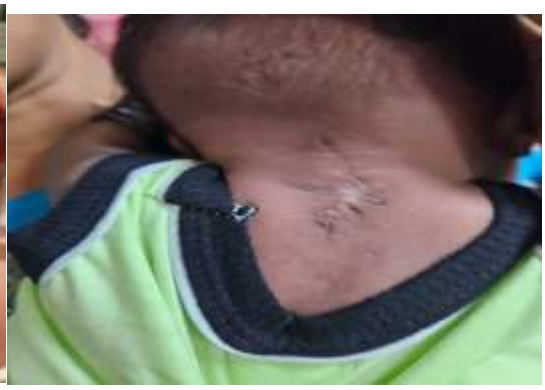
Case 1

A male neonate, was born via LSCS at 37 weeks of gestation due to a non-reactive NST. The birth weight was 2.760 kg, with an Apgar score of 8/10 at 1 minute and 9/10 at 5 minutes. The baby cried immediately after birth, requiring only routine resuscitation. Antenatally , the baby was diagnosed with cervical myelomeningocele (Aperta), confirmed by MRI, which revealed an open myelomeningocele through cervical spinal dysraphism with associated mild colpocephaly and syrinx of the cervical spine. USG findings showed ventriculomegaly with bilateral lateral ventricle dilation.

The baby was admitted to the NICU and underwent surgical repair of myelomeningocele on day 6 of life under intraoperative neuro monitoring. Postoperatively, the baby required invasive ventilation for 16 hours, followed by weaning to NIV on day 7 and room air by day 9. Feeding was initiated through an orogastric tube and gradually progressed to direct breastfeeding. Serial head circumference monitoring was performed, with the last recorded measurement at 36 cm. During the hospital stay, CSF culture revealed *Acinetobacter baumannii*, prompting a course of Inj. Meropenem for 10 days along with oral Ciprofloxacin for a total of 21 days.

At discharge on day 21 of life , the baby was stable, feeding well, and gaining weight, with a recorded weight of 3.190 kg, length of 48 cm, and head circumference of 36.5 cm. Hearing assessments (OAE and BERA) were normal. The discharge plan included exclusive breastfeeding for six months, immunization as per schedule, and regular neonatal and neurosurgery follow-up and CDC follow up for neurodevelopmental assessment





Case -2

A term male infant, was born via LSCS at 37 weeks and 6 days of gestation due to meconium-stained liquor. The birth weight was 2.586 kg, and the APGAR scores were 8/10 at 1 minute and 9/10 at 5 minutes. The baby cried immediately after birth and was shifted to the NICU due to a 3×3 cm cystic swelling in the lumbar region.

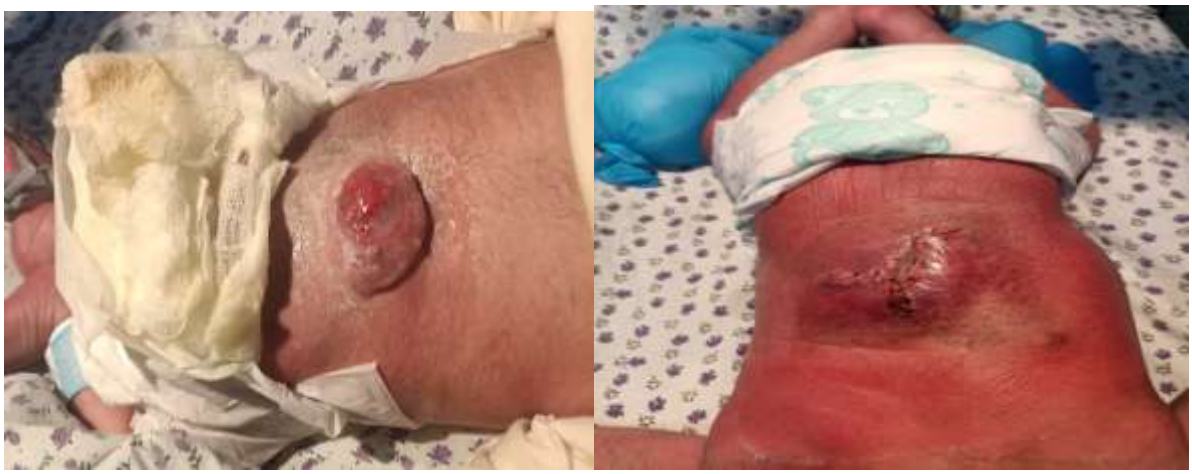
A neurosurgical evaluation led to MRI imaging, which revealed a lumbar lipomeningomyelocele with a tethered cord. No other brain anomalies were detected. The baby underwent myelomeningocele repair with detethering on day 10 of life (29/02/2024). Postoperatively, intravenous antibiotics (Inj. Piptaz) were administered for 5 days, and a surgical drain was placed and removed after 3 days. The recovery was uneventful. Additional screenings included an echocardiogram, which showed a small patent foramen ovale (PFO), an ophthalmologic evaluation that was normal, and a USG KUB that revealed mild renal pelviectasis, warranting a follow-up at 3 months.

The baby was initially fed direct breastfeeding (DBF), with paladai feeds introduced postoperatively. He remained euglycemic and showed appropriate weight gain. At discharge on day 28 of life , the baby weighed 2.720 kg, was feeding well, and was hemodynamically stable with normal oxygen saturation levels.. Follow-up visits were scheduled for neurosurgical, pediatric, and ophthalmologic evaluations, including USG KUB, BERA, and ROP screenings. The parents were counseled on neonatal care, exclusive breastfeeding, and danger signs. review, immunization, and developmental monitoring.

Case -3

A male infant, was born at 39 weeks gestation via elective LSCS due to a previous LSCS. Birth weight was 3.680 kg, and the baby had an APGAR score of 8/10 and 9/10 at 1 and 5 minutes, respectively. Antenatal USG detected an open spinal dysraphism with myelomeningocele and Chiari II malformation. Postnatally, the baby had respiratory distress, requiring NIV, intubation, and gradual weaning to room air by day 16. Echo revealed moderate PAH, necessitating inotropic support which was tapered by day 6. Due to a CSF leak from the myelomeningocele defect, early-onset sepsis was managed with IV antibiotics, including Meropenem, Vancomycin, and Colistin. Surgical repair of the myelomeningocele with ventricular tapping was performed on day 3. Postoperatively, the baby showed improved lower limb power (from 1/5 to 2/5), though tone remained decreased. Dressing was done regularly, and cultures indicated Serratia growth, treated with appropriate antibiotics . At discharge on day 22, the baby was tolerating feeds well, with a head circumference of 38.5 cm. Discharge advice included exclusive breastfeeding, vitamin D3 supplementation, physiotherapy, immunization per schedule, regular HC monitoring, neurosurgical follow-up for potential VP shunt placement if HC exceeds 40 cm, and wound care with dressing every alternate day. Further follow-up includes BERA testing, ROP assessment, and continued physiotherapy.

Baby was readmitted at 88 days of life with VP shunt displacement, wound discharge, and signs of hydrocephalus. The baby had a history of multiple medical conditions, including myelomeningocele, Chiari II malformation, sepsis, pneumonia, and failure to thrive. Initial investigations revealed meningitis due to *Enterobacter cloacae* and *Enterococcus faecalis*, requiring targeted IV antibiotics. Due to shunt failure, an external ventricular drain (EVD) was placed, and a revision VP shunt was inserted on 22/08/2024. The baby required respiratory support, blood transfusion, and nutritional supplementation. Postoperatively, the baby showed clinical improvement, tolerated feeds well, and remained seizure-free. At discharge, the baby was stable, feeding well on direct breastfeeding, with a functioning left VP shunt. Follow-up was advised for neurosurgical team.



Case -4

A term female neonate (AGA) born via elective LSCS at 38 weeks due to breech presentation, had a birth weight of 2.810 kg with good APGAR scores (8/10 at 1 min, 9/10 at 5 min. MRI Brain revealed an atretic occipital cephalocele over a dermoid cyst, with a small solid component and a fibrous stalk connecting to the posterior tentorium through a bony defect. Pediatric surgery and neurosurgery consultations were obtained, and follow-up was advised. USG abdomen showed gallbladder sludge, while cranial USG detected a well-defined cyst in the occipital region.). The baby developed neonatal jaundice with a peak SBR of 18.12 mg/dL on day 5, requiring phototherapy, after which levels reduced to below the cutoff. On discharge at day 7, the baby was stable, gaining weight, and feeding well. Parents were counseled on neonatal care ,follow-ups in pediatric surgery and neurosurgery OPD, as well as OAE and USG hip at six weeks, were advised and **plan for excision at 6months of age**

Summary of Case Series

Case No.	Gestational Age & Birth Details	Diagnosis	Surgical Management	Postoperative Course & Complications	Outcome & Follow-up
1	37 weeks, LSCS (Non-reactive NST), Male, 2.760 kg	Cervical Myelomeningocele (Aperta), Mild Colpocephaly, Syrinx, Ventriculomegaly	Myelomeningocele Repair (Day 6)	Required invasive ventilation (16 hrs), CSF culture: *Acinetobacter baumannii*, Treated with IV Meropenem & Ciprofloxacin	Discharged on Day 21, Stable, Feeding well, Regular Neurosurgical Follow-up
2	37+6 weeks, LSCS (Meconium-stained liquor), Male, 2.586 kg	Lumbar Lipomeningomyelocele, Tethered Cord, Mild Renal Pelviectasis	Myelomeningocele Repair with Detethering (Day 10)	Post-op IV Piptaz (5 days), Small PFO detected, Recovery uneventful	Discharged on Day 28, Stable, Regular Neurosurgical, Pediatric & Ophthalmologic Follow-up
3	39 weeks, LSCS (Previous LSCS), Male, 3.680 kg	Myelomeningocele, Chiari II Malformation, Moderate PAH	Myelomeningocele Repair & Ventricular Tapping (Day 3)	Required NIV, Inotropes for PAH, Early-Onset Sepsis (*Serratia*), Treated with Meropenem, Vancomycin, Colistin	Discharged on Day 22, Stable, VP Shunt Required, Readmitted at 88 Days for Shunt Displacement & Infection
4	38 weeks, LSCS (Breech Presentation), Female, 2.810 kg	Atretic Occipital Cephalocele over Dermoid Cyst, Neonatal Jaundice	No immediate surgery planned	Phototherapy for Jaundice, Gallbladder Sludge on USG, Stable on Exclusive Breastfeeding	Discharged on Day 7, Plan for Excision at 6 Months, Regular Pediatric & Neurosurgical Follow-up Plan for excision at 6 months of age

DISCUSSION

Singh B K et al included total of 25 babies in the study, with the majority born to mothers at a median age of 24 years (range: 19–36), and nearly one-third of the mothers were illiterate. Maternal periconceptional folic acid intake was reported in only five cases (21%). Two-thirds of the babies were male (64%), with a median age at admission of 9 days (range: 1–27). The majority had open neural tube defects (NTDs), with meningomyelocele being the most common type (88%), followed by occipital encephalocele (12%) and one case of a closed NTD with lipomeningomyelocele (4%). Hydrocephalus (76%) was the most frequently associated anomaly, followed by Arnold-Chiari malformation (56%). Motor weakness, including paraparesis or paraplegia, was observed in 84% of cases, while 44% had sensory deficits, and 48% had bowel and bladder dysfunction. Ventriculitis was the most common associated morbidity (38%). The primary surgical interventions performed included meningomyelocele (MMC) repair (33%) and ventriculoperitoneal (VP) shunt placement (24%). Of the total cases, 12 babies (48%) were discharged, 2 (8%) expired, and 11 (44%) left against medical advice.

Andronikou et al. reported five cases, with a majority having myelocystocele or meningocele, and a high incidence (80%) of Chiari type II malformation.

Habibi et al. studied 16 patients, predominantly presenting with stalk-type lesions, with 25% showing Chiari malformation and 50% developing hydrocephalus. Huang et al. examined 10 cases of myelocystocele, with lower rates of Chiari malformation (10%) and hydrocephalus (10%), but 90% of patients had normal neurological function.

Kasliwal et al. reported 10 cases, mostly myelomeningoceles, with 20% presenting Chiari malformation and 50% hydrocephalus, yet all patients retained normal neurological function. Pang and Dias observed nine cases, including myelocystocele and limited dorsal myeloschisis, with moderate rates of Chiari malformation (44.4%) and hydrocephalus (33%). Sun et al. studied eight cases, evenly split between myelocystocele and meningocele, with a higher prevalence of Chiari malformation (62%) and hydrocephalus (62%), though only 75% maintained normal neurological function.

Kıymaz et al . reported seven patients (one male, six female) showed a similar distribution of myelomeningocele and meningocele, with 43% having Chiari malformation and 42% hydrocephalus, while 86% retained normal neurological function. These findings reinforce the variability in presentation and prognosis, emphasizing the importance of early diagnosis and individualized management.

Conclusion

These cases highlight various congenital and neonatal conditions requiring specialized care and surgical intervention. Each infant presented with unique challenges, including neural tube defects, hydrocephalus, neonatal jaundice, and sepsis, necessitating multidisciplinary management. The importance of timely diagnosis, appropriate surgical interventions, and postnatal monitoring was evident in optimizing outcomes for these neonates. Comprehensive discharge planning, including exclusive breastfeeding, immunization, medication adherence, and follow-up with neurosurgical and pediatric teams, played a crucial role in ongoing care. These cases emphasize the significance of early detection, neonatal intensive care, and parental education in managing complex neonatal conditions and ensuring long-term well-being.

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