Anaesthetic Challenges & Management of Myelomeningocele Repair

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Abstract :

Myelomeningocele (meningomyelocele (MMC) or spina bifida) is a form of neural tube defect. Failure of neural tube closure early in intrauterine development results in spectrum of abnormalities ranging from spina bifida occulta, a relatively benign condition to meningomyelocele, an abnormality involving vertebral bodies, spinal cord and brainstem. These babies also have associated congenital anomalies like intestinal malrotation, renal anomalies, cardiac malformations and tracheoesophageal fistula. Anaesthesia for meningomyelocele always poses a challenge to anaesthesiologist as one has to deal with neonates and infants who already have so many anatomical and physiological differences compare to the adults or normal children. **Methodology:** To identify the anaesthetic challenges/ complications during perioperative and postoperative myelomeningocele repair. It was a retrospective study has been carried out on 20 children having age less than 2 year, operated during 1st July 2018 to 1st June 2019 at tertiary care hospital, Ahmednagar. **Results:** Overall complications were seen in 64.9% and the most common complications were respiratory like laryngospasm, bronchospasm, and hypoxemia and endobronchial intubation. Most of the complications were manageable through early and precise diagnosis. **Conclusion:** Children with meningomyelocele are prone to have peri-operative complications, hence Meticulous anaesthetic management is crucial for early repair and to prevent sequele of meningomyelocele.

Keywords: Myelomeningocele, Meningomyelocele, Spina bifida, Hydrocephalus, General anaesthesia

Introduction:

Myelomeningocele (MMC), one of the most common congenital malformation of the central nervous system, has been described as "the most complex treatable congenital anomaly compatible with life."[1] Failure of neural tube formation and closure may occur anywhere along the neural axis. MMC occurs in 0.4-1 per 1000 live birth and incidence varies with environmental and genetic factors.[1,2] In addition to the neural tube defect, several associated anomalies like hydrocephalus concerns the anaesthesiologist. Most infants with hydrocephalus are usually associated with Arnold- Chiari type II malformation (downward Displacement of the cerebellum into the brain stem and cervical canal with medullary kinking). This may cause cervical cord compression, during extension for intubation leading to brain stem compression. Airway management may be difficult in patients with significant hydrocephalus. Most patients show a diminished response to hypoxia, and may be more susceptible to post-operative apnoeic episodes.[4] The effect of positioning in neonates for induction of general anaesthesia must also be considered. No direct pressure should be applied to the exposed neural placode.[3] In addition, children with MMC have a higher incidence of intestinal malformations, renal anomalies, cardiac malformations and tracheoesophageal fistula.[5] Surgical management of MMC is challenging, not only for administration of anaesthesia but also for providing perioperative care owing to paediatric age group, comorbid conditions, and associated systemic abnormalities.[6]

Methodology:

All the available medical records of children who underwent excision and repair of MMC over a period of one years (July 2018 to June 2019) were analysed. For each child, data were collected by detailed review of records related to pre-anaesthetic evaluation, intraoperative course and postoperative complications. Pre-operative evaluation included age, sex, weight, height, Hb%, electrolytes, chest x-ray, site of lesion, CSF leak from the sac, neurological presentation, associated systemic abnormalities, venous and airway Assessment.

Inclusion Criteria :

1.Patients diagnosed as Myelomeningocele

2.Patients up to 2 year of age.

Exclusion Criteria :

1.Patients above 2 year of age.

All the patient had their MRI and CT scan of relevant part to see the extent of disease.**(Fig.1&2)** Intraoperative data included anaesthetic technique, intraoperative monitoring (heart rate, saturation, ventilation, BP, Urine output, temperature), fluids infused, blood loss and transfusion. Intraoperative parameter such as heart rate and blood pressure were recorded. Respiratory complications (hypoxemia, hypercarbia, bronchospasm, laryngospasm), endobronchial intubation, accidental extubations were noted. Miscellaneous complications like hypothermia, dislodgement of intravenous catheter were reviewed. Post-operative complications like nausea, vomiting, mechanical ventilation, surgical complications were reviewed.

Fig 1:

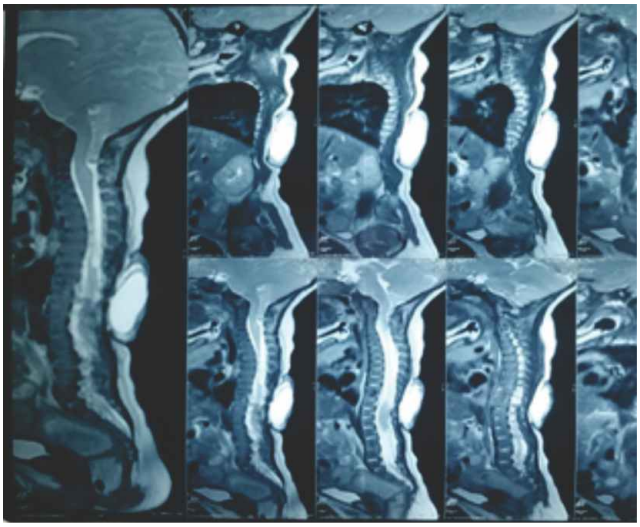

Fig 2:

Fig 1 & 2: Thoracic Myelomeningocele clinical & MRI pictures

Anaesthetic Technique : A standard anaesthetic technique was followed for all children. Intravenous line was secured with or without sedation. Those who needed sedation were given midazolam 0.25-5mg/kg body weight. Intravenous induction was done with thiopental sodium (3-5 mg/kg). Vecuronium was used to facilitate tracheal intubation. The intubation was performed mostly in supine position putting placode within the padding made in the shape of a doughnut. In patients associated with large hydrocephalus pillow behind shoulders were kept during intubation. Intubations in lateral position (5 cases) were done in patients with large MMC. Repair of MMC was done in prone position whereas in patients with large hydrocephalus VP Shunt was inserted in supine position followed by MMC repair in prone position. Anaesthesia was maintained with oxygen and Isoflurane with controlled Ventilation. Meperidine was used for analgesia. Isolite-P was the fluid of choice in most patients, Isolite -P were also used especially in neonates. At the end of surgery, residual neuromuscular block was reversed with calculated Dose of Neostigmine and atropine and the trachea was extubated when the patient fully awake.

Results :

Table 1: Age & Sex of the Children

Age	Sex		Total no.	Percentage
	Male	Female		
0-60 days	1	1	2	10
>60 days - 1 year	8	1	9	45
1 year - 2 year	3	6	9	45
Total	12 (60%)	8 (40%)	20	100

Table 2: Site of Myelomeningocele

Site	No. (%)
Lumber MMC	13 (65)
Lumbosacral	5 (25)
Cervical	1 (5)
Thoracic	1 (5)
Total	20

Table 3 : Associated Abnormalities

Abnormalities	No. (%)
Central nervous system abnormalities	
Hydrocephalus	6 (30%)
Chiari malformation	2 (10%)
Orthopaedic abnormalities	
Scoliosis	6 (30%)
Talipes Equino varus	4 (20%)
Flat/ high arched foot and tropic Ulcer	5 (25%)
Co-existing medical conditions	
Upper respiratory tract infections	5 (25%)
Electrolyte imbalance	5 (25%)

Table 4: Intra-Operative Complications

Complications	Intra-operative	No(%)
Cardiovascular		
Bradycardia	2	10
Tachycardia	1	5
Hypotension	1	5
Respiratory		
Bronchospasm	1	5
Hypoxemia	1	5
Endobronchial Intubation	1	5
Accidental Extubation	1	5
Laryngospasm	1	5

Table 5 : Post-operative complications

Complications	No (%)
Hydrocephalus	2 (10)
CSF leak	1 (5)
Wound infection	3 (15)
Wound dehiscence	2 (10)
Death	3(15)

Discussion :

MMC is the most common malformation of the CNS worldwide, with an incidence evaluated at 0.4-1 per 1000 live births, and is one of the leading causes of infantile paralysis along with lifelong disabilities including paraplegia, hydrocephalus, Chiari II malformation, incontinence, sexual dysfunction, skeletal deformities and mental impairment.[7] In our study out of a total 20 children operated, majority 90% (n=18) were above 60 days of age, 10% (n=2) were less than 60 days old and remaining patients were older than one year.**(Table 1)** Male child (n=12) were operated more than female child (n=8) with male : female ratio of 3:2. The study done in rural areas from Jaunpur to Ghazipur district in Eastern Uttar Pradesh show that the 19% cases of MMC recorded, the frequencies of a male child affected to that of the female child were in the a ratio of 1.38:1.[3]

The most common location of neural tube defect was found to be lumbar spine 65% (n=13) followed by lumbo sacral 25% (n=5) cervical and thoracic 1 each**(Table 2)**. Gok et al. study concluded that out of the 8 reported patients, most common location is lumbar site 75% & study conducted by Mowatt DJ et al found Large MMC was located more in the thoraco-lumbar area (67.5%).[4, 6]

Associated abnormalities **(Table 3)** Hydrocephalus was found to be most common seen in 6 (30%) children, who underwent ventriculoperitoneal shunt followed by MMC repair in the same setting. Chiari malformation II was found in 2 (10%) patients, all in association with hydrocephalus. Scoliosis seen in 6 cases (30%) diagnosed clinically by features of uneven shoulders, prominent ribs and typical look of the hip where one hip was higher than the other. Electrolyte imbalance was present in 5 (25%) of patient. Stephen A. Back study show that Hydrocephalus develops in 60% of patients with occipital, cervical, thoracic, or sacral lesions. Chiari malformation II was present in in 25% patients. Scoliosis present in 40% of patient.[5]

Among the 12 intraoperative complications respiratory complications was the most common as seen in 7 cases **(Table 4)**. Among respiratory complications Bronchospasm, hypoxemia, laryngospasm, accidental extubation were more common. Endobronchial intubation was found in 2 cases. Cardiac complications in the form of bradycardia, tachycardia and hypotension created problems during the operative procedure. Mowatt DJ, Thomson DN study Large MMC was located more in the thoraco-lumbar area (67.5%); 90% of the children showed severe motor deficit and 90% of them required shunt surgery, and 40% having severe respiratory problem and 30% having cardiac problem.[6]

Post-operative complications included wound infection 3(15%), hydrocephalus 2(10%), wound dehiscence 2 (10%), postoperative CSF leaking from repair 1(5%) were recorded. These postoperative complications were recorded as those cases had to undergo various revision operative procedures like, resuturing for wound dehiscence, debridement of wound infection, VP shunting for HCP, exploration of leaking CSF **(Table 5)**. Robinson S, Cohen AR et al compare to this study wound infection 9% is the most common complication, then hydrocephalus 4%, dehiscence 3% and CSF leak occur in 2% of patient.[8]

Conclusion :

The children with MMC have several associated CNS abnormalities which are very prone to have perioperative complications. Most of the complication manageable through early and precise diagnosis, meticulous preoperative preparations, vigilant intraoperative monitoring and preparation for anticipated complication.

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