



Original Article

MANAGEMENT OF MYELOMENINGOCELE : EIGHT YEARS EXPERIENCE IN DHAKA MEDICAL COLLEGE AND HOSPITAL

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Abstract

Introduction: Myelomeningocele is a single most common congenital malformation that affects the entire central nervous system and because of extensive internal CNS involvement, its management remains controversial. Proper management of affected children can lead to a meaningful and productive life, and poorly managed cases of myelomeningocele can be a devastating obstacle not only for patient but also for the patient's family.

Materials and methods: This was a descriptive study, started in January, 2007 and completed in December 2014 in the Department of Pediatric Surgery and Department of Neurosurgery of Dhaka Medical College & Hospital (DMCH). The medical records of 42 children with myelomeningocele operated at our center were reviewed retrospectively. The surgical & medical management protocol used for different sites of myelomeningocele was studied.

Results: The age of most of the patients at the time of myelomeningocele repair was between 45 days to 60 days; however, children with ruptured myelomeningocele were consistently repaired early after controlling infection. In our study, 16 patients (38.1%) of myelomeningocele with

hydrocephalus were operated for medium pressure ventriculoperitoneal (VP) shunt first, and then repair after 7 days to 10 days. VP shunt was done 7 days after repair of myelomeningocele in 11 patients (26.19%), who developed hydrocephalus postoperatively. All 8 patients with ruptured myelomeningocele (19.05%) were treated for ventriculitis first, then by repair. Seven patients (16.67%) with intact lesion were repaired between 4 weeks to 8 weeks of age.

Complications including CSF leak, wound infection, wound dehiscence, paraplegia, autonomic incontinence, shunt infection and malfunction after repair of myelomeningocele occurred in 27 patients (64.29%). Six patients (14.29%) died during treatment protocol. All the patients were followed up postoperatively for 6 months to 1 year.

Conclusion: Surgical intervention with or without VP shunt of patients with myelomeningocele produced good results.

Key words: Myelomeningocele; Repair; V-P shunt; Postoperative complications.

Introduction:

Myelomeningocele is the most common form of neural tube defect (NTD), a term used synonymously with spina bifida aperta, spinabifida cystica that affects the entire central nervous system and because of extensive internal CNS involvement, its management remains controversial¹⁻⁶. On the other hand, the potential for prevention of the myelomeningocele like neural tube defects in the developing countries, is still far from being fulfilled and there is great need to increase awareness among the general public regarding etiological factors of this disastrous anomaly and its prevention by appropriate prenatal care. Other needs to address are availability of medical facilities and legal, moral, economical, social

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and humanitarian issues. Proper management of affected children can lead to a meaningful and productive life, and poorly managed cases of myelomeningocele can be a devastating obstacle not only for patient but also for the patient's family. The management plan must address some important issues before implementation such as which of the patients will benefit from treatment, the timing of repair, the diagnosis of ventriculitis and its optimal treatment, identification of hydrocephalus and the best time and the best technique for CSF shunting. Initially, the ventricles may be normal or only slightly enlarged. However, after the NTD is closed, the ventricles often enlarge. The incidence of hydrocephalus associated with myelomeningocele ranges from 80% to 95%. The world literature is insufficient for accurate prediction of long-term prognosis for children with myelomeningocele and for formulary the comprehensive parameter regarding treatment indications. We are going to share our experience in the management of myelomeningocele with other health professionals.

Materials and methods:

The medical records of all children with myelomeningocele treated at DMCH during the year 2007- 2014 were reviewed retrospectively. The records of the patients were analyzed for sex, location and size of lesion, CSF leakage, head size at birth and at the time of procedures, presence of hydrocephalus, type of surgery either repair or CSF shunting, use of prophylactic antibiotic, postoperative complications, and total hospital stay. After admission, a thorough physical examination was performed that includes measurement of head circumference, anal sphincter function (anocutaneous reflex or anal wink); upper and lower extremity motor and sensory function; the site, level, and size of the myelomeningocele defect; and whether or not the defect was leaking CSF. The defect size in our series was an average 25.9 cm². In addition, the neonate was evaluated for signs and symptoms of hydrocephalus and other associated anomalies. The myelomeningocele defect was covered with sterile saline-soaked gauze to prevent desiccation of the exposed neural tissue. Broad-spectrum antibiotics were administered to reduce the risk of CNS infection. Head ultrasonography or computed tomography (CT) was performed in all the cases to evaluate the extent of ventricular enlargement and determine the need for shunt placement. Prophylactic

antibiotics were used with daily dressing of myelomeningocele depending upon the condition of skin over the lesion. CSF examinations were performed in only those cases where skin cover over the lesion was either infected or exhibited leakage. Myelomeningoceles were operated urgently in cases with evidence of CSF leakage after treating the ventriculitis depending upon CSF analysis. Repair was carried out using standard technique³ (separation of the neural placode from the intermediate zone of epithelium and reconstruction of the placode into a tube with preservation of all neural tissue; separation of the dura from the epidural space at the lateral margins of the defect and closure of the dura in a watertight but patulous fashion around the newly created NT, mobilization and midline approximation of the paraspinal muscles and fascia; and tension-free closure of the skin in the midline). Myelomeningoceles with hydrocephalus (n=16 - 38.1%) were treated with medium pressure VP shunt initially and then, repair after 7 days to 10 days. All other cases were repaired with standard procedures. Those who developed hydrocephalus postoperatively, treated with VP shunt insertion after 7 days of repair.

Results:

In our series of 42 cases there were 25 (59.52%) boys and 17 (40.48%) girls. The defect was located in the lumbosacral area 33 (78.57%) cases, in thoracolumbar area 2 (4.76%) cases, and the thoracic area 7 (16.67%). The defect size was an average 25.9 cm². The membrane was intact in 34 cases (80.95%) and ruptured in 08 cases (19.05%).

Table I

Frequency of Myelomeningocele according to region

Region	Percentage
Thoracic	07(16.67%)
Thoracolumbar	02 (4.76%)
Lumbosacral	33 (78.57%)

Table-II

Mode of presentation

Myelomeningocele	Percentage
Intact	34 (80.95%)
Ruptured	08 (19.05%)

We treated 08 (19.05%) cases of ruptured lesion with either free flow of CSF or minor leak having signs of ventriculitis with antibiotics first for controlling the infection, followed by repair as soon as possible. Hydrocephalus was diagnosed in 16 cases (38.1%) either before admission to hospital or after hospitalization, and all were treated with the insertion of VP shunt first, and then repair after 7 days to 10 days. In 26 cases (61.9%), hydrocephalus was absent initially but developed postoperatively in 11 cases (26.19%) where the shunt was placed after 7 days of repair. All other myelomeningoceles were repaired with standard techniques at an average age of 6 weeks. The average duration of myelomeningocele repair including anesthesia was 1 hour 30 minutes. All patients received prophylactic antibiotics. Six patients (14.29%) died in this series during treatment period.

Table-III
Associated hydrocephalus

Hydrocephalus	Percentage
Present	16 (38.1%)
Absent	26 (61.9%)

Table-IV
Postoperative complications

Complications	Percentage
CSF leak	23 (54.76%)
Wound infection and wound dehiscence	09 (21.43%)
Paraplegia	11(26.19%)
Autonomic incontinence	13 (30.95%)
Shunt infection	07 (16.67%)
Shunt malfunction	05 (11.90%)

Postoperative complications including CSF leak, wound infection and wound dehiscence, paraplegia, autonomic incontinence, shunt infection and malfunction occurred in 27 cases (64.29%). Uneventful postoperative period was observed in 09 patients (21.43%) and 06 patients (14.29%) died during postoperative period. The average hospital stay for all the cases was 15 days.

Discussion:

Developing countries are facing many problems; socioeconomic, cultural, educational and nutritional

that result in congenital anomalies of CNS such as NTDs more frequently than developed countries like the USA or Europe. NTD involves entire central nervous system and leads to disability or death. The most common form of neural tube defect is myelomeningocele, a term used synonymously with spina bifida aperta, spinabifida cystica and open neural tube defect. The prevalence of myelomeningocele has declined in developed countries of the world owing to both prenatal folate supplementation and to pregnancy termination following prenatal diagnosis. In United States before 1980 prevalence of myelomeningocele was 1-2/1000 live births, but more recently prevalence has declined to 0.44 per 1000 live births.^{11,12} Unfortunately, in developing countries prevalence is much higher, and acceptable prevalence data are not available, nor has the issue been addressed with the goal of eradication, or reduction of incidence. The cause of myelomeningocele is multifactorial and includes genetic predisposition, nutritional deficiencies, particularly folate and zinc, use of anti-epileptic drugs like carbamazepine or valproic acid, diabetes mellitus (type-1), pre-pregnancy obesity and possibility other non medical factors such as agricultural pesticides, radiation, hyperthermia and use of tobacco or drugs. In our series all patients had undergone CT scan of brain and US of the myelomeningocele to determine ventricular size, size of the lesion and other anomalies like abnormal shaped mid brain. We had 02 (4.76%) myelomeningoceles located at thoracolumbar region, 33 (78.57%) at the lumbosacral region, and 7 cases (16.67%) at thoracic region, and 16 cases (38.1%) associated with hydrocephalus. Our aim was to assure an independent and productive life for patients with myelomeningocele and their families with reference to normal intelligence, sphincter control, and power in lower limbs and other spinal anomalies like scoliosis and kyphosis due to tethering of the cord. It is generally accepted that repair ideally should be performed 72 hours after birth to avoid ventriculitis.^{4,5} In our series 08 cases (19.05%) were repaired early because of CSF leak. In rest of the cases, time of repair was between 45 days to 60 days, due to delayed referral of the cases as well as giving time to develop surrounding muscles and fascia to cover the gap. All cases were treated with antibiotics before repair. In all infected cases shunt surgery was delayed, until investigation and treatment for ventriculitis was successful. In our series, myelomeningoceles with hydrocephalus (n=16 -

38.1%) were treated with medium pressure VP shunt initially and then, repair after 7 days to 10 days. All other cases were repaired with standard procedures. Those who developed hydrocephalus postoperatively, treated with VP shunt insertion after 7 days of repair. The main purpose of myelomeningocele repair is to protect the functional tissue in the neural placode, to prevent loss of CSF and minimize the risk of meningitis by reconstructing the neural tube and its coverings with a stable soft tissue closure. To avoid complications, use of lumbar periosteal turn over flap and tissue expansion for delayed closure of large myelomeningocele has been advocated.^{3,12} Recently, rectal monitoring during repair of myelomeningocele has been performed to preserve neural tissues.⁷

In our series, postoperative CSF leak was found in 23 cases (54.76%), wound infection and wound dehiscence in 09 cases (21.43%), paraplegia in 11 cases (26.19%), autonomic incontinence in 13 patients (30.95%), shunt infection in 07 (16.67%) and shunt malfunction in 05 cases (11.90%). All the patients were followed up postoperatively for 6 months to 1 year. In our follow up we had not detect deterioration of neurological function in neurologically intact group.

Conclusions:

Children born with myelomeningocele usually have paralysis or weakness of the muscles below the level of their spine where the defect is. They also may not be able to control their bladder or bowels. Repair of a myelomeningocele is needed to prevent infection and further injury to the child's spinal cord and nerves. There is strong need of public health awareness program in all under developed countries with special reference to nutritional needs like folic acid deficiency. Early surgical repair of myelomeningocele with or without VP shunts shows very encouraging results.

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