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## Recommended Citation

Sattar M. Hashim, A., Ahmed, S., Jooma, R. (2008). Management of myelomeningocele. *Journal of Surgery Pakistan*, 13(1), 7-11.

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# MANAGEMENT OF MYELOMENINGOCELE

A SATTAR M. HASHIM, SHAHID AHMED, RASHID JOOMA

## ABSTRACT

- Objective* T To find out best possible protocol to provide productive life to children born with myelomeningocele
- Study design* Descriptive study
- Place & Duration of study* Department of Neurosurgery, Jinnah Postgraduate Medical Centre Karachi from December 2001 to December 2006.
- Patients and Methods* The medical record of 415 children with myelomeningocele operated at our center was 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 25-30 days; however, children with ruptured myelomeningocele were consistently repaired early. All paraplegic patients with dorsolumbar myelomeningocele were treated with either a low-pressure ventriculoperitoneal (VP) shunt only, direct repair or both. Patients with cervical, dorsal and lumbo-sacral myelomeningocele requiring VP shunt were operated either simultaneously for both procedures or with delayed insertions of a VP shunt after treatment of ventriculitis. All 16 patients with ruptured myelomeningocele (3.8%) were treated for repair as well as ventriculitis. Complications including CSF leak, wound infection or necrosis after repair of myelomeningocele occurred in 22 cases (5.3%). The postoperative follow up for all patients was 1- 2 years.
- Conclusions* Surgical intervention with a low-pressure VP shunt in large dorsolumbar myelomeningocele produced good results.
- Key words* Myelomeningocele, Spina bifida, Ventriculitis. Hydrocephalus.

## 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.<sup>1-10</sup> On the other hand, the potential for prevention of the myelomeningocele like neural tube defects in the third world 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 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.

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The management plan must address some important issues before implementation such as which of the patients will benefit from treatment, the timing of repair, which will be the best technique, the diagnosis of ventriculitis and its optimal treatment, identification of hydrocephalus and the best time and the best technique for CSF shunting.

We offered operative treatment to nearly every patient with just a few exceptions. 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 decided to share our experience in the management of myelomeningocele with other workers.

#### **PATIENTS AND METHODS:**

The medical record of all children with myelomeningocele treated at Jinnah Postgraduate Medical Centre during the year 2001- 2006 were reviewed retrospectively. The records of the patients were analyzed for sex, birth weight, location and size of lesion, CSF leakage, head size at birth and at the time of procedures, presence of hydrocephalus, timing of surgery either repair or CSF shunting, use of prophylactic antibiotic, presence of ventriculitis, nature of organisms, complications of wound healing, morbidity and mortality and total hospitalization time. After admission, prophylactic antibiotics were used with daily dressing of meningocele 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 and treated for ventriculitis depending upon CSF analysis.

Myelomeningocele repair was carried out using standard technique.<sup>4,5,6</sup> Large dorsal myelomeningoceles with myelodysplasia (n=155 - 37.3%) where standard procedure was difficult, were treated with low pressure VP shunt and repair 90 (58%); only low pressure VP shunt in 52 (33.5%) cases and meningocele-peritoneal shunt implemented in 13 (8.38%) cases. All other cases were repaired with standard procedures with VP shunt insertion simultaneously or after observation for 30 days or on confirmation of CT scan in 365 (88%) cases.

#### **RESULTS :**

In our series of 415 cases there were 205 (49.3%) boys and 210 (50.7%) girls. The defect was located in cervical area 21 (5%) cases; in the thoracic area 29(7%), throacolumbar area 340 (82%) cases, and in lower lumbosacral area 26 (61%) cases. In the thoracolumbar region out of 350 cases, 155 (37.35%) were paraplegic or exhibiting myelodysplasia and in 195 (46%) cases lower limb power was intact. The size of lesion, including the thoracolumbar region was measured in 150 cases; (the average area was 35.5 cm<sup>2</sup>). We treated 16 (3.8%) cases of ruptured lesion with either free flow of CSF or minor leak

having signs of ventriculitis, and they were repaired on emergency basis and treated with antibiotics.

All the myelomeningoceles were repaired with standard techniques at average age of 4 weeks due to either delayed referral from distant areas or due to infection that needed treatment before repair. The protocol was different in 155 (37.3%) cases located at thoracolumbar region having myelodysplasia (power in lower limbs grade 0-2). In this series 95 (61%) cases were repaired with standard techniques and simultaneous VP shunt procedure. In 48 cases (30.9%) where the defect was un-repairable we inserted a low-pressure VP shunt and left the lesion to be drained through ventricular system. In 12 (7.7%) cases of un-repairable large lesions we inserted a lumbo-peritoneal shunt to reduce the size.

The average duration of myelomeningocele repair including anaesthesia was 1 hour 20 minutes. All patients received prophylactic antibiotics. Postoperative complications including CSF leak, skin necrosis, and wound infection occurred in 22 cases (5.3%). The VP shunt infection was diagnosed in 17 cases (4%) and the most common organisms cultured were staphylococcus epidermidis, Klebsiella and Escherichia coli species. Postoperative suture removal time was delayed to 15 days in all cases except where skin flaps were closed without tension. The average hospitalization time for all the cases was 15 days.

Hydrocephalus was diagnosed in 365 cases (88%) either before admission to hospital or after hospitalization, and all were treated with the insertion of a ventriculoperitoneal shunt. In 291 cases (79.7%) the shunt was placed simultaneously before repair of myelomeningocele at one sitting and in 74 cases (20.2%) the VP shunt was placed after repair and observation of subsequent hydrocephalus with an average postoperative interval of 45 days. Five patients in this series died.

#### **DISCUSSION:**

Neurosurgeons need to refocus not only on the view under microscope but also on a view outside microscopic field of vision. We should focus on the disorders that affect large numbers of children.<sup>1</sup> Third world countries are facing many problems; socioeconomic, cultural, educational and nutritional that result in congenital anomalies of CNS like neural tube defects (NTD) more frequently than developed societies like the USA or Europe. NTD involves entire central nervous system and leads to disability or death. Children are the most valuable individuals in the lives of most people. Nothing else can cause the grief that the death of the child does, and few disasters cause the grief of a permanently brain damaged child.<sup>10-14</sup>

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.

As the term implies, there is some form of cyst apparent, even if it collapses shortly after birth, It occurs because spinal cord fails to fuse dorsally during primary neurulation during days 18-27 of human embryo genesis leaving a flat plate of neural tissue called a neural placode.<sup>4, 5, 10</sup>

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.<sup>4,15,16,17</sup> Unfortunately, in third world 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.

The defect involve neural tissue. Skin over the cystic lesion is not fully developed. The defect size in our series was an average 30.9 cms. In our series all patient had undergone ultrasonography to determine ventricular size and other anomalies like abnormal shaped mid brain; elongated cerebellum and obliteration of cisterna magna, characteristic of Chiari-II malformation. CT scan of brain and MRI of myelomeningocele were also done in selected cases i.e. 109(26%) of total cases, to exclude associated anomalies and determine the exact location and size of lesion. The role of MRI has been well documented.<sup>7,14,18,19,20</sup>

In our series 155 (37.3%) patients having myelodysplasia were paraplegic and incontinent. We repaired the lesion and placed low-pressure VP shunt, placed low-pressure VP shunt without repair and placed lumbo peritoneal shunt in some cases. Recently cord transaction has been done for repetitive symptomatic tethering to overcome severe pain in the mid thoracic and lumbar region. The results are encouraging at later stage of life to avoid delayed complications like spasticity and scoliosis.<sup>20, 21</sup>

In our series 340 (82%) myelomeningoceles were located at thoracolumbar region and 365 (88%) of the cases investigated showed presence of hydrocephalus and Chiari-II malformation. Twenty one (5%) were located in cervical region, and 29 cases (7%) in thoracic region, and non of them was associated with hydrocephalus or other anomalies. Our aim was to assure an independent and productive life for patients with myelomeningocele and their families with reference to normal intelligence, sphincter control, power in lower limbs and other spinal anomalies like scoliosis and kyphosis due to tethering of the cord.

We examined different parameters in detail between patients who either developed or came to hospital with complications like ventriculitis and those who did not. As for as normal intelligence is concerned we found ventriculitis as the single most important factor, diagnosed at different stages during the care of patient, either before repair of myelomeningocele, after repair, before VP shunt insertion or after placement of VP shunt. In our series ventriculitis was present in 43 cases (10%); 26 cases (61%) had ventriculitis related to myelomeningocele repair and in 17 cases (4%) due to VP shunt. Organisms found in these cases were usually gastro intestinal tract or skin colonizers. Almost all patients with ventriculitis suffered mental retardation and are under continued follow up.

We encountered 16 (3.8%) cases with ruptured myelomeningocele either due to birth trauma, a very thin placode and massive hydrocephalus or due to mishandling by medical professionals. One of the most important issues concerned with management protocol is the optimal timing of myelomeningocele repair. It is generally accepted that repair ideally should be performed 72 hours after birth to avoid ventriculitis.<sup>4,5</sup> In our series 16 cases 3.8% were repaired up to 48-72 hours because of CSF leak. In rest of the cases average time of repair was 28 days, due to delayed referral of the cases. All cases had cultures taken from the neural placode and only sterile cases were directly repaired; others were treated with antibiotics before repair. In all infected cases shunt surgery was delayed, until investigation and treatment for ventriculitis was successful.

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.<sup>17,6,4</sup> Recently, rectal monitoring during repair of myelomeningocele has been performed to preserve neural tissues.<sup>11</sup> In our series all cases were repaired with standard surgical techniques except for large dorsal myelomeningoceles with myelodysplasia. In 52 of 155 cases (33.5%) only a low-pressure VP shunt was inserted without repair of meningocele. There was satisfactory experience 100% shrinkage of myelomeningocele, saving operation time, deterring problem with wound closure due to very thin and inadequate skin, and minimize hospital stay due to poor wound healing. However, neurological deficits were irreversible because of cord tethering.<sup>12</sup> In 13 cases (8.5%) out of 155 with myelodysplasia and large sized myelomeningocele a lumbo-peritoneal shunt was inserted. All other cases were repaired with standard procedure; in 365 cases (88%) had a VP shunt.

In our opinion the choice of surgical method should be individualized based on shape, size and location of defect. In most circumstances meningocele resolve after C.S.F

diversion. Restoration of neural C.S.F dynamics can reduce flow into the sac resulting in meningocele size reduction. In our series there were wound healing problems including skin necrosis CSF leakage, and gross wound infection in 22 cases (5.3%). There was strong association of poor wound healing with ventriculitis. Out of total cases, 26 (6%) had ventriculitis, and in our experience the incidence of ventriculitis was strongly associated with wound infection of the myelomeningocele closure. There were VP shunt infections in 17 cases (4%) only and the rate was more common in patients previously treated for ventriculitis. There was a close relationship between wound healing and placement of VP shunt. The incidence of myelomeningocele repair breakdown was very low because of our protocol wherein all patients with hydrocephalus had a VP shunt before repair or simultaneously. To avoid C.S.F leak other factors were also considered like skin flaps; to avoid tension at skin edges.

In our series hydrocephalus was present in 365 case ((88%) and all were shunted. The incidence of ventriculitis in these patients after shunt insertion was present in 17 cases (4%). Different factors observed were responsible for shunt related infection including, age at shunt placement, local condition of scalp, duration of shunt surgery, use of prophylactic antibiotics and general condition of the patient.

Over all results in our series were encouraging; there were 5 deaths (1.2%) due to ventriculitis, ruptured meningocele and shunt infection. All other patients are under follow up and their associated anomalies such as hydrocephalus, Chiari malformation and other bony anomalies are being looked after with consultation from orthopedic surgeons. Following repair of myelomeningocele there is the possibility of tethered cord of delayed onset leading to neurological and urologic deficits.<sup>23,24,25</sup> In our follow up we have not detected deterioration of neurological function in our neurologically intact group.

#### CONCLUSIONS:

There is strong need of public health awareness programme in all under developed countries with special reference to nutritional needs like folic acid deficiency. Prenatal diagnosis with high resolution fetal ultrasonography should be done and the site and size should be determined for a recommendation of termination of pregnancy. Early surgical repair must be done to avoid further complications. Our method constructs a secure and water tight covering over spinal cord, provides durable soft tissue closure, and may be used in conjunction with other traditional skin or muscle flap techniques. Surgical treatment with a low-pressure VP shunts in patients with myelodysplasia having large dorsolumbar defects showed very encouraging results as compared to lumbo peritoneal shunts in these patients.

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