SHORT TERM OUTCOME OF SURGICAL MANAGEMENT OF PATIENTS WITH NEURAL TUBE DEFECT (SPINA BIFIDA)

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ABSTRACT

Objective: *To study short term out come of surgical management of patients with neural tube defect (Spina Bifida).*

Material and Methods: *This study was conducted at Paediatric Surgery unit, Lady Reading Hospital, Peshawar from January 2003 to December 2005. Patients with neural tube defect in whom surgery was indicated were included in this study except those who had complete paraplegia, total incontinence of urine and faeces and associated life threatening major congenital malformation. After discharge from hospital the patients were followed up for 2.5 years.*

Results: *Surgery was performed in 198 cases including 93 (46.97%) males and 105 (53.03%) females, ranging in age from one day to 18 months. There were 39 cases (19.70%) with meningocele, 144 cases (72.73%) with myelomeningocele, and 15 cases (7.58%) with lipomeningocele. Hydrocephalus observed in 42 cases (21.21%), was treated with V.P shunt. Clubfeet deformities due to neurological defects were found in 12 cases (6.06%). Incontinence of urine and faeces was found in 39 cases (19.70%). Postoperative complications were observed in 24 cases (12.12%) including wound infection 18 cases (9.09%), wound dehiscence 3 cases (1.52%) and CSF leakage 3 cases (1.52%). Thirty four patients died (17.17%) and 27 patients were lost to follow-up.*

Conclusion: *Management of neural tube defect requires proper assessment and appropriate decision for surgery. Proper selection of patients for surgery has a fair outcome*

Key Words: *Neural tube defects, Meningocele, Mylomeningocele, Lipomeningocele, management.*

INTRODUCTION

Spina bifida is a congenital defect of the spinal cord due to failure of fusion of vertebral arches with or without protrusion and dysplasia of the spinal cord and meninges.[1,2] The incidence of spina bifida is 3-5 per 1000 births, but there are wide geographical variations ranging from 1-2 per 1000 to 4-5 per 10003. Due to improved antenatal diagnosis and termination of pregnancy, the incidence is gradually falling in the developed countries.[4] The incidence of meningocele is 5% while mylomeningocele is 90-95% in all spina bifida cases.[5]

The factors predisposing to the spina bifida are both genetic and environmental. In genetic factors it has a polygenically inherited predispision.[6,7] The chance of siblings affected is 3 to 6%, which is 15 times in the general population. The incidence of mylomeningocele in first cousin of patient is about twice that of general population. Risk of mother having the second child affected is about 1 in 25 that is 4% and that after 2 affected children is about 1 in 10(10%)8. Neural tube defects affect females more than males[9]. There is increased incidence of mylomeningocele in certain populations such as Irish, British, Sikhs, and Egyptians. A clear correlation between maternal diet and neural tube defect has been determined, and 7-fold reduction achieved with folate and vitamin supplementation during pregnancy.[10-13] Rates are high for a baby conceived in March, April and May[14] (No specific factors identified). It is more common in the low social class and associated with high intake of potatoes (potatoes hypothesis).[15]

The presentation of these cases is usually with fluid filled sac commonly at the lumbosacral region, but it can be at any site from cervical to sacral region. The patient may have hydrocephalus, lower limb paralysis, incontinence of urine and faeces.[16-19] Multidisciplinary approach is adopted to manage these patients.

This study was conducted to study the short term out come of surgical management of patients with neural tube defect.

MATERIAL AND METHODS

This study was conducted over a period of three years from January 2003 to December 2005 in Paediatric Surgical Unit at Post-Graduate Medical Institute, Lady Reading Hospital, Peshawar. It is an 1100 bedded tertiary care hospital situated in the heart of the city receiving patients from all over the province and also from Afghanistan. It has all the main units along with radiological and pathological facilities. Paediatric Surgical Unit is a 31-bedded unit having two Paediatric Medical Units for medical assistance. We have O.P.D twice a week and we are receiving emergencies round the clock. Most of the neonates with spina bifida present as an emergency.

Patients with neural tube defect in whom surgery was indicated were included in this study except those who had complete paraplegia, total incontinence of urine and faeces and associated life threatening major congenital malformation. The preliminary Investigations, such as Hb%, TLC, DLC, blood sugar, HBS Ag, HCV, X-Ray spine and U/S skull are carried out. Wherever applicable the patients are started on Sofratulle dressing on the lesion, I/V antibiotics, vitamin K supplementation and where surgery was anticipated blood was also arranged. We counseled the parents about the prognosis of surgical and non-surgical management.

On all these patients undergoing the operation the following principals were followed.

- Elliptical skin incision given at the base of the sac
- Freeing the neural placode
- Placement of neural placode back into the spinal canal
- Reconstitution of the neural tube
- Identification and dissection of the dural edges
- Water tight closing of the dural sac
- Closer of the lumber fascia over the dural sac
- Skin closure may be
- Simple skin closure
- Relaxing flank incision
- S-Shape rotational flaps

After discharge from the hospital, the patients were followed serially in the Out Patient Department up for 2.5 years and also advised regarding post-operative and post-discharge management. In the follow up we usually look for the following complications:

1. Paralysis of lower limbs
2. Incontinence of urine and faeces
3. Hydrocephalus
4. CSF leakage from the wound
5. Wound infection

RESULTS

Surgery was performed in 198 cases from January 2003 to December 2005. There were 93 (46.97%) boys and 105 (53.03%) girls. Age of the patients ranged form one day to 18 months. Neonates were 135, infants were 45 and 18 cases were above the age of 12 months (Table 1). There

AGE DISTRIBUTION

Age	Frequency (n=198)	Percentage
Neonates	135	68.18
1 month to 1 year	45	22.73
More than 1 year	18	9.09

Table 1

were 39 cases with meningocele, 144 cases with mylomeningocele, and 15 cases were with lipomeningocele (Table 2). Occipital were 3, frontal were 3, cervical were 27, thoracic were 15, lumbar were 9, lumbosacral were 137, sacral were 3 and one was with double lesion, one at thoracic and the other at lumbosacral region (Table 3). Other associated findings were partial paralysis in 43 (21.72%) cases, hydrocephalus in 42 (21.21%) cases and Telepas equino varus in 12 (6.06%) cases. Incontinence of urine and faeces was found in 39 (19.7%) patients.

All these children underwent surgery for their primary and associated malformations as

TYPES OF NEURAL TUBE DEFECT

Type of defect	Frequency (n=198)	Percentage
Meningocele	39	19.70
Myelomeningocele	144	72.73
Lipomeningocele	15	7.5

Table 2

indicated. In 15/43(38.9%) patients paralysis increased and in rest of 28 cases there was some improvement in the paresis. In 42 cases hydrocephalus developed. Surgery for hydrocephalus was done in 42 cases, in which V.P Shunt was inserted in 12 (28.6%) patients before myelomeningocele repair and in 30 (71.4%) cases V.P. Shunt was inserted after myelomeningocele repair. In 3 cases shunt was put in after complication (CSF leakage from the wound). The number of patients with telepes equinus varus was 12 on which surgery was performed at the age of six months with partial improvement.

Post-repair complications were observed in 24 (12.12%) patients, which were wound infection in 18 cases (9.1%), wound dehiscence in 3 cases (1.52%) and CSF leakage in 3 cases {1.52%} (Table 4). Wound infection was treated by local/systemic antibiotics and frequent dressings. CSF leakage which was treated by socked pyodine gauze and V.P Shunt put in three cases because of persistent CSF leakage.

During the follow up, 34 (17.2%) patients died and 27 patients were lost to follow-up.

DISCUSSION

Mylomeningocele is such an obvious malformation and the complications associated with it are so noticeable that the condition must have been known since the earliest days of history.[20,21] Hypocrites was clearly aware of it. Morgagni 1550-1624 of Basle was the first to give an accurate description of the condition.[22] The great Dutch anatomist introduced the term spina bifida.[23] Nicolaus Tulp, who also appears to have

TYPES OF NEURAL TUBE DEFECT

Site involved	Frequency (n=198)	Percentage
Occipital	03	1.52
Frontal	03	1.52
Cervical	27	13.64
Thoracic	15	7.58
Lumbar	09	4.55
Lumbosacral	137	69.19
Sacral	03	1.52
Double lesion (Thoracic + Lumbosacral)	01	0.51

Table 3

been the first to try to cure the malformation by operation, his patient died and the postmortem dissection was beautifully illustrated in Tulp's book. It is believed that the illustrator was Tulp's friend Rembrandt.[24]

In the 17th Century both Tulp and later on Ruysch realized that the time-honored treatment of aspirating Myelomeningocele was dangerous and they attempted excisions with fatal results.[25] 1n 1811 Astley Cooper published his treatment by

POST OPERATIVE COMPLICATIONS

Complications	Frequency (n=198)	Percentage
Wound infection	18	9.09
Wound dehiscence	03	1.52
CSF leakage	03	1.52
Mortality	34	17.17

Table 4

repeated aspirations. The results of excision as well as aspiration of myelomeningoceles remained poor.[26] In 1872 Morton injected the sac with a sclerosing solution, a method which inspite of bad results, was recommended by a committee set up by the clinical society of London in 1882.[27] It was only when Bayer in 1892 introduced an operation in which a musculofascial flap was fashioned to cover the defect left by excising the myelomeningocele or encephelocele and that excision became the method of choice.[28-30]

Management of spina bifida depends on many factors. Before planning surgery patients are thoroughly evaluated for sac (site, skin cover, leakage of CSF), gross neurological deficit, other associated anomalies, socioeconomic status and acceptability of the baby by the family.[31]

In our study 151 cases were excluded because surgery was deferred due to:

a. Complete paraplegia, total incontinence of urine and faeces, and marked hydrocephalus.

b. Mortality in such group of patients is up to 60% within the first year of life.

c. Associated life threatening anomalies not compatible with life.

d. Rehabilitation of such patients in our community is very difficult.

e. Most of the parents belong to low socioeconomic class.

f. Lack of tertiary care health facilities.

g. State can not afford all expenses.

In those patients where surgery was indicated, the following protocol was followed in the unit. Those patients with CSF leakage were operated within 10 to 12 hours after antibiotic cover. Patients with exposed neural plaque were operated within 12 to 24 hours after birth. Sac covered with very thin skin were operated as early

as possible because of danger of rupture. Full thickness skin cover myelomeningocele were operated after neonatal age. Myelomeningocele with paralysed limbs if alive, were operated at the age of 8 months for nursing purposes. The aim of management was to have an ambulant child with no incontinence of urine and feaces, who is able to live independently and intelligently.

In our study Myelomeningocele was the common most (72.73%) neural tube defect which is consistent with other studies where Myelomeningocele (MMC) was present in 86.8% cases.[32]

In our study hydrocephalus was found in (21.21%) 42 cases. V.P Shunt was inserted in 12 (28.6%) patients before myelomeningocele repair and in 30 (71.4%) cases VP Shunt was inserted after myelomeningocele repair. This is contrary to the findings of Kumar R[33] et al who had hydrocephalus in 58.8% of children and who reported that 63.3% of children with overt hydrocephalus required shunt surgery prior to the definitive surgery; however, 23.3% of cases required a shunt after the MMC has been closed.

In our study partial paralysis was noticed in 21.72% cases, which is less than the 60 % reported by Saleem M et al.[34] This could be due to the fact that patients with complete paralysis were excluded from our study.

Complication rate of 12.12 % is acceptable. The most common complication was wound infection. Central nervous system infection is more common and the leading cause of death (44%) in patients with meningomyelocele.[35] Postoperative complications have been reported to be more frequent in patients with ruptured lesions, a larger size of defect and a lower birth weight.[32]

Mortality in our study was 17.17% which is comparable to 22.7% reported by Alatise OI et al.[33] Long term survival of the cases of open spina bifida is poor and almost half of them died around 30 years of age.[36] Of the survivors, 37% lived independently in the community, 39% drove a car, 30% could walk more than 50 metres and 26% were in open employment. However one-third still needed daily care.[36]

CONCLUSION

Spina bifida (neural tube defect) involves multiple organ systems and its management requires proper assessment and appropriate decision for surgery. Proper selection of patients for surgery has a fair outcome. Although there is no satisfactory way of treating the spina bifida patients, but efforts can be made to prevent the ongoing complications of the disease and multidisciplinary approach is required to manage these patients to improve the quality of life.

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