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PREDICTING OUTCOME FOLLOWING SURGERY FOR MYELOMENINGOCELE: A UPTH EXPERIENCE

P. O. Eghwrudjakpor, S. O. Wokocha, Augusta Eneh*
*Departments of Surgery and *Pediatrics, University of Port Teaching Hospital, Port Harcourt, Nigeria.*

ABSTRACT

Objectives: To evaluate outcome of treatment in children with myelomeningocele, and determine to what extent the clinical features at presentation can be utilized in prediction of this outcome.

Patients and methods: This study included patients with myelomeningocele admitted into the University of Port Harcourt Teaching Hospital over a two year period. All of them were operated upon by the same team using standard techniques.

Results: Neurological impairment was least severe and outcome of surgery was most favorable in those children with small and low-lying defects.

Conclusion: Management of the Neural Tube Defects, especially myelomeningocele, poses special problems. The results of surgery are not always satisfactory; and there is no consensus regarding surgical indications. The outcome following surgery probably depends on an interplay of several factors. Our findings, however, confirm that the most important predictors of severity and perhaps the postoperative outcome are the level and size of the defect.

***KEYWORDS*: Central Nervous System (CNS); Clinical features; Myelomeningocele (MMC); Outcome prediction; Surgery.**

INTRODUCTION

Treatment of children with myelomeningocele has undergone significant revolution over the years. From a more or less conservative attitude in the management of these patients, there now appears to be a trend towards more active intervention. Ethical, social, moral and religious issues, however, remain largely unresolved.

Characterized by herniation of the meninges and myelodysplastic neural tissue through a bifid spine, myelomeningocele is the commonest form of spina bifida. When it is treated early, the outcome in most patients can be expected to be reasonably acceptable because many of them will be able to achieve "community ambulatory status" - i.e. they can move about the community without resorting to wheelchairs[1]. The decision whether or not to treat these patients actively is, however, still the subject of intense controversy, with the result that even among neurosurgeons there is no complete agreement with regards to the indications for operation[2,3,4]. Although the protocols that were used for patient selection in the past[5,6,7] have largely been abandoned in favour of "aggressive management"[1] whereby surgery is offered to most patients without undue emphasis on their preoperative neurological status (as adjudged by the established criteria), predicting their outcome is still a significant problem.

*Correspondence: Dr. P. O. Eghwrudjakpor

In the neurosurgical unit of the University of Port Harcourt Teaching Hospital, our attitude is also one of "aggressive management". In this paper, we report our experience of these cases over a 2 years period and highlighted the relationship between the clinical features and outcome following treatment.

PATIENTS AND METHODS

Between January 1st, 2000 and December 31st, 2001, we operated on a total of 15 children with myelomeningocele. They came from geographical locations in the catchment area of the University of Port Harcourt Teaching Hospital. These include Rivers (8), Imo (4), Delta (1), Bayelsa (1) and Abia (1) states. Children with other neural tube defects including *cranium bifidum*, and other forms of spina bifida (myelocele and meningocele) were not included in this study. In addition to routine questions, relevant information obtained at the time of admission include the ages of the mothers; their pregnancy history including drug history, intrapartum illnesses and irradiation; family history; position of the affected children among their siblings and the presence of perinatal complications.

General physical and neurological examinations were carried out on all patients. In particular we sought after the presence of other congenital malformations outside the CNS. The neurological status at presentation was recorded and compared with postoperative changes. The head circumference was also measured at the same time and the rate of growth thereafter moni-

tored. All information obtained was duly documented in patients' folders. See Table 1.

Investigations requested for include x-rays of the skull, spine, and limbs when indicated. These were, however, not done in most cases due to financial constraints.

All the patients were operated upon by the same surgeons (POE and SW), and the same technique was employed in all cases. This involved: separation of the neural tissues from the superficial layers, their return into the spinal canal, canal reconstruction with duraplasty, approximation of paravertebral muscle fascia, where possible, and wound closure.

RESULTS

There was a total of 15 children; all of whom were confirmed to be full term deliveries. Of these, 9 were males and 6 were females. Their ages at presentation ranged from 1 day to 15 months (median of 5 days).

Position among siblings: We found no relationship between the position of patients among their siblings and the occurrence of MMC. 4 of the children were first, 3 were second, 3 were third and 2 were fourth among their siblings.

Antenatal history: All but one mother received proper antenatal care, either in a hospital or maternity home. Only 3 women did not take their routine antenatal drugs consistently. Besides malaria, none of the women agreed to having had significant illnesses during pregnancy.

Ages of the women who presented children with myelomeningocele ranged from 19 to 38 years (with a mean age of 28.8 years). See Fig 1.

Location of the defect: The lumbosacral region was the area most commonly involved (60%). In 2 children, the neck of the defect was quite broad and involved the thoracic, lumbar and sacral regions simultaneously.

Neurological status: Motor function was completely normal in 3 children. Among these, two presented with sacral myelomeningocele while the remaining one had a small lower lumbar defect. All the other children presented with some degree of neurological deficits – ranging from mild to severe, often asymmetrical paraparesis and/or incontinence of urine and faeces. Motor weakness was most severe in the children with thoracolumbosacral lesions. None of the children we operated on was completely paraplegic. See Table 1.

Associated congenital deformities outside the CNS: *Talipes equinovarus* was the most commonly encountered, being recorded in 40% of patients.

Radiological evaluation could be performed only in six children. Findings of spinal x-rays were consistent with rachischisis. 2 children also had kyphoscoliosis. In one of these, (A.M.) the skull x-ray also showed Lacunar Skull Deformity (LSD).

Fig.1: **Relationship between Maternal Age and Incidence of MMC**

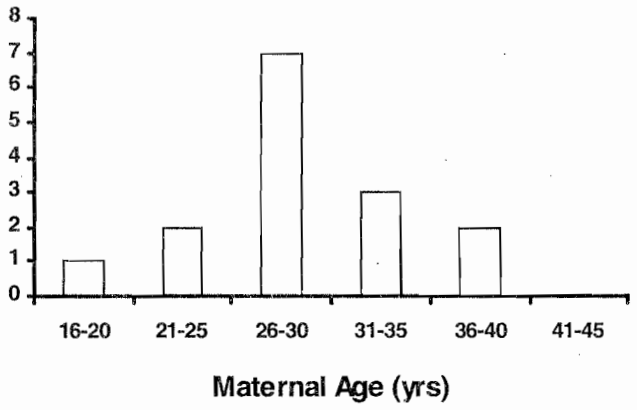

Table 1: **List of Myelomeningocele patients between 01/01/2000 and 31/12/2001**

No.	Name	Sex	Age	*H.C. (Cm)	Location of Defect	Size of Defect	**P.A.S.	***M.A. (yrs)	Motor Function	Antenatal History	Drug History	Associated Anomalies	Complications
1.	I.C.	M	1day	34.0	Thoraco-lumbosacral	9.0cm	3rd	29	Paraparesis	Booked	Routine	Talipes	CSF leakage
2.	E.K.	F	5days	36.5	Lumbar	3.0cm	2nd	27	Paraparesis	Booked	Routine	-	-
3.	O.N.	M	10days	33.0	Lumbosacral	3.5cm	8th	31	Paraparesis	Booked	Routine	-	Hydrocephalus
4.	S.B.	M	1day	33.0	Sacral	2.5cm	2nd	38	Normal	Booked	Routine	-	-
5.	A.O.	M	1day	35.0	Sacral	2.0cm	5th	27	Normal	Booked	Nil	Talipes	-
6.	A.M.	M	1day	34.0	Thoraco-lumbosacral	8.0cm	1st	27	Paraparesis	Unbooked	Nil	Kyphoscoliosis	CSF leakage
7.	A.U.	F	10days	35.0	Lumbosacral	5.0cm	4th	19	Paraparesis	Booked	Routine	Talipes	Sepsis
8.	O.G.	F	5days	33.5	Lumbosacral	4.0cm	3rd	38	Paraparesis	Booked	Routine	Talipes	-
9.	J.D.	F	15mns	39.0	Lumbosacral	3.5cm	1st	29	Paraparesis	Booked	Nil	Talipes	-
10.	O.A.	F	7mns	38.0	Lumbosacral	3.0cm	6th	26	Paraparesis	Booked	Routine	-	Hydrocephalus
11.	M.S.	M	1day	38.7	Lumbar	2.0cm	2nd	22	Normal	Booked	Routine	-	-
12.	U.F.	F	3wks	34.0	Lumbosacral	3.0cm	1st	29	Paraparesis	Booked	Routine	Kyphoscoliosis	Meningitis
13.	O.J.	M	7days	32.5	Lumbosacral	4.5cm	4th	32	Paraparesis	Booked	Routine	-	CSF leakage
14.	K.F.	M	4days	36.5	Lumbosacral	4.0cm	1st	25	Paraparesis	Booked	Routine	Talipes	Hydrcephalus
15.	O.B.	M	1day	37.0	Lumbosacral	3.5cm	3rd	33	Paraparesis	Booked	Routine	-	-

***H.C.** = **Head Circumference**
****P.A.S** = **Position Among Siblings**
*****M.A.** = **Mother's Age (Years)**

Post-operative complications include cerebrospinal fluid leakage, meningitis and hydrocephalus. CSF leakage and meningitis, when they occurred, were successfully managed conservatively. Of 3 children with hydrocephalus, 1 resolved spontaneously, and 1 had to have shunt insertion. The remaining one was lost to follow-up.

Clinical course/Follow up: The clinical course varied with the individual cases. Only 3 children could be followed up for more than 1 year. Average duration of hospital stay after surgery was approximately 10 days. This was slightly longer in children with cerebrospinal fluid leakage or meningitis, and shorter for uncomplicated cases. No new neurological deficits were observed in any child in the immediate post operative period or during follow up as outpatient. Of 3 children who were neurologically intact preoperatively, only 1 (S.B.) could be followed up for more than 8 months. His developmental milestones and rate of head growth remained normal. The child with the Lacunar Skull Deformity on the other hand, at the time of his last visit to clinic 11 months after surgery, was no longer incontinent of urine or faeces; but still had marked motor weakness of the lower limbs. He could barely sit without support, but was otherwise healthy.

DISCUSSION

Myelomeningocele is one of the commonly encountered congenital anomalies of the Central Nervous System (CNS). It comprises more than 70% of spina bifida cystica[8,9]. Unlike many other congenital disorders of the CNS, it is amenable to surgical intervention. Its occurrence has been associated with a number of factors including extremes of maternal age (especially mothers below 17 years and above 40 years of age). This was, however, not the case in this study. In fact, only one mother was less than 20 years and none of them was more than 40 years of age. Most of them indeed belonged to the twenty-six to thirty year age group (generally regarded as optimal for childbearing). Our findings are also at variance with some reports in the literature that suggest that the third child is more susceptible to developing spina bifida cystica[9]. In this study the disorder tended to be almost evenly spread across the various ages (Table 1), with a slight male preponderance (M:F ratio of 1.5 to 1.0).

Presentation to hospital was fairly early, with majority of them presenting during the first week of birth. This we believe is partly due to the increasing awareness that many of these children can be assisted to live reasonably acceptable lives after surgery. Also, in the joy of a new birth, many parents are not quite comfortable showing off a baby with a deformity. Indeed, the only child in the series who presented late (at 15 months) had actually been seen at another institution earlier, but due mostly to financial problems coupled with other logistic difficulties, the parents had to resort to unorthodox methods of treatment before coming to hospital as a final resort.

The lumbosacral region was found to be most commonly involved in this study, and all affected children presented with various degrees of neurological deficits. The paucity of neurological symptoms seen in the children with sacral and lumbar defects is almost certainly due to the fact that there was comparatively little neural involvement in these defects; and even then, the level was so low that motor and sphincteric actions were spared.

The finding of Lacunar Skull Deformity (LSD) in skull x-rays of children with myelomeningocele during the neonatal period has been associated with the occurrence of psychomotor retardation[10] as was the case in the only patient with this finding (A.M.) in the current series. However, it must be noted that in addition to LSD, this same patient also had a broad-based defect as well as severe orthopaedic complications all of which have equally been cited as negative prognostic features[5,7,8].

CONCLUSION

Outcome following treatment of children with myelomeningocele depends on several factors. The number of patients in this series is still quite small and follow-up is not long enough. However, from these preliminary results, it would appear that the most important determinants of the severity of neurological impairment, and therefore the pointers to possible outcome of surgery are the level and size of the defect. Clearly, a much larger sample is required to make the results more meaningful.

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