M Y E L O M E N I N G O C E L E :

A review of seventy one (71) cases and the difficulties encountered in the total management of these patients in the five year period, 1975 through 1979.

By Dr. Athanas Wambati Maingi
M.B.Ch.B.

A Thesis submitted in part fulfilment for the degree of MASTER OF MEDICINE (Surgery) of the University of Nairobi.

D E C L A R A T I O N S:

C A N D I D A T E

This thesis is my original work and has not been presented for a degree in any other University.

Sign _____

DR. A.W. MAINGI M.B,Ch.B.

S U P E R V I S O R :

This thesis has been submitted for examination with my approval as University Supervisor.

Sign _____

PROFESSOR NELSON W. AMORI F.R.C.S.

A C K N O W L E D G E M E N T S :

1. I, would like to thank Professor Nelson Awori for the great inspiring help which he gave me throughout the study of this work.

2. My thanks also are extended to Dr. J. Dar Senior Lecturer and Consultant Neurosurgeon in-charge of the Neuro-surgical unit, for the great help he gave me during the compiling of the work of this study, and also for the guidance and timely constructive and helpful comments. Many thanks are also due to other Neuro-surgeons of the unit, namely; Dr. Sande and Dr. Michael Kahwa, for the advice in the whole field of spina bifida.

3. I thank also the Sisters in-charge of the paediatric wards 1 and 2 for allowing me access to patients at all times. Special thanks in this respect go to Sister Wikonyo, the Sister in-charge of the Rehmutulla Wing ground floor.

4. Very special thanks to Mrs. Susan Ikui, the Records Officer who played a tremedous role in allowing me free access to the notes of all the patients included in the study.

5. Finally, but not least, to my beautiful wife, for the beautiful secreterial work.

C O N T E N T S :

S U M M A R Y:

During the period between January 1975 and December, 1979, one hundred and thirty two (132) new patients were seen in the neurosurgical clinic with some form of malformation of the cranium or spine. One hundred and eight (108) of them were spina bifida cystica and seventy one (71) of these had myelomeningocele of whom only thirty-seven (37) were selected for active treatment. Multidisciplinary approach for management of this condition was lacking, and the follow up was inadequate.

I N T R O D U C T I O N :

The aim of this paper is to analyse the spectrum of spina bifida problems, its management and follow up in the neurosurgical unit of the Kenyatta National Hospital. It is a retrospective study, in the greater part of it.

Spina bifida presents a tremedous initial shock to the parent looking at the deformity, and a well sustained fear of the future of the child. To the medical personnel confronted with the treatment of these children it is still a dilemma. The urge to guard many of these children from expected physical suffering and later on the mental anguish of being a cripple is great. A large number of doctors thus are not in favour of doing anything active on them. Large numbers of children alive following early closure of the defect and efficient treatment of hydrocephalus reach adolescence painfully aware of their deficiencies. Presence of such children cannot be considered a tribute to medical achievement, but can be interpreted as misuse of medical powers. Because of such a wide variation in thinking at different centres, the criteria of selection of children for treatment is still being sought.

HISTORICAL BACKGROUND:

Spina bifida has been shown to exist for more than 12,000 years (Ferembach, 1963). Hippocrates was aware of the condition. Tulpius, the teacher in Rembradts "Anatomy Lesson" described and illustrated it in 1652, and called it spina bifida. Von Ruysch in 1714 distiguished

...../cont.

INTRODUCTION CONT.

between paralytic and non-paralytic forms. Morgagni in 1761 first associated it with hydrocephalus and almost a century later, Cleland (1883) first showed and described what today is called Arnold-Chiari malformation. Arnold (1891) and later Chiari (1895) described what today is called a Lucuna Skull.

The entity and its associated abnormalities were clearly described and documented almost two hundred years ago. Yet inspite of the knowledge and advancement in medical technology, this condition still presents enormous problems in its management.

E M B R Y O G E N E S I S

The first attempt on the study of morbid anatomy and development of spina bifida were carried out in the nineteenth century by Professor Cleland of Glasgow and later by Von Recklinghausen. These studies because of the diversity of views on the development of this condition made it very controversial. Morgagni postulated that the primary defect was due to the obstruction of the central canal causing swelling and consequently rupture of the spinal cord, while Becland although accepting this 'hydrops' theory to a certain extend, attributed the development to torsion of the umbilical cord and persistance of excessive fluid in the central nervous system. Lanesti (1877) postulated the defects to be due to adhesions of the amnion to the back of the embryo, while Lebedoff (1881) thought the condition to be due to angulation of the spinal axis and consequent prevention of closure of the open neural tube. Later both Cleland and Von Recklinghausen changed their views. Cleland then suggested that the defect was due to overgrowth of neuroectoderm, while Von Recklinghausen advocated abnormal notochordal growth. Both however agreed that the condition resulted from failure of closure of neural tube due to some disturbance of the longitudinal growth of the embryo. This view is widely accepted at the present time. To this list has been added, quite recently, the hydromyelic theory by Gardner (1961). This theory was put forward after Gardner had observed a neighbour's child playing with a baloon whose surface had "popped up" at some weaker areas. This theory has been used to explain

..../4

the other congenital abnormalities of the central axis like the Dandi-walker cyst, Arnold Chiari malformation and syringomyelia. The embryology of the normal spinal cord is briefly outlined. The spinal cord develops from the neuro-ectoderm. Induction of formation of the neural tube depends on the influence of the underlying notochord. Three stages of development are recognised:-

1ST STAGE

NEURULATION : which takes place at around the 18th day of gestation and is completed by the 26th day, when the caudal neuropore closes. In other words differentration of the cells, formation of the neural tube and its folding, and then its closure in zipper fashion from the cervical region towards both ends (cranial and caudal) is complete between 18th and 26th days after conception. Failure of closure of caudal neuropore results in open myelomeningocele.

2ND STAGE

CANALIZATION OF CAUDAL CELL MASS (26 - 30 DAYS) :

This is the process which accounts for the development of the spinal cord below the level of 1st lumber vertebra. This caudal cell mass or tail bud, by its differentiation, also gives rise to other important structures like the kidneys. Vacuoles appear here, coelesce to form a canal which is continous with the more proximal one, and then the cell mass undergoes atrophy so that finally only the filum terminale can be seen - failure of this process may result in closed myelomeningocele or occult spinal dysraphism.

..../5

3RD STAGE:

The final stage is the development of the spinal cord coverings, and later the closure of the posterior vertebral arches at 40 - 60 days. Failure may result in spina bifida occulta or a simple meningocele.

HISTORICAL REVIEW OF MANAGEMENT:

In reviewing the literature, five phases are seen in the management of this condition. The simple meningocele will not be discussed here because its presentation, treatment and results still remain the same and devoid of any contraversies. As expected, it is the myelomeningocele of various extent which presents all the problems of spina bifida as we know today.

1ST PHASE:

No definite treatment was given for myelomeningocele until late 19th century, and therefore most of the children died during early infancy. This was particularly so because of lack of antibacterial agents and unavailability of proper surgical techniques.

2ND PHASE:

There was a brief period when some form of treatment was given. Morton (1877) in Glasgow treated these babies by injecting into the sac a solution of iodine in glycerine. Mayo Robson (1885) attempted surgical excision, but these techniques were unacceptable and were soon abandoned. Nothing further happened till the middle of 20th century when Sir John Fraser (1929) accepted that the results obtained were not worthy the effort.

..../6

3RD PHASE:

This phase lasted some twenty years upto 1958. During this time treatment was initially based on observation of the cases and then treating the baby who seemed to survive beyond the 1st year of life. By this time, backclosure was good, but a number of those whosurvived died of hydrocephalus which could not be controlled. In England, the attitude was aggressive in the 1950s,(Zachary in Sheffield, MaCnab in London., Naish (1956) first carried out the procedure of ileocutaneous ureterostomy for urine incontinence in London. The results of those who used selection (MaCnab in the London series) were better than the aggressive series of Zachary in Sheffield. The results were generally disappointing in both the series particularly survivors from Sheffield where the degree of mental and physical handicap in the survivors was unacceptable.

4TH PHASE:

This phase was born with the discovery of the Shunting system using a valve. This was developed by Holter an American Engineer and was first used by Spitz a surgeon on Holter's son; with very good results. Thus with this development all the spina bifida children were being referred to cetres for treatment on a treat-all basis. The news spread so much so that by the 1960s it was very difficulty for a doctor not to refer a child and for a surgeon not to operate in fear of adverse critism. Only three centres did not follow this routine. Oxford (Hide, 1972) Edinburgh (Stark and Drummond, 1973) and Melbourne (Smith and Smith, 1973).

...../ 7

After sometime the Sheffield series showed that many were dead within a year and those remaining were steadly dying. The survivors had severe mental and physical handicap, and others required numerous shunt revision procedures due to shurt complications. Thus the lesson of the 4th phase had been learned, and especially from the community point of view whereby such children were draining enormous national resources in education and upkeep.

5TH PHASE:

With the poor results of treatment so far, a principle of selection was in the mind of doctors. Lorber (1971) developed in Sheffield a criteria for selection. This criteria was generally accepted in many centres and has continued to be practised with very little modification the world over. Some centres nevertheless continued to criticise it for many years on moral, ethical or philosophical reasons.

...../8

METHODS:

A review of all the children with mid-line congenital anomalies of skull and spine referred to the Neurosurgical unit of the Kenyatta National Hospital (K.N.H), during the period from January, 1975 to December, 1979 was done. K.N.H. is the only centre in the country where free Neurosurgical care is given by full time personnel provided by the Medical School of the University of Nairobi. The patients, therefore, come from all districts and a number of them from neighbouring countries as well.

During the first visit, the child was examined thoroughly from the neurological point of view, and thorough general examination done. The initial examination was usually carried out by one of the Neurosurgeons, diagnosis of spina bifida cystica confirmed and any deformity of spine was noted. The tone, power, reflexes, sensation bladder or bowel incontinence presence or absence of foot deformity and head circumference noted. A general examination of the chest and abdomen was carried out and any disease present indentified and other congenital malformations excluded. Decision to treat or not to treat was then usually made on the basis of the findings.

At each subsequent visit during follow up, particular attention is paid to bladder or bowel incontinence, the presence of hydrocephalus by measuring the maximal occipital-frontal circumference of the head, and finally any new deformity of the limbs or worsening of the pre-existing one if present. This format of examination is routine and is usually carried out by one of the registrars attached to the unit. If there is anything significant found at follow up for

example appearance of new sign or symptom the Neurosurgeon is informed forthwith and usually re-examines the child, and gives advise on any further management.

One of the parents, usually the mother almost always accompanys the child throughout the follow up period. Sometimes advise would be given that the baby would be referred to his home district hospital for further follow up without having had to return to the unit. This was usually so when a decision of not to treat was made either at first visit or during follow up when results are not good warranting any further active treatment.

..../10

RESULTS:

During this period, a total of 132 babies and children were seen in the unit, having been referred with mid-line congenital anomalies. Of these 108 (82%) had spina bifida cystica.

TABLE I THE TYPE OF PRESENTATION OF SPINAL AND CRANIAL MALFORMATION:

TYPE	NO. OF CASES	PERCENT
SPINA BIFIDA CYSTICA	108	82
CRANIAL ENCEPHALOCELE	14	11
ANTERIOR MENINGOCELE	2	2
DIASTEMATOMYELIA	8	6
NO. OF PATIENTS	132	

One of the anterior meningocele was in a six month old child who had presented with persistent cough for which a plain chest Xray was ordered revealing a mediastinal mass. The other anterior sacral meningocele presenting as a pelvic mass.

The diastematomyelia patients presented much later than the others most of them in mid or late childhood. A couple of the patients presented in their adolescence. Three patients with diastematomyelia had stigmata of spina bifida on clinical examination, while the rest presented with progressive neurological deficit.

..../11

Lumbar and lumbo-sacral myelomeningocele account for 82% of all the meningoceles lumbar being the commonest site (47%).

TABLE 2 DISTRIBUTION OF THE LEVEL OF LESION IN 71 CASES WITH MYELOMENINGOCELE.

LEVEL OF LESION	NO. OF CASES	PERCENT
CERVICAL	3	4
THORACIC	2	3
THORACO-LUMBAR	3	4
LUMBAR	33	47
LUMBO-SACRAL	25	35
SACRAL	9	13
TOTAL NO. OF PATIENTS	71	

These 71 patients had a total of seventy-five lesions. Three patients had more than one lesion. One baby had a cervical and sacral lesion, while two babies each had a cervical and lumbar lesion. These three babies had severe neurological deficit and were not selected for treatment. More than two lesions were not seen in any one patient. Three meningoceles in the same patient are exceedingly rare; one such case was reported by Tryfonas (1973).

...../12

As expected some of the babies seen earliest were those born in the maternity wing of the Kenyatta National Hospital. The others were those born in the neighbouring Pumwani Maternity Hospital which is the main maternity hospital of the city's Municipality. Babies were seen from all corners of the country with degrees of delay sometimes not corresponding with the distances.

Only nine (13 %) of the patients with myelomeningocele had a reasonably comprehensive case summary as far as neurological examination is concerned. None had any orthopaedic examination as apparent from the case summary. The rest of the patients had in the letter of referal or internal consultation form (used in K.N.H), very sketchy notes such as "querry meningocele" or just "spina bifida for management". The majority had no information or examination of other systems, let alone family history or perinatal or post-natal sequency of events. This was true even of babies referred from the maternity unit of K.N.H.

...../13

Table 3 shows how soon after birth the babies could be seen in the neurosurgical unit.

TABLE 3 : TIME ELAPSED BEFORE CHILD HAD NEURO-SURGICAL ATTENTION.

TIME ELAPSED	NO. OF CASES	PERCENT
UNDER 2 DAYS	8	11
2 - 10 DAYS	22	31
10 - 30 DAYS	15	21
30 - 90 DAYS	10	14
OVER 90 DAYS	16	23
TOTAL NO. OF PATIENTS	71	

Of the 8 patients seen under the second day of life, 5 were selected for treatment and infact these babies were seen on their first day of life. The decision to treat these babies was made by the Neurosurgeons often without much participation in the decision making from the paediatrician or orthpaedic surgeon.

Thirty seven 37 (52%) babies were selected for treatment. The rest of the babies were not selected for treatment because of severe neurological deficit with incontinence of urine or stool or both, or because they had severe hydrocephalus or some othe lesion which was considered a contraindication to active treatment (Lorber, 1973).

.../14

It has been stated above that thirty four 34 (48%) out of the seventy one (71) myelomeningocele patients were not selected for treatment. Table 4 tries to show the reason where such a decision was made.

TABLE 4 : REASONS FOR NOT SELECTING 34 PATIENTS FOR TREATMENT.

REASON	NO. OF CASES	PERCENT
SEVERE PARALYSIS	30	88
SEVERE HYDROCEPHALUS	21	61
INCONTINENCE OF URINE	10	29
INCONTINENCE OF STOOL	9	26
SPINAL DEFORMITY	3	9
OTHER	3	9
TOTAL NO. OF PATIENTS	34	

Two of these patients had prolapsed uterus and five had bilateral talipes equinovarus in addition to one or more of the above in combination. There were three patients rejected under the "other" category. One week old baby from a district hospital, had severe vomiting from birth and was in very poor condition. This was thought to be due to malformation of the upper gastrointestinal tract. The baby also presented with pyrexia and moderately

...../15

severe brochopneumonia with persistent cough when being fed. This suggested a treacheo-oesophaged fistula. Before the diagnosis of multiple congenital malformation could be confirmed, the baby succumbed and died. The other two babies under "other" were 'rejected' because the parents refused any form of operation being undertaken on their babies, despite thorough explanation from the doctor and ward sister.

TIMING OF SURGERY:

The timing of surgery was more dependant on the time the child was seen first rather than in a definate policy. Even those who were seen on their first day of life, and selected for treatment, often could not get this treatment during their first two days of life. This was due mainly to shortage of operating time in theatre, or the unavailability of beds in the unit or that there was no consent for operation because in the African set-up, it is usually the father who must give consent for any operation. These babies were usually accompanied by their mothers or sometimes even by an escorting nurse. The ones operated on, were usually three days old and often much older. Table 5 illustrates this.

..../16

TABLE 5 : TIMING OF TREATMENT OF 37 CASES.

TIME ELAPSED BEFORE TREATMENT	NO. OF CASES	PERCENT
UNDER 2 DAYS	0	0
2 - 10 DAYS	10	27
10 - 30 DAYS	12	32
30 - 90 DAYS	8	22
OVER - 90 DAYS	7	19
TOTAL NO. OF PATIENTS	37	100

The range of delay was between 3 days and fourteen months, but the majority (59%) were treated under one month of age.

It cannot be said that all the thirty seven (37) patients selected for treatment were perfectly free of any problems. Eight (22%) of them had some varying degrees of disability as seen in figure 1 below:-

FIGURE 1 : MINOR DISABILITY IN 8 PATIENTS SELECTED FOR TREATMENT.

Paralysis of one leg 2

Paradoxical bladder (and or bowell) 4

Foot deformity 6

Hydrocephalus 4

Kyphosis/scoliosis 1

...../17

All these patients were treated under one week of life and no worsening of the original condition was observed while the patients were recovering in the ward. Two patients had ventriculoatrial shut inserted at the same sitting as back closure, but one developed respiratory problems and died hours post operatively. The other one was discharged without any problems. Two patients were treated conservatively with diamox and were for follow-up. One was lost for follow-up after only three months, and when she finally turned up after eight months was found to have severe hydrocephalus and completely blind. No. further surgical intervention was considered advisable.

Fifteen patients (41%) selected for treatment and considered to be perfect except for the back lesion, developed varying degrees of disability post-operatively. This is illustrated in figure 2. The operation timing was between five days and thirty three days. So these patients were really not treated as early as ideal.

FIGURE 2 : NEUROLOGICAL POST OPERATIVE COMPLICATIONS IN 15 PATIENTS PREVIOUSLY WITHOUT ANY DEFICIT.

Complete paralysis (Flaccid/or spastic)	.....	6
Bladder/bowel disfunction	.....	15
Foot deformity	.....	3
Hydrocephalus	.....	6

Some of them had more than one type of complication.

...../18

FOLLOW UP:

It became later clear that the follow up of these myelomeningocele patients was not consistent and that many of them were lost for follow up. Only 22 (31%) out of the seventy one patients were attending the neurosurgical clinic till their 1st birthday, and of these sixteen were the treated group. The rest had either died or that their parents sought traditional medicineman's care or they were not selected for treatment and therefore send back to the referring hospital with all the advise to the doctors and the parents having been outlined. The majority had been lost at 2 years. The longest follow up was four years and this child had been referred to the orthopaedic surgeons for management of the limb deformaties and for advise on braces.

...../19

D I S C U S S I O N :

It is clearly seen that myelomeningocele is still the more common type of spina bifida cystica in Kenya, as in other places. Knowing the natural history of this condition its management presents perhaps the greatest challenge to the doctor involved in their management.

Some degree of morbidity is always associated with myelomeningocele no matter what type of management or when such management, is given. This is particularly so in the developing countries like Kenya where other facilities such as deficiency in numbers and quality of staff, hospital beds and difficulties in communication are the order of the day. Like in any other centre, the first and the most difficult point in the approach to treatment of these babies is the decision; 'to treat or not to treat' and when.

Doran and Guthkelch (1961) having tried the new weapon of Spitz-Holter valve and the ventriculo-artial shunt in a large series, concluded their assessment of the whole problem of spina bifida with the words, "In the light of these findings, it can be argued that the most important remaining limit to the number of infants born with spina bifida cystica of all grades of severity who can lead worthwhile, even if restricted life is the amount of medical and surgical care which can be expended on them".

The first thing in this study is the delay in treatment which was due to difficulties in communication as most of these babies were referred from district and provincial hospitals. The delay was also caused by the doctors who had contact with the baby at the first hour of life, as they did not appreciate the erroneous problems in management of these cases. This is shown by the fact that the great

.../20

majority of these babies did not have adequate case notes i.e. thorough general examination, and adequate neurological or orthopaedic assessment, let alone measurement of the head circumference.

Delay at the unit is also evident and this was due to lack of hospital beds or operating time. When considering the surgical approach to management, timing is of a definite prognostic importance. Sharrard (1963) and his colleagues in Sheffield reported a controlled trial of early (under 48 hours) and delayed closure in infants with open myelomeningocele. They found that when closure was delayed, not only were infection and death more likely but paralysis tended to increase rapidly after birth. This study had a profound influence throughout the world, in the management of open meningomyelocele and the emphasis has been on early treatment of these cases.

The unit selected the cases on the guidelines outlined by Lorber (1971). On such selection criterion 37 (52%) were selected for treatment. The pitfall of the selection is that a multidisciplinary approach is not seen at the start in this hospital. In other words these babies do not routinely have a thorough paediatric, neurologic and orthopaedic assessment before the decision to treat or not to treat is made.

Practical experience in selective early operation is still limited and reports on its effectiveness emanate from centres such as Edinburgh and Melbourne in which the "Sheffield policy" or routine early closure was never accepted.

This policy was practised not only in Sheffield but also in many major centres in the 1960s. It was not until 1971 however, that the first fully documented follow up study appeared in Sheffield. In it, Lorber (1971) spelled out an appalling catalogue of suffering, disability and death. Only 41% had survived 7 years or more. Of the survivors(49%) were mainly or entirely wheelchair bound, and many of them had undergone numerous operation (average more than 12 operations). The majority had severe urinary tract and a third of the survivors had an I.Q of less than 80. Later series had lower mortality rate, but the disability of the survivors had increased.

The results of the selective early operations has been shown to have much superior results. In the Edinburgh series, (98%) of the babies were admitted within twenty-four hours of birth. On the basis of detailed paediatric, neurologic and orthopaedic assessment, (48%) were selected for early back closure. At the end of three months, more than (80%) of untreated infants had died, whereas more than (70%) of selected infants were alive, at 6 years. 25% of the survivors were mainly or entirely chairbound, 16% had upper urinary tract damaged and only 20% had a combination of severe mental and physical handicap. These figures are bad enough by any standards, but it will be seen that they are at least an improvement upon the figures given for Sheffield by Lorber (1971). Results from Melbourne (Smith and Smith 1973) are similar to those of Edinburgh.

..../22

Table 6 shows the comparison of results of routine early and selective early treatment.

TABLE 6 : RESULTS OF ROUTINE EARLY AND SELECTIVE EARLY OPERATION.

AUTHOR	NO. OF PATIENTS	% OF EARLY OPERATION	OVERALL 6–7 YEAR SURVIVAL	CATEGORY OF DISABILITY AT FOLLOW-UP 1 2 3 4 5
LORBER 1971	323	100	60	3 15 49 21 12
HUNT et al 1973	113	100	?	0 6 40 39 15
STARK & DRUMMOND 1973	163	48	42	0 25 56 13 6
SMITH & SMITH 1973	159	60–70	56	0 48 30 20 2

Categories of disability are based on those defined by Lorber (1971) and Stark Drummond, (1973).

Cat. 1 : no handicap

Cat. 2 : moderate handicap (normal intelligence and walking without calipers)

cat. 3 : severe physical handicap (walking with calipers or in wheelchair) - normal intelligence.

cat. 4 : severe physical handicap + moderate mental retardation (I.Q 60–75).

cat. 5 : severe physical handicap + severe retardation (I.Q 60).

...../23

The points of Lorber's criteria for selection are clear and easy to understand. It must be remembered that it is always not easy to examine a neonate even by the most experienced paediatric neurologist, but, the criteria are easy and do not require very sophisticated tools to carry out the examination. The points of the criteria are taken to understand that if any one of the points is found, it will amount to contradiction to treatment.

The criteria goes as follows:-

1. AT BIRTH:
   (a) Gross paralysis of the legs (paralysis of the legs below 3rd lumbar segmental level with at most hip flexors, adductors and quadriceps active).
   (b) Thoracolumbar or thoracolumbo-sacral lesion related to vertebral levels.
   (c) Kyphosis or scoliosis.
   (d) Grossly enlarged head with maximum head circumference of 2cm. more above 90th percentile related to birth weight.
   (e) Intracerebral birth injury.
   (f) Other gross congenital defects e.g cyanotic heart disease, ectopia vesicae or mongolism.

2. SPINA CLOSURE IN THE NEW BORN PERIOD.
   Meningitis or ventriculitis in an infant who already has serious neurological handicap and hydrocephalus.

...../24

3. LATER IN LIFE:

In any life threatening episode in a child who is severely handicapped by gross mental or physical defects.

The object of the criteria is to avoid treating those who would survive with severe handicaps and not to avoid treating those who would die early inspite of treatment given. There are difficulties in making this decision. For example there are people (Zachary, 1968, De Lange, 1974) who urg e that any life is better than no life. Or it is possible that a couple may have a myelomeningocele baby, and the only one after a long and desparate struggle. This couple may wish to have this child irrespective of whatever mental or physical handicap it may have. Rightfully therefore, the emotions of the parents should be considered before being told that their child cannot be offered any active treatment. The decision to treat or not to treat should be made by the most senior doctors, preferably a paediatrician who is looking after the child. The decision to offer any treatment should be taken after taking into consideration all the possible factors, including the most reasonable explanation in simple language to the parents about the possible and expected outcome of operation, should the decision be arrived at that no treatment is to be offered, the junior doctors and nurses should be put into the real perspective. they must be made to understand, without any ambiguity or exception, that nothing should be done which might prolong the survival of the infant, but that the child should be properly nursed and all suffering should be alleviated.

.. /25

Therefore:-

(1) The baby should be nursed in an ordinary cot with simple dressing of the back.

(2) Tube feeding, oxygen or resuscitation should be forbidden.

(3) If meningitis sets in, no antibiotics should be given.

(4) If hydrocephalus sets in rapidly, shunt procedure should be withheld because progressive hydrocephalus is an important cause of early death (Lorber lorber 1973, Lawrence 1974).

Although a policy of selective operation has now become standard in many centres, it is not universerly accepted. It is still being challenged by surgeons whose contributions in the field of spina bifida have been outstanding. Their main objectives are mainly on ethical grounds (Zachary, 1968, de lange, 1974).

Since the patients treated here were selected on a criteria widely used, the immediate post operative results should be comparable with those of other centres. The follow up here is inadequate and patients fail to attend after a certain period of attendance, and hence documentation is incomplete. Non-attendance is attributed to the poverty of the people in the rural areas, ignorance, low education level or difficulties in transportation due to bad roads etc. Or it is that the babies are dead within a short time from the second or third attendance in the follow up period ?. It is difficult to say. It is therefore not possible to know the fate of these children, the treated or the non-treated ones alike. It is an obligation to continue caring

...../26

for the treated child, so that all facilities must be employed without reservation. Just as the child who is not selected for treatment must not get any type of intervention which may prolong life, so does the child who is selected for treatment qualify, and very rightfully so, to continue having any intervention which is seen to promote or prolong life; and in as much comfort as possible. It is therefore for the benefit of the patient and also ethical and moral satisfaction of the doctor, to device a policy which makes follow up possible. This may help a great deal towards detecting the child who needs a revision of a procedure, or who needs a new procedure all together.

In this study it is seen that fifteen out of the thirty-seven (37) children operated upon had post-operative complications which must be viewed with concern because they are bad prognostic factors. These include hydrocephalus (6), bladder or bowel incontinence (15), complete paralysis of severe nature (8). This also means that a child was inflicted by more than one of these complications. These factors can be fatal in themselves even in short run. They are also usually the cause of fatality in the non-treated cases. It therefore means that if these cases are not given adequate and timely follow up their survival is not altered, and remains more or less the same as for those not treated.

The natural history of myelomeningocele is well documented by different groups of workers. For example, Lawrence described the outcome of 381 cases of myelomeningocele born in South Wales at a time when surgical

.../27

treatement was not practiced. At follow-up at 6 - 12 years only 10% of the patients were still alive and no fewer than 70% were, by any standard, grossly handicapped. It was estimated that without surgical treatment, even ignoring stillbirths and first day deaths, only 1 in 7 would reach school age and 1 in 70 would be fit to attend a normal school (Lawrence and Tew 1971). When the child is selected for treatment, intially the back closure is done. This prevents early meningitis which is the commonest cause of death in the immediate neonatal period. It also takes care of the possibility of ulceration of the covering of the sac which could lead to meningitis, septicaemia and death. It has been accepted that closure of the back, and more so if it done early (within 48 hours after birth) prevents the worsening of the neurologic deficit, and also minimuses the chances of infection. Other causes of death must be prevented and this almost always means surgical intervertion.

Hydrocephalus must be treated by some form of shunting procedure whereby cerebro-spinal-fluid is shunted from the ventricular system of brain to any of the sites mentioned below; right atrium, directly; peritoreim; or sometimes into some form of reservoir fashioned somewhere near the ventricular system. The commonest problem with any shunting procedure is the blockage of the system which may occur quite often; this means the doctor must be prepared to revise the procedure very many times and each time showing interest and paying as much attention to detail as the original procedure.

.../28

Incontinence of urine must be treated right from the beginning. Death from this may occur following an acute episode of urinary tract infection like pyelonephritis, or more commonly from progressive damage of the upper urinary tract due to recurrent infection, hydronephrosis or even later from progressive renal failure with or without hypertention. Hypertention may be the sole cause of death without any appreciable degree or renal failure. Urinary incontinence begins to cause bothersome problems during the school age period. It is quite a difficult problem to manage. It is wise to start with simple conservative measures for its control.

Urinary diversion would be avoided if such measures are successful. Or if they fail, then it would be more easier to accept diversion on the part of parents. Bladder training is started usually at the age of 3. The mother of the very young child is trained to do manual expression, and as the child grows older he takes the exercise. In the early stages of training some authorities recommend a high fluid intake and carefully selected adjuvant drug therapy (Hergardhet al, 1974).of carbachol and diazepam together. Portable urinals have social problems but they can be quite acceptable to boys, but girls normally do not accept them. With this manual expression is continued, catheter drainage has now been revived by some workers who have had bad experience with the diversion methods. (Duthie and Stark 1974, Forrest, 1974). A new type of catheter has been developed made of silicone-elastomer (silicath) which can be retained for 4-6 weeks.

.../29

Diversion methods for urinary incontinence has been used over the last two decades with a lot of enthusiasm after the first procedure was formed by Naish in 1956 in London. Some workers advocate ileal conduits while other advocate colonic conduits. Long term complications for children with ileal conduits urinary diversion have been observed (Dunn and Roberts, 1979). They observed over a period of 20 years, 82% of children develop complications and of the complications the more serious ones were progressive upper tract damage in (42%) severe recurent urinary tract infection, and severe psychological problems enough to warrant psychiatric treatment.

A similar follow-up for colonic conduits have been done. Studies show that this is much superior than ileal conduits in terms of occurence of late complications. Altwein et al (1977) observed late complications in only 14.3% of patients and only 20% in patients treated with ureterosigmoidostomy. Other workers report that there is no definite advantage of one method over the other (Elder et al, 1979).

PROGRESSIVE KYPHO-SCOLIOSIS is ultimately associated with recurrent chest infections and cor-pulmonale. These are associated with high mortality, but a major spinal deformity is relatively uncommon among the survivors of selective early operation. Extensive vertebral osteotomy for kyphosis used to be done at the time of back closure by Sharrard (1968), but results are generally not good because of the tendency to recurrence (Sharrard and Drennan, 1972). This is why kyphosis is regarded by many as contraindication to active treatment at birth. Established scoliosis is currently being managed causevatively

.../30

initially with a Milwaukee brace worn at night and then depending on improvement and progress. Delayed posterior spinal fusion is considered effective (Baker and Sharrard, 1973). It is therefore easy to appreciate that if survival of the treated cases is to be improved, not only must there be thorough and adequate follow up and continuous assessment, but the services of a paediatric surgeon, urologic surgeon and orthopaedic surgeon must be sought.

...../31

C O N C L U S I O N S :

THE FUTURE:

It is accepted that myelomeningocele arouses much psychological and social feelings to the parents, family and community. It is also quite true that the management of this condition is difficult, controversial and with generally poor results. The medical team caring for such a child may be beset with difficult moral, and ethical considerations before a decision of what type of management is best for a particular myelomeningocele patient. In a developing country like Kenya, resources are scarce and therefore there is inadequacy in manpower, hospital beds and therefore less patient time. Even when these things are available in adequate numbers, the patient population are generally poor and at best, of poor education making logical appreciation of the problem of management extremely difficult to apprehend, not to mention the associated difficult in follow up. The state is also bound by obligation to set aside financial resources and set up special institutions, while having available well trained and devoted general personnel to man such institutions.

The future therefore lies in prevention of myelomeningocele. It is agreed that the aetiology is multifactorial. Genetic factors play a major role in causation (Carter, 1974), and that racial variation is genetically determined. Consanguinity increases the risk of neural tube malformation and recurrence rate is quite high after a child has been born with spina bifida (Lorber, 1965; Carter and Roberts, 1967). Genetic factors are not the only things to go by because more-

...../32

zygous twins not only may be, but al ast invariably are discordant for spina bifida (Lawrence, Carter and David, 1968). The evidence for enviromental factors is still not complete and it is thought that the environment may just facilitate an already genetically determined condition to surface.

Genetic counselling should therefore be offered to all parents who have had an infant suffering from either anencephaly or spina bifida. This should be best done by the paediatrician. It is estimated that the offspring of survivors with spina bifida will have an incidence rate in the region of 3 - 4%, (Lorber, 1971C).

Ante natal diagnosis can now be made and so parents who have no religious or ethical objections to abortion can make use of facilities were available. Elevated levels of alpha-fetal protein (AFP) is diagnostic of anencephaly or open myelomeringocele when amniocentesis is done at 14-16 weeks of gestation (Brock and Sutcliffe, 1972). Recently maternal serum level of alpha fetal protein has been shown to be also diagnostic, although technically more difficult from the point of view of interpretation. Multiple pregnancy must be ruled out first using ultrasonography, because this is one of the causes of elevated levels of alpha-fetal protein.

Currently, some British medical researchers in five centres (Leeds University, Guys Hospital, Queens University, Belfast, Manchester University and Chester Hospitals) have come up with very interesting results. Their work was prompted by theobservation that there are

..../33

differing number of neural tube defects according to social class, and therefore incriminating nutritional factors. They have embarked on supplementing the risk mothers (those who have had one or more babies affected with some neural tube defect) with vitamins. They claim that they have reduced the expected incidence rate of about 5% to about 0.6%, a reduction of almost 90%. The researchers in their preliminary report say that although such reduction could be due to about four other reasons, the most likely is that the vitamins have prevented the defects. The final report is being eagerly awaited.

..../34

R E F E R E N C E S :

(1) ALTWEIN, J.E., et al (1977) Long Term Follow up of children with colonic Conduit Urinary Diversion and ureterosigmoidostomy Journal of Urology 118 832-836.

(2) BAKER R.H. & SHARRARD W.J.W. (1973) Correction of Lordoscoliosis in spina bifida by multiple spinal osteotomy and fusion with Dwyer fixation: a priliminary report.
Developmental Medicine and Child Neurology, supplement 29,12.

(3) BROCK D.J.H. & SUTCLIFFE R.G.: (1972) Alpha-fetoprotein in the antenatal diagnosis of anencephaly and spina bifida. Lancet ii,197.

(4) CARTER C.O: (1974) Clues to the aetiology of neural tube malformation. Developmental Medicine and Child Neurology, supplement 32,3.

(5) CARTER C.O. & FRASER ROBERTS J.A: (1967) The risk of recurrence after two children with central nervous syster malformations. Lancet i, 306.

(6) DE LANGE C.A: (1974) Selection in the management of patients with spina bifida aperta.
Developmental Medicine and Child Neurology, supplement 32,27.

(7) [illegible] P.A. & SMITH BLOCH A.M: (1951) Studies in spina bifida cystica. part 1: General survey and reassessment of the problem. Journal of Neurology, Neurosurgery and Psychiatry 24,131.

(8) DUNN J. & ROBERTS: (1979) The Long-term Results of Ileal Conduit Urinary Diversion in children.
British Journal of Urology 51,[illegible]61.

.../35

(8) DUTHIE E.J.W. & STARK G.D: (1974) Catheters for continence. A preliminary report on their trial in myelomeningocele. Developmental Medicine and Child Neurology, supplement 32,31.

(9) ELDER,D.D.,et al (1979) A Long-term Follow up of the Colonic Conduit Operation in children - British Journal of Urology 51,462-465.

(10) FEREMBACH D: (1963) Frequency of spina bifida occulta in prehistoric skeletons. Nature 199,100.

(11) FORREST E.M: (1974) The use of the Foley catheter for long-term urine collection in girls. Developmental Medicine and Child Neurology, supplement 32,54.

(12) [illegible] (1901) Rupture of the neural tube. The cause of myelomeningocele. Archives of Neurology 4,13.

(13) HIDE T.M., [illegible] P.P. & MILLS H.L: (1972) The outlook for the child with a myelomeningocele for whom early surgery was considered unjustifiable. Developmental Medicine and Child Neurology 14,196.

(14) LANCET, (1969) Editorial. Natural history of spina bifida, Lancet ii,34.

(15) LAURENCE K.M: (1974) Effect of early surgery for spina bifida survival and quality of life. Lancet i,301.

(16) LAURENCE K.M., CARTER C.C. & DAVIS P.A: (1960) Major central nervous system malformations in South Wales. - British Journal of preventive and Social Medicine, 22,146.

(17) LAURENCE K.M. & TEW B.J: (1971) Natural history of spina bifida cystica and cranium bifidum cysticum. Archives of Diseases in childhood 46,127.

(18) LORBER J: (1965) The family of spina bifida cystica pediatrics 35,589.

(19) LORBER J: (1971a) Results of treatment of myelomeningocele. An

..../36

analysis of 524 unselected cases with special reference to possible selection for treatment.
Developmental Medicine and Child Neurology 13,279.

(20) LORBER J: (1971c) What are the chances for the second generation ? Link 1,10.

(21) LORBER J: (1973b) Isosorbide in the medical treatment of infantile hydrocephalus. Journal of Neurosurgery 39,702.

(22) NERGARDH A., VON HEDENBERG., HELLSTROM B. & ERICSSON N.O: Continence training of children with neurogenic bladder dysfunction. Developmental Medicine and Child Neurology 14,47.

(23) SHARRARD W.J.W. & DREWAN J.G: (1972) Osteotomy - excision of the spine for lumbar kyphosis in older children with myelomeningocele. Journal of Bone and Joint Surgery [illegible],58.

(24) SHARRARD W.J.W., ZACHARY R.B., LORBER J. & [illegible] (1973) A controlled trial of immediate and delayed closure in spina bifida cystica. Archives of Disease in childhood 38,28.

(25) SMITH [illegible] SMITH E.B: (1973) Selection for treatment in spina bifida cystica. British Medical Journal iv,189.

(26) [illegible] TRYPHONAS H: (1973) Results of selective early operation in myelomeningocele. Archives of Disease in childhood 48,670.

(27) TRYPHONAS H: (1973) Three spina bifida defects in one child. Journal of Paediatric surgery 8,75-6.

(28) ZACHARY R.B: (1968) Ethical and social aspects of treatment of spina bifida. Lancet ii,274.