

Myelo-meningocele: A multi-disciplinary problem

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ABSTRACT

Background: Myelo-meningoceles are part of congenital afflictions of the spinal column. They arise from the failure of the neural tube to fuse properly during early embryonic growth. The causes and sequelae are multiple and, therefore, require multiple disciplines, to handle them. This study assessed the role of inter-disciplinary approach in the management of myelo-meningoceles. **Materials and Methods:** From 1975 to 2007, the author repaired 20 midline lumbar and lumbo-sacral myelo-meningoceles; 5 in Jamaica and 15 in Nigeria. There were 11 males and 9 females. Their ages, at operation, ranged from 1 to 168 days. All had urine and faecal incontinence and severe paraparesis to paraplegia. Skeletal deformities were present in 16 cases. The operations were carried out under routine general anaesthesia and in prone position. All cases were followed-up for up to 60 months, apart from one who died 4 days at home after discharge. **Results:** There were no deaths within the period of hospitalisation, usually about 14 days. Those followed-up have not made much improvement, though they were able to sit up without support and move around by shifting on their buttocks on the floor. **Conclusion:** We must continue to help these patients, but under the umbrella of specialised rehabilitation centres with the different specialists working together to make these patients attain a meaningful life and be useful to themselves and the society.

Key words: Disciplines, multiple, myelo-meningocele, problems

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INTRODUCTION

Spinal celes usually cause neurological deficits, skeletal deformities, ano-rectal and urinary bladder malfunctions, paraparesis, paraplegia and sensory loss below the cord level of involvement. Some factors postulated as aetiological, include Primary failure of neural tube closure,^{1,2} over distention and rupture of previously closed neural tube, disordered midline axial integration during gastrulation; that is the failure of the Hensen node to lay down properly, a single notochord, flanked by cohesive surrounding sheet of neuro-epithelium. Instead, paired notochordal anlagen develop from each half of the hensen node during gastrulation and two relatively independent hemineural plates, each developing into a hemicord, arise on either side of the node. This could induce malformation, which may disrupt neurulation resulting in either hemimyelomeningocele or myelomeningocele.³

Others include folate deficiency, Vitamin B-12 deficiency, teratogens, like valproic acid. The numbers 4, 5, 6 could also cause numbers 1, 2 and 3 mentioned above, ingestion of the leaves of *Mitragyna speciosa*, found in tropical South-East Asia,⁴ and anomalous ependyma could induce cord splitting and meningomyelocele.⁵

The need for the prevention of this lesion, which usually occurs during the first trimester of pregnancy, would, therefore, require the expertise of environmentalists, for clean and decent surroundings;⁶ dieticians/nutritionists to advice on the correct diet; obstetricians/embryologists to monitor the pre-cohabitation and antenatal periods, with a view, also, of detecting, *in utero*, such malformations and termination of the pregnancy encouraged.

The repair of the cele is carried out by the neurosurgeon/plastic and reconstructive surgeon; the urinary bladder problems are managed by the urologist;⁷⁻⁹ the ano-rectal ones are managed by the proctologist/entero-surgeon;¹⁰ while the orthopaedic surgeon/prosthetist takes care of the skeletal conditions, with the physiotherapist assisting.¹¹⁻²¹ Counselling of parents and guardians must not be left out.²² All of these procedures should be happening in one location of a special rehabilitation centre to promote good and adequate team work,

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without having to move the patient from one location to the other, or, doing one or some and leaving out the other(s). Maybe, habitation in and information from mars could eradicate this lesion with its multiple problems. The resultant inconveniences would hereby be minimised and the patient better integrated into the society.

Improper management of this case could lead to urinary tract infections, hydronephrosis, hypertension, cerebrospinal fluid leakage, atrophy of overlying tissue, growth out of proportion of sac and body, hip dislocations¹¹⁻¹⁸ kyphosis at the lesion level and death. Their chances of normal life of love, marriage and child bearing would be greatly jeopardised.

MATERIALS AND METHODS

From 1975 to 2007, the author repaired 20 midline lumbar and lumbo-sacral myelo-meningoceles [Figure 1] in Jamaica (5 lumbo-sacral myelo-meningoceles) and in Nigeria (2 lumbar and 13 lumbo-sacrals). There were 11 males and 9 females, with their ages ranging from 1 to 168 days. Only nine cases were referred by colleagues. The rest came to us on their own. All were midline and ulcerated. The patients all had sensory loss below L1 bilaterally, were paraplegic (but for one patient with severe paraparesis), urine and faecal incontinence. Sixteen (16) had skeletal deformities in form of pedes equinovarus and two were hydrocephalic [Table 1]. Laboratory tests included Venereal Disease Research Laboratory (VDRL) and retro-viral screening tests. Spinal X-rays were not done routinely to save costs. The spine and spinal cord levels were based on clinical findings. The operation [Figure 2], under routine general anaesthesia and in prone position, involved the separation of neural tissues from the overlying scarred tissue (Matrix), which was resected, then water tight closure of the dura, thereby enclosing the neural tissues, followed by skin closure [Figure 3]. Cultures of ulcerated wounds were done. Chloramphenicol and Vit. B Co. were given immediately post-operatively for 14 days. Stitches were removed 10 days post-operatively, apart from two cases, with persistent infection, where they were removed 4 and 6 days, respectively, post-operatively. All patients were discharged home about 14 days after the operations, to return to the out-patient's department 7 days later for follow-up checks. Physiotherapy of passive movements and gentle massage of the lower limbs was started on return to the ward.

RESULTS

The laboratory tests were within normal limits. The wound cultures yielded scanty growth of *Staphylococcus aureus*,

sensitive to ampicillin and chloramphenicol. The patients tolerated the procedures well and recovered from the anaesthesia without any additional gross neurological deficits. There were two wound dehiscences, following early removal of stitches due to continued wound infection. Healing was achieved with daily dressings. After about 60 months of follow-up [Table 2], the neurological and skeletal status remained as before the operations, although the wounds had healed in all and, the hydrocephalus in both patients arrested. They were all, however, able to sit up without support and shuffle around on their buttocks on the floor.

DISCUSSION

There was no recognised ante-natal care for their mothers, and so, did not have any vitamin B-12, or folic acid, the deficiency of any of which could have resulted in this malformation. There was also no history of intake of possible teratogens, like valproic acid or leaves of *mitragyna speciosa*,⁴ which could also have led to the malformation. However, they could have had native foods or concoctions containing teratogens. Teratogeniologists and embryologists could help with some information in this aspect. Environmentalists could assist identify any obnoxious elements in the surroundings, which are usually unhealthy.

Myelo-meningoceles are associated with neurological deficits and skeletal deformities. The careful repair

Table 1: Midline lumbar and lumbo-sacral myelo-meningoceles

Ulcerated	20
Midline lumbar	2
Midline lumbo-sacral	18
Bilateral loss of sensation below L1	20
Paraplegia	19
Paraparesis	1
Urine and faecal incontinence	20
Pedes equino-varus	16
Hydrocephalus	2

Table 2: Clinical presentation

Length of follow-up in months	3-60
Status when last seen.	Neurostatus: Unchanged in all. Urine and faecal incontinence: Unchanged in all. Pedes equino-varus: Unchanged. Hydrocephalus: Arrested in both. Wound infection/dehiscence: None. Could walk: None. Could sit up with support: 5 Could sit up without support: 14. Could shuffle around on buttocks: 14



Figure 1: Myelo-meningocele



Figure 2: Myelo-meningocele

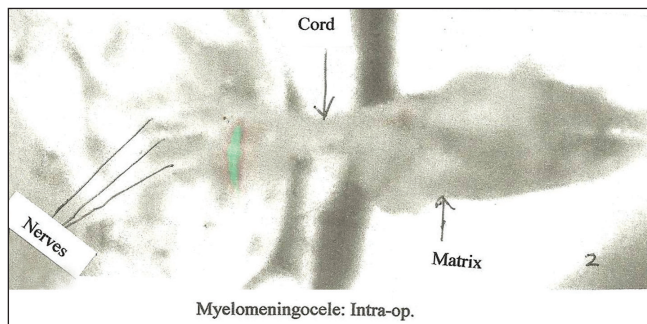


Figure 3: Myelo-meningocele

of these celes did not alter the neurological status post-operatively, since, it would appear, the damage to the nervous system was already done intra-uterine. This had to be done, not only for cosmetic reasons, but also to prevent further central nervous system infections and damage, cerebrospinal fluid leakage, atrophy of overlying tissue and growth out of proportion of sac and body. Stem cell therapy might be of use here. The hydrocephalus, fortunately,

was arrested in both cases. Otherwise, one cerebrospinal fluid shunting procedure or the other (ventriculo-atrial, ventriculo-peritoneal, etc.) would have been required. The persistent urine and faecal incontinence could be handled by diverting the urine and faeces into a colostomy bag, for example. Here, the services of specialists in this field would be required.⁷⁻¹⁰ The persistent skeletal deformities needed to have been taken care of by the orthopaedic surgeons/prosthetists,¹¹⁻²¹ even at this early stage. They need well trained physiotherapists. Our patients could not afford these specialists. They will eventually need institutional care, work in sheltered places, special schooling/education, job security to make them independent of charity and street begging. Their chances of normal life of love, marriage and child bearing will require a lot of counselling by experts.²² Our patients, when last seen, were still not old enough for this assessment.

The cost of investigations, treatment, follow-up, rehabilitation, reintegration into the society, is enormous and cannot be carried by their parents/relations alone.

CONCLUSION

It is most likely that for patients with myelo-meningoceles to make any meaningful livelihood and be integrated in and contribute positively to the society, a number of specialists must come together to their aid in well-organised specialised centres. Environmentalists must strive to sanitise our surroundings of obnoxious elements.⁶ Well-run ante-natal care must be encouraged. Hence, we are calling for such institutions to be established all over the world by governments, world health bodies, non-governmental organisations, philanthropists, etc. Nigeria and Nigerians must wake up and take the lead.

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