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Management of Spina Bifida Cystica at Ibadan

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Summary

Olumide, A. A. and Adeloye, A. (**1980**). *Nigerian Journal of Paediatrics* **7**(**2**), **46. Management of Spina Bifida Cystica at Ibadan**. Between January, 1973 and December, 1977, 132 children with spina bifida cystica were seen at the neurosurgical unit, and 70 per cent of these underwent surgery. The criteria used in selecting cases for surgery included a reasonable degree of motor function in the legs, imminent or actual rupture of the dural sac and the presence of social problems. Most of the patients were operated on as day cases. There was definite improvement in 40 per cent of the 37 patients with limb weakness, and in 47 per cent of the 38 patients with urinary incontinence. It is concluded that cooperation between neurosurgical, paediatric, orthopaedic and urological disciplines is essential if the child with spina bifida cystica is to receive maximum benefit from management.

THE clinical pattern of congenital malformations of the central nervous system (CNS) in Ibadan has been well documented (Adeloye and Odeku, 1972; Olumide, Odeku and Adeloye, 1975). These authors reported that spina bifida cystica formed the second largest group, accounting for 30–36 per cent of all CNS congenital malformations. Odeku (1967) and Adeloye (1971) have highlighted the problems of spina bifida cystica. The purpose of the present study was to document our experience in the surgical management of spina bifida cystica, including the long term results of surgery.

Materials and Methods

One hundred and thirty-two children with spina bifida cystica were seen at the Neurosurgical unit, University College Hospital (UCH), Ibadan, between January, 1973 and December, 1977. Of this number, 92 patients were operated on. Detailed history and clinical examination were carried out on each of the patients. Investigations included radiographs of the skull and spine in all cases, and air ventriculography in those with suspected or established hydrocephalus.

Patients selected for operation included those with reasonable degree of motor function in the legs, those in whom the sac of the spina bifida cystica had ruptured or was about to rupture, and those with social problems. In the last category, surgery was performed to facilitate the nursing care of a deformed but wanted child or on borderline cases who lived far away from Ibadan. Contraindications to surgery included flaccid paraplegia, advanced hydrocephalus, meningitis, and associated severe non-CNS malformations.

Results

Ninety-two (70 per cent) out of the 132 patients were selected for operation. These consisted of 56 males and 36 females, giving a male: female ratio of 3:2. The ages at presentation ranged between one day and one year (Table). Thirty-two patients (35 per cent) presented within the first week of life, and only three of these presented within the first 48 hours. Sixteen patients presented during the second week, while an average of seven other patients presented during the third, fourth, eighth and twelfth week, respectively.

TABLE

Spina bifida cystica: Age of operated patients at presentation in clinic

Age in weeks	*Number of Patients*
1	32
2	16
3	8
4	7
8	8
12	8
16	3
20	5
28	1
32	3
52	1

Level of Lesion

The lesion occurred at the lumbo-sacral region in 70 (76 per cent) of the 92 patients, the lumbar region in eleven (12 per cent), the sacral region in nine (10 per cent) and at the cervical region in two (2 per cent). The skin over the lesions was ulcerated in sixty (65 per cent) of the patients, and most of these were also infected.

Types of Lesion

The abnormalities were meningomyelocele (cord and roots within the sac) in 47 cases, myelomeningoceles (roots within sac) in 32, and simple meningoceles in 13 cases.

Associated Clinical Conditions

Other clinical problems associated with the malformation included urinary incontinence in 38 (41 per cent) of the patients, flaccid weakness of the lower limbs in 37 (40 per cent), patulous anus in 37 (40 per cent), hydrocephalus in 36 (39 per cent), sensory loss in 28 (30 per cent) and club foot in 26 (28 per cent). The most constant area of sensory impairment was in the saddle area (over the gluteal region). Club foot deformities were mainly equino-varus and calcaneo-varus types and were often bilateral. Laryngeal stridor occurred in three patients with associated hydrocephalus. Other congenital abnormalities included two cases of umbilical hernia, and one case each, of congenital heart disease and polydactyly.

Management

(*a*) *General*

All cases with ulceration of skin over the lesion received broadspectrum antibiotics and prophylactic anti-tetanus serum (ATS). The ulcer was dressed daily with Eusol. Surgery was performed only after adequate epithelisation of the sac.

(*b*) *Surgical*

Because of limited in-patient facilities, 74 (80 per cent) of the 92 patients were operated on as day cases, and sent home routinely on antibiotics. After an elliptical transverse or vertical skin incision, the rest of the surgery consisted of excision of redundant skin and dural sac, reconstitution of a new dural sac and approximation of the paraspinal muscle fascia in the midline. The skin is finally closed in one layer. We prefer a transverse incision for cosmetic reasons.

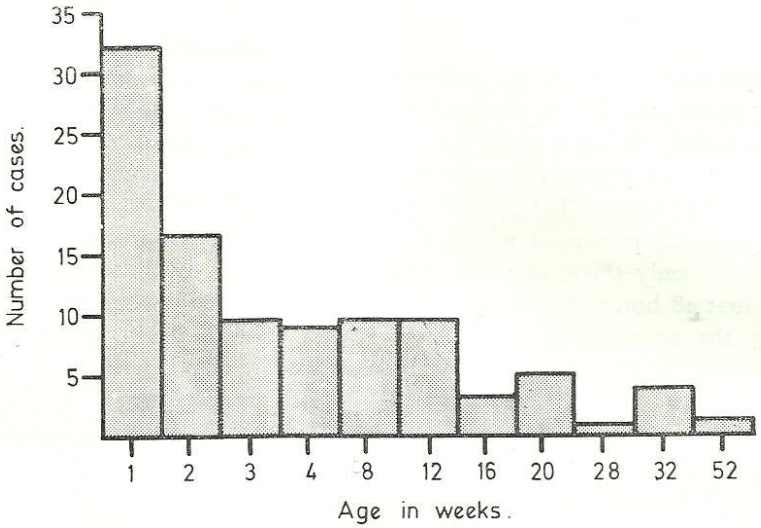

Fig. Histogram showing ages of operated patients at presentation in the clinic.

Most of the patients with associated hydrocephalus had spontaneous arrest, while a few of them arrested after intermittent ventricular tappings. Ventriculo-peritoneal shunts were performed on three cases before the repair of sac, while two cases were shunted after repair.

Post-operative Complications

Wound infection occurred in 12 (4 per cent) of the 92 patients. In the group of patients operated on as in-patients, wound infection occurred in seven (40 per cent) of the 18 patients; while in the group operated on as out-patients, wound infection occurred in five (6 per cent) of the 74 patients. It is significant that the incidence of wound infection was higher among those operated on as in-patients than among those operated on as day cases. Meningitis and ventriculitis occurred in two cases each, respectively. There was recurrence of the spinal swelling in three cases, while CSF leak occurred in one case. Post-operative hydrocephalus developed in ten patients who did not present with this complication prior to surgery.

Result of Surgery

The period of follow-up ranged between one month and six years. All patients were seen for at least a month after surgery. After six months, 37 patients (40 per cent) defaulted from follow-up. Eighteen (20 per cent) of the patients were followed up for more than a year. There was satisfactory improvement in 15 (40 per cent) of the 37 patients with limb weakness, and in 18 (47 per cent) of the 38 patients with urinary incontinence. Surgery did not appear to alter the extent of pre-operative sensory loss.

Discussion

It is now well established that early operation of spina bifida cystica decreases mortality and morbidity (Sharrad, 1963; Zachary, 1965; Mawdsley, 1966). Zachary (1965), advocates repair of the lesion within the first 48 hours of life since, according to this author, delay causes further neurological deficits. Guthkelch (1962) however, found no difference in muscle power

between children operated on early and those operated on late. It should be noted that his 'early' cases covered a much longer period than the first 48 hours of life. Here in Ibadan, it has not been possible to practise early repair in a majority of the cases who often present much later in life by which time infection had occurred. Inadequate in-patient facility is another factor against early operation here.

In Sheffield, Lober (1972) advocates selection of the cases for repair. Severe paraplegia, gross hydrocephalus, kyphosis and severe non-CNS malformations are contraindications to repair. In a developing country such as Nigeria, selection is determined by prevailing circumstances. Severely deformed cases born outside the hospital are never seen; those who present in hospital are usually not so disabled, so that on primarily social grounds, surgery is often considered.

The incidence of associated hydrocephalus is reported to be 70 per cent (Laurence, 1964). In the present small series, the incidence was 39 per cent which was due perhaps to strict selection criteria. In the past, there was an erroneous assumption that early operation with closure of the sac favoured development of hydrocephalus (Ingram and Matson, 1954). It has, however, been shown from ventriculographic and autopsy studies, that there is a high incidence of associated hind-brain malformations, notably Arnold-Chiari malformation, in spina bifida cystica (Zachary, 1965; Adeloye, 1976) which by itself is an important cause of congenital hydrocephalus.

Aetiology of spina bifida remains obscure. Recently, ante-natal diagnosis enabling selective termination of affected pregnancies has become feasible. Brock and Sutcliffe (1972) reported an association between high alpha-feto-protein levels in the amniotic fluid and anencephaly. A similar association has been demonstrated for spina bifida cystica (Alan *et al.*, 1973). Hagard, Carter and Milne (1976) examined the cost-benefit of introducing a programme for mass screening of pregnancies for the detection and abortion of foetuses with spina bifida cystica. They found that on economic grounds, screening may be worthwhile only in populations in which the incidence of spina bifida is high.

The treatment of a child with spina bifida cystica does not end with the repair of the sac. A multi-disciplinary approach is ideal. Most of these cases are first seen by the paediatrician. The orthopaedic surgeon corrects the club foot deformity, while the physiotherapist develops the functioning muscles to maximum performance. The urological surgeon gets involved about school age and may have to carry out urinary diversion operations in the presence of incontinence. Hence, cooperation between neurosurgical, paediatric, orthopaedic and urological disciplines is essential if the spina bifida patient is to benefit maximally.

Information on the epidemiology of spina bifida in developing countries is scarce. Unreliable hospital statistics and a high default rate from follow-ups, make an accurate study difficult. Thus, there is an urgent need for provision of reasonable hospital accommodation and rehabilitative facilities for improved handling of these unfortunate children.

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