Spina Bifida and Myelomeningocoele

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Spina Bifida is a congenital abnormality consisting of a separation of the spine of a vertebra into two halves. It may occur in various forms depending on associated features.

1. Spina Bifida Occulta

This consists of the bony deformity alone and is normally present in 5-10 per cent of people in the region of L.5.-S.1. Apart from skin blemishes over the area it very rarely causes any pathological symptoms.

2. Meningocoele

In this condition the spine of the vertebra is bifid and between the two halves a cyst of the meninges protrudes to form a lump on the back covered by skin. The cord is normal. The only problem is a cosmetic one and this may be corrected at any convenient time.

3. Myelomeningocoele

This consists of a deformed spinal cord over several segments surrounded by a cyst of the meninges.

This bulges between the bifid spines of several vertebrae and appears a as mass on the back usually uncovered by skin in its central portion. It may occur in any area of the spine but is most common in the lumbar area followed by the sacral and thoracic areas. It occurs at least twenty times as commonly as meningocoele, is the commonest single surgical abnormality seen on the first day of life and occurs in 2-3/1000 live births,

Since it is myelomeningocoele which constitutes the major therapeutic problem further remarks will be confined to this abnormality.

PATHOLOGY

Myelomeningocoele has several sequelae:

1. Because it is uncovered by skin it is liable to infection and untreated children die of meningitis or ventriculitis in a high percentage of cases.
2. In 80 per cent of cases there is an associated hydrocephalus. This is due to an abnormality of the base of the brain (the Arnold-Chiari phenomenon) consisting mainly of herniation of part of the cerebellum and medulla through the foramen magnum and arachnoid adhesions preventing the free circulation of C.S.F. If untreated, these children develop massive enlargement of the head and may be mentally defective.
3. The deformity of the spinal cord produces paraplegia below the level of the lesion which induces not only motor and sensory loss to the lower limbs of varying degree, but also deformity. The paraplegia also produces bladder paralysis with its secondary problems of incontinence or secondary infection. This infection may ultimately cause death after several years if unrelieved. The anus and rectum are also paralysed in these children producing not merely incontinence but often severe constipation because of loss of bowel sensation.

MANAGEMENT

The present-day approach to treatment is an aggressive one starting on the first day of life. The sac is surgically closed within hours of birth in the hope that this will prevent meningitis. It has also been shown by Sharrard *et al*[1] that early closure prevents deterioration in the paraplegia due to infection or drying out of the neural tube.

When this phase is over, a watch is kept for the development of hydrocephalus judged mainly by increased head circumference. If this is noted to be progressive (about 50 per cent of cases) drainage of the hydrocephalus is instituted.

This is achieved by ventriculo-atrial shunt, which consists of inserting a tube through a burr-hole in the skull into one of the lateral ventricles and then leading this subcutaneously into the jugular vein and threading it into the right atrium of the heart. The apparatus incorporates a pump which lies under the skin and can be palpated, and a valve which opens at a specific pressure and allows drainage of C.S.F. into the blood stream and prevents reflux of blood into the ventricle. The common valves in use are the Pudenz-Heyer and the Spitz-Holter.

These valves require frequent revision as they may block due to debris or clot at the distal end particularly when they are withdrawn from the atrium into the jugular vein with growth of the child. Most children only require these valves up to the age of two years when their hydrocephalus remits spontaneously but quite a few require them for the rest of their lives. A careful check must be made at regular intervals to ascertain that the valve is functioning. This can be done by palpating the pump and by head measurement before the sutures are closed. Once the sutures are united, the only signs of non-function are those of increased intracranial pressure, e.g. headache, drowsiness and vomiting. Once the hydrocephalus is controlled, attention is turned to the orthopaedic and urological aspects of the paraplegia which are dealt with in the following articles.

It is vitally important that the various disciplines concerned co-ordinate in the management of these children. To this end special clinics must be established where the specialists concerned can see these patients together and discuss their individual management. Such a combined clinic is held weekly at the Transvaal Memorial Hospital for Children in Johannesburg and is run by a paediatric surgeon, orthopaedic surgeon and urologist. An opthalmologist also attends because of the high incidence of squints. An orthopaedic technician, social worker, educational psychologist and physiotherapists complete the complement. Since the management is so complex, it is likely that, no matter how good a home this child comes from, it is best managed either as an in-patient or an out-patient in an institution specially geared to its problems and where expert nursing, physiotherapy, occupational therapy, and schooling can be provided under one roof. The Hope School and Homes have proved ideal for this purpose in Johannesburg. Because of the profound impact such a crippled child makes on his home, the various social problems should be attended to by a social worker.

RESULTS OF TREATMENT

If an energetic programme of treatment is instituted from birth, about half of the children with myelomeningocoele will survive and about 85 per cent of the survivors will have

reasonable intelligence. Because of their severe disability, it is likely that a large proportion will require some form of sheltered employment in the future.

THE ETHICAL PROBLEM

It might be argued that these children would be better left untreated in the hope that they would soon die. However, we know from the studies of Laurence[2] in Cardiff that, if completely untreated, about 20 per cent will survive anyway. These will then be severely disabled, many with large heads and tumours on their backs and with a high incidence of mental deficiency. Active treatment therefore produces more cripples but fewer mentally deficient children with less deformity. At present, therefore, active treatment appears the best alternative particularly since advances, e.g. electronically controlled bladders, etc. are a future prospect.

Selection of cases for treatment does not appear to be a tenable solution since there is no means of telling, at birth, which cases will either survive or do well on treatment. Should one then have left a child untreated and it survives, one might have missed the opportunity of achieving the best result as regards rehabilitation.

THE FUTURE

The embryological defect in myelomeningocoele occurs within the first 6 weeks of foetal life and, though many theories exist, the precise mechanism is obscure.

In many cases the abnormality probably occurs as a result of some pathology in the mother, e.g. virus disease, taking of drugs, etc. though this is not proven. However, it is certain that in a high proportion of cases a genetic factor exists, the transmission of which is ill-understood. Parents should be warned that after the birth of a child with myelomeningocoele the chances of having a further child with the abnormality is about 1:25. After a second child is born with the abnormality the chance of a defect in another child is about 1:8. The chances would obviously be increased too if there was a history of any central nervous system defect in the immediate family.

Since there is a genetic factor involved, the likelihood exists that myelomeningocoele will become more prevalent in future generations. We are unlikely to be able to prevent the condition occurring in the foreseeable future and should therefore make strenuous efforts to increase our therapeutic potential in the rehabilitation of these unfortunate children.

REFERENCES

1. SHARRARD, W. J. W., ZACHARY, R. B., LORBER, J. and BRUCE, A. M. (1963) Arch. Dis. Child., 38:18.
2. LAURENCE, K. M. (1966), Dev. Med. Child. Neurol., Suppl. No. 11, p. 10.