Problems of the Newborn

Spina Bifida and Hydrocephalus

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British Medical Journal, 1971, 4, 796-799

Congenital anomalies of the central nervous system account for a very large proportion of morphological defects in the newborn. The most common of these—spina bifida cystica—has a wide range of incidence in different countries, being highest in the western hemisphere, where in some areas the incidence is 4 per 1,000 births. A general practitioner in Britain with 2,000 patients on his list could expect a new case to occur in his practice once every five years.

Prompt and accurate advice to the family with a newborn child with such a major lesion and efficient handling of the infant will lay a good foundation for the future management of the child and of its family, whether or not the infant survives. Vacillation and changes of plan based on the apparent expediency of the moment and on emotion will undermine confidence and create guilt and anxiety so deep rooted that the family may breakdown.

Conversely 80% of infants with myelomeningocele also exhibit hydrocephalus.

The diagnosis will not be in doubt if the head size is big enough to impede labour, but most children with hydrocephalus have a normal head circumference at birth. Measurement of the head circumference (Fig. 1) should always be part of the routine neonatal examination, and the most valuable clue to congenital hydrocephalus is the disproportionate increase in this circumference during the first week or two after birth. The "setting sun" sign (Fig. 2) is often present even if the head circumference is initially normal. Palpably widened skull sutures are very suggestive of hydrocephalus, and in a full-term infant an enlarged posterior fontanelle and separated occipital and parietal bones at the lambdoid sutures are almost pathognomonic.

Encephalocele is a protrusion of the meninges through a midline defect in the occipital bone (Fig. 3). It usually contains some brain and may be associated with microcephaly.

FIG. 1—Head Circumference Chart.

Hydrocephalus

Hydrocephalus presenting in the newborn may be due to several different malformations. Four-fifths of the cases of congenital hydrocephalus in the United Kingdom are associated with spina bifida cystica and the Arnold-Chiari malformation.

Some very large encephaloceles may contain very little brain tissue and successful removal may be followed by normal development.

Spina Bifida

TYPES

Occult

The occult forms of spina bifida with hairy patches (Fig. 4), naevi, or fibro-fatty lumps (Fig. 5) over the spine do not need emergency treatment, but the infant should be carefully

Fig. 2

Fig. 3

Fig. 4

FIG. 2—Early "setting sun" sign. The sclera above the cornea was visible at times and the eyes were always more staring than would be expected in a 6-day-old infant. His ventriculogram showed gross dilatation of the ventricles. FIG. 3—An encephalocele. No neural tissue was found in the sac. Hydrocephalus was present. FIG. 4—A patch of hair overlies a spina bifida occulta.

Fig. 5

Fig. 6

Fig. 7

FIG. 5—Hamartoma of the cauda equina. A mainly lipomatous mass protrudes through the meninges and bifid spinous processes. FIG. 6—A flat "window" meningocele. FIG. 7—A high thoracic meningocele with a thin membranous sac.

examined early in life. Very rarely is there any neurological lesion initially, but any tethering of the cord may well cause damage as the child grows. Dimples in the midline of the back, other than the very common ones that lie directly over the coccyx, may be sinuses communicating with the meninges, and are a rare cause of recurrent meningitis. They again need early assessment.

Meningocele

The true meningocele takes several forms (Figs. 6 and 7) and may occur at any spinal level. It is not associated with neurological defects in the underlying cord, but is important because occasionally hydrocephalus is an associated lesion and because if the skin covering the protruding meninges is thin the sac may ulcerate and rupture and meningitis may follow. Early

treatment of any hydrocephalus and urgent operative closure of the back if rupture is likely will lead to survival free of disability.

Myelomeningocele

A myelomeningocele is characterized by part of the malformed spinal cord or cauda equina being exposed on the surface of the back. This neural plaque may be the dome of a protruding sac (Fig. 8) or lie flattened over a broad depression in the vertebral bodies (Fig. 9). There may be a considerable associated kyphosis and the bony defect may be of a few arches only or may affect almost the entire spinal column. The lumbosacral region is most usually involved, but occasionally cervical and thoracic lesions are seen.

Neurological complications are common and depend on the

FIG. 8—A cystic sacral myelomeningocele. A small plaque of neural tissue surmounts a sac of fluid.

FIG. 9—A thoracolumbar flat open myelomeningocele with kyphosis.

amount of spinal cord exposed and damaged. The assessment of the extent of the disturbance is very difficult. If the infant is cold, fewer muscle groups may appear to function than really can do so. If the legs themselves are stimulated, reflex activity may give a falsely optimistic view. Congenital dislocation of the hips and deformities of the feet and ankles are common, owing to imbalance of the muscles serving the affected joints. There will be areas of sensory loss in the skin below the level of the myelomeningocele. The trophic lesions that may develop because of this will not be present for at least some days. Disturbance of bladder function is not readily recognized in the neonate, but may be presumed if there is paralysis of the other muscles whose spinal centre is in the 2nd-4th sacral segments (lateral hamstrings, calf muscles, intrinsic muscles of the feet, the muscles of the pelvic floor, and sphincter ani).

Up to a quarter of infants with myelomeningocele are still-born and a further 10% may die from general perinatal causes. If untreated, nearly half will die from specific complications of the condition, but, the surprising fact is that 15% of the total will survive if given only basic care.

Management

There is no simple universal answer to the problem created by the unexpected birth of an infant with a major anomaly of the central nervous system. In the simpler lesions the indica-

tions for operative correction of the defect are not called into question, but for an infant with a myelomeningocele where some degree of handicap is inevitable there is considerable difference of opinion about the vigour with which he should be treated. If all children treated by early surgery had a normal unhandicapped life subsequently, or if all babies not treated were to die promptly, the problem would be solved. Efficient surgical management will give an overall survival rate of 60% or more, and some of the survivors will be severely handicapped. With no special treatment 15% will survive and these are not necessarily the least handicapped infants.

The number of survivors with no handicaps or minimal problems in a series of untreated infants is much smaller than the number of children of similar quality in an equivalent group of surgically treated ones. Some infants who have minimal handicaps must, therefore, either die if untreated, or their neurological damage progresses if the myelomeningocele is left exposed. If surgery is beneficial then all those who would survive without it must be given its benefits. To ensure this, and to reduce the number of severely handicapped children surviving, attempts have been made to draw up a list of criteria that make an infant "not worth operating on." A poor expectation of life and a poor quality of survival are certainly associated with extensive paraplegia, gross hydrocephalus, other associated malformations, established renal damage, and birth injury. Nevertheless, the infant who exhibits some or all of these characteristics may well survive, and the present evidence shows that his handicaps will have been increased by delay in treatment. In my opinion prompt surgical treatment of nearly all live-born infants with myelomeningocele is the best course open to us. The infant who does not have his myelomeningocele closed at birth and who survives, presents many more difficult problems of management in later years than the infant with a similar lesion subjected to operation.

DECISIONS AFTER BIRTH

The first and most important duty of the doctor on delivery of a live-born infant with myelomeningocele is to the parents. The mother is not usually in a position to give reasonable or informed consent to any proposed form of management, but obviously must be told something about her child's condition. The father is also often not capable of making a real decision of his own, and the burden of responsibility almost invariably falls on the doctor, who must explain the possible handicaps and disabilities and lead the parents to consent to the course of action which he honestly feels best. The initial discussion with the parents should always emphasize that to the best of our knowledge no action or omission on the part of the mother during the pregnancy caused the deformity. This may well need repetition on other occasions to try to allay her natural feelings of failure and guilt.

Early surgical closure of a myelomeningocele has certain clear advantages. Removing the swelling and closing the open wound makes nursing simpler and is usually essential if the child is to be looked after at home. The nervous tissue of the plaque is protected from further damage and its function is preserved. Provided primary healing occurs, the child is protected against infection of the meninges, and investigation of any associated hydrocephalus can proceed promptly, with consequent preservation of cerebral function and control of the size of the head. When the back has not been operated on the child will almost certainly have to be nursed in hospital for many months. Moreover, treatment of the hydrocephalus will have to be delayed because of the high risks of infection of the central nervous system if any valve is inserted in the presence of an infected back wound.

Ideally, unless associated abnormalities, birth injury, or other factors make it very likely that the infant is going to die

within a few days, urgent transfer to a surgical unit should be arranged. The neural plaque should be covered with a sterile dressing kept moist with sterile saline or aqueous chlorhexidine solution. The infant should be kept warm during transfer in a portable incubator, and the father should, if at all possible, accompany him to discuss his management further.

The back should be closed and any gross kyphosis corrected within 48 hours (preferably 24 hours) of birth as the risk of infection of the back and the meninges becomes too great if there is further delay. With satisfactory wound healing, full assessment can then begin of the urinary tract, of leg activity and deformity, and of the hydrocephalus, and appropriate early treatment started to minimize these disabilities.

Prognosis for the Child

The wide variety of form and extent of the lesions of the spinal cord, and the varying use of descriptive terms for them, make classification and comparison of different series of results difficult. But most experts agree that a policy of early surgical treatment of open myelomeningocele ensures that a higher proportion of children survive who are either normal or who have only minor handicaps. Varying degrees of paraplegia and urinary incontinence occur in half the cases, but are coped with remarkably well by those children who are of reasonable intelligence.

The results of treatment of hydrocephalus both in isolation and associated with myelomeningocele depend largely on the amount of associated brain damage or the presence of any other deficit. Nowadays the surgeon can almost always control the actual size of the head, so that the vast or unmanageable heads of untreated hydrocephalus should be a thing of the past. Measurements of the intelligence quotient, for what they are worth, have shown surprisingly good results in many of the treated children who are now reaching school age. No long-term detailed studies are yet available.

Prognosis for the Family

The arrival of a new child in a family always has profound effects on that family. If the child is malformed the effects are far reaching and may be disastrous. The fact of the infant's malformation is apt to produce feelings of inadequacy and guilt especially in the mother, and she needs reassurance and help. Early death may seem to solve a good many problems in the doctor's view, but not so obviously to the parents. If the child survives and is handicapped then the parents will continue to need a great deal of support and practical help and advice on how to handle him in relation to any siblings. Finally, the parents will always be anxious about the outcome of any future pregnancies. In Britain any pregnancy carries risk of 0.4% of spina bifida cystica. If a family already has one child with a major anomaly of the central nervous system, then the risks in future pregnancy are perhaps as high as 4%. If there are two or more close relatives who have had such lesions, the risk rises to 20%, which is very high indeed.

Conclusion

The proper course of treatment for the infant with a myelomeningocele is the subject of considerable debate at present. The survival rate of children with myelomeningocele with prompt surgery is about 60%, as opposed to 15% with no treatment. In my opinion early and vigorous treatment is justified in the great majority of cases by the considerable advantages accruing for the operated child compared with the increased disability in the unoperated child who survives.

Early treatment of hydrocephalus is again essential if brain damage is not to progress, and investigation and surgery in the neonatal period should be undertaken.

Small "closed" lesions of the spine are not associated with the same urgency, but early referral to a specialized unit for full assessment is desirable.

Scientific Basis of Clinical Practice

Electrical Activity of the Heart

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British Medical Journal, 1971, 4, 799-803

The heart is essentially a specialized muscle. Its function is to contract rhythmically and by its contraction move blood from an area of low hydrostatic pressure in the great veins to an area of high hydrostatic pressure in the aorta. So the heart is a muscular pump whose activity provides the work necessary to drive the blood round the circulation. As in any other sort of muscle, mechanical contraction depends on the electrical activity of the cells. To function efficiently as a pump, a certain sequence of events must occur—the heart cycle. This sequence of events is controlled by the electrical pro-

perties of the cells, and the heart beat owes its inherent rhythmicity to these characteristics.

Membrane Potential

Heart cells are like other muscle cells. They have similar constituents including actin and myosin filaments forming the contractile machinery, and a cell membrane. Like other cells there is an electrical potential difference between the inside and the outside of the heart cell, the inside being negative with respect to the outside in the resting state. The electrical properties of the cells depend largely on the properties of the membrane. This is selectively permeable to small inorganic ions, and its permeabilities to these ions varies during the heart cycle because of changes in the potential difference across the membrane. Conversely, the potential difference

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