Myelomeningocele

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Introduction

Myelomeningocele has, in the last 10 years, become one of the most important and commonest problems that a paediatric surgeon has to deal with. The abnormality has greatly affected the entire hospital service in relation to children and has made a great impact on the social services. There is no good evidence that this particular abnormality has become more common but there are a number of perfectly valid reasons why children with myelomeningocele should present a major problem in 1970 when they did not do so some 20 years ago. The simple answer is that many more of the infants born with this particular abnormality are now surviving than in the past. It is generally estimated that without treatment, about 10% of myelomeningocele infants will survive for at least a year (Laurence & Tew, 1971), and a small but significant number survive much longer; grossly deformed and often mentally retarded.

The reasons for the increased survival rate include the following:

(a) The overall advances in paediatric surgery and especially in neonatal anaesthesia.

(b) The introduction and development of antibiotics which can treat or prevent meningitis which used to be a common cause of death in this group of patients.

(c) The development and introduction of ventriculo-atrial drainage using shunts by Holter (Macnab, 1963) and Pudenz (1966).

(d) The concept introduced originally by Sharrard and his colleagues in 1962 (Sharrard *et al.*, 1963) that emergency closure of the spinal defect within 24 hr of birth produced not only a higher survival rate but also a better quality of survivor. They claimed that early closure prevented ascending meningitis, reduced the degree of paralysis and had no adverse influence on the development of hydrocephalus.

Largely as the result of the Sheffield school of teaching, emergency closure of myelomeningocele became an accepted and standard practice and the mortality has dropped to around 30% (Eckstein & Macnab, 1966). It is noted that of those children who die, 90% will die before their second birthday and indeed the large majority of these will die within the first 6 months.

The whole problem of myelomeningocele patients has been highlighted by a recent publication of Lorber (1971) analysing the results of so-called early closure and the fact that the large majority of the survivors of this treatment are grossly handicapped. A not insignificant proportion is mentally retarded. Lorber, like others before him, therefore suggests selection for treatment and gives criteria for such a selective process. The idea of selection, while sound at first sight, presents a great many problems. While simple facts such as hydrocephalus at birth, complete paraplegia and obvious kypho-scoliosis are regarded as contra-indications to surgery none of these criteria can be regarded as absolute. In my experience, quite a number of infants who had obvious hydrocephalus at birth and were treated with a ventriculo-atrial shunt at the same time as their myelomeningocele was repaired, have done remarkably well. Complete paraplegia by itself cannot really be regarded as an absolute contra-indication to treatment as the totally paralysed infant or child may be far better off than his partially paralysed counterpart who is likely to have multiple lower limb deformities caused by muscle imbalance. Those who advocate selection fail to recognize the simple fact that a great many of the so-called untreated myelomeningocele patients will survive for many months or even years and cause a great deal of hardship to their parents and their families. If selection is to be practised no hard-and-fast rules can be made and the individual infant must be assessed in relation to his individual family environment. A myelomeningocele baby of a 16-year-old unmarried mother may well be handled differently from the clinically identical infant of a couple who have tried for many years to have a child.

Myelomeningocele

The actual closure of the myelomeningocele is relatively straightforward. The neural plaque should be isolated and preserved but all membranous tissue should be excised so as to leave a clean skin edge. Primary closure of the dura is usually possible, and following wide mobilization to the flanks the skin can usually be closed producing a vertical suture line (Eckstein, 1971). Rotation flaps should be

avoided as these tend to slough and in any event subsequent scars will interfere with the wearing of calipers and urinary appliances. Muscle flaps are contra-indicated as these will tend to increase kyphoscoliosis later on, but in the occasional patient, a spinal osteotomy at the initial operation is justifiable to correct gross kyphosis and to allow primary skin closure (Eckstein & Vora, 1971), (Figs. 1 and 2).

Hydrocephalus

Between 80 and 90% of myelomeningocele patients will develop progressive hydrocephalus. The signs and symptoms of this complication or associated malformation include widening of the sutures, a tense fontanelle, rapidly increasing skull circumference, vomiting, drowsiness or a general failure to respond to stimuli. The diagnosis of hydrocephalus can obviously be confirmed by ventriculography but this procedure is not strictly necessary as a confident

FIG. 1. Myelomeningocele with gross kyphosis.

FIG. 2. Same patient after spinal osteotomy and primary skin closure; note that shunt has also been inserted at the same time.

diagnosis can usually be made on clinical grounds. The one essential pre-operative investigation before shunt therapy is an analysis of a specimen of cerebrospinal fluid with particular reference to its protein content on the one hand and to its sterility on the other. While a high protein content (1000 mg/100 ml) is no absolute contra-indication to shunt therapy, it is widely recognized that ventriculo-atrial shunts will not function satisfactorily in the presence of a high protein content.

We have found it useful in this situation to set up an experimental drainage system (using a discarded but functioning valve) with the patient's CSF and to assess the pressure at which the valve will function when the protein content of the CSF is raised. The effectiveness of the shunt is not directly correlated to the protein content of the CSF. Sterility of the CSF is obviously essential before a ventriculo-vascular shunt can be performed. Progressive hydrocephalus associated with active ventriculitis has to be managed by daily ventricular taps or by continuous drainage; neither procedure is without risk and a severe electrolyte depletion may result from prolonged external drainage of CSF.

The standard treatment of hydrocephalus today is the use of ventriculo-atrial shunts usually employing either the Holter or Pudenz system. Both shunt systems are similar in their working principles although their external appearance differs considerably. There is probably little to choose between these types of shunt although the writer has had considerably greater success using the Holter system. Both in the U.K. and on the Continent, paediatric surgeons tend to use the Holter system while neurosurgeons use the Pudenz system; the complication and failure-rate of large reported series is almost identical in both situations. The insertion of a ventriculo-atrial shunt is relatively simple and carries a negligible mortality but the complication rate is alarmingly high (Tsingoglu & Forrest, 1971). Approximately 12% of shunt-treated patients will develop bacteraemia due to colonization of the valve usually with *Staphylococcus albus*. Such bacteraemia is best treated by immediate shunt-replacement under full antibiotic cover (Nicholas, Kamal & Eckstein, 1970). Blockage of the shunt is common and may occur either at the proximal or distal end. There is a tendency for the choroid plexus to grow into the ventricular catheter and to block this and the use of the Rickham reservoir is said to reduce such blockage (Rickham, 1964). In any event, blockage of a ventricular catheter is relatively simple to correct surgically. The distal end of the catheter is likely to become blocked when, as a result of growth, it comes out of the right atrium into the superior vena cava. Clot formation then takes place around the catheter which seals it off from the circulation. Prophylactic

lengthening of the distal catheter has been recommended to avoid blockage of the distal end of the shunt. The position of the distal end of the shunt can be checked at all times, especially with the Holter system, as the tubing is radio-opaque. Other complications of shunt therapy such as perforation of the heart are uncommon and from a purely surgical point of view the technical problems start when all four jugular veins have been used (Dickson *et al.*, 1969). In such an event, the CSF can be shunted either into the peritoneal cavity, the pleural cavity or the heart, using either the azygos vein as a means of access or a direct cardiectomy. At least half the shunt-treated patients will require further surgery for their shunt.

Orthopaedic problems

The large majority of myelomeningocele patients will have a greater or lesser degree of paralysis in the legs, often associated with a considerable amount of muscle imbalance and subsequent deformity. The child with total paraplegia usually has straight legs and can be fitted with calipers relatively easily. Children with partial paralysis usually require multiple orthopaedic operative procedures to correct deformities such as talipes equinovarus, calcaneus deformity, genu recurvatum, fixed flexion of the knees or dislocation of the hip. In children with dislocation of the hip-joint, it is particularly important to remember that such dislocation is paralytic and not truly congenital and that conservative methods by manipulation or splintage are unlikely to succeed (Walker, 1968). While many myelomeningocele patients will ultimately be confined to wheelchairs it is a generally accepted policy that all children with myelomeningocele should be given the opportunity to walk in calipers provided their intelligence is adequate, and multiple orthopaedic surgical procedures are justifiable to make a particular patient suitable for ambulation. Research into the development and design of calipers is urgently needed.

Urinary continence

Although a previous study of mine had suggested that over 30% of myelomeningocele patients would develop normal urinary control (Eckstein, 1968), it is now clear that that particular series was selected and it is now generally agreed that at the most, 10% of myelomeningocele children will develop normal control. The problems associated with the neurogenic bladder are therefore important aspects of this congenital abnormality. Intravenous pyelography should be performed soon after birth and thereafter at 2-yearly intervals to detect upper tract dilatation. Regular urinalysis is essential to diagnose urinary tract infection so that this can be treated with suitable antibiotics. There is some evidence that such infec-

tion may in many of these particular patients be localized to the bladder only and in this situation, antibiotic therapy is indeed not necessary. On the other hand, if there is any clinical evidence of upper tract infection or radiological evidence of upper tract deterioration, then prolonged antibiotic therapy is essential. Upper urinary tract dilatation in infancy can often be arrested or improved by a transurethral resection of the external sphincter (Figs. 3 and 4). It should be noted that the internal sphincter or bladder neck in this particular group of patients is usually widely open and a bladder neck resection is therefore not only unnecessary but positively contra-indicated. The majority of male children with incontinence can ultimately be managed with a penile appliance, provided the penis is of adequate size. Since no suitable appliance exists for the female, incontinent females have to have a urinary diversion to the skin using either a loop of ileum or colon to provide a suitable conduit. While the present methods of urinary diversion, whether cutaneous ureterostomy or ileal conduit, carry a significant proportion of early complications, such diversions appear to function well for at least 5 years and possibly up to 10 years. There is, to date, insufficient information to allow us to predict the function of such conduits for periods exceeding 10 years.

FIG. 3. Excretion cystogram showing upper tract dilatation.

Fig. 4. Same patient, 5 months after transurethral resection of external sphincter.

Undoubtedly one of the most important recent developments in relation to urinary diversion to the skin is the introduction of plastic disposable appliances. The old rubber bags were difficult to clean and quite impossible to sterilize and should no longer be used. Stoma complications have become most unusual since the plastic disposable bags ('Car-shalton' by Eschman's or 'Mitcham' by Down Bros.) were introduced. Other essential features of the new disposable appliances include translucency so that the stoma can be inspected without removing the appliance and the use of a non-return flap valve to avoid the escape of large quantities of urine if the bag should accidentally come off. One of my patients with an ileal conduit diversion practises judo and many others play soccer or even rugby so that such a urinary diversion need not necessarily restrict the activities of these children.

Contrary to expectation, stone formation following congenital neuropathic bladder is relatively uncommon in contrast to this complication in patients with traumatic paraplegia. While it may well be that renal failure will play an important cause in the mortality in myelomeningocele patients in the years to come, there is to date no satisfactory evidence that this will indeed happen.

Bowel continence

Just as the majority of myelomeningocele patients have a neuropathic bladder, so a similar proportion have a neuropathic bowel. In practice, however, faecal incontinence is much less of a problem, largely because this group of patients tends to be constipated. At the most, only 50% of myelomeningocele patients have rectal incontinence as judged by social criteria and in a recent study, we were unable to correlate rectal function to the level of the neurological lesion, the degree of limb paralysis or indeed to bladder function. It appears that good rectal training was the most important factor in achieving social continence (Scobie, Eckstein & Long, 1970). Conscientious parents were the only common factor which improved bowel continence in this group of children.

Education

The education of myelomeningocele patients is giving rise to considerable concern. On the whole, myelomeningocele patients are of normal intelligence and their admission to normal schools is governed by the degree of paralysis and of incontinence on the one hand, while on the other hand the local facilities for schooling are of paramount importance. Approximately half of our myelomeningocele patients are attending normal schools at primary school level but there is likely to be an alarming drop when these same children reach the stage of secondary education. In this particular case, the problems are not related so much to the children as to the physical characteristics of the schools. While the majority of primary schools are relatively modern and one-level buildings, most secondary schools are old and require the negotiation of stairs to get pupils from one classroom to another. Such simple architectural problems are likely to interfere with schooling of these multiply handicapped children. There is no doubt whatever that the facilities for post-school vocational training for handicapped people are totally inadequate and it is only hoped that suitable facilities will be provided in time as the flood of myelomeningocele patients and multiply handicapped adolescents increases at a steady and alarming rate.

The fate of these unfortunate children born with myelomeningocele depends not only on whether we can provide the necessary hospital facilities and surgical expertise but also on whether the State will provide, in time, suitable facilities for vocational training and sufficient sheltered employment to justify the whole treatment of spina bifida and all its associated problems.

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