reduces the residual urine and therefore the opportunity for treatment with a urinary antiseptic such as a soluble sulphonamide or mandelic acid. The urine is examined for infection at frequent intervals.

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 clinical studies of Laurence 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 myelomeningocele 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 myelomeningocele 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 myelomeningocele 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


The Management of Urological Complications of Myelomeningocele

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The nerves which supply the bladder arise from the sacral portion of the spinal cord. Failure of development of this portion of the cord necessarily results in, at least, some degree of neurogenic bladder dysfunction.

The resultant stasis of urine in the bladder together with vesico-ureteric reflux of urine leads to persistent infection of the kidneys. The pyelonephritis would eventually destroy the renal substance if measures were not taken to prevent this chain of events. Cause of death of the child with a myelomeningocele, who survives the initial closure of the defect and hydrocephalus, is commonly renal failure due to pyelonephritis.

The approach adopted at the Spinal Defects Clinic, from the urological aspect, is essentially conservative. The regime consists of full urological investigation in the form of clinical examination and X-ray studies of the bladder and upper urinary tract. The bladder is filled with radiopaque dye in order to study its outline and confirm or refute the presence of vesico-ureteric reflux of urine. An intravenous pyelogram outlines the calyces, pelvis and ureters to establish the function of the kidneys and the degree of damage sustained. The urine is examined microscopically and cultured to isolate the infecting organism.

Treatment consists of clearing the urinary tract infection with an appropriate antibiotic, followed by a long-term treatment with a urinary antiseptic such as a soluble sulphonamide or mandelic acid. The urine is examined for infection at frequent intervals.

The most important bulwark of conservative treatment is suprapubic manual expression of the bladder. This reduces the residual urine and therefore the opportunity for infection to gain a foothold. The mother or nurse is instructed to empty the bladder at 2-hourly intervals during the day. When the child is older he is taught to express his own bladder.

X-rays of the bladder and kidneys are repeated at approximately 3-monthly intervals.

If the infection cannot be controlled by the means outlined, or if there is deterioration of the kidneys assessed by X-rays, the child is admitted to hospital for endoscopic examination. This examination often brings to light associated urological abnormalities with obstructive effects, e.g. urethral stenosis, urethral valves in boys or perhaps the presence of a calculus. Treatment of these lesions often improves the situation and the conservative regimen of 2-hourly expression and urinary antiseptics is again successful.

If, despite all the above measures, infection cannot be controlled and there is deterioration in the X-ray appearance of the urinary tract, a conduit is performed. This entails isolation of a loop of small intestine with its blood supply intact. The ureters are separated from the bladder and implanted into one end of the loop. The other end of the loop is brought out to the skin as an ileostomy.

Urine is constantly expressed through the stoma by the healthy, active bowel. A rubber bag is worn to collect the urine. It is kept in place with adhesives and a belt. There are 39 cases who require urological surveillance in the clinic, 18 males and 21 females. Eleven of these children have not responded to conservative management and conduits have been fashioned to provide free, low pressure drainage of the upper urinary tracts.