Urinary continence in open myelomeningocele

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SUMMARY  In a consecutive series of 200 neonates having undergone surgery for open myelomeningocele, 24 of the 106 survivors at 10 to 12 years of age proved to be continent of urine. These figures included 11 children who had considerable neurological involvement. 4 children did not acquire urinary control until after the age of 5 years. Therefore, we consider early urinary diversion in children with myelomeningocele, even in the presence of neurological deficit, to be indicated only for the prevention of progressive deterioration in renal function. Incontinence of urine itself is not an indication for urinary diversion in the first decade of life.

The chief handicaps of children with myelomeningocele are those related to hydrocephalus, to the function of the lower limbs, and to the urinary tract. The view is widely held that bladder control in spina bifida cystica is likely to be so poor that urinary diversion is needed in almost all cases. Indeed, children with myelomeningocele are thought to have such a poor prognosis with regard to bladder control that urinary diversion has been recommended for all children before 5 years of age. Whether or not this radical approach is justified can only be determined by a long-term study, and this report analyses bladder control in children born with open myelomeningocele at a time 10 to 12 years after closure of the lesion in the back.

Patients and methods

This series comprised an unselected group of 200 consecutive cases of open myelomeningocele operated on at the Children's Hospital, Sheffield, from 1962 to 1964, 194 of them being operated on in the first 36 hours of life. During that time, 2 other children were admitted to the hospital with open myelomeningocele but did not undergo operation because clinically they had sustained severe intracranial damage with probable ventricular haemorrhage and it was considered that they were unlikely to survive.

At the time of review between late 1974 and early 1975, 88 children were known to be dead, 6 were lost to detailed follow-up though known to be alive in the United Kingdom, and 106 children had survived for a minimum period of 10 years (see Table 1) about whom accurate information was available. 60% of the deaths occurred in the first 3 months of life and 90% during the first 2 years of life (Lister, 1971).

The total of 106 known survivors form the basis of this survey and have all been seen at the hospital at regular intervals over the past 10 to 12 years. Those with virtually no disability have been seen annually, and others more frequently. The level of the lesion was described in relation to the vertebrae involved and not to the neuromotor or neurosensory levels. The anatomical sites of the 106 lesions are given in Table 1.

The term 'continent' was used to indicate normal

<table>
<thead>
<tr>
<th>Site of lesion</th>
<th>No. of cases</th>
<th>No. continent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Female</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Lumbar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Female</td>
<td>12</td>
<td>4</td>
</tr>
<tr>
<td>Sacral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>8</td>
<td>3</td>
</tr>
<tr>
<td>Female</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>Thoracolumbar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Female</td>
<td>15</td>
<td>2</td>
</tr>
<tr>
<td>Lumbosacral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>18</td>
<td>3</td>
</tr>
<tr>
<td>Female</td>
<td>14</td>
<td>0</td>
</tr>
<tr>
<td>Thoracolumbosacral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Female</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>51</td>
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</tr>
<tr>
<td>Male</td>
<td>55</td>
<td>11</td>
</tr>
<tr>
<td>Female</td>
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</tbody>
</table>

Note: 24 of 106 have normal continence (22–23% of survivors)

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social bladder control by day and night, the ability to hold urine in the bladder over a couple of hours, and to keep normal underwear dry, but 4 children with occasional nocturnal enuresis and normal daytime control were included in the continent group. Those suffering from severe nocturnal enuresis even if they seemed to have good control during non-sleeping hours, and also those who were kept socially dry by means of manual expression of the bladder or by collecting apparatus such as a penile appliance, were classified as incontinent.

Results

According to the above criteria, 24 children had normal continence (23%) and 82 children were incontinent. However, among the 'incontinent', 46 were 'socially dry', 16 by the use of a penile appliance and 26 with the aid of urinary diversion. Of the remaining 4, one was mainly dry but occasionally wet during the day, one was wet only at night, one was maintained dry by bladder expression, and one was kept dry by bladder expression during the day but was wet at night. 36 children were frequently wet, sometimes in spite of apparatus designed to keep them dry; 3 boys were wet despite wearing a penile appliance and 4 girls and 2 boys did not have a satisfactory apparatus for maintaining dryness with ureteroleiostomies or ureterostomies. At the time of review none of the patients were being treated by bladder catheterization and 27 were wearing nappies. Very few of the patients in this series were kept socially dry by bladder expression, though this technique was being continued where indicated to ensure satisfactory emptying of the bladder.

Continence related to site of lesion. Table 1 gives the number of continent children according to the site of the lesion. The children with isolated thoracic, sacral, or lumbar lesions were more likely to be continent than those having more extensive lesions such as lumbosacral or thoracolumbosacral abnormalities. The proportion of girls and boys who were continent was approximately the same, 11 out of 55 girls, and 13 out of 51 boys, and it is noteworthy that thoracic lesions were more frequent in boys than in girls.

Loss of continence. Of those who were continent at the age of 5 years, only one lost control at a later date, a boy with a sacral myelomeningocele and anal stenosis who began to lose control at the age of 7 years. The residual volume of urine increased and he developed a number of urinary infections. At the age of 9 he had an external sphincterotomy and afterwards was kept dry with a penile appliance, and his urine became sterile. Although he can walk with below-knee calipers, he attends a special school on account of his urinary incontinence.

Delayed acquisition of continence. 4 children appeared to gain bladder control after the age of 5 years (see Table 2) but one of these children, a boy with a sacral lesion, had some nocturnal enuresis. All had a neurological deficit and could walk only with the aid of calipers. One girl with a thoracolumbar lesion also required the use of a Rollator.

<table>
<thead>
<tr>
<th>Site of lesion</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Lumbar</td>
<td>1 at 5 yr</td>
<td>0</td>
</tr>
<tr>
<td>Sacral</td>
<td>1 at 7 yr (E)</td>
<td>0</td>
</tr>
<tr>
<td>Thoracicolumbar</td>
<td>0</td>
<td>1 at 7 yr</td>
</tr>
<tr>
<td>Lumbosacral</td>
<td>1 at 7 yr</td>
<td>0</td>
</tr>
<tr>
<td>Thoracolumbosacral</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

E = a boy with enuresis.

Intravenous pyelography. All children had an intravenous pyelogram (IVP) during the neonatal period, which was repeated once or more before the age of 5 years. In the continent group, apart from some minor anatomical defects, all the urinary tract radiographs were normal.

Normal continence, lack of urinary symptoms, sterile urine on repeated culture, and a satisfactory IVP were accepted as evidence of normality in the urinary tract at the age of 5 years, and radiographic examination was not repeated in one boy and 5 girls after that age. Despite this general rule, 12 boys and 5 girls had further IVP films made between the ages of 5 and 12 years; none showed deterioration of the upper urinary tract. In addition, in one girl bilateral ureteric reflux was suspected and confirmed by micturating cystogram. She is the only girl in the continent group who had some nocturnal enuresis but had no other symptoms; she has never had urinary infection and is now being treated conservatively, though bilateral ureteric reimplantation has been considered.

Urinary tract infections. In 21 of the 24 continent children no evidence of urinary tract infection has been found in spite of repeated examinations by microscopy, routine biochemistry, and culture. One boy with occasional infection was treated with cotrimoxazole on a long-term basis; 2 girls have had one episode of urinary infection each, one being a girl with a thoracolumbar defect who achieved continence at the age of 7 years. None of these 3
children has been shown to have any abnormality of the upper urinary tract on IVP.

Associated hydrocephalus or neuromuscular involvement. Of the 24 continent children, 12 required a shunt for the treatment of hydrocephalus (later removed in 1 girl), and the other 12 have not shown a degree of hydrocephalus sufficient to merit a shunt. 13 children had only mild neurological involvement of the lower limbs, but 11 had considerable neurological involvement and 7 of these had severe kyphoscoliosis, including the 3 with lumbosacral lesions and the 2 with thoracolumbar lesions. If we exclude the 13 with mild neurological involvement from the total 106 cases, there were 93 patients with moderate or severe neurological involvement and among these 11 had normal urinary control (approximately 12%).

Discussion

Permanent urinary incontinence is a severe handicap and in the past has been regarded by some as one of the criteria for withholding immediate operative surgical treatment of the open lesion (Smith 1965; Matson, 1968). Therefore, it is important to assess the chances of acquiring bladder control in order to plan proper treatment.

Various authors (Burns, 1967; Eckstein, 1968; Stark, 1968; Mawdsley and Rickham, 1969; Lorber, 1971, 1972; Smith and Smith, 1973; Stark and Drummond, 1973; De Sanctis and Lattimer, 1974; Forrest, 1974; Levin, 1974; Negårđh et al., 1974) have assessed the chances of affected children gaining continence or reasonable control and quote figures of from 7% to 50%, the figures varying according to the type of lesion, the definition of continence, the period of follow-up, and possibly the optimism or otherwise of the author.

We have known of children acquiring control at the age of 11 or 12 years, and even know of one young man who only achieved control over micturition at 18 years when he started his training at medical school (not a case in this series). It is true that 4 children (3 boys, 1 girl) in this series who were incontinent at 5 to 6 years, and yet had normal upper urinary tracts, acquired control at a later date. Obviously, the small proportion of children acquiring control in some series may have been adversely influenced by a policy of very early urinary diversion before school age, and occasionally for ‘social reasons’ at the age of 2 or 3 years (Smith and Smith, 1973).

Urinary diversion itself is not without its problems and complications (Cook et al., 1968; Bakker and Cornil, 1972; Smith, 1972; Scott, 1973; De Sanctis and Lattimer, 1974; Woodburn, 1974), and in this series of 106 cases urinary diversion had been performed for social reasons alone before or at the age of 5 years in 8 girls. Of these, only one has been entirely free from problems and one is trouble free after revision of her stoma 2 years after the diversion. Of the remaining 6, 3 have inconvenience from encrustation of the loop, bleeding, and frequent infections, and 3 have additionally developed a degree of hydronephrosis, one with recurrent stones. This seems a high price to pay for ‘social dryness’.

Early diversion in boys is even more difficult to justify than in girls, because quite a number of boys who do not have normal continence can become socially acceptable by wearing a penile collecting appliance, the modern types giving satisfactory performance in older boys and young men.

The finding of nearly 23% of continent children at the age of 10 to 12 years in an unselected series of open myelomeningocele is a strong argument against early urinary diversion on surgical grounds alone, though diversion may be unavoidable on the grounds of upper tract deterioration. Admittedly, of the children with normal continence, 13 (54%) had only mild neurological involvement of the lower limbs, but in 11 (46%) of them there was considerable paralysis and these included 3 children (of a group of 22) with lumbosacral lesions and 2 children (of a group of 22) with thoracolumbar lesions. Therefore we now believe that even in a child with an extensive back lesion and major lower limb involvement, provided there is no progressive deterioration of the upper urinary tract, incontinence is not an indication for urinary diversion and other methods of management should be persisted with at least for the first decade of life.

In each child the state of the urinary tract should be considered carefully in an effort to detect the earliest signs of deterioration in the upper tracts, because it is this evidence which will be an indication for early surgical interference. Such surgery may include external sphincterotomy in boys (Thomas et al., 1973) or urethral dilatation or urethrotomy in girls, and though these measures cannot be guaranteed to achieve satisfactory emptying of the bladder they may prevent further deterioration for a time.

Lack of control of the bladder in males can be managed with a penile appliance in boys as young as 7 or 8 years of age, provided there is no evidence of bladder outlet obstruction. In girls, even those in whom it is thought that diversion may well be necessary, a trial with an indwelling balloon catheter in the bladder is well worthwhile (De Sanctis and Lattimer, 1974; Duthie and Stark, 1974; Forrest, 1974). Should this prove to be completely unsatis-
factory on account of irritation of the bladder, trauma to the mucosa, recurrent obstruction by debris, repeated infection which cannot be controlled by simple chemotherapy, or leakage of urine around the catheter, then the catheter can be abandoned without prejudice to alternative methods of management.

In a number of girls, not in this series, we have found bladder drainage by an indwelling catheter to be successful not only in keeping the child dry but also in reducing upper tract distension (Fig.). This may be a permanent method of management, or it may be used temporarily to delay diversion for a few years, or occasionally to give the child time to develop urinary control. We have not met serious problems of infection with indwelling catheters, though some authors (Rabinovitch, 1974) recommend intermittent catheterization.

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References


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