Spina bifida: do we have the right policies?

The enthusiastic acceptance of received dogma, especially when it is vigorously propounded, has been a common determinant of medical practice. The history of the management of neural tube defects in children illustrates this well, and examples may be drawn from our application of preventative measures, from our use of selection policies in the newborn, and from our allocation of resources to the management of older patients.

Prevention

The primary prevention\(^1\) of a disorder whose phenotype is determined by multiple genetic and environmental factors is unlikely to be straightforward. Historically, many ephemera in this field have been observed, advocated with enthusiasm, and discarded. Blight in potatoes consumed in pregnancy is perhaps the best known example. It is to be hoped that our current optimism concerning periconceptual vitamin supplements will not suffer the same fate. The onus, however, should be on us to establish the true worth of specific substances by clinical and laboratory research rather than our accepting unproved assertions and putting them widely into practice.

At the same time it is noteworthy that screening policies during pregnancy vary from region to region in the United Kingdom, with \(\alpha\) fetoprotein and ultrasound scanning being carried out in different ways and at different times. It is difficult to know how successful these policies are. In our experience failures of screening are still seen from time to time and it important that careful analysis of all such incidents be made.

Selection policies

There have been many changes in practice in the last two decades. Thus the emergency closure of myelocoeles, a procedure whose alleged principal purpose of preserving neurological function has never been substantiated by controlled studies, dominated neonatal management and determined future action for the great majority of affected children until the early 1970s.\(^2\) The resulting survival of many severely handicapped children led to the development of neonatal selection policies. Essentially, proponents of these argue that newborns with adverse criteria die while those without are likely to survive without major handicaps. Some care givers are discomforted by this approach, pointing out that many of the children who die have such potentially survivable conditions as infection, hydrocephalus, and incontinence.\(^3\) Even untreated, these children do not always die quickly. Gross\(^4\) describes a period of survival for totally untreated hydrocephalus in these circumstances of up to 137 days.

The paper from Cardiff\(^5\) in this issue of the Archives is therefore noteworthy. It is clear from this that not all infants with conventionally agreed adverse criteria die. Indeed, not only are there an appreciable number of survivors but also not all of these are grossly handicapped. Data from Liverpool support this concept. Table 1 shows the number of children in 1973–5 born with a myelocoele and to whom a selection policy was applied. These children were reviewed annually until recently. Table 2 indicates how many of these children survived.

As with the Cardiff series, the survivors of the 'selected out' group had more hospital admissions and a much longer period of time receiving inpatient treatment but at 5 years, assessment including measurement of intelligence showed that they were no worse overall than one would expect compared with children who had had closure at birth with a similar level of paralysis.

There are practical implications to these findings. We now reserve early primary closure for patients who, on established criteria,\(^6\) are likely to be minimally handicapped. This selection implies surgery within 24 hours of birth in order to minimise

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<th>Table 1</th>
<th>Number of myelocoeles in newborns for Mersey Region</th>
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<tr>
<td></td>
<td>1973</td>
</tr>
<tr>
<td>Operated ('selected in')</td>
<td>26</td>
</tr>
<tr>
<td>Unoperated ('selected out')</td>
<td>22</td>
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<th>Table 2</th>
<th>Five year survival of infants with myelocoele in Mersey Region</th>
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<tr>
<td></td>
<td>1973</td>
</tr>
<tr>
<td>Operated</td>
<td>22</td>
</tr>
<tr>
<td>Unoperated</td>
<td>8</td>
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the risk of infection. The disadvantage of such an emergency procedure is the difficulty of involving parents in speedy decision making when they often will wish to discuss all the long term implications at several interviews. To a degree, however, this is counterbalanced by subsequently regarding both 'selected in' and 'selected out' children in the same way. This implies that both groups, like all other handicapped children, merit continued evaluation and care.

So far as the 'selected out' children are concerned, we do not encourage a sedation policy and, as the Cardiff team points out, it is important that the parents are involved with the care of their baby. Some parents may reject their child but many will wish to take him home. The major concerns then become the size of the cystic lesion on the back and the presence and severity of hydrocephalus. It has been our practice in these babies to insert a shunt at 3 to 4 months of age when the back has epithelialised, and then carry out delayed primary back closure which makes nursing easier. Thereafter, their acute and chronic problems are managed using the same principles of relevant and comprehensive evaluation and care as would be applied to other children with handicaps.

Later childhood and adolescence

A proportion of children with spina bifida, whether 'selected in' or 'selected out' for surgery will continue to survive and need prolonged and expensive treatment. Judging from our present experience, this nettle has not been totally grasped and their care in school and especially later has been woefully underprovided. Very few of these will enter normal employment and many will need training well beyond school leaving age.7 Health facilities for adolescent and adult spina bifida patients in hospital and in the community are generally provided on an ad hoc basis,8 and a plea for appropriately organised care was made this year.9 Paediatricians should be reinforcing these pleas, for present overall provisions for older patients vary from poor to pathetic. Many in our experience gravitate back to the paediatric surgeon or physician and this is far from ideal.

Helping children with spina bifida and their families consumes resources and emotions. In prevention, management in infancy, and care in adolescence it may be that these can be more suitably allocated.

References


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