Long-term outcome in open spina bifida

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SUMMARY

Background: Doctors need reliable data on outcome in order to help parents faced with difficult decisions about termination of an affected pregnancy or treatment after birth.

Aim: To determine survival, health and lifestyle at the mean age of 30 years in a complete cohort of adults born with open spina bifida.

Design of study: Prospective cohort study.

Participants: Well-documented cohort of 117 consecutive cases of open spina bifida whose backs were closed non-selectively at birth between 1963 and 1971.

Method: Survivors (age range = 26 to 33 years) were surveyed by postal questionnaire and telephone interview. The main outcome measures were the health, independence and lifestyle of the survivors in terms of living in the community, driving a car and working in open employment.

Results: Ascertainment was 100%. Sixty (51%) had died, mainly the most disabled. Of the 57 survivors, 84% had a cerebrospinal fluid (CSF) shunt, 70% had an IQ of 80 or more, 37% lived independently in the community, 39% drove a car, 30% could walk more than 50 metres and 26% were in open employment. However one-third (19) still needed daily care, three were on respiratory support, two were blind, two had diabetes mellitus, and one was on dialysis. Mortality, disability and achievement reflected the neurological deficit that had been recorded in infancy in terms of sensory level. Attainment and independence were reduced in those who had needed revision of CSF shunt.

Conclusion: The survivors in this unselected cohort showed a wide range of outcome from apparent normality to very severe disability. This reflected both the extent of their original neurological deficit and events in the history of their CSF shunt.

Keywords: Open spina bifida; outcome; attainment; CSF shunt.

Introduction

In the 1960s, when this study started, babies with open spina bifida who owed their lives to the cerebrospinal fluid (CSF) shunt were not expected to survive into adulthood. Their future disability was unknown, as were its implications. Although early results of treatment led to optimistic forecasts, their physical and intellectual defects became more apparent as they grew older, and reviews became less encouraging.1-6 Data on long-term outcome of open spina bifida are vital for doctors helping parents with difficult decisions about termination of an affected pregnancy or treatment after birth. In the UK, most adults with spina bifida are cared for by general practitioners (GPs). The aims of this review were to record survival, health, lifestyle and attainments at the mean age of 30 years in a complete cohort of adults with spina bifida.

Method

Patients

In 1963 paediatricians in East Anglia agreed to refer all newborn babies with spina bifida to the Regional Neurosurgical Unit at Addenbrooke’s Hospital, Cambridge. Treatment was offered to all babies with open spina bifida without any attempt at selection, provided they were seen within a few hours of birth. Between 1963 and 1971, after a full neurological examination, 117 babies (50 male, 67 female) had their defects closed within 48 hours of birth. A CSF shunt was inserted for hydrocephalus when required.

Data collection

In 1997 all survivors were surveyed by confidential questionnaire and telephone interview. They were asked about their health, disability and attainments in terms of living independently, driving a car and working in open employment. For those who had died, causes of death were obtained from medical records and the Office of National Statistics. The study was approved by Cambridge Local Research Ethics Committee.

Statistical analysis

Patients had previously been classified into four groups according to sensory level to pin prick recorded in infancy.4 Those with intact sensation down to the knee (sensory level below L3) had a better short-term outcome than those with intact sensation no lower than the umbilicus (sensory level above T11). Mortality and measures of disability and attainment were compared in those with different sensory levels and CSF shunt histories using $\chi^2$ for trend.

Results

Ascertainment was 100%. Twenty-six (46%) of the 57 survivors responded to the questionnaire, and all survivors or a carer or relative were interviewed by telephone.
of the 57 survivors, nine had never had a shunt; of these, eight had little or no disability and a sensory level below L3. The remaining 48 had had a ventriculo-atrial shunt inserted. In 16 patients the shunt had never been revised. The other 32 patients had had a total of 113 revisions: for shunt insufficiency (mean = 4.5 revisions, range = 1 to 14). Elective revisions were carried out between the ages of two and 30 years (mean = 1.3 revisions, range = 1 to 3), and in 22 patients revisions were done only before the age of two years (mean = 1.3 revisions, range = 1 to 3).

**CSF shunts**

Of the 57 survivors, nine had never had a shunt; of these, eight had little or no disability and a sensory level below L3. The remaining 48 had had a ventriculo-atrial shunt inserted. In 16 patients the shunt had never been revised. The other 32 patients had had a total of 113 revisions: for shunt insufficiency (60%), infection (16%), detachment (13%), extrusion or leaking wound of back (5%), and unknown (6%). In ten patients revisions were done only before the age of two years (mean = 1.3 revisions, range = 1 to 3), and in 22 patients revisions were done before the age of two years (mean = 1.3 revisions, range = 1 to 3), and in 22 patients revisions were done only before the age of two years (mean = 1.3 revisions, range = 1 to 3).
Twenty-eight survivors (49%) had one or more attainments in terms of living independently in the community, driving a car (22 patients), or working in open employment (15 patients). Attainments were related to sensory level in infancy and to shunt history (Tables 1 and 3). Patients without a shunt or in whom the shunt was never revised were more likely to live independently, drive or work than those who needed revision, particularly after the age of two years when the cranial sutures had fused, rendering the intracranial contents more susceptible to pressure. Table 4 shows that late revisions of shunt (after the age of two years) were also associated with a birth head circumference greater than the 90th centile, a history of symptoms of raised intracranial pressure, visual defects, and need for daily care. Of the 11 survivors born with a birth head circumference greater than the 90th centile, nine needed revisions after the age of two years, ten had visual defects (of whom two were blind), seven had epilepsy, and five had an IQ lower than 80.

**Discussion**

**Principal findings**

By the mean age of 30 years, half the cohort had died and these were mainly the most disabled. One-third of the survivors lived independently, one-third needed some support and one-third needed daily care. Sensory level recorded in infancy predicted mortality, disability, and lifestyle in adulthood. Only about 10% of those who needed shunt revisions after the age of two years lived independently, drove a car, or worked in open employment.

**Strengths and weaknesses of the study**

The community basis of this study provides social as well as clinical data, enabling the realities of adulthood to be seen against the optimistic forecasts of the early years. Only 23 (40%) of the survivors were still attending hospital — mainly for urological care, pressure sores, or single items, such as surgical boots. Thus a hospital-based study would have given an incomplete perspective. As the patients grew older, the reduction in support, rehabilitation and encouragement from dedicated physiotherapists, parents and other carers revealed an outcome that was related to the patient’s own motivation in addition to the basic neurological deficit. A further strength of the study is that subjects were recruited consecutively without selection, and there was meticulous documentation of the clinical signs at birth. Patients who never require a CSF shunt, or in whom the open lesion proves at operation to be a simple meningocele, have a better outlook and their proportion in any cohort is crucial to data on outcome. In the present series, there were 11 such patients, of which four patients needed long-term anticonvulsants in the past, giving an overall incidence of epilepsy of 30%. Four patients needed regular analgesics for musculo-skeletal pain.

**Parenthood**

Seven women and one man had become parents. The man had minimal disability and no detectable sensory loss.

**Residence and dependency**

Twenty-one individuals (37%) lived independently in the community, ten of whom used wheelchairs. A further 17 (30%) were personally independent but had supervision and help when required. The remaining 19 (33%) needed help daily for dressing, shaving, toilet, or nursing care (mainly pressure sores). Thirteen of these still lived with their parents who were now aged 47 to 77 years; two women were in the care of their husbands; three were in residential establishments; and one was in sheltered accommodation with 24-hour attendance.

**Car drivers**

Thirty-one (54%) of the survivors had passed their driving test, but nine had discontinued driving because of lack of funds (four patients), or after an illness (three patients) or accident (two patients). Eleven were unfit to drive because of visual defects (three patients), epilepsy (three patients), or severe cognitive or perceptual defects (three patients). Lack of confidence, particularly in wheelchair users, was a potent reason for giving up driving or not attempting to drive in the first place.

**Employment**

Eleven men and four women were in open employment. All of them had an IQ higher than 80. Five did clerical work, two were teachers, two were engineers, two were unskilled manual workers, and the others were a business executive, accountant, van driver, and builder. Three were studying in addition to working full time. Four men and six women were in sheltered employment.

**Attainments**

Twenty-eight survivors (49%) had one or more attainments in terms of living completely independently in the community (21 patients), driving a car (22 patients), or working in open employment (15 patients). Attainments were related to sensory level in infancy and to shunt history (Tables 1 and 3). Patients without a shunt or in whom the shunt was never revised were more likely to live independently, drive or work than those who needed revision, particularly after the age of two years when the cranial sutures had fused, rendering the intracranial contents more susceptible to pressure. Table 4 shows that late revisions of shunt (after the age of two years) were also associated with a birth head circumference greater than the 90th centile, a history of symptoms of raised intracranial pressure, visual defects, and need for daily care. Of the 11 survivors born with a birth head circumference greater than the 90th centile, nine needed revisions after the age of two years, ten had visual defects (of whom two were blind), seven had epilepsy, and five had an IQ lower than 80.
whom ten survived to age 30 years. In contrast, those born with a birth head circumference above the 90th centile had a much worse outcome, which may have been related to the effect of prenatal raised intracranial pressure or cortical maldevelopment.9 The main limitation of the study is that the long follow-up implies treatments that have been superceded. Improvements in the diagnosis and management of neurological and renal problems have halved the mortality by the age of five years,10,11 but have less influence on long-term disability.

Comparison with other studies
Outcome in childhood of early operated spina bifida has been widely reported,1–5,11 but there are fewer studies of long-term outcome. A recent survey of a cohort of 118 adults aged 20 to 25 years with 24% mortality and 16% loss to follow-up, found continuing deterioration and a formidable number of neurosurgical and spinal operations.12 This is the only 30-year prospective study of open spina bifida with 100% ascertainment by the same independent observer.

Implications
These data may help health professionals who counsel parents of children with spina bifida. They show a range of possible outcomes in adulthood when parents may no longer be able or willing to look after their child. The profound effects on the family,13 and the implications of incontinence in the young adult must never be underestimated. Since, in this cohort, the most severely disabled died, modern treatment that reduces mortality may favour the survival of the more severely disabled.6 Those caring for patients with spina bifida need to know both their long-term potential and the limited benefits of treatment to focus on realistic goals.7

Table 2. Influence of sensory level and age on walking in 57 survivors with spina bifida.

<table>
<thead>
<tr>
<th>Sensory level in infancy</th>
<th>All survivors (n = 57)</th>
<th>Walkers at nine years (n = 32; 56%)</th>
<th>Walkers at 30 years (n = 17; 30%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Above T11</td>
<td>13</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>T11–L3</td>
<td>16</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>L4</td>
<td>8</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>L5–S2</td>
<td>6</td>
<td>6</td>
<td>5</td>
</tr>
<tr>
<td>No sensory loss</td>
<td>11</td>
<td>11</td>
<td>10</td>
</tr>
<tr>
<td>Asymmetrical loss</td>
<td>3</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

*Walkers* defined as able to walk more than 50 metres using aids if required. Survivors with lower sensory levels are more likely to be walkers — χ² for trend P<0.0001 for both age nine years and 30 years. Asymmetrical sensory loss excluded from the analysis.

Table 3. Lifestyle related to CSF shunt in 57 survivors at the mean age of 30 years.

<table>
<thead>
<tr>
<th>Living independently, n = 21 (37%)</th>
<th>No shunt (n = 9)</th>
<th>Shunt, a no revisions (n = 16)</th>
<th>Shunt, revised at age &lt;2 years (n = 10)</th>
<th>Shunt, revised at 2–30 years (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sheltered or in care</td>
<td>8</td>
<td>8</td>
<td>4</td>
<td>1¹</td>
</tr>
<tr>
<td>Driving a car, n = 22 (39%)</td>
<td>5</td>
<td>9</td>
<td>5</td>
<td>3²</td>
</tr>
<tr>
<td>Not driving</td>
<td>4</td>
<td>7</td>
<td>5</td>
<td>19</td>
</tr>
<tr>
<td>In open employment, n = 15 (26%)</td>
<td>3</td>
<td>6</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Sheltered employment or none</td>
<td>6</td>
<td>10</td>
<td>6</td>
<td>20</td>
</tr>
</tbody>
</table>

*Shunts (n = 48) were only inserted or revised in response to definite clinical need, such as symptoms or signs of raised intracranial pressure. χ² for trend: ¹P<0.0001; ²P<0.05.

Table 4. Features related to CSF shunt history in 57 patients with open spina bifida at the mean age of 30 years.

<table>
<thead>
<tr>
<th>No shunt (n = 9)</th>
<th>Shunt not revised (n = 16)</th>
<th>Shunt revised age &lt;2 yrs (n = 10)</th>
<th>Shunt revised age 2 yrs (n = 22)</th>
<th>χ² for trend</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth head circumference 90th centile, n = 11</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>9</td>
</tr>
<tr>
<td>History of symptoms of raised intracranial pressure, n = 25</td>
<td>0</td>
<td>1</td>
<td>5</td>
<td>19</td>
</tr>
<tr>
<td>Visual defects (mainly squint), n = 36</td>
<td>4</td>
<td>7</td>
<td>6</td>
<td>19</td>
</tr>
<tr>
<td>Daily care needed, n = 19</td>
<td>1</td>
<td>3</td>
<td>1</td>
<td>14</td>
</tr>
</tbody>
</table>

Implications
These data may help health professionals who counsel parents of children with spina bifida. They show a range of possible outcomes in adulthood when parents may no longer be able or willing to look after their child. The profound effects on the family,13 and the implications of incontinence in the young adult must never be underestimated. Since, in this cohort, the most severely disabled died, modern treatment that reduces mortality may favour the survival of the more severely disabled.6 Those caring for patients with spina bifida need to know both their long-term potential and the limited benefits of treatment to focus on realistic goals.7

The results have important practical implications for predicting long-term outcome in newborns. They suggest that babies with sensation below the knee (L3) are unlikely to be seriously disabled and could be achievers in adulthood. Babies who cry during a routine heel prick for the Guthrie test have a sensory level of S1 or below and are likely to remain community walkers at age 30. A similar response to pricking the saddle area (S2–4) forecasts the likelihood of bladder and bowel control.14

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Such simple tests should be routine, and may be particularly useful for management decisions in countries with less access to specialist investigation and treatment.

In the UK, the care of children with spina bifida depends mainly on paediatricians. Once they are adults, patients lose that comprehensive care.\textsuperscript{15} If they leave home they become separated from the support and care of parents whose experience of their condition is unique. The GP becomes the person most likely to be involved but may have limited experience of the particular problems presented by these patients. Chronic or intermittent shunt insufficiency has been misdiagnosed as sinusitis, dysphagia or painful neck, and referred to otolaryngologist, gastroenterologist or rheumatologist. Failure to recognise and treat shunt insufficiency promptly may be fatal, or may result in blindness or long-term dependency.\textsuperscript{8,16,17} Although the incidence of spina bifida is falling, survivors from the 1960s and 1970s will continue to require long-term support.\textsuperscript{15}

Conclusion
Survival in open spina bifida depends mainly on treatment. Disability and attainment depend on the severity of the original neurological deficit and on neurosurgical complications. A third of survivors to the age of 30 continue to need daily care.

References

Acknowledgements
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