ORIGINAL ARTICLE

Perineal sensation: an important predictor of long-term outcome in open spina bifida

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Objectives: To see if perineal sensation in infants with open spina bifida is associated with a better long-term outcome, particularly in terms of survival, renal-related deaths and incontinence.

Methods: We conducted a prospective cohort study on a complete cohort of 117 consecutive patients with open spina bifida, whose backs were closed non-selectively at birth between 1963 and 1971. A meticulous neurological examination in infancy showed that 33 (28%) of them had perineal sensation, defined as intact sensation to pinprick in at least one dermatome on one side in the saddle area (S2–4). Data recorded within 48 h of birth and during six reviews between 1972 and 2002 were used. Details of deaths were obtained from medical records and from the Office of National Statistics.

Results: By December 2005, 57% (67/117) of the cohort had died. There were 50 survivors with a mean age 38 years (range 35–41). More of those with perineal sensation survived than those without (23/33 v 27/84, $p < 0.001$). This difference was mainly caused by 19 renal deaths in those lacking perineal sensation. Crucially there were no renal-related deaths in those with perineal sensation (0/33 v 19/84, $p = 0.003$). Among the survivors, those with perineal sensation were more likely than the remainder to be continent of urine and faeces (10/23 v 1/27, $p < 0.001$ and 18/23 v 9/27, $p = 0.002$ respectively). They were also more likely to be able to walk at least 50 m (11/23 v 5/27, $p = 0.027$) and never to have had pressure sores (15/23 v 9/27, $p = 0.025$).

Conclusions: A simple assessment of perineal sensation in infancy predicts long-term outcome in terms of survival, renal prognosis and incontinence in open spina bifida.

Most people born with open spina bifida have a neuropathic bladder which may result in urinary incontinence, retention and infections, vesicoureteric reflux, renal damage and death.^{1–3} Studies of children with spina bifida show that those most severely affected, who have the most extensive neurological deficit, are most likely to die of urological complications.^{4–7} Continence is also related to neurological deficit,⁸ particularly in terms of sensory level.^{9–10} However, sensory testing is sometimes omitted from evaluation of urinary function and prognosis in open spina bifida because it can be difficult and occasionally confusing. By contrast, sensation in the perineum (sacral dermatomes 2, 3 or 4) is a more widely used clinical indicator—for example, in assessing possible spinal cord damage in spinal injury and prognosis for continence. There have been no prospective studies comparing long-term outcome over 40 years in infants born with open spina bifida with and without perineal sensation.

The Cambridge cohort comprises 117 consecutive cases of open spina bifida treated non-selectively from birth between 1963 and 1971 at Addenbrooke's Hospital, Cambridge, UK.⁵ Before closure of the back, sensation to pinprick was routinely tested and recorded from birth as part of a meticulous neurological examination. In 86 (81%) cases, hydrocephalus was controlled by insertion of a cerebrospinal fluid shunt. A further 11 patients died before a shunt could be inserted. The cohort has been prospectively followed up with reviews at mean ages of 4, 9, 18, 25, 30 and 35 years without loss to follow-up.^{5–9–11–14} The first two reviews were done at home and at school; the next two during hospital visits and at school; and the most recent ones in the community by a postal questionnaire backed up by a telephone call to the patient or carer. On the basis of previous experience,^{5–7} we hypothesised that cases with the clinical findings of perineal sensation in infancy

would have a better long-term outcome, particularly in terms of renal-related deaths and continence.

METHODS

Sensation was defined at birth and subsequently using a sterile needle starting from the most distal dermatomes. A positive response was recorded only if the child cried or made a facial grimace, indicating sensitivity to pinprick. In the absence of these, the response was recorded as negative even if there was reflex movement of the legs or an anal reflex.^{5–15} A positive response to pinprick anywhere in the perineal dermatomes, even if on one side only, was recorded as evidence of perineal sensation.

We used data from the reviews of the cohort^{5–9–11–14} backed by information on date and cause of death to December 2005 provided by The Office of National Statistics (reference MR564). In 36 cases (54%) autopsy reports were available. We drew Kaplan–Meier curves and compared survival and causes of death in those with and without perineal sensation using log rank tests. We compared urinary and faecal continence in survivors with and without perineal sensation using Fisher's exact test. As walking and pressure sores are related to neurological deficit,^{1–6–9} we also looked at these outcomes in the two groups.

RESULTS

A total of 67 (57%) of the cohort ($n = 117$) had died, a fifth before their first birthday and a third by age 5 years. Thereafter the death rate remained constant, with about 10% of the remainder dying every 10 years. There were 50 survivors (mean age 38 years, range 35–41) in 2005.

When the 117 babies were assessed in infancy, 84 (72%) were found to have no sensation to pinprick below the level of the

Figure 1 Survival related to perineal sensation in infancy in 117 consecutive cases of open spina bifida.

spinal lesion and no sensation in the perineum. The remaining 33 had some sensation in the perineum (sacral dermatomes 2–4). Of these, 16 had an incomplete neurological lesion with an island of sensory sparing in the perineum. Three had an asymmetrical lesion with perineal sensation on one side. In the remaining 14, there was no sensory loss in the perineum, legs or trunk. Apart from three exceptions, this last group tended to be less disabled.

Figure 1 and table 1 show better survival in those with perineal sensation. Of 33 born with perineal sensation, 23 (70%) survived to the mean age of 38 years compared with 27 (32%) of the 84 without perineal sensation ($p < 0.001$). The improved survival in those with perineal sensation was similar in those with no sensory loss and those with incomplete sensory loss: 64% (9/14) v 74% (14/19).

Figure 2 and table 1 show that the main reason for the poorer survival in those with no perineal sensation was due to renal related deaths. Of the total of 67 deaths, 19 (28%) were due to urological causes. All these deaths occurred in those lacking perineal sensation: nine before the age of 5 years, five at ages 5–20 years and five at ages 21–40 years. By contrast, there were no renal-related deaths in those with perineal sensation.

Figure 3 shows deaths due to non-renal causes in those with and without perineal sensation. From about the age of 5 years the lines run roughly parallel, suggesting a similar rate of deaths in both groups owing to other causes. Figure 4 shows that perineal sensation and renal deaths were also related to neurological deficit in terms of sensory level.

Figure 2 Renal deaths related to perineal sensation in infancy in 117 consecutive cases of open spina bifida.

Ten of the 23 survivors with perineal sensation were continent of urine without the use of aids or appliances (table 1). Of the remainder, six used intermittent catheterisation, four used pads, two had an indwelling catheter and one had had a urinary diversion. Of the survivors with no perineal sensation, one was continent of urine, eight used intermittent catheterisation, six had an indwelling catheter, five had a urinary diversion, two used a penile sheath and five used only pads. (Many used pads in addition to another method.) Those with perineal sensation were also more likely than the remainder to be continent of faeces, were able to walk at least 50 m and had never had sores. Three of the 23 male survivors have become fathers. Two, both with perineal sensation, conceived naturally. In the third patient without perineal sensation conception was by intracytoplasmic sperm injection.

DISCUSSION

This is the first study to conclusively show that perineal sensation in infancy predicts long-term survival, renal prognosis, and urinary and faecal continence in open spina bifida. The difference in survival in those with and without perineal sensation was mainly because of death from renal complications, and these deaths continued to occur throughout adulthood. Crucially, there were no renal deaths in those with perineal sensation.

Perineal sensory loss with associated bladder and urethral anaesthesia has practical implications. The patient may not be aware of a full bladder, nor may have the urge to empty it; an ordinary midstream urine specimen may be an impossible request. Acute urinary retention, cystitis or bladder calculus is unlikely to be accompanied by pain or dysuria. In addition, she or he may fail to notice skin damage to the anaesthetic perineum or penis, and self-catheterisation may be difficult, especially in females,² and more likely to cause trauma to the bladder or urethra. The protective function of sensibility to pain is an invaluable asset. It can be shown in infancy.⁵ When found in the perineum of babies with open spina bifida, it forecasts a better long-term outcome.

Strengths and weaknesses of the study

The strengths of this study are the long duration and 100% follow-up backed by meticulous recording of clinical findings in infancy. However, owing to the historical nature of the cohort, data on urodynamics and renal function are incomplete. In addition, improvements in the diagnosis and management of renal and neurological problems since these children were born have halved mortality by the age of 5 years.^{3 16} Finally, these results may not apply to different populations or ethnic groups, or to countries with very different forms of care.

Figure 3 Non-renal deaths related to perineal sensation in infancy in 117 consecutive cases of open spina bifida.

	Sensory level in infancy			
	<L3	L3–T11	>T11	Asymmetrical

Whole cohort	38	32	42	5
Perineal sensation*	24	6	0	3
Renal deaths*	0	4	15	0

Figure 4 Neurological deficit in terms of sensory level related to perineal sensation and renal deaths in 117 cases of open spina bifida. * χ^2 test for trend $p < 0.001$ (asymmetrical sensory level excluded from the analysis).

Table 1 Influence of perineal sensation in infancy on outcome at the mean age of 38 years in a complete cohort of 117 unselected cases of open spina bifida

	Perineal sensation n (%)	No perineal sensation n (%)	Relative risk (95% CI)
Whole cohort, n = 117	33	84	
Died, n = 67	10 (30)	57 (68)	0.45 (0.26 to 0.76)
Renal deaths, n = 19	0 (0)	19 (23)	0.064 (0.004 to 1.032)
Survivors in 2005, n = 50	23	27	
Urinary continence without aids, n = 11	10 (43)	1 (4)	11.74 (1.62 to 84.94)
Faecal continence, *n = 27	18 (78)	9 (33)	2.35 (1.32 to 4.17)
Walks ≥ 50 m, n = 16	11 (48)	5 (19)	2.58 (1.05 to 6.34)
Never had pressure sores, n = 24	15 (65)	9 (33)	1.96 (1.06 to 3.61)

*Faecal continence when clinically examined at school. (Later it was difficult to obtain reliable answers to this sensitive question. However, in six survivors, none of whom have perineal sensation, faecal incontinence remains a major problem. In those with perineal sensation, it is more easily controlled.)

Comparison with other studies

There have been few long-term prospective studies of open spina bifida. Lorber followed 270 consecutive, unselected patients with spina bifida to the age of 9 years,⁴ of whom 97 (36%) died. However, renal deaths (n = 13) occurred only in those with adverse prognostic criteria such as thoracolumbar lesions, severe paraplegia, gross hydrocephalus and kyphosis.^{5–7} This is similar to our findings. More recently, a prospective study from the USA of 118 cases treated non-selectively with 16% loss to follow-up found that mortality (24%) continued to rise in young adulthood and late deterioration was common.¹⁷ Of the survivors aged 20–25 years, only 15% were completely continent of urine without aids.

Most studies looking at urological outcome have been cross-sectional. A community-based study of 527 Nordic children aged 4–18 years with spina bifida found that only 8% had normal urinary control and 22% normal bowel control.¹⁸ Parents said urinary and faecal incontinence was a major

problem for a third of children. Stark suggested that neurological level was a useful predictor of urinary continence in children with spina bifida.⁸ As in our series, he observed that asymmetrical lesions with sparing of one leg usually implied normal bladder function. (This may help to explain the link between perineal sensation and walking found in our cohort, both of which are more common in those with low lesions.¹⁹) Although he focused mainly on motor level, he also noted the importance of sensation for urinary continence. More recently, the Aspine study, including 142 Dutch 16–25-year olds with open spina bifida, found that those with a sensory level below L5 were more likely than those with higher lesions to be continent of urine and faeces.¹⁰ In line with a study of British patients with spina bifida aged 9–48 years,²⁰ about a quarter of responders were reportedly clean and dry, which is similar to

What is already known on this topic

- Most people with open spina bifida have a neuropathic bladder which can lead to urinary incontinence, renal damage and death.
- These risks are greater in those with a more severe neurological deficit, particularly in terms of sensory loss.

What this study adds

- Simple assessment of perineal sensation in infancy predicts long-term outcome in terms of survival, renal prognosis and continence in open spina bifida. The only equipment needed is a sterile needle.
- Although the results may not be fully generalisable, this clinical finding may be particularly useful in countries with less access to sophisticated technology and where the incidence of spina bifida remains high.

our results. Improvements of continence rates with increasing age may be partly because of better survival in those less severely affected, who are also more likely to have perineal sensation. Higher rates of "social" continence have been noted.^{16 17}

Implications

Sensation is crucial to survival and function in open spina bifida and should not be ignored. A baby's single vocal response to pinprick in the perineum may provide the essential information to predict long-term outcome. It may be used to give parents of a baby with spina bifida some indication of likely survival and continence.¹ The only equipment needed is a sterile needle. This clinical finding may be particularly useful to guide decisions in countries with limited resources and less access to sophisticated technology, and where spina bifida may be relatively common.²¹

Perineal sensation may also be simpler to assess in infancy than at sensory level.^{5 15} Infants with perineal sensation have a better long-term outlook and are more likely to become continent. They are also more likely to be able to continue walking in adulthood if they have sensation down to the knees (L3).^{9 12} By contrast, those without perineal sensation will require more frequent urological monitoring, are unlikely to have physiological continence and are at continuing risk of renal-related death throughout adulthood. Finally, this unselected cohort with complete follow-up for nearly 40 years provides a baseline against which to compare modern management. However, despite the diversity of treatment over time,¹² the one thing that remains constant is the basic neurological deficit and this is the most important predictor of long-term prognosis.¹⁴

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