Outcome in people with open spina bifida at age 35: prospective community based cohort study
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The introduction of the cerebrospinal fluid shunt led to a fourfold increase in survival of babies with open spina bifida in the United Kingdom. In 1963 a prospective independent review was set up to record the results and implications of the new treatment. Such data are crucial to the dilemmas associated with termination of affected pregnancies or treatment at birth. We investigated survival, disability, health, and lifestyles in a complete cohort of adults with spina bifida.

Participants, methods, and results

Between 1963 and 1971, 117 babies had their backs closed at Addenbrooke’s Hospital, Cambridge, without any attempt at selection. Before closure of the back each baby had a full neurological examination. When necessary, hydrocephalus was controlled by the insertion of a ventriculoatrial shunt. In spring 2002 we reviewed the cohort by confidential postal questionnaire backed by a telephone call to the patient or carer. The Office for National Statistics provided information on deaths.

Ascertainment was 100%. Sixty three (54%) had died, mainly the most affected. Causes of death were cardiorespiratory (19) or renal (18) failure, hydrocephalus (10), central nervous system infection (10), convulsions (2), inhaled vomit (2), sudden infant death (1), and thrombocytopenic purpura (1). The mean age of the survivors was 35 years (range 32-38). The male:female ratio was 1:1.3, the same as at birth. Of the

What is already known on this topic

Around 11 000 people receive electroconvulsive therapy in England annually

Controversy exists as to whether treatment is beneficial and whether patients are satisfied with it

Patients’ views have never been systematically reviewed

What this study adds

At least one third of patients report significant memory loss after treatment

Routine neuropsychological tests to assess memory do not address the types of memory loss reported by patients

Reported patient satisfaction with electroconvulsive therapy depends on the methods used to elicit a response

Conclusion

Although clinical trials concluded that electroconvulsive therapy is an effective treatment, measures of efficacy did not take into account all the factors that may lead patients to perceive it as beneficial or otherwise. Studies of treatment are needed that are able to investigate a range of outcomes valued by patients, including factors that impact on effectiveness and satisfaction. Also important is loss of autobiographical memory, which is widely described but insufficiently systematically investigated.

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Comment

The outcome in this complete and unselected cohort ranged from apparent normality to severe disability. This reflected both the neurological deficit in terms of sensory level in infancy and the adverse events in the history of the shunt.\(^ 5\) Over one third of survivors continued to need daily care.

The community basis provides a fuller perspective than hospital based studies. Only a third of the survivors were still attending a hospital. Most were in the care of general practitioners, who had to manage problems associated with incontinence, pressure sores, sepsis, epilepsy, urinary and respiratory infections, hypotension, and obesity in addition to psychological distress or backache in the carer. Doctors need to know that headache, neck ache, drowsiness, deterioration in vision, or new eye signs may indicate shunt insufficiently, which requires prompt intervention to prevent serious long term consequences.\(^ 7\) The continuing needs of the large number of adult patients surviving from the era of non-selective treatment will have to be dealt with for many years. Given the limited benefits of treatment, the data we have gathered from this cohort provide those involved with counselling families affected by spina bifida with the clinical evidence to help them make informed decisions.

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54 survivors, 46 had a cerebrospinal fluid shunt, 39 had an IQ \( \geq 80 \), 16 could walk 50 metres or more with or without aids, and only 11 were fully continent. Thirty had had pressure sores, and 30 were overweight. Mortality and disability were related to neurological deficit (figure).

In terms of lifestyle, 22 survivors lived independently in the community, though seven depended on a wheelchair. They managed their own lives including transport, continence care, pressure areas, and all medical needs. Twelve lived in sheltered accommodation, where help was available if required, and 20 needed daily help, mainly from a parent (now aged 52-77) or partner or from social services. All these 20 were severely disabled: two were blind after shunt dysfunction, and two were on respiratory support. However 20 of the survivors drove cars, although a further nine had given up driving. Thirteen worked in open employment, five of them in wheelchairs. Seven women and two men had had a total of 13 children, none of whom had visible spina bifida.

One hundred years ago

A vaccination compliment

The wife of Major-General Chambers, of I.I. Trinity Gardens, Folkestone, recently paid the public vaccinator of Folkestone one of those delicate compliments of which only well-to-do women are capable. She apparently regards vaccination as too important an operation to be entrusted to an ordinary medical man, so, when she felt that she and her two daughters were in need of it, she selected one who may be regarded as a vaccination specialist. Many quite ordinary people, with whom we would not for a moment class the wife of a major-general, think and do the same, and the supreme delicacy of Mrs. Chambers’s conduct lies in the fact that, instead of being content to pay the public vaccinator a compliment in the surgery where he sees his official patients, she made it a direct and personal one by seeking him out with her two daughters in his private consulting room. Unfortunately, however, after this she rather spoiled the thing by refusing to pay his fees, and the intervention of another of H. M. officers, in the shape of the county court judge, had to be invoked before she rounded off her compliment by the noble fee of half a crown a head. Half a guinea was the fee that had been suggested, and it really seems more appropriate. Medical men are too modest a race really to enjoy the compliments so frequently paid by the public to their philanthropic instincts, and have too much artistic taste not to see that “billy” compliments like this should, if painted at all, be gilt with the finest gold. (BMJ 1905;i:1227).