

Oral presentation

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Expectation of life and unexpected death in open spina bifida: 40 year complete, non-selective longitudinal cohort study

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Background

There are few data on long term survival in open spina bifida. Predictions based on hospital studies with relatively short follow up may be over optimistic and omit unexpected deaths occurring in the community. We investigated survival, causes of death, and lifestyle in a complete cohort of open spina bifida at the mean age of 40 years.

Materials and methods

Participants comprised a well documented cohort of 117 consecutive cases of open spina bifida whose backs were closed non-selectively within 48 hours of birth between 1963 and 1971 at Addenbrooke's Hospital, Cambridge, UK. In 2007, all survivors were surveyed by postal questionnaire backed up by telephone interview with patient or carer. Details of deaths were obtained from the Office for National Statistics, medical records and autopsy reports.

Results

One in three (40/117) died before the age of 5 years. A further 26% (31/117) died over the next 35 years, over 10 times the national average. Half the deaths (16/31) after the age of 5 were sudden and unexpected. All occurred in the community and were followed by a coroner's autopsy. The most frequent causes of these unexpected deaths were epilepsy, pulmonary embolus, acute hydrocephalus and acute renal sepsis. In terms of neurological deficit only 17% (7/42) of those born with a high sensory level above

T11 survived compared with 61% (23/38) of those with a low sensory level below L3 ($p = 0.001$).

The mean age of the 46 (39%) survivors was 40 years (range 37 to 43). Fourteen (30%) could walk >50 metres, 37 (80%) had an IQ ≥ 80 , 38 (83%) had a cerebrospinal fluid shunt and 9 (20%) were continent of urine and faeces without pads or appliances. Fifteen (33%) worked in open employment, 21 (46%) drove a car and 14 (30%) lived independently. However 16 (35%) needed daily care.

Conclusion

This study indicates 'survival of the fittest' since most of those dying were severely affected. Doctors and care planners need to be aware that contrary to previous suggestions, there is a continuing high mortality throughout adult life, and many of the deaths are unexpected.