

ORIGINAL ARTICLE

Life expectancy in open spina bifida

Robert M. Shavelle  | Matthew K. Paige  | Jordan C. Brooks | David J. Strauss

Life Expectancy Project, San Francisco, CA, USA

Correspondence

Robert Shavelle, Life Expectancy Project, 1439 – 17th Avenue, San Francisco, CA 94122-3402, USA.

Email: Shavelle@LifeExpectancy.org

Abstract

Aim: To estimate life expectancies for individuals with open spina bifida, stratified by age, sex, functional ability, and specific impairments.**Method:** In the present study, extensive data from 1659 persons in California (1986–2019) were analysed using standard methods. Empirical mortality rates were calculated, and rates were also derived for various cohorts using logistic regression. Life tables were then constructed, from which life expectancies were obtained.**Results:** Survival varied significantly by ambulatory and feeding ability, and by bowel/bladder continence. For example, at age 5 years the life expectancy was 27 additional years for males in the most severely impaired group and 65 years in the least severely impaired, compared with 70 in the general population. There was a modest secular trend—mortality decreasing by roughly 1% per year—which is accounted for in the life expectancy calculations.**Interpretation:** Life expectancy in open spina bifida varies by age, sex, severity of disability, and bowel/bladder continence. Survival has improved over the past 30 years.

The survival of various birth cohorts with open spina bifida has been studied extensively.^{1–7} The subsequent prognosis for adults who had survived to a given age has also been documented.^{8–12} Such series have involved relatively small sample sizes, and have not reported results simultaneously stratified by multiple factors such as age, sex, and severity of impairment.

Spina bifida is commonly referred to as a ‘snowflake condition’ whereby individuals are affected differently depending on the size and location of the spinal lesion.¹³ Indeed, survival has been shown to vary by bony and sensory level, and some limited survival information stratified by such factors has been published.^{11,12} In addition, a study comparing survival in the USA and UK found that gross motor function was a key predictor of survival.⁵

Life expectancy (i.e. the average survival time amongst a group of similar persons) is valuable both as a summary measure of survival and health, and as a factor to consider when planning for the care of patients. Prior studies of spina bifida survival have reported survival probabilities over varying follow-up periods. To our knowledge, life expectancies have not been reported.

Using a large longitudinal database of patients with spina bifida in California, we examined the effect of functional abilities (walking, feeding) and continence on survival. Our

primary aim was to calculate life expectancies stratified by age, sex, functional group (ability/disability), and bowel/bladder continence (henceforth together referred to generally as impairments). We compared these with the general population values, and also with those of persons who have similar impairments instead due to an acquired spinal cord injury (SCI). We also examined whether, these factors being equal, long-term survival has improved over the past 30 years.

METHOD

Data from the California Department of Developmental Services (DDS) have been previously described in detail,¹⁴ and the present study is an update and expansion of prior work.⁵ Ethical approval was given by the authors' institutional review boards. Written informed consent was not required. The current cohort comprises 22 102 person-years from 1659 patients with open spina bifida who received DDS services between January 1986 and December 2019. We restricted to years before 2020 to avoid irregularities that may have arisen during the COVID-19 pandemic period. There were 422 deaths during the period. The DDS provides medical services, therapies, board and care, and respite services

Abbreviations: DDS, Department of Developmental Services; SCI, spinal cord injury.

to all persons (children and adults) in the State of California who have significant developmental disabilities. Those who are only mildly affected may not qualify for or seek such services, and thus may not be represented in the present sample.

Information was collected on the Client Development Evaluation Report,¹⁵ an instrument that contains over 200 psychological, medical, functional, behavioral, and cognitive items, and is completed approximately annually for all active DDS clients. Bony level (cervical, thoracic, or lumbosacral) and the presence of hydrocephalus were available through the spina bifida International Classification of Diseases, 9th Revision (ICD-9) or International Classification of Diseases, 10th Revision (ICD-10) codes.

Patients with spina bifida were identified by selecting the first Client Development Evaluation Report evaluation in the study period with ICD-9 code 741 or ICD-10 code Q05 as the etiology of developmental disability. Persons with spina bifida occulta (ICD-9: 756.17, ICD-10: Q760) were excluded. Mortality information was obtained from both the DDS and the California Department of Health Services, to which all deaths in the state are reported.

We computed empirical mortality rates (deaths divided by exposure time) and the associated life expectancies, stratified by decennial ages, for two walking groups (cannot walk, even with support vs walks with support, unsteadily alone for up to 10 feet, or independently alone at least 20 feet). Rates for ages 80 years and older were based on proportional life expectancy¹⁶ because only two patients had survived to this age. We used these rates to construct life tables¹⁷ from which we obtained life expectancies. These empirical rates and the resulting life expectancies provide a reference point for the results of the fitted survival model described below.

Survival analyses were performed principally using the pooled repeated observations method for analysis. In this method the unit of observation is a person-year. With each person-year we associated (1) a binary outcome variable indicating whether the person survived or died in that year, and (2) a set of explanatory variables, such as age, sex, and ambulatory ability. Logistic regression survival analysis was used to relate the outcome variable (survived/died) to the explanatory variables,²¹ as has previously been done in many similar applications.^{5,18–20}

After preliminary work, we ultimately modeled survival based on age, sex, calendar year, ambulation, feeding ability, and bowel/bladder control. In the 'Results', we note the effects of some lesser factors, and in the 'Discussion' we address the issue of possible changes with age in the values of patient covariates. Because we found high mortality at ages 0 to 3 years (see also, for example, Oakeshott et al.^{10,12}) and relative stability of function at ages 5 years and older (details below), we chose here to work with data on persons aged 5 years and older. This excludes children who died before the age of 5 years, who were likely to have been more medically compromised.

The four motor function groups based on ambulation and feeding ability were: (1) cannot walk or self-feed: those who (a) could not walk, even with handheld support (e.g. cane/

What this paper adds

- Life expectancies, rather than merely survival probabilities, were calculated.
- Survival varied significantly by age, sex, ambulatory and feeding ability, and bowel/bladder control.
- There was improvement in survival from 1986 to 2019.

crutch/walker), and (b) could not self-feed for most of their nutrition; (2) cannot walk, can self-feed: those who (a) could not walk, even with support, though (b) could self-feed for most of their nutrition; (3) some walking: those who could walk with handheld support or unsteadily alone at least 10 feet; (4) walks well independently: those who could walk well alone at least 20 feet, and balance well.

The three groups based on bowel/bladder control were: (1) no control of bowel or bladder; (2) some control, though not complete; (3) complete control of bowel and bladder. For the final life expectancy computations, the latter two bowel/bladder groups were combined for simplicity. We return to this issue in the 'Results' and 'Discussion'.

Logistic regression provides the odds of mortality in the groups, other factors (including age and sex) being equal. This method is closely related to the standard Cox proportional hazards regression model and Poisson regression.²² We used the fitted model to calculate age- and sex-specific mortality rates for persons in each group, keyed to reflect the most recent calendar year (2019). We then used these rates to construct life tables,¹⁷ from which the life expectancies were obtained.

We also report the corresponding life expectancies in the general population,¹⁷ and of similarly affected patients whose impairments resulted instead from a childhood- or adult-onset traumatic SCI.²⁰ The two SCI comparison groups are (1) persons with paraplegia, who generally cannot walk on their own, though retain full upper body function, and (2) those with an American Spinal Injury Association Impairment Scale Grade D injury, who generally require a cane/crutch to ambulate, some of whom have bowel/bladder dysfunction. Both comparison groups are of interest: the general population because it documents the reduction in life expectancy due to spina bifida, and SCI because it provides an external reference indicating the survival effect of disabilities and bowel/bladder control.

RESULTS

Demographic, clinical, and functional information at first evaluation is given in [Table 1](#), stratified by three levels of ambulation. The 1659 patients, of whom 50% were male, were first assessed at an average age of 13 years and followed for

TABLE 1 Demographics and functional information at first evaluation.

Variable	Value	Ambulatory ability			
		Cannot walk <i>n</i> = 1026 (62%)	Some walking ^a <i>n</i> = 363 (22%)	Walks well <i>n</i> = 270 (16%)	All <i>n</i> = 1659 (100%)
Age (years)	Mean (SD)	12 (11)	13 (11)	16 (13)	13 (12)
	0–4	45	32	24	38
	5–9	11	18	15	13
	10–19	24	25	31	25
	20–29	13	15	17	14
	30–39	6	6	6	6
	40–49	2	4	3	2
	50+	1	1	3	1
Sex	Male	47	51	57	50
	Female	53	49	43	50
Spina bifida region	Cervical	1	1	1	1
	Thoracic	5	1	0	3
	Lumbosacral	15	17	16	15
	Missing	79	81	82	80
Feeding tube	Yes	8	4	3	6
	No	92	96	97	94
Hydrocephalus	Yes	66	61	43	61
	No	20	28	31	24
	Missing	14	11	25	15
Epilepsy	Yes	22	22	20	22
	No	63	64	52	61
	Missing	15	14	28	17
Intellectual disability	None	29	39	29	31
	Mild	34	38	47	37
	Moderate	9	8	15	10
	Severe	6	6	2	5
	Profound	5	2	1	3
	Missing	17	7	7	13
Hand usage ^b	None	8	1	0	5
	Raking motion	15	5	1	10
	Uses thumb	14	10	5	12
	Uses fingers	63	84	94	73
Crawling/standing ^b	Does not crawl	32	2	0	20
	Crawls, creeps, or scoots	37	5	1	24
	Pulls to stand	5	5	0	4
	Stands with support	7	35	4	12
	Stands alone but is unstable	1	18	2	5
	Stands well alone	0	17	60	14
	Missing	18	17	33	20
Wheelchair use ^b	Cannot propel	18	9	2	14
	Assists	8	10	2	7
	Moves with difficulty	14	11	2	12

(Continues)

TABLE 1 (Continued)

Variable	Value	Ambulatory ability			
		Cannot walk <i>n</i> = 1026 (62%)	Some walking ^a <i>n</i> = 363 (22%)	Walks well <i>n</i> = 270 (16%)	All <i>n</i> = 1659 (100%)
Eating ^b	Moves smoothly	35	31	33	34
	Missing	25	39	61	34
	Cannot feed self	20	3	1	14
	Attempts to self-feed	5	4	1	4
	Finger-feeds without assistance	7	5	5	6
	Feeds using utensils, with spillage	17	19	17	17
Bowel/bladder ^b	Feeds without spillage	50	68	75	58
	None	79	64	42	70
	Some	11	19	23	15
	Full	5	16	34	12
Dressing ^b	Missing	4	1	1	3
	None	30	11	4	22
	Cooperates	32	29	23	30
	Puts on some clothes	17	22	18	18
	Puts on all; cannot tie shoes	6	15	17	10
	Dresses self completely	10	21	37	17
Word usage ^b	Missing	4	1	0	3
	None	27	10	6	20
	Simple words	26	31	29	28
	Complex words	19	24	26	22
	Broad vocabulary	27	35	38	31

All figures are column percentages except sample sizes and mean age (SD).

^aWalks with handheld support (e.g. cane/crutch/walker) or unsteadily alone at least 10 feet.

^bShown here are abbreviated descriptions of the variables and their levels. Details are given elsewhere¹⁵

an average of 13 years. Overall, 25% of the exposure time was to patients age 30 years and over. Approximately 38% of the group were ambulatory (22% with handheld support or unsteadily without, and 16% who walked well unsupported), while 62% were not.

The percentage who had no bowel or bladder control varied by ambulation: 79% of those who could not walk were impaired versus 64% in those who walked with limitations, and 42% of those who walked well alone. Data on the region of impairment (i.e. bony level) were largely missing (80%). In the non-missing data, 5% were cervical, 16% thoracic, and 79% lumbosacral.

The lowest functioning group (cannot walk or self-feed; 13% of the population) was also more severely impaired in other respects. Notably, we found that 31% required a feeding tube compared with only 3% in the other groups combined, 79% were non-verbal compared with 11%, and 44% had severe or profound intellectual impairment compared with 6%. These differences were each statistically significant ($p < 0.05$).

Table 2 shows the empirical mortality rates by age for two crude walking groups (cannot walk; can walk either with support or alone), along with the resulting life expectancies. As expected, mortality varied considerably by age and walking ability. The rates at ages 60 years and above were 6 to 10 times those at ages 5 to 9 years, and rates at all ages were 2 to 4 times higher for those who could not walk compared with those who could. The resulting crude empirical life expectancies varied accordingly: at age 5 years it was 56 years in those who could walk with support or alone, while only 35 years in those who could not.

The final logistic regression survival model is given in Table 3. The degree of functional impairment in walking, feeding, and continence was the most important factor. For example, mortality in those who could not walk or self-feed was 7.13 times that of those who could walk well unsupported ($p < 0.001$), and those with no bowel/bladder control had 1.54 times the risk of those with at least some control ($p = 0.002$). Males had 7% higher risk than females, other factors being equal. While this result was not

TABLE 2 Empirical deaths, person-years of exposure, mortality rates, standard errors of the rates, and resulting life expectancies.

Age	Cannot walk					Can walk with support or independently					Entire population				
	d	py	m	SE	e	d	py	m	SE	e	d	py	m	SE	e
5	41	2335	0.0176	0.0027	35	6	1631	0.0037	0.0015	56	47	3966	0.0119	0.0017	41
10	54	3717	0.0145	0.0020	33	10	2560	0.0039	0.0012	52	64	6277	0.0102	0.0013	38
20	89	3980	0.0224	0.0024	27	8	2264	0.0035	0.0012	44	97	6243	0.0155	0.0016	32
30	47	2032	0.0231	0.0034	23	14	1222	0.0115	0.0031	35	61	3254	0.0187	0.0024	26
40	38	944	0.0403	0.0065	17	7	564	0.0124	0.0047	29	45	1508	0.0298	0.0044	21
50	23	405	0.0568	0.0118	14	5	238	0.0210	0.0094	22	28	643	0.0435	0.0082	16
60	9	106	0.0849	0.0283	11	5	105	0.0476	0.0213	16	14	211	0.0664	0.0177	13
Total	301	13519				55	8584				356	22102			

Abbreviations: d, empirical deaths; e, resulting life expectancies; m, mortality rates; py, person-years of exposure; SE, standard errors.

TABLE 3 Logistic regression survival model.

Variable	Odds ratio (95% confidence interval)	<i>p</i>
Male	1.07 (0.86, 1.32)	0.56
Female	1.00	
Ages 5–9 years	1.00	
Ages 10–29 years	1.44 (0.97, 2.12)	0.07
Ages 30–39 years	2.45 (1.56, 3.87)	<0.001
Ages 40 years and older	2.87 (1.71, 4.82)	<0.001
Each year above age 40 years	1.06 (1.04, 1.09)	<0.001
Cannot walk or self-feed	7.13 (4.24, 12.00)	<0.001
Cannot walk; can self-feed	3.04 (1.91, 4.84)	<0.001
Walks with support or unsteadily alone	1.29 (0.74, 2.25)	0.36
Walks well independently	1.00	
No bowel or bladder control	1.54 (1.18, 2.01)	0.002
At least some bowel or bladder control	1.00	
Calendar year	0.99 (0.97, 1.00)	0.03

statistically significant ($p = 0.56$), the term was retained because inclusion of sex is necessary to facilitate comparisons with the general population. After controlling for the above factors, the effect of hydrocephalus was not statistically significant (odds ratio = 1.15, $p = 0.26$, not shown). There were no significant interaction effects ($p > 0.05$ in all cases).

In the final mortality model with terms for age, sex, walking/feeding ability, and bowel/bladder continence, there was a statistically significant secular (time) trend in survival ($p = 0.03$). The odds ratio was 0.99, indicating roughly 1% improvement (i.e. reduction) in mortality per calendar year, on average, over the 1986 to 2019 time period.

Life expectancies by age and sex for seven cohorts (by walking/feeding ability and bowel/bladder control) are given in Table 4, along with the associated general population life expectancies. As suggested by the survival model of Table 3, the resulting life expectancies are greatly affected by the level of impairment. They also decreased markedly

with age, and are modestly lower for males. For example, amongst patients who walk with support and have at least some bowel or bladder control, the life expectancies at age 10 years are 57 years and 58 years for males and females, compared with 65 years and 70 years in the general population. At age 50 years they are 25 years and 26 years compared with 28 years and 32 years.

Also shown in Table 4 are previously published life expectancies in two groups of patients with a SCI, leading to ostensibly similar physical impairments. At age 10 years, for example, males with American Spinal Injury Association Impairment Scale Grade D SCI have a life expectancy of 58 years, which is intermediate to the 57 to 61 additional years for those in the functionally similar spina bifida groups who can walk with support or alone and who have at least some bowel and bladder control. Conversely, males age 10 years with paraplegia due to SCI have a life expectancy of 49 years compared with only 37 to 44 years in patients with spina bifida who similarly cannot walk though can self-feed.

TABLE 4 Male life expectancies by age, functional level, and extent of bowel/bladder control.^a

Age	Cannot walk or self-feed	Cannot walk; can self-feed		Walks with support or unsteadily alone		Walks well independently		SCI ^b		General population
	BB: None	None	Any	None	Any	None	Any	Paraplegia	Grade D	
Males										
5	27	41	48	55	62	59	65	53	63	70
10	24	37	44	51	57	55	61	49	58	65
20	20	31	37	43	49	47	52	40	49	55
30	15	24	30	35	41	38	44	32	40	46
40	11	18	23	28	33	31	35	25	31	37
50	7	13	17	21	25	23	27	18	23	28
60	5	9	12	15	18	17	20	13	16	21
Females										
5	28	42	49	56	63	60	66	58	67	75
10	25	38	45	52	58	56	62	53	62	70
20	21	32	38	44	50	48	53	44	52	61
30	16	25	31	36	41	39	44	36	43	51
40	12	19	24	29	33	31	36	28	34	41
50	8	14	17	22	26	24	28	21	26	32
60	5	9	12	15	19	17	20	15	19	24

^aAny = some or complete control of bowel and bladder (BB). Note that this group is a mixture of those with some BB control and others with full control. Persons with full BB control had modestly higher life expectancies than those shown here for the 'Some+' group (results not shown). Conversely, those with limited though not full control had modestly lower life expectancies.

^bFor spinal cord injury (SCI) paraplegia and Grade D injuries, please see text for description.

The higher life expectancy in SCI may reflect that such patients rarely have a cognitive impairment, and most can live and work independently.

We also examined possible changes in patient functioning with age. For those aged 5 to 15 years at the start of follow-up, 80% did not change in their ambulatory ability, and 85% did not change with respect to continence. For those in the intermediate functional or continence groups, where changes in either direction were possible, such changes were roughly equally likely to be improvement or regression. At ages 40 years and over, regression was more common than improvement.

DISCUSSION

We confirmed that survival in spina bifida depends critically on the severity of impairment, especially motor function, feeding ability, and continence. The importance of neurological function in general on survival has been previously documented extensively in the population with spina bifida,^{5,12,23} and also in cerebral palsy,¹⁸ traumatic brain injury,¹⁹ and SCI.²⁰ In all of these conditions, greater function was associated with longer survival. In particular, prior spina bifida studies have found that more severe lesions,^{24–26} neurological level,¹² motor level,¹⁰ and perineal sensation²³ were highly associated with survival.

Regarding motor level specifically, this is not surprising as those with lesions at level L1 and above were more likely to

be immobile, wheelchair-bound, or incontinent, and to have hydrocephalus or a low IQ.¹² Yet prior researchers also found that observed motor deficits 'were not entirely predicted by motor level', and that 'this highlights the possible need for an improved spina bifida classification system that ... defines a particular individual's functional motor abilities'.²⁷ (p.1) The functional strata described here appear to meet this criterion.

Sensory level in infancy,²³ in addition to being correlated with long-term survival,²³ is correlated with continence and urological outcome.⁸ Bowel and bladder issues are the most common health issue in patients spina bifida,²⁸ and the most common reason for hospital admission is genitourinary procedures.²⁹ Bladder dysfunction is a leading cause of morbidity in the population with spina bifida, it puts them at higher risk of urinary tract infections, and may be a risk factor for bladder cancer because of both (1) exposure to radiation because of multiple and ongoing radiographic studies, and (2) intrinsic bladder issues concomitant with a neurogenic bladder in general.³⁰ Also, as noted by Oakeshott et al.,²³ (p.68) 'the protective function of sensibility to pain is an invaluable asset'.

Our findings regarding the associations of ambulation/feeding ability and bowel/bladder control with survival mirror results reported for Grade D patients with SCI,³¹ where the ability to walk and the need for bladder catheterization were also found to be key factors. The life expectancy comparisons here with those disabled by childhood- or adult-onset SCI may be appropriate because of the resulting similarities in impairments. Amongst patients with similar

functional and self-care abilities, we found that the etiology of disability (i.e. whether due to spina bifida or SCI) does not appear to have a major effect on survival. It has also been posited that a comparison with those with an acquired brain injury, such as traumatic brain injury, may be apt because of a similar cerebellar pathology,¹³ though we do not report those figures here.

Regarding the issue of improvement in survival over time, we found a modest trend: mortality decreasing by roughly 1% per calendar year. Similarly, both Glinianaia et al.⁶ and Shin et al.⁷ reported improved survival, the latter being restricted to 1-year survival after birth rather than longer term. Others have found improvements in the survival of with cerebral palsy.³² Conversely, no trends were found in the populations with SCI or traumatic brain injury.^{19,20}

Three other factors bear brief mention. In what follows we restrict attention to the non-missing data. First, in our population 64% had an intellectual disability. Similarly, yet using different terminology reflecting differing definitions, Mayes et al.³³ found that 60% have a learning disability, while Riedel et al.³⁴ reported that while persons with spina bifida often have cognitive impairments, only 26% (9/35) of their group had an IQ less than 70. Second, we found that 72% had hydrocephalus and thus presumably also required shunting. Oakeshott et al.¹⁰ reported 89%, while Bowman et al.² reported 86%, though theirs were both birth series and ours was the subset who had hydrocephalus at an older age. Finally, 27% in our cohort had epilepsy, similar to the 23% reported by Bowman et al.²

Limitations of the present study include significant missing values for region and hydrocephalus, and that the results apply only to those aged 5 years and older who require ongoing services or therapy. Further, the life expectancies calculated here are predicated on the assumption of no major change in motor function or continence with age. Because function is highly associated with survival, this assumption may have led to bias in the results. Notably, functional improvement is slightly more likely than regression in the younger ages, which suggests that the life expectancies in the lower functioning groups are, if anything, too low. On the other hand, we also found that regression is more common at older ages, making the life expectancies of the higher functioning groups, if anything, too high. Given the competing issues, and that the observed changes are modest in nature and span an average 13-year period, any resulting bias may be minimal. Further research on this issue is warranted. In addition, investigation of the causes of death would shed light on whether improvements in survival over the past 40 years are related to changes in what have previously been referred to as unexpected causes of death such as epilepsy, pulmonary embolisms, renal sepsis, or acute hydrocephalus.⁵

This is the first long-term study of patients with open spina bifida to report life expectancies by age, sex, and severity of impairment. We hope the results given here will aid

patients and caregivers alike in the proper planning for and treatment of those living with spina bifida.

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DATA AVAILABILITY STATEMENT

Research data are not available from the authors.

ORCID

Robert M. Shavelle  <https://orcid.org/0000-0001-5601-0759>

Matthew K. Paige  <https://orcid.org/0009-0009-2827-6192>

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