J Rehabil Med 2023; 55: jrm18244

ORIGINAL REPORT

SURVIVAL AND CAUSES OF DEATH IN ADULTS WITH SPINA BIFIDA IN SWEDEN: A POPULATION-BASED CASE-CONTROL STUDY

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Objective: **To analyse survival rates and causes of death in adults with spina bifida in Sweden compared with a matched control group.**

Design and methods: **This population-based study included 11,900 adults born between 1950 and 1997. Three national Swedish registers were used to identify individuals with a diagnosis of spina bifida and a matched control group without spina bifida in the period 1990–2015. International Classification of Diseases codes were used to identify causes of death. Survival analysis was conducted and causes of death in the 2 groups were compared.**

Results: **There was a lower probability of survival for people with spina bifida in all age groups ($p < 0.001$) compared with the control group. The most prevalent causes of death in people with spina bifida were congenital, respiratory, nervous, cardiovascular, genitourinary, and injuries. People with spina bifida had a higher probability of dying from congenital ($p < 0.001$), respiratory ($p = 0.002$), genitourinary ($p < 0.002$), and nervous-related ($p < 0.001$) and lower probability of injury-related deaths ($p < 0.001$).**

Conclusion: **Adults with spina bifida in Sweden have a lower survival rate compared with the general population, with the frequency of certain causes of death differing between the two groups. In order to reduce excess premature mortality, prevention and careful management of potentially fatal conditions are essential throughout a patient's lifespan.**

Key words: spina bifida; adults; transition to adult care; mortality; survival.

Accepted Sept 29, 2023

J Rehabil Med 2023; 55: jrm18244.

DOI: 10.2340/jrm.v55.18244

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LAY ABSTRACT

This study investigated survival and causes of death in adults with spina bifida, a neural congenital defect, in Sweden, compared with a group of individuals without spina bifida. National registers were used to identify individuals with spina bifida and a control group, and their causes of death were analysed. The results show that adults with spina bifida had a lower probability of survival in all age groups compared with the control group. The most common causes of death in people with spina bifida were related to their condition (identified as congenital), respiratory, nervous, cardiovascular, genitourinary issues, and injuries. People with spina bifida had a higher chance of dying from certain causes. In conclusion, adults with spina bifida in Sweden have a lower survival rate and different causes of death than adults without spina bifida. It is crucial to prevent and effectively manage potentially life-threatening conditions throughout a the lifespan of people with spina bifida. This study highlights the importance of addressing the healthcare needs of adults with spina bifida in order to reduce premature mortality.

Spina bifida (SB) is a neural tube defect resulting from the failure of fusion or development of the caudal neural tube. The prevalence of SB varies across time, geographical region, and ethnicity (1). The degree of disability depends on factors such as SB level of lesion and type. SB is categorized into open and closed. Open SB includes myelomeningocele (MMC; the most common and severe type of SB), myeloschisis, and hemimyelomeningocele, whereas closed SB includes meningocele and myelocystocele (2). Approximately 80% of individuals with SB have comorbid hydrocephalus (3, 4). The incidence of SB has decreased in the last decades, whereas the survival rate among infants has increased to approximately 75% (5). The prevalence in the adult population has increased since the 1960s due to advancements in surgery and medical care (6). People with SB often face health-related challenges related to comorbid and/or secondary conditions (7). These conditions involve body function and structure, activity limitation, and participation restriction (8). The transition to adult healthcare is often challenging and not well structured, with few multidisciplinary clinics available for adults. This results in a lack of follow-up, increased risk of hospitalization, an increase in preventable conditions, and, possibly, avoidable deaths (6, 9).

The current literature on causes of death in adults with SB is limited and research on mortality focuses mainly on children. In addition, most studies are

hospital-based rather than population-based and include older cohorts of individuals, thus not reflecting improvements in medical treatment and care. The most common causes of death identified in previous studies are infectious (including shunt-related), renal, cardio-pulmonary, and causes connected to the nervous system. A higher incidence of genitourinary cancers has also been reported; however, it is still an object of study. Individuals with SB also have an increased risk of secondary conditions, such as pressure injuries, hypertension, metabolic syndrome, and urological complications (5, 10–14). In the UK, the mortality rate of people with SB between 5 and 40 years was found to be 10 times higher than in the general population, and approximately half of the deaths were reported to be sudden and unexpected (5). Mortality has also been found to be associated with factors such as degree of disability, type of SB, level of lesion, presence of a shunt and Arnold Chiari malformation II, socioeconomic status, and parental education (5, 14–16). It is important to determine the causes of mortality and survival rates among adults with SB because if there is premature death caused by preventable causes, proactive and systematic work across the lifespan could potentially increase survival rates.

The aim of this study was to investigate the causes and timing of death in adults aged 18–65 years with SB compared with a matched control group in Sweden. It was hypothesized that adults with SB would have higher risk of early death and that the leading causes of death would differ compared with the general population. Finally, it was hypothesized that certain urological neoplasms would be more common causes of death in adults with SB than in those without SB.

METHODS

Study population and procedure

A population based, case-contro study was conducted including deaths that occured between the years 1990 and 2015 in Sweden. Participants with SB were identified by their diagnostic International Classification of Diseases (ICD) codes (741 for ICD-9 and Q05 for ICD-10). Eligible participants with SB over 18 years of age were identified for the period 1990–2015 in several Swedish national registers. Diagnostic codes in national registers may be overestimated due to, for instance, misdiagnosis (17). To address this, exclusion criteria were: individuals with diagnoses considered incompatible with SB (anencephaly and similar anomalies: 740, 740A, 740B, 740C; atresia and stenosis of large intestine, rectum, and anal canal: 751C; persistent cloaca: Q437; anencephaly: Q00; congenital absence, atresia, and stenosis of large intestine: Q42; other congenital malformations of nervous system-Arnold Chiari: Q07), individuals with other spinal diagnoses that lack a specific SB diagnosis (with codes Q05.1–8 and 741), and individuals with SB diagnosis only from the medical birth register but not from the national patient register (unless listed as a cause of death). A general population comparison group of adults without SB or cerebral palsy was drawn from the register of the total population at a 5:1 ratio, matched on sex, birth year, and area of residence. Yearly information for 1990–2015 from several national registers was linked to the current study population of cases and controls, including date and cause/s of death from the causes of death register, socioeconomic information from Statistics Sweden's Longitudinal Integrated Database for Health Insurance and Labor Market Studies, and diagnoses from the national patient register. The medical birth register and the combined national quality and follow-up register for MMC (MMCUP) were also used.

Measures and variables

Demographics and medical conditions. Demographics included age, sex, and foreign background. The ICD-specific codes of level of lesion and type of SB (741 followed by a number between 00 and 03, and 90 and 93, or Q05 followed by a number from 1 to 9, defining the level of lesion and the presence of hydrocephalus) were included. If more than 1 ICD code was reported for type of SB, the following criteria were used: (*i*) if an unspecified and a specified diagnosis were present, the more specific diagnosis was used; (*ii*) if 2 or more specific codes were present, the most frequent code was used; (*iii*) if 2 or more specific codes were present, and none was used more frequently than the others, the most recent diagnosis was used. The following comorbidities were included: hydrocephalus (codes G91, 331.3, 331.4, 331.5, 741.0, 742.3, Q05.0, Q05.1, Q05.2, Q05.3, Q05.4), scoliosis (M41, 737.3), kyphosis (M40.1, M40.2, 737.0, 737.1), and intellectual disability (mild: F70, 317, moderate: F71, 318.0, severe: F72, 318.1, profound: F73, 318.2, other/unspecified: F78, F79, 319).

Causes of death. Causes of death were classified into categories based on their ICD 9 and 10 codes. Each death certificate includes one underlying cause defined as the disease or injury which initiated the train of morbid events leading directly to death or the circumstances of the accident or violence that produced the fatal injury, and multiple contributing causes, which are used to indicate the chain of events leading to death (18). Using the European shortlist of causes of death (19), the causes were categorized as

congenital malformations and chromosomal abnormalities; diseases of the blood (-forming organs); immunological disorders; of the circulatory system; of the digestive system; of the genitourinary system; of the musculoskeletal system/connective tissue; of the nervous system and the sense organs; of the respiratory system; of the skin and subcutaneous tissue; endocrine, nutritional, and metabolic diseases; external causes of injury and poisoning; infectious and parasitic diseases; mental and behavioural disorders; neoplasms; and symptoms, signs, abnormal findings, ill-defined causes (diagnoses that cannot be classified elsewhere). The ICD-10 category "factors influencing health status and contact with health services" was also included. Causes that were found to be more frequent in adults with SB were then divided into subcategories to analyse the incidence of specific diseases or causes in that population. These subcategories were obtained by looking more specifically at ICD codes, where additional digits are used to specify the type of disease. Based on previous findings, neoplasms were also an object of focus and, to this end, the general categories were divided into subgroups based on their aetiology.

Statistical analysis

Causes of death. Descriptive statistics were used to describe the characteristics of the population. The frequency of each cause of death in cases and controls was summarized and 2-proportions Z-tests were used to assess the difference in the probability of death for each cause between the two groups. This analysis was conducted for underlying causes of death and for contributing causes of death in total as well as stratified by sex and age. Finally, we divided the most frequent causes of death in adults with SB into subcategories defined by more specific ICD codes and calculated the frequency of those in the entire study population. A 2-proportions Z-test was performed to compare the frequency of genitourinary neoplasms, which were of particular interest, in adults with and without SB.

Survival analysis. Survival analysis was conducted to compare survival in adults with SB and controls. First, the whole study population was included in the analysis and the time to event was the main parameter: individuals entered the study in 1990, and survival over 25 years (until 2015) was analysed. The same analysis was then conducted dividing the population into three age groups: 18–25, 26–45, and 46–65 years. Kaplan–Meier curves with log-rank tests were used to describe the likelihood of survival, followed by a Cox proportional-hazards model controlling for sex, place of birth, parental education, and comorbidities (scoliosis, kyphosis, intellectual disability, and hydrocephalus). Finally, the proportional hazard assumption was tested to confirm that the hazard ratios were constant over time. All statistical analyses were conducted using the statistical software R (v R 4.1.0, May 2021), and statistical significance was set at $p < 0.05$.

RESULTS

In total, 11,990 individuals, 1,943 cases and 10,047 controls, were included, of whom 54.5% were female and 45.5% male. The characteristics of the total study population by group are shown in Table I. 9.4% had a diagnosis of hydrocephalus, 4.5% scoliosis, 0.7% kyphosis, and 2.1% intellectual disability. During the study period, a total of 264 individuals died (151 cases and 113 controls), of whom 154 were male (84 cases and 70 controls) and 110 female (67 cases and 43 controls). Overall, these numbers correspond to 8.4% of cases, and 1.1% of controls.

The median age of death was 38.6 years in adults with SB and 40.2 years in controls. The frequency of deaths in each age group (18–25, 26–45, and 46–65 years) was 14.6%, 58.3%, and 27.1%, respectively, in people with SB and 23%, 35.4%, and 41.6% in people without SB. Among people with comorbidities, specifically hydrocephalus, scoliosis, intellectual disability, and kyphosis, the frequency of deaths was, respectively, 8.4%, 8.6%, 5.4%, 3.8% in those who also had SB. Among people with comorbidities who did not have SB, 4.6% of those with intellectual disability died.

Survival 25 years after inclusion in the study was 84.6% for cases compared with 97.8% for controls

Table I. Characteristics of the sample

	Persons with spina bifida	Persons without spina bifida
Total individuals in the sample, *n*	1,943	10,047
Socio-economic characteristics		
Sex		
Female, *n* (%)	1058 (54.5)	5482 (54.6)
Male, *n* (%)	885 (45.5)	4565 (45.4)
Foreign background		
Born in Sweden, *n* (%)	1670 (85.9)	9222 (91.8)
Born abroad, *n* (%)	273 (14.1)	825 (8.2%)
Parental education		
Higher education, *n* (%)	532 (27.4)	3114 (31.0)
Secondary education, *n* (%)	885 (45.5)	4667 (46.5)
Mandatory education, *n* (%)	268 (13.8)	1365 (13.6)
Information missing, *n* (%)	258 (13.3)	901 (8.9)
Comorbidities		
Hydrocephalus		
Yes, *n* (%)	1130 (58.2)	0
No, *n* (%)	813 (41.8)	10,047 (100)
Scoliosis		
Yes, *n* (%)	444 (22.9)	93 (0.9)
No, *n* (%)	1,499 (77.1)	9,954 (99.1)
Intellectual disability		
Yes, *n* (%)	184 (9.5)	65 (0.6)
No, *n* (%)	1759 (91.5)	9,982 (99.4)
Kyphosis		
Yes, *n* (%)	80 (4.1)	8 (0.1)
No	1843 (95.9)	10,039 (99.9)

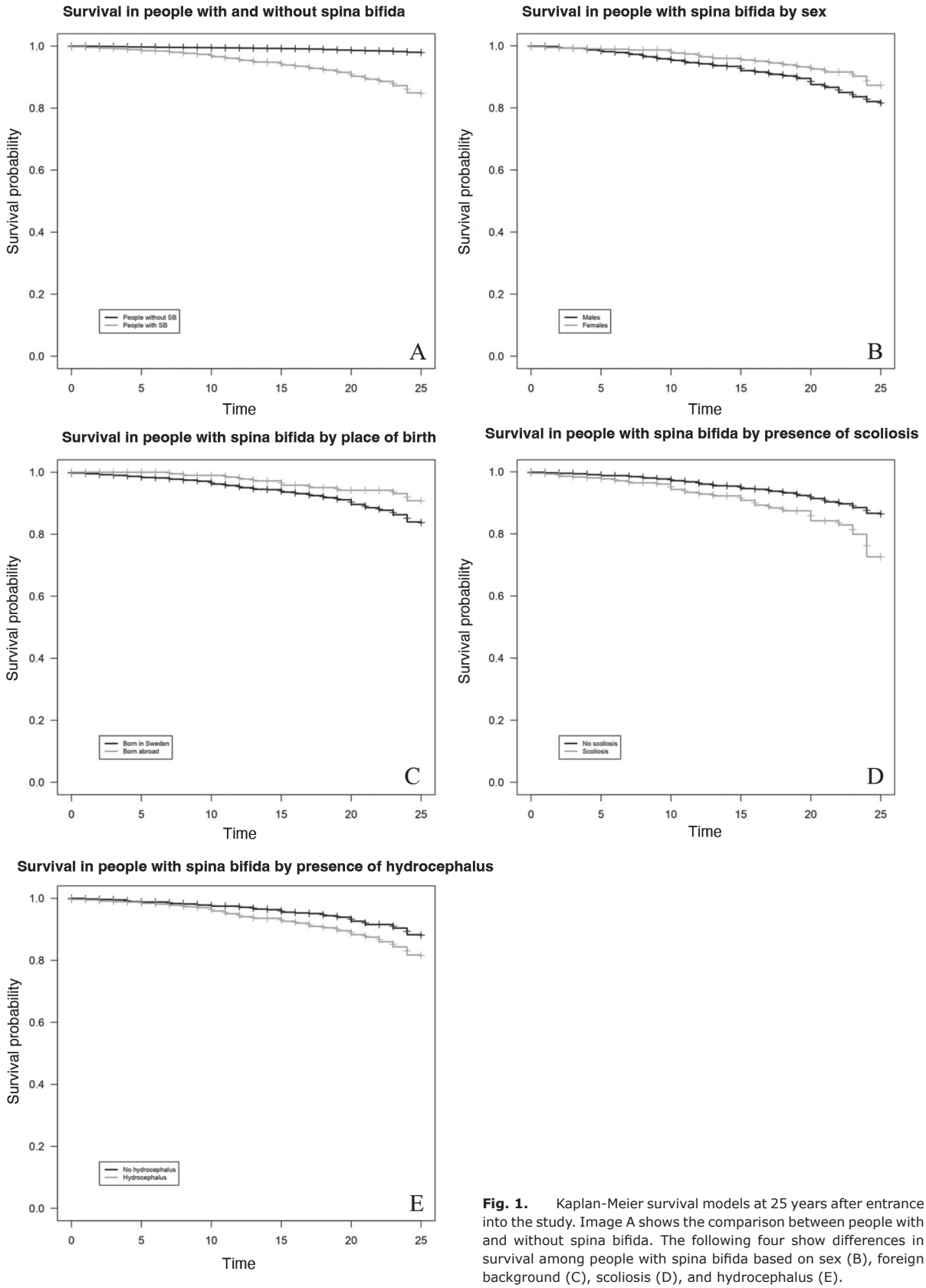

Fig. 1. Kaplan-Meier survival models at 25 years after entrance into the study. Image A shows the comparison between people with and without spina bifida. The following four show differences in survival among people with spina bifida based on sex (B), foreign background (C), scoliosis (D), and hydrocephalus (E).

(Fig. 1). Furthermore, survival was analysed in the three different age groups, which were 86.8%, 80.0%, 80.8% in the 18–25, 26–45, and 46–65 years age groups, respectively, after 25 years, compared with 98.7%, 97%, and 92.8% among controls. When analysing only adults with SB, the model showed lower survival rates in males, people born in Sweden, people with scoliosis, and those with hydrocephalus. Survival in these groups was 81.2%, 83.6%, 72.6%, and 81.3% after 25 years vs 87.3%, 90.8%, 86.3%, and 88.0% in their respective control groups.

Causes of death

Underlying causes of death. The most frequent underlying causes of death in adults with SB were congenital malformations and chromosomal abnormalities, diseases of the circulatory system, and neoplasms. Adults with SB had a higher probability of dying from congenital malformations and chromosomal abnormalities ($p<0.001$) and diseases of the genitourinary system ($p=0.02$), whereas controls died more frequently from external causes of injury and poison ($p<0.001$) and neoplasms ($p=0.02$) (Table II). However, adults with SB had a higher probability of having genitourinary-related neoplasms listed as a cause of death ($p<0.001$). Adults with SB were at higher risk of dying from congenital malformations and chromosomal abnormalities in all three age groups, which was also the case for the total population included in the study. No statistically significant difference was observed in frequency of genitourinary conditions by age groups, which is in contrast to what was noted for the entire study population. Controls had a higher frequency of death related to external causes of injury and poison in the age group 18–45 years ($p<0.001$). However, the difference in frequency was not significant after 45 years of age. There were no statistically significant differences observed in cases vs controls when comparing the frequency of causes by sex: both groups showed a higher probability of external causes of death in males ($p=0.02$ in cases and $p=0.01$ in controls). Probability of neoplasms was statistically significantly higher in females for controls ($p=0.01$).

Contributing causes of death. The study then investigated all contributing causes of death recorded in the death certificates. The number of causes of death listed ranged from 1 to 14, with a mean of 4.8 for adults with SB and 3.4 for controls. Overall, the most common causes listed for adults with SB were congenital malformations and chromosomal abnormalities, diseases of the respiratory system, and diseases of the nervous system and the sense organs (Table II). Adults with SB had a higher probability of congenital malformations and chromosomal abnormalities ($p<0.001$), diseases of the respiratory system ($p=0.002$), of the genitourinary system ($p<0.001$), of the nervous system ($p<0.001$), symptoms, signs, abnormal findings, and ill-defined causes ($p<0.001$), and factors influencing health status and contact with health services (e.g. "persons with potential health hazards related to family and personal history and certain conditions influencing health status" and "tobacco use") ($p=0.04$). Controls had a higher frequency of deaths by external causes of injury and poison ($p<0.001$). The right-hand column in Table II shows the listed

Table II. Frequency of causes of death reported in death certificates for persons with and without spina bifida. Right-hand column shows the contributing causes of death listed in persons who had spina bifida reported as an underlying cause of death in their death certificates

	Underlying cause		Contributing cause		
Causes of death	Frequency in persons with spina bifida (%)	Frequency in persons without spina bifida (%)	Frequency in persons with spina bifida (%)	Frequency in persons without spina bifida (%)	Contributing causes of death in persons with spina bifida as an underlying cause (%)
Congenital malformations and chromosomal abnormalities	**30.5**	**0.9**	**52.3**	**1.8**	31.5
Diseases of the blood (-forming organs), immunological disorders	0.0	1.8	2.6	2.70	0
Diseases of the circulatory system	17.2	12.4	31.8	21.2	8.0
Diseases of the digestive system	4.6	1.8	12.6	9.7	6.2
Diseases of the genitourinary system	**6.0**	**0.0**	**25.8**	**2.7**	9.7
Diseases of the musculoskeletal system/connective tissue	4.6	0.9	5.3	1.8	2.7
Diseases of the nervous system and the sense organs	4.6	3.5	**32.5**	**8.0**	17.7
Diseases of the respiratory system	3.3	2.7	**33.1**	**15.9**	14.2
Diseases of the skin and subcutaneous tissue	0.7	0.9	6.6	2.7	2.7
Endocrine, nutritional, and metabolic diseases	1.3	0.9	10.6	4.4	4.4
External causes of injury and poisoning	**10.6**	**46.9**	**24.5**	**50.4**	7.1
Factors influencing health status and contact with health services	0.0	0.0	**4.0**	**18.6**	4.4
Infectious and parasitic diseases	3.3	1.8	11.3	8.0	3.5
Mental and behavioural disorders	0.7	2.7	6.0	13.3	2.7
Neoplasms	**11.3**	**21.2**	14.6	22.1	1.8
Symptoms, signs, abnormal findings, ill-defined causes	1.3	1.8	**29.1**	**11.5**	13.3

Statistical significance shown in bold.

contributing causes of death of adults with SB as the underlying cause of death.

After categorizing neoplasms into different groups based on body systems, adults with SB with neoplasms listed as a cause of death were shown to have a higher frequency of genitourinary-related neoplasms ($p = 0.008$), 52.9% vs 8.3% among the controls. Furthermore, among contributing causes of death, 31.4% of neoplasm in adults with SB were of genitourinary type, compared with 5.3% in controls. All genitourinary neoplasms listed in controls involved female genital organs, whereas this type of neoplasm in adults with SB showed a different aetiology. Among underlying causes of death 88.9% of these neoplasms were urinary and 11.1% neoplasms of the male organs; among all causes of death, 81.8% were urinary and 18.1% involved male genital organs.

Survival

The Cox proportional hazard model showed that having SB increased the hazard of early death by a factor of 7.3 ($p < 0.001$), non-adjusted. When adjusting for sex, place of birth, and parental education, the hazard of early death increased to 7.4 ($p < 0.001$). Results from the model are shown in Table III.

A survival analysis was then conducted only for adults with SB, to investigate how different factors affected survival in the group. Sex, place of birth parental education, hydrocephalus and scoliosis were included in the model to investigate the effect of the different factors on survival. A statistically significant reduction in hazard ratios by a factor of 0.62 for females ($p = 0.003$) and 0.51 for people born outside Sweden ($p = 0.02$) were found. No statistically significant difference in survival was found when comparing parental education. Hazard ratios were increased by a factor of 2.1 for those with scoliosis ($p < 0.001$) and 1.6 (1.60 adjusted) for those with hydrocephalus ($p < 0.001$). The proportional hazard assumption was insignificant, indicating that the risk of death was proportional over time for the two groups and thus that the proportional hazard assumption holds.

Table III. Hazard ratios (HR) in persons with spina bifida and without spina bifida (controls), and in persons with spina bifida and possible contributing variables

	Variable	HR	*p*-value	95% CI
Persons with spina bifida and controls				
Unadjusted	Spina bifida	7.25	<0.001	5.68–9.25
Adjusted for sex, foreign background, and parental education	Spina bifida	7.42	<0.001	5.81–9.48
Persons with spina bifida				
Contributing variables	Female	0.56	0.003	0.45–0.85
	Born outside Sweden	0.53	0.03	0.28–0.92
	Hydrocephalus	1.64	0.003	1.18–2.29
	Scoliosis	1.98	<0.001	1.37–2.87
	Parental education			
	Maximum mandatory	Ref		
	Maximum secondary	0.78	0.23	0.51–1.18
	Maximum higher	0.63	0.069	0.38–1.03
	Missing information	0.66	0.10	0.400–1.09

95% CI: 95% confidence interval.

DISCUSSION

The results of this study confirmed the hypotheses that adults with SB have a higher rate of premature death compared with the general population and that listed causes of death between adults with SB and their counterparts without SB differ.

Adults with SB have lower survival in all age groups: the percentage who reached age 25, 45, and 65 years was consistently lower than the percentage of those in the general population. This finding is consistent with previous literature, although it varies in proportion. In a report by Oakeshott et al. (5), the mortality rate in individuals with SB was 10 times higher between ages 5 and 40 years. Although the current study confirms that mortality is higher, the proportion is lower. This can probably be explained by differences in age groups included. By only considering adults, individuals with more severe forms of disability or comorbidities may have died before reaching the threshold to be included in the study. Nevertheless, these findings confirm that this trend is still present in adulthood.

When considering underlying causes of death in adults with SB, this study observed the difference between what was classified as congenital malformations and chromosomal abnormalities (in 30.5% of the group), which includes SB, and other causes, with only 3 other categories (diseases of the circulatory system, injuries, and neoplasms) reaching over 10%. Because of this, the analysis of underlying causes does not completely capture the complexity of the situation as well as the analysis of contributing causes listed in the certificates. SB is a congenital disability and not a fatal disease as such. Nevertheless, physicians seem to list SB as a cause of death even though the actual cause was something else.

The findings of the current study on the most frequent causes of death in adults with SB are in accordance with the study hypothesis as well as with previous studies (12–14), with the five most prevalent causes being diseases of the respiratory (33.1%), nervous (32.5%), cardiovascular (31.8%), genitourinary system (25.8%), and injuries (24.5%). These results confirm that those diseases are among the most relevant causes of death for this group, both in hospital-based studies, as previously reported by Peyronnet et al. (12) and Dicianno et al. (13), and in population-based studies

based on register data, as that by Kancherla et al. (15). The type of urinary tract damage present in the current study differed from previous research, Peyronnet et al. (12) showed a higher frequency of upper urinary tract damage, whereas 80% of those in the current study were related to the kidneys. This may be the result of differences between the hospitalized and non-hospitalized populations. Moreover, even when considering neoplasms, a cause more frequent among controls overall, death caused by genitourinary cancers was more frequent in the SB population.

One important difference in the current findings compared with previous research is the lower prevalence of infections in terms of causes of death among adults with SB. Although 11.3% of the adults with SB had infections reported in their death certificates, this aetiology was not among the most prevalent in the current study population. This is in contrast to a study by Dicianno et al. (13), in which infections were the most frequent causes of death in inpatients. This apparent discrepancy can probably be explained by the type of population included in the study. It is plausible to assume that inpatients may be more likely to either present with an infection when they are admitted or develop one during their hospital stay. Furthermore, non-hospitalized individuals may be at a higher risk of dying from the complications of chronic diseases rather than acute ones. The discrepancy could also be explained by the way diseases were classified in the studies. In fact, the ICD classification used in the Swedish cause of death register classifies system-specific infections under their systematic category rather than in a general infection category.

As expected, causes of death in adults with SB were different from those listed in the general population. The results of the current study confirm that causes connected to the genitourinary, nervous, and respiratory systems are more frequent in adults with SB than in the control group. It is interesting to note that, while injuries are a frequent cause of death among adults with SB (with almost a quarter reporting it), the probability of it is higher in controls both among underlying causes and all listed causes.

With injuries being one of the most frequent causes of death in younger age groups in the general population, it is possible that adults with SB are more likely to die from other conditions that are less common in the general population, possibly as a result of the disability. The difference between the groups disappears in the older age group (46–65 years).

Strengths and limitations

This study has several limitations. Register-based data and the information on death certificates vary in quality. As there is no specific indication regarding the order in which causes of death are listed on death certificates, except for the underlying causes, it was not possible to determine precisely which causes were more relevant to the death and which ones were more secondary. Using ICD codes recorded in death certificates rather than clinical information may influence the accuracy of some of the information. For example, more specific diagnoses may have been missed, both when classifying the level of lesions and presence of hydrocephalus in SB, and specific causes of death. Specifically, in most adults with SB in the current study sample, the SB diagnosis was classified as "unspecified" rather than with a specific level of lesion. That information would have enabled more specific analyses of causes of death related to the level of lesion and the severity of SB. Furthermore, cause of death classification does sometimes not follow clinical classifications. For example, infections are often reported under their related system rather than in a general infection category, leading to an underestimation of the cause. Furthermore, the study was affected by left-truncated data: information from the register of causes of death was only available from 1990 and onwards; hence the study may have missed individuals who died before this date. The study population excludes individuals who died after 2015 and individuals born after 1997, who had not reached age 18 years by 2015, the last study year. Finally, the low number of comorbidities recorded among people in the current sample may have resulted in biased results regarding their causes of death or survival.

Nevertheless, the current study had several strengths. It was a population-based, register study, with a matched control group, meaning that it was possible to include data on an extensive cohort of individuals, not limited to a hospital setting or healthcare settings. This increases the generalizability of the findings. The focus of the study was mortality in adults with SB. As specific research on adults with SB is scarce in the literature, this study offers an important perspective, in particular given that survival to adulthood has been increasing in recent years. Moreover, the study cohort spans a long period of time, including individuals born up to 1997. This allows us to account for recent changes and improvements in treatment compared with older cohorts of individuals with SB. Also, the participants included in the study had a wide spectrum of severity, from more severe to milder cases, allowing us to study causes of death not restricted to a specific type or severity of disability.

Conclusion

This study demonstrates that, although treatments for SB have improved over the years, monitoring of

adults with SB for specific diseases and comorbidities that are more likely to become fatal is necessary. This would facilitate prevention and may result in a higher survival rate. As previous studies have demonstrated, close attention must be paid during the transition from paediatric to adult care. Practices for good follow-up and smooth transition should be initiated during paediatric care. In fact, many adults with SB lack appropriate care during this phase and in adulthood, something that often results in higher hospitalization rates and risk of death (5).

Interventions should be put in place to make sure that adults with SB have access to an adequate level of care and to multidisciplinary resources. The fact that many adults with SB and hydrocephalus are likely to have executive dysfunction must be taken into account when developing and implementing healthcare services for adults with SB.

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