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## Skin Ulcers and Mortality Among Adolescents and Young Adults With Spina Bifida in South Carolina During 2000–2010

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### Abstract

The authors investigated 48 deaths (7% death rate) among 690 adolescents and young adults with spina bifida in South Carolina during 2000–2010. The authors used Medicaid and other administrative data and a retrospective cohort design that included people with spina bifida identified using ICD-9 codes. Cox regression models with time-dependent and time-invariant covariates, and Kaplan–Meier survival curves were constructed. The authors found that 21.4% of the study group had a skin ulcer during the study period and individuals with skin ulcers had significantly higher mortality than those without ulcers ( $P < .0001$ ). People who had their first skin ulcer during adolescence had higher mortality than those who had the first skin ulcer during young adulthood ( $P = .0002$ ; hazard ratio = 10.70, 95% confidence interval for hazard ratio: 3.01, 38.00) and those without skin ulcers, controlling for other covariates. This study showed that age at which individuals first had a skin ulcer was associated with mortality.

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#### Author Contributions

BC was responsible for the design of data analysis, data analysis, and manuscript write-up. SM assisted in initial design of data analysis and the first version of the manuscript. YW and JAR assisted in data manipulation, and YW assisted in data analysis. JRM, JWH, OO, and LO contributed to the final critical review and the manuscript.

#### Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

#### Ethical Approval

The study was reviewed by the University of South Carolina Institutional Review Board, and the project received exempt status.

## Keywords

Adolescent; young adult; health; skin ulcer; spina bifida; survival analysis

Spina bifida is the most common permanently disabling birth defect.<sup>1</sup> About 1 out of every 1400 to 1500 newborns are affected with spina bifida each year in the United States.<sup>2</sup> This defect occurs when there is a failure of proper fusion of the spinal column, which can significantly impair neurologic functions. Many individuals with spina bifida also experience hydrocephalus, which can result in significant cognitive impairment and other problems with the central nervous system. The degree of disability ranges from mild to severe, based on the size and location of the spine lesion, and whether there is hydrocephalus.<sup>1</sup>

People with spina bifida typically need specialized medical care throughout their lives. The functional sequelae of spina bifida include lower body paralysis, bowel and bladder dysfunction, and cognitive impairment (if there is accompanying hydrocephalus that is not managed properly).<sup>3</sup> People with spina bifida typically have associated health conditions such as spinal cord dysfunction and hydrocephalus, resulting in elevated morbidity and mortality.<sup>4</sup> As a result, they frequently seek health care such as physician primary and specialty care (orthopedists, urologists, physiatrists, and neurosurgeons), nursing services, and physical and occupational therapy.<sup>5</sup> Long-term sustained health care is important for people with spina bifida to prevent adverse outcomes. Early death in this population is usually related to renal failure, urosepsis, or respiratory complications.<sup>6</sup> Some of these complications can be associated with long-term neurologic conditions such as symptomatic hydrocephalus and tethered cord syndrome which are the most serious neurologic conditions as people age with spina bifida.<sup>6</sup>

With improvements in medical care in the past few decades, increasing numbers of patients with spina bifida survive into adulthood. There have been innovations in prevention of complications from bowel and bladder dysfunction, improvements in skin care routines to prevent decubitus ulcers, better shunt care for people with hydrocephalus, and better surveillance of overall health. Understanding the transition from pediatric to adult health care can help identify health conditions or services that might increase survival of adolescents and young adults with spina bifida.<sup>7</sup> There is a rich literature on improving health care, transition and quality of life for people with spina bifida.<sup>8–11</sup> Oakeshott et al<sup>12</sup> conducted a longitudinal cohort study on survival and causes of death among infants with spina bifida. They found 26% died between the age of 5 and 35 years. McDonnell and McCann<sup>13</sup> investigated the death rate and factors contributing to death among attendees at a clinic for adults with spina bifida and hydrocephalus, where the focus was on shunt malfunction on morbidity, mortality, and sudden death. Recently, Wang et al<sup>14</sup> studied the survival of children with birth defects based on a 25-year population-based data in New York State. Among children born from 1983 to 2006, the survival probability (with 95% confidence interval) is 0.82 (0.80, 0.84). Kancherla et al<sup>15</sup> conducted a population-based study to examine cause-specific mortality among individuals with spina bifida, aged between 0 and 28 years. Among all births between 1983 and 2006, they found that mortality

in individuals with spina bifida was significantly higher than that in those with congenital hypertrophic pyloric stenosis. They also pointed out that although a majority of the deaths for people with spina bifida occurred within the first year of life, an increased risk of death endured in young adults with spina bifida (22 deaths out of 1988 individuals with spina bifida, aged between 16 and 25 years). The causes of death in this age group varied and no cause was identified for greater than 1 person other than multiple congenital anomalies (n = 2) and spina bifida (n = 4). Previous studies reported the pressure ulcer rates as high as 27% in pediatric intensive care units, 23% in neonatal intensive care units, and 20% to 43% among outpatients with spina bifida.<sup>16–19</sup> However, the authors have identified no studies that analyzed skin ulcers as a risk factor for mortality among adolescents and young adults with spina bifida.

The purpose of this study was to investigate risk factors associated with death among adolescents and young adults, 15–24 years of age, with spina bifida during a period that includes the transition from pediatric to adult health care. During the preliminary stages of the study the authors found that skin ulcers were the most common diagnosed condition in the study population, thus the authors considered the skin ulcers as the independent variable. People with spina bifida usually have a sensory and motor deficit in their lower limbs. Thus, toes and feet are the most common site of pressure ulcers.<sup>20</sup> The authors hypothesized that the presence of skin ulcers is a significant predictor of mortality, since these ulcers are likely markers of both the degree of impairment (immobility) and of suboptimal personal and medical care. A secondary purpose was to examine the most common risk factors associated with death for young spina bifida patients.

## Materials and Methods

### Data Sources

A retrospective cohort study was conducted using the state-level administrative data sources in South Carolina. This study is part of a larger project investigating the transition from pediatric to adult services for adolescents and young adults with rare health conditions.<sup>21</sup> The authors used 3 health databases for case ascertainment—Medicaid, State Health Plan and Uniform-billing Hospital Discharge Data, with a unique ID as a linkage for each patient. Vital records death certificate data were extracted using each case's unique ID. The study was approved by the South Carolina Department of Health and Human Services, the South Carolina Employee Benefit Administration, the South Carolina Data Oversight Council, the South Carolina Department of Health and Environmental Control and was granted exempt status by the institutional review board at the University of South Carolina.

### Case Identification

Individuals who were included in the study had to be between 15 and 24 years old for at least 1 year during the 2000 through 2010 time frame, thus they were born between 1975 and 1995. They also had to be enrolled in the South Carolina Medicaid program or State Health Plan for at least 1 calendar year during the study period. Spina bifida cases were identified based on medical billing data diagnosed with ICD-9-CM code (741.0 for spina bifida patients with hydrocephalus and 741.9 spina bifida patients without hydrocephalus).

The additional code (331.3 or 331.4) was also used to indicate the presence of hydrocephalus. In addition, a group of ICD-9 and Current Procedural Technology codes was utilized to identify individuals with shunt malfunction of shunt related procedures. The authors did not have a code for the spinal level of the spina bifida lesion. Among patients with spina bifida, those who received 1 or more of these codes were categorized as having hydrocephalus. During 2000–2010, 690 individuals with spina bifida aged 15–24 years were identified. Death and cause of death for the study participants were identified using the SC vital records with ICD-10 codes.

## Outcome

The outcome is the observed survival time in days and its vital status during the study period. The entry time is the earliest date indicated on the medical billing records when the individual interacted with the health care system. The end time is the last date recorded on the medical billing records or death date. The survival time is the interval between the entry time and the end time. The survival status is 1 if the patient is dead at the end time or 0 if the patient is alive at the end time.

## Explanatory Variables

**Skin ulcer**—The skin ulcer was identified based on medical billing ICD-9 codes (707.0–707.9) from inpatients and outpatients claims. Since the patients in this study were adolescents and young adults, the authors focused on the occurrence and incidence of skin ulcer during this period and its association with mortality. The authors investigated whether there was a difference of survival rate between people with and without skin ulcers, and between people having the first skin ulcer in adolescence and in young adulthood. To do so, the authors created a categorical variable based on time of the first skin ulcer occurrence. This variable was defined as 0 if the first skin ulcer time occurred in adolescence (15–19 years old), 1 if the first skin ulcer occurred during young adulthood (20–24 years old) and 2 for no skin ulcer.

**Time-dependent covariate**—As there are multiple skin ulcers occurring (yes/no) for some patients over multiple visits, which might have a significant effect on survival, the authors included a time-dependent covariate for skin ulcers in the analysis. Using time-varying explanatory variables, when appropriate, leads to more robust results because it utilizes all available data. As the diagnosis time for skin ulcers varied substantially across patients, the authors standardized the diagnostic intervals for all patients using 21-day windows.<sup>22,23</sup> The National Pressure Ulcer Long-Term Care Study used data from 95 long-term care facilities to collect and analyze data about pressure ulcer healing. They found that greater than 95% of episodes ranged from 5–14 days.<sup>23</sup> To be conservative, the authors applied the criteria for distinct episodes of skin ulcers to be at least 21 days apart. The time-dependent variable for skin ulcer across the time windows was coded as 1 if there was at least 1 skin ulcer diagnosis within a window and 0 otherwise. In addition, the number of skin ulcers for each patient was counted and used as a covariate in separate analysis.

**Other covariates**—The other explanatory variables include county of residence type (urban or rural), race (white, black, other), sex (male or female), receipt of food stamps (yes

or no), hydrocephalus (yes or no), primary care (yes or no) or number of primary care visits during the study period, rehabilitation therapy visit (yes or no) or number of therapy visits during the study period, emergency department (ER) visit (yes or no) or number of ER visits during the study period, medical specialist visit (yes or no) or number of medical specialist visits during the study period, and inpatient (IP) stay (yes or no) or number of IP stays during the study period. For the age of individuals, all years were considered if the age was between 15–24 during the 2000–2010 study period, where the minimum age was the age when enrolled in an insurance plan or 15 years if the person was eligible prior to age 15, and the maximum age was 24 years or age at death if the person died prior to reaching age 24. To investigate if there are other co-occurring birth defects in the study group, the authors used the ICD-9-CM codes to identify the potential birth defects including central nervous system, eye/ear, cardiovascular, orofacial, gastrointestinal, genitourinary, musculoskeletal, chromosomal, and other defects. Among those potential defects, 24 individuals had central nervous system defects, 1 patient had eye/ear defect and 2 had genitourinary defects. The other defects were not found among the patients. The central nervous system defect was included in the analysis. To adjust for the history of skin ulcer occurrence prior to the study period, the authors also included a binary variable, skin ulcer history, indicating if a patient had skin ulcer before entering the study. The mean age that an individual was first enrolled in the study is 16.64 years with standard deviation of 2.48 years. The mean age that a patient died or was at the end of the study is 21.18 years with standard deviation of 2.81 years. The mean duration that a person was in the study is 4.60 years with standard deviation of 2.66 years.

## Statistical Methods

Descriptive statistics were included to summarize the information in the data. Statistical tests were used to test the significance of the difference between the groups. For continuous variables, means were compared using t-tests. For categorical variables, the chi-square test (or Fisher's exact test for expected cell sample size < 5) was applied. For statistical modeling, the Cox proportional hazard model was first applied for the time to death with time-invariant explanatory variable, that is, the number of skin ulcers. Then the Cox proportional hazard model with the counting process method was used to deal with the time-varying covariate.<sup>24–27</sup> The survival curves were constructed by using the Kaplan–Meier nonparametric maximum likelihood estimates of the survival probability.<sup>28</sup> Differences in survival were compared with the log-rank test. All analyses were performed using SAS 9.3 (SAS Institute Inc, Cary, NC). Statistical significance was reached when the *P* value was < .05.

## Results

Table 1 and Table 2 present the descriptive summary of the categorical variables (Table 1) and the continuous or count variables (Table 2). The adolescence and young adulthood groups in Table 1 and Table 2 were defined based on the age when they entered the study. Forty-eight patients died among the cohort of 690 individuals with spina bifida during the period of the study (death rate 7%). Nearly 60% of the sample was female and 48% was white. About 18% of the sample used food stamps at some point during the study period.

About 21% of the sample had skin ulcer. This rate of skin ulcer in this study is comparable to the reported rates.<sup>16-19</sup> 13% had first skin ulcer during adolescence, and 8% had first skin ulcer during young adulthood. It is noted that among 55 patients having their first skin ulcer during young adulthood, 37 of them entered the study when they were adolescents but had their first skin ulcer during young adulthood (20–24), while 18 of them entered the study and had their first skin ulcer at young adulthood. Table 3 and Table 4 provide the frequency of key variables according to skin ulcer and vital status, respectively, among patients with spina bifida. Table 3 shows that the proportion of males having skin ulcers is significantly higher than that of females ( $P = .0002$ ). Also, patients with skin ulcers had significantly higher risk of death compared to patients without skin ulcers ( $P < .0001$ ). Patients with skin ulcers were significantly more likely to have hydrocephalus than patients without skin ulcers ( $P < .0001$ ). Individuals with skin ulcer history prior to the study period were more likely to have skin ulcers during their adolescence and young adulthood ( $P < .0001$ ). Among 114 patients who had skin ulcers prior to the study period, 77 patients had skin ulcers during their adolescence (66) and young adulthood (11), which is 52% of 148 patients who had skin ulcers during their adolescence and young adulthood. Individuals with and without food stamp enrollment had different risks of having skin ulcers, but the evidence in favor of the difference being significant was relatively weak ( $P = .05$ ). Table 4 shows, however, that the death proportions were significantly different between patients with and without food stamps ( $P = .009$ ). Skin ulcer history prior to the study period was significantly associated with mortality during adolescence and young adulthood ( $P < .0001$ ). There was a strong association between death and time of the first skin ulcer occurrence during adolescence and young adulthood ( $P = .0004$ ). In terms of anatomic locations of pressure ulcers, among 148 patients who have skin ulcers, 55 (37%) patients had a total of 1733 skin ulcers on their back; 43 (29%) patients had a total of 1302 ulcers on their hip; 60 (41%) patients had a total of 2598 ulcers on their buttock; 14 (9%) patients had a total of 167 ulcers on their ankle; 25 (17%) patients had a total of 722 ulcers on their heels; 49 (33%) patients had a total of 654 ulcers on other anatomical sites. There were 792 unspecified skin ulcers from 68 (46%) patients. Note that a patient can have multiple skin ulcers and the patients for different types of skin ulcers are not mutually exclusive.

Table 5 and Table 6 show the parameter estimates and hazard ratios in survival analysis based on the Cox regression model with time-varying skin ulcer diagnosis and the number of skin ulcers, respectively. Table 5 shows that age is a significant predictor of death among patients with spina bifida ( $P < .0001$ ). This implies that people at an older age (ie, age at start) had a significantly higher risk of death than those at a younger age. There is no significant difference of risk of death between male and female patients with spina bifida. Patients who had more inpatient stays had a higher risk of death than those with fewer inpatient stays ( $P = .002$ ; hazard ratio = 1.15, 95% confidence interval for hazard ratio: 1.06, 1.24). Patients with skin ulcer history prior to the study period had a higher risk of death than those who had no skin ulcer history, adjusting for the other covariates ( $P = .01$ ). People who had their first skin ulcer during adolescence had a much higher risk of death than people who had their first skin ulcer during young adulthood (reference group in the model) ( $P = .0002$ ; hazard ratio = 10.70, 95% confidence interval for hazard ratio: 3.01, 38.00). There was no significant difference in the risk of death between people who had the first



skin ulcer during young adulthood and those without skin ulcers ( $P = .07$ ; hazard ratio = 2.59, 95% confidence interval for hazard ratio: 0.93, 7.18). This illustrates that for patients with spina bifida, developing skin ulcers at a younger age could more likely lead to a higher risk of death. Table 6 shows the similar results, where the risk of death for people with more skin ulcers was not significantly different from that for those with fewer skin ulcers ( $P = .23$ ).

Figure 1 presents the Kaplan–Meier survival curves for people who had the first skin ulcer during adolescence (0), had the first skin ulcer in young adulthood (1), and had no skin ulcer (2). It shows that people who had their first skin ulcer in adolescence had a lower survival probability than people in the other 2 groups. The log-rank test for comparison of the 3 groups resulted in a  $P$  value = .036. To investigate the potential parity across the groups (eg, number of skin ulcers between males and females), the authors also included interactions into the model and none showed statistical significance. To examine confounding, the authors also fitted Cox models excluding the categorical variable for the first skin ulcer related variable (ie, time of the first skin ulcer occurrence) and number of skin ulcers separately. In addition, the authors conducted the stepwise selection process which consists of a series of alternating forward selection and backward elimination steps. All of the results (not shown) remained basically the same.

Among 48 deaths in patients with spina bifida, there were 20 underlying causes of death related to spina bifida including spina bifida, unspecified spina bifida with hydrocephalus, cerebral palsy, pneumonitis, and urinary tract infection. For the sake of simplicity the authors categorized contributing causes of death codes according to ICD-10 chapter, with the exception of spina bifida, which the authors maintained as a unique category. Up to 10 different conditions can be listed as contributing causes of death. The authors did not take order into account but instead included every listed contributing cause. The authors counted each category of contributing cause once, even if an individual had multiple causes listed within a particular category.

The appendix shows the top 10 contributing causes of death by ICD-10 mortality book chapter. Disease of the respiratory system was the most commonly reported category for contributing cause of death (23 out of 48). Spina bifida was second (20), followed by other diseases of the nervous system (17) and injury, poisoning and other consequences of external causes (15), diseases of the circulatory system (15) and diseases of the genitourinary system (15). Besides the direct contributing cause of death due to spina bifida, diseases of the genitourinary system are likely associated with spina bifida, which frequently results in a neurogenic bladder that can ultimately lead to renal failure. This information showed that the major causes of death reflect the course of spina bifida.

## Discussion

The authors investigated the risk factors that can have significant impact on the death of adolescent and young adult patients with spina bifida based on the data from South Carolina during the period 2000–2010. In the general population, the major cause of death during adolescence and young adulthood is injury. If adolescents and young adults with spina bifida

die more of spina bifida related conditions than injury-related conditions it would indicate that the course/severity of spina bifida is of more concern than injuries for patients with the condition. The authors used death certificates to identify the underlying and contributing causes of death for people with spina bifida who died during the study period.

The authors found that among patients with spina bifida, people with their first skin ulcer diagnosis during adolescence had significantly higher risk of death than those with their first skin ulcer diagnosis in young adulthood (estimated hazard ratio of 10.7) and those without skin ulcers. In contrast, patients with their first skin ulcer diagnosis in young adulthood had no significant difference of death risk from those without skin ulcers. Although the result showed that the association between the first skin ulcer occurred in adolescence and mortality was stronger than that from the other groups, it did not imply that the late skin ulcer occurrence is not important. As a potential indicator related to death, skin ulcers can indicate either worse functional status (eg, less able to ambulate, etc) or poorer care (eg, self-care, family care, nutrition care, hospital care, community care, etc).<sup>29</sup> These high risk factors for skin ulcers can lead to more frequent cause of mortality in adolescents than in young adults.

On the other hand, the number of skin ulcers seems to have no significant impact on the risk of death among patients with spina bifida. This is somewhat counterintuitive. One potential explanation for the lack of an association between number of skin ulcer diagnoses and risk of death would be repeated episodes of treatment for the same ulcer outside the 21-day window used for these analyses. For example, they might have been receiving regular wound care past the 21-day period. It is not possible, with the data available to us, to definitively distinguish new occurrences of skin ulcer from repeated episodes of treatment for the same ulcer. This lack of precision might have introduced a nondifferential misclassification bias in this analysis by using this variable, diminishing the authors' ability to test for association. To investigate if changing the length of the window could affect the result, the authors tried the 30-day window and 60-day window separately. The results from the Cox model with the newly defined windows are basically the same as those based on the model with the 21-day window.

Besides modeling the time-to-event data for spina bifida patients with the time-varying covariate (ie, using all time-dependent skin ulcers), the authors also fitted the Cox proportional model with the time-invariant covariate (ie, using the first skin ulcer). The model including time-invariant skin ulcer also did not provide the significant effect of skin ulcer on the risk of death ( $P = .66$ ).

There are several limitations in this study. First, the administrative data that the authors relied on were not collected for research. Thus, data from patients with spina bifida who were not in the list of the health care system were not collected. Second, since the data collected were from patients who had health insurance, those without health insurance were excluded, probably resulting in a sample in which most individuals were from a higher socioeconomic status. This type of bias might be the cause of the result in Table 4 where the proportion of deaths for spina bifida patients without food stamps is significantly higher than for those with food stamps ( $P = .009$ ). Third, as the medical claims were not directly linked



to clinical data or medical records, some patients with spina bifida were probably misclassified or missing in the dataset. Fourth, currently there are 2 types of insurance claims available in the system including Medicaid and the State Health Plan. An individual can have a claim in Medicaid, or in the State Health Plan, or in either of them over different time periods. Although the majority of insurance is Medicaid, the information of types of insurance is incomplete as the authors cannot identify whether an individual was covered by another payer (eg, 1 type of private insurance) that supplements Medicaid. In addition, it must be noted that, because this study included follow-up only through the age of 24, there was a shorter follow-up for individuals diagnosed with skin ulcers in young adulthood than for those diagnosed in adolescence. It is possible that an association between ulcers diagnosed in young adulthood and risk of death can be detectable if follow-up were extended further into adulthood. Nonetheless, although this is not a complete population based study, the data sources include people with public and private insurance in a state of over 4 million residents. The generalizability of these findings cannot be specified until another study is conducted in additional populations and hopefully, with all citizens included in the dataset.

Clinicians providing care for adolescents and young adults with spina bifida should be aware that occurrence of skin ulcer, especially during adolescence, can be an indicator of a higher risk for death. Skin ulcer can be a marker for a more severe underlying disability related to impaired mobility, suboptimal health care or personal care, or a deteriorating health status that eventually can lead to death. Additional research is needed to investigate the association between early skin ulcer occurrence and the increased risk of death.

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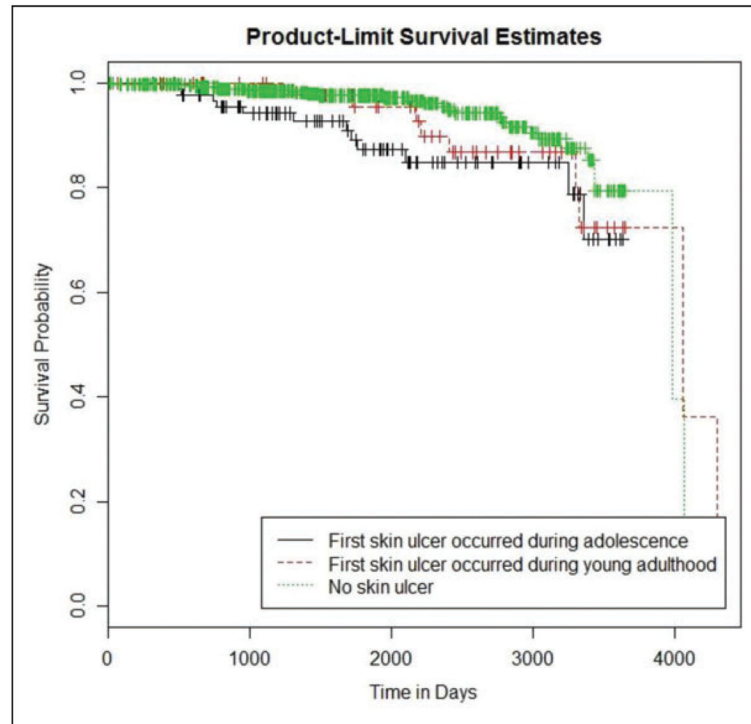
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## Appendix

### Top 10 Contributing Causes of Death (N = 48 Deaths/690 People With Spina Bifida)

Mortality by ICD-10 chapter title	Frequency excluding spina bifida
Diseases of the respiratory system	23
Spina bifida	20
Diseases of the nervous system	17
Injury, poisoning, and certain other consequences of external causes	17
Diseases of the circulatory system	15
Diseases of the genitourinary system	15
Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	14
Certain infectious and parasitic diseases	13
External causes of morbidity and mortality	13
Mental and behavioral disorders	10 <sup>a</sup>

<sup>a</sup>Cells containing 10 individuals were masked to protect patients' confidentiality.



**Figure 1.** Kaplan–Meier survival plots according to the time of occurrence of the first skin ulcer in 690 individuals with spina bifida during the follow-up period. The log-rank test for comparison of the 3 groups resulted in a  $P$  value = .036.

**Table 1**

Descriptive Summary of Categorical Variables for Adolescents and Young Adults With Spina Bifida.

Categorical variables	All (N = 690) n (%)	Adolescents (n = 576) n (%)	Young adults (n = 114) n (%)
Sex			
Female	408 (59.1)	336 (58.3)	72 (63.2)
Male	282 (40.9)	240 (41.7)	42 (36.8)
County type			
Rural	201 (29.1)	161 (28.0)	40 (35.1)
Urban	489 (70.9)	415 (72.0)	74 (64.9)
Race			
Black	215 (31.2)	178 (30.9)	37 (32.5)
Others	142 (20.6)	130 (22.6)	12 (10.5)
White	333 (48.2)	268 (46.5)	65 (57.0)
Food stamps			
Yes	126 (18.3)	126 (21.9)	0 (0.0)
No	564 (81.7)	450 (78.1)	114 (100.0)
Hydrocephalus			
Yes	527 (76.0)	447 (77.6)	80 (70.2)
No	163 (24.0)	129 (22.4)	34 (29.8)
Emergency department visit			
Yes	488 (70.7)	424 (73.6)	64 (56.1)
No	202 (29.3)	152 (26.4)	50 (43.9)
Inpatient visit			
Yes	394 (57.1)	345 (59.9)	49 (43.0)
No	296 (42.9)	231 (40.1)	65 (57.0)
Primary care visit			
Yes	622 (90.1)	541 (93.9)	81 (71.1)
No	68 (9.9)	35 (6.1)	33 (28.9)
Specialist visit			
Yes	498 (72.2)	442 (76.7)	56 (49.1)
No	192 (27.8)	134 (23.3)	58 (50.9)
Rehabilitation therapy visit			
Yes	372 (53.9)	353 (61.3)	19 (16.7)
No	318 (46.1)	223 (38.7)	95 (83.3)
Other visit			
Yes	>600 <sup>a</sup>	>560 <sup>a</sup>	>100 <sup>a</sup>
No	<10 <sup>a</sup>	<10 <sup>a</sup>	<10 <sup>a</sup>
Central nervous system defect			
Yes	24 (3.5)	>10 <sup>a</sup>	<10 <sup>a</sup>
No	666 (96.5)	>500 <sup>a</sup>	>100 <sup>a</sup>
Skin ulcer history			

Categorical variables	All (N = 690) n (%)	Adolescents (n = 576) n (%)	Young adults (n = 114) n (%)
Yes	114 (16.5)	97 (16.8)	17 (14.9)
No	576 (83.5)	479 (83.2)	97 (85.1)
Time of first skin ulcer occurrence			
First skin ulcer occurred during adolescence	93 (13.4)	93 (16.2)	0 (0.0)
First skin ulcer occurred during young adulthood	55 (8.0)	37 (6.4)	18 (15.8)
No skin ulcer	542 (78.6)	446 (77.4)	96 (84.2)
Death			
Yes	48 (7.0)	29 (5.0)	19 (16.7)
No	642 (93.0)	547 (95.0)	95 (83.3)

<sup>a</sup>Cells containing 10 individuals were masked to protect patient's confidentiality; the counts in the other lines were masked due to availability of the total number.

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**Table 2**

Descriptive Summary of Selected Variables for Adolescents and Young Adults With Spina Bifida.

<b>Continuous variable</b>	<b>All (N = 690) Mean (SD)</b>	<b>Adolescents (n = 576) Mean (SD)</b>	<b>Young Adults (n = 114) Mean (SD)</b>
Age (years)	16.6 (2.5)	15.7 (1.2)	21.5 (1.3)
Emergency department visits	4.7 (9.4)	5.1 (9.9)	3.0 (6.4)
Inpatient stays	1.8 (3.2)	2.0 (3.3)	1.1 (2.4)
Primary care visits	18.7 (27.0)	21.2 (28.6)	6.3 (8.4)
Medical specialist visits	9.3 (15.0)	10.3 (15.9)	4.6 (8.0)
Rehabilitation therapy visits	56.9 (176.9)	67.0 (191.9)	6.0 (20.4)
Number of other visits	196.9 (410.2)	212.2 (424.2)	119.6 (321.7)
Number of skin ulcers	7.6 (32.3)	8.3 (34.1)	4.5 (20.9)

The number of visits is the total number of visits during the follow-up period. SD, standard deviation.

**Table 3**

Two-Way Contingency Table for Skin Ulcer Indicator Among Adolescents and Young Adults With Spina Bifida.

Variable	Skin ulcer (%)		P value
	No	Yes	
Sex			
Female	340 (49.3)	68 (9.9)	.0002
Male	202 (29.3)	80 (11.6)	
Race			
Black	166 (24.1)	49 (7.1)	.23
Others	106 (15.4)	36 (5.2)	
White	270 (39.1)	63 (9.1)	
Food stamps			
No	435 (63.0)	129 (18.7)	.05
Yes	107 (15.5)	19 (2.8)	
Hydrocephalus			
No	150 (21.7)	15 (2.2)	<.0001
Yes	391 (56.7)	134 (19.4)	
County type			
Rural	164 (23.8)	37 (5.4)	.21
Urban	378 (54.8)	111 (16.1)	
Central nervous system defect			
No	>500 <sup>a</sup>	>100 <sup>a</sup>	.05
Yes	>10 <sup>a</sup>	<10 <sup>a</sup>	
Skin ulcer history			
No	505 (73.2)	71 (10.3)	<.0001
Yes	37 (5.4)	77 (11.2)	
Death			
No	515 (74.6)	127 (18.4)	<.0001
Yes	27 (3.9)	21 (3.0)	

<sup>a</sup>Cells containing 10 individuals were masked to protect patient's confidentiality; the counts in the other lines were masked due to availability of the total number.

**Table 4**

Two-Way Contingency Table for Death Among Adolescents and Young Adults With Spina Bifida.

Variable	Death (%)		P value
	No	Yes	
Sex			
Female	382 (55.4)	26 (3.8)	.47
Male	260 (37.7)	22 (3.2)	
Race			
Black	>150 <sup>a</sup>	>10 <sup>a</sup>	.09
Others	>100 <sup>a</sup>	<10 <sup>a</sup>	
White	302 (43.8)	31 (4.5)	
Food stamps			
No	>500 <sup>a</sup>	>40 <sup>a</sup>	.009
Yes	>100 <sup>a</sup>	<10 <sup>a</sup>	
Hydrocephalus			
No	> 150 <sup>a</sup>	< 10 <sup>a</sup>	.06
Yes	> 400 <sup>a</sup>	> 40 <sup>a</sup>	
County type			
Rural	>100 <sup>a</sup>	<10 <sup>a</sup>	.10
Urban	>400 <sup>a</sup>	>30 <sup>a</sup>	
Central nervous system defect			
No	>600 <sup>a</sup>	>30 <sup>a</sup>	.08
Yes	>10 <sup>a</sup>	<10 <sup>a</sup>	
Skin ulcer history			
No	549 (79.6)	27 (3.9)	<.0001
Yes	93 (13.5)	21 (3.0)	
Time of first skin ulcer occurrence			
First skin ulcer occurred during adolescence	>70 <sup>a</sup>	>10 <sup>a</sup>	.0004
First skin ulcer occurred during young adulthood	>40 <sup>a</sup>	<10 <sup>a</sup>	
No skin ulcer	>500 <sup>a</sup>	>20 <sup>a</sup>	

<sup>a</sup> Cells containing 10 individuals were masked to protect patient's confidentiality; the counts in the other lines were masked due to availability of the total number.

**Table 5**  
Results of Proportional Hazard Model With the Time-Varying Covariate for Adolescents and Young Adults With Spina Bifida.

Parameter	Parameter estimate	SE	Pr > ChiSq	Hazard ratio	95% confidence interval
Sex					
	F	0.41	.22	1.51	0.78 2.92
Race					
	Black	-0.52	.16	0.59	0.30 1.23
	Others	-0.16	.78	0.85	0.30 2.60
	White	—	—	—	—
Age					
		0.34	<.0001	1.41	1.24 1.60
Food stamps					
	No	0.64	.41	1.90	0.41 8.81
County type					
	Rural	-0.47	.24	0.62	0.28 1.37
Hydrocephalus					
	No	-0.45	.35	0.58	0.60 4.07
First skin ulcer occurred during adolescence					
	0	2.21	.0002	10.70	3.01 38.00
First skin ulcer occurred during young adulthood					
	1	—	—	—	—
	2	0.95	.07	2.59	0.93 7.18
No skin ulcer					
Primary care visits					
		-0.002	.80	1.00	0.99 1.01
Medical specialist visits					
		-0.03	.09	0.97	0.94 1.00
Emergency department visits					
		-0.02	.42	0.98	0.94 1.02
Inpatient stays					
		0.14	.002	1.15	1.06 1.24
Rehabilitation therapy visits					
		-0.0005	.63	1.00	0.998 1.00
Central nervous system defect					
	Yes	0.77	.15	2.15	0.74 6.25
Skin ulcer history					
	Yes	1.03	.01	2.79	1.26 6.18
Time-varying skin ulcer diagnoses					
		-0.61	.45	0.54	0.11 2.61

**Table 6**  
Results of Proportional Hazard Model With Number of Skin Ulcer Diagnoses for Adolescents and Young Adults With Spina Bifida.

Parameter	Parameter estimate	Standard error	Pr > ChiSq	Hazard ratio	95% confidence interval		
Sex	F	0.40	0.33	.23	1.50	0.77	2.88
Race	Black	-0.52	0.37	.16	0.59	0.28	1.23
Race	Others	-0.19	0.56	.74	0.83	0.27	2.53
Race	White	—	—	—	—	—	—
Age		0.34	0.06	<.0001	1.41	1.23	1.60
Food stamps	No	0.45	0.77	.56	1.57	0.35	7.09
County type	Rural	-0.45	0.39	.26	0.63	0.30	1.40
Hydrocephalus	No	0.32	0.48	.34	1.38	0.52	3.62
First skin ulcer occurred during adolescence	0	2.48	0.64	.0001	12.02	3.41	42.41
First skin ulcer occurred during young adulthood	1	—	—	—	—	—	—
No skin ulcer	2	0.84	0.52	.11	2.31	0.84	6.38
Number of primary visits		-0.001	0.006	.82	1.00	0.99	1.01
Number of medical specialists		-0.03	0.02	.05	0.97	0.94	1.00
Emergency department visits		-0.031	0.02	.17	0.97	0.93	1.01
Inpatient stays		0.15	0.04	.0009	1.16	1.07	1.26
Rehabilitation therapy visits		-0.0006	0.001	.57	1.00	0.997	1.001
Central nervous system defect	Yes	0.91	0.55	.10	2.48	0.83	7.33
Skin ulcer history	Yes	1.01	0.40	.01	2.73	1.24	6.04
Number of skin ulcer diagnoses		-0.005	0.004	.23	1.00	0.99	1.00