

## STATISTICAL BRIEF #21

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### Sickle Cell Disease Patients in U.S. Hospitals, 2004

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#### Introduction

Among the genetic-based anemias, sickle cell disease is a major cause of morbidity and mortality. Inheritance of a single copy of the sickle cell gene (trait or carrier state) is rarely associated with anemia or disease. In contrast, inheritance of two copies of the sickle cell gene causes severe, lifelong anemia. It is estimated that 2,000 infants are identified annually with sickle cell disease in the U.S., and estimates suggest 50,000 Americans, primarily of African descent, have sickle cell anemia.<sup>1</sup>

In addition to anemia, patients present with acute pain episodes (“crises”) and infections, as well as conditions due to chronic sickle cell disease, including cardiac, pulmonary, renal, and brain complications. Sickle cell crises often require hospitalization. Pain relief, antibiotics, and fluids comprise the standard supportive care for treatment of sickle cell anemia and painful crises. In March 1998, hydroxyurea was approved by the U.S. Food and Drug Administration for treatment of sickle cell disease in patients over 18 with at least three painful crises in the previous year. In clinical research studies, significant reductions in morbidity and mortality were associated with hydroxyurea therapy.<sup>2,3</sup>

This Statistical Brief presents data from the Healthcare Cost and Utilization Project (HCUP) on nationwide hospitalizations for sickle cell disease from 1994 through 2004. Patient characteristics, outcomes, costs of hospital stays, and associated diagnoses and procedures are examined for patients admitted for sickle cell disease. All differences between estimates noted in the text are statistically significant at the 0.05 level or better.

#### Findings

In 2004, there were an estimated 113,098 hospital stays during which sickle cell disease (SCD) was noted, of which nearly three-quarters were for adults with SCD. Table 1 compares patient and

#### Highlights

- The number of hospitalizations among adults with sickle cell disease (SCD) in 2004 was 83,149, which was equivalent to the number of hospitalizations in 1997.
- In 2004, one-fourth of hospitalized patients were children, but less than 2 percent were infants (age <1). About two-thirds of hospitalized patients with sickle cell disease were 18–44 years old.
- For adults with SCD, 31 percent of hospitalizations included blood transfusion. About 26 percent involved infections, 18 percent had substance-related and other mental disorders, and 14 percent had pulmonary conditions.
- The total hospital costs in 2004 for hospitalizations principally for SCD were approximately \$488 million. Among those hospital stays principally for SCD, 66 percent were paid by Medicaid and 13 percent were paid by Medicare, both public payers of health care.
- In 2004, the total number of in-hospital deaths among adults with SCD was 699, and among children 47. In 1998, in-hospital deaths among adults spiked to 758 from 516 in 1997. In contrast, the in-hospital deaths have remained relatively low and constant for children with SCD.

<sup>1</sup>Sickle Cell Disease Guideline Panel. *Sickle cell disease: screening, diagnosis, management, and counseling in newborns and infants*. Clinical Practice Guideline No.6. Rockville, MD: Agency for Healthcare Policy and Research, U.S. Department of Health and Human Services, 1993 (AHCPR publication 93-0562).

<sup>2</sup>Charache S, Terrin ML, Moore RD, Dover GJ, Barton FB, Eckert SV, et al. Effect of hydroxyurea on the frequency of painful crises in sickle cell anemia. Investigators of the Multicenter Study of Hydroxyurea in Sickle Cell Anemia. *N Engl J Med*. 1995 May 18;332(20):1317-22.

<sup>3</sup>Steinberg MH, Barton F, Castro O, Pegelow CH, Ballas SK, Kutlar A, et al. Effect of hydroxyurea on mortality and morbidity in adult sickle cell anemia: risks and benefits up to 9 years of treatment. *JAMA*. 2003 Apr 2;289(13):1645-51. Erratum in *JAMA*. 2003 Aug 13;290(6):756.

hospital characteristics for hospital stays with a principal diagnosis of SCD with all hospital stays where SCD was noted either as the principal or secondary diagnosis. Nearly 70 percent of the hospitalizations were principally for SCD.

From 1994 to 1997, there was a small, non-significant increase in hospitalizations for SCD in adults, from an estimated 70,636 to 84,721 (figure 1). During this same time, there was a small, non-significant decline for SCD hospitalizations among children, from an estimated 35,434 to 32,585. Similarly, the number of hospitalizations between 1999 and 2001 and between 2001 and 2004 in adults and children with SCD did not change significantly. Overall, the number of hospitalizations among adults in 2004 was 83,149, which was equivalent to the number of hospitalizations in 1997.

Less than 2 percent of hospitalizations for SCD were among infants (age <1) or adults ages 65–84. Two-thirds of hospitalizations for SCD were for patients ages 18–44, one-quarter were for 1–17 year olds, and approximately 10 percent were for 45–64 year olds. More than half the admissions were among females, and three-quarters of admissions came through the emergency department. The average length of stay was more than five days. More than half the hospitalizations were among patients originating from low-income areas. Over two-thirds of patients resided in large metropolitan areas, and 94 percent of stays occurred in metropolitan hospitals. Two-thirds of SCD hospital stays were in teaching institutions. The highest hospitalization rate per 10,000 black population occurred in the Northeast, followed by the Midwest, South, and West regions of the U.S.

SCD hospitalizations occur in just over half of U.S. hospitals (figure 2). Forty-three percent of all U.S. hospitals had no hospitalizations for SCD, and less than 5 percent had greater than 1000 hospitalizations. One-third of hospitals had between 1 and 100 hospitalizations, and approximately one-sixth had between 100 and 1,000 hospitalizations for SCD.

As shown in figure 3, the most common associated conditions among children hospitalized with SCD were infections (29 percent of hospitalizations) and pulmonary conditions (27 percent). About 21 percent of hospital stays for children with SCD involved blood transfusions. For adults with SCD, 31 percent of hospital stays involved blood transfusions, and 1 percent involved dialysis. Infections were noted in 26 percent of adult stays for SCD. Other common conditions among adult SCD hospitalizations included substance-related and other mental disorders (18 percent) and pulmonary conditions (14 percent).

Among those principally hospitalized for SCD, more than three-quarters of stays were billed to public payers (figure 4). Two-thirds of hospitalizations for SCD were billed to Medicaid, and an additional 13 percent were billed to Medicare. About 15 percent of hospitalizations for SCD were paid through private insurance, and 4 percent had no insurance. This compares with hospitalizations for all conditions in 2004, in which Medicaid was billed 25 percent, Medicare 33 percent, and private insurance 34 percent. The average cost for each hospitalization was \$6,223, and the total estimated cost for SCD hospitalizations in 2004 was \$488 million.

Overall, the number of in-hospital deaths in 2004 for hospitalizations principally for SCD was 238, an in-hospital death rate of 0.30 percent. There were an estimated 746 deaths for all hospitalizations that included SCD in the diagnoses, a death rate of 0.70 (table 1). Figure 5 shows the number of in-hospital deaths and death rate for SCD hospitalizations from 1994 to 2004, comparing children with adults. In 1994, for adults with SCD, there were an estimated 480 in-hospital deaths, which was a death rate of 0.68 per 100 hospitalizations. The number of in-hospital deaths, as well as death rate, was stable from 1994 to 1997. Between 1997 and 1998, there was a significant 47 percent increase in deaths and death rates for adult hospitalizations with SCD, to a peak of 758 deaths. From 1998 through 2004, the in-hospital deaths and death rate remained stable for adults. In contrast, the in-hospital deaths and death rate have remained relatively low and constant over the same time period for children with SCD.

Figure 6 shows the principal diagnoses associated with in-hospital death for stays involving sickle cell disease. About 28 percent of hospitalizations that resulted in death had SCD with crisis listed as the principal diagnosis. Infection (including sepsis and pneumonia) was the principal diagnosis for 22 percent of SCD hospitalizations that ended in death. Cardiac conditions (8 percent), respiratory failure (8 percent), stroke (5 percent), and gastrointestinal conditions (5 percent) were also leading principal diagnoses for SCD-related stays that resulted in death.

## Data Source

The estimates in this Statistical Brief are based upon data from the HCUP 1994–2004 Nationwide Inpatient Sample (NIS). Supplemental sources included data from the U.S. Census Bureau, Population Division, Annual Estimates of the Population for the United States, Regions, and Divisions.

## Definitions

### *Types of hospitals included in HCUP*

HCUP is based on data from community hospitals, defined as short-term, non-Federal, general and other hospitals, excluding hospital units of other institutions (e.g., prisons). HCUP data include OB-GYN, ENT, orthopedic, cancer, pediatric, public, and academic medical hospitals. They exclude long-term care, rehabilitation, psychiatric, and alcoholism and chemical dependency hospitals, but these types of discharges are included if they are from community hospitals.

### *Unit of analysis*

The unit of analysis is the hospital discharge (i.e., the hospital stay), not a person or patient. This means that a person who is admitted to the hospital multiple times in one year will be counted each time as a separate "discharge" from the hospital.

### *Region*

Region is one of the four regions defined by the U.S. Census Bureau:

- Northeast: Maine, New Hampshire, Vermont, Massachusetts, Rhode Island, Connecticut, New York, New Jersey, and Pennsylvania
- Midwest: Ohio, Indiana, Illinois, Michigan, Wisconsin, Minnesota, Iowa, Missouri, North Dakota, South Dakota, Nebraska, and Kansas
- South: Delaware, Maryland, District of Columbia, Virginia, West Virginia, North Carolina, South Carolina, Georgia, Florida, Kentucky, Tennessee, Alabama, Mississippi, Arkansas, Louisiana, Oklahoma, and Texas
- West: Montana, Idaho, Wyoming, Colorado, New Mexico, Arizona, Utah, Nevada, Washington, Oregon, California, Alaska, and Hawaii

### *Costs and charges*

Total hospital charges were converted to costs using cost-to-charge ratios based on hospital accounting reports from the Centers for Medicare and Medicaid Services (CMS).<sup>4</sup> Costs will tend to reflect the actual costs of production, while charges represent what the hospital billed for the case. For each hospital, a hospital-wide cost-to-charge ratio is used because detailed charges are not available across all HCUP States. Hospital charges reflect the amount the hospital charged for the entire hospital stay and does not include professional (physician) fees. For the purposes of this Statistical Brief, costs are reported to the nearest hundreds.

### *Diagnoses, ICD-9-CM, and Clinical Classifications Software (CCS)*

The principal diagnosis is that condition established after study to be chiefly responsible for the patient's admission to the hospital. Secondary diagnoses are concomitant conditions that coexist at the time of admission or that develop during the stay. All-listed diagnoses include the principal diagnosis plus these additional secondary conditions.

ICD-9-CM is the International Classification of Diseases, Ninth Revision, Clinical Modification, which assigns numeric codes to diagnoses. There are about 12,000 ICD-9-CM diagnosis codes.

CCS categorizes ICD-9-CM diagnoses into 260 clinically meaningful categories.<sup>5</sup> This "clinical grouper" makes it easier to quickly understand patterns of diagnoses and procedures.

CCS 61 was used to identify primary hospitalizations for sickle cell disease.

<sup>4</sup>HCUP Cost-to-Charge Ratio Files (CCR). Healthcare Cost and Utilization Project (HCUP). 2001–2003. U.S. Agency for Healthcare Research and Quality, Rockville, MD. [www.hcup-us.ahrq.gov/db/state/costtocharge.jsp](http://www.hcup-us.ahrq.gov/db/state/costtocharge.jsp)

<sup>5</sup>HCUP CCS. Healthcare Cost and Utilization Project (HCUP). August 2006. U.S. Agency for Healthcare Research and Quality, Rockville, MD. [www.hcup-us.ahrq.gov/toolsoftware/ccs/ccs.jsp](http://www.hcup-us.ahrq.gov/toolsoftware/ccs/ccs.jsp)

The ICD-9-CM codes used to identify all-listed sickle cell disease are 282.41 (Thalasemia Hb-S w/o crisis), 282.42 (Thalasemia Hb-S w/ crisis), 282.6 (Sickle cell anemia, Nos), 282.61 (Hb-S w/o crisis), 282.62 (Hb-S w/ crisis), 282.63 (Sickle cell/Hb-S disease), 282.64 (Sickle cell/Hb-S disease w/ crisis), 282.68 (Other sickle cell w/o crisis), and 282.69 (Sickle cell anemia nec). Several of these codes were added (282.41, 282.42) or modified (282.6, 282.61, 282.62, 282.63, 282.64, 282.68, 282.69) on October 1, 2003. The ICD-9-CM codes used to identify sickle cell disease w/ crisis are 282.42 (Thalasemia Hb-S w/ crisis), 282.62 (Hb-S w/ crisis), and 282.64 (Sickle cell/Hb-S disease w/ crisis).

In figure 2, associated conditions grouped together the most frequent principal CCS categories for SCD hospitalizations. Infections grouped pneumonia (CCS 122), urinary tract infection (CCS 159), fever (CCS 246), bacterial infection NOS (CCS 3), and other upper respiratory infections (CCS 126). Pulmonary conditions grouped asthma (CCS 128), respiratory failure (CCS 131), COPD (CCS 127), and other lower respiratory disease (CCS 133). Cardiac conditions grouped congestive heart failure (CCS 108), cardiomyopathy (CCS 97), and dysrhythmias (CCS 106).

In figure 3, associated conditions grouped together the most frequent principal CCS categories for SCD hospitalizations that ended in death. Infections grouped sepsis (CCS 2), pneumonia (CCS 122), urinary tract infection (CCS 159), fever (CCS 246), and HIV (CCS 5). Cardiac conditions grouped congestive heart failure (CCS 108), cardiomyopathy (CCS 97), and acute myocardial infarction (CCS 100). Gastrointestinal conditions grouped hepatitis (CCS 6), abdominal pain (CCS 251) and biliary tract disorders (CCS 149). Respiratory failure is CCS 131. Stroke is CCS 109. Kidney failure is CCS 157.

#### *Procedures and Clinical Classifications Software (CCS)*

The principal procedure is the procedure that was performed for definitive treatment rather than one performed for diagnostic or exploratory purposes (i.e., the procedure that was necessary to take care of a complication). If two procedures appear to meet this definition, the procedure most related to the principal diagnosis was selected as the principal procedure. Secondary procedures are additional procedures performed during the hospital stay. All-listed procedures include the principal procedure plus these additional secondary procedures.

All-listed CCS 222 identified blood transfusions, and all-listed CCS 58 identified dialysis.

### **About the NIS**

The HCUP Nationwide Inpatient Sample (NIS) is a nationwide database of hospital inpatient stays. The NIS is nationally representative of all community hospitals (i.e., short-term, non-Federal, non-rehabilitation hospitals). The NIS is a sample of hospitals and includes all patients from each hospital, regardless of payer. It is drawn from a sampling frame that contains hospitals comprising 90 percent of all discharges in the United States. The vast size of the NIS allows the study of topics at both the national and regional levels for specific subgroups of patients. In addition, NIS data are standardized across years to facilitate ease of use.

### **About HCUP**

HCUP is a family of powerful health care databases, software tools, and products for advancing research. Sponsored by the Agency for Healthcare Research and Quality (AHRQ), HCUP includes the largest all-payer encounter-level collection of longitudinal health care data (inpatient, ambulatory surgery, and emergency department) in the United States, beginning in 1988. HCUP is a Federal-State-Industry Partnership that brings together the data collection efforts of many organizations—such as State data organizations, hospital associations, private data organizations, and the Federal government—to create a national information resource.

For more information about HCUP, visit <http://www.hcup-us.ahrq.gov/>.

HCUP would not be possible without the contributions of the following data collection Partners from across the United States:

**Arizona** Department of Health Services  
**Arkansas** Department of Health & Human Services  
**California** Office of Statewide Health Planning & Development  
**Colorado** Health & Hospital Association  
**Connecticut** Integrated Health Information (Chime, Inc.)

**Florida** Agency for Health Care Administration  
**Georgia** GHA: An Association of Hospitals & Health Systems  
**Hawaii** Health Information Corporation  
**Illinois** Health Care Cost Containment Council and Department of Public Health  
**Indiana** Hospital&Health Association  
**Iowa** Hospital Association  
**Kansas** Hospital Association  
**Kentucky** Cabinet for Health and Family Services  
**Maryland** Health Services Cost Review Commission  
**Massachusetts** Division of Health Care Finance and Policy  
**Michigan** Health & Hospital Association  
**Minnesota** Hospital Association  
**Missouri** Hospital Industry Data Institute  
**Nebraska** Hospital Association  
**Nevada** Division of Health Care Financing and Policy, Department of Human Resources  
**New Hampshire** Department of Health & Human Services  
**New Jersey** Department of Health & Senior Services  
**New York** State Department of Health  
**North Carolina** Department of Health and Human Services  
**Ohio** Hospital Association  
**Oregon** Office for Oregon Health Policy and Research and Oregon Association of Hospitals and Health Systems  
**Rhode Island** Department of Health  
**South Carolina** State Budget & Control Board  
**South Dakota** Association of Healthcare Organizations  
**Tennessee** Hospital Association  
**Texas** Department of State Health Services  
**Utah** Department of Health  
**Vermont** Association of Hospitals and Health Systems  
**Virginia** Health Information  
**Washington** State Department of Health  
**West Virginia** Health Care Authority  
**Wisconsin** Department of Health & Family Services

For additional HCUP statistics, visit HCUPnet, our interactive query system at [www.hcup.ahrq.gov](http://www.hcup.ahrq.gov).

## References

For a detailed description of HCUP and more information on the design of the NIS and methods to calculate estimates, please refer to the following publications:

Steiner, C., Elixhauser, A., Schnaier, J. The Healthcare Cost and Utilization Project: An Overview. *Effective Clinical Practice* 5(3):143–51, 2002.

*Design of the HCUP Nationwide Inpatient Sample, 2004*. Online. August 8, 2006. U.S. Agency for Healthcare Research and Quality. [http://www.hcup-us.ahrq.gov/db/nation/nis/reports/NIS\\_2004\\_Design\\_Report.pdf](http://www.hcup-us.ahrq.gov/db/nation/nis/reports/NIS_2004_Design_Report.pdf)

Houchens, R., Elixhauser, A. *Final Report on Calculating Nationwide Inpatient Sample (NIS) Variances, 2001*. HCUP Methods Series Report #2003-2. Online. June 2005 (revised June 6, 2005). U.S. Agency for Healthcare Research and Quality. <http://www.hcup-us.ahrq.gov/reports/CalculatingNISVariances200106092005.pdf>

Houchens RL, Elixhauser A. *Using the HCUP Nationwide Inpatient Sample to Estimate Trends*. (Updated for 1988-2004). HCUP Methods Series Report #2006-05 Online. August 18, 2006. U.S. Agency for Healthcare Research and Quality. [http://www.hcup-us.ahrq.gov/reports/2006\\_05\\_NISTrendsReport\\_1988-2004.pdf](http://www.hcup-us.ahrq.gov/reports/2006_05_NISTrendsReport_1988-2004.pdf)

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AHRQ welcomes questions and comments from readers of this publication who are interested in obtaining more information about access, cost, use, financing, and quality of health care in the United States. We also invite you to tell us how you are using this Statistical Brief and other HCUP data and tools, and to share suggestions on how HCUP products might be enhanced to further meet your needs. Please e-mail us at [hcp@ahrq.gov](mailto:hcp@ahrq.gov) or send a letter to the address below:

Irene Fraser, Ph.D., Director  
Center for Delivery, Organization, and Markets  
Agency for Healthcare Research and Quality  
540 Gaither Road  
Rockville, MD 20850

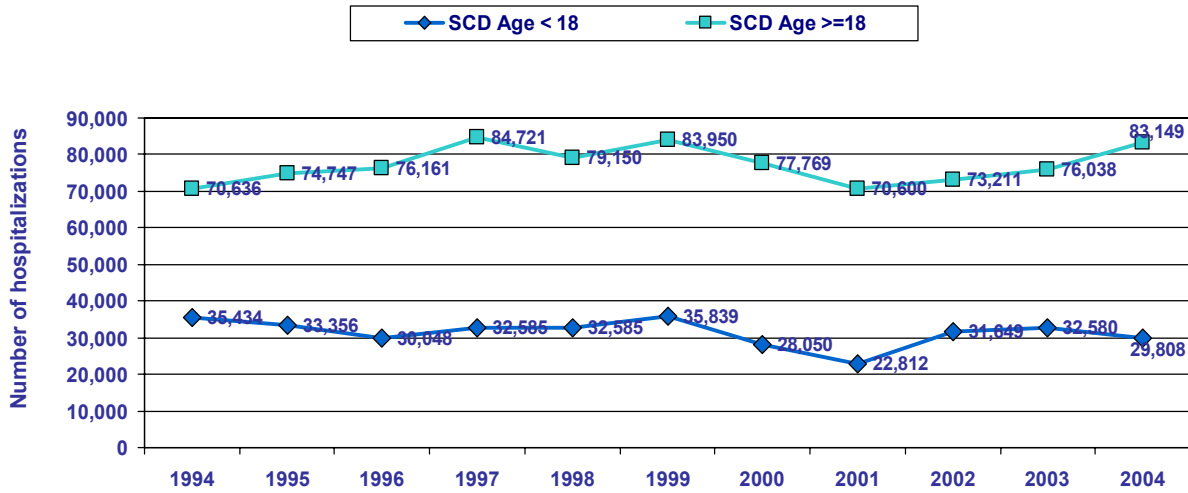
**Table 1. Patient and hospital characteristics associated with hospitalizations for sickle cell disease, 2004**

		Principal diagnosis of sickle cell disease	All-listed diagnosis of sickle cell disease
All discharges		78,524 (100%)	113,098 (100%)
Age group	<1	688 (1.0%)	2,087 (1.8%)
	1-17	18,621 (24.0%)	27,721 (24.5%)
	18-44	52,101 (66.0%)	69,902 (61.8%)
	45-64	6,730 (8.5%)	11,789 (10.4%)
	65-84	278 (0.5%)	1,376 (1.2%)
Sex	Male	36,838 (47%)	50,502 (45%)
	Female	41,359 (53.0%)	62,213 (55%)
In-hospital mortality (%)		0.30 (238 deaths)	0.70 (746 deaths)
Admitted through ED (%)		78.1%	72.8%
Average LOS (days)		5.4	5.5
Median income for zip code	Low (\$0-35,999)	43,068 (54.9%)	61,365 (54.3%)
	Not low (\$36,000+)	34,066 (43.4%)	49,881 (44.1%)
Residence	Large metro	54,675 (69.6%)	79,513 (70.3%)
	Small metro	17,198 (21.9%)	24,349 (21.5%)
	Nonmetro	6,408 (8.2%)	8,896 (7.9%)
Hospital Teaching status	Nonteaching	27,309 (35.0%)	37,861 (33.5%)
	Teaching	51,214 (65.0%)	75,236 (66.5%)
Hospital Location	Non-metropolitan	5,050 (6.0%)	6,620 (5.9%)
	Metropolitan	73,473 (94.0%)	106,477 (94.1%)
U.S region (rate per 10,000 black population)	Northeast	30.7	44.1
	Midwest	26.5	36.4
	South	19.3	28.3
	West	18.1	26.2

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample, 2004.



**Figure 1. Nationwide hospitalizations for children and adults with sickle cell disease (SCD), 1994–2004**

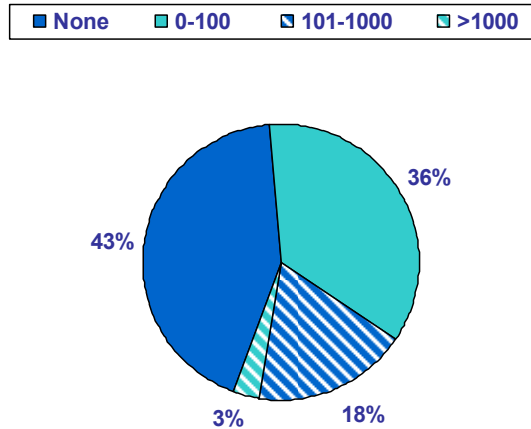


Note: Number of hospitalizations includes all-listed sickle cell disease diagnoses.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample



**Figure 2. Percentage of hospitals with none, 0–100, 101–1000 or >1000 SCD hospitalizations, 2004**

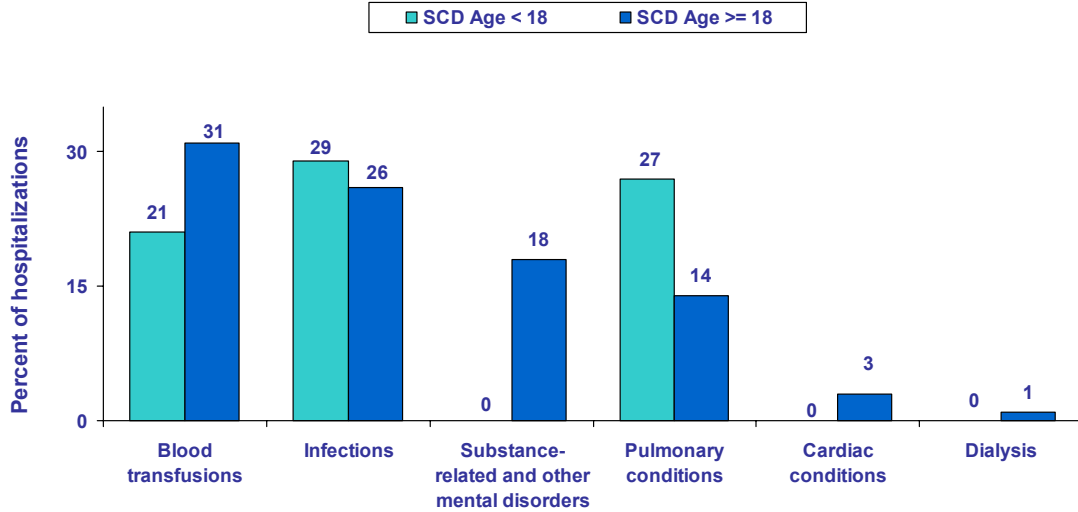


Note: Number of hospitalizations includes all-listed sickle cell disease diagnoses.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample



**Figure 3. Selected conditions and procedures associated with principal diagnosis of sickle cell disease**

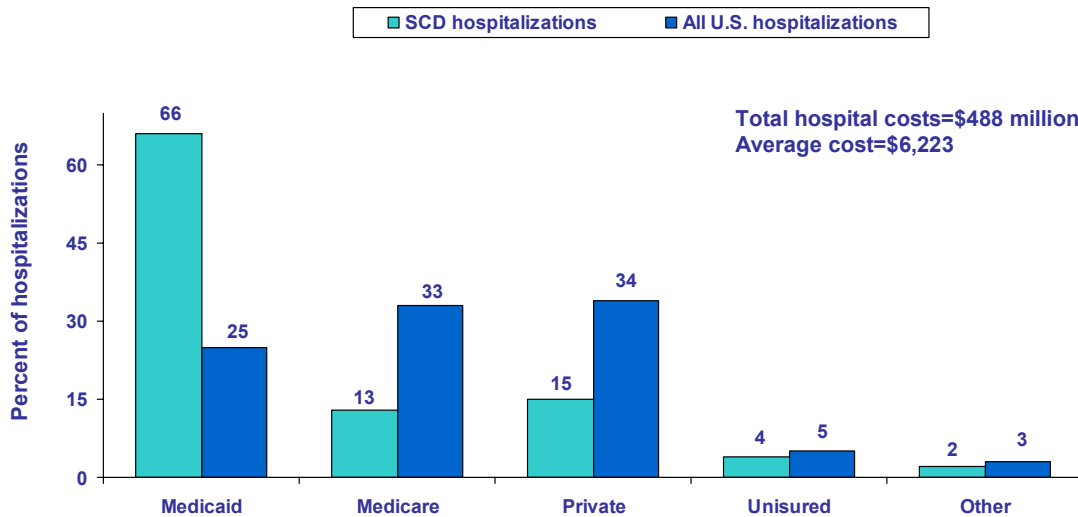


Note: Number of hospitalizations includes principal sickle cell disease diagnoses.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample



**Figure 4. Primary payer for SCD hospitalizations, 2004**



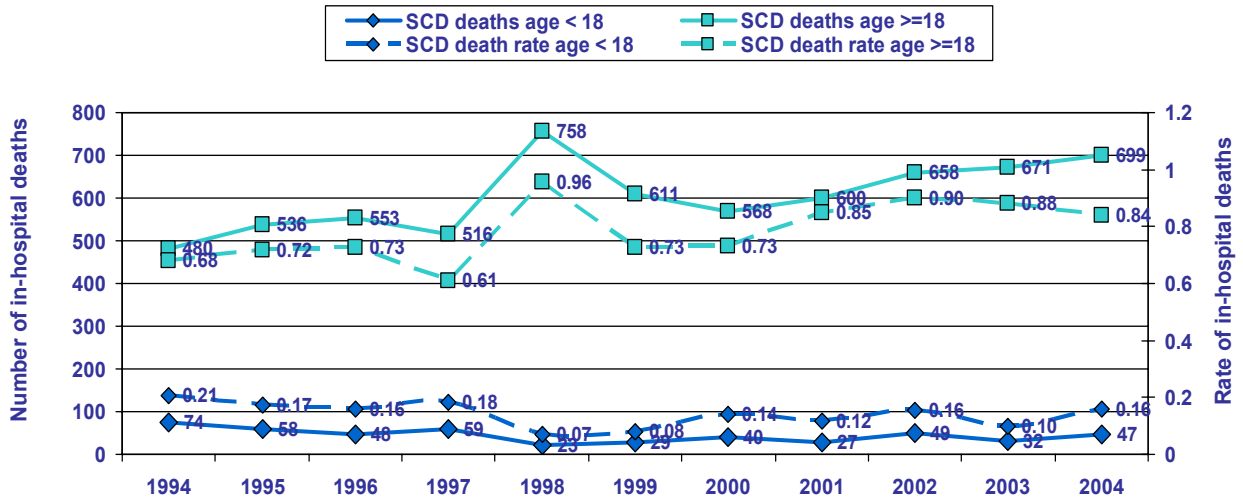
Note: Number of hospitalizations includes all-listed sickle cell disease diagnoses.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample





**Figure 5. In-hospital deaths and death rate for hospitalizations with SCD in children and adults, 1994–2004**

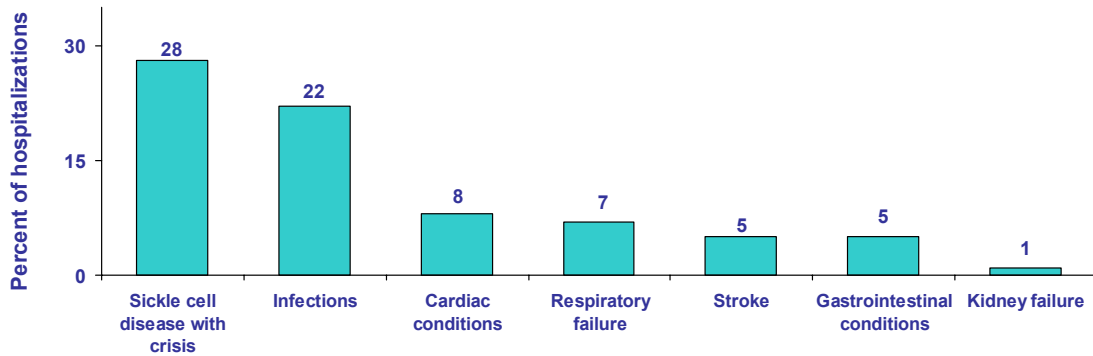


Note: Number of hospitalizations includes all-listed sickle cell disease diagnoses.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample



**Figure 6. Principal diagnosis associated with in-hospital death for hospital stays involving sickle cell disease, 2004**



Note: Number of hospitalizations includes all-listed sickle cell disease diagnoses.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample