Published in final edited form as:

Pediatr Blood Cancer. 2011 March ; 56(3): 413–419. doi:10.1002/pbc.22792.

Sickle Cell Disease in California: A Population-Based Description of Emergency Department Utilization

Julie A. Wolfson, MD, MSHS¹, Sheree M. Schrager, MS, PhD², Thomas D. Coates, MD^{1,3,5}, and Michele D. Kipke, PhD^{2,3,4}

¹Division of Pediatric Hematology-Oncology, Childrens Hospital Los Angeles

²Community, Health Outcomes, and Intervention Research Program, The Saban Research Institute, Childrens Hospital Los Angeles

³Department of Pediatrics, Keck School of Medicine, University of Southern California

⁴Department of Preventive Medicine, Keck School of Medicine, University of Southern California

⁵Department of Pathology, Keck School of Medicine, University of Southern California

Abstract

Background—Acute and chronic clinical manifestations of sickle cell disease (SCD) lead to significant healthcare utilization, especially of the emergency department (ED). Limited population-level data are available in SCD with the ability to connect patients to visits, leaving us with minimal description of utilization patterns.

Procedure—Using ED discharge data with links between patients and visits, we sought to describe the California SCD population and its ED utilization patterns across facilities. Non-public California Office of Statewide Health Planning and Development data employ unique patient identifiers, linking patients and visits.

Results—SCD patients of all ages are heavily reliant on Medicaid (46%). The majority of SCD Californians visit an ED more than once during a year (69%), but only a minority use more than one facility during a year (34%). However, adults with SCD have multiple visits and utilize multiple EDs in higher proportions than do children (72% vs. 60% and 40% vs. 21%, respectively). A higher proportion of visits to the ED are made by SCD adults, but a higher proportion of visits by children result in hospital admission. Uninsured adults outnumber uninsured children (16% vs. 5%).

Conclusions—ED utilization by the California SCD population is described on a population level. Utilization patterns by adults point towards increased utilization in the population no longer eligible for Title V pediatric coverage for their disease. Further investigation using population-level socioeconomic and geographic correlates is warranted to evaluate the factors leading to ED utilization in SCD.

Keywords

sickle cell disease; health services; utilization

Correspondence: Julie A. Wolfson, M.D., M.S.H.S., Pediatric Hematopoietic Stem Cell Transplantation, City of Hope National Medical Center, 1500 East Duarte Road, Machris 3, Duarte, CA 91010-3000, Phone: 626-301-8442, Fax: 626-256-8723, jwolfson@coh.org, jwolfson@post.harvard.edu.

The authors have no relevant conflicts of interest to disclose.

Introduction

Sickle cell disease (SCD) consists of a variety of inherited hemoglobinopathies, and in the setting of hypoxic, acidotic or dehydrated conditions results in abnormal, 'sickle-shaped' red blood cells [1]. Clinically, this results in a range of acute and chronic manifestations that include not only pain syndromes and related issues but also hemolytic anemia and its sequelae [2]. Furthermore, there are a multitude of major organ complications and sequelae; in addition to a risk of sudden death, patients grapple with multi-system organ complications including cardiovascular, pulmonary, cerebrovascular, renal, hepatobiliary, skin and developmental effects [2–3]. With the introduction of newborn screening and new and evolving evidence to manage acute sequelae, through the last five decades SCD has effectively shifted from a disease of children and young adults to a chronic disease of middle age [4–7]. In light of being both an acute and chronic disease, SCD is associated with significant pediatric and adult health care utilization [8–11]. In particular, high rates of emergency department (ED) utilization have been reported in single- and multi-institutional studies [8,11–16].

However, population-based data in SCD patients [17–19], especially in ED utilization [17], are quite limited. Most data sources are from single comprehensive care centers [8,13,20–27] or a conglomeration of these centers [15,19]; such data only describe patients who are already cared for at those centers. On the other hand, some publicly available data sets include a broader sample of sickle-cell patients, but these data generally lack patient identifiers such as social security number, medical record number, or a similar means by which to identify which ED visits belong to the same patient, or whether the same patient goes to one ED one day and a different ED the next [16,28–31]. Large hospital discharge surveys have been used to investigate utilization [10,23,28,32], relying on statistical methods and sampling weights in order to extrapolate to the entire population; however, this approach may be less appropriate in rare diseases such as SCD compared to more common diseases such as diabetes or asthma [33–34]. Furthermore, previous utilization studies in SCD have been limited to single insurance payers [11,14,35–39], and may evaluate only adult [13,31,40] or pediatric [20,26,32,35–37,39,41] populations. Finally, although state-based registries are likely more illustrative, these are limited in availability and may only represent half of a state's SCD population [15]. Without true population-based data, it is virtually impossible to know how often patients are using care, much less where they are going.

In the current report we investigated the population of Californians with SCD who use the ED and to describe their utilization of the ED. With the passage of Assembly Bill 2876, California hospitals and public health departments have access to the non-deidentified patient discharge data, ED discharge data, and ambulatory surgery data from the California Office of Statewide Health Planning and Development (OSHPD) [42]; these data include all discharges from non-federal hospitals. Using these non-public data from 2007, we were able to identify and track patients across multiple institutions and across time by means of patient-specific identifiers.

Methods

Data source

Data used for the current study were obtained from OSHPD; available databases included all California hospital discharges and ED visits from non-federal hospitals in the year 2007. The original data source comprises one year of ED visits to California hospitals from January to December 2007 (n=8,786,265) and one year of hospital discharges from January to December 2007 (n=4,012,775). The ED discharge dataset includes all ED visits in which

the disposition did not result in admission to the same hospital (i.e., admission to different hospital or discharge home or to other location). The inpatient dataset includes all ED visits that resulted in admission to the same hospital. Together, they represent all ED visits in OSHPD with no duplicate ED visits between the two datasets. In addition to administrative and demographic data, International Classification of Diseases, 9th Revision, Clinical Modification (hereafter “ICD-9”) codes are included for a primary and up to 24 secondary additional diagnoses. Visit-level OSHPD data are identified by record linkage numbers derived from the patient’s Social Security number, allowing separate visits by one patient to be linked to that patient.

Data selection

A three-step procedure was used to derive the visit-level data used in the present analysis. First, patients were selected from the original data sets described above on the basis of a relevant ICD-9 code pertaining to all variants of Sickle Cell Disease (282.60–282.69 and 282.41–282.42) in any of the primary or secondary diagnoses. Visits coded with only sickle-cell trait (282.50–282.59) and non-sickle-related thalassemia diagnoses were excluded. Second, all record linkage numbers corresponding to visits obtained in the first selection step were identified and all additional ED visits in 2007 identified by these record linkage numbers were added to the data set. This permitted us to investigate the total number of ED encounters from patients known to have SCD regardless of whether the condition was coded at the time of each encounter. Finally, the visits originating from OSHPD inpatient data were subjected to an additional selection on admission route: only inpatient visits for which the ED encounter resulted in admission to the same hospital were included. This ensured that there would be no duplication of visits from the ED discharge data, as both data sets potentially included visits from patients who were discharged from one facility’s ED and admitted directly to another hospital. This three-step procedure yielded 16,638 visits, encompassing all statewide ED encounters in 2007 from identified SCD patients as well as all SCD-related ED visits from unidentified individuals. Additionally, a patient-level data set (n=2,979) was created by removing the 840 visits without a record linkage number and aggregating the demographic information and counts of visits, admissions, and facilities used.

Visit-level measures

Measures retained from the original OSHPD data include: age in years; sex (male or female); ethnicity (Hispanic or non-Hispanic); and disposition (admit or discharge). Race (American Indian/Alaska Native, Black/African American, White, Asian/Pacific Islander, or other race) was mostly retained, with the exception that Asian race was coded separately from Hawaiian and Pacific Islander races in the ED discharge data but not the inpatient data. Thus, these groups were combined into a single category in the present analysis.

Additional characteristics were derived using information in the OSHPD data. Categorical indicators of age were derived by initially recoding patients into children < 21 years or adults ≥ 21 years, and then into six categories: 0–10, 11–20, 21–30, 31–40, 41–50, or 51+). Expected source of payment was reduced in the following manner: Medicare including Part A, Part B or HMO, Medicaid/Medi-Cal, other government or indigent source, or other source, were all further combined into public payer; HMO, PPO, POS, EPO, automobile insurance, Blue Cross/Blue Shield, worker’s compensation, and commercial insurance were further combined into private payer; self pay patients were considered uninsured. Finally, based on the county of residence, geographic residence was combined into Los Angeles County, Southern California (excluding Los Angeles County), and Northern California. In order to describe the frequency of clinical conditions presenting to the ED in our population,

we summed the frequencies of ICD-9 codes present in the primary and up to five secondary diagnoses, excluding ICD-9 codes used as inclusion criteria.

Patient-level measures

Aggregating demographic information yielded the following measures: Total number of ED visits, total number of admissions from the ED, total number of facilities utilized, mean age, sex; race, ethnicity, presence of SCD diagnosis during any visit, geographic residence (based on most recent county of residence). Additionally, a binary indicator of multiple facility utilization (1 if the total number of facilities was greater than 1; 0 otherwise) was created. Mean age was also recoded into the same binary and categorical measures as in the visit-level data. All borderline ages with trailing decimals were rounded into the higher age category, as the existence of a remainder indicates that the patient had an ED encounter at the older age sometime in 2007. For example, a patient with two visits, one at age 20 and one at age 21 would have a mean age of 20.5. We elected to group such patients in the 21–30 age category in the patient-level data because they would have aged out of pediatric comprehensive care during 2007, as California's Title V coverage is guaranteed only until a child's 21st birthday. Similar logic applies to other borderline age categorizations. In order to describe insurance coverage by individual rather than simply by visit, we chose the last visit of the year to describe the patient's payer for that year. With this method, if a patient lost eligibility and/or coverage during the year, then (s)he was described as self-pay even if the first visit of the year was expected to be paid for by Medi-Cal; thus, if a patient lost eligibility for Title V coverage during the year, the last visit of the year would reflect that occurrence.

Data inconsistencies

Aggregating from the visit to the patient level revealed inconsistencies in the demographic variables. Specifically, the same patients (as identified by record linkage number) were occasionally associated with conflicting indicators of sex, race, and/or ethnicity. Patients with inconsistent sex ($n = 11$) or race ($n = 277$) were assigned to "unknown/inconsistent" sex or race, respectively. Patients with inconsistent ethnicity ($n = 225$) were further investigated. Patients who had any visit in which they were identified as Hispanic were assigned Hispanic ethnicity, whereas those with consistently unknown or missing ethnicity, and those with inconsistencies between unknown and non-Hispanic, were assigned to "unknown/inconsistent" ethnicity; this was due to the fact that designation as Hispanic required intent whereas non-Hispanic or unknown could be passive designations. Once all inconsistencies had been examined and the person-level data set was finalized, events in the visit-level data set were reassigned sex, race, and/or ethnicity based on the revisions made to the person-level data (where visits were linked to patients by record linkage number).

Analysis

SPSS and Stata 10 were used for all statistical analyses. Chi-square tests and t-tests were performed to determine the statistical significance of differences between groups of adults and children as well as between males and females. The AB2876 request for OSHPD data was approved by the Committee for Protection of Human Subjects for the California Health and Human Services Agency (state IRB). This study was also approved by the Committee on Clinical Investigations (Institutional Review Board) of Childrens Hospital Los Angeles.

Results

Patient-level descriptive statistics are presented in Table I. The average age of SCD patients was 30.53 years old ($SD = 16.37$); the mean age of men in our sample was 28.6 years, and the mean age of women was 31.9 years ($t(2966) = -5.375$, $p < .001$). The sample itself was

comprised mostly of adults (72%), females (58%), African Americans (81%), non-Hispanics (86%), and California residents (97%). The majority of patients expected to pay for their visits with a form of public insurance (62%), with a smaller proportion covered by private insurance (25%) or uninsured (13%).

Patients averaged over five ED visits per year ($M = 5.30$, $SD = 12.06$); there was no significant difference in number of visits by gender ($t(2966) = -.634$, ns). Of these visits, nearly two per year resulted in admission to the hospital ($M = 1.85$, $SD = 3.06$). The distribution of visits was uneven among patients: Although most patients (96%) had fewer than 20 visits throughout the year, the remaining patients comprised a disproportionate number of visits in the sample. Indeed, 5% of the sample ($n = 154$) comprised 40% of the visits. A majority (69%) of the cohort had more than one visit in the year; furthermore, a third of the sample visited more than one hospital facility, although on average the number of facilities used was low ($M = 1.68$, $SD = 1.77$).

Visit-level descriptive statistics are presented in Table II. Similar to the person-level data, the majority of statewide ED encounters in 2007 came from adults (82%), females (58%), African Americans (74%), and non-Hispanics (75%). Additionally, most visits resulted in discharge (65%), and patients expected to pay for the majority of visits with a form of public insurance (72%). There were 305 facilities in the state visited by patients with SCD, with a mean number of visits per facility of 54.55 ($SD = 87.70$; range = 1 to 598).

The majority of the burden of SCD care at the ED fell on a public source (72%), with half (49%) falling on Medicaid, less on Medicare (18%), and the remainder other government programs. A smaller proportion was accounted for by private insurance (17%). Finally, 10% of SCD visits were by the uninsured.

A summary of the most common ICD-9 codes by age group is presented in Table III. The overall most common diagnoses associated with SCD patients visiting the ED were hypertension, limb pain and dehydration. In the adult (≥ 21 years) population, hypertension, tobacco use and limb pain emerged most frequently; however, fever, limb pain and pneumonia were more frequently present in children (< 21 years).

We also conducted investigations into whether ED populations or utilization patterns differed among children as compared to adults. We used the cutoff of 21 years of age to denote adults, as these patients have ‘aged out’ of California’s Title V coverage for children with special healthcare needs [43]. As illustrated in Table IV, adults had significantly more ED encounters than children ($t(2606) = 10.95$, $p < 0.001$), yet a smaller proportion was admitted to the hospital following an ED encounter ($\chi^2(1) = 69.80$, $p < 0.001$). Adults on average visited significantly more hospitals than children ($t(2918) = 11.30$, $p < 0.001$) and a higher proportion had multiple visits in the year ($\chi^2(1) = 41.87$, $p < 0.001$). Furthermore, a greater proportion of adults was uninsured, and a lower proportion of adults expected private insurance coverage for their ED care compared to children ($\chi^2(2) = 58.83$, $p < 0.001$). However, there were no significant differences in regional distribution of child and adult SCD patients ($\chi^2(2) = 3.02$, ns).

Discussion

There has been minimal evaluation of ED utilization at the population level [17]. Using state administrative data with access to patient identifiers in order to link visits to patients across both institutions and time, we have been able to briefly describe the California SCD population who visited the ED in one year and the utilization pattern across age groups.

Our demographic description conforms to expectations as a whole [40,44–46]. Our cohort, when compared to recent prevalence estimates [47–48], represents between 50% and 70% of the state's SCD population. Women in this population are older than men [8,45], African-Americans comprise the vast majority of patients, and Hispanic patients account for 8% of the cohort. These results are consistent with the rosters of the state's two largest SCD comprehensive care centers. We do have a significant amount of patients whose race is either unknown or inconsistent between visits. This may be due in part to the mechanism of data collection for race and ethnicity in an administrative database such as OSHPD. There are inconsistencies in race categories between true self-report and administrative databases, especially in non-white populations [49]. At hospital or ED registration, race is usually recorded by a hospital administrative employee who may or may not ask the patient for a self-description of his/her race, and in fact this response may have been chosen on behalf of the patient. Another complication with sociodemographic description based on administrative data is that nearly 5% of Californians in the 2000 census self-designated themselves as belonging to two or more races [50], yet the data collection form is confined to selecting one historically used race category per patient [51].

Comparing the insurance status of our patients to Californians as a whole, there were a comparable proportion of uninsured patients; uninsured patients represented 13% of our cohort and 13% of the general California population is uninsured [52]. However, the overwhelming majority of our cohort, and of SCD patients in general, belongs to a minority group [53]. If we compare the insurance status of our cohort to California minority populations, there appears to be a comparable proportion of uninsured African Americans (11%). However, whereas only 16% of general Californians are covered by Medi-Cal (Medicaid), 46% of patients in our cohort relied on Medi-Cal. This differential persists when comparing our cohort to African American (29%) and Latino (27%) Californians who rely on Medi-Cal [52].

As our cohort ages, patients comprise an increasing proportion of the visits and patient population. Similarly, the mean number of visits per year is significantly higher in adults than in children (6.29 vs. 2.78). Furthermore, the disposition of children and adults is significantly different; a smaller proportion of adult ED visits resulted in inpatient admission than did pediatric ED visits (34% vs. 42%). Center-based studies of disposition from the ED show over 60% of children and 33% of adults being admitted [8,54]. Our results, tracking patients across multiple facilities, show a similar differential between adult and pediatric disposition from the ED.

Not only multiple facility use, but also frequency of utilization have been key outcome measures in ED utilization [55]. Within our cohort, 21% of children and 40% of adults used more than one facility, with the remainder using only one facility for their ED visits. Similarly, 60% of children and 72% of adults visited the ED multiple times during the year, with the remainder having only one ED visit during the year. The utilization patterns in our cohort may indicate that a larger proportion of adults are 'shopping around' for care, without a single facility that they visit repeatedly, whereas children are more often tied to a single facility. This interpretation is consistent with the notion that children more likely have a medical home, or a health care setting in which care is coordinated, comprehensive and accessible, and directed by a physician, whether a generalist or subspecialist is more appropriate for the patient's condition(s) [56–58]; medical homes are known to decrease ED utilization [59]. On the other hand, this result may simply indicate a wider variety of facilities willing to see adults and a narrower range of facilities comfortable seeing children in the emergency setting. Further investigation into outpatient utilization patterns is necessary to compare the prevalence of a medical home in these populations.

Five percent of our cohort accounted for 40% of the total number of SCD ED visits in 2007. This is consistent with previous investigations of single payers and states [14,29,38] in which this highest tier of hospital utilizers was found to be sociodemographically similar to the remainder of health care utilizers but to differ diagnostically; while most high frequency utilization tapered off with time, a subset remained persistent [38]. In order to further characterize factors leading to extreme utilization in our cohort, this top tier of users warrants further investigation.

Despite the unique population descriptions that can be gleaned from such broad, linked data, there are inherent limitations to using administrative data. Numerous sites and modes of collection can lead to inconsistencies in reporting. Furthermore, there are limitations to the clinical accuracy of ICD-9 codes, especially in ED data when compared to inpatient problem lists or chart reviews [60–62]. Despite this fact, ICD-9 codes and administrative data are routinely used in clinical, epidemiologic and quality research [61]. In the absence of population-based longitudinal surveillance or population-level data, ICD-9 codes have been used here as a best available surrogate to describe this population and its utilization patterns.

Moreover, 840 visits were not associated with a record linkage number to identify the patient. As a result, although these patients are represented in the visit-level data, they may have been disproportionately excluded from the patient-level data. These patients did not likely have a Social Security number, the basis for deriving the record linkage number. Patients without record linkage numbers may include children who were not assigned a Social Security number at birth, undocumented immigrants, or patients seen in facilities where hospital policy did not compel obtaining Social Security number at registration.

Finally, using one year of ED discharge data limits our findings to those patients who visit the ED in one given year, although half of the SCD population may not do so [11]. Those patients who do not use the ED may have different characteristics and warrant investigation into both payer-level data and community-based case finding strategies in order to facilitate prospective surveillance. Despite the limitations of population-level administrative data, our study is one of very few to explore population-level data for both a pediatric and an adult population in order to evaluate ED utilization in SCD.

In summary, ED utilization by the California SCD population has been described on a population level. A higher proportion of our largely African American cohort relies on Medi-Cal than do African American Californians. Adults with SCD in California are uninsured in higher proportions than children with SCD; they also have multiple ED visits, use multiple facilities and are high-frequency ED utilizers in higher proportions than children. These adult utilization patterns point towards increased utilization in the population that has lost eligibility for Title V coverage for their disease; to better evaluate the premise that higher utilization patterns by age are consistent with lack of a medical home, correlation of population-level inpatient and outpatient utilization patterns is necessary. In order to assess the impact of insurance status and other factors on utilization, added population-level investigation is warranted, with data to account for socioeconomic and geographic characteristics.

Acknowledgments

This project was supported by the Los Angeles Basin Clinical and Translational Science Institute. The authors wish to thank Patricia Ganz MD, Ninez Ponce PhD, David Zingmond MD, PhD, Jack Needleman, PhD and Susan Claster, MD for their contributions to this project. The California Office of Statewide Health Planning and Development provided the data to Childrens Hospital Los Angeles.

References

1. Bunn HF. Pathogenesis and treatment of sickle cell disease. *N Engl J Med*. 1997; 337(11):762–769. [PubMed: 9287233]
2. Ballas SK, Lief S, Benjamin LJ, et al. Definitions of the phenotypic manifestations of sickle cell disease. *American Journal of Hematology*. 2010; 85(1):6–13. [PubMed: 19902523]
3. Orkin, SH.; Nathan, DG.; Ginsburg, D., et al., editors. *Nathan and Oski's Hematology of Infancy and Childhood*. Philadelphia: Saunders Elsevier; 2009.
4. Vichinsky E, Hurst D, Earles A, et al. Newborn screening for sickle cell disease: effect on mortality. *Pediatrics*. 1988; 81(6):749–755. [PubMed: 3368274]
5. Steinberg MH, Barton F, Castro O, 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; 289(13):1645–1651. [PubMed: 12672732]
6. Halasa NB, Shankar SM, Talbot TR, et al. Incidence of invasive pneumococcal disease among individuals with sickle cell disease before and after the introduction of the pneumococcal conjugate vaccine. *Clin Infect Dis*. 2007; 44(11):1428–1433. [PubMed: 17479937]
7. Hagar RW, Vichinsky EP. Major changes in sickle cell disease. *Adv Pediatr*. 2000; 47:249–272. [PubMed: 10959446]
8. Epstein K, Yuen E, Riggio JM, et al. Utilization of the office, hospital and emergency department for adult sickle cell patients: a five-year study. *J Natl Med Assoc*. 2006; 98(7):1109–1113. [PubMed: 16895280]
9. Mvundura M, Amendah D, Kavanagh PL, et al. Health care utilization and expenditures for privately and publicly insured children with sickle cell disease in the United States. *Pediatr Blood Cancer*. 2009; 53(4):642–646. [PubMed: 19492318]
10. Panepinto JA, Brousseau DC, Hillery CA, et al. Variation in hospitalizations and hospital length of stay in children with vaso-occlusive crises in sickle cell disease. *Pediatr Blood Cancer*. 2005; 44(2):182–186. [PubMed: 15390359]
11. Shankar SM, Arbogast PG, Mitchel E, et al. Medical care utilization and mortality in sickle cell disease: a population-based study. *Am J Hematol*. 2005; 80(4):262–270. [PubMed: 16315251]
12. Bilenker JH, Weller WE, Shaffer TJ, et al. The costs of children with sickle cell anemia: preparing for managed care. *J Pediatr Hematol Oncol*. 1998; 20(6):528–533. [PubMed: 9856672]
13. Hand R, Koshy M, Dorn L, et al. Health insurance status and the use of emergency and other outpatient services by adults with sickle cell disease. *Ann Emerg Med*. 1995; 25(2):224–229. [PubMed: 7832351]
14. Kauf TL, Coates TD, Huazhi L, et al. The cost of health care for children and adults with sickle cell disease. *Am J Hematol*. 2009; 84(6):323–327. [PubMed: 19358302]
15. Telfair J, Haque A, Etienne M, et al. Rural/urban differences in access to and utilization of services among people in Alabama with sickle cell disease. *Public Health Rep*. 2003; 118(1):27–36. [PubMed: 12604762]
16. Yusuf HR, Atrash HK, Grosse SD, et al. Emergency department visits made by patients with sickle cell disease: a descriptive study, 1999–2007. *Am J Prev Med*. 2010; 38(4 Suppl):S536–541. [PubMed: 20331955]
17. Brousseau DC, Owens PL, Mosso AL, et al. Acute care utilization and rehospitalizations for sickle cell disease. *JAMA*. 2010; 303(13):1288–1294. [PubMed: 20371788]
18. Fullerton HJ, Adams RJ, Zhao S, et al. Declining stroke rates in Californian children with sickle cell disease. *Blood*. 2004; 104(2):336–339. [PubMed: 15054044]
19. Haque A, Telfair J. Socioeconomic distress and health status: the urban-rural dichotomy of services utilization for people with sickle cell disorder in North Carolina. *J Rural Health*. 2000; 16(1):43–55. [PubMed: 10916314]
20. Frei-Jones MJ, Baxter AL, Rogers ZR, et al. Vaso-occlusive episodes in older children with sickle cell disease: emergency department management and pain assessment. *J Pediatr*. 2008; 152(2):281–285. [PubMed: 18206703]

21. Koshy M, Leikin J, Dorn L, et al. Evaluation and management of sickle cell disease in the emergency department (An 18-year experience): 1974–1992. *Am J Ther.* 1994; 1(4):309–320. [PubMed: 11835105]
22. Loureiro MM, Rozenfeld S, Sa Carvalho M, et al. Factors associated with hospital readmission in sickle cell disease. *BMC Blood Disord.* 2009; 9:2. [PubMed: 19250540]
23. Mayer ML, Konrad TR, Dvorak CC. Hospital resource utilization among patients with sickle cell disease. *J Health Care Poor Underserved.* 2003; 14(1):122–135. [PubMed: 12613072]
24. Nietert PJ, Abboud MR, Zoller JS, et al. Costs, charges, and reimbursements for persons with sickle cell disease. *J Pediatr Hematol Oncol.* 1999; 21(5):389–396. [PubMed: 10524452]
25. Okpala I, Thomas V, Westerdale N, et al. The comprehensiveness care of sickle cell disease. *Eur J Haematol.* 2002; 68(3):157–162. [PubMed: 12068796]
26. Teach SJ, Lillis KA, Grossi M. Compliance with penicillin prophylaxis in patients with sickle cell disease. *Arch Pediatr Adolesc Med.* 1998; 152(3):274–278. [PubMed: 9529466]
27. Yang YM, Shah AK, Watson M, et al. Comparison of costs to the health sector of comprehensive and episodic health care for sickle cell disease patients. *Public Health Rep.* 1995; 110(1):80–86. [PubMed: 7838948]
28. Davis H, Gergen PJ, Moore RM Jr. Geographic differences in mortality of young children with sickle cell disease in the United States. *Public Health Rep.* 1997; 112(1):52–58. [PubMed: 9018289]
29. Lanzkron S, Haywood C Jr, Segal JB, et al. Hospitalization rates and costs of care of patients with sickle-cell anemia in the state of Maryland in the era of hydroxyurea. *Am J Hematol.* 2006; 81(12):927–932. [PubMed: 16924648]
30. Strouse JJ, Jordan LC, Lanzkron S, et al. The excess burden of stroke in hospitalized adults with sickle cell disease. *Am J Hematol.* 2009; 84(9):548–552. [PubMed: 19623672]
31. Woods K, Karrison T, Koshy M, et al. Hospital utilization patterns and costs for adult sickle cell patients in Illinois. *Public Health Rep.* 1997; 112(1):44–51. [PubMed: 9018288]
32. Ellison AM, Bauchner H. Socioeconomic status and length of hospital stay in children with vaso-occlusive crises of sickle cell disease. *J Natl Med Assoc.* 2007; 99(3):192–196. [PubMed: 17393942]
33. Steiner C, Elixhauser A, Schnaier J. The healthcare cost and utilization project: an overview. *Eff Clin Pract.* 2002; 5(3):143–151. [PubMed: 12088294]
34. Machlin S, Cohen J, Elixhauser A, et al. Sensitivity of household reported medical conditions in the medical expenditure panel survey. *Med Care.* 2009; 47(6):618–625. [PubMed: 19433993]
35. Kuhlthau K, Ferris TG, Beal AC, et al. Who cares for medicaid-enrolled children with chronic conditions? *Pediatrics.* 2001; 108(4):906–912. [PubMed: 11581443]
36. Shankar SM, Arbogast PG, Mitchel E, et al. Impact of proximity to comprehensive sickle cell center on utilization of healthcare services among children with sickle cell disease. *Pediatr Blood Cancer.* 2008; 50(1):66–71. [PubMed: 16998856]
37. Sox CM, Cooper WO, Koepsell TD, et al. Provision of pneumococcal prophylaxis for publicly insured children with sickle cell disease. *JAMA.* 2003; 290(8):1057–1061. [PubMed: 12941678]
38. Carroll CP, Haywood C Jr, Fagan P, et al. The course and correlates of high hospital utilization in sickle cell disease: Evidence from a large, urban Medicaid managed care organization. *Am J Hematol.* 2009; 84(10):666–670. [PubMed: 19743465]
39. Raphael JL, Dietrich CL, Whitmire D, et al. Healthcare utilization and expenditures for low income children with sickle cell disease. *Pediatr Blood Cancer.* 2009; 52(2):263–267. [PubMed: 18837428]
40. Epstein AM, Ayanian JZ. Racial disparities in medical care. *N Engl J Med.* 2001; 344(19):1471–1473. [PubMed: 11346815]
41. Panepinto JA, O'Mahar KM, DeBaun MR, et al. Health-related quality of life in children with sickle cell disease: child and parent perception. *Br J Haematol.* 2005; 130(3):437–444. [PubMed: 16042695]
42. Office of Statewide Health Planning and Development. State of California; 2009. www.osphd.ca.gov

43. Medi-Cal Facts: California Children's Services & Medi-Cal. Medi-Cal Policy Institute: A Project of the California HealthCare Foundation; 2000. Report
44. Haywood C Jr, Lanzkron S. Sickle cell disease mortality in the United States: Age at death and contributing causes. *ASH Annual Meeting Abstracts*. 2007; 110(11):81.
45. Platt OS, Brambilla DJ, Rosse WF, et al. Mortality in sickle cell disease. Life expectancy and risk factors for early death. *N Engl J Med*. 1994; 330(23):1639–1644. [PubMed: 7993409]
46. Powars DR, Chan LS, Hiti A, et al. Outcome of sickle cell anemia: a 4-decade observational study of 1056 patients. *Medicine (Baltimore)*. 2005; 84(6):363–376. [PubMed: 16267411]
47. Hassell KL. Population estimates of sickle cell disease in the U. S *Am J Prev Med*. 2010; 38(4 Suppl):S512–521.
48. Brousseau DC, Panepinto JA, Nimmer M, et al. The number of people with sickle-cell disease in the United States: national and state estimates. *Am J Hematol*. 2010; 85(1):77–78. [PubMed: 20029951]
49. Boehmer U, Kressin NR, Berlowitz DR, et al. Self-reported vs administrative race/ethnicity data and study results. *Am J Public Health*. 2002; 92(9):1471–1472. [PubMed: 12197976]
50. Jones, NA.; Smith, AS. Census 2000 Brief: US Census Bureau. 2000. The Two or More Races Population: 2000. Report
51. Office of Statewide Health Planning and Development. State of California: CA.gov; 2010. <<http://www.oshpd.ca.gov/>>
52. [Accessed 2009] UCLA Center for Health Policy Research: California Health Interview Survey. 2009. <<http://www.chil.ucla.edu/main/DQ3/geographic.asp>>
53. Lorey FW, Arnopp J, Cunningham GC. Distribution of hemoglobinopathy variants by ethnicity in a multiethnic state. *Genetic Epidemiology*. 1996; 13(5):501–512. [PubMed: 8905396]
54. Frei-Jones MJ, Field JJ, DeBaun MR. Risk factors for hospital readmission within 30 days: a new quality measure for children with sickle cell disease. *Pediatr Blood Cancer*. 2009; 52(4):481–485. [PubMed: 19058209]
55. Zuckerman S, Shen YC. Characteristics of occasional and frequent emergency department users: do insurance coverage and access to care matter? *Med Care*. 2004; 42(2):176–182. [PubMed: 14734955]
56. American Academy of Family Physicians (AAFP), American Academy of Pediatrics (AAP), American College of Physicians (ACP) and American Osteopathic Association (AOA). [Accessed 2010] Joint Principles of the Patient-Centered Medical Home. Mar. 2007 <http://www.acponline.org/running_practice/pcmh/demonstrations/jointprinc_05_17.pdf>
57. Kirschner N, Barr MS. Specialists/Subspecialists and the patient-centered medical home. *Chest*. 2010; 137(1):200–204. [PubMed: 19505988]
58. Starfield B, Shi L. The medical home, access to care, and insurance: A review of evidence. *Pediatrics*. 2004; 113(5):1493–1498. [PubMed: 15121917]
59. Cooley WC, McAllister JW, Sherrieb K, et al. Improved outcomes associated with medical home implementation in pediatric primary care. *Pediatrics*. 2009; 124(1):358–364. [PubMed: 19564320]
60. Gorelick MH, Knight S, Alessandrini EA, et al. Lack of agreement in pediatric emergency department discharge diagnoses from clinical and administrative data sources. *Acad Emerg Med*. 2007; 14(7):646–652. [PubMed: 17554009]
61. Iezzoni LI. Assessing quality using administrative data. *Annals of Internal Medicine*. 1997; 127(8 Part 2):666–674. [PubMed: 9382378]
62. Kahn LH, Blustein J, Arons RR, et al. The validity of hospital administrative data in monitoring variations in breast cancer surgery. *Am J Public Health*. 1996; 86(2):243–245. [PubMed: 8633744]

Table I

Patient-level descriptive statistics (total N = 2,979 identified patients).

Variable	Range	Mean (SD)
Age (Years)	0 to 96	30.53 (16.37)
Number of Visits	1 to 311	5.30 (12.06)
Number of Admissions	0 to 34	1.85 (3.06)
Number of Facilities Utilized	1 to 37	1.68 (1.77)

Variable	Categories	N (%)
Age	Child (0–20)	841 (28)
	0–10	347 (12)
	11–20	494 (17)
	Adult (21+)	2138 (72)
	21–30	739 (25)
	31–40	576 (19)
	41–50	479 (16)
	51+	344 (12)
Sex	Male	1244 (42)
	Female	1724 (58)
	Unknown/Inconsistent	11 (0)
Race	Black/African American	2402 (81)
	Other Known Race	271 (9)
	Asian/Pacific Islander	16 (1)
	White	144 (5)
	Other Race	111 (4)
	Unknown/Inconsistent	306 (10)
Ethnicity	Hispanic	235 (8)
	Non-Hispanic	2557 (86)
	Unknown/Inconsistent	187 (6)
Health Insurance	Public	1858 (62)
	Private	730 (25)
	Uninsured	391 (13)
Multiple Visits	Made 1 ED Visit	926 (31)
	Made >1 ED Visit	2053 (69)
Multiple Facility Utilization	Utilized 1 Hospital Facility	1954 (66)
	Only One Visit	926 (31)
	Multiple Visits to Same Facility	1028 (35)
	Utilized >1 Hospital Facility	1025 (34)

Table II

Visit-level descriptive statistics (total N = 16,638 visits).

Variable	Categories	N (%)
Age	Child (0–20)	2970 (18)
	0–10	1185 (7)
	11–20	1785 (11)
	Adult (21+)	13668 (82)
	21–30	5498 (33)
	31–40	3380 (20)
	41–50	3183 (19)
	51+	1607 (10)
Sex	Male	6917 (42)
	Female	9669 (58)
	Unknown/Inconsistent	52 (0)
Race	Black/African American	12235 (74)
	Other Known Race	920 (6)
	Asian/Pacific Islander	33 (0)
	White	524 (3)
	Other Race	363 (2)
	Unknown/Inconsistent	3483 (21)
Ethnicity	Hispanic	2292 (14)
	Non-Hispanic	12427 (75)
	Unknown/Inconsistent	1919 (12)
Expected Source of Payment	Public Pay	11992 (72)
	Private Pay	2931 (18)
	Self Pay (No Insurance)	1714 (10)
	Unknown	1 (0)
Disposition	Admit	5843 (35)
	Discharge	10795 (65)
Geographic Residence	Los Angeles County (LAC)	6351 (38)
	Southern California (not LAC) *	3345 (20)
	Northern California **	6664 (40)
	Out-of-state/Missing/Unknown/Homeless	278 (2)

* Southern California (not LAC) Counties: San Diego, Orange, Riverside, San Bernardino, Ventura, Kern, Santa Barbara, San Luis Obispo and Imperial;

** Northern California: Remaining 48 counties

Table III

Most common ICD-9 codes.

Clinical Diagnosis* (ICD-9 code)	Number (%) of visits
<i>Entire cohort (n = 16,638 visits)</i>	
Hypertension (401.9)	958 (5.76)
Limb pain (729.5)	810 (4.87)
Dehydration (276.51)	765 (4.60)
Tobacco use (305.1)	716 (4.30)
Asthma (493.90)	655 (3.94)
Pneumonia (486)	647 (3.89)
Backache (724.5)	566 (3.40)
Urinary Tract Infection (599.0)	547 (3.29)
<i>Adults ≥ 21 (n = 13,669)</i>	
Hypertension (401.9)	931 (6.81)
Tobacco Use (305.1)	674 (4.93)
Limb Pain (729.5)	646 (4.73)
Dehydration (276.51)	619 (4.53)
Asthma (493.90)	541 (3.96)
Pneumonia (486)	485 (3.55)
Diabetes Mellitus (250.00)	478 (3.50)
Urinary Tract Infection (599.0)	477 (3.49)
<i>Children < 21 (n = 2,970 visits)</i>	
Fever (780.6)	301 (10.13)
Limb pain (729.5)	164 (5.52)
Pneumonia (486)	162 (5.45)
Dehydration (276.51)	146 (4.92)
Acute Upper Respiratory Infection (465.9)	145 (4.88)
Asthma (493.90)	114 (3.84)
Backache (724.5)	112 (3.77)
Abdominal Pain (789.00)	107 (3.60)
Acute Chest Syndrome (517.3)	107 (3.60)

* All diagnoses abbreviated. Table excludes ICD-9 codes used as inclusion criteria for selection into the study sample.

Table IV

Outcomes by age of patient.

Outcome	Child (< 21)	Adult (≥ 21)
Mean (SD) Number of Visits ***	2.78 (3.12)	6.29 (13.98)
Mean (SD) Number of Facilities Utilized ***	1.28 (0.67)	1.84 (2.03)
<i>Disposition ***</i>		
Number (%) of Visits Resulting in Discharge	1730 (58)	9065 (66)
Number (%) of Visits Resulting in Admission	1240 (42)	4603 (34)
<i>Multiple Utilization ***</i>		
Number (%) Who Made Only 1 Visit	335 (40)	591 (28)
Number (%) Who Made >1 Visit	506 (60)	1547 (72)
Number (%) Who Utilized 1 Hospital Facility	667 (79)	1287 (60)
Number (%) Who Utilized >1 Hospital Facility	174 (21)	851 (40)
<i>Expected Source of Payment ***</i>		
Number (%) of Public-Pay Visits	2138 (72)	9854 (72)
Number (%) of Private-Pay Visits	666 (22)	2265 (17)
Number (%) of Uninsured Visits	166 (6)	1548 (11)
Number (%) of Public-Pay Patients	577 (69)	1281 (60)
Number (%) of Private-Pay Patients	217 (26)	513 (24)
Number (%) of Uninsured Patients	47 (5)	344 (16)
<i>Geographic Residence (ns)</i>		
Number (%) in Los Angeles County	298 (36)	794 (38)
Number (%) in Southern California	205 (25)	530 (26)
Number (%) in Northern California	328 (40)	747 (36)

Significant difference between adults and children, $p < 0.001$; (ns) No significant difference between adults and children.