

# TCD Screening and Spending Among Children With Sickle Cell Anemia

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Children with sickle cell anemia are at high risk of stroke; without stroke prevention efforts, 10% will have a stroke before they are 18 years old.<sup>1,2</sup> These strokes are associated with morbidity and substantial health care spending, both in the short term and in the long term.<sup>3</sup> The impact of pediatric stroke is significant because of the large burden it places on children, families, and society.<sup>4-7</sup>

Preventive care can significantly reduce the risk of stroke in children with sickle cell anemia. Specifically, annual transcranial Doppler (TCD) screening can identify children with a high risk of stroke. For these children, chronic blood transfusions can reduce the risk of stroke by more than 90%.<sup>8,9</sup> Given the overwhelming evidence of the importance of TCD screening, an expert panel sponsored by the National Heart, Lung, and Blood Institute recommends that all children with sickle cell anemia receive annual TCD screening from age 2 years until at least age 16 years.<sup>10,11</sup>

Despite these recommendations, prior research indicates that annual TCD screening rates are suboptimal in children with sickle cell anemia enrolled in Medicaid.<sup>12-14</sup> For example, in a study of Medicaid claims data from 6 states, just 44% of such children received this screening in 2010.<sup>15</sup> It is unknown whether these findings generalize to privately insured children, who are estimated to represent 10% to 30% of children with sickle cell anemia.<sup>16-20</sup> The amount that families of privately insured children with sickle cell anemia pay out of pocket for TCD screening is also unknown. According to the principles of value-based insurance design, high-value services such as TCD screening should be subject to no or minimal cost sharing.<sup>21,22</sup> However, private plans often do not follow these principles when making coverage designs. Finally, it is unknown whether the receipt of TCD screening and out-of-pocket spending for this screening vary among families enrolled in high-deductible health plans (HDHPs) vs other plans. Several studies' findings suggest that high out-of-pocket spending and HDHP enrollment are associated with decreased rates of utilization of preventive services,<sup>23-33</sup> suggesting that families enrolled in HDHPs might have lower rates of TCD screening than those enrolled in other plans.

## ABSTRACT

**OBJECTIVES:** National guidelines recommend that children with sickle cell anemia receive annual transcranial Doppler (TCD) screening to assess stroke risk. Our objectives were to estimate the rate of TCD screening among privately insured children with sickle cell anemia, estimate out-of-pocket spending for TCD screening, and evaluate the association between TCD screening and enrollment in high-deductible health plans (HDHPs).

**STUDY DESIGN:** Cross-sectional.

**METHODS:** Using the 2009-2017 IBM MarketScan Commercial Database, we identified children aged 2 to 15 years who met a validated claims-based definition of sickle cell anemia. We calculated the proportion of children receiving annual TCD screening and out-of-pocket spending per TCD screen. Using logistic regression with generalized estimating equations, we modeled the receipt of annual TCD screening as a function of HDHP enrollment, controlling for demographics and year.

**RESULTS:** The 2519 children in the analysis accounted for 7197 person-years of enrollment; 14% of person-years were from HDHP enrollees. During 2009-2017, the proportion of children receiving TCD screening ranged from 40% to 44%. Median out-of-pocket spending for TCD screening was \$20 overall and \$65 among HDHP enrollees. Out-of-pocket spending exceeded \$100 for 27% of all screens and 42% of screens among HDHP enrollees. HDHP enrollment was not associated with TCD screening (adjusted odds ratio, 0.99; 95% CI, 0.85-1.15).

**CONCLUSIONS:** Among privately insured children with sickle cell anemia, fewer than half received annual TCD screening. Out-of-pocket spending exceeded \$100 for 27% of TCD screens. Although HDHP enrollment was not associated with TCD screening, additional studies are needed to assess whether cost sharing might deter this screening.

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## TAKEAWAY POINTS

- ▶ We aimed to determine how often privately insured children with sickle cell anemia receive annual transcranial Doppler (TCD) screening, and how much families pay out of pocket for this screening.
- ▶ In this cross-sectional analysis of administrative claims, fewer than half of privately insured children with sickle cell anemia received TCD screening. Median out-of-pocket spending for screening was \$20; out-of-pocket spending exceeded \$100 for 27% of screens.
- ▶ A substantial proportion of families of privately insured children with sickle cell anemia pay more than \$100 for TCD screening. Research is needed to determine whether there are cost-related barriers to accessing this high-value service.

The objectives of this study were as follows: (1) to estimate the rate of TCD screening among privately insured children with sickle cell anemia and (2) to estimate out-of-pocket spending for TCD screening.

## METHODS

## Data Source

This serial cross-sectional study utilized data from the 2009-2017 IBM MarketScan Commercial Database. This database includes claims data for nonelderly individuals with private employer-sponsored coverage across the United States. Data were derived from more than 120 employers, most of which are large firms, and more than 40 health plans. In 2019, MarketScan contained approximately 27 million beneficiaries, representing approximately 1 in 6 of those with employer-sponsored insurance in the United States.<sup>34</sup> Owing to this broad coverage, MarketScan is one of the most commonly used commercial claims databases in health services and policy research.

The database captures a broad spectrum of care, including inpatient, outpatient, and pharmacy services. Key data elements on inpatient and outpatient claims include *Current Procedural Terminology (CPT)* codes and *International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM)* and *International Classification of Diseases, Tenth Revision, Clinical Modification (ICD-10-CM)* diagnosis codes. Because data are derived from a limited data set, the study was not regulated as human subjects research by the Institutional Review Board of the University of Michigan; informed consent was not required.

## Study Population

The study population was children with sickle cell anemia aged 2 to 15 years. Although National Heart, Lung, and Blood Institute guidelines recommend TCD screening from 2 years “until at least age 16 years,” clinicians might feasibly interpret this text as either including or not including 16-year-olds.<sup>10,11</sup> Consequently, we cut off the sample at age 15 years. Children were identified as having sickle cell anemia based upon validated administrative claims-based definitions. These administrative claims-based definitions result from 2 separate validations. In Reeves et al,<sup>35</sup> newborn screening

records were used to test the accuracy of an *ICD-9-CM*-based algorithm for identifying children with sickle cell disease. Subsequently, the methods in that study were repeated to test the accuracy of an *ICD-9-CM*-based algorithm for identifying children with sickle cell anemia specifically. This study occurred during the process of National Quality Forum (NQF) endorsement of measures of preventive care for children with sickle cell anemia. The results of this additional validation are available on the NQF website. The case definition was 91.4%

sensitive and 80% specific in identifying children with sickle cell anemia (positive predictive value, 80.4%; negative predictive value, 91.3%). Second, in Reeves et al,<sup>36</sup> newborn screening records were used to test the accuracy of an *ICD-10-CM*-based algorithm for identifying children with sickle cell anemia. The case definition had a sensitivity of 94% and a specificity of 92%.<sup>35,37-39</sup> Given the results of these 2 validation studies, from 2009 through 2014, cases had an *ICD-9-CM* diagnosis code for sickle cell anemia (282.61, 282.62) on 3 or more claims during a calendar year. For 2016 through 2017, cases had an *ICD-10-CM* diagnosis code for sickle cell anemia (D57.00, D57.01, D57.02) or sickle cell disease without crisis (D57.1) on 1 or more outpatient claims during a calendar year. For 2015, cases were identified if they met either the *ICD-9-CM* case definition based on claims from January to September 2015 or the *ICD-10-CM* case definition based on claims from October to December 2015. Both case definitions were used owing to the transition to *ICD-10-CM* in the United States on October 1, 2015. Outpatient visits, emergency department visits, and hospitalizations were identified using validated procedure codes and revenue codes used in national quality measures. As sickle cell anemia is an inherited, lifelong disorder, a case status was assigned to a child in all eligible measurement years once the case criteria were met in any year.

To be eligible for sample inclusion, children had to be enrolled for at least 1 calendar year in the same plan and meet criteria for having sickle cell anemia in any year during the study period. Children could contribute multiple person-years of data, including from nonconsecutive years; only person-years in which children were continuously enrolled were included. Person-years were excluded if there were any claims with coordination of benefit payments during the year; if enrollees were in capitated plans, which provide encounter records that may not reliably report spending; or if out-of-pocket spending for any TCD screen was negative.

## TCD Screening

Receipt of TCD screening was identified using *CPT* codes 93886, 93888, 93890, 93892, and 93893.<sup>15</sup> Any child with at least 1 claim for a TCD screen within a calendar year was considered to have received TCD screening.<sup>11</sup>

### Out-of-Pocket Spending for TCD Screening

Out-of-pocket spending per TCD screen was defined as the sum of any deductible, coinsurance, or co-payments on claims containing a TCD procedure code. When multiple claims containing a TCD procedure code occurred on the same day (eg, separate professional and facility claims for a single procedure), out-of-pocket spending was summed across these claims. Out-of-pocket spending was inflated to 2017 US\$ using the Consumer Price Index medical care component.<sup>40,41</sup>

### Covariates

Age, sex, calendar year, rural residence, and US Census region were obtained from enrollment records. Data on US Census region were missing for 3.0% of enrollees, whereas data on rural residence were missing for 2.5% of enrollees. Multilevel multiple imputation was performed given that person-years were clustered within enrollees.<sup>42</sup> To construct a 2-level model, a deidentified beneficiary identifier was used as the cluster indicator to create subject-specific random effects. Health service encounters were identified, including outpatient visits, emergency department visits, and inpatient hospitalizations.

### Statistical Analysis

To characterize the study population, summary statistics on demographics, enrollment characteristics, and health care utilization were calculated. The proportion of children receiving TCD screening was calculated overall by study year (2009-2017) and by HDHP enrollment status. CIs were obtained by a procedure described in Clopper and Pearson<sup>43</sup> for the binomial distribution. Out-of-pocket spending per TCD screen was calculated overall and according to whether patients were enrolled in high-deductible vs non-high-deductible plans.

To explore the association between HDHP enrollment and receipt of TCD screening, a logistic regression model with generalized estimating equations and an exchangeable working correlation matrix structure was used to estimate the odds of receiving TCD screening as a function of HDHP enrollment, controlling for age, sex, indicators for study year, rural residence, and US Census region. Generalized estimation equations were used to account for the fact that children could contribute multiple person-years during the study period.<sup>44</sup>

### Sensitivity Analysis

In a sensitivity analysis, a doubly robust augmented inverse probability weights estimation method was used as an alternative method to account for selection bias in the analysis assessing the association between HDHP enrollment and receipt of TCD screening. A propensity score model was developed using logistic regression and the same set of covariates as in the primary analysis. Observations were weighted using the propensity scores, after which the same logistic regression model used in the primary analysis was fitted.<sup>45</sup> Analyses were performed using SAS version 9.4 (SAS Institute) and R version 3.5.1 (R Foundation for Statistical Computing).

**TABLE.** Demographic Characteristics of Children With Sickle Cell Anemia Aged 2-15 Years Continuously Enrolled in Private Insurance for at Least 1 Year, 2009-2017 (N=2519 children; 7197 person-years)

Characteristics	Category	n (%)
Sex	Male	3626 (50.4%)
	Female	3571 (49.6%)
Birth cohort	1994-1996	349 (4.9%)
	1997-1999	982 (13.6%)
	2000-2002	1526 (21.2%)
	2003-2005	1883 (26.2%)
	2006-2008	1470 (20.4%)
	2009-2011	727 (10.1%)
	2012-2014	260 (3.6%)
Urban/rural	Urban	6299 (87.5%)
	Rural	516 (7.2%)
	Unknown	382 (5.3%)
US Census region	Northeast	1179 (16.4%)
	Midwest	1206 (16.7%)
	South	4288 (59.6%)
	West	307 (4.3%)
	Unknown	217 (3.0%)
Plan type	Preferred provider organization	5075 (70.5%)
	Point of service	838 (11.6%)
	High-deductible health plan	992 (13.8%)
	Other	292 (4.1%)

## RESULTS

### Study Population

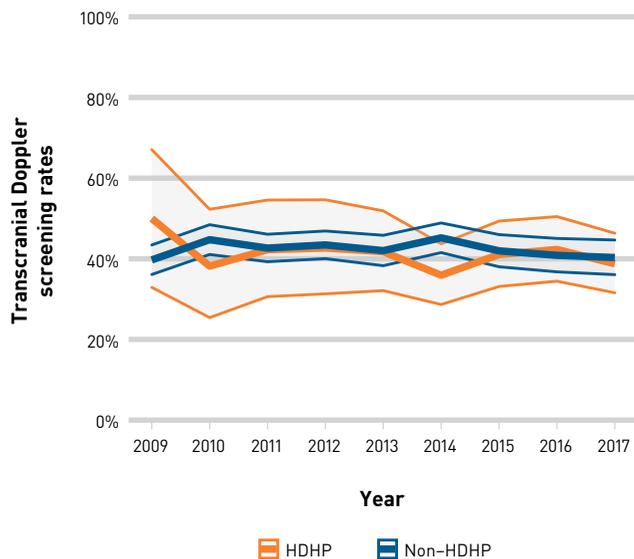
A total of 3227 children with sickle cell anemia aged 2 through 15 years were eligible to be included in the sample. These children contributed a total of 9591 person-years. Of these person-years, 593 were excluded owing to coordination of benefit payments, 1797 owing to coverage by capitated plans, and 4 owing to negative out-of-pocket spending for TCD screens. The remaining 7197 person-years were contributed by 2519 children; 708 (21.9%) of the 3227 eligible children were excluded from the sample entirely because they only contributed person-years in which exclusion criteria were not met.

Of the 2519 children in the final sample, 50.4% were male. The number of children enrolled each year varied from 699 (2017) to 908 (2011). The **Table** displays demographic characteristics at the person-year level. Across the study period, the mean (SD) annual number of outpatient visits per person-year was 7.1 (5.7), the mean (SD) annual number of emergency department visits per person-year was 1.1 (1.9), and the mean (SD) annual number of inpatient hospitalizations per person-year was 0.8 (1.3).

### TCD Screening Rates

TCD screening rates ranged from 40.0% (2017) to 44.3% (2010) overall. Children enrolled in an HDHP had TCD screening rates

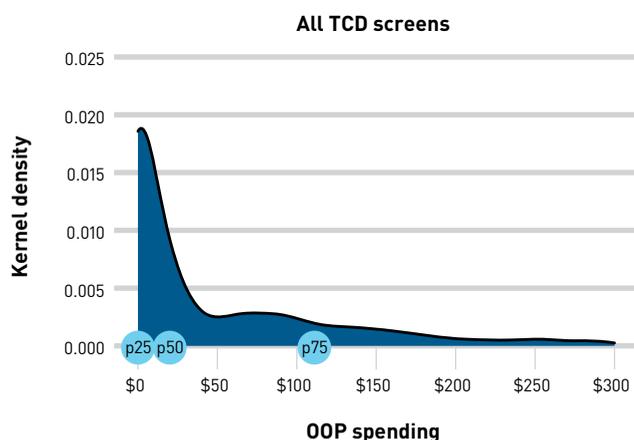
**FIGURE 1.** Annual Transcranial Doppler Screening Rates Among Privately Insured Children Aged 2-15 Years With Sickle Cell Anemia, 2009-2017, Stratified by Type of Health Plan (HDHP vs non-HDHP)<sup>44,a</sup>



HDHP, high-deductible health plan.

<sup>a</sup>Shaded areas represent 95% CIs obtained by a procedure described by Clopper and Pearson<sup>44</sup> for the binomial distribution.

**FIGURE 2.** Distribution of Inflation-Adjusted OOP Spending on Transcranial Doppler Screening (N = 2519 children; 7197 person-years)<sup>a</sup>



OOP, out-of-pocket.

<sup>a</sup>The shaded blue area represents the kernel density distribution of annual OOP spending per transcranial Doppler screening. p25, p50, and p75 correspond to the 25th, 50th, and 75th percentiles, respectively, of annual OOP spending per transcranial Doppler screening. Annual OOP spending was inflated to 2017 US\$ using the Consumer Price Index medical care component.

ranging from 35.9% to 50.0%. Those who were not enrolled in an HDHP had screening rates ranging from 39.7% to 45.2% (Figure 1<sup>44</sup>).

### Out-of-Pocket Spending for TCD Screens

Across the study period, out-of-pocket spending for TCD screens ranged from \$0 to \$1808 (median [IQR], \$20 [\$0-\$111]). This quantity was right-skewed, exceeding \$100 for 27% of screens (Figure 2). Aggregate out-of-pocket spending for TCD screens across the sample constituted 16.6% of aggregate total spending for these screens (ie, the sum of out-of-pocket spending and insurer reimbursement).

Out-of-pocket spending for TCD screens was higher for HDHP enrollees (median [IQR], \$65 [\$0-\$252]) compared with non-HDHP enrollees (median [IQR], \$18 [\$0-\$98]). Among TCD screens for HDHP enrollees and non-HDHP enrollees, out-of-pocket spending exceeded \$100 for 42% and 25%, respectively.

### Association Between HDHP Enrollment and TCD Screening

In adjusted analyses, enrollment in an HDHP was not associated with receiving TCD screening (adjusted odds ratio, 0.99; 95% CI, 0.85-1.15). A doubly robust, augmented inverse probability weights estimation method produced similar results (average treatment effect of HDHP enrollment on receipt of TCD screening, -0.01 percentage points; 95% CI, -0.05 to 0.03).

## DISCUSSION

We found that fewer than half of privately insured US children with sickle cell anemia received recommended annual TCD screening and that the rate of annual TCD screening changed little over time. Although most families were subjected to modest cost sharing for TCD screening, some experienced out-of-pocket spending exceeding \$100 for TCD screens.

This study provides the most recent national data on TCD screening rates among privately insured children. The low rates of TCD screening in our study are consistent with findings from older analyses of Medicaid claims and with an analysis of TCD screening rates among privately and publicly insured children at 28 clinical centers from 2012 through 2016.<sup>15,46</sup> However, our results stand in contrast to those of a single-center study in which privately insured children with sickle cell anemia were more likely to adhere to TCD screens that were ordered.<sup>46</sup>

Pediatric stroke is a devastating morbidity associated with sickle cell anemia. More than half of children experience neurological deficits at discharge after a stroke. For the majority of children, these deficits continue to persist for at least 1 year post stroke.<sup>4,5</sup> Further, health-related quality of life is significantly lower among children who have had a stroke compared with their healthier counterparts.<sup>5</sup> Rates of recurrence are high, with 25% of children having a recurrent stroke within 2 years of initial stroke.<sup>6</sup> TCD screening can accurately identify children at highest risk of stroke. Our study indicates that quality improvement efforts to increase

TCD screening initiated and implemented at the health plan level should include children with private insurance.

Health plans are uniquely positioned to implement quality improvement initiatives to promote TCD screening because they can track enrollees' health services across time and across hospital systems.<sup>47</sup> For example, children with sickle cell anemia may receive outpatient and emergency department care at separate institutions.<sup>48</sup> Although this fragmented care may not be available for review at either institution, the health plan would be able to link these encounters together for a child. Further, health plans have opportunities to manage and coordinate all preventive services for sickle cell anemia. This allows health plans to leverage their data to identify children with sickle cell anemia, assess gaps in quality of care for preventive services such as TCD screening, and implement solutions to close these gaps. Preventing strokes in children with sickle cell anemia will also eliminate the high costs associated with pediatric stroke for the health plans, both in the year following the stroke as well as across the patients' life span.<sup>3</sup> Finally, health plans are increasingly addressing social determinants of health to assist patients and families, such as providing transportation to TCD screening appointments.<sup>49</sup> Such interventions could increase TCD screening rates. As many health plans have both public and private insurance enrollees, solutions to improve TCD screening may be applied to children across payer types. Some strategies to reduce barriers to TCD screening, such as colocating screening within sickle cell outpatient clinics, could improve rates for both publicly and privately insured children.<sup>50</sup>

Multiple interventions may be needed to improve screening rates for privately insured children with sickle cell anemia. For example, a substantial proportion of children with sickle cell anemia lack access to hematologists, who may be more likely to order TCD screening than primary care pediatricians. However, it is unknown the degree to which deficits in TCD screening are driven by the lack of an opportunity to order this screen (eg, owing to lack of access), the lack of an order by clinicians conditional upon having such an opportunity, and the lack of completion of TCD screening by patients for whom this screening is ordered. Closing this knowledge gap is critical to inform the design of quality improvement interventions to increase TCD screening. Additionally, the families of many children with sickle cell anemia experience transportation and time costs when receiving TCD screening.<sup>51-53</sup> These considerations suggest that an approach addressing barriers at multiple levels is needed to increase rates of TCD screening.

Our results indicate that approximately 60% of TCD screens received by privately insured children with sickle cell anemia had some level of cost sharing. Overall, 27% of TCD screens had cost sharing exceeding \$100. For those enrolled in HDHPs, this proportion was even higher at 42%.

The fact that cost sharing for TCD screening can sometimes be substantial raises the concern that cost sharing might deter use of this high-value service. In our study, we do not find evidence to support this hypothesis, as enrollment in an HDHP was not associated with

lower rates of TCD screening. An important caveat, however, is that our study was cross-sectional in nature. Consequently, we could not account for unobserved differences between HDHP enrollees and non-HDHP enrollees that might affect the probability of TCD screening (eg, household income, disease severity, access to care). Future studies may be able to more rigorously assess the association between cost sharing and TCD screening by exploiting natural experiments, such as instances in which employers force patients to switch from non-HDHP to HDHP plans.<sup>54,55</sup>

## Limitations

This study has limitations. First, the database only included out-of-pocket spending among enrollees who completed TCD screening. Some families facing high potential cost sharing may have decided against TCD screening. However, this possibility could not be assessed because the database lacked information on benefit design or whether patients had met deductibles or annual out-of-pocket maximums at the time that TCD screening was being considered.

Second, the database is not nationally representative of all privately insured plans. Third, our study population, children with sickle cell anemia, was identified using administrative claims as opposed to the gold standard of newborn screening. Additionally, there may be differences in the rates of misclassification of patients with sickle cell anemia depending on whether the *ICD-9-CM* or *ICD-10-CM* version of the algorithm was used. However, we note that rates of TCD screening were similarly low both before and after the transition to *ICD-10-CM* in October 2015.

Finally, use of administrative claims to identify the study population excludes children who did not receive sickle cell–related care within a year. It is important to tailor interventions to include this population when considering opportunities to improve TCD screening rates.

## CONCLUSIONS

Privately insured children with sickle cell anemia have low rates of annual TCD screening. Cost sharing for this screening can be substantial for some patients. Although enrollment in an HDHP was not associated with TCD screening in this study, future research using alternative methods are needed to determine the precise relationship between cost sharing and TCD screening. ■

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for important intellectual content (SLR, SN, KJD, JLR, KPC); statistical analysis (SN, KPC); obtaining funding (SLR); administrative, technical, or logistic support (SN); and supervision (SLR, KPC).

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## REFERENCES

- Verduzzo LA, Nathan DG. Sickle cell disease and stroke. *Blood*. 2009;114(25):5117-5125. doi:10.1182/blood-2009-05-220921
- Ohene-Frempong K, Weiner SJ, Sleeper LA, et al. Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood*. 1998;91(1):288-294.
- Hamilton W, Huang H, Seiber E, Lo W. Cost and outcome in pediatric ischemic stroke. *J Child Neurol*. 2015;30(11):1483-1488. doi:10.1177/0883073815570673
- Goldenberg NA, Bernard TJ, Fullerton HJ, Gordon A, deVeber G; International Pediatric Stroke Study Group. Antithrombotic treatments, outcomes, and prognostic factors in acute childhood-onset arterial ischaemic stroke: a multicentre, observational, cohort study. *Lancet Neurol*. 2009;8(12):1120-1127. doi:10.1016/s1474-4422(09)70241-8
- Nossen MH, Aarsen FK, Akker SL, et al. Paediatric arterial ischaemic stroke: functional outcome and risk factors. *Dev Med Child Neurol*. 2010;52(4):394-399. doi:10.1111/j.1469-8749.2009.03580.x
- Lanthier S, Kirkham FJ, Mitchell LG, et al. Increased antidiabetic antibody IgG titers do not predict recurrent stroke or TIA in children. *Neurology*. 2004;62(2):194-200. doi:10.1212/wnl.62.2.194
- Massicotte MP, Bauman ME, Albers GW. Challenges in childhood arterial ischaemic stroke. *Lancet Neurol*. 2009;8(12):1079-1081. doi:10.1016/s1474-4422(09)70269-8
- Adams RJ, McKie VC, Hsu L, et al. Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. *N Engl J Med*. 1998;339(1):5-11. doi:10.1056/NEJM199807023390102
- Cherry MG, Greenhalgh J, Osipenko L, et al. The clinical effectiveness and cost-effectiveness of primary stroke prevention in children with sickle cell disease: a systematic review and economic evaluation. *Health Technol Assess*. 2012;16(43):1-129. doi:10.3310/hta16430
- National Heart, Lung, and Blood Institute. *The Management of Sickle Cell Disease*. 4th ed. National Heart, Lung, and Blood Institute; 2002. Accessed February 22, 2019. [https://www.nhlbi.nih.gov/files/docs/guidelines/sc\\_mngt.pdf](https://www.nhlbi.nih.gov/files/docs/guidelines/sc_mngt.pdf)
- Evidence-based management of sickle cell disease: expert panel report, 2014. National Heart, Lung, and Blood Institute. 2014. Accessed October 6, 2021. [https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%202016\\_0.pdf](https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%202016_0.pdf)
- Eckrich MJ, Wang WC, Yang E, et al. Adherence to transcranial Doppler screening guidelines among children with sickle cell disease. *Pediatr Blood Cancer*. 2013;60(2):270-274. doi:10.1002/pbc.24240
- Hussain S, Nichols F, Bowman L, Xu H, Neuner C. Implementation of transcranial Doppler ultrasonography screening and primary stroke prevention in urban and rural sickle cell disease populations. *Pediatr Blood Cancer*. 2015;62(2):219-223. doi:10.1002/pbc.25306
- Kanter J, Phillips S, Schlenz AM, et al. Transcranial Doppler screening in a current cohort of children with sickle cell anemia: results from the DISPLACE study. *J Pediatr Hematol Oncol*. 2021;43(8):e1062-e1068. doi:10.1097/MPH.0000000000002103
- Reeves SL, Madden B, Freed GL, Dombkowski KJ. Transcranial Doppler screening among children and adolescents with sickle cell anemia. *JAMA Pediatr*. 2016;170(6):550-556. doi:10.1001/jamapediatrics.2015.4859
- Yusuf HR, Atrash HK, Grosse SD, Parker CS, Grant AM. Emergency department visits made by patients with sickle cell disease: a descriptive study, 1999-2007. *Am J Prev Med*. 2010;38(suppl 4):S536-S541. doi:10.1016/j.amepre.2010.01.001
- Amendah DD, Mvundura M, Kavanagh PL, Sprinz PG, Grosse SD. Sickle cell disease-related pediatric medical expenditures in the U.S. *Am J Prev Med*. 2010;38(suppl 4):S550-S556. doi:10.1016/j.amepre.2010.01.004
- Boulet SL, Yanni EA, Creary MS, Olney RS. Health status and healthcare use in a national sample of children with sickle cell disease. *Am J Prev Med*. 2010;38(suppl 4):S528-S535. doi:10.1016/j.amepre.2010.01.003
- Mvundura M, Amendah D, Kavanagh PL, Sprinz PG, Grosse SD. 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. doi:10.1002/pbc.22069
- Robinson MR, Daniel LC, O'Hara EA, Szabo MM, Barakat LP. Insurance status as a sociodemographic risk factor for functional outcomes and health-related quality of life among youth with sickle cell disease. *J Pediatr Hematol Oncol*. 2014;36(1):51-56. doi:10.1097/MPH.0000000000000013
- Chernew ME, Rosen AB, Fendrick AM. Value-based insurance design. *Health Aff (Millwood)*. 2007;26(suppl 2):w195-w203. doi:10.1377/hlthaff.26.2.w195
- Norris HC, Richardson HM, Benoit MC, Shrosbree B, Smith JE, Fendrick AM. Utilization impact of cost-sharing elimination for preventive care services: a rapid review. *Med Care Res Rev*. 2022;79(2):175-197. doi:10.1177/10775587211027372
- Rezayatmand R, Pavlova M, Groot W. The impact of out-of-pocket payments on prevention and health-related lifestyle: a systematic literature review. *Eur J Public Health*. 2013;23(1):74-79. doi:10.1093/eurpub/cks034
- Gibson TB, Mark TL, Axelsen K, Baser O, Rublee DA, McGuigan KA. Impact of statin copayments on adherence and medical care utilization and expenditures. *Am J Manag Care*. 2006;12(suppl No. 12):SP11-SP19.
- Ye X, Gross CR, Schommer J, Cline R, St Peter WL. Association between copayment and adherence to statin treatment initiated after coronary heart disease hospitalization: a longitudinal, retrospective, cohort study. *Clin Ther*. 2007;29(12):2748-2757. doi:10.1016/j.clinther.2007.12.022
- Agarwal R, Mazurenko O, Menachemi N. High-deductible health plans reduce health care cost and utilization, including use of needed preventive services. *Health Aff (Millwood)*. 2017;36(10):1762-1768. doi:10.1377/hlthaff.2017.0610
- Beuwkes Buntin M, Haviland AM, McDevitt R, Sood N. Healthcare spending and preventive care in high-deductible and consumer-directed health plans. *Am J Manag Care*. 2011;17(3):222-230.
- Charlton ME, Levy BT, High RR, Schneider JE, Brooks JM. Effects of health savings account-eligible plans on utilization and expenditures. *Am J Manag Care*. 2011;17(1):79-86.
- Fronstin P, Sepulveda MJ, Roebuck MC. Consumer-directed health plans reduce the long-term use of outpatient physician visits and prescription drugs. *Health Aff (Millwood)*. 2013;32(6):1126-1134. doi:10.1377/hlthaff.2012.0493
- Haviland AM, Sood N, McDevitt R, Marquis MS. How do consumer-directed health plans affect vulnerable populations? *Forum Health Econ Policy*. 2011;14(1):20112006. doi:10.2202/1558-9544.1248
- Wharam JF, Zhang F, Landon BE, LeCates R, Soumerai S, Ross-Degnan D. Colorectal cancer screening in a nationwide high-deductible health plan before and after the Affordable Care Act. *Med Care*. 2016;54(5):466-473. doi:10.1097/MLR.0000000000000521
- Wharam JF, Graves AJ, Landon BE, Zhang F, Soumerai SB, Ross-Degnan D. Two-year trends in colorectal cancer screening after switch to a high-deductible health plan. *Med Care*. 2011;49(9):865-871. doi:10.1097/MLR.0b013e31821b35d8
- Wharam JF, Galbraith AA, Kleinman KP, Soumerai SB, Ross-Degnan D, Landon BE. Cancer screening before and after switching to a high-deductible health plan. *Ann Intern Med*. 2008;148(9):647-655. doi:10.7326/0003-4819-148-9-200805060-00004
- Health insurance coverage of the total population. Kaiser Family Foundation. Accessed December 15, 2021. <https://www.kff.org/other/state-indicator/total-population>
- Reeves S, Garcia E, Kleyn M, et al. Identifying sickle cell disease cases using administrative claims. *Acad Pediatr*. 2014;14(5):S61-S67. doi:10.1016/j.acap.2014.02.008
- Reeves SL, Madden B, Wu M, et al. Performance of ICD-10-CM diagnosis codes for identifying children with sickle cell anemia. *Health Serv Res*. 2020;55(2):310-317. doi:10.1111/1475-6773.13257
- Antibiotic prophylaxis among children with sickle cell anemia. National Quality Forum. Updated July 2, 2021. Accessed December 1, 2021. <http://www.qualityforum.org/QPS/3166>
- Hydroxyurea use among children with sickle cell anemia. National Quality Forum. Updated July 2, 2021. Accessed December 1, 2021. <http://www.qualityforum.org/QPS/3595>
- Transcranial Doppler ultrasonography screening among children with sickle cell anemia. National Quality Forum. Updated December 12, 2022. Accessed February 9, 2023. <http://www.qualityforum.org/QPS/2797>
- Dunn A, Grosse SD, Zuvekas SH. Adjusting health expenditures for inflation: a review of measures for health services research in the United States. *Health Serv Res*. 2018;53(1):175-196. doi:10.1111/1475-6773.12612
- CPI tables. Health Resources and Services Administration. Accessed November 25, 2019. <https://www.hrsa.gov/sites/default/files/hrsa/get-health-care/affordable/hill-burton/cpitables.pdf>
- Goldstein H, Carpenter J, Kenward MG, Levin KA. Multilevel models with multivariate mixed response types. *Stat Modelling*. 2009;9(3):173-197. doi:10.1177/1471082x080900301
- Clopper CP, Pearson ES. The use of confidence or fiducial limits illustrated in the case of the binomial. *JSTOR*. 1934;26(4):404-413. doi:10.2307/2331986
- Liang KY, Zeger SL. Longitudinal data analysis using generalized linear models. *Biometrika*. 1986;73(1):13-22. doi:10.1093/biomet/73.1.13
- Bang H, Robins JM. Doubly robust estimation in missing data and causal inference models. *Biometrics*. 2005;61(4):962-973. doi:10.1111/j.1541-0420.2005.00377.x
- Raphael JL, Shetty PB, Liu H, Mahoney DH, Mueller BJ. A critical assessment of transcranial Doppler screening rates in a large pediatric sickle cell center: opportunities to improve healthcare quality. *Pediatr Blood Cancer*. 2008;51(5):647-651. doi:10.1002/pbc.21677
- Warren LR, Clarke JM, Arora S, Barahona M, Arebi N, Darzi A. Transitions of care across hospital settings in patients with inflammatory bowel disease. *World J Gastroenterol*. 2019;25(17):2122-2132. doi:10.3748/wjg.v25.i17.2122
- Panepinto J, Owens P, Mosso A, Steiner CA, Brousseau D. Concentration of hospital care for acute sickle cell disease-related visits. *Pediatr Blood Cancer*. 2012;59(4):685-689. doi:10.1002/pbc.24028
- Raphael JL. Addressing social determinants of health in sickle cell disease: the role of Medicaid policy. *Pediatr Blood Cancer*. 2020;67(5):e28202. doi:10.1002/pbc.28202
- McCarville MB, Goodin GS, Fortner G, et al. Evaluation of a comprehensive transcranial Doppler screening program for children with sickle cell anemia. *Pediatr Blood Cancer*. 2008;50(4):818-821. doi:10.1002/pbc.21430
- Weisman JK, Diamond CE, Kappa S, Nickel RS. Transcranial Doppler screening adherence among children with sickle cell anemia seen in the emergency department. *J Pediatr*. 2020;217:172-176.e1. doi:10.1016/j.jpeds.2019.10.049
- Cabana MD, Kanter J, Marsh AM, et al. Barriers to pediatric sickle cell disease guideline recommendations. *Glob Pediatr Health*. 2019;6:2333794x19847026. doi:10.1177/2333794x19847026
- Crosby LE, Joffe NE, Davis B, et al. Implementation of a process for initial transcranial Doppler ultrasonography in children with sickle cell anemia. *Am J Prev Med*. 2016;51(1)(suppl 1):S10-S16. doi:10.1016/j.amepre.2016.01.021
- Galbraith AA, Ross-Degnan D, Soumerai SB, et al. High-deductible health plans: are vulnerable families enrolled? *Pediatrics*. 2009;123(4):e589-e594. doi:10.1542/peds.2008-1738
- Fendrick AM, Buxbaum JD, Tang Y, et al. Association between switching to a high-deductible health plan and discontinuation of type 2 diabetes treatment. *JAMA Netw Open*. 2019;2(11):e1914372. doi:10.1001/jamanetworkopen.2019.14372

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