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Performance of ICD-10-CM Diagnosis Codes for Identifying Children with Sickle Cell Anemia

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Abstract

Objective: To develop, test, and validate the performance of ICD-10-CM claims-based case definitions for identifying children with sickle cell anemia (SCA).

Data Sources: Medicaid administrative claims (2016) for children <18 years with potential SCA (any D57x diagnosis code) and newborn screening records from Michigan and New York State.

Study Design: This study is a secondary data analysis.

Data Collection/Extraction Methods: Using specific SCA-related (D5700, D5701, D5702) and non-specific (D571) diagnosis codes, 23 SCA case definitions were applied to Michigan Medicaid claims (2016) to identify children with SCA. Measures of performance (sensitivity, specificity, area under ROC curve) were calculated using newborn screening results as the gold standard. A parallel analysis was conducted using New York State Medicaid claims and newborn screening data.

Principal Findings: In Michigan Medicaid, 1,597 children had ≥1 D57x claim; 280 (18%) were diagnosed with SCA. Measures of performance varied, with sensitivities from 0.02-0.97 and specificities from 0.88-1.0. The case definition of ≥1 outpatient visit with a SCA-related or D571 code had the highest area under the ROC curve, with a sensitivity of 95% and specificity of 92%. The same definition also had the highest performance in New York Medicaid (n=2,454), with a sensitivity of 94% and specificity of 86%.

Conclusions: Children with SCA can be accurately identified in administrative claims using this straightforward case definition. This methodology can be used to monitor trends and use of health services after transition to ICD-10-CM.

Key Words: sickle cell anemia, administrative claims, ICD-10-CM

What this study adds:

- Accurate identification of children with sickle cell anemia using administrative claims is essential for tracking health services, receipt of preventive care, and outcomes.
- We developed, tested, and validated the performance of ICD-10-CM case definitions for identifying children with sickle cell anemia against newborn screening records in two large state Medicaid programs.
- The case definition of ≥1 outpatient visit with a sickle cell anemia-related or D571 code accurately identified children with sickle cell anemia.
- This methodology can be used to monitor trends and use of health services after transition to ICD-10-CM.

Introduction

Sickle cell disease is an inherited blood disorder that affects approximately 100,000 Americans.[1] There are many subtypes of sickle cell disease. Sickle cell anemia (SCA) is a collective term which includes subtypes hemoglobin (Hb) SS and Hbβ^0thalassemia.[2] Children with SCA are at risk for numerous morbidities, such as pain, serious infections, and stroke.[3] At least half will experience at least one hospitalization related to pain within a year.[4] Children with SCA are at 100 times the risk of infection, and 300 times the risk of stroke as compared to a child without SCA.[5,6]

The National Heart, Lung, and Blood Institute (NHLBI) developed evidence-based guidelines for preventive care among those living with sickle cell disease; many of these recommendations are targeted toward those with SCA.[2] For example, the NHLBI recommends that children with SCA receive the medication hydroxyurea to prevent pain crises, antibiotic prophylaxes to prevent infection, and transcranial Doppler (TCD) screening to identify children at the highest risk of stroke.[2] As a result of the comorbidities, complications, and receipt of preventive care, children with SCA experience numerous routine and acute encounters with the healthcare system.[7-19]

These encounters result in significant monetary and social costs for families and the healthcare system.[19]

The importance of the receipt of preventive care coupled with high healthcare utilization and cost emphasizes the need for population-based assessments of the quality of care among children with SCA. Such assessments may be particularly challenging as SCA is a rare disease. Given relatively frequent health services utilization among children with SCA, administrative billing claims, which are widely available for publically and commercially insured populations, may provide a useful data source for tracking population-based health and quality of care.[20]

Diagnosis codes, which are contained in administrative claims databases, are commonly used to identify children with chronic diseases such as asthma, diabetes and inflammatory bowel disease.[21-24] Several studies have demonstrated that administrative-claims using ICD-9-CM diagnosis codes can be used to accurately identify children with sickle cell disease.[7,25,26] However, these definitions are subject to two limitations in assessing health services among children with SCA: 1) identification of children with sickle cell disease more broadly as opposed to children with SCA; 2) use of ICD-9-CM diagnosis codes. Beginning in October 1, 2015, the Centers for Medicare and Medicaid Services (CMS) required the use of ICD-10-CM diagnosis codes in the United States.[27] Therefore, any definitions using ICD-9-CM diagnosis codes are no longer feasible for any assessments of administrative claims data post September 2015. To address these limitations, we developed, tested, and validated the performance of ICD-10-CM case definitions for identifying children with SCA. The goal of this analysis is to support evaluation of health services and receipt of preventive care among these children.

Methods

We developed, tested, and validated the performance of ICD-10-CM case definitions for SCA through the following 4-step process: 1) developing candidate SCA case definitions; 2) identifying a test population for the definitions; 3) testing the accuracy of

candidate case definitions; and 4) testing case definitions within an independent population.

1) Developing candidate SCA case definitions

The requisite transition from ICD-9-CM to ICD-10-CM diagnosis codes in the US has revealed several potential difficulties related to case definitions using administrative claims.[27] The transition introduced a coding nuance that is potentially quite impactful when evaluating health services related to SCA. The relevant ICD-10-CM diagnosis codes include three billable categories related to acute care for SCA: D5700 (HbSS with crisis); D5701 (HbSS with acute chest syndrome); and D5702 (HbSS with splenic sequestration). The ICD-10 diagnosis code D570 is non-billable per CMS.[28] There are no specific ICD-10 diagnosis codes that characterize care related to sickle cell *anemia* without a crisis. While there is an ICD-10-CM code (D571), this code is for sickle cell *disease* without crisis, but does not stipulate a specific subtype of sickle cell disease. Use of this general diagnosis code has unknown implications for accurate case identification of children with SCA; consequently, we developed and tested several case definitions that included the D571 diagnosis code.

We developed three mutually exclusive claims categories of health services related to SCA: outpatient visits, emergency department (ED) visits, and inpatient hospitalizations. Each category was defined using code sets to identify health services encounters from validated HEDIS® definitions based on Current Procedural Terminology (CPT) procedure codes, Uniform Billing Revenue codes, and ICD-10-CM diagnosis codes.[29] The HEDIS® definition for outpatient visits does not include visits for laboratory only. We also developed a case definition comprised of the overall count of SCA claims, irrespective of service type. The combinations of service categories within our ICD-10-CM case definitions was informed by our previous work validating ICD-9-CM case definitions.[7]

The health services categories were then used to develop 23 candidate case definitions, varying from a simple count of claims to various combinations of outpatient

visits and/or inpatient admissions. Definitions with simple counts of claims included any health services claim with a diagnosis code, irrespective of service or location (e.g., inpatient, outpatient, emergency department). Specific types of health services were considered, as these services can have different predictive values for identifying cases in other chronic conditions.[30,31] Counts of claims or service types ranged from at least one, to at least 3; previous findings indicate that little specificity would be gained by requiring additional claims and would result in loss of sensitivity.[7] Definitions with varying lengths between outpatient visits were also considered, consistent with other administrative claims-based definitions for chronic conditions.[32] It is important to note that definitions also varied by which ICD-10-CM codes were considered a SCA-related event; 14 case definitions only included D5700, D5701, and D5702 diagnosis codes while the remaining 9 definitions also included the non-specific D571 diagnosis code. Table 1 summarizes each candidate SCA case definition and the diagnosis codes upon which each is based. Case definitions are uniquely identified with letters (A-W) for subsequent reference.

2) Identifying a test population

Our test population included children in Michigan Medicaid ages 1 through 17 years who potentially had SCA, classified as those having at least 1 claim with a D57x diagnosis within the 2016 study period. Children were continuously enrolled for the entire calendar year and we excluded those with any other form of health insurance to maximize the completeness of health services claims within the study period.

Children meeting eligibility criteria were linked to Michigan birth certificates using child's name, birthdate, and sex; unlinked records were manually reviewed. Birth certificates were linked to NBS records using a previously validated method.[33] Children were classified as having: 1) confirmed SCA (HbSS, Hbβ^{0}Thalassemia); 2) not SCA; or 3) unknown status. An unknown NBS record status could result from situations whereby the child's Medicaid record cannot be linked to an electronic birth certificate (e.g., name change, born in another state, etc.), or as a consequence of an unconfirmed NBS result. In Michigan, any child with an abnormal hemoglobin result suggestive of disease on a

NBS is referred to a hematologist for confirmatory testing; these results are reported back to the NBS Program. Therefore, a child classified as having SCA in their Michigan NBS record has received confirmatory testing in addition to normal screening procedures. Michigan children with other subtypes of sickle cell disease, such as HbSC or HbSD, or normal hemoglobin, were classified as not SCA; those with unknown or undocumented NBS results were excluded from further analysis. For children in our testing population, descriptive statistics of age and sex were calculated both overall and by status (SCA / not SCA).

3) *Testing the accuracy of candidate case definitions*

Measures of performance were calculated for all 23 candidate case definitions among children in our test population with either confirmed SCA or not SCA, as determined through Michigan NBS testing and follow-up. Confirmed Michigan NBS results were used as our gold standard for measures of performance calculations. Measures of case definition performance included sensitivity, specificity, and area under the Receiver Operating Characteristic (ROC) curve.[34]

4) *Testing case definitions within an independent population*

Similar to the approach used in Michigan, we conducted a parallel analysis using the identical 23 candidate case definitions tested in Michigan in conjunction with claims and NBS data obtained from the New York State (NYS) Medicaid program. Children born in NYS in years 2006-2013 with sickle cell disease, as identified by NBS and confirmatory testing, were matched to corresponding Medicaid records as previously described.[35] The NYS population consisted of children ages 3-10 years who had at least 1 Medicaid claim with a D57x diagnosis during the 2016 study period; complete NYS NBS data were not available for children younger than 3 or older than 10 years of age. Children were continuously enrolled in NYS Medicaid for the entire calendar year with no other forms of health insurance within the study period.

Children were classified as having either confirmed SCA (HbSS, Hbβ^0Thalassemia), or not SCA. In NYS, any child with an abnormal hemoglobin result

on an NBS is referred to a hematologist for confirmatory testing; these results are reported back to the NYS NBS Program. Therefore, a child classified as having SCA within NBS records has been subject to the normal NBS procedures, as well as additional confirmatory testing. NYS data coding limitations required that children with other subtypes of sickle cell disease, normal hemoglobin and unknown NBS results be classified as not SCA. For each candidate case definition, measures of performance were calculated using NYS confirmed NBS results as the gold standard. Measures of performance included sensitivity, specificity, and area under the ROC curve.

Sensitivity Analyses: We performed two sensitivity analyses to assess the impact of additional codes and age upon our results. First, although the 3-digit code D570 is not billable per CMS guidance, we assessed occurrence of this code among children in the Michigan Medicaid study population.[28] Second, we assessed the sensitivity and specificities of our most accurate definition by stratifying the children into the following age groups within the Michigan Medicaid population: 1-5 years, 6-11 years, 12-17 years. We also assessed the sensitivity and specificity of the same case definition when the Michigan Medicaid study population was limited to ages 3-10 for a direct comparison to the NY Medicaid population.

Results

In Michigan, 1,597 children 1-18 years old had ≥1 D57x claim and were continuously enrolled in Michigan Medicaid in 2016 with no other forms of health insurance. Among these children, 280 (18%) were diagnosed with SCA and 1,177 (74%) were not SCA; 140 (8%) were excluded due to an unknown NBS result. Among the 1,457 children in the population with known NBS results, average age as of January 1, 2016 was 7.2 (SD=4.7) years, 49% were female, and 85% were recorded as non-Hispanic Black within Medicaid records (Table 2).

Measures of performance varied substantially across the 23 candidate case definitions tested in Michigan, with sensitivities ranging widely (0.03-0.97) and

specificities varying from 0.81-1.0 (Table 3). As expected, as definitions moved from less to more restrictive (i.e., one outpatient visit to two outpatient visits related to SCA), sensitivities decreased and specificities increased. Each definition that included a D571 diagnosis code in addition to the SCA diagnosis codes (D5700, D5701, D5702) had a substantially higher sensitivity; decreases in specificity were small when comparing definitions with and without the non-specific D571 diagnosis code. Area under the ROC curve ranged from 0.51 to 0.93. The case definition of ≥1 outpatient visit with a SCA-related or D571 code had the highest area under the ROC curve (0.93), with a sensitivity of 94% and specificity of 92%.

In NYS, there were 2,454 children that were 3-10 years old, had ≥1 D57x claim, and were continuously enrolled in NYS Medicaid in 2016 with no other forms of health insurance. Among these children, 505 (21%) were diagnosed with SCA, 1,949 (79%) were either not SCA or did not have an NBS result. Among the 2,454 children in the NYS Medicaid population, average age as of January 1, 2016 was 6.1 (SD=2.31) years, 48% were female, and 59% were recorded as non-Hispanic Black (Table 2).

Among the 23 candidate case definitions validated in NYS, sensitivities ranged widely, from 0.01-0.98, and specificities ranged from 0.81-1.0 (Table 3). Similar to Michigan, we found that definitions including a D571 diagnosis code had a higher sensitivity as compared to those with only the SCA diagnosis codes, with small decreases in specificity. Area under the ROC curve ranged from 0.51-0.90. The same case definition of ≥1 outpatient visit with a SCA-related or D571 code had the highest area under the ROC curve (0.90), with a sensitivity of 94% and specificity of 86%.

Sensitivity analyses: Consistent with CMS billing guidance, the non-billable ICD-10 code D570 was not found in any claim among our Michigan Medicaid study population. When the most accurate case definition (Definition O) was applied to stratified age groups, there were no clinically relevant changes in the sensitivities and specificities of the definition as compared to NBS records (Table 4).

Discussion

We conducted this study to develop, test, and validate a case definition for identifying children with SCA using administrative-claims data from two large state Medicaid programs. To our knowledge, this is the first US study to test the accuracy of using ICD-10-CM diagnosis codes to identify children with SCA. We found that our best performing case definition, one outpatient visit with a SCA (D5700, D5701, D5702) or D571 ICD-10-CM diagnosis code, had high sensitivity and specificity, in both our populations. Given the high accuracy of our case definition, our findings lend confidence that this definition will enable accurate, population-based assessments of the quality of care and health services among children with SCA when using administrative claims. Importantly, our results demonstrate that a high degree of accuracy in identifying children with SCA can be achieved in settings where NBS results are not available.

There are numerous case definitions developed to identify children with sickle cell disease using ICD-9-CM diagnosis codes that cannot be applied to claims data reporting ICD-10-CM diagnosis codes.[7] These ICD-9-CM definitions vary from a simple count of sickle cell disease-related diagnosis codes to combinations of outpatient and inpatient visits.[36-42] The majority of these ICD-9-CM definitions were developed to identify children with any form of sickle cell disease, as opposed to children with a specific diagnosis of SCA. The most accurate ICD-10-CM definition to specifically identify children with SCA is straightforward and easy to implement. It has been tested against the genetic gold standard of NBS records. The use of NBS records ensures the candidate case definitions are tested against a gold standard that has nearly zero misclassification of hemoglobin status. This gold standard is more accurate than those used in many other claims-based validation work, which often use medical records or registry data to confirm case status.[43-45] As health services researchers continue to identify opportunities for improvement in the care of these high-risk children using administrative claims, it would be advantageous to begin use of this validated and accurate case definition. Further, results obtained across studies using the same case definition may be more comparable than studies using a definition with unknown sensitivity and specificity.

Our study shows that including a D571 diagnosis code (sickle cell disease without crisis) was essential for identifying children with SCA, as results showed significantly improved sensitivity, with a small negative impact on specificity. This was a particularly surprising finding given the lack of specificity for the SCA subtype of this diagnosis code. For example, definitions O and J both require one outpatient visit related to SCA; however, definition O also allows for the outpatient visit to be coded with the non-specific D571 code. Definition J had a sensitivity of 16%; definition O had a sensitivity of 94%. Specificities for the two definitions were 99% (Definition J) and 92% (Definition O). The surprisingly high specificity for the non-specific D571 code presumably reflects the availability of specific ICD-10-CM codes for other subtypes of sickle cell disease without crisis, such as D57.20 (HbSC without crisis). It is also worthwhile to note that our specificity was calculated among the population of children with a claim reporting a D57x code. This would tend to understate the specificity, compared to the entire Medicaid population. Among the entire Medicaid population, the specificity would be over 99% due to the substantially increased number of true negatives.

This study has limitations; candidate case definitions were tested and validated in two state Medicaid programs. Michigan and NYS Medicaid programs may not be reflective of all of the nuances that exist between different state administrative claims databases, yet our study is strengthened by the consistency of results across two large, diverse Medicaid populations. Our results were also limited to children enrolled in Medicaid in a small timeframe; however, previous studies indicate that nearly 90% of children with SCA are enrolled in Medicaid at some point in time.[46] Our study is only able to identify children with SCA that utilize healthcare. Any child with SCA that does not have SCA-related healthcare needs would not be captured within this case definition, although we do expect this proportion of children to be small and would affect all case definitions equally. For example, we anticipate that less than 10 children with SCA would be without a D57x diagnosis code within a year; even when accounting for these children, the sensitivity of our case definition would continue to be over 90% in the Michigan Medicaid population. Finally, we did not test how this case definition would

perform among children outside of our study population age range. Specifically, we did not assess case definitions for identifying infants (i.e., <1 year of age) with SCA, or for individuals 18 years of age and older.

In conclusion, children with SCA can be accurately identified using this straightforward case definition. This methodology can be used to monitor trends and use of health services after transition to ICD-10-CM. Further studies should apply this definition to publically and commercially insured populations for prospective and continuous monitoring of opportunities to improve care among children with SCA, a vulnerable group of children at high risk for significant morbidity across the lifespan.

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Table 1. Candidate Case Definitions Developed for Identification of Children with Sickle Cell Anemia (SCA)

Definition	Description
Counts of SCA claims (D5700, D5701, D5702)	
A	≥1 claim
B	≥2 claims
C	≥3 claims
SCA-related emergency department (ED) visits (D5700, D5701, D5702)	
D	≥1 ED visit
E	≥2 ED visits
F	≥3 ED visits
SCA-related Inpatient Hospitalizations (D5700, D5701, D5702)	
G	≥1 hospitalization
H	≥2 hospitalizations
I	≥3 hospitalizations
SCA-related outpatient visits (D5700, D5701, D5702)	
J	≥1 outpatient visit
K	≥2 outpatient visits ≥30 days apart
L	≥3 outpatient visits, each ≥30 days apart
M	≥2 outpatient visits, ≥7 days apart

N	≥3 outpatient visits, each ≥7 days apart
SCA-related outpatient visits, including non-specific sickle cell code (D5700, D5701, D5702, D571)	
O	≥1 outpatient visit
P	≥2 outpatient visits, ≥30 days apart
Q	≥3 outpatient visits, each ≥30 days apart
R	≥2 outpatient visits, ≥7 days apart
S	≥3 outpatient visits, each ≥7 days apart
Counts of SCA claims, including non-specific sickle cell code (D5700, D5701, D5702, D571)	
T	≥1 claim
U	≥2 claims
V	≥3 claims
Combination	
W	≥2 SCA or D571 outpatient visits, each ≥7 days apart OR ≥1 SCA-related hospitalization

Table 2. Demographic Characteristics of Children with at least 1 D57x claim in 2016

		Michigan Medicaid		New York State Medicaid	
		Sickle cell anemia (SCA) (n=280)	**Not SCA (n=1,177)**	**SCA (n=505)**	**Not SCA (n=1,949)**
Sex	Male	137 (49%)	611 (52%)	259 (51%)	1023 (52%)
	Female	143 (51%)	566 (48%)	246 (49%)	926 (48%)
Race	Non-Hispanic Black	242 (86%)	1002 (85%)	328 (65%)	1114 (57%)
	Non-Hispanic White	7 (3%)	74(6%)	8 (2%)	53 (3%)
	Hispanic	<5	35 (3%)	78 (15%)	312 (16%)
	Unknown Race / Ethnicity	28 (10%)	59 (5%)	46 (9%)	353 (18%)
	Asian/Pacific Islander	<5	<5	<5	13 (1%)
	Other	N/A	N/A	44 (8%)	100 (5%)
	American Indian / Alaskan Native	<5	<5	<5	<5
Age (years), Mean (standard deviation)		8.7 (4.6)	6.8 (4.7)	6.5 (2.3)	6.0 (2.3)

Table 3. Measures of Performance for Candidate Case Definitions

Definition	Description	Michigan Medicaid			New York State Medicaid		
		Sensitivity	Specificity	Area under ROC Curve	Sensitivity	Specificity	Area under ROC Curve
Counts of sickle cell anemia (SCA) claims (D5700, D5701, D5702)							
A	≥1 claim	0.56	0.94	0.75	0.52	0.92	0.72
B	≥2 claims	0.48	0.96	0.72	0.33	0.97	0.65
C	≥3 claims	0.39	0.97	0.68	0.22	0.99	0.60
SCA-related emergency department (ED) visits (D5700, D5701, D5702)							
D	≥1 ED visit	0.45	0.95	0.70	0.22	0.96	0.59
E	≥2 ED visits	0.34	0.97	0.65	0.07	0.99	0.53
F	≥3 ED visits	0.20	0.98	0.59	0.03	1.00	0.51
SCA-related Inpatient Hospitalizations (D5700, D5701, D5702)							
G	≥1 hospitalization	0.33	0.99	0.66	0.34	0.98	0.66
H	≥2 hospitalizations	0.30	0.99	0.65	0.13	1.00	0.57
I	≥3 hospitalizations	0.25	0.99	0.62	0.07	1.00	0.53
SCA-related outpatient visits (D5700, D5701, D5702)							
J	≥1 outpatient visit	0.16	0.99	0.58	0.30	0.96	0.63
K	≥2 outpatient visits ≥30 days apart	0.05	1.00	0.53	0.04	1.00	0.52
L	≥3 outpatient visits, each ≥30 days apart	0.03	1.00	0.51	0.01	1.00	0.51
M	≥2 outpatient visits, ≥7 days apart	0.06	1.00	0.53	0.06	1.00	0.53

N	≥3 outpatient visits, each ≥7 days apart	0.03	1.00	0.51	0.03	1.00	0.51
SCA-related outpatient visits, including non-specific sickle cell code (D5700, D5701, D5702, D571)							
O	≥1 outpatient visit	0.94	0.92	0.93	0.94	0.86	0.90
P	≥2 outpatient visits, ≥30 days apart	0.85	0.97	0.91	0.62	0.96	0.79
Q	≥3 outpatient visits, each ≥30 days apart	0.70	0.99	0.85	0.45	0.98	0.71
R	≥2 outpatient visits, ≥7 days apart	0.86	0.97	0.91	0.70	0.95	0.82
S	≥3 outpatient visits, each ≥7 days apart	0.73	0.99	0.86	0.59	0.96	0.78
Counts of SCA claims, including non-specific sickle cell code (D5700, D5701, D5702, D571)							
T	≥1 claim	0.97	0.81	0.89	0.98	0.81	0.89
U	≥2 claims	0.94	0.91	0.92	0.89	0.89	0.89
V	≥3 claims	0.91	0.93	0.92	0.78	0.92	0.85
Combination							
W	≥2 SCA or D571 outpatient visits, each ≥7 days apart OR ≥1 SCA-related hospitalization	0.87	0.96	0.92	0.75	0.94	0.84

Table 4. Measures of Performance for Definition O, Stratified by Age in Michigan Medicaid in 2016, n=1457

		Sensitivity	Specificity	Area under ROC Curve
Age Ranges	1 through 5 years, n=735	0.96	0.93	0.94
	6 through 11 years, n=431	0.93	0.91	0.92
	12 through 17 years, n=291	0.95	0.90	0.92