Hemoglobin S Monitoring Prior to Chronic Transfusion Among Children with Sickle Cell Anemia

Section 1. Basic Measure Information

1.A. Measure Name

Hemoglobin S Monitoring Prior to Chronic Transfusion Among Children with Sickle Cell Anemia

1.B. Measure Number

0214

1.C. Measure Description

Please provide a non-technical description of the measure that conveys what it measures to a broad audience.

This measure assesses the percentage of children younger than 18 years of age identified as having sickle cell anemia (SCA) and on a chronic transfusion program who received monitoring of hemoglobin S (HbS) levels immediately prior to each transfusion during the measurement year. A higher proportion indicates better performance as reflected by appropriate treatment.

Approximately 2,000 infants are born with sickle cell disease (SCD) in the United States each year, a condition that occurs predominantly in people of African and Hispanic descent. Among children with SCD, approximately 11 percent experience a stroke by 20 years of age, and another 13 percent show evidence of silent infarcts (injuries sustained by the brain without clinical symptoms). This damage to the central nervous system is a devastating complication of SCD. It occurs most frequently in children with the hemoglobin variants associated with SCA, the most serious form of SCD. Prevention is a high priority. Regularly scheduled, ongoing blood transfusions have been found to reduce the percentage of HbS to 30 percent of total hemoglobin concentration. This reduction in HbS helps prevent a first stroke in high-risk patients and recurrent stroke in patients who have already had a first stroke. Chronic transfusion therapy has also been shown to ease other complications related to SCD. Clinical guidelines indicate that chronic transfusion protocols should include regular monitoring of the HbS level before each transfusion. However, there are no existing quality measures regarding the pre-transfusion monitoring of HbS in children with sickle cell anemia on a chronic transfusion program.

This measure uses medical record data and is calculated as the percentage of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of hemoglobin S (HbS) levels immediately prior to each transfusion.

1.D. Measure Owner

The Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC).

1.E. National Quality Forum (NQF) ID (if applicable)

Not applicable.

1.F. Measure Hierarchy

Please note here if the measure is part of a measure hierarchy or is part of a measure group or composite measure. The following definitions are used by AHRQ:

1. **Please identify the name of the collection of measures to which the measure belongs (if applicable). A collection is the highest possible level of the measure hierarchy. A collection may contain one or more sets, subsets, composites, and/or individual measures.**

   This measure is part of the Q-METRIC Sickle Cell Disease Measures collection.

2. **Please identify the name of the measure set to which the measure belongs (if applicable). A set is the second level of the hierarchy. A set may include one or more subsets, composites, and/or individual measures.**

   This measure is part of the Q-METRIC Sickle Cell Disease Medical Record Data set.

3. **Please identify the name of the subset to which the measure belongs (if applicable). A subset is the third level of the hierarchy. A subset may include one or more composites, and/or individual measures.**

   Not applicable.

4. **Please identify the name of the composite measure to which the measure belongs (if applicable). A composite is a measure with a score that is an aggregate of scores from other measures. A composite may include one or more other composites and/or individual measures. Composites may comprise component measures that can or cannot be used on their own.**

   Not applicable.

1.G. Numerator Statement

The eligible population for the numerator is the number of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of HbS levels immediately prior to each transfusion during the measurement year (January 1- December 31). Eligible children are restricted to those with sickle cell anemia, as determined by hemoglobin variants identified in Table 1 (see Supporting documents), with the

appropriate ICD-9 codes documented in the medical record. The procedure codes for blood transfusions and HbS level are identified in Table 2 (see Supporting Documents).

Documentation in the medical record must include, at a minimum, a note containing the date on which each treatment was administered. Transfusions must occur at intervals of 6 weeks or less for the entire measurement year, and blood draws to test for HbS levels must occur 24 hours or less before each transfusion.

1.H. Numerator Exclusions

Children with a diagnosis in the sampled medical record indicating one of the SCD variants listed in Table 3 (see Supporting Documents) should not be included in the eligible population unless there is also a diagnosis for a sickle cell variant listed in Table 1 (see Supporting Documents).

1.I. Denominator Statement

The eligible population for the denominator is the number of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program during the measurement year (January 1-December 31). Eligible children are restricted to those with sickle cell anemia, as determined by the hemoglobin variants identified in Table 1 (see Supporting Documents), with the appropriate ICD-9 codes documented in the medical record. The procedure codes for blood transfusions are identified in Table 2 (see Supporting Documents). Documentation in the medical record must include, at a minimum, a note containing the date on which each treatment was administered. Transfusions must occur at intervals of 6 weeks or less for the entire measurement year.

1.J. Denominator Exclusions

Children with a diagnosis in the sampled medical record indicating one of the SCD variants listed in Table 3 (see Supporting Documents) should not be included in the eligible population unless there is also a diagnosis for a sickle cell variant listed in Table 1 (see Supporting Documents).

1.K. Data Sources

Check all the data sources for which the measure is specified and tested.

Paper medical record; electronic medical record.

If other, please list all other data sources in the field below.

Not applicable.

Section 2: Detailed Measure Specifications

Provide sufficient detail to describe how a measure would be calculated from the recommended data sources, uploading a separate document (+ Upload attachment) or a link to a URL. Examples of detailed measure specifications can be found in the CHIPRA

Initial Core Set Technical Specifications Manual 2011 published by the Centers for Medicare & Medicaid Services. Although submission of formal programming code or algorithms that demonstrate how a measure would be calculated from a query of an appropriate electronic data source are not requested at this time, the availability of these resources may be a factor in determining whether a measure can be recommended for use.

See Supporting Documents for technical specifications for this measure.

Section 3. Importance of the Measure

In the following sections, provide brief descriptions of how the measure meets one or more of the following criteria for measure importance (general importance, importance to Medicaid and/or CHIP, complements or enhances an existing measure). Include references related to specific points made in your narrative (not a free-form listing of citations).

3.A. Evidence for General Importance of the Measure

Provide evidence for all applicable aspects of general importance:

- **Addresses a known or suspected quality gap and/or disparity in quality (e.g., addresses a socioeconomic disparity, a racial/ethnic disparity, a disparity for Children with Special Health Care Needs (CSHCN), a disparity for limited English proficient (LEP) populations).**

- **Potential for quality improvement (i.e., there are effective approaches to reducing the quality gap or disparity in quality).**

- **Prevalence of condition among children under age 21 and/or among pregnant women.**

- **Severity of condition and burden of condition on children, family, and society (unrelated to cost).**

- **Fiscal burden of measure focus (e.g., clinical condition) on patients, families, public and private payers, or society more generally, currently and over the life span of the child.**

- **Association of measure topic with children's future health – for example, a measure addressing childhood obesity may have implications for the subsequent development of cardiovascular diseases.**

- **The extent to which the measure is applicable to changes across developmental stages (e.g., infancy, early childhood, middle childhood, adolescence, young adulthood).**

Sickle Cell Disease Prevalence and Incidence

SCD is one of the most common genetic disorders in the United States (Kavanagh, Sprinz, Vinci, et al., 2011). The National Heart, Lung and Blood Institute (NHLBI) estimates that 2,000 infants are born with SCD in the United States each year (NHLBI, 2002). SCD affects 70,000-100,000

children and adults in the United States, predominantly those of African and Hispanic descent (Hassell, 2010).

Sickle Cell Disease Pathology and Severity

Vaso-occlusion (the sudden blockage of a blood vessel caused by the sickle shape of abnormal blood cells) is responsible for most complications of SCD, including pain episodes, sepsis, stroke, acute chest syndrome, priapism, leg ulcers, osteonecrosis and renal insufficiency (Steinberg, 1999). In addition, SCD can have hemolytic and infectious complications that result in morbidity and mortality in children with SCD (Kavanagh, et al., 2011).

Sickle Cell Disease Burden in Daily Life

The effect of SCD on children and families is significant; severe pain episodes and hospitalizations restrict daily activities and reflect negatively on school attendance and performance, as well as on sleep and social activities (Alvim, Viana, Pires, et al., 2005; Lemanek, Ranalli, Lukens, 2009). Although medical management of SCD continues to improve over time, 196 U.S. children died from SCD-related causes between 1999 and 2002 (Yanni, Grosse, Yang, et al., 2009).

Sickle Cell Disease Cost

In a study of health care utilization among low income children with SCD between 2004 and 2007, 27 percent of these children required inpatient hospitalization, and 39 percent used emergency care during a year. Of these children, 63 percent averaged one well-child visit per year, and 10 percent had at least one outpatient visit with a specialist (Raphael, Dietrich, Whitmire, et al., 2009). Patients with SCD use many parts of the health care system, incurring significant costs. In 2009, mean hospital charges for children with SCD and a hospital stay were $23,000 for children with private insurance and $18,200 for children enrolled in Medicaid (AHRQ, 2012). Kauf and colleagues estimate the lifetime cost of health care per patient with SCD to be approximately $460,000 (Kauf, Coates, Huazhi, et al., 2009). Estimates place the cost of chronic transfusion therapy at up to $400,000 per patient decade (Wayne, Schoenike, Pegelow, 2000). Researchers have suggested that this expense, along with the associated risks of ongoing transfusion and its lack of permanent benefit, should be considered as part of any cost/efficacy analyses regarding alternative SCD therapies, including hydroxyurea and bone marrow transplants (Wayne, et al., 2000).

Outcomes of Hemoglobin S Monitoring Prior to Chronic Transfusion

Stroke is a devastating complication of SCD, one that may occur without warning or in conjunction with other SCD-related illnesses. Approximately one child in 10 with SCD will have a stroke before adulthood, and the frequency of stroke is highest in these children before the age of 9 years (Ohene-Frempong, Weiner, Sleeper, et al., 1998; Pack-Mabien, Haynes Jr, 2009; Steinberg, 1999). The first line of prevention is a program of regular (chronic) blood transfusions, an approach that has been shown to reduce the risk of first and recurrent stroke in children. The marked decrease in stroke rates has made chronic transfusion therapy a standard of care (Aygun, Murray, Schultz, et al., 2009). Ongoing transfusions are used to address other complications of SCD, as well.

Used correctly, transfusions can prevent organ damage and save the lives of children with SCD. Used unwisely, transfusions can result in serious complications. Usually, the goal of chronic transfusion therapy is to maintain the HbS level at approximately 30 percent of total hemoglobin concentration. The pre-transfusion check of the HbS level helps ensure that the transfusions have been adequate to keep the HbS levels below 30 percent up to the time of the next transfusion. If they are higher at the pre-transfusion check, then the transfusion regimen must be altered to ensure that goal is met.

Reducing the number of sickled red blood cells (HbS) increases the oxygen-carrying capacity of the blood, as it is these particular red blood cells that cause the arterial blockages underlying many SCD complications. Transfusions are usually repeated every 3 or 4 weeks. Chronic transfusion therapy is indicated when the avoidance of potentially serious medical complications justifies the risk of transfusion, which include infection, iron overload, and alloimmunization (an immune response generated in one individual by an alloantigen from another) (NHLBI, 2002). Chronic transfusion therapy is provided to 90 percent of patients with sickle cell anemia. Neurologic injury causes serious morbidity in over 25 percent of patients with sickle cell anemia and is the most common indication for chronic transfusion programs. In addition to the use of long-term transfusion for stroke prevention, efficacy for the following conditions has expanded the indications for chronic transfusion: acute chest syndrome, chronic pain syndrome, pulmonary hypertension, anemia associated with renal failure, and end-stage organ failure. The use of prophylactic transfusion in patients with SCD has increased because of the profound effects that transfusions have on improving quality of life (Adams, McKie, Hsu, et al., 1998; NHLBI, 2002; Vichinsky, Luban, Wright, et al., 2001).

Intensive, ongoing efforts are needed to identify genetic and environmental factors that confer risk for SCD severity. This will help families decide and commit to appropriate therapies. Families should be made fully aware of all the morbid complications of SCD, especially the clinically silent changes in the central nervous system that can insidiously degrade a child's intellectual abilities. All therapies should be studied, including chronic transfusion, for the ability to address these problems (Reed, Vichinsky, 2001).

Performance Gap

Successful participation in a chronic transfusion program requires commitment from children and their families, which can make adherence challenging. Transfusions are scheduled at roughly monthly intervals; these frequent appointments are time consuming and require the presence of a parent or guardian, as well as regular access to dependable transportation. Children on a chronic transfusion program often need to be scheduled for other medical visits related to SCD; the stress of coordinating these appointments can be significant. There are also physical issues associated with transfusion, including repeated needle sticks and intravenous line access, which can be burdensome for children. If these demands reduce adherence to the transfusion program, the effectiveness of the transfusion protocol for the child may be compromised. Checking the HbS levels provides an indication of the appropriateness of the interval between transfusions, as well as an indication for the volume of transfusion required at each transfusion episode. One solution for time management issues is to schedule chronic transfusion during evenings or weekends to avoid interfering with school and work (NHLBI, 2002).

It is possible that not enough children with SCD are being directed toward chronic transfusion because of gaps in screening for stroke risk. A 2008 study at the Texas Children's Sickle Cell Center reported that only 45 percent of children with SCD were screened for stroke risk with annual transcranial Doppler (TCD) ultrasonography. Patients with private insurance were three times as likely to complete more than 50 percent of ordered TCD screenings as patients with Medicaid (Raphael, Shetty, Liu, et al., 2008). In a retrospective cohort study of children aged 2-16 years with SCD enrolled in Tennessee Medicaid, Eckrich and colleagues found that rates of TCD screening increased over time, with 2.5 percent receiving TCD screening in 1997 and 68.3 percent receiving screening in 2008. However, 31 percent of study participants received no TCD screening during the entire 11-year study period (1997-2008) (Eckrich, Wang, Yang, et al., 2013). Interviews with 36 caregivers of children with SCD revealed that 22 percent of caregivers had no knowledge of TCD screening, and 42 percent were unaware that TCD screening should be performed yearly (Bollinger, Nire, Rhodes, et al., 2011).

3.B. Evidence for Importance of the Measure to Medicaid and/or CHIP

Comment on any specific features of this measure important to Medicaid and/or CHIP that are in addition to the evidence of importance described above, including the following:

- **The extent to which the measure is understood to be sensitive to changes in Medicaid or CHIP (e.g., policy changes, quality improvement strategies).**

- **Relevance to the Early and Periodic Screening, Diagnostic and Treatment benefit in Medicaid (EPSDT).**

- **Any other specific relevance to Medicaid/CHIP (please specify).**

Sickle Cell Disease and Medicaid/CHIP

This measure is relevant to Medicaid because the majority of children with SCD are also enrolled in Medicaid. In 2009, 67 percent of children with SCD discharged from the hospital were enrolled in Medicaid, while 25 percent had private insurance (AHRQ, 2012). Medicaid enrollment often serves as a marker of poverty. The large number of children with SCD on Medicaid suggests some of these patients may be receiving suboptimum treatment because of unstable living situations, despite the provision of anticipatory guidance. These children may not be receiving stroke screenings regularly, and they may experience delays in being taken for medical care if family situations are such that work responsibilities, school commitments for siblings, or lack of transportation make seeking prompt medical attention difficult (Tanabe, Dias, Gorman, 2013).

3.C. Relationship to Other Measures (if any)

Describe, if known, how this measure complements or improves on an existing measure in this topic area for the child or adult population, or if it is intended to fill a specific gap in an existing measure category or topic. For example, the proposed measure may enhance an existing measure in the initial core set, it may lower the age range for an existing adult-focused measure, or it may fill a gap in measurement (e.g., for asthma care quality, inpatient care measures).

Currently, there are no quality measures for the diagnosis, assessment, or treatment of pediatric SCD.

Section 4. Measure Categories

CHIPRA legislation requires that measures in the initial and improved core set, taken together, cover all settings, services, and topics of health care relevant to children. Moreover, the legislation requires the core set to address the needs of children across all ages, including services to promote healthy birth. Regardless of the eventual use of the measure, we are interested in knowing all settings, services, measure topics, and populations that this measure addresses. These categories are not exclusive of one another, so please indicate "Yes" to all that apply.

Does the measure address this category?

a. **Care Setting – ambulatory:** Yes.
b. **Care Setting – inpatient:** No.
c. **Care Setting – other – please specify:** No.
d. **Service – preventive health, including services to promote healthy birth:** Yes.
e. **Service – care for acute conditions:** No.
f. **Service – care for children with special health care needs/chronic conditions:** Yes.
g. **Service – other (please specify):** No.
h. **Measure Topic – duration of enrollment:** No.
i. **Measure Topic – clinical quality:** Yes.
j. **Measure Topic – patient safety:** No.
k. **Measure Topic – family experience with care:** No.
l. **Measure Topic – care in the most integrated setting:** No.
m. **Measure Topic other (please specify):** Not applicable.
n. **Population – pregnant women:** Not applicable.
o. **Population – neonates (28 days after birth) (specify age range):** Yes; all ages in this range.
p. **Population – infants (29 days to 1 year) (specify age range):** Yes; all ages in this range.
q. **Population – pre-school age children (1 year through 5 years) (specify age range):** Yes; all ages in this range.
r. **Population – school-aged children (6 years through 10 years) (specify age range):** Yes; all ages in this range.
s. **Population – adolescents (11 years through 20 years) (specify age range):** Yes; adolescents 11-17 years.
t. **Population – other (specify age range):** Not applicable.
u. **Other category (please specify):** Not applicable.

Section 5. Evidence or Other Justification for the Focus of the Measure

The evidence base for the focus of the measures will be made explicit and transparent as part of the public release of CHIPRA deliberations; thus, it is critical for submitters to specify the scientific evidence or other basis for the focus of the measure in the following sections.

5.A. Research Evidence

Research evidence should include a brief description of the evidence base for valid relationship(s) among the structure, process, and/or outcome of health care that is the focus of the measure. For example, evidence exists for the relationship between immunizing a child or adolescent (process of care) and improved outcomes for the child and the public. If sufficient evidence existed for the use of immunization registries in practice or at the State level and the provision of immunizations to children and adolescents, such evidence would support the focus of a measure on immunization registries (a structural measure).

Describe the nature of the evidence, including study design, and provide relevant citations for statements made. Evidence may include rigorous systematic reviews of research literature and high-quality research studies.

This measure focuses on a clinical process (monitoring HbS levels prior to chronic transfusion) that, if followed using the results of the testing, produces a desirable clinical outcome (a reduced rate of stroke in children with SCD). The measure highlights where providers or health systems are falling short in providing health care maintenance for children with SCD.

The body of evidence addresses the prevention of first and recurrent stroke in children with SCD, using an ongoing regimen of regular transfusions to reduce the concentration of abnormal HbS to 30 percent of total hemoglobin at the pre-transfusion assessment. Maintaining this level requires regular monitoring of HbS levels prior to each transfusion. Evidence also covers the use of chronic transfusion to treat other conditions associated with SCD, including vaso-occlusive pain and acute chest syndrome. Table 4 (see Supporting Documents) summarizes several key sources of evidence for this measure, using the U.S. Preventive Services Task Force (USPSTF) rankings.

5.B. Clinical or Other Rationale Supporting the Focus of the Measure (optional)

Provide documentation of the clinical or other rationale for the focus of this measure, including citations as appropriate and available.

The abnormal hemoglobin found in sickle cells (HbS) causes red blood cells to develop a crescent (sickle) shape. Because these cells are stiff, sticky, and misshapen, they can block blood flow, which leads to many complications, including stroke. Stroke is a leading cause of death in children with SCD, occurring most often in those with sickle cell anemia (Ohene-Frempong, et al., 1998). In children with SCD, stroke is often caused by blockages of the intracranial internal carotid and middle cerebral arteries. Studies have shown that children with SCD have an approximately 11 percent risk of stroke by the time they are 20 years old (Adams, et al., 1998).

In children with sickle cell anemia, peak incidence of stroke occurs around age 7 years (Wang, Langston, Steen, et al., 1998).

When the oxygen supply to the brain falls below a critical level based on need, brain dysfunction occurs. There is evidence that oxygen demands are higher in children than in adults, making the child with SCD who has significant anemia at particular risk (NHLBI, 2002). Risk factors for stroke include a history of transient ischemia attack, elevated systolic blood pressure, elevated steady-state leukocyte count, severe anemia, and prior history of acute chest syndrome (Ohene-Frempong, et al., 1998).

A blood transfusion to reduce the concentration of hemoglobin S to 30 percent of total hemoglobin is a reliable method of preventing stroke and other complications of SCD. The increased volume of healthy red blood cells reduces the percentage of sickle cells in the blood, thereby reducing arterial blockages and increasing the oxygen-carrying capacity of the blood. Clinically, the microvascular perfusion of tissues is improved (NHLBI, 2002).

While not perfect, chronic transfusion is clearly an important therapy. Among children with sickle cell anemia and a first stroke who are not receiving chronic transfusion therapy, 50 percent have another stroke within 3 years, compared with 10 percent of those who receive regular transfusions (Steinberg, 1999). Chronic transfusion therapy does not completely eliminate risk and may need to be continued indefinitely.

There is the potential for problems from iron overload, infections, and alloimmunization. This has prompted some interest in a modified program allowing HbS to drift up to around 50 percent after several years of transfusion (Steinberg, 1999).

Section 6. Scientific Soundness of the Measure

Explain the methods used to determine the scientific soundness of the measure itself. Include results of all tests of validity and reliability, including description(s) of the study sample(s) and methods used to arrive at the results. Note how characteristics of other data systems, data sources, or eligible populations may affect reliability and validity.

6.A. Reliability

Reliability of the measure is the extent to which the measure results are reproducible when conditions remain the same. The method for establishing the reliability of a measure will depend on the type of measure, data source, and other factors.

Explain your rationale for selecting the methods you have chosen, show how you used the methods chosen, and provide information on the results (e.g., the Kappa statistic). Provide appropriate citations to justify methods.

This measure is based on medical records data; reliability testing is described here.

Data and Methods

Our testing data consisted of an audit of medical records from the three largest centers serving SCD patients in Michigan during 2012: Children's Hospital of Michigan (CHM, Detroit), Hurley Medical Center (Hurley, Flint), and the University of Michigan Health System (UMHS, Ann Arbor). Combined, these sites treat the majority of children with SCD in Michigan. Medical records for all children with SCD meeting the measure specification criteria during the measurement year were abstracted at each site. Abstracting was conducted in two phases; during Phase 1, 435 records were abstracted among the three sites. In Phase 2, an additional 237 cases were abstracted at one site. In total, 672 unique records were reviewed for children with SCD to test this measure.

Reliability of medical record data was determined through re-abstraction of patient record data to calculate the inter-rater reliability (IRR) between abstractors. Broadly, IRR is the extent to which the abstracted information is collected in a consistent manner. Low IRR may be a sign of poorly executed abstraction procedures, such as ambiguous wording in the data collection tool, inadequate abstractor training, or abstractor fatigue. For this project, the medical record data collected by two nurse abstractors were compared.

Measuring IRR at the beginning of the abstraction is imperative to identify any misinterpretations early on. It is also important to assess IRR throughout the abstraction process to ensure that the collected data maintain high reliability standards. Therefore, the IRR was evaluated during Phase 1 at each site to address any reliability issues before beginning data abstraction at the next site.

IRR was determined by calculating both percent agreement and Kappa statistics. While abstraction was still being conducted at each site, IRR assessments were conducted for 5 percent of the total set of unique patient records that were abstracted during Phase 1 of data collection. Two abstractors reviewed the same medical records; findings from these abstractions were then compared, and a list of discrepancies was created.

Three separate IRR meetings were conducted, all of which included a review of multiple SCD measures that were being evaluated. Because of eligibility criteria, not all patients were eligible for all measures. Therefore, records for IRR were not chosen completely at random; rather, records were selected to maximize the number of measures assessed for IRR at each site.

Results

For this measure, 8 of 435 unique patient records from Phase 1 of the abstraction process were assessed for IRR across the three testing sites. Overall, these eight records included 45 transfusions during the measurement year.

Table 5 (see Supporting Documents) shows the percent agreement and Kappa statistics for the measure numerator for each site and across all sites. The overall agreement for the measure is 98 percent, and the Kappa is 0.95, indicating that a high IRR level was achieved.

Discrepancies

If discrepancies between abstractors were found, the abstractors and a study team member reopened the electronic medical record to review each abstractor's response and determine the correct answer. For this measure, of the 45 entries reviewed for IRR, one discrepancy was found at one site. After discussion, a consensus result was obtained, and the inconsistent record was corrected for the final dataset.

6.B. Validity

Validity of the measure is the extent to which the measure meaningfully represents the concept being evaluated. The method for establishing the validity of a measure will depend on the type of measure, data source, and other factors.

Explain your rationale for selecting the methods you have chosen, show how you used the methods chosen, and provide information on the results (e.g., R2 for concurrent validity).

The validity of this measure was determined from two perspectives: face validity and validity of medical record data.

Face Validity

Face validity is the degree to which the measure construct characterizes the concept being assessed. The face validity of this measure was established by a national panel of experts and advocates for families of children with SCD convened by Q-METRIC. The Q-METRIC expert panel included nationally recognized experts in SCD, representing hematology, pediatrics, and SCD family advocacy. In addition, measure validity was considered by experts in State Medicaid program operations, health plan quality measurement, health informatics, and health care quality measurement. In total, the Q- METRIC SCD panel included 14 experts, providing a comprehensive perspective on SCD management and the measurement of quality metrics for States and health plans.

The Q-METRIC expert panel concluded that this measure has a high degree of face validity through a detailed review of concepts and metrics considered to be essential to effective SCD management and treatment. Concepts and draft measures were rated by this group for their relative importance. This measure was highly rated, receiving an average score of 8.2 (with 9 as the highest possible score).

Validity of Abstracted Data

This measure was tested using medical record data, which is considered the gold standard for clinical information; our findings indicate that these data have a high degree of face validity and reliability. We tested this measure among a total of 38 children younger than 18 years of age with sickle cell anemia (Table 6; see Supporting Documents). Overall, 45 percent of children with sickle cell anemia and on a chronic transfusion program received appropriate monitoring of HbS levels immediately prior to transfusion (range: 0 percent- 52 percent).

Section 7. Identification of Disparities

CHIPRA requires that quality measures be able to identify disparities by race, ethnicity, socioeconomic status, and special health care needs. Thus, we strongly encourage nominators to have tested measures in diverse populations. Such testing provides evidence for assessing measure's performance for disparities identification. In the sections below, describe the results of efforts to demonstrate the capacity of this measure to produce results that can be stratified by the characteristics noted and retain the scientific soundness (reliability and validity) within and across the relevant subgroups.

7.A. Race/Ethnicity

The measure was tested using medical records from the three largest centers serving SCD patients in Michigan during 2012: Children's Hospital of Michigan, Hurley Medical Center, and the University of Michigan Health System. Combined, these centers serve the vast majority of SCD patients in Michigan. While race and ethnicity data were not abstracted as part of the medical record review process, information is available from the State of Michigan for its entire population of births with an initial newborn screening result indicating SCD from 2004 to 2008. Table 7 (see Supporting Documents) summarizes the distribution across race and ethnicity groups for all SCD births in Michigan during that time period.

7.B. Special Health Care Needs

The medical records data abstracted for this study do not include indicators of special health care needs.

7.C. Socioeconomic Status

The medical records data abstracted for this study do not include indicators of socioeconomic status.

7.D. Rurality/Urbanicity

The medical records data abstracted for this study do not include indicators of urban/rural residence.

7.E. Limited English Proficiency (LEP) Populations

The medical records data abstracted for this study do not include indicators of LEP.

Section 8. Feasibility

Feasibility is the extent to which the data required for the measure are readily available, retrievable without undue burden, and can be implemented for performance measurement. Using the following sections, explain the methods used to determine the feasibility of implementing the measure.

8.A. Data Availability

1. What is the availability of data in existing data systems? How readily are the data available?

This measure is based on review of medical record data. The medical chart audit included records from the three largest centers serving SCD patients in Michigan during 2012: Children's Hospital of Michigan, Hurley Medical Center, and the University of Michigan Health System. Data were abstracted from medical record systems at two sites that use EHRs (both Epic systems) and from one site using paper charts.

Medical records for 100 percent of children with SCD meeting the measure specification criteria during the measurement year were abstracted from each hospital. In total, 672 unique records were reviewed; 38 records (6 percent) met denominator criteria for this measure.

Based on the abstracted chart data, the rate was calculated as the percentage of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of HbS levels immediately prior to each transfusion (45 percent). Measure numerator (17) divided by denominator (38); (see Table 6 in the Supporting Documents).

Medical record abstraction for this measure was accomplished with a data collection tool developed using LimeSurvey software (version 1.92, formerly PHPSurveyor). LimeSurvey is an open-source online application based in MySQL that enables users to develop and publish surveys, as well as collect responses. The tool was piloted to determine its usability and revised as necessary. The technical specifications for this measure also underwent revisions following pilot testing.

Data abstraction was completed by experienced nurse abstractors who had undergone training for each medical record system used, electronic and paper. Abstractors participated in onsite training during which the measure was discussed at length to include the description, calculation, definitions, eligible population specification, and exclusions. Following training, abstractors were provided with a coded list of potentially eligible cases from each of the sites. To abstract all pertinent data, two nurse abstractors reviewed the electronic and paper medical records. In addition to the specific data values required for this measure, key patient characteristics, such as date of birth and hemoglobin variant type, were also collected.

Abstraction Times

In addition to calculating IRR, the study team assessed how burdensome it was to locate and record the information used to test this measure by having abstractors note the time it took to complete each record. During Phase 1, on average, the abstractors spent 31 minutes per eligible SCD case abstracting the data for this measure, with times ranging from 2-240 minutes.

2. If data are not available in existing data systems or would be better collected from future data systems, what is the potential for modifying current data systems or creating new data systems to enhance the feasibility of the measure and facilitate implementation?

The proposed measure was determined to be feasible by Q-METRIC using medical record data from the three largest centers serving SCD patients in Michigan during 2012. Although paper charts were used at one of the sites, this was not found to be a barrier. In fact, the average time spent abstracting records for paper charts (30 minutes) was only slightly more than the 29-minute average reported at one center using EHRs and slightly less than the 31-minute average reported for the other site with EHRs.

8.B. Lessons from Use of the Measure

1. Describe the extent to which the measure has been used or is in use, including the types of settings in which it has been used, and purposes for which it has been used.

To our knowledge, this measure is not currently in use anywhere in the United States.

2. If the measure has been used or is in use, what methods, if any, have already been used to collect data for this measure?

Not applicable.

3. What lessons are available from the current or prior use of the measure?

Not applicable.

Section 9. Levels of Aggregation

CHIPRA states that data used in quality measures must be collected and reported in a standard format that permits comparison (at minimum) at State, health plan, and provider levels. Use the following table to provide information about this measure's use for reporting at the levels of aggregation in the table.

For the purpose of this section, please refer to the definitions for provider, practice site, medical group, and network in the Glossary of Terms.

If there is no information about whether the measure could be meaningfully reported at a specific level of aggregation, please write "Not available" in the text field before progressing to the next section.

Level of aggregation (Unit) for reporting on the quality of care for children covered by Medicaid/ CHIP†:

State level Can compare States

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
No.

Data Sources: **Are data sources available to support reporting at this level?**
No.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
Not applicable.

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
Not applicable.

Other geographic level: Can compare other geographic regions (e.g., MSA, HRR)

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
No.

Data Sources: **Are data sources available to support reporting at this level?**
No.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
Not applicable.

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
Not applicable.

Medicaid or CHIP Payment model: Can compare payment models (e.g., managed care, primary care case management, FFS, and other models)

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
No.

Data Sources: **Are data sources available to support reporting at this level?**
No.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
Not applicable.

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
Not applicable.

Health plan*: Can compare quality of care among health plans.

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
No.

Data Sources: **Are data sources available to support reporting at this level?**
No.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
Not applicable.

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
Not applicable.

Provider Level
Individual practitioner: Can compare individual health care professionals

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
No.

Data Sources: **Are data sources available to support reporting at this level?**
No.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
Not applicable.

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
Not applicable.

Provider Level
Hospital: Can compare hospitals

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
Yes.

Data Sources: **Are data sources available to support reporting at this level?**
Yes.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
The sample would include all children with clinical documentation of sickle cell anemia (see Table 1 in the Supporting Documents).

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
None identified.

Provider Level
Practice, group, or facility:** Can compare: (i) practice sites; (ii) medical or other professional groups; or (iii) integrated or other delivery networks

Intended use: **Is measure intended to support meaningful comparisons at this level? (Yes/No)**
No.

Data Sources: **Are data sources available to support reporting at this level?**
No.

Sample Size: **What is the typical sample size available for each unit at this level? What proportion of units at this level of aggregation can achieve an acceptable minimum sample size?**
Not applicable.

In Use: **Have measure results been reported at this level previously?**
No.

Reliability & Validity: **Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**
No.

Unintended consequences: **What are the potential unintended consequences of reporting at this level of aggregation?**
Not applicable.

Section 10. Understandability

CHIPRA states that the core set should allow purchasers, families, and health care providers to understand the quality of care for children. Please describe the usefulness of this measure toward achieving this goal. Describe efforts to assess the understandability of this measure (e.g., focus group testing with stakeholders).

This measure provides families with a straightforward measure to assess how well basic levels of comprehensive care are being provided for children with SCD. Low rates for dependable monitoring of HbS levels prior to each occurrence of chronic transfusion are easily understood to be unsatisfactory. The simplicity of the measure likewise makes it a straightforward guide for providers and purchasers to assess how well comprehensive care is managed in children with SCD.

This measure has not been assessed for comprehension. The primary information needed for this measure comes from medical records data and includes basic demographics, diagnostic codes,

procedure codes, and dates, all of which are widely available. The nurse abstractors testing the measure provided feedback to refine the abstraction tool and thus the specifications. These changes are reflected in the final documentation.

Section 11. Health Information Technology

Please respond to the following questions in terms of any health information technology (health IT) that has been or could be incorporated into the measure calculation.

11.A. Health IT Enhancement

Please describe how health IT may enhance the use of this measure.

Aspects of health IT, such as LOINC codes, will be able to be enhanced based on this measure. Health IT and process control dashboards may allow better compliance with these measures. Decision support could be created to focus on the checkout process, and computerized order entry (CPOE) could include order sets to enable easy ordering of these blood tests.

11.B. Health IT Testing

Has the measure been tested as part of an electronic health record (EHR) or other health IT system?

Yes.

If so, in what health IT system was it tested and what were the results of testing?

This measure was tested using EHR review conducted at two major SCD treatment facilities in Michigan using the Epic EHR system. The third facility used paper medical records for outpatient visits.

11.C. Health IT Workflow

Please describe how the information needed to calculate the measure may be captured as part of routine clinical or administrative workflow.

These data are likely already recorded in the EHR. Lab test orders or results should be easily recovered from the EHR with a date stamp matching that of the visit.

11.D. Health IT Standards

Are the data elements in this measure supported explicitly by the Office of the National Coordinator for Health IT Standards and Certification (ONC) criteria (see healthit.hhs.gov/portal/server.pt/community/healthit_hhs_gov__standards_ifr/1195)?

Yes.

If yes, please describe.

The ONC's Health IT Standards explicitly address the receipt of laboratory results and other diagnostic tests into EHRs, which are directly relevant to this measure. In addition, these standards indicate the requirement for EHRs to track specific patient conditions, such as SCD. The ONC standards include the following specific requirements in the Certification criteria (ONC, 2010) pertaining to Stage 2 Meaningful Use requirements include:

Stage 2 (beginning in 2013): CMS has proposed that its goals for the Stage 2 meaningful use criteria, expand upon the Stage 1 criteria to encourage the use of health IT for continuous quality improvement at the point of care. In addition, the exchange of information in the most structured format possible is encouraged. This can be accomplished through mechanisms such as the electronic transmission of orders entered using computerized provider order entry (CPOE) and the electronic transmission of diagnostic test results. Electronic transmission of diagnostic test results include a broad array of data important to quality measurement, such as blood tests, microbiology, urinalysis, pathology tests, radiology, cardiac imaging, nuclear medicine tests, and pulmonary function tests.

Incorporate clinical lab-test results into the EHR as structured data:

1. Electronically receive clinical laboratory results in a structured format and display such results in human readable format.

2. Electronically display in human readable format any clinical laboratory tests that have been received with LOINC® codes.

3. Electronically display all the information for a test report specified at 42 CFR 493.1291(C)(1) through (7).

Generate lists of patients by specific conditions to use for quality improvement reduction of disparities outreach:

4. Enable a user to electronically update a patient's record based upon received laboratory test results. Enable a user to electronically select, sort, retrieve, and output a list of patients and patients' clinical information, based on user-defined demographic data, medication lists, and specific conditions.

11.E. Health IT Calculation

Please assess the likelihood that missing or ambiguous information will lead to calculation errors.

Missing or ambiguous information in the following areas could lead to missing cases or calculation errors:

1. Child's date of birth.

2. ICD-9 codes selected to indicate sickle cell anemia/SCD.

3. Data and time of treatment.

4. Type of tests administered.

5. Date(s) of tests performed.

6. Care setting.

11.F. Health IT Other Functions

If the measure is implemented in an EHR or other health IT system, how might implementation of other health IT functions (e.g., computerized decision support systems in an EHR) enhance performance characteristics on the measure?

Displaying real-time updates of these indicators for patients and providers to see will be transformative for care, and would be one of the best ways to enhance performance. Alternatively, integrating these measures into views that might be seen by health care administrators will motivate changes in workflow or in operating models that can hardwire these orders into the provision of care for these patients.

Section 12. Limitations of the Measure

Describe any limitations of the measure related to the attributes included in this CPCF (i.e., availability of measure specifications, importance of the measure, evidence for the focus of the measure, scientific soundness of the measure, identification of disparities, feasibility, levels of aggregation, understandability, health information technology).

This measure assesses the percentage of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of hemoglobin S (HbS) levels immediately prior to each transfusion during the measurement year. A higher proportion indicates better performance as reflected by appropriate treatment.

This measure is implemented with medical record data; it was tested with electronic and paper medical records. The primary information needed for this measure includes date of birth, diagnosis codes, and procedure codes and dates. These data are available, although obtaining them may require a restricted-use data agreement. It also required the development of an abstraction tool and the use of qualified nurse abstractors. Continuing advances in the development and implementation of electronic medical records may establish the feasibility of regularly implementing this measure with data supplied by electronic medical records.

Section 13. Summary Statement

Provide a summary rationale for why the measure should be selected for use, taking into account a balance among desirable attributes and limitations of the measure. Highlight specific advantages that this measure has over alternative measures on the same topic that were considered by the measure developer or specific advantages that this measure has over existing measures. If there is any information about this measure that is important for the review process but has not been addressed above, include it here.

This measure, Hemoglobin S Monitoring Prior to Chronic Transfusion among Children with Sickle Cell Disease, assesses the percentage of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of

hemoglobin S (HbS) levels immediately prior to each transfusion during the measurement year. A higher proportion indicates better performance, as reflected by appropriate treatment. This measure was tested using medical record data. There are no existing quality measures for monitoring HbS prior to chronic transfusion in children with SCD.

Stroke is a devastating complication of SCD; one child in 10 with SCD will have a stroke before adulthood. Regularly scheduled, ongoing blood transfusions that reduce the percentage of HbS to 30 percent of total hemoglobin concentration help prevent a first stroke in high-risk patients and recurrent stroke in patients who have experienced a first stroke. Chronic transfusion therapy has also been shown to ease other complications related to SCD. Clinical guidelines indicate that chronic transfusion protocols should include regular monitoring of the HbS level before each transfusion. However, research suggests that participation in a chronic transfusion program may be challenging because of the time commitment involved. Lower levels of adherence may affect the effectiveness of the transfusion protocol for an individual child. The HbS level gives an indication of the appropriateness of the interval between transfusions, as well as an indication for the volume of transfusion required at each transfusion episode.

Q-METRIC tested this measure among a total of 38 children younger than 18 years of age with sickle cell anemia. Overall, 45 percent of children with sickle cell anemia and on a chronic transfusion program received appropriate monitoring of HbS levels immediately prior to transfusion (range: 0 percent-52 percent).

This measure provides families, providers, and purchasers with a straightforward means of assessing how well basic levels of comprehensive care are being provided for children with SCD. The primary information needed for this measure includes basic demographic data, dates, diagnostic codes, and procedure codes, all of which are widely available. Continuing advances in the development and implementation of health IT may establish the feasibility of regularly implementing this measure with data supplied by electronic medical records.

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Section 14: Identifying Information for the Measure Submitter

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expects that measures and full measure specifications will be made reasonably available to all interested parties. "Full measure specifications" is defined as all information that any potential measure implementer will need to use and analyze the measure, including use and analysis within an electronic health record or other health information technology. As used herein, "commercial use" refers to any sale, license or distribution of a measure for commercial gain, or incorporation of a measure into any product or service that is sold, licensed or distributed for commercial gain, even if there is no actual charge for inclusion of the measure. This statement must be signed by an individual authorized to act for any holder of copyright on each submitted measure or instrument. The authority of the signatory to provide such authorization should be described in the letter.