Anticipatory Guidance for Prevention and Management of Splenic Complications in Children with Sickle Cell Disease

Section 1. Basic Measure Information

1.A. Measure Name

Anticipatory Guidance for Prevention and Management of Splenic Complications in Children with Sickle Cell Disease

1.B. Measure Number

0227

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 4 years of age identified as having sickle cell disease (SCD) who received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care during the measurement year. A higher proportion indicates better performance, as reflected by appropriate guidance.

Approximately 2,000 infants are born with SCD in the United States each year, a condition that occurs predominantly in people of African and Hispanic descent. SCD is a chronic hematologic disorder, characterized by the presence of hemoglobin S (HbS). From infancy onward, the presence of this hemoglobin variant can lead to an array of serious medical conditions. The spleen is particularly subject to damage, and splenic problems can affect children with SCD from infancy onward. As splenic function is compromised by misshapen sickle cells, young children are susceptible to overwhelming infections (the damaged spleen cannot successfully clear bacteria) and to splenic sequestration (red blood cells are trapped in the spleen, leading to a precipitous fall in hemoglobin levels). Both conditions can escalate rapidly, ending in death if not addressed quickly. Clinical guidelines suggest that as part of comprehensive care, parents and primary caregivers should receive instructions about how to recognize splenic complications, assess spleen size, be vigilant about possible infection, and seek immediate care when symptoms warrant. However, there are no existing quality measures for anticipatory guidance regarding splenic complications in children with SCD.

This measure uses medical record data and is calculated as the percentage of children who received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications.

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 4 years of age identified as having SCD who received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care during the measurement year (January 1-December 31). Eligible children are restricted to those with SCD

variants identified in Table 1 (see Supporting Documents), based on appropriate ICD-9 codes, as documented in the medical record. Codes to identify outpatient care are listed in Table 2 (see supporting Documents).

Anticipatory guidance is any written or face-to-face verbal communication regarding the identification, prevention, and/or management of splenic complications as part of outpatient care with a patient, parent, or family member. Evidence of anticipatory guidance is determined through medical record review. Documentation in the medical record must include, at minimum, a note containing the date on which verbal or written anticipatory guidance was provided.

1.H. Numerator Exclusions

1. Children with evidence of a splenectomy should not be included in the eligible population.
2. Inpatient stays, emergency department (ED) visits, and urgent care visits are excluded from the calculation.
3. 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 4 years of age identified as having SCD who received outpatient care during the measurement year (January 1- December 31). Eligible children are restricted to those with SCD variants identified in Table 1 (see Supporting Documents), based on appropriate ICD-9 codes, as documented in the medical record. Codes to identify outpatient care are listed in Table 2 (see Supporting Documents).

1.J. Denominator Exclusions

1. Children with evidence of a splenectomy should not be included in the eligible population.
2. Inpatient stays, ED visits, and urgent care visits are excluded from the calculation.
3. 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.

Please see the Supporting Documents for detailed measure specifications.

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 Africa 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 the condition (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 children in the United States died from SCD-related causes between 1999 and 2002 (Yanni, Grosse, Yang, et al., 2009).

Sickle Cell Disease Cost

In a study of healthcare 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 healthcare 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 (Agency for Healthcare Research and Quality [AHRQ], 2012). Kauf and colleagues estimate the lifetime cost of healthcare per patient with SCD to be approximately $460,000 (Kauf, Coates, Huazhi, et al., 2009).

Outcomes of Anticipatory Guidance Regarding Prevention of Splenic Complications in Children with Sickle Cell Disease

SCD is a blood disorder that causes ongoing organ damage. The spleen, which filters blood, is particularly susceptible to the ravages of SCD, and the conditions associated with its dysfunction—overwhelming bacterial infection and splenic sequestration—are two of the main cause of death in young children with SCD. *Streptococcus pneumoniae* sepsis is the single most common cause of death in children with SCD, and mortality for splenic sequestration has been reported at 12 percent in first cases and 20 percent in second episodes (NHLBI, 2002). Parents and other primary caretakers who are educated about the symptoms and gravity of these splenic complications will be prepared to manage such situations in an appropriate and timely manner, thus improving their children's health outcomes. In the case of infection, this means taking febrile young children with SCD immediately for evaluation to a medical facility or ED where SCD expertise is available. For splenic sequestration, anticipatory guidance involves learning how to assess spleen size, being able to identify an enlarged spleen, and attending to the sudden onset of other symptoms such as pallor, weakness, and rapid heart rate and breathing. For pediatric patients who experience severe or repeated episodes of splenic sequestration, anticipatory guidelines indicate that chronic transfusion therapy or a splenectomy may be advised (American Academy of Pediatrics [AAP], 2002; NHLBI, 2002).

This measure assesses the percentage of children younger than 4 years of age identified as having SCD who received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care during the measurement year. The measure does not change across developmental stages.

Performance Gap

Fever is a common sign of illness in children, and it can be challenging to convince the parents of children with SCD about the importance of responding quickly to initially minor symptoms. But, the danger of overwhelming infection is constant, and fever may also signal acute splenic sequestration. Improved compliance with pneumococcal vaccination and penicillin prophylaxis has reduced the risk of mortality for children with SCD, and death from pneumococcal infection is rare at major SCD centers in the United States. That said, parents and other primary caregivers should not relax their vigilance about current recommendations regarding vaccinations, the use of antibiotics for prophylaxis, and anticipatory guidance about the indications and dangers of infection (NHLBI, 2002).

It is also important that providers take the time to listen to concerns voiced by the families of children with SCD so that guidance is provided in a manner that is sensitive to medical and psychosocial needs and that families have assistance in assessing available resources. Failure to consider and appreciate ethnic and cultural differences that may exist among providers, patients, and families contributes to misunderstanding and lack of trust. Education should be provided in an open, non-judgmental, mutually respectful environment. Providers should recognize that personal and cultural beliefs about illness, stress, and support systems affect the way that families respond to the challenge of raising a child with this chronic illness (Lane, Buchanan, Hutter, et al., 2001).

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).**

The majority of children with SCD are enrolled in Medicaid. In 2009, 67 percent of pediatric SCD patients discharged from the hospital were enrolled in Medicaid; only 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 prophylactic antibiotics to help prevent bacterial infections, or their caregivers may not keep regular track of spleen size. Further, if routine outpatient visits do not occur on a regular basis, caregivers may simply lack the knowledge provided by ongoing anticipatory guidance. In urgent situations requiring prompt medical attention, children may experience delays in being taken to clinics or an ED if family situations are such that work responsibilities, school commitments for siblings, or lack of transportation make seeking care 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).

There currently 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; birth to 28 days.
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; children ages 1 through 3 years.
r. **Population – school-aged children (6 years through 10 years) (specify age range):** No.
s. **Population – adolescents (11 years through 20 years) (specify age range): No.**
t. **Population – other (specify age range):** No.
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 (anticipatory guidance regarding identification, prevention, and/or management of splenic complications in children with SCD) that, if followed, results in a desirable clinical outcome (early detection and timely management of bacterial infections and splenic sequestration events). The measure highlights where providers or health systems are falling short in offering healthcare maintenance for children with SCD.

Clinical guidelines indicate that early and ongoing education should be provided to parents of children with SCD and other primary caregivers regarding the necessity of prompt medical evaluation and treatment of fever and indications of splenic sequestration. Table 4 (see Supporting Documents) summarizes several key sources of evidence for this measure, using the U.S. Preventive Services Task Force (USPSTF) rankings (criteria denoted as a note to Table 4).

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.

Sickle cells can obstruct the flow of blood to the spleen, which results in loss of function (asplenia) within the first few years of life. This not only compromises the spleen's ability to filter blood, it also undermines the availability of B-cells for antibody production. As the spleen suffers infarcts (tissue death due to lack of blood supply) and shrinks, children with SCD become susceptible to bacterial infections, especially pneumococcus, which is a major cause of death in this patient group. Parents should be aware of the importance of antibiotic prophylaxis in children under the age of 5 to prevent pneumococcal infection (AAP, 2002; Claster, Vichinsky, 2003; NHLBI, 2002).

Acute exacerbation anemia results from acute splenic sequestration complication, which is caused by intrasplenic trapping of red cells in a matter of hours. This produces a precipitous fall in hemoglobin levels and the potential for hypoxic shock. Acute splenic sequestration may be defined by a decrease of at least 2 g/dL from the steady-state hemoglobin concentration; evidence of increased erythropoiesis, such as a markedly elevated reticulocyte count; and an acutely enlarging spleen. Mild-to-moderate thrombocytopenia is often present. Severe cases rapidly progress to shock and death (AAP, 2002; NHLBI, 2002). Because the trapped red blood cells are removed from circulation, anemia and peripheral circulatory failure can develop over a matter of hours, making acute splenic sequestration one of the most dangerous complications of SCD (Emond, Collis, Darvill, et al., 1985).

Acute splenic sequestration has been reported in infants as young as 5 weeks of age, but most cases in children with SCD occur between 3 months and 5 years. Attacks are often associated with viral or bacterial infections; usual clinical manifestations are sudden weakness, pallor, rapid heart rate, rapid breathing, and abdominal fullness. Reported incidence of acute splenic sequestration ranges from 7.5 percent to 30 percent; 12 percent mortality has been reported for

first occurrences (NHLBI, 2002). Recurrent splenic sequestration is common, occurring in about half of patients who survive the first episode; a 20 percent mortality rate has been reported in these repeat cases. There are no clear prognostic factors, though the fetal hemoglobin level is lower at 6 months in children who develop this complication (NHLBI, 2002).

Treatment of acute splenic sequestration involves correcting the decreased blood volume with a red blood cell transfusion. Acute splenic sequestration can be fatal within a few hours, so emergent transfusion is necessary. Once transfusion begins, the red blood cells sequestered in the spleen are remobilized, the enlarged spleen decreases in size, and the hemoglobin level rebounds. Two approaches for managing recurrent splenic sequestration have been identified for young children with SCD: (1) chronic transfusion to keep HbS levels below 30 percent and (2) removal of the spleen to eliminate further episodes of sequestration. In general, the latter is appropriate for children over 2 years of age and the former for those younger (NHLBI, 2002). Elective splenectomy is well tolerated by children with SCD, and the risk of sepsis following removal of the spleen is low with prophylaxis and follow-up. Because episodes of splenic sequestration events tend to repeat at ever closer intervals, a policy of considering splenectomy after a first attack is justified (Emond, et al., 1985).

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 record data. Reliability testing is described in this section.

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 statistic. 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, 13 of 435 unique patient records (3 percent) from Phase 1 of the abstraction process were assessed for IRR across the three testing sites. Additionally, the records for patients with splenectomies were not eligible for review. Therefore, IRR was also assessed for this eligibility criterion. For the splenectomy criterion, 14 of 435 unique patient records (3 percent) from Phase 1 of the abstraction process were assessed for IRR across the three testing sites.

Table 5 shows the percent agreement and Kappa statistic for the measure numerator and the splenectomy eligibility criterion of this measure for each site and across all sites. The overall agreement for the numerator was 100 percent and the Kappa was 1.00, indicating a perfect IRR level was achieved. The overall agreement for the splenectomy eligibility criterion was 93 percent, and the Kappa was 0.00.

Discrepancies

When 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. After discussion, a consensus result was obtained, and inconsistent records were corrected for the final dataset.

For the splenectomy eligibility criterion, the abstractors agreed that no splenectomy had occurred for 13 of the 14 IRR records assessed for splenectomy, resulting in a high percent agreement (93 percent). However, since the abstractors found no splenectomy for those 13 records, when they did disagree, the Kappa statistic was 0.00. Upon review of the one IRR record where there was disagreement, no mention of a splenectomy was found. This 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 healthcare 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 8.0 (with 9 as the highest possible score).

Validity of Abstracted Data

This measure was tested using medical record data. This source is considered the gold standard for clinical information, and it has a high degree of face validity and reliability. This measure was tested among a total of 141 children younger than 4 years of age with SCD (Table 6; see Supporting Documents). Overall, 75 percent of children with SCD received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care (range 60-83 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

This measure was tested using medical records from the three largest centers serving SCD patients in Michigan during 2012: CHM, Detroit; Hurley, Flint; and UMHS, Ann Arbor. 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 did not include indicators of special healthcare needs.

7.C. Socioeconomic Status

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

7.D. Rurality/Urbanicity

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

7.E. Limited English Proficiency (LEP) Populations

The medical records data abstracted for this study did 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: CHM, Detroit; Hurley, Flint; and UMHS, Ann Arbor. Data were abstracted from medical record systems at two sites that use electronic health records (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; 141 records (21 percent) met denominator criteria for this measure.

Based on the abstracted chart data, the rate was calculated as the percentage of children younger than 4 years of age identified as having SCD who received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care (75 percent). Measure numerator (106) divided by denominator (141). (Table 6; see 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 specification 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 13 minutes per eligible SCD case abstracting the data for this measure, with times ranging from 3-30 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 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 (11 minutes) was only slightly more than the 10-minute average reported at one center using electronic medical records and much less than the 19-minute average reported for the other site with electronic medical records.

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 uses 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 younger than 4 years of age with clinical documentation of sickle cell disease presenting in an outpatient setting.

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 a straightforward measure to assess how well basic levels of comprehensive care are being provided for children with SCD. Low rates for the provision of anticipatory guidance 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, including anticipatory guidance, 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, dates, diagnostic codes, and procedure codes, 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.

In the short term, the predominant role of health IT in this measure is through displaying documentation templates and aggregating provider-captured anticipatory guidance information. Because most of this information is in one section of the EHR, it will be relatively easy to find and use data mining techniques to extract for the purposes of this measure. Over time, two phenomena may improve the use of the measure. First, it should be possible, given standards regarding ages and stages for providing this guidance, to develop patient-specific templates for documentation. These templates have been shown to improve compliance with recommended care practices, which will result in improved anticipatory guidance discussion. Second, the role of the patient and of patient portals is only beginning to emerge. It will likely be the case that these issues, as well as tools to help patients manage their illness, will be available through applications (apps) or personal health records that then communicate back to EHRs (or care coordinators) to improve the behaviors that these measures address.

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 electronic medical record review conducted at two major SCD treatment facilities in Michigan (outpatient records were paper at the third center).

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.

Anticipatory guidance, in general, comes in two forms: check box lists or “standardized” text created using documentation templates, and unstructured text arising from dictation or

potentially scanned documents in an EHR. This will be the primary way these data are captured in routine clinical workflow. Another, though less common, approach is to ask patients to complete forms before a visit. These forms, created by groups such as the American Academy of Pediatrics (Bright Futures) and customized for specialty-specific conditions, could be captured in any of the methods described above, and would be available to calculate the measure after neuro-linguistic programming techniques or data extraction in some other form took place.

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 ability to create patient-specific reminders for preventive services, broadly defined. While such reminders may be aimed at future appointments for services, they can also include prompts for patients to engage in activities to properly manage chronic conditions (CMS, 2012). In addition, these standards indicate the requirement for EHRs to track specific patient conditions, such as SCD. Consequently, patient reminders for activities to appropriately manage SCD could be achieved through these mechanisms, meeting the goals of anticipatory guidance preventive care. The ONC standards include the following specific requirements in the Certification criteria (ONC, 2010) pertaining to Stage 2 Meaningful Use requirements include:

> (h) Generate patient lists. 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 list, and specific conditions.

11.E. Health IT Calculation

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

1. Child's date of birth.
2. ICD-9 codes selected to indicate sickle cell disease.
3. Date and time of anticipatory guidance.
4. 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?

Performance on this measure could benefit from a number of health IT integration steps, including:

a. Documentation templates filled out by providers (or potentially scribes, in communication with providers during the visit) could improve provider behavior with respect to these issues during the visit.
b. Documentation templates created in specialty clinics could help with missed opportunities to provide this counseling in EDs, other clinic visits, home visits, or through patient-initiated contact with the health system via a patient portal or personal health application.
c. Active decision support before, during, or after the visit could prompt providers or patients about these issues.
d. EHRs could generate triggers to providers to provide this guidance (again) based on events that suggest a need to re-teach (such as after an ED visit for pain).

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 4 years of age identified as having SCD who received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care during the measurement year.

This measure is implemented with medical record data and 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, Anticipatory Guidance for Prevention and Management of Splenic Complications in Children with Sickle Cell Disease, assesses the percentage of children younger than 4 years of age identified as having SCD who received anticipatory guidance regarding the identification,

prevention, and/or management of splenic complications as part of outpatient care during the measurement year. A higher proportion indicates better performance, as reflected by appropriate guidance. This measure was tested using medical record data. There are no existing quality measures for anticipatory guidance regarding the identification, prevention, and/or management of splenic complications in children with SCD.

The spleen, which filters blood, is particularly susceptible to the ravages of SCD, and the conditions associated with its dysfunction — overwhelming bacterial infection and splenic sequestration — are two main cause of death in young children with SCD. Clinical guidelines suggest that infants with SCD should be started on prophylactic penicillin by 2 months of age and that parents should be educated about the importance of urgent medical evaluation and treatment for febrile illness and for signs and symptoms indicative of splenic sequestration. Pediatric patients with SCD who have a severe episode of acute splenic sequestration should be placed in a chronic transfusion program to keep HbS levels below 30 percent; splenectomy should be considered after the age of 2 years. However, because fever is a common occurrence in childhood, it can be challenging to convince families that this routine symptom may signal a serious condition requiring immediate response.

Q-METRIC tested this measure among a total of 141 children younger than 4 years of age with SCD. Overall, 75 percent of children with SCD received anticipatory guidance regarding the identification, prevention, and/or management of splenic complications as part of outpatient care (range: 60 percent-83 percent).

This measure provides a straightforward means of assessing how well basic levels of comprehensive care are being provided for children with SCD, including the provision of anticipatory guidance. The primary information needed for this measure includes basic demographics, dates, diagnostic codes, and procedure codes, all of which are widely available.

Continuing advances in the development and implementation of health information technology 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

First Name:	Gary L.
Last Name:	Freed, MD, MPH
Title:	Percy and Mary Murphy Professor of Pediatrics, School of Medicine
	Professor of Health Management and Policy, School of Public Health
Organization:	University of Michigan
Mailing Address:	300 North Ingalls, Room 6E08
City:	Ann Arbor
State:	MI
Postal Code:	48109
Telephone:	734-615-0616
Email:	gfreed@med.umich.edu

The CHIPRA Pediatric Quality Measures Program (PQMP) Candidate Measure Submission Form (CPCF) was approved by the Office of Management and Budget (OMB) in accordance with the Paperwork Reduction Act.

The OMB Control Number is 0935-0205 and the Expiration Date is December 31, 2015.

Public Disclosure Requirements

Each submission must include a written statement agreeing that, should U.S. Department of Health and Human Services accept the measure for the 2014 and/or 2015 Improved Core Measure Sets, full measure specifications for the accepted measure will be subject to public disclosure (e.g., on the Agency for Healthcare Research and Quality [AHRQ] and/or Centers for Medicare & Medicaid Services [CMS] websites), except that potential measure users will not be permitted to use the measure for commercial use. In addition, AHRQ 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.

AHRQ Publication No. 14(19)-P007-21-EF
June 2019