**Timeliness of Confirmatory Testing for Sickle Cell Disease**

**Section 1. Basic Measure Information**

**1.A. Measure Name**
Timeliness of Confirmatory Testing for Sickle Cell Disease

**1.B. Measure Number**
0132

**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 two major concepts:

1. The percentage of children who, having initially tested positive for sickle cell disease (SCD) through newborn screening, received confirmatory testing by 3 months of age.

2. The percentage of children whose confirmatory testing results were communicated to their families by 4 months of age. In children with SCD, illness and death can be reduced through early diagnosis, treatment of complications, systematic follow-up, and patient/parent education. In particular, timely confirmatory testing and prompt communication with families are steps that pave the way for initiating simple but life-saving treatments in young children with SCD. Neonatal screening for SCD is mandatory in the United States, but data are scarce that describe the proportion of children who receive confirmatory testing and the proportion of families who learn the results of that testing. This measure would highlight gaps where providers or health systems are falling short and encourage early and consistent treatment for all young children with SCD.

**1.D. Measure Owner**

**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 Measures administrative claims 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 numerator is the number of children receiving confirmatory testing for SCD and whose results are communicated to the family. Two rates will be reported:

1. The percentage of children who received confirmatory testing for SCD by less than or equal to 90 days of age.

2. The percentage of children with results of confirmatory testing communicated to the family by fewer than or equal to 120 days of age. For the purposes of this measure, SCD is restricted to hemoglobin screening results for a subset of conditions considered to be clinically significant (Table 1).
Table 1. Clinically Relevant Conditions for SCD Hemoglobin Screening

<table>
<thead>
<tr>
<th>Condition Name</th>
<th>Hemoglobin Screening Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hb S beta-thalassemia</td>
<td>Hb F,S,A</td>
</tr>
<tr>
<td>Hb SC-disease</td>
<td>Hb F,S,C</td>
</tr>
<tr>
<td>Hb SD-disease</td>
<td>Hb F,S,D</td>
</tr>
<tr>
<td>Hb SS-disease (sickle cell anemia)</td>
<td>Hb F,S</td>
</tr>
<tr>
<td>Hb SV-disease (unidentified variant)</td>
<td>Hb FSV</td>
</tr>
</tbody>
</table>

1.H. Numerator Exclusions

Children who died within 120 days of birth are excluded from the numerator.

Children with a diagnosis in a State’s newborn screening data indicating one of the SCD variants listed in Table 2 are specifically excluded from the numerator.

1.I. Denominator Statement

The denominator is drawn from all SCD cases reported in a State’s newborn screening program records within the measurement year.

1.J. Denominator Exclusions

Children who died within 120 days of birth are excluded from the numerator.

Children with a diagnosis in a State’s newborn screening data indicating one of the SCD variants listed in Table 2 are specifically excluded from the denominator.

Table 2. Excluded Initial Hemoglobin Screening Results

<table>
<thead>
<tr>
<th>Condition Name</th>
<th>Hemoglobin Screening Result</th>
<th>ICD-9 Code(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hb beta zero-thalassemia</td>
<td>Hb F only</td>
<td>282.49</td>
</tr>
<tr>
<td>Hb C-disease</td>
<td>Hb F, C</td>
<td>282.7</td>
</tr>
<tr>
<td>Hb C beta-thalassemia</td>
<td>Hb F,C,A</td>
<td>282.49</td>
</tr>
<tr>
<td>Hb D beta-thalassemia</td>
<td>Hb F,D,A</td>
<td>282.49</td>
</tr>
<tr>
<td>Hb E beta-thalassemia</td>
<td>Hb F,E,A</td>
<td>282.49</td>
</tr>
<tr>
<td>Hb E-disease</td>
<td>Hb F,E</td>
<td>282.7</td>
</tr>
<tr>
<td>Hb H-disease</td>
<td>Hb F,H</td>
<td>282.49</td>
</tr>
<tr>
<td>Hb SE-disease</td>
<td>Hb F,S,E</td>
<td>282.68, 282.69</td>
</tr>
<tr>
<td>Hb C-carrier</td>
<td>Hb F,A,C</td>
<td>282.7</td>
</tr>
<tr>
<td>Hb D-carrier</td>
<td>Hb F,A,D</td>
<td>282.7</td>
</tr>
<tr>
<td>Condition Name</td>
<td>Hemoglobin Screening Result</td>
<td>ICD-9 Code(s)</td>
</tr>
<tr>
<td>------------------------</td>
<td>-----------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>Hb E-carrier</td>
<td>Hb F,A,E</td>
<td>282.7</td>
</tr>
<tr>
<td>Hb S (sickle)-carrier</td>
<td>Hb F,A,S</td>
<td>282.5</td>
</tr>
</tbody>
</table>

1.K. Data Sources

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

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

State newborn screening data.

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 Specifications in the Supporting Documents for Q-METRIC Sickle Cell Disease Measure 1, Timeliness of Confirmatory Testing for Sickle Cell Disease.

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

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 components of the health care system and incur significant costs. In 2009, mean hospital charges for children with SCD who had 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, Coates, Huazhi,
et al. (2009) estimate the lifetime cost of health care per patient with SCD to be approximately $460,000.

**Outcomes of Timely Confirmatory Sickle Cell Disease Testing**

Children with SCD benefit significantly from early diagnosis of disease. In a group of patients diagnosed with SCD as newborns, the mortality rate was 1.8 percent compared with an 8 percent mortality rate among those diagnosed after 3 months of age (Vichinksy, Hurst, Earles, et al., 1988). Most of the study was carried out prior to the use of prophylactic antibiotics, which today is a standard for pediatric SCD treatment and significantly reduces the frequency of life-threatening infections. The lower mortality rate in the study is attributed to early diagnosis and treatment of complications, practices that are supported, in large part, by extensive follow-up and patient and parent education. Newborn screening for SCD followed by confirmatory testing for positive screening results, parental education, and comprehensive care reduces morbidity and mortality in infants and children with SCD (NHLBI, 2002).

This measure assesses timely confirmatory testing for a positive SCD newborn screen and the communication of those results to families. The measure does not change across developmental stages.

**Performance Gap**

There is significant variability and little tracking by more than half of States regarding this important issue. Neonatal screening for SCD is mandated in all 50 States, as well as in the District of Columbia, Puerto Rico, the U.S. Virgin Islands, and Guam. To our knowledge, there are no current published data that describe the proportion of children with a positive screen for SCD who receive confirmatory testing or what proportion of families with a child who receives a positive confirmatory test result have results communicated to them. However, in a study of 52 State Newborn Screening Program Follow-up Coordinators with 100 percent participation, 100 percent of primary care providers were notified of positive SCD screening results, while only 81 percent of hematologists, 73 percent of hospitals, and 40 percent of families were notified of positive findings (Kavanagh, Wang, Therrell, et al., 2008). In programs where communication with the families was inconsistent, the responsibility for providing this information often fell upon someone other than the State newborn screening program. The efficacy of ensuring timely confirmatory testing is unknown.

**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).
This measure is relevant to Medicaid/CHIP 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).

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.
g. Service – other (please specify): No.
h. Measure Topic – duration of enrollment: No.
i. Measure Topic – clinical quality: Yes.
k. Measure Topic – family experience with care: No.
l. Measure Topic – care in the most integrated setting: No.
m. Measure Topic other (please specify): No.

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; 29 days to 120 days.
q. Population – pre-school age children (1 year through 5 years) (specify age range): No.
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 two-part clinical process (confirmatory testing for infants with a positive newborn screening result and communication of those testing results to families) that, if followed, results in a desirable clinical outcome: reduced morbidity and mortality from SCD in infancy and early childhood when neonatal screening and confirmatory testing are linked to parental education and comprehensive care (NHLBI, 2002). This measure highlights where providers or health systems are falling short in timely confirmatory testing of all infants with a positive newborn screen for SCD and in communicating results to families. The body of evidence addresses the importance of confirmatory testing and communication of results. The evidence predominantly consists of clinical guidelines and expert consensus. Table 3 (see Supporting Documents) summarizes several key sources of evidence for this measure, using the U.S. Preventive Services Task Force (USPSTF) rankings (criteria denoted in the table).

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.

In patients with SCD, the mortality rate is highest in the first 5 years of life, with the greatest period of risk occurring between 6 months and 1 year of age (Vichinsky et al., 1988). Pneumonia
and splenic sequestration are but two of the potentially catastrophic complications that can occur in infancy; such complications have the potential to result in death if not recognized and treated promptly (Consensus Conference, 1987). Early detection of the disease through newborn screening followed by confirmatory testing and the successful communication of an SCD diagnosis to the family enables health care providers to offer preventive measures, such as antibiotics, and to teach lifesaving skills, such as how to detect an enlarged spleen or respond to a dangerous fever (Consensus Conference, 1987; Steinberg, 1999). This process leads to better outcomes for patients and their families and gives health care providers the advantage of a proactive approach in addressing serious care issues in their patients with SCD.

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.

Data/Sample

This measure is based on the gold standard data source for sickle cell disease: initial and confirmatory diagnosis information that is maintained by all State newborn screening programs in the United States. Q-METRIC tested this measure using newborn screening results from public health agencies in three States: Illinois, Michigan, and Wisconsin. Newborn screening data capture the vast majority of births in States—in Michigan, 97 percent of all children have newborn screening performed, while in Illinois the figure is nearly 99 percent. For remaining births, the parents opt out of newborn screening based on religious grounds. The measure was tested as specified, which requires an assessment among the entire population of a State’s birth cohort that had an initial newborn screen indicating SCD. This measure includes no sampling; consequently, no sampling error is introduced that would necessitate the calculation of measure reliability.

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 through face validity established by a national panel of experts and advocates for families of children with SCD. Face validity is the degree to which the measure construct characterizes the concept being assessed, which was established by combining two Q-METRIC SCD expert panels. The panel established a very high degree of face validity for this measure through a detailed review of concepts and metrics considered to be essential to effective SCD management and treatment. 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 expert panels included 14 experts providing a comprehensive perspective on SCD management and the measurement of quality metrics for States and health plans. From this group, concepts and draft measures were rated for their relative importance. This measure was among the most highly rated, with the expert panel’s average score of 8.7 (out of a maximum of 10). Two rating methods were used to minimize any potential bias due to outlier ratings; this measure received identically high ratings (8.7) using both methods. In addition, the expert panel members noted that not only was this measure important, but it could be accessed through State health departments. The measure as specified from State newborn screening program data was deemed to be the most valid, in contrast to candidate metrics that would be derived from provider data. Finally, the Q-METRIC SCD expert panel noted that this measure was a valid marker of important variations that they felt may exist across States; the panel noted that newborn screening programs can be highly variable across States, and this measure would likely reflect those variations.

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

Testing results show that SCD is largely concentrated among African American infants. Table 4 (see Supporting Documents) summarizes the distribution across race and ethnicity groups for each State. Note the degree to which information on the race and ethnicity of infants with SCD varies among States.

7.B. Special Health Care Needs

Among the States in which Q-METRIC conducted testing, newborn screening data for infants did not include indicators of special health care needs.
7.C. Socioeconomic Status

Although State newborn screening data for infants do not directly capture information on socioeconomic status, Medicaid eligibility, shown in Table 5, provides one proxy indicator using data available from two States (Illinois and Michigan).

<table>
<thead>
<tr>
<th>Insurance Status</th>
<th>Illinois (percent)</th>
<th>Michigan (percent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medicaid recipient</td>
<td>74.4</td>
<td>54.4</td>
</tr>
<tr>
<td>Non-Medicaid recipient</td>
<td>25.6</td>
<td>45.5</td>
</tr>
</tbody>
</table>

7.D. Rurality/Urbanicity

Table 6 summarizes the urban/rural distribution for our testing States and shows that the overwhelming majority of newborns with a positive screen for SCD reside in urban settings. This information was not available from Wisconsin’s newborn screening data.

<table>
<thead>
<tr>
<th>Residence</th>
<th>Illinois (percent)</th>
<th>Michigan (percent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rural</td>
<td>5.8</td>
<td>0.7</td>
</tr>
<tr>
<td>Urban</td>
<td>94.2</td>
<td>99.0</td>
</tr>
<tr>
<td>Unknown</td>
<td>n/a</td>
<td>0.3</td>
</tr>
</tbody>
</table>

7.E. Limited English Proficiency (LEP) Populations

This information is not available from newborn screening data in the three States in which this Q-METRIC measure was tested.

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 implemented using State universal newborn screening data for SCD. This testing is mandatory in all 50 States and the District of Columbia, Puerto Rico, the U.S. Virgin Islands, and Guam. Table 7 (see Supporting Documents) outlines the SCD newborn screening processes used by the three States in which this measure was tested by Q-METRIC (Illinois, Michigan, and Wisconsin). State newborn screening programs employ thin-layer isoelectric focusing (IEF, also
called hemoglobin electrophoresis) and high performance liquid chromatography (HPLC) techniques to examine capillary blood collected from a heel stick and absorbed onto filter paper. These tests identify types of hemoglobin and differentiate between SCD and sickle cell trait (Pack-Mabien, Haynes, 2009; USPSTF, 2007).

State public health departments have responsibility for newborn screening programs, including prompt and continuing follow-up to make sure infants identified through newborn screening receive early diagnosis, treatment, and management (National Newborn Screening and Genetics Resource Center [NSGRC], 2009). Some States with well-evolved programs have nurses or program specialists contact families of infants with positive SCD results for further testing and follow-up. Other States depend on the primary care provider to handle these tasks (Kavanagh, Wang, Therrell, et al., 2008).

All States conduct newborn screening for SCD; the birth cohorts for the three States in which this measure was tested by Q-METRIC are summarized in Table 8.

Table 8. Birth Cohorts by State, 2007-2011

<table>
<thead>
<tr>
<th>Annual Births</th>
<th>Illinois</th>
<th>Michigan</th>
<th>Wisconsin</th>
</tr>
</thead>
<tbody>
<tr>
<td>2007</td>
<td>180,530</td>
<td>125,172</td>
<td>72,757</td>
</tr>
<tr>
<td>2008</td>
<td>176,830</td>
<td>121,231</td>
<td>72,002</td>
</tr>
<tr>
<td>2009</td>
<td>171,163</td>
<td>117,309</td>
<td>70,824</td>
</tr>
<tr>
<td>2010</td>
<td>165,200</td>
<td>114,717</td>
<td>68,367</td>
</tr>
<tr>
<td>2011</td>
<td>161,312</td>
<td>114,159</td>
<td>67,811</td>
</tr>
</tbody>
</table>

The eligible population for this measure consists of those children who had an initial newborn screen indicating SCD. In the three States in which this measure was tested, the annual number of children with an initial screen for SCD ranged from 24 infants (Wisconsin) to 111 (Illinois). Performance on this measure varied within and between the three States tested (Table 9, see Supporting Documents). The overall average varied: 52.6 percent (Illinois), 47.4 percent (Michigan), and 80.7 percent (Wisconsin).

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 addresses two rates. The first rate — the percentage of children who, having initially tested positive for SCD through newborn screening, received confirmatory testing by 3 months of age — was determined to be feasible in the three States tested by Q-METRIC. In contrast, the second rate — the percentage of children with results of confirmatory testing communicated to families within 120 days of life — was determined to not be currently feasible using existing newborn screening data systems. However, significant initiatives are currently underway that will better support this measure through increased use of electronic health record (EHR) systems by primary care providers and specialists. Consequently, the availability of information regarding the communication of newborn screening results to parents
may substantially improve in the near future. For example, the use of personal health portals and health information exchange technologies will enable a subset of parents (current estimates are between 10 and 50 percent) to reliably receive information as it becomes available from their health care providers. In these cases, it will be important to refine the measure to include two distinct actions: “made available” and “received,” both of which are measurable using the EHR and usage logs.

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.

This measure is not currently in use in the three States in which Q-METRIC testing was conducted; further, it is not believed to be currently in use in any State.

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)

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?
All SCD cases reported in respective State.

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; this is the level at which results from newborn screening data for SCD are collected and maintained in the United States.

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)
Yes.

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?
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?
Yes.

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**

**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.

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**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, which assesses both the timeliness of confirmatory testing for SCD following a positive newborn screening and the communication of results to the family, is useful on multiple levels: First, it will give families the information they need to seek prompt early preventive treatment for SCD issues and to understand the importance of appropriate interventions for complications. Likewise, with early diagnosis, providers will be able to better address the serious medical challenges SCD raises for children, especially the very young, both in terms of prevention and in prompt attention to arising complications. For purchasers of care, early
diagnosis will result in more prompt institution of care that will both prevent and delay complications of this condition and thus likely lower long-term costs in the care of these patients.

This measure has not been assessed for comprehension. Because newborn screening is universally mandatory, State employees responsible for the procedures involved in this measure—screening, confirmatory testing, and communication of results—are familiar with the concepts of this process. In the three States where Q-METRIC tested the measure, employee comprehension was excellent.

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.

Information technology, such as mobile access to these repositories, may facilitate receipt of information important to this measure. For example, portals currently use secure messaging in a “content-agnostic” method to deliver messages to patients. It is possible that high-priority information can be delivered differently, thereby improving time to receipt. Second, as these technologies improve their interfaces to support small form-factor screens (e.g., smart phones), the time to patients’ receipt of important clinical information is likely to further decrease. Finally, health information exchange technology will allow rapid dissemination of results among caregivers, thereby improving communication of confirmatory testing from laboratories to primary care providers or specialists.

11.B. Health IT Testing

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

No.

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

This measure was tested using State newborn screening data.

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.

The key information needs of this measure are newborn screening results, confirmatory test ordering, confirmatory test results, and receipt of those results by providers and subsequently by families (parent or legal guardian). Newborn screening results currently arrive via hardcopy in many States and must be reviewed. Reviewing a laboratory result for newborn screening may be
captured by the EHR in the places where these results arrive electronically; or more typically, it may be documented in a note after review. Either of these methods (electronic capture or documentation in a note) will provide a time stamp for receipt of screening. Confirmatory testing is both documented by providers and ordered using electronic ordering systems, depending on the EHR. Results will be stored in the EHR and available for review. This review step may be audited, or results may be documented in an encounter summary.

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 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 into EHRs, which is directly related to the measurement of the timeliness and completeness of newborn screening results for children with SCD. The ONC standards include the following specific requirements in the Certification criteria (Federal Register, 2010):

(g) Incorporate laboratory test results:

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

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

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

(4) Update. Enable a user to electronically update a patient's record based upon received laboratory test results.

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. Newborn screening results for hemoglobin screening (initial and confirmatory), including the specific name of the hemoglobin condition.

3. Date of SCD screening results (initial and confirmatory).
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?

Implementation of an order entry system will allow easy access to date of ordering. This measure is based on the completion of confirmatory screening; knowing when the test was ordered will be important if the measure is low. Implementation of a reporting system for results or a secure messaging platform should greatly affect the ability to communicate results to patients or providers, thereby improving this measure.

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 two major concepts:

1. The percentage of children who test positive for SCD on newborn screening who receive confirmatory testing by 90 days of life.

2. The percentage of children with results of confirmatory testing communicated to families within 120 days of life.

Q-METRIC testing determined that the first concept is feasible and can be assessed using existing data systems. However, the second measure relies on tracking confirmatory testing results being communicated to parents, which currently is not documented in newborn screening data systems. However, emerging EHR advances may establish the feasibility of this measure using information from providers’ EHR systems.

References


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, Timeliness of Confirmatory Testing for Sickle Cell Disease, assesses two rates: (1) the percentage of children who, having initially tested positive for SCD through newborn screening, received confirmatory testing by 3 months of age; and (2) the percentage of children whose confirmatory testing results were communicated to their families by 4 months of age. Currently, there are no quality measures for the diagnosis, assessment, or treatment of pediatric SCD. Yet SCD is one of the most common genetic disorders in the United States, and its impact among children is great. In patients with SCD, the mortality rate is highest in early childhood, and infancy is a period of special risk. Young children with SCD experience infections, stroke, pain episodes, and hospitalizations; these events restrict sleep and daily activities, affecting a child’s ability to engage academically and socially.

Early diagnosis is highly effective in mitigating these challenges by promoting comprehensive care and parental education. Providers can proactively prescribe antibiotics to ward off overwhelming bacterial infections. Parents can learn lifesaving skills: how to detect an enlarged spleen or respond to a high fever. For purchasers of care, early diagnosis will result in prompt institution of care that will both prevent and delay complications, thus likely lowering health care costs over the long term. This proposed measure will support improved outcomes and appropriate use of health care resources. The measure received a high degree of support from the Q-METRIC SCD expert panel for addressing SCD management using a robust data source: State newborn screening data. Because newborn screening is mandatory in the United States, these data capture the vast majority of births.

At present, there is significant variability and little tracking by more than half of States regarding the subsequent steps of confirmatory testing and communication of results. To our knowledge, there are no current published data that describe the proportion of children with a positive screen for SCD who receive confirmatory testing, or what proportion of families are provided these positive results. Q-METRIC testing results for the first rate of the measure—the percentage of children who, having initially tested positive for SCD through newborn screening, received confirmatory testing by 3 months of age—varied from 52.6 percent (Illinois), 47.4 percent (Michigan), to 80.7 percent (Wisconsin). We regard the second rate—the percentage of children whose confirmatory testing results were provided to families—as aspirational. While this information currently is not readily available, it is our intention that the measure will promote better execution and capture of this crucial step. Health IT will be instrumental in facilitating this change, as system improvements will enhance the ease and speed with which important newborn screening information is communicated from labs to agencies and then to providers and families. Use of EHRs will likewise provide more accurate, consistent, and thorough documentation of confirmatory testing and communication activities.
The Q-METRIC expert panel noted that newborn screening programs can be highly variable across States; this measure would likely reflect those variations, an important first step in assuring consistent treatment for children nationwide. This measure is especially relevant to Medicaid /CHIP because the majority of children with SCD are also enrolled in Medicaid. These vulnerable young patients will benefit from prompt SCD diagnosis, medical attention, and family education.

Section 14: Identifying Information for the Measure Submitter

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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.

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