SECTION I.
BASIC MEASURE INFORMATION

I.A. Measure Name

Anticipatory Guidance for Prevention of Stroke in Children with Sickle Cell Disease

I.B. Measure Citation Information

Dombkowski KJ, Madden B, Shevrin CA, McCormick J, Freed GL for the Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium. Anticipatory Guidance for Prevention of Stroke in Children with Sickle Cell Disease. National Quality Measures Clearinghouse (NQMC). Rockville (MD): Agency for Healthcare Research and Quality (AHRQ). Published March 23, 2015.

I.C. Measure Description

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

This measure assesses the percentage of children younger than 18 years of age identified as having sickle cell disease (SCD) who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts 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. From infancy onward, the presence of this hemoglobin variant can lead to an array of serious medical conditions, including brain damage. Among children with SCD, approximately 11% experience a stroke by 20 years of age; approximately 13% show evidence of silent infarcts, that is, injuries sustained by the brain without clinical symptoms. Stroke can cause long-lasting complications, including cognitive deficits, mood and/or personality changes, physical weakness, and difficulties with language, vision, and swallowing. Silent infarcts are associated with learning disabilities. Assessing stroke risk in children with SCD is crucial; effective screening tools, such as transcranial Doppler (TCD) ultrasonography and magnetic resonance imaging (MRI), exist to identify those most at risk. Similarly, recognizing the symptoms of stroke and having a plan to seek immediate care are likewise essential in order to mitigate complications. Clinical guidelines indicate that families and other caregivers should receive anticipatory guidance about identifying, preventing, and managing strokes and silent infarcts. However, there are no existing quality measures for anticipatory guidance regarding stroke and silent infarct in children with SCD.

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

I.D. Measure Owner

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

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

Not applicable

I.F. Measure Hierarchy

Please use this section to note 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's National Quality Measures Clearinghouse and are available at http://www.qualitymeasures.ahrq.gov/about/hierarchy.aspx:

I.F.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.

I.F.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.

I.F.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

I.F.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

I.G. Numerator Statement

The eligible population for the numerator is the number of children younger than 18 years of age identified as having SCD who received anticipatory guidance regarding the prevention and/or management of stroke and silent infarcts 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, based on appropriate ICD-9 codes as documented in the medical record. Codes to identify outpatient visits are documented in Table 2.

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

Table 1: Codes to Identify Sickle Cell Disease

Condition Name	Hemoglobin Screening Result	ICD-9 Code(s)
Hb beta zero-thalassemia	Hb F only	282.49
Hb S beta-thalassemia	Hb F,S,A	282.41, 282.42
Hb SC-disease	Hb F,S,C	282.63, 282.64
Hb SD-disease	Hb F,S,D	282.68, 282.69
Hb SS-disease (sickle cell anemia)	Hb F,S	282.6, 282.61, 282.62

Table 2: Codes to Identify Outpatient Care

Description	CPT	HCPCS	ICD-9-CM Diagnosis
Office or other outpatient services	99201-99205, 99211-99215, 99241-99245		
Preventive medicine	99381-99385, 99391-99395, 99401-99404, 99411-99412, 99420, 99429	G0438, G0439	
General medical examination			V20.2, V70.0, V70.3, V70.5, V70.6, V70.8, V70.9

I.H. Numerator Exclusions (as appropriate)

1. Inpatient stays, emergency department visits, and urgent care visits are excluded from the calculation.
2. Children with a diagnosis in the sampled medical record indicating one of the SCD variants listed in Table 3 should not be included in the eligible population *unless* there is also a diagnosis for a sickle cell variant listed in Table 1.

Table 3: Excluded Sickle Cell Disease Diagnosis Codes

Condition Name	Hemoglobin Screening Result	ICD-9 Code(s)
Hb C beta-thalassemia	Hb F,C,A	282.49
Hb D beta-thalassemia	Hb F,D,A	282.49
Hb E beta-thalassemia	Hb F,E,A	282.49
Hb C-disease	Hb F,C	282.7
Hb E-disease	Hb F,E	282.7
Hb H-disease	Hb F,H	282.49

Condition Name	Hemoglobin Screening Result	ICD-9 Code(s)
Hb SE-disease	Hb F,S,E	282.68, 282.69
Hb C-carrier	Hb F,A,C	282.7
Hb D-carrier	Hb F,A,D	282.7
Hb E-carrier	Hb F,A,E	282.7
Hb S (sickle)-carrier	Hb F,A,S	282.5

I.I. Denominator Statement

The eligible population for the denominator is the number of children younger than 18 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, based on appropriate ICD-9 codes as documented in the medical record. Codes used to identify outpatient care are documented in Table 2.

I.J. Denominator Exclusions (as appropriate)

1. Inpatient stays, emergency department visits, and urgent care visits are excluded from the calculation.
2. Children with a diagnosis in the sampled medical record indicating one of the SCD variants listed in Table 3 should not be included in the eligible population *unless* there is also a diagnosis for a sickle cell variant listed in Table 1.

I.K. Data Sources

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

Data Source	
1. Administrative Data (e.g., claims data)	
2. Paper Medical Record	**X**
3. Survey – Health care professional report	
4. Survey – Parent/caregiver report	
5. Survey – Child report	
6. Electronic Medical Record	**X**
7. Other (If other, please list all other data sources in the field below.)	

SECTION II.
DETAILED MEASURE SPECIFICATIONS

Provide sufficient detail to describe how a measure would be calculated from the recommended data sources, either by uploading a separate document or by providing a link to a URL in the field below. 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.[1] 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 specifications document, *Q-METRIC Sickle Cell Disease Measure 7, Anticipatory Guidance for Prevention of Stroke in Children with Sickle Cell Disease,* at the end of this document. The SCD codebook used for medical record data abstraction is also included as a separate file.

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1 Initial Core Set of Children's Health Care Quality Measures: Technical Specifications and Resource Manual for Federal Fiscal Year 2011 Reporting. Available at http://www.medicaid.gov/Medicaid-CHIP-Program-Information/By-Topics/Quality-of-Care/Downloads/InitialCoreSetResourceManual.pdf and http://www.medicaid.gov/Medicaid-CHIP-Program-Information/By-Topics/Quality-of-Care/CHIPRA-Initial-Core-Set-of-Childrens-Health-Care-Quality-Measures.html.

SECTION III.
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).

III.A. Evidence for General Importance of the Measure

Provide evidence for all applicable aspects of general importance, including but not limited to the following:

- Addresses a known or suspected quality gap or disparity in quality (e.g., addresses a socioeconomic disparity, a racial/ethnic disparity, a disparity for Children with Special Health Care Needs (CSHCN) and/or a disparity for limited English proficiency (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 et al., 2011). The National Heart, Lung and Blood Institute 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 et al., 2010).

Sickle Cell Disease Pathology and Severity

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

Sickle Cell Disease Burden in Daily Life

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

Sickle Cell Disease Cost

In a study of health care utilization among low income children with SCD between 2004 and 2007, 27% of these children required inpatient hospitalization and 39% used emergency care during a year. Of these children, 63% averaged one well-child visit per year and 10% had at least one outpatient visit with a specialist (Raphael et al., 2009). Patients with SCD use many parts of the health care system, incurring significant costs. In 2009, mean hospital charges for children with SCD and a hospital stay were $23,000 for children with private insurance and $18,200 for children enrolled in Medicaid (AHRQ, 2012). Kauf et al. estimate the lifetime cost of health care per patient with SCD to be approximately $460,000 (Kauf et al., 2009).

Outcomes of Anticipatory Guidance Regarding Prevention of Stroke

Stroke is a devastating complication of SCD, one that may occur either without warning or as an accompaniment to other SCD complications. Approximately one child in 10 with SCD will have a stroke before adulthood, and the frequency of stroke is highest in these children before the age of 9 years (Ohene-Frempong et al., 1998, Steinberg, 1999; Pack-Mabien and Haynes, 2009). Ischemic strokes (those in which blood supply to the brain is blocked) occur more frequently in children with SCD than hemorrhagic (bleeding) strokes. The latter, while not unknown in children with SCD, occur more often in young adults. Children with SCD are also subject to silent infarcts, which are ischemic events that produce no symptoms but still damage the brain. Children with SCD also experience transient ischemic attack (TIAs), which are instances of brain dysfunction in which the symptoms resolve within 24 hours.

The brain is subject to serious damage as a result of stroke. Death from ischemic stroke is unusual, but motor and neuropsychological impairment is significant (Miller et al., 2001). Clinical manifestations seen in children who suffer strokes include subtle behavioral changes, academic difficulties, prolonged headaches, difficulties with speaking and language, weakness on one or both sides of the body, seizures, and gait disturbances (Pack-Mabien and Haynes, 2009). Silent infarcts can lead to learning disabilities, and TIAs are often precursors of full-blown stroke (NHLBI, 2002).

The first line of prevention is a program of regular (chronic) blood transfusions, which have been shown to reduce the risk of first stroke in children. However, to prevent one stroke, 100-200 children with SCD would need to be transfused (Adams, 2013). And because repeated transfusions are associated with toxicity, effort, and expense, this therapy is saved for high-risk patients (Miller et al., 2001). Primary prevention needs to hinge on a practical way to select those children who most need transfusion. TCD screening is a strong predictor of stroke risk and has been shown, in combination with chronic transfusion, to reduce the risk of first stroke by 92% (Adams, 2013, Adams et al., 1998). In the past 10 years, three major clinics have reported striking decreases in the occurrence of first stroke following TCD screening and chronic transfusion, proving the utility of TCD as a targeted intervention (Adams, 2013).

Even if they appear to be neurologically normal (that is, no evidence of clinical stroke), children with SCD may have a variety of anatomic and physiologic abnormalities involving the central nervous system that may be associated with deterioration in cognitive function. About 13% of children with SCD have silent brain lesions on MRI. These silent infarcts have an effect on learning and behavior and may increase the risk for clinical and subclinical damage to the CNS in the future (NHLBI, 2002).

For family members and other caretakers of children with SCD, then, stroke prevention is an important aspect of comprehensive care; understanding how to respond to the symptoms of stroke is likewise crucial. The provision of focused patient education and anticipatory guidance for families is an important step in averting or ameliorating potentially debilitating illnesses in these vulnerable patients. Home caregivers have a crucial role to play in the successful management of children with SCD, and it is important to emphasize the importance of this role at each parent education session. Educational materials and methods should be matched to the literacy level of the caregiver, and instructions should be provided on how to navigate the medical system. Information about lab values, physical findings, and medications should be easily accessible to the caregiver in case of an emergency (NHLBI, 2002).

This measure assesses the percentage of children younger than 18 years of age identified as having sickle cell disease (SCD) who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care during the measurement year. The measure does not change across developmental stages.

Performance Gap

Routine comprehensive care for children with SCD is essential to support their optimal health. These outpatient visits often provide the setting for health care providers to make sure parents and other primary caregivers receive anticipatory guidance about a range of important issues specific to managing this challenging condition. It is important, therefore, that all affected children receive dependable outpatient care. To characterize health care utilization in children with SCD, Raphael et al. (2009) studied administrative claims data from a managed care plan serving children with Medicaid and the State Children's Health Insurance Plan (SCHIP) for 2007-2009. The researchers found that a substantial proportion of children with SCD did not meet minimum guidelines for outpatient primary care and hematology comprehensive care. During the study period, only 63% of patients had one routine outpatient visit with a primary care provider and only 10% had a minimum of

one outpatient visit per year with a hematologist. These findings are concerning, as missed visits could mean lost opportunities for assessing stroke risk and providing anticipatory guidance and education about SCD and associated medical conditions.

As for screening for stroke risk, a 2008 study at the Texas Children's Sickle Cell Center reported that only 45% of children with SCD received annual TCD screenings and that patients with private insurance were three times more likely to complete more than 50% of ordered TCD screenings than patients with Medicaid (Raphael et al., 2008). In a retrospective cohort study of children aged 2-16 years old with SCD enrolled in Tennessee Medicaid, Eckrich et al. found that rates of TCD screening increased over time, with 2.5% receiving TCD screening in 1997 and 68.3% receiving screening in 2008. However, 31% of study participants received no TCD screening during the entire 11-year study period (1997-2008) (Eckrich et al., 2013). Interviews with 36 caregivers of children with SCD revealed that 22% of caregivers had no knowledge of TCD screening, and 42% were unaware that TCD screening should be performed yearly (Bollinger et al., 2011).

Another potential performance gap involves the manner in which anticipatory guidance is presented. It is important that providers take the time to listen to concerns voiced by the families of children with SCD so that information is presented in a way 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 between 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 et al., 2001).

III.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).[2]
- Any other specific relevance to Medicaid/CHIP (please specify).

Sickle Cell Disease and Medicaid/CHIP

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[2] The EPSDT is a comprehensive set of benefits available to children and youth under age 21 who are enrolled in Medicaid. For more information, see http://www.healthlaw.org/images/stories/epsdt/3-ESDPT08.pdf.

The Performance Gap section above detailed specific issues regarding the delivery of comprehensive care and screening for stroke risk to children on Medicaid diagnosed with SCD. More broadly, the measure has relevance because the majority of children with SCD are enrolled in Medicaid. In 2009, 67% of children with SCD discharged from the hospital were enrolled in Medicaid, compared with 25% who had private insurance (AHRQ, 2012). Medicaid enrollment often serves as a marker of poverty. The large number of children with SCD on Medicaid suggests some of these patients may be receiving suboptimum treatment because of unstable living situations, despite the provision of anticipatory guidance. These children may not be receiving stroke screenings regularly, and they may experience delays in being taken for medical care if family situations are such that work responsibilities, school commitments for siblings, or lack of transportation make seeking prompt medical attention difficult (Tanabe et al., 2013).

III.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 are currently no quality measures for the diagnosis, assessment or treatment of pediatric SCD.

References for Section III

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Adams RJ, McKie VC, Hsu L, et al. Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. *N Engl J Med* 1998; 339(1):5-11.

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Eckrich MJ, Wang WC, Yang E, et al. Adherence to transcranial Doppler screening guidelines among children with sickle cell disease. *Pediatr Blood Cancer* 2013; 60(2):270-274.

Hassell KL. Population estimates of sickle cell disease in the U.S. *Am J Prev Med* 2010; 38(4 Suppl):S512-521.

Kauf TL, Coates TD, Huazhi L, Mody-Patel N, Hartzema AG. The cost of health care for children and adults with sickle cell disease. *Am J Hematol* 2009; 84(6):323-327.

Kavanagh PL, Sprinz PG, Vinci SR, Bauchner H, Wang CJ. Management of children with sickle cell disease: a comprehensive review of the literature. *Pediatrics* 2011; 128(6):e1552-1574.

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Lemanek KL, Ranalli M, Lukens C. A randomized controlled trial of massage therapy in children with sickle cell disease. *J Pediatr Psychol* 2009; 34(10):1091-1096.

Miller ST, Macklin EA, Pegelow CH, Kinney TR, Sleeper LA, Bello JA, et al. Silent infarction as a risk factor for overt stroke in children with sickle cell anemia: a report from the Cooperative Study of Sickle Cell Disease. *J Pediatr* 2001; 139(3):385-390.

National Heart, Lung and Blood Institute. The Management of Sickle Cell Disease. In: National Institutes of Health, ed. Bethesda, MD, 2002.

Ohene-Frempong K, Weiner SJ, Sleeper LA, Miller ST, Embury S, Moohr JW, et al. Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood* 1998; 91(1):288-294.

Pack-Mabien A, Haynes Jr J. A primary care provider's guide to prevetive and acute care management of adults and children with sickle cell disease. *J Am Acad Nurse Pract* 2009; 21:25-257.

Raphael JL, Dietrich CL, Whitmire D, et al. Healthcare utilization and expenditures for low income children with sickle cell disease. *Pediatr Blood Cancer* 2009; 52(2):263-267.

Raphael JL, Shetty PB, Liu H, Mahoney DH, Mueller BU. A critical assessment of transcranial doppler screening rates in a large pediatric sickle cell center: opportunities to improve healthcare quality. *Pediatr Blood Cancer* 2008; 51(5):647-651.

Steinberg MH. Management of sickle cell disease. *N Engl J Med* 1999; 340(13):1021-1030.

Tanabe P, Dias N, Gorman L. Care of children with sickle cell disease in the emergency department: Parent and provider perspectives inform quality improvement efforts. *J Pediatri Oncol Nurs* 2013; 30(4):205-217.

Yanni E, Grosse SD, Yang Q, Olney RS. Trends in pediatric sickle cell disease-related mortality in the United States, 1983-2002. *J Pediatr* 2009; 154(4):541-545.

SECTION IV.
MEASURE CATEGORIES

CHIPRA legislation[3] 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,[4] 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.

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3 Children's Health Insurance Program Reauthorization Act of 2009. Public Law No. 111-3, 123 Stat. 8 (2009). Available at: http://frwebgate.access.gpo.gov/cgi-bin/getdoc.cgi?dbname=111_cong_public_laws&docid=f:publ003.111.

4 Under Section 214 of CHIPRA, States may elect to cover the following groups under Medicaid only or under both Medicaid and CHIP: pregnant women and children up to age 19 for CHIP or up to age 21 for Medicaid.

	Does the measure address this category [Yes/No drop-down]	
a. Care Setting – ambulatory	**Yes**	
b. Care Setting – inpatient	**No**	
c. Care Setting – other—please specify	**No**	**[Add the following choices: home, school, other community and public health settings, long-term care, other---drop-down or radio buttons]**
d. Service – preventive health	**Yes**	
e. Service – care for acute conditions	**No**	
f. Service - care for children with special health care needs/chronic conditions	**Yes**	
g. Service – health promotion and services to promote healthy birth	**No**	
h. Service-other (please specify)	**No**	
i. Measure Topic -duration of enrollment	**No**	
j. Measure Topic – clinical quality	**Yes**	
k. Measure Topic – patient safety	**No**	
l. Measure Topic – family experience with care	**No**	
m. Measure Topic – care in the most integrated setting	**No**	
n. Measure Topic – other (please specify)		n/a
o. Population – pregnant women		n/a
p. Population – neonates (28 days after birth) (specify age range) [o on new form]	**Yes**	Birth – 28 days
q. Population – infants (29 days to 1 year) (specify age range)[p on new form]	**Yes**	All ages in this range
r. Population – pre-school age children (1 year through 5 years) (specify age range)	**Yes**	All ages in this range
s. Population – school-age children (6 years through 10 years) (specify age range)	**Yes**	All ages in this range
t. Population – adolescents (11 years through 20 years) (specify age range)	**Yes**	Adolescents 11 through17 years

SECTION V.
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.

V.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 stroke in children with SCD), that, if followed, results in a desirable clinical outcome (decreased risk of stroke and prompt, appropriate care for emergent and/or existing cerebrovascular issues in children with SCD). The measure highlights where providers or health systems are falling short in providing health care maintenance, including anticipatory guidance for children with SCD.

The body of evidence addresses the prevention of first stroke in children with SCD, using TCD screening to identify those at highest risk followed by blood transfusions to reduce the concentration of abnormal hemoglobin (hemoglobin S) to 30% of total hemoglobin. Evidence also covers treatment for emergent ischemic and hemorrhagic stroke in this patient population and management of children with SCD who experience a silent brain infarct or TIA. Table 4 summarizes several key sources of evidence for this measure, using the US Preventive Services Task Force (USPSTF) rankings (criteria denoted in Table 4).

Table 4: Evidence Supporting Anticipatory Guidance of the Identification, Prevention and/or Management of Strokes and Silent Infarcts in Children with Sickle Cell Disease

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATION(S)
Clinical guidelines	• Prevention of a first stroke (primary prevention): Children with sickle cell disease (SCD) ages 2 to 16 years old should be screened for stroke risk using transcranial Doppler ultrasonography (TCD). Chronic transfusion should be strongly considered in those with confirmed abnormal TCD. If TCD is unavailable or technically inadequate, or if TCD results do not meet criteria for treatment in the presence of other strong indications of high risk, consideration should be given to intervention on an individualized basis unless enrollment in appropriate treatment trials is an option. (p. 93) • Children with ischemic stroke should undergoing acute evaluation with computed tomography (CT) scanning followed by intravenous hydration and exchange transfusion to reduce Hb S to less than 30% total hemoglobin. In most cases, this should be followed by chronic transfusion. (p. 93) • Children with intracranial hemorrhage should be evaluated for a surgically correctable lesion. Following this, chronic transfusion is recommended in cases of severe vasculopathy or unrepaired aneurysm. Acute hydration and short-term exchange transfusion may be beneficial as well. (p. 93) • Prevention of recurrent stroke (secondary prevention) involves chronic blood transfusion, with the target of reducing Hb S to less than 30% of total hemoglobin. The reduction in recurrent strokes is significant, but patients may still have a stroke despite adequate transfusion and low Hb S levels. (p. 88) • Current recommendations are that transfusion should be continued for at least 5 years or at least until the child reaches the age of 18. Chronic transfusion induces iron overload, which must be managed along with the transfusions. (p. 88) • For a child in whom a transient ischemia attack (TIA) is observed or strongly	III	National Heart Lung and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health. Bethesda, MD, 2002.

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATION(S)
	suspected, the presence of significant large vessel disease on imaging indicates the need for transfusion. Any other indication of significant risk to the brain should be followed by prophylactic treatment with transfusion. If a child's brain blood supply has failed once, even transiently, the patient is at significant risk of further deterioration. (p. 86) • The presence of silent brain lesions, assessed by magnetic resonance imaging (MRI), is associated with increased risk of clinical stroke. These lesions are evidence of brain injury and should prompt evaluation of the child for learning and cognitive problems; the cerebral vessels should be evaluated for primary stroke prevention. (p. 89) • While hemiparesis typically improves, cognitive deficits are often significant and long-lasting; formal testing should be carried out to identify rehabilitation and educational needs. (p. 85)		
Clinical guidelines	Any acute neurological symptoms other than mild headache should be evaluated immediately. Symptoms include weakness, impaired ability to communicate, seizures, loss of motor function, severe headache, stupor, and coma. • Initial evaluation should include a complete blood count, reticulocyte count, noncontrast CT or MRI to exclude hemorrhage. • Treatment includes anticonvulsants, if necessary; other supportive care for seizure or increased intracranial pressure, if present; and a program of chronic transfusions, usually initiated acutely by partial exchange transfusion or erythrocytapheresis. • Ischemic central nervous system injury can also present with nonfocal or soft signs, such as developmental delays or poor school performance. • Children at highest risk of stroke can be identified by screening with TCD ultrasonography. Those with positive findings may be candidates for primary stroke prevention and chronic transfusion.	III	American Academy of Pediatrics Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. Pediatrics. Mar 2002;109(3):526-535.

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATION(S)
	(all information, p.530)		
Randomized controlled trial	In the Stroke Prevention Trial in Sickle Cell Anemia (STOP), Adams et al. (1998) randomized 130 children with SCD and high stroke risk to receive either standard care or transfusions. The trial was ended early when the authors found that the risk of first stroke decreased by 92% when abnormal TCD screening results were followed by blood transfusions.	I	Adams RJ, McKie VC, Hsu L, et al. Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. N Engl J Med 1998; 339(1):5-11.
Randomized controlled trial	Adams et al. (2005) randomized 79 children with SCD and high stroke risk who had received transfusions for ≥ 30 months to receive either continued transfusions or no continued transfusions. The trial was ended before the planned enrollment of 100 children because discontinuation of transfusion was found to result in high rates of reversion to abnormal TCD results and subsequent stroke.	I	Adams RJ, Brambilla D. Discontinuing prophylactic transfusions used to prevent stroke in sickle cell disease. N Engl J Med 2005; 353(26):2769-2778.
Secondary analysis	Abboud et al. (2004) looked back at the STOP study data to determine the utility of magnetic resonance angiography (MRA) to predict stroke risk in children with SCD. The results suggested that TCD often detects flow abnormalities before stenotic lesions on MRA become evident.	II	Abboud MR, Cure J, Granger S, et al. Magnetic resonance angiography in children with sickle cell disease and abnormal transcranial Doppler ultrasonography findings enrolled in the STOP study. Blood 2004; 103(7):2822-2826.
Prospective cohort study	Siebert et al. (1998) followed 117 children with SCD over a period of 8 years. The authors found nine TCD factors that were significant for clinical disease. They recommended that TCD be used as initial screening for cerebrovascular disease in children with SCD.	II	Seibert JJ, Glasier CM, Kirby RS, et al. Transcranial Doppler, MRA, and MRI as a screening examination for cerebrovascular disease in patients with sickle cell anemia: an 8-year study. Pediatr Radiol1998; 28(3):138-142.
Clinical guidelines	• Children with conditional TCD screening results between 170 and 200 cm/second should get a second screen within 3 months. • Children with abnormal TCD results greater than 200 cm/second should have a confirming TCD and start transfusion therapy	III	Wang CJ et al. Quality-of- care indicators for children with sickle cell disease. *Pediatrics* 2011; 128:484-493.

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATION(S)
	within 1 month. • Children who experience a stroke, silent infarct, or TCD results of 170 cm/seconds or more should have an age-appropriate neurocognitive evaluation by a psychologist. • Children with SCD presenting with first time clinical stroke should be transfused.		
Clinical guidelines	Reducing the frequency of transfusion and permitting the Hb S concentration to rise to 50% of the total hemoglobin concentration after 4 years of intensive transfusion appear to be reasonable.	III	Steinberg MH. Management of sickle cell disease. *N Engl J Med* 1999; 340(13):1021-1030.
Clinical guidelines	Silent infarcts are strongly associated with increased risk of stroke in children with SCD SS (sickle cell anemia). A normal finding on MRI is reassuring. Performing MRI at an early age and in combination with TCD should improve the usefulness of silent infarct as a stroke predictor.	III	Miller ST, Macklin EA, Pegelow CH, Kinney TR, Sleeper LA, Bello JA, et al. Silent infarction as a risk factor for overt stroke in children with sickle cell anemia: a report from the Cooperative Study of Sickle Cell Disease. *J Pediatr* 2001; 139(3):385-390.
Clinical guidelines	Caretakers of young children with SCD should be educated about signs of TIA and advised to report them.	III	Ohene-Frempong K, Weiner SJ, Sleeper LA, Miller ST, Embury S, Moohr JW, et al. Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood* 1998; 91(1):288-294.
Clinical guidelines	Primary care providers should consult with a hematologist or sickle cell specialist and neurologist at a comprehensive sickle cell center for optimum management of patients who are at risk for CVA as documented by an abnormal TCD measuring greater than 170 cm/second or a previous history of stroke.	III	Pack-Mabien A, Haynes Jr J. A primary care provider's guide to preventive and acute care management of adults and children with sickle cell disease. *J Am Acad Nurse Pract* 2009; 21:25-257.

Note: USPSTF criteria for assessing evidence at the individual study level are as follows: I) Properly powered and conducted randomized controlled trial (RCT); well-conducted systematic review or meta-analysis of homogeneous RCTs. II) Well-designed cohort or case-control analytic study. III) Opinions of respected authorities, based on clinical experience; descriptive studies or case reports; reports of expert committees.

V.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.

Stroke

The abnormal hemoglobin found in sickle cells (hemoglobin S) causes red blood cells to develop a crescent (sickle) shape. Because these cells are stiff, sticky, and misshapen, they can block blood flow, which leads to many complications, including stroke. Stroke in this patient population is often caused by blockages of the intracranial internal carotid and middle cerebral arteries. A blood transfusion to reduce the concentration of hemoglobin S to 30% of total hemoglobin is a reliable method of preventing stroke. The increased volume of healthy red blood cells reduces the percentage of sickle cells in the blood that cause the arterial blockages.

It is only partly understood how SCD leads to cerebrovascular disease; the mechanism that plays the most prominent role is an intracranial cerebral artery vasculopathy (blood vessel disorder) that causes the vessels to narrow or become completely blocked. This unusual vascular disease process was confirmed 90 years ago as a cause of large brain infarctions in patients, usually children with SCD (Adams, 2013). It is further thought that in this patient population the disturbances associated with the increased cerebral blood flow and flow velocity seen in chronic anemia may cause cerebrovascular damage. Risk factors for stroke include a history of TIA, elevated systolic blood pressure, elevated steady-state leukocyte count, severe anemia, and prior history of acute chest syndrome (Ohene-Frempong et al., 1998).

Stroke is a leading cause of death in children with SCD, occurring most often in those with sickle cell anemia (SCD-SS). Work done by the Cooperative Study of Sickle Cell Disease has shown that ischemic stroke occurs in approximately 11% of patients with SCD-SS by the time they are 20 years old, with a recurrence rate of 14%. Hemorrhagic stroke occurs more often in young adults. A protective effect may operate early in life; children with SCD-SS younger than 2 years of age had the lowest incidence of stroke. However, incidence was higher in the 1-to-9 year age group compared with those ages 10 to 19 years old, suggesting that a subset of patients may have additional risk factors for early stroke (Ohene-Frempong et al., 1998). In children with SCD-SS, peak incidence of stroke occurs around age 7 (Wang et al., 1998).

When the oxygen supply to the brain falls below a critical level based on need, brain dysfunction occurs. Symptoms of this restriction in blood supply to the tissues (brain ischemia) include sudden numbness or weakness in the face, arm, or leg, especially on one side of the body; confusion, trouble speaking, or difficulty understanding speech; trouble seeing in one or both eyes; trouble walking, dizziness, loss of balance, or lack of coordination; and severe headache with no known cause (CDC, 2014). There is evidence that oxygen demands are higher in children that in adults, making the child with SCD who has significant anemia at particular risk (NHLBI, 2002).

While not perfect, chronic transfusion is a clearly important therapy. Among children with SCD SS and a first stroke who are not receiving chronic transfusion therapy, 50% have another stroke in 3

years compared with 10% of those who receive regular transfusions (Steinberg 1999). Work by the CSSCD noted no mortality after 62 ischemic strokes, but a 24% mortality rate after 38 hemorrhagic strokes within 2 weeks of the event (Ohene-Frempong et al., 1998).

Silent infarct

Silent infarcts are defined as areas of ischemic change in the central nervous system that are visible on MRI in patients with no history of neurological symptoms consistent with stroke. Research shows that very young children with SCD-SS and no history of stroke have evidence of infarction in the brain and/or stenosis of the major cerebral arteries similar to findings in older children; 17% of children with SCD SS ages 6 to 12 years old had silent infarcts (Wang et al., 1998).

MRI has demonstrated a strong association between silent infarcts and overt stroke; occurrence is 14-fold higher in those with silent infarct compared with those with a normal MRI (and thus a low risk of stroke). While 90%of CSSCD patient with silent infarct did not have a stroke during 5 years of follow-up, silent infarct is strongly associated with risk of stroke in children with SCD-SS and should receive appropriate treatment (Miller et al., 2001).

Transient ischemia attacks

A history of TIA is a strong risk factor for stroke; clinicians should regard TIA as a sign of cerebrovascular disease and use definitive diagnostic studies, initiating aggressive management to prevent occurrence of a full stroke. TIAs don't always precede a stroke, however, and mild TIAs in children may well go unnoticed (Ohene-Frempong et al., 1998). Further complicating the diagnosis of TIAs in very young children is that painful episodes can mimic the physical weakness caused by stroke. In cases where the history is unclear for the event actually being a TIA, caution is advised, especially if long-term transfusion is being considered.

Transcranial Doppler Ultrasonography

TCD screenings are an inexpensive technology with reproducible results that can be used to detect abnormal blood flow velocities in children; excessively high velocities are indicative of the stenosis and lesions that lead to stroke. As the velocity of blood flow increases in the intracranial internal carotid and middle cerebral arteries, so does the risk of stroke. NHLBI guidelines suggest that children with SCD ages 2 years and older be screened annually with TCD. The STOP randomized trial established the following cut points: Blood flow velocity greater or equal to 200 cm/second in one of these arteries is considered an abnormal finding; children with this reading should receive periodic blood transfusions and repeat TCD screenings to assess blood flow velocity. A blood flow velocity between 170 and 199 cm/second is considered conditional or marginal; children with those results should be rescreened every 4 months. Blood flow velocities under 170 cm/second are considered normal; those children can continue to be screened annually (Adams et al., 1998).

References for Section V

Adams RJ. Toward a stroke-free childhood in sickle cell disease: The 2013 Sherman Lecture. *Stroke* 2013; 44:2930-2934.

Adams RJ, McKie VC, Hsu L, et al. Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. *N Engl J Med* 1998; 339(1):5-11.

Centers for Disease Control and Prevention. Stroke signs and symtoms. http://www.cdc.gov/stroke/signs_symptoms.htm. Accessed April 4, 2014.

Miller ST, Macklin EA, Pegelow CH, Kinney TR, Sleeper LA, Bello JA, et al. Silent infarction as a risk factor for overt stroke in children with sickle cell anemia: a report from the Cooperative Study of Sickle Cell Disease. *J Pediatr* 2001; 139(3):385-390.

National Heart, Lung and Blood Institute. The Management of Sickle Cell Disease. In: National Institutes of Health, ed. Bethesda, MD, 2002.

Ohene-Frempong K, Weiner SJ, Sleeper LA, Miller ST, Embury S, Moohr JW, et al. Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood* 1998; 91(1):288-294.

Steinberg MH. Management of sickle cell disease. *N Engl J Med* 1999; 340(13):1021-1030.

Wang WC, Langston JW, Steen RG, Wynn LW, Mulhern RK, Wilimas JA, et al. Abnormalities of the central nervous system in very young children with sickle cell anemia. *J Pediatr* 1998; 132(6):994-998.

SECTION VI.
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.

VI.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 below.

Data and Methods

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

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

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

IRR was determined by calculating both percent agreement and Kappa statistics. While abstraction was still being conducted at each site, IRR assessments were conducted for 5% 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, 22 of 435 unique patient records (5%) 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 for each site and across all sites. The overall agreement for this measure is 77% and the Kappa is 0.47.

Table 5: Agreement and Kappa Statistics for Sickle Cell Disease for Inter-Rater Reliability at Three Sites

Site	Total Records	N Agreed (%)	Kappa Statistic
Hospital #1	5	60%	0.30
Hospital #2	5	60%	0.17
Hospital #3	12	92%	0.63
All Sites	22	77%	0.47

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. When consistent differences were noted between the abstractors, clarification was provided and the abstraction tool modified, where appropriate.

For this measure, abstractors disagreed 5 of 22 times about the presence or absence of anticipatory guidance for the prevention of stroke, resulting in an agreement of 77% and a Kappa statistic of 0.47. During discussion in the review meetings, it was discovered that one of the abstractors had missed one case of anticipatory guidance, and had considered a transcranial Doppler ultrasound and an MRI visit to be anticipatory guidance about stroke. Therefore, retraining was necessary. It was reiterated that although the measure specification included "the identification, prevention and/or management of stroke/silent infarcts", there had to be anticipatory guidance provided, not just a clinical assessment. Text was added to the data collection tool to clarify the definition of anticipatory guidance.

VI.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., R^2 for concurrent validity). Provide appropriate citations to justify methods.

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

Face Validity

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

The Q-METRIC expert panel concluded that this measure has a high degree of face validity through a detailed review of concepts and metrics considered to be essential to effective SCD management and treatment. Concepts and draft measures were rated by this group for their relative importance. This measure was highly rated, receiving an average score of 7.8 (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; our findings indicate that these data have a high degree of face validity. This measure was tested among a total of 500 children younger than 18 years of age with sickle cell disease (Table 6). Overall, 20% of children with SCD received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care (range: 14%-58%).

Table 6: Anticipatory Guidance Regarding Stroke for Children with Sickle Cell Disease

Site	Rate	Numerator	Denominator
Hospital #1	15%	10	65
Hospital #2	58%	38	66
Hospital #3	14%	52	369
All Sites	20%	100	500

SECTION VII.
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.

VII.A. Race/Ethnicity

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

Table 7: Race/Ethnicity for Newborns with SCD in Michigan, 2004-2008 (n=294)

White			Black			Asian or Pacific Islander			Other	
Non-Hispanic	Hispanic	Total	Non-Hispanic	Hispanic	Total	Non-Hispanic	Hispanic	Total	Unknown	Total
2%	1%	3%	81%	1%	82%	1%	0%	1%	15%	100%

VII.B. Special Health Care Needs

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

VII.C. Socioeconomic Status

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

VII.D. Rurality/Urbanicity

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

VII.E. Limited English Proficiency (LEP) Populations

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

SECTION VIII.
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.[5] Using the following sections, explain the methods used to determine the feasibility of implementing the measure.

VIII.A. Data Availability

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

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

Medical records for 100% 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; 500 records (74%) met denominator criteria for this measure.

Based on the abstracted chart data, the rate was calculated as the percentage of children younger than 18 years of age identified as having SCD who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care (20%). Measure numerator (100) divided by denominator (500). (See Table 6 in the Validity section above).

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

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[5] The definition is adapted from: Centers for Medicare & Medicaid Services Quality Measurement and Health Assessment Group glossary, as part of the Measures Management System Measure Development Overview. Available at: http://www.cms.gov/MMS/19_MeasuresManagementSystemBlueprint.asp#TopOfPage. Accessed February 6, 2012.

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 15 minutes per eligible SCD case abstracting the data for this measure, with times ranging from 1-45 minutes.

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

The proposed measure was determined to be feasible by Q-METRIC using medical record data from the three largest centers serving SCD patients in Michigan during 2012. Although paper charts were used at one of the sites, this was not found to be a barrier. In fact, the average time spent abstracting records for paper charts (14 minutes) was only slightly more than the 13-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.

VIII.B. Lessons from Use of the Measure

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

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

VIII.B.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

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

Not applicable

SECTION IX.
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 Section XVI. 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. Table IX-1 shows the questions (in columns) about the measure's use at different levels of aggregation for quality reporting (in rows) included in the CHIPRA PQMP Candidate Measure Submission Form (CPCF).

Table IX-1. Questions about the measure's use at different levels of aggregation for quality reporting

Level of aggregation (Unit) for reporting on the quality of care for children covered by Medicaid/CHIP†	**Intended use: Is measure intended to support meaningful comparisons at this level? (Yes/No)**	**Data Sources: Are data sources available to support reporting at this level?**	**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?**	**In Use: Have measure results been reported at this level previously?**	**Reliability & Validity: Is there published evidence about the reliability and validity of the measure when reported at this level of aggregation?**	**Unintended consequences: What are the potential unintended consequences of reporting at this level of aggregation?**
State level*: Can compare States	☐Yes ☒No	No	Not applicable	No	No	Not applicable
Other geographic level: Can compare other geographic regions (e.g., MSA, HRR)	☐Yes ☒No	No	Not applicable.	No	No	Not applicable
Medicaid or CHIP Payment model: Can compare payment models (e.g., managed care, primary care case management, FFS, and other models)	☐Yes ☒No	No	Not applicable	No	No	Not applicable
Health plan*: Can compare quality of care among health plans.	☐Yes ☒No	No	Not applicable	No	No	Not applicable
Provider-level*						
Individual practitioner: Can compare individual health care professionals	☐Yes ☒No	No	Not applicable	No	No	Not applicable
Hospital: Can compare hospitals that serve SCD patients.	☒Yes ☐No	Yes.	The sample would be comprised of all children with clinical documentation of sickle cell disease [see Table 1] presenting in an outpatient setting.	No.	No.	None identified.
Practice, group, or facility:** Can compare: (i) practice sites; (ii) medical or other professional groups; or (iii) integrated or other delivery networks	☐Yes ☒No	No	Not applicable	No	No	Not applicable

† There could be other levels of reporting that could be of interest to Medicaid agencies such as markets and referral regions.

* Required in CHIPRA legislation. ** There is no implication that measures that are applicable at one level are automatically applicable at all three of the levels listed in this row.

SECTION X.
UNDERSTANDABILITY

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

This measure provides families with a straightforward measure to assess how well basic levels of comprehensive care are being provided for children with SCD. Low rates for 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, 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 XI.
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 calculation of the measure.

XI.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 to 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.

XI.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 using the Epic electronic health records system. The third facility used paper medical records for outpatient visits.

XI.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, or 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.

XI.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: http://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. 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 (Federal Register 2010) pertaining to Stage 2 Meaningful Use requirements:

> (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.

XI.E. Health IT Calculation

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

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

1. Child's date of birth
2. ICD-9 codes selected to indicate sickle cell anemia/SCD
3. Date and time of anticipatory guidance
4. Care setting

XI.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 on the measure?

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

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 emergency departments, 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).

References for Section XI

Health information technology: Initial set of standards, implementation specifications, and certification criteria for electronic health record technology." Fed Regist **75**(8): 2013-2047.

Electronic Health Record Incentive Program—Stage 2 Fed Regist **77**(171): 53968- 54162.

SECTION XII.
LIMITATIONS OF THE MEASURE

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

This measure assesses the percentage of children younger than 18 years of age identified as having sickle cell disease (SCD) who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care during the measurement year. A higher proportion indicates better performance as reflected by appropriate guidance.

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.

In future implementations, there are considerations that may further strengthen this measure and potentially ease the burden of data collection. Specific feedback from our medical record abstractors suggested that it would be helpful to clarify in the measure specification whether information about the *identification* of stroke also qualifies as anticipatory guidance for prevention of stroke. Although our testing results for this measure do not include this change, it should be considered prior to subsequent implementation of this measure.

SECTION XIII.
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 of Stroke in Children with Sickle Cell Disease*, assesses the percentage of children younger than 18 years of age identified as having sickle cell disease (SCD) who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts 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 stroke/silent infarcts in children with SCD.

Stroke is a devastating complication of SCD; one child in 10 with SCD will have a stroke before adulthood. Clinical guidelines suggest that to prevent a first stroke, children with SCD ages 2 to 16 years old should be screened for stroke risk using transcranial Doppler ultrasonography (TCD). Chronic transfusion should be strongly considered in those with confirmed abnormal TCD. Prevention of recurrent stroke (secondary prevention) also involves chronic blood transfusion, with the target of reducing Hb S to less than 30% of total hemoglobin. The presence of silent brain lesions, assessed by magnetic resonance imaging (MRI), is associated with increased risk of clinical stroke. These lesions are evidence of brain injury and should prompt evaluation of the child for learning and cognitive problems, as well as for primary stroke prevention. However, studies have shown that many children are never screened for stroke and that many do not receive adequate comprehensive outpatient care, the setting in which anticipatory guidance is provided.

Q-METRIC tested this measure among a total of 500 children younger than 18 years of age with SCD. Overall, 20% of children with SCD received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care (range: 14%-58%).

This measure provides families, providers, and purchasers with a straightforward means of assessing how well basic levels of comprehensive care are being provided for children with SCD, 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.

SECTION XIV.
IDENTIFYING INFORMATION FOR THE MEASURE SUBMITTER

Complete information about the person submitting the material, including the following:

a. Gary L. Freed, MD, MPH

b. Percy and Mary Murphy Professor of Pediatrics, School of Medicine; Professor of Health Management and Policy, School of Public Health

c. University of Michigan

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g. Signed written statement guaranteeing that all aspects of the measure will be publicly available, as defined in the Public Disclosure Requirements.

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 (Section XIV: Identifying Information for the Measure Submitter).

This work was funded by the Agency for Healthcare Research and Quality (AHRQ) and the Centers for Medicare & Medicaid Services (CMS) under the CHIPRA Pediatric Quality Measures Program Centers of Excellence grant number U18 HS020516. AHRQ, in accordance to CHIPRA 42 U.S.C. Section 1139A(b), and consistent with AHRQ's mandate to disseminate research results, 42 U.S.C. Section 299c-3, has a worldwide irrevocable license to use and permit others to use products and materials from the grant for government purposes, which may include making the materials available for verification or replication by other researchers and making them available to the health care community and the public, if such distribution would significantly increase access to a product and thereby produce substantial or valuable public health benefits.

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Sickle Cell Disease

Measure 7: Anticipatory Guidance for Prevention of Stroke in Children with Sickle Cell Disease

Description

The percentage of children identified as having Sickle Cell Disease who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care during the measurement year. A higher proportion indicates better performance as reflected by appropriate guidance.

Calculation

This measure requires medical record data and is calculated as follows:

The percentage of eligible children who received anticipatory guidance regarding the identification, prevention and/or management of stroke/silent infarcts (numerator divided by denominator).

Definitions

Intake period January 1 through December 31 of the measurement year.

Anticipatory guidance Any written or face-to-face verbal communication regarding the identification, prevention and/or management of stroke/silent infarcts as part of outpatient care with patient, parent, or family member.

Outpatient care A Health Maintenance Exam (HME) or an Evaluation and Management (E&M) visit with primary care provider or a specialist (see Table 7-A).

Table 7-A: Codes to Identify Ambulatory or Preventive Care Visits

Description	CPT	HCPCS	ICD-9-CM Diagnosis
Office or other outpatient services	99201-99205, 99211-99215, 99241-99245		
Preventive medicine	99381-99385, 99391-99395, 99401-99404, 99411-99412, 99420, 99429	G0438, G0439	
General medical examination			V20.2, V70.0, V70.3, V70.5, V70.6, V70.8, V70.9

Eligible Population

The determination of eligible population for this measure requires medical record data.

Ages Younger than eighteen years of age during measurement year

Event/Diagnosis Diagnosed with sickle cell disease as documented in the medical record (see Table 7-B).

NOTE: See exclusions noted below; there are several sickle cell variants that may be recorded under the 282.49 ICD-9 code that do not qualify for inclusion (see Table 7-C). Medical records for cases with ICD-9 code 282.49 should not be reviewed unless a diagnosis of Hb beta zero-thalassemia can be confirmed.

Table 7-B: Codes to Identify Sickle Cell Disease

Condition Name	Hemoglobin Screening Result	ICD-9 Code(s)
Hb beta zero-thalassemia	Hb F only	282.49
Hb S beta-thalassemia	Hb F,S,A	282.41, 282.42
Hb SC-disease	Hb F,S,C	282.63, 282.64
Hb SD-disease	Hb F,S,D	282.68, 282.69
Hb SS-disease (sickle cell anemia)	Hb F,S	282.6, 282.61, 282.62

Specification

Denominator The eligible population for the denominator is the number of children younger than 18 years of age identified as having SCD who received outpatient care during the measurement year.

Numerator The eligible population for the numerator is the number of children younger than 18 years of age identified as having SCD who received anticipatory guidance regarding the prevention and/or management of stroke and silent infarcts as part of outpatient care during the measurement year.

Documentation in medical record must include, at a minimum, a note containing the date on which verbal or written anticipatory guidance was provided.

Exclusions

- Inpatient stays, emergency department visits, urgent care visits.
- Children with diagnosis in the sampled medical record indicating one of the sickle cell disease variants listed in Table 7-C should not be included the eligible population *unless* there is also a diagnosis for a sickle cell variant listed in Table 7-B.

Table 7-C: Excluded Sickle Cell Disease Diagnosis Codes

Condition Name	Hemoglobin Screening Result	ICD-9 Code(s)
Hb C beta-thalassemia	Hb F,C,A	282.49
Hb D beta-thalassemia	Hb F,D,A	282.49
Hb E beta-thalassemia	Hb F,E,A	282.49
Hb C-disease	Hb F,C	282.7
Hb E-disease	Hb F,E	282.7
Hb H-disease	Hb F,H	282.49
Hb SE-disease	Hb F,S,E	282.68, 282.69
Hb C-carrier	Hb F,A,C	282.7
Hb D-carrier	Hb F,A,D	282.7
Hb E-carrier	Hb F,A,E	282.7
Hb S (sickle)-carrier	Hb F,A,S	282.5