



Measure Fact Sheet – The AHRQ-CMS Pediatric Quality Measures Program (PQMP)

## Timeliness of Confirmatory Testing for Sickle Cell Disease

## Timeliness of Antibiotic Prophylaxis for Children with Sickle Cell Anemia

**Measure Developer: Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC), University of Michigan**

Numerator	Denominator	Exclusions	Data Source(s)
<b>Timeliness of Confirmatory Testing for Sickle Cell Disease</b>			
The number of children who, having initially tested positive for sickle cell disease (SCD) through newborn screening, received confirmatory testing within 90 days of birth.	The number of children who initially tested positive for SCD reported in a State's newborn screening program within the measurement year.	Children who died within 120 days of birth; or children with a diagnosis in the State newborn screening data indicating an excluded SCD variant.	State newborn screening data.
<b>Timeliness of Antibiotic Prophylaxis for Children with Sickle Cell Anemia</b>			
The number of children with a newborn screen positive for sickle cell anemia who received appropriate preventive antibiotic prophylaxis within 90 days of birth.	All sickle cell anemia cases reported in a State's newborn screening program within the measurement year.	Children who died within 90 days of birth; children who were placed in the neonatal intensive care unit within 90 days of birth.	State newborn screening data.



## Measure Importance

Sickle cell disease (SCD) is one of the most common genetic disorders in the United States.<sup>1</sup> The National Heart, Lung, and Blood Institute (NHLBI) estimates that 2,000 infants are born with SCD in the United States each year.<sup>2</sup> SCD affects 70,000-100,000 children and adults in the United States, predominantly those of African and Hispanic descent.<sup>3</sup> The condition is chronic, lifelong, and associated with a decreased lifespan.

Children with SCD benefit significantly from early diagnosis,<sup>4</sup> which is facilitated by timely confirmatory testing. Communicating results to families provides the foundation of a lifetime of appropriate care for this complex disease.

There are multiple subtypes of SCD; the subtype conferring the most clinical risk is sickle cell anemia. Prompt initiation and consistent use of antibiotics in young children with sickle cell anemia increases survival rates through the prevention of overwhelming bacterial infections.<sup>2,5</sup>

## Evidence Base for Focus of the Measures

Evidence indicates that confirmatory testing and successful communication with families enable health care providers to offer preventive measures and teach life-saving skills to those who care for children with SCD.<sup>6</sup> Antibiotic prophylaxis, initiated as early as possible in infants with sickle cell anemia and continued daily until the child is 5 years of age, reduces susceptibility to serious infection. NHLBI recommends that infants with sickle cell anemia be started on twice-daily penicillin as early as possible and remain on preventive antibiotics until age 5.<sup>2,5,7</sup>

## Advantages of the Measures

- These measures fill a gap in pediatric quality measurement.<sup>8</sup> There are no existing quality measures for diagnosis, assessment, or treatment of pediatric SCD or sickle cell anemia.
- Measuring confirmatory testing and receipt of antibiotic prescriptions within the first 90 days of life are both feasible using existing data systems.
- State newborn screening data have greater validity than provider-generated screening data and increase the sensitivity and specificity of these measures.
- These measures are publicly available for noncommercial use.

## Levels of Aggregation Applicable to the Measures<sup>9</sup>

These measures are intended for aggregation and comparison of performance at the State level.

## Reliability and Validity of the Measures

- These measures are based on the gold standard data source for SCD: initial and confirmatory diagnosis information that is maintained by all State newborn screening programs in the United States.
- These measures were tested using the entire birth cohort for three States over a 5-year period among those who had an initial newborn screen indicating SCD. These measures did not include sampling, and therefore, no sampling error was introduced that would require the calculation of measure reliability.

- The face validity of these measures was ranked highly by nationally recognized technical experts (8.6/8.7 out of 9 in each of two rating methods).
- Testing found 58 percent agreement between sickle cell anemia cases reported in newborn screening systems and Medicaid claims systems in one State in 2010; this increased substantially to 80 percent agreement in 2011.

## Measure Development and Testing

These measures of timeliness used newborn screening results from the public health agencies of three different States (Illinois, Michigan, and Wisconsin) to test validity and feasibility.

## Selected Results from Tests of the Measures

- The percentage of newborns who received confirmatory testing within 90 days of birth ranged from 47.4 to 80.7 percent in the three States in which this measure was tested.
- While data were insufficient to test how often confirmatory testing results are communicated to parents, this aspect of newborn screening is essential to the successful care of infants with SCD. Family communication of testing results is an aspirational measure that is intended to shape and improve practice.
- Data regarding the timeliness of antibiotic prophylaxis were available in only two of three States.
- The annual rates in the two States ranged between 36.4 percent and 61.0 percent over the period 2007 through 2011.

## Caveats

- Some States may not currently track antibiotic prescriptions in conjunction with their newborn screening data systems. In those jurisdictions, this measure will be more reliant upon advances in health information technology (IT). Such advances will enable wider availability of electronic prescription results through increased use of electronic health records (EHRs) and information sharing through health information exchange (HIE) technologies.
- These two measures would have to be specified for use in EHRs and HIEs.

## More Information:

- AHRQ: [CHIPRAqualitymeasures@ahrq.hhs.gov](mailto:CHIPRAqualitymeasures@ahrq.hhs.gov)
- Q-METRIC: <http://chear.org/qmetric>  
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- Coming soon: Link to measure details on the AHRQ Web site.

For more information about the PQMP, visit [www.ahrq.gov/CHIPRA](http://www.ahrq.gov/CHIPRA).

## Notes

<sup>1</sup>Kavanagh PL, Sprinz PG, Vinci SR, et al. Management of children with sickle cell disease: a comprehensive review of the literature. *Pediatrics* 2011; 128(6):e1552-74.

<sup>2</sup>National Heart, Lung and Blood Institute. The management of sickle cell disease. In: National Institutes of Health, ed. Bethesda, MD, 2002; No. 02-2117.

<sup>3</sup>Hassell KL. Population estimates of sickle cell disease in the U.S. *Am J Prev Med* 2010; 38(4 Suppl):S512-21.

<sup>4</sup>Vichinsky, E., Hurst, D., Earles, A., et al. Newborn screening for sickle cell disease: effect on mortality. *Pediatrics* 1988; 81(6):749-55.

<sup>5</sup>Gaston MH, Verter JL, Woods G, et al. Prophylaxis with oral penicillin in children with sickle cell anemia. A randomized trial. *N Engl J Med* 1986; 314(25):1593-9.

<sup>6</sup>Steinberg MH. Management of sickle cell disease. *N Engl J Med* 1999; 340(13):1021-30.

<sup>7</sup>Hirst C, Owusu-Ofori S. Prophylactic antibiotics for preventing pneumococcal infection in children with sickle cell disease. *Cochrane Database Syst Rev* 2010; 1-21.

<sup>8</sup>Dougherty D, Schiff J, Mangione-Smith R. The Children's Health Insurance Program Reauthorization Act quality measures initiatives: moving forward to improve measurement, care, and child and adolescent outcomes. *Acad Pediatr* 2011; 11(3Suppl):S1-S10.

<sup>9</sup>The Children's Health Insurance Program Reauthorization Act required measures developed under this program to "permit comparison of quality and data at a State, plan, and provider level." The measure developer identified the intended levels of aggregation and comparison as reported here.

The Children's Health Insurance Program Reauthorization Act (CHIPRA) called for establishment of a Pediatric Quality Measures Program (PQMP) as a followup to identifying the initial core set of children's health care quality measures. This measure fact sheet was produced by the Agency for Healthcare Research and Quality, based on information provided by the AHRQ-CMS CHIPRA Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC), University of Michigan, which was funded by an AHRQ-CMS award. A listing of all submitted CHIPRA Centers of Excellence measures can be found at [www.ahrq.gov/chipra](http://www.ahrq.gov/chipra). All CHIPRA COE-developed measures are publicly available for noncommercial use.



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