

Q-METRIC Sickle Cell Disease Measure 1: *Timeliness of Confirmatory Testing for Sickle Cell Disease*

Graphics for Section VIII Feasibility

VIII.A. Data Availability

TABLE 7: Summary of SCD Screening Process for Illinois, Michigan, and Wisconsin

Process Step	Illinois	Michigan	Wisconsin
Initial screening	<p>HPLC testing is done to determine the presence of abnormal Hgb in whole blood.</p> <p>Lab results are phoned to program staff immediately. The physician or hospital of record is called to recommend immediate referral. Specimen report, disorder fact sheet and medical specialist list are faxed and mailed to the physician or hospital, along with a recommendation that specialist and family be contacted immediately. Regular phone follow-up continues until case is confirmed.</p>	<p>HPLC testing is followed by IEF for all specimens with abnormal results on the HPLC run.</p>	<p>IEF and extended gradient HPLC are used.</p> <p>Abnormal results are phoned to the pediatrician, and a report is issued.</p>
Confirmatory testing	<p>All abnormal SCD screening results require confirmatory blood tests, sometimes including testing of parents and siblings.</p>	<p>All children with abnormal results on IEF are referred to SCDA-MI, which is responsible for ensuring all children with positive initial screening results receive confirmatory testing, counseling, and appropriate treatment. The confirmatory test (Hgb electrophoresis) is required preferably before 3 months of age.</p>	<p>A blood collection kit is sent to the pediatrician at approximately 3 weeks of age for collection of a whole blood specimen at 4 weeks of age. If the specimen is not returned by 2 months of age, the pediatrician is queried about the delay. The specimen is logged and linked to the initial specimen. Confirmatory testing uses IEF and extended gradient HPLC.</p>

Process Step	Illinois	Michigan	Wisconsin
Communications	<p>Referral to a pediatric hematologist for evaluation and diagnostic testing is recommended within the first month of life and should not be delayed.</p> <p>The Illinois NBS program does not track information on the timeliness of communicating NBS confirmatory testing to parents.</p>	<p>SCDAA will attempt to contact the family, inform them of the diagnosis, assist in obtaining the required confirmatory test, provide education and assist in initiating penicillin prophylaxis, when indicated.</p> <p>The Michigan NBS program does not track information on the timeliness of communicating NBS confirmatory testing to parents.</p>	<p>Results are phoned to the pediatrician, and the report is mailed to the pediatrician and the specialist in the area.</p> <p>The Wisconsin NBS program does not track information on the timeliness of communicating NBS confirmatory testing to parents.</p>
Comments	N/A	<p>The SCDAA-MI reports specific data elements back to the state NBS program for all children with initial screening results indicative of disease, creating a Michigan NBS roster for SCD. This roster includes individual-level data such as birth date, confirmatory results, date of confirmatory testing, and initiation of penicillin prophylaxis.</p>	<p>All cases of SCD are kept in a spreadsheet, including demographics, specimen information, and initial and confirmatory test results. Date infant is seen at the sickle cell clinic is added; a prescription for penicillin is provided at first visit if the medication has not already been started.</p>

Abbreviations: SCD = sickle cell disease; HPLC = high performance liquid chromatography; Hgb = hemoglobin; IEF = iso-electric focusing; SCDAA-MI = Sickle Cell Disease Association of America-Michigan Chapter; NBS = newborn screening.

Table 8: Birth Cohorts by State, 2007-2011

Annual births	Illinois	Michigan	Wisconsin
2007	180,530	125,172	72,757
2008	176,830	121,231	72,002
2009	171,163	117,309	70,824
2010	165,200	114,717	68,367
2011	161,312	114,159	67,811

Table 9: Percentage of Children Testing Positive for SCD on Newborn Screening who Received Confirmatory Testing by 90 Days of Life

State	2007		2008		2009		2010		2011	
	n	%	n	%	n	%	n	%	n	%
Illinois	108	27.8%	111	58.6%	106	68.9%	93	60.2%	101	65.4%
Michigan	74	45.9%	69	52.2%	70	44.3%	84	46.4%	73	47.9%
Wisconsin	27	88.9%	38	86.8%	34	85.3%	35	71.4%	24	70.8%