

**SUPPLEMENTARY BOX. Medical claims codes and algorithm used to identify persons with sickle cell anemia and define transcranial Doppler ultrasound screening, hydroxyurea use, and indication of severe complications — IBM MarketScan Multi-state Medicaid database,<sup>\*,†</sup> United States, 2010–2019**

Diagnosis or intervention	Definition/Algorithm	Codes
<b>Sickle Cell Anemia (SCA)</b>	3 or more claims with ICD versions 9 or 10 CM diagnosis codes for SCA or SCD without crisis (ICD 10 only) on different service dates within any 1-year period, 2010–2019	<p>ICD-9-CM: 282.61 (HbSS disease without crisis), 282.62 (HbSS disease with crisis)</p> <p>ICD-10-CM: D5700 (HbSS with crisis), D5701 (HbSS with acute chest syndrome), D5702, (HbSS with splenic sequestration), D571, (sickle cell disease without crisis)</p>
<b>TCD Screen</b>	A medical claim with a procedure code corresponding to TCD in 2019 (or 2014 for selected analyses of change in TCD screening)	<p>Procedure Codes:</p> <p>93886 Transcranial Doppler study of the intracranial arteries; complete study   93888 Transcranial Doppler study of the intracranial arteries; limited study   93890 Transcranial Doppler study of the intracranial arteries; vasoreactivity study   93892 Transcranial Doppler study of the intracranial arteries; emboli detection without intravenous microbubble injection   93893 Transcranial Doppler study of the intracranial arteries; emboli detection with intravenous microbubble injection</p>
<b>Hydroxyurea Use</b>	At least one pharmacy code claim with a National Drug Code corresponding to hydroxyurea in 2019 (or 2014 for selected analyses of change in hydroxyurea use)	<p>00003083050 - 00003-0830-50 HYDREA: 100 Capsule 500 MG &gt; BRISTOL-MYERS SQUIBB CO.</p> <p>00003633517 - 00003-6335-17 DROXIA: 60 Capsule 200 MG &gt; BRISTOL-MYERS SQUIBB CO.</p> <p>00003633617 - 00003-6336-17 DROXIA: 60 Capsule 300 MG &gt; BRISTOL-MYERS SQUIBB CO.</p> <p>00003633717 - 00003-6337-17 DROXIA: 60 Capsule 400 MG &gt; BRISTOL-MYERS SQUIBB CO.</p> <p>00054224725 - 00054-2247-25 HYDROXYUREA: 100 Capsule 500 MG &gt; ROXANE LABORATORIES, INC.</p> <p>00054824725 - 00054-8247-25 HYDROXYUREA: 100 Capsule 500 MG &gt; ROXANE LABORATORIES, INC.</p> <p>00555088202 - 00555-0882-02 HYDROXYUREA: 100 Capsule 500 MG &gt; TEVA PHARMACEUTICALS USA</p> <p>00603394621 - 00603-3946-21 HYDROXYUREA: 100 Capsule 500 MG &gt; QUALITEST PHARMACEUTICALS</p> <p>00677168001 - 00677-1680-01 HYDROXYUREA: 100 Capsule 500 MG &gt; UNITED RESEARCH LABORATORIES, INC.</p> <p>00904539460 - 00904-5394-60 HYDROXYUREA: 100 Capsule 500 MG &gt; MAJOR PHARMACEUTICALS</p> <p>00904693961 - 00904-6939-61 HYDROXYUREA: 100 Capsule 500 MG &gt; MAJOR PHARMACEUTICALS</p> <p>38779135402 - 38779-1354-02 HYDROXYUREA: 1 Powder &gt; MEDISCA, INC.</p> <p>38779135404 - 38779-1354-04 HYDROXYUREA: 1 Powder &gt; MEDISCA, INC.</p> <p>38779135405 - 38779-1354-05 HYDROXYUREA: 1 Powder &gt; MEDISCA, INC.</p> <p>38779135407 - 38779-1354-07 HYDROXYUREA: 1 Powder &gt; MEDISCA, INC.</p> <p>38779135408 - 38779-1354-08 HYDROXYUREA: 1 Powder &gt; MEDISCA, INC.</p> <p>42291032101 - 42291-0321-01 HYDROXYUREA: 100 Capsule 500 MG &gt; AVKARE, INC.</p> <p>49452365001 - 49452-3650-01 HYDROXYUREA: 1 Powder &gt; SPECTRUM PHARMACY PRODUCTS</p> <p>49452365002 - 49452-3650-02 HYDROXYUREA: 1 Powder &gt; SPECTRUM PHARMACY PRODUCTS</p> <p>49452365003 - 49452-3650-03 HYDROXYUREA: 1 Powder &gt; SPECTRUM PHARMACY PRODUCTS</p> <p>49452365004 - 49452-3650-04 HYDROXYUREA: 1 Powder &gt; SPECTRUM PHARMACY PRODUCTS</p> <p>49884072401 - 49884-0724-01 HYDROXYUREA: 100 Capsule 500 MG &gt; PAR PHARMACEUTICAL</p> <p>51285054802 - 51285-0548-02 HYDROXYUREA: 100 Capsule 500 MG &gt; DURAMED PHARMACEUTICALS, INC.</p> <p>51552085104 - 51552-0851-04 HYDROXYUREA: 1 Powder &gt; FAGRON INC</p> <p>51552085106 - 51552-0851-06 HYDROXYUREA: 1 Powder &gt; FAGRON INC</p>

		<p>51552085109 - 51552-0851-09 HYDROXYUREA: 1 Powder &gt; FAGRON INC  51927265500 - 51927-2655-00 HYDROXYUREA: 1 Powder &gt; PROFESSIONAL COMPOUNDING CENTERS  54569037800 - 54569-0378-00 HYDREA: 100 Capsule 500 MG &gt; A-S MEDICATION SOLUTIONS LLC  54569037801 - 54569-0378-01 HYDREA: 60 Capsule 500 MG &gt; A-S MEDICATION SOLUTIONS LLC  54569037802 - 54569-0378-02 HYDREA: 30 Capsule 500 MG &gt; A-S MEDICATION SOLUTIONS LLC  54569571500 - 54569-5715-00 HYDROXYUREA: 100 Capsule 500 MG &gt; A-S MEDICATION SOLUTIONS LLC  54868136700 - 54868-1367-00 HYDREA: 100 Capsule 500 MG &gt; PHYSICIANS TOTAL CARE  54868477300 - 54868-4773-00 HYDROXYUREA: 30 Capsule 500 MG &gt; PHYSICIANS TOTAL CARE  54868477301 - 54868-4773-01 HYDROXYUREA: 100 Capsule 500 MG &gt; PHYSICIANS TOTAL CARE  54868477302 - 54868-4773-02 HYDROXYUREA: 50 Capsule 500 MG &gt; PHYSICIANS TOTAL CARE  54868477303 - 54868-4773-03 HYDROXYUREA: 60 Capsule 500 MG &gt; PHYSICIANS TOTAL CARE  55154714300 - 55154-7143-00 HYDROXYUREA: 10 Capsule 500 MG &gt; CARDINAL HEALTH, INC.  57866443601 - 57866-4436-01 HYDROXYUREA: 30 Capsule 500 MG &gt; VIBRANTA, INC.  58063097909 - 58063-0979-09 MYLOCEL: 60 Tablet 1 GM &gt; MGI PHARMA, INC.  60429026501 - 60429-0265-01 HYDROXYUREA: 100 Capsule 500 MG &gt; GOLDEN STATE MED. SUPPLY (GSMS) INC  68084028401 - 68084-0284-01 HYDROXYUREA: 100 Capsule 500 MG &gt; AMERICAN HEALTH PACKAGING  68084028411 - 68084-0284-11 HYDROXYUREA: 1 Capsule 500 MG &gt; AMERICAN HEALTH PACKAGING  71770010060 - 71770-0100-60 SIKLOS: 60 Tablet 100 MG &gt; MEDUNIK USA, INC.  71770010560 - 71770-0105-60 SIKLOS: 60 Tablet 100 MG &gt; MEDUNIK USA, INC.  71770012030 - 71770-0120-30 SIKLOS: 30 Tablet 1000 MG &gt; MEDUNIK USA, INC.</p>
<p><b>Indication of Severe Disease:</b> one or more hospitalizations for acute chest syndrome in current or any previous year</p>	<p>At least one inpatient claim with ICD versions 9 or 10 CM diagnosis codes for acute chest syndrome between 2010 and 2019.</p>	<p>ICD-9-CM: 517.3 Acute chest syndrome ICD-10-CM: D57411 Sickle-cell thalassemia with acute chest syndrome   D5701 Hb-SS disease with acute chest syndrome   D57211 Sickle-cell/Hb-C disease with acute chest syndrome   J99 Respiratory disorders in diseases classified elsewhere   D57811 Other sickle-cell disorders with acute chest syndrome.</p>
<p><b>Indication of Severe Disease:</b> highest number hospitalizations for pain crises in current or any previous year</p>	<p>Maximum count of inpatient claims with ICD versions 9 or 10 CM diagnosis codes for pain in calendar year between 2010 and 2019.</p>	<p>ICD-9-CM: 282.42 Sickle-cell thalassemia with crisis   282.62 Hb-SS disease with crisis   282.64 Sickle-cell/Hb-C disease with crisis   282.69 Other sickle-cell disease with crisis ICD-10-CM: D5700 Hb-SS disease with crisis, unspecified   D570: (Non-Billable Dx) Hb-SS disease w crisis   D5709: Hb-SS disease w crisis w other specified complication   D57218: Sickle-cell/Hb-C disease w crisis w other specified complication   D57219 Sickle-cell/Hb-C disease with crisis, unspecified   D5721: (Non-Billable Dx) Sickle-cell/Hb-C disease w crisis   D57418: Sickle-cell thalassemia, unspecified, w crisis w other specified comp   D57419 Sickle-cell thalassemia with crisis, unspecified   D5741: (Non-Billable Dx) Sickle-cell thalassemia w crisis   D57438: Sickle-cell thalassemia beta zero w crisis w other specified comp   D57439: Sickle-cell thalassemia beta zero w crisis, unspecified   D5743: (Non-Billable Dx) Sickle-cell thalassemia beta zero w crisis   D57458: Sickle-cell thalassemia beta plus w crisis w other specified comp   D57459: Sickle-cell thalassemia beta plus w crisis, unspecified   D5745: (Non-Billable Dx) Sickle-cell thalassemia beta plus w crisis   D57818: Other sickle-cell disorders w crisis w other specified complication   D57819 Other sickle-cell disorders with crisis, unspecified   D5781: (Non-Billable Dx) Sickle-cell disorders NEC w crisis</p>

**Abbreviations:** CM = Clinical Modification; ICD-9-CM = *International Classification of Diseases, Ninth Revision, Clinical Modification*; ICD-10-CM = *International Classification of Diseases, Tenth Revision, Clinical Modification*; SCA = sickle cell anemia; SCD = sickle cell disease; TCD = transcranial Doppler.

\* MarketScan Medicaid data used in this analysis were available for claims beginning in 2010. The primary sample selected for this analysis included children who were continuously enrolled in Medicaid in 2019. However, because the number of a child's healthcare visits in any given year varies widely, the algorithms used to identify which of the children in the initial 2019 sample had SCA and to classify SCA disease severity used children's claims data for 2019 or any previous data year (i.e., claims between 2010 and 2019).

† Because there are no specific ICD-10 diagnosis codes that characterize care related to SCA without a crisis, the more general code D571 (SCD without crisis) was included in the SCA algorithm. Despite the inclusion of this code, the algorithm demonstrated high sensitivity (91% and 78%), high specificity (93% and 92%), and high area under the ROC curve (92% and 85%), in a validation study in which the algorithm was applied to children enrolled in Medicaid in 2 states and compared to their newborn screening genotyping results.