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 FS,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	282.61, 282.62

Table 2. Blood Tests for the Emergency Department Management of Sickle Cell Disease

Definition	Procedure Code	Short Description	Long Description
Pulse oximetry reading	0Y4306	Pulse oximetry	Pulse oximetry
Complete blood count	85025	Complete CBC w/auto diff WBC	Blood count; complete (CBC), automated (HGB, HCT, RBC, WBC and platelet count) and automated differential WBC count
Complete blood count	85027	Complete CBC automated	Blood count; complete (CBC), automated (HGB, HCT, RBC, WBC and platelet count)
Complete blood count	85014	Hematocrit	Blood count; hematocrit (HCT)
Complete blood count	85018	Hemoglobin	Blood count; hemoglobin (HGB)
Reticulocyte count	85044	Manual reticulocyte count	Blood count; reticulocyte, manual
Reticulocyte count	85045	Automated reticulocyte count	Blood count; reticulocyte, automated
Reticulocyte count	85046	Reticulocyte/HgB concentrate	Blood count; reticulocytes, automated, including one or more cellular parameters (EG, reticulocyte hemoglobin content (CHR), immature reticulocyte fraction (IRF), reticulocyte volume (MRV), RNA content), direct measurement
Blood culture	87040	Blood culture	Isolation and identification of microorganisms and susceptibility testing, when appropriate. Other isolated organisms, i.e., anaerobes, yeast, etc.) may be referred for identification and/or susceptibility testing if medically indicated AND a separate culture procedure has NOT yielded the same organism(s).

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
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

Table 4. Evidence Supporting Appropriate Emergency Department Blood Testing in Children with Sickle Cell Disease

Type of Evidence	Key Findings	Level of Evidence (USPSTF ranking*)	Citation(s)
Clinical guidelines	All children with SCD who have fever (greater than 38.5°C or 101°F) and other signs of infection should be evaluated promptly. The younger the child, the higher the index of suspicion. In a child with no obvious sources of infection, a minimum evaluation should include blood culture, complete blood count, reticulocyte count, and chest x-ray for children under 3 years of age. Immediately after the blood is taken, the child should be given broad-spectrum antibiotics, preferably intravenously (p. 28). Ideally, children with SCD are followed at a practice or center that allows for comprehensive management of their disease. These facilities should have 24-hour access to medical consultants, hematology and microbiology laboratories, and a blood bank, among other services (p. 29).	III	National Heart, Lung, and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health, Bethesda, MD; 2002.
Clinical guidelines	A child with fever or pallor and listlessness should always be initially evaluated, if possible, at a site where complete blood cell (CBC) and reticulocyte counts, blood cultures, intravenous antibiotics, and red blood cell transfusions are readily available. Because patients with SCD develop splenic dysfunction as early as 3 months of age, they are at high risk for septicemia and meningitis with pneumococci and other encapsulated bacteria. Thus, all patients with temperature greater than 38.5°C require rapid triage and physical assessment, urgent CBC and reticulocyte counts, blood culture (plus cerebrospinal fluid analysis and other cultures as indicated), and prompt administration of a broad-spectrum parenteral antibiotic, such as ceftriaxone sodium, cefuroxime, or cefotaxime sodium (p. 529).	III	American Academy of Pediatrics Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. Pediatrics 2002; 109(3):526-35.
Clinical guidelines	Infectious processes are a serious cause of morbidity in the child with SCD, as these patients have decreased immunologic function due to chronic microinfarcts within the spleen. Functional asplenia is found in more than 90% of SCD patients by age 5; therefore, greater vigilance for occult bacteremia, meningitis, and sepsis must be practiced … the presence of fever in patients with SCD should be treated with a higher index of suspicion for systemic infection than in the non-SCD patient (p.222).	III	Taylor S, et al. Emergency nursing care of pediatric sickle cell patients: Meeting the challenge. Pediatr Emerg Care 2001; 17(3):220-5.

	Assessment in the ED of pediatric patients with SCD complications should start a pulse oximetry reading (compared, if possible, to the patient's normal pulse ox reading, if known) … Laboratory tests should include a complete blood count to assess the level of anemia, reticulocyte count to assess the body's response to anemia, and a blood culture for febrile patients (pp. 221-222).		
Clinical guidelines	For children with SCD who present with fever or history of fever (greater than or equal to 38.5°C), a complete blood count, reticulocyte count, and blood culture should be obtained. Children with SCD with a fever greater than or equal to 38.5°C should be given parenteral broad spectrum antibiotic treatment within 60 minutes of triage.	III	Wang CJ, et al. Quality of care indicators for children with sickle cell disease. Pediatrics 2011; 128:484-93.

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.

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

Site	Eligibility criteria/measure numerators	Number of records reviewed	N agreed (%)	Kappa statistic
Hospital #1	Fever criterion	3	100	1.00
	Pulse oximetry	0	-	-
	Complete blood count	0	-	-
	Reticulocyte	0	-	-
	Blood Culture	0	-	-
Hospital #2	Fever criterion	7	100	1.00
	Pulse oximetry	4	100	1.00
	Complete blood count	4	100	1.00
	Reticulocyte	4	100	1.00
	Blood Culture	4	100	1.00
Hospital #3	Fever criterion	19	84	0.68
	Pulse oximetry	10	50	0.00
	Complete blood count	10	100	1.00
	Reticulocyte	10	100	1.00
	Blood Culture	10	100	1.00
All sites	Fever criterion	29	90	0.79
	Pulse oximetry	14	64	0.10
	Complete blood count	14	100	1.00
	Reticulocyte	14	100	1.00
	Blood Culture	14	100	1.00

Note: This measure includes a numerator that assesses whether the patient received all four required tests within 60 minutes of initial contact in the ED. ITT calculations are not included for the overall numerator, since it would be redundant with IRRs calculated for the individual numerators.

Table 6. Appropriate Outpatient Blood Testing for Children with Sickle Cell Disease

Site	Pulse Oximetry Numerator		Complete Blood Count Numerator		Reticulocyte Numerator		Blood Culture Numerator		Overall Numerator		Denominator
	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	
Hospital #1	100%	10	20%	2	20%	2	30%	3	10%	1	10
Hospital #2	100%	14	21%	3	21%	3	7%	1	7%	1	14
Hospital #3	39%	39	26%	26	25%	25	44%	44	10%	10	99
All Sites	51%	63	25%	31	24%	30	39%	48	10%	12	123

Table 7. Race/Ethnicity for Newborns with SCD in Michigan (percent), 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