CME

Which Febrile Children With Sickle Cell Disease Need a Chest X-Ray?

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Abstract

Objective: Controversy exists regarding which febrile children with sickle cell disease (SCD) should receive a chest x-ray (CXR). Our goal is to provide data informing the decision of which febrile children with SCD presenting to the emergency department (ED) require a CXR to evaluate for acute chest syndrome (ACS).

Methods: Retrospective chart review of children ages 3 months to 21 years with SCD presenting to the ED at one of two academic children's hospitals with fever ≥38.5°C between January 1, 2010, and December 31, 2012. Demographic characteristics, respiratory symptoms, and laboratory results were abstracted. The primary outcome was the presence of ACS. Binary recursive partitioning was performed to determine predictive factors for a diagnosis of ACS.

Results: A total of 185 (10%) of 1,837 febrile ED visits met ACS criteria. The current National Heart, Lung, and Blood Institute (NHLBI) consensus criteria for obtaining a CXR (shortness of breath, tachypnea, cough, or rales) identified 158 (85%) of ACS cases, while avoiding 825 CXRs. Obtaining a CXR in children with NHLBI criteria or chest pain and in children without those symptoms but with a white blood cell (WBC) count ≥18.75×10^9/L or a history of ACS identified 181 (98%), while avoiding 430 CXRs.

Conclusion: Children with SCD presenting to the ED with fever and shortness of breath, tachypnea, cough, rales, or chest pain should receive a CXR due to high ACS rates. A higher WBC count or history of ACS in a child without one of those symptoms may suggest the need for a CXR. Prospective validation of these criteria is needed.

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The National Heart, Lung, and Blood Institute (NHLBI) recommends urgent medical evaluation for all children with sickle cell disease (SCD) who develop a fever ≥38.5°C.[1] As part of the medical evaluation, all febrile children with SCD should have a complete blood count and blood culture obtained and receive intravenous antibiotics.[1] Furthermore, those children with shortness of breath, tachypnea, cough, and/or rales should have an immediate chest x-ray (CXR) to investigate for acute chest syndrome (ACS). The NHLBI expert panel proposed this CXR recommendation for ACS utilizing a consensus process, citing insufficient evidence to determine which children were at increased risk of having ACS.

ACS is the second leading cause of hospitalization in children with SCD, accounting for up to 25% of premature deaths in this patient population.[2–5] Patients with ACS frequently present with fever due to the underlying infectious or noninfectious etiology of the disease such as pulmonary infarction or microvasculature sludging.[6–8] Given the high susceptibility of patients with SCD to serious bacterial infections,[9] it is critical that any fever be considered a potential emergency situation. The high morbidity of ACS mandates that evaluation for ACS is performed when indicated. A previous study by Vichinsky et al.[2] found that 99% of cases of ACS had a new pulmonary infiltrate on CXR and, therefore, positive CXR findings in the setting of fever can be considered diagnostic for ACS.

A single-site study concluded that history and physical examination are not sensitive enough to determine which febrile SCD patients require a CXR.[10] Utilizing a

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prospectively collected physician questionnaire, Morris et al.[10] found that 61% of ACS cases were not clinically suspected based on physical evaluation. Specifically, no symptoms or examination findings were able to identify patients with ACS, and 57% of the patients had completely normal examination findings. Based on these results, the authors recommend a low threshold for obtaining a CXR for all febrile SCD patients. However, it is important to note that despite these known study results, the NHLBI expert panel did not recommend a CXR for all febrile patients with SCD.

Due to these conflicting recommendations in the literature, significant variation exists in clinical practice. A recent analysis of the 2010 Pediatric Health Information System demonstrated that the percentage of febrile children with SCD who received a CXR in the emergency department (ED) ranged from less than 40% to more than 90% across 36 tertiary care children's hospitals in the United States.[11] This variation in care suggests that there is an absence of data-driven evidence supporting the clinical guidelines for CXR utilization in the ED. To our knowledge, this project will be the first multicenter study to provide data that will aid providers in deciding which patients presenting to the ED with SCD and fever require a CXR to evaluate for ACS.

METHODS

Study Design

We conducted a retrospective chart review of all ED visits made by children ages 3 months to 21 years with a diagnosis of SCD or sickle cell crisis between January 1, 2010, and December 31, 2012, at two children's hospitals. Both sites are academic medical centers with a comprehensive sickle cell center and an ED staffed by pediatric emergency medicine physicians. The study was reviewed and approved as expedited research with a waiver of informed consent by the institutional review board at both centers.

Methods and Measurements

Our inclusion criteria were febrile children with SCD presenting to the ED. Charts were included as having SCD based on ICD-9-CM discharge diagnosis codes for various diagnoses of SCD (282.41, 282.42, 282.6, 282.61, 282.62, 282.63, 282.64, 282.68, 282.69), which include both crisis and noncrisis codes. All charts were retrieved via an electronic data extraction using these ICD codes. As it has been shown that using ICD-9-CM codes to identify fever in patients with SCD is unreliable, we individually reviewed all retrieved charts for documented fever.[12] Any chart with a documented fever $\geq$38.5°C within the past 24 hours at home, in the ED triage record, or the ED physician note, was included. If the chart did not provide a specific time when the fever started but instead indicated that the fever began within the past 24 hours (e.g. "yesterday," "last night," "this morning," or "one day of fever"), the chart was included. We had 100% agreement when 80 charts were reviewed by two reviewers for the fever criteria.

Visits were excluded due to antibiotic pretreatment as this would influence testing. Patients were classified as pretreated if they received ceftriaxone within 24 hours in the same ED or if noted in the chart to have been received elsewhere. Visits were not excluded for use of penicillin, amoxicillin, or azithromycin at home, as these were believed to be for prophylactic use.

Demographic characteristics and signs and symptoms of a respiratory illness were abstracted from the patient chart. Triage vitals were used for all vital signs. The patient's genotype was retrieved from the sickle cell database maintained by the comprehensive sickle cell center at each institution. All respiratory symptoms were documented (Table 1), with specific categorization into groupings consistent with high-risk symptoms as defined by the NHLBI guidelines for evaluation of ACS.[1] A patient was considered to have shortness of breath if any of the following was documented in the chart: trouble breathing, shortness of breath, decreased air movement, decreased breath sounds, increased work of breathing, respiratory distress, shallow breathing, retractions, or chest tightness. The absence of documentation of respiratory symptoms was treated as negative for analysis. A patient was classified as having tachypnea based on the age-based 99th percentile Lancet criteria.[13] A patient was classified as "ill-appearing" if the ED documentation contained one of the following terms: ill-appearing, toxic, limp, unresponsive, gray, cyanotic, apnea, weak cry, poorly perfused, grunting, listless, lethargic, or irritable. Terms such as slightly irritable, mildly irritable, cranky, fussy, mottled, slightly delayed capillary refill, tired, and sleepy were not considered ill-appearing.[14] A patient was recorded as having a central line if it was noted in the patient's chart or if there was documentation that a central line was accessed. If the patient received a complete blood count or reticulocyte count in the ED, these laboratory results were abstracted. Finally, history of ACS was recorded if noted in the ED documentation; absence of documentation was treated as negative for analysis.

Data were abstracted by a trained medical student and research assistant at one site and a single medical student at the second site under the supervision of the site principal investigators (PI). The reviewers were blind to the study hypothesis. An initial set of five charts was collaboratively abstracted with each site's study PI to ensure an accurate understanding of all data elements. A formal coding manual and custom Access database (Microsoft Access, 2010) were used to standardize abstraction between sites. Site PIs periodically met with abstractors to review the abstraction process and answer questions. Coding decisions were communicated between sites to maintain consistency.

Outcomes

The primary outcome was the presence of ACS. Radiology reports from all ED-obtained CXRs were reviewed and categorized into a priori defined categories as: normal, infiltrate (if the impression included the words ACS, pneumonia, infiltrate or consolidation or opacification, atelectasis (if the impression explicitly stated so), atelectasis versus infiltrate (impression includes the words atelectasis and infiltrate), and other (not meeting any of the previous four categories). All radiology reports classified as other were individually reviewed a second time by the medical student abstractors and

Table 1
Demographics Characteristics and Clinical Findings

	ACS (−), *n* = 1,652	ACS (+), *n* = 185	p-value	OR (95% CI)
Age (y)	3.2 (1.4–9.1)	5.0 (2.2–10.2)	<0.001	1.0 (1.0–1.1)
Female	828 (50.1)	98 (53.0)	0.46	1.1 (0.8–1.5)
Severe disease (Hb-SS/Hb-SB^0)	1,070 (64.8)	129 (69.7)	0.18	0.8 (0.6–1.1)
Fever in the ED	1,199 (72.6)	151 (81.6)	<0.01	1.7 (1.1–2.5)
Hypoxia (triage pulse ox <93%)	83 (5.0)	33 (17.8)	<0.001	4.1 (2.7–6.3)
Central line	26 (1.6)	0 (0)	0.10*	0.3 (0.0–2.5)
Ill-appearing	27 (1.6)	13 (7.0)	<0.001	4.6 (2.3–9.0)
History of ACS	586 (35.5)	104 (56.2)	<0.001	2.3 (1.7–3.2)
ED laboratory values				
WBC count ($\times 10^9$/L)	14.1 (9.8–19.5)	18.5 (13.1–25.2)	<0.001	1.1 (1.0–1.1)
Neutrophils (%)	65 (49–75)	69 (61–77)	<0.001	4.8 (1.9–12.2)
Hemoglobin (g/dL)	9.3 (8.0–10.5)	8.3 (7.4–9.8)	<0.001	0.7 (0.7–0.8)
Platelets ($\times 10^9$/L)	372 (271–478)	381 (280–517)	0.71	1.0 (1.0–1.0)
Reticulocytes (%)	8 (4–14)	10 (4–16)	<0.01	13.9 (1.6–120.4)
Respiratory symptoms				
Shortness of breath†	111 (6.7)	43 (23.2)	<0.001	4.2 (2.8–6.2)
Tachypnea (Lancet)‡	299 (18.1)	74 (40.0)	<0.001	3.0 (2.2–4.2)
Cough	662 (40.1)	139 (75.1)	<0.001	4.5 (3.2–6.4)
Rales/crackles	0 (0)	2 (1.1)	0.01*	10.0 (8.7–11.5)
Chest pain	129 (7.8)	41 (22.2)	<0.001	3.4 (2.3–5.0)
Wheezing	42 (2.5)	13 (7.0)	0.001	2.9 (1.5–5.5)
Sore throat	196 (11.9)	16 (8.6)	0.19	0.7 (0.4–1.2)
Rhinorrhea	559 (33.8)	63 (34.1)	0.95	1.0 (0.7–1.4)
Congestion	150 (9.1)	22 (11.9)	0.21	1.4 (0.8–2.2)
Sneezing	11 (0.7)	1 (0.5)	1.00*	0.8 (0.1–6.3)
Respiratory symptom composites				
NHLBI guideline§‖	854 (51.7)	158 (85.4)	<0.001	5.5 (3.6–8.3)
NHLBI guideline or chest pain	894 (54.1)	162 (87.6)	<0.001	6.0 (3.8–9.3)
NHLBI guideline or wheezing	858 (51.9)	158 (85.4)	<0.001	5.4 (3.6–8.2)
NHLBI guideline or chest pain or wheezing	897 (54.3)	162 (87.6)	<0.001	5.9 (3.8–9.3)
Any respiratory symptoms¶	1,198 (72.5)	173 (93.5)	<0.001	5.5 (3.0–9.9)

Data are reported as median (IQR) or *n* (%).
ACS = acute chest syndrome; IQR = interquartile range; NHLBI = National Heart, Lung, and Blood Institute; WBC = white blood cell.
*Calculated using Fisher's exact test.
†One or more of the following: trouble breathing (*n* = 119), shortness of breath (*n* = 12), decreased air movement (*n* = 1), decreased breath sounds (*n* = 3), increased work of breathing (*n* = 21), respiratory distress (*n* = 1), shallow breathing (*n* = 1), retractions (*n* = 1), or chest tightness (*n* = 7).
‡Data from Fleming et al.[13]
§Guideline recommends immediate CXR in children whose SCD is accompanied by shortness of breath, tachypnea, cough and/or rales.[1]
‖All NHLBI consensus panel guideline composites were calculated using the Lancet criteria for tachypnea.
¶Any of the respiratory symptoms noted in the "Respiratory symptoms" subsection of this table.

none of these radiology reports were consistent with ACS. ACS was defined based on our a priori classifications as the occurrence of one of three scenarios: 1) radiology reported infiltrate on CXR, 2) infiltrate versus atelectasis on CXR with patient admitted to the inpatient unit and having a discharge diagnosis of ACS or pneumonia, or 3) no ED CXR obtained but a CXR within 48 hours read as positive for infiltrate. Forty-eight hours was conservatively chosen to indicate that the ACS was probably present at the initial visit and not simple disease progression.

Data Analysis

With 3 years of medical record review at each site, we estimated that approximately 2000 febrile visits would be reviewed. If ACS occurred in 15% this would yield 300 cases of ACS. Using the standard 10 outcomes per explanatory variable, our sample size would be adequate to develop a parsimonious decision rule. We first created a composite variable using the NHLBI guideline for performing a CXR in febrile children with SCD (Table 1, section footnote).[1] Additional composite variables were created incorporating wheezing and chest pain with the NHLBI criteria. The variables abstracted from the patient chart and these composites were compared between those with ACS and those without ACS using a chi-square test for categorical variables (and Fisher's exact test where indicated due to small sample sizes) and a Wilcoxon rank-sum test for nonparametric continuous variables with a Type I error rate of <0.05. Similar to previous studies, all visits were treated as independent occurrences for analysis.[10,11,16] We performed binary recursive partitioning using classification and regression tree (CART) analysis to determine predictive factors for a diagnosis of ACS (CART PRO 6.0, Salford Systems).[15] Recursive partitioning progressively divides patients into subpopulations that only have a particular outcome and is therefore a method to use

when trying to develop a decision rule with high sensitivity.[17–19] We also calculated the sensitivity, specificity, positive and negative predictive values, positive and negative likelihood ratios, 95% confidence intervals (CIs), and area under the curve (AUC) for the NHLBI criteria and the final primary analysis CART model.

Finally, we performed a secondary analysis using the same outcomes, but including only those children who had a CXR obtained during the ED visit. This analysis was performed to evaluate whether risk factors and recursive partitioning analyses would be consistent when applied to all patients versus those who had a CXR obtained.

RESULTS

There were a total of 6,400 visits made by 1207 children ages 3 months to 21 years with SCD between January 1, 2010, and December 31, 2012 (Figure 1). Of these visits, 1,881 (29.4%) had a documented fever ≥38.5°C. Forty-four visits were excluded due to treatment with ceftriaxone within 24 hours of the visit. Therefore, the final sample included 1,837 febrile ED visits made by 697 children with SCD.

Demographic characteristics are shown in Table 1. Fifty percent of the population was female. The median age was 3.5 years (interquartile range [IQR] = 1.5 to 9.2 years); 65% had either genotype HbSS or genotype HbSβ^0.

Prevalence of ACS

A total of 185 (10%) of 1,837 visits met criteria for ACS. One-hundred forty-two were diagnosed based on CXR showing infiltrate at initial visit, 32 visits had a CXR of atelectasis versus infiltrate and had a discharge diagnosis of ACS, and 11 patients had no ED CXR obtained but had a CXR positive for infiltrate within 48 hours.

Symptoms Related to ACS

Ninety-four percent of visits made by patients with ACS had one or more respiratory signs or symptoms. The percentage of patients diagnosed with ACS who had each clinical or laboratory finding are listed in Table 1. The most common respiratory finding in these patients was cough (75.1%) followed by tachypnea (40.0%). Only 7.0% of patients with ACS were documented as ill-appearing in the ED and 56.2% had a history of ACS.

Evaluation of the composite variables listed in Table 1 showed 158 patients with ACS (85%) had one or more of the respiratory findings included in the NHLBI recommendations (shortness of breath, tachypnea, cough, and/or rales). Relying on the NHLBI guideline alone would have missed 27 ACS diagnoses, but avoided 825 CXRs. When chest pain was added, 162 (88%) of the patients with ACS were identified, and 781 CXRs were avoided.

CART Analysis

The CART model identifying patients at increased risk of ACS is shown in Figure 2. In our CART model, the first split was the NHLBI consensus criteria of shortness of breath, tachypnea, cough, and/or rales plus chest pain. A total of 1,056 (57.5%) of the 1,837 total patients with fever met these criteria; 162 (15.3%) of those were diagnosed with ACS. The test characteristics for the model are shown in Table 2. Of the 781 who did not meet these criteria, 23 (2.9%) were diagnosed with ACS. Next, the CART analysis split this lower risk group based on white blood cell (WBC) count. Of the 781, a

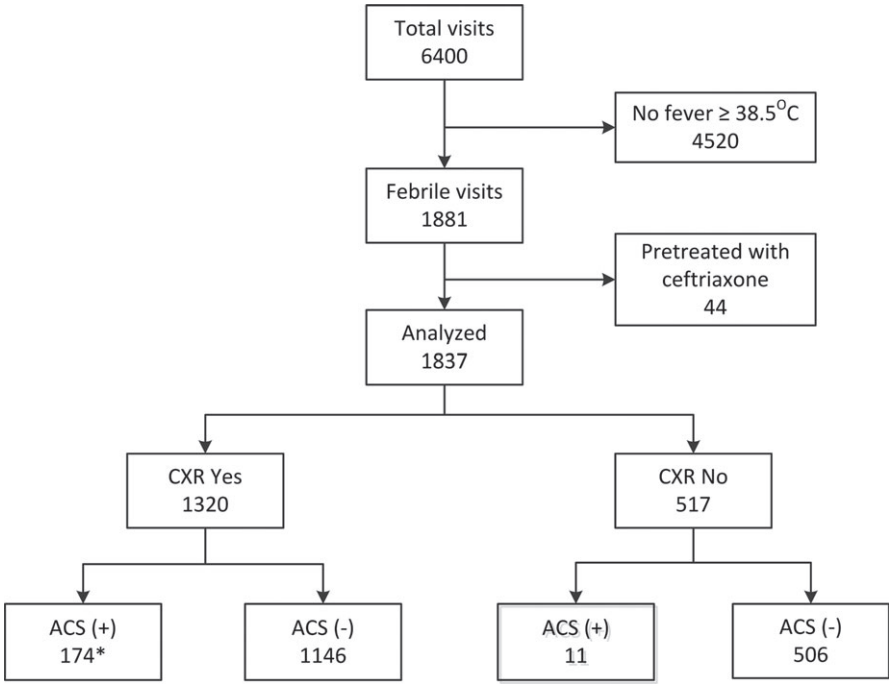

Figure 1. Flow diagram of sample population. *Infiltrate on CXR = 142; infiltrate versus atelectasis on CXR with discharge diagnosis of ACS = 32. ACS = acute chest syndrome; CXR = chest x-ray.

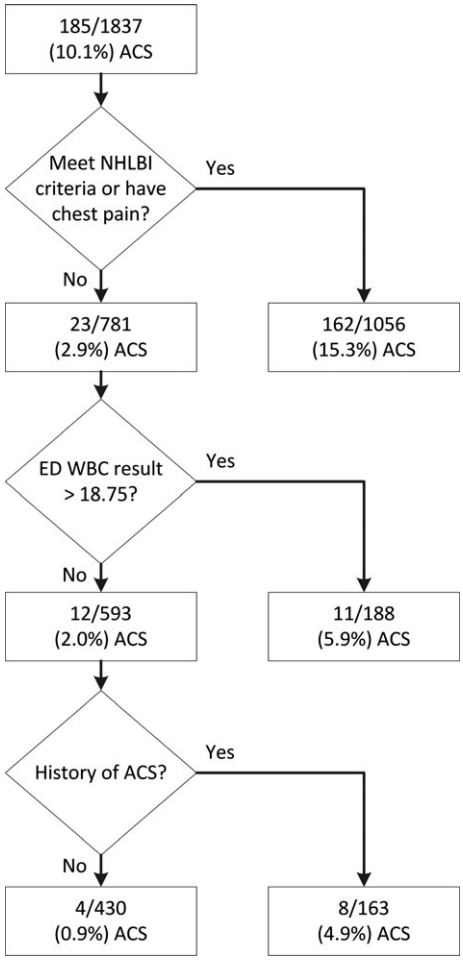

Figure 2. CART model for CXR utilization. ACS= acute chest syndrome; CART = classification and regression tree; CXR = chest x-ray; NHLBI = National Heart, Lung, and Blood Institute; WBC = white blood cell.

total of 188 (24.1%) had a WBC count >18.75 × 10^9/L; 11 (6%) of those had ACS. Including those with a history of ACS, we identified 181 (98%) of 185 cases of ACS while avoiding 430 CXRs. Clinical characteristics and outcomes of the four patients missed by the CART model are shown in Table 3.

Of the 517 visits at which an ED CXR was not performed (Table 1), 170 (33%) had at least one of the risk criteria noted in the NHLBI guideline. When the additional risk criteria identified by the CART model were added, 333 visits (64%) met at least one of the risk criteria for obtaining an ED CXR. More specifically, of the 11 visits at which an ED CXR was not performed but subsequently developed ACS, nine (82%) met one or more of the CART model criteria for obtaining a CXR.

Secondary Analysis in Those With a CXR Obtained

Chest x-rays were performed at 1,320 (71.9%) visits. Of those children who received a CXR, 174 (13.2%) had ACS. The clinical characteristics, laboratory values and respiratory symptoms and the outcome of ACS are shown in Table 4. A CART analysis on this subpopulation revealed the primary split again to be the NHLBI

Table 2
Test Characteristics of Model in Predicting ACS

	Sensitivity (95% CI)	Specificity (95% CI)	Positive Predictive Value (95% CI)	Negative Predictive Value (95% CI)	Positive Likelihood Ratio	Negative Likelihood Ratio	AUC
NHLBI guideline or chest pain	87.6% (81.9%–92.0%)	45.9% (43.5%–48.3%)	15.3% (13.2%–17.7%)	97.1% (95.6%–98.1%)	1.6	0.3	0.667
NHLBI or chest pain or WBC count >18.75 × 10^9/L	93.5% (88.9%–96.6%)	35.2% (32.9%–37.5%)	13.7% (12.0%–16.0%)	98.0% (96.5%–99.0%)	1.4	0.2	0.643
NHLBI or chest pain or WBC count >18.75 × 10^9/L or history of ACS	97.8% (94.6%–99.4%)	25.8% (23.7%–28.0%)	12.9% (11.2%–99.8%)	99.1% (97.6%–99.8%)	1.3	0.1	0.618

ACS = acute chest syndrome; AUC = area under the curve; NHLBI = National Heart, Lung, and Blood Institute.

Table 3
Clinical Characteristics and Outcomes of Patients Missed by CART Model

	Case 1	Case 2	Case 3	Case 4
Age (y)	1.8	0.6	2.0	2.2
Sex	Male	Male	Female	Female
Genotype	Hb-SC	Hb-SS	Hb-SC	Hb-SC
Fever in ED	Yes	Yes	No	Yes
WBC count ($\times 10^9$/L)	13.3	8.9	8.9	15.3
Hemoglobin (g/dL)	10.3	9.1	10.6	10.3
Respiratory symptoms	None	None	None	None
ED CXR impression	No CXR	No CXR	Infiltrate vs. atelectasis	Infiltrate
Disposition	Hospitalized	Hospitalized	Hospitalized	Discharged*
Length of stay (days)	2	7	1	0
Final diagnosis	ACS	ACS, bacteremia	ACS	Viral syndrome

ACS = acute chest syndrome; CXR = chest x-ray; WBC = white blood cell.
*Patient was called to return to ED the next day after radiologist read CXR as infiltrate. Patient had no respiratory distress, hypoxia or fever and was discharged home on oral antibiotics with a final diagnosis of ACS.

Table 4
Demographics Characteristics and Clinical Findings in the Subset of Children With a CXR Performed

	ACS (–), *n* = 1,146	ACS (+), *n* = 174	p-value	OR (95% CI)
Age (y)	2.7 (1.3–8.3)	5.1 (2.2–10.3)	<0.001	1.0 (1.0–1.1)
Hypoxia (triage pulse ox < 93%)	77 (6.7)	33 (19.0)	<0.001	3.2 (2.1–5.1)
Ill-appearing	25 (2.2)	12 (6.9)	<0.001	3.3 (1.6–6.7)
History of ACS	397 (34.6)	99 (56.9)	<0.001	2.5 (1.8–3.3)
ED laboratory values				
WBC ($\times 10^9$/L)	14.2 (9.8–19.6)	18.5 (13.1–25.9)	<0.001	1.1 (1.0–1.1)
Neutrophils (%)	63 (48–75)	68 (61–77)	<0.001	6.5 (2.4–17.3)
Hemoglobin (g/dL)	9.2 (8.0–10.4)	8.3 (7.4–9.8)	<0.001	0.8 (0.7–0.9)
Respiratory symptoms				
Shortness of breath*	107 (9.3)	43 (24.7)	<0.001	3.2 (2.1–4.7)
Tachypnea (Lancet)†	233 (20.3)	73 (42.0)	<0.001	2.8 (2.0–4.0)
Cough	551 (48.1)	134 (77.0)	<0.001	3.6 (2.5–5.2)
Rales/crackles	0 (0)	2 (1.1)	0.01‡	7.7 (6.7–8.8)
Chest pain	117 (10.2)	41 (23.6)	<0.001	2.7 (1.8–4.0)
Wheezing	40 (3.5)	13 (7.5)	<0.05	2.2 (1.2–4.3)
Respiratory symptom composites				
NHLBI guideline§‖	689 (60.1)	153 (87.9)	<0.001	4.8 (3.0–7.7)
NHLBI guideline or chest pain	721 (62.9)	157 (90.2)	<0.001	5.4 (3.3–9.1)

Data are reported as median (IQR) or *n* (%).
ACS = acute chest syndrome; CXR = chest x-ray; IQR = interquartile range; NHLBI = National Heart, Lung, and Blood Institute; WBC = white blood cell.
*One or more of the following: trouble breathing (*n* = 119), shortness of breath (*n* = 12), decreased air movement (*n* = 1), decreased breath sounds (*n* = 3), increased work of breathing (*n* = 21), respiratory distress (*n* = 1), shallow breathing (*n* = 1), retractions (*n* = 1), or chest tightness (*n* = 7).
†Data from Fleming et al.[13]
‡Calculated using Fisher's exact test.
§Guideline recommends immediate CXR in children whose SCD is accompanied by shortness of breath, tachypnea, cough and/or rales.[1]
‖All NHLBI consensus panel guideline composites were calculated using the Lancet criteria for tachypnea.

consensus criteria of shortness of breath, tachypnea, cough, and/or rales plus chest pain. A total of 878 (66.5%) of patients met these criteria and 157 (17.9%) of those were diagnosed with ACS. Of the 442 who did not meet these criteria, 17 (3.8%) were diagnosed with ACS. The next CART split was the combined WBC count >18.75 $\times$ 10^9/L or history of ACS. A total of 196 children met one of these criteria; 15 (7.7%) were diagnosed with ACS. The remaining 246 children met none of these criteria, and two (0.8%) had a diagnosis of ACS.

DISCUSSION

Our multicenter study provides data to guide future CXR utilization in children with SCD and fever who present to the ED. Our results provide data to support the consensus panel assertion that shortness of breath, tachypnea, cough, and/or rales are associated with higher rates of ACS. The current NHLBI consensus criteria would have identified 85.4% of patients subsequently diagnosed with ACS. Adding chest pain to the NHLBI consensus panel recommendations would allow

for the capture of 87.6% of children with ACS, and would find ACS in approximately 15% of patients in whom a CXR was obtained. Those children not meeting NHLBI consensus guidelines had a 2.9% chance of ACS, a risk that could be decreased with routinely collected additional information concerning WBC and history of ACS.

When other respiratory signs and symptoms such as hypoxia, wheezing, sore throat, rhinorrhea, and congestion were included in the CART analysis, the patients identified with ACS did not improve substantially. In isolation (without any positive NHLBI consensus panel symptoms) genotype or clinically ill-appearing were not associated with risk of ACS to warrant obtaining a CXR.[3,5]

The current criterion standard for determining the necessity of a CXR for febrile children with SCD, the NHLBI consensus panel guideline, does not recommend a CXR for all febrile patients with SCD. In our analysis, it would miss 27 ACS diagnoses. Our data suggest that there is strong evidence to add chest pain to these high-risk criteria, while a high WBC count and history of ACS can further identify patients at risk for ACS. Given the previously documented variability in practice, it is clear that data-driven evidence is needed to further determine which patients require a CXR.[11,20] While neither the NHLBI guideline nor our model identified all instances of ACS, our model provides additional evidence of high-risk groups that might be more likely to benefit from obtaining a CXR. This model does not exclude providers from ordering CXRs on all febrile patients, but may provide evidence that might expand the CXR usage to other high-risk groups for providers who currently utilize the NHLBI criteria in determining on which children to obtain a CXR.

As a retrospective chart review, our study is limited by the information abstracted from the patient chart. In particular, whether the patient had a fever relied on medical record documentation. However, chart review has been previously shown to identify patients with fever more accurately than using ICD-9-CM coding.[12] Although our study focused on a small subset of patients within the larger SCD population, we collected data from two academic medical centers, each with a comprehensive sickle cell center. Therefore, the results may be generalized to other sites with similar patient demographics. Additionally, our treatment of missing documentation for respiratory symptoms and history of ACS as negative may have minimized the effect of these variables in our model. Most importantly, these results have not been validated in another population and that would be required before any substantial change in clinical practice could be recommended.

CONCLUSION

Based on our multisite study, we providence evidence that patients with sickle cell disease presenting to the ED with a fever who also have shortness of breath, tachypnea, cough, rales, or chest pain should receive a chest x-ray to investigate for ACS. To further maximize the number of patients accurately diagnosed with acute chest syndrome, a white blood cell count greater than 18.75×10^9/L or a history of acute chest syndrome in a patient who does not have one of the above respiratory symptoms may be used to determine a high-risk group for acute chest syndrome. If validated, these data can help guide a provider when deciding if a patient with sickle cell disease presenting with fever needs a chest x-ray.

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