This form is for acute chest syndrome and follows to find standard elements in various definitions. This is the same as sickle cell related acute pain episodes data which are collecting standard variables for vaso-occlusive crisis diagnosis. All pain related diagnoses could follow the same format as above: Symptom, Vitals (including pulse oximetry), Physical Exam, Diagnostic Laboratory and Therapy. The elements with astericks (\*) are Core.

**\*Acute Chest Syndrome**

[ ]  Yes

[ ]  No

[ ]  Unknown

**If yes, indicate the following:**

Symptoms

1. Pleuric Chest Pain
[ ]  Yes
[ ]  No
[ ]  Unknown
2. Chest pain
[ ]  Yes
[ ]  No
[ ]  Unknown
3. Wheezing
[ ]  Yes
[ ]  No
[ ]  Unknown
4. Cough
[ ]  Yes
[ ]  No
[ ]  Unknown
5. Dyspnea
[ ]  Yes
[ ]  No
[ ]  Unknown

Vitals

1. \*Temperature (highest on day of diagnosis) \_\_\_\_\_\_\_\_
2. \*Heart rate (highest on day of diagnosis) \_\_\_\_\_\_\_\_
3. \*Respiratory rate \_\_\_\_\_\_\_
4. \* Systolic blood pressure on day of diagnosis \_\_\_\_\_\_\_
5. \*\*SpO2 (O2 saturation) in room air (FiO2 = 0.21) decreased by 2% or more from baseline
6. Oximetry
[ ]  Yes
[ ]  No
[ ]  Unknown
	1. \*O2 Saturation Value: \_\_\_\_\_\_\_\_
7. \*PaO2 < 60 mmHg [ ]  Yes [ ]  No [ ]  Unknown
	1. \*PaO2 Value: \_\_\_\_\_\_\_

Physical Exam

1. Rales on lung auscultation
[ ]  Yes
[ ]  No
[ ]  Unknown
2. Intercostal retractions
[ ]  Yes
[ ]  No
[ ]  Unknown
3. Nasal flaring or use of accessory muscles of respiration
[ ]  Yes
[ ]  No
[ ]  Unknown
4. Wheezing
[ ]  Yes
[ ]  No
[ ]  Unknown

Diagnostic

Laboratory

1. Leukocytosis
[ ]  Yes
[ ]  No
[ ]  Unknown
2. Decreased hemoglobin
[ ]  Yes
[ ]  No
[ ]  Unknown
3. Decreased platelet count
[ ]  Yes
[ ]  No
[ ]  Unknown

Imaging[[1]](#endnote-2)

1. Development of new infiltrate on chest x-ray and/or perfusion defect demonstrable on lung radioisotope scan
[ ]  Yes
[ ]  No
[ ]  Unknown
[ ]  Unable to perform due to pregnancy *(this question may not be applicable now but in
 previous definitions)*
2. A new pulmonary infiltrate involving at least one complete lung segment that is consistent with the presence of alveolar consolidation, but excluding atelectasis
[ ]  Yes
[ ]  No
[ ]  Unknown
[ ]  Unable to perform due to pregnancy
3. Pulmonary infiltrate
[ ]  Yes
[ ]  No
[ ]  Unknown
[ ]  Unable to perform due to pregnancy
4. Radiographic evidence of consolidation. A new segmental (involving at least one complete segment) radiographic pulmonary infiltrate

[ ]  Yes
[ ]  No
[ ]  Unknown
[ ]  Unable to perform due to pregnancy

Therapy

1. Transfusion
[ ]  Yes
[ ]  No
[ ]  Unknown
	1. If yes, type

 [ ]  Simple

 [ ]  Exchange

Severity

1. Admitted to Hospital
[ ]  Yes
[ ]  No
[ ]  Unknown
2. ICU
[ ]  Yes
[ ]  No
[ ]  Unknown
3. Mechanical Ventilation
[ ]  Yes
[ ]  No
[ ]  Unknown
	1. The length of time received mechanical ventilation
4. Respiratory support

Non-mechanical ventilatory support:
[ ]  Simple nasal cannula
[ ]  Face mask O2 (e.g. ventimask, non-rebreather)

Noninvasive mechanical ventilatory support:
[ ]  CPAP
[ ]  SiPAP
[ ]  BiPAP
[ ]  High flow nasal cannula (HFNC)

Invasive mechanical ventilatory support (delivered by ETT or trach):
[ ]  Conventional mechanical ventilation
[ ]  HFOV

1. If no mechanical ventilation…
[ ]  CPAP
[ ]  Nasal cannula oxygen
[ ]  Face mask oxygen

**Rapid Progression Acute Chest Syndrome Module**

1. Decreased oxygen saturation requiring at least 3 L of oxygen to maintain oxygen hemoglobin saturation at least 90% or intubation and medical ventilator within 24 hours of onset of respiratory symptoms.

[ ]  Yes
[ ]  No
[ ]  Unknown

1. Worsening anemia was arbitrarily defined as a decrease in hemoglobin by >= 2 g/dL from baseline.

[ ]  Yes
[ ]  No
[ ]  Unknown

1. Thrombocytopenia (or decrease in platelet count) as defined as a platelet count 150,000/mcl or a 50% decrease from baseline.

[ ]  Yes
[ ]  No
[ ]  Unknown

1. Multiorgan failure (defined as dysfunction of two or more organs by the following criteria [10]: respiratory failure (respiratory distress and at least 3 L of oxygen to maintain oxygen hemoglobin saturation at least 90%), acute renal insufficiency (an increase in the serum creatinine concentration of 50% from baseline; or oliguria of <0.5 mL/kg/hr for more than 6 hr) [17], altered mental status, other neurologic symptoms (new focal neurologic deficit, seizure, confusion, blurred vision),he patic insufficiency (at least two of the follow features: alanine aminotransferase >70 U/L, total bilirubin >2 times upper limit of normal, direct bilirubin > 2 times the upper limit of normal), and prothrombin time prolonged by more than 3 sec [10]. Aspartate aminotransferase was not included because this may be elevated in the setting of hemolysis (Chaturvedi, S; Ghafuri, DL; Glassberg, J; Kassim, AA; Rodeghier, M; DeBaun, MR. Rapidly progressive acute chest syndrome in individuals with sickle cell anemia: a distinct acute chest syndrome phenotype. Am. J. Hematol., 2016 vol. 91(12) pp. 1185-1190)

[ ]  Yes
[ ]  No
[ ]  Unknown

## General Instructions

This form is for acute chest syndrome and follows to find standard elements in various definitions. This is the same as sickle cell related acute pain episodes data which are collecting standard variables for vaso-occlusive crisis diagnosis. All pain related diagnoses could follow the same format as above: Symptom, Vitals (including pulse oximetry), Physical Exam, Diagnostic Laboratory and Therapy.

Pain FDA definition follows: Pain that lasts 4 hours; No explanation other than vaso-occlusive; and, required therapy with parental opioids or ketorolac in a medical setting. “Sickle cell-related pain crises were defined as acute episodes of pain, with no medically determined cause other than a vaso-occlusive event, that resulted in a medical facility visit and treatment with oral or parenteral narcotic agents or with a parenteral nonsteroidal anti-inflammatory drug.”

Paid longer than 4 hours
[ ]  Yes
[ ]  No

[ ]  Unknown

No other medically determined cause
[ ]  Yes
[ ]  No
[ ]  Unknown

Medical facility visit

[ ]  Yes

[ ]  No

[ ]  Unknown

Treatment with oral or parenteral narcotic agents or with a parenteral non-steriodal antiinflammatory drug.

[ ]  Yes

[ ]  No

[ ]  Unknown

Exploratory: If female: Dysmenorrhea

[ ]  Yes

[ ]  No

[ ]  Unknown

Pediatric consideration:

Bronciolitis definition in patient 2-3 years of age is the same as adult with Acute Chest Syndrome which is life-threatening event.

Suggestion if there is more than one event:

1. Pain + ACS = ACS
2. Pain alone = VOC

**OR**

1. ACS and VOC considered as one event and not separated at all.

## Specific Instructions

Please see the Data Dictionary for definitions for each of the data elements included in this CRF Module.

**References**

1. Ballas SK, Lieff S, Benjamin LJ, Dampier CD, Heeney MM, Hoppe C, Johnson CS, Rogers ZR, Smith-Whitley K, Wang WC, Telen MJ; Investigators, Comprehensive Sickle Cell Centers. Definitions of the phenotypic manifestations of sickle cell disease. Am J Hematol. 2010 Jan;85(1):6-13. doi: 10.1002/ajh.21550. PMID: 19902523; PMCID: PMC5046828.

2. Castro O, Brambilla DJ, Thorington B, et al. The acute chest syndrome in sickle cell disease: Incidence and risk factors. The Cooperative Study of Sickle Cell Disease. Blood. 1994;84:643–649.

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6. Stuart MJ, Setty BN. Sickle cell acute chest syndrome: Pathogenesis and rationale for treatment. Blood 1999;94:1555–1560.

7. Davies SC, Luce PJ, Win AA, et al. Acute chest syndrome in sickle-cell disease. Lancet 1984;1:36–38.Bernard GR, Artigas A, Brigham KL, et al. The American-European Consensus Conference on ARDS. Definitions, mechanisms, relevant outcomes, and clinical trial coordination. Am J Respir Crit Care Med 1994;149:818–824.

Johnson CS. The acute chest syndrome. Hematol Oncol Clin North Am 2005;19:857–879.

Rackoff WR, Kunkel N, Silber JH, et al. Pulse oximetry and factors associated with hemoglobin oxygen desaturation in children with sickle cell disease. Blood 1993;81:3422–3427.

Vichinsky E, Williams R, Das M, et al. Pulmonary fat embolism: a distinct cause of severe acute chest syndrome in sickle cell anemia. Blood 1994;83:3107–3112.

1. Consideration of pregnant women needs to be made. [↑](#endnote-ref-2)