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**

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

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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)