Guideline Evidence

Guideline Topic: Management of Acute Chest Syndrome in Pediatric Sickle Cell Patients
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Date of Creation: 4/27/18 Sugg Update: 2020
Search Criteria: Acute chest syndrome, sickle cell, pediatrics, sickle cell emergencies
Databases: Ovid, PubMed.
Key Guidelines (Dates) Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.
Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014

#	Recommendation	Source	Classification	Level of Evidence
1	Evaluate people with SCD who develop acute onset of lower respiratory tract disease signs and or symptoms (cough,shortness of breath, tachypnea, retractions, or wheezing) with or without fever for ACS. This should include a chest x-ray and measurement of oxygen saturation by pulse oximetry.	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services	I	Level B
2	Treat people with SCD who have ACS with an intravenous cephalosporin, an oral macrolide antibiotic, and supplemental oxygen (to maintain oxygen saturation of greater than 95 percent).	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	I	Level B
3	All patients with ACS should be given prompt and adequate pain management.	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services	I	Level B
4	In people with HbSC disease or HbS β+-thalassemia with ACS, decisions about transfusion should be made in consultation with an SCD expert.	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	I	level C
5	Pulmonary embolism, fluid overload, opiate narcosis and hypoventilation may cause or trigger ACS and should be considered when diagnosis is made.	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services	I	Level B
6	Encourage use of incentive spirometry while awake.	Report 2014 US Department of Health and Human Services Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	I	Level B
7	Bronchodilators should be used if theres are clinical features suggestive of history of asthma or evidence of acute bronchospasm.	Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	II	Level B
8	In people with SCA, give simple blood transfusion (10 mL/kg red blood cells) to improve oxygen carrying capacity to people with symptomatic ACS whose hemoglobin concentration is >1.0 g/dL below baseline. If baseline hemoglobinis 9 g/dL or higher, simple blood transfusion may not be required.	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	II	Level B
9	In all persons with SCD, perform urgent exchange transfusion—with consultation from hematology, critical care, and/or apheresis specialists—when there is rapid progression of ACS as manifested by oxygen saturation below 90 percent despite supplemental oxygen, increasing respiratory distress, progressive pulmonary infiltrates, and/or decline in hemoglobin concentration despite simple transfusion.	Evidence Based Medicine in Sickle Cell Disease, Expert Panel Report 2014 US Department of Health and Human Services Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	II	Level B
10	Blood should be sickle-negative and fully matched.	Guideline on the management of acute chest syndrome in sickle cell disease. BJH guideline. March 2015.	I	Level A
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CPQE Guideline Evidence, cont.

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