

Sickle Cell Disease and Hereditary Hemolytic Anemias

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REFERENCES

1. <http://www.who.int/bulletin/volumes/86/6/06-036673/en/>. (Modell B, Darlison M: Bulletin of the World Health Organization. Global Epidemiology of Haemoglobin Disorders and Derived Service Indicators.) Accessed July 7, 2018.
2. Weatherall D: The inherited disorders of haemoglobin: an increasingly neglected global health burden. *Indian J Med Res* 134: 493, 2011. [PMID: 22089613]
3. Ware RE, de Montalembert M, Tshilolo L, Abboud MR: Sickle cell disease. *Lancet* 390: 311, 2017. [PMID: 2815939]
4. <http://www.cdc.gov/NCBDDD/sicklecell/data.html>. (Centers for Disease Control and Prevention: Sickle cell disease, data and statistics.) Accessed February 7, 2018.
5. Sheth S, Licursi M, Bhatia M: Sickle cell disease: time for a closer look at treatment options? *Br J Haematol* 162: 455, 2013. [PMID: 23772687]
6. Quinn CT, Rogers ZR, McCavit TL, et al: Improved survival of children and adolescents with sickle cell disease. *Blood* 115: 3447, 2010. [PMID: 20194891]
7. Kassim AA, DeBauw MR: Sickle cell disease, vasculopathy, and therapeutics. *Annu Rev Med* 64: 451, 2013. [PMID: 23190149]
8. Rees DC, Williams TN, Gladwin MT: Sickle-cell disease. *Lancet* 376: 2018, 2010. [PMID: 21131035]
9. Hsia CC: Respiratory function of hemoglobin. *N Engl J Med* 338: 239, 1998. [PMID: 9435331]
10. Stuart MJ, Nagel RL: Sickle-cell disease. *Lancet* 364: 1343, 2004. [PMID: 15474138]
11. Simon E, Long B, Koyfman A: Emergency medicine management of sickle cell disease complications: an evidence-based update. *J Emerg Med* 51: 370, 2016. [PMID: 27553919]
12. Mousa SA, Al Momen A, Al Sayegh F, et al: Management of painful vaso-occlusive crisis of sickle-cell anemia: consensus opinion. *Clin Appl Thromb Hemost* 16: 365, 2010. [PMID: 20530056]
13. https://www.ncbi.nlm.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf (National Institutes of Health, National Heart, Lung, and Blood Institute: Evidence-based management of sickle cell disease. Expert Panel Report, 2014.) Accessed July 7, 2018.
14. Okomo U, Meremikwu MM: Replacing fluids to treat acute episodes of pain in people with sickle cell disease. *Cochrane Database Syst Rev* 7: CD005406, 2017. [PMID: 28759112]
15. Okomo U, Meremikwu MM: Fluid replacement therapy for acute episodes of pain in people with sickle cell disease. *Cochrane Database Syst Rev* 6: CD005406, 2012. [PMID: 22696351]
16. Han J: Use of antiinflammatory analgesics in sickle cell disease. *J Clin Pharm Ther* 42: 656, 2017. [PMID: 28695614]
17. Hagedorn JM, Monico EC: Ketamine infusion for pain control in acute pediatric sickle cell painful crises. *Pediatr Emerg Care* 2016 Nov 29. [Epub ahead of print] [PMID: 27902670]
18. Palm N, Floroff C, Hassig TB, Boylan A, Kanter J: Low-dose ketamine infusion for adjunct management during vaso-occlusive episodes in adults with sickle cell disease: a case series. *J Pain Palliat Care Pharmacother* 23: 1, 2018. [PMID: 29791238]
19. Vichinsky EP, Neumayr LD, Earles AN, et al: Causes and outcomes of the acute chest syndrome in sickle cell disease. National Acute Chest Syndrome Study Group. *N Engl J Med* 342: 1855, 2000. [PMID: 10861320] [Erratum in: *N Engl J Med* 343: 824, 2000.]
20. Paul RN, Castro OL, Aggarwal A, Oneal PA: Acute chest syndrome: sickle cell disease. *Eur J Haematol* 87: 191, 2011. [PMID: 21615795]
21. Desai PC, Ataga KI: The acute chest syndrome of sickle cell disease. *Expert Opin Pharmacother* 14: 991, 2013. [PMID: 23534969]
22. Miller AC, Gladwin MT: Pulmonary complications of sickle cell disease. *Am J Respir Crit Care Med* 185: 1154, 2012. [PMID: 22447965]
23. Stuart MJ, Setty BN: Acute chest syndrome of sickle cell disease: new light on an old problem. *Curr Opin Hematol* 8: 111, 2001. [PMID: 11224686]
24. Styles LA, Aarsman AJ, Vichinsky EP, Kuypers FA: Secretory phospholipase A2 predicts impending acute chest syndrome in sickle cell disease. *Blood* 96: 3276, 2000. [PMID: 11050014]
25. Mekontso Dessap A, Deux JF, Abidi N, et al: Pulmonary artery thrombosis during acute chest syndrome in sickle cell disease. *Am J Respir Crit Care Med* 184: 1022, 2011. [PMID: 21836136]
26. Cecchini J, Fartoukh M: Sickle cell disease in the ICU. *Curr Opin Crit Care* 21: 569, 2015. [PMID: 26539931]
27. Howard J, Hart N, Roberts-Harewood M, Cummins M, Awogbade M, Davis B; BCSH Committee: Guideline on the management of acute chest syndrome in sickle cell disease. *Br J Haematol* 169: 492, 2015. [PMID: 25824256]
28. Knight-Madden JM, Hambleton IR: Inhaled bronchodilators for acute chest syndrome in people with sickle cell disease. *Cochrane Database Syst Rev* 7: CD003733, 2012. [PMID: 22786487]
29. Alhashimi D, Fedorowicz Z, Alhashimi F, Dastgiri S: Blood transfusions for treating acute chest syndrome in people with sickle cell disease. *Cochrane Database Syst Rev* 1: CD007843, 2010. [PMID: 20091653]
30. Bernini JC, Rogers ZR, Sandler ES, et al: Beneficial effect of intravenous dexamethasone in children with mild to moderately severe acute chest syndrome complicating sickle cell disease. *Blood* 92: 3082, 1998. [PMID: 9787142]
31. Griffin TC, McIntire D, Buchanan GR: High-dose intravenous methylprednisolone therapy for pain in children and adolescents with sickle cell disease. *N Engl J Med* 330: 733, 1994. [PMID: 8107739]
32. Ogunlesi F, Heeney MM, Koumbourlis AC: Systemic corticosteroids in acute chest syndrome: friend or foe? *Pediatr Respir Rev* 15: 24, 2014. [PMID: 24268617]
33. Vichinsky EP, Neumayr LD, Earles AN, et al: Causes and outcomes of the acute chest syndrome in sickle cell disease. *N Engl J Med* 342: 1855, 2000. [PMID: 10861320]
34. De Gracia-Nieto AE, Samper AO, Rojas-Cruz C, et al: Genitourinary manifestations of sickle cell disease. *Arch Esp Urol* 64: 597, 2011. [PMID: 21965257]
35. López Revuelta K, Ricard Andrés MP: Kidney abnormalities in sickle cell disease. *Nefrologia* 31: 591, 2011. [PMID: 21959727]
36. Olujohungbe A, Burnett AL: How I manage priapism due to sickle cell disease. *Br J Haematol* 160: 754, 2013. [PMID: 23293942]
37. Pearson HA, Gallagher D, Chilcote R, et al: Developmental pattern of splenic dysfunction in sickle cell disorders. *Pediatrics* 76: 392, 1985. [PMID: 2412200]
38. Brousse V, Elie C, Benkerrou M, et al: Acute splenic sequestration crisis in sickle cell disease: cohort study of 190 paediatric patients. *Br J Haematol* 156: 643, 2012. [PMID: 22224796]
39. Jenkins T: Sickle cell anemia in the pediatric intensive care unit: novel approaches for managing life-threatening complications. *AACN Clin Issues* 13: 154, 2002. [PMID: 12011590]
40. Olabode JO, Shokunbi WA: Types of crises in sickle cell disease patients presenting at the haematology day care unit (HDCU), University College Hospital (UCH), Ibadan. *West Afr J Med* 25: 284, 2006. [PMID: 17402517]
41. Tsitsikas DA, Gallinella G, Patel S, et al: Bone marrow necrosis and fat embolism syndrome in sickle cell disease: increased susceptibility of patients with non-SS genotypes and a possible association with human parvovirus B19 infection. *Blood Rev* 28: 23, 2014. [PMID: 24468004]
42. Strouse JJ, Lanzkron S, Urrutia V: The epidemiology, evaluation and treatment of stroke in adults with sickle cell disease. *Expert Rev Hematol* 4: 597, 2011. [PMID: 22077524]
43. Switzer JA, Hess DC, Nichols FT, Adams RJ: Pathophysiology and treatment of stroke in sickle disease: present and future. *Lancet Neurol* 5: 501, 2006. [PMID: 16793922]
44. Gueguen A, Mahevas M, Nzouakou R, et al: Sickle-cell disease stroke throughout life: a retrospective study in an adult referral center. *Am J Hematol* 89: 267, 2014. [PMID: 24779035]
45. Kugler S, Anderson B, Cross D, et al: Abnormal cranial magnetic resonance imaging scans in sickle-cell disease. Neurological correlates and clinical implications. *Arch Neurol* 50: 629, 1993. [PMID: 8503800]
46. Strouse JJ, Cox CS, Melhem ER, et al: Inverse correlation between cerebral blood flow measured by continuous arterial spin-labeling (CASL) MRI and neurocognitive function in children with sickle cell anemia (SCA). *Blood* 108: 379, 2006. [PMID: 16537809]
47. Venkataraman A, Adams RJ: Neurologic complications of sickle cell disease. *Handb Clin Neurol* 120: 1015, 2014. [PMID: 24365368]
48. Ohene-Frempong K, Weiner SJ, Sleeper LA, et al: Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood* 91: 288, 1998. [PMID: 9414296]
49. Adams RJ, McKie VC, Hsu L, et al: Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. *N Engl J Med* 339: 5, 1998. [PMID: 9647873]
50. Powers WJ, Rabinstein AA, Ackerson T, et al: 2018 guidelines for the early management of patients with acute ischemic stroke: a guideline for healthcare professionals from the American Heart Association/American Stroke Association. *Stroke* 49: e46, 2018. [PMID: 29367334]
51. Glassberg J: Evidence-based management of sickle cell disease in the emergency department. *Emerg Med Pract* 13: 1, 2011. [PMID: 22164362]
52. Ramakrishnan M, Moisi JC, Klugman KP, et al: Increased risk of invasive bacterial infections in African people with sickle-cell disease: a systematic review and meta-analysis. *Lancet Infect Dis* 10: 329, 2010. [PMID: 20417415]
53. Hirst C, Owusu-Ofori S: Prophylactic antibiotics for preventing pneumococcal infection in children with sickle cell disease. *Cochrane Database Syst Rev* 9: CD003437, 2012. [PMID: 22972063]
54. Voskaridou E, Christoulas D, Terpos E: Sickle-cell disease and the heart: review of the current literature. *Br J Haematol* 157: 664, 2012. [PMID: 22530942]
55. Gerry J, Bulkley B, Hutchins G: Clinicopathologic analysis of cardiac dysfunction in 52 patients with sickle cell anemia. *Am J Cardiol* 42: 211, 1978. [PMID: 150786]
56. Barrett OJ Jr, Saunders DE, McFarland D, et al: Myocardial infarction in sickle cell anemia. *Am J Hematol* 16: 139, 1984. [PMID: 6230006]
57. Manci EA, Culberson DE, Yang YM, et al: Causes of death in sickle cell disease: an autopsy study. *Br J Haematol* 123: 359, 2003. [PMID: 14531921]
58. Aslam AK, Rodriguez C, Aslam AF, et al: Cardiac troponin I in sickle cell crisis. *Int J Cardiol* 133: 138, 2009. [PMID: 18178271]
59. Pavlu J, Ahmed RE, O'Regan DP, et al: Myocardial infarction in sickle-cell disease. *Lancet* 369: 246, 2007. [PMID: 17240292]
60. Sherman SC, Sule HP: Acute myocardial infarction in a young man with sickle cell disease. *J Emerg Med* 27: 31, 2004. [PMID: 15219301]
61. Novelli EM, Huynh C, Gladwin MT, Moore CG, Ragni MV: Pulmonary embolism in sickle cell disease: a case-control study. *J Thromb Haemost* 10: 760, 2012. [PMID: 22417249]

62. Buckner TW, Key NS: Venous thrombosis in blacks. *Circulation* 125: 837, 2012. [PMID: 22331921]
63. Naik RP, Streiff MB, Lanzkron S: Sickle cell disease and venous thromboembolism: what the anticoagulation expert needs to know. *J Thromb Thrombolysis* 35: 352, 2013. [PMID: 23435703]
64. Lim MY, Ataga KI, Key NS: Hemostatic abnormalities in sickle cell disease. *Curr Opin Hematol* 20: 472, 2013. [PMID: 23817169]
65. Novelli EM, Huynh C, Gladwin MT, et al: Pulmonary embolism in sickle cell disease: a case-control study. *J Thromb Haemost* 10: 760, 2012. [PMID: 22417249]
66. Shah N, Thornburg C, Telen MJ, Ortel TL: Characterization of the hypercoagulable state in patients with sickle cell disease. *Thromb Res* 130: e241, 2012. [PMID: 22959127]
67. Delaney KM, Axelrod KC, Buscetta A, et al: Leg ulcers in sickle cell disease: current patterns and practices. *Hemoglobin* 37: 325, 2013. [PMID: 23600469]
68. Martí-Carvajal AJ, Knight-Madden JM, Martínez-Zapata MJ: Interventions for treating leg ulcers in people with sickle cell disease. *Cochrane Database Syst Rev* 11: CD008394, 2012. [PMID: 23152256]
69. Hassell KL, Eckman JR, Lane PA: Acute multiorgan failure syndrome: a potentially catastrophic complication of severe sickle cell pain episodes. *Am J Med* 96: 155, 1994. [PMID: 8109600]
70. Venkata Sasidhar M, Tripathy AK, Viswanathan K, et al: Thrombotic thrombocytopenic purpura and multiorgan system failure in a child with sickle cell-hemoglobin C disease. *Clin Pediatr (Phila)* 49: 992, 2010. [PMID: 19525485]
71. Shelat SG: Thrombotic thrombocytopenic purpura and sickle cell crisis. *Clin Appl Thromb Hemost* 16: 224, 2010. [PMID: 18840631]
72. Tsaras G, Owusu-Ansah A, Boateng FO, Amoateng-Adjepong Y: Complications associated with sickle cell trait: a brief narrative review. *Am J Med* 122: 507, 2009. [PMID: 19393983]
73. Kotila TR: Sickle cell trait: a benign state? *Acta Haematol* 136: 147, 2016. [PMID: 27423233]
74. Piel FB, Howes RE, Patil AP, et al: The distribution of haemoglobin C and its prevalence in newborns in Africa. *Sci Rep* 3: 1671, 2013. [PMID: 23591685]
75. Dalia S, Zhang L: Homozygous hemoglobin C disease. *Blood* 122: 1694, 2013. [PMID: 24137818]
76. Lim JI: Ophthalmic manifestations of sickle cell disease: update of the latest findings. *Curr Opin Ophthalmol* 23: 533, 2012. [PMID: 23047170]
77. Pereira SA, Brener S, Cardoso CS, Proietti AB: Sickle cell disease: quality of life in patients with hemoglobin SS and SC disorders. *Rev Bras Hematol Hemoter* 35: 325, 2013. [PMID: 24255615]
78. El-Hazmi MA, Al-Hazmi AM, Warsy AS: Sickle cell disease in Middle East Arab countries. *Indian J Med Res* 134: 597, 2011. [PMID: 22199098]
79. Peters M, Heijboer H, Smiers F, Giordano PC: Diagnosis and management of thalassae mia. *BMJ* 344: e228, 2012. [PMID: 22277544]
80. Forget BG, Bunn HF: Classification of the disorders of hemoglobin. *Cold Spring Harb Perspect Med* 3: a011684, 2013. [PMID: 23378597]
81. Taher AT, Weatherall DJ, Cappellini MD: Thalassaemia. *Lancet* 391: 155, 2018. [PMID: 28774421]
82. Sankaran VG, Nathan DG: Thalassemia: an overview of 50 years of clinical research. *Hematol Oncol Clin North Am* 24: 1005, 2010. [PMID: 21075277]
83. Taher AT, Musallam KM, Cappellini MD, Weatherall DJ: Optimal management of β thalassaemia intermedia. *Br J Haematol* 152: 512, 2011. [PMID: 21250971]
84. Maakarion JE, Cappellini MD, Taher AT: An update on thalassemia intermedia. *J Med Liban* 61: 175, 2013. [PMID: 24422369]
85. Borgna-Pignatti C, Gamberini MR: Complications of thalassemia major and their treatment. *Expert Rev Hematol* 4: 353, 2011. [PMID: 21668399]
86. Chou ST, Liem RI, Thompson AA: Challenges of alloimmunization in patients with haemoglobinopathies. *Br J Haematol* 159: 394, 2012. [PMID: 23034087]
87. Musallam KM, Angastiniotis M, Eleftheriou A, Porter JB: Cross-talk between available guidelines for the management of patients with beta-thalassemia major. *Acta Haematol* 130: 64, 2013. [PMID: 23485589]
88. Fisher SA, Brunskill SJ, Doree C, et al: Desferrioxamine mesylate for managing transfusion iron overload in people with transfusion-dependent thalassemia. *Cochrane Database Syst Rev* 8: CD004450, 2013. [PMID: 23963793]
89. Fisher SA, Brunskill SJ, Doree C, et al: Oral deferasirox for iron chelation in people with thalassemia. *Cochrane Database Syst Rev* 8: CD004839, 2013. [PMID: 23966105]
90. Meerpohl JJ, Antes G, Rucker G, et al: Deferasirox for managing iron overload in people with thalassemia. *Cochrane Database Syst Rev* 2: CD007476, 2012. [PMID: 22336831]
91. Luzzatto L, Nannelli C, Notarso R: Glucose-6-phosphate dehydrogenase deficiency. *Hematol Oncol Clin N Am* 30: 373, 2016. [PMID: 27040960]
92. Nkhoma ET, Poole C, Vannappagari V, et al: The global prevalence of glucose-6-phosphate dehydrogenase deficiency: a systematic review and meta-analysis. *Blood Cells Mol Dis* 42: 267, 2009. [PMID: 19233695]
93. Hagag AA, Badraia IM, Elfarargy MS, et al: Study of glucose-6-phosphate dehydrogenase deficiency: 5 years retrospective Egyptian study. *Endocr Metab Immune Disord Drug Targets* 18: 155, 2018. [PMID: 28982343]
94. Youngster I, Arcavi L, Schechmaster R, et al: Medications and glucose-6-phosphate dehydrogenase deficiency: an evidence-based review. *Drug Saf* 33: 713, 2010. [PMID: 20701405]
95. Belfield KD, Tichy EM: Review and drug therapy implications of glucose-6-phosphate dehydrogenase deficiency. *Am J Health Syst Pharm* 75: 97, 2018. [PMID: 29305344]
96. Gulbis B, Eleftheriou A, Angastiniotis M, et al: Epidemiology of rare anemias in Europe. *Adv Exp Med Biol* 686: 375, 2010. [PMID: 20824457]
97. Da Costa L, Galimand J, Fenneteau O, Mohandas N: Hereditary spherocytosis, elliptocytosis, and other red cell membrane disorders. *Blood Rev* 27: 167, 2013. [PMID: 23664421]
98. Gallagher PG: Abnormalities of the erythrocyte membrane. *Pediatr Clin North Am* 60: 1349, 2013. [PMID: 24237975]
99. Perrotta S, Gallagher PG, Mohandas N: Hereditary spherocytosis. *Lancet* 372: 1411, 2008. [PMID: 189404675]
100. Barcellini W, Bianchi P, Fermo E, et al: Hereditary red cell membrane defects: diagnostic and clinical aspects. *Blood Transfus* 9: 274, 2011. [PMID: 21251470]
101. Bolton-Maggs PH, Stevens RF, Dodd NJ, et al: Guidelines for the diagnosis and management of hereditary spherocytosis. *Br J Haematol* 126: 455, 2004. [PMID: 15287938]
102. Casale M, Perrotta S: Splenectomy for hereditary spherocytosis: complete, partial, or not at all? *Expert Rev Hematol* 4: 627, 2011. [PMID: 22077527]