

## References

### References:

1. **John P. Greer**, John Foerster, John N. Lukens. Wintrobe's Clinical Hematology (2003). 11<sup>th</sup> edition. New York. Lippincott Williams & Wilkins Publishers.
2. **A. Victor Hoffbrand**, Daniel Catovsky, Edward G.D. (2005) Postgraduate Haematology. 5<sup>th</sup> edition. Oxford. Blackwell Publishing Ltd.
3. **A. V. Hoffbrand**, P.A. H. Moss, J.E. Pettit. (2006). 5<sup>th</sup> edition. Essential Haematology Victoria, Australia. Blackwell Publishing Ltd.
4. **Barbara J. Bain**. Blood Cells A Practical Guide. 4th edition. Oxford, UK. Blackwell Publishing Ltd. (2006).
5. **Betke K**, Marti HR, Schlicht L. (1999). Estimation of small percentage of fetal Hemoglobin. Nature.
6. **John P. Greer**, John Foerster, John N. Lukens. (2003). Wintrobe's Clinical Hematology. 11th edition. New York. Lippincott Williams & Wilkins Publishers.
7. **Reinhold Munker**, Erhard Hiller, Jonathan Glass, Ronald Paquette. (2007). Modern Hematology. 2nd edition. New Jersey. United States of America. Humana Press Inc.
8. **Barbara J. Bain**. Haemoglobinopathies diagnosis. (2006). 2nd edition Oxford Blackwell Science Ltd.
9. **Ronald Hoffman**, Edward Benz, Sanford Shattil, Bruce Furie, Harvey Cohen. (2008). Hematology: Basic Principles and Practice. 5th edition. New York. Churchill Livingstone.
10. **Vinay Kumar**, Ramzi S.Cotran, Stanley L.Robbins. (2003) Basic pathology.7 the edition. International edition china Saunders.
11. **Bernadette F. Rodak**. Diagnostic Hematology. (1995)1st edition. USA. W.B. Saunders Company.
12. **Mary Louise Turgeon**. Clinical Laboratory Science (2007) 5the China Mosby.
13. **Robert S.Hillman**, Kenneth A. Ault, Henry M.Rinder.Hematology in Clinical Practice (2005) 4th International edition Mc Graw Hill.
14. **Fifty nine World Health Organization assembly Provisional agenda** item 11.4 (A59/9). (2006). *Sickle cell anemia. World Health Organization*. Geneva, Switzerland. Report by the Secretariat.

## References

15. **Teebi. Ahmad S**, Farag. Talaat. I. (1999). *Genetic Disorders among Arab Populations*. 1<sup>st</sup> edition. London. Oxford University Press.
16. **Samir K Ballas, Franca B Barton, Myron A Waclawiw**, *Health and Quality of Life Outcomes* 2006, **4**:59doi:10.1186/1477-7525-4-59
17. **F. Kern William**. (2002). *PDQ Hematology*. 1<sup>st</sup> edition. Oklahoma City. B.C. Decker.
18. **Hoffbrand. A.V**, P.A. H. Moss, J.E. Pettit. (2008). 6<sup>th</sup> edition. *Essential Haematology* Victoria, Australia. Blackwell Publishing Ltd.
19. **Hillman. Robert S**, Ault .Kenneth A., Rinder .Henry .M. (2005). *Hematology in Clinical Practice*. 4<sup>th</sup> edition. United States of America. McGraw-Hill Companies.
20. **Mears JG**, Beldjord C, Benabadji M, et al. (1981). *The sickle gene polymorphism in North Africa*. Blood. 58(3):599-601
21. **Frank Firkin**, Colin Chesterman, David Pennington and Bryan Rush. (1989). *De Gruchy Clinical Haematology in Medical Practice*. 5th edition. London. Blackwell Publishing Ltd
22. **Barbara A.Brown**.Hematology: Principles and Procedures (1993) 6<sup>th</sup> U.S.A Lea & Febiger.
23. **Douglas C.Tkachuk**, J .V. Hirschman, James R.McArthur.atlas of Clinical Hematology. (2002) China W.B.Sounders Company.
24. **Dinah V.Parums**, *Essential Clinical Pathology* (1996). 1st Australia .Blackwell Science Ltd.
25. **Frank Firkin**, Colin Chesterman, David Pennington & Bryan Rush. (2002). 5th India Blackwell Science Ltd.
26. **World Health Organization**. (2000). turning the tide of malnutrition: Responding to the challenge of the 21st century. Geneva, Switzerland.
27. **Joseph J. Mazza** .Manual of Clinical Hematology. (2002) 3 rd U.S.A Lippincott. Williams & Wilkins.
28. **Alvin H. Schmaier**, Lilli M. Peteruzzelli. *Hematology for Medical Student* (2003). 1st edition. U.S.A Lippincott Williams & Wilkins.
29. **Drew Provan**, Gribben. John. G. (2005). *Molecular Hematology*.2nd edition. United States of America. Blackwell Publishing Ltd.

## References

30. **Weatherall DJ**, Clegg JB, Higgs DR et al. (2001) the Haemoglobinopathies. In: Scriver CR, Beaudet AL, Sly WS et al., Eds. *The Metabolic Basis of Inherited Disease*, 8<sup>th</sup> edn. New York: McGraw-Hill.
31. **E. Anne Stiene Martin**. Cherty. A. Latspeich. Steininger. Jonhn A. Koepke. *Clinical Hematology Principle, procedure, Correlation*. (1998). 2nd edition. Lippincott.
32. **Wong SC**, Huisman TH. (1972). Further evidence for non-linkage of the haemoglobin structural loci in man. *Clin Chim Acta*. Elsevier. Netherlands. 38(2):473-4
33. **Greer. John P**, John Foerster, Lukens. John N. (2003). *Wintrobe's Clinical Hematology*. 11th edition. New York. Lippincott Williams & Wilkins Publishers.
34. **Livingstone FB**. (1967). Abnormal haemoglobin in human populations. A summary and interpretation. Chicago: Aldine.
35. **J Pagnier**, J G Mears, O Dunda-Belkhodja, et al. (1984). Evidence for the multicentric origin of the sickle cell hemoglobin gene in Africa. *Proceedings of the National Academy of Sciences U S A*. 81(6): 1771–1773.
36. **SE Antonarakis**, CD Boehm, GR Sergent, et al. (1984). Origin of the  $\beta$  S-globin gene in blacks: the contribution of the recurrent mutation or gene conversion or both. *Proceedings of the National Academy of Sciences U S A*. 81(3): 853–856.
37. **Wainscoat JS**, Bell JI, Thein SL, et al. (1983). Multiple origins of the sickle mutation: evidence from beta S globin gene cluster polymorphisms. *Molecular biology & medicine*. ENGLAND. 1(2):191-7.
38. **Kan YW**, Dozy A M. (1978). Polymorphism of DNA sequence adjacent to the human  $\beta$ -globin structural gene; its relation to the sickle mutation. *Proceedings of the National Academy of Sciences U S A*. 75(11):5631-5.
39. **Kan YW**, Dozy A M. (1980). Evolution of the hemoglobin S and C genes in world populations. *Science*. (4454):388-91.
40. **Flatz G**. (1967). Hemoglobin E: distribution and population dynamics. *Humangenetik*. Springer Verlag. Germany. 3(3):189-234.
41. **Roth EF**, Friedman M, Ueda Y, et al. (1978). Sickling rates of human AS red cells infected in vitro with *Plasmodium falciparum* malaria. United States. *Science*. 202(4368):650-2.

## References

42. **Sick K**, Beale D, Irvine D, et al. (1967). Haemoglobin G Copenhagen and haemoglobin J Cambridge. Two new beta-chain variants of haemoglobin A. *Biochim Biophys Acta*. Elsevier Pub. Co. Netherlands. 140(2):231-42.
43. **Iheanyi E Okpala**. (2004). *Practical Management of Haemoglobinopathies*. 1st edition. Oxford. Blackwell Publishing Ltd.
44. **Marshall A**. Lichtman, Ernest Beutler, Thomas J. Kipps, Uri Seligsohn, Kenneth Kaushansky, Josef T. Prchal. (2005) *Williams Hematology*. 7th edition. United States of America. McGraw-Hill Professional.
45. **John P**. Greer, John Foerster, John N. Lukens. *Wintrobe's Clinical Hematology*. (2003). 11th edition. New York. Lippincott Williams & Wilkins Publishers.
46. **Griffith P**. Rodgers & Neal S. Young. *BETHESDA Handbook of Clinical Hematology*. (2005). 1st edition U.S.A Lippincott Williams & Wilkins.
47. **Dunston T**, Rowland R, Huntsman RG, Yawson GI. (1972). Sickle-cell haemoglobin C disease and sickle-cell beta thalassaemia in white South Africans. *South African medical journal*. South African Medical Association. South Africa. 46 (39):1423-6.
48. **Emmanuel C**. Besa, Patricia M. Catalano, Jeffrey A. Kanta, Leigh C. Jefferies. *NMS Hematology* (1992) 1st edition .Egyptian Edition. MASS publishing Co.
49. **Nagel RL**. (2004). Beta-globin-gene haplotype, mitochondrial DNA, the Y-chromosome: Their impact on the genetic epidemiology of the major structural Haemoglobinopathies. *Cellular and molecular biology*. C.M.B. Association. France. 50(1):5-21.
50. **Ronald Fisher**, JBS Haldane, & Sewall Wright. (2004). *Random Mutations and Evolutionary Change*. USA. Hildegard Adler.
51. **James V. Neel**. (1949). *The Inheritance of Sickle Cell Anaemia*. United States. Science. 110(2846):64-66.
52. **Bertles JF**, Rabinowitz R, Dobler J. (1970). Hemoglobin interaction: modification of solid phase composition in the sickling phenomenon. *United States. Science*. 169 (943):375-7.
53. **Hoffbrand. A. Victor**, Catovsky Daniel, G.D Edward. (2005) *Postgraduate Haematology*. 5th edition. Oxford. Blackwell Publishing Ltd.
54. **V. Hoffbrand**, P.A. H. Moss, J.E. Pettit. *Essential Haematology* (2006). 5th edition Victoria, Australia. Blackwell Publishing Ltd.

## References

55. **N.C.Hughes Jones, S.N.Wickramasinghe & C.Hatton.** Lecture notes on Hematology. (200)7th edition. Australia. Blackwell Publishing Ltd.
56. **Philip Lanzkowsky.** (2005). Manual of Pediatric Hematology and Oncology. 4th edition. London. UK. Elsevier Academic press.
57. **William F. Kern.** PDQ Hematology. (2002). 1st edition. Oklahoma City. B.C. Decker.
58. **Al Arrayed SS, Haites N.** (1995). Features of sickle-cell disease in Bahrain. East Mediterr Health J.; 1(1):112-9.
59. **Mary Louise Turgeon.** Clinical Hematology Theory and Procedures. 2<sup>nd</sup> (2008)
60. **Benjamin LJ,** Dampier CD, Jacob AK, et al. *Guideline for the Management of Acute and Chronic Pain in Sickle-Cell Disease.* APS Clinical Practice Guidelines Series, No. 1. Glenview, IL, 1999.
61. **Platt OS,** Thorington BD, Brambilla DJ, et al. Pain in sickle-cell disease. Rates and risk factors. *N Engl J Med* 1991; 325:11-6.
62. **Ballas SK,** Carlos TM, Dampier C, et al. *Guidelines for Standard of Care of Acute Painful Episodes in Patients with Sickle Cell Disease.* Pennsylvania Department of Health, 2000.
63. **Vichinsky EP,** Johnson R, Lubin BH. Multidisciplinary approach to pain management in sickle-cell disease. *Amer J Pediatr Hematol Onc* 1982; 4:328-33.
64. **Benjamin LJ,** Swinson GI, Nagel RL. Sickle cell anemia day hospital: an approach for the management of uncomplicated painful crises. *Blood* 2000; 95:1130-7.
65. **Walco GA,** Dampier CD. Pain in children and adolescents with sickle-cell disease: a descriptive study. *J Pediatr Psychol* 1990; 15:643-58.
66. **Wong DL,** Hackenberry-Eaton M, Wilson D, et al. *Whaley and Wong's Nursing Care of Infants and Children;* 6<sup>th</sup> edition. St. Louis: Mosby-Year Book, Inc., 1999:1153.
67. **Wong DL,** Baker CM. Pain in children: comparison of assessment scales. *Pediatr Nurs* 1998; 14:9-17.
68. **Styles LA,** Vichinsky E. Effects of a long-term transfusion regimen on sickle cell-related illnesses. *J Pediatr* 1994; 125:909-11.

## References

69. **Leikin SL**, Gallagher D, Kinney TR, et al. Mortality in children and adolescents with sickle cell disease. Cooperative Study of Sickle Cell Disease. *Pediatrics* 1989; 84:500-8.
70. **Gill FM**, Sleeper LA, Weiner SJ, et al. Clinical events in the first decade in a cohort of infants with sickle cell disease. Cooperative study of sickle cell disease. *Blood* 1995; 86:776-83.
71. **Bjornson AB**, Falletta JM, Verter JI, et al. Serotype specific immunoglobulin G antibody responses to pneumococcal polysaccharide vaccine in children with sickle cell anemia: effects of continued penicillin prophylaxis. *J Pediatr* 1996; 129:828-35.
72. **Gaston MH**, Verter JI, Woods G, et al. Prophylaxis with oral penicillin in children with sickle cell anemia. A randomized trial. *N Engl J Med* 1986; 314:1593-9.
73. **Williams LL**, Wilimas JA, Harris SC, et al. Outpatient therapy with ceftriaxone and oral cefixime for selected febrile children with sickle cell disease. *J Pediatr Hematol Oncol* 1996; 18:257-13.
74. **Platt OS**. The febrile child with sickle cell disease: a pediatrician's quandary. *J Pediatr* 1997; 130:693.
75. **Brown KE**, Young NS, Barbosa LH. Parvovirus B19: Implications for transfusion medicine. Summary of a workshop. *Transfusion* 2001;41:130-5
76. **Rednam KRV**, Jampol LM, Goldberg MF. Scatter retinal photocoagulation for proliferative sickle cell retinopathy. *Am J Ophthalmol* 1982; 93:594-9?
77. **Charache S**. Eye disease in sickling disorders. *Hematol Oncol Clin North Am* 1996; 10:1357-62.
78. **National Institute of Health**, National Heart, Lung, and Blood Institute Division of blood diseases and Resources NIH; 2002 4th Editions.
79. **DeSimone J**, Heller P, Hall L, Zwiers D. 5-Azacytidine stimulates fetal hemoglobin synthesis in anemic baboons. *Proc Natl Acad Sci USA* 1982; 79:4428-4431.
80. **Ley TJ**, DeSimone J, Noguchi CT, et al. 5-Azacytidine increases gamma-globin synthesis and reduces the proportion of dense cells in patients with sickle cell anemia. *Blood* 1983; 62:370-380.

## References

81. **Platt OS**, Orkin SH, Dover G, Beardsley GP, Miller B, Nathan DG. Hydroxyurea enhances fetal hemoglobin production in sickle cell anemia. *J Clin Invest* 1984; 74:652-656.
82. **Rodgers GP**, Dover GJ, Noguchi CT, Schechter AN, Nienhuis AW. Hematologic responses of patients with sickle cell disease to treatment with Hydroxyurea. *N Engl J Med* 1990; 322:1037-1045.
83. **El-Hazmi MAF**, Warsy AS, al-Momen A, Harakati M. Hydroxyurea for the treatment of sickle cell disease. *Acta Haematol* 1992; 88:170-174.
84. **Charache S**, Terrin ML, Moore RD, et al. Effect of Hydroxyurea on the frequency of painful crises in sickle cell anemia. *N Engl J Med* 1995; 332:1317-1322.
85. **Charache S**, Barton FB, Moore RD, et al. Hydroxyurea and sickle cell anemia -- clinical utility of a myelosuppressive "switching" agent: the Multicenter Study of Hydroxyurea in Sickle Cell Anemia. *Medicine (Baltimore)* 1996; 75:300-326.
86. **Hackney AC**, Hezier W, and Gullledge TP, et al. Effects of Hydroxyurea administration on the body weight, body composition and exercise performance of patients with sickle-cell anaemia. *Clin Sci (Colch)* 1997;92:481-486
87. **Styles LA**, Lubin B, Vichinsky E, et al. Decrease of very late activation antigen-4 and CD36 on reticulocytes in sickle cell patients treated with Hydroxyurea. *Blood* 1997; 89:2554-2559.
88. **Adragna NC**, Fonseca P, Lauf PK. Hydroxyurea affects cell morphology, cation transport, and red blood cell adhesion in cultured vascular endothelial cells. *Blood* 1994; 83:553-560.
89. **Bridges KR**, Barabino GD, Brugnara C, et al. A multiparameter analysis of sickle erythrocytes in patients undergoing Hydroxyurea therapy. *Blood* 1996;88:4701-4710
90. **Charache S**. Eye disease in sickling disorders. *Hematol Oncol Clin North Am* 1996; 10:1357-1362.
91. **Steinberg MH**, Nagel RL, Brugnara C. Cellular effects of Hydroxyurea in Hb SC disease. *Br J Haematol* 1997; 98:838-844.
92. **Steinberg MH**, Lu Z-H, Barton FB, Terrin ML, Charache S, Dover GJ. Fetal hemoglobin in sickle cell anemia: determinants of response to Hydroxyurea: Multicenter Study of Hydroxyurea. *Blood* 1997; 89:1078-1088.
93. **De Montalembert M**, Belloy M, Bernaudin F, et al. Clinical and hematological response of sickle cell children to treatment with Hydroxyurea. *Blood* 1994; 84: Supple 1:219A-219A.abstract.

## References

94. **Scott JP**, Hillary CA, Brown ER, Mickiewicz V, Labotka RJ. Hydroxyurea therapy in children severely affected with sickle cell disease. *J Pediatr* 1996; 128:820-828.
95. **Jayabose S**, Tugal O, Sandoval C, et al. Clinical and hematologic effects of Hydroxyurea in children with sickle cell anemia. *J Pediatr* 1996; 129:559-565.
96. **Ferster A**, Vermeylen C, Cornu G, et al. Hydroxyurea for treatment of severe sickle cell anemia: a pediatric clinical trial. *Blood* 1996; 88:1960-1964.
97. **Najean Y**, Rain J-D. Treatment of polycythemia Vera: use of <sup>32</sup>P alone or in combination with maintenance therapy using Hydroxyurea in 461 patients greater than 65 years of age. *Blood* 1997; 89:2319-2327.
98. **Nand S**, Stock W, Godwin J, Fisher SG. Leukemogenic risk of Hydroxyurea therapy in polycythemia Vera, essential thrombocythemia, and myeloid metaplasia with myelofibrosis. *Am J Hematol* 1996; 52:42-46
99. **Triadou P**, Maier-Redelsperger M, Krishnamoorthy R, et al. Fetal haemoglobin variations following Hydroxyurea treatment in patients with cyanotic congenital heart disease. *Nouv Rev Fr Hematol* 1994; 36:367-372
100. **Claster S**, Vichinsky E. First report of reversal of organ dysfunction in sickle cell anemia by the use of Hydroxyurea: splenic regeneration. *Blood* 1996; 88:1951-1953.
101. **Ferster A**, Bujan W, Corazza F, et al. Bone marrow transplantation corrects the splenic reticuloendothelial dysfunction in sickle cell anemia. *Blood* 1993; 81:1102-1105.
102. **Campbell PJ**, Olatunji PO, Ryan KE, Davies SC. Splenic regrowth in sickle cell anaemia following hypertransfusion. *Br J Haematol* 1997; 96:77-79.
103. **Voskaridou E**, Kalotychou V, Loukopoulos D. Clinical and laboratory effects of long-term administration of Hydroxyurea to patients with sickle-cell/ $\beta$ -thalassaemia. *Br J Haematol* 1995; 89:479-484.
104. **Loukopoulos D**. Current status of thalassemia and the sickle cell syndromes in Greece. *Semin Hematol* 1996; 33:76-86.
105. **El-Hazmi MAF**, al-Momen A, Kandaswamy S, et al. On the use of Hydroxyurea/erythropoietin combination therapy for sickle cell disease. *Acta Haematol* 1995; 94:128-134.
106. **Glibber JG**, Wandersee NJ, Little JA, Ginder GD. 5'-Flanking sequences mediate butyrate stimulation of embryonic globin gene expression in adult erythroid cells. *Mol Cell Biol* 1991; 11:4690-4697.
107. **Perrine SP**, Ginder GD, Faller DV, et al. A short-term trial of butyrate to stimulate fetal-globin-gene expression in the  $\beta$ -globin disorders. *N Engl J Med* 1993; 328:81-8.

## References

108. **Sher GD**, Ginder GD, Little J, Yang S, Dover GJ, Olivieri NF. Extended therapy with intravenous arginine butyrate in patients with  $\beta$ -Haemoglobinopathies. *N Engl J Med* 1995; 332:1606-1610.
109. **Dover GJ**, Brusilow S, Charache S. Induction of fetal hemoglobin production in subjects with sickle cell anemia by oral sodium phenylbutyrate. *Blood* 1994; 84:339-343.
110. **Blau CA**, Constantoulakis P, Shaw CM, Stamatoyannopoulos G. Fetal hemoglobin induction with butyric acid: efficacy and toxicity. *Blood* 1993; 81:529-537.
111. **Atweh GF**, Dover GJ, Faller DV, Stamatoyannopoulos G, Fournarakis B, Perrine SP. Sustained hematologic response to pulse butyrate therapy in beta globin disorders. *Blood* 1996; 88: Suppl 1:652a-652a.abstract.
112. **Clark MR**, Mohandas N, Shohet SB. Hydration of sickle cells using the sodium ionophore Monensin: a model for therapy. *J Clin Invest* 1982; 70:1074-1080.
113. **Orringer EP**, Brockenbrough JS, Whitney JA, Glosson PS, Parker JC. Okadaic acid inhibits activation of K-Cl cotransport in red blood cells containing hemoglobins S and C. *Am J Physiol* 1991;261:C591-C593.
114. **Berkowitz LR**, Orringer EP. Effect of cetiedil, an in vitro antisickling agent, on erythrocyte membrane cation permeability. *J Clin Invest* 1981; 68:1215-1220.
115. **Wolff D**, Cecchi X, Spalvins A, Canessa M. Charybdotoxin blocks with high affinity the Ca-activated  $K^+$  channel of Hb A and Hb S red cells: individual differences in the number of channels. *J Membr Biol* 1988; 106:243-252.
116. **Brugnara C**, de Franceschi L, Alper SL. Inhibition of  $Ca^{2+}$ -dependent  $K^+$  transport and cell dehydration in sickle erythrocytes by clotrimazole and other imidazole derivatives. *J Clin Invest* 1993; 92:520-526.
117. **De Franceschi L**, Saadane N, Trudel M, Alper SL, Brugnara C, Beuzard Y. Treatment with oral clotrimazole blocks  $Ca^{2+}$ -activated  $K^+$  transport and reverses erythrocyte dehydration in transgenic SAD mice: a model for therapy of sickle cell disease. *J Clin Invest* 1994; 93:1670-1676.
118. **Brugnara C**, Gee B, Armsby CC, et al. Therapy with oral clotrimazole induces inhibition of the Gardos channel and reduction of erythrocyte dehydration in patients with sickle cell disease. *J Clin Invest* 1996; 97:1227-1234.
119. **De Franceschi L**, Bachir D, Galactéros F, et al. Oral magnesium supplements reduce erythrocyte dehydration in patients with sickle cell disease. *J Clin Invest* 1997; 100:1847-1852.

## References

120. **De Franceschi L**, Rouyer-Fessard P, Alper SL, Jouault H, Brugnara C, Beuzard Y. Combination therapy of erythropoietin, Hydroxyurea, and clotrimazole in a  $\beta$  thalassemia mouse: a model for human therapy. *Blood* 1996; 87:1188-1195.
121. **Goldberg MA**, Brugnara C, Dover GJ, Schapiro L, Charache S, Bunn HF. Treatment of sickle cell anemia with Hydroxyurea and erythropoietin. *N Engl J Med* 1990; 323:366-372.
122. **Rodgers GP**, Dover GJ, Uyesaka N, Noguchi CT, Schechter AN, Nienhuis AW. Augmentation by erythropoietin of the fetal-hemoglobin response to Hydroxyurea in sickle cell disease. *N Engl J Med* 1993; 328:73-80.
123. **Charache S**, et al. 1995. Effect of hydroxyurea on the frequency of painful crises in sickle cell anemia. Investigators of the Multicenter Study of Hydroxyurea in Sickle Cell Anemia. *N. Engl. J. Med.* 332:1317–1322.
124. **Charache S**. 1997. Mechanism of action of hydroxyurea in the management of sickle cell anemia in adults. *Semin. Hematol.* 34:15–21.
125. **Lewis Mitchell**, Barbara J. Bain, Imelda Bates. (2006). *Practical Haematology*. 10th edition. Philadelphia, United States of America. Elsevier Ltd.
126. **Sysmex America Inc.** (2003). Sysmex Kx 21N. Document Number 10-1023 06/05 1M: (TOA Medical Electronics Co 1991).
127. **Atweh, G.F.**, and Schechter, A.N. 2001. Pharmacologic induction of fetal hemoglobin: raising the therapeutic bar in sickle cell disease. *Curr. Opin. Hematol.* 8:123–130.  
Richard E. Glove, Edward D, [Eugene P. Orringer](#). *Molecular Pharmacology* June 1, 1999 vol. 55 no. 6 1006-1010.
128. **Masoud Nahavandi**; Melville Q. Wyche; Elliott Perlin; Fatemeh Tavakkoli; Oswaldo Castro: Department of Anesthesiology, Center for Sickle Cell Disease, [Hematology](#), Volume 5, Issue 4 December 2000 , pages 335 – 339.
129. **Gladwin, M.T.**, et al. 2002. Nitric oxide donor properties of Hydroxyurea in patients with sickle cell disease. *Br. J. Haematol.* 116:436–444.
130. **C. Alvin Head**, Carlo Brugnara, Ricardo Martinez-Ruiz, Robert M. Kacmarek, Kenneth R. Bridges, David Kuter, Kenneth D. Bloch, and Warren M. Zapol. Department of Anesthesia and Critical Care, Hematology and Oncology Unit, and Cardiovascular Research Center of the Department of Medicine, Massachusetts General Hospital, Department of Pathology, Children’s Hospital, Department of Hematology, Brigham and Women’s Hospital, Harvard Medical School, Boston, Massachusetts 02115, *10 June 1997*.
131. **Chernecky & Berger**: *Laboratory Tests and Diagnostic Procedures*, 5th edition.

## References

132. **Betke K**, Marti HR, Schlicht L: Estimation of small percentage of fetal haemoglobin. *Nature* 1959; 184:1877-1878.
133. **Dacie John**, S.M. Lewis, B.G. Bain & I. Bates. (2001). *Practical Haematology*, 9th edition.
133. **John J. Strouse**, Sophie Lanzkron, Mary Catherine Beach, Carlton Haywood, MS, Haeseong Park, Catherine Witkop, Renee F. Wilson, Eric B. Bass, MD, Jodi B. Segal. Hydroxyurea for Sickle Cell Disease: A Systematic Review for Efficacy and Toxicity in Children. *PEDIATRICS* Vol. 122 No. 6 December 2008, pp. 1332-1342 (doi:10.1542/peds.2008-0441)
134. **Ballas SK**, Barton F, Castro O, Bellevue R, Investigators of the multicenter study of hydroxyurea in sickle cell anemia. Narcotic analgesia use among adult patients with sickle cell anemia [abstract]. *Blood* 1995;86(10 Suppl 1):642a. Ballas SK, Barton F, Castro O, Koshy M, Bellevue R. Pattern of narcotic analgesic consumption among adult patients with sickle cell anemia [abstract]. *Proceedings of the National Sickle Cell Disease Program 21st Annual Meeting*; 1996 March. 1996:63. Ballas SK, Barton FB, Waclawiw MA, Swerdlow P, Eckman JR, Pegelow CH, et al. Hydroxyurea and sickle cell anemia: effect on quality of life. *Health and Quality of Life Outcomes* 2006;4:59. Ballas SK, Marcolina MJ, Dover GJ, Barton FB. Erythropoietic activity in patients with sickle cell anaemia before and after treatment with hydroxyurea. *British Journal of Haematology* 1999;105(2):491-6.
135. **Zimmerman SA**, Schultz WH, Davis JS et al. Sustained long-term hematologic efficacy of hydroxyurea at maximum tolerated dose in children with sickle cell disease. *Blood* 2004;103(6):2039-45.
136. **El-Hazmi MA**, Warsy AS, al-Momen A et al. Hydroxyurea for the treatment of sickle cell disease. *Acta Haematol* 1992;88(4):170-4.