CHAPTER 14

FUTURE PROSPECTIVES

Future prospectives

During the project AT-4-074 a major consideration was directed towards epidemiological studies, investigations of disease pathophysiology, clinical presentation and study of the molecular nature of the genetic defects, gene-gene interactions, interaction with environmental factors, and investigation of ways to decrease the suffering of these patients by better management and care strategies. Programmes for control and prevention were devised and implemented. The conception of the SAS group played a role in the success of these activities. In addition, the designation of our unit as W.H.O. Collaborating Centre for Haemoglobinopathies, Thalassaemias and Enzymopathies played a significant role in the Referral & Consulting Scheme which have enable toward a better health care delivery, improved awareness, improved management, vaccination and prophylaxis strategies and genetic counselling. This has also help in standardization and continuation of such activities at the national level. Symposia were held to bring update the knowledge and recent developments in these fields. Workshops have been carried out to train more personnel to conduct efficient studies on blood genetic disorders, not only in Saudi Arabia but in other Middle Eastern countries.

It is contemplated that our efforts will enable effective health care delivery and will contribute positively to overcome the burden created by blood and other genetic problems.

A similar pattern of investigation for determining the magnitude of other genetic problems is complementary to these investigations. The model that has been arrived at during this study may be followed for development of facilities and strategies for the study of other genetic problems prevalent in the Saudi population.

Multifactorial and polygenic disorders are of similar chronic nature to that of red

cells genetic disorder and require very similar approach for control and prevention as well as care provision to the effected individuals and share the significance of finding preclinical markers. Molecular epidemiology and telecommunications may be of relevance and would result in unveiling of possible diagnostic probes and enable monitoring of disease states, respectively.

Studies at the level of the Arabian peninsula, including Gulf states and other neighbouring countries, will allow comparative studies and will widen the spectrum and further use of facilities. The establishment of the Arab Group for the Study of Red Cell Genetics Disorders and the W.H.O. Task Force and more recently the establishment of the panel of experts for genetic diseases for the World Health Organization would enable the exploration of areas of prevention, control and management at a regional and international level.

On the one hand, the progress made over the last years based on the acquisition of molecular methodologies and better control of infectious:disease has focussed attention on genetic disorders and other non-communicable diseases. It is, therefore, timely to gather efforts, expertise, and lessons from the studies hitherto carried out to widen the scheme to explore further the multifactorial and polygenic disorders in different population groups and tribes and to contribute to the advancement of knowledge to lay the basis for better diagnostic care and prevention strategies. On the other hand, enrichment of knowledge through study of human diversity and normal human genome will pave the way for interaction and learning experiences with the international community.

To this end we recommend a comprehensive approach for investigation, care, management, control and prevention of genetic disorders making use of the results, the

facilities and the expertise that have been developed as a result of this project.

CHAPTER:15

PAPERS PRESENTED IN NATIONAL & INTERNATIONAL JOURNALS FROM THE RESULTS OF PROJECT AT-4-074

15.1. Published/accepted papers

1. **El-Hazmi, M.A.F**. and Warsy, A.S. (1984): Aspects of sickle cell gene in Saudi Arabia – interaction with glucose-6-phosphate ehydrogenase deficiency. Hum Genet 68: 320- 323.

2. **El-Hazmi, M.A.F.** and Warsy, A.S. (1985): Glutathione Reductase Deficiency in Association with Sickle Cell and Thalassaemia Genes in Saudi Populations. Hum. Hered. 35: 326-332.

3. **El-Hazmi, M.A.F.** and Warsy, A.S. (1985): Glucose-6-phosphate Dehydrogenase Deficiency - Genetic, Pathophysiologic and Therapeutic:Aspects.Annals of Saudi Medicine 5(4): 213-223.

4. **El-Hazmi, M.A.F.** and Warsy, A.S. (1985): Hepatitis in Glucose-6-Phosphate Dehydrogenase Deficients - A Case for Concern in Saudi Arabia?:Saudi Medical Journal 6(6): 519-522.

5. **El-Hazmi, M.A.F.** (1985): Clinical manifestation and laboratory findings of sickle cell anaemia in association with α-thalassaemia in Saudi Arabia.Acta haemat. 74: 155-160.

6. **El-Hazmi, M.A.F.** and Warsy, A.S. (1986): Glucose-6-phosphate dehydrogenase polymorphism in Saudi population. Hum. Hered. 36: 24-30.

7. **El-Hazmi, M.A.F.**, Al-Swailem, A.R., Al-Faleh, F.Z. and Warsy, A.S. (1986): Frequency of glucose-6-phosphate dehydrogenase, pyruvate kinase and hexokinase deficiency in the Saudi population. Hum. Hered. 36: 45-49.

8. **El-Hazmi, M.A.F.**, Al-Faleh, F.Z. and Warsy, A.S. (1986): Epidemiology of Viral Hepatitis among Saudi population. 1. A Study of Viral Markers in Khaiber. Saudi

Medical Journal 7(2): 122-129.

9. **El-Hazmi, M.A.F.** (1986): Infections in sickle cell disease.Annals of Saudi Medicine 6(1): 33-41.

10. **El-Hazmi, M.A.F.** (1986): β-Globin gene polymorphism in the Saudi Arab population. Hum. Genet. 73: 31-34.

11. **El-Hazmi, M.A.F.** (1986): Studies on Sickle Cell Heterozygotes in Saudi Arabia - Interaction with α-Thalassaemia.Acta haemat. 75: 100-104.

12. **El-Hazmi, M.A.F.** (1986): Hepatitis B markers in Saudi Arabia: A comparative study in different regions.Annals of Saudi Medicine 6(3): 185-191.

13. **El-Hazmi, M.A.F.** and Warsy, A.S. (1986): Double Heterozygote Leftward/ Rightward Deletion Type Alpha-Thalassaemia in Saudi Arabs. Hum. Hered. 36: 222- 226.

14. **El-Hazmi, M.A.F.** (1986): Leftward deletion α-thalassaemia in the Saudi Arabian population. Hum. Genet. 74: 219-222.

15. **El-Hazmi, M.A.F.**, Jabbar, F.A., Al-Swailem, A.R. and Warsy, A.S. (1986): β-Globin gene polymorphism in Saudis - Triple Hpa I fragments. Hum. Genet. 74: 313-315.

16. **El-Hazmi, M.A.F.** and Warsy, A.S. (1986): On the Nature of Sickle Cell Disease in the South- Western Province of Saudi Arabia.Acta haemat. 76: 212- 216.

17. Warsy, A.S. and **El-Hazmi, M.A.F.** (1987): Glucose-6-phosphate Dehydrogenase Deficiency in Saudi Arabia - A Review. Saudi Medical Journal 8(1): 12-20.

18. **El-Hazmi, M.A.F.** (1987): The Distribution and nature of hemoglobinopathies in Arabia. In: William P. Winter (Ed.): Hemoglobin Variants in Human Populations.

CRC Press:Inc. Boca Raton, Florida.pp 65-77.

19. **El-Hazmi, M.A.F.** and Warsy, A.S. (1987): Interaction between glucose-6-phosphate dehydrogenase deficiency and sickle cell gene in Saudi Arabia. Trop. geogr. Med. 39: 32-35.

20. Warsy, A.S. and **El-Hazmi, M.A.F.** (1987): Bacterial Infections and the Sickling Disorders - A Review. Saudi Medical Journal 8(3): 228-237.

21. El-Hazmi, M.A.F. and Warsy, A.S. (1987): The Normal Range for Red Cell Pyruvate Kinase in the Saudi Population. Saudi Medical Journal 8(3): 246-249.

22. El-Hazmi, M.A.F. and Warsy, A.S. (1987): Glucose-6-phosphate Dehydrogenase Deficiency in Blood Donors. Saudi Medical Journal 8(3): 325-326 (Letter to the Editor)

23. El-Haazmi, M.A.F. and Al-Swailem, A.R. (1987): Sickle Cell-β°-Thalassaemia in Saudi Arabia. Hum. Hered. 37: 211-216.

24. El-Hazmi, M.A.F. and Warsy, A.S. (1987): Beta-Globin Gene Polymorphism in Saudis: 5.6 Hpa I Fragment. Hum. Hered. 37: 237-240.

25. El-Hazmi, M.A.F. and Warsy, A.S. (1987): A normal reference range for erythrocyte glucose-6-phosphate dehydrogenase in a Saudi population. Med. Lab. Sci. 44: 125-129.

26. El-Hazmi, M.A.F., Jabbar, F.A., Al-Faleh, F.Z. and Warsy, A.S. (1987): Triple Alpha Genes in Association with Sickle Cell and Beta-Thalassaemia Gene in the Saudi Population. Acta haemat. 77: 161-165.

27. El-Hazmi, M.A.F., Jabbar, F.A., Al-Faleh, F.Z., Al-Swailem, A.R. and Warsy, A.S. (1987): The haematological, biochemical and clinical presentation of

haemoglobin S in Saudi Arabia - (i) Haematological and clinical expression. Trop. geogr. Med. 39: 157-162.

28. El-Hazmi, M.A.F. (1987): α-Thalassaemia in Saudi Arabia: deletion pattern. Hum. Genet. 76: 196- 198.

29. **El-Hazmi, M.A.F.**, Jabbar, F.A. and Warsy, A.S. (1987): Cholesterol and triglyceride level in patients with sickle cell anaemia. Scand. J. Clin. Lab. Invest. 47: 351-354.

30. **El-Hazmi, M.A.F.** (1987): Haplotypes of Alpha-Globin-Gene in the Saudi Population - The Triplicated Gene ($\alpha\alpha\alpha^{anti\ 3.7/}$). Hum. Hered. 37: 314-318.

31. **El-Hazmi, M.A.F.**, Al-Swailem, A.R. and Warsy, A.S. (1987): Liver Function Tests in Sickle Cell Anemia Patients: A Case-Control Study in Saudi Arabia. Amer. J. Med. Sci. 30(6): 371-376.

32. **El-Hazmi, M.A.F.** (1987): Haemoglobinopathies, Thalassaemias and Enzymopathies in Saudi Arabia: The Present Status. Acta haemat. 78: 130- 134.

33. **El-Hazmi, M.A.F.**, Jabbar, F.A., Al-Faleh, F.Z., Al-Swailem, A.R. and Warsy, A.S. (1988): The Haematological, Biochemical and Clinical Presentation of Haemoglobin S in Saudi Arabia (ii) Biochemical Expression. Saudi Medical Journal 9(1): 66-71.

34. **El-Hazmi, M.A.F.** (1987): Haemoglobinopathies and Allied Disorders in Saudi Arabia. Proc. CMRC Symp Appl. M.R., Riyadh 1983. pp. 35-47.

35. **El-Hazmi, M.A.F.** (1987): 'Reference Values for Laboratory Parameters in Saudi Arabia.Proc. CMRC Sym. Appl. M.R. pp 3-16.

36. **El-Hazmi, M.A.F.** (1987): Haemoglobinopathies, Thalassaemias and Enzymo-

pathies in Saudi Arabia: The Present Status. Acta haemat. 78: 130- 134.

37. **El-Hazmi, M.A.F.**, Jabbar, F.A., Al-Faleh, F.Z., Al-Swailem, A.R. and Warsy, A.S. (1988): The Haematological, Biochemical and Clinical Presentation of Haemoglobin S in Saudi Arabia (ii) Biochemical Expression. Saudi Medical Journal 9(1): 66-71.

38. **El-Hazmi, M.A.F.** and Warsy, A.S.(1988): Correlation between red cell glucose-6-phosphate dehydrogenase level and haematological parameters. Med. Lab. Sci. 45: 255-260.

39. **El-Hazmi, M.A.F.** and Warsy, A.S. (1988): Glucose-6-Phosphate Dehydrogenase Deficiency in Saudi Arabia - A Study in Al-Ula. Hum. Hered. 38: 317-322.

40. **El-Hazmi, M.A.F.** (1989): Sickle Cell Anaemia Patients and Sickle Cell Trait Cases in Saudi Arabia. (Letter to the Editor). Saudi Medical Journal 10(1): 78-79.

41. **El-Hazmi, M.A.F.** (1989): Haemoglobinopathies in Saudi Arabia. (Letter to the Editor).Saudi Medical Journal 10(2): 161-163.

42. **El-Hazmi, M.A.F.**, Jabbar, F.A., Al-Faleh, F.Z., Al-Swailem, A.R. and Warsy, A.S. (1989):The haematological, biochemical and clinical presenta- tion of haemo-globin S in Saudi Arabia - (ii) Biochemical Expression (Letter to the Editor). Saudi Medical Journal 10(2):160-161.

43. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989): Glutathione Reductase in the South-Western Province of Saudi Arabia - Genetic Variation vs Acquired Deficiency. Haematologia 22(1): 37-42.

44. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989): Glutathione Reductase Deficiency in Saudi Arabia. Annals of Saudi Medicine 9(3): 263-266.

45. **El-Hazmi, M.A.F.** (1989): Xmn I Polymorphism in the Gamma-globin Gene Region among Saudis. Hum. Hered. 39: 12-19.

46. **El-Hazmi, M.A.F.** (1989): Hepatitis B virus in Saudi Arabia. Journal of Trop. Med. & Hyg. 92: 56-61.

47. **El-Hazmi, M.A.F.** (1989): Viral hepatitis. A health problem in Saudi Arabia. Proceedings of the 7e Colloque de Pont-a-Mousson, Biologie Prospective. John Libbey Eurotext, France. pp 963-966.

48. **El-Hazmi, M.A.F.** (1989): Anti-Delta in HBsAg Positive Saudis. Saudi Medical Journal 10(4): 286- 289.

49. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989): The effect of glucose-6-phosphate dehydrogenase deficiency on the haematological parameters and clinical manifestations in patients with sickle cell anaemia. Trop. geogr. Med. 41: 52-56.

50. **El-Hazmi, M.A.F.**, Al-Faleh, F.Z. and Warsy, A.S. (1989): Plasma Uric Acid, Urea and Creatinine in Sickle Cell Disease. Saudi Medical Journal 10(6): 471-476.

51. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989): Glucose-6-Phosphate Dehydrogenase and Pyruvate Kinase Levels in Anaemic Patients. (Letter to the Editor):. Saudi Medical Journal 10(6): 523.

52. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989): Glucose-6-Phosphate Dehydrogenase Deficiency in Blood Donors. (Letter to the Editor). Saudi Medical Journal 10(6): 524.

53. **El-Hazmi, M.A.F.** (1989): Sickle Cell Anaemia Patients and Sickle Cell Trait Cases in Saudi Arabia. (Letter to the Editor). Saudi Medical Journal 10(6): 529.

54. El-Hazmi, M.A.F. (1989): Regional variation in sickle cell anaemia in Saudi

Arabia. (Letter to the Editor). Annals of Saudi Medicine 9(5): 511-513.

55. **El-Hazmi, M.A.F.** and Warsy, A.S. (1990): Frequency of Glucose-6-Phosphate Dehydrogenase Phenotypes and Deficiency in Al-Baha. Hum Hered 39: 313-317.

56. **El-Hazmi, M.A.F.** (1990): Red cell genetic abnormalities in different regions of Saudi Arabia. Second proceedings of the CMRC Workshop on 'Applied Medical Research - Past, Present and Future'. King Saud University, Riyadh, 1985. pp 1-10.

57. **El-Hazmi, M.A.F.** and Warsy, A.S. (1990): Frequency of glucose-6-phosphate dehydrogenase phenotypes in Saudi population. Second Proceedings of the CMRC Workshop on 'Applied Medical Research - Past, Present and Future'. King Saud University, Riyadh, 1985. pp 11-16.

58. **El-Hazmi, M.A.F.**, Al-Momen, A.K. and Al-Swailem, A.R. (1990): The Values of Haematological and Biochemical Parameters in α-Thalassaemia.2 in Saudi Arabia.Saudi Medical Journal 11(1): 55-58.

59. **El-Hazmi, M.A.F.** The Frequencies of Hb S: α- and β-thalassaemia in Saudi Arabia: Preliminary National Values. (Letter to the Editor). Saudi Medical Journal 11(2): 161.

60. **El-Hazmi, M.A.F.** (1990): Microheterogeneity of glucose-6-phosphate dehydrogenase phenotypes. Med. Lab. Sci. 47: 26-29.

61. **El-Hazmi, M.A.F.**, Al-Faleh, F.Z. and Warsy, A.S. (1990): Red cell genetics and environmental interactions - Hepatitis B virus. Second proceedings of the CMRC Workshop on'Applied Medical Research - Past, Present and Future'. King Saud University, Riyadh, 1985. pp 17-28

62. **El-Hazmi, M.A.F.** (1990): Microheterogeneity of glucose-6-phosphate dehydro-

genase phenotypes. Med. Lab. Sci. 47: 26-29.

63. **El-Hazmi, M.A.F.** and Warsy, A.S. (1990): Correlation between red cell pyruvate kinase activity and the haematological parameters. Med. Lab. Sci. 47: 80-84.

64. **El-Hazmi, M.A.F.** (1990): Beta-Globin Gene Haplotype in Saudi Sickle Cell Anaemia Patients. Hum. Hered. 40: 177-186.

65. **El-Hazmi, M.A.F.**, Bahakim, H.M., Babikar, M.A., Al-Swailem, A.M. and Warsy, A.S. (1990): Symptom-Free Intervals in Sicklers - Does Pneumococcal Vaccination and:Penicillin Prophylaxis Have a Role?:Journal of Tropical Pediatrics 36: 56-62.

66. **El-Hazmi, M.A.F.** (1990): Hepatitis A antibodies: prevalence in Saudi Arabia. Journal of Trop. Med. & Hyg. 92: 427-430.

67. **El-Hazmi, M.A.F.** and Warsy, A.S. (1990): Frequency of glucose-6-phosphate dehydrogenase variants and deficiency in Arabia. Gene Geography 4: 15-20.

68. **El-Hazmi, M.A.F.**, Bahakim, H.M., Al-Swailem, A.M. and Warsy, A.S. (1990): The Features of Sickle Cell Disease in Saudi Children. Journal of Trop. Paediatrics 36: 148-155.

69. **El-Hazmi, M.A.F.**, Al-Swailem, A.R. and Warsy, A.S. (1990): Case Studies on haemoglobin S heterozygotes with severe clinical:manifestation. Journal of Trop. Paediatrics 36: 223-229.

70. **El-Hazmi, M.A.F.**, Al-Swailem, Bahakim, H.M., F.A., Al-Faleh, F.Z. and Warsy, A.S. (1990): Effect of thalassaemias, G-6-PD deficiency and Hb F on the nature of sickle cell disease in south-western Saudi Arabia. Trop. geogr. Med. 42: 241-247.

71. **El-Hazmi, M.A.F.**, Warsy, A.S. and Al-Momen, A.K. (1991): Glycosylated

haemoglobins in normal individuals and patients with red cell genetic diseases. Practical Diabetes Digest: 73-75.

72. **El-Hazmi, M.A.F.**, Jabbar, F.A., Al-Faleh, F.Z., Al-Swailem, A.R. and Warsy, A.S. (1991): Pattern for sickle cell, thalassaemia and glucose-6- phosphate dehydrogenase deficiency genes in north- western Saudi Arabia. Hum. Hered. 41 (1): 26-34.

73. **El-Hazmi, M.A.F.** (1991): Establishment of Laboratory 'Reference Values'. Saudi Medical Journal 12(3): 167-171

74. **El-Hazmi, M.A.F.** and Warsy, A.S. (1991): Glucose-6-phosphate dehydrogenase in Saudi population. Proceedings of the lst Internation Conference on `Human Genetics & Physical Anthropology' 9-12 December, 1989, Cairo, **Egypt.** pp: 77-87.

75. **El-Hazmi, M.A.F.** and Warsy, A.S. (1991 Glucose-6-Phosphate dehydrogenase variants and sickle cell genes in Al-Qunfuda. Trop. & Geogr. Med. 43: 174-179.

76. **El-Hazmi, M.A.F.** and Warsy, A.S. (1991): Sickle cell gene - Interaction with other red cell genetic abnormalities. Proceedings of the 1st International Conference on `Human Genetics & Physical Anthropology' 9-12 December, 1989, Cairo, **Egypt.** pp 21-48.

77. **El-Hazmi, M.A.F.** and Warsy, A.S. (1991):The Clinical, Haematological and Biochemical Expression of Haemoglobin S (HbS) in the Eastern Saudi Arabia. Journal of Islamic Academy of Sciences 4: 149-158.

78. **El-Hazmi, M.A.F.** (1992):α-Thalassaemia in Saudi Arabia - Restriction endonucleases as diagnostic tools. Saudi Medical Journal 13(4): 333-339.

79. **El-Hazmi, M.A.F.** (1992):Clinical and haematological diversity of sickle cell disease in Saudi children. Journal of Tropical Pediatrics 38: 106-112.

80. **El-Hazmi, M.A.F.** and Warsy, A.S. (1992):The frequency of glucose-6-phosphate dehydrogenase phenotypes and sickle cell gene in Al-Qasim. Annals of Saudi Medicine 12(5): 463-467.

81. **El-Hazmi, M.A.F.** (1992):Genetic Diseases in Arabia - A Model for National Awareness and Care Programme. Saudi Medical Journal 13 (6): 514-520.

82. **El-Hazmi, M.A.F.** (1992):Haemoglobinopathies, thalassaemias and enzymopathies in Saudi Arabia Saudi Medical Journal 13(6): 488-499.

83. **El-Hazmi, M.A.F.**, Bahakim, H.M. and Warsy, A.S. (1992):DNA polymorphism in the β-globin gene cluster in Saudi Arabs - Relation to severity of sickle cell anaemia. Acta Haematol 88: 61-66.

84. **El-Hazmi, M.A.F.**, Bahakim, H.M. and Al-Fawaz, I. (1992):Endocrine functions in sickle cell anaemia patients. Journal of Tropical Pediatrics 38: 307-313.

85. **El-Hazmi, M.A.F.** (1992):Heterogeneity and variationof clinical and haematological expression of Hb S in Saudi Araba. Acta haematol 88: 67-71.

86. **El-Hazmi, M.A.F.** and Warsy A.S. (1992):Triple alpha genes ($\alpha\alpha\alpha^{anti\ 3.7/}$) in a patient with sickle cell anaemia. Hum Hered 42: 360-366

87. **El-Hazmi, M.A.F.,** Warsy, A.S., Al-Momen, A.K. and Harakati, M. (1992): Hydroxyurea for treatment of sickle cell disease. Acta Haematol 88: 170-174.

88. **El-Hazmi, M.A.F.** (1993): The relationship of the genetic heterogeneity of sickle cell gene to clinical manifestation. Journal of Tropical Pediatrics 39: 23-29.

89. **El-Hazmi, M.A.F.** and Warsy, A.S. (1993):The frequency of Hb S and glucose-6-

phosphate dehydrogenase phenotypes in relation to malaria in western Saudi Arabia. Saudi Medical Journal 14(2): 121-125.

90. **El-Hazmi, M.A.F.**, Bahakim, H.M., Warsy, A.S., Al-Momen, A.K., Al-Wazzan, A., Al-Fawaz, I. and Huraib S. (1993): Does Gγ/Aγ ratio and Hb F level influence the severity of sickle cell anaemia.Molecular and Cellular Biochemistry Journal 124: 17-22.

91. **El-Hazmi, M.A.F.**, Warsy, A.S., Bahakim, H.M. and Al-Swailem, A.R. (1993): Glucose-6-phopshate dehydrogenase deficiency and sickle cell genes in two regions of Western Arabia. Annals of Saudi Medicine 13(3): 250-254.

92. **El-Hazmi, M.A.F.** (1993): Genetic Engineering and its Application to Medicine. Proceedings of the **Symposium on `DNA** Technology & Genetic Engineering in Human Diseases'. 13-14 March, 1990, College of Medicine, Riyadh, Saudi Arabia. pp 17-34.

93. **El-Hazmi, M.A.F.** Pathogenesis and Laboratory Diagnosis of Molecular Defects in the Thalassaemia. Proceedings of the **Symposium on `DNA** Technology & Genetic Engineering in Human Diseases'. 13-14 March, 1990, College of Medicine, Riyadh, Saudi Arabia. pp 49-64.

94. Warsy, A.S. and **El-Hazmi, M.A.F.**Pathogenesis and Laboratory Diagnosis of Molecular Defects in the Hemoglobinopathies. Proceedings of the **Symposium on `DNA** Technology & Genetic Engineering in Human Diseases'. 13-14 March, 1990, College of Medicine, Riyadh, Saudi Arabia. pp 65-81.

95. **El-Hazmi, M.A.F.** and Warsy, A.S. DNA Polymorphism for Genetic Diagnosis. Proceedings of the **Symposium on `DNA** Technology & Genetic Engineering in

Human Diseases'. 13-14 March, 1990, College of Medicine, Riyadh, Saudi Arabia. pp 91-101.

96. **El-Hazmi, M.A.F.** and Warsy, A.S. (1993):On the molecular interactions between thalassaemia and sickle cell gene. Journal Tropical Pediatrics 39: 209-213.

97. **El-Hazmi, M.A.F.** (1992):Blood Genetic Disorders.Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management', 18-20 April 1992, Riyadh, Saudi Arabia.

98. **El-Hazmi, M.A.F.** and Krishnamoorthy, R. The locus control region of the β-globin gene domain. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management'.

99. Warsy, A.S., **El-Hazmi, M.A.F.**, Al-Swailem, A.R. and Al-Swailem, A.M. The spectrum of β-thalassaemia mutations in the Saudi population. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

100. **El-Hazmi, M.A.F.**, Warsy, A.S., Al-Swailem, A.M. and Al-Swailem, A.R.Genetic Modulators of sickle cell anaemia. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

101. **El-Hazmi, M.A.F.**, Bahakim, H.M., Al-Fawaz, I., Al-Momen, A.K., Al-Wazzan, A., Huraib, S., Al-Harakati, M., Warsy, A.S. and Desouki M.Variations in the clinical patterns, laboratory values and complications of Saudi sickle cell disease. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

102. Al-Wazzan, A., Bahakim, H., Al-Momen, A.K., Al-Fawaz, I. and **El-Hazmi,**

M.A.F. The pulmonary manifestation in sickle cell anaemia. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

103. Al-Fawaz, I., **El-Hazmi, M.A.F.**, Bahakim, H.M. and Warsy, A.S. Cholelithiasis in sickle cell cell disease. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

104. Huraib, S., Al-Momen, A.K., **El-Hazmi, M.A.F.**, Al-Harakati, M., Warsy, A.S., Bahakim, H.M. and Fawaz, I: The pattern of renal involvement in sickle cell disease. Proceedings of the symposium on `Blood Genetic Disorders: Patho-physiology and Management' (In press).

105. Khashogi, T., Al-Nuaim, L., Ba-Aqeel, H. and **El-Hazmi, M.A.F.** Effects of the maternal sickle cell heterozygosity on the fetal outocme. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management'(In press).

106. **El-Hazmi, M.A.F.**, Al-Fawaz, I., Apawoye, A.D., Al-Magamci, M.S., El-Agib, A., Al-Swailem, A.R. and Rub, S. Treatment of sicklecell anaemia patients with piracetam - Results from a multicenter trial. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

107. **El-Hazmi, M.A.F.,** Al-Momen, A.K., Husraib, S., Al-Harakati, M., Al-Wazzan A. and Warsy, A.S. The clinical, haematological and biochemical modulation by fetal haemoglobin induction in sickle cell disease patients by hydroxyurea with or without rHUEpo. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

108. Warsy, A.S.. **E;-Hazmi, M.A.F.**, Al-Swailem, A.R. and Al-Swailem, A.M. Red cell enzymopathies in Arabia. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

109. **El-Hazmi, M.A.F.**, Mohareb, F., Al-Wazzan, A., Al-Harakati, M., Huraib, S., Al-Fawaz, I., Al-Swailem, A.R. and Abdel-Raheem, K. An awareness programme for blood genetic disorders. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

110. **El-Hazmi, M.A.F.**, Al-Harakati, M., Huraib, S., Al-Wazzan, A., Mohareb, F., Al-Fawaz, I. and Al-Swailem, A.R. Psycho-social effects of the haemoglobin disorders on the patient/family unit. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' (In press).

111. **El-Hazmi, M.A.F.**, Warsy, A.S., Bahakim H.M. and Al-Swailem, A.R.Glucose-6-phosphate dehydrogenase deficiency and sickle cell genes in Makkah - Saudi Arabia. Journal of Tropical Pediatrics (In press).

112. **El-Hazmi, M.A.F.,** Al-Swailem, A.R., Al-Fawaz, I., Warsy, A.S. and Al-Swailem, A.M.Diabetes Mellitus in Children Suffering from β-Thalassaemia. Journal of Tropical Medicine.

113. **El-Hazmi, M.A.F.**, Warsy, A.S., Al-Swailem, A.R., Al-Faleh, F.Z. and Al-Jabbar, F.A.Genetic Compounds - Hb S, Thalassaemias and Enzymopathies Spectrum of Interactions. Journal of Tropical Pediatrics (In press).

114. **El-Hazmi, M.A.F.** and Warsy, A.S.Iron-Endocrine Patterin in Patients with β-Thalassaemia. Journal of Tropical Medicine (In press).

115. **El-Hazmi, M.A.F.**Haemoglobin Disorders: A Global Extension of a Model Health

Care Programme. World Health Forum (In press).

116. **El-Hazmi, M.A.F.**, Warsy, A.S. and Bahakim, H.The Blood Proteins C&S in the Sickle Cell Disease. Acta haematologica (In press).

117. **El-Hazmi, M.A.F.**Genetic red cell disorders in Saudi Arabia - A multifaceted problem. HEMOGLOBIN (In press).

118. **El-Hazmi, M.A.F.**, Al-Swailem, A.R. and Warsy, A.S.Glucose-6-phosphate dehydrogenase deficiency and sickle cell genes in Bisha - Saudi Arabia. Tropical & Geographical Medicine (In press).

119. **El-Hazmi, M.A.F.,** Warsy, A.S. and Al-Fawaz, I.The pituitary-thyroid-gonadal axis in Saudi β-thalassaemia patients. Proceedings of the International Mediterranean Conference on Endocrine Disorders in Thalassaemia (In press).

120. **El-Hazmi, M.A.F.,** Warsy, A.S., Addar, M.H.N. and Habib Z.Fetal hemoglobin level - Effect of gender, age and haemoglobin disorders.

121. **El-Hazmi, M.A.F.,** Warsy, A.S., Al-Swailem, A.R. and Bahakim, H.Red Cell Genetic Disorders and Plasma Lipids.

122. **Submitted El-Hazmi, M.A.F.** and Al-Swailem, A.R.Molecular Defects in β-Thalassaemias in the Population of Saudi Arabia.

123. **El-Hazmi, M.A.F.,** Al-Swailem, A.R., Warsy, A.S., Sulimani, R. and Al-Swailem. A.M.Prevalence of Diabetes Mellitus in Saudi Arabia

124. **El-Hazmi, M.A.F.,** Sulaimani, R.A., Warsy, A.S., Al-Swailem, A.R., Baraimah, N. and Adeeb, A.B.Asp 57 and Arg 52 of the HLA-DQ as genetic markers for insulin dependent diabetes mellitus in Saudis.

125. **El-Hazmi, M.A.F.,** Al-Momen, A.K., Kandaswamy, S., Huraib, S., Harakati, M.,

Al-Mohareb, F/ and Warsy, A.S.The pharmacological manopulation of fetal haemoglobin - Trails using hydroxyurea and rHuEpo.

126. **El-Hazmi, M.A.F.,** Al-Momen, A.K., Kandaswamy, S., Huraib S., Harakati, M., Al-Mohareb, F. and Warsy. A.S.On the use of hydroxyurea/erythropoietin combination therapy for sickle cell disease.

127. **El-Hazmi, M.A.F.,** Warsy, A.S. and Al-Swailem, A.R.Spectrum of Beta-Thalassaemia Mutations in the Arab Populations.

128. **El-Hazmi, M.A.F.,** Al-Momen, A.K., Warsy, A.S., Al-Harakati, M., Huraib, S. and Al-Desouki, M.Potential molecular therapy of sickle cell anaemia.

129. **El-Hazmi, M.A.F.** and Warsy, A.S. β^S-Globin Gene Haplotypes in Saudi Arabia.

130. **El-Hazmi, M.A.F.,** Warsy, A.S. and Al-Swailem, A.R. A comparison of the frequency of eleven beta-thalassaemia mutations in the Arab populations.

131. **El-Hazmi, M.A.F.** and Warsy, A.S. Plasma Uric Acid in Saudi Arabs.

CHAPTER 16

PAPERS PRESENTED

IN NATIONAL

AND INTERNATIONAL

CONFERENCES

DURING THE

PROJECTAT-4-074

1. **El-Hazmi, M.A.F.** (1983): Haemoglobinopathies and allied disorders in Arabia - State of the Art. The First CMRC Workshop, King Saud University, 10-12 May, 1983, Riyadh, Saudi Arabia.

2. **El-Hazmi, M.A.F.** (1983): Association of Hb F and its subunits with different haemoglobin disorders. International Society of Haematology European African Division 7th Meeting. Barcelona, 4-9 September, 1983, Barcelona, Spain.

3. **El-Hazmi, M.A.F.** (1983): Molecular variants of red cell enzymes in Saudi Arabia - G- 6-PD deficiency. International Society of Haematology European and African Division 7th Meeting. Barcelona, 4-9 September, 1983, Barcelona, Spain.

4. **El-Hazmi, M.A.F.**, Al-Faleh, F.Z., Jabbar, F.A., Swailem, A.R. (1983): The frequency of red cell glucose-6-phosphate dehydrogenase, pyruvate kinase and hexokinase deficiency in Riyadh, Hafouf and Khaiber - Comparative studies. 8th Saudi medical Conference, 30 October - 3 November, 1983, Riyadh, Saudi Arabia.

5. **El-Hazmi, M.A.F.**, Al-Faleh, F.Z., Jabbar, F.A. (1983) Incidence and prevalence of hepatitis with and without G-6-PD deficiency in Saudi Arabia. 8th Saudi Medical Conference, 30 October - 3 November, 1983, Riyadh, Saudi Arabia.

6. **El-Hazmi, M.A.F**. (1983): α-Thalassaemia in Arabia - Clinical manifestation and laboratory diagnosis. 2nd African, Mediterranean Near East Congress of Clinical Chemistry, 20-24 November, 1983, Cairo, Egypt.

7. **El-Hazmi, M.A.F.** (1984): The biochemical genetics of sickle cell anaemia and common haemoglobinopathies. Symposium on Haemoglobinopathies, Thalassaemias and Enzymopathies. King Saud University, College of Medicine, 2-3 May, 1984, Riyadh, Saudi Arabia.

8. **El-Hazmi, M.A.F**., Jabbar, F.A., Al-Faleh, F.Z., Swailem, A.R. and Warsy, A.S. (1984):: The clinical, haematological and biochemical expression of haemoglobin S in Saudis. Symposium on Haemoglobinopathies, Thalassaemias and Enzymopathies. King Saud University, College of Medicine, 2-3 May, 1984, Riyadh, Saudi Arabia.

9. **El-Hazmi, M.A.F.** (1984): Recent trends in molecular therapy of sickle cell anaemia. Symposium on Haemoglobinopathies, Thalassaemias and Enzymopathies. King Saud University, College of Medicine, 2-3 May, 1984, Riyadh, Saudi Arabia.

10. Warsy, A.S., and **El-Hazmi, M.A.F.** (1984): Enzymopathies affecting the red cell metabolism - Historical perspective and current status. Symposium on Haemoglobinopathies, Thalassaemia and Enzymopathies. King Saud University, College of Medicine, 2-3 May, 1984, Riyadh, Saudi Arabia.

11. **El-Hazmi, M.A.F.** (1984): Biochemical Variables in blood donation: Red cell enzyme. Symposium on "Blood Utilization and Blood Banking'. 17-19 September, 1984, Jeddah, Saudi Arabia.

12. **El-Hazmi, M.A.F.** (1984): Students and faculty views of a medical college environment in Saudi Arabia. Annual Meeting of the Association for the Study of Medical Education (ASME):, University of Leicester, U.K.

13. **El-Hazmi, M.A.F.** (1984): Infections in Haemoglobinopathies. Symposium on Infectious Diseases. 11-12 November, 1984, Riyadh, Saudi Arabia.

14. **El-Hazmi, M.A.F.** (1985): Genetic red cell abnormalities in different regions in Saudi Arabia. The Second CMRC Workshop, King Saud University, 3-5 February,

1985, Riyadh, Saudi Arabia.

15. **El-Hazmi, M.A.F.**, Al-Faleh, F.Z., and Warsy, A.S. (1985): Red cell genetic and environmental interactions - Hepatitis B virus. The Second CMRC Workshop, King Saud University, 3-5 February, 1985, Riyadh, Saudi Arabia.

16. **El-Hazmi, M.A.F**. and Warsy, A.S. (1985): Frequency of G-6-PD phenotypes in the Saudi population. The Second CMRC Workshop, King Saud University, 3-5 February, 1985, Riyadh, Saudi Arabia.

17. **El-Hazmi, M.A.F.** (1986): β-Globin gene polymorphism in Arabia.International symposium on "The Impact of Biotechnology on Diagnostic". 16-18 April, 1985, Rome, Italy.

18. **El-Hazmi, M.A.F.** (1985): α-Thalassaemia in Saudi Arabia - Restriction endonucleases. International Symposium on 'The Impact of Biotechnology on Diagnostic'. 16-18 April, 1985, Rome, Italy.

19. **El-Hazmi, M.A.F.** (1985): Red cell genetic disorders - interaction with hepatitis B virus. 37th National Meeting of the American Association for Clinical Biochemistry, 21-26 July, 1985, Atlanta, Georgia. U.S.A.

20. **El-Hazmi, M.A.F.** (1987): Haemoglobinopathies, Thalassaemias and Enzymopathies in Saudi Arabia: The Present Status. Haematology Meeting, 8-10 September, 1986, St. Bartholomew's Hospital, London, U.K.

21. **El-Hazmi, M.A.F.** and Warsy, A.S. (1987): Enzymopathies in Saudi Arabia. Second National Meeting of Chemistry, College of Science,, King Saud University, 7-9 March, 1987, Riyadh, Saudi Arabia.

22. Babiker, M.A., Bahakim, H.M. and **El-Hazmi, M.A.F.** (1987):

Hepatitis B and A markers in the Saudi population. Second National Meeting of Chemistry, College of Science, King Saud University, 7-9 March, 1987, Riyadh, Saudi Arabia.

23. **El-Hazmi, M.A.F.**, Al-Swailem, A.R., Al-Faleh. F.Z., Jabbar, F.A. (1987): Alpha-thalassaemia gene in Saudi Arabia - Genetic study and clinical manifestations. Second National Meeting of Chemistry, College of Science, King Saud University, 7-9 March, 1987, Riyadh, Saudi Arabia.

24. **El-Hazmi, M.A.F.** and Warsy, A.S. (1988) Molecular analysis of β-globin gene haplotypes in Saudi Sickle Cell Anaemia patients. 40th National Meeting of the American Association for Clinical Chemistry. 24-28 July, 1988, New Orleans, Louisiana, U.S.A.

25. **El-Hazmi, M.A.F.**, Babiker, M., Bahakim, H.M., Al-Swailem, A., Warsy, A.S. (1989): National history of sickle cell disease in south-western Saudi Arabia. 4th Annual Paediatric Symposium, King Fahad National Guard Hospital, 26-27 March, 1989, Riyadh.

26. **El-Hazmi, M.A.F.** (1989): Clinical manifestations and molecular pathology of sickle cell disease. 4th Annual Paediatric Symposium, King Fahad National Guard Hospital, 26-27 March, 1989, Riyadh.

27. **El-Hazmi, M.A.F.**, Babiker, M., Bahakim, H.M., Al-Swailem, A., Warsy, A.S. (1989): Symptom-free intervals in sicklers: Does phenomococcal vaccination have a role? 4th Annual Paediatric Symposium, King Fahad National Guard Hospital, 26-27

March, 1989, Riyadh.

28. **El-Hazmi, M.A.F.** (1989):

Anti-delta and HBeAg in hepatitis B surface antigen positive individuals. 3rd Saudi Symposium on Gastroenterology and Hepatology, College of medicine, King Saud University, 26-28 February, 1989, Riyadh.

29. **El-Hazmi, M.A.F.** (1989):

Red cell enzyme levels in association with genetic and acquired diseases. 8th International Congress of Clinical Enzymology, 30 May - 1 June, 1989, Toronto, Canada.

30. **El-Hazmi, M.A.F.** (1989):

Haemoglobinopathies and thalassaemias - What's new in diagnostic procedure. The First International Conference on Human Genetics and Physical Anthropology, 4-7 December 1989, Cairo, Egypt.

31. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989):

Sickle cell gene - interaction with other red cell genetic abnormalities. The First International Conference on Human Genetics and Physical Anthropology, 4-7 December 1989, Cairo, Egypt.

32. **El-Hazmi, M.A.F.** and Warsy, A.S. (1989):

Glucose-6-phosphate dehydrogenase in Saudi population. The First International Conference on Human Genetics and Physical Anthropology, 4-7 December 1989, Cairo, Egypt.

33. **El-Hazmi, M.A.F.** (1990):

Association between malaria and red cell genetic disorders in Saudi

Arabia.Symposium on "Endemic Diseases in the Kingdom", 9-10 January 1990, Al-Baha, Saudi Arabia.

34. **El-Hazmi, M.A.F.** (1990):

Heterogeneity in the genetic basis of sickle cell gene - its relation to clinical manifestation. First Symposium on "Sickle Cell Disease", 14-15 February 1990, Qatif Central Hospital, Qatif, Saudi Arabia.

35. **El-Hazmi, M.A.F.**, Bahakim, H, Al-Momen, A.K., Al-Wazan, A., Al-Fawaz, I., Huraib, S. and Al-Nasser, A. (1990): Management of Sickle Cell Disease. First Symposium on "Sickle Cell Disease, 14-15 February, 1990, Qatif Central Hospital, Qatif, Saudi Arabia.

36. **El-Hazmi, M.A.F.**, Warsy, A.S. and:Al-Momen, A.K. (1990): Glycosylated haemoglobin in normal individuals and patients with red cell genetic disease.Fourth Kuwait International Medical Science Conference on `Diabetes Mellitus', 4-8 March, 1990, Kuwait.

37. **El-Hazmi, M.A.F.** (1990):

Recombinant DNA Technology in Diagnosis of Haemoglobinopathies and thalassaemias. 2nd Paediatric Symposium, 19-20 June 1990, King Fahad Hofouf Hospital, Al-Hofouf, Saudi Arabia.

38. **El-Hazmi, M.A.F.**, Bahakim, H., Al-Momen, A.K., Warsy, A.S., Al-Wazzan, A., Al-Fawaz, A. and Huraib S. (1990): Influence of coexisting G-6-PD deficiency and α-thalassaemia on the severity of sickle cell disease in Saudi Children. 2nd Paediatric Symposium, 19-20 June 1990, King Fahad Hofouf Hospital, Al-Hofouf, Saudi Arabia.

39. **El-Hazmi, M.A.F.** (1990):

Advances in diagnosis and management of haemoglobinopathies.Paediatrics Symposium, 17-18 October, Madina, Saudi Arabia.

40. **El-Hazmi, M.A.F.** (1990):

The molecular pathology and diagnosis of the thalassaemia. First Anglo- Egyptian Conference on `Bioscience and Technology' 10-15 November, 1990, Alexandria, Egypt.

41. **El-Hazmi, M.A.F.** (1990):

Clinical and haematological expression of HbS in Saudis.Joint meeting of the International Society of Hematology (ISH) and the American Society of Hematology (ASH),28 November - 4 December 1990, Boston, USA.

42. **El-Hazmi, M.A.F.**, Bahakim, H., Warsy, A.S., Al-Momen, A.K., Al-Wazzan, A., Al-Fawaz, I and Huraib S. (1990): Does G /A:ratio and HbF level influence the severity of sickle cell anaemia?:Joint meeting of the International Society of Hematology (ISH):and the American Society of Hematology (ASH):, 28 November - 4 December 1990, Boston, USA.

43. **El-Hazmi, M.A.F.**, Bahakim, H. and Warsy, A.S. (1990): DNA polymorphism in the β-globin gene cluster in Saudis. Joint meeting of the International Society of Hematology (ISH): and the American Society of Hematology (ASH):, 28 November - 4 December 1990, Boston, USA.

44. **El-Hazmi, M.A.F.**, (1991): Molecular basis of the thalassaemia in Arabian ethnic population.Meeting of the Egyptian Society of Clinical Chemistry, 27-28 June 1991, Alexandria, Egypt.

45. **El-Hazmi, M.A.F.**, and Warsy, A.S. (1991): Iron-endocrine pattern in patients with β°-thalassaemia. 43rd National Meeting of the American Association for Clinical Chemistry (AACC):, 28 July - 1 August 1991, Washington DC, USA.

46. **El-Hazmi, M.A. F.** Bahakim H. and Al-Fawaz, I. (1991): Endocrine Functions in Sickle Cell Anaemia. 43rd National Meeting of the American Association for Clinical Chemistry (AACC):, 28 July - 1 August 1991, Washington DC, USA.

47. **El-Hazmi, M.A. F.** Warsy, A.S., Al-Momen, A.K. and Al-Harakati, M. (1991): Hydroxyurea for Treatment of Sickle Cell Disease.43rd National Meeting of the American Association for Clinical Chemistry (AACC):, 28 July - 1 August 1991, Washington DC, USA.

48. **El-Hazmi, M.A. F.** Warsy A.S. and Al-Momen, A.K. (1991): Alpha Thalassaemia, $\alpha\alpha\alpha^{anti\ 3.7/}$ and Sickle Cell Gene - Molecular Interactions.XIth Meeting of the International Society of Haematology (European and African Division):, 31 August - 6 September, 1991 Basel - Switzerland.

49. **El-Hazmi, M.A. F.** Al-Momen, A.K., Warsy, A.S., Harakati, M., Huraib S. and Desouki M. (1991): Potential Molecular Therapy of Sickle Cell Anaemia.XIth Meeting of the International Society of Haematology (European and African Division):, 31 August -6 September, 1991, Basel - Switzerland.

50. **El-Hazmi, M.A.F.**, (1991): Genetic diseases in Arabia - A model for a national awareness and care programme.International Congress on Genetic Diseases of Hemoglobin. 6-8 November 1991, Nice - France.

51. **El-Hazmi, M.A. F.** Huraib, S., Al-Momen, A.K., Warsy, A.S. (1991): Management of severe sickle cell disease - A role for erythropoietin and

hydroxyurea.International Congress on Genetic Diseases of Hemoglobin. 6-8 November 1991, Nice - France.

52. **El-Hazmi, M.A. F.** Warsy, A.S. and Al-Fawaz, I. (1991): Molecular pathology of β-thalassaemias in Arabia.International Congress on Genetic Diseases of Hemoglobin. 6-8 November 1991, Nice - France.

53. **El-Hazmi, M.A.F.**, and Warsy, A.S. (1991): Diabetes mellitus in patients with β-thalassaemia. The Alexandria International Diabetes Days.8-15 November 1991, Alexandria, Egypt.

54. **El-Hazmi, M.A. F.** Al-Wazzan, A. and Opawoye, A.D. (1992): Immunological aspects of the haemoglobinopathies.6th Annual Pediatric Symposium.8-9 February 1992, Riyadh, Saudi Arabia.

55. **El-Hazmi, M.A. F.** Al-Fawaz, I. and Warsy, A.S. (1992): Red cell deformability and the effect of piracetam in sickle cell syndrome.6th Annual Pediatric Symposium.8-9 February 1992, Riyadh, Saudi Arabia.

56. **El-Hazmi, M.A. F.** Al-Momen, A.K., Warsy, A.S. and Bahakim, H.M. (1992): The blood proteins C & S in the sickle cell disease.6th Annual Pediatric Symposium.8-9 February 1992, Riyadh, Saudi Arabia.

57. **El-Hazmi, M.A.F.** (1992): Molecular markers of the severity of sickle cell anaemia.Symposium on Hemoglobinopathies. 26 February 1992, Qatif, Saudi Arabia.

58. **El-Hazmi, M.A.F.**, (1992): Blood Genetic Disorders. Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

59. **El-Hazmi, M.A.F.**, and Krishnamoorthy, R. (1992): The locus control region of the β-globin gene domain. Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

60. Warsy, A.S., **El-Hazmi, M.A. F.** Al-Swailem, A.R. and Al-Swailem, A.M. (1992): The spectrum of β-thalassaemia mutations in the Saudi population. Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

61. **El-Hazmi, M.A. F.** Warsy, A.S., Al-Swailem, A.M. and Al-Swailem, A.R. (1992): Genetic modulators of sickle cell anaemia. Symposium on Blood Genetic Disorders -Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

62. Perichon, B., Lapoumeroulie, C., Elion, J., Ragusa, A., **El-Hazmi, M.A.F.**, and Krishnamoorthy, R. (1992): β-thalassaemiaand sickle cell anemia: Epistatic effect of different DNA sequence elements dictating the clinical and haematological phenotype. Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

63. **El-Hazmi, M.A. F.** Bahakim, H.M., Al-Fawaz, I., Al-Momen, A.K., Al-Wazzan, A., Huraib, S., Al-Harakati, M., Warsy, A.S. and Desouki, M. (1992): Variations in the clinical patterns,laboratory values and complications of Saudi sickle cell disease. Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

64. Al-Wazzan, A., Bahakim, H., Al-Momen, A.K., Al-Fawaz, I. and **El-Hazmi, M.A.F.**, (1992): The pulmonary manifestation in sickle cell anaemia. Symposium

on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

65. Al-Fawaz, I., **El-Hazmi, M.A. F.** Bahakim, H.M. and Warsy, A.S. (1992): Cholelithiasis in sickle cell disease. Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

66. Huraib, S., Al-Momen, A.K., **El-Hazmi, M.A. F.** Al-Harakati, M., Warsy, A.S., Bahakim, H.M.and Al-Fawaz, I. (1992): The pattern of renal involvement in sickle cell disease.Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

67. **El-Hazmi, M.A. F.** Al-Fawaz, I., Apawoye, A.D., Al-Magamci, M.S., El-Agi, A., Al-Swailem, A.R. and Rub, S. (1992): Treatment of sickle cell anaemia patients with piracetam - Results from a multicentre trial.Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

68. **El-Hazmi, M.A. F.** Al-Momen A.K., Huraib, S., Al-Harakati, M., Al-Wazzan, A. and Warsy, A.S. (1992): The clinical, haematological and biochemical modulation by fetal haemoglobin induction in sickle cell disease by hydroxyurea with or without rHuEpo.Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

69. Warsy, A.S., **El-Hazmi, M.A. F.** Al-Swailem, A.R. and Al-Swailem, A.M. (1992): Red cell enzymopathies in Arabia.Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

70. **El-Hazmi, M.A. F.** Al-Mohareb, F., Al-Wazzan, A., Al-Harakati, M., Huraib, S.,

Al-Fawaz, I., Al-Swailem, A.R. and:Abdel-Rahim, K. (1992): An awareness programme for blood genetic disorders.Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

71. **El-Hazmi, M.A. F.** Al-Harakati, M., Huraib, S., Al-Wazzan, A., Mohareb, F., Al-Fawaz, I. and Al-Swailem, A.R. (1992):: Psycho-social effects on thehaemoglobin disorders on the patient/family unit.Symposium on Blood Genetic Disorders - Pathophysiology & Management. 18-20 April 1992, Riyadh, Saudi Arabia.

72. **El-Hazmi, M.A. F.** Warsy, A.S. and Al-Fawaz, I. (1992): The pituitary-thyroid-gonadal axis in Saudi β-thalassaemia patients.International Mediterranean Conference on Endocrine Disorders in Thalassaemia. 7-9 May, 1992, Rende, Italy.

73. **El-Hazmi, M.A.F.**, (1992): Evaluation of serum erythropoietin and iron status in the hemoglobinopathies. 24th Congress of the International Society of Haematology, 23-27 August, 1992, London, UK.

74. **El-Hazmi, M.A. F.** Al-Momen, A.K., Warsy, A.S., Al-Harakati, M. and Al-Mohareb, F. (1992): Hydroxyurea and/or erythropoietin - Single and combination therapy for treatment of sickle cell syndrome. 24th Congress of the International Society of Haematology, 23-27 August, 1992, London, UK.

75. **El-Hazmi, M.A. F.** Warsy, A.S., Al-Swailem, A.R., Al-Faleh, F.Z. and Jabbar, F.A. (1992): Genetic Compounds - Hb S, Thalassaemias and Enzymopathies Spectrum of Interactions. 24th Congress of the International Society of Haematology, 23-27 August, 1992, London, UK.

76. **El-Hazmi, M.A.F.**, (1992): Red cell genetic disorders in Saudi Arabia. 32nd Scientific Week, 11 November, 1992, Damascus, Syria.

77. **El-Hazmi, M.A.F.**, (1992): Human Haemoglobin Disorders: A Global Health Problem. 32nd Scientific Week, 11 November, 1992, Damascus, Syria.

78. **El-Hazmi, M.A.F.**, (1922): Molecular biology and clinical correlation in sickle cell anaemia.International Symposium on Paediatrics, 17 February, 1993, Jeddah, Saudi Arabia.

79. **El-Hazmi, M.A.F.**, (1993): The clinical significance of gene interactions in sickle cell disease. 5th International Conference on Thalassaemias and Haemoglobinopathies, 29 March - 3 April, 1993, Nicosia, Cyprus.

80. **El-Hazmi, M.A.F.**, (1992): The Haemoglobin F gene(s) switching pattern in Saudi Arab population. 5th International Conference on Thalassaemias and Haemoglobinopathies, 29 March -3 April, 1993, Nicosia, Cyprus.

81. **El-Hazmi, M.A. F.** Al-Swailem, A.R., Bahakim, H.M., Warsy, A.S. and Al-Swailem, A.M. (1993): Red cell genetic disorders in Saudi Arabia. 5th International Conference on Thalassaemias and Haemoglobinopathies, 29 March - 3 April, 1993, Nicosia, Cyprus.

82. **El-Hazmi, M.A. F.** Al-Momen, A.K., Kandaswamy S., Huraib, S., Harakati, M., Mohareb F. and Warsy, A.S. (1993): The pharmacological manipulation of fetal haemoglobin - Trials using hydroxyurea and rHuEpo. 5th International Conference on Thalassaemias and Haemoglobinopathies, 29 March - 3 April, 1993, Nicosia, Cyprus.

83. **El-Hazmi, M.A. F.** Al-Fawaz, I. Bahakim, H.M., Warsy, A.S., Hawsawi, Z and Mohammad A.A. (1993): Beta-Thalassaemia Major in Saudi Arabia. 5th International Conference on Thalassaemias and Haemoglobinopathies, 29 March -

3 April, 1993, Nicosia, Cyprus.

84. **El-Hazmi, M.A. F.** Al-Momen, A.K., Kandaswamy, S., Harakati, M., Huraib, S., Mohareb, F. and Warsy, A.S. (1993): Recombinant Human Erythropoietin (rHuEpo) and Hydroxyurea (H.U.) Combination Therapy in Sickle Cell Disease. Second International EPREX Nephrology Congress, 29 April - 3 May, 1993, Luzern, Switzerland.

85. **El-Hazmi, M.A. F.** Warsy, A.S., Al-Fawaz, I., Habib, Z. (1993): Fetal hemoglobin level - Effects of gender, age and hemoglobin disorders. 1993 National Meeting of the American Association of Clinical Chemistry (AACC), 11-15 July, 1993, New York, NY, U.S.A.

86. **El-Hazmi, M.A. F.** Warsy,A.S., Al-Swailem, A.R., Al-Swailem, A.M. and Bahakim, H. (1993): Red cell genetic disorders and plasma lipids. 1993 National Meeting of the American Association of Clinical Chemistry (AACC), 11-15 July, 1993, New York, NY, U.S.A.

87. **El-Hazmi, M.A. F.** Al-Momen, A.K., Harakati, M., Huraib, S., Mohareb F and Warsy, A.S. (1993):: Effect of hydroxyurea with or without rHuEpo for the treatment of sickle cell disease. 14th Annual Congress of Alexandria Faculty of Medicine, 27-29 July, 1993 Alexandria, Egypt.

88. **El-Hazmi, M.A.F.** (1993): Is r-HuEPO + H. Urea more effective than r-HuEPO alone in Sickle Cell Anaemia? International Hemoglobinopathy Seminar, 19-20 November, 1993, Athens.

CHAPTER 17

LIST OF

REFERENCES

Abraham EC and Huisman TH. Differences in affinity of variant beta chains for alpha chains: a possible explanation for the variation in the percentages of beta chain variants in heterozygotes. Hemoglobin 1977; 1:861-73.

Acquaye J, Ganeshaguru K, Sejeny S, Hassounah F, Samuel A, Omer A. A study of alpha thalassaemia families in western Saudi Arabia. Trop geogr Med 1985; 37: 319-327.

Acquaye JK, Omer A, Ganeshaguru K, Sejeny SA, Hoffbrand AV. Non-benign sickle cell anaemia in western Saudi Arabia. Br J Haemat 1985; 60: 99-108.

Adeloya A, Odeku EL. The nervous system in sickle cell disease. Afr. J Med Sci 1970; 1:33-48.

Ager JAM and Lehmann H. Observations on some 'fast' haemoglobin. K.J.N. and Barts Br. Med J 1958; 1: 929.

Akinyanju O and Akinsete I. Leg ulceration in sickle cell disease in Nigeria. Trop Geogr Med 1979; 31: 87-91.

Al-Awamy BH, Niazi GA, El-Mouzan MI, AlTorki MT, Naeem MA. Newborn screening for sickle cell haemoglobinopathy and other inherited erythrocytic disorders in the eastern province of Saudi Arabia. Saudi Med J 1986; 7: 502-509.

Al-Awamy BH, Niazi GA, El-Mouzan MI, Al-Torki MT, Naeem MA. Relationship of haemoglobin F and alpha-thalassaemia to severity of sickle cell anaemia in eastern province of Saudi Arabia. Ann Trop Paediatr 1986; 6: 261-265.

Al-Awamy BH, Wilson WA, Pearson HA. Splenic function in sickle cell disease in the eastern province of Saudi Arabia. J Pediatr 1985; 104: 714-717.

Al-Awamy BH, Niazi GA, Al-Mouzam, M.I., Al-Torki, M.T., Naeem M.A. Alpha-thalassaemia and Hb E in the eastern province of Saudi Arabia. Birth Defects: Original Article Series 1988; 23 (5B): 371-376.

Al-Awamy BH, Niazi GA, Naeem MA, Wilson JB, Huisman THJ. Haemoglobin Handsworth x18 (A16) Gly-Arg in a Saudi newborn. Haemoglobin 1985; 9: 183-186.

Al-Awamy BH, Niazi GA, Al-Mouzan MJ, Chen SS, Wilson JB, Webber BB, Huismann THJ. Hb F-Dammam or α2γ2 79(EF)3 Asp- Asn. Haemoglobin 1985; 9: 171-173.

Al-Awamy BH, Niazi GA, Wilson JB, Huisman THJ. Hb Setif alpha 94(GI) aspartic acid - Tyrosine observed in Saudi Arabian family. Haemoglobin 1985; 9: 87-90.

Al-Awamy BH, Niazi GA. Sickle Cell trait in the eastern province of Saudi Arabia: Effect of concurrent α-thalassaemia. Saudi Med J 1987; 8: 253-258.

Al-Awamy BH, El-Mouzan MI, Niazi GA, Al-Torki MT. Sickle cell anaemia in early childhood in the eastern province of Saudi Arabia. General Directorate of Research Grants Program, King Abdulaziz City for Science and Technology, Riyadh (Publication No. 49), 1411 AH (1991 AD).

Al-Fawaz, I., El-Hazmi, M.A.F., Bahakim, H.M. and Warsy, A.S. Cholelithiasis in sickle cell cell disease. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management', 1993 (In press).

Al-Khatti A, Umevera TT, Clow T, et al. Erythropoietin stimulates F reticulocyte formation of sickle cell anaemia. Trans. Assoc. Am. Physician 1989; 101: 54-61.

Alleyne SI, D'Hereux Rauseo R and Serjeant GR. Sexual development and fertility of Jamaican female patients with homozygous sickle cell disease. Arch Intern Med

1981; 141: 1295-7.

Allison AC. Genetic factors in resistance to malaria. Ann NY Acad Sci 1961; 91: 710-729,

Angastionitis M. Cyprus: Thalassaemia program. Lancet 1990; 336: 1119-1120.

Antonarakis SE, Boehm CD, Giardian PJV, Kazazian HH. Non-random association of polymorphic restriction sites in the β-globin gene cluster. Proc Natl Acad Sci USA 1982; 79: 137-141.

Antonarakis SE, Kazazian HH, Orkin SH. DNA polymorphisms and molecular pathology of the human globin gene cluster. Hum Genet 1985: 69: 1-4.

Ashcroft MT and Desai P. Mortality and morbidity in Jamaican adults with sickle cell trait and with normal haemoglobin followed up for twelve years. Lancet 1976; 2: 784-6.

Babiker MA, Taha SA. Two different patterns of sickle cell disease in children in Saudi Arabia. Ann Trop Paedr 1982; 2: 179-181.

Babiker MA, El-Hazmi MAF, Al-Jobori AM, Obeid H, Bahakim HM. Splenic function in children with sickle cell disease: Two different patterns in Saudi Arabia. Scand. J. Haematol 1985; 35: 191-193.

Ballas SK, Castillo R, Gay RN, Adach K, Effects of gender on Hb F level in sickle cell anaemia. Inter Soc Haematol 1992; Abstract: 648-668.

Ballas SK and Smith ED. Blood cell changes during the evolution of the sickle cell painful crises. Blood, 1992; 79: 2154-2163.

Bank A, O'Donnell JV, and Braverman AS. Globin chain synthesis in heterozygotes for beta chain mutations. J Lab Clin Med 1970; 76: 616-21.

Bayoumi RA, Omer A, Samuel APW,Saha N, Sebai ZA, Sabaa HMA. Haemoglobin and erythrocytes glucose-6-phosphate dehydrogenase variants among selected tribes in western Saudi Arabia. Trop Geogr Med 1979; 31: 245-252.

Betke M, Marti HR, Schlicht L. Estimation of small percentage of foetal haemoglobin. Nature 1959; 184: 1877-1878.

Beutler E. Hemolytic anaemia in disorders of red cell metabolism. Plenum Medical Book Company. New York 1978; 23: 168.

Beutler E. Red Cell Enzyme defects. Hemat Pathol 1990; 4: 103-114.

Beutler E. Red cell enzyme defects. Hematologic Pathology 1990; 4(3): 103-114.

Beutler E. Red Cell Metabolism. A Manual of Biochemical Methods. 2nd ed. Grune & Stratton, New York 1975.

Blume KG, Lohr GW, Praetsch O, Rudiger HW, Wendt GG. Beitrag zur populations - genetic der pyruvatkinase menschlicher erythrocyten. Hum Genet 1968; 6: 261.

Bohrer SP. Acute long bone diaphysical infarcts in sickle cell disease. Br. J Radiol 1970; 43: 685-97.

Booker CR, Scott RB, and Ferguson AD. Studies in sickle cell anemia. XXII Clinical manifestations of sickle cell anemia during the first two years of life. Clin Pediatr 1964; 3: 111-5.

Bowman JE (Ed). Distribution and evolution of hemoglobin and globin loci. Elsevier. New York 1983.

Brittenham G, Lozoff B, Harris JW, Kan YW, Dozy AM, Nayud WVS. Alpha globin gene number: Population and restriction endonuclease studies. Blood 1980; 55: 706-708.

Brittenham G, Lozoff B, Harris JW, Mayson SM, Miller A and Huisman THJ. Sickle cell anemia and trait in Southern India: Further studies. Am. J. Hematol 1979; 6: 107-23.

Brittenham G. Genetic model for observed distribution of proportions of haemoglobin in sickle cell trait. Nature 1977; 268: 635-6.

Brozovic M and Anionwu E. Sickle cell disease in Britain. J. Clin Pathol. 1984; 12: 1321-1326.

Burlew K, Evans R, Oler C. The impact of a child with sickle cell disease on family dynamics. Ann. N.Y. Acad. Sci. 1989; 565: 161-171.

Chandrasoma P, Taylor CR. Concise pathology. Appleton and Lange, California, 1993: 373

Charache S, Dover GJ, Mayer MA, Moore JW. Hydroxyurea induced augmentation of fetal haemoglobin production in patients with sickle cell anaemia. Blood 1987; 69: 109-116.

Coa A, Galanello R, Monni G, Olla G. Cossu P, Ristaldi MS. The prevention of thalassaemia in Sardinia. Clin. Genet 1989; 36: 277-282.

Cockshott WP. Dactylitis and growth disorders. Br. J Radiol 1963; 36: 19-26.

Cooper DN, Smith BA, Cooks HJ, et al. An estimate of unique DNA sequence heterozygosity in the human genome. Hum Genet 1985; 69: 210-225.

Cordes W. Experiences with plasmochin in malaria (preliminary reports). 15th Annual Report: United Fruit Co. (Med Dept), Boston, 1926; 66-71.

Dacie JV, Lewis SM. Practical haematology, 6th ed. Churchill Livingstone, Edinburgh, 1991.

Dacie JV and Lewis SM. Practical haematology. 5th ed., Churchill Livingstone, Edinburgh, 1975.

Darnell JE: Variety in the level of gene control in eukaryotic cells. Nature 1982; 297: 365.

DeSimone J, Kleve L, Longley MA and Shaeffer J. Rapid turnover of newly-sythesized β^S chains in retoculocytes from individuals with sickle cell traint. Biochem Biophys Res Comm 1974; 57: 248-54.

DeSimone J, Adams JG 3d, Shaeffer J. Evidence for rapid loss of newly synthesized haemoglobin S molecules in sickle cell anaemia and sickle cell trait. Br. J. Haematol 1977; 35: 373-385.

Dierks P, Van Ooyen A, Cochran MD, Dobkin C, Reiser J, Weissman C: Three regions upstream from the cap site are required for efficient and accurate transcription of the rabbit β-globin gene in mouse 3 T6 Cells. Cell 1983; 32: 695.

Diggs LW and Ching RE. Pathology of sickle cell anemia. Southern Med J 1934; 27: 839-45.

Diggs LW. Sickle cell crises. Am J Clin Pathol 1965; 44: 1-19.

Dincol G, Aksoy M, Erdem S. Beta-thalassaemia with increased haemoglobin A_2 in Turkey. Study in 164 thalassaemia heterozygotes. Hum. Hered. 1979: 29:272.

Dover GJ, Humphries PK, Moore JG, Ley NS, Charache SH, Nienhuis AW. Hydroxyurea between cytotoxicity and F cell production. Blood 1986; 67: 735-738.

Economidou J, Constantoulakis M, Angustaki O, et al. The incidence of hepatitis associated antigen and antibody in patients with thalassaemia in Greece. Vox Sang 1970; 19: 401.

Edelstein SJ, Telford JN et al. Structure of fibers of sickle cell hemoglobin. Proc Natl Acad Sci USA 1973; 70: 1104.

Edwards CRW, Bouchier IAD, Davidson's principle and practice of medicine. 16th edition, Churchill Livingstone, London, 1991.

Efremov GD, Wilson JB, Huisman THJ. The clinical heterogeneity of human hemoglobin F: Direct evidence for the existence of three types of -chains, the IG, IA, TA chains. Biochim. Biophys Acta 1979; 579: 421.

Efstratiadis A, Posakony JW, Maniatis T, Lawn RM, O'Connell C et al. The structure and evolution of the human β-globin gene family. Cell 1980; 21: 653.

Eisenstein MI, Posner AC and Friedman S. Sickle cell anemia in pregnancy. A review of the literature with additional case histories. Am J Obstet Gynecol 1956; 72: 622-34.

El-Hazmi, M.A.F. and Lehmann, H. Haemoglobin Riyadh $\alpha_2\beta$]120) $_2$GH3[Lys _ Asn) - A new variant found in association with α-thalassaemia and iron deficiency. Hemoglobin 1976-77; 1(1): 59-74.

El-Hazmi, M.A.F. and Lehmann, H..Human Haemoglobins and Haemoglobinopathies in Arabia: Hb O Arab in the Saudi Arabia. Acta haemat. 1980; 63: 268-273.

El-Hazmi, M.A.F. Human Haemoglobin and Haemoglobinopathies in the Arabian Peninsula - Studies at the Molecular Level. Proceedings of the 4th Saudi Medical Meeting, Dammam. 1980; pp 317-324.

El-Hazmi, M.A.F. The Red Cell Genetics and Environmental Interactions - A Tehamat--Aseer Profile. Saudi Medical Journal 1985; 6(2): 101-112.

El-Hazmi, M.A.F. Incidence and Frequency of Haemoglobinopathies and Thalassaemia in the North-West Sector of Arabia. Saudi Medical Journal 1985; 6(2): 149-162.

El-Hazmi, M.A.F. The Distribution and nature of hemoglobinopathies in Arabia. In: William P. Winter (Ed.) Hemoglobin Variants in Human Populations. CRC Press Inc. Boca Raton, Florida. 1987; pp 65-77.

El-Hazmi, M.A.F. Haemoglobinopathies and Allied Disorders in Saudi Arabia. Proc. CMRC Sym. Appl. M.R., Riyadh 1987. pp. 35-47.

El-Hazmi, M.A.F. Haemoglobinopathies, Thalassaemias and Enzymopathies in Saudi Arabia: The Present Status. Acta haemat. 1987; 78: 130-134.

El-Hazmi, M.A.F. Studies on Sickle Cell Heterozygotes in Saudi Arabia - Interaction with α-Thalassaemia. Acta haemat. 1986; 75: 100-104.

El-Hazmi, M.A.F. and Warsy, A.S. Glucose-6-phosphate Dehydrogenase Deficiency - Genetic, Pathophysiologic and Therapeutic Aspects. Annals of Saudi Medicine 1985; 5(4): 213-223.

El-Hazmi, M.A.F. Infections in sickle cell disease. Annals of Saudi Medicine 6(1): 33-41.

El-Hazmi, M.A.F. Clinical manifestation and laboratory findings of sickle cell anaemia in association with α-thalassaemia in Saudi Arabia. Acta haemat. 1985; 74: 155-160.

El-Hazmi, M.A.F. and Warsy, A.S. Glucose-6-Phosphate Dehydrogenase Deficiency in Blood Donors. A Need for Reassessment. Saudi Medical Journal 1986; 7(3): 250-254.

El-Hazmi MAF, Jabbar FA, Al-Faleh FZ, Al-Swailem AR, Warsy AS. The haematological, biochemical and clinical presenta tion of haemoglobin S in Saudi Arabia (i) Haematological and clinical expression. Trop Geogr Med 1987; 39: 157-162.

El-Hazmi MAF, Jabbar FA, Al-Faleh FZ, Al-Swailem AR Warsy AS. The haematological, biochemical and clinical presentation of haemoglobin S in Saudi Arabia (ii) Biochemical Expression. Saudi Med J 1988; 9(1): 66-71.

El-Hazmi MAF, Bahakim HM, Al-Swailem AM, Warsy AS. The features of sickle cell disease in Saudi children. J Trop Paedr 1990; 36: 148-155.

El-Hazmi MAF, Warsy AS. On the nature of sickle cell disease in the south-western province of Saudi Arabia. Acta haemat 1986; 76: 212-216.

El-Hazmi MAF, Al-Swailem AR, Jabbar FA, Al-Faleh FZ, Warsy AS. Effect of thalassaemia, G-6-PD deficiency and Hb F on the nature of sickle cell disease in south-western Saudi Arabia. Trop Geogr Med 1990; 42: 241-247.

El-Hazmi MAF, Bahakim HM, Babiker MA, Al-Swailem AR, Warsy AS. Symptom-free intervals in sicklers - Does pneumococcal vaccination and pencillin prophylaxis have a role? J Trop Pediatr 1990; 36: 56-62.

El-Hazmi MAF, Warsy AS. Glucose-6-phosphate dehydrogenase deficiency in blood donors. A need for reassessment. Saudi Medical Journal 1986; 7(3): 250-254.

El-Hazmi MAF, Al-Swailem AR, Al-Faleh FZ, Warsy AS. Frequency of glucose-6-phosphate dehydrogenase, pyruvate kinase and hexokinase deficiency in the Saudi population. Hum Hered 1986; 36: 45-49.

El-Hazmi MAF, Warsy AS. Glutathione reductase deficiency in association with sickle cell and thalassaemia genes in Saudi population. Hum Hered 1985; 35: 326-332.

El-Hazmi MAF, Warsy AS. Aspects of sickle cell gene in Saudi Arabia - interaction with glucose-6-phosphate dehydrogenase deficiency. Hum Genet 1984; 68: 320-323.

El-Hazmi MAF, Warsy AS. Interaction between glucose-6-phosphate dehydrogenase deficiency and sickle cell gene in Saudi Arabia. Trop geogr Med 1987; 39: 32-35.

El-Hazmi MAF, Warsy AS. The effect of glucose-6-phosphate dehydrogenase deficiency on the haematological parameters and clinical manifestations in patients with sickle cell anaemia. Trop geogr Med 1989; 41: 52-56.

El-Hazmi MAF, Warsy AS. Beta-globin gene polymorphism in Saudis: 5.6 Hpa I fragment. Hum Hered 1987; 37: 237-240.

El-Hazmi MAF, Jabbar FA, Al-Swailem AR, Warsy AS. β-globin gene polymorphism in Saudis - Triple Hpa I fragments. Hum Genet 1986; 74: 313-315.

El-Hazmi MAF. β-globin gene haplotypes in Saudi sickle cell anaemia patients. Hum Hered 1990; 40: 177-186.

El-Hazmi MAF. β-Globin gene polymorphism in the Saudi Arab population. Hum Genet 1986; 73: 31-34.

El-Hazmi MAF. α-thalassaemia in Saudi Arabia: deletion pattern. Hum Genet 1987; 76: 196-198.

El-Hazmi MAF. Abnormal hemoglobins and allied disorders in the Middle East -Saudi Arabia. In: James E. Bowman (ed.). Distribution and Evolution of hemoglobin and Globin Loci. Elsevier Science Publishing Co. Inc. New York, 1983; pp 239- 249.

El-Hazmi MAF. Haemoglobin disorders: A pattern for thalassaemia and haemoglobinopathies in Arabia. Acta haemat 1982; 68: 43-51.

El-Hazmi MAF. Human haemoglobin and haemoglobinopathies in the Arabian Peninsula - Studies at the molecular level. Proceedings of the 4th Saudi Medical Meeting, Dammam. 1980; pp 317-324.

El-Hazmi MAF. Infections in sickle cell disease. Ann Saudi Med 1986; 6(1): 33-41.

El-Hazmi MAF. On the nature of sickle-cell disease in the Arabian Peninsula. Hum Genet. 1979; 52: 323-335.

El-Hazmi MAF. Some aspects of the sickle cell gene in Saudi Arabia. Proceedings of the First International Conference on Sickle Cell Disease: A World Health Problem. Bethesda, Mary land. 1976; pp 64-66.

El-Hazmi MAF. The clinical and haematological expression of HbS in Saudis. Blood 1990; 76(10) Suppl 1: 59a.

El-Hazmi, M.A.F. Haemoglobinopathies, thalassaemias and enzymopathies in Saudi Arabia Saudi Medical Journal 1992; 13(6): 488-499.

El-Hazmi, M.A.F., Warsy, A.S. and Bahakim, H. The Blood Proteins C&S in the Sickle Cell Disease. Acta haematologica.

El-Hazmi, M.A.F. and Warsy, A.S. Iron-Endocrine Patterin in Patients with β-Thalassaemia. Journal of Tropical Medicine.

El-Hazmi MAF, Lehmann H. Interaction between iron deficiency and α-thalassaemia: the in vitro effect of haemin on α-chain synthesis. Acta haemat 1978; 60: 1-9.

El-Hazmi MAF and Warsy AS. A normal reference range for erythrocyte glucose-6-phosphate dehydrogenase in a Saudi population. Med Lab Sci 1987a; 44: 125-129.

El-Hazmi MAF and Warsy AS. The normal range for red cell pyruvate kinase in the Saudi population. Saudi Med J 1987b; 8(3): 228-237.

El-Hazmi MAF. Thyroxine carriage and haemoglobinopathies as example of molecular pathology. Ph.D. thesis, University of Cambridge, Cambridge, U.K. 1977

Embury SH, Miller JA, Dozy AM, Kan YM, Chan V, Todd D. Two different molecular organization account for the single α-globin gene of the α-thalassaemia.2 genotye. J Clin Invest 1980; 66: 1319-1329.

Embury SH, Gholson MA, Gillette P, Rieder RF and the National Cooperative Study of Sickle Cell Disease. The leftward deletion α-thalassaemia.2 haplotype in a black subject with haemoglobin SS. Blood 1985; 65: 769-771.

Embury SH, Dozy AM. Correlation of α-globin phenotype with haematological parameters in sickle cell trait. Blood ((Supp.I) 1979; 54: 53.

Embury SH, Andree M, Dozy MT, Miller J, et al. Concurrent sickle cell anaemia and -thalassaemia . New Engl. J. Med 1982; 306: 270-274.

England TM and Frazer PM. Differentiation of iron deficiency from thalassaemia trait by routine blood count. Lancet 1973; i: 449.

Ennis JT, Serjeant GR and Middlemiss JH. Homozygous sickle cell disease in Jamaica. Br. J. Radiol. 1973; 46: 943-50.

Fabricius E and Rejewsky MF. Determination of hydroxyurea in mammalian tissues and blood. Rev. Europ. Eludes. Clin. et Biol. 1971; XVI; 679-683.

Feldenzer J, Mears JG, Burns AL, Natta C, Bank A. Heterogeneity of DNA fragment associated with the sickle globin gene. J Clin Invest 1979; 64: 751-755.

Felice A, Abraham FC, Miller A, Cope N, Gravery M, Huisman THJ. Post-transitional

control of human haemoglobin synthesis in the cell. New York: Liss 1978; 131-53.

Finch JT, Perutz MF et al. Structure of sickled erythrocytes and of sickle cell hemoglobin fibers. Proc Natl Acad Sci USA 1973; 70: 718.

Galanello R, Melis MA, Rugger R, Addis M, et al. °-thalassaemia trait in Sardinia. Hemoglobin 1979; 3: 33.

Garcia SC, Moragon AC, Lopez-Fernandez ME. Frequency of pyruvate kinase and glucose-6-phosphate dehydrogenase deficiency in Spanish population. Hum Hered 1979; 29: 310-313.

Gaston MH. Sickle cell disease: An overview. Semin Roentgenol 1987; 3: 150-159.

Gelpi AP. Glucose-6-phosphate dehydrogenase deficiency in Saudi Arabia: A Survey. Blood 1965; 25: 486.

Gelpi AP. Glucose-6-phosphate dehydrogenase deficiency, the sickling trait and malaria in Saudi Arabian children. J Paediatr 1967; 71: 138-146.

Gelpi AP. Migrant populations and the diffusion of the sickle cell gene. Ann. Intern. Med. 1973; 79: 258-264.

Gelpi AP. Sickle cell disease in Saudi Arabia. Acta haematol 1970; 43: 89-99.

Goldberg MA, Brunara C, Dover GJ, et al. Treatment of sickle cell anaemia with hydroxyurea and erythropoietin. New Engl J Med 1990; 323: 366-372.

Golding JSR, Maclver JE and Went LN. The bone changes in sickle cell anaemia and its genetic variants. J Bone Jt Surg 1959; 41B: 711-8.

Gonttas A, Fessas P, Tsevrenis H, Xefteri E. Description d'unenouvelle variate d'anaemic hemolytique congenitale (Elude hematologique, electrophoretique et genetique Sang. 1955; 26: 911.

Graham C, Maude GH and Serjeant GR. Delayed menarche in homozygous sickle cell disease. W Ind Med J 1986; 35: 18-22.

Hardisty RM, Weatherall DJ (eds). Blood and its disorders. 2nd edition. Mosby, 1982.

Harris JW. Studies on the destruction of red blood cells. VIII. Molecular orientation in sickle cell hemoglobin solutions. Proc Soc Exp Biol Med 1950; 75: 197.

Herrick JB. Pecular elongated and sickle-shaped and red blood corpuscles in a case of severe anemia. Arch Intern Med 1910; 6: 517-21.

Heywood JD, Karon M, Weissman S. Studies of in vitro synthesis of heterogenic hemoglobins. J. Clin. Invest. 1964; 43: 2368-2376.

Higgs DR, Pressley L, Clegg JB, Weatherall DJ, Serjeant DR. α-thalassaemia in Black populations. Johns Hopkins Med J 1980; 146: 300-310.

Higgs DR, Wood WG, Jarman AO, Vickers MA, Wilkie AOM, Lamb J, Vyas P, Bennett JP. The α-thalassaemias. Ann N.Y. Acad Sci 1990; 612: 15-22.

Higgs DR, Aldridge BE, Lamb J, Clegg JB, et al. The interaction of alpha-thalassaemia and homozygous sickle cell disease. New Engl. J. Med. 1982; 306: 1441-1446.

Huisman THJ, Trimodality in the percentage of -chain variant in heterozygotes: the effect of the number of active Hb and structural loci. Hemoglobin 1977; 1: 349-382.

Huisman THJ, Schroeder WA, Reese A, Wilson JB, et al. The T chain of human fetal hemoglobin at birth and in several abnormal hematological conditions. Pediatr. Res. 1977; 11: 1102-1105.

Huraib, S., Al-Momen, A.K., El-Hazmi, M.A.F., Al-Harakati, M., Warsy, A.S., Bahakim, H.M. and Fawaz, I. The pattern of renal involvement in sickle cell disease. Proceedings of the symposium on `Blood Genetic Disorders: Pathophysiology and Management' 1993 (In press).

Ingram VM. Gene mutations in human haemoglobin. The chemical difference between normal and sickle cell haemoglobin. Nature (London) 1957; 180: 326.

Ingram VM. Hemoglobin and its abnormalities. Monograph in American Lecture Series. Springfield. Ill Charles C Thomas. Publishers 1961.

Itano HA. Quantitative and qualitative control of adult haemoglobin synthesis - a multiple allele hypothesis. Am J Hum Genet 1953; 5: 34-45.

Jeffrey AJ. DNA sequence variants in the G, A, and -globin genes of man. Cell 1979; 18: 1-10.

Kaine WN. Sickle cell anaemia in children in Eastern Nigeria. A detailed analysis of 210 cases. East Afr. Med. J. 1982; 59: 742-749.

Kamel K and Awny AY. Origin of the sickling gene. Nature 1965; 16: 428-30.

Kan YW, Dozy AM. Polymorphism of DNA sequence adjacent to human -globin structural gene: relationship to sickle mutation. Proc Natl Acad Sci USA 1978; 75: 5631-5635.

Kan YW, Dozy AM. Evolution of the haemoglobin S & C genes in world populations. Science 1980; 209: 388-391.

Kattamis C, Ladis V. Metaxatou-Mavromati A. Hemoglobin F & A2 in Greek patients with homozygous β and β/δβ thalassaemia in abnormal haemoglobins and thalassaemia: Diagnostic Aspects (ed. R.M. Schmidt). Academic Press, New York p 209.

Kazazian HH, Antonarakis SE. The varieties of mutation. In: Childs B, Holtzman NA, Kazazian HH, Valle DL. Molecular genetics in Medicine. Elsevier New York 1988 43-67.

Kazazian HH, Boehm CD. Molecular basis and prenatal diagnosis of β-thalassaemia. Blood 1988; 72: 1107-1116.

Kazazian HH Jr, Dowling CE, Boehm CD, Warran TC, Economous EP, Katz J, Antonarakis SE. Gene defects in β-thalassaemia and their prenatal diagnosis. Ann N. Y. Acad Sci. 1990; 612: 1-14.

Kazazian HH Jr, Orkin SH, Antonarakis SE, Cheng T-C, Sexton JP. β-thalassaemia mutations differ among various ethnic groups. Blood 1983; 62: 68.

Kazazzian HH Jr. The thalassaemia syndromes: Molecular basis and prenatal diagnosis in 1990. Semin Hematol 1990; 27: 209-228.

Kekwick RA, Lehmann H. Sedimentation characteristics of the -chain haemoglobin (haemoglobin 'Barts'). Nature (Lond.) 1960; 187: 158-

Khashogi T. Effects of maternal sickle cell trait on fetal outcome. Symposium on Blood genetic disorders pathophysioogy and management. 15-17 Shawwal, 1412 (18-20 April, 1992), College of Medicine, King Saud University, Riyadh.

Knox-Macauley HHM, Weatherall DJ, Clegg JB, Pembury ME. Thalassaemia in the British. Br. Med J 1973; iii: 150.

Konotey-Ahulu FD. Effect of environment on sickle cell disease in West Africa, epidemiologic and clinical consideration. In: Abraham H, Bertles JF, Wethers DL (eds). Sickle cell disease, diagnosis, management, education and research. St. Louis, Mosby, 1973.

Konotey-Ahulu FID. Patterns of clinical haemoglobinopathy. E Afr. Med J 1969a; 46: 118-9.

Konotey-Ahulu FID. The sickle cell diseases. Arch Intern Med 1974; 133: 611-619.

Konotey-Ahulu. Anaesthetic deaths and the sickle cell trait. Lancet 1969b 1:267-8.

Konotey-Ahulu. Computer assisted analysis of data on 1,697 patients attending the sickle cell/haemoglobinopathy clinic of Korle Bu Teaching Hospital, Accra, Ghana. Clinical features. I. Sex, genotype, age, rheumatism and dactylitis frequencies. Ghana Med J 1971; 10: 241-60.

Koshy M, Ensuah R, Koranda A, et al. Leg ulcers in patients with sickle cell disease. Blood 1989; 74: 1403-1408.

Kumar RJ and Clark M (ed). Clinical Medicine. A text book for medical students and doctors. ELSS London 1991.

Kunkel LM, Smith KD, Boyer SH et al. Analysis of human Y-chromosome specific reiterated DNA in chromosome variants. Proc Natl Acad Sci USA 1977; 74: 1245-1247.

Lambotte C. Hand-foot syndrome in sickle cell disease. Am J Dis Child 1962; 104: 200-1.

Lauer J, Shen C-KJ, Maniatis T. The chromosomal arrangement of human α-like globin genes. Sequence homology and α-globin gene deletions. Cell 1980; 20: 119.

Lawn RM, Efsratiadis A, 0'Connel C, Maniatis T. The nucleotide sequence of the human β-globin gene. Cell 1980; 21: 647-651.

Leary JJ, Brigati DJ, Ward DC. Rapid and sensitive colorimetric method for visualizing biotin-labeled DNA probes hybridized to DNA or RNA immobilized on nitrocellulose. Blo-blots. Proc Natl Acad Sci USA 1983; 80: 4045-4049.

Lehmann H, Marangin G, Mourant AE. Distribution of haemoglobin in Saudi Arabia. Nature 1963; 198: 492-493.

Lehmann H, Huntsman RG. An introduction to the structure and function of haemoglobin. Clinic in Haematology 1974; 3(2): 217-223.

Lehmann H, Huntsman RG. Man's Haemoglobins. North Holland Publishing Company. Amsterdam 1966.

Lehmann H. Distribution of the sickle cell gene. A new light on the origin of the East Africans. Eugencis Review 1954a; 46: 101-21.

Lehmann H. Origin of the sickle cell. S. Afr J Sci 1954b; 50: 140-1.

Lie-Injo LE, Solai A, Herrera AR, Nicohaisen L, Kan YW, Wan WP, Hasan K. Hb Barts level in cord blood and deletion of α-globin genes. Blood 1982; 59: 370-376.

Lie-Injo LE, Lie HG, Ager JAM, Lehmann H. α-thalassaemia as a cause of hydrops foetalis. Br. J. Haematol 1962; 8:1.

Lie-Injo LE and Jo BH. A fast moving haemoglobin in hydrops foetalis. Nature 1960a 185: 698.

Lie-Injo LE and Jo BH. Hydrops foetalis with fast-moving haemoglobin. Br. Med.. J.

1960b; ii: 1649.

Liebhaber SA, Goossens M, Poon R, Kan YM. The primary structure of the α-globin gene cloned from normal human DNA. Proc Natl Acad Sci USA 1980; 77: 7054-7058.

Leibhaber SA, Grossens M, Kan YM. Homology and concerted evolution at the is α_1 and α_2 loci of human α-globin. Nature 1981; 290: 26-29.

Livingston FB. Abnormal hemoglobins in human populations. Aldine Publishing Company, Chicago 1967.

Livingstone FB. The malaria hypothesis. In JE Bowman (ed.) Distribution and Evolution of Hemoglobin & Globin Loci. Elsevier, New York 1983.

Luzzatto L, Usanga EA, Reddy S. Glucose-6-phosphate dehydrogenase deficient red cells: resistance to infection by malarial parasites. Science 1969; 164: 839-842.

Mackey JP and Vivarelli F. Tanganyika Annual Report of the Medical Laboratory. cited in Mackey, J.P. and Vivarelli F (1954).

Malamos B, Fessas P and Stamatoyannopoulos G. Types of thalassaemic trait carriers as revealed by a study of their incidence in Greece. Br. J. Haematol 1962; 8: 5.

Malaria Control Program in the Kingdom. Malaria Control Services, Ministry of Health, Kingdom of Saudi Arabia. Bahr Al-Olum Press, Riyadh, 1983.

Maniatis T, Fritsch EF, Lauer J, Lawn RM. The molecular genetics of human hemoglobins. Ann Rev Genet 1980; 14: 145-178.

Maniatis T, Fritsch EF, Lauer J, Lawn RM, Proudfoot NJ, Shander HM, Shen C-KJ. The structure and chromosomal arrangement of human globin genes. In: Stamatoyannopoulos G, Nienhuis AW (ed). Organization and Expression of globin genes. Alan R Liss Inc New York 1981: 15-31.

Marengo-Rowe AJ. Rapid electrophoresis and quantitation of haemoglobin on cellulose acetate. J Clin Pathol 1965; 18: 790-792.

Marinone G and Bernasconi C. Studio electroforetico quantitative su blocco d'amido' dellemoglobina normale and dei talassemici. Haemotologica 1957; 42: (Supp.I)

Mazza U, Saglio G, Ceppio FC, Camaschella E, et al. Clinical and haematological data in 254 cases of beta-thalassaemia trait in Italy. Br. J. Haematol 1976; 33: 91.

Mentzler WC. Differentiation of iron deficiency from the thalassaemia trait. Lancet 1973; i: 882.

Modell CB and Berdoukas. The clinical approach to thalassaemia. Grune & Stratton, New York, 1981.

Modell CB. Management of thalassaemia major. Br. Med. Bull. 1976; 32: 270.

Myers RM, Maniatis T, Lerman LS. Detection and localization of single base changes by denaturing gradient gel electrophoresis. Methods Enzymol 1987; 155: 501.

Myers RM, Lumelsky N, Lerman LS, Maniatis T. Detection of single base substitutions in total genomic DNA. Nature 1985; 9:313: 495

Myers RM, Fischer SG, Lerman LS, Maniatis T. Nearly all single base substitutions in DNA fragments joined to a GC-Calmp can be detected by denaturing gradient gel electrophoresis. Nucleic Acads Res 1985b; 13: 3131-Nienhuis AW, Angnou NP, Ley TJ. Advances in Thalassaemia Research. Blood 984; 63: 738-758.

Pembrey ME, Weatherall DJ, Clegg JB, Bunch C, Perrine RP. Haemoglobin Barts in Saudi Arabia. Br J Haemat 1975; 29: 221-234.

Pembrey ME, Weatherall DJ, Perrine RP, Clegg JB. Population with benign sickle cell disease. Model for management of sickling disorders. Quart J Med 1975; 44: 633-634.

Pembrey ME, Wood WG, Weatherall DJ, Perrine RP. Fetal haemoglobin production and the sickle cell gene in the oasis of eastern Saudi Arabia. Br J Haemat 1978; 40: 415-429.

Pembrey ME, Perrine RP, Wood WG, Weatherall DJ. Sickle (beta S) thalassaemia in eastern Saudi Arabia. Am J Hum Genet 1980; 32: 26-41.

Perrine RP, Pembrey ME, John P, Perrine S, Shoup F. Natural history of sickle cell anaemia in Saudi Arabia. Ann Intern Med 1978; 88: 1-6.

Perrine RP, Brown MJ, Clegg JB, Weatherall DJ, May A. Benign sickle cell anaemia. Lancet 1972; ii: 1163-1167.

Perrine RP, John P, Pembrey M, Perrine S. Sickle cell disease in Saudi Arabs in early childhood. Arch Dis Childhood 1981; 56: 187-192.

Perrine RP. Cholelithiasis in sickle cell anemia in a Caucasian population. Am J Med 1973; 54: 327-32.

Perutz MF, Lehmann H. Molecular pathology of human haemoglobin. Nature 1968; 219: 902.

Pongpipat D, Suvette V, Assateerawatt A. Host defence in thalassaemia and the effect of splenectomy. 1. incidence of hepatitis B surface antigen and antibody. J. Med Assoc. Thailand 1978; 61: 75.

Poortrakul P, Wasi P and Na-Naorn S. Haematological data in 312 cases of -thalassaemia trait in Thailand. Br. J. Haematol 1973; 24: 703.

Portnoy BA and Herion JC. Neurological manifestations in sickle cell disease. Ann Intern Med 1972; 76: 643-52.

Powars D, Wilson B, Imbus C, Pegelow C and Allen J. The natural history of stroke in sickle cell disease. Am J Med 1978; 65: 461-71.

Powars DR, Weiss JN, Chan LS, Schroeder WA. Is there a threshold level of fetal hemoglobin that ameliorates morbidity in sickle cell anemia. Blood 1984; 63: 921-926.

Powars DR. The natural history of sickle cell disease in the first ten years. Semin Hematol 1975; 12: 267-285.

Pressley L, Higgs DR, Clegg JB, Perrine RP, Pembrey ME, Weatherall DJ. A new genetic basis for hemoglobin H disease. N Engl J Med 1980; 303: 1383-1388.

Proudfoot JN, Maniatis T. The structure of a human α-globin pseudogene and its relationship to α-globin gene duplication. Cell 1980; 21: 537.

Proudfoot NJ, Brownlee GG. 3' Non-coding region sequences in eukaryotic messenger RNA. Nature 1976; 263: 211.

Reid HL, Barnes AJ, Lock PJ, Dromandy JA, Dormandy TL. A simple method for measuring erythrocyte deformability. J. Clin. Invest. 1977; 855: 858.

Robinson AR, Robson M, Harrison AP, Zuelzer WW. A new technique for the differentiation of hemoglobin. J Lab Clin Med 1957; 50: 745-752.

Rodgers GP, Dover G.J. Noguchi CJ, Schechter AN, Nienhuis AW. Hemotologic responses of patients with sickle cell disease to treatment with hyroxyurea. New Engl J Med 1992; 322: 1037-1045.

Rodgers GP. Recent approaches to the treatment of sickle cell anaemia. JAMA Middle East 1992: 53-57.

Salditt-Georgieff M, Darnell JE: A precise termination site in the mouse β major transcription unit. Proc Natl Acad Sci USA 1983; 80: 4694.

Schanfield MS, Scalaise G, Economidou I, Modell CB, et al. Immunogenetic factors in thalassaemia and hepatitis B infection. A multicentre study. Dev. Biol. Stand. 1975; 30: 257.

Schechter AN, Noguchi CT, Rodgers GP. Sickle cell anaemia. In: Stamatoyannopoulos G, Niehnuis AW, Leder P, Majerus PW (eds). The molecular basis of blood diseases. Philadelphia PA, W.B. Saunders Co. 1987; 179-218.

Schroeder WA, Huisman THJ, Shelton JR, et al. Evidence for multiple structural genes for the chains of human fetal hemoglobin. Proc. Natl. Acad. Sci. USA 1968; 60: 537-544.

Sears DA, Kraus AP. Avascular necrosis of the hip in the sickle cell hemoglobinopathies. J Canad Ass Radiol 1983; 34: 136-9.

Sears DA. the morbidity of sickle cell trait. A review of the literature. Am J Med 1978; 64: 1021-36.

Sebes JI and Kraus AP. Avascular necrosis of the hip in the sickle cell hemoglobinopathies. J. Canad. Ass. Radiol. 1983; 34: 136-9.

Sejeny SA, Ganeshaguru K, Acquaye JK, Agyei-Obese S, Samuel APW, Omer A. Level of Hb A_2 and fetal haemoglobin in some alpha and beta thalassaemia heterozygous states. Saudi Med J 1986; 7: 355-362.

Serjeant GR, Grandison Y, Lowrie Y, Mason K, Phillips J, Serjeant BE and Vaidya S. The development of haematological changes in homozygous sickle cell disease: a cohort study from birth to 6 years. Br J Haematol 1981; 48: 533-43.

Serjeant GR. Leg ulceration in sickle cell anemia. Arch Intern Med 1964; 133: 690-4.

Serjeant GR. Sickle cell disease. Oxford Medical Publication. Oxford 1988: pp 349.

Sheffield VC, Cox DR, Lerman LS, Myers RM. Attachment of a GC-Clamp to genomic DNA fragments by the polymerase chain reaction results in improved detection of single base changes. Proc Natl Acad Sci 1989; 86: 232-

Shelton JB, Shelton JR,Schroeder WA. High performance liquid chromatographic separation of globin chains on a large-pore C4 column. J Liquid Chromatography 1984; 7: 1969-1977.

Shine I, & Lal S. A strategy to detect β-thalassaemia minor. Lancet 1977; 1: 692-694.

Siegling JA. In discussion of paper by Chung SMK and Ralston EL presented at Annual Meeting of Am. Orthop. Ass. Colorado Springs, May 23-6, 1966.

Silvertroni E, Bianco I, Muzzolin M, et al. Studio biochimico, elettroforetica and spettroto-metrico dell'emoglobina de malati di anemic microcitica Costituzionale and dimorbo di cooley. Experientia 1957; 13: 705.

Sorrentino B, Ney P, Bodine D, Nienhuis AW. A 46 bp enhancer sequence within the locus activating region is required for induced expression of the γ-globin gene during erythroid differentiation. Nucleic Acids Res. 1990; 18: 2721-2733.

Southern EM. Detection of specific sequences among DNA fragment separated by electrophoresis. J Mol Biol 1975; 98: 503-517.

Stamatoyannopoulos MD, Nienhuis A (ed). The regulation of haemoglobin switching The John Hopkins University Press, London, 1991.

Stetson CA Jr. The state of hemoglobin in sickled erythrocytes. J Exp Med 1966; 123: 341.

Stevens CE, Silbert JA, Miller DR, Dienstag TL, et al. Serologic evidence of hepatitis A & B virus infections in thalassaemia patients on a retrospective study. Transfusion. 1978; 18: 356.

Stevens MCG, Padwick M and Serjeant GR. Observations on the natural history of dactylitis in homozygous sikcle cell disease. Clin Pediatr 1981; 20: 311-7.

Tanaka KR, Clifford GO and Axelrod AR. Sickle cell anemia (homozygous S) with aspetic necoris of femoral head. Blood 1956; 11: 998-1008.

Ten State Nutrition Survey, 1968-1970, DHEW Publication No. (HSM) 72-8134 Vol. I-V, Washington, 1970.

Troungos C, Krishnamoorthy R, Lambardo T, Sortino G, Cacciola E, Labie D. A leftward deletion α^{+}-thalassaemia found in East Sicily in conjunction with heterozygous β-thalassaemia. Hum Genet 1984; 67: 216-218.

Tuck SM, Studd JWW and White JM. Pregnancy in women with sickle cell trait. Br J Obstet Gynaecol 1983; 90: 108-11.

Umemura T, Papayannopoulos TH, Stamatoyannopoulos G. Fetal hemoglobin synthesis in vivo: Direct evidence for control at the level of erythroid progenitors. Proc. Natl. Acad. Sci, USA 1988; 85: 9287.

Vierucci A, London WT, Blumberg BS, et al. Australia antigen and antibody in transfused children with thalassaemia. Arch. Dis Child 1972; 47: 760.

W.H.O. Working Group on Community Control of Hereditary Anaemias, W.H.O. Bulletin, 1982; 60: 643.

Warsy AS, El-Hazmi MAF. Bacterial infections and the sickling disorders - A review. Saudi Med J 1987; 8(3): 228-237.

Warsy AS, El-Hazmi MAF. Glucose-6-phosphate dehydrogenase deficiency in Saudi Arabia - A Review. Saudi Medical Journal 1987; 8(1): 12-20.

Warsy AS, El-Hazmi MAF. Molecular therapy of sickle cell anaemia - State of art. Saudi Med J 1985: 6(3): 257-263.

Warsy AS. Frequency of glucose-6-phosphate dehydrogenase deficiency in sickle cell disease. A study in Saudi Arabia. Hum. Hered. 1987; 35: 143-147.

Weatherall DJ, Clegg JB. The Thalassaemia syndrome (3rd edn.). Blackwell Scientific Publications, Oxford 1981.

Weatherall DJ, Fiorelli G, Gorini S (ed). Advances in red blood cell biology. New York Raven Press 1982: 297-308.

Weatherall DJ, Clegg JB, Blankson J, McNeil JR. A new sickling disorder resulting from interaction of the genes for haemoglobin S and thalassaemia. Br J Haemat 1969; 17: 517-526.

Went LN, Maclver JE. Thalassaemia in the West Indies. Blood 1961; 17-166.

White JG. The structure of sickled hemoglobin in situ. Blood 1968; 31: 561.

Wilimas J, Goff JR, Anderson HR, Langston JW and Thompson E. Efficacy of transfusion therapy for one to two years in patients with sickle cell disease and cerebrevascular accidents. J Pediatre 1980; 96: 205-8.

Williams WJ, Beutler E, Erslev AJ, Lichtman MA. Textbook of Hematology. McGraw Hill Book Co. New York 1983

Winter WP (Ed). Hemoglobin variants in human population. CRC Press, Boca Baton 1987.

Wolfort FG, and Krizek TJ. Skin ulceration in sickle cell anemia. Plast Reconstr. Surg 1969; 43: 71-7.

Wong SC, Ali MAM, Boyadjian SE. Sickle cell trait in Canada, Trimodel distribution of Hb S as a result of interaction with α-thalassaemia gene. Acta Haemat 1981; 65: 159-163.

Wood DH. Cerebrovascular complications of sickle cell anaemia. Stroke 1978; 9: 73-75.

Wood GW, Pembrey ME, Serjeant GR, Perrine RP, Weatherall DJ. Hb F synthesis in sickle cell anaemia: a comparison of Saudi Arab cases with those of African origin. Br J Haemat 1980; 45: 431-445.

World Health Organization - Technical Report No. 509. Treatment of haemoglobinopathies and allied disorders. Geneva, W.H.O, 1972.

World Health Organization Scientific Group. Haemoglobinopathies and allied disorders. Technical Report Series 338, Geneva. World Health Organization, 1966.

World Health Organization Working Party on the Community Control of Hereditary Anaemias 1985; Update of the progress of Haemoglobinopathies Control.

World Health Organization. Community approaches to the control of hereditary disease. Geneva, Oct. 1985.

World Health Organization. Report of a W.H.O. meeting on advances in diagnosis, treatment and prevention of hereditary diseases. Geneva No. 1989.

World Health Organization. Report of the Vth Annual Meeting of the W.H.O. Working Group on the feasibility study on Hereditary Disease Community Control Programme. (Hereditary Anaemias: Alpha Thalassaemias). Herakleion (Crete), Oct. 1987.

CHAPTER 18

APPENDICES

16. 1 Equipment used during the period of Project AT-4-074

- Amino Acid Analyser
  - LKB 4400
  - LKB 4400
- Analyzers Chemistry
  - American Monitor
  - Dimension
  - Hitachi
  - Sincrone
  - Atomic Absorption Perkin Elmer
- Balances
  - Analytical Sartorious
  - Analytical Mettler
  - Toploading Mettler Sartoriou
- Beta Counters
  - Mark III Searle
  - Packard
- Centrifuges
  - Ultra Centriscan
  - Ultra Super Speed 65
  - Ultra Sorval OTD65 Dupont
  - High Speed Coolspin
  - High Speed J21B Beckman

- High Speed J6 Beckman
- Bench Top Heraecus Digifuge
- Bench Top Heraecus Labofuge
- Bench Top heraecus Microfuge

♦ Computers

- Tektronix 4051
- IBM KSU Main Frame
- Compaq 486 Deskpro
- Compaq 386
- AST 386 Primium

♦ DNA Extractor

- 341 Applied Bio System

♦ Electrophoresis

- Submarine Gel BRL
- Vertical Gel LKB Biorad Hoffer
- Multiphore LKB
- Flat bed Helina Dessaga Corning
- Flat bed Biorad Pharmacia
- Highvoltage CAMAG SAVANT

♦ ELISA

- IMX
- Amerlite

♦ F P L C

- Pharmacia

♦ Fraction Collector

- MUltirack LKB
- FRAC 300 Pharmacia
- MSE Model 328

♦ Freeze Dryers

- Beta
- Virtis

♦ Gamma Counters

- Compugamma 1282 LKB

♦ Haematology

- Coulter ZF
- Coulter S
- Co-oximeter 282
- ZNP 400 Hematoflourometer
- Zeiss Microscope with Camera
- Hema Tek Slide Stainer
- Hemo Scan

♦ HPLC

- Variant 5000 Vista Series

♦ Incubators

- Corning ACI
- Forma Scientific

- Illuminated Cooled Orbital Galankam
- CO2 Incubator Forma Scientific

♦ Luminometer

- Luminometer 1251 LKB

♦ Ovens

- Vaccume Oven LTE Qualvac
- Heracus
- Horo

♦ pH Meters

- Backman 4500
- Beckman Select Ion 5000
- Radiometer PHM83
- Radiometer PHM84

♦ Scanners

- Densitometer
- Cliniscan
- Corning 710 Flourometer

♦ Sequencers

- Beckman Peptide Sequencer
- DNA SEquencer LKB

♦ Spectrophotometers

- Spectrophotometer Zeiss
- DMR 11 Zeiss

- Zeiss DM 4

♦ Synthesizer

- DNA RNA Applied Bio System

♦ Thermocycler

- Parkin Elmer
- Statagen

♦ Water Baths

- Kolterman
- Grant

♦ Water Purification Systems

- Buchr
- Millipore

LIST OF CHEMICALS USED DURING THE PROJECT

1. Amicon concentrator
2. Amino acid calibration std.
3. Agarose NA
4. Acetonitrile HPLC grade
5. AME medium
6. Aziridin pure
7. Auto ICS pepettor tips
8. Auto ICS flowcell and stirring bars
9. Auto ICS dilution segment
10. Auto ICS buffer
11. Auto diluent
12. Acetonitrile HPLC grade
13. ACE kits 787011
14. ACE std 817759
15. Albumin A119
16. Alpha 1 antichymotrypsin A22
17. Alpha 1 antitrypsin A012
18. Alpha 1 Fetoprotine A008
19. Alpha 2 macroglobulin A003
20. Alpha 2 microglobulin A256
21. ACES
22. Antitrypsin norpartigen
23. Agarosa Prep.
24. Amberlite MB-1
25. Apolipoprotien B std, human serum
26. Apolipoprotein control serum Apo A A
27. AMES hematrak staining kits.
28. Alcohol swabs
29. Alpha D glucose 1 phosphate NA2 salt

30. Angiotensine
31. Adenosine
32. Ammonium acetate purise
33. Agar agar
34. Ampholine PAG plate
35. Ampicilline trihydrate
36. Aldolase
37. Ausab hepatitis kits
38. Ausria hepatitis kits
39. Anti-delta RIA kits
40. AVA II restriction enzyme
41. Adenosine 5' gamma P32 triphosphate
42. Bijou bottle
43. Bind silane
44. Benzene
45. B-D glucose-1-phosphate NA salt
46. Borate buffer for fluorimetry
47. Blotting pads
48. Blotter pads
49. Bgl II restriction enzyme
50. Bam HI restriction enzyme
51. Beta-thal quick column kits
52. Bind silane
53. Ceruloplasmin A03
54. Chemical resistant tubes
55. Citrate gel for acid electrophoresis
56. Citrate buffer for acid electrophore
57. Carboxylase
58. Coulter cuvattes
59. Corab hepatitis

60. Constant boiling HCL
61. Citric acid anhydrous
62. Cotton
63. Chloroform resistant tubes w/screw C
64. Coulter accuvates
65. Co-oximeter control
66. Citrate gel
67. Citrate buffer
68. Calibration II for PF8
69. Ceruliplasmin A031
70. Calibrator standard 4310-122
71. Critikan syringe driver syringe mind
72. Column PD-10 G-25 medium
73. Checker slide kits
74. Control serum
75. DCTP
76. DNA storage tubes, clear-yellow-red-B
77. D-fructose-6-phosphate (Ba salt)
78. Diethyl maleta technical
79. Deae 24-cellulose SS servacel
80. Dextran sulphate 500 BA salt
81. Dipotassium hydrogen phosphate (Anhyd)
82. DNA synthesis RPN 1256 (Amersham)
83. DNA synthesis RPN 1257 (Amersham)
84. Diluent Co-oximeter
85. DATP S35
86. DNA sequencing kits
87. Ethyl acetate
88. EDTA
89. Ependorf bottles

90. Filter with holders
91. Free fatty acid kits
92. Ficoll 400
93. Filter O rings
94. Fetal hemoglobin test kits
95. Falcon graduated tubes
96. Fetal hemoglobin staining kits
97. Filter paper 3M
98. Ferritin
99. fisher polybags
100. Fluorosine detection reagent kits
101. Floe resister
102. Fructose 6 phosphate barium salt
103. Gloves
104. G-6-PD isoenzyme reagent kits
105. Glucagone
106. G-6-PD diagnostic kits
107. Glucortise (AMES)
108. Granulocite microphage growth factor
109. Glucose determination strips
110. Gene AMP TM DNA amplification reagen
111. Gene Amplification reaction tubes
112. Gel bond paq
113. G6PDH test combination
114. Glyceraldehyde 3 phosphate
115. Glyceraldehyde 3 phosphate dehydrogen
116. Glycerophosphate dehydrogenase
117. Glutathione reductase
118. Glutathione reduced (GSH)
119. Hybond N

120. Haemoglucotest
121. Hydroxyurea
122. Hanks balanced salt solution
123. Human serum albumin
124. Hydrea CA 500
125. Heptafluorobutyric acid
126. Heptane
127. Haptaglobin
128. Hemopaxin
129. Haemoterge
130. Hb F Quick plate kits
131. Hb F Quick plate control kits
132. Hematak Pak staining kits
133. Insulin C peptide 1125 test kits
134. Iscoves modified dulbecome medium
135. Isotone II
136. Isoterge
137. IgA nor partigen
138. IgG nor partigen
139. IgM nor partigen
140. Immobiline dry plates pH 5-6
141. Immobiline dry plates pH 4-6
142. Immobiline dry plates pH 3-6
143. Immobiline dry plates pH 6.2
144. Immobiline dry plates pH 9.3
145. Lactic acid kits
146. Lactaglobulin A
147. Low melting point agrose
148. Lize III
149. M Partigen apolipoprotein

150. Minoton LMC
151. Microscopic glass slides
152. Membrane filters
153. Mst II
154. Micropipettes
155. Miilipore Millex HA 0.45
156. Micro tubes
157. 4 Methylumbellifery phosphate
158. N-butyl chloride
159. N Propanol HPLC grade
160. Norpartigen apolipoprotein B
161. Nootrophil syrup
162. Nootrophil cap 400 mg
163. Nootrophil IV injections
164. N-Propanol HPLC grade
165. Nujol mineral oil
166. Nick translation kits
167. NADP
168. Alpha NADP reduced NA salt
169. NADPH
170. Nucleotide phosphorylase 1
171. Norpartigen A antitrypsin
172. Norpartigen C3
173. Norpartigen C4
174. Oligonucleotides (Primers)
175. Opthalmodehydre OPA
176. Prealbumin
177. Polyberen
178. Protein hydrolysate kits
179. Phosphate buffer saline

180. Polyvinyl pyrolidone
181. PDP
182. pH buffer solutions
183. Opthalmodehyde OPA
184. Pyruvic acid NA salt
185. Pentafluoropropionic acid
186. Penacyle bromide
187. Pyridine
188. Polaroid filns Pos Neg
189. Pasteur pipettes longform glass
190. Pasteur pipettes longform plastic
191. Plastic wrap
192. Pipettes transfer
193. Quardral 0.1m
194. Restriction endonuclease Scq 1
195. Ribose 5-phosphate disodium salt
196. Repal silane
197. Reagents for American monitor
198. Romi 1640 medium
199. Reagent pack properdin factor B
200. Starch (smithies) Toronto Starch
201. Sodium laurylsulphate purum
202. Sephadex G25 and G50
203. Sperimidine
204. Sicke Thal Quick column
205. Sicke Quick Test
206. Solman Sperm DNA
207. Slyci tek control normal and abnormal
208. Sample cups
209. Sodium citrate buffer

210. Sodium hydroxide solution
211. Std serum apolipoprotein A
212. Tetan plates
213. TDX iron assay
214. TDX TIBC assay
215. Trifluoroacetic acid HPLC grade
216. Tetra hydrofurane
217. Transferrin norpartigen
218. Vacutainers
219. Xmn I
220. Zepoglobin