

REFERENCES

1. Arthur W. Neienhuis, Nicholas P. Anagnou, and Timothy J. Ley (1984) : Advances in thalassemia research. Blood 63(4): 750.
2. Yenchitsomanus P., Baramée A., Fucharoen S., Pootraku P., Wasi P. (1982) : Heterogeneity of Southeast Asian beta-thalassemia in Thailand. J. Trop. Med. Public Health. 13: 618-627.
3. Furbetta M., Tuveri T., Rosatelli C., Angius A., Falchi M.M., Cossu P., Meloni A., Giagu N., Cao A. (1983) : Molecular mechanism accounting for milder types of thalassemia major. J. Pediatr. 103: 35-39.
4. Triadou P., Lapoumeroulie C., Girot R., Labie D. (1983) : α -Globin Loci in homozygous β -thalassemia intermedia. Hum Genet. 63: 338-340.
5. Wainscoat J.S., Kanavakis E., Wood W.G., Letsky E.A., Huehns E.R., Marsh G.W., Higgs D.R., Clegg J.B., Weatherall D.J. (1983) : Thalassemia intermedia in Cyprus : the interaction of α - and β -thalassemia. Br. J. Haematol. 53: 411-416.
6. Weatherall D.J., Pressley L., Wood W.G., Higgs D.R., Clegg J.B. (1981) : Molecular basis for mild forms of homozygous beta thalassemia. Lancet I : 527-529.
7. Fritsch E.F., Lawn R.M., and Maniatis T. (1980) : Molecular cloning and characterization of the human beta-like globin gene cluster. Cell. 19: 959.
8. Lauer J., Shen C.K.J., and Maniatis T. (1980) : The chromosomal

- rearrangement of human alpha-like globin genes : Sequence homology and alpha-globin gene deletions. Cell. 20: 119.
9. Deisseroth A., Nienhuis A., Turner P., Velez R., Anderson W.F., Lawrence J., Creagan R., Kucherlapati R. (1977) : Localization of the human α -globin structural gene to chromosome 16 in somatic cell hybrids by molecular hybridization. Cell. 12: 205-218.
 10. Liebhaber S.A., Cash F.E., and Ballas S.K. (1968) : Human alpha globin gene expression : The dominant role of the α_2 locus in mRNA and protein synthesis. J. Biol. Chem. 261(32): 15327.
 11. Diesseroth a., Neinhuis, Lawrence J., Giles R., Turner P., Ruddie F.H. (1978) : Chromosomal localization of human α -globin gene on human chromosome 16 in somatic cell hybrids. Proc. Natl. Acad. Sci. U.S.A. 75: 1456-1460.
 12. Jeffrey A.J., Craig I.W., Francke U. (1979) : Localizations of the $G\gamma$ -, $A\gamma$ -, δ - and β -globin genes on the short arm of human chromosome 11. Nature. 281: 606-608.
 13. Schroeder W.A., Huiman T.H.J., Shelton J.R., Shelton J.B., Kleihauer E.F., Dozy A.M., and Robberson B. (1968) : Evidence for multiple structural genes for the γ chain of human fetal hemoglobin. Proc. Natl. Acad. Sci. U.S.A. 60: 537.
 14. Spritz R.A., de Riel J.K., Forget B.G.; Weissman S.M. (1980) : Complete nucleotide sequence of the human δ -globin gene. Cell. 21: 639-646.
 15. Stamatoyannopoulos G., Nienhuis A.W., Leder P., Majerus P.W. (1987): The molecular basis of blood diseases. Philadelphia, W.H.

Saunders Company, 67.

16. Weatherall D.J., and Clegg J.B. (1981) : The Thalassemia syndromes. 3rd edition. Boston, Blackwell Scientific Publication.
17. Weatherall D.J. (1984) : The Thalassemia. In : William W.J., Beutler E., Erslev A.J., Lichtman M.A. (eds). Hematology. 3rd edition, McGraw-Hill, New York, 403-521.
18. Haig H., Kazazian Jr., and Corinne D.B. (1988) : Molecular basis and prenatal diagnosis of β -thalassemia. Blood. 72(4): 1108.
19. Steinberg M.H. (1988) : Review : Thalassemia : Molecular pathology and management. Am. J. Med. Sci. 296(5): 310.
20. Orkin S.H., Old J.M., Weatherall D.J. and Nathan D.G. (1978) : Partial deletion of beta-globin gene DNA in certain patients with beta-thalassemia. Proc. Natl. Acad. Sci. U.S.A. 76: 2400.
21. Najean U., Descryver F., Henni T., Girot R. (1985) : Red cell kinetics in thalassemia : It's use for a prospective prognosis. Br. J. Haematol. 59: 533-539.
22. Orkin S.H., Kazazian H.H. Jr. (1984) : The mutation and polymorphism of the human β -globin gene and its surrounding DNA. Annu. Rev. Genet. 18: 131-171.
23. Orkin S.H. (1982) : Linkage of β -thalassemia mutations and globin gene polymorphisms with DNA polymorphisms in human β -globin cluster. Nature. 296: 627-631.
24. Thein S.L., Hesketh C., Wallace R.B., and Weatherall D.J. (1988) : The molecular basis of thalassemia major and thalassemia intermedia in Asian Indians : application to prenatal diagnosis.

- Br. J. Haematol. 70: 225-231.
25. Wainscoat J.S., Bell J.I., Old D.J., Weatherall D.J., Furbetta M., Galanello R., Cao A (1985) : Globin gene mapping studies in Sardinian patients homozygous for β^0 thalassemia. Mol. Biol. Med. 1: 1-10.
 26. Wainscoat J.S., Old J.M., Weatherall D.J. and Orkin S.H. (1983) : The molecular basis for the clinical diversity of β -thalassemia in Cypriots. Lancet i : 1235-1237.
 27. Liebhaber S.A., Kan Y.W. (1980) : Molecular pathology of $\alpha\alpha$ -thalassemia. Birth Defects. 18(7): 34-44.
 28. Proudfoot N.J., Gil A., Maniatis T. (1982) : The structure of the human zeta-globin gene and a closely linked, nearly identical pseudogene. Cell. 1: 553-563.
 29. Liebhaber S.A., Goossens M.J., Kan Y.W. (1981) : Homology and concerted evolution at the α_1 and α_2 Loci of human $\alpha\alpha$ -globin. Nature. 290: 26-29.
 30. Dozy A.M., Kan Y.W., Embury S.H., Mentzer W.C., Wong W.C., Lukin B., Davis J.R., and Koenig H.M. (1979) : $\alpha\alpha$ -globin gene organization in black precedes the severe form of $\alpha\alpha$ -thalassemia. Nature. 280: 605-607.
 31. Embury S.H., Miller J.A., Dozy A.M. (1980) : Two different molecular organization account for the single alpha-thalassemia 2 genotype. J. Clin. Invest. 66: 1319.
 32. Embury S.H., Gholson M.A., Gillette P., Rieder R.F. (1985) : The leftward deletion $\alpha\alpha$ -thalassemia 2 in a black subject with HbSS.

- Blood. 65: 769-771.
33. Higgs D.R., Hill A.V.S., Nicholls R., Goodbourn SEY, Ayyub H., Teal H., Clegg J.B., Weatherall D.J. (1985) : Molecular rearrangement of the human α -globin gene cluster. Ann. NY. Acad. Sci. 445: 46-46.
 34. Winichagoon P., Higgs D.R., Goodbourn SEY, Clegg J.B., Weatherall D.J., Wasi P. (1984) : The molecular basis of α -thalassemia in Thailand. EMBO. 3: 1813-1818.
 35. Pressley L., Higgs D.R., Clegg J.B., Weatherall D.J. (1980) : Gene deletion in α -thalassemia prove that the 5' ψ -locus is functional. Proc. Natl. Acad. Sci. U.S.A. 77: 3586-3589.
 36. Soplhocleous T., Higgs D.R., Aldridge B., Trent R.J., Pressley L., Clegg J.B., Weatherall D.J. (1981) : The molecular basic for the haemoglobin Bart's hydrop fetalis syndrome in Cyprus. Br. J. Haematol. 47: 153-155.
 37. Pressley L., Higgs D.R., Aldridge B., Metaxatou-Marromati A., Clegg J.B., Weatherall D.J. (1980) : Characterization of a new α -thalassemia 1 defect due to a partial deletion of the α -globin complex. Nucleic Acids Res. 8: 4889-4894.
 38. Orkin S.H., Michelson A. (1980) Partial deletion of the α -globin structural gene in human α -thalassemia. Nature. 286: 538-540.
 39. Nicholls R.D., Higgs D.R., Clegg J.B., Weatherall D.J. (1985) α^0 thalassemia due to recombination between the α_1 -globin gene and an AluI repeat. Blood. 65: 1434-1438.
 40. Fischel-Ghodsian N., Vickers M.A. et al. (1988) : Characterization

- of two deletions that remove the entire human zeta-alpha globin gene complex (—^{THAI} and —^{FIL}). Br. J. Haematol. 70: 233.
41. Felber B.K., Orkin S.H., and Hamer D.H. (1982) : Abnormal RNA splicing causes one form of alpha-thalassemia. Cell. 29: 982.
 42. Pirastu M., Saglio G., Chang J.C., Cao A., and Kan Y.W. (1984): Initiation codon mutation as a cause of alpha-thalassemia. J. Biol Chem. 259: 12315.
 43. Higgs D.R., Goodbourn S.E.Y., Lamb J., Clegg J.B., and Weatherall D.J. (1983) : Alpha-thalassemia caused by a polyadenylation single mutation. Nature. 306: 398.
 44. Goossens M., Lee K.Y., Liebhaber S.A., and Kan Y.W. (1982): Globin structural mutant alpha (Leu → Pro) is a novel cause of alpha-thalassemia. Nature. 296: 864.
 45. Clegg J.B., Weatherall D.J., Milner P.F. (1971) : Hemoglobin constant spring - a chain termination mutant? Nature. 234: 337.
 46. Michelson A.M. and Orkin S.H. (1980) : The untranslated regions of the duplicated human alpha-globin genes are unexpectedly divergent. Cell. 22: 371.
 47. Suominen A.I., Karp M.T., Mantasala P.I. (1984) : Fractionation of DNA with Sephacryl S-1000. Biochem. Inter. 8(2): 209-215.
 48. Gomez-Marquez J., Friere M., and Segade F. (1987) : A simple procedure for large-scale purification of plasmid DNA. Gene. 54 : 255-259.
 49. Southern E.M. (1975) : Detection of specific sequences among DNA fragments separated by gel electrophoresis. J. Mol. Biol. 98:

503-517.

50. Cohen S.N., Chang A.C.Y., Boyer H.W., and Helling R.B. (1973) : Construction of biologically functional bacterial plasmids in vitro. Proc. Natl. Acad. Sci. U.S.A. 70 : 3240-3244.
51. Davies K.E. (1986) : Fetal DNA analysis. In : Old J. (ed.). Human genetic disease : a practical approach, IRL press, Oxford, 1.
52. Cao A. (1988) : Diagnosis of β -thalassemia intermedia at presentation. Birth Defects. 23(5B): 219-226.
53. Weatherall D.J., Wainscoat J.S., Thein S.L., Old J.M., Wood W.G. Higgs D.R., and Clegg J.B. (1985): Genetic and molecular analysis of mild forms of homozygous β -thalassemia. Ann. NY. Acad. Sci. 445: 70.
54. Lynch J, Tate V.E., weatherall D.J., Fucharoen S., Tanphaichitr V. S., Isarangkura P., Seksarn P., Laosombat V., Kulapongs P., and Wasi P. (1988) : Molecular basis of β -thalassemia in Thailand. Birth Defects. 23(5A) : 71-79.
55. Higg D.R., Vickers M.A., Wilkie A.O., Pretorius M., Jarman A.P., and Weatherall D.J. (1989) : A review of the molecular genetics of the human α -globin gene cluster. J. Am. Soc. Hematol. 73(5): 1081-1104.