REFERENCES

The system of references used in this study is in accordance with the Vancouver Style[242] set in 1979 by the International Steering Committee of Medical Editors (ISCME) and with American National Standard for Bibliographic References.[243]

1. Smith ME, ed. Retina and Vitreous. San Francisco: American Academy of Ophthalmology, 1988; 4: 74-140.

2. Marmor MF. Retinitis Pigmentosa, A symposium on terminology and methods of examination. Ophthalmology (Rochester) 1983; 90(2): 126-31.

3. Merin S, Auerbach E. Retinitis pigmentosa. Surv Ophthalmol 1976; 20: 303-46.

4. Heckenlively JR, ed. Retinitis Pigmentossa. Philadelphia: JB Lippincott, 1988: 1-269.

5. Boughman JA, Caldwell RJ. Assessment of clinical variables and counselling needs in patients with retinitis pigmentose. Am J Med Genet 1982; 12: 185-93.

6. Mark OMT, ed. Retinal Diseases. Philadelphia: J.B. Lipincott, 1988; 272-83.

7. Fishman G. Hereditary retinal and choroidal diseases: Electroretinogram and electrooculogram findings. In: Peyman GA, Sanders DR, Goldberg MF, eds. Principles and practice of Ophthalmology. Volume 2. Philadelphia, London Toronto: WB Saunders Co, 1980; 2: 857-904.

8. Krill AE, ed. Hereditary Retinal and choroidal Diseases. New York: Harper & Row, 1977; 2: 479-576.

9. Karpe G: The basis of clinical electroretinography. Acta Ophthalmol 1945; 23: 1-14.

10. Bhattacharya SS, Clyton JE, Haper PS: A genetic linkage study of a Kindred with X-Linked retinitis pigmentosa. Br J Ophthalmol 1985; 69: 340-7,

11. Bhattacharyass, Wright AF, Clayton JF, Price WH, Phillips CI, Mckeown CME, Jay M, Bird AC, Pearson PL, Southern EM, Evans HJ. Close genetic Linkage between X-Linked retinitis Pigmentosa and a restriction Fragment length polymorphism identified by recombinand DNA Probe L 1.28. Nature 1984; 309: 253-5.

12. Boughman JA, Fishman GA. A genetic analysis of retinitis pigmentosa. Br J Ophthalmol 1983; 67(7): 449-54.

13. Ammann F, Klein D, Franceschetti H. Genetic and epidemiological investigation on pigmentary degeneration of the retina and allied disorders in Switzerland. J Neurol Sci 1965; 2: 183-96.

14. HU DN. Genetic aspects of retinitis Pigmentosa in China. Am J Med Genet 1982; 12: 51-6.

15. Matsunaga N. Annual report to Retinitis Pigmentosa study group, Ministry of Health and Welfare, Government of Japan: 1983; 119-25.

16. Boughman JA, Connally PM, Nance WE. Population genetic studies of retinitis pigmentosa. Am J Hum Genet 1980; 32: 223-35.

17. Bunker CH, Berson EL. Prevalence of retinitis pigmentosa in Maine. Am J Ophthalmol 1984; 97: 357-65.

18. Berson EL. Electroretinographic Findings in retinitis pigmentosa. JPN J Ophthalmol 1987; 31: 327-48.

26. Anderson SRM, Tso MOM, Fishman GA. A histopathologic study of retinitis pigmentosa. Opthalmic Paediatr Genet 1982; 1: 151-168.

27. Fishman GA. Retinitis pigmentosa, genetic percentages. Arch Ophthalmol 1978; 96: 822-6.

28. Hruby K. Retinitis pigmentosa, chibret Int J Ophthalmol 1983; 1(2): 9-22.

29. Smelser G, ed. Fundamentals and principles of Ophthalmology. San Francisco: American Academy of Ophthalmology, 1988; 1: 149-194.

30. Ismail SR, ed. Human Genetics. Alexandria: Dar EL-Maaref, 1985; 146-97.

31. Colier E, Maumenee IH, Berman ER, eds. Genetic Eye Diseases; Retinitis pigmentosa and other Inherited Eye Disorders. New York: Liss AR, 1982: 3-25.

32. Segal BS. Retinitis pigmentosa. Chibret Int J Ophthalmol 1985; 3(2): 15-23.

33. Korra A, Zeidan A, Sahn F, Zawawi A. Retinitis pigmentosa. A clinical and genetic study. Bull Ophthalmol Soc Egypt 1980; 73: 95-101.

34. Sabri N, Hussein H, EL-Hifnawy M. A genetic study in retinitis pigmentosa. Bull Ophthalmol Soc Egypt 1988; 81: 555-63.

35. Berson EL, Rosner B, Simonoff E. Risk factors for genetic typing and detection in retinitis pigmentosa. Am J Ophthalmol 1980; 89(6): 763-75.

36. Thompson JS, ed. Genetics in Medicine. Philadelphia: WB saunders Company, 1980, 70-90.

37. Spiro R, Weleber R, Kimberling W. Serum IGM in retinitis pigmentosa: A genetic study. Clin Genet 1978; 13: 295-304.

38. Jay M. On the heredity of retinitis pigmentosa. Br J Ophthalmol 1982; 66: 405-16.

39. Pearlman I, Auerbach E. The relationship between visual sensitivity and rhodopsin density in retinitis pigmentosa. Invest Ophthalmol Vis Sci 1981; 20(6): 758-65.

40. Young RW. Visual cell renewal systems and the problem of retinitis pigmentosa. Adv Exp Med Biol 1977; 77: 93-113.

41. Ripps H, Brin KP, Wale RA. Rhodopsin and visual threshold in retinitis pigmentosa. Invest Ophthalmol Vis Sci 1978; 17(8): 735-45.

42. Bird AC. Retinitis pigmentosa: A review. Trans Ophthalmol Soc NZ 1977; 29: 51-8.

43. Szamier RB, Berson EL. Retinal ultrastucture in advanced retinitis pigmentosa. Invest Ophthalmol Vis Sci. 1977; 16: 947-62.

44. Kolb H, Goura SP. Electron microscopic observations of human retinitis pigmentosa, dominantly inherited. Invest Ophthalmol 1974; 13: 489-98.

45. Rayborn ME, Moorhead LC, Holly field JG. A dominantly inherited chorioretinal degeneration resembling sector retinitis Pigmentosa. Ophthalmology 1982; 89: 1441-54.

46. Szamier RB. Ultra structure of the preretinal membrane in retinitis Pigmentosa. Invest Ophthalmol 1981; 21: 227-36.

47. Wolter JR. Retinitis Pigmentosa.Arch Ophthalmol 1957; 57: 539-53.

48. Taylor D, Lake BD, Marshall J. Retinal abnormalities in ophthalmologic Lipidosis. Br J Ophthalmol Vis Sci 1981; 65: 484-88.

49. Hansen RI, Friedman AH, Gartner S, Henkind P. The association of retinitis pigmentosa with preretinal macular glisis Br J. Ophthalmol 1977; 61: 597-600.

50. Szamier RB, Berson EL, Klein R. Sex-Linked retinitis Pigmentosa: Ultra structure of photoreceptors and pigment epithelium Invest. Ophthalmol Vis Sci 1979; 18: 145-160.

51. Gartner S, Henkind P. Pathology of retinitis pigmentosa. Ophthalmology (Rochester) 1982; 89(12): 1424-32.

52. Rodrigues MM, Wiggert B, Hackett J. Dominantly inherited retinitis pigmentosa: Ultrastructure and biochemical analysis. Ophthalmology 1985; 92: 1165-172.

53. Bloome MA, ed. Manual of Retinal and choroidal Dystrophies. New York: Accrofts, 1986; 67-75.

54. Dowling JE, ed. Retinitis pigmentosa:Clinical Implications of Current Research. New York: Plenum, 1977; 85-120.

55. Quigky HA, Anderson DR. The histological basis of optic disc pallor in experimental optic atrophy. Am J Ophthalmol 1977; 83: 709-17.

56. Gerstein DD, Dantzker DR. Retinal vascular changes in hereditary visual cell degeneration. Arch Ophthalmol 1969; 81: 99-105.

57. Hollyfield JG, Frederick JM, Tabor GA. Metabolic studies on retinal tissue from a donor with a dominantly inherited chorioretinal degeneration ressembling sectorial retinitis pigmentosa. Ophthalmology 1986; 91: 191-6.

58. Heredia CD, Vich JM, Hugnet J, Garcia-Calderon JV, Garcia-Calderon PA. Altered cellular immunity and suppressor cell activity in patients with primary retinitis pigmentosa. Br J Ophthalmol 1981; 65: 850-4.

59. Kollarits CR, Mehelas TJ, Shealy TR, Zahn JR. Von Hippel tumours in sibilings with retinitis pigmentosa. Ann Ophthalmol 1982; 14(3): 256-9.

60. Experimental Eye Research Symposium. The pigment epithelium: Its relationship to the retina in health and disease. I. Exp Eye Res 1976; 22: 395-568.

61. Young RW. Visual cells and the concept of renewal. Invest Ophthalmol 1976; 15: 700-25.

62. Young RW. Shedding of discs from rod outer segments in the Rhesus monkey. J Ultrastruct Res 1977; 34: 190-230.

63. Ishikawa T, Yamada E. The degradation of the photoreceptor outer Segments within the pigment epithelial cell of rat retina. J Electron Microscop. 1970; 19: 85-91.

64. Feeney L. Lipofuscin and melanin of human retinal pigment epithelium: Fluorescence, enzyme Cytochemical and ultrastructural studies. Invest Ophthalmol Vis Sci 1978; 17: 583-600.

65. Hogan MJ. Role of the retinal pigment epithelium in macular disease. Trans Am Acad Ophthalmol Otolaryngol 1972; 76: 64-80.

66. Bok D, Hall MO. The role of the pigment epithelium in the etiology of the inherited retinal dystrophy in the rat. J Cell Biol 1971; 49: 664- 82.

67. Berson EL. Light deprivation for early retinitis Pigmentosa: An hypothesis. Arch Ophthalmol 1971; 85: 521-29.

68. Hayes KC, Carey RE, Schmidt SY. Retinal degeneration associated with taurine deficiency in the cat. Science 1975; 188: 949-51.

69. Walt RP, Kemp CM, Lyness L. Vitamin A treatment for night blindness in primary biliary Cirrhosis. Br Med J 1984; 288: 1030-31.

70. Maraini G, Fadda G, Gozzoli F. Serum levels of retinol-binding protein in different genetic types of retinitis pigmentosa. Invest Ophthalmol 1975; 14(3): 236-7.

71. Massoud WH, Bird AC, Perkins ES. Plasma vitamin A and beta carotene in retinitis pigmentosa. Br J Ophthalmol 1975; 59: 200-4.

72. Futterman S, Swanson D, Kalina RE. Retinol in retinitis pigmentosa: Evidence that retinol is in normal concentrarion in serum and the retinol binding protein complex displays unaltered fluorescence properties. Invest Ophthalmol 1974; 13(10): 798-801.

73. Campbell DA, Tanks EL. Biochemical findings in human retinitis pigmentosa with particular relation to vit A deficiency. Br J Ophthalmol 1962; 46: 151-164.

74. Bergsma DR, Wolf ML. A therapeutic trial of vitamin A in patients with pigmentary retinal degenerations: A negative study Adv Exp Med Biol 1977; 77: 197-209.

75. Ripps H, Brin KP, Weale RA. Rhodopsin and visual threshold in retinitis pigmentosa. Invest Ophthalmol Vis Sci 1978; 17: 735-45.

76. Grouras P, Carr RE, Gunkel RD. Retinitis pigmentosa in abetalipoproteinaemia effects of vitamin A. Invest Ophthalmol Vis Sci 1971; 10: 784-93.

77. Rahi AHS. Autoimmunity and the retina. I. Antigenic specificity of photoreceptor cells. Br J Ophthalmol 1970; 54(7): 441-4.

78. Galbraith GMP, Fudenberg HH. One subset of patients with retinitis pigmentosa has immunologic defects. Clin Immuno-pathol 1984; 31: 254-60.

79. Kumar M, Gupta RM, Nema HV. Role of autoimmunity in retinitis pigmentosa. Ann Ophthalmol 1983; 15(9): 838-40.

80. Rahi AHS. Autoimmunity and the retina. II. Raised serum IgM levels in retinitis pigmentosa. Br J Ophthalmol 1973; 57(12): 904-9.

81. Jain IS, Singh KP, Dhir SP. Serum ceruloplasmin and immunoglobulins in retinitis pigmentosa patients. Ann Ophthalmol 1981; 13(6): 733-4.

82. Brinkman CJJ, Pinckers AJLG, Broek-huyse RM. Immune reactivity to different retinal antigens in patients Suffering from retinitis pigmentosa. Invest Ophthalmol Vis Sci 1986; 19(1): 743-50.

83. Char DH, Bergsma DR, Rabson AS, Albert DM, Herberman RB. Cell mediated immunity to retinal antigens in patients with pigmentary retinal degenerations. Invest Ophthalmol 1974; 13(3): 198-203.

84. Chen MC, Marak GE, Pilkerton AR. The incidence of HLA-SD antigens in recessive retinitis pigmentosa. Br J Ophthalmol 1978; 62: 172-3.

85. Rocha H, Antunes L. Primary retinitis pigmentosa: autoimmunity. Ophthalmology 1977; 1: 153-6.

86. Galbraith GMP, Pandy JP, Fudenberg HH, Ferrara GB. HLA, Immunoglobulin allotypes and cell mediated immunity in retinitis pigmentosa: A family study. J Immunogenet 1984; 11: 165-9.

87. Pruett RC, Albert DM. Vitreous degeneration in myopia and retinitis pigmentosa. Bull Soc Belge Opthalmol 1987; 23: 211-28.

88. Ross DF, Fishman GA, Gilbert LD, Anderson RJ. Variability of visual field measurements in normal subjects and patients with retinitis pigmentosa. Arch Ophthalmol 1984; 102: 1004-10.

89. Pruett RC. Retinitis Pigmentosa:Clinical observations and corelations. Trans Am Ophthalmol Soc 1983; 81: 693-735.

90. Pearlman JT, Axelrod RN, Tom A, Angeles L. Frequency of central visual impairment in retinitis pigmentosa. Arch Ophthalmol 1977; 95;894.

91. Berson EL, Sandberg MA, Rosner BB, Birch DG, Hanson AH. Natural course of retinitis pigmentosa over a three-year interval. Am J Ophthalmol 1985; 99; 240-5l.

92. Carr RE, Noble KG. Retinitis pigmentosa. Ophthalmology (Rochester) 1981; 88(2): 165-72.

93. Bec P, Mathis A, Adam P. Retinitis pigmentosa associated with astrocytic hamartomas of the optic disc. Opthalmologica 1984; 189: 135-8.

94. Rajacich GM, Heckenlively JR. The cup-disc ratio in retinitis pigmentosa. Invest Ophthalmol Vis Sci 1982; 22: 55.

95. Pruett RC. Retinitis pigmentosa: A biomicroscopical study of vitreous abnormalities. Arch Ophthalmol 1975; 93: 603-8.

96. Krauss HR, Heckenlively JR. Visual field changes in cone-rod degenerations. Arch Ophthalmol 1982; 100: 1784-1790.

97. Pearl JT. Retinitis pigmentosa: An improved clinical approach. Doc Ophthalmol Proc Ser 1977; 235-38.

98. Hyvarinen L, Maumenee AE, Kelley J, Cantollino S. Fluorescein angiographic findings in retinitis pigmentosa. Am J Ophthalmol 1971; 71(1): 17-26.

99. Bastek JV, Heckenlively JR, Straatsma BR. Cataract surgery in retinitis pigmentosa patients. Ophthalmology (Rochester) 1982; 89(8): 880-4.

100. Francois J. Chorioretinal heredodegeneration. Proc Soc Med 1961; 54: 1109-18.

101. Marmor MF. Visual Loss in retinitis pigmentosa. Am J Ophthalmol 1980; 89(5): 692-8.

102. Levy NS. Early diagnosis and evolution of dominant retinitis pigmentosa. Am J Ophthalmol 1978; 86(4): 553-6.

103. Szamier RB, Berson EL, Klein R, Meyers S. X-Linked retinitis pigmentosa: Ultrastructure of photoreceptors and pigment epithelium. Invest Ophthalmol Vis Sci 1979, 18(2): 145-60.

104. Bird AC. X-Linked retinitis pigmentosa. Br J Ophthalmol 1975; 59(4): 177-99.

105. Fishman GA. Retinitis pigmentosa. Visual loss. Arch Ophthalmol 1979; 96: 1185-8.

106. Heckenlively JR. Preserved para-arteriole retinal pigment epithelium (PPRPE) in retinitis pigmentosa. Br J Ophthalmol 1982; 66: 26-30.

107. Lorentzen SE. Drusen of the optic disc: A clinical and genetic study. Acta Ophthalmol 1966; 90: 1-180.

108. Spencer WH. Drusen of the optic disc and aberrant axoplasmic transport. Ophthalmology 1978; 85: 21-38.

109. Newman NM, Stevens RA, Heckenlively JR. Nerve fibre layer loss in diseases of the outer retinal layer Br J Ophthalmol 1987; 71: 21-6.

110. Newman NM. Ophthalmoscopic Observations of the retinal nerve fibre layer: Ophthalmology (Rochester) 1977; 83: 786-96.

111. Ayesh I, Sanders MD, Friedmann AL. Retinitis pigmentosa and Coats' disease. Br J Ophthalmol 1976; 60: 775-7.

112. Lanier JD, Mccrary JA, Justia J. Autosomal recessive retinitis pigmentosa and Coats' disease. Arch Ophthalmol 1976; 94: 1737-42.

113. Bressler NM, Gragoudas ES. Neovascularization of the optic disc associated with atypical retinitis pigmentosa. Am J Ophthalmol 1985; 10: 431-3.

114. Kaugman SJ, Goldberg MF. Autosomal dominant vitreoretinal choroidopathy. Arch Ophthalmol 1982; 100: 272-8.

115. Butner RW. Retinitis pigmentosa and retinal neovascularization; a case report. Ann Ophthalmol 1984; 16(9): 863-5.

116. Wilensky JT. Glaucoma. In: Peyman GA, Sanders DR, Goldberg MF, eds. Principles and practice of ophthalmology. Philadelphia, London, Toronto: WB Saunders Co,1984;1: 671-737.

117. Takahashi M, Jalkh A, Hoskins J. Biomicroscopic evaluation and photography in liquified vitreous in some vitreoretinal disorders. Arch Ophthalmol 1981; 99: 1555-9.

118. Fagerholm PP, Philipson BT. Cataract in retinitis pigmentosa. An analysis of cataract surgery results and pathological lens changes. Acta Ophthalmologica 1985; 63: 50-8.

119. Spalton DJ, Hitchings RA, Hunter PA, eds. Atlas of clinical Ophthalmology. The Retina, Retinal Dystrophies. Philadelphia: JB Lippincot, 1984, reprinted 1986; 16: 17-24.

120. Carr RE. Discussion of retinitis pigmentosa cataracts. Ophthalmology 1982; 89: 883-4.

121. Pau H. Differential Diagnosis of Eye Diseases. Philadelphia: W.B Saunders, 1978; P285-94.

122. Gonin J. Le scotome annulaire dan la degenerescence pigmentaire de la retina. Ann Oculist 1901; 125: 101-130.

123. Biro I. Symmetrical development of pigmentation as a specific feature of the fundus pattern in retinitis pigmentosa. Br J Ophthalmol 1979; 63: 90-6.

124. Sunga RN, Sloan LL. Pigmentary degeneration of the retina: Early diagnosis and natural history. Invest Ophthalmol 1967; 6: 309– 25.

125. Parrish RK, Schiffman J, Anderson DR. Static and kinetic visual field testing. Arch Ophthalmol 1984; 102: 1497-1502.

126. Sandberg MA, Berson EL. Visual acuity and cone spatial density in retinitis pigmentosa. Invest Ophthalmol Vis Sci 1983; 24(11): 1511-3.

127. Fishman G, Vasquez V, Fishman M, Berger D. Visual loss and foveal lesions in Usher's syndrome. Br J Ophthalmol 1979; 63: 484-8.

128. Seiff SR, Heckenlively JR, Pearlman JT. Assessing the risk of retinitis pigmentosa with the age of onset data. Am J Ophthalmol 1982; 94: 38-43.

129. Sieving PA, Fishman GA. Refractive errors of retinitis pigmentosa patients. Br J Ophthalmol 1978; 62: 163-7.

130. Rajacicn GM. The usefulness of refractive errors in diagnosing RP subtypes. Invest Ophthalmol 1983; 24: 244-5.

131. Keith CG. Bardet-Biedl Syndrome. Aus J Ophthalmol 1984; 12(2): 143-8.

132. Fishman G, ed. The Electroretinogram and Electro-oculogram in retinal and choroidal Diseases. Minnesota: American Academy of Ophthalmol and Otolaryngol 1975; 9-41.

133. Fishman GA. Basic Principles of Clinical Electro-retinography. Retina 1985; 2: 123-6.

134. Golloway NR, ed. Ophthalmic Electrodiagnosis. Philadelphia: WB Saunders, 1975; 3-160.

135. Stark J. A clinician's guide to electrophysiology of the retina. Aus J Ophthalmol 1984; 12: 171-81.

136. Fishman GA. Electroretinography and Inherited Macular Dystrophies. Retina 1985; 5: 172-8.

137. Krill AE, ed. Hereditary retinal and choroidal diseases. Vol 1. Evaluation. New York: Harper & Row, 1977; 1: 73-297.

138. Sandberg MA. Hereditary retinitis pigmentosa: Electroretinography, fundus reflectometry and psychophysics. Int Ophthalmol Clin 1980; 20(1): 39-52.

139. Marmor MF. The electroretinogram in retinitis pigmentosa. Arch Ophthalmol 1979; 97: 1300-4.

140. Berson El, Gouras P, Gunkel RD. Rod responses in retinitis pigmentosa, dominantly inherited. Arch Ophthalmol 1968; 80: 58-67.

141. Rothberg DS, Weinsteins GW, Hobson RR, Nork TM. Electroretinography and retinitis pigmentosa. No discrimination between genetic subtypes. Arch Ophthalmol 1982;100: 1422-6.

142. Van-Lith GHM. The macular function in the ERG. Doc Ophthalmol proc ser 1976; 10: 405-15.

143. Bjork A, Karpe G. The electroretinogram in retinitis pigmentosa. Acta Ophthalmol 1951; 29: 361-76.

144. Armington JC, Schwab GJ. Electroretinogram in nyctalopia. Arch Ophthalmol 1954; 52: 725-33.

145. Armington JC, Gouras P. Detection of the ERG in retinitis pigmentosa. Exp Eye Res 1961; 1: 74-80.

146. Berson EL, Gouras P, Hoff M. Temporal aspects of the ERG. Arch Ophthalmol 1969; 81: 207-14.

147. Berson EL, Rosen Job, Simonoff EA. Electroretino-graphic testing as an aid in detection of carriers of X-chromosome linked retinitis pigmentosa. Am J Ophthalmol 1979; 87: 460-8.

148. Arden GB, Carter RM, Hogg CR. A modified ERG technique and the results obtained in X-linked retinitis pigmentosa. Br J Ophthalmol 1983; 67: 419-30.

149. Kanski JJ, ed. Clinical Ophthalmology. London: Butter Worths, 1989; P369-89.

150. Berson EL. Electrical phenomena in the retina, in Moses RA ed : Adler's physiology of the Eye, St. Louis: CV Mosby Co, 1981, 466-529.

151. Berson EL, Goldstein EB. The early receptor potential in sex-linked retinitis pigmentosa. Invest Ophthalmol 1970; 9: 58-63.

152. Steinberg RH. Interactions between the retinal pigment epithelium and neural retina. Doc Ophthalmologica 1985; 60: 327-46.

153. Sockol S. Visually evoked potentials: Theory, techniques and clinical applications. Surv Ophthalmol 1976; 21: 18-44.

154. Carr RE, Siegel IM. Visual Electrodiagnostic Testing:A pratical guide for the clinician. Baltimore.: Williams & Wilkins Co, 1982; 60-74.

155. Alexander KR, Fishman GA. Prolonged rod dark adaptation in retinitis pigmentosa. Br J Ophthalmol 1984; 68: 561-9.

156. Weinstein GW, Lowell GG, Hobson RR. A comparison of electroretinographic and dark adaption studies in retinitis pigmentosa. Doc Ophthalmol Proc Ser 1976; 10: 291-302.

157. Weinstein GW, Maumenee AE, Hyvarinen L. On the pothogenesis of retinitis pigmentosa. Ophthalmologica 1971; 62: 82-97.

158. Verriest G. Les deficiences acquises de la discrimination chromatiques. Mem Acad Roy Med Belg 1964; 2: 35-327.

159. Fishman GA, Young RSL, Vasquez V, Lourenco P. Colour vision defects in retinitis pigmentosa. Ann Ophthalmol 1981; 13(5): 609-18.

160. Heckenlively JR, Martin DA, Rosales TO. Telangiectasia and optic atrophy in cone-rod degenerations. Arch Ophthalmol 1981; 99: 83-91.

161. Mallick KS, Zeimer RC, Fishman GA, Blair NP, Anderson RJ. Transport of Fluorescein in the ocular posterior segment in retinitis pigmentosa. Arch Ophthalmol 1984; 102: 691-6.

162. Berkow JW, Kelley JS, Orth DH, eds. Fluorescein Angiography: A guide to the interpretation of Fluorescein Angiograms. San Francisco: American Academy of Ophthalmology, 1984; 9-101.

163. Don wong RBP. Techniques of Fundus photography. New York: Eastman Kodak, 1976; 39-59.

164. Hyvarinen L, Maumenee AE, George T. Fluorescein Angiography of the choriocapillaris. Am J.Ophthalmol 1969; 67(5): 653-66.

165. Novotny HR, Alvis DL. A method of photographing fluorescence in circulating blood in the human retina. Circulation 1961; 24: 82.

166. Cunha-Vaz J, Maurice DM. The active transport of fluorescein by the retinal vessels and the retina. J Physiol 1967; 191: 467.

167. Bird AC. X-linked retinitis pigmentosa. Br J Ophthalmol 1975; 59: 177.

168. Gieser DK, Fishman GA, Cunha-Vaz J. X-linked recessive retinitis pigmentosa and vitreous fluorophoto metryy. Arch Ophthalmol 1980; 93: 307-10.

169. Fishman GA, Maggiano JM, Fishman M. Foveal lesions seen in retinitis pigmentosa. Arch Ophthalmol 1977: 96(11): 1993-6.

170. Fishman GA, Fishman M, Moggiano J. Macular lesions associated with retinitis pigmentosa. Arch Ophthalmol 1977; 95: 798-803.

171. Krill AE, Newell FW, Chishti M. Fluorescein studies in diseases affecting the retinal pigment epithelium. Am J Ophthalmol 1968; 66-470.

172. Fetkenhour CL, Choromokos E, Weinstein J, Shoch D. Cystoid macular oedema in retinitis pigmentosa. Trans Am Acad Ophthalmol & Otolaryngol 1977; 83: 515-21.

173. Fogle JA, Welch RB, Green WR. Retinitis pigmentosa and exudative vasculopathy. Arch Ophthalmol 1978;96: 696-702.

174. Spalton DJ, Bird AC, Cleary PE. Retinitis pigmentosa and retinal oedema. Br J Ophthalmol 1978; 62: 174-82.

175. Wright BE. Cystoid macular oedema of non retinal vascular origin. Bull.N.Y. Acad. Med 1982; 58(6): 507-18.

176. Spallone A: Retinal fluorescein leakage in retinitis pigmentosa. Am J Ophthalmol 1985; 102: 408

177. Godel V, Regenbogen L. Unilateral retinitis pigmentosa and pit of optic disc. Arch Ophthalmol 1976;94: 1417-8.

178. Hyvarinen L, Maumenee AE, Kelly J. Fluorescein Angiographic Findings in Retinitis Pigmentoso. Am J Ophthal-mol 1971; 71(1): 17-26.

179. Krill AE, Archer D, Newell FW. Fluorescein Angiography in Retinitis Pigmentosa. Am J Ophthalmol 1970; 69(5): 827-35.

180. Geltzer AI, Berson EL. Fluorescein Angiography of Hereditary Retinal Degenerations. Arch Ophthalmol 1969; 81: 776-82.

181. Schauwvlie GHEP, Delaey JJ. Fluorophotometry in Retinitis Pigmentosa. Bull Soc Belge Ophthalmol 1985; 215: 87-94.

182. Best M, Toyofuku H, Galin MA. Ocular hemodynamics in retinitis pigmentosa. Arch Ophthalmol 1972; 88-123-30.

183. Heckenlively JR, Friederich F. Retinitis Pigmentosa in the Navajo. Matab Pediatr Syst Ophthalmol 1981; 5: 201-6.

184. Fish G, Greyr, Sehmi KS. The dark choroid in posterior retinal dystrophies.Br J Ophthalmol 1981; 65: 359-63.

185. Eagle LRC, Lucier AC, Bernardino VB. Retinal pigment epithelial abnormalities in fundus flavimaculatus. Ophthalmolgy 1980; 87: 1189: 1200.

186. Fishman GA, Weinberg AB, McMahon TT. X-linked recessive retinitis pigmentosa:Clinical characteristics of carriers. Arch Ophthalmol 1980; 104: 1329-35.

187. Ricc H, Amman A, Ammann F, Franceschetti A. Reflet taptoderversilde (phenomenomne de Mizuno inverse) chez descon-ductrices dertinopathie pigmentaire recessive lie du sexe. Bull Soc Fran Ophthalmol 1983; 76: 31.

188. Newsome DA. Retinal Fluorescein Leakage in Retinitis Pigmentosa. Am J Opthalmol 1986; 101: 354-60.

189. Ffytche TJ. Cystoid maculopathy in retinitis pigmentosa. Trans Ophthalmol Soc UK 1972; 92: 265.

190. Merin S. Macular cysts as an early sign of tapetoretinal degeneration. J Paediatr Ophthalmol 1970; 7: 225-28.

191. Newsome DA, Blacharski PA. Grid photocoagulation for Macular Edema in Patients with Retinitis pigmentosa. Am J Ophthalmol 1987; 103: 161-6.

192. Green WR. Retina. In: Spencer WH, ed. Ophthalmic pathology. An Atlas and Textbook. 3rd ed. Philadephia: WB Saunders, 1985, 989-1210.

193. Gass JDM. A fluorescein angiographic study of macular dysfunction secondary to retinal vascular disease. Arch Ophthalmol 1968; 80: 535.

194. Bresnick GH. Diabetic maculopathy: A critical review highlighting diffuse macular edema. Ophthalmology 1983; 90: 1301.

195. Allen AWJr, Gass JDM. Contraction of a perifoveal epiretinal membrane simulating a macular hole. Am J Ophthalmol 1976; 82: 684-91.

196. Albert D, Geltzer AJ. Retinitis Punctata Albescens in a negro child studied with fluorescein angiography. Arch Ophthalmol 1969; 81: 170-5.

197. Schauwvlie P, Delaey JJ. Fluorophotometry: Indications and technique. Bull Soc Belge Ophthalmol 1985; 214: 53-64.

198. Miyake Y, Hiroshi I. Vitreous fluorophotometry in patients with cone-rod dystrophy. Br J Ophthalmol 1984; 68: 489-93.

199. Travassos A, Fishman G, Cunha-Vaz J. Vitreous fluorophotometry studies in retinitis pigmentosa. Graefe's Arch Clin Exp Ophthalmol 1985; 222: 237-40.

200. Fishman GA, Cunha-Vaz J, Salzano T. Vitreous fluorophotometry in patients with retinitis pigmentosa.Arch Ophthalmol 1981; 99: 1202-7.

201. Foxman SG, Wirtschafter JD, Letson RD. Leber's Congenital amourosis and high hyperopia: a discrete entity. In: Henkind P, ed. ACTA: XXIV International Congress of Ophthalmology. Philadelphia: JB Lippincott Co, 1983; 1: 55-58.

202. Pearlman JT, Flood TP, Seiff SR. Retinitis pigmentosa without pigment. Am J Ophthalmol 1976; 81(4): 417-9.

203. Hellner KA, Rickers J. Familiary bilateral segmental retinopothia pigmentosa. Ophthalmologica 1973; 166: 327-41.

204. Omphroy CA. Sector retinitis pigmentosa and chronic angle-closure glaucomo: a new association. Ophthalmologica 1984; 189: 12-20.

205. Bastek JV, Foos RY, Heckenlively J. Traumatic Pigmentary retinopathy Am J Ophthalmol 1981; 92: 621-4.

206. Cogon DG. Pseudoretinitis pigmentosa. Arch Ophthalmol 1969; 81: 45-53.

207. Franois J. The differential diagnosis of tapeto retinal degeneration. Arch Ophthalmol 1958; 59: 86-120.

208. Ellis D, Heckenlively J. Retinitis punctata albescens: Fundus appearance and functional abnormalities. Retina 1983; 3: 27-31.

209. Vannas S, Setala M. On atypical night blindness. Acta Ophthalmol 1958; 36: 849-59.

210. Galindez I. Retinitis punctata albescens. Am J Ophthalmol 1954; 37: 436-8.

211. Carr RE, Ripps H, Siegel IM.Visual pigment kinetics and adaptation in fundus albipunctatus. Doc Ophthalmologica proc ser 1974; 4: 193-206.

212. Marmor MF. Dystrophies of the retinal pigment epithelium. In: Zinn KM, Marmor MF, eds. The Retinal Pigment Epithelium. Cambridge: Harvard Press, 1979; 424-53.

213. Marmor MF, Defining fundus albipunctatus. Doc Ophthalmologica Proc Ser 197 ;13; 227-34.

214. Fishman GA, Buckman G, Van Every T. Fundus Flavimaculatus: A clinical classification. Doc Ophthalmol Proc Ser 1977; 13: 213-20.

215. Bird AC. Management of retinitis pigmentosa.Trans Ophthalmol Soc UK 1984; 103: 494-5.

216. Fonda G. Low vision correction in retinitis pigmentosa and associated diseases. Int J Rehab Research 1980; 3(1): 39-43.

217. Editorial. Treatment of retinitis pigmentosa. Br Med J 1976; 1: 58-9.

218. Marrow MF. The management of retinitis pigmentosa and allied disorders. Ophthalmol Digest 1979; 41: 13-20.

219. Berson EL. Light deprivation for early retinitis pigmentosa. Arch Ophthalmol 1971; 85: 521-9.

220. Miller D, Weiter J. Clinical Light Damage to the Eye. New York: Springer-Verlog, 1987; 79-127.

221. Epstein RL. Inborn metabolic disorders and the eye. In: Peyman GA, Sanders DR, Goldberg MF, eds. Principles and practice of ophthalmology. Volume 3. Philadelphia, London, Toronto: WB Saunders co, 1980; 3: 1707-76.

222. Fishman GA, Kumar A, Joseph ME, Torok N, Anderson RJ. Usher's syndrome. Ophthalmic and neuro-otologic findings suggesting genetic heterogeneity. Arch Ophthalmol 1983; 101: 1367-74.

223. Isaeff WB, Nis Wonger J.Survey of 336 deaf students for retinitis pigmentosa. Ann Ophthalmol 1981; 13(10): 1131-2.

224. Merin S, Abraham FA. Usher's and Hallgren's syndromes. Acta Genet Med Gemellol (Roma) 1973; 23: 49-55.

225. Bloom TD, Fishman GA. Usher's syndrome. CNS defects determined by computed tomography. Retina 1983; 3: 108-13.

226. Campo RV, Aabreg TM. Ocular and Systemic mainfestations of the Bardet-Biedl syndrome. Am J Ophthalmol 1982; 94: 750-6.

227. Keith CG. Bardet-Biedl Syndrome. Aus J Ophthalmol 1984; 12(2): 143-8.

228. Kornzweig A. Bassen-Kornzweig syndrome: Present status. J Med Genet 1970; 7: 271-6.

229. Gouras P, Carr RE, Gunkel RD. Retinitis pigmentosa in abetalipoproteinemia: Effects of vitamin A. Invest Ophthalmol 1971; 10: 784-93.

230. Judisch GF, Rhead WJ, Miller DK. Abetalipoproteinemia: Report of an unsual patient. Ophthalmolgica 1984; 189: 73-9.

231. Carr RE. Primary Retinal Degenerations. In Duane TD, Jaeger EA, eds. Clinical Ophthalmology. Philadelphia: Harper and Row, 1985; 3(24): 10-13.

232. World Health Organization. Methods of Assessments of Avoidable Blindness. WHO Offset Publication No.54. Geneva: WHO, 1980.

233. Vaughan D, Asbury T, eds. General ophthalmology. 10th ed. Los Altos, California: Lange Medical Publications, 1983; 340-1.

234. Michaels D, ed. Basic Refraction Techniques. New York: Raven Press, 1988; 10-23.

235. Sarhan AE, Elshabrawy M, Kashlan KM, eds. Introduction to statistical methods in medicine and public health. Alexandria: Dar EL-Maaref, 1962; 20-32.

236. Hill BA, ed. Principles of Medical Statiatics. London: Lancet, 1971; 260-84.

237. Fonda G. Low Vision Correction in retinitis pigmentosa and associated diseases. Int J Rehabil Res 1980; 3: 34-43.

238. Heckenlively JR. The frequency of posterior subcapsular cataract in the hereditary retinal degenerations. Am J Ophthalmol 1982; 93: 733-8.

239. Schepens CL. Clinical aspects of pathologic changes in the vitreous body. Am J Ophthalmol 1964; 28: 8-21.

240. Sidd RJ, Fine SL, Owens SL. Idiopathic preretinal gliosis. Am J Ophthalmol 1982; 94: 44-8.

241. Sternberg PJ, Landers MB, Walbarsht M. The negative coincidence of retinitis pigmentosa and proliferative diabetic retinopathy. Am J Ophthalmol 1984; 97(6): 788-9.

242. International Steering Committee of Medical Editors. Uniform requirements for manuscripts submitted to biomedical journals. Am Rev Respir Dis 1979; 119: 3-10.

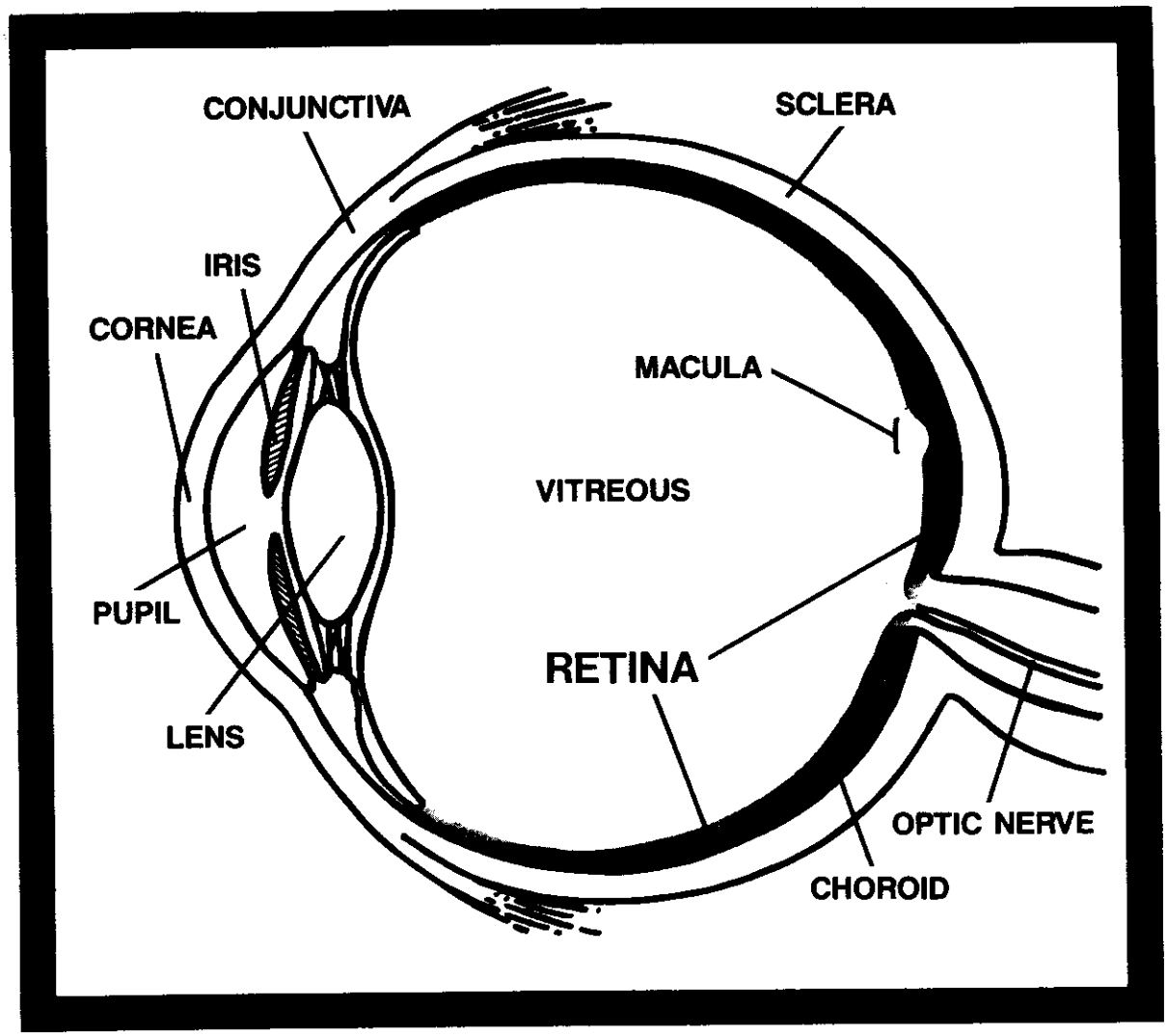
243. American National Standards Institute. American national standard for bibliographic references, Z39.29-1977. New York: American National Standards Institute, 1972.

Answers To Your Questions About Retinitis Pigmentosa

& Other Retinal Degenerative Diseases.

RP™

RP Foundation Fighting Blindness
National Headquarters



1. WHAT IS RETINITIS PIGMENTOSA (RP)?

RP is the name given to a group of diseases which affect the retina. The retina, located in back of the eye, acts like the film in a camera. It is a delicate layer of cells which picks up the picture and transmits it to the brain—where "seeing" actually occurs. In RP, the retina begins to degenerate, which first causes peripheral vision and night vision to diminish, and usually only decades later, central vision.

2. WHAT ARE THE EARLY SYMPTOMS OF RP?

Often, one of the earliest symptoms of RP is difficulty seeing at night or in dimly lit places (night blindness). Later there is a very gradual loss of side (peripheral) vision, resulting in "tunnel vision" in some cases. The symptoms of RP generally worsen over a period of years. When other members of a family are also affected, the rate of vision loss is usually similar to that experienced by the other affected members.

3. CAN RP BE EASILY DIAGNOSED?

Because each case of RP can be so different, sometimes it is easy to diagnose and other times it is not. Generally, RP is most difficult to diagnose in the earliest stages. As the disease advances, some characteristic changes in the appearance of the retina begin to occur. Depending on the stage of the disease, various tests may need to be done. Electroretinogram (ERG), visual field and other tests of visual function may be required to determine the precise state of the retina. Whenever RP is suspected, a patient should have a thorough evaluation by an ophthalmologist who is familiar with retinal degenerations.

4. WHAT CAUSES RP?

We know that certain cells in the retina—the rods and cones—stop functioning. However, we do not know what causes this degeneration. Research provides us with the opportunity to seek solutions.

5. WHO IS DOING THE RESEARCH ON RP?

Through the efforts of the RP Foundation Fighting Blindness, the number of highly qualified scientific investigators working full time on RP and related diseases continues to grow. One reason research efforts in this type of retinal disease are difficult is because there is no means of safely removing living tissue from the delicate eye in order to study it.

6. WHAT IS AN RP RESEARCH CENTER?

The RP Foundation Fighting Blindness supports extensive research at a number of major medical centers and hospitals around the world. The purpose is to find the causes, means of prevention and treatments of retinal degenerative diseases such as retinitis pigmentosa, Usher's syndrome and macular degeneration. Most of these centers also provide clinical evaluations and special testing. Even with these evaluations, it is important for patients with RP to remain in the care of their own eye doctor for general eye care.

7. DO THESE RESEARCH CENTERS STUDY OTHER RETINAL DEGENERATIONS?

Researchers at RP centers also study many other related diseases which affect the retina. Often, the term RP is used to encompass all of these. Research toward understanding one retinal degeneration may contribute to the understanding of related retinal disorders.

8. IS THERE A TREATMENT TO ARREST THE PROGRESSION OF RP?

Currently there is no way to halt the degeneration of the retina. Many speculative attempts at "treatments" have been undertaken over the past years, but there is no convincing scientific evidence to demonstrate any benefits from these attempts at treatment. In fact, most of these are unregulated experiments on humans. Our scientists follow up all reasonable treatment possibilities in their vigorous search for a retinal therapy.

9. IF I AM DIAGNOSED WITH RP, HOW QUICKLY WILL IT PROGRESS?

Most RP patients have a very gradual progression of symptoms, often over a period of many years or even decades. Vision often remains stable between annual examinations.

10. CAN RP CAUSE BLINDNESS?

Blindness, to most people, refers to a complete loss of sight. Although some RP patients with advancing age do become blind in this sense, most will retain at least some vision, and are classified as "legally blind." Each individual case differs.

11. WHAT IS MEANT BY THE TERM "LEGAL BLINDNESS"?

Legally blind individuals are those whose best visual sharpness or acuity (with glasses, if needed) is 20/200 or worse in the better eye; or whose visual field, regardless of visual acuity, is restricted to a 20 degree diameter (10 degree radius).

12. IS RP ASSOCIATED WITH OTHER PROBLEMS?

Usually it is not; however, some people with RP also have hearing loss present at birth or shortly thereafter. When this occurs it is called Usher's syndrome. On rare occasions, RP may be associated with other syndromes.

13. WHAT ABOUT CATARACTS AND RP?

It is not unusual for an individual with RP to develop a cataract, which is a clouding of the lens of the eye. Cataracts that significantly interfere with vision can be surgically removed. Whether or not surgery improves the vision often depends on the extent of retinal changes. Because surgery is not for everyone, it is necessary to discuss its advisability with an ophthalmologist.

14. DOES NIGHT BLINDNESS MEAN A PERSON HAS RETINITIS PIGMENTOSA?

No. Night blindness can be a symptom of a number of retinal disorders besides RP, and in some cases does not represent a progressive eye disorder. Diagnosis requires expert evaluation.

15. CAN AN EYE TRANSPLANT BE PERFORMED?

No. The retina (the layer of cells in the eye affected by RP) is connected to the brain, and, within the limits of current scientific knowledge, cannot successfully be transplanted. Although you may hear people talk about "eye transplants", they are referring to corneal transplants, which have no relationship to RP.

16. WHAT ARE THE EFFECTS OF VITAMIN TREATMENTS?

As far as our scientific advisers can ascertain, it is not known if vitamin treatments are helpful in treating retinitis pigmentosa. There is no proven vitamin deficiency or shortage in patients with RP. Thousands of people affected by RP have taken large quantities of vitamins, especially vitamin A, without proven beneficial effect. However, scientists are continuing to study this matter.

17. DOES LIGHT AFFECT THE VISUAL LOSS IN RP?

There is no scientific evidence that normal levels of light increase visual loss. Persons with RP may use their eyes in ordinary light without restriction, although many are more comfortable avoiding bright lights. As a precaution, individuals with RP and other retinal degenerations are encouraged to protect their eyes from long-term exposure to bright sunlight until more is learned. Good quality sunglasses are useful for bright days outdoors.

18. THERE ARE TIMES WHEN MY VISION SEEMS BETTER AND TIMES WHEN IT SEEMS WORSE. DOES THIS HAPPEN TO OTHERS?

Yes, it does. A number of factors might account for good days or bad days for someone with RP. Some people feel they see better on cloudy days; others feel they do not see as well on cloudy days. Fatigue and emotional stress may also temporarily affect vision. All people with RP have days when, for no apparent reason, they seem to function better or worse than others.

22. IF MY CHILD IS DIAGNOSED AS HAVING A RETINAL DEGENERATIVE CONDITION SUCH AS RP, WHEN AND WHAT SHOULD I TELL HIM?

There are no hard and fast rules for when and what to explain to a child about RP. A child's need for information varies depending on age and maturity. Although children are very perceptive and quickly sense they have a visual problem, they may be able to absorb only a little information about it at a time. It is usually best to answer their questions as frankly and positively as possible, without offering more information than your child has requested. Help your child understand that some of his or her limitations may be due to the visual impairment, without using the condition as a crutch or excuse for not reaching a reasonable goal. You can reassure your child about two concerns that are frequently voiced. RP will not change the appearance of one's face or eyes. Also, most children with RP can complete their education in a regular school system.

23. CAN I DRIVE IF I HAVE RP?

This is a difficult question. Certainly many people with RP do drive legally, and drive well. Legal requirements vary widely from state to state. It would be best to discuss your visual limitations and driving with your own ophthalmologist.

24. WHAT IF MY FAMILY NEEDS HELP IN COPING WITH RP?

Families with RP may find it helpful to discuss their questions and concerns with other people who have similar problems and experiences. The RP Foundation, through its affiliates, tries to offer this opportunity. Professional guidance from a medical social worker, school counselor, psychologist, psychotherapist or genetic counselor may also be very beneficial. A list of service agencies and resources appears at the end of this booklet.

25. IF MY RP REACHES AN ADVANCED STAGE, WILL I BECOME DEPENDENT ON OTHERS?

Many aids, services and techniques are available to provide increased mobility and independence to people with RP. Some of these include orientation and mobility training, control of illumination and special lenses. A department of vocational rehabilitation or commission for the blind or visually impaired in your state offers training programs to assist in adapting to such situations as traveling, housekeeping, employment and education. Check your telephone directory under state government for these listings.

26. WILL MY CAREER BE AFFECTED?

People with retinal degenerative diseases can continue to lead productive lives and pursue career goals. By determining appropriate aids, training and other job modifications in the chosen field, potential problems can be identified and forestalled. Vocational counseling, offered through educational institutions, or state and local agencies mentioned above, can be very beneficial when planning or maintaining a career. When RP is diagnosed early, a person can often take full advantage of educational and career guidance.

27. WHAT ARE LOW VISION AIDS?

Low vision aids are devices which help people maximize the use of their remaining vision. These aids may be:

OPTICAL AIDS, such as Corning and NOIR glasses, the Fresnel Prism, telescopes, microscopes and night vision aids;

NON-OPTICAL AIDS, such as the Wide Angle Mobility Light, paper guides, large print typewriter and adjustable stands; and,

ELECTRONIC AIDS, such as Apollo Laser and Visualtek closed-circuit TV, reading machines and talking computers.

To determine which aids may help you the most, it is wise to obtain a thorough low vision evaluation from a specialist or a low vision clinic.

28. WHAT ASSISTANCE DOES THE ITT NIGHT VISION AID OFFER INDIVIDUALS WITH NIGHT BLINDNESS?

The Night Vision Aid, developed and manufactured by ITT, is a hand-held device that may help some individuals at a particular stage in their RP when they have functional daytime vision. This aid is expensive, but in some areas costs may be covered by health insurance or by state departments of rehabilitation. For further information, including a list of current Night Vision Aid Screening Centers in the United States and Canada, contact Night Vision Aid Distributors at 1401 Mt. Royal Avenue, 4th Floor, Baltimore, Maryland, 21217, or call 301-225-9400.

29. CAN I DONATE MY EYES TO THE FOUNDATION FOR RESEARCH?

In 1980, the Foundation initiated a National Retina Donor Program to meet the acute, increasing need of our researchers to study human retinal tissue. Valuable retinal tissue is obtained after death from

persons affected by retinal degenerations or members of their families. A very reliable retina donor notification and retrieval system has been developed. Eyes are prepared and shipped to our researchers as soon as possible after death to maximize the specimen's usefulness in biochemical research. The Retina Donor Program has already greatly enhanced research efforts. This precious anatomical gift involves no cost to the donor's family or estate. Contact the RP Foundation for additional information.