

CONTENTS

| | |
|-------------------------------------------|------------------|
| <i>Illustrations</i> | <i>page</i> xiii |
| <i>Foreword</i> | xvii |
| <i>Introduction</i> | xix |
| <i>Chapter I. GENERAL: AETIOLOGY</i> | 1 |
| Definition | 1 |
| Title | 1 |
| Retrospect | 2 |
| Summary of Questionnaire | 3 |
| AETIOLOGY | 4 |
| Incidence | 4 |
| Bilateral incidence | 5 |
| Influence of sex | 6 |
| Time of origin | 7 |
| Age of onset | 7 |
| Relationship to juvenile glaucoma | 8 |
| Influence of heredity | 11 |
| Heredity in congenital glaucoma | 12 |
| Influence of consanguinity | 14 |
| Anticipation | 15 |
| Summary | 16 |
| References | 17 |
| <i>Chapter II. DIFFERENTIAL DIAGNOSIS</i> | 19 |
| Myopia | 19 |
| Tears in Descemet's membrane in myopia | 22 |
| Anterior staphyloma | 26 |
| Keratectasia | 27 |
| Keratoconus | 27 |
| Megalocornea | 28 |
| Incidence | 37 |
| Inheritance | 38 |
| Theories of origin | 39 |
| Megalophthalmia | 45 |
| Summary | 46 |
| References | 47 |

| | |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------|
| <i>Chapter III. THE STRUCTURE AND DEVELOPMENT OF THE INVOLVED TISSUES: THEIR EMBRYOLOGY AND THEIR COMPARATIVE ANATOMY</i> | <i>page</i> 50 |
| The development of these tissues | 50 |
| The sclera | 50 |
| The cornea, its size, its curvature | 51 |
| The deepening of the angle of the anterior chamber | 53 |
| The development and the retrogression of the associated mesoderm | 53 |
| The structure of these tissues | 62 |
| The angle of the anterior chamber | 62 |
| The scleral furrow and the scleral roll or spur | 63 |
| The canal of Schlemm: the afferent arteriolar supply to Schlemm's canal | 69 |
| The meshwork of the angle: termination of Descemet's membrane | 72 |
| Iris processes: the anterior attachment of the ciliary muscle | 75 |
| The comparative anatomy of involved tissues | 77 |
| Conclusions | 96 |
| References | 97 |
| <i>Chapter IV. THE PATHOLOGY OF CONGENITAL GLAUCOMA</i> | 99 |
| A. Interference with function | 99 |
| Refraction | 99 |
| Vision | 104 |
| Intra-ocular pressure | 105 |
| B. Alterations in structure | 106 |
| The orbit and the globe: size of angle and degree of distension | 106, 110 |
| The sclera | 111 |
| The cornea: increase in size, thinning of cornea, changes of curvature, opacities of the cornea, age of onset of the tears, congenital pupillary synechiae and corneal defects | 119 |
| The uveal tract: the iris, aniridia, the ciliary body, the ciliary and vorticosae veins, the choroid | 127 |

CONTENTS

ix

| | |
|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------|
| The lens: alteration in shape and position, aplasia | <i>page</i> 136 |
| The retina and the optic nerve | 139 |
| The anterior chamber | 141 |
| The angle of the anterior chamber: the angle in chronic glaucoma, the angle and Schlemm's canal, persistent or aberrant meshwork in the angle, defects of Schlemm's canal, posteriorly placed Schlemm's canal, rudimentary development of scleral spur, peripheral anterior synechiae | 142–152 |
| Summary | 153 |
| C. Association of hydrophthalmia with other anomalies | 153 |
| Ocular defects: non-ocular anomalies | 153 |
| Associated hydrophthalmia | 156 |
| 1. Generalised neurofibromatosis or von Recklinghausen's disease: clinical manifestations, onset and distribution of ocular lesions, summary of reported cases, ciliary neurofibromata, summary of examined specimens, cranial and intra-cranial lesions, signs of intra-cranial disease, association with hemihypertrophy, the escape of the eye in hemihypertrophy and atrophy, other ocular changes in neurofibromatosis | 158–179 |
| 2. Hydrophthalmia and facial naevi: summary of cases, angle of filtration, vascular changes in the uvea, other ocular changes, summary of examined specimens, cranial and intra-cranial lesions, disorders of growth, hemihypertrophy, meningo-cutaneous angiomatosis, intra-cranial angiomatous malformations that may be associated with facial naevi and glaucoma, contrast with neuro-retinal angiomatosis | 180–206 |
| 3. Causes of hydrophthalmia in association with facial naevi and neurofibromatosis: mechanical theories, neuro-vascular theories, a nervous origin, other theories | 211–218 |
| Summary | 221 |
| References | 222 |

| | |
|-----------------------------------------------------------------------------------------------------------------|-----------------|
| <i>Chapter V. PATHOGENESIS</i> | <i>page</i> 230 |
| The theories of origin of hydrophthalmia | 230 |
| 1. The developmental theory | 231 |
| The significance of ocular anomalies | 232 |
| The significance of the association with non-ocular anomalies | 233 |
| The significance of persistent and aberrant mesoblastic tissue in and round the anterior chamber | 234 |
| The significance of the meshwork in the angle | 236 |
| Hydrophthalmia and neurofibromatosis | 239 |
| Hydrophthalmia and facial naevi | 240 |
| Aniridia | 240 |
| Microphthalmia | 241 |
| The significance of an “absent” canal of Schlemm | 242 |
| The significance of iris processes | 244 |
| The significance of peripheral anterior synechiae | 245 |
| The significance of congenital pupillary synechiae and congenital corneal defects | 248 |
| Congenital glaucoma in the lower mammals | 259 |
| Experimental evidence of interference with filtration | 260 |
| 2. Inflammation as a cause | 260 |
| Introduction | 260 |
| The significance of the signs of ocular inflammation | 261 |
| The significance of the state of the choroid | 266 |
| The significance of lowered resistance of the scleral coat | 267 |
| The significance of endophlebitis | 268 |
| The significance of the state of the vitreous | 270 |
| The significance of the association with general disorders: syphilis, early incidence of interstitial keratitis | 272–3 |
| 3. Other theories of origin | 279 |
| The association with nervous and endocrine disorders: hypersecretion, Angelucci’s theory | 279 |
| Two main types of hydrophthalmia | 280 |
| Summary | 285 |
| References | 289 |

CONTENTS

xi

| | |
|----------------------------------------------------------------------|-----------------|
| <i>Chapter VI. TREATMENT</i> | <i>page</i> 292 |
| History of treatment | 292 |
| Medical treatment: miotics | 294 |
| Operative treatment | 299 |
| Iridectomy | 299 |
| Paracentesis of the cornea | 305 |
| Posterior sclerotomy | 306 |
| Anterior sclerotomy | 308 |
| Incision of the angle: the operation of de Vincentiis (1893–5) | 312 |
| The theory of fistulisation | 314 |
| Lagrange's sclerecto-iridectomy | 318 |
| Holth's sclerectomy | 322 |
| Corneo-scleral trephining | 323 |
| Holth's iridencleisis | 330 |
| Iridotaxis | 332 |
| Heine's cyclodialysis | 332 |
| Other operations: extraction of the lens | 334 |
| The relative merits of various operations | 335 |
| Multiple operations | 337 |
| The question of operation | 338 |
| The time for operation | 338 |
| Tension after operation | 340 |
| Treatment of complications: cataract, detachment of the retina | 341–2 |
| Summary | 342 |
| References | 343 |
| <i>Chapter VII. PROGNOSIS</i> | 347 |
| The final picture. A permanent cure | 347 |
| I. The spontaneous arrest of congenital glaucoma: mild cases | 348 |
| II. Results of operative treatment in different series | 354 |
| III and IV. Vision of unoperated and operated patients in later life | 354 |
| V. Influence of age at operation on vision and tension | 355 |

| | | |
|-----|------------------------------------------------------------------------------------------------------------------|----------|
| xii | CONTENTS | |
| | <i>Chapter VII. PROGNOSIS (contd.)</i> | |
| | VI. Influence of size of cornea on visual prognosis | page 356 |
| | Optic disc | |
| | VII. Analysis of cases with final vision of at least 6/12 | 358 |
| | VIII. The prognosis of hydrophthalmia when associated with neurofibromatosis and facial naevi | 359 |
| | IX. Juvenile mortality | 361 |
| | Information obtained from Questionnaire | 361 |
| | Summary | 362 |
| | References | 363 |
| | <i>Chapter VIII. GENERAL REFLECTIONS</i> | 365 |
| | <i>Index</i> | 371 |
| | <i>Tables at back</i> | |
| | Analysis of Early Specimens | |
| | Analysis of Specimens over 2½ years and under eleven | |
| | Analysis of Specimens over eleven years | |
| | available for download from www.cambridge.org/9781107625518 | |