REVIEW OF PRIMARY CONGENITAL GLAUCOMA

Essay
Submitted for partial fulfillment of
Master degree (M.Sc.) In ophthalmology

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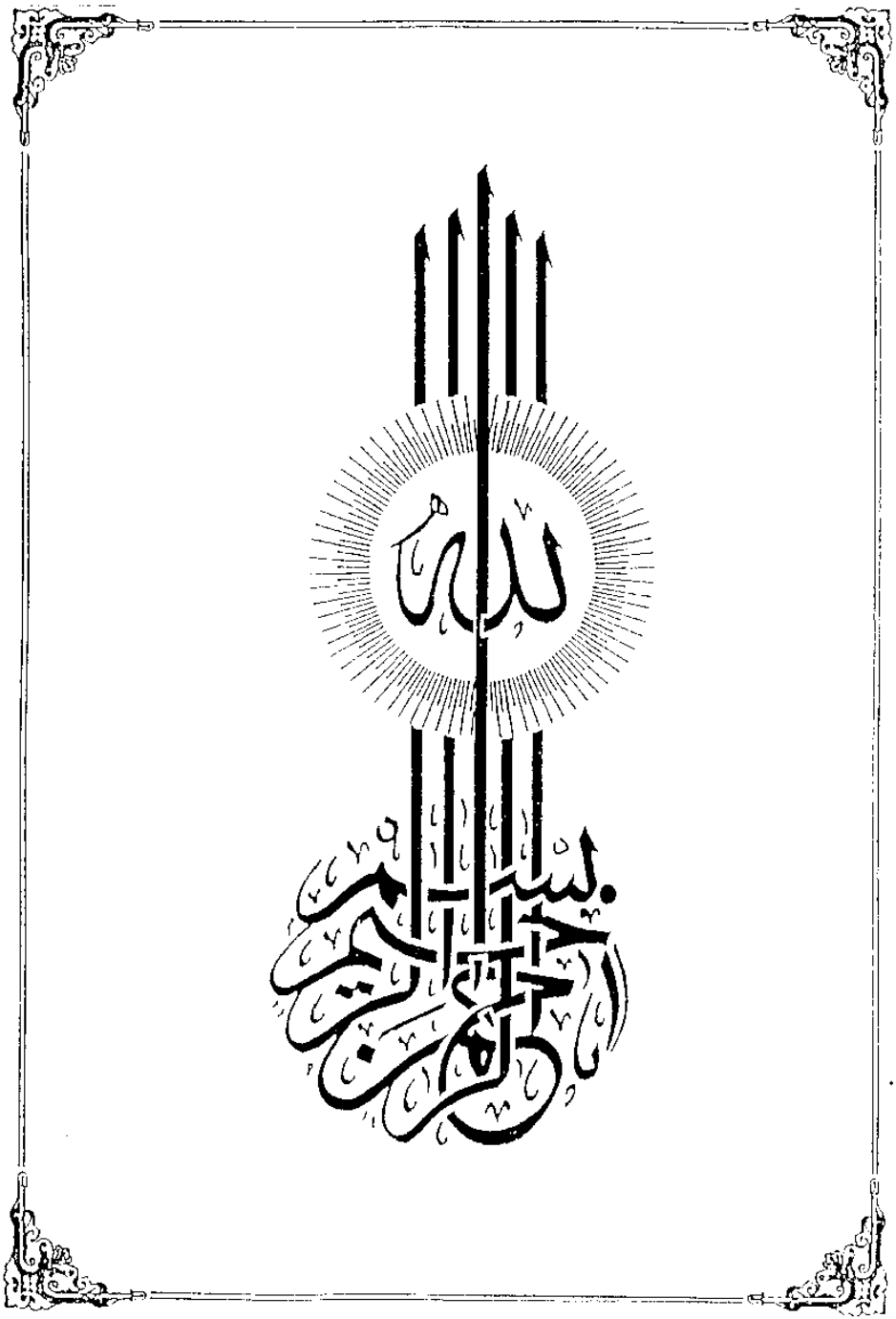


Acknowledgment

The challenge of a study may not be easy to go through without the sincere guidance of experienced people who take to their heart the responsibility of education, doing so they give so much of their time and knowledge to help a candidate achieve his goal. The path is full of deviations and, so many times, irrelevance yet the devotion of such people helps straightening up the thoughts and clearing the logistics and sequence of relations.

I have been lucky to have this kind of guidance from Professor Magdy El-Barbary whose patience and care carried me safely towards my goal. His clear vision encouragement and systematic thoughts have so many times taken me from my distractions and back to the theme. The careful advice that has been given to me by Dr Sherif Embabi has been so valuable. His discussions and vivid remarks helped me in producing this study.

I hereby thank my tutors; Professor El-Barbary and Dr Embabi deeply for all what they have done.

Contents

Introduction

Primary congenital glaucoma has special clinical features; the child's eye is expansile and the anterior segment expands with pressure if the onset is in the first three years, the visual system is still developing and visual loss can occur not only from corneal scarring and optic nerve damage but also as a result of amblyopia, the third feature is that you are dealing with a child and this needs teamwork both in elucidating the signs of the disease and providing clues to adequacy of glaucoma treatment (Freedman, 1998). Its pathogenesis is still disputed; most observers have not been able to document ultrastructurally a continuous endothelial membrane, as initially advanced by Barkan. A location for the gene responsible for its development has been mapped to chromosome number 2 (Sarfarazi, 1995). The primary, definitive treatment is surgical. Both goniotomy and trabeculotomy ab externo give similarly good results in the majority of patients, but in the Middle East trabeculectomy gives good results as well (Krieglstein, 1986).

The prognosis in this disease is related to the time of its initial presentation, initial surgical intervention, degree of optic nerve damage as well as nature and quality of corneal enlargement and astigmatism. The inability to easily quantitate visual acuity and extent of visual loss in neonates necessitates following up of patients by measurement of corneal diameter, intraocular pressure and axial length. However, even these data should not be relied upon exclusively to determine the quality or quantity of success in primary infantile glaucoma (Anderson, 1983).

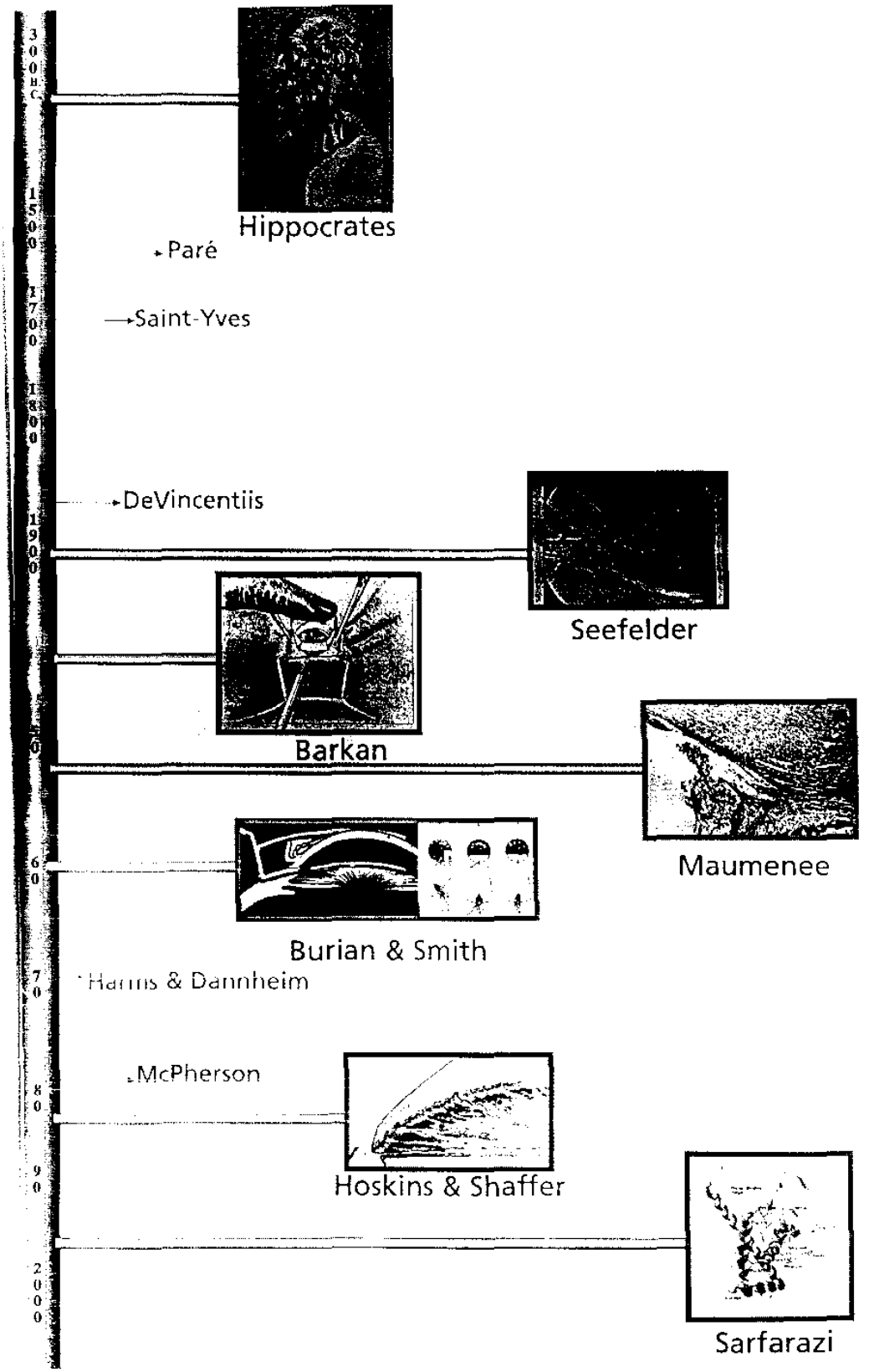
SECTION 1

History

The early writers, such as Hippocrates, Celsus and Galen, recognized congenital enlargement of the globe. They did not associate it with elevated intraocular pressure and they included in a single clinical entity all those conditions where the globe appeared to be of unusual size, exophthalmos amongst them. In the 16th century Ambroise Paré (1517-1590) introduced the term "ox-eye" to describe the enlargement of the globe: "Oeil de beuf (Buphthalmos) est une maladie d'oeil quand il est gros et eminent, sortant hors la teste, comme on voit les boefs les avoir." In 1722 Saint-Yves attempted to classify the various forms of ocular enlargement in "De la grosseur demesuree du Globe de l'oeil." He included (1) the naturally large eye, (2) exophthalmos and (3) increase in the size of the eye due to an abundance of aqueous humor (deLuise and Anderson, 1983; Luntz, 1984).

The first reference to elevated intraocular pressure in association with enlargement of the globe is credited to Berger in 1744. Despite numerous descriptions of buphthalmos in the sixteenth century, especially those of Paré, and pathologic observations of *Schiess-Gemesus* in 1862, it remained for *von Muralt* to segregate this disorder as a type of glaucoma in 1869. Both he and von Graefe in 1869 believed that the corneal enlargement was primary and that the ocular hypertension resulted from damage to the corneal nerves. Other authors felt that the glaucoma was secondary to uveal inflammation (Raab, 1876; Gallenga, 1885).

In the early part of the 20th century, the changes in the angle were shown to be primary and the inflammatory changes secondary by Reis in 1905 and Seefelder in 1906. The distinction between a physiologically large eye or cornea and true glaucomatous enlargement of the eye was firmly



lished by Kayser in 1914 and Kestenbaum in 1919.
Barkan in 1949 described a persisting fetal membrane overlying the trabecular meshwork. This was confirmed by Worst (1966) who called it "the ... Barkan's membrane." Recent pathologic studies have found no evidence of Barkan's membrane, but have shown abnormal trabecular meshwork and ciliary body (Maul, 1980; Anderson, 1981).

Other investigators pursued the anatomico-pathologic aspect of primary congenital glaucoma especially JR Anderson, Kluyskens, Allen and Maumenee. And the need to classify the different developmental glaucomas was developed especially noted by Haas in 1955 when he noticed different response to surgery in different cases (Luntz et al., 1984).

The importance of early change in the optic nerve was stressed by Shaffer and Heatherington (1969). Earlier authors felt that anterior scleral distention protected the optic nerve from the effects of raised intraocular pressure(Luntz and Harrison, 1996).

DeVincentiis (1891) was the first to suggest an operation to incise the tissues of the anterior chamber angle to treat congenital glaucoma. Success was reported for many cases despite the limitations imposed by the technology available at that time. The incision frequently was made into Descemet's membrane or into the ciliary body rather than into the trabecular meshwork, and these incisions were unsuccessful. Scarring of the incision was also reported and could have resulted from too deep an incision of the meshwork, thus cutting into scleral tissue (Luntz et al., 1984).

Fig. 1-1. Diagramatic representation of the historical events in congenital glaucoma. *From top to bottom* Hippocrates (Rutkow, 1993), Section in angle of anterior chamber by Seefelder (Anderson, 1939). Goniotomy as shown by Barkan in 1948. Section in angle of anterior chamber by Maumenee (Maumenee, 1959). Trabeculotomy as shown by Burian and Smith simultaneously in 1960. Classification of developmental glaucomas by Hoskins and associates in 1984. Sarfarazi and co workers in 1995 map the main gene for primary congenital glaucoma.

Otto Barkan in 1938 modified this technique and named it goniotomy. The prognosis for congenital glaucoma was so poor that Anderson in 1939 said *"The future of Hydrophthalmia is dark. Little hope of preserving sufficient sight to permit the earning of livelihood can be held out to them*".

Seefelder in 1920 also said *"I know of no case of operated hydrophthalmia where undiminished sight has been retained till later life."* In reviewing several reported surgical series. he found that one patient in three was blind after surgery. One patient in three had visual acuity less than 6/60, and one in three had visual acuity better than 6/60. After age 25, no patient had better than 6/24 vision. In the collected series of unoperated patients, only one in four had vision better than 6/60. Two of four were blind by the age of 12. Sixty percent of patients 25 to 50 years of age were blind.

Before Barkan introduced goniotomy in 1938 there were several surgical techniques being practiced, but did not give good results. They include: sclerectomy, iridectomy, sclero-iridectomy, iridencliesis, iridotstasis & cyclodyalysis. The introduction of goniotomy started a new era for congenital glaucoma and the surgical techniques pioneered by Barkan in 1938, have remained essentially unchanged today; an alternative procedure, microsurgical technique in the form trabeculotomy, was independently and simultaneously described by *Burian* and *Smith* in 1960.
Gregersen and Kessing in 1977 compared results obtained by macrosurgery (i.e., goniotomy, trephine, and diathermy) to those obtained with trabeculotomy.

Other surgical modalities were tried, but are not practiced anymore as. Goniopuncture, Goniotripssy, trephine and Iridectomy with scleral cautery.

In 1973 Kwitko summarized the surgical procedures for congenital glaucoma as follows:

Trabecular Surgery

I. Trabeculotomy: the slitting open of the trabecular meshwork obstruction
   a. Internal transverse trabeculotomy (de Vincentiis, Barkan)
   b. Internal direct trabeculotomy, goniotripsy (Kwitko)
   c. Trabeculotomy ab externo (Smith, Mein and Burian, Harms and Dannheim)

II. Trabeculectomy (Cairns, Dellaporta)

By-pass Surgery

I. Iridectomy with scleral cautery (Scheie)
II. Trephine

Combined Trabeculum and By-pass Surgery

Goniopuncture (Scheie)

Recently the gene responsible for primary congenital glaucoma has been mapped to chromosome number 2 by Sarfarazi and associates in 1995. Gene therapy is in the horizons.