PRIMARY CONGENITAL GLAUCOMA

INTRODUCTION

"Primary infantile glaucoma is a condition in which the eye have an isolated maldevelopment of the trabecular meshwork without other associated developmental ocular anomalies or ocular disease that lead to increase intra-ocular pressure during the first years of life". "Primary congenital glaucoma" as a specific syndrome was described by **Shaffer and Weiss (1970)** as follows: It is the most common hereditary glaucoma of childhood, inherited as an autosomal recessive trait, with a specific angle anomaly consisting of absence of angle recess due to an anterior iris insertion directly into the trabecular surface. There are no other major anomalies of development. Corneal enlargement, clouding and tears in Descemet's membrane result from elevated intraocular pressure.

Morin (1986) gave a clinical description of primary congenital glaucoma, as such "Primary infantile glaucoma is characterized by an enlarged cornea and elevated intra-ocular pressure that causes optic nerve damage. It occurs in infancy and early childhood and is due to a genetically determined developmental defect of the trabecular meshwork which is present at birth but may manifest itself at a later date. The most common form of congenital glaucoma is primary infantile glaucoma.

It may be present at birth or may develop within the first year of life.If glaucoma develops later, after 3 years from birth when the globe enlargement can no longer occurs, it is known as juvenile glaucoma or late onset congenital glaucoma (Morin, 1986).

Shaffer (1976) defines primary congenital glaucoma as an elevated intraocular pressure usually within the first three years of life due to maldevelopment of the trabecular meshwork. Hoskins(1981) defines primary congenital glaucoma (primary infantile glaucoma): as glaucoma in infancy associated with an isolated trabeculo-dysgenesis that responds well to goniotomy and may be inherited in a recessive mode.

The many congenital glaucomas are uncommon or rare disorders. Their importance lies in the long lifetime course, and the potential severity of the disease (Weiss 1978). He defined congenital glaucoma as elevated intraocular pressure present at birth sufficient to cause damage to the optic nerve and/or secondary anterior segment changes.

The lack of an ideal method to save vision for those afflicted in the past led Anderson (1939) to state that "The future of patients with hydrophthalmia is dark, little hope of preserving sufficient sight to permit earning a living can be held out to them". In accordance with Anderson, Barkan (1942) stated that congenital glaucoma or hydrophthalmia (infantile glaucoma or buphthalmos) is per-

haps the most hopeless and certainly the most pathetic of ocular conditions requiring surgery. The end result, with or without operation is frequently blindness, and more often than not enucleation of one or both eyeballs is required.Despite aggressive management, congenital glaucoma still produces severe visual loss because of a combination of amblyopia, corneal scaring, refractive error and optic nerve damage (Morgan 1981).

Congenital glaucoma is one of the most important causes of blindness in early childhood. As any vision during child's formative years is worth fighting for, even if it is ultimately lost later in severe cases. Prompt and appropriate treatment is the difference between continuing vision for a child or a whole lifetime of darkness (R. Shaffer 1983).

In place of the gloom and despair in the last thirty years Valvo (1968) stated that "Fortunately with early diagnosis and microscopic surgical techniques the large majority of these eyes can be controlled if not completely cured. However in few patients who continue to show poor response to surgery such operations will delay the inevitable blindness, and allow the child to develop visual images that will be valuable to him in later life".