

# The Glaucomas

Volume I - Pediatric Glaucomas

Bearbeitet von  
Roberto Sampaolesi, Jorge Zarate, Juan Roberto Sampaolesi

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# Primary Congenital Glaucoma

# 1

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## History

Hippocrates, 400 years before Christ, and Celsus and Galen, 100 years after Christ, noted the phenomenon of buphthalmos but did not relate it to glaucoma.

In 1561, Ambroise Paré wrote: “Oeil de boeuf est une maladie d’œil quand il est gros et éminent, sortant hors de la tête, comme voits les bœufs les aboir,” which is one of the first descriptions of buphthalmia. The observations of Schiess-Gemuseus in 1863 and 1884 [1, 2] are also important, but in fact it was Von Muralt [3] who related this alteration to a type of glaucoma. These observations were later confirmed by Von Hippel, Parsons, and especially by Seefelder [4] and Seefelder and Wolfrum [5], who demonstrated the true pathogeny of this disease.

Taylor [6, 7] was the first to publish Carlo De Vincentiis’s surgical technique for the treatment of glaucoma [8, 9]. Designed in Naples and known as the “incision of the angle formed by the iris and the cornea” or internal sclerotomy, this technique had the same requirements as goniotomy, since it was performed with a small sickle-shaped knife (the De Vincentiis knife), specially manufactured to prevent aqueous humor from overflowing. Ocular fixation was good and the incision was nontraumatic and superficial to prevent damage to other structures of the chamber angle. He used the technique for all sorts of glaucomas and it

was the first blind goniotomy, though it is actually an ab interno trabeculotomy. It was then abandoned for 30 years, perhaps because it was reported only in a local Italian journal and because its author died too soon. In 1900, Scalinci [10] presented 13 cases of congenital glaucoma successfully operated using this technique.

The technique was forgotten until Otto Barkan [11] revived it as an operation for congenital glaucoma and called it goniotomy (cutting the angle).

Among those who studied the anatomopathological aspects of this disease are Kluysken [12], Shaffer [13], and Allen et al. [14]. One step forward was the introduction of trabeculotomy by Burian, Harms, and Paufigue, which improved the prognosis of congenital glaucomas greatly. For the first time in 1987, R. Sampaolesi [15] introduced combined surgery: trabeculotomy and trabeculectomy in a single surgical session for refractory congenital glaucomas. In 2005, he applied Koslov’s technique of nonpenetrating deep sclerectomy to find the Schlemm, changing Harms’s operation for the latter (see Chap. 15).

## Concept

Congenital glaucoma is an infrequent disease, an inherited developmental defect, occurring within the 1st year of life and referred to the ophthalmologist within the first 24 months.

It is characterized by a congenital anomaly of the chamber angle at the level of the trabecular meshwork, which obstructs the aqueous humor outflow pathways, leading to high intraocular pressure (IOP) and to an early elongation of the eyeball, corneal enlargement, and corneal edema. If immediate and proper surgical treatment is not provided, it produces progressive impairment leading to serious damage of the entire eye, particularly in the optic nerve.

Congenital glaucoma is a complex disease. It is completely different from simple adult glaucoma, due to the anatomical and physiological features of the eyes of newborns. From its clinical manifestations, its pathophysiology, and anatomopathological findings, its immediate cause has been suggested to lie with

goniodysgenesis caused by arrested development of the chamber angle.

In this chapter, only cases in which the disease presents from birth to 24 months of age will be discussed. These are pure congenital glaucomas, with definite clinical features, progression, and anatomopathology. They have been named primary congenital glaucomas, primary infantile glaucomas, or developmental glaucomas.

Goniodysgenesis may develop more mildly than is seen in congenital glaucoma and, in this case, ocular hypertension will occur progressively and later, because of factors that remain unknown. These cases belong to completely different clinical forms known as juvenile glaucoma, late congenital glaucoma, or goniodysgenesis, which manifest at 5, 10, or 18 years and are even more frequent in adults until the age of 40 years.

In our experience [15–17] and in that of Kwitko [18] and Walton [19], a dividing line between infantile congenital glaucomas and late congenital glaucomas can be drawn at the age of 4 years, because from this age, the axial length can no longer grow as a consequence of elevated IOP. Glaucomas associated with ocular and systemic malformations belong to an independent group (see Chap. 22).

Infantile congenital glaucoma is the most severe of glaucomas, which means that just as in the acute glaucomas in adults, surgery has to be performed immediately, without losing time. Shaffer did the examination under general anesthesia in the same room where surgery would be performed. Before the examination under general anesthesia, he asked the parents for their approval to proceed with surgery if the diagnosis was positive.

The degree of ocular damage depends on the length of the period between the appearance of the first clinical manifestations and surgery, or on the failure of surgery to regulate IOP. Damage may include glaucomatous optic disc cupping; visual field loss; ocular distension with acquired refractive errors and macular disorders; Descemet membrane and endothelium tears, which, if located centrally, cause severe visual loss (in this case a corneal graft is needed immediately); peripheral retinal disorders; anisometropia; amblyopia with or without strabismus, etc.

Since perimetry, visual acuity, and macular function tests are useless for diagnosis and monitoring the progression of this disease so early in life, echometry, applanation tonometry with pachymetry, or better yet with Pascal tonometer measurements, and gonioscopy have become the most valuable tools for these purposes.

In addition to all the postoperative checks, visual acuity must be constantly monitored with the preferential looking test.

These advances developed during the last two decades have changed the attitude of ophthalmologists, who are now optimistic when they encounter this disease, since early diagnosis can be made with family education and the cooperation of the pediatrician.

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## Epidemiology

The prevalence in the population is 8:100,000 children [20] (congenital glaucoma occurs in 1 out of 10,000 births). In 80% of cases, it is bilateral. It affects males in 70% of cases. It is the most frequent cause of early blindness of congenital origin: 50% of cases with blindness from glaucoma.

The most complete papers on the subject are those authored by Anderson [21]; Westerlund [22]; Kluyskens [12]; Gallenga and Mateucci [23]; Van der Helm [24]; Carvalho and Calixto [25]; Shaffer [26]; Kwitko [18]; Jerndal et al [27], and De Luise and Anderson [28] (Table 1.1).

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## Heredity

Most cases are sporadic, nonhereditary, and nonfamilial. From 10% to 12% have a family tendency and an autosomal recessive heredity pattern as reported by François [34] and Duke Elder (1964) [35].

It is striking that in family cases, father and son are the members affected, an uncommon trend in autosomal recessive heredity. In 1972, Merin and Morin [36] studied 64 families and concluded that heredity is multifactorial both in congenital and in open-angle glaucoma.

This is consistent with the results obtained by Demanais in 1981 [37]. In identical twins, both are affected by congenital glaucoma [38], though Fried et al. [39] described the case of a pair of monozygotic twins where only one was glaucomatous; this suggests a role of nongenetic factors.

Kluyskens [12] was the first to create genetic maps according to goniodysgenesis.

The cases studied by Jerndal et al. from 1970 to 1974 [27] demonstrated that goniodysgenesis is a dominant disease in congenital glaucoma. When the disease runs in the family, cases of congenital glaucoma, late congenital glaucoma (juvenile glaucoma), and adulthood congenital glaucoma occur. Manifestations are varied, as shown by one family studied by Jerndal et al., in which the father married twice: from one marriage, he had one son with congenital glaucoma and from the other, one with late congenital glaucoma. The father, aged 46 years, has glaucoma with severe goniodysgen-

**Table 1.1** Frequency, bilaterality and gender

Frequency	0.01%–0.07% of ocular diseases	Anderson 1939 [21]	
	0.008% ± 0.0012% of the population	Westerlund 1947 [22]	
	0.0056% of the population	Van der Helm 1963 [24] with Sturge-Weber and Krause syndrome not included	
Bilaterality	Gros 1987 [29]	116 cases	64% bilateral
	Seefelder 1906 [30, 31]	47 cases	67% bilateral
	Brons 1937 [32]	127 cases	81.4% bilateral
	Anderson 1939 [21]	94 cases	86% bilateral
	Van der Helm 1963 [24]	630 cases	75.3% bilateral
	Sampaolesi 1991 [33]	875 cases	78% bilateral
Gender	Van der Helm 1963 [24]	425 males (68%)	202 females (32%)
	Sampaolesi 1991 [33]	595 males (68%)	280 females (32%)

esis. This chapter and the others on pediatric glaucomas, will describe several families whose family trees are consistent with those studied by Jerndal et al.

From a practical point of view, when their first child is diagnosed with congenital glaucoma, parents want to know the risk of having another child with the same disease. The answer is that one out of four children is affected, though this is not actually predictable (See Chap. 7) and the chance of a second child having the disease is small: 1%–3%.

## Prevalence

Primary congenital glaucoma occurs in all ethnic groups. The birth prevalence, however, varies worldwide:

- 1:5,000–22,000 in Western countries;
- 1:2,500 in the Middle East;
- 1:1,250 in the Rom (Gypsy) population of Slovakia [40];
- 1:3,300 in the Indian state of Andhra Pradesh, where the disease accounts for approximately 4.2% of all childhood blindness [41].

In Saudi Arabia and the Rom population of Slovakia, primary congenital glaucoma is the most common cause of childhood blindness [40, 42].

## Etioopathogenesis

The study of etioopathogenesis is based on the clinicopathological correlation of gonioscopic findings in relation to the pathological anatomy of specimens obtained during surgery (when combined surgery was required). We were pioneers in the study of trabeculectomy specimens and the French authors followed.

Proper interpretation of the pathology and etioopathogenesis of pediatric congenital glaucomas is based on:

1. Knowledge of the embryological development of the chamber angle;
2. Knowledge of the normal chamber angle in children, its gonioscopic appearance, and its variations within normality;
3. Pathological gonioscopic findings in pediatric congenital glaucoma;
4. Pathological anatomy of the specimens obtained from combined surgery procedures performed in cases of refractory glaucoma;
5. Correlation between the gonioscopic picture and the pathological anatomy.

In addition to the items above, it should be remembered that trabeculectomy specimens always belong to very severe or advanced cases within the first 24 months of age, since either goniotomy or trabeculotomy are per-

formed in mild cases. In this large group of patients, anatomopathological verification is therefore impossible.

Another factor also leading to misinterpretations is disagreement as to the nomenclature used: wholly different words are used to refer to the same element, when they are actually synonyms. All the following terms refer to the anomalous tissue obstructing the trabecular meshwork and preventing the aqueous humor from reaching its natural outflow pathways:

1. Pectinate ligament: a term adopted by comparison with the structure located at the chamber angle in ungulates (horses).
2. Anterior iris insertion (or high insertion of the iris). This is a misinterpretation, since this anomalous tissue overlaps with the iris root, reaching the scleral spur and covering the ciliary body band or, even further, it extends up to the Schwalbe line and covers the trabecular meshwork. It should be kept in mind that the iris root never shifts and, even in congenital glaucomas, it inserts at the usual place at the ciliary body band, which is made up of the inner surface of the ciliary muscle. It is simply an apparent high insertion of the iris.
3. Fetal mesoderm.
4. Pathological mesodermal remnants. From now on we will use this term here and the reason for our choice will be explained later.

A review of useful literature with a summary of the findings of each author follows:

- Raab [43] reported the first demonstration of an obstruction of the iridocorneal angle in a congenital glaucoma case.
- Taylor [44, 45] was the first to publish the surgical technique conceived by Carlo De Vincentiis, from Naples, designated by the author as *l'incisione dell'angolo irideo* (incision of the iridic angle). This was the first blind goniotomy.
- Scalinci [46] presented 13 cases of pediatric congenital glaucoma operated on with this method, with successful results. This technique was not actually put into practice until 1938, by Barkan.
- Barkan [47–49] proposed a goniotomy with visual guidance by means of a gonioscopic contact lens he designed, in order to remove “an imperforated membrane covering the angle of the anterior chamber, and preventing the aqueous humor from outflowing, thus leading to ocular hypertension.”
- Barkan [50] described a “transparent or semi-transparent membrane in a vertical position from Schwalbe’s line to the iris in the angle. The vertical position of this tissue is in contrast with the hori-

zontal position of the iris. After goniotomy, the iris falls backwards, as if it had had a high insertion, and uncovers Schlemm’s canal with its normal anatomic relationships with the other structures. Therefore, the angle, free from any obstructing tissue, is available for the aqueous humor.”

- Barkan [51] described the pathological anatomy in order to correlate it with the gonioscopic findings. He describes “a membrane lining the inside of the angle from Schwalbe’s line to the iris,” and he reports the presence of mesodermal remnants inside or under this membrane.
- Maumenee [52] described the absence or aplasia of the spur, but this is one of the rare quotations in the literature that has not been verified by other authors. We have always found the spur in more than 300 specimens studied.
- Shaffer [53] described an “abnormal mesodermal reticulum” in a case with apparent high insertion of the iris.
- Hansson and Jerndal [54] demonstrated that the chamber angle in congenital glaucoma resembles that of a normal fetus at stage 200–240 mm, 7 months of gestation.
- Sampaolesi et al. [17] conducted a study with light microscopy and surface electron microscopy, where they described what they called pathological mesodermal remnants obstructing the trabecular meshwork covered by a membrane, which are stained with dark silver colorants (Gomori’s stain) due to the large amount of reticulin fibers contained in them, which are the same as those on the tissue obstructing the chamber angle in normal fetuses at month 7 of gestation (200–240 mm). This morphology of the chamber angle in primary or pure congenital glaucomas resembles the morphology of the normal developing chamber angle.
- Anderson [55–57] makes one of the most interesting contributions: his explanation of the movement of the different components of the chamber angle during its formation and, fundamentally, the mechanism causing the ciliary muscle to shift forward in congenital glaucoma, which will be discussed later.
- Allen et al. [58] hypothesized that the formation of the chamber angle may be due to what they termed cleavages (separation between mesodermal layers) caused by an uneven growth of the structures of the chamber angle from the 5th month. But Kupfer and Kaissner-Kupfer [59], some years later, demonstrated that this theory was based on a critical mistake, thus invalidating it.

The theory of migration of neural crest cells has been considered [56], but Alvarado has reported otherwise [60, 61].

There are other important papers in the literature, such as those by Smelser and Ozanics [62]; Mann [63]; Holmberg [64], and Maul et al. [65], confirming the findings detailed above.

A very important paper has been specially reserved for the end of this chapter because of its great value: in 1906, Seefelder [4] and Seefelder and Wolfrum [66], for the first time described the pathology of congenital glaucoma as a detention in development at the 7th month of gestation; they presented the pathological anatomy of congenital glaucoma and compared it with the histology of a normal fetus at the 7th month of gestation in order to show their similarity. More recently, Worst [67] published a similar image in his book; it is a specimen published by Castelli [68] and interpreted by himself. Finally, according to Jerndal and colleagues [27], of the different theories – cleavage, atrophy, and resorption – postulating a detention in development is the most consistent with the way of thinking of current authors.

The original papers on the pathological anatomy of our specimens will be discussed in Chap. 15.

Our research into congenital glaucoma was conducted following the following steps:

- 1960–1970: IOP (normal and pathological);
- 1970–1980: echometry in the diagnosis and follow-up;
- 1980–1990: functional results in operated primary congenital glaucomas;
- 1982–1983: optic disc changes in congenital glaucoma;
- 1990–2006: confocal tomography of the optic nerve head;
- 1983–2006: surgical methods to apply according to the type of chamber angle: type I, type II, refractory glaucoma, and according to the echometric values. New evaluation of the anatomical and functional results of surgery 12–35 years after surgery.

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