CORNEAL CHANGES IN PRIMARY CONGENITAL GLAUCOMA

BY *J. D. Morin,* MD AND (BY INVITATION)
W. R. Coughlin, MD

IN A RECENT REVIEW ON PRIMARY CONGENITAL GLAUCOMA (PCG),[1] IT WAS observed that there was marked variation in the presenting clinical picture and that this depends on the age of clinical onset (AO), time of diagnosis (TDx) and control of intraocular pressure (IOP). This review suggested the following:

CLINICAL MANIFESTATIONS OF PRIMARY CONGENITAL GLAUCOMA

1. The earlier the age of clinical onset, the greater the change in corneal chord length or corneal diameter (CD) between the diagnosis and follow-up.
2. A close parallel exists among changes in intraocular pressure, corneal diameter and cup-disc ratio, such that for each 10 mm Hg increase in IOP, there is a corresponding increase of 0.5 mm CD and 0.2 increase in the cup-disc ratio.
3. Good control of IOP (≤ 25 mm Hg) between the AO and extended follow-up significantly affects the corneal chord length.

   (a) So that in the presence of a sustained increased IOP (ie, 25 mm Hg) there is 2.5 times greater increase in the CD than there is when the IOP is maintained below that level from the age of onset until the extended follow-up. (b) A decrease in the chord diameter (≤ 2.0 mm) was present in 11 of 23 cases of early onset PCG over an extended period of time during which the IOP was kept below 25 mm Hg, and when IOP was above that level, there were no cases of apparent corneal shrinkage.

The above observations led to the following questions:

(a) Will good control of IOP prevent, minimize, or reverse the anticipated increase in corneal diameter in the developing eye? (b) If there is a parallel change between changes in IOP, CD, and C/D, may one safely monitor changes in corneal chord length as an index of underlying disease progression? (c) In cases of uncontrolled PCG, is there a critical period in terms of clinical onset preferentially inducing changes in the cornea and/or sclera? (d) Are there additional etiological factors operating within any given reference period, such as differences in the biochemical framework of the cornea and sclera (ie, mature/immature collagen, presence or absence of cross linkage) which might influence the development and manifestations of PCG.

A study was undertaken to:

(a) develop a simple, practical and accurate method of measuring corneal chord length which would provide a permanent record of the cornea size as it is at that point in time. (b) conduct a comparison study of apparent corneal chord length shrinkage using the standard conventional caliper technique and the corneal contact lens color photographic method. (c) re-evaluate PCG in terms of AO to ascertain whether or not there is a critical period in the developing eye, such that underlying pathophysiological changes might preferentially affect the clinical presentation of the cornea and/or sclera.

METHOD

A series of congenital glaucoma cases were monitored by the "corneal contact lens color photography, for measuring changes in corneal chord length" which consisted of the following:

1. A contact lens of known chord length (eliminates the need to use standard base curves).
2. A color photograph is taken with an appropriately sized contact lens sitting on the underlying cornea.
3. The corneal chord length is measured on a tangent screen onto which is projected the color transparency photograph of the contact lens sitting on the cornea. The

limbal margin used as a reference landmark for the chord length measurement.

The side of the limbal margin used as a reference point is not critical providing the same point (medial or lateral limbal zone) is used for subsequent measurement. Having the color transparencies from the original and interim follow-up monitoring periods available, one can then re-calculate all measurements using the same reference base for comparison.

It is important to use a contact lens that differs in chord length from that of the underlying cornea. Unless the two diameters differ, it is impossible to appreciate the true boundary of the underlying cornea when the color transparency is projected on the tangent screen. One may use a contact lens that is either larger or smaller than the cornea so that the projected image of the cornea and the lens can readily be accurately measured on the screen.

The distance between the camera and the patient as well as between the projector and the tangent screen is not critical for neither distance is used to calculate the corneal chord length. Use of a simple formula based on ratios of the cornea and lens to their projected ratio enables one to calculate the chord length of the cornea.

Example:

$$\frac{CCL}{CCL_{ts}} = \frac{CLL}{CLL_{ts}}$$

Where CCL = corneal chord length
CCL_{ts} = corneal chord length on tangent screen
CLL = contact lens chord length
CLL_{ts} = contact lens chord length on tangent screen

Knowledge of three of the four variables enables one to calculate the fourth.

This technique enables one to monitor changes in the corneal chord length and it provides a permanent record for subsequent monitoring of changes.

RESULTS

A comparison study of apparent corneal shrinkage using the conventional caliper technique and the corneal contact lens color photographic method were made on ten cases of PCG in order to ascertain whether the previously reported findings of a reduction in corneal chord length were real or apparent. Three examples are seen in Table I.

TABLE I: COMPARISON CHANGES IN CORNEAL CHORD LENGTH

	Follow-up EUA	Caliper Method	Contact Lens Method
A	Initial exam	13.5 mm	14.5 mm
	6 mos.	13.8 mm	14.0 mm
	12 mos.	14.5 mm	14.0 mm
B	Initial exam	13.8 mm	12.5 mm
	2 mos.	13.5 mm	12.0 mm
	11 mos.	14.3 mm	12.0 mm
C	Initial exam	14.0 mm	14.0 mm
	4 mos.	14.8 mm	14.5 mm
	10 mos.	14.5 mm	14.9 mm

Analysis of these ten cases revealed that the reduction in corneal chord length was real and not artifactual. Although the reduction was less than 1 mm in two of the three cases illustrated, it is true that the significance is even more readily apparent when one realizes that this reduction in chord length occurred at a time when the normal corneal maturation and enlargement is taking place. The actual reduction in chord length obtained following the control of intraocular pressure is most likely greater.

A more detailed study of 61 cases of PCG suitably matched for pairs (age of clinical onset—follow-up duration) was carried out in an attempt to correlate the age of onset with changes in intraocular pressure and corneal chord length (Table II).

TABLE II: FOLLOW-UP MONITORING OF CORNEAL CHORD LENGTH IN PRIMARY CONGENITAL GLAUCOMA

Range of clinical onset:	birth — 47 mos. of age
Range of follow-up:	3 mos. — 120 mos.
Changes in corneal chord length:	
— Unilateral increase	9 cases
— Bilateral increase	24 cases
— Unilateral decrease	8 cases
— Bilateral decrease	6 cases
— No significant change	14 cases

A comparison of the result of unilateral vs bilateral PCG reveals a marked difference in the chord length behavior. The results are illustrated in Table III as mean values of an increase (+) or decrease (−) for matched age of onset group in unilateral and bilateral cases.

TABLE III: MEAN CHANGES IN CORNEAL CHORD LENGTH

Age of clinical onset:	0-3 mos	4-12 mos	13-24 mos
Unilateral PCG	−1.30 mm	−0.40 mm	−1.00 mm
Bilateral PCG	−1.00 mm	−0.40 mm	−1.30 mm
Unilateral PCG	+0.66 mm	+0.66 mm	+0.60 mm
Bilateral PCG	+1.33 mm	+1.86 mm	+0.70 mm

DISCUSSION

If one is to attach significance to any corneal chord length, one must do so in light of accepted normal growth patterns. The cornea undergoes a maturation from birth to reaching its adult diameter around three years of age. Normal growth range was derived from a review as seen in the table from "The Eye in Childhood."[2] A normal maturation range from 9.0 to 11.80 mm horizontal diameter represents the anticipated growth in a normal healthy eye.[3] Whether such maturation occurs in PCG is unknown. But as it may, any change in corneal diameter during the first five years of life must be considered in light of such anticipated maturation correction factor. Apparent increase in chord length may in fact represent normal maturation growth and true increase may be considerably less. Conversely, an apparent decrease in corneal length during the first five years of life may actually represent less than the actual amount. However, to say that good control would result in true reduction of corneal chord length at a time when normal maturation would increase it is difficult to fathom. What is important to realize is that any reduction in chord length represents an even greater amount than that amount recorded. It is not known whether the natural maturation growth phase is linear and at any one time what the true correction factor is. For this reason, we elected not to use maturation correction factor to calculate the change in chord length realizing however that an increase recorded may be too high while the decrease shrinkage may be too low.

In either case, knowing the initial and final measurements provide one with an objective parameter for monitoring changes in PCG, providing the method employed is accurate and consistent.

The conventional caliper technique to monitor changes in corneal diameter is simple to operate. Unfortunately, there are many disadvantages which prevent us from detecting minor changes in chord length. There is marked variation in readings taken by independent observers. The scale readings are gross and at best one does not have a permanent objective measurement of the cornea available for future evaluation. There is no permanent record of corneal diameter at that point in time. Any error made at measuring the chord length at that particular examination is permanent and not available for evaluation.

Whether the distention of the eye is due to the unique factor of high acid of soluable collagen fraction in the sclera is uncertain. However, if one can establish a specific enzyme defect in the scleral collagen or in the cross-linkage formation the door to the prevention of buphthalmos is open, along with research into the causes of myopia.

SUMMARY

A control study for monitoring changes in horizontal corneal diameter has been described. A decrease in horizontal diameter was obtained at a time when the normal eye was continuing to grow irrespective of the age of onset and the clinical presentation of the disease, suggesting that another factor may be used to monitor the process of PCG.

REFERENCES

1. Morin JD, Merin MD, Sheppard MD: Primary congenital glaucoma—A survey. *Can J Ophthalmol* 9:17, 1974.
2. Glaucoma in children. In the Ophthalmologic Staff of the Hospital of Sick Children, Toronto: *The Eye in Childhood*. Chicago, Year Book Medical Publishers, 1967, p 261.
3. Duke-Elker: *System of Ophthalmology*. London, Henry Kimpton, 1961, vol II, pp 93-94.

DISCUSSION

DR A. EDWARD MAUMENEE. I wish to thank Doctors Morin and Coughlin for sending me a copy of their presentation well in advance of the meeting and allowing me to discuss their interesting paper.

The increase in corneal diameter, stretching of the globe producing myopia, and increased cup/disc diameter in children who have elevated intraocular pressure has been well documented for many years. This has been thought due to a greater elasticity of the collagen fibers in children rather than a basic abnormality in the collagen or ground substance in congenital glaucoma. The same stretching of the globe may occur in eyes of normal children up to two to four years of age who have developed secondary glaucoma from trauma.

The diagnosis of congenital glaucoma and the follow-up of these patients is frequently difficult. First, because the accuracy of tonometry is altered by the general anesthesia or sedation needed to take the pressure, by the thickness and consistency of the cornea, the altered corneal diameter and the marked variation in intraocular pressure from day to day or even hour to hour in congenital glaucoma.

The appearance of the angle of the anterior chamber on gonioscopy may be suggestive of congenital glaucoma but is certainly not always diagnostic.

The cupping of the disc may be helpful but large cups of 0.5 to 0.7 of the disc can occur in 5 to 10% of normal children.

Therefore, the size of the corneal diameter has frequently been used both in the diagnosis and the follow-up of children with congenital glaucoma. Doctor Morin's technique of using photography and a contact lens appears to be more accurate and gives a better documentation of these changes than the previously used technique of caliber measurements. It should be pointed out, however, that both the corneal curvature and limbal markings in children particularly those with congenital glaucoma may differ from those of adults. Thus, in a normal infant, the radius of the corneal curvature is less than that of an adult and therefore protrudes forward from the scleral spur more abruptly than an adult. In congenital glaucoma the external limbus or insertion of the conjunctiva and Tenon's capsule into the cornea slides forward up to 5 mm in front of Schwalbe's line in buphthalmic eyes. Thus, it is frequently difficult to determine the true corneal diameter or chord length if the external limbus is used rather than transillumination to detect the point of insertion of the iris into the scleral spur.

Doctor Morin's observations of shrinkage of the corneal diameter in controlled congenital glaucoma are new to me. The data in his paper which he sent to me were insufficient for me to analyze his observations. It has been well documented that the cup/disc ratio decreases when the intraocular pressure is controlled in congenital glaucoma. This occurs very rapidly after the intraocular pressure is reduced and is thought to be due to a shrinkage of the scleral canal of the optic nerve. It will be interesting to see a more detailed presentation of his data as to the time of shrinkage of the corneal diameter after pressure control and the other factors such as age of onset, duration of the glaucoma, the degree of elevation of pressure, the corneal diameter, etc. which might influence this shrinkage.

I wish to thank Doctors Morin and Coughlin again for allowing me to discuss their very interesting observations.

Dr Marshall M. Parks. I wish to add another parameter to this discussion and also ask the author a question. It came to my attention several years ago that one of the best parameters available in following infants after glaucoma surgery was by refraction. Similar to the disc-cup ratio change after normalization of the pressure also reversal or reduction of the myopic refractive error occurred. The myopia reduction occurred over a period up to two years in the children below three years of age. As a result of this observation I ask the author if any correlation was made between the corneal size change he reported and a change in the cycloplegic refraction error among the patients evaluated following normalization of their pressure.

DR ARTHUR JAMPOLSKY. The author has pointed out the inaccuracy inherent in the caliber method of measuring corneal diameters, and has recommended a photographic disc method. However, since some of these materials may not be readily available, I would like to point out an alternative for accurately measuring corneal diameters (or pupillary diameters), which is accurate to within one tenth of a millimeter. This simple method takes advantage of the extremely accurate alignment mechanism of the visual system, and also the ability to very accurately match symmetrical halves.

If one matches a series of blackened half-circles (over-laying the lower half of the cornea) so as to match the upper corneal half, and complete the circle, one has taken advantage of some of the most accurate psycho-physical measurements that it is possible to make.

One "aligns" the continuity of the nasal and temporal corneal (or pupillary) borders, as well as match the upper and lower symmetrical halves. This can be done quickly and easily with extreme accuracy and rulers such as these are easily made part of every clinician's armamentarium.

DR PAUL LICHTER. I would like to ask Doctor Morin if in his study he has had the opportunity to evaluate the optic disc relative to the changes in the cornea. He mentioned in his abstract that these were correlated with intraocular pressure and I wondered if he noticed any difference in the sensitivity of the optic disc changing with elevated pressure vs the change in the size of the corneal chord length.

DR J. DONALD MORIN. I thank Doctor Jampolsky for his suggestion of an accurate alignment method to measure the corneal diameter. When I initially started dealing with children with congenital glaucoma I was befuddled as to the accuracy of the measurements. They were as different as the observers that were doing them. If we have another method of trying to accurately make this measurement I'll be very happy to try and use it. In respect to the data base, [slide] I just wanted to show where we got some of this data. We were using these various data that we obtained in a large survey and we found that there was a significant difference in the amount of corneal change. Here we are seeing an increase of 1.25 mm in someone who's intraocular pressure was greater than 24 mm Hg whereas when the intraocular pressure was controlled the corneal diameter was increased by about 0.5 mm. As far as the relation to age of onset we broke them down into under three months, three months to three years and over three years. As I study this disease, I think it could be categorized differently. I think we should start looking at the child who was born with congenital glaucoma, the child who developed the disease between two months and a year and then from a year on because behavior patterns and the prognosis for a good therapeutic result seems to revolve around the age of onset more than anything else. The corneal diameter starts at about 11.96 mm in these youngsters at birth to three months and goes to 13.7 mm in contrast to those who are over three years of age when it starts about 12.3 mm and stays about the same during the subsequent follow-up.

Doctor Lichter asked me about the results of correlating the intraocular pressure and corneal diameter. If you notice that the corneal diameter does not change when you are able to reduce the intraocular pressure by about 25 mm Hg that is not to say that most of the youngsters with congenital glaucoma had an intraocular pressure in the range of 50 mm Hg but that when there is a profound decrease in the intraocular pressure we notice that there is a marked decrease in the size of the cornea. Although we really didn't see anybody with a corneal diameter change of 4 mm we did see a change of 2 mm and finally we did correlate it with the cup-disc ratio. When the corneal diameter decreases the cup-disc ratio also decreases. If there is a decrease of 2 mm then the change in the cup-disc ratio is on the order of -1 or more so I think that the accurate measurement of cornea is really becoming important and the ways to do this I think are really just starting to open up with photographic methods and easier methods. In respect to Doctor Parks' question no, we haven't been able to accurately refract these patients. We seem to get hazy media and we just cannot refract them. What we would like to do and what we are about to embark upon is an accurate measurement by the ultrasound method trying to see if the axial length of the eye changes.