

Comparative evaluation of RetCam vs. gonioscopy images in congenital glaucoma

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Purpose: To compare clarity, exposure and quality of anterior chamber angle visualization in congenital glaucoma patients, using RetCam and indirect gonioscopy images. **Design:** Cross-sectional study Participants. Congenital glaucoma patients over age of 5 years. **Materials and Methods:** A prospective consecutive pilot study was done in congenital glaucoma patients who were older than 5 years. Methods used are indirect gonioscopy and RetCam imaging. Clarity of the image, extent of angle visible and details of angle structures seen were graded for both methods, on digitally recorded images, in each eye, by two masked observers. **Outcome Measures:** Image clarity, interobserver agreement. **Results:** 40 eyes of 25 congenital glaucoma patients were studied. RetCam image had excellent clarity in 77.5% of patients versus 47.5% by gonioscopy. The extent of angle seen was similar by both methods. Agreement between RetCam and gonioscopy images regarding details of angle structures was 72.50% by observer 1 and 65.00% by observer 2. **Conclusions:** There was good agreement between RetCam and indirect gonioscopy images in detecting angle structures of congenital glaucoma patients. However, RetCam provided greater clarity, with better quality, and higher magnification images. RetCam can be a useful alternative to gonioscopy in infants and small children without the need for general anesthesia.

Key words: Congenital glaucoma, gonioscopy, Retcam

Congenital glaucoma results from an anomaly of the filtration angle, which may be secondary to arrested or abnormal development of the trabecular meshwork, both preventing egress of the aqueous. The long-term prognosis in congenital glaucoma is dependent on the type and extent of angle anomalies in each case (Hoskins) and therefore imaging of the angle becomes essential part of management.^[1]

Direct gonioscopy has been recommended in infants with congenital glaucoma. This is difficult, requiring examination under anesthesia, a handheld microscope, a special focal illuminator and expertise. High intraocular pressures induce corneal epithelial and stromal edema in infantile eyes and Haab striae, i.e., breaks in Descemet membrane, which causes irregular astigmatism and central corneal opacification, preventing adequate visualization of the angle and anterior segment through the central cornea, and therefore adequate direct gonioscopy.

The RetCam is an imaging modality designed to obtain wide field photographs of the fundus, but has been used in glaucoma management to image the optic disc and the anterior chamber angle.^[2] RetCam gonioscopy is performed through the peripheral part of cornea, and provides a novel method to investigate anterior chamber angles, with/without general anesthesia in a child.

This pilot study was performed to compare the visualization and photography of angle structures in older congenital glaucoma patients using the indirect gonioscopy and

RetCam 120, to determine the agreement between the two methods and visibility of angle structures on masked comparison of two methods.

Materials and Methods

Consecutive congenital glaucoma patients aged ≥ 5 years were recruited from the Congenital Glaucoma clinic at a tertiary ophthalmic hospital, over 6 months.

A detailed medical and surgical history was obtained from each patient and parent, followed by a thorough systemic and ocular examination, including optic disc evaluation and applanation tonometry. Patients having leukomatous corneal opacity and those having any other ocular pathology were excluded from the study. Uncooperative patients and those who did not give consent were also excluded. Patients with nebular central opacities (Haab's striae) were included in the study.

Both procedures were explained to the child and guardian, and written informed consent was obtained. The study was approved by the Hospital Institutional Review Board and adhered to tenets of the Declaration of Helsinki.

Gonioscopy was performed in all cases by a single examiner using a Goldmann 2 mirror indirect gonioscopy lens at a magnification of $\times 16$ on Haag-Streit slit lamp fitted with camera using EyeCap software, and four photographs each depicting nasal, temporal, superior and inferior angles were taken to document 360° of the angle.

RetCam imaging was performed the same day by an observer masked to gonioscopy findings. The procedure was carried out in supine position, with RetCam images taken, in each eye. After ensuring the cooperation of the patient, topical anesthetic drops were applied and speculum was placed. After application of a coupling gel, a 130 degree wide field lens was held close to the limbus, opposite the angle to be imaged. The probe was slowly

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tilted anteriorly, to obtain a clear view of the angle. The foot pedal was used to adjust the focus and brightness to get a sharp and clear image. Images were saved for subsequent analysis.

Images from goniophotography and RetCam imaging were graded by two masked glaucoma specialists, separately, as per guidelines below:

1. Clarity of trabecular meshwork:
 - Grade 1: All structures perceived
 - Grade 2: All structures not perceived
2. Quality of details seen:
 - Grade 1: All angle landmarks clear and well focused in all quadrants
 - Grade 2: Some angle structures clear, others blurred in some quadrant
 - Grade 3: Angle landmarks could not be ascertained in any quadrant

Statistical analysis was done to determine agreement by kappa statistics using SPSS software 11.5 version.

Results

Forty congenital glaucoma patients were screened, 4 had a completely opaque cornea, 7 had pseudophakia and 4 were uncooperative for the tests and therefore excluded from the study. Twenty five patients completed the study, 6 females and 19 males with age range of 5-32 years. (less than 9 yrs: 8, 9 to 16 years: 12, more than 16 years: 5)

There were 20 patients having primary congenital glaucoma, 3 with Axenfeld Rieger's syndrome and 2 having Aniridia. In 15 patients, both eyes were included and in 10 patients 1 eye was eligible.

The horizontal, white to white corneal diameter recorded was 12 to 14.5 mm (average 13 mm), and the C:D ratios were 0.4-0.9 (average 0.62). Of the 40 eyes studied, 24 had documented Haab striae, and 8 had some macular opacification of the central cornea. Surgery had been performed in 24 eyes, 5-18 (average 12) years prior to this study. All eyes studied had an IOP of <15 mm Hg at the time of the examination. Since we were only interested in the quality of angle photographs, we included children post surgery also.

Primary congenital glaucoma eyes showed multiple filiform iris processes in 95% of patients, with an anterior insertion of the iris into the mid-trabecular meshwork or at Schwalbe's line, in 80% [Figs. 1 and 2]. All eyes with Axenfeld Rieger's syndrome had a prominent Schwalbe's line and iridocorneal adhesions at the mid-peripheral cornea [Fig. 3]. The trabecular meshwork could not be visualized in the two Aniridia patients having glaucoma, due to extensive anterior synechiae of the iris stump [Fig. 4 and Table 1].

Clarity of the angle structures was Grade 1 in 37/40, 92.5% of eyes, by RetCam, compared to 30/40, 75%, on indirect gonioscopy $P = 0.73$. All structures of the angle were well focused, Grade 1 quality images in 31/40, 77.5% eyes by RetCam versus 19/40, 47.5% eyes by indirect gonioscopy, $P = 0.137$ [Table 2].

There was excellent interobserver agreement regarding the grading of the angle structures visualized by RetCam and indirect gonioscopy individually, as recorded by both observers, $P \leq 0.001$ [Table 3]. There was good agreement

between the two methods, on the extent of angle imaged by both methods, $P = 0.0011, 0.0005$. However there were significant differences between the two methods with regard to image clarity and details of angle structures [Table 4].

Discussion

Gonioscopy in congenital glaucoma is essential for determining the cause of raised IOP in that eye, as well as prognosticating the long-term course of the disease. Direct gonioscopy is

Table 1: Angle anomalies seen in different types of congenital glaucoma

Diagnosis	No.	Description of angle	No/percentage of eyes affected (%)
Primary congenital glaucoma	20	Wrap around iris	5/25
		Iris insertion at Schwalbe's line	4/20
		Mid TM insertion	12/60
		Posterior TM insertion	9/45
		Filiform iris processes	19/95
Axenfeld-Rieger's syndrome	3	Iridocorneal adhesions	3/100
		Prominent Schwalbe's line	3/100
Aniridia with glaucoma	2	Rudimentary iris stump	2/100
		Ciliary processes visible	2/100
		Closed angle	2/100

Table 2: Retcam and slit lamp gonioscopy grading

Parameters	Slit lamp		RetCam	
	Observer 1 (%)	Observer 2 (%)	Observer 1 (%)	Observer 2 (%)
Clarity of images				
Grade 1	30/40 (75)	27/40 (67.5)	37/40 (92.5)	37/40 (92.5)
Grade 2	10/40 (25)	13/40 (32.5)	3/40 (7.5)	3/40 (7.5)
Quality of details seen				
Grade 1	19/40 (47.5)	13/40 (32.5)	31/40 (77.5)	31/40 (77.5)
Grade 2	15/40 (37.5)	21/40 (52.5)	7/40 (17.5)	8/40 (20)
Grade 3	6/40 (15)	6/40 (15)	2/40 (5)	1/40 (2.5)

Table 3: Inter-observer agreement grading of angle structures

Parameters	Agreement (%)	Kappa	Probability
RetCam clarity	95.00	0.6396	0.0000
Indirect gonioscopy clarity	87.50	0.6970	0.0000
RetCam quality of images	92.50	0.7935	0.0000
Indirect gonioscopy quality of images	67.50	0.4810	0.0000

recommended for gonioscopy in congenital glaucoma, but this cannot be done in many such patients due to the presence of Haab

Table 4: Agreement between two methods

Parameters	Agreement (%)	Kappa	Probability
Indirect gonioscopy/ RetCam clarity			
Observer 1	72.50	0.0435	0.3645
Observer 2	65.00	0.0036	0.4872
Indirect gonioscopy/ RetCam extent of angle			
Observer 1	72.50	0.4647	0.0011
Observer 2	77.50	0.5161	0.0005
Indirect gonioscopy/ RetCam quality of images			
Observer 1	47.50	0.0604	0.2829
Observer 2	45.00	0.1398	0.0632

striae, central corneal edema or central corneal opacification. It requires a binocular, hand-held biomicroscope, focal illuminator and great expertise. Additionally, direct gonioscopy allows only 2-3 clock hours of the angle to be well focused at a time, with alignment of the illuminator and microscope essential to avoid reflections. The indirect gonioscope allows examination of about 180 degrees of the angle, with brighter illumination, relatively easy focus and flexible magnification by the slit lamp. To permit a masked comparison, of RetCam vs. gonioscopy, we needed best possible recorded images, and therefore compared indirect gonioscopy images to RetCam images in older congenital glaucoma patients as a pilot study.

Of the 40 eyes studied, 24 had problems in central corneal transparency, 8 opacification and 24 Haab striae. All 25 patients had undergone a MMC augmented trabeculotomy with trabeculectomy surgery and had a well-controlled IOP. Clarity in visualization of the angle structures was significantly better by RetCam images, 92.5% versus 75%, on indirect gonioscopy images on Eye cap. The RetCam provided a panoramic view, which could be easily interpreted by the observers.



Figure 1: The superior anterior chamber angle in indirect gonioscopy. Angle structures appear hazy in high magnification

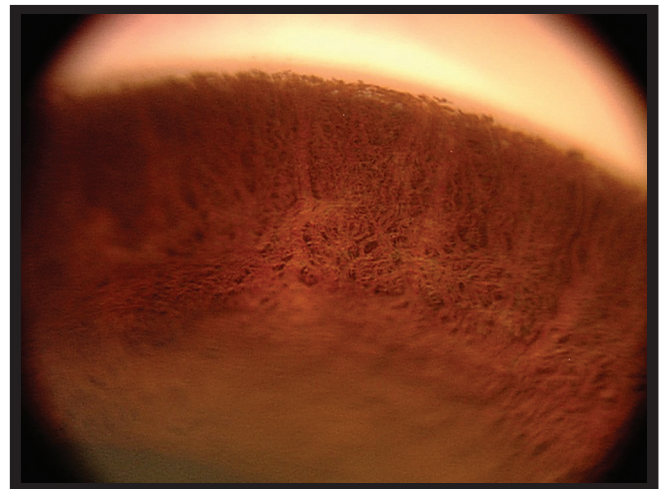


Figure 2: The same (as in Fig. 1) superior anterior chamber angle on RetCam showing forest of iris processes, wrap around iris and mid-trabecular meshwork insertion

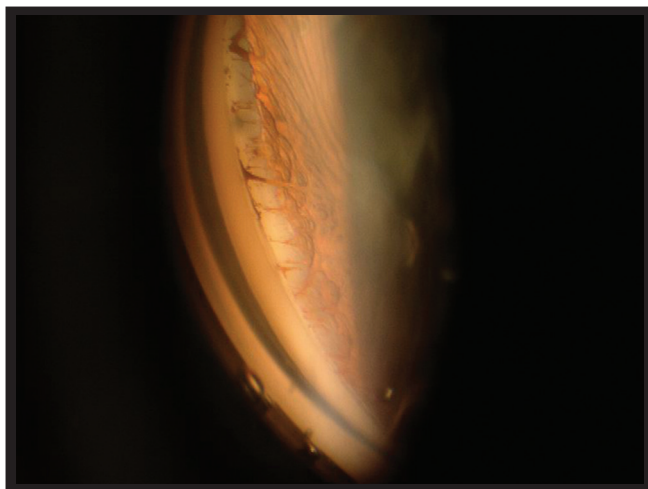


Figure 3: Gonioscopy image shows anterior chamber angle in a patient of Axenfeld–Rieger’s syndrome with prominent Schwalbe’s line and iridocorneal adhesions

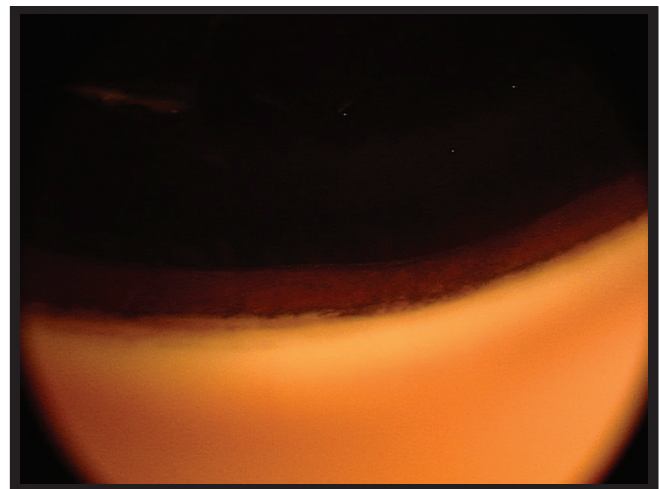


Figure 4: RetCam image shows rudimentary iris stump in Aniridia anterior chamber angle with no angle structures visible

In our study, structures of the anterior chamber angle were significantly better focused, in 77.5% eyes by RetCam images, versus 47.5% eyes by indirect gonioscopy images. There were magnified, well-focused images with a greater depth of focus obtained by RetCam, which were not always possible on indirect gonioscopy images, as magnification beyond $\times 16$ and reflections often made focusing difficult and resultant images blurred. We also found good agreement between RetCam images and gonioscopy images in identification of angle structures in patients of congenital glaucoma. In patients with primary congenital glaucoma or trabeculogenesis, we saw 20% had iris insertion at Schwalbe's line, 60% had mid-trabecular meshwork insertion, 45% had posterior trabecular meshwork insertion and 95% had filiform processes. Histopathological studies have shown the abnormalities in congenital glaucoma such as an anterior insertion of the iris and ciliary body on the trabecular meshwork with underdevelopment of the angle recess, longitudinal fibers of the ciliary muscle inserting directly into the corneoscleral meshwork and thickened and taut trabecular beams in the meshwork. Hoskins classification of developmental glaucomas is based on the anatomical contribution of different tissues – a) abnormality of the trabecular meshwork alone – isolated trabeculodysgenesis, b) abnormality of both trabecular meshwork and iris – irido trabeculodysgenesis, or with associated anomalies of the cornea – corneo trabeculodysgenesis.^[3]

Slit-lamp gonioscopy using Eye cam was more taxing to the young patient, as well as more time consuming, as they often resisted the placement of the gonioscopes and their larger and flatter corneas allowed air bubbles to frequently enter, requiring repeated placement of the lens, prolonging the procedure. In comparison when patient was cooperative, RetCam gonioscopy required the patient to look in a specific direction. Even in the presence of Bell's phenomenon, it is still possible to view the superior angles and part of the nasal and temporal angles.

There are two earlier studies, using RetCam/Eye cam in determining angle closure in adult patients, and evaluating the role of peripheral iridotomy in widening angles. They found the RetCam to be equally informative.^[4,5]

The limitations of our study were that a small number of eyes were analyzed in this pilot study and the use of indirect gonioscopy images in comparison to RetCam images. RetCam is highly expensive machine and requires highly trained observer with cooperative patients.

We were looking for a detailed evaluation of the insertion of iris, presence, type and extent of iris processes, abnormal vessels, Schwalbe's line, iridocorneal adhesions and pigmentation. Indirect and direct gonioscopy are excellent methods to visualize the angle details. However we found that the images procured by slit-lamp gonioscopy were not as clear and magnified as obtained with RetCam 120.

RetCam provides a direct view of the angle with excellent optical quality through the peripheral cornea. The direct and indirect gonioscopes use light passing through the central cornea which is often affected by Haab striae. This may explain the discrepancies between the two methods in our study, where congenital glaucoma commonly causes corneal edema associated with striae in the central cornea. The difference in findings between RetCam and gonioscopy could also be due to other factors such as patient positioning, variability of illumination and the optical principle employed.

It fulfills the long felt need for a single machine to record the angle and additionally the fundus in children with congenital glaucoma. This pilot study gives an alternative, complementary to gonioscopy for documentation of anterior chamber angles in high magnification even in central opacities.

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