ISSN: 2090-9047
Online ISSN: 2536-9466
Review Article/JORS
Khalil A, 2017, 1(2):7-27

JOURNAL OF OPHTHALMOLOGY AND RELATED SCIENCES

DEVELOPMENTAL GLAUCOMA

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***Submitted**: 2-12-2017; **Revised:** 18-12-2017; **Accepted:** 30-12-2017*

ABSTRACT

Primary congenital glaucoma is an early onset developmental glaucoma (EODG) with unique features in its pathophysiology, clinical presentation and management. Intraocular pressure (IOP) measuring is tricky in both technique and interpretation of its values. Optic disc cupping plays a more central part in follow-up of EODG than in follow-up of adult glaucoma. Management of EODG is essentially surgical. Angle surgeries; trabeculotomy and goniotomy, have the highest success rates in literature. Both of these time-honored techniques have new developments to enhance their outcomes. Other conventional techniques that are successful in adults like trabeculectomy, non-penetrating surgery and valve surgery show poorer results in infants.

The “developmental glaucomas” is a term used to describe those glaucomas caused by maldevelopment of the eye's aqueous outflow system. Depending on the severity of this maldevelopment the resultant elevated intraocular pressure (IOP) may occur at birth or anytime thereafter. Most cases present at birth or within the first year of life. The classification used here is based on the Guidelines of the Japanese Glaucoma Society[1], which names early onset developmental glaucoma (EODG) to include primary congenital glaucoma and postnatal onset (infantile) glaucoma, in which morphological anomalies are limited to the anterior chamber angle.

Pathophysiology of developmental glaucoma

In normal development[2], the corneoscleral coat grows faster than the uveal tract during the last trimester, leading to a posterior migration of the ciliary body attachment from Schwalbe's line (5th month) to the scleral spur (9th month), and then to a location behind the scleral spur (postnatally). In developmental glaucoma, the insertion of the anterior ciliary body and iris overlaps the trabecular meshwork, similar to the late fetal position. The trabecular sheets are perforated. The ciliary body band seen on gonioscopy provides an indicator of the development of the iridocorneal angle[3].

Light microscopy of the angle shows abnormal deposits of ground substances and basement membrane like substance in between trabecular beams **(Figure 1)**. In an ultrastructural study, Tawara and Inomata[4] observed a thick subcanalicular tissue with a structure similar to that seen in the endothelial meshwork beneath the inner wall of Schlemm's canal in all EODG eyes. They detected abnormal deposits of ground substances that resembled basement membrane. The intertrabecular spaces are seen filled with extracellular matrix composed of basal lamina like material and fine fibrillar material **(Figures 2, 3)**. The high success rate of trabeculotomy which cuts directly into the trabecular sheets supports these histopathological findings. The so-called Barkan membrane[5] obstructing the angle has not been proved clinically or histopathologically by subsequent studies[2,6].

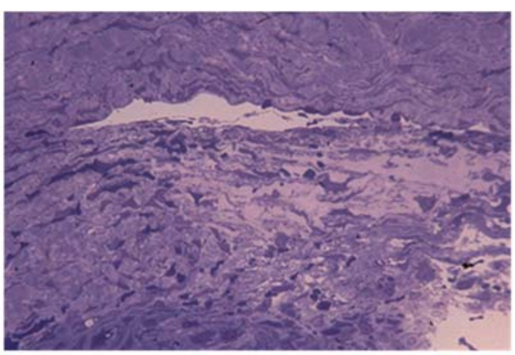


Figure 1. A light micrograph from 4-month-old boy with EODG, showing abnormal deposits of ground substances and basement membrane like substance in between trabecular beams (SC: Schlemm's canal, TM: trabecular meshwork, AC: anterior chamber). (Courtesy of Akihiko Tawara M.D., D.Sc. University of Occupational & Environmental Health, Kitakyushu, Japan)

Figure 2. Low magnified electron microscopic picture showing extracellular matrix in the subcanalicular tissue of EODG. (Courtesy of Akihiko Tawara M.D., D.Sc. University of Occupational & Environmental Health, Kitakyushu, Japan)

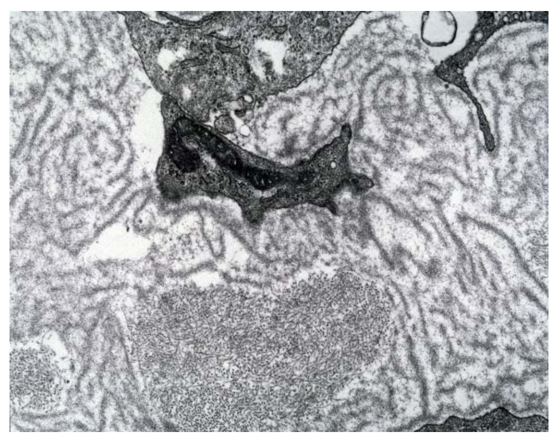


Figure 3. High magnifies electron microscopic picture showing basal lamina like material and fine fibrillar material in the subcanalicular tissue (Courtesy of Akihiko Tawara M.D., D.Sc. University of Occupational & Environmental Health, Kitakyushu, Japan)

Incidence and Genetics

The disease is bilateral in 75% of the cases, males more commonly affected (65%).

There is sporadic occurrence in 90% of the cases, the remainder; autosomal recessive with variable penetrance. Its incidence is 1:10.000 births in the West and 1:2500 births in the Middle East. More than 80% present before 1 year. There are a number of genes that have been linked with some developmental forms of glaucoma, most notably PITX2 and FOXC1 that are associated with anterior segment dysgenesis; Rieger syndrome, irido-gonio-dysgenesis, and iris hypoplasia[7]. There is significant phenotypic variability in patients with PITX2 mutations, both within and between families, with EODG as a common occurrence[8]. FOXC1 encodes a fork-head transcription factor and is typically found in patients with anterior segment disease[9]. Severity (phenotypes) can be predicted depending on the genotype[10]

Presentation and differential diagnosis:

Epiphora, photophobia and blepharospasm **(Figure 4)**

These are often the first symptoms to bring the parents with their child. These symptoms warrants careful examination with glaucoma in mind. I have seen infants with a moderate degree of glaucoma who were referred because of persistent epiphora after nasolacrimal probing. Nasolacrimal duct stenosis or obstruction is not uncommon, but is usually associated with signs of conjunctivitis and is more often unilateral. Some corneal dystrophies (Meesman's and Reis-Buckler) can produce photophobia and epiphora because of associated epithelial disturbances.

Figure 4. Classically described EODG with photophobia and epiphora.

Grayish discoloration (cloudiness of the cornea)

This is usually the presenting symptom of more advanced cases **(Figure 5).** This should be differentiated from dystrophy (Congenital hereditary endothelial dystrophy), and sclerocornea, in which scleral tissue is seen extending into the cornea. Fine vessels usually penetrate into the cornea **(Figure 6)**. A within normal corneal diameter and absence of epiphora and photophobia should hint to the diagnosis. Corneal enlargement without cloudiness **(Figure 7)** is a less often presenting symptom. This should be differentiated from megalocornea, which is congenital non-progressive corneal enlargement without evidence of previous or concurrent ocular hypertension. The angle is within normal and cornea crystal clear.

Figure 5. Bilateral corneal haze & enlargement.

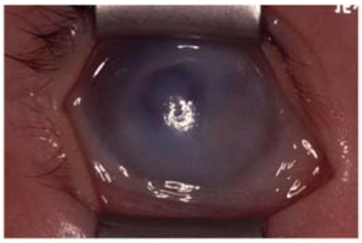


Figure 6. Sclerocornea (Courtesy of Akihiko Tawara M.D., D.Sc. University of Occupational & Environmental Health, Kitakyushu, Japan)

Figure 7. Parents are often peaceful with larger corneas of their infants seeing it as a sign of beauty, and often overlooking the mild corneal cloudiness seen in the right eye of this infant.

Examination

Examination is ideally done under general anesthesia (EUA), to make a full examination at no rush or haste. This is at least necessary for a first full examination for decision making and follow up examinations as thought necessary. Regular follow up examinations can then adequately be made with sedation (chloral hydrate), which will easily enable measuring the IOP and having a glimpse on the disc.

The normal infant eye

The normal IOP in the newborn well falls in the 1 digit mmHg readings[11]. The newborn disc has almost no cup. The cornea is clear, its horizontal diameter in a full-term newborn is 10 to 10.5 mm and increases to the adult diameter of approximately 11.5 to 12 mm by 2 years of age. In the newborn infant, the ciliary body is seen as a distinct band immediately anterior to the iris insertion. The presence of this band usually distinguishes the normal infant eye from one with gonio-dysgenesis. An invisible or very narrow ciliary body band represents an underdevelopment of the angle[3]. Insertion of the infant iris into the angle wall is flat compared with the adult angle recess configuration. Recession of the angle, which turns the iris posteriorly before inserting into the ciliary body, does not occur until the first 6 to 12 months of life.

Corneal diameter measurement and evaluation of corneal edema

A diameter greater than 12 mm in an infant is highly suggestive of EODG[12]. Measurements of the cornea are made in the horizontal meridian with calipers **(Figure 8).** Microscopic early corneal edema **(Figure 9)** or more pronounced cloudiness and breaks in Descemet's membrane should be looked for. Corneal cloudiness varies from some faint haze to marked corneal discoloration hindering proper visualization of anterior segment structures. It is usually reversible after IOP control **(Figure 10).** Rupture lines of Descemet’s membrane (Haab’s striae) with associated localized areas of edema can be seen in moderate to severe untreated cases **(Figure 11).** When the edema related to these lines subsides, glassy lines can be seen long after glaucoma control.

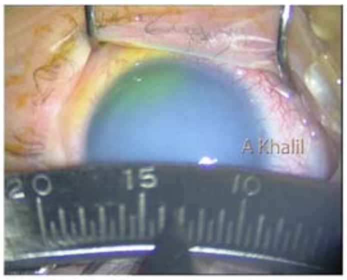


Figure 8. Measuring of the white to white corneal diameter

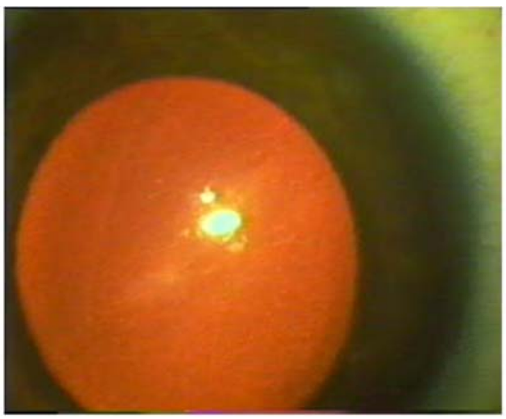


Figure 9. Microscopic corneal edema (epithelial microbullae seen by retroilluminatoin against red reflex) can be an early sign of EODG with postnatal onset.

Figure 10. Corneal cloudiness clearing in the left eye 5 days after surgery as compared to the right still cloudy non-operated eye (watering is caused by child crying rather than epiphora).

Figure 11. Rupture lines of Descemet's membrane (Haab's striae) with associated localized areas of edema.

Measuring the IOP

The normal IOP in newborns and young infants is much lower than in adults. A time honored value of 21 as an upper limit cannot be taken peacefully in infants, a most valuable observation made first by Sampaolesi et al in the late sixties[13], followed by several observers[14-16] **(Figure 12).**

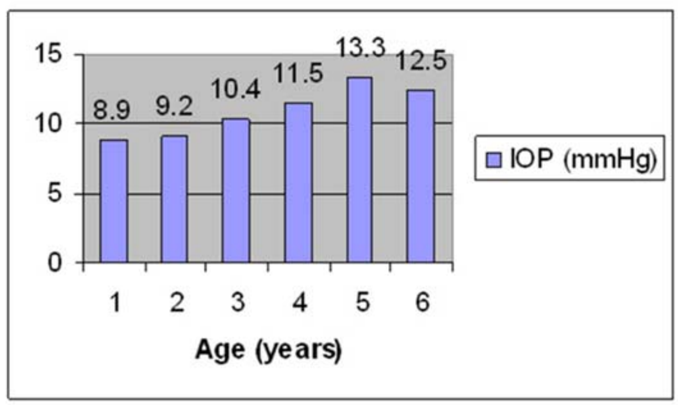


Figure 12. IOP measurements in infants and children up to 6 years (Data from Sampaolesi et al[13]).

IOP measurement in infants **(Figure 13 a-c)** is associated with several potential flaws,

i. Anesthetic considerations
EUA is usually required. Most general anesthetics lower IOP by variable amounts and at variable times after administration.

ii. Device Considerations
The normal IOP, measured in the supine position with the Perkins, was found to be lower in infants and young children than in adults. On the other hand, with the Pneumotonometer, the supine pediatric IOP was found to be higher than that measured by the Perkins and was not significantly different from the adult sitting IOP[15]. The Schiotz measurements were significantly higher than those obtained with the Perkins and the Tono-pen tonometers[17].

iii. Anatomical considerations
The large globe size relative to the small palpebral fissure renders IOP measuring very tricky, gentle lid retraction by examiner's finger-tips without exerting any direct pressure on the eye can easily lead to posterior bowing of the lateral canthus increasing the reading. Facial differences make the ideal lid opening different between different infants. It was recently demonstrated that central corneal thickness was significantly thinner in eyes with EODG than in normal controls[18]. Goldmann applanation tonometry was developed assuming a corneal thickness around 500 μm[19]. Thicker corneas tend to give higher IOP measurements[20] and thinner corneas tend to give lower measurements[21].

To obtain the most relevant readings, the following guidelines might be helpful,

i. Standardization of examination parameters; tonometer used, anesthetic agent, examination setting, etc.

ii. Obtaining at least 5 readings for each eye, with full separation from the patient's eye after each reading, the lowest repeatable reading is used.

iii. A reading of 18 in a newborn should not be passed as normal. When will trained by both mother and surgeon, it is often possible to measure IOP in children at office.

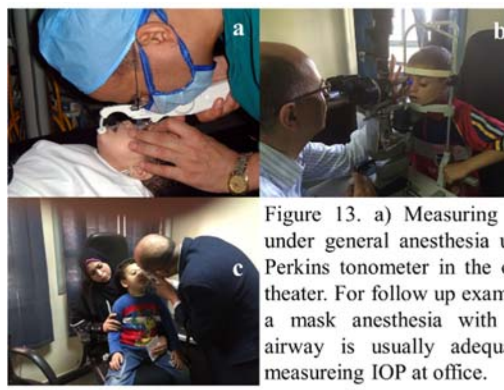


Figure 13. a) Measuring the IOP under general anesthesia using the Perkins tonometer in the operation theater. For follow up examinations, a mask anesthesia with an oral airway is usually adequate. b,c) measureing IOP at office.

Optic nerve head evaluation **(Figure 14a, b)**
The newborn disc has almost no cup. Cupping larger than 0.3, especially if asymmetric between two eyes, is strong evidence that the disc is under pressure and may be glaucomatous[22]. Changes in the optic disc occur readily with changes in IOP in infants. This is caused by increased tissue elasticity in the infant eye translating IOP increase into mechanical distortion in the disc supporting elements, and posterior

bowing of the lamina cribrosa. This cupping is easily reversible typically within 4-6 weeks after normalization of IOP. The younger the child, the faster the reversibility[23,24]. If left untreated, neuronal loss eventually ensues, with irreversible damage. An increasing cup size indicates inadequate intraocular pressure control. Therefore, it is vital to make careful drawings or take photographs for subsequent comparison. Together with regression of photophobia, reversibility of optic disc cupping is one of the best criteria for surgical success in most cases.

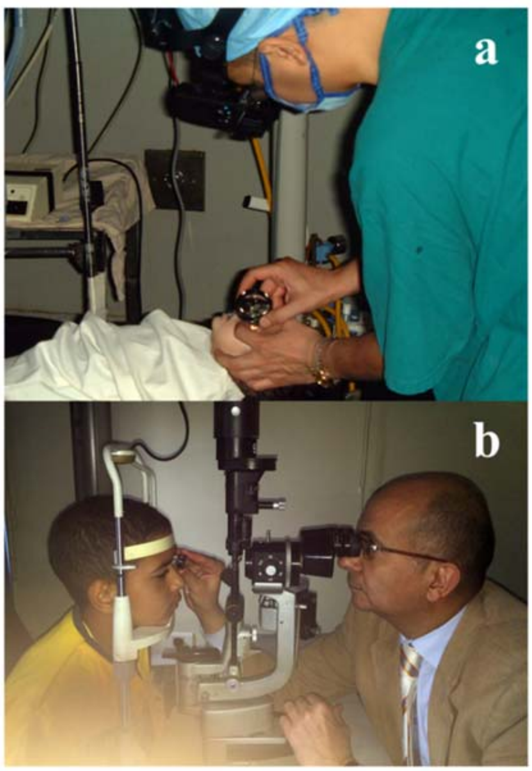


Figure 14. Optic disc evaluation under anesthesia (a), and at office (b).

Gonioscopy

Gonioscopy **(Figure 15)** is performed by use of a gonioprism, a Koeppe or a barkan operating lens (a truncated koeppe lens) and binocular microscope for the evaluation of gonio-dysgenesis.

Axial length (AL) measurements

Measuring the axial length was proposed as a diagnostic and follow-up procedure[25]. Similar to corneal diameter, AL increases in a disproportionate manner to age in EODG. It is still thought by others that corneal diameter is a more sensitive tool in this context[26]

Figure 15. Gonioscopy, flat insertion of the iris into Schwalbe's line.

Cycloplegic refraction

After control of the glaucoma, cycloplegic refraction is done to rule out any associated refractive error and/or anisometropia with subsequent amblyopia and strabismus. Though usually to the myopic side, hyperopic shift in buphthalmic eyes can occur. Prompt amblyopia management is as important as glaucoma control to achieve the best visual outcome in these eyes **(Figure 16).**

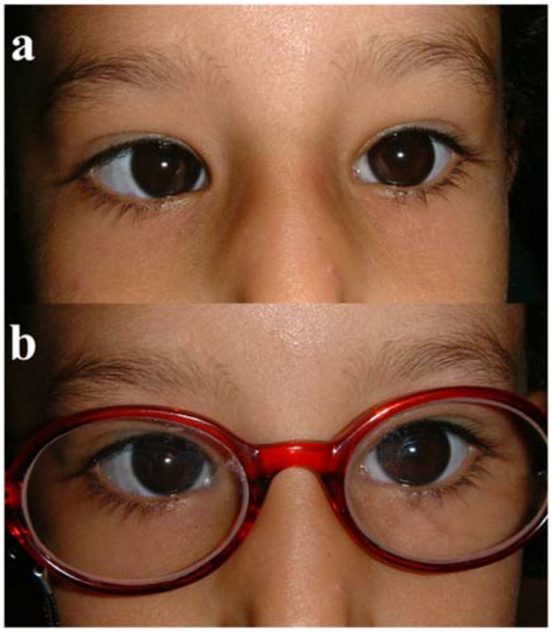


Figure 16. a) This case presented 3 years after bilateral successful trabeculotomy for EODG, with anisometropia (one eye myopic, the other low hyperopic) and strabismus. b) Glasses correction and amblyopia management could achieve a bilateral 0.8 visual acuity.

Management

Management of EODG is a challenging responsibility, which starts with early detection, and offering optimum surgical intervention followed by a life-long follow up. Prognosis is related to the time of its initial presentation, initial surgical intervention, degree of optic nerve damage and later visual rehabilitation[27]. Surgery is the first line of therapy for EODG since this type of glaucoma results from abnormal anatomical development of the angle, and its surgical correction is recommended. Accumulated experience shows the effectiveness of surgery, while the long-term effectiveness and complications of different anti-glaucoma drugs have not been well studied.

Surgical Management

i. Trabeculotomy and goniotomy remain the first line surgical procedures for EODG[28-33]. A surgeon anticipating to manage these cases should be familiar with either technique. Goniotomy and trabeculotomy act to enhance aqueous outflow by cutting through the trabecular meshwork, which is the main site for resistance for aqueous outflow in these eyes **(Figure 17a)**. The rates of successful IOP control with these procedures are relatively high and the incidence and severity of intra-operative and postoperative complications are small in comparison with all other procedures. They directly attack the faulty site with minimal surgical trauma, as compared with trabeculectomy **(Figure 17b)**.

ii. Trabeculectomy

The decision to perform this procedure must be made carefully because in infants and children, filtering bleb formation may be difficult despite intraoperative use of antimetabolites. The use of mitomycin C (MMC) leads to a lower IOP level, but also leads to a greater incidence of resultant hypotony maculopathy. Late bleb-related ocular infection does occur in children after trabeculectomy with MMC and is characterized by abrupt onset, bleb infiltration, and rapid progression, leading to significant late visual loss[34-36]. The addition of trabeculectomy, deep sclerectomy to trabeculotomy looks to be a handsome synergism and has been advocated by some authors[37-40]. Kubota et al[41], did not find an additive effect for sinusotomy when added to trabeculotomy. It is hardly conceivable why trabeculectomy, known to induce proliferation in younger eyes, should be added to a potentially successful trabeculotomy. In a series of 30 eyes with previously failed primary surgery, 18 eyes were treated by trabeculotomy and 12 eyes were treated by trabeculotomy-trabeculectomy. There was no statistically significant difference in the final outcome between the 2 groups (Khalil, unpublished data).

iii. Deep sclerectomy

Deep sclerectomy lowered the pressure below 16 mmHg in 56% of EODG eyes[42]. Although deep sclerectomy may reduce the IOP in patients with refractory EODG, there was a specific risk profile associated with it and all eyes were ultimately classified as failures[43]. Deep sclerectomy leaves the trabecular beams, which are the main site for resistance to outflow in EODG, untouched. **(Figure 17c).**

iv. Aqueous shunt implantation

Aqueous shunt implantation offers a greater chance of successful glaucomacontrol in the first 2 years of life, compared with trabeculectomy with MMC. However, the enhanced success is associated with a higher likelihood of postoperative complications requiring surgical revision, most commonly tube repositioning[44]. They can be used as a last resort in refractory cases with multiple previous surgeries and compromised angle structures.

Goniotomy versus trabeculotomy

Trabeculotomy and goniotomy seem to be in some ways equivalent, and both are particularly successful in previously unoperated cases of EODG. Goniotomy, however, does not have a good success rate when done below 1 month or over 2 years of age[28]. It is usually associated with a relatively high rate of recurrence[45], and multiple goniotomies are needed to achieve a success rate similar to that of trabeculotomy[46]. Trabeculotomy is probably a more demanding technique with which it may be more difficult to achieve a technically perfect procedure than it is with goniotomy. In a relatively large case series of 317 eyes that underwent different surgical modalities, trabeculotomy gave the best results[47] and it was shown to have an excellent long-term results[33]. Even with goniotomy advocates[29], trabeculotomy is generally preferred in children over the age of three years, in situations where corneal clouding prevents adequate visualization of the trabecular meshwork, and in patients with aniridia[48]. A 360 degrees trabeculotomy was suggested[49], but added to its possible technical difficulties, it is commonly followed by extreme hypotony[50]. Microcatheter-assisted trabeculotomy[51] is another relatively new technique for trabeculotomy. *Goniotomy* involves the creation of a superficial incision into uveal TM, allowing the iris root to move posteriorly and presumably relieving the mechanical obstruction to aqueous outflow. This procedure is performed under direct visualization through a gonioscopic contact lens and the operating microscope. There have been some reports on endoscopic goniotomy overcoming the inherent drawback of difficult visibility caused by corneal cloudiness[52, 53], with no much later reports about further results.

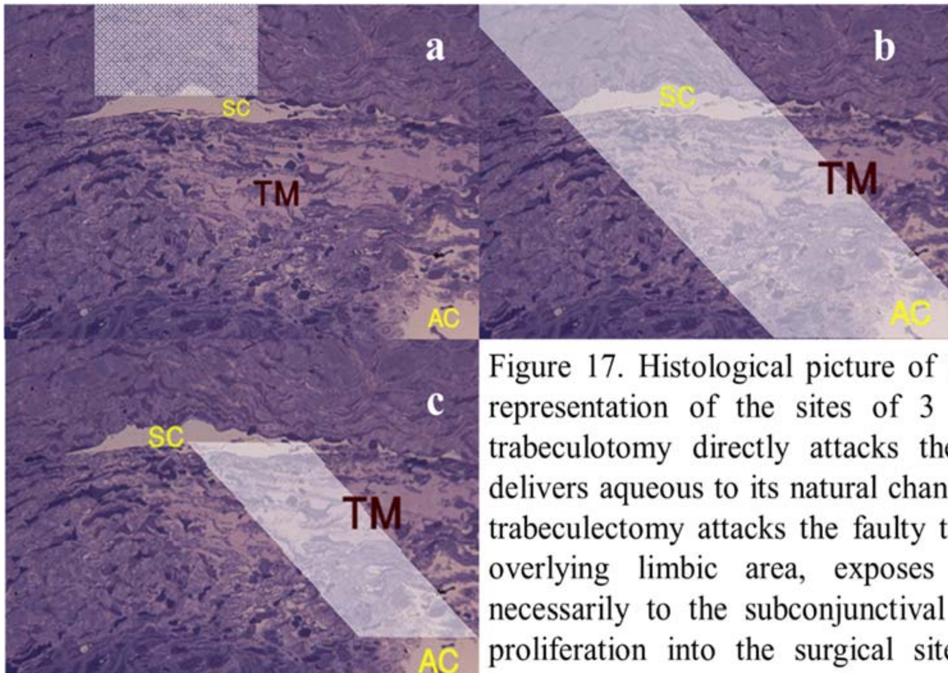


Figure 17. Histological picture of fig 1 with diagrammatic representation of the sites of 3 surgical procedures; a) trabeculotomy directly attacks the faulty trabeculum, & delivers aqueous to its natural channel; Schlemm's canal. b) trabeculectomy attacks the faulty trabeculum together with overlying limbic area, exposes the surgical site unnecessarily to the subconjunctival space & invites active proliferation into the surgical site. c) Deep sclerectomy attacks deep scleral tissue leaving the faulty trabeculum intact. SC: Schlemm's canal, TM: trabecular meshwork, AC: anterior chamber.

Trabeculotomy is my first choice for surgery in all cases of EODG, regardless any previous surgery, as long as there is a sound non-scarred 120 degrees of limbic circumference. Bad prognostic signs include; eyes with corneal diameter 14.5 or more, advanced congenital cases, and multiple previous surgeries. Several techniques have been described for performing trabeculotomy. I prefer the use of 3 sets of variable curvature handle-less probes, which can adapt to varying corneal diameters. Coming with different curvatures helps in gentle and accurate probing of the canal. An unsuitable curvature used might not pass the whole length; and gives a false block impression. They are very light weight with no handle to directly transmit force to them, rendering it difficult to spoil the 2 core steps in the procedure; by either forceful faulty insertion, or traumatic forceful rotation into the AC if it is not correctly inserted.

Technique **(Figures 18 a-d):**
A 6-0 corneal traction, is taken to expose the superior limbic area. A limbal based conjunctival incision is made 6-8 mm in length and 4-5 mm from the limbus, and pulled away by two 8-0 retraction sutures. A superficial scleral flap is carried out well into the clear cornea **(Figure 19).** A radial incision is **(Figure 20)** gradually deepened over the canal of Schlemm, which demonstrates considerable variation in its location among newborn-infant eyes with different globe sizes. It is usually located more posteriorly in larger globes, and can often be found stretched,

well underneath the (white) of sclera. Opening of the canal is recognized by one or more of the following; gentle egress of aqueous **(Figure 21)**; a gush or an efflux of aqueous **(Figure 22)** denotes opening the AC rather than the canal, direct (dry) visualization of the canal (**Figure 23)**; most commonly in stretched out eyes, associated afterwards by exudation of aqueous. In congested eyes, egress of blood from the canal site can be the main sign of its opening.

Probe insertion **(Figure 24-26)**:
The lip of the radial incision is gently held by the non-dominant hand, while the trabeculotomy probe is held by the dominant hand and gently introduced into the cut end of the canal. The internal probe can usually be visualized in its tight path in the canal. The external probe is always there to assess the conformity of the trabeculotome to the limbic circumference. A correctly placed trabeculotome only moves along its axis, not perpendicular to it. It cannot be rotated posteriorly! If it does, then it is probably in the suprachoroidal space **(Figure 27)**. When in the canal, it is a blunt pin in a conforming tube! Cutting the soft trabecular meshwork does not involve any force. On rotating the trabeculotome into the AC, the tip makes the first cut, and appearance in the AC, then follows the rest of the internal probe **(Figures 28, 29)**. This tactile lag between

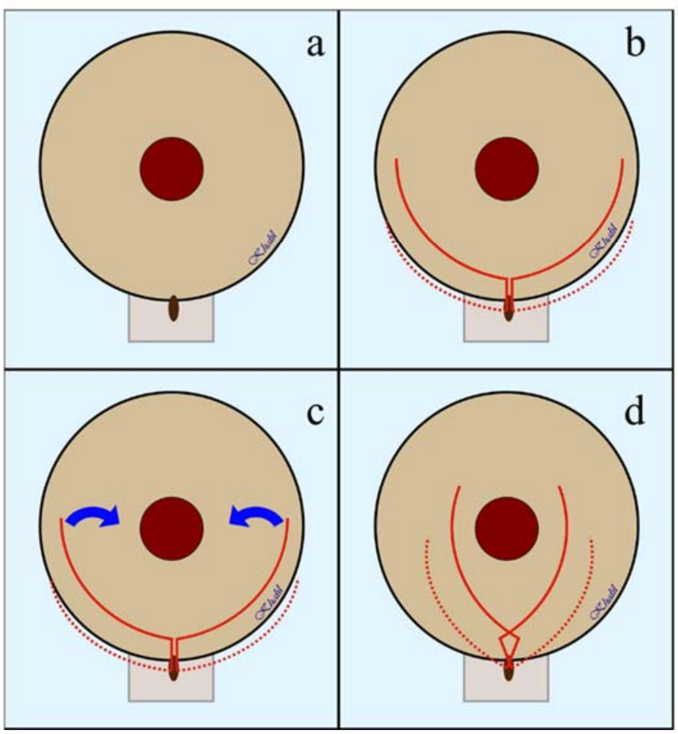


Figure 18. a,b) Diagrammatic representation of the layout of surgery; surgery is usually carried out at the 12 o'clock position, unless this site is jeopardized by previous surgery, in which case a lateral (or even inferior) approach can be used. Trabeculotomy probes are introduced into both sides of the cut ends of Schlemm's canal. Both probes should be introduced before rotating the first probe to reduce the difficulty of inserting the second probe in a collapsed canal. c,d) Probes are then rotated, one after another into the anterior chamber. In doing so, the trabecular meshwork is severed along the length of the probes connecting the Schlemm's canal directly to the anterior chamber.

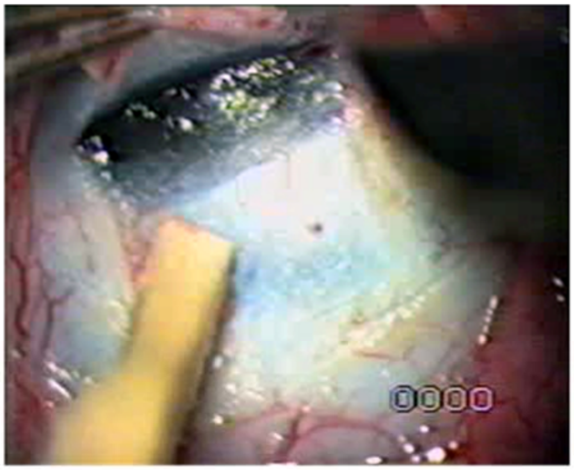


Figure 19. Intra-scleral dissection is carried out well into the clear cornea. Position of the canal is so variable in eyes with EODG.

Figure 20. A radial incision is started, gradually deepening over the presumed location of the canal of Schlemm till it is reached.

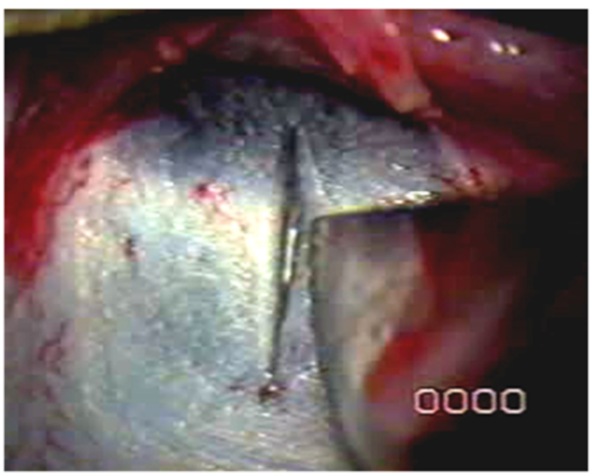


Figure 21. Gentle egress of aqueous is a good indication of opening the canal.

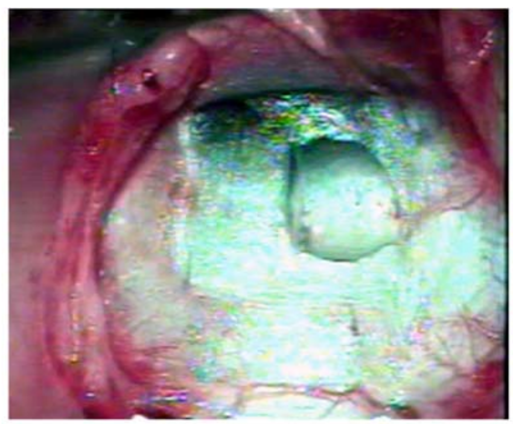


Figure 22. A (gush) of aqueous during dissection signifies opening the AC rather than the canal. Finding the canal becomes more demanding, but possible.

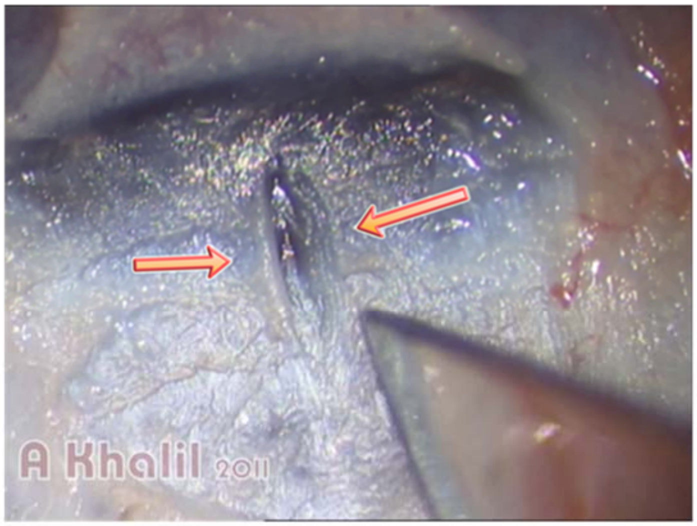
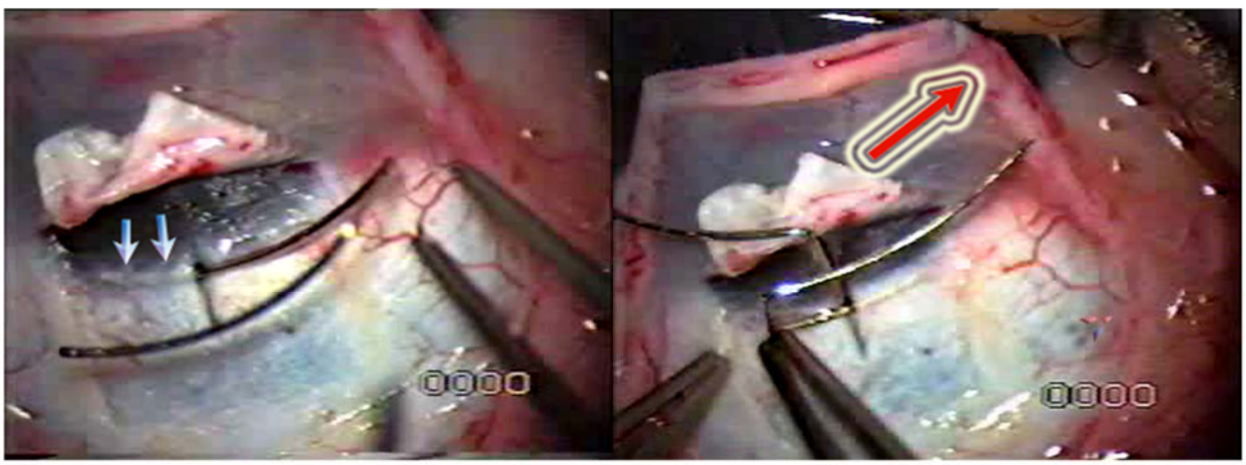


Figure 23. Direct (dry) visualization of the canal.



Figures 24,25. Both probes are gently (knocked) to their course, the internal probe can usually be visualized in its tight path in the canal (arrows). The external probe is always there to assess the conformity of the trabeculotome to the limbic circumference, and that it has not gone astray. There should be minimal resistance; significant resistance means the probe is in the wrong place.

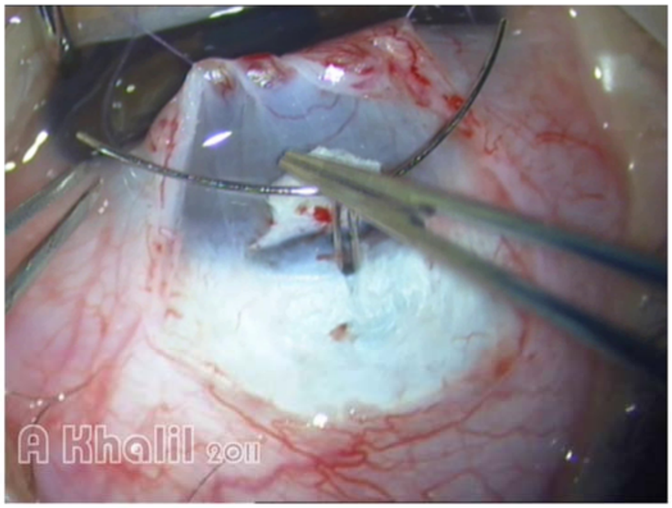


Figure 26. The 2 probes nicely in place; the outer probes are parallel to the limbus. They are not freely mobile either anteriorly into the AC, or posteriorly into the supra-ciliary space.

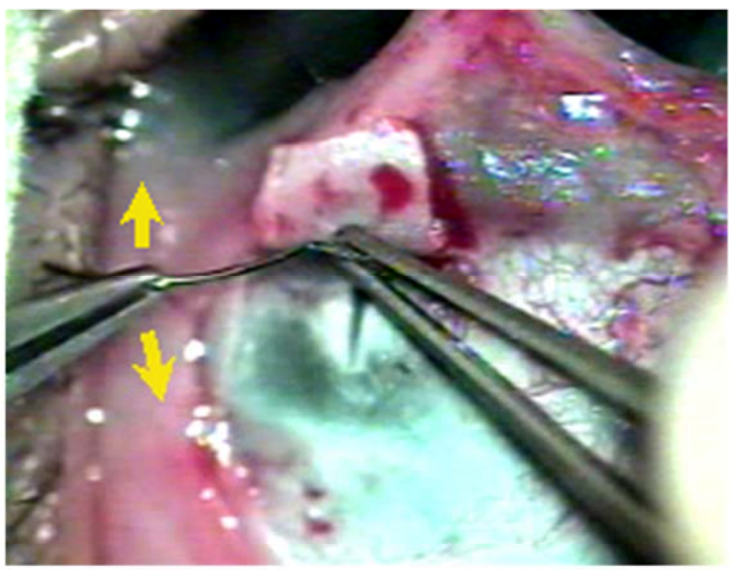
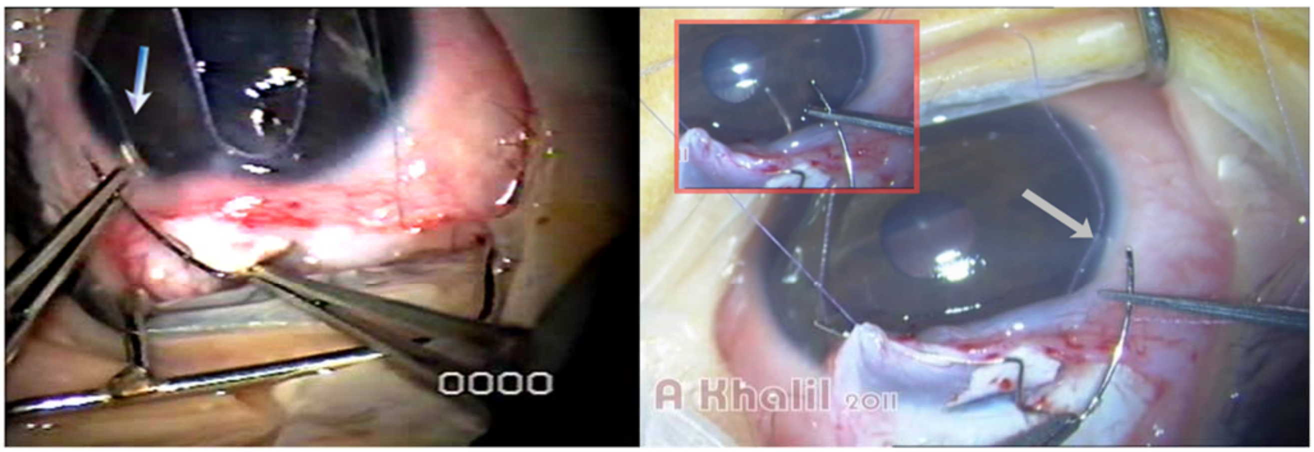


Figure 27. The outer probe betrays a faulty insertion into the supra-ciliary space; the trabeculotome is freely mobile posteriorly. This has to be removed completely, and re-inserted carefully.



Figures 28,29. On rotating the correctly placed trabeculotome into the AC, the tip makes the first cut, and appearance in the AC (arrows), then follows the rest of the internal probe (inset). This tactile lag between appearance of only the tip first, with no limbal or corneal distortion, and then the rest of the probe (Khalil's sign), is an important sign of success. If rotation meets no resistance at all, with simultaneous appearance of the whole length of the internal probe in the AC means that it was lodged in the anterior chamber angle rather than in the canal.

appearance of only the tip first, with no limbal or corneal distortion, and then the rest of the probe (Khalil's sign), is an important sign of success. The need for force, with corneal or limbal distortion simply means that the trabeculotome is not properly placed **(Figure 30)**.

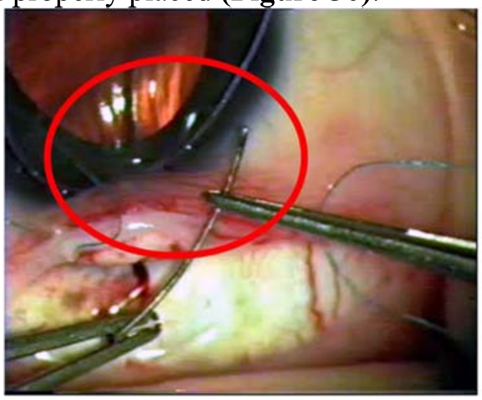


Figure 30. Corneal wrinkles on rotating the trabeculotome imply its faulty insertion in scleral lamellae.

On the other hand, if rotation meets no resistance at all, with simultaneous appearance of the whole length of the internal probe in the AC means that it was lodged in the anterior chamber angle rather than in the canal. After rotating of the full length of the probe into the AC, the probe is gently withdrawn, paying care not to touch the iris-lens. This is especially important with the second probe when the anterior chamber gets shallower. Hyphema on rotating the trabeculotome is not uncommon, but usually self-limited, and absorbed by the second post-operative day. Injection of air into the AC helps to control a more active bleeding is very rarely needed. Scleral flap is closed tightly by interrupted 10/0 monofilament. The conjunctiva is then closed by running 8/0 virgin silk or vicryl **(Figure 31)**.

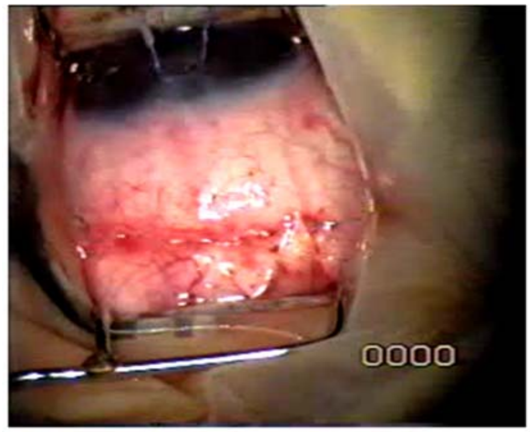


Figure 31. Conjunctiva wound site at the conclusion of the procedure.

Medical Treatment

Poor efficacy, a greater potential for adverse systemic side effects (especially in infants and very young children), difficulties with administration and compliance, and the lack of proven long-term safety profiles in children have contributed to the minimal role of glaucoma medications in the pediatric population. Medical treatment is used as an auxiliary means to temporarily control IOP till surgery is performed or in the rare instance when repeated surgeries fail to control IOP.

i. Beta adrenergic antagonists

Include timolol 0.25% and betaxalol. These agents have been shown to decrease the IOP in pediatric patients. Systemic side effects can be severe and have included asthma, bradycardia, and apnea[54].

ii. Carbonic anhydrase inhibitors

Include both oral and topical formulations. Acetazolamide administer-ed orally is very effective at lowering IOP but can result in a decreased appetite, diminished energy, and a metabolic acidosis. Topical dorzolamide or brinzolamide are viable alternatives to oral carbonic anhydrase inhibitors, with significantly fewer side effects but less effective IOP reduction[55]. Most of the patients show disappointingly little IOP effect from *latanoprost*, but some children, particularly older children and those with juvenile-onset open-angle glaucoma, do have a significant ocular hypotensive effect with it[56-58]. Systemic and ocular side effects in children on latanoprost are infrequent and mild.

iii. Cholinergic agonists, such as *pilocarpine*, effectively constrict the pupil but have a limited effect on IOP presumably due to the altered anatomy of the anterior chamber angle[59].

iv. Finally, ***brimonidine*** should be used with caution in young children because of the potential for CNS depression[60].

Outcome parameters

As delineated partially in the examination section, successful management of EODG cannot mean simply achieving a pressure below 21mmHg without medication! It is to the contrary, a postoperative pressure of 21 in a few months infant is a definite failure. Successful control means all of the following Regression of corneal edema Regression of optic disc cupping (below 1 year), or at least its stabilization (above 1 year) Stabilization of corneal diameter and/or axial length Last and least; an IOP less than 16 mmHg.

Follow-up

The inability to easily measure visual acuity and extent of visual loss in neonates makes these parameters less helpful in following patients than measurement of corneal diameter and intraocular pressure. However, even these data should not be relied upon exclusively to determine the quality or quantity of success in the follow up of EODG. When embarking on the management of these eyes, the ophthalmologist should bear in mind and inform the family of the patient that there should be a lifelong follow up. The criteria mentioned under outcome parameters should be assessed frequently in the first year post-operatively. A suggested regimen would be 1 week, 1, 3, 6 months postoperatively then a 6 monthly examination till the age of 4 years. Afterwards, a yearly examination should be sufficient. This is a flexible schedule which can be modified according to individual cases. Refractive problems should be anticipated and dealt with promptly. It is near meaningless to save an eye from sight threatening glaucoma, and lose it.

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