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ADVANCES IN OPHTHALMOLOGY AND OPTOMETRY

Current Management of Pediatric Glaucoma

Sushmita Kaushik, MS*, Deepika Dhingra, MS, DNB, FICO, Surinder S. Pandav, MS

Advanced Eye Centre, Postgraduate Institute of Medical Education and Research, Sector 12, Chandigarh 160012, India

Keywords

• Childhood glaucoma • Congenital glaucoma • Examination under anesthesia • Goniotomy • Trabeculotomy

Key points

- The management of childhood glaucoma has greatly improved in terms of a better classification system, development of newer diagnostic modalities, better anesthesia techniques, and medical and surgical treatment.
- The interest in ocular biometry (axial length, pachymetry, etc.) and imaging techniques (ultrasound biomicroscopy, handheld optical coherence tomography, etc.) has increased over time, resulting in better diagnosis and treatment.
- The importance of examination under sedation for the frequent evaluation of children with glaucoma is now recognized, as this helps avoid the adverse effects of repeated general anesthesia.
- The role of oral propranolol has been described in children with Sturge-Weber syndrome with choroidal hemangioma to prevent intractable choroidal effusions and exudative retinal detachments.
- Glaucoma drainage devices and cyclophotocoagulation (endoscopic, transillumination) procedures are increasingly used for children with multiple failed surgeries.

*Corresponding author. Advanced Eye Center, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
E-mail address: sushmita_kaushik@yahoo.com
Twitter: @KaushikSushmita (S.K.); @Insandeepika19 (D.D.)

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INTRODUCTION

Childhood glaucoma is a treatable cause of blindness provided it is recognized, diagnosed, and treated in time [1]. WHO has estimated that it is responsible for blind years (second only to cataract) [2]. Although congenital glaucoma is a rare disease in terms of prevalence among ophthalmic diseases (0.01% to 0.04%) [3], it accounts for 4.2% to 5.0% of blindness in the pediatric population [4,5] and 2% to 15% of individuals in blind institutions. Prompt diagnosis and surgical treatment can prevent blindness in most of these infants. Preservation of any vision during a child's formative years is important, to avoid a lifetime of blindness.

The fundamental pathophysiology of all childhood glaucoma, regardless of the cause, is impaired outflow through the trabecular meshwork, causing an increased intraocular pressure, which leads to optic neuropathy, ocular enlargement, and corneal changes including corneal edema, haab's striae, or opacity; this could be due to a developmental abnormality (nonacquired) or due to acquired causes such as trauma, surgery, inflammation, etc. It is clear that childhood glaucoma per se is an umbrella term that comprises a vast variety of diseases including those that occur at birth, those that are developmental in nature but manifest later, and those that are due to acquired causes [6]. It is imperative to know exactly what condition one is dealing with, because the treatment and prognosis depends largely on what the underlying disease is.

There has been a growing interest in childhood glaucoma in recent years, probably partly due to greater survival of small infants with the developments in neonatal care and the greater dissemination of modern ophthalmic care to hitherto underdeveloped regions. Unlike adult glaucoma, the management of childhood glaucoma is difficult owing to the varied nature and prognosis of the disease and the need of ensuring normal visual development of the immature growing eye. Pediatric glaucoma is difficult to classify because children often present with a variety of ocular or systemic findings frequently attributable to underlying genetic defects.

The management of childhood glaucoma has improved in many ways, which include better classification methods and understanding of the disease, newer diagnostic modalities, improvements in anesthesia procedures, and surgical treatment options that have improved significantly since goniotomy was first described by Barkan [7] and trabeculotomy was first described by Burian and Smith [8,9]. Because of rapid developments in molecular biology techniques, it is now much easier to understand the pathophysiology of the disease by unraveling the underlying genetic abnormality.

In this chapter the authors look at recent advancements seen in the world of childhood glaucoma. This would include the development of a novel unified classification system, newer surgical procedures, and the exciting potential of genetic research in this condition.

Standardized nomenclature

For all phenotypically and genotypically heterogeneous diseases, a universally accepted nomenclature and easy-to-use classification helps to develop

standards of care and promotes widespread collaboration and development of new advancements. The Childhood Glaucoma Research Network (CGRN) has developed a standardized nomenclature and classification system that was ratified by a consensus statement at the IXth World Glaucoma Association at Vancouver, 2013 [10] and was later adopted by the American Academy of Ophthalmology [11]. This classification will be used during the course of this review of pediatric glaucoma.

DIAGNOSIS

The age of diagnosis depends on the national criteria for pediatric patients.

- United States: younger than 18 years
- United Kingdom, Europe, Asia: younger than 16 years

Table 1 depicts the diagnostic criteria adopted by the Childhood Glaucoma Research Network (CGRN) [10].

CLASSIFICATION

Childhood glaucoma had no uniform classification system. The terms congenital glaucoma and developmental glaucoma were used interchangeably. Some

Table 1
Definitions of glaucoma and glaucoma suspect as per Childhood Glaucoma Research Network classification

Glaucoma	Glaucoma suspect
Intraocular pressure (IOP)-related damage to the eye At least 2 of the following criteria are present: 1. IOP > 21 mm Hg; investigator discretion is required for children who are examined under anesthesia due to variable effects of anesthesia on IOP measurement 2. Optic disc changes: • Optic disc cupping or progressive increase in cup-disc ratio • Cup-disc asymmetry of $\geq$ 0.2 or focal rim thinning 3. Corneal findings: Haab's striae or diameter $\geq$ 11 mm in newborn, > 12 mm in child < 1 y, or > 13 mm any age 4. Progressive myopia, myopic shift, or an increase in axial length out of keeping with normal growth 5. Reproducible visual field defect consistent with glaucomatous optic neuropathy	No IOP-related damage to the eye At least 1 of the following criteria are present: 1. IOP > 21 mm Hg on 2 separate occasions 2. Suspicious optic disc appearance for glaucoma, that is, increased cup-disc ratio for size of optic disc 3. Suspicious visual field for glaucoma 4. Increased corneal diameter or axial length in setting of normal IOP

Adapted from Beck A, Chang TC, Freedman S. Section 1: Definition, classification, differential diagnosis. In: Weinreb RN, Grajewski A, Papadopoulos M, Grigg J, Freedman S, editors. World Glaucoma Association Consensus Series-9: Childhood Glaucoma. Amsterdam, The Netherlands: Kugler Publications; 2013. pp. 3–10.

of the earlier classifications proposed by various investigators include the following:

- Hoskins and colleagues [12] (classified the disease as per anatomic defects such as isolated trabeculodysgenesis or associated dysgenesis of the iris and/or cornea)
- Schaffer and colleagues [13] (isolated congenital glaucoma, associated with congenital anomalies or acquired glaucoma)
- Walton and colleagues [14] (addition of other disorders in the classification by Schaffer and colleagues)

The CGRN classification [10] is the currently used standard classification system, and it has classified childhood glaucoma into 4 broad segments, as depicted in Table 2.

Fig. 1 depicts the CGRN Flowchart that is, a guide to reaching a diagnosis for any child presenting with glaucoma.

Representative pictures of the most commonly seen entities of childhood glaucoma are shown in the following pictures (Figs. 2–5).

DIAGNOSTIC TECHNIQUES

Intraocular pressure measurement and current understanding of its role in pediatric glaucoma

Applanation tonometry is the gold standard for IOP measurement. The Goldmann or Perkins applanation tonometer (GAT/PAT) (in the outpatient department/examination under anesthesia, respectively) is commonly used. However, there are many children who are cooperative for slit-lamp examination but do not cooperate for GAT. For these cases, many newer tonometers have been developed such as tonopen, noncontact tonometry or iCare tonometer (Tiolat Oy, Helsinki, Finland) [15–17]. The Tonopen (Reichert Inc, New York, USA) requires the use of topical anesthetic, and the readings are reliable only with lower IOP levels less than 20 mm Hg [15]. With higher IOP, the instrument usually tends to overestimate the IOP with discrepancy as high as 12 mm Hg [16].

The rebound tonometer by iCare is very light in touch, does not require topical anesthesia, and can be performed more easily in younger children as well (Fig. 6). It has been found to have readings within 3 mm Hg as measured with applanation tonometer, and the readings are usually higher than applanation readings [17].

Despite the different modalities available, the measurement of IOP in children is nevertheless challenging. Examination under anesthesia is often necessary for pediatric patients. Different agents used for sedation or general anesthesia have reported having varied effects on IOP [18]. Also, the modalities used for airway management, hemodynamic factors, tonometry technique, and body positioning can all affect IOP measurements. IOP measurement in children is also potentially influenced by other factors such as the type of tonometer used to record IOP, the cooperation of children, eye movement, and the

Table 2
Childhood Glaucoma Research Network classification

	Secondary childhood glaucoma			
Primary childhood glaucoma (isolated angle anomalies) No other ocular or systemic associations	Associated with congenital nonacquired ocular anomalies	Associated with congenital nonacquired systemic anomalies	Acquired glaucoma	Secondary acquired glaucoma post-cataract surgery
1. Primary congenital glaucoma • Neonatal-onset glaucoma (0–1 mo of age) • Infantile glaucoma (1–24 mo of age) • Late-onset or late recognized (after 2 y) 2. Juvenile glaucoma	Glaucoma associated with ocular anomalies in addition to angle dysgenesis 1. Axenfeld-Rieger anomaly (syndrome if systemic associations) 2. Peters anomaly 3. Ectropion uveae 4. Congenital iris Hypoplasia 5. Aniridia 6. Persistent fetal vasculature 7. Microphthalmos 8. Microcornea 9. Ectopia lentis	Glaucoma and associated nonacquired systemic features 1. Sturge-Weber syndrome 2. Homocystinuria 3. Mucopolysaccharidoses 4. Weill Marchesani syndrome 5. Axenfeld-Rieger syndrome 6. Phacomatoses 7. Neurofibromatosi 8. Congenital rubella syndrome	This group includes secondary glaucoma due to various acquired reasons other than cataract surgery: 1. Uveitis 2. Trauma 3. Steroid-induced 4. Tumors 5. Retinopathy of prematurity 6. Prior ocular surgery other than cataract surgery	Glaucoma after cataract surgery has been given a separate place considering the high frequency of glaucoma following cataract surgery in children

Adapted from Beck A, Chang TC, Freedman S. Section 1: Definition, classification, differential diagnosis. In: Weinreb RN, Grajewski A, Papadopoulos M, Grigg J, Freedman S, editors. World Glaucoma Association Consensus Series-9: Childhood Glaucoma. Amsterdam, The Netherlands: Kugler Publications; 2013. pp. 3–10.

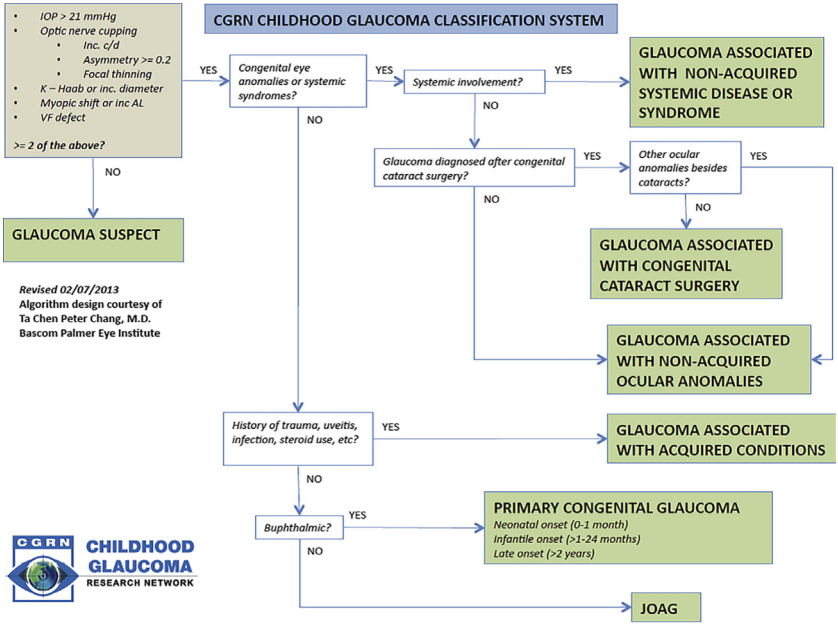

Fig. 1. CGRN childhood glaucoma classification system. (*From* Robert N Weinreb. Childhood glaucoma: the 9th consensus report of the World Glaucoma Association [2013]; Used with permission of Kugler Publications.)

status of the cornea such as edema or opacities [19]. In fact, it is now recognized that the IOP is among the least accurate parameters measured when assessing a child for glaucoma. It is increasingly accepted that the diagnosis of childhood glaucoma should never be made based on IOP alone.

Ocular biometry

The interest in ocular biometry, including axial length and pachymetry, measurements has gradually increased over the years due to the greater understanding regarding the value of these parameters in the clinical evaluation as well as decision-making of children with glaucoma. In a normal eye, corneal power, axial length, anterior chamber depth, and lens power are the major

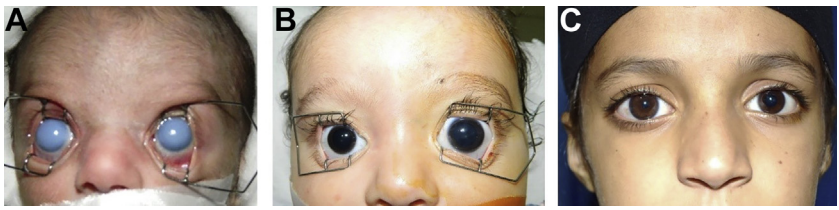

Fig. 2. Primary congenital glaucoma. Neonatal onset (*A*). Infantile onset (*B*). Late onset (*C*).

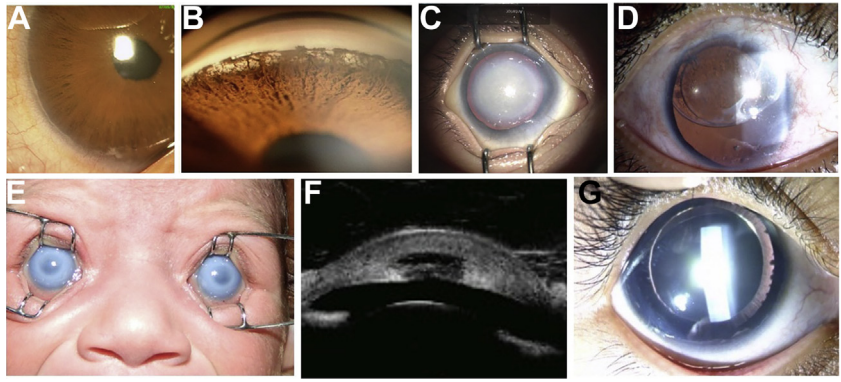

Fig. 3. Glaucoma secondary to nonacquired ocular conditions. (*A, B*) Posterior embryotoxon and typical iris processed seen on gonioscopy in Axenfeld-Reiger syndrome. (*C*) Neonatal onset congenital ectropion uveae. (*D*) Aniridia with aniridia keratopathy and IOL seen in the subluxated capsular bag. (*E*) Bilateral Peters anomaly. Not the clear area in the region of the Descemet membrane and posterior stromal defect. (*F*) UBM of the same baby seen in (*E*) showing the posterior stromal defect clearly. (*G*) Microspherophakia.

refractive components of the eye, with axial length typically being the single most crucial factor. Other than corneal stretching and axial elongation of the eyeball, the effect of congenital glaucoma on these variables has not been widely studied [20,21]. Eyes with glaucoma usually have larger axial lengths at presentation, which decrease after IOP control and then follow a normal curve as expected for age [22,23]. Biometry could be of immense importance both for the diagnosis of congenital glaucoma in children with borderline IOPs and to detect glaucoma in the fellow eye of patients with presumed unilateral disease. It also may have an essential role in the follow-up of patients with congenital glaucoma who had undergone surgery.

Patients with congenital glaucoma are known to have thick corneas in the presence of corneal edema, which becomes thinner after IOP control and resolution of corneal edema [24,25]. IOP measurement could also be affected by corneal biomechanical factors in additional to the anatomic thickness. The corneal hysteresis and resistance factor are reportedly decreased in congenital glaucoma, which may affect IOP readings [24]. On the contrary, many eyes with microcornea have thicker corneas [25], which may result in fallaciously higher IOP readings.

It is clear that it is important to rely on numerous other factors such as optic disc evaluation, corneal diameter, and axial length measurements for optimum assessment of the glaucoma status in children.

Refractive error

Refractive changes in children depend on many factors such as genetics, reading habits, and the environment [26,27]. In children with glaucoma, IOP is another factor contributing to the refractive change due to the eye's

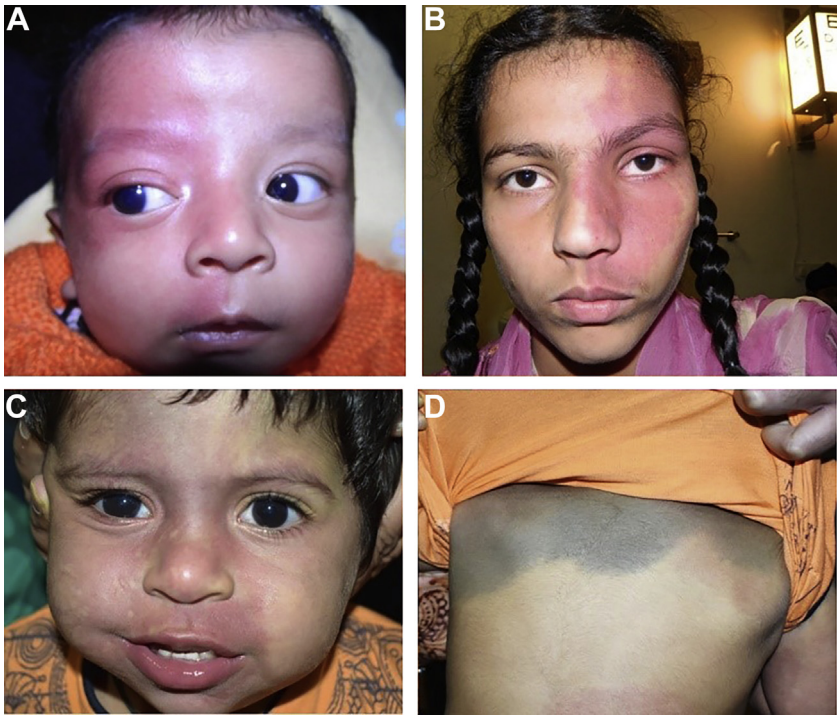

Fig. 4. Glaucoma secondary to nonacquired systemic diseases. Sturge-Weber syndrome. Glaucoma may occur in infancy (*A*), causing buphthalmos, or in late childhood (*B*), causing raised IOP and disc cupping. (*C, D*) Klippel Trenaunay syndrome showing the port-wine stain crossing the midline (*C*) and large pigmented area on the back (*D*) and limbs.

enlargement. Every millimeter increase in axial length contributes to −2.5 D of myopia, and every dioptre increase in corneal curvature contributes to 1.0 D of myopia [28]. The anterior chamber depth and lens thickness also contributes to the refractive changes [29].

Ultimately, the refractive error in an individual is determined by an interplay of corneal/lens power and axial length. In cases of pediatric glaucoma, it has been found that myopia/hypermetropia are not always proportionate to the axial length [30]. There are many factors responsible for emmetropization in childhood glaucoma as well. Corneal enlargement and corneal flattening, a decrease in the axial diameter of the lens due to ocular enlargement and backward movement of the lens, are some of the factors that may be responsible to counteract myopia due to an increase in the axial length.

Ultrasound biomicroscopy

Ultrasound biomicroscopy (UBM) has a great role to play in cases of significant corneal opacification where anterior chamber details are not clear. It can be performed in the outpatient department in cooperative children but needs to be

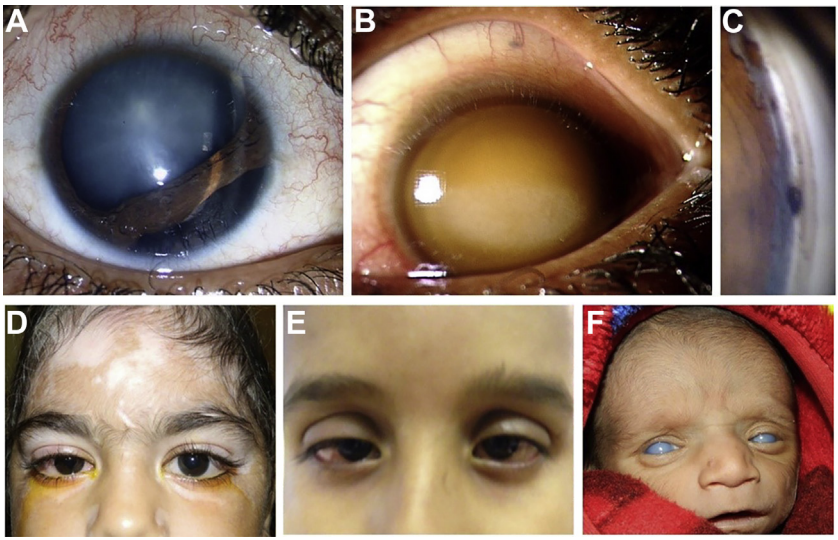

Fig. 5. Acquired childhood glaucomas. Posttraumatic glaucoma showing iridodialysis and traumatic cataract (*A*), corneal blood staining (*B*), and angle recession on gonioscopy (*C*). (*D*) Glaucoma secondary to uveitis in a child with Vogt-Koyanagi-Harada disease. (*E*) Steroid-induced glaucoma in a child with vernal keratoconjunctivitis. (*F*) Neonatal glaucoma in congenital rubella syndrome. Note the typical ''monkey facies.''

performed under anesthesia/sedation in uncooperative children. For neonates and cases with smaller eyes and orbits, a clear scan soft sleeve technique is useful (Fig. 7).

Another potential utility of the UBM is to look for the Schlemm canal and the outflow channels to predict the success rate with angle surgery [31].

Handheld optical coherence tomography

The handheld spectral-domain optical coherence tomography (Envisu 2300, Bioptigen Inc., Research Triangle Park, NC, USA) has been found feasible to look for the anterior segment and posterior segment structures in younger children without sedation or anesthesia [32,33]. It has great potential for accurate assessment of anterior segment structures, and the developing angle, and promises to be of great help in creating a normative database of retinal nerve fiber layer and macular thickness in children. For example, it could be of immense value in prognosticating a baby with aniridia by diagnosing the foveal hypoplasia [34].

Genetic testing

Genetic testing is very important in childhood glaucoma, although it is very complex, as there are many genes that may be responsible for a set of findings and genetic diseases with varied findings. Moreover, primary congenital glaucoma cases are sporadic in greater than 90% cases and familial in less than 10%

Fig. 6. Measurement of IOP using a handheld rebound tonometer while the baby is comfortable in his mother's lap.

cases. For genetic testing, there are single gene tests (eg, CYP1B1 for primary congenital glaucoma [PCG], FOXC1 and PITX2 for Axenfeld-Rieger and Peters anomaly; PAX6 for aniridia), multiple gene panels (eg, a set of genes for early onset glaucoma), or whole exome or genome sequencing (for cases where the disease is considered to have a genetic defect, but the disease is complex and genes responsible are not known) [35,36].

Genetic testing, although logistically difficult to offer all patients, has a great role in certain situations, as illustrated in the following section:

1. Positive family history so as to predict which family member has a risk of disease depending on the presence or absence of a mutation in a particular gene, and thus plan for the follow-ups of asymptomatic members can be made accordingly
2. Genetic counseling by predicting the risk of transfer of gene and the disease development

Fig. 7. UBM using a clear scan probe in a neonate with corneal opacity.

3. To predict the prognosis, as PCG cases with CYP1B1 mutations have been found to have a severe disease compared with those without [35].
4. To diagnose a complex case where the diagnosis is not certain so as to prognosticate the disease and for genetic counseling

ANESTHESIA

Childhood glaucoma cases require repeated anesthesia to monitor IOP, corneal diameter, anterior segment, and posterior segment examination until the children start cooperating for slit-lamp examination and IOP measurement. Repeated anesthesia exposures have been shown to affect brain development and can affect their cognition, behavior, and memory [37]. Many sedative agents including chloral hydrate, pedicloryl, midazolam, and ketamine are used for short-term procedures, with variable success rates [37,38]. A newer sedative agent dexmedetomidine, α-2 agonist, has been found to have better success rates compared with chloral hydrate for the ophthalmic examination of children. It does not cause gastrointestinal side effects as with chloral hydrate and respiratory depression as with midazolam. It can be administered intranasally or intravenously. The intranasal route has been found to have better acceptability, as it does not cause any irritation and avoids the cannulation. A recent study [39] on the evaluation of intranasal 3.5 μg/Kg dexmedetomidine reported success rates of 77.4% with dexmedetomidine alone and 100% when rescue drug midazolam 0.25 mg/kg intranasal was administered. Fig. 8 depicts the examination procedure.

TREATMENT

Medical treatment

The mainstay of treatment in nonacquired childhood glaucoma is surgical. Medical treatment remains useful as a temporizing measure for IOP control

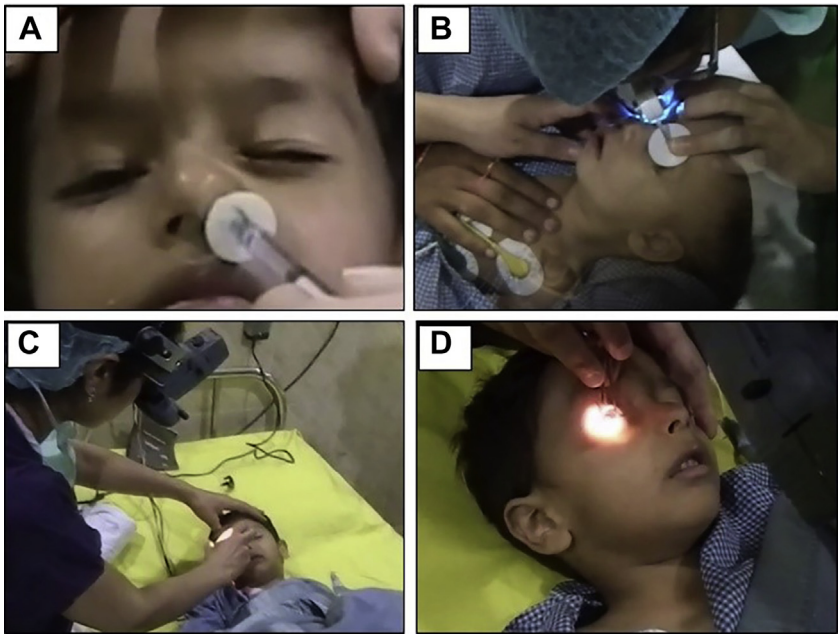

Fig. 8. The examination procedure of a child under sedation using intranasal dexmedetomidine. (*A*) Administration of the drug using mucosal atomization device with half dose in each nostril. (*B*) Intraocular pressure measurement using Perkin applanation tonometer with the child in mother's lap. (*C*) Indirect ophthalmoscopy for the examination of anterior and posterior segments. (*D*) Corneal diameter measurement.

until the surgery is performed or to supplement inadequate IOP control after surgery. There has been an improvement in the available formulations of timolol maleate for use in infants. Topical timolol 0.5% can cause apnea in smaller children due to increased systemic absorption. Timolol 0.25% gel formulation once a day, betaxolol 0.25%, and now timolol maleate 0.1% gel form has been found to have a better safety profile. Timolol 0.1% formulation is gel-based with carbomers (carbopol) and polyvinyl alcohol and has good retention with similar IOP lowering as of timolol 0.5% without systemic side effects [40,41].

Although the exact mechanism of action and duration of therapy is not known, oral propranolol has been shown to be effective in Sturge-Weber syndrome to prevent/treat choroidal effusions/exudative retinal detachments due to diffuse choroidal hemangioma. A dose of 2 mg/kg/d has been effective, by the presumed mechanisms of inhibiting angiogenesis, apoptosis of proliferating endothelial cells, or vasoconstriction etc. [42,43].

Surgery

Advances in childhood glaucoma surgeries are mainly guided by trials first conducted in adults [44]. Barkan [7] first described goniotomy with unsuccessful

outcomes in adults and then demonstrated good outcomes in children. Goniotomy became the surgical treatment of choice for children with good outcomes. However, a reasonable cornea clarity was the main prerequisite to perform the surgery to visualize the angle. Subsequently, trabeculotomy was described as an ab-externo technique to rupture the inner wall of Schlemm canal, independently by Burian [8] using the trabeculotome and by Smith using nylon filament [9]. Combined trabeculotomy with trabeculectomy was first described by Maul and colleagues [45], which soon gained popularity because of its higher success rates and faster corneal clearing, which is so important in children to ensure visual rehabilitation and prevent amblyopia.

However, it was a sobering fact that despite the best of surgical technique, many surgical procedures failed. Glaucoma in children could be very refractory to one surgical procedure. Glaucoma drainage devices were introduced as a viable treatment for refractory glaucoma or for glaucomas where the conjunctiva was deemed to be too scarred for a trabeculectomy to succeed. The Baerveldt Glaucoma implant (BGI) and Ahmed Glaucoma Valve implants were introduced in 1990 and 1993, respectively [46]. Recently the Aurolab aqueous drainage implant has been introduced for clinical use by Aurolab, Madurai, India. This implant is a low-cost, nonvalved glaucoma drainage device (GDD) designed as the BGI with a 350 mm^2 plate area, which has shown encouraging results at par with the established implants [47].

Advances in surgical techniques are basically the modifications in the original techniques described years back, some of which are described as follows:

Modifications in goniotomy

a. Coaxial endoscopic goniotomy has been described for cases with corneal edema and was first described in humans by Medow and colleagues in 1997 [48]. It uses an endoscope (EndoOptiks, Little Silver, NJ) with a special blade mounted on the endoscope itself, or one can also use an MVR blade (Visitec, Sarasota, FL) through a separate incision with anterior chamber maintained by using either a viscoelastic or an anterior chamber maintainer.
b. Other modifications in the goniotomy technique involve the use of an electrosurgical device called Trabectome or Kahook Dual Blade to ablate/excise the inner wall of Schlemm canal, which prevents the closure of the cleft [49].
c. Gonioscopy-assisted transluminal trabeculotomy (GATT) has been described by Dr Davinder Grover in cases where an initial goniotomy cleft is created and an illuminated microcatheter or a prolene suture that is cauterized at the tip are used to pass through the Schlemm canal for 360° treatment of the angle [50].

Modifications in trabeculotomy

A 360° trabeculotomy has been found to have higher success rates with the achievement of lower IOP compared with conventional trabeculotomy by treating greater extent of the angle. It can be performed using either 6–0 prolene suture (introduced by Beck and Lynch) or by an illuminated microcatheter iTrack (iScience Interventional, Menlo Park, CA), which helps with the direct visualization of the illuminating tip [51].

Glaucoma drainage devices

The use of GDDs has increased over the years and are found useful for the failed trabeculectomy cases or multiple failed glaucoma surgeries. It has fewer bleb-related complications, but pediatric eyes are more prone to complications of tube migration, retraction, tube corneal touch, and endophthalmitis compared with adults [47,52]. Small modifications have ensured greater safety in implantation of GDDs in children:

- Tube elongation using angiocatheter or especially available extension devices
- Tube shortening by cutting the longer tubes using lesser invasive methods from within the anterior chamber

Cyclophotocoagulation

This procedure is reserved for cases with poor visual potential or multiple failed surgeries. It is difficult in children, as the location of ciliary processes changes with the globe enlargement due to anatomic changes in a buphthalmic eye and, moreover, ciliary processes regenerate more in pediatric cases. Many developments have been made in the technique in the form of the use of an endoscope for the coagulation of ciliary processes under direct visualization [53]. Using transillumination to localize the site of ciliary processes has been shown to improve the chances of success with this modality.

Despite the rapid strides in the diagnosis and management of childhood glaucoma, there is much to explore in this field. Research continues for the better care of patients with childhood glaucoma in terms of disease control with visual as well as vocational rehabilitation.

CLINICS CARE POINTS

- Childhood glaucoma is a potentially blinding condition unless recognized and treated in time.
- The basis of all childhood glaucomas, whatever may be the cause, is raised IOP due to reduced aqueous outflow.
- Because the infant's eye is elastic, the consequences of this raised IOP includes secondary effects such as globe enlargement, progressive myopia, and corneal changes due to breaks in the Descemet membrane and stromal edema.
- The treatment of childhood glaucoma, especially in infancy is usually surgical.
- Angle surgery is the most commonly performed surgery and includes goniotomy, trabeculotomy performed ab-externo or ab-interno.
- Cases of cloudy corneas may need a trabeculectomy combined with trabeculotomy.
- Refractory childhood glaucomas often require glaucoma drainage devices or cyclophotocoagulation.
- The underlying cause is important to diagnose, as many developmental glaucomas may be associated with other systemic abnormalities.

- Unlike in adult glaucomas, childhood glaucoma cannot be treated by control of raised IOP alone but also requires intensive amblyopia treatment to ensure visual rehabilitation.
- Advances in genetic technology have also opened up techniques such as Next-Gen sequencing, which allows genotype characterization and opens up avenues for appropriate genetic counseling.

Disclosure

Nothing to declare.

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