ORIGINAL ARTICLE

Profile of Childhood Glaucoma Attending a Tertiary Eye Care Center in Northern India

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Abstract

Purpose: To ascertain the prevalence and clinical features of the various types of childhood glaucoma at a tertiary eye care hospital in Northern India.

Materials and methods: Retrospective chart review of all children less than 16 years of age with childhood glaucoma who presented from 1st April 2014 to 31st March 2019, who was diagnosed to have any subtype of childhood glaucoma as per Childhood Glaucoma Research Network (CGRN) classification and advised appropriate management.

Results: Out of 405 children with childhood glaucoma, 36% had primary glaucoma, whereas the rest had secondary glaucoma. Primary congenital glaucoma (PCG) was the most common form of primary glaucoma. Glaucoma associated with acquired conditions was the most common cause of secondary glaucoma. Primary glaucoma was mostly bilateral in contrast to secondary glaucoma. The most common age of presentation with primary glaucoma was <1 year of age, and in children with secondary glaucoma was 11–16 years. On presentation, 80% of eyes had intraocular pressure (IOP) of >20 mm Hg and 70% had cupping of >0.7. Eyes with PCG were primarily managed surgically.

Conclusion: In our cohort, PCG was the most common primary childhood glaucoma. Traumatic glaucoma was the most common secondary glaucoma. Since childhood glaucoma is an important cause of visual morbidity in children, its timely diagnosis and prompt management are essential to prevent irreversible visual loss.

Clinical significance: Understanding the disease pattern, their presenting features, and the proportion of different types of childhood glaucoma can help in planning appropriate eye care services, create awareness and better allocate resources to plan appropriate management strategies. Screening programs and counseling of parents should also be strengthened.

Keywords: Childhood glaucoma, Childhood Glaucoma Research Network classification, Primary congenital glaucoma, Primary glaucoma, Secondary glaucoma.

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Introduction

Glaucoma is an important cause of visual impairment in children which is reversible in the early stages and irreversible in later stages. Childhood glaucoma is estimated to cause a significant percentage of blindness (1.2% in Great Britain, 3% in North Eastern India, and 7% in Southern India).[1–3] Childhood glaucoma can be classified as primary or secondary childhood glaucoma as per CGRN Classification (2013).[4]

Various studies conducted all over the world to determine the epidemiology of childhood glaucoma has reported variable prevalence, and there were differences in the characteristics of various subtypes. In the United Kingdom, Papadopoulos et al. evaluated 99 children and found that 52 had secondary glaucoma with an annual incidence of PCG—5.41 in 1,00,000 live births in Great Britain and 3.31 in 1,00,000 live births in Ireland.[5] In Toronto, Taylor et al. reviewed 306 children and found out that the most common childhood glaucoma was congenital (38%), followed by aphakic glaucoma (20%).[6]

In Saudi Arabia, Alanazi et al. reviewed 180 patients and found that 80% had PCG.[7] In China, Fang et al. reviewed 734 patients and found that PCG was the leading subtype (47.55%), followed by primary juvenile glaucoma (17.03%) and traumatic glaucoma (11.03%).[8] In Olmsted county, Aponte et al. reviewed 30 children and found the incidence to be 2.29/1,00,000. Secondary glaucomas were the most common, while congenital and juvenile forms were rare.[9] In Dallas, United States of America, Fung et al. reviewed 376 eyes of 239 patients, of whom 19% had PCG, 4% had primary juvenile glaucoma, 45% had secondary glaucoma (posttrauma and aphakia), and 31% were glaucoma suspects.[10] From Beijing, Qiao et al. evaluated 1,055 patients with childhood glaucoma. Congenital glaucoma was the most common subtype (46%), followed by traumatic (12%) and aphakic glaucoma (9%).[11] A prospective study from a tertiary center in South India found that 56% of children had primary glaucoma—PCG was the most common childhood glaucoma.[12] Reported prevalence of PCG varies between 23 and 80%.[5–13] Some studies reported a higher prevalence of

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PCG[6,7,11–13] whereas others had a higher number of children with secondary glaucoma.[5,8–10]

Understanding the disease pattern, its presenting features, and the proportion of different types of childhood glaucoma can help in planning appropriate eye care services and training to create awareness and better allocate resources to plan appropriate management strategies. To the best of our knowledge, there is no such study that evaluates the prevalence and clinical features of the various types of childhood glaucomas childhood glaucoma in Northern India. This study aims to delineate the profile of childhood glaucoma presenting to a tertiary eye care hospital in the region.

Materials and Methods

A retrospective review of medical records of all the children 16 years of age with childhood glaucoma who presented to our tertiary eye hospital in Northern India from 1st April 2014 to 31st March 2019 was performed. Subjects who had had adequate Outpatient department or one examination under anesthesia (EUA) whereby all the relevant investigations of glaucoma would have been done, the patient was diagnosed to have any subtype of childhood glaucoma as per CGRN classification and advised appropriate management, were included for this study. The study was approved by the institutional review board, and the study protocol adhered to the tenets of the Declaration of Helsinki.

The definition of childhood glaucoma (two or more of the following findings) as per CGRN classification[4] is:

- Intraocular pressure (IOP) of >21 mm Hg on repeated testing or when IOP was <21 in the presence of corneal changes or under medications for lowering IOP.
- Optic disk cupping: progressive increase in the cup-disk ratio, cup-disk asymmetry of ≥0.2, or rim thinning.
- Corneal findings: Haab's striae or corneal diameter ≥11 mm in newborns, >12 mm in children <1 year of age, and >13 mm at any age.
- Progressive myopia or myopic shift coupled with an increase in ocular dimensions beyond normal growth.
- A reproducible visual field defect consistent with glaucomatous optic neuropathy.

Subcategories of childhood glaucoma as per CGRN classification[4] are:

- Primary childhood glaucoma.
  - Primary congenital glaucoma (PCG)—neonatal glaucoma detected at birth to 1 year)/infantile glaucoma detected >1–3 years of life.
  - Juvenile open-angle glaucoma (JOAG)—glaucoma in children from 3–16 years of age, excluding the categories as mentioned in secondary glaucoma or children with typical signs of PCG.
- Secondary childhood glaucoma.
  - Glaucoma associated with nonacquired ocular abnormalities—Peter's anomaly, Rieger anomaly, and aniridia.
  - Glaucoma associated with nonacquired systemic abnormalities—Down syndrome and Phakomatosis.
  - Glaucoma associated with acquired conditions—trauma and steroids.
  - Glaucoma postcataract surgery (pseudophakic/aphakic).
- Glaucoma suspects.

Following details were noted from medical records—the age at presentation, presenting complaints, history of consanguinity, family history, laterality, history of the acquired cause, visual acuity, IOP measurement (by Goldmann Applanation Tonometry in older and cooperative children/Perkin's tonometer in children undergoing EUA), anterior segment and posterior segment findings, management details. Visual acuity in all the children was assessed in our records as fixing and following light for nonverbal children, Teller acuity chart, or Snellen chart for verbal children. Evaluation under anesthesia was done using Sevoflurane in our hospital. For all those children in whom the disc couldn't be seen due to corneal edema, findings for glaucomatous changes in the disc were noted postoperatively or after IOP was adequately lowered. Few children also underwent visual field evaluation whenever possible.

Patients >16 years of age at presentation who were lost to follow-up and could not undergo all the relevant investigations or with incomplete/lost records were excluded from our study.

Descriptive statistics included mean and standard deviation for scale variables. Categorical variables were summarized as percentages. All the calculations and charts were generated using Microsoft Excel 2016 and Statistical package for the Social Sciences version 24.

Results

On a retrospective chart review, 584 eyes of 405 patients were found to have childhood glaucoma. The details of the clinical characteristics of children with various subtypes of glaucoma are given in Table 1. Out of 405 children, 144 children (249 eyes—36%) had primary glaucoma, whereas 261 children (335 eyes—64%) had secondary glaucoma. No children were glaucoma suspects in our study. Out of 144 children with primary glaucoma, 114 children (79%) had PCG, whereas 30 children (21%) had JOAG. Glaucoma associated with acquired conditions was the most common cause of secondary glaucoma in our study affecting 181 eyes of 160 children (61.3% of all the secondary glaucoma cases). Most common causes among the acquired conditions were trauma (25.7%), postvitreoretinal surgery (13.8%), retinoblastoma (9.9%), and steroid-induced glaucoma (6.1%). Glaucoma following cataract surgery was seen in 38 eyes of 28 children (10.7% of all secondary glaucoma cases), of which 21 eyes of 15 children had aphakia (5.7%) and 17 eyes of 13 children were pseudophakic (4.98%). Glaucoma associated with nonacquired systemic/syndromic diseases affected 17 eyes of 11 children (4.2%). Sturge–Weber syndrome and Rubella were the most common syndromic associations seen in our study. In glaucoma associated with nonacquired ocular anomalies, 99 eyes of 62 children (23.7%) were affected, and the majority were iris/corneal pathology. The most common corneal pathology was Peter's anomaly, whereas aniridia was the most common iris pathology.

Of the 405 children, 179 children (358 eyes—44%) had bilateral involvement, whereas the rest had unilateral glaucoma (226 eyes—56%). Of the 144 children with primary glaucoma, 105 children (73%) and 72 children (28%) out of 261 children with secondary glaucoma had bilateral involvement (Table 1).

The total number of males in our study was 277 (68%), while females were 128 (32%). Male:female ratio was close to 2:1 in primary glaucoma (1.82:1) as well as in nonacquired ocular anomalies (2.4%). The male:female ratio was 3.9:1 in secondary glaucoma due to acquired conditions, whereas the ratio was 0.8:1 in children having glaucoma following cataract surgery. The mean age at presentation was 3 months in PCG, 12 years in JOAG, and 11 years in uveitis/trauma/steroid induced/postvitreoretinal surgery and post-penetrating keratoplasty patients. Mean age at presentation in children with secondary glaucoma following retinoblastoma was 3.14 years, 6.50 years following retinopathy of prematurity (ROP), and 9.4 years in children with secondary glaucoma following cataract

Table 1: Clinical characteristics of children (number, laterality, and gender) with various types of childhood glaucoma

			Laterality of glaucoma			*Gender*	
Diagnosis (as per CGRN classification)	*Number of subjects*	*Number of eyes*	*Right eye affected*	*Left eye affected*	*Both eyes affected*	*Male*	*Female*
Primary glaucoma	**144**	**249**	**24**	**15**	**105**	**93**	**51**
PCG	114	192	22	14	78	82	32
JOAG	30	57	2	1	27	11	19
Secondary glaucoma	**261**	**335**	**94**	**95**	**73**	**185**	**77**
Nonacquired ocular anomalies	**62**	**99**	**9**	**16**	**37**	**37**	**25**
Axenfeld Rieger anomaly/syndrome	7	11	1	2	4	4	3
Peter's anomaly	18	30	2	4	12	8	10
Ectropion uveae	2	3	1	0	1	0	2
Aniridia	18	33	0	3	15	13	5
Microspherophakia	3	6	0	0	3	2	1
Congenital hereditary endothelial dystrophy	2	4	0	0	2	2	0
Anterior staphyloma	2	2	1	1	0	1	1
Coat's disease	9	9	3	6	0	6	3
Retinal detachment	1	1	1	0	0	1	0
Nonacquired systemic disease/anomalies	**11**	**17**	**3**	**2**	**6**	**7**	**4**
Down's syndrome	1	1	1	0	0	1	0
Marfan syndrome	1	2	0	0	1	0	1
Rubella	2	3	1	0	1	2	0
Sturge Weber syndrome	3	3	1	2	0	1	2
Phacomatoses	1	2	0	0	1	0	1
Rubenstein–Tayabi	1	2	0	0	1	1	0
Stickler syndrome	1	2	0	0	1	1	0
Ectodermal dysplasia	1	2	0	0	1	1	0
Acquired conditions	**160**	**181**	**77**	**64**	**20**	**128**	**33**
Uveitis	2	3	1	0	1	0	2
Trauma (blunt)	54	54	32	22	0	49	5
Trauma (penetrating)	13	13	5	8	0	9	4
Steroid-induced	16	31	0	1	15	12	4
Retinoblastoma	26	27	11	14	1	20	6
ROP	2	2	1	1	0	2	0
Postvitreo-retinal surgery	36	39	21	14	2	29	8
Post-penetrating keratoplasty	11	12	6	4	1	7	4
Following cataract surgery	**28**	**38**	**5**	**13**	**10**	**13**	**15**
Aphakic	15	21	2	7	6	6	9
Pseudophakic	13	17	3	6	4	7	6
Total	**405**	**584**	**24**	**15**	**105**	**278**	**128**

The bold digits are the major headings according to the CGRN classification system, the others are the categories under the bold subheadings

surgery. At presentation, 59 children (14.57%) were ≤12 months of age (mean—4.3 ± 2.15 months, median—4.16 months, and range—8 days to 8.3 months), 112 children (27.65%) were between 1 and 5 years (mean—2.97 ± 1.5 and median—3 years), and 234 children (57.78%) were aged between 6 and 16 years (mean—11.23 ± 3.3 and median—11 years). The most common age of presentation in children with primary glaucoma was <1 year of age (32.64%), and in children with secondary glaucoma was 11–16 years (42.15%).

Most common presenting complaints among children with primary glaucoma were lacrimation (35.34%), photophobia, and diminution of vision, whereas in children with secondary glaucoma was a diminution of vision (64%) (Table 2). Of these, 28 children (19.44%) with primary glaucoma had already undergone glaucoma surgery before visiting our hospital. Positive family history of glaucoma was found in 10.42% of the patients (15 patients) with primary glaucoma and 5% (13 patients) with secondary glaucoma. Among primary glaucoma, positive family history was seen equally in PCG as well as JOAG patients. Among secondary glaucoma, association with positive family history was seen most commonly in patients with Aniridia, Axenfeld Rieger anomaly, and Peter's anomaly. A history of consanguineous marriage was present in seven out of 15 children with primary glaucoma (4.86% of patients with positive family history).

In total, 230 eyes (39.4%) had visual acuity better than 20/40, 10% (59 eyes) had visual acuity between 20/60 and 20/200, 2.2% eyes (13 eyes) had visual acuity between 20/200 and counting fingers, whereas 6.51% eyes (38 eyes) had hand motions or

Table 2: Presenting complaints in various types of childhood glaucoma

Glaucoma category	*Diminution of vision (%)*	*Leucocoria (%)*	*Lacrimation (%)*	*Photophobia (%)*	*Enlargement of the eyeball (%)*	*Squinting of eyes (%)*	*Pain (%)*	*Second opinion (%)*	*Others (%)*
Primary glaucoma	25.30	10.44	35.34	25.30	8.03	2.01	–	23.69	4.02
PCG	21.35	13.54	45.83	32.81	10.42	1.56	–	18.75	–
JOAG	38.60	–	–	–	–	3.51	–	40.35	17.54
Secondary glaucoma	63.28	9.85	5.37	6.27	2.09	2.09	2.69	15.22	2.69
Nonacquired ocular anomalies	50.51	10.10	15.15	16.16	4.04	6.06	3.03	15.15	3.03
Nonacquired systemic diseases	41.18	11.76	11.76	11.76	5.88	–	–	29.41	–
Acquired conditions	69.61	11.60	0.55	0.55	1.10	0.55	3.31	13.26	3.31
Following cataract surgery	76.32	–	–	5.26	–	–	–	18.42	–
Total	47.09	10.10	18.15	14.38	4.62	2.05	1.54	18.84	3.25

Table 3: Presenting IOP and IOP at last follow-up in patients with childhood glaucoma

	IOP at presentation			*IOP at the last follow-up*					
Glaucoma category	*≤20 mm Hg (%)*	*21–30 mm Hg (%)*	*>30 mm Hg (%)*	*1–10 mm Hg (%)*	*11–20 mm Hg (%)*	*21–30 mm Hg (%)*	*>30 mm Hg (%)*	*Cannot be assessed*	*Lost to follow- up*
Primary glaucoma	73 (29.32%)	104 (41.7%)	72 (28.92%)	46 (18%)	153 (61%)	9 (4%)	7 (3%)	2 (1%)	32 (113%)
PCG	61 (31.77%)	75 (39.06%)	56 (29.17%)	38 (20%)	112 (58%)	4 (2%)	6 (3%)	2 (1%)	30 (16%)
JOAG	12 (21.05%)	29 (50.88%)	16 (28.07%)	8 (14%)	41 (72%)	5 (9%)	1 (2%)	–	2 (4%)
Secondary glaucoma	41 (12.24)	125(37.31%)	169 (50.45%)	43 (13%)	206 (61%)	28 (8%)	10 (3%)	26 (8%)	22 (7%)
Nonacquired ocular anomalies	15 (15.15%)	46 (46.46%)	38 (38.38%)	10 (10%)	59 (60%)	11 (11%)	6 (6%)	4 (4%)	9 (9%)
Nonacquired systemic diseases	6 (25.29%)	6 (35.39%)	5 (29.41%)	2 (12%)	10 (59%)	–	–	–	5 (29%)
Acquired conditions	11 (6.08%)	64 (35.36%)	106 (58.56%)	25 (14%)	111 (61%)	12 (7%)	4 (2%)	21 (12%)	8 (4%)
Following cataract surgery	9 (23.68%)	9 (23.68%)	20 (52.63%)	6 (16%)	26 (68%)	5 (13%)	–	1 (3%)	–
Total	114 (19.52%)	229 (39.21%)	241 (41.27%)	89 (15%)	359 (61%)	37 (6%)	17 (3%)	28 (5%)	54 (9%)

less. Around 244 eyes (41.8%) had recorded vision as fixing and following the light.

The details of IOP at the presentation are given in Table 3. About 20% of the eyes with childhood glaucoma presented with IOP ≤20 mm Hg (mean—15.63 ± 4.4, median—17, and range—2–20 mm Hg), whereas 80% of the eyes presented with IOP >20 mm Hg (mean—32.80, median—30, and range—21–72 mm Hg). Around 70% of the eyes with primary glaucoma and 88% with secondary glaucoma presented with IOP >20 mm Hg. In eyes with PCG, around 68% of the eyes presented with >20 mm Hg (mean—31.78 ± 9.4, median—28.5, and range—21–70 mm Hg), whereas in JOAG, 79% of the eyes presented with high IOP (mean—29.47 ± 7, median—28, and range—21–50 mm Hg). In eyes with glaucoma due to acquired causes, 94% of the eyes had a presenting IOP >20 mm Hg (mean—33.97 ± 9.6, median—33.5, and range—21–72 mm Hg), whereas 76% of the eyes with glaucoma post cataract surgery had IOP >20 mm Hg (mean—30.1 ± 9.5, median—32, and range—21–48 mm Hg). Among the acquired causes of secondary glaucoma, all the patients presented with IOP >20 mm Hg in eyes with glaucoma secondary to uveitis, trauma, ROP, and postcorneal surgery.

On fundus evaluation, around 70% of the eyes with childhood glaucoma had a cupping of >0.7:1 on presentation. Around 61% of the eyes with PCG and 96.5% of those with JOAG had a cupping of >0.7:1. In eyes with secondary glaucoma, around 71% had a cupping of >0.7:1 whereas 23.6% had no fundus view. In eyes with acquired glaucoma, the percentage of eyes with cupping >0.7:1 was 79.3, 87, 82, 75, and 87% in glaucoma posttrauma, steroid-induced, postvitreoretinal surgery, postcorneal surgery, and postpediatric cataract surgery, respectively.

Of the 584 eyes, 272 eyes (46.6%) were treated medically, 282 eyes (48.29%) required surgical intervention, and 30 eyes (5%) were treated by laser/cyclodestructive treatment. About 82 eyes (14%) were also advised to undergo surgical/cyclodestructive procedures. The treatment details are given in Tables 4 and 5. In PCG eyes, 135 eyes (70%) underwent a surgical intervention—the most common being trabeculectomy and trabeculectomy (87%), followed by tube implant (10%). Rest 57 eyes (30%) were managed medically after the primary surgical intervention was done somewhere else or surgery was advised, as in 30 eyes (16%). In eyes with secondary glaucoma, 53% were managed medically, 40% underwent surgery, whereas 7% underwent cyclodestructive procedures or peripheral iridotomy. About 13% of the patients have further advised a surgery or cyclodestructive procedure. About 67% of JOAG eyes, 54% of eyes with nonacquired ocular anomalies, 59% with systemic diseases, and 74% of eyes with glaucoma following

Table 4: Management in various types of childhood glaucoma

Glaucoma category	*Medical management (%)*	*Surgical intervention (%)*	*Laser procedure/cyclodestructive procedure (%)*	*Advised surgery/ cyclodestructive procedure (%)*
Primary glaucoma	95 (38)	150 (60.24)	4 (2)	38 (15)
PCG	57 (30)	135 (70)	–	30 (16)
JOAG	38 (67)	15 (26)	4 (7)	8 (14)
Secondary glaucoma	177 (53)	132 (40)	26 (7)	44 (13)
Nonacquired ocular anomalies	53 (54)	34 (34.34)	12 (12.12)	13 (13)
Nonacquired systemic diseases	10 (59)	7 (41.18)	–	6 (35)
Acquired conditions	86 (48)	84 (44.41)	11 (6)	22 (12)
Following cataract surgery	28 (74)	7 (18.4)	3 (8)	3 (8)
Total	272 (46.58)	282 (48.29)	30 (5.14)	82 (14.04)

Table 5: Surgical management details in various types of childhood glaucoma

Glaucoma category	*Trabeculectomy (%)*	*Trabeculectomy + trabeculotomy (%)*	*Tube implant (%)*	*Iridectomy (%)*	*Lens extraction (%)*	*Enucleation (%)*	*Total*
Primary glaucoma	15 (11)	123 (82)	12 (9)	–	–	–	150
PCG	4 (3)	119 (88.15)	12 (10)	–	–	–	135
JOAG	11 (73)	4 (27)	–	–	–	–	15
Secondary glaucoma	38 (28)	15 (11.36)	39 (29)	6 (4)	8 (6)	26 (19)	132
Nonacquired ocular anomalies	11 (28)	8 (23.53)	4 (10)	5 (13)	4 (10)	2 (5)	34
Nonacquired systemic diseases	–	4 (57.14)	2 (33)	–	1 (17)	–	7
Acquired conditions	26 (31)	3 (3.57)	27 (33)	1 (1)	3 (4)	24 (29)	84
Following cataract surgery	1 (14.29)	–	6 (85.71)	–	–	–	7
Total	53 (19)	132 (48)	51 (18)	6 (2)	8 (3)	26 (10)	282

cataract surgery were managed medically. In secondary glaucoma posttrauma, 32 eyes (48%) were managed medically, whereas 31 eyes (46%) required surgery—the most common being tube implantation followed by trabeculectomy. Around eight eyes (12%) were also advised surgery/cyclodestructive procedure. In steroid-induced glaucoma, 17 eyes (55%) were medically managed, 13 eyes (42%) underwent surgery, whereas five eyes (16%) were advised surgery/cyclodestructive procedure. In children with retinoblastoma, 89% underwent enucleation, whereas the rest were advised enucleation/chemoreduction as soon as possible. In eyes with secondary glaucoma postvitreoretinal surgery, 31 eyes (79%) were managed medically, and six eyes (15%) were managed surgically—tube implantation being the surgery of choice. In 12 eyes with glaucoma post-penetrating keratoplasty, seven eyes (58%) were managed surgically—tube implantation being the most common procedure, whereas three eyes (25%) were managed by cyclodestructive procedure.

In PCG and JOAG groups, there was a significant reduction in mean IOP postmedical/surgical intervention with 78% (mean—3.1 ± 5 and range—1–16 mm Hg) and 86% (mean—12.02 ± 5, range—4–16 mm Hg, and median—3.5 mm Hg) of the eyes respectively having IOP ≤ 20 mm Hg (Table 5). In eyes with secondary glaucoma, 13% of the eyes had an IOP of ≤10 mm Hg, whereas 61% of the eyes had an IOP of 11–20 mm Hg. In total, 74% of the eyes had an IOP ≤ 20 mm Hg (mean—13.89 ± 3.45, median—14, and range—2–20 mm Hg) after the management of secondary glaucoma. Follow-up duration was also variable among various groups, with 49% of the children having a follow-up duration of ≤6 months (mean—2.86 months), 18% with a follow-up of up to 1 year (mean—10.07 months), and the rest 32% with a follow up of 1–5 years (mean—30.39 months).

Discussion

Various authors have used variable age cutoffs, IOP, visual fields, and other criteria to diagnose and monitor response to glaucoma treatment. Because of this, the reported prevalence and outcomes of the disease are not uniform. Also, there is scant data about childhood glaucoma from the northern part of India, especially that which conforms to the CGRN classification. Our study aims to bridge this lacuna in the understanding of childhood glaucoma in the region.

In our study, there were 405 children (584 eyes) with childhood glaucoma. The most common subtype of glaucoma in our study was found to be secondary glaucoma (64%). Previously, some studies have reported a higher prevalence of PCG,[6,7,11–13] whereas others had a higher number of children with secondary glaucoma.[5,8–10] These differences may be due to various factors—race, socioeconomic status, education, awareness, and availability of healthcare services. Also, the major drawback of these studies is that variable definition systems/age groups are used for childhood glaucoma. In our cohort, secondary glaucoma was found to be the most common subtype. This may be because our tertiary care center receives many referrals for complicated cases, while primary glaucoma is managed by other smaller centers as well in North India.

In our study, the majority of primary glaucoma cases were PCG (79%), followed by JOAG (21%). Among the secondary glaucoma

majority were glaucoma associated with the acquired condition (61%)—the most common being trauma, postvitreoretinal surgery, retinoblastoma, and steroid-induced glaucoma. Trauma was the most common cause of secondary glaucoma in our study. Thus, preventing trauma, advising protective eyewear, encouraging safe and supervised practices in sports, and educating parents to prevent access to sharp objects/toys can help in decreasing the incidence of posttraumatic glaucoma. Postvitreoretinal and postcorneal surgery was also found to be important causes of glaucoma in our study. Educating parents about the likelihood of glaucoma development and lifelong follow-up can help in the early diagnosis and management of these cases. Retinoblastoma was also an important cause in our study, thus highlighting the importance of strengthening awareness programs in the community. Retinoblastoma can also present as a masquerade of PCG with corneal edema/lacrimation/buphthalmos and can be missed, so detailed evaluation, including ultrasound B-scan, is warranted in all patients with poor fundus visibility.

Most of the cases of childhood glaucoma had unilateral involvement (56%). Primary glaucoma mostly had bilateral involvement (73%), while secondary glaucoma was mostly unilateral (72%). About 68% of the children with childhood glaucoma were males. This preponderance of boys can be explained by the gender bias that exists in our patriarchal society—girls are often neglected, and the healthcare-seeking behavior for females is generally poorer. In addition, boys are more actively involved in indoor as well as outdoor activities, making them more susceptible to trauma.

Around 58% of children with glaucoma were aged 5–16 years. Thus, awareness about the disease pathology is very important for parents as well as schoolteachers. Also, the school screening programs should be strengthened and conducted on a regular basis.

Most common presenting complaints were lacrimation, photophobia, and diminution of vision in primary glaucoma. Since early diagnosis and management can help in reversing the disease pathology, awareness among the parents about the presenting complaints is essential. Most frequent complaint in secondary glaucoma was a diminution of vision, thus stressing the fact that awareness should be created among the parents of children with decreased vision to get their children evaluated by local ophthalmologists and seek expert opinion by specialists as and when required.

Positive family history of glaucoma was seen in 10.42% of patients with primary glaucoma, and about 50% of these also had a consanguineous marriage. This highlights the importance of counseling parents with a family history of glaucoma. Genetic counseling should also be advocated. Around 70% of the eyes with primary glaucoma and 88% of eyes with secondary glaucoma had IOP of >20 mm Hg. Amongst primary glaucoma children, 68% of eyes with PCG and 79% of eyes with JOAG had IOP of >20 mm Hg. Although there is a higher baseline IOP in JOAG/secondary glaucoma, children may be asymptomatic. Among children with secondary glaucoma due to acquired causes, all eyes presented with IOP of >20 mm Hg, highlighting the morbidity associated with these conditions, thus making their management difficult.

About 50.5% of the eyes with childhood glaucoma had a visual acuity worse than 20/200 indicative of significant visual disability associated with childhood glaucoma. At presentation, 80% of the eyes presented with an IOP of >20 mm Hg and 70% of the eyes had a cupping of >0.7:1, highlighting the morbidity associated with childhood glaucoma. Around 96.5% of those with JOAG had a cupping of >0.7:1, thus highlighting visual morbidity associated with JOAG, thus making fundus examination in all children who present for routine evaluation to an ophthalmologist essential.

In the PCG group, 70% of the eyes were managed surgically. Rest 30% was managed medically after the primary surgical intervention was done somewhere else or surgery was advised (16%). In eyes with secondary glaucoma, 53% were managed medically, and 40% underwent surgery. About 13% of the patients were further advised of a surgical intervention or cyclodestructive procedure. Since PCG is a surgical condition, most of our patients were managed by surgical intervention. However, secondary glaucoma, even in children, has medical management as the mainstay and surgical intervention is advised when medical management is not effective/feasible. There was a significant reduction in IOP (<20 mm Hg) postmanagement of glaucoma in all the groups—PCG (78%), JOAG (86%), and secondary glaucoma (76%) thus highlighting the importance of management in these cases.

A major limitation of this study is its retrospective nature. Also, there is little data to support the geographic localization of areas with a higher prevalence of childhood glaucoma. That said, this study has a large sample size, and the CGRN classification, as well as the profiling of childhood glaucoma patients at presentation, provides a wealth of data for epidemiologists and policymakers alike. Given that it is the first study of this nature from Northern India, it can aid in the formulation of guidelines for strengthening screening programs, as well as the management of childhood glaucoma.

In conclusion, understanding the disease pattern, its presenting features, and the proportion of different types of childhood glaucoma can help in planning appropriate eye care services, creating awareness, and better-allocating resources to plan appropriate management strategies. Screening programs and counseling of parents should be strengthened, especially in conditions that predispose to secondary glaucoma in children.

References

1. Durnian JM, Cheeseman R, Kumar A, et al. Childhood sight impairment: a 10-year picture. Eye (Lond) 2010;24(1):112–117. DOI: 10.1038/eye.2009.32
2. Bhattacharjee H, Das K, Borah RR, et al. Causes of childhood blindness in the northeastern states of India. Indian J Ophthalmol 2008;56(6):495–499. PMID: 18974521.
3. Dorairaj SK, Bandrakalli P, Shetty C, et al. Childhood blindness in a rural population of southerh India: prevalence and etiology. Ophthalmic Epidemiol 2008;15(3):176–182. DOI: 10.1080/09286580801977668
4. European Glaucoma Society Terminology and guidelines for glaucoma, 4th edition-chapter 2: classification and terminology. Br J Ophthalmol 2017;101(5):73–127.
5. Papadopoulos M, Cable N, Rahi J, et al. The British Infantile and Childhood Glaucoma (BIG) Eye Study: Invest Ophthalmol Vis Sci 2007;48(9):4100–4106. DOI: 10.1167/iovs.06-1350
6. Taylor RH, Ainsworth JR, Evans AR, et al. The epidemiology of pediatric glaucoma: the Toronto experience. J AAPOS 1999;3(5):308–315. DOI: 10.1016/s1091-8531(99)70028-5
7. Alanazi FF, Song JC, Mousa A, et al. Primary and secondary congenital glaucoma:baseline features from a registry at King Khaled Eye specialist Hospital, Riyadh, Sudi Arabia. Am J Ophthalmol 2013;155(5):882–889. DOI: 10.1016/j.ajo.2012.12.006
8. Fang Y, Long Q, Guo W, et al. Profile of pediatric glaucoma patients in Shanghai Eye, Ear, Nose and Throat Hospital. Chin Med J (Engl) 2014;127(8):1429–1433. PMID: 24762583.
9. Aponte EP, Diehl N, Lohney BG. Incidence and clinical characteristics of childhood glaucoma: a population based study. Arch Ophthalmol 2010;128(4):478–482. DOI: 10.1001/archophthalmol.2010.41

10. Fung DS, Roensch MA, Kooner KS, et al. Epidemiology and characteristics of childhood glaucoma: results from the Dallas Glaucoma Registry. Clin Ophthalmol 2013;7:1739–1746. DOI: 10.2147/OPTH.S45480
11. Qiao CY, Wang LH, Tanq X, et al. Epidemiology of hospitalized pediatric glaucoma patients in Beijing Tongren Hospital. Chin Med J (Engl) 2009;122(10):1162–1166. PMID: 19493464.
12. Senthil S, Badakere S, Ganesh J, et al. Profile of childhood glaucoma at a tertiary centre in South India. Indian J Ophthalmol 2019;67:358–365. DOI: 10.4103/ijo.IJO_786_18
13. Barsoum-Homsy M, Chevrette L. Incidence and prognosis of childhood glaucoma. A study of 63 cases. Ophthalmology 1986;93(10):1323–1327. DOI: 10.1016/s0161-6420(86)33569-3