The British Infantile and Childhood Glaucoma (BIG) Eye Study

Maria Papadopoulos,1 Noriko Cable,2 Jugnoo Rahi,3,4 Peng Tee Khaw,1,2 and the BIG Eye Study Investigators5

PURPOSE. Pediatric glaucoma is a rare, potentially blinding condition, yet, in the United Kingdom, there is a paucity of contemporary epidemiologic and clinical data regarding this condition. The British Infantile and Childhood Glaucoma (BIG) Eye Study is the first national population-based study conducted to examine the incidence, detection patterns, current management, and intraocular pressure (IOP) control at 1 year in children with newly diagnosed glaucoma in the United Kingdom.

METHODS. A prospective study was conducted wherein children in the United Kingdom and Republic of Ireland aged ≤16 years with newly diagnosed primary or secondary glaucoma, were identified by consultant ophthalmologists through active surveillance from December 2001 until November 2002. Eligible cases were re-evaluated 12 months after notification.

RESULTS. Of the 99 eligible children with newly diagnosed glaucoma, 47 had primary and 52 secondary glaucoma. The annual incidence of diagnosis of primary congenital glaucoma (PCG) in Great Britain was 5.41 in 100,000 (1/18,500) live births and in the Republic of Ireland, 3.31 in 100,000 (1/30,200). The incidence of PCG in children of Pakistani origin was almost nine times that of Caucasians. IOP control of ≤21 mm Hg was achieved in 94% with medications (60% without medications) in cases of PCG and in 86% with medications (28% without medications) in cases of secondary glaucoma.

CONCLUSIONS. The British annual incidence of PCG diagnosis is comparable to that reported for other similar populations. Ethnic minorities from South Asia are at significantly increased risk of PCG. Successful IOP control in PCG after surgery in Britain is comparable to that in the published literature. (Invest Ophtalmol Vis Sci. 2007;48:4100–4106) DOI:10.1167/iovs.06-1350

G laucoma in infancy and childhood is a potentially blinding condition characterized by elevated IOP. Clinical research suggests that prognosis is largely dependent on early, accurate diagnosis and successful treatment involving intraocular pressure control to a level where progression is unlikely, along with the prevention of amblyopia.1 As it is uncommon, epidemiologic studies are difficult to undertake. It is estimated that a consultant ophthalmologist in a nonspecialist center in the Western world will expect to see a new case of PCG approximately every 5 years.2 As a result of its relative rarity, PCG is sometimes misdiagnosed or suboptimally treated, allowing irreversible corneal and optic nerve damage to occur. Consequently, it accounts for a disproportionate percentage (up to 18%) of children in blind institutions around the world.3,4 Overall glaucoma is responsible for 5% of blindness in children worldwide.5

Pediatric glaucoma is classified as primary when an isolated idiopathic developmental abnormality of the anterior chamber angle exists and secondary when aqueous outflow is reduced due to either a congenital or an acquired ocular disease or systemic disorder.6 Primary pediatric glaucoma includes PCG (isolated trabeculodysgenesis) and juvenile open-angle glaucoma (JOAG). PCG is the commonest glaucoma in infancy,7,8 but it has a variable reported incidence worldwide. A higher prevalence has been observed in genetically inbred populations and in certain ethnic and religious groups in which parental consanguinity, especially cousin-cousin marriages, is common.9–11 PCG has been reported to occur more frequently in males than in females12–14 and is reported to be bilateral in 70% to 80% cases.15,16 Familial cases tend to have an equal sex distribution.10,11,17 Secondary pediatric glaucoma is commonly associated with anterior segment dysgenesis, developing in 50% of cases.18 Glaucoma associated with aniridia is usually due to progressive angle closure, presenting often in childhood with an incidence ranging from 6% to 75%.19 Aphakic glaucoma can occur early or years after initial uneventful surgery, and has a variable incidence from 5% to 41%, depending on the age of surgery, corneal diameter and technique.20–22 Phacomatoses commonly associated with glaucoma are Sturge-Weber syndrome23 and Klippel-Trenaunay-Weber syndrome. The glaucoma seen in inflammatory disorders is multifactorial with the reported incidence as high as 38% in children with juvenile idiopathic arthritis.24 Surgery is the definitive treatment modality for the control of IOP in pediatric glaucoma, although medical treatment is first line in secondary glaucoma, because it is often successful in reducing IOP short term. The available surgical procedures have various indications, with both advantages and disadvantages and potentially good success rates, especially when performed at referral centers where there is sufficient volume to ensure both skillful surgery and safe anesthesia. However, the approach to management and success, as determined by IOP control, may vary around the world. We report, to our knowledge, the first prospective, national population-based study of pediatric glaucoma in the United Kingdom, including its incidence, detection patterns, and current clinical management practices and their short-term outcome in IOP control.

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The study investigators are listed in the Appendix.

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MATERIALS AND METHODS

In the United Kingdom, pediatric glaucoma is managed by glaucoma specialists with a pediatric interest and by pediatric ophthalmologists. Children eligible for the study were identified through the British Ophthalmic Surveillance Unit (BOSU), which was established in 1997 to facilitate the study of uncommon ophthalmic disorders. Within this scheme, all consultant ophthalmologists in the United Kingdom and the Republic of Ireland receive a report card each month to complete and return to the BOSU, either noting all eligible children seen during the previous month or confirming that they have no new cases to report.26 For a 12-month period (December 1, 2001, to November 30, 2002) active surveillance was undertaken whereby consultant ophthalmologists reported all children aged 16 years or less with newly diagnosed primary or secondary glaucoma. Glaucoma was defined as the presence of a combination of signs consistent with high IOP (≥21 mm Hg), either in the past or at present such as: disc cupping >0.3 or disc asymmetry ≥0.2, progressive disc cupping, buphthalmos (prominent, enlarged eye), enlarged corneal diameter (>11 mm in newborn, >12 mm in a child <1 year, or >13 mm in a child of any age), corneal edema, Descemet’s membrane splits (Haab’s striae), visual field defects, and progressive myopia. The presence of elevated IOP was not always necessary to enable the inclusion of spontaneously arrested glaucoma cases—that is, for example, enlarged eyes with Haab’s striae but normal IOP and optic discs. Cases with isolated disc cupping of >0.3 or disc asymmetry ≥0.2 in the absence other signs may have represented physiological cupping and so were excluded.

On notification, a standardized questionnaire developed specifically for this study was sent to the reporting ophthalmologist, to gather demographic and socioeconomic data and information on clinical presentation and initial management. Nonresponding ophthalmologists were reminded by mail and telephone. One year after notification, information regarding further management and IOP control was again represented by questionnaire. In recognition of the difficulties of assessing visual function and the degree of optic nerve damage in young patients, often with cloudy or scarred corneas, we defined therapeutic success, as in prior reports, as the lowering of IOP to ≤21 mm Hg.

Statistical Analysis

The incidence of diagnosis of PCG in the first year of life was determined for each country by dividing the number of newly diagnosed cases presenting in the first year of life by the number of live births for 2002. In addition, the incidence by ethnic groups and sex (according to the U.K. Office for National statistics taxonomy) were similarly calculated along with relative rates. Socioeconomic status was determined using a regional method based on postal codes in Great Britain. The Carstairs Index was used to assess the level of deprivation, based on the postal code of each child provided through the Web site at the Manchester Information and Associated Service. This score is a composition of four variables: unemployment, overcrowding, low social class, and non-car ownership. The score was obtainable only for people in Great Britain as a whole.

RESULTS

Ninety-five percent of notifications were associated with a completed questionnaire. Ninety-nine eligible children were identified: 47 with primary and 52 with secondary glaucoma (Table 1). Of these, three were lost to follow-up (one with PCG, two with secondary glaucoma). There were no cases reported from Northern Ireland; therefore, our data are based on the populations of Great Britain (England, Scotland, Wales) and the Republic of Ireland.

In a comparison of ethnic groups and type of glaucoma, 75% of Asian children (Indian, Pakistani, Bangladeshi) had primary glaucoma compared with 33% to 45% of children of other ethnic origins (Fig. 1).

When the proportion of primary to secondary glaucoma was compared by ethnicity, Caucasian children predominated in both groups. Children of Pakistani, Bangladeshi, and black origin from deprived areas were overrepresented, especially in those with primary glaucoma. Approximately 80% of Asians and blacks were living in the most deprived areas.

Descriptive Analyses were Undertaken in Relation to Detection and Clinical Management

The study was approved by Moorfields Eye Hospital local research and ethics committee. There was no direct patient contact and all data were handled in accordance with prevailing guidelines on data confidentiality and protection.

![Figure 1. Ethnic groups and glaucoma type (n = 99).](attachment:image-url)
73% of the children (Table 4).

Detection and Presentation. The diagnosis of primary glaucoma was made as a consequence of parental concern in 40% (two with mitomycin C [MMC] and one with 5 fluorouracil 6%(4/68) of cases, 75% of which were with an antimetabolite lotomy 16%). Trabeculectomy was the primary surgery in formed in 87% (59/68) of cases (goniotomy 71%, trabecu-

Primary Glaucoma

Of the 47 children with primary glaucoma, 45 had PCG and 2 had JOAG. The onset of PCG was congenital (≤3 months of age) in 38%, infantile (4 months–2 years) in 56%, and juvenile (2–16 years) in 6%. The data reported in the following sections refer to cases of PCG (73 eyes) unless stated otherwise.

Incidence. The annual incidence of newly diagnosed PCG in Great Britain by age 1 year was 5.41 in 100,000 live births (1/18,500), and in the Republic of Ireland it was 3.31 in 100,000 live births (1/30,200; Table 2).

An important variation by ethnic origin was found in the incidence of PCG. The highest incidence in a defined ethnic group was in children of Pakistani origin—almost nine times that of Caucasians, followed by Bangladeshi and Indians (Table 3). The incidence in Caucasians was the lowest of all.

The overall incidence did not vary by sex. Sixty-two percent of PCG cases were bilateral, and there was no difference regarding laterality in unilateral cases. Overall, a family history of childhood-onset glaucoma was found in 11% of PCG cases. By ethnicity, 50% (1/2) of black patients had a family history of childhood-onset glaucoma but only 9% of Asians and, similarly, 10% of Caucasians. There was parental consanguinity in 16% of cases (all Asian), with 67% of all Pakistani children in our study being from consanguineous marriages.

Detection and Presentation. The diagnosis of primary glaucoma was made as a consequence of parental concern in 73% of the children (Table 4).

Overall, 40% of the children with PCG presented to the referring or treating ophthalmologist by 3 months of age, 57% by 6 months, and 79% within the first 12 months. The mean age of presentation for children with PCG was 11 months (SD, 22.5; range, 0–127). More than half of the Asian children presented within the first 3 months of age, as opposed to the Caucasian children, 52% of which presented in the first 6 months of age. This difference was not statistically significant.

Primary cases were mostly referred to the treating ophthalmologist by a colleague ophthalmologist by 3 months of age, 57% referring or treating ophthalmologist by 6 months, and 79% within the first 12 months. The mean age of presentation for children with PCG was 11 months (SD, 22.5; range, 0–127). More than half of the Asian children presented within the first 3 months of age, as opposed to the Caucasian children, 52% of which presented in the first 6 months of age. This difference was not statistically significant.

Management. Because PCG is a rare condition, these children were largely treated at tertiary centers (89%): Moorfields Eye Hospital, London (45%); Great Ormond Street Hospital for Children, London (19%); Manchester Royal Eye Hospital (19%); and Birmingham Children’s Hospital (6%). Examination under anesthesia (EUA) was performed as part of the initial management in 7% of cases. The common anesthetics used were sevoflurane (41%) and ketamine (41%), followed by isoflurane (10%). The IOP was measured in 98% of cases, most commonly by a Perkins tonometer (78%) followed by a handheld device (19%) (Tonopen; Medtronics, Jacksonville, FL), and by noncontact devices (3%). The optic disc was visible in 73% of affected eyes. Refraction was performed in 56% of the children, and correction by glasses was prescribed in 9%. Ultrasound was performed in 36% of the patients. Visual fields were tested in only one patient who had JOAG.

Of the 73 eyes affected with PCG, information regarding management was available for 72 eyes, of which 94% (68/72 eyes) had surgery and 6% (4/72) had medical treatment only. Eighty-one percent of the patients had medication before surgery, and, on average, 1.7 eye drops were prescribed (range, 1–5 drops). Pilocarpine was the commonest drug prescribed, followed by betaxolol, latanoprost, dorzolamide, and combined timolol with dorzolamide. Acetazolamide usage was 22%. Almost 50% of affected eyes underwent surgery within 1 week of diagnosis, 71% within 3 weeks and 75% within 4 weeks. The median duration from the diagnosis to surgery was 2 weeks. Angle surgery was the commonest procedure, performed in 87% (59/68) of cases (goniotomy 71%, trabeculotomy 16%). Trabeculectomy was the primary surgery in 6%(4/68) of cases, 75% of which were with an antimetabolite (two with mitomycin C [MMC] and one with 5 fluorouracil

### Table 2. Annual Incidence of Diagnosis of PCG in the First Year of Life

<table>
<thead>
<tr>
<th></th>
<th>Great Britain</th>
<th>England</th>
<th>Scotland</th>
<th>Wales</th>
<th>Republic of Ireland</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cases</td>
<td>45</td>
<td>29</td>
<td>6</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Live births in 2002</td>
<td>646,887</td>
<td>565,709</td>
<td>51,270</td>
<td>30,205</td>
<td>60,503</td>
</tr>
<tr>
<td>Incidence (per 100,000)</td>
<td>5.41</td>
<td>5.13</td>
<td>11.70</td>
<td></td>
<td>3.31</td>
</tr>
<tr>
<td>95% CI</td>
<td>3.62–7.20</td>
<td>3.26–6.99</td>
<td>2.34–21.07</td>
<td>0–7.89</td>
<td>0–7.89</td>
</tr>
</tbody>
</table>

Source: Office for National Statistics

### Table 3. Incidence of PCG in Children ≤16 Years of Age by Ethnicity and Sex in Great Britain

<table>
<thead>
<tr>
<th>Ethnic group</th>
<th>Cases</th>
<th>Population</th>
<th>Incidence per 100,000 (95% CI)</th>
<th>Relative Rate (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>White</td>
<td>29</td>
<td>10,398,802</td>
<td>0.28 (0.25–0.31)</td>
<td>Reference*</td>
</tr>
<tr>
<td>Pakistani</td>
<td>6</td>
<td>2,433,944</td>
<td>2.46 (1.84–3.08)</td>
<td>8.82 (3.66–21.24)</td>
</tr>
<tr>
<td>Bangladeshi</td>
<td>1</td>
<td>100,884</td>
<td>0.99 (0.38–1.60)</td>
<td>3.55 (0.48–20.09)</td>
</tr>
<tr>
<td>Indian</td>
<td>2</td>
<td>229,524</td>
<td>0.87 (0.49–1.25)</td>
<td>3.12 (0.75–13.09)</td>
</tr>
<tr>
<td>Black</td>
<td>2</td>
<td>4,227,736</td>
<td>0.47 (0.26–0.67)</td>
<td>1.68 (0.40–7.03)</td>
</tr>
<tr>
<td>Chinese</td>
<td>1</td>
<td>28,326</td>
<td>3.53 (1.34–5.72)</td>
<td>12.66 (1.72–92.95)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>26</td>
<td>5,869,500</td>
<td>0.04 (0.03–0.07)</td>
<td>Reference*</td>
</tr>
<tr>
<td>Female</td>
<td>17</td>
<td>5,588,000</td>
<td>0.03 (0.02–0.05)</td>
<td>0.73 (0.39–1.34)</td>
</tr>
<tr>
<td>Socioeconomic status</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Upper quintiles (1, 2, 3)</td>
<td>18</td>
<td>6,874,500</td>
<td>0.26 (0.22–0.30)</td>
<td>Reference*</td>
</tr>
<tr>
<td>Lower quintiles (4, 5)</td>
<td>22</td>
<td>4,583,000</td>
<td>0.48 (0.42–0.54)</td>
<td>1.83</td>
</tr>
</tbody>
</table>

n = 41. Two children of Asian origin were excluded, as their race was unknown. Source: Office for National Statistics.

* Reference refers to the baseline group to which the other categories were compared.
Other 7% 8%

Surveillance of families with known
Clinical surveillance of children
with known ocular disease
Children health surveillance/screening 18% 19%

FIGURE 2.

uveitis (60% of cases) and, surprisingly, by phacomatoses (67% was 1.5:1, due to the high proportion of girls affected by segment dysgenesis (10%; Table 1). The ratio of boys to girls caused by phacomatoses (23%), uveitis (19%), and anterior affected. Most (31%) cases were lens-related, followed by those secondary glaucoma, 69 eyes were

In univariate analyses, there was no significant association between achieving IOP control (≤21 or ≤17 mm Hg) at 1 year and initial IOP, sex, ethnicity, family history, time to surgery from diagnosis, corneal diameter, age of diagnosis, procedure performed, and total number of procedures. Surgical complications were rare. One (1.5%) patient sustained lens trauma during the first procedure.

With regard to the management of JOAG, one patient was treated only medically and the other had a 5FU trabeculectomy in one eye and cycloidee laser treatment in the other.

Secondary Glaucoma

In the 52 patients with secondary glaucoma, 69 eyes were affected. Most (31%) cases were lens-related, followed by those caused by phacomatoses (23%), uveitis (19%), and anterior segment dysgenesis (10%; Table 1). The ratio of boys to girls was 1.5:1, due to the high proportion of girls affected by uveitis (60% of cases) and, surprisingly, by phacomatoses (67% of cases). All patients with juvenile idiopathic arthritis were girls but only half of those with idiopathic uveitis were girls. Most cases (67%) of secondary glaucoma were unilateral.

Six percent of the patients had a family history of childhood-onset glaucoma. These children had anterior segment dysgenesis (Rieger anomaly), aphakic glaucoma (congenital cataracts), and neurofibromatosis. Most (80%) of them were Caucasian. There was a history of consanguinity in four (8%) patients: three Asian and one Caucasian.

Detection and Presentation. Glaucoma was mainly detected through clinical surveillance—that is, routine follow-up of patients with diagnoses known to be associated with glaucoma (52%), such as uveitis, aphakia, pseudophakia, and the phacomatoses (Table 4). Disease was detected in 19% of cases when the children failed routine screening tests in which either the diagnosis associated with glaucoma was made or children presented with opaque corneas or buphthalmos in the perinatal period. As opposed to primary cases, only 11% of secondary cases were detected as a result of parental concern (difference between the two groups, 54%; 95% CI: 36%–69%,  P < 0.0001), and these were mostly cases of phacomatoses. No cases were detected as a result of a family history of glaucoma. The mean age at presentation was 46 months (SD 56; range, 0–178), as opposed to 11 months in primary glaucoma (F = 15.64, P < 0.001). In 51% of the children, the disease manifested in the first year of life and, in 53%, after more than 5 years of life. Most of the children with secondary glaucoma were referred to the treating ophthalmologist by a colleague for definitive management (58%). Pediatricians referred 31% and general practitioners only 7%.

Management. Similar to those with PCG, these patients were mainly managed at tertiary centers (80%): Moorfields Eye Hospital, London (40%); Great Ormond Street Hospital for Children, London (22%); Birmingham Children’s Hospital

TABLE 4. Mode of Detection Patterns

<table>
<thead>
<tr>
<th></th>
<th>Primary*</th>
<th>Secondary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parental concern</td>
<td>73%</td>
<td>11%</td>
</tr>
<tr>
<td>Child health surveillance/screening</td>
<td>18%</td>
<td>19%</td>
</tr>
<tr>
<td>Clinical surveillance of children with known ocular disease</td>
<td>0%</td>
<td>52%</td>
</tr>
<tr>
<td>Surveillance of families with known glaucoma history</td>
<td>2%</td>
<td>0%</td>
</tr>
<tr>
<td>Other</td>
<td>7%</td>
<td>8%</td>
</tr>
</tbody>
</table>

* In one case, mode of detection was unknown.

Data are percentage of the total cases.

† Information missing in five cases.

‡ Information missing in four cases.

![Figure 2. Percentage of patients with PCG undergoing surgery for IOP control within the first year after diagnosis (n = 73 eyes).](http://iovs.arvojournals.org/pdfaccess.ashx?url=/data/journals/iovs/932943/ on 04/03/2017)
Pediatric glaucoma is a potentially blinding disease that is associated with elevated IOP. Prognosis is largely related to the timing of presentation, and so early diagnosis and prompt surgical treatment significantly influence the visual outcome. Over the last 50 years there has been a dramatic improvement in the prognosis of this disease because of the introduction of techniques such as angle surgery and trabeculectomy along with anti-fibrotic treatment. We undertook this prospective, U.K. population-based survey to determine the incidence of glaucoma in children and establish detection patterns, clinical management and successful IOP control at 1 year. In the absence of a second independent source of cases, we were unable to assess completeness of ascertainment using capture-recapture analysis but the number of new cases reported was within the anticipated range.

Our findings regarding the annual incidence of diagnosis of PG in the first year of life in Great Britain (equivalent to 1 in 18,500 live births) are consistent with those reported in other Western countries, although direct comparisons are difficult because of methodological differences. One of the major findings of our study was ethnic origin as a major risk factor for PG: South Asian children of consanguineous parents were at particular risk. For instance, Pakistani children had almost nine times the risk of PG when compared with white Caucasian children.

In PCG, clouding of the cornea is the most frequent physical sign first recognized by parents or doctors. Therefore, it is not surprising that the diagnosis of PCG most often occurred as a result of parental concern and that these children were more likely to present to accident and emergency for immediate medical attention than were children with secondary glaucoma. Since the timing of diagnosis and subsequent surgical intervention is important to the visual prognosis, it is crucial that general practitioners and casualty officers be familiar with the relevant signs and the need for prompt referral. Secondary pediatric glaucoma was mainly detected through routine follow-up of patients with diagnoses known to be associated with glaucoma. This emphasizes the need for strict surveillance of children at risk.

Despite recent major advances in the genetics of PCG, our knowledge is incomplete. Our finding of a family history in PG in 11% of cases agrees with that published in the literature of between 4% and 40%. However, most cases of PCG appear to be sporadic. Autosomal recessive inheritance with variable penetrance ranging from 40% to 100% is reported in familial cases, especially where there is a high incidence of the disease due to parental consanguinity. In this study, the incidence of consanguinity in PCG was 16%, and in all cases the parents were Asian. This may account for the disproportionate number of PCG in the Asian ethnic group.

In this study, the incidence of consanguinity in PCG was 16%, and in all cases the parents were Asian. This may account for the disproportionate number of PCG in the Asian ethnic group. The increased incidence of PCG in consanguineous marriages emphasizes the importance of inquiring about consanguinity and screening siblings if it is present, along with offering these couples genetic counseling.

In most cases, PCG presents at <6 months of age, >80% within the first year. Our findings are in agreement, in that in 40% of the cases PCG presented within 3 months of birth and in 75% within the first 12 months. In only three (6%) cases did it present in children older than 5 years. The mean age of presentation was 11.3 months, similar to the 10.6 months found by Barsoum-Homys and Chevrette. Although it is a rare condition, PCG is an important diagnosis that should not be missed in the first year of life.

An appropriate anesthetic is vital in the assessment of children with pediatric glaucoma. As a general rule, all anesthetics lower the IOP, with the possible exception of ketamine, chloral hydrate, and N2O. In certain subtle cases of glaucoma, this effect can have a profound impact on the timing of the diagnosis and the visual outcome. Ketamine hydrochloride is believed to result in IOP measurements that are commensurate with those taken in awake infants. Of the children with PCG undergoing an EUA in this survey, 41% had ketamine anesthesia. Fewer EUAs were performed in children with sec-
ondary glaucoma than in those with primary glaucoma, consistent with the older age of those with secondary glaucoma.

The management of pediatric glaucoma has improved considerably over the past few decades. As this is a rare condition, we found that most cases were managed at specialist centers: Moorfields Eye Hospital (42.5%); Great Ormond Street Hospital for Children (20.5%); Royal Eye Hospital, Manchester (13.5%); and Birmingham Eye Hospital (8%). The commonest surgery for PCG in our study was angle surgery, which was successful in controlling IOP with medications in 88% of cases. This success rate falls within the range reported for goniectomy, with IOP control at 1 year after multiple operations of 72% to 93.5%, and is similar to that of trabeculectomy. It is important to highlight that there was only a 1-year follow-up in our study and that these glaucomas are known to relapse with time.

Enlarged ocular dimensions are thought to influence surgical prognosis, as they are a measure of structural damage the angle has sustained over time. A corneal diameter of $\geq 14$ mm in diameter has been associated with worse prognosis in some reports. We and others have not been able to find this association. For example, Haas was unable to find an association with corneal diameter in those infants presenting at birth or within two months, in which case the poor prognosis may instead be explained by the severity of the disease. Other risk factors for failure include family history and being female.

We failed to find an association between IOP control and initial IOP, sex, ethnicity, family history, time to surgery from diagnosis, corneal diameter, age of diagnosis, procedure performed, total number of procedures, and achieving IOP control of $\leq 21$ mm Hg. However, the small number of cases in our study may have limited our power to detect true associations that had existed.

Although the mainstay of treatment for pediatric glaucoma is surgery, medical treatment is often first-line in secondary glaucoma. Hence, our finding that the mean time from diagnosis to surgery was significantly longer in children with secondary glaucoma (median 4 weeks) than in those with primary glaucoma (median 2 weeks) was not surprising. This finding also explains why those with secondary glaucoma were prescribed more eye drops than those with primary glaucoma. Trabeculectomy was the most common surgery in these children in preference to angle surgery, probably because of its poorer success rate in secondary glaucoma. Seventy-one percent of children were receiving medications after 1 year of follow-up. The poor success rate without medications in 28% reflects the aggressive nature of secondary glaucoma and the fact that medications are first line. However, success with medical treatment was 86%, suggesting that topical medications are a critical component of management in pediatric glaucoma, despite the lack of formal licensing in children. Furthermore, it highlights the need for more research on the efficacy of topical medications in children.

**CONCLUSION**

The incidence of PCG in our study was consistent with that found in the literature for similar populations. South Asian children are at major risk of PCG. This finding is likely to reflect a higher incidence of monogenic disease in consanguineous families and highlights the importance of screening siblings and offering genetic counseling in these cases. As parental concern was the main reason leading to the diagnosis of PCG, healthcare professionals should ensure that parental reports are heeded. Stringent surveillance of children at risk of secondary glaucoma is needed, as it led to the diagnosis of glaucoma in most cases in our survey. When comparing primary and secondary glaucoma, significant differences were found. Primary cases were more likely to be treated earlier and surgically with preference to goniectomy. More secondary glaucoma cases were managed medically before surgery and with a greater number of topical medications. The most frequent topical treatment was pilocarpine in primary glaucoma and latanoprost in secondary glaucoma. Sixty percent of children with primary glaucoma and 28% with secondary glaucoma achieved IOP control ($\leq 21$ mm Hg) at 1 year without additional medication. Thus, although prognosis has improved for these disorders over the years, further improvements in clinical practice are still required.

**References**


**APPENDIX**

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