Primary Congenital Glaucoma: Trends in Presentation Over 3 Decades at a Tertiary Eye Care Center in India

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Precis: In this study, huge time delay was observed between first primary congenital glaucoma (PCG) symptom recognition by parents to ophthalmological diagnosis and this delay was unchanged over time. Bilateral afferent and mean presenting intraocular pressure (IOP) remained unaltered.

Purpose: To compare the trends in presentation of patients with PCG over 3 different time periods at 10-year intervals (1998, 2008, and 2018) managed by a single surgeon at one of the largest tertiary eye care centers in South India.

Methods: Retrospective analysis of 313 eyes of 172 patients diagnosed to have PCG and treated at the L V Prasad Eye Institute (LVPEI) between January and December of the 3 specified years. Data collected included age at symptom recognition and at presentation, sex, affected eye, signs and symptoms, time between onset of symptoms and diagnosis (diagnostic delay), corneal clarity, and IOP.

Results: Of the 172 patients, 48 (27.9%), 76 (44.2%), and 48 (27.9%) belonged to 1998, 2008, and 2018, respectively. Bilateral afferent (79% to 84%) was common and there was no sex predilection. The median age at recognition of first symptoms by caregivers/parents was 1 day (interquartile range, 1 to 30), and at diagnosis was 120 days (interquartile range, 30 to 378). Overall, presentation to LVPEI was delayed by >3 months in 80 (46.5%) children, and there was no statistically significant difference in the duration of delay over time (P = 0.13). There was no significant difference in the mean presenting IOP over time (P > 0.05).

Conclusions: We provide unique single center-based information of trends in the presentation of patients with PCG over 3 decades. Diagnostic delay was common, and a large number of characteristics of patients with PCG in South India have remained unchanged, particularly, age at onset and at diagnosis and laterality.

Key Words: primary congenital glaucoma, surgical intervention, combined trabeculectomy-trabeculotomy, delay, India

Primary congenital glaucoma (PCG) is a rare disease associated with idiopathic isolated developmental anomaly of the anterior chamber angle. Based on the onset of the disease, PCG is classified into 3 types: neonatal onset (0 to 1 mo), infantile onset (<1 to 24 mo), and late onset (> 24 mo). stool The classical presentation of PCG consists of a triad of symptoms that include photophobia, tearing, and blepharospasm. Common signs of PCG include corneal edema, corneal edema, buphthalmos, elevated intraocular pressure (IOP), and Haab's striae. stool PCG manifests at birth or within few months to years after birth. stool Despite its low incidence, PCG may comprise >5% of the general glaucoma population. stool It may occur up to 10 times more frequently in certain ethnic groups in different geographic locations where consanguineous relationships or inbreeding is common. In Southern India, the prevalence of PCG is estimated at 1 in 3300 live births, accounting for 4.2% of overall childhood blindness.

Early diagnosis and prompt microsurgical intervention are the prerequisites to optimize the visual and surgical outcomes in PCG. Factors such as earlier onset of the disease, and delay in surgical intervention have previously been linked to poor outcomes in PCG. This resulted in established treatment paradigm that early intervention within a few days to a few weeks of life improves outcomes in PCG. Furthermore, the chances of success are highest with the first surgical procedure. In 2013, the Childhood Glaucoma Research Network, an international consortium of glaucoma specialists, summarized the consensus statements for glaucoma surgery in children and stated that children with most engaged caregivers often have the best outcomes.

As mentioned earlier, a successful outcome is highly dependent on the age at surgery in children with PCG. However, the chances of early treatment are sometimes hindered by a prolonged interval between the onset of symptoms and the parents of children with PCG seeking medical care (patient delay), or between hospital presentation and the eventual diagnosis (doctor delay). Furthermore, logistical challenges within hospitals might cause unwanted delays in treatment (system delay). These barriers are important to address as the development of visual impairment at an early age has far reaching implications on a child's overall development and life. Surgical delays which result in poor outcomes burden both the families affected and the society as a whole. Our hospital is a tertiary eye care center located in South India, with a dedicated children's eye care facility for childhood glaucoma. Our center has been operational for over 3 decades now.

Over 2 decades ago (1998), we reported the clinical characteristics and surgical outcomes of 120 children with PCG seen consecutively at our center over a 5-year period between 1990 and 1995. A decade later (2007), we reported the clinical characteristics and long-term surgical outcomes of 624 eyes of 360 children with PCG treated consecutively at our center over a 15-year period between 1990 and 2004. Given that engaged

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parents/caregivers achieve the best outcomes, it is important to understand the role of different types of delay, if any, that influence the outcomes of children with PCG. To the best of our knowledge, there is paucity of literature regarding this. Therefore, the aims of the present study were to analyze the trends in presentation characteristics, and different types of delays, and compare them across 3 different time periods at 10-year intervals (1998, 2008, and 2018).

**METHODS**

A retrospective chart review of a prospectively maintained database (L V Prasad Eye Institute Congenital Glaucoma Registry\textsuperscript{18}) at the L V Prasad Eye Institute (LVPEI), South India, was conducted to identify all the children who were surgically treated for PCG (unilateral or bilateral) by the same glaucoma specialist in a single glaucoma clinic between 1998 and 2018. We excluded patients with secondary childhood glaucomas. All the patients underwent primary combined trabeculotomy-trabeculectomy.\textsuperscript{1,6,16,19} In case of bilateral PCG, the choice of simultaneous bilateral combined trabeculotomy-trabeculectomy was offered to all patients after thoroughly discussing the risks with the parents.\textsuperscript{20} From the database, 3 specific years with 10-year intervals—1998, 2008, and 2018 were identified. Patient information was collected from electronic (2018) as well as paper charts (1998 and 2008). This study was approved by the Institutional Review Board of the LVPEI, Hyderabad, India and conducted in accordance with the tenets of the Declaration of Helsinki.

Data collected included age at which symptoms were first noticed by caregivers/parents, sex, laterality, age at first ophthalmological consultation, age at surgery, type and number of symptoms, place of residence, date of first consultation at LVPEI, and date of glaucoma surgery. In addition, we collected the socioeconomic status of the family (based on a matrix that includes a number of variables such as type of house ownership, family income, number of items in the household, etc.) used by our center to classify patients into different categories. This is in accordance with the center’s policy that enables everyone who needs the care to receive it, regardless of their ability to pay for it.\textsuperscript{21} Clinical characteristics that were collected included corneal clarity and diameter, presence/absence of Haab’s striae, IOP, disc evaluation, gonioscopy findings, and type and number of preoperative topical medications.

We estimated the long-term success rates from the 1998 and 2008 cohorts of those patients who completed at least 10 years of follow-up at our center. We defined success a priori as (1) postoperative IOP ≤ 21 mm Hg with or without medications, (2) no additional surgery required to control IOP, and (3) no devastating or severe complication. Best-spectacle corrected visual acuity, postoperative IOP, and (3) no devastating or severe complication.

Statistical analysis was performed using MedCalc for Windows, version 12.5.0 (MedCalc Software, Ostend, Belgium). Participants’ demographic and clinical characteristics were summarized as mean and SD, median and interquartile range (IQR), and by counts and percentages as appropriate. The \( \chi^2 \) was used to compare proportions between groups, and 1-way analysis of variance was used to compare \( >2 \) groups. The comparison of continuous variables between the 2 groups was carried out using \( t \) test and Mann-Whitney test as appropriate. A \( P \)-value of \( <0.05 \) was considered statistically significant.

**RESULTS**

**Demographic and Clinical Characteristics**

One hundred seventy-two children (313 eyes) diagnosed with PCG in the 3 time periods of the study (1998, 2008, 2018) were identified. Of these, 48 (27.9\%) belonged to the first time period (1998), 76 (44.2\%) belonged to the second (2008), and 48 (27.9\%) belonged to the third time period (2018). Patient demographics are summarized in Table 1. Overall, bilateral affliction was common, and 53\% were male and 47\% were female with PCG. About one-half of the parents of children with PCG belonged to the lower socioeconomic strata in 1998 and 2008, as compared with 33.3\% in 2018. Significantly higher proportion of parents of children with PCG presented to our center from outside the states of Andhra Pradesh and Telangana for treatment in 2018 as compared with the other time periods, that is, 1998 (\( P = 0.0001 \)) and 2008 (\( P = 0.003 \)). The proportion of children with a history of parental consanguinity was significantly lower in 2018 as compared with those in 1998 (\( P = 0.001 \)) and in 2008 (\( P = 0.001 \)). There was no significant difference in the presenting and last follow-up IOP across the 3 time periods as determined (\( P > 0.05 \) both).

**Timeline**

Caregivers consistently recognized the eye problem within a week of the child’s birth in almost two-third of the cases across all the 3 time periods (Table 2). Of these,

| Year (Count, %) | 1998 (87 Eyes) | 2008 (140 Eyes) | 2018 (86 Eyes) |
|-----------------|---------------|-----------------|----------------|
| **No. patients**| 48 (27.9)     | 76 (44.2)       | 48 (27.9)      |
| **Laterality**  |               |                 |                |
| Unilateral      | 9 (18.8)      | 12 (15.8)       | 10 (20.8)      |
| Right eye       | 2 (22.2)      | 7 (58.3)        | 5 (50)         |
| Left eye        | 7 (77.8)      | 5 (41.7)        | 5 (50)         |
| **Bilateral**   | 39 (81.2)     | 64 (84.2)       | 38 (79.2)      |
| **Sex**         |               |                 |                |
| Male            | 28 (58)       | 37 (26)         | 27 (56)        |
| Female          | 20 (42)       | 39 (51)         | 21 (44)        |
| **Parental consanguinity** | | | |
| Parental consanguinity | 17 (35)      | 25 (33)         | 3 (6)          |
| **IOP at presentation (mm Hg)** | | | |
| Mean ± SD       | 27.0 ± 7.41   | 26.8 ± 8.12     | 25.6 ± 7.72    |
| Range           | 1042          | 868             | 1250           |
| **IOP at last follow-up (mm Hg)** | | | |
| Mean ± SD       | 13.4 ± 3.79   | 15.1 ± 8.61     | 14.2 ± 5.42    |
| Range           | 1030          | 852             | 1126           |
| **Corneal clarity at presentation, eyes** | | | |
| Edema           | 60 (68.9)     | 105 (75)        | 54 (62.7)      |
| Haab’s striae   | 9 (10.3)      | 5 (3.6)         | 13 (15.1)      |
| No. of eyes on AGM at presentation | 33 (37.9)     | 53 (37.8)       | 46 (53.5)      |
| Location of residence* | | | |
| Outside Andhra Pradesh/Telangana | 12 (25) | 32 (42.1) | 37 (77.1) |

*Andhra Pradesh/Telangana represent 2 South Indian states and L V Prasad Eye Institute is located in Telangana. Telangana was a part of the undivided Andhra Pradesh until 2014. AGM indicates antiglaucoma medication; IOP, intraocular pressure.
mothers were the first person to observe the initial symptoms in one-half of the children with PCG. The eye problems most commonly observed by the caregivers and parents included whitish appearance of the cornea and an inability to open the eyes in daylight (Table 1). Overall, the median age at the time of recognition of first symptoms was 1 day of age of the child (range, 1 to 7300 d; IQR, 1 to 30) (Table 3). It should be noted that the group included 1 adult, aged 20 years (7300 d) who self-presented due to decreased vision in 1 eye. There was no significant difference in the median age at the time of recognition of first symptoms by the caregivers across the 3 time periods (P > 0.05) (Table 3). Also there was no statistically significant difference in the age of the children at which symptoms were recognized by the caregivers or parents between the unilateral (median, 3 d) and bilateral PCG groups (median, 1 d) (P = 0.33).

Of the 172 children, only 23 (13.4%) children presented to our hospital within a week of recognition of the eye problem by the caregivers, and 40.1% of the children presented within 3 months of recognition (Table 2). However all of them underwent glaucoma surgery at our center within a week of their diagnosis. The component time periods contributing to the delay in presentation are provided in Table 2. The median age at the time of diagnosis at our center was 120 days (range, 1 to 8030 d; IQR, 30 to 378). It should be noted that the group included 1 adult, aged 22 years (8030 d) who self-presented and was diagnosed to have bilateral late-recognized PCG. There was no statistically significant difference in the median age of the children at the time of diagnosis between the 3 time periods (P = 0.23) (Table 3).

Across all the time periods, the consultations with local ophthalmologists took place within 1 week of symptom recognition by the caregivers/parents (Table 4). Despite this, there was a large delay from the day the caregivers or parents recognized the eye problem of the child to the day when they brought their children to our center for management across all the 3 time periods (Tables 2, 3; Fig. 1). The presentation to our hospital was delayed by 1 week to 3 months in 40.1% of the children with PCG, and by > 3 months in 46.5% of the children. However, there was no statistically significant difference in the duration of delay between the 3 time periods (P = 0.13).

In the 1998 cohort of PCG patients (n = 48), 19 (39.5%) completed at least 10 years of follow-up. The mean (SD) IOP at last follow-up was 15.36 mm Hg (4.68) and ranged from 10 to 30 mm Hg in the treated eyes. The median best-spectacle corrected visual acuity (logMAR) was 0.30 (Snellen equivalent, 20/40) and ranged from 0 to 3.0 (Snellen equivalent, 20/20 to no light perception) in the treated eyes. Success rate was 90.9% over a mean (SD) follow-up of 15.63 (4.25) years. Five patients underwent repeat glaucoma surgery; 4 underwent trabeculectomy with mitomycin-C, and 1 patient underwent transcleral cyclophotocoagulation.

In the 2008 cohort of PCG patients (n = 76), 26 (34.2%) completed at least 10 years of follow-up. The mean (SD) IOP at last follow-up was 16.33 mm Hg (4.96) and ranged from 10 to 30 mm Hg in the treated eyes. The median best-spectacle corrected visual acuity (logMAR) was 0.18 (Snellen equivalent, 20/30) and ranged from 0 to 3.0 (Snellen equivalent, 20/20 to no light perception) in the treated eyes. Success rate was 86.9% over a mean (SD) follow-up of 11.31 (2.79) years. Six patients underwent repeat glaucoma surgery; 5 underwent trabeculectomy with mitomycin-C, and 1 patient underwent transcleral cyclophotocoagulation.

**DISCUSSION**

In this study, almost two-third of the caregivers or parents recognized the classical features of PCG such as corneal edema and photophobia within the first week after birth of their child, regardless of the time period studied.

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### TABLE 2. Distribution of 172 Children With Primary Congenital Glaucoma According to Age at Time of First Symptoms, Age at Time of Diagnosis, and Overview of Delay Time From First Symptoms to Diagnosis

| Year | n | <1 wk | 1 wk-3 mo | >3 mo | <1 wk | 1 wk-3 mo | >3 mo | <1 wk | 1 wk-3 mo | >3 mo |
|------|---|-------|----------|-------|-------|----------|-------|-------|----------|-------|
| 1998 | 48 | 32 (66.7) | 11 (22.9) | 5 (10.4) | 0 | 20 (41.7) | 28 (58.3) | 2 | 4 (2) | 21 (43.7) | 25 (52.1) |
| 2008 | 76 | 48 (63.1) | 17 (22.4) | 11 (14.5) | 12 (15.8) | 27 (35.5) | 37 (48.7) | 15 | 9 (7.9) | 29 (38.1) | 32 (42.1) |
| 2018 | 48 | 30 (62.5) | 6 (12.5) | 12 (25) | 4 (8.3) | 17 (35.4) | 27 (56.2) | 6 | 12.5) | 19 (39.6) | 23 (47.9) |
| Total | 172 | 110 (63.9) | 34 (19.8) | 28 (16.3) | 16 (9.3) | 64 (37.2) | 92 (53.5) | 23 | 13.4 | 69 (40.1) | 80 (46.5) |

*Age of the child when caregivers first noticed symptoms.
LVPEI indicates L V Prasad Eye Institute (tertiary eye care centre; see text for details).
However, <10% of them presented without any delay to our hospital. Our results are in accordance with a previous study from Denmark consisting of 118 PCG patients identified over 4 decades (1977 to 2016).22 Overall, in our study, we did not observe any sex predilection for PCG. However, higher risk of PCG among male sex has been reported from other regions of the world.22–24

Parental concern has been reported to be most common route for seeking consultation with the ophthalmologist in cases of children with PCG.8,22 In our study, parental concern (mostly mother) was significantly higher (100%) in the recent year (2018) as compared with the previous time periods (56.2% in 1998 and 51.3% in 2008). One important reason for the early recognition by the caregivers and parents could be attributed to the severe disease phenotype associated with obvious corneal clouding and other presenting symptoms. Furthermore, higher rate of parental consanguinity noted in our study population has been reported to be associated with severe disease phenotype.10,25

The majority of the caregivers/parents in our study (>80%) sought prior consultation with an ophthalmologist across all the time periods before presenting to our hospital. Despite such high prior consultation rates, there was unnecessary delay of >3 months in 46.5% of the children with PCG presenting to our hospital across all the 3 time periods. Several reasons have been cited as barriers to effective management of PCG in developing countries such as India.7,26–31 These barriers include delayed presentation, severe phenotype, limited access to health care, long travel distances, poor socioeconomic status, and cultural fear of surgical treatment in a young infant.26 Given the retrospective nature of our study, we did not have data regarding health care access, exact travel distances, and any cultural issues, etc. However, review of our database revealed that a relatively higher proportion (about one-half) of the parents of children with PCG belonged to the lower socioeconomic strata in 1998 and 2008, as compared to one-third (33.3%) in 2018. In this group of caregivers/parents, it is plausible that reasons such as the cost of travel to the clinic, follow-up cost, and the wage-earning hours of the parents may have contributed significantly to the delay in seeking care at our tertiary eye care center. Also, sex inequalities within the household still prevent many mothers to bring their child to an eye care professional for examination or to a hospital for surgery in a timely manner. In many developing countries, women have low literacy rates and little or no financial independence; they often need permission and assistance from their husbands, male relatives or community members to take their children to the hospital. In addition, hospitalization and surgical procedures in a very young child in general instil fear and anxiety among parents and this could be could be reduced if health care professionals at different levels could transmit clear information about benefits of early glaucoma surgery.

We observed that a significantly higher proportion of parents of children with PCG presented to our hospital from outside the state of Telangana (our hospital is located in this state) and its neighbouring state of Andhra Pradesh for treatment in 2018 as compared with the other time periods. In the present study, we did not find laterality (unilateral vs. bilateral) to be associated with diagnostic delay. Although caregivers/parents in both these groups recognized the symptoms of PCG early enough in their children, they presented late to our hospital perhaps due to the reasons listed earlier. However, there was no delay in glaucoma surgery in either of these groups and simultaneous bilateral surgery was performed in bilateral affliction. We have previously demonstrated the several advantages offered by simultaneous bilateral surgery in bilateral PCG.20

Interestingly, 40.1% of the children with PCG were brought by the caregivers/parents to our hospital between 1 week and 3 months of recognition of symptoms, and underwent surgery within a week of diagnosis. This perhaps indicates that these children did not have significant delay in accessing our services, thereby, improving their chances of better visual and surgical outcomes. In our previous study, we investigated the surgical outcomes of 157 children with PCG (299 eyes) who underwent glaucoma surgery within 6 months of birth between 1990 and 2001 at our hospital.19 We found good IOP control and 40.8% of the children achieved normal visual acuity (≥20/60) in the better eye over a mean follow-up of 26.5 ± 25.1 months.19

In the present study, overall 80 children (46.5%) with PCG had delayed presentation to our hospital by >3 months from recognition of symptoms by the caregivers/parents. This delay occurred despite prior consultation with the local ophthalmologist within 1 week of symptom recognition; however, there was no delay in the surgical intervention in these children at our center. Delayed presentation has been associated with poor IOP control and suboptimal visual outcomes. Nonetheless, we noted good long-term IOP control over a mean follow-up of 15.6 and 11.3 years in the 1998 and 2008 cohorts, respectively. The visual outcomes are

### TABLE 4. Distribution by Persons Who First Noticed the Symptoms of Primary Congenital Glaucoma

| Year | Suspicion by Family Member* | Prior Medical Consultation† |
|------|----------------------------|-----------------------------|
| 1998 | 27 (56.2)                  | 40 (83.3)                   |
| 2008 | 39 (51.3)                  | 62 (81.6)                   |
| 2018 | 48 (100)                   | 47 (97.9)                   |

*Family members include parents and immediate family members (caregivers such as grandparents, uncles, and aunts on paternal and maternal side).

†Prior medical consultation took place within 1 week of suspicion of symptoms by family member.

![FIGURE 1. Scatter plot showing the relationship between age (in days) at recognition of symptoms by the caregivers/parents of children with PCG and the delay in diagnosis (in days). Note that logarithmic scales have been used for both the axes.](image-url)
comparable to our previous study of 624 eyes of 360 children patients operated for PCG over a 15-year period.17 Furthermore, our visual outcomes compare favourably with those reported by Zagora et al18 from Australia. However, it is difficult to ascertain the impact of the delay on the surgical outcomes given that in 53.5% of the children the delay was limited to up to 3 months. In the remainder of the patients with a delay of > 3 months, the duration of delay was highly variable preventing any meaningful comparisons.

The surgical management of PCG remains a demanding task. In a developing country such as India, the availability of a limited number of trained personnel with appropriate skill sets (surgeon, anaesthesiologist, nursing staff, other professionals) for microsurgical treatment of PCG and postoperative care is a serious concern.26 This is supported by our finding that children with PCG were referred to our hospital (a tertiary eye care center) by local ophthalmologists in 1998, and the pattern remains unaltered even after 3 decades. This shortage of trained providers along with the necessary infrastructure can be overcome by training adequate numbers of pediatric glaucoma surgeons who not only will diagnose the condition, but who will also be competent surgically, and be able to follow-up the children with PCG for life.33

Although there were strengths to this study, for example, recruitment at a single-site (large tertiary care center with a dedicated pediatric eye care facility) and a single pediatric glaucoma specialist with experience in the diagnosis and care of childhood glaucoma caring for all patients over 3 decades, this study does carry some limitations given its retrospective design. There is a risk of information bias as the data were obtained solely from medical and electronic records. Consequently, we could not gather information from the caregivers and parents regarding various logistical issues, attitudinal considerations, values, beliefs, and cultural barriers that they may have faced. These factors negatively impact the decision of caregivers and parents to seek professional help immediately once symptoms have been detected.

In conclusion, this is the first single-site, single surgeon study from India to our knowledge that has extensively studied the trends in presentation of PCG cases to a tertiary eye care center. Our findings demonstrate that there was no sex predilection for PCG. Bilateral affliction, and more severe disease are the characteristic features of PCG in India. Although disease recognition occurs early by the caregivers and parents of children with PCG followed by guidance and referral by local ophthalmologists, delay in presentation to tertiary eye care center remains a significant problem in India. Bridging strategies that would link communities to hospital need to be developed so as to reach parents and caregivers of children with PCG living in remote areas of the country, who cannot come to the hospital because of poor socioeconomic conditions of their households and the need to commute long distances. Educational efforts to increase the knowledge of the caregivers about the need for early intervention for PCG should also be warranted to ensure the early presentation of these children for glaucoma surgery. These needs are great in all societies but perhaps greatest in the developing countries such as India.

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