Long-term Outcomes of Surgical Treatment of Unilateral Primary Congenital and Infantile Glaucoma

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SUMMARY

Aim: To evaluate the long-term outcomes of surgical treatment in unilateral primary congenital glaucoma (PCG), with a focus on stabilizing intraocular pressure (IOP) and structural and functional parameters of the affected eye.

Material and Methods: A retrospective analysis was performed on 20 eyes of 20 patients with unilateral PCG operated on between 2002–2022 by combined trabeculotomy-trabeculectomy (CTT) or trabeculectomy (TBE) with/without MMC at the Department of Pediatric Ophthalmology, University Hospital Brno. The evaluated parameters included age at the time of diagnosis and surgery, type and number of procedures, pre- and postoperative IOP, presence of buphthalmos and Haab's striae, corneal diameter, cup-to-disc ratio (C/D), spherical equivalent (SE), best-corrected visual acuity (BCVA), use of medication, visual field findings, and retinal nerve fiber layer (RNFL) thickness on OCT. Surgical success was defined as IOP ≤ 21 mmHg without the need for further glaucoma surgery.

Results: Mean age at the time of diagnosis was 7.9 months, with 85% diagnosed before 6 months; mean period of follow-up monitoring was 12.3 years. CTT was performed in 55% and TBE in 45% of patients, with mean age at the time of surgery of 10 months. All eyes presented with buphthalmos and Haab's striae; mean corneal diameter was 13.1 mm. The C/D ratio decreased from 0.53 to 0.26 ($p = 0.002$) and IOP from 30.1 to 14.7 mmHg ($p < 0.0001$). At the final follow-up examination, 85% of patients required no topical medication (mean 0.7; range 0–4). One additional procedure was needed in 10% of cases, with an overall primary surgical success rate of 90%. Mild to moderate myopia was the most common refractive outcome (65%). Mean BCVA was 0.6, with ≥ 0.5 achieved in 65% of patients. Mean RNFL thickness was 82.9 µm; significant thinning below 80 µm with corresponding visual field defects was observed in 3 eyes.

Conclusion: With early diagnosis and appropriate surgical treatment, unilateral PCG has a favorable long-term prognosis. CTT and TBE provide stable IOP control, partial reversal of glaucomatous cupping, and good visual function. Refractive correction, amblyopia prevention, and careful monitoring of the other eye are essential

Key words: unilateral primary congenital glaucoma, combined trabeculotomy with trabeculectomy (CTT), reversal of glaucomatous cupping

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INTRODUCTION

Primary congenital glaucoma (PCG) is a rare but serious ocular pathology in childhood, involving defective development of the structures of the intraocular fluid drainage system, trabeculodysgenesis, and a subsequent increase of intraocular pressure (IOP). In most cases the disease is manifested bilaterally [1–5], while unilateral forms are less common and represent only 15–35% of all patients with PCG [6]. Untreated or late diagnosed PCG leads to the development of structural and subsequently functional changes to the affected eye. This typically involves a reduction of corneal transparency, the presence of Haab's striae (double thin lines of cracks in the Descemet's membrane of the cornea), increase of corneal diameter (≥ 12 mm during the first year of life), progressive enlargement of the globe (buphthalmos or hydrophthalmos), elongation of the axial length of the globe, occurrence of glaucomatous cupping of the disc of the optic nerve (ON) and subsequent functional damage to vision. According to the age of manifestation, congenital glaucoma is classified as primary congenital (up to 3 months of age) and infantile glaucoma (up to 3 years of age) [7,8].

The foundation of PCG management is surgical treatment. The most frequently used techniques include trabeculotomy, trabeculectomy and a combination thereof – combined trabeculotomy with trabeculectomy (CTT). The aim of the primary surgical procedure is to attain stable, low intraocular pressure with a minimum requirement for supplementary medication, minimization of complications and preservation of good visual functions over the course of long-term monitoring. Topical

antiglaucoma medication usually plays a complementary role in PCG, and is used primarily before planned surgery and/or postoperatively in the case of insufficient long-term compensation [1–4,8]. Unilateral cases of PCG are rare and are more frequently distinguished by asymmetrical differentiation of eye growth, which increases the risk of anisometropia and secondary amblyopia [6]. These factors may fundamentally influence the long-term functional outcome of treatment. When evaluating the results, it is therefore essential to assess not only the success rate of reducing intraocular pressure, but also the structural and functional parameters of the affected eye, which determine the resulting visual functions. To date only a relatively small number of studies have been published focusing exclusively on unilateral forms of PCG [6].

The aim of this study is to assess the long-term results of surgical treatment of unilateral primary congenital glaucoma in patients treated at the Department of Pediatric Ophthalmology, University Hospital Brno in the period of 2002–2022. The retrospective analysis covers 20 eyes of 20 children with unilateral primary congenital and infantile glaucoma. It assesses the long-term compensation of intraocular pressure following the primary surgical procedure, as well as the structural and functional outcomes of the affected eye over the course of long-term observation.

MATERIAL AND METHODS

The retrospective analysis included 20 patients (17 of male sex, 3 of female sex) with unilateral primary congenital and infantile glaucoma, who were surgically treated at the Department of Pediatric Ophthalmology, University Hospital Brno in the period of 2002–2022. The development of glaucoma in the patient's other eye was not recorded in any of the patients during the period in question.

The evaluated parameters covered the type and number of surgical procedures, age at the time of diagnosis, age at the time of surgery, preoperative and final IOP, presence of buphthalmos and Haab's striae, preoperative corneal diameter, preoperative and final value of optic nerve disc cupping expressed as cup/disc (C/D) ratio, final refraction – spherical equivalent (SE), best corrected visual acuity (BCVA) at the end of observation, number of applications of topical antiglaucoma treatment at the last follow-up examination, findings on the perimeter, and optical coherence tomography (OCT) – retinal nerve fiber layer (RNFL) thickness in the affected eye.

Intraocular pressure was measured preoperatively and subsequently during the course of long-term observation. In uncooperative children the measurement was performed under general anesthesia. The instruments used included the tonometer TONO-PEN AVIA (Reichert) and/or Goldmann applanation tonometer. Corneal diameter was measured using a standardized caliper. The anterior segment was examined biomicroscopically. Optic nerve disc cupping (C/D ratio) was evaluated ophthalmoscopically with the use of an indirect ophthalmoscope (Omega 500 Heine) or with the aid of fundus photography (NIDEK Retina Scan Duo). Refraction was determined with the aid of a manual autorefractokeratometer (NIDEK ARK-30) and/or automatic autorefractokeratometer (NIDEK Tonoref III). Final BCVA was evaluated with the aid of a Snellen chart (images, Pflüger hooks or standard type) in a decimal value. A quantitative evaluation of the RNFL was performed with the aid of the OCT instrument Cirrus HD-OCT (Carl Zeiss). The XLSTAT program (Addinsoft, France) was used for the statistical data processing. The preoperative and final values of IOP and C/D ratio were compared with the aid of a paired Student t-test with a level of significance of $p < 0.05$. The success rate of the primary surgical procedure was defined as IOP compensation of ≤ 21 mmHg or without the need for antiglaucoma medication and without the need for further antiglaucoma surgery at the end of the observation period.

In most cases we chose a uniform procedure of combined trabeculotomy with trabeculectomy (CTT) or trabeculectomy (TBE) alone, with or without the use of mitomycin (MMC) to prevent scarring and to ensure the long-term through flow of the filtration opening. Following peritomy of the conjunctiva and preparation of a scleral flap, a radial incision was made in the middle of the scleral bed perpendicular to the corneal limbus, after clarification of the mouth of the sinus venosus right-sided and left-sided trabeculectomy probes were carefully inserted into the Schlemm's canal with corresponding curvature (modification according to Filouš), and subsequently rotated 90° into the anterior chamber, thereby causing ablation of the trabecular tissue within a scope of 120 to 180 degrees. This was followed by a standard trabeculectomy procedure with or without application of MMC, with basal iridectomy and suture of the scleral flap and conjunctiva (Figures 1, 2, 3).

RESULTS

The cohort incorporated 20 eyes of 20 pediatric patients, of whom 17 were boys (85%) and 3 were girls (15%). Mean age at the time of diagnosis was 7.9 months (range 1–35 months). In 17 patients (85%) glaucoma was diagnosed before 6 months of age. The mean period of monitoring was 12.3 years (range 3–21 years). Eleven children (55%) underwent combined trabeculotomy with trabeculectomy (CTT), the remaining nine patients (45%) underwent trabeculectomy (TBE), in one case TBE was supplemented with the application of mitomycin C (MMC). Mean age at the time of surgery was 10 months (range 2–40 months). Mean preoperative corneal diameter in the affected eye was 13.1 mm (range 12–14 mm). Buphthalmos and Haab's striae of the cornea were present in all the affected eyes. The demographic data, preoperative and clinical characteristics in the observed cohort are summarized in Table 1.

Mean preoperative size of optic nerve disc cupping (C/D ratio) was 0.53 (range 0.2–0.9). A partial rever-

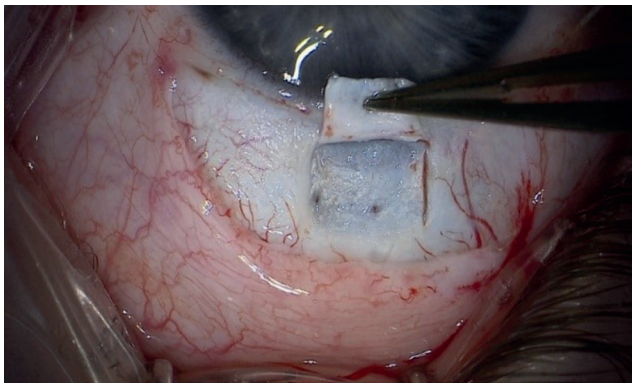


Figure1. Scleral flap during CTT or TBE

CTT – Combined Trabeculotomy and Trabeculectomy, TBE – Trabeculectomy

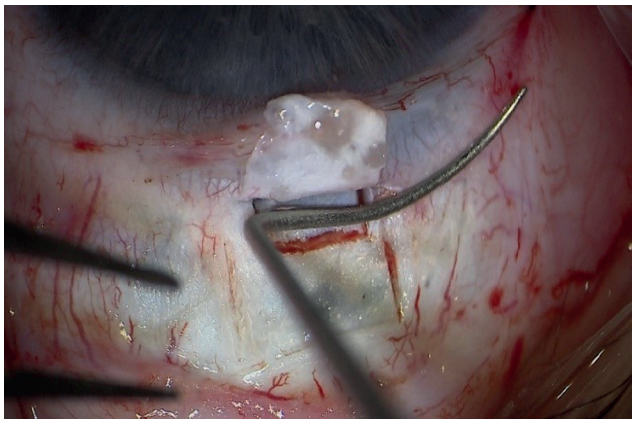


Figure 2. Trabeculotomy probe prepared for insertion into Schlemm's canal during CCT

CTT – Combined Trabeculotomy and Trabeculectomy

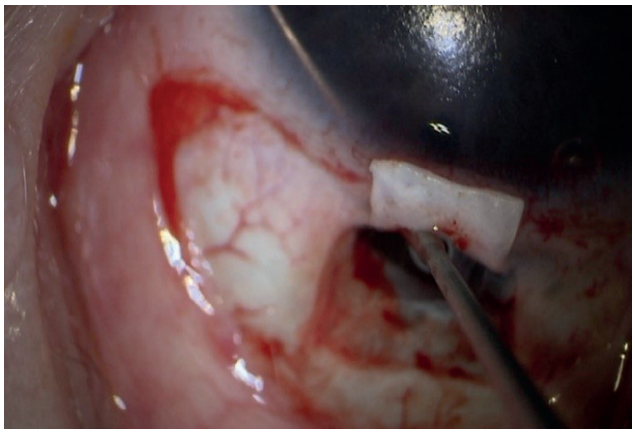


Figure 3. Introduction of the trabeculotomy probe into Schlemm's canal during CTT

CTT – Combined Trabeculotomy and Trabeculectomy

sal of glaucomatous cupping was achieved postoperatively, with a mean final C/D ratio of 0.26 (0.1–0.7) (p = 0,002), as illustrated in Table 2. Graph 1 shows a manifest decrease of C/D values following surgical treatment. The median preoperative C/D was 0.6, while after surgery this was reduced to the value of 0.3. This shift indicates partial reversibility of glaucomatous

Table 1. Demographic data and preoperative and intraoperative clinical characteristics of the monitored patients with unilateral congenital glaucoma (n = 20)

Gender n (%) Male Female	 17 3	 85 % 15 %
Affected eye n (%) Right Left	 7 13	 35 % 65 %
Mean age at diagnosis (months)	7.9 ±9.4	(range 1–35)
Mean age at surgery (months)	9.9 ±11.2	(range 2–40)
Reoperative corneal diameter (mm)	13.1 ±0.5	(range 12–14)
Preoperative presence of buphthalmos and Haab's striae, n (%)	20	100 %
Mean preoperative intraocular pressure (mm Hg)	30.1 ±4.4	(range 22–40)
Mean cup-to-disc ratio (optic disc cupping)	0.53 ±0.20	(range 0.2–0.9)
Mean preoperative number of topical antiglaucoma medications	2	(rozmezí 1–3)
Type of surgery number of eyes, n (%) CTT TBE TBE + Mitomycin C	 11 8 1	 55 % 40 % 5 %
Mean follow-up duration (years)	12.3 ±5.9	(range 3–21.5)
Mean age at the end of follow-up (years)	12.4 ±6.2	(range 3.5–22)

CTT – combined trabeculotomy and trabeculectomy, TBE – trabeculectomy ONH – optic nerve head, C/D – cup-to-disc ratio

cupping of the optic nerve disc in children following effective reduction of intraocular pressure.

Mean preoperative intraocular pressure (IOP) was 30.1 ±4.4 mmHg, whereas the final mean IOP after surgery was reduced to the value of 14.7 ±3.0 mmHg ($p < 0.0001$). (Table 2) Graph 2 documents a pronounced reduction of IOP following surgical treatment. The median preoperative IOP was 30 mmHg (range 25–40), whereas postoperatively its final value was reduced to 15 mmHg (range 10–20). The dispersion of postoperative values is markedly lower, which indicates the stabilization and homogenization of IOP postoperatively in the long-term observation period. The results confirm both a statistically and clinically significant reduction of IOP following surgical intervention (Table 2).

The mean number of applications of adjuvant topical antiglaucoma therapy at the end of the observation period was 0.7 (range 0–4). At the end of the observation period 15 patients (85%) no longer required topical anti-

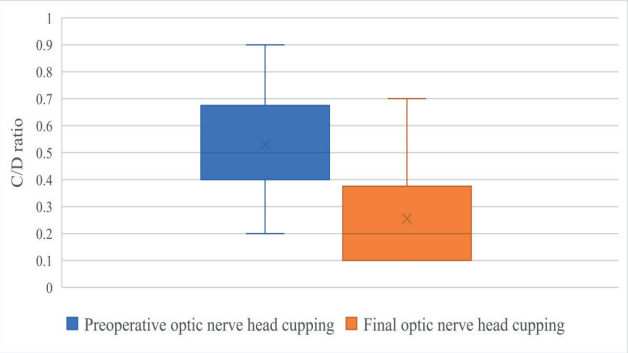
glaucoma therapy. Only two patients (10%) required one additional antiglaucoma operation during the long-term observation period. Success of the primary surgical procedure, defined as IOP compensation of ≤ 21 mmHg with our without the need for antiglaucoma medication and without the need for further antiglaucoma surgical intervention at the end of the observation period, was achieved in 90% of eyes.

Final refraction, expressed as spherical equivalent (SE) in the affected eye was most often within the band of mild to medium myopia in 13 eyes (65%) at the end of the observation period. Four eyes (20%) manifested severe myopia and three eyes (15%) had hypermetropic refraction (Graph 3). One patient underwent scleroplasty and a laser refractive procedure for severe myopic anisometropia, and one patient underwent surgery for divergent strabismus. All the children received pleoptic and orthoptic care at our clinic during the period of development of visual functions. Graph 3 clearly illustrates the distribution of final refraction (SE) in the observed eyes of the cohort. In most eyes myopic refraction predominated. The majority of the patients (n = 13) had values within the band of medium to mild myopia. This result corresponds to the trend in which congenital glaucoma frequently leads to myopic shift of refraction.

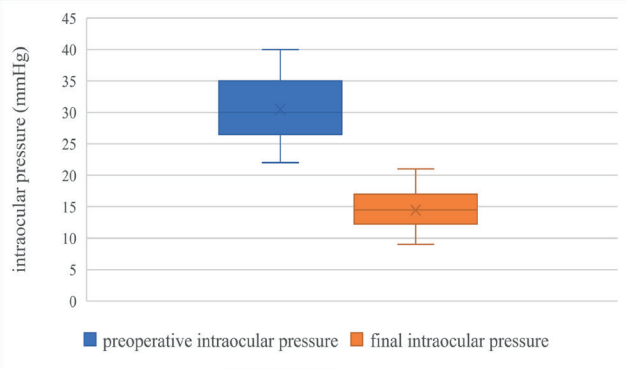
Mean best corrected visual acuity (BCVA) in the affected eye at the end of the observation period was 0.6 decimally (range 1.0–0.01). Thirteen patients (65%) attained final BCVA of 0.5 or better at the end of the observation period. Five patients (25%) had final BCVA within the range of 0.4 to 0.1, and only two patients manifested final BCVA of worse than 0.1, as summarized in Table 3.

An examination of the visual field (VF) and RNFL thickness was performed on 12 patients (60%) at the end of the observation period with the aid of optical coherence tomography (OCT). The measured RNFL values in 12 eyes reflect the long-term structural results of unilateral primary glaucoma. Mean RNFL thickness was 82.9 um, which is an only slightly lower value than the referential range for the healthy population (90–110 µm) depending on the used OCT method and patient age. A more significant reduction of RNFL beneath 80 µm with correlating blind spots in the VF was recorded in only 3 of these 12 eyes (25%).

The detailed mean RNFL values, with distribution in the individual quadrants is summarized in Table 4. The highest mean values were determined in the superior



Graph 1. Preoperative and final optic nerve head cupping (C/D ratio) in the monitored group of 20 eyes of 20 patients with unilateral congenital glaucoma

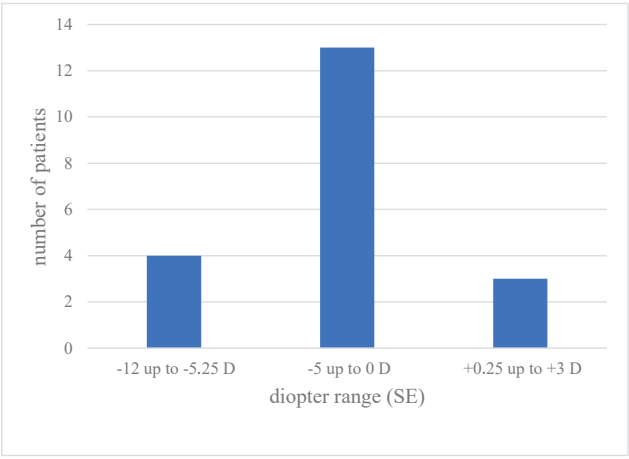


Graph 2. Preoperative and final intraocular pressure in the monitored group of 20 eyes of 20 patients with unilateral congenital glaucoma

Table 2. Comparison of the mean initial and final intraocular pressure values and optic nerve head cupping in the monitored group (n = 20)

	Initial IOP before surgery mean ±SD	Initial preoperative IOP mean ±SD	p-value *
Intraocular pressure (mmHg)	30.1 ±4.4 (range 22–40)	14.7 ±3.0 (range 9–21)	< 0.0001
Optic disc cupping, C/D	0.53 ±0.20 (range 0.2–0.9)	0.26 ±0.18 (range 0.1–0.7)	0.002

**– Paired Student' t-test, ONH – optic nerve head, IOP – intraocular pressure, C/D – cup/disc ratio, SD – standard deviation*



Graph 3. Distribution of final refraction, spherical equivalent (SE) in the affected eye in the monitored group

Table 3. Final best-corrected visual acuity (BCVA) of the affected eye in the group of monitored patients (n = 20)

BCVA* (snellen, decimal)	Number of eyes	%
< 0.1	2	10
0.1	3	15
0.4–0.2	2	10
0.9–0.5	8	40
1.0	5	25

Table 4. Mean retinal nerve fiber layer (RNFL) thickness values at the end of the follow-up period by individual quadrants, measured by OCT in 12 eyes in the monitored group of patients with unilateral congenital glaucoma

RNFL	Average ±SD; µm
Average value (ave)	82.9 ±9.0 (range 68–102)
Superior quadrant (S)	99.8 ±8.9 (range 63–134)
Inferior quadrant (I)	97.5 ±7.6 (range 82–120)
Temporal quadrant (T)	66.7 ±7.2 (range 57–73)
Nasal quadrant (N)	66.1 ±8.0 (range 54–79)

SD – standard deviation

quadrant 99of .8 ±8.9 µm, whereas the lowest values were in the nasal quadrant of 66.1 ±8.1 µm. The topographic distribution corresponds to the physiological profile of the RNFL with predominance of the superior and inferior quadrant, with slight reduction of the absolute values across all quadrants. This image may reflect a slight diffuse atrophy of axons of the retinal ganglion cells, as a consequence of glaucoma suffered at an early age. However, in the majority of the patients these structural changes were not projected into functional affliction of sight, and 75% of these 12 evaluated eyes manifested a physiological finding on the perimeter. In this study the RNFL values were not correlated with the contralateral healthy eye.

DISCUSSION

In our cohort of 20 patients with unilateral PCG, the mean length of follow-up observation was 12.3 years. Mean intraocular pressure was reduced from 30.5 mmHg to 14.5 mmHg at the end of the observation period, which represents a reduction of more than 50%. At the same time a partial reversal of optic nerve disc cupping (C/D ratio) was achieved, from 0.53 to 0.26. These results confirm the long-term effectiveness of surgical treatment also in the case of unilateral PCG. Our results show that both a combined technique (CTT) and trabeculectomy (TBE) alone produce long-term favorable results in the compensation of IOP and morphology of the optic nerve disc. Combined procedures are indicated in the literature as more appropriate for early manifesting or advanced forms, in which faster compensation takes place, whereas TBE provides comparable results in later manifestations. Our results support the conclusions of the studies conducted by Mandal and Sood et al. [2,3], i.e. that the effectiveness of both methods is comparable over the long term, while the experience of the surgeon and postoperative care are of fundamental importance.

Reversal of glaucomatous cupping after attaining compensation of intraocular pressure is a common phenomenon in pediatric patients. In younger children this process is more pronounced thanks to the greater plasticity of the lamina cribrosa, which enables partial restoration of the anatomy of the optic nerve disc after reduction of IOP [9,10]. Our observed decrease of the C/D ratio by 0.27 corresponds to the data from the literature, where Wu et al. [9] described a significant reduction of cupping after TBE in the majority of children with PCG [9]. Meirelles et al. [1] confirmed a similar effect especially in patients operated on before reaching 1 year of age.

In our cohort glaucoma did not develop in the other eye of any of the patients. However, according to Majumar et al. [6], glaucoma may develop also in the contralateral eye within the first 5 years of observation in a certain proportion of children with originally unilateral affliction. According to some authors, the other eye therefore cannot be considered entirely healthy, but rather as potentially manifesting forme fruste glaucoma. As a result it is essential to ensure long-term monitoring of both eyes [5,6].

The observations we have to date indicated that primary congenital glaucoma associated with mutations of genes CYP1B1 and LTBP2 is manifested predominantly bilaterally [11–13], whereas no unequivocal genetic association has yet been confirmed in unilateral cases [14]. None of our patients underwent a genetic test.

A peculiar feature of unilateral forms of PCG is an increased risk of anisometropia and amblyopia, which was manifested in practically all the patients in our cohort. Clinical practice must incorporate not only surgical treatment, but also systematic refractive and orthoptic care. A patient in whom correction of a refractive error and pleoptic treatment were not commenced in a timely manner scored the worst BCVA values, which underscores the importance of timely comprehensive rehabilitation of visual functions [4].

A limitation of our study is its retrospective character and the relatively small number of patients. Nevertheless, our results confirm that timely surgical treatment leads to long-term and stable compensation of intraocular pressure and to partial reversal of structural changes while preserving good functional results in long-term monitoring. An interesting subject for future examination would be a structural analysis of the contralateral healthy eye in cases of unilateral primary congenital glaucoma.

CONCLUSION

In the case of timely diagnosis and an adequate surgical solution, unilateral primary congenital and infantile glaucoma have a favorable long-term prognosis. The primary surgical procedure is effective over the long term

in as many as 90% of cases. Combined trabeculotomy with trabeculectomy, as well as trabeculectomy alone, are effective surgical methods which lead to long-term compensation of intraocular pressure and partial reversal of glaucomatous cupping of the optic nerve disc. In long-term observation, in addition to monitoring the compensation of intraocular pressure, it is important to place emphasis on monitoring the development and correction of refractive error, anisometropia and amblyopia. With the assistance of pleoptic and orthoptic care, it is possible to achieve very good long-term functional results in children with unilateral primary congenital glaucoma.

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