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Innovative Strategies in Managing Early-Onset Primary Congenital Glaucoma

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Abstract

Congenital glaucoma has the potential to damage the visual nerve fibers, leading to permanent blindness. This case report aims to provide timely and appropriate management upon diagnosis. The case involves a 5-month-old baby girl delivered via spontaneous normal delivery at 36-37 weeks, with a birth weight of 3100 grams, who presented with complaints of left eye enlargement since one month of age, persistent watery eyes, and photophobia when exposed to sunlight. Diagnoses included Primary Congenital Glaucoma, Congenital Cataract, and Exotropia Oculi Sinistra. The patient received preoperative medication, underwent trabeculotomy-trabeculectomy surgery, and had CRS screening and Rubella serological examination. Early detection and operative intervention to regulate intraocular pressure can preserve optimal visual function throughout adulthood.

1. Introduction

Primary Congenital Glaucoma (PCG) is a condition that has the potential to induce harm to the optic nerve fibers, ultimately leading to irreversible blindness [1]. Primary congenital glaucoma is a rare disorder that impacts infants and young children [2, 3]. Its prevalence is notably high in certain populations around the world, particularly in areas where inbreeding and consanguinity are common [4]. The prevalence of pediatric glaucoma accounts for approximately 4.2% of all cases of childhood blindness [2]. It typically manifests bilaterally, with males constituting 65% of the affected population [5]. Nearly all instances are sporadic, although 10% exhibit a hereditary component, commonly following an autosomal recessive pattern with varying modes of inheritance [4].

PCG may present with symptoms in infancy or early childhood, such as excessive tearing, sensitivity to light, and involuntary eye muscle spasms [5]. In addition to increased IOP (Intra Ocular Pressure), common ocular features include corneal cloudiness, Haab's striae, and enlarged corneal diameter. Generally, patients with glaucoma necessitate ongoing treatment, management, and evaluation to prevent blindness [2, 5, 6]. Glaucoma-induced blindness is irreversible but can be averted through early detection, although patients often realize it too late. In chronic glaucoma, blindness progresses gradually, often escaping the patient's notice [7]. In contrast, prompt action is crucial in acute glaucoma to administer appropriate therapy and prevent blindness [8].

Children's eyes differ from those of adults [7]. In adults, glaucoma is diagnosed based on optic nerve pathology, whereas in children, it is characterized by intraocular pressure-related damage to the entire eye [5, 9].

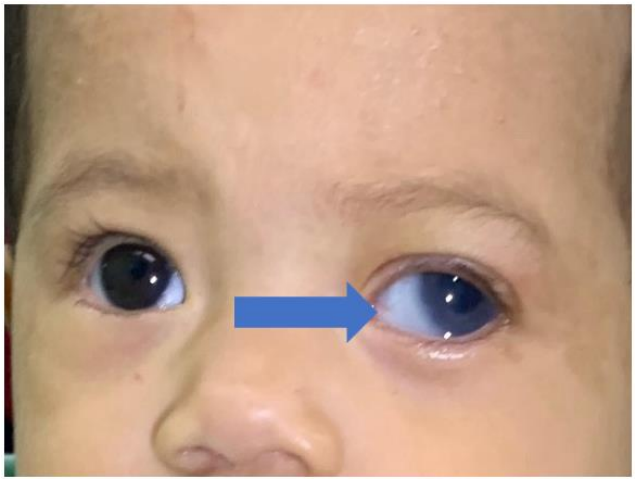

Figure 1. Diagnosis of Primary Congenital Glaucoma of the Oculi Sinistra and Exotropia of the Oculi Sinistra.

Table 1. Ophthalmology examination.

Oculi Dextra	Oculi Sinistra
• Visus: Fixed and follow the object	• Visus: Fixed and follow the object
• Palpation IOP: N	• Palpation IOP: N+
• Palpebra: Calm	• Palpebra: Spasms
• Conjunctiva: Quiet	• Conjunctiva: Quiet
• Cornea: Clear (10mm)	• Cornea: Macrocornea (12mm), Hazy
• Anterior chamber: Normal	• Anterior chamber: In
• Pupils: Round, 2-3 mm, Light reflex (+)	• Pupils: Round, 5-6 mm, Light reflex (-)
• Lens: Clear	• Lens: Clear

Therefore, this case report aims to provide timely and adequate management according to the diagnosis and comprehensive management steps for primary congenital glaucoma, including optical rehabilitation after surgery. Trabeculotomy – Trabeculectomy. This approach resulted in improved vision and was safe for children or infants.

2. Cases

A 5-month-old female, Acehnese, on 2 February 2024, was brought by her parents to the pediatric ophthalmology department of General Hospital Dr. Zainoel Abidin with chief complaints of an enlarged left eye since one month of age (Figure 1). The patient's eye also exhibited constant watering and sensitivity to sunlight. The baby was born with spontaneous normal delivery at 36-37 weeks, with a birth weight of 3100 grams. Throughout the gestational period, the patient's mother abstained from all medication except for the vitamins prescribed by her obstetrician. The patient's prenatal course was unremarkable, and there was no evidence of any infections during the pregnancy. The mother did not report experiencing fever or a reddish skin rash. According to the patient's family, there was no history of the child's eyes exhibiting a cat's-eye glow.

The patient had no history of NICU admission. She received complete immunization and was exclusively breastfed. The patient was irritable and presented with photophobia and blepharospasm. Tears were coming out of her left eye. Furthermore, there was no reported history of trauma, familial occurrence of the same disease, or any systemic illnesses. The patient's overall condition was good. Table 1 describes the visual acuity test of the patient.

The patient was diagnosed with Primary Congenital Glaucoma, Congenital Cataract, and Left Ocular Exotropia. Preoperative medicinal therapy was administered, including 0.25% Beta blocker eye drops two times daily, one drop in the left eye, and Artificial tears four times daily, one drop in the left eye. 5 A laboratory examination, chest X-ray, consultation with pediatrics, cardiology, and anesthesia were undertaken to acquire approval for surgery under general anesthesia. CRS screening, Otorhinolaryngology consultation, and Rubella serological examination were also performed. According to the American Society of Anesthesiologists (ASA), the patient had pre-anesthesia physical state II. Combined Trabeculotomy - Trabeculectomy with general anaesthesia after giving informed consent to the patient's parents and obtaining approval from the anaesthesia department was carried out on 7 February 2024.

IOP measurement using the Perkins tonometer was conducted immediately after anesthesia. The intraocular pressure (IOP) in the right eye measured 19 mmHg, while in the left eye, it was 35 mmHg (Figure 2). corneal diameter measurement using calipers during an EUA (examination under anaesthetic). The corneal diameter of the right eye was 10.5 mm, whereas that of the left eye was 13 mm. The pupil diameter in the right eye measured 2.5 mm, while in the left eye, it was 5.5 mm. The posterior capsule of the lens appeared cloudy, with a diameter of 4 mm (Figure 3).

After measuring the IOP, pupil, and corneal diameter, we performed surgery on the patient using trabeculotomy—trabeculectomy (Figure 4) and the irrigated aspiration technique (Figure 5). During the surgery, we found obstruction of the trabecular meshwork and narrowness of Schlemm's canal. These intraoperative findings guided us in performing the Trabeculotomy—trabeculectomy procedure to reduce IOP and preserve visual function (Figure 6).

Postoperative medical management included oral antibiotics administered three times daily at a dosage of 0.5 ml, oral antipyretic analgesics administered three

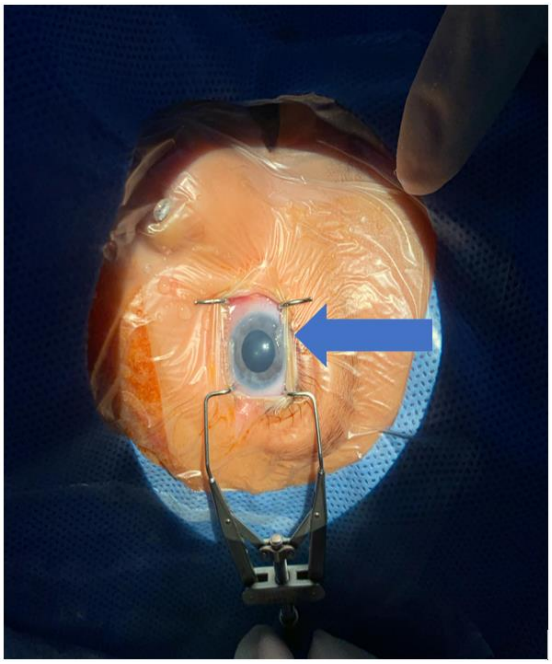

Figure 2. IOP OD 19 mmHg, OS 35 mmHg (Perkins tonometer).

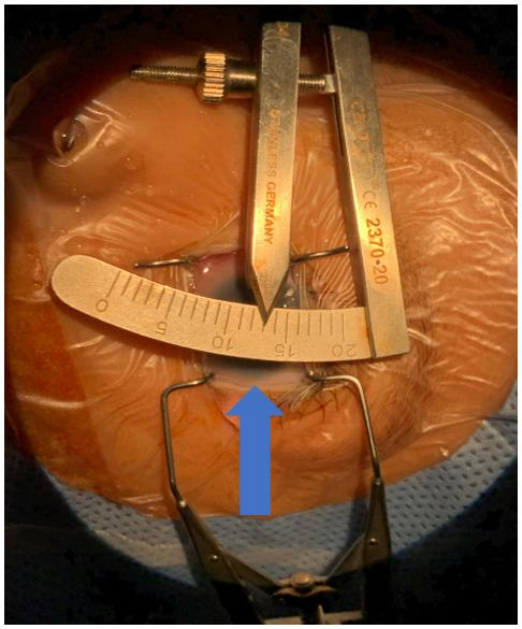

Figure 3. Cornea OD 10.5 cm OS 13 mm, Pupil OD 2.5 mm, OS 5.5 mm, Posterior capsule of lens cloudy (4 mm).

times daily at a dosage of 0.7 ml, topical antibiotics applied eight times daily to the left eye, and topical anti-inflammatory medication applied eight times daily to the left eye. The patient was discharged after no complaints on the second postoperative day. Furthermore, the patient is scheduled for outpatient care at the Department of Pediatric Ophthalmology and Strabismus in RSUD dr. Zainoel Abidin Banda Aceh within a week and one month after surgery. After one month, the patient's corneal diameter looked normal in both eyes, with no complications such as infection, increased eye redness, pain, vision changes, or excessive tearing.

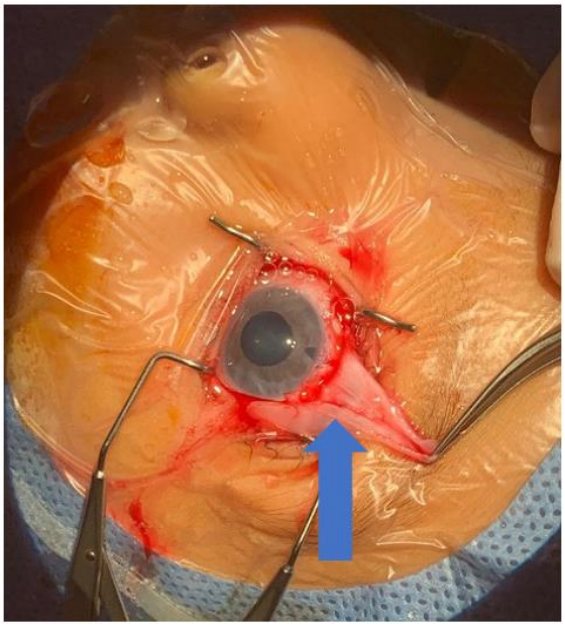

Figure 4. Trabeculotomy - Trabeculectomy procedure.

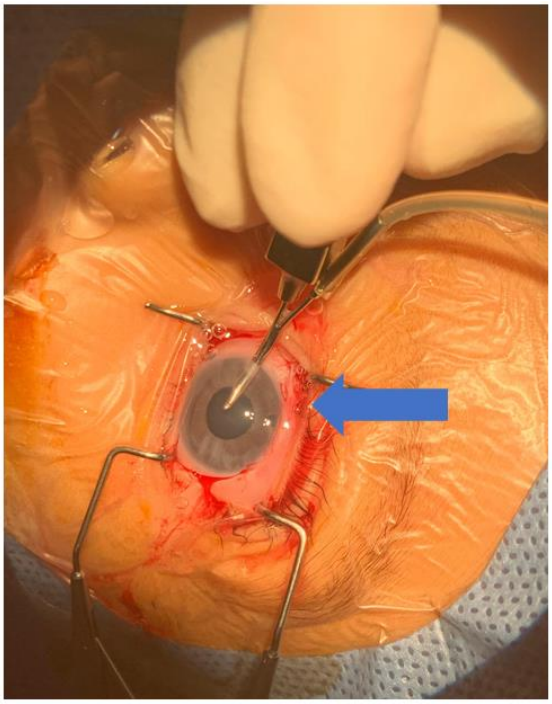

Figure 5. Irrigation aspiration.

3. Discussions

PCG is a rare genetic congenital disorder that affects children at birth [9]. Epidemiologically, the disease is more prevalent in boys, constituting 65% of cases, compared to girls, and it affects both eyes (bilateral) in 70% of cases [5]. The increased incidence in males is attributed to the influence of the hormone testosterone on the development of the trabecular meshwork [10]. However, it can also occur in girls unilaterally, as in this case. The typical triad for PCG includes excessive tearing, sensitivity to light, and involuntary eyelid spasms, all of

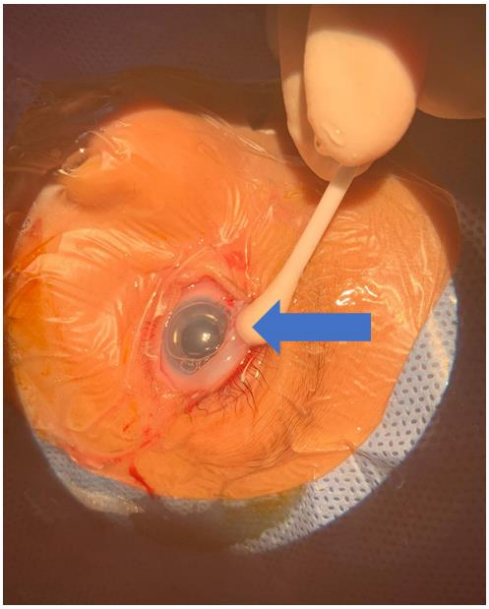

Figure 6. Operation complete.

which are present in this patient. Additional signs include an enlarged eye and corneal clouding [7, 9].

According to Wright and Spiegel, patients with primary congenital glaucoma experience symptoms of lacrimation, photophobia, blepharospasm, buphthalmos, and corneal clouding as this case also showed these symptoms and signs [9]. The classic triad of symptoms is linked to the rapid expansion of the infant's eye under high pressure, leading to corneal enlargement and often causing breaks in the Descemet membrane (Haab striae), resulting in corneal edema and opacification [5]. Associated signs include deep anterior chamber, buphthalmos, myopia, and optic nerve cupping. In extreme cases, the lens can dislocate [5, 7].

Operative treatment serves as the primary modality in managing primary glaucoma in children, with medication employed as adjunctive therapy [5]. Angle surgery (goniotomy, trabeculotomy-trabeculectomy) has a high success rate [5, 8]. There are various surgical techniques available for pediatric glaucoma [7]. In this instance, trabeculotomy-trabeculectomy was selected, taking into account the child's hazy corneal media and age being less than one year. Additionally, the pathophysiological basis of pediatric glaucoma, specifically abnormalities in the angle of the anterior chamber of the eye (trabecular meshwork), underpins the choice of trabeculotomy-trabeculectomy [5].

The choice of surgical procedure depends on the status of the cornea [11]. Combined trabeculotomy-trabeculectomy has been advocated to manage moderate to severe congenital glaucoma [12]. This procedure can provide access to dual outflow via Schlemm's canal and trabeculectomy fistula but has the disadvantage of damaging the conjunctiva [12]. Mandal et al. have reported only minor postoperative complications in their previous studies [13]. Sayed et al. have reported that surgical management of PCG is fraught with complications in both the early and late postoperative periods, including a shallow anterior chamber, hyphema, and thin cystic blebs [14]. However, Marwa et al. found that Combined Trabeculotomy Trabeculectomy proves to be a successful surgical intervention for PCG patients without complications that threaten vision [15]. Other factors that influence the prognosis of PCG include severity of disease, time of presentation, and time of surgery [7, 9, 12, 16].

Surgical treatment for PCG has a good prognosis; research that was conducted by Farid et al. revealed that Surgical management of PCG younger than one year of age achieved a good success rate with a low rate of visually significant postoperative complications [12]. Research by Malek et al. also revealed that 62 patients' mean IOP reduced from 22.68±3.99 mmHg to 9.75±3.88 mmHg ($P<0.0001$). The complete success rate was 91.6%, 88.4%, 84.7%, 71.6%, 59.7%, and 54.3%, respectively, at first, second, fourth, sixth, eighth, and tenth year [17]. This research aligns with the theory that if congenital glaucoma is diagnosed between 3 and 12 months of age and the corneal diameter is less than 14 mm, the prognosis is favorable [18]. The prognosis for visual function, in this case, is uncertain, as the condition was diagnosed at the age of 5 months and has received appropriate management [12]. However, unilateral cases typically have a less favorable prognosis compared to bilateral cases [19].

During the surgical procedure, a cataract measuring 4 mm in diameter was detected in the posterior capsule. However, the decision was made not to treat the cataract in the patient due to concerns that postoperative epithelialization and inflammation could lead to Posterior Capsular Opacity and synechiae, resulting in a secondary increase in Intraocular Pressure [20]. Moreover, the ratio of cataract diameter to pupil diameter still permits sufficient light entry. Special attention is being paid to closely monitoring the progression of cataracts.

While this study aimed to present the comprehensive management of primary congenital glaucoma for infants, it is important to acknowledge several limitations of our study. Firstly, as a single case report, the findings may not represent broader patient populations and should be interpreted cautiously. Additionally, the retrospective nature of the study design introduces the potential for recall bias and incomplete documentation of clinical details after the surgery was performed. Therefore, while this case report offers valuable clinical insights, further

research using larger sample sizes and prospective study designs is warranted to validate these findings and establish broader conclusions.

4. Conclusions

Congenital glaucoma can manifest as an isolated condition or as part of a systemic disorder, often accompanied by other ocular abnormalities. This necessitates thorough examination and multidisciplinary management. Early detection and timely, appropriate surgical intervention to regulate intraocular pressure can preserve good visual function throughout adulthood. In this case, the patient was diagnosed with Primary Congenital Glaucoma. The surgery was performed on the left eye. The patient's visual function improved, and the lacrimation and blepharospasm were reduced. Post Trabeculotomy-trabeculectomy surgery medications in children or newborns require anti-inflammatories and antibiotics; also, for post-operative management, we pay attention to the complications that might occur, as described in this case.

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Ethical Clearance: This article was approved by the Health Research Ethics Committee of RSUD dr. Zainoel Abidin.

Informed Consent Statement: Informed consent was obtained from the patient who had undergone this procedure.

Data Availability Statement: This article includes all the data supporting the results.

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Conflicts of Interest: All the authors declare no conflicts of interest.

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