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From Bright to Brightness: Mastering the Management of Bilateral Congenital Cataracts

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Abstract

Congenital cataract is the leading cause of vision loss in newborns and children, and it is caused by an opacity of the eye's lens that occurs at birth or within the first few months of life. Congenital cataracts may develop as unilateral or bilateral. Around 50% of cases of it are considered as idiopathic. A 4-month-old boy was brought by his parents to the Department of Pediatric Ophthalmology and Strabismus in RSUD dr. Zainoel Abidin Banda Aceh with a chief complaint of a white spot on the patient's right eye. The spot had appeared since birth and became more visible when he was 2 months old. The patient's overall condition was good. Both eyes' visual acuity was blink reflex. The ocular pressure in both eyes was normal. An ophthalmologic examination revealed a fully clouded right eye lens and aphakia in the left eye. The patient had undergone cataract surgery on his left eye on September 6th, 2023. He was diagnosed with Congenital Cataract Oculus Dexter (OD) and Aphakia Oculus Sinister (OS) before surgery. The patient underwent right eye cataract surgery with aspiration irrigation (AI), Primary Posterior Capsulotomy (PPC), and Anterior Vitrectomy (AV) without Intraocular Lens (IOL) implantation within the next two weeks. Following the surgery, the patient's visual function improved to fix and follow the object, the pupils looked round in both eyes, aphakia, and no membrane development. The patient was prescribed glasses for optical rehabilitation.



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1. Introduction

Cataracts are a major cause of blindness worldwide, particularly in developing countries [1]. Although most cataracts appear in elderly populations, a small percentage may manifest in children, with pediatric cataracts responsible for most childhood blindness. Congenital cataract is an ophthalmic condition characterized by the opacity of the lens in the eye that is

present at birth or within the first few months of life. The onset of this condition at an earlier age poses a greater risk of amblyogenic deprivation, resulting in reduced visual acuity. Therefore, early diagnosis and prompt intervention are essential to prevent or minimize the complications associated with congenital cataracts [2, 3].

Congenital cataract is the primary cause of vision loss in infants and children. The World Health Organization

(WHO) reports that there are 2.2 billion visually impaired people worldwide, with 65.2 million of them having cataracts. Congenital cataracts can be detected at birth or during the first ten years of life. According to the WHO, more than 14 million children are bilaterally blind due to cataracts, which accounts for over 50% of all causes of blindness globally. In developed countries, congenital cataracts occur in 1-6 out of 10,000 live births, while in developing countries it is between 5-15 out of 10,000 births [4]. The approximate pooled prevalence of cataracts in children either due to congenital or developmental factors in Indonesia was 0.60, Bangladesh was 0.92, Iran was 1.47, Bhutan was 2.01, China was 3.68, Thailand was 4.27, India was 4.47, Malaysia was 5.33, Nepal was 5.42, Vietnam was 9.34, and Cambodia was 10.86 [5].

Congenital cataracts can develop in one or both eyes and around 50% of cases are idiopathic. Cataract surgery in children is not the same as in adults. The cortex and nucleus of the lens of a child's eye are softer than those of an adult, so cataract surgery can be performed using the aspiration irrigation technique. Posterior Capsular Opacification (PCO) and glaucoma are major complications of cataract surgery in children. However, PCO can be prevented by performing surgery with Primary Posterior Capsulotomy (PPC), Anterior Vitrectomy (AV), and postoperative medication. Amblyopia should be treated immediately after congenital cataract surgery, with regular follow-up tests to achieve adequate visual acuity [6–8]

The eyes of children and infants are not miniatures of adult eyes. Due to the differences in anatomy and physiology, the management of cataract in children and infants requires special considerations. In this case report, we present comprehensive management steps for congenital cataracts, including optical rehabilitation after cataract surgery. The procedure used an irrigated aspiration technique, PPC and AV. This approach resulted in improved vision and was safe for children or infants with bilateral congenital cataract.

2. Cases

A 4-month-old boy was brought by his parents to the Department of Pediatric Ophthalmology and Strabismus in RSUD dr. Zainoel Abidin Banda Aceh with a chief complaint of a white spot on the patient's right eye. The parents said that the spot had appeared since birth and became more visible when he was 2 months old. The patient was not able to focus on objects around him. The patient had been diagnosed with a bilateral congenital cataract in early September and had undergone cataract surgery on his left eye on September 6th, 2023.

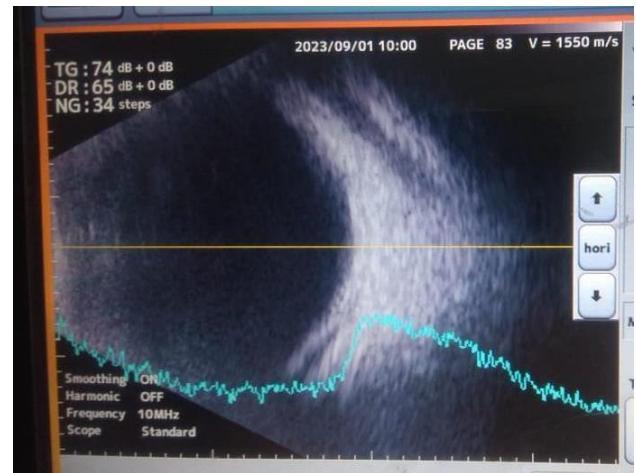


Figure 1. Ultrasonography examination on the patient's right eye.

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 patient was delivered via cesarean section, with a birth weight of 2900 grams. Upon delivery, there was no history of cyanosis. The patient had a history of NICU admission for ten days due to shortness of breath. The patient received complete immunization and he was exclusively breastfed. The patient was seen actively reacting to sounds, babbling, spontaneously smiling, laughing, and lifting his head. However, the patient's eyes could not follow moving objects and lacked focus on her parents and others. There was no family history of the same condition.

The patient's overall condition was good. Both eyes' visual acuity was blink reflex. The ocular pressure in both eyes was normal. An ophthalmologic examination revealed a fully clouded right eye lens and aphakia in the left eye. Because the posterior section could not be examined due to a clouded lens, an ultrasonography examination was performed and the results were within normal ranges. (Figure 1).

On September 20th, 2023, the patient was scheduled for surgery with Aspiration Irrigation (AI), PPC, and Anterior Vitrectomy (AV) on his right eye (Figure 2). A laboratory examination, chest X-ray, consultation with pediatrics, cardiology, and anesthesia were undertaken to acquire approval for surgery under general anesthesia. The patient had pre-anesthesia physical state II according to the American Society of Anesthesiologists (ASA). The patient was diagnosed with Congenital Cataract Oculus Dexter (OD) and Aphakia Oculus Sinister (OS) before surgery. Following surgery, the patient was diagnosed with Aphakia Oculus Dexter. After surgery, the patient

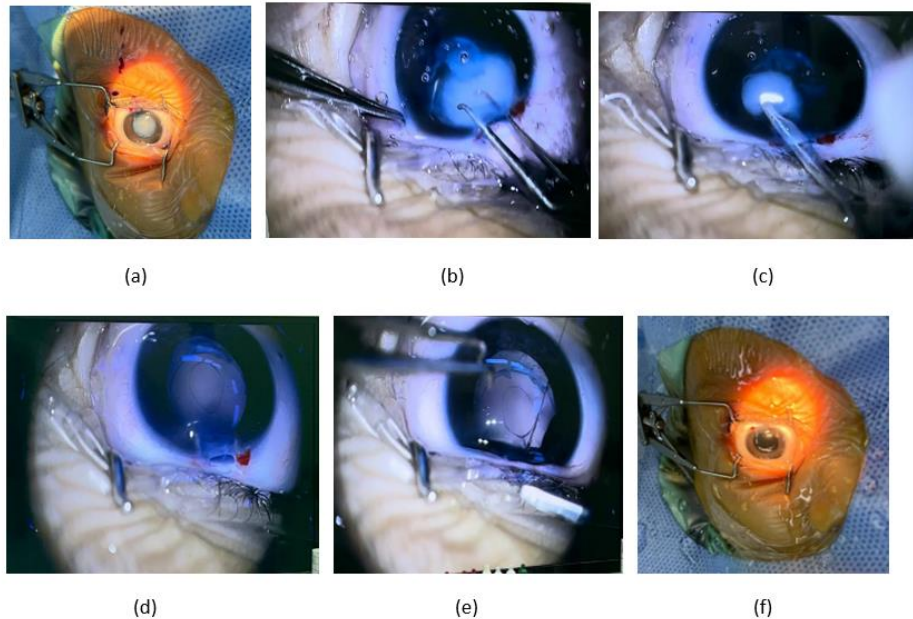


Figure 2. The steps of congenital cataract surgery on a 4-month-old boy's right eye: a) a clinical image of the patient taken intraoperatively (before surgery) revealed a cloudy lens with anterior capsular fibrosis; b) The anterior lens capsule was stained using trypan blue then Continuous Curvilinear Capsulorhexis (CCC) was performed; c) Irrigation and aspiration of the lens nucleus and cortex; d) Primary Posterior Capsulotomy (PPC) of the posterior lens capsule and Anterior Vitrectomy (AV) were performed, e) The corneal-scleral incision was stitched using nylon 10.0, f) The surgery was done, the cortex and nucleus were clear.

was prescribed antibiotic syrup, analgesic syrup, antibiotic eye drops six times a day, anti-inflammatory eye drops eight times a day. The patient was discharged after no complaints on the first 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 visual function improved to fix and follow the object, the pupils looked round in both eyes, aphakia, and no membrane development. The patient has been prescribed glasses with +20 diopter (D) accommodation for optical rehabilitation.

3. Discussions

In this case, the 4-month-old boy had a chief complaint of a white spot on the patient's right eye. The spot had appeared since birth and became more visible when he was 2 months old. An ophthalmologic examination revealed a fully clouded right eye lens and aphakia in the left eye. The patient had undergone cataract surgery before on his left eye. Because of these findings, the patient was diagnosed with congenital cataract. Cataracts are particularly frequent in the elderly and are associated with the aging process, but they can also occur as congenital defects. Congenital cataracts can be unilateral or bilateral and have a good prognosis if treated early. Almost half of all congenital cataracts are sporadic, with an unknown cause. A prenatal history of maternal

illnesses such as TORCHS (Toxoplasmosis, Rubella, Cytomegalovirus, Herpes Simplex, HIV, and Syphilis) infection during the first trimester of pregnancy, and the usage of drugs throughout pregnancy, are essential to determine the cause of congenital cataracts. Cataracts are also common in preterm and underweight newborns. [2,4] Genetic cataracts account for 8-29% of all congenital cataracts, with autosomal dominant inheritance being the most frequent. Many professionals advise genetic counseling when evaluating congenital cataracts [5, 9]

A white patch, or leukocoria, will be noticed on physical examination of the pupils of infants with congenital cataracts [4, 10]. At that moment, the child's visual function is the blink reflex, which should be able to follow the light employed as a source of stimulus (fix and follow the light) [11]. Congenital cataracts can lead to complications such as deprivation amblyopia. Bilateral cataracts are associated with nystagmus, whereas unilateral cataracts are associated with strabismus. If the fundus is unable to be examined due to cataracts, ocular ultrasonography (B-scan) can be conducted to establish the absence of retinal detachment and other intraocular masses. An ultrasound examination can also determine the length of the eyeball [12, 13].

The treatment of a cataract is determined by whether it is unilateral or bilateral, the existence of other eye disorders, and the time of onset. The majority of children with cataracts require surgery, with only a few exceptions

that are treated conservatively. A complete eye examination, including evaluation of the cataract morphology, is essential to identify the optimal timing and method of treatment [8]. The location of opacities in the lens and the morphology of the cataract can provide information about the cause of the cataract. Cataracts in children are classified according to their morphology as anterior polar, posterior polar, nuclear, lamellar, zonular, sutural, or cerulean [2].

The treatment of congenital cataracts is classified into two categories: non-surgical and surgical treatment. Pediatric cataracts are managed non-surgically by observation and pupil dilatation. Cataracts less than 3 millimeters in size or placed pericentrally are evaluated so that they do not interfere with the visual axis. If the cataract visibly measures more than 3 millimeters in diameter and is centrally situated, surgery is conducted [13]. Cataract surgery in infants or children is not the same as in adults. This is because infants and children have anatomical differences, such as a thicker and more elastic anterior lens capsule, a thicker vitreous, and a soft lens. These differences can make the surgery more challenging to manage. This makes way easier to escape into the lens's posterior capsule during the Continuous Curvilinear Capsulorhexis (CCC) procedure, which should be avoided due to the necessity of capsular support for lens implantation [14].

Because the cortex and nucleus of the lens are softer in children's eyes than in adult eyes, cataract surgery can be performed using the irrigation aspiration approach. However, the cortex in infant eyes is stickier, making aspiration difficult for some infants [8]. Because of the tendency of pediatric lens epithelial cell re-proliferation, it is critical to remove the entire lens cortex [12]. Despite significant advancements in pediatric cataract surgery techniques, Posterior Capsular Opacity (PCO) remains the most prevalent post-cataract complication, affecting nearly 100% of newborns. PCO can be avoided by doing surgery with a PPC and an AV. PPC and AV techniques have the disadvantages of requiring specialized skills and experience, extra surgical time, loss of vitreous, a high rate of macular edema, and retinal detachment [15].

This patient was 4 months old. As a result, no IOL was implanted following cataract removal, and the patient was given glasses for optical rehabilitation. The absolute error in children is higher than in adults. There are special considerations when it comes to IOL insertion during infant or pediatric cataract surgery where age and corneal diameter are important. Children under one year of age are considered not recommended for IOL implantation due to the shallow capacity of the eye chamber, which can cause difficulty during IOL insertion,

the possibility of secondary glaucoma, a higher risk of inflammation, which increases the risk of PCO, and the high risk of complications and unpredictable refractive changes due to rapid eyeball growth at the age of less than two years. Intraocular lens (IOL) implantation can be also considered in pediatric eyes with an axial length greater than 17 mm and a corneal diameter greater than 10 mm, so the lens capsule is large enough to accommodate the IOL. The residual anterior and posterior capsules also merge more easily in the absence of IOL implantation, so *Soemmering's* ring is less likely to obstruct the visual axis [2, 11, 16–18].

Children experience higher inflammation after cataract surgery than adults. Younger age, a history of uveitis, intraoperative iris manipulation, and extended surgery are all factors that may aggravate inflammation. Therefore, children or newborns require more aggressive anti-inflammatory medications after cataract surgery. Topical antibiotics, corticosteroids, and cycloplegics are typically administered for many weeks following surgery [19]. Optical rehabilitation plays an essential role in supporting the formation of shadows on the retina. This is one of the efforts to prevent and treat amblyopia. Optical rehabilitation with contact lenses or glasses, as well as amblyopia therapy, should start immediately after surgery, taking into consideration postoperative recovery. Eyeglasses are the safest and simplest approach to correction for infants with bilateral aphakia. Glasses may be easily adjusted to accommodate the refractive change that occurs in the eye. Infants require glasses for near vision due to their visual activity being predominantly focused at close range. Contact lenses can also be used in patients with bilateral Aphakia, however, they require more maintenance [11, 20].

The eyes should be evaluated on the first day and a week after surgery to rule out any potential complications such as infection, increased intraocular pressure, visual acuity, refraction, anterior segment, and visual axis opacities. It is also necessary to evaluate the posterior region. Furthermore, the child is examined on a regular basis for strabismus, secondary glaucoma, and retinal detachment. Children who have cataract surgery as babies and children with small eyes are more likely to develop glaucoma. However, glaucoma often develops years after surgery [11].

4. Conclusions

Congenital cataract is the primary cause of vision loss in infants and children. Both unilateral and bilateral congenital cataracts should be removed as soon as they are detected so that the child's brain can experience vision and also prevent amblyopia. There are several

considerations in performing cataract surgery in pediatrics compared to adult cataract surgery due to anatomical and physiological differences in the eye such as thicker cornea and curvier curvature, shallower anterior chamber, soft nucleus, different eyeball length increases in three stages making it difficult to calculate the power of the lens to be implanted, and the high proliferation and epithelialization that causes PCO. In this case, the patient was diagnosed with bilateral congenital cataract. The surgery was performed on the left eye initially, followed by the right eye in the following two weeks. the patient's visual function improved to fix and follow the object, the pupils looked round in both eyes, aphakia, and no membrane development. Post-cataract surgery medications in children or newborns require more aggressive anti-inflammatories, also post-operative visual rehabilitation should not be overlooked, as described in this case.

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