Case Report

Successful Management of Complex Congenital Cataract and its Complication

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Abstract

Management of cataract and its complication after surgery in a child with complex congenital cataract where accompanied by other ocular abnormality can pose a great surgical challenge. This paper reported a case in dr. Cipto Mangunkusumo Hospital on 2021.This case report described a case of an 18-year-old girl with slowly decreasing vision after undergoing surgery charity on the right eye (RE) around 4 months ago due to cataract which she had since birth. Best corrected visual acuity (BCVA) preoperatively of the RE and LE was 6/60 and 0,5/60 respectively. Ocular assessment was pseudophakia with Posterior Capsular Opacity (PCO) on the RE and cataract on the left eye (LE) accompanied by microcornea, nystagmus and coloboma of iris and retina on both eyes. Secondary posterior capsulotomy surgery was done on the RE and cataract extraction, primary posterior capsulotomy (PPC) on the LE with anterior vitrectomy (AV) on both eyes. Last follow-up postoperatively showed a significant improvement of uncorrected visual acuity on her RE from 0.5/60 preoperatively to 6/30, meanwhile only subjective improvement of VA was found on her LE. This case can achieve visual improvement with promptly surgical treatment without complication and routine monitoring still needs to be done.
Keywords: *congenital, coloboma, microcornea, cataract surgery, pseudophakia, complication.*

Keberhasilan Tata Laksana Katarak Kongenital Kompleks dan Komplikasinya

Abstrak

Tata laksana katarak dan komplikasi pasca-operasi pada pasien anak dengan katarak kongenital kompleks disertai kelainan okular lain seperti koloboma dan mikrokornea merupakan operasi dengan tantangan tersendiri. Makalah ini melaporkan kasus di Rumah Sakit dr. Cipto Mangunkusumo pada tahun 2021. Kasus ini mendeskripsikan seorang anak berusia 18 tahun dengan keluhan penurunan tajam penglihatan mata kanan secara bertahap sejak tiga bulan terakhir setelah menjalani operasi katarak di acara bakti sosial karena ada bintik putih di bagian hitam mata yang dimiliki sejak lahir. Tajam penglihatan terbaik pra-operasi 6/60 di mata kanan dan 0,5/60 di mata kiri. Pada pemeriksaan mata ditemukan pseudofakia, kekeruhan kapsul posterior di mata kanan dan katarak di mata kiri disertai mikrokonea, koloboma iris dan retina, serta nistagmus di kedua mata. Dilakukan operasi mata kanan berupa kapsulotomi posterior sekunder dan mata kiri berupa ekstraksi katarak, kapsulotomi posterior primer disertai vitrektomi anterior di kedua mata. Terdapat peningkatan tajam penglihatan signifikan pasca- operasi saat kontrol terakhir pada mata kanan pasien yaitu dari 0,5/60 menjadi 6/30. Pada mata kiri perbaikan tajam penglihatan hanya secara subjektif. Kasus ini dapat mencapai perbaikan penglihatan dengan operasi yang tepat tanpa komplikasi dan diperlukan pemantauan rutin pasca-operasi.
Kata kunci: *kongenital, koloboma, mikrokornea, operasi katarak, pseudofakia, komplikasi.*

Introduction

Cataract represents a leading cause of blindness worldwide and the estimated prevalence of congenital cataract is 2 to 3 per 10,000 live births. Childhood cataract is one of the most important causes of blindness and severe visual impairment in children and is responsible for 5–20% of pediatric blindness worldwide. Cataract in children may be congenital or acquired, unilateral or bilateral. There was no difference in the prevalence based on gender.[1]

Congenital cataract may occur alongside other ocular anomalies, such as microcornea and coloboma. Coloboma can involve any part of the eye caused by the failure of embryonic fissures to close during the fifth week of gestation that characteristically leads to an inferonasal defect. Microcornea is a clear cornea of normal thickness with a diameter less than 9 mm in the newborn and less than 10 mm after 2 years of age. It may be the result of overgrowth of the tips of the optic cup and may be inherited in an autosomal dominant or recessive manner.[2,3]

Cataract surgery in eyes with microcornea and coloboma can present greater intraoperative challenges and pose higher risk of complications.[4] Early diagnosis and referral for surgery are important for successful management. This case report wants to highlight the importance of early and proper cataract surgery including handling of complication and patient's monitoring in bilateral complex congenital cataracts to give favourable surgical outcome and avoid childhood blindness finally.

Case Illustration

This paper reported a case in dr. Cipto Mangunkusumo Hospital on 2021. An 18-year-old girl presented to our tertiary eye health care hospital with slowly decreasing vision after undergoing surgery charity on the right eye (RE) around 4 months ago due to cataract which she had since birth. At four months old of age, the parents noticed that their child had no respond when stimulated by an object and complaint of a whitish appearance on both of her eyes. The patient had visited the ophthalmologist and a surgery was advised but she never had a surgery due to the economic status.

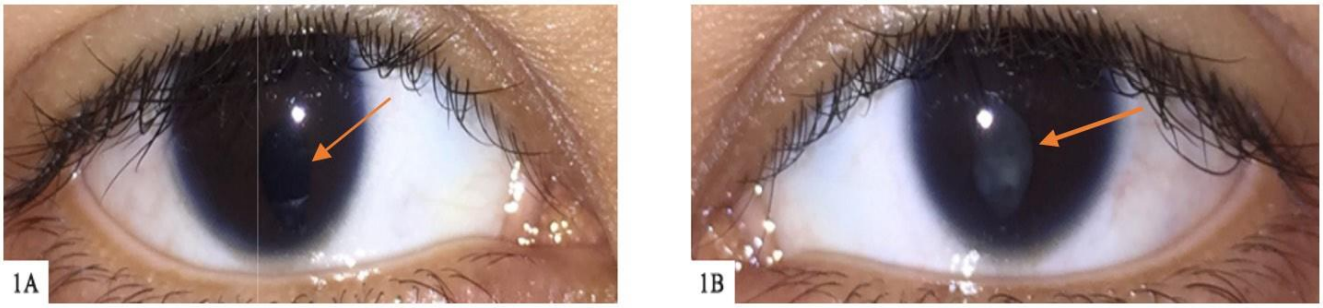

Figure 1. Pre-operative view showed microcornea and iris coloboma (orange arrow) on both eyes with (A) IOL on the RE and (B) cataract on the LE.

Ophthalmology examination revealed best corrected visual acuity (BCVA) was 6/60 on right eye (RE) and 0,5/60 on LE. The intraocular pressure (IOP) was 18 mmHg on RE and 15 mmHg on LE (within normal limit). Anterior segment showed microcornea (corneal diameter 9 mm for right eye and 8 mm for left eye), iris coloboma, and nystagmus on both of her eyes (Figure 1A and 1B). There was intraocular lens (IOL) with posterior capsular opacity (PCO) on the RE covering the visual axis and lens opacity of the LE (Figure 2A and 2B). Posterior segment showed retinal coloboma on RE meanwhile for LE it was hard to be evaluated due to lens opacity. Ocular USG examination was done on LE and there was also retinal coloboma (Figure 3).

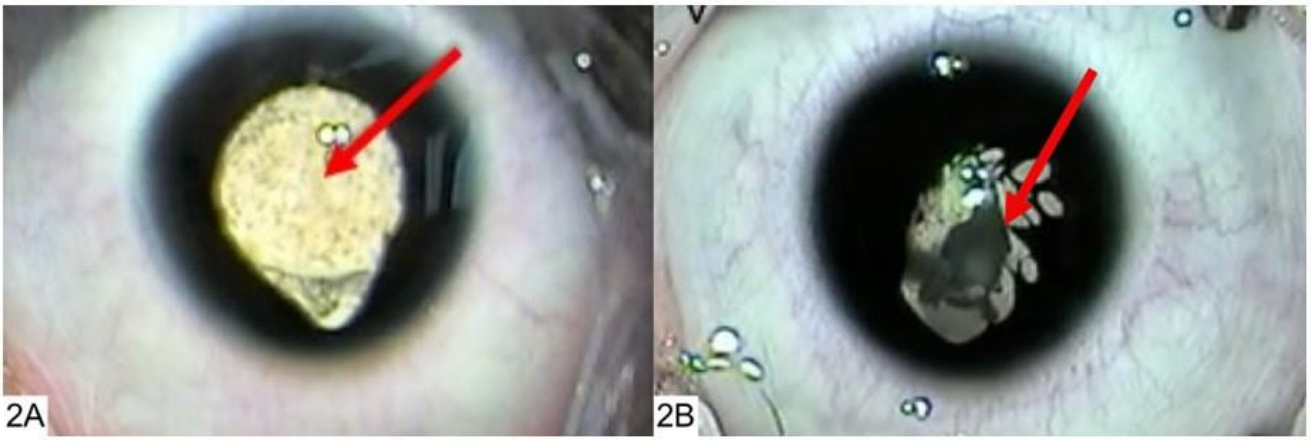

Figure 2. Intraoperative view showed microcornea, iris coloboma on both eyes with (A) IOL and PCO on the RE and (B) cataract on the LE in red arrow.

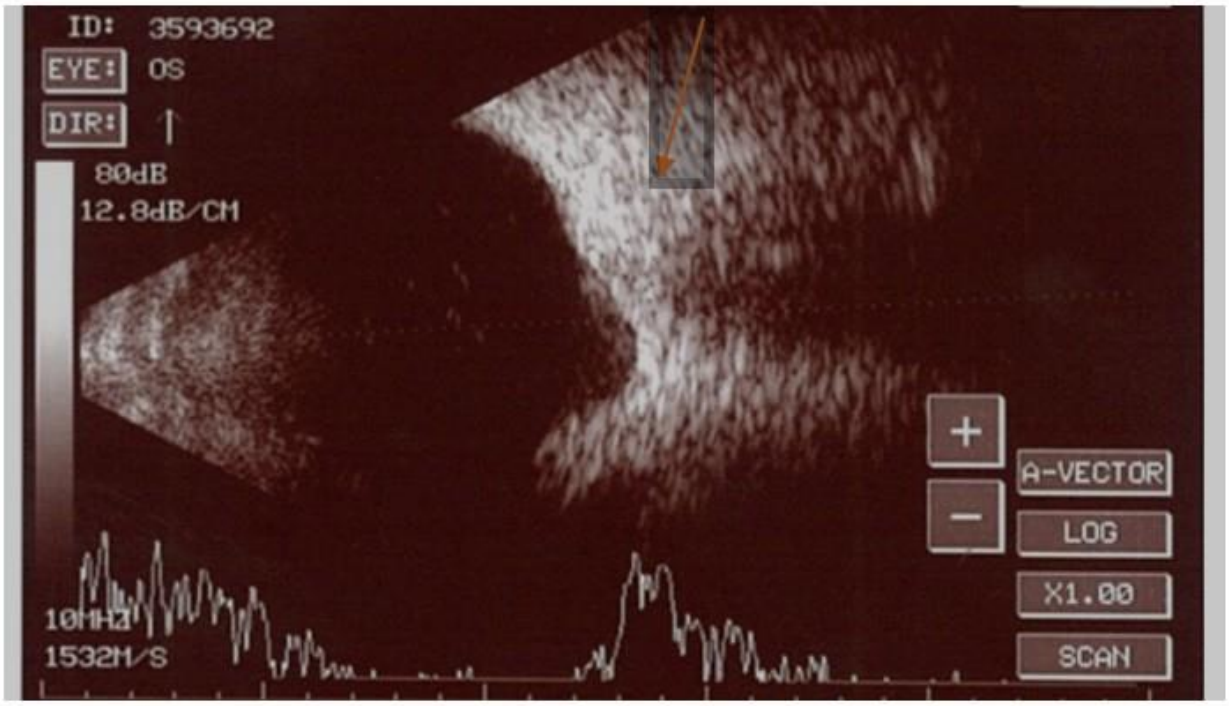

Figure 3. Retinal coloboma (orange arrow) on the LE with ocular USG examination.

She was diagnosed with congenital cataract on the LE, pseudophakia and PCO on the RE, bilateral coloboma of iris and retina, microcornea, and sensory nystagmus on both eyes. The patient was planned to undergo secondary posterior capsulotomy on the RE and cataract extraction, primary posterior capsulotomy (PPC) of LE in general anaesthesia two weeks after the RE's surgery. Anterior vitrectomy (AV) was performed on both eyes. The result of surgery showed no opacity on visual axis of both eyes (Figure 4). Unfortunately, retinal coloboma was found on both eyes with retinal photo examination and it was more advanced on LE (Figure 5). The uncorrected visual acuity of her right eye improved to 6/30. However, there is only subjective improvement of visual acuity on the LE due to wide retinal coloboma. There was no complication in postoperative follow-up.

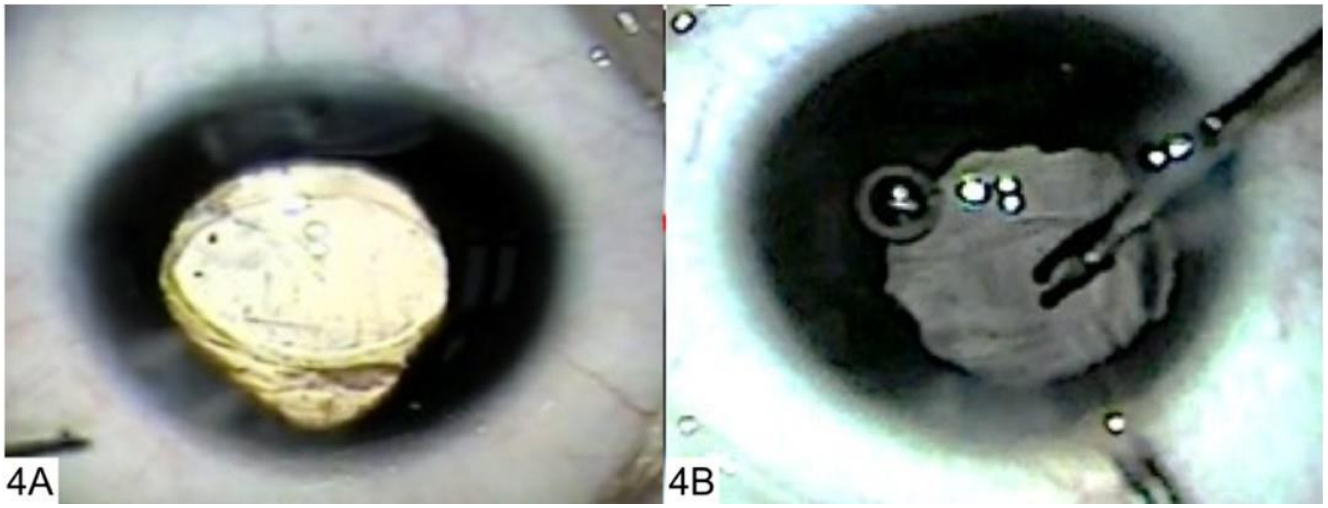

Figure 4. Clear visual axis after second surgery on RE (4A) and first surgery on LE (4B).

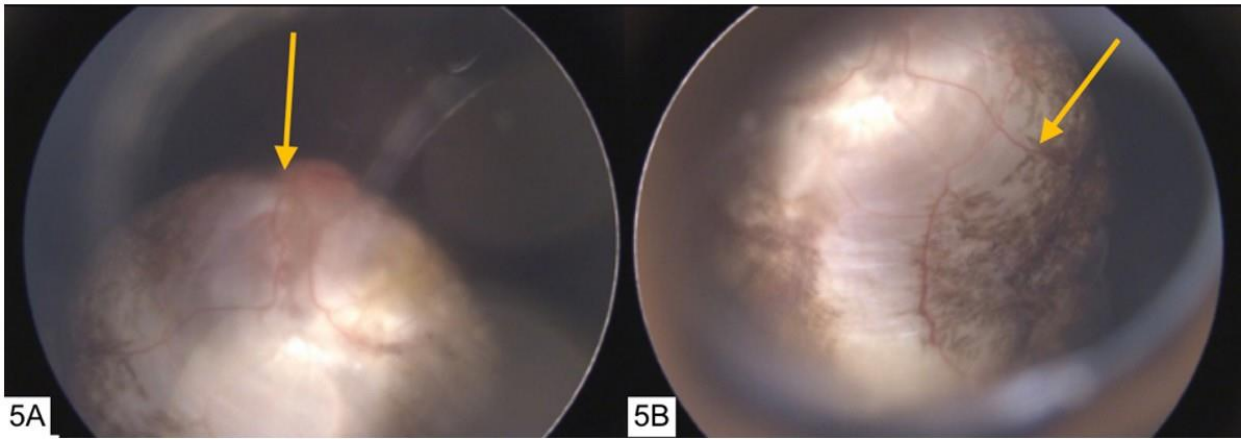

Figure 5. Bilateral retinal coloboma (yellow arrow), RE (5A) and wider on LE (5B).

Discussion

Congenital cataract that happened due to gene mutation can be accompanied by other ocular anomalies. Genes such as PAX6, FOXC1, and PITX2 play an important role in ocular formation. The gene defect cause ocular abnormalities in the formation of structures and eye sizes. Jamieson et al[5] found that MAF transcription factor genes play role in the formation of polarized and posterior polar cataracts accompanied by microcornea and iris coloboma without microphthalmia. In our case, she had bilateral congenital cataract accompanied by microcornea and coloboma of retina and iris on both eyes.

Surgical management of eyes with microcornea, coloboma and congenital cataract is complex and technically difficult. Several considerations need to be pondered before performing cataract surgery in this case. The first one is the time of surgery; early surgery is associated with better visual acuity outcomes. A critical period to perform cataract surgery in children with bilateral congenital cataracts is before age of 8 weeks. At that time is the longer time frame during which the developing visual system retains plasticity and can thus be modified before full visual maturation. This period may minimize the effect of visual deprivation on the developing visual system and provide for optimal rehabilitation of visual acuity.[6,7] In this patient, early surgery should be possible when the diagnosis had been established in four months of age, meanwhile the surgery was done too late where it has passed the golden period of eye development in children and the effect of visual deprivation have manifested. The timing of surgery and duration of visual deprivation are important factors in the development of nystagmus. Prolonged duration of the cataract (>6

weeks) is associated with a significant risk for both strabismus and nystagmus. Nystagmus has also been reported to be more frequent in bilateral rather than unilateral cataract. In this case, the patient was known to get cataracts since the age of 4 months and the cataract surgery was performed at the age of 18 years on eye condition with nystagmus. The presence of nystagmus was shown to be associated with worse visual outcome in children with bilateral congenital cataracts.[8] In addition, the presence of retinal coloboma give poor prognosis of vision in this patient moreover on the LE.

Complex congenital cataract surgery can pose greater intraoperative challenges and risk of higher postoperative complications because of the associated ocular malformations compared to congenital cataract surgery alone. Intraoperative complications might be associated with shallow anterior chamber, zonular deficiency and poor pupil dilation which could be managed well in this case. Postoperative complications are the development of secondary glaucoma especially in eyes with microcornea and PCO formation. The prevalence of postoperative glaucoma is 15-45%. Increased IOP as an early sign of glaucoma often arise especially in one year postoperatively.[9] Although IOP rise was not found in this case, nevertheless routine follow-up is required and important to monitor IOP.

PCO occurs rapidly in young children because of intense inflammation process after surgery. PPC and AV surgical techniques which can prevent the development of PCO, should be performed at the time of cataract surgery in a child.[10] In this case, patient was referred for further treatment (secondary PC and AV surgery) due to PCO on her RE. Previously, she underwent cataract surgery with minimal facility and limited operative resources at the mass cataract surgery charity where surgical risk were higher and could endanger the patient's vision function. Therefore, PCO had developed in four months after surgery that worsen her vision. PPC and AV were performed simultaneously with cataract surgery for the prevention of PCO on the LE, so secondary surgery can be greatly reduced.[11] In addition, these eyes should be handled with great caution due to the increased risk of poor surgical results.

The third one is the implantation of IOL. Visual rehabilitation after pediatric cataract surgery remains a challenge, essential and need such as glasses, aphakic contact lens wear and IOL implantation which give best correction of visual acuity.[12] Some controversies remain over the benefit of IOL implantation in young children. Correction of aphakia after congenital cataract surgery with primary IOL implantation results in improvement of visual acuity and binocular vision outcome, less occurrence of strabismus, but a higher rate of complications requiring reoperation.[13-14] IOL implantation must be conducted carefully to avoid possible complications. The right eye's IOL implantation with microcornea was performed on this case. It could be very challenging due to a small cornea and shallow anterior chamber. This condition can lead to glaucoma. However, the use of posterior chamber IOL was considered as a safe technique.[9] In this case, on follow up examinations showed good IOP results and the patient was planned to have a periodically visit schedule. Patients were also given glasses to maximize visual acuity on the RE.

Conclusion

Complex congenital cataract should be regarded as a major surgical challenge and managed with great caution to prevent the increased risk of complications and poor surgical results. Early cataract surgery is required to reduce the probability of visual deprivation. Furthermore, the technique of surgery and implantation of IOL must be considered deliberately because they also play a big role in the outcome and complication of the surgery. Routine follow-up needs to be done in order to monitor the possibility of postoperative complications. Timely referral to eye health center with pediatric ophthalmology team sub-specializing in this field is required to manage such cases.

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