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# Bilateral Preexisting Congenital Posterior Capsular Defects with Accompanying Membranes

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#### **Abstract**

**Purpose:** To present three cases having bilateral congenital posterior capsular defects accompanying bilateral congenital cataracts.

**Cases:** Similar to the previous reports there were characteristic demarcation of thickened margins on the posterior capsule defects and white dots on the anterior vitreous face in all of our cases. In addition to previous reports, we detected a semi-transparent membrane at the location of the posterior capsule defect bilaterally in all of our cases.

**Observations:** This membrane was loosely attached to the borders of the posterior capsular opening and we removed it with vitreus cutter in two cases and with forceps in the other. In two cases the membranes covered the entire posterior capsular defect area; but in one case the membrane covered only the half of the defect. The cases were managed by standard irrigation – aspiration and anterior vitrectomy.

**Conclusion:** Ophthalmologists should be aware that in some congenital cataracts, they may notice congenital posterior capsular defects with accompanying membranes.

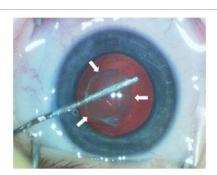
#### Introduction

Bilateral congenital cataract is the most common cause of treatable childhood blindness, accounting for 5% to 20% of blindness in children worldwide. In developing countries, the prevalence of blindness from cataract is higher—about 1 to 4 per 10 000 children [1]. Mostly bilateral congenital cataracts are idiopathic and about one third of cases are hereditary. Other causes include metabolic disorders and intrauterine infections [2,3]. Also, congenital cataracts can be associated with systemic abnormalities such as trisomy 21 and Turner's syndrome [4,5].

Preexisting posterior capsule defect (PCD) in congenital cataract is a lens abnormality first reported by Vasavada [6]. This condition manifests in infants as a congenital cataract with an existing defect in the posterior capsule. Interestingly, in these cases cataract development is seen after some months from birth [6]. Osher and Vajpayee previously described preexisting PCDs in cataract of adults or that caused by trauma [7,8]. In our study, we reported three cases of bilateral PCDs and posterior capsular membranes accompanying congenital cataract.

## Case 1

An 8-month-old boy presented with an abnormal red reflex in both of his eyes. His parents reported us that they did not see any abnormality in his eyes at birth; but at about three months age his eyes showed a somewhat abnormal light reflex and poor fixation. The intrauterine and perinatal events were unremarkable. No abnormalities were noted on routine pediatric examination. There was no family history of congenital cataract. Visual acuity testing showed poor fixation in both eyes. On examination under general anesthesia, both eyes had nuclear cataract with posterior cortical extension of the opacity. Pre-operative IOP values measured with Schiotz tonometer was 16.5 mmHg in both of his eyes. Previous to surgery, an echography was performed in order to rule out any retinal abnormalities. Cataract surgery was performed on both eyes at the age of 9 months with one week interval in between. Intraoperatively in both eyes, at the end of the aspiration, a preexisting posterior capsule defect (PCD) and a membranous tissue were seen at the center (Figure 1). This easily movable semi-transparent membrane was located nearly at the same place of PCD. It was above the anterior vitreus. It was not connected posteriorly to the vitreus fibers, but it was loosely attached to the borders of posterior capsule defect. Its size was nearly the same with the size of the posterior capsule defect. We removed it with vitreus cutter. There were small white dots on the anterior surface of the vitreus; but the margins of the posterior capsular defect were not much thickened. In both operations, anterior vitrectomy was performed and triamcinolone acetonide injection into the anterior chamber was done to lessen the post –operative inflammatory reactions. An intraocular lens (IOL) was not implanted into the eyes and the patient was left aphakic bilaterally. On the last follow-up, anterior segment examination was unremarkable except for the presence of aphakia with clear visual axis. Intraocular pressure



**Figure 1:** The left eye of Case 1 after aspiration of lens. A membranous opacity is seen above the iris spatule. White arrows indicate the borders of the membrane.

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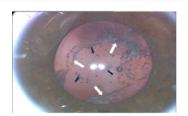
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**Figure 2:** The right eye of Case 2 after aspiration of lens. A membranous opacity is seen at the posterior capsular area. Blue arrows indicate the borders of the membrane.



**Figure 3:** The right eye of Case 3 after aspiration of lens. A membranous opacity is seen at the posterior capsular area. Dark arrows indicate the borders of the membrane and white arrows indicate the borders of the posterior capsule defect.

was 12 mmHg in both eyes and fundus examination was within normal limits. Spectacles were prescribed for visual rehabilitation. One year later from the first operations, we performed secondary IOL implantations. The patient was prescribed glasses for near vision. In the follow-up period, no additional problems occurred.

### Case 2

A 15-month-old boy presented with poor visual acuity in both of his eyes. The intrauterine and perinatal events were unremarkable. No abnormalities were noted on routine pediatric examination. Visual acuity testing showed poor fixation in both eyes. On examination under general anesthesia, the anterior segments were observed to be normal, except for the presence of a white mature cataract bilaterally. There was no family history of congenital cataract. Previous to surgery, an echography was performed in order to rule out any retinal abnormalities. The infant underwent cataract surgery on the right eye at 16 months of age and one week later, the left eye was operated for congenital cataract. Before the operations, we did not notice any abnormality at the posterior capsule. But, intra-operatively in both eyes, at the end of the aspiration, a preexisting posterior capsule defect (PCD) was seen at the center. Additionally, there was a semi-transparent membrane located nearly at the same place of PCD (Figure 2). Its size was nearly the same with the size of the posterior capsule defect. It was loosely attached to the surrounding posterior capsule. It has no connections to the vitreus fibers or ciliary processes. It was more transparent than the membrane seen in case 1. It was also thinner than the membrane seen in case 1. We detached this membrane easily with a forceps by pulling it. Margins of the posterior capsule defect were slightly thickened and there were some white dots on the anterior vitreous face. In both operations, anterior vitrectomy was performed and triamcinolone acetonide injection into the anterior chamber was done. Intraocular lens (IOL) implantation was not performed and the eyes were aphakic at the end of the operation. On the last follow-up, anterior segments were normal except aphakia bilaterally. Intraocular pressures were 11 mmHg in both eyes and fundus examination was within normal limits bilaterally. Postoperatively, spectacles were prescribed for visual rehabilitation. Nine months later from the first operations, we performed secondary IOL implantations. The patient was prescribed glasses for near vision. In the follow-up period, no additional problems occurred.

## Case 3

A 36-month-old boy presented with poor visual acuity in both of his eyes. No abnormalities were noted on routine pediatric examination. Best corrected visual acuity was counting fingers for the right eye and 20/80 for the left eye. In biomicroscopical examination, the anterior segments were observed to be normal, except for the presence of a white mature cataract in the right eye and lamellar cataract in the left eye. There was no family history of congenital cataract. Previous to surgery, an echography was performed in order to rule out any retinal abnormalities. The child underwent cataract surgery on the right eye at 36 months of age and one month later, the left eye was operated for congenital cataract. At this case, we noticed the defects on the posterior capsules under biomicroscope preoperatively. But we detected the semi-transparent membrane per-operatively in the right eye. Its size was nearly half of the size of the posterior capsule defect (Figure 3). It was firmly attached to the surrounding posterior capsule in one side and loosely attached to the underlying vitreus in the other sides. We removed this membrane with vitreus cutter. Margins of the posterior capsule defect were much thickened when compared with the other two cases. There were some white dots on the anterior vitreous face. In both operations, anterior vitrectomy was performed and triamcinolone acetonide injection into the anterior chamber was done. Implantation of IOL was performed in this case and optic capture was done (haptics were in the sulcus, optic was in the capsular bag). On the last follow-up, anterior segments were normal except pseudophakia bilaterally. Intraocular pressures were 12 mmHg in both eyes and fundus examination was within normal limits bilaterally. The patient was prescribed glasses for near vision.

# Discussion

Posterior capsular breaks existing prior to surgery may be detected for the first time during lens surgery. Such preexisting breaks are often seen with traumatic cataracts, intraocular tumors and in cases of posterior polar cataracts [9]. Bilateral congenital preexisting posterior capsular defects (PCDs) in the absence of a posterior polar cataract or posterior lenticonus were first reported by Vajpayee [10]. The etiology of preexisting PCD is unknown; but intrauterine abnormalities of the embryonic lens may take role in its pathogenesis.

One possible diagnosis for these three cases may be posterior lenticonus that is a rare ectasia of the posterior capsule of the lens. It occurs unilaterally in most cases. But we eliminated the diagnosis of posterior lenticonus due to biomicroscopy, retinoscopy and intraoperative findings. Usually in posterior lenticonus there is an appearance of an oil drop on retinoscopy [11]. Since posterior lenticonus may be associated with PCD and anterior hyaloid opacifications, it might be thougt that the membranes we had detected might be opacification of the anterior hyaloid. But anterior positioning of the membranes and the absence of any posterior protuberance of the lens indicate us that this may be a different abnormality.

Vasavada noted that a swift transformation of a previously nonmature congenital cataract to a white cataract, observed in conjunction with white crystalline particles floating in the anterior vitreous, is an indication of a preexisting posterior capsule defect [6]. Different from a fresh iatrogenic posterior capsule break, in a preexisting posterior capsule defect, there is a characteristic demarcation of thickened margins on the posterior capsule defect and there are white dots on the anterior vitreous face [6]. These findings were also observed in our cases; but in contrast to some previous reports, we did not observe 'fish tail' sign in our cases. Vasavada described this sign as an appearance of the anterior vitreous opacities moving like a fish-tail seen when the globe is moved with a forceps [6].

It was stated that male predominance and unilateral cataract were more frequent in PCD cases [6]. In our report, all of our patients were males but PCD was bilateral. In our cases, when the eyes were not dilated, the cataracts looked like totally white mature cataracts. But after pupil dilatation, we saw that the density of the opacity was greatest at the center, leaving the periphery transparent. Interestingly, the area of lens opacity was proportional with the place of PCD in all eyes.

In addition to previous reports, we saw membranous opacities at the place of PCD. In case 1 and case 2, these membranes were not attached firmly to the surrounding tissues and were located in the place of the absent posterior capsules; but in case 3 the membrane was firmly attached to the posterior capsule in only one side. Although the physical characteristics and appearances of the membranes seen in both cases were not identical, we think that they should be classified in the same disorder. In our opinion, the membranes might be remnant of the absent posterior capsule. We removed the membranes with vitreous cutter in case 1 and case 3 and with a forceps in case 2. We did not encounter any difficulty in removing these membranes. It would have been good if we had been able to send the 'membrane' from one of the cases for microscopic and pathological examination; but, we think that it was not much necessary since the location and appearance of these membranes indicated us that they should be posterior capsule remnants.

In all the operations, the cataractous lenses were removed with routine irrigation-aspiration method. Anterior vitrectomy was applied through limbal ports. In spite of the fact that PCD could be detected after the aspiration of the lens, we considered not to perform hydrodissection in these cases; since it may enlarge the posterior capsule defect. Despite the availability of adequate posterior capsular support, we did not implant intraocular lenses because of the young age of our patients in case 1 and case 2. It might be better if we implanted IOL's at the same session, but we performed IOL implantation at about one year later from the first operations in order to put exact diopters that would not necessitate IOL exchange in future. In case 3, IOL optic capture was done (haptics were in the sulcus, optic was in the bag). We did not encounter any complications intra- and post-operatively. Although contact lenses were better in aphakic visual rehabilitation;

we prescribed spectacles due to difficulties in contact lens application in smaller children and also the patients tolerated spectacles well. At the last follow-up, the central visual axis was clear in all eyes. This membrane formation did not cause a higher percentage of visual axis opacification; it might be due to performing a good anterior vitrectomy.

It would be said that these 3 cases of PCD might not add much to the known literature, as it is a well-described entity by Vasavada. But, while doing surgery, when a surgeon sees these membranes accompanying PCD, the suggestions and methods described in the text should help the surgeon in completing the operation without any delay and complications.

In conclusion, although bilateral congenital posterior capsular defects accompanying bilateral congenital cataracts share some similar features with each other; they differ in some aspects: presence of a membrane, type of cataract, appearance of anterior vitreous opacities and additional posterior capsular opacification. But management strategy and prognosis are similar in all subtypes.

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