

# Black Diaphragm Intraocular Lens Implantation in Patients with Aniridia

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## Abstract

**Purpose:** To review the management outcomes of black diaphragm intraocular (BDI) lens implantation in Arab patients with aniridia.

**Methods:** Patients with aniridia undergone BDI lens implantation at our institution between 2013 and 2014 were included. Uncorrected visual acuity (UCVA), manifest refraction, and best-corrected visual acuity (BCVA) were evaluated before and 6 months and yearly after BDI lens implant surgery until the last visit. Intra- and postoperative complications were noted.

**Results:** Our series comprised 14 patients (8 males) with aniridia. The median duration of follow-up was 30 months (25% quartile). Ocular parameters, refractive status, and vision were all significantly improved at the last follow-up compared to the preoperative values ( $P < 0.05$  for all comparisons). All patients reported a significant decrease in photophobia and glare. Postoperatively, 11 eyes (78%) gained 2 or more lines of UCVA. At the last follow-up, BCVA increased by 2 or more lines in all cases. Early postoperative complications included main wound leakage (one eye) and anterior chamber hyphema (one eye). Late ( $\geq 6$  months) complications included corneal decompensation (one eye), failed penetrating keratoplasty graft (2 eyes), and subluxation of a scleral fixated BDI lens (one eye). Surgical interventions performed to manage complications included penetrating keratoplasty in 2 eyes with corneal decompensation and failed graft (one each), and re-suturing of a subluxated intraocular lens (one eye).

**Conclusion:** BDI lenses seem to be a safe and effective iris prosthetic with intraocular lens combination surgery for patients with congenital or traumatic aniridia. Periodic evaluation and prompt management of complications are recommended.

**Keywords:** Aniridia; Black Diaphragm Intraocular Lens Implantation; Visual Disturbances

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## INTRODUCTION

Complete or partial aniridia can be congenital or secondary to trauma. The lack of iris tissue causes an increase in spherical and chromatic aberrations, resulting in photophobia and glare.<sup>[1]</sup> Persistent visual disturbances

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often result in a debilitating decrease in vision related quality of life. Hence, management of aniridia will benefit overall quality of life.

The procedures used for the management of aniridia can potentially reduce photophobia, hence increasing visual function. They include coloured contact lenses, corneal tattooing, corneal stromal implants, and black diaphragm intraocular (BDI) lenses.<sup>[2]</sup> Implantation of BDI lenses reduces peripheral corneal optical aberrations and the related visual symptoms.<sup>[3]</sup> Encouraging results, including improved cosmesis and visual acuity, have been reported with BDI lens implants.<sup>[4]</sup> However, postoperative complications of BDI lens implantation have been reported, such as the progression of glaucoma, corneal epithelial disorders, and cystoid macular oedema.<sup>[4,5]</sup> Numerous case series have discussed the advantages of the management of aniridia.<sup>[6-9]</sup> To the best of our knowledge, the management of aniridia in Arab patients has not been reported. This is a relatively unique population with aniridia because these patients typically reside in regions with bright sunlit conditions throughout the year. The current study presents the outcomes of BDI lens implantation in Arab patients with aniridia.

## METHODS

This retrospective chart review evaluated patients with aniridia who had undergone BDI lens implantation at our institution between 2013 and 2014. The study was approved by the hospital research ethics committee (IRB 1350-R). BDI lenses were implanted only in cases in whom intraocular pressure (IOP) had been controlled prior to considering surgery. Patients with a follow-up period of less than 6 months were excluded.

Detailed ocular examination performed prior to surgery included measurement of uncorrected visual acuity (UCVA), manifest refraction, best-corrected visual acuity (BCVA), slit lamp examination of the anterior segment, and applanation tonometry (Goldmann). The posterior segment was assessed with slit lamp biomicroscopy with a +20 D and +90 D Volk lenses. Patients underwent B-Scan ultrasonography of the posterior segment and specular microscopy of the cornea, as clinically warranted. The same ophthalmic examination was performed postoperatively.

All surgery was performed under standardized sterile conditions. An anaesthetist delivered peribulbar anaesthesia for all but one patient who received general anaesthesia. A lid speculum was inserted to maximize globe exposure. In phakic eyes, phacoemulsification was performed. After phacoemulsification and IOL insertion, the two capsular rings (Morcher type 50 C; Morcher GmbH, Stuttgart, Germany) were inserted in the capsular bag in aniridia segments.<sup>[10]</sup> An ophthalmic viscoelastic device was injected into the anterior chamber at the start of surgery and removed at the end of the procedure,

and the corneal wound was sutured. In aphakic eyes, a scleral fixation technique was used to secure the BDI lens. Different types of Morcher BDI lenses were implanted. All eyes were targeted for emmetropia using the Holladay II IOL formula. Patients used topical fluoroquinolone eyedrops 4 times daily for 2 weeks and prednisolone acetate 1% eyedrops 4 times daily. Topical steroid administration was tapered slowly over 4 weeks. Operative follow-up was performed at 1 day, 2 weeks, 6 weeks, 3 months, 6 months, and yearly.

A pretested data collection form was used to record the type of aniridia, visual acuity, IOP, topical glaucoma medications, and other surgical interventions. The median (minimum to maximum) was used for reporting UCVA. Preoperative and postoperative UCVA were compared with the Wilcoxon signed-rank test. A *P* value < 0.05 was considered statistically significant.

## RESULTS

The study sample comprised 14 patients with aniridia (6 females and 8 males). The mean age of the patients was  $26.43 \pm 7.31$  years (range, 14–41 years). The sample included 9 right eyes and 5 left eyes, of which 2 were cases of congenital aniridia and 12 were cases of traumatic aniridia. Eight eyes had complete aniridia and 6 had a partial iris defect. Twelve of the eyes were aphakic and 2 were phakic, both with cataracts. The 12 aphakic eyes had undergone previous surgical intervention, including primary repair in 3 eyes, repair of traumatic graft dehiscence in 5 eyes, complicated cataract surgery in 3 eyes, and therapeutic penetrating keratoplasty, pars plana vitrectomy, pars plana lensectomy, and iridectomy in one eye for fungal endophthalmitis. All patients complained of photophobia.

The following types of BDI lenses were implanted: 67 B (one eye), 67 G (2 eyes), 67 L (2 eyes), 67 F (2 eyes), and 68 (5 eyes). The median duration of follow-up was 30 months [25% quartile, 24 months (minimum 6 months and maximum 108 months)].

Data on vision and ocular status preoperatively and at last postoperative follow-up are compared in Table 1. Postoperatively, all patients reported a significant decrease in photophobia and glare. Ocular parameters, refractive status, and vision were all significantly improved at the last follow-up compared to the preoperative values (all *P*s < 0.05). Postoperatively, 11 eyes (78%) gained  $\geq 2$  lines of UCVA. At the last-follow up, BCVA increased by 2 lines or more in all cases.

No intraoperative complications occurred. Early postoperative complications included wound leakage, requiring re-suturing in one eye, and anterior chamber hyphema in one eye, which resolved spontaneously within 5 days with topical prednisolone acetate 1%. Late (6 months or later) postoperative complications included corneal decompensation in one eye, failed

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penetrating keratoplasty graft in 2 eyes, and subluxation of a scleral fixated BDI lens in one eye. Postoperative surgical interventions included penetrating keratoplasty in 2 eyes (corneal decompensation, failed graft) and re-suturing of a subluxated IOL in one eye. None of these 3 eyes exhibited any additional sequelae after the postoperative surgical intervention. The complete details on the management outcomes of late postoperative complications of BDI for aniridia have been summarized in Table 2.

**DISCUSSION**

In this small case series, we noted that the black iris diaphragm or BDI lens improved visual acuity and refractive status and reduced the visual disturbances in eyes with aniridia. Appropriate postoperative management of complications can ensure restoration of vision.

The incidence of congenital aniridia is 1 in 60,000.<sup>[11]</sup> Thus, the Saudi population could include as many as 325 cases of aniridia.<sup>[12]</sup> Aniridia following trauma has been documented; however, traumatic aniridia is rare and is caused by intraocular surgery in most cases.<sup>[7,13]</sup> These patients could also benefit from BDI lens implantation.

In the current study, vision improved significantly following BDI lens implantation and management of complications, indicating that this type of IOL are safe and effective in eyes with aniridia. These eyes are likely to have additional ocular comorbidities, including amblyopia, which could impede the attainment of maximal postoperative vision in some cases.<sup>[4]</sup>

In our series, the majority of eyes were aphakic and only 2 cases underwent cataract surgery and BDI lens implantation. Controversy remains regarding the use of clear lens extraction and IOL implantation in the

**Table 1. Vision, refraction and ocular status before and at the last follow up after black diaphragm implantation in eyes with aniridia**

	Number of eyes	Level	Before BDI implantation	After BDI implant	P
Uncorrected vision	14	Median	20/400	20/100	< 0.001*
		25% quartile	20/400	20/125	
		Minimum	Light perception	20/600	
		Maximum	20/100	20/50	
Best corrected vision	7 before and 14 after implantation	Median	20/50	20/30	< 0.001*
		25% quartile	20/80	20/40	
		Minimum	20/125	20/100	
		Maximum	20/30	20/20	
Intraocular pressure	14	Mean	18.6	17.8	0.002*
		Standard deviation	5.3	2.1	
Refractive error (spherical)	14	Median	8.0	-0.15	< 0.001*
		25% quartile	5.5	-1.0	
		Minimum	0.0	-2.0	
		Maximum	24.0	0.0	
Refractive error (cylinder)	8	Median	-3.0	-0.7	< 0.001*
		25% quartile	-4.0	-1.0	
		Minimum	-8.0	-7.5	
		Maximum	0.0	0.0	
Glaucoma medications	14	0	4	14	< 0.001*
		1	10	0	

BDI, black iris diaphragm, \*P<0.05 is statistically significant

**Table 2. Management outcomes of late postoperative complications of black diaphragm implantation for aniridia**

Complication	Time of complication after BDI implant (months)	Vision before management	Surgery	Vision after surgery	Duration of follow up after surgery (months)
Corneal decompensation	6	20/300 (UCVA)	Penetrating Keratoplasty	20/100 (UCVA)	2
failure of Graft done before BDI	12	2/200 (UCVA)	Penetrating Keratoplasty	20/50 (UCVA)	12
Subluxated IOL due to trauma	36	20/80 (BCVA)	Re-suturing of IOL	20/20 (BCVA)	17

BDI, black diaphragm intraocular; IOL, intraocular lens; BCVA, best corrected visual acuity; UCVA, uncorrected visual acuity

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management of high myopia.<sup>[14]</sup> In cases of aniridia, even a clear lens carries the risk of weak zonules and thinning of the lens capsule.<sup>[15]</sup> Hence, removal of the lens and IOL implantation are more favourable options.<sup>[15]</sup>

In the current study, the IOL power was calculated using the Holladay II IOL formula. Although distance emmetropia was targeted in all eyes, most of the cases had hyperopic spherical residual refractive errors with myopic astigmatism. This outcome may have been due to suturing, intraocular manipulation of the capsular bag, capsular ring insertion for sectorial aniridia, or inaccurate calculation of the IOL power. Additional studies are warranted to determine the causes of postoperative astigmatism and the methods necessary to address this issue.

None of the patients in the current study had postoperative glare or photophobia. However, these data were collected subjectively. Further investigation is required to obtain patient feedback using adequate survey instruments, including the addition of an objective measure of preoperative and postoperative glare, contrast sensitivity, and other visual disturbances.

In the current study, postoperative IOP did not vary significantly. None of the patients required medical therapy for elevated IOP postoperatively. Increased IOP is a known complication of BDI lens implant surgery.<sup>[3,6,7]</sup> Aslam et al reported that IOP increased on day 1 following surgery, reaching as high as 43 mmHg.<sup>[5]</sup> This could have been due to compression of the trabecular meshwork by the lens haptics.<sup>[7]</sup> It could also have been due to incomplete removal of ophthalmic viscoelastic device following BDI lens implantation. We recommend proactive management to avoid such IOP spikes because elevated IOP can affect the stability of the lens and the corneal wound.

In the current study, corneal decompensation occurred in one eye, which was successfully managed with penetrating keratoplasty. Corneal decompensation is a known complication of prosthetic iris implants.<sup>[3,16]</sup> Dong et al<sup>[8]</sup> reported that the long-term postoperative complications of BDI lens implantation were related to the severity of ocular damage prior to the surgery. They postulated that displacement or forward migration of the IOL causes corneal decompensation.<sup>[8]</sup>

Three eyes in the current study were successfully managed for late postoperative complications. Closer monitoring and understanding of the risk of intraocular manipulation at different stages of BDI lens implantation surgery may aid in the detection of complications and timely management could restore visual function.

A large series study conducted by Lockington et al<sup>[17]</sup> noted only one case of postoperative subluxation, out of 47 cases.

Our study had some limitations. Data on postoperative endothelial cell count were unavailable; however, clinical evidence of endothelial damage was not observed in these eyes. Because of the small number of eyes in our series, the outcomes could have been influenced by statistical errors and the results should be interpreted with caution.

In conclusion, BDI lens implantation seems to be a safe and effective method for patients with congenital or traumatic aniridia. The postoperative reduction in refractive error and the diminished visual disturbances benefit patients. Periodic evaluation and prompt management of complications are recommended.

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Nil.

**Conflicts of Interest**

There are no conflicts of interest.

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