African Vision and Eye Health
ISSN: (Online) 2410-1516, (Print) 2413-3183

Management of paediatric ectopia lentis at an Indonesian referral hospital: A retrospective review

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Dates:
Received: 20 Apr. 2022
Accepted: 01 June 2022
Published: 27 July 2022

How to cite this article:
Barliana JD, Yulia DE, Soeharto DA. Management of paediatric ectopia lentis at an Indonesian referral hospital: A retrospective review. Afr Vision Eye Health. 2022;81(1), a768. https://doi.org/10.4102/aveh.v81i1.768

Background: Ectopia lentis in children is rare and often associated with systemic conditions. Left untreated, it can lead to visually debilitating complications. Although there are various techniques available, no current consensus for treatment of paediatric ectopia lentis exists.

Aim: To describe the management and visual outcomes of paediatric ectopia lentis in an academic referral hospital.

Setting: This study was conducted at Cipto Mangunkusumo National Central General Hospital, Jakarta, Indonesia.

Methods: A retrospective review was conducted on the medical records of all paediatric ectopia lentis patients treated at our hospital, from 2011 to 2020. Primary outcomes include uncorrected visual acuity (UCVA), best-corrected visual acuity (BCVA), treatment, surgical technique, selection of intraocular lens (IOL), and postoperative complications.

Results: This study included 66 eyes of 39 patients. Average age at initial presentation was 7.8 (2–17) years. Aetiology was mostly non-traumatic, whilst four were secondary to trauma. All eyes except one underwent surgery, and IOLs were inserted in 57.4% of the non-traumatic cohort, with iris-claw IOL as the most popular choice (85.7%). Uncorrected visual acuity and BCVA significantly improved in pseudophakic and aphakic groups ($p < 0.05$). Nearly 60% achieved a BCVA of 6/12 or greater, and the proportion was higher amongst pseudophakic group. Ten eyes had post-operative complications, with a median onset of 393 (1–1095) days after surgery.

Conclusion: With varied techniques used to manage paediatric ectopia lentis, the choice of technique was dependent on individual patient condition and surgeon preference. Whilst the proportion of eyes with BCVA of ≥ 6/12 was higher amongst the pseudophakic eyes than aphakic eyes, IOL implantation entails the risk of complications, whereas no complications occurred in those left aphakic. Therefore, no particular technique can be considered marginally superior to the other, and larger studies are needed.

Keywords: ectopia lentis; children; pseudophakia; IOL; ophthalmic surgery.

Introduction

Ectopia lentis is a rare finding in children that pertains to a displaced crystalline lens because of weak zonules. Apart from trauma, ectopia lentis is often found in systemic disorders, such as Marfan's syndrome, homocystinuria, or Weil–Marchesani syndrome.[1] If untreated, complications can arise, including luxation to the anterior or posterior chamber, corneal endothelium damage, glaucoma, and retinal detachment.[2,3]

In treating paediatric ectopia lentis, surgery may not be needed if visual rehabilitation and maturation are achieved through a conservative approach.[3,4,5] However, a previous study reported ametropic amblyopia in 50% of patients despite maximal conservative management.[4] Indications for surgery include low best-corrected visual acuity (BCVA) or progressively severe subluxation.[3,4,5] Surgical intervention involves rendering the eye aphakic by removing the crystalline lens. Following this, visual rehabilitation presents the next therapeutic challenge. Options include aphakic spectacles, contact lenses, or intraocular lens (IOL) implants. Aphakic spectacles are often difficult for children to tolerate because of visual disturbances, including aniseikonia, prismatic distortion, and constriction of visual fields, and the discomfort in wearing spectacles themselves. Contact lenses rely on compliance of wear and pose a higher risk of epithelial erosion than IOLs. Good visual outcome is dependent on compliance with amblyopia therapy in the first several years of life. With this, intolerance towards contact lenses or spectacles indicates IOL implantation.[3,6,7]

Intraocular lens used in children include angle-supported anterior chamber intraocular lens (ACIOL), iris-fixated IOL, posterior-chamber IOL (PCIOL) centred with a capsular tension ring (CTR), or scleral-fixated IOL (SFIOL). Each technique has its strengths and drawbacks. Anterior chamber intraocular lens is the most conventionally used IOL but is often related to endothelial cell loss, corneal decompensation, and pupillary ectopia. Because of these problems, posterior-chamber SFIOLs are generally preferred over ACIOLs in aphakic children with inadequate capsular support for PCIOL implantation. Another viable option for eyes lacking capsular support is iris-claw IOLs, which can be implanted anteriorly or retropupillary.[4]

This study aimed to describe the management and visual outcomes of children with ectopia lentis in an academic referral hospital in Indonesia.

Methods

Numerical data were described with mean (standard deviation [s.d.]), or median (minimum-maximum), depending on normality according to the Kolmogorov–Smirnov test. Categorical variables were presented in frequency and percentage, *n* (%). Visual acuity was presented in decimals based on Snellen chart. Further analysis of data on surgery separated subjects into the following sub-groups: traumatic and non-traumatic ectopia lentis, as well as pseudophakia (IOL implant) and aphakia. Measure of association was reported as the difference between means (*p*-value) and s.d. with 95% confidence interval (CI). Data were inserted into Microsoft Office Excel v 16.48 and analysed using IBM SPSS (Statistical Package for the Social Sciences) version 26.

Ethical considerations

A retrospective descriptive analysis was done on the medical records of all paediatric patients with ectopia lentis treated at Cipto Mangunkusumo National Central General Hospital, Jakarta, Indonesia, from January 2011 to December 2020. Patients were excluded if medical records were inaccessible. This study was granted ethical approval by the Health Research Ethics Committee-University of Indonesia/Cipto Mangunkusumo Hospital (Protocol Number: 21-05-0517) and in accordance with the Declaration of Helsinki. Extracted data include age, gender, aetiology, laterality, uncorrected visual acuity (UCVA), best-corrected visual acuity (BCVA), treatment, surgeon, surgical technique, and selection of IOL. Postoperative UCVA and BCVA were recorded at each patient's last follow-up, and complications and onset were noted.

Results

This retrospective study included 66 eyes of 39 patients: 29 were bilateral and 10 were unilateral (Table 1). Average age at initial presentation was 7.8 ± 3.5 years, ranging from two to 17 years. Aetiology was mostly unspecified, with 65% having unconfirmed or unidentified underlying systemic diseases.

TABLE 1: Distribution of subject characteristics.

Characteristic	Value		
	n	%	Mean ± s.d.
Number of patients (eyes)	39	66	-
Laterality			
Bilateral	29	74.4	-
Unilateral	10	27.5	-
Age at presentation (years)	-	-	7.8 ± 3.5
Gender			
Male	22	56.4	-
Female	17	42.5	-
Etiology			
Unspecified/unconfirmed	26	66.7	-
Marfan syndrome	8	20.5	-
Trauma	4	10.3	-
Homocystinuria	1	2.6	-
Duration of follow-up (months)	-	-	16.5 ± 21.3

s.d., standard deviation.

TABLE 2: Surgery on non-traumatic ectopia lentis (*n* = 61).

Characteristic	*n*	%
Surgical technique		
IOL implant	35	57.4
Primary	27	77.1
Secondary	7	20.0
CTR	1	2.9
Without IOL implant	26	42.6
ICCE with vitrectomy	25	41.0
ICCE without vitrectomy	1	1.6
Type of IOL		
Iris claw IOL	30	85.7
SFIOL	4	11.4
PCIOL	1	2.9

IOL, intraocular lens; CTR, capsular tension ring; ICCE, intracapsular cataract extraction; SFIOL, scleral-fixated IOL; PCIOL, posterior-chamber IOL.

Nine patients had confirmed systemic diseases: eight patients with Marfan's syndrome and one with homocystinuria as confirmed by the paediatrics department. Four patients were secondary to trauma. Average duration of follow-up was 16.5 ± 21.3 (0.5–96) months.

There were 62 eyes with non-traumatic ectopia lentis (Table 2). Only one eye did not undergo surgery as the patient tolerated conservative management with spectacles and achieved BCVA of 6/7.5. Overall average age at surgery was 7.9 ± 3.3 (3–17) years. Intraocular lenses were inserted in 35 eyes (57.4%) whilst 26 eyes (42.6%) were left aphakic. Average age at surgery was 8.3 ± 2.8 (5–14) years for pseudophakic eyes, and 7.3 ± 3.9 (3–17) years for eyes left aphakic. Amongst the 25 patients left aphakic, 24 eyes (96.2%) underwent intracapsular cataract extraction (ICCE) and anterior vitrectomy, whilst one eye underwent ICCE without anterior vitrectomy.

Amongst 35 pseudophakic patients, the majority had primary IOL insertion (27 eyes, 77.1%), where IOL implantation was done in the same procedure as ICCE and anterior vitrectomy. Seven eyes (20.0%) underwent secondary IOL insertion, where ICCE and anterior vitrectomy were conducted in the first surgery, followed by a separate procedure for secondary IOL implantation. One eye underwent irrigation aspiration and CTR with IOL implantation. Iris-claw IOL was the most

TABLE 3: Surgery in traumatic ectopia lentis (n = 4).

Characteristic	Value			
	n	%	Mean ± s.d.	Range
Age at operation (years)	-	-	6.0 ± 2.9	2–9
Surgery technique				
Secondary iris-claw IOL implant	1	25	-	-
Lensectomy + anterior vitrectomy	2	50	-	-
Irrigation aspiration, synechiolysis, cataract extraction, PPL + anterior vitrectomy	1	25	-	-

IOL, intraocular lens; PPL, pars plana lensectomy; s.d., standard deviation.

TABLE 4: Ocular parameters in non-traumatic ectopia lentis.

Characteristic	Value		
	n	Mean ± SD	Range
Preoperative UCVA	50	0.05 ± 0.05	0.0–0.15
Postoperative UCVA	39	0.2 ± 0.2	0.03–0.5
Preoperative BCVA	26	0.3 ± 0.2	0.02–0.8
Postoperative BCVA	54	0.6 ± 0.3	0.2–1.0

UCVA, Uncorrected visual acuity; BCVA, Best corrected visual acuity; SD, standard deviation.

popular IOL choice (30 eyes, 85.7%), followed by SFIOL (four eyes, 11.4%) and PCIOL (one eye, 2.9%).

There was a minority of traumatic ectopia lentis in this study, with four eyes of four patients (Table 3) and all eyes underwent surgery. Average age at surgery was 6.4 ± 2.7 years. Surgical technique was variable: one eye (25%) had secondary iris-claw IOL implant, two eyes (50%) had lensectomy with vitrectomy, and one of these eyes also underwent endolaser. Finally, one eye (25%) had three separate surgeries: irrigation-aspiration of coagulum and fibrin, followed by synechiolysis and cataract extraction, and lastly, the eye was fitted with a rigid gas permeable lens.

Visual acuity of the non-traumatic cohort was compared between baseline and after surgery (Table 4). Most eyes had improved uncorrected visual acuity (UCVA) after IOL implantation, with average postoperative UCVA (0.2 ± 0.2) higher than its baseline preoperatively (0.05 ± 0.05), and this was statistically significant ($p < 0.05$) in both aphakic and pseudophakic groups. However, three eyes that were left aphakic (4.8%) had UCVA that did not improve, where two eyes had unchanged UCVA, and one eye experienced a decrease in UCVA. Out of 61 eyes, comparison between UCVA could only be done in 34 eyes because of incomplete data.

Improvement between mean preoperative BCVA (0.3 ± 0.2) and mean postoperative BCVA (0.5 ± 0.3) was also found in the non-traumatic cohort, and this was also statistically significant ($p < 0.05$) in both aphakic and pseudophakic groups. Only one eye did not improve in BCVA, and this patient had pre-existing esotropia. Out of 61 eyes, comparison between BCVA could only be done in 26 eyes because of incomplete data.

Moreover, 32 (59.3%) of the eyes in this cohort achieved BCVA of 6/12 or greater, 23 eyes (71.9%) of which received IOL implant. The proportion of eyes with BCVA greater than or equal to 6/12 was higher amongst the pseudophakic group (23 eyes, 72.0%) than the aphakic group (nine eyes, 40.9%).

Comparison between preoperative and postoperative visual acuity was also done in the traumatic ectopic lens group. Average postoperative UCVA (0.008 ± 0.00) was not higher than its baseline preoperatively (0.01 ± 0.008). Average postoperative BCVA (0.2 ± 0.3) was slightly higher than its baseline preoperatively (0.13 ± 0.1). Of the three eyes that were analysed, one eye experienced an improvement in BCVA, whilst two eyes did not. Because of the limited sample size and missing data, proper statistical analysis could not be done.

From the 65 eyes in this study that underwent surgery, 10 eyes (15.4%) had postoperative complications, with a median onset of 393 (1–1095) days after surgery. Four eyes were that of patients with Marfan's syndrome. Almost all complications occurred in the eyes with iris-claw IOL, except for one eye that had PCIOL. There were no post-operative complications in those who were left aphakic.

Intraocular lens de-enclavation occurred in four eyes (6.2%), with a median onset of 421 days (393–730 days). Two cases were immediately re-enclavated; another was managed by explanting the iris-claw IOL and switching to an SFIOL, whilst one case was left for observation. All resulted in good visual outcome.

Elevated intraocular pressure (IOP) occurred in three eyes (4.6%), with a median onset of 14 days (1–14 days). One case of elevated IOP one day after surgery was immediately treated with peripheral iridectomy. Another case was treated with glaucoma medication. Both cases had good visual outcomes. One case developed secondary glaucoma with severe inflammation two weeks after surgery and was treated with glaucoma medication, steroids, and antibiotics, and the eye recovered within a week.

Retinal detachment was reported in one eye three years after iris-claw implant and was treated with endolaser and silicone oil. Visual axis opacity was reported in one case 20 months following PCIOL implant and was treated with laser. Another eye was reported to have vitreous bleeding and corneal imbibition, which developed into phthisical eye in one week; treatment involved antibiotics and corticosteroid eyedrops. Endophthalmitis was found in one eye three weeks after surgery and was identified to have occurred because of external factors. This patient was treated with intravitreal antibiotics.

Discussion

To date, no consensus has been established for the best management of ectopia lentis in children, as treatment plans are typically made on a case-by-case basis. In this study, nearly all eyes underwent surgery, except for one patient of the non-traumatic cohort. This patient was able to achieve good visual outcome through spectacle correction, and thus, conservative management without surgery was deemed adequate, which is in line with previous recommendations.[3,4] In eyes that underwent surgery, nearly 60% had IOL implantation. Average age at first surgery amongst those left aphakic was slightly

younger and consisted of a wider range than the IOL group (3–17 years vs 5–14 years). Intraocular lens implantation in younger children is less recommended because of immaturity and inflammation at infancy. Despite the study population consisting of relatively older children who were suitable for IOL implantation, the choice of technique was not determined solely by age; IOL implant was not required in children who tolerated and achieved acceptable BCVA with conservative correction.[8,9] Amongst the pseudophakic eyes, nearly 78% underwent primary IOL implantation. Previous literature recommended primary IOL implantation if patients were at least 2–3 years of age, whilst those younger were recommended to be first left aphakic and corrected conservatively.[5,10] Other considerations in delaying IOL implant include elevated risk of retinal detachment associated with Marfan's syndrome, which may be exacerbated by lensectomy. Also, axial myopia is a common feature in Marfan's, in which case leaving them aphakic might be preferable.[11]

Iris-claw IOL seems to be preferred in our referral centre because of its relatively simple technique and faster operation time whilst delivering equally good outcomes compared to other alternatives. However, iris-claw is associated with potential corneal endothelial cell decompensation, and there have been reports of de-enclavation occurring often.[12,13,14,15,16] Unlike this study, SFIOLs are relatively more popular in current literature in the paediatric population, and its placement is considerably more physiological; however, complications, such as hyphema and vitreous haemorrhage, are common as scleral fixation requires direct involvement of vascular tissue, albeit such complications were not found in the one patient in this study who had SFIOL.[4]

This study demonstrated an overall good visual outcome, with a statistically significant improvement in UCVA and BCVA after surgery, with or without IOL implantation. Good visual outcomes in paediatric ectopia lentis patients who underwent surgery with or without IOL implantation have been shown in previous studies.[13,15,16,17,18,19] In the non-traumatic cohort, 59.32% achieved BCVA of 6/12 or greater, the proportion of which was higher amongst pseudophakic eyes (72.0%) in comparison to aphakic eyes (40.9%). Contrastingly, previous studies have exemplified that 70.0% – 100.0% of aphakic patients from primary lensectomy achieved 6/18 or better, and as high as 93.0% achieved 6/12 or better.[4,7,20] Previous studies stated that compliance towards visual rehabilitation is critical in optimal correction[6]; the less optimal visual outcome reported in this study may be explained by the lack of individual compliance towards spectacle use in the aphakic patients.

Comparable rates of pseudophakic eyes with good visual acuity were reported in previous studies, with 71% and 80% of the respective cohorts with retropupil iris-claw that achieved BCVA of 6/12 or greater.[5,21] Studies on patients who received SFIOL reported slightly lower proportions, with 39.0%, 52.3%, and 61.6% who had BCVA greater or equal to 6/12.[6,21,22] Epley et al. notably conducted a subgroup analysis in patients left aphakic after 9 years of age, thereby after the amblyopic age range, and revealed a much higher proportion of 83% that achieved BCVA of 6/12 or greater.[6] Similarly, this study's finding of a larger proportion of pseudophakic eyes that achieved at least 6/12 visual acuity may be attributed to the cohort of older children.

Ten eyes in this study experienced post-operative complications, with a median onset of 393 days. Almost all complications occurred in the eyes with iris-claw IOL, with the exception of one eye with PCIOL. This higher rate in the former group may be attributed to 87.5% of the pseudophakic patients being implanted with iris-claw IOL.

Intraocular lens de-enclavation occurred in four eyes with iris-claw IOL, with onset from surgery ranging from 393 to 730 days. Onset of iris-claw IOL de-enclavation after surgery varies widely between studies, ranging from one day to 84 months post-operatively.[1,15,16,17,18,23] Whilst most of this study's cases immediately underwent re-enclavation, one case had IOL explantation and exchange, and this process was similarly reported in Gonnermann et al., where they described a case of retropupil iris-claw IOL dislocation into the anterior chamber, which required explantation and IOL exchange.[24] Intraocular lens detachment in SFIOL was also reported by Sen et al. in 6.8% of their cohort with an average onset of 8.15 (3–14) years.[22]

Elevated IOP occurred in three eyes, on average occurring after two weeks. This rate is comparable with the findings from previous studies, which reported three eyes after SFIOL implant.[22] Two studies on anterior iris-claw each reported one case of glaucoma, which required peripheral iridectomy that occurred at three days and five months post-surgery, respectively.[17,18] Similarly, studies on retropupil iris-claw reported one case that required trabeculectomy,[21] one that was transient,[25] and one that was controlled within a week with topical timolol and oral acetazolamide.[23] Furthermore, one study stated that acute angle-closure glaucoma following anterior iris-claw IOL implantation occurred in 0.0% – 7.0% of patients, lower than that in secondary scleral-fixated PCIOL implantation (0.0% – 30.7%).[26]

Visual axis opacity was reported in one case 20 months following PCIOL implant using CTR. Surgeons generally avoid performing primary posterior capsulotomy and CTR in the same procedure because of its technical challenge. Therefore, one of the most common complications following paediatric cataract surgery with CTR is visual axis opacification which requires a second surgery.[8] Another study mentioned posterior capsule opacification requiring intervention occurred in 83.8% of patients (31 out of 37) during the median follow-up of 27 months.[4]

Retinal detachment was reported in one eye three years after iris-claw implant. This patient was suspected of having Marfan's syndrome, in which retinal detachment is a common finding. Fan et al.'s study reported 17.2% of their cohort of anterior iris-claw in adults and children with Marfan's

syndrome eyes experienced retinal detachment 1–48 months after surgery.[10] Another study reported one case of retinal detachment secondary to severe blunt trauma to the eye.[16] Retinal detachment is also found in cases of SFIOL, where it was observed to occur on average 17.3 months after surgery.[22]

Other complications found in the patients of this study include vitreous bleeding and corneal imbibition. Other studies on SFIOL reported five eyes with transient anterior uveitis less than 1 month after surgery,[27] and another reported one eye with uveitis-glaucoma-hyphema syndrome.[28] Sen et al. reported one eye after SFIOL implant with uncontrolled glaucoma that developed ciliary staphyloma and progressed to hypotonic maculopathy. This eye was treated with a scleral patch graft; however, this failed, and the eye became phthisical. The same study reported two eyes with vitreous haemorrhage and hyphema, and both were treated conservatively.[22] Acute inflammation post-surgery, marked by anterior chamber cells, was seen in 60% of patients in both iris-claw and SFIOL groups in a study on adults, and reported one patient with uveitis after SFIOL implantation.[29]

In the present study, ectopia lentis secondary to trauma on average, did not experience much improvement in visual acuity after surgery. Albeit only one eye had an IOL inserted, and the issue of limited sample size prohibits proper statistical analysis. A study by Gawdat et al. which discussed seven post-traumatic patients reported that visual acuity increased and remained stable within a year of follow-up after anterior iris-claw implants.[17]

Limitations of this study include the wide variety of techniques used in the management of paediatric ectopia lentis. Because the number of patients that underwent each technique was not equal, proper statistical analysis between techniques could not be properly conducted. However, including all the different surgical techniques was considered necessary to accurately portray the many potential approaches one can use in the treatment of paediatric ectopia lentis. This referral centre's choice of technique varied according to each surgeon's preference. Another limitation is the missing data, as visual acuity could not be measured in uncooperative patients. Another limitation is that the aetiology in most patients was unspecified. Although underlying Marfan's was often suspected in these patients, proper diagnosis could not be made in some of these patients. As a result, the researchers were unable to truly evaluate the risk entailed in patients with Marfan's, such as the inherent risk towards retinal detachment and progressive refractive error.

Conclusion

In conclusion, this study reports the varied techniques used to manage paediatric ectopia lentis in an Indonesian national referral centre, where choice of technique was highly dependent on each patient's condition and surgeon preference. Iris-claw IOL was the most common choice, and these patients had good visual outcome. Pseudophakic eyes had a higher proportion of eyes with BCVA greater than or equal to 6/12 in comparison to aphakic eyes. However, pseudophakic eyes entail risk of complications, whereas no complications occurred in those left aphakic. Therefore, not one technique can be considered as marginally superior to the other, and larger studies are needed.

Acknowledgements

Competing interests

The authors declare having no competing interests and no financial or personal relationships that may have inappropriately influenced the writing of this article.

Authors' contributions

J.D.B. and D.E.Y. contributed to the initial concept and idea. J.D.B., D.E.Y., and D.A.S. contributed to the design, literature search, data acquisition, data analysis, statistical analysis, and all three authors discussed the results and contributed to manuscript preparation, editing, and manuscript review. J.D.B. and D.E.Y. supervised the entire process. J.D.B. served as guarantor.

Funding information

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data availability

Data that support the findings of this study are available upon request from the corresponding author, J.D.B. The data are not publicly available at this time because of human data that can compromise the privacy of research participants.

Disclaimer

Views expressed in this submitted article are that of the authors own and do not necessarily reflect the official position of the institution affiliated with the authors.

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