Original Research Article

Visual outcome of pars plana vitrectomy with lensectomy with scleral fixated IOL implantation in ectopia lentis

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Abstract

Background: Patients with ectopia lentis present with low visual acuity, diplopia, pain. Cataract surgery with normal in the bag posterior chamber intra ocular lens (PCIOL) surgery for these patients is difficult due to weak capsular support. In traumatic cases, sometimes vitreous is seen in anterior chamber, these patients need vitrectomy with removal of lens and implantation of scleral fixated PCIOL. **Aim:** To evaluate visual outcome of pars plana vitrectomy with lensectomy with scleral fixated IOL implantation in ectopia lentis. **Material and Methods:** This prospective study included 50 eyes of 45 patients with ectopia lentis. All patients underwent lensectomy with 3 port pars plana vitrectomy with scleral fixated IOL implantation. The scleral fixated posterior chamber intraocular lens (SF-PCIOL) used were AUROLAB. Patients evaluated on post-operative 1, 7, 30 and 90 days. **Results:** 43 eyes (86%) had final best corrected visual acuity better than 6/24 and 46 eyes (92%) had BCVA 6/36 and better. Secondary glaucoma(10%), anterior uveitis (6%), vitreous hemorrhage(4%) and CME(2%) were found to be few of the complications. Patients had post-operative average refractive error of -0.78 sphere and-1.245 cylinder at 3 month follow up. **Conclusion:** Lensectomy with pars plana vitrectomy and scleral fixated IOL implantation is a safe and effective treatment for ectopia lentis with inadequate capsular integrity

Key Word: Ectopia lentis, Lensectomy with pars plana vitrectomy, scleral fixated IOL implantation, visual outcome, complications

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INTRODUCTION

Human lens constitutes one of the most important refractive media in eye. Proper development of lens and appropriate position in eye allows focusing of refractive rays on retina and achieve good visual acuity. Change in lens position due to incomplete development or due to trauma resulting in ectopia lentis can result in severe deterioration of vision.[1] Trauma is the most common cause of acquired lens displacement. Non traumatic ectopia lentis is commonly associated with Marfan syndrome, homocystinuria, aniridia, and congenital glaucoma. Less frequently, it appears with Ehlers-Danlos syndrome, hyperlysinemia, and sulfite oxidase deficiency.[2] Patients with ectopia lentis present with low visual acuity, diplopia, pain (due to raised intraocular pressure), these patients have weak zonular support with ectopic and weak capsular bag.[3] Cataract surgery with normal in the bag posterior chamber intra ocularlens (PCIOL) surgery for these patients is difficult due to weak capsular support. In traumatic cases, sometimes vitreous is seen in anterior chamber, these patients need vitrectomy with removal of lens and implantation of scleral fixated PCIOL.[4] The present study was conducted to evaluate visual outcome of pars plana vitrectomy with lensectomy with scleral fixated IOL implantation in ectopia lentis.

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MATERIAL AND METHODS

This prospective study included 50 eyes of 45 patients with ectopia lentis. Written informed consent was obtained from every patient. Approval from Institutional ethical committee was taken prior to the commencement of the study.

Inclusion criteria

- patients with ectopia lentis above 2 years of age with inadequate capsular support
- patients of congenital or acquired ectopia lentis
- patients undergoing lensectomy with pars plana vitrectomy with scleral fixated IOL
- implantation

Exclusion criteria

- Patients with ectopia lentis undergone phacoemulsification or SICS with CTR
- Patients having traumatic retinal detachment, optic neuropathy and otherco-existing retinal pathology.
- Patients with globe perforation

Patients were evaluated for history of trauma, systemic illness, similar complaints in parents or siblingsand duration of vision loss. Examination was done by visual acuity with best correction /aphakic correction, slit lamp examination, indirect ophthalmology, Bscan (if indicated) and intraocular pressure by non-contact tonometry. Systemic examination was done in view of suspected Marfan's syndrome features inpatient and signs such as height/arm span, wrist sign, thumb sign were observed. CVS examination was done. Patients were diagnosed according to Ghent's criteria.46 Investigations like Hb, CBC, Blood sugar, RFTs, ECG were done. Special investigations like IOL power calculation by IOL master, 2D echo, homocysteine level if indicated were also done. All patients underwent lensectomy with 3 port pars plana vitrectomy with scleral fixated IOL implantation. The scleral fixated posterior chamber intraocular lens (SF-PCIOL) used were AUROLAB. Single piece polymethyl meth acrylate (PMMA) with modified C haptics with at least 2 dialling holes in the haptics, with an optic diameter of at least 6.5mm, with over all diameter 13.5mm. Suture used were 10-0 polypropylene suture with double armed straight needles, 10-0nylon suture with micro point spatulated needle. Patients evaluated on post-operative day 1, 7, 30 and 90 in terms of visual acuity on Snellen chart on every visit, auto-refraction on day 90, refraction and best corrected visual acuity on day 90, Slit lamp examination, Intraocular pressure measurement with non-contact tonometer, Indirect ophthalmoscopy. All patients were given tablet ciprofloxacin 500 mg twice daily and tablet ranitidine 150 mg twice daily for 5 days started on 1 day prior to surgery. Injection dexamethasone 8 mg for adults and according to weight in children given intramuscular on a day of surgery. Locally in operated eye, eyedrop containing combination of of loxacin and prednisolone acetate instilled in tapering dose till 5 weeks, atropine eyedrop twice a day 5 weeks and anti-glaucoma medications given if IOP was raised.

RESULTS

A total of 50 eyes of 45 patients between the ages of 0 to 80 yrs with ectopia lentis undergoing lensectomy + pars plana vitrectomy with SFIOL. Out of 45 patients, 37 (82.6%) were males and 8 (17.4%) were female with a male to female ratio of 4.6:1.Ages ranging from 7yr to 78yrs with an average age of 41.6yrs with 70% of patients within the age group of 20-60yrs. Out of 45 patients (50 eyes), 5 patients (10 eyes) had Marfan's syndrome, 40 eyes were due to Trauma. Out 40 eyes of traumatic ectopia lentis, pre-operatively, 24 eyes (60%) hadvisual acuity of <6/60, 12 eyes (30%) had visual acuity of 6/60 to 6/36, and 4 eyes(10%)had visual acuity of 6/24 to 6/18. After 3 months of surgery, 1 eye (2.5%) had visual acuity <6/60, 5 eyes (12.5%) had visual acuity 6/60-6/36, 9 eyes(22.5%) had visual acuity 6/24 to 6/18, and 25(62.5%) eyes had visual acuity of 6/12 or better.

Table 1: Visual acuity in traumatic ectopia lentis

Visual acuity	Pre-op	Post-op at 3 month
<6/60	24(60%)	1(2.5%)
6/60-6/36	12(30%)	5(12.5%)
6/24-6/18	4(10%)	9(22.5%)
6/12-6/6	0	25(62.5%)

Out of 10 eyes of Marfan's syndrome pre-operatively, 4 eyes(40%) had visual acuity of <6/60, 3 eyes(30%) had visual acuity 6/60 to 6/36, and 3 eyes(30%) had visual acuity of 6/24 to 6/18.After 3 months of surgery, 1 eye (10%) had visual acuity of 6/36, 1 eye (10%)had visual acuity 6/24, 8 eyes (80%) had visual acuity 6/12 or better.

Table 2: Visual acuity in Marfan's syndrome cases

Visual acuity	Pre-op	Post-op at 3 month
<6/60	4 (40%)	0
6/60-6/36	3 (30%)	1(10%)
6/24-6/18	3 (30%)	1(10%)
6/12-6/6	0	8(80%)

Out of 50 eyes, 42 eyes had cataractous dislocated lens and 8 eyes had non cataractous lens. All traumatic (38) cases had cataractous lens changes. 4 eyes (2 patients) out of 10 eyes (5 patients) of Marfan's syndrome had cataractous lens changes and 6 eyes (3 patients) had non-cataractous lens.

Table 3: Lens opacity

	Traumatic	Marfan's syndrome	Total
Cataractous	40 (80%)	4 (8%)	44 (88%)
Non- cataractous	0	6 (12%)	6 (12%)
Total	40 (80%)	10 (20%)	50 (100%)

Out of the 50 eyes who underwent the procedure, 28 (56%) eyes showed a VA of <6/60 pre-operatively compared to only 2 eyes (4%) having vision <6/60 with 37 eyes (74%) having a VA of 6/24 or better at the end of 1 week postoperatively. At the end of 1st week, 16(32%) patients had a VA better than 6/12 and only 2(4%) patients had VA poorer than 6/60 at the end of 1 week compared to 28 patients (56%) who had VA poorer than 6/60 pre-operatively. At one week post-operative, we observed of 28 (56 %) patients who had visual acuity <6/60 pre-operatively, 5 (10%) patients improved to 6/12-6/6, 10 (20%) patients improved to 6/24-6/18 and 11 (22%) patients had BCVA of 6/60-6/36. Only 2 (4%) patients had vision deterioration or similar vision as pre-operative. In 15 (30%) patients with pre-operative vision 6/60-6/36, 9 (18%) patients improved to 6/24-6/18 and 6 patients (12%) improved to 6/12-6/6. Out of 7 (14%) patients with preoperative vision of 6/24-6/18, 2 patients remained 6/24-6/18 and 5 (10%) patients improved to 6/12-6/6. At the end of 1 month, 31(62%) patients had a VA better than 6/12 and only 2(4%) patients had VA poorer than 6/60 at the end of 1 week compared to 28 pts(56%) who had VA poorer than 6/60 pre-operatively. 6 Patients (12%) regain vision 6/60-6/36 and 11 (22%) patients improved to 6/24-6/18. In our study, at the end of 1 month, we found out of 28 patients with preoperative vision <6/60, only 2 remained <6/60. 6 (12%) patients improved to 6/60 -6/36, 8 (16%) patients improved to 6/24 - 6/18. Out of 15 patients (30%) pre-operative vision 6/60-6/36, 3 (6%) and 12 (24%) patients improved to 6/24-6/18 and 6/12-6/6 respectively. All 7 patients (14%) with preoperative vision 6/24-6/18 improved to 6/12 -6/6 after a month post operatively. One (2%) patient remained with vision <6\60 due to preexisting corneal opacity. 6 (12%) improved to 6\60 -6\36, while 3 patients remained with vision 6/60 due to preexisting nebular corneal opacity while 10 (20%) patient improved to 6\24 –6\18, 33 (66%) patients improved to 6 \12 -6\6. At the end of 3 months, 33(66%) patients had a vision better than 6/12. Out of 28 (56%) patients with preoperative visual acuity <6\60, 27 (54%) patients improved to >6\60 and only 1 patient remained with vision<6\60 (due to preexisting nebulomacular corneal opacity), 3 patients with 6\60 vision had preexisting nebular corneal opacity. Out of 15 patients (30%) patients with vision 6\60 -6\36, 3 (6%) patients improved to 6\24-6\18 and 12 (24%) patients gained vision >6\12 and all 7 (14%) patients with pre-opearative vision 6\24 - 6\18 improved to 6\12- 6\6. 3 eyes of 2 patients with Marfan's syndrome had preexisting amblyopia, so didn't show much improvement in BCVA. In our study, patients had average refractive error of -0.78 sphere and -1.245 cylinder. Minimum refractive error of -0.25 sphere and -0.75 of cylinder was observed. Maximum refractive error of -2.0 sphere and -3.5 cylinder was observed. Out of 50 eyes 5(10%) had raised IOP initially, managed with anti-glaucoma medication but 2 (4%) remained with IOP at the end of 3 months, advised anti-glaucoma medication lifelong. Other complications include Anterior uveitis in 3 eyes(6%), vitreous hemorrhage in 2 eyes(4%), IOL decentration in 2 eyes(4%),CME in one eye(2%). All of those resolved on treatment except 2 raised IOP. None of the patients in our study developed endophthalmitis, retinal detachment.

DISCUSSION

Management of ectopia lentis includes medical and surgical management. Surgical management includes lensectomy with CTR ring, ACIOL, iris supported claw lenses and SFIOL for placement of lens. Pars-plana vitrectomy + lensectomy+ SFIOL is a standard procedure for ectopia lentis. Indications for surgery include visual acuity less than 6/18, monocular diplopia, forward sub-luxation of the lens to the anterior chamber, or rapidly progressing posterior subluxation of the lens. However, the procedure is associated with certain complications such as dropped lens fragments, retinal detachment and vitreous haemorrhage. The procedure may be difficult due to increased mobility of the lens while performing lensectomy. The technique of Pars Plana Vitrectomy with SFIOL helps to create an attachment of the ectopic lens to the ciliary sulcus through scleral fixation sutures, thereby reducing the chance of dropped lens or lenticular fragments during lensectomy. By manipulating the sutures, the lens is made more visible in the pupillary area, aidingthe process of lensectomy and avoiding accidental damage of the pupil, ciliary body and retina. Further, preplaced scleral sutures facilitate subsequent fixation of an IOL. In present study, out of 50 eyes 10 eyes were of marfanoid patient and 40 had given history of trauma. Preoperatively 28 eyes (56%) Visual acuity of less than 6\60, 15 eyes(30%) had VA between 6\60 to 6\36, 7 eyes(14%) had VA between 6/24 to 6/18.At the end of 3 months, all of the patients had improved visual acuity as compared toprevious visual acuity. 33(66%) patients had a vision better than 6/12. Out of 28 (56%) patients with preoperative visual acuity <6\60, 27 (54%) patients improved to>6\60, and only 1 patient remained with vision <6\60 (due to pre-existing nebulomacular corneal opacity), 3 patients with 6\60 vision had preexisting nebular cornealopacity. So, 46 eyes(92%)

improved to visual acuity better than 6/36 ($p<0.001$). This shows improvement in visual acuity after a month is statistically highly significant. Gradual improvement in vision with in a firs tweek post-operative data about 8 patients having low vision compared to pre-operative vision with only 4 (8%) patients with vision on third month follow up. All these 4patients had pre-existing corneal opacity because of visual improvement was not above the line of significance. Out of 10 eyes of Marfan's syndrome in our present study, 3 eyes were amblyopic and hence did not improve above 6/18.In our study, patients had average refractive error of -0.78 sphere and -1.245cylinder.Minimum refractive error of -0.25 sphere and -0.75 of cylinder was observed. Maximum refractive error of -2,0 sphere and -3.5 cylinder was observed. Out of 50 eyes of 45 patients operated,5 patients had raised IOP compared to preoperative IOP which was managed with anti-glaucoma topical eye drops (Timolol0.5%or brimonidine0.2%) and acetazolamide 250 mg twice daily, but 2 (4%) patients remained persistent with raised intra ocular pressure despite of anti-glaucoma drops and had to continue treatment long term basis. These patients evaluated further by perimetry and OCT disc for RNFL thickness and further continued with anti-glaucoma medications in operated eye for lifelong and advised follow up every month. On first week follow up 3 patients out of 50 eyes found to have anterior uveitis on slit lamp examination these patients were treated with of loxacin+prednisolone eyedrops 1hourly for first week, then gradually tapered over 5 weeks and uveitis resolved on a month follow up. 2 (4%) patients had low vision on first week follow up and on examination found to have diffuse vitreous hemorrhage which was mostly due to trauma to ciliary body while taking suture to fix SFIOL. These patients further evaluated serially by BSCAN for condition of posterior segment at each visit and observed. Vitreous hemorrhage resolved on its own in a time period of a month. 2 (4%) patients on 1st week follow up had found to have IOL decentration but patient remained with good visual acuity with refractive correction. These patients were evaluated by UBM for position of haptics and no active intervention was done for same. One patient on a month follow up came with complain of decreased visual acuity. On slitlamp biomicroscopy, with 78D lens observed to have CME which was confirmed on OCT and treated with topical Nepafenac eye drop thrice daily and steroids. CME completely resolved on 3rd month follow up. Johnston RL *et al* found on mean follow-up of 20 months that preoperative best-corrected visual acuity was 20/40 or better in 36% (23 of 63 eyes), 20/60 to 20/120 in 33% (21 of63 eyes), and 20/200 or worse in 31% (19 of 63 eyes) improving to 20/40 or better in76% (48 of 63 eyes), 20/60 to 20/120 in 18% (11 of 63 eyes), and 20/200 or worse in6% (4 of 63 eyes) at final follow-up. Mitra *et al* in their study observed that at the end of 1 month VA was 6/6 in 5cases (25%) and 6/12 in 7 cases (35%). 4 bilateral congenital cases had amblyopia with6/18 vision in 4 cases (20%) and 6/36 in 4 cases (20%). No reduction in the visual acuity was noted at the six-month follow-up examination.[5]Omulecki *et al* did Pars plana vitrectomy technique combined with the outside-in method for scleral fixation of PC IOL on Forty eyes of 36 patients, and found the best corrected postoperative visual acuity was between 6/6 and 6/8 in 32 eyes, between 6/12and 6/18 in 5, and between 6/24 and 1/60 in 3 eyes. They observed one case of IOLdecentration.[6]Chaudhry *et al* studied 28 eyes which underwent pars plana lensectomy (PPL), parsplana vitrectomy (PPV), and scleral fixation of posterior chamber intraocular lens forclosed globe injuries and observed out of these 28 eyes, 19 eyes (79%) improved to6/24 or better.[7] Omulecki W and colleagues did Pars plana vitrectomy +lensectomy+ SFIOL on a family of father and his two sons(total of 6 eyes) with Marfan's syndrome. All eyes achieved good visual acuity (20/20-20/25). Time of observation ranged between 8 and 20 months.[6] A study by Omulecki showed that postoperatively, transient hypotony was observed in10 eyes, fibrinous reaction in the anterior chamber in 1 eye, dispersed blood in the vitreous cavity in 8 eyes, hyphema in 1 eye, corneal edema in 1 eye, vitritis in 3 cases,and slight dislocation of the scleral-fixated IOL in 1 case. Intraocular pressure was elevated in 11 eyes preoperatively; postoperatively, it normalized in 7 eyes withoutmedication.[6] In the study by Mitra *et al* there was no case of dislocated nuclear fragments and accidental injury to the iris or ciliary body. Scant vitreous bleeding on the first post operative day was seen in 3 cases (15%); these cases also had anterior uveitis, which cleared within a week. Scleral suture erosion was not seen in any of the 20 eyes. Lens tilt in 3 cases (15%) and small decentration in 2 cases (10%) were seen, however these did not seriously compromise the visual outcome.[4] Johnston RL *et al* in their study have mentioned preoperative complications included iatrogenic retinal breaks in 3 cases, difficulty with a fixation suture in 1 case, and mild vitreous hemorrhage in 1 case. Post operative complications included retinal detachment in 2 cases, choroidal hemorrhage in 1 case, intermittent pupil capture in 9 cases, self-limiting vitreous hem orrhage in 3 cases, and late intraocular lenses dislocation in 1 case.[8] Pars plana vitrectomy for the dislocated lens removal may be combined with simultaneous scleral fixation IOL implantation. The procedure is safe and gives goodvisual rehabilitation.

CONCLUSION

From our observations it can be concluded that lensectomy with pars plana vitrectomy and scleral fixated IOL implantation is a safe and effective treatment for ectopia lentis with inadequate capsular integrity.

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