Outcomes of Lensectomy in Hereditary Lens Subluxation

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Purpose: To evaluate the results of pars plana lensectomy in patients with hereditary lens subluxation.

Method: Hospital records of patients with hereditary lens subluxation who had undergone pars plana lensectomy at Labbafinejad Medical Center, Tehran-Iran from 1996 to 2003 were reviewed. Patients with more than 6 months of follow up were included. Underlying disorders, best corrected visual acuity (BCVA) before and after surgery, intraocular pressure (IOP), postoperative refraction and complications were evaluated.

Results: Overall, records of 87 eyes of 49 patients including 27 male and 22 female subjects were reviewed. Mean follow up duration was 20±18 months. Underlying disorders leading to lens subluxation included Marfan syndrome (79.5%), Weill-Marchesani syndrome (8.2%), simple ectopia lentis (8.2%), and homocystinuria (4.1%). The most common indication for surgery was non-correctable refractive error (92.1%). Mean BCVA was 1.13 LogMAR (20/250) preoperatively, which improved to 0.26 LogMAR (20/30-20/40) postoperatively (P<0.001). BCVA ≥20/40 was achieved in 82.8% of cases after surgery. Angle-supported anterior chamber intraocular lens (ACIOL) was implanted in 85.1% of the eyes. Prophylactic band was applied in 63 eyes (72.4%). Retinal detachment developed in four eyes (4.6%) and was successfully treated.

Conclusions: Lensectomy/anterior vitrectomy with implantation of an angle-supported ACIOL in patients with hereditary lens subluxation improves vision significantly without considerable complications.

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INTRODUCTION

Hereditary lens subluxation, also named ectopia lentis or subluxated lens, may be an isolated disorder or may occur in association with multisystem disorders or an inborn error of metabolism. The most common associated disorders include Marfan syndrome, Weill-Marchesani syndrome, homocystinuria, sulfate oxidase deficiency, Ehlers-Danlos syndrome, and hyperlysinemia.[1] The mechanism for lens dislocation is an abnormality in lens zonules.[2]

Dislocation of the lens results in irregular myopic astigmatism, which can cause amblyopia in childhood. Forward lens dislocation can cause acute glaucoma and corneal endothelial cell loss. Dislocation of the lens into the vitreous cavity may result in severe vitritis and

retinal detachment.[2]

Reduced vision is managed initially by optical correction using spectacles or contact lenses, however lensectomy may become necessary when visual acuity is not adequately correctable by optical means.[3] Rehabilitation of the aphakic eye is the next challenge after a successful operation in these cases. One option is to implant an intraocular lens (IOL) which has been reported to incur serious complications.[4] The aim of this study was to evaluate the results of pars plana lensectomy in patients with hereditary lens subluxation.

METHODS

A retrospective review was performed on records of patients who had undergone pars plana lensectomy for hereditary lens subluxation at Labbafinejad Medical Center, Tehran-Iran, over a 7-year period (1996 to 2003). Patients with unilateral involvement and those with previous eye trauma were not included. We also excluded cases with complete lens dislocation into the vitreous cavity because the latter required a different surgical procedure. Variables included age, sex, etiology of lens subluxation, indication for surgery, pre- and postoperative best corrected visual acuity (BCVA), postoperative refraction, intraoperative, early (less than 2 weeks) and late (after 2 weeks) postoperative complications and length of follow up.

All patients had undergone pars plana lensectomy. In eyes with pathologic changes in the retinal periphery predisposing to retinal detachment, a prophylactic encircling band was used. The band was sutured 9.5-10 mm posterior to the limbus in each quadrant and the free ends were fixed in the superotemporal quadrant. The sclerotomies were sutured after performing the procedure. In eyes with no contraindication for IOL implantation such as young age (< 4 years) or peripheral anterior synechiae (PAS), an angle-supported ACIOL with Z-shaped haptics was implanted. The IOL was inserted through a limbal incision which was later sutured with 10-0 nylon.

RESULTS

Eighty-seven eyes of 49 patients (27 male and 22 female) with mean age of 13.8 ± 9.1 (range 3-38) years underwent pars plana lensectomy. Etiologies for lens subluxation included Marfan syndrome (39 patients, 79.6%), simple ectopia lentis, Weill-Marchesani syndrome (each in 4 patients, 8.2%) and homocystinuria (2 patients, 4.1%). Indications for surgery included uncorrectable refractive error (92.1%), anteriorly dislocated lens (5.7%) and flat AC (2.2%). Direction of the displacement was superotemporal in 57 eyes (65.5%), superonasal in 11 eyes (12.6%), inferonasal in 9 eyes (10.4%) and inferotemporal in one eye (1.2%). The lens was completely dislocated into the AC in 5 eyes (5.7%).

An ACIOL was implanted in 74 eyes (85.1%) and a prophylactic band was applied in 63 eyes (72.4%). Peripheral iridectomy (PI) was performed in 76 eyes (87.4%) intraoperatively to prevent pupillary block. Three eyes which had not undergone PI developed pupillary block postoperatively. These cases were successfully managed with Nd:YAG laser PI, and achieved controlled IOP without medication at final follow up.

Preoperatively mean BCVA was 1.13±0.32 LogMAR (20/250) which improved to 0.26±0.35 LogMAR (20/30 to 20/40) after a mean follow up of 20±18 (range 6-80) months (paired t test, $P<0.001$). Of 87 eyes, 72 (82.8%) achieved visual acuity of 20/40 or better. Causes of postoperative BCVA <20/40 included amblyopia (9 eyes), retinal detachment (4 eyes) and glaucoma (2 eyes).

Postoperative IOP was in the normal range (6-21 mmHg) in all cases except for 2 eyes of one patient with Weill-Marchesani syndrome in which IOP was high because of PAS formation due to long-standing flat AC.

Table 1 summarizes intraoperative and early and late postoperative complications. Intraoperative complications were few, not serious and resolved during follow up. All cases of sterile inflammation improved with topical or systemic corticosteroids. Both cases of hypotony were due to leakage from the

sclerotomy sites which stopped after a few days with IOP reaching normal levels. Malpositioned IOLs included two cases of mild IOL capture which did not require any treatment and one case of haptic-corneal touch that underwent IOL exchange with the same type of IOL. All cases of refractive over-correction (postoperative myopic refraction ≥-1.5 D) occurred in eyes with prophylactic bands despite the fact that the myopic effect of the band was compensated by decreasing 1.5 to 2 D from the calculated IOL power. Nine cases of under-correction (postoperative hyperopic refraction ≥+1.5 D) included 5 eyes with and 4 eyes without prophylactic bands (P= 0.23). Overall retinal detachment (RD) occurred in 4 eyes (4.6%). Three eyes of 2 patients with Marfan syndrome developed RD 6, 18 and 20 months postoperatively. These eyes underwent scleral buckling and achieved final BCVA of 20/60-20/120. The fourth case of RD was localized which was treated by barrier laser and achieved final BCVA of 20/30. All cases of RD occurred in eyes with prophylactic bands.

Table1 Complications of pars plana lensectomy

Complications		**No**	**%**
Intraoperative:			
	Hyphema	1	1.2
	Vitreous hemorrhage	1	1.2
Early Postoperative:			
	Sterile inflammation	6	6.9
	Pupillary block 3	3	3.5
	Hypotony	2	2.3
	High IOP	2	2.3
Late Postoperative:			
	Over-correction	10	11.5
	Under-correction	9	10.4
	Retinal detachment	4	4.6
	Malpositioned IOL	3	3.5
	CME	1	1.2

IOL: intraocular lens, CME: cystoid macular edema

DISCUSSION

Surgical management of the subluxated lens has historically been reported to be associated with serious intraoperative complications and poor visual prognosis. In 1967, Jarret[5] reported visual improvement in 58% of patients and Cross[6] in 1973 reported this outcome in 51%. Following the developments in vitrectomy instrumentation and advances in microsurgical techniques since 1980, surgical results have greatly improved.[7] Halpert[8] and Antoby[9] reported final visual acuity of 20/40 or better in 80% and 84% of their patients, respectively. In agreement to these recent studies, 82.8% of our cases achieved this level of vision. The most prevalent cause for limited final visual acuity (<20/40) in our study and the latter two studies[8,9] was amblyopia.

IOL implantation after lens extraction in eyes with lens subluxation has been reported to be associated with severe complications such as glaucoma and corneal decompensation.[8,9] Antoby[9] reported two cases of scleral fixated IOLs. One was complicated by dislocation into the vitreous cavity after 3 years and the other led to explantation of the lens due to severe vitritis. We performed IOL implantation in 85% of our cases (74 eyes) using an angle-supported ACIOL with Z-shaped haptics and the results were encouraging. We observed only 3 cases of malpositioned IOLs after a mean follow

up of 20 months. These included 2 cases of mild IOL capture with no need for treatment and one case of haptic-corneal touch necessitating IOL exchange. Based on these results, it seems that a suitable and properly-positioned ACIOL, does not incur significant complications. Advantages include alleviating aphakia-related problems, especially amblyopia.

The incidence of RD was high in earlier studies: 15% and 25% in the series reported by Jarret[5] and Cross[6], respectively. This figure decreased to a much lower range of 3% to 4% in recent studies.[8,9] RD occurred in 4 eyes (4.6%) of our series including one case of localized RD. These eyes achieved final visual acuity of 20/60 to 20/120. Interestingly, all these 4 eyes had prophylactic bands. We do not know how much the prophylactic band was able to prevent RD and whether the incidence of RD would have been increased if eyes with degenerative changes in the retinal periphery had not received a prophylactic band.

Over-correction was seen merely in eyes with prophylactic bands, although the myopic effect was compensated by decreasing 1.5 to 2 D from the calculated IOL power. There is no method for exact prediction of the myopic effect of an encircling element. We recommend limiting the use of prophylactic bands to cases at risk for RD.

In the current series under-correction was observed in 9 eyes including 5 eyes (7.9%) with and 4 eyes (16.7%) without prophylactic bands (P= 0.23). It seems that under-correction can be the result of random errors in IOL power calculation. Considering the limited cooperation of the majority of these cases due to young age during biometry, we recommend careful IOL calculation repeated 2 or 3 times.

In conclusion, pars plana lensectomy with implantation of an angle-supported ACIOL with Z-shaped haptic leads to significant improvement in vision with an acceptably low rate of complications in hereditary lens subluxation. IOL implantation is safe in these eyes and avoids aphakia-related problems and amblyopia.

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