



A Contemplation Of Pediatric Primary Ectopia Lentis

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*A*bstract

PURPOSE: To study the etiology, complications and visual outcome with optical and surgical management in pediatric primary ectopia lentis.

MATERIALS AND METHODS: This prospective, non comparative, interventional study was conducted in the Department of Pediatric Ophthalmology at College of Ophthalmology and Allied Vision Sciences / King Edward Medical University, Mayo Hospital Lahore, from July 2010 to December 2011. Thirty eyes of thirty patients, 4 to 12 years of age, having primary crystalline lens sub-luxation were included in the study. Only one eye of each patient was included in the study. Optical management group and surgical management group were identified and catered. Patients with secondary sub luxation of crystalline lens were excluded. Treatment options used were optical correction, endo-bag limbal lensectomy and anterior vitrectomy with aphakic correction by glasses and contact lenses , and endo-bag lens aspiration with posterior chamber intraocular lens implantation with or without capsular tension ring followed by amblyopia therapy .

RESULTS: Thirty eyes of thirty patients were included in the study. Eighteen patients were male and twelve patients were female. All had bilateral sub-luxated lenses. Marfan's syndrome was encountered in 14(46.7%) patients, Weil Marchesani syndrome in 4(13.3%) patients, Homocystinuria in 2(6.7%) patients, Hereditary ectopia lentis in 4(13.3%) of patients and simple ectopia lentis in 6(20%) of patients. Endobag lensectomy was performed on 20(66.7%) eyes while lens aspiration with posterior chamber intraocular lens implantation with capsular tension ring was done in 3(10%) eyes and in 3(10%) eyes lens aspiration with posterior chamber intraocular lens implantation was done without capsular tension ring. Pre operative best corrected visual acuity was HM-6/60 in 14(46.7%) eyes, 6/36-6/24 in 10 (33.3%) eyes and 6/18-6/12 in 6(20%) eyes. After six months best corrected visual acuity was 6/6-6/9 in 6(20%) eyes , 6/12-6/18 in 16(53.3%) eyes , 6/24-6/36 in 4(13.3%) eyes and 6/60-HM in 4 (13.3%) eyes . Twenty six (86.7%) eyes showed post-operative best corrected visual acuity improvement from 2 to 6 lines on snellen's visual acuity chart.

CONCLUSION: Endobag limbal lensectomy is a safe and effective technique for management of ectopia lentis in pediatric age group . Endobag lens aspiration with posterior chamber intraocular lens implantation with capsular tension ring is a welcome consideration in management of pediatric ectopia lentis

KEY WORDS: Ectopia lentis , Subluxation of lens , Dislocation of lens , Aphakia , Endo bag lensectomy , Capsular tension ring .

surgery. Optical dispensation was revisited and amblyopia therapy was optimized as and where indicated.

RESULTS:

Out of 30 patients 18 were male and 12 were female (Table 1). Age range was between 4 -12 years (Table 2). 20 eyes were right and 10 were left (Table 3). 14 (46.7%) patients were with Marfan's syndrome, 4 (13.3%) patients were having Weill Marchesani syndrome, 2 (6.7%) were with Homocystinurea, 4 (13.3%) were hereditary ectopia lentis and 6 (20%) were with simple ectopia lentis (Table 6). In 18 (60%) eyes there was subluxation while in 12 (40%) eyes there was dislocation (Table 4). Anterior dislocation was seen in 8 (26.7%) eyes while posterior dislocation was seen in 4 (13.3%) eyes. Cataract was seen in 16 (53.3%) eyes (Table 5).

Optical correction was given in 4 (13.3%) patients, endobag limbal lensectomy was performed in 20 (66.7%) patients, lens aspiration with posterior chamber IOL implantation with capsular tension ring was done in 3 (10%) patients and lens aspiration with posterior chamber IOL implantation without capsular tension ring was done in 3 (10%) patients (Table 7).

TABLE 1: GENDER DISTRIBUTION (n=30)

GENDER	NO OF PATIENTS
MALE	18 (60%)
FEMALE	12 (40%)

TABLE 2: AGE DISTRIBUTION

AGE IN YEAR	NO OF PATIENTS
1-4	4 (13.3%)
5-8	18 (60%)
9-12	8 (26.7%)

TABLE 3: DISTRIBUTION OF LATERALITY

SIDE	NO OF EYES
RIGHT EYE	20 (66.6%)
LEFT EYE	10 (33.3%)

TABLE 4: DISTRIBUTION OF LUXATION (n=30)

EXTENT OF LUXATION	NO OF PATIENTS
SUBLUXATION	18 (60%)
ANTERIOR DISLOCATION	8 (26.7%)
POSTERIOR DISLOCATION	4 (13.3%)

TABLE 5: DISTRIBUTION OF LENS DEFORMATION (n=30)

LENS DEFORMATION	NO OF CASES
CATARACTOUS LUXATION	16 (53.3%)
CLEAR LENS LUXATION	14 (46.7%)

TABLE 6: ETIOLOGY OF ECTOPIA LENTIS

ETIOLOGY	NO OF PATIENTS
Marfan's syndrome	14 (46.7%)
Weill Marchesani syndrome	4 (13.3%)
Homocystinuria	2 (6.7%)
Hereditary ectopia lentis	4 (13.3%)
Simple ectopia lentis	6 (20%)

TABLE 7: INTERVENTION EXECUTED

INTERVENTION	NO OF PATIENTS
Optical correction	4 (13.3%)
Limbal lensectomy	20 (66.7%)
Endo bag lens aspiration with PCIOL implant	6 (20%)

TABLE 8: BEST CORRECTED VISUAL ACUITY BEFORE INTERVENTION

VISUAL ACUITY	NO OF PATIENTS
HM-6/60	14 (46.7%)
6/36-6/24	10 (33.3%)
6/18-6/12	6 (20%)

TABLE 9: BEST CORRECTED VISUAL ACUITY 6 MONTHS AFTER INTERVENTION

VISUAL ACUITY	NO OF PATIENTS
6/6-6/9	6 (20%)
6/12-6/18	16 (53.3%)
6/24-6/36	4 (13.3%)
6/60-HM	4 (13.3%)

Preoperative best corrected visual acuity was HM-6/60 in 14 (46.7%) eyes, 6/36-6/24 in 10 (33.3%) eyes, and 6/18-6/12 in 6 (20%) eyes (Table 8).

Six months postoperatively best corrected visual acuity was 6/6-6/9 in 6 (20%) eyes, 6/12-6/18 in 16 (53.3%) eyes, 6/24-6/36 in 4 (13.3%) eyes and 6/60-HM in 4 (13.3%) eyes (Table 9).

There was a significant improvement in BCVA



postoperatively in all eyes of all patients except 4(13.3%).

Four (13.3%) eyes presented with glaucoma. Three eyes had controlled IOP after lensectomy and one eye needed trabeculectomy with mitomycin C as a secondary procedure. Anterior uveitis was recorded in 6(20%) eyes postoperatively and was successfully controlled medically.

In patients treated with limbal lensectomy, aphakia was corrected with glasses in 16 eyes and soft contact lenses in 4 eyes. There was no significant difference in final visual acuity outcome in these groups.

No surgical complication was encountered peroperatively in any of the cases. There was retinal detachment in one eye within three months after surgery which was repaired with sclera buckling successfully.

DISCUSSION:

Ectopia lentis is a disease the history of which can be traced to the dawn of eternity. It can occur at any age¹². It is the most common congenital lenticular anomaly second only to cataract².

Early diagnosis and treatment of ectopia lentis are essential because of associated ocular repercussions and potentially life threatening systemic volcanos. In the absence of familial predilection its diagnosis may be delayed.

Cornerstone of management in ectopia lentis is visual rehabilitation by optometric optimization or surgical intervention as needed. If good vision can be achieved with optical correction and there is no evidence of complications, surgical intervention can be held in abeyance.

Modern microsurgical techniques yield very good results with limbal or pars plana lensectomy for ectopia lentis^{13,14}. Either approach is effective and the selection depends on the comfort of the surgeon and familiarity with the technique⁴. In this study we used endo-bag limbal lensectomy approach on 26 eyes. A closed system endosurgical technique ensures globe maintenance, preserves normal anatomical relationships and prevents scleral and vitreous collapse¹⁵.

The methods for visual rehabilitation adjunctive to surgery include glasses, contact lenses, and IOLs. Recently, intraocular lens implantation has emerged as an effective and safe method for the correction of aphakia, and several authors have reported satisfactory results with this technique^{16,17}. However, the number of patients in these studies was small and the follow-up period was too short to ensure the safety

and effectiveness of the technique. Intraocular lens implantation has benefits over contact lenses, including reduced chances of corneal epithelial erosion, infection, displacement of the contact lens, and lack of cooperation in both patients and families.

In this study endo-bag lens aspiration with posterior chamber IOL implantation with or without CTR was done in 6(20%) eyes. No complication was observed and BCVA improved in all eyes. Pfeifer and Mikek¹⁸ described a surgical technique based on the use of Cionni endocapsular tension ring, dry irrigation aspiration of lens material, centration of the capsular bag and foldable IOL implantation into the bag with no serious intra operative or postoperative complications. The final BCVA improved in 9 of 11 eyes. More studies on use of the modified CTR are required to provide pseudophakia as an ideal treatment of ectopia lentis in pediatric age group.

However, anterior chamber intraocular lenses should not be used, especially in pediatric patients, due to the increased risk of corneal endothelial damage and angle-closure glaucoma in the long term¹⁹. Scleral fixation of posterior chamber IOLs²⁰ seems to be a safer method, but postoperative complications are more likely. Therefore, we must carefully weigh the benefits and risks of this surgery before deciding on the approach and timing for IOL implantation.

We corrected aphakia with glasses in 16 eyes and with soft contact lenses in 4 eyes in patients treated with limbal lensectomy. No significant difference was seen in the visual outcome with regard to method of postoperative visual rehabilitation. Glasses were also given in remaining patients treated with optical correction and endo bag lens aspiration with posterior chamber IOL implantation with or without CTR to obtain BCVA. No complications related to glasses or contact lenses occurred, and no patients changed their method of visual rehabilitation due to poor cooperation. Undoubtedly, occlusion therapy should be initiated whenever amblyopia is involved, regardless of the method of visual rehabilitation²¹.

Our sample size was small and follow up period too did not land into adulthood. Capsular bag preservation, bag expansion and with-in the bag IOL implantation are the horizons to be further explored and standardized with a larger sample size and longer follow up period.

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