**Original Article**

# A Review of 100 Cases of Ectopia Lentis with Glaucoma: Its Types, Presentation, Management and Visual Prognosis

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**Purpose:** To examine the clinical profile of glaucoma associated or caused by ectopia lentis, its types and management and visual prognosis.

**Material and Methods:** The medical record of 100 consecutive patients suffering from Ectopia lentis and glaucoma admitted in Eye Unit of Khyber Teaching Hospital, Peshawar from June 2002 to December 2007 were reviewed retrospectively. The clinical records were analyzed. The main parameters were type of glaucoma, its management, visual prognosis and IOP control.

**Results:** No age and sex was exempted. Main types of glaucoma were pupillary block 26.92% (35 eyes), angle anomaly 18.46% (24 eyes), glaucoma due to lens in the anterior chamber 16.92% (22 eyes), traumatic glaucoma 14.61% (19eyes), phacolytic glaucoma in 10% (13 eyes) followed by mixed type of glaucoma 16.16% (18 eyes). In 87.69% (114 eyes) surgical intervention done as peripheral iredectomy in 33.08% (43 eyes), trabeculectomy 13.85% (18 eyes), phacotrabeculectomy in 9.23% (12eyes), lensectomy in 23.08% (30 eyes) and lensectomy with vitrectomy in 8.46% (11 eyes).

Glaucoma was completely relieved in 74.62% cases while 25.48% (43 eyes) needed antiglaucoma medication. Visual acuity was improved in 70.77% (92 eyes). Amblyopia, optic disc cupping and optic atrophy were the main causes of visual failure.

**Conclusion:** Glaucoma in ectopia lentis patients is very common. In more than 80 % cases need surgical intervention like crystalline lens removal plus glaucoma surgery. Even after surgery chances of refractory glaucoma is there.

Ectopia lentis or displaced crystalline lens syndrome may be congenital or acquired .In either case it is associated with a number of ocular and systemic complication. The ocular complications are mainly glaucoma, cataract and uveitis. Lund and Sjontoft conducted study in1950 and found glaucoma in 25% of cases. Various type of glaucoma's associated with ectopia lentis are, pupillary block, angle anomaly, phacolytic, traumatic angle recession, primary angle closure, primary open angle and mixed type. Glaucoma is a serious blinding disease as WHO data in 1990 has reported is the third leading cause of blindness in the world.

The management of glaucoma associated with ectopia lentis is often difficult and challenging. The current management strategies until lately were aimed at reduction of intra ocular pressure by topical eye drops containing beta-adrenergic antagonist, cholinergic agonists, adrenergic agonists or carbonic anhydrase inhibitor and prostaglandins analougues or in combination with systemic carbonic anhydrase inhibitors and hyperosmotics. Failure of medical therapy is followed by laser or surgical intervention. The main surgical interventions are peripheral iridectomy, trabeculectomy with or without antimeta-

bolites, lensectomy with or without vitrectomy and phacotrabeculectomy.

## MATERIAL AND METHODS

All the patients of ectopia lentis associated with glaucoma (intraocular pressure more than 22mmHg) admitted to eye ward from June 2002 to Dec.2007 were studied. The history charts chosen were analyzed for relevant data. This includes age, sex and visual acuity. History revealed ectopia lentis to be congenital or acquired. Ocular examination was done with respect to visual acuity, anterior chamber examination i.e. corneal edema, peripheral anterior syncline, anterior chamber depth, iris atrophy, lens dislocation (luxation)/ subluxation and its site. Intraocular pressure measurement with applanation/ goldmann tonometer taken at the time of admission and discharge. Gonioscopy finding were recorded. Posterior segment assessment including disc cupping, neovascularization, atrophy and hemorrhage. Systemic examination was performed with specific anomalies of cardiovascular system, musculoskeletal system and nervous system in view.

Where needed electrocardiography, echocardiography, full blood count, ESR, urine examination, bleeding time, clotting time, sodium nitroprusside test, homocystine level in urine and plasma were also done.

The treatment modalities were studied. Anti glaucoma therapy was given to control glaucoma. In majority cases it was not successful. Surgical intervention was done. These were mainly iridectomy, trabeculectomy, lensectomy, phacotrabeculectomy and vitrectomy.

## RESULTS

One hundred and thirty eyes of one hundred consecutive patients having ectopia lentis with glaucoma, hospitalized for management in Khyber teaching hospital were studied. Out of these one hundred patients, seventy-seven were male and twenty three were female. Bilateral glaucoma was present in thirty patients and unilateral in seventy (Table 1).

### Age distribution

No age was exempted. Up to age 10 years there were 19 patients (19%), in 11-19 years age group 37 patients (37%), in 20 to 39 years group 22 patients (22%) while 40 years and above 22 patients (22%) (Table 2).

### Type of glaucoma

Pupil block glaucoma in 35 eyes (26.92%), angle anomaly in 24 eyes (18.46%), glaucoma due to lens in the anterior chamber in 22 eyes (16.92%), traumatic glaucoma in 19 eyes (14.61%), phacolytic glaucoma in 13 eyes (10%) mixed type glaucoma in 09 eyes (6.92%), primary open angle and closed angle in 05 eyes (3.85%) and 03 eyes (2.31%) respectively (Table 3).

**Table 1:** Sex and Glaucoma

| Sex | No of patients n (%) |
|---|---|
| Male | 77 (77) |
| Female | 23 (23) |
| Total | 100 (100) |

**Table 2:** Ages and Glaucoma

| Age (years) | No of patients n (%) |
|---|---|
| Up to 10 | 19 (19) |
| 11-19 | 37 (37) |
| 20 to 9 | 22 (22) |
| 40 to 59 | 17 (17) |
| 60 and above | 05 (05) |
| Total | 100 (100) |

**Table 3:** Types of glaucoma

| Types | No. of eyes n (%) |
|---|---|
| Angle anomaly | 24 (18.46) |
| Pupil block | 35 (26.92) |
| Photolytic | 13 (10) |
| Glaucoma with lens in anterior chamber | 22 (16.92) |
| Angle closure glaucoma | 03 (14.61) |
| Open angle glaucoma | 05 (2.31) |
| Trauma and angle recession | 19 (3.85) |
| Lensectomy and vitrectomy | 09 (6.92) |

**Therapeutic intervention**

In majority cases ectopia lentis with glaucoma was unable to be controlled with anti glaucoma medication. In 114 eyes (87.69%) needed surgical intervention as peripheral iredectomy in 43 eyes (33.08%), lensectomy in 30 eyes (23.08 %), trabeculectomy in 18 eyes(13.85%), phacotrabeculec-tomy in 12 eyes (9.23%) and lensectomy with vitrectomy in 11 eyes (8.46%) eyes (Table 4).

Glaucoma was controlled in 97 eyes (74.62%) while in 43 eyes (25.48%) it remained uncontrolled and needed additional anti glaucoma medication (Table 5).

Visual acuity was improved to 6/6-6/12 in 69 eyes (53.85%), up to 6/36 in 23 eyes (17.69%), up to 3/60 in 12 eyes (9.23%) and perception of light in 26 eyes (20%) (Table 6).

**Table 4:** Management and glaucoma

| S.No | Type of treatment | No of eyes n (%) |
|---|---|---|
| A | Medical | 16 (11.3) |
| B | Surgical<br>•Peripheral/iridectomy<br>•Trabeculectomy<br>•Lensectomy<br>•Phaco Trabeculectomy<br>•Lensectomy and virectomy | 114 (87.69)<br>• 43 (33.08)<br>• 18 (13.85)<br>• 30 (23.8)<br>• 12 (9.23)<br>• 11 (8.46) |

**Table 5:** Glaucoma status

| Glaucoma Status | No of eyes n (%) |
|---|---|
| Controlled | 97 (74.62) |
| Uncontrolled | 33 (25.48) |

**Table 6:** Visual prognosis and glaucoma

| Vision | Eyes n (%) |
|---|---|
| PL | 26 (20) |
| Pl + 3/60 | 12 (9.23) |
| Up to 6/60 | 23 (17.69) |
| Up to 6/12 | 69 (53.08) |

## DISCUSSION

The crystalline lens is implicated as causative element in producing several forms of glaucoma. These include lens dislocation/subluxation (ectopia lentis), lens intumescent cataract glaucoma, classical pupillary block glaucoma, lens particle glaucoma, phacoanaphylaxis and phacolytic glaucoma[1]. Various studies showed glaucoma association is an established fact[2-3].

Glaucoma has been reported in patients with ectopia lentis as part of the syndromes like marfan, Weill marchesani, homocytineurea and aniridia[4]. Fibrilin -1 (FBN-1) gene mutations screening analysis shows ectopia lentis with secondary glaucoma is positive as compare to normal control people.

This type of genetic and molecular understanding provide information for genetic counseling[5].

Glaucoma in ectopia lentis patients has been reported as 41.5%[6]. Cross and Jerret has reported the presence of glaucoma as 25 %[7] while in another study the glaucoma has been reported as 25 % in marfan syndrome[8]. Maumenee reported glaucoma as 129/1000 in patients of ectopia lentis as compared to normal population where it was 40/1000.

The ectopia lentis is mainly associated with secondary type of glaucoma although primary angle closure and open angle glaucoma has also been reported[9].

The secondary glaucoma's are associated or aggravated by angle anomaly in congenital type and traumatic angle recession in traumatic type. No age and sex is exempted from ectopia lentis and glaucoma. Glaucoma was noticed in 59% in age group of 20-39 years. Ectopia lentis and its induction of gluacoma appear in second and third decade of life because of changes in zonules and hperplastic cillary body. This causes forward displacement of iris accompanied by unequal lens zonuler tension leading to angle closure or pupillary block glaucoma or it may appear due to abnormal structure of anterior chamber[10].

The crystalline lens is the main culprit causing raised intraocular pressure particularly when it is displaced anteriorly, causing pupil block glaucoma. This condition was noticed in 26.92 % of cases.

In our study pupilary block glaucoma was highest followed by angle anomaly and lens in the anterior chamber with glaucoma. In study of Harrison et al angle anomaly and pupillary block were the main glaucoma variety[11].

Ectopia lentis or lens displaced syndrome associated with glaucoma remains a therapeutic challenge for ophthalmologists. If it is of congenital origin then presence of other ocular condition like angle anomaly and aniridia or systemic condition like

homocytinuria further complicate the condition. On other hand traumatic variety with hypaema and angle recession is also difficult to manage.

Medical treatment may be helpful to control intraocular pressure temporarily in cases like angle closure, open angle or traumatic glaucoma. But insuffient to control glaucoma in majority of types,[12] which need surgical intervention like peripheral iredectomy, trabeculectomy/ phacotrabeculectomy, anterior vitrectomy and lensectomy.

In our study 11.3% cases of glaucoma were controlled with medication while 88.7 % cases needed surgical intervention. In 74.62% glaucoma was cured. While 25.48% cases still needed medication.

Peripheral iredectomy is useful in pupullary block glaucoma. If peripheral iredectomy fails, lensectomy with or without intraocular implantation is done. The intraocular pressure becomes normal[13].

Harrison and his colleagues studied forty five patients with ectopia lentis in homo cystinuria. Sixty two patients had lens into the anterior chamber or pupil block glaucoma. Eighty-four surgical procedures were done. Peripheral iredectomy was not successful alone. Lensectomy and trabeculectomy were the choice of operation[11].

In another case angle closure glaucoma with progressive myopia due to ectopia lentis was treated with propyhlactic lensectomy. That proven most effective[9].

Kluppel et al operated 23 eyes having decreased visual acuity and uncontrolled secondary glaucoma. Main surgical procedure was lensectomy and transcleral posterior chamber intraocular lenses implantation suture technique. Visual acuity improved in 17/23 eyes (73.91%). The decrease visual acuity in remaining 26.09 % eyes were due to amblyopia. Gluacoma was controlled in 100 % of cases[14].

80% Weill Marchesani Syndrome have glaucoma, which responds poorly to medical treatment alone. Peripheral iredectomy, PPL (Pars plana lensoctomy), lensectomy with anterior vitrectomy and trabeculectomy are surgical procedure of choice[15].

Lensectomy done in patients having "glaucoma with spherophakia" where glaucoma was uncontrolled with patent peripheral iredectomy and medical treatment. Right eye visual acuity was 6/6. Intra ocular pressure was normal without glaucoma therapy. Left eye visual acuity was 6/9 and normal intra ocular pressure[13].

In polish journal klin oczna it has been reported that in 116 eyes of ectopia lentis parsplana lensectomy, vitrectomy and scleral fixation intraocular implant-tation done. Visual acuity improved in 89% of cases while glaucomatous optic atrophy and amblyopia were the main cause of decreased visual acuity[16].

In our study visual acuity improved in 92 eyes (70.77%) while in 38 eyes (29.23%) the glaucomatous optic cupping was the main cause of decreased visual acuity. The glaucoma was relieved in 74.62% eyes.

In conclusion ectopia lentis is very challenging clinical field. Glaucoma association makes it further complex. In more than 80 % cases need surgical intervention like crystalline lens removal plus glaucoma surgery. Even after successful surgery chances of refractory glaucoma is there.

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