Original Article

Ectopia Lentis – Etiology and Management in Children

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Purpose: To study the etiology, mode of presentation and management strategies of ectopia lentis in children less than 10 years of age.

Study Design: Interventional case series.

Place and Duration of Study: The Children's Hospital and Institute of Child Health, Lahore and department of ophthalmology, KEMU, Lahore from January 2010-January 2015.

Material and Methods: All patients diagnosed as having non-traumatic ectopia lentis were included in the study. The children were evaluated for vision threatening complications of ectopia lentis. A complete systemic examination was done by the pediatrician to diagnose any associated inherited systemic syndrome. The family members of the child were also examined for ectopia lentis. Surgery was planned in all eyes where there was displacement of the lens in anterior chamber with or without pupil block.

Results: A total of 54 eyes of 27 patients diagnosed as having non-traumatic ectopia lentis were included in the study. There were 15 males, 12 female patients with 24 patients falling in the age bracket of 0 – 7 years. Seventeen out of the twenty – seven patients were diagnosed as having inherited systemic syndrome, Marfan being the most common. Majority of the patients presented to the eye department only when there was pain in the eye due to increase in IOP.

Conclusion: Every child patient with ectopia lentis is a candidate for a thorough ocular and systemic examination for early recognition of vision threatening as well as life threatening systemic problems. The importance of involvement of parent with amblyopia management cannot be overemphasized for visual rehabilitation of the child.

Key words: Ectopia lens, subluxation of lens, Lensectomy.

Ectopia Lentis means partial displacement or complete dislocation of the lens from its normal position, trauma being the most common cause[1]. Non-traumatic ectopia lentis can occur as an isolated entity or may be part of the spectrum of a variety of inherited disorders[2,3]. In familial or idiopathic ectopia lentis there are no associated systemic or ocular findings[4].

High refractive errors and optical aberrations because of the lens not being in place reduce vision[5]. When there is a uniform loosening of the lens zonules a change in the shape of the lens results in myopia. Anterior dislocation of the lens may cause pupil block and an increase in IOP. A posteriorly displaced lens has the potential of causing damage to the retina.

The ocular complications related to ectopia lentis can result in permanent decrease in vision, especially if this occurs in amylogenic age. Prompt recognition and treatment are required to decrease the burden of blindness from complicated ectopia lentis. When a pediatric ophthalmologist comes across a child with ectopia lentis, he/she should consider the wide spectrum of inherited disorders associated with this entity[2,6]. The child should be referred to appropriate

subspecialty so as to address the possible life – threatening disease.

The child should be monitored for amblyopia, and should be kept on a close follow-up with frequent cycloplegic refraction and correction of the visual error with appropriate glasses. In cases of marked lens displacement surgical intervention should be considered followed by visual rehabilitation.

MATERIAL AND METHODS

A prospective interventional study was conducted in the department of Ophthalmology, KEMU and department of pediatric ophthalmology, Children's Hospital & Institute of Child Health (CH-ICH), Lahore from January 2010 – January 2015. Patients were seen either at KEMU or CH&ICH and were managed respectively in either of the two institutes. All patients who presented with non-traumatic ectopia lentis were included the study.

Patient demographic information recorded on the prescribed form included age, gender and ethnicity. After complete ocular and systemic examination, note was made of the mode of presentation, BCVA, state and position of the lens, and underlying etiology. The patient was referred to the department of pediatric medicine for systemic evaluation and appropriate management.

Conservative management was done in non-significant subluxation of lens, Cycloplegic refraction was done and appropriate glasses prescribed. The parents were counseled as regards the vision threatening potential of the disease and importance of follow-up.

Surgical management: Surgery was planned in all eyes where there was displacement of the lens in anterior chamber with or without pupil block. Relevant specialty was kept on board. Within the bag lensectomy followed by anterior vitrectomy was the procedure of choice. Postoperatively a close watch was kept on the signs of any complication. The parents of the child were advised as regards use of topical antibiotics, steroids and cycloplegic drops. At the time of discharge, the patients were given appropriate glasses. A follow-up schedule was given to the parents.

RESULTS

A total of 27 patients age 3 months to 11 years were included in the study, mean age being 4.27 years with a standard deviation of 2.2. Male were 56% whereas females were 44%, (graph 1). Seventeen patients (62.9%) were diagnosed as having inherited systemic syndrome, Marfan being the most common (9 patients) (Table 1). Most of the patients presented to the eye department only when there was pain in the eye due to increase in IOP (31 eyes – 57.4%) (Graph 3).

Table 1: Underlying etiology of ectopia lentis.

Underlying etiology	N (Patients) N (%)
Marfan syndrome	9 (33.33)
Weil Marchesani	5 (18.5)
Homocystinuria	3 (11.11)
Idiopathic	6 (6.8)

Table 2: Ocular associations

Ocular associations	N (Eyes) N (%)
Axial myopia	18 (33.33)
Glaucoma	8 (14.8)
Cataract	4 (7.4)
Aniridia	2 (3.7)

Table 3: Management Strategies of ectopia lentis

Management	N (Eyes) N (%)
Conservative	21 (38.8)
Pars plana lensectomy	27 (50)
Irrigation Aspiration with IOL	3 (5.5)
Irrigation Aspiration with CTR+IOL	3 (5.5)

DISCUSSION

Ectopia lentis is the second most common congenital lenticular anomaly resulting in compromised vision. There is disruption of the zonular fibers resulting in subluxation of the crystalline lens[7]. Genetic alterations being a frequent cause of ectopia lentis[8].

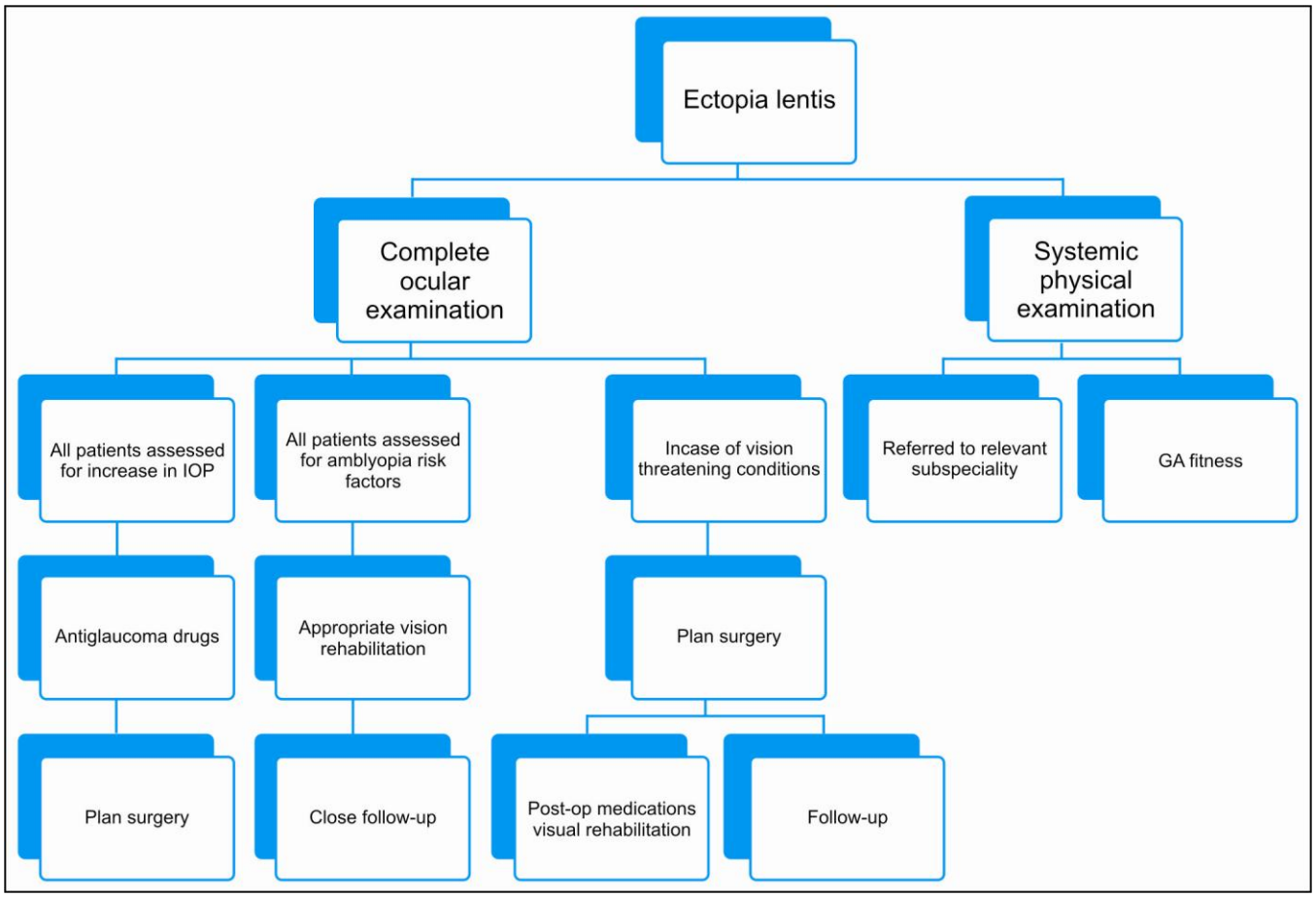
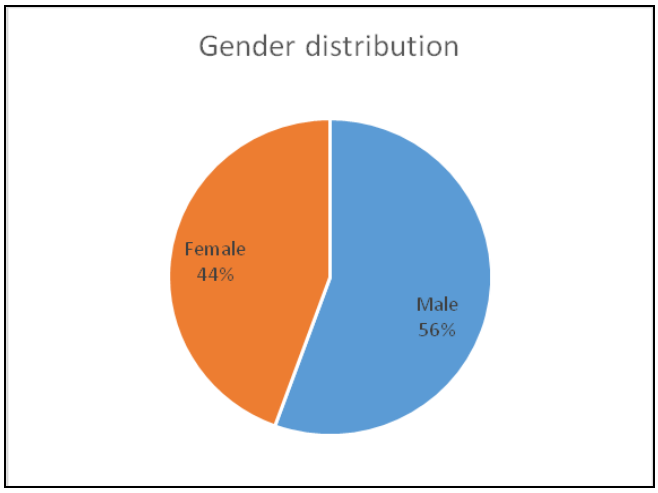
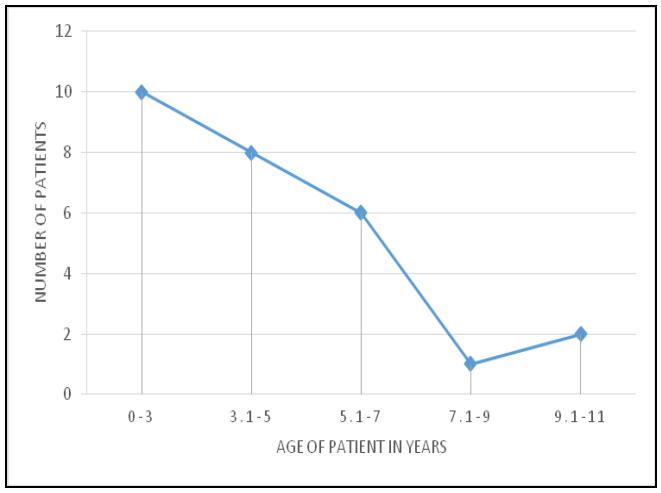


Fig. 1: Protocol of ectopia lentis management.

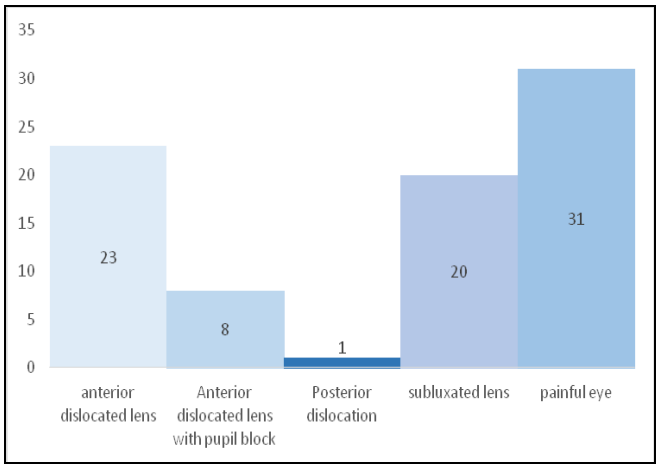


Graph 1: Gender distribution.

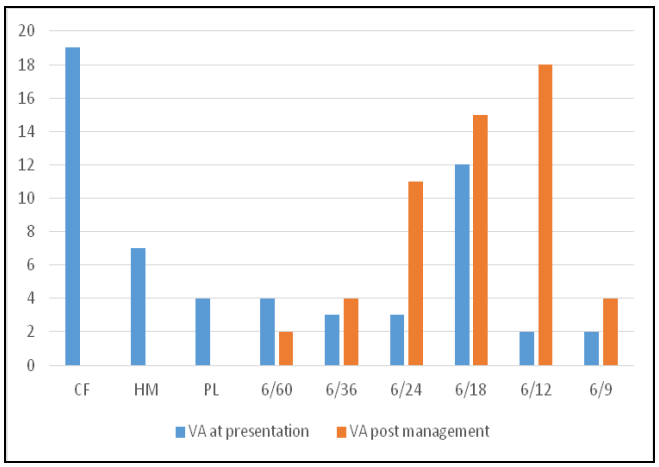


Graph 2: Age range.

This entity can be isolated or associated with inherited systemic syndromes such as Marfan syndrome[9], Weill Marchesani syndrome[10], Homocystinuria, Hyperlysinemia and sulfite oxidase



Graph 3: Mode of presentation.



Graph 4: Pre and post management visual acuity.

deficiency accounting for 75% of the cases. Marfan syndrome however, the most common cause of inherited ectopia lentis, is caused by a mutation in FBN[1,11,12]. In accordance with literature in this series inherited systemic disorders account for 63% of cases, Marfan's syndrome (33%) being most common of all systemic association. There have been reports in the literature of its association with Ehler – Danlos syndrome, Sturge – Weber syndrome, and Stickler syndrome[13].In this study however, no patient was found to be effected with these entities.

The most common mode of presentation in our series in decreasing order of frequency was anteriorly dislocated lens (43%) with and without pupil block, followed by subluxation of lens (37%). A high percentage of patients reported only when they developed severe ocular pain (57.4%). This may be due to the fact that majority come from far-flung areas having limited access to health facilities.

When dealing with a patient with ectopia lentis it is essential to make a comprehensive plan regarding diagnosis, management, visual rehabilitation and follow-up[14]. All the relevant subspecialties should be taken on board for timely diagnosis and management of associated systemic problems. The biggest challenge a pediatric ophthalmologist faces is when to plan for surgery and how to manage the visual rehabilitation of the compromised eye of the child. If there is a possibility of achieving visual improvement and maturation in the very sensitive amblyogenic age of the child with glasses, surgery can be delayed or even avoided. Despite maximum conservative management Romano et al, report ametropic amblyopia in 50% of patients with familial ectopia lenses[15].In our series 31.4% eyes had vision less the 6/24.

In cases where the vision is severely compromised surgical removal of the culprit lens should be planned[16]. A thorough ocular examination should be undertaken prior to surgery. Looking at the lens under undilated and dilated condition would help the surgeon assess the direction and location of the zonular dysfunction as well as degree and direction of subluxation. Retina should be carefully examined for any associated pathology especially in cases of Marfan syndrome[17].

In the pre-automated vitrectomy era, surgery for ectopia lentis in small eyes was met with high rates of complications e.g. vitreous loss followed by retinal detachment. Since the availability of the close system, automated, irrigation – vitrectomy there has been a remarkable improvement in the methodology and outcome with minimum complications in the hands of an experienced surgeon. Both limbal, anterior approach as well as pars plana approach has been advocated by researchers[18]. The key to the success of the surgical procedure is the meticulous removal of vitreous gel and to make sure that there is no incarceration of the vitreous in the wound. In our series no significant postsurgical complication was noted.

Postoperatively the pediatric ophthalmologist is faced with the next big challenge of visual rehabilitation[19]. Various options include aphakic glasses, contact lens, angle supported anterior chamber IOL implant, posterior chamber iris fixated IOL implant, posterior chamber capsule placed IOL with or without capsular tension ring. However, each

of these modalities has its inherent limitations and complications. In our series 88% of patients showed improvement in BCVA of 6/18 or better. The role of proper counseling and involvement of parents and compliance with amblyopia therapy cannot be overemphasized[20].

CONCLUSION

Ectopia lentis should not be considered as an isolated entity. The child patient should be taken as a whole in order to rule out the presence of associated systemic problems so as to decrease the morbidity and mortality

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REFERENCES

1. **Scothorn DM, Sporn A, Terry JE.** Ectopia lentis secondary to physical abuse in a traumatized, elderly individual. J Am Optom Assoc. 1991; 62: 630-3.
2. **Shortt AJ, Lanigan B, O'Keefe M.** Pars plana lensectomy for the management of ectopia lentis in children. J Pediatr Ophthalmol Strabismus, 2004; 41: 289–94.
3. **Chandra A, Charteris D.** Molecular pathogenesis and management strategies of ectopia lentis. Eye (Lond), 2014; 28:162–8.
4. **Sadiq MA, Vanderveen D.** Genetics of ectopia lentis. Semin Ophthalmol, 2013; 28: 313–20.
5. **Wen Y, Wu-Chen, Robert D, Letson M.** functional and stuctural outcomes following lensectomy for ectopia lentis. J Americn Assoc Pediatr Ophthalmol strabismus, 2005; 9:353–7.
6. **Hsu HY, Sean L. Edelstein JTL.** Surgical management of non-traumatic pediatric ectopia lentis: A case series and review of the literature. Saudi J Ophthalmol. 2012; 3: 315–21.
7. **Ahram D, Sato TS, Kohilan A, Tayeh M, Chen S, Leal S, et al.** A homozygous mutation in ADAMTSL4 causes autosomal - recessive isolated ectopia lentis. Am J Hum Genet. 2008; 84: 274–8.
8. **Neuhann TM.** Hereditary ectopia lentis . Klin Monbl Augenheilkd, 2015; 232: 259–65.
9. **Zadeh N, Bernstein JA, Niemi AK, Dugan S, Kwan A, Liang D, et al.** Ectopia lentis as the presenting and primary feature in Marfan syndrome. Am J Med Genet Part A. 2011; 155: 2661–8.
10. **Chu BS.** Weill-Marchesani syndrome and secondary glaucoma associated with ectopia lentis. Clin Exp Optom. 2006; 89: 95–9.
11. **Kainulainen K, Karttunen L, Puhakka L, Sakai L, Peltonen L.** Mutations in the fibrillin gene responsible for dominant ectopia lentis and neonatal Marfan syndrome. Nat Genet, 1994; 6: 64–9.
12. **Chandra A, Patel D, Aragon-Martin JA, Pinard A, Collod-Broud G, Comeglio P, et al.** The revised ghent nosology; reclassifying isolated ectopia lentis. Clin Genet. 2015; 87: 284–7.
13. **Chandra A Aragon Martin JA; Child,AH Arno G; Charteris D et. al.** Alternative diagnoses with ectopia lentis. Eye (Lond). 2012; 26: 481–2.
14. **Hoffman RS, Snyder ME, Devgan U, Allen QB, Yeoh R, Braga-Mele R.** Management of the subluxated crystalline lens. J Cataract Refract Surg. 2013; 39: 1904–15.
15. **Romano PE., Kerr NC HGM.** Bilateral ametropic functional amblyopia in genetic etopia lenti:its relation to the amount of subluxatio, an indicator for early surgical management. Binocul Vis Strabismus, 2002; 17: 235–41.
16. **Neely DE, Plager DA.** Management of ectopia lentis in children. Ophthalmol Clin North Am. 2001; 14: 493–9.
17. **Nahum Y, Spierer A.** Ocular Features of Marfan Syndrome: Diagnosis and Management. Isr Med Assoc J, 2008; 10: 179–81.
18. **Simon MA, Origlieri CA, Dinallo AM, Forbes BJ, Wagner RS, Guo S.** New Management Strategies for Ectopia Lentis. J Pediatr Ophthalmol Strabismus, 2015; 52: 269–81.
19. **Melissa A. Simo, Catherine A, Anthony M. Dinallo, Forbes PJ, Wagner RS.** Suqin Guo M. New Management strategies for ectopia lentis. J Pediatr Ophthalmol Strabismus, 2015; 52: 269–81.
20. **Epley K.D., Shainberg MJ., Lueder GTTl.** Pediatric secondary lens implantationin the absence of capsular support. J AAPOS. 2001; 5: 301–6.