Ectopia lentis: can we tell who will do well?

E. Qian[1], M. Ali Khan[1], A. Seyed-Safi[2], T. Khanam[2], A. Chandra[3], D. Charteris[2], P. Maghsoudlou[1]
1 University of Bristol, Bristol, 2 Moorfields Eye Hospital, London, 3 Southend University Hospital

Introduction

Ectopia lentis is the dislocation of the lens from its natural position, usually occurring due to a defect of its zonular attachments associated with conditions such as Marfan's syndrome and homocystinuria[1,2]. Though surgical intervention is the only definitive choice, optimal management has remained elusive[3]. There are many small studies comparing surgical methods in its treatment, but overlying factors that help prognosticate are lacking[4,5]. A retrospective study of our cohort of patients presenting with ectopia lentis is explored for factors which may impact patient outcomes.

Methods

Data of all patients presenting to Moorfield's Eye Hospital with ectopia lentis from 1969 to 2020 was collected retrospectively including demographics, presentation, visual acuity, biometry, treatment history, outcomes, and follow up visual acuities up. Student's t test was used for parametric comparison test and Chi squared tests were used for non-parametric tests.

Demographics

	Best final vision better than 0.3 LogMAR (n=66)	Best final vision 0.3 LogMAR or worse (n=29)	p-value
Age at presentation	25.74 years	21.12 years	P=0.288
Below 15 years	38.5%	51.7%	P=0.208
Female	45.6%	48.0%	P=0.800
Follow up	12.62 years	17.17 years	P=0.068

Biometry

	<0.3 LogMAR	≥0.3 LogMAR	
Refractive error	89.7%	100%	P=0.432
Axial length	24.85 mm	27.3 mm	P<0.01
Anterior chamber depth	3.84 mm	3.55 mm	P=0.290
Keratometry	41.55 D	42.17 D	P=0.143

Presentation

	<0.3 LogMAR	≥0.3 LogMAR	
Best VA	0.31	0.77	P<0.0001
VA difference (R/L)	0.44	0.38	P=0.633
Diagnosis			P=0.084
Bilateral presentation	93.9%	96.6%	P=0.600
Subluxed lens over visual axis	7.3%	14.3%	P=0.344
Ocular comorbidities (RD, glaucoma, amblyopia)	34.8%	51.7%	P=0.122
Cardiac pathology	40.9%	24.1%	P=0.116

Results

95 patients were included in analysis. The average best binocular vision was vision overall was 0.22 LogMAR. 66 patients achieved better than 0.3. This cohort compared to those that were 0.3 or worse. At presentation, axial length and best VA were significantly difference between these groups. Surgical treatment nor complications seemed to make a difference.

Considering the outcome of eyes individually, diagnosis, and treatment type were found additionally as factors that differed between the two groups. Improvement by final follow up was also significant, interesting 1-year differences was not.

Treatment

	<0.3 LogMAR	≥0.3 LogMAR	
Surgical treatment	71.2%	51.7%	P=0.066
Complications	55.6%	50.0%	P=0.716

Outcomes

	<0.3 LogMAR	≥0.3 LogMAR	
VA 1 year (best)	0.19	0.66	P<0.0001
VA change (R/L)	-0.14	-0.19	0.592
Final VA (best)	0.06	0.58	Reference
VA change (R/L)	0.52	0.38	P=2.78
Final VA improvement	-0.25	-0.19	P=0.419

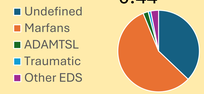
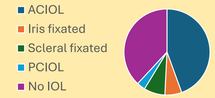
Discussion

Axial length and initial VA has been previously found to be linked to poorer outcomes with ectopia lentis[6]. Higher axial lengths are associated with Marfan syndrome and ADAMTSL mutations[6,7], which were both found in our cohorts. However, there was a high proportion of undifferentiated patients which may decrease the signal seen in the patient cohort. Considering eyes separately which may have artificially increased our power showed that diagnosis distribution was statistically different with a larger Marfan cohort in the better group and a larger ADAMTSL mutation cohort in the second group.

Discussion contd.

Using data of each eye individually also gave us a signal in that surgical treatment was more often undertaken in eyes that had a better result. Where surgical lens extraction was occurred, more patients were left aphakic in the worse cohort which may have contributed to the final VA being worse, or perhaps simply had poorer visual potential.
Unfortunately, retrospective data is incomplete, and conclusions of causality cannot be drawn here. Additional signal when considering eyes separately calls for larger cohorts and more data to better assess predictive factors for outcome of ectopia lentis.

Individual eyes

	<0.3 logMar (n=97)	≥0.3 logMar (n=88)	
Age at presentation	26.52 years	21.27 years	P=0.063
Female	46.4%	47.7%	P=0.856
Axial length	24.56 mm	26.86 mm	P<0.05
Anterior chamber	3.88mm	3.62mm	P=0.271
Keratometry	41.66 D	41.57 D	P=0.872
VA at presentation	0.44	0.92	P<0.0001
Diagnosis			P<0.01
Surgical treatment	66.0%	37.5%	P<0.001
Approach by pars plana vitrectomy	86.8%	70.4%	P=0.075
IOL type			P<0.0001
Complications for follow up	41.27	45.45	P=0.694
Complications for procedure	17.86	34.48	P=0.080
VA at 1 year	0.26	0.86	P<0.0001
VA change at 1 year	-0.18	-0.09	P=0.300
Final VA	0.08	0.91	reference
Final VA change	-0.37	-0.03	P<0.001

Conclusion

This retrospective study identifies presenting visual acuity and axial length as key prognostic factors in ectopia lentis outcomes.
Patients with better initial vision and shorter axial lengths achieved superior visual results, with 69.5% achieving vision better than 0.3 LogMAR. These findings may help guide clinical decision-making and patient counselling.

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University of Bristol
Moorfields Eye Hospital NHS Foundation Trust
Southend University Hospital NHS