Ectopia Lentis Moorfields Study: Visual and functional outcomes

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Background

- **Ectopia lentis** (EL) describes the abnormal displacement of the lens from its central position[1]. It is a well-described phenomenon which can have a significant impact on vision.

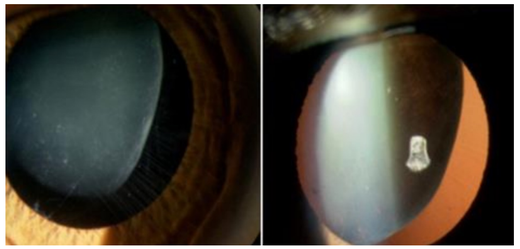


Figure 1.1, Image of lens displacement. Reproduced from Chandra and Charteris (2012)[2]

- Lens subluxation may be managed conservatively, however surgical removal of the displaced lens may be indicated. This may be accompanied by the insertion of an artificial intraocular lens[3,4] (Wagoner et al 2003, Cheung et al 2019).
- At present there are **no national or regional guidelines** for the management of ectopia lentis. To begin to address this, a cohort of patients with ectopia lentis under the care of Moorfields eye hospital was identified, and this study was performed in order to assess the current management of these patients and the long-term outcomes of this care.

Methods

A retrospective review of patients with ectopia lentis, presenting to Moorfields between 1979 and 2019, was carried out. Baseline demographics, ocular co-morbidities, surgical details, and visual acuity outcomes were obtained, in addition to whether referral to cardiology or genetics services was documented. All analyses were carried out using SPSS.

Results

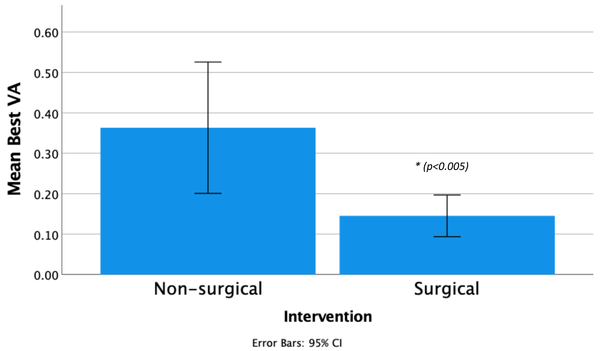
Cohort:

- Data was collected from the notes of **108** patients (Male = **57**, Female = **41**)
- Mean age at first review was **23.7** years (SD=**19.1**)
- Mean BCVA (best corrected visual acuity) at presentation was **0.44** LogMAR (SD=**0.45**)
- Among those with a confirmed diagnosis, the most commonly identified cause of EL was Marfans syndrome. (*Table 1.1, right*).

Cause of ectopia lentis	Number of patients
Marfans syndrome	47
Homocysteinuria	1
Weill-Marchesiani syndrome	0
Ehlers Danlos syndrome	2
Isolated ectopia lentis	10
Traumatic ectopia lentis	7
Unconfirmed diagnosis	41
Total	**108**

Management and visual outcomes:

- **65** patients (**60.1**%) underwent surgical management of EL after presentation to Moorfields, while the remainder were monitored and managed conservatively.
- The most recently recorded BCVA for each patient was documented. Among those having undergone surgical intervention, the final BCVA was significantly reduced compared to those managed conservatively (**0.15** (SD=**0.21**) vs **0.36** (SD=**0.48**), (*p<0.005*), (*Figure 1.2, left*).
- The mean follow-up period was **15.4** years (SD **11.1**).



Complications:

- Of the **65** patients undergoing surgical management of EL, **33** (**50.8**%) had a documented surgical complication requiring a follow-up appointment (e.g. post-op uveitis), among which **22** (**33.9**%) had a documented complication requiring a corrective procedure (e.g. repositioning/exchange of IOL). (*Table 1.2, right*).

Intervention	Number of patients	Complication requiring follow up	Complication requiring corrective procedure
No surgery	43	0	0
Lensectomy only	28	14	5
ACIOL	25	14	11
Iris-fixed IOL	5	0	0
Scleral-fixed IOL	4	4	4
PCIOL	3	1	1
Total	**108**	**33**	**22**

Discussion and Conclusion

- The **majority** of patients presenting to Moorfields with ectopia lentis were managed surgically, and the **BCVA** was significantly better than those managed conservatively.
- However it is also noted that more than a half of patients had a **recorded complication** relating to the surgical procedures performed.
- While a **strength** of this current study is that it comprises a large cohort of patients presenting to a specialist centre, it is **limited** by the absence of a broader measure of the impact of surgical interventions on patient satisfaction and quality of life.

Conclusion:

- This finding highlights the need to **develop guidelines** for the management of ectopia lentis to **optimise outcomes** for patients.

References:

1.Chandra, A, & Charteris, D. (2014). Molecular pathogenesis and management strategies of ectopia lentis. *Eye*. **2.** Chandra, Aman, Aragon-Martin, J. A., Hughes, K., Gati, S., Reddy, M. A., Deshpande, C., Cormack, G., Child, A. H., Charteris, D. G., & Arno, G. (2012). A Genotype-Phenotype Comparison of ADAMTSL4 and FBN1 in Isolated Ectopia Lentis. *Investigative Ophthalmology & Visual Science* **3.** Wagoner, M. D., Cox, T. A., Ariyasu, R. G., Jacobs, D. S., Karp, C. L., & American Academy of Ophthalmology. (2003). Intraocular lens implantation in the absence of capsular support: A report by the American Academy of Ophthalmology. *Ophthalmology*, **4.** Cheung, C. S., & VanderVeen, D. K. (2019). Intraocular Lens Techniques in Pediatric Eyes with Insufficient Capsular Support: Complications and Outcomes. *Seminars in Ophthalmology*.