Bilateral ectopia lentis in opposite quadrants in a child with Marfan syndrome

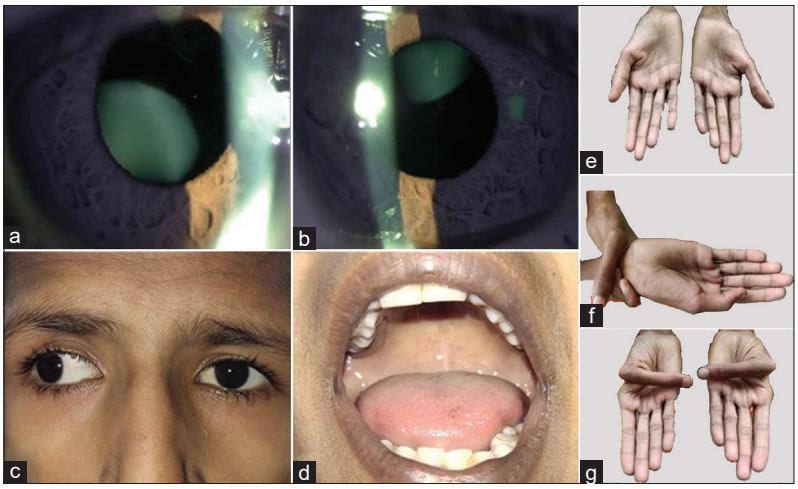

Figure 1: (a and b) Anterior segment photograph demonstrating infero-temporal lens subluxation in right eye (a) and superior lens subluxation in left eye (b). (c-g) Clinical images showing right eye exotropia, bilateral enophthalmos, and malar hypoplasia (c); high arched palate (d), arachnodactyly (e), positive wrist sign (f), and thumb sign (g)

A 15-year-old girl presented for evaluation of right eye exotropia since childhood. Ocular examination revealed best-corrected visual acuity of 20/40 in both eyes (refractive correction of -12DS/-3DC × 40° in right and -11DS/-2.5DC×130° in left eye); alternate divergent strabismus (40°) with left eye predominant fixation and ectopia lentis in opposite quadrants [Fig. 1a and b]. Fundus showed tessellated appearance with lattice degeneration. Physical examination revealed increased arm span to height ratio (1.052), decreased body upper to lower segment ratio (0.77), pectus carinatum and other clinical features [Fig. 1c-g] compatible with Marfan Syndrome (MFS). Echocardiography revealed aortic root dilatation. She was diagnosed as ectopia lentis associated with MFS.[1,2] Due to good visual acuity, she was kept under close ophthalmological follow-up and referred to cardiology for immediate management. The index report demonstrates an unusual ocular presentation (opposite lens subluxation in same individual) in a patient of MFS, which is rarely reported in literature.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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References

1. Loeys BL, Dietz HC, Braverman AC, Callewaert BL, De Backer J, Devereux RB, *et al*.The revised Ghent nosology for the Marfan syndrome. J Med Genet 2010;47:476-85.
2. Nemet AY, AssiaEI, Apple DJ, Barequet IS. Current concepts of ocular manifestations in Marfan syndrome. Surv Ophthalmol 2006;51:561-75.

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