Case Report

Cleft lip and palate with ectopia lentis et pupillae

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ABSTRACT

A 7-year-old male child with a history of bilateral cleft lip and palate presented with ectopia lentis et pupillae; there has been no previous reported association between cleft lip and palate and ectopia lentis.

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Cleft lip is a common genetic abnormality. Cleft lip with or without cleft palate occurs in approximately one out of 1,000 live-births.[1] Ectopia lentis et pupillae is a rare cause of ectopia lentis characterized by the lens and pupil being displaced in opposite directions.[2]

Case Report. A 7-year-old male child referred from the plastic surgery department presented with decreased visual acuity, and history of multiple plastic surgery for bilateral cleft lip and palate, and tongue-tie. There was no previous history of eye surgery. The child is a product of normal delivery; there was no history of maternal infections or drug exposure during pregnancy. His mother and father were completely normal with no consanguinity among parents. He has one brother and one younger sister who are completely normal. The family denies any congenital problem in the family.

Examination revealed facial asymmetry, scars of bilateral cleft lip and palate surgeries; he also had micrognathia, as shown in **Figures 1, 2, & 3**. There were no other systemic skeletal abnormalities.

Ophthalmic examination showed visual acuity of 6/60 in the right eye and 6/60 in the left eye, normal adnexa, and hypertelorism. Extra ocular muscle motility was normal, and there was no squint. Pre operative refraction was: Right eye: +6.0-5.0x70=6/36 and left eye: +12.0-1.0x90=6/12. Slit lamp examination revealed normal corneas, deep anterior chambers, the pupils were oval and displaced inferiorly in both eyes. The lens was partially displaced superiorly in the right eye and was completely displaced from the visual axis in the left eye. After dilatation the dislocated lens in the left eye was seen superiorly (**Figures 4, 5, & 6**). Fundoscopy showed normal fundus and periphery, no retinal detachment (**Figure 7**). Ultrasound for both eyes showed bilateral subluxated lenses with flat retinae (**Figure 8**). He underwent right pars plana lensectomy and vitrectomy for lenticular astigmatism, his postoperative refraction was: Right eye: +12.0-0.5x80=6/12 and left eye: +12.0-1.0x90=6/12.

Discussion. Ectopia lentis is a recessively inherited condition which is not associated with systemic abnormality, in some patients the pupil is also displaced in the opposite direction to the lens (ectopia lentis et pupillae).[3] However, our patient denied a history of any similar condition in the family. The pupils are oval or slit like showing more than the normal 0.5 mm nasal and downward eccentricity. They are usually displaced in a direction opposite to the lens that is towards the most defective zonular fibers. The irides often have an atrophic appearance and may dilate poorly,[4] this goes

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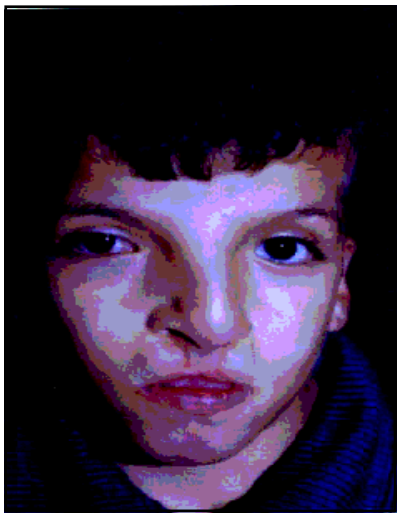

Figure 1 - General appearance of the child with obvious post-operated cleft lip, the ectopic pupils also seen.

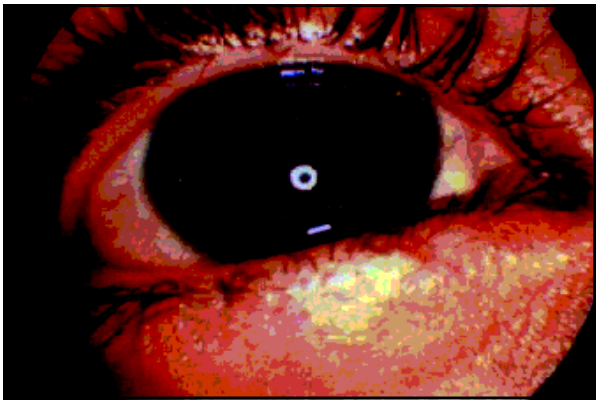

Figure 4 - Close color photograph of the right eye (aphakic) showing the ectopic downward displaced pupil.

Figure 2 - Cleft lip, post operative.

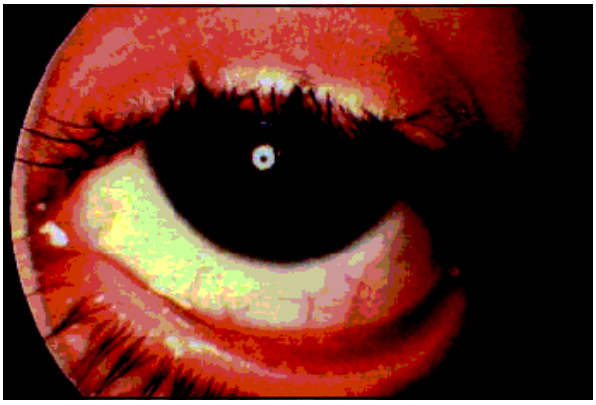

Figure 5 - Close photograph of the left eye showing ectopic pupil, before dilatation.

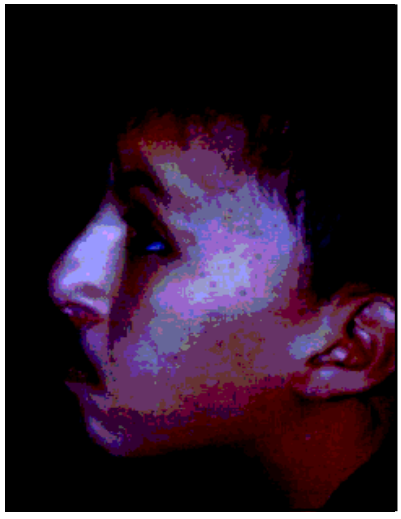

Figure 3 - Side view showing micrognathia with normal external ears.

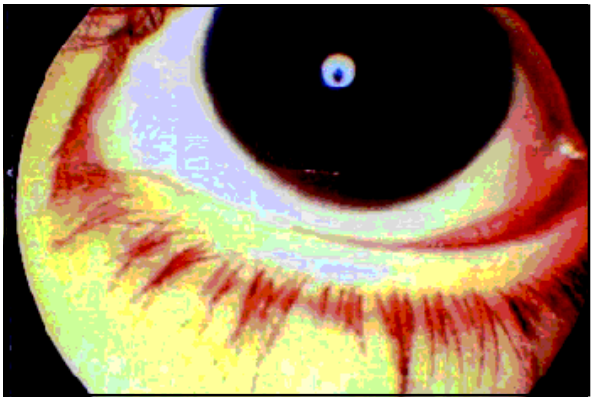

Figure 6 - Close color photograph of the left eye, after dilatation, showing (ectopia lentis et pupillae); downward displaced pupil with clear shadow of upward positioned lens.

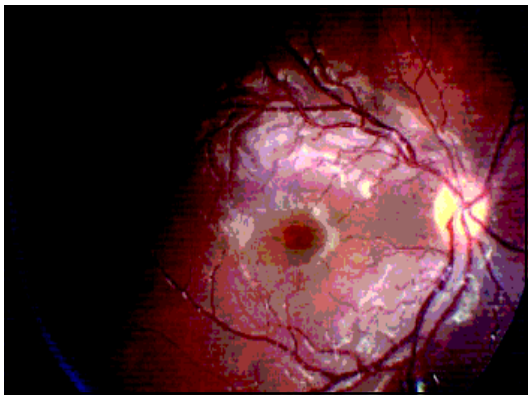

Figure 7 - Colored photograph of right fundus showing normal retina.

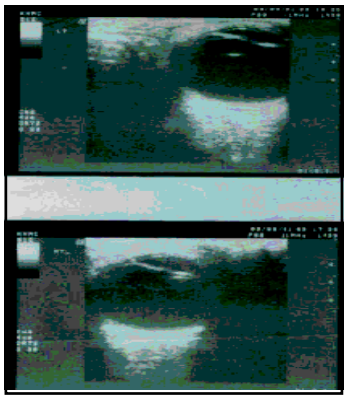

Figure 8 - Echography of both eyes preoperatively showing the shadow of lens displaced superiorly close to the visual axis in the right eye and superiorly away from the visual axis in the left eye.

with the clinical manifestations of our patient. A cleft of the lip and palate are distinct entities closely related embryologically functionally and genetically.[5] Although both may appear to occur sporadically, the inheritance of susceptibility genes allowing the formation of cleft in certain individuals appears important in the formation of clefts.[5] Associated malformations are especially derived from the first branchial arch.[5] Searching through Medline and literature, no association has been reported between cleft lip and palate and ectopia lentis.[1-5]

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