KJO
pISSN: 1011-8942 eISSN: 2092-9382

Korean J Ophthalmol 2010;24(5):314-317
DOI: 10.3341/kjo.2010.24.5.314

Case Report

Two Cases of Mirror-Image Eye Anomalies in Monozygotic Twins

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We report two cases of mirror image anomalies in two different pairs of monozygotic twins. In case 1, the twins exhibited mirroring of strabismus and refractive errors. Twin 1 had 35 prism diopters (PD) right intermittent exotropia at distant fixation and myopic anisometropia that was spherical 2.00 diopters more myopic in the right eye. Twin 2 had 35 PD left intermittent exotropia at distant fixation and her left eye was more myopic by – spherical 1.00 diopters. In case 2, the twins were diagnosed with infantile nystagmus with upbeat jerk. Twin 1 exhibited a habitual head turn of 30° to the left with dampening of her nystagmus in dextroversion. Twin 2 also exhibited abnormal head position, but in his case the habitual turn was 30° to the right. We believe that this is the first report describing mirror imaged intermittent exotropia with anisometropia and infantile nystagmus with opposite abnormal head positions in pairs of monozygotic twins.

Key Words: Anisometropia, Identical twins, Infantile nystagmus, Intermittent exotropia, Mirror image anomaly

Only 8% of twins are monozygotic twins, and the incidence of identical twins is 3/1,000 deliveries. The splitting of the zygote commonly occurs between the 8- and 16-cell stages of embryonic development. If the splitting process is delayed, the mirror-image phenomenon (i.e., expression of features or anomalies on opposite sides) may occur as a result [1]. Such mirror pathologies may result from genetic or environmental factors that occur during the acquisition of the symmetry of the zygote and before its split into two monozygotic embryos [2]. The mirror-image phenomenon is observed only in identical twins, and is observed in approximately 25% of monozygotic twins [3]. The mirror-image phenomenon rarely affects ophthalmic features [1]. To the best our knowledge, there have been no reported cases of intermittent exotropia with anisometropia (case 1) or infantile nystagmus (case 2) in monozygotic twins.

Received: January 4, 2010 *Accepted*: April 13, 2010

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Case Report

Case 1

A pair of 8-year-old twins were confirmed to be monozygotic twins by an obstetrician. The twins were born by Caesarean section at intrauterine 40 weeks, and weighed 2.3 and 2.4 kg. Neither twin exhibited any medical disorders during childhood and there was no known family history of ocular motility disorders.

The twins were treated at a pediatric ophthalmic clinic to evaluate low vision. The uncorrected visual acuity of twin 1 was 0.1 in the right eye and 0.5 in the left eye. The uncorrected visual acuity of twin 2 was 0.4 in the right eye and 0.1 in the left eye.

The corrected visual acuity of twin 1 was 1.0 in both eyes. Cycloplegic refraction was -sph 2.00 -cyl 1.50 axis 180 in the right eye and -cyl 1.75 axis 180 in the left eye. Ocular motility tests revealed 35 prism diopters (PD) right intermittent exotropia at distant fixation and 35 PD alternate intermittent exotropia at near fixation. The worth four dot test revealed right eye suppression at near and distant fixation.

The corrected visual acuity of twin 2 was also 20/20 in both eyes. Cycloplegic refraction was -cyl 1.50 axis 180 in the right eye and -sph 1.00 -cyl 1.00 axis 180 in the left eye.

Fig. 1. Postoperative image of 8-year-old identical twins who underwent 8.5 mm bilateral lateral rectus recessions at 1 month of ago. Before surgery, Twin 1 had 35 prism diopters (PD) right intermittent exotropia, while twin 2 had 35 PD left intermittent exotropia at distant fixation.

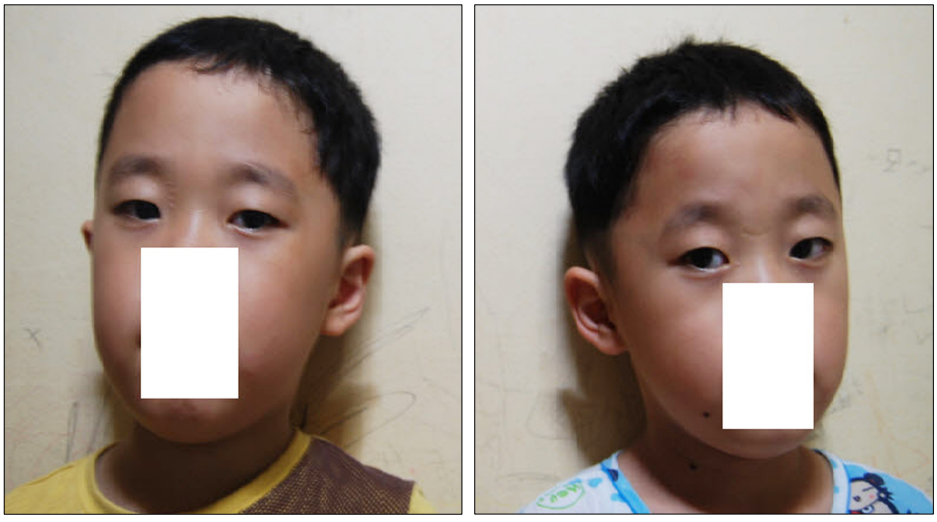

Fig. 2. These identical twins exhibited the up beat jerk type of infantile nystagmus. Twin 1 (right) had a habitual head turn of 30˚ to the left with dampening of nystagmus in dextroversion, while twin 2 (left), had a habitual head turn of 30˚ to the right.

Ocular motility tests revealed 35 PD left intermittent exotropia at distant fixation and 35 PD alternate intermittent exotropia at near fixation. The Worth four dot test revealed left eye suppression at near and distant fixation.

Both twins were prescribed corrective glasses at their first visit. At follow-up 2 months later, both twins showed 35 PD alternate intermittent exotropia at near and distant fixation. After more than 2 months, both twins underwent strabismus surgery with 8.5 mm recession of the bilateral lateral rectus muscles on the same day. At postoperative 1 month, both twins exhibited orthophoria at near and distance (Fig. 1).

Case 2

6-year-old identical twins were referred to our ophthalmology clinic when they were 3 years old due to nystagmus and abnormal head position that was first noted after 1 year of age. The twins had been born at intrauterine week 40 with birth weights of 2.8 and 3.2 kg. Neither twin exhibited any medical disorders after birth except nystagmus. There was no known family history of ocular motility disorders.

The twins were diagnosed with infantile nystagmus. The characters of nystagmus in both twins were frequency of approximately 2 Hz and upbeat jerk type nystagmus that was bilaterally symmetric in both eyes. The nystagmus was dampened at near fixation and showed no response to the horizontal optokinetic nystagmus test. The twins did not exhibit any oscillopsia. The extraocular movement exam revealed orthophoria at near and distant fixation in both twins. The refractive errors of the twins were classified as mild hyperopia in both eyes that did not require correction with glasses.

Twin 1 exhibited a habitual head turn of 30˚ to the left with dampening of nystagmus in dextroversion, increase of nystagmus in primary position, and maximal nystagmus in levoversion. The electronystagmography (ENG) of twin 1 showed that a null zone was present at his right 30˚ gaze, which was consistent with his left head turn (Fig. 2).

Twin 2 exhibited a habitual head turn of 30˚ to the right. The nystagmus was dampened in levoversion, increased in primary position, and maximized with dextroversion. This preferred eye position was consistent with the results of ENG exam, which revealed a null zone at his left 30˚ gaze (Fig. 3).

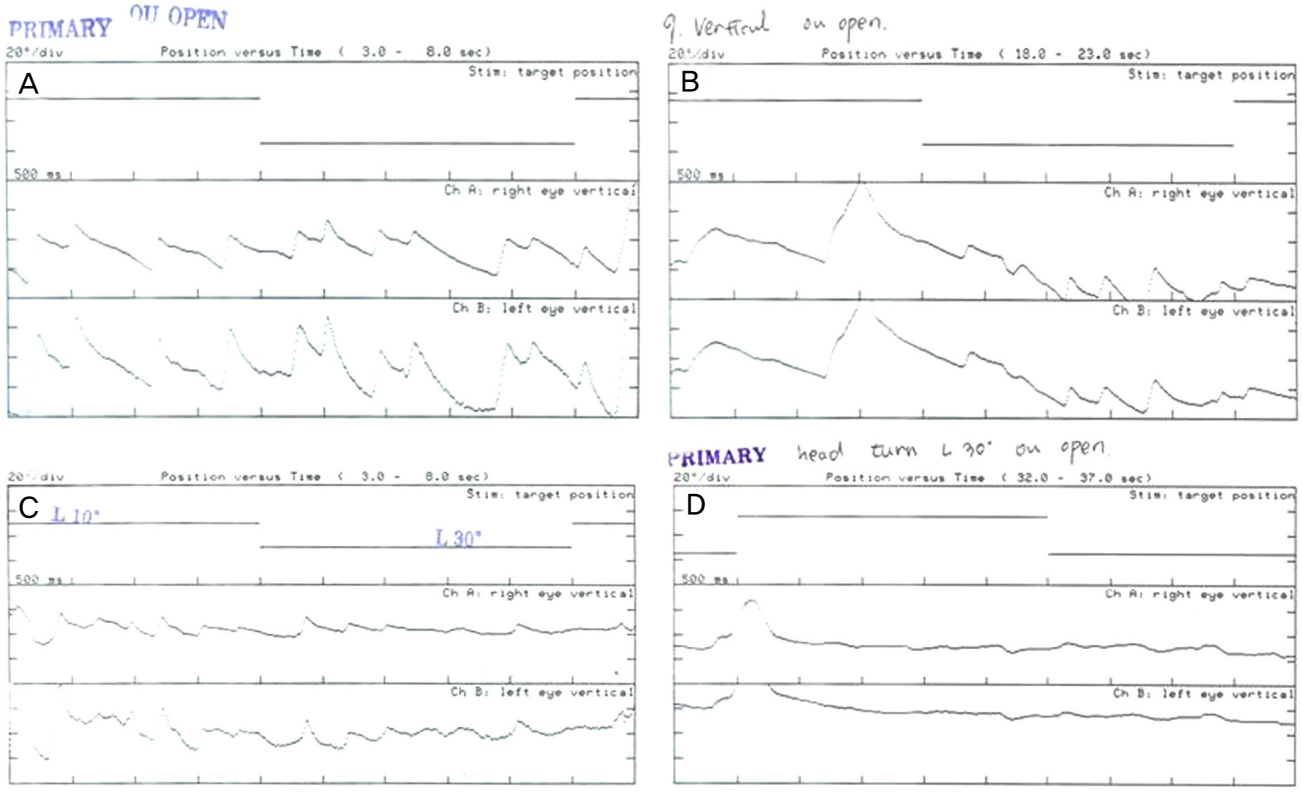

Fig. 3. Vertical electronystagmography of identical twins with infantile nystagmus shows that twin 1 had upbeat jerk type nystamus (B) with dampening of nystagmus in dextroversion (D), and that twin 2 also had up beat jerk type nysgagmus (A) with dampening in levoversion (C). This result is consistent with the preferred abnormal head positions of these identical twins.

The treatment plans for both twins were mirror-image a ugmented Kestenbaum surgery for treatment of their abnormal head positions.

Discussion

In case 1, both twins exhibited mirroring of strabismus and refractive errors. The degrees of exotropia were 35 PD in both, but the deviating eye was the right eye in twin 1 and the left eye in twin 2, which may have reflected the poor visual acuity of the more myopic eye in each twin. After receiving prescription glasses, the visual acuity of both twins was assessed at 20/20 in both eyes and their intermittent exotropia transformed into the alternate type.

In case 1, we treated both twins with 8.5 mm bilateral lateral rectus recession and obtained good postoperative outcomes. However, in case 2, the twins were treated differently, with mirror-image augmented Kestenbaum surgery, for their abnormal head positions.

In case 2, the twins exhibited mirroring of abnormal head positions and preferred eye positions. Even though they both exhibited upbeat jerk type bilaterally symmetrical nystagmus, they exhibited horizontal abnormal head postion. According to Alexander's law, patients with up beat jerk nystagmus are expected to raise their chins. Therefore, the infantile nystagmus observed in case 2 was neither the typical type nor did it conform to Alexander's law [4].

A recent report described a case of mirror image myopic anisometropia in a brother and sister pair in Korea [5]. We believe that case 1 is the first report of mirror image intermittent exotropia with anisometropia, and that case 2 is the first report of mirror image abnormal head position in monozygotic twins with infantile nystagmus. These completely symmetric mirroring phenomena may constitute evidence that intermittent exotropia, anisometropia and infantile nystamus are not acquired diseases, but are genetic in origin. The results of this report may also help resolve the issue of the timing of the extablishment of zygotic symmetry in human embryology.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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