

Concordant Esotropia and Bilateral Hypermetropia in Young Monozygotic Twins

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ABSTRACT

Monozygotic twins with concordant essential infantile esotropia and bilateral hypermetropia are reported. They are discussed in relation to the importance of early surgical

alignment before the age of two years and the influence of high hypermetropia on visual development post operatively.

Keywords: Infantile esotropia, congenital esotropia, hypermetropia, timing, surgery

INTRODUCTION

Essential infantile esotropia, or congenital esotropia, is a large angle esotropia occurring in the first six months of life^{1,2}. It is generally associated with anomalies including dissociated vertical deviation, inferior oblique over-action, latent nystagmus, limitation of abduction associated with crossed fixation and asymmetrical optokinetic nystagmus^{1,2}. The majority of patients require surgical correction of the angle of deviation as it can lead to muscle and conjunctival contracture, and because the size of the esotropia is cosmetically unacceptable¹. It has also been established that patients are likely to develop a subnormal form of binocular single vision, usually in the form of a microtropia, if operated on before the age of two years^{1,3,4}.

The twins who are reported on have concordant (the presence of the same trait in both twins) essential infantile esotropia and bilateral hypermetropic refractive errors. They will be discussed in relation to the genetic influence on the development of essential infantile esotropia and hypermetropia, the importance of early surgical alignment and the influence of hypermetropia on visual development in patients with essential infantile esotropia.

CASE REPORT

Monozygotic twin girls, Twin 1 and Twin 2, were born prematurely at thirty-three weeks gestation. They had a family history of squint and amblyopia in a maternal aunt.

The twins were referred for squint surgery by a private ophthalmologist, and were first seen at fifteen months of age. The parents had first noticed the twins' squints at three months of age. Both twins had been wearing glasses full-time and undergoing occlusion of the amblyopic eye for half an hour per day for the past six months, at the direction of the referring ophthalmologist. Twin 1's current glasses prescription was R +5.00/-2.00x180°, L +4.00/-1.00x180°, and Twin 2's was R +4.50/-1.50x175°, L +4.00/-1.00x100°.

The twins were booked for strabismus surgery at their initial appointment. Results of orthoptic testing were reasonably consistent at each subsequent visit. Findings were as follows:

Twin 1

On examination, Twin 1 fixed and followed a light stimulus and displayed objection to cover of the left eye. Cover test with and without glasses revealed a moderate to large right esotropia which briefly held fixation, measuring 40Δ by Krimsky reflections. Ocular movements showed a slight limitation of right abduction and right inferior oblique over-action. Latent nystagmus was also noted.

Twin 1 was instructed to undertake patching of the left eye for six hours per day, which proved difficult to achieve. Consequently patching was substituted for Atropine 1% to be instilled into the left eye every second day. It was initially found that Twin 1 would not take up fixation with the left eye occluded; however over a period of three months of amblyopia treatment Twin 1's mother began to notice the alternation to a left esotropia on occasion, indicating that the right visual acuity had improved.

Pre-operatively the right esotropia was measured to be 40Δ

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by Krimsky reflections (patient cooperation could not be gained for a prism cover test). A bimedial rectus recession was performed to correct the esotropia at 23 months of age. Post-operatively, an esotropia of 4 Δ was measured by Krimsky reflections. Ocular movements appeared full, the inferior oblique over-action previously noted appearing to have resolved.

Twin 2

Twin 2 also fixed and followed a light stimulus but did not object to cover of either eye, suggesting near equal visual acuity. Cover test with and without glasses showed a small to moderate left esotropia, at times holding fixation/alternating. Measurement by Krimsky reflections with and without glasses was approximately 30 Δ . Ocular movements showed cross fixation on versions, again indicating near equal visual acuity. Twin 2 was instructed to undertake two hours of patching of the right eye per day for treatment of amblyopia, with which she was compliant.

At her pre-operative orthoptic assessment, the size of the deviation was measured by prism cover test to be 40 Δ . A left recess/resect procedure was undertaken to correct the strabismus when Twin 2 was 18 months old. Post-operatively no deviation was apparent upon cover testing. Ocular movements showed a restriction of the left eye on abduction and a right inferior oblique over-action.

DISCUSSION

Twin studies have shown a 94.1% concordance rate of infantile esotropia in monozygotic twins as compared to a 26% concordance rate in dizygotic twins, suggesting a strong genetic component⁵. The higher concordance rate of concomitant strabismus found between dizygotic twins than siblings suggests that environmental factors also play a part in the development of concomitant strabismus⁶. Environmental risk factors for the development of childhood strabismus have been identified to be advanced maternal age, maternal smoking during pregnancy, and low birth weight (<1500g).^{6,7} Hypermetropia has also been shown to be a risk factor for the development of childhood strabismus⁸. A twin study on the genetics of refractive error predicted the heritability of hypermetropia to be 89%⁸. These twins illustrate concordance in both hypermetropic refractive error and essential infantile esotropia.

There are two main theories relating to the development of essential infantile esotropia. Worth's sensory theory states that the development of infantile esotropia is due to a congenital deficit of fusion, and as such the restoration of normal binocular single vision is not possible^{1,2}. Chevasse later put forward the theory that binocular reflexes gradually develop in the first five years of life, and that the development of infantile esotropia is due to interference with the development of the conditioned binocular reflexes^{1,2}.

It has been well established that there is an increased likelihood of patients developing a subnormal form of binocular single vision if operated on before the age of two years, usually in the form of a microtropia^{1,3}. Various studies have advocated that surgical alignment before six months of age results in a finer form of stereoacuity than those aligned after the age of six months^{9,10}, but a weakness to these results is that these studies could not take into account the possibility of spontaneous resolution of the esotropia¹¹. Conversely there has also been a study on the effect of surgical alignment on functional binocular status in adults and children over the age of eight years who previously had had no surgical treatment for their infantile esotropia.¹² The study found that 88% of patients surgically realigned to within 8 Δ of orthotropia who had shown a suppression response pre-operatively on testing with Bagolini striated glasses demonstrated binocular function on testing with Bagolini striated glasses post-operatively¹². Generally it is recognised that the earlier surgical alignment is performed, a higher degree of binocular function will be attained^{9,10,12}.

The presence of latent nystagmus, dissociated vertical deviation (DVD) and amblyopia can be barriers to the proper development of sensory fusion and stereopsis. Consequently, bifoveal binocular single vision is unobtainable after successful surgery for essential infantile esotropia and a microtropia is the expected outcome¹. Early surgery also can lessen any mechanical component from muscle and connective tissue contracture¹.

The measurements of the angle of the esotropia in our twins taken at each visit were reasonably consistent, suggesting that they were reliable enough to base squint surgery on. Surgery to correct the strabismus was booked at the twins' first appointments, with the aim of performing surgery as soon as possible. As the twins were aged fifteen months at their first appointment, it was important that surgery was not delayed beyond their second birthday.

As a result of surgery, Twin 1 was left with a residual esotropia of 4 Δ , a good result considered that ocular alignment tends towards divergence with age. Twin 2's surgical result was of orthotropia, and while this may initially be a good result cosmetically, in the long run a consecutive exotropia may develop due to the tendency for ocular divergence with age if binocular single vision and fusion fail to develop. For this reason a residual esotropia of 5-10 Δ would have been the preferable surgical outcome. As both twins have only recently undergone surgical correction, the long-term stability of ocular alignment is as yet unknown.

It has been advocated that alignment between 15 Δ esotropia and 10 Δ exotropia is acceptable, but a small residual esotropia if 5 Δ -10 Δ is preferred¹. Kushner & Fisher reported that the ideal surgical outcome was that of orthotropia, followed by a residual small-angle esotropia, the least preferable being a consecutive small-angle exotropia¹³. They found

that patients who were orthotropic six months after surgery showed better alignment and binocularity five years after surgery than those patients who were esotropic or exotropic six months after surgery¹³.

A refractive element to the esotropia does not appear to be present in either twin as the size of the deviation, measured by the Krimsky method, was the same with and without glasses. In this case the hypermetropic correction has been prescribed to ensure normal visual development. The prevalence of high hypermetropia (> +2.50D) in essential infantile esotropia has been found to be 14.4%¹⁴. Surgery should be performed to correct the amount of the deviation that the hypermetropic correction does not control after 2-3 months of spectacle wear¹⁴, this being the total deviation in the twins. To prevent the development of a superimposed accommodative esotropia, the hypermetropic correction should be continued to be worn subsequently to surgery due to the presence of subnormal binocular single vision¹⁴.

These twins give an example of early surgical alignment (before two years of age) for essential infantile esotropia. They also demonstrate the co-existence of high hypermetropia with essential infantile esotropia, and its influence on visual development.

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