

## Spontaneous Resolution of Early Onset Esotropia: Two Case Studies

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### ABSTRACT

The case studies of two infants who both presented with a large angle early onset esotropia that resolved completely within the first 12 months of life are presented. Spontaneous resolution of early onset esotropia is uncommon and

these two cases highlight the importance of examining infants carefully and repeatedly prior to early surgical intervention.

**Keywords:** Infantile esotropia, spontaneous resolution, binocular development

### INTRODUCTION

**E**arly onset esotropia, also referred to as congenital esotropia or infantile esotropia, is characterised by the presentation of an esotropia within the first 6 months of life. Children with early onset esotropia typically exhibit minimal refractive error, and can later develop any of the following associated features, including inferior oblique (IO) overaction, dissociated vertical deviation (DVD) or latent nystagmus.<sup>1</sup>

Spontaneous resolution of early onset esotropia is uncommon. Reporting possible cases of spontaneous resolution reiterates the importance of the delicate maturation of the visual system that occurs during infancy and highlights the importance of examining infants carefully and repeatedly prior to surgical intervention. This paper presents two cases of patients diagnosed with early onset esotropia who demonstrated spontaneous resolution.

large right esotropia on corneal reflections which measured approximately 30 degrees using Hirschberg's method. Her visual behaviour showed central, steady and maintained fixation and following ability with each eye, and left fixation preference on cover test.

Miss P's right eye was unable to abduct beyond the midline on ductions whilst presenting a visually attentive stimulus (toy and examiner's face) and by performing post-rotational optokinetic nystagmus testing. There was no significant refractive error, no nystagmus, and no other significant clinical findings. Due to the limitation of abduction, a magnetic resonance imaging (MRI) was performed to exclude any pathology, particularly along the VI cranial nerve, but no pathology was detected.

At aged 8 weeks, Miss P's esotropia measured up to 50 prism dioptres using the Krimsky method. Her right abduction, tested as previously, had improved to beyond midline, and was graded as a -2 limitation. She maintained left fixation preference and treatment of 30 minutes left occlusion daily was recommended.

At aged 3 months, Miss P had been compliant with occlusion as instructed. Her parents had noted a significant improvement in her esotropia. Alternate prism cover test measured a 10 prism dioptre esophoria. Her right abduction was full, with no evidence of amblyopia or suppression. She was able to fuse a 20 prism dioptre base-out prism presented over either eye. It was recommended that occlusion be ceased.

At Miss P's most recent visit, at aged 6 months, she was orthophoric with no evidence of abduction deficit or amblyopia. A normal fusional response was noted on 20 prism dioptre base-out prism testing.

### CASE REPORTS

#### CASE REPORT 1

Miss P presented aged 4 weeks with a history of an esotropia since birth. There was no significant family history, a normal pregnancy and birth. Miss P, other than her right esotropia, was a healthy thriving infant meeting all her developmental milestones

On examination, she was alert and cooperative. All testing was performed for near fixation only. She demonstrated a

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## CASE REPORT 2

Master J presented at aged 5.5 months with a history of an esotropia since birth. There was no significant family history, a normal pregnancy and birth. Master J, other than his esotropia, was also a healthy thriving infant meeting all of his developmental milestones.

On examination, Master J was alert and cooperative. All testing was performed for near fixation only. On cover test, a large alternating esotropia was measured to be approximately 40 prism dioptres using the Krimsky method. His visual behaviour showed central, steady and maintained fixation and following ability with each eye and no fixation preference on cover test.

Ocular motility showed a mild left abduction deficit which was graded as a -1 limitation. Testing of abduction was performed similarly to Miss P. He had no significant refractive error, no nystagmus and no other clinical defect. Due to the limitation of abduction, MRI was also performed however, no pathology was found. Due to the alternating nature of the esotropia, no occlusion was recommended.

Master J was reviewed at 9 months of age. Alternate cover testing for near showed orthophoria. Ocular motility showed full abduction of both eyes. There was no fixation preference and therefore no evidence of amblyopia. A normal fusional response was noted on 20 prism dioptre base-out prism testing.

## DISCUSSION

Spontaneous resolution of an early onset esotropia is uncommon. The most recent Pediatric Eye Disease Investigator Group (PEDIG) study that aimed to identify the probability of spontaneous resolution concluded that 27% of patients are likely to spontaneously resolve by 6 months of age.<sup>2</sup> The study included infants aged less than 20 weeks, examined by an ophthalmologist and diagnosed with an esotropia. Of the 27% of patients who spontaneously resolved, most presented with a variable or intermittent esotropia and measured less than 40 prism dioptres. The authors acknowledged that only one patient with a constant deviation greater than 40 prism dioptres spontaneously resolved, therefore concluding it is less likely that patients with a constant esotropia measuring greater than 40 prism dioptres will spontaneously resolve.

Few cases of spontaneous resolution of early onset esotropia have been reported by other authors. Shon et al<sup>3</sup> reported three cases with spontaneous recovery of early onset esotropia. Their patients presented at less than 6 months of age with an esotropia less than 40 prism dioptres. They resolved at less than 12 months of age. Of particular interest was that with long-term follow-up all three patients later demonstrated poor stereoacuity and the associated phenomenon of DVD and

IO overaction, which developed between the ages of 39 and 59 months.

In our cases, Miss P and Master J presented at ages 4 weeks and 5.5 months respectively. Both infants had a clinically identified large angle esotropia that was constant at their initial examination, and by reports from the parents was present since birth. Both infants were healthy and thriving. Miss P resolved at less than 6 months of age and Master J at less than 9 months of age. Master J's parents reported resolution when he was aged approximately 7 months. The clinical findings suggest there is a high probability that these patients had an early onset esotropia and demonstrated a phenomenon of spontaneous resolution that is rarely observed.

Why did these infants' esotropia resolve? Some key issues for discussion include identifying the characteristics and causes of an early onset esotropia; the pathway that allows for resolution including visual, macular and binocular maturation, which may actually be what hinders ocular alignment. Further issues include the timing of surgical intervention, the importance of carefully measuring the angle of deviation at more than one visit to observe any reduction in angle size, counselling of parents and follow-up of patients with spontaneous resolution in light of the potential for development of poor stereoacuity, DVD and IO overaction.

The aetiology of early onset esotropia is undefined. Ocampo and Foster<sup>4</sup> summarised some schools of thought. Early researchers hypothesised that excessive tonic convergence was a major contributing factor in the development of early onset esotropia. Others suggested that fusion was defective at birth and irreparable which resulted in no drive for orthophoria or binocularity.

Thorn et al<sup>5</sup> and Chino et al<sup>6</sup> later discovered that the necessary neurons for binocularity are present at birth, however they are immature. The maturity of the neurons and associated pathways occurs in stages during infancy, and can be impeded, incomplete, delayed or cease development.<sup>7</sup> Thorn<sup>5</sup> identified the neurons in the primary visual cortex that are involved in binocular functions. These neurons were found to mature at approximately 3 months of age, irrespective of ocular alignment. Fawcett, Wang and Birch<sup>7</sup> also summarised the significant research that has defined discrete stages of binocular development and stages of vulnerability for incompleteness or cessation. They reported that at aged 3 months the average infant's binocular maturity begins and continues at a fast rate until 8 to 18 months. The progress then slows and persists until approximately 3 years of age.

Studies in binocular development suggest that despite a manifest deviation, infants have the potential for developing binocular functions. This is a possible reason for the spontaneous resolution seen in our cases, whereby their binocular development matured such that the drive

for orthophoria was eventually achieved. It is however not possible in a clinical setting to determine the maturity of the binocular cells without a cortical analysis.

Patients with a persistent manifest deviation, require surgical intervention within the "window of opportunity" or crucial stages of binocular development to allow for any potential for developing binocular functions. By classifying the stages of binocular development, this isolates the primary reason why many ophthalmologists and orthoptists advocate early surgical intervention for infants with early onset esotropia; to optimise the potential for developing binocular functions. However, there is some debate that delayed surgical intervention is better for achieving more accurate postoperative alignment due to the improved accuracy in the measurement of strabismus.<sup>8</sup> For our patients, early surgical intervention was discussed with the proviso that the patient fulfilled several criteria prior to intervention. These criteria included that all pathology be excluded, a constant manifest esotropia and stable measurements over at least two consecutive visits. For both Miss P and Master J, any associated pathology was excluded, however, the angle of deviation reduced and resolved and surgery was unwarranted.

Another contributing factor that may influence spontaneous resolution of ocular alignment in infancy is macular maturity. The macular development commences in utero with initial rapid growth in the first 3 months, then slows as the rest of the retina develops. At 8 months the foetus' macula resumes development at the same rate as the rest of the retina. At birth, the macular development is incomplete. It is suggested that macular development is completed by 4 months postnatal age.<sup>9</sup>

If macular maturation is delayed, unstable alignment or strabismus may be evident. It is possible that delayed macular maturation may have occurred with the cases reported. On clinical examination fixation from either eye was central, steady and maintained, suggesting good macula function. Also the macula appeared normal on dilated indirect ophthalmoscopic examination. However, without electrodiagnostic analysis, delayed macular maturation is difficult to determine.

Miss P gave an indication of possibly having delayed macular and visual maturation with the evidence of fixation preference. By occluding the left eye, the right eye was allowed more visual stimulation. This may also have contributed to the drive for orthophoria and spontaneous resolution of her early onset esotropia.

## CONCLUSION

The most likely diagnoses based on the available data and clinical evidence is that both infants exhibited spontaneous recovery of early onset esotropia. Both presented with a constant esotropia at birth and resolution occurred within the critical stages of binocular development. It can be suggested that spontaneous resolution occurred due to the intricate maturing of binocular functions and/or of the macula and thus was sufficient that orthophoria was established. Both children will continue to be monitored with close interest, particularly in regards to the possible demonstration of poor stereoacuity, DVD and IO overaction.

Irrespective of whether these children had delayed binocular and macular maturation, they highlight the importance of keeping in mind that patients initially identified with an early onset esotropia may spontaneously resolve. The occurrence of spontaneous resolution of early onset esotropia also reiterates the advantages of carefully measuring the angle of deviation on more than one occasion prior to surgical intervention.

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## REFERENCES

1. Ansons AM, Davis H. *Diagnosis and Management of Ocular Motility Disorders*. 3rd Ed. Oxford: Blackwell Science Ltd; 2001.
2. Pediatric Eye Disease Investigator Group. Spontaneous resolution of early-onset esotropia: experience of the Congenital Esotropia Observational Study. *Am J Ophthalmol* 2002;133(1):109-118.
3. Shon MA, Hahn KH, Han SH, Hwang JM. Spontaneous resolution of infantile esotropia. *J AAPOS* 2001;5(1):44-47.
4. Ocampo VV, Foster CS. Esotropia, Infantile. 2010 [updated 2010, cited 2010 1st Dec] Available from <http://emedicine.medscape.com/article/1198876-overview>.
5. Thorn F, Gwiazda J, Cruz AA, et al. The development of eye alignment, convergence, and sensory binocularity in young infants. *Invest Ophthalmol Vis Sci* 1994;35(2):544-553.
6. Chino YM, Smith EL, Hatta S, Cheng H. Postnatal development of binocular disparity sensitivity in neurons of primate visual cortex. *J Neurosci* 1997;17(1):296-307.
7. Fawcett SL, Wang YZ, Birch EE. The critical period for susceptibility of human stereopsis. *Invest Ophthalmol Vis Sci* 2005;46(2):521-525.
8. Birch EE, Stager DR. Long-term motor and sensory outcomes after early surgery for infantile esotropia. *J AAPOS* 2006;10(5):409-413.
9. Mukherjee PK. *Pediatric Ophthalmology*. New Delhi: New Age International Publishers; 2005.