Consecutive Exotropia Following Surgically Corrected Cyclic Esotropia

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INTRODUCTION

Cyclic esotropia was first described by Costenbader and Mousel in 1964.1 It is a rare form of strabismus featuring a regularly recurring manifest convergent squint which follows a rhythmic pattern, often occurring on alternate days.2 The condition

ABSTRACT

Introduction: Cyclic esotropia is a rare form of strabismus consisting of regular intervals of esotropia alternating with periods of orthophoria in a rhythmic/cyclic manner. In the vast majority of cases, surgery appears to permanently correct the esotropia, with no sequelae after years of follow-up. We report a case of consecutive exotropia in a patient five years after bilateral medial rectus recessions for cyclic esotropia.

Methods: A case report involving review of a clinical chart.

Results: A two-year-old male presented with right esotropia and mild amblyopia. He was treated with patching and following resolution of the amblyopia he developed a cyclic esotropia. Surgical correction was performed for the full amount measured on a “manifest” day. Following the surgery, he was orthophoric and demonstrated binocular vision. He remained stable for five years, and then returned with occasional diplopia and an intermittent exotropia.

Conclusion: Cyclic esotropia is a rare disorder of ocular motility that spontaneously appears and disappears at regular intervals. After surgical correction, the deviation disappears and recurrence of esotropia is very infrequent. We present the first reported case of consecutive exotropia following surgical correction of a cyclic esotropia.

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adheres to a definite pattern unrelated to visual activity, illness or interruption of fusion. This distinguishes it from other forms of intermittent strabismus. The cyclical nature of the esotropia has many postulates, with the leading one implicating the “biological clock” or circadian rhythm.

Surgery is almost always curative for the condition. Costenbader noted in the first presented cases of cyclic esotropia that when surgery was performed the patients became constantly orthophoric. Moreover, Windsor et al. states that “cyclic strabismus differs from other rhythmic disease states in that peripheral manipulation of the ocular muscles uniformly eliminates the problem.”

Although surgery is corrective of cyclic esotropia for most patients, more recently recurrences of strabismus have been observed. Cahill et al. reported a patient who manifested a recurrence of esotropia in a cyclical fashion after surgical correction. They concluded that their case supports the hypothesis that surgery in cyclic esotropia only serves to remove peripheral signs of an as-yet-unknown central cyclical disorder, analogous to removing the hands of the clock while leaving the clock running. We present a similar yet different recurrence that may also support this conclusion.

CASE

A 25-month-old male was referred to our clinic in 1991 because of intermittent right esotropia. The patient was otherwise healthy and neurologically normal, reaching his developmental milestones. Visual fixation indicated right amblyopia, being uncentral, steady and unmaintained in the right eye and central, steady and maintained in the left eye. Krimsky testing revealed a right esotropia of 20–30°. Ocular versions were full. We were unable to accurately perform tests for binocular vision at this visit. Fundus examination was normal. Cycloplegic refraction was +0.75 in each eye. The patient was started on patching of the left eye three hours per day. On examination one month later, visual fixation was now normal in the distance. The patient continued to manifest a moderate right esotropia of 25°. The patient underwent tapering of his patching.

At follow-up four months later, the patient’s eyes were orthophoric and visual fixation remained normal in each eye. Fusion was demonstrated at near on Worth 4-dot testing, and the patient achieved 550 seconds of arc of stereopsis on Lang II™ testing. However, the patient’s mother had noted a transition for two months alternating between straight and turned eyes on a 24-hour cycle. The patient was then seen on consecutive days in order to assess this cyclical pattern. On a turned day, visual fixation remained normal, with a left esotropia of 35° at near and distance, but with no fusion on Worth 4-dot and no stereopsis on Lang or Titmus® testing. On the straight days, visual fixation was normal, the eyes straight with the ability to overcome 20° base-out on prism vergence testing at near, and binocular vision was present with fusion at near and distance on Worth 4-dot light testing and 200 seconds arc of stereopsis on Titmus® testing (fly positive, 2/3 animals, 0/9 circles).

This cyclical pattern continued for two years and then the patient deteriorated to a constantly present right esotropia now increased in amount to 45°, with a “V” pattern and overacting inferior oblique muscles on ocular version testing, and no stereoacuity. The patient, however, maintained equal visual acuity of 20/30 in each eye. The patient underwent bilateral medial rectus recessions of 6 mm in each eye along with inferior oblique myectomies under general anesthesia. At six months follow-up after surgery, the patient was orthophoric with 20/20 vision bilaterally, and he recovered binocular vision, fusing at near and distance on Worth 4 dot light testing as well as appreciating 100 seconds of arc of stereopsis on Titmus® testing (fly positive, 3/3 and 1/9). The parents were pleased with the results of surgery.

The patient remained orthophoric until December 1999 (five years after surgery), now noticing his left eye drifting out at times with some diplopia. Unaided visual acuity was 20/20 right and 20/70 left, improving to 20/20 left with refraction (+0.25 sphere right and –2.25 sphere left). He showed an intermittent left exotropia of 16° in the distance and 12° at near. No stereopsis was demonstrated. The patient was given glasses, but no other treatment.
DISCUSSION

Cyclic esotropia is a rare condition occurring in one in 3000–5000 strabismus patients. First described by Costenbader and Mousel in 1964, the condition is still somewhat a mystery but has gained new insight in recent years. Cyclic esotropia has generally been described as a regularly recurring manifest convergent strabismus that follows a rhythmic pattern, often appearing on alternate days. It has been postulated that the condition is related to the circadian rhythm or biological clock and that surgery is curative for the condition.

Cyclic forms of strabismus leave many questions unanswered, such as whether strabismus patients can be predisposed to developing a cyclical pattern, whether cyclical strabismus patients are predisposed to developing strabismus later in life, and what etiology underlies this unique disorder. Although recent reports have documented acquired cyclic esotropia with no strabismus history previously, a conclusive analysis will likely require an understanding as to the etiology of the condition. Similarly, determining whether cyclical strabismus patients are truly predisposed to strabismus in future years or whether the risk is equal in all patients will be strengthened when the causative factors are elucidated.

A further question often raised regarding this unique condition, is whether a peripheral or a central mechanism is involved in its manifestation, as well as the role surgery plays in correcting this disorder. Chamberlain, Hess, and others have reported the remarkable and permanent recovery after surgery for this condition, possibly supporting a hypothesis of an underlying peripheral involvement in the etiology of cyclic esotropia. Helveston further added that if a first operation is unsatisfactory, repeat surgeries can produce excellent cosmetic and functional results.

The success of surgery in maintaining orthophoria might suggest a peripheral mechanism underlying this condition; however, reports of recurrence of cyclic esotropia after surgical correction contribute to the theory that there may be a central mechanism to cyclic forms of strabismus. Richter postulated that “ocular surgery removed the hands from the clock without altering the clock itself.” Further supporting the theory of a central mechanism, Cahill notes in his 1999 report of recurrent cyclic esotropia that the case “supports the hypothesis that the underlying central disorder was unaffected by the surgical procedure on the peripheral site” and that in the end “surgery only serves to remove peripheral signs of an as-yet-unknown central cyclical disorder analogous to removing the hands of the clock while leaving the clock running.”

Our case may serve to further support this hypothesis since this is, to our knowledge, the first reported case of consecutive exotropia occurring after surgical correction of cyclic esotropia, although our patient did develop myopic anisometropia sometime after his surgery and this may be a confounding factor. Therefore, the possibility exists that although our patient had good results cosmetically and functionally for five years before reverting to a consecutive exotropia, the surgical treatment only affected a temporary peripheral manifestation of a central process. In other words, our surgery merely removed the hands from the clock, while the clock ticked on.

REFERENCES


**Key words:** consecutive exotropia, cyclic esotropia, strabismus surgery

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