



A Post-Surgical Stereovision Surprise in an Adult With an Exotropia Since Infancy Previously Managed, at Two Years With Surgery

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CASE REPORT

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ABSTRACT

Aim: To describe an unexpected sensory outcome in an adult male who is seeking ocular re-alignment for a psychosocially symptomatic large non-specific exotropia with suppression. The primary diagnosis was infant onset exodeviation of unclear diagnosis, was managed with bilateral strabismus surgery at two years of age, little memory of follow-up.

Result: Measurable binocular single vision (BSV) was demonstrable following surgery at 17 years of age, albeit slowly between two weeks and six months postoperatively and subsequently enhanced. His newly acquired sub-optimal BSV led to symptomatic occupation-associated asthenopia. Following two subsequent operations over a 15-year period, he has stable, symptom-free ocular realignment within three prism diopters of orthophoria and performing tasks that require extended periods of near-vision activity.

Conclusion: Delayed high levels of stereovision were unexpectedly achieved in an adult with infant onset exotropia with pre-operative sensory suppression that was surgically aligned to near orthophoria. The re-establishment of BSV in such a clinical scenario has to attain a level that is robust enough to meet an individual's social and occupational needs.

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INTRODUCTION

The lack of a reliable history is all too common in childhood onset strabismus cases that re-present itself later in life to the adult motility service. This can present management and prognostic challenges in patients with infant onset exotropia, where the spectrum of sensorimotor outcomes following strabismus surgery in adulthood are not well reported, when compared to patients presenting with consecutive exotropia (Lal & Holmes 2002; Morris, Scott & Dickey 1993) or post-surgery deteriorated intermittent exotropia (Baker 2008; Lim et al. 2008). We describe the 20-year management of a male with a surgically treated infant onset exodeviation who re-presented in adulthood with a psychosocially symptomatic exotropia. The latter was surgically managed. He subsequently benefited from protracted ongoing care including two further procedures for a decompensating exotropia associated with a maldeveloped binocular single vision system that impacted him socially and professionally.

CASE REPORT

A 16-year-old male presented to the adult motility clinic with a psychosocially symptomatic constant exotropia, which had been troubling him, especially in photographs, for some years. He never experienced diplopia and requested surgical management of his strabismus. His past ophthalmic history included bilateral lateral rectus recessions for a left divergent squint age two-years. Hospital records of his previous strabismus surgery were not available. The duration of post-operative follow-up was thought to be brief but was otherwise unknown. His parents recall the exodeviation to be present in the first year of life but did not clearly recall whether it was intermittent or constant. He was otherwise fit and well with no medical history of note.

Corrected visual acuity was 0.0 Log MAR in either eye (OD -5.75/+1.50×085, OS -4.50/+0.50×085) with a stable refractive error during follow-up in our department. Slit lamp examination revealed conjunctival scarring over the right lateral rectus. An alternate prism and cover test (APCT) measured 45 prism diopters (PD) exotropia at six meters and 40 PD exotropia for near viewing. Ocular motility testing revealed bilateral minor adduction deficits (*Figure 1*). Synoptophore measurements were negative for simultaneous perception while the Worth 4 Dot test (W4D) found alternating suppression at both near and distance. Peripheral testing was not performed.

The patient underwent surgery, namely a bilateral 4 mm medial rectus muscle resection and right lateral rectus recession of 3 mms'. The lateral rectus muscle was found at 13 mm from the limbus and further recessed to 16 mm from the limbus (Operation 2). As no BSV potential had been demonstrated, simultaneous bilateral inferior oblique muscle weakening was not performed for the mild inferior oblique overaction and asymptomatic V-pattern.

On the first day post-operatively, he was pleased with the outcome of reduced angle 4 PD exotropia for both near and distance. At the second post-operative visit at two-weeks, he had further improved and was able to momentarily control to an 8 PD exophoria for near and distance in primary position but was not able to demonstrate stereopsis, motor fusion or simultaneous perception.

Six months post operatively, at the third post-operative visit, the patient remained pleased with the outcome. The angle in primary position was 14 PD exophoria at 6m and 10 PD exophoria for near (*Figure 2*). The patient could converge to 8 cm then either eye diverged without diplopia. His prism fusion range at near and distance was 4 PD base-out (BO) to 10 PD base-in (BI) with 55" (arc/second) stereopsis using the Frisby Stereo test.

Over the next 15 years, he had two more surgeries (Operations 3 and 4) for recurring asthenopia symptoms (headaches and sometimes blurred vision and/or

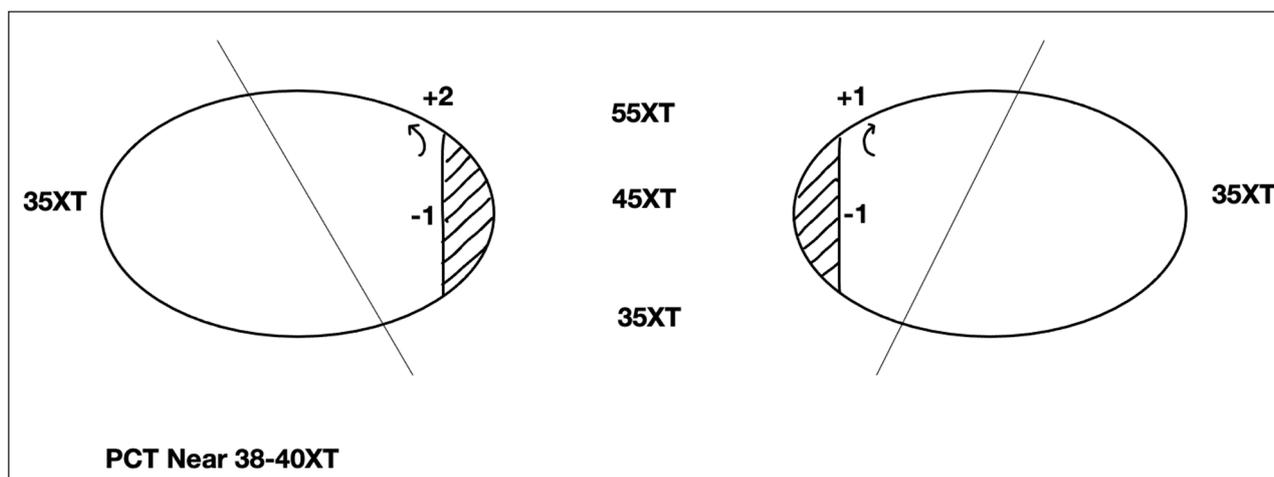


Figure 1 Ocular motility assessment aged 17 years (pre-operation 2), with 6m distance measurements.

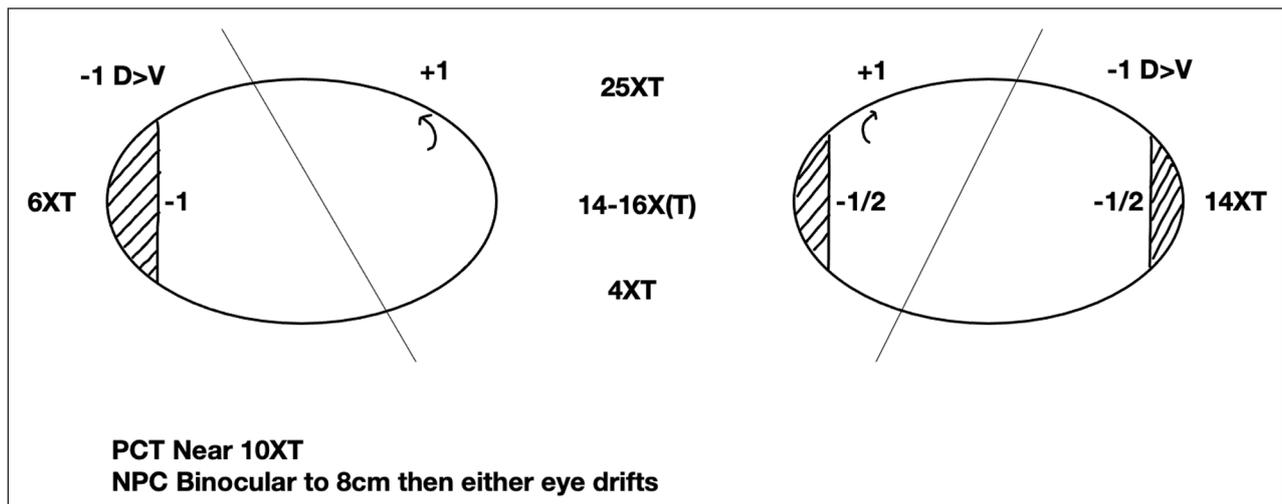


Figure 2 Ocular motility assessment aged 17 years (post-operation 2/pre-operation 3), with 6m distance measurements.

diplopia) related to sustained near-vision tasks whilst working long hours in the software industry. At age 28 years, these included bilateral 10 mm inferior oblique muscle recessions and left lateral rectus re-recession from 12.5 mm to 16 mm's from the limbus (Operation 3) for a decompensating primary position exophoria associated with persistent symptom-producing mild bilateral overacting inferior oblique muscles and with an increase in the magnitude of the up-gaze exotropia. The rationale for symmetrical additional inferior oblique recessions was to collapse the pattern, thereby reducing the larger incomitant up-gaze exodeviation that could exceed his fusional convergence amplitudes. Following Operation 3 he could converge to 12–14 cm before the left eye diverged. At that time, a fusion range at 6 meters could not be measured as the patient decompensated on introduction of a prism. His asthenopia symptoms initially reduced, only to recur circa three years later. At age 32, he decided to proceed with further surgery, namely a 4 mm right medial rectus re-resection (Operation 4) for recurrent symptomatic decompensating exophoria during the course of his aforementioned work (**Figure 3**).

Circa four years following his fourth operation, he remains happy with his sensorimotor status. He has a 2 PD exophoria and 2 PD right hypophoria for both distance and near (**Figure 4**). Bagolini striated glasses showed a BSV response for near and distance in free space. Prism fusion range for near measured 14 PD BO to 4 PD BI. The W4D showed BSV for near and distance alternating at distance with right suppression. His stereopsis measured 55" arc (Frisby), and his convergence was binocular to 6 cm at the last recorded assessment. The small and increased post-operative right abduction underaction may have helped with this ongoing stability in his alignment and symptomatic improvement, albeit at the expense of a slight non-symptoms producing increase in right horizontal gaze incomitance (Deacon et al 2014; Graeber & Hunter 2015).

Following the three strabismus procedures in adulthood his documented stereopsis improved, the chronology of such stereoacuity and surgical intervention is presented in (**Figure 5**).

DISCUSSION

Exodeviations account for approximately one quarter of all childhood strabismus cases (Buck et al. 2009), intermittent exotropia (X(T)) being the most common (Govindan et al. 2005). Other causes include exotropia (XT) associated with abnormalities of the central nervous system, convergence insufficiency, congenital XT and XT associated with sensory deprivation or paralytic pathology (Govindan et al. 2005). There are many studies of childhood onset strabismus re-presenting in adulthood, although not specifically infant onset exodeviations. Previous studies highlight the functional improvement that surgical correction to within 15D in primary position can bring (Dickmann et al. 2013; Tomac et al. 2020; Morris, Scott & Dickey 1993).

The term congenital XT is often used interchangeably with infantile XT, encompassing constant XT's presenting before reaching one-year of age without other ocular pathology in an otherwise healthy child

In our case, we believe the initial differential diagnoses would be limited to infantile XT or intermittent exotropia X(T). Infantile XT in healthy children is far more uncommon than X(T) (Petrunak 2017). Given the close proximity of caregiving tasks for babies and their ability to control at near, it is understandable that the distance exodeviation although present, can be often missed until the child is a little older. In such young patients, it can be difficult to distinguish between these two entities.

Hunter et al. (2001) found that, except for the larger angle observed in congenital XT, distinction between the latter and intermittent exotropia can be difficult in infants

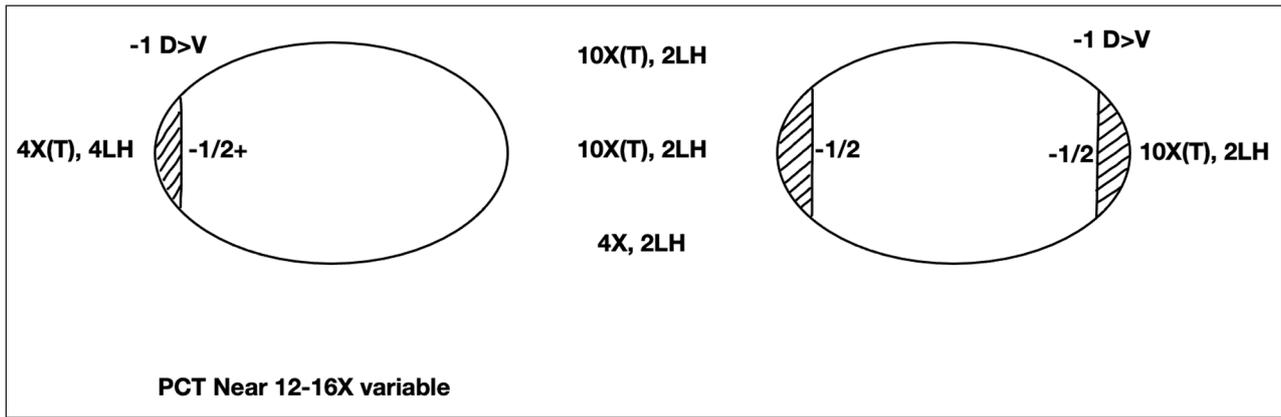


Figure 3 Ocular motility assessment aged 32 years (pre-operation 4), with 6m distance measurements.

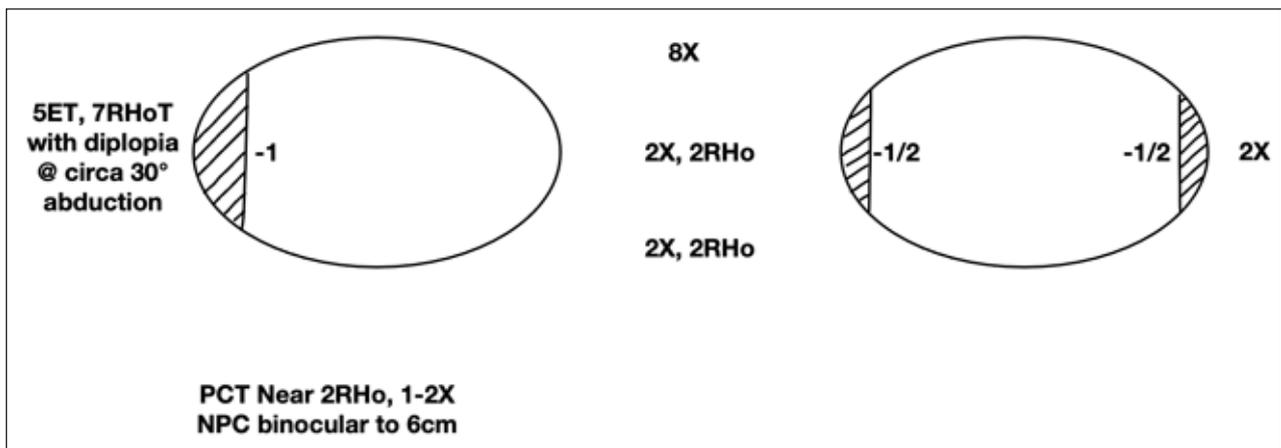


Figure 4 Ocular motility assessment aged 36 years (4 years post-operation 4), with 6m distance measurements.

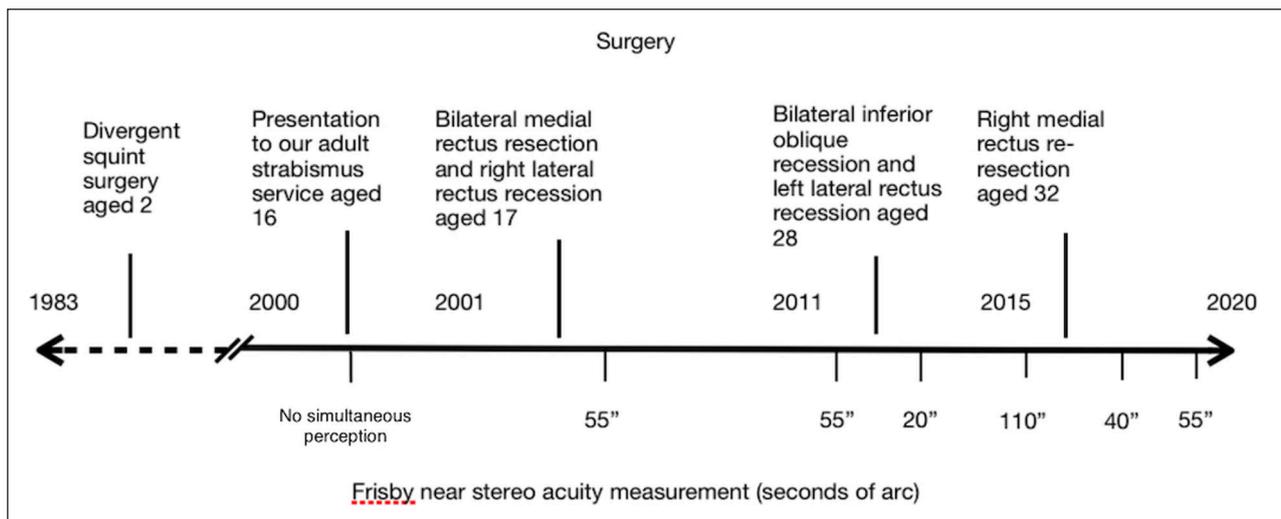


Figure 5 Timeline summary of surgery versus stereoacuity.

presenting within the first 12-months of life, where rates of strabismus associated abnormalities including A- or V-patterns, oblique muscle overaction, nystagmus or dissociated vertical deviation (DVD) are similar between both groups. They raised the question of whether they are simply variable expressions of the same disease where

most infantile XTs' would have an intermittent phase initially while those that are intermittent at presentation can become constant if control is poor.

Maturation of binocular sensory function is almost complete by 18-months of age (Birch 2003), however maturation of binocular vision and susceptibility or

vulnerability to deterioration are two separate entities. Developmentally the critical period for susceptibility of stereopsis extends further, beginning at birth, peaking around three to four months of age with continuing susceptibility throughout early childhood to 4.6 years (Fawcett, Wang & Birch 2005). Our case re-presented at age 16, and despite excellent visual acuity in both eyes, synoptophore assessment did not illicit the potential for binocular single-vision potential that subsequently materialised following realignment surgery. This is a clinical scenario that is perhaps recognised in orthoptic practice more frequently than is reported in the literature. Whilst not often performed in patients with a history of infant onset exotropia, a prolonged Prism Adaptation Test (PAT) compliantly performed in a motivated adult may in this situation have shown potential for BSV by allowing more time pre-operatively to re-establish a BSV response than the synoptophore evaluation (Prism adaptation study research group 1990).

Andalib et al. (2015) performed a review of childhood onset strabismus undergoing surgery in adulthood, specifically looking at prognostic factors for sensory outcomes. This more general, non-targeted review included exotropes and esotropes that were not subcategorised further. All had absent stereopsis pre-operatively. Post operatively, it initially appeared that the exotropes had a greater improvement in stereopsis (55.8%) when compared to the esotropes (41.2%). Similarly the non-amblyopic groups (30.4%) showed greater improvement in stereopsis than the amblyopic group (9.1%), however neither of these were statistically significant. The only prognostic factor identified for improvement of stereopsis was the post-operative surgical alignment of orthotropia. Our case met all three positively reported prognostic factors, namely a non-amblyopic exotrope that achieved post-surgery orthophoria.

Koc & Sefi-Yurdakul (2016) looked specifically at visually mature exotropes. Aside from the intermittent nature at presentation, they found other positive predictors of stereoacuity included onset after one year of age and the absence of aniso-acuity. They also believed alphabet patterns, inferior oblique overaction and vertical strabismus, to have a negative predictive role for a positive fine stereoacuity response (better than 60" arc). Our patient had two negative predictive factors based on the latter authors findings, namely a historic exotropia onset in the first year of life and a modest V-pattern that was managed with strabismus surgery (Operation 3).

Limited data has been published investigating binocularity in infantile XT, specifically comparing constant infantile XT and X(T). However, Saunders & Trivedi (2008) suggest that at least gross binocularity is achievable in both subgroups, with X(T) more able to achieve measurable stereopsis or even bifoveal fixation than XT. Hunter et al. (2001) retrospectively analysed 13 patients with XT presenting before the age of one year and split

their cohort into constant XT and X(T) groups where 92% underwent surgery for constant exotropia or progressive loss of control of X(T). Post-operatively, 27% needed further surgery within the follow up range of 13–158 months. Six of 13 had no BSV, while seven obtained gross BSV, where two of the seven achieved better than 200", both being in the X(T) group. Suh et al. (2013) evaluated the outcomes of surgery in 45 children with early onset XT before one year of age, who they split into constant XT and X(T) subgroups. Ten patients achieved Titmus test stereoacuity greater than 60" of arc, one in the XT versus nine in the X(T) group. These three studies find that a minority of children with a diagnosis of infant onset exodeviation can develop demonstratable BSV following early childhood surgery with seemingly higher grades of stereoacuity achievable in those young children with intermittent exotropia. We believe that our case did at least experience BSV for some period before and/or after his early childhood surgery during the recognized developmental period of stereovision maturation and vulnerability (Fawcett, Wang & Birch 2005), before decompensating to a constant exotropia with establishment of suppression that was reversed by successful motor re-alignment surgery in adulthood. Our case has maintained measurable stereoacuity for almost two decades, with, on each occasion of subsequent surgeries (Operations 3 & 4), a slight reduction in stereovision preceding a further surgical intervention and improvement post-operatively (**Figure 5**). Fluctuations of stereo-acuity, even in those who are visually mature can occur and seemingly there is residual neural plasticity (Andalib et al. 2015).

Convergence fusional reserves are linked to control of X(T) (Fu et al. 2015, Hatt et al. 2011). On initial presentation to clinic, our patient had poor convergence and no fusion. However, following Operation 3, he could converge to 12–14 cm before the left eye diverged. At that time, a fusion range at six meters could not be measured as the patient decompensated on introduction of a prism, consistent with his sub optimal BSV development. Following Operation 4, his binocular convergence and fusional convergence reserves at near had further improved.

Although data is limited and mainly retrospective; a minority of patients with infant-onset exotropia, good visual acuities and no demonstratable pre-operative binocular single vision (BSV) potential can develop gross BSV or even measurable stereovision when managed surgically in adulthood. In our patient, this re-established itself gradually, between two weeks and six months post-operatively. The limited literature suggests that the prognosis is consistently better for the infantile onset intermittent exotropia group. Paradoxically, the successful alignment outcome of our particular patient's psychosocially symptomatic exotropia latterly presented him with secondary asthenopia related BSV challenges largely precipitated by the specific requirements of his

career. This necessitated further surgeries to achieve near orthophoria alignment and binocular convergence to 6 cm so as to enable his restored sub-optimal fusion to better meet the demands of his occupation.

ETHICS AND CONSENT STATEMENT

Written patient consent was obtained for publication.

COMPETING INTERESTS

The authors have no competing interests to declare.

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