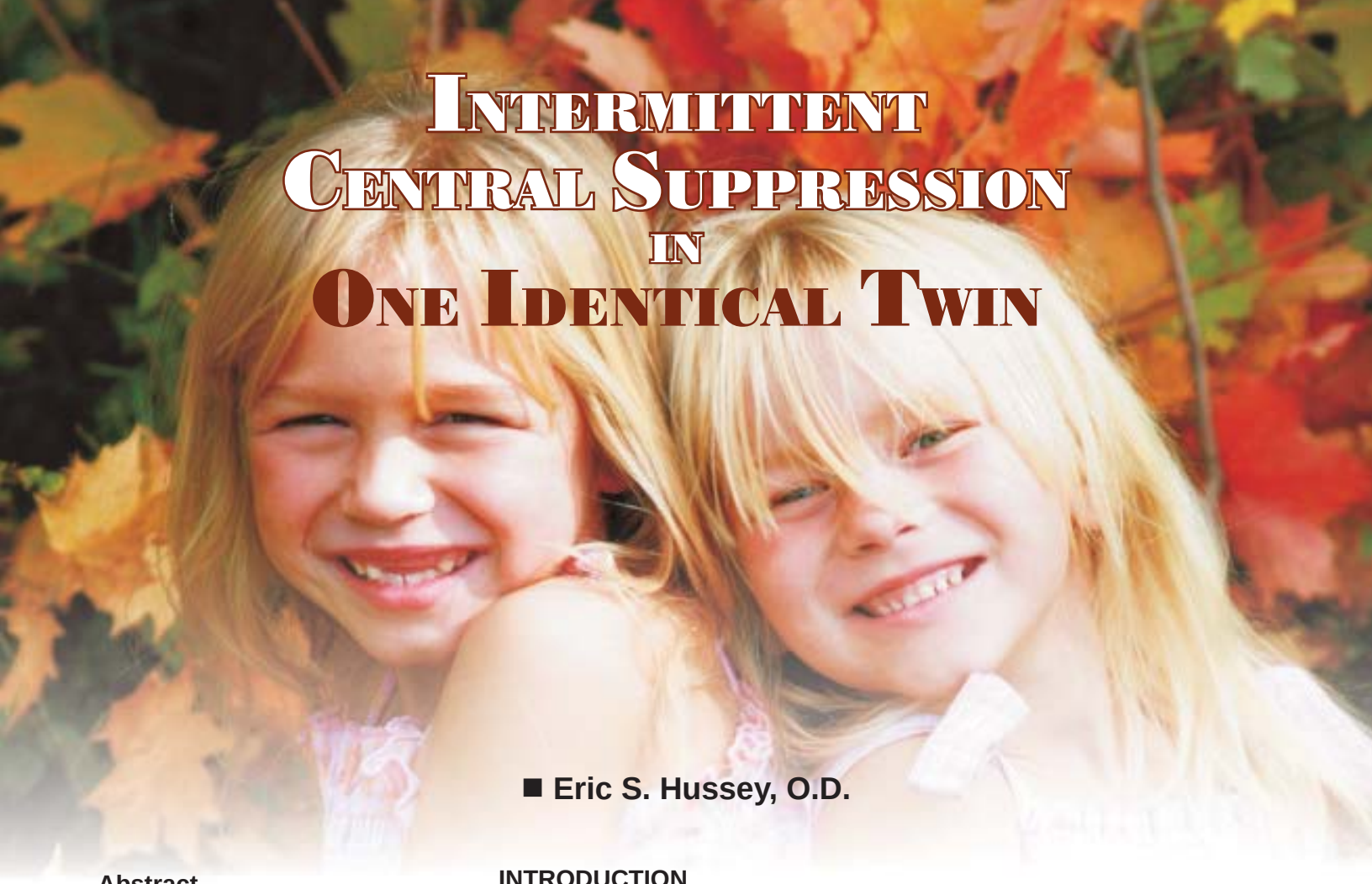

Intermittent Central Suppression in One Identical Twin

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Abstract

Central suppression is found with different types of strabismus and amblyopia. Intermittent central suppression (ICS) is an intermittent loss of visual sensation in the area of central vision occurring in the absence of strabismus and amblyopia. Suppression found in either strabismus or amblyopia is considered a cortical competitive inhibition. The same mechanisms and risk factors that influence suppression in strabismus and amblyopia might be expected to be applicable in ICS. In addition, there is evidence that there are genetic markers for both strabismus and amblyopia. The heritability of ICS, however, has not been reported. A case study is presented. Data, derived from identical (monozygotic) twins shows one twin to exhibit ICS while the other does not, despite inheriting identical genes. This twin data may help to separate the characteristics of ICS from the more constant and possibly more inheritable defect of strabismic/amblyopic suppression.

Key Words

amblyopia, diplopiaphobia, intermittent central suppression, monozygotic twins, strabismus

INTRODUCTION

Intermittent central suppression (ICS) is defined as a repetitive, intermittent loss of central visual sensation in the absence of strabismus and/or amblyopia.[1] ICS has been implicated in a number of problems, from visual dyslexia,[1] to diplopia,[2] to errors in marksmanship,[3] to problems laying bricks;[4] all in patients who demonstrate binocular function and normal, equal corrected acuities.

In strabismus and amblyopia the neurological location of the suppression is identified as the occipital visual cortex.[5] Binocular cells of the cortex are thought to either fuse, or "fight" the impulses coming from the two eyes. Bielschowsky, referring to strabismus and amblyopia, stated (1938 but published in 1943): "There is a fundamental difference between seeing with two eyes and binocular vision."[6] The same should be said concerning non-strabismics with intermittent central suppression (ICS).

The strabismic/amblyopic "fight" occurs when the two eyes' images cannot be fused cortically. The non-matching of the sensory information between the two eyes triggers a suppression, a phobic type response to diplopia. Suppression compensates for the intolerable condition of diplopia and, if early enough in development, potentially produces amblyopia.[5] This sequence of events is thought to start with loss of normal physiologic (retinal) rivalry, with the non-dominant eye losing its image an ever-increasing percentage of the time. The term "diplopiaphobia" was coined by Bielchowsky to describe this cortical desire to remove diplopia.[6]

Suppression is assumed to be compensating for diplopia, but diplopia does not always create suppression. Strabismic suppression appears to be age related. Adult patients suffering from strabismus as a result of traumatic brain injury often exhibit intractable diplopia they cannot suppress.[7,8] For those patients with diplopia resulting from traumatic strabismus, the ability to develop suppression could be very useful in dealing with the abruptly appearing diplopia. Therapeutic interventions for diplopia in later-developing strabismus would only temporarily be necessary. Clinicians could trust that time would remove the second image through development of suppression as a reliable

sequela to the diplopia. Waiting for suppression development would therefore be an appropriate treatment for diplopia in traumatic strabismus.[7]

The inability of adults to develop constant suppression offers evidence that suppression is a problem of binocular development rather than strictly a function of a "cortical on-off toggle switch."[5,9-12] With ICS there might be a lack of visual signal coordination from below the cortex. ICS, unlike a strabismic suppression, would not rely solely on a cortical diplopiaphobia to produce stand-alone cortical neural inhibition. Rather, ICS might include inhibition at the level of the lateral geniculate nucleus.[1,13-16]

Bielschowsky discussed the necessity of adequate and intact sensory stimuli for the development of motor fusion and binocularity.[6] Diplopiaphobic suppression can be considered the result of an error in motor accuracy (i.e., strabismus). The suppression development triggered by motor error prevents the matching of complementary sensory stimuli from the two eyes. Complementary sensory stimuli are considered necessary to develop precise motor fusion. Precise motor fusion, in turn, is required to develop binocularity. The one supports the other. The occurance of strabismus triggers a downhill spiral of inhibition and lack of fusion. This lack of correct neural development begins with the original eye turn. It does not allow normal sensory fusion to develop. If this is true then why, in the absence of strabismus (ICS), would a "phobic" cortical response be expected? This question has yet to be answered. One possiblility is that it could be inherited.

The heritability of strabismus is no doubt multi-factorial. Children whose parents' families included several relatives with strabismus showed an almost 50% strabismus rate, if those children were also highly hyperopic.[17] In contrast, only 10% of the same group showed strabismus if they had small amounts of hyperopia.[17] The American Optometric Association Clinical Guidelines refer to strabismus, amblyopia and high hyperopia as having some genetic basis.[18,19] Monozygotic twins are genetically identical and show 73% concordance for strabismus, whereas non-identical (dizygotic, fraternal) twins show 35% concordance.[20] These percentages suggest heredity is a significant determinant in strabismus. No studies of heredity have been reported on suppression as a sensory defect separate from any strabismus or amblyopia. If strabismus (that is, diplopia) is the triggering factor in the development of suppression, and heredity is a significant determinant in strabismus; then heredity might also be a factor in the development of ICS.

The diagnosis of ICS is found and measured with vectographic test targets at near and at distance.[16,21,22] The targets include the modified Borish diamond target at near and distance targets from the standard distance projected (AO) vectographic adult slide. Individual targets on the adult (AO) distance vectographic slide include OD and OS acuity targets, a vectographic clock-dial astigmatism test, distance fixation disparity, and alternating letter-alternating eye between lines, all within a bilateral framework. Within the limits of patient response abililties and possible pathologies that might be present (e.g., unilateral cataract), all patients are tested with the same battery of vectographic targets, probing for suppression. Not everyone can respond reliably to questioning about targets "disappearing" or "blacking out" with suppression.

If similar ICS findings in identical twins are measured, then this would indicate a possible inherited characteristic. The following case study of monozygotic twins suggests that there is not a strict genetic explanation for non-strabismic intermittent central suppression.

Case Findings

SA and RA are monozygotic, 10-year-old, twin girls who were in fifth grade in September of 2001, when they were examined. Both SA and her identical twin sister, RA, took piano lessons during 2001. SA's piano teacher referred her for a vision examination. Their teacher noticed that RA was progressing in reading music, while SA would not try to read music, preferring to play by memory. Attempts to get SA to read music generated great frustration for the child. Academically SA lagged behind RA in all subjects.

Eye health examinations were within normal limits for both girls and automated visual fields were within normal limits for 10-year-olds. A routine analytical vision examination procedure using vectographic polarized targets was performed. Both girls responded appropriately to vectographic testing that probed for ICS as a primary visual sensory diagnosis, while also testing accommodative and convergence function.[21]

Refractive, accommodative and vergence findings are summarized in Table 1. For these twins, many of the standard analytical test findings were very similar. The most significant differences between the two were:

1) SA's reduced accommodative range- SA was unable to clear 20/20 letters at near in the phoropter without additional plus lens power beyond her distance subjective refraction – and
2) her slightly more prominent fatigue in base-out vergences compared to her twin sister. (The examination routine is to perform three consecutive sets of near vergence findings to assess fatigue effects. Neither child exhibited fatigue in the base in direction.)

Clear differences between the twins are shown in the binocular sensory data, as measured with vectographic binocular targets (Table 2). SA was diagnosed with ICS based upon these findings. Her identical twin sister, RA, did not show ICS on any of the same analytical examination subtests. SA showed an intermittent alternating suppression. Repetitive suppression periods were about two seconds in length spaced by similar periods of about two seconds of simultaneous bilateral sight. Distance stereopsis scores were normal for both girls, and SA's near

Table 1. Refractive, accommodative and vergence summary. Accommodative lens powers are gross spherical powers as read from the phoropter, not net values

		SA	RA
Distance Refraction	OD	+1.00-1.00x100 20/20-	+0.75-0.50x105 20/20-
	OS	+0.75-1.00x75 20/20	+0.75-0.50x75 20/20
Accommodative Range / PRA	(20)	+1.75 blur-out* +2.25 recovery	-0.75 blur-out +0.25 recovery
Base Out Break/ Recovery, near	1st Finding 3rd Finding	24/6 fatigues to 12/-6	30/6 fatigues to 20/6
Base In Break/Recovery, near		18/6 (no fatigue effect x3 sets)	16/4 (no fatigue effect x3 sets)

** Extra plus was needed to obtain 20/20 acuity at near.*

Table 2. Intermittent Central Suppression Diagnostic points			
		SA	RA
Intermittent Central Suppression (ICS) Responses	Modified Borish Diamond	suppression Right side (2 sec off/2 sec on)	No ICS found
	Vectographic Distance Acuity	Suppression OD Acuity Letters	No suppression
	Vectographic Distance Clockdial	Alternate Suppression (more Right side)	No suppression
	Stereopsis	4/4 Distance 9/9 Near (Wirt Rings)	4/4 Distance (near Stereo not tested)

stereopsis score was normal.[22] RA's near stereopsis was not measured.

Each child was prescribed her distance refraction (Table 1). SA's ICS was treated both with traditional anti-suppression therapy procedures such as stereoscope drawing procedures (e.g., cheiroscopic tracings and VO stars) and diplopic eye movement procedures (prism-dissociated rotations), as well as with electronic rapid alternate occlusion procedures.[2,3,4] Analysis of these anti-suppression therapies indicates two fundamental components of anti-suppression therapy. The two components are bilateral sensory input combined with a motion stimulus.[3,4] Electronic rapid alternate occlusion delivers a strong bilateral motion stimulus. The technique uses liquid crystal shutter lenses to create visual flicker motion.[23] Direct alternation between the eyes at approximately 5 Hz is fast enough that the central vision reads the signal as continuous; that is, bilateral.

As the ICS was reduced, accommodative therapies (bilateral accommodative rock procedures) were introduced with successively increased demand. After accommodation normalized, vergence was treated with standard therapies (e.g., the aperture rule[a]).

In-office therapy was carried out in 2002. Twenty-nine sessions eliminated SA's suppression, as well as normalizing both convergence and accommodation function. The academic ability of SA, as well as her ability to read sheet music, improved as therapy was successfully completed. As of July 2008, her twin sister RA continues to show no ICS.

DISCUSSION

What might be causes for one twin to have ICS and not the other? The clinical findings including the refractive status, accommodative resting state (dissociated cross cylinder finding) and near vergence ranges are all quite similar. There appears to be a fatigue factor revealed with repeat testing of positive relative vergence, as that function deteriorates over time. Beyond SA's vergence fatigue, the positive relative accommodation measure is the only finding, other than the ICS, that appears to differentiate the twins.

In some cases, base-out vergence fatigue is associated with ICS. In another case study ICS was viewed as the primary diagnosis.[2] It was treated in isolation using electronic rapid alternate occlusion. Base-out vergences improved with the elimination of the ICS. No other vergence-specific therapy was administered. The presence of ICS may explain some of the difference in vergence findings between these twins. Similarly, some differences in accommodative findings might be expected if there is ICS-induced sensory instability in one twin and not the other.[13]

SA was the twin who was born first. Is it possible that she underwent more birth trauma? Possibly, with the second birth (RA), the birth canal was more widely open. It is also possible that at an early age some accident or environmental event occurred in the home to one child and not the other. Trauma might explain the accommodation-convergence differences since those functions are known to be affected in traumatic brain injury.[8]

Another alternative explanation might be that there are differences in nearpoint stress between the twins and this difference could explain the different accommodative and convergence measures. Both girls however, have attended the same schools at the same time**,** throughout their school careers. They have been reared in the same home with identical free-time activities. Since they are monozygotic twins, their responses to similar nearpoint stresses would be expected to be substantially the same. Those factors would tend to speak against the nearpoint stress explanation for the girls' differences in accommodative and convergence behavior.

Seventy-three percent of strabismic monozygotic twin pairs are concordant for the condition, therefore 27% are not.[20] This difference between twins may be an expression of non-concordance. Given the concordance of the other visual characteristics, this explanation seems less likely, but can't be entirely dismissed.

Some strabismics will alternate fixation and therefore the suppressing eye corresponds to the turned eye. On vectographic testing, the strabismic will typically report seeing only the target associated with the fixating eye. The non-strabismic patient with ICS reports intermittency and alternation of the targets while binocularly viewing vectographic targets.[21,22] Non-strabismic ICS does not appear to be a cortex-based "fuse or fight" inhibition that begins with a vergence error (diplopia). This study suggests that ICS also does not share the hereditary tendency of strabismic/amblyopic suppression.[17,20]

Are ICS and the constant suppression seen in strabismus and amblyopia two different phenomena? Strabismic/amblyopic suppression is constant while ICS shows a temporary loss of the visual percept centrally. The differences in intermittency, alternation and volitional control might be explained if different brain structures serve as the loci of suppression in strabismic versus non-strabismic intermittent suppression. I have suggested ICS occurs at or near the lateral geniculate nucleus (LGN).[1-4,13,16,24] Other studies have shown binocular inhibitory interaction at the LGN that may form a basis for sub-cortical suppression,[14,15] whereas strabismic suppression is considered to be cortical.[5,6,22]

The differences between ICS, and the constant suppression of strabismus and amblyopia might explain the differences in symptomology. ICS is characterized by repetitive, but intermittent central suppressions. The "off-then-on" ICS suppressions could result in intermittent losses of sensory feedback for eye aiming. Repetitive seconds-long losses (seen in ICS) of sensory feedback might allow a fixation drift of the eyes. This drift would exhibit itself clinically as variable and inaccurate eye aiming. Any aiming inaccuracies would require correction during the binocular vision periods, when the suppression has resolved.

Through the time course of ICS, four visual circumstances may be sequentially produced by the ICS and the consequent aiming inaccuracy. These circumstances would be, in order: a suppression of central

visual input monocularly, biocular sensory input but without precise alignment, biocular sensory input with motion as the alignment error is detected and corrected by a vergence movement and lastly, precisely aligned binocularity. Repetitively cycling through those four visual states would create a visually unstable world for those exhibiting ICS.[25]

Visual instability caused by the variable fixation could be expected to cause difficulty with tasks requiring stable perception of detail such as normal reading, or also in reading music. The constant suppression of strabismus, on the other hand, would not be expected to cause a problem with reading since the visual sensation is stable, albeit less than optimal.[22] Constant strabismic suppression would be expected to be associated with the level of deprivation that could produce a clinical amblyopia in the developing visual system.

CONCLUSIONS

The strabismus/amblyopia model for suppression is a cortical competitive inhibition, triggered by diplopia and retinal rivalry ("diplopiaphobia"). This produces one dominant and one cortically-blocked signal. This model does not explain the intermittency or the alternation of the ICS. The strabismus-derived suppression model also fails to explain the production of ICS in an adult apparently from cervical trauma.

This case study of monozygotic twins offers evidence that ICS is not an absolute genetic manifestation. Further, ICS appears to have a different mechanism for suppression than that of strabismus and/or amblyopia.

This case also demonstrates that intermittent central suppression is treatable with specific anti-suppression therapies. The only documented treatment for intermittent central suppression is specific anti-suppression therapy. Anti-suppression therapy is, therefore both appropriate and medically necessary.

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Source

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