Article ▸ Vision Therapy for Convergence Insufficiency Co-Incident with Duane Retraction Syndrome

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ABSTRACT

Background: Duane retraction syndrome (DRS) is a congenital ocular motility disorder characterized by limited abduction and/or limited adduction, globe retraction, and narrowing of the palpebral fissure on adduction. Because of adduction limits, these patients may also exhibit convergence insufficiency (CI). Symptomatic individuals can often benefit from optometric vision therapy (OVT). Although OVT does not treat the DRS itself, these procedural interventions can alleviate symptoms by improving vergence and accommodative ability.

Case: A 10-year-old girl was referred for complaints of asthenopia at near and intermittent horizontal diplopia on side gaze. She exhibited signs of intermittent alternating exotropia at near and poor pursuit eye movements. A visual skills evaluation revealed DRS and CI; with the latter being managed successfully with OVT. Symptoms were relieved entirely with a relatively short course of therapy for the CI, despite the presence of adduction limitations due to congenital mis-wiring of the lateral rectus.

Conclusion: Optometric vision therapy is not a common treatment consideration for patients with DRS, but underlying binocular vision anomalies may cause some of these patients to be symptomatic. Appropriate testing should be performed to reveal these visual skills deficits and referrals should be made for OVT when indicated. It is important to understand that a disease or pathological state can have one or more associated functional overlays that can be successfully treated with a resultant improved quality of life.

Keywords: convergence insufficiency, Duane retraction syndrome, optometric vision therapy, binocular vision

Introduction

Duane retraction syndrome (DRS) is an uncommon but well recognized condition, occurring in about 1-4% of all cases of strabismus.[1-3] It is caused by a congenital mis-wiring of the lateral rectus by aberrant innervation from cranial nerve III, which results in a co-contraction of the lateral and medial recti.[2-4] The syndrome is typically diagnosed at an early age and often is simply monitored. In cases where there is a large strabismus and/or compensating head posture, surgery may be indicated to attempt to improve the head turn, but there have been no clinical trials supporting the use of surgery for DRS.[1,2] Yoked or compensating prisms are also accepted treatment options in some cases.[5-7] There have been only a few reports in the literature suggesting optometric vision therapy (OVT) as a management option for these patients.[5,6]

This case highlights a patient with a more subtle presentation of DRS that had been previously unrecognized. A careful evaluation of ocular motilities to reveal any non-concomitancy was critical to proper diagnosis and patient education in this case, and should be emphasized. The importance of educating DRS patients on all treatment options, including OVT, is evident. No treatment can restore normal ocular motilities in all fields of gaze, but OVT may be a desirable option for patients seeking a functional improvement by eliminating symptoms and improving signs caused by associated binocular vision disorders.

Case Report

A 10-year-old girl, SA, was referred to Pacific University's Vision Therapy Service by her primary care optometrist. The referring optometrist noted signs of intermittent alternating exotropia at near and poor pursuit eye movements. SA complained of asthenopia at near and intermittent horizontal diplopia on side gaze. She reported that although she enjoyed reading and was performing well in school, she felt that her eyes were not working together well. Her eyes would tire easily and hurt or feel sore while reading, more noticeably with the right eye than the left eye. This would cause her to lose her place and re-read the same lines. Ocular history was positive for anisometropia corrected with glasses. Her ocular health was unremarkable. SA's personal medical history was remarkable for depression currently treated with fluoxetine.

At the initial office visit, entering visual acuities through her habitual spectacle correction prescribed one year ago were:

OD . . . . -1.75 -0.25 x 180 . . . . . 20/200

OS . . . . . +0.50 -0.25 x 005 . . . . . 20/40

A trial frame assessment of visual acuities through her updated spectacle correction from her primary care examination two weeks prior was:

OD . . . . -3.50 -0.25 x 180 . . . . . 20/20-2

OS . . . . . -0.50 -0.50 x 010 . . . . . 20/20

Near visual acuities through this refraction were 20/25 OD, 20/25 OS, 20/20 OU. Extraocular muscle version testing

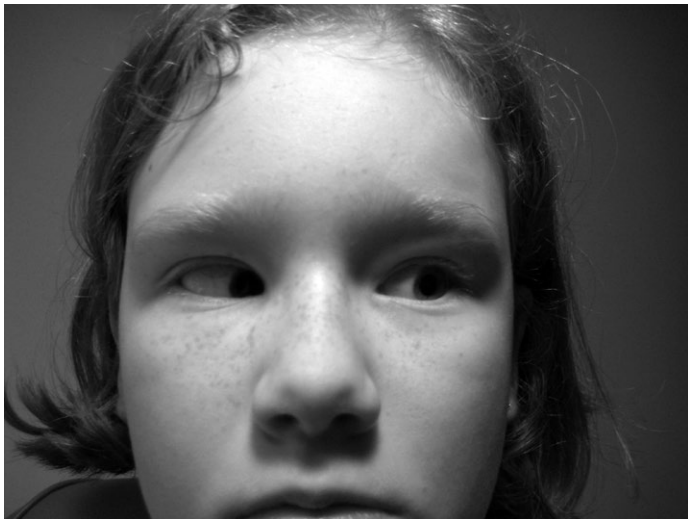

Figure 1: *Limited abduction OS in left gaze (graded 2 on 4 point scale)*

Figure 2: *Mild adduction deficit OS with globe retraction and narrowing of palpebral fissure on right gaze*

showed full range of motion of the right eye but revealed an abduction deficit (graded as 2 on a 4-point scale), widening of the palpebral fissure on attempted abduction, and globe retraction with narrowing of the palpebral fissure on adduction of the left eye (Figures 1 & 2). This presentation is consistent with DRS Type I. Pupils were equal, round, and reactive to light with no APD. Confrontation visual fields were full OD, OS.

Vision skills testing was performed over two office visits. At the initial visit, minimal binocular vision testing was performed through a control lens that had the most recent spectacle correction. Convergence insufficiency (CI) was suspected based on exophoria greater at near than far, reduced near point of convergence, and inadequate positive fusional vergence ability at near.

SA presented to her second office visit for additional visual skills testing after wearing her new spectacle prescription for three weeks. She reported an improvement in her symptoms at near with the new glasses. The Convergence Insufficiency Symptom Survey (CISS) was administered. A score of 16, which is the lowest score in children considered to be symptomatic for convergence insufficiency was noted. Her best corrected far visual acuities through the new spectacles were 20/15-2 OD and 20/15-3 OS. Pertinent binocular vision and accommodative findings are summarized in Table 1. Other accommodative testing showed normal values. A Developmental Eye Movement Test (DEM) was administered to test for oculomotor dysfunction, and the patient was found to have appropriate automaticity and oculomotor skills.

The patient was diagnosed with DRS Type I and CI. The diagnosis of CI was based on her symptomatic CISS score, exophoria greater at near than distance, decreased positive fusional vergence range, and difficulty with plus lenses and base out prism on facility testing. During our consultation with the patient and parents, we explained that DRS is a congenital anomaly that is typically monitored with no active treatment; however, CI is a binocular vision dysfunction that has been shown to be most effectively treated with optometric vision therapy. We explained that although SA's symptoms were considered borderline with her new spectacle correction, our findings showed deficiencies in her visual skills which could potentially make her more symptomatic over time. We recommended an office-based OVT program with an estimated treatment time of 10 sessions. The treatment prognosis was judged to be good to excellent for eliminating symptoms of CI. In addition, we explained that it is possible that some of SA's symptoms may be due to the aniseikonia induced by the three diopters of anisometropia corrected for by spectacle lenses. We recommended that the patient return to her primary care optometrist for a contact lens fitting to correct more effectively for the anisometropia by minimizing the image size difference between the eyes. The family agreed to pursue OVT and to consider the contact lens fitting.

Active Vision Therapy

A program of office-based vision therapy was initiated and continued for eight weeks. Office visits involved a five minute review of the previous week's home therapy and approximately 40 minutes of active vergence and accommodative activities. Home therapy consisted of 20 to 30 minutes of prescribed therapy daily. SA's compliance with home activities was excellent.

The following sequence of activities was performed over the eight weeks of active therapy through SA's updated full-time spectacle correction. The therapy plan was consistent with those described in binocular vision texts by Griffin and Grisham[8] and Scheiman and Wick.[9]

Phase I: Gross convergence and monocular accommodative activities were prescribed initially. Brock string techniques including smooth bead push-up, bead jumps, and imaginary bug on a string were gradually achieved. Monocular accommodative rock (MAR) was built up to +2.25D and

Table 1: Clinical findings at initial evaluation and therapy outcome at completion of therapy and at 3 month progress evaluation

	Initial Evaluation	8 week Progress Evaluation	3 month Progress Evaluation
Cover Test	1-2 ΔXP at far 30Δ XP′	2Δ XP at far 14Δ XP′	2Δ XP at far 20Δ XP′
Near Point of Convergence (break/recovery)	8cm/20cm on 1st trial TN on 5th trial with effort	1cm/4cm x 5 repetitions	3cm/6cm x 5 repetitions
Vergence ranges (by prism bar at near)	BI: x/25/10 BO: x/4/2	BI: x/30/20 BO: x/40/25	BI: x/25/20 BO: x/35/25
Vergence facility (8Δ BI/BO)	6 cpm BO side more difficult	10 cpm Equal difficulty with BO/BI	10 cpm Equal difficulty with BO/BI
Accommodative facility (+/-2.00)	7 cpm OU Plus side more difficult	8.5 cpm OU Equal difficulty with +/-	9 cpm OU Equal difficulty with +/-
Stereoacuity	Randot Stereotest Forms : 250 arc seconds Circles : 20 arc seconds	Super Stereotest: 1A : 20 arc seconds 2A : 30 arc seconds 3A : 20 arc seconds	Super Stereotest: 1A : 20 arc seconds 2A: 20 arc seconds 3A : 30 arc seconds

XP = Exophoria; TN = To Nose; BI = Base-In; BO = Base-Out; cpm = cycles per minute

-6.00D lenses performed at eight cycles per minute. For this activity, SA initially used three-letter groupings as the target. She progressed to using age appropriate reading material by first switching lenses with every word and later by switching every sentence and then paragraph for more of a sustained focus.

Phase II: Relative vergence was introduced using variable Tranaglyphs at near. SA began therapy using variable Tranaglyphs and progressed to more central, detailed targets. Positive and negative fusional vergence ranges were worked on in equal time allotments. Once strong smooth fusional vergence ability was attained, jump demand was emphasized using "look-aways" and later using "BIM/BOP" (minus lens "rocks" with base-in and plus lens "rocks" with base-out training). VTS3 computerized vergence therapy (HomeTherapy Systems, Gold Canyon, Arizona) was also performed in-office as a reward for good compliance with traditional therapy activities. Random dot targets were emphasized and presented equally base-in and base-out. Binocular accommodative rock (BAR) was concurrently trained using the patient's chosen reading material and built up to +/-2.00D at 8.5 cycles per minute.

Phase III: Open space vergence training was accomplished using the aperture rule trainer and lifesaver cards. Both fusional convergence and divergence abilities were emphasized in equal time allotments. Opaque lifesaver cards were used for chiastopic training, while transparent lifesaver cards with a file folder septum (similar to a Remy Separator) were used for orthopic training. Additionally, SA was instructed to perform the lifesaver cards with horizontal head movements side to side to help expand her range of single binocular vision.

Therapy Outcome

A progress evaluation on the eighth week of office-based therapy revealed improvement in all visual skills areas to normal values. These improvements as compared to the initial clinical findings are summarized in Table 1. CISS was repeated and showed an improvement in symptoms with an asymptomatic score of 10.

SA was released from active therapy and home maintenance therapy was prescribed. This home therapy consisted of open space fusional convergence and divergence using lifesaver cards. She was instructed to perform this activity one to three times per week for three months with "look aways" to maintain jump vergence ability and head movement side to side to expand range of single binocular vision.

Three Month Progress Evaluation

SA reported comfortable vision through her habitual spectacle correction and ability to perform home maintenance therapy one time per week with ease. The CISS was re-administered and confirmed that SA was asymptomatic with a score of 4. Clinical findings remained relatively stable, as summarized in Table 1.

SA reported having a contact lens evaluation with her primary care optometrist the week prior. Her parents asserted that the contact lens fitting would be completed in the next month, but they were concerned about whether rigid gas permeable or soft contact lenses were a better choice. We encouraged a spherical soft contact lens fitting and discussed the option of soft multifocal lenses if an attempt at myopia control was a goal of contact lens treatment in addition to correction of anisometropia to reduce aniseikonia.

SA and her parents were educated on the stable findings and success of her OVT program and released from our care. We recommended that she continue maintenance therapy with the lifesaver cards one time per month and to return to our OVT service only as needed if asthenopia or difficulty with maintenance therapy occurred. Finally, we encouraged

her to continue her contact lens care and annual complete eye examinations with dilated fundus exams through her primary care optometrist.

Disussion

Duane retraction syndrome is a congenital anomaly of the sixth cranial nerve and nucleus, resulting in limited abduction and/or adduction of the affected eye and globe retraction and narrowing of the palpebral fissure on adduction.[1,2] Females (58%) are more commonly affected than males, and it occurs more frequently in the left eye (59%) than the right eye, although it can also be seen bilaterally (18%).[1] Most DRS cases have no other congenital abnormality; however, these patients do have a 10-20% greater risk of various other ocular lesions and congenital malformations.[1-3] Examples include crocodile tears, ocular dermoids, morning-glory syndrome, hearing defects, and Goldenhar syndrome.[1,3]

The retraction phenomenon of DRS had previously been thought to be structural in origin; however, it is now accepted to be caused by an absent or maldeveloped 6th cranial nerve nucleus in the brainstem.[2,3] Any structural anomalies have been shown to be secondary.[2] During the 4th-8th week of embryonic development, agenesis of the abducens nucleus and nerve results in anomalous innervation of the lateral rectus (LR) by cranial nerve III (CN III), allowing for co-contraction of the horizontal recti.[2,3] The degree of transfer of CN III fibers intended for the medial rectus (MR) to the LR determines what type of presentation the patient exhibits.[4] The traditional classification of DRS provided by Huber can be understood by the extent of aberrant innervation to the LR.[3,4] In DRS Type I, the most common classification seen clinically, there is a small amount of fiber transfer to the LR. When co-contraction occurs, these patients are able to adduct the affected eye, but have limited abduction due to the weak innervation of the LR. DRS Type III involves a greater transfer of nerve fibers, resulting in a more equal distribution to the MR and LR, which presents clinically as both limited abduction and adduction. Finally, the limited adduction with near-normal abduction seen in DRS Type II requires a large nerve fiber transfer. In this rare presentation, the LR over-powers the MR on co-contraction.

The continuum that exists between the three DRS types has brought up the question of abandoning the Type I, II, III classification system in recent years, as it is now understood that all presentations are truly Type III to some degree.[2-4] Our patient was initially diagnosed with DRS Type I due to an abduction deficit with retraction of the globe and narrowing of the palpebral fissure on adduction. On subsequent examination, there appeared to be a mild adduction deficit as well, indicating that she may have been more appropriately diagnosed as Type III. Careful examination of ocular motilities is likely to reveal both abduction and adduction deficits to some degree in most DRS patients.[2,4]

Accepted treatment options for DRS include monitoring, prism, and, in certain cases, surgery.[1,2,5,6] Prism can be prescribed as fusion prism to eliminate diplopia or in a yoked form to help reduce a compensating head turn.[5] Surgery is typically reserved for significant strabismic deviations in primary position or anomalous head positions that are cosmetically or functionally problematic.[1,2] Jampolsky suggests, "The risk/benefit ratio in DRS is considerably different than in usual strabismus surgeries."[2] Due to the abnormal adduction and abduction patterns from anomalous innervations of the LR, significant over-corrections may occur unexpectedly. Because of the increased risk of unexpected outcomes with surgery, other treatment options are often favored.[1,5] In many cases, patients are simply educated about the congenital origin of the disorder and monitored. Several authors have reported success with OVT in patients with DRS.[5,6] Griffin and Carlson reported on a case of bilateral DRS where the patient achieved both subjective and objective improvements with OVT.[6,8] Cook writes that "the goal is to obtain the strongest possible fusion in primary gaze, and the largest possible zone of binocular vision" and he "has found that it is possible to: 1) increase both sensory and motor fusion in primary gaze, 2) counsel the patient on how to avoid cosmetic issues, and 3) increase the range of motion in some cases."[5]

Our case supports the importance of testing for binocular vision anomalies in symptomatic patients, even if another ocular problem is uncovered that may account for the subjective complaints. Entrance testing and refraction revealed DRS and anisometropia, to which the patient's symptoms may have been fully attributed. Utilization of the CISS as a tool to quantify symptoms, as well as performing appropriate testing to assess the accommodative and vergence systems, is critical to proper diagnosis and patient education.[10,11] It should be noted that although the prognosis for CI treatment with OVT is normally excellent, we were slightly more guarded due to the presence of an innervational anomaly with DRS. Signs that the patient's prognosis was positive were the improvement of her receded NPC on repetition and the excellent stereopsis that she exhibited. These findings indicated that she was already able to converge and achieve single binocular vision with effort, but that these skills could be strengthened with training to facilitate her visual comfort.

Summary

The current understanding of DRS supports the likelihood that all DRS patients exhibit both abduction and adduction deficits to some degree. Although the innervational etiology of the syndrome prevents the possibility of curing DRS by any means of treatment, the presence of adduction deficits with concurrent CI cannot

be ignored. Some of these individuals may be symptomatic and effectively managed with OVT, as shown by the excellent outcome with a relatively short course of vision therapy for our patient. Optometric vision therapy is not a common treatment consideration for patients with DRS, but this case and others support the recommendation of OVT for symptomatic patients.

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