Article • Optometric Vision Therapy in the Management of Amblyopia Associated with DRS-Type II: A Case Report

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ABSTRACT

Background: Duane retraction syndrome (DRS) 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. Management to restore full-fledged ocular motility has been elusive.

Case Report: An 8-year-old male, RK, visited our clinic with the complaint of outward deviation of the left eye, headache, and asthenopic symptoms. Upon examination, he was diagnosed with DRS-Type II with amblyopia. Vision therapy was started, which lasted for 3 months. A post-vision therapy binocular vision assessment showed marked improvement of visual acuity and other binocular parameters despite the presence of adduction limitations.

Conclusion: Symptomatic patients with DRS may have amblyopia with compromised binocular vision functions. Optometric vision therapy must be considered as a first-line treatment, which helps patients to improve oculomotor control (saccades and pursuits), accommodative and vergence skills, stereopsis, visual-motor skill, and spatial processing.

Keywords: amblyopia, binocular vision, Duane retraction syndrome, optometric vision therapy

Introduction

A rare but well-known congenital restrictive strabismus entity is Duane retraction syndrome (DRS), which is a type of congenital cranial disinnervation disorder (CCDD).[1] It is characterized by co-contraction of the horizontal recti on attempted adduction, causing globe retraction along with variable amounts of upshoot or downshoot.[1,2] Globe retraction in patients with severe ocular motility limitation was first described by Heuck;[3] later Stilling,[4] Turk,[5] Bahr,[6] Sinchair,[7] Wolff,[8] and others provided detailed descriptions of DRS. The detailed description summarizing the findings made by previous studies was published by Alexander Duane in 1905,[1] for whom the retraction syndrome has been named. However, in European literature, the syndrome is referred to, perhaps more appropriately, as Stilling-Turk-Duane retraction syndrome.

DRS is believed to result from a failure of innervation of the lateral rectus muscle by a hypoplastic or absent sixth nerve nucleus, due to which a paradoxical innervation[9] to the lateral rectus is generated by the axons that supply the medial rectus. A common teratogenic stimulus during the 8th week of gestation[10] has been of etiologic significance in sporadic cases of DRS, although autosomal dominant inheritance patterns[11,12] have been noted, along with chromosomal abnormalities.[13,14] Despite being of rare occurrence, DRS is thought to account for 1-5%[15] of all strabismus, about 0.1%[16] among the general population. It is unilateral in 85% of cases; most often the left eye is affected in comparison to the right eye.[15] Bilateral cases are usually associated with asymmetric involvement. DRS has a female preponderance (60%).[16]

The retraction syndrome has been classified on the basis of the type of strabismus present as exotropic DRS, esotropic DRS, and orthotropic DRS.[17] The simplicity of this classification makes it favorable for surgical planning. However, a more-accepted classification system based on electromyographic results was proposed by Huber,[18] categorizing DRS into three classes: DRS-Type I, DRS-Type II, and DRS-

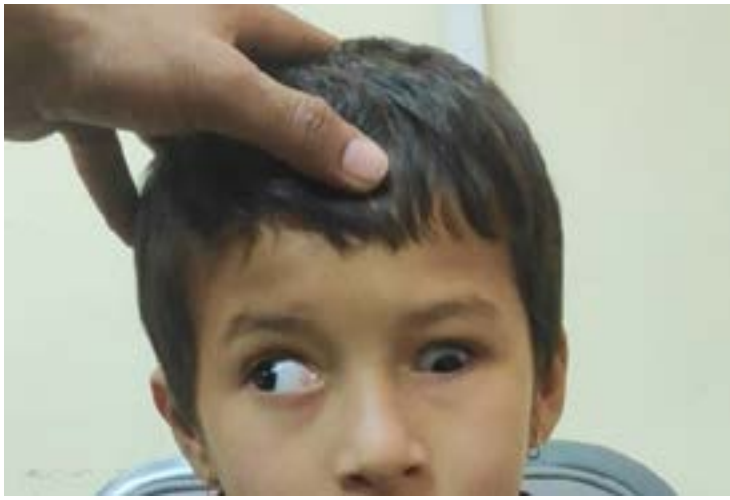

Figure 1. *Limited adduction OS in right gaze with narrowing of the palpebral fissure*

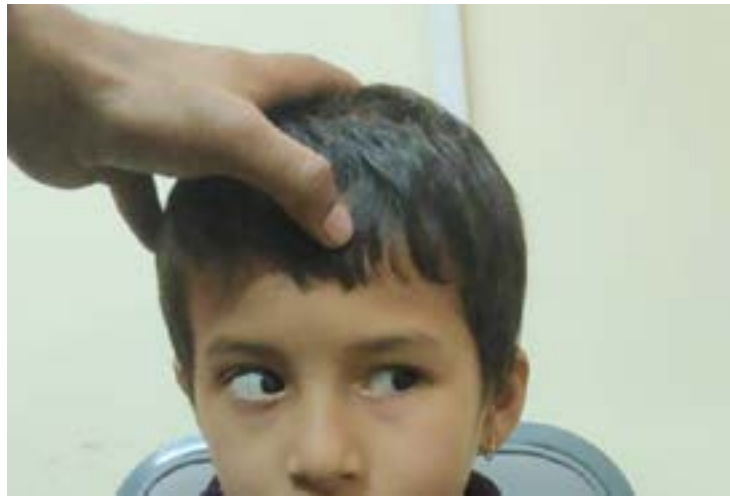

Figure 2. *Normal abduction of the left eye*

Type III. The order of frequency is Type- I (78%), Type III (15%), and Type II (7%),[19] rendering DRS-Type II to be rarest among all.

The management approach for DRS has been focused primarily on surgical methods in the presence of abnormal head posture and significant deviation in primary position, although the results are often disappointing; therefore, no clinical trials have supported the use of surgery for DRS.[20,21] There have been only a few reports in the literature suggesting optometric vision therapy (OVT) as a management option for these patients.[22,23] Here, we present a case report on DRS-Type II with amblyopia that was managed successfully with optometric vision therapy (OVT). Our case is unique in the sense that our patient is male (despite the female preponderance) presenting with a Type II variety (the least frequent type). This necessitates the importance of educating DRS patients on all treatment options, including OVT. Although no treatment can restore normal ocular motilities in all fields of gaze, 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

An 8-year-old male, RK, visited our clinic with the complaint of an outward deviation of the left (OS) eye since birth. RK's parents noted that he was reluctant to perform any near activities, including reading and writing, as he soon got headaches and eyestrain. An abnormal head tilt was also noted by the parents. There was no history of any ocular or birth trauma or family history of strabismus.

On general examination, the patient seemed otherwise normal, except for the occasional head tilt towards the right. The visual acuity was 6/6 OD and 6/9 OS, with no pinhole improvement, suggestive of amblyopia. While taking the distance binocular visual acuity, we noted a strong right eye fixation preference, with a face turn to the right. Dry retinoscopy revealed low myopia OU. The anterior segment and posterior examination were normal, except for a grade I RAPD OS. The colour vision and contrast sensitivity, measured with Ishihara pseusoisochromatic test plates and the Pelli-Robson contrast sensitivity chart, respectively, were found to be normal. The cyclopentolate was administered, and wet retinoscopy was performed, which revealed plano OD and -0.75 DS OS.

On the second visit, post-mydriatic treatment revealed plano OD with 6/6 and -0.50 DS OS with 6/9. He was given a spectacle Rx to wear full time. After two weeks of adaptation, a comprehensive binocular vision assessment was perfromed. Extraocular muscle version testing showed full range of motion of the right eye but revealed an abduction deficit and globe retraction with narrowing of the palpebral fissure on adduction of the left eye (Figures 1 & 2). Based on the binocular vision assessment, the patient was diagnosed with DRS-Type II and amblyopia. Table 1 shows the results from the binocular vision assessment.

Management Plan

The management plan included eye alignment in the primary position, elimination of the abnormal head posture and amblyopia, and improvement of overall binocular function. Office-based vision therapy, 12 sessions per month, was initiated.

Vision Therapy

Each office therapy session lasted one hour. Visual acuity was measured on every alternate session. Binocular vision was assessed every month. Oculomotor therapy (fixation, saccades, pursuits, eye-hand coordination), monocular acuity activities OS, and monocular accommodative activities OS

Table 1. Pre- and Post-Vision Therapy Binocular Vision Evaluation

Test	Initial Evaluation	Post-VT Evaluation (1 month)	Post-VT Evaluation (3 months)
Visual acuity (aided)	OD: 6/6, N6 OS: 6/9, N6	OD: 6/6, N6 OS: 6/6, N6	OD: 6/6, N6 OS: 6/6, N6
Stereoacuity	400 seconds of arc	250 seconds of arc	50 seconds of arc
Cover test	D: 15$^{\Delta}$ LXT N: 20$^{\Delta}$ LXT	D: 10$^{\Delta}$ LXT N: 15$^{\Delta}$ LXT	D: 10$^{\Delta}$ LXT N: 10$^{\Delta}$ LXT
Amplitude of accommodation	OD: 10 D OS: 8.3 D OU: 10 D	OD: 12.5 D OS: 10 D OU: 12.5 D	OD: 13.3 D OS: 12.5 D OU: 14.28 D
Accommodative facility (+/-2.00)	OD: 10 cpm OS: 8 cpm OU: 10 cpm	OD: 13 cpm OS: 10 cpm OU: 14 cpm	OD: 16 cpm OS: 15 cpm OU: 16 cpm
Near point of convergence	8 cm	7 cm	6 cm
Negative fusional vergence (NFV)	D: ×/8/6 N: ×/12/8	D: ×/10/8 N: ×/14/10	D: ×/14/12 N: ×/20/16
Positive fusional vergence (PFV)	D: ×/10/8 N: ×/14/10	D: ×/16/12 N: ×/16/12	D: ×/16/12 N: ×/20/14
Vergence facility (8$^{\Delta}$ BI/BO)	7 cpm	9 cpm	12 cpm

and OD were started, followed by binocular activities for vergence and yoked prism therapy to reduce a compensatory head turn. The patent underwent a total of 35 therapy sessions over a period of three months. Upon completion of the 35th session, his visual acuity improved to 6/6 OS. Appendix A shows the sequential vision therapy program. A comprehensive binocular vision assessment was done at the end of the therapy sessions. Table 1 shows the comprehensive binocular vision assessment pre- and post-vision therapy.

Post-VT, RK was given maintenance therapy for vergence (opaque eccentric circles without the help of a pointer, pencil push-ups, Brock string twice daily) and accommodation (accommodative rock OD and OS separately, twice daily).

After the three-month follow-up visit, the improved visual acuity and other parameters were well maintained.

Discussion

DRS 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.[21] The aims of management in DRS include the elimination or improvement of any abnormal head position (face turn), the treatment of significant or slight ocular misalignment, the reduction of severe globe retraction, and the improvement of upshoots and downshoots. Surgery is typically reserved for significant strabismic deviations in primary position or anomalous head positions that are cosmetically or functionally problematic.[21] Due to abnormal adduction and abduction patterns, the risk/benefit ratio in DRS is considerably different than in usual strabismus surgeries.[21] Because of the increased risk of unexpected outcomes with surgery, other treatment options are often favored.[22] 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.[22-24] Griffin and Carlson reported on a case of bilateral DRS where the patient achieved both subjective and objective improvements with OVT.[23] Tira et al. reported a case of convergence insufficiency associated with DRS that was successfully managed using OVT.[24]

The goal in OVT is to obtain the strongest possible fusion in primary gaze with the largest possible zone of binocular vision. Treatment of DRS must go beyond surgery and include fixation stability, oculomotor control (saccades and pursuits), accommodative skills, contrast sensitivity, stereopsis, visual motor skills, and spatial processing.

The prevalence of amblyopia in DRS patients ranges from 3% to 48% in the literature.[4,9,10] In our case, amblyopia was present, which was successfully managed using active OVT. Our case supports the importance of doing a comprehensive binocular vision evaluation in patients with DRS. Symptomatic patients must undergo binocular vision evaluation to determine whether any binocular anomalies are present. Well-designed OVT could help to improve overall binocular vision skills.

Conclusion

Although OVT is not a common treatment consideration for DRS, it will definitely help the patient improve their binocular function and overall visual

performance by increasing both sensory and motor fusion in primary gaze. DRS patients might present with amblyopia and binocular vision dysfunctions, with both abduction and adduction deficits to some degree. Effective OVT will help in improving visual acuity as well as overall binocular skills. The symptomatic patient can also be successfully managed with optometric vision therapy. One should do a comprehensive binocular and accommodative assessment before initiating therapy. An excellent outcome can be obtained through a well-designed vision therapy program.

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Dahal M, Shyangbo R, Kaiti R. Optometric vision therapy in the management of amblyopia associated with DRS-Type II: A case report. Optom Vis Perf 2022;10(1):26-30.

Appendix: Sequential In-Office Vision Therapy Program

Sessions 1-7
- 12-pointed star with Russell rings
- Wolff wand activities for fixation
- Wayne fixator (near-far-near)
- Hart chart
- Balance board activities
- Walking rail with distance Hart chart
- Pegboard activities
- Pointer-in-straw
- Swinging Marsden ball
- Monocular loose lens rock with plus & minus with near acuity-appropriate word rock card

Sessions 8-14
- Chalkboard circles and swirls
- Wolff wand pursuits
- Balance board activities
- Walking rail with distance Hart chart
- Pegboard activities
- Pointer-in-straw
- Hitting Marsden ball with VMC stick
- Marsden ball with minus lens
- Brock string
- Monocular loose lens rock with plus & minus with near acuity appropriate word rock card

Sessions 15-21
- Walking rail with distance Hart chart
- Pegboard activities
- Pointer-in-straw
- Hitting Marsden ball with VMC stick
- Yoked prism (all directions)
- Marsden ball with minus lens
- Tranaglyphs: convergence only
- Aperture rule: single aperture, convergence only
- Brock string
- Red/green bar reading with red/green goggles

Sessions 22-28
- Mechanical rotator
- Tracking lifesaver card
- Balance board activities
- Walking rail activities
- Chalkboard activities
- Yoked prism (all directions)
- Tranaglyphs: both convergence & divergence
- Aperture rule: single aperture with plus lens binocularly
- Brock string with base in & base out prism
- Red/green bar reading with red/green goggles
- Monocular loose lens rock plus & minus in each eye
- Binocular accommodative facility
- Bernell-o-scope
- Pegboard with balance board
- Marsden ball with VMC stick

Sessions 29-35
- Yoked prism (all directions)
- Tranaglyphs: both convergence & divergence, followed by Vectogram
- Aperture rule: single aperture with plus lens binocularly followed by double aperture
- Brock string with base in & base out prism
- Red/green bar reading with red/green goggles
- Monocular loose lens rock plus & minus in each eye
- Binocular accommodative facility
- Bernell-o-scope
- Pegboard with balance board
- Marsden ball with VMC stick