

Education Article

Challenges in Refractive Correction in Duane Retraction Syndrome

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Article info:

Received: 22 Feb 2026

Accepted: 29 Apr 2026

Citation: Khorrami-Nejad M, Nashee Jaber H. Challenges in Refractive Correction in Duane Retraction Syndrome. *Journal of Modern Rehabilitation*. 2026;20(3):?-?

Running title: Refractive Correction in Duane Retraction Syndrome

Abstract:

Duane Retraction Syndrome (DRS) is a congenital cranial dysinnervation disorder characterized by limitation of horizontal ocular movements, globe retraction, and narrowing of the palpebral fissure on attempted adduction. Although surgical alignment has traditionally been emphasized in the management of DRS, refractive abnormalities and amblyopia represent critical determinants of long-term visual outcome that are often underrecognized. Large clinical series demonstrate that unilateral involvement predominates, with type I being the most common subtype, and consistent female and left-eye preponderance. Across studies, refractive errors are frequently encountered, with hyperopia and hyperopic astigmatism most commonly reported, although myopia, astigmatism, and anisometropia are also prevalent. Amblyopia affects approximately one-fifth of patients overall and occurs more frequently in bilateral disease. Both strabismic and anisometropic mechanisms contribute, reflecting the combined impact of ocular misalignment, suppression, and unequal refractive input during critical periods of visual development. Accurate refractive assessment in DRS poses unique clinical challenges. Abnormal head posture, ocular motility limitation, co-contraction of horizontal recti, and fixation instability—particularly in children with amblyopia—may compromise the reliability of subjective refraction. Subtle interocular differences in unilateral cases can be amblyogenic and require careful detection. Careful refraction, with meticulous attention to head position and fixation stability, is therefore essential. Early identification and timely optical correction are fundamental to preventing avoidable visual

impairment. Refractive management should be regarded as a cornerstone of comprehensive DRS care, complementing surgical and orthoptic strategies to optimize visual development and functional outcomes.

Keywords: Duane retraction syndrome; refractive error; amblyopia; anisometropia; abnormal head posture

Duane Retraction Syndrome (DRS) is a congenital cranial dysinnervation disorder characterized by limitation of horizontal eye movements, globe retraction, and narrowing of the palpebral fissure on attempted adduction. First described by Duane in 1905, DRS accounts for approximately 1–4% of all strabismus cases and most commonly presents as a unilateral condition with left-eye and female predominance.(1, 2)

Large clinical series have demonstrated that unilateral involvement constitutes nearly 80–90% of cases, with type I being the most prevalent subtype, followed by types III and II.(3-5) Bilateral DRS occurs in approximately 10–20% of patients and differs from unilateral disease particularly in the pattern of primary position deviation, with esotropia more frequently observed in bilateral cases.(4, 5) Beyond ocular motility abnormalities, DRS is commonly associated with refractive errors, amblyopia, abnormal head posture, upshoots and downshoots, and occasional ocular or systemic anomalies.(4, 5)

Although surgical strategies and alignment outcomes have been widely discussed, refractive aspects of DRS have received comparatively less emphasis. Given the congenital onset of DRS and the importance of early visual development, optimal refractive assessment and timely correction are critical. This editorial reviews the epidemiological profile of DRS and focuses on the challenges in refractive management in these patients.

Epidemiological and Clinical Profile of DRS

Multiple large series have clarified the demographic and clinical spectrum of DRS. In a study of 331 patients, unilateral disease accounted for 88% of cases, with type I representing the majority of unilateral presentations.(4) A larger series of 441 patients similarly reported unilateral involvement in 88% and bilateral involvement in 12% of cases, with type I being the most common subtype.(5) Female preponderance and left-eye involvement are consistent findings across studies.(4, 5) In unilateral DRS, types I and III show significant left-eye dominance, whereas type II demonstrates no consistent laterality.(3-5)

Upshoots and downshoots are frequent clinical features, particularly in types II and III.(4) Abnormal head posture is also common, especially in unilateral disease, serving as a compensatory mechanism to maintain binocular single vision.(6) Long-standing abnormal head posture may lead to facial asymmetry, particularly contralateral to the direction of head turn.(7)

These epidemiological patterns are clinically relevant because they influence refractive evaluation, amblyopia risk, and management strategies.

Refractive Errors in DRS

Refractive errors are common in DRS and may significantly influence visual development. Hypermetropia is frequently reported, although myopia and astigmatism are also observed.^{4, 6} In a 441-patient series, 31.5% had hypermetropia or hyperopic astigmatism, whereas 22.2% had myopia or myopic astigmatism.(5) Some studies have also highlighted the prevalence of anisometropia in DRS.(8, 9)

Anisometropia may arise from differences in axial length or corneal curvature between affected and unaffected eyes. Chronic co-contraction of horizontal recti and globe retraction could theoretically influence corneal biomechanics, potentially contributing to astigmatic changes. Although definitive causality remains unproven, clinical observations suggest a higher cylindrical component in some unilateral cases.(10)

Importantly, refractive error in DRS may not correspond to the degree of ocular deviation. Even orthotropic patients can exhibit significant ametropia. Therefore, refractive assessment should be systematic and independent of alignment status.(11)

Amblyopia: Prevalence and Mechanisms

In a large cohort of 582 patients, amblyopia was identified in 20.1% overall, affecting 18.5% of unilateral cases and 36.5% of bilateral cases, indicating a markedly higher risk when both eyes are involved.(9) Within unilateral DRS, amblyopia occurred in 16.4% of Type I, 14.9% of Type II, and 19.5% of Type III patients, without substantial intertype variation.(9) Similarly, in a series of 312 patients, amblyopia was observed in 19.3% of cases, with a slightly greater frequency in Type III DRS.(10)

Earlier literature also documented considerable variability. Kirkham reported amblyopia in approximately 25% of patients and highlighted a strong association with anisometropia.(10) Conversely, Tredici and von Noorden described lower rates of amblyopia (3%) and anisometropia (17%), suggesting heterogeneity among populations.(8) In a smaller retrospective study, anisometropia was present in 16% of patients, and amblyopia was uncommon, primarily attributable to anisometropic mechanisms.(12)

More recent large-scale data emphasize the contribution of both strabismus and refractive asymmetry. In the 582-patient review, strabismic amblyopia was the predominant subtype (62.4%), although 37.6% of amblyopic patients also exhibited anisometropia.(9) Mechanistically, chronic ocular misalignment, suppression, and unequal refractive input during critical visual development likely interact to impair cortical visual maturation.

Given this substantial and multifactorial risk, comprehensive cycloplegic refraction and early amblyopia screening remain essential in all DRS patients, particularly in bilateral disease and in those with significant anisometropia.

Challenges in Accurate Refraction

Refractive management in DRS is uniquely challenging for several reasons:

- 1. Abnormal Head Posture:** Patients often adopt a compensatory face turn to maintain binocular fusion.(6) Performing refraction without correcting for habitual head posture may lead to measurement errors. Cycloplegic refraction should ideally be performed in primary head position while ensuring stable fixation.
- 2. Ocular Motility Limitation:** Restricted abduction or adduction can interfere with fixation during retinoscopy and subjective refraction. Co-contraction during attempted gaze shifts may cause transient instability, complicating accurate endpoint determination.(10)
- 3. Fixation Instability and Amblyopia:** Children with amblyopia may exhibit eccentric fixation, further reducing reliability of subjective responses.(13) Objective techniques such as cycloplegic retinoscopy and autorefractometry under controlled conditions are critical.
- 4. Bilateral Asymmetry:** In unilateral DRS, asymmetry between eyes may be subtle but clinically significant.(9) Small anisometropic differences can be amblyogenic and should not be underestimated.

Conclusion

DRS is a complex congenital ocular motility disorder with distinct epidemiological and clinical patterns. While surgical management has traditionally been emphasized, refractive abnormalities and amblyopia represent critical and potentially modifiable determinants of long-term visual outcome.

Unilateral and bilateral DRS differ in alignment characteristics and demographic distribution, yet both carry significant risk of refractive error and amblyopia. Abnormal head posture, motility restriction, and fixation instability pose practical challenges in obtaining accurate refraction.

Early identification, meticulous cycloplegic assessment, and timely optical correction are essential to prevent avoidable visual impairment. Refractive management should therefore be considered a cornerstone—not an adjunct—of comprehensive care in patients with DRS.

Acknowledgements

Artificial intelligence applications (GPT-5.2) were utilized for English language editing to improve clarity, grammar, and overall readability. However, the authors conducted all scientific content and interpretations without relying on AI tools.

Authors' Contributions

Both authors contributed to the conceptualization of the editorial topic. Masoud Khorrami-Nejad, drafted the initial manuscript. Both authors approved the final version of the manuscript and agree to be accountable for all aspects of the work.

Funding

No funding was received for the preparation of this editorial.

Competing Interests

The authors declare that they have no competing interests.

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