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CASE SERIES

Type I Duane's Retraction Syndrome: Clinical Spectrum, Motility Profiles, and Individualized Management Outcomes — A Case Series of Six Patients from a Tertiary Eye Care Centre

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Abstract

Background: Duane's retraction syndrome (DRS) is a congenital cranial dysinnervation disorder accounting for up to 4% of strabismus presentations. Type I DRS, the most prevalent subtype, is characterised by abduction limitation, globe retraction, and palpebral fissure narrowing on adduction. Misdiagnosis as acquired abducens palsy remains a clinical challenge.

Methods: Six consecutive patients with clinically diagnosed Type I DRS presenting to a South Indian tertiary eye care centre (January 2024–December 2025) underwent comprehensive ophthalmic evaluation including nine-gaze motility assessment, prism bar cover test, cycloplegic refraction, and standardised photographic documentation.

Findings: Five females and one male (age range 11–39 years) were included. Bilateral involvement was present in four patients. All six demonstrated the cardinal triad of Type I DRS. Adduction upshoot was present in four patients (66.7%). Primary position deviation included orthotropia (n=4), exotropia (n=1), and esotropia (n=1). Five patients were managed conservatively; one underwent medial rectus recession achieving orthotropia at three-month follow-up.

Interpretation: Most Type I DRS patients are managed conservatively. Surgery, reserved for primary position deviation and amblyopia, yields excellent functional and cosmetic outcomes without restoring motility.

Key words: : Duane's retraction syndrome, congenital cranial dysinnervation disorder, strabismus, abduction limitation, globe retraction, palpebral fissure narrowing, adduction upshoot, medial rectus recession, ocular motility, case series

1 | INTRODUCTION

Duane's retraction syndrome is a congenital, non-progressive ocular motility disorder first described by Alexander Duane in 1905 and classified by Huber in 1974 into three types based on the pattern of duction restriction. The underlying pathophysiology involves developmental agenesis or hypoplasia of the abducens nucleus and sixth cranial nerve, with consequent aberrant innervation of the lateral rectus muscle by a branch of the oculomotor nerve. Upon attempted adduction, simultaneous co-contraction of the medial and lateral recti causes retraction of the globe into the orbit, producing the pathognomonic palpebral fissure narrowing that distinguishes DRS from all other causes of horizontal strabismus (1, 2).

Type I DRS — characterised by marked limitation of abduction with relatively preserved adduction — constitutes 75–80% of all DRS cases and accounts for up to 4% of paediatric strabismus presentations. The condition affects approximately 1 in 1,000 indi-

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viduals, with a female predominance (~60%) and left-eye predilection for unilateral cases. Bilateral involvement, though recognised, is less frequently documented in case-based reports.

The most clinically significant diagnostic pitfall for Type I DRS is misidentification as acquired abducens (sixth cranial nerve) palsy, particularly in adults, leading to unnecessary neuroimaging, lumbar puncture, or systemic investigation. Recognition of the pathognomonic globe retraction and palpebral fissure narrowing on adduction — features absent in abducens palsy — is sufficient to establish the diagnosis clinically (3, 4).

We present six patients with Type I DRS evaluated at a tertiary eye care centre, documenting detailed clinical profiles, nine-gaze motility assessments with photographic evidence, and individualized management outcomes.

2 | METHODS

Study Design and Setting

This retrospective, observational case series was conducted at a tertiary eye care centre in South India. Six consecutive patients with clinically diagnosed Type I DRS presenting between January 2024 and December 2025 were identified from clinical records and included. Diagnosis was established by a consultant paediatric ophthalmologist and strabismologist on the basis of clinical criteria: abduction restriction greater than adduction restriction, globe retraction, and palpebral fissure narrowing on adduction.

Clinical Evaluation Protocol

All patients underwent the following:

- BCVA assessment using Snellen chart (Cardiff acuity cards for young children)
- Cycloplegic refraction (1% cyclopentolate, two drops at ten-minute intervals)
- Hirschberg corneal reflex testing
- PBCT in primary position at distance (6 m) and near (33 cm)
- Nine-gaze ocular motility assessment with grading of abduction and adduction limitation on a −1 to −4 scale (−1 = mild, −4 = complete)
- Documentation of globe retraction, palpebral fissure narrowing, and upshoot/downshoot on adduction
- Standardised nine-gaze photographic motility panel
- Dilated fundus examination (1% tropicamide)

Surgical Candidacy Criteria

Surgical intervention was considered for patients with: (a) primary position deviation ≥10 PD with or without diplopia, (b) functionally significant AHP (>15° face turn), or (c) cosmetically unacceptable esotropia or exotropia in primary gaze. Amblyopia was treated pre-operatively prior to any surgical decision.

Case Presentations

A total of six patients (five female, one male; age range 11–39 years) were included in this series. Demographic data and clinical characteristics of all patients are summarised in Table 1 , and detailed nine-gaze ocular motility findings are provided in Table 2 .

Case 1

A 39-year-old woman presented with a longstanding history of restricted horizontal eye movements noticed since childhood, without diplopia or head tilt (Table 1**, Case 1**). BCVA was 6/6 in both eyes with minimal refractive error. Cover test revealed orthotropia in primary position at both distance and near. Ocular motility demonstrated marked bilateral abduction limitation (−3 OD, −3 OS) with full adduction, globe retraction on adduction bilaterally, and palpebral fissure narrowing in both eyes on adduction; no upshoot or downshoot was elicited (Table 2 **, Case 1**). No abnormal head posture was present. In the absence of primary position deviation or functional impairment, conservative management with annual follow-up was recommended.

Case 2

A 14-year-old girl was referred from routine school screening for suspected left eye weakness (Table 1**, Case 2**). BCVA was 6/6 bilaterally on cycloplegic refraction, which revealed low bilateral hyperopia (+0.75 DS OU). Cover test confirmed orthotropia in primary position. Motility assessment revealed moderate bilateral abduction limitation (−2 OD, −2 OS) with preserved adduction, globe retraction, palpebral

fissure narrowing, and bilateral adduction upshoot (Table 2 , **Case 2**). No abnormal head posture was adopted. Optical correction was prescribed and the patient was placed on six-monthly follow-up. Amblyopia was not detected.

Case 3

A 35-year-old man presented with a cosmetic complaint of his left eye appearing ”sunken” on side-gaze, a feature he recalled from early childhood (Table 1**, Case 3**). BCVA was 6/6 OU, and no significant refractive error was identified. Cover test demonstrated orthotropia in primary position with no diplopia. Motility assessment revealed marked left eye abduction limitation (−3 OS), globe retraction, palpebral fissure narrowing on adduction, and an adduction upshoot; the right eye demonstrated unrestricted movement in all cardinal positions (Table 2 **, Case 3**). The nine-gaze photographic motility panel for this patient — illustrating the full triad of marked abduction limitation, globe retraction, palpebral fissure narrowing on adduction, and superior globe displacement (upshoot) — is presented in Figure 1 . Given orthotropia in primary gaze and absence of functional or postural impairment, conservative management was advised.

Case 4

A 24-year-old woman was referred by her optometrist for evaluation of restricted eye movements bilaterally (Table 1**, Case 4**). BCVA was 6/6 OU with myopic astigmatism (−1.50/−0.75 × 180° OD; −1.25/−0.50 × 175° OS). Cover test confirmed orthotropia in primary position. Motility demonstrated moderate bilateral abduction limitation (−2 OD, −2 OS) with an additional mild bilateral adduction limitation (−1 OU) — an atypical but recognised feature of Type I DRS attributed to mechanical lateral rectus co-contraction; globe retraction and palpebral fissure narrowing were present bilaterally, but no upshoot or downshoot was elicited (Table 2 **, Case 4**). Optical correction was prescribed and conservative management with follow-up was instituted given the absence of primary position deviation, AHP, or functional deficit.

Case 5

A 17-year-old girl was referred by a general ophthalmologist for evaluation of a right eye turn noticed by her family (Table 1**, Case 5**). BCVA was 6/6 OU with no significant refractive error on cycloplegic refraction. PBCT revealed a right exotropia of 12 PD in primary position at distance, without AHP. Motility showed moderate right eye abduction limitation (−2 OD), globe retraction, palpebral fissure narrowing on adduction, and an adduction upshoot; the left eye was entirely normal in all positions (Table 2 **, Case 5**). In view of the small angle of primary position exotropia (12 PD), preserved binocular single vision, and absence of AHP, conservative management with three-monthly reviews was agreed upon with the patient and family, with surgery deferred pending deviation stability.

Case 6

An 11-year-old girl was brought by her parents with concerns regarding her left eye turning inward since early childhood, associated with cosmetic embarrassment (Table 1**, Case 6**). BCVA was 6/6 OD and 6/9 OS, confirming amblyopia in the left eye. Cycloplegic refraction revealed hyperopia (+1.50 DS OD; +2.00 DS OS). PBCT demonstrated a left esotropia of 15 PD in primary position at distance without AHP. Motility assessment revealed marked left eye abduction limitation (−3 OS), mild adduction limitation (−1 OS), globe retraction, palpebral fissure narrowing, and an adduction upshoot; the right eye was unremarkable (Table 2 **, Case 6**). The pre-operative nine-gaze photographic motility panel for this patient — demonstrating primary position esotropia, marked left eye abduction limitation, prominent globe retraction with palpebral fissure narrowing, and adduction upshoot — is presented in Figure 2 . Following 12 weeks of pre-operative amblyopia therapy (part-time occlusion), which improved BCVA OS to 6/7.5, the patient underwent left medial rectus recession (5.5 mm) under general anaesthesia. Intraoperative forced duction testing confirmed a restricted medial rectus. Operative details, surgical parameters, and post-operative outcomes at the three-month review are detailed in **Table 3**. Orthotropia in primary position (residual <5 PD) was achieved postoperatively, with final BCVA improving to 6/6 OS. Globe retraction, palpebral fissure narrowing, and motility restriction persisted post-operatively, as expected, and the patient and family were counselled accordingly.

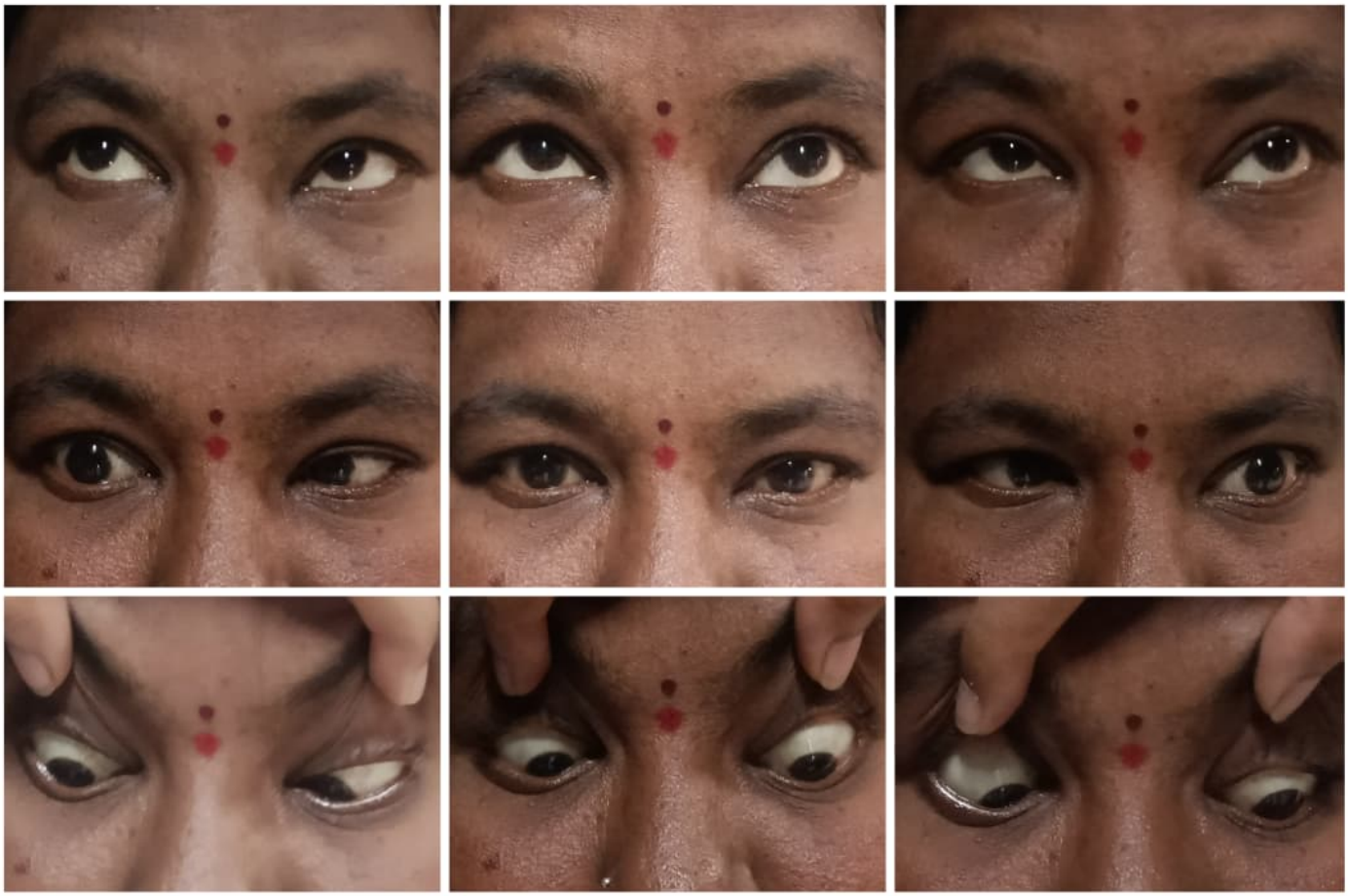

Fig. 1: Nine-gaze ocular motility photographic panel (Case 3: 35-year-old male, unilateral left Type I Duane's retraction syndrome). The composite panel demonstrates: (centre) full binocularalignment in primary position with orthotropia; (left column) marked limitation of left eye abduction with the right eye in primary gaze; (right column) globe retraction and palpebral fissure narrowing of the left eye on adduction; and (top-right) superior displacement (upshoot) of the left globe in the adduction–elevation position. Right eye motility is unrestricted in all positions.

3 | DISCUSSION

This case series documents six patients with Type I DRS, providing detailed nine-gaze motility profiles, photographic documentation, and individualized management outcomes from a South Indian tertiary eye care centre. Several clinically important observations merit discussion.

Demographic and epidemiological patterns. Female predominance (five of six patients, 83.3%) and the broad age range at presentation (11–39 years) are consistent with published epidemiological data. The overall female-to-male ratio in DRS approximates 3:2, though some referral-based series report proportions approaching the 5:1 excess observed in this cohort, likely reflecting ascertainment bias at specialist centres. Bilateral involvement in four of six patients (66.7%) is notably higher than the commonly cited bilateral rate of 15–20% in Type I DRS and warrants cautious interpretation given the

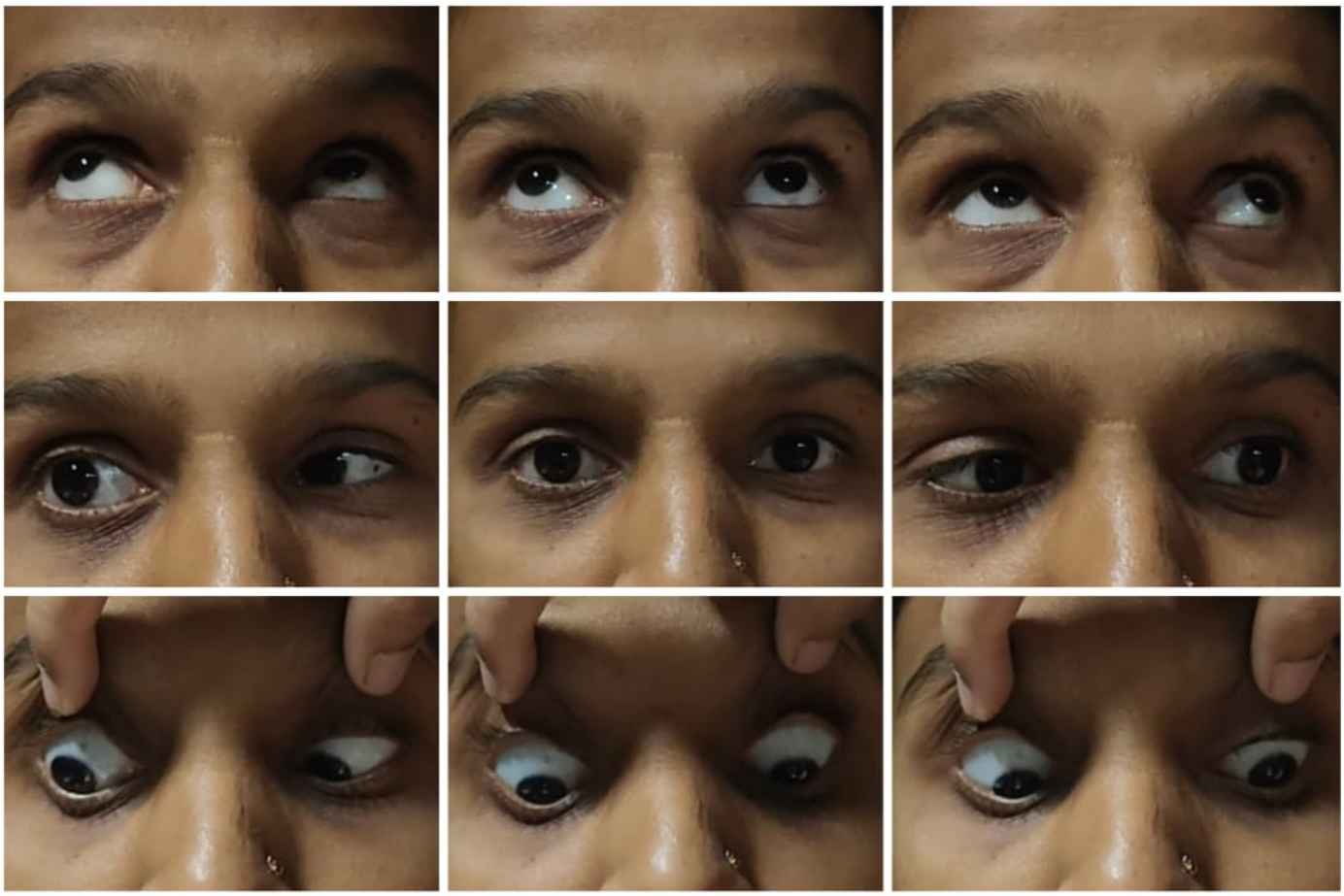

Fig. 2: Nine-gaze ocular motility photographic panel (Case 6: 11-year-old female, unilateral left Type I Duane's retraction syndrome, pre-operative). The panel demonstrates: (centre) left esotropia in primary position; (left column) marked limitation of left eye abduction; (right column) prominent globe retraction and palpebral fissure narrowing of the left eye on adduction; and (top-right) adduction upshoot of the left eye. Mild limitation of adduction is also present. Right eye motility is unrestricted.

Table 1. Demographic and Clinical Characteristics of Patients

Case	Age (years)	Sex	Laterality	Primary Position Deviation	Abnormal Head Posture	BCVA OD / OS	Refractive Error	Management
1	39	F	Bilateral	Orthotropia	Absent	6/6 / 6/6	Minimal	Conservative
2	14	F	Bilateral	Orthotropia	Absent	6/6 / 6/6	+0.75 DS OU	Conservative + optical Rx
3	35	M	Left	Orthotropia	Absent	6/6 / 6/6	Minimal	Conservative
4	24	F	Bilateral	Orthotropia	Absent	6/6 / 6/6	Myopic astigmatism OU	Conservative + optical Rx
5	17	F	Right	Exotropia 12 PD (RE)	Absent	6/6 / 6/6	None	Conservative, close follow-up
6	11	F	Left	Esotropia 15 PD (LE)	Absent	6/6 / 6/9*	Hyperopia OU	Surgical: left MR recession 5.5 mm

*Pre-operative BCVA OS 6/9 (amblyopia); improved to 6/6 OS at three-month post-operative follow-up following amblyopia occlusion therapy and surgery.BCVA = best-corrected visual acuity; OD = right eye; OS = left eye; PD = prism dioptres; RE = right eye; LE = left eye; MR = medial rectus; Rx = prescription.

Table 2. Ocular Motility Findings Across Nine Cardinal Positions

Case	Abduction Limitation	Grading (−1 to −4)*	Adduction Limitation	Globe Retraction	PF Narrowing on Adduction	Upshoot on Adduction	Downshoot on Adduction
1	Marked (bilateral)	−3 OD / −3 OS	Absent	Present (bilateral)	Present (bilateral)	Absent	Absent
2	Moderate (bilateral)	−2 OD / −2 OS	Absent	Present (bilateral)	Present (bilateral)	Present (bilateral)	Absent
3	Marked (left)	−3 OS	Absent	Present (left)	Present (left)	Present (left)	Absent
4	Moderate (bilateral)	−2 OD / −2 OS	Mild (bilateral) −1 OU	Present (bilateral)	Present (bilateral)	Absent	Absent
5	Moderate (right)	−2 OD	Absent	Present (right)	Present (right)	Present (right)	Absent
6	Marked (left)	−3 OS	Mild (left) −1 OS	Present (left)	Present (left)	Present (left)	Absent

*Limitation graded on a −1 to −4 scale: −1 = mild, −2 = moderate, −3 = marked, −4 = complete absence of movement.PF = palpebral fissure; OD = right eye; OS = left eye.

Table 3. Surgical Case — Operative Parameters and Post-operative Outcomes (Case 6)

Parameter	Detail
Patient age at surgery	11 years
Pre-operative deviation (primary position)	Left esotropia 15 PD
Pre-operative BCVA OS	6/9 (amblyopia)
Pre-operative amblyopia therapy	Part-time occlusion ×12 weeks
BCVA OS post-amblyopia therapy (pre-op)	6/7.5
Procedure performed	Left medial rectus recession, 5.5 mm
Anaesthesia	General anaesthesia
Intraoperative forced duction test	Restricted medial rectus confirmed
Post-operative deviation (3 months)	Orthotropia (residual <5 PD)
Post-operative BCVA OS (3 months)	6/6
Globe retraction post-operatively	Persistent (expected; counselled)
Adduction upshoot post-operatively	Persistent (no change)
Complications	None

PD = prism dioptres; BCVA = best-corrected visual acuity; OS = left eye.

small sample; however, it may reflect a genuine pattern in South Indian patients or referral clustering of bilateral cases at a tertiary centre (5, 6) .

Motility spectrum and upshoot. All six patients demonstrated the cardinal triad of Type I DRS — abduction limitation, globe retraction, and palpebral fissure narrowing on adduction — consistent with the neuroanatomical basis of abducens nerve hypoplasia with aberrant oculomotor co-innervation of the lateral rectus (Figures 1 and 2). Abduction limitation was graded as marked (−3) in three patients and moderate (−2) in three, suggesting a spectrum of abducens hypoplasia rather than uniform nerve agenesis. Mild adduction limitation (−1) was present in Cases 4 and 6, an atypical but recognised feature attributable to lateral rectus co-contraction mechanically resisting adduction or to partial vertical co-innervation anomalies (7).

Adduction upshoot was present in four of six patients (66.7%), exceeding the widely cited rate of approximately 43% in larger DRS series (Table 2). The upshoot in DRS is explained by two non-exclusive mechanisms: (a) the ”leash effect,” whereby a tight, fibrotic lateral rectus acts as a mechanical tether that deflects the globe superiorly during adduction; and (b) anomalous superior rectus or inferior oblique co-innervation via aberrant oculomotor nerve branches. Differentiating these mechanisms — using the co-contraction test and intraoperative forced duction — has direct surgical implications, as the Y-splitting procedure (addressing the leash) differs from standard recession when anomalous vertical innervation predominates (8, 9) .

Primary position deviation and AHP. Four

patients were orthotropic, one was exotropic (12 PD), and one was esotropic (15 PD) in primary gaze. Notably, no patient in this series adopted an abnormal head posture, even those with primary position deviation. AHP is reported in approximately 75–80% of esotropic Type I DRS patients; its absence in Case 5 and Case 6 may reflect small deviation magnitude (12 and 15 PD respectively), intact fusional reserves, or early cortical adaptation (8, 9).

Management and surgical outcomes. Five of six patients were managed conservatively, consistent with established evidence that the majority of Type I DRS — particularly orthotropic cases — require no surgical intervention. Surgery in DRS has clearly defined, limited goals: elimination of primary position deviation, correction of AHP, and improvement of cosmesis. It does not restore abduction, eliminate globe retraction, or normalise motility. In Case 6, medial rectus recession of 5.5 mm achieved orthotropia (residual <5 PD) with full amblyopia recovery (6/6 OS) — underscoring that pre-operative amblyopia management is essential before surgical planning and contributes substantially to visual outcome (10, 11).

Differential diagnosis. The most critical clinical mimic is acquired abducens palsy. Key differentiating features favouring Type I DRS include: congenital onset; non-progressive course; globe retraction on adduction (absent in abducens palsy); palpebral fissure narrowing on adduction; and orthotropia or minimal deviation in primary gaze with no diplopia. Awareness of these distinctions prevents unnecessary neuroimaging, lumbar puncture, or misdiagnosis (12, 13).

Limitations. This series is limited by its small sample size (n=6), single-centre retrospective design, absence of MRI orbit and brain to confirm abducens nerve agenesis, and lack of genetic testing (SALL4, CHN1, MAFB). Long-term follow-up beyond three months is unavailable for the surgical case. A prospective multi-centre registry approach would strengthen epidemiological and outcomes evidence for DRS in South Asian populations. (14, 15)

4 | CONCLUSION

Type I Duane's retraction syndrome presents with a consistent and clinically recognisable phenotype: abduction limitation, globe retraction, palpebral fissure narrowing on adduction, and — in the majority of patients — upshoot on adduction. The condition is non-progressive and most patients, especially those in orthotropia, can be managed conservatively with reassurance and optical correction. Surgical intervention should be reserved for primary position deviation, functionally significant AHP, or amblyopia, with realistic counselling that motility will not be restored. Early and accurate clinical recognition remains the cornerstone of appropriate management and avoids unnecessary systemic investigation.

Clinical Message

Type I DRS has a characteristic and reproducible clinical presentation that distinguishes it from acquired abducens palsy. Careful nine-gaze motility examination and photographic documentation enable confident diagnosis. Most patients require observation alone; timely and targeted surgical intervention in selected cases achieves excellent functional and cosmetic outcomes without attempting to restore ocular motility.

Declaration of Patient Consent

The authors confirm that written informed consent was obtained from all adult patients and from the parents or legal guardians of minor patients for participation in the study, photographic documentation, and publication of clinical information, with full assurance of anonymity.

Data Sharing Statement

De-identified case-level data supporting this report are available from the corresponding author on reasonable request.

5 | CONFLICTS OF INTEREST

None declared.

6 | FUNDING

None.

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