

Clinical Features of Duane Retraction Syndrome: A New Classification

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Purpose: We sought to provide a new classification system for Duane retraction syndrome (DRS) according to type and angle of strabismus during primary gaze and to analyze the clinical features of each DRS type.

Methods: The medical records of 65 DRS patients who visited the department of pediatric ophthalmology at Seoul National University Children's Hospital between 2010 and 2017 were retrospectively analyzed. Patients whose angle of exotropia at primary gaze exceeded 3 prism diopters (PDs) were classified as "Exo-Duane," those whose angle of strabismus at primary gaze did not exceed 3 PDs were classified as "Ortho-Duane," and those whose angle of esotropia at primary gaze exceeded 3 PDs were classified as "Eso-Duane."

Results: Among 65 DRS patients, Ortho-Duane was the most common (53.8%) type, followed by Eso-Duane (33.8%) and Exo-Duane (12.3%). The mean age at diagnosis was significantly higher in the Exo-Duane group than the Ortho-Duane or Eso-Duane group ($p = 0.003$ and $p < 0.001$, respectively). A predominance of left eye involvement was observed in the Ortho-Duane (62.9%) and Eso-Duane (90.9%) groups. The frequencies of upshoot, downshoot, fissure narrowing, and globe retraction were not significantly different among the subgroups. Head-turn was more frequent in Eso-Duane patients than in Exo-Duane or Ortho-Duane patients ($p = 0.001$ and $p < 0.001$, respectively). Myopia accounted for the most common refractive error among Exo-Duane patients (71.4%), while hyperopia was found more often in both Ortho-Duane (64.7%) and Eso-Duane (85.0%) patients. The majority of patients showed gross stereoacuity (93.1%), and a large proportion had good stereoacuity (Exo-Duane 60.0%, Ortho-Duane 81.3%, Eso-Duane 87.5%).

Conclusions: Our newly proposed classification of DRS according to type and angle of strabismus at primary gaze was practically useful and showed potential for use as an objective guideline in the clinical setting.

Key Words: Classification, Duane retraction syndrome

Duane retraction syndrome (DRS) is a congenital ocular-movement disorder accompanied by various horizontal

gaze limitations, palpebral fissure narrowing, globe retraction, and excessive upshoot and downshoot [1,2]. Among several classifications that represent the pathogenesis and clinical characteristics of DRS [2], Huber's classification is currently one of the most widely used [3,4] and proposes three types of DRS according to presenting electromyographic (EMG) pattern as follows: Huber type I DRS, characterized by definite abduction limitation and normal

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or mild adduction limitation; Huber type II DRS, characterized by definite adduction limitation and normal or mild abduction limitation; and Huber type III DRS, characterized by both abduction and adduction limitations [1,4,5].

However, clinicians face practical challenges in applying Huber's classification in actual cases since it is based on the EMG pattern presented; moreover, atypical EMG patterns and the diversity of possible clinical findings make it difficult for examiners to clearly distinguish subtypes. Thus, studies to develop a more practically useful classification system are needed. In the present research, we newly classified DRS into three types according to type and angle of strabismus at primary gaze and analyzed the clinical features of each type.

Materials and Methods

A retrospective analysis of the medical records of 65 DRS patients who visited the department of pediatric ophthalmology at Seoul National University Children's Hospital between 2010 and 2017 was carried out. Diagnosis of DRS was based on the finding of horizontal gaze deficit(s) together with one or more of the following characteristics: (1) palpebral fissure narrowing, (2) globe retraction, and (3) excessive upshoot and/or downshoot. Patients with history of prior ophthalmic surgery, ocular trauma, or orbital disease and those without a record of angle of strabismus at primary gaze were excluded from this study.

An analysis of epidemiological data (sex, age at diagnosis, laterality of the involved eye, strabismus surgery performed after diagnosis, and other systemic or central nervous system diseases) and clinical findings (angle of deviation at primary gaze, upshoot, downshoot, palpebral fissure-narrowing, globe retraction, abnormal head position, refractive status, and stereoacuity) was conducted. The angle of deviation was measured using the alternate prism cover test, and the refractive status was evaluated with cycloplegic refraction. Also, stereoacuity was assessed with the Worth four-dot test and Randot stereotest.

In this study, we propose a new classification system for DRS that sorts patients into three types according to the following criteria: patients whose angle of exotropia at primary gaze exceeded 3 prism diopters (PDs) were classified as "Exo-Duane," patients whose angle of strabismus at primary gaze did not exceed 3 PDs were classified as "Or-

tho-Duane," and patients whose angle of esotropia at primary gaze exceeded 3 PDs were classified as "Eso-Duane." The numerical criteria of 3 PDs is an arbitrary value. The standard was set within a narrow range since most patients with DRS present with a small angle of deviation at primary gaze less than 10 PDs, and abnormal head posture is concurrent even with a small angle of deviation; moreover, globe retraction, upshoot, or downshoot are often accompanied by only a small amount of ocular movement. Angle of strabismus was the measurement value at first medical examination.

Regarding stereoacuity, patients with DRS with stereoacuity of 100 arcseconds or less were considered to have good stereoacuity, while those with stereoacuity greater than 100 arcseconds were considered to have poor stereoacuity.

Strabismus surgery was indicated for cosmetic purposes in patients with large face turn, strabismus in the primary position, excessive upshoot or downshoot in adduction, or severe globe retraction.

Statistical analyses were performed using descriptive statistics, Fisher's exact test, the Kruskal-Wallis test, and/or the Mann-Whitney *U*-test where appropriate to compare results among the three DRS types. Also, analysis of covariance was used for age adjustment, and the Wilcoxon signed rank test was applied to compare preoperative and postoperative stereoacuity. All analyses were performed using the IBM SPSS Statistics ver. 23.0 (IBM Corp., Armonk, NY, USA), and comparisons were regarded as statistically significant for *p*-values less than 0.05. Bonferroni correction was applied for group-wise comparisons of each subtype. This study was approved by the institutional review board of Seoul National University Hospital (1907-184-1051) and the study protocol followed the tenets of the Declaration of Helsinki. Informed consent was waived due to the retrospective nature of the study.

Results

Epidemiology and comparison among the subgroups

A total of 65 patients was included, and the baseline demographics of these patients are shown in Table 1. Of 65 patients with DRS, eight (12.3%) were classified as Exo-Duane, 35 (53.8%) were classified as Ortho-Duane, and 22

Table 1. Epidemiology of Duane retraction syndrome patients

	Total	Exo-Duane	Ortho-Duane	Eso-Duane	<i>p</i> -value
No. of patients	65 (100.0)	8 (12.3)	35 (53.8)	22 (33.8)	-
Sex					
Male	37 (56.9)	4 (50.0)	17 (48.6)	16 (72.7)	0.234*
Female	28 (43.1)	4 (50.0)	18 (51.4)	6 (27.2)	
Age at diagnosis (yr)	4.4 ± 4.6 (0–19)	10.4 ± 5.6 (2–18)	4.3 ± 4.5 (0–19)	2.8 ± 2.4 (0–8)	0.003†
Involved eye					
Right eye	16 (24.6)	4 (50.0)	11 (31.4)	1 (4.5)	0.026*
Left eye	46 (70.8)	4 (50.0)	22 (62.9)	20 (90.9)	
Both eyes	3 (4.6)	0 (0.0)	2 (5.7)	1 (4.5)	
Surgery	29 (44.6)	7 (87.5)	9 (25.7)	13 (59.1)	0.001*
Systemic disease	7 (10.8)	1 (12.5)	4 (11.4)	2 (9.1)	1.000*
CNS disease	1 (1.5)	0 (0.0)	0 (0.0)	1 (4.5)	0.462*

Values are presented as number (%) or mean ± standard deviation (range).

CNS = central nervous system.

*Fisher's exact test; †Kruskal-Wallis test.

(33.8%) were classified as Eso-Duane. There were 37 (56.9%) male patients and 28 (43.1%) female patients, with no significant difference in sex proportion among subgroups ($p = 0.234$). Overall mean age at diagnosis was 4.4 ± 4.6 (range, 0–19) years, and mean age at diagnosis was 10.4 ± 5.6 (range, 2–18) years for Exo-Duane, 4.3 ± 4.5 (range, 0–19) years for Ortho-Duane, and 2.8 ± 2.4 (range, 0–8) years for Eso-Duane. Whereas no significant difference in age at diagnosis was noted between the Ortho-Duane and Eso-Duane populations ($p = 0.268$), a significantly higher value was obtained in the Exo-Duane group than in the Ortho-Duane or Eso-Duane group ($p = 0.003$ and $p < 0.001$, respectively). The left eye was affected in 46 (70.8%) patients, followed by the right eye in 16 (24.6%) patients and both eyes in three (4.6%) patients. Predominance of left-eye involvement was also observed in 22 (62.9%) and 20 (90.9%) patients of the Ortho-Duane and Eso-Duane groups, respectively.

Of the 65 patients included, 29 (44.6%) underwent strabismus surgery after diagnosis of DRS, comprising seven (87.5%) Exo-Duane patients, nine (25.7%) Ortho-Duane patients, and 13 (59.1%) Eso-Duane patients. In subgroup comparison for proportion of patients who underwent strabismus surgery, a statistically significant difference was observed ($p = 0.001$).

With regard to associated comorbidities, seven (10.8%) patients—one (12.5%) Exo-Duane patient, four (11.4%) Or-

tho-Duane patients, and two (9.1%) Eso-Duane patients ($p = 1.000$)—had systemic diseases such as Goldenhar syndrome, Noonan syndrome, and asthma. Among the entire cohort, central nervous system disease was found only in one (4.5%) patient in the Eso-Duane group.

The overall total mean angle of deviation was 7.7 ± 10.2 (range, 0.0–40.0) PD. Separately, the Exo-Duane group presented findings of 20.6 ± 7.5 (range, 10.0–32.5) PD, the Ortho-Duane group showed findings of 0.06 ± 0.3 (range, 0.0–2.0) PD, and the Eso-Duane demonstrated findings of 15.3 ± 8.7 (range, 4.0–40.0) PD for angle of deviation.

Clinical characteristics and comparison among the subgroups

1) Abnormal ocular movement, head position, and hyperopia at primary gaze

Several characteristics of DRS are shown in Table 2. Upshoot was observed in 23 (35.4%) patients, and downshoot was seen in 10 (15.4%) patients. Additionally, palpebral fissure narrowing was observed in 30 (46.2%) patients, and globe retraction on adduction was recorded in six (9.2%) patients.

Abnormal head position was detected in 30 (46.2%) patients; of these, 25 (38.5%) showed head turn, which was the most common feature; two (3.1%) showed head tilt; and three (4.6%) showed both head turn and head tilt. The fre-

quency of normal head position was significantly higher in the Ortho-Duane group than in the Eso-Duane group ($p < 0.001$) but was not statistically different between that of the Exo-Duane group and the Ortho-Duane group or the Eso-Duane group ($p = 0.028$ and $p = 0.345$, respectively). No significant difference was observed in head tilt among the

three subtypes ($p = 0.344$). Head turn was significantly more frequent in the Eso-Duane group than in the Exo-Duane or Ortho-Duane groups ($p = 0.001$ and $p < 0.001$, respectively); however, the frequency of head turn between the Exo-Duane and Ortho-Duane groups was not significantly different ($p = 1.000$) (Table 2).

Table 2. Abnormal ocular movement, head position, and hypertropia at primary gaze in Duane retraction syndrome patients

	Total (n = 65)	Exo-Duane (n = 8)	Ortho-Duane (n = 35)	Eso-Duane (n = 22)	p-value
Abnormal ocular movement					
Upshoot	23 (35.4)	4 (50.0)	13 (37.1)	6 (27.3)	0.535*
Downshoot	10 (15.4)	1 (12.5)	7 (20.0)	2 (9.1)	0.704*
Fissure narrowing	30 (46.2)	5 (62.5)	13 (37.1)	12 (54.5)	0.295*
Retraction	6 (9.2)	1 (12.5)	2 (5.7)	3 (13.6)	0.474*
Abnormal head position					
Normal head position	35 (53.8)	3 (37.5)	28 (80.0)	4 (18.2)	<0.001*
Head tilt	2 (3.1)	1 (12.5)	1 (2.9)	0 (0.0)	0.344*
Head turn	25 (38.5)	1 (25.0)	6 (17.1)	18 (81.8)	<0.001*
Head tilt and turn	3 (4.6)	3 (25.0)	0 (0.0)	0 (0.0)	0.001*
Hypertropia at primary gaze	6 (9.2)	4 (50.0)	1 (2.9)	1 (4.5)	0.001*

Values are presented as number (%).

*Fisher's exact test.

Table 3. Refractive error and status of Duane retraction syndrome patients

	Total (n = 61)	Exo-Duane (n = 7)	Ortho-Duane (n = 34)	Eso-Duane (n = 20)	p-value (adjusted p-value)	
Refractive error (SE)						
Involved eye	0.90 ± 1.5 (-2.5 to 5.5)	-0.48 ± 0.72 (-1.5 to 0.75)	0.68 ± 1.3 (-2.5 to 4.3)	1.8 ± 1.6 (-0.25 to 5.5)	0.001* (0.014 [†])	
Normal eye	0.73 ± 1.5 (-2.0 to 5.0)	-0.84 ± 0.94 (-1.9 to 0.25)	0.56 ± 1.3 (-2.0 to 4.0)	1.6 ± 1.6 (-0.38 to 5.0)	0.001* (0.009 [†])	
Average	0.81 ± 1.5 (-1.8 to 5.1)	-0.66 ± 0.73 (-1.3 to 0.50)	0.62 ± 1.2 (-1.8 to 4.0)	1.7 ± 1.6 (-0.13 to 5.1)	0.001* (0.008 [‡])	
Refractive status						
Involved eye	Emmetropia	3 (4.9)	0 (0.0)	3 (8.8)	0 (0.0)	1.000 [‡]
	Hyperopia	42 (68.9)	1 (14.3)	22 (64.7)	19 (95.0)	0.043 [‡]
	Myopia	16 (26.2)	6 (85.7)	9 (26.5)	1 (5.0)	0.011 [‡]
Normal eye	Emmetropia	4 (6.6)	0 (0.0)	3 (8.8)	1 (5.0)	1.000 [‡]
	Hyperopia	41 (67.2)	2 (28.6)	22 (64.7)	17 (85.0)	0.001 [‡]
	Myopia	16 (26.2)	5 (71.4)	9 (26.5)	2 (10.0)	0.001 [‡]
Average	Emmetropia	3 (4.9)	0 (0.0)	2 (5.9)	1 (5.0)	0.861 [‡]
	Hyperopia	41 (67.2)	2 (28.6)	22 (64.7)	17 (85.0)	0.043 [‡]
	Myopia	17 (27.9)	5 (71.4)	10 (29.4)	2 (10.0)	0.013 [‡]

Values are presented as mean ± standard deviation (range) or number (%).

SE = spherical equivalent.

[†]Kruskal-Wallis test; [‡]Analysis of covariance; ^{*}Fisher's exact test.

Hypertropia at primary gaze was detected in six (9.2%) patients, comprising four (50.0%) Exo-Duane patients, one (2.9%) Ortho-Duane patient, and one (4.5%) Eso-Duane patient ($p = 0.001$).

2) Refractive error

As of refractive error, the highest proportions of hyperopia were obtained in 42 (68.9%) patients and 41 (67.2%) patients for the subgroups of involved eyes and normal eyes, respectively; followed by myopia as the second most common refractive status in 16 (26.2%) and 16 (26.2%) patients; and emmetropia as the least common refractive status in four (6.6%) and three (4.9%) patients (Table 3). During subgroup comparison, proportion of hyperopia was significantly higher in the Eso-Duane group than in the Exo-Duane group ($p = 0.011$); meanwhile, no significant difference was found between the Ortho-Duane and Eso-Duane or Exo-Duane groups ($p = 0.108$ and $p = 0.105$, respectively). With regard to myopic refractive error, the proportion of myopia was higher in Exo-Duane group than in Eso-Duane group with statistical significance ($p = 0.005$), while that between Ortho-Duane and Exo-Duane or Eso-Duane group was without statistically significant difference ($p = 0.079$ and $p = 0.174$, respectively). When comparing between subgroups following age adjustment, the average refractive error showed no statistically significant difference among subgroups (Exo-Duane vs. Or-

tho-Duane, $p = 0.557$; Exo-Duane vs. Eso-Duane, $p = 0.022$; Ortho-Duane vs. Exo-Duane, $p = 0.028$; $\eta^2=0.056$).

3) Stereoacuity

In the entire cohort, the mean stereoacuity was 2.1 ± 0.5 (range, 1.3–3.8) log arcseconds, and the mean age at the time of examination was 7.3 ± 5.0 (range, 3–21) years. A better value of stereoacuity was obtained in the Ortho-Duane population (2.0 ± 0.3 [range, 1.3–2.6] log arcseconds) in comparison with either the Exo-Duane (2.4 ± 0.7 [range, 1.9–3.5] log arcseconds) or Eso-Duane (2.2 ± 0.7 [range, 1.7–3.8] log arcseconds) population; however, the difference was not statistically significant ($p = 0.380$) (Table 4). In 27 (93.1%) patients comprising the majority, gross stereoacuity was observed as follows: four (80.0%) Exo-Duane patients, 16 (100.0%) Ortho-Duane patients, and seven (87.5%) Eso-Duane patients ($p = 0.192$). In 23 (82.1%) patients, good stereoacuity was observed as follows: three (60.0%) Exo-Duane patients, 13 (81.3%) Ortho-Duane patients, and seven (87.5%) Eso-Duane patients ($p = 0.574$).

Data of postoperative stereoacuity were collected in seven patients and are shown in Table 5. The mean age at the time of examination was 5.9 ± 2.9 (range, 4–10) years. A mean stereoacuity of 2.1 ± 0.3 (range, 2.0–2.6) log arcseconds and positive findings of gross stereoacuity were obtained in all seven patients (100%), and good stereoacuity was observed in five (71.4%) of these seven patients. No

Table 4. Stereoacuity of Duane retraction syndrome patients

	Total (n = 29)	Exo-Duane (n = 5)	Ortho-Duane (n = 16)	Eso-Duane (n = 8)	p-value
Stereoacuity (log arcsec)	2.1 ± 0.5 (1.3–3.8)	2.4 ± 0.7 (1.9–3.5)	2.0 ± 0.3 (1.3–2.6)	2.2 ± 0.7 (1.7–3.8)	0.380*
Gross Stereoacuity, positive	27 (93.1)	4 (80.0)	16 (100.0)	7 (87.5)	0.192†
Good stereoacuity	23 (82.1)	3 (60.0)	13 (81.3)	7 (87.5)	0.574†
Poor stereoacuity	5 (17.9)	2 (40.0)	3 (18.8)	1 (12.5)	

Values are presented as mean \pm standard deviation (range) or number (%).
*Kruskal-Wallis test; †Fisher’s exact test.

Table 5. Postoperative stereoacuity of Duane retraction syndrome patients who underwent strabismus surgery

	Total (n = 7)	Exo-Duane (n = 1)	Ortho-Duane (n = 1)	Eso-Duane (n = 5)	p-value
Stereoacuity (log arcsec)	2.1 ± 0.3 (2.0–2.6)	2.0 ± 0.0 (2.0–2.0)	2.0 ± 0.0 (2.0–2.0)	2.1 ± 0.3 (2.0–2.6)	0.627*
Gross stereoacuity, positive	7 (100.0)	1 (100.0)	1 (100.0)	5 (100.0)	1.000†
Good stereoacuity	5 (71.4)	1 (100.0)	1 (100.0)	3 (60.0)	1.000†
Poor stereoacuity	2 (28.6)	0 (0.0)	0 (0.0)	2 (40.0)	

Values are presented as mean \pm standard deviation (range) or number (%).
*Kruskal-Wallis test; †Fisher’s exact test.

significant differences in mean stereoacuity, rate of positive gross stereoacuity, and good stereoacuity of the patients were observed among the three groups ($p = 0.627$, $p = 1.000$, and $p = 1.000$, respectively). When comparing stereoacuity before and after strabismus surgery in two patients whose preoperative and postoperative stereoacuity data were both available, the mean stereoacuity improved from 2.7 ± 1.0 (range, 2.0–3.5) log arcseconds before surgery to 2.0 ± 0.0 (range, 2.0–2.0) log arcseconds after surgery without statistical significance ($p = 0.317$).

Discussion

In our study, Ortho-Duane comprised the largest proportion of patients (53.8%) among the three subgroups, followed in order by Eso-Duane (33.8%) and Exo-Duane (12.3%), which is a different finding from previous reports. O'Malley et al. [6], Shauly et al. [7], and Lee and Chang [8] instead reported that the majority of patients had esotropia (or esodeviation), and decreasing proportions of patients had orthotropia and exotropia (or exodeviation) in order, whereas Isenberg and Urist [9] and Park et al. [10] reported that orthotropia was the most common group followed equally by exotropia and esotropia. The differences in results among the studies may be due to the arbitrary criteria used to define the angle of strabismus in our study as compared to the other studies. Regarding average refractive errors, no statistically significant differences among the subgroups after age adjustment were observed, with a partial eta squared (η^2) value of 0.056, which means that age had a small effect on refractive error in our patients.

Further, we also did not detect any sex preponderance, which is similar to the findings of previous studies on Korean patients with DRS by Lee and Chang [8] and Park et al. [10] but inconsistent with those reporting a female preponderance among non-Korean patients with DRS [5,11-13]. The disparity of results among research groups may be related to demographic differences among the patients involved in each study and also be attributable to the small number of patients included in our study.

We observed in our cohort that unilateral cases were more frequent than bilateral cases, and the left eye was more commonly involved than the right eye, findings similar to those of previous studies [5,11-13]. Bilateral DRS accounted for 4.6% of all patients, which was less than the

range of 10% to 24% of all DRS cases noted in previous reports [11,12,14]. This difference in results between our study and others may be due to the limited number of patients in our study, as mentioned above.

In our study, 46.2% of all patients showed abnormal head position, with 38.5% of patients showing head turn, which is less than the proportion of 50% of patients reported by Lee and Chang [8] and that of 54.6% reported by Kekunnaya et al. [13] but more than that of 16.7% reported by Park et al. [10]. With regard to abnormal head positions, head turn was more frequent than head tilt (38.5% vs. 3.1%). Since horizontal deviation rather than vertical deviation such as hypertropia at primary position is more common in patients with DRS, compensatory head posture which is mainly head turn rather than head tilt may occur more frequently. In each subgroup, the proportion of patients with normal head posture was higher in the Ortho-Duane group than in the Exo-Duane or Eso-Duane group, though the former comparison was without a statistically significant difference ($p = 0.028$ and $p < 0.001$, respectively). Conversely, the proportion of patients with head turn was higher in the Exo-Duane or Eso-Duane group than in the Ortho-Duane group, with no significant difference for the former comparison ($p = 1.000$ and $p < 0.001$, respectively). These differences in the proportion of patients in regard to head posture among the groups could also be explained by the compensatory mechanism used to achieve binocular vision in the same context.

The proportion of patients accompanying hypertropia at primary gaze was 50.0% in the Exo-Duane group, which was largest, followed by 4.5% of the Eso-Duane and 2.9% of the Ortho-Duane groups. Although patients with DRS are known to have no vertical movement deficits, this result indicates that hypertropia at primary gaze could present as a mild form of upshoot in Exo-Duane and Eso-Duane groups.

With regard to refractive error, hyperopia was the most common, found in 67.2% of patients. This result is in agreement with those of most preceding studies [11]. However, the results of analysis by subtype revealed that myopia was most common in Exo-Duane group, whereas hyperopia was most common in Ortho-Duane and Eso-Duane groups. Since only seven patients were included in the Exo-Duane subgroup, generalization of these results to all DRS patients has some limitations.

In our study, 93.1% of patients had gross stereoacuity

and 82.1% had good stereoacuity, which is consistent with the results of former studies that reported DRS patients have good stereoacuity since they try to maintain fusion with compensatory head posture [2]. However, the comparison of stereoacuity between subgroups showed no statistically significant differences. All seven patients with available postoperative stereoacuity data showed gross stereoacuity, and 71.4% of them had good stereoacuity after surgery; nevertheless, it was difficult to evaluate whether surgery was a significant influencing factor for improvement of stereoacuity since records of stereoacuity both before and after the surgery were available in only two patients and evaluation of stereoacuity was not possible due to the young age of some of the patients. In the two patients with available data, mean stereoacuity was improved without statistical significance after surgery ($p = 0.317$). Further studies with long-term follow-up periods and larger numbers of patients are required to better evaluate the effect of strabismus surgery on improvement of stereoacuity.

DRS is one of congenital cranial dysinnervation disorders, which are congenital, nonprogressive, sporadic, or familial developmental anomalies of the cranial nerves characterized by abnormal eye, eyelid, and/or facial movements [15]. The traditional Huber's classification proposes three subtypes of DRS according to EMG pattern and corresponding ocular movement deficits. However, Huber's classification has several limitations. First, EMG studies are not feasible in all cases in practice. Second, diversity of ocular presentations and atypical EMG patterns, which arise from a wide range of pathophysiology of DRS, prevent ease of classification of the subtypes clinically. Third, Huber's classification does not encompass various characteristics of DRS such as vertical deviations and anomalous head positions [11]. Lastly, in Huber's classification, definite, or normal or mild adduction and/or abduction limitation(s) are somewhat subjective judgments that could differ among examiners; as such, a novel classification that enables better communication between ophthalmologists is required. In this regard, our study has strengths in that it proposed a clinically more convenient and objective new classification method of DRS using the angle of strabismus at primary position rather than Huber's traditional classification and analyzed various features of DRS. Moreover, to the best of our knowledge, our study is one of only a few studies to analyze demographics and

characteristics of DRS in an Asian population except one patient, which allows for comparison of differences between Asian and Caucasian populations. Although there were several studies that classified DRS patients based on deviation at primary position [16,17], our classification system is differentiated from them in that we used the criterion of 3 PD at primary gaze in classifying subgroups. Our study has limitations due to the relatively small number of patients included and because the data were collected through a retrospective analysis of medical records. Further studies analyzing changes of characteristics over time in larger populations and using magnetic resonance imaging may provide more information in understanding the pathophysiology of DRS.

In conclusion, the newly proposed classification of DRS according to type and angle of strabismus at primary gaze was practically useful and showed potential for validity as an objective guideline in the clinical setting.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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