

Type 4 Duane syndrome



Joshua A. Schliesser, MD,^a Derek T. Sprunger, MD,^b and Eugene M. Helveston, MD^b

PURPOSE	To identify cases of synergistic divergence whose characteristics suggest that this entity is a form of Duane syndrome.
METHODS	The records of all patients with a Duane syndrome diagnosis, including standardized eye position photographs, from the E-Consultation program of Cybersight, Orbis International were analyzed.
RESULTS	A total of 350 Duane syndrome cases were identified. Of these, 19 (5%) had features consistent with synergistic divergence, or type 4 Duane syndrome. Of the 19, 16 (84%) were male, 15 (79%) had palpebral fissure narrowing, all had anomalous head posture, and 18 (95%) were exotropic. Only 9 (47%) patients were reported to have undergone surgery.
CONCLUSIONS	Synergistic divergence is a rare entity with features similar to those of Duane syndrome. We suggest that this entity be classified as type 4 Duane syndrome, because it has unique findings and an innervation pattern that differs from the other 3 recognized types. (J AAPOS 2016;20:301-304)

Duane syndrome encompasses a spectrum of strabismus entities that have as common features misinnervation and cocontraction of extraocular muscles in addition to enophthalmos with palpebral fissure narrowing.¹⁻³ Other findings include an abnormal compensatory head posture and vertical up- or downshoots. Historically, Duane syndrome has been classified into three types. Type 1 includes patients with poor eye abduction and esotropia. Type 2 includes patients with poor eye adduction and exotropia. Type 3 includes patients with both poor eye adduction and abduction. Duane syndrome has been considered a continuum of diseases with variability in misinnervation and resultant clinical features.¹ Initially described in the nineteenth century by multiple authors, Duane was the first to obtain a relatively large case series of patients and described abnormal motility, head position, globe retraction, and pseudoptosis.⁴ Using electrophysiology testing, Huber demonstrated paradoxical misinnervation of rectus muscles and further classified Duane syndrome into the three classic subtypes.⁵

Simultaneous abduction or synergistic divergence has previously been reported in the literature.^{1-3,6,7} These case reports demonstrate typical findings of a Duane syndrome variant as well as an extreme form of misinnervation.

Patients typically have exotropia in primary gaze with synergistic divergence of the affected eye despite expected adduction (Figure 1). The purpose of this study was to identify cases of synergistic divergence whose characteristics suggest a fourth subtype of Duane syndrome using a large, international telehealth database.

Subjects and Methods

Cybersight is a telehealth and education program of Orbis International with a database of worldwide cases of Duane syndrome presented from 2003 to 2012. The Cybersight website has an extensive template for entering and uploading patient information, greatly aiding the consultation process. Review of electronic or e-consultation cases provides an important method for collecting a large number of patients to retrospectively assess cases provided by expert consultants. In the present study, the records of Duane syndrome cases were reviewed retrospectively to obtain the following data: patient age and sex, ophthalmologic history, medical history, and patient/family description of eye problem. Examination results, including visual acuity, head position, refraction, ductions and versions, and laterality of abnormality were also noted. Lastly, an assigned mentor via the Cybersight consultation service analyzed patient photographs of eye alignment of the 9 diagnostic positions of gaze, including right and left head tilt. Multiple mentors volunteer as consultants and were involved in these electronic consultations. The Indiana University School of Medicine Ethics Committee approved this study. This study and all of the data collection performed conform to all local laws and were compliant with the principles of the Declaration of Helsinki.

Results

A total of 350 cases of Duane syndrome were identified. Of these, 179 patients (51%) had type 1 Duane syndrome;

Author affiliations: ^aDixie Ophthalmic Specialists at the Zion Eye Institute, St. George, Utah; ^bIndiana University School of Medicine, Indianapolis, Indiana

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Correspondence: Joshua A. Schliesser, MD, 1791 East 280 North, St. George, UT 84790 (email: josh.schliesser@gmail.com).

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FIG 1. External photograph of patient with type 4 Duane syndrome. A, Exaggerated abduction with attempted adduction. B, Exotropia in 1°. C, Relatively normal abduction.

Table 1. Type 4 Duane syndrome characteristics

Characteristic	No.
Alignment in primary position	
Exotropia	13
Exotropia and hypertropia	5
Orthotropia	1
Anomalous head posture	19
Palpebral fissure narrowing	15

81 (23%), type 2; and 71 (20%), type 3. Features of synergistic divergence were observed in 19 patients (5%) with Duane syndrome, 13 of whom were initially diagnosed with type 2 or Duane variant. On careful review of images, the telemedicine consultant identified synergistic divergence.

Of the 19 identified with synergistic divergence, 16 were males. Laterality was equally divided. This entity was observed early in life, with parents reporting an average age of onset of 0.5 years (range, 0-4). The average age of presentation for evaluation was 11.5 years (range, 1-26). Most patients were exotropic and showed narrowing of palpebral fissure on attempted adduction of the involved eye. An anomalous head posture was identified in all patients (Table 1). Infraduction deficit was observed in 6 patients. One patient had a supraduction deficit. Dramatic upshoots were only observed in 2 patients.

Neuroimaging had been performed on 4 of the 19 patients, and only 1 had an abnormality detected by the radiologist. The abnormality was described as “an asymmetry of the right half of the midbrain.” One patient with Goldenhar syndrome and 1 patient with cerebral palsy also had synergistic divergence. Nystagmus was observed in 1 patient. Anisometropia was observed in 3 patients (Table 2).

Eye muscle surgery was performed in 9 cases. On forced duction testing, patients were reported to have a tight lateral rectus muscle. Bilateral lateral rectus recession was performed in 3 patients; single lateral rectus recession, in 5 patients; and vertical muscle transposition with single lateral rectus recession in 1 patient (Table 3). Postoperative data was available for 7 patients, of whom 6 had good alignment in primary position, with decrease of head turn after a single surgery. The 1 patient who had a significant amount of residual exotropia and head turn had undergone ipsilateral lateral rectus recession. All patients had persisting synergistic divergence after surgery.

Table 2. Associated neurologic, systemic, or other ocular findings observed in patients with type 4 Duane syndrome

Finding	No.
Abnormal MRI—midbrain finding	1
Goldenhar syndrome	1
Cerebral palsy	1
Nystagmus	1
Anisometropia	3

Table 3. Surgical interventions performed on patients with type 4 Duane syndrome

Surgery	No.
Bilateral LR recession	3
Single LR recession	5
Single LR recession with vertical rectus transposition	1

LR, lateral rectus muscle.

Discussion

What we have identified as a fourth type of Duane syndrome has been called “simultaneous abduction,” “synergistic divergence,” the “splits,” and “perversion of the extraocular muscles.”^{1,3,6,7} It has previously been described in the literature, but only through case reports.

We characterize type 4 Duane syndrome as exotropia in primary gaze, head turn opposite the involved eye, essentially full abduction of the involved eye, and absent adduction of involved eye with simultaneous abduction in gaze opposite the involved eye and narrowing of the palpebral fissure. Figure 2 shows the potential innervation differences to the medial and lateral rectus muscles that can result in the pathology seen with Duane syndrome.³ Wilcox and colleagues³ performed electromyographical testing on a single patient with synergistic divergence and found evidence of anomalous innervation that was on the continuum of Duane syndrome. Similar findings have been reported in overcorrected patients who have undergone ipsilateral medial rectus recession for type 1 Duane syndrome.⁸ None of our patients had a history of previous medial rectus surgery.

In the case of simultaneous abduction the oculomotor nerve sends nerve fibers to the lateral rectus muscle; the signal causes the eye to abduct and cocontract when it should simply adduct. Several questions remain unanswered including: Does the oculomotor “dividing” weaken the medial rectus muscle? Does this contribute to the wide

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