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ARTICLE

Comparison of ocular torsion between congenital and acquired unilateral superior oblique palsy

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Abstract

Background/Objectives To compare ocular torsion between congenital and acquired unilateral superior oblique palsy (USOP).

Subjects/Methods Retrospective review was performed on medical records of 163 USOP patients. Qualitative fundus torsional states in both eyes were determined based on locations of optic disc and fovea. Disc–fovea angles (DFA) were collected in both eyes for quantitative analysis. DFAs and the proportions of qualitative fundus torsional states in paretic and non-paretic eyes were compared between USOP patients and normal controls, and between congenital and acquired USOP patients.

Results This study included 90 patients with congenital USOP, 73 patients with acquired USOP, and 66 normal controls. Most control subjects showed no torsion in both eyes (93.9%), whereas 61.1% of congenital and 46.5% of acquired USOP patients showed extorsion in either eye. More patients with congenital USOP showed fundus extorsion in the non-paretic eye (24.4% versus 12.3%) or both eyes (20.0% versus 6.8%), compared with patients with acquired USOP ($P = 0.007$). DFAs of paretic and non-paretic eyes were larger in USOP patients than in normal controls ($P < 0.001$, for both congenital USOP versus control and acquired USOP versus control). DFAs of non-paretic eyes were larger in congenital USOP patients than in acquired USOP patients (10.3° versus 8.5°, $P = 0.018$).

Conclusions Congenital USOP showed greater fundus extorsion in the non-paretic eye, compared with acquired USOP. Fundus photographs of both eyes are necessary to understand the ocular torsion in USOP patients and the variations in fundus torsion with varying USOP aetiology.

Introduction

Unilateral superior oblique palsy (USOP) induces torsional misalignment in both eyes [1, 2]. Sensory tests for ocular torsion, such as the double Maddox rod test, Lancaster test, and examination with an amblyoscope, sufficiently demonstrate extorsional sensory states in patients with USOP [1, 3, 4]. However, the paretic eye in USOP does not always show extorsion on fundus photography [1, 5–7]. Fundus extorsion may be found in the paretic or non-paretic fellow eye or in both eyes. Several studies have investigated the torsional state in USOP patients, but they included small sample sizes and described fundus torsional states in congenital [5–7] or acquired USOP patients without comparing them [3, 4]. Furthermore, a few studies have investigated the fundus torsional state in both eyes. To date, the variations in fundus torsional states of both eyes with varying USOP aetiology remain unclear.

This study was conducted to describe the torsional states of both eyes on fundus photographs in USOP. We also compared the torsional state of both eyes between congenital and acquired USOP patients.

Subjects and methods

We retrospectively reviewed medical records of 163 consecutive patients with USOP who were treated during the period from 2007 to 2015 in a tertiary referral medical

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centre. The study protocol was approved by the institutional review board at Asan Medical Center and conformed to the tenets of the Declaration of Helsinki.

USOP was diagnosed on the basis of clinical findings, which included hypertropia in the paretic side, over-elevation or under-depression in adduction of the paretic eye, anomalous head posture, or positive Bielschowsky head tilt test result. We excluded patients with any signs of bilateral SOP, such as the reversal of hypertropia on diagnostic fields or head tilt. We also excluded patients with masked bilateral SOP, skew deviation, congenital syndrome, craniosynostosis, and/or craniofacial anomaly.

We stratified patients on the basis of congenital or acquired USOP. Congenital USOP was diagnosed based on clinical information, such as photographic evidence of persistent anomalous head posture, facial asymmetry, large vertical fusional amplitude, absence of torsional diplopia, or magnetic resonance imaging evidence of SO muscle volume asymmetry. Congenital USOP was further stratified into childhood-onset USOP and decompensated USOP, based on the presence of diplopia. Acquired USOP was also diagnosed based on clinical information, such as acute onset vertical diplopia, cardiovascular risk factors, or the presence of torsional diplopia. Acquired USOP was further stratified based on aetiology, such as microvascular, traumatic, or intracranial aetiology. Traumatic USOP was diagnosed when a patient had acute onset USOP and definite evidence of head trauma prior to symptom onset. USOP with intracranial pathology was diagnosed when a patient had magnetic resonace imaging evidence of intracranial pathologies, such as tumours or haemorrhages, which interrupted the trochlear nerve pathway. Microvascular USOP was diagnosed in patients with cardiovascular risk factors, such as diabetes, hypertension, or hyperlipidaemia, after exclusion of congenital USOP, traumatic USOP, and USOP with intracranial pathology.

A TRC-50DX fundus camera (Topcon Medical System, Tokyo, Japan) was used to acquire fundus photographs. By using an internal fixation target, all subjects' eyes were aligned to the primary position during fundus photography. Unwanted head tilting of subjects was prevented by aligning the subject's lateral canthi to side marks on the fundus camera. When a subject showed severe ocular torsion, we repeated fundus photography to confirm consistency.

As mentioned in a previous study [8], the vertical position of fovea is typically located between the centre and inferior margin of the optic disc. When the vertical position of the fovea was located below the inferior margin of the optic disc, the eye was determined to have ocular extorsion. Similarly, when the vertical position of the fovea in an eye was located above the geometric centre of the optic disc, the eye was determined to have ocular intorsion.

A disc–foveal angle (DFA) was measured in accordance with a protocol used in previous studies [8, 9]. Imaging software (PetaVision image software, Asan Medical Center, Seoul, Korea) and a built-in protractor were used to measure the DFA [10]. DFA measurements were repeated four times and the average of the four measurements was used for analysis. DFA was expressed as a positive number when the vertical position of the fovea was located below the centre of the optic disc. DFA was expressed as a negative number when the vertical position of the fovea was located above the centre of the optic disc.

We included data regarding fundus torsion and DFA from fundus photographs of both eyes in 66 normal control subjects. Normal control subjects had no systemic disease affecting ocular alignment, no ocular disease, and no strabismus; their refractive error ranged from emmetropia to −5.0 diopters. These control subjects were used for comparison of DFA and ocular torsional state with USOP patients.

Statistical analysis was conducted with SPSS version 21.0 (IBM Corporation, Armonk, NY, USA). P values < 0.05 were considered statistically significant. We compared differences in the proportions of sex, paretic side, and fundus torsional state between congenital and acquired USOP by using Pearson's χ^2 test. In addition, we compared differences in age, amount of hyperdeviation in primary position, and DFAs of paretic and non-paretic eyes between congenital and acquired USOP by using independent t-tests.

Results

This study included 90 congenital and 73 acquired USOP patients. There were no statistical differences in sex and paretic-side laterality between the two groups ($P = 0.191$ and $P = 0.146$, respectively, Pearson's χ^2 test). Patients with congenital USOP were younger than patients with acquired USOP. Hyperdeviation in the primary position was larger in patients with congenital USOP than in patients with acquired USOP (Table 1).

Qualitative ocular torsional state is shown in Fig. 1. Only 6.2% of control subjects showed fundus extorsion in either eye. In contrast, 61.1% of patients with congenital USOP and 46.5% of patients with acquired USOP showed fundus extorsion in paretic or non-paretic eyes. Congenital USOP showed fundus extorsion relatively more frequently in the non-paretic side, compared with acquired USOP (24.4% versus 12.3%). The proportion of patients who showed bilateral eye extorsion was greater in congenital USOP than in acquired USOP (20.0% versus 6.8%); proportions were statistically different between the two groups ($P = 0.007$, Pearson's χ^2 test) (Fig. 1).

Table 1 Baseline characteristics of subjects

		n	Sex (female: male)	Paretic side (right:left)	Age (years, mean ± SD)	Hyperdeviation (prism diopters, mean ± SD)
Congenital USOP (90 patients)	Childhood onset	58	22:36	20:38	8.7 ± 4.7	11.22 ± 5.91
	Decompensated	32	14:18	13:19	44.1 ± 17.6	11.75 ± 7.26
	Congenital total	90	36:54	33:57	21.3 ± 20.3*	11.41 ± 6.35*
Acquired USOP (73 patients)	Microvasculopathy	48	14:34	22:26	62.8 ± 8.4	5.72 ± 3.92
	Trauma	15	3:12	10:5	53.1 ± 18.6	8.40 ± 7.41
	Intracranial	10	5:5	3:7	52.8 ± 17.9	5.80 ± 4.26
	Acquired total	73	22:51	35:38	59.4 ± 13.2*	6.29 ± 4.92*

SD standard deviation, *USOP* unilateral superior oblique palsy

*$P < 0.05$ on independent *t*-test between congenital and acquired USOP

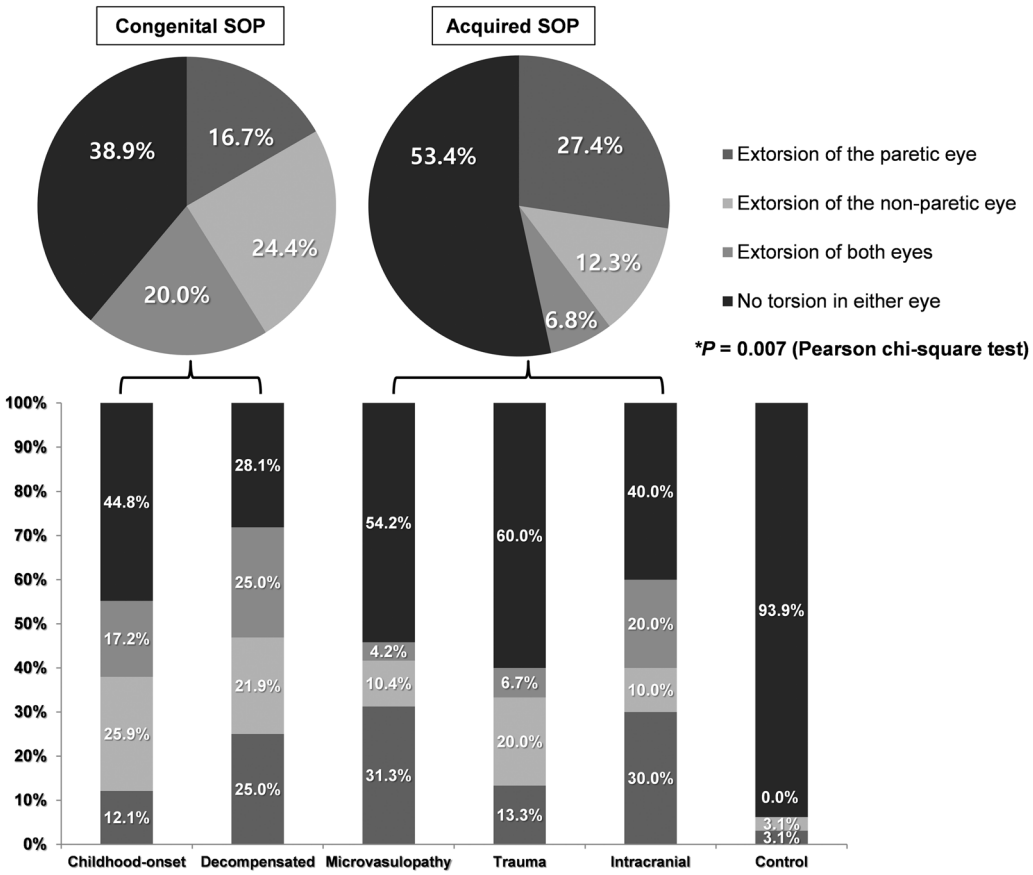

Fig. 1 Qualitative torsional state, based on aetiology. Congenital unilateral superior oblique palsy (USOP) patients showed relatively greater extorsion in non-paretic eyes and in both eyes, compared with acquired USOP patients. Acquired USOP patients showed greater extorsion in paretic eyes ($P = 0.007$, Pearson's χ^2 test, two-sided test)

DFA comparison based on aetiologies is shown in Fig. 2. The paretic, non-paretic, and sum DFAs of both eyes in USOP were larger than those of eyes in control subjects ($P < 0.001$, for both, control versus congenital USOP and control versus acquired USOP, independent *t*-tests). In a comparison between congenital and acquired USOP, DFAs in non-paretic eyes were statistically different ($P = 0.018$, independent *t*-test). DFAs in paretic eyes and sum DFAs of both eyes were not different between the two groups (Fig. 2).

As shown in Fig. 2, decompensated congenital USOP showed large DFAs in paretic and non-paretic eyes. When compared with other subgroups, decompensated congenital USOP showed larger DFAs in non-paretic eyes, compared with microvascular USOP ($P = 0.009$, independent *t*-test) and traumatic USOP ($P = 0.048$, independent *t*-test).

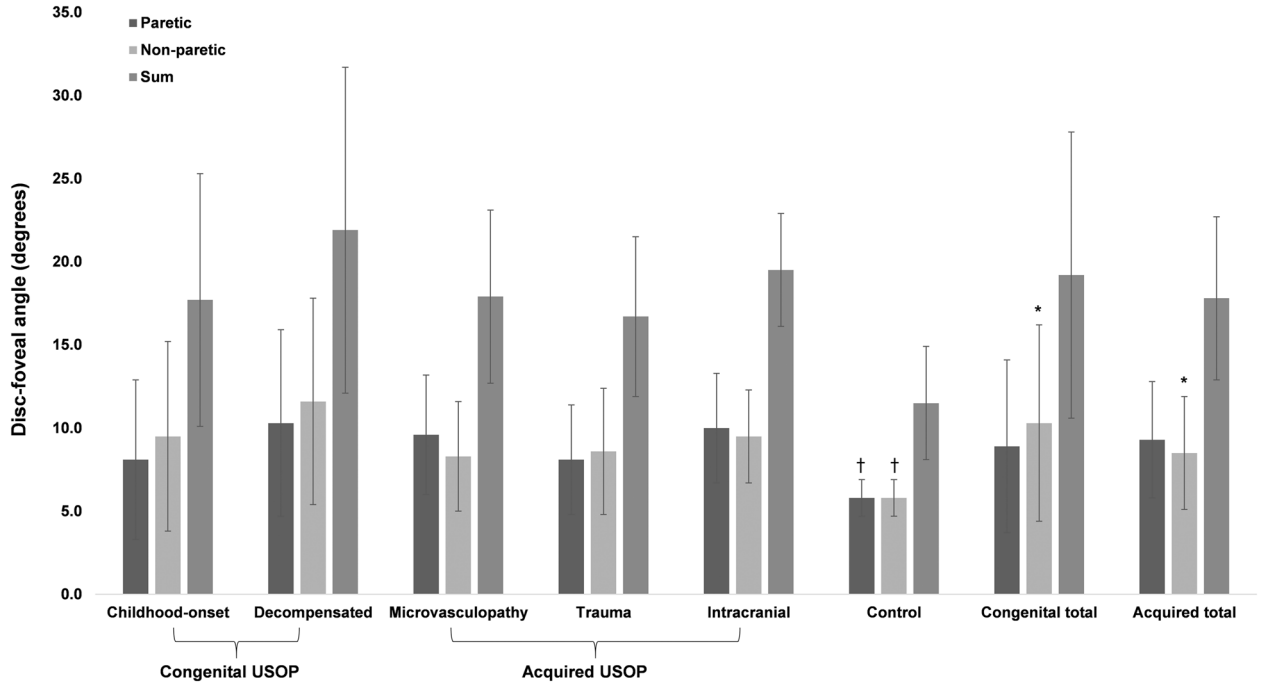

Fig. 2 Disc–-foveal angle (DFA, mean ± standard deviation) based on aetiology in unilateral superior oblique palsy (USOP) patients. Paretic, non-paretic, and sum (both eyes) DFAs in USOP patients showed greater extorsion than those in control subjects ($P < 0.05$, independent *t*-test). Non-paretic DFAs in congenital USOP patients showed greater extorsion than those in acquired USOP patients. *P-value = 0.018, independent *t*-test. Dagger indicates mean DFA of both eyes is indicated in control subjects

The sum DFA of both eyes in decompensated congenital USOP tended to be larger than that of eyes in childhood-onset ($P = 0.027$, independent *t*-test), microvascular ($P = 0.039$, independent *t*-test), and traumatic USOP ($P = 0.058$, independent *t*-test). DFAs of paretic eyes in decompensated USOP showed no statistical differences, compared with DFAs of paretic eyes in other subgroups.

Discussion

In this study, we have defined the fundus torsional state of both eyes, based on aetiology, in patients with USOP. Patients with USOP, regardless of their aetiologies, showed greater fundus extorsion in both paretic and non-paretic eyes, compared with normal controls. Congenital USOP revealed greater fundus extorsion in the non-paretic eye, compared with acquired USOP. Those findings were statistically valid, both qualitatively and quantitatively.

Lefevre et al. [5] described the fundus torsional state in 31 congenital superior palsy patients, using the same standard for fundus torsion that was used in our current study. They reported that 60.5% of the patients showed fundus extorsion in paretic (47.4%) or non-paretic (28.9%) eyes, which indicated that presumably 15.8% of the patients showed fundus extorsion in both eyes; the mean DFAs of paretic (10.7°) or non-paretic (8.8°) eyes were larger than those of normal controls (6.3°). These findings were comparable with our study. Although the proportion of congenital USOP patients with non-paretic eye extorsion (28.9%) in the study by Lefevre et al. was less than that in the present study (44.4%, including the cases of fundus extorsion in the non-paretic eye alone and in both eyes), it was still greater than the proportion of acquired USOP patients with non-paretic eye extorsion (19.1%, including 6.8% of acquired USOP patients with both eye extorsion) in this study.

No previous studies have directly addressed the fundus torsional state of both eyes in acquired USOP patients. Choi et al. [11] reported DFAs of paretic (12.67°) and non-paretic (9.59°) eyes in 22 acquired USOP patients. Lee et al. [4] reported that fundus torsional states were present in 61% of the paretic eyes of 31 acquired USOP patients. These findings suggested that the paretic eye tended to show fundus extorsion more frequently than the non-paretic eye in acquired USOP, consistent with the results of the present study.

The reason for differences in fundus torsional state in paretic and non-paretic eyes between congenital and acquired USOP remains unclear. Kushner and Hariharan [1] elegantly demonstrated a lack of immediate motor correction for torsional misalignment, even in ocular fixation change. They showed prolonged occlusion (~60 min) for the non-affected eye (i.e., long-term fixation with the

affected eye in USOP patients), induced fundus torsional change in the affected eye. Kim et al. [7] reported that ocular dominance might induce fundus extorsion in the non-dominant eye, regardless of the laterality of congenital USOP with SO atrophy. These findings suggested that long-term ocular dominance might affect fundus torsional state, regardless of the laterality of palsy. Fundus torsional state in congenital USOP might be affected by ocular dominance for a longer duration than in acquired USOP. Therefore, the probability of fundus extorsion in the non-paretic eye might be higher in congenital USOP than in acquired USOP.

Decompensated USOP showed relatively larger DFAs than other subgroups, as shown above. Decompensated congenital USOP exhibits a relatively long-term latent deviation, compared with other subgroups; it might be affected by ocular dominance for a longer duration than any other subgroups of USOP, including childhood-onset USOP. Guyton [12] revealed that long-term ocular misalignment could permanently change extraocular muscles. Repetitive sensorial and motor adaptation to torsional misalignment, and resultant permanent changes in extraocular muscles, especially inferior oblique muscle contracture, would aggravate fundus extorsion in decompensated USOP.

In this study, the DFA of normal control subjects was smaller than in paretic and non-paretic eyes of USOP patients, regardless of aetiology. This difference between normal control subjects and USOP patients suggests that USOP should be regarded as a binocular torsional disorder, rather than a monocular extorsional disorder. As demonstrated in a prior study [13], binocular torsional movements, such as cycloversion or cyclovergence, were present in this study, although ocular torsion was mediated by neural mechanisms to some degree [14]. A previous study [15] graphically demonstrated binocular torsional movements of cycloversion and cyclovergence in USOP patients using scleral search coils. USOP might initially induce paretic ocular extorsion, and afterward, ocular dominance [7], cyclofusion [15], or a neural adaptation mechanism [16] in the non-paretic eye, increasing the total amount of extorsion in both eyes; this may result in the "spread of torsional comitancy," similar to the "spread of comitancy" in long-term paralytic strabismus.

This study had a few limitations. It was conducted in a retrospective manner, such that several biases related to its retrospective nature might affect the findings in this study. A relatively large number of SOP patients were included in this study, but additional samples might be needed to confirm the findings suggested in this study. We did not collect ocular dominances from the study patients, so we could not conclude whether the variability of the fundus torsional state was solely affected by ocular dominance. A prospective study regarding the relationship between ocular dominance and fundus extorsion is needed to definitively evaluate this hypothesis.

In conclusion, congenital USOP showed greater fundus extorsion in the non-paretic eye, compared with acquired USOP. The fundus torsional state of both eyes in USOP might vary based on aetiology. It might be necessary to review fundus photographs of both eyes to understand the ocular torsional nature of USOP in each patient.

Summary

What was known before

- There were no reports on the etiological variations in fundus torsional states in both eyes with varying USOP aetiology.

What this study adds

- Congenital USOP showed greater fundus extorsion in the non-paretic eye compared to acquired USOP.
- Fundus photographs of both eyes are necessary to understand the ocular torsion in USOP patients.
- The fundus torsional state of both eyes in USOP varies with the aetiology.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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