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Comparison of Muscle Volume Between Congenital and Acquired Superior Oblique Palsies by Magnetic Resonance Imaging

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Abstract: Magnetic resonance imaging (MRI) studies of superior oblique (SO) muscles have revealed a high incidence of SO muscle atrophy/hypoplasia in congenital SO palsy patients. It has also been reported that long-standing acquired SO palsy patients show atrophic SO muscles in the affected eye. The purpose of this study was to compare the incidence of SO muscle atrophy/hypoplasia in congenital and acquired SO palsy by utilizing MRI. Coronal MRI image planes were taken from 29 cases of unilateral congenital SO palsy and 9 cases of acquired unilateral SO palsy patients. The SO muscle bellies were traced and their sizes were measured from each image plane. The total volume of the affected superior oblique muscle was compared with that of the normal fellow eye. The mean volume of the affected superior oblique muscle to that of the normal muscle was 45.3% (SD = 30.1) in the congenital group and 65.8% (SD = 22.7) in the acquired group. The volume reduction of the SO muscle in congenital SO palsy patients appears to be mainly a congenital abnormality rather than a secondary change, as seen in acquired SO palsy patients. **Jpn J Ophthalmol 1998;42:466–470** © 1998 Japanese Ophthalmological Society.

Key Words: Extraocular muscles, eye movement, MRI, superior oblique palsy.

Introduction

Superior oblique (SO) palsy is one of the most common causes of vertical deviation in primary gaze. Anatomical variations of the SO tendon have been widely recognized and a new classification of congenital SO palsy has been advocated.[1,2] Recently, magnetic resonance imaging (MRI) has been accepted as a useful technique to evaluate extraocular muscle size[3] and has revealed that SO abnormalities are not limited to the tendon but include the affected SO muscle belly, showing it to be atrophic or hypoplastic.[4,5] Similar atrophy of the SO muscle belly has been observed in long-standing acquired SO palsy.[6] It is still unclear if the muscle shrinkage observed in congenital SO palsy is a primary defect, a denervational atrophy, or both. In order to obtain better knowledge of the etiology of congenital SO palsy, we measured the SO muscle volume in both congenital and acquired SO palsy patients.

Materials and Methods

Magnetic resonance imaging was performed on 31 unilateral congenital SO palsy patients, 9 unilateral acquired SO palsy patients, and 5 volunteers without strabismus as control. Diagnosis of SO palsy was made by clinical examination and based on the presence of hypertropia, greatest in the nasal field of the involved eye, and a positive head tilt test. Patients who were seen by us as infants or whose parents had noticed abnormal head posture or strabismus without trauma from childhood were diagnosed as having a congenital palsy. Two patients who developed contralateral SO palsy after the initial surgery were diagnosed as masked bilateral SO palsy and were removed from the study. In adult patients, medical history of strabismus, old photographs, existence of large fusional vergence amplitudes or facial asymme-

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try helped differentiate congenital from acquired palsy.[7] In total, 29 unilateral congenital superior oblique palsy patients were studied, consisting of 18 men and 11 women. Age at the initial visit ranged from 6 months to 53 years, with MRI being performed on patients between 3 and 53 years of age.

The SO tendon was explored in 10 patients during surgery. Eight patients were found to have an anomalous insertion of the SO tendon: three in class IV (Cases 1, 12, and 28), 1 in class III (Case 10), 1 in class II (Case 13), 2 in class I (Cases 2 and 7) using the new classification by Helveston,[1] and 1 had a branched tendon (Case 3).

The acquired SO palsy group consisted of 7 men and 2 women, ages 16–68 years. All patients had symptoms for over 6 months and 8 of 9 patients had surgery. The causes of SO palsy included trauma, pons hemorrhage, and unknown etiology. There is a possibility of bilateral palsy in traumatic SO palsy patients; however, none of our patients showed evidence of bilateral involvement either before or after strabismus surgeries.

In each subject, 3-mm thick gapless coronal image planes were obtained spanning the orbit from the apex to the equator of the globe by using a surface coil over the scanned orbit and head. We employed three MRI systems; Sigma 1.5 Tesla (General Electric, Milwaukee, WI, USA), Visart 1.5 Tesla (Toshiba, Tokyo), and MRT200 1.5 Tesla (Toshiba, Tokyo). In all patients, spin echo (SE) T1-weighted images in the coronal planes were taken with a 256×256, 256×192, or 224×256 matrix over a 15 or 16 cm^2 field of view. We understand it is very important to control eye and head positions during MRI, but most of our study patients were too young to keep their eyes open and fixate on one position long enough. In order to keep them stable, young patients were sedated by triclofos sodium (Tricloryl, Glaxo, UK) during MRI scanning. Adult patients, older children, and control subjects were awake and instructed to close their eyes while MRI was performed. In each patient, five to seven image planes showing the SO muscle belly were obtained.

Image planes showing the SO muscle were scanned by an image scanner with 300 dpi resolution, and transferred to a personal computer (Macintosh Quadra 840 or Power Macintosh 7900/120, Apple Computer Inc., Cupertino, CA, USA). From each slice, the SO muscle, the medial rectus muscle (MR), the lateral rectus muscle (LR), and the inferior rectus muscle (IR) were traced and the number of pixels were counted (Adobe Photoshop ver. 3.0J, Adobe Systems, San Jose, CA, USA). The total number of pixels covered by each muscle were considered to represent muscle volume. In order to minimize the variations caused by the image quality or intersubject differences of muscle volume, the volume of the affected muscle was compared to that of the normal side as a percentage rather than using actual muscle volume. Data analysis was performed with the computer software StatView - J4.02 (Abacus Concepts, Berkeley, CA, USA).

Results

Patients' characteristics and the measurements of muscle volume are summarized in Tables 1 to 3.

Muscle Atrophy

The mean ratio of the muscle volume of the SO, MR, LR, and IR of the left eye to the right eye in control subjects was approximately 1.0. The mean ratio of muscle volume of the MR, LR, and IR of the paretic eye to the normal eye was approximately 1.0 for both congenital and acquired SO palsy patients. Using a 2 standard deviation of the SO in control subjects as a criterion, we classified the muscle atrophy as follows: no atrophy was more than 75%; mild was 50–75%; moderate was 25–50%; and severe was less than 25%. In the congenital SO palsy group, the mean percent volume of the affected SO muscle was 45.3% (SD = 30.1) of the normal SO muscle. Five patients (17.2%) were classified as having severe atrophy, 12 patients (41.3%) were moderate, 3 patients (10.3%) were mild, and 7 patients (24.1%) were classified as having no atrophy (Figure 1).

In the acquired cases, the mean percent volume of the SO muscle was 65.8% (SD = 22.7). No patients showed severe atrophy. Three patients (33.3%), a 60-year-old woman who was in an automobile accident 2 years previously (Case 6), a 43-year-old man with pons hemorrhage 1.5 years earlier (Case 7), and a 27-year-old man who had a sports accident 12 years earlier (Case 3) were classified as moderate. Two patients (22.2%) showed mild atrophy and the remaining 4 patients (44.4%) were classified as having no atrophy (Figure 1). There was a significant difference in the percent volume of the SO muscle between congenital and acquired SO palsy (Mann-Whitney *U*-test, $P = 0.031$).

Age and Duration of Palsy

There were no statistically significant differences between the age at MRI and muscle atrophy in the

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Table 1. Patients' Characteristics and Results of Measurements in Congenital Superior Oblique (SO) Palsy

Patient No.	Side	Age at MRI (years)	Performed Surgery		% Volume of Muscle Size of Paretic Side Compared to That of Normal Fellow Eye				Surgical Findings
			First	Second	SO	MR	LR	IR	
1	L	4	LIO	LSR, LMR	31.4	106	110	102	Absent
2	R	2	RIO	RSO tuck	97.5	92.9	76.8	96.5	Loose
3	L	9	LIO	LSO tuck, LSR	35.4	106	82.4	75.6	Branched
4	L	3	LIO		28.9	113	113	97.2	
5	R	7	RIO,R-R		35.6	96.9	89.7	87.7	
6	R	10	RIO	RSR	41.7	93.2	109	141	Normal
7	L	3	LIO,LSO tuck		12.8	78.9	108	132	Loose
8	L	9	LIO		8.1	88.0	127	123	
9	L	7	LIO		34.4	103	104	87.5	
10	L	3	LIO	LSO tuck	12.9	86.2	107	93.1	In Tenon
11	R	4	RIO,RLR		76.7	113	98.6	90.2	
12	L	6	LIO,LSR		0	89.4	111	99.1	Absent
13	R	32	RIO,RSR		55.3	111	103	89.5	Nasal insertion
14	R	28	RIO		42.4	104	96.1	110	
15	L	50	LIO	LSO tuck	21.1	125	114	86.4	
16	L	10	LIO		49.0	120	123	98.4	
17	L	9	LIO		94.1	87.1	86.5	125	
18	R	7	RIO		76.7	105	82.9	110	
19	R	9	RIO		26.4	88.8	79.8	102	
20	R	2	RIO		73.8	107	97.0	80.0	
21	L	2	LIO		59.6	94.4	133	145	
22	L	7	LIO		92.2	119	93.7	118	
23	R	8	RIO	RSR	36.5	119	99.0	82.8	
24	L	53	R-R		28.7	80.3	83.5	86.6	
25	L	3	RIO		47.7	109	92.9	90.3	
26	R	3	RIO		92.1	105	112	96.1	
27	L	3	LSR		13.3	103	105	124	Normal
28	R	1	RIO,RSR,BLR		0	89.3	92.5	95.6	Absent
29	R	10	R-R	RIO	90.3	111	108	146	
Mean					45.3	101.5	101.3	103.8	
SD					30.1	12.5	14.1	19.8	

BLR: bilateral lateral rectus muscle recession, IR: inferior rectus muscle, L: left, LIO: left inferior oblique muscle weakening, LMR: left medial rectus muscle recession, LR: lateral rectus muscle recession, LSO: left superior oblique muscle, LSR: left superior rectus muscle recession, MR: medial rectus muscle recession, MRI: magnetic resonance imaging, R-R: recess-resect of horizontal muscles, RIO: right inferior oblique muscle, RLR: right rectus muscle recession, RSO: right superior oblique muscle. RSR: right superior rectus muscle recession.

congenital and the acquired groups. In the acquired group, there was no relationship between the duration of palsy and the severity of muscle atrophy.

Discussion

The patients examined in this study were younger than in the previously published MRI reports on SO.[4–6,8] The younger age reduces the accuracy of the measurements of the muscle volume, but still gives us evidence that the muscle abnormality in congenital SO palsy is as common a finding as the tendon anomaly. In addition, most of the volume reduction is an inborn abnormality rather than a secondary change.

Demer and Miller[4] reported that the normal SO muscle belly changes its volume significantly when it contracts, but the paretic SO muscle does not. In our study, most of the congenital patients were sedated because they were too young to be stable during MRI. In addition, controls and all the acquired group patients were awake but simply closed their eyes during MRI. The ocular position with the lid closed is more likely to be in up gaze rather than in down gaze because of Bell's phenomenon. When patients fixate in the down-gaze position, the percent volume of the affected SO muscle to the normal SO is expected to become smaller than that in the primary or in the up-gaze position. From these consid-

Table 2. Patients' Characteristics and Results of Measurements in Acquired Superior Oblique (SO) Palsy

Patient No.	Side	Duration of Palsy	Age at MRI (years)	Performed Surgery	% Volume of Muscle Size of Paretic Side Compared to Normal Side			
					SO	MR	LR	IR
1	R	12 y	24	RIO, RSR	92.9	110.0	98.6	84.7
2	R	9 y	24	RIO,RMR	94.7	93.0	102.0	94.8
3	R	12 y	27	RIO	49.5	115.0	107.0	87.0
4	L	6 m	62	LSR, RIR	64.6	108.0	106.0	103.0
5	R	11 y	24	RIO	78.9	90.5	79.9	118.0
6	L	1 y	60	LIO	39.7	91.1	103.0	103.0
7	R	1 y	43	IO, H-I	29.9	119.0	105.0	109.0
8	R	1 y	16	Spontaneous recovery	77.2	81.0	140.0	116.0
9	L	8 m	10	Spontaneous recovery	64.7	93.0	136.0	67.0
Mean					65.8	100.0	109.0	98.2
SD					22.7	13.0	18.7	16.4

H-I: Harada-Ito, IO: inferior oblique, LIO: left inferior oblique muscle weakening, LSR: left superior rectus muscle recession, m: month, RIO: right inferior oblique muscle weakening, RIR: right inferior rectus muscle recession, RMR: right medial rectus muscle recession, RSR: right superior rectus muscle recession, y: year.

erations, our data might be underestimating the volume reduction, not overestimating it, in both congenital and acquired SO palsy.

Unlike the report by Ozkan et al,[8] in this study reduction of the affected muscle volume was more commonly observed and more severe in the congenital group than in the acquired group. There are several explanations for this difference.

Even without evidence, bilateral palsy should always be taken into consideration. Because we compared the muscle volume between the palsied and normal side, rather than comparing the actual muscle volume, if muscle atrophy is present in both eyes, the asymmetry may be obscured. As Nishida et al[9] reported, individual muscle volume measurements on MRI vary widely, depending on the age and body weight. Because our study subjects consisted of young children and adults, a comparison of actual muscle volume was not expected to reflect true muscle atrophy. On the contrary, in the acquired cases, the patients differed in age at injury, in cause and duration, and in the severity of palsy. It is possible that these factors influenced the incidence of muscle atrophy.

We performed MRI not on all the congenital SO palsy patients on whom we operated, but often after the failure of the initial surgery. Anomalous tendon was observed more often than reported by Helveston[1] and Lo[2]. Our congenital group may be biased by being more severe cases of SO palsy, which may increase the incidence of the atrophy of SO muscle.

Table 3. Percent Volume (Left/Right) of Extraocular Muscles in Normal Subjects

Volunteer No.	Sex	Age at MRI (years)	% Volume of Muscle Size of Left Eye to Right Eye			
			SO	MR	LR	IR
1	Male	28	113.0	100.0	93.5	120.0
2	Female	38	92.9	102.0	104.0	103.0
3	Male	27	94.9	91.9	112.0	105.0
4	Male	29	89.1	94.3	114.0	117.0
5	Male	28	97.1	111.0	92.7	110.0
Mean			97.4	99.8	103.0	11.0
SD			9.21	7.42	9.94	7.72

SO: superior oblique muscle, MR: medial rectus muscle, LR: lateral rectus muscle, IR: inferior rectus muscle.

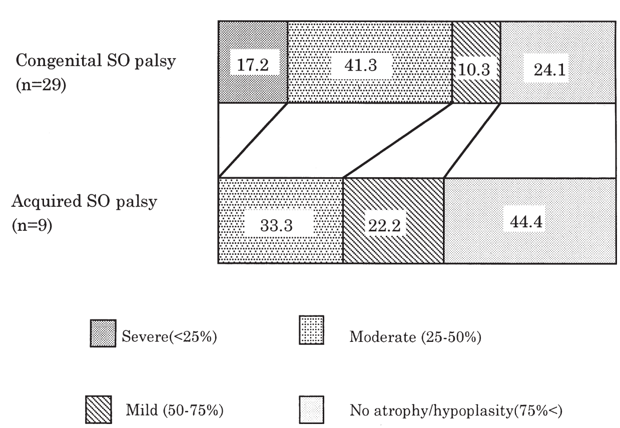

Figure 1. Distribution of percent volume of palsied superior oblique (SO) muscle to normal SO muscle. There was significant difference between congenital and acquired SO palsy patients in percent volume of affected SO muscle to SO of normal fellow eye. (Mann-Whitney U-test, $P = 0.031$).

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It is important to note that not all the congenital SO palsy patients showed hypoplastic SO muscle belly. This may be related to the limitations of the imaging technique in this study. Demer et al[5] report, one half to one third of patients clinically diagnosed with congenital or acquired SO palsy have normal contraction of the SO muscle and are considered to be masquerading as SO palsy. In this study, one fourth of congenital SO palsy patients did not show morphological changes in the SO muscle. This finding suggests that the etiology of congenital SO palsy is not simple and some causes are neurological paresis or, as Demer suggests, are masquerading as SO palsy[5]. Magnetic resonance imaging may enable us to make a valid diagnosis of SO palsy so that the clinical diagnosis of SO palsy can be confirmed.

In conclusion, atrophy/hypoplasity of the SO muscle was observed more often and was more severe in congenital than in acquired SO palsy cases. Morphological changes in the SO muscle belly in congenital cases are more likely an inborn abnormality rather than denervational atrophy. Magnetic resonance imaging, when combined with other clinical findings, gives us useful information to differentiate congenital from acquired SO palsy.

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