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AN ELECTROMYOGRAPHIC STUDY OF *TORTICOLLIS MUSCULARIS CONGENITA*

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The unilateral affection of musculus sternocleidomastoideus in patients with torticollis muscularis congenita often combines with a cranial asymmetry concave to the side of the affection and with an elevation of the shoulder on the affected side. Whether this is secondary to the affection of the sternocleidomastoid muscle or is due to an involvement of the facial and the trapezoid muscle on the same side is still a matter of dispute. The sternocleidomastoid and the trapezoid muscle originate from the same branchial archs, and this may indicate a common affection of the muscles from a primary involvement of the branchial arch (Lidge et al. 1957).

However, this does not explain the affection of the facial muscles, unless the development defect is supposed to be neurogenic (Schubert 1926) with a congenital defect in the nuclei of the accessorius and the facial nerves which lie near each other.

In 19 infants with congenital torticollis Baxter et al. 1961 compared the electromyographic findings during full effort with histological findings. The motor unit activity was normal or slightly diminished in 5 patients, moderately diminished in 10 and severely diminished in 4. The degree of destruction of muscle fibers was related to the loss of fibers as indicated by electromyography. Tönnis (1964) studied the duration amplitude and shape of motor unit potentials. In 2 of the 10 patients he examined the trapezius muscle in addition to the sternocleidomastoid. In 7 patients with advanced torticollis the duration and amplitude of motor unit potentials were reduced and the incidence of polyphasic potentials was increased. The two trapezoid muscles showed no electromyographic involvement, and he considered congenital torticollis to be a primary developmental defect of the sternocleidomastoid muscle.

The aim of the study presented in this report is to contribute to an

elucidation of the pathogenesis of congenital torticollis by an electromyographic study of the sternocleidomastoid, trapezius, and facial muscles.

MATERIAL AND METHODS

The study comprised 31 patients, 16 males and 15 females, 8–59 years old, 18 less than 20 and 13 more than 20 years old. Congenital torticollis, facial asymmetry or other congenital diseases occurred in the near family of 8 patients. In 24 patients there was a slight facial asymmetry and in 10 a slight asymmetry of the shoulder girdle. One patient had a congenital eye disease.

In 29 patients the sternocleidomastoid muscles were examined on the affected and the contralateral side and in 27 patients, in addition, the trapezoid muscles. The facial muscles (m. frontalis, m. orbicularis oris and m. triangularis) were investigated in 19 patients. In 16 patients electromyography was performed 4–18 years after a tenotomy.

Recording. The leading-off area of the platinum core of the concentric electrodes was 0.07 mm², and the external diameter of the cannula 0.65 (DISA 13k03) and 0.45 mm (13k50). Their impedance was $50\text{k}\Omega < -45^\circ$ at 150 Hz and $7\text{k}\Omega < -40^\circ$ at 5000 Hz measured in saline with a voltage of less than 10 mV. A 3 channel electromyograph (DISA 3A) was used for recording, two channels connected to two electrodes inserted in the muscle of the affected side, the third channel connected to an electrode inserted into the contralateral muscle. The patients were examined horizontally and in a resting position. Recordings from different motor units were obtained by inserting the electrodes in three different points of the muscles and at each point in three different depths. In the facial muscles recordings were made from only two points in each muscle. The pattern of activity and its amplitude during full effort was studied when the patient was asked to activate the right and left muscle simultaneously and then by asking the patient to turn the head against resistance to the right and to the left. The average duration of a representative number of individual motor unit potentials and their average amplitude was determined during slight voluntary effort as was the incidence of polyphasic potentials. These parameters were evaluated according to the criteria described by Buchthal (1957).

RESULTS

Sternocleidomastoideus muscle. In 8 of the 13 patients in whom a tenotomy had not been performed, full effort resulted in an interference

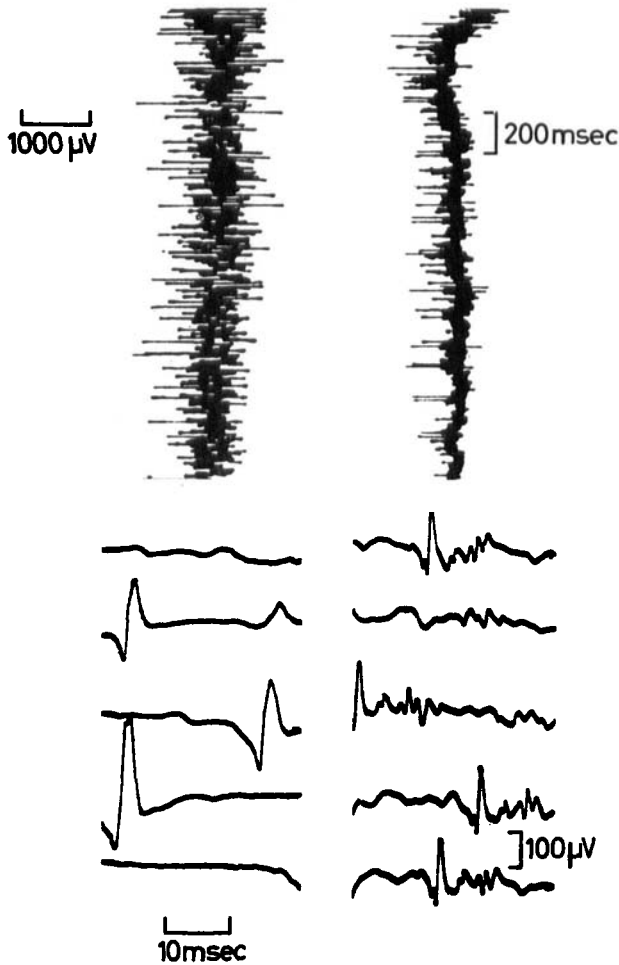


Figure 1. Electromyogram in a patient with congenital torticollis. Above: Pattern of activity during full effort. Below: Randomly sampled motor unit potentials. Right: Affected, left contralateral sternocleidomastoid muscle.

pattern, in 7 with a reduced amplitude as compared to the contralateral side. In 4 the reduction in amplitude was 30–80 per cent; they had an advanced wasting of the muscle. Five patients showed a mixed pattern, indicating loss of some motor units (Figure 1). Fibrillation potentials or other types of spontaneous activity did not occur, except when the patients kept the head straight, when rhythmic bursts of motor unit activity occurred at a frequency of about 12 per sec.

Table 1. Electromyographic findings in the sternocleidomastoid muscles in patients with congenial torticollis.

Pt	Age years	Per cent difference*) in mean duration	Per cent difference*) in mean amplitude	Per cent incidence of polyphasic potentials**	
				affected side	not affected side
not tenotomized					
PH	8	— 6	—10	14	14
PDA	9	—35	—22	20	17
FN	10	+ 2	—24	25	12
WN	10	—30	—61	17	13
JH	10	—45	—28	16	6
FH	10	— 2	— 3	25	0
VJ	11	0	—44	22	17
IBK	12	— 6	—32	25	14
BB	18	— 7	+ 7	14	7
JP	25	—19	—15	32	16
PM	32	+ 2	—10	17	5
DP	36	—42	—42	12	0
EN	42	+13	—15	19	7
after tenotomy					
GR	11	—11	—12	11	11
HC	11	— 7	—49	0	0
KOL	15	—20	+ 9	23	23
LL	17	+ 9	—32	0	0
RV	18	0	+ 2	5	0
KB	18	— 4	—13	8	8
IR	18	— 2	—29	10	10
BD	19	—28	—30	10	0
GL	22	+ 5	— 7	18	0
KD	23	—32	—33	6	6
HP	25	—24	+ 9	23	23
GJ	26	—22	—29	0	0
IB	26	—27	—38	8	0
IP	33	—15	—19	13	0
GH	33	— 2	+15	10	5
COV	59	+10	+13	8	7

*) between the affected and the non-affected side

**) of all randomly sampled motor unit potentials

The mean duration of motor potentials was $13.5 \pm$ per cent shorter in the muscle of the affected side than in the contralateral muscle and the mean amplitude was 23 ± 5 per cent decreased. The incidence of polyphasic potentials averaged 20 ± 1.5 per cent in the muscles of the affected side as compared to 10 ± 1.7 per cent in the contralateral muscle (Table 1).

In 16 patients in whom a tenotomy of the sternocleidomastoid muscle had been performed years ago, there was an interference pattern with full effort in 8 patients, a mixed pattern in 7 and discrete motor unit activity in 1. The amplitude of the pattern of activity was reduced in 7.

The mean duration of motor unit potentials was 10 ± 3.5 per cent shorter in the muscle of the affected and tenotomized side than in the contralateral muscle, and the amplitude of randomly sampled motor unit potentials was 15 ± 5 per cent lower. Polyphasic potentials occurred in 9 ± 2 per cent of all potentials in the muscles of the affected side, in 6 ± 2 per cent in the contralateral muscles (Table 1).

Trapezoid muscle. In 17 of the 27 patients studied there was an interference pattern during full effort and in 10 a somewhat reduced degree of activity. The average duration and amplitude of randomly sampled motor unit potentials were the same in the muscle of the affected side as in the contralateral muscle. The incidence of polyphasic potentials was 7 ± 1.5 per cent compared to 2 ± 0.5 per cent in the contralateral muscle.

Facial muscle. In 19 patients studied, the pattern of activity during full effort and the duration and amplitude of randomly sampled potentials were the same in the muscle of the affected side as in the contralateral muscle, whether or not there was a facial asymmetry.

DISCUSSION

The interference or mixed pattern of decreased amplitude in the sternocleidomastoid muscle of the affected side as well as the diminished duration and amplitude of motor unit potentials and the increased incidence of polyphasic potentials are consistent with the assumption of a primary myogenic origin of the congenital torticollis. Baxter et al. (1961) and Tönnis (1964) have reported similar findings; they related the electromyographic changes to the increased proliferation of connective tissue observed in biopsies from the affected sternocleidoid muscles.

Randomly distributed loss of muscle fibers and their replacement by

connective tissue can account for the electromyographic changes. Such changes can occur either secondary to a fibrositis on a vascular or inflammatory basis or to a defect in development. The familiar incidence is in favour of the latter. Döring (1939) reported a familiar occurrence in 15 per cent of patients with torticollis and found it often together with other malformations. In 8 of our 31 patients there was familiar occurrence of torticollis or other asymmetries. In 10 there was a shoulder and in 24 facial asymmetry in addition to the torticollis. These asymmetries are probably secondary to the developmental defect since there was no electromyographic evidence of a reduction in activity in the trapezoid and facial muscles.

SUMMARY

Electromyography of neck and facial muscles in patients with congenital torticollis showed a diminished duration and amplitude of the motor unit potentials as well as an increased incidence of polyphasic potentials in the sternocleidomastoid muscle of the affected side. This is consistent with a myogenic affection of the muscle.

There were no electromyographic abnormalities in the trapezoid and facial muscles. The shoulder and facial asymmetry is considered to be secondary to the changes in the sternocleidomastoid muscle.

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