Postural laterality in torticollis and torsion dystonia

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SUMMARY Tests of postural rotational laterality were carried out in 63 patients suffering from torticollis (53) or torsion dystonia with torticollis (10) and in 33 normal subjects. There was a correlation between the predominant direction of postural functions and the direction of the rotatory component of torticollis. Torticollis and torsion dystonia may be due to the release of a postural rotational laterality in pre-disposed subjects and not to a presumed asymmetric lesion of the brain-stem.

Stereotyped movements are a striking feature in axial dyskinesias (torsion dystonia and torticollis): the involuntary movement of the head, trunk and limbs is repeated in the same plane and in the same direction ("richtungsbestimmte Bewegungen"¹). Another characteristic sign of the axial dyskinesia is the rotational asymmetry of the non-volitional movement: a more or less marked turning movement to the right or to the left is present in every patient, whereas flexion, extension or tilting of the head (and trunk) may not always be present.²

Post-mortem findings contrast with the stereotyped and asymmetric clinical findings. A decreased number of ganglia cells in cortex, cerebellum and basal ganglia (mainly in caudate nucleus and putamen) in both halves of the brain has been reported by many authors.³⁻⁹ Only Hassler and Dieckmann¹⁰ described unilateral cell loss in the left thalamus in a case of predominantly left-sided torsion dystonia. Other authors were unable to find any pathomorphological changes which could be correlated with torticollis and torsion dystonia.¹¹⁻¹³ Histological sections from earlier reports were re-examined by Zeman and Dyken¹⁴ (from cases of torsion dystonia^{9 15}) and by Tarlov¹³ (from cases of torticollis¹⁶⁻¹⁸) with the conclusion that no pathological changes could be found. In some cases described as examples of torticollis or torsion dystonia, the clinical condition originally was reported under other titles,¹⁹⁻²¹ then later quoted.²²

What is the reason for the stereotyped rotational postures in patients suffering from torticollis and

torsion dystonia? It is possible that a pre-existing rotational laterality is unmasked by the disease. Accordingly, postural and related motor and sensory functions were tested in a group of patients with torticollis and in a control group of normal subjects.

Material and methods

Sixty-three adult patients (33 men and 30 women) aged between 17 and 57 years at the time of the onset of the illness (mean age 34.3 years, SD 9.58 years), 53 of whom had isolated torticollis and 10 of whom had torticollis with dystonic movements of the limbs, were divided into two groups according to the direction of rotation of the head: L, that is left-rotated ($n = 33$) and R, that is right-rotated ($n = 30$). The direction of rotation of the head was established by integrated fine-wire electromyography of the neck muscles (the sternocleidomastoid, the scalenus, the splenius, the levator scapulae, the trapezius and the cervical semispinal muscles of both sides), and by photographs taken from four sides of patients in the standing position. The objection that the laterality of motor functions could be the result of (and not the reason for) the asymmetric involuntary posture was irrelevant because not only patients with marked dyskinesia were tested (54%), but also those who had partially or completely recovered (46%).

The laterality of the hand was tested by 16 manoeuvres. The laterality of the auditory system was examined by means of a dichotic listening test,²³ in which 20 pairs of three-digit figures were synchronously delivered to both ears. For vestibular functions, the duration, the total amplitude, and the maximum velocity of the low phase of the caloric nystagmus elicited by the technique of Fitzgerald and Hallpike²⁴ were registered by means of electronystagmography in total darkness, without a fixation point. Only nystagmus elicited by cold water (20°C) was of sufficient intensity and suitable for statistical evaluation in all cases (the instability of the head due to dyskinetic movements caused artificial dysrhythmia and

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Received 11 December 1980, revision received 21 July 1981
Accepted 8 August 1981

inhibition of rotational and 44°C nystagmus). The laterality of turning of the body was examined by 16 tests without the subject realising the aim of the tests (the direction of turning back in the examination of walking, the direction of quick turning during the examination of stability in the standing and in the squatting position, all tests being repeated with eyes open and closed). The same vestibular functions and tests of turning of the body were examined in the control group of 33 adult normals (group N) aged between 17 to 45 years (mean age 29.2 years, SD 6.18 years). The mean values in each group were ascertained and the differences between groups L, R and N were statistically evaluated by Student's *t* test.

To confirm the constancy of the individual vestibular laterality, directional preponderance and "canal paresis" pattern of the caloric nystagmus (20°C and 44°C) were re-evaluated in 20 patients (10 L and 10 R) tested two years earlier. All had either completely recovered or exhibited a considerable reduction of dyskinesia; their EMG however retained asymmetric activity at rest (fig 1).

Results

No difference in hand laterality was observed between the torticollis groups L and R and the normal subjects. There was no correlation between the predominant hand and the direction of torticollis. Ninety-two per cent of the subjects were right-handed.

No difference in ear laterality was observed between the torticollis groups L and R and the normal subjects. There was no correlation between the predominant ear and the direction of torticollis. The right ear was dominant in 73% of subjects.

Nystagmus elicited by cold water was exaggerated bilaterally in groups L and R, exceeding mean values obtained in normals by 2%-17% for the duration, by 15%-70% for the total amplitude, and by 12%-62% for the maximum velocity of the slow phase (table 1). Nystagmus elicited by cold water in the torticollis groups L and R was asymmetric. Nystagmus with the slow phase in the direction of the rotation of the head was of longer duration, of greater amplitude and of higher velocity (figs 2, 3). There was a correlation between the greater nystagmus and the direction of torticollis (table 1). The

Fig 1 EMG pattern in a man previously suffering from torticollis with rotation of the head to the right. The dyskinesia disappeared, the prime-mover (*m. semispinalis cervicis* of the right side) is, however, still hyperactive at rest.

direction of nystagmus asymmetry was the same in all 20 patients re-examined, the slow phase of the preponderating nystagmus being consonant with the direction of torticollis in 17 of 20 patients.

In walking, standing and squatting tests, there was a greater tendency to turn to the left side in the torticollis group L and in normal subjects. There was a correlation between the preferred "turning

Table 1 Mean values of nystagmus evoked by cold water (20°C) in patients with torticollis to the left (L, n = 33) and to the right (R, n = 30) and in the control group (N, n = 33). Significance of differences

Group		Nystagmus with slow phase to the right			Nystagmus with slow phase to the left			Difference L/R Student's <i>t</i> -test	Significance
		L	R	N	L	R	N		
Duration in seconds	$\bar{x}$	126.7	145.0	124.6	146.4	134.3	124.4	2.23	p < 0.05
	SD	25.5	20.1	31.8	20.1	44.9	35.0		
Total amplitude in degrees	$\bar{x}$	185.4	193.9	161.5	250.3	182.4	146.8	3.29	p < 0.01
	SD	89.3	110.8	91.4	104.9	116.5	104.6		
Maximum velocity of the slow phase in degrees per sec	$\bar{x}$	22.1	23.4	19.6	30.5	22.0	18.7	3.47	p < 0.001
	SD	10.7	13.0	10.9	12.6	14.0	10.8		

Fig 2 Nystagmus pattern in a man suffering from torticollis with rotation of the head to the right. The total amplitude of the caloric nystagmus with slow phases to the right (TA_1) predominates.

direction" and the direction of torticollis ($p < 0.001$, table 2).

Table 2 Rates of turns about in walking, standing and squatting positions

Group	L	N	R
Turns to the left	89%	71%	46%
Turns to the right	11%	29%	54%
Total sum of tests	100%	100%	100%

Discussion

Maier²⁵ described experiments in which albino rats were faced with an insoluble problem in a Lashley

jumping apparatus. They were forced to jump and whatever solution they chose, they were "punished" in half of their jumps. Such rats tended to develop "behaviour-fixations"; after some time, they commonly jumped, for example, always to the right, regardless of the stimulus situation, even if the food was visible on the left-hand side. Maier states: "It appears that the fixated group develops some kind of adjustment to the test situation and is therefore able to prevent emotional tensions". The onset of an audiogenic epileptic seizure in rats is manifested by the tonic deviation of the head and tail to one side followed by the running in the circle in the same direction.

In some dogs placed into a standard empty room another stressful condition evokes an epileptic

Fig 3 Nystagmus pattern in a man suffering from torticollis with rotation of the head to the left. The total amplitude of the caloric nystagmus with slow phases to the left (TA_2) predominates.

“whirling fit”.²⁶ The rotational character of the seizure is its prominent feature and the direction of the turning is in most dogs constant. The “left-sided fit” begins with the turning of the head to the left, followed by circling to the left and then the animal falls on its left side and generalised convulsions appear.²⁷ During the interictal period, the “left-sided” dog walks in the cage predominantly in circles to the left. This stereotyped repetitive walking in circles of constant direction was described mainly in laboratory and zoo-reared foxes, dingos and wolves.²⁸ It was concluded that both the laterality of seizures and the motor stereotypy was based on a predominant asymmetry of postural functions.²⁹

Data is available to suggest that in healthy man

also exists a postural asymmetry (laterality) of rotational motor stereotypes. A predisposition for turning to the left prevails. In radiographs of the occipital condyles and C 1 vertebrae, Jirout³⁰ found a symmetrical position in 30% of healthy subjects, but in 57% the condyles were deviated to the right (that is the head rotated to the left), and in 13% the condyles were deviated to the left. In EMG studies there is a prevalence of activity of the left-sided semispinalis neck muscles (prime-movers of ipsilateral rotation of the head) in right-handed standing subjects.³¹ Rotational movements in basic motor patterns of the axis of the body are predominantly associated with tilting of the body to the other side, so the predominance of left-sided rotation is in

accord with the stabilographic observations.³² The authors showed in healthy subjects a tendency to tilt to the right, while other authors³³ showed that lateral oscillations of the vertical axis tended to be to the right side. In walking and stepping tests most right-handed people deviate to the right.³⁴

Asymmetry of postural functions in healthy subjects also has been shown in caloric, rotatory and optokinetic nystagmus (physiologic directional preponderance of nystagmus). The mean frequency of optokinetic nystagmus to the left is higher (3.57/s) than to the right (3.17/s).³⁵ In right-handed subjects, Jongkees³⁶ showed a directional preponderance of postrotational nystagmus to the left; Cigánek³⁷ found a directional preponderance of optokinetic nystagmus to the left. Caloric nystagmus to the left has been found to be larger and of longer duration³⁸ than nystagmus to the right and the velocity of its slow phase greater.³⁶

Most subjects with spasmodic and also with congenital torticollis rotate to the left (table 3).

Table 3 Direction of rotational component of torticollis

Authors	Type of torticollis	To the left	To the right
Arseni and Sandor, 1960 ³⁹	spasmodic	28	13
Sorensen and Hamby, 1965 ⁴⁰	spasmodic	43	28
Hamby and Schiffer, 1969 ⁴¹	spasmodic	27	23
MacDonald, 1969 ⁴²	congenital	68	34
Ling and Low, 1972 ⁴³	congenital	105	45
Bösch, 1972 ⁴⁴	congenital	229	158
Stejskal and Tománek	spasmodic	71	57

In our study, the direction of the rotational component of torticollis was found to parallel the rotational postural laterality. The dyskinesia may be regarded as due to pathological release of a pre-existing postural laterality.

The cooperation of the Data Processing Department of the UVN Hospital, Prague, is gratefully acknowledged.

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