

## SHORT REPORT

# Characteristics of dystonic movements in primary and symptomatic dystonias

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**Objective:** To compare clinical characteristics of the involuntary movements in primary and symptomatic dystonias.

**Patients and methods:** 132 consecutive patients with the diagnosis of primary dystonia and 51 consecutive patients with secondary dystonia caused by well defined structural lesion(s) of the central nervous system, with particular emphasis on the characteristics of involuntary movements.

**Results:** Eight variables with the highest risk contribution to either symptomatic or primary dystonias were identified: dystonic movement in secondary dystonia was much more frequently presented at rest, whereas the presence of dystonic tremor, chronic inflammatory process, or peripheral trauma located in the region that is later affected by dystonia, as well as the use of sensory tricks and development of spontaneous remissions, classified the affected patients more often in the category of those with primary dystonia.

**Conclusion:** The study identified several clinical features that may be helpful in differentiating primary from secondary dystonia.

Classification of dystonia according to aetiology differentiates between primary and secondary dystonias.<sup>1</sup> Virtually any structural or metabolic lesions of the brain, particularly involving putamen, other basal ganglia, rostral brainstem, and upper cervical lesions, have been associated with dystonia.<sup>2</sup> Sudden onset of occurrence at rest, cranial onset in childhood, onset in the legs during adulthood, hemidystonia, fixed postures, and early speech involvement are suggestive of secondary (symptomatic) dystonia.<sup>3</sup>

Taking the advantage of examining a relatively large group of patients with symptomatic dystonia caused by defined structural brain lesions,<sup>4</sup> we compared clinical characteristics of the involuntary movements in primary and symptomatic dystonias in order to define predictors for affiliation to one of these two groups.

## PATIENTS AND METHODS

The study comprised 132 consecutive patients (52% males) with the diagnosis of primary dystonia who were recruited for a genetic study at the Institute of Neurology (Belgrade) in the period from 1998 to 2001, and 51 consecutive patients (49% males) with secondary dystonia caused by well defined structural lesion(s) of the central nervous system (CNS), diagnosed in the period from 1991 to 2001. The group with secondary dystonia has been previously described in detail.<sup>4</sup> In brief, the group was composed of patients with clearly verified CNS lesion that correlated with the time of dystonia manifestation, substantiated by magnetic resonance imaging and/or computed tomography. Stroke was the main aetiology (34 patients; 66%), followed by brain tumours (six patients; 12%), head trauma with brain contusion (three patients; 6%),

multiple sclerosis (three patients; 6%), arterio-venous malformations (two patients; 4%); encephalitis (two patients; 4%), and one patient (2%) with thalamotomy for coarse postural tremor. There were no differences between patients with primary and secondary dystonia either in age at onset (41.0 (SD 17.6) and 42.1 (SD 14.4) years, respectively), or the duration of dystonia (11.1 (SD 7.5) and 9.2 (SD 5.4) years, respectively). Positive family history for dystonia was registered in 21 patients (16%) with primary, and in none of patients with secondary dystonia.

All the patients with primary dystonia fulfilled the following criteria: (a) presence of dystonia with different distribution (focal, segmental, multifocal, generalised); (b) normal prenatal, perinatal, and postnatal development; (c) lack of diagnostic criteria for “dystonia-plus” syndromes or other neurodegenerative diseases; (d) lack of data about toxin or drug exposure; and (e) normal neurological examination except for dystonia. Patients were observed and examined by two neurologists (VSK and MS) independently, with particular emphasis on the nature of involuntary movements.

## RESULTS

Patients with primary dystonia had mainly focal dystonia (96 patients; 73%), and less frequently segmental (26 patients; 20%) and generalised (10 patients; 7%) dystonia. Patients with secondary form had focal dystonia in 43% of cases (22 patients), followed by hemidystonia (16 patients; 31%), generalised (8 patients; 16%), and segmental dystonia (5 patients; 10%). Therefore, the appearance of focal dystonia was more frequently seen in patients with primary dystonia ( $p < 0.001$ ), while hemidystonia was present only among symptomatic cases.

Occurrence of dystonia at rest was noted in 41 (80%) patients with secondary, and in only 55 (42%) patients with primary dystonia ( $p < 0.001$ ). The site of symptom onset did not differ in various subgroups based on dystonia distribution among patients with idiopathic and secondary forms of the disease.

Jerky clonic movements were more frequently noticed in primary cases ( $p = 0.001$ ; table 1), while slow, athetoid movements were more common in symptomatic cases ( $p < 0.001$ ). Dystonic tremor of the affected body part was observed in 60% and 24% of cases with primary and secondary form of the disease, respectively ( $p < 0.001$ ; table 1).

Sensory tricks were used by 55 (42%) of patients with primary, and by only two (4%) of those with secondary dystonia ( $p < 0.001$ ; table 1). Complete or partial spontaneous remissions were registered in 34 (26%) and two (4%) of cases with primary and symptomatic form of the disease, respectively ( $p = 0.001$ ).

**Abbreviations:** CNS, central nervous system

**Table 1** Clinical characteristics of dystonic movements in patients with primary (n = 132) and symptomatic dystonia (n = 51)

	Patients with symptomatic dystonia	Patients with primary dystonia	p Value
Tonic type of dystonic movement	28/51 (55%)	68/132 (52%)	0.666
Clonic (jerky) dystonic movement	6/51 (12%)	44/132 (33%)	0.001*
Athetoid movements	17/51 (33%)	5/132 (4%)	0.000*
Dystonic tremor	12/51 (24%)	79/132 (60%)	0.000*
Dystonia at rest/action dystonia	41/10	55/77	0.000*
Postural tremor	11/51 (22%)	17/132 (13%)	0.143
Other involuntary movements	4/51 (8%)	18/132 (14%)	0.280
Sensory tricks	2/51 (4%)	56/132 (42%)	0.000*
Spreading of dystonia	17/51 (33%)	65/132 (49%)	0.05*
Spontaneous remission	2/51 (4%)	34/132 (26%)	0.001*
Diurnal fluctuations	12/51 (24%)	68/132 (52%)	0.001*
Degree of disability	48/51 (94%)	129/132 (98%)	0.069
Pain	32/51 (63%)	67/132 (51%)	0.145
Head trauma	2/51 (4%)	6/132 (5%)	0.853
Peripheral injuries	1/51 (2%)	25/132 (19%)	0.003*

\* Statistically significant; values present ratio of affected v total number of patients, with percentage in parenthesis.

Some form of chronic inflammatory process (such as conjunctivitis, furunculosis, phlegmona) affecting the site of dystonia presentation and antedating its occurrence was observed in 20 (15%) patients with primary, while such cases were not found among those with secondary forms. Also, peripheral injuries preceded dystonia in 25 (19%) and in only one (2%) patients with primary and symptomatic dystonia, respectively ( $p = 0.003$ ; table 1).

The results of the logistical univariate regression analysis revealed differences in clinical features between these two groups. When all significant variables identified by univariate regression analysis were jointly analysed with multifactorial analysis, seven variables with the highest risk contribution to either symptomatic or primary dystonias were identified: focal distribution of dystonia (odds ratio (OR) 3.33; 95% confidence interval (CI) 1.0 to 10.9); dystonia at rest (OR 11.96; 95% CI 2.8 to 51.8); jerky clonic movements (OR 6.42; 95% CI 1.7 to 25.3); athetoid movements (OR 0.04; 95% CI 0.01 to 0.2); dystonic tremor (OR 5.04; 95% CI 1.4 to 18.3); sensory tricks (OR 24.82; 95% CI 2.5 to 244.0); and diurnal fluctuations (OR 6.86; 95% CI 1.8 to 25.4).

## DISCUSSION

Our study has identified several significant differences between primary and secondary dystonia: dystonic movement in secondary dystonia is much more frequently presented at rest, while the presence of dystonic tremor, chronic inflammatory process, or peripheral trauma located in the region later affected by dystonia, as well as the use of sensory tricks and development of spontaneous remissions, classified the affected patients more often in the category of those suffering from primary dystonia. Prevalence of dystonia appearance at rest suggests a secondary form of the disease.<sup>5</sup> In both groups, focal dystonia was the most common type of dystonia, although significantly more frequently in patients with primary dystonia (80% v 42%;  $p < 0.001$ ). However, hemidystonia was registered only in patients with the secondary form of the disease. This is in accordance with the data showing that at least 75% of patients with hemidystonia have a contralateral damage of the basal ganglia.<sup>3</sup> Interestingly, contrary to suggestion that primary

dystonia has gradual onset and slow progression, while symptomatic dystonia has abrupt onset followed by a rapid progression,<sup>6</sup> we were not able to observe any difference between the groups in either the mode of onset or pace of progression.

Patients with secondary dystonia had a slightly higher incidence of dystonic posturing (that is, fixed dystonic postures), and appearance of slow, athetoid movements. However, those with primary dystonia had more frequently jerky clonic contractions. Also, dystonic tremor was observed in 60% of patients with primary and in only 24% of those with secondary dystonia.

Spontaneous remissions and the ability to suppress dystonic movement by sensory tricks also differentiate primary from secondary dystonias. Both phenomena were more frequently registered in primary dystonia, confirming the suggestion that they are typical for its clinical course.<sup>7</sup>

Data about chronic inflammatory process antedating dystonia of the same region were obtained in a survey of detailed histories in 15% of patients with primary and in none with secondary dystonia. Similar data have been obtained for peripheral injuries. Peripheral trauma induced dystonia differs in patients with primary and those with secondary dystonia, since in the latter it appears after a short latency of only a couple of days, involuntary movements appear either at rest or during sleep, there is a slight response to anticholinergics, and there are no efficient sensory tricks.<sup>8</sup> It was suggested that peripheral injury may cause primary dystonia particularly in genetically<sup>9</sup> or otherwise predisposed patients.<sup>10</sup>

About 75% of all patients with dystonia suffer from primary dystonia, while secondary dystonias cover the remaining quarter of all cases.<sup>3</sup> Our study identified several clinical features that may be helpful in differentiating primary from secondary dystonia.

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