Movement Disorders
Vol. 24, No. 12, 2009, pp. 1725–1731

Viewpoint

Is this Dystonia?

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Abstract: Torsion dystonia is characterized by sustained muscle contractions causing twisting and repetitive movements and abnormal postures. The diagnosis can be made difficult, delayed, and often misled by several factors: variability of dystonia presentation, uncertain recognition of the specific physical signs, lack of diagnostic tests, wide etiological spectrum, and coexistence of other movement disorders. Diagnostic tools are of limited assistance for the diagnosis of dystonia, which remains based on clinical diagnostic skills. We propose here, a new diagnostic algorithm to systematize the clinical diagnostic workout. A correct recognition of the physical signs that constitute the hallmark of most dystonia syndromes provides the grounds to perform a structured diagnostic sequence and share a consistent methodology. This clinical algorithm may be enhanced by adding diagnostic tools for dystonia, once their diagnostic value is assessed. © 2009 Movement Disorder Society

Key words: dystonia; diagnosis

CURRENT DIFFICULTIES IN THE DIAGNOSIS OF DYSTONIA

Dystonia syndromes are among the most commonly observed movement disorders in clinical practice.[1] Dystonia *musculorum deformans* was recognized as a new nosologic variety for the first time by Oppenheim, who identified some of its distinctive features in four Jewish children.[2] His clinical series is now considered an *ante litteram* report of the DYT1 phenotype. The phenomenology of dystonia coincided with the generalized phenotype for many decades, until June 1975, when an international conference chaired by Stanley Fahn in New York laid the way to the modern era and recognized the clinical features of focal forms of dystonia.[3] Marsden and coworker's[4–8] intuition then provided the intellectual glue for lumping together focal dystonia entities that were previously considered independent nosologic forms (e.g., blepharospasm, torticollis, "spastic" dysphonia, and writer's cramp). In 1984, an ad hoc committee of the Dystonia Medical Research Foundation documented the occurrence in all forms of dystonia "of sustained muscle contractions, frequently causing twisting and repetitive movements, or abnormal postures"[9]; later, it was recognized that the association of slow tonic posturing with faster (phasic) movements, sometimes resembling tremor, is the clinical hallmark of this movement disorder.[9–11] These characteristics still constitute the clinical core of dystonia, but their application to the wide-ranging phenomenology of different dystonia forms is a difficult task, because of lack of validated criteria and of a diagnostic algorithm.

The current diagnostic uncertainty has several reasons. First, the clinical appearance of primary dystonias is varied[12] and nonprimary cases represent a further diagnostic challenge, because the clinical presentation may show less typical features of dystonia sometimes intermixed with additional clinical signs.[13] As shown in Table 1, dystonia is the only physical sign of primary dystonia syndromes, whereas in nonprimary cases, it is associated with other movement disorders or other neurological signs. Moreover, there

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Potential conflict of interest: None reported.

Received 15 December 2008; Revised 26 February 2009; Accepted 17 March 2009

Published online 24 June 2009 in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/mds.22597

TABLE 1. *Combination of physical signs observed in different dystonia syndromes listed by etiological classification*

Dystonia syndromes	Physical signs observed
Primary (or idiopathic) dystonias	Dystonia
Dystonia plus syndromes	Dystonia, parkinsonism, myoclonus
Paroxysmal dystonias	Dystonia, chorea, myoclonus
Heredo-degenerative dystonias	Dystonia, parkinsonism, chorea, myoclonus, spasticity, cerebellar features, dysautonomia, cognitive impairment, epilepsy
Symptomatic dystonias	Same as for heredo-degenerative dystonias plus focal neurological signs, if present

is evidence that the clinical presentation of some secondary dystonias is different from typical primary torsion dystonia[11,12]; to date, no studies comparing the clinical features of primary versus nonprimary forms have been published.

Second, a number of conditions can be mistaken for dystonia because of sustained postures. To this regard, Fahn proposed to use the term "pseudodystonia" to encompass "disorders that can mimic torsion dystonia, but are not generally considered to be a true dystonia."[14] Uncertainties on terminology have remained since, and bridging terms, such as "spastic dystonia," have further contributed to vagueness of nomenclature.[15,16] Thence, the rule that, as a medical term, "dystonia" should point to a coherent and unique phenomenological set, appears not to be fulfilled. Third, there is a lack of consensus on the lower phenotypic edge of dystonia (so called *formes frustes*[17]). In such cases, the patients may play down mild clinical findings (a common occurrence in family studies[18]) or the examining neurologist may be uncertain whether to call attention to a mild phenomenology, such as excessive blinking[19] or occasional posturing.[20] The consistency and specificity of dystonia features need to be assessed in every patient. An European consensus panel acknowledged the lack of a specific diagnostic algorithm and recommended referral to expert observation in all uncertain cases to achieve a correct diagnosis of dystonia.[12]

The present review aims to answer the simple question "Is this dystonia?" that may come to the mind of the nonexpert examiner, by proposing a new diagnostic algorithm. Once validated, this will allow systematizing knowledge into a practical set of diagnostic criteria to serve as a guide for a simple and unambiguous recognition of the physical signs of dystonia.

CLINICAL FEATURES OF DYSTONIA

The clinical features of dystonia have been highlighted in recent years.[11,13] There is a need, however, to systematize current knowledge into objective criteria to serve as a basis for a diagnostic algorithm. Table 2 proposes a systematized description of the cardinal physical signs observed in dystonia. These features have to be looked for in all movement disorders, either fast or slow, also when the immediate impression is that of a "tremor," "tic," "chorea," or "myoclonus". The arguments supporting this systematization are succinctly reviewed here.

TABLE 2. *Clinical criteria for the physical signs observed in patients with dystonia*

Physical sign	Description
Dystonic postures	• A body part is flexed or twisted along its longitudinal axis (not available for blepharospasm or laryngeal dystonia) • A sensation of rigidity and traction is present in the affected part *Incomplete phenomenology: at least one feature; complete phenomenology: both features*
Dystonic movements	• A twisting nature or a pull in a preferred direction is detected (also when the movement appears as tremor) • Movements are repetitive and patterned (i.e., consistent and predictable) • Movements are often sustained at their peak to lessen when a given posture (usually opposite to the preferred direction) is identified ("null point") *Incomplete phenomenology: at least one feature; complete phenomenology: at least two features*
Gestes antagonistes (tricks)	• Alleviation of dystonia occurs during the *geste* movement, usually soon after its start • Alleviation may last for as long as the *geste* or slowly reverses spontaneously before its end • The *geste* movement is natural and "elegant," never forceful • The *geste* movement does not push or pull the affected body part, but simply touches it (sensory trick) or accompanies it during alleviation of dystonia *Present: all features*
Mirror dystonia	• At least three different types of repetitive tasks (e.g., finger sequence, normal writing, or piano-like movements) are performed at low and fast speed in the nonaffected limb *Present: at least during one task*
Overflow dystonia	• Dystonic movement or dystonic postures extend beyond the commonly involved body region • It is observed at least once, ipsilaterally or contralaterally, either by inspection or EMG mapping, in coincidence with the peak of dystonic movements *Present: both features*

Dystonic Postures and Dystonic Movements

The term "torsion" dystonia was used by Marsden[5] to identify the combination of dystonic postures and movements representing the hallmark physical signs observed in dystonia syndromes. The term "fixed" dystonia is used, instead, to identify conditions where dystonic postures, but no movements, are observed, as in the definition of dystonia originally proposed by Denny-Brown.[15] A dystonic posture flexes or twists a body part along its longitudinal axis. Dystonic movements are usually irregular and sustained at their peak, but at times are intermixed with regular, tremor-like movements, also called dystonic tremor.[21,22] Typically, dystonia produces a movement disorder that is predictable, as it involves one or more body regions, is consistent in pattern, has a directional quality and often has overlying spasms. In most instances, clinical observation is sufficient to identify dystonic postures (tonic component) and movements (phasic component).[11] The distinction between "phasic" dystonic movements that are mixed up with "tonic" postures was also recognized by the ad hoc committee[9] and applied to the clinical assessment of patients undergoing ablative stereotactic surgery.[23] However, a number of conditions (such as neuromuscular diseases, spasms, tonic seizures, etc.) may produce abnormal postures that mislead the observing clinician.[13] Furthermore, phasic dystonic movements can easily be mistaken for tremor (dystonic tremor) or myoclonus and viceversa.[21,22,24]

Dystonia has other unique activation/deactivation features that can be recognized if appropriately looked for by the examining neurologist,[11,13] namely, the *gestes antagonistes* (or sensory tricks), mirroring, and overflow.

Gestes Antagonistes (Tricks)

They have been typically described in patients with cervical dystonia who reduce or even abolish dystonic posturing while making one or more specific voluntary movements,[25,26] but can occur in any body districts.[27–29] The mechanism of action of the *ges*tes is debated[30] and their efficacy may diminish as disease progresses[31]; still, they are very characteristic and particularly useful for diagnostic purposes. Their phenomenology contrasts with dystonic postures and movements, as they are never forceful, instead natural and elegant, and their action is not associated to any mechanical correction by counterpressure.[32] The *geste* movement must have a typical appearance before it can be used as a criterion to support the diagnosis: alleviation of dystonia occurs immediately and affects both tonic and phasic components. It has been observed that *gestes* do not improve non-dystonic essential head tremor,[33] are uncommon in early post-traumatic dystonia[34] and have atypical phenomenology in patients with psychogenic movement disorders.[35]

Overflow and Mirroring

These are two related clinical phenomena that prove particularly helpful in cases with mild or inconstant phenomenology.[11] Overflow is an unintentional muscle contraction which accompanies, but is anatomically distinct from, the primary dystonic movement[20]; it is usually observed at the peak of dystonic movement as a spread of activity to adjacent, otherwise unaffected, body regions.[11] Overflow is considered a clinical expression of specific physiopathological abnormalities occurring in dystonia.[36,37] Mirror dystonia occurs on the affected body side when a specific task is performed by the homologous opposite normal body part. The specific task(s) capable to elicit mirror dystonia must be identified to appreciate this feature (Table 2). A typical example, observed in almost half of the patients with writer's cramp, is mirror dystonia of the dominant hand while writing with the opposite unaffected hand.[38]

DIAGNOSTIC TOOLS

Since the introduction of EMG analysis of dystonic movements, emphasis has been placed on sustained simultaneous contractions of agonists and antagonists.[39] EMG analysis can complement clinical observation in many ways. First, allows distinguishing the three types of abnormal activation (tonic, phasic, and tremulous) that correspond to the homonymous clinical features.[40] Tonic activity is characterized by an interference pattern with only slight variations of amplitude and density, phasic patterns consist of mostly synchronous bursts of activity in the dystonic muscles of variable duration (from 250 ms to several seconds), tremulous activity consists of rhythmic bursts in several muscles with duration between 50 and 300 milliseconds. Furthermore, multiple simultaneous EMG recordings (mapping) can be performed at rest or during activation/deactivation tasks to detect features of dystonia with greater detail than allowed by visual inspection (Fig. 1). EMG mapping detects coactivation of agonist muscles, that is typically observed in limb dystonia and with less confidence in other focal forms (e.g., cervical dystonia[41]). As reported by the American Academy of Neurology, EMG evaluation is useful to differ-

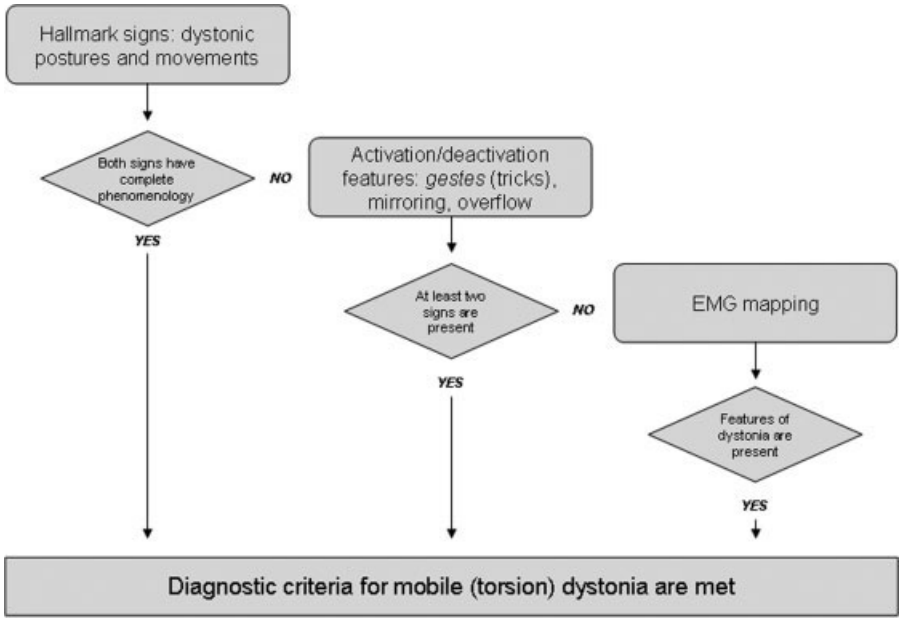

FIG. 1. Flow chart for the diagnosis of mobile (torsion) dystonia. This stepped approach allows reaching a clinical diagnostic level based on the criteria listed in Table 2.

entiate different types of tremors from myoclonus and dystonia, and helps identify the muscles involved and the actual disease topography.[42] The complementary value of EMG analysis to physical examination is confirmed by the observation that, without the aid of EMG, in cervical dystonia 41% of dystonic muscles would be unrecognized and 25% of inactive muscles would be erroneously judged by inspection.[43]

Attempts to quantify activation/deactivation tasks have been performed in patients with blepharospasm. It has been reported that blink rate both at rest and during conversation is higher in patients compared with controls,[44] and that 76% of patients blinked more at rest than during conversation, whereas 74% of controls blinked more during conversation than at rest. This reversal in the normal pattern of blinking[45] suggests that conversation may attenuate blepharospasm similarly to a *geste antagoniste*. The sensitivity and specificity in discriminating patients and controls were found to be best with a resting blink rate above 27 blinks per minute. These criteria can be helpful when observing patients with cranial involvement. A limitation is that activation/deactivation maneuvers have not been quantified for tasks involving the limbs. Other neurophysiological studies have also identified additional features which are characteristically seen in dystonia. Circuit studies have shown that dystonia is characterized by increased excitability at different CNS levels: intracortical (as demonstrated by paired transcranial magnetic stimulation[46–48] and silent period study[49,50]), brainstem (shown by cranial reflex pathway studies[51]), and spinal cord (demonstrated by H reflex recovery curve and reciprocal inhibition[52–54]). These evidences indicate that the distinctive electrophysiological abnormalities of dystonia have a wider somatic representation than visible by clinical inspection. However, there are no prospective analyses of the sensitivity and specificity of these techniques, or of their correlation to clinical features, thus limiting their diagnostic applicability. Furthermore, some studies suggest that the predictive value of reflexological studies may be low: it has been shown that the blink reflex recovery cycle does not discriminate between focal dystonia and focal tics,[19,55] and there is evidence that the R2 recovery index is normal in one third of patients with blepharospasm and in two third with torticollis.[56]

In addition to neurophysiological tests, neuroimaging holds promises as a diagnostic tool in dystonia. Conventional MRI is critical for the assessment of patients with secondary dystonias.[13] Research with new imaging techniques, instead, has a diagnostic potentiality in primary forms. Reduced white matter integrity has been observed by DTI-MRI in the subgyral region of the sensorimotor cortex in patients with DYT1 dystonia[57] and fractional anisotropy has been found to be reduced in patients with primary dystonia.[58] White

matter changes have also been identified in patients with focal primary dystonias,[59,60] and it remains to be evaluated if these MRI features can be used to complement the clinical diagnosis. In addition, functional neuroimaging studies can reveal abnormalities across a broad network of regions in primary dystonias. Abnormal activity has been reported in premotor, supplementary motor cortices, and somatosensory motor cortex. However there is a discrepancy among studies showing overactivity of the premotor and prefrontal motor planning cortex and underactivation of the primary sensorimotor cortex[61–63] as well as overactivation of the primary sensorimotor cortex and underactivation of the prefrontal motor areas.[64]

DIAGNOSTIC ALGORITHM

On the basis of the evidence succinctly reviewed here, EMG mapping is the only clinical tool with a proven value for the diagnosis of dystonia to complement clinical examination. Thence, the new algorithm proposed in Figure 1 leads to the diagnosis of dystonia based on clinical observation and EMG complementation. When the full-house phenomenology is observed, the clinical diagnosis is plainly achieved by direct physical examination (left path). Otherwise, additional clinical signs are necessary (middle path). The observation of EMG abnormalities typical of dystonia is helpful when the clinical features are considered insufficient to the diagnosis (right path).[12] The clinical features need not be severe, but the objective criteria (Table 2) are to be met in all cases. The algorithm proposed here provides a mean to reliably estimate the phenomenology of dystonia also when it is minimal. Neurophysiological tests are not routinely recommended unless necessary.

In our experience, this methodology can be easily and effectively implemented in most forms of dystonia, either primary or secondary, and is particularly helpful in clinical genetic studies to identify the specific features of the affected individuals. A systematic application of this algorithm to different etiological forms of dystonia is warranted to assess its specificity and sensitivity. Some dystonia types may be more difficult to ascertain than others. Paroxysmal dyskinesias provide a particularly challenging setting, because dystonia occurs in brief episodes with normalcy in between and can coexist with chorea and myoclonus.[65,66] The implementation of this flow chart depends on the possibility to detect episodes of dystonia in these patients. Fixed dystonia, instead, remains out of the scope of this analysis. Fixed, immobile dystonic postures are characterized by a strong limitation of passive range of motion, absence of *geste antagonistes*, and association with other diseases, such as Parkinson's disease[67] or complex regional pain syndrome.[68]

This algorithm complements without overlapping the available rating scales that score topography, provoking factors, severity and duration of dystonia,[69–71] but do not assess dystonic postures and movements, mirroring, or overflow. One scale for cervical dystonia[70] specifically appraises the efficacy of sensory tricks (*gestes*), but does not consider whether the phenomenology of tricks is typical, a task left to the diagnostic algorithm. In keeping with complementarities, the present description does not consider pain, a common feature of cervical dystonia,[72] that is measured by a dedicated rating scale[70] and is not used for diagnostic purposes. In recent years, vigorous efforts have been engaged in classification of dystonia[73] and the foundations for the assessment of the clinical features of dystonia have been laid.[11,13] Nevertheless, the mechanisms involved in the pathophysiology of dystonia, the wide range of clinical presentations and the absence of a unique and unequivocal gold standard diagnostic tool make the diagnosis of dystonia firmly linked to the patient's history and the physical examination by a skilled neurologist. In such scenario, the proposal of a simple algorithm firmly based on the clinical features of proven diagnostic value may provide a shared diagnostic methodology. Future studies will allow reckoning its specificity and sensitivity, and may further enhance it by adding other diagnostic tools once their diagnostic value is assessed.

Acknowledgments: This work was supported, in part, by grants from the Italian Ministry of University and Research and the Italian Ministry of Health.

Author Roles: Alberto Albanese: Manuscript: Conception, writing of the first draft, review and critique; Stefania Lalli: Manuscript: Bibliographic search, review and critique.

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