

“Spastic dystonia” or “Inability to voluntary silence EMG activity”? Time for clarifying the nomenclature

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We thank Lumsden and colleagues for their comments on our paper “Spastic dystonia in stroke subjects: prevalence and features of the neglected phenomenon of the upper motor neuron syndrome” (Trompetto et al. 2019). We agree with them that precise terminology is essential for unambiguous communication and sharing of knowledge. Since this goal is far from being achieved in the field of the positive phenomena of the upper motor neuron syndrome (UMNS), we agree that the time has come to find a nomenclature able to capture the essential and unique features of the phenomenon so far reported as “*spastic dystonia*”, thereby eliminating eventual confusion. Realistically, this should be matter of dedicated reflections, but we welcome the opportunity offered by Lumsden and colleagues (Lumsden et al. 2019).

The main points of the subject (and debate) are the following.

First, the currently used term “*spastic dystonia*” refers to the inability of patients with UMNS to voluntary silence muscle activity on command (Gracies 2005; Sheean and McGuire 2009; Trompetto et al. 2014). This inability leads to spontaneous tonic muscle contractions, which prove stretch-sensitive (Gracies 2005), and ultimately amplify the velocity-dependent hypertonia (Marinelli et al. 2017; Trompetto et al. 2019).

Second, as an inability to relax muscles at will, the phenomenon falls in the most recent definition of dystonia, “a movement disorder characterized by sustained or intermittent muscles contractions causing abnormal, often repetitive, movements, postures, or both” (Albanese et al. 2013).

Third, since “*spastic dystonia*” may occur in the absence of velocity-dependent hypertonia (Denny-Brown 1966), which is the hallmark of spasticity, why qualifying this phenomenon as “*spastic*”? But: if this is not “*spastic*”, should it be simply “dystonia”, “secondary dystonia”, or “dystonia in the context of the UMNS”?

As delineated above, the debate prompts comments that move from the phenomenological to the pathophysiological level. We shortly review strengths and weaknesses for each point, and postpone eventual solution to more thorough review.

From a phenomenological point of view, we consider as undisputed that dystonia and “spastic dystonia” manifest similarly, therefore the involuntary muscle activity we are dealing with is dystonic in nature. But it is only one nature of the abnormal movement, the second one being its exquisite sensitivity to muscle lengthening.

Equally undisputed is the clearly distinct pathophysiology between “dystonia” and “*spastic dystonia*”. The overwhelming majority of papers using the term “dystonia” refers to clinical conditions stemming from structural or functional diseases of the basal ganglia, unrelated to the activity of the stretch reflex. On the contrary, “*spastic dystonia*” is found in patients having lesions outside the basal ganglia, exhibiting clear abnormality in the excitability of the stretch reflex

circuitry. Because definitely distinct pathological mechanisms give rise to muscle overactivity sharing similar phenomenology, the term “dystonia” seems inadequate for describing the involuntary muscle contraction we are dealing with.

Even from a nosological perspective, the term “dystonia” appears inappropriate. By definition, “dystonia” is a *movement disorder*, whereas “*spastic dystonia*” is not. Indeed, a *movement disorder* (meaning to indicate a disease of the basal ganglia) is defined as a “neurological syndrome in which there is either an excess of movement or a paucity of voluntary and automatic movements, unrelated to weakness or spasticity” (Fahn et al. 2011). It is apodictic how much “*spastic dystonia*” can be related to both weakness and spasticity. Corollary is that spastic dystonia can neither be considered *tout-court* “a subset of dystonia” (Lorentzen et al. 2018).

For old-fashioned “motorist” neurologists, the term “*spastic dystonia*” reflects perfectly the double nature of the stretch-sensitive (although not stretch-induced), spontaneous, tonic muscle contraction that can be observed in the absence of movement (as sole inability to relax) or during movement (as associated postural reactions) in patients affected by UMNS. None of the semantic nuances, such as “dystonic spasticity” (Lorentzen et al. 2018), or “dystonia with spasticity” or “dystonia accompanying the Upper Motor Neuron Complex” (as suggested by these respected authors) is likely to improve the understanding or the clarity of this similarly old-fashioned concept. But as time goes on, the audience of physicians dealing with UMNS patients grows, and we see the need for changing the term “*spastic dystonia*”. Nevertheless, we must acknowledge the difficulty to find an equally efficacious two-word descriptor, probably reflecting the peculiarity and complexity of this motor phenomenon (sensitivity to - but not dependency from peripheral input; occurrence both in the absence and during movement; ability to cause or not abnormal postures).

The letter by Lumdsen et al. (2019) proves that “*spastic dystonia*” seems no more conducive to the recognition of the phenomenon that it wants to identify, probably explaining why it is “the neglected phenomenon of the UMNS” (Trompetto et al. 2019). A newly proposed definition as a “stretch- and effort-unrelated sustained involuntary muscle activity following central motor lesions” (Lorentzen et al. 2018) certainly avoids confusion in terms of dystonia, but it fails to convey the inability to relax muscles, it overestimates the “*un-relation*” to muscle stretch, and uncouples this involuntary muscle activity from velocity-dependent hypertonia, which is the more evident clinical sign in these patients (Marinelli et al. 2017; Trompetto et al. 2019).

Aside from shading dystonia vs. spasticity and *vice versa*, we consider that “inability to voluntary silence EMG activity” describes well both the phenomenological and pathophysiological level, the positive phenomenon of the UMNS currently referred to as “*spastic dystonia*”. The ensuing stretch-sensitive spontaneous tonic muscle contractions favor/promote velocity-dependent hypertonia, and

may produce abnormal postures. This physiological construct would bring the concept to the clinicians that two are the causes of velocity-dependent hypertonia: spasticity (Lance 1980) and “inability to voluntary silence EMG activity”. Such a distinction would help prevent the overdiagnosis of “spasticity” whenever the clinical examination reveals the presence of velocity-dependent hypertonia, and will strengthen the idea that only the use of surface-EMG can discriminate between muscles that are relaxed prior to passive stretch (i.e., muscles affected by spasticity), and those showing stretch-sensitive spontaneous tonic contractions (i.e., muscles affected by “inability to voluntary silence EMG activity”).

In conclusion:

- spasticity and “inability to voluntary silence EMG activity” are distinct positive phenomena of the UMNS
- both the muscles affected by spasticity and those affected by “inability to voluntary silence EMG activity” show velocity-dependent hypertonia
- spasticity and “inability to voluntary silence EMG activity” are not clinical signs, because they cannot be distinguished on a clinical base alone
- surface-EMG is necessary to distinguish spasticity from “inability to voluntary silence EMG activity”
- pathophysiology of “inability to voluntary silence EMG activity” in UMNS is currently undefined.

Conflict of interest

None of the authors have potential conflicts of interest to be disclosed.

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