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Letter to the Editor

"Spastic Dystonia", "Dystonia with Spasticity" or "Dystonia accompanying the Upper Motor Neuron Complex"? A reconciliation of nomenclature is needed

We read with interest the study by Trompetto and colleagues examining the prevalence of "spastic dystonia" in stroke subjects (Trompetto et al., 2019). It has been our observation in children and young people experiencing arterial ischemic stroke that a mixed motor picture typically develops over time, with negative and positive motor phenomena commonly coincident. Careful assessment of this motor profile is essential, identifying which components are contributing to the functional difficulties experienced by the patient and in what manner to guide appropriate intervention. Consistent with the findings in this cohort of 23 patients with hemiparetic stroke, spasticity, i.e. a velocity dependent increase in tone, typically contributes less to functional difficulties in comparison to the impact of involuntary posturing of the affected limbs. Indeed, the Trompetto cohort exhibit the phenomenon of a maintained heightened EMG discharge during a maintained stretch, thus demonstrating a 'length-dependent' increased EMG output over baseline (a classic dystonic response to imposed stretch).

As Trompetto and colleagues acknowledge, the terminology with which the involuntary muscle contraction described in this cohort of subjects remains a matter for debate (Trompetto et al., 2019). Precise nomenclature and terminology are essential for the sharing of knowledge in both the clinical and research settings. Whilst we entirely agree with the authors' definition of "spasticity" as a dynamic phenomenon (i.e. a velocity dependent increase in tone), we would welcome discussion on the utility of the term "spastic dystonia". Definitions of dystonia have evolved over time, with the most recent consensus definition by Albanese and colleagues defining dystonia as "*a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures or both*" (Albanese et al., 2013). The definition for "spastic dystonia" Trompetto and colleagues reference, *spontaneous tonic muscle contraction occurring at rest*, at an operant level is not distinct from this broader definition of dystonia – i.e. clinical phenomena fulfilling the criteria for definition of "spastic dystonia" would equally fulfil the criteria for "dystonia". At a phenomenological level, within these current definitions "spastic dystonia" may be considered as a subset of "dystonia", as has been acknowledged by some authors (Lorentzen et al., 2018). Whilst not fully understood, the pathophysiology resulting in the hypertonicity described as "spastic dystonia" is clearly distinct from that underlying "spasticity". Furthermore, as elegantly demonstrated by Trompetto and colleagues, "spastic dystonia" frequently occurs entirely in the absence of "spasticity". Given these observations, applying the adjective "spastic" to this form of dystonia is misleading, potentially confusing the important message that persistent involuntary muscle contraction should be distinguished from spasticity in clinical practice.

It is our belief that the labelling of subjects with motor disorders as "spastic" or "dystonic" is overly reductive, and a more nuanced approach to defining and describing motor impairment is required, beginning with the acknowledgement that these patients have a disorder of ***movement*** and ***posture***. It is telling that the published consensus paper by Albanese and colleagues does not contain the word "spasticity" (Albanese et al., 2013). Equally, it has been our observation that reports in recent years focusing on "spastic dystonia" typically make no reference to a broader definition of dystonia (Gracies, 2005; Trompetto et al., 2019). A reconciliation of nomenclature is required, to ensure that clinicians and researchers alike are working with a common language, and also to a common purpose. Research into the pathophysiology of spasticity and dystonia must not become the domains of entirely separate clinical research communities.

It is unclear at present to what extent different forms of dystonia share common or overlapping pathophysiological processes. We agree with Trompetto and colleagues that the use of EMG, and indeed other neurophysiological techniques, can facilitate a greater understanding of the nature of a given individual's hypertonicity. Inclusion of neurophysiological assessments should be encouraged, both as an extension of the clinical examination and in the research domain. Over time it may become clear that beyond a common clinical phenomenology, there are distinct and disparate disorders of motor functioning which may be identified, enabling classification and categorization of different elements of a given patient's hypertonicity on a more objective level, as enunciated in the recent *operational definitions* within the *'European consensus on the concepts and measurement of the pathophysiological neuromuscular responses to passive muscle stretch'* (van den Noort et al., 2017). We would urge for a broad application of the term dystonia until such pathophysiological clarity is resolved. When a combination of spasticity and dystonia is identified, terms such as "dystonia with spasticity" or "dystonia with features of the upper motor neuron syndrome" should be considered in preference to "spastic dystonia".

The study by Trompetto and colleagues provides important data, which contributes to advancing our understanding of the fundamental contributors to disturbed function in patients with motor disorders. We look forward to further discussion and debate within the clinical and research communities to arrive at a mutually beneficial reconciliation of nomenclature currently used across the field.

Conflict of interest

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

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