

Dystonia After Severe Brain Injuries

In their recent publication in *Movement Disorders*, Albanese et al.¹ reported the results of a consensus update on the phenomenology and classification of dystonias. They encouraged clinicians and researchers to test their innovative classification scheme in various clinical settings. Motivated by their report, we aimed to assess their classification of dystonias among patients in a vegetative or minimally conscious state after experiencing traumatic brain injury (TBI), vascular disease, or cerebral anoxia.

Of the 132 consecutive patients who were admitted to our Department of Rehabilitation in a vegetative or minimally conscious state after sustaining severe brain injury, 28 patients developed dystonias. According to axis I of the new classification, we observed the following: (1) focal dystonias (i.e., cervical dystonias, blepharospasms, and oromandibular dystonias); (2) hemidystonias; and (3) generalized dystonias. Dystonias were more frequent in patients who suffered brain injury caused by cerebral anoxia (32%), compared to patients with TBI (24%) or cerebrovascular disease (10%). Generalized dystonias were prevalent in patients with cerebral anoxia, whereas focal dystonias were predominant after TBI. During the rehabilitation period, some patients showed improvements in their dystonic patterns, whereas the conditions of other patients remained stationary (static dystonia) or worsened (progressive dystonia).

Some characteristics observed in our patients and data reported in the literature may contribute additional insights to the new classification. First, onset of acquired dystonias may occur with a bimodal temporal distribution. Previous studies defined early-onset acquired dystonias as events that occurred over a short time interval (weeks or months) after cerebral damage.²⁻⁴ Late-onset acquired dystonias were considered to occur after a longer interval of time (≥ 1 year).^{4,5} We followed up patients in the postacute rehabilitation phase; thus, we only evaluated early-onset acquired dystonic events. Second, the clinical course of dystonia showed three distinct temporal patterns: worsening; static; or improving. Notably, improvements in acquired dystonia were previously described in patients who suffered from movement disorders caused by traumatic injury² or by stroke.³

In light of these observations, we suggest some updates to the new classification. In particular, we recommend that (1) “early- or late-onset acquired dystonia” be added as a new clinical descriptor, (2) disease course descriptors be changed

to “worsening, static, or improving,” and (3) “cerebral anoxia” be added as a possible cause of acquired dystonia. ■

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Reply: Dystonia After Severe Head Injuries

We thank Dr. Boccagni and colleagues for their comments and appreciate their evaluation of the new classification of dystonia in a series of patients with vegetative and minimally conscious states. Based on such experience, which refers to a very specific and infrequent clinical scenario, they make three proposals.

The first is to add a new clinical descriptor called “early-onset or late-onset acquired dystonia” related to the time course after the causative lesion. In the new classification, we aimed at separating clearly phenomenology (Axis I) from etiology (Axis II). Age at onset is listed among the phenomenological features, quite distinct from etiology. Boccagni and colleagues, instead, propose a new phenomenological descriptor related to etiology: early or late onset after injury. Although this type of information may be interesting, it is often impossible to determine time from insult because an

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Relevant conflicts of interest/financial disclosures: Nothing to report.

Received: 11 July 2013; Revised: 23 September 2013;
Accepted: 20 January 2014

Published online 3 March 2014 in Wiley Online Library
(wileyonlinelibrary.com). DOI: 10.1002/mds.25840

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Relevant conflicts of interest/financial disclosures: Nothing to report.

Received: 4 December 2013; Revised: 12 January 2014;
Accepted: 20 January 2014

Published online 3 March 2014 in Wiley Online Library
(wileyonlinelibrary.com). DOI: 10.1002/mds.25861

insult may not be evident and the time of onset after an injury may be confused with age at onset. This addition could only be used for some acquired dystonia forms, such as delayed-onset progressive dystonia after a static lesion, a relatively rare and specific occurrence. We did not include a number of other possible descriptors that only refer to specific applications, considering that they could be added for particular situations. We do not agree with adding this item and reiterate that the separation of phenomenology from etiology is an important feature of the new classification.

A second suggestion by Boccagli and colleagues is to change the description of disease course from “static or progressive” to “worsening, static, or improving.” “Worsening” is considered synonymous with “progressive” and the substitution, we feel, would not make a meaningful difference. The suggestion of adding “improving,” or even “relapsing,” is interesting and may warrant further consideration, although it deals with a very infrequent occurrence and it may be difficult to dissociate spontaneous improvements from responses to intervening therapeutic interventions. Also, the proposed change relates to a definable single insult and may not be useful in the overall scheme. In practice, additions to the disease course description would be easy to make.

The final suggestion is to add “cerebral anoxia” to the possible causes of acquired dystonia. Although we have not explicitly listed this diagnosis in the etiological list, we mentioned a vascular etiology and acknowledge that the list of possible causes did not intend to be exhaustive, because this classification aimed only at providing a structure for classifying different etiologies, and we only listed some examples. Certainly, cerebral anoxia and any other cause of acquired dystonia should be included in any comprehensive list of etiologies.

At the present time, making changes to the new classification seems premature. We, however, encourage testing it in real clinical practice, as Boccagni and colleagues have done. A review may be performed after 1-2 years of implementation. It is likely that the special needs of other types of dystonias may lead to additional suggestions for change to the classification system, which might be addressed sometime in the future, rather than piecemeal, when more consensus regarding what changes are most compatible with the overall needs will be gathered. ■

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