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“Complex” Dystonia Is Not a Category in the New 2013 Consensus Classification

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The Task Force for Nomenclature of Genetic Movement Disorders recently proposed a new system of naming genetically determined movement disorders.¹ These recommendations apply to a wide spectrum of movement disorders, including parkinsonisms, dystonias, choreas, paroxysmal movement disorders, cerebellar ataxias, and spastic paraplegias. We agree that current genetic classifications are cumbersome and “unsuitable” for accurate and comprehensive categorization of genetic movement disorders.

We appreciate that the task force addressed the discordance between phenotype and list designation by assigning more appropriate phenotype-prefix relationships. They propose 29 DYT symbols for inherited dystonias that occur in isolation or combined with other movement disorders, designated by an additional symbol: PARK, CHOR, or NBIA. The task

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force focuses on “prominent features” of diseases linked to mutations in a specific gene. It remains unclear, however, whether prominent features are the “most common” or rather the “most severe” reported phenotype. The terms “prominent” and “common” are subjective, therefore matter to dispute, and were not used in the 2013 consensus classification for the dystonias.

Concerning dystonia, the task force review mentions implementing the recently published new classification but mistakenly refers to 3 dystonia subgroups of isolated, combined, and complex and incorrectly suggests that these categories correspond to the previous terms of primary, dystonia-plus and secondary dystonia.

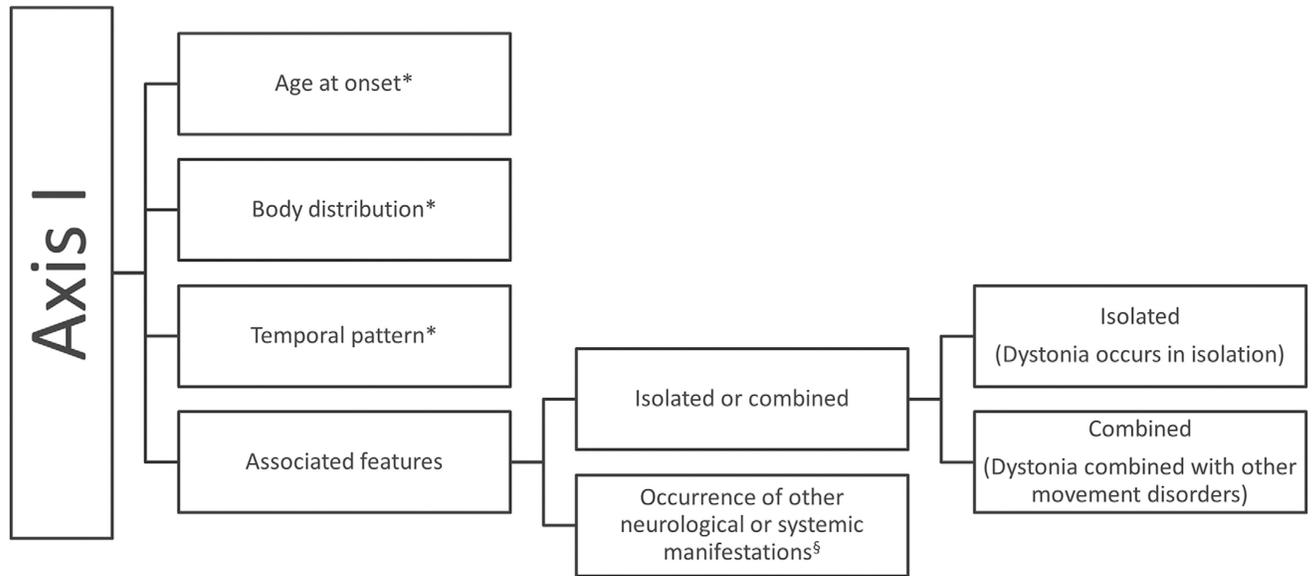
The 2013 consensus classification of dystonia has 2 separate axes: clinical and etiological. Axis I provides a guide to patient evaluation based on clinical characteristics, allowing construction of a syndromic (phenotypic) diagnosis that then guides etiological differential diagnosis, including genetic etiologies. In considering whether there are any associated movement disorders, there are only 2 subgroups: isolated and combined dystonia (disorders in which dystonia frequently coexists with other movement disorders).² The task force review added a third category of “complex” dystonias (in which dystonia dominates the clinical picture, but this occurs in the context of a complex phenotype including symptoms other than movement disorders). In the consensus classification of dystonia, other neurological or systemic features are a separate item of axis I, in recognition that these can be present both in patients with simpler, isolated dystonia phenotypes and those with more complex combined phenotypes. The tripartite definition proposed by the task force review does not accommodate for genetic conditions with isolated dystonia phenotypes that occur in combination with extraneurological manifestations (eg, liver involvement in Wilson disease presenting with isolated dystonia).

To clarify the distinction between associated movement disorder features and coexisting neurological or systemic manifestations in the 2013 classification of dystonia, here we provide an explanatory figure to prevent further misinterpretations of the new classification scheme of dystonia.

The main innovation of the 2013 dystonia classification is the distinction between clinical features and etiology, which is separated into 2 separate axes. A similar distinction is being adopted for new classifications of other movement disorders. This also fits well with the proposed new genetic nomenclature, in which the “prefix” denotes a broad clinical syndrome (phenotype) and the “suffix” the gene. However “complex” dystonia is not a category in axis I of the dystonia classification, and genotype falls under the etiological axis II of inherited dystonia.

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**FIG. 1.**

Clinical characteristics listed under axis I. The distinction of isolated and combined dystonias is reported under “Associated features.” *Further subdivisions of this axis I category are not shown here and can be found in the published classification²; §Other neurological or systemic manifestations may coexist with either isolated or combined dystonia.