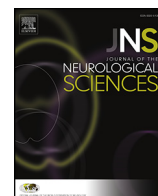




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## Review Article

## Scales for hyperkinetic disorders: A systematic review

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## ABSTRACT

Hyperkinetic movement disorders represent a heterogeneous group of disorders in which involuntary movements are the prevalent clinical symptoms. The five main categories of hyperkinetic disorders are tremor, dystonia, tics, myoclonus and drug-induced dyskinesia.

The severity of hyperkinetic disorders is assessed by all clinicians when they examine a patient; quantifying the severity also provides a means of studying the natural history of a given disorder and the possible effect of new therapeutic interventions. This means that good rating instruments are required in both everyday practice and experimental settings. Unfortunately, the clinical evaluation of these disorders is complicated by the inherent nature and variability over time of involuntary movements. A number of scales have been proposed over the years to study the various hyperkinetic disorders. The aim of this review is to systematically identify all the clinical scales that have been proposed and to classify them according to the criteria developed by the Movement Disorder Society (MDS) task force for rating scales in Parkinson's disease. On the basis of this methodology, a scale may be defined as 'Recommended', 'Suggested' or 'Listed' in decreasing order of value.

We found that, although numerous scales aimed at assessing hyperkinetic disorders have been published, their variability in terms of clinimetric properties, availability and effort required to administer them is high. In this evaluation, we identified scales defined as 'Recommended' for the assessment of all forms of hyperkinetic disorders. The situation highlighted by our analysis varies considerably, with several 'Recommended' scales being available for some conditions such as tics or dystonia, but only one being available for myoclonus. This gap needs to be filled by the scientific community through both the development of new clinical tools and the refinement of existing ones.

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## Contents

1.	Introduction	0
2.	Scales for hyperkinetic disorders	0
2.1.	Tremor	0
2.1.1.	Fahn–Tolosa–Marin Tremor Rating Scale	0
2.1.2.	Washington Heights–Inwood Genetic Study of Essential Tremor Rating Scale	0
2.2.	Dystonia	0
2.2.1.	Blepharospasm Disability Index	0
2.2.2.	Cervical Dystonia Impact Scale	0
2.2.3.	Toronto Western Spasmodic Torticollis Rating Scale	0
2.2.4.	Craniocervical Dystonia Questionnaire	0
2.2.5.	Voice Handicap Index	0
2.2.6.	Vocal Performance Questionnaire	0
2.2.7.	Fahn–Marsden Dystonia Rating Scale	0
2.3.	Chorea	0
2.3.1.	Unified Huntington's Disease Rating Scale	0

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2.3.2.	Abnormal Involuntary Movement Scale	0
2.3.3.	Modified motor score of the Unified Huntington's Disease Rating Scale	0
2.4.	Tics	0
2.4.1.	Global Tics Rating Scale	0
2.4.2.	Hopkins Motor and Vocal Tic Scale	0
2.4.3.	Motor Tic, Obsessions and Compulsions, Vocal Tic Evaluation Survey	0
2.4.4.	Premonitory Urge for Tics Scale	0
2.4.5.	Shapiro Tourette Syndrome Severity Scale	0
2.4.6.	Tourette's Disorder Scale	0
2.4.7.	Tourette Syndrome–Clinical Global Impression	0
2.4.8.	Tourette Syndrome Global Scale	0
2.4.9.	Unified Tic Rating Scale	0
2.4.10.	Yale Global Tic Severity Scale	0
2.5.	Myoclonus	0
2.5.1.	Unified Myoclonus Rating Scale	0
2.6.	Drug-induced dyskinesia	0
2.6.1.	TDRS: Simpson Tardive Dyskinesia Rating Scale	0
2.6.2.	DIS-Co: Dyskinesia Identification System—Coldwater	0
2.6.3.	ESRS: Extrapyramidal Symptoms Rating Scale	0
3.	Conclusions	0
	Acknowledgments	0
	References	0

## 1. Introduction

Hyperkinetic movement disorders represent a heterogeneous group of disorders in which unwanted (involuntary) movements are the prevalent clinical symptoms. These disorders are usually linked to basal ganglia dysfunction [1]. The five main categories of dyskinesia are tremor, dystonia, tics, myoclonus and drug-induced dyskinesia.

The severity of hyperkinetic disorders is assessed by all clinicians when they examine a patient. Quantifying the severity also provides a means of studying the natural history of a given disorder and the possible effect of new therapeutic interventions. In recent decades, a number of mechanical and electronic devices, including accelerometers, have been developed jointly by physicians and engineers to measure involuntary movements; more recently, computerized devices have also been designed [60]. The advantages of mechanical and electronic measurements are objectivity and consistency even when they are performed by different clinicians. However, as these measurements do not appear to be as sensitive as clinical measurements, hyperkinetic disorders continue to be assessed largely by clinical methods. This means that good rating instruments are required in both everyday practice and experimental settings. Unfortunately, the clinical evaluation of these disorders is complicated by the inherent nature and variability over time of involuntary movements. A number of scales have been proposed over the years to study the various hyperkinetic disorders. The aim of this review is to systematically identify all the clinical scales that have been proposed and to classify them according to the criteria developed by the Movement Disorder Society (MDS) task force on rating scales for Parkinson's Disease (PD) [65]. The systematic review of the PD rating scales carried out by this task force was conducted according to an established methodology [30]. This process includes scale identification, selection and appraisal strategies, using terminology and definitions developed ad hoc [30]. On the basis of this methodology, a scale is defined as 'Recommended' if it has been applied to that specific disease population, if there are data on its use in studies other than those collected by the group that developed the scale, and if it has been studied clinimetrically and found to be valid, reliable and sensitive to change. A scale is defined as 'Suggested' if it has been applied to specific populations, but only one of the other criteria applies. A scale is defined simply as 'Listed' if it satisfies only one of the three criteria used to define 'Recommended' scales. Owing to the relative lack of proven treatments for hyperkinetic disorders, the clinimetric criterion for rating dyskinesia scales does not categorically require responsiveness to be

established. Indeed, if a scale fulfills the requirements of reliability and validity, the criterion is considered to be satisfied, although the absence of responsiveness is noted as a weakness of that scale. This classification has been successfully used to assess the validity of the scales used for both motor and non-motor aspects of PD [16].

This review will follow the same pattern for each one of the conditions studied. Only published or in press peer-reviewed papers or published abstracts from main neurological meetings were evaluated. The Medline database on PubMed was searched for relevant papers and all the scales used to measure a given disorder were identified (as of Medline last accessed on the 10th of November 2014) using the following query: "Hyperkinetic disorders" and 'Assessment' or 'Scales' or 'Questionnaires'. For each scale, a search was conducted for the following terms 'Tremor' and 'Assessment' or 'Scales' or 'Questionnaires', 'Dystonia' and 'Assessment' or 'Scales' or 'Questionnaires', 'Chorea' and 'Assessment' or 'Scales' or 'Questionnaires', 'Tics' and 'Assessment' or 'Scales' or 'Questionnaires', 'Myoclonus' and 'assessment' or 'Scales' or 'Questionnaires', 'Drug-induced dyskinesias' and 'Assessment' or 'Scales' or 'Questionnaires'. In addition for each scale, a search was conducted for the terms 'Tremor', 'Dystonia', 'Chorea', 'Tics', 'Myoclonus', 'Drug-induced dyskinesias' and the name of the scale. All scales have been reported in a specific table, though only those defined as 'Recommended' according to the aforementioned criteria will be appraised and discussed in detail in the following text.

## 2. Scales for hyperkinetic disorders

### 2.1. Tremor

Twelve scales have been assessed for tremor evaluation ([3,4,5,23, 25,41,43,44,54,71,101,116] (Table 1), but only two of them, the Fahn–Tolosa–Marin Tremor Rating Scale (FTM–TRS) and the Washington Heights–Inwood Genetic Study of Essential Tremor (WHIGET), reached recommendation status (Table 2).

#### 2.1.1. Fahn–Tolosa–Marin Tremor Rating Scale

**2.1.1.1. Scale description.** The Tremor Rating Scale (TRS) was developed by Fahn, Tolosa and Marin to quantify rest, postural and action/intention tremors [23]. This scale also evaluates voice tremor as well as handwriting and other tasks, such as hygiene and dressing, assessing the impact of tremor on patients' daily life. The TRS is divided into three

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