

Fahn–Marsden Rating Scale

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Glossary

Dystonia – Sustained muscle contractions of agonists and antagonists causing involuntary twisting movements and postures.

Reliability – Consistency of the assessment or measurement of a given tool (e.g., a scale), at different times or by different evaluators.

Validity – Degree to which a test (e.g., a scale) measures what it is intended.

Definition and History

Dystonia is a hyperkinetic movement disorder characterized by sustained twisting and posturing movements, which may affect single or multiple body areas. While there are characteristic electrophysiologic features of dystonia, such as muscle agonist–antagonist cocontraction, these do not yet allow quantification of severity of dystonia in different body sites. Further, the features of dystonia are not static and vary according to activity. In order to better characterize the clinical course, and to assess the efficacy of treatments, scales to capture both the severity of the dystonia (including the dynamic features) and the functional disability of the dystonia are needed. Scales may encompass dystonia affecting all body sites, such as the Fahn–Marsden (F–M) rating scale, which may be used for generalized, segmental, or focal dystonia, or may be focused on one body area, such as cervical dystonia.

Both the structure and validity and reliability of the F–M rating scale were first reported in 1985. The scale was subsequently first used in the context of a therapeutic trial with trihexyphenidyl for the treatment of torsion dystonia.

The F–M rating scale is composed of two sections: a movement scale based on the neurological examination and a disability scale based on the patient's opinion of his disability in activities of daily living. The movement scale is further divided in nine body regions: eyes, mouth, speech and swallowing, neck, right arm, left arm, trunk, right leg, and left leg. Individual scores are obtained for each body region; from 0 to 8 for the eyes, mouth, and neck, and from 0 to 16 for the other body parts. The individual scores are calculated for each body region

using a formula that takes a provoking factor, a severity factor and a weighting factor into account. The eyes, mouth, and neck are 'down-weighted' because when involved, these regions were not suggested to cause as much disability. The provoking factor is scored from 0 to 4 as following: 0 – No dystonia, 1 – Dystonia on particular action, 2 – Dystonia on many actions, 3 – Dystonia on action of a distant body part (overflow) or intermittently at rest, and 4 – Dystonia at rest. For speech and swallowing, the provoking factor is slightly different, and is based on frequency. The severity factors are scored similarly for all regions except speech and swallowing. Some site-specific criteria apply according to the specific abnormal movement or posture caused by the dystonia (e.g, bending of the trunk with truncal dystonia, blinking or spasms of eye closure with blepharospasm). In general, the severity factors range from 0 to 4, with 0 – No dystonia, 1 – Slight dystonia, 2 – Mild dystonia, 3 – Moderate dystonia, and 4 – Severe dystonia, with different ratings for speech and swallowing. Once individual scores for each body part are calculated, they are summed to obtain the movement scale score. The scale ranges from minimum of 0 to maximum of 120.

For the disability score, seven activities of daily living are rated according to the patient's perception of his/her disability: speech, writing, feeding, eating, hygiene, dressing, and walking. The scores are from 0 to 4 (except for walking, from 0 to 6), and scores are task specific. In general, scores are determined according to the following scale: 0 – Normal, 1 – Slight difficulty, 2 – Some difficulty, 3 – Marked difficulty, and 4 – Severe difficulty, unable to perform the activity. The scale for walking is slightly different, ranging from 0 to 6; with a score of 6 assigned if the subject is wheelchair bound. Once the scores for all the individual activities of daily living are obtained, they are summed to obtain the total disability score, which ranges from 0 to 30.

Metrics of the Scale: Reliability and Validity

The validity of the scale assesses how well the scale reflects a gold standard rating. As clinical assessment of dystonia remains the de facto 'gold standard,' the validity of the motor portion of the scale reflects how well the scores correlate with the clinical impression of dystonia

severity. For example, when the dystonia is perceived as severe by an experienced neurologist, a valid scale would lead to a high score. The converse is true for a dystonia perceived as mild by the clinician. On the other hand, the reliability of the scale corresponds to how reproducible the score is, among different examiners (interrater reliability), but also from the same examiner at different times (intrarater reliability).

p0030 The validity and reliability of the F–M rating scale were first evaluated by Burke and colleagues in 1985. The validity was evaluated by comparing the F–M score with the global clinical impression of severity and with the disability score. The reliability was assessed by first, comparing evaluations of the 10 patients on two occasions by two examiners (intrarater validity) and second, by examining the correlation of the ten evaluations of the three examiners. The scale was shown to be both reasonably valid and reliable for patients with primary torsion dystonia. However, only a small number of patients were assessed, and because dystonia is so heterogeneous, the broader spectrum of dystonia was not tested. Further, the degree of agreement for individual body sites was not reported.

p0035 Two large multicenter studies have since evaluated the F–M scale. Comella and colleagues evaluated the reliability of the scale using 20 videotaped patients, and 25 dystonia experts, and compared the scale to the UDRS (Unified Dystonia Rating Scale) and GDS (Global Dystonia Rating Scale). They concluded that all three scales were internally consistent, showed good to excellent interrater reliability (intraclass correlation coefficient, 0.71–0.78), but that the provoking factor in the F–M demonstrated less interrater agreement than the motor severity ratings. Further, they found that the GDS (which includes rating for proximal and distal limbs separately, and does not include subjective speech and swallowing ratings, and does not include weighting factors) was easier to administer than the F–M. Krystkowiak and colleagues further demonstrated reliability in a prospective assessment of the F–M scale used to track severity and disability of primary generalized dystonia treated with deep brain stimulation.

s0015 **Applications of the Scale**

p0040 The F–M scale was first used to assess the efficacy of trihexyphenidyl as a treatment for primary and secondary dystonia. It is now primarily used to assess the efficacy of deep brain stimulation surgery in both primary and secondary dystonia, usually in cases with generalized dystonia.

Problems with the Scale

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As noted, the F–M rating scale has several limitations. The major drawback in using the F–M scale is lack of ease in administration, which relates to separately determining provoking and severity factors. The GDS was developed as an alternative to the F–M, and has been shown to be at least as reliable and valid but simpler and easier to apply. The UDRS was developed to address the F–M scale's limitations regarding the flexibility to report specific sites as the rated areas are smaller and more defined, and shows great promise. The F–M scale is also limited in its applicability for some dystonia plus syndromes and secondary dystonias as it is focused on dystonia, and additional movement disorders, such as myoclonus and features of secondary dystonia, such as that due to Wilson disease, or spasticity associated with other forms of secondary dystonia are not captured. In these situations, disease specific scales, such as the Wilson Disease Scale, may be more applicable.

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See also: Basal Ganglia (00015); Animal Models for Dystonia (00095); Dystonia (00104); DYT 1 (00109); DYT11, DYT15 – Myoclonus-Dystonia (00111); Dyt12 – Rapid Onset Dystonia Parkinsonism (00112); DYT2-Autosomal Recessive Generalized Dystonia (00114); Dyt4 Autosomal Dominant Type Dystonia or Whispering Dysphonia (00116); DYT 5 (00117); Hypnic Jerks (00126); Dystonia in Amish-Mennonite and Mennonite Families (00128); Generalized Primary Torsion Dystonia (00134); Wilsons Disease (00430).

Further Reading

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Relevant Websites

www.wemove.org – Worldwide Education and Awareness for Movement Disorders

MOVE: 00099

Non-Print Items

Abstract:

The Fahn–Marsden scale (F–M, also known as the Burke–Fahn–Marsden scale, BFM) is a reliable and valid scale used to quantify dystonia symptoms and signs. It is composed of two sections: a movement scale and a disability scale. It has been used in multiple studies to quantify the efficacy of treatments for dystonia, most frequently deep brain stimulation studies.

Keywords: Assessment; Burke–Fahn–Marsden; Dystonia; Rating scale

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