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ORIGINAL ARTICLE

Validation of the Hemifacial Spasm Grading Scale: a clinical tool for hemifacial spasm

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Abstract

Background To create an objective rating tool for hemifacial spasm (HFS) and validate it on a cohort of patients.

Methods A panel of movement disorders specialists elaborated, through the Delphi method, the Hemifacial Spasm Grading Scale (HSGS). The validity of the scale was tested in a longitudinal, prospective observational study, with standardized video recording protocol before and after botulinum neurotoxin (BoNT) treatment. The video recordings obtained from each patient were then independently assessed with HSGS by three blinded raters. The scale was compared to patient-reported HFS-7 scale and to the clinical grading of spasm intensity scale.

Results Intra-rater reproducibility ranged between ICC 0.73 (95% CI = 0.54–0.86) and 0.83 (0.68–0.92) and inter-rater reproducibility between 0.62 (95% CI = 0.44–0.77) and 0.82 (0.69–0.90). HSGS scores correlated with clinical grading of spasm intensity scale scores, but not with HFS-7. HSGS confirmed BoNT efficacy, with scores lowering at 1 month from treatment.

Conclusions HSGS represents an objective, quick and reliable scale for the assessment of HFS, and might be useful to monitor BoNT treatment efficacy over time.

Keywords Hemifacial spasm · Scale · Botulinum toxin · HSGS

Introduction

Hemifacial spasm (HFS) is characterized by unilateral, tonic, and clonic contractions of the muscles innervated by the ipsilateral facial nerve [1, 2]. HFS is an uncommon neurological condition, affecting 10 in 100.000 people [3, 4] and is frequently due to neurovascular facial nerve compression [5, 6]. Despite being considered a benign condition, HFS represents a chronic, progressive, and potentially disabling disorder if not opportunely treated. To date, together with microvascular decompression surgery, botulinum neurotoxin (BoNT) is considered a first-line effective and safe treatment [7–9]. Over the past 20 years, a number of assessing tools were adopted in several studies on BoNT treatment [10–14]. However, poor reproducibility and lack of standardized grading of clinical symptoms limited the clinical application of HFS scales [10, 13, 15–17]. Such limitation highly affected the comparison between trials evaluating BoNT effect on HFS. To date, no scale has been proven to be useful as a standardized evaluation of HFS [10]. In particular, existing scales are flawed by poor reproducibility, lack of objective assessment or video recordings, poor standardization of assessment procedure, no clear definition of frequency and severity of HFS, or enrollment in the validation study of patients with HFS and blepharospasm as well [10]. Thus, an objective tool to assess symptoms and treatment efficacy is an unmet need for patients with HFS. The aim of this study was to create an objective rating tool for HFS, validating it on an outpatient cohort of patients evaluated before and after BoNT treatment.

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Methods

Construction of the Hemifacial Spasm Grading Scale

Core principles

First, a group of five movement disorder neurologists with experience in BoNT treatment defined, through the Delphi method, the main items crucial to evaluate HFS. The working group defined the HFS Grading Scale items by executing a structured consensus-driven Delphi method, with iterative surveys and meetings to develop a proposal based on each expert opinion. The group convened nine times from January 2015 to July 2015. Each meeting was followed by formal decision for each proposal, with decision-making using a 3/5 majority vote for a final decision on measures to be included in the scale. Unanimous agreement was first reached on the three main virtues of the scale: (i) accuracy in assessing the severity of the disturbance; (ii) quick administration in a clinical routine setting; (iii) easiness of use. Then, a list of phenomenological aspects of HFS severity was drawn. Clinical features were grouped into three main features better describing involuntary muscular contractions: localization, intensity, and frequency. Four independent drafts of the scale were created by each member of the group, compared, and merged together until a final unanimous agreement was reached.

Scale construction

In the resulting version of the scale, each of the three features (localization, intensity, and frequency) was studied in depth, in order to properly grade different degrees of severity. Concerning localization, consensus was achieved assigning higher scores for severity to HFS spreading to both upper and lower-half face muscles. On the contrary, lower scores were attributed to HFS forms confined to the upper-half face muscles, such as periorbital muscles (e.g., orbicularis oculi), or to the lower face muscles, even though very uncommon [1]. Regarding intensity, greater score was assigned to sub-continuous/continuous muscular contractions, leading to eyelid closure, while lower score was given to rare or singular contractions during clinical assessment. Concerning frequency, the highest score was assigned to spontaneous HFS visible for > 50% of the assessment period, median score to spontaneous HFS with presentation < 50% of the assessment period, while the lowest score was attributed for HFS presenting only when elicited by a motor stimulus. Several scores for each single item were proposed, and strengths and weaknesses of each were discussed and singularly evaluated. After a unanimous agreement was reached, the final version of HFS Grading Scale (HSGS) was approved (Table 1). The Delphi method led to a unanimously accepted version of the HSGS scale composed of three core features: localization, intensity, and frequency. Localization limited to the upper or lower face muscles, intensity confined to single jerks, and frequency as low as only elicited by muscle activation all received the minimum score of 1. A score of 2 was assigned in case of localization to both the upper and lower face muscles, or intensity characterized by sub-continuous jerks (spasm), while high frequency with spontaneous contraction was graded 3 or 5 if present in respectively less and more than 50% of the assessment time. Total score was calculated by adding single-item scores.

Table 1 The Hemifacial Spasm Grading Scale (HSGS)

Hemifacial spasm	Yes–no
Localization	
- Isolated upper face (e.g., orbicularis oculi)/lower face muscles	1
- Involvement of both the upper and lower face muscles	2
Intensity	
- Single jerks	1
- Sub-continuous jerks (spasm)	2
Frequency	
- Muscular contractions provoked by motor activation	1
- Spontaneous contractions	
< 50% the time	3
> 50% the time	5
Total	____/9

Standard protocol approvals, registrations, and patient consents

To validate the HSGS and verify its clinometric properties on a sample of patients suffering from HFS, an observational longitudinal cohort study was designed and approved by the local ethics advisory committee (CEAS Umbria, n. 2771/16). Written informed consent to participate and authorization for disclosure of videos were obtained from all participants.

Enrolment of the study population

Inclusion criteria were (i) age > 18 years, (ii) clinical diagnosis of HFS formulated by a neurologist with experience in movement disorders, and (iii) HFS present for at least 3 months before HSGS assessment. Patients with severe sensory and/or cognitive deficits were excluded from the study. Overall, 35 consecutive subjects affected by HFS and receiving BoNT treatment at the Movement Disorders Outpatient Clinic of the Neurology Department were enrolled in the study. For every participant, demographic (age, gender), clinical (disease side and duration, duration of treatment, number of treatments, history of neurological disease, chronic illness, current

medication use, and QoL related to HFS by means of HFS7 [16]), and neuroimaging data were collected.

Longitudinal assessment, video recording, and blind assessment

Specific assessment of HFS severity in patients undergoing BoNT treatment was performed according to a standardized procedure. A 90-s video recording was taken for each patient at baseline (T0) and 1 month after treatment (T1) to evaluate peak improvement and possible side effects. Video recording followed a standardized procedure: patients were asked to stay seated on a chair in a closed, quiet room, in front of the camera; during the recording, they were asked to be silent, at rest, with eyes opened for 30 s, then to talk freely, answering simple questions (i.e., date and place of birth, actual and previous occupation) to evaluate the appearance of HFS after slight physiological stimuli; finally, patients were asked to repeatedly squeeze both eyes while grinding their teeth for three times, to assess HFS after maximal motor stimuli. Collected video recordings were then randomly and anonymously assigned to at least three neurologist specialists in movement disorders and with experience in HFS BoNT treatment. HFS severity was assessed on each video by means of the HSGS and clinical grading of spasm intensity scale [12], an objective assessment tool that has been used to monitor BoNT effect [12]. HSGS scores were compared to clinical grading of spasm intensity scale scores both at baseline and after treatment, to assess correlation before and after the use of BoNT and to define the efficacy of the treatment itself [12].

Validation process and assessment of reliability

HSGS reliability was determined by inter-rater and intra-rater administration. For inter-rater reliability, all 70 video recordings (35 performed at T0 and 35 at T1) were randomly and anonymously submitted to three raters (movement disorders neurologists with experience in BoNT treatment blinded to patients' demographic, neuroradiological, and clinical data beyond those derived from the video), who independently filled in the HSGS for each video. For intra-rater reliability, 2 months after the first viewing, one of the three raters reviewed all 70 video recordings, re-administering the HSGS. The raters also performed on each recording the clinical grading of spasm intensity [12]. Scores obtained from clinical grading of spasm intensity were then related to HSGS scores.

Statistical analysis

The statistical analysis was performed with the R software v3.0. Sample descriptive statistics were calculated. Continuous variables were described with median and interquartile range while categorical variables with frequencies and percentages. For all items (localization, intensity. and frequency) and for the total score, the hypothesis of inter-rater and intra-rater agreement through intra-class correlation coefficients (ICC) was verified. Correlation between HSGS and clinical grading of spasm intensity scale [12] was measured by means of Spearman correlation coefficients. Reduction of single-item and total HSGS scores at peak effect of toxin treatment (i.e., 1 month after injection) compared with baseline has been tested by means of the Mann–Whitney U test. In all analyses, a significant level of 5% was considered.

Data availability Anonymized data will be shared by request from any qualified investigator. The video will be made available by request from any qualified investigator if approved by our Research Ethics Board.

Results

Thirty-six patients have been consecutively enrolled in our study (18 males and 18 females) (Table 2).

One patient retrieved consent and was excluded. Overall, 35 patients received HFS diagnosis and were enrolled and have completed the study. Mean age was 66.0 years, mean disease duration was 10 years (5.0–17.5), and mean BoNT treatment duration was 7 years (3.0–13.0). The left side was affected in 21 patients and one patient had a bilateral presentation. Neurovascular conflict was found in 10 of 21 undergoing brain magnetic resonance imaging. Five patients did not undergo any neuroradiologic exam at the time of enrolment. Incobotulinum toxin A was used for all treatments, with a mean dose of 11.0 UI distributed on six injection sites. We collected standardized 90-s video recording at baseline and at follow-up, all assessed by independent blind raters through HSGS. All patients receiving the lower score on localization item had isolated periorbital muscle involvement. HSGS

Table 2 Demographic and clinical characteristics. Median (Q1–Q3) or *n* (%)

Variable (*n* = 35)	Median (IQR)
Age (years)	66.0 (59.0–75.0)
Gender (M)	18/35 (51.4)
Disease duration (years)	10.0 (5.0–17.5)
BoNT dose per treatment (UI)	11.0 (5.0–17.5)
Treatment duration (years)	7.0 (3.0–13.0)
Injection sites number	6.0 (4.0–7.5)
Neurovascular conflict (yes)	10/27 (37%)
Right side	13/35 (37.1%)
Left side	21/35 (60.0%)
Bilateral	1/35 (2.9%)

demonstrated good reproducibility between raters, with ICC ranging from 0.73 (95% CI = 0.54–0.86), for the item "frequency" at T0, to 0.83 (0.68–0.92), for the item "frequency" at T1 (Table 3). Inter-rater reproducibility was satisfactory as well (HSGS total score ICC 0.74 at T0, 0.77 at T1) (Table 3).

Even though not validated [12], further comparison with a HFS scale confirmed significant correlation between the ratings of the two scales. In particular, for each rater, the HSGS overall scores consistently correlated with the score of the clinical grading of spasm intensity [12] (Table 4). No correlation was found between HSGS and HFS-7 [16] overall score, and HSGS also for BoNT efficacy. Indeed, HFS was also scored with HSGS after 1 month from BoNT treatment, demonstrating a significant reduction of single-item (localization $p = 0.024$, intensity $p = 0.049$, frequency $p = 0.003$) and overall HSGS scores ($p = 0.002$) compared with baseline (Table 3).

Table 4 Correlation between HSGS and clinical grading of spasm intensity [12] (Spearman)

Rater #	Item	Clinical grading of spasm intensity [12]	*p* value
1	HSGS-T0	0.79	< 0.001
	HSGS-T1	0.83	< 0.001
2	HSGS-T0	0.61	< 0.001
	HSGS-T1	0.77	< 0.001
3	HSGS-T0	0.84	< 0.001
	HSGS-T1	0.61	< 0.001

Discussion

Despite being frequent among movement disorders centers, HFS lacks the standardized assessment tools reserved for other diseases such as Parkinson's disease. To date, although several rating scales have been used in clinical trials addressing BoNT treatment efficacy, no validated tool has been provided for the assessment of HFS, which is often evaluated with incomplete scales reserved for blepharospasm [10]. Differently, the SMC grade, derived from surgical studies, is a validated clinical scale [18]. However, this tool, designed to evaluate patients who are candidates for surgery, is not able to grade mild and moderate condition, such as the increased blinking rate caused, for example, by external stimuli or primary localization to the lower face muscles. The differences of clinical objective variations even in mild forms and the possibility of localization of involuntary muscular contractions either in the upper (e.g., orbicularis oculi) or in the lower face muscles are important to optimize the BoNT treatment.

The lack of a standardized questionnaire, mostly due to the complexity in assessing a highly heterogeneous and episodic in nature movement disorder, leads to unsatisfactory follow-up and BoNT planning. Patient reporting, despite being crucial to define outcome, is influenced by several factors, including socioeconomic status, gender, marital status, and acquired information on their disease [10, 19]. Thus, a standardized reliable, objective, and simple tool to assess HFS might be of great help for clinicians in both detecting HFS at baseline and evaluating BoNT efficacy at follow-up. With this study,

Table 3 Intra-rater and inter-rater agreement and follow-up vs baseline comparison

Time	Variable	Intra-rater*,#	Inter-rater*,#	Baseline vs follow-up§
T0	HFS—Localization	0.74	0.55	1.89 ± 0.32
		(0.54–0.86)	(0.36–0.72)	2 (1–2)
	HFS—intensity	0.73	0.62	1.58 ± 0.51
		(0.54–0.86)	(0.44–0.77)	2 (1–2)
	HFS—frequency	0.76	0.74	3.63 ± 1.64
		(0.58–0.87)	(0.60–0.85)	5 (1–5)
	HFS—total	0.81	0.74	7.11 ± 2.13
		(0.66–0.90)	(0.60–0.85)	8 (3–9)
T1	HFS—localization	0.78	0.68	1.42 ± 0.77
		(0.59–0.89)	(0.50–0.82)	2 (0–2)
	HFS—intensity	0.74	0.49	1.16 ± 0.69
		(0.52–0.86)	(0.26–0.69)	1 (0–2)
	HFS—frequency	0.83	0.82	2.00 ± 1.56
		(0.68–0.92)	(0.69–0.90)	1 (0–5)
	HFS—total	0.82	0.77	4.58 ± 2.36
		(0.65–0.91)	(0.62–0.88)	5 (0–8)

*ICC (95% CI); # all *p* values were < 0.001; § Mean ± SD median (min–max)

T0, baseline; *T1*, follow-up visit

we provide an objective scale for HFS, constructed according to the Delphi method by movement disorders specialists, and validated with standardized procedures in an appropriate cohort of patients suffering from HFS. The HSGS, whose grading performance has been further confirmed through the comparison with the clinical grading of spasm intensity scale [12], represents a quick and reliable tool to assess clinical features of HFS. Indeed, even though patient-reported symptoms and quality of life remain crucial, clinical assessment is mandatory to verify treatment response and HFS evolution over time. Accordingly, the HSGS has been developed to be intuitive for clinicians, who have to take care of only three main items to be graded: localization, intensity, and frequency of the HFS during each visit. We have chosen a simple and intuitive definition of the extension of the disease, assigned a specific score according to muscle group involvement, distinguished between isolated or combined periorbital muscles and lower-half face muscles, intensity of muscle contraction, and discerned single-muscle contractions from repetitive/subcontinuous contractions, and frequency of presentation of the clinical manifestation.

The present study does not have the aim to use the HSGS score as assessment of outcomes of BoNT treatment. Further studies are needed to explore this possibility. However, in our opinion, also a patient showing single contractions, isolated to the upper or lower face muscles, that are visible only after motor activation (HSGS lower score of 3), with a slight disability in social or functional contexts (HFS-7 grade 1), may benefit from BoNT treatment. Indeed, both single-item and total HSGS mean scores were significantly reduced after BoNT treatment compared with baseline. Thus, HSGS might be considered for further refinement and use in trial setting, where BoNT efficacy will eventually need to be certified.

Interestingly, no correlation was observed between HSGS and HFS-7 scores. Such findings, considering that HFS-7 is a subjective evaluation tool based on patient reporting, might suggest that the two paradigms are pointing in different directions. The HSGS, strictly clinical, might give objective data for clinical use and BoNT treatment scheduling, while HFS-7 might point to quality of life as the main issue. However, as all scales depend on patient self-reporting, also HFS-7 might be flawed by the emotional and physical status of the patient in a certain moment, by patient gender, by socioeconomic/sociodemographic status, and by depressive symptoms [17, 18].

Limitations to this study can be found in the small cohort recruited, all having an idiopathic form of HFS. However, since consecutive recruitment, strict follow-up, and standardized blinded rating of video recordings were performed, such factors might have poor impact on an objective scale of HFS grading. Meantime, correlation with other patient-reported scales will refine the complexity between subjective and objective measures among HFS patients. Moreover, no differences in the long-term effect of BoNT treatment using the HSGS scale were observed. This may be due to the low number of patients, which was sufficient for the validation process, but not enough to highlight outcome differences. In conclusion, our study provides a simple, quick, and reproducible assessment scale for HFS, allowing clinicians to better evaluate disease course and possibly monitor BoNT efficacy, to better tailor treatment to patients.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

Ethical standards The study protocol was approved by the Ethics Committee (CEAS, Umbria) in accordance with the principles stated in the Declaration of Helsinki. Written informed consent was obtained from all the participants.

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