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Can the Jankovic-assessment be used as an alternative to electromyography? A cross-sectional study on facial dystonia patients treated with Botulinum toxin

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ABSTRACT

Purpose: This study aims to quantitatively compare the Jankovic assessment (JA) with electromyography (EMG)-based measures for assessing changes in facial movements in patients with facial dystonia.
Materials and methods: Thirteen patients (five males and eight females) affected with different forms of facial dystonia (hemifacial spasm and synkinesis) participated in this study. All patients were treated with Botulinum Toxin (BTX) and evaluated with the JA scale and EMG-based measures, including motor unit potentials (MUP) latency and presence of polyphasic potentials before and after BTX injection. Correlation between the JA scores and the EMG-based measures was calculated. Statistical analysis was performed with the Pearson test.
Results: Correlation between the JA scores and the EMG-based measures was found to be statistically significant, both before and after treatment with BTX.
Conclusion and relevance: JA scores significantly correlated with more objective EMG-based measures, suggesting that the JA scale can be used to assess facial movement changes, for example elicited by a treatment such as BTX injection. Thus, in facial dystonia patients, the JA scale may be used for evaluating treatment outcomes as a valid and low-cost alternative to EMG.

1. Introduction

In clinical practice, clinical scales are typically used for evaluating facial movement impairments such as those due to facial palsy [1,2] or facial synkinesis [3]. For example, the Jancovic assessment (JA) measures the severity of hemifacial spasm/synkinesis [4]. These clinical assessments are cheap and easy to perform but suffer from poor accuracy and high inter-subject variability [5]. To overcome these limitations, several computer-based methods, such as for example eFACE [6], have been proposed and shown to be more accurate than clinical scales [6–8], but they require specialized equipment [6] and suffer from a relatively high learning training curve [7]. Needle EMG can also be used to accurately evaluate facial movements [9,10]. Its sensitivity and specificity for evaluating muscular function has been confirmed by several studies [10–13]. In assessments of facial palsy, EMG is considered to be superior to electroneurography (ENG) because it can reliably identify nerve damage even 1 month after the facial palsy onset (differently than ENG which is accurate only when used within 20 days from facial palsy onset) [11,12]. Furthermore, EMG can reliably and accurately evaluate impairments in muscular movements in acute facial palsy (the first 15 days after FP onset) as it has been recently shown [13]. However, needle EMG is used only rarely due to its invasiveness [10]. Methods based on combinations of clinical scales with EMG have also been proposed; for example, Monini et al. combined the House Brackman scale with EMG for facial paralysis assessment [14], but ultimately in clinical practice, despite their limitations, clinical assessments remain the gold standard methods for evaluating facial movements [1–4].

How clinical scales for facial movements assessment compare to more objective methods, such as those based on EMG recordings, has been little investigated [15]. Thus, this study aims to compare the JA scale [4] with EMG-based measures for the purpose of assessing facial

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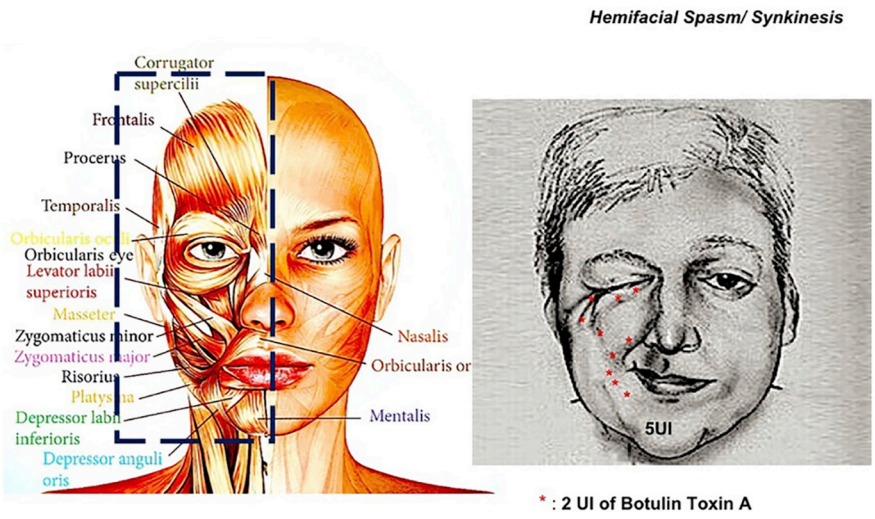

Fig. 1. The left panel shows facial muscles. In the right panel, the red cross shows the BTX injection point, while '5UI' shows the BTX injection point in the mentalis muscle which was treated with more units of BTX. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

movement impairments in patients with facial dystonia, and specifically synkinesis, blepharospasm and hemifacial spasm, treated with Botulinum Toxin (BTX).

2. Materials and methods

This study was carried out in the Department of Otolaryngology of Silvestrini University Hospital, a tertiary referral center, from January 2017 to September 2018. All study procedures were approved by the Internal Revision Board (IRB) of the hospital and conducted in accordance with the ethical principles of the Declaration of Helsinki and the national regulations for non-surgical studies (note, no identification number is released for this type of studies).

Thirteen subjects (five males and eight females, average age 52.1 years (CI 95%: 30–75; SD: 13.5)) affected with different forms of facial dystonia (hemifacial spasm and synkinesis) were enrolled in this study. All subjects signed a written, informed consent. None had been previously treated with BTX. All subjects were treated with a BTX A (Allergan Botox A, www.allergan.com) injection, which was administered by the same physician, a facial plastic surgeon with > 10 years of experience (Figs. 1-3).

For each subject the following data was collected: age, sex, disease (hemifacial spam or synkinesis), origin of the disease, administered units of BTX for dystonia to be resolved, time to achieve a BTX effect and side effects. Furthermore, patients were evaluated before BTX injection and 1 month (4 weeks) after BTX injection using the JA scale [4] and EMG as detailed below.

The JA scores were as follows: 0 = no spam or synkinesis; 1 = Minimal, increased blinking or spasm only with external stimuli. Sporadic episodes of spasm; 2 = Mild, but spontaneous eyelid fluttering/spontaneous facial movement (without spasm) definitely noticeable and possible embarrassing for the patient but with no functional disability. Duration of fluttering < 1 s; 3 = Moderate, very noticeable spasm, mild movement impairment. Spasm of duration longer than 1 s, but eye open/ face relaxed > 50% of waking time as self-reported; and 4 = Severe, incapacitating spasm. Closed eye (blepharospasm) and hemifacial spasm > 50% of waking time as self-reported.

Needle EMG was recorded from a minimum of twenty pairs of muscle fibers from the *orbicularis oculi* (*OOculi*) and *orbicularis oris muscles* (*OOris*) as in [16,17] using Surpass EMG/EP/IOM equipment (www.emsbiomed.com) in manual modality. Motor unit potentials (MUPs) waves' morphology (presence or absence of polyphasic potentials), amplitude and latency were collected and analyzed by the same neurophysiologist with > 10 years of experience. The presence of polyphasic MUPs was defined "pathological" if we found > 20% of polyphasic potentials MUPs during the recording.

To prepare the data for statistical analysis, the EMG measures were classified as follows. EMG recorded pre-treatment was classified as 0 or 1 (normal MUP wave morphology or presence of polyphasic potentials

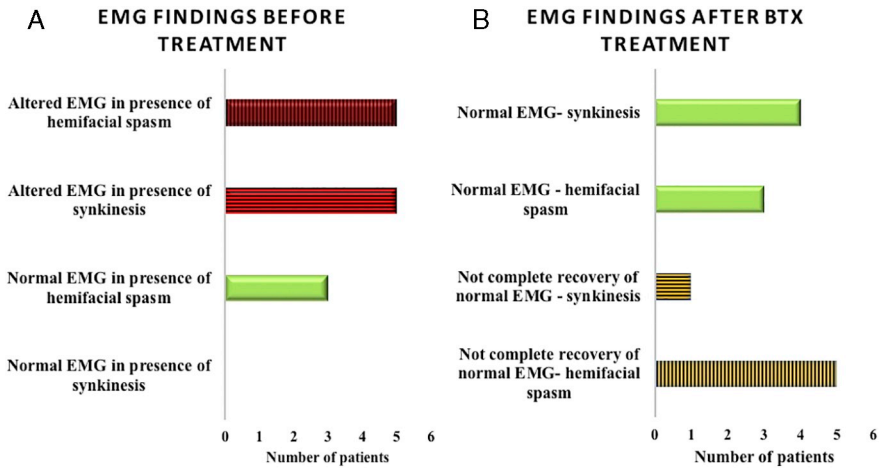

Fig. 2. A) The figure summarizes the signal features observed in patients' EMG recorded prior to BTX treatment. B) The figure summarizes the signal features observed in patients' EMG recorded one month after BXT treatment.

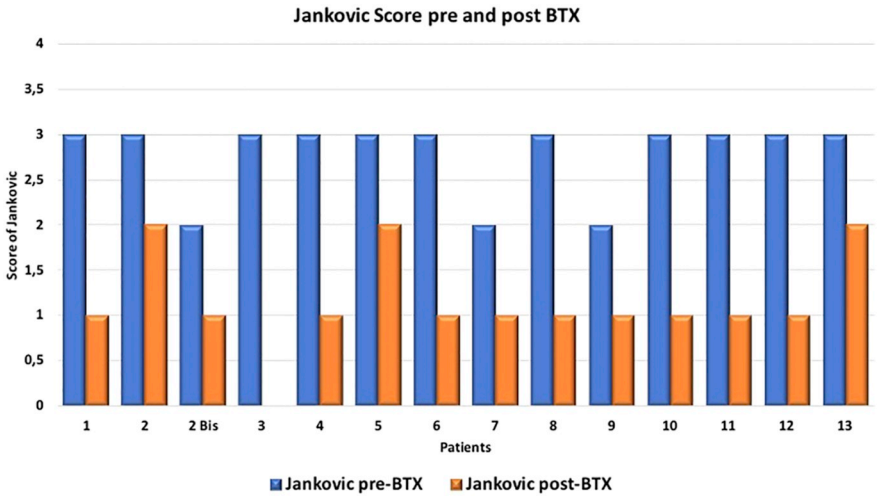

Fig. 3. The bar graphs show the pre-treatment and post-treatment JA scores for each patient. "2 bis" refers to the patient who underwent a second BTX injection in the orbicularis oculi muscle due to the persistence of spams.

over 20%, respectively). EMG recorded post treatment was classified as 0, 1, or 2 (normal MUP wave morphology or reduction of the number of polyphasic potentials > 60%; reduction of the number of polyphasic potentials < 60%; or no change compared to pre-treatment baseline, respectively).

Statistical analysis was performed using STATA®. A two-tailed *t*-test was used to evaluate the difference between pre-treatment and post-treatment JA scores. Analysis of JA scores was performed both including all patients (hemifacial spasm, synkinesis) and separately for patients' subgroups (e.g., hemifacial spasm patients only). A Chi-Square test was used to evaluate the change in the EMG measures that occurred after the BTX injection compared to pre-treatment. JA scores and EMG measures were correlated using a Pearson test both before and after BTX treatment. For all tests the level of significance was set to 0.05.

3. Results

Eight patients presented with hemifacial spasm. Among them, two patients had a progressive disease, while in six patients a hemifacial spasm affecting all hemiface muscles with the same severity had presented suddenly. Five patients suffered from unilateral facial synkinesis developed after recovery from facial palsy (Table 1).

Table 1
The table summarizes the patients' demographics, patients' symptoms, and ethiopatogenesis. Note, hemifacial spasm labeled as "P" indicates a progressive form of hemifacial spasm.

Patient number	Age	Sex	Ethiopatogenesis	Symptomatology
1	54	*M*	Post -surgical facial palsy (VIII nerve schwannoma)	Synkinesis
2	60	*F*	Post recovery facial palsy due to Herpes oticus	Synkinesis
			2nd treatment with BTX	Persistent synkinesis
3	47	*M*	Post recovery Bell's palsy	Synkinesis
4	30	*F*	Facial palsy	Synkinesis
5	46	*M*	Neurovascular impingement	Hemifacial spasm
6	75	*M*	Neurovascular impingement	Hemifacial spasm
7	35	*F*	Facial palsy post parotidectomy	Synkinesis
8	60	*F*	Neurovascular impingement	Hemifacial spasm (P)
9	55	*M*	Unknown	Hemifacial spasm (P)
10	45	*F*	Neurovascular impingement	Hemifacial spasm
11	38	*F*	Neurovascular impingement	Hemifacial spasm
12	63	*F*	Neurovascular impingement	Hemifacial spasm
13	70	*F*	Neurovascular impingement	Hemifacial spasm

The pre-treatment average JA score across all patients was 2.78 (CI 95%: 2–3: SD: 0.42). The lowest score (score of 2) was observed only in two patients, one who suffered from hemifacial synkinesis due to recurrent episodes of facial palsy and one affected by a progressive form of hemifacial spasm. In 10 out of 13 patients we observed a pathological number of polyphasic MUPs prior to treatment. All these 10 patients displayed abnormally long MUPs latency. In the remaining three patients the MUPs displayed normal wave morphology. The three patients with normal EMG had a JA score of 2.

A single BTX injection was able to resolve the symptoms in 12 patients. One patient completely recovered only after a second BTX injection (6 IU) in the *orbicularis oculi muscle.* This particular patient suffered from a severe form of synkinesis (JA score of 3) which arose after recurrent, frequent episodes of facial palsy (3 episodes in a year). None of the patients developed any side effects, including any allergy, immediately or 7 days after the injection. 1 month after BTX treatment, JA scores significantly decreased across all patients (average JA score: 1.14; CI 95%: 0–2; SD: 0.53; *t*-test: $p < 0.00001$). Analysis of JA scores in subgroups showed a statistically significant improvement in synkinesis (*t*-test: $p = 0.01$) and hemifacial spasm (t-test: $p < 0.00001$) patients.

Changes in EMG also indicated a significant improvement (χ: $p = 0.00001$). Specifically, compared to pre-treatment baseline, seven patients (4 with synkinesis and 3 with hemifacial spasm) no longer displayed pathologic polyphasic MUPs; also, the number of polyphasic MUPs was reduced by 50% in four patients (1 with synkinesis and 3 with hemifacial spasm); by 70% in one patient with hemifacial spasm; and by 80% in one patient with hemifacial spasm. Latency became normal in 11 patients (84.6%) and remained slightly increased in 2 patients (15.4%) with hemifacial spasms (see Table 2).

JA scores and EMG measures were significantly correlated both pre-treatment (Pearson: $r = 1$; $p < 0.00001$) and post-treatment (Pearson: $r = 0.7455$; $p = 0.002884$).

4. Discussion

This study showed that the JA scores and the EMG measures showed a statistically significant change when comparing pre-treatment to post-treatment data ($p < 0.00001$ and $p = 0.00001$ for JA and EMG respectively). Furthermore, these measures were significantly correlated, both pre-treatment ($p < 0.00001$) and post-treatment ($p = 0.002884$).

BTX A has been successfully used for the treatment of muscle dystonia in different body' districts and for managing the muscle stiffness that arises from spasmodic neuronal disease [18–20]; for example, it is

Table 2
The table details the signal features observed in patients' eye and mouth MUPs recorded prior to BTX treatment.

Patient number	Pre-treatment EMG
1	Pathological polyphasic MUPs (> 20%) that had normal amplitude and prolonged latency
2	Pathological polyphasic MUPs (> 20%); all MUPs showed an increased amplitude and prolonged latency
	Eye: Polyphasic MPUs with slightly reduced amplitude. Mouth: Pathological polyphasic MPUs (> 30%) with slightly increased amplitude
3	Pathological polyphasic MUPs (> 40%) with normal amplitude and prolonged latency
4	Pathological polyphasic MUPs (> 50%) with prolonged latency and increased amplitude
5	Pathological polyphasic MUPs (30%), with increased amplitude and prolonged latency
6	Pathological polyphasic MUPs (70%), normal amplitude and prolonged latency
7	Polyphasic MUPs with slightly increased amplitude
8	Pathological polyphasic MUPs (> 20%) normal amplitude and prolonged latency
9	Polyphasic MUPs with slightly prolonged latency and increased amplitude
10	Pathological polyphasic MUPs (50%), with normal amplitude and prolonged latency
11	Pathological polyphasic MUPs (60%), with increased amplitude and prolonged latency
12	Pathological polyphasic MUPs (> 20%), with prolonged latency and increased amplitude
13	Eye: Polyphasic MUPs with slightly reduced amplitude; Mouth: Polyphasic MUPs with slightly increased amplitude

commonly injected in the head and neck areas for treating muscular facial spasms [21–25], headache [26], and dysphonia [27]. Despite being widespread and having proven to be effective in these indications [21–27], the use of BTX remains controversial [28], also due to the reported stiffness increase in muscles injected with the toxin [28]. In this study we found that all patients treated with BTX achieved complete resolution of their symptoms (hemifacial spasm or synkinesis) one month after the injection, regardless of the cause of their primary disease (facial palsy, vascular impingement, or unknown) as shown by their JA scores and EMG recordings. Also, none of the subjects reported any of the side effects described in literature [25,28]. These results support the efficacy of BTX for treating facial spasm [29,30] and synkinesis.

By inhibiting release of acetylcholine at the muscular junction [31,32], BTX causes a chemical temporary myectomy [33,34] that blocks aberrant muscular movements, which leads to resolution of muscular spasms regardless of their origin [35]. In this study, efficacy of treatment was confirmed by the decrease of the JA scores and the improvement in the EMG measures at 1 month after treatment. The changes we observed in the EMG were indicative of recovery of motor unit function. For example, the progressive reduction of polyphasic MUPs was indicative of a reduction of the excitation threshold, elicited by the block of acetylcholine re-uptake at the level of muscular junction. Such a reduction occurred regardless of the origin of the aberrant excitation, whether spontaneous as in hemifacial spams or associated to a normal facial expression (e.g., simultaneous smiling and closing of the eye) in the affected hemiface [35] as in synkinesis.

We found that a single BTX injection was generally sufficient to achieve a resolution of muscular spasms. In fact, only one patient needed a second injection (6 IU, injected into the *orbicularis oculi muscle*) to achieve complete spasm relief. This finding is consistent with the results reported by Filipo et al. who showed the effectiveness of multiple BTX injections for treating muscular spasms caused by facial synkinesis [36].

Overall, the results of this study suggest that JA scores are a valid alternative to EMG in the evaluation of outcome of BTX injections in patients with facial spasm or synkinesis. We believe that establishing the validity of methods for facial assessment, such as the JA scale, can help clinicians assess those treatments whose therapeutic effect is still being debated, such as BTX. Finally, the use of JA may allow a significant reduction of cost of neurologist visits given that a neurological consultation can cost up to 50% more when electrophysiological tests are performed [37]. While such costs may differ depending on the country, it has been shown that the minimum cost increase added by electrophysiological tests is 10% [37]. Future studies on wider samples should be performed to confirm the specificity and sensitivity of the JA scale.

4.1. Limitations of the study

The major limitation of this study is the limited sample size. Other limitations are the presence of patients suffering from two different forms of facial dystonia (hemifacial spasm and synkinesis) and the spasms' unknown origin in one patient.

5. Conclusions

This study shows that JA scores, which are subjective measures, correlate with more-quantitative EMG-based measures in the assessment of BTX treatment outcome in patients with hemifacial spam or facial synkinesis. This correlation suggests that the JA scale may be a valid measure of outcome of BTX treatments when EMG is not available.

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Declaration of competing interest

None of the authors declares conflict of interest.

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