L’INOCULO DI TOSSINA BOTULINICA

NELLE DISTONIE

(L’IMPORTANZA DEL MONITORAGGIO EMG)

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ISSUES & OPINIONS

NEEDLE EMG GUIDANCE FOR INJECTION OF BOTULINUM TOXIN

Needle EMG Guidance Is Useful

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Needle EMG Guidance Is Rarely Required

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-La sola valutazione clinica di una distonia è insufficiente a localizzare con precisione i muscoli da inoculare (soprattutto i muscoli profondi) : sensibilità solo 0.35, specificità solo 0.74 (Brans et al. Neurology, 1998)

-**L'importanza di centrare con l'inoculo la zona delle placche motorie (PM)**

a) L'inoculo di tossina nella regione delle PM produce la paresi maggiore (Shaari and Sanders,Muscle Nerve , 1993).

b) Lo spostamento di solo 0.5 cm dell'inoculo dalla zona delle PM riduce del 50% l'efficacia della BTX (Childers et al., Muscle Nerve, 1998). L'effetto dell'inoculo di BT può essere aumentato se tale inoculo viene eseguito esattamente nella zona della placca neuromuscolare (Lapakti et al., 2010).

-**Riduzione con l'EMG della diffusione ai muscoli vicini:** la diffusione dell' inoculo varia da 2.5 a 4.5 cm dal muscolo bersaglio. Diffusione= deficit di muscoli non coinvolti nell'iper-attività (specie per le distonie). In pazienti con distonia all'arto superiore la diffusione a muscoli non interessati con trattamento senza guida EMG era presente in circa il 63% dei casi ; l'effetto era una risposta sub-ottimale nel 15 % dei casi (Ross et al., 1997)

-**La tecnica della stimolazione ad ago dei fascicoli muscolari è utile nei pazienti in sedazione, nei pazienti con deficit del controllo motorio fine (ad es. nei pazienti con esiti post-stroke)**

-**Con EMG minori effetti collaterali in casi di torcicollo spasm. (disfagia) rispetto all'inoculo senza EMG**. Nelle distonie nei pazienti trattati con tecnica EMG maggiore effetto clinico rispetto a quelli trattati senza tecnica EMG (Cornella et al., Neurology 1992)

- **Il beneficio del trattamento con tecnica EMG supera nettamente gli svantaggi: il costo addizionale dell' EMG è inferiore rispetto al costo della BTX. L'esatta localizzazione della BTX porta a una minor dose consumata ed a un maggiore effetto. Minor dose di BTX =minore probabilità di sviluppare anticorpi neutralizzanti la BTX.**

EFFETTO DELLA TOSSINA BOTULINICA (BT) SUL SNP CONTROLLATO MEDIANTE ESAME EMG

- **L'ampiezza del CMAP del m. EDB inizia a ridursi a 48 ore dall'inoculo con BT. Massimo decremento del CMAP a 7-21giorni dall'inoculo. Il decremento rimane per almeno 90 gg. (Hamjian and Walker, 1994; Eleopra et al. 1998).**
- **EMG utilizzabile come test per la risposta terapeutica del paziente prima d'iniziare il trattamento con BT e/o per verificare la mancanza di risposta terapeutica da cause secondarie (Cordivari et al., 2006)**
- **La denervazione spontanea del muscolo compare a circa 20 gg nel muscolo normale (Van Putten et al., 2002) e l'atrofia muscolare inizia a 10-20 gg dall'inoculo di BT con un picco a 42 gg nel muscolo spastico (Hamjian and Walker, 1994)**
- **Recentemente è stato provato che l'effetto dell'inoculo di BT può essere aumentato se tale inoculo viene eseguito esattamente nella zona della placca neuromuscolare (Lapakti et al., 2010). Con ciò possibile ottimizzazione dell'efficacia, risparmio e riduzione di effetti collaterali come deficit su muscoli vicini**

SERIAL NEUROPHYSIOLOGICAL STUDIES OF INTRAMUSCULAR BOTULINUM-A TOXIN IN HUMANS

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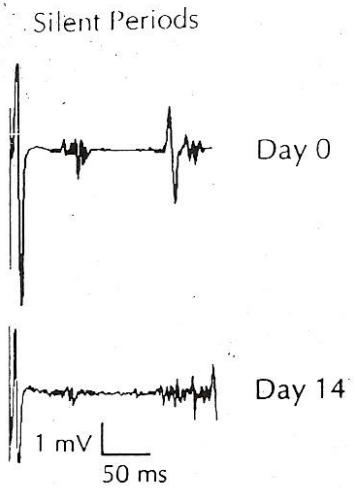

FIGURE 1. The amplitude of the recorded silent period of the EDB diminishes 2 weeks after administration of botulinum toxin, but its duration does not change. Note that the voluntary potential at 50 ms has also decreased in amplitude but not duration.

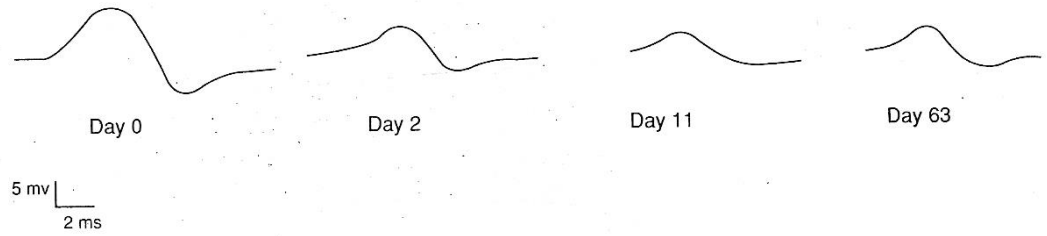

FIGURE 2. Serial EDB CMAP recordings of 1 individual following botulinum toxin administration. The decline in amplitude and area begins by day 2. It returned to baseline by day 100 in only 2 of 10.

Secondary Nonresponsiveness to Botulinum Toxin A in Cervical Dystonia: The Role of Electromyogram-Guided Injections, Botulinum Toxin A Antibody Assay, and the Extensor Digitorum Brevis Test

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Abstract: We studied 20 patients with cervical dystonia who had started to respond poorly to botulinum toxin A (BTXA) injections after an initial good response. All patients had extensor digitorum brevis (EDB) tests performed in addition to BTXA immunoprecipition assay (IPA) and mouse bioassay (MBA) antibody testing. The patients were reexamined and then treated with carefully placed electromyogram (EMG)-guided BTXA. Nine patients had a good clinical response to EMG-guided injections and all of these patients showed an obvious decrement on the EDB test. All were BTXA blocking antibodies (Abs)–negative via IPA and MBA (apart from one patient who had low BTXA antibodies titers using IPA but no antibodies by MBA). In the other 11 patients, there was a poor clinical response to EMG-guided BTXA injections. Seven of these 11 had small EDB decrement and BTXA antibodies using IPA, suggesting resistance to BTXA. Of the remaining four patients, two had obvious EDB decrement and low antibody titers via IPA (one of them had no antibodies via MBA), while the other two patients showed obvious decrement on the EDB test and no antibodies via IPA. This study shows that the EDB test correlates better with the clinical response than the antibody assays and that EDB decrement does not always correlate quantitatively with the BTXA antibody titers. In patients with secondary nonresponsiveness, it is recommended that an EDB test is the initial investigation of choice. In those patients where the EDB test does not demonstrate resistance to BTXA, a reexamination of the patients and carefully placed injections under EMG guidance may improve results.

Improvement after BTXA treatment:

Decrement >73%

No improvement after BTXA treatment:

Decrement<23%

EDB Test

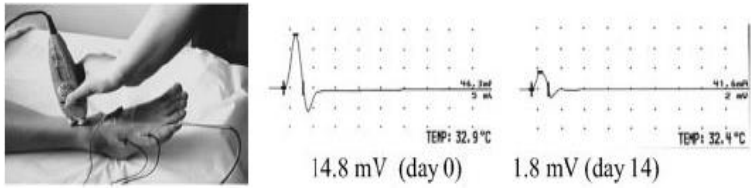

FIG. 1. EDB cMAP is recorded on the EDB muscle with a supramaximal stimulation of the peroneal nerve. Test is performed before and 2 weeks after BTXA injection into the EDB muscle. Note different amplitude calibration on the two traces.

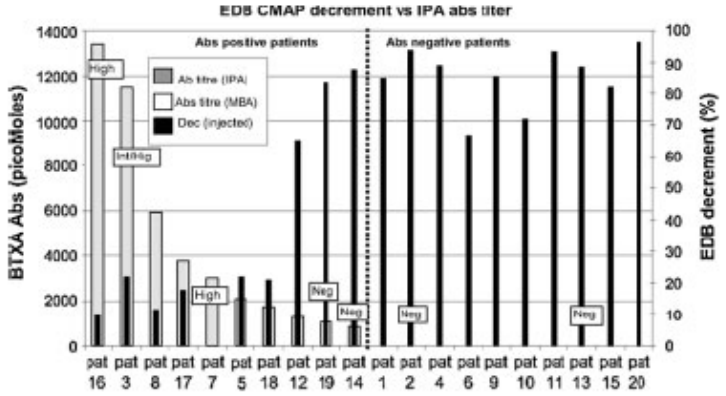

FIG. 2. Histogram showing EDB cMAP decrement in Abs-positive patients via IPA. Comparison with MBA assay titer in seven patients.

Needle EMG Guidance Is Rarely Required

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MUSCLE & NERVE November 2001

Table 1. Disadvantages of routine needle EMG guidance of botulinum toxin injections.

1. Need for additional equipment (hollow EMG needle, EMG machine and connecting cables)
2. More painful (larger diameter needles) ?
3. More time-consuming
4. More expensive
5. Determination of initial placement of EMG electrode is still dependent on clinical examination
6. If a muscle is obviously contracting and/or is hypertrophied, needle EMG is redundant (unless it is the antagonist muscle, which can be determined only by clinical examination, not by EMG)
7. The exact anatomic location of the EMG needle tip cannot be verified ?
8. Needle EMG does not differentiate between contractions produced by agonist versus antagonist muscles (and may thus lead to injection of the wrong muscle) ??
9. Needle EMG may be misleading—patients often "tense" otherwise uninvolved muscles during EMG
10. Single bolus EMG-guided injection leads to a greater risk of spread to adjacent, uninvolved unwanted muscles ???
11. The results of BTX treatment without needle EMG are so good that the small additional improvement (even if proven) does not justify the routine use of EMG ??

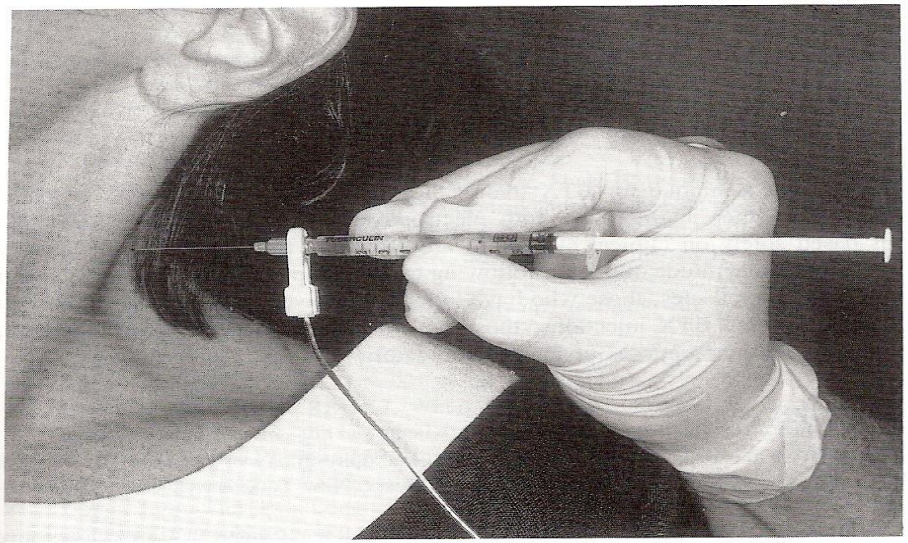


Figure 2 A demonstration of the electromyography (EMG)-assisted technique. In this figure, the sternocleidomastoid muscle is being targeted for injection. The tuberculin syringe containing botulinum toxin is attached directly to the electrode, and will be injected through the electrode when a full recruitment pattern appears on the EMG oscilloscope.

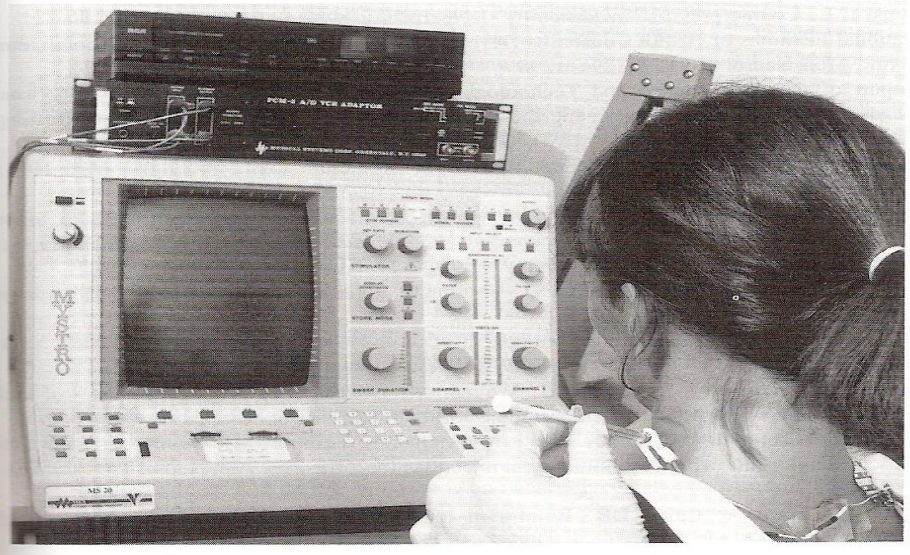
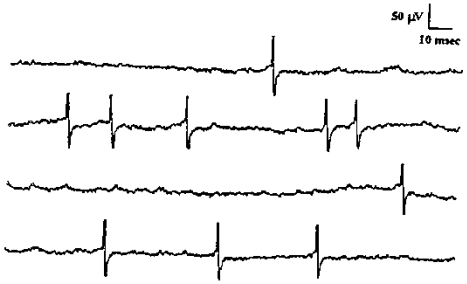


Figure 3 The position of the patient and examiner. When the electromyography (EMG)-assisted technique is used, the patient is seated in front of the EMG machine, and the treating physician is positioned behind the patient, permitting a full view of the EMG oscilloscope tracing.

Attività d'inserzione

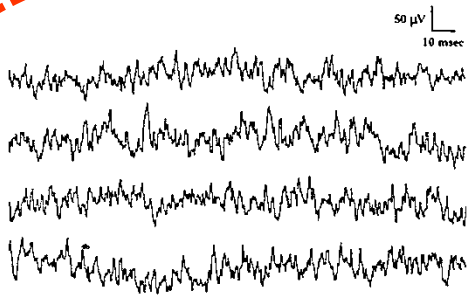
Normal insertional activity.

Potenziali di placca



End-plate spikes.

Rumore di placca con potenziali di placca in miniatura (MEPP)



End-plate noise.

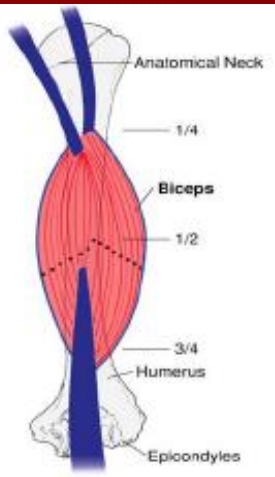


Fig. 1 Biceps

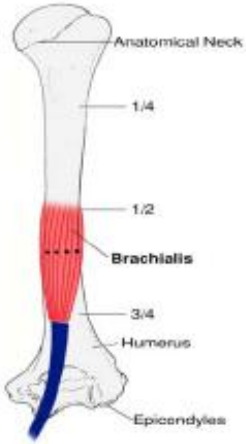


Fig. 2 Brachialis

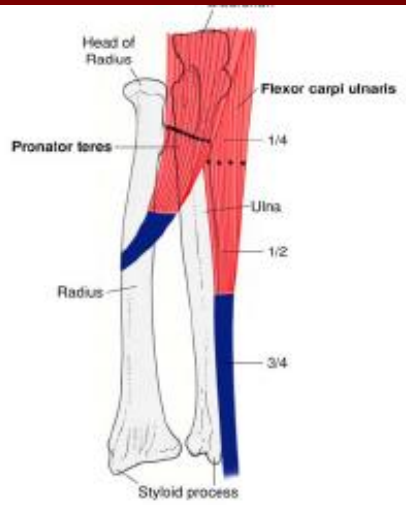


Fig. 3 Pronator teres and Flexor carpi ulnaris

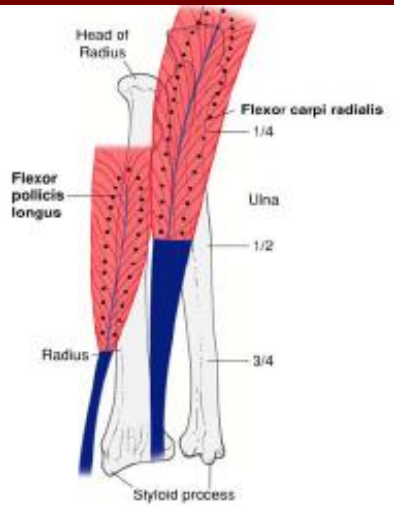


Fig. 4 Flexor pollicis longus and Flexor carpi radialis

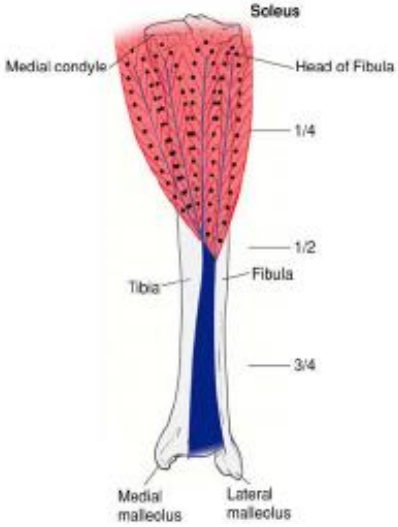


Fig. 7 Soleus

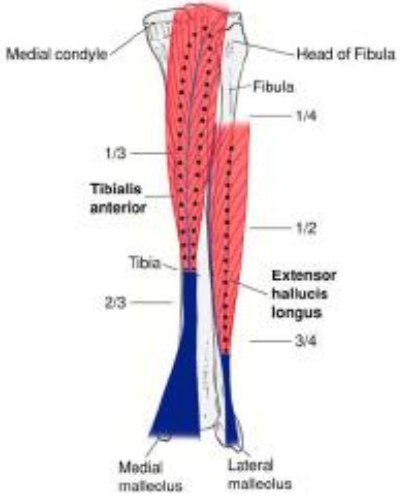


Fig. 8 Extensor hallucis longus

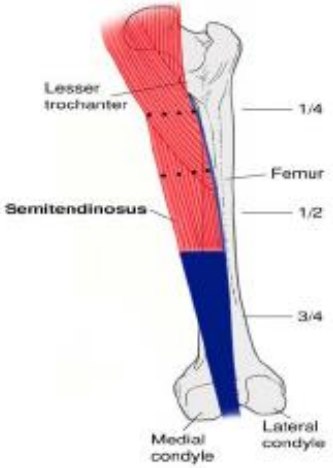


Fig. 11 Semitendinosus

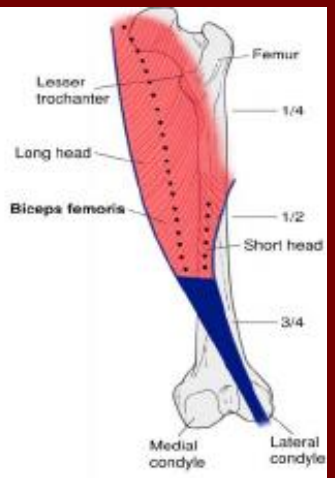
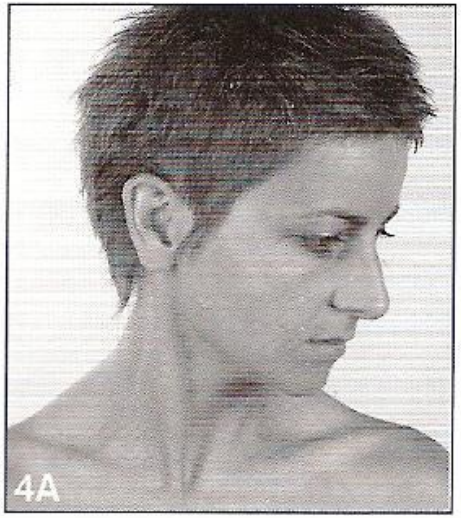


Fig. 12 Biceps femoris

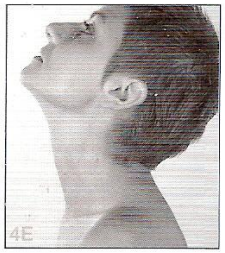
Endplate Zone Location of Commonly Injected Muscles

NAME OF MUSCLE	PENNATION	LOCATION OF MUSCLE FIBERS	PRESUMED NEUROMUSCULAR JUNCTION
Triceps	Bipennate	3 heads. Long head upper ¾ of humerus from just distal to the head of the humerus, lateral head upper ¾ of the humerus measuring from anatomical neck and short head little more than the lower ¼ of the humerus measuring from anatomical neck.	Oval bands distributed throughout the muscle.
Extensor digitorum	Multipennate	Almost the upper ¾ of the forearm measuring from the radial head	Oval bands distributed throughout the muscle.
Extensor carpi radialis longus	Unipennate	Upper ¾ of the radius measuring from the olecrenon of the ulna.	Horizontal band at about the upper ¼ of the radius measuring from the radial head.
Extensor carpi radialis brevis	Unipennate	Little above ¾ of the radius measuring from the olecrenon of the ulna.	Horizontal band little above the upper ¼ of the radius measuring from the radial head.
Flexor digitorum profundus	Multipennate	Upper 2/3rd of ulna measuring from the radial head.	Oval bands distributed throughout the muscle.
Flexor digitorum superficialis	Multipennate	Upper ¾ of ulna and radius measuring from the radial head.	Oval bands distributed throughout the muscle.
Pronator teres	Unipennate	Little above the upper half of the ulna measuring from the olecrenon.	Horizontal band at about the upper ¼ of the ulna measuring from the olecrenon.
Flexor pollicis longus	Bipennate	Upper half of ulna measuring from the olecrenon.	Oval bands distributed throughout the muscle.
Adductor longus	Unipennate	Upper ⅓ of femur measuring from femoral head.	Horizontal band at about the upper ¼ of the femur.
Adductor brevis	Unipennate	Horizontal fibers in area of the lesser trochanter of femur and slightly below it.	Vertical band at about the area where a vertical line dropped from the femoral neck would lie
Rectus femoris	Diverging bipennate	Cover about upper 2/3 of femur measuring from Greater trochanter of femur .	Oval bands distributed throughout the muscle.
Vastus medialis	Unipennate	Neck of femur to slightly above the medial condyle.	Vertical band along the muscle.
Vastus lateralis	Unipennate	Greater trochanter of femur to about the lateral condyle of the femur.	Vertical band along the muscle.
Gracilis	Unipennate	Just distal to the head of the femur to the ¾ of the femur measuring from the head of the femur.	Two horizontal bands one at about ¼ and second at about ½ of distance measured from the femoral head.
Sartorius	Strap muscle.	Greater trochanter of femur to little below the medial condyle	Distributed all over the muscle
Flexor hallucis longus	Unipennate	Upper 1/4 to little below the middle of the fibula measuring from the fibular head.	Vertical band along the muscle .
Flexor digitorum longus	Bipennate	Middle 3rd of tibia measuring from the medial tibial condyle.	Oval bands distributed throughout the muscle.
Extensor digitorum longus	Unipennate	Upper ¾ of fibula measuring from fibular head.	Vertical band along the muscle fibers

4.1 Spasmodic Torticollis

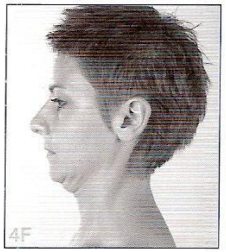


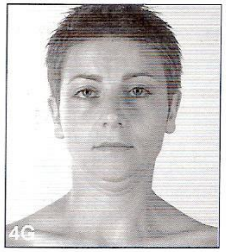
4.3 Retrocollis/Posterior Shift



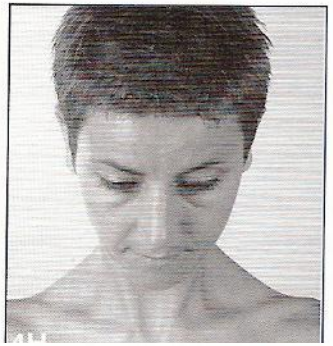
Involved Muscles in Retrocollis

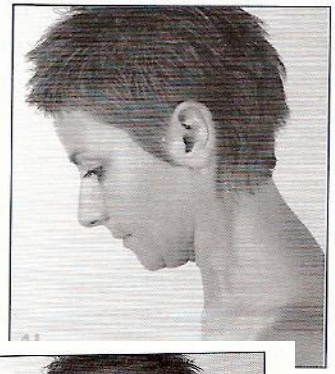
*** Mm. splenius capitis (see page 120)
*** Mm. semispinales capitis (see page 119)
** Mm. scaleni posteriores (see page 118)
** Mm. trapezii, pars descendens (see page 122)
* Mm. longissimi colli

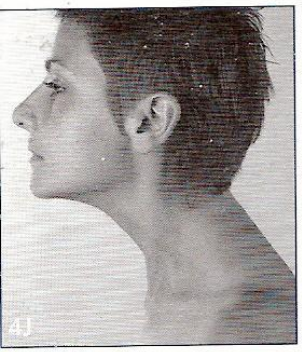


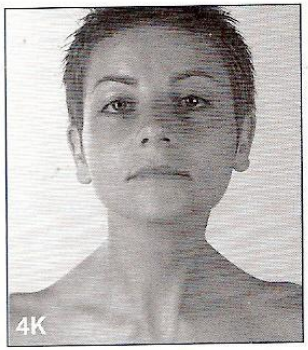


4.4 Anterocollis/Anterior Shift

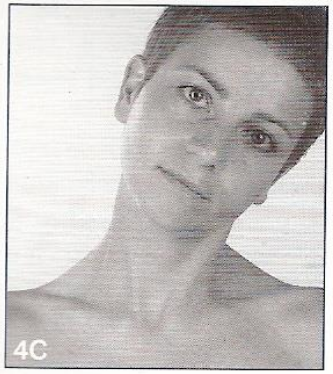


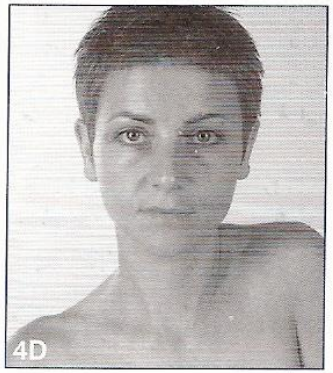






4.2 Laterocollis/Dystonic Scapular Elevation





Electromyographic evaluation of cervical dystonia for planning of botulinum toxin therapy

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Keywords:
botulinum toxin, cervical dystonia, electromyography, evaluation, therapeutic use

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The success of botulinum toxin (BT) injections for treatment of cervical dystonia depends on precise identification of dystonic muscles and on quantification of their dystonic involvement. Conventionally, this is attempted by clinical examination analysing the dystonic head position. In this presentation, a more systematic approach is sought by using an electromyography (EMG)-based evaluation procedure.

In 10 consecutive patients with cervical dystonia not previously exposed to BT clinical examination, analysing the dystonic head position was performed to classify patients into four groups with similar dystonic head positions. Additionally, a 2-channel concentric needle EMG was used to measure the amplitudes of dystonic and maximal voluntary activities in sternocleidomastoid (SCM), splenius capitis (SC) and trapezius/semispinalis capitis (T/SS) muscles bilaterally. The ratio between both amplitudes, the dystonia ratio, was used to quantify dystonic muscle involvement.

In all patients dystonia ratios could be calculated. In patients with similar head positions, EMG evaluation revealed different qualitative and quantitative dystonic involvement patterns. In six patients, there were discrepancies in identification of dystonic muscles between clinical examination and EMG evaluation. EMG evaluation excluded dystonic involvement in five patients. All excluded muscles were SCM. In one of these patients, additional T/SS involvement was detected by EMG evaluation. In one patient, SC involvement was revealed by EMG evaluation. All dystonic muscle involvement detected by EMG evaluation represented genuine dystonic muscle coactivation rather than compensatory muscle activity.

The EMG evaluation presented allows quantitative and qualitative identification of dystonic muscle involvement which cannot be achieved by clinical examination. Both pieces of information may be helpful for optimization of BT therapy.

Common function of muscle	Muscle
Head extensors	Levator scapulae Semispinalis capitis Splenius capitis Erector spinae Rectus capitis posterior, major and minor Trapezius
Head flexors	Sternocleidomastoid Anterior scalenes Platysma Digastrics Other suprahyoid† and infrahyoid muscles† Longus colli†
Common muscles that tilt the head (all ispislateral)	Sternocleidomastoid Anterior scalene Middle and posterior scalene Levator scapulae Splenius capitis/cervicis Semispinalis capitis Oblique capitis superioris Intertransversarii Trapezius
Common muscles that rotate the head	Contralateral Sternocleidomastoid Trapezius Levator scapulae Semispinalis Ipsilateral Splenius capitis Oblique capitis inferioris
Muscles that elevate the shoulder	Levator scapulae Trapezius

† Botulinum toxin injections pose significant risk for swallowing difficulties.

Retrocollis

Antecollis

Laterocollis

Torticollis

Shoulder elevation

	Outcome group		
	Good effect (%)	Unsatisfactory effect (%)	Total population (%)
	52 (67)	26 (33)	78 (100)
Changes in complexity pattern[a]			
No change	38 (76)	12 (24)	50 (100)
Less complex	10 (67)	5 (33)	15 (100)
More complex	4 (31)	9 (69)	13 (100)
Severity of head deviation prior to treatment[b]			
Mild	10 (59)	7 (41)	17 (100)
Moderate	27 (66)	14 (34)	41 (100)
Severe	15 (75)	5 (25)	20 (100)

[a]Pearson's $\chi^2 = 9.498$, $P = 0.009$.
[b]Pearson's $\chi^2 = 1.108$, $P = 0.575$.

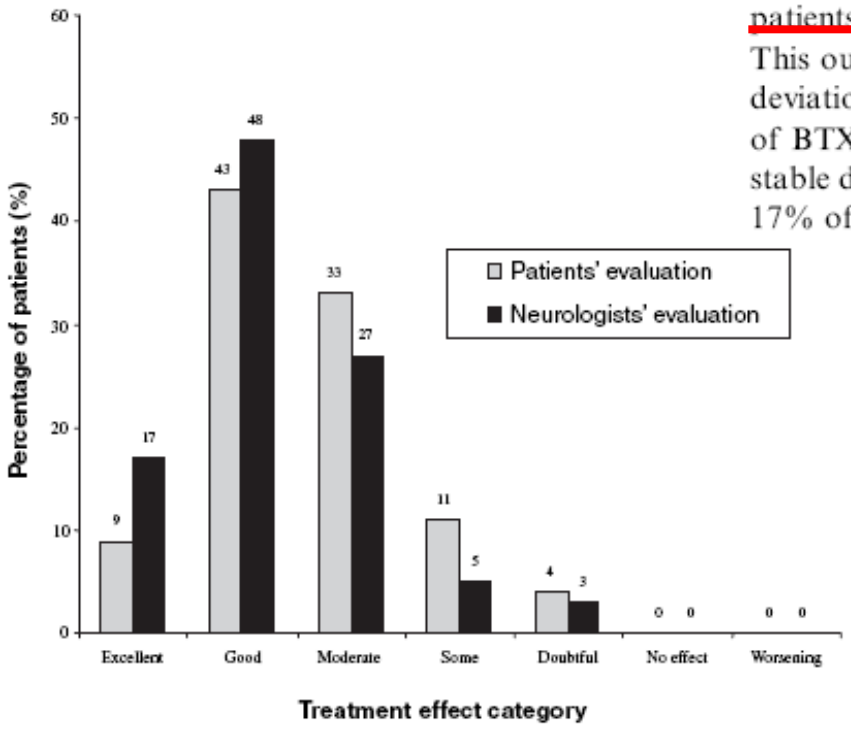


Figure 1 Long-term efficacy of botulinum toxin A as evaluated by the patients ($n = 76$, gray columns) and by the treating neurologists ($n = 71$, black columns). The columns represent the percentage of patients in each treatment effect category.

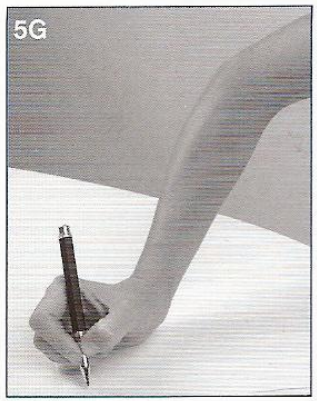
The course of cervical dystonia and patient satisfaction with long-term botulinum toxin A treatment

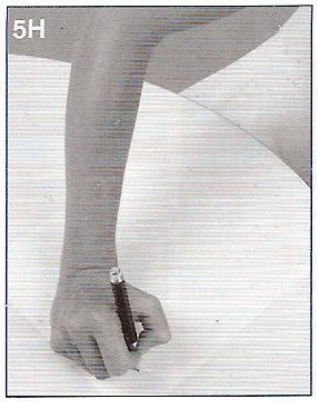
I. M. Skogseid and E. Kerty

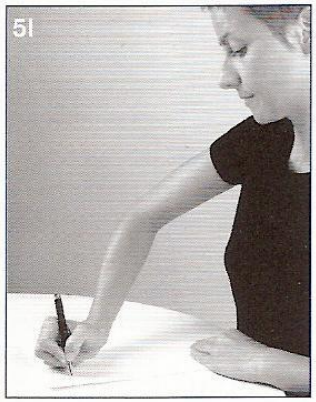
European Journal of Neurology 2005, **12:** 163–170

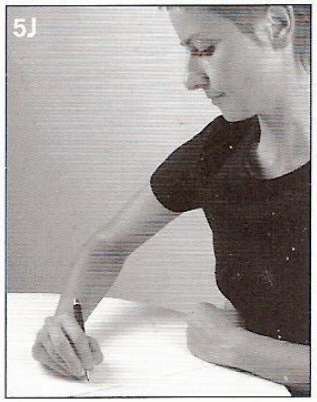
In 78 patients with idiopathic cervical dystonia (CD), we studied the course of the disease and the patients' satisfaction with long-term botulinum toxin A (BTX) treatment (median 5.5 years, range 1.5–10). On a seven-point scale ranging from excellent to worsening, the effect of treatment was scored as excellent or good by 52% of patients and moderate by 33%. The independent scores of the treating neurologists were excellent or good in 65% and moderate in 27%, respectively, and correlated well with the patients' scores. The 'Global Burden of Disease', as expressed on Visual Analog Scales (VAS, 0–10) before and at evaluation of treatment, was reduced by a median of 4 in individual patients. By combining these outcome measures, 67% of the patients were characterized as having a good effect, and 33% an unsatisfactory effect. This outcome (good or unsatisfactory effect) was independent of the severity of head deviation or complexity pattern of CD prior to treatment, the delay from onset to start of BTX treatment, or the number of treatments. The complexity pattern remained stable during treatment in 64% of the patients, became less complex in 19%, whereas 17% of the patients developed more complex patterns.

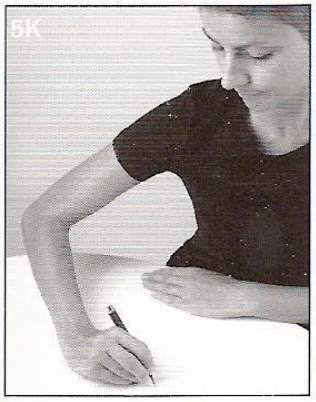
Extension Type

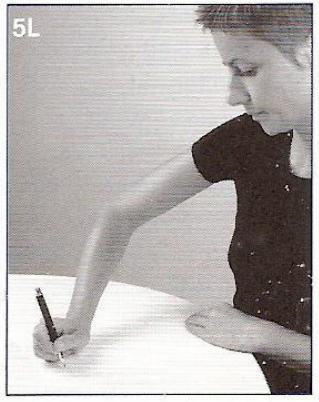




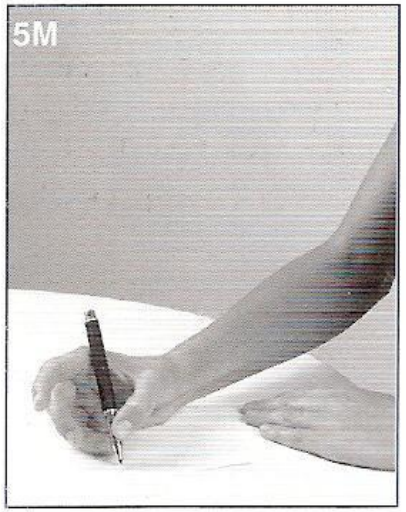


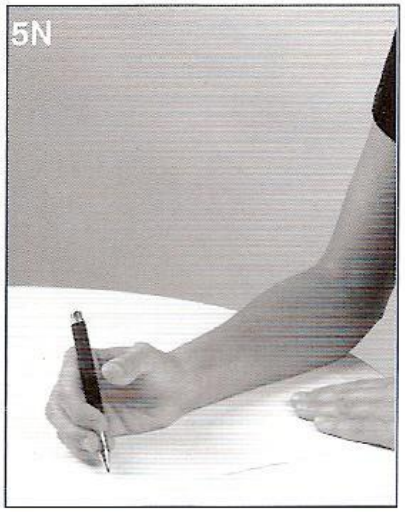


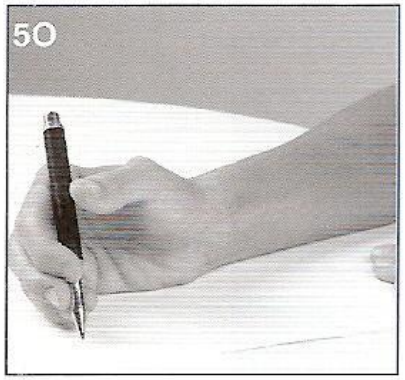


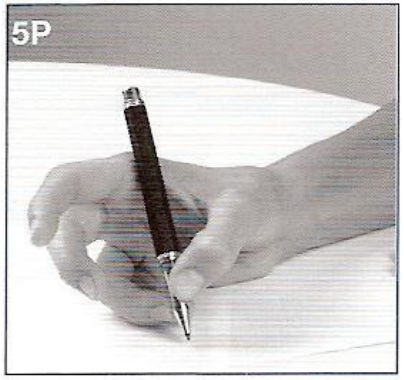


B 5 Action-induced Dystonias of the Hand









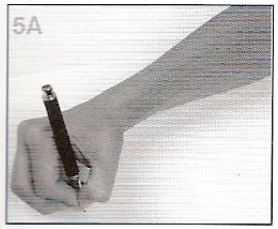
Involved Muscles

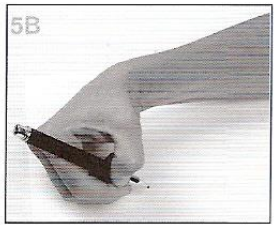
*** M. extensor carpi radialis (see page129)
*** M. extensor carpi ulnaris (see page 130)
** M. extensor digitorum communis (see page 131)
** M. extensor pollicis longus
** M. adductor pollicis (see page 141)
* M. pronator quadratus

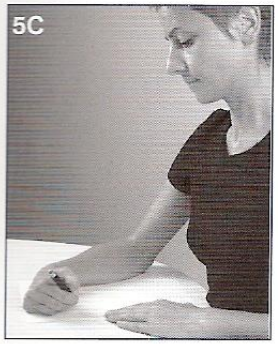
Flexion Type

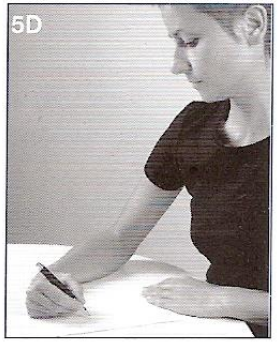
Involved Muscles

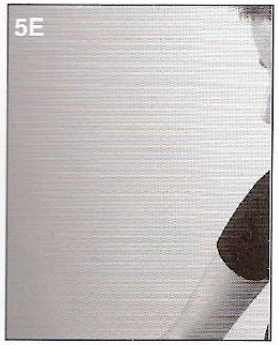
*** M. flexor carpi radialis (see page 132)
** M. flexor carpi ulnaris (see page 133)
** M. flexor digitorum superficialis (see page 135)
** M. flexor digitorum profundus (see page 134)
* M. palmaris longus
* M. flexor pollicis longus (see page 136)
** M. flexor pollicis brevis (see page 140)

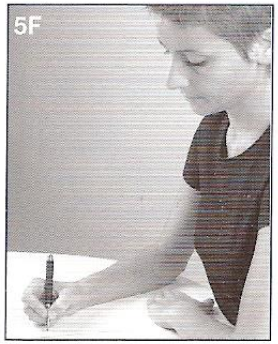












Factors predicting improvement in motor disability in writer's cramp treated with botulinum toxin

R Djebbari, S T du Montcel, S Sangla, J S Vidal, G Gallouedec, M Vidailhet

J Neurol Neurosurg Psychiatry 2004;75:1688–1691

scores. Patients with a pronation/flexion pattern of dystonia showed the best and the most sustained improvement. Primary writing tremor was little improved. There was a correlation between the self assessment score and the Burke-Fahn-Marsden score. Benefit was maintained over time

Conclusions: These results have implications for the identification of patients most likely to benefit from BTX injections.

Table 2 Severity and disability scores of the Burke-Fahn-Marsden scale before and after treatment

Scores	Before treatment	After treatment	
Severity	2.8 (1.8)	1.8 (1.7)	p<0.0001
Disability	1.9 (0.8)	1.1 (1.0)	p<0.0001

Scores are expressed as mean (SD); scores were obtained at the "first visit" and at the "last visit".

Dystonia disability scale: subscore for handwriting (tremor or dystonia)

0, Normal
1, Slight difficulties, legible.
2, Almost illegible.
3, Illegible.
4, Unable to grasp to maintain hold on pen.

Table 3 Predictive factors for improvement in disability and severity

	Disability			Severity		
	β	SE	p Value	β	SE	p Value
Sex			NS			NS
Age			NS	0.03	0.01	<0.006
Duration of evolution			NS			NS
Extension of the thumb	0.56	0.25	<0.04			NS
Flexion and pronation of forearm	0.66	0.29	<0.03			NS
Other affected muscles			NS			NS
Myoclonus			NS			NS
Mirror movements			NS			NS
Physiotherapy			NS			NS
Number of muscle injected (less than *v* more than 3)			NS			NS

β: parameter estimate. For categorical variables, the improvement is increased of a β factor when the tested factor is present. For continuous variables, the improvement is increased of a β factor per unit increase of the tested factor.

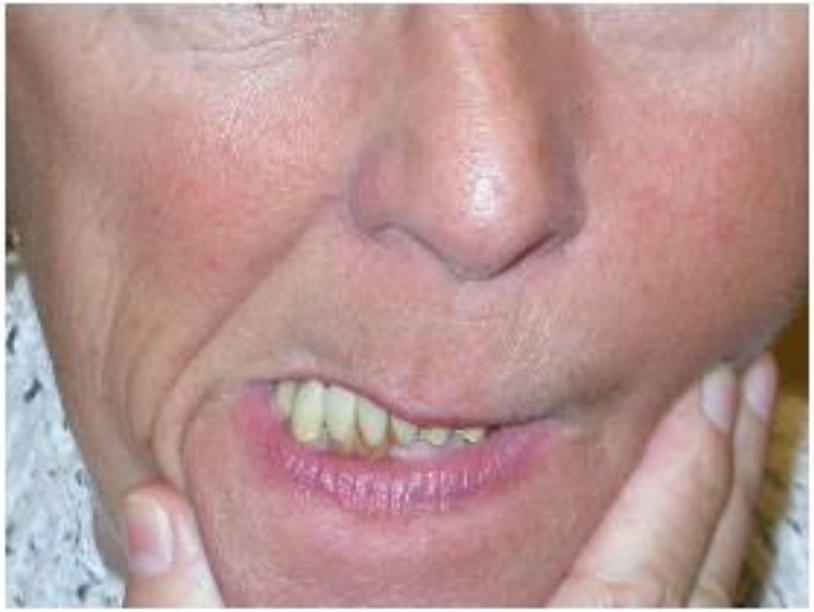
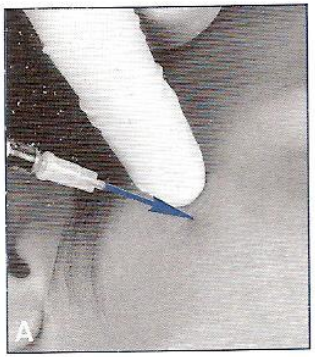


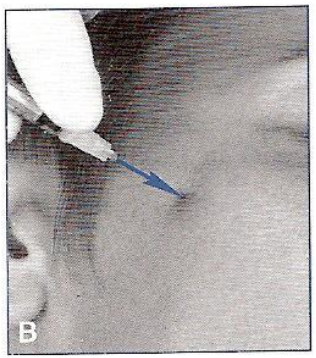
Figure 5
Patient with OMD: Pathologic movements of the mandible are evident, patients use so called "gestes antagonistiques" to break the dystonic activity of the jaw muscles.

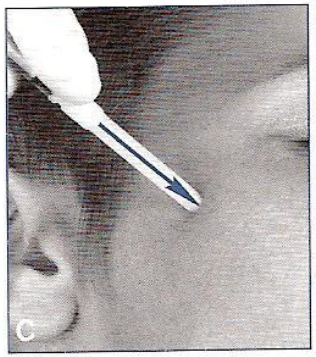
M. Pterygoideus Lateralis Procedure

Using EMG control, injections are given preauricular through the semilunar incisure while the patient keeps the mouth slightly open with relaxed muscles of mastication.



As the needle touches the lateral lamina of the pterygoid process, slightly pull needle back, tilt it towards dorsocranial and then push it further forward. The electromyographic pattern of action potentials obtained while the patient actively pushes the lower jaw forward (rostal) or towards the contralateral side helps to ascertain correct placement of the needle.





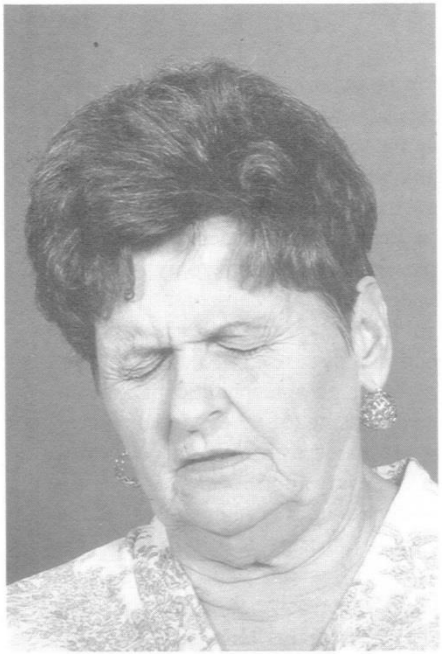
As to be expected, the treatment does change the pattern of lower jaw movements. The translative component during opening of the mouth decreases. However, at the most, if at all, patients may find it a little more difficult to bite off pieces of food because of the reduced distance between the cutting edges of upper and lower incisors. Otherwise, patients will be unaware of the restrictions in mouth opening.

Pterigoideo Laterale (Mov. di lateralità e stabilizzazione della mandibola)

EMG Pteriogoideo Laterale

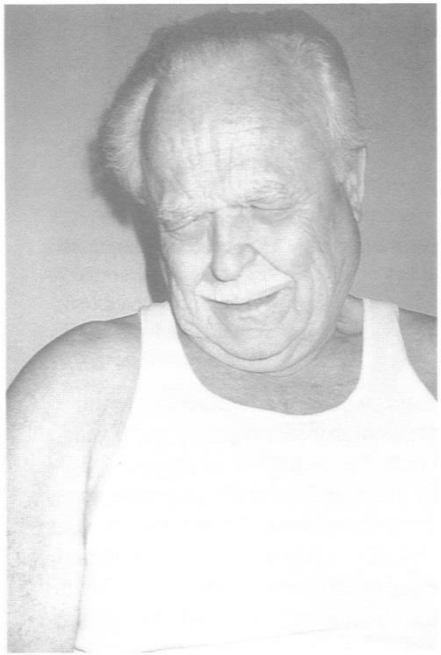
Pterigoideo Mediale (Elev. Mandibola)

EMG Pterigoideo Mediale



(a)

(b)



(c)

(d)

Table 1 CNS Diseases Associated with Eyelid Closure

- Blepharoclonus
  - Primary
  - Secondary
    - Hydrocephalus
    - Multiple sclerosis
- Chorea
  - Huntington's disease
  - Sydenham's chorea
- Dystonia
  - Primary
  - Secondary
    - Acute dystonic reaction
    - Levodopa-induced dyskinesia
    - Midbrain-diencephalic lesion
    - Parkinsonism
      - Multiple system atrophy
      - Parkinson's disease
      - Postencephalitic parkinsonism
      - Progressive supranuclear palsy
    - Tardive dystonia
    - Tourette's syndrome
    - Wilson's disease
    - X-linked dystonia-parkinsonism
    - Others
- Essential tremor
- Eyelid "freezing"
  - Acquired hepatocerebral degeneration
  - Creutzfeldt-Jakob disease
  - Hallervorden-Spatz syndrome
  - Multiple system atrophy
  - Neuroacanthocytosis
  - Parkinson's disease
  - Postencephalitic parkinsonism
  - Progressive supranuclear palsy
  - Wilson's disease
- Myoclonus
- Seizure disorder
- Tourette's syndrome
- Whipple's disease

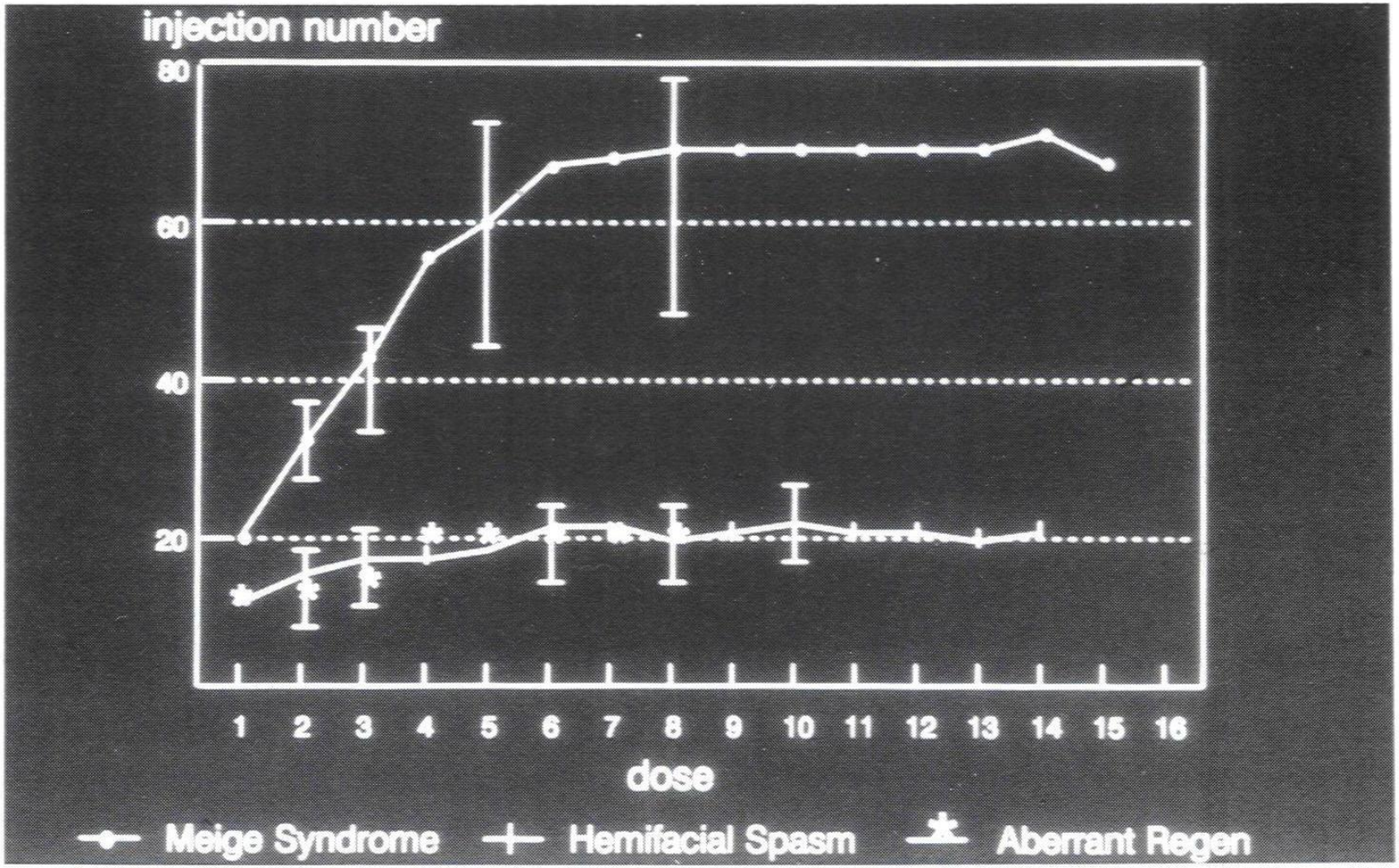


Figure 7 Dose requirement after multiple injection periods over an average of 3–4 years for various syndromes causing blepharospasm (Meige syndrome, aberrant regeneration of the facial nerve, and hemifacial spasm).

Caratteristiche comuni delle disfonie spasmodiche (DSP)

-I sintomi compaiono in assenza di anomalie strutturali della laringe (laringiti, polipi c. vocali , ulcere delle c. vocali , Ca laring. ecc..)

-Non sono presenti riduzioni dei movimenti delle c.vocali durante compiti non legati all'emissione di parole (respiro , tosse, ecc..)

-I sintomi si osservano essenzialmente nel parlare e non durante attività particolari come fischiare , cantare o l'emissione vocale in falsetto

-I sintomi sono azione-indotti; cioè, essi sono più evidenti nel movimento volontario

-I sintomi peggiorano sempre più con la durata del discorso e con l'esecuzione di discorsi in cui è presente un elevato stress emotivo. Molti pazienti riferiscono un peggioramento della voce alla fine della giornata

-In genere aspetti emozionali o riflessi della voce , quali tossire, ridere e piangere non sono coinvolti dal disturbo

-Per lo più colpisce età comprese fra 30 e 60 anni e nel 60% il sesso femminile

Caratteristiche nell'insorgenza delle DSP

Usualmente sono disordini di natura idiopatica, sebbene siano riportati in Letteratura casi di DSP secondari a traumi cranici

Inizio in genere graduale, da 1 a 2 anni , che segue ad un'infezione delle vie respiratorie superiori in almeno il 30 % dei casi

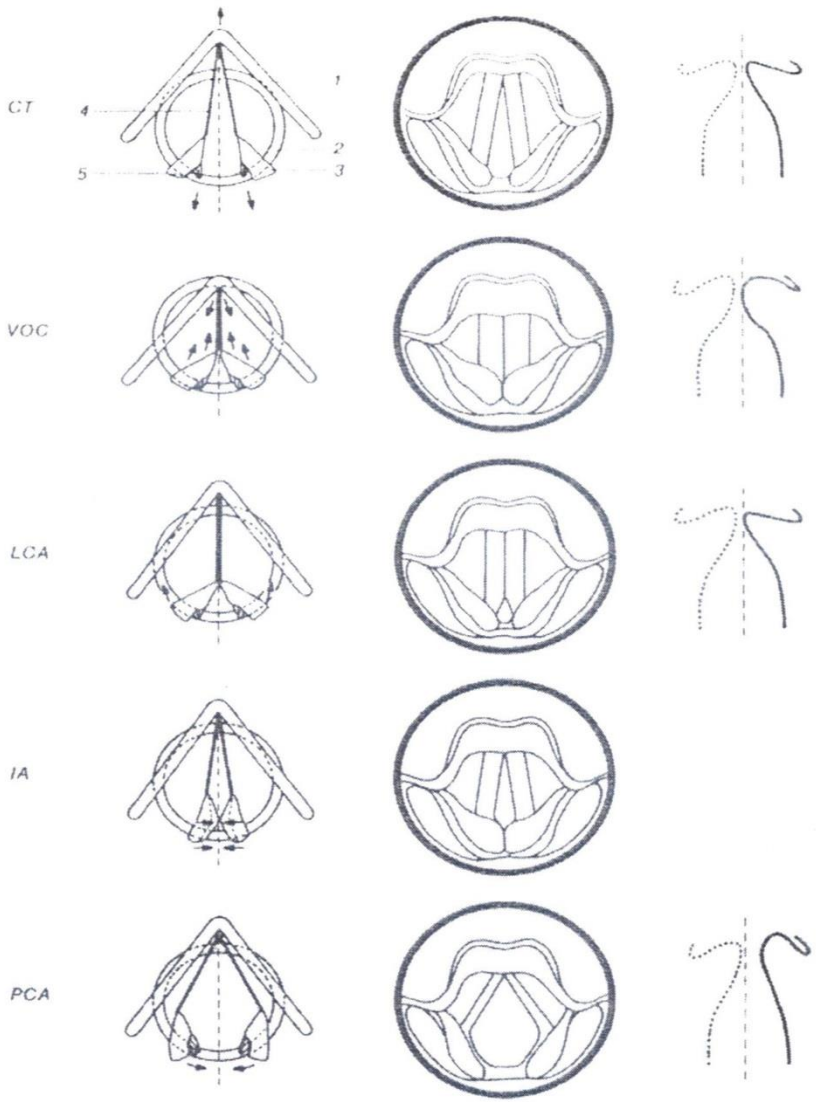
Frequente l'inizio in seguito a intensi stress emotivi. In genere i disturbi inizialmente compaiono solo durante situazioni di elevato stress emotivo

Sono stati riferiti recuperi spontanei nel 14% dei casi (ma in questo caso vi è il sospetto che le forme inquadrate siano essenzialmente psicogene)

Diagnosi Differenziale nelle DSP

E' fondamentale che la valutazione di tali patologie venga eseguita da un Team composto da Otorinolaringoiatra, Foniatra, Logopedista e Neurologo (Neurofisiopatologo)

- **Esclusione di varie patologie laringeee (Laringoscopia a fibre ottiche): polipi delle c.v., ca Laringeo, ulcere c.v. , laringiti , paralisi delle corde vocali di tipo centrale o periferico (paralisi del n.L ricorrente o superiore, SLA, Neuropatie ereditarie , ecc..)**
- **MSA (paralisi in abduzione o distonia delle corde vocali), M. di Wilson, M. di Parkinson, PSP, SM, Disordini cerebellari, patologie muscolari infiammatorie o degenerative (distrofie oculo-faringea, polimiosite ecc.)**
- **Disfonie psicogene o di conversione (?): DD estremamente difficile soprattutto per le forme di DSP in abduzione , presenti anche a voce bisbigliata**


CT
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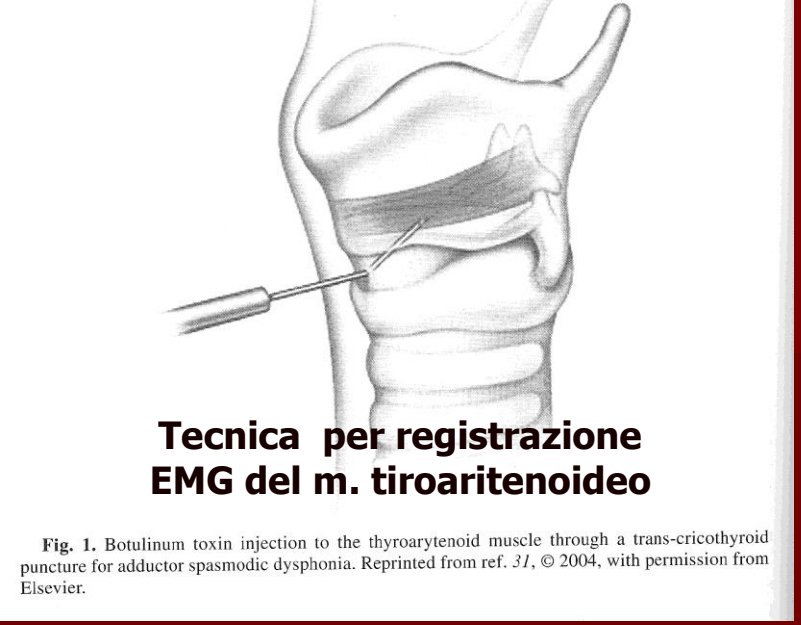
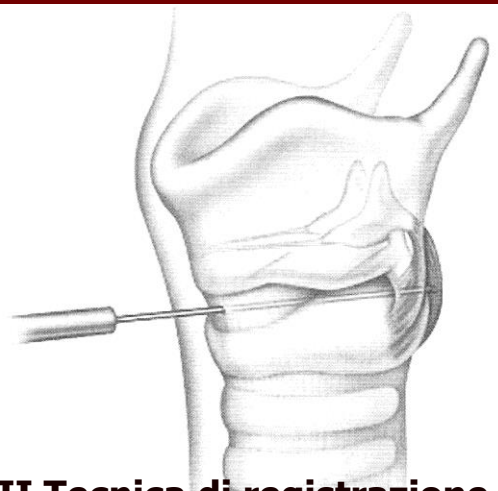


Tecnica per registrazione EMG del m. tiroaritenoideo

Fig. 1. Botulinum toxin injection to the thyroarytenoid muscle through a trans-cricothyroid puncture for adductor spasmodic dysphonia. Reprinted from ref. *31*, © 2004, with permission from Elsevier.

I tecnica per registrazione EMG del m. cricoaritenoideo posteriore

Fig. 2. Laryngeal rotation technique for botulinum toxin injection to the posterior cricoarytenoid muscle for abductor spasmodic dysphonia. Reprinted from ref. *31*, © 2004, with permission from Elsevier.



II Tecnica di registrazione EMG del m. cricoaritenoideo posteriore

Fig. 3. An alternate approach to the posterior cricothyroid muscle through the posterior cricoid lamina. Reprinted from ref. *31*, © 2004, with permission from Elsevier.

a) Normal

b) Abductor dysphonia

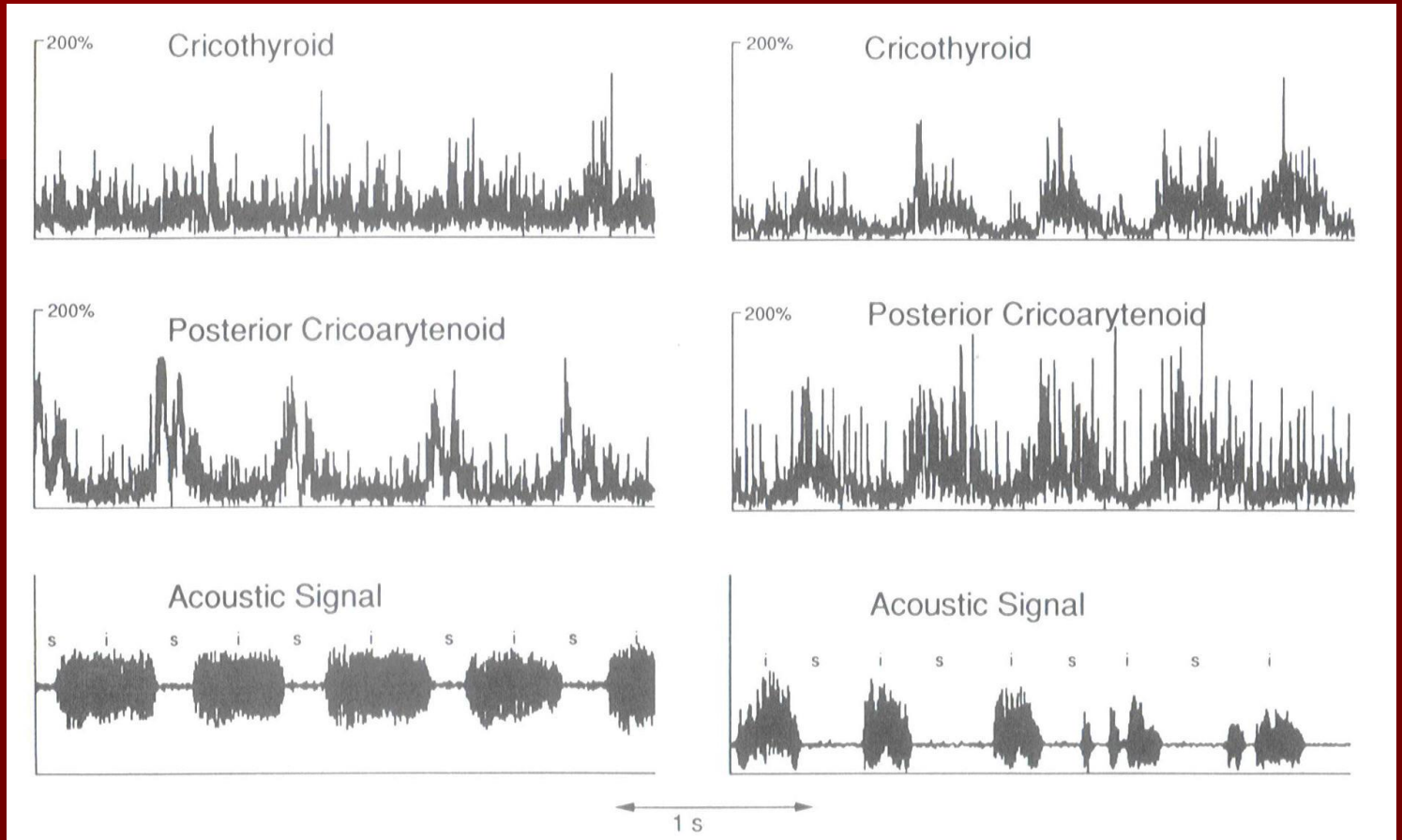
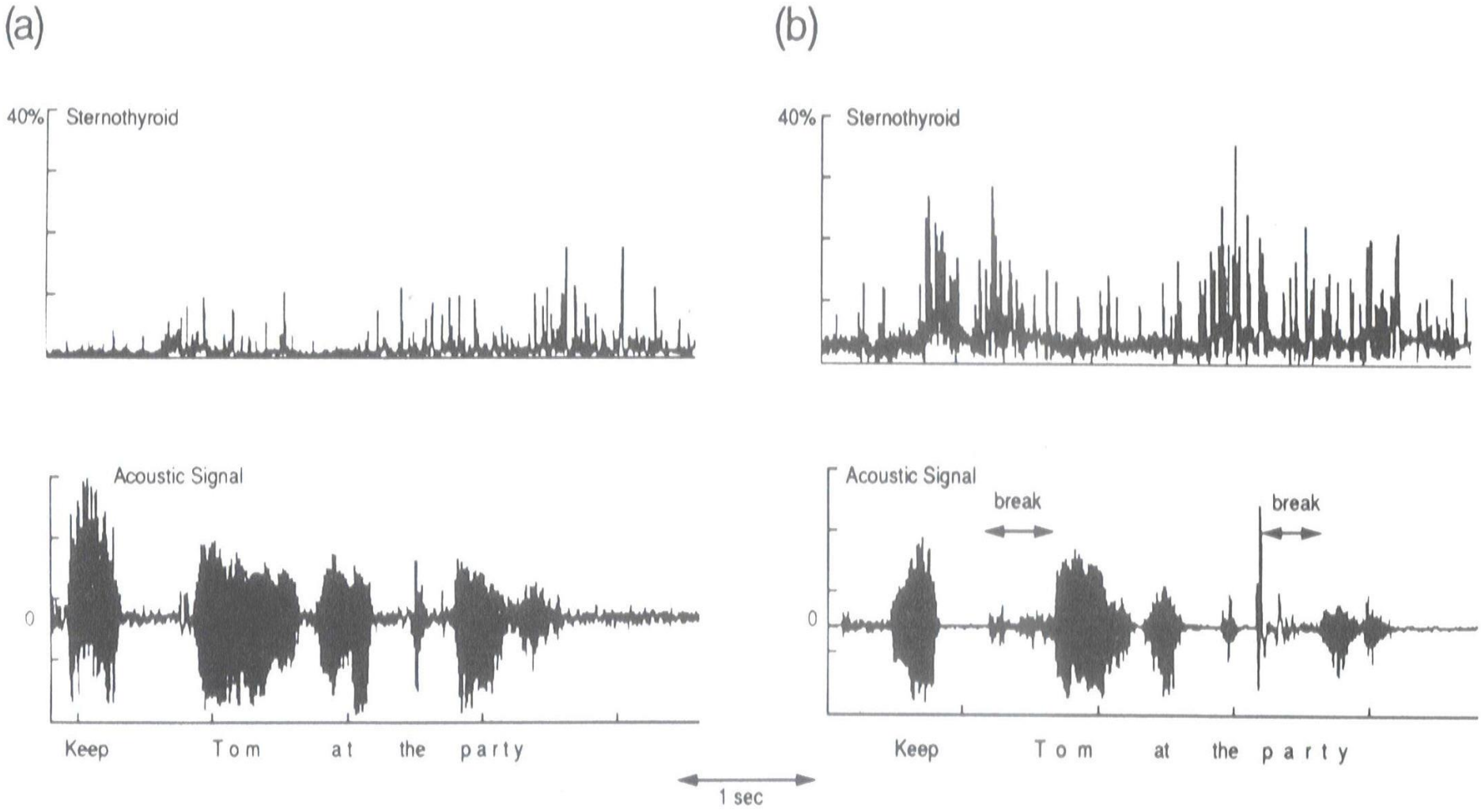


Figure 5 Prolonged bursts involving both the cricothyroid and posterior cricoarytenoid muscles during attempts at repetition of the syllable *si* in a patient with abductor spasmodic dysphonia (b) compared with a normal control (a). The prolonged bursts of these two muscles result in prolonged durations of vocal-fold opening for the *s* and shorter intervals of voicing for the vowel /i/.

NORMAL

ADDUCT. DYSPHONIA



Cricoaritenoideo post.: abduttore CV

EMG Cricoaritenoideo post.

Cricoaritenoideo post.: abduttore CV

EMG Cricoaritenoideo post.

Tiroaritenoideo: adduttore-tensore CV

EMG tiroaritenoideo

Cricoarit. Laterale: adduttore CV

EMG Cricoarit. Laterale

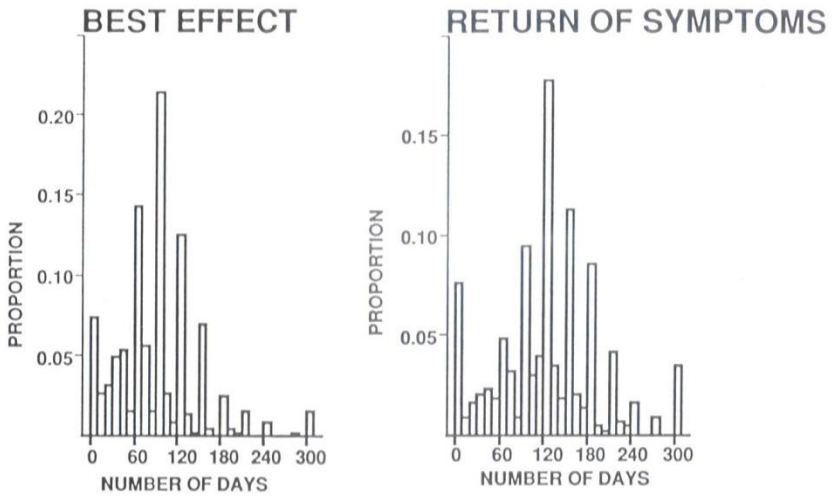


Figure 1 Frequency histograms showing the distribution of patient responses for the duration in days of their best effect following injection and the number of days before the return of symptoms. Includes the data from 503 injections.

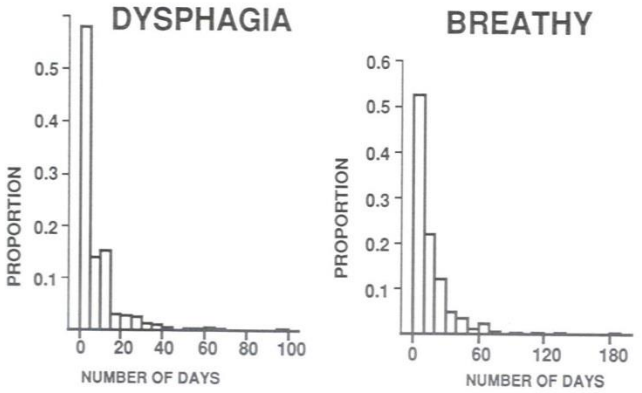
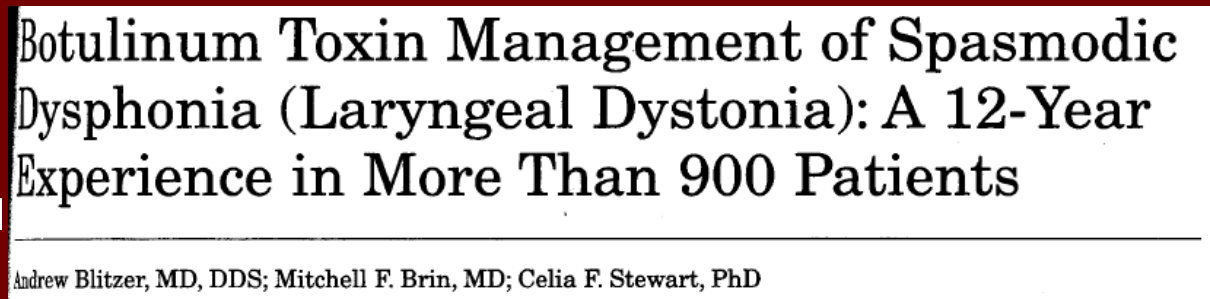


Figure 2 Frequency histograms showing the distribution of patient responses for the duration in days of dysphagia following injection and the duration in days of breathiness following injection. Includes the data from 503 injections.

EFFETTI COLLATERALI DEL TRATTAMENTO CON TOSSINA BOTULINICA DELLA DISFONIA ABDUTTORIA ED ADDUTTORIA

- Tirage e/o stridor nel 28 % dei casi
- Respiro "corto" nel 65% dei casi
- Disfagia 27-60% dei casi

Tali sintomi iniziano mediamente a 3 giorni dall'inoculo e regrediscono nel giro di 1-2 settimane


Botulinum Toxin Management of Spasmodic Dysphonia (Laryngeal Dystonia): A 12-Year Experience in More Than 900 Patients

Andrew Blitzer, MD, DDS; Mitchell F. Brin, MD; Celia F. Stewart, PhD

Laryngoscope, 108:1435–1441, 1998

Accuracy of muscle localization without EMG: Implications for treatment of limb dystonia

Abstract—Although botulinum toxin is an effective treatment for focal dystonia, the importance of electromyography (EMG) in identifying muscles and guiding injections is unclear. The authors examined the accuracy of muscle localization in 38 muscles in patients with focal hand dystonia without EMG guidance. Only 37% of needle placement attempts reached the target muscles or muscle fascicles. This study demonstrates that EMG guidance is needed for correct localization of desired muscles.

NEUROLOGY 2002;58:805–807

F.M. Molloy, MD; H.A. Shill, MD; A. Kaelin–Lang, MD; and B.I. Karp, MD

Neurology 2002;58;805

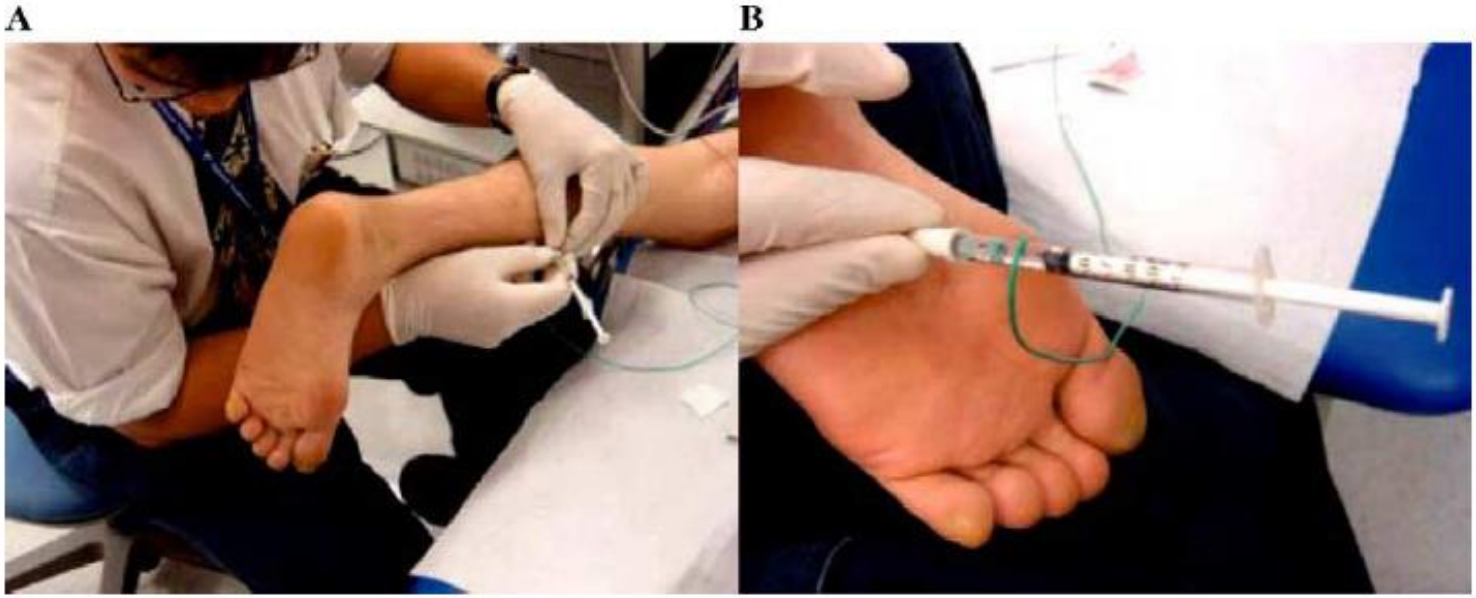


Fig. 1. Botulinum toxin injections under EMG guidance with electrical stimulation of the flexor digitorum longus (A) and brevis (B).

High doses of botulinum toxin effectively treat disabling up-going toe

M.M. Kurtis, A.G. Floyd, Q.P. Yu, S.L. Pullman •

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Involuntary up-going toe can be a disabling consequence of dystonia or spasticity. In this study, we treated eight patients with botulinum toxin (BTx) in the extensor hallucis longus (EHL) and applied objective and subjective outcome measures to determine treatment efficacy. Using 100% higher doses than generally reported, patients noted 62 ± 20% mean benefit and scores on a modified Fahn• Marsden Dystonia Scale decreased significantly by 1.8 ± 0.6 (p = 0.010). High doses (up to 160 BTx A units) into the EHL were safe and dosage correlated highly and significantly with treatment efficacy (• = 0.859, p = 0.006).

Table 1
Patient data

Pt	Age at onset (years)	Age started BTx (years)	Gender	Diagnosis	Follow-up (years)	Severity pre-BTx	Severity at max benefit	Subjective treatment response (%)	Initial EHL dose (U)	Stable EHL dose (U)	Duration of effect (months)	Total dose (U)
1	60	62	M	Task specific foot dystonia	11	2	0	90	60	160	7• 8	160
2	8	12	M	Generalized dystonia	11	4	1	80	35	160	2.5	400
3	58	60	F	Post-stroke hemiparesis	1	4	1	80	100	100	2.5	300
4	74	79	F	Post-stroke hemiparesis + late-onset dystonia	1	4	2	60	60	80	3	400
5	67	69	F	Post-stroke hemiparesis	7	3	1	60	60	75	3	400
6	55	58	M	Parkinson disease: off dystonia	3	3	1	50	50	50	3	200
7	7	48	M	Generalized dystonia	13	2	1	50	40	40	3	240
8	12	36	M	Generalized dystonia	2.5	4	3ß	30	50	75	3.5	400

Pt = Patient; U = Botox® equivalent units.

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Assessment: Botulinum neurotoxin for the treatment of movement disorders (an evidence-based review)

Report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology

ABSTRACT

Objective: To perform an evidence-based review of the safety and efficacy of botulinum neurotoxin (BoNT) in the treatment of movement disorders.

Methods: A literature search was performed including MEDLINE and Current Contents for therapeutic articles relevant to BoNT and selected movement disorders. Authors reviewed, abstracted, and classified articles based on American Academy of Neurology criteria (Class I–IV).

Results: The highest quality literature available for the respective indications was as follows: blepharospasm (two Class II studies); hemifacial spasm (one Class II and one Class III study); cervical dystonia (seven Class I studies); focal upper extremity dystonia (one Class I and three Class II studies); focal lower extremity dystonia (one Class II study); laryngeal dystonia (one Class I study); motor tics (one Class II study); and upper extremity essential tremor (two Class II studies).

Recommendations: Botulinum neurotoxin should be offered as a treatment option for the treatment of cervical dystonia (Level A), may be offered for blepharospasm, focal upper extremity dystonia, adductor laryngeal dystonia, and upper extremity essential tremor (Level B), and may be considered for hemifacial spasm, focal lower limb dystonia, and motor tics (Level C). While clinicians' practice may suggest stronger recommendations in some of these indications, evidence-based conclusions are limited by the availability of data. *Neurology*® 2008;70:1699–1706

UTILITA' DELL'ELETTROMIOGRAFIA CLINICA NEL TRATTAMENTO CON TOSSINA BOTULINICA

Metodo per la corretta localizzazione dei punti per l'inoculo per la tossina botulinica

Indagine preliminare, complementare all'inquadramento clinico-fisiopatologico per decidere i muscoli da inoculare

Indagine complementare a quella clinica per valutare gli effetti del trattamento con tossina botulinica