LA DISFONIA SPASMODICA: LA DIAGNOSI E LE TECNICHE DI TRATTAMENTO CON TOSSINA BOTULINICA

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SD diagnosis and classification should be carried out by a medical team: Neurologist, Otorhinolaryngologist, Speech Therapist

I step : differential diagnosis with vocal cord (VC) diseases

Exclusion of laryngeal diseases: VC polyps, Laryngeal Ca , VC ulcers, Laryngitis

II step: differential diagnosis between primary and secondary forms of motor disorders of VC

MSA (paralysis in VC abduction or VC dystonia), Wilson disease, Parkinson's Disease, PSP, MS, cerebellar disorders, inflammatory or degenerative muscle diseases (oculopharyngeal dystrophies , polymyositis, etc.) , central and peripheral palsy of VCs

III step: differential diagnosis between functional and organic SD

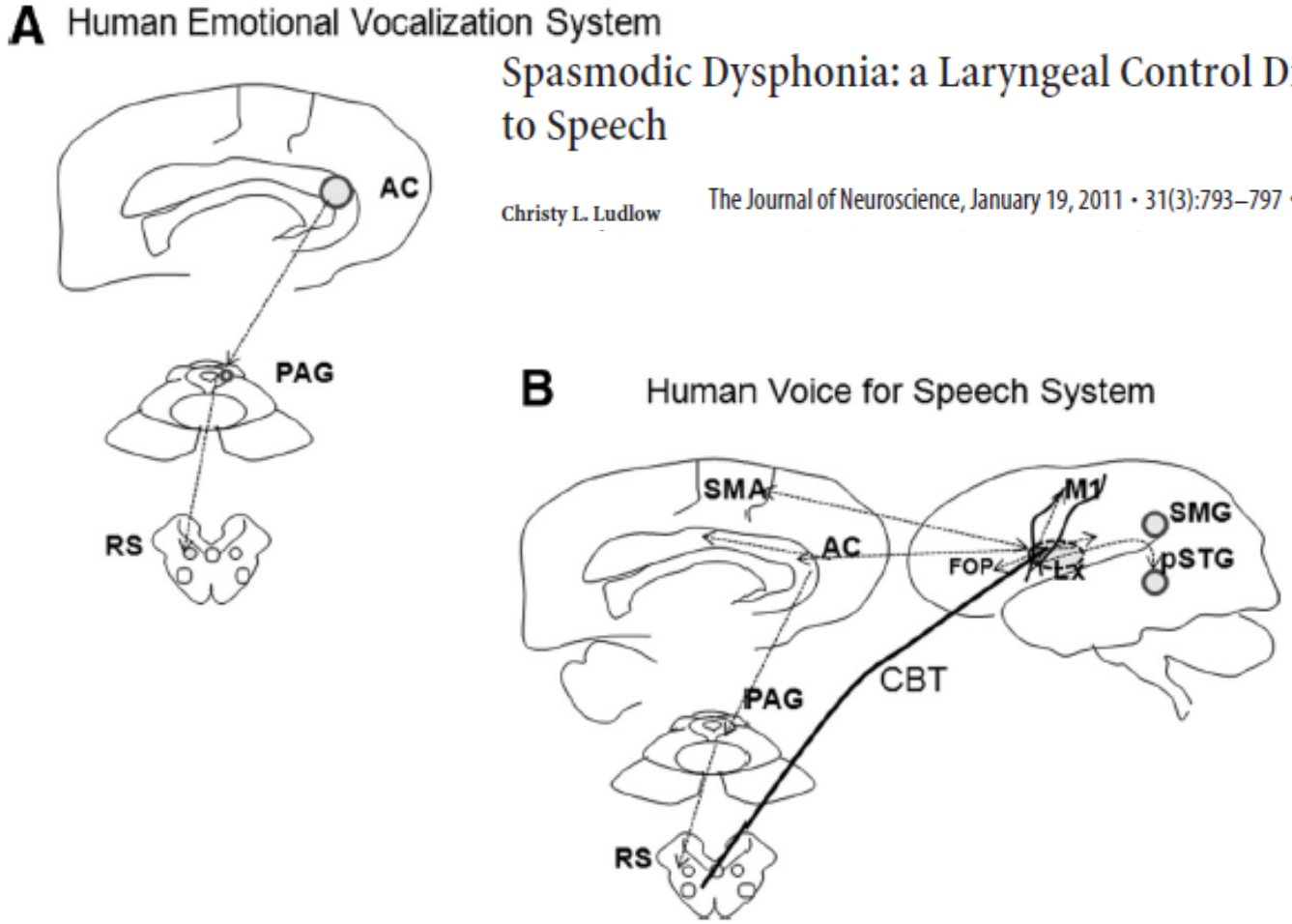
Psychogenic voice disorders (?): DD extremely difficult especially for forms of abd SD

Presbyphonia

- age related structural changes of the vocal folds- may cause glottal gap during voice production
- "presbyphonia is not a common disorder and should be a diagnosis of exclusion made only after careful medical and speech evaluation"

INSTRUMENTAL SET UP FOR DIAGNOSIS OF SPASMODIC DYSPHONIA

- Acoustic analysis
  - ex. Visi-pitch,
- laryngostroboscopic recordings
- electroglottography (EGG)
  - Assesses laryngeal functioning
- electromyography (EMG)
  - Assesses muscle function
- videofluoroscopic assessment
  - Swallowing test



Spasmodic Dysphonia: a Laryngeal Control Disorder Specific to Speech

Christy L. Ludlow

The Journal of Neuroscience, January 19, 2011 • 31(3):793–797 • 793

Figure 1. Illustration of the overlap and differences in the neural systems involved in emotional vocalization and in voice production for speech communication. ***A***, A schematic diagram of the human emotional vocalization system, which includes the anterior cingulate (AC), the periaquaductal gray (PAG), and the reticular system (RS) in the medulla with input to the laryngeal motor neurons in the nucleus ambiguous, based on the work of Jürgens (2002a,b) in the squirrel monkey. ***B***, A schematic diagram of the human voice for a speech system based on the study by Kuypers (1958), transcranial magnetic stimulation (Rödel et al., 2004), and functional neuroimaging (Schulz et al., 2005; Loucks et al., 2007; Chang et al., 2009; Simonyan et al., 2009), and includes the laryngeal motor cortex (Lx), the direct corticobulbar tract to the motor neurons (CBT), the frontal opercular speech system (FOP), the primary motor cortex (M1), the supplementary motor area (SMA), the posterior superior temporal gyrus (pSTG), and the supramarginal gyrus (SMG).

Movement Disorders of the Vocal Cord

- Adductor Spasmodic Dysphonia (Add SD)
- Abductor Spasmodic Dysphonia (AbdSD)
- Mixed Spasmodic Dysphonia (MixSD)
- Singer's Dysphonia (SingD)
- Vocal or Laryngeal tremor (V/L tremor)
- Respiratory Spasmodic Dysphonia (RSD)
- Stridor
- Spastic Dysarthria (SpDis)

Table 1. Pathophysiology of Spasmodic Dysphonia

Knowledge	Needs
Cortical involvement: somato-sensory and perisylvian	Better understanding of specific pathways
Subcortical involvement: basal ganglia and thalamus	Better localization, especially within possible brainstem areas
Genetic influence	Possible DYT6/THAP1

Spasmodic Dysphonia: An Evidence-Based Clinical Update

Balaji Rangarathnam, PhD, CCC-SLP, and Gary H. McCullough, PhD

Vol. 20, No. 12 December 2013 JCOM

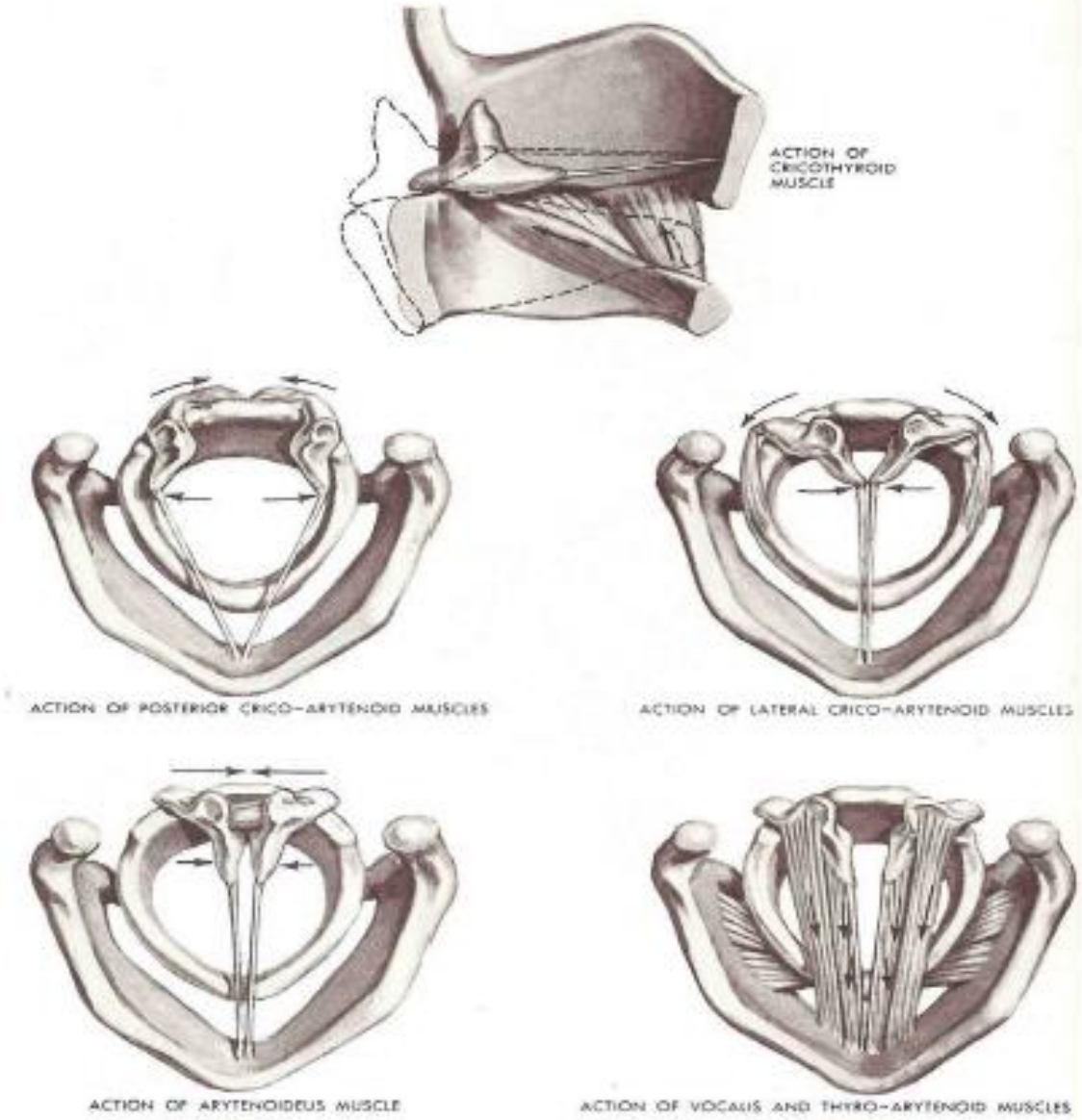
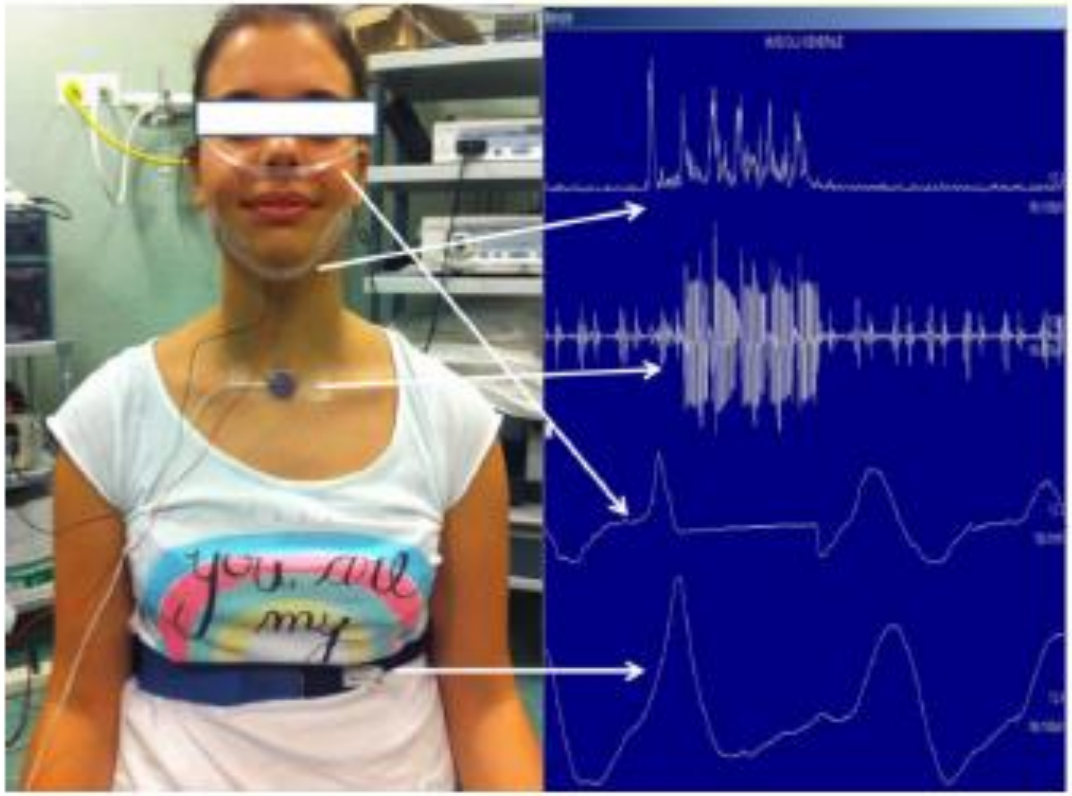


Fig. 3 - Azione dei muscoli intrinseci della laringe



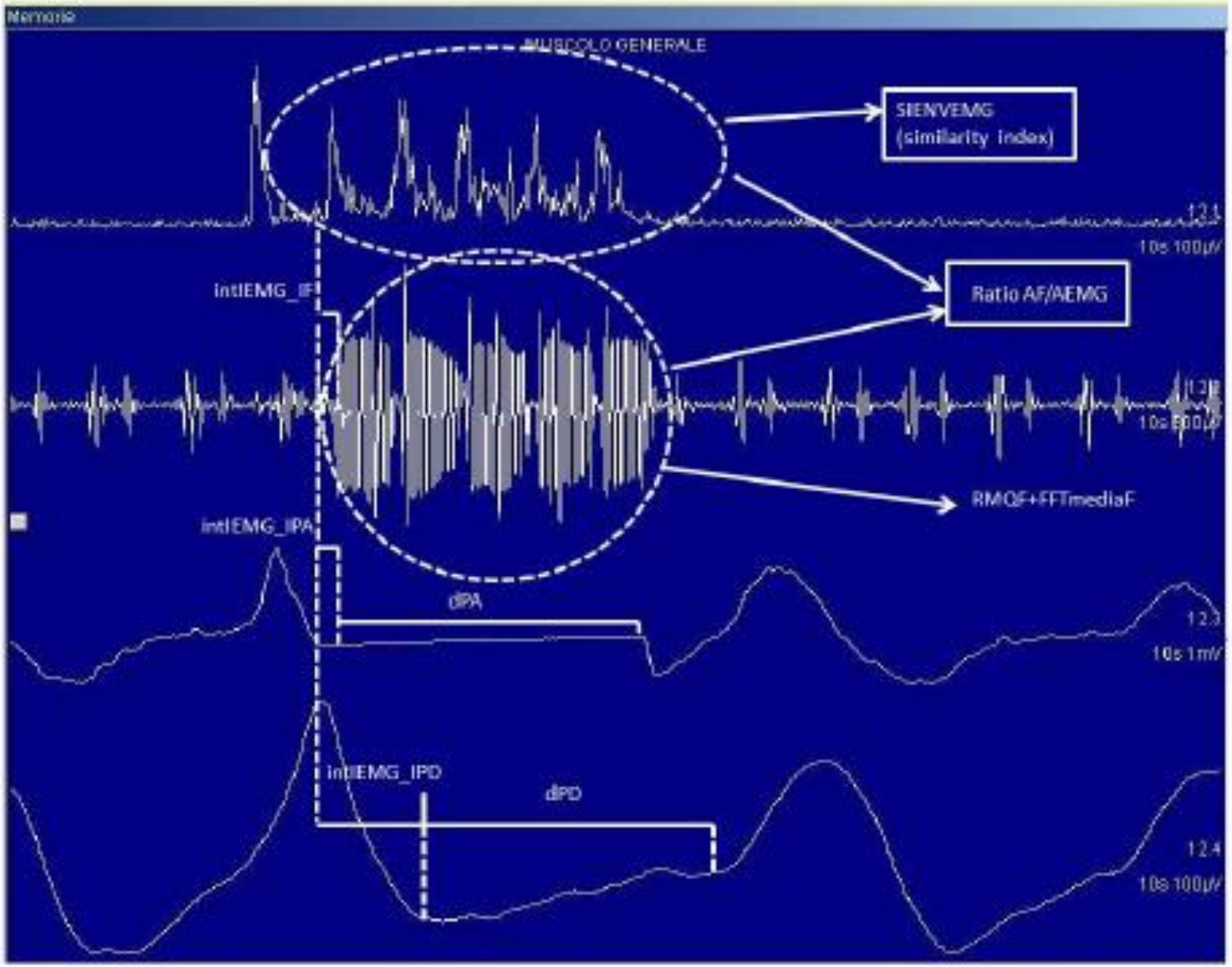
Rectified Surface-EMG from submental/suprahyoid muscles

Piezoelectric trasducer for tracheal air flux recording during the voice

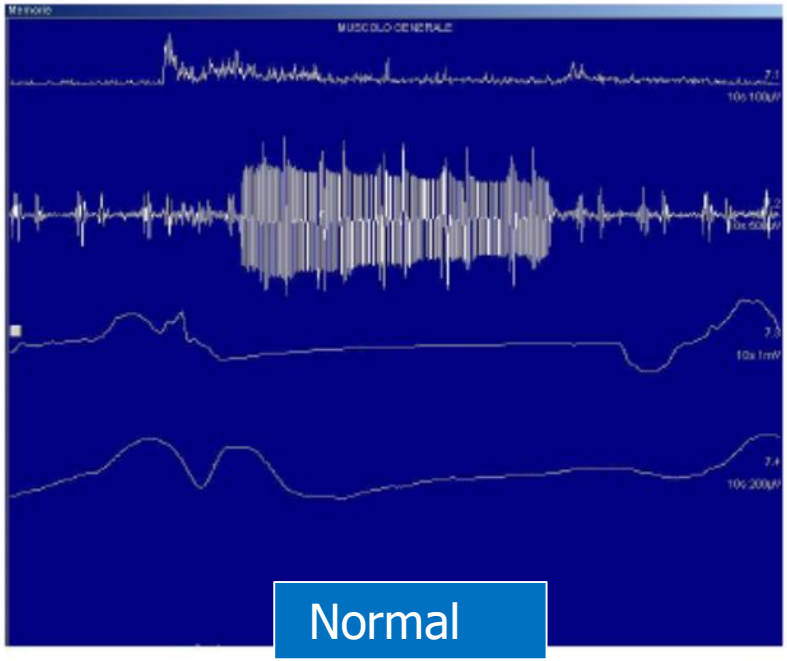
Piezoelectric transducer for nasal air flux recording during the voice

diaphragmatic belt with piezoelectric transducer for recording of breathing during voice output

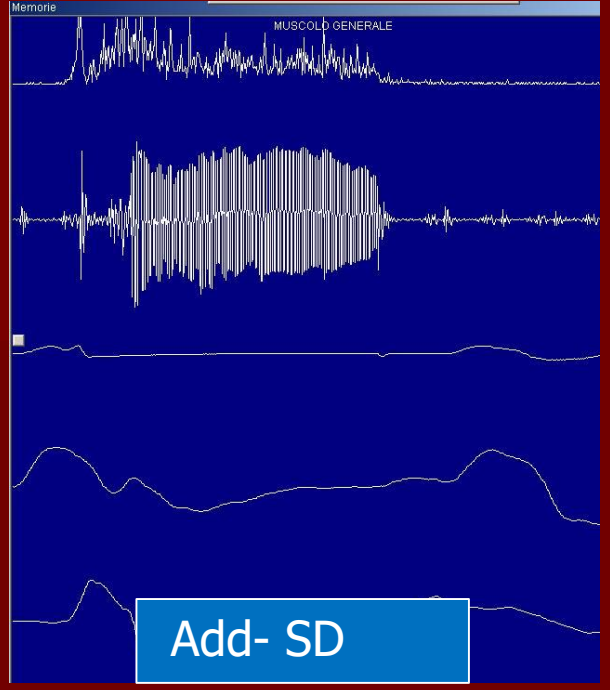
Electrokynesiographic investigation of the voice (4 channels for recording phonemas)

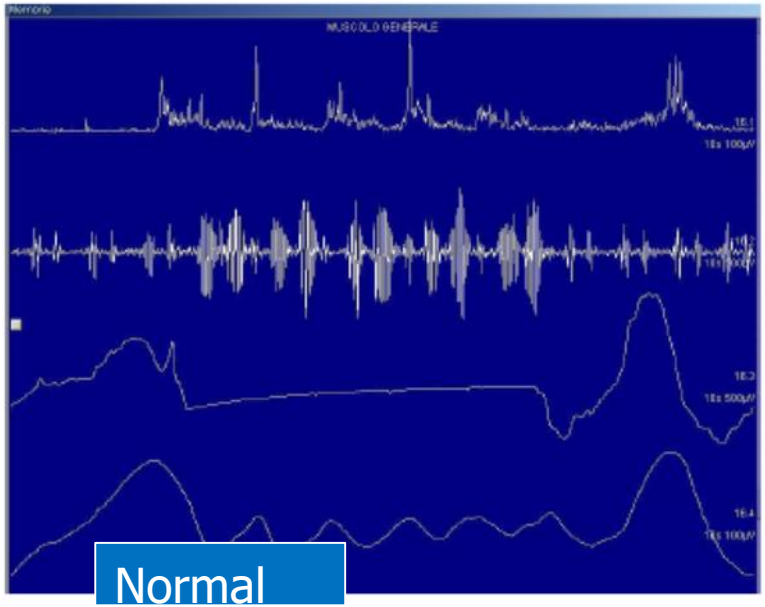


Electrokynesiographic investigation of the voice in normal for recording phonemas :
pa..pa..pa..pa..pa

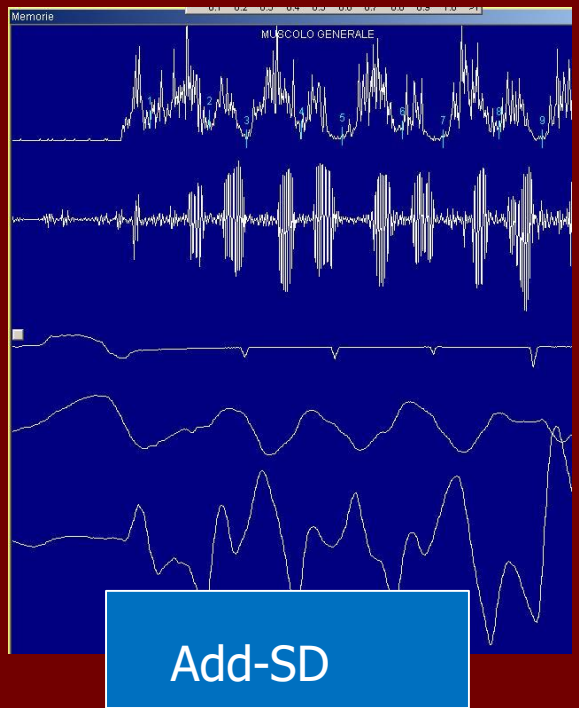


« aaaaaaaaaa»





« iu..iu..iu..iu..iu»



a) Normal

b) Abductor dysphonia

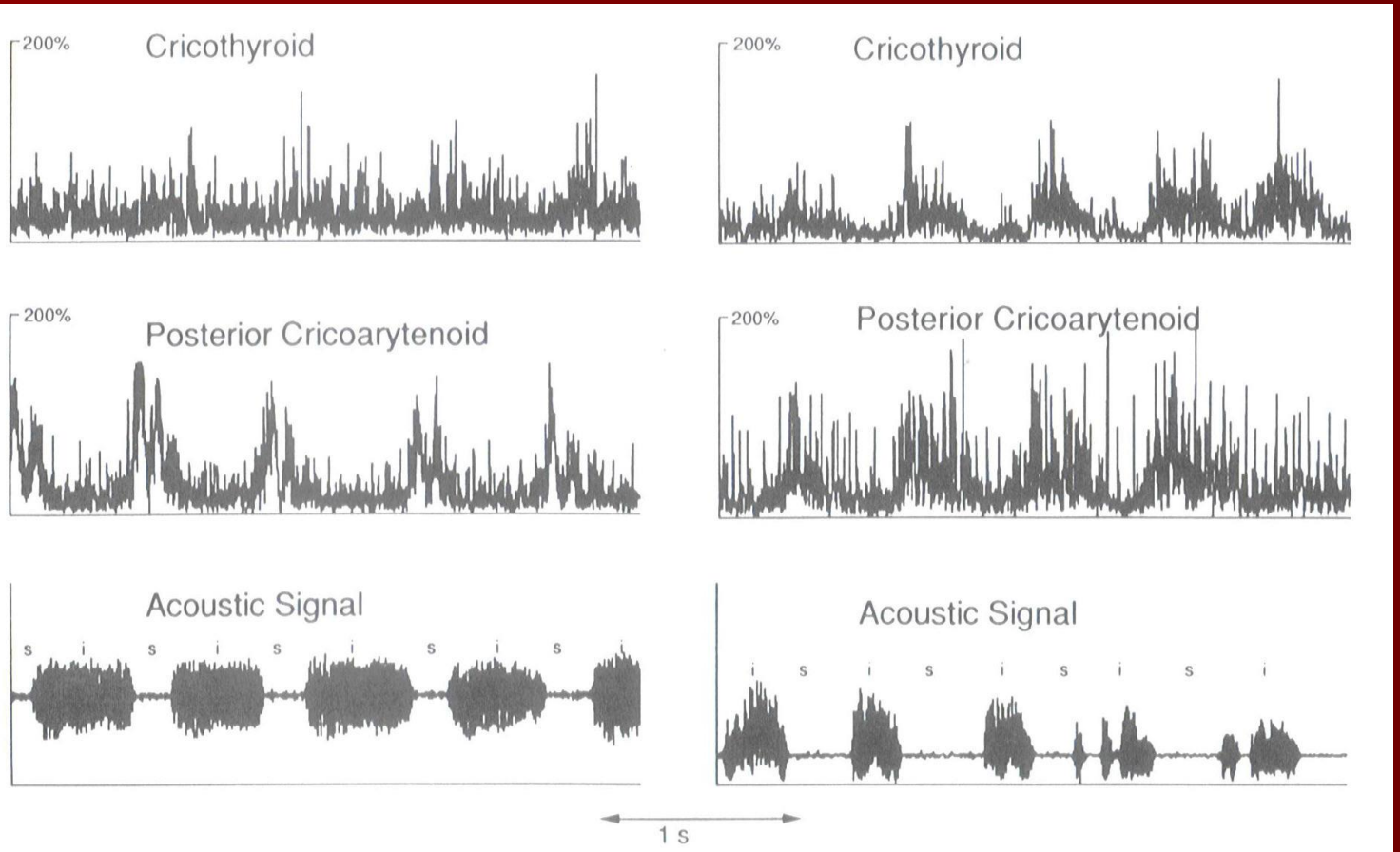


Figure 5 Prolonged bursts involving both the cricothyroid and posterior cricoarytenoid muscles during attempts at repetition of the syllable *si* in a patient with abductor spasmodic dysphonia (b) compared with a normal control (a). The prolonged bursts of these two muscles result in prolonged durations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

Adductor Spasmodic Dysphonia (SD)

The most frequent form of SD (about 85% of SD patients)

The voice is hoarse, tense with sudden explosions of sound in the intermediate vowels of a word.

These aspects are related to hyper adduction of VCs resulting in a sudden glottic closure with air flow stop which disrupts the phonation.

These changes are particularly evident in words like "eat" or phrases like

"Eels we eat every day."

Abductor Spasmodic Dysphonia (SD)

This form is less common than adductor SD (approximately 15% of patients with SD)

Voice aspirated and strained by sudden breakage of the sound emitting aspirated phonemes with persistent extreme abduction of the aspirated consonants.

The abduction of the VCs interferes with their closure and interferes with the sound of the vowel following a consonant

The voice is “breathless"

If the form is severe there are moments with complete aphonia.

Mixed Spasmodic Dysphonia (SD)

Extremely rare (0.5-1%)

The patients show symptoms of both forms of adductor and abductor SDs

The diagnosis of mixed form is important for predicting the response to treatment

Poor response to BTX treatment that often produces side effects

(the inoculum of the muscle cricoarytenoideus posterior can lead to a slight improvement of symptoms, while the inoculation of the muscle tyroarytenoideus produces mild dyspnea which can exacerbate speech disorder)

VOCAL OR LARYNGEAL TREMOR

Common symptom of several neurological disorders :
20-30% of cases of Essential Tremor
Dystonic tremors: Parkinson's disease and atypical Parkinsonisms, SD etc..

Rhythmic movements, involuntary and oscillating of VCs clinically identifiable as

"bleating voice"

Not only intrinsic muscles of the larynx but also frequently the extrinsic laryngeal muscles, pharyngeal muscles, diaphragm, palatal muscle, thoracic muscles and muscles of the abdominal wall are involved

BTX can be effective mainly in forms of SD with tremor .

Respiratory Spasmodic Dysphonia

Very rare

It's also known as laryngeal abductor –respiratory dysphonia or respiratory SD

The voice is mostly normal until the patient performs an inspiration: during this phase you can hear a sound like suffocation (transient and tight VC abduction during inspiration)

Good response to BTX treatment of the abductor muscles of VC

Annals of Otology, Rhinology & Laryngology 115(2):89-92.

Singer's Dystonia: First Report of a Variant of Spasmodic Dysphonia

Ajay Chitkara, MD; Tanya Meyer, MD; Anat Keidar, PhD; Andrew Blitzer, MD, DDS

Most patients display adductor hyperfunction in the mid-range of the singing voice with strain, fatigue, roughness, and loss of vibrato.

Some patients show abductor spasms with a breathy break in the midrange transition and occasional compensatory hyperadduction

Some patients show breaks in their speaking voice after initial presentation in the singing voice.

Treatments for SDs

- **<u>BTX injection ('gold standard therapy' for SD)</u>**

- Voice therapy (alone may be effective for mild forms of SD)

- Medical Therapy (may be occasionally effective)
  a) anticholinergic drugs (tryhexyphenidil, benztropine mesylate etc..)
  b) β-blockers non-selectives (Propanolol)
  c) Central myorelaxant such as baclofen
  d) Benzodiazepines (Diazepam, clonazepam etc..)

- Surgical interventions (selective denervation of recurrent laryngeal nerve in intrinsec laryngeal muscles; selective, thyroplasty, partial thyroarytenoid myectomy)

(This measure should be considered as the "last resort" for unresponsive and very severe cases)

What dose of botulinum toxin should SDs receive?

There is no standard BTX dose.

The dose for each patient with SD has to be customized for the patient due to the severity of SD, the patient's voice demands, and the response to BTX.

The most common doses range 1.0 - 2.5 units (IncobotulinumtoxinA, OnabotulinumtoxinA) or 3.0-8.0 units (AbobotulinumtoxinA) placed in the right and left vocal fold muscles for the treatment of AddSD.

The range for a unilateral injection for AddSD is typically from 2.5 to 10 units (IncobotulinumtoxinA, OnabotulinumtoxinA) or 8.0-30 units (AbobotulinumtoxinA)

The most common doses range 1.0 - 2.0 units (IncobotulinumtoxinA, OnabotulinumtoxinA) or 3.0-7.0 units (AbobotulinumtoxinA) placed in CAP muscles of one side for the treatment of AbdSD. No increases over these ranges.

Should SDs receive unilateral or bilateral Botulinum toxin injections?

There are no standard rules.

Most commonly, bilateral injections are done for AddSD. This is based on the notion that the disorder is bilateral and symmetric.

However, in certain patients a unilateral injection may be preferable to minimize side effects.

For AbdSD, it is typical to inject either one side only or one side with a large dose and the other side a small dose.

The patient will then return approximately three weeks following this procedure for a check, depending upon the response to the first injection in order to decide the time of the second injection

Most common complications are due to

- ***Inaccuracy of injection***
- ***Inappropriate dosing of botulinum toxin***

SIDE EFFECTS

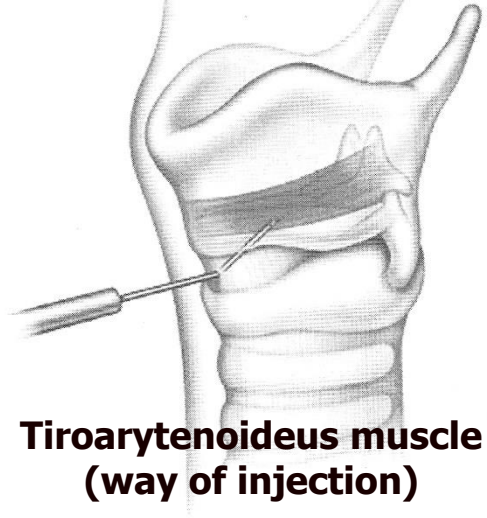
Difficulty in swallowing This is probably the most common side effect. Some temporary change in swallowing is reported by up to 17% of patients treated. In the vast majority of cases, this is more of an inconvenience than a danger.

Minor bleeding and bruising at the injection site This may occur, especially in those patients who take aspirin or blood thinners. Serious bleeding has not been reported.

There are side effects specific to each type of SD as a result of over-dosing the affected muscles

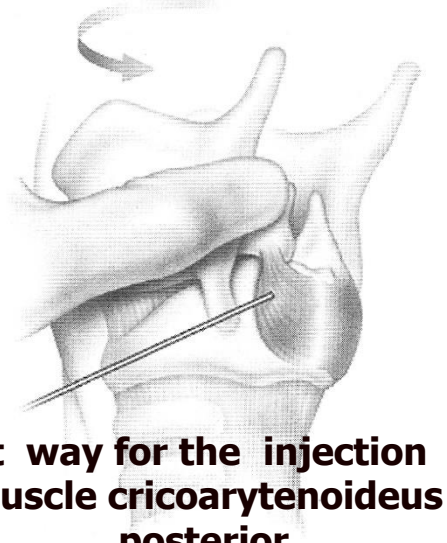
In AdSD: breathy voice and difficulty when drinking liquids. Both of these occur when the vocal folds treated are not able to come together completely

In AbSD: some breathing restriction because the treated vocal fold is not able to move aside fully.



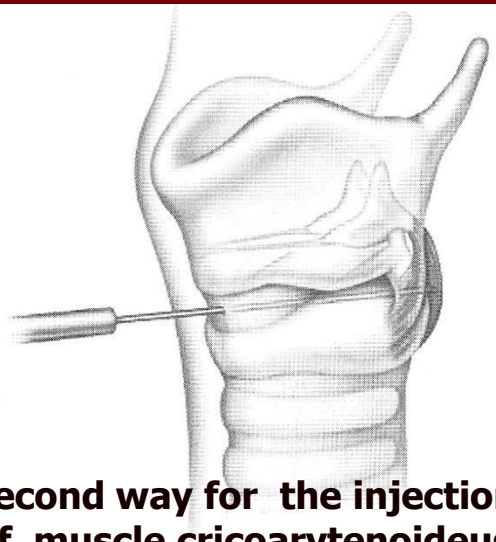
Tiroarytenoideus muscle (way of injection)

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.



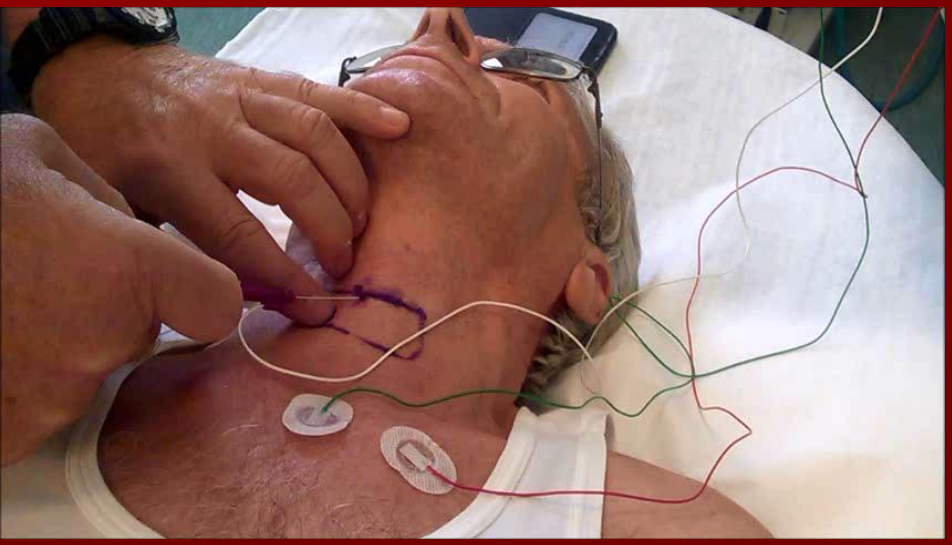
First way for the injection of muscle cricoarytenoideus posterior

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.



Second way for the injection of muscle cricoarytenoideus posterior

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

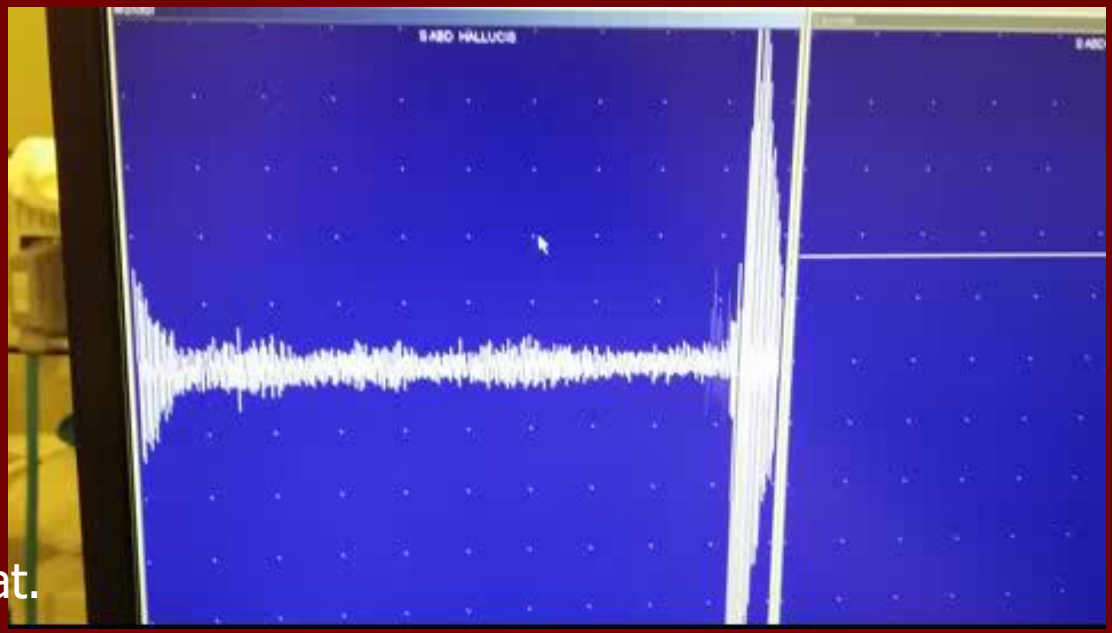


Tyroarytenoideus m.

Dystonic tremor of VC

Cricoarytenoideus lateralis m.

Muscle cricoarytenoideus lat.

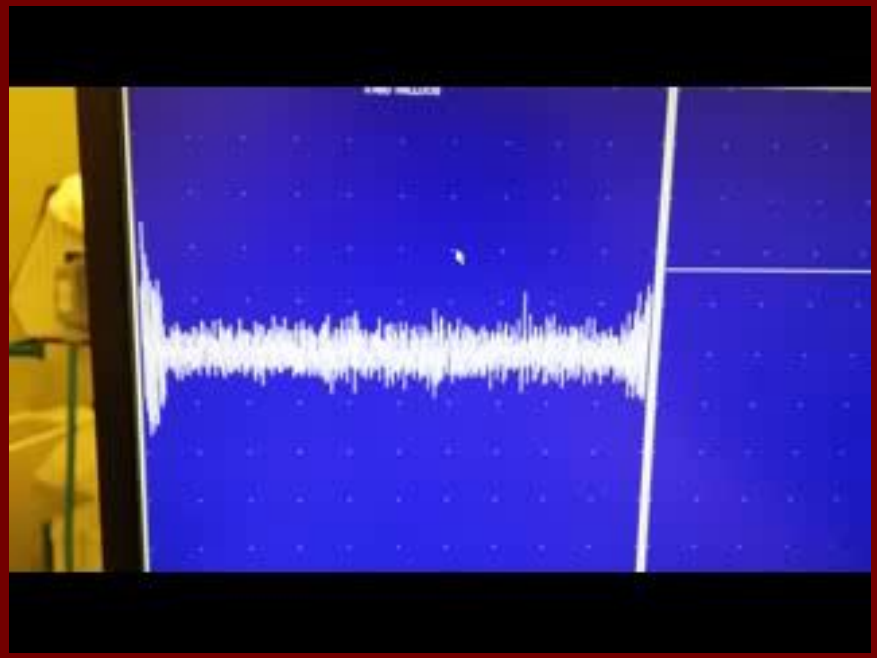


EMG of muscle cricoarytenodeus lat.

Cricoaritenoideo post.: abduttore CV

EMG Cricoaritenoideo post.

Cricoaritenoideo post.: abduttore CV
Throgh cricothyroid membrane



EMG Cricoaritenoideo post.

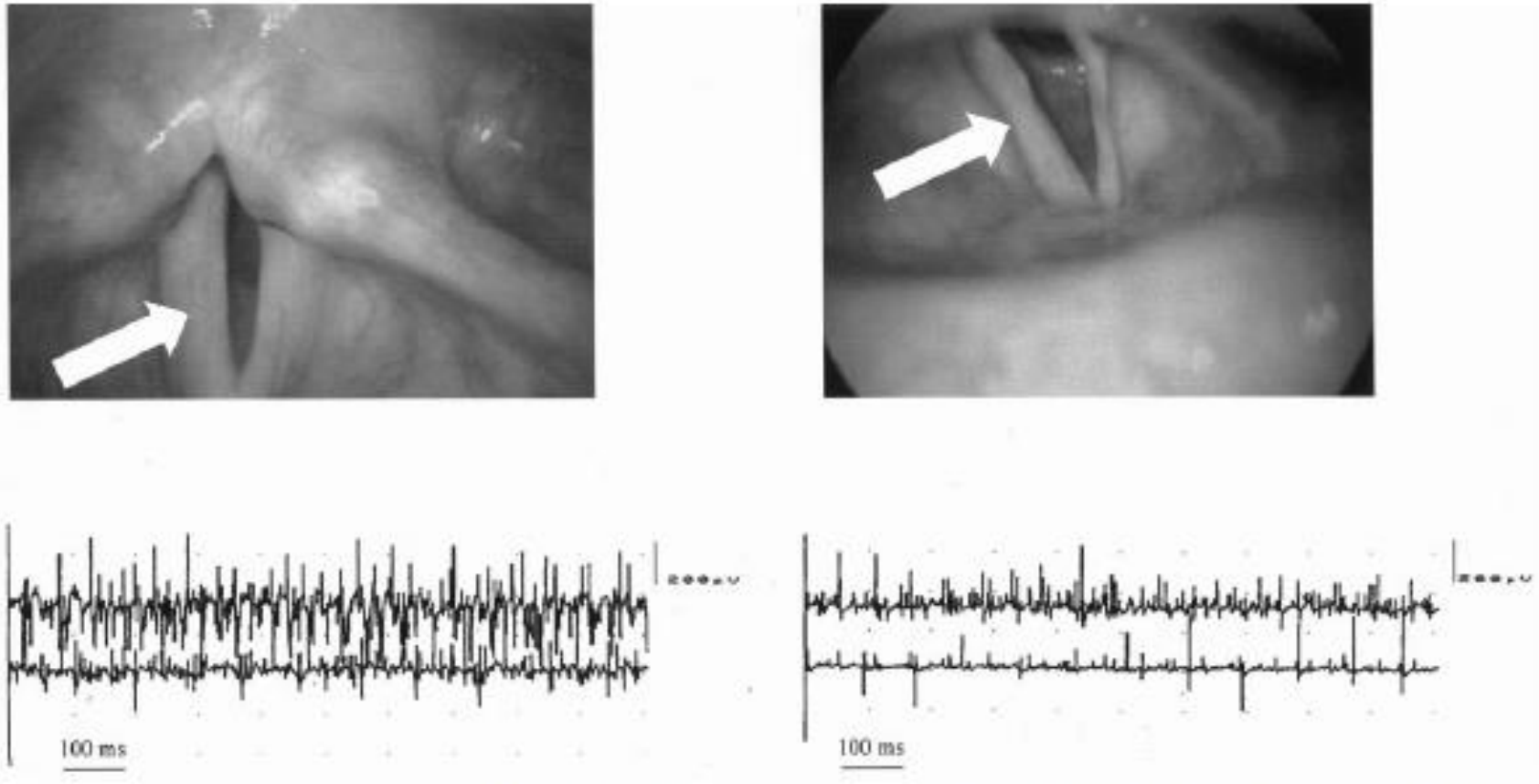


Figure. Laryngoscopy (top) and electromyogram (EMG) activity (bottom) before and after injection of botulinum toxin. Patient 2 before (left) and after (right) injection of botulinum toxin into the right thyroarytenoid muscle (arrow). Simultaneous EMG recording was performed from the right posterior cricoarytenoid muscle (top) and the right thyroarytenoid muscle (bottom). One month after injection, laryngoscopy showed the right vocal cord abducted; EMG activity of the right thyroarytenoid muscle was markedly reduced. The slight reduction of EMG activity observed in the right posterior cricoarytenoid muscle could be explained by either a "balance" mechanism (reduced activity of the antagonist muscle) or diffusion of botulinum toxin from the site of inoculation.

Not paralysis, but dystonia causes stridor in multiple system atrophy

Ilaria Maria Merlo, MD; Antonio Occhini, MD; Claudio Pacchetti, MD; and Enrico Alfonsi, MD

NEUROLOGY 2002;58:649–652

Specific patterns of laryngeal electromyography during wakefulness are associated to sleep disordered breathing and nocturnal stridor in multiple system atrophy

E. Alfonsi [a, *], M. Terzaghi [b], G. Cosentino [c], C. Tassorelli [a], R. Manni [b], N. Pozzi [d], R. De Icco [a], G. Bertino [e], M. Todisco [a], E. Alvisi [a], M. Fresia [a], C. Pacchetti [d], R. Zangaglia [d], P. Prunetti [a], A. Moglia [a]

Parkinsonism Relat Disord. 2016 Oct;31:104-109.

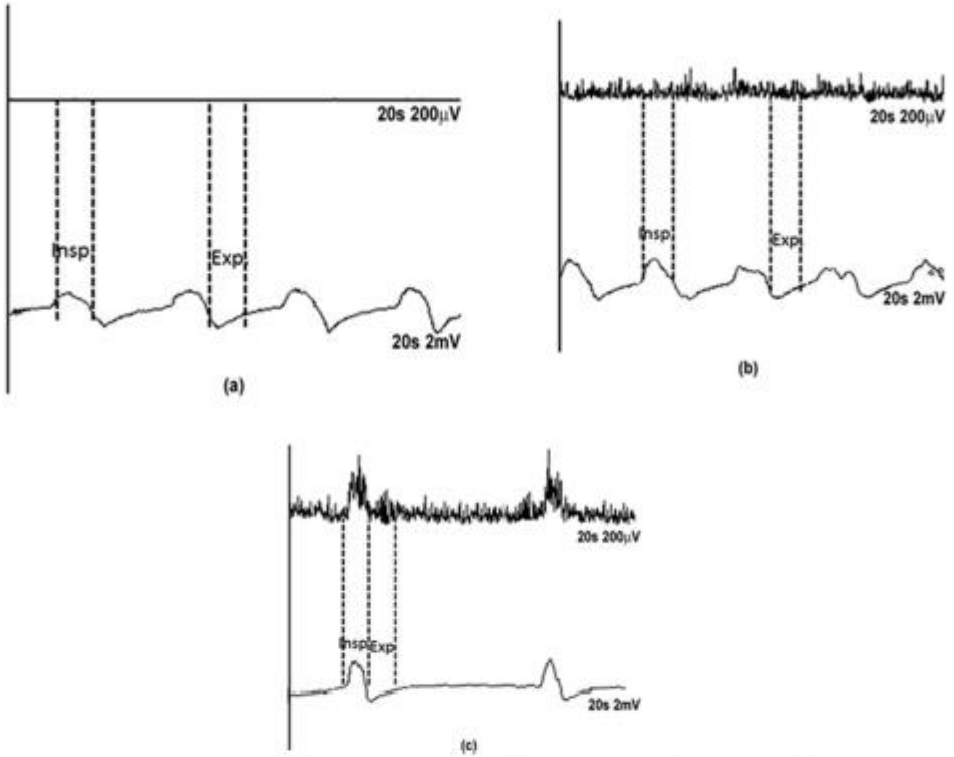


Fig. 1. Kinesiologic EMG investigation of the vocal cords: a) normal pattern; b) tonic EMG activity of thyroarytenoid muscle (adductor of the vocal cords) during quiet breathing; c) paradoxical activation during inspiration with tonic EMG activity during quiet breathing of vocal cord adductors.

Conclusions: Electromyographic/kinesiologic investigation of VC muscles during wakefulness provides additional information on the pathophysiology of the respiratory abnormalities in MSA patients that could be useful for guiding the choice of the best appropriate treatment and care.

Spastic Dysarthria

- Caused by bilateral damage to UMN
  - Degenerative disease, vascular causes, TBI, unknown

- Hypertonia, hyperreflexia, spasticity, neuropath. reflexes

- Speech: slow, effortful, may be hypernasal, imprecise artic., hoarseness, strain-strangle, monopitch/pitch breaks, short phrases, monoloud

- Spastic Dysarthria
  - Breathy, slow, monotone, monoloud, effortful, hoarse

SPASTIC DYSARTHRIA

- Slow and monotonus speech
- Rhinolalia
- Dysphagia is frequently associated
- Drooling is frequently associated
- Spastic laugh and cry
- Archaic reflexes (palm-chin, grasping)
- Global deficit of tongue motility
- Sometimes vocal cord spasms during breathing with inspiratory "tirage"
- (paradoxical adduction of vocal cords during inspiration)

SPASMODIC DYSPHONIA

- It appears or it is accentuated with emotions
- Vocal fatigue caused by an increased effort in speaking
- Movements or tensions in spasm of the body following voice output (especially in addSD)
- No abnormalities during breathing or swallowing

Thank you for your attention

Pavia: «Old Bridge»

Pavia: Old University