Diagnosis (Dx) and Treatment (Tx) of Phonatory Laryngeal Movement Disorders (PhLMD) : The Dystonia Group

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In this presentation I will
1: define phonatorykaryngeal movement disorders (PhLMD)
2: present Hx perspectives of PhLMD
3: present Dx relevant physiologic models of symptoms of PhLMD
4: discuss evidence based Tx options and outcomes
5: Provide some clinical examples
1: General Definition of PhLMD & Movement Disorders (MD)

PhLMD are a sub-group of MDs. MDs are comprise neurological disorders that involve inappropriate action(s) of motor and movement systems. Some of the more commonly known diseases include Parkinson's disease, tremors, dystonias, dyskinesias, Huntington's Chorea, and Tourette's syndrome. PhLMD are those conditions that involve disorders of voice and speech production.
In this presentation I will concentrate on how to define phonatory symptoms representative of PhLMD and dysphonias with subjectively similar sound: The discussion covers

1: laryngeal dystonias
2: dyskinesias
3: vocal tremors

I will also elaborate on efficacious Diff Diagnosis (Dx) and treatment (Tx) protocols to rule out mimicking phonatory disorders to avoid non-productive Tx, specifically I will focus on

1: MTD
2: Ventricular Dysphonia
3: Psychogenic Dysphonias.
General Definition of **Dystonia**:
Dystonia represents abnormal muscle tone accompanied by tightening, spasming, twisting of that muscle or a muscle group during action, resulting in impaired intended voluntary activity.
Dystonia can be focal, segmental or generalized, idiopathic, induced, or linked.

**Phonatory dystonia** affects muscles involved in voice production

**Speech dystonia** affects muscles involved in speech production: These can be mixed in one patient
Here are some examples of clinical manifestations of various dystonias
The Laryngeal (Phonatory type) Dystonia discussed here is the group of Voice Disorders (VoD) known as Spasmodic Dysphonia (SD). SD is sub-classified into Adductor Spasmodic Dysphonia (ADD SD) & Abductor Spasmodic Dysphonia (ABD SD). Mixed ADD/ABDSD has been proposed, but we do not agree to existence of this form. SD can be Mixed or confused with Vocal Tremor (VoT), that is not a form of dystonia.
Historical Confusion:
A VD originally described in 1871 by Traube, as a form of “hysterical dysphonia” somehow became a model for what we now consider SD. Traube suggested that SD is a psychiatric VP, though already at the beginning of 1900, a neurologic model was postulated (Rheti, 1932).

All Tx of SD (including all modalities failed for over 100 y)
The 1st successful Tx of ADDSD that involved unilateral recurrent laryngeal nerve (RLN) resection was introduced by Dedo in 1976. This Tx was based on a testable physiologic model.
Since the original Traube’s mention of “something”, any VoP that showed no laryngeal pathology and sounder “bizzare” became “a model” of SD. Consequently more than 300 semantic descriptors have been used to classify SD, all on the basis of the sound alone!!! (Izdebski, 1980)

This vast and fuzzy Dx protocol is typically blamed on that, the Dx of all types of SD is possible only by ear (Aronson et at, 1991, Chhetri, et al 2008, Dedo, 1976, Izdebski et al, 1979-2008) and that no unequivocal Dx protocols exist. However, this is not really so, moreover, the ear is a sensory system like the eye, hence there is nothing wrong in using the ear, if one knows what to listen for.
The issue is not that the Dx is by sound, the problem is that symptoms are typically tested, without regard to a physiologic model of symptoms (Izdebski, 1983), model that is capable of predicting symptoms in a systematic and reproducible way. This apparent “diagnosis by ear” need not to lead to unverifiable impressions of what is and is not SD, as long as testing is done in accordance to the principles of laryngeal phonatory biomechanics. Not doing so, only perpetuates the more than 100 y old diagnostic mess, and hence affects the Tx choices.
Symptoms, Dx&Tx Controversies

This pervasive absence of using clinically verifiable and predictable acoustic model to R/O specific MPhLDs is particularly disturbing in light of overlapping and mimicking symptoms of many VoD, hence influencing Tx types, Tx choices, and Tx outcomes, and continuously confusing outcomes.

Using the argument of changing phonation by applying treatment is not equivalent to establishing diagnostic criteria, as it is known that i.e. many modalities, including Botox are capable of effecting the workings of any muscle (Blitzer).
Therefore, having a uniform Dx Model is of utmost import to Diff Dx, as vocal symptoms overlap among many voice disorders, specifically among the ADDSD, ABDSD, muscular tension dysphonia (MTD), vocal tremor (VoT0 and PsychogeneicDysphonia (PsDy). Hence, improper Dx leads to wrong Tx.
Historical Perspective

**ADDSD**: Strangled

**ABDSD**: Breathy Voice
In the modern literature ADDSD is understood as caused by inappropriate hyper-adduction during voicing (Dedo & Izdebski, Izdebski and Dedo, 1976-2008) causing strained, strangled phonation and breaks in the specific voice range, on specific vocal laryngeal gestures, while the non-phonatory laryngeal tasks are not affected. Symptoms are not random, not representing Come & Go profile, and are eliminated or enhanced predictably using specific phonatory target stimulation tasks. To R/O SD vs mimicking VoP, we were the first to introduce task specificity, & a strict symptom taxonomy, specifying which tasks do, and which tasks do not cause, elicit or enhance symptoms. We proposed a physiologically verifiable MODEL of symptomatology (Izdebski, 1982), based on laryngeal biomechanics, to avoid erroneous Dx.
Other clinicians reports are less strict than ours in their definition of ADDSD or ABDSD symptoms, (until most recently, 2006-2008) “Sometimes, the voice will even be fluent while laughing or singing, but not during connected speech. Sulica, 2008)

Others suggest speech related tasks that do not reflect symptomatic reality (Koufman), and only recently our early suggestions are being implemented by others (Chettri, et al 2008)

This Dx controversy seems to account for the wide discrepancies regarding the outcomes of the unilateral RLN section reported in the literature. (Dedo & Izdebski, 1984, 20%), Aronson & DeSanto 1981, 40 % of failure rate)
The ABDSD, is understood as an opposite condition to ADDSD, caused by vocal folds coming apart (hyper-abduct) during connected speech. Cause is unknown for both, but brain stem (BS) models prevail. Severity is also reported to be variable from day to day, worse with strangers or on the phone, and if so, severity is physiologically driven.
OUR DIFF DX MODEL
ADDSD & ABDSD = FOCAL LARYNGEAL DYSTONIA (BASAL GANGLIA)
ADD + ABDSD
May be accompanied by Vocal Tremor*
(Cerebellum driven*)
SYMPTOMS
ACOUSTIC & VISUAL
ADHD Symptom’s Model

Faulty reading at the level of brainstem of sensory motor information from end-organ causing faulty loops and exaggerated efferent responses.
ADDSD

• Acoustic Symptoms

Overpressure and vocal arrests

Present within specific constraints dictated by laryngeal phonatory biomechanics and afferent efferent feedback loop mediated at basal ganglia levels

Symptoms are present
within 1st 1-1.5 octave only and more evident on all voiced segments
other phonatory levels normal
Vocal Tremor (VoT) not primary symptom
All other laryngeal tasks are asymptomatic
Test Protocol
Target phonatory tasks capable of eliciting systematic and predictable laryngeal responses capable of differentiating ADD vs ABD SD vs VoT vs MTD vs verteiccular dysphonia (VeD) vs PsDy
ADDSD Test Protocol

Fo Levels & intensity targets
Voiced (Early one morning... vs Voiced/Voiceless segments (He saw half a shape...)
Random Speech (The rainbow is a division of...)
Whisper Speech (Above in whisper mode)
Falsetto Speech (Above in falsetto mode)
Inspiratory Speech (above on ingressive phonation)
S/Z ratio
Non-verbal tasks: Whistle, Valsalva
Blow, Cough, Laughter
Other test: Delayed auditory feedback (DAF), RLN Block with Xylocaine
ADDSD Test Qualification

Results: No symptoms above 1.5 octave (falsetto), whisper, blowing, whistle, inspiratory speech, no symptom elimination on DAF, cough, laughter Vo, (random), S (none) Z symptomatic Unequivocal physiologic test: U RLN Block or Botox or Voice Tx
ADDSD Test Protocol

Symptomatic Targets

Voicing affected below/within 1.5 octave, on all voiced segments, z, predictably on a random sample, on DAF
Organische u Neurogen Stimmstörungen

Phonetogramm

ADD SD

No Organic Phonatory Restrictions

"Die Berufszeit" AVI // 5. bis 7. Juli 1996 // Salzburg // Austria
ALL-VOICED UN-PARALYZED

PCA

IA

TA

CT

V

500 msec
ALL-VOICED UN-PARALYZED FALSETTO

PCA
IA
TA
CT

Figure 3. Laryngeal EMG and voice traces during the subject’s reading of a segment from the all-voiced sentence in falsetto register (Mean $f_0 = 480$ Hz).
Figure 4. EMG of right laryngeal muscles and subject’s voice traces while reading a segment of the all-voiced sentence during temporary paralysis of the left laryngeal muscles (Mean $f_0 = 227$ Hz).
ADDSD

Treatment

Interruption of the feedback loop (Temp or Perm)
ADDSD

Treatment

Neuro-Destructive*
Permanent: Surgical RLN section (full or partial)
Temporary: Nerve Crush or Chemical (Botox) (outcomes may be supported by VTx)

* No other form of treatment works
ADDSD

- Treatment Outcomes
  Surgical RLN section*
  82% Treatment Efficacy
  18% Recurrence less severe <than before RLN section
  12% Breathy persistence

*(28 years of follow-up)
Negative Outcome ADDSD18% Recurrence

- Reason & Re-Treatment
- Re-innervation: No Re-animation
- Re-resection of RLN
- Laser C0₂ Thinning
- Botox
- VoTx

*(28 years of follow-up)*
# Recurrence After L RLN Section and Telfon Overinjection

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TREMOR VS ADDSD

A

B

C

SDm

SDf

SDf
Definition of LMD causing VP.

We define LMD as having neurologic or physiologic etiology.
Definition of LMD causing VP.

We define LMD as having neurologic or physiologic etiology.
HIGH TO LOW GLISSANDO

/spastic dysphonia/

TREMOR
Negative Outcome ADDSD
12% Breathiness
- Reason
Poor Medialization (Glottic Gap)
Tx of Breathiness: VoTx, Medialization or both
*(28 years of follow-up)*
ABDSD Model
Unclear (?)
Target Tasks
Voiceless-Voiced
“He….” =>F0 drop
ABDSD Tx Model

Tx frustrating and unclear (?)

PCA => Poor outcomes

CT=>Fo drop =>Better

Best =>TA +CT Muscle den + load

(Koufman => Best outcome per our experience)
Tremor (VoT)
CNS site
VoTTx
1: Botox
2: Load increase
Mixed ADDSD + VoT
ADDSD gone
VoT reduced
(RLN or Botox)
Botox Rationale
Denervation that avoids permanent paralysis
(Full or Partial)
(100 % failure 3-6 months)
Long Term Dx
Botox Controversies
Dosage
Methods
Location
Botox Controversies

Dosage

From .5 (Bilat) to 5-20 (Unilat)
Botox Controversies

Methods

Monitoring (Visual, EMG) vs No Monitoring
Botox Controversies

Location

Unilat vs Bilat

TA (ADD)

PCA (ABD) / CT

2 x CT / 1 TA (VoT)
"Why not to do Bilateral Injection and why to use L-EMG Monitoring during Chemical Denervation of the Laryngeal Muscles in ADD-SD"

Raul Mata Cruz, MD
Oakland, CA, Kaiser Permanente Medical Center

Krzysztof Izdebski, FK, MA, PhD, CCC-SLP, FASHA
Pacific Voice & Speech Foundation, SF, CA
Our Rationale

- Unilateral technique
- L-EMG monitoring
Hypothesis
ADD-SD is mediated equally bilaterally, hence unilateral denervation is sufficient

Izdebski, Journal of Voice
1992
Proof
L-EMG, physiologic and acoustic studies of VF activity pre & post block of RLN

Shipp, Izdebski, Reed, Morrissey, J Hearing & Speech Disorders 1985
R ILM PRE L-RLN Block

R ILM POST L-RLN Block

Figure 4. EMG of right laryngeal muscles and subject’s voice traces while reading a segment of the all-voiced sentence during temporary paralysis of the left laryngeal muscles (Mean $f_0 = 227$ Hz).
Proof

Unilateral restoration of nl L-EMG signal after blocking the contralateral side

Shipp, Izdebski, Reed, Morrissey, J Hearing & Speech Disorders 1985
Furthermore

- Acoustic studies of voice quality after RLN block of either side gave the same result regardless of sidedness.
- As the temporary paralysis recovers, the voice changes from breathy and aperiodic to normal and then to spasmodic.

Shipp, Izdebski, Reed, Morrissey, J Hearing & Speech Disorders 1985

Proof
Clinical results from unilateral L or R RLN sectioning

Izdebski, Shipp, Dedo, Otolaryngol H&N Surgery 1981
Dedo, Izdebski, Ann Otol Rhinol Laryngol. 1984
Unilateral Injection

Conclusions

- Sufficient to relieve spasmodic symptoms
- Results in restored normal activity in the opposite VF- minimizing breathiness, aperiodicity & aspiration risks
Bilateral Injection

Conclusions

- Relieves spasmodic symptoms
- But results in a larger, flaccid glottic gap, resulting in:
  - Weaker/breathier voice
  - Higher aspiration risk
- Anecdotal patient reports confirm
Bilateral Injection
Other Considerations

- 2x Patient discomfort
- 2x Injection related complications
- 2x Treatment failures
L-EMG vs Touch-Point Localization

- Lack of prospective comparative data
- Which gives most objective evidence of correct placement?
- Esp. important with bilat. injection
• Touch-Point localization
  Anatomical & tactile
  Fiberoptic visual?
  2-3 data points

• L-EMG localization
  Anatomical & tactile
  Electrophysiologic (high pitch, low pitch, valsalva, inspiration)
  6 data points
Conclusions

- Unilateral injection is sufficient, safe & superior technique
- L-EMG maximizes target hit rate based on the redundancy of confirmatory information prior to injection
For further questions regarding this presentation please contact Dr. Izdebski at kizdebski@pvsf.org or go to www.pvsf.org