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Hypokinetic Dysarthria in Parkinson's Disease: Current Insights and Clinical Implications

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Learning Objectives

After this course, participants will be able to:

1. List factors that may contribute to hypokinetic dysarthria in speakers with Parkinson's disease.
2. Describe at least two challenges for the assessment of speech in individuals with Parkinson's disease and how these challenges can be addressed.
3. List at least two behavioral modifications that are evidence-based treatment options for hypokinetic dysarthria in speakers with Parkinson's disease without deep brain stimulation (DBS).
4. Describe the impact of deep brain stimulation (DBS) on speech in speakers with Parkinson's disease.
5. Describe how behavioral modification treatments can be modified for speakers with Parkinson's disease and deep brain stimulation (DBS).

Overview of Presentation

- Introduction
- General overview: Hypokinetic dysarthria and Parkinson's disease
- Article 1: Factors that may contribute to hypokinetic dysarthria in speakers with Parkinson's disease (Sapir, 2014)
- Article 2: Behavioral modifications as treatment approaches for hypokinetic dysarthria in speakers with Parkinson's disease (Tjaden et al., 2014)
- Article 3: The impact of DBS on speech in persons with Parkinson's disease and treatment options (Tripoliti et al., 2011)
- Conclusions
- Questions & Answers

Hypokinetic dysarthria

- Darley, Aronson, & Brown, 1969
  - Reduced vocal loudness and vocal decay (hypophonia)
  - Hoarseness, harshness, breathiness
  - Reduced prosodic pitch and loudness inflection
  - Imprecise consonants and distorted vowels
  - Short rushes of speech (festinations)
  - Hesitation
  - Dysfluency
  - Voice tremor
- Is hypokinetic dysarthria special?
  - Dysarthria commonly associated with slowed speech, but speakers with hypokinetic dysarthria can exhibit an accelerated speaking rate and festinations
  - Dysfluencies typically not observed in other types of dysarthria

Parkinson's disease

- Parkinson's disease is on the rise worldwide
  - In 2005 4.1 million people with PD
  - 2030 predicted 8.7 million people
  - ~1.5% of individuals 60+ years old
  - In the future, more people will have PD for longer
  - Up to 90% of people with PD will develop dysarthria

  - Not only in older adults, some forms of PD begin in the 20s and 30s
  - 10-20% of all diagnosed cases of PD are under age of 50
  - Rate of progression is slower in young onset PD

[e.g., Sapir, 2014; Parkinson's Foundation retrieved at pdf.org; American Parkinson's Disease Association, National Young Onset Center retrieved at youngparkinsons.org]

Article 1: Factors contributing to hypokinetic dysarthria in talkers with PD

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Review Article

Multiple Factors Are Involved in the Dysarthria Associated With Parkinson's Disease: A Review With Implications for Clinical Practice and Research

Shimon Sapir[a]

[Sapir, S. (2014). Multiple factors are involved in the dysarthria associated with Parkinson's disease: A review with implications for clinical practice and research. *Journal of Speech, Language, and Hearing Research, 57*, 1330-1343]

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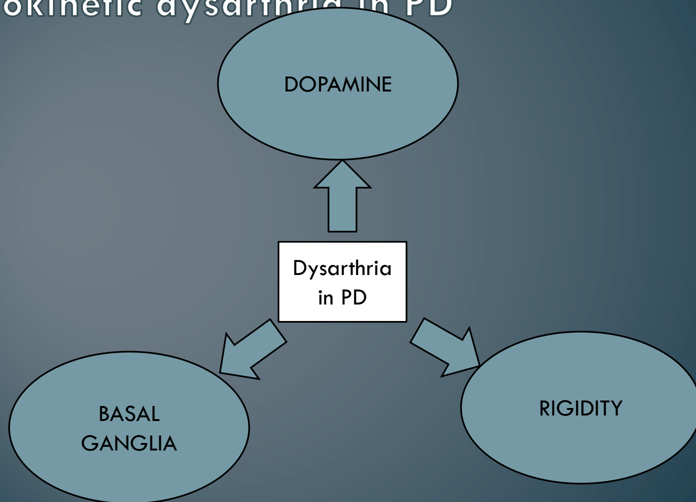
Factors commonly considered as contributors to hypokinetic dysarthria in PD



Basal Ganglia Pathology

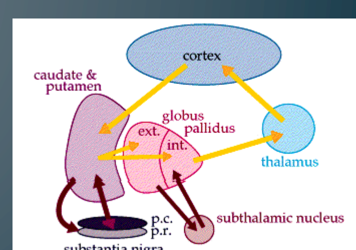
- Striatum (caudate nucleus & putamen)
- Globus pallidus (external and internal)
- Subthalamic nucleus
- Substantia nigra (pars compacta and pars reticulata)

Loss of dopamine in striatum results in adaptive responses in GP, STN, SNpc, and ultimately thalamus

Classic signs of PD

rest tremor
rigidity
bradykinesia
hypokinesia
postural instability

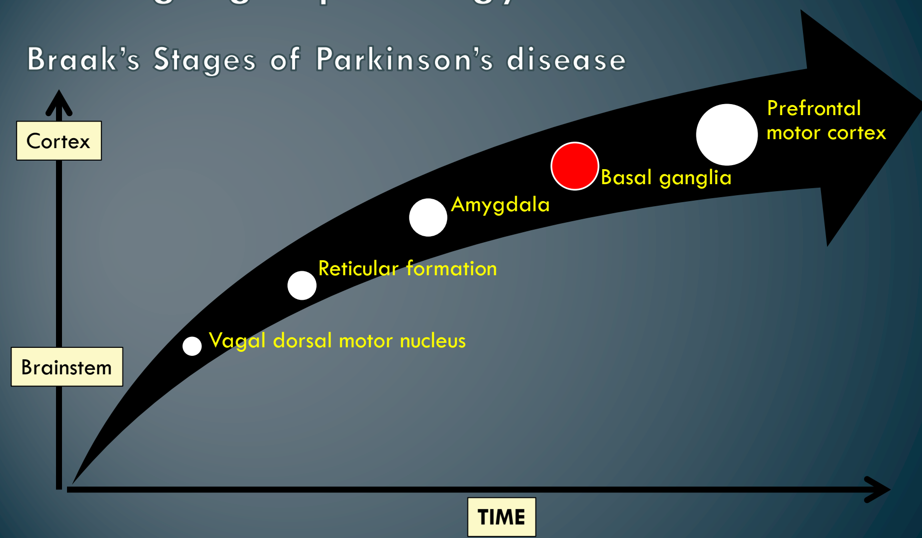
http://dcadmh-loosescrews.wikispaces.com/2.+Overiew+of+Parkinson%27s+Disease



http://www.bioon.com/bioline/neurosci/course/cerebell.html

Is hypokinetic dysarthria explained by basal ganglia pathology?

Braak's Stages of Parkinson's disease



[e.g., Burke et al., 2008]

Is hypokinetic dysarthria explained by dopamine loss?

- Mixed findings on changes in voice/speech in response to dopamine treatment
  - Voice and speech changes may be too subtle to be detected
  - Dopamine might be used up for limb movements

[e.g., Sapir, 2014]

Is hypokinetic dysarthria explained by muscle rigidity?

- Definition
  - Resistance to passive movement throughout the range of motion
  - Abrupt, transient reduction in resistance (cogwheel phenomenon)
- Rigidity has been rarely examined in orofacial muscles
  - Lip stiffness correlated with movement amplitude (Hunker et al., 1982)
  - Lip stiffness did NOT correlated with movement amplitude (Caligiuri, 1987)
- In the limb system, hypokinesia and bradykinesia are conceptualized separately from limb rigidity

[e.g., Caligiuri, 1987; Hunker, Abbs, & Barlow, 1982]

Is hypokinetic dysarthria explained by muscle rigidity?

- Stimulability and task-dependent performance do not support rigidity as a contributing factor to hypokinetic dysarthria
  - Loud speech
  - Complexity of speech task (short sentence repetitions vs. conversational speech during dual tasking)

[e.g., Connor & Abbs, 1991]

Possible factors and interactions contributing to hypokinetic dysarthria in PD

- **Scaling amplitude**
- **Internal cueing**
- **Sensory processing**
- Temporal processing
- **Automaticity**
- Emotive prosody
- **Vocal vigilance**
- Initiation
- Pre-programming

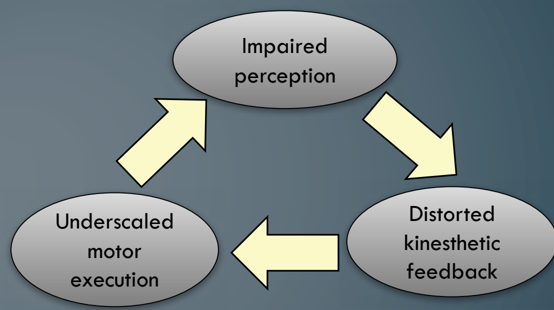
[Sapir, 2014]

Sensing, scaling, and sustaining effort

- Significantly higher self-ratings of effort for general daily activities and for speech tasks compared to healthy controls
- Handgrip and tongue elevation constant-pressure tasks showed faster decay curves in PD group compared to controls
- May in part explain hypokinesia/movement decay and hypophonia/vocal decay in speech and speech-like tasks

[e.g., Solomon & Robin, 2005]

Simplified hypothetical mechanism underlying hypokinetic dysarthria in PD



Impaired perception of motor performance provides wrong kinesthetic feedback, which results in underscaled motor execution

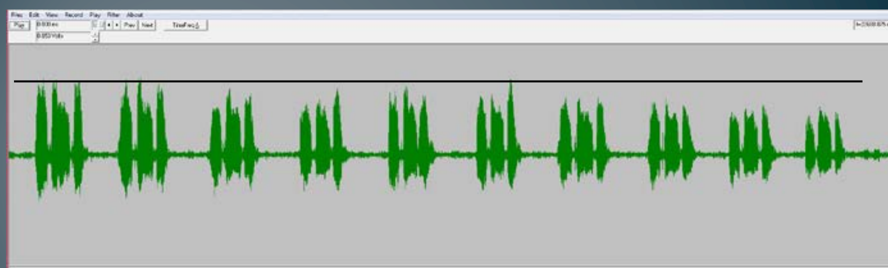
[Sapir, 2014]

Scaling amplitude

- Underscaling and decay of force in the limb system
  - Shuffling gait
  - Micrographia
  - Reduced grip forces
- Underscaling and decay of force in the speech system
  - Respiratory drive
  - Vocal fold excursion
  - Orofacial movements

[see Sapir, 2014 for review]

Example of mild vocal decay during the early stages of speech deterioration



Internal cueing

- In the limb system
  - External cueing improves stride length, size of letters in handwriting tasks
- In the speech system
  - Ability to adjust vocal loudness with explicit instructions (speak louder!), but not able to adjust vocal loudness independently
- In cognitive tasks
  - Difficulty with set-shifting when tasks completed without external cues, but normal performance with external instructions

[see Sapir, 2014 for review]

Sensory processing

- In the limb system
  - Poor integration of cortical, cerebellar, and basal ganglia information in the thalamus
  - Poor conscious perception/ elevated sensory threshold
- In the speech system
  - Reflex responses to auditory feedback perturbation (pitch-shift and loudness shift perturbations) abnormally strong
    - Dysfunctional mechanisms of error detection and correction in auditory feedback
  - Misperception of their own vocal loudness
    - Reporting their own voice as too loud when cued to speak louder to reach *normal* loudness level

[see Sapir, 2014 for review]

Summary and implications for clinical practice

- Currently mechanisms of hypokinetic dysarthria is only poorly understood
  - Questionable if muscle rigidity and dopamine loss can explain speech deficits
  - Voice and speech changes can occur before limb motor symptoms are noticeable; however, special instrumentation may be needed to detect these changes (acoustic and kinematic analysis)
- Abnormal sensory processing, increased attention to action, and disrupted internal cueing mechanisms may be related to hypokinesia and movement decay

Summary and implications for clinical practice

- Speech assessments are challenging because…
  - Speech performance can fluctuate depending on required linguistic complexity and competing motor tasks
    - Speech evaluation in controlled, quiet environment may not provide insight in every day functioning
  - Assessment of various speech conditions
    - Dual tasks such as walking and talking
    - Automatic speech task (counting, days of the week,…) vs. paragraph reading, picture description, conversation
- Early onset and subtle speech changes may be difficult to detect when using speech intelligibility measures and speech severity ratings
  - Integration of smart phone apps in speech assessments to measure SPL, pitch range during speech task, duration of breath groups,
  - Recording and acoustically analyzing speech samples (free downloadable software PRAAT or wavesurfer)
    - Vowel space area
    - SPL meter, Voice test (F0, jitter, shimmer)

Summary and implications for clinical practice

- Speech treatments may be most effective if
  - they address internal cueing deficits, perception of own speech output, and attention to speech production
  - practice is intense
  - they address all speech systems = global approach

[Sapir, 2014]

Speech behavioral modification as treatments for talker with PD and hypokinetic dysarthria

- It is currently unknown, which behavioral modification is most effective
- Little guidelines for clinicians

Article 2: Clear, loud, or slow speech?

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Research Article

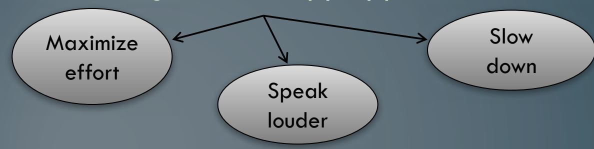
Impact of Clear, Loud, and Slow Speech on Scaled Intelligibility and Speech Severity in Parkinson's Disease and Multiple Sclerosis

Kris Tjaden,[a] Joan E. Sussman,[a] and Gregory E. Wilding[a]

[Tjaden, K., Sussman, J.E., & Wilding, G. (2014). Impact of clear, loud, and slow speech on scaled intelligibility and speech severity in Parkinson's disease and multiple sclerosis. *Journal of Speech, Language, and Hearing Research, 57*, 779-792]

Global therapy approaches for hypokinetic dysarthria in speakers with IPD

- What is a global therapy approach?



- Does not focus on specific speech sounds
  - Across the time domain (ongoing)
- Does not focus on a specific speech component
  - Across subsystems (respiration, phonation, resonance, articulation)

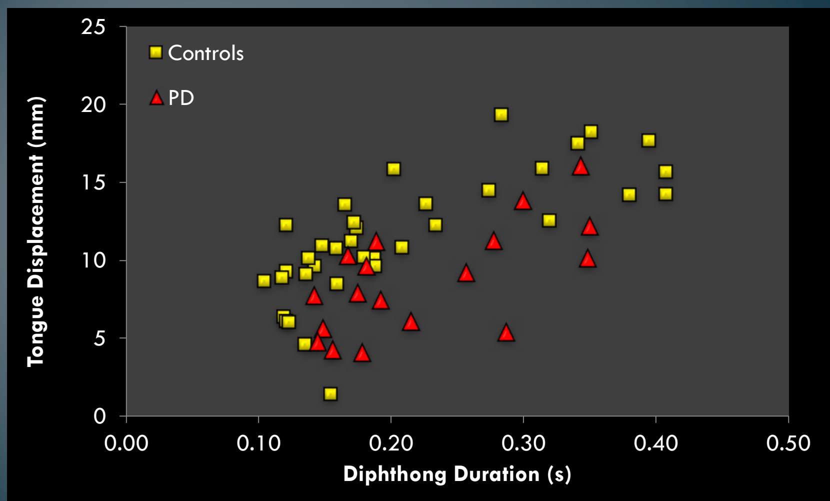
[Tjaden et al., 2014]

How do global therapy approaches work?

- Rationale for slow speech
  - May give the speaker time to reach target position (e.g., tongue to palate contact)
  - May enhance coordination among speech subsystems (phonation/articulation)
  - May give the listener time to process the distorted acoustic signal
  - May mark word boundaries better (e.g., lexical segmentation) for listener

[see Tjaden et al., 2014 for review]

Some data



[Mefferd, 2015]

How do global therapy approaches work?

- Rationale for loud speech
  - Positively impacts vocal quality and the range of F0 (intonation patterns)
  - Improves articulatory behavior (larger articulatory excursions)
  - May positively impact audibility for listener

[e.g., see Tjaden et al., 2014 for review]

How do global therapy approaches work?

- Rationale for increased effort (clear speech)
  - Hyperarticulation, or increased articulatory effort, has been shown to elicit **slower and louder** speech
  - May provide combined benefits of slow/loud speech in clear speech approach with specific focus on articulation

[see Tjaden et al., 2014 for review]

Impact of clear, loud, and slow speech on perceptual measures

[Tjaden, Sussman, & Wilding, 2014]

- Purpose of research study
  - To compare the effect of rate reduction, increased loudness, and clear speech (high articulatory effort) on intelligibility and speech severity in individuals with PD and Multiple Sclerosis (MS)

Impact of clear, loud, and slow speech on perceptual measures

[Tjaden, Sussman, & Wilding, 2014]

- 78 speakers
  - 32 controls
  - 16 speakers with PD
  - 30 speakers with Multiple Sclerosis (MS)
- Task
  - Read sentences at habitual, slow, loud, and clear speech
  - Sentences contained 7-9 words
- 100 Listeners
  - 50 listeners scaled speech intelligibility
  - 50 listeners scaled speech severity
- Task
  - Listen to sentences in multi-babble noise and scale intelligibility/severity

Impact of clear, loud, and slow speech on perceptual measures

[Tjaden, Sussman, & Wilding, 2014]

- Results – Task Performance
  - Loud speech increased SPL levels 7-10db
  - Clear speech increased SPL levels 3-4dB
  - Slow speech elicited 49-29% rate reduction
  - Clear speech elicited 19-37% rate reduction
  - Loud speech did not elicit rate reduction in the PD group

Impact of clear, loud, and slow speech on perceptual measures

[Tjaden, Sussman, & Wilding, 2014]

- Results – Task and Group Effects on Speech Intelligibility
  - Speakers with PD: loud = clear > habitual = slow speech
  - Speakers with MS: loud = clear > habitual = slow speech
  - Control Speakers: loud = clear > habitual > slow speech
- Results – Speech Intelligibility and Speech Severity
  - High correlations between scaled speech intelligibility and scaled speech severity for speakers with PD and MS during habitual speech
  - Correlations were significantly weaker for other speaking conditions

Discussion

- Improvements in intelligibility were about 7-11% in response to clear/loud speech
- Speakers with mostly mild speech impairment were studied
- Does this approach work as well for more severe speakers?
  - Future studies are needed
- Why does slow speech not work for mild dysarthria?
  - Perhaps negatively affects prosodic patterns? Disrupts the coordination of articulatory movements and coarticulation?
- Why does clear/loud speech work for mild/moderate dysarthria?
  - Segmental changes (increased articulator/acoustic contrast), vocal quality, enhanced prosodic features (pitch range)
    - Segmental changes: clear > loud speech
    - Vocal quality/prosodic features: loud >clear

[Tjaden et al., 2014]

Summary and conclusions

- Speakers with mild dysarthria can improve speech intelligibility using loud or clear speech
- Slow speech appears to negatively impact speech intelligibility in speakers with mild/moderate dysarthria due to PD and MS
- Caution: these findings are based on intelligibility ratings in multi-babble noise

[Tjaden et al., 2014]

Article 3: Speech treatments for speakers with IPD and deep brain stimulation

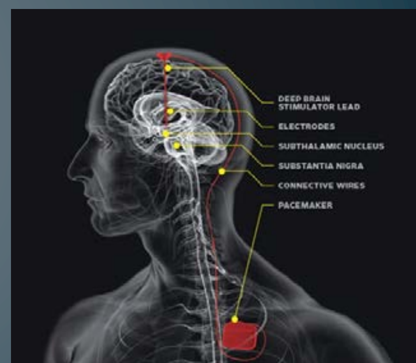
Treatment of dysarthria following subthalamic nucleus deep brain stimulation for Parkinson's disease

Elina Tripoliti, **Laura Strong**, **Freya Hickey**, **Tom Foltynie**, **Ludvic Zrinzo**, **Joseph Candelario**, **Marwan Hariz**, and **Patricia Limousin**
UCL, Institute of Neurology, Sobell Department, Unit of Functional Neurosurgery, Queen Square, London, WC1N 3BG, UK

[Tripoliti, E., Strong, L., Hickey, F., Foltynie, T., Zrinzo, L., Candelario, J., Hariz, M., Limousin, P. (2011). Treatment of dysarthria following subthalamic nucleus deep brain stimulation for Parkinson's disease. *Movement Disorders, 26*(13), 2434-2436.]

Deep Brain Stimulation – What is it?

- Electrode stimulating specific parts of the basal ganglia
  - Internal globus palidus (GPi-DBS)
  - Subthalamic nucleus (STN-DBS)



http://www.neurosurgery.uthscsa.edu/display_patients.php?ps_id=27&pg=

[e.g., Skodda et al., 2014; Tripoliti et al., 2014]

Basal Ganglia Pathways

Direct pathway – Excitatory to motor cortex

Motor cortex → Putamen → Globus pallidus internal / Substantia nigra (pars reticulata) → Thalamus → Motor cortex → **Movement**

Indirect pathway – Inhibitory to motor cortex

Motor cortex → Putamen → Globus pallidus external → Subthalamic nucleus → Substantia nigra (pars reticulata) → Thalamus → Motor cortex → **No Movement**

excites | inhibits | disinhibits

[e.g., Calabresi et al., 2014]

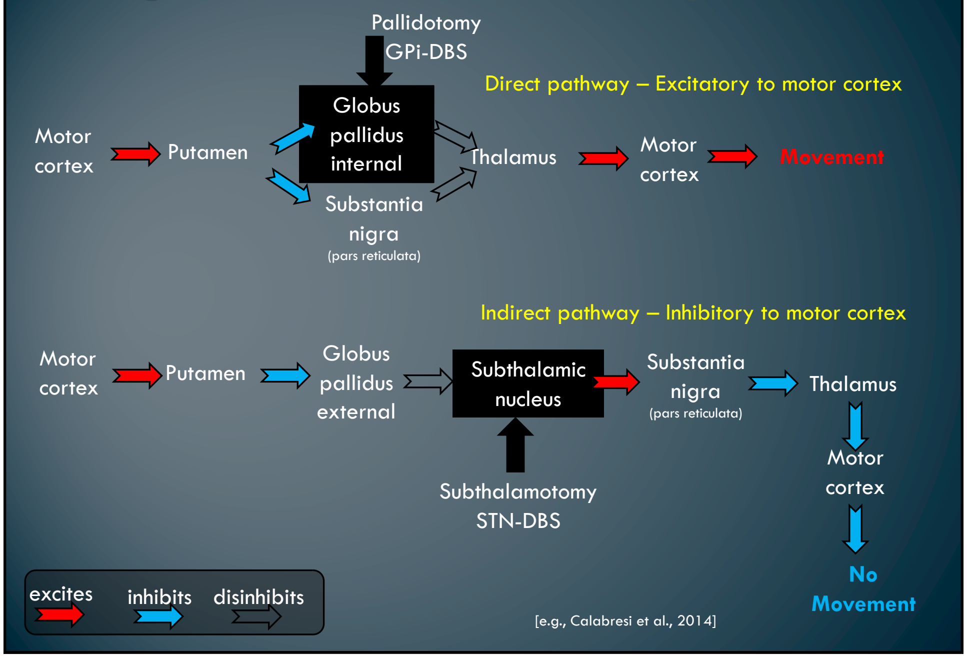
Basal Ganglia Pathways

- Substatia nigra pars compacta
  - Supplies dopamine to the basal ganglia circuits
  - Stimulation of this region does NOT result in any movements
  - Degeneration of this region has HUGE impact on movement
- Substatia nigra pars reticulata
  - Ultimately conveys the signal of the basal ganglia pathways to the thalamus (which then communicates with the motor cortex)
  - Its role is inhibitory

What happens in PD?

- Cells in the pars compacta of the substantia nigra degenerate
- Decline in dopamine supply
- Disturbed communication between BG structures
- Results in excessive inhibitory activity of the globus pallidus internal (GPi) and the subthalamic nucleus
  - Bradykinesia
  - Hypokinesia

PD surgical treatments - How do they work?



Does DBS affect speech?

- DBS effects on speech vary from person to person
  - 10-20% of patients with STN experience worsening of speech
  - Others showed that UPDSR speech score showed no significant change under STIM-ON condition vs. STIM-OFF condition, but there was a trend of worsened speech under STIM-ON vs. STIM-OFF
  - In some patients with PD improved articulation was noted under STIM-ON vs. STIM-OFF

[e.g., Barbe et al., 2014; Flora et al., 2010; Muecke et al., 2014; Skodda et al., 2014]

DBS-related speech deterioration

- Patient-related risk-factors for worsening of speech after DBS:
  - Longer disease duration
  - Overall more severe limb and speech motor symptoms
  - Need for strong stimulation in the left hemisphere
  - Electrode placement within the medial and/or posterior portion of the STN on the left side

[e.g., Tripoliti et al., 2014]

Treatment options for dysarthria in speakers with DBS

[Tripoliti et al., 2011]

- Rationale for the study
  - Surgical treatment does not improve speech intelligibility
  - Behavioral speech treatment is needed for patients with PD and DBS
  - What to do?!
- Research question
  - How effective is LSVT® Loud for patients with DBS?
- Methods
  - 10 patients with PD with bilateral STN-DBS
  - 10 patients with PD with medication intake
  - Pre/post treatment design and 6-month follow up

LSVT® – standardized exercise for PD

- Exercise regimen to slow the progression of motor symptoms and neural degeneration
- Targets movement amplitude, recalibration of the sensorimotor system, self-cueing, and attention to motor action
- LSVT® is consistent with principles of activity-dependent neuroplasticity
  - Specificity – targeting hypokinesia (movement amplitude)
  - Intensity – ramping up dosage of treatment
  - High repetition rate – mass practice
  - Saliency of treatment tasks – translation of structured motor exercise into daily functional activities
  - Lasting treatment outcomes – recalibrate motor and perceptual systems to help self-cueing

[e.g., Fox, Ebersbach, Ramig, & Sapir, in press]

Treatment options for dysarthria in speakers with DBS

[Tripoliti et al., 2011]

- Tasks
  - Sustained phonation /a/ for three repetitions
  - Sentence Intelligibility Test
  - 60-seconds monologue
- Measures
  - SLP dB
  - Intelligibility
  - Perceived severity ratings for six speech characteristics clusters (based on Darley, Aronson, & Brown, 1975)

Treatment options for dysarthria in speakers with DBS

[Tripoliti et al., 2011]

- Results
  - Sustained phonation
    - Significant increase in vocal loudness in medication group only from pre-treatment to follow-up session
  - Sentence Intelligibility Test
    - No sign. changes in speech intelligibility from baseline to post-treatment or follow-up in either group
      - Medication group went from 95% to 98%
      - DBS group went from 89% to 83%
  - Monologue
    - Improved respiratory, phonatory, and overall perceptual ratings for post-treatment and at 6-month follow-up **in medication group only**
    - Deterioration of respiration in the DBS group post-treatment and at 6-month follow-up

Treatment options for dysarthria in speakers with DBS

[Tripoliti et al., 2011]

- Further findings:
  - Common speech characteristics of persons with DBS
    - Strained-hoarse voice
    - Excess loudness
    - Monoloudness
    - Monopitch
    - Reduced stress
    - Imprecise consonants
    - Distorted vowels
    - Insufficient breath support →Short breath groups

Treatment options for dysarthria in speakers with DBS

[Tripoliti et al., 2011]

- Discussion
  - Use of LSVT® for speakers with PD who are treated with medication supported by findings of study
  - Not so much for patients treated with DBS
- Why does LSVT® appear not to work for patients with DBS?
  - Different pathophysiology of dysarthria
  - Different sensory processing?
  - Different ability to learn new motor skills?
  - Spread of current to non-motor areas possibly negatively impacting neighboring areas in the brain
- Limitations
  - Small number of participants in both groups

LSVT ®Loud for PD following STN-DBS

[Spielman, J. et al., 2011]

- 12 participants with PD
  - 4 with bilateral STN-DBS
  - 8 with medication
- Tasks
  - Sentence repetitions
  - Paragraph reading
  - Monologue
- Measures
  - vocal intensity
  - Vowel articulation index (VAI)
  - Voice Handicap Index

LSVT ®Loud for PD following STN-DBS

[Spielman, J. et al., 2011]

- Results
  - Both groups of participants improved in response to LSVT®Loud
    - Vocal intensity
    - Vowel articulation index
    - Voice Handicap Index (but PD-DBS group rated themselves worse before and after tx compared to medicated group)
- Discussion
  - Findings are interpreted as evidence of the effectiveness of LSVT®Loud for speakers with PD with and without STN-DBS

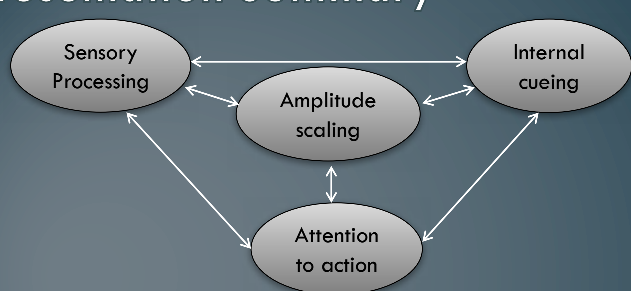
What to do with patients with DBS?

- Assess speech characteristics
  - If consistent with hypokinetic dysarthria begin trial of behavioral therapy (loud/LSVT®)
- Ways to modify speech therapy for patients with PD and DBS
  - Little research studies available at this time
  - If possible, communicate with neurologist → adjustment of voltage possible?
  - If strained-hoarse voice/excess loudness is noted, visual biofeedback may be helpful → app on ipad to show SPL meter with designed range, identify target intensity range
  - Shift of focus to clear speech (articulation) and increased pause times between words (clear lexical boundaries)

Future Directions

- Is LSVT® the best behavioral approach for speakers with PD (with/without DBS)?
  - How about effortful/clear speech?
  - Comparative studies needed!
  - How can we better predict treatment outcomes before treatment begin?
    - Less trial and error in therapy

Presentation Summary



- Behavioral modifications can improve speech function, particularly loud speech and clear speech
- DBS can worsen speech function
- LSVT may not be effective for some patients with STN-DBS intervention
  - Behavioral modifications may include the use of visual biofeedback to reduce excessive loudness, a focus on clear speech, and increased pausing
- Much more research is needed
  - Small n studies difficult to interpret
  - Heterogeneity of patients with PD and mixed success
  - Studies comparing treatment approaches – which one is the best?

Time for Questions

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