Auditory Considerations for Individuals with Multiple Sclerosis

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“Multiple sclerosis (MS) is an unpredictable, often disabling disease of the central nervous system that disrupts the flow of information within the brain, and between the brain and body.”

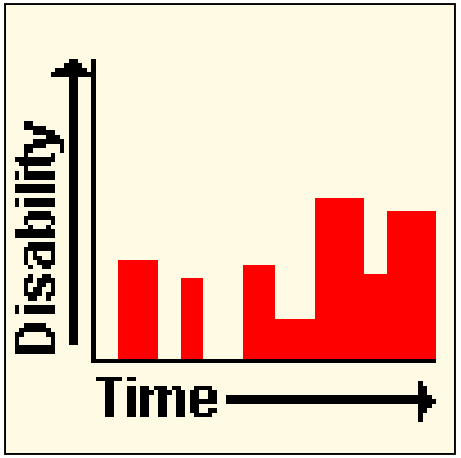
-National Multiple Sclerosis Society

https://www.nationalmssociety.org/What-is-MS

Four Courses of Multiple Sclerosis

1) Relapsing-Remitting (RR) MS:
   - Worsening of symptoms (relapse) that occurs with increasing frequency, along with periods of reduced symptoms (remission)
   - 85% initially diagnosed with this type of MS

Graphic on this slide (and the next two) is used with Dr. Eric Chudler's permission. The original can be found here: http://faculty.washington.edu/chudler/ms.html



The National Multiple Sclerosis Society website provides excellent information regarding MS. For details on MS types: https://www.nationalmssociety.org/What-is-MS/Types-of-MS

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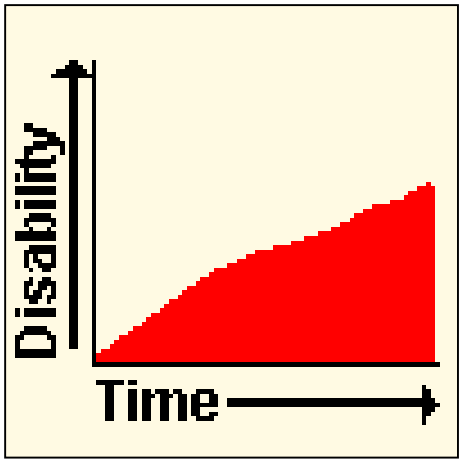
Four Courses of Multiple Sclerosis

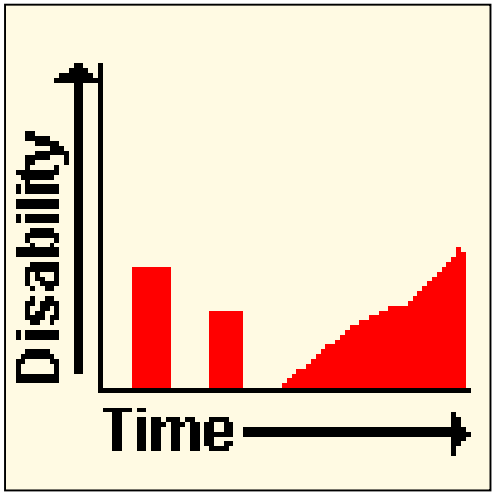
2) Primary-Progressive MS:
   - Steady progression of symptoms with few periods of remission
   - 15% are diagnosed with this type of MS



https://www.nationalmssociety.org/What-is-MS/Types-of-MS

Four Courses of Multiple Sclerosis

3) Secondary-Progressive MS:
   - Begins as RRMS but disease worsens to a point where there is no remission of symptoms



https://www.nationalmssociety.org/What-is-MS/Types-of-MS

Four Courses of Multiple Sclerosis

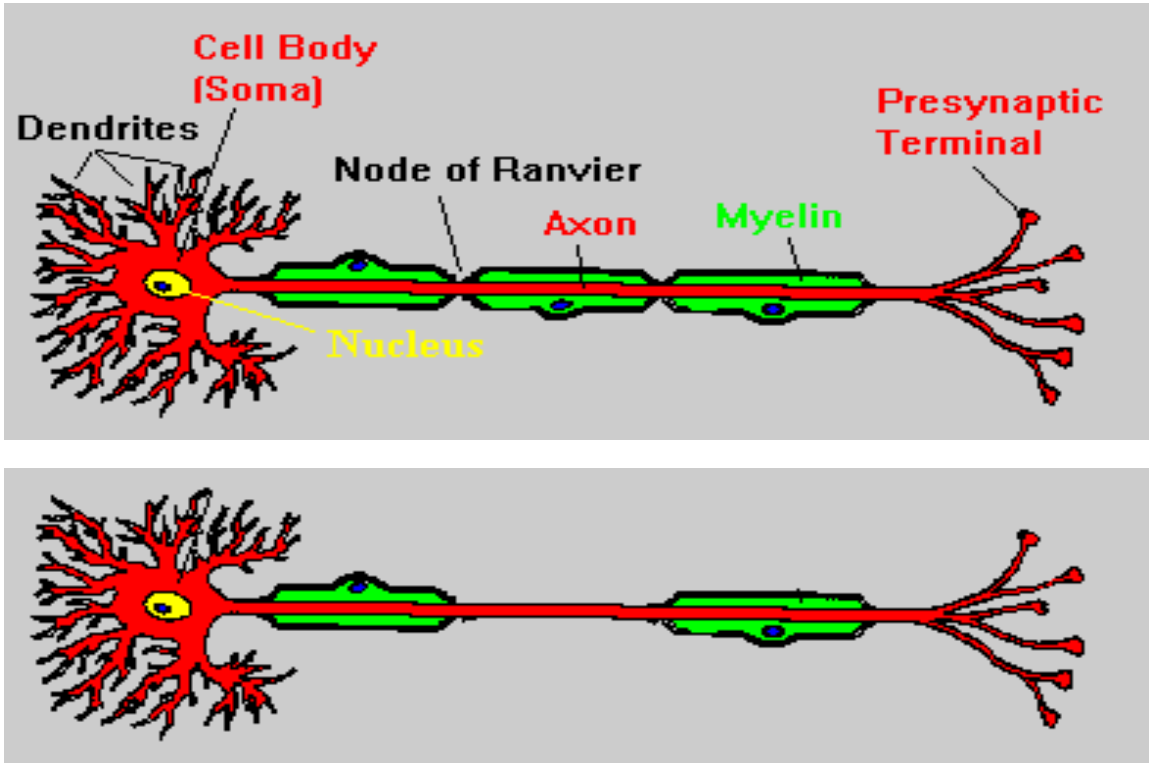
4) Clinically-Isolated Syndrome (CIS):

- Note change in course types occurred in 2013 – one course was eliminated and another was added[1]
- This the first presentation of symptoms that shows neurological signs consistent with MS
- Those who experience CIS might not progress to MS

[1]Dublin et al (2014). Defining the clinical course of multiple sclerosis: The 2013 revisions. *Neurology,* 83(3): 278-286.

https://www.nationalmssociety.org/What-is-MS/Types-of-MS

How Does MS Affect Neurons?



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Epidemiology of Multiple Sclerosis

- Numbers: 2.3 million world wide
- Age of onset: 20-50 years (usually)
- Geography: higher prevalence further from the equator
- More women than men
- Genetic component to MS
- Ethnicity: more common in Caucasians with northern European ancestors

https://www.nationalmssociety.org/What-is-MS/Who-Gets-MS#section-0

Treatment

- This is no cure for MS
- Medications:
  - to modify the disease course
  - to treat exacerbations (e.g., high-dose corticosteroids)
  - to manage symptoms
- Other treatment options may be considered:
  - rehabilitation (for functional improvements)
  - complementary/alternative medicine (symptoms)
    - need to review the evidence

http://www.nationalmssociety.org/Treating-MS

Common Symptoms of MS

- Fatigue
- Walking/Gait Problems
- Numbness/Tingling
- Spasticity
- Weakness
- Vision Problems
- Dizziness/Vertigo
- Sexual Problems
- Bowel/Bladder Problems
- Pain
- Cognitive Changes
- Emotional Changes/Depression

http://www.nationalmssociety.org/Symptoms-Diagnosis/MS-Symptoms

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Symptoms of MS (not so common)

- Speech Problems
- Swallowing Problems
- Tremor
- Seizures
- Breathing Problems
- Itching
- Headache
- Hearing Loss

http://www.nationalmssociety.org/Symptoms-Diagnosis/MS-Symptoms

Pure-Tone Hearing Loss

- There have been several case studies of individuals with MS presenting with sudden SNHL, ranging from mild to severe
- In most cases, the hearing thresholds eventually returned to normal or previous levels
- How about auditory function, as measured by the ABR, in these cases?
  - did not return to normal in some cases
  - did return to normal in some cases
  - some improvement was noted in others

Please feel free to email the presenter for the list of references re: the material on this slide as it is extensive.

Hypothesis: The pure-tone hearing loss was caused by a lesion and/or associated swelling at the lower portion of the central auditory pathway or on the cochlear nerve

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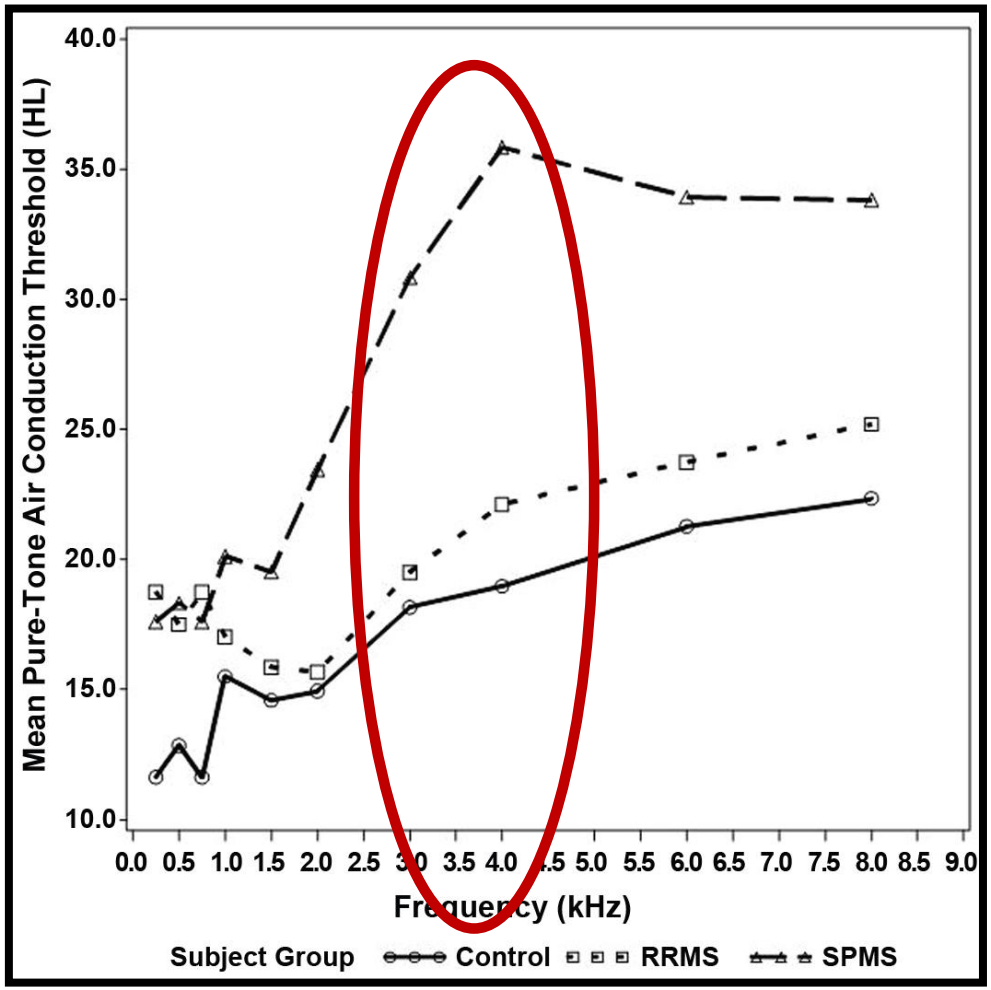
Pure-Tone Hearing Loss



Subjects: 26 with RRMS, 21 with SPMS, and 49 without MS

Results: MS had significantly poorer hearing in the lows (.25-.75 kHz) and highs (3-8 kHz) than controls

Lewis et al (2010). Audiometric hearing status of individuals with and without multiple sclerosis. *JRRD,* 47(7): 669-678.

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Pure-Tone Hearing Loss

- Larger study: 73 subjects with MS and 73 without MS (age and gender matched)
- **NOTE:** most (n=57) with MS had RRMS
- No significant difference (p=0.19) between those with MS and those without on pure-tone thresholds
- No significant meaningful correlations between pure-tone hearing thresholds and lesion activity
- Few lesions present in areas specific to audition

Doty et al (2012). Pure-tone auditory thresholds are not chronically elevated in multiple sclerosis. *Behav Neurosci,* 126(2): 314-324

Disease-Modifying Medications

- Five of the 15 currently FDA approved disease-modifying medications are a form of interferon
- Reports of interferon use and sudden hearing loss in patients with hepatitis
- Question: Do those taking interferon have poorer thresholds than those who do not?
- Answer: An ototoxic effect could not be ruled out at 8000 Hz

Lewis et al (2014). Does interferon beta-1-a impact pure-tone hearing Sensitivity among individuals with multiple sclerosis? *J Neursosci Nurs,* 46(6): 351-360.

Subjective Auditory Complaints

- When hearing loss is not a presenting symptom, it is often not evaluated, or asked about, by MS practitioners
- 23% of the subjects had measurable HL and 14% reported subjective hearing loss[1]
- 18% (n=6) had HL (but no information was provided regarding their subjective hearing problems) and 33% (n=11) with hearing thresholds WNL reported subjective hearing problems[2]

[1]Grénman (1985). Involvement of the audiovestibular system in multiple sclerosis: and otonuerologic and audiologic study, *Acta Otolaryngol Suppl*, 420: 1-95.
[2]Musiek et al (1989). Electrophysiologic and behavioral auditory findings in multiple sclerosis, *Am J Otol*, 10:43-350.

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Hypothesis: Lack of correlation with HL and auditory complaints due to CANS dysfunction

Musiek et al (1989). Electrophysiologic and behavioral auditory findings in multiple sclerosis, *Am J Otol*, 10:43-350.

(Central) Auditory Processing

Test	Abnormal Findings (%)
Masking Level Difference	50%
Low Pass Filtered Speech	7%
Staggered Spondaic Word Test	22%
Dichotic Digits	33%
Frequency Patterns	20%

Musiek et al (1989). Electrophysiologic and behavioral auditory findings in multiple sclerosis, *Am J Otol*, 10:43-350.

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Dichotic Listening

Test	Abnormal Findings (%)
Staggered Spondaic Word test	10%
Dichotic CV test	80% (REA)
Synthetic Sentence Identification – Ipsilateral Competing Message	25%

Other studies have reported not only a LE deficit but also a RE enhancement (e.g., Rubens et al., 1985; Lindeboom & Ter Horst, 1988; Wisehart et al., 1995).

Jacobson et al. (1983). Dichotic paradigms in multiple sclerosis. *Ear Hear,* 46(6): 311-317.

Dichotic Listening: Imaging Studies

- Poorer dichotic listening has been associated with callosal atrophy[1, 2, 3]

[1]Pelletier et al (1993). Functional and magnetic resonance imaging correlates of callosal involvement in multiple sclerosis. *Arch Neurol,* 50(10): 1077-1082.

[2]Gadea et al (2002). Dichotic listening and corpus callosum magnetic resonance imaging in relapsing-remitting multiple sclerosis with emphasis on sex differences. *Neuropsychology,* 16(2): 275-281.

[3]Berlow et al (2012). Magnetic resonance imaging correlates of dichotic listening performance in multiple sclerosis. *Sem Hear,* 33(3): 283-294.

Temporal Processing

Test	Abnormal Findings (%)
MLD	38%
Gap Detection	13%

- Subjects with poorer behavioral performance had more abnormalities on their auditory-evoked potentials
  - MLD = ABR/MLR
  - Gap-detection = LLR

Hannley et al (1983) also found a relationship between MLD and Wave III of the ABR

Hendler, Squires, Emerich (1990). Psychophysical measures of central auditory dysfunction in multiple sclerosis: neurophysiological and neuroanatomical correlates. *Ear Hear,* 11(6): 403-416.

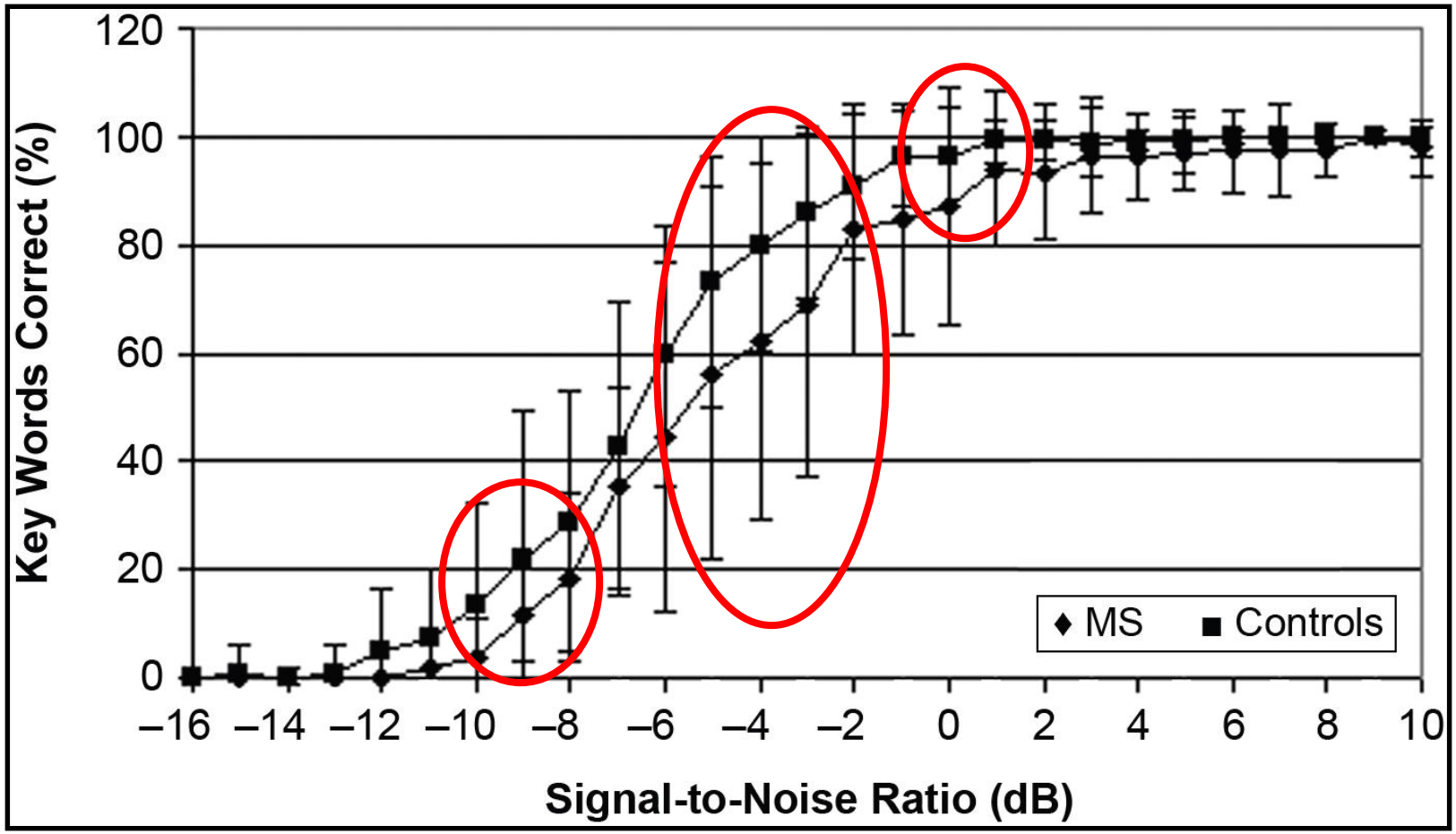
Temporal Processing

- 16 subject with MS:
  - 7 with isolated brainstem auditory pathway lesions
  - 4 with only forebrain auditory lesions
  - 1 caudal and rostral pathway lesions
  - 4 with no lesions in auditory pathway

Test	Result
Gap Detection	40% outside normal limits
SIN - Continuous	No difference btwn groups
SIN - Interrupted	Difference btwn groups

Rappaport et al (1994). Auditory temporal resolution in multiple sclerosis. *J Otolaryngol,* 23(5): 307-324.

Speech Recognition in Noise



Lewis et al (2006). Some effects of multiple sclerosis on speech perception in noise: preliminary findings. *JRRD,* 43(1); 91-98.

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Contralateral Acoustic Reflex

- Musiek et al (1989): 26% of subjects (who had hearing WNL) had abnormal results

- Jacobson et al (1983):
  - 5% (n=1) with abnormal reflexes
  - 35% with positive decay

- Jerger et al (1986): 71% identification rate with the acoustic reflex test

Jerger et al (1986). Patterns of auditory abnormality in multiple sclerosis. *Audiology,* 25(4-5): 193-209.

Auditory Brainstem Response (ABR)

- Musiek et al (1989): 62% of his subjects had abnormal results on at least one ABR measure, 42% had bilateral abnormalities
- In fact, several studies have noted abnormalities in ABR waveforms in patients with MS
- Abnormalities in Wave V seem to be common in the literature but other abnormalities have been noted
- An increase in abnormality may be seen with increases in stimulation rate (Rappaport et al, 1994)

Due to the large number of citations on this slide (bullet points #2 and #3), please contact the author for the references.

Auditory Brainstem Response (ABR)

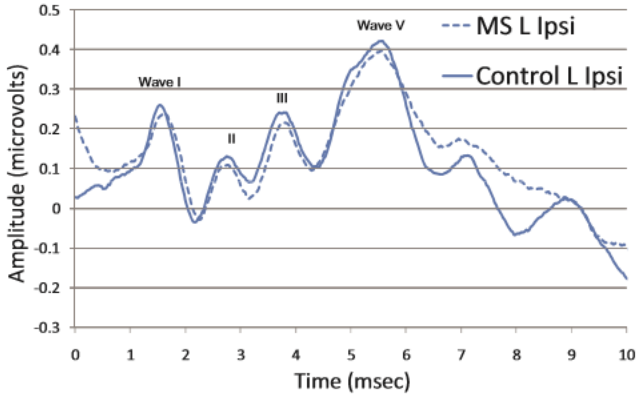

Figure 1 Grand average auditory brain stem responses in response to clicks presented to subjects' left ear. Ipsi, ipsilateral.

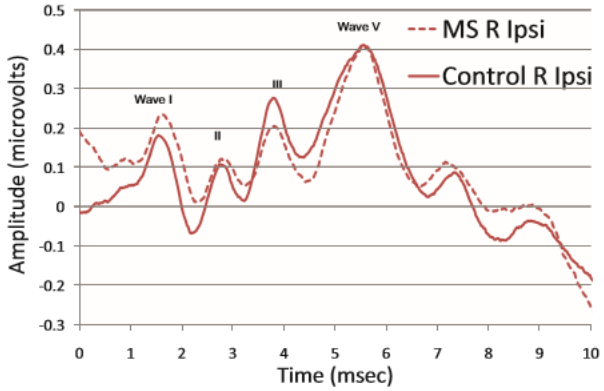

Figure 2 Grand average auditory brain stem responses in response to clicks presented to subjects' right ear. MS, multiple sclerosis; R, right; Ipsi, ipsilateral.

Folmer et al (2012) Electrophysiological measures of auditory processing in patients with multiple sclerosis. *Semin Hear,* 33(3): 274-282

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Auditory Brainstem Response (ABR)

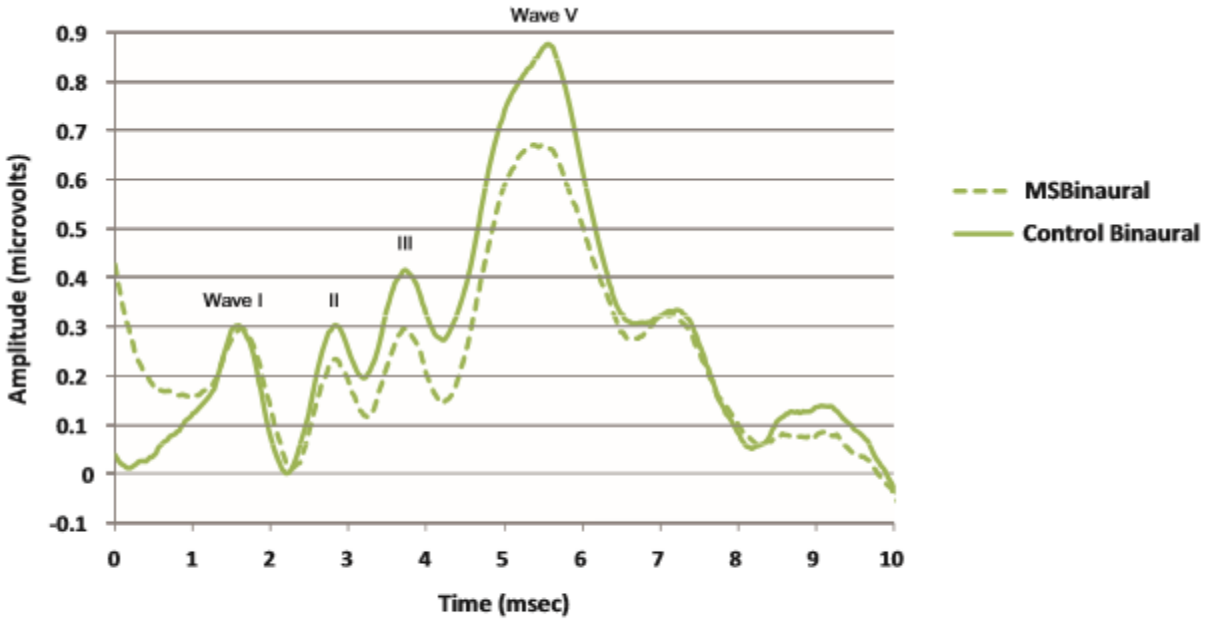

Figure 3 Grand average auditory brain stem responses in response to clicks presented to both ears simultaneously. MS, multiple sclerosis.

Folmer et al (2012) Electrophysiological measures of auditory processing in patients with multiple sclerosis. *Semin Hear,* 33(3): 274-282

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Middle Latency Response (MLR)

- Hendler et al (1990) reported no MLR abnormalities if the ABR was normal. Of those with unilateral ABR abnormalities, 33% had abnormal MLR findings.
- Rappaport et al (1994) reported that 31% of their subjects had abnormal MLR findings. Three had no waveforms and the other two had delayed latencies of Pa peaks. 80% of these had brainstem lesions.

Rappaport et al (1994). Auditory temporal resolution in multiple Sclerosis. *J Otolaryngol,* 23(5): 307-324.

Middle Latency Response (MLR)

- Celebisoy et al. (1996) evaluated ABR and MLR in 30 subjects with MS
  - 60% had ABR abnormalities
  - 73% had MLR abnormalities
  - 50% had both ABR and MLR abnormalities
  - MLR abnormalities were found in 7out of 12 subjects with normal ABR

Late Latency Response (LLR)

- Polich et al (1992) reported that auditory P300 latency was greater and P300 amplitude was smaller from MS patients compared to control subjects.
- Others have also noted an increase in latency.

Aminoff J, Goodin D (2001). Long-latency cerebral event-related potentials in multiple sclerosis.*J Clin Neurophysiol,* 18(4): 372-377.

Piras et al (2003). Longitudinal study of cognitive dysfunction in multiple sclerosis: neuropsychological, neuroradiological and neurophysiological findings. *J Neurol Neurosurg Psychiatry,* 74(7): 878-885.

Mangano et al (2006). Cognitive impairment and neurophysiological correlates in MS. *J Neurol Sci,* 245(1-2): 117-122.

Gonzalez-Rosa et al (2006). Differential cognitive impairment for diverse forms of multiple sclerosis. *BMC Neurosci,* 19:7-39.

Whelan et al (2010). A high-density ERP study reveals latency, amplitude, and topographical differences in multiple sclerosis patients versus controls. *Clin Neurophysiol,* 121(9): 1420-1426.

Polich et al (1992). P300 in multiple sclerosis: a preliminary report. *Int J Psychophysiol,* 12(2): 155-163.

Late Latency Response (LLR)

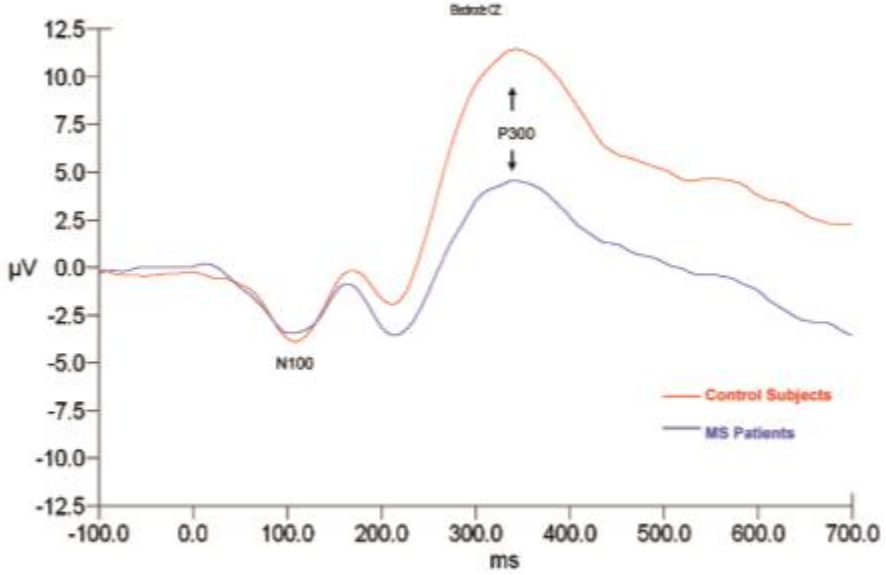

Figure 4 Grand average auditory event-related potentials (recorded at electrode Cz) in response to 1000-Hz tones (target) presented to subjects' right ear during the "oddball" protocol.

Folmer et al (2012) Electrophysiological measures of auditory processing in patients with multiple sclerosis. *Semin Hear,* 33(3): 274-282

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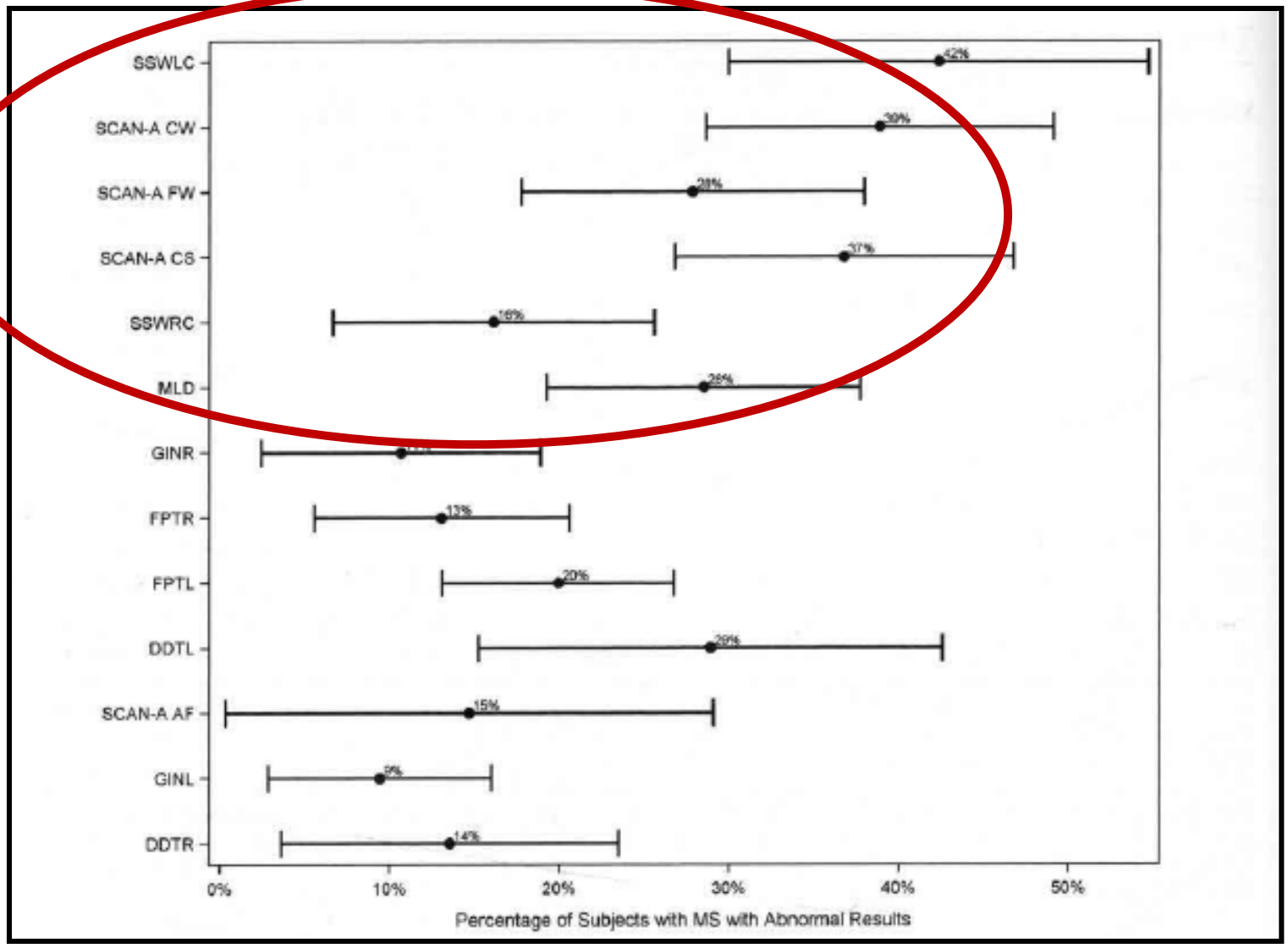

Lewis et al (2012). Preliminary identification of central auditory processing screening tests for individuals with multiple sclerosis. *Sem Hear, 33(3): 261-273.*

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Wrap-Up

- In cases of sudden SNHL:
  - See patient right away
  - Must have them seen for medical evaluation right away as well

Audiologic Evaluation:

- Comprehensive audiometric evaluation
- Immittance audiometry (including reflexes)
- Otoacoustic emissions (to help determine site)
- Consider ABR

Musiek et al. (2012). *Disorders of the Auditory System.* San Diego: Plural Publishing.

Wrap-Up

- Serial audiograms:
  - Check for stability (prior to considering amplification)
  - Changes in disease activity/progression
  - Monitoring if on certain medications (Lewis et al, 2014)
- Self-assessment of auditory function
- Assessment of central auditory function:
  - Behavioral tests
  - Auditory-auditory potentials

Musiek et al. (2012). *Disorders of the Auditory System.* San Diego: Plural Publishing.

Due to time constraints, this presentation did not cover dizziness/vertigo related to MS or auditory rehabilitation specific to this population. Since the author has expertise in the area of auditory rehabilitation on this topic, attendees are welcome to contact her for more information at the email address below. Additionally, as mentioned previously, a full citation list may be requested.

Questions?

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