Binaural Hearing and Speech Laboratory

INVESTIGATING AUDITORY MATURATION IN DOWN SYNDROME

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INTRODUCTION

Down Syndrome: A developmental disorder with high prevalence of hearing loss

- **One in 707 live births in the United States.[1]**
- **Intellectual disability and Language difficulties are characteristic phenotypes.[2]**
- **Up to 75% of people with Down's syndrome experience hearing impairment at some point in their lives.[3,4]**
- **Among typically developing (TD) children, chronic hearing loss affects the maturation of auditory perceptual functions such as spatial hearing and speech intelligibility in noise.[5,6]**
- **Auditory perceptual abilities depend on the complex interrelationship between auditory, cognitive, and linguistic factors — factors that are impacted in individuals with DS and may have a compounding adverse impact on auditory perception (Figure 1).**

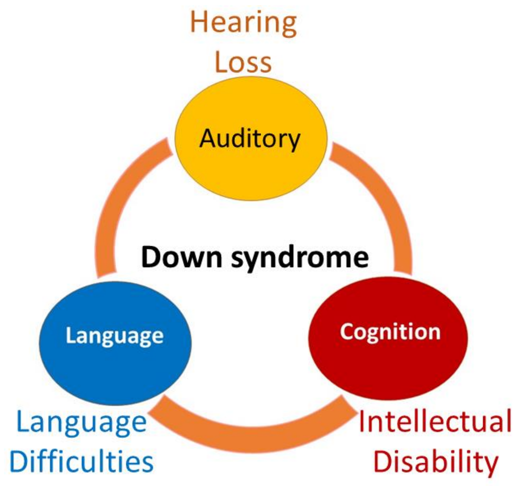

Figure 1: Complex interactions of auditory, cognitive and linguistic functions in DS

Findings from a previous study in the lab involving young adults with DS[7]

1. Differential maturation of 'where' and 'what' auditory functions in young adults with DS
2. Hearing loss and verbal working memory predicted speech intelligibility

- # DS young adult (age18-24), N=19
- Seven participants with DS had mild or severe hearing loss in one or both ears.
- * TD data from previous studies using similar task set up.[8,9,10]

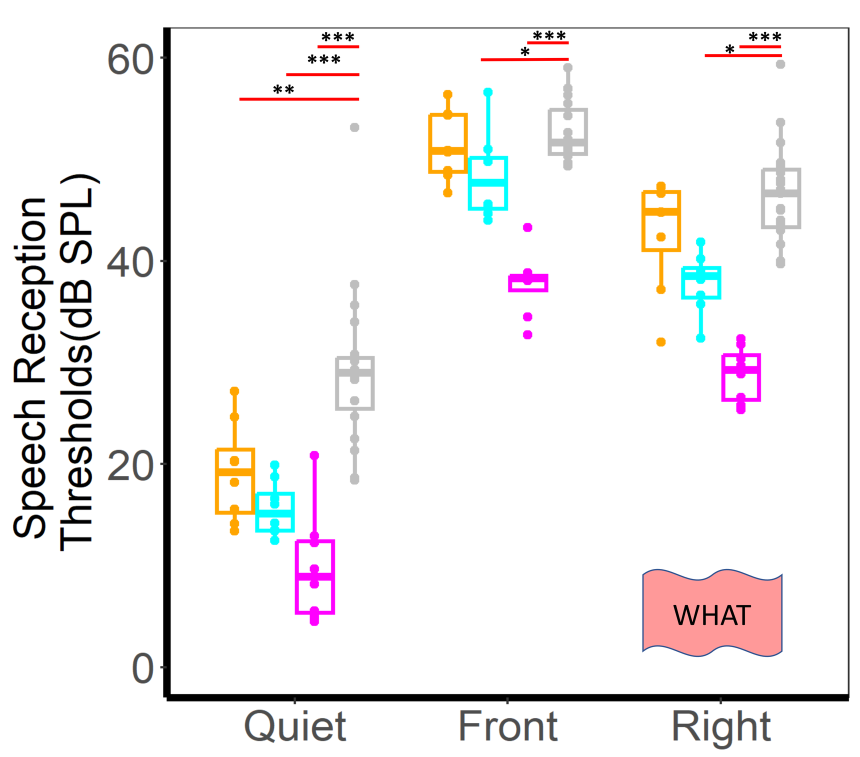

Figure 2: Speech reception thresholds in *quiet, front,* and *right* conditions. See Methods for task details. *p<0.05; **p<0.01; ***p<0.001

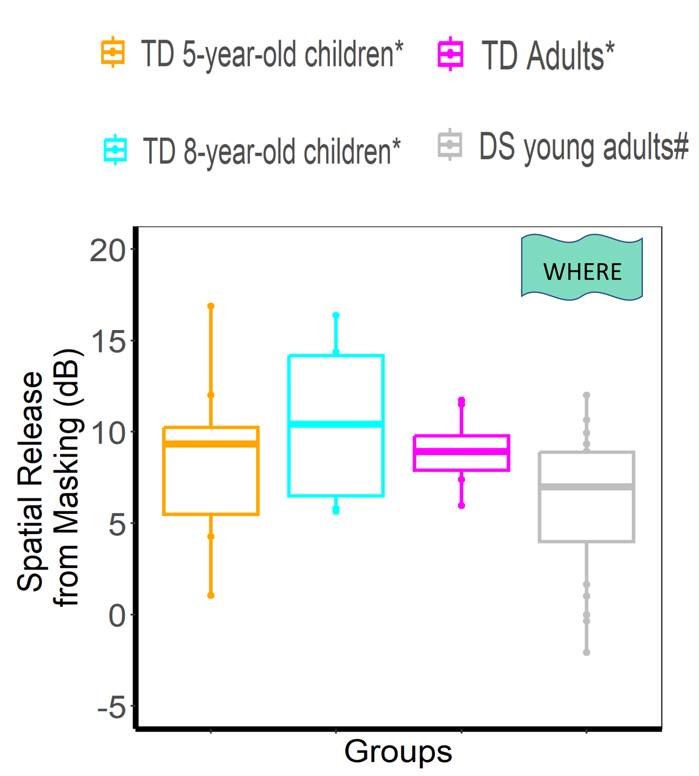

Figure 3: Spatial release from masking. See Methods for task details.

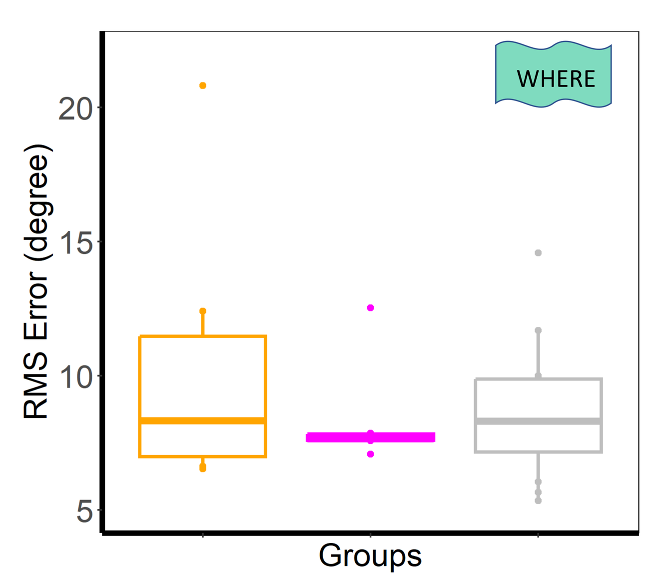

Figure 4: Root Mean Square (RMS) errors in a sound location identification task with 15 loudspeakers.

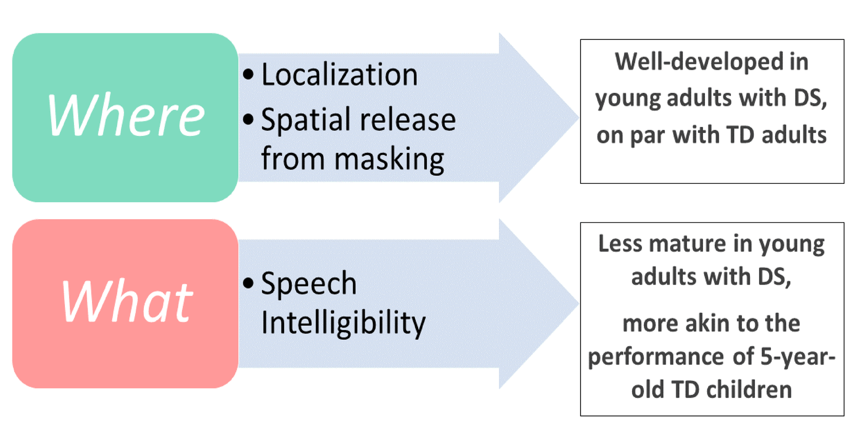

Figure 5: Model summarizing findings of differential maturation of 'where' and 'what' auditory functions in DS

STUDY HYPOTHESES

The 'what' domain of auditory perception in DS is impacted by immaturity in the auditory system, hearing loss, and cognitive deficits.

1. Impairments in 'what' perception will be associated with greater hearing loss and lower cognitive abilities in DS.
2. Objective assessment of auditory maturation, using cortical auditory evoked potentials after compensating for hearing loss, will reveal maturational delays in DS that would account for their functional abilities on tasks evaluating 'what' auditory perception.

STUDY AIMS

1. To characterize the developmental trajectory of speech perception in quiet and noisy conditions, and speech-related auditory processing in TD children, adolescents, and adults.
2. To characterize the maturational status of these measures in young adults with DS by comparing with the typical auditory developmental trajectory.
3. To examine relationships between auditory maturational delays, hearing loss, and cognitive abilities in DS.

PRELIMINARY OBSERVATIONS TO DATE

METHODS

Study Details

- Study Sample
  - TD Children (age range 10-12), N=3
  - TD Adolescents (age range 14-16), N=4
  - TD young adult (age 22), N=1
  - DS young adult (age 24), N=1
- Pure-tone air conduction audiometry at octave frequencies between 250-8000 Hz
  - All TD participants passed a hearing screening at 20 dB HL
  - Thresholds for young adult with DS are shown in Figure 6

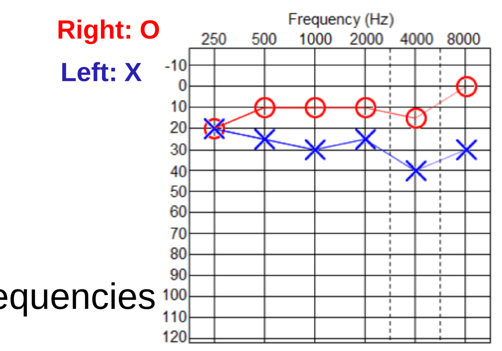

Figure 6: Audiogram of the participant with DS

Speech Intelligibility Test

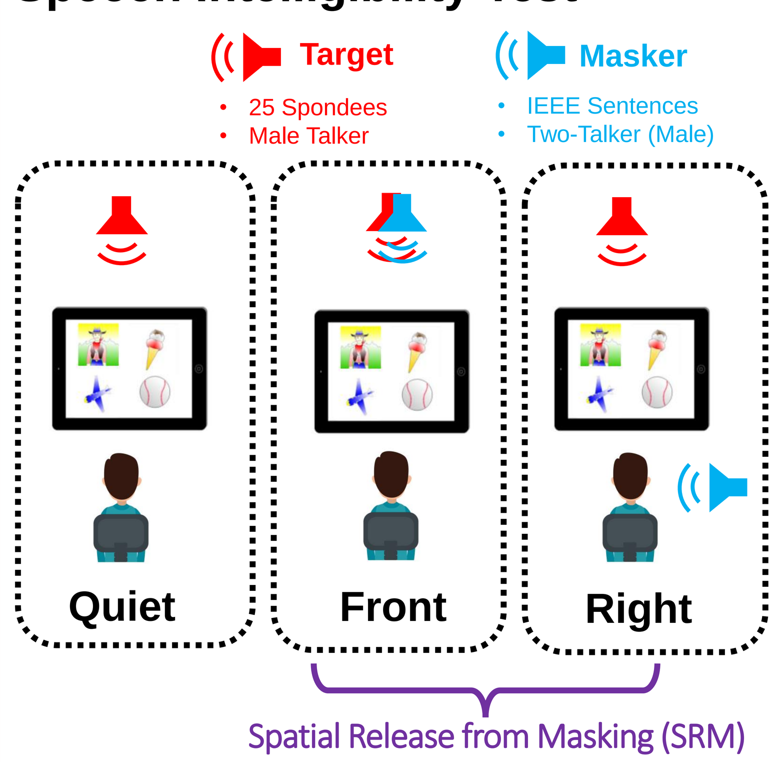

Figure 7: Children's Realistic Index for Speech Perception (CRISP) Task

- A closed-set four alternative force choice task[11] (Figure 7)
- Each target spondee had a corresponding picture used by participants to indicate response
- Maskers were at 55 dB SPL
- Target was initially presented at 60 dB SPL and then varied following an adaptive tracking procedure (3-down/1-up rule)
- **Speech reception threshold (SRT):** point on the psychometric function where performance was estimated at **79.4%**
- **Spatial release from masking (SRM): $SRT_{front} - SRT_{right}$**

EEG Recording

- Cortical auditory evoked potentials (CAEPs) were collected in response to two 80 ms naturally spoken /m/ & /s/ phonemes, presented in a random order (150 trials each) during a passive listening task.
- /m/ is a low frequency sound whereas /s/ is a high frequency sound. See Figure 8.
- Stimuli were presented through ER2 insert earphones in each ear separately (Figure 9) with a variable inter-stimulus interval of 1000-1500 ms, using E-Prime 3.0 software (Psychology Software Tools, PA, USA).
- In TD individuals, stimuli were presented at a level of 65 dB SPL; for the participant with DS, the sound presentation level was compensated for hearing loss in the left ear.

Stimuli

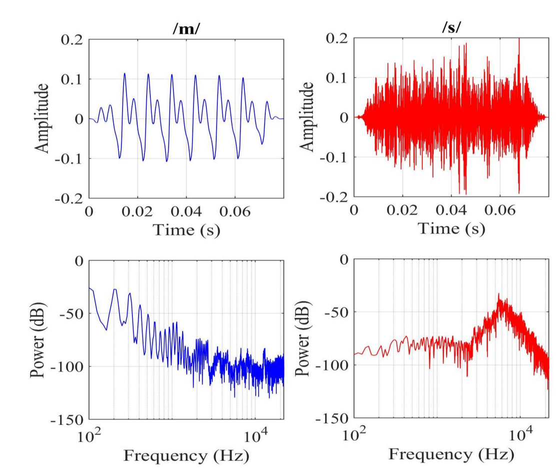

Figure 8: Waveforms and power spectra of /m/ and /s/ stimuli

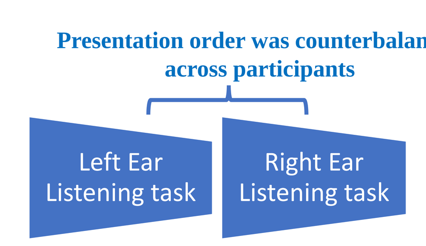

Figure 9: EEG acquisition conditions

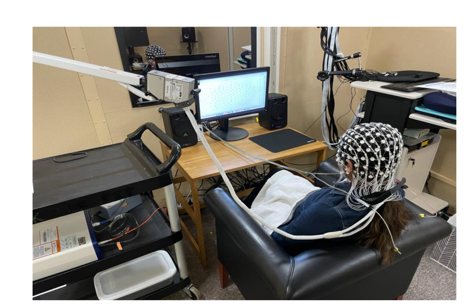

Figure 10: 128- channel geodesics EEG acquisition system

EEG was recorded using a 128-channel Hydrocel Geodesic Sensor Net with NetAmp 400 amplifiers (Electrical Geodesics Inc., Magstim EGI, Eugene, OR, USA). See Figure 10.

Verbal Working Memory Assessment

Wechsler Intelligence Scale for Children Fourth Edition (WISC-IV; Wechsler, 2003) – Digit Span Forward and Backward were used to assess simple and complex verbal working memory, respectively.[12]

RESULTS

Behavior

Maturational delays in speech intelligibility, spatial release from masking, and verbal working memory capacity were observed in the young adult participant with DS.

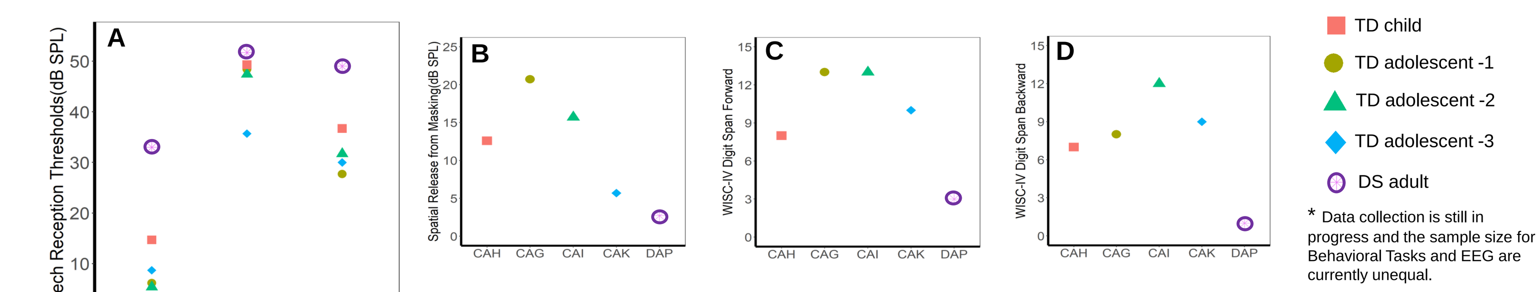

Figure 11: Speech reception thresholds in *quiet, front,* and *right* conditions (A). Spatial release from masking (B). WISC-IV Digit Span Forward total score (C). WISC-IV Digit Span Backward total score (D).

EEG

- **Clear N1-P2 response was observed in all participants.**
- **CAEP responses in the young adult participant with DS potentially indicate auditory maturational delays.**

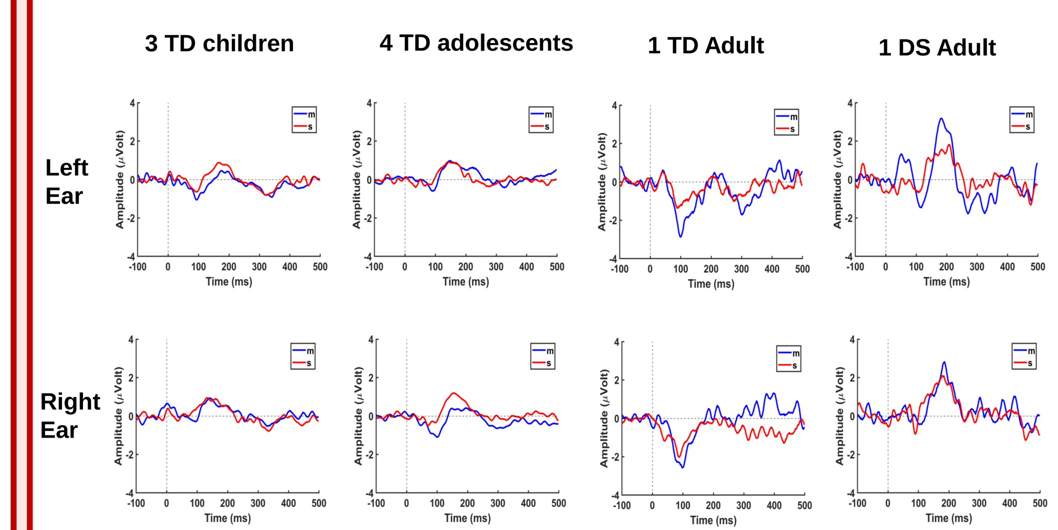

Figure 12: CAEP waveforms at Cz for /m/ and /s/ stimuli. The left ear condition is shown in the upper panel and right ear condition is shown in the lower panel.

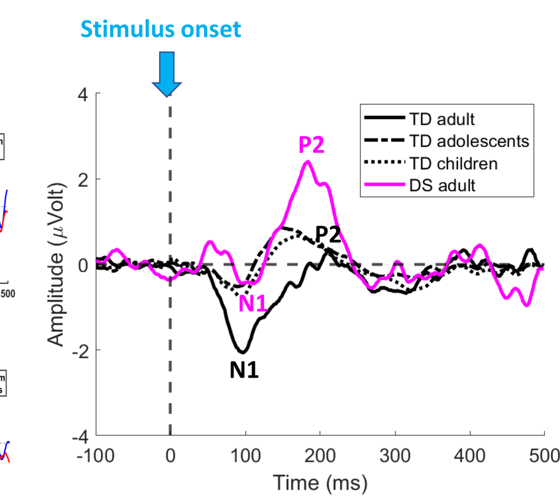

Figure 13: CAEP waveforms at Cz averaged across phonemes (/m/, /s/) and ears.

SUMMARY

- Age-related maturational changes in speech intelligibility, verbal working memory scores, and CAEP responses were observed among TD children and adolescents.

1. In comparison with 1 TD child (10-year-old) and 3 adolescents (14-16 years old), the young adult participant with DS demonstrated
   a) Higher SRTs in the quiet condition and in the presence of speech maskers.
   b) SRM was smaller, i.e., less benefit from spatial cues in segregating target speech from the spatially separated masker.
   c) Lower verbal memory scores in both simple and complex test conditions.
2. CAEP responses showed effects of stimulus type, i.e., different responses for low frequency /m/ and high frequency /s/ phonemes. Future research will investigate the association between hearing thresholds and CAEP characteristics across stimulus types.
3. In support of our hypothesis, a less mature N1 peak with longer latency and a large P2 peak present in an adult participant with DS may indicate immaturity in the auditory system. In individuals with DS, previous studies have reported longer N1 latencies and higher P2 amplitudes during passive listening paradigms.[13]

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