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.¹⁴
- 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).

**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 those 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.

**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.

**METHODS**

**Brain Imaging**

- T1-weighted structural images were obtained using a 3.0-Tesla Tesla Siemens Tim Trio MRI scanner.
- Diffusion tensor imaging (DTI) was acquired using a 32-channel head coil.
- Cortical thickness maps were calculated using FreeSurfer software.

**Behavior**

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

**RESULTS**

- Higher SRTs in the quiet condition and in the presence of speech maskers.
- 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).
- Each target spondee had a corresponding picture used by participants to indicate response.

**SUMMARY**

- Age-related maturational changes in speech intelligibility, verbal working memory scores, and CAEP responses were observed among TD children and adolescents.
- In comparison with 1 TD child (10 year-old) and 3 adolescents (14-16 years old), the young adult participant with DS demonstrated

  1. Higher SRTs in the quiet condition and in the presence of speech maskers.
  2. Lower verbal memory scores in both simple and complex test conditions.
  3. 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.
- 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