Canadian Academy of Audiology
Academie Canadienne d'audiologie

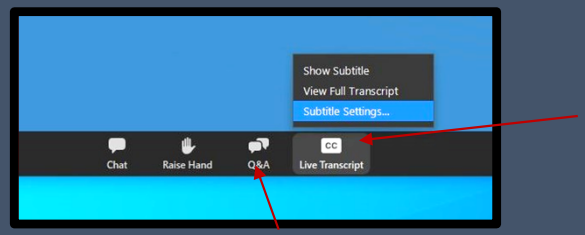
Audiological considerations and speech perception outcomes for children and adults living with Down syndrome

Speaker: Lori Leibold, Ph.D.
Director, Center for Hearing Research
Boys Town National Research Hospital

Moderator: Stephen Lomber, Ph.D.
Professor of Physiology, McGill University

September 16, 2024

CAA Webinars Include Live Zoom Transcription



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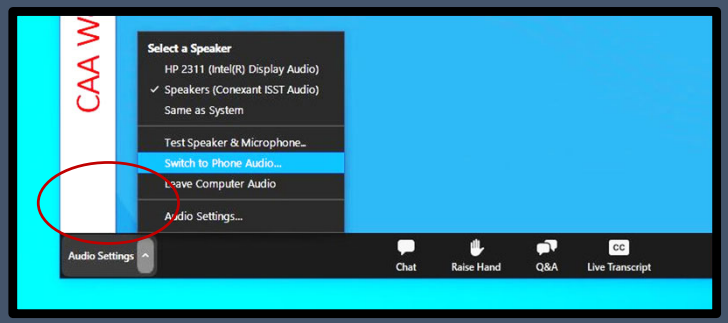

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Canadian Academy of Audiology is a professional association dedicated to enhancing the role of audiologists as primary hearing health care providers through advocacy, education and research.

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Moderator: Stephen Lomber, Ph.D.

Stephen G. Lomber, Ph.D. is a Professor of Physiology at McGill University and directs the Cerebral Systems Laboratory. Work in his lab examines cortical plasticity in the presence and absence of acoustic input, and following the initiation of auditory processing through the means of cochlear prosthetics.

Dr. Lomber is the Scientific Program Chair for the Annual Meeting of the Association for Research in Otolaryngology (ARO). He is a past chair of the Gordon Research Conference on the Auditory System and the International Conference on Auditory Cortex, and a CAA board member.

Speaker: Lori Leibold, Ph.D.
Director, Center for Hearing Research
Boys Town National Research Hospital

Lori Leibold is the Director of the Center for Hearing Research and leads the Human Auditory Development Laboratory at Boys Town National Research Hospital in Omaha, Nebraska. Her background is in audiology and developmental psychoacoustics. Her research is focused on understanding how and when hearing and speech perception develop across infancy and childhood, which includes studies evaluating the speech perception and auditory abilities of infants, children, and adults living with Down syndrome. Working with a team of scientists, clinicians, and community engagement specialists, she is involved in multiple efforts to increase participation rates, promote programmatic longevity, and improve hearing health outcomes for individuals living with Down syndrome.

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Audiological Considerations and Speech Perception Outcomes for Children and Adults Living with Down Syndrome

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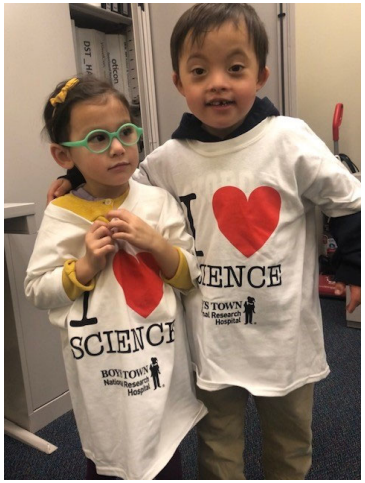

Project INCLUDE

BOYS TOWN National Research Hospital®

Support and Funding

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Our Team: Research

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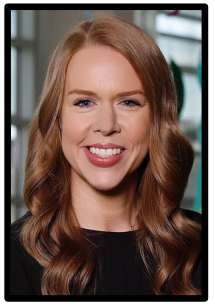

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Our Team: Community Advisory Board

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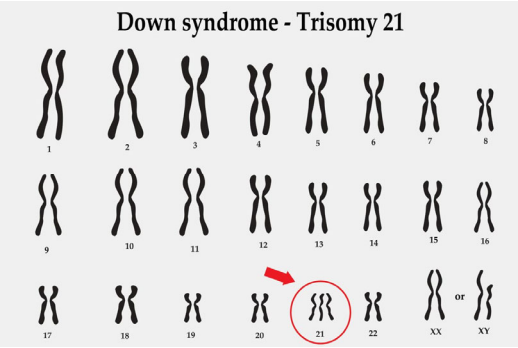
COMMUNITY ADVISORY BOARD GOALS

- represent the interests of individuals with Down syndrome and their families in current and future Project INCLUDE activities
- make sure that Project INCLUDE research activities are inclusive of many different abilities
- share Project INCLUDE research results in ways that are meaningful to individuals with Down syndrome and their families

Down Syndrome: Quick Facts

- ~1 in 700 live births
- Known genetic cause (trisomy 21)
- Predisposed to certain medical conditions
- Appear to be ‘protected’ against other medical conditions
- Extensive heterogeneity in phenotypic outcomes

Down Syndrome: Prior to the 1970s and 1980s



- Average lifespan <30 years
- Most individuals were institutionalized
- Lack of access to education and medical care
- Funding for and inclusion in research was limited

Down Syndrome: Today

risingkites.org

- Average lifespan ~60 years
- Are not typically institutionalized
- IQ scores have increased by 20 points
- Most individuals with Down syndrome learn to read and write
- **Extensive heterogeneity** in phenotypic outcomes

The NIH INCLUDE Project

INvestigation of Co-occuring conditions across the Lifespan to Understand Down syndromE

The overarching goal of the INCLUDE Project is to improve the health and quality of life of people with Down syndrome.

NIH National Institutes of Health
Turning Discovery Into Health

DS-Connect®
The Down Syndrome Registry

INCLUDE
Data Coordinating Center

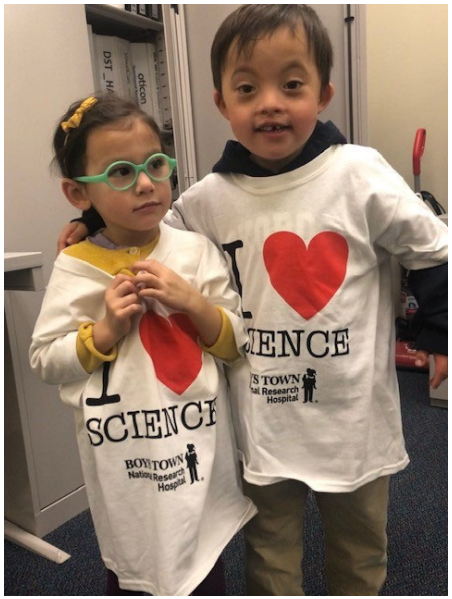
Medical Problems Common in Down Syndrome

Bull et al. 2022 (Table 1)

Condition	%
Hearing problems	75
Vision problems	60-80
Obstructive sleep apnea	50-79
Otitis media with effusion	50-70
Congenital heart disease	40-50
Feeding difficulty	31-80
Respiratory infection	20-36
Dermatologic problems	56
Hypodontia & delayed dental eruption	23
Congenital hypothyroidism	2-7

Condition	%
Antithyroid antibody positive	13-39
Thyroid disease by adulthood	50
Gastrointestinal atresias	12
Seizures	1-13
Hematologic problems	1-10
Autoimmune conditions	13-39
Symptomatic atlantoaxial instability	1-2
Autism	7-19
Hirshsprung disease	<1

Project INCLUDE: Boys Town



https://youtu.be/wUywFj1SOT4

- Characterize hearing and improve audiological assessment across the lifespan
- Characterize early development of speech perception and selective listening
- Identify factors that support functional hearing

Methods: Overview

OTOSCOPY

Tip: Recommend wax removal prior to visiting the lab (or clinic).

We will look in your ear.

You will play a listening game.

AUDIOMETRIC THRESHOLDS

- modified Hughson-Westlake procedure
- air conduction at 0.25-8 kHz and at 11.2 and 16 kHz
- bone conduction testing when air conduction thresholds >20 dB HL (0.25-4 kHz)

WIDEBAND ACOUSTIC IMMITTANCE

- broad frequency range (0.25-8 kHz)
- 21.5 clicks/second
- at ambient pressure and at static pressures from -300 to +200 daPa

The computer makes a funny sound.

You will pick the word you hear.

MASKED SPEECH RECOGNITION

- word recognition in background noise or speech
- picture-pointing response
- target words and masker presented from same speaker (colocated) or from different speakers (spatially separated)

LANGUAGE AND COGNITION

- receptive vocabulary (PPVT-V)
- non-verbal intelligence (Stanford-Binet Intelligence Scales)
- excecutive function (BRIEF)

You will point at pictures.

Down Syndrome: Hearing loss

- Prevalence: 50-80% across studies Ëfin”vn» nl#i%#M£fi#nfi#a#Suafik n#¨˘·˘½
- Hearing loss is often conductive, secondary to otitis media with effusion (Nightengale et al. 2017)
- High rates of mixed and sensorineural hearing loss throughout the lifespan (DeSchrijver et al. 1990)
- Evidence suggesting early onset presbycusis (Buchanen 1990)

Consequences of Hearing Loss: Children

- Inconsistent auditory access, often leading to reduced cumulative auditory experience and language delay (Tomblin et al. 2015)
- Speech-in-noise difficulties (Qju}ılnfin#a}#'ÆÆ¸)
- Reduced spatial hearing abilities (S£~ i}ı#å#Pa¡jn"˘·¸)
- Effects of chronic otitis media often persist even years after resolution Ë9a}#n#a}#'ÆÆ˝½

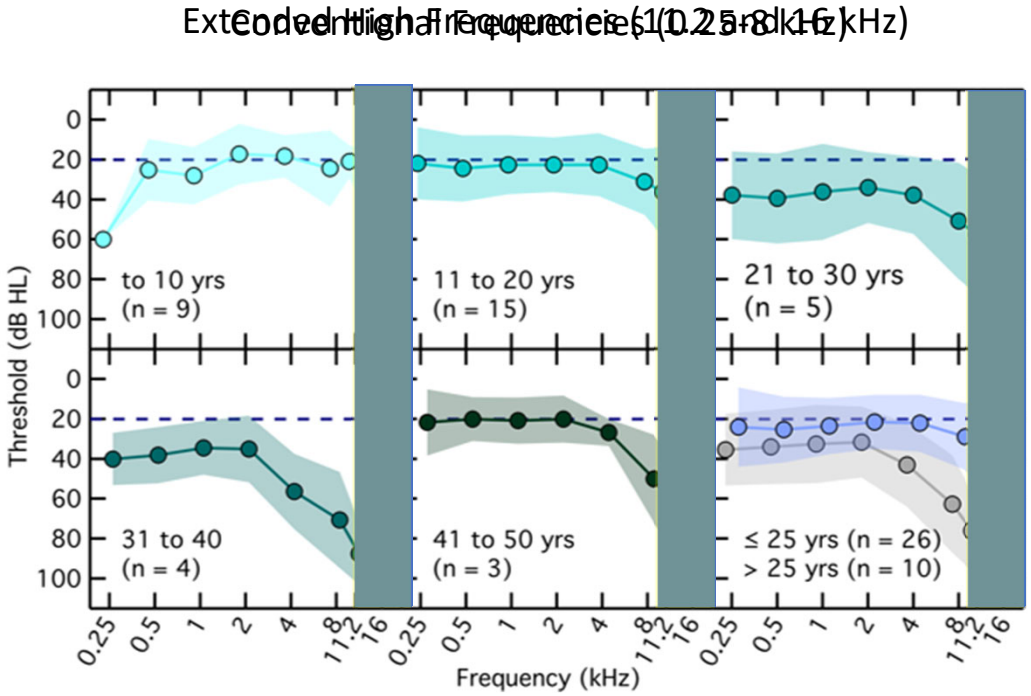
Down Syndrome: Other Potential Barriers to Real-World Communication Outcomes

- Impairments in executive function, particularly working memory and selective attention/inhibition (Lanfranchi et al. 2010)
- Language delays (Abbeduto et al. 2007; Martin et al. 2009)
- Speech production difficulties, reduced intelligibility (Kent & Vorperian 2012; Wilson et al. 2019)

Study Findings: Audiological Outcomes

- Participants: 36 individuals with Down syndrome
- Age range: 6-47 years
- Presented at the 2024 meeting of the American Auditory Society ïM£fïnfïnïa}#¨˘¨¸½

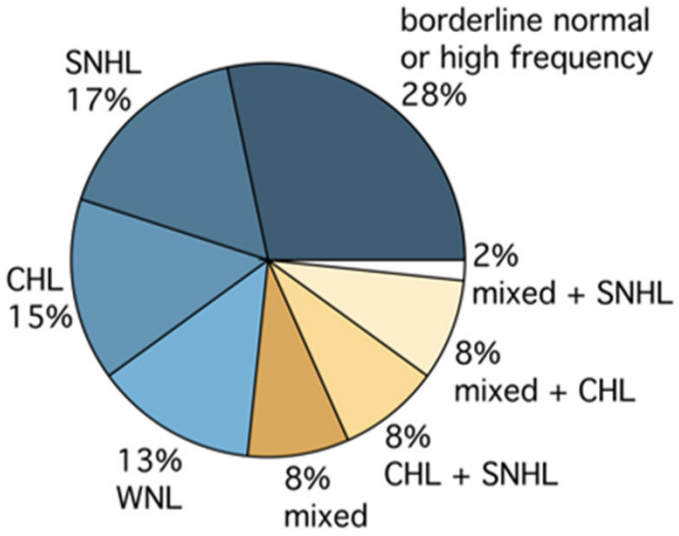
Results: Air Conduction Thresholds



87% of participants had hearing loss

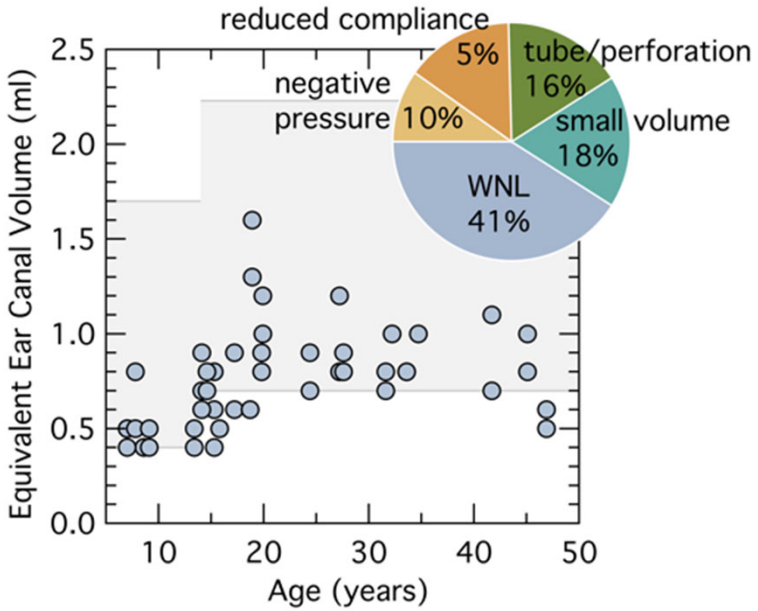
Porter et al. 2024 (American Auditory Society)

Results: Type of Hearing Loss



Porter et al. 2024 (American Auditory Society)

Results: Middle Ear Dysfunction



Porter et al. 2024 (American Auditory Society)

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- 87% of ears tested had some type of hearing loss
  - Conductive hearing loss was not the most common type observed
- Better-ear speech intelligibility index (SII) ranged from 15-99 (average = 84.2)
- Hearing aid use was low relative to age-matched peers who are neurotypical

More Comprehensive Data Coming Soon!

9th International Pediatric Conference
A Sound Foundation Through Early Amplification

October 27-30, 2014

San Diego, California (or live streaming)

October 28th	
14:10 PM - 14:45 PM	**Behavioral hearing assessment in children with developmental differences** Angela Bonino (USA)
14:45 PM - 15:15 PM	**Coffee Break**
15:15 PM - 15:50 PM	**Management of Children with Down Syndrome in the Audiology Clinic** Heather Porter (USA)

Study Findings: Speech Recognition

Two main goals:

1. Characterize maturation of masked speech recognition in 5- to 25-year-olds with Down syndrome
2. Identify factors that support masked speech recognition

Participants

- 39 individuals with Down syndrome (5-27 years)
- 63 individuals who are neurotypical (4-25 years)

Receptive Vocabulary and Working Memory

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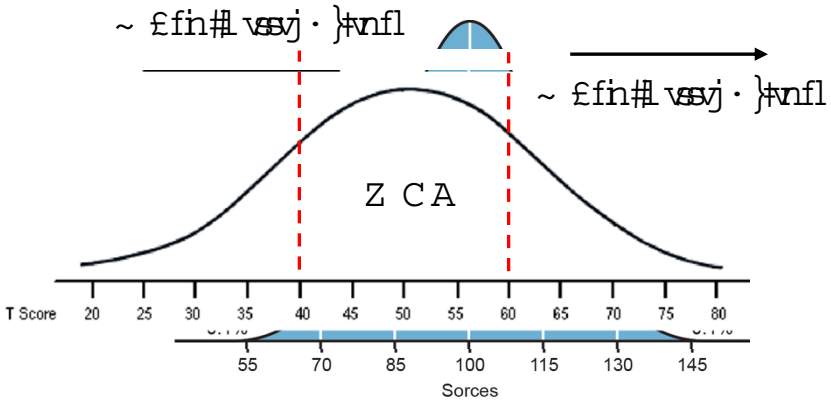
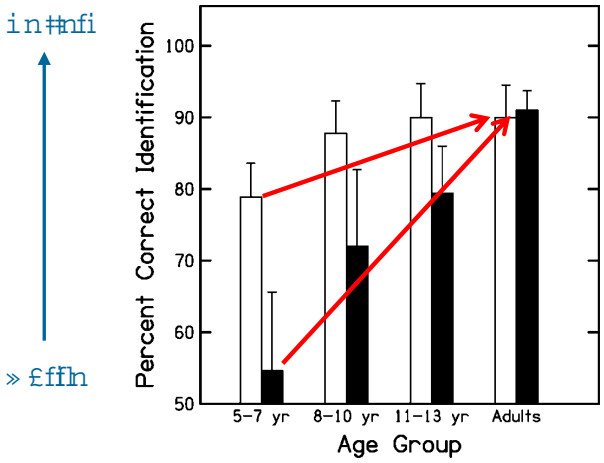


Masked Speech Perception



Speech Recognition in Competing Speech

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Age = 11 years

Age = 4 ¾

Speech Recognition in Competing Speech

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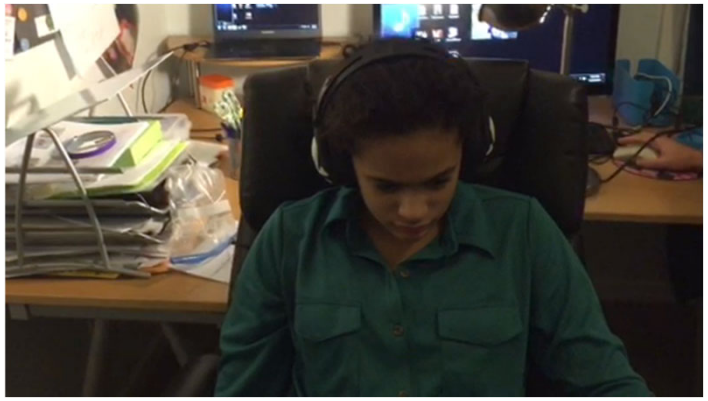
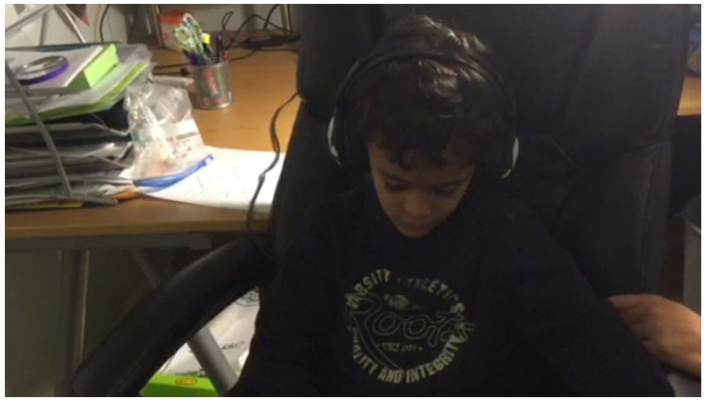
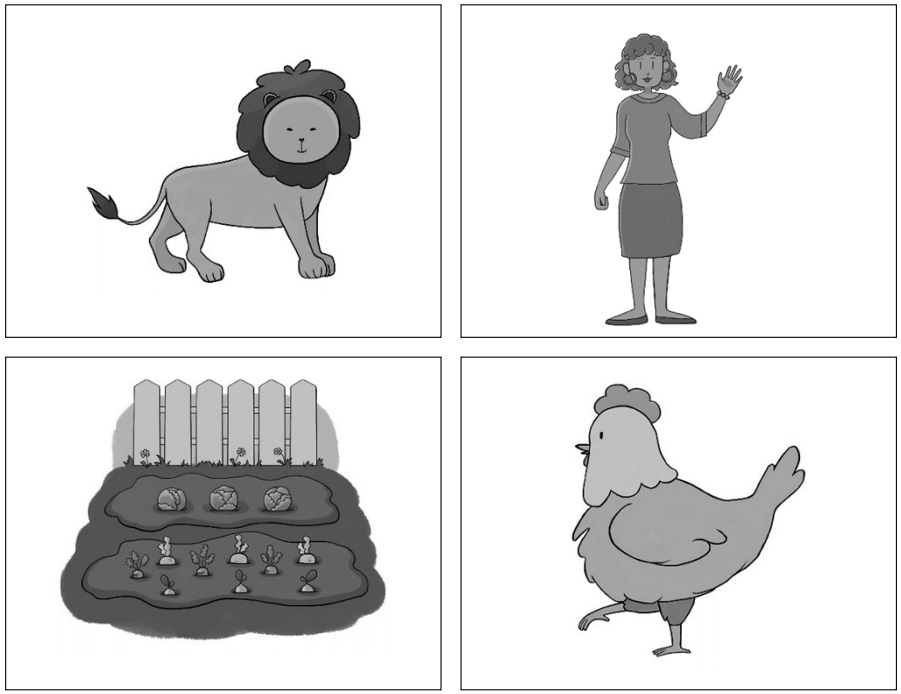


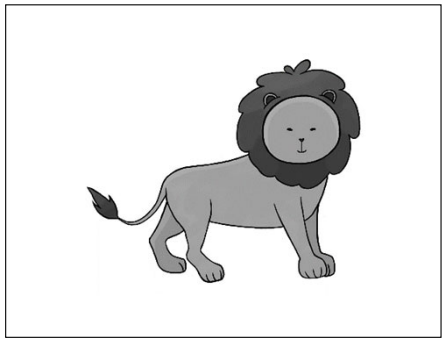


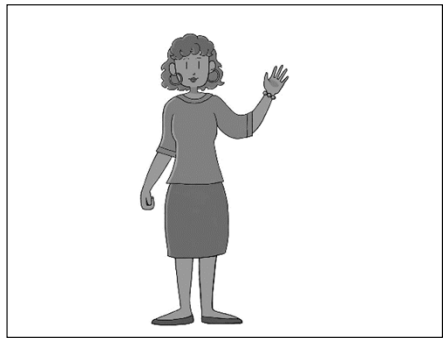
Speech recognition procedure

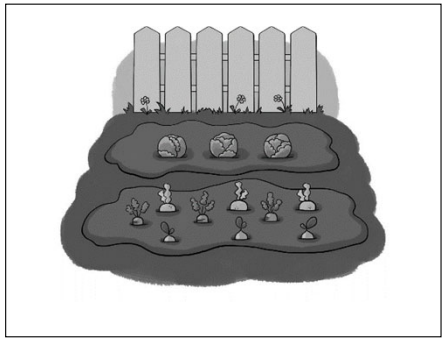
(based on Calandruccio et al., 2014)

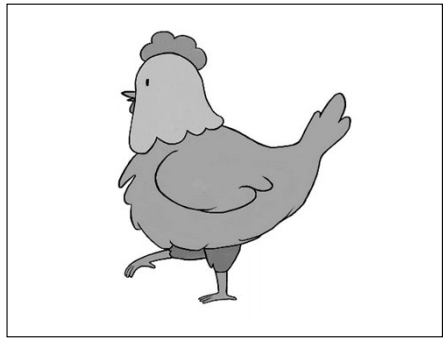


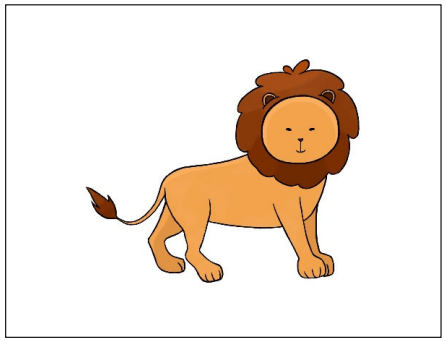
ChEgSS
Children's English and Spanish Speech Recognition Test

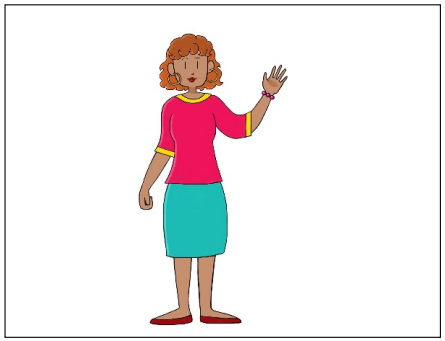


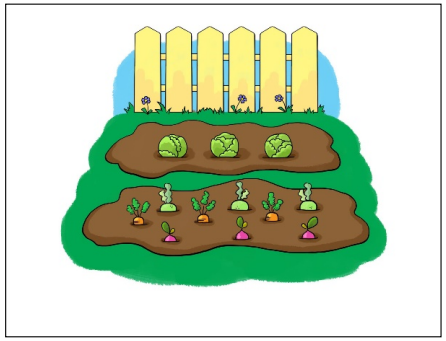


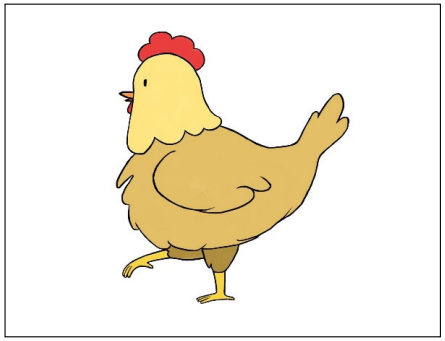












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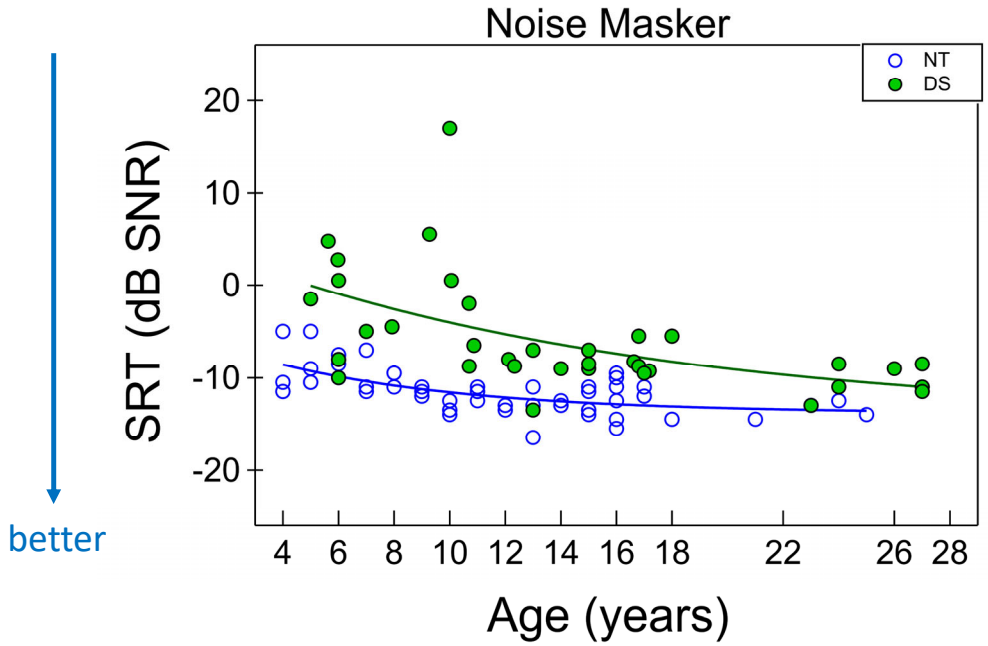

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<u>High prevalence of hearing loss</u>

- Almost universal at extended high frequencies
- Approximately half of the sample had middle ear dysfunction
- Under-utilization of amplification

<u>Greater difficulty understanding masked speech relative to peers who are neurotypical</u>

- Appear to ‘catch up’ in noise masker
- Performance gap increases with age in speech masker

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Questions?

Contact - Contact@CanadianAudiology.ca

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For those attending this session live you will receive a thank you for attending email. That is your record of attendance and CEU.

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UPCOMING WEBINAR: UPDATE ON INFANT HEARING HEALTH SERVICES IN CANADA: 2024 REPORT CARD – WEDNESDAY, NOV. 13, 2024 AT 1PM ET +

UPCOMING WEBINAR: OPPORTUNITIES AND CHALLENGES FOR ASSISTIVE LISTENING WITH BLUETOOTH LE AUDIO AND AURACAST ON WEDNESDAY, OCT 16, 2024 AT 1PM ET +

UPCOMING WEBINAR: AUDIOLOGICAL CONSIDERATIONS AND SPEECH PERCEPTION OUTCOMES FOR CHILDREN AND ADULTS LIVING WITH DOWN SYNDROME – WEDNESDAY, SEPT 18, 2024 AT 1PM ET +

ARCHIVED WEBINAR: TOOLS TO HELP DETERMINE WHEN PATIENTS SHOULD BE REFERRED FOR A COCHLEAR IMPLANT CANDIDACY EVALUATION WITH TERRY ZWOLAN – AIRED MARCH 20, 2024 AT 1PM ET +

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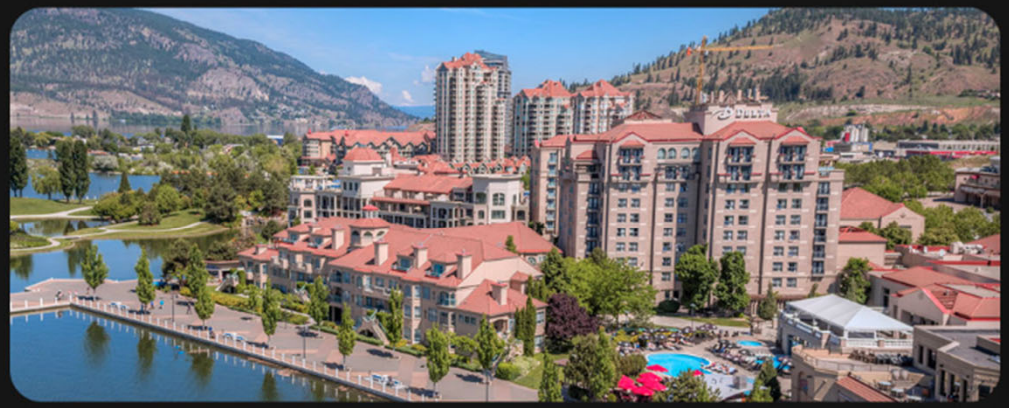
CONFERENCE2024



SAVE THE DATE

Oct 6–9, 2024

The Delta Grand Okanagan Resort → Kelowna, British Columbia

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Thank You