



# **Risk factors and etiology**

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## **Risk Factors**

- Maternal risk factors for severe microtia/atresia in China: A case-control study. (Liu et al., 2018 )
- Sociodemographic, health behavioral, and clinical risk factors for anotia/microtia in a population-based case-control study. (Ryan et al., 2019)
- Does Prematurity Play a Role in Newborn Microtia-Anotia?
   (Shehan et al., 2022)
- Maternal occupation as a nail technician or hairdresser during pregnancy and birth defects, National Birth Defects Prevention Study, 1997–2011. (Siegel et al., 2021)
- Population-based birth defects data in the United States, 2011–2015: A focus on eye and ear defects. (Stallings et al., 2018)

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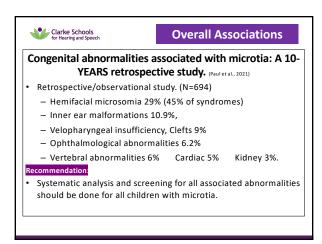
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## Suggested Risk Factors

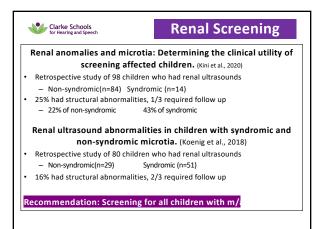
- History of miscarriage (5.5x higher with 3 or more), progesterone use, bleeding/cramping during pregnancy,
- Exposure to chemicals, such as pesticides, formaldehyde
- Prematurity
- Non-singleton birth
- One or both parents Hispanic, mother born outside the US
- Mother's binge drinking, drinking 300mg or more of caffeine daily, or smoking 5 or more cigarettes daily.
- Mother: Type 1 or Type 2 diabetes before pregnancy (Ryan et al., 2019)
- · Anemia in first trimester

## Associated Medical and Health Needs

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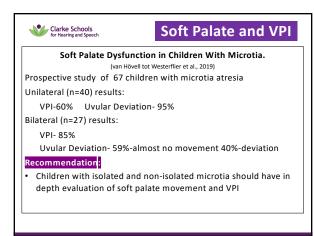
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## Clarke Schools Cr Hearing and Speech Congenital heart defects in patients with isolated microtia: evaluation using colour Doppler echocardiographic image (Guo et al., 2021) N=804 isolated microtia/atresia (ages 1-31 yrs) CHD found (n=52), with 9 having 2+ CHD – Atrial septal defect, ventricular septal defect, patent ductus arteriosus No difference based on type of microtia/atresia CHD higher in m/a population than general population CHD might not be found at birth for isolated m/a Recommendation: Doppler echocardiogram at birth

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Screened prior to any surgery



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## **Evidence Informed Considerations**

- Children with microtia/atresia should be referred to a craniofacial team for a thorough, systematic analysis and screening for all associated abnormalities related to microtia should be considered.
- Children with non-syndromic or isolated microtia with CHD may have been missed at birth. They should be screened prior to any surgery.
- Detailed family and pregnancy history should be collectedinformation may help parents.
- Children with isolated and non-isolated microtia need in depth evaluation of soft palate movement, hypernasality and VPI

## IMPACT OF AURAL ATRESIA ON HEARING AND RECOMMENDATIONS ON BONE CONDUCTION DEVICES

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Clinical consensus document for fitting non-surgical transcutaneous bone conduction hearing devices to children. (Bagatto et al., 2021)

Pediatric Bone Conduction Working Group Purpose: Address the assessment, selection, and fitting for non-surgical bone conduction devices for children under age 5

- Goal is to fit within 1 month of diagnosis
- Children as young as 2 months
- Bilateral m/a....bilateral BCDs (sequential, based on age)

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Poor speech recognition, sound localization and reorganization of brain activity in children with unilateral microtia-atresia. (Liu et al., 2021)

27 children with unilateral m/a, 27 matched controls

M/A children

- Poorer speech recognition and spatial listening
- No difference in cognitive scores
- Brain/neural activity via fMRI
  - Increase in visual networks
  - Decrease in auditory and attention networks

Acquisition limitations of bone conduction hearing devices in children with unilateral microtia and atresia Alexander, N. L., Silva, R. C., Barton, G., & Liu, Y-C. C. (2020)

- 94 children-unilateral microtia/atresia, conductive HL
- 67 completed ABR testing & counseling on benefits of BCD
- 50 fit. Mean age: 20.8 months/median age: 15.9 months
- Barriers:
  - not showing for ABR
  - communication barriers
  - insurance coverage
  - parent concern about child use

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Are children with unilateral hearing loss more tired? Carpenter, D., Dougherty, W., Sindhar, S., Friesen, T.-N., Lieu, J., & Kesser, B. W. (2022).

- Compare listening fatigue in UCHL, USNHL, LRNH
- (USNHL) (n=27) (UCHL) (n=42) (LRNH) no hearing loss

## Child report:

Overall: UNSHL > UCHL, LRNH UCHL  $\approx$  LRNH **Parent Report** 

Parents of UCHL reported higher levels of fatigue

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## **Bilateral Application of Bone-Conduction Devices**

den Besten et al., 2020:

 Bilateral BCD fitting provides more support for sound lateralization and localization than one device, though lateralization is more likely than localization

Caspers et al., 2021:

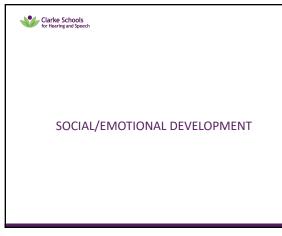
• Experienced adult BCD users could lateralize sound and 1/3 localized sound accurately

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## **Evidence Informed Considerations**

- Fit BCD within 1 month of diagnosis, age is not a barrier
- Fit 2 BCDs for bilateral m/a, sequential if needed
- Barriers need to be addressed:
  - Families from vulnerable backgrounds need more support to move from dx to fitting
  - Insurance/funding is a significant barrier
- CHL has less listening fatigue than SNHL. CHL may not be aware of their own listening needs.

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## Behavioral Adjustment of Preschool Children With and Without Craniofacial Microsomia. (Johns et al., 2021)

- Overall, similar to children without CFM
- 20% children: may demonstrate "Internalizing Behaviors" (emotionally reactive, somatic complaints, anxiety/depression, withdrawn)
- Parent concerns about stress and anxiety , such as being too dependent
- Speech and peer relationship concerns could be misdiagnosed as ASD

RX: early screening/interventions, parent intervention models ASD caution

## Behavioral performance and self-report measures in children with unilateral hearing loss due to congenital aural atresia. (Cañete et al., 2021)

- Worse when noise is directed to good ear, speech to CAA
- Required higher SNR in the hearing ear
- Sound localization impaired
- Self-reported higher classroom difficulties-especially for listening in noise or listening activities. "Quiet" classrooms aren't quiet
- Parents and older children report difficulties in all areas of SSQ
- RX: Preferential seating- not sufficient for classrooms

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### Parental Reports of Intervention Services and Prevalence of Teasing in a Multinational Craniofacial Microsomia Pediatric Study. (Johns et al., 2021)

- 169 children, average 10 years old
- · 40% parents reported their child being teased
  - 49% reported "some of the time"
  - 39% reported "almost never"
- Teasing started around age 5 (US), age 7 (South America)
- 85% teasing occurred at school
- No correlation with amplification, HL, IEP services
- Higher teasing for children with m/a and mandibular hypoplasia

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## Exploring the Experiences of Adults With Microtia: A Qualitative Study. (Hamlet & Harcourt, 2020)

- Challenges/Anxiety-hiding ear, others finding out, disclosing
- Surgery as adult isn't a quick fix for anxiety, self-esteem issues
- Adults did not view microtia as a negative impact on life
- With age, insecurity lessens. Family/friends made a difference

## Recommendations

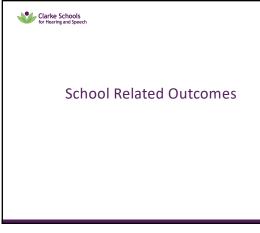
- Psychosocial supports for parents & young children considering reconstruction
- Assessment of patients/parents' expectations for reconstruction and realistic outcome counseling
- Psychosocial support/interventions related to visible differences

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## **Evidence Informed Considerations**

- Early screening and interventions are needed to address anxiety, communication impacts on peer relationships, independence, visible differences, self-esteem for children and for parents
- Collaboration with ASD professionals for appropriate diagnosis
- Classrooms are never quiet, preferential seating is not enough
- Schools play a role in supporting children before teasing starts

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Hearing-related quality of life, developmental outcomes and performance in children and young adults with unilateral conductive hearing loss due to aural atresia. (Smit et al., 2021)

29 participants, ages 7-19 years, 3 age groups -Standardized measures: quality of life, behavior, communication, overall development

- 44.8% report using amplification
- SSQ-quality of life lower than general population
- Social/Emotional, Behavior, Language-within average range
- 34% repeated a grade and 2 were in special ed. class

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## Impact of unilateral congenital aural atresia on academic Performance: A systematic review. (van Hövell tot Westerflier et al., 2018)

- Only two studies could be identified
  - High selection bias
  - No standardized outcome measures
- Other studies were excluded based on:
  - SNHL included or not specific if AA dx.
  - Type of article (ie narrative review)
  - Multiple studies using same participants
  - Missing full texts, unable to obtain

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# Outcomes for School-Aged Children with

Aural Atresia. (Hyland et al., 2020)

- 10 children with AA (8 UL) vs 10 control
- Language, reading, classroom, quality of life
- All had early amplification (5 current/7 FM) & LSL support
- Results:
- \* 90% AA  $\geq$  average Vocabulary, Language and Reading
- 70% AA ≥average functional communication
- HEAR-QL-significant differences from control group

   Experiences difficulties hearing in different situations
  - Frequency of HL having a negative impact on their feelings

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## **Evidence Informed Practices**

- Provide early amplification and intervention to improve language and academic outcomes
- AS IF-quality of life impact needs support beyond amplification
- Psychosocial Development-development of healthy selfesteem and of communication strategies for social situations
- In absence of research-address amplification, intervention and academic support and psychosocial needs for positive outcomes

Questions?

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