

From Research to Practice Using Current Research to Guide Supports for Children with Microtia/Atresia

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LEARNING OUTCOMES



Discuss important findings from recent research related to young children with microtia/atresia.



Apply findings from recent research to support families of children with microtia/atresia



Use research on social, emotional, linguistic, & academic outcomes for children with microtia/atresia to identify research influenced best practices.



(Rapin,
1979)



(van Hövell tot
Westerflier et al., 2018)

- Studies meeting research standards=**0**
- Studies that looked at language acquisition or language quality=**0**
- Overall, variables, such as SES, weren't considered
- Rx: need for prospective multidisciplinary studies to evaluate the actual impact of conductive hearing loss

- Studies meeting research standards=**0**
- Current research is sparse, inconclusive and has a significant risk of bias.
- Rx: high quality studies on the effects on academic performance are needed

Focuses, Trends, and Developments in Craniofacial Microsomia From 1992 to 2022: A Bibliometric Analysis. (Liu, Z. & Teng, L., 2023)

hemifacial microsomia; first and second branchial arch syndrome; otomandibular dysostosis; Goldenhar syndrome; oculo-auriculo-vertebral spectrum(OAVS))

1992 (16) > 2022 (60)

No.	Research areas	Count	% of 949
1	Surgery	434	45.732
2	Dentistry/Oral Surgery	243	25.606
3	Genetics Heredity	157	16.544
4	Pediatrics	100	10.537
5	Otorhinolaryngology	69	7.271
6	Ophthalmology	44	4.636
7	Clinical Neurology	37	3.899
8	Radiology Nuclear Medicine Medical Imaging	26	2.740
9	Medicine Research Experimental	24	2.529
10	Anesthesiology	20	2.107



Common Questions


What caused this?
Was it something I did?

Why didn't they
find this when I
was pregnant?


How will they do in
school?

Are there other
medical issues?

Does my child
need a BCD?


Will my child get
picked on or
bullied?







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What caused this?
Was it something I did?

What we know

- 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
- Mother's binge drinking, drinking
- 300mg or more of caffeine daily
- Smoking 5 or more cigarettes daily.
- Anemia in 1st trimester
- Mother: Type 1 or Type 2 diabetes before pregnancy



What's New?

(Chen et al., 2022)

- Genital infections during pregnancy
- Teratogenic drugs usage
- Fathers-older, smokers, have chronic diseases and expose toxins/chemicals
- Increased risk + low folic acid or age

(Lowry et al., 2023)

- Significant increase 1997-2019

(Noroña et al., 2024)

- 99.1% had hearing loss (98.5 conductive)
- 83%-lived at high altitudes (>2500-3500m)
- 19% family history
- Ecuador rate: 17 per 10,000 births

(Shehan et al., 2022)

- More likely to be born prematurely
- Race- Asian+M/A>prematurity





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**Why didn't they see this when I was
pregnant?**

Studies

- Lamanna, B., Dellino, M., Cascardi, E., Rooke-Ley, M., Vinciguerra, M., Cazzato, G., Malvasi, A., Vitagliano, A., Nicolì, P., Di Cosola, M., Ballini, A., Cicinelli, E., & Vimercati, A. (2023). Efficacy of Systematic Early-Second-Trimester Ultrasound Screening for Facial Anomalies: A Comparison between Prenatal Ultrasound and Postmortem Findings. *Journal of Clinical Medicine*, 12(16), 5365.
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- Zhang, X., Zheng, W., Feng, Y., Yu, N., Qin, J., Li, K., Yan, G., Zou, Y., & Li, B. (2023). The role of MRI in the prenatal diagnosis and classification of fetal microtia. *European Radiology*.
<https://doi.org/10.1007/s00330-023-09816-5>

Findings

- Ultrasound: 2D preferred, 2nd trimester is the best time to observe the fetal ear morphology
- MRI accuracy: microtia (93.68%) and atresia (93.10%)





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Are there other medical issues?

What we know

Screen for:

- Renal anomalies
- Doppler echocardiogram at birth and screened prior to any surgery
 - CHD higher in m/a population but not be found at birth for isolated m/a
- In-depth evaluation of soft palate movement and VPI
- Vertebral anomalies
 - rib graft, could lead to chest wall deformity complications



What's New?

- Cholesteatoma
 - Atresia: 1.7% (4/238) in atresia
 - Stenotic EAC 43(203/473)
 - <5% of MA or stenosis-most were stenosis (Kalmanson et al., 2023)
- Spinal Anomalies (Alexander et al., 2023)
 - 425 microtia patients-18.4% spinal imaging
 - Scoliosis, fusion defects, rib deformities
 - Scoliosis-increased rate for syndromes
- Genitourinary malformations (Gao et al., 2023)
 - 163/3143 patients (5.2%) had malformations
 - hydronephrosis, renal cyst, and abnormal testicular development
- Hematuria (Sun et al., 2023)
 - Girls with > incidence than boys and control
- Velopharyngeal insufficiency (A. E. Tio et al., 2023)
 - CFM, VPI and hypernasality: 12.5% and 55%





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Amplification Parents' Needs

What we know

- 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.



Studies

- Hearing impairment and ear anomalies in craniofacial microsomia: A systematic review. (Rooijers et al., 2022)
- Incidence of audiologic or otolaryngologic evaluation in patients with external ear anomalies. (Patel et al., 2023)
- Relationship between congenital malformation of the outer ear and hearing. (Zhang et al., 2023).
- Optimal choice for improving the hearing in children with unilateral microtia and atresia: Softband or adhesive adapter? (Liu et al., 2022).
- Signal transparency of remote microphone technology in pediatric bone conduction device users. (Sanchez et al., 2023).
- Early information and clear recommendations to parents positively influence the use of bone anchored hearing systems for young children with unilateral microtia/atresia. (Kazemir et al., 2022).
- The role of bone-anchored hearing devices and remote microphones in children with congenital unilateral hearing loss. (Lazzerini et al., 2023)
- Anxiety, depression, stress, and self-esteem in Turkish parents of children with microtia. (Turhan et al., 2023)
- Early experiences of parents of children with craniofacial microsomia. (Johns et al., 2024).



What's New?

- **Adhesive vs. Softband (Liu et al., 2022).**
 - **Functional gain: (adhesive) 20.63 dB HL**
 - **Functional gain: (softband) 26.39 (3.15) dB HL**
- **Remote Microphone Use (Sanchez et al., 2023)**
 - **Remote mic: Significant Improvement**
 - **Coupling of receiver to remote mic negatively affects signal transparency-no improvements**
- **BCD for Unilateral Conductive HL**
 - **Hearing threshold-moderate improvements**
 - **Speech recognition-significantly improvement**
 - **Localization-wide individual variations.**
- **Parent Needs**
 - **Parents-varied experiences with rx for amplification**
 - **Children with Unilat M/A-not using BCD full-time**
 - **Earlier info./strong recommendations> earlier trial,ongoing usage.**
 - **Many parents experience stress and depression. Information and intervention re: concerns for future help.**



Evidence Informed Considerations

- Consider using personal remote mic only (check!!!)
- Monitor children with stenosis EAC actively for cholesteatoma
- Screen for vertebral anomalies before rib graft reconstruction.
- There is a need for greater consistency in what, when, and how MA parents receive information and recommendations.
- Refer to a craniofacial team for a thorough, systematic analysis and screening for all associated abnormalities related to microtia



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- NYS EHDI Advisory Committee
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- Employed by Clarke Schools for Hearing and Speech