

Psychological Aspects and Current Options for Patients with Microtia and Atresia

An Educational Audiology Association Presentation

Sponsored by the



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Presented by: Melissa Tumblin

A parent of a child born with right sided Microtia and a unilateral hearing loss caused by Atresia.

Parent Advocate

Founder of the Microtia and Atresia Support Group

Founder of Ear Community, a 501(c)(3)

Board of Director Member for Hands & Voices

Past Family Advisory Council Member for Children's Hospital Colorado

Medical Device Marketing Professional

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Presentation Goals

1. Describe the difference between congenital microtia and congenital aural atresia
2. Describe the impact of unilateral hearing loss and psychological aspects on the pediatric patient and adults with Microtia and Atresia
3. Describe the current options for helping pediatric patients become more audible with the use of bone conduction hearing devices and FM systems

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Microtia and Atresia in Review



- Microtia is a congenital deformity affecting the outer ear (pinna) where the ear does not fully develop during the first trimester of pregnancy.
- Aural Atresia is the absence or closure of the external auditory ear canal.

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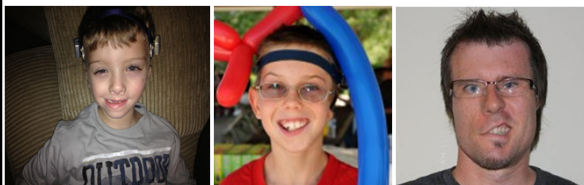
Microtia and Atresia in Review



Grade I Grade II Grade III Grade IV

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Microtia and Atresia in Review



Hemifacial Microsomia

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Microtia and Atresia in Review

- 1 out of every 6,000 to 12,000 births.
- More common in males.
- More common for the right ear to be affected.
- Microtia and Atresia can be unilateral or bilateral.
- Conductive hearing loss due to Atresia.
- Ethnicities affected by Microtia/Atresia.
- In 2012, approximately 4 million babies were born in the United States with about 500 to 600 of these births having Microtia and Atresia. It is estimated that 39,000 people live in the United States with Microtia/Atresia.
- Genetic???

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Microtia and Atresia in Review



Microtia ears are beautiful ears

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Microtia and Atresia in Review



Prosthetic Ear

- Realistic
- Non-surgical option
- Temporary

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Microtia and Atresia in Review



Rib Graft: The Gold Standard

- Age 5
- 3 or > surgeries
- Biological material

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Microtia and Atresia in Review



Medpor

- Age 3
- Achieved in one stage
- Polyethylene

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Microtia and Atresia in Review



Canalplasty (atresia repair)

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Microtia and Atresia in Review

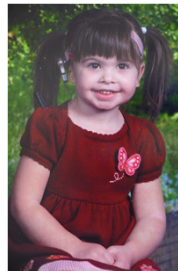
New technology around the corner...

1. Stem cell research
2. 3D printing



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My Child Has Microtia and Atresia – Now What?



3.5 years old, Grade III Microtia and Atresia of the right ear.

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Experiences with Microtia/Atresia

“My daughter, Eden, was born with RMA in September (2010). **At our first ENT visit ‘the best’ doctor in Omaha wasn’t going to mention a BAHA until I brought it up.** Eden is being fitted for her first BAHA in 2 weeks.”

- Jay, Nebraska (9/2010)

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Experiences with Microtia/Atresia

“...My daughter Makayla was born with right ear microtia and atresia. When she was born, I had no idea what was going on, what caused her condition or [found] physicians that were familiar with Microtia. No one advised me on hearing devices or options other than reconstructive surgery. **After 9 years, Makayla is finally getting a bone conductive hearing device. I am appalled that no audiologists and ENTs have suggested them to us before.**”

- Cate, Texas (10/2012)

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Experiences with Microtia/Atresia

“I am both a professional in the field...and the parent of a child with bilateral microtia/atresia. This population of children are, overall, overlooked, undeserved, poorly researched, and under amplified. **That statistics for the incidence of microtia/atresia are ridiculously inaccurate- and yet no one is correcting them. The advice given to parents is often inadequate and often outdated.** The youngest child to date that we have fit with a baha was 21 days old. The second youngest was about 6 weeks old. If the advice for other types of hearing loss focus on the earlier the better and two are better than one, why are the families of these children not counseled the same way?”

- Meredith, New York (10/2012)

Experiences with Microtia/Atresia

“I wanted to take the time to say how wonderful both the Ear Community and the Microtia and Atresia Community Support groups are. In addition, they have been extremely helpful, informative, as well as comforting. My daughter was born with Atresia in both ears. **I work as a school psychologist and know the implications of hearing loss, specifically on speech development, academic performance, and social skills. The psychologist in me knew she would be just fine, especially with all the available resources and technology. The parent in me, as you know experienced a number of emotions. Life is challenging and no parent wants their child to struggle.**

- Lorryn, New Jersey (5/2013)

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Experiences with Microtia/Atresia

"Once I found out about a BAHA, I called my hearing center and got a loaner within a month. **The Audiologists say the earlier the better and nobody ever even mentioned a hearing device for babies.**"

- Casey, Texas (11/2012)

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Experiences with Microtia/Atresia

"My daughter is 3 1/2 years old and for the first time today I feel like we have direction and are getting somewhere with her microtia and atresia. **Today Sophia was fitted for a soft band Baha which we weren't informed was an option....I was also able to get in touch with a doctor at Shriners Hospital who made us feel very hopeful about having medpor surgery done which again I did not know about.**"

- Melissa, Illinois (5/2011)

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Experiences with Microtia/Atresia

"I thought there was no one like me that was born with Microtia. I ALWAYS wanted to meet people just like me. For many years up until my age now (33) in Columbia, SC I thought that I was alone. I don't have much anger but I am frustrated with what I have going on and that's hearing."

- Angela, South Carolina (12/2012)

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Experiences with Microtia/Atresia

"In spite of believing for my entire life that I was deaf in my left ear, I learned there may now be a possibility of hearing! There is more testing required, and will require the BAHA. I am not sure what will come of this. **I am still in shock over this news, and can only imagine how life changing this can be for others!"**

- Angela, Canada (10/2011)

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Experiences with Microtia/Atresia

"We both cried when we realized that Natalie would be able to meet other children just like her and that I would be able to meet other parents who share the same experience. Natalie has chronic ear infections and I was worried that she couldn't hear. **For the past five years, I was told by our doctors that Natalie could not have a Baha until she was five years old.** Even though I was sad to learn that she could have been aided sooner with a soft band at the picnic, I am happy to say that Natalie finally does have a Baha today and she loves it!"

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- Bliss, Colorado (10/2012)

Experiences with Microtia/Atresia

"Our 3 year old daughter...has unilateral Microtia. We have been seeing Dr. [] at [] since she was born. It wasn't until about a week ago we found out on our own about Medpor as well as your website. **We wished we would have been told about other options besides rib graft and prosthetics.**

- Scott, Colorado (8/2012)

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Experiences with Microtia/Atresia

"My daughter, Clark, is now 21 months old and has RMA. She does not have a BAHA yet, but we are working on it. **It is not "standard practice" with her group of audiologists to treat unilateral loss with a BAHA. We have been told that there is not enough research to support this, but I disagree.** We have an appointment with her ENT on Wednesday, so we hope to get closer.

- Janae, North Carolina (5/2013)

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Experiences with Microtia/Atresia

"Noah has RMA and he passed his newborn hearing test too. Later as a toddler I realized he was having hearing and speech problems. **We didn't find out he had atresia till he was almost five. Because of that newborn hearing screen I felt that not only did it give a false sense of hope but it delayed in getting the help he needed.**"

- Amanda, Texas (1/2013)

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Experiences with Microtia/Atresia

"When my daughter was born, the Pediatrician saw her ear during the exam but she "passed" the hearing test in both ears! We even received a copy of the results. **She has stage three [Microtia and Atresia] with no canal opening at all, so it was obvious she couldn't have passed.**"

- Misty, Arizona (1/2013)

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Where do I begin?

1. Diagnosed with Microtia and Atresia
 - a. The mother should never blame herself
 - b. Informed about helpful resources
2. Renal ultrasound and EKG/ECG
3. Diagnostic assessments:
 - a. New born hearing screening
 - b. ABR and (AOE) Auto-acoustic evaluations
4. Meet with an ENT and Audiologist
 - a. Discuss Microtia and Atresia
 - b. Discuss options and hearing solutions
 - c. Discuss monitoring hearing loss

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Unilateral Hearing Loss

Is unilateral hearing loss misunderstood?

• YES!

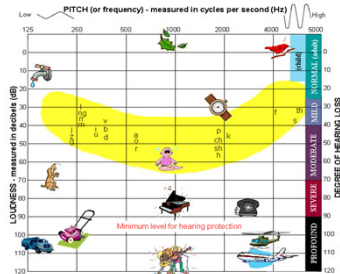
- Parents
- Patients

Confusion, whispers, localizing sound, sounds never heard before, not realizing the sounds of life, available technology...

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Unilateral Hearing Loss

- The critical years of development (birth to 3)



Speech banana chart sourced from the AAA, www.Audiology.org

Unilateral Hearing Loss

Is one good ear good enough?

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Unilateral Hearing Loss

“The Left and Right Ears Are Not Created Equal”

By, Dr. Yvonne Sininger, PhD and Dr. Barbara Cone-Wesson, PhD
(Science, 10/10/2004)

“We always assumed our left and right ears worked exactly the same way. As a result, we tended to think it didn't matter which ear was impaired in a person. Now, we see that it may have profound implications for the individuals speech and language development.”

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Unilateral Hearing Loss

“The Left and Right Ears Are Not Created Equal”

By, Dr. Yvonne Sininger, PhD and Dr. Barbara Cone-Wesson, PhD
(Science, 10/10/2004)

“We were intrigued to discover that the clicks triggered more amplification in the baby's right ear, while the tones induced more amplification in the baby's left ear. Our findings demonstrate that auditory processing starts in the ear before it is ever seen in the brain. Even at birth, the ear is structured to distinguish between different types of sound and to send it to the right place in the brain.”

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Unilateral Hearing Loss

"Unilateral Hearing Loss in Children: Challenges and Opportunities"

By, Oyler, R. and McKay, S.
(The ASHA Leader, 1/22/2008)

"Studies show that even before they enter school, children with UHL are at risk for speech and language delays. Although the average age for first words was found to be within normal limits (12.7 months), the average age for the first two-word utterances was found to be significantly delayed in children with UHL (Kiese-Himmel, 2002)."

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Unilateral Hearing Loss

"Supplement to the JCIH 2007 Positions Statement: Principles and Guidelines for Early Intervention After Confirmation that a child is Deaf or Hard of Hearing"

(American Academy of Pediatrics, Vol. 131, No. 4, 4/2013, pg.9)

"Research indicates that there are sensitive periods for the development of auditory skills and spoken language, specifically, the first 5 years of a child's life are critical for development in these areas. To optimize this short time period of a child's life, families and infants/children who are D/HH require the highest level of provider skills at the very beginning of the child's life."

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Spatial Hearing/Sound Localization

"Why Our Two Ears Process Sound Differently"

By, Dr. Neil Bauman, Ph.D.
(4/2/2006)

"Therefore, only having one ear detracts from our ability to hear, understand, and appreciate sound."

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Spatial Hearing/Sound Localization

“Unilateral Hearing Loss – How Serious Is it?”

By, Jane Madell, Editor
(Hearing and Kids, 3/2012)

“Brain research has demonstrated that language is a left brain activity and the majority of fibers from the right ear go to the left side of the brain so, if a child is going to have a hearing loss in one ear, it would be better if it were the left one. (If only we could control this.)”

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Spatial Hearing/Sound Localization

“Understanding Single Sided Deafness:
Evaluation and Treatment for Professionals”

By, Dr. George Cire, AuD
(Online Cochlear HOPE Series 8/2012)

“First, you lose the ability to have spatial hearing, or what we traditionally think of as localization in the horizontal plane. Our two-eared auditory system is precisely oriented for using specific cues that allow us to localize sound, and we use a two-ear input system to make that happen. When one ear is taken out of the equation, it creates some confusion within the brain.”

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Spatial Hearing/Sound Localization

“The loss of spatial hearing not only heightens communication concerns, but it also creates safety concerns.”

By, George Cire, AuD
(Online HOPE Series 8/2012),



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Challenges in the school classroom

“Unilateral Hearing Loss in Children: Challenges and Opportunities”

By, Bess & Thorpe (1984), Oyler, Oyler, & Matkin (1988) and McKay, S.
(The ASHA Leader, 1/22/2008)

“Children with UHL were approximately 10 times more likely to fail a grade than their normal hearing peers.”

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Challenges in the school classroom

“One third of children with unilateral hearing loss had to repeat a grade by 3rd grade.”

As demonstrated by, Bess et al (1998), Bess et al (1986)
English and Church (1999)

“While a unilateral hearing loss may not be the same kind of emergency that a bilateral hearing loss is, auditory brain development is just as critical for these children.” By, Jane Madell, Editor, (Hearing and Kids, 3/2012).

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Challenges in the school classroom

“Children with unilateral hearing loss are more likely to struggle in school”

By, Dr. Judith E. C. Lieu, MD, ENT, July 20, 2013,
(Washington University School of Medicine)

“...an invisible impairment. That is the problem, because the child appears normal, it is easy to forget that they have an impairment. A child with unilateral Atresia who fails to pay attention in class or keeps talking when he or she should be quiet, might be labeled a problem student when the real issue is that he or she simply does not hear the teacher.”

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Challenges in the school classroom

“Children with unilateral hearing loss are more likely to struggle in school”

By, Dr. Judith E. C. Lieu, MD, ENT, July 20, 2013,
(Washington University School of Medicine)

“Compared with children who have Atresia of the left ear, those with the condition on the right may be more likely to have problems learning in school.” - In previous published studies, nearly 35 percent of those with right unilateral hearing loss had repeated a grade, compared with fewer than seven percent of children with left unilateral hearing loss.”

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Challenges in the school classroom

IEPs...Why do so many families not know about one?

IFSP Transition (EI)

IEP and 504 school plans

...Families need to be better informed.

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Treatment Options

Bone Anchored Hearing Devices and Systems

1. Bone Anchored Hearing Aid (BAHA)
2. Bone Conduction Hearing Aid (BCHA)
3. Bone Conduction Hearing Device
4. Osseointegrated Device (OSI)
5. Sound Processor




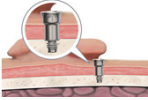
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Treatment Options

Bone Anchored Hearing Devices and Systems

- Implantable Surgery after 5 years of age
- Mandated by FDA

Permission to post the following images has been granted by Oticon Medical.
Permission to post Oticon Ponto Pro snapped in has been granted by patient.
Healing cap image was sourced from www.entkent.com



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Treatment Options

Bone Anchored Hearing Devices and Systems



Cochlear BP100
BAHA3

Oticon Medical
Ponto Pro

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Treatment Options

Bone Anchored Hearing Devices and Systems

Softband Head Band

- Can be used on children under 5 years of age



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Treatment Options

Bone Anchored Hearing Devices and Systems

Why do professionals not agree with each other?

- Standard or personal preference?
- Recommended age?
- Regional?

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Helpful Tips for Wearing a BAHA

Try not to get frustrated and don't give up!

1. Make it fun
2. Better when in upright position
3. Take the stress out (what's easiest for you works).
4. Stay in control

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Helpful Tips for Wearing a BAHA



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FM Systems

- Helps with speech
- Helps in the classroom setting
- Helps allow children to meet and exceed expectations in the classroom



Pictured: Oticon Medical Ponto Pro w/Amigo FM System and Phonak Inspiro FM System with Oticon Ponto Pro

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Other Helpful Technology

Helpful Hearing Devices for Conductive Hearing Loss

- Oticon Medical Ponto, Ponto Pro, and Ponto Pro Power, www.OticonMedical.com
- Cochlear BP100, BP110 Power, www.Cochlear.com
- Sophono Alpha 1 and Alpha II, www.Sophono.com
- MedEl Soundbridge and Bonebridge, www.Medel.com/us
- Sonitus Medical Soundbite, www.SoundBiteHearing.com
- BMH Tech Contact Mini, www.BHM-TECH.at
- TransEar TransEar, www.TransEar.com
- FM Systems by Phonak Inspiro, www.Phonak.com and the Oticon Medical Amigo, www.OticonMedical.com

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Psychological Aspects

Facial challenges/differences: not just dealing with hearing loss...

- Public stares, teasing, low self esteem or lack of confidence, ashamed to ask for help.
- ADHD or not?
- Employer discrimination?
- Surgery: guilt and making decisions
- * One good ear is good enough...you'll be fine.

Helpful to find a mentor or someone in the same situation.

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Ear Community

- A 501(c)(3) global nonprofit organization that helps children and adults born with Microtia and Atresia since August 16, 2012.
- Launched on February 8, 2012

- Emotional Support
- Connect with others
- Learn about options
- Helpful resources
- Learn about Hearing Loss

www.EarCommunity.com

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2013 Ear Community Picnics



Orlando, Florida – April 27, 2013
Columbus, Ohio – June 22, 2013
London, England – June 29, 2013
Barcelona, Spain – July 6, 2013
Chicago, Illinois – July 13, 2013
Broomfield, Colorado – July 27, 2013
Los Angeles, California – August 10, 2013

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Ear Community



Global Microtia and Atresia Summer Family Picnics

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Ear Community Testimonials

"My family just wanted to give you and your team the biggest of thanks for this past weekend's family picnic in Orlando! Not only did the girls have a great time with all the activities and food, my wife and I found it very informative and well worth the time. Seeing the look on my daughter's face when she tried on the BAHA device from Ociton for the first time brought tears to our eyes. The coolest part was when the representative from Cochlear allowed us to experience what it sounds like to her wearing the device. A special thanks to Dr. Reinisch who sat with us for nearly 30 minutes discussing the Medpor technique. We were totally against that prior to meeting with him and he has opened our eyes to the possibility of going that route. You have no idea how much your group, website and information means to us and I want to personally thank you for all that you do! I wanted to share a photo of my beautiful daughter, Eva enjoying her bubbles and sporting her Microtia Kids Rock shirt. She had to wear it all day at the picnic and then as soon as we got home, Mom had to wash it so she could wear it to school today."

Mark and Lisa Johnson,
Florida (4/2013)

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Ear Community Gives Back

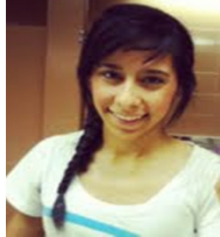
BAHA Recipients



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Ear Community Gives Back

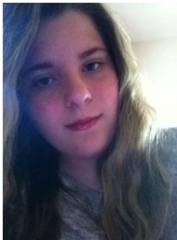
College Scholarship Recipients



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Ear Community Gives Back

Financial Assistance



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Helpful Documents

Document: Comparative Review for the Cochlear BP100/BAHA3, Oticon Ponto Pro, and Sophono Alpha 1 (S) Soft Band Head Band BAHA Models, August 2011, revised October 2011. Posted to various surgeon websites and on hand at medical centers that specialize in Microtia and Atresia repair and can be found on the Ear Community website under the Hearing Loss menu tab, by Melissa Tumblin

Document: "10 Helpful Tips for BAHA Wearers," posted to various websites, and support groups that help with Early Intervention service programs, and can be found on the Ear Community website under the Hearing Loss menu tab, September 2011, by Melissa Tumblin

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Helpful Articles

Article: "Falling Through the Cracks With Microtia/Atresia," published by Hands & Voices, Winter 2010, Volume XIV – Issue 2 of "The Communicator," and can be found under the About menu tab of the Ear Community website, by Melissa Tumblin.

Link to article:

http://www.handsandvoices.org/articles/early_intervention/V14-2_cracks.htm

Article: "Mixed Feelings: Making Surgical Decisions for Children With Microtia and Atresia," summer 2011, Volume XIV - Issue 4, pg. 19 of the National issue for "The Communicator" published by the Hands & Voices Organization, by Melissa Tumblin

Article: "A Little Love Can Go a Long Way" in the "Facing Forward" newsletter for the AboutFace Organization's international newsletter, April 2012, Vol. 10, ISS.2, by Melissa Tumblin

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Press Releases and Interviews

Press Release: On February 8, 2012, a press release announcing the launch of the Ear Community website was picked up by multiple business and news wire entities such as: Healthy Hearing, AudiologyOnline, Hearing Review, Advance for Hearing Practice Management, Hearing Health Matters, etc...

Interview: Interview with "Melissa Tumblin, Founder, Ear Community" through AudiologyOnline, archived, July 16, 2012. Link to interview:

<http://www.audiologyonline.com/interviews/interview-with-melissa-tumblin-founder-6955>

Webinar: Accredited Educational session through AudiologyOnline (1 CEU), course #22454 "Microtia and Atresia: A Parent's Perspective", April 23, 2013, archived link: <http://www.audiologyonline.com/audiology-ceus/course/microtia-aural-atresia-parent-s-22454>

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References

Photos – permission granted by parents and adults pictured. Butterfly image sourced from Bugguide.net. Microtia grades courtesy of Dr. Sheryl Lewin (Medpor surgeon). Medpor photos courtesy of Dr. Sheryl Lewin. Rib Graft photos courtesy of Dr. Burt Brent. Canalplasty photo courtesy of Dr. John Reinisch (Medpor) and Dr. Joseph Roberson (ENT/Otologist). Prosthetic ear photos courtesy of Greg Gion.

Shared Experiences – commentary from Ear Community and the Microtia and Atresia Support Group, slides 15 – 26.

"The Left and Right Ears Are Not Created Equal," by, Dr. Yvonne Sininger, Ph.D (Science, 10/10/2004), slide 31.

"Unilateral Hearing Loss in Children: Challenges and Opportunities," by, Oyler, R. and McKay, S., (The ASHA Leader, 1/22/2008), slide 32 and 38.

"Supplement to the JCIH 2007 Positions Statement: Principles and Guidelines for Early Intervention After Confirmation that a child is Deaf or Hard of Hearing" (Pediatrics, The Official Journal of the American Academy of Pediatrics), Vol. 131, No. 4, 4/2013, pg.9), slide 33.

"Why Our Two Ears Process Sound Differently," by, Dr. Neil Bauman, Ph.D, (4/2/2006), slide 34.

"Unilateral Hearing Loss – How Serious Is It?" by, Jane Madell, Editor, (Hearing and Kids, 3/2012), slide 35 and 39.

"Understanding Single Sided Deafness: Evaluation and Treatment for Professionals," by, Dr. George Cire, Ph.D, AuD, (Online HOPE Series 8/2012), slide 36 and 37.

"Unilateral Hearing Loss in Children: An Update for the 1990s." by English, Christina, and Church, Gerald. Language, Speech, and Hearing Services in Schools 30 (1):26–31, slide 39.

I am hopeful that we may continue to learn from one another and collaborate in hopes of making things easier for the next family who has a child born with Microtia and Atresia.

Especially, children (and adults) with unilateral hearing loss.

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Thank you!

Ear Community

www.EarCommunity.com

E-mail: EarCommunity@gmail.com

The Microtia and Atresia Support Group on Facebook

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