HEAR MAPS a New Classification for Congenital Microtia/Atresia Based on the Evaluation of 742 Patients

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ABSTRACT

Objective:
- Describe anatomical and radiological findings in 742 patients evaluated for congenital aural atresia and microtia by a multidisciplinary team.
- Develop a new classification method to enhance multidisciplinary communication regarding patients with congenital aural atresia and microtia.

Study Design: Retrospective chart review with descriptive analysis of findings.

Setting: Multidisciplinary tertiary referral center

Patients: Patients with congenital atresia and microtia evaluated from January 2008 to January 2012.

Intervention: Data analysis and description of new classification method for congenital atresia and microtia.

Results: We developed a new classification method based on the acronym HEAR MAPS (Hearing, Ear [microtia], Atresia grade, Remnant earlobe, Mandible development, Asymmetry of soft tissue, Paralysis of the facial nerve and Syndromes). We used this method to evaluate 742 congenital atresia and microtia patients between 2008 and January of 2012. Grade 3 microtia was the most common external ear malformation (76%). Pre-operative Jahrsdoerfer scale was 9 (19%), 8 (39%), 7 (19%), and 6 or less (22%). Twenty three percent of patients had varying degrees of hypoplasia of the mandible. Less than 10% of patients had an identified associated syndrome.

Conclusion:
Patients with congenital aural atresia and microtia often require the intervention of audiology, otology, plastic surgery, craniofacial surgery and speech and language professionals to achieve optimal functional and aesthetic reconstruction. Good communication between these disciplines is essential for coordination of care. We describe our use of a new classification method that efficiently describes the physical and radiologic findings in microtia/atresia patients to improve communication amongst care providers.

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3. Park S. "Aural Atresia of the External Ear", Available at http://emedicine.medscape.com/article/845632-overview#showall
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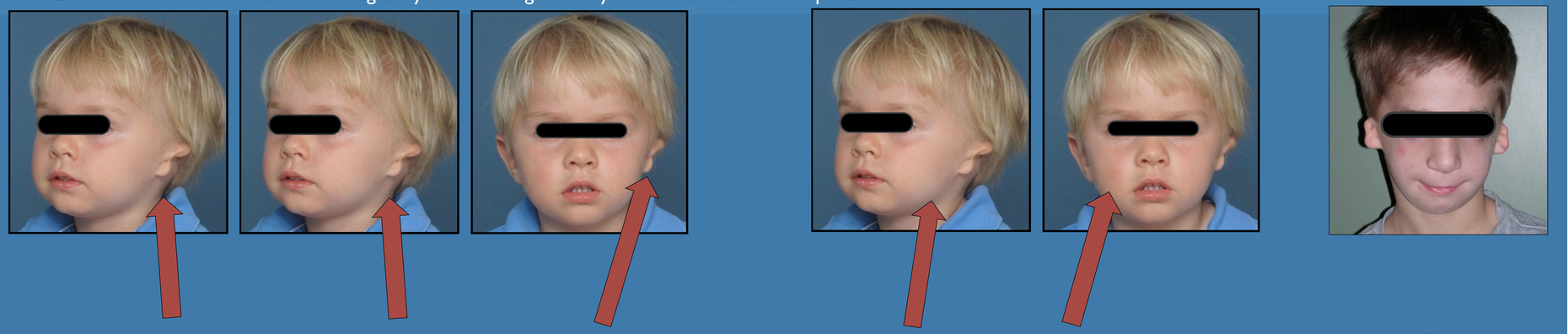
INTRODUCTION

Patients with congenital aural atresia and microtia are inherently complex and require the services of multiple medical specialties[1]. Unfortunately, communication between providers is not always optimal due to subspecialization and geographical separation. The multitude of classifications systems used by different subspecialties compounds this problem[2,3]. Perhaps the most widely used atresia classification method, the Jarhsdoerfer's 10-point scale, only discounts one point if the external ear is abnormal. It has no reference to the degree of malformation or any associated craniofacial involvement[3]. Similarly the plastic surgery literature presents multiple classification methods (Marx, Weerda, Tanzer, Fukuda, Firmin, Aguilar and Jahrsdoerfer) that describe abnormalities of the pinna but do not address concurrent abnormalities of the external auditory canal or middle ear[3]. At our institution the reconstructive team consists of an otologist/neurotologist, a plastic surgeon, a craniofacial surgeon, and a team of audiologists; all of whom have separate offices and convene in the operating theater. As communication is essential for coordination of care we realized that we needed to improve and streamline the interaction amongst team members and sought to create a new method building upon the strength of existing classifications. We created a classification based on the acronym HEAR MAPS (Hearing, Ear [microtia], Atresia grade, Remnant earlobe, Mandible development, Asymmetry of soft tissue, Paralysis of the facial nerve and Syndromes) and have used it to evaluate patients since 2008 with several modifications during the years resulting in the system described in this report.

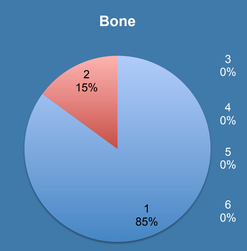


H E A R M A P S

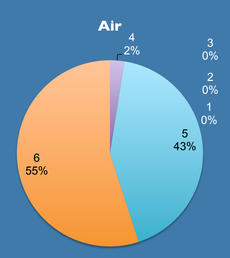
Hearing:
We record the patient's PTA2 (Average of thresholds at 500, 1000, 2000 and 3000Hz) for both bone and air thresholds for both ears.

Hear Bone/Air

Grade 1	0-20
Grade 2	20-30
Grade 3	30-40
Grade 4	40-50
Grade 5	50-60
Grade 6	>60

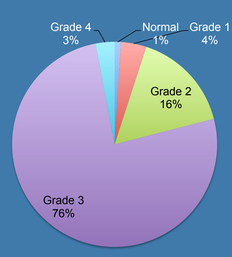


Average Bone PTA2: 12.98

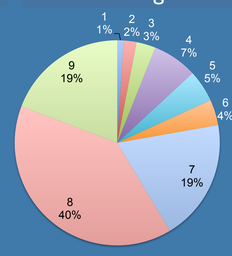


Average Air PTA2: 63.11

Ear (microtia):
We use the modified Marx's 4-point scale for the classification of microtia[3].

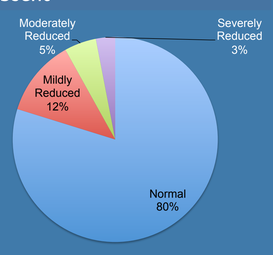


Atresia Score:
We use the Jahrsdoerfer 10-point grading scale based on CT findings[2].



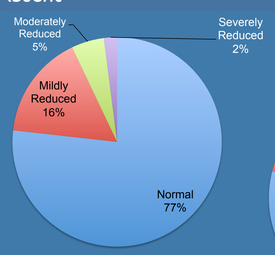
Remnant Ear Lobe

Grade 1	Normal
Grade 2	Mildly Reduced
Grade 3	Moderately Reduced
Grade 4	Severely Reduced/ Absent



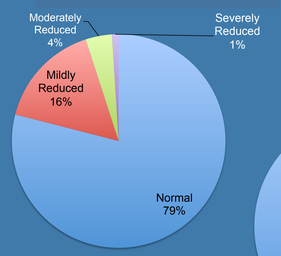
Mandible Asymmetry

Grade 1	Normal
Grade 2	Mildly Reduced
Grade 3	Moderately Reduced
Grade 4	Severely Reduced/ Absent

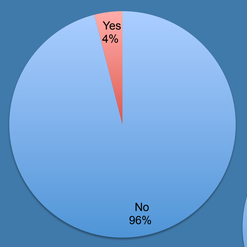


Asymmetry Soft Tiissue

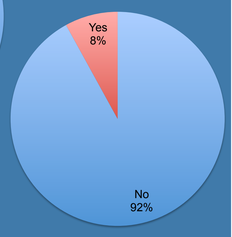
Grade 1	Normal
Grade 2	Mildly Reduced
Grade 3	Moderately Reduced
Grade 4	Severely Reduced/Absent

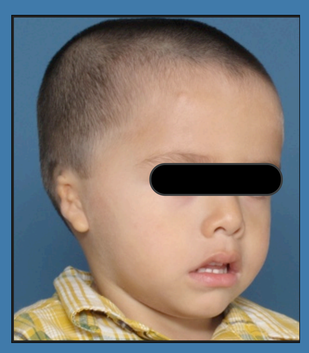


Paresis Facial Nerve:
We use the House-Brackmann facial nerve grading scale. Facial nerve paresis should alert the otologist to abnormal anatomy of the facial nerve

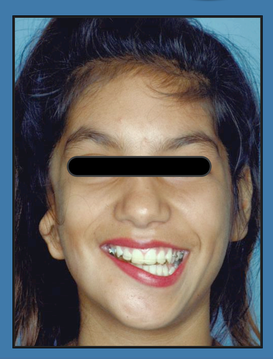


Syndrome:
We use a binary system for the presence of an associated syndrome. Common syndromes we have encountered in our series are Goldenhar Syndrome (4%), Treacher Collins (2%), Branchio-oto-renal (1%), Chromosome 13 deletion (1%) and CHARGE (0.3%)

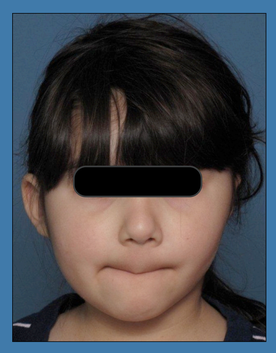




Right $H_{1/6}E_3A_9R_1M_1A_1P_1S_1$. 4 year-old male with isolated unilateral atresia/microtia. Audiogram showed normal bone scores with a maximal conductive hearing loss. CT scan was graded as 9 on Jahrsdoerfer scale. He eventually underwent atresia and microtia repair



Right $H_{1/6}E_3A_4R_1M_1A_1P_3S_1$. 15 year-old female with right atresia/ microtia and facial nerve paresis of the lower branches. CT revealed the patient was not a candidate for atresiaplasty and underwent microtia repair and selective chemodenervation of the contralateral facial nerve. The patient was offered a bone anchored hearing device.



Left $H_{1/6}E_3A_5R_1M_1A_2P_1S_1$. 3 y/o female with left sided CAA. She was deemed a borderline candidate for atresiaplasty. Notice the soft tissue asymmetry with a symmetric chin suggesting normal mandibular development.

DISCUSSION

Best outcomes are achieved when a multidisciplinary team creates a comprehensive plan tailored to the patient. The creation of such teams poses communication challenges. This new grading system has served two interrelated purposes for our team: standardization of evaluation and communication enhancement.

The current system allows providers to understand what a colleague from a different specialty needs to address. For example a Plastic Surgeon can quickly realize that the patient in front of him with a Jahrsdoerfer score of 4 is not a good candidate for atresiaplasty and other means of hearing will be recommended. Armed with this knowledge, rib or medpor microtia reconstruction planning can go forward. It has also been our experience that this system facilitates education of team members, trainees and other patient providers.

Although the system allows for the majority of the data to be shared in the subject line of an email, it is not intended to replace provider-to-provider communication and planning.

CONCLUSIONS

Patients with congenital aural atresia and microtia often require the intervention of audiology, otology, plastic surgery, and craniofacial surgery to achieve optimal function and aesthetic reconstruction. Accurate communication between these different providers is essential for coordination of care. We propose a new classification and communication method that efficiently describes the physical and radiologic findings in patients with congenital aural atresia and microtia to improve communication and patient care amongst healthcare providers.