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HEAR MAPS a 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.

Methods: Retrospective chart review with descriptive analysis of findings arising from the evaluation of patients with congenital atresia and microtia between January 2008 and January 2012 at a multidisciplinary tertiary referral center.

Results: We developed a 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 consecutive 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 esthetic 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.

1. Introduction

The development of the pinna is tightly connected to the development of the external auditory canal and the middle ear by complex interactions between ectoderm, mesoderm and endoderm originating from the first and second branchial apparatus [1]. Congenital aural atresia (CAA) and microtia occur in approximately 1:10,000–20,000 births as a result of the aberrant development of these structures. As a consequence, there is hypoplasia or aplasia of the external ear (pinna and external auditory canal) and the middle ear [2]. The inner ear's distinct origin from the otocyst as a distinct invagination of the ectoderm of the first branchial groove makes it less likely to be involved in cases of CAA [1,3]. CAA is also associated with congenital anomalies of the heart, lungs and kidneys. Additionally, several syndromes involving the head and neck are commonly associated with CAA, including Goldenhar's, Treacher Collin's, and Branchio-oto-renal syndrome amongst others.

Patients with CAA are inherently complex and require the services of multiple medical specialties. 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. For example, the otology literature has multiple classification methods for atresia including Altmann's, Schuknecht's, De la Cruz's and Chiossone's, none of which address the pinna [2]. Perhaps the most widely used 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 [4]. Similarly the plastic surgery literature presents

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multiple classification methods (Marx [5], 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 [6].

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. Communication is essential for coordination of care. We soon 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 that resulted in the system described in this report. With this manuscript we describe the classification to the community and share our clinical findings.

2. Materials and methods

Institutional Review Board exemption status was obtained for this project from the Western Institutional Review Board (Olympia, Washington). All photographs in this manuscript have written consent by the patients or caregiver on file. Patients were evaluated by the senior authors (JBR, JFR and SAS) between the years of 2008 and 2012. Demographic and medical data were recorded in a password-protected Bento database (FileMaker, Inc., Santa Clara, CA) with each patient encounter. This information was de-identified and exported into Excel (Microsoft Corporation, Redmond, Washington) for retrospective review. Inclusion criteria were patients with CAA evaluated by the senior authors during the previously mentioned time frame. Descriptive analysis was performed with Excel.

3. Results

The patient population consisted of 742 consecutive patients evaluated for CAA and microtia between January 2008 and January 2012 by the senior authors (JBR/JFR/SAS). Table 1 describes the HEAR MAPS classification system. Demographics of the patient population are summarized in Table 2. Clinical findings are described in Table 3.

3.1. Hearing

Hearing assessment was performed and recorded in all patients before any surgical intervention. Following the guidelines from the Committee on Hearing and Equilibrium we record the patient's audiogram by calculating PTA2 (average of thresholds at 500, 1000, 2000 and 3000 Hz) for both bone and air thresholds [7]. We report Bone PTA2 separated by a slash from the Air PTA2. We have found it to be important to record bone and air thresholds for both ears given that in our series 15% of patients had a mild sensorineural hearing loss in the ear with CAA and up to 22% of CAA patients present with inner ear anomalies on CT evaluation which may result in associated hearing loss of either a conductive, sensorineural or mixed nature [3]. We chose not to report PTAs for the contralateral ear for ease of use but will make a note of any contralateral hearing loss for obvious clinical reasons (Table 3).

3.2. Ear (microtia)

We used the modified Marx's 4-point scale for the classification of microtia [5] (Fig. 1).

Table 1
HEAR MAPS classification.

Hear bone/air (PTA2 dB HL)	
Bone PTA2/Air PTA2	
Ear (microtia)	
Grade 1	Normal
Grade 2	Mild malformation
Grade 3	Moderate malformation
Grade 4	Anotia
Atresia Jahrsdoerfer CT scale	
Grade 1–10	
Remnant earlobe	
Grade 1	Normal
Grade 2	Mildly reduced
Grade 3	Moderately reduced
Grade 4	Severely reduced/absent
Mandible	
Grade 1	Normal
Grade 2	Mildly reduced
Grade 3	Moderately reduced
Grade 4	Severely reduced/absent
Asymmetry soft tissue	
Grade 1	Normal
Grade 2	Mildly reduced
Grade 3	Moderately reduced
Grade 4	Severely reduced/absent
Paresis of the facial nerve House–Brackmann scale	
Grade 1–6	
Syndrome	
Grade 1	None
Grade 2	Yes

3.3. Atresia score

We used Jahrsdoerfer 10-point grading scale based on CT findings to determine atresiaplasty candidacy [4].

3.4. Remnant earlobe

We graded the Remnant earlobe as normal, mildly, moderately or severely reduced. In cases in which the earlobe is significantly displaced the earlobe is still graded based on its size and a separate note is made to this effect. The size of the remnant lobe has important implications for the microtia reconstruction planning.

3.5. Mandible asymmetry

We evaluated the mandible for ipsilateral mandibular hypoplasia. We grade the hemimandible as normal, mildly, moderately or severely reduced. This classification is especially useful for our craniofacial surgeon to determine if mandibular distraction procedures are anticipated in the patient.

3.6. Asymmetry soft tissue

We grade the soft tissue asymmetry as normal, mildly, moderately or severely reduced (Fig. 2). We evaluate the facial symmetry to see if there is hemifacial hypoplasia but it is difficult

Table 2
HEAR MAPS.

Demographic characteristics of the patient population	
Age	
Median	5
Range	(1 month-69 years)
Gender	
Male	63%
Female	37%
Side	
Right	332 (53%)
Left	189 (30%)
Bilateral	110 (17%)

Table 3
Clinical findings of the patient population.

Hearing $n=41$[a]		
	Bone	Air
Average PTA2	12.98	63.11
Ear (microtia grade) $n=432$ (no data 310)		
Normal		6 (1%)
Grade 1		17 (4%)
Grade 2		69 (16%)
Grade 3		329 (76%)
Grade 4		11 (3%)
Atresia score $n=498$ (no data 244)		
Jahrsdoerfer score		
1		5 (1%)
2		11 (2%)
3		17 (3%)
4		35 (7%)
5		24 (5%)
6		21 (4%)
7		95 (19%)
8		195 (39%)
9		95 (19%)
Remnant lobe $n=311$ (no data 431)		
Normal		248 (79%)
Mildly reduced		38 (12%)
Moderately reduced		17 (5%)
Severely reduced		8 (3%)
Mandible $n=320$ (no data 422)		
Normal		246 (76%)
Mildly reduced		53 (16%)
Moderately reduced		16 (5%)
Severely reduced		5 (2%)
Asymmetry soft tissue $n=306$ (no data 436)		
Normal		242 (79%)
Mildly reduced		49 (16%)
Moderately reduced		12 (4%)
Severely reduced		4 (1%)
Pre-operative paresis $n=209$ (no data 533)		
No		200 (96%)
Yes		9 (4%)
Syndrome $n=355$ (no data 387)		
No		326 (92%)
Yes		29 (8%)

[a] Of note hearing was the last category added therefore we present the average PTA2 for the patients operated during the last year.

sometimes to determine if the asymmetry is solely due to soft tissue, bone or a combination. This distinction is critical because soft tissue asymmetries can be corrected with fillers such as autogeneous abdominal fat.

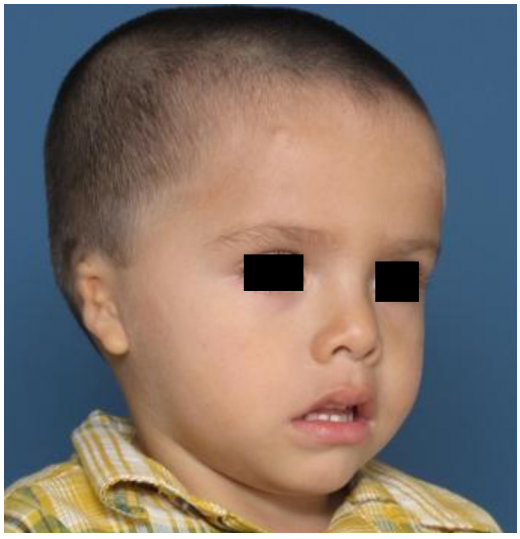

Fig. 1. $H_{1/6}E_3A_9R_1M_1A_1P_1S_1$. 4 year-old male with isolated unilateral atresia/microtia. Audiogram showed normal bone scores with a maximal conductive hearing loss. CT scan was graded as 9 on Jahrsdoerfer scale.

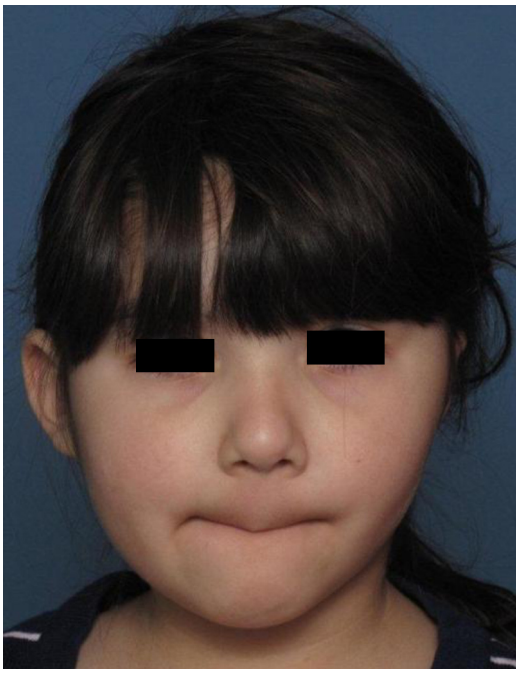

Fig. 2. $H_{1/6}E_3A_5R_1M_1A_2P_1S_1$. 3 year-old female with left sided CAA. She was deemed a borderline candidate for atresiaplasty. Notice the soft tissue asymmetry with a symmetric chin.

3.7. Paresis facial nerve

We used the House–Brackmann facial nerve grading scale. Facial nerve paresis should alert the otologist to abnormal anatomy of the facial nerve [8] (Fig. 3).

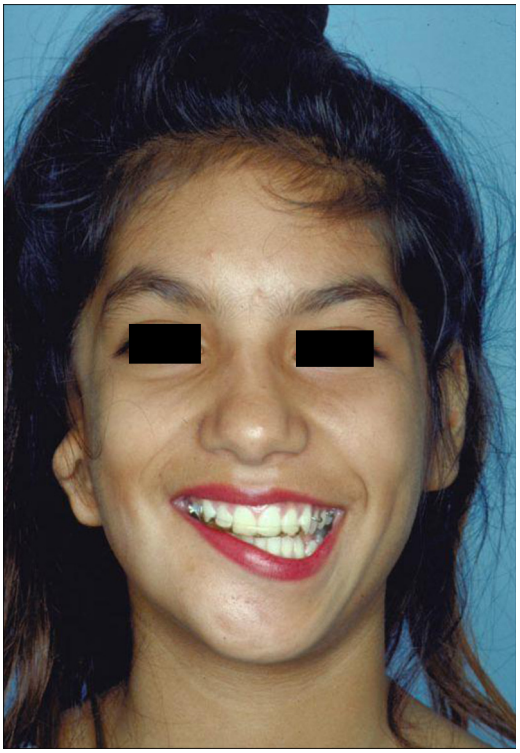

Fig. 3. $H_{1/6}E_3A_4R_1M_1A_1P_3S_1$. 15 year-old female with right CAA and facial nerve paresis of the lower branches. CT revealed the patient was not a candidate for atresiaplasty and underwent microtia repair and selective chemodenervation of the contralateral facial nerve. The patient was offered a bone anchored hearing device.

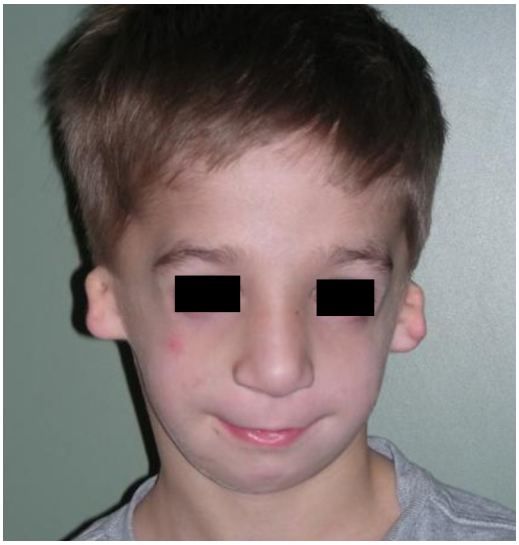

Fig. 4. (Right) $H_{3/6}E_3A_4R_2M_2A_2P_1S_2$. (Left) $H_{2/6}E_3A_5R_2M_3A_3P_1S_2$. 8 year-old male with history of Treacher Collins.

3.8. Syndrome

We used a binary system for the presence of an associate 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%) (Fig. 4).

4. Discussion

Best outcomes are achieved when a multidisciplinary team creates a comprehensive plan tailored to the patient with CAA with or without other associated craniofacial abnormalities. The creation of such teams poses communication challenges given the significant subspecialization and demands to discuss individual patients' needs in busy schedules. This new grading system has served two interrelated purposes for our team: standardization of evaluation and communication enhancement. By standardizing our evaluation, we assure that all patients have a complete evaluation regardless of which specialist meets them first. Additionally discussions amongst providers have proven to be more focused and time efficient. The system has increased its value over time with successive refinements.

Individual specialties have grading systems that go beyond our current system. For example, jaw abnormalities can be graded with much more detail frequently needed for surgical decision-making. The current system, however, allows providers to understand what a colleague from another specialty needs to address in short format. As an example, a plastic surgeon can quickly determine that a Jahrsdoerfer grade of 4 on a CT scan is not a good candidate for canalplasty and other means of hearing will be recommended. Armed with that knowledge, rib graft or medpor reconstruction planning can go forward. Following the same logic knowledge about soft tissue asymmetry or absent ear lobe can help the plastic surgeon in surgical planning. The presence of osseous hypoplasia lets the craniofacial surgeon know that his/her involvement is necessary in this case.

It has also been our experience that the current system facilitates education of team members, trainees and other patient providers who may not be as familiar with this rare syndrome and its treatment. To date, this grading system has been used mainly with those physicians providing surgical care to the patient but can easily extend to Pediatricians, Speech and Language Pathologists, Orthodontists, Audiologists, etc. on the treatment team.

There are several limitations in this project. Although the data was prospectively collected not all data points were available for all patients since some data might be unavailable at the time of consultation (i.e. CT Scan in a 6 month old) or some items of the acronym were added on later. We acknowledge that any attempt to summarize a patient's medical history in an acronym will be an oversimplification and will necessarily leave out essential information. However, acronyms have successfully been used in medicine throughout time. Obstetricians, for example, have long used acronyms to describe a woman's reproductive health.

This system is not intended to replace direct provider-to-provider communication but to allow concise exchange of information with other providers. Based on our extensive experience treating patients with congenital aural atresia and microtia we think that other providers might find this system useful.

5. Conclusion

Patients with CAA often require the intervention of audiology, otology, plastic surgery, and craniofacial surgery to achieve optimal function and esthetic reconstruction. Accurate communication between these different providers is essential for coordination of care. We propose a 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.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at http://dx.doi.org/10.1016/j.ijporl.2013.07.002.

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