ISSN: 2340-3438

Edita: Sociedad Gallega de Otorrinolaringología.

Periodicidad: continuada.

Web: www: sgorl.org/revista

Correo electrónico:

actaorlgallega@gmail.com

Acta Otorrinolaringológica Gallega

Artículo Original

Type III Microtia: A retrospective study of clinical and radiological findings

Microtia tipo III: estudo retrospetivo dos achados clínicos e imagiológicos

Mariana Donato[1], Vera Silva[2], Filipe Correia[1], Nelson Gilberto[1], Ricardo Santos[1], Assunção O´Neill[1], Pedro Escada[1]

[1]ENT Department, Hospital de Egas Moniz, Centro Hospitalar Lisboa Ocidental, Lisbon, Portugal

[2]Neurorradiology Department, Hospital de Egas Moniz, Centro Hospitalar Lisboa Ocidental , Lisbon, Portugal

Recibido: 14/8/2018 Aceptado: 9/10/2018

Abstract

Objectives: To assess the multiple ear malfomartion that can be found in a patient with type III microtia and to present the demographic findings and the audiological evaluation of a group of patients.

Methods: Retrospective study, including all patients with type III microtia referred to the Childhood Hearing Loss Unity of a tertiary hospital during a period of 8 years. Patients' clinical history, audiologic evaluation and computed tomography (CT) results were collected from individual case files. CT findings were distributed into 5 groups of abnormalities: external ear, middle ear, mastoid, inner ear and fallopian canal.

Results: Ten patients with type III microtia were included. The right ear was the most affected. Six patients had associated genetic syndromes. All patients with only one side affected had a conductive hearing loss on the affected ear with normal hearing in the contralateral ear. Regarding the groups of CT findings: the external auditory canal was atresic in nine patients and one patient had a membranous stenosis; in

Correspondencia: Mariana de Miranda Lemos Donato
Hospital de Egas Moniz, Centro Hospitalar Lisboa Ocidental, Portugal
Correo electrónico: marianamldonato@gmail.com

the middle ear, there were malformations of the tympanic membrane, ossicular chain and the tympanic cavity width. The most common ossicular chain anomaly was a malleus-incus fusion. Abnormalities of mastoid size and pneumatization were also observed. The Fallopian canal was abnormal in six patients. No anomalies of tegmen tympani, oval and round window or inner ear were found in our study.

Conclusion: This study highlights the anatomic variations of the temporal bone that can be associated with type III microtia. It is a valuable addition to the available knowledge of this rare condition, since it allows us to predict which changes are most expected to be found in these patients and permit an easier reading of the imaging exam.

Keywords: Congenital Microtia; Ear Diseases/diagnosis; Ear Diseases/diagnostic imaging

Resumo

Objetivo: Avaliar as múltiplas malformações do ouvido possíveis nos doentes com microtia tipo III e apresentar os achados demográficos e a avaliação audiométrica de uma amostra destes doentes.

Métodos: Estudo retrospetivo, incluindo todos os doentes com microtia tipo III referenciados para o departamento de Surdez Infantil dum hospital terciário durante 8 anos. A avaliação clínica, avaliação audiológica e os resultados da tomografia computorizada (TC) foram obtidos através de consulta dos processos clínicos. Os achados da TC foram distibuídos em 5 grupos de anomalias: ouvido externo, ouvido médio, mastoide, ouvido interno e canal de Falópio.

Resultados: Foram incluídos 10 doentes com microtia tipo III. O ouvido direito foi o mais afetado. Seis doentes tinham síndromes genéticas associadas. Todos os doentes com apenas um lado afetado apresentavam hipoacusia de condução ipsilateral com audição normal no ouvido contralateral. Relativamente aos grupos de achados de TC: o canal auditivo externo era atrésico em nove doentes; um doente apresentava estenose membranosa; no ouvido médio, registaram-se malformações da membrana timpânica, da cadeia ossicular e da largura da cavidade timpânica. A malformação da cadeia ossicular mais frequente foi a fusão incudo-maleolar. Na mastoide, observam-se alterações do tamanho e da pneumatização. O canal de falópio estava alterado em seis doentes. Não foram observadas malformações do tégmen timpânico, das janelas oval e redonda ou do ouvido interno.

Conclusão: Este estudo realça as alterações anatómicas do osso temporal que podem estar associadas a microtia tipo III. É uma adição importante ao conhecimento atual, uma vez que permite predizer quais as alterações que são expectáveis nestes doentes, assim como possibilita uma leitura mais fácil do exame de imagem.

Palavras chave: Micrótia congénita; Doenças do ouvido/diagnóstico; Doenças do ouvido/ diangósitco imagiológico

Introduction

Ear malformations affect approximately 1/3800 live births and correspond to 50% of the malformations of the ear, nose and throat area.[1] They can affect the outer, middle and inner ear, frequently in combination.[1]

The term microtia describes a spectrum of congenital anomalies of the auricle that range from mild structural abnormalities to complete absence (anotia). The incidence of microtia in Europe and North-America is 0.02% of the newborns.[2-8]

The etiology of microtia is not completely known.[1, 4-7] A number of causes have been implicated, including teratogens, vascular insults and genetic factors.

There is no universal classification used for microtia, which makes it difficult to standardize the clinical findings.[1, 4-6] The Weerda classification grades microtia as type I (most structures of a normal auricle are recognizable); type II (some structures of a normal auricle are recognizable); type III (none of the structures of a normal auricle are recognizable).[3, 9]

Much has been published regarding surgical treatments of microtia and their outcome, but there is a lack of information regarding the middle and inner ear malformations that can be found in this patients.[4]

The main objective of this study is to assess the multiple ear malformation (external auditory canal, middle ear, mastoid, inner ear and fallopian canal) that can be found in a patient with type III microtia. We also aim to present the demographic findings and the audiological evaluation of a group of those patients.

Methods

Patient selection

A retrospective study was performed including all patients with type III microtia referred to the Childhood Hearing Loss Unity of the Otorhinolaryngology Department of Centro Hospitalar Lisboa Ocidental (tertiary referral hospital), between January of 2007 and December of 2015.

To classify microtia, the Weerda classification was used.

In order to obtain more consistent data, children with type I and type II microtia were excluded. Patients with incomplete information in the individual case files were also excluded.

Patients' clinical history, audiologic evaluation and computed tomography (CT) results were collected from individual case files. The information collected is listed in table 1. Experienced otorhinolaryngologists reviewed the data.

The work was carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki).

Image analysis

The available CT studies were examined by a neuroradiologist. Findings were systematically evaluated in accordance with a predefined sheet and were distributed in 5 main anatomical sites (table 2). All listed structures were evaluated for both ears.

Table 1: Information collected from the individual case file

Information collected from the individual case files
1. Gender
2. Age
3. Side of affected ear
4. Associated genetic syndromes
5. Audiological tests (pure tone audiogram, speech audiogram and brain evoked response audiometry)
6. Computed tomography (CT) results

Table 2: Predefined sheet used to evaluate the computed tomography (CT) findings

CT results
1. External ear: pinna and external auditory canal
2. Middle ear: tympanic cavity (size and morphology), tegmen tympani, tegmen mastoideum, middle ear ossicles (presence and integrity), oval and round window
3. Mastoid (size and pneumatization)
4. Inner ear
5. Fallopian canal

Results

Forty-nine cases were retrieved, 39 of those were excluded due to non-compliance with the aforementioned criteria. The study group consisted in 10 patients with type III microtia with the main characteristics summarized in table 3.

Demographic findings

Six patients were female and four were males, the average age at the time of the first appointment was 4 years old. Only one patient had bilateral microtia. In the remaining 9 patients with only one side affected, the right ear was the most affected (6 cases).

Associated genetic syndromes

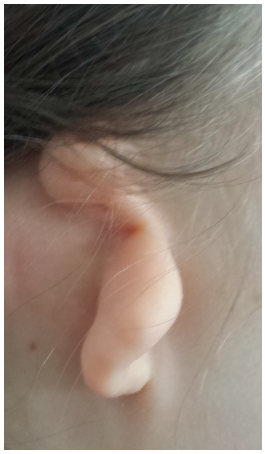
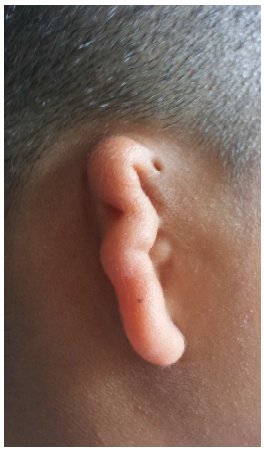
Microtia was an isolated finding in four patients while it was associated with genetic syndromes in six: five patients had oculo-auriculo-vertebral syndrome (OAVS) and one patient had Treacher-Collins syndrome.

Table 3. The main characteristics of the population of patients with type III microtia studied (OAVS: oculo-auriculo-vertebral syndrome; BER: brain evoked response audiogram)

	Age	Gender	Ear	Associated genetic syndromes	Audiological exams			CT findings			
					Type of exam	Result	Speech audiometry	Outer ear	Middle ear	Mastoid	Fallopian canal
Patient 1	1	Female	Right	OAVS	BER	RE: Absent V wave at 60dB;	--	Abnormal pinna; external auditory canal atresia	Malleus-incus fusion; absent incudostapedial joint; incus adherent to the lateral tympanic cavity wall	Anterior displacement of the mastoid	Anterior displacement of the mastoidal segment
Patient 2	4	Female	Right	OAVS	Audiogram	RE: bone conduction threshold of 10 dB with ABG 60 dB; LE: PTA 0 dB	RE: S curve with an threshold of intelligibility of 90 dB	Abnormal pinna; external auditory canal atresia	Hypopnematized tympanic cavity; malleus-incus fusion	Hypopnematized mastoid cavity	Antero-lateral displacement of the mastoidal segment
Patient 3	1	Female	Right	Isolated	BER	RE: Absent V wave at 60dB; LE V wave at 10 dB	--	Abnormal pinna; external auditory canal atresia	Malleus-incus fusion; dysmorphic incus	--	--
Patient 4	6	Male	Left	OAVS	Audiogram	RE: PTA 5 dB; LE: Bone conduction threshold of 5 dB ABG 55 dB	RE: S curve with an threshold of intelligibility of 70 dB	Abnormal pinna; external auditory canal atresia	Hypopnematized tympanic cavity; malleus-incus fusion; absent incudostapedial joint	Hypopnematized mastoid cavity	Anterior displacement of the mastoidal segment
Patient 5	1	Male	Left	Isolated	BER	RE: V wave at 10 dB; LE Absent V wave at 60 dB	--	Abnormal pinna; external auditory canal atresia	Malleus-incus fusion	--	--
Patient 6	8	Female	Left	Isolated	Audiogram	RE: PTA 0 dB; LE: Bone conduction threshold of 5 dB ABG 60 dB	LE: S curve with an threshold of intelligibility of 70 dB	Abnormal pinna; external auditory canal atresia	Dysmorphic incus	--	--
Patient 7	4	Male	Bilateral	Treacher-Collins	Audiogram	RE: Bone conduction threshold of 10 dB ABG 60 dB; LE: Bone conduction threshold of 5 dB ABG 70 dB	RE: S curve with an threshold of intelligibility of 55 dB; LE: S curve with an threshold of 75 dB	Abnormal pinna; membranous stenosis of the EAC	Hypopnematized tympanic cavity; malleus-incus fusion; absent incudostapedial joint; malleus and incus adherent to the lateral tympanic cavity wall	Hypopnematized mastoid cavity	Anterior displacement of the mastoidal segment
Patient 8	1	Female	Right	OAVS	BER	RE: Absent V wave at 50dB; LE: V wave at 10 dB	--	Abnormal pinna; external auditory canal atresia	Hypopnematized tympanic cavity; malleus-incus fusion; incus adherent to the posterior tympanic cavity wall	Hypopnematized mastoid cavity	Displacement of the tympanic and mastoidal segment
Patient 9	9	Female	Right	Isolated	Audiogram	RE: Bone conduction threshold of 5 dB ABG 65 dB; LE: PTA 0 dB	RE: S curve with an threshold of intelligibility of 80 dB	Abnormal pinna; external auditory canal atresia	Dysmorphic incus	--	--
Patient 10	1	Male	Right	OAVS	BER	RE: V wave at 10 dB; LE Absent V wave at 70 dB	--	Abnormal pinna; external auditory canal atresia	Hypopnematized tympanic cavity; malleus-incus fusion; absent stapes supra-structure	Hypopnematized mastoid cavity	Displacement of the tympanic and mastoidal segment

Physical examination

All selected patients presented with severe deformity of the pinna precluding the recognition of any of the normal structures (figure 1). None of the patients had an identifiable external auditory canal. Patients with OAVS also presented a facial asymmetry with hypoplasia of the ipsilateral jaw. The patient with Treacher-Collins syndrome had bilateral microtia, downward-slanting eyes, notched lower eyelids and micrognathia.

A  B 

Figures 1a and 1b: Ears with type III microtia.

Audiological evaluation

All patients with only one side affected had a moderate-to-severe conductive hearing loss on the affected ear with normal hearing in the contralateral ear. The patient with bilateral microtia (patient 4) had symmetrical moderate-to-severe conductive hearing loss.

Computed tomography

External ear: All patients had an abnormal pinna consistent with type III microtia (figures 2 and 3). The external auditory canal was atresic in nine patients (figure 3). One patient had a membranous stenosis (figure 4).

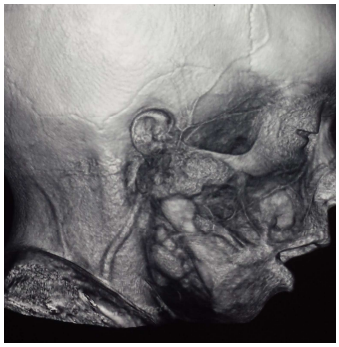
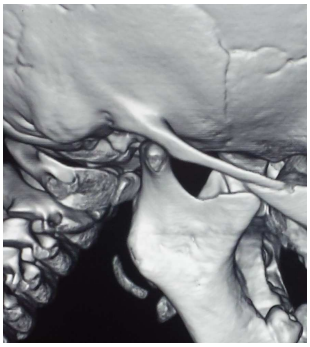
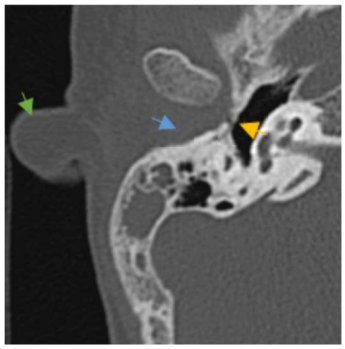
A  B 

Figure 2a and 2b: 3D TC reconstruction showing an abnormal pinna (a) and the absence of the external auditory canal (b).

Figure 3: (axial CT image) EAC with membranous (blue arrow) and osseous atresia (yellow arrow) with an abnormal pinna (green arrow).



Middle ear:

Tympanic membrane: a bony plate was the most common finding at the level of tympanic membrane (nine patients) (figure 5).

Tympanic cavity width: the tympanic cavity was hypopneumatized (figure 6) in five patients (four of the five patients with OAVS and in the patient with Treacher-Collins syndrome). The patients with microtia as an isolated finding had no anomalies on the tympanic cavity width.

Ossicular chain: a malleus-incus fusion was the most common ossicle anomaly, present in eight patients (figures 7 and 8). The incudostapedial joint was absent in three patients (figure 8). The ossicles were adherent to the tympanic cavity walls in three patients (figure 9). There was a dysmorphic incus in three patients (figures 7 and 8) and an agenesis of the stapes supra-structure was present in one patient (figure 10).

No anomalies of tegmen tympani, tegmen mastoideum, oval and round window were found.

Mastoid: the mastoid was hypopneumatized (figure 6) in five patients (four of the five patients with OAVS and in the patient with Treacher-Collins syndrome). The other patient with OAVS had an anterior displacement of the mastoid cavity. As with the tympanic cavity, in isolated microtia there were no anomalies on the mastoid width.

Inner ear: None of the patients had inner ear anomalies.

Fallopian canal: six patients displayed anomalies: all had displacement of the mastoid segment (figure 11) and two of them had a caudal displacement of the tympanic segment.

Figure 4: (axial CT image) EAC with a membranous stenosis (blue arrow) and a soft tissue plug in place of the tympanic membrane (yellow arrow).

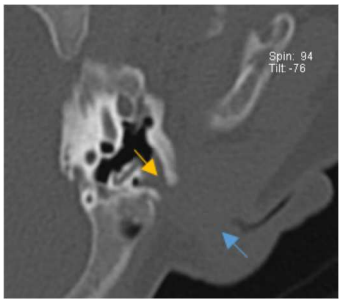


Figure 5: coronal CT image showing a bony plate at the level of the tympanic membrane (blue arrow).

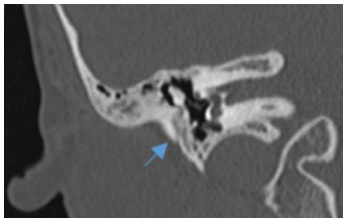
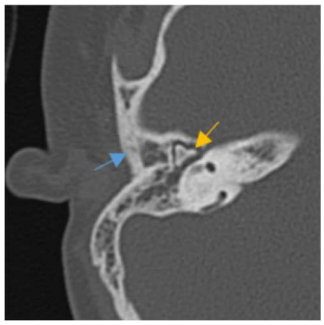
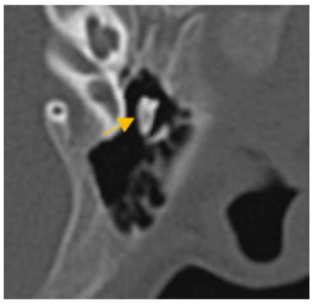


Figure 6: (axial CT image) mastoid (blue arrow) and tympanic cavity hypopneumatized (yellow arrow).



A



B

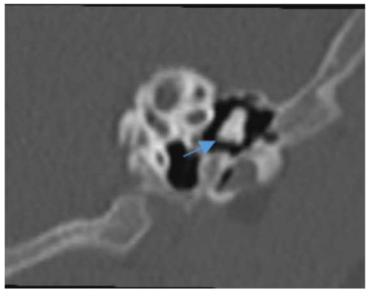
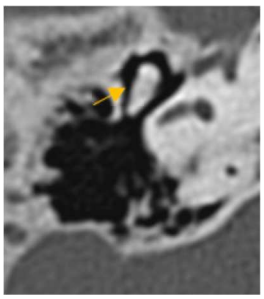


Figure 7a (axial CT scan) and 7b (coronal CT scan) partial malleus-incus fusion (blue arrow) and dysmorphic incus (particularly the lenticular and long process of incus) (yellow arrow).

A



B

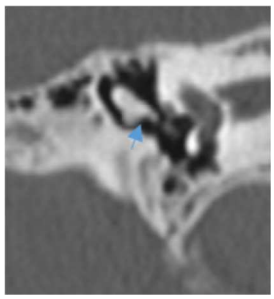
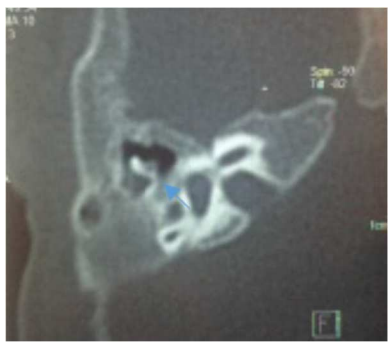


Figure 8 a) and b) Axial and coronal CT scan: malleus-incus fusion (yellow arrow), dysmorphic incus and absence of the incudostapedial joint (blue arrow).

Figure 9 (axial CT scan) incus is adherent to the posterior tympanic wall (blue arrow).



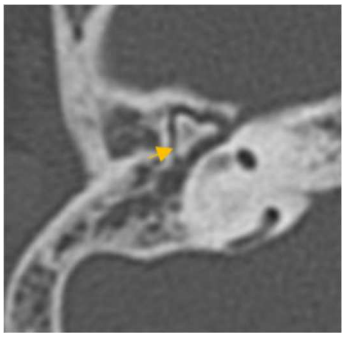
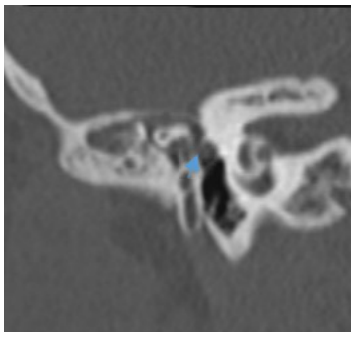
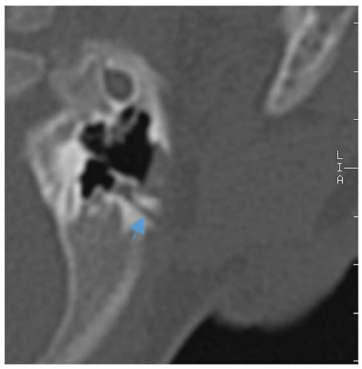
A  B 

Figure 10a (axial CT) and figure 10b 10b (coronal CT): displasic ossicular chain (yellow arrow) with and agenesis of the stapes supra-structure (blue arrow). The mastoid and tympanic cavity are hypopneumatized.

Figure 11: Axial CT images: mastoidal segment of the fallopian canal is more anterior and external (blue arrow).



Discussion

Microtia is associated not only with aesthetic problems but also with psychosocial trauma for the child and for the family.[2, 3] If the clinical presentation is bilateral, there is greater risk of significant language delays and of development attention deficit disorders, which can be worsened if the patient has some lack of visual accuracy because it will be very difficult to use glasses.[2-4]

Demographic findings

A female preponderance was found, which is contrary to most studies described in the literature for both European and North Americans populations.[3, 5, 6] It is difficult to draw conclusions about these findings, since our study focused on only ten patients. Microtia is often described as a unilaterally condition (79–93% of cases) affecting most frequently the right ear, which was also observed in the presented cohort.[3-6, 10]

Associated genetic syndromes

Although the majority of cases are isolated and nonsyndromic, microtia can be associated with other anomalies or with an identifiable syndrome pattern. The most common syndromes associated with microtia are OAVS, Treacher-Collins and Townes–Brock syndromes.[1-5] In our study, the majority of patients had an identifiable syndrome, probably because the cases were retrieved from a tertiary reference center for childhood deafness.

The five patients with OAVS presented a hemifacial microsomia characterized by a unilateral microtia and hypoplastic jaw. This disease is a rare congenital condition, where the head structures derived from the first and the second pharyngeal arches are incompletely developed in one or both sides.[11] It mostly results in jaw and ear abnormalities with external and medium ear involvement. The phenotype may vary from isolated facial asymmetry to eye or spine involvement in the most severe cases.[1, 2, 10, 11] The patient with Treacher-Collins syndrome had bilateral microtia, downward-slanting eyes, notched lower eyelids and micrognathia. This rare autosomal dominant syndrome is associated, in the large majority of the patients, with a mutation in the TCOF1 gene. This gene encodes a protein that plays an important role in the early embryonic development of bone and soft tissues of the face, leading to symmetrical morphological abnormalities.[2, 12-14] Patients affected by this syndrome have no associated developmental delay or neurologic disease, so they often face social challenges throughout their life because of the physical appearance.[13, 14]

Audiological results

The hearing screening in a newborn with microtia is of vital importance because some patients may have hearing loss in the nonmicrotic ear. If the nonmicrotic ear passes in this screening, additional testing can be delayed until the age of 6 to 7 months.[3] In our study, objective testing (brain evoked response audiometry) was used for children younger than 3 years and pure tone audiometry and speech audiometry were performed in older children. Similar to the literature, all patients presented a moderate-to-severe conductive hearing loss on the affected side. [3, 9]

Computed tomography findings

High-resolution CT of the temporal bone is the standard imaging exam to study the ear in cases of microtia.[15] It offers excellent visualization of the osseous anatomy of the temporal bone, being more suitable for displaying the changes of the external auditory canal, middle ear (including the ossicles and the mastoid cavity), inner ear, and fallopian canal.[9, 16, 17] The short time of image acquisition and the fact that this exam does not require contrast are some of the advantages of the CT scan that are especially important when studying a pediatric population, albeit the use of ionizing radiation.[17]

Temporal bone anatomy is complex and the presence of multiple anomalies in the same patient demands some insight into the embryological development of the temporal bone. Most congenital temporal bone anomalies are caused by either premature arrest of its normal development or complete failure of formation.[16]

The outer and middle ear have a different embryological origin from the inner ear.[1] The outer and middle ear are derived from the first and second branchial arches, first branchial cleft and first pharyngeal pouch.[16, 18] The inner ear is derived from the otic capsule.[16]

The CT findings were distributed into 5 groups.

The first group includes external ear abnormalities. All patients with type III microtia had an atresia of the external auditory canal (a bony atresia in nine patients and a membranous atresia in one patient). This is in agreement with the study by Qin *et al*. about the anatomic variants on the CT scan in congenital aural atresia and stenosis, where 86% of ears with third-degree microtia had an atresia.[19]

The second group includes middle ear anomalies and was subdivided in: malformations of tympanic mem-

brane; tympanic cavity width; and ossicular chain.

Patients with a bony atresia had a bony plate at the level of tympanic membrane whereas the patient with the membranous atresia had a soft-tissue plug at this position, findings that are in agreement with previous studies.[19]

The tympanic cavity was hypopneumatized in all patients with OAVS and in the patient with Treacher-Collins syndrome, also in accordance with the literature.[12, 20]

There were several anomalies in the ossicular chain, most of them involving the malleus and incus. The ossicles are derived from the first and second branchial arches. The most accepted theory is that the upper ossicular chain (malleus head, incus body and incus short process) arises from the first branchial arch, while the lower ossicular chain (malleus manubrium, incus long process and lenticular process) comes from the second arch.[9, 16] There is some controversy about the origin of the stapes. At this moment it is thought that the stapes has a dual origin, with its supra-structure derived from the second arch crest and the stapedial footplate being composed of cells of both neural crest and mesoderm.[21] The most common anomaly reported in the literature is the malleus-incus fusion,[19] which was also the most common anomaly found in this study.

Similar to the tympanic cavity, the mastoid was hypopneumatized in four of the five patients with OAVS and in the patient with Treacher-Collins syndrome, in accordance with the literature.[12, 20] The other patient with OAVS had an anterior displacement of the mastoid. The patients with microtia as an isolated finding had no anomalies of the mastoid cavity width.

Malposition of the fallopian canal has been reported in almost all children with microtia, especially in severe grades of microtia, since it depends on the development of the middle ear, mastoid process and tympanic ring.[22, 23] In the literature, the most common finding is an anterior displacement of the mastoid segment of the canal.[16, 18] In our study, the fallopian canal was abnormal in six patients. All patients with an abnormal fallopian canal had an anterior displacement of the mastoid segment, but there were also two patients with tympanic segment anomalies.

No anomalies of the inner ear were found in this study. These results are consistent with the earlier embryological development of the inner versus outer and middle ear structures.[16]

Conclusion

This study highlights the anatomic variations of the temporal bone that can be associated with type III microtia. It is a valuable addition to the available knowledge of this rare condition, since it allows an anticipation of the most expected changes in these patients and an easier interpretation of the imaging exam.

Conflict of Interest Statement: None.

Funding Source: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1- Bartel-Friedrich S. Congenital Auricular Malformations: Description of Anomalies and Syndromes. Facial Plast Surg. 2015; 31: 567-80

2- Gendron C, Schwentker A, van Aalst JA. Genetic Advances in the Understanding of Microtia. J Pediatr Genet. 2016; 5: 189-97

3- Kelley PE, Scholes MA. Microtia and congenital aural atresia. Otolaryngol Clin North Am. 2007; 40: 61-80

4- Luquetti DV, Leoncini E, Mastroiacovo P. Microtia-anotia: a global review of prevalence rates. Birth Defects Res A Clin Mol Teratol. 2011; 91: 813-22

5- Luquetti DV, Heike CL, Hing AV, Cunningham ML, Cox TC. Microtia: epidemiology and genetics. Am J Med Genet A. 2012; 158A: 124-39

6- van Nunen DP, Kolodzynski MN, van den Boogaard MJ, Kon M, Breugem CC. Microtia in the Netherlands: clinical characteristics and associated anomalies. Int J Pediatr Otorhinolaryngol. 2014; 78: 954-9

7- Lei L, Zhenzhong L, Lin L, Bo P. Uncovering the pathogenesis of microtia using bioinformatics approach. Int J Pediatr Otorhinolaryngol. 2017;99: 30-35

8- Wroblewska-Seniuk K, Dabrowski P, Greczka G et al. Sensorineural and conductive hearing loss in infants diagnosed in the program of universal newborn hearing screening. Int J Pediatr Otorhinolaryngol. 2018; 150: 181-186

9- Bartel-Friedrich S, Wulke C. Classification and diagnosis of ear malformations. GMS Curr Top Otorhinolaryngol Head Neck Surg. 2007; 6: Doc05

10- Tasse C, Böhringer S, Fischer S et al. Oculo-auriculo-vertebral spectrum (OAVS): clinical evaluation and severity scoring of 53 patients and proposal for a new classification. Eur J Med Genet. 2005; 48: 397-411

11- Manara R, Brotto D, Ghiselli S et al. Cranial nerve abnormalities in oculo-auriculo-vertebral spectrum. AJNR Am J Neuroradiol. 2015; 36: 1375-80

12- Rosa F, Coutinho MB, Ferreira JP. Ear malformations, hearing loss and hearing rehabilitation in children with Treacher Collins syndrome. Acta Otorrinolaringol Esp. 2016; 67: 142-7

13- Vincent M, Geneviève D, Ostertag A, S. Marlin, D. Lacombe, D. Martin-Coignard et al. Treacher Collins syndrome: a clinical and molecular study based on a large series of patients. Genet Med. 2016; 18: 49-56

14- Chang CC, Steinbacher DM. Treacher collins syndrome. Semin Plast Surg. 2012; 26: 83-90

15- Dedhia K, Yellon RF, Branstetter BF, Egloff AM. Anatomic variants on computed tomography in congenital aural atresia. Otolaryngol Head Neck Surg. 2012; 147: 323-328

16- Tekes A, Ishman SL, Baugher KM et al. Does microtia predict severity of temporal bone CT abnormalities in children with persistent conductive hearing loss? J Neuroradiol. 2013; 40: 192-197

17- DeMarcantonio M, Choo DI. Radiographic Evaluation of Children with Hearing Loss. Otolaryngol Clin North Am. 2015; 48: 913-932

18- Jacob R, Gupta S, Isaacson B et al. High-resolution CT findings in children with a normal pinna or grade I microtia and unilateral mild stenosis of the external auditory canal. AJNR Am J Neuroradiol. 2015; 36: 176-180

19- Qin FH, Zhang TY, Dai P, Yang L. Anatomic Variants on Computed Tomography in Congenital Aural Atresia and Stenosis. Clin Exp Otorhinolaryngol. 2015; 8: 320-328

20- Berio A, Garlaschi G, Mangiante G, Piazzi A. Oculo-auriculo-vertebral spectrum with craniosynostosis and osteo-cartilagineous multiple defects: a diffuse chondro-membranous-osteo-dysplasia. Pediatr Med Chir. 2015; 37: 123

21- Thompson H, Ohazama A, Sharpe PT, Tucker AS. The origin of the stapes and relationship to the otic capsule and oval window. Dev Dyn. 2012; 241: 1396-404

22- Takegoshi H, Kaga K, Kikuchi S, Ito K. Facial canal anatomy in patients with microtia: evaluation of the temporal bones with thin-section CT. Radiology. 2002; 225: 852-858
23- Goldsztein H, Roberson JB. Anatomical facial nerve findings in 209 consecutive atresia cases. Otolaryngol Head Neck Surg. 2013; 148: 648-652