Correlation Between Audiological, Clinical and Radiological Findings in Patients with Microtia

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ABSTRACT
Background: Microtia is a congenital abnormality of the auricle. The main problems of patients with microtia are hearing loss and cosmetic abnormalities of the affected ear. Most patients with microtia have normally formed inner ears and have no sensorineural hearing loss. High resolution computed tomography (HRCT) scan and hearing assessment are important for evaluation of patient with microtia.
Objective: The aim of the current work was to evaluate the relation between audiological, clinical and radiological findings in patients with microtia.
Patients and Methods: This is a descriptive cross-sectional study of 30 patients with microtia, who were selected from outpatient clinic of El-Hussein University Hospital, Cairo, Egypt, from August 2018 to September 2019. All subjects were subjected to full history taking, Otological examination, general examination, basic audiological evaluation, and radiological evaluation. Relations were evaluated between hearing level, clinical degree of microtia according to **Marx *et al.*** **[1]** classification and CT scoring as assessed by the CT scoring system of **Jahrsdoerfer *et al.*** **[2]**.
Results: The hearing level correlated significantly with severity of microtia. However, there was no significant association between degree of microtia and CT scoring as assessed by the CT scoring. Also, no significant correlation between hearing level and the total CT scoring.
Conclusion: The principle "The better developed the external ear, the better the hearing level" has been proved in Egyptian cases with microtia. It may not be possible to predict external and middle ear abnormalities based on microtia grades. Therefore, both audiology testing and radiology investigation are two independent evaluations before hearing reconstruction surgery.
Keywords: Microtia, external auditory canal, middle ear anomalies, temporal bone, atresia, hearing, high-resolution computed tomography, pure tone audiometry.

INTRODUCTION

Microtia is a congenital anomaly of the ear that ranges in severity from mild structural abnormalities to complete absence of the ear (anotia) and can occur as an isolated birth defect or as part of a spectrum of anomalies or a syndrome. Microtia is often associated with hearing loss and patients typically require treatment for hearing impairment and surgical ear reconstruction **[3]**.

Abnormalities of the middle ear structure associated with microtia include stapes deformity, absence of oval or round windows, aberrant course of facial nerve, poor pneumatization of the middle ear space, and fusion of malleus and incus **[4]**.

The incidence of inner ear abnormalities associated with microtia is estimated between 10% and 47%. This condition is often accompanied by various temporal bone anomalies **[5]**.

Mostly, microtia is a unilateral anomaly with a right-side dominance **[6]**. The reported prevalence varies among regions, from 0.83 to 17.4 per 10,000 births, and considered to be higher in Hispanics, Asians, Native Americans, and Andeans. The etiology of microtia and the cause of this wide variability in prevalence are poorly understood. Strong evidence supports the role of environmental and genetic causes **[3]**.

It is possible that the anatomical anomalies such as fixation of the stapes or round window closure have an effect on the hearing level. So, high-resolution computed tomography (HRCT) is indispensable for the surgical planning because it provides important anatomic information **[5]**.

More than 80% of patients with microtia have aural atresia resulting in conductive hearing loss, with air conduction hearing typically reduced by 40–65 dB, whereas bone conduction is normal in 90% of the affected ears **[7]**.

There are several grading systems for microtia. In the Marx classification, all of the features of a normal auricle are present in grade I, but the pinna is smaller than normal. In grade II, some anatomical structures are still recognizable. In the most common form, grade III (the peanut-shell type), only a rudiment of soft tissue is present **[8]**.

Jahrsdoerfer *et al.* **[2]** developed a grading system based on temporal bone HRCT and appearance of the auricle for determining good surgical candidacy and accurate prediction of the surgical outcome. Nevertheless, the relationships between preoperative hearing level and abnormalities of the temporal bone have not been elucidated.

The occurrence of microtia is of public health importance in part due to the psychosocial sequelae,

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including the stigma associated with malformations of the ear and the burden of undergoing multiple surgeries [9].

AIM OF THE WORK

The aim of the current work was to evaluate the relation between audiological, clinical and radiological findings in patients with microtia.

SUBJECTS AND METHOD

This descriptive cross-sectional study included a total of 30 patients with microtia (unilateral and bilateral), attending at outpatient clinic of El-Hussein University Hospital, Cairo, Egypt. This study was conducted between August 2018 to September 2019.

Ethical approval and written informed consent :
The study was approved by the Al-Azhar University Academic and Ethical Committee and a written informed consent was obtained from all patients.

Exclusion criteria: Patients with other anomalies associated with systemic syndromes such as Treacher Collins and Goldenharr syndromes and those younger than 3 years of age in whom pure-tone audiometric examination was difficult.

Equipment: Pure Tone Audiometer model MAICO53. Immittance meter model GSI 39 with a probe tone 226Hz. The HRCT images of the temporal bone were obtained using the CT system (General Electric, Milwaukee, USA). Coronal and sagittal reformations of 1-mm thickness were obtained for all patients.

All subjects were submitted to the following:

Full history taking. Otological examination to detect the clinical degree of microtia. The severity of microtia was classified into grades I, II, or III according to Marx's classification [1]. In brief, grade I microtia exhibits only mild deformity, with the auricle being slightly smaller than normal, each part of which can be clearly distinguished. In grade II microtia, the size of the auricle is one half to two thirds of the normal size and its structure is only partially retained. In grade III microtia, the auricle is severely malformed and usually exhibits a peanut shape. Pure tone audiometry was done at frequencies 250, 500, 1000, 2000, 4000 and 8000 Hz. Immitancemetry was performed to in the unaffected ear (in unilateral cases) to exclude otitis media or any middle ear pathology. Acoustic reflex threshold was done on 500, 1000, 2000 and 4000 Hz ipsilaterally and contralaterally. Bone conduction was tested at frequencies 500, 1000, 2000 and 4000 Hz. Speech Audiometry also was done to detect the speech reception threshold (SRT) using Arabic Bisyllabic words for adults **Soliman** [10] and word recognition scores using Arabic phonetically balanced words **Soliman *et al.*** [11]. The HRCT images of the temporal bone have been obtained and used for grading of associated temporal bone anomalies according to the CT scoring system of **Jahrsdoerfer *et al.*** [2].

The anomalies of the temporal bone were graded according to parameters of Jahrsdoerfer CT scoring system: 1) subtotal of parameters related to ossicular development (presence/absence of the stapes, malleus/incus complex, and incudo-stapedial connection [full mark, 4 points]), 2) those related to windows connected to the cochlea (presence/absence of the oval and round windows [full mark, 2 points]), 3) those related to aeration of the middle ear cavity, such as the middle ear space and mastoid (full mark, 2 points), and 5) those related to the facial nerve route (full mark, 1 point) **Jahrsdoerfer *et al.*** [2].

Statistical analysis

The recorded data were analyzed using the statistical package for social sciences, version 20.0 (SPSS Inc, Chicago, Illinois, USA). Quantitative data were expressed as mean± standard deviation (SD). Qualitative data were expressed as frequency and percentage.

The following tests were done:

- Independent-samples t-test of significance was used when comparing between two means.
- Chi-square (x^2) test of significance was used in order to compare proportions between two qualitative parameters.
- The confidence interval was set to 95% and the margin of error accepted was set to 5%. The p-value was considered significant as the following:
- Probability (P-value)
  - P-value <0.05 was considered significant.
  - P-value <0.001 was considered as highly significant.
  - P-value >0.05 was considered insignificant.

RESULTS

This study was conducted on 30 patients with microtia; 17 male (56.7%) and 13 females (43.3%). The mean age of the study group was 7.2 years ranged from 3-19 years. Incidence of bilateral microtia was 6.6%. The degree of microtia was not different between unilateral and bilateral cases.

Table (1): Distribution of Gender in the studied group.

Variables		Studied group (N = 30)	
Gender	Male	17	56.7%
	Female	13	43.3%

Table (2): Distribution of the affected side in the studied group.

Variables		Studied ears (N = 32)	
Side	Right	20	62.5%
	Left	12	37.5%

Table (3): Mean and standard deviation of air-bone gap (ABG), air conduction (AC) & bone conduction (BC) of right ears with clinical degree of microtia in the studied group.

Right ears		Clinical Degree			P-value
		I (n = 2)	II (n = 2)	III (n = 16)	
ABG	Mean	10.00	31.88	45.55	< 0.001
	±SD	0.00	0.88	7.68	
AC	Mean	32.08	45.00	61.51	< 0.001
	±SD	0.59	3.54	6.16	
BC	Mean	21.25	13.13	16.02	0.086
	±SD	1.77	2.65	3.66	

**No statistically significant difference (p-value > 0.05) between mean average of BC of right ears and clinical degrees in the studied group.
**Statistically significant difference (p-value < 0.001) between mean average of ABG and AC of right ears and clinical degrees in the studied group.

Table (4): Mean and standard deviation of ABG, AC & BC of left ears with clinical degree of microtia in the studied group.

Left ears		Clinical Degree		P-value
		II (n = 2)	III (n = 10)	
ABG	Mean	30.63	48.00	< 0.001
	±SD	0.88	2.30	
AC	Mean	46.25	61.83	< 0.001
	±SD	1.77	1.23	
BC	Mean	16.88	14.13	0.075
	±SD	0.88	1.32	

**No statistically significant difference (p-value > 0.05) between mean average of BC of left ears and clinical degrees in the studied group.
**Statistically significant difference (p-value < 0.001) between mean average of ABG and AC of left ears and clinical degrees in the studied group.

Table (5): comparison of SRT (dBHL) & WR% of right ears as regard clinical degree in the studied group.

Right ears SRT, WR		Clinical Degree			P-value
		I (n = 2)	II (n = 2)	III (n = 16)	
SRT (dBHL)	**Mean**	32.5	47.5	63.13	**< 0.001**
	±SD	3.54	3.54	6.02	
WR %	**Mean**	100.0	100.0	99.75	0.894
	±SD	0.0	0.0	1.0	

Statistically significant difference **(p-value < 0.05) between clinical degrees (I, II, III) as regard SRT (dBHL) at right side of the studied group.
No statistically significant difference **(p-value > 0.05) between clinical degrees (I, II, III) as regard WR% at right side of the studied group.

Table (6): comparison of SRT (dBHL)& WR% of left ears as regard clinical degree in the studied group.

Left ears SRT, WR		Clinical Degree		P-value
		II (n = 2)	III (n = 10)	
SRT (dBHL)	**Mean**	50.0	64.0	< 0.001
	±SD	0.0	3.16	
WR%	**Mean**	100.0	100.0	------
	±SD	0.00	0.0	

statistically significant difference **(p-value < 0.001) between clinical degrees (II, III) as regard SRT (dBHL) at Left side of the studied group.**No statistically significant difference **(p-value > 0.05)** between clinical degrees (I, II, III) as regard WR% at left side of the studied group.

Table (7): Mean average of HRCT Score of right ears with clinical degree of microtia.

Right ears HRCT		Clinical Degree			P-value
		I (n = 2)	II (n = 2)	III (n = 16)	
HRCT Score	Mean	9.0	8.50	8.88	0.345
	±SD	0.0	0.71	0.34	

No statistically significant difference **(p-value > 0.05) between clinical degrees (I, II, III) as regard HRCT score at right ears of the studied group.

Table (8): Mean average of HRCT Score of left ears with clinical degree of microtia.

Left ears HRCT		Clinical degree		P-value
		II (n = 2)	III (n = 10)	
HRCT Score	Mean	**8.0**	**8.9**	**0.533**
	±SD	**1.4**	**0.3**	

**No statistically significant difference (p-value > 0.05) between clinical degrees (II, III) as regard HRCT Score at left side of the studied group.

Table (9): Correlation between HRCT Score and mean average of (ABG, AC & BC) in the studied group.

Rt. Side Variables	(r)	p-value	Lt. Side Variables	(r)	p-value
HRCT Score vs ABG	**-0.02**	**0.932**	**HRCT Score vs ABG**	**- 0.48**	**0.11**
HRCT Score vs AC	**0.03**	**0.898**	**HRCT Score vs AC**	**- 0.45**	**0.13**
HRCT Score vs BC	**0.14**	**0.556**	**HRCT Score vs BC**	**0.47**	**0.12**

**No statistically significant (p-value > 0.05) correlation between HRCT Score and mean average of (ABG, AC & BC) on the right ears.
**No statistically significant (p-value > 0.05) correlation between HRCT Score and mean average of (ABG, AC & BC) on the left ears.

DISCUSSION

This study was conducted on 30 patients with microtia; 17 male (56.7%) and 13 females (43.3%). The mean age of the study group was 7.2 years ranged from 3-19 years. Incidence of bilateral microtia was 6.6%. The degree of microtia was not different between unilateral and bilateral cases.

Microtia has been reported to occur predominantly in male individuals (male-female ratio, 2:1). Furthermore, the incidence of bilateral microtia is reported at 10% to 30%, with right ear involvement in 55% to 65% of unilateral cases **Jafek *et al.* [12], Schuknecht [13]** and **Ishimoto *et al.* [14].**

In the current study the male cases accounted for 56.7%, which was consistent with the phenomenon of male predominance [5, 14, 15, 16, 17, 18].

As regard laterality, this study revealed that the right ear was affected in 62.5% of cases which matched with the previously documented results that reported about right-side predominance in microtia cases [14, 15, 16, 17].

The current study revealed that third degree microtia was the commonest: 26 cases (86.6%), second degree: 4 cases (13.3%) and first degree: 2 cases (6.6%). This was in agreement with previously reported studies [14, 1517].

Ishimoto *et al.* [14] reported that most cases 62 (88%) were with third degree microtia. Also, 329 cases (76%) were with third degree microtia as reported by **Roberson *et al.* [17]. Jin *et al.* [15]** reported that the distribution of clinical degree was as follows: 68 cases (32.6%) grade 0, 32 cases (15%) grade 1, 24 cases (11.5%) grade 2 and 82 cases (39%) grade 3.

In our study, conductive hearing loss was detected in 29 (96.6%) cases, only one case suffered from mixed hearing loss. As regard degree of hearing loss, there were 24 (80%) cases with moderately severe, 2 (6.6%) mild, 5 (16.6%) moderate and one case with severe hearing loss. This agreed with previously reported studies [19, 20, 21] and disagreed with **Jin *et al.* [15].**

Calzolari *et al.* [19] reported that the majority of children (22 ears; 62.9%) had conductive hearing loss; sensorineural and mixed hearing losses were found in four (1 I .4%) and seven (20%) cases respectively.

Ishimoto [20] reported that the hearing level appeared to be affected by the extent of these two abnormalities in patients with microtia. According to the results, the hearing level in microtia with atresia ranges from about 60 to 70 dB in the affected ear, while that in microtia with stenosis ranges from about 54 to 64 dB.

Asma *et al.* [21] reported that the best hearing level for the studied ears was 50 dB which is moderate hearing loss and the worst hearing was 90 dB which is categorized as profound hearing loss based on World Health Organization (WHO) grading of hearing impairment. Four ears had moderate hearing loss, 18 had severe hearing loss and four had profound hearing loss. Most of the studied ears had hearing level of 70 dB preoperatively. The mean of preoperative hearing level was 68.59 dB.

On the other hand, **Jin *et al.*** [15] reported that mixed hearing loss was detected in 55 (53.4%) patients and conductive hearing loss is the rest.

In this study there was Statistically significant difference **(p-value < 0.001)** between Pure-tone average (average of air conduction thresholds at 250 up to8000 Hz), air-bone gap average (at 500 up to 4,000 Hz) and SRT (dBHL) of both ears and clinical degrees in the studied group. However, there was no statistically significant difference **(p-value > 0.05)** between mean average of bone conduction and WR% of both ears and clinical degrees in the studied group. This result was in agreement with [15, 16, 18] However, disagreed with [5, 22]**.**

Jin *et al.* [15] reported that hearing loss deteriorated as the grade of microtia increased. However, **Patil *et al.*** [16] reported that the association between grade III microtia and degree of hearing loss was significant. However, no significant association was found between lower grades of microtia (grade I and II) and degree of hearing loss.

These findings were expected as reported by **Jin *et al.*** [15], the poorer the condition of the external ear the greater the hearing loss due to more anatomical disorders affecting conductive pathway of sound. With respect to the bone-conduction hearing level, the reason for this finding may be that sound waves were transmitted to the cochlea through the skull, irrespective of the state of external ear.

Chen *et al.* [18] reported that the hearing level correlated significantly with external ear abnormalities of Schuknecht's classification. They concluded that the better developed the external ear, the better hearing level.

On the other hand, **Okajima *et al.*** [22] concluded that the conductive hearing loss did not deteriorate as the grade of microtia increased. Also, **Ishimoto *et al.*** [5] reported that there was no relationship between hearing level and severity of microtia scored by Marx classification.

Later on, **Ishimoto** [20] reported that patients with severe microtia when the ear is peanut shaped (Marx's grade III) have more severe hearing loss than patients with an almost normal outer ear but when the relationship between hearing level and the development of the outer ear according to Marx's classification was analyzed there was no relation. Therefore, patients with severe abnormality of the outer ear do not always experience severe hearing loss. These different results could be explained in terms of racial variability.

The present study revealed that there was no statistically significant difference **(p-value > 0.05)** between clinical degrees (I, II, III) as regard HRCT score at both ears of the studied group. This agreed with **Patil *et al.*** [16] and disagreed with **Ishimoto *et al.*** [14] and **Chen *et al.*** [18].

Chen *et al.* [18] reported that they found significant relationship between the development of EAC and all the subgroups of middle ear development: ossicular chain development, windows connected to the cochlea, aeration development of the middle ear, and facial nerve aberration. That is to say, the better developed the external auditory canal, the better developed the temporal bone.

These different results may be explained by limitation of our sample size and different grading systems used in classification as **Ishimoto *et al.*** [14] used in their study modified CT scoring system of **Jarhsdoerfe *et al.*** [2] instead of the classic one used in the current study. **Chen *et al.*** [18] used Schuknecht's grading system for evaluation of external ear abnormalities while we used Marx's classification.

In this study there was no statistically significant **(p-value > 0.05)** correlation between HRCT Score and Pure-tone average (average of air conduction thresholds at 250 up to 8000 Hz) and air-bone gap average (at 500 up to 4,000 Hz) of both sides. This agreed with **Ishimoto *et al.*** [5] and **Asma *et al.*** [21].

Ishimoto *et al.* [5] reported that there is no significant relationship between hearing level and the total CT scoring (full mark, 10 points) as assessed by the CT scoring system of **Jarhsdoerfe *et al.*** [2]. This finding does not mean, however, that specific factors related to temporal bone anomaly do not influence hearing level. Two parameters (i.e, those related to ossicular development and to windows connected to the cochlea) were closely related to hearing levels. Especially the hearing level was affected by window abnormalities. The explanation could be that the sound wave applied to the stapes does not result in a bulk shift of the cochlea scalae, preventing displacement of basilar membrane in otherwise normal cochleae. The traveling wave with all its linear and nonlinear effects could not be launched in a classic manner.

Asma *et al.* [21] reported that there was no significant correlation between preoperative hearing level (HL) with HRCT score based on a Jarhsdoerfe grading system.

CONCLUSION

Microtia is predominant in males. The right side is more likely to be affected. Wide range of anatomic abnormalities are present in the external and middle ear in patients with microtia.

The principle "The better developed the external ear, the better the hearing level" has been proved in Egyptian cases with microtia.

Preoperative assessment of these (as well as other abnormalities including atretic plate pneumatization and aberrant facial and carotid canal with HRCT imaging of temporal bone) plays an important role in planning appropriate management.

Despite of importance of clinical grading of microtia, it may not be possible to predict these abnormalities based on microtia grades, thus emphasizing the importance of imaging in all patients regardless of the clinical grade of microtia.

No prediction of the status of the middle ear could be made based on the hearing level. Therefore, both audiology testing and radiology investigation are two independent evaluations before hearing reconstruction surgery.

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