ORIGINAL ARTICLE

Craniofacial, Temporal Bone, and Audiologic Abnormalities in the Spectrum of Hemifacial Microsomia

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Objectives: To evaluate the clinical, audiologic, and temporal bone computed tomograpic findings in patients with hemifacial microsomia and to use the OMENS (each letter of the acronym indicates 1 of the following 5 dysmorphic manifestations: O, orbital asymmetry; M, mandibular hypoplasia; E, auricular deformity; N, nerve involvement; and S, soft tissue deficiency) grading system to assess possible correlations between the severity of dysmorphic features with the type of abnormalities in the temporal bone and with degree of hearing deficit.

Design: Retrospective study.

Setting: Tertiary care children's hospital.

Patient: Forty patients with hemifacial microsomia.

Result: Mandibular hypoplasia and auricular abnormalities were the most common clinical manifestations, present in 39 patients (97%) and 38 patients (95%), respectively. Conductive hearing loss was noted in 35 patients (86%) and sensorineural hearing loss in 4 patients (10%). Facial nerve weakness was present in 20 patients (50%). Twenty patients had unilateral aural atresia, 12 patients had unilateral aural stenosis, and 7 patients had bilateral anomalies. Moderate hypoplasia or atresia of the middle ear was noted in 36 patients (90%) and ossicles were malformed in 30 patients (75%). Hypoplasia of the oval window was the most common inner ear abnormality.

Conclusions: Severity of craniofacial features (total OMENS score) significantly correlated with the degree of temporal bone abnormality, but no correlation was noted with the degree or type of hearing loss. We recommend the following: (1) use of the OMENS classification system for documentation and analysis of dysmorphic finding in hemifacial microsomia; (2) complete audiologic evaluation in all patients with hemifacial microsomia regardless of the type of craniofacial abnormalities; and (3) temporal bone computed tomography for further evaluation of hearing deficit.

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HEMIFACIAL microsomia (HFM) is a term coined by Gorlin and colleagues.[1,2] This disorder has also been called "otomandibular dysostosis,"[3] "first branchial arch syndrome,"[4] "second branchial arch syndrome,"[5] "oculoauriculovertebral sequence,"[6] "Goldenhar syndrome,"[7] "lateral facial dysplasia,"[8] and "craniofacial microsomia."[9] Hemifacial microsomia manifests in a highly variable phenotype. It is the second most common craniofacial malformation after cleft lip and cleft palate. Any structures derived from the first and second pharyngeal arches can be affected. Although unilateral presentation is preponderant, bilateral anomalies are also seen in 30% of these patients.[10]

This study analyzed the severity of the craniofacial features, temporal bone abnormalities, and audiologic findings in patients with HFM. We also sought to assess a possible relationship between the clinical and dysmorphic features of these patients with the abnormalities in the temporal bone computed tomographic (CT) scans and the severity and type of hearing loss.

RESULTS

CLINICAL FINDINGS

There were 17 male (43%) and 23 female patients (57%), ranging in age from 2 to 37 years (mean age, 12 years). None of these patients had chromosomal anomalies, a history of teratogenesis, or a family history of craniofacial abnormalities. There were 17 (43%) left-sided, 16 (40%) right-sided, and 7 (17%) bilateral presentations (**Table 2**).

The 5 major OMENS features (ie, orbital, mandible, ear, nerve, and soft tissue) are listed in Table 1. Abnormal orbital size

PATIENTS AND METHODS

PATIENTS

The medical records of 162 patients with the diagnosis of HFM seen in the Craniofacial Center at The Children's Hospital, Boston, Mass, were reviewed. Only those patients with complete medical records who had undergone temporal bone CT scan and audiologic workup were included in the study. Forty patients were eligible for this retrospective study.

Medical records were reviewed and data were recorded for the following: the patient's sex, age, family history, facial nerve function, history of otologic disease, and audiologic evaluation. Temporal bone CT scans were reviewed for abnormalities of the external auditory canal (EAC), middle ear, ossicles, inner ear, mastoid development, facial nerve canal, and mandibular condyle.

The OMENS classification system[11] was used to grade the abnormal components in patients with HFM. Each letter of the acronym indicates 1 of the 5 major dysmorphic manifestations: O, orbital asymmetry; M, mandibular hypoplasia; E, auricular deformity; N, nerve involvement; and S, soft tissue deficiency. Each of these 5 anatomical features was graded from 0 to 3 (ie, 0 indicates none; 3, worse) according to the severity using physical examination findings, photographs, and radiographs, including posteroanterior, lateral, and cephalometry. A total OMENS score was obtained by summing the 5 anatomical features (**Table 1**).

A single neuroradiologist (C.D.R.) reviewed the temporal bone CT scans. For analysis, radiographic findings of each anatomical category were assigned an ordinal integer score. Data were recorded on the following: EAC (normal [0], stenosis [1], or atresia [2]), middle ear (normal [0], hypoplastic [1], or atretic [2]). ossicles (normal [0], malformed and fused [1], or unidentified [2]), mastoid (normal [0], poor pneumatization [1], or absent pneumatization [2]), facial nerve (normal [0], displaced [1], or could not be identified [2]), condyle (normal [0], displaced [1], or hypoplastic [2]), and inner ear abnormality (normal [0] or abnormal [1]). A total radiographic score was obtained by summing the scores for each category.

STATISTICAL METHODS

Individual and total OMENS scores were compared with the total radiographic score, type, and degree of hearing loss. Patients manifesting unilateral and bilateral anomalies were analyzed separately. For patients with bilateral involvement comparisons were performed on the basis of side rather than patient since each side was evaluated separately. For analysis, the degree of hearing loss was assigned an ordinal integer score, ie, 0 (normal), 1 (mild), 2 (moderate), 3 (moderate-severe), 4 (severe), or 5 (profound).

Since numerical results were ordinal scores rather than continuous variables, nonparametric procedures were used for all analyses. Correlation between the OMENS scores and both the hearing loss and the radiographic score was assessed using the Spearman rank correlation coefficient (ρ). Comparison of OMENS scores on the basis of type of hearing loss was performed using the Kruskal-Wallis procedure.[5] Patient summaries were expressed as medians, ranges, and frequency distributions. Owing to the nature of this study, the conservative Bonferroni correction for multiple testing was not applied. However, to provide a higher degree of protection to the experimentwise type I error rate, we considered the results to be statistically significant if $P<.01$. Statistical analysis was performed using SAS version 6.12 (SAS Institute, Cary, NC).

Table 1. OMENS Classification System*

Orbit	Facial nerve†
O0 Normal orbital size, position	N0 No facial nerve involvement
O1 Abnormal orbital size	N1 Upper facial nerve involvement (temporal or zygomatic branches)
O2 Abnormal orbital position	N2 Lower facial nerve involvement (buccal, mandibular, or cervical)
O3 Abnormal orbital size, position	N3 All branches affected
Mandible	Soft tissue
M0 Normal	S0 No obvious tissue or muscle deficiency
M1 Small mandible and glenoid fossa with short ramus	S1 Minimal soft tissue or muscle deficiency
M2 Ramus short and abnormally shaped	S2 Moderate soft tissue or muscle deficiency
Subdivisions A and B are based on relative positions of the condyle and temporomandibular joint (TMJ)	S3 Severe soft tissue or muscle deficiency
2A Glenoid fossa in anatomically acceptable position	
2B TMJ inferiorly, medially, and anteriorly displaced, with severely hypoplastic condyle	
M3 Complete absence of ramus, glenoid fossa, and TMJ	
Ear	
E0 Normal ear	
E1 Minor hypoplasia and cupping with all structures present	
E2 Absence of external auditory canal with variable hypoplasia of concha	
E3 Malpositioned lobule with absent auricle, lobular remnant usually inferior anteriorly displaced	

**OMENS indicates the following: O, orbital asymmetry; M, mandible hypoplasia; E, auricular deformity; N, facial nerve involvement; and S, soft tissue deficiency (table adapted from Vento et al[11]).*

†Other involved nerves were analyzed, ie, the trigeminal nerve and hypoglossal nerve.

Table 2. Patient Status

Patient No./ Age, y/Sex	Affected Side	Audiogram Results*	OMENS Classification†
1/10/M	L	R (mild-profound SNHL), L (profound MHL)	O0 M2a E3 N1 S1
2/26/F	L	L (moderate-severe CHL)	O0 M3 E3 N2 S2
3/10/M	L	L (severe CHL)	O0 M2a E3 N1 S2
4/4/M	L	L (moderate-severe CHL)	O0 M1 E1 N0 S0
5/20/F	R	R (profound SNHL)	O0 M2a E3 N1 S0
6/15/F	L	L (severe CHL)	O0 M2b E3 N1 S1
7/13/M	R	R (severe mixed), L (mild CHL)	O0 M2b E2 N0 S1
8/11/F	R	R (severe CHL), L (high-frequency SNHL)	O0 M2b E0 N0 S1
9/15/M	R	R (severe CHL)	O0 M2b E3 N3 S3
10/13/M	R	R (severe CHL)	O0 M2a E3 N0 S0
11/9/M	R	R (moderate-severe CHL)	O2 M2b E3 N0 S2
12/3/F	R	R (severe CHL)	O0 M0 E3 N0 S0
13/4/M	L	L (severe CHL)	O0 M2b E3 N2 S1
14/7/F	L	Normal	O0 M2b E1 N0 S0
15/18/F	L	L (profound CHL)‡	O0 M3 E0 N (hypoglossal) 1 N2 S2
16/21/M	R	R (moderate CHL)‡	O2 M2a E1 N (hypoglossal) 1 N3 S2
17/7/M	R	R (severe CHL)	O0 M2b E3 N2 S2
18/3/M	L	L (severe CHL)	O0 M2b E1 N0 S0
19/12/F	L	L (severe CHL)	O0 M2a E1 N2 S2
20/9/F	L	Bilateral severe CHL	O3 M2b E3 N3 S2
21/4/F	R	R (profound SNHL)	O0 M2b E3 N0 S0
22/2/M	L	L (mild CHL)	O0 M2a E2 N0 S0
23/7/F	R	R (severe CHL)	O0 M2b E3 N0 S0
24/25/F	R	R (severe CHL)	O0 M3 E3 N0 S0
25/37/F	L	L (mild CHL)	O0 M2b E1 N0 S0
26/22/F	L	L (severe CHL)	O0 M3 E3 N2 S2
27/4/F	L	L (severe CHL)	O3 M3 E3 N2 S2
28/7/F	L	L (severe CHL)	O0 M1 E2 N0 S0
29/17/F	R	R (severe CHL), L (moderate CHL)	O0 M2a E3 N3 S3
30/19/M	L	L (moderate-severe CHL)	O0 M1 E3 N0 S1
31/4/M	R	R (profound CHL)‡	O0 M2b E2 N3 S1
32/12/F	R	R (profound CHL)‡	O0 M2b E2 N0 S0
33/12/F	R	R (moderate-severe CHL)	O0 M1 E2 N0 S0
34/19/M	Bilateral	Bilateral-moderate severe CHL	O0 M1 E3 N0 S0
35/11/F	Bilateral	Bilateral-moderate severe CHL	O0 M2b E3 N3 S0
36/17/F	Bilateral	Bilateral severe CHL	O3 M2a E2 (R) N0 S0
37/13/F	Bilateral	Bilateral severe CHL	O0 M2a (R) 2b (Lt) L E3 N2 (L) S1
38/14/M	Bilateral	Bilateral moderate MHL	O0 M1 E1 N1 S2
39/22/F	Bilateral	R (normal borderline), L (severe CHL)	O0 M2a E1 N0 S0
40/9/M	Bilateral	Bilateral moderate-severe CHL	O0 M3 E2 N3 (R) S0

**CHL indicates conductive hearing loss; SNHL; sensorineural hearing loss; MHL, mixed hearing loss; and profound CHL (maximum measurable CHL).*

†OMENS indicates the following: O, orbital asymmetry; M, mandible hypoplasia; E, auricular deformity; N, facial nerve involvement; and S, soft tissue deficiency (table adapted from Vento et al[6]). For bilateral patients if no side is mentioned that would indicate a bilateral manifestation.

‡Profound CHL levels indicated by electrophysiologic test results.

and position were noted in 4 patients (12%) with unilateral HFM and 1 patient (14%) with bilateral HFM. Mandibular hypoplasia, ranging from a small short ramus to complete absence of a ramus, was noted in 32 patients (97%) with unilateral HFM and 7 patients (100%) with bilateral HFM. Unilateral HFM as defined in Table 1 presented as follows: E1 (n=6 [18%]), E2 (n=6 [18%]), and E3 (n=19 [58%]). Bilateral HFM presented as E1 (n=2 [29%]), E2 (n=2 [29%]), and E3 (n=3 [42%]). Six patients (15%) had preauricular tags.

Facial nerve weakness as defined in Table 1 was documented in 16 patients (48%) with unilateral HFM. Four patients had weakness of N1 (frontal or zygomatic), 7 patients had weakness of N2 (ie, buccal, marginal mandibular, or cervical), and 5 had weakness of N3 (complete facial nerve). Two of these patients also had weakness of the unilateral hypoglossal nerve (CN12). Four patients (57%) with bilateral HFM had facial nerve paresis. Two of these patients had bilateral and 2 had unilateral manifestations of facial nerve weakness (Table 2). Soft tissue or muscle deficiency was noted in 19 patients (58%) with unilateral HFM and 2 patients (29%) with bilateral HFM.

A history of chronic otitis media was noted in 20 patients (60%) with unilateral HFM and 3 patients (43%) with bilateral HFM. Fifteen patients (45%) with unilateral HFM and 1 patient (14%) with bilateral HFM had undergone placement of tympanostomy tubes. Five patients (15%) with unilateral HFM and 5 patients (71%) with bilateral HFM were successfully using hearing aids.

AUDIOLOGIC FINDINGS

Patients with unilateral HFM (Table 2) initially had the following: normal hearing (n=1 [3%]), mild conductive hearing loss (CHL) (n=2 [6%]), moderate CHL (n=1 [3%]), moderate-severe CHL (n=5 [15%]), severe CHL

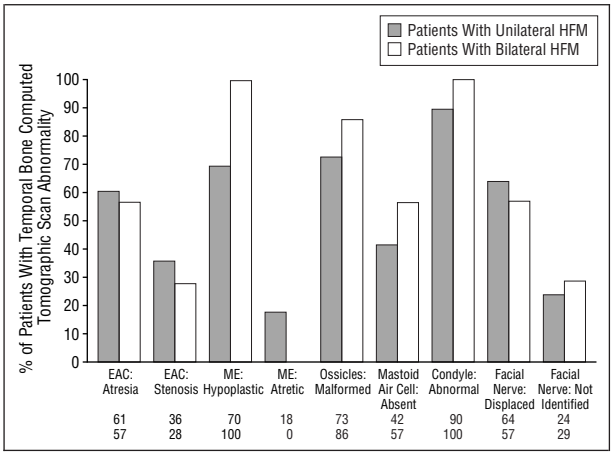

Figure 1. *HFM indicates hemifacial microsomia; EAC, external auditory canal; and ME, middle ear. All values are expressed as percentages of patients.*

(n=1442%]), profound CHL (n=3 [9%]), profound sensorineural hearing loss (SNHL) (n=2 [6%]), and bilateral moderate to severe CHL (n=2 [6%]). Three patients (9%) initially had bilateral hearing loss (Table 2). Patients with bilateral HFM (Table 2) initially had the following: bilateral moderate to severe CHL (n=3 [43%]), bilateral severe CHL (n=2 [29%]), bilateral moderate mixed hearing loss (n=1 [14%]), or unilateral severe CHL (n=1 [14%]).

RADIOLOGICAL FINDINGS

Atresia of the EAC was noted in 20 patients (61%) with unilateral HFM and in 4 patients (57%) with bilateral HFM. Canal stenosis was observed in 12 patients (36%) with unilateral HFM and in 2 patients (28%) with bilateral HFM. Only 1 patient with unilateral HFM had a normal EAC. One patient with bilateral HFM had atresia on one side and stenosis on the other (**Figure 1** and **Figure 2**).

Patients with unilateral HFM had the following findings: normal middle ear (ME) (n=4 [12%]), hypoplastic ME (n=23 [70%], and atretic ME (n=6 [18%]). All patients with bilateral HFM had hypoplastic MEs. Ossicles were malformed and fused in 24 patients (73%) with unilateral HFM and 6 patients (86%) with bilateral HFM. Ossicles could not be identified in 8 patients (24%) with unilateral HFM because of severe atresia of the ME.

Mastoid air cells were absent in 14 (42%) and poorly developed in 8 patients (24%) with unilateral HFM; and absent in 4 (57%) and poorly developed in 2 (29%) patients with bilateral HFM. Thirty patients (90%) with unilateral HFM and 7 patients (100%) with bilateral HFM had displacement and some degree of condylar hypoplasia.

Abnormalities of the facial nerve canal were noted in 35 (88%) of the 40 patients. Anterior displacement of the facial nerve, most commonly in the mastoid segment, was seen in 21 patients (64%) with unilateral HFM and in 4 patients (57%) with bilateral HFM. The facial nerve canal could not be identified in 8 patients (24%) with unilateral HFM and in 2 patients 29% with bilateral HFM.

Hypoplastic oval window was the most common inner ear abnormality and was noted in 12 patients (36%) with unilateral HFM and in 2 patients (29%) with bilateral HFM. An abnormal cochlea was noted in 1 patient with unilateral HFM and in 1 patient with bilateral HFM; these 2 patients had profound SNHL and bilateral moderate mixed hearing loss, respectively. Other inner ear abnormalities noted in patients with unilateral HFM were as follows: hypoplastic vestibule (n=2), hypoplastic semicircular canals (n=1), atretic facial nerve recess (n=1), and atretic round window (n=2).

STATISTICAL RESULTS

Patients' demographics and summarized data are listed in **Table 3** and **Table 4**. Patients with unilateral HFM had a significant correlation between the "E" score (auricular abnormality) and the radiographic score (ρ+0.57, $P<.001$). However, no significant correlation was found for other individual scores (orbit, mandible, nerve, or soft tissue). Total OMENS score (severity of craniofacial features) correlated significantly with the total radiographic score for both unilateral ($\rho=0.47$, $P=.006$) and bilateral ($\rho=0.83$, $P<.001$) presentations. All significant ρ values were positive, indicating that the high total OMENS scores are associated with high radiographic scores and vice versa. No statistically significant correlation between the OMENS scores and the degree or type of hearing loss was observed for either unilateral or bilateral presentations. The *P* values for all statistical tests are summarized in Table 3 and Table 4.

COMMENT

The incidence of HFM has been estimated at 1 in 5600 births.[11] It usually manifests with varying degrees of hypoplasia and asymmetry of bony and soft tissue of the face that is usually unilateral but not uncommonly bilateral—7 patients in our series. It has been suggested that there is a 3:2 predilection for males and the right side of the face.[12] In our sample, no evidence of sex preference or dominance of facial asymmetry to either side was noted; confirming our earlier study of 121 patients.[11]

The cause of HFM is thought to be pathogenically heterogeneous. It is as if the defective genes, teratogens, and vascular anomalies singly or collectively can cause disruption of normal development leading to a wide spectrum of the anomalies seen in these patients. Poswillo[13] devised a chemically induced murine phenocopy and showed that focal hemorrhage from the stapedial artery supplying the first and second pharyngeal arches could be the primary cause. However, it is unlikely that embryonic hematoma formation could account for the wide range of HFM features, particularly those outside the craniofacial regions. Johnston and coworkers[14,15] showed that exposure to thalidomide and retinoic acid could lead to anomalies similar to HFM. Accutane (13-*cis* retinoic acid) interferes with proliferation and migration of neural crest cells, causing facial and cardiac abnormalities similar to the HFM. Otani et al[16] proposed a transgenic mouse model with an insertional mutation on chromosome 10. Hemifacial microsomia can also occur in patients with chromosomal disorders such as trisomy 18, trisomy 7, 9 mosaicism, and terminal deletion of 22q. There are also pedigrees with HFM,

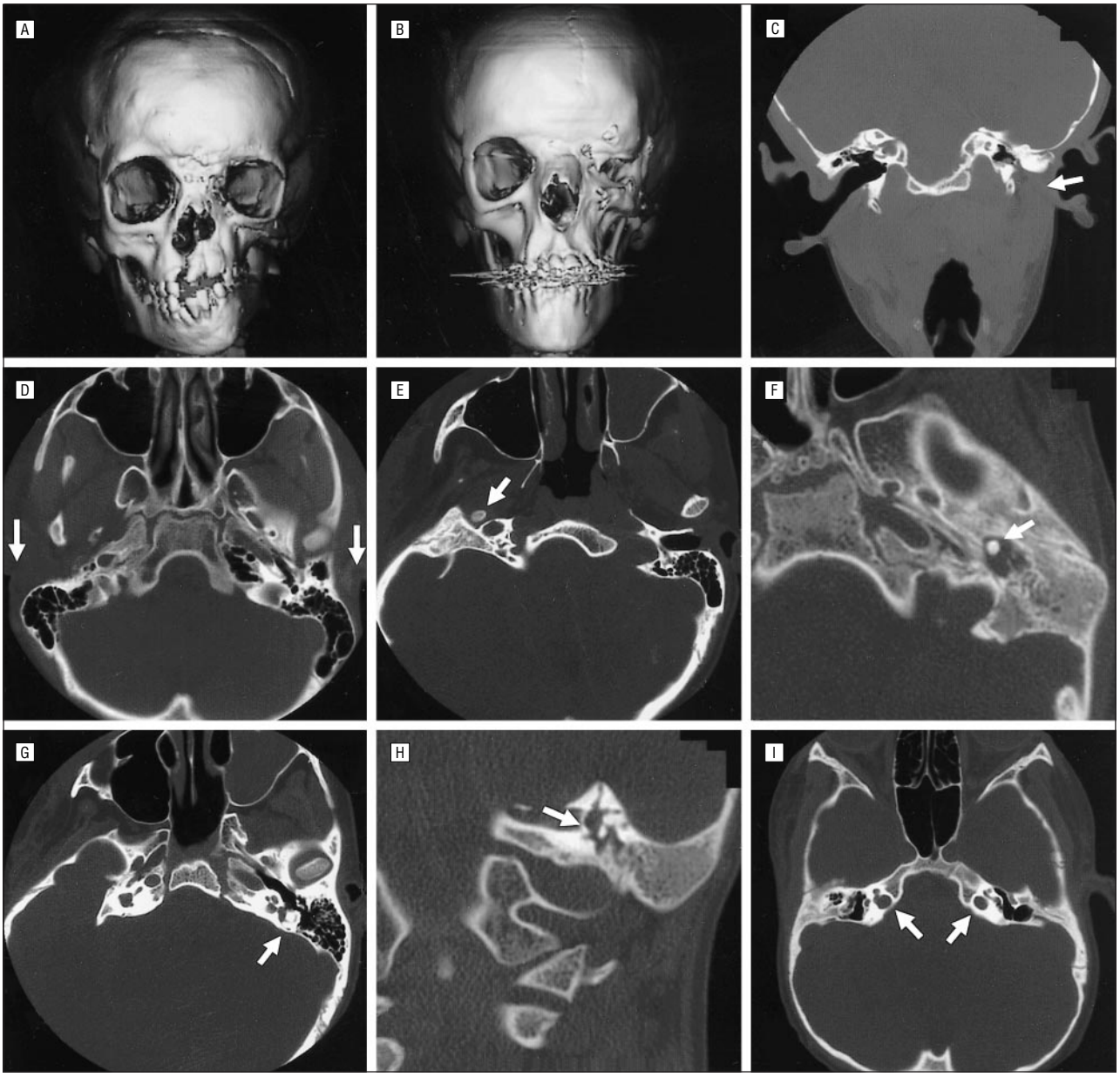

Figure 2. *A-B, A 3-dimensional image of hemifacial microsomia. C, View of unilateral external auditory canal stenosis (arrow). D, View of bilateral external auditory canal stenosis (arrows). E, View of hypoplastic condyle (arrow). F, View of malformed and fused ossicles (arrow). G, view of abnormal semicircular canal (arrow). H, View of hypoplastic oval window (arrow). I, View of abnormal cochlea (arrows).*

and in some affected families the history is consistent with autosomal dominant or recessive inheritance.[17-20]

Several classification systems have been proposed to document and analyze the clinical manifestations of HFM. Mandibular hypoplasia seems to be the element central to all these schemes. Pruzansky[21] described the 3 types of mandibular abnormalities; however, he did not account for other associated manifestations of HFM. Harvold et al[22] described 5 types of mandibular anomalies, but did not include auricular, neural, and orbital anomalies. David et al[12] presented the skeletal, auricular, and soft tissue (SAT) system modeled after the TMN classification of tumors. However, this system does not permit independent evaluation of the orbital and cranial nerve abnormalities. The OMENS system, proposed in 1991,[11] is an expansion of Pruzansky's classification. It includes 5 of the major craniofacial manifestations of HFM and allows independent grading of the dysmorphic features. In this system, each anatomical abnormality is graded from 0 (normal) to 3 (most severe). Cousley[23] compared the SAT with the OMENS system and concluded that the OMENS system was more sensitive to the wide phenotypic heterogeneity of HFM. The OMENS system was later expanded by Horgan et al[24] to OMENS-Plus to include the extracraniofacial anomalies.

The association between HFM and anomalies of the other organs (ie, central nervous system, cardiac, pulmonary, renal, gastrointestinal, and skeletal) has been well documented. Sporadic malformations present in a frequency of 1 in 1000 births (ie, congenital lip and palate, tracheoesophageal fistula, atrial septal defect, and tetralogy of Fallot). The term "associated anomaly" is used

when a defect occurs in 10% to 15% of the cases and is pathogenically related to the primary abnormality. Associated anomalies in patients with HFM are reported as: central nervous system, 5% to 15%[24,25]; cardiac, 45% to 55%[26] and 26.4%[3]; genitourinary, 5% to 6%[25] or 10%[24]; pulmonary and gastrointestinal, 10%[24]; and skeletal, 41%.[24]

Patients with HFM can present with a wide range of anomalies; however, facial nerve weakness and hearing loss are the most common functional deficits. The prevalence of facial nerve palsy with HFM varies in range as seen from the following sources: 10%, Grabb[27]; 19%, Converse et al[28]; 25%, Murray et al[29]; 22%, Bassila and Goldberg[30]; 45%, Vento et al[11]; and 22%, Carvalho et al.[10] In our series 20 (50%) of the 40 patients showed some degree of facial nerve weakness, and 2 patients also had unilateral hypoglossal nerve weakness. Facial nerve weakness could be due to the deficiency of the mesoderm of the pharyngeal arches, neural ectoderm, or a combination of both. Evaluations of the temporal bones have shown the following: (1) abnormal course and exit point of the facial nerve over the temporamandibular joint[28]; (2) development of the nervous intermedius component of the facial nerve, and no development of the mastoid component[31]; (3) hypoplasia of the entire course of the facial nerve.[32]

The most common type of hearing deficit in HFM is conductive loss—34 patients (86%) in our series. However, the presentation of SNHL in these patients remains underappreciated. The overall incidence of congenital SNHL in the general population is 0.001% to 0.004%, and 3% to 4% in patients with craniofacial syndromes.[30] The incidence of SNHL in our patients was 10% (4 patients) consistent with other reports, 11% (Carvalho et al[10]) and 16% (Bassila and Goldberg[30]).

Hemifacial microsomia can manifest as a structural abnormality in any tissue derived from the first and second pharyngeal arches. The mandibular and auricular deformities are the main components of the dysmorphic features. In our series, 39 patients (97%) had mandibular hypoplasia and 38 patients (95%) showed auricular abnormalities. Our data showed a significant correlation between the severity of the external auricular anomalies ("E" score) and the extent of temporal bone abnormalities in patients with unilateral HFM. We also found that the overall severity of craniofacial features (total OMENS score) correlated significantly with the degree of the temporal bone abnormalities (total radiographic score) for patients with unilateral and bilateral HFM. Thus, in patients with abnormal hearing, temporal bone CT scan must be obtained to further assess the degree of stenosis and atresia of EAC, status of the ossicular chain and ME, and inner ear anomalies. The timing of the temporal CT scan must be individualized based on the the age of the patient, severity or worsening of the hearing status, and no otologic surgery. At our craniofacial center, a 3-dimensional CT study is always done in preparation for mandibular elongation in the age range of 5 to 7 years. Whenever possible, a combined study for temporal bone abnormalities and type of mandibular hypoplasia should be done concurrently.

Table 3. Patient Demographics

Patient Characteristic	Type of Hemifacial Microsomia	
	Unilateral (n = 33)	Bilateral (n = 7)
Median age (range), y	11 (2-37)	14 (9-22)
Median total OMENS score (range)*	6 (2-13)	5 (3-8)
Median total radiographic score (range)	8 (2-12)	9 (5-9)
Sex		
F	19	4
M	14	3
Side affected		
L	16	
R	17	
Severity of hearing loss		
Normal	1	1
Mild	2	0
Moderate	1	2
Moderate-severe	5	6
Severe	18	5
Profound	6	0
Type of hearing loss		
Conductive	28	11
Sensorineural	2	0
Mixed	2	2
None	1	1

**OMENS indicates the following: O, orbital asymmetry; M, mandible hypoplasia; E, auricular deformity; N, facial nerve involvement; and S, soft tissue deficiency (table adapted from Vento et al[11]).*

Table 4. Tests of Association (*P*) Predictors*

Outcome	O	M	E	N	S	Total
Unilateral Hemifacial Microsomia						
Hearing loss†	.27	.42	.23	.18	.79	.29
Total radiology†	.13	.02	<.001	.28	.35	.006
Hearing loss type‡	.85	.95	.38	.72	.31	.69
Bilateral Hemifacial Microsomia						
Hearing loss†	.08	.39	.52	.40	.60	.37
Total radiology†	.99	.15	.36	.02	.10	<.001
Hearing loss type†	.74	.15	.14	.55	.022	.27

**OMENS indicates the following: O, orbital asymmetry; M, mandible hypoplasia; E, auricular deformity; N, facial nerve involvement; and S, soft tissue deficiency (table adapted from Vento et al[11]).*

†P value was based on Spearman's correlation coefficient (ρ).

‡P value was based on Kruskal-Wallis test.

Also, our data show that there is no correlation between the severity of the dysmorphic features of HFM and the degree or type of hearing loss. Patients with minimal dysmorphic features can present with a moderate-severe degree of hearing loss. Failure to appreciate this finding could delay proper and timely diagnosis, thereby prolonging sensory deprivation and delayed speech and language acquisition. Thus, we strongly recommend a complete audiologic evaluation of every child with a diagnosis of HFM, regardless of the type or the severity of the clinical manifestations.

CONCLUSIONS

Our results confirmed that evaluation and interpretation of data using the OMENS classification system provide a logical and comprehensive manner to document, independently analyze, and compare the major dysmorphic features of HFM. Furthermore, we agree with Cousley[23] that an asterisk be added to the "E" component of the acronym (OME*NS) to indicate the type and degree of hearing loss.

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