

# Distribution and phenotypes of hemifacial microsomia and its association with other anomalies

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**Objective:** To investigate the distribution and phenotypes of hemifacial microsomia (HFM) and its association with other anomalies. **Methods:** This study included 249 Korean patients with HFM, whose charts, photographs, radiographs, and/or computed tomography scans acquired during 1998–2018 were available from Seoul National University Hospital and Dental Hospital. Prevalence according to sex, side involvement, degree of mandibular deformity, compensatory growth of the mandibular body, and Angle's classification, and its association with other anomalies were statistically analyzed. **Results:** Prevalence was not different between male and female patients (55.0% vs. 45.0%,  $p > 0.05$ ). Unilateral HFM (UHFM) was more prevalent than bilateral HFM (BHFM) (86.3% vs. 13.7%,  $p < 0.001$ ). Although distribution of the Pruzansky–Kaban types differed significantly in patients with UHFM (I, 53.0%; IIa, 18.6%; IIb, 24.7%; III, 3.7%;  $p < 0.001$ ), no difference was observed in occurrence between the right and left sides (52.6% vs. 47.4%,  $p > 0.05$ ). Among patients with BHFM, prevalence of different Pruzansky–Kaban types on the right and left sides was greater than that of the same type on both sides (67.6% vs. 32.4%,  $p < 0.05$ ). Despite hypoplasia of the condyle/ramus complex, compensatory growth of the mandibular body on the ipsilateral side occurred in 35 patients (14.1%). Class I and II molar relationships were more prevalent than Class III molar relationships (93.2% vs. 6.8%,  $p < 0.001$ ). Forty-eight patients (19.3%) had other anomalies, with 50.0% and 14.4% in the BHFM and UHFM groups ( $p < 0.001$ ). **Conclusions:** Patients with HFM require individualized diagnosis and treatment planning because of diverse phenotypes and associations with other anomalies.

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**Key words:** Distribution, Phenotype, Hemifacial microsomia, Association with other anomalies

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## INTRODUCTION

Hemifacial microsomia (HFM) is the second most common facial anomaly, with a broad spectrum of phenotypes.<sup>1-6</sup> Although it usually affects one side of the face, ears, mouth, and mandible, both sides are sometimes affected. HFM has heterogeneous etiological factors related to the alteration of mesodermal development as well as the increased susceptibility to vascular disruption.<sup>7-9</sup> Most patients with HFM require long-term multidisciplinary medical and dental care to rehabilitate occlusal function and improve facial aesthetics.<sup>10-14</sup>

Vento et al.<sup>15</sup> proposed the orbit, mandible, ear, nerve, soft tissue (OMENS) classification system, which grades 5 anatomic features of HFM according to severity on a scale from 0 to 3. Horgan et al.<sup>16</sup> refined the OMENS classification system into the OMENS-plus classification system to account for the presence of extra-craniofacial anomalies (e.g., central nervous system, cardiac, pulmonary, renal, gastrointestinal, and vertebral deformities). Tuin et al.,<sup>17</sup> in their association study between structures, suggested that deformities of structures that mainly developed from the first branchial arch (i.e., the orbit, mandible, and soft tissue) show a similar degree of severity. This also holds true for structures mainly derived from the second branchial arch (i.e., the facial nerve and ear). However, studies have shown that mandibular and ear deformities are the major features of the HFM classification system.<sup>18,19</sup>

The degree of mandibular deformity evaluated using the Pruzansky-Kaban classification system<sup>10,11,20,21</sup> might be one of the most important factors in treatment planning and prognosis prediction of orthodontic treatment using unilateral functional appliances or surgical interventions, including mandibular distraction osteogenesis, orthognathic surgery, and costochondral graft.<sup>4,22,23</sup> If patients with HFM show compensatory growth of the mandibular body despite hypoplasia of the condyle/ramus complex on the ipsilateral side, surgical intervention should be delayed until the completion of growth because of the possibility of severe chin point deviation to the non-affected side. In addition, Angle's classification is another important issue in orthodontic diagnosis and treatment planning. If patients with HFM show a Class III molar relationship, distraction osteogenesis of the mandible cannot be performed because of the possibility of anterior crossbite.

The Korean National Health Insurance Service (KNHIS) plans to provide insurance benefits for orthodontic treatment to patients with HFM in the near future. Basic data on the distribution and phenotypes of patients with HFM can provide useful information for diagnosis, treatment planning, and prognosis evaluation. Several previous studies conducted in other countries have in-

vestigated the clinical characteristics of patients with HFM from the perspective of plastic surgery.<sup>19,24-27</sup> However, no clinical demographic studies have investigated the distribution and phenotypes of Korean patients with HFM from an orthodontic perspective. Therefore, the purpose of this retrospective study was to investigate the distribution and phenotypes of HFM and its association with other anomalies using clinical data.

## MATERIALS AND METHODS

The study sample comprised 249 Korean patients with HFM (mean age at first consultation,  $7.07 \pm 3.99$  years), who visited the Multi-disciplinary Clinic, Department of Pediatric Plastic and Reconstructive Surgery, Seoul National University Children's Hospital (SNUCH) and the Department of Oral and Maxillofacial Surgery and Department of Orthodontics, Seoul National University Dental Hospital (SNUDH) during 1998–2018. The inclusion criteria were as follows: (1) patients diagnosed with HFM and followed-up or treated and (2) patients whose charts, clinical photographs, cephalometric and panoramic radiographs, and/or three-dimensional computed tomography (3D-CT) images were available. HFM prevalence according to sex, side involvement, degree of mandibular deformity, compensatory growth of the mandibular body, and Angle's classification, and its association with other anomalies were investigated. This study was reviewed and approved by the Institutional Review Board of the SNUDH (ER119025).

Since Vento et al.<sup>15</sup> confirmed the concept that mandibular deformity is the cornerstone of the anomaly, the degree of mandibular deformity has been evaluated using the Pruzansky-Kaban classification system. In this system, Type I indicates that the ramus/condyle complex has a normal shape but small size. In Type IIa, although the ramus/condyle complex is hypoplastic and abnormally shaped, the glenoid fossa is placed at the right position and the temporomandibular joint is functional. Type IIb indicates that the glenoid fossa is placed at the inferiorly, medially, and anteriorly altered positions with a severely hypoplastic ramus/condyle complex. Type III indicates the complete absence of the ramus/condyle complex and the glenoid fossa.<sup>11,20</sup>

Compensatory growth of the mandibular body was assessed using panoramic radiographs, posteroanterior cephalometric radiographs, and/or 3D-CT images. Compensatory growth of the mandibular body was identified if the lower dental midline was deviated to the unaffected side in patients with unilateral HFM (UHFM) and to the less severely deformed side in patients with bilateral HFM (BHFM). Angle's classification was assessed via molar relationships by using intraoral photographs or orthodontic models.

Descriptive statistics, chi-square goodness-of-fit test, chi-square independence test, and Fisher’s exact test were used for statistical analysis using SPSS for Windows, ver. 12.0 (SPSS Inc., Chicago, IL, USA). A *p*-value of less than 0.05 was considered significant.

## RESULTS

### Distribution of sex and sidedness

No difference was observed in HFM prevalence between male and female patients ( $n = 137$  [55.0%] vs.  $n = 112$  [45.0%],  $p > 0.05$ ; Figure 1). UHFHFM was more prevalent than BHFM ( $n = 215$  [86.3%] vs.  $n = 34$  [13.7%],  $p < 0.001$ ; Figure 1). Right-sided UHFHFM ( $n = 113$ ) was the most common, followed by left-sided UHFHFM ( $n = 102$ ) and BHFM ( $n = 34$ ) (Figure 1). However, patients with UHFHFM showed no difference in occurrence between the right and left sides ( $n = 113$  [52.6%] vs.  $n = 102$  [47.4%],  $p > 0.05$ ; Table 1).

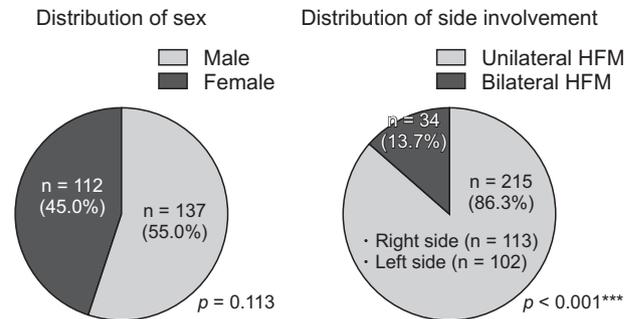
### Distribution of the Pruzansky–Kaban types in patients with unilateral hemifacial microsomia

A significant difference was observed in the distribution of the Pruzansky–Kaban types in these patients (Type I,  $n = 114$  [53.0%]; Type IIa,  $n = 40$  [18.6%]; Type IIb,  $n = 53$  [24.7%]; and Type III,  $n = 8$  [3.7%];  $p <$

0.001; Table 1).

### Distribution of the Pruzansky–Kaban types in patients with bilateral hemifacial microsomia

Patients with different Pruzansky–Kaban types on the right and left sides were more common than were those with the same type on both sides ( $n = 23$  [67.6%] vs.  $n = 11$  [32.4%],  $p < 0.05$ ; Table 2).



**Figure 1.** Distribution of sex and side involvement in patients with hemifacial microsomia (HFM). The chi-square goodness-of-fit test was performed ( $***p < 0.001$ ).

**Table 1.** Distribution of the Pruzansky–Kaban types and side involvement in patients with unilateral hemifacial microsomia

Unilateral type (n = 215)	Pruzansky–Kaban classification		Side involvement		
	Type	<i>p</i> -value	Right side	Left side	<i>p</i> -value
I	114 (53.0)	< 0.001***	65	49	-
IIa	40 (18.6)		22	18	
IIb	53 (24.7)		23	30	
III	8 (3.7)		3	5	
Sum	215		113 (52.6)	102 (47.4)	0.453

Values are presented as number (%).

The chi-square goodness-of-fit test was performed ( $***p < 0.001$ ).

**Table 2.** Distribution of the Pruzansky–Kaban types in patients with bilateral hemifacial macrosomia ( $n = 34$ )

Degree of the condyle/ramus complex deformity between the right and left sides				Sum	<i>p</i> -value
Same	Different				
Both I 7 (20.6)	I-IIa 7 (20.6)	I-IIb 3 (8.8)	I-III 3 (8.8)	20 (58.8)	-
Both IIa 1 (2.9)	IIa-IIb 6 (17.6)	IIa-III 1 (2.9)	-	8 (23.5)	
Both IIb 3 (8.8)	IIb-III 3 (8.8)	-	-	6 (17.6)	
Both III 0 (0)	-	-	-	0 (0)	
Sum 11 (32.4)	Sum 23 (67.6)			34	0.040*

Values are presented as number (%).

The chi-square goodness-of-fit test was performed ( $*p < 0.05$ ).

**Compensatory growth of the mandibular body**

Despite hypoplasia of the condyle/ramus complex, compensatory growth of the mandibular body on the ipsilateral side occurred in 35 patients (14.1%; Table 3). No difference in prevalence of compensatory growth of the mandibular body was observed between patients with UHFM and BHFМ (n = 33/215 [15.3%] vs. n = 2/34 [5.9%], p > 0.05; Table 3).

**Angle's classification**

Class I and II molar relationships were more prevalent than were Class III molar relationships (n = 232 [93.2%] vs. n = 17 [6.8%], p < 0.001; Table 4). No difference in the prevalence of Class III molar relationships was observed between patients with UHFM and BHFМ (n = 12/215 [5.6%] vs. n = 5/34 [14.7%], p > 0.05; Table 4).

**Table 3.** Distribution of compensatory growth of the mandibular body at the ipsilateral side in patients with UHFM and BHFМ

Variable	UHFM	BHFМ	p-value
Compensatory growth	33	2	0.187
No compensatory growth	182	32	
Sum	215	34	

Fisher's exact test was performed.

UHFM, Unilateral hemifacial macrosomia; BHFМ, bilateral hemifacial macrosomia.

**Table 4.** Distribution of Angle's classification in patients with UHFM and BHFМ

Angle's classification	UHFM	BHFМ	p-value <sup>†</sup>	Class I and II vs. Class III	
				Sum	p-value <sup>‡</sup>
Class I and II	203	29	0.064	232	< 0.001***
Class III	12	5		17	
Sum	215	34		249	

UHFM, Unilateral hemifacial macrosomia; BHFМ, bilateral hemifacial macrosomia.

\*\*\*p < 0.001.

<sup>†</sup>Fisher's exact test was performed.

<sup>‡</sup>The chi-square goodness-of-fit test was performed.

**Table 5.** Distribution of the association with other anomalies in patients with UHFM and BHFМ

Variable	UHFM	BHFМ	p-value
Association with other anomalies	32	17	< 0.001***
No association with other anomalies	183	17	
Sum	215	34	

UHFM, Unilateral hemifacial macrosomia; BHFМ, bilateral hemifacial macrosomia.

The chi-square independence test was performed (\*\*\*p < 0.001).

**Association with other craniofacial and extra-craniofacial anomalies**

Forty-nine patients with HFM (19.7%) had other anomalies (n = 32 with UHFM and n = 17 with BHFМ; Table 5). The association with other anomalies was more prevalent in patients with BHFМ than in those with UHFM (n = 17/34 [50.0%] vs. n = 32/215 [14.9%], p < 0.001; Table 5).

Eight kinds of craniofacial anomalies were observed in 42 patients (n = 42/48, 87.5%) and six kinds of extra-craniofacial anomalies were observed in 9 patients (n = 9/48, 18.8%) (Table 6). Three patients had concomitant craniofacial and extra-craniofacial anomalies (Table 6). In terms of the number of associated anomalies, one anomaly was the most common (n = 37/49, 75.5%), followed by two anomalies (n = 11/49, 22.4%) and three anomalies (n = 1/49, 2.0%) (Table 6). Among the recorded anomalies (n = 61 in total; Table 6), relatively high associations were found with Goldenhar syndrome (n = 14/61, 23.0%), Tessier no. 7 craniofacial clefts (n = 10/61, 16.4%), missing teeth (n = 8/61, 13.1%), plagiocephaly (n = 8/61, 13.1%), and vertebral anomaly (n = 7/61, 11.5%; 3 with Klippel-Feil syndrome, 3 with scoliosis, and 1 with torticollis). A relatively low degree of association was found with oral cleft (n = 3/61, 4.9%; 2 with cleft palate and 1 with unilateral cleft lip and palate), cardiac anomaly (n = 3/61, 4.9%), Parry-Romberg syndrome (n = 3/61, 4.9%), micrognathia (n = 1/61, 1.6%), microstomia (n = 1/61, 1.6%), digit anomaly (n = 1/61, 1.6%), and chromosome anomaly (n = 1/61, 1.6%).

Table 6. Summary of the association with other anomalies

Number and kinds of associated anomaly	One anomaly		Two anomalies		Three anomalies	
	UHFM	BHFM	UHFM	BHFM	UHFM	BHFM
1	Goldenhar syndrome* 4	Goldenhar syndrome* 6	Goldenhar syndrome* and #7 craniofacial cleft* 1	Goldenhar syndrome* and #7 craniofacial cleft* 1	Goldenhar syndrome*, #7 craniofacial cleft*, and missing teeth* 1	-
2	#7 craniofacial cleft* 4	#7 craniofacial cleft* 2	Klippel-Feil syndrome† and scoliosis† 1	Goldenhar syndrome* and Klippel-Feil syndrome† 1	-	-
3	Plagiocephaly* 6	Plagiocephaly* 1	#7 craniofacial cleft* and missing teeth* 1	Klippel-Feil syndrome† and plagiocephaly* 1	-	-
4	Oral cleft* 2	Oral cleft* 1	Parry-Romberg syndrome* and missing teeth* 1	Scoliosis† and missing teeth* 1	-	-
5	Torticollis† 1	Cardiac anomaly† 1	Cardiac anomaly† and digit anomaly† 1	Microstomia* and micrognathia* 1	-	-
6	Missing teeth* 3	Missing teeth* 1	Cardiac anomaly† and chromosome anomaly† 1	-	-	-
7	Parry-Romberg syndrome* 2	-	-	-	-	-
8	Scoliosis† 1	-	-	-	-	-
9	Macrostomia* 1	-	-	-	-	-
10	Bell's palsy* 1	-	-	-	-	-
Sum (n = 49)	25	12	6	5	1	0
	37 (75.5)		11 (22.4)		1 (2.0)	

UHFM, Unilateral hemifacial macrosomia; BHFM, bilateral hemifacial macrosomia; -, none.

\*Craniofacial anomalies.

†Extra-craniofacial anomalies.

**Table 7.** Summary of the results of sex, type, side involvement, and the Pruzansky-Kaban types in patients with unilateral HFM from previous studies

Author (yr)	Country (No. of subject)	Study design	Sex		Type		Side involvement*				Unilateral HFM			
			Male (%)	Female (%)	Unilateral HFM (%)	Bilateral HFM (%)	Right side (%)	Left side (%)	Pruzansky-Kaban classification†		Pruzansky-Kaban classification†			
									Normal (M0) (%)	Type I (M1) (%)	Type IIa (M2A) (%)	Type IIb (M2B) (%)	Type III (M3) (%)	
Vento et al. <sup>15</sup> (1991)	USA (154)	Retrospective	44.8	55.2	85.0	16.0	52.9	47.1	11.0	40.0	22.0	17.0	10.0	
Poon et al. <sup>19</sup> (2003)	Australia (65)	Retrospective	54.1	45.9	76.0	24.0	55.3	44.7	9.0	30.0	27.0	23.0	11.0	
Fan et al. <sup>28</sup> (2005)	USA (198)	Retrospective	50.5	49.5	85.0	15.0	52.9	47.1	-	-	-	-	-	
Park et al. <sup>24</sup> (2014)	Korea (100)	Retrospective	63.0	35.0	85.0	15.0	51.8	48.2	0	59.0	21.0	18.0	2.0	
Tuin et al. <sup>30</sup> (2015)	USA (105)	Retrospective	58.1	41.9	77.1	22.9	58.4	41.6	-	-	-	-	-	
Xu et al. <sup>25</sup> (2015)	USA, China, Sweden, Korea, Japan, Taiwan, Italy (1,121)	Meta analysis	52.2	47.8	86.4	13.6	52.8	47.2	-	-	-	-	-	
Caron et al. <sup>27</sup> (2017)	Netherlands, UK, USA (755)	Retrospective	54.0	46.0	88.6	11.4	55.4	44.6	9.2	40.9	26.3	14.1	9.5	
This study (2019)	Korea (249)	Retrospective	55.0	45.0	86.3	13.7	52.6	47.4	-	53.0	18.6	24.7	3.7	

HFM, Hemifacial macrosomia.

\*Calculated from the data.

†Classification was modified from the data.

## DISCUSSION

### Distribution of sex and sidedness

The lack of a difference in the prevalence of HFM between male and female patients was in agreement with the results of previous studies, despite differences in geographical and ethnic factors (Table 7).<sup>15,19,25,27,28</sup>

In the present study, the prevalence of UHFM was higher than that of BHFM. Moreover, no difference was observed in occurrence between the right and left sides among patients with UHFM. These results were also consistent with those of previous studies despite differences in geographical and ethnic factors (Table 7).<sup>15,24,25,27,28</sup>

### Distribution of the Pruzansky–Kaban types in patients with unilateral hemifacial microsomia

The distribution pattern of the Pruzansky–Kaban types in patients with UHFM was similar to that reported by Park et al.<sup>24</sup> However, these results differed from those of previous studies conducted in other countries (Table 7).<sup>15,19,27</sup> The reasons for this difference could be the following. First, the subjects of this study were Korean patients with HFM. Second, in this study, patients with HFM who had skin tags or microtia and insignificant skeletal deformity were considered as showing the M1 (Type I) pattern. Park et al.<sup>24</sup> suggested that patients with microtia could be considered as those with HFM Type I, because all patients with microtia tend to have minimal mandibular deficiency. Therefore, the total number of patients with the M0 (normal) and M1 (Type I) patterns in previous studies (51% in Vento et al.<sup>15</sup>; 50.1% in Caron et al.<sup>27</sup>; Table 7) would be similar to that in our study (53%, Table 7). Third, Takahashi–Ichikawa et al.<sup>29</sup> reported that panoramic radiography was a reliable modality for evaluating patients with HFM Type I, while 3D-CT was preferred for evaluating patients with HFM Type II. Therefore, we recommend the use of two modalities for the clinical assessment of HFM types.

### Distribution of the Pruzansky–Kaban types in patients with bilateral hemifacial microsomia

No previous studies have reported the differences in the Pruzansky–Kaban types between the right and left sides in patients with BHFM. According to the results of this study, different Pruzansky–Kaban types on the right and left sides were more prevalent than was the same type on both sides. This is a novel finding that should be considered, especially when planning distraction osteogenesis of the mandible on both sides.

### Compensatory growth of the mandibular body

Despite hypoplasia of the condyle/ramus complex, compensatory growth of the mandibular body on the

ipsilateral side occurred in 14.1% of patients with HFM (Table 3). This novel finding should be considered during diagnosis, treatment planning, and prognosis prediction for orthodontic and surgical interventions. If compensatory growth of the mandibular body occurs at the affected side, the lower dental midline and chin point would be shifted to the unaffected side. Therefore, when orthodontic treatment using unilateral functional appliances or surgical interventions, including mandibular distraction osteogenesis or costochondral graft, is performed, matching the lower dental midline and chin point to the facial midline is difficult.

### Angle's classification

The finding that Class III molar relationships occurred in 6.8% of patients with HFM has clinical implications. Since patients with HFM usually have simultaneous hypoplasia of the condyle/ramus complex and mandibular body, the origin of the Class III molar relationships should be investigated in future studies. In addition, when patients with HFM show Class III molar relationships, distraction osteogenesis of the mandible cannot be performed at preadolescent and adolescent ages owing to the possibility of anterior crossbite. Therefore, orthognathic surgery should be considered after the completion of growth.

### Association with other craniofacial and extra-craniofacial anomalies

Since the association with other craniofacial and extra-craniofacial anomalies was more prevalent in patients with BHFM than in those with UHFM, attention should be paid to the general medical condition of patients with BHFM for ensuring proper treatment planning and prognosis prediction.

In the present study, eight kinds of craniofacial anomalies were observed in 42 patients (87.5%) and six kinds of extra-craniofacial anomalies were observed in 9 patients (18.8%) (Table 6). Moreover, Goldenhar syndrome was the most commonly associated craniofacial anomaly. However, Goldenhar syndrome tends to be subjectively over diagnosed in patients who show more severe HFM features.<sup>30</sup> The percentage of patients with HFM and extra-craniofacial anomalies in the current study was lower than that reported by Horgan et al.<sup>16</sup> (55.4%), Park et al.<sup>24</sup> (41.4%), and Caron et al.<sup>27</sup> (35.0%). Park et al.<sup>24</sup> reported no significant correlation between the total OMENS score and extra-craniofacial anomalies, despite significant correlations between the orbit and mandible, mandible and soft tissue, and facial nerve and soft tissue.

Despite the potential limitations of data collected from a single university hospital, we obtained meaningful basic information on the distribution and pheno-

types of Korean patients with HFM who were diagnosed and followed-up or treated during the last two decades (1998–2018). The results of this study might serve as the basis for setting guidelines on health care utilization for the orthodontic treatment of patients with HFM. Nevertheless, nationwide multicenter studies with long-term follow-up and systematic statistical analyses have to be conducted to obtain information necessary for providing the sophisticated insurance plans of the KNHIS for orthodontic treatment of patients with HFM.

## CONCLUSION

Patients with UHFM show a significant difference in the distribution of the Pruzansky–Kaban types. In patients with BHFM, different Pruzansky–Kaban types on the right and left sides were more prevalent than was the same type on both sides. Compensatory growth of the mandibular body on the ipsilateral side occurred in 14.1% of patients with HFM. Class III molar relationships occurred in 6.8% of patients with HFM. Other anomalies were more prevalent in patients with BHFM than in those with UHFM. Because of the diverse phenotypes of HFM and its association with other anomalies, patients with HFM require individualized diagnosis and treatment planning.

## CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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