Otolaryngology–Head and Neck Surgery (2007) 136, 33-37

ORIGINAL RESEARCH

Velopharyngeal insufficiency in hemifacial microsomia: Analysis of correlated factors

Emi Funayama, MD, PhD, Hiroharu H. Igawa, MD, PhD, Noriko Nishizawa, MD, PhD, Akihiko Oyama, MD, PhD, and **Yuhei Yamamoto, MD, PhD,** Sapporo and Takamatsu, Japan

OBJECTIVE: To investigate the incidence of unilateral hypodynamic palate (UHP) and velopharyngeal insufficiency (VPI) in hemifacial microsomia (HFM), and to determine the dysmorphic manifestations having significant associations with UHP/VPI in HFM.

STUDY DESIGN: This was a nonrandomized study of 48 patients with unilateral HFM without cleft palate. The correlation between each anomaly and UHP/VPI was analyzed statistically. In addition, we observed 4 HFM patients with cleft palate to examine the influence on cleft palate speech.

RESULTS: The incidence of UHP in HFM was 50.0% and that of VPI was 14.6%. All the VPI patients had UHP. Severe micrognathia and soft tissue deficiency, macrostomia, and mental retardation were significant risk factors for developing VPI in HFM. Moreover, UHP exacerbated speech in HFM with cleft lip and palate.

CONCLUSIONS: Significant correlations were detected between VPI and HFM. This finding should be helpful in the overall management of HFM.

The terms "hemifacial microsomia" (HFM) and "first and second branchial arch syndrome" have been used interchangeably. A wide range of severity and a variety of facial anomalies derived from first and second branchial arch deficiency on the affected side have been described.[1] HFM is often characterized by a lack of development of the external and middle ear, mandible, zygoma, maxilla, and soft tissue, including the facial and masticatory muscles. The palatal musculature is occasionally less developed. As Luce et al[2] demonstrated, some HFM patients can have speech problems, primarily velopharyngeal insufficiency (VPI), which is related to the associated unilateral hypodynamic palate (UHP). Some studies have reported large series of VPI patients without cleft palate,[3-6] in which one category consists of HFM. Dellon et al[7] also emphasized that even unilateral microtia is not a separate entity, but rather a part of the syndrome of HFM, documenting a high incidence of unilateral hypodynamic palate among microtia patients. Therefore, HFM patients have various speech impairments related to UHP/VPI, but the correlated clinical factors that we need to consider have not been determined. Here, we undertook a prospective study to ascertain the frequency of UHP/VPI in HFM patients and statistically determine the phenotypes correlated with it. Moreover, we examined cleft palate with HFM and observed the velopharyngeal pattern and function compared to cleft palate without HFM. The ultimate purpose of this study is to develop clinical means to detect speech problems in HFM patients sooner and initiate the appropriate treatment.

MATERIAL AND METHODS

Subjects

This was a nonrandomized prospective study that reviewed 48 patients, aged 4 to 67 years, with the diagnosis of unilateral HFM or microtia without cleft palate seen at

From the Departments of Plastic and Reconstructive Surgery (Drs Funayama, Oyama, and Yamamoto); and Otolaryngology (Dr Nishizawa); Hokkaido University Graduate School of Medicine, Sapporo; and the Department of Plastic and Reconstructive Surgery, Faculty of Medicine, Kagawa University, Takamatsu (Dr Igawa).

Reprint requests: Emi Funayama, Department of Plastic and Reconstructive Surgery, Hokkaido University Graduate School of Medicine, Address: Kita-15, Nishi-7, Kita-ku, Sapporo 060-8638, Japan.

E-mail address: emi-f@bc.iij4u.or.jp.

0194-5998/$32.00
doi:10.1016/j.otohns.2006.08.020

Hokkaido University Graduate School of Medicine, Japan. In addition, we reviewed 4 HFM patients with repaired unilateral cleft palate (after primary palate push-back), consisting of 2 cleft soft palate (SCP) and 2 unilateral cleft lip and palate (UCLP). The study was performed in accordance with the guidelines of the local institutional review board, and the Helsinki Declaration of 1975.

Evaluating and Grading Each Manifestation

We attempted to determine the manifestations in HFM that are most closely related to UHP and VPI, excluding cleft palate cases. First, all the patients were evaluated and graded separately according to the OMENS classification,[8] which separates HFM into the 5 major dysmorphic manifestations: O, orbital asymmetry; M, mandible hypoplasia; E, ear deformity; N, facial nerve weakness; and S, soft tissue deficiency. Four levels of severity exist for each anatomic category, and the levels are expressed as a numeric scale from 0 to 3, with 0 representing normal and 3 the most severe malformation. This system allows the separate evaluation of each anatomic component independently. In addition to these 5 major dysmorphic manifestations, we examined and recorded the presence of other associated malformations, such as macrostomia, epibulbar dermoids, and vertebral or rib anomalies. For epibulbar dermoids (n = 1), vertebral (n = 1), and rib anomalies (n = 0), too few malformations were found for statistical analysis; consequently, we could only investigate the correlation with macrostomia. Furthermore, we chose to examine the presence of mental retardation because we had experienced a VPI case in microtia exhibiting a slight degree of this condition, and congenital VPI is known to be relevant to it.[1,3]

Intraoral Examination and Speech Assessment

Each patient underwent a direct intraoral examination to observe the soft palate and uvula movement, as well as an articulatory test. Experienced speech-language pathologists conducted all of the perceptual speech evaluations. Patients old enough to have sufficient speech production were evaluated. When any speech problem was detected (in voice production), then a hearing test (for acoustic properties), speech development test (including the perception of the speech sound), and videonasopharyngoscopy (for direct velopharyngeal evaluation) were performed routinely. Velopharyngeal function was rated on a quartile scale published previously (0 = normal resonance; I = mild, infrequent nasal turbulence or mild, fleeting hypernasality, no treatment indicated; II = VPI symptoms that were psychosocially significant, but amenable to change or mild to moderate in character, trial period of speech therapy indicated; III = VPI symptoms severe enough to warrant surgical or prosthetic secondary palatal management).[9]

Statistical Analysis

The relationship between the degree of severity of each anomaly using the OMENS level and the proportion of UHP/VPI was evaluated using Spearman's rank correlation coefficient. In addition, to determine whether nonrandom associations were present between the presence of VPI and each anomaly described above, statistical analyses were performed using Fisher's exact test.

Examination of Cleft Palate

We also examined HFM patients with cleft palate and observed their velopharyngeal pattern and function using videonasopharyngoscopy. The treatment process and speech result were analyzed clinically, and compared to cases without HFM, considering other clinical manifestations due to HFM.

RESULTS

Non–Cleft Palate Group

Of the 48 patients, 24 (50.0%) had UHP. Of these 24 patients, seven (29.2%) demonstrated VPI (in all cases, the VPI grade was II). All VPI patients had UHP. All the VPI patients underwent speech assessment, including endoscopic examination, and they all had asymmetric VPI caused by unilateral abnormal or decreased palatal mobility. All the patients (n = 48) were analyzed statistically to investigate the correlation between the degree of severity of each deformity and VPI, as well as UHP, using Spearman's rank correlation. As mentioned above, we use a score from 0 to 3 to assess the severity of each major anatomic category in the OMENS classification (orbit, mandible, ear, nerve, soft tissue). As the severity of the mandibular hypoplasia worsened, a significant increase occurred in the proportion of UHP/VPI. The same trend was found for the soft tissue deficiency. For the other factors, the correlation coefficients were smaller. Little correlation was observed between the degree of microtia and the proportion of UHP/VPI (Table 1).

In addition to the severity in the OMENS classification, the correlations between VPI and macrostomia and mental retardation were also analyzed. Seven patients with macrostomia were evaluated. Five of the 7 (71.4%) had UHP. Of these 5, 3 patients developed VPI. The macrostomia cases showed a high proportion of UHP/VPI compared to the patients without macrostomia (Table 2). Five patients were mentally retarded. One had relatively severe mental retardation, while 4 exhibited slight degrees of retardation. All the mentally retarded patients had UHP. Interestingly, 100% of them had VPI (Table 2). To determine whether nonrandom associations existed between the presence of VPI and each anomaly described above, Fisher's exact test was performed. Consequently, soft tissue deficiency ($P < 0.01$), mandible hypoplasia ($P < 0.05$), macrostomia ($P < 0.05$), and mental retardation ($P < 0.01$) all had significant associations with the presence of VPI (Table 3). Combined,