Int. J. Oral Maxillofac. Surg. 2016; 45: 679–682
http://dx.doi.org/10.1016/j.ijom.2015.12.003, available online at http://www.sciencedirect.com

International Journal of Oral & Maxillofacial Surgery

Clinical Paper
Cleft Lip and Palate

Cleft characteristics and treatment outcomes in hemifacial microsomia compared to non-syndromic cleft lip/palate

K. M. Dentino[1], A. Valstar[2], B. L. Padwa[1]
[1]Department of Plastic and Oral Surgery, Boston Children's Hospital, Boston, MA, USA; [2]Erasmus University, Rotterdam, The Netherlands

K. M. Dentino, A. Valstar, B. L. Padwa: Cleft characteristics and treatment outcomes in hemifacial microsomia compared to non-syndromic cleft lip/palate. Int. J. Oral Maxillofac. Surg. 2016; 45: 679–682.

Abstract. The goal of this study was to describe the clinical characteristics and treatment outcomes of patients with hemifacial microsomia (HFM) and cleft lip/palate (CL/P), and to compare them to a historic cohort of patients with non-syndromic CL/P treated at the same centre. A retrospective review of patients with HFM and CL/P was performed; the main outcome measures assessed were cleft type/side, surgical outcome, midfacial retrusion, and speech. Twenty-six patients (13 male, 13 female; mean age 22.7 ± 14.9, range 1–52 years) with cleft lip with/without cleft palate (CL±P) were identified: three with cleft lip (12%), two with cleft lip and alveolus and an intact secondary palate (8%), and 21 with cleft lip and palate (CLP) (81%; 15 unilateral and six bilateral). Four patients (19%) had a palatal fistula after palatoplasty. Twelve of 22 patients aged >5 years (55%) had midfacial retrusion and two (9%) required a pharyngeal flap for velopharyngeal insufficiency (VPI). Fisher's exact test demonstrated a higher frequency of complete labial clefting ($P = 0.004$), CLP ($P = 0.009$), midfacial retrusion ($P = 0.0009$), and postoperative palatal fistula ($P = 0.03$) in HFM compared to non-syndromic CL±P. There was no difference in VPI prevalence. This study revealed that patients with HFM and CL±P have more severe forms of orofacial clefting than patients with non-syndromic CL±P. Patients with HFM and CL±P have more severe midfacial retrusion and a higher palatal fistula rate compared to patients with non-syndromic CL±P.

Key words: cleft lip/palate; hemifacial microsomia.

Accepted for publication 3 December 2015
Available online 6 January 2016

Hemifacial microsomia (HFM) is estimated to occur in one of every 5600 live-births and is the second most common craniofacial birth defect after isolated cleft lip/palate (CL/P).[1–3] HFM is a highly variable malformation affecting first and second pharyngeal arch derivatives, including the orbit, maxilla, mandible, ear, cranial nerves, and soft tissues, with a broad spectrum of severity.[4,5] Patients with HFM can have extracraniofacial malformations, most commonly central nervous system, cardiac, and skeletal abnormalities,[6] or additional craniofacial anomalies such as hemipalatal neuromuscular deficiency[5,7] and orofacial clefts.[1,2,8,9]

The reported incidence of orofacial clefting in patients with HFM varies, ranging from 18% to 61% when atypical clefts and macrostomia are included.[1,2,8] Fan et al. found that cleft lip with/without cleft palate (CL±P) occurs in 10% of HFM patients and therefore should also be considered part of the expanded HFM spectrum.[2] They showed that the side and severity of labiopalatal clefting correlates with the severity and predominant side of HFM, suggesting a common etiopathogenesis.[2] HFM is less often associated with isolated cleft palate (Veau type I or II), but patients with HFM frequently have hemipalatal neuromuscular deficiency and velar hypoplasia, which can lead to hypernasality and velopharyngeal insufficiency (VPI).[2,5,7]

There is limited information on cleft characteristics and treatment outcomes in patients with HFM and CL/P. This study was performed to summarize the clinical features and treatment outcomes of patients with HFM and CL/P and to compare them to a historic cohort of patients with non-syndromic CL/P treated at the same tertiary care centre.[10,11]

Materials and methods

This was a retrospective, descriptive study of the clinical characteristics and postoperative outcomes of patients with HFM and CL/P. After approval by the institutional review board, the charts of all patients with a diagnosis of HFM and CL/P treated at the study institution between 1962 and 2013 were identified and reviewed. Subjects with other syndromes, atypical or lateral facial clefting, or macrostomia were excluded. Patients with HFM who required a palatoplasty because of hemipalatal neuromuscular deficiency, velar hypoplasia, or hypoplastic palatal shelves were not considered to have a Veau type I or II cleft palate and therefore were excluded from the analysis.

Descriptive characteristics included age, sex, HFM and cleft laterality (left, right, bilateral), cleft type (complete, incomplete, unilateral, bilateral, or asymmetric), postoperative palatal fistula, midfacial retrusion, and occurrence of VPI.

Speech–language pathologists specializing in cleft care performed perceptual assessments using the Pittsburgh Weighted Values for Speech Symptoms Associated with Velopharyngeal Incompetence.[12] The need for a pharyngeal flap was based on the review of videofluoroscopy and Pittsburgh scale scores by the speech–language pathologist and plastic surgeon. Midfacial retrusion was assessed by the need for Le Fort I maxillary advancement for the correction of sagittal maxillary hypoplasia by examination of intraoral and extraoral photographs; this was done by a single surgeon (B.L.P.).

The outcomes of patients with HFM and CL/P were compared to published data for a historic cohort of patients with non-syndromic CL/P treated at the same tertiary care centre.[10,11] Good et al. studied a cohort comprising 177 patients, 111 male and 66 female, with non-syndromic CL/P.[10] All had completed facial growth at the time of the analysis (males ≥18 years, mean age 21.5 years; females ≥16 years, mean age 20.8 years). Sullivan et al. studied a cohort of 449 patients who had non-syndromic cleft palate, with or without cleft lip (CP±L).[11] This included 246 males and 203 females who had palate repair between 1976 and 2004 and were at least 4 years of age at the time of the study.

Statistical analysis

Patient characteristics were summarized and descriptive statistics calculated using SPSS/SAS (IBM SPSS Statistics version 20.0 (IBM Corp., Armonk, NY, USA); SAS 9.3 software (SAS Institute Inc., Cary, NC, USA)). Associations between categorical variables (postoperative palatal fistula, midfacial retrusion, and the occurrence of VPI) in patients with HFM and CL/P versus the historic cohort of patients with non-syndromic CL/P[10,11] were analyzed using Fisher's exact test. Continuous data were expressed as the mean ± standard error, and comparisons between groups were performed using a χ^2 analysis or Fisher's exact test when appropriate. The resulting *P*-values were two-tailed and considered significant at a value of $P \leq 0.05$.

Results

Demographics

Twenty-six subjects with HFM and cleft lip, with or without cleft palate (CL±P) were included in this study; there were no patients with HFM and isolated cleft palate (CP). Thirteen subjects were male and 13 were female, and the mean age at time of analysis was 22.7 ± 14.9 years (range 1–52 years). The predominant side of HFM was equally distributed between right ($n = 9$), left ($n = 9$), and bilateral ($n = 8$), and there was also an equal distribution of right ($n = 9$), left ($n = 9$), and bilateral labial clefts ($n = 8$). In 22 subjects (85%), the side of the cleft occurred on the predominant side of HFM, and in four patients (15%) with unilateral HFM, the unilateral cleft occurred on the opposite side.

This series included three patients (12%) with cleft lip (CL) only (two (8%) unilateral incomplete and one (4%) bilateral incomplete), two patients (12%) with cleft lip and alveolus (CLA) with an intact secondary palate (one (4%) unilateral complete and one (4%) bilateral asymmetric), and 21 patients (81%) with cleft lip and palate (CLP) (13 (50%) unilateral complete, two (8%) unilateral incomplete, and six (24%) bilateral complete) (Table 1). There were no patients with isolated CP.

Postoperative outcomes

The 26 patients with CL±P underwent nasolabial repair at a mean age of

Table 1. Cleft types.

Cleft type	Number of patients
CL (intact secondary palate)	5
Unilateral complete cleft lip and alveolus	1
Bilateral asymmetric cleft lip and alveolus	1
Unilateral incomplete cleft lip	2
Bilateral incomplete cleft lip	1
CLP	21
Unilateral complete	13
Unilateral incomplete	2
Bilateral complete	6
CP	0

CL, cleft lip; CLP, cleft lip and palate; CP, cleft palate.