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# Velopharyngeal valving during speech, in patients with velocardiofacial syndrome and patients with non-syndromic palatal clefts after surgical and speech pathology management

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ABSTRACT

*Background:* Velocardiofacial syndrome (VCFS) is the most common genetic syndrome associated with cleft palate. There are reports describing several anomalies associated with the palatal cleft in patients with VCFS, which can affect the characteristics of the velopharyngeal insufficiency (VPI) in these cases.
*Objective:* The purpose of this study is to assess velopharyngeal sphincter function during speech, using videonasopharyngoscopy (VNP) and videofluoroscopy (VF), in patients with VCFS, as compared with patients with non-syndromic palatal clefts (NSCP).
*Material and method:* Twenty patients with VCFS corroborated by a FISH test were studied. All patients showed a palatal cleft. All patients had received previous management including speech therapy and palatal repair. These patients underwent a thorough clinical speech evaluation, including VNP and VF. Twenty patients with NSCP matched by sex, type of cleft and within the age range of the patients with VCFS were studied as controls.
*Results:* From the patients with VCFS, seventeen patients showed a submucous cleft palate. Three patients showed sub-total cleft of the secondary palate. Fourteen patients (70%) showed a coronal velopharyngeal closure pattern. Six patients (30%) showed a circular pattern. In contrast, 10 patients (50%) from the NSCP group showed a circular pattern, two of them showed a Passavant's ridge. Seven patients (35%) showed a coronal pattern and 3 patients (15%) showed a saggital pattern. Mean velum (V) and lateral pharyngeal wall (LPW) motion were significantly decreased in patients with VCFS (V = 46% vs 71%; LPW = 14% vs 30%; $P < 0.001$). Size of the defect during speech was significantly increased in patients with VCFS (34.57% vs 67.37%; $P < 0.001$).
*Conclusion:* Velopharyngeal valving during speech is significantly different in patients with VCFS as compared with patients with NSCP. Several anomalies associated with the palatal cleft in patients with VCFS can explain these differences. Thus, the surgical approach for repairing a palatal cleft should consider these differences. Moreover, surgical planning should be performed according to the specific findings of the velopharyngeal sphincter in order to improve speech outcome.
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## 1. Introduction

Velocardiofacial syndrome (VCFS) is one the most common multiple anomaly syndromes in humans. The inheritance pattern was confirmed to be autosomal dominant since the early reports in the 1980 [1–3]. Its genetic pattern was found in 1992 when a microdeletion of chromosome 22 at band q11.2 was demonstrated [4]. This syndrome is now recognized as the most common syndrome associated with cleft palate and velopharyngeal insufficiency (VPI). Moreover, VCFS constitutes 8% of patients with clefts of the secondary palate [5,6]. The most common forms of palatal anomalies in VCFS are submucous cleft palate and occult submucous cleft palate [6]. These clefts can be difficult to identify without videonasopharyngoscopy and/or videofluoroscopy. Many reports suggest that individuals with VCFS have hypernasal speech in the absence of a cleft. However, occult submucous cleft palate is an anomaly that can go undetected until an endoscopic examination of the nasal surface of the velum is performed, identifying an absence (agenesis) or hypoplasia of the musculus uvulae [6]. Although frequency estimates of VPI among individuals with non-syndromic submucous cleft palate is a topic which has created controversy in the related scientific literature, with numbers from 9% to 47% or 51%, in our center, in studies of children with and without resonance and speech disorders, the frequency of VPI in

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cases of non-syndromic submucous cleft palate is actually quite low, probably under 10% [5]. In contrast, patients with VCFS who have a submucous cleft, show VPI in over 70% of the cases. There are several factors contributing to the high frequency of VPI in VCFS. Platybasia, small adenoids, tonsil hypertrophy, hypotonia, congenital velar shortening and abnormalities of pharyngeal muscles have been reported [5–7].

Besides hypernasal speech and nasal emission as a consequence of velopharyngeal dysfunction, other speech disorders and language delay are also common manifestations of VCFS. A high percentage of the speech disorders are associated with cleft palate [5,6,8,9].

The goal in treating VPI is to restore a functional seal of the velopharyngeal sphincter during speech. The aim is to achieve a balanced nasal resonance during articulation. Several surgical options have been reported including Wardill push-back procedure, Furlow's palatoplasty, minimal incision palatopharyngoplasty, which includes specific surgical repair of the levator veli palatine muscle, and other augmentation techniques including fat injection and other synthetic materials injections [5,10]. In addition, individualized velopharyngeal surgery is commonly performed when simple palatal repair fails to completely correct VPI. The most frequently reported procedures are customized pharyngeal flaps and sphincter pharyngoplasties.

There are reports that in non-syndromic submucous cleft palate, a minimal incision palatopharyngoplasty is a safe and reliable procedure for correcting VPI. The use of additional individualized velopharyngeal surgery is usually performed as a second option in unsuccessful cases. However, it has been reported that VPI in cases of VCFS requires a different approach [5,10,11]. It seems that velopharyngeal function during speech is somehow different in patients with VCFS.

The purpose of this study is to assess velopharyngeal sphincter function during speech using videonasopharyngoscopy (VNP) and videofluoroscopy (VF) in patients with VCFS, as compared with patients with non-syndromic palatal clefts (NSPC).

## 2. Material and methods

This study was carried out at the Cleft Palate Clinic of the Hospital Gea Gonzalez in Mexico City. The protocol was approved by the Research Committee and the Bioethics Committee of the Hospital. All patients with VCFS who were evaluated at the clinic from January 2002 to December 2009 were studied.

The active group was assembled with 20 patients with VCFS. All patients had a positive FISH (Fixed In Situ Hybridization) test corroborating a 22q11.2 deletion. We selected patients who showed postoperative VPI. Thirteen patients were females, seven patients were males. Seventeen of these patients showed a submucous cleft palate, whereas 3 patients showed incomplete clefts of the secondary palate. The 3 patients with incomplete clefts of the secondary palate, underwent a minimal incision palatopharyngoplasty according to the surgical routine of the cleft palate clinic as reported earlier, between 4 and 10 months of age [10,12,13]. The 17 patients with submucous cleft also underwent surgical repair of the palate with the same surgical technique, between the ages of 4 and 7 years of age. These patients were operated on at these latter ages, because the presence of velopharyngeal insufficiency (VPI) had to be determined before surgery was indicated [13]. All patients (20 patients) included in the active group for this paper, showed residual VPI after surgery. That is, VPI was demonstrated after surgical repair by speech and resonance clinical evaluation, videonasopharyngoscopy and videofluoroscopy.

From 20 patients included in the active group, 13 patients showed compensatory articulation errors associated with VPI. These patients underwent speech intervention with a speech pathologist which had several years of experience treating patients with VPI and compensatory articulation. Speech therapy was provided according to the protocol routinely used in the clinic as reported earlier [10,12–14]. All these patients received speech therapy until they were able to imitate selected speech samples used in our clinic [10,14–16] as modeled by the speech pathologist. At this point in time, the patient was considered as ready for undergoing a VNP and VF for assessing velopharyngeal function during speech.

All patients with VCFS included in the active group underwent VNP and VF for assessing velopharyngeal sphincter during speech, according to the parameters used in our clinic as described in previous reports [10,14–16]. All procedures were performed by the same phoniatrist (first author of this paper). Besides the phoniatrist, the speech pathologist in charge of each patient was present during the procedures in order to model and facilitate correct production of selected speech samples.

A control group was assembled including 20 patients with non-syndromic cleft palate. These patients were matched by gender and type of cleft with the patients with VCFS included in the active group. Furthermore, the patients included in the control group were within the age range of the patients with VCFS. All these patients showed residual VPI demonstrated by the same protocol as described for the active group. However, although some patients had received speech therapy for different periods of time, none of them showed compensatory errors at the time they were selected for the study.

All patients included in the control group were subjected to VNP and VF with the same protocol used in patients with VCFS.

Age range in patients with VCFS was 8–17 years. Mean age was 11.25; standard deviation was 2.92; median age was 10.50. The age of the patients included in the control group ranged from 8 to 16 years. Mean age was 10.95; standard deviation was 2.72; median age was 10.00. A Mann–Whitney test demonstrated a non-significant difference between the groups ($P = 0.763$).

All videonasopharyngoscopies and videofluoroscopies were evaluated in detail using the videotapes. Velopharyngeal closure pattern, velum motion, lateral pharyngeal wall motion, presence or absence of Passavant's ridge, size of the closure defect and form of the defect were evaluated in all cases. All recordings were assessed by 2 independent examiners. Concordance between the examiners was 100% when the closure pattern was determined. None of the patients showed a Passavant's ridge.

It should be pointed out that assessment of velum motion, lateral pharyngeal wall motion and size of the defect has shown significant variability in previous reports [5,10,15,17]. Nonetheless, they are useful clinical estimates of velopharyngeal function during speech. Thus, in order to enhance efficacy of the evaluations, two examiners assessed all measurements on videotape recordings. Each case was discussed by both examiners together until a consensus was reached. Mean velum motion, lateral pharyngeal wall motion and size of the defect between groups were compared using a Student's *t*-test.

None of the patients with VCFS had been subjected to adenoidectomy or tonsillectomy when they were recruited for the study. Also, none of the patients with NSCF underwent any of these surgical procedures.

## 3. Results

From the group of patients with VCFS, fourteen patients (70%) showed a coronal velopharyngeal closure pattern during speech. Six patients (30%) showed a circular pattern. Passavant's ridge was not observed in any of the cases. None of the patients showed a saggital pattern.

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