

The Occult Submucous Cleft Palate and the Musculus Uvulae

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This report describes *nasopharyngoscopic findings* in 20 patients with small central gaps in the velopharyngeal sphincter. All 20 patients were found to have V-shaped midline defects on the nasal surface of the velum. This finding is contrasted with the finding of a *musculus uvulae* bulge on the nasal surface of the velum in 20 normal subjects. The contribution of the *musculus uvulae* to *velopharyngeal valving* and the entity of the occult *submucous cleft palate* are discussed.

In a recent investigation, Shprintzen et al. (1976) reported on the use of multi-view video fluoroscopy in the evaluation of the velopharyngeal (VP) sphincter in a series of 150 patients with hypernasal speech.

They found that 19 patients (13%) appeared to make velopharyngeal closure on lateral view, but frontal, base, and oblique views showed the presence of small central gaps in the VP sphincter. Oral examination of these patients showed no abnormality of the lip, hard or soft palate, or uvula. Specifically, there were none of the stigmata of submucous cleft palate evident. Subsequently, nasendoscopic evaluation of these patients uniformly revealed defects on the nasal surface of the velum, which appeared to contribute directly to the failure in velopharyngeal valving.

It is the purpose of this paper to describe velopharyngeal insufficiency (VPI) resulting from small central gaps in the VP sphincter in a series of patients, to relate this to the spectrum of overt and occult submucous cleft palate, and to compare the findings to a group of normal subjects.

Method

Twenty patients ranging from 4-2 to 36-4 years of age with a mean age of 10-6 were

isolated who demonstrated 1) hypernasal resonance during speech, 2) small central gaps in the velopharyngeal sphincter on multi-view video fluoroscopy, and 3) normal palatal morphology on oral examination. These patients were examined nasoendoscopically to visualize directly and record the movements of the velopharyngeal sphincter in connected speech. Twenty normal subjects ranging in age from 7-0 to 55-0 years of age with a mean age of 21-4 years were examined and recorded nasoendoscopically performing the same speech tasks.

Experimental Procedure

The technique employed for multi-view video fluoroscopy has previously been described in detail (Skolnick, 1969; Shprintzen et al., 1975). The examinations in lateral, frontal, base, and left and right oblique projections were recorded on a video tape recorder (Sony EV 210) with simultaneous sound and with slow motion and stop-frame capabilities.

Nasoendoscopic examinations were carried out using a flexible fibre-optic naso-pharyngoscope (Machida). The fibroscope utilized was a forward viewing instrument which could be deflected 90° downward at the distal tip by turning the control knob on the instrument eye piece (Figure 1). The scope is 3.9 mm. in diameter at the distal end. A xenon cold light source (Machida) provided sufficient light intensity for filming with an 8 mm. motion picture camera with simultaneous

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magnetic sound recording (GAF 250×1). The nose was prepared using a one-half per cent phenylephrine solution and two per cent tetracaine spray. After adequate topical anesthesia had been obtained, the instrument was passed along the inferior meatus until the choanal orifice was seen. Manipulation of the scope tip allowed an optimal view of the VP sphincter to be obtained. The patients were examined in a sitting position without restraint (Figure 2). A standard sample of speech and nonspeech tasks was used for both video fluoroscopic and nasendoscopic studies (Shprintzen et al., 1977).

The size of the gap in the VP sphincter was measured by inserting catheters of known diameter into the area of closure of the VP sphincter via the opposite nasal cavity. The diameter of the gap was compared directly under vision with that of the catheter and the area of the defect derived. Oral evaluations were made by at least six experienced examiners who noted the integrity of the lip, dental arch, palate and uvula. Judgment of hyper-

nasality from live voice was made independently by two experienced speech pathologists using a four-point scale (normal, mild, moderate, and severe). Interjudge agreement was 0.95. In cases of disagreement, the least severe ranking was chosen.

Results

DEGREE OF V.P.I. The gaps in the VP sphincter during connected speech in the 20 patients with hypernasal speech ranged from 1.0 mm.² to 12.0 mm.² with a mean of 5.0 mm.². All 20 patients in the normal group demonstrated complete closure of the VP sphincter.

VELAR DEFECTS. The most striking finding was a midline V-shaped defect and absence of musculus uvulae bulge on the nasal surface of the velum, seen in all 20 patients with hypernasal speech (Figures 3, 4, 5). All of these subjects were noted to have good velar mobility and good lateral pharyngeal wall motion on video fluoroscopic examination. Observation of the 20 normal subjects showed a clearly defined musculus uvulae bulge in the midline of the velum. The bulge was most pronounced at the point of contact between velum and posterior pharyngeal wall and definitely contributed to VP closure (Figure 6).

ORAL EXAMINATION. Oral examination revealed no abnormality of the lip, dental arch, hard and soft palates, or uvula in either group of patients. Trans-illumination of the palate was negative in all cases.

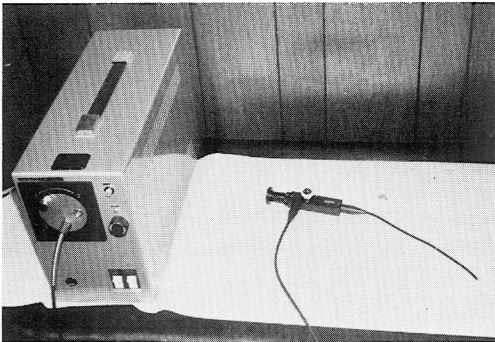


FIGURE 1. Machida 3.9 mm. pharyngo-laryngoscope with xenon light source.



FIGURE 2. Nasopharyngoscopic examination with observer viewing through teaching attachment.

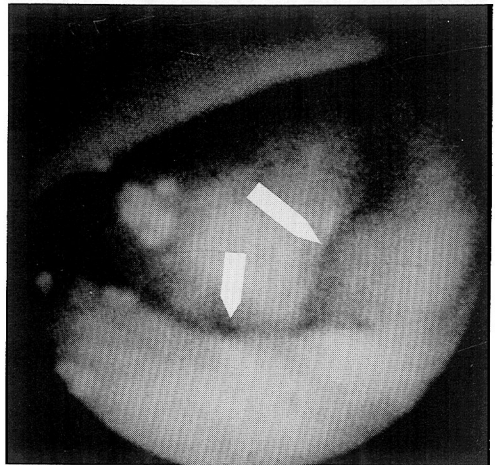


FIGURE 3. Nasopharyngoscopic photograph showing midline V-shaped defect on superior surface of velum and hypoplastic Eustachian tube orifice.

SPEECH. Of the 20 patients with hypernasality, eight patients were judged to have mild hypernasal resonance, six moderate, and six severe hypernasal resonance.

VIDEO FLUOROSCOPY. In "lateral" view, 14 of the 20 subjects appeared to achieve VP closure. However, air bubbles were seen in the barium coating of the nasopharynx, indicating VPI. Frontal and base views confirmed the presence of small central gaps in the sphincter. The type of motion in the velum and lateral pharyngeal wall for all 20 patients did not differ markedly from that seen in normal patients.

Discussion

Over the past few years, there have been several hypotheses advanced concerning the

muscles involved in velopharyngeal valving (Dickson, 1972; Dickson and Dickson, 1972; Shprintzen, 1974 and 1975; Skolnick et al., 1976; Zagzebski, 1976). Dickson (1972), with data collected from anatomical preparations of the velum and base view cinefluoroscopy postulated that levator palati was the only muscle responsible for VP closure, activating both velar and lateral pharyngeal wall motion. Shprintzen et al. (1974 and 1975) hypothesized that a combination of the levator palati and the upper most fibres of the superior constrictor were responsible for closure of the velopharyngeal sphincter. More recently, Shprintzen et al. (1977) reported that we may be mistaken in looking for a single mechanism of velopharyngeal valving, indicating a variable contribution of the levator palati and

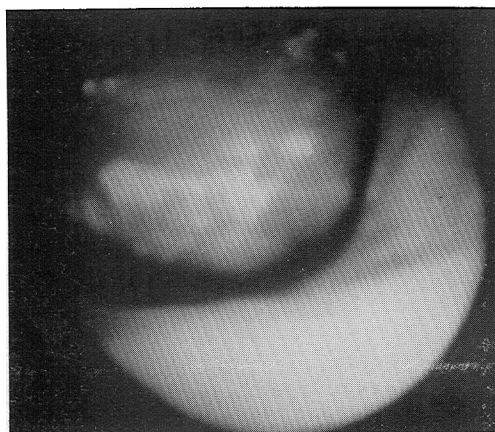


FIGURE 4. Nasopharyngoscopic photograph showing flat surface on superior velum indicating absent musculus uvuli.

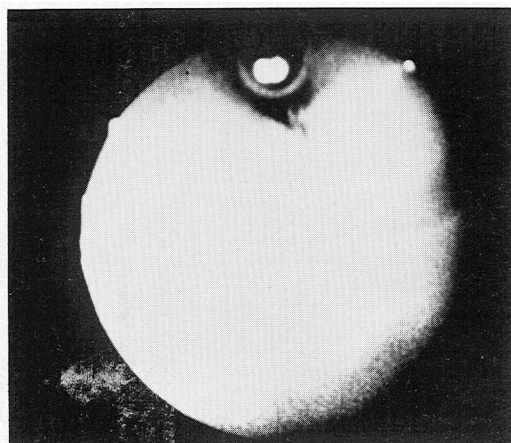


FIGURE 5. Nasopharyngoscopic photograph showing small central gap in VP sphincter with air bubble in mucous.

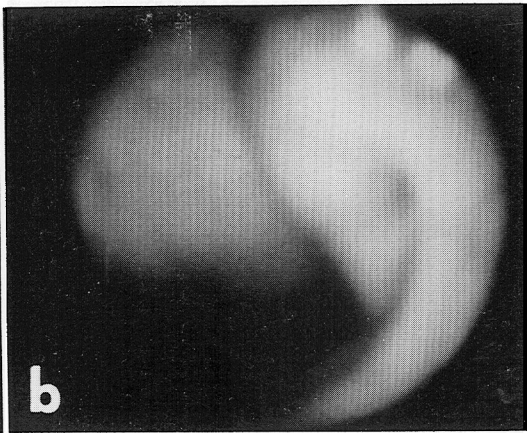
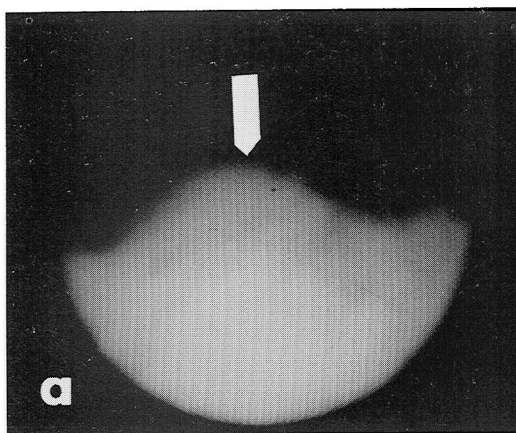


FIGURE 6. Nasopharyngoscopic photograph showing normal subject with prominent musculus uvulae bulge (a) and Eustachian tube orifice (b).

superior constrictor to account for the observation of multiple valving patterns (Skolnick et al., 1973; Zwitman et al., 1975).

None of the studies mentioned above have attributed any role to the musculus uvulae in VP valving. However, Pigott (1969a), and Pigott et al. (1969b), investigating the uses of nasendoscopy in studying the velopharyngeal valve, observed the nasal surface of the velum in 25 cleft palate and 25 normal subjects. He consistently observed in the normal patients a large ridge occupying the central one-third of the soft palate rising to a height almost equal to its width. He attributed this bulk to the musculus uvulae (M.U.) and noted its importance in contributing to velopharyngeal valving during speech movements. Many of his cleft palate subjects showed absence of the M.U. bulge, and he speculated that had it been present, four of his patients would probably have attained VP closure.

A recent study on the morphology of the musculus uvulae by Azzam and Kuehn (1977) shows the M.U. to be a paired muscle running dorsal to the levator sling and exhibiting its most cohesive form at this point. Their study suggests that M.U. contributes substantially to the area of the "velar eminence" and when contracted adds bulk to the nasal surface of the velum, thus facilitating VP closure.

From these studies and from our own observations, we feel that a strong case can be made for the functional importance of the M.U. muscle in normal velopharyngeal valving. It is, therefore, of considerable interest to us that we have encountered a group of patients with velopharyngeal insufficiency, all of whom show a small central gap in the VP sphincter, and all of whom show a deficiency on the nasal surface of the velum. Of interest is the fact that all of the 20 pathologic subjects were misdiagnosed prior to nasoendoscopic and video fluoroscopic evaluation. The referring diagnoses for the 20 subjects with hypernasality included:

- a) palatal paresis—8
- b) CPI—7
- c) velopharyngeal disproportion (short palate)—3
- d) idiopathic hypernasality—1
- e) Hysterical conversion reaction—1

Evaluation techniques utilized to reach the above misdiagnoses included oral manometry, phonation cephalometrics, lateral view

cineradiography, listener judgment, and oral inspection.

On video fluoroscopy and on oral examination, there were no other detectable abnormalities in this group of patients. The palates and uvulae appeared normal. It is our hypothesis that the V-shaped defect noted on the nasal surface of the velum in these patients represents either absence or hypoplasia of the M.U.

Based on the findings reported above, it is interesting to consider Kaplan's (1975) article on occult submucous cleft palate. He demonstrated that subtle defects of palatal musculature may occur and give rise to velar dysfunction and hypernasality, without showing the classic stigmata of an overt submucous cleft palate. However, he stresses that definitive diagnosis is dependent on intra-operative explorations of the soft palate muscles, and the demonstration of abnormal insertion of the levator palati onto the hard palate. In this context, it is interesting to review the electromyographic study of Chaco and Yules (1969) who demonstrated an absence of motor units in the midline of the palates of patients with velopharyngeal insufficiency of unknown origin. Oral examination in these patients failed to reveal any abnormalities and yet the electromyographs were characteristic of submucous cleft palate.

It is our conclusion that the entities of the occult submucous cleft palate and the functional importance of the M.U. have been established. It is our hypothesis that the two can be related in that absence of the M.U. may result in VPI and constitutes a variety of the occult submucous cleft palate.

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