A Concealed Problem: Submucous Cleft Palate and Treatment of Related Speech Disturbances

Mert Çalış, Figen Özgür
Department of Plastic, Reconstructive and Aesthetic Surgery, Hacettepe University School of Medicine, Ankara, Turkey

Abstract

Submucous cleft palate is a specialized subgroup of cleft pathologies, with a worldwide incidence of approximately 1 in 600 live births. Besides the classical triad of bifid uvula, muscle diastasis (zona pellucida) and a notch at the posterior hard palate (defined by Calnan), the anatomical presentation of submucous cleft palate may differ. Unlike overt cleft palate, because of the concealed anatomical appearance, submucous cleft pathologies may be ignored during routine evaluations. For cases with a minor deformity, speech therapy may be initiated as the first-line treatment; however, for cases necessitating surgical treatment, speech therapy must be initiated later. There are various surgical options for the treatment of submucous cleft palate. However, when speech results are taken into consideration, the timing of surgery appears to be much more important than the option selected for surgical treatment. In contrast, we believe that for cases with severe velopharyngeal insufficiency, a pharyngeal flap must contribute to surgical treatment as the first-line option.

Keywords: Submucous cleft palate, velopharyngeal insufficiency, surgical treatment

INTRODUCTION

Submucous cleft palate is a specific sub-group of cleft palate pathologies which demonstrates a mean incidence approximately in one of every 600 live births globally.1 Although it was first described by Roux in 1825, the pathology is acknowledged to have been first described as “submucous” by Kelly in 1910. In 1954 Calnan introduced a systematic description that defined a clinical viewpoint which described its components as bifid uvula, muscle diastasis (zona pellucida), and notching of the bone structure in the hard posterior palate.2

INCIDENCE

Two large-scale field studies are reported in literature that were conducted for identifying the mean incidence rate of submucous cleft palate in the population. In one of these, Weatherley-White et al.3 report to have identified submucous cleft palate in nine schoolchildren out of 10,836 (0.08%). Of the nine, speech dysfunction due to velopharyngeal incompetence was observed in only one child. In another field study, Garcia-Velasco et al.4 report to have identified submucous cleft palate in only one schoolchild out of 6,000 (0.02%). Considering the difficulty in diagnosing the submucous cleft palate and that it can be diagnosed only in the presence of a developing speech dysfunction, it is indeed not possible to establish the actual incidence rate.5

ANATOMY

Apart from the classic triad described by Calnan6, anatomic presentation of the submucous cleft palate can occur in different ways (Figure 1). There is, however, an occult/discrete type not defined by Calnan’s triad that presents a normal anatomy of the soft palate in intraoral assessment and occurs together with speech dysfunction that arises from the incompetence of the musculus uvular (VPI) (Figure 2).7 These cases can be diagnosed only by a nasopharyngoscopic assessment.

Another anatomic variable is the notching seen in the hard palate. A study by Sommerlad et al.8 shows that the notching in the hard palate did not occur at the same rate in all submucous cleft palate cases, and describes three types proportionate to the notching of the hard
palate. Further, Mori et al.\(^9\) show that the speech dysfunction and the VPI findings accompanying bone deformity can be surprisingly disproportionate. Most extreme VPI findings were identified in the patient group with minimal notching of the hard palate, which is defined as type-1.

**CLINICAL PRESENTATION OF SUBMUCOUS CLEFT PALATE**

Unlike classical types of cleft palate, submucous cleft palate cases can be overlooked in routine assessment because of their vague anatomic appearance. These cases are often diagnosed during an advanced assessment performed based on speech dysfunction, adaptation and other social difficulties that occur during kindergarten or school years.\(^{10}\) Incomplete closure between the nasal and oral cavities, and lack of sufficient pressure in the oral cavity leads to hypernasality and articulation disorders. Articulation disorders can vary depending on the structure of the cleft, and their severity increases in proportion to the muscle diastasis. Diagnosis and treatment can be delayed in some extreme cases, since it can be perceived as an articulation-specific speech dysfunction.

Another difficulty which patients with submucous cleft palate can experience, like patients with other types of cleft palate, is eustachian dysfunction due to the incompetence of the soft palatal muscles in providing for the airflow in the eustachian tube.\(^{11}\) This outcome can lead to a chronical imbalance in the pressure of the middle ear and to recurrent serous otitis attacks, including its chronic types. In such cases conductive hearing loss is inevitable in the long run.\(^{12,13}\)

Moreover, based on studies, secondary serous otitis is thought to have a predilection for submucous cleft palate versus other types of cleft palate, which cannot be typically explained by a eustachian dysfunction but additionally involve a problem of the tube dilatation mechanisms. Ventilation tube insertion should be considered at early stages to prevent a potential loss in hearing. The significant improvement observed by Pensler et al.\(^{14}\) to be achieved with respect to serous otitis in submucous cleft palate patients following a repositioning of the levator supports this outcome.

Another problem submucous cleft palate patients can encounter is swallowing difficulty secondary to a chronic nasal reflux that occurs in early ages. Therefore, a detailed examination must be considered in patients presenting swallowing difficulty.\(^2\)

**ASSESSMENT OF VELOPHARYNGEAL INCOMPETENCE**

Assessment of the velopharyngeal incompetence bears importance for identifying the underlying cause and for planning the appropriate treatment. VPI is ideally assessed by a multidisciplinary team consisting of a plastic surgeon, a speech-linguistics therapist, an audiologist, a developmental specialist, ear-nose-throat specialist, and an orthodontist. Ideally, all team members will form a council that functions by simultaneous exchange of information.

Findings of a velopharyngeal incompetence secondary to submucous cleft palate often are hypernasality and associated compensatory articulation disorders.\(^{15}\) Speech disorder is often caused by the inability of the velopharyngeal unit associated with a cleft palate to use the valve mechanism for resisting intraoral pressure increase, hence its inability to prevent airflow into the nasal cavity. This outcome leads to hypernasality.\(^{16}\)

Although VPI is initiated subjectively by a perceptive evaluation performed by a specialized audiologist after clinically taking the patient’s history, it should be confirmed by objective techniques such as pressure-airflow assessments, acoustic analysis (nasometer) and nasopharyngoscopy, or functional magnetic resonance imaging.\(^{17}\) These techniques will be more beneficial in treatment planning and follow-up rather than in diagnosis.

Methods used in Assessing Velopharyngeal Insufficiency\(^{18,19}\)
the pathologic segment for treating submucous cleft palate, in which they performed an excision and reconstruction of the soft palate and muscle dehiscence. In their study it does not constitute a solution for an accompanying shortness of the soft palate and muscle dehiscence. Although simple excision and primary reconstruction of the submucous segment seem to be among the surgical options, patients in which they simultaneously added a pharyngeal flap, and have reported better speech results in the latter group.

Another possible approach following the excision of the submucous segment is to combine one of the described palatoplasty procedures-e.g. V-Y pushback or von Langenbeck palatoplasty. In his 1954-article describing his triad, Calnan has suggested that the only treatment of a submucous cleft palate was the excision of the submucous followed by a V-Y pushback palatoplasty for extending the palate. He reports achieving normal speech levels in 13 of his 18 patients whom he had treated using this approach.

Another study demonstrating that the treatment of the submucous cleft palate can be more challenging than the classic cleft palate was conducted by Massengill et al. In this study, velopharyngeal incompetence was identified postoperatively in four out of twelve patients that had undergone V-Y pushback palatoplasty for the reconstruction of a complete cleft palate, whereas, velopharyngeal incompetence was observed in nine of the twelve patients following the reconstruction of a submucous cleft palate using the same surgical procedure. Although the measurements performed using cinefluoroscopy in both patient groups returned similar palatal lengths, this difference was considered to be associated with the lesser palatal heights and the younger age range of the patients in the submucous cleft palate group. In fact, this outcome demonstrates that the timing of the surgical procedure bears high importance with respect to the achieved speech results regardless of the technique used. Comparing reconstruction procedures performed in earlier years and later years based on the results reported in the literature, it can be confidently said that better results can be achieved with an early reconstruction procedure with respect to velopharyngeal incompetence and associated articulation disorders and that timing is a more significant parameter than the surgical technique.

Pensler et al. defined the age of two as a critical threshold for the timing and demonstrated that submucous cleft palate reconstructions performed before the age of two returned better outcomes compared to those performed in the later years. While the timing of the surgery is inarguably important in submucous cleft palate cases, given the difficulties in its diagnosis and further that it is often diagnosed upon its clinical presentation - usually with speech dysfunction-at school age, diagnosis, and thereby surgical treatment of such cases before the age of two is not always possible. In the same study researchers indicate the necessity for performing intravelar veloplasty in addition to palate lengthening techniques as part of the reconstruction of the submucous cleft palate.

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**Indirect Methods**
1. Perceptive evaluation
2. Acoustic Assessment - Nasometer
3. Accelerometer
4. Pressure-Flow assessments
5. Phototransduction
6. Electromyography (EMG)
7. Low cost methods: Mirror, pipette,

**Direct Methods**
1. Ultrasonography
2. Computerized Tomography
3. Magnetic Resonance Imaging
4. Videofluoroscopy
5. Nasendoscopy (Nasopharyngoscopy)

Although occult submucous cleft palate patients do not demonstrate the physical examination findings seen in classical submucous cleft cases, nasopharyngoscopy assessment has identified a flattening and central depression on the nasal side of the soft palate that was described as “gull wing”.

When submucous cleft palate is diagnosed in later years, speech therapy and surgical treatment should be considered in a complementary manner. While speech therapy can be initiated prior to a surgery in a limited number of cases, it should necessarily be scheduled after the surgery in cases that require surgical intervention. Cases that are treated in later years may require longer therapies, families of patients should be informed to that end. Motivation of both the patient and the family is highly important for a successful therapy.

**SURGICAL TREATMENT OPTIONS IN SUBMUCOUS CLEFT PALATE**

Although the anatomy appears to be less affected in a submucous cleft palate than in a classic cleft palate, its surgical indication is determined by velopharyngeal incompetence and speech dysfunction.

Major options in the surgical treatment of submucous cleft palate are:
1. Excision and primary reconstruction
2. Palatoplasty options (von Langenbeck, two-flap palatoplasty, V-Y pushback)
3. Intravelar veloplasty
4. Pharyngeal Flap
5. Furlow’s Z-Plasty
6. Multiple techniques combined with a pharyngeal flap

Although simple excision and primary reconstruction of the submucous segment seem to be among the surgical options, it does not constitute a solution for an accompanying shortness of the soft palate and muscle dehiscence. In their study of the 1970s, Crikelair et al. have compared their patients in which they performed an excision and reconstruction of the pathologic segment for treating submucous cleft palate, with patients in which they simultaneously added a pharyngeal flap, and have reported better speech results in the latter group.

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length and contribute to the reconstruction of the nasal mucosa, it will also ensure the relation between the soft palate and the posterior pharyngeal wall for treating velopharyngeal incompetence. Assessing the postoperative speech levels achieved in patients who underwent a submucous palatal reconstruction, Abyholm et al. report to have found better speech results in patients treated with a von Langenbeck palatoplasty and a simultaneous pharyngeal flap than those treated with only a von Langenbeck palatoplasty.

Another approach that appears to be an alternative to this approach which includes a pharyngeal flap for the reconstruction of submucous cleft palate cases is the Furlow’s double-opposing Z-plasty (Figures 3a, b). In one of the widest-ranging studies in literature, Chen et al. 25 of the 37 submucous cleft palate cases included in the study, 32 cases that satisfied specific criteria were treated with a Furlow’s double-opposing Z-plasty procedure, and out of these 32 cases velopharyngeal incompetence is reported to remain in only one patient. Providing effective lengthening of the palate using the double-opposing Z-plasty reconstruction technique described by Furlow, and delaying a pharyngeal flap procedure to a second intervention for the surgical treatment of a possible developing velopharyngeal incompetence seems to be a viable option.

The results achieved in the speech levels of 29 patients with submucous cleft palate who were surgically treated at the Hacettepe University Hospitals with Furlow’s palatoplasty and a pharyngeal flap, and simultaneous intravelar veloplasty, were assessed by different phoneme levels using a nasometer. Although the results achieved by the two techniques were found comparable, pharyngeal flap and simultaneous intravelar veloplasty showed significantly better results in patients who had preoperatively presented higher nasality. In our clinical approach we believe that using a pharyngeal flap as part of the first reconstruction procedure will be suitable in patients who are found to present severe VPI findings in preoperative evaluation. It is evident that favorable speech results can be achieved also with Furlow’s palatoplasty in standard submucous cleft palate cases, and that scheduling a pharyngeal flap to secondary surgeries will be more beneficial in avoiding potential side effects cause by the non-use of a flap, such as obstructive sleep apnea.

CONCLUSION

Submucous cleft palate is usually diagnosed in later years as a result of a developing speech dysfunction. In children older than age two, resistant articulation disorders that require long-term speech therapy can persist despite a surgical treatment. Therefore, diligent examination of the palate and nasopharyngoscopic assessment, if suspected, should be performed during infancy. While surgery in early years comes forth as more important than a surgical treatment, Furlow’s double-opposing Z-plasty technique or a pharyngeal flap should be preferred to provide for an optimal length of the soft palate in the surgical treatment of these patients.

Figure 3. a, b. (a) Preoperative view of submucous cleft palate patient. (b) Postoperative view of the 6-year old patient whose submucous cleft palate was reconstructed with Furlow’s double-opposing Z-plasty technique. Extension achieved in the soft palate is significant.
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REFERENCES