
Treatment of Submucous Cleft Palate With Selective Use of the Furlow Z-Palatoplasty

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Purpose: Submucous cleft is an uncommon entity that can be complicated by functional abnormalities, specifically velopharyngeal incompetence (VPI), secondary to abnormal palatal muscular insertion. This study aims to characterize our experience using the Furlow Z-palatoplasty for the treatment of VPI in patients with submucous clefts.

Methods: A retrospective chart review was conducted looking at 24 patients diagnosed with symptomatic submucous clefts between 2000 and 2007 at Children's Hospital of New Orleans. Demographics such as age, gender, diagnosis, need for surgical correction, type of operation, complications, presence of genetic syndromes, need for secondary surgery, and need for myringotomy tubes were examined.

Results: The average age at initial surgery for the entire study population was 6.2 years. The success rate of our Furlow procedure was 66.7%, with 33.3% requiring secondary pharyngeal flaps. The genetic syndromic patient population had an average age at initial surgery of four years and experienced a lower primary success rate of 50%. The non-syndromic patient population had an average age at initial surgery of 7.3 years, with an 85.7% primary success rate.

Conclusions: Our data supports the notion that Furlow Z-palatoplasty is an effective procedure in the treatment of submucous cleft palate with VPI, frequently without the need for secondary surgical procedures in the majority of patients, particularly those patients without syndromes.

INTRODUCTION

First described by Roux more than 100 years ago, submucous cleft is an uncommon condition that is defined by the presence of three anatomical features: bifid uvula, palatal muscular diastasis, and notching of the hard palate.¹⁻³ Kaplan further described the "occult submucous cleft" when he found that the same functional problem can occur in the presence of abnormal muscular insertion without the presence of the classic triad.⁴ These similar conditions both can result in velopharyngeal incompetence (VPI) due to the abnormal insertion of the palatal musculature onto the hard palate instead of forming a continuous muscular sling across the midline.

True submucous cleft occurs uncommonly, with incidence reports ranging from 1:1,200 to 1:20,000.³⁻⁹ Also, the incidence of submucous cleft in patients demonstrating VPI has been reported to be between 27% and 62%.^{4,10} Due to the relative scarcity of these patients, there is a paucity of literature addressing this condition, and its diagnosis and treatment remain a challenge for the physicians that care

for these patients.

In the setting of submucous cleft with associated VPI, numerous procedures have been employed for surgical correction, including pharyngeal flaps, V-Y pushback palatoplasty, minimal incision palatopharyngoplasty, and Furlow Z-palatoplasty.^{3,9,11-13} A consensus approach to treatment remains elusive, however. At our institution, for patients who are unresponsive to speech therapy, the Furlow Z-palatoplasty is used for initial surgical management. Pharyngeal flaps are reserved as secondary procedures for patients in whom the initial Z-palatoplasty is unsuccessful.

The purpose of this study is to report our experience using the Furlow Z-palatoplasty for primary correction of VPI in the submucous cleft population. The efficacy of this procedure was characterized based on subjective improvements in speech and the need for secondary procedures.

METHODS

A retrospective chart review was conducted for those patients diagnosed with submucous cleft that were seen

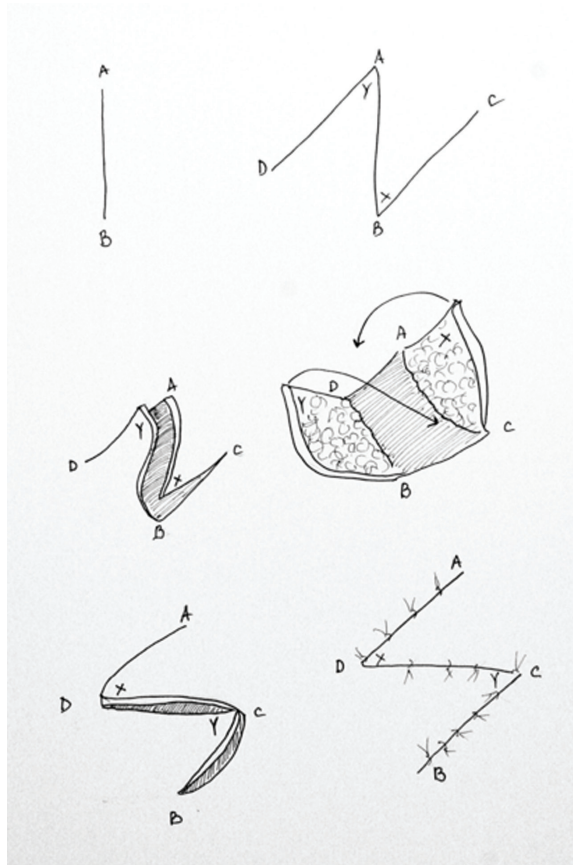


Figure 1: This schematic illustrates the basic concept of a z-plasty, whereby local tissue flaps are juxtaposed in such a manner as to lengthen the original AB tissue segment.

by the Children's Hospital of New Orleans Craniofacial and Cleft Team between 2000 and 2007. This study was approved by the Institutional Review Board. Operative notes and charts were reviewed for data regarding the patients' age, gender, diagnosis, type of operation, need for myringotomy tubes, subsequent complications, length of follow-up, need for secondary procedures, and presence of genetic syndromes. In addition, speech therapy notes were carefully reviewed to track changes in patients' speech patterns before and after surgery.

SURGICAL PROCEDURE

The classic Furlow Z-platoplasty, as described for overt cleft palates, was used for primary surgical management in this series.¹⁴⁻¹⁵ Double opposing z-plasties were designed on the oral and nasal surfaces of the soft palate. The levator and tensor musculature were elevated with a posteriorly based flap that included the nasal mucosa on the right side and oral mucosal on the left side. The interdigitating z-plasties were then closed, restoring the integrity of the palatal muscular sling and effectively lengthening the soft palate.

Patients were evaluated before and after surgery by the speech pathologists on our multi-disciplinary team. Assessments were made on each follow-up visit to evaluate correction of VPI and to determine whether the patient would require further surgical correction. Nasal escape, compensatory substitutions, volume/pitch changes, nasal resonance, tongue placement, oral-nasal balance, and articulation were all assessed.

RESULTS

A total of 24 patients with the diagnosis of submucous cleft palate were treated by our team during the period of time included in this study (Figure 1). Seventeen of the 24 patients required surgical intervention to correct their submucous cleft due to the presence of velopharyngeal incompetence. Fifteen of those 17 patients underwent Furlow Z-platoplasty. Five of those 15 patients later required a secondary pharyngeal flap for persistent VPI, yielding a 67% success rate for correction of VPI by Furlow Z-platoplasty in this group. The other two surgical patients underwent a primary pharyngeal flap, as it was felt that a Furlow Z-platoplasty would have been unsuccessful due to their individual palatal anatomic idiosyncrasies.

The average age at surgery for all 17 surgical patients was 6.2 years, and this group was followed on average for 3.75 years after their initial procedure. Seven of the 24 patients with submucous cleft did not require surgical correction of any kind, as their condition improved with speech therapy alone.

Eleven of the 24 patients had previously been diagnosed with a genetic syndrome (Figure 2). Eight of these 11 patients had VPI that required surgical correction with the Furlow procedure, which provided adequate oral-nasal balance in four of the eight patients, for a 50% success rate in this subgroup. The other four patients demonstrated persistent VPI and required a secondary pharyngeal flap to successfully improve their speech. The average age at surgery for the 11 syndrome-associated patients was four years.

Among the 13 non-syndromic, submucous cleft palate patients, seven had sufficient VPI to require a primary Furlow Z-platoplasty (Figure 3). Six of these 7 cases (86%) resulted in functional oral-nasal balance, with only one (14%) requiring secondary pharyngeal flap for persistent VPI (Figure 4). The average age at surgery for this group was 7.3 years.

Eustachian tube dysfunction and treatment were also analyzed in our submucous cleft population. Thirteen of the 24 (54%) patients required placement of pressure equalizing (PE) tubes by an otolaryngologist. Nine of the 11 syndromic patients (82%) ultimately required PE tube placement, compared with only 4 of the 13 non-syndromic patients (31%).

DISCUSSION

Dysfunction of the velopharyngeal sphincter from

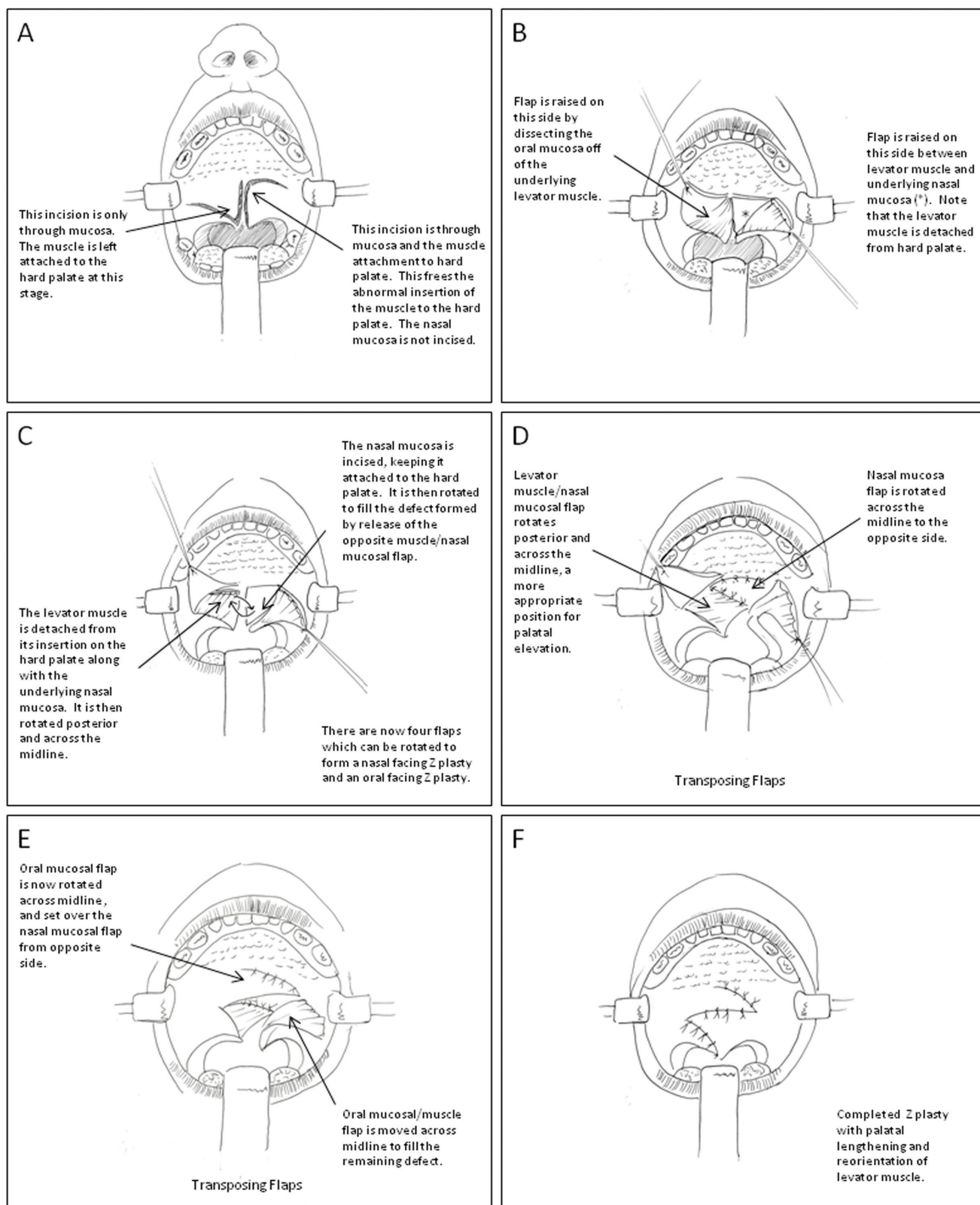


Figure 2: This schematic illustrates the steps involved in the Furlow Z-platoplasty repair of submucous cleft palate, the purpose of which is to recreate anatomically appropriate attachments of palatal musculature, and to lengthen the palate.

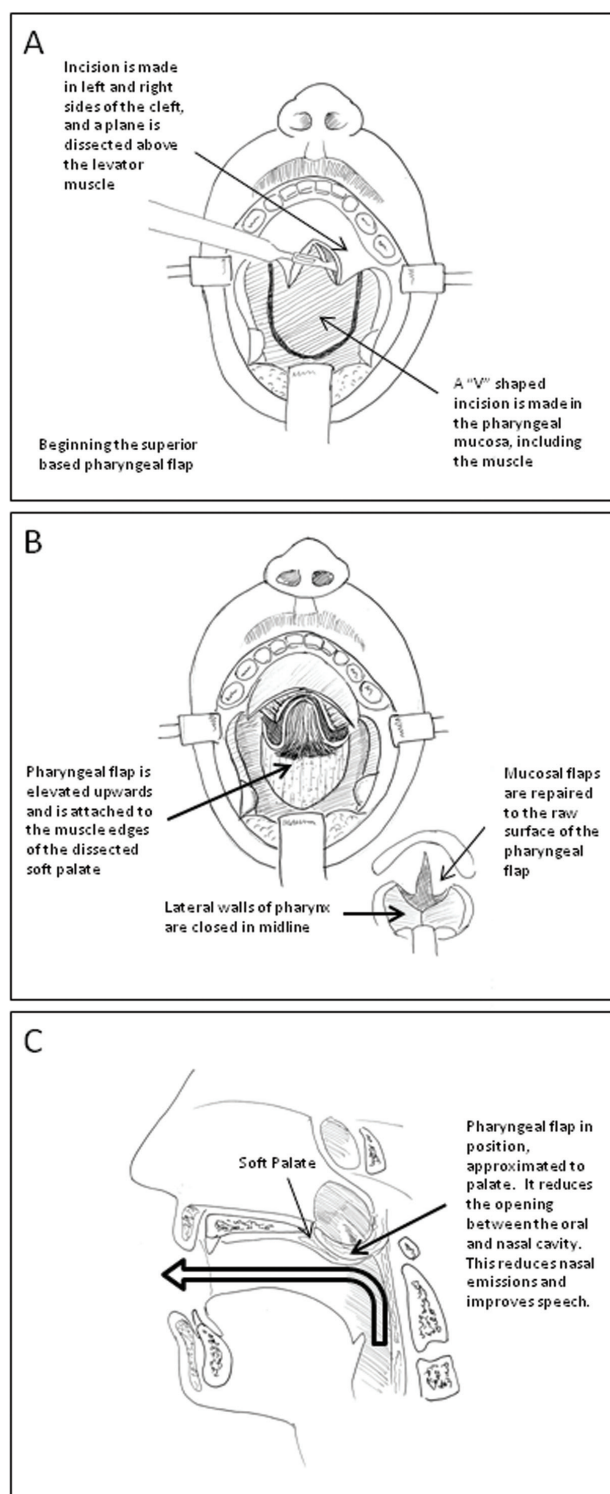


Figure 3: This schematic illustrates the steps involved in pharyngeal flap repair of submucous cleft palate. This repair is not an anatomic repair but rather, reduces the size of the opening between the oral and nasal cavities to reduce the symptoms of velopharyngeal insufficiency.

obvious abnormalities of the pharyngeal wall are readily seen and diagnosed early. However, submucous cleft palate is a relatively rare variant of palatal cleft. Due to the subtle anatomic variations associated with this condition, it may not be recognized on routine well child exams, and diagnosis is often delayed until the child is older, at which time VPI, compensatory corrections, and articulation errors may already be present.¹¹ There have been many attempts to account for this inconsistency, and a number of approaches for evaluation of VPI have been outlined in the literature. Bunke et al. utilized multiple techniques, including detailed intraoral exams, subjective speech evaluations, flow studies involving simultaneous intranasal and intraoral pressure measurements, and lateral cephalometric X-rays.²⁶ Videonasendoscopy and videofluoroscopy have also been described as effective diagnostic modalities.^{9,11} At our institution, the degree of VPI is established by subjective speech evaluation performed by a speech pathologist and the plastic surgeon. We also use nasopharyngeal endoscopy to further characterize patients with equivocal evaluations. In these patients, it is important that language be sufficiently developed to permit proper assessment of oral-nasal balance so that effective characterization of VPI can be achieved.

In conjunction with the inconsistencies that exist regarding diagnosis of submucous clefts, debate remains regarding the optimal timing of surgical treatment, and a number of approaches have been reported. In his experience, Calnan advocated treatment similar to that of an overt cleft palate (assuming the diagnosis is made in time), with correction at about one year of age.² Porterfield advocated repair of all patients at 16 to 18 months of age.¹⁶ There have also been several studies that reported no correlation at all between age at surgery and successful postoperative outcomes, raising further questions about the need for and timing of surgical intervention.^{13,17} Ultimately, most authors recommend waiting until speech has adequately developed so that appropriate evaluation of VPI may be performed before considering surgical correction.^{5,9,11,13,17}

A variety of operative procedures have been described for the surgical treatment of submucous clefts. Calnan reported improvements in speech using a procedure that employed VY retroposition and lengthening of the velum.² Kaplan used the combination of a VY pushback with a superiorly based pharyngeal flap and also reported improvements in speech in that patient series.⁴ In his review of 22 submucous clefts, Cleveland suggested the necessity of a pharyngeal flap to achieve successful correction of speech.¹⁹ Porterfield also suggested the combination of a primary pharyngeal flap in conjunction with a palatal procedure in the primary correction of submucous clefts.¹⁹ However, this procedure is not without risk for significant complications, including airway obstruction, bleeding, flap dehiscence, persistent oral-nasal imbalance, and obstructive sleep apnea.²⁰⁻²⁵ In contrast, there is significant evidence that palatoplasty alone is adequate treatment for VPI.¹¹

Similar to other reported protocols, at our institution patients with marginal insufficiency are treated with an

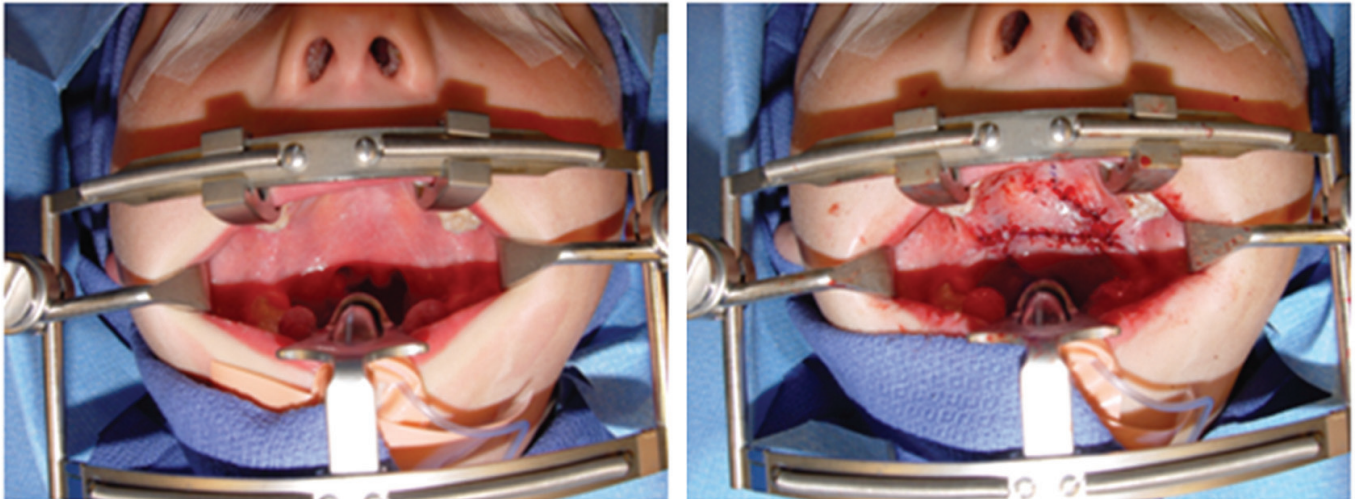


Figure 4: (Left) Patient with submucous cleft palate and VPI requiring Furlow Z-palatoplasty. (Right) The same patient after Furlow Z-palatoplasty.

interventional trial of speech therapy for 6 to 12 months.²⁶ In those patients for whom surgery is indicated, the Furlow Z-palatoplasty is used as our primary repair procedure.¹³ The Z-palatoplasty offers the advantage of directly correcting the dysfunctional anatomy, creating a functional velar musculature while simultaneously lengthening the palate, and avoids undermining the mucosa of the hard palate. Our patients are followed closely after the procedure and only undergo secondary pharyngeal flap if it is clear that there is persistent or recurrent VPI.

The patients in our series experienced a 67% success rate for correction of VPI with primary Z-palatoplasty, 33% of the patients ultimately required a secondary pharyngeal flap. Interestingly, when we stratified our patient population to account for association of genetic syndromes, we noticed a higher rate of failure in the syndromic (50%) when compared to non-syndromic patients (14%). The average age at surgery (4 years in syndromic patients vs. 7.3 years in non-syndromic patients) was almost doubled in the non-syndromic patient population. The need for PE tubes was also more frequent in the syndromic patients. This may indicate that the functional deficit was less severe in the non-syndromic population or that population may potentially have been able to better compensate for their VPI, and thus, further delayed diagnosis.

CONCLUSION

We found the Furlow Z-palatoplasty to be an effective procedure in the treatment of patients with submucous cleft palate and VPI. With this procedure, one can achieve adequate correction of VPI and avoid secondary corrections in the majority of patients, particularly in those patients without evident genetic syndromes. We were able to achieve functional oral-nasal balance in two-thirds of our patients. Through its success at our institution, we recommend the

use of the Furlow Z-palatoplasty in the correction of velopharyngeal incompetence.

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