The Occult Submucous Cleft Palate

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Classic submucous cleft palate is identified by the triad of a bifid uvula, a furrow along the midline of the soft palate, and a notch in the posterior margin of the hard palate (Figure 1). These are the obvious overt physical signs of an underlying anatomic abnormality—the insertion of the levator and other palate muscles onto the hard palate instead of forming a sling across the midline. As a result of this muscle malposition, velar function may be abnormal and velopharyngeal incompetence may result. We now recognize that muscle malposition can occur in the absence of the triad of overt signs. This condition is designated "occult" submucous cleft palate.

We believe that isolated cleft of the secondary palate, submucous cleft palate, and occult submucous cleft palate are variations in expression of the same embryologic disorder; that there is, in fact, a continuous spectrum of severity of muscle malformation and actual clefting. However, we would exclude cleft palate associated with craniostenosis (Apert's syndrome), branchial arch syndromes (Treacher-Collins), mandibular micrognathia (Pierre-Robin), and cleft palate with cleft lip from these considerations because they are probably embryologically distinct conditions. (*2*).

Cleft uvula reportedly occurs in 0.3-10% of the "normal" population without velopharyngeal incompetence and without other physical signs of submucous cleft. (*13*) Too, it has been shown that the classic submucous cleft palate also occurs in 1-10% of the population without speech dysfunction. (*14*) We believe that these are also microforms of secondary palate and submucous clefts which have compensated velopharyngeal function. The terms congenital short palate (*3*) and congenital insufficiency (*7, 11*) may also be variant presentations of occult submucous cleft palate. 75% of our patients had a short soft palate and, therefore, could be termed congenital short palate. However, all of these were subsequently shown to be submucous clefts.

We have reviewed 240 cases of velopharyngeal incompetence without cleft lip or cleft palate (Figure 2) and have identified 41 cases of classic submucous cleft and 23 cases of occult submucous cleft. The other 167 cases of non-cleft velopharyngeal incompetence were due to a variety of other causes but most were probably patients with occult submucous clefts or other microforms that were not diagnosed because of our initial lack of appreciation of this problem. Patients with velopharyngeal incompetence are classified into four categories:

1) ANATOMIC DISPROPORTIONS WITH NORMAL PALATE MUSCLE FUNCTION. Most of these patients have either short soft palate or deep nasopharynx. Many,

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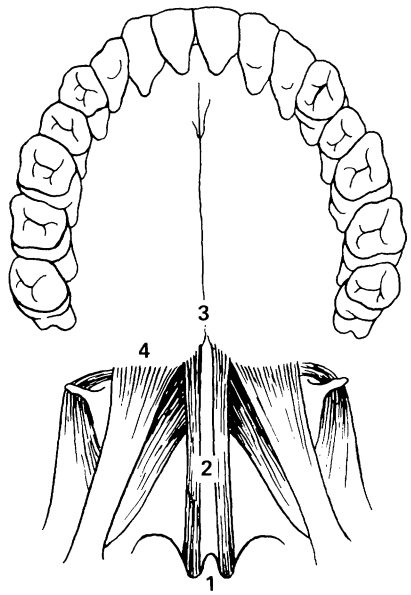


FIGURE 1. The classic submucous cleft showing the overt signs and the abnormal muscle position.

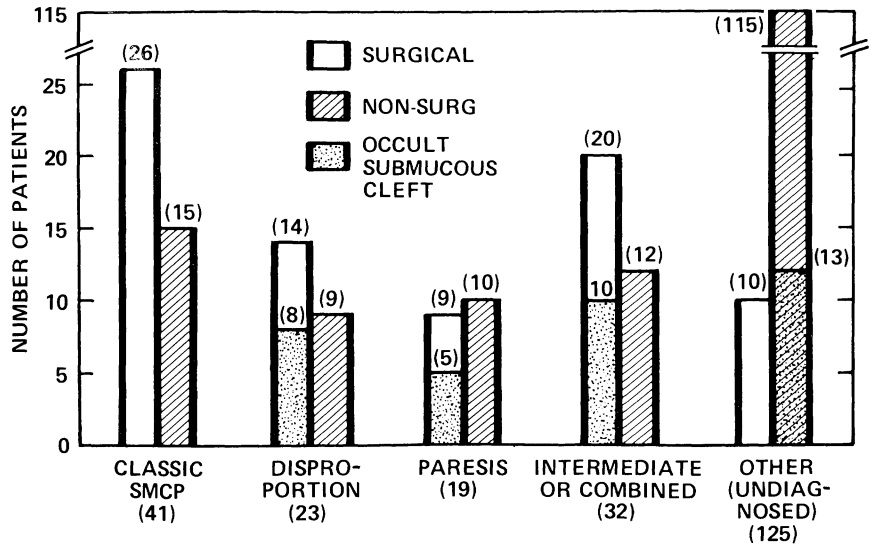


FIGURE 2. Non-cleft velopharyngeal incompetence is subdivided according to the mechanism causing dysfunction. The occult submucous cleft palate (speckled bar) represents about 50% of all of our diagnosed cases. Over one-half of the cases were undiagnosed. These were patients seen during the first five years of the 10-year study when we did not yet recognize the existence of occult submucous cleft palate and also included patients with mild velopharyngeal incompetence not requiring surgery which, therefore, could be definitively identified.

if not all, with short soft palates probably were occult submucous clefts. However, because of our initial lack of awareness of occult clefts, we did not make a diagnosis. Many of the patients with deep nasopharynx had had a prior

tonsillectomy and adenoidectomy. Eight cases of occult submucous cleft palate were identified in this group.

2) MUSCLE DYSFUNCTIONS WITH NORMAL ANATOMIC PROPORTIONS. These patients have palate muscle paresis due to any one of the primary neuromuscular diseases. Five cases of occult submucous cleft palate were in this group.

3) INTERMEDIATE. These patients had both anatomic and dynamic abnormalities. Ten cases of occult submucous cleft were in this group.

4) INDETERMINATE. These are patients who did not clearly fit into any of the major categories. Thirteen cases were suspected occult submucous clefts, but they did not have surgery to confirm the diagnosis.

Diagnosis of Occult Submucous Cleft

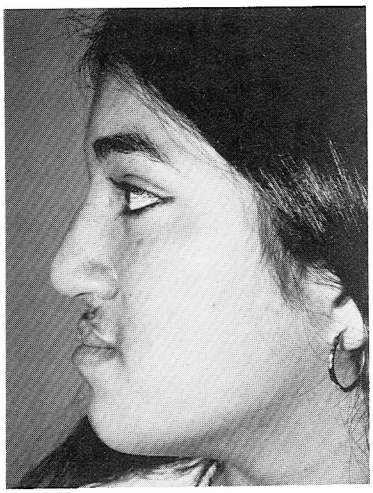
We have reviewed our experience and can now, in many cases, make a presumptive diagnosis of occult submucous cleft palate based on 1) characteristic facial features; 2) cephalometric analysis; and 3) cinefluorographic voice studies. Ultimately, however, the *definitive diagnosis is dependent upon the intra-operative exploration of the soft palate muscles.* The approach to the diagnosis, selection of surgical candidates, and methods and results of surgery will be discussed.

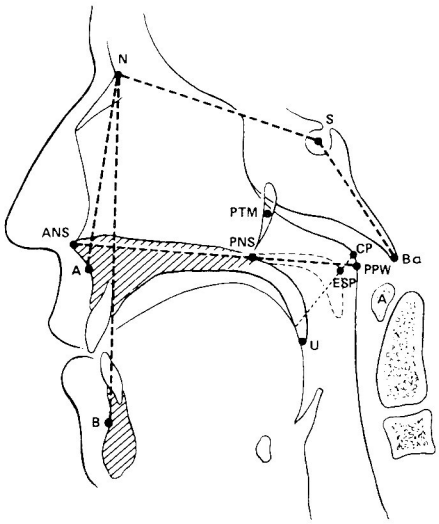
1. FACIAL FEATURES. There are characteristic facial features seen in patients with classic submucous cleft, the occult submucous cleft, and in some patients with clefts of the secondary palate. These features should suggest the diagnosis of occult (or classic) submucous cleft. Not all of these features occur in every patient. Some of the patients with proven occult submucous cleft palate had none of these signs and some had all of them. These deformities are listed with the frequency indicated in parentheses:

a) maxillary hypoplasia—"dish face" (75%)
b) lip contour deformity at vermilion border—"gull wing" (75%)
c) drooping of oral commissure (25%)
d) dynamic facial muscle abnormality (25%)
   paranasal bulge—horizontal
   lateral lip bulge—vertical
   hypoanimation—a "dull" face or expression
e) external ear abnormality—flat arc of superior helix (10%)
f) alveolar arch abnormalities (5%)

It is apparent that all of these features (except the ear anomaly) are related to mesodermal deficiencies of the maxillary process of the first branchial arch. This is consistent with the fact that the mesoderm (muscle) of the soft palate is also embryologically derived from the maxillary component of the first branchial arch. However, the motor innervation of the maxillary process comes from the hyoid arch (VIIth nerve) and the pharyngeal plexus (IXth and Xth nerves). Many of these facial features are seen in other cranial and branchial arch syndromes. Thus, these are characteristic of maxillary hypoplasia and not necessarily specific to submucous cleft.

a) Maxillary hypoplasia (Figure 3). The tendency towards flattening of the





POINTS:

A = ANTERIOR TUBERCLE OF ARCH OF ATLAS
ANS = ANTERIOR NASAL SPINE
Ba = BASION, ANT. LIP OF FORAMEN MAGNUM ON BASISPHENOID
CP = CLOSURE POINT, PROBABLE POINT OF MIDSAGITTAL CONTACT OF PALATE AGAINST POSTERIOR PHARYNGEAL WALL
ESP = MOST POSTERIOR POINT OF SOFT PALATE KNEE IN MAXIMUM ELEVATION
N = NASION, ANTERIOR LIMIT OF FRONTO-NASAL SUTURE
PNS = POSTERIOR NASAL SPINE, USUALLY ESTIMATED AT JUNCTIONS OF PTM WITH HARD PALATE
PPW = POSTERIOR PHARYNGEAL WALL AT LEVEL OF HARD PALATE PLANE
PTM = PTERYGOMAXILLARY FISSURE
S = SELLA TURCICA, MIDPOINT
U = TIP OF UVULA (OR POSTERIOR EDGE OF SOFT PALATE)

LINEAR MEASUREMENTS:

ANS→PNS = HARD PALATE LENGTH
PNS→PPW = NASOPHARYNGEAL DEPTH (IN HARD PALATE PLANE)
PNS→U = SOFT PALATE LENGTH
PNS→ESP = EFFECTIVE SOFT PALATE LENGTH
ESP→CP = VELOPHARYNGEAL GAP

ANGULAR MEASUREMENT:

N→S→Ba = CRANIAL BASE ANGLE
S→N→A = MAXILLARY POSITION
S→N→B = MANDIBULAR POSITION

FIGURE 3. The "dish face" maxillary hypoplasia cephalometric measures of SNA and hard palate length are normal.

maxilla is somewhat difficult to appreciate in young children, but becomes more apparent in the teenager and adult. Slight flattening occurs primarily over the maxilla, but minimally involves the premaxilla (central lip and alveolus).

Standard cephalometric measurements (SNA-SNB, hard palate length) are within 2 standard deviations of normal for age but are consistently less than the average for age. (*12*)

b) *Lip contour deformity along the vermilion border* (Figure 4). At the lip junction of the frontonasal (premaxilla) and the maxillary processes there is a deficiency of mesoderm that results in paired furrows in the margin of the lip vermillion. We describe this deformity as having a gull-wing contour. This deformity is more common in the submucous cleft classification Groups 1 (short soft palate), 3 (classic submucous cleft) and 4 (combined), and not typical of Group 2 (paretic).

c) *Drooping of the oral commissures* (Figure 5). The normal lip has a horizontal relation when a line is drawn from one oral commissure through the inferior aspect of the vermilion "tear drop" and the opposite commissure. Children with occult submucous cleft palate, particularly those in Group 2 (paretic palates), have a downward displacement of the commissures relative to the center of the lip which resembles a frown. This contour is probably secondary

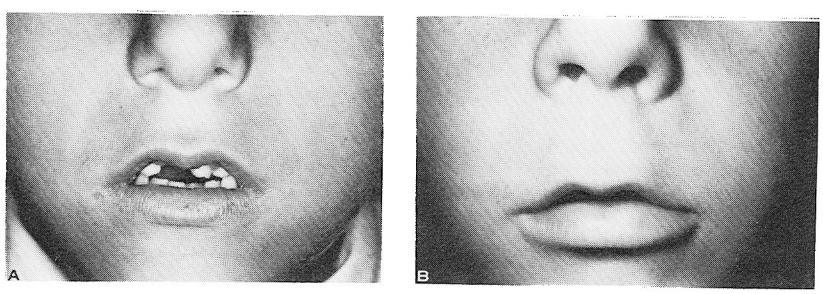


FIGURE 4. Lip contour deformity ("gull wing") along the vermilion border.

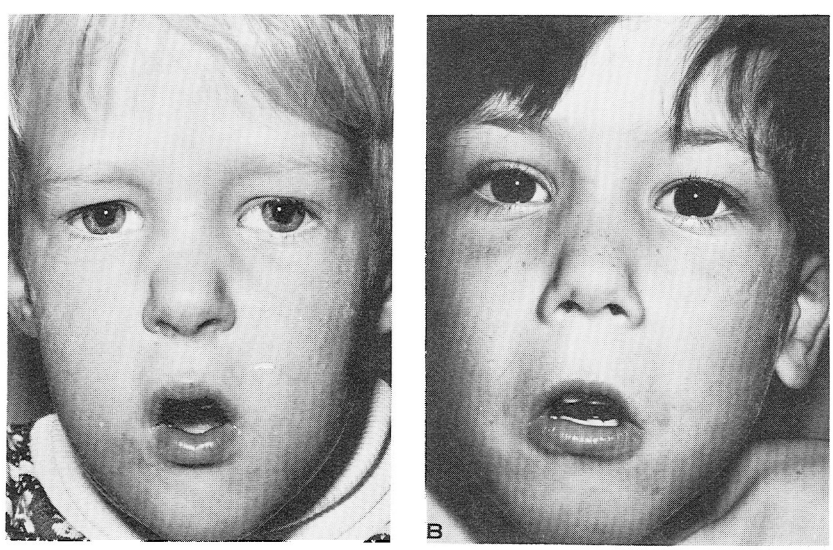


FIGURE 5. Drooping of the oral commissure results in a convex lip orifice instead of horizontal. A frown-like appearance exists probably due to facial muscle dysfunction.

to hypoactivity of the buccal maxillary facial muscles that elevate the commissures.

d) Dynamic facial muscle abnormality (Figure 6). We have already indicated that the lip abnormalities of vermilion contour and downward commissures are a result of muscle deficiencies or dysfunction and can be identified when the muscles are static. Two additional muscle dysfunctions can be identified only during facial animation: a bulge of muscle extending horizontally from the nasal ala across the nasolabial crease to the cheek (nasal grimace), and a bulge of muscle extending vertically from the nasal ala and floor to the lateral lip. This latter deformity is reminiscent of the labial muscle displacement seen in cleft lip. The orbicularis muscles do not form a complete ring but, rather, are divided superiorly at the site of the lip cleft (or in the case of submucous cleft palate) and vertically into the nose. This can only be noted during lip puckering.

Moreover, many of the patients have facial muscle hypoanimation or a dull, expressionless facies. Facial muscle hypoanimation occurs during periods of unawareness; the patient cannot smile normally (Figure 7).

e) External ear abnormality. The normal helical rim begins at the inset of the pinna in the temporal region and curves superiorly to form the dome of a C. A few patients with occult submucous cleft palate have a flat superior helix. It should also be noted that some of these children also had neck webbing. Because of the relative infrequency of these findings, we question whether they are a specific finding of submucous cleft palate syndrome.

f) Alveolar arch abnormality. A few of the patients also had a submucous cleft of the embryologic primary palate as well as the secondary palate. These patients have notches in the alveolar arch between the upper lateral incisor and canines.

2. CEPHALOMETRIC FEATURES. Standard lateral head x-rays were used to measure many parameters of facial skeletal configuration and growth. Of most significance were the hard palate length, soft palate length, and nasopharyngeal depth measurements. Also measured were the SNA and SNB angles.

These studies demonstrated that in nearly all cases (90%) hard palate length was less than the average for age but was within 1 standard deviation. Nasopharyngeal depth was usually greater than average (75%), but only 10% were more than 1 standard deviation above normal. The soft palate length was

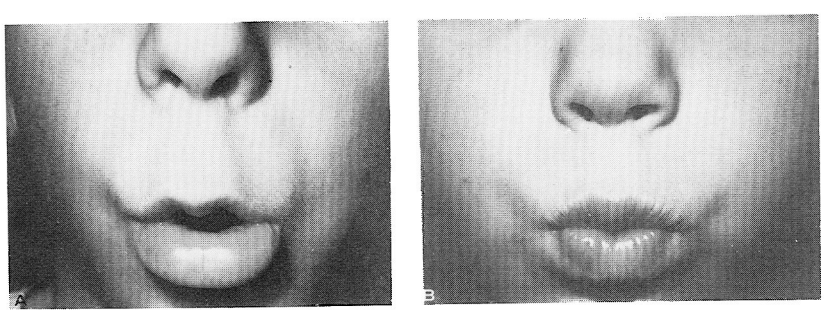


FIGURE 6. Some facial muscle abnormalities can only be identified during animation: lateral nasal bulge and vertical lip bulge.

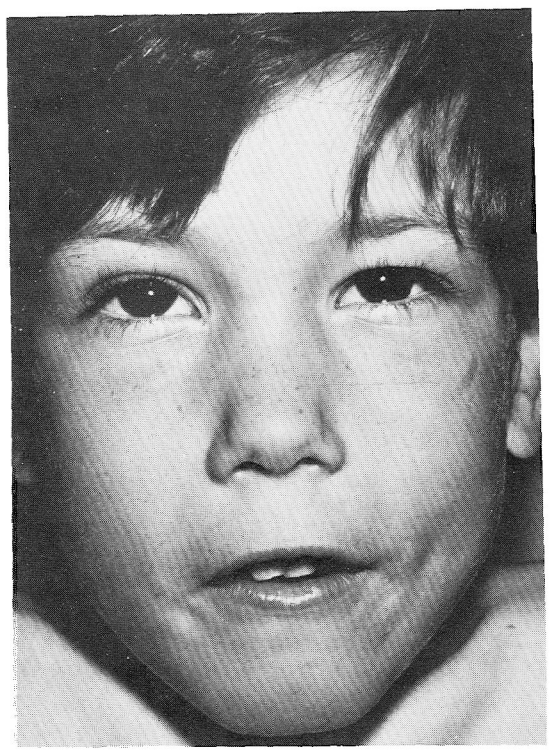


FIGURE 7. Paretic muscles prevent normal smile.

short by 2 standard deviations in 75% of the cases, and short by 1 standard in 90%.

The SNA and SNB angles were within 2 standard deviations of normal in 95% of the patients, but in every case the SNA-SNB difference was less than the average for age. Maxillary-mandibular occlusion was normal. These measurements showed a distinct but minimal tendency toward maxillary hypoplasia.

3. CINEFLUOROGRAPHIC FEATURES. Cinefluorography does not directly assist in making a diagnosis of occult submucous cleft, but rather confirms the speech pathologist's diagnosis of velopharyngeal incompetence and helps determine whether speech therapy or surgery should be the initial treatment. Furthermore, if surgery is required, the type of surgery is highly influenced by the cine evaluation of palate mobility and by the amount of velopharyngeal incompetence. This relation between cine study and choice of surgical procedure is described in another section.

Lateral cine voice studies showed marked variability. 20% of patients had normal palate mobility, "knee" formation, quickness and amplitude of motion, but still failed to achieve velopharyngeal closure. 40% of patients had normal palate function with apparent velopharyngeal closure during "model" speech (counting from one to ten, repeating specific words or phonemes), but showed velopharyngeal incompetence during spontaneous speech (when they had to describe a picture and were forced to focus on the subject matter rather than concentrate on the speech production). A fatigue pattern was not demonstrated.

40% of submucous cleft palate patients with velopharyngeal incompetence have consistent decreased palatal mobility and/or amplitude of motion on lateral cineradiography.

Cine voice studies revealed the tethering effect of abnormal levator insertions into the posterior border of the hard palate which we believe to be the key defect of submucous cleft palate. The levators contract isometrically against an immobile insertion and may appear to be paretic. We call this a "pseudoparesis." Most of our patients have obtained marked improvement in palatal motion after a surgical procedure that releases and repositions the abnormal levator insertions.

From his cine studies, Hoopes (*9*) concluded that the more anteriorly the levator was inserted the greater was the degree of velopharyngeal incompetence for a given depth of nasopharynx. Our experience confirms this impression. Weatherley-White (*16*) was able to define the relationship of soft palate mobility to speech performance using cineradiography. He found that in submucous cleft palate patients with "fast" palates normal speech occurred, while in patients with "slow" palates velopharyngeal incompetence was more common.

In summary, the diagnosis of occult submucous cleft palate can be suspected on the basis of any one of a constellation of oro-facial physical features, cephalometrics, and cinefluorographic voice studies. We have tried oral x-ray studies to visualize a notch in the hard palate and tried palate EMGs, but we have not found that either study can confirm or exclude the presence of levator muscle displacement due to occult submucous cleft palate. Facial and extremity EMGs are helpful to diagnose the patients with a suspected primary neuromuscular disease who present with palate paresis. Transillumination was not of value because muscle diastasis is rarely complete. Therefore, the definitive diagnosis is dependent upon intra-operative exploration of the palate.

Patient Management

The patients are seen by a multidisciplinary team of pediatricians, speech pathologists, orthodontists, oral surgeons, social workers, and plastic surgeons. Other specialists including neurologists, neurosurgeons, ophthalmologists and otolaryngologists are consulted for special problems. The objectives of the evaluation are listed below:

1) Confirm the presence of velopharyngeal incompetence by speech analysis and cinefluorography.

2) Grade the severity of speech dysfunction. We use a 5-point scale for acceptability, intelligibility, nasality, and articulation of specific phonemes (Figure 8).

3) Determine the degree of palate mobility. We use a 5-point scale with descriptive terms that characterize the severity of the velopharyngeal gap and normalcy of velar movement (Figure 9).

4) Make a presumptive diagnosis of the etiology and mechanisms of velopharyngeal incompetence.

Based on this evaluation, patients with a speech rating of 2—regardless of the anatomic situation or severity of cinefluorographic velopharyngeal incompe-

SPEECH RATING

	ARTICULATION PLOSIVE-FRICATIVE	OVERALL ACCEPTABILITY
1. NORMAL	NORMAL	NORMAL
2. MILD, PROBABLY NOT PERCEPTIBLE TO LAYMAN	0–10% INCORRECT	MILD, NOT SOCIALLY OR EDUCATIONALLY DISTRACTING
3. MODERATE, CONSISTENTLY PERCEPTIBLE TO LAYMAN	10–50% INCORRECT	MODERATE
4. MODERATE-SEVERE, 50% UNINTELLIGIBLE	50–75% INCORRECT	MODERATE-SEVERE UNACCEPTABILITY 50% UNINTELLIGIBLE
5. SEVERE, SPEECH GENERALLY UNINTELLIGIBLE	75–100% INCORRECT	SEVERE, GENERALLY UNINTELLIGIBLE

FIGURE 8. These are the key ratings by speech pathologists, although complete speech and language examination is also performed.

CINEFLUOROGRAPHIC PALATE MOBILITY RATING

1. BROAD CONTACT, NORMAL QUICKNESS, AMPLITUDE
2. TOUCH CLOSURE – INTERMITTANT INCOMPETENCE – NEAR NORMAL MOBILITY
3. NARROW OPEN – AMPLITUDE AND/OR QUICKNESS DIMINISHED
4. MODERATELY OPEN – SLOW MOVEMENT – NO "KNEE" CONFIGURATION
5. WIDE OPEN – PARETIC

FIGURE 9. Palate mobility is characterized by this scale. This cine evaluation is important in determining indications for surgery and type of surgical procedure.

tence—are recommended to continue speech therapy for at least 6 months and until age 6 or 7. Most of these patients improve with maturity and therapy (Figure 10), and because they do not have surgery we can only make a presumptive diagnosis of occult submucous cleft palate. Patients with ratings of 4 or 5 require surgical therapy, including diagnostic palate exploration. An exception is the child 2–4 years of age who has not had speech therapy and needs time to mature and who on cinefluorography shows near normal palate mobility (Grade 1, 2, or 3). Patients with a 3 rating are usually in the surgical category but there are individual considerations.

Surgical Therapy

Palate exploration (Figure 11).

1) A midline incision extends from the distal centimeter of the hard palate to the proximal centimeter of the soft palate.

2) The oral mucoperiosteum is lifted laterally with a periosteal elevator and the oral mucosa of the soft palate is dissected sharply with scissors. Extreme care must be taken to avoid cutting into the muscle or damaging the mucosa.

Findings (Figure 12)

The diagnosis of occult submucous cleft palate can be made by the appearance of palate bone and muscle insertions although there is a much greater variability in the proportion of levators that insert onto the hard palate. It is infrequent that all the muscles attach to the palatine bone. In most cases 75–90% of the muscle inserts on the bone, but some of the muscle meets in the midline (Type A). In a few cases (10%), nearly all the muscle meets at the midline, yet there is no palatine aponeurosis (Type C).

Approach

1) If the diagnosis of occult submucous cleft palate is not confirmed (i.e., muscle anatomy is normal), the following plan is generally followed:

a) Patients with normal palate mobility—retropharyngeal implant (*1, 4, 15*)

b) Patients with palate paresis—obturating pharyngeal flap (*5, 8*)

2) If there is an occult submucous cleft (levators insert onto the hard palate with or without a notch in the hard palate), we have reconstructed the palate by levator muscle reconstruction, palate pushback, and high superiorly based pharyngeal flap inserted into the raw surface of the nasal side of the palate (Fig-

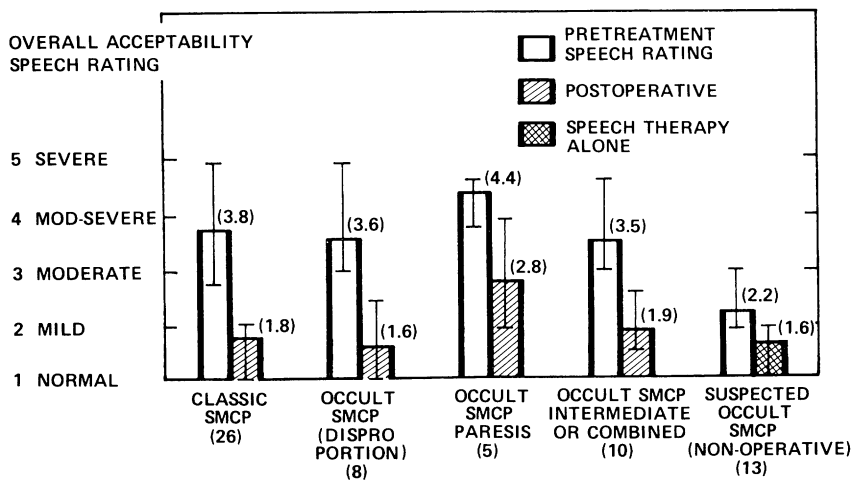


FIGURE 10. The results of treatment of classic and occult submucous cleft. Normal or very mild speech defectiveness was achieved in most cases. Patients with the worst initial speech ratings and palate paralysis had the least improvement. Speech therapy alone was beneficial for patients with minimal speech defectiveness and cinefluorographic palate mobility ratings of 1 or 2.

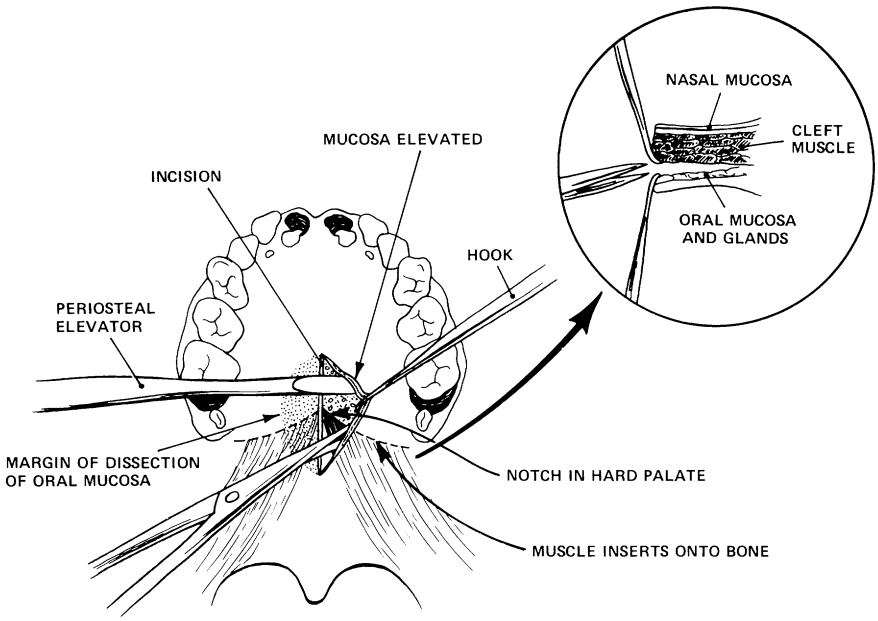


FIGURE 11. Palate exploration is through a midline incision to elevate the mucosa and observe the muscle and bony palate.

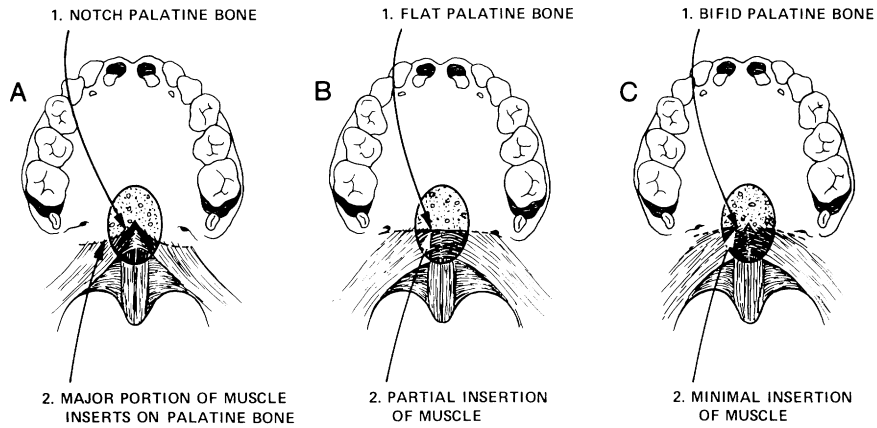


FIGURE 12. The findings at surgery are variable, but the typical forms of occult cleft with muscle insertion are shown. Type C and Type B may be "congenital short palate."

ure 13). Steps for this repair are:

a) oral mucoperiosteal V-Y pushback
b) transection of the attachment of the palate muscle and nasal mucosa
c) separation of the oral palate mucosa from the palate muscle
d) reconstruction of levator muscle sling
e) elevation of a high superiorly based pharyngeal flap
f) lining of the raw surface of the pushback with the pharyngeal flap.

The detailed methods of levator reconstruction for repair of clefts of the secondary palate (*10*) and the methods for palate pushback and superiorly based

pharyngeal flap have been described (*6*). We now recognize that some of the minimal forms of submucous palate clefts with near normal palate mobility can be treated with retropharyngeal implants. Our choice of surgical procedure was based on an attempt to evaluate a single method.

Results (Figure 10)

Twenty-six patients with classic submucous cleft palate and twenty-three patients with occult submucous cleft had the combined palate pushback, levator muscle reconstruction and pharyngeal flap surgery and a two-year postoperative follow-up. Thirteen patients with presumed occult submucous cleft palate or classic submucous cleft palate had speech therapy only.

1) Classic submucous cleft (26 cases)—the average pre-operative speech rating was 3.8 (moderate-severe) and postoperatively was 1.8 (mild-normal).

2) Occult submucous cleft with palatopharyngeal disproportion but normal palate function (8 cases)—pre-operative ratings were 3.6 and postoperatively were 1.6.

3) Occult submucous cleft with muscle dysfunction (5 cases)—the average pre-operative rating was 4.4 and 2.8 postoperatively.

4) Occult submucous cleft—Intermediate (10 cases)—3.5 rating pre-operatively and 1.9 postoperatively.

5) Occult cleft, Non-operative (13 cases)—initial ratings averaged 2.2 and post-speech therapy were 1.6.

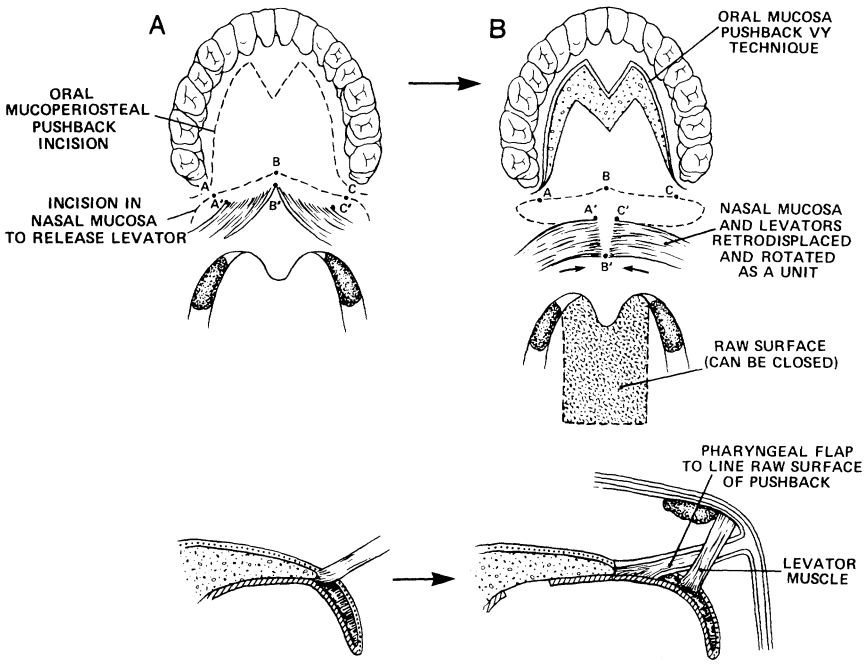


FIGURE 13. Our preferred method for levator muscle repositioning, palate pushback and superiorly based pharyngeal flap to line the palate pushback and prevent reattachment of palate muscles to the palatine bone.

Summary

We have studied 41 patients with classic submucous cleft and 32 cases with occult submucous cleft. Both groups have the same anatomic abnormality that leads to velar dysfunction—the insertion of the palate muscles onto the hard palate rather than onto the midline soft palate raphe. However, the occult submucous cleft palate does not have the classic triad of bifid uvula, hard palate bony notch, and furrow in the midline of the soft palate. Characteristic facial features, cephalometric x-rays, and cine voice studies can help make a presumptive diagnosis of occult submucous cleft palate. Surgical management includes a diagnostic palate exploration to identify muscle configuration followed by levator muscle sling reconstruction, palate pushback, and pharyngeal flap. Excellent speech results are obtained except with patients having palate paresis.

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