V. Hodgepodge

43. Congenital Velopharyngeal Incompetence and Submucous Cleft Palate

CONGENITAL velopharyngeal incompetence has as its most important manifestation *hypernasal speech without a cleft of the velum*. On examination, often to the surprise of the surgeon, there is at first sight a normal-looking palate with good mobility of the velum. More careful scrutiny reveals that in action the velum does not reach to the posterior pharyngeal wall, either because of a short palate or because of an abnormal backward position of the posterior pharyngeal wall. Of the many possible causes of velopharyngeal incompetence, one can be submucous cleft palate; however, not all submucous clefts have velopharyngeal incompetence.

Surgeons of the nineteenth century generally considered the submucous cleft of the hard palate the cardinal cause of open nasal speech. Some surgeons of that era, however, including Billroth, Passavant, von Langenbeck and Wolff, were convinced that late, spontaneous closure of palate clefts was frequent, without or after incomplete or unsuccessful surgical intervention. Robert Ivy himself reported such a case.

SUBMUCOUS CLEFT PALATE

In 1825 P. J. Roux of Paris first called attention to submucous cleft palate. In his *Mémoire* he recorded that, in 1823, he had been consulted by a young girl who nasalized so badly that her

speech was unintelligible. She had a cleft of the posterior portion of the velum, and there was a faulty union of the osseous tissue of the hard palate under an intact mucosa.

In 1846 Demarquay exhibited a dissection in which there was a cleft of the velum and a cleft of the bony palate which was filled with fibrous tissue and covered with intact mucosa.

Henri P. J. Winters of Utrecht University considers Gustav Passavant the rightful claimant of priority for describing congenital velopharyngeal incompetence. In 1862 Passavant reported a young female patient with a cleft lip and a spontaneously healed small cleft in the soft palate who three years later revealed the velum closed but a broad, deep submucous cleft of the hard palate and open nasal speech:

> She visibly could not bring the velum into contact with the posterior pharyngeal wall.

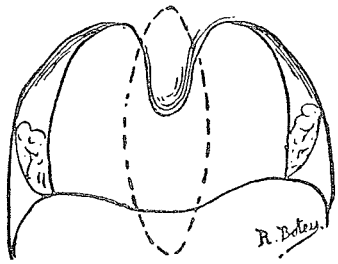
In 1865 Passavant reported a man with a submucous cleft of the hard palate, an intact velum, a small bifid uvula and heavy open nasal speech. Further evidence suggesting Passavant's genuine insight into the true physiology is given by his 1865 use of "*insufficienz*" in characterizing open nasality in speech after surgical closure of clefts of the soft palate.

In 1864 von Langenbeck described three types of bony clefts with the mucosa intact: (1) cleft of the velum with bony palate almost entirely absent, (2) cleft of the velum and hard palate in which the fissure in the bony palate was more extensive than in the soft tissues and (3) cleft of the velum associated with a fine split in the midline or on either side of the vomer, again with mucosa intact.

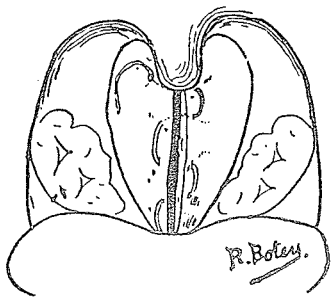
In 1869 Notta reported an 11-year-old girl with cleft uvula and bony cleft to incisive foramen with mucosa intact, who spoke with a distinct nasal twang. In 1870 Ulysse Trélat recognized the association of anteroposterior brevity of the bony palate and notching of the palate bones but attributed the nasal intonation to the anteroposterior shortening and lateral narrowing of the hard palate.

Lermoyez in 1892 defined "l'insuffisance vélo-palatine," or congenital velopharyngeal insufficiency, as a developmental disturbance with a healthy, mobile, normal-looking short velum

which was *always* associated with a submucous cleft of the posterior hard palate and a bifid uvula. Gutzmann in 1899 expressed doubts as to this invariable association. In 1893 Mears of Philadelphia was the first American to note this condition.

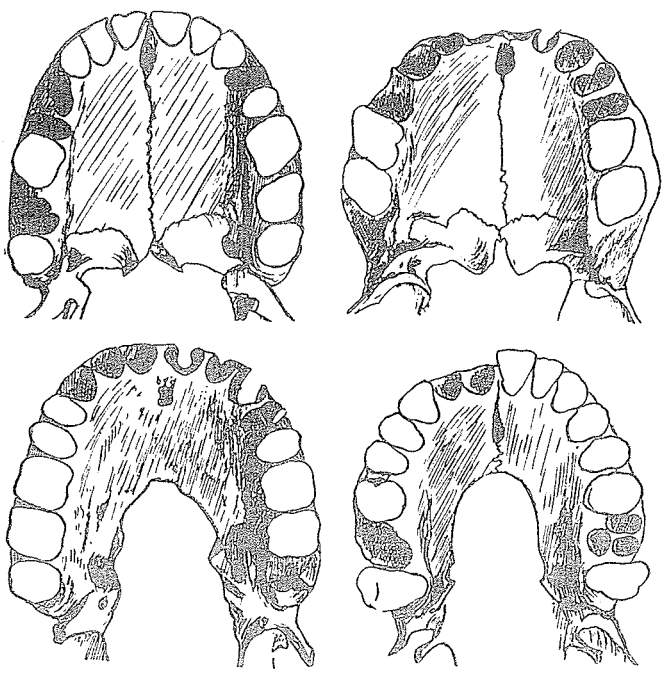


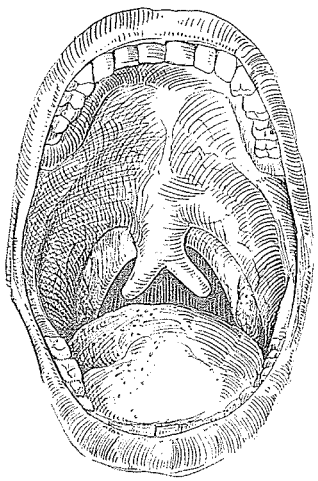
In 1907 Ricardo Botey of Barcelona devised an operation for correction of the deformity. It involved elliptical excision of one or two vertical sections of posterior pharyngeal wall and closure with a continuous mattress suture, as diagramed.



Adam Brown Kelly, chief of the Throat and Nose Clinic at Victoria Infirmary in Glasgow, Scotland, was described as being a combination of modesty, geniality and humanity who "always remained an unrepentant laryngologist." He reveled in research and was the first, in 1910, to coin the phrase *submucous cleft palate.* He made a very complete study of eight cases and 11 cases of what he described as muscular insuffiency of the palate, in which all 19 presented rhinolalia aperta. He also observed a further 18 submucous cleft patients with normal speech. Measuring and comparing the hard and soft palate and the nasopharyngeal opening of these and normal subjects, he decided that both the hard and soft palate were short in submucous clefts. Kelly published sketches to demonstrate the variety of missing portions of the posterior hard palate. He was also the first to acknowledge that there could be hypernasality in the absence of the stigmata of submucous cleft palate.

Adam Brown Kelly





In 1922 and 1923 Miloslav Seeman of Prague contributed several papers on submucous clefts. In 1927 Alexander Limberg of Leningrad introduced the terms *fissura ossea occulta* for submucous cleft of the hard palate and *fissura muscularis occulta* for submucous cleft of the soft palate.

In 1933 George Dorrance of Philadelphia, in his description of submucous cleft palate, remarked that the bony deficiency in the posterior part of the bony palate could vary from a large V-shaped defect to a mere notch to no loss of bone at all. Even with no bony defect, Dorrance pointed to the submucous cleft in the muscular tissue and in the palatal aponeurosis. He observed that the palatal mucosa was always intact but the velum was pulled forward with marked shortening in the anteroposterior diameter of the palate. He presented a sketch from Karl Peter showing a split uvula and he noted that the insertions of the levator palatini muscles were displaced forward and thus were unable to raise the velum upward and backward to the desired point of contact with Passavant's cushion for velopharyngeal closure.

In 1954 James Calnan, while at Oxford, wrote a learned treatise on submucous cleft palate. He noted the varying deficiency in the bone of the posterior edge of the hard palate, the mobile but markedly shortened velum and the bifid uvula. The absence of muscle union and of a median raphe down the midline of the velum presented a translucent zone seen in the mouth when a beam of light was flashed from above. During phonation, this area broadened owing to the pull of the tensor and levator muscles.

Calnan excised the submucous cleft in eight cases and reported

lack of muscle union across the cleft or poorly developed muscle fibres which lack orientation lying in a matrix of fibrous tissue. Mucous glands may present between the muscle fibres.

By cineradiography, as described by Ardran and Tuckey in 1951, Calnan studied the submucous cleft palate during speech and found that the velum failed to occlude the nasopharyngeal isthmus when it should, but its mobility and degree of elevation were not markedly impaired. The failure seemed to depend on the shortness of the velum, but when the posterior pharyngeal wall came forward, the velum moved upward and occluded the aper-

ture at a much lower level than normal.

Calnan was quite dogmatic about treatment:

In our experience there is only one treatment for submucous clefts with rhinolalia and that is surgical excision of the submucous portion of the cleft and V-Y retroposition of the soft palate on the lines described by Kilner.

If normal speech does not develop in three months, speech therapy is begun. Out of 17 cases, 15 achieved normal speech. Of the two remaining cases, one was successfully treated with a Hynes pharyngoplasty and in the other a Hynes procedure was planned. Calnan's stand on submucous cleft in infancy is of interest:

The correct time at which to treat patients with submucous cleft (as with obvious cleft palate) is at or about one year of age, before the development of speech. As has been mentioned, patients are not sent for treatment or diagnosis until much older. This is due to a failure of diagnosis which could be improved: all patients with cleft lip should receive a thorough examination of the palate. If a submucous cleft can be diagnosed with confidence, then the rational treatment is to excise the fibrous cleft and retropose the palate.

Six patients with this condition have been seen and treated in infancy in this Department. Four now have normal speech without the need for speech-training: the remaining two seem to be developing normal baby-talk.

William Porterfield

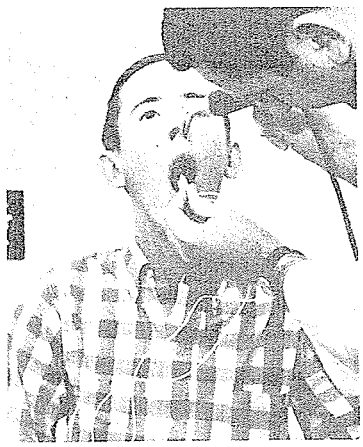
In 1965 H. William Porterfield and John C. Trabue of Columbus, Ohio, endorsed Calnan's classic triad of (1) bifid uvula, (2) midline soft palate muscle separation with intact mucosa and (3) midline notching in the posterior edge of the bony palate as prerequisites for diagnosis of submucous cleft palate. They reported 18 submucous cleft palate cases out 505 cleft palate patients and made recommendations as to what to do:

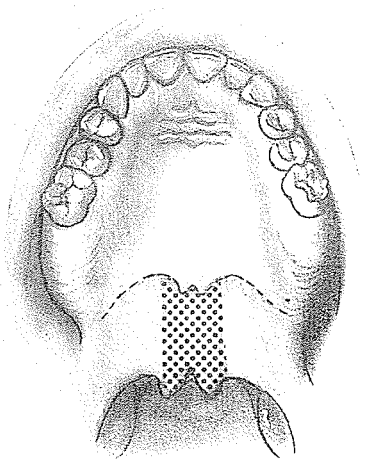
In view of our experience of a delay in seeing these cases and inferior speech results, we advocate the following: (1) Recognition of this defect in early infancy by the initial examining physician. (2) Surgical repair at 16 to 18 months by the pushback technique.

DIAGNOSTIC AIDS

In 1966 several publications dealt with submucous cleft palate, paying special attention to diagnosis and its guide to surgery.

Raymond Massengill





Previous techniques used for diagnosis of submucous cleft palate had ranged from Olin's palpation of the bony defect to Van Riper's use of radiography. Raymond Massengill, dedicated speech pathologist at Duke University, who raises roses and Tennessee walking horses, was stimulated in 1966 to devise a simple light instrument with an extension to be passed into the nostril and over the nasal side of the palate. The light penetration of the submucous cleft is studied by oral photometry with a photocell placed under the palate. Readings establish the extent of the submucous cleft. Variations of this overt diagnostic technique, presented in *Plastic and Reconstructive Surgery*, are used in many clinics today.

In 1967 Thomas Rees, D. Wood-Smith, C. Swinyard and J. Converse of New York University studied 12 submucous cleft palate cases with electromyography and diagramed the typical zone (dotted region) of absent to diminished electrical activity. They reported in *Plastic and Reconstructive Surgery*:

1. Electromyography is a useful diagnostic adjunct in the submucous cleft palate.
2. Electromyographic "mapping" of the muscle deficiency in the submucous cleft palate can serve as a guide to the surgeon in selecting the appropriate width of nonfunctioning soft palate to be excised.
3. The muscle deficiency present in the submucous cleft palate would seem to warrant the addition of muscle tissue as a component part of the repair. A pharyngeal flap helps to achieve this objective.

Indeed, their best speech results, they found, occurred in the two Veau-Wardill pushback operations with a superior pharyngeal flap added.

In 1979 Randall G. Michel, G. J. Baylin, A. S. Hall, I. H. Pipkin and W. R. Hudson of Duke University reported palatal tomography to be of benefit in diagnosis of occult hard palate defects. Although not advocating wide clinical use of this technique, they noted:

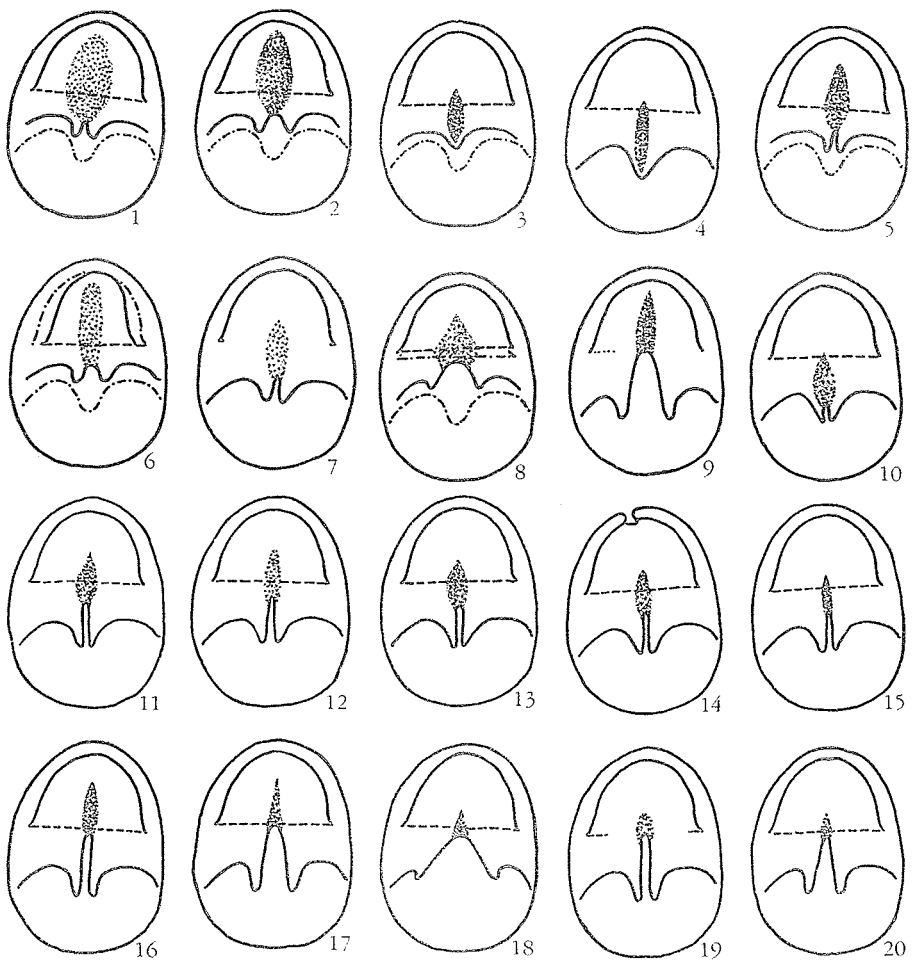
Nine of the 12 patients were found by tomography to have palatal defects that had not been detected either by cineradiography or by clinical investigation, including physical examination of the palate.

Defining SMCP

In 1970 George F. Crikelair, P. Striker and B. Cosman of the College of Physicians and Surgeons, Columbia University, New York, presented a provocative stand against limiting the diagnosis of submucous cleft palate to Calnan's triad. They stated:

> Actually, there is no such single grouping of findings that can be defined as a submucous cleft palate.
>
> As recognized by the classical authors, from von Langenbeck through Dorrance, there is a variety of submucous defects in which a submucous zone (either large or small) is present together with (or rarely, without) an actual cleft—which, in turn (when present), may be large or small. The submucous area may be in the hard palate, the soft palate or both; the palatal bony defect may vary from near total absence to apparent normality (without even a notching of the posterior margin). This wide spectrum of anatomical appearances is borne out in our experience; the only constant feature of the "submucous cleft palate" is the presence of a submucous zone.

They presented their stand diagramatically in *Plastic and Reconstructive Surgery*, with the dotted area being the submucous



defect, the dashed line the posterior edge of the hard palate, and the dashed and dotted line the normal outline of the alveolar ridge or posterior edge of the hard palate or the posterior edge of the velum.

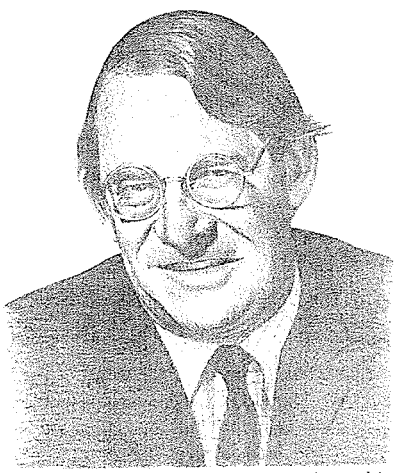
Crikelair's team, with a surgical experience of 20 significant submucous cleft palate cases, opposed the accepted dogma of excision of the submucous zone and palate pushback.

When the submucous area is large its excision may leave little tissue for palate closure of any kind—and predispose to breakdown and fistula formation. On the other hand, it is clear that significant submucous defects are not always associated with short soft palates and/or short hard palates. Consequently the insistence on a push-back procedure in all cases is hard to justify.

Our experience demonstrates that good speech results may be achieved without excision of the submucous section of the defect and/or without a push-back procedure. Where the palate does appear to be short, a primary pharyngeal flap may be used (without submucous zone excision). Where the submucous portion is smaller, narrower, and in the soft palate, and where the palate segments are ample, excision and a simple von Langenbeck closure can give good results. . . . It is thus possible to approach therapy in each instance of submucous defect freely.

a good point

In 1969, 1970 and 1974 John E. Hoopes and others of Johns Hopkins University confirmed by cineradiography the early finding of Dorrance that the anatomical defect in submucous cleft palate was the insertion of the levator palatini muscles too far forward to elevate the soft palate effectively. He defined the factors responsible for velopharyngeal incompetence in submucous cleft palate as (1) a short soft palate and (2) an anteriorly displaced levator insertion. The more anterior the levator insertion, the greater the velopharyngeal incompetence.



Chris Weatherley-White

Asymptomatic SMCP

Another surgeon interested in submucous cleft palate is R. C. A. Weatherley-White of Denver, who seems to spend most of his free time in the air. Born in India into a third-generation Indian Medical Service family, he served as a paratrooper in the 82nd Airborne Division, was once United States National Inter-Collegiate Sky Diving Champion and is now learning to fly aerobatically. His first contact with plastic surgery was through Sir Archibald

McIndoe's visit to the R.A.F. Hospital at Holton, and he later trained with Richard Stark in New York.

In 1972 Chris Weatherley-White, C. Sakura, L. Brenner, J. Stewart and J. Ott of the University of Colorado found nine submucous cleft palate cases in a study of 10,836 Colorado school children in the Denver area. This presented an incidence of 1:1,200, but only one child had mildly abnormal speech and it was corrected by therapy alone. In these and 52 other submucous cleft palate patients referred, the combination of a relatively short palate with lessened mobility and demonstrable easy fatigue with effort was noted. Speech proficiency was not related to the degree of muscular clefting. Only four patients had surgery, and this involved the complete excision of the diastasis ("zona pellucida") combined with pharyngeal flaps in all, plus Veau-Wardill push-back in two. Hypernasality was corrected in three and improved in one.

Weatherley-White wrote in 1976:

not all submucous clefts require surgery

In our epidemiological studies on submucous cleft palate, I am constantly reinforced in our initial premise by the fact that I see so many asymptomatic submucous cleft palates. My aphorism concerning this lesion would be *to follow all children with submucous cleft palate very carefully with frequent routine speech evaluations.* Surgery should be done if there are signs of velopharyngeal incompetence; this should obviously be done as soon as the diagnosis is made, to prevent neurological pathways becoming stratified and causing a persistence of the speech problem.

In 1973 Raymond Massengill, K. Pickrell and M. Robinson of Duke University reported on a comparison of 12 submucous cleft palate patients with a random group of 12 postoperative cleft palate patients. The groups were matched according to age and sex, and all had some type of pushback procedure with what was presumed to be adequate length—either a Veau, a Dorrance, a Dorrance with an island flap, a Wardill or a Wardill with an island flap. In their small series, the submucous cleft palate patients had a higher percentage of velopharyngeal incompetence than the controls. This was partially explained by the later diagnosis of the patients, after they presented speech problems, and resultant greater palatal height in the control than in the submucous cleft palate groups.

In 1970 John Hoopes with A. Dellon, J. Fabrikant and A. Soliman of Johns Hopkins University suggested:

The procedure of an island flap pushback alone has proved disappointing in the management of submucous cleft palate, for which reason the combined procedure [island flap and pharyngeal flap] is utilised. The difference in the type of closure obtained with an island flap pushback *versus* a pharyngeal flap is essentially the difference between active and passive closure, respectively.

They reasoned that, in addition to the increase in active closure achieved with the posterior displacement of the levator muscles by the island, extra passive aid was given by the tethering of the pharyngeal flap.

In 1974 N. Culf, J. Chong and L. Cramer of Temple University stated that one of the ideal candidates for the double sandwich island flaps was the submucous cleft palate patient and reported 10 cases so treated. They wrote:

In cases of submucous cleft with significant separation of the levator muscles, the muscle bundles are easily approximated through the transverse incision at the junction of the hard and soft palate.

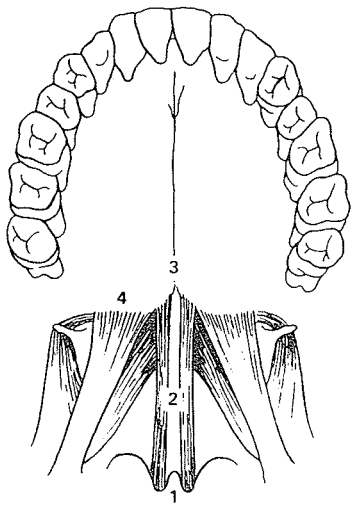
F. L. F. Innes of Norwich, England, trained by Kilner and Peet and influenced by the work of Calnan, is convinced that early cleft palate closure is ideal and is concerned that submucous clefts escape detection. He wrote in 1976:

Most of the patients do not reach me until a year or two after they have gone to school. This condition is still too often diagnosed much too late with the result that the unfortunate child is greatly handicapped.

The educational and psychological problems of late diagnosis are by themselves bad enough but there are in addition physical problems. The soft palate elements in such patients are very poor and underdeveloped. The standard Kilner-Wardill operation is probably adequate for children with submucous clefts who are presented for operation at about the age of one year, but if there is any doubt about the result of this operation by itself, an island flap . . . can be easily introduced. For older patients, the Kilner-Wardill operation is not adequate. Something more is required because in such patients the pharynx will have become excessively large. The pharynx seems to grow too wide and too deep without the influence of proper palatal muscle action in the front of the palatopharyngeal isthmus.

In these cases Innes advocated the combination of a pushback procedure aided by an island flap and complemented with reduction of the pharynx by the Hynes pharyngoplasty.

In 1975 in the *Cleft Palate Journal* E. N. Kaplan of Stanford University noted that the classic submucous cleft palate with Calnan's triad represents the obvious overt physical signs of an underlying anatomical abnormality—the insertion of the levator and other palate muscles onto the hard palate instead of forming a sling across the midline. He stated:



> 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. . . . However, we would exclude cleft palate associated with craniostenosis . . . branchial arch syndromes . . . mandibular micrognathia . . . and cleft palate with cleft lip . . . because they are probably embryologically distinct conditions.

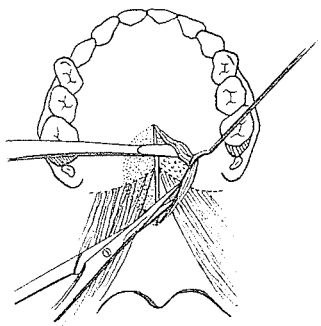
Kaplan reviewed 250 cases of velopharyngeal incompetence without cleft lip or cleft palate and identified 41 cases of classic submucous cleft and 23 cases of occult submucous cleft. Here are his diagnostic aids:

1. Facial features suggestive of occult or classic SMCP:
   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%).
2. Cephalometric studies revealed that in 90% of patients, the hard palate length was less than average but within one standard deviation of normal; in 75% nasopharyngeal depth was greater than average and only 10% fell more than one

standard deviation away from normal, but in 90% the soft palate was short by one standard deviation and 75% were short by two standard deviations.

3. Cinefluorographic studies were recommended to confirm velopharyngeal incompetence and to help determine need for therapy or surgery.

Kaplan noted:



Ultimately, however, the *definitive diagnosis is dependent upon the intra-operative exploration of the soft palate muscles.*

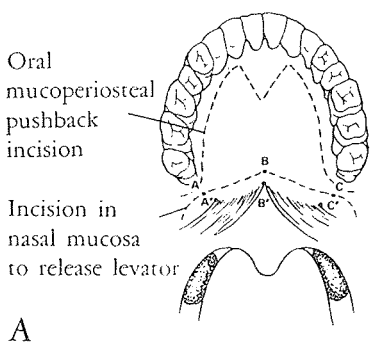
He then elaborated:

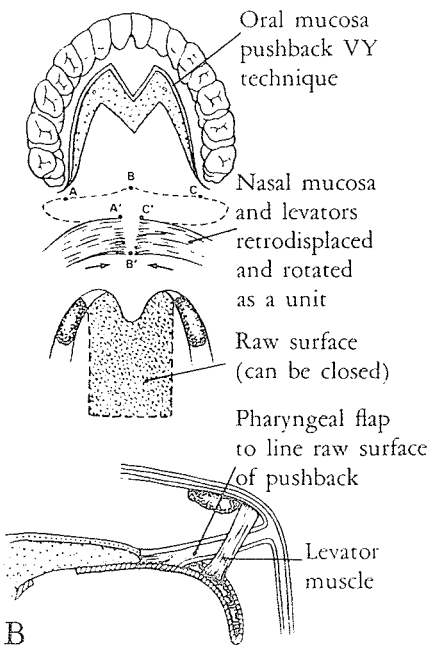
1. A midline incision extends from the distal centimeter of the hard palate to the proximal centimeter of the soft palate.
2. The oral mucoperiosteum is lifted laterally with a periosteal elevator and the oral mucosa of the soft palate is dissected sharply with scissors. Extreme care must be taken to avoid cutting into the muscle or damaging the mucosa.

He admitted to great variation of levator muscle insertion but noted that 75 to 90 percent of the levator muscles inserted on the bone with some muscle meeting in the midline.

Kaplan outlined his surgical 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.
   b) Patients with palate paresis—obturating pharyngeal flap.





2) If there is an occult submucous cleft . . . we have reconstructed the palate by levator muscle reconstruction [A], palate pushback, and high superiorly based pharyngeal flap inserted into the raw surface of the nasal side of the palate [B].

This is the method described in 1965 by Dibbell, Laub, Jobe and Chase, applied to occult submucous clefts by Kaplan with the addition of reconstruction of the levator muscle sling.

In 1975 Minami, Kaplan, Wu and Jobe of Stanford University reported finding that 44 percent of submucous cleft palate patients with velopharyngeal incompetence had decreased palatal mobility on lateral cineradiography. They also pointed to the tethering effect of the abnormal levator insertions into the posterior hard palate as the key defect in SMCP. As they wrote:

The levators contract isometrically against an immobile insertion; thus, they may appear to be paretic.

Roland Minami

They also reported four patients with none of the classic findings of SMCP who, after palatal exploration, revealed in each case a very small bony notch, unmistakable abnormal levator palatini insertions into the posterior border of the hard palate and diminished palatal mobility.

Treatment: As pointed out by Weatherley-White, normal speech will develop without any treatment in about 90 percent of SMCP patients, making early treatment inadvisable. For cases that do come to surgery, the Stanford group advocates, as standard procedure, *pushback* to correct the short palate and release the abnormal levator attachments and *superiorly based pharyngeal flap* to line the raw area and hold the backward displacement with a partial obturation of the velopharyngeal gap. To this they have added *reconstruction of the levator sling.*

In 1976 Frank E. Abyhǫlm of Oslo, Norway, reported 47 submucous cleft palate patients, operated on at the average age of 10.8 years during 1965 to 1974. Eleven had had tonsillectomy and/or adenoidectomy prior to diagnosis. He noted better results when the patient was operated on under 7 years of age and from his experience advocated von Langenbeck or pushback palate closure with levator muscle sling construction and a superiorly based pharyngeal flap.

Meanwhile, Porterfield had continued his interest in the palate

and submucous clefts. In 1976 he recalled the problems faced 20 years ago and felt encouraged by the improvements that have followed:

> My venerable old Chief, Harold Trusler of Indianapolis, had a reputation for being a gruff, tough old fellow, but who beneath the surface, was a very gentle and kind man. He would look at a previously operated palate that demonstrated much scar and immobility, turn away and say to the gathered audience, "that surgeon just had a bad pair of hands." To me, this merely points out what has been accomplished in these years.

The Calnan Controversy

Eleven years after his first paper, Porterfield, with Mohler and Sandel in 1976, still held rigidly to Calnan's triad as requirements for a case to be admitted to the inner sanctum of "submucous cleft palate." In a direct attack:

> We would take issue with the statements of Crikelair *et al* in which they described submucous defects in association with overt cleft palate deformities and designated that deformity also as a SMCP. We feel the strict criteria of Calnan are valid ones for delineating the submucous cleft palate problem.

Meanwhile, Porterfield became aware that a pushback at 16 to 18 months was *not* sufficient treatment of SMCP. He began backtracking with an "ink had hardly dried" revision, suggesting in 1976 that a primary pharyngeal flap, or a superiorly based pharyngeal flap combined with a von Langenbeck palatoplasty if the palate seemed short, would be a more efficacious procedure.

This, of course, excited a 1977 Letter to the Editor from Crikelair and Cosman to Porterfield in reference to submucous cleft palate diagnosis and treatment. Excerpts from the letter follow:

> It is a simple fact that submucous defects can occur in the hard palate without any involvement of the soft palate. It has also been observed that extensive submucous defects in the hard and soft palate may coexist with clefts of the soft palate considerably larger than a mere bifid uvula. Such cases have been amply documented [Crikelair et al., Roux, Demarquay, Trélat, and Veau]. What name could one give to these defects, if not "submucous cleft palate?"
>
> The restrictive criteria of Calnan serve to perpetuate the view that submucous cleft palate is a wholly different entity from cleft palate, rather than one of its manifestations. This concept has led Calnan, Porterfield, and

others to the too severe stricture concerning the necessity for excision of the submucous portion of the defect, a view which Dr. Porterfield and his colleagues now agree is in error.

It would have been generous of Dr. Porterfield *et al* to have pointed this out—and to have indicated, as they now also show, good speech may be achieved by the simple addition of a pharyngeal flap to the submucous cleft palate, without any other manipulation.

Porterfield's 1977 reply to the Crikelair-Cosman letter ended:

My principal reason for disagreeing with them . . . is, however, simply that of being a "purist." I think that the original description of Calnan should be retained, that it should not be confused by extraneous modifications.

What's so sacrosanct about Calnan's three criteria?

To throw another dimension into submucous cleft palate, M. Fára of Prague stated in 1977:

All our 105 patients with submucous cleft manifested medial cleft palate with vomer in the central line. . . . The wider the cleft in the palatal plates, the greater the parting of the velar muscles and the substituting attachments of these muscles to the palatal plates are functionally less valuable. Every fourth patient with submucous cleft was afflicted with the syndrome of developmental shortening of the palate: special physiognomy with striking hypomimia and decreased intellect.

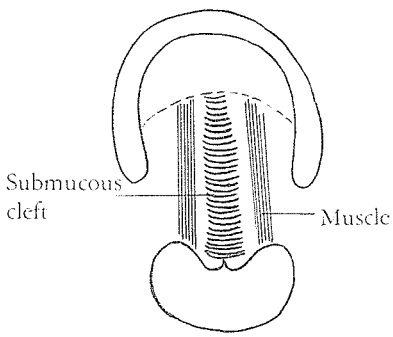
A SIMPLER APPROACH

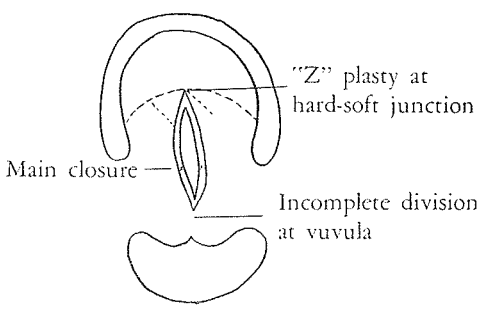
Dennis Walker of Johannesburg, who devised a different approach to the various submucous cleft palate problems, remembered a visit to the theater at 149 Harley Street when Gillies, in a mischievous mood, called through to Sir Archibald McIndoe in the next room, to ask whether Walker might watch him work:

McIndoe said, "yes, of course," and then the wicked old maestro engaged my attention so that I was forced to stay with him, until McIndoe looked at me *sideways* when I eventually went to watch him.

Gillies' gamesmanship

With a touch of Gillies' flexibility, Walker designed his attack on what he considered the specific problems of the submucous cleft, leaving the uvula, when not bifid, intact.





CONGENITAL HARD PALATE HOLES

Defects affecting only the hard palate are among the rarest found in clefts. They appear as oval holes in the midline, not usually

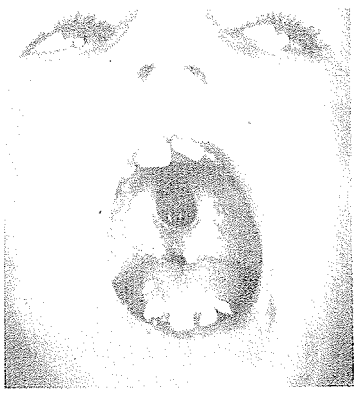
extending the whole length of the palate plates. Their occurrence is usually thought to be connected with a submucous cleft. Evidently there are exceptions.

In 1966 J. B. Lynch, S. R. Lewis and T. G. Blocker of the University of Texas, Galveston, reported a case of a Caucasian male with a moderately wide cleft of the hard palate extending from the incisive foramen to the junction of the hard and soft palates and an unattached, underdeveloped vomer. The alveolus was intact and the soft palate normal with microscopic sections of its midline revealing

> muscle fibers extending to the midline, which indicated that the muscular portion of the soft palate was, in fact, normally developed and did not represent a submucous cleft.

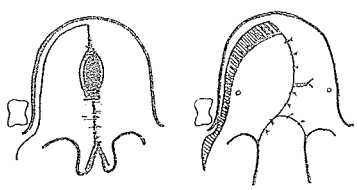
This defect, they felt, was not explained by any modern concepts of embryology.

It is more common, however, for this hard palate defect to accompany a submucous cleft palate. Both Trélat and Veau suggested that the phenomenon was a prenatal rupture of a submucous cleft, either spontaneous or artificial, and Fára endorsed their stand, estimating the occurrence of the opening at the time when the intrauterine growth of the head was reaching completion. In 1954 Calnan presented one of these cases with a hard palate hole accompanying a submucous cleft palate.



In 1971 in *Plastic and Reconstructive Surgery* Miroslav Fára of Charles University, Prague, reported five cases of congenital defects in the hard palate associated with typical complete submucous clefts and, in addition, slightly underdeveloped maxillae and marked hypoplasia of the palate plates.

TREATMENT



One of the cases reported by Fára was treated by F. Burian in 1935 with a mucoperiosteal rotation flap and inferiorly based pharyngeal flap.

Fára himself used a more refined approach. Excision of the midline submucous cleft area revealed the following:

On the sides of the excision, numerous cross-sectioned muscle fibers are seen; toward the midline they become rare, with an oblique or longitudinal change in direction. In the midline, however, the muscle fibers are completely absent and one sees only dense connective tissue.

Fára emphasized the importance of

proper retropositioning of the palate, after detaching the muscle insertions from the posterior margins of the hypoplastic palatal plates, and suturing the muscles in the midline with a primary pharyngofixation [using a superiorly based flap.]

PATHOGENESIS OF SUBMUCOUS CLEFT

Dynamic David Poswillo of the Royal College of Surgeons of England Research Establishment in Downe, Kent, in 1974 reported an intriguing study of exogenous factors in the etiology and pathogenesis of complete and submucous cleft palate. A series of pregnant mice were given phenytoin in pediatric suspension at the rate of 150 mg per kilogram by gastric intubation from day 12 to day 16 of pregnancy. Examination of 100 consecutive fetuses at day 18.5 revealed 16 with complete cleft palate, 15 with submucous cleft palate and 69 normal. Serial study of the mouse fetuses in which SMCP had been induced supported the hypothesis of interference with mesodermal differentiation. A centripetal gradient of differentiation in the palatal shelf was described, commencing at the nasal foramen and extending to the uvula. When this gradient was disturbed by teratogens, after fusion of the palate, either SMCP plus bifid uvula, or bifid uvula alone resulted, the anomaly being determined by the stage of onset in relation to the anteroposterior gradient risk.

Study of the animal defect helped identify the causal mechanism in man. The findings supported the proposal that the teratogenosensitive period of palatogenesis in man should be regarded as extending from early embryogenesis to about the twelfth week of development. Until that time, agents can act to interfere with the developing palatal plates and the velar mesoderm in such a way that SMCP and bifid uvula, microforms of cleft palate, could result.

OTHER CAUSES OF VELOPHARYNGEAL INCOMPETENCE

In 1933, in his remarkable book *The Operative Story of Cleft Palate,* George Dorrance of Philadelphia discussed congenital insufficiency of the palate and categorized six varieties:

1. Normal appearance but inability of velum to approximate the pharyngeal wall because of anteroposterior shortening of the hard palate and velum.
2. Velum normal but hard palate short.
3. Hard palate normal but velum short.
4. Hard palate normal but submucous cleft of the velum.
5. Velum normal in appearance but with submucous cleft extending into the hard palate.
6. Palate insufficiency after successful cleft closure with the velum too short to reach the posterior pharyngeal wall.

Dorrance noted that children with congenital shortening of the palate usually learn to speak later than normal children, or speak indistinctly. Speech deficiency improves with time but is often associated with compensatory mechanisms such as development of compressor naris muscles and hypertrophy of the faucial and pharyngeal tonsils. His dissertation on diagnosis by symptoms was impressive.

1. Rhinolalia aperta or open nasalizing where vowels take a nasal tone ("ah" becomes "an"), consonants become altered with exception of "M" and "N" and sigmatismus, or the inability to produce the letter sound "S."
2. Shortness of breath in speaking due to air loss through the nose.
3. Inability to whistle.
4. Inability to hiss.
5. Inability to blow out a candle flame.
6. Mouth breathing becomes a habit presenting a vacant expression.
7. Pronounced facial movements with such muscles as the nasal compressor and corrigators to turn the patient into a "face-talker."
8. Diminished hearing due to improper ventilation of the middle ear.
9. Disturbed deglutition.
10. Fast talk avoiding difficult words.
11. Intra-oral examination reveals velopharyngeal insufficiency due to overall shortness of the palate and possible weakness in the superior pharyngeal constrictors.
12. Intranasal examination reveals insufficient velopharyngeal closure.

13. Bifid uvula in some cases.

14. Palpation through normal mucosa reveals various examples of submucous cleft in the muscles of the soft palate and the bone of the hard palate.

15. Irregularity and crowding of teeth, congenital absence of maxillary incisor tooth or the association with cleft lip.

In reference to nasal escape Dorrance stated:

The most reliable test is to hold a piece of cotton in front of the anterior nares while the patient makes efforts to pronounce non-nasal letter sounds. The escaping air makes the cotton move when this mechanism is insufficient.

DIFFERENTIAL DIAGNOSIS

Dorrance warned that congenital insufficiency of the palate should not be confused with palsy of the velum, "stomatolalia" and speech defects due to loss of teeth or faulty lingual articulation. Palsy of the palate is recognizable by the absence of the palatal reflex, inability of the velum to move and its lack of response to faradic stimulation. Stomatolalia or rhinolalia clausa is a condition of speech in which the letter sounds lack their nasal resonance. Dorrance added that speech defects due to loss of teeth are correctable by dentures and, if due to faulty tongue habits, are benefited by methodical speech training. He recommended his "push-back" operation of the palate as the best treatment of congenital insufficiency.

In 1954, while still at Oxford University James Calnan outlined, in addition to submucous cleft palate, other causes of velopharyngeal insufficiency:

1. Congenital short palate.
2. Cerebral agenesis of the supranuculear bulbar origin.
3. Paralysis of the palate (infectious or viral, which usually clears after recovery from the infection).
4. "Tonsillectomy palate" in which the surgeon has taken a portion of the soft palate along with the tonsils.
5. Functional rhinolalia where the patient "talks down the nose" for no apparent reason.
6. Rhinolalia following adenoidectomy.

Great interest has been shown in the condition of velopharyngeal incompetence following adenoidectomy.

In 1958 A. G. Gibb published his findings on a series of 19 patients seen by him in Scotland with permanent nasal escape following removal of tonsils and adenoids. He found that there had been 27,734 operations for removal of tonsils and adenoids in a population of 62,000 from 1950 to 1957. The incidence of permanent speech defect was 1 in every 1,459. Yet Calnan reviewed the Scottish series and indicated the probability that five were submucous cleft palate patients and four were mentally retarded with possible congenitally large pharynx. Gibb, he noted, felt that the soft palate was short and considered this factor the cause of nasality after adenoidectomy.

In 1971 James Calnan of the Royal Postgraduate Medical School, London, reported that between 1951 and 1968 he had a series of 19 patients with permanent nasal escape during speech following removal of tonsils and adenoids. All had had normal speech prior to the T & A and were above average intelligence. Radiological studies demonstrated a fully mobile soft palate with a gap between it and the posterior pharyngeal wall. When the gap was occluded by a cartilage implant behind the posterior wall of the pharynx, speech returned to normal. Cephalometric measurements suggested that the essential defect was a pharynx deeper than normal. He stated:

> A good analogy would be a size 7 foot in a size 8 shoe: both are "normal" but of little use together.

Calnan acknowledged that the disproportion between palate and pharynx was probably not possible to diagnose before adenoidectomy.

In 1975 Roland Minami, Ernest Kaplan, George Wu and Richard Jobe of Stanford University reported on 23 patients with hypernasality following removal of adenoids and tonsils. In most cases, they noted, velopharyngeal incompetence after a T & A is transient and disappears in a few weeks after compensation for the missing adenoid prominence. A marginally adequate mechanism may not be able to cope and will "unmask" the presence of

a submucous cleft or other pathology. Surgical experience with the standard Stanford pushback with superiorly based pharyngeal flap achieved improvement of severe to moderate hypernasality to minimal with the best prognosis in patients with normal amplitude and quickness of palatal motion. Nine patients had no surgery and, out of six of these followed three years, two showed improvement.

Noting the extensive reports of good results in such cases with retropharyngeal augmentation, Minami and his colleagues admitted favoring this approach "with a velopharyngeal gap of 5 mm. or less." When there was preexisting hypernasality worsened by T & A, surgery, they felt, should be directed toward correction of the underlying cause.

PROPHYLAXIS

It is important that children with congenital palatal incompetence who have only minimal hypernasality not be subjected to a standard adenoidectomy. Subtelny and Koepp-Baker advised that when adenoidectomy was absolutely indicated because the adenoid tissue covered the orifices of the Eustachian tubes, a lateral band adenoidectomy could be done, leaving the bulk of adenoid tissue in the midportion of the pharynx undisturbed.

CONGENITAL LARGE PHARYNX

By 1971 James Calnan of London had added *congenital large pharynx* to his list of six groups besides submucous cleft palate which can result in velopharyngeal incompetence. He reported a personal series of 41 patients seen over an 18-year period presenting nasal escape with apparently normal palatopharyngeal mechanisms. Extensive clinical and cephalometric studies revealed the pharynx to be larger than normal, justifying the term *congenital large pharynx.*

Calnan found no improvement with the Wardill-Kilner V-Y procedure, lack of nasal resonance with the Hynes procedure, extrusion of Teflon implants and the best results with autogenous costal cartilage implanted in the retropharyngeal area above the

arch of the atlas. His overall result was 60 percent normal speech obtained after the various types of surgery.

In 1975 Minami, Kaplan, Wu and Jobe of Stanford University divided velopharyngeal incompetence without overt cleft palate into the "Big Four":

1. Acquired palatopharyngeal disproportion after removal of adenoids and tonsils.
2. Abnormal anatomy of levator palati muscles—submucous cleft palate.
3. Palatal paresis.
4. Other causes—mental retardation, congenitally short palate, congenitally large pharynx and possible "occult" SMCP.

In 1979 W. S. Hagstron, R. W. Parsons, S. J. F. Landa and M. C. Robson of the University of Chicago, reported two cases of familial velopharyngeal incompetence caused by myasthenia gravis. They suggested consideration of: "myasthenia gravis (electromyography or testing with edrophonium) when the cause of neuromuscular dysfunction cannot be definitely established."

PALATAL PARESIS

The discussion of the treatment of palatal paresis by Minami et al. is of special interest. They noted that surgery has included pharyngeal flaps, citing Randall, Bakes and Kennedy (1960), J. C. Hardy et al. (1961), and Crikelair, Kastein and Cosman (1970); unilateral pharyngeal flap limited to affected side in unilateral palatal paresis, suggested by Broadbent and Swinyard (1959); and temporalis muscle and fascial sling reported by Kiehn et al. (1965). The obvious objective in a paralyzed palate is the construction of an almost complete obturator. If this can be accomplished by Hogan's wide, lined pharyngeal flap closing off the lateral ports to 3 mm., then that is the method of choice. Palatal lifts and obturating types of prosthesis have been used successfully and may be the treatment of choice, at least in patients with cerebral palsy, as noted by Gibbons and Bloomer (1958), Lang and Kipfmueller (1969), Gonzales and Aronson (1970) and Hardy et al. (1969).

In all types and degrees of palatal paralysis resulting in velopharyngeal incompetence, the pharyngeal flap, by reduction,

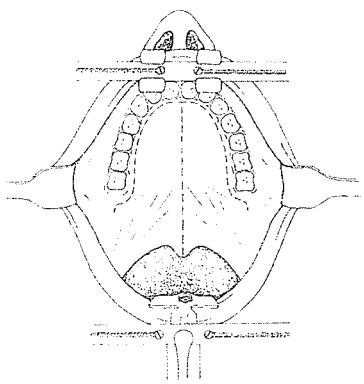
constriction and obturation of the velopharyngeal aperture, has earned an important place in the surgical treatment. Its contribution is great even to such rare conditions as congenital suprabulbar paresis described in 1950 by Worster-Drought, in which the motor outflow to the tongue, lips, palate, pharynx and larynx is affected. In the mildest form of this syndrome, the soft palate suffers most with a marked rhinolalia. In 1958 Wynn Williams of Nottingham, using the Rosenthal pharyngeal flap, reported good results on 49 percent of the patients.

At the 1973 International Congress on Cleft Palate in Copenhagen, C. H. Waar of the University Hospital Dijkzigt, Rotterdam, advocated surgical fixation of the uvula to the lower adenoid region for speech improvement in paralysis of the soft palate. The chance of improvement is far better if, during phonation, a constriction of the pharyngeal musculature is noted. Waar cited two successful cases, a 5-year-old girl with bilateral paralysis of the glossopharyngeal nerve but no other neurological symptoms, and a 30-year-old male with dysarthria, hemiplegia and open nasality as a result of a traffic accident. After long, unsuccessful speech therapy, a uvula fixation was followed shortly by a definite reduction in nasality.

In 1977 at the Third International Cleft Palate Congress in Toronto, Donnell F. Johns, Ph.D., and Kenneth E. Salyer of the University of Texas Southwestern Medical School, Dallas, determined the width of their obliterating superiorly based pharyngeal flap by observing and marking the most medial excursion of the lateral pharyngeal walls. Using this width flap, they were able to report 14 successes out of 15 cases of neurogenic velopharyngeal incompetence.

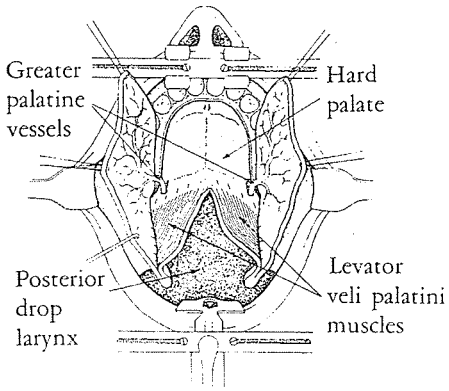
Jack Fisher

In 1975 alert Jack C. Fisher of the University Hospital, San Diego, and M. Edgerton of the University of Virginia Hospital, Charlottesville, reported the combined use of the levator retrodisplacement and pharyngeal flap for congenital palate insufficiency. They presented their argument:

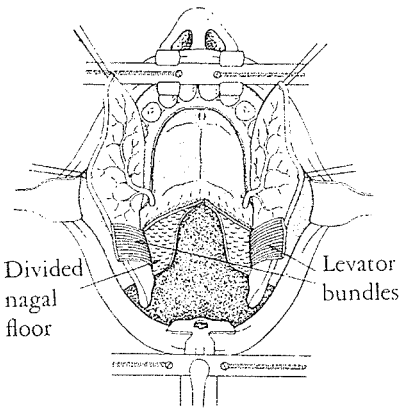


> The retrodisplacement principle has been combined with a pharyngeal flap in a manner which provides muscular union between the LVPM [levator veli palatini muscle] in the midline of the soft palate and the fibers of the superior constrictor in the posterior pharyngeal wall. Continuity of muscular tissue at the borders of each new lateral velopharyngeal portal thus provides

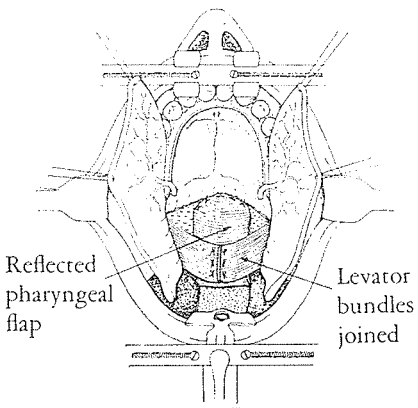
an anatomic configuration with the potential for functional sphincter action.



Acceptable indications for this procedure include non-cleft palate insufficiency, clefts of the soft palate with short levator insertions, and previously repaired clefts with persistent nasality.

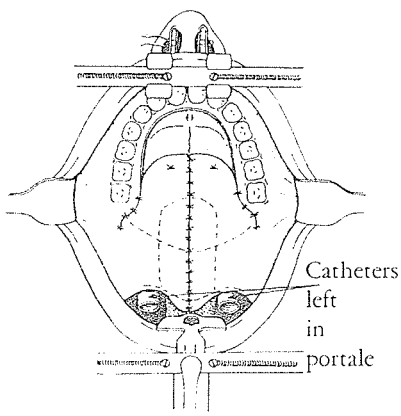


They use this approach in primary palate surgery, combining it with an extensive pushback. Their outline of the procedure in the *Cleft Palate Journal* began with elevation of mucoperiosteal flaps, freeing of vascular bundles and division of the nasal floor in the midline. This was followed by division of levator muscle bundles from the hard palate insertion as well as from the mucoperiosteum and nasal floor in order to permit retrodisplacement. Then a superiorly based pharyngeal flap was turned into the nasal defect and the levator bundles joined in the midline at the base of the flap. After completion of the palate pushback, catheters were left in each portal and fixed to the columella.



Here are Fisher's remarks in 1978 in reference to the paper:

First of all, it was the first honest admission that levator retrodisplacement alone in children with non-cleft velopharyngeal incompetence isn't very good very often. Second, it suggests a theoretic means for establishing a dynamic sphincter around the newly formed velopharyngeal portals. As of today, that is still speculative and unproven. I'm not sure my convictions are still strong that one can produce a dynamic sphincter with inherent muscular function. I was reminded of this when I listened carefully to Otto Kriens in Toronto last year. Surely, he has applied the levator repositioning principle most avidly, but he feels it is of advantage only within the first few months of life. I wonder if we have any chance at all of redirecting the function of those muscles if we wait until the child is 8 or 9 or 13? Thus, Levator Retrodisplacement with Pharyngeal Flap asks more questions than it answers.



PUTTING IT ALL IN PERSPECTIVE

After 150 years, finally, at the 1977 Third International Congress on Cleft Palate in Toronto, Samuel Pruzansky, with S. Peterson-Falzone, J. Laffer and P. Parris of the University of Illinois Center for Craniofacial Anomalies, imposed order on the chaos of the loosely grouped and ill-defined cases of hypernasality in the

absence of an overt cleft:

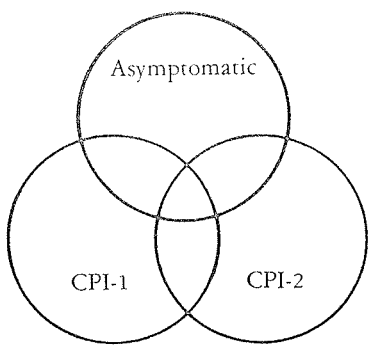
Hypernasality may be due to 1. disorder of the nervous system, 2. end organ defects, or 3. a combination of both.

All these cases of hypernasality were grouped under the heading of "Congenital Velopharyngeal Incompetence" (CPI) because the causative factors are present at birth. The incompetence (hypernasality) cases were divided into two groups, CPI(1) and CPI(2). In CPI(1), one or more of the triad of associated structural defects are present in a frequency of: bifid uvula, 85 percent; dehiscence of velar muscles, 65 percent; submucous cleft of the hard palate, 73 percent. Pruzansky noted that these associated defects are not causes of hypernasality, and one, two, or all three can be present without hypernasality. In CPI(2), no visible or palpable stigmata are present.

Radiographic examination of both types revealed one or more of the following: short or thin velum, platybasia, craniovertebral anomalies contributing to a deep pharynx, paucity of adenoid and early involution of adenoid.

CPI referral over the past 25 years has shown an increase in absolute number and in proportion to overt clefts, with an incidence of 49 percent CPI(1) and 40 percent CPI(2).

A study of 20 families of each group revealed the incidence of inheritance. CPI(1) showed a family history in 16.8 percent and CPI(2) in 22.2 percent, with an overlapping. Family history of clefts was seen in CPI(1) in 18 percent and in CPI(2) in 9.1 percent. It not only runs in families; it runs in syndromes!



As noted by S. Peterson-Falzone, S. Pruzansky, J. Laffer and P. Parris:

Both [CPI] types may occur in conjunction with a number of known craniofacial malformation syndromes, including mandibulofacial dysostosis, Klippel-Feil, von Recklinghausen, hemifacial microsomia, familial craniovertebral malformations, and others. Patients with Apert syndrome and Crouzon disease frequently exhibit the stigmata associated with CPI type 1 but do not show hypernasality due to decreased depth of the pharynx and abnormal size of the soft palate. In the presence of an overwhelming constellation of malformations, the hypernasality may be overlooked.