Current Concepts of Treatment of Ear Disease in Cleft Palate Children and Adults

RICHARD B. YULES, M.D.
Worcester, Massachusetts 01609

Introduction

Hearing plays an extraordinarily important role in rehabilitation in the cleft palate patient. Hearing is responsible for input which provides for the child's education, development and social adjustment. The processed hearing, speech, is severely hindered in patients who have significant hearing loss.

The discussion of hearing in the cleft palate patient must include—1) How much of a loss really exists in the cleft palate population? 2) What is the etiology of this loss and when does it start? 3) What is the relationship of the hearing loss to the initial and secondary cleft palate surgical procedures? 4) What type of therapy is available for both the initial and subsequent treatment of ear disease in the cleft palate population? 5) How does hearing affect the success of speech therapy and what methods are available to circumvent poor speech results in a poor hearing patient?

Hearing Loss in the Cleft Palate Populations. *Reviews* (*1*) by this author have demonstrated that over 50% of the adult cleft palate population suffers from a hearing loss. This author has presented results from over 14 studies on the incidence of conductive hearing loss in the cleft palate patient population. The patients involved in this collective study range in age from one month to 48 years of age. The number of patients employed in these studies, numbered 1,604. The criteria for a conductive hearing loss ranged from a 10 decibel to 30 decibel loss. The vast majority of these surveyed papers demonstrated hearing loss in over 50% of the population examined. This 50% figure is, of course, far in excess of the United States norm which has been currently established by Dr. Schein at .2% in his most recent census. These data led to the inescapable conclusion that the cleft palate population suffers in excess from what might be expected with a significant, debilitating, hearing loss. That this loss is conductive is important since conductive losses fall within the realm of current medical and/or surgical therapy providing the cause and/or lesion can be identified.

The Etiology and Beginning. A conductive loss must of necessity fall within the outer ear or middle ear space. There has been no significant increase of outer ear congenital abnormality demonstrated in the cleft palate population (*2*). In fact, all of the pathology appears to fall within the middle

ear. That the cleft palate patient is born and/or shortly develops serous effusions within the middle ear space, has been well documented by Stool and Randall in 1967 (*3*), and confirmed by Paradise and Bluestone (*4*) shortly thereafter. Stool and Paradise studied 94 patients ages two days through twenty months with a mean percentage of 95% serous effusions discovered. The findings of these investigators suggest that mucoid middle ear fluid is found in less than 20% of control children. These investigators also biopsied middle ear contents and found granulation tissue suggesting that the process of middle ear fluid formation might be taking place even *in utero*. That the cleft palate population suffers from a high incidence of serous effusions has now been widely accepted. The reason for the fluid formation in the middle ear space of cleft palate babies and children is unknown. Although it is possible that some mechanism provides for secretion of fluid into the middle ear of these babies, it seems more likely that the primary cause is eustachian tube dysfunction with subsequent irritation and/or absence of adequate drainage of the eustachian tube and/or middle ear space. Since it is apparent that the two muscles (tensor palatine and levator palatine) are defective at the palate end, there is no reason to expect that their function would be adequate on the eustachian tube end. There is, of course, the possibility that the absence of palate cover over the eustachian tube during swallowing provides a direct link to irritation of the eustachian tube mucous membrane with subsequent obstruction. For whatever reason, as will be shown shortly, when the eustachian tube-palatal-muscular-complex and/or eustachian tube-palatal-cover is restored, the incidence of ear disease in these patients falls sharply. This raises the question then of when for maximal effect on the hearing organ should surgery be initiated in the cleft palate population?

The Effect of Surgery. Data comparing hearing loss in patients who have undergone primary palate surgery at different ages and have undergone different surgeries are difficult to find. A large study by Masters (*5*) of 172 patients demonstrated hearing loss in only 31% when the palate was repaired between 0 and 17 months, a greater loss of 43% when the palate was repaired between 18 and 23 months, and the largest loss of 54% when the palate was repaired after 24 months of age. These data suggest that closure of the cleft and re-establishment of a cover for the eustachian tube by the palate itself and/or the re-establishment of the eustachian tube-palatal-muscle complex decreases the incidence of hearing loss, and we must infer the incidence of middle ear pathology. The establishment of adequate cover for the eustachian tube and good functioning of the eustachian tube opening musculature is suggested by the Masters study which demonstrates an incidence of 47% hearing loss when a simple palate repair is effected (VonLangenbeck repair), but when the palate is both closed and lengthened a hearing loss of only 29%. The highest incidence of hearing loss in this study was when only a prosthesis was employed leading to an incidence of loss of 78%. There seems little doubt based on these data, that the earlier

and more effectively the palate is both closed and lengthened in order to provide both adequate cover and function for the eustachian tube, the less the resultant conductive hearing loss in the cleft palate population.

In view of the fact that the most popular surgical procedure currently employed for *secondary* palate lengthening would now appear to be a palatal pushback accompanied by some sort of pharyngeal flap, one cannot help but question as to whether or not this pharyngeal flap might not interfere with eustachian tube function and then lead to a higher incidence of ear disease in the patient who has undergone secondary repair. Studies designed to look at this question are few and far between. This author has evaluated hearing in 69 patients who underwent pharyngeal flaps. These patients, age 5 to 48 years had all had previously repaired cleft palates; 58% of the patients had a preoperative air bone gap of greater than 15 decibels in the speech range. A similar loss was present in only 46% one year postoperatively, a statistically significant improvement. These data correlate well with other studies such as those performed by Graham and Lierle (*6*), and Aschan (*7*). Graham and Lierle's study of 43 patients, age 7 to 26 years, found no significant hearing loss and no ear infection following pharyngeal flap surgery; these investigators concluded that ear disease did not result from pharyngeal flap surgery. Aschan found an incidence of hearing loss of 33% one year postoperatively in a patient population who had a 40% incidence preoperatively. His conclusion was consistent with my data which suggest that if anything, pharyngeal flaps decrease middle ear pathology and help improve subsequent hearing. This hypothesis is not conclusive since both studies lacked evaluation of parallel patient populations whose hearing might have been improving even without pharyngeal flap surgery. The bulk of evidence points to the conclusion that surgery performed early and adequately assists middle ear function, probably by assisting eustachian tube function. The question remains as to how the ear disease can be treated when it exists in spite of adequate palatal correction.

Early Treatment. Serous effusions in the middle ear space of childhood populations are presently treated in this country by 1) decongestants, 2) tonsillectomy and/or adenoidectomy, and/or 3) middle ear aeration via paracentesis and/or tympanic membrane aeration tubes. The theory behind decongestants (either local or systemic) is that they will establish eustachian tube drainage by removing edema of the mucous membrane around the eustachian tube and/or treat the primary cause of the swelling such as allergy with the use of an antihistamine. Decongestants have had notoriously poor results in the cleft palate population although there are presently no good studies comparing patients who have been maintained on decongestants against the patients who have not been maintained on decongestants. That such studies are lacking is probably silent evidence that no otologist is going to gamble by not placing his patients on decongestants as a first line of defense against serous effusions.

Tonsillectomy and/or adenoidectomy is a questionable procedure for

serous effusions even in the non-cleft palate population. That no data in a cleft palate population should be available is therefore not surprising. None-the-less, it is the feeling on the part of most otologists that a tonsillectomy and/or adenoidectomy in the cleft palate patient population is contraindicated because it opens even wider the oronasal port making the patient's speech frequently worse and seeming to have no effect on the subsequent middle ear disease. There are otologists who do advocate lateral band adenoidectomy, a procedure which spares the bulk of the midline adenoidal mass against which the palate can unfold during speech while at the same time presumably decompressing the eustachian tube by removing obstructive adenoidal tissue in the lateral fossa where its effect on speech is less pronounced.

A review (*8*) of 81 patients published by this author demonstrated a rather clear cut relationship between velopharyngeal incompetence and tonsilloadenoidectomy. In view of these results and in the absence of any data demonstrating a positive effect on speech and/or hearing by adenotonsillectomy, it is the opinion of this author that the procedure is contraindicated in the average cleft palate child unless infection is the basic problem. Serous effusions can presently be handled by more conservative means.

The single best procedure currently available for treatment of middle ear effusions is drainage via the tympanic membrane with a paracentesis. There are those because of the recurrent nature of the serous effusions who would advocate not draining the fluid and simply fitting the patient with a hearing aid until he reaches adolescence. In spite of this vocal minority, the vast majority of otologists feel strongly that treatment of middle ear effusions in the palate population is best handled by draining the fluid away so that the middle ear for at least a while becomes aerated. There are those who prefer to drain the ear by repeated aspirations without the placement of middle ear aeration tubes, but the vast majority of America's otologists would opt for the establishment of middle ear aeration for as long as is possible with a given procedure; this requires a tympanic membrane middle ear aeration tube. Middle ear ventilating tubes have been employed in the past, but were not popularized until a report of Armstrong in 1954 (*9*). Since that time, many different shapes, materials and points of insertion have been described. Regardless of the feeling of the otologist as to which tube and/or which material (be it teflon, silastic, polyethylene or stainless steel) and regardless of the shape (be it a button, an arrow, a straight tube, a phlanged tube) and/or its location (posterior inferior or anterior inferior) the purpose of the tube is the same—aeration of the middle ear. There are no good data to suggest when these tubes should be inserted for maximal long term results. It is safe to say that the state of medicine at this time suggests treatment of the cleft palate population in the following manner: As soon as serous effusions can be documented either by pneumotoscopy and/or by impedance audiometry, aspiration of the fluid with insertion of the surgeon's favorite tube should take place. When these tubes fall out,

as is always the case, tubes should not be immediately re-inserted but the effusion, should it recur, should be allowed to recur. Following treatment with decongestants which are usually unsuccessful, the tube may then be re-inserted in several months. The theory behind this approach is that repeated tube placements will not weaken the tympanic membrane unnecessarily, yet at the same time the fluid will not be allowed to become so thick and tenacious that it can not be removed and/or leads to a permanent thinning and/or collapse of the tympanic membrane. This tube re-insertion is continued until adolescence, until a permanent perforation of the tympanic membrane results, and/or until no effusions recur.

Since repeated tube insertions have become the rule rather than the exception, one can but ask the question as to why a *permanent* tube has not been developed. There have been attempts to develop such a tube. Initially, a malleus clip tube was employed consisting of a small bore lumen polyethylene or silastic tube with a small stainless steel clip. The tube was inserted on either side of the malleus and clipped into place. It was the experience of people employing these tubes that the malleus necrosed in some cases and/or the tube plugged in others. The most recent attempt on the part of this author was in a study designed jointly with Dr. Herbert Silverstein who developed a tube which was inserted by drilling through the middle ear bone itself in order to fix it firmly in place. Tubes were inserted in 18 cleft palate patients who had serous effusions. Although the tubes themselves remained beautifully in place, it was our experience over a three year interval that every single tube plugged by either filling -1) with crusting secretions which could not be conveniently cleaned with a straight pick and/or local medications or, 2) as was discovered in three years in ears which were explored-filled along the median aspect with granulation tissue and/or in one case cholesteatoma debris. Thus, our experience to date is that no permanent aeration tube has been successful in maintaining permanent middle ear aeration in the cleft palate child. A recent tube described by Crabtree (*10*) which has a mesh which allows it to adhere to the medial aspect of the tympanic membrane may offer some hope for a safe tube which can be inserted with ease and which can be rapidly extracted should it occlude.

Late Ear Disease. In spite of decongestants and middle ear aeration, a significant percentage of cleft palate patients come to adulthood with poorly aerated ears: Some have atelectatic tympanic membranes collapsed over the ossicular structures in some cases fixing them; some have small and/or large tympanic membrane perforations; and some have cholesteatoma—the result of epithelial ingrowth into the diseased middle ear. Although there are no data in cleft palate patients to demonstrate at what age correction of this middle ear disease should take place, the concensus of otologists is that middle ear reconstruction should wait until adolescence because it is at this time that eustachian tube function even in the cleft palate population appears to have maximally developed. The sole exception to this statement

might be widespread destruction occurring from cholesteatoma in the child, a case where the surgeon might be forced to eradicate the disease before destruction of the total ear takes place.

Although the older literature is replete with a variety of procedures, current otologic surgery has centered around mastoid tympanoplasties with ossicular reconstruction. Although surgeons will differ with regard to whether or not the posterior canal should or should not be left up and/or whether or not the reconstruction of the ossicular chain would be secondarily staged, the ultimate goal would appear to be agreed upon: elimination of any middle ear and/or mastoid disease and reconstruction of the sound conduction apparatus with as little hearing loss as is possible. The type of procedure which is employed is dictated in the final analysis by the amount and/or type of disease present. When the middle ear space is collapsed and under a 20 decibel air-bone gap is present, most otologists would opt for no intervention. When a 30 decibel or greater loss is apparent, a tympanoplasty is indicated. When a perforation exists with no apparent drainage and/or epithelial ingrowth, a tympanoplasty is also indicated. When chronic ear drainage and/or cholesteatoma formation is apparent, then a mastoid tympanoplasty is indicated. The tympanoplasty with or without a mastoidectomy can be classified into five classical groups: Type I, Type II, Type III, Type IV, and Type V. The "Type" number refers to the position of the tympanic membrane graft in relation to the ossicular chain. The status of mastoid tympanoplasties has been beautifully reviewed by Silverstein (*11*) in 1972. A Type I tympanoplasty places the graft on the malleus. A Type II tympanoplasty places the graft over the incus. A Type III tympanoplasty places the graft onto the head of the stapes. A Type IV tympanoplasty places the graft onto the footplate of the stapes. A Type V tympanoplasty places the graft in continuity with the inner ear space either via a horizontal semicircular canal fistula or through a fat graft placed in the oval window in place of the stapes. The most common grafting material currently in vogue is temporalis fascia, the technique for its use being nicely described in the 1970 monograph by Dr. Hough (*12*). In the cleft palate population because of the history of poor middle ear aeration, tympanic membrane ventilation tubes are frequently employed through the graft at the time of surgery. The success rates of mastoid tympanoplasty in the cleft palate patient population are currently not available, probably because of the small series in any given otologist's hands. None-the-less, a success rate of 75% in eliminating disease and/or establishing serviceable hearing which is accepted for the non-cleft patient population is probably no different from the success rate in the cleft palate population itself.

Hearing's Effect on Speech. The ultimate integrated output of the patient's hearing, speech, is seriously affected by a patient's hearing loss. It is well established that patients with over a 30 decibel air-bone gap tend to speak more hypernasally than their peer palate popluation. Why this should be so is not clear. Perhaps more palatal pharyngeal incompetence is

associated with the more severe hearing loss; therefore, patients with poor hearing speak more hypernasally simply because they have more palate pathology. That this hypothesis is incorrect has been well established by studies which I have reported as well as results from Skolnik (*2*), Aschan (*7*), and Loeb (*13*). Evaluation of my 69 patients demonstrated in spite of adequate velopharyngeal closure as demonstrated by cineradiography and respirometry studies, patients who had a hearing loss of 30–40 decibels almost always had rhinolalia; whereas, patients with under a 20 decibel hearing loss had a postoperative success rate of over 60%. My data are consistent with Aschan who found a six fold incidence of hypernasal speech in his postoperative patients with a 30 decibel or greater hearing loss. Loeb demonstrated even more dramatically the difference in postoperative patients, the incidence of hypernasality 90% in patients with poor hearing with only 55% in those with reasonably normal hearing. There can be little doubt that for whatever reason, hearing, especially an air bone gap of over 30 decibels, which is defective hinders the patient from correcting this hypernasal speech.

If it is the inability of the patient who is hard of hearing to hear his hypernasality which accounts for his poor speech, then it is reasonable to offer a sensory cue other than hearing to assist the development of normal nasal speech. It is for this reason that a visual cue conditioner demonstrating nasality vs non-nasality has been developed and employed by this author (*14*). The results of any large series remains yet to be seen.

Summary

This author has demonstrated that the incidence of conductive hearing loss in adults who have had cleft palates approximates 50%. That their ear disease arises during infancy has been well documented by Stool and Paradise. The serous otitis media which develops in infancy and persists through adolescence is presently best treated by tympanic aeration tubes. Whether or not a limited adenoidectomy is a useful adjunct in treatment remains to be conclusively demonstrated. That cleft palate repair and/or pharyngeal flap surgery is helpful in the eventual outcome of ear disease would seem to be so. The end product of recurrent serous effusions and/or ear infections can be a chronic draining ear and/or cholesteatoma which following the attainment of adolescence is best treated by mastoid tympanoplasty with ossicular reconstruction. The effect of hearing on the speech of palate patients is discussed and a new method for using visual cues in helping palate patients speak less hypernasally offered.

reprints: *Richard B. Yules, M.D.*
470 Pleasant Street
Worcester, Massachusetts 01609

References

1. Yules, R. B. Hearing in cleft palate patients. *Arch Otolaryng. 91*, 319–323, 1970.

2. Skolnik, E. M. Otologic evaluation in cleft palate patients. *Laryngoscope 68*, 1908–1959, 1958.
3. Stool, S, and P. Randall Unexpected ear disease in infants with cleft palate. *Cleft Palate J. 4*, 99–103, 1967.
4. Paradise, J. L. and C. D. Bluestone, Diagnosis and management of ear disease in cleft palate infants. *Trans. Amer. Acad. Ophthal. Otolaryng. 73*, 709–714, 1969.
5. Masters, F. W., H. G. Bingham, and D. W. Robinson, The prevention and treatment of hearing loss in the cleft palate child. *Plast. reconstr. Surg. 25*, 503–509, 1960.
6. Graham, M. D., and D. M. Lierle, Posterior pharyngeal flap palatoplasty and its relationship to ear disease and hearing loss. *Laryngoscope 72*, 1750–1755, 1962.
7. Aschan, G. Hearing and nasal function correlated to postoperative speech in cleft palate patients with velopharyngoplasty. *Acta Otolaryng 61*, 371–379, 1966.
8. Chaco, J., and R. B. Yules, Velopharyngeal incompetence post tonsillo-adenoidectomy: An electromyographic study . *Acta Otolaryng. 68*, 276–278, 1969.
9. Armstrong, B. W., A new treatment for chronic secretory otitis media. *Arch. Otolaryng. 59*, 653–657, 1954.
10. Crabtree, J. A., Permanent tympanic ventilating tube. *Otolaryng Clinics N. Amer.* pp. 61–65, 1970.
11. Silverstein, H., Surgery for chronic suppurative otitis media. *N. E. J. Med. 287*, 287–290, 1972.
12. Hough, J. V. D., Tympanoplasty with the interior fascial graft technique and ossicular reconstruction. *Laryngoscope 80*, 1385–1413, 1970.
13. Loeb, W. J., Speech hearing, and the cleft palate. *Arch. Otolaryng. 79*, 20–30, 1964.
14. Yules, R. B., and R. A. Chase, A. training method for reduction of hypernasality in preech. *Plast. reconstr. Surg. 43*, 180–185, 1969.