

5. Ear Disease and Hearing Loss in Cleft Palate

CLEFT palate has long been associated with ear disease and hearing loss. In 1878 A. Alt was the first to mention a case of otorrhea in a deaf-mute patient with a cleft palate. He closed the cleft, and the otorrhea healed. The patient was able to hear again and then learned to speak.

Conductive deafness associated with cleft palate has been reported to occur in between 27 and 50 percent of patients: Halfond and Ballenger, 50 percent; Nylén, 40 percent; Skolnik, 39 percent; Pagnamenta, 30 percent; Spriestersbach, 29 percent; and Meissner, 27 percent.

Possible causes considered in this association of cleft palate and ear disease have varied among repeated infections following the lack of an intact palatal partition between the oral and nasal cavities, disturbance by the cleft of the normal muscle physiology necessary for adequate Eustachian tube function, timing and type of surgery, scarring after surgery, effect of hamular fracture, actual abnormal Eustachian tube anatomy, and cranial base deformity.

REFLUX

The presence of a cleft in the palate was thought to allow reflux up the Eustachian tube causing otitis media. Even when treated with antibiotics without drainage, the purulent material was merely converted to a sterile exudate which remained as a foreign body resulting in conductive hearing loss.

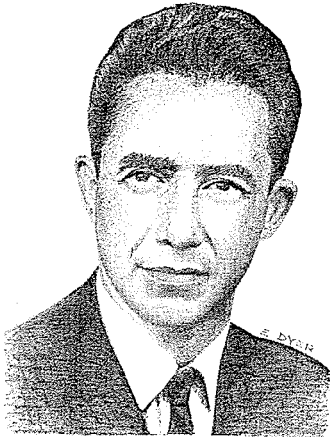
LACK OF MUSCLE FUNCTION

As more thought and research were directed to this problem, blame was laid on the abnormal insertions of the musculature which, disrupted by the cleft, prevented normal action and adversely affected the function of the Eustachian tube. Surgeons started to look to closure of the cleft in the palate as early as possible as a step toward partially improving the muscle function.

EARLY SURGERY ADVOCATED

In 1960 Frank Masters, Hal Bingham, and Dave Robinson of the University of Kansas Medical Center began to study hearing loss in cleft palate patients and added normal hearing to normal speech and normal facial growth as goals of cleft palate treatment. They deduced:

The chronic recurring middle ear infection with its subsequent audiometrically detectable hearing deficit is a manifestation of altered eustachian physiology which in the cleft palate child is almost inevitable as the extrinsic musculature of the eustachian apparatus is not intact. Thus, early restoration of the dynamic physiology of the eustachian apparatus by creation of a normal muscular insertion appears to be the best method available to reduce the appalling incidence of hearing loss.

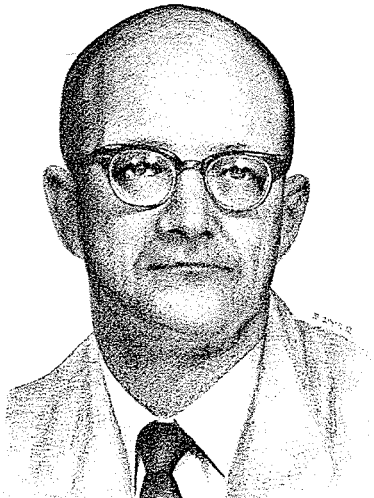


Beverly Armstrong

TREATING THE EAR DISEASE ITSELF

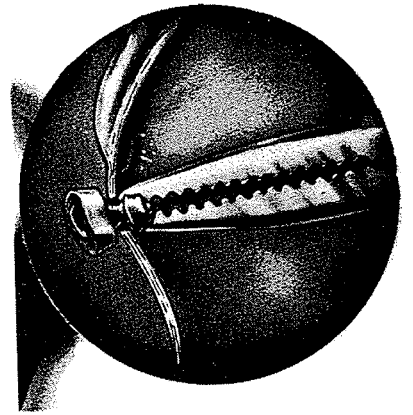
In 1954 Beverly W. Armstrong of Charlotte, North Carolina, first described insertion of *pressure equalization* (PE) tubes to maintain aeration following myringotomy, aspiration and tympanostomy for middle ear fluid.

In 1966, in the *Cleft Palate Journal*, James Donaldson of the University of Washington, Seattle, published an illustration of insertion of the Silastic tube after myringotomy, with the inner flange passing through the tympanic membrane and the outer flange securing it in place. He reported that, out of 702 cleft palate patients examined, 98 (13.9 percent) had middle ear pathology requiring one or more myringotomies. The infant was not suspect, but ages ranged from under 1 up to 15 years, with the largest number in the 3- to 4-year period. To establish middle ear aeration in these patients with inadequate Eustachian tube



James Donaldson

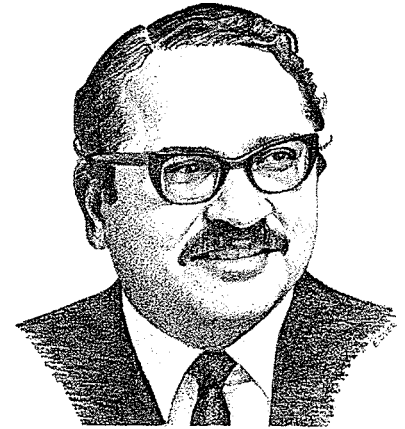
function he advocated insertion of Silastic tubes. Although the tubes tended to be extruded sooner than desired, temporary middle ear aeration frequently improved the appearance of the tympanic membrane, allowing it to assume its normal position rather than a retracted one.



EAR PATHOLOGY IN THE INFANT

Still no one suspected ear pathology in the infant. In 1958 Skolnik had stated that the incidence of pathology was 6 percent in cleft palate children younger than a year old and increased to 60 percent in preschool children.

In 1967 Sylvan E. Stool and Peter Randall at the Children's Hospital, Philadelphia, reported on bilateral examination of the middle ears of 25 cleft palate infants under general anesthesia through a binocular operating microscope after myringotomy. Ninety-four percent of these ears contained mucoid material, and each of 10 biopsy specimens revealed the presence of granulation tissue. They suggested:



Sylvan Stool

A reasonable approach to the problem of otitis media in the cleft palate infant is to examine these patients with magnification whenever they are having reconstructive surgery. The removal of abnormal material from the middle ear and the ventilation of this cavity should allow pneumatization of the ear to proceed in a more normal manner and should aid significantly in preventing subsequent hearing loss.

Interested in how it all began, I asked Peter Randall, who had long been concerned with the miserable problems of draining ears in children with clefts and the difficulty of finding cooperative otolaryngologists. Here are his reminiscences, written to me in 1973:

On September 11, 1963, Sylvan arrived on the scene. As you know, he is a somewhat rotund, jolly genius who had gone from medical school in Texas through his Pediatric training in Seattle, Salt Lake City and Boston and through a complete Otolaryngology residency at the University of Colorado in Denver. Sylvan arrived in Philadelphia fresh out of his residency being Board qualified in both Pediatrics and Otolaryngology with a lot of enthusiasm, absolutely no practice whatsoever and a perfectly beautiful, brand new Zeiss operating microscope. The day after he arrived we had our big monthly Cleft Palate Clinic and Sylvan—who had wondered how long it

would be before he saw his first patient—found one ear after another with problems beyond description. He described it as an “Otolaryngologist’s Paradise.” Within a very few days, he was in the operating room pushing his new Zeiss scope from one room to another asking me if I would mind if he looked at the ears of a child whose cleft lip I was about to tackle. This was the first time we had even thought of looking at the ears in an infant this young (about three months of age), and I thought he was really just trying to get experience with his new operating room toy. The child had had no known trouble with his ears, but Sylvan immediately said, “Oh my gosh, look at this!” which I did but frankly I didn’t see very much. Then he asked if I would mind if he made a little hole in the eardrum and I asked how little was a little hole and why he wanted to do it, and he said that it was very little, would heal very quickly, and the reason was that he was sure there was a lot of trouble behind that particular drum. I shudder to think of my lack of informed consent, but under the circumstances it seemed that this would be the best thing to do for that child at that point. After myringotomy he began sucking out thick, inspissated, mucoid “glue.” I thought that he had opened into a mucocele. He then asked if I would mind if he looked at the other ear which I readily agreed to, and the same problems were found on that side as well.

As luck would have it we had a second cleft lip to follow in the next room, and it wasn’t long before Sylvan was asking if I would mind if he looked at that child as well. At that stage of the game we were taking Silastic intravenous catheters and cutting them into small pieces to use for insertion in the myringotomy openings. The various “buttons” were not yet available at Children’s Hospital.

We were soon misled by a child whom we examined at about six weeks of age and actually found granulation tissue within the middle ear space, which led us to believe that much of this material was infected. However, Charles Bluestone has since shown that virtually all of it is not infected in these infants. We set out to try to study the incidence, and couldn’t really figure out how to get a “control group.” Then one day, I pulled a dirty trick on Sylvan and asked him to look at the ears of a child who had a cleft of the lip but no cleft of the palate. After he did the myringotomy and found that this was a perfectly normal drum with an air-containing middle ear space, he asked, “What is the matter with this child? His ears are perfectly all right.” Then it was obvious that our control group should come from those patients with clefts of the lip only. We found that 87% of our infants with cleft palate had thick inspissated mucus in their middle ear space and only about 28% of those with clefts of the lip only had any kind of fluid in the middle ear space and this was usually quite watery.

Buddy Bluestone had been working along the same lines with Jack Paradise at Pittsburgh, but I’m fairly sure it did not antedate our work in

1963. It's interesting that they came up with the same incidence figures exactly that we had, but they carried it one step further in that they brought back the children who did not have the mucoïd material in the middle ear on one and even two successive occasions and showed that eventually 100% of these children get into trouble with collection in the middle ear space. Bluestone has done a very careful study of the fluid, its characteristics and studies on the Eustachian tube.

Almost simultaneously, the team of Paradise and Bluestone had directed its interest toward this ear problem. Jack L. Paradise, pediatrician at the University of Pittsburgh, in an occasional escape from crying babies, soothes his nerves by restoring antique oriental rugs. Enticed into the Cleft Palate Clinic by Betty Jane McWilliams to look into the problem of feeding and nutrition in young cleft infants, he consistently discovered bilateral secretory otitis media.

C. D. Bluestone, Alpha Omega Alpha at the University of Pittsburgh School of Medicine, became interested in the cleft palate problem in 1959 by Sam Pruzansky, while interning at the University of Illinois. In 1964 he joined the cleft palate team as otolaryngologist at the University of Pittsburgh and by 1966 was confirming Paradise's findings by myringotomy with an operating microscope. In 1969 Paradise, Bluestone and Felder reported the universality of otitis media in infants with cleft palate:

Bilateral secretory or suppurative otitis media was found without exception in 50 infants with cleft palate 20 months of age or younger. By contrast, in a group of 100 infants without cleft palate chosen at random, otitis media was present during 22 percent of 274 clinic episodes they experienced during an 8-month period. . . . Infants with cleft palate who received myringotomy and suction, but without insertion of plastic tubes, developed early re-accumulation of fluid which required repetition of surgery. The course of infants who received myringotomy with tubes was generally satisfactory; but in some, otorrhea, early extrusion of tubes, or both occurred.

Chronic secretory or suppurative otitis media probably develops in all infants with cleft palate, and usually within the first month of life. In such infants, myringotomy accompanied by the insertion of tubes appears to be the best method of treatment now available. This procedure may be performed promptly and repeated as often as necessary to maintain middle ear aeration. It is hoped thereby to avoid the scarring, ossicular damage, cholesteatoma, or suppurative intracranial complications which otherwise



Jack Paradise



Charles Bluestone

might eventually develop in some patients. Also, if surgery is not performed, hearing impairment will probably persist throughout infancy or longer, with untoward effects on well-being and function and with serious implications for intellectual, speech and emotional development.

Then Jack Paradise in 1970 studied 200 children under 12 months of age—100 with cleft palate, 100 without clefts. He found serous fluid in the middle ear in 100 percent of the cleft and 20 percent of the non-cleft children.

Also in 1970, Joyce Heller, Irving Hochberg and Gastone Milano of Newark State College, Union, New Jersey, studied 60 cleft palate and 60 non-cleft palate children aged 3 to 12 years. They found:

Cleft palate children have significantly poorer hearing sensitivity than non-cleft children, but all threshold deviations were within normal hearing limits for both.

There was an improvement in hearing sensitivity in both cleft palate and non-cleft palate children as a function of increasing age and this was most evident above six years.

There was a significantly greater incidence of conductive hearing impairment (significant air-bone gap) and aural pathology in cleft palate children than in non-cleft palate children.

Approximately 50 percent of the cleft palate children whose otoscopic findings were positive showed unilateral abnormalities.

SUBVERSIVE SUBMUCOUS CLEFT PALATE

In 1971 La Vonne Bergstrom and William Hemenway of the University of Colorado Medical Center used otolaryngological and audiometric examination to evaluate 58 patients with submucous clefts of the palate. Recurrent or chronic disease of the middle ear in 39 percent ranged from serous otitis media to cholesteatoma. Thirty-four percent had conductive hearing loss, and 25 percent had either pure sensorineural or mixed hearing losses. Half of the patients with middle ear disease did not have speech disorders, and hence submucous cleft palate might not be suspected on that basis. Submucous cleft palate should suggest the possibility of accompanying, perhaps

asymptomatic, middle ear disease, and unresolving middle ear disease might be the reason to suspect SMCP.

ADVOCATES OF EARLY CLEFT CLOSURE

In 1972 Charles Bluestone, Jack Paradise, Quinter Beery and Ronald Wittel studied 22 infants with unoperated cleft palates who received myringotomies with tube insertions during the first three months and between the ages of 18 and 24 months had secondary palate closure with a V-Y pushback with or without an island flap. They evaluated Eustachian tube protective function by roentgenographic studies after instillation of radiopaque media into the nasopharynx and noted:

Infants with unrepaired palate clefts were conditionally able to clear radiopaque fluid media in a prograde direction from the Eustachian tube and middle ear into the nasopharynx, but there was obstruction to retrograde flow from the nasopharynx into the Eustachian tubes. Following surgical repair of the palate, there was improvement in prograde clearance and, in over half the ears tested, retrograde flow appeared normal.

Abnormal distensibility of the Eustachian tube in infants with cleft palate was suggested by the results of tests following repair of the cleft. This distensibility may be a function of reduced tubal stiffness (increased compliance) which may in turn contribute to functional obstruction of the tube by rendering its opening more difficult.

Their conclusion:

Prior to closure of the palate, middle ear fluid is present in all untreated infants. Since this seems to be related to functional obstruction of the Eustachian tube, ventilation of the non-aerated middle ear cavity is indicated. A previous study has shown that closure of the secondary palate is often followed by a reduction in the prevalence of middle ear disease. The present investigation confirms this finding and suggests a relationship to improvement in Eustachian tube function. It would, therefore, seem worthwhile to reconsider repairing the palatal defect at as early an age as possible, especially when otorrhea through a tympanostomy tube has failed to respond to medical treatment. Following repair of the palate, recurrence of middle ear effusion warrants repetition of myringotomy and insertion of tympanostomy tubes. However, if Eustachian tube function is improved following palate repair, or if otorrhea develops through the tympanostomy tubes, their removal on a trial basis may be indicated.

*First reference
to possible
specific
Eustachian
tube pathology*

In 1973 Paulsen of Denmark reported:

Ear disease seems a little easier to control in patients with incomplete than with complete palate clefts, and a striking finding in patients with all types of clefts has been the improvement in overall otologic status following palate repair.

In 1975 E. R. Soudijn and A. J. C. Huffstadt of the State University Hospital, Groningen, studied 132 standard cleft palate patients before and after palate surgery. They found "glue" in the ears of 94 percent of the presurgical cases and reported:

Six months after the closing of the soft palate, the percentage of glue ears appeared to have already been reduced to 65. There is of course a tendency for decrease of aural pathology with advancing years, but this does not occur until the age of four, according to Graham. In our patients the highest age was only 19 months.

The most interesting aspect of our study however is the relatively high percentage (30%) of glue ears in children with cleft lips and in children of the control group. . . . In babies with a cleft palate the pharyngeal mechanism appears to be so severely disturbed, that in 94% of the cases glue cannot be drained from the aural cavity.

They concluded:

Probably all children's middle ears contain glue at birth. In the cleft palate babies glue is present in 94%.

Inspection of the eardrums, eventually followed by myringotomy in cleft palate babies is indicated periodically.

Closing of cleft palate by surgery leads to improvement of tubal function.

In 1974 Margareta Korsan-Bengtson and Olle Nylén of Göteborg, Sweden, studied the ears and hearing of 60 children aged 8 to 15 years who had had a Wardill-Kilner palate closure at 16 to 20 months and, if the cleft was complete, had had a Johanson-Ohlsson bone grafting at 8 to 10 months. Hearing and middle ear function was normal in 33 out of the 60. Screening audiometry revealed the incidence of conductive hearing loss exceeding 20 decibels at two or more frequencies to be 3 to 5 percent. The corresponding percentage of operated cleft palate children was 20 percent. This figure was considered very favorable since 75 percent of these children had serious middle ear disease before surgical closure of the palate. They concluded:

*normal
glue!*

*In our cleft
palate clinic in
1978, Arlene
Earley has
found no fluid
in the middle ear
of cleft palate
babies 3 to 4
weeks of age*

It would thus seem justified to recommend early palate closure, even from an otological point of view, in order to prevent tubal dysfunction with middle ear changes and hearing loss.

In 1977 Lee Dellon wrote:

In 1969 the first patient we corrected the levator of told me when he awoke that he could hear better. Post-op audiograms demonstrated correction of his conductive hearing loss. All we had done was alter his levator's insertion. I believe long-term follow-up will show that if levator retrodisplacement is included in the primary palatal repair, not only will speech results be improved but also hearing loss diminished, and I have written this limerick to emphasize it.

Oh, Levator Veli Palatini,
Thou art more than wet linguini,
For when velum quakes
and Eustachius awakes
'Tis 'cause thou art in-between!

Medical art student Susan Seif and Dellon have written a 1977 study of "Interrelationships of the Levator and Tensor Veli Palatini Muscles and the Eustachian Tube: An Anatomic Reconstruction from Serially Sectioned Fetal Heads." It discounts the tensor as a tube opener but acknowledges the part played by the levator:

← speculation?

Indeed during isometric contractions the TVP increased girth will exert a pressure radially inward against the paratubal tissue. . . . The levator veli palatini muscle (LVP), as it goes from lateral and inferior to the medial plate of the Eustachian tube cartilage medially into the mobile soft palate, is in a unique position to elevate the medial tube cartilage. An isotonic LVP contraction elevates the soft palate posteriorly and the tubal cartilage medially. During this LVP muscle contraction the paratubal tissue would be compressed radially inward from below, while the radially inward "closing" pressure exerted by the "resting" medial cartilage would be relieved. . . . The incidence of hearing loss diminishes with increasing age because . . . with growth and development the LVP origin moves laterally (increasing its effectiveness in releasing the pressure of the medial cartilage plate on the tubal lumen) and the insertion moves downward and forward (increasing velar excursion and, again, increasing LVP effectiveness as a tubal "opener."

speculation?

NOT ALL CLEFT CLOSURES CURE

In 1962 D. C. Spriestersbach, Dean Lierle, Kenneth Moll and William Prather of the State University of Iowa studied hearing in 163 cleft lip and palate patients and found the incidence of loss and magnitude of threshold deviations significantly greater in the

youngest age groups (33 to 77 months) than in any of the older groups. They noted no significant correlation between physical management (type or time of surgery) and hearing loss.

Unfortunately, closure of the cleft in the palate, although usually of benefit, does not invariably cure the ear problem. This failure has been blamed by some on certain aspects of the surgery, such as hamular fracture, tensor tendon division or other traumatic and scarring procedures responsible for preventing the development of normal tubal function. A 1920 study by A. R. Rich of Johns Hopkins University utilized a palatal incision to allow visualization of the tubal orifices. Rich found these orifices to be closed normally at rest but opened during the swallowing, yawning and sneezing reflexes.

The levator palatini, the palatopharyngeus, the internal pterygoid and the superior constrictor muscles of the pharynx, when either cut or stimulated, were found to exert no influence whatever upon the patency of the orifice or lumen of the tube.

The tensor palatini was the only muscle functionally related to the Eustachian tube. Contraction of this muscle was always accompanied by a dilatation of the tubal orifice and lumen. Relaxation or division of the tensor palatini was followed by a passive return of the tubal walls to the condition of approximation which they normally occupy when at rest. This work, of course, eventually pointed an accusing finger at fracture of the hamulus or division of the tensor tendon during cleft palate surgery.

*Should the
name tensor
palatini really
be tensor tuba
auditoria ?!*

HAMULAR FRACTURE

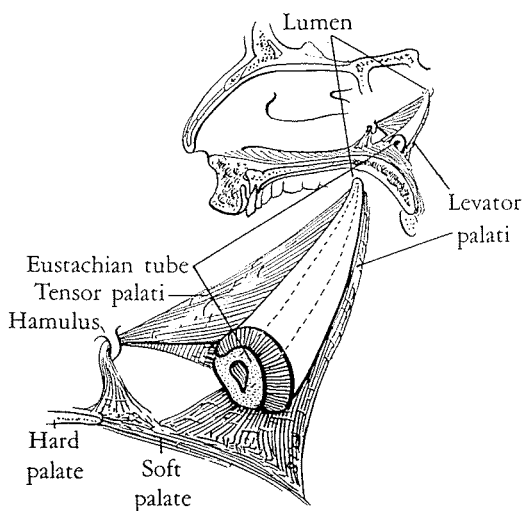
Fracture of the pterygoid hamulus during cleft palate closure has been blamed for perpetuating hearing impairment. Newmann in 1968, Graves and Edwards in 1944, and McMyn in 1940 all demonstrated that in addition to the main tensor veli palatini, which hooks around the hamulus to insert into the soft palate, a smaller component of this muscle arises from the Eustachian tube cartilage and inserts into the hamulus and the end of the hard palate. Skolnik in 1958 and M. A. Ross in 1971 confirmed that hamular fracture is related to occurrence of otitis media. Politzer,

as early as 1862, pointed out that the muscular relations of the Eustachian tube of the dog correspond closely to those of man. In 1962 Holborow's experiment in dogs concluded that "the integrity of the tensor is essential for tubal opening," and in 1971 Odoi, Proud and Toledo, after unilaterally "expunging" the hamulus in dogs, reported development of middle ear effusions.

Yet modern human studies raise much doubt as to whether hamular fracture affects ear physiology. In 1968 M. Bennett, R. H. Ward and C. A. Tait found no such increase after tensor tendon division. In 1972 Bluestone, Paradise, Beery and Wittel, following unilateral hamular osteotomy and infracture on 12 patients, noted:

No difference in Eustachian tube function was observed as a result of this procedure, and although follow-up has been short, recurrences of middle ear effusion have also appeared unrelated.

In 1973 in the *Cleft Palate Journal* Barrett Noone, Peter Randall, Sylvan Stool, Ralph Hamilton and Richard Winchester of the University of Pennsylvania presented a sketch of their



rendition of tube, tensor and hamulus anatomy. They then reviewed a randomized series of 89 patients undergoing soft palate closure between 1963 and 1969 in which unilateral hamular fracture was alternated between the right and left sides. They reported:

An evaluation of the development of clinical middle ear disease and documented hearing loss by audiogram during a three-year postoperative fol-

low-up period demonstrated no difference between the ear on the side of hamulotomy compared to the opposite ear.

OTHER SURGICAL TRAUMA

In 1966 Otto Kriens of Bremen, while studying the anatomy of palatal musculature in Prague, concluded that the levator muscle elevated the medial edge of the tubal cartilage and additional opening was effected by the pull of the tensor muscle. In the cleft palate, the pull of the tensor muscle was in deviated craniolateral motion which did not elevate the medial edge of the Eustachian tube. Kriens warned against surgical intervention near the epipharyngeal portion of the tube. He felt that fracture of the hamulus might disrupt the musculotendinous apparatus near the tube, and, even worse, dissecting and packing the space of Ernst could disrupt the equilibrium of the muscles in this area, to be further compounded by resultant scarring.

EFFECT OF PHARYNGEAL FLAP ON HEARING

Evidently pharyngeal flaps do not affect hearing. M. D. Graham and D. Lierle found in 1962 that the pharyngeal flap procedure did not aggravate an existing hearing loss. In 1966 G. Aschan concluded that surgical reconstruction by velopharyngoplasty helps to restore tubal functioning and thus hearing. G. W. Leworthy and H. Schliesser reported in 1975:

The application of a pharyngeal flap did not decrease the preoperative hearing acuity in 96 per cent of our 53 patients.

T & A

To remove or not to remove the tonsils and adenoids, that is a question even in the normal child. Recurrent nasal discharge, repeated attacks of otitis media and respiratory obstruction are accepted as criteria for a T & A. In 1965 N. I. Chalot noted improvement in hearing in 75 percent of cleft palate patients after tonsillectomy and adenoidectomy.

Yet in the non-cleft child or in the unrecognized submucous cleft case, removal of the adenoids can result and has resulted in the disastrous production of hypernasal speech. In the cleft palate child the need for any extra prominence in the posterior pharyngeal area to aid in velopharyngeal closure causes hesitation in the random removal of the adenoids. Some surgeons are enthusiastic about partial adenoidectomy in the treatment of otitis media, preserving speech by removing only the lateral portion of the adenoid under direct vision. This maneuver, however, is not so easy. As suggested by Stool, it may be possible to preserve the child's adenoid tissue if the ear disease is treated via the tympanic route first.

The removal of the tonsils is slightly less hazardous, provided the surgeon shells out the tonsil carefully, preserving the anterior and especially the posterior pillars and avoiding excessive post-operative scarring.

POSSIBLE ANATOMICAL CAUSES OF INADEQUATE TUBE FUNCTION

According to Wilma Maue-Dickson of the University of Miami, there is no indication that the extrapalatal anatomy of the tensor veli palatini muscle is abnormal in cleft palate. Even in severe cleft of the palate the presence of a well-defined palatal aponeurosis (the tendon of tensor) can be demonstrated in the palatal tags in histological sections. In 1975 an anatomical study by Maue-Dickson showed that human fetuses with cleft palate consistently have (1) narrower and smaller auditory tube lumina which are more widely separated than in normals, (2) greatly enlarged auditory tube cartilages, also more widely separated than in normals, (3) more widely separated pterygoid plates than in normals, and (4) significantly reduced pharyngeal height but greatly increased pharyngeal width—all of which were shown in cross section in Chapter 2. One conclusion from these data is that the space between the lateral pharyngeal wall and the side wall of the cranium is substantially reduced and that the tube may suffer a mechanical disadvantage as a result. The problem may be reduced by craniofacial growth, which sometimes relieves stress

Latham, Long and Latham (1980) found in a 3-D study of a 5 mo. cleft palate infant: The levator muscle appeared to be in a position to obstruct the auditory tube during muscle contraction.

on the tube. This suggestion is consistent with the observation that children with cleft palate typically have reduced middle ear problems within the first few years of life.

P A T H O G E N E S I S

It is the responsibility of the Eustachian tube to ensure that the external and the middle ear air pressures remain the same. Anything that interferes with evacuation of embryonic tissue from the middle ear and subsequent pneumatization of the temporal bone, or with the function of the Eustachian tube to prevent it from supplying the middle ear with air, will result in conductive hearing loss.

There are several clinical stages in the pathogenesis of ear disease, as noted by Stool. The *exudative stage* has acute, subacute and chronic phases.

The *viscid stage* results from failure of resolution of the exudative stage. The middle ear is filled with thick, tenacious mucoid substance. The audiogram usually reveals an average loss of 3 decibels due to failure of movement of the tympanic membrane. Treatment involves removal of the viscid material from the middle ear via a myringotomy and substitution of a prosthesis for the function of the Eustachian tube.

In the *adhesive stage* the tympanic membrane is adherent to the structure of the middle ear. It is usually seen in later childhood when the previous stage was not resolved. The membrane becomes atrophic and flaccid and collapses against the medial wall of the middle ear with a loss of 40 to 60 decibels. Desquamated epithelium gets caught and collected in the adhesive pockets of the tympanic membrane. Treatment includes myringotomy, aspiration of fluid and insertion of tubes coupled with eversion of invaginated pockets. The replacement of atrophic membrane with grafts may be of value. The best treatment for this stage is prevention.

The most serious complication is the formation of a cholesteatoma. In the cleft palate this is acquired during the adhesive stage when invaginated pockets collect squamous debris. Diagnosis may depend on x-ray demonstration of a radiolucent area.

Removal of the abnormal tissue is mandatory for cures and to prevent extension into surrounding structures including the brain.

In 1975 Jack Paradise took an international view of middle ear problems associated with cleft palate, stating:

Eighty years have elapsed since Gutzmann in Germany first observed that approximately half of all patients with cleft palate suffer from significant reduction in auditory acuity. . . . The mucous membrane lining becomes markedly thickened, the epithelial cells undergo metaplasia, and there is a great increase in the secretion of mucus.

Noting that even children with normal palates commonly experience secretory otitis media, he listed a string of international investigators studying the histological and biochemical abnormalities that characterize this sterile inflammatory process in the middle ear. Sade (Israel), Senturia (U.S.A.), Paparella (U.S.A.), Lupovich (U.S.A.), Gunderson and Gluck (Norway) and Mogi (Japan) have all contributed to a better understanding of the middle ear changes.

PULLEN

Fred Pullen of the University of Miami School of Medicine, while training at the Massachusetts Eye and Ear Infirmary in Boston, became infatuated with otology under Harold Schuknecht. Pullen's nasal fracture during a Michigan Golden Gloves tournament, a skilled light touch and his center-of-the-target attack on clinical problems made him the perfect otorhinolaryngology member of our Cleft Palate Team. Here is his 1977 stand:

The Eustachian tube connects the middle ear cavity to the nasopharynx. In the adult, the anterior two-thirds is cartilaginous and the posterior third bony, but in the infant the bony portion is relatively longer. The direction of the tube in the adult inclines superiorly with the horizontal plane at an angle of 30° to 40° , but in the infant this inclination is only 10° . The lumen of the Eustachian tube is shaped like two cones with the apex of each directed toward the middle. The aural orifice of the tubes is oval in shape, measuring 5 mm high and 2 mm wide in the adult. The nasopharyngeal orifice in the adult is a vertical slit at right angles to the base of the skull, but in the infant this opening is oblique due to the more horizontal position



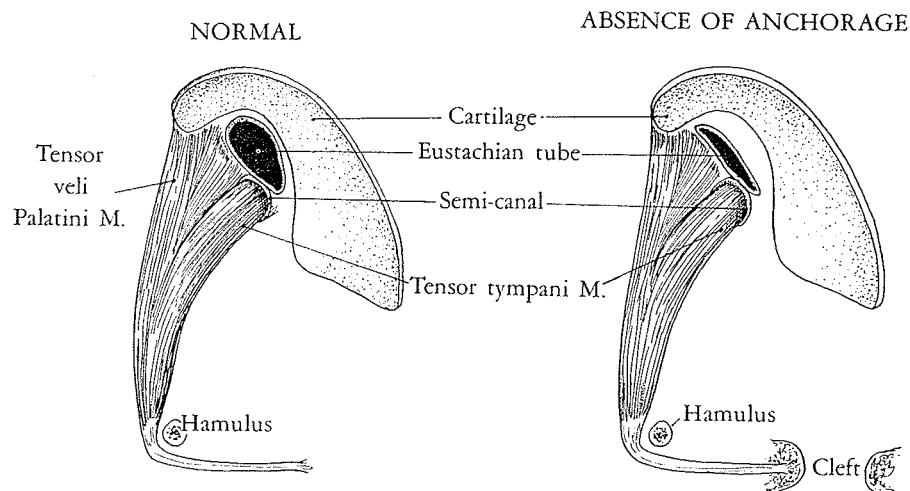
Fred Pullen

of the cartilage. The diameter of the orifice is 8 to 9 mm in the adult and 4 to 5 in the infant. In the newborn, the nasopharyngeal orifice lies in the plane of the hard palate, but in the adult it is situated 10 mm above this plane. The middle portion of the Eustachian tube, or isthmus, is not sharply constricted, but is relatively long, with gradual widening at each end, forming the aural and nasopharyngeal orifices. The diameter of the isthmus in the adult is 1 to 2 mm; but in the infant it is somewhat larger. The mucosal lining of the cartilaginous portion is similar to that of the nasopharynx and contains mucous glands. The mucosa in the protympanic portion of the Eustachian tube is similar to that of the middle ear and contains both mucus producing elements and cilia. The function of the Eustachian tube appears to be two-fold: clearance of secretions and ventilation of the middle ear.

Usually the Eustachian tube is closed, but it opens during swallowing, yawning and sneezing, permitting the air pressure in the middle ear to equalize with atmospheric pressure. This opening mechanism is muscular and involves the nasopharyngeal orifice. The tensor palati is the only muscle related to tubal function, and closure is the result of relaxation of the tensor palati with passive approximation of the tubal walls.

The tensor tympani muscle originates from the cartilaginous portion of the Eustachian tube and the adjoining part of the greater wing of the sphenoid, as well as from the bony canal in which it is contained. It passes backward through the canal, enters the middle ear as a slender tendon which bends laterally around the processus cochleariformis and is inserted into the neck of the malleus and into the tympanic membrane directly.

In 1970, A. J. Lupin demonstrated by dissections that the tensor palati and tensor tympani muscles have fibers that intermingle. Both of these muscles have fibers originating in the area of the greater wing of the sphenoid near the spine, and also both have fibers originating from the Eustachian tube. There is intermingling of these fibers on the undersurface



of the sphenoid spine. The common development of these muscles is from the mandibular arch with a common nerve supply from the mandibular division of the trigeminal nerve. There are three branches to the three divisions of the tensor palati muscle and one to the tensor tympani. These two muscles are, in fact, continuous and, indeed, the tensor tympani muscle is a continuation of the muscle fibers from the anterior portion of the tensor palati with additional slips from the base of the sphenoid bone and from the periosteum lining its semi-canal. The contraction of the medial part of the tensor palati muscle unrolls the tubal cartilage by depressing the lateral lamina, thus opening the nasopharyngeal orifice of the tube.

It is postulated here that the contraction of the tensor palatini is accompanied by a similar contraction of the tensor tympani muscle, thereby exerting a slight medial or inward movement of the tympanic membrane. This combined movement thereby acts as a "pump" mechanism to help clear the air or "push" secretions out of the Eustachian tube into the nasopharynx. Tympanometry measurements of the ear were performed at -150 mm of water pressure thereby causing the tympanic membrane to be pulled laterally into the external auditory canal. Upon voluntary contraction of the tensor palati and tympani muscles, the pressure in the outer ear canal increased by 3 to 5 mm, proving a medial motion of the tympanic membrane thereby substantiating this "pump" mechanism. The function of the tensor tympani muscle is therefore to provide 3 to 5 mm of positive pressure in the middle ear and Eustachian tube upon the opening of the Eustachian tube by the tensor palati. This pressure need not be more than that when one realizes the minute movements of the tympanic membrane which produce hearing. Air is thereby forced out of the Eustachian tube upon swallowing and new air then rushes into the tube and middle ear, providing a continuous cycle of high oxygen containing air in the middle ear. The function of the tensor tympani muscle has always been a mystery. Clearly now the function and importance of this muscle has become elucidated.

In patients with cleft palate there is no opposing action of the opposing tensor muscles, the Eustachian tubes remain closed and the pump mechanism does not function. If the Eustachian tube fails to open, the middle ear cannot be adequately aerated and fluid usually accumulates in it.

Identification of Serous Otitis Media

Pneumatic otoscopy as noted by J. Northern in 1976 is the accepted final criterion and absolute reference in the evaluation of ear disease. However, physicians vary tremendously in ability to interpret their observations during otoscopy and pneumotoscopy. A case history can be unreliable, and hearing tests conducted during infancy are rather complicated. Consequently, an early recognition of any disturbances in early childhood mainly depends upon an improved diagnosis, with the aid of a microscope.

also Speculated
on by D. Dickson
in 1976

Pullen's
pump

This is
a first.

This "pump"
theory was
proven by
Donald Kammerer
of Pittsburgh
in Laryngoscope
1978

In regard to audiology, traditional hearing tests are not sufficient to identify otitis media, and changes in hearing sensitivity do not necessarily relate to changes in the otologic disease process. Fifty percent of 408 ears with serous otitis media would have passed as "normal" on school hearing tests conducted at the accepted screening level of 25 dB HL.

Acoustic Impedance Measurements

The most sensitive screening technique for identification of serous otitis media is tympanometry. Tympanometry is defined as an objective technique for measuring the compliance (or mobility) of the tympanic membrane while varying air pressure in the external auditory canal. Tympanic membrane mobility is of particular importance since almost any pathological condition located on, or medial to, the eardrum will influence its movement. Tympanometry, compared to otoscopy, is totally objective, and eardrums noted to have normal mobility by pneumatic otoscopy examination can be shown to have abnormal mobility with tympanometry. Acoustic impedance contributes especially meaningful information about middle ear disease in cleft palate children and should be a routine part of their evaluation. In 1975, F. Bess, H. Lewis and D. Cieliczka noted that in cleft palate children identification of middle ear problems is possible with impedance when the audiometric and otologic examinations were normal.

Management of Otologic Problems

The treatment of infants with cleft palates must be directed toward the correction of effects as well as causes. Before repair of the secondary palate, elimination of middle ear fluid and aeration of the middle ear are the responsibility of the otologist and can be accomplished by myringotomy, aspiration and insertion of tympanostomy or pressure equalization tubes. The insertion is usually made in the anterior superior or inferior quadrant. The middle ear is then aspirated with a suction tube and the PE tube inserted.

This procedure should probably be performed soon after birth and close follow-up is necessary at least every two months. When spontaneous extrusion of tubes occurs, repeat myringotomy and reinsertion should be carried out. During the first year of life, these infants ordinarily have one or more reconstructive procedures on the lip or primary palate requiring general anesthesia. This provides an excellent opportunity for re-evaluation of the tympanic membranes, by means of the operating microscope, and for reinsertion of tympanostomy tubes if necessary. Infants should probably sleep in the prone position; when awake and supine, it may be helpful for the head and neck to be kept elevated at least 20° above the horizontal.

The tympanostomy pressure equalization tubes should be reinserted as many times as necessary until adequate functioning of the Eustachian tube is

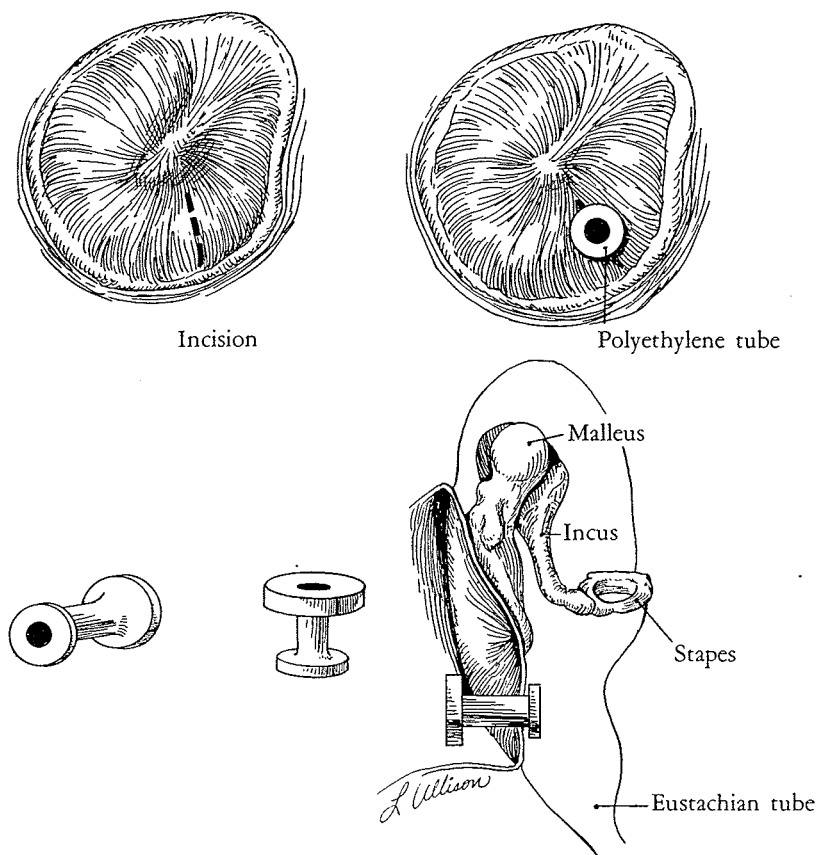


Illustration of use of a polyethylene tube as a pressure equalization tube.

demonstrated by a well-pneumatized middle ear upon repeated examinations. Occasional autoinflation of the middle ear may be performed to maintain the pneumatized middle ear. It is only continuous and meticulous attention to the aeration of the middle ear which will prevent serious and permanent otologic disability, and as long as air is there, I am not concerned about early closure of the palate.

THE PULLEN PLUG

Fred Pullen has devised a plug to allow children wearing tubes to swim, and in Florida swimming is important. He noted in 1978:

A high molecular weight polymer plastic cork (Richards Manufacturing Company, Memphis) which allows air but not liquids to pass in, is inserted into the P.E. tube. These little patients can then go swimming without fear of otitis media from getting water in the middle ears. This plug has been used for two years in over 200 cases with only one infection.

Thus, it can be said that cleft palate infants have an extremely high incidence of fluid in the middle ear, and whether this is due to the cleft in the normal tensor mechanism, abnormal tube anatomy or the relatively restrictive and compromising craniofacial relations, certain points become clear:

1. If fluid in the middle ear is ignored, hearing loss is increased, especially in low frequencies with inability to hear hypernasality. (Deaf children often speak with hypernasality.) The hypernasal cleft palate speech, not being detectable to the patient, reduces his chances of correction, even after surgical production of a competent velopharyngeal sphincter, thus setting up a *vicious cycle* for the speech therapist.

2. Early examination under the microscope, myringotomy, suction of accumulated fluid and insertion of tubes are important.

3. Repeated examinations under the microscope during the cleft surgery and reinsertion of tubes when necessary are essential.

4. As long as aeration of the middle ear is maintained, there is no hurry for palate surgery as far as hearing is concerned.

5. Craniofacial growth is probably responsible for improved tubal function, and therefore time is an ally if aeration is maintained.

6. It is possible that freeing, uniting and retrodisplacing the levator is the part of the surgery that benefits the Eustachian tube function.