Middle Ear Problems Associated with Cleft Palate

An Internationally-oriented review*

JACK L. PARADISE, M.D.
Pittsburgh, Pennsylvania 15261

Prevalence of Hearing Impairment and Ear Disease

The present state of our knowledge concerning middle ear problems in patients with cleft palate comprises important contributions from at least nine nations. Eighty years have elapsed since Gutzmann (*11*) in Germany first observed that approximately half of all patients with cleft palate suffer from significant reductions in auditory acuity. That observation has withstood the test of time; its essential correctness has been confirmed repeatedly, particularly during the past 20 years or so (*4*). Moreover, as shown by Pannbacker (*24*) (U.S.A.) and Walton (*41*) (U.S.A.), the prevalence of hearing impairment if relatively minor losses are included is probably considerably higher than 50%. The nature of this hearing loss has been identified as mainly conductive, although more recently, Bennett (*3*) (U.S.A.) has pointed out that among young adults with cleft palate, sensorineural losses are also to be found.

Conductive losses would be expected from consideration of the types of eardrum abnormalities found in these patients: scarring, distortion of landmarks, adhesions, retraction, and perforation. Prior to 1966, representative studies such as those of Skolnik (*35*) (U.S.A.) and Aschan (2) (Sweden) reported middle ear abnormalities of some sort or other in perhaps 70 to 80% of all patients with cleft palate, but the populations studied consisted mainly of adults and children, not infants.

In 1966 and 1967 came Stool and Randall's observation (*38*) in Philadelphia, and our own (*29*), independently, in Pittsburgh, that otitis media is present in virtually all infants under the age of two years with unrepaired cleft palates. The middle ear effusions we found in these infants varied widely in viscosity (*18*), and most were sterile, but occasionally suppuration was present. More recently, the observation that young infants with cleft palate are universally affected with middle ear disease has been re-

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From the University of Pittsburgh Cleft Palate Center; the Departments of Pediatrics and Community Medicine of the University of Pittsburgh School of Medicine; and the Children's Hospital of Pittsburgh.

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ported from Germany by Koch and co-workers (*15*), and essentially similar observations are being currently reported from the Netherlands by Soudyn and Huffstadt (*37*), and from Norway by Møller (*23*).

Nature of the Disease in the Middle Ear

Even in children with normal palates, secretory otitis media is a very common disorder, and investigators in several countries have studied the histological and biochemical abnormalities which characterize this sterile, inflammatory process in the middle ear. Sadé and co-workers (*32, 33, 40*) (Israel); Senturia et al. (*34*) (U.S.A.); Paparella and Lim and co-workers (*25, 17, 26, 16, 12*) (U.S.A.); Lupovich et al. (*18*) (U.S.A.); Gunderson and Glück (*10*) (Norway); and Mogi et al. (*22*) (Japan) have all contributed to our current level of understanding of the changes that occur in the middle ear. The mucous membrane lining becomes markedly thickened, the epithelial cells undergo metaplasia, and there is a great increase in the secretion of mucus.

Pathogenesis in Relation to Eustachian Tube Function

It seems natural that attention to the middle ear disease of cleft palate patients should not have been limited to phenomenologic observation, but should also have been directed at pathogenesis. Early on, this attention focussed on the Eustachian tube. On the basis of anatomic studies, Holborow in England postulated (*13, 14*) that the tube was unable to open properly and ventilate the middle ear, so that tympanic liquid accumulated. Similarly, in a report published in 1964, Paulsen (*30*) in Denmark, using impedance measurement techniques which had been developed by his countrymen Metz (*21*) and Terkildsen and Nielsen (*39*), concluded also that children with cleft palate suffered from Eustachian tube dysfunction. More recently, roentgenographic studies of infants and young children with unrepaired clefts by Bluestone and others of our group have shown that there is obstruction to the normal flow of instilled radiopaque contrast medium from the nasopharynx into the nasopharyngeal portion of the Eustachian tube; conversely, however, if contrast medium is injected via a tympanostomy tube into the middle ear, it flows more or less normally into the nasopharynx (*4, 6*). In further studies, employing a modification of a technique first developed by Flisberg (*9*) in Sweden, we found that the Eustachian tube is unable to completely equilibrate air pressure differentials between the middle ear and the nasopharynx (*5*). These latter observations therefore tend to support the earlier concept (*13, 14, 30*) that although the Eustachian tube in patients with cleft palate is anatomically patent, functional obstruction results from impairment of its opening mechanism. We believe that this impairment is in turn related to increased compliance of the tubal wall. Certain changes in these X-ray findings in the direction of normal occur in some patients following palate repair (5), but they are not consistent, and their significance is not certain.

Implications of Middle Ear Disease

What is the importance, in pragmatic terms, of all these observations to the patient with a cleft palate? Since we know that in patients generally, chronic secretory otitis media predisposes to middle ear infection, and can ultimately result in middle ear adhesions, scarring, and even, as described by Buckingham (*7*) (U.S.A.), cholesteatoma, it seems reasonable to infer that some of the structural middle ear alterations and associated conductive hearing losses noted in older cleft palate patients might be the end-result of untreated inflammatory disease earlier in life. The recent observation by Paparella et al. (*27*) (U.S.A.) that inflammation in the middle ear can lead to pathologic changes in the inner ear suggests that the sensorineural losses found in some patients with cleft palate (*3*) may also originate from chronic disease in the middle ear.

Moreover, it seems reasonable to assume, both on acoustic grounds and on the basis of experience in older children and adults with middle ear effusions, that the persistent effusions of infants with cleft palate cause conductive hearing losses of variable degree, and that these persistent losses may in turn interfere with aspects of intellectual, language, speech, and emotional development, at least in some infants. It is therefore intriguing to speculate that the restrictions of language skill (*19*) and the psychological problems (*20, 36*) found by McWilliams and Smith (U.S.A.) to be prevalent in older children with cleft palate might be traceable, at least in part, to hearing loss in infancy and early childhood.

Early Treatment: Preliminary Results and Unanswered Questions.

In Pittsburgh we began in 1966 to undertake an aggressive treatment program with regard to the middle ear effusions of young infants with cleft palate, with two main purposes in mind. First, we hoped to institute and maintain normal hearing acuity throughout infancy and early childhood, thereby assuring optimal conditions for speech, language, and related areas of development; and secondly, we hoped to avoid later otic and auditory handicaps. We scheduled infants for myringotomy at the earliest practicable time, usually within a few weeks of initial examination. Liquid found in the middle ear at myringotomy was aspirated with a small suction tip, and a bi-flanged polyethylene tube was then inserted through the incision and left in place. Whenever tympanostomy tubes became blocked or were extruded, and middle ear effusion recurred, myringotomy and tube insertion were repeated.

Armstrong (U.S.A.) had of course originated this now popular method of treating chronic secretory otitis media some 10 years earlier (*1*), and at about the same time that we were beginning to apply the method to patients with cleft palate in Pittsburgh, Donaldson (*8*) and Stool (*38*) were also beginning to do so in Iowa City and Philadelphia, respectively.

Our experience to date indicates that with this regimen most infants

and young children can be maintained in satisfactory otologic status most of the time. Ear disease seems a little easier to control in patients with incomplete than with complete palatal clefts, and a striking finding in patients with all types of clefts has been the improvement in overall otologic status following palate repair (*28*). Paulsen (*31*) has recently made similar observations in Denmark.

Because the treatment program requires frequent examinations and usually several myringotomies, and because otorrhea through tympanostomy tubes is a common complication (though usually fairly easily controlled), it is important to determine whether this program results in significant, long-term benefits. In order to determine this, groups of patients who receive this type of routine, aggressive otologic management beginning in early infancy must be compared with respect to otic and auditory status, and language and intellectual function, with otherwise similar groups who have been treated otologically only when they complained of ear pain, or developed purulent otorrhea, or became obviously hard of hearing. We are currently involved in this type of comparative study, but conclusive results are not yet available.

Broader Implications

The problem of middle ear disease in patients with cleft palate is of more than parochial interest. Middle ear infections and their complications and sequelae in subjects with *normal* palates constitute a problem of immense proportion throughout the world. However, as in subjects with clefts, understanding of pathogenetic mechanisms is incomplete, and prevailing methods of therapy leave much to be desired and therefore remain issues of controversy. Attempts at prevention are best described as primitive.

Since infants and children with cleft palate constitute a natural model for the study of otitis media, improved understanding of pathogenesis and long-term effects, and improved methods of treatment in patients with clefts could have broadly beneficial pediatric impact. It is in this broader light that the contributions listed here from workers in many countries should be viewed.

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