Research Revisited

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Stool SE, Randall P. (1967). Unexpected ear disease in infants with cleft palate. Cleft Palate J 4:99–103.

In 1963, after completing a residency in otolaryngology and having previously completed a residency in pediatrics, I joined the staff of Children's Hospital of Philadelphia as the first geographic full-time otolaryngologist, determined to make a career in pediatric otolaryngology. My second day at that institution, the Cleft Palate Clinic met; I attended, introduced myself to Peter Randall, and told him that I was interested in children with ear disease. I examined the 20 children who came to the clinic and found that there was a very high prevalence of draining ears, cholesteatoma, and middle ear effusion. My findings were very interesting—I felt I had found an otolaryngologic gold mine and that there would be enough work there to keep me busy for several weeks. Unfortunately, I also found that the clinic met only once a month.

This left a large amount of free time, which at that time I had available, and so on Thursday mornings I would go to the operating room, to Dr. Randall's surgery. As he was operating, I noticed that during the palate repair he inserted a finger to fracture the pterygoid hamulus. I told him that this was a very bad thing to do because it caused ear disease. He asked me how I knew that, and I replied that my professor had said that plastic surgeons ruin the ears when they fracture the hamulus and alter the Eustachian tube function. Peter then questioned me as to whether my professor was always correct. As I reflected on this question, I thought he was not always correct but seldom in doubt.

When I looked at the child's ears I found that, indeed, this child already had ear disease. It then seemed appropriate to start looking at all of the ears of children undergoing cleft lip or palate surgery, and after several weeks of doing this, I found that even the very young children who were having lip adhesions already had ear disease.

Consequently, we started to do myringotomies every Thursday morning on all of Dr. Randall's patients, and I found a high prevalence of fluid in the middle ears of children who had cleft palate. In contrast, those children who had cleft lip only (without cleft palate) had a relatively low prevalence of ear disease. It was possible during this time for me to obtain some of the fluid for examination, and occasionally I was able to remove some tissue from the middle ear cleft. Examination of the tissue invariably revealed granulation tissue and, occasionally, well organized fibrosis.

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We accumulated a significant series of these patients and, in 1965, presented the findings at the meeting of The American Cleft Palate Association in New York. Subsequently, after many rewrites, we submitted a paper to Hugh Morris (then Editor of *The Cleft Palate Journal*) regarding ear disease in cleft palate. Hugh called after receiving the paper; he felt a more appropriate title would be "unexpected ear disease," because, indeed, we had not expected to find such a high prevalence of disease in the very young infant.

Several years later I had the opportunity to meet Jack Paradise and Charles Bluestone of Pittsburgh and found that they had been making similar observations. They preferred to call the problem "the universality of ear disease" in infants with cleft palate.

Another observation was that the cephalometric films of these patients revealed poor pneumatization of the temporal bone. Review of the rather sparse literature on cleft palate showed that Harvold had observed in his thesis an incidental finding that pneumatization was strikingly abnormal and that the cell structure of the system was compact. This also seemed to be an important finding, and consequently, when one of the junior medical students asked me whether I had any projects that might be of interest, I suggested we look through the cephalometric films of the Growth Center. Dr. Krogman agreed that we could examine these films, and we selected a group of children with clefts and children without clefts. Using a planimeter, we measured pneumatization, and these measurements revealed that there was significantly decreased pneumatization of the temporal bone in the children with cleft palate. These observations were published in *The Cleft Palate Journal* in 1969.

When Dr. Shprintzen recently asked me to review this early work, one of his questions was, "How good were these observations." I feel that the observations of middle ear disease in the infant with cleft palate and of the failure of pneumatization have been corroborated by a number of sources. There have been articles, published mostly in the English language literature from the United States and Europe, that substantiate the findings of ear disease in very young infants. The observation of failure of pneumatization, while not studied extensively in the cleft palate literature, has been studied extensively in the otologic literature. It appears that both of these findings are valid.

Some of the other observations that were made have also been of interest. Initially, I attempted to obtain cultures of the middle ear fluid, and invariably the cultures showed no growth. When I moved to Pittsburgh, we continued the attempt to culture middle ear fluids. Dr. Casselbrant (who

had joined the staff in Pittsburgh) initiated a study of the microbiology of middle ear fluids of infants with cleft palate. Her technique involved sterilizing the external canal, obtaining a specimen, and having the microbiologist come to the operating room, where the specimen was planted immediately. When this technique was used there was a significant increase in positive findings, and indeed, the organisms found in the middle ear of infants with cleft palate were much the same and virtually in the same proportion as those found in the chronic middle ear effusions of children without cleft palate.

It seemed obvious to me at the time that identification of a disease process early in life facilitated timely intervention that might prevent some of the complications and sequelae of long-term ear disease, which I noted my first day in the Cleft Palate Clinic. It also seemed obvious that the proper approach would be to ventilate the middle ear using a myringotomy and tube procedure; with ventilation the middle ear would permit the temporal bone to be pneumatized and, it is hoped, keep the tympanic membrane and middle ear healthier. We subsequently started a program whereby all children with cleft palates were examined with a microscope while they were under anesthesia and were then seen regularly for otologic and audiologic examination. When tympanometry eventually became available, it was utilized to diagnose middle ear disease.

It is my impression that this approach is now being used in a number of centers throughout the world, and although it is difficult to evaluate the impact, studies by Dr. Paradise reveal that the effect on language development caused by chronic middle ear disease may be favorably influenced by myringotomy with tubes. A number of other studies, such as anatomic and functional studies of the Eustachian tube, have been stimulated because of these early observations. We have also been able to show, using auditory brainstem response testing, that infants have a conductive hearing loss secondary to the fluid in the middle ear.

Dr. Shprintzen asked whether I felt we had solved the problem of ear disease in children with clefts. My response has to be that, initially, I felt that frequent ventilation of the ears should be able to solve the problem of failure of pneumatization and decrease the amount of hearing loss. I now feel that the problem has only been partially solved, because it has become apparent that cleft palate is a complex abnormality; even though we ventilate the ears, we have not completely eliminated middle ear disease in this population. There are still some children in whom middle ear effusion accumulates in spite of treatment. It is my impression, however, that there is considerably less irreversible chronic ear disease. The difficulty at present is that we have no way of predicting which of the children will recover and how many times it will be necessary to ventilate the ears. To paraphrase Drs. Paradise and Bluestone, it is possible to say, "The disease is universal; however, the treatment has not become universal." There remains a need to understand more about the pathogenesis, pathophysiology, and epidemiology before we can develop a satisfactory universal method of therapy that will prevent the socioeconomic consequences of ear disease that are secondary to the anatomic abnormality.

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