Controversies in the Evaluation and Management of Otosclerosis

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KEYWORDS

• Otosclerosis • Stapedectomy • Stapedotomy • Bisphosphonates • Lasers
• Implantable hearing devices • Middle ear actuators • Barotrauma

KEY POINTS

- Although genetic loci have been identified and the measles virus implicated in the development of otosclerosis, the exact mechanism of the bone remodeling associated with otosclerosis remains uncertain.
- Systemic treatments to prevent the progression of cochlear otosclerosis have been limited; however, new-generation bisphosphonates may be more effective, but not without risks.
- Proper preoperative evaluation minimizes the likelihood of stapes surgery on patients with a dehiscent superior semicircular canal or concomitant Meniere's disease.
- Although stapedotomy in children with juvenile-onset otosclerosis is effective, it is not without risk. Consequently, in children with unilateral disease, delaying surgery until adulthood may be preferred.
- Innovations in technology, such as middle ear actuators, may provide patients with far-advanced otosclerosis an alternative to traditional stapedotomy or cochlear implantation.

HISTORICAL PERSPECTIVE

Dating back to the times of Kessel[1] and Politzer,[2] controversies have surrounded the etiology and management of the entity that Politzer[2] first termed, *otosclerosis*. What was originally believed to be a condition attributed to "chronic interstitial middle ear catarrh" with secondary stapes fixation was discovered by Politzer[2] to be a primary disease of the labyrinthine capsule, which he referred to as "otosclerosis." Despite his publication of histologic evidence of otosclerosis in 16 cases of stapes fixation, it took almost half a century for Politzer's views to gain universal acceptance. Even today, controversy still surrounds the precise etiology of otosclerosis. Based on

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Fowler's[3] study in identical twins showing almost a 100% concordance rate of otosclerosis and the more recent identification of 10 genetic loci (*OTSC*1–10) associated with otosclerosis, most investigators concur that otosclerosis is an inherited disease that is transmitted in an autosomal dominant pattern with an incomplete penetrance rate of 20% to 40%.[3–5] The exact cause of the abnormal bone remodeling that produces the otosclerotic foci remains uncertain; however, ultrastructural and immunohistochemical evidence of measles-like structures and antigenicity in active otosclerotic lesions published by McKenna and colleagues[6,7] has implicated the measles virus in the formation of the otosclerotic foci. In addition, measles ribonucleic acid has been demonstrated in fresh footplate specimens as well as archival ones. This hypothesis may be further strengthened by the decline in otosclerosis after introduction of the measles vaccination.[8] Research is ongoing to clarify the role of the measles virus in the development of otosclerosis.

The controversies surrounding the etiology of otosclerosis pale in comparison to the controversies that have been associated with its management. Kessel,[1] who is considered the "Father of Stapes Surgery," was under the mistaken opinion that the hearing loss associated with otosclerosis was caused by increased pressure in the inner ear fluids. He theorized that by removing the stapes, he could relieve that pressure. Prior to testing his hypothesis in humans, he removed the columella, which is the stapes equivalent, in 2 pigeons. As he described it, clear fluid drained from their ears for 8 days until a membrane formed to seal the oval window. According to Kessel,[1] the pigeons did not experience any vertigo or hearing loss. Based on these "pre-clinical animal observations," he performed stapes mobilizations and stapes removal in humans. He reported "some improvement in hearing and no serious complications."[1] Obviously, his perceptions were not consistent with the perceptions of other clinicians. In many cases, the hearing improvement lasted only for a period of days to weeks, and there was always the risk of labyrinthitis and rarely meningitis. Consequently, stapes surgery fell into disrepute and was vehemently criticized by the leading otologists, Politzer, Siebenmann, and Moure, who in 1899 at the Sixth International Congress of Otology declared, "Stapes surgery is useless, often mutilating, and dangerous." They went on to say, "The question of surgical therapy for otosclerosis was interred with great pomp at the 1894 International Conference in Rome. There is no reason to revive it."[9,10]

Fortunately, more than 50 years later, Dr John Shea[11] revived stapes surgery. But even this revival was not without controversy. It was only with the assistance of Dr Howard P. House,[12] that Dr Shea was able to present his stapedectomy technique before the Triological Society in Montreal, Canada. At 11:00 AM on Thursday, May 17, 1956, Dr House was scheduled to moderate the panel, "Symposium - The Operation for the Mobilization of the Stapes in Otosclerotic Deafness." Just before the panel was to begin, Dr Shea told Dr House about his first patient who had successfully undergone a stapedectomy. Knowing that something like this would be extremely controversial, Dr House told Dr Shea that he would call on him as the last discussant from the audience. Dr Shea took the opportunity to approach the podium and present his stapedectomy experience to the membership of the Triological Society. Before Dr Shea could be unduly criticized by the audience, Dr House explained that unfortunately the time for the symposium had expired and brought down his gavel, ending the session.[12]

CURRENT CONTROVERSIES

Evaluation

With this historical perspective as a backdrop, the current controversies as they relate to the evaluation and management of otosclerosis are reviewed.