Clinical Records

In-the-ear hearing aids within auricular prostheses

J. J. HOMER, M.D., F.R.C.S. (ORL-HNS), A. C. ROBERTS, PH.D.*, C. L. WENGRAF, F.R.C.S.

Abstract

We report a child with bilateral congenital microtia in whom cosmetic and auditory rehabilitation has been effected using in-the-ear hearing aids within prostheses overlying the rudimentary external auditory meati after canaloplasty. This novel method of auditory rehabilitation has not been reported before and is suitable in selected cases. The prostheses themselves were successfully secured using a prosthetic contact adhesive we have developed that offers excellent retention, little if any skin reaction, and high patient acceptability.

Key words: Hearing Aids, Maxillofacial Prosthesis; Tissue Adhesive

Case report

An otherwise healthy male neonate presented with bilateral microtia, stenotic external auditory meati (Figure 1) and facial nerve paresis involving mainly the lower branches. His older sibling had identical congenital deformities, although there was no other family history. He was diagnosed as having branchio-oto-renal syndrome, a rare (1 in 40 000 live births) autosomal dominant disorder of high penetrance but highly variable expression. The features of this syndrome can include external, middle- and inner-ear abnormalities, branchial sinuses, fistulae or cysts, renal anomalies and other associated abnormalities including facial nerve paralysis.¹

He was assessed as a neonate by a multidisciplinary team including paediatrics, otolaryngology, plastic surgery, audiology and maxillo-facial prosthetics. Computed tomography (CT) showed no cochlear abnormalities, a stenotic external auditory meati approaching abnormal, poorly pneumatized mesotympani having two ossicles and a rudimentary tympanic membrane. Brain-stem evoked response audiometry (BSERA) estimated a threshold of 70 dB, with a suprathreshold reduction of latencies suggesting a primarily conductive deafness (later confirmed with pure tone audiometry). His initial hearing rehabilitation was effected using a bone conducting hearing aid held on with a headband. This worked very well and his early speech development was normal.

At one-year-old, the vestigial auricular hillocks were excised and he underwent a right canaloplasty. At 18 months of age, prosthetic auricles were made to be secured with prosthetic contact adhesive and the patient accepted wearing these very well. With the right canaloplasty being successful and trouble free, it was thought that an in-theear hearing aid could be moulded to the canal and prosthesis overlying it, in such a fashion that the aid could be fitted onto the prosthesis (i.e. be removable from it) and then the prosthesis-aid unit adhered onto the skin

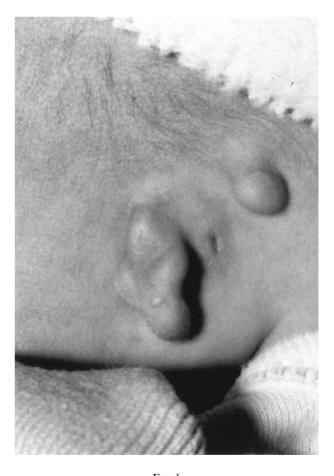


FIG. 1 Patient as neonate demonstrating microtia and stenotic external auditory meatus.

From the Department of Otolaryngology-Head and Neck Surgery, Hull Royal Infirmary, Hull, the Academic Surgical Unit*, University of Hull and Department of Maxillo-facial Prosthetics, St Luke's Hospital, Bradford, UK. Accepted for publication: 8 June 2001.





FIG. 3 Hearing aid and prosthesis shown in detail apart.

FIG. 2 Hearing aid/prosthesis *in situ*.

(Figures 2–4). This was achieved at two years of age and was immediately successful. Aided hearing thresholds, averaged from 500 Hz to 2 KHz, were 35 dB. Bone conduction thresholds, averaged 20 dB for the corresponding frequencies.

Over the next few years, he was closely monitored with audiometry, and speech and language development assessment. The latter was tested using the Reynell language development scales and showed progress commensurate with his age. At five year of age, he underwent a left canaloplasty. This had been delayed on account of the extreme stenosis of the canal. This again was successful and he now wears bilateral prostheses with in-the-ear aids within. His general, social and speech development has been normal. His speech and language development was further assessed at the age of seven years using the Test of Reception of Grammar (TROG) and British Pictorial Vocabulary skills, at which he scored above the 50th percentile for his age. He wears the prosthesis-hearing aid units everyday for all activities, including sports except swimming, without any retention problems at all or skin reaction to the adhesive.

Discussion

A prosthesis secured via osseo-integrated skin-penetrating implants has, in recent years, become popular for the cosmetic rehabilitation for patients with external ear loss, whether congenital or acquired.^{2,3} This has arisen because the results from surgical reconstruction tend to be disappointing.^{4,5} In the pre-osseo-integration era, the

results from using tissue adhesive to secure auricular and other prostheses gained a reputation for being sub-optimal, particularly because of poor retention, skin irritation, deterioration of the prosthesis (caused by the adhesive) and difficulties with positioning.^{4,6}

However, advances in the science of tissue adhesive technology have been made. One of us has developed a prosthetic contact adhesive that approaches the ideal, i.e. that it should have good bonding, be removable from skin and prosthesis (no build up), be non-irritating and have no deforming effects on the prosthesis itself.⁷ The adhesive is a dimethyl polysiloxanebase contact adhesive formulated for high spreadability and is compatible between the interface of skin and prosthetic material. It is readily removed with a companion formulated cleaner, but the adhesive remains effective without oxidization. It provides stability and secure fixation and has been used over a wide spectrum of facial and body prostheses.

In the population of patients from the West, North and East Yorkshire area that the Bradford maxillofacial prosthetic service serves, there are a number of patients who enjoy a high level of satisfaction with tissue adhesivesecured prothetic auricles. Most patients wear the prostheses all day every day, together with sporting activities including swimming in a number of patients. The results are such that we feel that this system is superior for most patients. Patients are able to receive and start wearing their adhesive-secured prostheses within a month of first being seen in the clinic. For paediatric patients, the prostheses can be introduced from around the age of 18 months. Generally most patients do not need regular clinic appointments but require a replacement prosthesis every year or so owing to 'wear and tear'. All our patients have the option for osseointegration and so far, none have chosen to convert since the introduction of the improved prosthetic contact adhe-

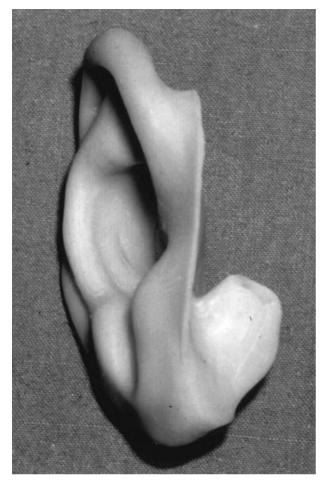


FIG. 4 Hearing aid and prosthesis shown in detail together.

sive. Although in large regional centres, the complication rate of surgery for osseo-integrated implants is low,⁵ this procedure is still invasive and can be associated with problems, particularly skin reactions and infection.⁸ In our experience, such problems as well as difficulty in maintaining the abutment has lead to patients' requests for removal of the implants (C Vize, personal communication; M Carroll, personal communication).

The placement of an in-the-ear conduction hearing aid in the prosthesis itself overlying the surgically-widened external auditory meati is, to our knowledge, a novel idea, and one that has worked well in this case. The feasibility of this system relied on successful canaloplasty and the presence of a rudimentary tympanic membrane and middle ear. The aided thresholds (35 dB) in a child with a bone conduction (BC) threshold of 0 dB represented adequate auditory rehabilitation. The published results of bone anchored hearing aids (BAHA), the normal form of auditory rehabilitation for patients with this type of congenital deformity,⁹ are marginally superior to this (aided thresholds of 20 dB with normal BC thresholds).¹⁰ BAHA is the gold standard in this situation and it is essential to closely monitor patients' progress with speech and language development if any alternative rehabilitation methods are employed.

Finally we endorse the view that the provision and choice of maxillo-facial prostheses and bone anchored hearing aids should be carried out in large regional centres in a multidisciplinary environment.⁵

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Address for correspondence:

Dr Jarrod J. Homer, B.Med.Sci., M.D., F.R.C.S., (ORL-HNS), Head and Neck Fellow, ENT Department, Princess Alexandra Hospital, Ipswich Road, Woollonggabba, Brisbane QLD 4102, Australia.

E-mail: jarrod@homer67.freeserve.co.uk

Dr J. Homer takes responsibility for the integrity of the content of the paper.

Competing interests: None declared.

Declaration of interest: Professor Roberts developed the prosthetic contact adhesive used in this case report