Advances In The Management Of Hearing Problems

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Introduction

The recent advances in the management of hearing problems are in three areas; firstly, early detection and diagnosis of hearing loss in children so that rehabilitation can be started whenever appropriate. Secondly, prostheses can be made from better tolerated material and employed for surgical reconstruction of the ossicular chain in the middle ear. Finally, the rehabilitation of those patients who have sensorineural problems can be enhanced by the programmable hearing aids. For the unfortunate patients who suffer from profound deafness, a cochlear implantation can be carried out to enable them to regain functional hearing. *(HK Pract 1996;18 (4): 141-146)*

序言：

最新的醫學研究對聽覺不靈患者有三大貢獻：
1) 對失聰兒童作出及早診斷，令復康計劃得以適時推行。 2) 用最新材料幫助重建中耳的「聽小骨」。 3) 「程序指令式的助聽器」可幫助「感覺神經耳聾」的病人。
最新的耳蝸移植手術更可令耳聾者如願得償，重拾聽覺的樂趣。

Hearing assessment

There is a general consensus that in infants and children with hearing loss, early intervention reduces the long-term communicative, social, and economic consequences[1,2]. Significant strides have also been made in the development of objective electrophysiologic techniques such as the otoacoustic emissions (OAEs) and auditory brainstem response (ABR).

Otoacoustic emissions

As early as 1948 it was thought that there was an active feedback mechanism within the cochlea which promoted signal processing by the cochlea for filtering, amplification and gaining control of sound signal[3]. This hypothesis was proven to be correct in 1978 when David T. Kemp discovered otoacoustic emissions (OAEs)[4]. OAEs are defined as vibrational energy that is generated in the cochlea and travels all the way through the middle ear structures, to be transduced as sound at the tympanic membrane (the reverse course of the normal sound conduction into the inner ear). Hence, specially designed equipment placed in the outer ear can pick up these spontaneous OAEs.

OAEs are believed to come from the outer hair cells of the cochlea with the intensities lying between 10 and 20dB sound pressure levels with frequencies ranging from 0.8 to 2.5kHz. There are great intersubject variations in both intensity and frequencies of the observed OAEs, but the signals from both ears of the same object are likely to be similar[5].

OAEs have been proven to be closely related to the functional status of the inner ear and are not detectable when hearing loss exceeds 40-50dB. Coupling the above with the fact that the procedure of detecting OAEs is noninvasive, it has become a useful objective screening test, especially for infants, children and people in whom it is difficult to carry out other tests. OAEs test for both ears can be completed in 10 minutes in a reasonably quiet room,

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a sound isolated chamber is not necessary[6]. Uncooperative children may be sedated to facilitate proper placement of the probe and a satisfactory result can be obtained.

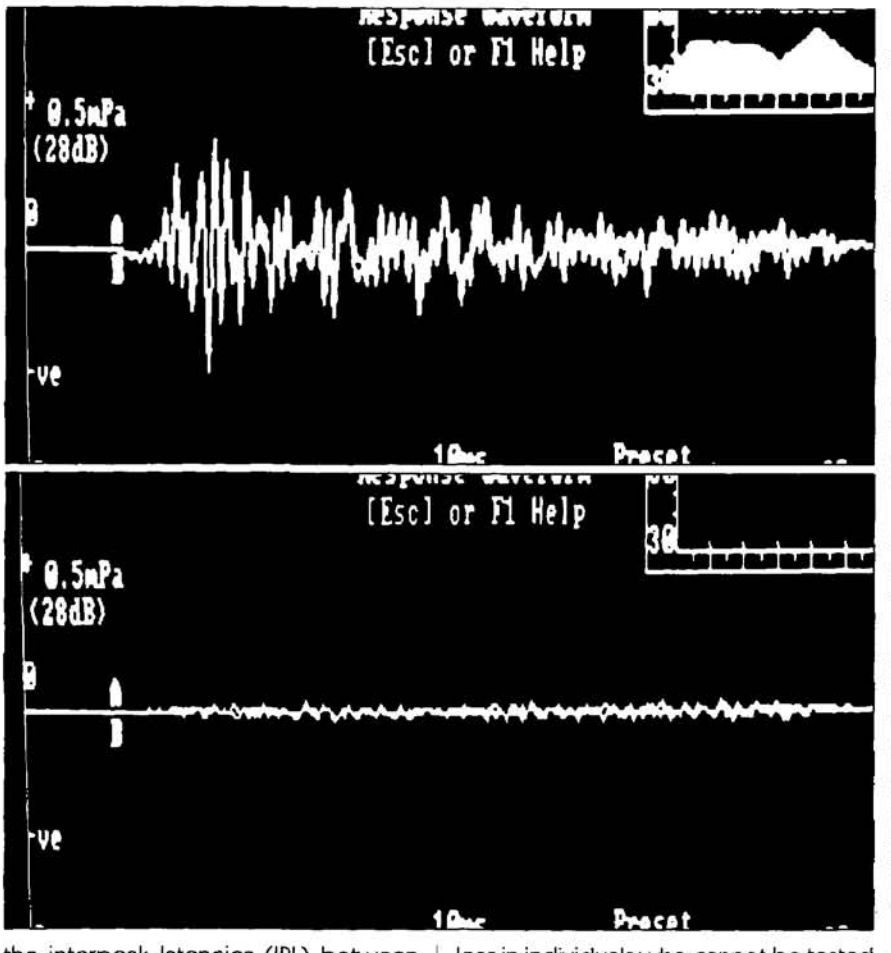
All the peripheral auditory structures are involved in the generation and transduction of OAEs and all these pathways are screened with the test. Mild to moderate hearing loss has been shown to significantly reduce OAEs and the results are not difficult to evaluate (**Figure 1**). OAE tests are thus probably the most sensitive auditory screening now available, although other studies are required to diagnose the exact pathology. The presence of OAEs excludes significant dysfunction of the peripheral auditory system, but normal OAEs may be detected in patients with retrocochlear pathology or CNS dysfunction[7]. OAEs should be used in conjunction with other tests, such as the ABR test to achieve a comprehensive auditory assessment.

Auditory brainstem response

Auditory brainstem response (ABR) testing was first reported in human in 1970[8] and it is now an established method of evaluating peripheral auditory function. Short-duration auditory stimuli in the form of clicks are presented to the ear under test and this results in the synchronous activation of the auditory pathway in such a way that the far-field potentials can be recorded from the scalp using a computerized averaging technique designed to detect small signals. ABR reflects the activity of the auditory pathway structures from the distal auditory nerve to the midbrain and has been found to have wide clinical application[9].

Amplitude measures of the ABR are not useful clinically due to their large intra- and intersubject variations. On the other hand, the latency measurements of the ABR wavepeaks are useful in the diagnosis of pathologies along the auditory pathway. The three latency measurements most frequently examined are the absolute latency of wave V; the interaural latency difference (IDL) between the ears of wave V; and the interpeak latencies (IPL) between waves I and III, I and V, and III and V. For example in patients with acoustic neuroma, the absolute latency of wave V is lengthened and IDL is even more sensitive and specific[10]. (**Figure 2**)

Figure 1: **Upper:** Normal otoacoustic emissions tracing
Lower: Diminished otoacoustic emissions indicates hearing loss



Recent advances in ABR testing include the application of frequency-specific stimuli to evaluate a distinct area of the basilar membrane[11]. This test is useful when a certain degree of hearing loss is suspected in a patient, or when a child 'failed' some form of screening test. The frequency-specific ABR testing is the most effective means of approximating the audiogram for peripheral auditory functioning. However, it cannot indicate whether there is an understanding of the auditory signal as ABR testing is limited to the evaluation of subcortical structures only.

ABR bone conduction testing is valuable for determining the presence of a conductive component in a hearing loss in individuals who cannot be tested with traditional methods. The bone oscillator may be placed on the mastoid or the forehead and as in behavioural bone conduction tests the response to the bone-conducted stimuli is biaural. Reliability of ABR bone conduction testing is comparable to that of air conduction test using clicks[12] and the ABR threshold for air-conducted and bone-conducted clicks are essentially equal. Thus, this electrophysiological air-bone gaps can be used to estimate the degree of conductive hearing loss in the same way as ordinary pure-tone audiograms[13].

Implants in middle ear surgery

One of the common causes of conductive deafness is the loss of the continuity of the ossicular chain. This can be related to trauma, the presence

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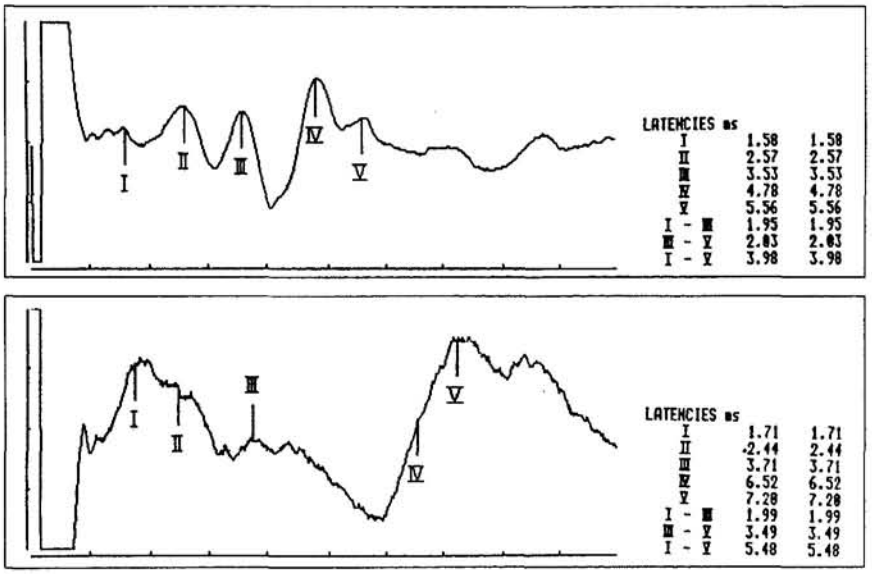
Figure 2: **Upper:** Normal auditory brainstem response tracing
Lower: Latency of wave V is lengthened in a patient with acoustic neuroma



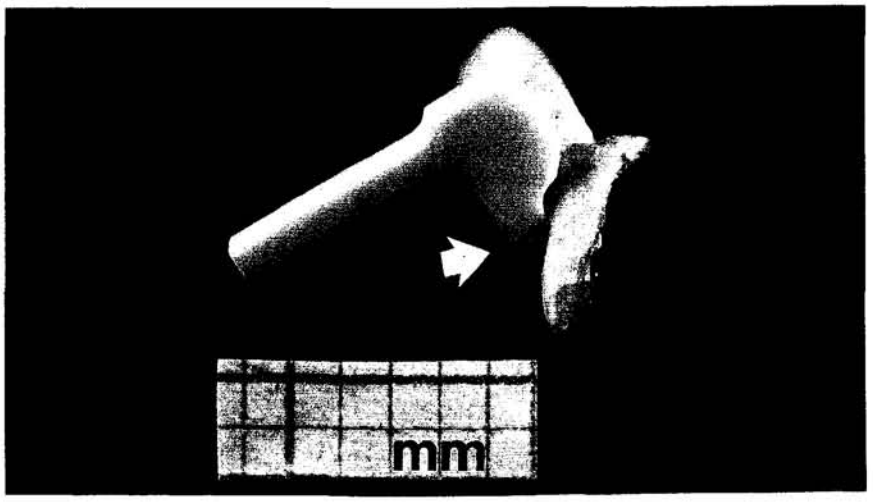
of cholesteatoma or necrosis of one of the ossicles due to infection. The incus is the ossicle most commonly affected by these processes. One of the themes of reconstructive surgery of the middle ear is to restore the continuity of the ossicular chain and most of the time it is concerned with the replacement of the incus or the suprastructure of the stapes. Initially, a short segment of polyethylene tube was used as it produces very little tissue reaction and its diameter permits it to fit one end onto the head of the stapes while the opposite end can be easily cut in a V shape manner to fit into the manubrium of the malleus[14]. The early and short-term effects of the reconstruction were astonishing and the patients often experience a dramatic improvement of hearing. However, as the polyethylene tube is in contact with the eardrum, it gradually erodes the tympanic membrane leading to extrusion and hearing loss. Other tissue such as the bone[15], cartilage[16] and a number of other materials such as Teflon, nylon, solid plastic and metal have all been employed and each is beset with its own problems including graft failure, implant extrusion and hearing loss[17-18].

The patient's own incus was first used for reconstruction of the ossicular chain in 1960[19] and homograft incus has been used with success[20]. This procedure involves the creation of a notch on the short process of the incus so that the manubrium of the malleus can be placed into the notch to reduce slipping of the prosthesis. The long process of the incus is then manipulated to fit onto the head of the stapes or the foot plate. Homografts used in this way have a high success rate and a low incidence of extrusion. The shortcomings of using cadaveric incus are that firstly each homograft has to be harvested by the surgeon and a lot of time is required. Secondly, with the recent AIDS epidemic there is a fear of transmission of the disease whenever homografts are used. Thus there is a need for an inert and low rejection incus prosthesis.

Hydroxylapatite is a calcium bioceramic that has the same chemical composition as living bone, $Ca^{10}(PO^4)^6(OH)^2$. Since 1970, it has been used as prostheses in other specialties such as orthopaedic surgery, plastic surgery and otolaryngology. Studies on both porous and dense hydroxylapatite in animals showed that all implants were integrated into the middle ear and were covered with normal mucosa[21]. There were no signs of extrusion. The application of these prostheses in human give similar results. For patients who had the hydroxylapatite implant and were re-explored later, the prosthesis was noted to be encased in a mucosal envelop, there was no evidence of granulation adhesion, softening or graft deterioration. The implant could be taken out modified and reinserted to give good functional results[22].

The few advantages with the hydroxylapatite incus prosthesis are that the notch on the body of the prostheses allows it to interlock with the manubrium of the malleus to prevent slipping (**Figure 3**). Secondly, the body of the prosthesis is in contact with the tympanic membrane, sound wave is directly disseminated from the eardrum through the implant to the stapes thus making the transmission of sound energy

Figure 3: The malleus sits over the notch of the incus Prosthesis (arrow)



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more efficient. Thirdly, the extreme biocompatibility of hydroxylapatite does not lead to any adhesions which may impede the movement of the implant. In fact, the false joint formed between the prosthesis and the other ossicles facilitates the motion of the implant and prevents fixation.

Programmable hearing aids

The first amplification system used by mankind is probably by putting the palm of the hand behind the ear and this has been shown to provide about 14dB of amplification at 1500Hz[23]. The basic principle of the contemporary hearing aid is as follows: the sound energy is picked up by a microphone and this is converted to electrical signals which correspond to the variation of the incoming sound energy and pressure. This electrical signals are then augmented by a transistor amplifier which is usually positioned together with the microphone. The signals are then amplified by the main amplifier and delivered to a receiver which converts the electrical signal back to sound. The gain of the amplifier can be modified with a volume control. The frequency response and the maximum output of the hearing aid can also be preset. These adjustments contribute to the trimming or potentiation of the input sound energy.

With the programmable hearing aid, the computer memory chip in the form of a memory module replaces the conventional trimmer and potentiometer function of a hearing aid so that a more varied or precise control is now possible[24]. The memory nodule responses to an external microprocessor which can access those memorylocations within the chip which represent different electrical acoustical performances. The patient carries the external microprocessor in the form of a remote controller and can change the performance of the hearing aid at different environment to obtain the most appropriate amplification of sound.

Cochlear implant

It has been known since 1800 that electrical stimulation can produce auditory sensations. This was demonstrated by Volta who inserted a metal rod into his ear and attached the rod to an electrical circuit[25]. This concept has been employed for the management of patients with profound sensorineural hearing loss. An electronic device can be implanted directly into the cochlea and this carries a coded electrical stimulus that bypasses the damaged or missing hair cell in the cochlea and directly stimulates the remaining auditory neural component such as the spiral ganglion cells or axons.

The modern cochlear implant has a microphone that collects external sound energy between 0.1 and 6kHz and converts it to an electrical signal. A processor modifies the signal and amplified to an acceptable level to match the narrow electric dynamic range of the ear. The processed signal is subsequently passed onto an external transmitter which then transmits this signal across the skin with electromagnetic induction or radiofrequency transmission to an implanted internal receiver. Electric current then flows along the active and return electrodes that have been implanted into the cochlea stimulating the auditory nerve, thereby producing the sensation of sound. The implanted electronic receiver is covered in hard Silastic and the electronic circuits are further sealed with a ceramic or titanium case. The active electrodes are insulated with Teflon and there has not been any documented rejection of the cochlear implant.

The 22-channel cochlear implant (Nucleus, Melbourne) is currently the most commonly used multi-electrode cochlear implant[26]. The latest system has a speech processor Spectra 22 which focuses on the coding of both spectral and temporal content. Filtered acoustic signals are divided into 20 frequency bands. The output are assigned tonnotopically to any of the twenty two electrodes. Upto 10 maxima can be selected and stimulation rate can then be optimised according to the number of maximum selected.

Patients who have undergone cochlear implantation require long periods of postoperative audiological training to obtain the maximum benefit. The implantees have to be motivated and have reasonable expectations. Strong family support is also essential. The selection of candidates for cochlear implantation involves professional input from various disciplines, social workers, speech therapists, audiologists, surgeons and in case of children, teachers of the deaf. In general, children aged 2 years or more with bilateral profound hearing loss and after at least 6 months training with properly prescribed hearing aids does not show any appreciable benefit, are suitable candidates. There should also be no medical contraindication to surgeryand the patient is highly motivated with the appropriate expectation of the outcome of the implantation. It is important that the patient, after implantation, be enrolled in a aural-verbal programme for further development of auditory skill.

In general, children who are postingually deafened show the greatest improvement in hearing. This is followed by children who are congenitally or deafened early in life and received cochlear implant in early childhood. Those patients who had an early onset of deafness but did not receive implant until later in adolescence had only a limited improvement in subsequent speech development[27,28].

From 1989 to July 1995, in the ENT unit of the Department of Surgery, The University of Hong Kong, in conjunction with the Hong Kong Society for the Deaf, we have performed a total of ten cochlear implantations. Seven patients were adults and three were children, their age ranged from 3 years to 45 years (median 36 years).

The operation of cochlear implantation was carried out under general anaesthesia. With a post-aural

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incision, a cortical mastoidectomy was carried out and the active electrode of the Nucleus 22 cochlear implant was then inserted through the round window into the cochlea via a posterior tympanotomy (**Figure 4**). The internal receiver connected with the active electrode was then fixed onto the mastoid bone (**Figure 5**). All the patients had a smooth postoperative course and were discharged home after removing the stitches at about seven days after the operation. Speech and audiological rehabilitation usually took place approximately 8 weeks after the operation (**Figure 6**)

Standard audiological and speech perception and discrimination assessments in all the ten patients showed marked improvement after the implantation. They were able to distinguish male or female voice, recognise open-set bisyllable words and discriminate close-set vowels and consonants. They could also comprehend every day sentence and environmental sound. Two of the implantees were able to use the telephone and carry out selective

Figure 4: Post-operative X-ray showing the electrode of the cochlear implant in the cochlear (arrow)

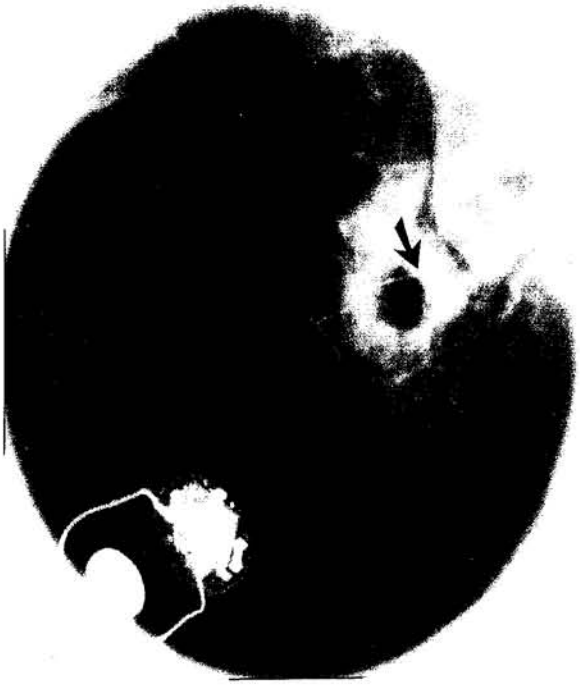

Figure 5: The internal receiver (R) are fixed onto the mastoid bone. The electrode (arrow) enters the cochlea through a posterior tympanotomy. All these will be covered with the scalp flap (S)

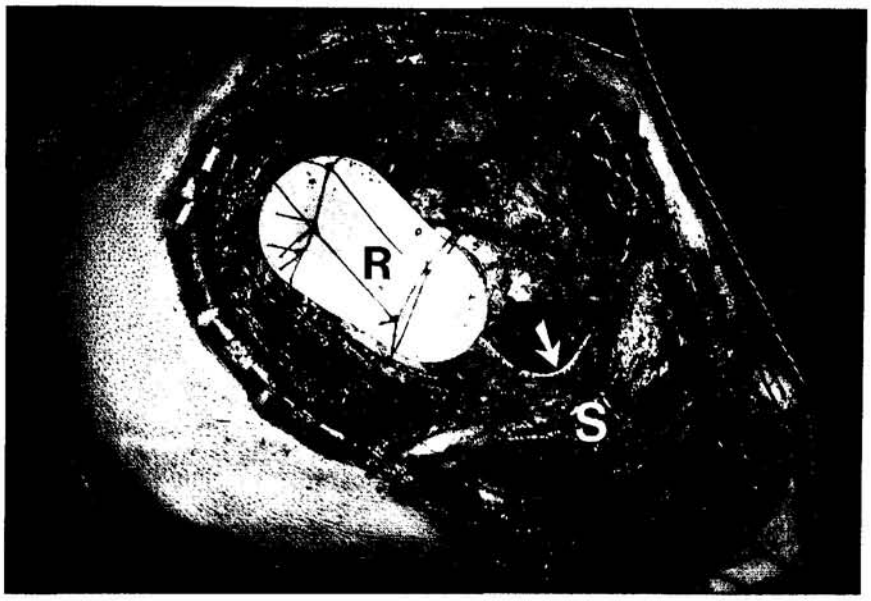

Figure 6: Patient wearing the speech processor (P) and the external transmitter (inset arrow)

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conversation. Despite the fact that Cantonese is largely a tonal language the patients were able to discriminate the six contrastive tones with significant results. Cochlear implant is versatile in its application to different language and has contributed significantly to the rehabilitation of the few Chinese patients with profound deafness. This gain is much more significant in children. ■

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