

Managing Hearing Loss in Children and Adults: Singapore Context

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Abstract

Hearing loss is common in Singapore, with as many as 360,000 persons affected. With Singapore's ageing population, this prevalence is expected to increase. Modern hearing devices have the potential to restore hearing in most patients. Cochlear implants can restore hearing in those with even profound to total hearing loss, by stimulating the auditory nerve directly without the need for viable cochlear hair cells. A holistic approach to managing deafness is necessary, including addressing psychosocial issues. Many of the problems experienced by the hearing-impaired are caused by the ignorance and lack of understanding from other people around him/her. "Reverse education therapy" refers to the effective education of these people, to directly benefit the hearing-impaired. In children, hearing restoration together with early detection and effective rehabilitation enable up to 80% to 90% of those born with significant hearing loss to cope well in mainstream education. The infrastructure to support them and facilitate their integration into mainstream society is necessary, and some of these are in place in Singapore. Leisure time noise-induced hearing loss in children is becoming increasingly important. In adults, societal support and understanding are just as important, particularly towards the elderly and pre-lingually deafened adults. Public education in Singapore is necessary, including that on the recognition of the abilities of successfully treated individuals, as well as that on showing more compassion, support and patience towards those who have not benefited sufficiently from treatment. In the future, there is potential for the clinical application of cochlear hair cell/auditory nerve preservation and regeneration.

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Introduction

In the management of hearing loss, advances in technology have led to more reliable screening tools, better diagnostic equipment and sophisticated hearing devices. These, in turn, have resulted in earlier detection and more effective restoration of hearing loss. Disability resulting from deafness affects not only the individual concerned, but also his/her family, friends and colleagues. As one of the commonest major disabilities, it has a great impact on society and is therefore an important public health concern. Besides the availability of medical expertise and hearing devices, overall management strategies must also include accessibility to these medical services, rehabilitation, education and social support.

Prevalence

Up to 6 in 1000 are born with sensori-neural hearing loss.¹ One in 1000 has a severe to profound condition.² This

makes it one of the commonest major congenital disabilities. Late-onset sensori-neural hearing loss occurs in about 15% to 20% of preschool children with sensori-neural hearing loss.³ Before the end of adolescence, 4 in 1000 of these children would have developed severe to profound hearing loss.² In Singapore, universal newborn hearing screening results showed that 3.8 per 1000 newborns have hearing loss. Therefore, assuming a birth rate of 40,000 per year, there are nearly 3000 young persons 19 years of age and below born with hearing loss in Singapore. This figure excludes many more with acquired hearing loss.

Prevalence estimates indicate that 1 out of 11 people in the general population are hearing-impaired.⁴ Given the population of 4 million in Singapore, there may be as many as 360,000 persons with hearing loss here. The prevalence is expected to increase in future because of the ageing population in Singapore. Singapore is the second fastest ageing nation in Asia after Japan,⁵ and persons over 65

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years of age will increase from 7% of the total resident population in the year 1999, to 18% in 2030. Age-related hearing loss is so common that about 50% of the population is expected to have significant hearing loss by 80 years of age.⁶

Causes of Hearing Loss

Congenital deafness can be due to genetic (about 50%), acquired (25%) or unknown (25%) causes.³ Of those due to genetic inheritance, 75% to 80% are autosomal recessive, 15% to 20% autosomal dominant, 1% to 2% x-linked and a few mitochondrial (maternal). The majority (70%) of genetic causes are non-syndromic. Non-genetic congenital deafness can be the result of prenatal infections like rubella and cytomegalovirus. Deafness developing later in life can also be genetic in nature or acquired, such as from diseases, illnesses, ototoxicity and trauma.

Deafness may be conductive, sensori-neural or both. Conductive deafness affects the sound-conducting mechanism, anywhere from the external to inner ear. Sensori-neural deafness is the result of damage to the cochlea (inner ear) or retro-cochlear pathways (auditory nerve and central nervous system).

Effects of Hearing Loss

Children

It is well recognised that hearing is critical to speech and language development, communication and learning. Hearing loss causes delay in the development of receptive and expressive communication skills (speech and language). The resultant language deficit also causes learning problems and reduced academic achievement. A study in Singapore revealed that children whose congenital hearing loss had been detected, and had received late intervention, generally fared more poorly in their Primary Six Leaving Examinations than their normal hearing peers.⁷

Adults

Most hearing-impaired people suffer certain social, psychological and physical problems as a result of their hearing loss. Psychological consequences may include embarrassment, frustration, depression, insecurity, low self-esteem and a lack of self-confidence. Social consequences include communication difficulties, inattentiveness and a lack of concentration. Communication difficulties often lead to social isolation, and also have an impact on vocational choices.

Modalities of Treatment

Medications

Some patients with conductive hearing loss resulting from middle ear infections recover after a course of

antibiotics and nasal decongestants. Sensori-neural hearing loss resulting from Meniere's disease may respond to diuretics, and that caused by syphilitic otitis may improve with antibiotics and steroids.

Surgical Restoration of the Physiological Hearing Mechanism

Conductive hearing loss can generally be rectified by appropriate surgery, so that the mechanical energy from sound can reach the inner ear, where it is converted to electrical energy by cochlear hair cells. For example, tympanic perforations can be repaired (myringoplasty) and the stapes in otosclerosis can be replaced (stapedectomy).

Surgery generally cannot restore physiological sensori-neural hearing, as the loss is usually a result of irreversible cochlear hair cell damage. An exception is sensori-neural hearing loss due to disturbed cochlear function from a perilymph fistula, where successful surgical repair can restore inner ear function. Surgery, however, can restore sensori-neural hearing in a non-physiological way, by means of a cochlear implant, which works by stimulating the cochlear nerve directly without the need for viable cochlear hair cells.

Hearing Aids and Assistive Devices

Modern digital hearing aids have added features and processing schemes previously not possible with analogue technology, making hearing amplification better and more comfortable.⁸ It is still a challenge, however, to develop hearing aids capable of enabling patients to understand speech in noisy environments. Besides better functioning, there is a trend towards increasing miniaturisation, which has progressed from body-worn aids, to aids worn behind the ear and in the ear canal. FM systems can be used to enhance the effectiveness of hearing aids, by improving signal-to-noise ratio.

Newer hearing aid technologies include those implanted into the middle ear. The potential advantages of implantable hearing aids are the elimination of feedback and the occlusion effect, as well as improved fidelity and cosmesis. There are also bone-anchored hearing aids, where the hearing aid is fitted onto a titanium screw, which bio-integrates with the mastoid bone. This type of hearing aid is useful for patients who are not able to wear a hearing aid in the ear canal, such as those with chronic ear infections or atretic ear canals.

Besides devices that amplify sounds, there are assistive tools which can help the profoundly deaf cope better in day-to-day activities. These include silent vibrating alarm clocks and doorbells with flashing lights.

Cochlear Implants

Some patients have cochlear hair cells so severely damaged that conventional hearing aids are inadequate. Such patients

may benefit from the cochlear implant, which does not rely on viable cochlear hair cells to work. The first adult and first child in Singapore were implanted in the Singapore General Hospital in 1989 and 1997 respectively. Since then, more than 200 patients have been implanted and managed by SGH. This includes adults as old as 80 years of age, and children as young as 1 year of age. A special type of hearing implant is the brainstem implant, which may be considered for patients with damaged auditory nerves.

Cochlear implants and other hearing devices can be costly. In 2001, the Ministry of Health, under its Health Service Development Programme, started a 4-year pilot project which subsidises up to 80% of the cost of a cochlear implant. There are other funds available in Singapore to help needy patients purchase these costly devices, such as the Assistive Technology Fund (Ministry of Community Development, Youth and Sports) and the Children's Medical Fund (National Kidney Foundation).

Holistic Approach to Treatment

Besides treating the cause of hearing loss and attempts at hearing restoration, other issues which contribute to the well-being of the patient have to be addressed. For example, factors such as parental concerns, educational issues and psychosocial effects need to be considered in a child who requires a cochlear implant. Even the psychological trauma of simply undergoing ear surgery should not be overlooked.

Many problems faced by the hearing-impaired are the result of the ignorance of family members, friends, peers, teachers and others around them. In order to fully and truly understand the hearing-impaired, these people must be effectively educated on the limitations faced by the hearing-impaired. This should include experiencing the sounds actually heard by the hearing-impaired, in simulated conditions. Merely directing therapy at the hearing-impaired is not enough. Effective education directed at people around the hearing-impaired results in therapeutic benefits to the hearing-impaired themselves, and it can aptly be referred to as "reverse education therapy".

Specific Issues in Children

Prevention

Prevention is always the best policy, even for congenital deafness. As maternal rubella is one of the most important causes of congenital deafness, the existence of a national rubella immunisation programme in Singapore has played a major role in preventing deafness. Over the last decade, a better understanding of the genes involved in deafness has led to more accurate genetic counselling and ultimately to more effective prevention of deafness.⁹

Historically, noise-induced threshold shift is not

considered a common cause of childhood hearing problems. However, with more frequent leisure time noise exposure, such as in discotheques, a significant number of young persons have noise-induced deafness.¹⁰ In USA, 12.5% of persons 6 to 9 years of age are estimated to have noise-induced hearing threshold shifts in 1 or both ears.¹¹ In Singapore, a study among National Service conscripts revealed a hearing loss prevalence of almost 40 in 1000, especially those with frequent exposure to loud noise.¹² Therefore, in today's context, hearing conservation education programmes for children should be considered.¹³

Early Detection

For optimal speech and language development, all infants with hearing loss should be identified by 3 months of age and should receive intervention by 6 months of age.¹⁴ In recent years, universal newborn hearing screening programmes have become available in public and private hospitals in Singapore where obstetric services are provided. Existing childhood hearing screening programmes carried out in schools, polyclinics and private clinics should continue, as they can detect delayed hearing loss. However, instead of conducting mass hearing screening for 7-year-olds in Primary One, it should be done earlier as it is now known that plasticity for speech and language development is before 5 to 6 years of age.

Early Intervention

After the hearing deficit in the child has been appropriately addressed, the child should be enrolled in a suitable rehabilitative program. In Singapore, 3 main rehabilitative approaches are available, namely auditory-verbal, auditory-oral and total communication. Singapore General Hospital's Listen & Talk Programme provides holistic management using the auditory-verbal approach to prepare the hearing-impaired child for mainstream education.

Mainstream Support

There are special schemes available in Singapore to provide support in mainstream schools and in the community. The Ministry of Community Development, Youth and Sports has the Early Intervention Programme for Infants and Children, a centre-based programme providing social, educational and rehabilitative services for special needs children of up to 6 years of age. In 2003, the Integrated Childcare Centre Programme was introduced, where 6 childcare centres have trained teachers to provide a curriculum tailored to the needs of these children while allowing them to engage in the same group activities as other children. Itinerant School Service is a community integration programme piloted by the Singapore Association for the Deaf, to help hearing-impaired children in schools.

Among all the major disabilities, congenital deafness is

one where modern medical intervention can arguably make the biggest difference to the lives of those affected. With appropriate modern holistic management, at least 80% to 90% of children born with significant deafness can potentially cope well in mainstream education, integrate into mainstream society successfully and contribute effectively to society.⁷ Public education is very much needed in this aspect.

Specific Issues in Adults

There are 4 types of hearing loss in the adult population of Singapore which deserve special mention:

Noise-induced Deafness

Loud noise of 85 dB for more than 8 hours per day over prolonged periods of time can result in noise-induced deafness. In Singapore, where legislative procedures are in place to ensure that cases of noise-induced deafness are reported to the Ministry of Manpower, employers take the necessary preventive measures and workers are compensated for noise-induced deafness.¹⁵ Noise-induced deafness is the leading occupational disease in Singapore, with over 500 new cases detected by the Department of Industrial Health yearly.¹⁶

Patients with Nasopharyngeal Carcinoma

In this part of the world, nasopharyngeal carcinoma is common among Chinese adults. Nasopharyngeal carcinoma can present as hearing loss due to middle ear effusion, which the primary health physician must be mindful of. As radiotherapy is the primary modality of treatment and because the ear lies in the radiation fields, hearing loss in patients who have undergone radiotherapy is common. Radiation-induced hearing loss can be both conductive or sensori-neural in nature.¹⁷ Conductive hearing loss is usually due to middle ear effusion, which is the result of eustachian tube damage. Sensori-neural hearing loss occurs if the cochlear hair cells or retro-cochlear auditory pathways are damaged by radiation.

Age-related Hearing Loss (Presbycusis)

Hearing loss is the third most prevalent chronic condition in older adults and has important effects on their physical and mental health.¹⁸ Acceptance of hearing aids and hearing rehabilitation by the elderly is generally poor.¹⁹ In Singapore, almost 70% of the elderly who need hearing aids are not keen on having them, although 40% experience negative psychosocial effects as a result of the handicap.²⁰ As the adverse effects are contributed to a large extent by ignorance on the part of family members, peers, professionals and the public at large, the education of these people is important. In the primary healthcare setting, it is important to realise that besides age-related sensori-neural hearing loss, there

are many other forms of hearing loss e.g., ear wax and chronic middle ear infections, for which effective treatments exist.

Pre-lingually Deafened Adults

These are adults who are born deaf or are deafened before the acquisition of speech and language. In the past, certain technologies, which might have enabled them to hear enough to develop speech and language, may have been unavailable. Even if such technology had been available, there would invariably have been some who would not have derived significant benefit from it. They therefore generally communicate by total communication (including sign language). As they have exceeded the critical age of brain plasticity for the development of speech and language, the benefit they can potentially derive from current medical technology is limited. It is important that society does not forget this group, and helps to facilitate their integration into the community. Education is needed for the public to show more compassion, patience and understanding towards their disabilities.

Future Directions

The technology used in various aspects of managing deafness will continue to advance rapidly. Better technology will lead to more accurate screening and diagnostic tools, as well as improved hearing devices. For example, hearing aids are expected to provide clearer speech perception in background noise, better binaural hearing/localisation of sounds and become smaller, with longer battery life.

In cell and molecular biology, the regeneration/repair/protection of auditory hair cells and nerves are exciting and rapidly evolving fields.²¹ Gene therapy has been shown to be effective in providing long-term support to auditory hair cells. During the last few years, much progress has been made in the molecular basis of presbycusis, with the aim of instituting preventive therapies in the elderly.²²

Conclusion

Hearing loss is common in Singapore, both in children and adults. With an ageing population, more people are expected to experience hearing loss in the near future. A holistic approach to the effective management of hearing loss is necessary. This includes not only the restoration of hearing, but also addressing other aspects such as prevention, early detection, rehabilitation and psychosocial issues. As a significant part of the problems faced by the hearing-impaired is caused by the ignorance and lack of understanding of those around him/her, the effective education of these people is important. Although some of the social infrastructure required to support the hearing-impaired in the community is in place in Singapore, more public education is needed. In particular, the public should

recognise the ability of successfully treated individuals, and show more compassion, support and patience towards those who have not adequately benefited from treatment. In the future, there is potential for the clinical application of cochlear hair cell/auditory nerve preservation and regeneration.

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QUESTIONS

1. In Singapore, the following statements are true:
 - a) 6 in 10,000 are born with hearing loss
 - b) Nearly 3,000 persons below 19 years of age suffer from hearing loss since birth
 - c) There are about at least 350,000 persons suffering from hearing loss
 - d) By 80 years of age, about 50% will experience significant hearing loss
2. The following statements about the causes of deafness are true:
 - a) In congenital deafness, about 50% are genetic in origin
 - b) The majority of congenital causes is syndromic in nature
 - c) Deafness can be classified as conductive, sensori-neural or mixed
 - d) Sensori-neural deafness are due to cochlear or retro-cochlear causes
3. In the treatment of deafness
 - a) Many types of conductive deafness can be rectified by surgery
 - b) Cochlear implantation is indicated in patients with profound deafness due to cochlear damage
 - c) “Reverse education therapy” refers to effective education directed at people around the hearing-impaired
 - d) Future treatment modalities may include hair-cell regeneration and gene therapy
4. In childhood hearing loss
 - a) Immunisation programmes are not effective preventive measures
 - b) Children born with hearing loss should be detected by 3 months and intervened by 6 months of age for optimal speech and language development
 - c) Universal newborn hearing screening programmes are recommended for early detection of congenital deafness
 - d) With early detection and effective early intervention, at least 80% of these children can cope well in mainstream education
5. In Singapore, the following statements about hearing loss in adults are true:
 - a) Noise-induced deafness has been a leading occupational disease in Singapore
 - b) Patients with nasopharyngeal carcinoma can present with hearing loss and often suffer from deafness after radiotherapy
 - c) In helping pre-lingually deafened adults, effective education of family members, friends, professionals and the public is essential
 - d) In elderly patients with deafness, most are keen to have hearing aids

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