

SCREENING OF HEARING IMPAIRMENT AND ASSOCIATED EFFECTS ON ADAPTIVE BEHAVIOUR IN ADULTS WITH DOWN SYNDROME

V. P. Prasher

Introduction

High prevalence rates of hearing impairment in adults with learning disability (LD) (Kropka and Williams, 1986; Stein *et al.*, 1987; Yeates, 1989) and in children with Down syndrome (DS) (Dahle and McCollister, 1986, Roizen *et al.*, 1993) have been reported. Recent studies investigating hearing impairment in adults with DS are given in TABLE I. Overall, these studies show that hearing impairment occurs in the majority of people with DS, up to 95% of ears tested (Evenhuis *et al.*, 1992). The cause may be conductive, sensorineural or a combination of both types.

Although measures used to assess hearing impairment (audiometry, tympanometry) have generally been similar, the criteria for hearing impairment, age of sample assessed, institutionalised or community residents, and use of matched individuals have led to

varying prevalence rates being reported. Further, cooperation of persons with testing is an important source of variability. Yeates (1992) found one-third of her sample were unable to comply with subjective testing. In some of those that underwent objective testing general anaesthesia had to be given.

Supplementing the medical findings, several researchers have investigated the consequential impact of hearing impairment on the person's level of functioning. Individuals with impaired hearing have been shown to have impaired social functioning (Wright *et al.*, 1991; Lonigan *et al.*, 1992) and hearing impairment may be a factor in the decline in cognitive functioning (Libb *et al.*, 1985; Saxon and Witriol, 1976). This latter point is highly relevant to the possible mis-diagnosis of dementia in individuals with DS (Prasher and Krishnan, 1993).

* Dr. V. P. Prasher, M.B.Ch.B., M.Med.Sc., M.R.C.Psych.

Lecturer in Psychiatry, Department of Psychiatry, University of Birmingham, Queen Elizabeth Psychiatric Hospital, Mindelsohn Way, Edgbaston, Birmingham, B15 2QZ, United Kingdom

Fax No.: UK 0121-627 2832

* For Correspondence

TABLE I
Prevalence studies investigating audiological abnormalities in people with Down syndrome

Authors	Sample Size	Age Group	Control Group	Measure Used	Findings
Brooks <i>et al.</i> (1972)	100	1-59 years	MR controls	Pure tone or speech audiometry/verbal speech	73% of DS and 25% of controls had hearing loss
Balkany <i>et al.</i> , (1979)	107	2 months-60 years	no	Pure tone speech audiometry	Hearing loss in 78% of ears tested. 83% conductive and 17% sensorial
Keiser <i>et al.</i> , (1981)	51	15-51 years	no	Pure tone audiometry/ tympanometry/verbal speech	74% hearing loss. 32% conductive, 45% sensorineural & 23% mixed
Evenhuis <i>et al.</i> , (1992)	35	35-62 years	no	Impedance or pure tone or evoked response audiometry	95% of ears tested demonstrated impairment
Yeates (1992)	77	adults	MR controls	Audiometry/distraction tests/Brainstem Evoked and Post Aural Evoked Responses	43% had hearing loss requiring a hearing aid. 64% had mixed loss

MR = mental retardation

Prevalence of hearing impairment is likely to be high in elderly adults with DS, but this still needs to be confirmed. Age related prevalence rates have not been established. The possible detrimental effects of the presence of cerumen on hearing need to be confirmed. The possible effects of hearing impairment on adaptive functioning require further investigation. This research study investigates these important areas of interest.

Methodology

Adults with DS, aged over 16 years, involved in a large longitudinal study investigating healthcare for adults with

DS participated in this study. The DS cohort comprises hospital and community residents, living in a wide geographical region with a wide age range of individuals. Clinical and cytogenetic studies were previously undertaken to confirm all individuals had DS. Severity of LD was assessed by reviewing previously reported intelligence test results, previous level of functioning determined by review of medical notes, from carer interview and from mental state examination of the individual. Severity of LD was classified using ICD-10 criteria (WHO, 1992).

Hearing was assessed using information from several sources; primary carer, response to whisper speech and

response to distraction (response to audible speech from behind whilst attention focused on carer). The latter tests were repeated on three occasions. Test response was graded from no impairment (response to all tests), borderline impairment (response >50% of tests), moderate impairment (response <50% of tests) and severe impairment (no response to testing). Otolaryngological examination for presence of cerumen was undertaken after the hearing assessment and with excess cerumen (cerumen group) diagnosed if the tympanic membrane of both ears was not visible. If one or both membranes could be seen individuals were classified as having no excess cerumen (no cerumen group). Therefore, in this study "bedside screening" was undertaken to assess the prevalence of "functional" hearing loss rather than using instruments previously used to measure "physiological" loss.

Each individual underwent a detailed medical examination and a psychiatric examination for psychiatric disorders. Adaptive behaviour was assessed using the Adaptive Behaviour Scale (Nihira, 1974) (ABS). The ABS is a widely used scale with Part I domains assessing independent functioning and Part II assessing maladaptive behaviour. The domain for medication was excluded from Part II. Overall mean total scores for the DS group with impairment of hearing were compared to the scores for the age-LD matched DS group living in the same type of residence with no hearing impairment and no other significant medical or psychiatric pathology (eg. hypothyroidism, visual loss, dementia, depression). Groups

were divided into below and above the age of 40 years. Similar comparison was made for maladaptive behavioural problems using Part II of the ABS. Statistical analysis was performed using analysis of variance, t test and Chi-squared.

Results

Two-hundred and one adults participated in this study, sex distribution was 102 (50.7%) male and 99 (49.3%) female. The mean age of the sample population was 42.22 years (standard deviation [SD] 12.51; standard error [SE] 0.88). The minimum age for the total sample was 16 years and the maximum age 76 years. Over 110 individuals (55%) were aged 40 years and over, and 17 (8.4%) were over 60 years of age.

The majority of the individuals were living in the community, with 73 (36.3%) living in supervised community units and 85 (42.3%) living in their family homes. Forty-three (21.4%) were resident in the hospital setting. Thirty-eight (18.9%) individuals had mild LD, 134 (66.7%) had moderate and 27 (13.4%) had severe impairment, and two were unknown.

Eight individuals were uncooperative with audiological testing. Of the remaining 193 (96%) individuals over two-thirds had no detected hearing impairment (TABLE II). Twenty-three (11.9% tested) had moderate or severe clinical impairment. A significant association was found between hearing impairment and increasing age (TABLE III; ANOVA; F ratio= 6.3; P<0.001).

TABLE II
Hearing Acuity Findings in Sample

Finding	Frequency N	Percentage %
No impairment	138	68.7
Borderline impairment	32	15.9
Moderate impairment	20	10.0
Severe impairment	3	1.5
Not examined	8	3.9
TOTAL	201	100

TABLE III
Mean Ages for Differing Hearing Acuities

Hearing Assessment	Frequency	Mean Age (SD) Years
No impairment	134	40.0 (11.6)
Borderline impairment	32	47.7 (11.5)
Moderate impairment	20	48.0 (11.8)
Severe impairment	7	47.7 (11.5)
TOTAL	193	

Excess cerumen was found in the ears of 38 (20%) of 186 individuals examined. There was a statistically significant association between presence of cerumen and increasing age. The mean age for the no cerumen group (N = 148) was 41.2 years (SD 12.3) and for the cerumen group (N = 38) was 46.5 years (SD 10.7), (t test = 2.44; P<0.05). There was also a significant association between the presence of excess cerumen and hearing loss (Chi-squared test, Pearson's value = 15.1, P<0.001).

The detrimental effect of hearing impairment on adaptive functioning for the 2 groups (age below 40 years and 40 years and over) is shown in FIGURE 1. For both groups persons with hearing impairment scored significantly lower on the assessment of independent adaptive behaviour than persons with no hearing impairment. For the groups below the age of 40 years, individuals with hearing impairment also had significantly greater maladaptive problems (FIGURE 2).

FIGURE 1
Independent (Part I) Scores for Down syndrome groups with and without hearing impairment

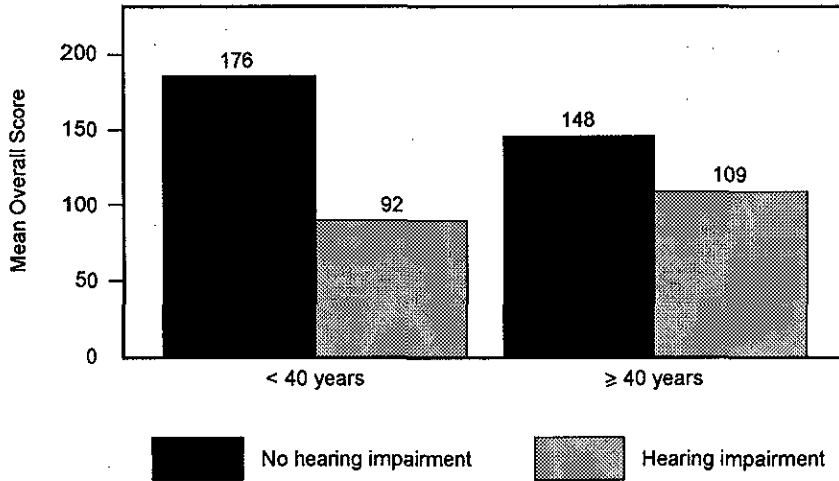
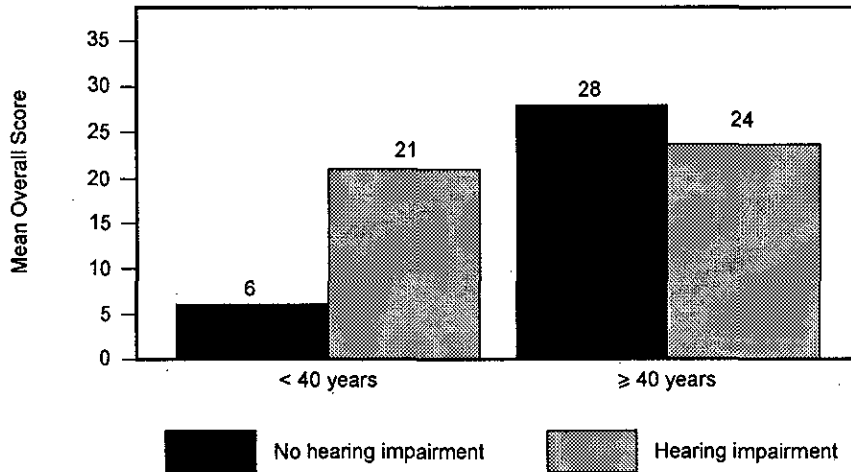


FIGURE 2
Maladaptive Behaviour (Part II) Scores for Down syndrome groups with and without hearing impairment



Discussion

Significant clinical hearing impairment was found in 12% of the sample tested, with evidence of some degree of impairment in 28% of the sample. These rates are lower than those previously reported, 43-95% (TABLE I). However, previous studies have used audiometry and tympanometry as measures. Such techniques are appropriate for diagnosis and detect early changes of hearing impairment. Although the rate in the present study was found to be lower than in other studies, and probably underestimated the exact prevalence, it highlights a high prevalence of hearing impairment in adults and especially in older adults with DS. In this study emphasis was put on functional hearing impairment rather than physiological impairment. Further physiological studies, using audiometry and tympanometry are envisaged to validate these functional findings.

As in the non-LD population, an association between hearing impairment and increasing age was found for this DS population ($P < 0.001$). An association between hearing loss and presence of excess cerumen was confirmed by this study ($P < 0.001$) as was an association between presence of cerumen and age ($P < 0.05$).

Hearing impairment leads to a loss of adaptive skills for all ages and to an increase in maladaptive behaviour for individuals below the age of 40 years. This confirms earlier concerns that deterioration in adaptive functioning cannot be assumed to be due to dementia in

people with DS unless the presence of hearing impairment has been excluded.

It is important that physical and psychiatric morbidity is recognised early in people with DS and subsequently managed correctly. Regular "bedside screening" for hearing loss is essential with subsequent referral for further audiological assessment if necessary. Early diagnosis will enable individuals to communicate to the best of their ability, to function in society to their best potential and prevent loss of adaptive skills. This study highlights the need for such regular routine screening.

Summary

Although an increase in the prevalence of hearing impairment in adults with Downs syndrome is accepted, satisfactory research data is lacking. This study investigated hearing impairment in 201 Down syndrome adults. Twenty-eight percent had some evidence of hearing impairment with 12% evidence of moderate to severe impairment. Hearing impairment was found to be related to age and the presence of cerumen. The detrimental effects of impaired hearing acuity on adaptive behaviour were demonstrated. Recommendations for healthcare services are discussed.

References

- Balkany, T.J., Downs, M.P., Jafek, B.W. and Kraticek, M.J. (1979). Hearing loss in Down's syndrome. *Otolaryngology - Head and Neck Surgery*, 87, 372-384.
- Brooks, D.M., Wooley, H. and Kanjilal, G.C. (1972). Hearing loss and middle ear disorders in patients with Down's syndrome (mongolism). *Journal of Mental Deficiency Research*, 16, 21-29.
- Dahle, D. J. and McCollister, F. P. (1986). Hearing and otologic disorders in children with Down syndrome. *American Journal of Mental Deficiency*, 90, 636-642.
- Evenhuis, H. M., van Zanten, G. A., Brocaar, M. P. and Roerdinkholder, W.H.M. (1992). Hearing-loss in middle-aged persons with Down syndrome. *American Journal on Mental Retardation*, 97, 47-56.
- Keiser, H., Montague, J., Eold, D., Maune, S. and Patterson, D. (1981). Hearing loss of Down syndrome adult. *American Journal of Mental Deficiency*, 5, 467-472.
- Kropka, B. I. and Williams, C. (1986). The epidemiology of hearing impairment in people with mental handicap. In: Ellis D. Ed. *Sensory impairment in mentally handicapped people*. London: Croom Helm, 35-60.
- Libb, J. W., Dahle, A.J., Smith, K., McCollister, F.P. and McLain, C. (1985). Hearing disorders and cognitive function of individuals with Down syndrome. *American Journal of Mental Deficiency*, 90, 353-56.
- Lonigan, C.J., Fischel, J. E., Whitehurst, G. J., Arnold, D. S. and Valdez-Menchaca, M. C. (1992). The role of otitis media in the development of expressive language disorder. *Developmental Psychology*, 28, 430-440.
- Nihira, K., Foster, R., Shellhaas, M. and Leyland, H. (1974). *AAMD Adaptive Behaviour Scale, 1974 Revision*, Washington, D. C.: American Association on Mental Deficiency.
- Prasher, V.P. and Krishnan, V.H.R. (1993). Mental disorders and adaptive behaviour in people with Down's syndrome. *The British Journal of Psychiatry*, 162, 848-849.
- Roizen, N. J., Walters C., Nicol, T. and Blondis, T.A. (1993). Hearing loss in children with Down syndrome. *The Journal of Pediatrics*, 123, S9-S12.
- Saxon, S. A. and Witriol, E. (1976). Down's syndrome and intellectual development. *Journal of Pediatric Psychology*, 1, 45-57.
- Stein, L. K., Kraus, N., Ozdamar, O., Cartee, C., Jabaley, T., Jeantet, C. and Reed, N. (1987). Hearing loss in an institutionalised mental retarded population. *Archives of Otolaryngology-Head and Neck Surgery*, 133, 32-35.
- World Health Organisation (1992). The ICD-10 Classification of Mental and Behavioural Disorders. *Clinical Descriptions and Diagnostic Guidelines*. Geneva: WHO.
- Wright, P.F., Thompson, J. and Bess, F.H. (1991). Hearing, speech, and language sequelae of otitis media with effusion. *Pediatric Annals*, 20, 617-621.
- Yeates, S. (1989). Hearing in people with mental handicaps; a review of 100 adults. *Mental Handicap*, 17, 33-7.
- Yeates, S. (1992). Have they got a hearing loss? A follow-up study of hearing in people with mental handicaps. *Mental Handicap*, 20, 126-133.