Language and Communication in Adults With Down Syndrome and Dementia of the Alzheimer Type: A Review

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Abstract

Language problems are one of several key diagnostic clinical features of dementia of the Alzheimer type (DAT). There is a large empirically derived literature describing the changes in language and communication in individuals with DAT over the course of its relentless progression. Relatively less is known, however, about the language and communication changes associated with the onset and evolution of DAT among individuals with Down syndrome (DS). The purpose of this brief review is to present what is known currently about the language and communication of adults with DS and DAT. The review concludes with the call for additional experimental research and multi-perspective observational studies of the unique changes in language and communication in individuals with DS and DAT.

It is widely accepted that individuals with Down syndrome (DS) over 40 years of age are at great risk for developing dementia of the Alzheimer type (DAT). Zigman, Schupf, Sersen, and Silverman (1995) showed that individuals with DS are significantly more likely to have DAT than are individuals with other developmental disabilities. Estimates of the percentage of individuals with DS who have DAT range from 4% in the community (Devenny, Silverman, Hill, Jenkins, Sersen & Wisniewski, 1996) to 88% in institutional settings (Evenhuis, 1990). Other estimates of the prevalence of DAT in individuals with DS indicate 3.4% in the 30 to 39 year old group, 10.3% in 40 to 49 year olds, and 40% in the 50 to 59 year cohort (Holland, Hon, Huppert, Stevens & Watson, 1998). Malamud (1964; 1972) first reported that one hundred percent of autopsied brains of individuals with DS over the age of 40 present with the neuropathological changes associated with DAT, a finding confirmed by others (Visser,

Aldenkamp, van Huffelen, Kuilman, Overweg, & van Wijk, 1997; Whalley, 1982; Wisniewski, Wisniewski, & Wen, 1985). Defining the clinical profile of dementia in adults with DS is a challenge, despite the neuropathological evidence showing the presence of DAT in adults with DS (see summary by Mann, 1993). Diagnosing DAT in adults with DS is difficult due, in part, to the underlying developmental cognitive and language problems, to possible hearing and vision impairments, to potential thyroid dysfunction, to the presence of depression and co-existing psychiatric disorders, and to the heterogeneity of baseline cognitive, language, psychosocial, and behavioural skills (Burt, Loveland, & Lewis, 1992; Dalton, Seltzer, Adlin & Wisniewski, 1993; Evenhuis, van Zaten, Brocaar, & Roerdinkholder, 1992; Lai, 1992; Lai & Williams, 1989; Lowe & Temple, 2002).

Cognition, DS and DAT

Evidence from neuropsychological investigations supports the position that DAT does occur in adults with DS who show specific impairments in visuospatial planning and working memory (Crayton, Oliver, Holland, Bradbury, & Hall, 1998; Devenny, Krinsky-McHale, Sersen & Silverman, 2000; Haxby, 1989; Roeden & Zitman, 1997). Moreover, data suggest that cognitive changes in adults with DS are not necessarily a function of age-related declines (Jordens, Evenhuis, & Janssen, 1997). Diagnostic criteria for DS and DAT now exist based on the ICD-10 (WHO, 1990), recognized diagnostic methods (Aylward, Burt, Thorpe, Lai, & Dalton, 1995), and practice guidelines for clinical assessment and care (Janicki, Heller, Seltzer, & Hogg, 1996). These criteria include the presence of progressive cognitive and language changes.

Language, Communication, DS and DAT

It is well known that language and communication changes are among the earliest symptoms marking the onset of DAT in adults. The Diagnostic and Statistical Manual of Mental Disorders 4th Edition (DSM-IV) (American Psychiatric Association, 1994) and the National Institute of Neurological and Communicative Disorders and Stroke and Alzheimer's Disease and Related Disorders Association (NINCDS-ADRDA) (McKhann, Drachman, Folstein, Katzman, Price & Stadlan, 1984) research criteria and Evenhuis's (1992) screening instrument for DAT include the early presence of "aphasia" (sic). There is wide-recognition and acceptance among scientists, clinicians, and family and professional caregivers that language and communication problems mark the onset of DAT, change precipitously over its course, and

create undue emotional, social and functional strains on those with DAT and those who provide for their care (e.g., Bayles, Tomoeda & Trösset, 1992; Orange, 1991). Notwithstanding our knowledge of the language and communication in DAT, we are only beginning to understand how the language and communication of adults with DS is affected by the onset and progression of DAT and how it differs from the profiles observed in individuals with DS only.

Comparative studies of language in DS. The language of adults with DS often is described relative to the performance of children, adolescents and young adults with DS, or to adults with developmental disabilities of other origins (Campbell-Taylor, 1993; Chapman, Seung, Schwartz & Bird, 2000; Cooper & Collacott, 1995; Rasmussen & Sobsey, 1994; Rondal, 1988; Sabsay & Kernan, 1993). Overall, these studies showed that expressive language can remain relatively stable or decline marginally with age (Carter-Young & Kramer, 1991; Copper & Collacott, 1995), and can reflect skill levels of younger adults with DS (Copper & Collacott, 1998). It is generally acknowledged, though, that the expressive language of adults with DS is characterized by reduced mean length of utterance (i.e., short sentences), auxiliary verbs errors (e.g., 'be' and 'have'), fractured word order (i.e., syntax), and simplification of grammar (Rondal, 1988; Rondal & Lambert, 1983; Sabsay & Kernan, 1993). Adults with DS exhibit problems repeating sentences containing elementary grammar and syntax, and carrying out one-, two- and three-step commands (Ashman, 1982; Kernan, 1990). Functional neuroimageing positron emission tomography (PET) data show significant reductions in cerebral glucose metabolism in primary language areas (i.e., left superior temporal and left inferior frontal) of young adults with DS compared to healthy age- and sex-matched controls (Azari, Horwitz, Pettigrew, Grady, Haxby, Giacometti, & Schapiro, 1994), suggesting age-related changes in neurolinguistic competence.

Language, ageing and DS. Recent studies that examined the effects of ageing on language in adults with DS show that receptive vocabulary is stronger than auditory comprehension of syntax, and that receptive language is weaker overall than expressive language (Carter Young & Kramer, 1991; Cooper & Collacott, 1995; Rasmussen & Sobsey, 1994). Moreover, speaking rate slows with ageing among adults with DS (Das & Mishra, 1995). Haxby (1989) and Rasmussen and Sobsey (1994), however, did not find age-related differences in cross-sectional analysis of language between young adults with DS and older adults with DS only. Longitudinal analysis of DS adults over 40 years of age indicate significant losses in communication skills, particularly in receptive language (Collacott &

Cooper, 1997; Haxby, 1989; Rasmussen & Sobsey, 1994). Studies also showed the onset of dyspraxia, a central nervous system motor sequencing disorder that affects speech production and can negatively influence accurate gesture use (Dalton & Fedor, 1998; Haxby, 1989). Carter Young and Kramer (1991) found a significant association between advancing age and comprehension of spoken language and a similar association between advancing age and verbal expression in their study of age-related language decline in adults with DS. Specific receptive language deficits included a diminished ability to attend to auditory stimuli, poor word discrimination, poor comprehension of the meaning of spoken language, and reduced ability to follow verbal directions. The investigators questioned whether these declines in language are the result of the onset of DAT or whether they are part of a normal ageing process particular to DS. Similarly, Cooper and Collacott (1995) found that receptive language abilities in individuals with DS 40 to 49 years of age were significantly lower than the receptive language skills of younger individuals with DS.

Language, communication in adults with DS-DAT. Comparisons of the language of adults with DS and DAT versus adults with DS only are limited primarily to case studies or to pilot investigations. Dalton and Crapper-McLachlan (1986) reviewed studies of the clinical manifestations of DAT in individuals with DS and found little information regarding "… language-based dysfunctions in DS studies of dementia." (p. 662). A case study of oral confrontation naming of pictures in an individual with DS and dementia showed significant longitudinal declines over a 20-month period (Kledaras, McIlvane, & Mackay, 1989). Naming performance and error patterns were consistent with those described in individuals with DAT (i.e., semantically empty terms – "thing" and "stuff", category label use rather than category specific exemplars – "bird" vs "cardinal", and visually related items – "ball" for "orange"). The authors concluded that confrontation naming tests can be useful in documenting and characterizing the progression of DAT in individuals with DS. Two participants with DS and dementia in Haxby's study (1989) exhibited naming scores at the lower end or below young participants' range and significantly lower verbal comprehension than adults with DS only. Four individuals with DS and postmortem confirmed DAT exhibited declines in communication skills, based on caregiver descriptions (Rasmussen & Sobsey, 1994). Roeden and Zitman (1997) found significant longitudinal relationships between declines in receptive and expressive language skills and the presence of dementia among 14 of 28 DS participants with dementia. Written language did not show any significant change overtime, a finding attributed to low performance of participants at the onset of the study (i.e., floor effect).

Magnetic resonance imaging (MRI) investigations confirm that adults with DS and dementia have more generalized cerebral atrophy, mesial temporal lobe shrinkage, third ventricle enlargement, and reduced volumes of their left cerebral hemisphere hippocampus and left amygdala than adults with DS only (Pearlson, Breiter, Aylward, Warren, Grygorcewicz, Frangou, Barta, & Pulsifer, 1998). These neuroimaging data provide further evidence of a neurolinguistic basis for language and communication changes in adults with DS and dementia.

Moss, Tomoeda and Bayles (2000) investigated the relationship of age to language performance in 22 participants with DS only and 2 individuals with DS and dementia. They found that chronological age had a significant inverse relation to mental age and language performance. Lower scores of linguistic expression (e.g., repetition, naming, sentence completion, object description) and comprehension (e.g., following spoken commands, reading comprehension of words and sentences) all correlated significantly with older age. The language skills of the two individuals with DS and dementia were significantly poorer than the 22 participants with DS only and the mean scores of the entire standardization sample of individuals with early and middle clinical stage of DAT from the Arizona Battery for Communication Disorders of Dementia (Bayles & Tomoeda, 1993). Moss and colleagues noted that poor reading levels of individuals with DS, along with older chronological age and low mental age, placed them at risk for expressive language scores similar to or worse than individuals with DAT.

Conclusions

It is generally accepted that receptive language skills are more greatly affected by ageing in adults with DS than are expressive language skills (Carter Young & Kramer, 1991; Collacott & Cooper, 1997; Cooper & Collacott, 1995; Moss et al., 2000). Moreover, expressive language of adults with DS, which may remain relatively intact over time, can act as a measure of their overall ability level and reflect skill level of younger adults with DS (Cooper & Collacott, 1998). To date, however, the effect of DAT on the language and communication performances of adults with DS has not been controlled in many studies. Whether changes in receptive language are age-related, DAT-related, or a combination of both, remain largely unknown. Few studies thus far have examined systematically and in detail the language and communication in adults with DS and DAT, compared the language performance of adults with DS and DAT versus adults with DS only, described specific language functions in adults with DS and DAT, or

explored relationships between language and adaptive behaviour in adults with DS and DAT. Despite preliminary reports that language and communication problems are present in individuals with DS and may be a harbinger of the onset of DAT, there is little detailed information on the language and communication profile of adults with DS and DAT. Preliminary reports describing strategies about how to maintain communication with individuals with DS and DAT need to be verified empirically to establish solid evidence-based clinical practice patterns (McCallion, 1999). There is a clear and definite need for further cross-sectional and longitudinal experimental studies as well as multi-perspective observational field investigations of the language and communication in individuals with DS and DAT.

Acknowledgments

References

American Psychiatric Association (1994). *Diagnostic and statistical manual of mental disorders (4th ed.).* Washington: American Psychiatric Association.

Ashman, A.F. (1982). Coding, strategic behaviour, and language performance of institutionalized mentally retarded young adults. *American Journal of Mental Deficiency, 86,* 627-636.

Aylward, E. Burt, D Thorpe, L. Lai, F. & Dalton, A.J. (1995). *Diagnosis of dementia in individuals with intellectual disability: Report of the AAMR-IASSID working group for establishment of criteria for the diagnosis of dementia in individuals with developmental disability.* Washington: American Association on Mental Retardation.

Azari, N.P., Horwitz, B., Pettigrew, K.D., Grady, C.L., Haxby, J.V., Giacometti, K.R., & Schapiro, M.B. (1994). Abnormal pattern of cerebral glucose metabolic rates involving language areas in young adults with Down syndrome. *Brain and Language, 46,* 1-20.

Bayles, K.A., & Tomoeda, C.K. (1993). *Arizona battery for communication disorders of dementia.* Tucson, AZ: Canyonlands Publishing.

Bayles, K.A., & Tomoeda, C.K., & Trösset, M.W. (1992). Relation of communication abilities of Alzheimer's patients to stage of disease. *Brain and Language, 42,* 454-472.

Burt, D.B., Loveland, K.A., & Lewis, K.R. (1992). Depression and the onset of dementia in adults with mental retardation. *American Journal on Mental Retardation, 96,* 502-511.

Campbell-Taylor, I. (1993). Communication impairments in Alzheimer disease and Down syndrome. In J.M. Berg, H. Karlinsky & A.J. Holland (Eds.), *Alzheimer disease and Down syndrome and their relationship* (pp. 155-171). Oxford: Oxford University Press.

Carter Young, E., & Kramer, B. (1991). Characteristics of age-related language decline in adults with Down's syndrome. *Mental Retardation, 29,* 75-79.

Chapman, R.S., Seung, H., Schwartz, S.E., & Bird, E.K. (2000). Predicting language production in children and adolescents with Down's syndrome: The role of comprehension. *Journal of Speech, Language and Hearing Research, 43,* 340-350.

Collacott, R.A., & Cooper, S.A. (1997). A five-year follow up study of adaptive behaviour in adults with Down syndrome. *Journal of Intellectual & Developmental Disability, 22,* 187-197.

Cooper, S.A., & Collacott, R.A. (1995). The effect of age on language in people with Down's syndrome. *Journal of Intellectual Disability Research, 39,* 197-200.

Cooper, S.A., & Collacott, R.A. (1998). Expressive language and general ability in Down's syndrome: A pilot study. *British Journal of Developmental Disabilities, 44,* 48-52.

Crayton, L. Oliver, C., Holland, A., Bradbury, J., & Hall, S. (1998). The neuropsychological assessment of age related cognitive deficits in adults with Down's syndrome. *Journal of Applied Research in Intellectual Disabilities, 11,* 255-272.

Dalton, A.J., & Fedor, B.L (1998). Onset of dyspraxia in ageing persons with Down syndrome: Longitudinal studies. *Journal of Intellectual & Developmental Disability, 23,* 13-24.

Dalton, A.J., & Crapper-McLachlan, D.R. (1986). Clinical expression of Alzheimer's disease in Down's syndrome. *Psychiatric Clinics of North America, 9,* 659-670.

Dalton, A.J., Seltzer, G.B., Adlin, M.S., & Wisniewski, H.M. (1993). Association between Alzheimer disease and Down syndrome: Clinical observations. In J.M. Berg, H. Karlinsky, & A.J. Holland (Eds.), *Alzheimer disease and Down syndrome and their relationship* (pp. 53-69). Oxford: Oxford University Press.

Das, J.P., & Mishra, R.K. (1995). Assessment of cognitive decline associated with ageing: A comparison of individuals with Down syndrome and other etiologies. *Research in Developmental Disabilities, 16,* 11-25.

Devenny, D.A., Krinsky-McHale, S.J., Sersen, E., & Silverman, W.P. (2000). Sequence of cognitive decline in dementia in adults with Down's syndrome. *Journal of Intellectual Disability Research, 44*(6), 654-665.

Devenny, D.A., Silverman, W.P., Hill, A.L. Jenkins, E., Sersen, E.A., & Wisniewski, K.E. (1996). Normal ageing in adults with Down's syndrome: A longitudinal study. *Journal of Intellectual Disability Research, 40*(3), 208-221.

Evenhuis, H.M. (1990). The natural history of dementia in Down's syndrome. *Archives of Neurology, 47*(3), 263-267.

Evenhuis, H.M. (1992). Evaluation of a screening instrument for dementia in ageing mentally retarded persons. *Journal of Intellectual Disability Research, 36,* 337-347.

Evenhuis, H.M., Kengen, M.M., & Eurlings, H.A. (1990). *Dementia Questionnaire for Mentally Retarded Persons.* Zwammerdam: Hooge Burch, Institute for Mentally Retarded People.

Evenhuis, H.M., van Zanten, G.A., Brocaar, M.D., & Roerdinkholder, W.H.M. (1992). Hearing loss in middle-age persons with Down syndrome. *American Journal on Mental Retardation, 97,* 47-56.

Haxby, J.V. (1989). Neuropsychological evaluation of adults with Down's syndrome: Patterns of selective impairment in non-demented old adults. *Journal of Mental Deficiency Research, 33,* 193-210.

Holland, A.J., Hon, J., Huppert, F.A., Stevens, F., & Watson, P. (1998). Population based study of the prevalence of presentation of dementia in adults with Down syndrome. *British Journal of Psychiatry, 172,* 493-498.

Janicki, M.P., Heller, T., Sletzer, G.B., & Hogg, J. (1996). Practice guidelines for the clinical assessment and care management of Alzheimer's disease and other dementias among adults with intellectual disability. *Journal of Intellectual Disability Research, 40,* 374-382.

Jordens, W.C., Evenhuis, H.M., & Janssen C.G.C. (1997). Ageing and cognitive decline in people with Down's syndrome. *British Journal of Developmental Disabilities, 43,* 79-84.

Kernan, K.T. (1990). Comprehension of syntactically indicated sequences by Down syndrome and other mentally retarded adults. *Journal of Mental Deficiency Research, 34,* 169-178.

Kledaras, J.B., McIlvane, W.J., & Mackay, H.A. (1989). Progressive decline of picture naming in an ageing Down syndrome man with dementia. *Perceptual and Motor Skills, 69,* 1091-1100.

Lai, F. (1992). Clinicopathological features of Alzheimer disease in Down syndrome. In L. Nade and C.J. Epstein (Eds.), *Down syndrome and Alzheimer disease* (pp. 15-34). NY: Wiley-Liss.

Lai, F. & Williams, R. (1989). A prospective study of Alzheimer disease in Down's syndrome. *Archives of Neurology, 46,* 849-853.

Lowe, C. & Temple, V. (2002). Identifying hearing loss in adults with developmental disabilities. *Journal of Speech-Language Pathology and Audiology, 26,* 20-26.

Malamud, N. (1964). Neuropathology. In H.A. Stevens & R. Herber (Eds.), *Mental retardation: A review of research* (pp. 429-452). Chicago: University of Chicago Press.

Malamud, N. (1972). Neuropathology of organic brain syndromes associated with ageing. In C.M. Gaitz (Ed.), *Ageing and the Brain* (pp. 63-87). New York: Plenum.

Mann, D.M.A. (1993). Association between Alzheimer disease and Down syndrome: Neuropathological observations. In J.M. Berg, H. Karlinsky, & A.J. Holland (Eds.), *Alzheimer disease and Down syndrome and their relationship* (pp. 71-92). Oxford: Oxford University Press.

McKhann, G., Drachman, D., Folstein, M., Katzman, R., Price, D., & Stadlan, E.M. (1984). Clinical diagnosis of Alzheimer's disease. *Neurology, 34,* 939-944.

McCallion, P. (1999). Maintaining communication. In M.P. Janicki & A.J. Dalton (Eds.), *Dementia, ageing, and intellectual disabilities: a handbook* (pp. 261-277). Philadelphia, PA: Brunner/Mazel.

Moss, S.E., Tomoeda, C.K., & Bayles, K.A. (2000). Comparison of the cognitive-linguistic profiles of Down syndrome adults with and without dementia to individuals with Alzheimer's disease. *Journal of Medical Speech-Language Pathology, 8*(2), 69-81.

Orange, J.B. (1991). Perspectives of family members regarding communication changes. In R. B. Lubinski (Ed.), *Dementia and communication* (pp. 168-186). Philadelphia: Mosby.

Pearlson, G.D., Breiter, S.N., Aylward, E.H., Warren, A.C., Grygorcewicz, M., Frangou, S., Barta, P.E., & Pulsifer, M.B. (1998). MRI brain changes in participants with Down syndrome with and without dementia. *Developmental Medicine & Child Neurology, 40,* 326-334.

Rasmussen, D.E., & Sobsey, D. (1994). Age, adaptive behaviour, and Alzheimer disease in Down syndrome: Cross-sectional and longitudinal analysis. *American Journal on Mental Retardation, 99,* 151-165.

Roeden, J.M., & Zitman, F.G. (1997). A longitudinal comparison of cognitive and adaptive changes in participants with Down's syndrome and an intellectually disabled control group. *Journal of Applied Research in Intellectual Disabilities, 10,* 289-302.

Rondal, J.A. (1988). Language development in Down's syndrome: A life-span perspective. *International Journal of Behavioural Development, 11,* 21-36.

Rondal, J.A., & Lambert, J.L. (1983). The speech of mentally retarded adults in a dyadic communication situation: Some formal and informative aspects. *Psychologic Belgica, 23,* 49-56.

Sabsay, S., & Kernan, K.T. (1993). On the nature of language impairment in Down syndrome. *Topics in Language Disorders, 13,* 20-35.

Visser, F.E., Aldenkamp, A.P., van Huffelen, A.C., Kuilman, M., Overweg, J., & van Wijk, J. (1997). Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Don syndrome. *American Journal on Mental Retardation, 101,* 400-412.

Whalley, L.J. (1982). Dementia of Down's syndrome and its relevance to aetiological studies of Alzheimer's disease. *Annals of New York Academy of Sciences, 396,* 39-53.

Wisniewski, K.E., Wisniewski, H.M. & Wen, G.Y. (1985). Occurrence of neuropathological changes and dementia of Alzheimer's disease in Down's syndrome. *Annals of Neurology, 17,* 278-282.

World Health Organization (1990). *International statistical classification of diseases and related health problems.* Geneva: World Health Organization.

Zigman, W.B., Schupf, N.S., Sersen, E., & Silverman, W. (1995). Prevalence of dementia in adults with and without Down syndrome. *American Journal on Mental Retardation, 100*(4), 403-412.

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