Editorial

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In this issue, the first three papers address issues which have relevance to aspects of language in individuals with Down syndrome and the remaining papers address health and medical issues. The first paper was presented at the 3rd International Conference on Language and Cognition in Down Syndrome, Portsmouth, 2000, and the poster abstracts from that conference are included in this issue. The conference papers are being published in full in this journal. Many have already been published in volume 7 and the remaining two will be in later issues of Volume 8.

In the first paper, Jean Rondal and Annick Comblain of the University of Liege, Belgium, consider the language functions of French speaking individuals with Down syndrome from mid adolescence to approximately 50 years of age. They report the findings of a cross-sectional study of individuals at mean ages of 16, 27 and 44 years as well as a four year longitudinal study. They also draw on data on productive language from one of their earlier published studies and they conclude that they find no evidence of change in the grammatical, lexical or phonological aspects of receptive or productive language over the time periods that they have studied.

Their interest in language changes over time is linked with a concern that the scientific community develop measures sensitive enough to study the cognitive changes which may accompany normal ageing in individuals with Down syndrome or be indicators of the onset of a pathological decline associated with Alzheimer disease. In their longitudinal study, Rondal and Comblain with colleagues, measured the cerebral metabolic rate, CMR, of the individuals in the study at yearly intervals using PET scans. There was a significant decline in CMR over the period of the study but no related decline in language skills. The authors point out that the life expectancy of individuals with Down syndrome has increased dramatically and that some 50% will live to 55 years or beyond and some 15% to 68 years or older. They argue that more services should be developed to ensure that the quality of life of adults is good and is maintained into old age. They indicate that speech and language therapy for adults is developing in a number of countries and that its effectiveness needs to be evaluated - the adults that they have studied have shown no significant change in their language skills and perhaps progress would be possible with appropriate therapy. There have been two recent publications which have indicated that therapy can produce change in the language of adults (Leddy & Gill, 1999; Jenkins, 2001).

The authors point out that their data from their cross-sectional study spanning some 30 years of age may need to be interpreted with caution as the younger groups may have had more effective education and therapy and therefore better speech and language skills than the older groups had achieved as adolescents. For example, if this were the case it could mask any progress that the young adults had actually made since adolescence. On the issue of measuring changes with age, the authors rightly stress the need to consider different aspects of language and to recognise that the phonological, syntactical, lexical and pragmatic aspects vary independently. Lexical knowledge and pragmatic skills may continue to develop in adult life, but it may be more difficult to improve phonological and syntactical skills. The current study provides no evidence of progress in syntax from mid adolescence yet an increase in MLU for some individuals older than 16 years has been reported in another longitudinal study (Chapman, 2001). Clearly more longitudinal research is needed to clarify these issues and such studies need to document the education, therapy and social experience of participants during the time of the study to aid interpretation of their results.

The second paper is concerned with an issue that is relevant to both spoken and written language, phonological awareness. Helen Fletcher and Sue Buckley, of the Department of Psychology, the University of Portsmouth, UK report the findings of an investigation of the phonological awareness skills of a group of 17 children with Down syndrome (mean age 12 years 2 months) and any links between these and their reading skills. Phonological awareness, the ability to identify the component sounds in spoken words, has been shown to be an important component of reading skills in typically developing children. It is a necessary prerequisite for developing the use of alphabetic strategies for decoding and spelling words as a reader.

Measures of phonological awareness include tasks which require children to recognise rhyming words, words which begin with the same sound - alliteration, to blend sounds at the phoneme level to create a word e.g. c-a-t = cat, and to break words into their component phonemes. These four tasks, rhyming, alliteration, blending and segmenting were

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presented to the children, supported by picture material, so that for the first 3 tasks the child could select the correct answer from a choice of pictures but the segmenting task required the child to produce the sounds in the words. The children performed well on the first 3 phonological awareness tasks but found segmentation much more difficult.

On standardised reading and spelling tasks, the group had mean age-equivalent scores of 7 years 2 months and on reading comprehension a mean age-equivalent score of 6 years 3 months. Tests of alphabetic skills, non-word reading and spelling were included. The children's ability to blend phonemes was the only skill to correlate positively with non-word reading or spelling abilities, suggesting that the ability to hear and manipulate sounds at the phoneme level is the skill that is significantly linked to alphabetic reading strategies. However, there were children in the group who were able to score well on the blending task but still showed no alphabetic reading skill when confronted with non-words, indicating that phonemic awareness is a necessary but not sufficient skill for the development of an alphabetic reading strategy. It is likely that children need specific instruction in the use of their phonemic knowledge for decoding unfamiliar words and for spelling.

Typically developing children who can read and spell at a 7 year level would usually find the phonological awareness and alphabetic tasks used in this study easy, indicating that children with Down syndrome use mainly a logographic strategy to read for longer than typically developing children, as has recently been suggested by the data from another study (Kay-Raining Bird, Cleave & McConnell, 2000).

The third paper in this issue, by Lee Bennetts and Mark Flynn of the Department of Speech and Language Therapy, University of Canterbury, New Zealand addresses the issue of the effects of background noise, such as in a classroom, on the speech perception of children with Down syndrome. The authors identify the need to investigate the hearing abilities of children with Down syndrome in the classroom environment. Drawing on published studies, they point to the fact that learning in the classroom is heavily dependent on the ability to process spoken language. They estimate that children spend at least 45% of their time in listening activities and that the majority of school instruction is verbal. Children with Down syndrome may be at a significant disadvantage in the typical mainstream classroom, given their risk of mild to moderate hearing loss. Sound-field amplification systems, which reduce the signal-to-noise ratio for the listener, have been shown to assist the learning and 'on-task' time of typically developing children as well as those with hearing impairments. In an exploratory study with 4 children with Down syndrome, aged 5 years 8 months to 7 years 3 months, the authors demonstrated the effectiveness of sound-field amplification in supporting accurate speech perception. The children only had mild hearing losses, but as background noise increased their accuracy in identifying familiar words decreased significantly. With sound-field amplification providing a 10dB enhancement, there was no decrease in performance as background noise level increased.

These children had better hearing than the majority of children with Down syndrome, yet their speech perception was markedly affected by listening against background noise. It can be expected that this effect will be greater for those with more severe conductive or sensori-neural losses. More research is needed to see if the effect of the sound-field amplification is equally effective when children have a greater degree of hearing loss, and, as the authors point out, research in the classroom is needed to evaluate the system in situ. In the classroom, a larger number of variables could be measured including, for example, comprehension tasks, time on task, listening spans, as well as speech perception. This could be a very important area of research as this technique is not expensive to implement and could significantly benefit children in the classroom. The authors also point out that there are benefits from using the system for the teacher in protecting his or her voice as well as potentially improving the learning outcomes for the students. The use of sound-field amplification in the homes of children with Down syndrome should also be considered. One successful phonological training study has reported the use of amplification (Cholmain, 1994) in the home.

The next three papers address health and medical issues. The first of these by Corrado Romano, Rosa Pettinato, Letizia Ragusa, Concetta Barone, Antonino Alberti and Pinella Faila reports a summary of the findings of study which has investigated links between zinc levels and immune system antibodies in 120 individuals with Down syndrome. Zinc is known to play a central role in the immune system and in resistance to infections in the typically developing population. A number of studies have reported reduced zinc levels in some individuals with Down syndrome and this literature is reviewed in this paper. Some studies report negative consequences of low zinc levels for health, others do not. The authors also review studies of zinc supplementation and conclude that the results of different studies are conflicting - some claim benefits for growth and resistance to infection, others report no significant effects of supplementation.

In this study of 120 individuals, ranging in age from 4 months to 48 years, zinc levels were normal for 80% of the group and low for 20%. The researchers then compared the two groups on a variety of measures including growth hormone secretion, antigliadin antibodies, thyroid function markers and immunoglobulins. They found no significant differences on any of the many measures that they used except in the IgG4 antigliadin antibody. They conclude that the state of zinc deficiency in their sample was not associated with any dysfunction in growth hormone secretion, proneness to coeliac disease, thyroid function or immune function.

The next paper is a short report of an epidemiological study of the incidence of oesophageal atresia (incomplete development of the oesophagus) in Down syndrome. The authors, Sebastiano Bianca, Marco Bianca and Guiseppe

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Ettore of the University of Catania and Azienda Ospedaliera Garibaldi, Catania, Sicily, reviewed data from the Sicilian Registry of Congenital Malformations from 1991 to 1998. They found an incidence of 0.9% for oesophageal atresia in a population of 333 babies with Down syndrome born during this period compared with and incidence of 0.13% in the rest of the infant population. This is a 30 fold increase in incidence. This is higher than reported previously in the literature.

The final paper is from Lital Keinan Boker and Joav Merrick from the University Medical Centre, Utrecht, The Netherlands and National Institute of Child Health and Human Development, Tel Aviv University, Israel respectively. This is also an epidemiological study of the incidence of cancers, including leukaemias, in individuals with Down syndrome. The study population comprises of two groups, those born before 1979 and those born from 1979 and 1995. These two groups represent the total population of individuals with Down syndrome resident in Israel from 1948. In the younger age group aged 1 to 17 years all the 7 cases of cancer reported for the group had suffered from leukaemia - a 25 fold increase in expected cancer incidence for this age group. In the older group, 17 cases of cancer were found, 4 leukaemias and 13 solid tumours. This represents a 7 fold increase in expected incidence for this age group. The solid tumours were varied and included a breast cancer, a pituitary tumour, two cases of ovary cancer, Hodgkin's disease, a malignant melanoma, an oesophageal cancer, two biliary tumours, a sarcoma, a seminoma and two cases of gastric cancer. The breast cancer incidence is significantly lower than might be expected and the gastric cancer incidence is higher. The authors compare their data with the literature and identify the need for further studies of the incidence of cancer in this population.

Rondal and Comblain (this issue) point out that the population of adults with Down syndrome is expected to double by 2025, due to increased longevity, therefore it is urgent that the research community turn their attention to all aspects of the health-care, cognitive and social needs of adults.

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