

Editorial

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In this issue, we have papers on four independent topics, the effects of probable Alzheimer disease on speech, language and memory function, the links between early reading skills and phonological awareness, the effects of visual feedback on movement and sleep related breathing disorders in adults with Down syndrome.

The first paper, entitled *Françoise, a fifteen year follow-up*, from Jean Rondal and his colleagues M. Elbouz, M. Ylieff, and L. Docquier, at the University of Liège in Belgium presents an individual case study that describes in detail the changes in language and memory function of Françoise, a 47 year old lady with Down syndrome. Françoise will already be well known to many in this field as she developed exceptionally good spoken language abilities in her earlier years and Rondal published a detailed study of her language, memory and cognitive skills in 1995.^[1] In 2000, the day centre that Françoise attended became concerned about changes in her behaviour and requested further investigation. This case study is of particular interest because Françoise had exceptionally good language abilities for a person with Down syndrome. It is therefore possible to identify subtle changes in her abilities which would not be evident in adults with more typical language abilities. The assessments revealed that in 2002, Françoise had relatively preserved articulatory skills though more dysfluencies and slower speech rate than previously. She showed considerable word finding difficulties and a significant deterioration in syntactic comprehension abilities. Her MLU had decreased but she still had basically preserved morphosyntax in her spontaneous speech, maintaining correct French word order and correctly marking gender, number, case, tense or aspect. Her short-term memory span for digits and words had decreased, and her sentence span had decreased very significantly. Rondal and colleagues review in detail the findings with regard to loss of language and memory skills in Alzheimer patients in the typical population and identify that Françoise's profile is similar – relatively spared syntactic production skills but greater difficulty with semantic comprehension of sentences. The authors suggest that the difficulty in sentence comprehension tasks may be linked to impairment of central executive function rather than verbal short-term memory per se. They also provide a useful review of the studies of ageing and cognitive change in individuals with Down syndrome, identifying

well preserved skills in the majority. They discuss possible explanations of the profile of language deterioration seen in both Françoise and typical adults with probable Alzheimer disease and suggest that differential effects on implicit or explicit memory systems or differential effects on a modular language system could be explanations at this point in our knowledge of the processes involved. This paper is a unique and valuable addition to the literature on dementia in Down syndrome.

The second paper, entitled *Early phonological awareness and reading skills in children with Down syndrome*, from Esther Kennedy and Mark Flynn of the Ministry of Education and the University of Canterbury, New Zealand, explores the phonological skills of beginning readers. In typical developing children, phonological awareness skills have been shown to be both predictors of reading ability in children, and to develop with reading instruction and reading progress. The authors point out that some researchers, including the Editor and colleagues, have suggested that children with Down syndrome make less use than typical children of the phonological (alphabetic) information provided by the letters in words when reading in the early years and rely to a greater extent on the whole word visual (logographic) features. Children with Down syndrome begin to show more phonological awareness and the ability to read non-words when they reach word reading skills of 7 to 8 years i.e. as a result of reading experience.^[2] Other researchers argue that a minimum level of phonological awareness and of short-term memory skills (digit span) are prerequisites for reading. These conflicting views are difficult to resolve as tasks designed to measure phonological awareness, particularly when involving judgements of rhyme or requiring phonemic isolation and blending are frequently too difficult for children with Down syndrome who are beginning readers. This paper reports the findings of a study of reading and phonological awareness skills of 9 children with Down syndrome. In addition to developing tests to assess rhyme awareness, alliteration, phoneme blending, phoneme isolation and non-word reading skills, the researchers also include measures of consonant production skill, speech perception skills and hearing. The children, as in other studies, found the phonological awareness tasks difficult, with only 3 able to score above chance on phoneme blending, 1 child on the rhyme task, 4 on phoneme isolation and

alliteration, and 3 on non-word reading. In contrast, all 9 children scored above chance on letter naming and 7 on letter sound knowledge. Alliteration (requiring identification of initial sounds in words only), letter naming and letter sound knowledge were the only skills significantly correlated with word reading abilities. Letter names and letter sounds are taught to children and the initial sounds likely to be the first phonemes identified in printed words. It can be argued therefore that these skills are all the result of reading instruction and improve with reading success. Interestingly, speech perception skills were correlated with speech intelligibility but not pure tone audiometry results, indicating the importance of assessing speech perception skills in addition to traditional hearing assessments, to guide therapy programmes. Speech perception skills were not correlated with any other measures. It may be the poor speech perception and speech sound production skills in children with Down syndrome that delay their development of an awareness of the sounds which make up words, until they receive explicit instruction on phonemes as part of literacy teaching. Explicit speech sound discrimination activities and games designed to teach them to be aware of alliteration and rhyme during their preschool years might be beneficial. This study, while limited by the numbers of children participating, should encourage others to continue to develop methods to explore these issues further and the authors propose directions for further research.

The third paper, entitled *Performing movement sequences with knowledge of results under different visual conditions in adults with Down syndrome*, from Naznin Virji-Babul, Jennifer Lloyd and Geraldine Van Gyn of the Down Syndrome Research Foundation and the University of Victoria, Canada, addresses the issue of the role of visual information when learning movements. Until recently the study of the development of the movement skills of children and adults with Down syndrome has been a neglected area. Many authors identify 'clumsiness', slowness and variability of movements as characteristic of individuals with Down syndrome but little is known about the reasons for this profile. In this paper, the authors compare the performance of 10 adults with Down syndrome with 14 neurologically 'typical' adults when required to carry out a task which involved completing a sequence of button presses as quickly as possible. Reaction times and movement times were measured under 2 conditions – *full visual feedback*, with full view of arms and hands, and *no visual feedback*, when participants could not see arms and hands but the buttons on the board were still visible. The results indicate that neither movement time nor reaction time were affected by visual feedback for either group of participants. The authors suggest that this indicates that individuals with Down syndrome do not have any proprioceptive deficits as proprioceptive feedback alone allowed them to perform the task successfully. However, fingertip positions must have been visible as they touched buttons in the 'no visual feedback' conditions, and perhaps this was sufficient visual information for success in this task. The typical adults showed no improvement in performance over the trials in the study but the adults with

Down syndrome showed considerable improvement, still continuing at the final 20th trial. This confirms the findings of other researchers which indicate that many adults with Down syndrome have not fully developed their motor skills as they show considerable improvements with training. As the authors point out, this improvement during the study means that in future, research designs should allow all participants to reach their best performance levels before making comparisons between groups.

The final paper, entitled *Sleep related breathing disorders in adults with Down syndrome* from Onofria Resta, Maria Barbaro and colleagues from the Universities of Bari and Foggia in Italy, provides information on the type and incidence of sleep disorders in adults. In this study, the nocturnal respiratory patterns of 6 adults were monitored for an 8 hour night period. Oxygen saturation levels and breathing patterns were monitored in addition to the information provided by a 12 channel polysomnograph. The study indicated high levels of sleep disturbance. Five of the 6 adults had sleep apnoea, the majority (89.2%) of the apnoeic episodes were obstructive rather than central (10.8%). Some 70% of the apnoeic episodes induced significant oxygen desaturation. The authors compare their findings with the small number of other published studies, some of which indicate that, as in the general population, age and weight are risk factors in sleep apnoea in adults with Down syndrome, although this was not demonstrated in this study. The authors do not comment on the possible effects of such a high incidence of sleep apnoea and accompanying oxygen desaturation on the health, learning abilities or day-time behaviour of those affected. This would seem an important area for future study – for both adults and children with Down syndrome – as there is already some evidence of effects of sleep disturbance on daytime behaviour in children.^[3]

References

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