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THE NATIONAL DOWN SYNDROME SOCIETY'S 11TH INTERNATIONAL DOWN SYNDROME RESEARCH CONFERENCE ON COGNITION AND BEHAVIOUR

(Florida, November 1997)

Sue Buckley

The National Down Syndrome Society held its 11th International Down Syndrome Research Conference on Cognition and Behaviour in Florida in November 1997. I was invited to participate in the 3 day meeting, giving me the opportunity to share this summary with readers. I have attempted to review the main points from most of the papers. Many of the presenters have published some or all of their work, so if any reader would like more information on any topic please contact me and I will provide references or put them in touch with authors. I have grouped the papers into topic areas, starting with infancy and moving to adult issues, with medical/genetic research at the end.

Infancy

The first presentation by Dr Rathe Karrer and Dr Jennifer Hill Karrer of the Smith Mental Retardation Centre, University of Kansas outlined their research on attention and cognition in infants. They explained how they are using measures of the electrical activity of the brain called event-related potentials (ERPs) to study cognitive development in infants. ERPs are measured by electrodes placed on the baby's scalp. It is a harmless and non-invasive technique that can be used to measure brain responses to stimuli presented to the baby. They are looking at responses to visual stimuli such as attention, response to novelty and recognition memory in infants with and without Down syndrome. They are engaged in longitudinal studies and may be able to relate the ERP data to different rates of progress in the children.

ERP measures are being widely used in other centres in the UK and USA to study brain development in infants and children with Down syndrome and to chart the effects of different rates of functional development on specialisation in the brain. It is even argued that the technique can be used to evaluate the

effects of different types of intervention or input on the development of the brain.

Speech and language

The next presentation by Dr Carolyn Mervis of the Department of Psychology, University of Louisville, Kentucky, reviewed her research on vocabulary acquisition and on children's ability to master category concepts. She pointed out that the ability to form concepts and to categorise is critical to a child's ability to understand and to organise their experience of the world. She has collected detailed longitudinal data on five children with Down syndrome and they showed two rates of progress with two children progressing like typical children and three progressing more slowly, highlighting the variability to be expected in their development.

A second paper on language development was presented on the second day by Dr. Libby Kumin, Department of Speech/Language Pathology, Loyola College, Maryland. She has collected data on 115 children from two to five years by asking their parents to complete the McArthur Communicative Development Inventory. This asks parents to identify the words and sentences that their child is using from a checklist. Mean vocabularies grew from 55 words at age two by approximately 100 words per year to 391 words at five years. However, variability at each age was very large (from 8 to 226 words for different children at two to 62 to 611 at five years). She also reported on the emergence of early grammatical markers such as plurals, possessives and past tenses. These also showed wide variation in the age they were achieved by different children.

Both the papers on language illustrated the very different rates of language development seen in different children with Down syndrome. Research which identifies the reasons for these different rates of progress is

needed if we are to become more effective with individually targeted interventions matched to children's specific needs.

Early communication and later social skills

Dr Marion Sigman, from the School of Medicine, University of California Los Angeles, reported the findings of a longitudinal study of 70 children with Down syndrome followed from pre-school to middle school years. In the pre-school years, the children's ability to initiate interactions and to share in joint attention situations (e.g. to attend to same object or activity with a communication partner) correlated highly with verbal language development. Followed up eight to ten years later, those with better interactive skills as infants were better socially integrated in the school playground. Dr Sigman therefore emphasized the importance of following the baby's lead and talking about what he or she is looking at to encourage these joint interaction skills. She also observed that schools do not use playtimes as positively as they could to teach and foster social interactions between children.

Reading and language

There were two papers on reading and speech and language development. In the first, Dr Anne Fowler of the Haskins Laboratories, New Haven, Connecticut reviewed the research on language development, considering variation and progress in the four different aspects of language skill. These are 1) phonology - how words are pronounced, 2) grammar, syntax and morphology- how sentences are constructed, 3) lexical knowledge or vocabulary- the names and meanings of words 4) pragmatics - the rules governing the way that conversations are conducted. Dr Fowler pointed out that not only is there wide variation in overall language skill between children and adults with Down syndrome, but there is also variation in the levels of difficulty experienced within the four aspects of language for individuals. She also pointed out that current research findings needed to be interpreted with caution as we have only just begun to tap the full potential of persons with Down syndrome. With these caveats in mind, she reported that typically, children with Down syndrome have more difficulty with grammar and phonology than with vocabulary and pragmatics. She argued that phonological memory (auditory short-term memory - often measured by digit span tasks) is a core problem affecting language learning. For example in one study of young adults,

correct use of verbal auxiliaries (e.g. is and are in he is running, they are running) was predicted by digit span, those with better digit spans having more correct use of grammar. She also pointed out that some individuals achieve good language and some good literacy skills also, the good readers also having better digit spans than most people with Down syndrome.

My paper on stimulating reading and language through the school years addressed some of the issues raised by Dr Fowler. Drawing on two longitudinal data sets collected with colleagues here at The Sarah Duffen Centre and in the Department of Psychology, University of Portsmouth, I argued that the better digit spans and better speech and language skills of readers may be the consequence of learning to read rather than causal factors. Studies of typically developing children would support the view that this may be the case and that progress in either short memory, or speech and language or reading skill will have a beneficial effect for the other two skills - this is referred to as reciprocal causation or 'bootstrapping'. I also illustrated the range of reading, spelling and comprehension progress being made by children in our ongoing longitudinal study and the rate at which they are becoming alphabetic readers (e.g. able to use phonic knowledge to 'sound out' an unfamiliar word). It appears that they move from relying on logographic (relying on visual memory of whole words) to alphabetic strategies once they reach the reading levels of a typical seven year old on our tests. This is very encouraging especially as the children with Down syndrome are likely to have less good sound production and sound discrimination abilities to support the learning of phonics.

Short-term memory

One of the factors thought to be causally linked to some of the language learning difficulties is poor auditory (or verbal) short-term memory development in children with Down syndrome. Dr Chris Jarrold, Dr Alan Baddeley and Dr Alexa Hewes, of the University of Bristol, UK, presented their recent research on this topic as a poster presentation. They compared the performance of children with Down syndrome, children with moderate learning disability and typically developing children, all matched for vocabulary comprehension, on verbal and visual spatial short-term memory tasks. The children with Down syndrome have equivalent visual-spatial spans but significantly lower verbal spans when compared to the other two groups of children.

This difference was not related to hearing loss or speech articulation rate for the children with Down syndrome and they also showed no 'primacy' effect e.g. relatively good performance on the first item in a list. The authors conclude that their results suggest a problem in the phonological loop component of the working memory system - perhaps rapid trace decay or increased interference from subsequent items entering memory. This is an important area for further work as this system is critical for the support of all learning.

Adolescence and early adulthood

Three papers discussed the needs of adolescents and young adults with Down syndrome. Dr Don Van Dyke from the University of Iowa Hospital and Clinic, Iowa City presented an overview. He identified that the major task of adolescence is separation from the family. In early adolescence the main preoccupations for young people are growth and puberty. For young people with Down syndrome puberty is usually reached at the usual age, the growth spurt may be earlier and less and obesity may be a risk. This is a time when appearance and self-image become important, so it is important to encourage good hygiene, a pride in appearance and fashionable clothes. Friendships and the peer group become important and leisure interests and social competence will help the young person to be included. Dr Van Dyke emphasised the need for good health care, including sex education, contraception and sexual health care to be available to adults and adolescents with Down syndrome.

In a paper entitled Living in the Community, Dr Siegfried Pueschel discussed the needs of adults if they are to participate fully in the community. He pointed out that we need to consider planning for the transition from adolescence to adulthood, for post-secondary education, for living and recreational options. We need to consider social skills, relationships and sexuality, medical care and mental health issues. Dr Pueschel emphasised the need for forward planning to build up self-esteem and the social and practical skills that will be needed for meaningful participation in the community. He also discussed employment and living options and the need for preparation for relationships. He particularly stressed the value of leisure and recreational activities, arguing that they can build self-confidence, improve motor skills and physical fitness, increase independent functioning, nurture creativity and self expression and provide opportunities for friendships.

Adult mental health

Dr William Cohen from Pennsylvania Children's Hospital, Pittsburgh, shared his experience of treating behavioural problems of young people with Down syndrome at his specialist Centre. About one third had some difficulty. The most common problems were defiant or aggressive behaviours and adjustment difficulties - typical adolescent difficulties. One person had depression, one panic attacks and another an eating disorder. For all adolescents with disabilities, adolescence is a particularly difficult time as they come to realise the real impact that their disability is having on their lives when they see peers and siblings moving on in ways that they are unable to achieve. In the young adults, less than one third had any problems and none were aggressive. Depression, anxiety, eating disorders and obsessive compulsive disorder were the type of problems seen, again problems occurring in a minority of the rest of the population of this age. Dr Cohen gave examples of responses to treatments for his patients and drew attention to one important issue. People with Down syndrome may be inclined to talk to themselves when under stress. You and I will be worrying silently in our heads, they are doing this worrying out loud and it is important that it is not misinterpreted as a psychotic symptom.

A further paper on psychiatric disorders in adults with Down syndrome was presented by Anna Mies and Kendra Moses of Southern Illinois University School of Medicine giving similar outcomes in their clinic population. They pointed out that some 20% to 30% of adults with Down syndrome may suffer from a mental disorder at some time. They are less vulnerable than adults with learning disabilities from other causes, but may be more likely to have depression or dementia in later life. It is important that doctors and carers realise that a mental illness can occur and can be treated, especially as some folk will have limited communication skills and their difficulties be misinterpreted, so go untreated or be treated inappropriately.

Issues for older adults

Dr Dennis McGuire and Dr Brian Chicoine, from the Lutheran General Hospital, Glenview, Illinois shared the results of their experience of working with some 600 adults at their Adult Down Syndrome Centre. They had information on their self-help, social and communication skills as well as their health care needs. Some 10% of adults needed help

with all daily living skills and a further 5% needed prompts. Of the rest 38% were mostly independent, 40% needed some support in their lives and 7% were able to manage selfcare but were not socially or practically independent. While 75% of adults had communication which was understood by caregivers most of the time, only 28% could be understood most of the time by unfamiliar others. When it came to expressing feelings, 78% could express them well non-verbally but only 39% verbally and so many caregivers found it difficult to interpret the expressed emotions of the person with Down syndrome that they were caring for. Especially vulnerable in this respect were those with mental health problems (about 30% of the group).

The most common health issues were sensory deficits - of 602 people, 312 had impaired vision and 247 had a significant hearing loss and 351 had impacted wax in their ears! Overweight was an issue for 423 people, 222 were hypothyroid, 6 had vitamin B12 deficiency and 11 were diabetic. 41 had developed seizures and only 33 suffered from sleep apnoea suggesting it is less common in adults with Down syndrome than in children. Of the 300 women in the study, 49 had passed the menopause and 89 suffered from dysmenorrhea.

Of those with mental health problems, 87.5% were described as having a 'reversible' or treatable disorder and only 12.5% had Alzheimers dementia. (This meant that only 26 of the total of 579 had dementia and they were all over 40 years of age). This study also drew attention to the frequent occurrence of self-talk, which was reported as used by 79% of the adults. Eighty percent were friendly and social, with only 20% described as anti-social or inappropriate in their behaviour at times.

Medical/genetic research

There were two papers in which the progress in understanding the genetics of Down syndrome was reviewed. One emphasized the search for links between genetic profiles and cognitive profiles, the other explained the potential of trisomic mouse models for testing some of these hypotheses. Both these speakers gave similar presentations at the Vancouver conference in April 1998, which I will be reporting on in the next issue, so I will include a little more detail then.

Two papers discussed the search for effective pharmacological treatments for some of the

abnormalities in biochemical functioning and the health risks associated with Down syndrome. One was presented by Dr Alberto Costa of the Jackson Laboratory, Bar Harbor and the other by Dr George Capone of the Kennedy Krieger Institute, Baltimore. These were both speculative papers, discussing the areas in which research might be worthwhile based on our current knowledge and the enormous methodological difficulties facing such research. Dr Capone pointed out that there is a need to develop a valid and reliable protocol for accurately measuring the development of children with Down syndrome before the effectiveness over time of any interventions can be reliably evaluated.

The American Down Syndrome Medical Interest Group, which includes many of the most expert medical practitioners, biochemists and geneticists in this field, had its meeting at this conference. (A number of these experts are also parents of children with Down syndrome themselves). This meant that there was discussion of health guidelines, areas that need further research and exchange of good practice. (There was also extensive discussion of the problems being caused by the promotion of Targeted Nutritional Interventions ('TNI') despite the lack of evidence for the wild claims being made for its efficacy by those selling it or those using it in their private practices). The view of this expert group, which is to not recommend the use of 'TNI', can be found on the NDSS website.

My views on this are made clear elsewhere in this issue and on our website. The most distressing aspect of this whole sorry debate is the unnecessary stress and anxiety being caused to new parents by the conflicting information that they are being bombarded with at present.

The Author:

Sue Buckley is Professor of Developmental Disability at the Department of Psychology at the University of Portsmouth, UK. She is also Director of The Centre for Disability Studies at The University of Portsmouth and Director of Research and Information Services at The Down Syndrome Educational Trust. She also serves as a Non-Executive Director of the Portsmouth and South East Hampshire District Health Authority and on the boards of the European Down Syndrome Association and the International Down Syndrome Federation.