From scientific research to intervention in Williams syndrome

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Book review of "Understanding Williams syndrome: Behavioural Patterns and Interventions", by Eleanor Semel and Sue Rosner. ISBN0-8058-2618-1, Lawrence Erlbaum Associates, 2003. 456 pages. (£26.51 on amazon.co.uk)

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Over the last few years, one of my main areas of scientific research in psychology has been into Williams syndrome (WS). Initially this was while working with Professor Annette Karmiloff-Smith at the Institute of Child Health and now in my position as a lecturer at Birkbeck College. My work has included investigation of language acquisition WS and recently has focused on visuo-spatial skills (some of you may have met my PhD student, Dagmara Annaz, who is investigating face recognition abilities in WS, Down's syndrome, and autism).

To parents of children and adults with WS, as well as teachers and clinicians, the results of theoretical research can sometimes seem far removed from everyday concerns of coping with the disorder. For children and adults with WS, going down stairs, tying shoelaces, and learning handwriting are the kinds of challenges to be addressed. How has scientific research helped with these kinds of issues?

Recently, a new book has been published by Eleanor Semel and Sue Rosner, entitled 'Understanding Williams Syndrome: Behavioural Patterns and Interventions'. This book summarises the current state of scientific knowledge on WS but importantly it also highlights links to the treatments and interventions that are available. It is aimed at multiple audiences: parents, carers, teachers, clinicians, and researchers.

I thought I would take this opportunity to offer a brief review of the book, since I believe it offers a valuable illustration of the way in which treatment and intervention depend on a solid foundation of basic science. For those interested in a scientific perspective on the book, I have also written a review for the journal Cortex, which can be downloaded from my website (www.psyc.bbk.ac.uk/people/academic/thomas_m/). However, in the rest of this article, I want to focus on the types of interventions recommended for WS and to place them in the context of current scientific knowledge.

In their book, Semel and Rosner offer a synthesis of current knowledge of WS from the perspective of Professor Ursula Bellugi's research laboratory at The Salk Institute in San Diego, one of the main US research laboratories involved in investigating the disorder. Semel and Rosner complement the results of scientific studies by clinical observations and personal accounts, making extensive use of two large-scale parental questionnaires to identify frequent and not-so-frequent characteristics of the disorder. The chapters are organised into a consideration of four main aspects of WS: language skills and problems, perceptual and motor performance, specific aptitudes (social skills, curiosity, memory, and musicality) and behavioural problems (fears and anxieties, distractibility, impulsivity, poor adaptability, low frustration tolerance, and atypical activity). In each case, consideration is given to avenues for intervention. Five basic forms of intervention are described: (1) task-specific interventions; (2) naturalistic training situations; (3) compensatory strategies; (4) environmental manipulations; (5) control mechanisms. In addition, Semel and Rosner discuss the professional specialists to whom parents might appeal, such as physical therapists, speech therapists, and occupational therapists. In the following paragraphs, I will use examples to illustrate each type of intervention, all drawn from the book.

Task-specific interventions focus on specific training and guided learning experiences in the particular area of weakness. For example, one characteristic weakness in WS is in visuo-spatial and motor skills. These include motor movements in response to visual patterns but also subtler aspects of visual perception, for instance as revealed by problems in deciding which of two lines is longer. Although language is a relative strength in WS, research has revealed that problems in visual-spatial

awareness appear to feed through into difficulties understanding some aspects of language. In particular, these include spatial terms such as prepositions (e.g., behind, in, under) and relational terms (e.g., before-after, first-second, all-some). Difficulties understanding these terms can make it hard to follow instructions, for instance in the classroom. Semel and Rosner describe a task-specific intervention for spatial language problems called the 'barrier task'. The child or adult with WS sits across a table from the instructor or parent. On the table are half a dozen items of special interest to the individual with WS. The instructor has a duplicate set of items and there is a barrier in the middle of the table so that the individual with WS cannot see the other person's items or their placement. Training then consists of the instructor asking the individual to perform some spatial arrangement of the items, such as "Put the cat *under* the table". Both the instructor and the individual construct the arrangement and then the barrier is lowered. The displays are compared and the individual is given feedback if necessary. Various spatial or relational terms can be practised in this way, and when the individual with WS is familiar with the game, they can even reverse roles so that the individual sets the problems for the instructor.

Naturalistic training situations take advantage of everyday tasks to practise areas of weakness. Almost all individuals with WS experience problems in the development of their motor skills, both in terms of large-scale motor movements such as walking on cobble stones (where limitations in spatial awareness and laterality have a severe impact) and in terms of fine motor skills such as holding and using a pencil (where weaknesses in visuo-spatial functioning undermine performance). This can lead to difficulties in everyday activities, such as eating, grooming, tool use, sports, and handwriting. Such abilities can, however, be practised in loosely structured informal

situations. For example, the child's awareness of body parts and types of movement can be bolstered in games like "Simon Says" and "Hoky Coky", where body positions must be imitated, and left and right sides of the body differentiated. Kitchen tasks also require eye-hand coordination, such as using a rolling pin to prepare dough, or a biscuit cutter to produce shapes such as circles or letters. This could be used also to bolster spatial language by naming the shapes and letters as your child cuts them out.

Compensatory or mediational strategies attempt to build on existing strengths in WS to find different ways to succeed in areas of weakness. For example, verbal strengths can be exploited to buttress deficiencies in motor tasks or in spatial memory. Motor tasks such as drawing can be aided by instructing individuals with WS to associate parts of a picture with verbal cues and verbalise the sequence of drawing parts of the picture. Thus the letter 'I' might be illustrated by a green dot at the top and a red dot at the bottom, and the individual instructed to "Start at the green dot," "Go straight down the street to the red dot," and "Stop at the red dot". The individual with WS can then guide his or her own movements using the learned verbal cue. Verbal skills can also be used to scaffold motor tasks through semantic associations. For example, the task of folding towels or clothes can be aided by instructions such as "make the corners kiss," "make it into a box", "make a sandwich", or "take the ear of the rabbit (corner) and put it on top of the other ear". Similarly, spatial memory can be aided by verbalisation, for instance using descriptions out loud of travel routes to help individuals find their way. By labelling landmarks and creating verbal instructions, the individual can be given more confidence in getting from home to school, work, or a friend's house and back again. However, some care is necessary with verbal mediation because successful verbal performance may mask a persistent lack of

deeper understanding. Thus Semel and Rosner note how in the area of arithmetic, memorisation of number facts and calculation routines may be futile if individuals with WS do not understand basic principles of mathematics such as one-to-one correspondence between numbers and quantities. As Annette's student, Daniel Ansari, has shown, individuals with WS can learn to count even to 1000 without alas understanding what counting is for and that the last number when counting a sequence of objects represents the totality of the set.

Environmental manipulations usually follow an analysis of precisely the aspects of a task that are causing difficulties. The aim then is to manage the environment in such a way as to improve task learning or performance. For example, walking down stairs can prove very difficult for individuals with WS. Research into visuo-spatial-motor skills has indicated that individuals with WS can be distracted by irrelevant visual information and can have difficulty in separating figures from backgrounds. In other words, they struggle to extract the relevant visual information from a scene needed to guide their motor movements. Control of lighting to avoid shadows and reduction of extraneous information can be useful. For descending stairs, Semel and Rosner advise that it is sometimes helpful to instruct individuals with WS to look straight at the stairway wall and hold onto the stair railing, initially with both hands. They suggest that glancing at the corner of the stair rise or placing masking tape at the edge of each step are other ways of coping. Handwriting is another motor skill where managing the task environment can aid performance. Suggestions include the use of larger, smooth flowing pens that are easier to grip, use of lined paper, and use of a slanted writing surface. Specialist pharmacies or websites for motability carry such equipment. One of the difficulties for children with WS is that they are sometimes left handed and

efforts to comply with teaching procedures for right-handed children can produce awkward writing styles, such as hooking their hand around the pen. Research has indicated that handedness and laterality as a whole is a problem area for individuals with WS. The tendency in WS towards confused or mixed handedness and perhaps even elevated levels of left-handedness has been viewed by researchers as an indirect indicator of possible anomalies in brain organisation – that is, the way the left and the right sides of the brain divide up the cognitive tasks between them during early development. Problems confusing left and right suggest certain task manipulations may be useful in learning motor tasks. For example, children are often helped in learning skills such as tying shoelaces and doing up buttons by adults modelling the behaviour and requiring the child to imitate the model. Laterality difficulties suggest that the best way to model motor behaviours is to sit next to the child rather than across from them in a mirror configuration. Sitting next to the child prevents them from having to reverse left and right to reproduce the behaviour.

Control mechanisms are a form of direct instruction or rechannelling to guide the behaviour of individuals with WS, including reinforcement of their successful efforts. These mechanisms can take several forms. For instance, since individuals with WS are often good at conversation, 'talking' can be used as a method to control behaviour. Thus Semel and Rosner suggest that many individuals with WS respond well to "reasoning", so that they can often be "talked out" of behaving inappropriately, "talked into" conforming to acceptable standards of behaviour, and "talked through" the establishing of new or improved forms of behaviour. In using rewards to encourage certain behavioural patterns, it is important to tailor such rewards to the particular interests of individuals with WS. Thus Semel and Rosner note that, in

contrast to other developmental disorders, the 'hyper' sociable cognitive profile of individuals with WS means that social praise can often be more effective as a reward than traditional treats such as sweets. And their unusual facility with face recognition can be exploited in the classroom by using face stickers as a reward rather than gold stars.

Control mechanisms can also be applied to dealing with problems in certain areas of language. Although language is often a strength, there can be difficulties with an area of language called 'pragmatics'. Pragmatics refers to the practical use of language in terms of its social and communicative roles. For instance, pragmatics would include the 'rules' of good conversation: taking turns, sticking to the point, and so forth. Individuals with WS have fewer problems with what Semel and Rosner call the 'feeling' aspects of pragmatics (social sensitivity: e.g., making eye contact, sensitivity to non-verbal cues) but can struggle in some areas, such as exhibiting excessive greeting behaviours, problems with sticking to the point, and with answering questions appropriately. Nevertheless, problems such as excessive use of greeting behaviours in WS are amenable to control mechanisms. Thus Semel and Rosner describe how in class, one student with WS can be assigned the role of "official class greeter", whereby the nominated individual is allowed to greet people on a specified list (but no others). Other roles can be assigned, such as class historian or classroom host/hostess. Roles can then serve to rechannel behaviour, providing opportunities to greet others but within specified limits.

There is much more in Semel and Rosner's book on the theme of intervention and the examples described here can only give a flavour. But the authors do draw some general lessons. They are as follows: (i) capitalise on the use of control mechanisms;

(ii) exploit the power of verbal mediation strategies; (iii) reduce external pressures and demands; (iv) enlist the services of qualified specialists; and (v) optimise the potential for growth. Notably, Semel and Rosner recommend intervention even in areas of relative strength in WS, such as language. The intention is to strengthen an area with the potential for greater strength and thereby maximise its possible use in supporting other weaker abilities. However, the authors are also realistic in accepting that a number of features in WS remain difficult to treat, including auditory hypersensitivity, visuo-spatial-motor integration skills, and various behavioural problems. (Although with regard to auditory hypersensitivity, I should mention that Josephine Marriage, formerly funded by the WSF, did find a solution for many sufferers. She used a treatment developed for tinnitus. A small in-the-ear device allows the individual to reduce or increase sound levels gradually. Not only did this help with hyperacusis but it also appeared to have a wider calming effect). Finally, there are other aspects of the book which I do not have space to discuss, such the degree of variability found in the behavioural features of WS, advice on possible pharmacological interventions in WS, and advice on the best educational classes for children with WS to be in at various ages.

Overall, the book represents an important contribution, both as a sourcebook and as a confirmation of the importance of parallel and interacting programmes of basic science and intervention. As well as a continuation of these streams, Semel and Rosner are right to point out that our next need is a systematic evaluation of the effectiveness of various treatment methods.

Lastly, it is worth noting that a book such as Semel and Rosner's is inescapably frozen in time. Already the field is moving forward, in terms of the psychology, brain mechanisms, and genetics of WS. To give some examples, Semel

and Rosner suggest that in understanding language use in WS, the next topics to investigate include language difficulties with word-finding, semantic relations, and unusual word choice. They particularly highlight the field of academic skills in WS as requiring further research, including work on learning to read and on number concepts. Each of these three areas has been the subject of recent scientific papers from the Neurocognitive Development Unit, respectively by myself on language development, by Dr. Emma Laing (now at Cardiff University) on learning to read, and by Dr. Daniel Ansari (now at Darmouth College, New Hampshire, USA) on number concepts, all in collaboration with Professor Annette Karmiloff-Smith. The on-going challenge is to integrate scientific findings with interventional approaches, but I believe that Semel and Rosner's book demonstrates how a concrete framework is already in place to take this enterprise forward. Basic science provides a stable foundation on which to recommend interventions and to understand why they may work. But basic science would not be possible without the invaluable co-operation of the many children and adults with WS and their families who have helped us with the projects. For that, I would once again like to express our gratitude. And to remind everyone that there is plenty of work left to do!

MT.

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Recent scientific papers on WS by members of the Neurocognitive Development Unit:

- Ansari, D., Donlan, C., Thomas, M. S. C., Ewing, S., Peen, T., & Karmiloff-Smith, A. (2003). What makes counting count? Verbal and visuo-spatial

contributions to typical and atypical number development. Journal of Experimental Child Psychology, 85, 50-62.

- Karmiloff-Smith, A., Grant, J., Ewing, S., Carette, M. J., Metcalfe, K., Donnai, D., Read, A. P., Tassabehji, M. (2003). Using case study comparisons to explore genotype-phenotype correlations in Williams-Beuren syndrome. Journal of Medical Genetics, 40(2), 136-140.
- Laing, E., Hulme, C., Grant, J., & Karmiloff-Smith, A. (2001). Learning to read in Williams syndrome: Looking beneath the surface of atypical reading development. Journal of Child Psychology and Psychiatry, 42, 729-739.
- Thomas, M. S. C., Dockrell, J. E., Messer, D., Parmigiani, C., Ansari, D., & Karmiloff-Smith, A. (submitted for publication). Speeded naming, frequency and the development of the lexicon in Williams syndrome. Language and Cognitive Processes.