Williams syndrome: Fractionations all the way down?

(Review of Eleanor Semel and Sue Rosner's Understanding Williams Syndrome.

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In healthy adults, there is some case to be made that discrete cognitive components or functional circuits underlie different high-level abilities such as language, face recognition, visuo-spatial skills, and perhaps even social cognition. The case is based mainly on the selective loss of these abilities after certain sorts of brain damage. What is the developmental origin of such components? Imagine a scenario where a gene or set of genes was responsible for building each cognitive component. Here, despite their lowly biochemical role, the fingers of the genes would reach into the large-scale structure of the cognitive system. Evidence for this scenario might come from neurogenetic developmental disorders. If we found a disorder where the relevant gene(s) had been mutated and the individual came to exhibit a developmental a deficit restricted to the cognitive domain for which the gene(s) were responsible, this would constitute evidence for direct links between the genotype and the phenotype.

Williams syndrome was once proposed as a developmental disorder that could fit this role. In Understanding Williams Syndrome, Eleanor Semel and Sue Rosner provide a fascinating overview on how the disorder has fared as a test of direct genotype-phenotype links.

Williams syndrome (WS) is a rare disorder characterised by a unique pattern of behavioural, cognitive, emotion, and physical limitations and strengths. In addition to clinical characteristics such as heart abnormalities, short stature, and a facial dysmorphology, WS is also marked by an uneven cognitive profile. There is some degree of learning disability (IQs are typically between 50-60) but language skills are much stronger than visuo-spatial skills in the disorder. Indeed, although initially delayed, the language of individuals with WS sometimes seems precocious in their use of unusual words and conversational flourishes. Individuals with WS appear sociable and empathetic. They demonstrate particular (relative) facilities in face

recognition, in storytelling, and in musicality; but then particular weaknesses in dealing with numerical concepts, spatial cognition and in abstract reasoning. Despite the complexity of this uneven profile, the genetic underpinnings of WS are becoming increasingly well understood: there is a deletion of approximately 25 contiguous genes from one copy of chromosome 7 (see Semel & Rosner, 2003, Chapter 8; Karmiloff-Smith, Grant et al., 2003, for additional recent findings).

In their book Semel and Rosner offer a synthesis of current knowledge of WS from the perspective of The Salk Institute in San Diego, one of the main research laboratories involved in investigating the disorder. The book seeks to integrate scattered research findings on behaviour, brain, and genetics, along with clinical observations and personal accounts. Extensive use is made of two large-scale parental questionnaires to overcome the small sample sizes inevitable in the scientific studies carried out on this rare disorder.

The book is aimed at several audiences: researchers but also parents, teachers, and clinicians. As in any such endeavour, it is sometimes frustrating for one audience when another is being addressed, but the advantage for any one audience is an invaluable insight into the concerns of the others. The meat of the book is a consideration of the main strengths and weaknesses of individuals with WS. This includes sections on language, perceptual-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). However, the book also plays a crucial role in relating research to intervention. It details the range of intervention strategies available to help children and adults with WS, from task-specific interventions and naturalistic training situations to compensatory strategies and environmental manipulations. The reader is

left with a sense of encouragement that with this growing body of (sometimes very practically informed) knowledge, the prospects for individuals with the disorder to lead fulfilling lives are increasingly bright.

However, the same cannot be said for the hopes of researchers that WS would offer evidence of direct links between genotype and phenotype. Here is an example from five years ago of one such proposal, on the disparity between language and visuo-spatial skills: "Presumably [one of the lost genes in WS] plays an important role in the development of the neural networks used in spatial reasoning, possibly in the parietal lobes. The other missing genes, perhaps, are necessary for the development of other parts and processes of the brain, though not for language or face perception" (Pinker, 1999, p. 260-261). Semel and Rosner's review suggests that the type of clean fractionation between whole cognitive modules necessary to identify gene-module links is simply not present in WS. Rather fractionations appear to occur 'all the way down'. That is to say, fractionation occurs within domains to a degree of specificity of cognitive structure that seems beyond the reach of anything like targeted gene expression.

The idea that genes determine the large-scale architecture of the cognitive system relies on the assumption that genes target a particular level of cognitive granularity. This notion is expressed in Pinker's proposal, where the granularity is at a level that differentiates a 'language' component, a 'spatial reasoning' component, and a 'face perception' component. Other proposals have suggested that perhaps the language component might split into a 'grammar' and 'lexicon' sub-component open to differential genetic influence (Clahsen & Almazan, 1998). This granularity of analysis might caste the WS profile as follows: language – develops normally; face recognition – develops normally; social cognition – develops normally; musical

ability – develops normally; visuo-spatial cognition – develops atypically; memory – develops atypically; numerical cognition – develops atypically; problem solving – develops atypically; and so forth. However, as the evidence accumulates, Semel and Rosner paint a picture in which every one of these domains reveals more fine-grained levels of fragmentation on closer analysis.

Let us take the example of language discussed in Chapter 2. Language as a domain is viewed as a relative strength in WS. However, within language, individuals with WS seem to be more advanced in grammar than pragmatics. But within grammar, more errors appear in morphosyntax (verb tense agreement, personal pronouns) than in syntax (complex sentence forms such as passives and conditionals). Moreover, recent evidence suggests that even within syntax, there is greater difficulty with repeating certain types of sentence structure than others (Grant, Valian, & Karmiloff-Smith, 2002). Development is uneven within pragmatics too: there is relatively good performance in what Semel and Rosner term the 'feeling' functions of communication (social sensitivity: e.g., making eye contact, sensitivity to non-verbal cues), which contrasts with problems in other areas such as greeting behaviours, topic maintenance, and question answering. In the domain of semantics, a relative strength in category concepts (e.g., animals vs. clothing) contrasts with problems understanding semantic relational concepts such as spatial-temporal terms. Even within category concepts, recent evidence has indicated differential naming problems across categories (Temple et al., 2002; Thomas et al., 2003).

Semel and Rosner discuss other fractionations in Chapter 5 in the context of specific aptitudes within WS. Although sociability is a strength in WS, within sociability there is a fractionation between friendliness and success with adults, and the disinterest or ineptness shown when interacting with peers. There is a

fractionation between their sensitivity and understanding of others, and the difficulty they can exhibit in respecting the private space of peers. Within the domain of memory, there are fractionations between relative skill in verbal memory (e.g., in digit span) but poor performance in visuo-spatial memory (e.g., Corsi span). Within verbal (phonological) memory itself, Semel and Rosner note a fractionation between a strength in learning words but not in learning to read phonologically similar words. There is a strength in remembering semantically salient items like poems, stories, and songs over long periods, but not in learning or retaining facts over a few minutes. In musicality, in a few musically trained individuals in WS, there is a strength in composing, transposing, and performing music but a difficulty in reading music and playing instruments. To these we may add the domain of numeracy, where children with WS reveal a weakness in understanding number concepts, but mental-age appropriate learning of the count sequence (Ansari et al., 2003). And the highly salient dissociation between weaknesses in some visuo-perceptual skills (e.g., deciding which of two lines is longer) and a strength in recognising faces, discussed by Semel and Rosner in Chapter 4.

How are we to explain this level of fractionation? Recent theoretical approaches propose that many of the observed fractionations are the consequences of the processes of cognitive developmental acting on a neonatal brain that has been constructed with (perhaps subtly) altered initial neurocomputational biases. This theoretical framework has been called ‘neuroconstructivism’ (Karmiloff-Smith, 1998; Karmiloff-Smith & Thomas, 2003). The domain of face recognition provides a good example to illustrate the idea.

Face processing is a relative strength in WS, with performance on some tasks at chronological-age-appropriate levels. A fascination in faces forms part of the

'hyper-social' profile of the disorder and has been identified from an early age (Jones et al., 2001). Faces, then, are likely to be a stimulus to which children with WS have a good deal of exposure and to which rewards are attached. One might conclude that face recognition develops normally in WS, perhaps in its own, self-contained, 'preserved' module. However, both behavioural and brain studies have revealed that this is not the case. In terms of behavioural studies, research has indicated that face recognition in WS is characterised by a reliance on individual features, whereas expert recognition in normal development is characterised by the increasing use of second-order configurations or combinations of features to distinguish between individual faces (Karmiloff-Smith, 1997; Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen, 1999; though see Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003). Indeed, recent work has suggested that the ability of individuals with WS to process configurations is in fact as poor as their general visuo-spatial processing ability – the very domain from which face recognition is supposed to dissociate (Karmiloff-Smith, Thomas, et al., 2003). In terms of brain studies, brain activity has been recorded during face recognition in WS, using event related potentials (ERPs). Expert face recognition in healthy adults is associated by scalp voltage waveforms that are specialised for human faces (compared to, say, monkey faces or to cars), and predominantly localised to the right hemisphere. However, ERPs elicited in adolescents and adults with WS were found to be distributed across both hemispheres and did not distinguish between human faces, monkey faces, and cars (Grice et al., 2001, 2003; Mills et al., 2000). Face recognition appeared neither as localised nor as specialised.

However, localisation and specialisation – what one might take to be hallmarks of a functional module – turn out to be an emergent aspect of face

recognition in typical development rather than a precursor to it. ERP experiments indicate that such hallmarks are absent in young infants (de Haan, 2001; Johnson & de Haan, 2001). Across development, infant processing of human upright faces becomes increasingly localised to the fusiform gyrus in the right hemisphere, and increasingly specialised in the form of the activation of a predominantly right-lateralised waveform component (the infant equivalent of the adult N170) whenever the older infant is presented with an upright human face (Halit, de Haan & Johnson, 2003). By 12 months of age, the electrophysiology of the infant brain when processing faces begins to look relatively similar to that of adults, although development of the N170 continues throughout childhood to adolescence (Taylor, McCarthy, Saliba, & Degiovanni, 1999). In a recent review of the neuropsychology of face processing, de Haan (2001) concluded that the cortical specialisation for face processing observed in normal adults is achieved through a gradual experience-driven specialisation of an initially more general-purpose visuo-spatial processing system.

The story on face processing in WS, then, could turn out to be something like as follows. Genetic effects during brain development in WS generate initial cortical structures with different neurocomputational biases – overall processing is poorer, but the circuits have greater potential to process isolated information (features) than configurations. Due to a socio-emotional reward system operating elsewhere in the WS brain, the atypical visuo-spatial system is exposed to many faces. The visuo-spatial system then follows an atypical developmental trajectory but, because a significant proportion of faces can be identified by individual features (unlike, perhaps, other visual patterns) and because the input of faces is frequent, the system is able to achieve a reasonable level of competence. However, the system is unable to

achieve the neural organisation, specialisation, and localisation usually associated with this level of behavioural competence.

Semel and Rosner argue that such atypical cerebral organisation may be a more widespread aspect of WS. They point to evidence from lateralisation studies suggesting confused handedness and a lack of clear-cut dominance. They cite Bellugi et al.'s (1988) report that individuals with WS show greater prevalence of left-handedness in daily activities than is usual for people without a family history of left dominance and parental questionnaire results indicating increased confusion of left and right in motor tasks and body awareness.

It is unlikely that genetic effects during brain development in neurogenetic disorders are uniform across the entire brain, but this does not mean that they will be highly region specific. Differential effects are likely to be graded rather than targeting certain circuits, particularly with regard to higher cortical functions (Kingsbury & Finlay, 2001). For example, in their review of brain mechanisms in Chapter 8, Semel and Rosner note how there is evidence of a greater ratio than usual of anterior to posterior (parietal + occipital) tissue in WS. While this is consistent with deficits in visual processing associated with the posterior occipital lobes, an altered anterior/posterior ratio is far from domain-specific in cognitive terms. In short, the granularity of genetic differences in cortex is likely to be far coarser than cognitive modules.

Nevertheless, Semel and Rosner describe a highly differentiated final cognitive profile. This endstate pattern of relative strengths and weaknesses is likely the result of complex processes of development, attenuating or exaggerating initial neurocomputational differences. The usual emergence of an interactive network of neural systems may be perturbed by several factors: by the differing effect of the

atypical computational biases on the ability of various areas to process the signal with which they are provided (by the initial large scale input-output connectivity of the brain); by anomalies in the emergence of specialised circuits through pruning or competition; by compensatory changes during interactions between different brain regions; and by the atypical subjective environment to which the individual with the disorder is exposed (see Mareschal et al., in press, for discussion). The developmental result is a granularity of subsequent fractionations likely to be considerably finer than cognitive modules.

Such ideas are now being explored using several innovative methods (see Karmiloff-Smith, Scerif, & Thomas, 2002, for discussion). For instance, the cognitive deficits found in adults with WS are being traced back to their origins in infancy, to reveal the role of the developmental process in generating deficits. Thus, Paterson et al. (1999) have demonstrated how the relative competencies of infants, children, and adults with WS and Down's syndrome alter across development, as the abilities follow different atypically constrained trajectories. Semel and Rosner discuss the work of Bellugi and colleagues, which demonstrates the relative trajectories of language, face processing, and visuo-spatial processing in WS from childhood through to adulthood. Face processing is always higher than would be expected for their mental age. Language in WS is not an early strength (see Laing et al., 2002) but starts to accelerate from late childhood and adolescents onwards. By contrast, visuo-spatial processing is markedly impaired at all ages, with the acquisition curve flattening out almost completely by adolescence (Bellugi et al., 2000; see Farran & Jarrold, 2003, for discussion of recent findings).

Another new approach to investigating the disorder is the use of computational modelling techniques. In this work, connectionist models of developing cognitive

systems have their initial computational biases disrupted. This permits exploration of the consequences of the initial conditions on the subsequent acquisition of cognitive domains such as language, and the role that development can play in compensating for or exaggerating initial anomalies (e.g., Thomas & Karmiloff-Smith, 2002a, b, 2003). In particular, an identical deficit introduced either at the outset of learning or at the end of learning can give rise to very different outcomes.

Overall, the hope is that as our understanding of the atypical constraints that shape development in neurogenetic disorders increases, so too will our understanding of the constraints that act on normal development. Echoing previous authors such as Karmiloff-Smith (1998) and Mervis and Klein-Tasman (2000), Semel and Rosner conclude that the behavioural characteristics of WS are 'manifestations of a developmental process involving cascades of genes, brain mechanisms, and transactions with the environment at all levels, including interventions' (p.375).

There is plenty more in Semel and Rosner's book than I have had space to describe. For instance, the authors attempt to identify sub-groups within WS and to distinguish prototypical from associated features of the disorder. Much of the content is aimed at parents and teachers for practical use. To give but a flavour, if laterality is a problem for a child with WS, then when you are teaching her a common motor skill such as doing up buttons or tying shoe laces, sit next to her rather than across from her in a mirror configuration. This means the child doesn't have to mentally reverse left and right to copy the behaviour!

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 more recent work (language development: Thomas et al., 2003; learning to read: Laing et al., 2001; number concepts: Ansari et al., 2003).

To the theorist, however, this book is sufficient to confirm early intuitions that WS can provide an important window on genotype–phenotype relations. The difficulty is that (perhaps unsurprisingly from the neuroconstructivist stance) our first peek through this window has revealed nothing simple or direct about the connections between the genetic and cognitive levels.

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Williams syndrome is a genetic condition that causes various developmental and health problems such as ADHD, anxiety, phobias, a short nose with a broad tip, full cheeks, and a wide mouth with full lips. Genetic causes, treatments, and life expectancy information are provided.Â What other names do people use for Williams syndrome? Signs/Symptoms. What are the signs and symptoms of Williams syndrome? Causes. What genetic and chromosomal changes are related to Williams syndrome? Williams syndrome occurs when a person has an entire chromosome arm missing from their DNA. Symptoms include cardiac problems and shortness of breath.Â That's why in their first ten months, babies with Williams Syndrome may be colicky and irritable. Fortunately for all the parents out there, this problem normally sorts itself out with age. Advertisement. Irregularities In The Teeth. Kids with Williams Syndrome may wind up seeing the inside of the dentistâ€™s office more than other children. Your dentist may have to tackle issues like tooth decay, gaps, and missing teeth to start. The good news for parents is that normal dental hygiene can help with managing many of these concerns. Williams Syndrome: Fractionations All the Way Down?. Cortex, Vol. 42, Issue. 7, p. 1053.Â Patterns of syntactic development in children with Williams syndrome and Down's syndrome: Evidence from passives and whâ€questions. Clinical Linguistics & Phonetics, Vol. 21, Issue. 9, p. 705. Williams syndrome: fractionations all the way down? Review of Understanding Williams Syndrome, by Eleanor Semel and Sue Rosner. ISBN 0-8058-2618-1, Lawrence Erlbaum Associates, 2003, 456 pages. Price: US $ 49.95, UK Â£ 34.50. In healthy adults, there is some case to be made Rosner, 2003, Chapter 8; Karmiloff-Smith et al., that discrete cognitive components or functional 2003, for additional recent findings). circuits underlie different high-level abilities such In their book Semel and Rosner offer a as language, face recognition, visuo-spatial skills, synthesis of current knowledge of WS from t @article{Thomas2006WilliamsSF, title={Williams Syndrome: Fractionations All the Way Down?}, author={M. Thomas}, journal={Cortex}, year={2006}, volume={42}, pages={1053-1057} }. M. Thomas. Published 2006. Psychology. Cortex. View on Elsevier. bbk.ac.uk.Â I. The Neurocognitive Profile of Williams Syndrome: A Complex Pattern of Strengths and Weaknesses. U. Bellugi, L. Lichtenberger, W. Jones, Z. Lai, Marie St. George. Psychology, Medicine. Journal of Cognitive Neuroscience. 2000. 643. PDF.