

Dethroning the Myth: Cognitive Dissociations and Innate Modularity in Williams Syndrome

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Despite increasing empirical data to the contrary, it continues to be claimed that morphosyntax and face processing skills of people with Williams syndrome are intact. This purported intactness, which coexists with mental retardation, is used to bolster claims about innately specified, independently functioning modules, as if the atypically developing brain were simply a normal brain with parts intact and parts impaired. Yet this is highly unlikely, given the dynamics of brain development and the fact that in a genetic microdeletion syndrome the brain is developing differently from the moment of conception, throughout embryogenesis, and during postnatal brain growth. In this article, we challenge the intactness assumptions, using evidence from a wide variety of studies of toddlers, children, and adults with Williams syndrome.

Neurocognitive studies of developmental disorders never turn out to be as straightforward as they first promise. Studies of Williams syndrome (WS) are no exception. The pioneering work of Bellugi, Lichtenberger, Jones, Lai, and St. George (2000) seemed to point to some clear-cut dissociations in the cognitive architecture of WS. Language and face processing appeared to be preserved in the face of both general retardation and particularly serious problems with visuospatial cognition, number skills, planning, and problem solving (Bellugi, Wang, & Jernigan, 1994). Researchers in the field of WS have been cautious about these claims, couching them in terms of relative strengths and weaknesses rather than absolute ones (Bellugi et al., 2000; Karmiloff-Smith, 1998; Karmiloff-Smith

et al., 1997; Klein & Mervis, 1999; Mervis, 1999; Vicari, Carlesimo, Brizzolara, & Pezzini, 1996; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996). By contrast, secondary sources regarding WS data, cited in writings by linguists, psychologists, and philosophers, have often used WS to bolster claims about innate and independently functioning modules, some of which are intact and others impaired (e.g., Bickerton, 1997; Pinker, 1994, 1999). This emanates from a view, held explicitly or implicitly, that behavioral deficits found in the phenotypic outcome of individuals with genetic disorders are direct windows on the initial state, that is, the innate modular structure of the cognitive system (Baron-Cohen, 1998; Leslie, 1992; Temple, 1997; see Karmiloff-Smith, 1998, for critical discussion). As Baron-Cohen (1998) put it,

I suggest that the study of mental retardation would profit from the application of the framework of cognitive neuropsychology. In cognitive neuropsychology, one key question running through the investigator's mind is 'is this process or mechanism intact or impaired in this person'?

The notion that an ability is necessarily intact in a genetic disorder when behavior falls within the normal range fails to consider the psychological processes underlying overt behavior. This kind of reasoning negates the role of development in producing phenotypic outcomes and treats the end-state cognitive system as if it were a normal system with some components missing and others intact. In other words, it is based on the neuropsychology model of brain damage to previously normal adults and can, in our view, be very misleading when applied to developmental disorders. Further, as far as WS is concerned, the nativist literature frequently misrepresents the empirical findings, treating relative strengths as absolute strengths. In the first part of this article, data relating to the phenotypic outcome of two areas hailed as intact in WS—language and face processing—are examined. In the second part, we look at the early cognitive state in toddlers with WS and consider its relationship to the adult end state.

LANGUAGE AND WILLIAMS SYNDROME

In our view, it remains questionable as to whether any aspect of language—syntax, semantics, phonology, or pragmatics—is intact in WS. Yet a number of researchers have tried to demonstrate that language, in particular morphosyntax, is preserved in WS and functions independently of other cognitive systems. Rossen, Jones, Wang, and Klima (1995), for example, claimed that “Williams syndrome presents a remarkable juxtaposition of impaired and intact mental capacities: linguistic functioning is preserved in WS while problem solving ability and visuospatial cognition are impaired.” Likewise, Pinker (1991) claimed that:

Although IQ is measured at around 50, older children and adolescents with WS are described as hyperlinguistic with selective sparing of syntax, and grammatical abilities are close to normal in controlled testing. This is one of several kinds of dissociation in which language is preserved despite severe cognitive impairments.

Not all researchers make such sweeping claims, but many linguists of a Chomskyan persuasion nonetheless try to find an aspect of WS language that is spared and, by extension, innately specified. For example, Clahsen and Almazan (1998) argued for a double dissociation of innate mechanisms, on the basis of their claim that in WS lexical memory is impaired and syntax is intact, whereas in specific language impairment (SLI) the opposite obtains. These authors used evidence from a number of syntactic elicitation and comprehension tasks. These included tests of past tense formation, expressive language, and the interpretation of passive sentences and of anaphoric and reflexive pronouns. Performance of the individuals with WS on the latter two tasks was at ceiling. However, ceiling effects are notoriously difficult to interpret because they can simply suggest that a task is not sensitive enough. Furthermore, Clahsen and Almazan's arguments were based on a very small sample of children with WS ($n = 2$ for mental age [MA] 5 years and $n = 2$ for MA = 7 years), together with considerable interindividual variation among the few participants. Strong claims about the cognitive architecture of a syndrome cannot be made on the basis of such sparse data.

The main claim of the Clahsen and Almazan (1998) study was that individuals with WS have a specific deficit in forming irregular past tenses (e.g., creep–crept) but intact performance in forming the regular past tense (e.g., walk–walked). Because this important claim was based on such a small sample with individual variation, we carried out a much broader, in-depth study of past tense formation (Thomas et al., 2001), comparing the performance of 21 participants with WS on two past tense elicitation tasks with that of four typically developing control groups at ages 6 years, 8 years, 10 years, and adult. Given that WS language is seriously delayed initially, Thomas et al. (2001) argued that it is not sufficient to show that irregular past tense formation is poorer than regular past tense formation, because this is also true of some stages of typical development. Rather it is necessary to demonstrate that the level of past tense formation is poorer than would be expected in individuals with WS for their actual level of language development. The study showed that when performance was related to chronological age (CA) using regression analyses, individuals with WS showed a somewhat greater disparity between irregular and regular verbs compared to the controls. However, when verbal MA was controlled for, the WS group displayed no selective deficit in irregular past tense formation. Moreover, we could not replicate the Clahsen and Almazan (1998) control data. At no age did our controls show high levels of irregularization of novel verbs that rhyme with irregular verbs (see also van der Lely & Ullman [2001] for similar control data results to ours). Furthermore, our

results also highlighted how potentially misleading small samples such as in the Clahsen and Almazan (1998) study can be. As individuals, a few of our participants with WS performed very poorly on both regular and irregular verbs, whereas a few others displayed very high performance on both. If these high performers had by chance constituted the very small *N* of the Clahsen and Almazan study, then the authors would have had to draw totally different conclusions from the ones drawn. Our findings on a much larger population of 21 individuals with WS are inconsistent with the view that people with WS are selectively impaired on irregular past tense forms. Indeed, as a group, there was no selective deficit for irregulars and the WS results could be placed on the typical developmental pathway found in younger individuals. The results were in fact consistent with the hypothesis that the WS language system is delayed because it has developed under different constraints. Mervis and her collaborators (e.g., Klein & Mervis, 1999) have also concluded that the best way to characterize WS language is that it is delayed, revealing patterns typical of younger children.

A number of findings now suggest, however, that the WS language system is not only delayed but also develops along a different trajectory compared to controls, with individuals with WS placing relatively more weight on phonological information and relatively less weight on semantic information. For example, during the early acquisition of language, the naming spurt in WS precedes fast-mapping ability, whereas in typical development these two are closely associated (Mervis & Bertrand, 1997). These same authors also showed that the naming spurt in WS does not coincide with exhaustive category sorting, an index of children's maturing semantic representations, which suggests that vocabulary growth relies less on semantics than in the typically developing case (Mervis & Bertrand, 1997). Further, although local semantic organization looks normal in WS in terms of priming effects (Tyler et al., 1997) and in terms of category fluency (Scott et al., 1995), global semantic organization remains at the level of young children and never reaches the mature state even in relatively high functioning adults with WS (Johnson & Carey, 1998).

A number of other studies of oral and written language also point to a reduced contribution of semantics in language development in WS. For example, Karmiloff-Smith et al. (1997) found that when participants with WS were monitoring sentences for a target word, they did not show sensitivity to subcategory violations, suggesting that in WS semantic information may become available too slowly to be integrated with the online processing of syntax. A recent study of reading in WS came to similar conclusions about the role of phonology over semantics. The group with WS displayed equal levels of reading for both concrete and abstract words (Laing, Hulme, Grant, & Karmiloff-Smith, 2001). By contrast, the controls found concrete, imageable words much easier to read. In addition, the study showed that imageability effects are weaker in people with WS. Finally, Grant et al. (1997) used the Children's Nonword Repetition task (Gathercole,

Willis, Baddeley, & Emslie, 1994) with participants with WS. They showed that, despite a vocabulary test age of 8 years, when learning new words people with WS behaved like 4- to 5-year-olds and did not show the pattern seen from 6 years onward in the typically developing population. Like very young children, the participants with WS were less influenced by the semantics of the words that the nonce terms resembled and relied more on phonology. Taken together, these different studies suggest that, unlike typical development, semantics seems to place somewhat less of a constraint compared to phonology in the way in which WS language develops over time.

We have so far suggested that semantics may play a somewhat less important role in WS lexical development than in typical controls and that this aspect of WS language develops atypically. However, it remains possible that WS syntax is intact, as many have claimed (e.g., Bickerton, 1997; Clahsen & Almazan, 1998; Pinker, 1999). There are, however, a number of lines of evidence that cast doubt on this. First, vocabulary levels are usually better than syntactic levels in WS on various standardized tasks, and both are significantly below CA (Karmiloff-Smith et al., 1997). Second, even in very simple imitation tasks, participants with WS show impairment with complex syntactic structures like embedded relative clauses. A recent study by Grant, Valian, and Karmiloff-Smith (2002) showed that despite having a mean vocabulary test age of 9 years, the participants with WS performed significantly worse on relative clauses than the 6- and 7-year-old controls and worse than even the 5-year-olds on three of the four sentence types. Length of sentence did not explain the results because the shortest of the sentence types was the most difficult for the WS group who performed at ceiling on non-embedded filler sentences of varying length. These findings are inconsistent with the view that WS syntax is intact. Even in an area of relatively simple syntax-grammatical concord over sentence elements—which normal French-speaking children acquire easily and early—people with WS show impairment. Karmiloff-Smith et al. (1997) studied the ability of a group of French-speaking participants with WS to use grammatical gender agreement. The results showed that although the children with WS learned the local gender marker (correct article) for a nonce term easily (in fact, more easily than control children), their capacity for gender agreement across sentence elements such as agreement on adjectives or pronouns was seriously impaired. Even for known words, the WS group made double the number of errors of the young controls. This suggests that memory for local verbal material (article + noun) is good, but processing of sentential syntax (gender agreement across sentence elements) is not. Studies of Italian-speaking children have also revealed that grammatical gender is a particular problem, with children with WS displaying errors never encountered in typical development (Volterra et al., 1996). Several studies (e.g., Klein & Mervis, 1999) now suggest that the problems that people with WS have with semantics and syntax can often be camouflaged by their good verbal memory.

Despite these and numerous other linguistic data from studies of WS, the myth that WS morphosyntax is intact continues to thrive. This is clear from the following quotation from Pinker's (1999) recent book, where he contrasts individuals with SLI and WS, respectively: "The genes of one group of children impair their grammar while sparing their intelligence; the genes of another group of children impair their intelligence while sparing their grammar" (p. 262).

It is in our view theoretically misleading and empirically inaccurate to claim that grammar is spared in this clinical population. WS grammar is relatively good compared to some other clinical groups and relatively good compared to WS spatial deficits, but no better than their MA would predict. These are relative descriptions, not absolute ones. One of the crucial features of WS language is that in infancy and toddlerhood it is initially seriously delayed (Mervis, Morris, Bertrand, & Robinson, 1999; Mervis, Robinson, & Pani, 1999; Singer Harris, Bellugi, Bates, Jones, & Rossen, 1997). Now, if the WS infant brain presented with an intact morphosyntactic module, as many such quotations suggest, then this severe delay would surely be surprising. But given the empirical facts, it is not. The myth of intact WS language needs to be dethroned and buried once and for all. This does not mean that the WS cognitive architecture is uninteresting. On the contrary, we need to understand why the language of people with WS language is initially so delayed (Laing et al., 2002; Nazzi, Paterson, & Karmiloff-Smith, in press) and why it develops atypically. We will look at the issue of early development, with respect to language, number, and spatial cognition, in the third part of this article. Prior to doing so, we consider another aspect of the WS cognitive architecture—face processing—that is also claimed to be intact.

FACE PROCESSING SKILLS IN WILLIAMS SYNDROME

As with language, initial claims about face processing in WS suggested an innately specified face processing module that is intact. Indeed, Bellugi, Birchle, Jernigan, Trauner, and Doherty (1990) asserted, "we find in the WS population *normal* face processing capacities with at floor performance on spatial tasks," and Rossen et al. (1995) claimed to have found "*selective preservation* [italics added] of face recognition in Williams syndrome." There is no doubt that people with WS are very proficient at face processing. One might ask if face processing in adults with WS is modular, and the reply could be affirmative, that is, it has become modularized with development. One might also ask: Is it an intact module? But this is the wrong question because it negates development and the possibility that the cognitive processes underlying proficient WS face processing are different from those of typically developing controls. Indeed, several studies (Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen, 1999; Karmiloff-Smith, 1998; Udwin & Yule, 1991) have replicated Bellugi's earlier work and revealed normal

or near-normal behavioral scores on standardized tasks like the Benton Facial Recognition Test (Benton, Hamsher, Varney, & Spreen, 1983) and the Rivermead Behavioural Memory Test (Wilson, Cockburn, & Baddeley, 1985). But these same studies have seriously challenged the notion that the behavioral success displayed in WS face processing capacities is normal. It has been shown that whereas typically developing controls use predominantly configural processes to recognize faces, people with WS tend to use predominantly componential or featural processes and do less well when a task forces configural processing (Wang, Doherty, Rourke, & Bellugi, 1995). Under certain circumstances, they are capable of using global or configural processing, particularly in low-level perceptual tasks (Birhle, Bellugi, Delis, & Marks, 1989; Mervis et al., 1999; Pani, Mervis, & Robinson, 1999), but they show a stronger tendency toward featural processing in many low-level and higher level visuospatial tasks, including face processing.

In an elegant set of studies using faces, buildings, and geometric shapes, Deruelle et al. (1999) showed that when faces and buildings are inverted, typically developing controls display a significant inversion effect for faces (they are faster and more accurate for upright faces) but not for buildings. By contrast, although the performance of the group with WS decreased slightly with inverted faces, this decrease was not significantly greater than that observed for buildings. The lack of the face inversion effect is not attributable to a floor effect because the WS accuracy scores were similar to those of their MA matches who did exhibit an inversion effect with faces. Furthermore, although the WS group ranged from 7 to 23 years, there was no trend with age toward the typical pattern. This finding was also supported using another set of geometric stimuli. Deruelle et al. gave participants the choice between similarity on the basis of configuration or similarity on the basis of features. For example, a square composed of four tiny circles might be placed with a square composed of four tiny squares (same configuration, different features) or a rectangle composed of four tiny circles (same features, different configuration). Control participants of either the same CA or the same MA tended to choose patterns of the same configuration, whereas the WS group showed no such preference. With a match-to-sample design using a set of schematic faces in which either configuration or features were changed, the WS group did not differ in the number of errors made on local features, but showed severe deficits compared to the controls in the configural trials. The authors conclude that individuals with WS display a selective configural processing deficit compared to both CA and MA matches. Their face processing proficiency stems from a deviant developmental pathway and does not reveal the functioning of a so-called normal, intact module. So, it is not the case that people with WS have an intact face processing module and an impaired space processing module. Both follow atypical developmental trajectories.

Imaging studies focusing on the electrophysiology of face processing in WS also support the notion of a differently developing expertise rather than an intact module. In a face recognition (match/mismatch) event-related potentials (ERP) study of 18 adults with WS, Mills et al. (2000) found abnormalities in the early waveform (100 and 200 msec post stimulus onset) of each of the participants with WS. This was not found in any of the controls. The authors suggest that these differences index abnormalities in face perception that may be specific to WS. Another study also points to abnormalities in face processing in WS. Using high-density ERP and a simplified task of face perception, Grice et al. (2001) tested 18 individuals with WS ($M CA = 21.4$ years) and also found waveform differences compared to CA-matched controls that indicated both deviance and delay. The N170 face-sensitive component was abnormal in the WS group and, unlike controls, was not increased in amplitude to inverted faces. There was also less right lateralization than for controls. In addition, unlike the control group, there was no difference in the N170-equivalent component to human faces or monkey faces. This finding suggests that the individuals in the WS group are not specialized for human faces in the same way as are controls. These data again refute the idea of an 'intact' module. Rather, they suggest that people with WS have either an incomplete or a different form of modularization for face processing.

EARLY DEVELOPMENT IN WILLIAMS SYNDROME

We now turn to early development with respect to these two areas of relative proficiency in WS language and face processing. Our aim (Paterson, 2000; Paterson, Brown, Gsödl, Johnson, & Karmiloff-Smith, 1999) is to challenge some of the deeply engrained assumptions in cognitive neuropsychology and developmental cognitive neuroscience about the use of developmental disorders for bolstering nativist claims. The assumption—which we will call the Modular Continuity Hypothesis—often remains implicit in writings, but is in fact part and parcel of the logic of the argument and stems, as we suggested in the Introduction, from adult neuropsychology models. It holds that the brain is organized into innate (genetically determined) mental/neural modules that have the same potential for dissociation across the human lifespan. In other words, it is assumed that there is a transparent relationship between phenotypic outcomes and genes, with the expectation that the same dissociations observed in the adult steady state hold during the period in which these abilities emerge.

Is the inference that the WS phenotypic end state supports the case for innate modularity justified? In other words, can one assume the state of early development simply from the pattern of proficiencies and impairments in the phenotypic outcome in the adult, without studying their developmental trajectories? It is known that WS and Down syndrome (DS) display different cognitive profiles in

the end state (Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993; Klein & Mervis, 1999; Wang, Doherty, Hesselink, & Bellugi, 1992), although, using more subtle measures, Klein and Mervis (1999) discovered a number of hitherto neglected similarities between WS and DS at 9 to 10 years of age. In adulthood, however, it remains clear that vocabulary levels of people with WS are better than those with DS and that both syndromes show serious impairment in the domain of number (Bellugi et al., 1994; Paterson, 2000).

Paterson (2000; Paterson et al., 1999) purposely chose two tasks—one language-related, one number-related—which could be designed to be as similar as possible for both very young children and adults. For number, numerosity judgments were required; for language, receptive vocabulary measures were taken. The domains of vocabulary and number were purposely chosen because in the phenotypic end state it had been claimed that individuals with WS show greater proficiency in vocabulary than individuals with DS, and that both syndromes are seriously impaired in number. If the phenotypic end state can be directly used to assume the pattern obtaining in infancy, then the infant profiles should resemble the adult profiles across these two syndromes. Paterson (2000) first examined adult abilities. She tested participants with WS and DS who were matched on CA and on MA from the British Ability Scales, $t(13) = 2.05$, $p > .06$. She showed that these adults had significantly different scores on a vocabulary test, the British Picture Vocabulary Scales (BPVS), with a smaller discrepancy between CA and test age on the BPVS for adults with WS than for those with DS, $t(6) = 2.55$, $p < .05$, as had been demonstrated in previous work. Numerosity judgment tasks had not been hitherto used with adults with either WS or DS. Participants were required to judge which of two numbers (either Arabic numerals 1–10 or dots displays) displayed on a computer screen is the larger. Reaction times and accuracy were measured. In the normal case, a distance effect is always apparent: Numbers very close (like 7 and 8) take longer for a decision as to which is the larger than numbers that are far apart (like 7 and 2). Paterson demonstrated that adults with WS and DS performed differently on the numerosity judgment tasks. The adults with DS, although slower overall, showed a clear-cut effect as evidenced by typically developing controls. By contrast, the adults with WS performed significantly worse than the matched adults with DS on several number tasks and, although there was a trend in the right direction, this WS group did not show the distance effect. There was a significant difference between the WS and DS groups for the discrepancy between reaction times to close and far pairs (Mann–Whitney $U = -11$, $p < .05$). So the phenotype in the adult end state was DS significantly worse than WS on vocabulary, WS significantly worse than DS on numerosity judgments.

An attempt was made to devise similar number tasks with adults and toddlers. In both cases, the tasks involved making a comparison between two numerosities. In the toddler case, an implicit same/different judgment was required, whereas the

adults had to decide which of the numerosities was the larger. Likewise, vocabulary comprehension was measured in both adults and toddlers.

Paterson et al. (1999) used a preferential looking paradigm to examine numerosity and vocabulary in 65 toddlers between 13 and 36 months, divided into groups of toddlers with WS, atypical controls with DS matched for CA and MA on the Bayley Infant Scales II (Bayley, 1993), typically developing MA controls (also matched on the Bayley), and typically developing CA controls. If the use of the so-called Modularity Continuity Hypothesis were justified, then these young children should show a similar profile of cognitive abilities and impairments to adults in each of the syndromes. However, this was not the case. The toddlers with WS and DS were equally impaired and performed significantly worse than CA controls on the language task, despite the fact that adults with WS were significantly better than the adults with DS. For vocabulary, then, both atypical groups of toddlers performed like the MA controls, that is, at approximately half their CA. By contrast, although the adults with WS were more impaired than the adults with DS with numerosity judgments, the toddlers with WS showed unimpaired performance on the numerosity judgment task. They performed like the CA controls, whereas the toddlers with DS were seriously impaired and did not even reach the level of the MA controls. Again, the pattern in early development differed considerably from that observed in adulthood. Caution must therefore be exercised when making claims about innate modules based on phenotypic outcomes. These data suggest either that the learning trajectories of the two syndromes are different in development or, in the language example, that children with DS are subject to increasing deficit in linguistic skills compared to their counterparts with WS, who retain a relatively stable pattern of delay throughout development. Whichever turns out to be the case, it is only via developmental studies following infants and toddlers from the initial state through childhood and adulthood that this question can be properly addressed. Alas, in WS, a syndrome characterized by initial feeding problems and failure to thrive, it is very hard to test infants in the very early phases of postnatal life. Even as late as 20 months, however, the toddler profile is different from the resulting adult profile. This again stresses the need to focus on the process of development itself when studying developmental disorders (Karmiloff-Smith, 1998) and that claims regarding starting states cannot necessarily be based on patterns found in phenotypic outcomes. In other words, although it is always possible that in some cases early developmental patterns turn out to show the same profile of abilities and impairments as the adult end state, this cannot be taken for granted without empirical verification. Furthermore, although the adult profile concerns higher level cognitive domains, it is probable that impairments in infancy are related to much lower level processing mechanisms.

With respect to language, we are left with an intriguing question as to why onset in WS is so delayed, given the relative proficiency in later life. We have argued

that this may in part be due to an imbalance between semantics and phonology leading to weak semantic representations. It may also in part be due to abnormality of nonlinguistic precursors to language. For instance, Mervis and Bertrand (1997) showed that, unlike typical development, in WS naming precedes pointing. Thus, early naming in WS may be more like sound production and less like language production. Results from recent work in our lab also suggest that pointing and joint attention, which underpin certain aspects of normal language acquisition, are significantly reduced and atypical in toddlers with WS compared to MA and CA controls (Laing et al., in press). We are also examining early speech perception and have shown that infants with WS have deficits in early speech segmentation abilities (Nazzi et al., in press). It is in our view crucial to examine the array of prelinguistic abilities that the typically developing child brings to the language-learning task. It is clear that throughout development, however, from the early stages through to the adult end state, WS language is not intact at any level that researchers have hitherto seriously examined. Let us reiterate, the myth of intact language should be buried and the search for intact modules ceased, so that more subtle research can be pursued to discover the developmental language trajectories followed by individuals with WS. The lack of intactness is unsurprising if we recall that the WS brain is qualitatively different from the normal brain in terms of brain anatomy (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999), brain chemistry (Rae et al., 1998), and computational processing (Grice et al., 2001; Mills et al., 2000). Nor is it therefore surprising that seemingly normal behavioral outcomes even in adults turn out to be underpinned by different linguistic processes from the normal case (Karmiloff-Smith, 1998).

What about face processing in early development? Can that provide clues to the atypical processing style used by adults with WS? First, it is known that normally developing infants already show a right hemisphere superiority for configurational face processing from 4 to 5 months of age onward (Deruelle & de Schonen, 1998; de Schonen & Mathivet, 1990). So the right hemisphere bias in normal adults is already present early in infancy. Second, by 1 month of age, typically developing infants show a novelty preference for new faces over old faces in a preferential looking task. By 3 months of age, not only do they display a novelty preference, but they also show a prototype effect (de Haan, Johnson, Maurer, & Perrett, 2001). Take the following experimental situation. Four faces are displayed one after another and then the infant is given a choice between a fifth face (not previously seen by the infant, but a prototype morphed from the previous four faces) and one of the already-seen faces. In this situation, 3-month-olds (but not yet 1-month-olds) treat the morphed face as more familiar than the already seen face. This means that by 3 months, infants do not simply learn the details of exemplars, but build up a prototypical representation of the configuration of previously processed faces. In a pilot study, Brown (2000) found that infants with WS

did not show the prototypical effect. This suggests that infants and toddlers with WS tend to learn exemplars and may lack the generalization processes necessary to form prototypes. If this preliminary finding holds, the WS lack of prototype extraction could well be due to a tendency early in development toward featural rather than configural processing. Because it has been shown that toddlers with WS spend more time than typically developing or DS controls fixated on faces (Mervis et al., 2003), the early infant processing style may explain the phenotypical outcome in WS adult face processing. It may also be a clue as to why infants with WS succeed in discriminating small numerosities compared to infants with DS: The former group's seemingly normal performance may actually rely on a focus on qualitative detail rather than quantity.

Yet again we need to bury the myth of what at first blush seemed like an intact face processing module in adults with WS. Face processing follows a different developmental trajectory in this clinical population.

It has always been recognized that children and adults with WS show clear behavioral deficits in visuospatial tasks outside face processing. Our lab has also pursued spatial cognition in infancy (see also Atkinson et al., 1997). In a study of saccade planning, Brown and colleagues (Brown, 2000; Brown et al., in press) showed that toddlers with WS (with a mean CA of 29 months, range 23–37 months) have atypical spatial representations for planning visually guided actions. Saccades in healthy controls and in CA/MA-matched toddlers with DS (with a mean CA of 29 months, range 24–37 months) are executed within body-centered spatial coordinates. By contrast, toddlers with WS displayed evidence of deficits in saccade planning, suggesting in their case a greater reliance on subcortical processing mechanisms than the other groups. So once again, be it face processing or visuospatial processing, the WS brain proceeds along an atypical developmental pathway. Moreover, in both the prototype face processing and saccade planning studies, toddlers with WS displayed sticky fixation, that is, they looked longer at all the displays than participants with DS and the CA- and MA-matched typically developing controls. It is therefore possible that subsequent focus on features is the result of an early inability to disengage. To reiterate, it is not that one module—face processing—is spared and the other module—visuospatial processing—is impaired. Both domains develop atypically in WS.

CONCLUDING COMMENT

In our view, the notion of direct impairment to higher level cognitive modules is unlikely to explain the phenotypic outcome in WS or other genetic disorders. We anticipate that impairments will be in the form of lower level mechanisms traceable back to early infancy. There may be cases where infant and adult impairments turn out to be similar, but our new work stresses the importance of not taking for

granted the idea that the infant start state will display the same profile as the adult end state. As we stated in the Introduction, neurocognitive studies of developmental disorders never turn out to be as straightforward as they first promise. We have shown that studies of WS are no exception, and that it is time that the myths of static, intact modules be dethroned in favor of studying the complex dynamics of developmental trajectories.

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