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Dans Enfance 2003/3 Vol. 55 , pages 237 à 249
Éditions Presses Universitaires de France

ISSN 0013-7545
ISBN 2130533639
DOI 10.3917/enf.553.0237

Language and non linguistic cognition in children with Williams syndrome : A complex interaction

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SUMMARY

The approach taken in this paper is to use language, especially narratives as a means to better understand the relation of language to other cognitive domains: spatial cognition and affect. Using the disparate developmental trajectories and separability as a starting point, we first concentrate on studies of language development in children with WMS and then use narratives as a context to explore later language and the intersection of language with these other cognitive domains: spatial cognition and then affective expression.

Key-words : Language development, Spatial cognition, Affect, Williams Syndrome.

RÉSUMÉ

Langage et cognition non linguistique chez les enfants atteints du syndrome de Williams : une interaction complexe

Dans cet article, nous avons pris le parti d'utiliser le langage, et en particulier la narration, comme un moyen de mieux comprendre la relation du langage avec d'autres domaines cognitifs comme la cognition spatiale et l'affect. En prenant pour point de départ la disparité des trajectoires développementales et la possibilité qui s'offre ainsi de les séparer, nous nous concentrons d'abord sur les études concernant le développement du langage chez les enfants atteints du syndrome de Williams, puis nous utilisons la narration comme contexte d'exploration du langage plus avancé et de ses connections avec d'autres domaines cognitifs : la cognition spatiale et l'expression des affects.

Mots clés : Développement du langage, Cognition spatiale, Affect, Syndrome de Williams.

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ENFANCE, n° 3/2003, p. 237 à 249

1 / INTRODUCTION

Interest in Williams syndrome, a rare genetic disorder, stems from the uneven cognitive profile that is associated with the syndrome, including apparent dissociations in cognitive functions. Individuals with WMS are characterized by a unique physiological and behavioral profile characterized by excessive sociability with relatively spared linguistic abilities in the face of mild to moderate mental retardation and significant impairments in spatial cognition (Bellugi, Lichtenberger, Jones, Lai, & St-George, 2000). In this paper, we review a series of formal and informal studies of language development in English speaking children and adolescents with Williams syndrome (WMS) and then investigate how their use of language intersects with other cognitive domains to better understand the relation between language and non-linguistic cognition.

Williams syndrome is a rare developmental disorder that typically results in specific facial features, a characteristic heart defect, and mild to moderate mental retardation. Recent studies are also finding a characteristic cognitive and neuroanatomical profile for WMS. This sporadic disorder (approximately 1 in 20,000 live births) has as its genetic basis a hemizygous deletion encompassing the elastin gene locus on chromosome 7 (*i.e.,* one copy of a small set of genes, including elastin, LIM1-kinase, syntaxin 1a, and other surrounding genes) and this deletion is present in 96% or more of clinically diagnosed WMS individuals (Ewart, Morris, Atkinson, Weishan, Sternes, Spallone, Stock, Leppert, & Keating, 1993; Frangiskakis, Ewart, Morris, Mervis *et al.,* 1996; Korenberg, Bellugi, Salandanan, Mills, & Reiss, 2003; Morris, Thomas, & Grenberg, 1993).

2 / GENERAL COGNITIVE FUNCTIONING

Across both verbal and non-verbal standardized conceptual and problem-solving tasks, subjects with WMS demonstrate a consistent, serious impairment in general cognitive functioning. Most individuals with WMS typically perform significantly below what is considered "normal" for their age, with global standard scores on IQ tests ranging from 40 to 90 with a mean of 55 (Bellugi, Klima, & Wang, 1996). In Figure 1 we see the range of Wechsler Full Scale IQ (Wechsler, 1974; Wechsler, 1981) of 82 subjects with WMS, compared to a typical normal distribution of scores for Full Scale IQ. The range of performance on IQ tests, shown in Figure 1, also depicts the variability within WMS group; overall, their mean is shifted downward from the normal distribution into the mild to moderate range of mental retardation.

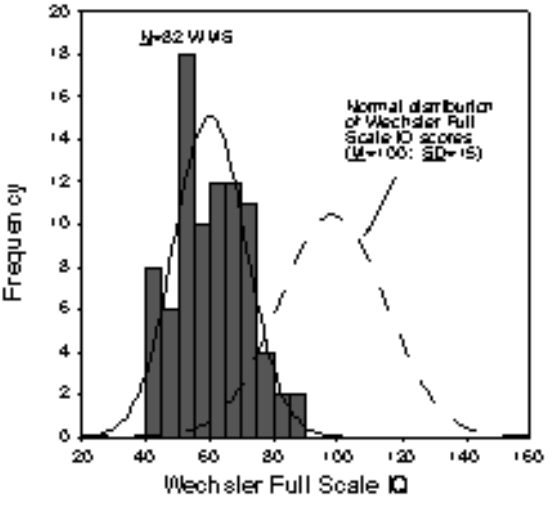

Fig. 1. — Distribution of IQ's in Williams Syndrome
Wechsler Full Scale IQ's in WMS range from 40 to 90,
and they are fairly normally distributed,
with a mean IQ of approximately 55 (SD=11)

3 / DISTINCT DEVELOPMENTAL TRAJECTORIES ACROSS COGNITIVE DOMAINS

In spite of the global cognitive impairment as measured by IQ, previous studies have shown that WMS results in a characteristic profile of specific strengths and deficits, across as well as within cognitive domains. To further explicate this uneven profile, Jones, Rossen and Bellugi (1995) examined development in a large cross-sectional group of 71 participants with WMS, ages 5 to 29 years (mean age 14;9 years). They used standardized measures of receptive vocabulary, visual-motor integration and face recognition, to look at the developmental patterns across cognitive domains using individuals with DNS as a control group.

The WMS group exhibited a distinctive profile on each of the three tasks during school-age, adolescence, and adulthood (see Fig. 2). In the WMS cohort, distinct development trajectories were found for each target cognitive domain. Specifically, in the area of receptive vocabulary, children with WMS were delayed initially, but then showed improvement with increasing age. Development in this domain continued well into adulthood. Contrasting strikingly with their vocabulary development, the WMS group showed marked impairment and a relatively flat profile in the visuo-spatial domain. Finally, with respect to the development of face recognition abilities, the

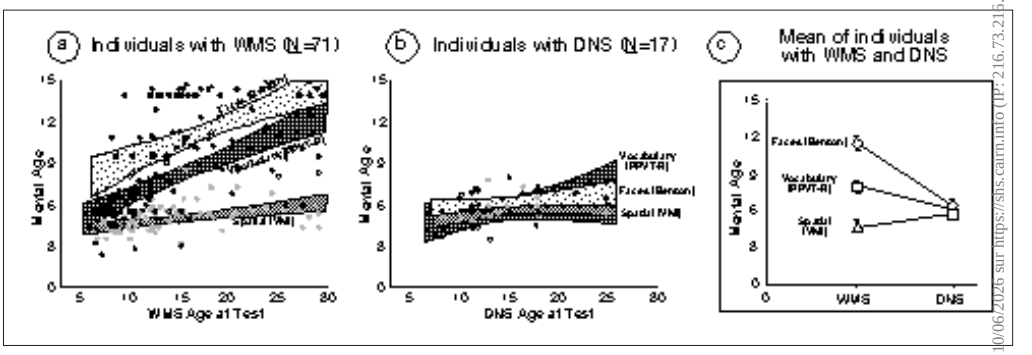

Fig. 2. — Distinct Trajectories in Cognitive Domains in Williams Syndrome but not in Down Syndrome

Developmental trajectories of contrasts between language, face and space processing in WMS are shown. *a)* Subjects of all ages with WMS show distinctly different trajectories in three domains: lexical knowledge, spatial cognition and face processing. On a standardized test of vocabulary, subjects with WMS start with low scores and then show a sharp increase with age. On a spatial task that involves copying geometric shapes, the performances of subjects with WMS are consistently below those of subjects with DNS, and plateau at an early age. On a task of face processing, subjects with WMS perform extremely well even at very young ages. *b)* Subjects with DNS showed essentially the same developmental trajectory across the three domains. *c)* In contrast to the DNS group, subjects with WMS show three distinctly different trajectories.

developmental trajectory in WMS was distinct from both those of the visual-spatial and of receptive vocabulary abilities. For face recognition, young children with WMS performed at a higher level, than expected for their age, and continued to perform well throughout development. Thus, in contrast to the other domains, face recognition is a relative strength for WMS throughout development. Contrasting with these three disparate developmental trajectories, the performance of the DNS group shows a coherent profile across domains with all three areas developing comparably.

4 / CONTROVERSIAL ISSUES IN LANGUAGE AND COGNITION

In the past 15 years increasing interest in WMS has spawned numerous studies focusing on language and its relation to other aspects of cognition; this issue has sparked much debate and the controversy remains unresolved (Clahsen & Almazan, 1998; Karmiloff-Smith, Grant, Berthoud, Davies, Howlin, & Udwin, 1997; Pinker, 1994, 1997; Stevens & Karmiloff-Smith, 1997; Bates, 1997; Levy, 1996; Volterra, Caselli, Capirci, Tonucci, & Vicari, 2003; Vicari, Caselli, Gagliardi, Tonucci, & Volterra, 2002). Broadly, some have presented WMS presented as a prime example of the

modularity of language in which language is independent, in significant respects, from other general cognitive systems. Others argue that since the cognitive functioning of adults with WMS is similar, in many respects, to young school aged children, such a level of cognitive functioning provides a sufficient substrate of cognitive abilities for the development of complex syntax. As such, WMS does not represent a dissociation between language and general cognitive functions. Levy (1996) argues that there may be "uniquely preserved accessing privileges for language which may enable individuals with WMS to reach levels of performance that they cannot reach through other modalities". Although these questions are unresolved, most scholars agree that language (particularly morphology and syntax) is a relative strength in WMS, and this profile makes WMS distinctive from other syndromes which include mental retardation (Volterra, Capirci, Pezzini, & Sabbadini, 1996; Karmiloff-Smith, 1998; Bellugi, Lichtenberger, Jones, Lai, & St-George, 2000).

The approach taken in this paper is to use language, particularly spoken narratives, as a means to better understand the relation of language to other cognitive domains: spatial cognition and affect. Using the disparate developmental trajectories and separability noted above as a starting point, we first concentrate on studies of language development in children with WMS and then use narratives as a context to explore later language and the intersection of language with these other cognitive domains: spatial cognition and then affective expression.

The WMS individuals who participated in these studies come from of a large group of more than 500 clinically- and genetically-diagnosed individuals with Williams syndrome, contrasted individuals with Down Syndrome (DNS) and typically developing individuals (TD) from three large studies based in California. Children with WMS were identified on the basis of established diagnostic criteria, including distinctive facies, a specific heart defect, mental retardation, and absence of one copy of the gene for elastin on chromosome 7 (see AAP, 2001 for diagnostic criteria). For those over age 12, the initial clinical diagnosis was confirmed by molecular genetic testing (fluorescence in situ hybridization, or FISH). In the following studies, we include a variety of groups for comparison: typically developing individuals who are matched with WMS individuals on chronological age (CA) or mental age (MA); and individuals with DNS, matched in age and Full Scale IQ to the WMS group. Because both Williams and Down groups are genetically-based disorders involving mental retardation, they have been chosen as comparison groups.

5 / EXPRESSIVE LANGUAGE IS A RELATIVE STRENGTH IN WILLIAMS SYNDROME

A / First words to early grammar

The global cognitive impairment typical of adolescents and adults with WMS contrasts dramatically to the ease and facility with which they use complex language, a characteristic not seen in other groups of mentally retarded individuals (Rossen, Klima, Bellugi, Bihrle, & Jones, 1996). Here we review some of the work in English (see Volterra, Capirci, Pezzini, & Sabbadini, 1996 ; Volterra, Caselli, Capirci, Tonucci, & Vicari, 2003; Vicari, 2002; and Pirchio, Caselli, & Volterra, this volume, for Italian, Karmiloff-Smith, Grant, Berthoud, Davies, Howlin, & Udwin, 1997; and Bernicot, Lacroix, & Reilly, this volume for French). In the development of language, WMS children appear to follow a distinct sequence. To capture the initial stages of language development, one study used a parent report, the MacArthur Communicative Development Inventory (CDI; Fenson, Dale, Reznick, Thal, Bates, Hartung, Pethick, & Reilly, 1993), with a sample of 54 children with WMS and 39 children with DNS (ranging in age from 12 to 76 months). They found no significant differences in the onset of first words between children with WMS and DNS; both groups showed significant delay compared to controls, and the children, especially the WMS group, showed a great deal of variability (Singer-Harris, Bellugi, Bates, Jones, & Rossen, 1997). Interestingly, in several studies of these

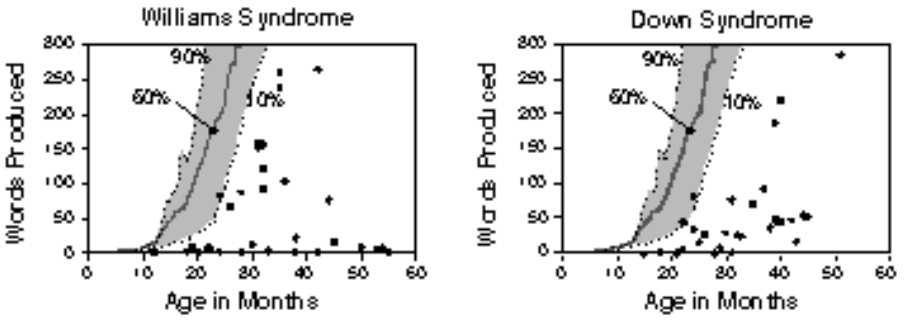

Fig. 3. — Development of First Words in Young Children with Williams and Down Syndrome

The total number of words produced by very young children (infants and toddlers) on the MacArthur Communicative Development Inventory (CDI) for children with WMS (left) and DNS (right) shows equally delayed onset of words in both groups. Shaded area represents trajectories of normal children in a large study using the CDI (Fenson *et al.,* 1993).

early stages, children with DNS had a larger repertoire of gestures than those with WMS. As Figure 3 shows, both WMS and DNS groups are equally delayed in the onset of first words.

With the emergence of grammar, the young children with WMS display marked and rapid development, however those with DNS do so only marginally; they tend to plateau quite early. The last section of the Macarthur CDI parent report includes the parents' writing the child's three longest sentences. In this instance, when matched for age and vocabulary, the sentences from the WMS children are significantly longer and more complex than those of the DNS group. Other researchers have found this linguistic advantage for WMS children over DNS from even earlier in development (Mervis & Robinson, 2000).

B / The development of grammar

Once children begin to produce sentences, linguistic competence is often assessed by morphological and syntactic performance. In this section we present data from a sentence repetition task as well as from narratives, a spontaneous discourse context, to explore these later stages of language development.

Looking at a wide age spectrum (age 3-16) using a sentence repetition task (Carrow Elicited Language Inventory, CELI), groups of children with WMS (N=68) and a small number of DNS (N=11), that were functioning at about the same level of general cognitive ability, were contrasted with typically developing controls (N=59). The number of morphosyntactic errors in the sentences were examined for each group. Typically developing children showed decreasing errors from ages three to eight years, and on average, achieved a perfect score between ages eight and nine. Similar to other aspects of language, the WMS group also progressed on this tasks with age, but slower than the TD group. Morphosyntactic errors decrease from ages 4-14; with near perfect scores by ages 14-16 years for the WMS group. However, for the DNS group, the slope was flatter as the number of errors decreased more slowly; they continued to continued to commit morphosyntactic errors at age 16.

To summarize so far, the language of children with WMS does not map directly onto that of adolescents with WMS. That is, language development in children with WMS is initially delayed; they show much individual variability; and their performance in childhood is below chronological controls, but well above that of DNS who have comparable intellectual impairments.

6 / THE DOMAIN OF SPATIAL COGNITION AND SPATIAL LANGUAGE

As clearly shown above, visuo-spatial cognitive tasks present a significant challenge for individuals with WMS. Whether copying an illustration or doing free drawing, when participants with WMS are asked to draw, the product typically has poor cohesion and lacks overall organization. Even into adulthood, individuals with WMS tend to plateau and remain basically at the level of typically developing five-year-olds (Beret, Lai, Hickok, Stiles, & Klima, 1997). It has often been noted that WMS individuals will verbally mediate their path through the drawings. This permits us to add the individual's own verbal labels to interpret the components of the drawing. Using narratives to better understand the relation between spatial language and spatial cognition, we look at spatial errors and descriptions of scenes that include spatial relations. Given WMS relative fluency with language, we wondered if their spatial deficits would manifest linguistically, and in fact pervade their language, or, if their language skills would allow them to circumvent or overcome their problems with space. If spatial language is dependent on spatial cognition per se, such that it maps directly onto the world, we would expect to see impaired performance in the use of spatial language. However, if language can develop semi-independently, then we should not see spatial errors above and beyond other types of morphological errors.

The present narrative study includes stories told by adolescents of the wordless picture book, *Frog, where are you?* (Mayer, 1969). As the repetition studies demonstrate, by adolescence, most individuals with WMS are making few errors. Therefore, with this general level of morphosyntactic proficiency, if spatial cognition influences language, we would expect errors to be primarily spatial in nature. To address these questions, we looked at spatial language in 27 adolescents with Williams Syndrome (ages 11-15; mean IQ=55); 14 adolescents with Down Syndrome (matched for age and IQ) and 24 Typically Developing adolescents (matched for age). Spatial language was considered from two different perspectives: spoken narratives (focusing on types of morphological errors, specifically spatial prepositional errors); and specific scene descriptions (which critically include spatial information).

When we look at morphological errors in the narratives, overall, as expected, the WMS group make few errors in this age group, however, the older group (13-15) does better than the younger group (11-13) reflecting continued development. As a whole, the WMS group does not significantly differ from the typically developing controls; the DNS group however, makes significantly more errors than either the WMS or the TD control group. In this discourse context, and in this age group, the WMS group has

attained proficiency in English morphosyntax, however, we found a variety of problems encoding spatial relationships.

To look more closely at how the WMS group mapped language onto spatial relations, we chose four pictures from the Frog story in which the spatial information was especially pertinent, and we looked at how these pictures were described by the WMS adolescents, by typically developing adolescents, and also by an additional group of 20 adults. Overall, the WMS group was less likely to explicitly encode the static spatial relations typically included by the control groups. For example, in the picture below, descriptions from 92% TD adolescents mentioned the boy looking into a hole in the tree and 85% mentioned the beehive falling down. In contrast, for the WMS group, only 25% mention looking in the hole and only 25% mention that the beehive fell or was knocked down. Moreover, in their descriptions, 30% of the WMS group (as compared to zero in the control group) made some kind of error in their description.

Overall, despite their apparent proficiency with morphology and syntax, we found that the WMS group often omitted pertinent spatial information, and made more errors than overall morphological performance would predict when spatial descriptions were called for, thus reflecting in language their non-linguistic spatial deficits. The data do suggest that spatial language is in fact dependent on an intact system of spatial cognition and that the two systems very much work in concert. Nonetheless, in everyday conversation, these spatial errors are rather subtle, suggesting that the linguistic skills of the WMS group do allow them to talk about space more competently than their visuo-spatial skills might predict.

7 / THE INTERSECTION OF LANGUAGE AND AFFECT

Children, adolescents and adults with WMS are extremely friendly, attracted to strangers and frequently display a type of hypersociability. Even as infants, in social situations, they show more positive and less negative behavior than typically developing infants and toddlers (Jones, Bellugi, Lai, Chiles, Reilly, Lincoln, & Adolphs, 2000). Many individuals with WMS display a strong impulse towards interpersonal social contact and enhanced affective expressivity, although their social behavior may not always be appropriate (Einfeld, Tonge, & Florio, 1997). Here we look at how children and adolescents with WMS use their linguistic skills to engage others socially. Again we turn to narratives of *Frog, where are you?* (Mayer, 1969) to investigate the intersection of language and social behavior in individuals with WMS. The most obvious characteristic differentiating narratives from subjects with WMS from those with DNS and age-matched or mental age matched controls is in their abundant use of narrative enrichment or eva-

luative devices during this story-telling task (Reilly, Klima, & Bellugi, 1990; Losh, Bellugi, Reilly, & Anderson, 2001; Reilly, Bernicot, Vicari, Lacroix, & Bellugi, in press). These enrichment devices provide the evaluation of the narrator, that is, his or her perspective on the information explicitly presented in the pictures themselves. Using language and prosody, the emotions, motivations and thoughts of the story characters are expressed, as well as those of the "narrator"–in this case the particular WMS individual. Individuals with WMS use an abundance of affective expression both prosodically and lexically; they appear to be able to manipulate affective linguistic devices for the purposes of story-telling. Affective prosody was measured by noting how frequently paralinguistic affective expression was used, including changes in pitch, volume and vocalic lengthening (Reilly, 1992). Affectivity in lexical devices was measured by the relative frequency of evaluative devices (cf. Labov & Waletsky, 1967; Bamberg & Reilly, 1996), such as exclamations, intensifiers, sound effects, and emotion words, used to engage the audience. This heightened profile of linguistically encoded affect is also found in Italian and French speaking children and adolescents with WMS (see Bernicot *et al,* this volume, and Reilly, Bernicot Vicari, Lacroix, & Bellugi, in press). This profile was found to be significantly different from the stories of subjects with DNS, as well as from those of typically developing children or other clinical groups (Reilly, Bates, & Marchman, 1998; Reilly *et al.,* in press).

As is often noted, a primary characteristic of individuals with WMS appears to be an impulse for affective expression and social contact. Their abundant use of affective language in narratives reflects the confluence of strengths of the WMS profile, affective expressivity, sociability, and language.

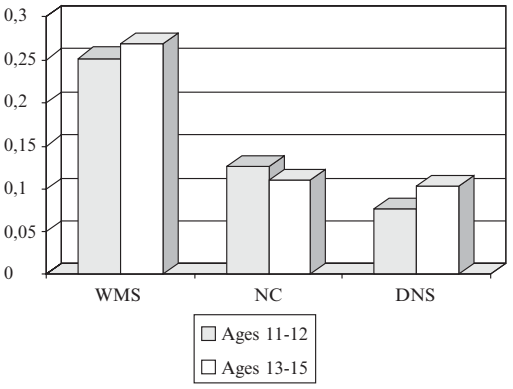

Fig. 4. — Evaluation in Narratives from Children with Williams Syndrome: The Confluence of Affect and Language

8 / CONCLUSION

Noting the fractionated behavioral profile in individuals with Williams Syndrome, in this paper, we have looked at language development from first words to grammar in children with Williams Syndrome and then how particular aspects of cognition intersect with language, an area of strength. We examined how spatial cognition, an area of pronounced weakness in those with WMS intersected with language and found that impaired spatial cognition does indeed influence linguistic performance; linguistic proficiency does not fully conceal or mask spatial difficulties. We also investigated linguistically encoded affective expression, as a means of capturing the intersection of language and sociability in WMS. The findings suggest that as soon as language becomes available, individuals with WMS use it effectively and characteristically for social purposes. Thus, in demonstrating how language interacts with other aspects of the social-cognitive profile of individuals with WMS, we have found that expressive language, a clear strength, does indeed reflect other aspects of their cognitive profile. These data should provide clues to the long-standing theoretical issues in the functional organization of language and non-linguistic cognition.

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Remerciements.—The research described here was supported by Grants from the National Institutes of Health (NICHD P01 HD 33113 to UB, P50 NS22343 to Ursula Bellugi and Judy Reilly, as well as a grant from the James S. McDonnell Foundation to UB). The authors are especially grateful to the subjects and their families for their spirited participation in these studies.