Narratives in French and American Children with Williams Syndrome

Judy Reilly[1, 2], Agnes LaCroix[2], Josée Poirier[1], Josie Bernicot[2],
Ursula Bellugi[3], & Edward Klima[3]

San Diego State University[1], Université of Poitiers[2],
Salk Institute for Biological Studies[3]

To appear in a Special Issue of L'HOMME ET LANGAGE,
in memory of Elizabeth A. Bates

Introduction

Williams Syndrome (WS) is a rare genetically based disorder that occurs in 1/20,000 live births. Its characteristic medical, psychological, neuropsychological and neuroanatomical profile and stems from a hemizygous deletion of about 20 genes and transcription factors on chromosome 7 including Elastin, Lim1kinase, Syntaxin1a, GTF2I and GTF2IRD1 among others (Ewart et al., 1993; Frangiskasis et al., 1993; Botta et al., 1999; Korenberg, et al., 2000, Korenberg, et al, 2003 Hirota, et al, 2003). One of the most striking aspects of individuals with WS, and one that has intrigued developmental cognitive neuroscience, is the distinctive cognitive profile they present: Individuals with WS are moderately retarded, with IQs typically in the 50–70 range; their impairments in visuo-spatial, planning and numerical/arithmetic abilities stand in marked contrast to their apparent fluency with language, extreme sociability and positive affect (Atkinson, et al., 2001; Bellugi, et al., 2001; Gianotti & Vicari, 1999;Vicari, Bellucci, Carlesimo, 2001; Losh et al 2001, Jones, et al., 2000; Reilly et al 2004; Vicari, et al., 2002). Relying on these two communicative systems, language and affect, individuals with WS are known to charm, flatter and socialize with any available adult.

From the very first encounters with children with WS, medical professionals and researchers alike have been struck with their verbal facility. Von Arnim and Engel (1964) in one of the earliest papers on record to describe individuals with Williams wrote, "The salient features of the psychological structure are an unusual command of language combined with an unexpectedly polite, open and gentle manner," (P.367). As a result, over the past 20 years, much ink and energy has gone into discussions regarding their apparent linguistic proficiency. Some have taken WS apparent facility with language as example of modularity (Clahsen & Almazon, 1998; Levy, 2004; Pinker, 1997) whereas as others have provided extensive data disconfirming this claim, (Bates, 2004; Bates, Tager-Flusberg, Vicari, Volterra, 2001; Karmiloff-Smith, Brown, Grice et al, 2003; Karmiiloff-Smith, Grant, Berthoud et al, 1997). Still other researchers have shown that language itself for the WS group includes interesting strengths and weaknesses (Bellugi, Lichtenberger et al, 2001). Whereas most researchers agree that productive language of WS adolescents and adults is an undisputable strength in their profile, when we look at younger children with WS, the emergence of language is initially delayed (Singer-Harris et al., 1997; Morris & Mervis, 2000); initial milestones may reflect an atypical sequence of development (Mervis & Klein Tasman, 2000), acquisition is slow (Bellugi, Lichtenberger et al, 2001; Reilly et al 2004); that WS children may rely more on phonology rather than semantics (Mervis, Morris, Bertrand, Robinson, 1999; Vicari, Carlesimo et al, 1996); that WS children are delayed with respect to morphology and syntax Bellugi, et al, 2000); that variability across subjects is high (Losh, Bellugi, Reilly, 2000); and finally, that by adolescence, most WS are very good talkers The bulk of this work has been conducted in English and Italian; a few studies have included French speaking children with WS (Karmiloff-Smith et al 1997, Bernicot et al., 2003; Lacroix et al., 2004; Reilly et al., 2005) and several recent contributions come from Hungarian (Pleh, Lukacs & Racsmany, 2003) and Hebrew (Levy, 2004). The results with respect to morphosyntactic proficiency in these languages rich in inflectional morphology have been conflicting. Specifically for French, Karmiloff-Smith and colleagues (1997) have found that WS had difficulty with gender, whereas Monnery and colleagues found that French speaking WS performed well on tests of gender, but had difficulty in lexical retrieval (2002). Studies of Hebrew and Spanish also found that gender was not a problem for the WS group (Levy & Hermon 2003). Given these conflicting conclusions, it has been difficult to tease apart those aspects of performance that are specific to a particular language from those which are characteristic of Williams Syndrome. In this paper we will look at narrative data from French speaking children and adolescents with WS with an eye to broadly comparing these results to

their English speaking counterparts. We will look not only at structural, i.e., grammatical, proficiency, that is mastery of morpho-syntax, but also how the *use* of language by children with WS might inform our understanding of their phenotype, using French, a language richer in inflectional morphology than English.

Because narratives are common in everyday social interaction, and even children as young as 3 years have an idea of 'what a story is,' (Appleby, 1978) narratives provide an excellent context for eliciting expressive language data. Narratives permit us to look not only at the child's proficiency with particular linguistic structures, but also with how she uses those structures to construct a coherent and cohesive story. According to Labov and Waletzky (1967), a *good* story entails not only a plot or the *referential* information (information about the plot and characters), but also its meaning, or significance, according to the narrator, that is, the *evaluative* aspect of a narrative. Given their apparent linguistic strengths and sociability, narratives represent an appropriate context to investigate language in individuals with WS. Although similarities have long been noted in the acquisition of different languages in typically developing (TD) children, clear language specific aspects have also been recognized (Slobin 1985, 1992, 1997). Additionally, within discourse, we know that the rhetorical choices, that is, the conventions for using particular structures in discourse also differs across cultures and languages (Berman & Verhoeven, 2002; Jisa et al, 2002). Thus comparing profiles of language development of French and English speaking individuals with WS within a discourse context will begin to differentiate those aspects of linguistic behavior that are common to WS from those which are more closely tied to the particular language that the children are learning and to the culture in which they are living.

Our past studies of narratives with English speaking children with WS have yielded interesting findings. In the school aged group (Losh et al, 2001; Reilly et al 2004), the WS children tell stories of comparable length to the TD children, however they make many more morpho-syntactic errors than their chronologically matched controls. In fact, quantitatively, the WS group make as many morphological errors as age matched children with Language Impairment (LI) (Reilly et al, 2004). With respect to recruiting complex syntax, they are delayed, although they show the typical developmental pattern of initially relying on coordination and then later using more subordination (Reilly et al 1998, Losh et al., 2001). And lastly, comparing the structure of their stories to those of both the LI group and the TD group, the WS stories are somewhat impoverished, that is, they include fewer components of the story, even though they were looking at the pictures as they told the story. However, if we look at evaluative devices, that is, those indices reflecting the narrator's perspective, the WS group far outstrips both the typically developing children and the LI group. Interestingly, in the adolescent group (Reilly et al 1991), we find few errors, a richer use of syntax and again, an extensive use of evaluation. With the American children as background, we now turn to a group of French speaking children and adolescents with Williams Syndrome.

Methods

Participants: A total of 36 French speaking children and adolescents participated in the study**:** Twelve (12) children and adolescents with WS, diagnosed by a geneticist, and recruited with the help of the regional Associations du Syndrome de Williams in France. The participants in the Williams Syndrome group ranged in age from 6-18 (mean age=12.3), and their IQs ranged from 41-74, with a mean of 65. Two groups of typically developing children from local schools in Poitiers were used as controls: 12 typically developing children matched for chronological age (CA), gender and socio-economic status; and 12 typically developing children matched for mental age (MA), gender and socio-economic status.

Procedure: Similar to others who have used this task (e.g., Berman & Slobin, 1994, Jisa & Kern, 1998) our narrative task consisted of telling a story from a wordless picture book *Frog, Where Are You?* (Mayer, 1969). This 24-page "quest" story is about a boy, his dog, and a frog, and the story begins with the boy and the dog in the bedroom looking at a frog in a jar. In the morning, they wake up to find that the frog has disappeared; the remainder of the story consists of their search for the frog. During their search, the boy

and the dog encounter various obstacles, but in the end, they find the frog with a mate and baby frogs. Children first look through the book and then are asked to tell the story as the pictures appear. The children's narratives were both audiotaped and videotaped; stories were transcribed using the CHAT format from the CHILDES system (MacWhinney, 2000; Sokolov & Snow, 19940). Utterance boundaries were determined by intonation contours as well as pause length.

Coding. Our coding scheme, which was originally developed by Reilly, Bates & Marchman (1998) has now been used to understand narrative discourse in variety of populations (Losh et al 2001, Reilly et al, 2004; Losh and Capps, 2003) Here we will focus on morpho-syntax and evaluation.

1. Overall Story Length — Number of Propositions. Children's stories vary in length, thus to neutralize these differences, we have created proportions to compare performance (e.g., the frequency of morphological errors). Thus, stories were first coded for length as measured by number of propositions. In this analysis, a proposition is defined as a verb and its arguments, with a proposition corresponding roughly to a distinct, single event. Each clause in a complex sentence was considered to represent one event, thus, one proposition. For example, the utterance, "Le petit garçon cherche la grenouille parcequ' il voulait la trouver," counted as two propositions, as would "Le petit garçon cherche la grenouille; il voulait la trouver." In contrast, "Il voulait sortir" was counted as one proposition. In sum, as a mechanism to control for varying story lengths, the number of propositions in a story was used as a denominator for more detailed explorations of linguistic and evaluative performance, as presented below.

2. Coding Morpho-Syntax. All errors of commission or omission were tallied. Subcategories of morphological errors included the following (examples):

1)Errors in agreement Gender: " **le** fenêtre est ouverte" la grenouille est **beau**;
Number: " le chien et le garçon **il** est heureux " ;

2) Verb auxiliaries "le garçon **a** monté sur le rocher ";

3) Verb tense "une petite fille qui **dormant**";

4) Prepositional errors " il a mis le bocal **avec** la tête"»;

5) Missing obligatory constituents "Le petit garçon qui cherche " ;

6) Addition of a superfluous word "l'enfant dit au **le** chien"

Results and Discussion

Morpho-syntax

Initially we compared the length of the children's stories and found that the stories of the WS children were shorter than their CA matched ($p<.05$) controls but did not differ significantly from their MA matched controls. Using their story lengths, the proportion of morphosyntactic errors (over the number of propositions) was computed for all children. as is seen in Figure 1. The children with WS perform similarly to both their chronological-age matched and mental-age matched controls. However, the two control groups differ from one another: The MA controls make significantly more errors than the CA controls ($F(1,33) = 4.85$, $p= 0.0346$). In sum, although the performance of the WS group does not differ significantly from the typically-developing children, they have not yet reached the proficiency level of the CA children.

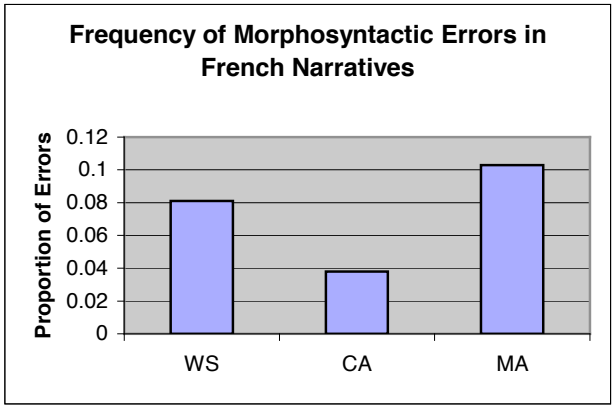

Figure 1. Morphosyntactic Errors in French Children and Adolescents

French speaking children wihWS make few errors; they fall between their chronological and mental aged matched controls. Comparatively, English-speaking WS children make significantly more morpho-syntactic errors than their chronological age matched and their mental age matched controls. Looking at Figure 2, it is clear that there is little variability in the French groups and both the WS and TD groups are almost at floor. In contrast, for English, the WS group shows massive variability well into adolescence whereas the proportion of errors for the TD group diminishes significantly with age.

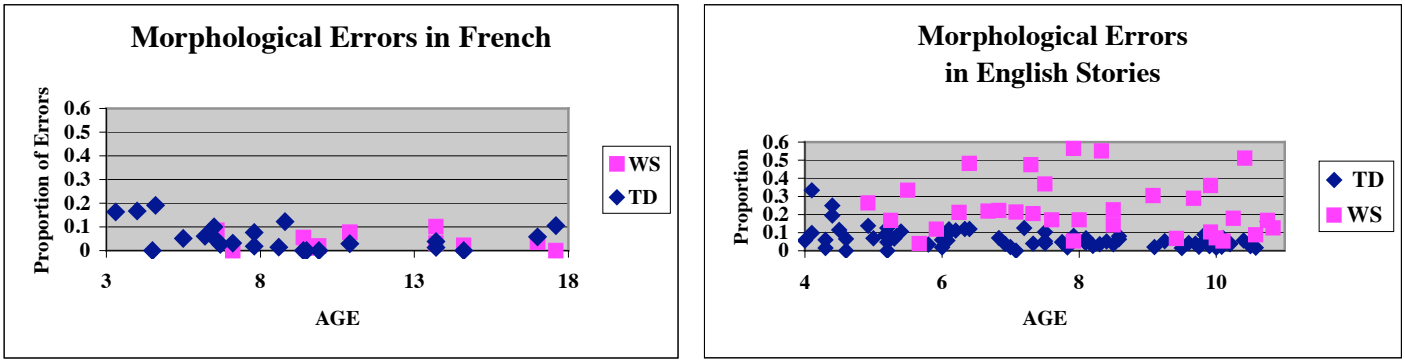

Figure 2: Variability in morphosyntactic performance (proportion of errors) for children with WS in English as compared to French. English data is from 30 children with WS and 73 TD children.

Why are the French children with WS doing do much better than their English counterparts? One possible factor is that in spite of the increased morphology of French, there is much homophony. Stories include predominantly third person pronouns and in French the singular and the plural (il/ils) are homophonous as are the verbs in the 2nd group: consider il cherche and ils cherchent. To understand these differing levels of linguistic proficiency more in depth systematic studies will be required. Regardless, these cross-linguistic data from the same task demonstrate that the language acquisition profiles for WS children learning English and French are quite different, and that mastery of morphology and syntax are not defining features of the Williams group.

Language and Space

Given the very few errors of the French speaking children and adolescents with WS, it was striking how many of the errors appeared to be with prepositions. In light of WS frequently noted visuospatial impairments, the use of prepositions may represent an area in which other cognitive abilities influence language performance. Looking across the groups, more than half the WS (7/12) made such errors whereas for the TD groups, only 25%-33% made at least one or more prepositional error. This discrepancy between WS and TD children with respect to errorful prepositions merited closer investigation.

Going back to the texts, we coded all the prepositions appearing in the narratives and divided the French prepositions into two categories: spatial and non-spatial according to *Grammaire du français contemporain* (Larousse). Prepositions such as *au dessus* and *sur* were classified as 'spatial', whereas prepositions such as *avant, après*, and *par* were classified as 'non-spatial'. As a group, children with WS used a total of 198 prepositions, 79 (39.9%) of which were classified as 'spatial' and 119 (60.1%), as 'non-spatial'. Of these 198, there were 13 errors, and these included both semantic and morphosyntactic errors. Although small, the number of preposition errors hid a clear pattern: 100% of these errors were with spatial prepositions (see Figure 3 below). Hence, the nature of the prepositions had a significant impact on the children's ability to use them correctly ($t(11) = 3.463$, $p = 0.005$). The WS group is the only one

affected by the nature of the prepositions. Specifically, no significant difference between spatial and non-spatial prepositions was found for either control group.

Figure 3. Spatial Errors whn looking at picture from *Frog, Where are you (Mayer, 1969) :*
Pour pas que l'animal qui sort ***dans le*** *trou l'entende.*
Y'a quelqu'un là-dedans ***sous*** *le trou?*

If problems with such prepositions reflected spatial deficits, we would expect to see a similar profile with other linguistic markers if they too described spatial relations. Another type of spatial markers includes certain adverbs and adverbial locutions. The WS group used a total of 13 adverbs and 8 adverbial locutions that are classified by the *Grammaire du français contemporain* (Larousse) as spatial (e.g.: *ici, dedans, dehors)*. Errors were found with 42.9% (9/ 21) of these elements. Overall, if we look across all the linguistic elements that encode spatial relations, for the WS group, the ratio of erroneous to correct uses of spatial elements (prepositions, adverbs and adverbial locutions) was 22:77. That is, close to 1 spatial element out of 4 was incorrectly used. In sum, the WS struggle with markers of spatial relations, significantly more than do TD children.

An additional cue that the visuospatial impairment in WS children permeates their language use is a tendency to avoid describing the spatial relations altogether. In the storybook are two pictures (page 12-13) that depict events involving spatial relations (a beehive falling down (1) and a boy in a tree looking in a hole (2)). We tallied the topic(s) the children chose to talk about. In the WS group, none of the five younger children (i.e. less than 10 year old) mentioned any of the events requiring the description of a spatial relation. The seven older children (more than 10 year old) mentioned at least one, and only two of the WS group mentioned both events. In contrast, only five of all CA controls did not mention both events, and no child omitted the two altogether. Thus, it seems that WS children (especially the younger ones) choose to avoid talking about spatial relations.

In sum, although French-speaking children with WS appear to show morpho-syntactic proficiency comparable to the TD group, they still struggle with certain elements that TD do not. Specifically, WS children seem to struggle with spatial markers, be they prepositions, adverbs or adverbial locutions, so much so that they tend to avoid discussing topics relating to spatial relations. When they do describe these relations, they make numerous errors using the relevant markers. Note that the apparent difficulty in using spatial markers is not due to the establishment of any kind of relation between two elements: no errors were found with prepositions, adverbs and adverbial locutions that referred to an agent (*par*), indicated a purpose (*pour*), conjoined two elements (*avec)* or

described a temporal relation (*après*). Thus, in spite of their relative linguistic proficiency, it does appear that their use of language reflects other aspects of their cognitive profile, in this case, a weakness in visuospatial processing.

Interestingly, we have recently found that in the English narratives of adolescents with WS, when their language is relatively good, errors in the use of spatial prepositional constitute a relatively high proportion of the total errors (Reilly, et al, 2001; Lichtenberger et al, 2002). Such findings in naturalistic speech are complemented by the experimental studies of Landau who has also found deficits in spatial language for English speaking individuals with WS (Landau & Zukowski, 2003; Lakusta & Landau, 2004). Overall then, it appears that the visuospatial impairments evident in those with WS permeate their use of language, and this occurs across languages and across cultures. As such, how language is used can serve as an index characterizing their neuropsychological profile. Finally, we turn to another aspect of language use, evaluation.

Language and Sociability: Evaluative Language

From our first study of narratives in English speaking adolescents with WS, we noticed that their story-telling was filled with affective prosody and evaluative language (Reilly et al, 1991). In a large scaled study with school aged children, the pattern was similar (Losh et al 2001). For the French stories, we used a similar coding system and here we focus on social evaluative devices: Using phrases or exclamations to capture addressee attention, e.g. sound effects, character speech, as in "Tiens, le garcon il tombe dans l'eau!"

Looking at the French group, it is clear that enhanced use of social evaluative devices is also a characteristic of French children and adolescents with WS. As can be seen in Figure 4, the WS group uses twice the proportion of social evaluation as their MA matched controls and ten times the amount of their CA matched controls.

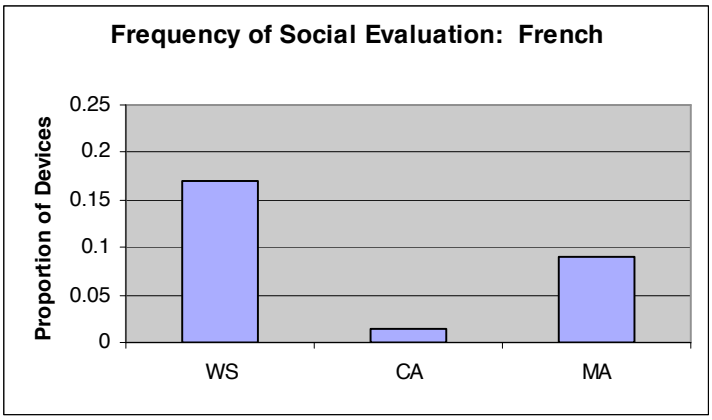

Figure 4: Frequency of Social Evaluative Devices in Narratives from French speaking children with Williams Syndrome and their controls, both chronological and mental age matched.

Let us return to the question "why are people who first meet an individual with WS struck by their 'good' language?" It is our conjecture that it is how people with WS use language rather than the structural aspects that are most striking. As we saw above from the spatial errors, the visuo-spatial impairment that characterizes WS permeates their language: they appear to avoid explicitly indicating important events in the story when spatial information was a core element, and when they did use prepositions to encode spatial relations, almost a third were errorful. Thus, similar to English speaking children with WS, the French group also reflects this profile. Another distinctive characteristic of those

with WS is their exuberant sociability and affiliative drive, and just as their spatial impairment is reflected in language use, language can also serve as means to socially connect. In both the French stories and the American stories, the WS groups use far more social evaluative language than we see in any typically developing groups. Thus, again, the striking sociability of individuals with Williams Syndrome is expressed by how they manipulate and exploit the language available to them. Although their excessive sociability and strong affiliative drive characterize the WS groups in both the French and American studies, it is also clear that the individual cultures also play a role. As can be seen in Figure 5, in the French groups, both WS and TD use less evaluation than their American counterparts, suggesting that the cultural rules for expression of emotion and evaluation are different and that even with a genetically based syndrome, children are shaped by these cultural conventions.

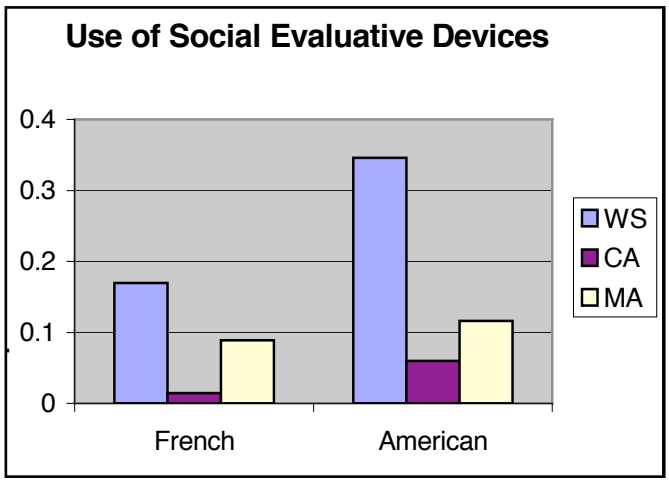

Figure 5: Using Social Evaluative language in narratives from French and American children with Williams Syndrome and their chronological and mental age matched controls.

Conclusions

In this paper, we have looked at narratives from French and American children and adolescents with Williams Syndrome. By comparing behaviors across languages and cultures, we were able to better delineate the phenotype of WS, separating what might be language and culture specific from what might in fact characterize this rare genetically based syndrome. With respect to structural aspects of language, specifically morphosyntactic proficiency in this quasi-naturalistic context, the French and English speaking groups show very different profiles. Those children with WS who are acquiring French perform similarly to controls whereas those learning English make numerous errors and are significantly delayed. However, if we focus on how individuals with WS *use* language, we find that spatial relations indeed appear to pose specific problems, which are reflected in the larger proportion of errors in spatial than non-spatial prepositions in this population. And significantly, both the English and French groups of children with WS are particularly adept at using language to socially connect with their addressees. How language is used then serves as an index of the phenotype of Williams Syndrome, underscoring their cognitive limitations as well as their great propensity for social interactions, involving heightened affective language. These results across languages and cultures indeed provide evidence for a characteristic of WS behavior that is beyond the borders of typical normal behavior and mark an aspect of a "Williams phenotype": an irrepressible affiliative drive, or hypersociability. Taken together with other behavioral results of social behavior in studies of Williams individuals (Doyle, et al, 2004; Hirota, et al, 2003; Doyle et al, 2004), these findings also lend support to the hypothesis of a specific genetic basis for overly social behavior in Williams syndrome.

References

Atkinson J, Shirley L, Braddick O, Nokes L, Mason A and Braddick F. Visual and visuospatial development in young children with Williams syndrome. Developmental Medicine and Child Neurology 2001; 43: 330-7.

Appleby, A. (1978). The child's concept of story. Chicago: University of Chicago Press.

Bates EA. Explaining and interpreting deficits in language development across clinical groups: where do we go from here? Brain Lang. 2004 Feb;88(2):248-53.

Bates E, Tager-Flusberg H, Vicari S, Volterra V.Debate over language's link with intelligence.Nature. 2001 Oct 11;413(6856):565-6.

Bellugi U, Korenberg, J R, & Klima E S. (2001) Williams syndrome: An exploration of neurocognitive and genetic features. In M. Posner (Ed.) Journal of Clinical Neurosciences Research, Special Issue 1: 217-229.

Bellugi, U., Litchenberger, L., Jones, W., Lai, Z., & St. George, M., (2001). The neurocognitive characterization of Williams Syndrome: A complex pattern of strengths and weaknesses. In U. Bellugi & M. St. George (Eds.), Journey from cognition to brain to gene: Perspectives from Williams syndrome (pp. 1-42). Cambridge, MA: MIT Press.

Bellugi U, Lichtenberger L, Mills D, Galaburda A, Korenberg JR.Bridging cognition, the brain and molecular genetics: evidence from Williams syndrome.Trends Neurosci. 1999 May;22(5):197-207. Review.

Bernicot, J. La Croix, A. & Reilly, J. (2003). La narration chez les enfants atteints du syndrome de Williams: aspects structuraux et pragmatiques. In J. Reilly & Bernicot, (Eds.) Le développement du langage chez les enfants atypiques. *Numero Special d'Enfance*, 3, pp.265-282.

Berman, R. & Slobin, D. (1994). Relating events in narrative: A cross-linguistic developmental study. Hillsdale, NJ: Lawrence Erlbaum.

Berman, Ruth A., Verhoeven, Ludo, 2002. Crosslinguistic perspectives on developing text production abilities in speech and writing. Written Language and Literacy 5, 1–44.

Botta A, Sangiuolo F, Calza L, Giardino L, Potenza S, Novelli G, & Dallapiccola B. Expression analysis and protein localization of the human HPC- 1/syntaxin 1A, a gene deleted in Williams syndrome. Genomics 1999; 15: 525-8.

Capirci, O., Sabbadini, L., & Volterra, V. (1996). Language development in Williams syndrome: A case study. Cognitive Neuropsychology.

Chevalier, J-C; Blanche-Benveniste, C.; Arrivé, M; Peytard, J., (1964). Grammaire du français contemporain (Larousse) Librairie Larousse : Paris.

Clahsen H, Almazan M. Syntax and morphology in Williams syndrome.Cognition. 1998 Sep;68(3):167-98.

Doyle, T.F., Bellugi, U., Korenberg, J.R. & Graham, J. (2004). "Everybody in the world is my friend": Hypersociability in young children with Williams syndrome. American Journal of Medical Genetics, 124A, 263-273.

Doyle, T. F., Bellugi, U., Reiss, A. L., Galaburda, A. M, Mills, D. L., & Korenberg, J. R. (2004). Genes, neural systems, and cognition: Social behavior of children with Williams syndrome: Observing genes at play? [Abstract/Poster]. Society for Neuroscience 34th Annual Meeting Abstracts, 666.11.

Ewart A K, Morris C A, Atkinson D, Jin W, Sternes K, Spallone P, Stock A D, Leppert M, Keating M T. Hemizygosity at the elastin locus in a developmental disorder, Williams syndrome. Nature Genetics 1993; 5: 11-6.

Frangiskakis J M, Ewart A K, Morris C A, Mervis C B, Bertrand J, Robinson B F, Klein B P, Ensing G J, Everett L A, Green E D. LIM-Kinase-1 hemizygosity implicated in impaired visuospatial constructive cognition. Cell 1996; 86: 59-69.

Giannotti A. & Vicari S. (1999). La Sindrome di Williams: Aspetti clinici e riabilitativi. Franco Angeli Ed., Milano.

Hirota, H., Matsuoka, R., Chen, X.N., Salandanan, L.S., Lincoln, A., Rose, F., Sunahara, M., Osawa, M., Bellugi, U., & Korenberg, J.R. (2003). Williams syndrome deficits in visual spatial processing linked to GTF2IRD1 and GTF2I on Chromosome 7q11.23. Genetics in Medicine, 5(4), 311-321.

Jisa, H & Kern, S. (1998). Relative Clauses in French Children's Narrative Texts, Journal of Child Language, 25, 3, Oct, 623-652

Jisa, H., Reilly, J.S., Verhoeven, L.,Baruch, E., & Rosado, E. (2002). Passive Voice construction in written texts: A cross-linguistic study. Written Language and Literacy, 2, 5:163-182.

Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). Hypersociability in Williams syndrome: An addition to the behavioral phenotype. In U. Bellugi and M. St. George, Linking Cognitive Neuroscience and Molecular Genetics. *Special Issue: Journal of Cognitive Neuroscience*, 12(1), 30-46.

Karmiloff-Smith A, Grant J, Berthoud I, Davies M, Howlin P, Udwin O. Language and Williams syndrome: how intact is "intact"? Child Dev. 1997 Apr;68(2):246-62.

Karmiloff-Smith A, Thomas M. What can developmental disorders tell us about the neurocomputational constraints that shape development? The case of Williams syndrome. Dev Psychopathol. 2003 Fall;15(4):969-90.

Karmiloff-Smith A, Brown JH, Grice S, Paterson S. Dethroning the myth: cognitive dissociations and innate modularity in Williams syndrome. Dev Neuropsychol. 2003;23(1-2):227-42.

Korenberg, J.R., Bellugi**, U.,** Salandanan, L.S., Mills, D.L., & Reiss, A.L. (2003). Williams syndrome : A neurogenetic model of human behavior. In *Encyclopedia of the Human Genome*. London: England. The Nature Publishing Group.

Korenberg, J., Chen, X-N., Hirota, H., Lai, Z., Bellugi, U., Burian, D., Roe, B., & Matsuoka, R. (2000). Genome structure and cognitive map of Williams syndrome. In U. Bellugi and M. StGeorge, Linking Cognitive Neuroscience and Molecular Genetics. *Journal of Cognitive Neuroscience,* 12(1), 89-107.

Kreiter, J., Bellugi, U., Lichtenberger, L., Klima, E.S., Reilly, J.S., & Kikuchi, D. (2002, November). Gregarious language in narratives by adolescents with Williams syndrome. Poster session at the American Speech-Language Hearing Association Convention, New Orelans, LA.

Labov, W. (1984). Intensity. In D. Schiffrin (Ed.) Meaning, form and use in context: Linguistic applications. Georgetown University Round Table on Languages and Linguistics, pp. 43-70. Washington, D.C.: Georgetown University Press.

Labov, W., & Waletzky, J. (1967). Narrative analysis: Oral versions of personal experience. In J. Helm (Ed.), Essays on the verbal and visual arts (pp. 251-263); reprinted in 1997**.**

Lacroix, A, Bernicot, J., & Reilly, J. (2004). Conversations et etats mentaux chez les enfants atteints du Synbdrome de Williams. In M. Guidetti & V. Laval (Eds.) Developmental Pragmatics, *Numero Special de Psychologie française,*161-176.

Landau, B. & Zukowski, A. (2003). Objects, mothions and paths: spatial lanague in children with Williams Syndrome. Developmental Neuropsychology, 23, 105-137.

Lakusta L, Landau B. (2005).Starting at the end: the importance of goals in spatial language. Cognition.96(1):1-33.

Levy Y. (2004). A longitudinal study of language development in two children with Williams syndrome.J Child Lang. May;31(2):287-310.

Levy Y, Bechar T.Cognitive, lexical and morpho-syntactic profiles of Israeli children with Williams syndrome.Cortex. 2003 Apr;39(2):255-71.

Lichtenberger, E.O., Bellugi, U., Kikuchi, D.K., & Klima, E.S. (2002, July). The intersection of language and space: representing spatial prepositions. Poster presentation Williams Syndrome Association Professional Conference, Long Beach, CA.

Losh, M., Bellugi, U. Reilly, J and D. Anderson. (2001). The integrity and independence of evaluation in narratives: Evidence from children with Williams Syndrome. Narrative Inquiry. Vol 10(2), 265-290.

Losh M, & Capps L. (2003).Narrative ability in high-functioning children with autism or Asperger's syndrome. J Autism Dev Disord. 2003 Jun;33(3):239-51.

MacWhinney, B., (2000). Tools for Analyzing Talk, Transcription Format and Programs., The CHILDES Project. Mahway, NJ: Lawrence Erlbaum.

Mayer, M. (1969). Frog, where are you? New York: Dial Books for Young Readers

Mervis CB.Williams syndrome: 15 years of psychological research.Dev Neuropsychol. 2003;23(1-2):1-12.

Mervis CB, Klein-Tasman BP.Williams syndrome: cognition, personality, and adaptive behavior. Ment Retard Dev Disabil Res Rev. 2000;6(2):148-58. Review.

Mervis, CB, Morris, CA, Bertrand, J &Robinson, BF. (1999). Williams Syndrome: Findings from an intergrated program of research. In H. Tager-Flusberg (eD). Neurodevelopmental disorders: Contributions to a new framework from the cognitive neurosciences. Cambridge, MA: MIT Press.

Monnery, S. Seigneuric, A. Zagar,D., Robichon, F. (2002). A linguistic dissociation in Williams syndrome: Good at gender agreement but poor at lexical retrieval. Reading and Writing, 15, 589-612.

Morris CA, Mervis CB.Williams syndrome and related disorders.Annu Rev Genomics Hum Genet. 2000;1:461-84..

Pinker, S. (1994). The Language Instinct. New york: William Morrow Co.

Pleh C, Lukacs A, Racsmany M.Morphological patterns in Hungarian children with Williams syndrome and the rule debates.Brain Lang. 2003 Sep;86(3):377-83.

Reilly, J., Bates, E., & Marchman, V. (1998). Narrative discourse in children with early focal brain injury. Brain and Language. 61(3), 335-375.

Reilly, J., Bellugi, U., Lichtenberger, E., Kreiter, J., & Zamora, A. (2001, July). Talking about space in Williams syndrome. In J. Reilly (Chair), Symposium on Language in Children and Adolescents with Williams Syndrome in Italian, English, and French.

Reilly ,J., Bernicot, J., Vicari, S., Lacroix, A., & Bellugi, U., (2005). Narratives in Children with Williams Syndrome: A Cross Linguistic Perspective. In D. Ravid and H. Bat-Zeev Shyldkrot

(Eds.). *Perspectives on language and language development: essays in honor of Ruth A. Berman*. Dordrecht, the Netherlands: Kluwer, 303-312.

Reilly, J., Klima, E., & Bellugi, U. (1991). Once more with feeling: Affect and language in atypical populations. In D. Cicchetti (Ed.), Development and Psychopathology. Cambridge, MA: Cambridge University Press.

Reilly, J., Losh, M. Bellugi, U., & Wulfeck, B. (2004) Frog Where Are You?: Narratives in children with specific language impairment, early focal brain injury and Williams Syndrome, *Brain and Language,88,*229-247.

Robinson BF, Mervis CB, Robinson BW. (2003). The roles of verbal short-term memory and working memory in the acquisition of grammar by children with Williams syndrome. Dev Neuropsychol.23(1-2):13-31.

Singer-Harris, N.G., Bellugi, U., Bates, E., Jones, W., & Rossen, M.L. (1997). Contrasting profiles of language development in children with Williams and Down syndromes. In D. Thal & J. Reilly (Eds.), "Origins of Language Disorders." Special Issue of *Developmental Neuropsychology,345-370*.

Slobin, D.I. (Ed.), 1985, 1992, 1997. The Crosslinguistic Study of Language Acquisition, Vol. 1: The data (1985); Vol. 2: Theoretical issues (1985); Vol. 3 (1992);Vol. 4 (1997); Vol. 5: Expanding the contexts (1997). Erlbaum, Hillsdale, NJ.

Sokolov, J. and C. Snow, eds. (1994). Handbook of Research in Language Development using CHILDES. 1994, Lawrence Erlbaum: Hillsdale, NJ.

Von Arnim, G., Engel, P. (1964). Mental retardation is related to hypercalcemia, *Developmental Medical and Child Neurology* 6:366-377.

Volterra V, Capirci O, Pezzini G, Sabbadini L, Vicari S.Linguistic abilities in Italian children with Williams syndrome.Cortex. 1996 Dec;32(4):663-77.

Vicari S., Bellucci S., Carlesimo G.A. Procedural learning deficit in children with Williams syndrome. Neuropsychologia, 2001, 39, 665-677.

Vicari S., Caselli M.C., Gagliardi C., Tonucci F., Volterra V. (2002). Language acquisition in special population: A comparison between Down and Williams syndromes. Neuropsychologia, 2002, 40, 13, 2461 - 70.

Figures:

1. Morphological errors in French: WS.CA. MA

2.Scatter plots: English and French Morphological Errors

3.Book picture with spatial errors

4.French social evaluation

5. Social Evaluation in American and French children