

THE NARRATIVE PROFILE IN WILLIAMS SYNDROME: THERE IS MORE TO STORYTELLING THAN JUST TELLING A STORY

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Introduction

Facing the multiplicity of internal and external stimulation, individuals are left with the central task of constructing meaning out of their experience. This is accomplished by integrating both internal and external reality into a coherent, yet complex and diverse narrative plot.

Narrative is probably one of the most distinguishable human capacities involving the coordination among a diversity of neurocognitive processes (Gonçalves *et al.*, 2004; Rubin and Greenberg, 2003). Both in narrative production and comprehension, a complex interaction of linguistic,

cognitive, affective and social abilities is present (Capps *et al.*, 2000; Reilly *et al.*, 2004). Among others, narrative ability has been associated with executive temporal ordering (Labov and Waletzky, 1967; Mar, 2004), theory of mind (Astington, 1990; Fletcher *et al.*, 1995; Gallagher *et al.*, 2000), lexical, semantic and prosodic language devices (Losh and Capps, 2003), as well as memory (Frisk and Milner, 1990a,b; Rubin and Greenberg, 2003; Wheeler *et al.*, 1997) and emotional processes (Gernsbacher *et al.*, 1992; Oatley, 1999; Partiot *et al.*, 1995).

Additionally, given its associative nature, narrative seems to be dependent of a distributed network of intercortical and

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cortical-subcortical connections (Cozolino, 2002; Siegel, 1999). Previous studies have shown that narrative comprehension and production require the activation of multiple brain regions that are not restricted to the areas typically associated with sentence processing (Mar, 2004).

The importance of studying narrative is related with its contributions to the analysis of spontaneous language production and to the exploration of language development, in its structural (e.g., grammatical, syntactic) and content dimensions (e.g., inferences), both in typical and atypical populations (e.g., Losh and Capps, 2003; Losh *et al.*, 2000; Reilly *et al.*, 1998, Reilly *et al.*, 2004). Studies with neurodevelopmental disorders with a genetic basis are useful in elucidating the complex interactions between genes, environment, and cognition, and also in understanding how these atypical constraints affect narrative development.

Williams Syndrome (WS), a genetic disorder characterized by a deletion on chromosome 7 q11.22-23, has been presented as an intriguing syndrome where an apparent preservation of narrative production and language coexist with profound intellectual deficits, specially visual-spatial and executive functioning impairments (Bellugi *et al.*, 2000; Martens *et al.*, 2008).

Most of the initial interest in WS research was fostered by this apparent dissociative pattern of neurodevelopment (Bellugi *et al.*, 1990). In fact, there seems to be evidence for some type of expressive language preservation (Bellugi *et al.*, 1997) and receptive vocabulary knowledge (e.g., Brock *et al.*, 2007) in WS when compared with other genetic syndromes characterized by mental retardation (e.g., Down Syndrome). However, a detailed investigation of language subcomponents has demonstrated several atypicalities,

namely in terms of syntax, morphology, lexical-semantic processing, and pragmatics, with evidence also of an atypical developmental pathway (Bello *et al.*, 2004; Capirci *et al.*, 1996; Clahsen and Almazan, 1998; Jarrold *et al.*, 2000; Karmiloff-Smith *et al.*, 1997; Smith *et al.*, 1998; Smith *et al.*, 2002; Laing *et al.*, 2002; Laws and Bishop, 2004; Mills *et al.*, 2003; Neville *et al.*, 1994; Stevens and Karmiloff-Smith, 1997; Temple *et al.*, 2002; Thomas *et al.*, 2006).

Concerning narrative production in WS, several studies have also pointed out the existence of several problems (Gonçalves *et al.*, 2004; Heinze *et al.*, 2007; Jones *et al.*, 2000; Losh *et al.*, 2000; Reilly *et al.*, 2004). For example, Reilly and colleagues (2004) found significant difficulties on cognitive measures of structural and thematic narrative dimensions (e.g., use of cognitive inferences) by individuals with WS, suggesting a failure to integrate the different elements of narrative. However, the same authors found significantly greater amount of evaluative (social engagement) devices in WS narratives, in comparison with other developmental disorders. Some authors have also proposed the influence of pragmatic aspects of WS social profile on tasks of lexical access, explaining the rare-word usage (Thomas *et al.*, 2006).

Together these findings suggest a dissociation between the expressive component and the cognitive dimension of narrative. In fact, in spite of the proficiency shown in the use of some linguistic forms (e.g., morphosyntactic abilities) and social tools (evaluative devices), the more cognitive dimensions of narrative (e.g., reference to the motivations and goals of story's characters, linked to theory-of-mind ability, integration of the different episodic and thematic elements of the story) seem to be compromised, probably due to the cognitive deficits

that characterize WS (Reilly *et al.*, 2004). This parallels other domains of language, where the expressive domain seems to be dissociated from the receptive domain. For example, although it seems that many children with WS are able to use metaphors, analogies, similes and idioms (Semel and Rosner, 2003), they present difficulties comprehending figurative language (e.g., differentiating between lies and jokes) and answering to second-order knowledge questions (Sullivan *et al.*, 2003).

It is important to note that WS cognitive profile occurs in a background of atypical brain development and organization. In fact, neuro-imaging studies have brought evidence for neuroanatomic abnormalities in these patients, namely cerebral hypoplasia, despite a distinct topographic distribution of volume reductions and preservations (Meyer-Lindenberg *et al.*, 2005; Reiss *et al.*, 2000; Reiss *et al.*, 2004). In addition, cortical and thickness profile abnormalities (Kippenhan *et al.*, 2005; Thompson *et al.*, 2005; Gaser *et al.*, 2006; Schmitt *et al.*, 2001; Van Essen *et al.*, 2006) with morphological changes in cerebral shape (Schmitt *et al.*, 2001), central sulcus and sylvian fissure (Eckert *et al.*, 2006; Jackowski and Schultz, 2005) have also been documented. Particularly interesting is the finding of the presence of bilateral patterns of symmetry in WS (Van Essen *et al.*, 2006), namely in brain regions that are highly asymmetric and lateralized in normal development, such as superior temporal gyrus (Sampaio *et al.*, 2008) and perisylvian cortices (Eckert *et al.*, 2006), strongly related with language processes, and thus with narrative production. Thus, these reports suggest delayed or abnormal brain developmental trajectories that underlie specific patterns of cognitive function in WS.

In sum, the study of narrative in WS

brings back one of the most controversial themes about neurocognitive development: the existence or not of neurocognitive dissociation phenomena (e.g., Quartz and Sejnowsky, 1997; Thomas and Richardson, 2006). Contrary to the initial claims of cognitive dissociation and language preservation in WS (Bellugi *et al.*, 1988, Bellugi *et al.*, 1990, Bellugi *et al.*, 1992; Thal *et al.*, 1989; Wang and Bellugi, 1994), more recent studies showed that such a complex neurocognitive process as narrative production (a highly cognitive associative and neural distributed task) is not spared in the context of overall mental retardation with significant deficits in most of the cognitive and neurological processes (Bellugi *et al.*, 2000; Losh *et al.*, 2000; Reilly *et al.*, 2004).

This study aimed to specifically explore the structure, diversity and complexity of the narrative profile of WS, complementing previous studies on narrative production in this genetic syndrome (e.g., Reilly *et al.*, 2004). By using a new scoring system, structural (coherence), process (complexity) and content (multiplicity) aspects of fictional narrative production in WS were then compared with those of a typical development group. This analysis aimed to provide a more detailed analysis of narrative profile in WS, focusing on its social, emotional and (meta)cognitive dimensions, and giving less attention to its linguistic dimensions (e.g., grammatical competence).

Method

Participants

A group of twelve participants (4 female and 8 male), diagnosed with WS, with an age range between 9 and 31 years, ($M = 16.5$, $SD = 6.88$) was compared with

a typical development group, individually matched on chronological age ($M = 17.4$, $SD = 7.52$), gender and socio-economic level, measured through an adapted version of Graffar Scale (Graffar, 1956).

Participants with WS were recruited at a Genetic Medical Institute (Portugal) and Genomic Foundation in Galicia (Spain). WS diagnoses were made by fluorescent in situ hybridisation (FISH) confirmation of elastin gene deletion (Korenberg *et al.*, 2000). Exclusion criteria included the presence of severe sensorial or speech disorder, as well as co-morbidity with severe psychopathology not associated with the syndrome. Controls were typically developing individuals without evidence of psychiatric, neurological disorder or cognitive impairment. Each participant and their guardians gave written informed consent for their participation in the study via consent forms, after a detailed description of the study.

Given the rare incidence of this syndrome, a small number of participants were recruited. These are part of a sample of individuals who have been recruited by our laboratory for the last 5 years.

The choice of a control group matched for chronological age rather than mental age was due to several reasons. First, our major aim was to compare a group of individuals with WS to a group of typically developing individuals and to understand which differences exist in the narrative profile of individuals with WS and, in particular, what deviates from typical development. Second, control participants who are matched on IQ (e.g., Down Syndrome or nonspecific mental retardation) generally have language abilities that are inferior to those of individuals with WS.

Third, as pointed out by Levitin and colleagues (2002), full-scale IQ measures

are not a reliable comparison measure, due to the profile of cognitive fractionation that characterizes WS.

By comparing narratives of WS with chronologically matched typically developing individuals we expected to identify which aspects of narrative structure, process and content are more affected by development constraints and which are more resilient.

Instruments

To assess general cognitive functioning (Full Scale IQ), participants 8-16 years of age were administered the Wechsler Intelligence Scale for Children-Third Edition (WISC-III) (Wechsler, 1991), while participants over 16 years old were administered the Wechsler Adult Intelligence Scale- Third Edition (WAIS-III) (Wechsler, 1997).

In order to elicit narrative production the pictures book "Frog, where are you?" (Mayer, 1969) was used, as in previous studies (e.g., Capps *et al.*, 2000; Losh *et al.*, 2000; Reilly *et al.*, 1998, Reilly *et al.*, 2004). This is a storybook without words, composed by a set of images with the aim of eliciting a story. This task has been used in several language studies, mainly because of the multiplicity of processes, contents and structural elements that can be elicited by the images (Reilly *et al.*, 1998). The procedure followed by this study was based on the instructions proposed by Reilly and colleagues (2004): (1) presentation of the book, with the following instruction: "This book tells the story of a boy, a frog and a dog; I want you first to see these images and then tell me the story while you see again the images"; (2) the participant turns the pages; (3) the participant tells the story while observing

TABLE I
Socio-Demographic Characteristics

	Williams Syndrome (N = 12)				Control Group (N = 10)			
		Range	M	SD		Range	M	SD
Gender		-	-	-		-	-	-
Female	8				7			
Male	4				3			
Age		9-32	16.5	6.88		9.32	17.4	7.52
Socio-Economical Level*		5-3	3.85	0.77		5-2	3.77	0.89

* Graffar Scale (1 - high level; 5 - low level)

the images. All the narratives obtained were videotaped, transcribed and analysed in terms of its structure, process and content, based on specific coding systems developed by Gonçalves and co-workers (2002) (see TABLE II for a more detailed description of the coding systems used).

All the systems evaluate different subdimensions of the narrative structure, process and content in a 5-point Likert scale. Besides the score for each individual subdimension, a global score can be obtained for the three narrative major dimensions (content, process or structure) by summing each subdimension scores corrected for the deviations, using the following formula: $[3 \pi_i + \text{sgn}(\pi_i - 3)(\pi_i - 3)^2] + 4$ (where π_i =value of each parameter). Acceptable levels of inter-rater reliability (86%-96%) and internal consistency (alpha values from .66 to .93) have been described for this coding systems (Gonçalves *et al.*, 2002).

These systems have been applied to different clinical groups (e.g., agoraphobia, depression, and eating disorders), as well as to typically developing individuals of different ages (see Gonçalves *et al.*, 2002a,b; Moreira *et al.*, 2008).

The *System for the Assessment of the*

Structural Coherence of Narrative (Gonçalves and Henriques, 2000a), based on the narrative structure models of Labov and Waletzky (1967) and Baeger and McAdams (1999), was designed for the assessment of the narrative coherence using a coding system composed by four subdimensions: orientation, structural sequence, evaluative commitment and integration. It is worth noting that the dimension of evaluative commitment differs, in some extent, from the concept of "evaluation" measured in other studies (Reilly *et al.*, 2004): in this context, evaluative commitment makes reference to the emotional states of the narrator and to his engagement with the task of story telling (e.g., use of onomatopoeias; interjections; hesitations; modulation of emotional prosody), not including references to the mental states of the characters (this is assessed separately, as emotional and cognitive subjectifying). *The System for the Assessment of Narrative Process Complexity* (Gonçalves and Henriques, 2000b), based on the systems developed for the analysis of narrative process in oral narratives previously developed by Angus and colleagues (1999) and Gonçalves (1995), is aimed at assessing the level of complexity in the narrative process using four indexes:

objectifying, emotional subjectifying, cognitive subjectifying and metaphorizing. Finally, the *System for the Assessment of Narrative Content Diversity* (Gonçalves and Henriques, 2000c) was designed to assess the diversity of the narratives in terms of themes, events, settings and characters.

Procedure

After collecting data about sociodemographic characteristics, diagnosis, clinical history, and general cognitive functioning, all participants were administered the narrative induction task. Participants were presented with the 24-page wordless picture book, *Frog, where are you?* (Mayer, 1969) and asked to tell the story to the experimenter, while they looked through the book. Participants' narratives were videotaped and transcribed. They were then analyzed according to the structure, process and content systems described above.

Three psychologists, blind to the study hypotheses, were involved in the process of collecting socio-demographic data, conducting global cognitive assessment and administrating the narrative induction task. Six additional psychologists, equally blind to the participants' diagnosis, coded the narratives (two judges for each coding system). The observers were trained (at least 60 hours) in narratives' coding, based on the Narrative Analysis System (Gonçalves *et al.*, 2002). Inter-rater agreement was calculated for all the narratives using the Within Class Correlation Coefficient (Everitt and Hay, 1992) and all discrepancies were solved by consensus. Inter-rater agreement before consensus was above 80% for all the subdimensions analyzed.

Results

There was no significant group differences with respect to socio-demographic characteristics, including age ($t(19) = .291, p > .05$) and socio-economic status – Graffar Index ($Z = -.932; p > .05$) (see TABLE I).

General Cognitive Functioning

Mean distribution of Full Scale Intelligence Quotient (FSIQ) in WS was found to be within the moderate mental retardation interval, with equally low scores in verbal and performance IQ. In comparison, as expected, normal development participants showed significantly higher levels in terms of General IQ, as well as Verbal and Performance IQ (see TABLE II).

Additionally, no significant differences were found between Verbal and Performance IQ ($t(11) = 1.410, p > .05$). Finally, an heterogeneous profile was found for the different IQ subscales (see TABLE III), with very low scores obtained for the block design subtest ($M = 1.00; SD = 0.00$).

Narrative Measures

Global Narrative Measures

In terms of global narrative measures, we found a significant lower quality of WS narratives, when computing mean scores for all the three narrative dimensions: structure, process and narrative content (see Appendix). Overall, WS narratives were characterized by low levels of structural coherence and process complexity, even though with moderate levels of content diversity. In comparison,

TABLE II
Subdimensions of narrative structure, process and content

Subdimension	Description	Question answered	Example (shown in bold)
Structural coherence			
<i>Orientation</i>	Does narrative make reference to: - characters? - the social / spatial / temporal / personal context where behaviours take place? - past relevant events that have contributed for the occurrence of current behaviours? - relevant events that have occurred after the central event?	What is the context of the narrative?	This story is about a boy, a dog, and a frog who lived together in the same house. Once, when the boy was sleeping, the frog escaped jumping through the window.
<i>Structural sequence</i>	Does narrative make reference to: - an initial event? - an internal response to the event? - an action? - the associated consequences?	And then, what happened?	It was night, the boy and the dog were sleeping , but not the frog. The frog quietly approached the window and jumped. In the morning, when the boy woke up, he didn't see the frog and started yelling , calling for him.
<i>Evaluative commitment</i>	Does narrative make reference to: - the emotional states of the narrator? - the extent of his commitment with the narrative?	Why have the narrative been told?	Wow! Look! Finally they found the frog! They were all so happy!
<i>Integration</i>	Are the elements of narrative described in an integrated/coherent manner?	Is the guideline of discourse clear?	This story is about a boy, a dog and a frog. Once, the frog disappeared and the boy and the dog went both after the frog. After a series of disadvantages, they found the frog. The frog went after his family and he finally found his parents, brothers, and sisters. In the end, all were happy.
Narrative process complexity			
<i>Objectifying</i>	Does narrative make reference to: - Sensorial elements related with the episode's description? In what extent?	What are the sensorial experiences of the characters?	The bees went after the dog. Ohhh... It must have been very painful to feel those needles in his skin.
<i>Emotional subjectifying</i>	Does narrative make reference to: - Emotional states related with specific events? In what extent?	What are the emotional experiences of the characters?	After several adventures and risks, they finally found the frog. They were all very happy with that: the boy and the dog were happy because they found the frog, and the frog was very happy for having found his family.
<i>Cognitive subjectifying</i>	Does narrative make reference to: - the cognitions, ideas, thoughts and plans of the characters referred? In what extent?	What are the cognitive experiences of the characters?	While he was calling for the frog, the boy was thinking about what could have happened to him and where he might have been.
<i>Metaphorizing</i>	Does narrative make reference to: - the meanings constructed by the narrator, in order to make sense of the episodes described?	In what way does the narrator make sense of the events described?	In the end, they were all very happy. The boy learned that when you really want something, you have to work hard for it but, in the end, you'll be rewarded. And his reward was to finally find the frog.
Narrative content diversity			
<i>Themes</i>	How many themes are introduced in the narrative?	What are the thematic contexts introduced in the narrative?	
<i>Events</i>	How many action sequences are described?	And then, what happened?	Very curious, the dog was trying to catch the hive , when suddenly it felt down and the bees started chasing the dog.
<i>Scenarios</i>	Does narrative make reference to: - the environment that surrounds the events described?	What is the context where action takes place?	While the boy was at home , sleeping, the frog jumped from the window.
<i>Characters</i>	How many (real or imagined) characters are introduced in the narrative?	Who are the agents of the actions described?	The boy and the dog tried to find the missing frog.

controls' narratives presented significantly higher levels of structural coherence, process complexity and content diversity (see TABLE V).

Narrative Structural Coherence

As stated previously, WS narratives were characterized by poor and significantly lower levels of structural coherence than those obtained for typical development participants.

When we compared each of the four subdimensions of narrative structure (see TABLE VI), WS showed "very low" or "low" levels of orientation, structural

sequence and integration. In contrast, the subdimension of evaluative commitment approached moderate levels (equal or superior to a mean score of 3).

In comparison, the typical development group presented significantly higher scores for each one of structure parameters: orientation, structural sequence, evaluative commitment and integration.

Narrative Process Complexity

As found for narrative structure, the complexity of narrative process in WS was significantly inferior to that observed in the typical development group. In

TABLE III
Cognitive profile of patients with WS and controls

	Williams Syndrome (N=12)				Control Group (N=10)				t	p
	Min.	Max.	M	SD	Min.	Max.	M	SD		
Full Scale Score	40	61	47.42	6.07	84	132	100.70	13.71	11.394	<.001
Verbal IQ	46	73	53.25	7.63	82	131	100.50	14.40	9.341	<.001
Performance IQ	46	61	50.08	4.98	84	123	101.80	13.74	11.298	<.001

TABLE IV
IQ subtests scores for participants with WS and controls

IQ Subtests	Williams Syndrome (N=12)		Control Group (N=10)		t	p
	M	SD	M	SD		
Information	2.00	1.21	8.50	2.01	8.956	<.001
Similarities	3.00	2.41	10.20	2.20	7.313	<.001
Arithmetic	2.50	3.09	10.10	3.11	5.727	<.001
Vocabulary	2.42	2.07	10.00	3.53	5.995	<.001
Comprehension	2.17	2.55	12.30	3.47	7.673	<.001
Picture Completion	3.00	3.69	11.00	2.16	6.319	<.001
Coding B	1.67	0.89	10.50	1.43	16.962	<.001
Picture Arrangement	2.00	1.48	8.70	2.71	6.000	<.001
Block Design	1.00	0.00	11.10	2.96	10.787	<.001
Object Assembly	2.27	1.79	10.10	3.41	6.482	<.001

TABLE V

Global scores for narrative structure, process and content in participants with WS and controls

Narrative Dimensions	Williams Syndrome (N=12)		Control Group N=10)		<i>t</i>	<i>p</i>
	M	SD	M	SD		
Structure	19.83	12.60	61.40	11.97	7.918	<.001
Process	5.00	4.05	31.80	14.98	5.493	<.001
Content	35.17	11.74	47.40	6.26	3.118	<.01

TABLE VI

Scores for the submissions of narrative structure and coherence in participants with WS and controls

Structure Parameters	Williams Syndrome (N=12)		Control Group N=10)		<i>t</i>	<i>p</i>
	M	SD	M	SD		
Orientation	1.75	0.62	4.10	0.74	7.984	<.001
Structural Sequence	1.75	0.62	4.40	0.52	10.293	<.001
Evaluative Commitment	2.67	1.16	4.10	0.74	3.523	<.001
Integration	1.50	0.67	4.10	0.74	8.557	<.001

general, WS narratives were characterized by very low levels (in fact, absolute ground levels with $M = 1$ and $SD = 0$) of emotional subjectifying, cognitive subjectifying and metaphoryzing and low levels of objectifying. These results contrast with those obtained by typical developing controls, who reveal significantly higher levels for all the subdimensions: objectifying; emotional subjectifying; cognitive subjectifying; and metaphoryzing (see TABLE VII).

Narrative Content Diversity

This was the dimension in which participants with WS obtained higher values. However, it is important to note that, even in terms of content diversity, the scores were still significantly lower than those obtained by typically developing controls.

Comparing the different sub-

components of narrative content in WS (see TABLE VIII), the moderate levels of content diversity obtained seem to be influenced by the relatively higher levels of characters' diversity. Very low scores were observed in terms of the diversity of scenarios, events and themes. As observed for the other narrative dimensions, normal controls presented significantly higher results in terms of the diversity of characters, scenarios and events.

It is important to note that "themes diversity" is the only variable where participants with WS are not significantly different from typical development controls. In other words, both typical development and WS narratives presented low diversity of themes. This finding may have been influenced by the nature of the narrative induction task, not particularly effective in eliciting diversity of themes.

In order to account for the effects of age and IQ, we additionally performed an analysis of covariance (see TABLE IX). Data

TABLE VII

Scores for the subdimensions of narrative process and complexity in participants with WS and controls

Process Parameters	Williams Syndrome (N=12)		Control Group N=10)		t	p
	M	SD	M	SD		
Objectifying	1.92	0.79	4.30	0.66	7.615	<.001
Emotional Subjectifying	1.00	0	2.50	0.85	5.582	<.001
Cognitive Subjectifying	1.00	0	2.00	1.16	2.739	<.01
Metaphorizing	1.00	0	1.50	0.52	3.000	<.01

TABLE VIII

Scores for the subdimensions of narrative content and diversity in participants with WS and controls

Content Parameters	Williams Syndrome (N=12)		Control Group N=10)		t	p
	M	SD	M	SD		
Characters	4.08	1.17	4.92	0.32	2.329	<.05
Scenarios	2.25	0.62	3.10	0.74	2.888	<.05
Events	2.58	0.79	3.40	0.70	2.566	<.05
Themes	1.83	0.58	2.00	0.00	1.000	n.sig

n.sig indicates not significant

showed a group effect for the majority of the narrative variables, with the exception of evaluative commitment and narrative content subcomponents, after controlling for the effects of age and IQ.

IQ seems to be associated with the performance on specific subdimensions of narrative process, namely: emotional subjectifying, cognitive subjectifying and metaphoryzing. Not differently, age seems to impact several narrative process dimensions, more specifically cognitive subjectifying and metaphoryzing. On the contrary, no significant effects of IQ and age on structural coherence and content diversity were found ($p > .05$).

Discussion

First of all, concerning intellectual functioning, the present study confirms a moderate mental retardation in WS, with generalized low scores for both the verbal and performance components. These results contrast with previous studies that rather found a dissociation between verbal and performance IQ (Howlin *et al.*, 1998; Jarrold *et al.*, 1998).

The main objective of the present study was to conduct a more detailed analysis of the several components of narrative production in participants with WS, when compared with typically developing controls. In general, the results obtained did not find support for

TABLE IX

The independent effects of age and IQ on narrative performance (structure, process and content) and the effects of group, after covariating age and IQ

Parameter	Effects (f)		
	Group	Covariates	
		Age	IQ
Structure parameters			
Orientation	9.049*	4.027	0.030
Structural Sequence	11.651**	0.893	0.009
Evaluative Commitment	2.384	0.06	0.233
Integration	18.146**	2.579	1.595
Process parameters			
Objectifying	16.076**	0.592	2.280
Emotional Subjectifying	44.298**	2.612	16.361**
Cognitive Subjectifying	14.545**	9.068*	7.129*
Metaphorizing	21.589**	7.751*	11.189**
Content parameters			
Characters	0.050	0.591	0.961
Scenaris	0.000	1.311	1.104
Events	0.549	0.454	2.936
Themes	0.29	0.954	0.229

* $p < .05$; ** $p < .005$

the claim of a spared narrative production in WS, suggesting a poor quality of narrative production in terms of structural coherence and process complexity, even though with moderate levels of content diversity.

It is important to note that the WS narrative profile (relationship between structure, process and content levels) seems to differ from the typical development group. In fact, while controls' narratives seem to be based on higher levels of structural coherence, participants with WS seem to privilege the diversity of contents. Together, these findings suggest that participants with WS not only show significantly lower levels of narrative coherence, complexity and diversity but also that their narrative profile seems to

privilege the diversity of narrative content at the expense of narrative coherence.

However, a detailed analysis of the different narrative subdimensions revealed some interesting data. Considering the different components of narrative structure, we found moderate levels for the "evaluative commitment" subdimension. In fact, this is the only variable of narrative structure where we could not find significant differences between individuals with WS and typically developing controls. It is worth noting that this component is related with the level of emotional commitment of the narrator with the narrative as evidenced by the richness of paralinguistic devices used (e.g., prosody modulation, emphatic stress and other "audience hookers"). This

may suggest a relative preservation of the social-expressive component of narrative construction, consistent with previous studies (Jones *et al.*, 2001; Reilly *et al.*, 2004).

The relative preservation of prosodic aspects of narrative production in WS contrasts with the difficulties found on tasks of prosody comprehension (Catterall *et al.*, 2006; Plesa-Skwerer *et al.*, 2006, Plesa-Skwerer *et al.*, 2007), supporting the dissociation between expressive and receptive aspects of language in this genetic disorder. As suggested by Capps and colleagues (2000), it might be that individuals with WS show an ability to reproduce by rote some evaluative devices, without a fully understanding of their influence on the listener's attention.

Concerning the complexity of the narrative process, the present results seem to be consistent with the finding that individuals with WS make fewer or no inferences of motivations and mental states relative to controls (Reilly *et al.*, 2004), which may be related with ground level scores, although they contrast with some studies showing WS ability in identifying internal states (emotional and cognitive) of the story characters. For example, Jones and co-workers (2001) found that children with WS are significantly more efficient in the identification of the mental and affective states of story characters when compared with Down Syndrome children and normal controls matched on mental age. Nevertheless, posterior studies (Plesa-Skwerer *et al.*, 2006, Plesa-Skwerer *et al.*, 2007) showed that emotion recognition is not spared in WS, since, for example, the recognition of facial and vocal emotional expressions pose difficulties for these individuals. On the one hand, the extreme low levels found for the WS group along with the low levels found for the typical development group may question the

discriminative power of the system used in assessing process complexity. On the other hand, this may be a consequence of differences in the matching process relative to previous works, since in our study a control group matched on chronological age (and not on mental age) was used. Also, the broad age range may have obscured developmental patterns in narrative production, namely the sophistication of theory of mind abilities with increasing age (Perner and Land, 1999).

An interesting finding was the effect of age and IQ on this narrative dimension. In fact, the ability to infer mental states (cognitive and affective) or to make reference to meanings in order to make sense of the events described is an important developmental acquisition, dependent on developmental processes, as well as on general cognitive abilities (as, for example, prefrontal functioning) (Huizinga *et al.*, 2006; Kobayashi *et al.*, 2007; Saarni, 1999; Segalowitz and Davies, 2004). However, due to the small sample size, caution is needed in the interpretation of these statistical findings.

Finally, higher scores were found for the diversity of narrative content, even though still within the moderate range and not significantly different from controls, after controlling for age and IQ. It is interesting to note that these results seem to be influenced by the high score found for the diversity of characters, a subdimension, once again, related to the social dimension of narrative production.

In sum, individuals with WS seem to produce narratives that are significantly less coherent, diverse and complex than typically developing controls. Contrasting with the reliance on coherence as a central device for narrative construction, individuals with WS tend to rely more on the diversity of content as a major

narrative device. Additionally, they seem to compensate for their deficiencies in narrative ability by relying on some social markers of the narrative, such as an emotional commitment with the storytelling (i.e., evaluative commitment).

Overall, these findings are consistent with previous studies showing that narrative abilities are impaired in WS. They add further evidence for a better understanding of the narrative profile in WS, suggesting fractionations within the narrative profile of individuals with WS, where a relative preservation of the social-expressive dimension of narrative (e.g., indexed by scores of evaluative commitment, diversity) coexists with the impairment of more cognitive (e.g., such as references to mental states, motivations and goals of the characters, as indexed by emotional and cognitive subjectifying) and metacognitive aspects of narrative (e.g., integrating individual actions into an overarching theme, as indexed by integration; making meaning from the narrated actions, as indexed by metaphoryzing).

However, some limitations of this study make the generalization of the current results more difficult, namely the broad age range of the participants and the small size of our samples.

Future studies should address these limitations and include other control groups, namely a mental-age or language-related matched control group, to allow the differentiation between the aspects of narrative skills related to language delay in WS from those that are specific of its narrative profile.

In spite of these limitations, the current findings are consistent with the idea that narrative is a complex neurocognitive function. It would be expected that a process that implies an associative and distributed neurocognitive functioning

would be impaired in neurodevelopmental disorders, where brain develops abnormally since the beginning (Annaz *et al.*, 2008; Kamirloff-Smith, 1998, Kamirloff-Smith, 2007; Kamirloff-Smith *et al.*, 2003; Yeo *et al.*, 2007). This was indeed the case in our study. Individuals with WS seem to be affected in overall narrative production, relying on certain narrative devices as a compensatory alternative for most of their deficits. The relative strength of their story-telling devices (evaluative commitment and diversity of characters) may give an apparent idea of effective story telling abilities but, as our data shows, there is much more in story telling than just telling a story.

Summary

Williams Syndrome (WS) is a neurodevelopmental disorder that is characterized by a distinctive neurocognitive and behavioural phenotype, where relative cognitive strengths (e.g., language, narrative production, and face processing) coexist with severe deficits in other cognitive domains (e.g., visual-spatial processing).

By using a new scoring system, this study aimed to explore structural (coherence), process (complexity) and content (multiplicity) aspects of fictional narrative production in WS, taking typical development as reference. In this way, it aimed at providing more evidence on the narrative profile of WS, complementing previous studies.

Results showed that narratives in individuals with WS are significantly less coherent, diverse and complex relative to controls. Contrasting with typically developing controls' reliance on structural coherence, individuals with WS tend to rely more on the diversity of narrative

content as a major narrative device. Additionally, these participants seem to compensate their deficiencies in narrative ability by relying on some social markers of the narrative, such as the emotional commitment with the story telling (i.e., evaluative commitment). Together, these findings bring additional support for the dissociation between expressive/social and cognitive/metacognitive aspects of narrative production in WS.

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Example 2

Narrative transcription of a girl with Williams Syndrome (age = 27)

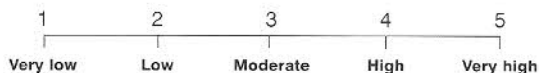
WS narrative transcription

Subject's data: Age = 27; IQ = 61; Gender = female

«Once upon a time, there was a frog, a frog named João, a dog named Faisca and a boy named Hélder. Here it shows the frog jumping up the bottle. João, Hélder and Faisca are sleeping. Hélder wakes up and Faisca remembers to jump up to Hélder's shoulders . Hélder is... I guess he is peeping through the garment and the dog is putting the muzzle inside the bottle. The dog with the bottle and the boy calling for someone... Hélder is near the window and the dog falls down with the bottle. Hélder goes to the dog and the dog caresses him and licks his ear, which is an **act of tenderness**. Here, we see Hélder calling for someone and Faisca, but Faisca is looking at the bees. Faisca, Faisca tries to jump to the bees and the boy starts calling an animal named... well, Beethoven, this little mouse. Here, it shows the dog turned to the tree trying to call the bees' attention , isn't it? And the boy is covering the nose because the animal must have a bad smell. Here the bees are already going after Faisca, aren't they? And the boy, Hélder, hides himself. Here the owl talks with the boy, the boy falls down and the bees go after Faisca. Here Hélder, the boy, is hiding himself and the owl... And here it's already the boy calling for the dog. In the mean time, a deer appears and goes against Hélder, isn't it? And here it goes against Hélder and the dog is barking, isn't it? **Ah!** They don't realize the cliff and the dog falls and Hélder falls, in other words, Faisca and Hélder fall. They fall into the water, don't they? And then the dog goes under Hélder's head. Here Hélder orders him to make less noise:

- Xiiiiuuuu!

And then they climb the trunk of a tree. Here they are already happy, they see two frogs and also... Hélder and Faisca are saying goodbye to the frogs. Hélder and Faisca are also very happy.»



<i>Orientation</i>	2	<i>Objectifying</i>	2	<i>Characters</i>	5
<i>Structural Sequence</i>	2	<i>Emotional Subjectifying</i>	1	<i>Scenarios</i>	2
<i>Evaluative Commitment</i>	3	<i>Cognitive Subjectifying</i>	1	<i>Events</i>	3
<i>Integration</i>	1	<i>Metaphorizing</i>	1	<i>Themes</i>	2
Total¹ (Structural coherence)	22	Total (Process complexity)	6	Total (Content diversity)	42

¹ After the application of the formula: $[\sum \text{pi} + \text{sgn}(\text{pi} - 3) (\text{pi} - 3)2] + 4$

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