

# Can the language of mentally challenged individuals give us an insight into typical language development?

**Evidence from Williams Syndrome** 

JASNA PLANČIĆ

SUPERVISOR Allison Louise Wetterlin

**University of Agder, [2017]** Faculty of Humanities and Education Department of Foreign Languages and Translation



Table of contents

1	Introduction		3
2	Theoretical Background		
	2.1 Nativism (domain-specific perspective)		
	2.2 Constructivism (domain-general perspective)		
	2.3 An alternative to domain-specific and domain-general approach		
	2.3.1	Neuroconstructivism	8
3	Modularity The	eory	10
	3.1 Beyond modularity		11
	3.1.1	Pre-specified Modules vs. a Process of Modularization	11
	3.1.2	Developmental trajectories	12
	3.1.3	Static vs. Dynamic approach	13
4	Williams syndrome		14
	4.1 Early stud	ies on WS	15
	4.2 Studies on	WS in the 1980s	16
	4.3 Language studies on WS from the 1990s until today		17
	4.3.1	Syntax in WS	18
	4.3.2	Morphology in WS	20
	4.3.3	Phonology and prosody in WS	22
	4.3.4	Semantics and pragmatics in WS	23
	4.4 Summary.		25
5	The onset of vo	cabulary acquisition	27
	5.1 Pre-linguis	tic skills	27
	5.1.1	Referential pointing	
	5.1.2	Joint attention	29
	5.1	.2.1 Dyadic vs. Triadic Interaction	29
	5.2 Lexical constraints		
	5.2.1	The whole object assumption	
	5.2.2	The taxonomic assumption	
	5.2.3	The mutual exclusivity assumption	
	5.3 Vocabular	y Spurt	
	5.4 Summary.		
6	Production vs.	Comprehension	

	Referances	
10	Suggestions for further research	
9	Conclusion	
8	Alternative approach to language acquisition	61
	7.2 Dissociation of language and other cognitive skills?	58
	7.1 Spatial language and spatial cognition	56
7	Language and other cognitive abilities – cross domain interactions	54
	6.3 Summary	51
	6.2.2 Semantic priming	48
	6.2.1 Event-Related Potentials	
	6.2 On-line experiments of language comprehension	
	6.1.3 Homonyms	42
	6.1.2 Speeded naming and spontaneous speech	41
	6.1.1 Word-definition	40
	6.1 Off-line experiments of language comprehension	40

#### Acknowledgements

I would like to express my sincere gratitude to my supervisor, Professor Allison Wetterlin, for all the time she invested in me by reading my work and giving me feedback. I thank her for all the meetings and discussions I had with her, as well as, for her constant support and encouragement.

I would also like to thank my colleague, Solveig Eriksen, for many long and inspiring talks we had during our Master's studies. I am grateful to her for making my studying in Norway enjoyable and homelike.

Next, I would like to thank my partner, Rade, for being there for me whenever I needed someone to share my thoughts with and for believing in me even when I, myself, did not.

Last but not least, I have to express my profound gratitude to my family for all the love and support they have given me. I particularly want to thank my sister, Maja, who spent hours in helping me understand medical and biological component of my thesis. Now, as always, she was my best mentor in these domains.

# 1 Introduction

One of the greatest mysteries in the language literature is how children learn language. From the middle of the 20th century until today there have been many theoretical speculations about language acquisition. The major issue among these different theories is the nature-nurture debate. Are there any innate mechanisms in the brain which are specialized for language learning or do children learn language in much the same way as they learn how to walk or write? When it comes to the studies of typical child development, researchers' opinions regarding language acquisition are divided; some support the nativist point of view (Nativism) while the others support the empirical point of view (Constructivism). However, in the studies of atypical child development researchers seem to be favoring the nativist stance.

In this paper, I will look at the studies that have investigated language acquisition of people with Williams Syndrome (henceforward WS). WS is a rare neurodevelopmental disorder, which is usually characterized by intact verbal intelligence alongside severe cognitive impairments. The discovery of WS and its unusual cognitive profile has intrigued many linguists. In the search for the explanation of the WS language phenomenon, the constructivist approach seems to be inadequate because their main argument is that language development is inseparable from the rest of the cognition. On the other hand, the nativist approach offers a very plausible explanation. Nativists argue that the uneven cognitive profile of WS confirms their theory that language is innate and independent from the rest of the cognition. However, these are not the only alternatives. Relatively recently a new approach to language acquisition (Neuroconstructivism) has been developed. Neuroconstructivism embraced both the nature and nurture point of view. By doing so, it challenged the nativist claim that WS can be taken as evidence for dissociation of language and the rest of the cognition.

The aim of this thesis is to investigate whether the language of mentally challenged individuals, such as individuals with WS, can give us an insight into the language acquisition of typically developing children. To accomplish this investigation, I will discuss three main issues surrounding the debate about WS language. First, I will examine whether WS language skills are indeed significantly better than the rest of their cognitive skills. Second, I will try to find out whether the language development of WS children follows the same path as the language development of normal children. And third, I will investigate whether WS non-

verbal cognitive skills have any influence on their language skills. I will approach these issues by analyzing and comparing various language studies done on WS individuals.

I will start this endeavor by presenting the main arguments which stand behind the domain-specific (Nativism) and domain-general (Constructivism) approach to language acquisition. Then, I will show that there is an alternative to these two approaches. This alternative is domain-relevant approach (Neuroconstructivism) to language acquisition.

Next, in chapter three, I will discuss how nativists use Fodor's *Modularity theory* when they attempt to explain unusual cognitive profile of WS people. At the same time, I will present how the inconsistencies in WS cognition are explained from the neuroconstructivist point of view.

In chapter four, I will first introduce the reader to the main characteristics of WS by explaining its cause and by describing the physical, psychological and cognitive profile of individuals born with this syndrome. Then, the review of the empirical studies on WS will follow. This review will be presented in chronological order, and the main focus will be on language studies of English-speaking individuals with WS. After analyzing the findings from these studies, in the summary of this chapter, I will address the first issue about WS language.

Chapter five will address the second issue about WS language. Here, I will compare the vocabulary acquisition of infants with WS to the vocabulary acquisition of typically developing infants.

Chapters six and seven will closely examine the semantics of WS language. In the summary of the chapter seven, I will address the third issue about WS language.

Finally, in chapter eight, I will discuss whether WS language development can be a window into the typical language development.

# 2 Language Acquisition- Theoretical Background

The majority of linguists who study normal and atypical language development from the Chomskyan nativist to the most domain-general constructivist - agree that both genes and environment play a part in child language acquisition. One major difference between the theories is in the claim in what way and to what degree genes and environment contribute to developmental outcomes. Nativists argue that language is innate and a domainspecific module in the human brain. For them, the environment acts simply as a trigger for recognizing and setting native-language parameters of universal grammar (Chomsky, 1972, 1980, 1986; Pinker 1991, 1994). In constructivist's view, on the other hand, the environment is a driving force in language acquisition (Bruner, 1983; Nelson 1985; Clark 1993; Goldberg, 1995; 2006; Tomasello, 2003). Constructivists acknowledge that the ability to learn language is specific to humans. Nevertheless, they claim that the input plays a crucial role in language acquisition (Ambridge & Lieven, 2011). These two divergent theories, however, are not the only alternatives. Karmiloff-Smith (1992) suggests that the nativist and constructivist approach should not necessarily be mutually exclusive. She argues that normal and atypical language development "encompasses the dynamics of a rich process of interaction between mind and environment" (Karmiloff-Smith, 1998, p. 9). Therefore, she proposes a reconciliation of nativism and constructivism displayed in the theoretical framework of neuroconstrucitivism.

In the following sections, I will first present the basic theoretical assumptions of nativism and constructivism, before I introduce the relatively new theoretical approach to language acquisition, neuroconstructivism.

# 2.1 Nativism (domain-specific approach)

The nativist approach to language acquisition is based on Chomsky's (1972, 1986) innateness hypothesis. The innateness hypothesis rests on the claim that the newborn brain is genetically endowed with the underlying system of grammatical rules.

This approach has been especially influential since the late 1950s when Chomsky stated that behaviorism (which argues that the infant brain is a tabula rasa or a blank paper on which linguistic experience imprints its structure) could not explain language acquisition (Chomsky, 1959). Chomsky (1980) argued that it is impossible that language is learned

solely through the input because there is not enough linguistic information available in the environment to account for the complex grammar children eventually acquire. In addition, Chomsky (1972, 1986) noted that adult's everyday language could be full of unfinished sentences, mistakes, and slip of the tongues. However, even with the limited input, children become proficient speakers who can produce and process an infinite number of novel sentences. In other words, according to nativists, the primary linguistic data to which children are exposed to is often very poor to justify the rich linguistic knowledge that is gained in the end. This is also called *Poverty of the Stimulus* argument.

Chomsky (1986) also emphasized that children, regardless of their language background, go through the similar stages of language development, beginning with single words utterances, advancing to the more complex combination of words until finally reaching fully-formed adult-like sentences. This development is typically very rapid, with first words appearing at around twelve months, first word-combinations at around eighteen months, and by the age of thirty months, children have usually acquired most of the essential grammatical rules of their language. The instance of uniformity and rapidity of language acquisition is another argument nativists use to claim that some aspects of language must be "genetically imprinted" in the human brain (cf. Radford, 2004; Gleason & Ratner, 2009).

According to Chomsky (1986), humans possess a language faculty for processing language. Theoretically speaking, the language faculty "provides children with a genetically transmitted algorithm for developing a grammar" (Radford, 2004, p. 11). Nativists argue that the language faculty is an autonomous area, separated from all other cognitive areas in the brain (see Fodor, 1983; Pinker, 1991, 1994). In other words, "the brain mechanisms by which the child learns language are not only considered innate, but also entirely domain specific - that is, dedicated only to language learning" (Karmiloff-Smith, & Karmiloff, 2001, p. 5). In nativist view, therefore, children are born with an innate language module. This will be further discussed in chapter 3.

# 2.2 Constructivism (domain-general approach)

The constructivist approach to language acquisition advocates that essentially all aspects of language (including grammatical categories) are learned or "constructed on the basis of input together with general cognitive, pragmatic and processing constraints" (Goldberg, 2006, p. 3). Contrary to nativists, constructivists reject the idea that children are born with an innate knowledge of grammar (see Goldberg, 1995, 2006).

The constructivist theory has its roots in the general concept of psychological empiricism. The idea that linguistic knowledge is acquired only through observation, experience and evidence originates from the psychological approach to child development of Jean Piaget (1952). For Piaget and his disciples, such as Sinclair (1971, 1987), language acquisition involves domain-general learning processes which operate across all areas of linguistic and nonlinguistic cognition. In their view, the infant comes to the world with the "knowledge-empty" mind, in which all aspects of cognition are "emergent products of a self-organizing system that is directly affected by its interaction with the environment" (Karmiloff-Smith, 1992, p. 9). The way in which children learn language is not different from the way in which they learn to use other skills since all areas in the brain are considered to be interconnected. For instance, Sinclair (1971) proposed that the child's ability to set Russian dolls one inside the other forms the basis for the child's later ability to comprehend how sentences can be embedded within one another. Hence, according to constructivists, the general cognitive development is a prerequisite for the normal language development (see section 5.1).

Most constructivists study language from the socio-pragmatic perspective (e.g., Nelson, 1985; Clark, 1993; Tomasello, 2003), which was initially proposed by Jerome Bruner. Bruner (1983) maintains that the role of social interaction is fundamental to the children's ability to learn language. In social interaction, (initially in a mother-infant relationship, and later in an interaction with the rest of the child's social environment) children start to recognize the rules for communication. One of the earliest signs of communication, from the socio-pragmatic point of view, is "the [children's] ability to make social-pragmatic inferences regarding a speaker's focus of attention and his or her communicative intentions" (Ambridge & Lieven, 2011, p. 3). This ability is commonly called joint attention. Many linguists nowadays recognize the children's maturity to establish joint attention as a very significant step for the normal development of their vocabulary (see section 5.1.2).

We see that, from the both cognitive and socio-pragmatic perspective of the constructivist approach, the brain mechanisms responsible for language learning are domaingeneral. That is, according to constructivists, children use the same mechanisms for language learning as they do for learning about other aspects of the world.

# 2.3 An alternative to domain-specific and domain-general approach

The nature-nurture debate has been going on for many years now. During all these years, nativist and constructivist stances on language acquisition seem to be irreconcilable. A developmental neuropsychologist, Karmiloff-Smith (1992), argued that this debate is not a constructive one. Instead of opting for nativism or constructivism, she suggests that these theories should be observed as complementary in fundamental ways and that an ultimate theory of language acquisition, and human cognition in general, should include aspects of both nativism and constructivism. Therefore, as an alternative, Karmiloff-Smith (1998) proposes the domain-relative framework implemented by neurocostructivism.

#### **2.3.1** Neuroconstructivism (domain-relevant approach)

Neuroconstructivist theory is primarily a theory which focuses on the study of language development. From the developmental perspective, neuroconstructivism tries to embrace both innate predispositions and the interaction with the environment. Karmiloff-Smith (1992), one of the firmest advocates of neuroconstructivism, argued that, with certain provisos, the reconciliation of nativism and constructivism is possible. She suggests the following modifications.

On the one hand, she criticizes "the exclusive focus of nativists like Fodor and Chomsky on biologically specified modules [because such stance] leaves little room for rich epigenetic-constructivist processes" (Karmiloff-Smith, 1992, p. 10). Karmiloff-Smith (1992) argues that children, in the process of language development, become active participants in the construction of their own knowledge. Her claim is that the nativist insistence on a specified language module neglects the importance of learning process, i.e. the process of gradual language development. Therefore, she suggests that an important addition to nativism would be Piaget's view of the developmental process.

On the other hand, Karmiloff-Smith (1992) argues that Piagetian domain-general sensorimotor development alone cannot account for the acquisition of language. Syntax, for instance, does not simply stem from free play or problem solving with toys because, as Karmiloff-Smith explains, neither lining up of objects creates the basis for word order nor does nesting of one toy inside another create the basis for embedded sentences. Therefore, she suggests that constructivism should add some innate domain-specific predispositions which would grant the epigenetic process a head start in language development. In other

words, she proposes that initial language endowment needs to include some innate biological constraints. However, these constraints have to be less detailed and less specific than nativists claim.

As mentioned above, neuroconstructivism strongly opposes nativist explanation of language competence in terms of built-in core knowledge because it disregards the prospect of developmental changes in the brain. Many studies of early brain damage show that human brain is far more flexible than the strict modularity would suggest (see Tillema et al., 2008; Ilves et al., 2014). In this regard, neuroconstructivists stress that "the brain is not prestructured with ready-made representations [rather] it is channeled to progressively develop representations via interactions with both the external environment and its own internal environment" (Karmiloff-Smith, 1992, p. 10). Furthermore, a clear distinction has to be made between the domain-specific and domain-relevant proposal of what is innate. On the one hand, nativists propose a genetic blueprint for pre-specified modules that are present at birth, and on the other, neuroconstructivists propose initial constraints or predispositions that direct attention to relevant environmental inputs. In contrast to domain-specific approach, the domain-relevant approach proposes that "the brain starts out with a number of constraints each of which is somewhat more relevant to the processing of certain kinds of input [e.g., language, face processing] over others, but which [can] become domain-specific over developmental time" (Karmiloff-Smith, 2015, p. 93). In short, instead of nativist prespecified modules, Karmiloff-Smith (1992) proposes a more progressive process of modularization.

In the next chapter, I will first briefly summarize Fodor's Modularity theory, then in section 3.1, I will return to the subject of neuroconstructivism where the proposal of a progressive process of modularization will be discussed in more detail.

# **3** Modularity Theory

The concept of modularity comes from the idea that a system as a whole can consist of a set of parts, each of which has a different specialized function. This concept is at the center of many debates led by cognitive and neuroscientists, and generally by theoreticians who deal with the structure of the mind/brain. Broadly speaking, modularity theory tries to explain to what degree the cognitive domains can be considered separable. In other words, can cognitive domains operate independently of one another? The exact definition of a module varies across disciplines and theoretical approaches.

The most fervent proponent of modularity theory is Jerry Fodor (1983). He claims that certain cognitive processes are self-contained or modular. A module, according to Fodor, is:

"...a closed system (encapsulation) that is not available to conscious awareness (unconscious) is mediated by a dedicated neural system (localization) and deals exclusively with one particular information type (domain-specificity)..."

#### (Ambridge & Lieven, 2011, p. 359)

In linguistics, the concept of modularity is tightly linked to the field of developmental neuropsychology. The main objective of this discipline has been to use patterns of developmental impairment to explain a model of normal state processes and brain structure. In this manner, linguists of a nativist persuasion (e.g., Pinker, 1991, 1999; Pinker & Prince, 1994; Flavell et al., 1993; Levy, 1996) use research findings of clinical cases of individuals with neurodevelopmental disorders, to argue for a disassociation between language other cognitive domains, i.e., to argue for the existence of language module.

The strongest evidence which nativists use to account for language module comes from the examples of the so-called double dissociation (DD). The underlying logic of DD is following: "Due to genetic impairment, child A fails to learn skill X but learns skill Y. Another Child, B, shows the opposite pattern, where he learns Y but not X" (Elsabbagh & Karmiloff-Smith, 2004, p. 5). In line with this logic, researchers deduce that skills X and Y are built upon a different set of genes. Thus, a child who is missing a particular set of genes is assumed to miss the necessary module to develop the skill which is normally built upon that particular set of genes. Pinker (1999) described the example of DD as follows:

"...Overall, the genetic double dissociation is striking [...] The genes of one group of children [Specific Language Impairment] impair their grammar while

sparing their intelligence; the genes of another group of children [Williams syndrome] impair their intelligence while sparing their grammar"

(p. 262).

Many linguists share Pinker's stance on the subject in question (e.g., Bellugi et al. 1994, 2000; Clahsen & Almazan, 1998). Moreover, as the investigations of WS started to develop into more detailed experimental studies (i.e., studies that observe all aspects of language separately), the researchers started to propose more specific, within-language dissociations (cf. section 4.3). Nevertheless, in recent years, the modularity theory has been strongly challenged, especially by the relatively new approach to language acquisition, the above-discussed Neuroconstructivism (cf. section 2.3.1).

# **3.1 Beyond modularity**

As presented in the previous section, the existence of neurodevelopmental disorders with uneven cognitive profiles (such as WS) has led researchers to divide the mind/brain into separate modules. Thus, we have a module for number, face processing, space, language, etc. (Pinker, 1999; Bellugi et al., 1988). Moreover, they argue that each module is being processed within a specialized area of the brain. These modular, domain-specific explanations have also been generalized to studies of typically developing children. Consequently, when comparing cognitive abilities of typically and atypically developing children, different abilities of individuals with disorders are commonly described as either intact or impaired. Neuroconstructivists do not agree with this.

#### **3.1.1** Pre-specified modules vs. the process of modularization

The straightforward application of modularity theory to apparent selective behavioral failures in developmental disorders raises a number of issues. All these issues come from the fact that development is an adaptive process. During this adaptive process individual's cognitive system is trying to "optimize his or her interactions with the physical and social environments" (Thomas et al., 2013, p. 485). Thus, behavioral deficits or achievements that appear during development must be explained in terms of the developmental process itself. With this in mind, neuroconstructivists suggest that the explanations should embrace the key concepts which characterize developmental theories. They are the following:

- (1) **plasticity** the idea that the cognitive system changes its structure in response to experience,
- (2) **interactivity** the idea that cognitive components interact with each other across development,
- (3) **redundancy** the idea that a given task can be accomplished equally adequately by more than one system,
- (4) **compensation** the idea that certain behaviors can be delivered in different ways (perhaps not as well) by alternative systems.

(Thomas et al., 2013, p. 485)

When something goes awry in the process of development, the cognitive system aims to adapt itself to the existing situation by using alternative paths. However, this does not mean that the successful developmental outcomes should be described as intact. From the neurocostructivist point of view, the proper explanation of the behavioral deficits or achievements in developmental disorders should include identifying developmental trajectories and analyzing how innate constraints have been altered by the disorder during different developmental times (see Bishop, 1997; Karmiloff-Smith, 1998; Temple, 1997; Thomas, 2013).

#### **3.1.2** Developmental trajectories

Neuroconstrutivists stress that, in order to explain developmental outcomes more accurately, it is important to chart full developmental trajectories (Karmiloff-Smith, 1998). By looking at developmental trajectories, researchers can assess how changes occur from infancy onwards. Moreover, by analyzing the trajectories, they can discover how, at different developmental times, the interaction among the various brain parts may change. For example, a process that is essential at Time 3 may be irrelevant for a process at Time 5. However, the developmental outcome. For this reason, neuroconstructivists insist that developmental timing is a vital factor that needs to be considered when studying language development, especially in the atypical case (Karmiloff-Smith, 2009).

Some studies on WS language development have revealed that individuals with WS, when compared to other mentally disabled groups, show different language results in infancy or early childhood than in adulthood (see Volterra et al., 1996; Paterson et al., 1999; Paterson, 2000). These findings imply that it is not enough to study the end state of language

development and then arrive at a conclusion that language in WS is intact or impaired. For example, the subtle differences found during the process of language acquisition in WS cannot confirm that their language is intact (Paterson, 2000). These differences can only indicate that the brain found its way, by using different cognitive processes, to adapt its mechanisms and arrive at a more or less successful outcome.

#### 3.1.3 Static vs. Dynamic approach to language acquisition

To fully comprehend the neuroconstructivist approach one should also understand that the interaction between nature and nurture, proposed by this school of thought, is a complex interaction between genes and environment. Karmiloff-Smith (1998) clarifies that:

"...on the gene side, the interaction lies in the outcome of the indirect, cascading effects of interacting genes and their environments and, on the environment side, the interaction comes from the infant's *progressive* selection and processing of different kinds of input"

(p. 390)

From both nativist and constructivist point of view, the notion of 'environment' is perceived as a static factor. This is, according to Karmiloff-Smith (1998), another adjustment that needs to be made when endeavoring to understand language acquisition. She explains that both normal and atypical language development is, by all means, a dynamic process. Thus, the way children process language inputs is probably altered repeatedly under the influence of the development itself. For this reason, Karmiloff-Smith (1998) emphasized that the very research has to have a dynamic approach to language acquisition. Applying the static approach to atypical language acquisition might yield questionable conclusions.

# 4 Williams Syndrome

Williams syndrome (WS) was first identified in 1961 by a New Zealand cardiologist John C. R. Williams. He described it as a condition involving serious heart defect, mental retardation, and distinctive facial features (Williams, Barrett-Boyes, & Lowe, 1961). In some of the earlier articles, WS was also defined as the syndrome with idiopathic infantile hypercalcaemia because it involves abnormal calcium metabolism (Beuren, Apitz, & Harmanz, 1962; von Arnim & Engel, 1964). Two decades after the initial recognition, researchers discovered many significant characteristics of WS which helped them understand the psychology and biology of individuals born with this syndrome. However, it was not before 1993 that the genetic basis of WS was identified (Ewart et al., 1993). Today, it is known that WS is caused by a micro-deletion of approximately 20 contiguous genes on the long arm of chromosome 7 (Korenberg et al., 2000). The estimated occurrence of this neurodevelopmental disorder is 1 in 20,000 live births (Morris et al., 1988). The more recent study, however, rates its prevalence of nearly 1:7,500 (see Strømme et al., 2002).

The lack of the genetic material causes a number of physiological and psychological symptoms which characterize WS. Some of the frequent health issues related to WS are cardiovascular and renal abnormalities, general "failure to thrive" in infancy, and abnormal sensitivity to certain sounds - hyperacusis (von Arnim & Engel, 1964; Morris et al., 1988; Hagerman, 1999). The physical appearance of WS individuals is marked by low height, slight build, and specific facial morphology. Facial features of people with WS are sometimes referred to as "pixie-like" or "elfin" facies (Jones & Smith, 1975). WS individuals typically have a broad forehead, blue eyes with a stellate iris pattern, full cheeks, a short-upturned nose, a wide mouth, prominent lips, and small and widely spaced teeth (Semel & Rosner, 2003). The most common psychological characteristic of WS individuals is a pronounced friendliness and an obvious predisposition to empathic behaviors (Doyle et al., 2004; Frigerio et al., 2006).

At the cognitive level, WS children show evident developmental delays and learning disabilities. Nevertheless, what researchers find intriguing is the fact that in the adult phenotype WS individuals exhibit uneven cognitive profile (Bellugi et al., 1994). Namely, an overall IQ score (on the Wechsler Intelligence Scale for Children – Revised, WISC-R) of WS individuals is ranging from 40 to 90. With these wide-ranging scores, they fall into the category of moderately to mildly intellectually disabled (Mervis & Becerra, 2007). Yet, when

one examines their scores on different IQ subtests separately, it turns out that not all their cognitive abilities are equally impaired. For instance, WS subjects show very good results on language tests (Bellugi et al., 1988) while their visuo-spatial abilities are very limited (Atkinson et al., 2001; Farran & Jarrold, 2003). Furthermore, the results WS subjects achieve on standardized tests of face recognition usually fall into the range of normal performance (Gagliardi et al. 2003), while they exhibit severe deficits in numerical cognition, problem-solving and planning (Bellugi et al. 1988, 1994; Paterson et al. 1999; Paterson, 2000). Bellugi and her colleagues (1994) argued that the low overall IQ score of WS individuals simply camouflages the obvious discrepancies in their specific cognitive abilities. They claimed that some of WS true intellectual potentials could not be mirrored in their general IQ scores.

In the following sections, I will give an overview of the investigations on WS population with the special focus on language studies. This will be a chronological review, beginning with the earliest studies on WS from the 1960s and 1970s and finishing with the more specific language studies on WS conducted during the 1990s until the present day.

# 4.1 Early studies on WS

Early investigations of WS were usually multifaceted studies which presented a rather broad description of WS. These studies observed medical, physiological, behavioral, cognitive and linguistic aspects of the WS profile. One such study was conducted by von Arnim and Engel in 1964. They investigated the profiles of four WS individuals who were between 5 and 15 years of age. The participants' IQ scores ranged from 43 to 56. In the summary of their study, von Arnim and Engel (1964) characterized the participants as individuals who have physical growth deficiency, poor motor coordination, and periodic signs of unreasonable anxiety, but on the other hand excellent social skills and an "unusual command of language" (p. 367). They also pointed out that "loquacity combined with friendliness and a great ability to make interpersonal contacts makes them appear brighter and more intelligent than in fact they are" (Arnim & Engel, 1964, p. 375).

A decade later, Jones and Smith (1975) published their research data on 14 WS subjects. The participants in this study were children and adults with WS who were aged 3 months to 23 years. Their full IQ scores ranged between 41 and 80. This study described WS behavior as "friendly, loquacious, and cocktail party manner" alongside mental deficiency (Jones and Smith, 1975, p. 719).

In 1978, Bennett et al. conducted a study of WS in which they used the McCarthy Scales of Children's Abilities (McCarthy, 1972). This was the first endeavor to systematically present measured data on individuals with WS. The study included seven children with WS who were 4;6–8;5 years of age. Their overall IQ scores ranged between 30 and 81 (mean 53.9). The results of this study showed that all seven participants scored better on tests of verbal ability than on the tests of fine motor and gross motor abilities. Bennett et al. (1978) concluded that WS language skills were superior as opposed to their poor motor skills and cognitive impairments.

Even though these pioneering studies of WS offered a very general picture of WS profile, they tended to propose the possible cognitive dissociations in this atypical population, and thus they initiated this debate which is still ongoing.

# 4.2 Studies on WS in the 1980s

In the 1980s, the research data on WS started to accumulate which intensified the controversy about intact language skills in the face of poor non-verbal cognition in WS. For example, contrary to the earlier studies, Kataria et al. (1984) did not report an advantage of verbal skills over gross motor skills in their study. Moreover, they did not confirm that WS individuals have an unusual command of language. Pagon et al. (1987) presented similar results as Kataria et al. (1984). They tested nine individuals with WS (aged between 10 and 20) on the Wechsler Intelligence Scale for Children (WISC; Wechsler, 1976) and reported that seven participants scored above the floor on the verbal scale (floor 45). The participants' verbal IQs were between 47 and 85. However, five of those seven also scored above the floor on the non-verbal tests where their scores ranged from 45 to 69. The ultimate differences between verbal and non-verbal scores of participants in this study did not reach statistical significance. Pagon et al. (1987) noted that only one participant from their study showed a significant verbal advantage which was statistically irrelevant. In addition, the data from another two studies, which used different IQ tests, showed no significant verbal advantage in WS individuals either. In one study, Crisco et al. (1988) administered the Illinois Test of Psycholinguistic Abilities to 22 children with WS, while in the other Greer et al. (1997) administered the Stanford-Binet Intelligence Scale to 15 WS subjects.

The data from work by Udwin and colleagues (Udwin & Yule, 1990, 1991; Udwin, Yule, & Martin, 1987) were not in accordance with the data from the Pagon et al.'s (1987)

study, even though they employed the same IQ measures, the WISC. In contrast to Pagon et al. (1987), however, Udwin and colleagues provided a much larger group of WS participants. They presented the data of 44 individuals with WS who were between 6 and 16 years of age. The scores of a small number of the participants were below the floor on both the verbal and non-verbal scales. However, the scores of the rest of the participants point to a statistically significant dominance of verbal IQs. The verbal IQs of the participants were between 45 and 109 (mean 62.4), while their non-verbal IQs were between 45 and 73 (mean 55.9).

As we can see, most of the studies in the 1980s, which used standardized IQ tests in investigating WS population, did not discover major differences between their linguistic abilities and their other cognitive skills. The exceptions are the investigations of Udwin and colleagues (Udwin & Yule, 1990, 1991; Udwin, Yule, & Martin, 1987) who reported a rather marginal linguistic dominance. Bellugi et al. (1992), however, pointed out a problem with the standardized IQ measures used in these early studies on WS. They emphasized that the vocabulary tests of both the WISC and the Stanford-Binet scales require the participants to give a well-formed definition of words. Little or no credit was given for responding with broad descriptions or exemplars, which is usually the type of the response individuals with WS provide. In contrast, the formal vocabulary tests used during the 1990s and later on only require the participants to name a picture. The scores of individuals with WS on such tests are much higher (see section 4.3.4). Moreover, the vocabulary tests used in early studies include a comprehension subpart which involves practical problem-solving. Skills for problemsolving are profoundly impaired in WS population (see section 4). Conversely, the comprehension part in the formal vocabulary tests used today requires only pointing to one out of four given pictures. All things considered, Bellugi et al. (1992) argued that the low scores of individuals with WS can be explained by the fact that the vocabulary tests used in early studies are not purely linguistic in nature. According to Bellugi et al. (1992), such vocabulary tests require metalinguistic knowledge, which places additional cognitive demands on WS subjects (for further discussion see section 6.2.2).

# 4.3 Language studies on WS from the 1990s until today

From the 1990s onwards, the majority of the studies that investigated linguistic skills in WS were detailed experimental studies. Standardized IQ measures were only used to provide an initial assessment for the purposes of matching WS subjects to other control groups. This new approach to WS research provided systematically measured data from all domains of WS language including syntax, morphology, phonology, lexical semantics, and pragmatics. In the following sections, each of these domains will be reviewed. We will see that the in-depth experimental investigations of WS language abilities have taken the modularity debate one step further, suggesting that the dissociation does not only exist on the cognitive level but also within the language itself. All the findings I incorporated in this review are the findings from various studies conducted on English speaking individuals with WS.

#### 4.3.1 Syntax in WS

In a number of studies, Bellugi and her collaborators (1988; 1994; 1999; 2000) argued that syntactic abilities are relatively strong in WS. These studies examined adolescents and adults with WS, whose results were contrasted with age- and IQ-matched individuals with Down syndrome (DS). Discussing their experimental data, Bellugi et al. (2000) stated that "the general cognitive impairment [found in individuals with WS] stands in stark contrast to their facility and ease in using sentences with complex syntax" (p. 12). Since this is not common in other mentally disabled groups, they proposed that syntactic skills in WS are "spared" or "relatively preserved". For instance, Bellugi et al. (1994) reported that the WS subjects have a much better comprehension of negative clauses, reversible passives, and conditionals than their DS matches. Also, they showed that individuals with WS perform significantly better than the DS matches when required to detect syntactic anomalies and to correct ungrammatical sentences. For example, when given semantically anomalous sentences or sentences violating transformational rules (e.g., subject-auxiliary inversion), the WS subjects were able to convert them into acceptable English sentences.

Examiner:	The picture that painted the person was very kind.
WS subject:	The picture that was painted by the man was very kind

Examiner:Were delivered the flowers by the messenger?WS subject:Were the flowers delivered by the messenger?

(Bellugi et al., 1988, p. 201)

Similarly, Clahsen and Almazan (1998) reported that some more complex syntactic skills, such as the principle of binding, are intact in individuals with WS. In short, the principle of binding explains the syntactic conditioning of referential dependencies between anaphoric

elements (including personal and reflexive pronouns) and their antecedents (Chomsky, 1986). Binding Principle A regulates the distribution of reflexives, whereas Binding Principle B regulates the distribution of personal (non-reflexive) pronouns.

**Principle A**: a reflexive must be locally bound, i.e. it must have a local antecedent.

e.g.,  $Mowgli_i$  is tickling himself<sub>i</sub>/\*<sub>j</sub> j = Baloo bear

**Principle B**: a pronoun must be locally free. e.g.,  $Mowgli_i$  is tickling  $him *_i/_j$  j = Baloo bear

(Adapted from Clahsen and Almazan, 1998)

The experiment Clahsen and Almazan (1998) conducted involved a yes/no sentence-picture decision in which the test stimuli did or did not match with the pictures shown. The subjects were first introduced to the characters (Mowgli and Baloo Bear). The material used in the study were two pictures. In one picture Mowgli was tickling Baloo bear while in the other Mowgli was tickling himself. In the experiment, the subjects were presented with one of the two pictures and the experimenter asked, "Is Mowgli tickling him/himself?". The subjects were requested to reply yes/no respectively. The results showed that the WS subjects performed at ceiling level in both syntactic tasks, like their TD controls.

Interestingly, however, it has not been shown yet that individuals with WS outperform typically developing (TD) controls in syntactic abilities when matched on mental age (MA). For example, Joffe and Valocosta (2007) showed that individuals with WS perform worse than MA-matched TD controls on comprehension of passive sentences and on production of wh-questions. Furthermore, Perovic and Wexler (2007) suggested that some complex aspects of grammar, particularly the aspects which are acquired later in typical development, may be unachievable for individuals with WS. This study examined two syntactic structures, binding and raising. The participants were divided into two groups of children with WS, a group of 6-12 year olds and a group of 12-16 year olds. Their results were compared to the results of TD controls matched on non-verbal MA, verbal MA, and grammar. Ultimately, Perovic and Wexler (2007) reported that the knowledge of the principle of binding in both groups of children with WS was in line with the TD controls. However, they also reported that both groups of children with WS showed considerably poor comprehension of raised constructions (e.g., Homer seems to Lisa to be wearing a hat.) in contrast to unraised constructions (e.g., It seems to Lisa that Homer is wearing a hat.). Raising constructions involve moving ('raising') an argument from a lower non-finite sentence (embedded or subordinate clause) to the higher

subject position (of a matrix or main clause) (cf. Perovic and Wexler, 2007). Similarly to the above-mentioned binding experiment, Perovic and Wexler (2007) used a sentence-picture matching task to test the comprehension of the principle of raising in WS subjects. The subjects were requested to point to one of the two pictures that matched the sentence uttered by the experimenter. To convey the meaning of the raising verb, they used the graphic convention of thought bubbles. For example, the target picture that goes with the stimulus sentence '*Homer seems to Lisa to be wearing a hat*' would present the character of Lisa with a thought bubble in which the other character, Homer, is depicted wearing a hat. Both WS groups who participated in this study showed very poor comprehension of sentences which involved raised constructions.

Since the typical development suggests that the principle of binding is usually mastered until the age of 6 (see Guasti, 2002), while the principle of raising is not mastered before the age of 8 or 9 (see Hirsch & Wexler, in press), Perovic and Wexler (2007) suggested that syntactic abilities of WS population do not exceed the 7-year-old level of normal language development.

#### 4.3.2 Morphology in WS

Empirical data regarding morphology in WS mostly comes from the investigation of regular vs. irregular morphology (see Bromberg et al., 1994; Clahsen & Almazan, 1998; Thomas et al., 2001). Clashen and Almazan (1998) examined the use of English past tense formation. They tested only four children with WS (aged 11;2-15;4) and compared them with roughly matched TD controls on MA. The participants were tested on items of existing irregular verbs (e.g., swim, feed, drive, etc.), novel irregular verbs with stems which rhymed with existing irregulars (e.g., shrim (shram), cleed (cled), crive (crove)), existing regular verbs (e.g., flush, rush, look), and novel verbs with stems which do not rhyme with any existing irregular verb (e.g., spuff, dotch, cug). In the experiment, each verb was first presented in the context of a sentence where the experimenter said, e.g., "Every day I feed my dog", Then, the experimenter started a second (test) sentence, e.g., "Just like every day, yesterday I \_\_\_\_\_ my dog". Finally, the subject was asked to repeat this test sentence and to fill in the missing word. The final results of this study showed that, relative to their roughly matched TD controls, the WS subjects present a selective deficit in irregular past tense formation. Taking these results into consideration, Clashen and Almazan (1998) concluded that the use of regular morphology in WS individuals is intact, while their use of irregular

morphology is impaired. This dissociation between regular and irregular morphology, in a genetic disorder such as WS, has been interpreted as evidence that the innate language faculty consists of two mechanisms. One computational, rule-based mechanism responsible for the application of grammatical rules (such as 'add -*ed* to regular verbs', as in *walk-walked*), and one associative memory mechanism responsible for the retrieval of stored forms from the mental lexicon (as in *sing-sang*). Clahsen and Almazan (1998) thus argued that individuals with WS possess intact computational system (grammar), while their lexical system is impaired.

Thomas et al. (2001) used the same material as Clahsen and Almazan (1998), but a different approach to investigate the performance of WS subjects on English past tense formation. Their aim was to have a larger group of participants and to control for verbal MA between WS subjects and their TD controls. Basically, Thomas et al. (2001) employed a developmental trajectory approach when they analyzed the data from the study. Developmental trajectory approach suggests that it is vital to chart full developmental trajectory before analyzing study data (for the explanation of the term developmental trajectory see section 3.1.2). They examined 21 WS subjects, aged 10;11-53;3, mean chronological age (CA) 22;8 and mean general IQ 45. The performance of the WS subjects was contrasted with four different-age groups of TD controls: three groups of children (6, 8 and 9-year-olds), and one group of adults (age range 17;3–45;0). The chart of these four TD groups demonstrated how the use of English past tense morphology changes across different ages (i.e. across different developmental times). At the age of 6, TD children show significantly better performance on regular morphology, relative to irregular morphology. Nevertheless, this difference decreases with their age. The same tendency is seen among the WS subjects. However, since the verbal MA of WS individuals is usually much lower than their CA, Thomas et al. (2001) matched the controls to their verbal MA as precisely as possible. Finally, by comparing the developmental trajectories of the WS subjects and their controls, Thomas et al. (2001) discovered that the WS subjects' performance on English past tense formation was somewhere between the performance of the 6-year-old and 8-year-old group of TD controls, which was completely in line with their verbal MA. Therefore, it is not surprising that WS population shows better results in regular than in irregular past tense formation since the TD 6-year-olds show that as well.

Thomas et al.'s study indicates how important it is to follow developmental trajectories when investigating atypical language acquisition. It also calls into question the hypothesis that WS individuals can be taken as evidence for the existence of two separate

mechanisms of the language faculty. According to this study, the selective deficit in irregular past tense formation of WS individuals is only the result of their low verbal MA.

#### 4.3.3 Phonology and prosody in WS

Phonological abilities in WS have been of little interest to researchers so far. This is probably because the vast majority of WS population displays fluent and well-articulated speech, unlike other intellectually challenged populations. The articulatory problems, such as the mispronunciation of tricky multisyllabic words or phrases and stuttering, are infrequent (Udwin and Yule, 1990).

In contrast to the phonological abilities, the prosodic abilities of individuals with WS have attracted more attention. For example, Reilly et al. (1990) tested a small group of adolescents with WS and with DS on a story-telling task *"Frog where are you?"*. From their performance, they evaluated that adolescents with WS use far more affective-expressive prosody in contrast to adolescents with DS. The evaluations were made based on the analysis of the sentences produced by the participants during story telling. Several instances of affective vocal prosody were looked at: pitch changes, vocalic lengthening, and modifications in volume. The example of one such evaluation of WS affective prosody is the following:

# "He said, FRO#:GGIE, come back#again"." = high pitch : = lengthened vowel CAPITALS = increased volume # = stress on preceding syllable

(Reilly et al., 1990, p. 379)

As we can see, this evaluation was based on impressionistic data only, no instrumental measuring was used. Setter et al. (2007) conducted a larger, instrumental study, where they measured acoustic variables in the speech of the participants. Their aim was to compare the pitch range and vowel durations of WS children to those of TD children and to find out whether children with WS are perceived differently to CA and LA (receptive language abilities) matches by a group of phonetically naïve listeners. As the material for the experiment, they used the same story telling task (*"Frog where are you?"*) as Reilly et al. (1990). They found out the following. First, with the use Laryngograph, they discovered that

WS group had much higher as well as wider pitch range than either of the two TD groups (CA and LA). Second, the spectrographic analysis of vowel durations showed that the children with WS used similar vowel durations as the LA control group. This indicates that WS individuals may be delayed in comparison to TD children of their own age in this aspect of speech. And third, the naïve listeners recruited for the study perceived the speech of WS subjects as much more emotionally involved than the speech of TD children of a similar LA and CA. Thus, the results are consistent with the proposals from the study by Reilly et al. (1990).

The fact that individuals with WS show different performance from younger TD controls on two aspects of speech (pitch range and the intensity of emotional involvement), indicates that their prosodic development is atypical. This phenomenon can be explained by the neuroconstructivist view (Karmiloff-Smith, 1998), which suggests that, when the development of cognition is disturbed by a gene mutation (as is in WS), the brain starts to follow an atypical trajectory (in order to adapt itself), which can consequently create an uneven development of different cognitive skills (see section 3.1.1). However, this view does not exclude the chance that some aspects of development in WS population will resemble those seen in typical development, even if the developmental path was different. If we assumed that development in WS is just delayed, but follows a typical trajectory, then WS individuals should perform in an identical way as younger TD children on any cognitive task. According to the data from the two above reviewed studies, this is not the case with prosodic skills of WS population.

# 4.3.4 Semantics and pragmatics in WS

Judging by the huge corpus of literature on WS in the field of semantics, lexical abilities of WS population have intrigued researchers the most. This may be due to the fact that receptive vocabulary is one of the aspects of language in which individuals with WS tend to perform better than it would be expected by their MA (Bellugi et al., 1988; Rossen et al., 1996; Clahsen et al., 2004). Namely, various formal vocabulary tests (e.g., the Peabody Picture Vocabulary Test-Revised (PPVT-R), the British Picture Vocabulary Scale (BPVS), and the Gardner Expressive One Word Vocabulary Test (EOWPVT)) demonstrate that the vocabulary performance of adolescents and adults considerably surpasses their overall MA (Bellugi et al., 1988; Rossen et al., 1996; Tyler et al., 1997; Mervis et al., 1999). This is surprising because it is a unique example among groups with mental disorders (Reilly et al.,

1990). However, Volterra et al. (1996) who investigated younger children with WS (9-, 10year-olds) reported that their vocabulary skills do not appear so extraordinary at this age. Instead, Volterra et al. (1996) pointed to an initial delay in receptive vocabulary in children with WS, relative to their TD controls matched on MA. The advantage of individuals with WS, in contrast to the vocabulary skills of DS individuals, becomes noticeable only in their early adolescence (Rossen et al., 1996). Therefore, Rossen et al. (1996) suggested that the remarkable vocabulary in adolescents and adults with WS are a result of developmental changes, not a result of intact language ability.

Even though individuals with WS display a very rich vocabulary, their lexical skills do not seem entirely normal. For example, Bellugi et al., 1994 reported that individuals with WS have an unusual choice of words (e.g., "I will have to *evacuate* the glass", meaning I'll have to *empty* the glass [p. 32]). They also argued that individuals with WS use a lot more low-frequency words than either DS or TD children (e.g., when asked to name as many animals as they know, some of their answers were "yak", "chihuahua" "ibex", "newt" [p. 32]). Furthermore, Bellugi et al. (1988) noted some unusual responses that individuals with WS provided on a word-definition task (e.g., "Sad is when someone dies; someone is hurt, like when you cry" [p. 197). These various findings have led to the hypothesis that lexical-semantic processing in WS is impaired or atypical (see Rossen et al., 1996; Bellugi et al., 1988, 1994).

Some of the recent studies challenged the hypotheses that individuals with WS possess abnormal vocabularies. Several studies revealed that, when the conversational context is controlled for, i.e. when the individuals with WS are required to talk about a specific topic, instead of the topics they are specially interested in (e.g., animals), they neither use many unusual words nor do they use more low-frequency words than it would be predicted for their level of general intelligence (Volterra et al., 1996; Jarrold et al., 2000; Stojanovik and van Ewijk, 2008;).

Figurative speech is another interesting area in WS language research. Studies concerning this domain have reported that most of the WS subjects can properly use and interpret metaphors, idioms, similes and other simple forms of figurative language. However, although they demonstrate the ability to understand word meaning and concepts that are relatively concrete, studies have found that the vast majority of them present difficulties with understanding more complex forms of figurative language, such as the words with multiple, ambiguous or abstract meanings (Annaz et al., 2009; Mervis et al., 2003).

Last but not least, it is important to mention investigations regarding pragmatic abilities in WS. Initially, researchers noted that individuals with WS are "loquacious" (Arnim & Engel, 1964), "highly social" (Reilly et al., 1990), that their communicative skills are "impressive" (Karmiloff-Smith, 1992), and that they possess "relative strength in expressive language" (Morris et al., 1988). However, more recent studies have indicated that the remarkable expressiveness of WS population can hide many deficits in their pragmatics (see Jones et al., 2000; Laws & Bishop, 2004; Stojanovik, 2006).

The expressive language of individuals with WS is usually examined via story telling tasks and interviews. Story telling tasks revealed that, unlike DS and TD individuals, individuals with WS use a lot of evaluation devices (i.e., 'audience hookers', e.g., "Guess what happened next?", "Suddenly", "Lo and behold") to engage their listeners (Rossen et al., 1996, p. 373). In addition, their narratives are usually long and descriptive, although less coherent and less complex than the narratives of their TD matches (Gonçalves et al., 2011). Interviews, on the other hand, discovered, more vulnerability in the pragmatics of WS population. For example, Laws and Bishop (2004) showed that individuals with WS use significantly more stereotyped phrases in communication than children with DS or SLI. Stojanovik (2006) identified the lack of proper interaction in communication in WS subjects. She reported that children with WS often have difficulties responding appropriately to interlocutor's questions, providing or requesting relevant information, and clarifying their responses, if needed. She also noted that they sometimes even have trouble staying on a topic of conversation. However, they also have an overwhelming need for social interaction, so they will do anything to keep the conversation going (Jones et al., 2000).

## 4.4 Summary

It is evident from the above-reviewed studies that there is a lack of consensus in the literature findings on language advantage over other cognitive domains in individuals with WS. We can also see an inconsistency in data on strengths and weaknesses within the different language domains in WS population. The exact basis for these discrepancies is yet to be found. However, it seems apparent that the use of different instruments for measurement and evaluation of language skills in WS can deliver different results. The differences might depend on the degree of cognitive demands that the chosen instrument places on WS subjects. For instance, we saw that the standardized vocabulary tests used in the earlier studies on WS were more demanding than the ones used in more recent studies.

Furthermore, the inconsistencies in data from the within-language studies can suggest the linguistic heterogeneity of WS population. In such case, every endeavor to characterize WS language profile as one homogeneous linguistic profile might be questionable (see Stojanovik et al., 2006). In addition, since all the language studies reviewed in this chapter include the investigations on English speaking individuals with WS, cross-linguistic studies would be useful in providing more information from each language domain. For example, languages with the high degree of inflections might reveal more precise information about WS strengths and weaknesses in the domain of morphology. Nevertheless, several conclusions about language in WS can be drawn from this review. First, the studies conducted on younger children with WS confirm that the language acquisition in WS population is delayed, relative to typical language acquisition. Second, the investigations of WS syntactic abilities imply that individuals with WS, even though significantly better in this domain than other mentally disabled groups, probably do not exceed the language development of a 7-year-old child. Third, the investigations (using developmental trajectory approach) of WS morphological skills indicate that the development of morphology also plateaus at the age of 7. Since the developmental trajectory of TD individuals shows that the normal development of irregular morphology continues even after that age, the hypothesis that WS is evidence for the existence of the two separate mechanisms of language faculty is arguable. Fourth, the investigations of WS vocabulary skills show us that the WS child's performance differs from the WS adult performance in these language domains. Namely, the performance of individuals with WS in these domains is much poorer in childhood than in adulthood, when compared to the performance of individuals with DS. This again shows how important it is to chart full developmental trajectories in order to properly analyze atypical language development. Fifth, the studies of prosodic skills in WS indicate that the language of WS in not only delayed but also follows an atypical developmental path. Lastly, the investigations of WS pragmatic abilities suggest that the remarkable expressiveness and enormous social drive of individuals with WS conceal many deficits in their language.

In the following chapter, I will further investigate whether WS children follow the same language development as normal children. I will do so by discussing the findings from different language studies on individuals with WS and comparing them to the data from the literature on typical language development. Since covering every aspect of language would exceed the scope of this paper, I decided to focus on vocabulary acquisition. I chose this aspect of language specifically because the development of vocabulary can be followed from the very early age.

# 5 The onset of vocabulary acquisition

Although it seems that children learn to speak very quickly and almost effortlessly, learning language is not an easy task. In the past, linguists used to place the onset of language at about twelve months, when children start to produce their first identifiable words. Innovative research techniques, however, show that language roots can be found much earlier than that. In fact, language acquisition is said to start in mother's womb, continues throughout childhood, and lasts even beyond adolescence (e.g. see Karmiloff-Smith, 2001). During this long journey of language acquisition, the vast majority of children successfully deal with various challenges. But what happens if the infant brain processing is affected by a gene abnormality? Can language stay relatively spared even though the general cognition is severely impaired due to a gene mutation? In the previous chapter, we saw that the language of individuals with WS is hypothesized to be one such example. The linguistic performance of WS individuals on various vocabulary tests is significantly better than their mental age would predict. But does this necessarily mean that individuals with WS follow normal language development from its earliest milestones? In order to investigate this, I will compare the research findings on early vocabulary acquisition of children with WS to the research findings on early vocabulary acquisition of TD children. Ideally, the comparison (between WS individuals and TD individuals) should begin with the studies that have been done on very young infants. However, researchers have been unable to test young infants with WS due to the medical problems they face in their early months of life. Therefore, this comparison will begin with studies that investigated infants at approximately 10 months of age. First, I will discuss how the pre-linguistic behaviors of infants with WS differ from the pre-linguistic behaviors of TD infants. In section 5.2, I will talk about lexical constraints which help TD children recognize a word meaning, and then show how these lexical constraints function in children with WS. Lastly, in section 5.3, I will compare the occurrence of vocabulary spurt in TD children and children with WS.

# 5.1 **Pre-linguistic skills**

Vocabulary acquisition is one of the first challenges every child faces. According to the social-pragmatic approach to language acquisition, "the process of word learning is constrained by the child's general understanding of what is going on in the situation in which she hears a new word" (Tomasello & Akhtar, 2000, p. 5). In other words, in order to build vocabulary, the infant must find a way to discern what the utterances she hears are referring to. To do so, she has to be able to analyze the linguistic input, but also to make use of the non-linguistic cues which indicate to objects or actions around her. In this complex task of disambiguating meaning, any clues available, such as pointing or movement of the specific object into child's focus of attention, is helpful. From the social-pragmatic point of view, pre-linguistic skills infants display, such as referential pointing and the establishment of joint attention, are strongly related to the onset of normal lexical development.

#### 5.1.1 Referential pointing

Referential pointing together with babbling is considered to be one of the first signs of infants' communication. Pointing towards an object or an action (that is being referred to in a conversation) can come from either an adult (parent, caregiver, etc.) or from the child. In normal development, the child's ability to understand the referential pointing of others comes prior to her own ability to point. Infants will look at the object being pointed at when they are approximately nine months old, while their own referential pointing will begin by ten months of age (Murphy & Messer, 1977). Typically developing children begin to point at objects in the environment several weeks before they can produce their first words. Pointing towards an object enables the child to draw the adult's attention to it, which in return usually elicits labeling from the adult. This helps the child to comprehend some words before she is able to produce them orally. Referential or declarative pointing, thus, can be explained as a substitute for the infant's ability to name objects at the early stage of language development (Paterson, 2000).

Several studies on WS have shown that, unlike TD infants, infants with WS do not point towards objects or actions before they produce their first words. Mervis and Bertrand (1997) reported that referential pointing in children with WS appears approximately six months after their first spoken word. They also noted that in contrast to TD 9-month-olds, children with WS do not respond to the pointing of others either. Furthermore, Laing et al. (2002) examined whether the lack of pointing in WS children could be caused by the impairments in fine motor skills. They discovered that, despite the lack of pointing, children with WS did exhibit pincer grip. As Butterworth and Morissette's (1996) study demonstrated, TD children use pincer grips before they begin to point. Since this relationship between pincer grip and pointing is not the same for the children with WS, Laing et al. (2002) suggested that the lack of pointing found in children with WS could not be caused by the impairments in fine motor skills. Especially because a pincer grip, per se, requires much more precision than pointing. Thus, the lack of pointing in WS can rather be interpreted as the lack of understanding of non-linguistic referential clues at this early stage of the language development.

#### 5.1.2 Joint attention

Referential pointing is a medium of establishing joint attention between the child and an adult. Plainly speaking, joint attention refers to a situation when a child and adult are focused on the same thing (Ambridge & Lieven, 2011). Baldwin (1995) argued that during this process both of the participants need to be aware that the focus of attention is shared.

#### 5.1.2.1 Dyadic versus triadic interaction

It is very important that we understand a distinction between dyadic and triadic interaction. Dyadic interaction involves the simple eye contact between the child and her interlocutor. On the other hand, triadic interaction, or joint attention, includes triadic coordination between the child, his interlocutor, and an object or an action. Joint attention goes beyond child's initial ability to participate in a face-to-face dyad (Laing et al., 2002). The skills that the child needs to possess in order to be able to coordinate a moment of joint attention are gaze shifting, reaching, and referential pointing with babbling. Normally, these skills that eventually render joint attention emerge during the period of 6 and 18 months of age (Bakeman & Adamson, 1984). Moreover, joint attention can be initiated and responded to by either of the participants of the dyad (Akhtar & Gernsbacher, 2007).

Many investigations on TD children demonstrated that the child's ability to engage in joint attention is related to her subsequent language acquisition. Probably the most famous experimental study of joint attention in word learning is the one conducted by Baldwin (1993). He tested three different-age groups of children (1;2-1;3, 1;4-1;5, 1;6-1;7). All children were first presented with two toys, and then they were given one to play with. The other toy remained with the experimenter. One half of the children from the each group was involved in a *direct labeling* condition, while the other half was involved in a *discrepant labeling* condition. In the *direct labeling* condition, the experimenter looked at the toy the child was playing with and said, e.g., *It's a toma*. In the *discrepant labeling* condition, the

experimenter looked at his own toy when he produced labeling, while the child was still attending his/her own toy. After the initial session, all children were asked which toy was a toma. The children from the direct labeling condition all correctly associated the word toma with the toy they were playing with. The children from the *discrepant labeling* condition responded differently depending on their age. The youngest group made significantly more incorrect associations, while the group aged 1:4-1:5 was at chance. However, the experiment showed that 77% of children aged 1;6-1;7 correctly associated a novel word to a toy even when the toy the experimenter was referring to was not the one they were attending to. Thus, Baldwin reported that TD children are able to interpret other's people intentions by carefully following the direction of their eye gaze. He argued that starting from approximately 16-18 months of age TD children understand enough about joint attention so that it can block their incorrect mapping of a word to its referent. These findings indicate that TD children make use of non-linguistic cues to interpret the referential context of communication. Moreover, a longitudinal study conducted by Carpenter et al. (1998) investigated a large group of TD children (aged 9-15 months) and revealed a positive correlation between the amount of time mothers and their children spent in joint attention engagement and the size of the children's early vocabulary.

On the other hand, children with WS seem to be impaired in establishing joint attention. Laing et al. (2002) tested joint attention skills of 13 toddlers with WS, aged 17-55 months. The results of the experiment showed that the participants with WS were able to engage only in dyadic interaction. Contrary to a control group of TD toddlers matched on MA, toddlers with WS showed the absence of ability to either initiate or to respond to joint attention. Bertrand et al. (1993) conducted a longitudinal study in which they investigated joint attention skills of only one girl with WS. She was observed from the age of 1;8 to 2;8. The study reported that the girl did not start to engage in joint attention until well after the vocabulary spurt. Moreover, they noted that she showed little interest in toys used in the study. Instead of playing with the given toys, she preferred to focus on the experimenter's face, which was consequently a distractor for establishing a triadic interaction. Thus, since toddlers with WS appear to be incapable of engaging in joint attention before they start to produce words, there must be some other way they initially establish lexical reference.

## **5.2.** Lexical Constraints

Although the ability to employ non-verbal cues, such as pointing or joint attention gives toddlers a head start in word learning, these cues may not always be available or may not be enough. If there are no explicit clues, how do children figure out the meaning of a spoken word? Every word can have an indefinite number of possible meanings and "[i]f children had to consider, evaluate, and rule out an unlimited number of hypotheses about each word in order to figure out its meaning, learning word meanings would be hopeless" (Markman, 1990, p. 57). Markman (1990) identifies the problem of vocabulary acquisition as the problem of induction. According to her, children assume that words are used in certain ways. These default assumptions are called lexical constraints. Lexical constraints guide children to learn words by offering them a limited number of hypotheses they should consider as possible word meanings. Markman (1990) suggests three possible constraints on word meanings:

- (1) **The whole object assumption** which leads children to interpret novel terms as labels for objects not parts, substances, or other properties of objects
- (2) **The taxonomic assumption** which leads children to consider labels as referring to objects of like kind, rather than to objects that are thematically related
- (3) **The mutual exclusivity assumption** which leads children to expect each object to have only one label.

(p. 57)

These lexical constraints help children rule out a number of possible assumptions regarding word's meaning. However, they cannot entirely solve the problem of word learning. They are only assumed to be a part of the early word learning mechanism, thus, as children grow lexical constraints fade away or become replaced with some other learning mechanism. Moreover, Markman (1990) does not explicitly say whether she considers these lexical constraints to be innate, although she does refer to them as "possessed knowledge". Thus, it is not exactly clear whether the lexical constraint approach to vocabulary acquisition is a nativist approach.

#### 5.2.1 The whole object assumption

The whole object constraint guides the child to assume that a novel word refers to a whole object rather than to its parts or to the materials it is made of. Thus, when the child hears a word *rabbit* (possibly in the presence of a rabbit) she will infer that this word refers to a rabbit as a whole, not to the rabbit's tail, paws or its fur (cf. Ambridge & Lieven, 2011).

Markman and Wachtel (1988) conducted a study on TD children aged 3;0–4;3 in which they demonstrated that children associate novel words with whole objects and not with the parts of these objects. In the study, children were shown pictures of unfamiliar objects (e.g., *a lung*) and given an unfamiliar label (e.g., *bronchus*). On the request to say "Which one is the bronchus?" the whole thing (experimenter circles the whole lung) or just this part (experimenter points to the bronchus) 80% of the children chose "the whole thing". This is interpreted as evidence that children operate with the whole object constraint during their word learning process.

Stevens and Karmiloff-Smith (1997) replicated and extended the study by Markman and Wachtel (1988). Their aim was to test whether lexical constraints on word learning could be found in WS individuals. The experimental group consisted of 14 WS subjects whose CA ranged from 7;5 to 31;5 (mean 20;1). Their vocabulary MA ranged from 6;8–16;4 (mean 9;11) whereas their non-verbal MA ranged from 3;6–9;0 (mean 6;4). The results of the experimental group were contrasted with the results of four TD control groups (a group of 3-, 5-, 7- and 9-year-olds). In regard to TD control, Stevens and Karmiloff-Smith (1997) reported that the results from all four groups were comparable to the results from the Markman and Wachtel's (1988) study. TD children across all ages from 3 to 9 years seem to be guided by the rule of the whole object constraint. On the other hand, the picture for the WS subjects was very different. Unlike in TD controls, the whole object constraint does not seem to be present in WS subjects. They were randomly choosing both "the whole object" and "a part of the object" options. In other words, WS subjects showed no significant difference in favor of "the whole object" responses. This suggests that, relative to normal development, the vocabulary acquisition of WS individuals is not constrained by the whole object bias.

#### 5.2.2 The taxonomic assumption

After establishing that a label (e.g., *shoe*) refers to a whole object (e.g., *the child's own shoe*) the child has to figure out how to extend it to other objects (e.g., *her mother's shoe*). This is not an easy task. The child must figure out that the label (*shoe*) she learned can refer to other objects 'of the same kind' (e.g., *a high-heeled shoe, a sandal, etc.*). In this endeavor, children are guided by the taxonomic constraint (cf. Ambridge & Lieven, 2011).

In theory, a label might be extended to objects that are related either thematically, where objects are linked by causal, temporal, spatial or some other theme (e.g., *shoes and feet, cows and milk, etc.*), or taxonomically, where objects are linked by the same taxonomic category (e.g., *dogs, cows, and rabbits* all belong to the same taxonomic category of *animals*). However, experimental studies have shown that children favor extending labels to objects that are taxonomically related (cf. Ambridge & Lieven, 2011; Guasti, 2002).

Markman and Hutchinson (1984) tested 4- and 5-year-olds in an experiment in which two groups of children were assigned two different conditions: the no-word condition and the novel-word condition. In both conditions, participants were first shown a target toy (e.g., a police car). Then, two other toys were placed next to the target toy, one on the right and the other one on the left. One toy was thematically related to the target toy (e.g., a policeman), whereas the other was an object that belongs to the same superordinate category as the target toy (e.g., a truck). In the no-word condition, the participants were shown the target toy, and the experimenter asked, "See this?" Then, he showed the two other toys and asked, "Can you find another one?" In the novel word condition, the experimenter pointed to the target toy and labeled it with the nonsense word by saying, e.g., "This is wug". Then he placed the other two toys on the table, and while holding the target toy, he would say, "Can you find another wug that is the same as this?" If labeling stimulates children to choose the object of the same kind, it can be supposed that in the no-word condition children would randomly choose either the thematically or the taxonomically related toy, while in the novel-word condition they would be biased towards the taxonomically related toy. The results from the Markman and Hutchinson's (1984) experiment showed that the children who were assigned the no-word condition made a taxonomic choice only 25% of the time, whereas the children who were assigned the novel-word condition made a taxonomic choice 65% of the time. Since the distinction between the two conditions was the presence versus the absence of a label for the objects, Markman and Hutchinson (1984) concluded that the presence of the novel word invites children to relate objects taxonomically rather than thematically. This further suggests that the process of word learning in TD children is constrained by the taxonomic bias.

Let us now see what studies have found concerning the taxonomic constraint in individuals with WS. Steven and Karmiloff-Smith (1997) conducted a similar experiment as the one of Markman and Hutchinson (1984) mentioned above. They tested 12 WS subjects aged 8;6–30;11 (mean 19,9). Their vocabulary MA was assessed to be in the range 4;5–16;4 (mean: 9;1) while their non-verbal mental age was assessed to be in the range 3;6–8;3 (mean: 5;4). TD controls were divided into two groups: a group of three-year-olds and a group of

nine-year-olds. The results in relation to the TD children confirmed the Markman and Hutchinson's (1984) findings. Namely, both groups of TD controls showed a significant advantage of taxonomically related choices over the thematically related choices in the presence of a novel word, as opposed to the no-word condition. Contrary to TD children, there was no significant difference in the responses of WS subjects between the no-word condition and the novel-word condition. That is, no greater number of taxonomic responses was made in either of the conditions. Therefore, Steven and Karmiloff-Smith (1997) argue that the individuals with WS lack the taxonomic constraint. However, they report that there were a few individual cases of WS subjects who did show a taxonomic bias. The surprising fact was that those subjects did not have better vocabulary scores than the rest of the subjects who did not show a taxonomic bias. This finding indicates that WS individuals can acquire relatively good levels of vocabulary even though their vocabulary development is not guided by the taxonomic constraint.

#### 5.2.3 The mutual exclusivity assumption

We have seen that children initially assume that a novel word refers to the whole object. However, they eventually have to learn words that refer to the part of the objects. As Markman (1990) argues, this is the moment when the whole object constraint must be superseded by the new assumption. Namely, if the child had already learned that the word *rabbit* refers to the rabbit that stands in front of her, she would assume that a novel word uttered in the presence of the rabbit (e.g., *tail*) is a label that refers to some part of the rabbit, not the rabbit itself. This assumption is defined by the mutual exclusivity constraint.

The mutual exclusivity constraint guides children to assume that "each object will have one and only one label" (Markman, 1990, p. 66). The assumption that words are not synonymous may be initially helpful in vocabulary acquisition. However, we know that a single object can be referred to both with its own label and with the label of the categories it belongs to (e.g., a parrot is also a pet, an animal, and a bird). For this reason, at one point of their vocabulary acquisition, children must supersede the mutual exclusivity constraint as well.

As it was mentioned earlier, the constraints Markman (1990) proposes cannot entirely solve the induction problem of vocabulary acquisition. The proposed constraints are not absolute prohibitions. They are only tendencies of children to favor certain choices at the very start of their vocabulary acquisition. In other words, they only help children limit the indefinite number of hypotheses for possible word meanings that is imposed on them at the onset of their vocabulary development. Other than that, the scope of these constraints is very restricted. For example, they cannot clarify how children acquire the meaning of abstract objects, verbs, and prepositions (cf. Ambridge & Lieven, 2011; Guasti, 2002).

Let us now return to the mutual exclusivity constraint to see how TD children deal with it in practice. Similarly to the above-mentioned experiment on the whole object constraint, Markman and Wachtel (1988) conducted the study on 3- and 4-year-olds to investigate the influence of the mutual exclusivity constraint. This time, however, the participants were not presented with the picture of an unfamiliar object. Instead, they were first taught the name for the unfamiliar object in the picture (e.g., *lung*) which was later used in the experiment. Thus, in the initial training, the experimenter gave a short presentation to each child by saying, e.g. "This is a lung. People have two lungs in their body, and they use them for breathing." The rest of the study followed the same procedure as the whole object constraint study. Children were presented with the picture from the training, and the experimenter asked about, e.g. "Which one is a *bronchus*? The whole thing [experimenter circles whole lung] or just this part [points to the bronchus]." The results showed that children chose "just this part" in 85% of the time. Therefore, the evidence shows that children successfully override the whole object constraint when a novel word is uttered in the presence of a familiar object.

Steven and Karmiloff-Smith (1997) investigated the mutual exclusivity constraint in WS population. Their study included 10 WS subjects who were 8;0–30;5 years of age (mean: 19;6). The vocabulary MA of these subjects was in the range 6;8–16;4 (mean: 9;9) whereas their non-verbal MA was between 5;3 and 11;0 (mean: 7;6). The results were contrasted with the two groups of TD controls: a younger group of three-year-olds and the older group of nine-year-olds. The procedure of the experiment was the same as in the Markman and Wachtel's (1988) study. The study demonstrated that, like the TD controls, the WS group successfully excluded the possibility that the familiar object (for which they already have a label) can have another label and therefore demonstrated that they are sensitive to mutual exclusivity constraint. These results are somewhat surprising. We have seen earlier in this chapter that WS individuals are not sensitive to either the whole object constraint or to the taxonomic constraint. However, the mutual exclusivity constraint seems to be present in their word learning process in the same way as it is in the TD controls.

#### 5.3 Vocabulary Spurt

Most children start to produce first words by their first birthday. At that age, their expressive vocabulary usually consists of three to four different words. From then on, the number of new words in their vocabulary slowly grows until they reach about 75-150 words. At this point, there is a sudden increase in the rate at which new words are acquired. This occurrence is characterized as the vocabulary spurt (cf. Ambridge & Lieven, 2011).

Gopnik and Maltzoff (1987) noted that the explosion in vocabulary acquisition occurs when the child is about 18 months old. They also argued that this experience coincides with the development of certain cognitive abilities, such as spontaneous exhaustive sorting. Spontaneous exhaustive sorting refers to the child's ability to sort the objects that belong to a particular category. Furthermore, Mervis and Bertrand (1994) argue that the vocabulary spurt is strongly related to children's fast mapping ability. Fast mapping means that the child is able to map novel words to objects for which she still does not have a name. For example, if three objects lie on the table, two familiar and one unfamiliar, the child will pick up the unfamiliar one on hearing a novel name. This realization that all objects should have a name is called Novel Name - Nameless Category (N3C) principle. Before children start to use this principle, they can only rely on other people who assist them in mapping by explicitly providing a connection between a new word and its referent. For instance, pointing to an object and labeling it at the same time or labeling an object that the child is already attending to. However, when children acquire the N3C principle they no longer need to rely on adults to provide an explicit connection between a label and its referent. In other words, even if the direct connection is not given, the child is still able to map a novel label to an object that is present, but for which she does not yet have a name.

Gopnik and Meltzoff, (1987) found that 16-20 month children who were able to sort various objects into categories could also use the N3C principle, whereas the ones who could not sort spontaneously had not yet begun to fast map. They explained that these two abilities (spontaneous exhaustive sorting and fast mapping) occur at the same time because they reflect parallel insights, one insight being cognitive (all objects belong to some category) and the other insight being linguistic (all objects have a name). Finally, it seems that when these to abilities meet the vocabulary starts to grow rapidly.

In contrast to typical development, children with WS do not produce first words before the age of 2. However, the vocabulary spurt in WS children often appears long before their ability to sort objects into categories. In fact, it was found that the onset of the

vocabulary spurt in WS children occurs 6–12 months prior to the emergence of exhaustive sorting (Paterson, 2000). Mervis et al., 1997, reported that some WS children had a vocabulary size of more than 500 words before they displayed the ability to spontaneously sort objects into categories. Despite these discrepancies, Steven and Karmiloff-Smith (1997) demonstrated that once WS children began to use spontaneous exhaustive sorting, they were also capable of fast mapping, which demonstrates that these two abilities (one cognitive and one linguistic) are indeed closely related.

As it appears, children with WS learn word labels before understanding how to map those labels to objects. This suggests that WS individuals might be memorizing the phonological sequences of words while their referential understanding of those words might be very limited. In other words, they might be focusing less on semantics and more on phonology in their early vocabulary development, in contrast to TD children (Mervis et al., 1997).

#### 5.4 Summary

The comparison of the literature on vocabulary acquisition presented above points to some clear differences in the way WS and TD language develops in the early stages. We saw that the social-interaction skills (such as referential pointing and the establishment of joint attention) play an important role in helping TD infants make associations between social referencing and language. These skills do not seem to function in the same way in WS infants. Referential pointing, which TD infants use to request labeling for things around them, appears quite late in the WS development. In fact, it appears after the onset of their language production, which is not the case in normal development. Since the possibility that the lack of pointing is caused by fine motor impairments is excluded, we are left to speculate whether or not WS infants comprehend non-linguistic referential clues at this early stage of the language development. Another useful pre-linguistic behavior that typically helps children associate words with their meanings is the ability to establish joint attention. Although WS individuals are described as very sociable, this characteristic might have some negative influence on their language development. WS individuals tend to be obsessed with people's faces which usually draws their attention away from other things happening around them. In this way, they fail to engage in joint attention which is, according to social-pragmatic view, very important for the

normal onset of vocabulary development since it can give initial clues for the correlation between words and their references. It is possible that the lack of pre-linguistic skills slows down the language acquisition of WS individuals which would explain the delay in the onset. However, the insensitivity of older children and adults with WS to lexical constraints indicates that WS individuals continue to follow a different path in vocabulary acquisition. If we assume that lexical constraints are the part of the innate linguistic knowledge, or as Markman (1990) describes them "possessed knowledge", we can see that WS individuals do not rely much on that knowledge in their vocabulary acquisition. The above-reviewed studies show that while all TD children between 3 and 9 years of age display fast mapping and make use of the whole object, the taxonomic and the mutual exclusivity constraints, WS individuals are only sensitive to mutual exclusivity and fast mapping. Are these two biases enough for normal language development? Further research is needed to discover that.

Taking both pre-linguistic behaviors and lexical constraints into consideration, it is clear that TD children acquire words only after they establish some kind of semantic relation between labels they hear and objects that exist in their environment. If WS children fail to do so, how can we explain their remarkable scores on vocabulary tests in adolescence and adulthood? Some studies indicate that people with WS rely more on phonology than on semantics when they learn new words (see Grant et al., 1997; Karmiloff-Smith et al., 1997; Laing et al., 2001). Moreover, many studies point to the fact that lexico-semantic processing in WS population is aberrant (Bellugi et al., 1988, 1994; Rossen et al., 1996; Udwin et al., 1987; Volterra et al., 1996; Neville, Mills, & Bellugi, 1994; Neville, Holcomb, & Mills, 1989). In the next chapter, I will first review some of these studies. Then, I will introduce a study that suggested that, even though the semantic processing in WS individuals is organized in the same way as in TD individuals (Tyler et al., 1997). Lastly, I will discuss what such findings might say about the connection between language and the rest of the cognition.

## 6. **Production vs. Comprehension**

Although significantly delayed and very poor at the beginning, once the vocabulary of individuals with WS starts to develop it gradually improves. By the time the individuals with WS reach adulthood, they have built up an impressive size of vocabulary. Adult WS language is often characterized as being filled with elaborate and "erudite-sounding" vocabulary items, which is not apparent in other mentally challenged groups (Reilly et al., 1990; Volterra et al., 1996). Here is what a person with WS (age 32) replied when asked what he would want other people to know about WS.

"We are respectable, loving, understanding people. With pride, and dignity, and grace. And people should understand that Williams Syndrome people don't really care about the bad things in life and what goes on in the world because we as people have enough of problems of our own, living day to day... First of all I am a human being, and I am a man, and I am an older adult. And I want people to realize that I do have feelings, I'm not a freak... So, respectability is a main factor in learning how to deal with people with Williams Syndrome or any other syndrome."

(Bellugi et al., 2001, xiv)

This individual with WS evidently has quite large and varied vocabulary. Researchers often describe people with WS as eloquent and verbose. However, do they understand everything they say? Is everything they say semantically structured and processed in the same way as for people classified as TD? Many investigations claimed that although WS individuals show remarkable vocabulary skills in adolescence and adulthood, their lexical semantics are not without anomalies. The experimental works of Bellugi et al. (1988) and Rossen et al. (1996) showed that the choice of words WS people use is often unusual, particularly because they tend to use more low-frequency words. They also reported that word-definition and speeded naming tasks reveal certain lexico-semantic deficits in WS language. Another work that presented evidence for aberrant semantic processing in WS is the work by Neville and collaborators (Neville, Mills, & Bellugi, 1994; Neville, Holcomb, & Mills, 1989). They measured neuropsychological correlates to semantic processing by using Event Related Potentials (ERPs) and found different temporal patterns in the brain activity of WS subjects in contrast to TD controls. However, despite the claims that semantic processing in WS is abnormal, Tyler et al. (1997) argued that the semantic organization of WS lexicon is normal. They conducted a study in which they measured semantic priming effects in word-word associations and found no significant difference between the results of WS subjects and TD

controls. In this chapter, I will first look into all these findings and then discuss how they might be related to the debate about cognitive dissociations.

#### 6.1 Off-line experiments of WS language comprehension

#### 6.1.1 Word-definition

The first clue that the WS lexical semantic processing is aberrant comes from experiments using a word-definition task. The word-definition task investigates the ability of a subject to explain the meaning of a given word. The responses in this task are scored for accuracy and completeness. The highest scores are received if the subject provides a precise definition in which the most salient features of the word are included, while the responses that include the superordinate categories or some descriptive characteristics of the word are scored lower.

Bellugi et al. (1988) tested the comprehension of three adolescents with WS (Van - age 11, IQ 50; Cristal – age 15, IQ 49; Ben – age 16, IQ 54) on the Oral Vocabulary subtest of the Test of Language Development in which the subject are required to give oral definitions for 20 common English words. Here are their responses when asked what the word *sad* means.

Van:	"Sad is when someone dies; someone is hurt, like when you cry."
Crystal:	"Sad means that someone hurts your feelings, or when someone starts
	crying."
Ben:	"When you lost somebody that you love and care about. It means
	something happens to you like your grandmother died or some part of
	your family or your cousin."

(Bellugi et al., 1988, p. 197)

It is evident that their definitions tend to be lengthy and descriptive, but they lack the most salient feature of the word *sad*. The precise definition of the word *sad* should include '*it is a* (*type of*) feeling'. Here is another definition that illustrates only a very distant association to the target word in the form of a rather loose description.

Experimenter: "What does *poor* mean?" Crystal: "Means they are hungry; they are dying from exposure."

(Bellugi et al., 1988, p. 197)

Generally, WS individuals score very low on word-definition tasks. However, Bellugi et al. (1988) reported that in a spontaneous speech WS individuals are able to use all the words from the test correctly. Bellugi et al. (1992) emphasized that word-definition tasks include metalinguistic demands that WS people are unable to fulfill. We will see in section 6.2.2 that a similar observation was made by Tyler et al. (1997).

#### 6.1.2 Speeded naming and spontaneous speech

The second clue to possible aberrant semantic processing in WS comes from the speeded naming tasks. In speeded naming tasks, the subject needs to provide as many words as possible on a requested topic within a limited period of time. The speed at which names of items can be accessed provides information about the semantic abilities of the subject. Furthermore, this task can also provide the insight of how the mental lexicon of the subject is organized.

Bellugi et al. (1988) administered the Clinical Evaluation of Language Functions -Producing Word Associations test. In separate trials, the subjects were asked to name as many animal names as they could within 60 seconds. The results were evaluated quantitatively (i.e., by observing a total number of different words produced) and qualitatively (i.e., by investigating the number of subcategories within each semantic category and the number of shifts between them). The following are examples of the responses from the WS subjects for animal names:

- Van: Sea lion, zebra, hippopotamus, lizard, beaver, kangaroo, chihuahua, crocodile, tiger, owl, turtle, reptile, frog, giraffe
- Crystal: Koala bear, antelope, moose, anteater, lion, tiger, rat, bear, giraffe, elephant
- Ben: Buffalo, leopard, sabretooth tiger, condor, vulture, turtle, bear, snake, giraffe, lion, bull, dog, cat, tiger

(Bellugi et al., 1988, p. 198)

It is noticeable that the subjects did not have difficulties accessing numerous members of a given semantic category. Thus, quantitatively WS subjects do not present any semantic deficits. However, qualitatively their semantic organization appears to be unusual. Bellugi et al. (1988) noted that their responses were surprising because they did not only provide general subclasses of items but also very specific, rare and nonprototypical animals. Similar tendency to use unusual words was observed in their spontaneous speech. For example:

"The bees *abort* the beehive", meaning: "The bees *leave* the beehive"

(Rossen et al., 1996, p. 384)

"I'll put the earrings and you can *buckle* them", meaning: "...you can *fasten* them"

(Bellugi et al. 1994, p. 32)

Rossen et al. (1996) interpreted these unusual word choices of WS individuals as a clue for lexico-semantic anomalies. Contrary to this interpretation, Semel and Rosner, (2003) argued that such choices might be slightly off the mark semantically, but again not inappropriate in terms of context.

#### 6.1.3 Homonyms

A third indication that WS semantic processing is atypical emerges from the responses of WS individuals to tasks that include homonyms. Homonyms are words that are spelled or pronounced the same but have different meanings. Each homonym (e.g., *bank*) has at least two meanings: one more frequently used, primary meaning (associated with *money*) and the other less frequently used, secondary meaning (associated with *river*). In a situation where the meaning of a homonym is not contextualized most people tend to interpret it by choosing its primary meaning.

Rossen et al. (1996) used three different tasks to assess the WS comprehension of homonyms: free association, similarity judgment, and definitions. In the free association task, the homonym was read to the subjects, and then they were asked to say the first word that comes to their mind. In the similarity judgment task, the subjects were given a word triad (e.g., BANK-RIVER-MONEY) and then they were asked to "pick two that go best together". In the definition task, the subjects were simply asked to say everything they know about the meaning of the target homonym. The results showed that WS subjects performed much like the TD controls in the free association task. They associated the target homonym with its primary meaning more frequently. By contrast, in the other two tasks, WS subjects were using the secondary meaning more often than the TD controls. While TD controls demonstrated a clear preference for primary meanings, WS subjects chose randomly between primary and secondary meanings. Rossen et al. (1996) concluded that individuals with WS are less sensitive to word frequency than TD individuals, which again implies that their semantic processing is aberrant. He further suggested that this aberrant semantic processing is probably caused by an abnormal lexical organization.

#### 6.2 On-line experiments of WS language comprehension

#### **6.2.1 Event-Related Potentials**

The first experimental study which directly measured brain activity during the semantic processing of WS individuals was conducted by Neville and collaborators (Neville, Mills, & Bellugi, 1994; Neville, Holcomb, & Mills, 1989). They investigated the processing of semantic information in sentences (in people with WS) by looking at Event-Related Potentials (ERPs). This study differed greatly from the previously-reviewed behavioral (off-line) investigations because it directly measures real-time language processing by recording changes in the temporal patterns of brain activity. Before I present this investigation, I will briefly introduce some basic facts about the nature of this kind of research.

Human brains store enormous amounts of various kinds of information about the world. In the normal brain, these different pieces of information are rapidly retrieved from the 'semantic memory' as a reaction to a linguistic signal, such as a spoken, written or signed word. What is fascinating is the fact that the brain of a TD language user is able to perform this action in only hundreds of milliseconds. This is approximately the amount of time the brain needs to make sense of a word heard in an ongoing discourse (Kutas & Federmeier, 2000). When we observe the linguistic behavior of TD people, retrieving information from the semantic memory seems to be an effortless process. However, if we look at semantic processing from the brain perspective, we realize that it is actually achieved by complex neural mechanisms.

Uncovering the underlying brain mechanisms that enable semantic processing is essential for a better understanding of human cognition, and thus for language faculty itself. Off-line studies, like the ones mentioned above, analyze different language behaviors of people and then evaluate whether the semantic information is processed normally or not during language comprehension. However, such studies cannot explain how the brain constructs meaning and how this meaning is retrieved during language comprehension. Also, they cannot explain how fast the process of retrieving semantic information is, which is again an important fact in evaluating whether the process itself is abnormal or not.

Neuroimaging techniques can provide us with the information on brain activity during semantic processing. Hemodynamic-based neuroimaging methods, such as Functional magnetic resonance imaging (fMRI) and positron emission tomography (PET) imaging, provide excellent spatial resolution. Hence, they can show the location of a brain activity during semantic processing with certain precision. However, their temporal resolution is

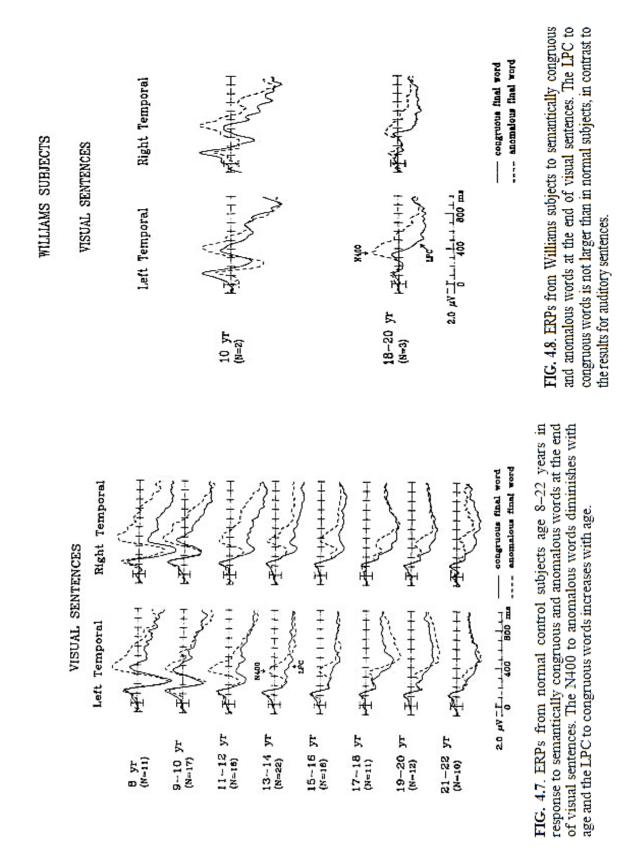
limited because they cannot capture events occurring faster than a second or two. Since the semantic processing, like any other brain processing, is happening within milliseconds, fMRI and PET imaging are not capable of capturing it. Electroencephalography (EEG), however, provides a very high temporal resolution which can capture events occurring within milliseconds (cf. Dale & Sereno, 1993; Kutas & Dale, 1997). Therefore, EEG studies can illustrate the temporal changes during the brain processing (in this case semantic processing), and they do so in the form of event-related potentials (ERPs). ERP is the recorded measure of brain response to some sensory, cognitive or motor event. (Kutas & Federmeier, 2000).

In 1980, Kutas and Hillyard used the EEG method to look at semantic processing of TD individuals. They monitored the electrophysiological response to a semantically anomalous word (e.g., pencil) in a sentence context (e.g., He shaved off his mustache and pencil). The participants in the experiment were requested to silently read 160 seven-word sentences presented one word at a time. A random 25% of those 160 sentences ended in a semantically inappropriate, but syntactically correct word. They discovered that the unexpected sentence ending elicits a negative-going brain wave that peaks around 400 msec (N400). The N400 is an electrophysiological brain component of "time-locked EEG signals" referred to as ERPs (Kutas & Federmeier, 2000). In the same study, Kutas and Hillyard (1980) found out that the N400 was not caused by any type of violation at the end of a sentence. For instance, a semantically expected but physically unexpected word (because in capital letters) at the end of a sentence (e.g., He shaved off his mustache and BEARD) elicited a positive-going brain wave that peaks around 300 msec (P300). Moreover, more recent investigations showed that a syntactic violation (e.g., He shaved off his mustache and swimming) elicits a positive component P600 (see Coulson, King, & Kutas, 1998). These different findings indicate that the N400 is particularly related to semantic processing, and is not simply a response to any unexpected stimulus. Hence, neurolinguists suggest that the N400 provides a window into our knowledge of the semantics of words.

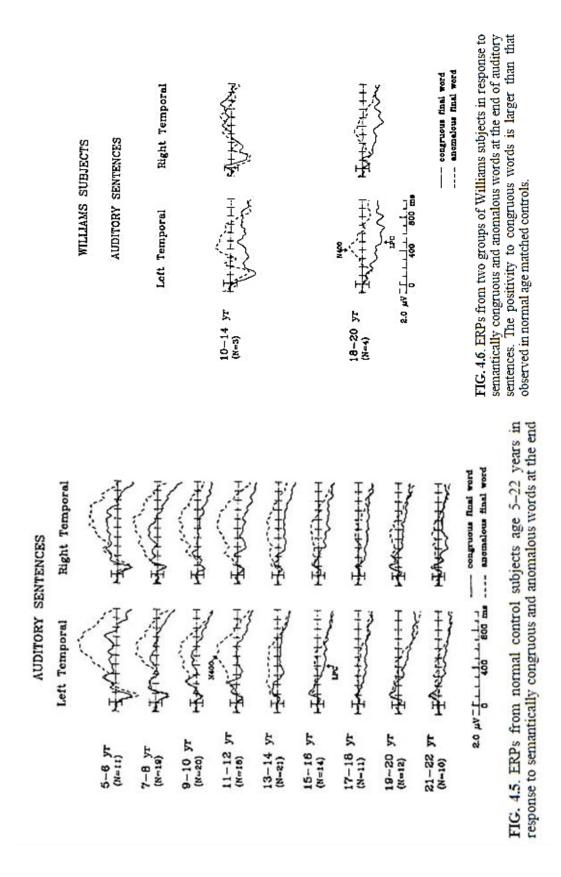
Now that we have an idea of what ERP studies are and how they are used to measure brain activity, let us return to the investigations by Neville and collaborators (Neville, Mills, & Bellugi, 1994; Neville, Holcomb, & Mills, 1989). Neville, Mills, and Bellugi (1994) replicated and modified the Kutas and Hillyard's (1980) experiment. They observed two small groups of subjects with WS: one group of 4 children (10-14 year olds) and one group of 4 adults (18-20 year olds). Their ERPs were compared to the ERPs of TD subjects (5-22 year olds). The participants were requested to read or listened to sentences presented one word at a time at their own pace that ended in either a highly semantically probable way (e.g., *I have* 

five fingers on my hand) or an anomalous way (e.g., I have five fingers on my water). In contrast to Kutas and Hillyard (1980), Neville, Mills, and Bellugi (1994) tested brain reactions on both visual and auditory stimuli (see the figures below). In the visual modality, the TD subjects showed a gradual decrease of the amplitude of N400 to unexpected words and a loss of N400 to expected words with increasing age. The WS subjects displayed similar responses to visual stimuli. However, the pattern of the ERPs to auditory stimuli in WS individuals was different from the one found in TD controls. In TD subjects, the unexpected auditory word at the end of a sentence elicited a large negativity response (N400) and a loss of N400 to expected words by 15 years, when ERPs are characterized by a late positive component (LPC). WS subjects displayed unusually enhanced positivity to expected sentence endings than TD subjects of corresponding ages. For auditory stimulus, results from WS subjects showed a difference in timing of the activation of the brain response, as well as, the difference in the distribution of reactions across the brain regions, relative to TD controls. In TD controls, the N400 effect was larger from the right than from the left hemisphere for both auditory and visual stimulus exposure, and this asymmetry was increased with age. The WS subjects did not show this pattern of asymmetry in the auditory modality. In WS subjects, the N400 effect was larger over the left hemisphere. (Neville, Mills, & Bellugi 1994).

We can see that ERPs can reveal both the timing and location of semantic processing during language comprehension. The results of Neville, Mills, and Bellugi's (1994) study show that both the timing and the location of semantic processing in WS population display certain abnormalities. Moreover, other researchers who used ERPs for the investigation of WS language comprehension have presented comparable findings to the ones I reviewed here (e.g., see George, Milles, & Bellugi, 2000; Mills et al., 2003; Fisherman et al., 2010). Therefore, it can be concluded that the findings from the on-line studies based on ERPs are in line with the hypothesis made based on the off-line studies mentioned in previous sections.



(Taken from Neville, Mills, & Bellugi, 1994, p. 93-94)



(Taken from Neville, Mills, & Bellugi, 1994, p. 90-91)

#### 6.2.2 Semantic priming

All hitherto mentioned studies point to an abnormality in the semantic processing of the WS population. Rossen et al. (1996) argued that the malfunction in WS semantic processing is probably caused by an abnormal lexical organization. Tyler et al. (1997) conducted an on-line study to investigate whether such argument can be confirmed by looking at the directly measured effects of semantic priming.

Semantic priming refers to an occurrence when the recognition of a word is speeded by the prior presentation of a semantically related word. This event is assumed to be driven by the semantic overlap between prime and target word. For example, it has been demonstrated that when a person is presented with the word *cat* (prime word) before being exposed to a word *dog* (target word), he/she needs less time to recognize that the word *dog* is an actual English word because the two words are semantically related. In other words, the meanings of these two words are referencing each other in some way. However, if the prime word (e.g., *cat*) is semantically unrelated to the target word (e.g., *vase*) a person needs more time to recognize the target word (cf. Meyer & Schvaneveldt, 1971). Hence, priming can influence word recognition in two different ways. It can speed up target word processing which is called *facilitation*, or it can slow it down which is referred to as *inhibition* (Gulan & Valerjev, 2010).

How can such an occurrence be explained? I mentioned in the previous section that human brain is retrieving lexical information very rapidly. As an explanation for this, linguists assume that all the different lexical information that we are storing in our brains are semantically organized. This further suggests that that the retrieval of semantically related words in succession should be faster than the retrieval of semantically unrelated words. Of course, faster triggering of a word recognition can be provoked by many other connections as well. For instance, words that sound similar but don't have any semantic relation to each other (e.g. *cheese* and *please*) can trigger faster recognition of one another by phonetic association. This is called phonetic priming. Possibilities for priming word recognition are numerous, however, for the sake of the Tyler et al.'s (1997) experiment, I will stay within the frame of semantic priming.

Semantic priming is considered to be a very reliable method for investigating semantic organization in the mental lexicon because it illustrates automatic activation of semantic information. This is the reason Tyler et al. chose this method for their experiment. Their aim was to avoid placing any extra cognitive demands on WS subjects. As they

explained, most of the studies that investigated language comprehension of WS individuals used explicit tasks, where subjects have to access semantic information in a controlled, explicit manner. Such explicit tasks include sorting, matching, abstract thinking, etc. All these things require cognitive involvement that goes beyond "purely linguistic knowledge" (cf. Bellugi et al., 1992; Tyler et al., 1992; for further discussion see chapter 7). In contrast, the task that Tyler et al. (1997) used examined semantic knowledge implicitly. The subjects were not required to explicitly retrieve the meanings of the words they heard, i.e., no overt responses were requested from them. According to Tyler et al. (1997) implicit tasks, such as semantic priming, can "reveal sparing of semantic knowledge which, on the basis of explicit tasks, is assumed to be inaccessible or lost" (p. 518).

The semantic priming experiment that Tyler et al. (1997) conducted included 12 WS subjects aged 14;1–30;5. Their IQs ranged from 45-87, and their vocabulary MA ranged from 7;1–16;4. Their results were compared to 20 TD subjects who were slightly older in CA, ranging in age 18-40 years. The study examined two aspects of semantic organization: category structure and functional attributes. Category structure examines whether WS lexicon has an organized structure of taxonomically related words (e.g., *desk-chair*), whereas functional attributes examine whether WS individuals associate different thematically related concepts (e.g., *baby-crib*) in the same way as TD individuals (cf. section 5.2.2).

The participants were tested using a primed monitoring task, in which they were requested to press a reaction time button as soon as they heard a target word which was prespecified by the examiner. Each hearing trial involved listening to a list of words played with a constant interval between each spoken word. The lists of words were short, varying from 5-10 words. The target word was always the last word on the list. In one version of the experiment the target word (e.g., *umbrella*) was immediately preceded by the related prime word (e.g., *rain*), while in the other version it was immediately preceded by the unrelated control word (e.g., *essay*). The rest of the words on the lists (filler words) preceding the prime-target pair were all semantically unrelated to the prime-target pair. Also, none of them started or ended with the same phonological sequences as the target word so that phonological priming could be avoided (see an example of these lists below).

		Functional pair		
hen-farm				
related:	larder	10	nrelated:	larder
	sun			sun
	health			health
	essay			essay
	hen			clam
	FARM			FARM
	chisel			chisel
sweater-1	warm			
related:	ghost	10	nrelated:	ghost
	racket			racket
	sweater			pliers
	WARM			WARM
	punch			punch
		Category pair		
mouse-ha				
related:	bed	10	nrelated:	bed
	sugar			sugar
	brick			brick
	vivid			vivid
	half			half
	proceed			proceed
	mouse			clown
	HAMSTER			HAMSTER
	foot			foot

TABLE II Examples of the Materials

(Taken from Tyler et al., 1997, p. 521)

Before each hearing trial, the participants were told the target word they were supposed to monitor for. In order to measure the time participants need to recognize a target word, a timing pulse was set at the onset of each target word, and it was stopped when the subject pressed the response button. If the participant's reaction times to detect target words were faster when they were preceded by the related prime word than when preceded by the unrelated control word, then the priming effect is displayed.

Tyler et al. (1997) reported that the reaction times of WS subjects for semantically related pairs were only slightly slower than the ones of TD controls, which was statistically insignificant. Similar to those the of TD controls, the monitoring latencies of the WS subjects were significantly faster in the semantically related condition than in the semantically unrelated condition. Moreover, both taxonomically and thematically related word-pairs primed strongly in WS individuals (see the table below). This indicates that WS individuals are able to retrieve various kinds of semantic information rapidly when they hear a word, and then to use this information to facilitate subsequent words which are related in different ways.

Finally, contrary to Rossen et al.'s (1996) proposal, Tyler et al. (1997) found no evidence that the lexical organization of WS individuals is impaired.

	Related	Unrelated	Difference
Controls	011015		
Overall	365	437	72
Functional	370	445	75
Category	361	428	67
ws			
Overall	513	582	69
Functional	500	564	65
Category	527	600	73

#### Semantic organization in Williams syndrome

TABLE III Mean Monitoring Latencies (ms) for the Controls and WS Subjects

(Taken from Tyler et al., 1997, p. 523)

#### 6.3 Summary

Around the age of two, individuals with WS start to produce their first words. In time, their vocabulary increases and already in adolescence their vocabulary knowledge is reported to be much better than one could predict judging by the level of their IQs. This was not found in any other intellectually challenged population. In everyday speech, WS adults display erudite-sounding vocabulary. However, researchers surprisingly varied and are still questioning whether they understand all the words they use. Findings from several investigations suggested that the semantic processing of WS people is abnormal (Bellugi et al., 1988; Bellugi et al., 1994; Rossen et al., 1996). In three of these investigations, researchers used off-line experiments to probe the vocabulary comprehension of WS individuals. The first experiment tested their ability to give oral definitions of 20 common English words. The second experiment investigated their semantic organization via the speeded naming task. And the third experiment explored their comprehension of homonyms. Off-line experiments showed that individuals with WS have difficulties in precisely defining very common words, although they are able to correctly use them in their spontaneous

speech. They also demonstrated that WS people are capable of naming numerous words that belong to the same category in limited time. However, these numerous words that belong to the same category are often unusually organized in a sense that various basic subordinates are mixed with rare and nonprototypical category members. Moreover, the off-line experiments also showed that WS individuals are prone to insert some low-frequency words in their sentences which sometimes might seem semantically inappropriate. Their insensitivity to word frequency was also displayed in the experiment where the comprehension of homonyms was tested. Besides these clues that come from various off-line experiments, findings that indicate malfunctioning of WS semantic processing came from one on-line study as well (Neville et al., 1994). This on-line study directly observed WS brain responses to semantically inappropriate sentence endings by using ERPs. This method enabled the researchers to find out how WS brain reacts to semantic anomalies in contrasts to normal brain. The data from this on-line experiment showed that both the timing and the nature of real-time semantic processing in WS brain are to some degree different from the ones found in the normal brain. Finally, despite all the evidence that imply that semantic processing is not without anomalies, researchers that employed semantic priming task claim that semantic organization of WS individuals is not impaired (Tyler et al., 1997). They found no significant difference between WS and TD individuals in time they need to automatically access the semantic information.

How can we explain the discrepancies in the findings of these various studies on WS language comprehension? Is it possible that WS individuals possess unimpaired semantic memory and yet abnormal semantic processing? First, we must say that the hypothesis made based on the observations of explicit language behaviors should always be taken with caution. In other words, data found in the off-line studies mentioned above are not really reliable sources when it comes to deciding how the different brain processes in WS are functioning. However, if we look at the results of the two on-line studies presented above they might seem conflicting. Both of these studies aimed to examine automatic processes which are involved in language comprehension. Nevertheless, each had a different focus. Neville et al. (1994) focused on examining the WS automatic activation of brain responses on semantically appropriate versus inappropriate sentence endings, which involved the processing of words in a sentence. Tyler et al. (1997), on the other hand, focused on testing the WS semantic organization of the mental lexicon, which involved automatic activation of individual word meanings. Thus, Neville et al. (1997) observed the processing of words in isolation.

Taking into consideration findings of both studies, we can assume that the semantic memory of WS people is structured normally since they do not show any difficulties in accessing individual word meanings. However, when WS people start to integrate word meanings into sentential contexts, problems with the semantic processing arise. Such interpretation could also explain why they have far better scores on vocabulary comprehension tests that involve simple word to picture matching than on the ones that involve giving word explanations. In the former, they only need to access individual word meanings while in the latter they need to integrate different pieces of semantic information into sentential context. In the following chapter, I will discuss what the nature of the WS's unusual lexico-semantic processing might be. That is, whether the impairments in different parts of the cognition in WS individuals maybe interfere with their knowledge of language.

# 7 Language and other cognitive abilities – cross domain interactions

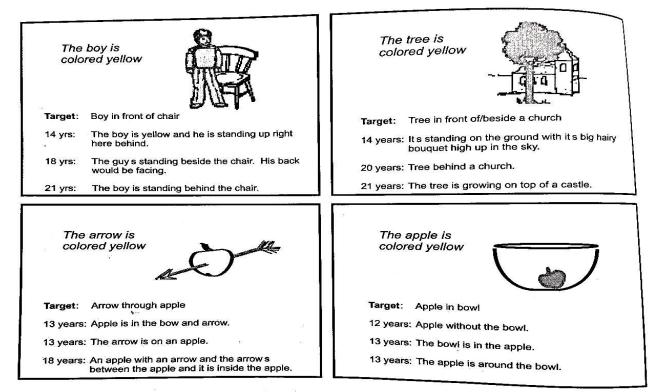
If the semantic organization of WS mental lexicon is functionally equivalent to the one of TD, why is the semantic processing of WS different from the one in TD? Tyler et al. (1997) argued that most of the tasks used for investigation of WS language comprehension involved more cognitive demands than WS individuals can keep track of due to their general mental disability. Their aim was to avoid this. If they succeeded to employ the task that examines "purely linguistic knowledge", we might assume that WS language comprehension shows anomalies whenever lexico-semantic processing requires interaction with other cognitive domains.

The first indication that WS lexico-semantic processing shows abnormalities when language interacts with other cognitive domains emerges from word-definition tasks. If we assume that a word-definition task involves the ability of the subject to make transformations in the properties of concepts, then we can say that such task (apart from involving the activation of appropriate lexico-semantic information) involves the non-linguistic ability of problem-solving. If this is so, it shouldn't be a surprise that WS adolescents and adults perform poorly in word-definition tasks since most of them also have difficulties solving simple transformational tasks from Piagetian number or mass conservation problems (Bellugi et al., 1988).

The second indication comes from the spontaneous speech of WS individuals. The retrieval of the appropriate semantic information and the integration of that information into sentential context might involve planning and sorting of the relevant words (semantic information) from the mental lexicon before their integration in context. Planning and sorting are again cognitive skills that are impaired in WS people (see Bellugi et al. 1988, 1994). Thus it is possible that the effect of this impairment is reflected in WS language.

Finally, third and maybe the strongest indication that WS individuals' lexico-semantic processing breaks down when other cognitive domains need to be included is their comprehension and usage of semantic relational terms. WS people show severe impairments in visuo-spatial intelligence (see Atkinson et al., 2001; Farran & Jarrold, 2003). Accordingly, it has been found that they also have difficulties comprehending and using directional (e.g., *right, left, up, down*), special (e.g., *in, on, above, below*), and positional (e.g., *top, bottom, high, low*) terms (Semel & Rosner, 2003). Bellugi et al. (2000) reported that WS subjects (10-

41 year-olds) make far more errors than their MA matches on the test specifically designed to probe their knowledge of spatial prepositions. In picture matching part of that test, WS subjects selected wrong picture referent for spatial prepositions (such as in front of, between, above) 11,5% of the time, while their TD controls made such errors only 0,2% of the time. In the part of the test where subjects are required to describe a picture containing spatial relations between two objects, WS individuals performed even worse (see below the examples of the errors they made; figure 1.14). They produced erroneous picture descriptions 30% of the time, as opposed to their TD controls who had a significantly lower error rate.



#### Figure 1.14

Figure 1.14 Experimental tasks were developed to investigate the intersection of language and spatial representation in Experimental tasks were developed to investigate the investigate and spatial representation in WMS, including production and comprehension tasks (modeled after Bowerman, 1996). Shown here are WMS, including production and comprehension and electron at the Bowerman, 1996). Shown here are sample errors made by WMS individuals ages 12 and older on a task involving production of spatial preposample errors made by WMS individuals agos is and crucial stimuli pictures, one of the objects in the picture is sitions and language about space. Note that on the date item, as in the picture of an arrow passing through colored yellow (but here is shaded) to represent the target item, as in the picture of an arrow passing through colored yellow (but here is shaded) to represent the mapping between language and spatial representa-an apple. Individuals with WMS exhibit difficulty in the mapping between language and spatial representaan apple. Individuals with wives exhibit dimension, and the arrow's between the apple and it is tion, e.g., 'The arrow is on the apple,' or 'An apple with an arrow and the arrow's between the apple and it is

(Taken from Bellugi et al., 2000, p. 23)

After this initial investigation by Bellugi et al. (2000), many researchers started to examine the interaction between WS visuo-spatial cognition and WS language (e.g., Landau & Zukowski 2003; Phillips et al., 2004; Landau & Hoffman, 2005; Lakusta, Wagner, & Landau, 2007; Lukacs, Pleh, & Racsmany, 2007). In the following sections, I will present Phillips et al. (2004) investigations and then discuss how their findings might be interpreted when it comes to the hypothesis that WS is evidence for a dissociation between language and other cognitive skills.

#### 7.1 Spatial language and spatial cognition

Similarly to Bellugi et al.'s (2000) investigation, Phillips et al. (2004) administered two studies to examine the interaction between language and visuo-spatial cognition in WS by testing individuals' comprehension of spoken language with a spatial component.

In the first study, they employed Test for Reception of Grammar (Bishop, 1983). The TROG is a standardized measure designed to test grammatical comprehension. It consists of 20 different tasks (blocks from A-T) each of which probes different grammatical constructions. In each task, the participant is presented with four pictures and is asked to point to the one that best corresponds to a spoken word, phrase, or sentence. The TROG is usually used for the assessment of verbal MA of WS individuals so that they could be matched and compared with the control groups in different experiments. When researchers use the TROG for these purposes, they simply assess the overall performances of subjects and convert them to the verbal MA values. Phillips et al. (2004) used the TROG for different purposes. They aimed to directly examine the possibility that spatial items within the TROG might be actually affecting WS individuals' overall performance on this test.

Phillips et al. (2004) estimated that there are three blocks within the TROG that could be considered to contain a spatial component. These three blocks are:

- (1) Block K, which tests the understanding of *longer*, *bigger*, *taller than*; (e.g., *The toothbrush is longer than the pencil.*)
- (2) Block M, which tests the understanding of prepositions *in* and *on*; (e.g., *The apple is in the basket*.)
- (3) Block P, which assesses understanding of prepositions *above* and *below*. (e.g., *The bird is below the flower*.)

(Adapted from Phillips et al., 2004)

The prediction of the study was that if individuals with WS have difficulties with the comprehension of spatial terms, as a result of their spatial cognitive impairments, then they ought to find these three blocks of the TROG especially difficult.

The first study tested the performance of 32 individuals with WS (8-38 year olds) and compared it to the performance of two control groups; a group of 32 children with moderate learning difficulties (MLD), and a group of 32 TD children. Both control groups were matched on verbal MA with WS subjects. The results showed that the subjects with WS indeed have particular difficulty in comprehending the sentences from the blocks; K, M, and P of the TROG. They made significantly more errors on these blocks (that contain spatial component) than both the TD and the MLD controls, who actually find these blocks quite easy. Contrary to this, the performance of WS subjects on other blocks (the ones that do not contain spatial component) was in line with the ones of TD and MLD controls. Thus, the findings confirm that individuals with WS, who have severe impairments on visuo-spatial components.

Since the TROG was not originally designed for an assessment of comprehension of spatial and non-spatial terms, and since it only has 3 blocks with a spatial aspect, Phillips et al. (2004) created a test that could specifically be used for these purposes. This test was named 'Test For Receptive Understanding of Spatial Terms' (TRUST). It was loosely based on the TROG, but it included more items with a spatial component. So besides the spatial terms that are used in the TROG (*in, on, above, below and longer/bigger*), the TRUST also assessed the comprehension of the terms *behind* (e.g., *The apple is behind the cup*), *in front of (e.g., The dog is in front of the house*) and *shorter/smaller* (e.g., *The snowman is shorter than the penguin*). For the sake of the comparison, the TRUST incorporated four tasks from the TROG that do not include spatial terms or constructions. These are:

- (1) Block L, which tests the understanding of <u>reversible passives</u>; (e.g., *The rabbit is pushed by the snail.*)
- (2) Block O, which tests the understanding of <u>X but not Y</u>; (e.g., *The pig is muddy but not pink.*)
- Block Q, which tests the understanding of not only X but also Y;
   (e.g., Not only the hat but also the boot is gray)
- (4) Block S, which tests the understanding of <u>neither X nor Y</u>. (e.g., *The strawberry is neither blue nor red.*)

#### (Adapted from Phillips et al., 2004)

Apart from these four blocks from the TROG, the TRUST also included one novel task that does not include spatial component and that is the comprehension of the comparatives *'darker than/lighter than'*. This task was included as a direct comparison for the spatial comparatives (e.g., *longer than/shorter than*).

This second study included 15 WS individuals whose performance was compared to two control groups matched on vocabulary MA. The TRUST was presented in practically the same way as the TROG, except that the pictures were presented on a computer screen. The results from this second study supported the results from the first study. They confirmed that, relative to the TD controls, the WS subjects seemed to have specific difficulties in comprehending spoken descriptions of spatial relationships. The results also showed that, compared to the controls, the WS subjects made significantly fewer errors on non-spatial as opposed to spatial terms. In fact, the WS subjects performed comparably to the TD matches on the non-spatial terms. This indicates that WS subjects' comprehension of non-spatial terms was is in line with their level of receptive vocabulary.

Phillips et al. (2004) made an interesting observation in this second study. They noticed that when WS individuals fail to select the correct picture in this task with a spatial component, they tend to choose the picture that depicts the exact opposite of the target spatial term. For example, for the spoken description *The dog is behind the house* they tend to choose the picture that depicts the dog that is standing in front of the house. If we take a closer look at the above sample errors of WS subjects from Bellugi et al. (2000) study, we can notice the same occurance. Phillips et al. (2004) called this type of error a systematic error. According to them, systematic errors suggest that WS individuals do not have any particular difficulty in comprehending the broad sense of what terms, such as *behind* and *in front of* refer to, rather they have difficulties "either in discriminating these terms from one another, or in representing the ordering of the relation between the two objects" (Phillips et al., 2004, p. 24).

All in all, finding from both studies by Phillips et al. (2004) provide strong evidence that people with WS experience a specific problem in understanding of language that includes a spatial component.

#### 7.2 Dissociation of language and other cognitive skills?

Since the earliest studies on WS, researchers have emphasized that people with WS show an imbalance in their cognitive profile. Various studies have found that the language skills of WS individuals are superior to the rest of their cognition (see chapter 4). Such

findings have led to the hypothesis that WS is evidence that verbal and non-verbal cognitive domains develop independently (e.g., Bellugi et al., 1994, 1996; Flavell et al., 1993; Levy, 1996; Pinker 1994). However, the more detailed studies of WS revealed many inconsistencies within the language domain itself and, as we saw in the previous two sections, some of these inconsistencies seem to be influenced by the non-verbal cognition. This might point to the fact that verbal and non-verbal domains do not function independently, but are rather involved in a complex process of interactions. Several authors have suggested that the language strengths of WS people might help them facilitate their non-linguistic performance (e.g., Bellugi et al., 1988, have argued that people with WS often talk their way through drawing tasks, as if they are using their verbal strengths to mediate visuo-spatial performance), while only few have considered whether non-verbal deficits of WS people actually impinge on their language performance.

The findings from Phillips et al. (2004) studies presented in the previous section provide a clue that WS people's difficulties in spatial awareness can constrain certain aspects of their language performance (for similar findings see also Landau & Zukowski 2003; Landau & Hoffman, 2005; Lakusta, Wagner, & Landau, 2007). This could be evidence that language and non-language domains are not necessarily separate from one another, as has typically been suggested. If this is so, the new question arises. In what way do spatial cognition and language interact in WS? Phillips et al. (2004) suggested two possible answers to this question. The first answer is that this "interaction is a reflection of conceptual difficulties in the comprehension of space rather than in any fundamentally linguistic difficulty with spatial language per se" (Phillips et al., 2004, p. 28). In other words, it should not be surprising that WS individuals have problems comprehending language with a spatial component when they have little knowledge of space. For instance, even a person with an excellent level of language abilities might have difficulties in making sense of language that incorporates terms from an subject area that he/she knows little about, e.g., quantum physics. The second answer is that "it may be that spatial difficulties in Williams syndrome interact in a more fundamental way with the 'on-line' comprehension of spoken descriptions of space" (Phillips et al., 2004, p. 28). That is, it is possible that general mental disability of individuals with WS limits their ability to construct spatial 'mental models' so that they could be able to understand the verbal descriptions of space. Thus, in contrast to the first interpretation, the second interpretation suggests that, even with an extensive teaching of what space is, WS people might not be able to fully grasp the notion of space because their mental abilities are constraining them, which in turn constrains their ability to understand spatial language.

The debate whether the verbal and non-verbal cognition of WS people interact or function separately clearly needs further investigations. The new investigations should include tests with language terms that have components from other impaired areas of WS cognition. For example, language terms or phrases that involve numbers or time, because these are also areas in which WS people show severe impairments (Bellugi et al., 1988; Paterson, 2000). However, taking into consideration that language is multifaceted, proving that WS lexico-semantic processing is abnormal because it is dependent on non-verbal cognitive domains still does not prove that all aspects of WS language interact with nonverbal cognitive domains. As we have seen earlier in this paper, some researchers proposed that the knowledge of grammar is intact in WS individuals (Clahsen & Almazan, 1998).

# 8. Is Williams Syndrom language a window into normal language development?

We have seen in chapter four that the studies which reported a significant advantage of WS language skills over their non-verbal skills were mostly conducted on WS adolescence and adults. These findings led researchers of nativist persuasion to claim that WS is evidence for the existence of language module. However, more recent studies, which employed the neurochostructivist approach to language acquisition, challenged this claim. These studies tested much younger children with WS and always aimed to include a larger number of WS participants who were then strictly matched on verbal MA to controls (cf. chapter 4). The results from these studies that followed developmental trajectories indicate that it is wrong to make conclusions about the initial state of the brain only by looking at its end state. As neuroconstructivists noted, the straightforward application of modularity theory to apparent strengths of WS adult brain evades the fact that the initial state of WS brain might be different from its adult state (cf. section 3.1.1). The data from studies conducted on infants and younger children with WS confirm this theory (cf. chapter 5). They demonstrate that WS language is delayed, but they do not report that it is better than it might be predicted considering their IQ levels. As Rossen et al. (1996) noted, the first significant advantages in WS vocabulary are not evident before the early adolescence. Moreover, the more detailed experimental studies of WS semantics suggest that even when WS individuals begin to show verbal advantages this still does not prove that their language skills are without anomalies (cf. chapters 6 and 7).

Since my analysis of the various language studies on WS individuals strongly indicates that WS language is neither intact nor does it follow typical development, my opinion is that it seems inappropriate to draw conclusions about typical language development based on WS language development.

# 9. Conclusion

By analyzing various studies on WS language, I found out several important facts about WS language. First, in contrast to TD children, the language development of WS children is delayed. This delay might occur due to their lack of typical pre-linguistic behaviors. Second, even after this delayed onset, WS children do not seem to follow the same language development. Some researchers suppose that the insensitivity of WS individuals to the lexical constraints indicate that they rely more on phonetics than on semantics when they learn new words. Third, I found out that, even though WS adults are very fluent speakers, they might not always understand everything they say. Several experiments indicate that there might be some abnormalities in their semantic processing. These abnormalities are usually shown on tests that are assumed to be more cognitively demanding. If this is true, we cannot say that language is entirely separate from the rest of the cognition. This further suggests that it is wrong to claim that WS is evidence for dissociation between language and other cognitive skills.

## **10.** Suggestions for further research

Language acquisition has always puzzled linguists. Many theories have been proposed, however, the nature-nurture debate has always remained. With the development of neuroconstructivism, this debate might finally be resolved. Nevertheless, neuroconstructivism indeed offers a new perspective on how we should observe language development, particularly the language development of people with cognitive impairments. We are witnessing that the new technological developments are shifting the focus of theoretical approaches to language acquisition from an observation of language behavior to a direct inspection of brain reactions to linguistic stimuli. This should not come as a surprise since the brain is a source of all our behaviors and experiences. In this thesis, I looked at the study which used ERPs to test the timing and location of brain responses to a word stimulus. Using such methods seems to be very useful, particularly in language investigations of people with a mental disorder like WS. My thesis examined ERP studies only in regards to WS semantics, however, it would be very interesting to find out whether the same or similar techniques have been used to test WS phonetics, especially because it has been proposed that individuals with WS might be relying more on phonetics than on semantics when they learn new words. Furthermore, since my thesis has mainly been focused on WS semantics, further research should encompass other aspects of WS language, such as syntax and morphology. Several studies I looked at suggested that even though WS individuals demonstrate good syntactic abilities these abilities do not exceed the 7-year-old level of normal language development. Finally, my investigation was based on studies conducted on English-speaking individuals with WS. In order to better understand the nature of WS language cross-linguistic studies should be included also.

#### References

- Akhtar, N., & Gernsbacher, M. A. (2007). Joint Attention and Vocabulary Development: A Critical Look. *Language and Linguistic Compass (vol.1, issue 3)*, 195-207.
- Ambridge, B., & Lieven E. V. M. (2011). Child Language Acquisition: Contrasting Theoretical Approaches. Cambridge: Cambridge University Press.
- Anderson, M. (1998). Mental retardation general intelligence and modularity. *Learning and individual differences*, *10*, 159-178.
- Annaz, D., Van Herwegen, J., Thomas, M., Fishman, R., Karmiloff-Smith, A., & Rundblad,
  G. (2009). Comprehension of metaphor and metonymy in children with Williams
  syndrome. *International Journal of Language and Communication Disorders*, 44(6), 962-978.
- Atkinson, J., Anker, S., Braddick, O., Nokes, L., Mason, A., & Braddick, F. (2001). Visual and visuospatial development in young children with Williams syndrome. *Developmental Medicine and Child Neurology*, 43, 330–337.
- Bakeman, R., & Adamson, L. (1984). Coordinating attention to people and objects in motherinfant and peer-infant interaction. *Child Development*, 55, 1278–1289.
- Baldwin, D. A. (1993). Early referential understanding infants ability to recognize referential acts for what they are. *Developmental Psychology*, 29(5), 832-843.
- Baldwin D. A. (1995). Understanding the link between joint attention and language. In: C.
  Moore, P. J. Dunham (Eds.) *Joint Attention: Its Origins and Role in Development* (pp. 131–158). Hillsdale, NJ: Erlbaum.
- Bellugi, U., Bihrle, A., Neville, H., Jernigan, T., & Doherty, S. (1992). Language, cognition, and brain organization in a neurodevelopmental disorder. In M. Gunnar & C. Nelson (Eds.), *Developmental behavioral neuroscience* (pp. 201-232). Hillsdale, NJ: Lawrence Erlbaum.
- Bellugi, U., George, M. St., Galaburda, A. M., Korenberg, J. R., Mills, D. L., Reiss, A. L. (2001). *Journey from Cognition to Brain to Gene: Perspectives form Williams Syndrome*. Cambridge MA, London England: The MIT Press.
- Bellugi, U., Lichtenberger, L., Jones, W., Lai, Z., & St. George, M. (2000). The neurocognitive profile of Williams syndrome: a complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*, 12, 7-29.

- Bellugi, U., Lichtenberger, L., Mills, D., Galaburda, A., & Korenberg, J.R. (1999). Bridging cognition, the brain, and modular genetics: evidence from Williams syndrome, *Trends in Neurosciences*, 22(5), 197-207.
- Bellugi, U., Marks, S., Bihrle, A.M., & Sabo, H. (1988). Dissociation between language and cognitive functions in Williams Syndrome. In D. Bishop & K. Mogford (Eds.), *Language development in exceptional circumstances* (pp. 177-189). London: Churchill Livingstone.
- Bellugi, U., Wang, P., Jernigan T. (1994). Williams syndrome: An Unusual Neuropsychological Profile. In Broman, S., Grafman, J. (Eds.), *Atypical cognitive deficit in developmental disorders: Implications for brain function* (pp. 23– 56). Hillsdale, N. J.: Lawrence Erlbaum Associates.
- Bennett, F., La Veck, B., & Sells, C. (1978). The Williams elfin faces syndrome: the psychological profile as an aid in syndrome identification, *Paediatrics*, 61(2), 303-306.
- Bertrand, J., Mervis, C., Rice, C.E., & Adamson, L. (1993). Development of joint attention by a toddler with Williams syndrome. Paper presented at the Gatlinberg Conference on Research and Theory in Mental Retardation and Developmental Disabilities, Gatlinberg.
- Beuren, A. J., Apitz, J., & Harmanz, D. (1962). Supravalvular aortic stenosis: a complex syndrome with mental retardation and a certain facial appearance. *Circulation*, 26, 1235-1240.
- Bishop, D. V. M. (1983). Test For Reception of Grammar. Manchester, U.K.: Medical Research Council.
- Bishop, D. V. M. (1997). Cognitive neuropsychology and developmental disorders: uncomfortable bedfellows. *Quarterly Journal of Experimental Psychology*, 50A, 899-923.
- Bromberg, H., Ullman, M., Marcus, G., Kelly, K., & Coppola, M. (1994). A dissociation of memory and grammar: Evidence from Williams syndrome. Paper presented at the Eighteenth Annual Boston University Conference on Language Development.
- Bruner, J. (1983). Child's Talk: Learning to Use Language. Oxford: Oxford University Press.
- Butterworth, G., & Morissette, P. (1996). Onset of pointing and the acquisition of language in infancy. *Journal of Reproductive and Infant Psychology*, *14*, 219–231.

- Carpenter, M., Nagell, K., & Tomasello, M. (1998). Social cognition, joint attention and communicative competence from 9 to 15 months of age. *Monographs of the Society for Research in Child Development*, 60(4), i-vi, 1-143.
- Chomsky, N. (1959). A Review of B. F. Skinner's Verbal Behavior. Language, 35(1), 26-58.
- Chomsky, N. (1972). Language and Mind. New York: Harcourt Brace Jovanovich.
- Chomsky, N. (1980). Rules and representations. Oxford: Blackwell.
- Chomsky, N. (1986). *Knowledge of language: Its nature, origin, and use*. New York: Praeger.
- Clahsen, H., & Almazan, M. 1998. Syntax and morphology in Williams syndrome. *Cognition*, 68(3), 167-198.
- Clahsen, H., Ring, M., & Temple, C. (2004). Lexical and morphological skills in English speaking children with Williams syndrome, in S. Bartke and J. Siegmüller (Eds.), *Williams syndrome across languages*, (pp. 221-244) Amsterdam: Benjamins.
- Clark, E. V. (1993). The Lexicon in Acquisition. Cambridge: Cambridge University Press.
- Coulson, S., King, J., & Kutas, M. (1998). Expect the unexpected: event-related brain response to morphosyntactic violations. *Language and Cognitive Processes*. 13(1), 21–58.
- Crisco, J. J., Dobbs, J.M., & Mulhern, R.K. (1988). Cognitive processing of children with Williams syndrome. *Developmental Medicine and Child Neurology*, *30*(5), 650-656.
- Dale, A. M., & Sereno, M. I. (1993). Improved localization of cortical activity by combining EEG and MEG with MRI cortical surface reconstruction. *Journal of Cognitive Neuroscience 5*, 162–176.
- 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.
- Elsabbagh, M., & Karmiloff-Smith, A. (2004). Modularity of mind and language. *The Encyclopedia of Language and Linguistics*, 2, 1-25.
- Ewart, A. K., Morris, C. A., Atkinson. D., Weishan, J., Stemes, K., Spallone, P., Stock, D. A., Leppert, M., & Keating, M.T. (1993). Hemizygosity at the elastin locus in a developmental disorder, Williams Syndrome. *Nature Genetics*, 5, 11-16.
- Farran, E. K., & Jarrold, C. (2003). Visuo-spatial cognition in Williams syndrome: reviewing and accounting for the strengths and weaknesses in performance. *Developmental Neuropsychology*, 23, 175–202.

- Fishman, I., Yam, A., Bellugi, U., Lincoln, A., & Mills, D. (2010). Contrasting patterns of language-associated brain activity in autism and Williams syndrome. *Social Cognitive* and Affective Neuroscience, doi:10.1093/scan/nsq075
- Flavell, J. H., Miller, P. H., & Miller, S. A. (1993). Cognitive development. Englewood Cliffs, NJ: Prentice Hall.
- Fodor, J. A. (1983). *The Modularity of Mind: An Essay on Faculty Psychology*. Cambridge, MA: MIT Press.
- Frigerio, E., Burt, D. M., Gagliardi, C., Cioffi, G., Martelli, S., Perrett, D. I., et al. (2006). "Is everybody always my friend?" Perception of approachability in Williams syndrome. *Neuropsychologia*, 44, 254–259.
- Gagliardi, C., Frigerio, E., Burt, D. M., Cazzaniga, I., Perrett, D. I., & Borgatti, R. (2003). Facial expression recognition in Williams syndrome. *Neuropsychologia*, *41*, 733–738.
- Gleason, J. B., & Ratner, N. B. (2009). The development of language. Boston: Pearson.
- George, St. M., Mills, D. L., & Bellugi, U. (2000). ERPs during auditory language comprehension in Williams syndrome. The effects of word frequency, imageability and length on word class. Neuroimage, 11, 357.
- Goldberg, A. E. (1995). *Constructions: A Construction Approach to Argument Structure*. Chicago: University of Chicago Press.
- Goldberg, A. E. (2006). *Construction at Work: The Nature of Generalization in Language*. New York: Oxford University Press.
- Gonçalves, Ó. F., Pinheiro, A. P., Sampaio, A., Sousa, N., Férnandez, M., & Henriques, M. (2011). Autobiographical Narratives in Williams Syndrome: Structural, Process and Content Dimensions. *Journal of Developmental and Physical Disabilities*, 23(4), 289-302.
- Gopnik, A., & Meltzoff, A. (1987). The development of categorization in the second year and its relation to other cognitive and linguistic developments. *Child Development 58*, 1523–1531.
- Grant, J., Karmiloff-Smith, A., Gathercole, S. E., Paterson, S., Howlin, P., Davies, M., et al. (1997). Phonological short-term memory and its relationship to language in Williams syndrome. *Journal of Cognitive Neuropsychiatry*, 2(2), 81–99.
- Greer, M. K., Brown, F. R., Shashidhar, G., Choudry, S. H., & Klein, A .J. (1997). Cognitive, adaptive, and behavioural characteristics of Williams syndrome. *American Journal of Medical Genetics (Neuropsychiatric Genetics)*, 74(5), 521-525.

- Guasti, M. T. (2002). *Language Acquisition: The Growth of Grammar*. Cambridge MA: The MIT Press.
- Gulan, T., & Valerjev, P. (2010). Semantic and related types of priming as a context in word recognition, *Review of Psychology*, 17(1), 53-58.
- Hagerman, R. J. (1999). *Neurodevelopmental disorders: Diagnosis and treatment*. New York: Oxford University Press.
- Hirsch, C. & Wexler, K. (in press). The late development of raising: what children seem to think about *seem*. In W. Davis & S. Dubinsky (Eds.), *New Horizons in the Analysis of Control and Raising*. Springer.
- Ilves, P., Tomberg, T., Kepler, J., Laugesaar, R., Kaldoja, M-L, Kepler, K., & Kolk, A. (2014). Different plasticity patterns of language function in children with perinatal and childhood stroke. *Journal of Child Neurology*, 29(6), 756-764.
- Jarrold, C., Hartley, S., Philips, C., & Baddeley, A.D. (2000). Word fluency in Williams syndrome: Evidence for unusual semantic organisation? *Cognitive Neuropsychiatry*, 5(4), 293–319.
- Joffe, V., & Varlokosta, S. (2007). Patterns of syntactic development in children with Williams syndrome and Down's syndrome: evidence from passives and wh-questions. *Clinical Linguistics and Phonetics*, 21, 705-727.
- Jones, K. L., & Smith, D. W. (1975). The Williams elfin facies syndrome. The Journal of Pediatrics, 86, 718-723.
- Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*, 12(S1), 30-46.
- Karmiloff-Smith, A. (1992). *Beyond Modularity: A Developmental Perspective on Cognitive Science*. Cambridge, MA: The MIT Press.
- Karmiloff-Smith, A. (1998). Development itself is the key to understanding developmental disorders. *Trends in Cognitive Sciences*, *10*(2), 389-398.
- Karmiloff-Smith, A. (2009). Nativism Versus Neuroconstructivism: Rethinking the Study of Developmental Disorders. *Developmental Psychology*, 45(1), 56–63.
- Karmiloff-Smith, A. (2015). An Alternative to Domain-general or Domain-specific Frameworks for Theorizing about Human Evolution and Ontogenesis. AIMS Neuroscience, 2(2), 91-104.

- Karmiloff-Smith, A., Grant, J., Berthoud, I., Davies, M., Howlin, P., & Udwin, O. (1997).
  Language and Williams syndrome: How intact is "intact"? *Child Development*, 68, 246–262.
- Karmiloff-Smith, A., & Karmiloff, K. (2001). *Pathways to Language: From Fetus to Adolescence*. Cambridge, MA & London, England: Harvard University Press.
- Kataria, S., Goldstein, D. J., & Kushnik, T. 1984. Developmental delays in Williams ("elfin facies") syndrome. *Applied Research in Mental Retardation* 5(4), 419-23.
- Korenberg, J. R., Chen, X. N., Hirota, H., Bellugi, U., Burian, D., Roe, B., et al. (2000). Genome structure and cognitive map of Williams syndrome. *Journal of Cognitive Neuroscience*, 12, 89–107.
- Kutas, M., & Dale, A. (1997). Electrical and magnetic readings of mental functions. In M. D. Rugg (Ed.), *Cognitive Neuroscience* (pp. 197–242). Psychology Press
- Kutas, M., & Hillyard, S. A. (1980). Reading senseless sentences: brain potentials reflect semantic incongruity. *Science* 207, 203–205.
- Kutas, M., & Federmeier, K. D. (2000). Electrophysiology reveals semantic memory use in language comprehension. *Trends in Cognitive Sciences*, 4(12), 463-470.
- Laing, E., Butterworth, G., Ansari, D. Gsödl, M., Longhi, E., Panagiotaki, G., et al. (2002). Atypical development of language and social communication in toddlers with Williams syndrome. *Developmental Science*, 5(2), 233–246.
- Laing, E., Hulme, C., Grant, J., & Karmiloff-Smith, A. (2001). Learning to read in Williams syndrome: Looking beneath the surface of atypical reading development. *Journal of Child Psychology and Psychiatry*, 42, 729–739.
- Lakusta, L., Wagner, L., & Landau, B. (2007). Conceptual foundations of spatial language: Evidence for a goal bias in infants. *Language Learning and Development*, 3(3), 179-197.
- Landau, B., & Hoffman, J. E. (2005). Parallels between spatial cognition and spatial language: Evidence from Williams syndrome. *Journal of Memory and Language*, 53, 163-185.
- Landau, B., & Zukowski, A. (2003). Objects, motions and paths: Spatial language in children with Williams syndrome. *Developmental Neuropsychology*, 23(1&2), 105-137.
- Laws, G., & Bishop, D. (2004). Pragmatic language impairment and social deficits in Williams syndrome: a comparison with Down's syndrome and specific language impairment. *International Journal of Language and Communication Disorders*, 39(1), 45-64.

Levy, Y. (1996). Modularity of language reconsidered. Brain and Language, 55, 240-263.

Lukacs, A., Pleh, C., & Racsmany, M. (2007). Spatial language in Williams syndrome: Evidence for a special interaction? *Journal of Child Language*, *34*(2), 311-343.

- McCarthy, D. (1972). McCarthy *scales of children's abilities*, San Antonio, TX: Psychological Corporation.
- Markman, E. M. (1990). Constraints Children Place on Word Meanings. Cognitive Science 14, 57-77.
- Markman, E. M. & Hutchinson, J. E. (1984). Children's sensitivity to constraints on word meaning: taxonomic versus thematic relation. *Cognitive Psychology*, 16, 1-27.
- Markman, E. M., & Wachtel, G. F. (1988). Children's use of mutual exclusivity to constrain the meanings of words. *Cognitive Psychology* 20, 121–157.
- Mervis, C. B., & Becerra, A. M. (2007). Language and communicative development in Williams syndrome. *Mental Retardation and Developmental Disabilities Research Reviews*, 13, 3-15.
- Mervis, C. B., & Bertrand, J. (1997). Developmental relations between cognition and language: Evidence from Williams Syndrome. In L. B. Adamson & M. A. Romski (Eds.), *Research on Communication and Language Disorders: Contributions to Theories of Language Development*. New York: Brookes.
- Mervis, C.B., Morris, C. A., Bertrand, J, & Robinson, B. F. (1999). Williams Syndrome:
  Findings from an integrated program of research. In H. Tager-Flusberg (Ed.) *Neurodevelopmental disorders* (pp. 65-110). Cambridge, MA: MIT Press.
- Mervis, C. B., Robinson, B. F., Rowe, M.L., Becerra, A. M., & Klein-Tasman, B.P. (2003).
  Language abilities of individuals with Williams syndrome. In L. Abbeduto (ed.),
  International Review of Research in Mental Retardation 27, (pp. 35-81). San Diego,
  CA: Academic Press.
- Mervis, C. B., Golinkoff, R. M., & Bertrand, J. (1994). Two-year-olds readily learn multiple labels for the same basic-level category. *Child Development*, 65, 1163–1177.
- Meyer, D. E., & Schvaneveldt, R. W. (1971). Facilitation in recognizing pairs of words: Evidence of dependence between retrieval operations. *Journal of Experimental Psychology*, 90, 227-234.
- Mills, D., Llamas, T., St George, M., Doyle, T. F., Neville, H., & Korenberg, J. R. (2003). Electrophysiological signatures of abnormal auditory language processing in infants, children and adults with Williams syndrome. San Diego: Institute for Neural Computation, University of California.

- Morris, C. A., Demsey, S. A., Leonard, C. O., Dilts, C., & Blackburn, B. L. (1988). Natural history of Williams syndrome: Physical Characteristics. *Journal of Pediatrics*, 113(2), 318–326.
- Mundy, P. & Gomes, A. (1998). Individual differences in joint attention skill development in the second year. *Infant Behaviour and Development*, 21(3), 469–482.
- Murphy, C. M., & Messer, D. J. (1977). Mothers, infants and pointing: A study of gesture. InH. R. Schaffer (Eds.), *Studies in Mother-Infant Interaction* (pp. 325-354). New York: Academic Press.
- Neville, H. J., Holcomb, P. J., & Mills, D. M. (1989). Auditory, sensory and language processing in Williams syndrome: An ERP study. *Journal of Clinical and Experimental Neuropsychology*, 11(52).
- Neville, H. J., Mills, D. L., & Bellugi, U. (1994). Effects of altered auditory sensitivity and age of language acquisition on the development of language-relevant neural systems: Preliminary studies of Williams syndrome. In S. Broman and J. Grafman (Eds.), *Cognitive Deficits in Developmental Disorders: Implications for Brain Function* (pp. 67-83).Hillsdale, NJ: Lawrence Erlbaum.
- Nelson, K. (1985). *Making Sense: The Acquisition of Shared Meaning*. New York and London: Academic Press.
- Pagon, R., Bennet, F., La Veck, B., Stewart, K., & Johnson, J. (1987). Williams syndrome: Features in late childhood and adolescence. *Paediatrics*, 80(1), 85-91.
- Paterson, S. J. (2000). The development of language and number understanding in Williams Syndrome and Down's Syndrome: Evidence from the infant and mature phenotypes. Unpublished doctoral thesis, University College London.
- Paterson, S. J., Brown, J. H., Gsödl, M. K., Johnson, M. H., & Karmiloff-Smith, A. (1999). Cognitive modularity and genetic disorders. *Science*, 286, 2355–2358.
- Perovic, A., & Wexler, K. (2007). Complex grammar in Williams syndrome. *Clinical Linguistics and Phonetics*, 21, 729–745.
- Phillips, C. E., Jarrold, C., Baddeley, A. D., Grant, J., & Karmiloff-Smith, A. (2004). Comprehension of spatial language terms in Williams syndrome: Evidence for an interaction between domains of strength and weakness. *Cortex*, 40, 85-101.
- Piaget, J. (1952). *The Origins of Intelligence in Children*. New York: International University Press.
- Pinker, S. (1991). Rules of language. Science, 253, 530-535.
- Pinker, S. (1994). The language instinct. London: Penguin Books.

Pinker, S. (1999). Words and rules. London: Weidenfeld & Nicolson.

- Pinker, S., & Prince, A. (1994). Regular and irregular morphology and the psychological status of rules of grammar. In S. D. Lima, R. L. Corrigan, & G. K. Iverson (Eds.), *The reality of linguistic rules*. Amsterdam: Benjamin.
- Radford, A. (2004). *Minimalist syntax: exploring the structure of English*. Cambridge: Cambridge University Press.
- Reilly, J., Klima, E. S., & Bellugi, U. (1990). Once more with feeling: affect and language in atypical populations. *Development and Psychopathology*, 2(4), 367-391.
- Rossen, M., Klima, E. S., Bellugi, U., Bihrle, A., & Jones, W. (1996). Interaction between language and cognition: evidence from Williams syndrome, in J.H. Beitchman, N. Cohen, M. Konstantareas, & R. Tannock (Eds.), *Language, learning, and behavior disorders: developmental, biological, and clinical perspectives* (pp. 367-392). New York: Cambridge University Press.
- Semel, E., & Rosner, S.R. (2003). Understanding Williams Syndrome: Behavioral Patterns and Interventions. New Jersey: Lawrence Erlbaum Associates.
- Setter, J., Stojanovik, V., van Ewijk, L., & Moreland, M. (2007). Affective prosody in children with Williams syndrome, *Clinical Linguistics & Phonetics*, 21(9), 659-672.
- Sinclair, H. (1971). Sensorimotor action patterns as a condition for the acquisition of syntax. In R. Huxley & E. Ingram (Eds), *Language Acquisition: Models and Methods* (pp. 121-135). London: Academic Press.
- Sinclair, H. (1987). Language: a gift of nature or a home-made tool? In S. Modgil & C. Modgil (Eds.), Noam Chomsky: *Consensus and Controversy* (pp. 173-180).
  London: The Falmer Press.
- Stevens, T., & Karmiloff-Smith, A. (1997). Word learning in a special population: do individuals with Williams syndrome obey lexical constraints? *Journal of Child Language*, 24, 737–765.
- Stojanovik, V. (2006). Social interaction deficits and conversational inadequacy in Williams syndrome. *Journal of Neurolinguistics*, *19*(2), 157-173.
- Stojanovik, V., & van Ewijk, L. (2008). Do children with Williams Syndrome have unusual vocabularies? *Journal of Neurolinguistics*, 21(1), 18-34.
- Stojanovik, V., Perkins, M. & Howard, S. (2006). Linguistic heterogeneity in Williams syndrome. *Clinical Linguistics and Phonetics*, 20, 547-552.
- Strømme, P., Bjørnstad, P.G., & Ramstad, K. (2002). Prevalence estimation of Williams syndrome. *Journal of Child Neurology*, 17(4), 269–271.

Temple, C. (1997). Developmental cognitive neuropsychology. Hove, UK: Psychology Press.

- Thomas, M., Grant, J., Barham, Z., Gsödl, M., Laing, E., Lakusta, L., Tyler, L., Grice, S., Paterson, S., & Karmiloff-Smith, A. (2001). Past tense formation in Williams syndrome. *Language and Cognitive Processes*, 16(2), 143-176.
- Thomas, M., Purser, H., & Richardson, F. (2013). Modularity and Developmental Disorders. *The Oxford Handbook of Developmental Psychology*, *2: Self and Other*, 481-505.
- Tillema, J., Byars, A., Jacola, L., et al. (2008). Cortical reorganization of language functioning following perinatal left MCA stroke. *Brain and Language*, 105(2), 99– 111.
- Tomasello, M. (2003). Constructing a Language: A Usage-Based Theory of Language Acquisition. Cambridge, MA: Harvard University Press.
- Tomasello, M., & Akhtar, N. (2000). Five questions for any theory of word learning. In R. Golinkoff & K. Hirsh-Pasek (Eds.) *Becoming a word learner: A debate on lexical acquisition* (pp. 115-136). New York: Oxford University Press.
- Tyler, L. K. (1992) Spoken Language Comprehension: An Experimental Approach to Normal and Disordered Processing. Cambridge. MA: MIT Press.
- Tyler, L. K., Karmiloff-Smith, A., Voice, J. K., Stevens, T., Grant, J. Udwin, O., Davies, M., & Howlin, P. (1997). Do individuals with Williams syndrome have bizarre semantics? Evidence for lexical organization using an on-line task. *Cortex*, 33(3), 515-527.
- Udwin, O., & Yule, W. (1990). Expressive language of children with Williams syndrome, American Journal of Medical Genetics, 37(S6), 108-114.
- Udwin, O., & Yule, W. (1991). A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*, *13*(2), 232-244.
- Udwin, O., Yule, W., & Martin, N. (1987). Cognitive abilities and behavioural characteristics of children with idiopathic infantile hypercalcaemia. *Child Psychology and Psychiatry*, 28(2), 297-308.
- Volterra, V., Capirci, O., Pezzini, G., Sabbadini, L., & Vicari, S. (1996). Linguistic abilities in Italian children with Williams syndrome. *Cortex*, *32*, 663–677.
- von Arnim, G. & Engel, P. (1964). Mental retardation related to hypercalcaemia. *Developmental Medicine and Child Neurology*, *6*, 366-377.
- Wechsler, D. (1976). Wechsler intelligence scale for children-revised. Windsor: NFER.
- Williams, J. C. P., Barrett-Boyes, B. G., & Lowe J. B. (1961). Supravalvular aortic stenosis. *Circulation*, 24, 1311-1318.

Ypsilanti, A., Grouious, G., Alevriadou, A., & Tsapkini, K. (2005). Expressive and receptive vocabulary in children with Williams and Down syndromes. *Journal of Intellectual Disability Research*, 49(5), 353-364.