Chapter 6
Williams Syndrome

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6.1 Introduction

When encountering a person with Williams syndrome (WS) for the first time, one can be overwhelmed by an impression of instant and unbounded friendliness. This experience is often hard to reconcile with the understanding that the same person is likely to have intellectual disability, and has serious difficulties making sense of the social world. This is one of the many paradoxes that have made WS a fascinating target of investigations across several research fields – from genetics to the philosophy of language – over the last decades. The chapter opens with a few historical notes about how WS – a relatively rare neurodevelopmental disorder of genetic aetiology – came to be at the center of heated theoretical debates concerning the relations between language, cognition, and social behavior, and ultimately about the structure of the human mind. The brief excursion into the history of research on this syndrome provides a context for understanding the current state of knowledge about pragmatics in WS, another area of seemingly paradoxical capacities demonstrated by people with this intriguing neurodevelopmental disorder.

In the next section of the chapter, the neuropsychological and behavioral profile of individuals with WS is outlined, with a particular focus on two domains that play a critical role in the development of pragmatic skills: language and social-cognition. These domains had initially been considered to be 'intact' or 'spared' in people with WS, despite their 'severe' intellectual disability' (Von Arnim & Engel, 1964; Bellugi et al., 1988). Later research findings, however, indicated that this interpretation was premature, and that the linguistic, cognitive and social-behavioral phenotypes associated with WS consist of a complex mixture of strengths and deficits within these domains (for reviews see Järvinen-Pasley et al., 2008; Martens et al., 2008; Mervis, 2006; Plesa Skwerer, 2017).

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The rest of the chapter presents an overview of the main assessment methodologies that have been used so far in research investigating pragmatic functioning in WS. These range from parent-report questionnaires to qualitative conversational analyses. Results of these varied methodological approaches have converged toward demonstrating that pragmatics represents an area of particular difficulty for people with WS across the lifespan. What may account for these pragmatic difficulties is then discussed in light of developmental findings about the early socio-communicative behaviors shown by infants and young children with WS. The final sections of the chapter address the clinical implications of the research findings reviewed and suggest potential directions for future research.

6.2 Williams Syndrome – A Brief Research History

The history of research into WS provides clues about why this rare neurodevelopmental disorder with a relatively well-understood genetic basis gained such prominence in theoretical debates about fundamental human capacities in cognition, language, communication and social behavior more generally. The syndrome was first described in the early 1960s by physicians in New Zealand (Williams et al., 1961) and Germany (Beuren et al., 1962), who independently noted the remarkably similar medical and cranio-facial characteristics of a group of patients with a common heart condition – supravalvular aortic stenosis – and with developmental delay/mental retardation. Several psychological commonalities were also noted in the clinical description of these cases and soon the condition was recognized as a particular developmental disorder, which eventually came to be known as Williams syndrome, or Williams-Beuren syndrome.

In their original clinical observations of six children with infantile hypercalcemia[1] described in 1964, von Arnim and Engel highlighted "an unusual command of language" as a salient feature of the children's "psychological structure", which appeared to be in sharp contrast to "the severe mental retardation that is invariably present" (Von Arnim & Engel, 1964, p. 367). Considering the "mental similarities" between these children, the authors stated: "Their IQ is about 40–50 but they show outstanding loquacity and a great ability to establish interpersonal contacts" (p. 376). At face value, such statements resonate strongly with the view of independence of language or social cognition from other cognitive domains, a theoretical perspective that became very influential in the 1980s after the publication of Fodor's (1983) seminal work, *Modularity of Mind*. According to this view, language and other core knowledge domains constitute innately specified modules that operate independently and, therefore, may be selectively 'spared' or 'impaired'.

Early studies of the cognitive phenotype associated with WS emphasized dissociations between domain-specific abilities (e.g. "language" and "cognition"; see

[1] A condition involving excessive blood calcium levels.

Bellugi et al., 1988, 1992, 1994), and the syndrome started to be cited as providing evidence in support of the modularity of mind hypothesis (Clahsen & Temple, 2003; Jackendoff, 1994; Piattelli-Palmarini, 2001; Pinker, 1999). This interpretation, with its strong theoretical implications for understanding the organization of cognitive systems more generally (Anderson, 1998; Pinker, 1994, 1999), led to a surge in research on the structural aspects of language (morphology, phonology, syntax, semantics) in WS, and to assertions of 'intact language' in the face of severe intellectual disability/cognitive impairment (Bellugi et al., 1988, 1990, 1994; Rossen et al., 1996; Pinker, 1994).

Over the past two decades, however, initial claims of preserved language, independent from other domain-specific abilities (e.g. non-verbal cognition), have been replaced by a more nuanced view of uneven linguistic and cognitive profiles developing interdependently, with relative strengths and deficits both across and within domains, in the mature WS phenotype (Brock, 2007; Mervis, 1999, 2004, 2006; Mervis & Bertrand, 1997). Nevertheless, descriptions of people with WS as having 'striking language' and strong social skills alongside severe cognitive deficits persisted for decades in the literature and are still prevalent to this day in the popular media (Finn, 1991; Dobbs, 2007; Mervis & John, 2010). One consequence of this prevailing view of the WS behavioral phenotype has been a relative lack of clinical interest in targeting pragmatic language in interventions for people with WS. Moreover, in early studies, the pragmatic skills of individuals with WS were described as a particular strength, often by comparison to the significant impairments that are found almost universally among people with autism, regardless of their structural language abilities (Karmiloff-Smith et al., 1995).

Research on language in WS underwent a period of intense focus on the morphosyntactic skills of individuals with WS (Bellugi et al., 1988, 1997; Clahsen & Almazan, 1998; Levy & Bechar, 2003; Perovic & Wexler, 2007; Ring & Clahsen, 2005; Zukowski, 2004) in the hope of documenting the functional independence and modular organization of such capacities. However, initial claims of 'intact' language or 'preserved' components of language, presumed to be under genetic specification, developing independently from other non-linguistic domains, failed to be validated by empirical findings. A similar mixed picture of abilities and clear deficits, more in line with individuals' overall level of cognitive functioning than with notions of 'selective sparing', eventually emerged from research on social cognition and social perception in WS, as will be described later in the chapter.

Inconsistencies between initial and later findings about the language and social cognitive abilities of people with WS can be explained by methodological shortcomings of earlier empirical research on WS, such as small samples, with participants from a wide age range, and problematic choices of control groups or group-matching procedures, given that the majority of investigations relied on group-comparison designs (for critiques see Brock et al., 2009; Martens et al., 2008; Mervis, 2004; Mervis & Robinson, 2003). For instance, individuals with Down syndrome (DS) were often selected as a contrast group for participants with WS in studies of language abilities (Bellugi et al., 1988, 1994, 1999; Vicari et al., 2002) even though many aspects of language are particular weaknesses in people with DS,

relative to their overall cognitive abilities. Many studies compared participants with WS with individuals developing typically, matching groups on mental age, which inevitably required the inclusion of much younger typically developing (TD) children as a contrast group for older participants with WS. Finally, the view of a fractionated cognitive profile with syndrome-specific 'peaks' and 'valleys' in abilities was almost entirely missing any developmental or cultural dimensions (Karmiloff-Smith et al., 1997; Paterson et al., 1999; Thomas, 2005).

Later research studies, using appropriate comparison groups, larger samples, and taking developmental processes into account, have shown that neither language (or any structural aspect of language, e.g. grammar, morphosyntax) nor social cognition or social perception proved to be 'intact' domains in WS (Karmiloff-Smith et al., 2003). In light of these findings, research interest gradually shifted toward investigating how people with WS used language for social purposes, given their distinctive profile of social engagement. Thus, the study of pragmatic language in WS had a late start, but it is growing rapidly, encompassing a variety of assessment approaches and settings. So far, studies on WS over the last almost three decades have yielded a complex cognitive and behavioral profile that shows significant heterogeneity across many domains of abilities, including pragmatics (Laws & Bishop, 2004; Porter & Coltheart, 2005). However, despite substantial individual variability, the majority of people with WS can be characterized by a distinctive profile of neurogenetic, psychological and behavioral phenotypes, outlined next.

6.3 Phenotypic Characteristics of Williams Syndrome

Williams syndrome is among several neurodevelopmental disorders of known genetic aetiology associated with a unique cognitive and social-behavioral phenotype, in which language and social communication appear to be distinctive features. While initially considered to be a very rare disorder, current estimates indicate that it occurs in approximately 1 in 7500 live births and in about 6% of individuals with intellectual disability of genetic aetiology (StrØmme et al., 2002). The syndrome's genetic basis is a hemizygous contiguous deletion of approximately 28 genes in chromosome band 7q11.23[2] (Hillier et al., 2003; Osborne, 2006). The deletion region includes one copy of the ELASTIN gene and elastin deficiency is considered to account for some of the vascular and connective tissue abnormalities associated with WS. The condition can be diagnosed at or shortly after birth by using a fluorescence *in situ* hybridization probe for the missing ELASTIN gene, a technique which has been available since the 1990s.

A characteristic set of physical, medical and behavioral attributes commonly seen in people with WS include connective tissue and cardiovascular abnormalities

[2] The deletion refers to loss of one of the two copies of a segment of the long arm of chromosome 7, which includes a number of adjacent missing genes.

(Ewart et al., 1993), distinctive craniofacial features, infantile hypercalcemia, sensory modulation problems such as hypersensitivity to sound, failure to thrive or growth deficiencies, and premature aging (Morris, 2006, 2010). The majority of individuals with WS have intellectual disability in the mild to moderate range (mean full-scale IQ between 50–60), although there is wide variability in intellectual functioning within the population (Mervis & John, 2010; Porter & Coltheart, 2005). On standardized assessments of cognitive functioning, verbal ability scores tend to be better than non-verbal (performance) IQ scores, although the discrepancy is not universal, and it depends in part on the type of test administered (Jarrold et al., 1998; Martens et al., 2008; Mervis & John, 2010). However, global assessments of cognitive functioning may mask a checkered pattern of markedly uneven abilities within particular domains. Initial research on WS highlighted severe deficits in spatial cognition, number processing and problem-solving alongside apparent strengths in language, face processing and social cognition (Donnai & Karmiloff-Smith, 2000; Mervis et al., 2000).

A distinctive personality and social-behavioral profile, which has been consistently described as 'hypersocial' (Jones et al., 2000; Järvinen et al., 2013; Porter et al., 2007) with an 'undercurrent of anxiety' (Mervis & Klein-Tasman, 2000), is demonstrated by both children and adults with WS. At all ages people with WS show an exaggerated 'affiliative drive' manifested in unusually friendly, affectionate, outgoing, gregarious, and empathic behavior (Doyle et al., 2004; Fidler et al., 2007; Gosch & Pankau, 1997; Järvinen-Pasley et al., 2010; Klein-Tasman & Mervis, 2003; Klein-Tasman et al., 2011; Plesa Skwerer & Tager-Flusberg, 2016). Despite these characteristics, individuals with WS have significant difficulties in social functioning, in forming and maintaining friendships, and often develop high levels of anxiety and social isolation as they age (Davies et al., 1998; Einfeld et al., 2001; Elison et al., 2010).

6.3.1 Language Profile in Williams Syndrome

Pragmatic language abilities emerge at the intersection of linguistic and social-cognitive skills. Both language and social cognition have been extensively studied in WS (see Brock, 2007; Brock et al., 2009; Järvinen et al., 2013; Mervis & Becerra, 2007; Plesa Skwerer & Tager-Flusberg, 2006, 2011 for reviews). The history of research in these domains presents a similar evolution from initial assumptions of either 'intact' or 'sparing' of abilities, to accumulating evidence of significant difficulties. Over time, the theoretical controversies that fueled continued interest in examining the language abilities of individuals with WS have shifted from debates about modularity and the independence of language from other aspects of cognition, to discussions of developmental trajectories and sources of heterogeneity in linguistic and non-linguistic communicative abilities in this disorder (Thomas & Karmiloff-Smith, 2003; Stojanovik et al., 2006; Van Herwegen et al., 2011).

To date, comprehensive characterizations of the language phenotype associated with WS have been based on analyses of performance on a variety of standardized language tests or psycholinguistic tasks, administered in the context of cross-sectional research designs (see Brock, 2007; Mervis & Becerra, 2007 for reviews). A growing body of evidence from research across several languages (see Bartke & Siegmüller, 2004) suggests that overall language skills in individuals with WS are commensurate with their mental-age levels, with certain areas of specific abilities showing better performance than expected based on non-verbal cognitive functioning (e.g. receptive vocabulary, particularly knowledge of concrete words). Within the language domain, according to Mervis and Velleman (2011), "concrete vocabulary and phonological skills are relative strengths, grammatical abilities are at the level expected for overall intellectual abilities and relational language and pragmatics are clear weaknesses" (p. 99).

From a developmental perspective, the linguistic profile of individuals with WS is characterized by considerable delay in language onset and slow development over infancy and toddlerhood (Mervis & John, 2012), yet "relatively verbose, intelligible and fluent speech in late childhood and adulthood" (Krishan et al., 2015, p. 82). By the time children with WS reach school age, their expressive language, including vocabulary, syntactic forms and fluency appear to be relative strengths (Mervis & Becerra, 2007). However, these advances in structural language do not lead to adequate conversational and discourse abilities, despite the appearance of speech proficiency demonstrated in various contexts by most school-age children, adolescents and adults with WS. Using language efficiently in social contexts involves not just the availability of a well-developed linguistic system (e.g. varied lexicon, ability to comprehend and produce complex syntactic forms), but more importantly, it requires the ability to attribute mental states to the people involved in the communicative interaction, an aspect of social understanding to which I turn next.

6.3.2 Social Cognition in Williams Syndrome

For communicative exchanges to be successful, both the speaker and the hearer need to rely on a set of socio-cognitive skills commonly referred to as 'mentalizing abilities' or 'theory of mind' (ToM). These include the ability to infer the communicative intent of a speaker, based on attributing mental states to the communication partners, the ability to monitor the knowledge state and informational needs of the speaker and hearer, and the ability to perceive and interpret non-verbal cues such as facial expressions, eye gaze, and gesture to aid in the interpretation of verbal messages and of other communicative exchanges.[3] Impairments in any aspect

[3] Having knowledge about the social and conversational rules that apply to verbal interactions in one's culture, as well as other skills related to efficient use of contextual information, attention monitoring, planning, and the abilities usually subsumed under the label 'executive function' are

of these competencies may compromise the pragmatic processes that underlie the effective use of language in social contexts. Strong relations between ToM and social communication skills have been reported both for TD children (Astington, 1990; Tomasello, 1995) and for a range of clinical populations (Abbeduto et al., 2004; Hale & Tager-Flusberg, 2005; Losh et al., 2012; see Cummings, 2013 for a review).

Initially, based on their heightened motivation for social engagement and relatively good language skills, it was hypothesized that people with WS would have a good understanding of the social world, showing domain-specific sparing in social cognition or ToM (Karmiloff-Smith et al., 1995; Tager-Flusberg et al., 1998). One early study (Karmiloff-Smith et al., 1995) that included older individuals with WS (ranging from 9 to 23 years) reported that the majority of them passed standard first-order false belief and higher-order, ToM-related tasks, which are often failed by individuals with autism (based on prior studies). The authors concluded that ToM might be an "islet of preserved ability" in WS (p. 202). However, given the age of the individuals with WS tested and the lack of a matched, non-autistic comparison group (Brock et al., 2009; John et al., 2009), this interpretation remains problematic.

In fact, later studies, based on age-appropriate ToM tasks, and including appropriate control groups, found that the performance of children and adolescents with WS on ToM tasks was similar to that of participants with intellectual disability (ID) matched on age and IQ. In a series of studies probing systematically different types of mentalizing abilities, Tager-Flusberg and Sullivan (2000) found that participants with WS did not perform any better than a group of individuals with Prader-Willi syndrome (PWS) and a group of participants with non-specific aetiology of ID matched on age, IQ, and standardized language measures to the WS group. This was demonstrated on three different first-order false belief tasks, on second-order belief reasoning (Sullivan & Tager-Flusberg, 1999), on distinguishing between lies and jokes (Sullivan et al., 2003), and on using trait information to attribute intentionality (Plesa Skwerer et al., 2006). Similar findings have been reported when non-verbal ToM tasks, using a picture sequencing method, were used to assess understanding of pretence, intention or false belief (Porter et al., 2007; Santos & Deruelle, 2009). Thus, across a variety of studies and task formats (language-based or non-verbal ToM tasks), the performance of individuals with WS in social reasoning was no better than predicted by mental age.

While findings for tasks of social reasoning have been generally consistent across studies, there is less agreement about the social perception abilities of people with WS (Hepburn et al., 2011; Plesa Skwerer, 2017). Once more, researchers hypothesized that WS would be a paradigmatic case illustrating a dissociation between 'social cognitive and social perceptual components of theory of mind' (Tager-Flusberg & Sullivan, 2000). Based on the apparent emotional sensitivity of people with WS, it was assumed that, while having difficulties with inferential

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also critical in communication processes. However, given the paucity of research on WS addressing these areas, the discussion of findings presented in this chapter remains focused on the contributions of language and theory of mind-related abilities to pragmatic functioning in WS.

aspects of social information processing, individuals with WS would nevertheless show 'sparing' of social perception abilities, such as being proficient at recognizing facial and vocal expressions of emotion, skills important for social communication. Experimental studies, however, revealed that on explicit measures of emotion recognition from faces or voices, children and adults with WS performed no better than comparison groups matched on mental age on tasks of discriminating, matching, or labeling expressions of emotion (Gagliardi et al., 2003; Plesa Skwerer et al., 2006; Porter et al., 2007).

Overall, evidence from a growing number of studies involving different methodological approaches indicates that the ability to decode mental state information at the perceptual or at the cognitive, inferential level of mentalizing is impaired in WS. Such impairments, together with the distinctive language and personality profile characteristic of people with WS, likely shape their use of language in context as a social communicative tool. The following sections of the chapter provide a description of the features of pragmatic language that have been examined so far in individuals with WS, organized by the type of assessment or methodological approach used to evaluate pragmatic language in WS.

6.4 Assessing Pragmatic Language Abilities in Williams Syndrome

Pragmatic skills in children, adolescents and adults with WS have been assessed mostly in the context of cross-sectional research designs, involving groups of TD participants and several different clinical populations, including Down syndrome (DS), autism spectrum disorders (ASD) and specific language impairment (SLI). More recently, researchers have started to use longitudinal and developmental trajectory approaches to investigate syndrome-specific features of pragmatic development (John et al., 2012; Thomas et al., 2010) but these types of studies are still rare in the literature on WS. Relevant findings based on each type of methodological approach used in research so far are described next.

6.4.1 Caregiver Report-Based Measures

One of the assessment instruments commonly used to evaluate children's pragmatic abilities based on caregiver report is the Children's Communication Checklist (CCC; Bishop, 1998 and CCC-2; Bishop, 2003). The checklist includes 10 subscales, of which four comprise items directly related to pragmatic language behaviors (i.e. Inappropriate Initiation, Stereotyped Language, Use of Context, and Nonverbal Communication). The rest of the subscales comprise items related to structural language (four subscales) and autism symptomatology (two scales – Social Relations

and Interests). Based on this parent-rated assessment, Laws and Bishop (2004) found evidence of significant communication impairment in a sample of 19 children and young adults (aged 6–25 years) with WS relative to TD, DS and SLI control groups. In their study, 79% of the participants with WS scored in the range considered to be indicative of pragmatic impairment, compared to 50% of the participants with DS and 41% of the children with SLI. The WS group differed from the TD controls in all areas of pragmatic competence covered by the subscales – inappropriate initiation of conversation, coherence, stereotyped conversation, use of context, and development of conversational rapport – and scored worse than the two clinical groups in two domains: the use of stereotyped language and inappropriate initiation of conversation.

Using the revised version of the same instrument (CCC-2; Bishop, 2003), Philofsky et al. (2007) investigated the pragmatic language profiles of school-age children with WS and children with ASD, developmental disorders that are often considered to exemplify contrasting social phenotypes. When compared to parent ratings of the abilities of TD school-age children in the control group, both clinical groups showed communication impairment. Overall, the WS and ASD groups did not differ from each other on the General Communication Composite summary score (designed to identify communication impairment that could be based on either structural or pragmatic language deficits, or both). However, when considering the purely pragmatic subscales of the CCC-2, the children with WS appeared less impaired than the ASD group on Coherence, Stereotyped Language, Nonverbal Communication and Social Relations subscales, although no significant group differences were found on Inappropriate Initiation, Use of Context, and Interests subscales.

An examination of the item-level ratings on the CCC-2 showed that the WS children were considered by their caregivers as more skilled at use and understanding of affective expressions, prosody, learned phrases, social responsiveness, empathy and social relatedness with others, suggesting relative strengths in some areas of pragmatics compared to the children with ASD. These findings provide a complex picture of similarities and subtle differences in various aspects of pragmatic functioning across developmental disorders, pointing to types of communication impairments and strengths that may be syndrome-specific. Further research, using a combination of assessment measures and contexts, is needed to be able to determine whether particular features of pragmatic language differentiate individuals with WS from other neurodevelopmental disorders, while accounting for general cognition as well as for structural language level (Martin et al., 2017).

6.4.2 Standardized Direct Assessments of Pragmatic Skills

Few studies have tested directly the pragmatic abilities of people with WS using individually-administered standardized tests of pragmatic language. These types of instruments usually require the responder to make judgments about social situations

based on brief stories accompanied by pictures, or pictures alone. One standardized test that has been given to children and adolescents with WS is the Test of Pragmatic Language-2 (TOPL-2; Phelps-Terasaki & Phelps-Gunn, 1992, 2007), which probes pragmatic reasoning in several domains relevant to successful communication (e.g. abilities to appraise and monitor physical context, audience, topic, visual-gestural cues, purpose/speech acts and abstraction). Hoffmann et al. (2013) compared directly the performance of school-age children and adolescents with WS (aged 8–16 years) on the TOPL-2 with parent ratings of the same participants' communication abilities on the CCC-2 (Bishop, 2003). While there were no significant differences in terms of overall scores between the TOPL-2 and the CCC-2, the two types of assessment yielded different results with respect to the number of participants each classified as having pragmatic language impairment (PLI). In particular, the TOPL-2 identified significantly more participants with WS as meeting cut-off for PLI than did the CCC-2 instrument (70% vs. 30%, respectively).

This pattern of results was in direct contrast to findings reported by Volden and Phillips (2010) for a group of children with ASD who had age-appropriate structural language skills. Using the same instruments, they found that the CCC-2 classified significantly more of the children with ASD as having PLI than did the original TOPL (Phelps-Terasaki & Phelps-Gunn, 1992) assessment (81% versus 56%, respectively). Whether the reason for the discrepant findings in the two studies is related to syndrome-specific aspects of pragmatic impairment or to other factors remains unclear. The TOPL-2, a test of a participant's ability to "view a social situation as an objective bystander" (Hoffmann et al., 2013, p. 200), draws on meta-pragmatic skills, and may rely more heavily on social cognitive rather than linguistic abilities, which may have rendered the test more challenging for the cognitively impaired children and adolescents with WS.

Given the differences in the rate of PLI detected among the WS participants by the two assessment instruments, it is critical that future studies of pragmatic skills involve multiple methods and measures in order to provide a comprehensive evaluation of pragmatic functioning in this population. Evaluations that include more ecologically valid contexts of assessment, based on quantitative and qualitative analyses of real-life interactions with different types of communication partners, both familiar and unfamiliar, should complement standardized and caregiver-report measures. Semi-structured conversations with researchers and narrative elicitation tasks have already provided a wealth of information about the distinctive communication style exhibited by people with WS, while revealing a checkered picture of strengths and weaknesses in their socio-communicative skills.

6.4.3 *Elicited Narratives and Conversation*

Early reports of unusual language proficiency in WS were based on comparing narratives produced by a small group of adolescents with WS to those of two comparison groups: an age-and IQ-matched group of adolescents with DS and a group of younger, mental age-matched TD children. In the first study to use a wordless picture book – *Frog, Where Are You?* (Mayer, 1969) – to elicit narratives from four adolescents (aged 10–18 years) with WS, Reilly et al. (1990) reported that the WS participants showed more prosodic features of speech (e.g. instances of pitch changes, vocalic lengthening, modifications in volume) than did either comparison group, and produced more evaluative language (including references to the emotional and mental states of story characters, character speech or sound effects, exclamatory phrases, and emphatic markers meant to capture the attention of the listener) than did the DS group.

Follow-up studies using the same picture book (Losh et al., 2000; Reilly et al., 2004) compared larger groups of school-age children with WS to age-matched groups of TD children and to children with specific language impairment (SLI). These studies found a similar abundant use of evaluative language in the narratives produced by the children with WS relative to either the SLI or the TD groups. However, the children with WS made significantly more grammatical errors than the TD group, and their grammatical performance was no better than that of the children with SLI. An analysis of the type of evaluative devices used by the children with WS indicated a higher prevalence of 'social engagement device' (e.g. sound effects, character speech, and "audience hookers"), but fewer cognitive inferences than in the narratives of the comparison groups.

Several researchers have reported a lower use of linguistic devices that contribute to narrative coherence and cohesion (cohesive ties, grammatical markers, and complex syntactic structures) in stories told by children with WS, relative to both TD and SLI comparison groups, even when individuals with WS produced longer narratives overall. On narrative measures that tap cognitive inferencing skills, such as story structure and integrating themes, children with WS usually scored significantly lower than comparison groups across narrative elicitation studies in several languages (Diez-Itza et al., 2018; Lacroix et al., 2007; Lorusso et al., 2007; Losh et al., 2000; Stojanovik et al., 2004). These findings suggest that the excessive use of social engagement devices in narratives, which has been observed across different languages and cultures, including English, French, Spanish, Italian, Dutch and Portuguese (Reilly et al., 2005; Gonçalves et al., 2010; Jones et al., 2000; Lacroix et al., 2007; Van Den Heuvel et al., 2016), may mask difficulties with both macrostructural and cognitive aspects of narrative performance in WS, while reflecting the most enduring characteristic of the children, adolescents and adults with this syndrome – their heightened sociability (Losh et al., 2000; Järvinen et al., 2013). The contrast between the strong desire to engage an audience and the ability to do so competently is also reflected in the tendency of individuals with WS to use an abundance of affective prosody and social evaluation even in the second

story-telling instance to the same listener, or when not being paid attention to, which suggests a lack of pragmatic sensitivity, because the speaker with WS does not take into account the state of the listener (Järvinen-Pasley et al., 2008).

The strong motivation to keep a social interaction going and to capture the attention of a conversational partner seems to shape the speech style exhibited by people with WS across a variety of situations and often irrespective of their familiarity with the interlocutor. People with WS tend to engage conversationally with strangers just as readily as with people they know very well, and rarely adjust their style of speech based on what they know about the person they are speaking with (Järvinen-Pasley et al., 2010; Jawaid et al., 2012; Jones et al., 2000). For instance, several researchers provided anecdotal reports of participants with WS 'turning the tables' on the experimenter during interviews and asking personal questions, often accompanied by poor turn-taking and topic maintenance (Jones et al., 2000; Harrison et al., 1995; Semel & Rosner, 2003; Stojanovik et al., 2001). Udwin and Yule (1990) remarked on the conversational inadequacies demonstrated by a significant proportion of school-age children with WS in naturalistic interactions. Based on 30 min of conversation with a researcher, they found that 37% of the children with WS in their study met criteria for hyperverbal speech (also referred to as 'cocktail party speech'). This involves fluent speech with an excessive use of stereotyped phrases, an overfamiliar manner, introduction of irrelevant personal experiences and perseverative responding.

Several other studies included qualitative analyses of dyadic conversational interactions conducted mainly with researchers as the conversational partner. It should be noted that these studies were either case-reports or included a small number (4–12) of participants with WS. Stojanovik (2006) used semi-structured conversations around photographs depicting everyday situations to probe the abilities of five school-age children with WS to talk about their own experiences related to the topic, and to respond to the researcher's questions. Compared to a group of children with SLI matched on receptive vocabulary and grammatical ability, and to a group of TD peers, the children with WS were less likely to give adequate responses to the interlocutor's request for information or clarification, or to produce responses that would advance the conversation, even though they were likely to produce extended responses (e.g. more than yes/no replies). However, significantly more of their utterances were inadequately informative, providing too little information or misinterpreting what the speaker meant, and tending to overrely on the conversational partner's lead.

Similar findings have been reported for French-speaking children with WS observed in parent-child interactions or in conversation with an examiner. Lacroix et al. (2007) found that, compared to TD controls, their participants with WS showed less turn-taking in conversations, more non-contingent responding, and provided less information in response to an adult's request for clarification. While these studies provide generally consistent descriptions of the deficits shown by people with WS in conversation across several languages, the predictors and correlates of the pragmatic language difficulties reported remain to be systematically investigated.

Recently, Rossi and Giacheti (2017) examined associations between speech-language, general cognitive functioning (IQ) and behavior problems in a sample of Brazilian children and adolescents with WS (aged 8–18 years). Besides administering tests of receptive vocabulary, syntactic comprehension, IQ, and a parent questionnaire involving ratings of behavioral problems (Child Behavior Checklist-CBCL; Bordin et al., 2001, 2013), the authors analyzed 30-minute conversations with a researcher, which were coded for five speech-language characteristics considered to be indicative of pragmatic difficulties: clichés; echolalia; perseverative speech; exaggerated prosody; and monotone speech. They reported that 61.5% of the participants with WS showed perseverative speech in conversation, while 50% used exaggerated prosody, 42.3% used clichés, and 19.2% used echolalia during the conversation task. On the CBCL questionnaire, 73.8% of the participants were rated by their parents as 'talking too much'. The two types of repetitive verbal behavior coded during conversation – perseverative speech and echolalia – were negatively correlated with performance on standardized assessments of language, while exaggerated prosody use in conversation was associated with higher ratings of problem behaviors on the CBCL (Total problem scale).

These associations between lower language performance, increased reliance on prosody and paralinguistic devices in conversation, and behavioral dysregulation in individuals with WS may provide insight into why so many people with WS experience serious difficulties in forming and maintaining social relationships, especially with peers, despite their strong need for social contact and relatively proficient expressive language. Hargrove et al. (2013) analyzed spontaneous conversations between adolescents with WS and age-matched TD peers with respect to their use of paralinguistic features such as laughter, sound effects, fillers and repetitions/reformulations. While the TD peers produced significantly more fillers and reformulations than the adolescents with WS did, the WS group used an abundance of sound effects and laughter, which may be used to mask difficulties with the content of their conversations: while superficially engaging, their contributions to topic maintenance were largely irrelevant. As these authors commented, 'What may be engaging or "cute" in younger children could be viewed as awkward or immature by adolescent communicative partners', and may end up 'isolating the adolescents with WS from peers rather than engaging them' (p. 157).

In summary, at all ages, people with WS tend to engage in conversation readily and enthusiastically, but often lack the tools to initiate and maintain a meaningful conversational exchange. In a variety of discourse contexts, such as storytelling, picture descriptions, and biographical interviews involving questions about participants' interests, family, or everyday activities, children and adolescents with WS were more likely than control groups to use more adult vocabulary, social phrases and an overly-familiar conversational style. Their exaggerated use of prosodic and paralinguistic features of speech, instead of reflecting expressive communication competence, may serve mainly a social function: this speech style appears to be an attempt to compensate for the inability to contribute meaningfully to the substance of a conversation, while trying to hold the audience engaged in the interaction. Over time, this characteristic conversational style may have the opposite

effect of that intended by the speaker with WS, and might, in fact, contribute to the serious difficulties that people with WS have with developing friendships and meaningful social relationships, especially as they age.

6.4.4 Experimental and Laboratory-Based Tasks

Recently, researchers have started to use experimental tasks to deconstruct the complex pattern of pragmatic deficits demonstrated by people with WS across naturalistic discourse contexts. Such tasks enable investigators to focus on discrete pragmatic skills (e.g. signaling non-comprehension of a message, providing information to clarify a message or conversational repair skills) and may lead to identifying particular sources of pragmatic deficits and strengths. So far, two areas of pragmatic competence have been probed experimentally in individuals with WS: referential communication and interpretation of non-literal language.

In the first study to examine the referential communication abilities of children with WS, John et al. (2009) focused on the listener's role. They investigated whether 6- to 12-year-old children with WS were able to recognize and verbalize the referential problem when a communication partner's messages were inadequate. In the experimental setting used (modeled after Abbeduto et al., 2008), the child and the experimenter were separated by an opaque barrier, and the child was required to identify a referent (a picture) from an array of items based on the information conveyed by the speaker. The researchers examined children's verbal responses to messages that were ambiguous, or included a word the child did not understand, or conveyed an impossible action because the requested item was not available.

The children with WS indicated that there was a problem with the speaker's message less than half the time, and they had difficulty verbalizing the nature of the referential problems encountered. The type of message inadequacy influenced their responding. They performed better in the 'impossible condition', reporting the problem 55% of the time, whereas they communicated a problem with an ambiguous message or with a message containing an unknown word only about 20% of the time. Children's performance on false-belief tasks and age were significant predictors of the likelihood and effectiveness of their verbalizing that a speaker's message was inadequate in the more difficult conditions (ambiguous and unknown word). The strong relationship found in this study between theory of mind and listener-role referential communication skills in children with WS is consistent with findings from research on TD children and on individuals with other neurodevelopmental disorders (Abbeduto et al., 2004; Losh et al., 2012; see Matthews et al., 2018 for a review), adding to the evidence of close connections between pragmatic skills and social cognitive skills across typical and atypical development.

Asada et al. (2010a) focused on the 'speaker role' in a study involving an object-choice situation used to elicit verbal requests and corrections/reformulations from the children engaged in the task. These researchers examined the communication

repair skills of 4- to 11- year-old children with WS relative to those of MA-matched TD children. After the children indicated their choice of a preferred object, the experimenter proceeded to give them verbal feedback (i.e. whether the child's choice was understood or misunderstood) and one of the objects (i.e. the desired or undesired object). Thus, four conditions ensued from the combination of the experimenter's verbal feedback and object-giving action. While the overall frequency of verbalizations across the four conditions was similar across groups, children with WS produced fewer requests and rejections of the wrong object than did the TD group, particularly when they were verbally misunderstood. In contrast to the TD children who increased their corrections in the verbal misunderstanding condition compared to the correct understanding condition, the children with WS did not vary their verbalizations for clarification based on condition. Their behavior suggested that they often failed to take into account the perspective of the interaction partner, even when it was explicitly mentioned verbally. These results indicate that children with WS have difficulty in using communication repair skills, such as providing verbal corrections in order to share what they meant with others, to establish mutual understanding.

In another study based on a modified referential communication paradigm, Plesa Skwerer et al. (2013) probed experimentally the ability of children with WS to monitor the informational adequacy of both the communication partner's message and of the children's own verbal messages. The researchers used a collaborative game format, in which a child and a researcher took turns in placing small objects on a large mat illustrated with their pictures to 'build' a toy-size 'farm' or 'wildlife park'. The game partners had to indicate to each other the objects they needed help with (e.g. for those that were outside their reaching space), and to collaborate in the selection of the items. Some items were of the same identity but distinguishable by a relevant attribute (e.g. white bear and brown bear) while others were unique. When in the 'builder'/speaker role, the researcher sometimes expressed insufficiently informative indirect requests for items of the same type (e.g. "I think I need the bear now") directed toward the 'helper'/hearer.

The children with WS (ranging in age from 5 to 13 years) performed significantly worse than a group of TD children matched for chronological age (CA), but similarly to a group of TD controls matched for verbal mental age (VMA) on quantitative measures of pragmatic comprehension (i.e. number of clarification requests) when in the 'helper'/hearer role. However, they were less likely than both comparison groups to take into account the available visual information about the objects (i.e. to look both in the partner's and in their own space) before interpreting the intended referent of the partner's request. When they *did* realize that the speaker provided insufficient information to help them distinguish between objects of the same type, the children with WS were more likely to use pointing or picking up and showing one of the objects than to verbalize their request for clarification. In contrast to this reliance on gesture and guessing, the TD children showed a developmental trend toward using increasingly definite verbal questions to obtain the needed information. Another tactic used by the children with WS when confused about the partner's message was to comment or ask personal questions unrelated to

the collaborative game. This exchange illustrates such a common situation. The experimenter makes an indirect and ambiguous request for a non-unique item (there are two toy dogs similar in shape and color but different in size):

> Experimenter: I need the dog now…
> Child with WS: (after a brief pause, looking at E's hand) Oh, I know what your ring means. You are married, right?

When the participant played the role of the 'builder'/speaker, the children with WS provided fewer adequately informative referents (i.e. mentioning the distinguishing feature of a non-unique item) than both the CA and the VMA controls did, and significantly fewer of them provided adequately informative referents on all the relevant trials (Ammerman, 2013). However, as the game progressed, learning effects were observed for the WS children, who did not differ from either comparison group in conversational repair attempts (offering appropriate information when notified that the listener needs more information). As the game continued, the children with WS were just as likely as the control groups to improve their responding to the feedback provided by the game partner, and to repair communication breakdowns, especially after the partner modeled a clarification response. This finding suggests that scaffolding could have beneficial effects for children with WS, who appear to be able to learn from specific feedback and improve their communication strategies during social interaction.

Longitudinal studies are needed to determine whether such gains in communication adequacy remain temporary and are circumscribed to the context of the particular interaction or may endure and contribute to lasting improvements in pragmatic functioning. So far, only one study has examined longitudinal relations between aspects of pragmatic behavior demonstrated by 4-year-old children with WS during a play session with their mother, and their conversational ability about 6 years later. John et al. (2012) found that the ability to verbally contribute new information within a social interaction at age 4 years showed stability from preschool to primary school age in children with WS. Differences in this pragmatic skill at school age were predicted by children's ability to pair verbalizations with eye contact in triadic interactions (secondary intersubjectivity) as preschoolers. When taken together with the previously mentioned findings about the potential role of scaffolding for improving communicative strategies in WS, these longitudinal findings underscore the importance of establishing pragmatic language outcomes as intervention targets for young children with WS, to maximize their beneficial effect for acquiring and consolidating pragmatic skills in this population.

For effective communication speakers are also required to consider the attentional focus of their interlocutor, to ensure that their message is processed as intended. Typically developing children by age two are able to track speakers' intentions and to take into account what they attend to, or what they know in a particular situation (O'Neill, 1996; Moll & Tomasello, 2006). Asada et al. (2010b) investigated the ability of children with WS (mean CA = 10; 2 years) to evaluate the attentional focus of a partner in communication and to modify their verbal message accordingly with a view to sharing information. The children in their study were

asked to complete simple actions with toys while the experimenter either attended to them or did not pay attention to them during the time they accomplished the task.

While a comparison group of TD children who were individually matched on vocabulary age to the participants with WS verbalized more about their accomplishment when they were not attended to than when they were attended to, the children with WS showed the opposite pattern, meaning that their verbalizations did not take into account the attentional state of the listener. The authors interpreted the verbal behavior of the children with WS as a violation of the pragmatic rule "to speak the appropriate amount according to other's state" (Asada et al., 2010b, p. 456). They suggested several possible explanations for the atypical verbal communication pattern demonstrated by the children with WS, including possible impairment in the ability to direct or share attention to something (i.e. declarative function deficit), or impaired understanding of the 'seeing-leads-to-knowing' principle (socio-cognitive deficit), or higher motivation to interact verbally when they are attended to, regardless of the goal of the communication. More research is needed to disentangle the potential contribution of each of these factors to pragmatic deficits, or to clarify how their combination may impact the quality of communication efforts in people with WS.

In summary, across different types of referential communication tasks, it appears that children with WS have difficulties with a set of processes involved in efficient communication, including evaluating the informational adequacy of messages, requesting and providing verbal corrections or clarifications when needed, understanding the attentional focus and state of the hearer and, more generally, evaluating and taking into consideration the perspective of another during a social-communicative interaction. This promising line of research should be continued with investigations of whether and how each of these discrete pragmatic behaviors and processes may be turned into targets for interventions aimed at improving pragmatic functioning in people with WS.

6.4.5 Non-literal Language Comprehension and Production

Comprehending and using figurative language are important aspects of pragmatic competence because they involve the ability to distinguish between intended meaning and the 'surface' expression (literal meaning) in the process of utterance interpretation. This process can be challenging as it usually relies on background knowledge and the ability to draw links and find similarities between often conceptually disparate domains (Keil, 1986). Not surprisingly, comprehension and production of figurative language represent late achievements in typical development, continuing to progress throughout childhood (see Falkum, 2019 for a review). Several studies have examined comprehension of non-literal language in WS, including metaphors and metonyms (Annaz et al., 2009; Van Herwegen et al., 2013), perceptual simile (Thomas et al., 2010), idiomatic expressions (Mervis et al., 2003; Lacroix et al., 2010), irony (Sullivan et al., 2003), jokes (Krishan et al., 2017;

Sullivan et al., 2003) and sarcasm (Karmiloff-Smith et al., 1995; Godbee & Porter, 2013).

The majority of evidence from these studies points to significant delays shown by individuals with WS in interpreting language in context when the intended meaning differs from the literal meaning. Some researchers have suggested that comprehension of different forms of non-literal language by individuals with WS follows an atypical developmental trajectory. Across several studies, it was found that comprehension of metaphor and novel metonymy was not only delayed but did not increase with increasing chronological age in WS, in contrast to the pattern of performance of the TD participants (Annaz et al., 2009; Thomas et al., 2010; Van Herwegen et al., 2013). This seems at odds with reports of increasingly rich expressive language used by individuals with WS as they age. As will be discussed later, many individuals with WS do *produce* figurative language, especially as they increase their vocabulary knowledge, but it is possible that they do so without understanding its meaning (Bertrand et al., 1994; Thomas et al., 2010).

Being able to interpret appropriately the communicative intent implied in various forms of non-literal language such as jokes, ironic statements, lies and sarcastic remarks is important for relating to peers in everyday discourse. Sullivan et al. (2003) tested the ability of adolescents with WS to distinguish between lies and ironic jokes, using a series of short stories in which a character's final statement was false, but the communicative intent was either deception/lying or irony/joking. Adolescents with WS and two comparison groups, one of adolescents with PWS, the other of adolescents with non-specific aetiology of ID, matched on age, IQ and verbal abilities to the WS group, were asked to classify the characters' statements as lies or jokes, and to justify their responses. While almost all of the participants in the three groups were unable to identify which intentionally false utterances were intended as ironic jokes, classifying them as lies instead, the WS group differed from controls in their justification responses. They usually referred back to the facts of the story, instead of using mental state-based explanations, as did the control groups, which suggests that social-cognitive impairments may be a major source of these adolescents' pragmatic deficits in interpreting non-literal language.

Godbee and Porter (2013) presented stories in which characters made non-literal comments that were either sarcastic in intent, or voicing a metaphor or a simile, to participants with WS (ranging in age from 5;4 years to 43;8 years) and to TD controls who were individually matched either for mental age (MA) or for chronological age (CA) with the WS participants. In this study, the individuals with WS performed worse than their CA-matched controls in explaining 'what did the story character mean' on all forms of non-literal language comprehension, but their performance was not significantly different from that of the MA-matched controls, although sarcasm comprehension was particularly poor (at floor) in the WS group.

Only one study to date has focused on humor comprehension, in relation to mental state language use, in adolescents with WS and those with DS. These adolescents were compared to two groups of TD participants, one matched on MA and another matched on CA to the clinical groups. When asked to explain 'what was funny?' about 23 humorous cartoons, both groups with intellectual disabilities obtained

scores indicating lower levels of humor comprehension than the CA controls, but were not different from each other, or from the MA controls, either in humor comprehension scores or in total mental state language use. These results also indicate that the relatively better expressive language abilities of the older participants with WS compared to other individuals with intellectual disability such as DS, PWS or much younger TD participants, did not play a compensatory role in their performance on tasks of non-literal language comprehension.

Using a developmental trajectory approach, Naylor and Van Herwegen (2012) found that, during a fictional narrative task, 7–18 year-olds with WS *produced* a similar amount of figurative expressions as did a TD comparison group, an apparent contrast to the poor performance usually demonstrated on tasks of figurative language comprehension. Semel and Rosner (2003) also noted the use of idioms and figurative language by individuals with WS during conversations. What could explain this discrepancy between understanding and production of figurative language by individuals WS? Anecdotal reports as well as qualitative analyses of conversations (Udwin & Yule, 1990; Jones et al., 2000) have suggested that their use of idioms, social phrases and various forms of figurative language often appears to be somewhat inappropriate to the social context, raising doubts about their conceptual understanding of the expressions produced (Bertrand et al., 1994). Parents of individuals with WS have also reported instances of using language they clearly do not understand, which may explain the impression of unusual vocabulary (e.g. low-frequency word choices) that has been anecdotally reported to be a distinctive feature of the speech produced by adolescents and adults with WS (Bellugi et al., 1992; Rossen et al., 1996).

Thomas et al. (2010) suggested that using low-frequency words, or peppering their speech with clichés, idioms and figurative language they have previously heard and memorized, may be ways in which individuals with WS attempt to capture the attention of an 'audience' and to keep the social interaction going. In this case, the production of figurative language may be a pragmatic device serving social ends for individuals with WS and may not reflect the conceptual understanding required by a meaningful use of these forms of speech. In short, the findings reviewed so far strongly suggest that people with WS tend to enroll all the linguistic tools available to them in the service of social engagement, sometimes at the expense of a meaningful and socially appropriate use of verbal content in their communicative attempts.

6.5 Developmental Precursors and Correlates of Pragmatic Skills

Why is pragmatics an area of language functioning that is particularly problematic for children, adolescents and adults with WS? High interest in social engagement and relatively good structural language achieved by school age, despite a delayed

onset, would be expected to sustain and bolster the development of pragmatic skills. To gain insight into the possible origins of pragmatic deficits in WS, we need to understand the developmental relations among a complex set of abilities that interact across developmental time (Karmiloff-Smith, 1998), resulting in the profile of pragmatic language functioning described in this chapter. This set of relevant skills and processes involve early socio-communicative behaviors emerging in infancy and their relationship with later developments in the structural aspects of language, as well as in social perception and social cognition, attention monitoring processes and executive functioning, behavior regulation abilities and learning processes underlying the acquisition of cultural knowledge. There has been little or no research involving individuals with WS in some of these areas, and studies of infants or young children with WS are still scarce. However, a number of key findings that have emerged from recent research could shed light on the developmental origins of the pragmatic language profile associated with this intriguing neurodevelopmental disorder.

Pragmatic language shows a protracted development in TD individuals and many skills continue to emerge through adolescence and beyond. However, a variety of pragmatic skills emerge early in life, starting with pre-verbal turn-taking around 8–9 months (Ninio & Snow, 1996) followed by a sequence of processes closely related to social interaction. In describing this sequence, Adams (2002) notes that "early social exchanges revolve around objects which are the focus of joint attention followed by rapid development of communicative acts between 14 and 32 months" (p. 975). By contrast, from early in life, the attention of infants with WS revolves almost exclusively around the people they interact with, as reflected in their atypical eye contact and limited gaze following away from the partner's face during social exchanges. In one of the first studies to focus on social interactive behaviors in WS, Mervis et al. (2003) observed that a 10-month-old girl with WS displayed unusually prolonged and intense-looking behavior toward her play partner (mother or unfamiliar adult) compared to both developmental-age and chronological-age matched TD female infants.

Similar unusual eye contact was reported in other contexts where infants, toddlers and young children with WS directed their attention almost exclusively to the people present, at the expense of sharing and coordinating attention between their social partners and surrounding objects and events (Thurman & Fisher, 2015; Mervis et al., 2003; Laws & Bishop, 2004). During semi-structured interactive assessments of early socio-communicative abilities, such as the Early Social Communication Scales (ESCS; Mundy & Hogan, 1996), toddlers with WS showed less object-related behaviors (declarative and instrumental pointing, reaching, requesting toys and response to joint attention bids) than a group of TD toddlers matched on developmental age (Laing et al., 2002). While they engaged readily in dyadic interactions and used more social interactive behaviors (requests for tickling, turn-taking behaviors, eye contact not related to objects) than the control group, the toddlers with WS showed significant impairments in triadic interactions relative to MA-matched controls. Eye contact was used more in dyadic interaction and less for

social referencing or in combination with requesting or reaching behaviors by the toddlers with WS compared to controls.

This finding was corroborated by a later study involving preschoolers with WS (Thurman & Mervis, 2013), who were compared to age- and gender-matched children with DS in their social-referencing behaviors and its associated component abilities – initiating eye contact, gaze following and emotional responsivity. More specifically, Thurman and Mervis (2013) found that children with WS were less likely to initiate eye contact (unsolicited) and to follow another person's gaze in triadic situations than were children with DS, although both groups showed difficulty utilizing the communicative significance of facial expressions of fear in social-referencing processes.

Similar difficulties with both initiation of, and response to, joint attention were demonstrated by children with WS during the administration of the Autism Diagnostic Observation Schedule (ADOS; Lord et al., 1999) – a semi-structured standardized assessment designed to elicit behaviors that are directly relevant to the diagnosis of Autism Spectrum Disorder (ASD). The ADOS involves a series of interactive activities, appropriate for a child's developmental level/language and age that create opportunities for observing and evaluating joint referencing, social relatedness, communication skills, and restricted and repetitive behaviors. Several studies in which individuals with WS were administered the ADOS (Klein-Tasman et al., 2007; Lincoln et al., 2007; Tordjman et al., 2012) indicated some overlap in their socio-communicative functioning with that of children with ASD. While the proportion of children with WS who met the cut-off for a classification of ASD differed across studies (ranging from 10% to 50%), reports of abnormalities demonstrated by children with WS in the use of gestures, declarative pointing, initiating joint attention and showing objects were consistent across studies. At the same time, compared to the children with autism, those with WS showed relative strengths in their quality of social overtures, social smiling and directing facial expressions and vocalizations to another.

Besides the prelinguistic, socio-communicative difficulties described above, children with WS have been reported to begin to talk before they begin to either point or to show objects in triadic interactions, which is an atypical developmental sequence. The use of referential language prior to the onset of communicative gesture use has been described both in longitudinal and in cross-sectional studies of infants with WS, and has been observed both by parent report and in structured laboratory settings (Laing et al., 2002; Mervis & Bertrand, 1997; Singer-Harris et al., 1997). Even preschoolers with WS demonstrate a lack of ability to use gaze-shift to infer a partner's communicative intent (John & Mervis, 2010) or to evaluate the attentional focus of a partner (Asada et al., 2010b). Researchers have pointed out that this pattern of relations between prelinguistic and linguistic developments in WS appears to be atypical not just relative to the normative trajectory mapped for TD children, but also relative to the sequence of communicative developments found in other neurodevelopmental disorders, including DS and ASD (Mervis & John, 2010). The impact of this atypical course of communicative development on the acquisition of pragmatic skills in WS remains to be investigated systematically.

In summary, the pragmatic difficulties demonstrated by individuals with WS are likely to be rooted in the delays shown by young children with WS in establishing joint attention and secondary intersubjectivity during social interactions. Such difficulties and delays in the use of eye gaze, gesture and directing and sharing attention around objects and events, in combination with the failure to use communicative gestures to express intentions prior to the onset of language, are likely to trigger cascading effects on later socio-communicative developments and, in particular, on the trajectory of pragmatic skill acquisition in WS.

6.6 Clinical Implications

As noted at the beginning of this chapter, early research reports on individuals with WS emphasized their 'striking language proficiency' despite their cognitive deficits, so it is not surprising that the need for interventions targeting the use of language in social contexts by people with WS was not fully recognized until fairly recently. Even though teacher- and parent-report descriptions of 'incessant chatter', 'an old fashioned and formal style of speech, including the use of stock phrases' (Udwin et al., 1987, p. 306), 'poor turn-taking and topic maintenance, inappropriate responses, repetitive phrases and hyperverbalization' (Meyerson & Frank, 1987, p. 260) have been noted in some of the earliest published studies about WS, clinicians and speech therapists rarely prioritized addressing these speech peculiarities until recently, when their impact on the social-adaptive functioning of individuals with WS has started to be recognized and documented (Howlin et al., 2010).

Pragmatic abnormalities often have negative consequences on social skills and interpersonal relationships, interfering with the ability of individuals with WS to engage with peers and to participate in age-appropriate social activities, and may even put them at risk for social victimization (Elison et al., 2010; Jawaid et al., 2012). Understanding and taking into account how pragmatic deficits impact the adaptive skills of individuals with WS is a necessary step toward establishing targeted goals for intervention. Because recent research has uncovered significant heterogeneity in abilities among individuals with WS, despite the many common strengths and challenges discussed so far, it is possible that different factors, learning and social experiences may differentially contribute to progress in pragmatic language development for different children with WS. As Mervis and Velleman (2011) suggest, to determine the intervention needs of a child with WS, the child needs to be carefully observed in interaction with caregivers, teachers and other children in a variety of settings, in addition to conducting formal assessments of the child's language level.

This is important for several reasons. From an intervention-planning standpoint, it is critical to take into account the atypical developmental sequence between the onset of referential communicative gestures and referential expressive language in WS. It may not be the case that once a child with WS is already talking, "basic

referential gestures have been mastered" (Mervis & Velleman, 2011, p. 100), as is the case in typical development. If this misleading assumption is made, early intervention planning may miss opportunities to address some of the non-verbal skills (e.g. ability to establish triadic joint attention, comprehension and production of communicative gestures) that are essential for the development of pragmatic language competence. Also misleading may be the fluency and good articulation that many children with WS demonstrate once they have acquired basic expressive vocabulary, because these verbal characteristics are sometimes taken to signal that speech/language therapy is no longer needed. Such verbal strengths may in fact mask serious difficulties with the pragmatic aspects of communication, which can persist into school years and later in life, and are likely to hinder successful functioning in the social world.

In one of the first comprehensive books published about WS, Semel and Rosner (2003) suggested several practical goals for interventions aimed at improving language pragmatics. They indicate that mediational strategies could be effective in modifying behavior such as, for instance, inappropriate greeting behaviors, which are displayed by individuals with WS "with almost every new person they meet" (p. 89), inappropriate requests for attention, persistent questioning, topic perseveration or poor turn-taking/turn-yielding. These strategies involve pointing out the social role differences of various types of people (e.g. strangers, acquaintances, family members, service people, professionals, etc.) when encountered in various contexts, and modeling appropriate behaviors (e.g. role-playing alternative ways of communicating, videotaping mock situations and having the instructor provide specific feedback, training to use "self-talk" to help restrain from compulsive greeting, etc.). Semel and Rosner (2003) suggest that interventions using "modeling, role playing, puppetry, playacting, or improvisational dramatization" (p. 94) may be especially effective, given the personality characteristics of people with WS, who tend to be dramatic in their emotional expressions and to crave social praise.

One of the most salient characteristics of the behavioral phenotype of people with WS is an openly declared, strong love for music (Thakur et al., 2018). Their affinity for music and rhythm may be used toward therapeutic goals, such as improving the quality and structure of conversational exchanges (e.g. teaching turn taking/turn yielding and the appropriate use of prosodic features of speech). The use of music in interventions targeting pragmatic language outcomes may be a particularly beneficial approach for individuals with WS. Conducting studies to evaluate systematically the efficacy of various intervention protocols aimed at improving pragmatic functioning in people with WS should be a priority for future research.

6.7 Directions for Future Research

This review of what is currently known about the pragmatic language profile of people with WS suggests several potentially interesting avenues for future research. First, identifying the predictors of progress in pragmatic language development in WS should be an important research goal because this knowledge could contribute directly to informing the design of pragmatic language intervention protocols for this population. This effort implies a systematic investigation of the developmental relationships between particular types of pragmatic processes and the trajectories of structural language, sociability, and non-verbal cognition as they interact across developmental time in individuals with WS. So far, in addition to extensive investigations of linguistic abilities in WS, research has focused mainly on socio-cognitive (ToM) and socio-perceptual processes that may directly influence the acquisition of pragmatic skills. But very little is known about how executive functions and inferencing abilities, memory processes, behavior regulation skills or the acquisition of cultural knowledge (e.g. learning about, and internalizing socially appropriate norms and rules of communication) contribute to particular aspects of pragmatic functioning in people with this intriguing syndrome.

Second, it should be noted that there are still major gaps both in the phenotypic characterization of the pragmatic profile of people with WS, and in understanding the course of development of pragmatic skills in WS. As described in this chapter, interest in pragmatic language in WS has surged in the last decade, yet many aspects of pragmatic functioning, including different types of speech acts, conversational implicatures and discourse processes remain to be investigated systematically in this population. Research on the trajectory of pragmatic skill acquisition and its complex relations with other features of cognitive and behavioral functioning in WS is extremely limited. Therefore, longitudinal studies with larger samples or studies based on cross-syndrome comparisons of developmental trajectories of particular abilities are much needed in the field of neurodevelopmental disorders research. Such studies are critical for identifying syndrome-specific phenomena, including potential specificity in the social uses of language, and may open avenues for eventually linking genetic abnormalities to brain development and to behavioral outcomes.

Although recently researchers have acknowledged the substantial individual variability within domains of skills found in the WS population, little is known about possible gender differences in pragmatic language in WS. A similar discussion of potential cultural differences in the pragmatic profiles of people with WS is timely. Previous cross-cultural research on perceived sociability and on narrative production in children with WS has revealed a rich set of similarities and differences in phenotypic profiles in WS across languages and cultures (Zitzer-Comfort et al., 2007; Reilly et al., 2005). Conducting cross-cultural research using a multitude of assessment instruments, as well as through naturalistic observations in a variety of different contexts, could be a particularly useful avenue for distinguishing between syndrome-specific and cultural/linguistic influences on pragmatic functioning in

people with WS. Using multiple methods and conducting research across different settings and time points could provide valuable insights not only for understanding the development of pragmatic language and social communication in WS, but also for designing and timing appropriately the implementation of interventions, in order to maximize their beneficial effects for the adaptive, social and even emotional functioning of people with WS.

6.8 Summary

Despite its relatively short history, research on pragmatic language skills in WS has addressed a number of important pragmatic concepts, including communicative intentions, reference resolution, informational adequacy of messages and clarification requests, conversational repair strategies, non-literal language, and a range of discourse functions examined in narrative tasks and in dyadic conversations. This chapter reviewed some of the key findings and methodological approaches used to investigate a number of discrete pragmatic skills and behaviors demonstrated by children, adolescents and adults with WS in several settings, including lab-based experimental tasks, narrative elicitations, conversations and dyadic interactions with an adult, standardized tests and parent-report questionnaires. Probably one of the most interesting findings of this research is the consistency with which people with WS seem to use their language abilities primarily for social engagement, while the content of their communication appears to be secondary to their dyadic interaction goals. This social use of language may be a syndrome-specific feature of the WS behavioral phenotype. Although much remains to be learned about the complex interplay of linguistic, cognitive, social, and cultural factors shaping pragmatic functioning in WS, research conducted so far has made significant strides towards characterizing the profile and the precursors of pragmatic language in people with a neurodevelopmental disorder that continues to challenge our understanding of the organization and development of the human mind.

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