Formative Experiences

The Interaction of Caregiving, Culture, and Developmental Psychobiology

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CAMBRIDGE
UNIVERSITY PRESS

CAMBRIDGE UNIVERSITY PRESS
Cambridge, New York, Melbourne, Madrid, Cape Town, Singapore,
São Paulo, Delhi, Dubai, Tokyo

Cambridge University Press
32 Avenue of the Americas, New York, NY 10013-2473, USA

www.cambridge.org
Information on this title: www.cambridge.org/9780521895033

First published 2010

Printed in the United States of America

A catalog record for this publication is available from the British Library.

Library of Congress Cataloging in Publication data

Formative experiences : the interaction of caregiving, culture, and developmental psychobiology / edited by Carol M. Worthman . . . [et al.].
p. cm.
Includes bibliographical references and index.
ISBN 978-0-521-89503-3 (hardback)
1. Parenting – Cross-cultural studies. 2. Developmental psychobiology.
I. Worthman, C. M. (Carol M.), 1948– II. Title.
HQ755.8.F67 2010
649'.1–dc22 2009033929

ISBN 978-0-521-89503-3 Hardback

For children and their families, everywhere: their lives, our future. (CMW)

. . . May we as clinicians, researchers, policy makers, teachers, and parents come together to be inspired by and with them [those children and their families, everywhere] to encourage the most wonderful of formative experiences for generations to come! (DSS)

SIX

We Are Social – Therefore We Are

The Interplay of Mind, Culture, and Genetics in Williams Syndrome

Carol Zitzer-Comfort, Judy Reilly, Julie R. Korenberg, and Ursula Bellugi

INTRODUCTION

Williams syndrome (WS) is a rare neurodevelopmental disorder arising from a hemideletion in chromosome band 7q11.23, including the gene for elastin (ELN) and approximately 20 surrounding genes (Ewart et al., 1993; Korenberg, Chen, et al., 2000; Korenberg, Bellugi, Salandanan, Mills, & Reiss, 2003; and Korenberg et al., 2008). More than 95% of individuals clinically diagnosed with WS are estimated to have deletions that fall within the same breakpoints (Perez-Jurado, Peoples, Kaplan, Hamel, & Franke, 1996; Korenberg et al., 2003) (see Figure 6.1). Physical characteristics of WS include specific facial and physical anomalies; a variety of cardiovascular difficulties, commonly supravalvular aortic stenosis; mild to moderate mental retardation; failure to thrive in infancy; and small stature (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Morris & Mervis, 1999, 2000; and Korenberg et al., 2008).

CHARACTERISTIC FEATURES OF WILLIAMS SYNDROME

The basic anatomy of the brain in people with WS is normal, but the total volume is somewhat reduced. The areas that seem to be best preserved include the frontal lobes and a part of the cerebellum called the neocerebellum, as well as parts of the temporal lobes known as the limbic area, the primary auditory area, and the planum temporale (Lenhoff, Wang, Greenberg, & Bellugi, 1997).

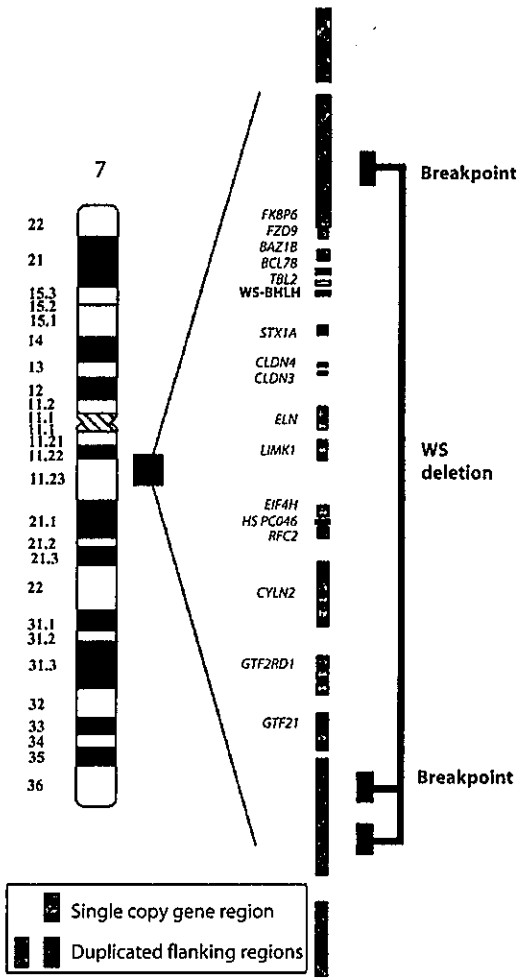


Figure 6.1. The ideogram represents the region of chromosome 7, band 7q11.23, which is commonly deleted in WS. This region is expanded at the right to illustrate its genomic organization, a region of largely single-copy genes flanked by a series of genomic duplications. Bars at the end of the bracket indicate the regions used in the common breakpoints. From Järvinen-Pasley, A., Bellugi, U., Reilly, J., Mills, D. L., Galaburda, A., Reiss, A. L., et al. (2008). Defining the social phenotype in Williams syndrome: a model of linking gene, the brain, and cognition. *Development and Psychopathology*, *20*(1), 1–35. Copyright 2008 by Cambridge University Press. Reprinted with permission.

Figure 6.2. Photographs of children with Williams syndrome (WS). Reproduced with parental permission.

Lenhoff et al.'s (1997) widely circulated overview of the syndrome noted that:

The Salk group's examination of brains by magnetic resonance imaging and by autopsy supports the probability that the chromosomal deletion responsible for Williams syndrome alters the brain in a more complicated way. The deletion seems to produce anatomical changes (such as abnormal clustering of neurons in visual areas) that yield deficits in visual-spatial abilities. But the chromosomal defect appears to spare a network that includes structures in the frontal lobes, the temporal lobe and the cerebellum. This preserved network, then, may serve as a neuroanatomical scaffolding for the unexpectedly strong language abilities of Williams people. (p. 72)

This research is, indeed, exciting and yields new insight into the complexity of the relationship among the brain, genes, and the environment. Neuroanatomical studies, alone, cannot account for the variability nor for the uniqueness of WS; thus, these types of studies must be combined with the newly emerging cross-cultural research.

In addition to the physical characteristics (Figure 6.2), adults with WS often display characteristic patterns of cognitive strengths and weaknesses, that is, comparatively strong language abilities coupled with profound deficits in visuospatial construction (Bellugi et al., 2000). Williams syndrome thus presents a compelling model for the investigation of the impact of genetics on behavior because its genetic basis is well defined and circumscribed and results in an uneven cognitive profile phenotypic of the syndrome.

A consistent behavioral characteristic of WS is the heightened affiliative behavior (see Jones et al., 2000; Bellugi, Järvinen-Pasley, Doyle, Reilly, Reiss, & Korenberg, 2007; Järvinen-Pasley et al., 2008; and Mervis & Klein-Tasman, 2000, for reviews). Almost since it was characterized as a syndrome, anecdotal observations have held that persons with WS are outgoing (Von Arnin & Engel, 1964). Descriptions of hypersociability in adults and children with WS, such as "gregarious personality" and "indiscriminate friendliness" have been reported around the world (Udwin, Yule, & Martin, 1987; Gosch & Pankau, 1994; Bjornstad, 1994; Kotzot et al., 1995; Einfeld, Tonge, & Florio, 1997; Ruangdaraganon, Tocharoentanaphol, Kotchabhakdi, & Khowsathit,1999; Battin, Lancombe, Taine, & Goizet, 2000; Nakaji, Kawame, Nagai, & Iwata, 2001).

One salient phenotypic feature of the syndrome, evident from infancy and extending into adulthood, is increased interest in social interactions (Mervis & Klein-Tasman, 2000; Jones et al., 2000; Meyer-Lindenberg, Mervis, & Berman, 2006; Järvinen-Pasley et al., 2008). In individuals with WS, gregariousness is accompanied by social disinhibition, even toward people not considered approachable (Bellugi, Adolphs, Cassady, & Chiles, 1999; Järvinen-Pasley et al., 2008; Frigerio et al., 2006). In spite of this exuberant sociability, WS individuals may have pronounced difficulty in making lasting friendships, as well as pragmatic difficulties (Tager-Flusberg & Sullivan, 2000; Laws & Bishop, 2004). Despite widespread reports of an unusually intense social drive among these subjects and a growing literature of studies of social behavior in WS, almost no studies to date have examined the effects of different cultural settings on the social behavior of people with WS. In this chapter, we present two studies investigating the sociability of individuals with WS across different cultures. The first focuses on the social use of language in narratives of American, French, and Italian children, and adolescents with WS. The second explores social behavior in American and Japanese children with WS.

THE SOCIAL USE OF LANGUAGE IN WS ACROSS CULTURES

In our past studies, we investigated the narratives of English-speaking children and adolescents with WS (Losh, Bellugi, Reilly, & Anderson, 2000;

Reilly, Klima, & Bellugi, 1990; Reilly, Losh, Bellugi, & Wulfeck, 2004; Kreiter et al., 2002). The developmental profile is one of prolific talkers who have a somewhat delayed but continuing mastery of English morphology and syntax in the face of more impoverished narrative structure (Losh et al., 2000; Reilly et al., 2004). Here, we present a summary of our narrative studies with American children and adolescents with WS.

Language as an Index of Sociability in Williams Syndrome

We presented individuals with WS and their typically developing peers with the same wordless picture book: *Frog, Where Are You?* (Mayer, 1969). The subjects were asked to tell the story to the experimenter. Because this picture book contains no words and provides a rich context for language production, it has been used extensively in cross-linguistic work (Berman & Slobin, 1994) and across typically and atypically developing populations (Reilly et al., 2004; Losh et al., 2000). After the stories were told, they were coded using a scheme designed to assess both grammatical proficiency and use of evaluative language (Reilly et al., 1990; Reilly et al., 2004).

The stories were first coded for length as measured by the number of propositions presented; a proposition is defined as a verb and its arguments, roughly corresponding semantically to a single event. Each clause in a complex sentence was considered to represent one event, and therefore one proposition. Morphological errors were tallied and categorized by type, as were the frequency and types of complex syntax recruited. Proportions were created for all measures.

To capture the social aspects of the narration, we coded the stories for the use of social evaluative language, using a definition of evaluation that draws from William Labov's work on narratives. Evaluation is language that reflects the narrator's attitude or perspective (Labov & Waletzky, 1967). Specifically, evaluative devices are linguistic tools used to attribute emotions or motivations to characters in a story, build suspense, and maintain audience involvement and interest. Examples include emphatics, intensifiers (e.g., *really, very,* and *so*), character speech, direct quotes, and sound effects. Because the language of WS appeared so extensively colorful and attention-getting, we added to our coding schemes a special category of evaluative devices termed "audience hookers," which are intended to capture and maintain the listener's attention. For indices of both language structure (grammatically correct clauses) and language use (evaluative language), proportions were created using story length, as measured by the number of propositions, as the denominator. Two independently trained researchers conducted transcription and coding, and reliability was at or above 90%.

The Social Use of Language in Adolescents with WS

In one of the first language studies on WS (Reilly et al., 1990), we asked adolescents with WS, aged 10–18 years, to narrate the *Frog* story; control groups included age- and IQ-matched adolescents with Down syndrome (DS), and mental-age-matched typically developing (TD) children. The stories were then analyzed for grammar and evaluative language. The adolescents with WS were relatively proficient, specifically in their use of grammar. Also, the WS group used evaluative language significantly more frequently than did the TD controls or the adolescents with DS. Table 6.1 provides examples of evaluative language in WS and Figure 6.3 shows the enriched linguistic affect in WS, as compared to DS and mental-age matched controls. Both the figure and the table provide a taste of the richness of social language among those with WS.

The Social Use of Language in Children With WS in the United States

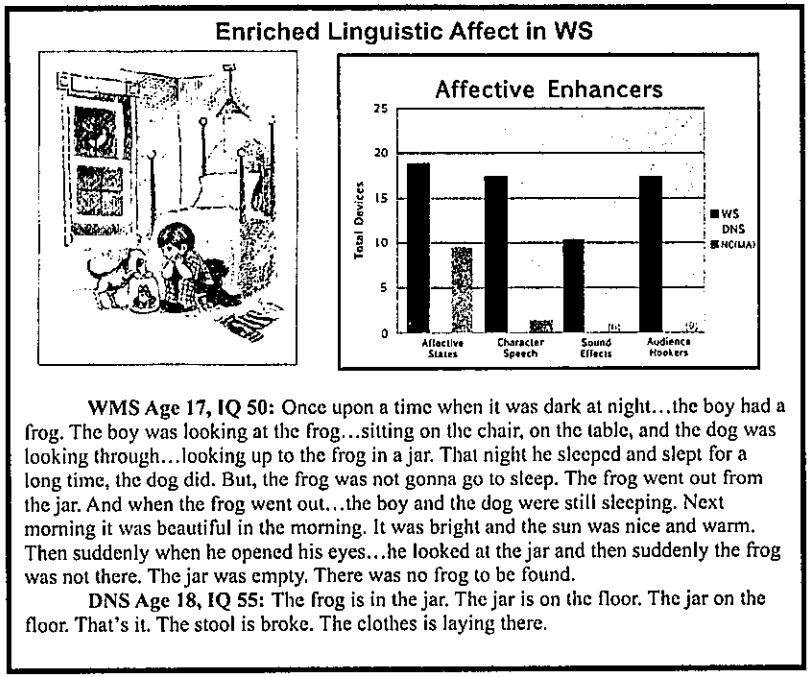
Because the adolescents in our early study had mastered English grammar by and large, we wanted to know if their excessive use of evaluative language was characteristic of the WS group as a whole, and if so, when and how it developed. To address these questions, our next studies included larger and younger samples of children and adolescents. For example, data from 35 children with WS ages 4–12 years, and 70 chronologically age-matched TD children are shown later (Losh et al., 2000; Reilly et al., 2004). Using the coding scheme described earlier, Figure 6.4a shows the mastery of morphosyntax in the narratives of children with WS, compared with age-matched controls. Unlike the adolescents, children with WS are consistently impaired in the proportion of grammatically correct clauses, compared with controls.

In contrast to this developmental lag vis-à-vis the acquisition and use of morphosyntax, one of the most striking aspects of narratives told by WS children is the frequent and pervasive use of what we have termed social evaluation. That is, evaluative devices designed to engage and maintain the listener's attention, such as the use of character voice, intensifiers, and what we have called "audience hookers" (e.g., exclamations, sound effects, and rhetorical questions). As we have found across studies, the WS group recruits these social evaluative devices significantly more frequently than their TD peers (see Figure 6.4b). Thus, whereas morphosyntactic development in this younger group varies, with some children in the normal range and others significantly below (Reilly et al., 2004), the use of social evaluation in their narratives is significantly higher for every single subject that we have studied, compared with controls (Losh et al., 2000, Reilly et al., 2004).

Table 6.1. *Examples from WS of Evaluative Language*

AFFECTIVE STATES	WS: -And ah! he was amazed!
	WS: -The boy looks suspicious.
	WS: -And then he was happy because he had a big family.
	WS: -The dog gets worried and the boy gets mad.
	WS: -And the poor dog was just tired, walking slowly.
	WS: -The next morning, he was sad because the frog left.
CHARACTER SPEECH	WS: -He goes, 'Ouch! oh uh get outta here bumblebees!'
	WS: -And the dog licked him and said, 'Thank you for saving me.'
	WS: -And then the frogs all sat up and the frog goes 'ribbit.'
	WS: -And the boy said, 'Goodbye, Mrs. Frog. Goodbye, Mr. Frog. Goodbye, many frogs. I might see you if I come around again.'
	WS: -He said, 'Wow!, look at these, a female and a male frog and also lots of baby frogs.'
	WS: And the dog licked him and said: "Thank you for saving my life."
SOUND EFFECTS	WS: -And the light goes 'ching.'
	WS: -And 'boom,' millions of bees came out and tried to sting him.
	WS: -Suddenly splash! The water came up.
	WS: -He was looking for the frog, and 'boom,' he broke it.
AUDIENCE HOOKERS	WS: -Suddenly, the frog jumped out!
	WS: -Ouch! that hurt!
	WS: -Gadzooks! The boy and the dog start flipping over.
	WS: -And ah! he was amazed!
	WS: -Lo and behold! they find him . . . with a lady.
	WS: -And all of a sudden, the boy saw a female frog with the frog that he lost.
	WS: -Well, what do you know? A frog family! Two lovers. END
	WS: -And when the frog went out. . .the boy and the dog were still sleeping. Next morning it was beautiful in the morning. It was bright and the sun was. . .really bright and was nice and warm.

Consistent with our previous findings, these results demonstrate the excessively social use of language in WS. Whereas structural language proficiency varies across individuals, as soon as children with WS are able to produce simple narratives, they exploit their linguistic abilities maximally for social purposes. This, perhaps, is one of the most striking characteristics of WS. Talking with a person with WS is an experience one does not soon forget.



WMS Age 17, IQ 50: Once upon a time when it was dark at night...the boy had a frog. The boy was looking at the frog...sitting on the chair, on the table, and the dog was looking through...looking up to the frog in a jar. That night he sleeped and slept for a long time, the dog did. But, the frog was not gonna go to sleep. The frog went out from the jar. And when the frog went out...the boy and the dog were still sleeping. Next morning it was beautiful in the morning. It was bright and the sun was nice and warm. Then suddenly when he opened his eyes...he looked at the jar and then suddenly the frog was not there. The jar was empty. There was no frog to be found.

DNS Age 18, IQ 55: The frog is in the jar. The jar is on the floor. The jar on the floor. That's it. The stool is broke. The clothes is laying there.

Figure 6.3. (1) Enriched linguistic affect in Williams syndrome (WS), as compared to Down syndrome (DS) and mental-age matched controls. (2) Story openings by WS and DS children. (1) and (2) adapted from Reilly, J., Klima, E. S., & Bellugi, U. (1990). Once more with feeling: Affect and language in atypical populations. *Development and Psychopathology*, 2, 367–391. Copyright 1990 by Cambridge University Press. Illustrations reprinted from *Frog, Where Are You?* by Mercer Mayer, copyright ©1969 by Mercer Mayer. Used by permission of Dial Books for Young Readers, A Division of Penguin Young Reader Group, A Member of Penguin Group (USA) Inc., 345 Hudson Street, New York, NY 10014. All rights reserved.

NATURAL BORN STORYTELLERS

Individuals with WS Are Unrivaled in Their Use of Social Language

We recently extended our studies to investigate the specificity of excessive social language use across different populations, including individuals with neurodevelopmental disorders other than WS. We compared and contrasted age-matched WS children and adolescents with three groups, individuals with language impairment (LI), with early focal lesions (FL), and with high functioning autism (HFA), as well as a group of TD children. Whereas the neurodevelopmentally disabled

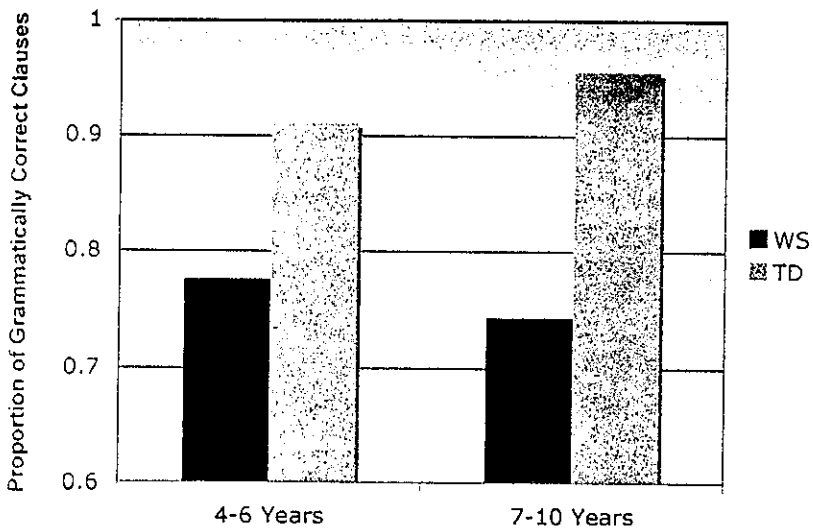


Figure 6.4a. Language structure: Grammatically correct clauses. The mastery of morphosyntax in the narratives of children with Williams syndrome (WS) compared with their age-matched typically developing (TD) controls. Note that at both developmental ages shown, the WS are impaired compared to the controls in the proportion of grammatically correct clauses. From Järvinen-Pasley, A., Bellugi, U., Reilly, J., Mills, D. L. Galaburda, A, Reiss, A. L., et al. (2008). Defining the social phenotype in Williams syndrome: A model of linking gene, the brain, and cognition. *Development and Psychopathology*, 20(1), 1–35. Copyright 2008 by Cambridge University Press. Reprinted with permission.

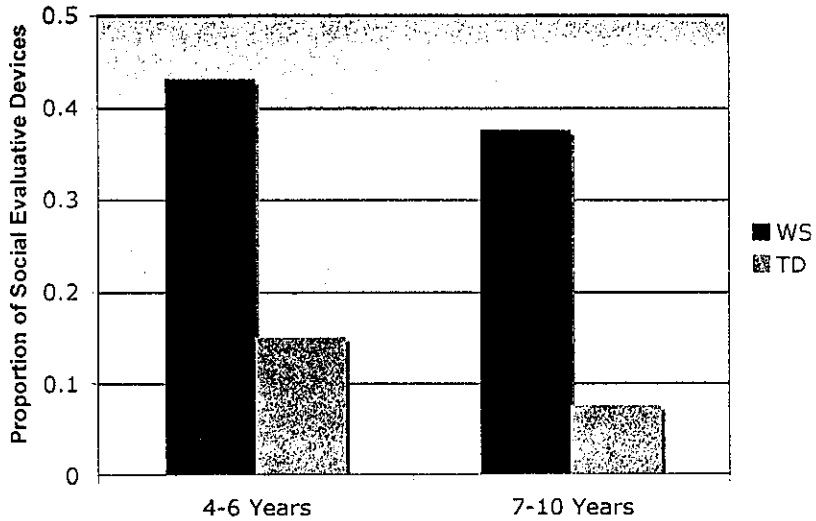


Figure 6.4b. Language use: Social-evaluative language in narratives. The use of social evaluation in the narratives of children with WS and typically developing children (TD). Contrasted with the mastery of morphosyntax, where children with WS lag behind, in the proportion of social evaluative devices, the WS are significantly higher than their matched normal controls. From Järvinen-Pasley, A., Bellugi, U., Reilly, J., Mills, D. L. Galaburda, A, Reiss, A. L., et al. (2008). Defining the social phenotype in Williams syndrome: A model of linking gene, the brain, and cognition. *Development and Psychopathology*, 20(1), 1–35. Copyright 2008 by Cambridge University Press. Reprinted with permission.

groups showed differentially impaired acquisition of grammatical structure, individuals with WS far exceeded any other group, including TD individuals, in their overabundance of social evaluative language (Reilly et al., 2004). As Figure 6.5 demonstrates, our findings strongly suggest that excessively social evaluative language may indeed be uniquely characteristic of WS, in contrast with other populations.

Comparisons Across Genres

Our narrative analyses have drawn from the characterization of narratives by Labov and Waletzky (1967), who described different functions of narrative as *referential* (information pertaining to plot) and *evaluative* (the narrator's perspective and attitude toward events). The latter is largely related to the story's significance to the narrator. Given this theoretical perspective, it is possible that the particular genre used in the studies, that is, narrative, was responsible for the distinctive profile of WS. To control for this possibility, we coded and analyzed biographical "warm-up" interviews from adolescents with WS, DS, and mental-age-typical controls. In these warm-up interviews, conducted at the beginning of testing sessions, experimenters asked questions about the individual's family, friends, school, siblings and pets (Harrison, Reilly, & Klima, 1995). Interviews, as a genre, display a certain structure: The interviewer asks questions and the interviewee responds. Individuals with WS were the only group to *turn the tables* on the experimenter, often reversing the normal roles of interviewer and interviewee by asking questions, making evaluative comments, and even using personal flattery. For example, when asked about family, a WS adolescent said, "I have a sister. Do you have a sister? How old is she?" Another child with WS said to the examiner, "Where do you live? What do you like to eat for dinner? Do you have a boyfriend? I think you are beautiful." Similar

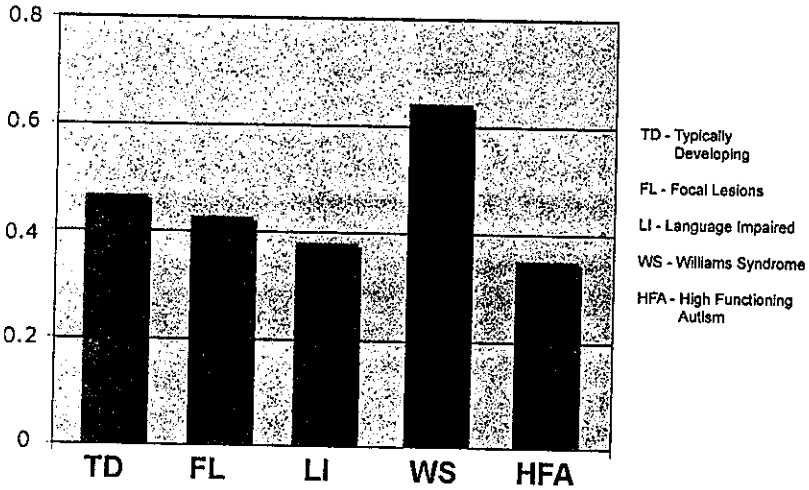


Figure 6.5. Uniqueness of WS language evaluation across groups. In a separate study, we have examined the use of grammatical structure and the social use of language devices from the same narrative across different populations. Shown here are the proportions of social evaluative language in stories from typically developing children (TD) at the far left, followed by those from children with early focal lesions (FL), language impaired children (LI), children with Williams syndrome (WS) in black, and finally individuals with high functioning autism (HFA). Note that the proportion of social language in WS is significantly higher than any other population, including normal controls.

to social evaluative devices, such personal questions and comments function to "hook" an audience, engaging the interlocutor's attention. These complementary data suggest that the social use of language, apparent in the narratives of WS individuals, is not genre-specific, but is a much more pervasive and general phenomenon in this group.

THE INTERSECTION OF GENETICS, CULTURE, AND SOCIAL LANGUAGE IN WS

The studies of language structure and the social use of language in WS described above, which were conducted in the United States, led us to critically important questions that we address in this paper. We have seen that the extensive use of social evaluation, the intersection of language and affect, is pervasive in WS at all ages and across genres, as contrasted with other developmental disorders as well as with age- and mental-age-matched controls. An important issue, not heretofore addressed, is the degree to which this quality, noted in various studies with WS, appears in different cultures and linguistic environments. Thus, our current research focuses on the following questions: "How do different cultures and languages influence the social use of language in WS?" "Given significant differences in languages and cultures, is this WS profile of exuberant sociability consistent outside of the United States?" "What is the effect of different cultural settings on the social language of this genetically based syndrome?" We know from anecdotes, personal experiences, stories, and studies that cultures can vary widely in the socialization of children, the expression of emotion, the use of gestures, the manner of greeting strangers, and the structure of spoken language. Cross-linguistic and cross-cultural studies would provide opportunities to separate some effects of the genetic basis for WS from environmental and cultural effects.

These questions led us to our current study, in which we chose cultures and environments that contrast with American culture in noteworthy ways. We chose Italy and France, using our contacts in those two countries to initiate the studies (Reilly, Bernicot, Vicari, Lacroix, & Bellugi, 2005). Both Italy and France are romance cultures, yet they contrast in their display rules for emotion: The French are considered rather reserved, whereas the Italians are more effusive. In fact, comparative studies found American mothers to be more expressive than French mothers with infants (Suizzo, 2004; Bril, Dasen, Sabatier, & Krewer, 1999). French children are taught to control their emotional expressions, to be quiet and discreet in public, and to express their emotions appropriately and correctly. This discretion is nicely reflected in the French proverb, "Vivre heureux, vivons cachés" (To live happily, we live hidden). In contrast, Italian is categorized as a "high-gesture" language (Kendon, 1995a, 1995b), reflecting the overall increased expressivity of Italian culture. This expressivity has also been documented in a comparative study of children's story books (Shatz, Dyer, Marchetti, & Massaro, 2006), in which the authors found that Italian translations differed from the English version in three ways: heightened emotional intensity, more specific expression of mental states, and more explicit expression of social awareness or responsibility. Such findings reflect not only an increased expressivity, but also a higher value placed on sociability in Italian culture.

Social Use of Language in Children and Adolescents with WS in Italy

The first non-U.S. group of individuals we studied were 17 Italian children with WS aged 10–16 years, and their mental-age-matched TD peers. As noted above, Italian has been categorized as "high-gesture" (Kendon, 1995) in that speakers frequently recruit gestures in their social interactions.

As in other languages, evaluation can be conveyed lexically and paralinguistically in Italian (i.e., by gestures and intonation); however, an Italian speaker also can use suffixes that convey evaluative content. For example, a boy is *un ragazzo*, a nasty boy is *un ragazzaccio*, a little boy is *un ragazzino* and a nasty little boy is *un ragazzinaccio*.

Given the richness of Italian forms and the frequency of gesture, one might ask whether Italian children with WS use social evaluation or affective language in a manner similar to that of TD Italian children, that is, whether all Italian children rely heavily on social evaluation, with little difference in the WS group. To address this question, we again collected, transcribed, and coded narratives from the Italian version of *Frog, Where Are You?* Looking at morphology and syntax, we found that Italian children with WS, like their American counterparts, made more errors and used less complex syntax than age-matched controls, but they also used significantly more social evaluation than did the TD Italian children.

Social Use of Language in Children and Adolescents with WS in France

Our second non-U.S. group is composed of 12 children and adolescents with WS aged 6–16 years who live in France, along with chronological age-matched TD peers. When we examined the use of evaluation in the French group, (Reilly, Bernicot, et al., 2005; Reilly, LaCroix, et al., 2005), we again found significantly more social evaluation in the WS group than in the control group. The findings for French subjects are consistent with the findings in English and Italian subjects with respect to the effect of the genetic basis of WS. That is, French children with WS use significantly more social evaluation than their typically developing peers, just as American and Italian WS children do. To give a taste of the rich and unique nature of children's use of social evaluation, Table 6.2 below includes examples from the *Frog* stories, highlighting the use of evaluative language among subjects with WS in the three countries.

Thus, all three groups of WS children and adolescents from different linguistic and cultural backgrounds are significantly more socially expressive than their respective control groups. Importantly, however, the results also show a significant effect of culture: Typically developing Italian children show the highest social evaluation, whereas the French are lowest in that parameter and the Americans are in between (Reilly, Bernicot, et al., 2005).

These comparative results suggest that in spite of the clear propensity of individuals with WS to recruit high levels of social evaluation, the nature of each culture and its conventions for demonstrating sociability and conveying emotion also help determine how the social behavior of WS children and adolescents is expressed in language.

Table 6.2. *Evaluative Language in English, Italian, and French*

Examples of Evaluative Language in English	
But, phew! (signaling relief), it was just a little bit swampy	
He said "wow, look at these . . . a female and a male frog and also lots of baby frogs	
And lo and behold . . . Some frogs came out of the bushes	
Here's the frog and he's in love! And he says "Hooray! Hooray! Hooray! I found my froggie!" And then he "Byeeee!"	
Examples of Evaluative Language in Italian	
"Rana, . . . (ride) raaana dove sei?" "Bow wow".	*"Frog . . . (laughs) frooog, where are you?" "Bow wow". The dog fell with that thing Boom! And then "Where did you get to?"*
Il cane cadeva con questa cosa Bum!	
E poi "dove sei finita?"	
Questo rana simpatiche!	*What a funny frog!*
Trovano un piccolino	*They found a little bitty one*
E poi c'e il gufo che sta cosi triste	*And then there is the owl that is so sad*
Examples of Evaluative Language in French	
Le garcon dit "mince le bocal va etre casse"	*The boy said: Darn! The jar is gonna break*
Et puis il s'enerve finalment et le chien il est content	*and then he gets upset finally and the dog's happy*

DISCUSSION: THE INTERPLAY OF MIND, GENETICS, CULTURE, AND SOCIAL LANGUAGE IN WS ACROSS CULTURES

In this first section of the chapter, we have used narratives as a context to explore the intersection of affect and language in children and adolescents with WS to better understand the phenotype and how cultural conventions might modulate its expression. It appears that in spite of the culture and the resources of the language, or lack thereof, children and adolescents with WS are characterized by their frequent and extensive use of social evaluation in their stories. Although language studies are one avenue for investigating the impact of culture on a genetically based syndrome, we continued to seek additional avenues to investigate the interplay of nature and nurture in WS across cultures. We turn now to the use of a different measurement tool, parental questionnaires, to assess the social drive in WS in contrasting cultures, the United States, and Japan.

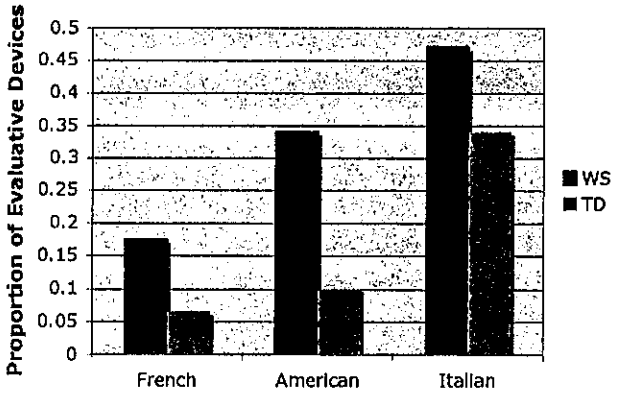


Figure 6.6. The effect on social evaluative language in Williams syndrome (WS) and typically developing (TD) controls. Note that in both the figure and the table, WS individuals use significantly higher social language than controls, but at the same time, the culture has an effect as well. From Järvinen-Pasley, A., Bellugi, U., Reilly, J., Mills, D. L. Galaburda, A, Reiss, A. L., et al. (2008). Defining the social phenotype in Williams syndrome: A model of linking gene, the brain, and cognition. *Development and Psychopathology*, 20(1), 1–35. Copyright 2008 by Cambridge University Press. Reprinted with permission.

THE SOCIAL DRIVE OF WILLIAMS SYNDROME ACROSS CULTURES

The French, Italian, and American language studies discussed earlier led to more questions about the impact of "nurture" on the expression of a genetic syndrome. We were particularly interested in the expression of hypersociability in WS among various cultural groups in the United States and abroad. As with language studies, which had been conducted in the United States only, the affiliative drive that is typical of WS had not been examined across cultures.

Nobody questions that the color of our eyes is encoded in our genes. When it comes to behavior, however, the concept of "DNA as fate" quickly breaks down. It has long been accepted that both genes and the environment shape human behavior. But just how much sway the environment holds over our genetic destiny has been difficult to untangle. By comparing the social behavior of WS children – known for their innate drive to interact with people – across cultures that have differing social mores, we are beginning to learn the answer. Overall, a consistent result has emerged: Regardless of age, language, or cultural background, the social phenotype of individuals with WS is shaped by both genes and gene-environment interactions.

Assessing the Social Drive in WS and TD Controls Using the SISQ

In a previous study (Doyle, Bellugi, Korenberg, & Graham, 2004), we assessed sociability among a large group of children in the United States using a parental report questionnaire, the Salk Institute Sociability Questionnaire (SISQ). SISQ is designed to ask parents to rate their child's tendencies to approach others, to remember names and faces, to please other people, and to empathize with or comment on others' emotional states. It also asks about the child's general behavior in social situations and the tendency for other people to approach their child.

The questionnaire was completed by 64 parents of WS children aged 2–12 years; control groups included 31 parents of children with Down syndrome (DS) and 27 parents of typically developing (TD) age-matched controls. Results showed that children with WS were rated overall as significantly more social than DS children or TD subjects. In addition, children with WS were rated significantly higher in approaching strangers than the other two groups, and higher with respect to social-emotional behaviors than DS children (but not different when compared with age-matched TD subjects). Significant differences in social behavior were reported from the earliest ages assessed, with WS children exceeding both comparison groups.

These findings provide initial evidence that differences in hypersociability, particularly an attraction to strangers, cannot be attributed simply to cognitive impairment, resulting in a lack of understanding of the social conventions governing others (both WS and DS children are cognitively impaired); nor can they be attributed to developmental factors (see also Jones et al., 2000).

SOCIAL BEINGS BY NATURE

Our previous studies suggest the involvement of genetic predisposition in the expression of hypersociability in WS; thus, exploring sociability across cultures can provide keen insight into the interplay of temperament (in a disorder with a known genetic basis) and culture. This study examines the ways in which social behavior in WS, which is thought to have a genetic predisposition, might be mediated by cultural expectations in both Japan and the United States. Because both the genetic phenotype of WS and the presence of excessive friendliness toward strangers or overly social behavior in that syndrome are well documented within the United States, it is of great interest to know whether the expression of hypersociability is influenced by cultural and societal mores, particularly by the factors that

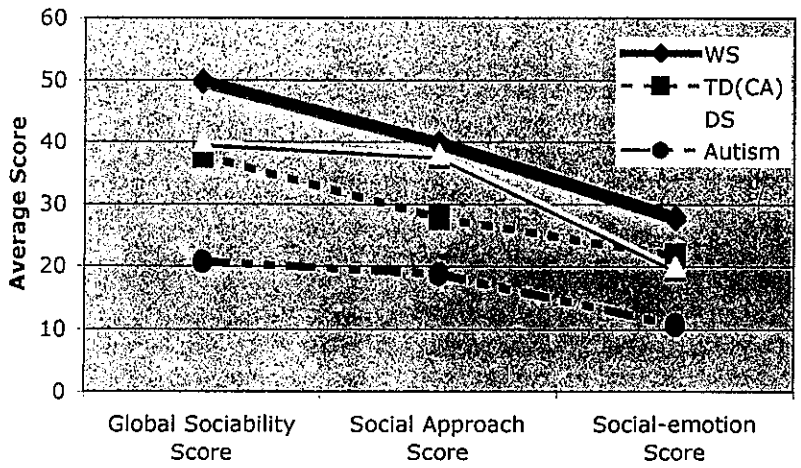


Figure 6.7. Parental characterization of sociability contrasting WS, DS, Autism, and TDs (SISQ). Individuals WS are consistently rated higher by their parents in social behaviors using the Salk Institute Sociability Questionnaire than chronological age (CA)-matched individuals with autism, DS, or TD. From Järvinen-Pasley, A., Bellugi, U., Reilly, J., Mills, D. L. Galaburda, A, Reiss, A. L., et al. (2008). Defining the social phenotype in Williams syndrome: A model of linking gene, the brain, and cognition. *Development and Psychopathology*, 20(1), 1–35. Copyright 2008 by Cambridge University Press. Reprinted with permission.

prescribe appropriate social behavior. It is intriguing to explore ways in which children with developmental disorders – especially those disorders with a known genetic basis – might be affected by social mores of two vastly differing cultures.

THE SOCIAL DRIVE OF WS IN THE UNITED STATES AND JAPAN

Genetics and Culture at Play

To determine the extent to which the affiliative drive in individuals with WS is universal, we settled on two countries with markedly contrasting cultures for this research study: the United States and Japan. These two cultures, respectively, have often been contrasted as exemplars of "individualistic" and "collectivist" societies. Differences between them can be summed up by the following distinctive proverbs: In America, "The squeaky wheel gets the grease." In Japan, "The nail that stands out gets pounded down" (Markus & Kitayama, 1991). In their landmark paper examining culture and the self, Markus and Kitayama further note that "People in Japan and America may hold strikingly divergent construals of the self, others, and the interdependence of the two. American examples stress attending to the self, the appreciation of one's difference from others, and the importance of asserting the self. The Japanese examples emphasize attending to and fitting in with others and the importance of harmonious interdependence with them" (p. 224).

This "individualistic" self-view in western cultures leads to defining one's self in terms of one's own feelings and actions, placing emphasis on the ways in which the individual is unique. Asian cultures emphasize more of a "collectivistic" self-view in which one defines oneself in terms of relationships with others (see Triandis, 1989, 1995).

Because of the contrasting viewpoints of "self" in the two countries, such differences could be expected to lead to variations in socialization of children in the two countries. Hess et al. (1986) note that "In Japan, a child is thought to be good if he or she is 'obedient' (*sunao*), 'mild and gentle' (*otonasii*), and 'self-controlled' (*jiseishin ga aru*). In the United States, the good child is assertive, socially competent with peers, and courteous" (p. 158). Moreover, parental reports in these countries have shown that mothers in Japan rate their children as shyer and less sociable than comparable ratings by mothers in the United States (Stevenson et al., 1990). (For further discussion on differences between United States and Japanese socialization and child rearing practices, see Conroy, Hess, Azuma, & Kashiwagi, 1980; LaFreniere et al., 2002; Lebra, 1994; Masataka, 2002; White, 1993; Zahn-Waxler, Friedman, Cole, Mizuta, & Hiruma, 1996.)

It is, therefore, of great interest to know how the expression of hypersociability is also influenced by cultural and societal mores, particularly the factors prescribing appropriate social behavior. Although a number of research groups have been studying the strengths and weaknesses of the cognitive profiles of individuals with WS in various countries, little or nothing is known about the effects of different cultures and environments on people with WS. For example, how does genetics influence the expression of sociability in these subjects across cultures that have varying expectations for appropriate behavior?

ASSESSING SOCIABILITY IN WS USING A PARENTAL QUESTIONNAIRE

Across Cultures and Languages

Participants included the parents of 24 children living in Japan and 24 in the United States. Twelve of the children in each sample had WS and 12 were TD. The children ranged in age from 3 years to 13 years; males and females were equally represented. We studied age-matched and gender-matched pairs, with one American and one Japanese child in each pair. The study

was conducted via questionnaires given to the parents of the 48 children. In the United States, participants included parents of WS children who were attending a meeting of the Williams Syndrome Association and parents of TD children who attend school near the Salk Institute in California. In Japan, the Japanese Williams Syndrome Foundation collected the WS data, whereas TD data were collected through the laboratory of co-author Dr. Nobuo Masataka (Zitzer-Comfort, Doyle, Masataka, Korenberg, & Bellugi, 2007). To ensure consistency between the English and Japanese versions of the SISQ for cross-cultural comparison, two individuals, fluent and literate in both English and Japanese, independently translated the SISQ from English into Japanese and then back-translated from Japanese to English.

Evaluating Sociability in WS Children in the East and West

The Salk Institute Sociability Questionnaire (SISQ) was developed to assess specific aspects of social behavior commonly reported among people with WS; results from the SISQ were first reported in Jones et al. (2000). The SISQ has been used in a variety of different contexts and across age groups. Moreover, the Salk Institute's Laboratory for Cognitive Neuroscience (LCN) has collected data on over 80 adolescent and adult individuals with WS; parents of 44 of these individuals had completed both the SISQ and another standardized parent report instrument, the Multidimensional Personality Questionnaire (MPQ) (Tellegen, 1985). LCN studies find that the SISQ overall scores show high correlations with the MPQ in the WS cohort on social dimensions such as Social Potency and Social Closeness, but no correlation with either other MPQ measures or IQ. Bonnie Klein-Tasman has also used the MPQ with a different cohort of individuals with WS (Klein-Tasman & Mervis, 2003) and found that, like the Salk Institute team, the distinctiveness of the WS personality appears to lie in focusing on others, a pattern characterized by an eagerness to interact with others as well as high levels of tension and sensitivity. This distinctiveness of the WS social phenotype provides the groundwork for the present study of cross-cultural influences upon social behavior.

DISCUSSION: NATURE AND NURTURE OF SOCIABILITY AMONG INDIVIDUALS WITH WS

The SISQ consists of both quantitative and qualitative items. Quantitative items ask parents to rate their child's specific social behavior on a 7-point Likert scale. Qualitative items ask the parents to provide a descriptive response. These items assess Global Sociability by yielding three subscales: a tendency to approach strangers, a tendency to approach familiars, and social emotional behavior (such as tendency to empathize with others, accuracy of emotional evaluations of others, eagerness to please others, and ability to remember names and faces of others).

Items assessing social approach behavior ("Social-Emotional") consist of statements such as "How would you compare your child's tendency to approach strangers with an average child of the same age?" or "How would you describe your child's general behavior in social situations?" Results for the first question are rated on a scale ranging from 1 ("approaches much less") to 7 ("approaches much more"), whereas the scale for the second question ranges from 1 ("very shy and inhibited") to 7 ("extremely outgoing"). Qualitative items include "Describe your child's typical reactions when meeting someone for the first time (please give examples)"; or "Give some examples of your child's socializing with strangers." The social approach items were grouped for analysis into two types. Those that assess a child's tendency to approach family members or others who are encountered frequently ("Approach Familiars") and those that assess a child's tendency to approach people unknown to them ("Approach Strangers"). The Social-Emotional score was the sum of four items; the Approach Familiars score was the sum of three items; the Approach Strangers score was the sum of five items; and all 12 items added up to the Global Sociability score.

Quantitative Data Analysis

The quantitative data were analyzed by a 2×2 analysis of variance (ANOVA) with Diagnostic Category (WS children versus TD children) and Culture (American versus Japanese) as independent variables, and Global Sociability as the dependent variable. Wilks' Lambda criterion was used to assess significance. Figure 6.8 shows the data distribution for the Global Sociability score, combining all questionnaire items, and the data for each of the three subscales.

As Figure 6.8 shows, both American and Japanese WS children rated significantly higher on Global Sociability than did the TD children; thus, there was a very strong effect for Diagnostic Category (WS or TD). At the same time, however, there was a significant effect of Culture, in that parents of U.S. children tended to rate their children higher in Global Sociability than did parents of Japanese children, regardless of the diagnostic category (WS or TD).

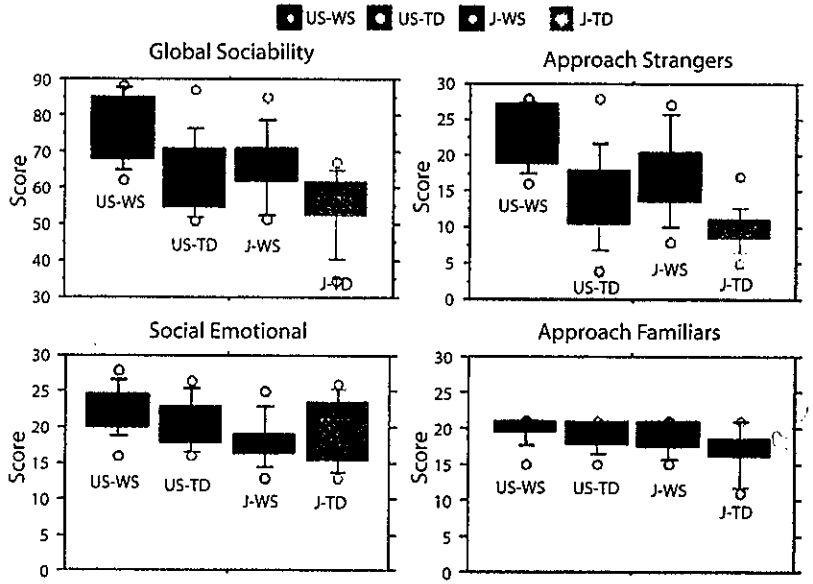


Figure 6.8. Summary of comparisons across Diagnostic Category (Williams vs. typically developing) and Culture (Japanese vs. U.S.) for the quantitative analysis of the SISQ. Graphs display horizontal lines at the 10th, 25th, 50th, 75th, and 90th percentiles. The top box shows Global Sociability, encompassing all parts of the SISQ combined. The Japanese WS mean scores are significantly higher than the Japanese TD means; and the U.S.-WS means are significantly higher than the U.S.-TD means, a major effect of Diagnostic Category. Whereas the U.S.-WS means are significantly higher than all other means, the Japanese-WS are almost on a par with the U.S.-TD, and there is also a significant effect of Culture (U.S. vs. Japanese) as well. These patterns do not hold for Social-Emotional items. Note that on Approach Familiar items, the four groups are almost indistinguishable, at near ceiling. On the Approach Strangers items, the significant effects of both Diagnostic Category and Culture are strongly observed. From Zitzer-Comfort, C., Doyle, T., Masataka, N., Korenberg, J., & Bellugi, U. (2007). Nature and nurture: Williams syndrome across cultures. *Developmental Science*, 10(6):755–762.

Comparison of scores across Cultures indicates a major difference for Approach Strangers, but not for Approach Familiars (scores for Approach Familiars were consistent across groups and cultures) or Social-Emotional items. Similarly, comparison of scores across Diagnostic Category indicates a major difference for Approach Strangers, but not for the other two subscales. Overall, the significant between-group differences in Global Sociability resulted primarily from higher Approach Strangers ratings for both Diagnosis and Culture.

Qualitative Analysis of Sociability Data

The SISQ also asks parents for qualitative descriptions of their child in various social situations, e.g., "Describe your child's typical reactions when meeting someone for the first time (please give examples)." Table 6.3 presents samples of qualitative responses from the various age groups across the cultural and diagnostic groups discussed in this study.

The examples presented in Table 6.3 demonstrate that specific behaviors described by parents of children with WS to illustrate approaching strangers and socializing with them are very similar in both cultures. Nevertheless, Japanese parents rated their children lower on the 7-point scale than did U.S. parents.

The differences in quantitative scores for Japanese WS children may more accurately reflect parental attitudes or sensitivities than the actual behaviors of the children: Perhaps cultural influence is exerted more on parents' *ratings* than on the expression of the behavior, or perhaps the stigma of having a "different" child in Japan affects how parents rank their child's degree of sociability. Yet this explanation does not account for the lower Global Sociability and Approaching Strangers scores for Japanese TD children. To resolve possible discrepancies caused by reliance on parental reports, with the objective of understanding the interplay of phenotype and culture in WS, further cross-cultural observational studies are needed that involve more participants and a deeper look at interactions with strangers.

CONCLUSION AND FUTURE RESEARCH DIRECTIONS: THE SOCIAL NATURE OF WS – WHERE GENETICS AND CULTURE MEET AND CONVERGE IN THE EAST AND WEST

Williams syndrome provides a compelling model for investigating the effects of genotype, phenotype, and environmental interactions. The genetic basis of WS is by now well known and has been documented in great detail (unlike, for example, the basis of autism). The genetic phenotype thus involves the absence of one copy of a small set of genes on chromosome 7, an absence that occurs in nearly all those clinically identified with WS (Korenberg et al., 2003; Korenberg et al., 2008). Currently members of our research group (Korenberg, Reiss, Reilly, Bellugi, et al.) are working together to begin to link genotype and phenotype in WS, and the hunt is on to link specific genes within the WS region with brain development and behavioral functions. An initial yet powerful approach involves intensive examination of some specific cases of individuals with specific smaller deletions (see Doyle, Bellugi, Korenberg, et al., 2004; Doyle, Bellugi,

Table 6.3. *Sample Responses to Qualitative Item 5: "Describe your child's typical reactions on meeting a stranger for the first time"*

Age Range	J TD	J WS	US TD	US WS
(3–4 years)	She flinches and comes to parents	He says words that are used in greeting like "hello" and "what are you doing?" It happened frequently when he just learned the words. Although it is not happening as frequently as before, when he wants to brag about something, he still approaches strangers and starts talking to them.	Shy. Makes only minimal eye contact. Tries to hide behind familiar people	Searches them out to attain eye contact. Reaches for their hands o pull them down to his level. Big, happy smiles.
(5–8 years)	She often holds my hand tightly, stays behind me, and observes the person. She will greet if told to do so. She observes how I, her mother, respond to the person and tries to correspond the situation	She always greets them by saying "Hi! I'm M—" energetically. If someone talks to her, she happily starts to talk about different things	Warms up to children but not to adults for some time.	Introduces self with, "Hi, I'm J... May I ask a question or two?"
(9–13 years)	She more likely watches the person from a distance rather than talking to him/her. She has a little difficulty greeting even after she is introduced to the person. She tries to minimize her words when she has to answer	He starts talking about himself by looking straight into a person's eyes. He says, "What are you doing." Or "I'm K.—" He still greets people by saying "hello" to strangers passing by.	Sometimes shy with adults and/or disinterested. With friends, she will introduce herself and is very friendly	13, is always engagin She asks numerous questions and inquires as to a person's living arrangements. She has invited strange to our home for dinner.

Reiss, Galaburda, Mills, & Korenberg, 2004; Korenberg et al., 2003; Hirota et al., 2003; Dai et al., 2008 for examples). The approach already has led to the hypothesis that specific genes near the end of the deletion in WS may be related to expression of social behavior (Korenberg et al., 2007; Bellugi, Järvinen-Pasley, Reilly, et al., 2007; Salk Press Release, 2007).

The focus on both consistency and variability of sociability in WS children permits consideration of the expression of sociability not only in WS individuals with typically sized deletions but also in those with atypical deletions. For example, the large-scale study of development of sociability in young WS, DS, and TD subjects described earlier included data from a young WS child. This child had a smaller-than-typical deletion that retained between one to three genes in the telomeric region that are almost invariably deleted in the "classic" allele. This child had typical medical and cognitive diagnostic characteristics for WS; however, her sociability scores, most significantly, those for approaching strangers, were significantly lower than the mean of the WS group, implicating specific genes in the emergence of this behavior in WS (Doyle, Bellugi, Reiss, et al., 2004; Korenberg et al., 2007).

The relative lack of variability in hypersociability among WS children, which can be gleaned both from parental reports and experimental as well as observational measures, suggests that the behavioral feature of hypersociability in approaching strangers may be strongly influenced by the genetic deletion (see also Klein-Tasman & Mervis, 2003), tempered by environmental factors. The early age of onset of WS and the case of the child with the atypical deletion further support this conclusion.

The results of the comparative Japan–U.S. study support a genetic "proportional stamp" on the expression of social behavior in WS across cultures; children with WS in both cultures showed more affinity for approaching strangers and rated higher in Global Sociability than TD children in their respective countries. That study, using a common instrument, aimed to examine social behavior among WS children in the two countries to investigate how cultural expectations or mores influence social behavior. Our results suggest that the WS social and genetic phenotype may influence affiliative behavior toward strangers, even among children who are subject to a cultural expectation of cautiousness towards strangers. Despite differences in upbringing and cultural expectations regarding social interaction, both Japanese and Americans with WS were rated significantly higher in Global Sociability and in tending to approach strangers than their TD counterparts. However, the sociability of American TD children was on par with Japanese WS children – whose social behavior is considered out

of bounds by Japanese standards – a sign that cultural expectations clearly influence social behavior. By both quantitative and qualitative measures, these differences are evidence of nature's stamp on culture's milieus, and the reverse.

The same can be said of the use of social evaluation in language in WS. Because social language use varies across cultures in TD individuals, language studies also provide a glimpse of the exciting interaction of culture and genetics in WS. As demonstrated in the first section of this chapter, WS children across cultures consistently engage their listeners with "audience hookers," turn the tables on interviewers, and outperform their respective control groups in using social evaluative language. Given this persistent profile, the atypical expressive use of language in narratives may well be a "marker" of the WS phenotype; it is also intriguing with respect to the contribution of genes to neural systems that underlie social behavior (Doyle, Bellugi, Korenberg, et al., 2004). Most importantly, it becomes clear that the form and intensity of social behavior in WS is influenced by an individual culture's display rules and social conventions for *expressing* sociability. Thus language in a genetically based syndrome, in particular its structure and social uses, proves a productive tool in studying the complex interplay between our genes and our environment.

The expression of sociability in WS and the inimitable nature of social language in WS may not be "either/or" phenotype/culture; they are, rather, "both/and." The particular features of WS allow us to investigate the dual influences of nature and nurture; thus, continuing to examine WS across cultures and across domains will be an important avenue for further exploration.

ACKNOWLEDGMENTS

This is a review chapter that borrows heavily from our work and writing from several papers (Doyle, Bellugi, Korenberg, et al., 2004; Zitzer-Comfort et al., 2007; and Järvinen-Pasley et al., 2008). All illustrations are the copyright of Dr. Ursula Bellugi and the Laboratory for Cognitive Neuroscience.

We thank Dr. Anna Järvinen-Pasley and Yvonne Searcy for their contributions to the analyses. We also would like to thank the many families who contributed to this research in four countries. Without the continuing support of families living with Williams syndrome, our understanding of the interplay of nature, nurture, and the human condition would not be nearly as advanced as it is today. In Japan, Mr. Sugimoto of the Japanese Williams Syndrome Association, was particularly helpful as he translated many of the materials into Japanese and acted as a liaison for the Japanese families who chose to participate. In Italy, the data were gathered by Stefano Vicari in collaboration with the Ospedale Bambino Gesù in Santa Marinella (Rome); and in France, data were gathered by A. Lacroix in children's homes through the Williams Syndrome Family Associations and the Laboratoire, Langage et Cognition (LaCo, Université de Poitiers-CNRS).

The research was supported in part by NICHD HD 33113 to UB, JRK and NIND NS 22323 to UB, JR.

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