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Nature and nurture: Williams syndrome across cultures

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INTRODUCTION

Williams Syndrome (WS) is a rare neurodevelopmental disorder arising from a hemizygous deletion in chromosome band 7q11.23, including the gene for elastin (ELN) and approximately 20 surrounding genes (Ewart, Morris, Atkinson, Jin, Sternes, & Spallone, 1993; Korenberg, Bellugi, Salandanan, Mills, & Reiss, 2003; Korenberg, Chen, Hirota, Lai, Bellugi, & Burian, 2000).

It is estimated that more than 95% of individuals clinically diagnosed with WS have deletions that fall within the same breakpoints (Perez Jurado, Peoples, Kaplan, Hamel, & Franke, 1996). 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). In addition, adults with WS often display a characteristic pattern of cognitive strengths and weaknesses, that is, comparatively strong language abilities coupled with profound deficits in visuospatial functioning (Bellugi, et al., 2000; Mervis, Robinson, Bertrand, Morris, Klein-Tasman, & Armstrong, 2000).

In addition to the typical cognitive profile, a consistent behavioral characteristic of WS is heightened affiliative behavior as defined, in part, as “being ‘overly friendly’ [or ‘hypersocial’] compared to age-matched controls, and more ‘unreserved with strangers’ as compared to children with non-specific mental retardation at the same chronological ages” (see Doyle, Bellugi, Korenberg, & Graham, 2004; Jones, Bellugi, Lai, Chiles,

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3 Reilly, & Lincoln, 2000; Klein-Tasman & Mervis, 2003; Mervis & Klein-Tasman, 2000,
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5 for reviews). Almost since its characterization as a syndrome, anecdotal observations
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7 have been made that persons with WS were outgoing (Von Armin & Engel, 1964).
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9 “Gregarious personality,” “indiscriminate friendliness,” and other similar descriptions of
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11 hypersociability in adults and children with WS have been reported (e.g., Battin,
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13 Lancombe, Taine, & Goizet, 2000; Bjornstad, 1994; Einfeld, Tonge, & Florio, 1997;
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15 Franceschini, Guala, Vardeu, Signorile, Franceschini, & Mastroiacovo, 1996; Kotzot,
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17 Bernasconi, Brecevic, Robinson, Kiss, & Kosztolanyi, 1995; Nakaji, Kawame, Nagai, &
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19 Iwata, 2001; Ruangdaraganon, Tocharoentanaphol, Kotchabhakdi, & Khowsathit, 1999;
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21 Teo, Chan, Yong, Ng, Wong, & Knight, 1997; Udwin, Yule, & Martin, 1986; van Hagen,
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23 Govaerts, de Coo, Gille, Nieuwint, & Madan, 2001). Individuals with WS are highly
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25 social, and yet they may have great difficulty sustaining friendships (Gosch & Pankau,
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27 1994). Moreover, people with WS have been described as being socially fearless;
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29 however, the remarkable social phenotype may be coupled with a strong undercurrent of
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31 anxiety (Meyer-Lindenberg, Mervis, & Faith Berman, 2006), which suggests that
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33 sociability in WS is exceptionally complex and requires further study.
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41 In a previous study (Doyle, et al., 2004), we assessed sociability among a large
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43 group of children in the United States using a parental report questionnaire, the Salk
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45 Institute Sociability Questionnaire (SISQ). SISQ items ask parents to rate the tendency of
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47 their child to approach others, their general behavior in social situations, their ability to
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49 remember names and faces, their eagerness to please other people, their tendency to
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51 empathize with or comment on others’ emotional states, and the tendency for other
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53 people to approach their child. The questionnaire was completed by 64 parents of
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3 children with WS between the ages of 2 and 12 years, 31 parents of children with Down
4 syndrome (DS), and by 27 parents of typically developing (TD) age-matched controls.
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6 Results showed that children with WS were rated as being significantly more social
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8 overall than were those with DS or TD subjects. In addition, children with WS were
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10 rated significantly higher in approach towards strangers as compared to either of the other
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12 two groups, and higher than DS (but not different from age-matched TD) with respect to
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14 social-emotional items. Significant differences in social behavior were reported from the
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16 earliest ages assessed, with WS exceeding both comparison groups. These findings
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18 provide initial evidence that differences in hypersociability, particularly the attraction to
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20 strangers, cannot be attributed simply to cognitive impairment, resulting in a lack of
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22 understanding of the social conventions governing others, as both WS and DS are
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24 cognitively impaired; nor can they be attributed to developmental factors (see also Jones,
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26 et al., 2000).
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34 Because both the genetic phenotype of WS and the presence of excessive
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36 friendliness toward strangers or overly-social behavior in WS are well documented
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38 within the U.S., it is of great interest to know whether the expression of hypersociability
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40 is influenced by cultural and societal mores, particularly those factors prescribing
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42 appropriate social behavior. It would be interesting to explore ways in which children
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44 with developmental disorders--particularly those disorders with a known genetic basis--
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46 may be affected by social mores of such vastly differing cultures: the United States and
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48 Japan.
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53 The cultures of the United States and Japan have often been contrasted as
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55 exemplars of "individualistic" and "collectivist" societies, respectively. Differences
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3 between the two cultures can be summed up by the following vastly differing proverbs:
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5 “In America, ‘The squeaky wheel gets the grease.’ In Japan, ‘The nail that stands out gets
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7 pounded down’” (Markus & Kitayama, 1991, p. 224). In their landmark paper examining
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9 culture and the self, Markus and Kitayama further note that “People in Japan and
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11 America may hold strikingly divergent construals of the self, others, and the
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13 interdependence of the two. American examples stress attending to the self, the
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15 appreciation of one’s difference from others, and the importance of asserting the self.
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17 The Japanese examples emphasize attending to and fitting in with others and the
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19 importance of harmonious interdependence with them” (p. 224). This “individualistic”
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21 self-view in western cultures, leads to defining one’s self in terms of his or her own
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23 feelings and actions; thus, placing emphasis on the ways in which the individual is
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25 unique. Alternatively, Asian cultures emphasize more of a “collectivistic” self-view in
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27 which one defines him or herself in terms of relationships with others. The emphasis in
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29 this view is on ways in which one relates to others (see Triandis, 1989, 1996). Because of
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31 the contrasting ways in which individuals conceive of “self” in the United States and in
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33 Japan, one might expect such differences to lead to variations in the ways in which
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35 children in the two countries are socialized. Hess, Azuma, Kashiwagi, Dickson, Nagano,
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37 and Holloway (1986) note that “In Japan, a child is thought to be good if he or she is
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39 ‘obedient’ (*sunao*), ‘mild and gentle’ (*otonasii*), and ‘self-controlled’ (*jiseishin ga aru*).
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41 In the United States, the ‘good child’ is assertive, socially competent with peers, and
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43 courteous” (p. 158). Moreover, parental reports contrasting children in these countries
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45 have shown that mothers in Japan rate their children as being shyer and less sociable than
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47 do mothers in the United States (Stevenson, Lee, Chen, Stigler, Hsu, & Kitamura, 1990).
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(For further discussion on differences between U.S. and Japanese socialization and child-rearing practices, see Conroy, Hess, Azuma, & Kashiwagi, 1980; LaFreniere, Masataka, Butovskaya, Chen, Auxiliadora-Dessen, & Atwanger, 2002; Lebra, 1994; Masataka, 2002; White, 1993; Zahn-Waxler, Friedman, Cole, Mizuta, & Hiruma, 1996).

Our previous studies suggest the involvement of a genetic predisposition in the expression of hypersociability in WS (Bellugi, Adolphs, Cassady, & Chiles, 1999; Doyle et al., 2004a, 2004b; Jones, et al., 2000). Thus, exploring sociability across cultures can provide keen insight into the interplay of temperament (in a disorder with a known genetic basis) and culture. While a number of research groups have been studying the cognitive strengths and weaknesses of individuals with WS in various countries, there have been no studies that directly explore social behavior in WS across cultures. 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.

METHOD

Participants. Participants included the parents of children living in Japan (24) and the United States (24). Twelve of the children in each sample had Williams syndrome (WS) and 12 were typically developing (TD). The children from each culture were matched individually with regard to age (between 3y, 3m and 13y, 7m) and gender (males and females are equally represented). Mean ages and standard deviations for each experimental group are displayed in Table 1.

----Table 1 about here----

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The U.S. participants were parents of children with WS attending a meeting of the Williams Syndrome Association, and the parents of TD children who attend school near the Salk Institute in California. The Japanese WS data were collected by Mr. Sugimoto, a member of the Japanese Williams Syndrome Association, who ensured that each person completing the questionnaire understood the materials, including the informed consent form, and was willing to participate. Japanese TD data were collected through the laboratory of co-author Dr. Nobuo Masataka of Kyoto University.

Procedures. The Salk Institute Sociability Questionnaire (SISQ) is an instrument specifically developed to assess specific aspects of social behavior commonly reported among people with WS first reported in Jones et al., 2000. The SISQ has been used in a variety of different contexts and across age groups as indicated previously. Moreover, the Salk Institute lab has collected data on over 80 adolescent and adult individuals with WS; of these, 44 had completed both the SISQ and another standardized parent-report instrument, the Multidimensional Personality Questionnaire (MPQ; Tellegen, 1985). The Salk 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, whereas there is no correlation with other MPQ measures, nor with IQ. The MPQ has also been used by Klein-Tasman with a different cohort of individuals with WS (Klein-Tasman & Mervis, 2003). Similar to the findings from the Salk team, Klein-Tasman and Mervis report using the MPQ that the distinctiveness of the WS personality appears to lie in the focus of WS 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

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3 phenotype provides the groundwork for the present study of cross cultural influences
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5 upon social behavior.
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8 To ensure consistency between the English and Japanese versions of the SISQ for
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10 the cross-cultural comparison in this study, two individuals, fluent and literate in both the
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12 English and Japanese languages, independently translated the SISQ from English into
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14 Japanese and then back-translated from Japanese to English.
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17 18 19 **RESULTS**

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21 The SISQ consists of both quantitative and qualitative item types. Quantitative
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23 items ask parents to rate their child's specific social behavior on a 7-point Likert scale.
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25 Qualitative items instead ask the parents to fill in a descriptive response. The items yield
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27 three subscales: tendency to approach strangers, tendency to approach familiars, and
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29 social emotional behavior (such as tendency to empathize with others, accuracy of
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31 emotional evaluations of others, eagerness to please others, and ability to remember
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33 names and faces of others). The three subscales together comprise Global Sociability.
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35 Items assessing social approach behavior consist of statements such as "How would you
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37 compare your child's tendency to approach strangers with an average child of the same
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39 age?" on a scale ranging from 1 ('approaches much less') to 7 ('approaches much more');
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41 or "How would you describe your child's general behavior in social situations?" on a
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43 scale ranging from 1 ('very shy and inhibited') to 7 ('extremely outgoing'). Qualitative
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45 items include "Describe your child's typical reactions when meeting someone for the first
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47 time (please give examples);" or "Give some examples of your child's socializing with
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49 strangers." The social approach items were grouped for analysis into two types: those
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51 that assess the child's tendency to approach family members or others encountered
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3 frequently (yielding an “Approach Familiars” score) and those that assess the child’s
4 tendency to approach people unknown to them (an “Approach Strangers” score. The
5 Social-Emotional score was the sum of four items; Approach Familiars score was the
6 sum of three items; Approach Strangers was the sum of five items; and the Global
7 Sociability score was the sum of all twelve items.
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15 Quantitative Data Analysis

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17 The quantitative data were analyzed by a 2x2 analysis of variance (ANOVA) with
18 Diagnostic Category (Williams syndrome children versus Typically Developing children)
19 and Culture (American versus Japanese) as independent variables and the summary
20 Global Sociability score as the dependent variable. Wilks’ Lambda criterion was used to
21 assess significance. This analysis revealed a significant main effect of Diagnostic
22 category (WS versus TD), $F(1, 44) = 18.07, p < .0001$ and a significant effect of
23 Culture (Japanese versus American) $F(1, 44) = 11.10, p < .002$. The Diagnostic
24 Category by Culture interaction failed to reach significance, $F = .42, p = > .05$, suggesting
25 that the pattern of differences between the diagnostic groups was similar across the two
26 cultures. Figure 1 shows the data distribution for the Global Sociability overall
27 composite score combining all questionnaire items, and the data for each of the three
28 subscales.
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46 ---Figure 1 about here---

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48 As can be seen from the figure, both the American and Japanese children with WS were
49 rated significantly higher on Global Sociability than were the typically developing
50 children; thus, there was a very strong effect for Diagnostic Category (WS or TD). At the
51 same time, there was also a significant effect of Culture, in that parents of children in the
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3 U.S. tended to rate their children as higher in Global Sociability than did parents of
4 children in Japan, regardless of their diagnostic category (WS or TD). Comparison of
5 scores across Cultures indicates a major difference for Approach Strangers, but not for
6 Approach Familiars (scores were nearly at ceiling) nor Social Emotional items.
7 Similarly, comparison of scores across Diagnostic Category indicates a major difference
8 for Approach Strangers, but not for the other two subscales. Overall, these results
9 illustrate that the significant between-group differences in Global Sociability were
10 primarily contributed to by higher ratings of “Approach Strangers” for both Diagnosis
11 and Culture.
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24 Qualitative Analysis of Sociability Data.

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27 The SISQ also asks parents to provide qualitative descriptions of their child in
28 various social situations with items such as, “Describe your child’s typical reactions
29 when meeting someone for the first time (please give examples).” Table 2 presents
30 sample qualitative responses from the two cultural and diagnostic groups selected at
31 different ages for this question.
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39 ---Table 2 about here---

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41 Qualitative responses were further examined in order to address the problems of
42 reference group effects (Heine, Lehman, Peng, & Greenholtz, 2002) and cultural
43 response biases that could be reflected in the use of a Likert-style questionnaire (Chen,
44 Lee & Stevenson, 1995). For this analysis, all responses across categories and cultures
45 were presented in randomized order not revealing their sources, and were translated into
46 both languages. Consistent with the analysis reported in Doyle et al. (2004a), the raters
47 were blind to the participants’ diagnostic classification and cultural membership. Two
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3 independent American, and two independent Japanese, raters, blind to the subjects'
4 identities, categorized all the responses into three behavioral groups: Shy, In-between or
5 Social. While all parents (12) of the U.S.-WS children provided an example to this
6 questionnaire form, only five parents of U.S.-TD children did so. There were 10
7 responses for the J-WS group, and 11 responses for the J-TD group. As a cross-cultural
8 check, we also asked two Japanese raters to categorize the qualitative responses in the
9 same way. Table 3 shows the frequencies of the ratings in percentages from both the
10 American and Japanese raters for the four participant groups within and across the two
11 cultures.
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25 ----Table 3 about here---

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27 Comparing the two, despite the small number of subjects, some trends emerge.
28 American and Japanese raters both categorized the responses from WS individuals as
29 more social regardless of culture, and both groups of raters categorized more of the TD
30 responses as “Shy.” However, even here, there is a trend toward cultural difference. The
31 Japanese raters categorized some of the WS responses as “Shy,” and some of the J-TD
32 responses as “Social,” whereas none of the American raters did so. Thus, the qualitative
33 data complemented the quantitative data, in that there is a strong effect for Diagnostic
34 Category, but some differences are observed among the cultural groups with respect to
35 the frequencies of responses characterized as Shy, Social, and In-between.
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50 **DISCUSSION**

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53 Williams Syndrome provides a compelling model for investigating the effects of
54 genotype, phenotype and environmental interactions. The genetic basis of WS is by now
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3 well known and documented in great detail (unlike, for example, the basis of Autism).
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5 The genetic phenotype thus involves absence of one copy of a small set of genes on
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7 chromosome 7, occurring in nearly all clinically identified WS (Korenberg et al, 2003;
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9 Morris & Mervis, 1999). Currently several groups are working to begin to link genotype
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11 and phenotype in Williams, and the hunt is on to link specific genes within the WS region
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13 with the medical characteristics, brain development and behavioral functions (see Doyle,
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15 et al., 2004a, 2004b; Korenberg, et al., 2003; Meyer-Lindenberg, Hariri, Munoz, Mervis,
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17 Mattay, & Morris, 2005; Meyer-Lindenberg, et al., 2006, for examples).
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22 In our studies, we highlight the consistency and variability of sociability in
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24 children with WS, which in turn allows for consideration of the expression of sociability
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26 not only in those Williams individuals with typical size deletions but also in Williams
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28 children with atypical deletions. Included in the large scale study of development of
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30 sociability in young WS, DS, and TD described above, was the data from a young child
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32 with WS who had a smaller deletion than is typical of the syndrome, retaining between
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34 one to three genes in the telomeric region that are almost invariably deleted in the
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36 “classic” deletion. This child had the typical medical and cognitive diagnostic
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38 characteristics for WS. However, her sociability scores especially those for approaching
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40 strangers, were significantly lower than the mean of the WS group, implicating specific
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42 genes in the emergence of this behavior in WS (Doyle, et al., 2004a, 2004b). The relative
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44 lack of variability in hypersociability among children with WS gleaned both from
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46 parental report and experimental as well as observational measures, combined with the
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48 early age of onset, and the results obtained for the child with the atypical deletion,
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50 suggests that the behavioral feature of hypersociability in approaching strangers may be
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3 strongly influenced by the genetic deletion (see also Klein-Tasman & Mervis, 2003), as
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5 well as tempered by environmental factors.
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8 To date, there have not been investigations of hypersociability across cultures in
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10 Williams syndrome. A cross-cultural study of expressiveness in language conducted by
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12 Reilly, Bernicot, Vicari, Lacroix, and Bellugi, (2003) with children in France, Italy, and
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14 the U.S. found that the form and intensity of Williams' social language behavior is
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16 influenced by the individual culture's social tenets for expressing sociability. Reilly, et
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18 al., reported that all groups of children with WS used more evaluative language than their
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20 TD controls; however, the effect was culturally specific. In other words, while
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22 individuals with WS across cultures and languages demonstrate atypically expressive use
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24 of language, this increase in evaluative language is relative to the baseline observed in the
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26 cultures of the respective individuals.
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32 The results of the present study support a genetic "proportional stamp" on the
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34 expression of social behavior in WS across cultures as children with WS in the U.S. and
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36 in Japan showed more affinity for approaching strangers and were rated higher on Global
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38 Sociability than TD children in their respective countries. The present study was
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40 conducted to examine, using a common instrument, social behavior among children with
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42 WS in Japan and the United States in order to investigate the ways in which social
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44 behavior is influenced by cultural expectations or mores. These results suggest that the
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46 WS social and genetic phenotype may influence the degree of expression of affiliative
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48 behaviors towards strangers, even among children for whom a cultural expectation of
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50 cautiousness towards strangers exists. We observed, by both quantitative and qualitative
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3 measures, nature's stamp on culture's milieus. In other words, the expression of
4 sociability in WS may not be "either/or" phenotype/culture, it is rather "both/and."
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8 Alternative explanations for the results reported here may still be made. It is
9 interesting to note in Table 2 that the examples given for specific behaviors described by
10 parents of children with WS to illustrate approaching and socializing with strangers are
11 very similar, regardless of culture. Nevertheless, Japanese parents rated their children
12 lower on the 7-point scale than did U.S. parents; thus, an alternative explanation for the
13 difference in quantitative scores obtained for Japanese children with WS as compared to
14 their U.S. peers may be that cultural influence is exerted most on parent's *ratings* rather
15 than on the expression of the behavior itself. Perhaps the stigma of having a "different"
16 child in Japan affected the ways in which parents ranked their child's degree of
17 sociability. Thus, further cross-cultural observational studies of children with WS
18 interacting with strangers are needed to resolve possible discrepancies caused by reliance
19 on parental reports in order to gauge the interplay of phenotype and culture in WS.
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36 This survey of WS in U.S. and Japanese cultures examines the social behaviors
37 that typify WS, and finds that despite the differences in upbringing between the two
38 cultures, individuals with WS are found to be more sociable in approaching strangers
39 than typically developing individuals, thus, demonstrating the impact of the WS
40 phenotype. The unique nature of WS allows us to investigate the dual influences of
41 nature and nurture; thus, continuing to examine Williams syndrome across cultures will
42 be an important avenue for further exploration.
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References

- Battin, J., Lacombe, D., Taine, L., & Goizet, C. (2000). Williams syndrome (microdeletion 7q11.23), model of behavioral phenotype. *Bulletin de l'Academie Nationale de Medecine (Paris)*, *184*, 105-115.
- Bellugi, U., Adolphs, R., Cassady, C., & Chiles, M. (1999). Towards the neural basis for hypersociability in a genetic syndrome. *Neuroreport*, *10*, 1653-1657.
- Bellugi, U., Lichtenberger, L., Jones, W., Lai, Z., & St George, M. (2000). The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*, *12*, 7-29.
- Bjornstad, P. (1994). Williams-Beuren syndrome in Norway. *Tidsskr Nor Laegeforen*, *114*, 25-8. (Article in Norwegian.)
- Chen, C., Lee, S.Y., & Stevenson, H.W. (1995). Response style and cross-cultural comparisons of rating scales among East Asian and North American students. *Psychological Science*, *6*, 170-175.
- Conroy, M., Hess, R., Azuma, H., & Kashiwagi, K. (1980). Maternal strategies for regulating children's behavior: Japanese and American families. *Journal of Cross-Cultural Psychology*, *11*, 153-172.
- Doyle, T.F., Bellugi, U., Korenberg, J.R. & Graham, J. (2004a). "Everybody in the world is my friend": Hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics*, *124*, 263-273.
- Doyle, T. F., Bellugi, U., Reiss, A. L., Galaburda, A. M., Mills, D. L., & Korenberg, J. R. (2004b). Genes, neural systems, and cognition: Social behavior of children with Williams syndrome: Observing genes at play (Abstract/Poster). *Society for Neuroscience 34th Annual Meeting Abstracts*, 666.11.
- Einfeld, S.L., Tonge, B.J., & Florio, T. 1997. Behavioral and emotional disturbance in individuals with Williams syndrome. *American Journal of Mental Retardation*, *102*, 45-53.
- Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W., Sternes, K., & Spallone, P. (1993). Hemizyosity at the elastin locus in a developmental disorder, Williams syndrome. *Nature Genetics*, *5*, 11-16.

- 1
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3 Fatimilehin, I. & Nadirshaw, Z. (1994). A cross-cultural study of parental attitudes and
4 beliefs about learning disability (mental handicap). *Mental Handicap Research*, 7,
5 202-207.
6
7
8 Franceschini, P., Guala, A., Vardeu, M. P., Signorile, F., Franceschini, D., &
9 Mastroiacovo, P. (1996). The Williams syndrome: An Italian collaborative study.
10 *Minerva Pediatrica*, 48, 421-428.
11
12
13 Gosch, A., & Pankau R. (1994). Social-emotional and behavioral adjustment in children
14 with Williams syndrome. *American Journal of Medical Genetics*, 53, 335-339.
15
16
17 Heine, S.J., Lehman, D.R., Peng, K., & Greenholtz, J. (2002). What's wrong with cross-
18 cultural comparisons of subjective Likert scales? The reference group effect.
19 *Journal of Personality and Social Psychology*, 82, 903-918.
20
21
22 Hess, R., Azuma, H., Kashiwagi, K., Dickson, W P., Nagano, S., & Holloway, S.
23 (1986). Family influences on school readiness and achievement in Japan and the
24 United States: An overview of a longitudinal study. In H. Stevenson, H. Azuma,
25 & K. Hakuta (Eds.), *Child development and education in Japan* (pp. 147-166).
26 New York: Freeman.
27
28
29 Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., & Lincoln, A. (2000). II.
30 Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*, 12, 30-
31 46.
32
33
34 Klein-Tasman, B. P., & Mervis, C. B. (2003). Distinctive personality characteristics of 8-
35 , 9-, and 10-year-olds with Williams syndrome. *Developmental Neuropsychology*,
36 23, 269-290.
37
38
39 Korenberg, J. R., Chen, X. N., Hirota, H., Lai, Z., Bellugi, U., & Burian, D., (2000). VI.
40 Genome structure and cognitive map of Williams syndrome. *Journal of Cognitive*
41 *Neuroscience*, 12, 89-107.
42
43
44 Korenberg, J.R., Bellugi, U., Salandanan, L.S., Mills, D.L., & Reiss, A.L. (2003).
45 Williams syndrome: A neurogenetic model of human behavior. In *Encyclopedia*
46 *of the Human Genome*. London, England: The Nature Publishing Group.
47
48
49 Kotzot, D., Bernasconi, F., Brecevic, L., Robinson, W. P., Kiss, P., & Kosztolanyi, G.
50 (1995). Phenotype of the Williams-beuren syndrome associated with
51 hemizyosity at the elastin locus. *European Journal of Pediatrics*, 154, 477-482.
52
53
54 LaFreniere, P., Masataka, N., Butovskaya, M., Chen, Q., Auxiliadora-Dessen, M., &
55 Atwanger, K. (2002). Cross-cultural analysis of social competence and behavior
56 problems in preschoolers. *Early Education and Development*, 13, 187-199.
57
58
59
60

- 1
2
3 Lebra, T.S. (1994). Mother and child in Japanese socialization: A Japan-U.S. comparison.
4 In P. Greenfield & R. Cocking (Eds.), *Cross-cultural roots of minority child*
5 *development* (pp. 259-274). Hillsdale, NJ: Erlbaum.
6
7
8 Markus, H., & Kitayama, S. (1991). Culture and the self implications for cognition,
9 emotion, and motivation. *Psychological Review*, 98, 224-253.
10
11
12 Masataka, N. (2002). Low anger-aggression and anxiety-withdrawal characteristic to
13 preschoolers in Japanese society where 'Hikkikomori' is becoming a major social
14 problem. *Early Education and Development*, 13, 187-199.
15
16
17 Masataka, N. (2001). Working memory and sentence comprehension in children with
18 Williams syndrome. *Cognitive Studies: Bulletin of the Japanese Cognitive*
19 *Science Society*, 8, 25-36.
20
21
22 Mervis, C.B., & Klein-Tasman, B.P. (2000a). Williams syndrome: cognition,
23 personality, and adaptive behavior. *Mental Retardation & Developmental*
24 *Disabilities Research Review*, 6, 148-158.
25
26
27 Mervis, C.B., Robinson, B.F., Bertrand, J., Morris, C.A., Klein-Tasman, B.P., &
28 Armstrong, S.C. (2000b). The Williams syndrome cognitive profile. *Brain and*
29 *Cognition*, 44, 604-628.
30
31
32 Meyer-Lindenberg, A., Hariri, A. R., Munoz, K. E., Mervis, C. B., Mattay, V. S., &
33 Morris, C. A. (2005). Neural correlates of genetically abnormal social cognition in
34 Williams syndrome. *Nature Neuroscience*, 8, 991-993.
35
36
37 Meyer-Lindenberg, A., Mervis, C. B., & Faith Berman, K. (2006). Neural mechanisms in
38 Williams syndrome: A unique window to genetic influences on cognition and
39 behaviour. *Nature Reviews. Neuroscience*, 7, 380-393.
40
41
42 Morris, C. & Mervis, B. (1999). Williams syndrome. In S. Goldstein & C. Reynolds
43 (Eds.) *Handbook of neurodevelopmental and genetic disorders in children* (pp.
44 555-590). New York: Guilford Publications.
45
46
47 Nakaji, A., Kawame, Y., Nagai C., & Iwata, M. (2001). Clinical features of a senior
48 patient with Williams syndrome. *Rinhso Shinkeigaku*, 41, 592-8.
49
50
51 Perez Jurado, L.A., Peoples, R., Kaplan, P., Hamel, B.C., & Franke, U. (1996).
52 Molecular definition of the chromosome 7 deletion in Williams syndrome and
53 parent-of-origin effects on growth. *American Journal of Human Genetics*, 59,
54 781-792.
55
56
57 Plomin, R., DeFries, J.C., McClearn, G.E., & McGuffin, P. (2001). *Behavioral genetics*,
58 *4th ed.* New York: Worth.
59
60

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57
58
59
60
- Reilly, J., Bernicot, J., Vicari, S., Lacroix, A., & Bellugi, U. (2003). Narratives in children with Williams syndrome: A cross linguistic perspective. In D. Ravid & H.B. Shyldkrot (Eds.), *Perspectives on language and language development* (pp. 303-312). Dordrecht, Netherlands: Kluwer Publishers.
- Ruangdaraganon, N., Tocharoentanaphol, C., Kotchabhakdi, N., & Khowsathit, P. (1999). Williams syndrome and the elastin gene in Thai patients. *Journal of the Medical Association of Thailand = Chotmai het Thangphaet*, 82, 174-178.
- Stevenson, H., Lucker, G., Lee, S., & Stigler, J. (1987). Poor readers in three cultures. In Super, C. (Ed.), *The Role of Culture in Developmental Disorder* (pp. 153 – 177). San Diego: Academic Press, Inc.
- Stevenson, H. W., Lee, S-Y., Chen, C., Stigler, J.W., Hsu, C-C., & Kitamura, S., (1990). Contexts of achievement: a study of American, Chinese, and Japanese children. *Monograph of the Society for Research in Child Development, Vol. 55*, 80-81.
- Tellegen, A. (1985). Structures of mood and personality and their relevance to assessing anxiety, with an emphasis on self-report. In A. H. Tuma & J. D. Maser (Eds.), *Anxiety and the anxiety disorders* (pp. 681–716). Hillsdale, NJ: Lawrence Erlbaum Associates, Inc.
- Teo, S., Chan, D., Yong, M., Ng, I., Wong, K., & Knight, L. (1997). Williams syndrome--the Singapore General Hospital experience. *Department of Paediatric Medicine, Singapore General Hospital, Singapore. Academy of Medicine Singapore*, 26, 360-364.
- Triandis, H. C. (1989). The self and social behavior in differing cultural contexts. *Psychological Review*, 96, 506-520.
- Triandis, H. C. (1996). The psychological measurement of cultural syndromes. *American Psychologist*, 51, 407-415.
- Udwin, O., Yule, W., & Martin, N. (1987). Cognitive abilities and behavioural characteristics of children with idiopathic infantile hypercalcaemia. *Journal of Child Psychology and Psychiatry*, 28, 297-309.
- van Hagen, J. M., Govaerts, L. C., de Coo, I. F., Gille, J. J., Nieuwint, A. W., & Madan, K. (2001). Williams syndrome: New insights into genetic etiology, pathogenesis and clinical aspects [Williams-syndroom: nieuwe inzichten in genetische etiologie, pathogenese en kliniek]. *Nederlands Tijdschrift Voor Geneeskunde*, 145, 396-400.
- Von Armin, G. & Engel, P. (1964). Mental retardation related to hypercalcaemia. *Developmental Medicine and Child Neurology*, 6, 366-377.

1
2
3 White, M. (1993). *The material child: Coming of age in Japan and America*. New York:
4 Free Press.
5

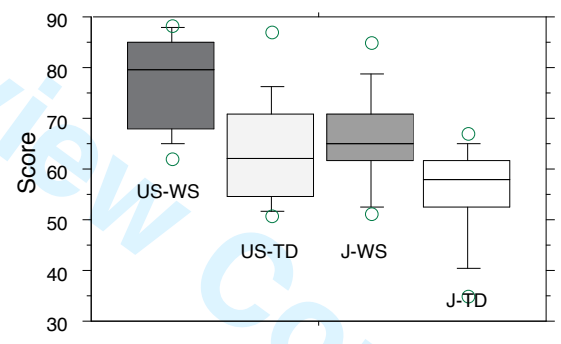
6
7 Zahn-Waxler, C., Friedman, R.J., Cole, P.M., Mizuta, I., & Hiruma, N. (1996). Japanese
8 and United States preschool children's responses to conflict and stress. *Child*
9 *Development*, 67, 2462-2477.
10
11
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15
16
17
18
19
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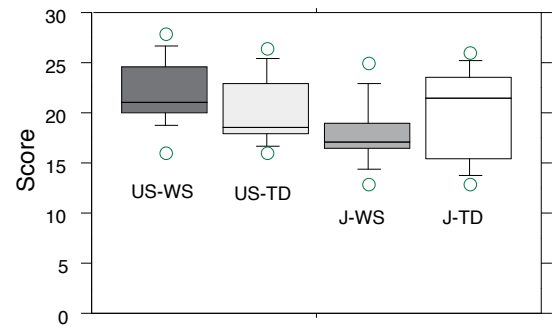
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US-WS US-TD J-WS J-TD

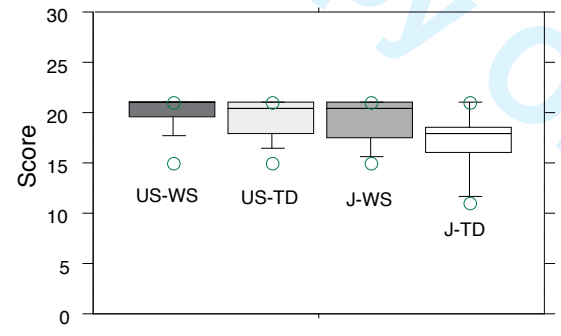
Global Sociability



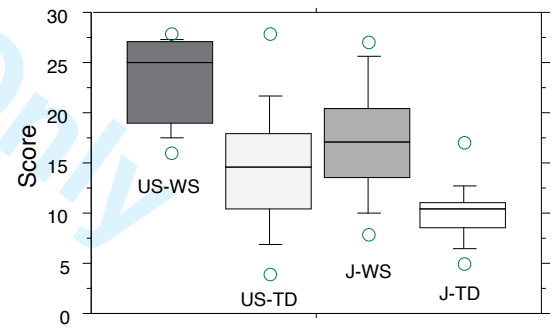
Social Emotional



Approach Familiars



Approach Strangers



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6 **Figure 1.** Summary of Comparisons across Diagnostic Category (Williams versus
7 Typically Developing) and Culture (Japanese versus US) for the quantitative analysis of
8 the SISQ. Graphs display horizontal lines at the 10th, 25th, 50th, 75th, and 90th centiles.
9 The top box shows Global Sociability, encompassing all parts of the SISQ combined.
10 The Japanese WS mean scores are significantly higher than the Japanese TD means; and
11 the US-WS means are significantly higher than the US-TD means, a major effect of
12 Diagnostic Category. Whereas the US-WS means are significantly higher than all other
13 means, the J-WS are almost on a par with the US-TD, and there is also a significant effect
14 of Culture (US versus Japanese) as well. These patterns do not hold for Social Emotional
15 items. Note that on Approach Familiar items, the four groups are almost
16 indistinguishable, at near ceiling. On the Approach Strangers items, the significant
17 effects of both Diagnostic Category and Culture are strongly observed.
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Table 1. Age Means and Standard Deviations for Experimental Groups

		WS	TD
Japan	Mean (SD)	7.22 (3.48)	7.35 (3.37)
United States	Mean (SD)	7.30 (3.04)	7.23 (3.10)

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Table 2. Table 2. 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 to 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, responds 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 sometime.	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.	H. is always engaging. She asks numerous questions and inquires as to a person’s living arrangements. She has invited strangers to our home for dinner.

Table 3. Summary of Data from Japanese and U.S. Raters for Item 5. Responses were randomized across Diagnostic Category and Culture. Raters from US and Japan, blind to the purposes of the study, were asked to categorize each response as “Shy”, “Social” or “In between.”

US Raters	Percentage of responses			
	J-TD	J-WS	US-TD	US-WS
Characterized as				
Shy	45	0	30	0
In between	55	35	50	8
Social	0	65	20	92

Japanese Raters	Percentage of responses			
	J-TD	J-WS	US-TD	US-WS
Characterized as				
Shy	41	30	20	4
In between	45	15	50	4
Social	14	55	30	92

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