

Behavioral Assessment of Social Anxiety in Females with Turner or Fragile X Syndrome

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Social skills impairment in children with Turner or fragile X syndrome has been documented using parental reports. Anxiety, shyness, and difficulty understanding social cues have been reported for females with Turner syndrome; whereas social withdrawal, avoidance of social interactions, and anxiety are often reported for females with fragile X syndrome. Social interaction anxiety in these two populations may be a framework for understanding the difficulty these children experience in social situations. In the present study, 29 females with Turner syndrome and 21 females with fragile X syndrome ages 6–22 years were compared to females in a comparison group, on a videotaped role-play interaction. Behavioral indices examined included eye-contact maintenance, duration of speech, and body discomfort as observed during the brief interaction. Three of eight such behavioral measures of social skills differentiated the participant groups from each other. Specifically, participants with fragile X required more time to initiate interactions than did participants in either of the remaining groups; and females with Turner syndrome made fewer facial movements than did females in the fragile X or comparison group. Self-report and parental ratings did not suggest higher levels of anxiety in females with Turner or fragile X syndrome, but did reflect higher levels of social difficulty. The authors discuss these findings in terms of understanding the nature of social dysfunction in females with Turner or fragile X syndrome.

KEY WORDS: Fragile X females; Turner syndrome; social anxiety.

INTRODUCTION

Turner and fragile X syndromes are developmental disorders associated with neuropsychological and behavioral deficits. Each of these syndromes has been posited as a potential etiology of nonverbal learning disability (Tsatsanis & Rourke, 1995), in view of associated neurocognitive and social skills difficulties. In the present study, we examined potential group differences on very limited aspects of social functioning, using a behavioral approach that has not yet been used

to study social functioning in females with Turner or fragile X syndrome. Specifically, we evaluated behavioral indices of anxiety during the initiation and maintenance of a simulated brief social interaction with an unfamiliar person. The rationale for this study is presented below, following a review of each disorder.

Turner and Fragile X Syndromes: Genotypes and Phenotypes

Turner and fragile X syndromes have similarities and differences at both the genotype and phenotype levels. Turner syndrome results from the total or partial absence of one of the two X chromosomes typically present in females, whereas fragile X syndrome is most often associated with a single gene mutation at location Xq27.3 (Verkerk *et al.*, 1991). Turner syndrome occurs exclusively in females (see Ross & Zinn, 1999;

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Rovet & Buchanan, 1999, for a review) and is not associated with mental retardation. Fragile X occurs in both males and females. The vast majority of males (Bailey, Hatton, & Skinner, 1998) and approximately half of females (Rousseau *et al.*, 1994) with fragile X have mental retardation (see Hagerman, 1999; Mazzocco, 2000, for a review). In view of marked contrasts in rates of mental retardation across males and females with fragile X and the greater comorbidity of social problems among individuals with mental retardation, the sample included in the present study is limited to females without mental retardation who had IQ scores above 70.

Neuropsychological characteristics of females with Turner or fragile X syndrome include visual-spatial processing difficulties (Mazzocco, Baumgardner, Freund, & Reiss, 1998; McCauley, Kay, Ito, & Treder, 1987; Reiss & Freund, 1990), dyscalculia, right-left disorientation, constructional dyspraxia, and attention problems (Reiss & Freund, 1990; Rovet, 1993; Turk, 1992; Lesniak-Karpiak, Barakat, & Ross, 2001). Expressive language deficits have also been noted in persons with fragile X (Fisch, 1993; Hagerman & Sobesky, 1989), and verbal fluency deficits have been reported in individuals with Turner syndrome (Waber, 1979). In addition to neuropsychological deficits, females with either syndrome may exhibit social or emotional problems, including anxiety or depression (McCauley, Ito, & Kay, 1986; McCauley, Feuillan, Kushner, & Ross, 2001; Lachiewicz, 1992; Lachiewicz & Dawson, 1994; Ross *et al.*, 1996; Rovet & Ireland, 1994).

Parents of children and adolescents with fragile X rate their children as more anxious or withdrawn, or as showing more avoidant behaviors than children without this disorder, when completing parent rating questionnaires. Lachiewicz (1992) reported that social difficulties are discernable in children with fragile X as early as during the preschool years. During social interactions, individuals with fragile X demonstrate poor eye contact, shyness, difficulty initiating and maintaining conversations, hand flapping, and tangential and perseverative speech (Hagerman & Sobesky, 1989; Turk, 1992). Some of these aberrant verbal and nonverbal behaviors are also observed during social interactions among individuals with social phobia or other forms of social dysfunction (Stravynski & Greenberg, 1989). The presence of such behaviors in children with fragile X suggests that their social impairments may be related to social discomfort or social anxiety.

As a group, females with Turner syndrome also demonstrate impaired psychosocial profiles as assessed by self-report measures or parental ratings of child be-

haviors (Bender, Puck, Salbenblatt, & Robinson, 1984; McCauley *et al.*, 1986; McCauley *et al.*, 2001; Ross *et al.*, 1996; Rovet, 1993; Rovet & Ireland, 1994). Females with Turner syndrome receive lower parental ratings than do constitutionally short-statured girls on the Child Behavior Checklist subscales of social competence (McCauley *et al.*, 1986, 2001). Although physical appearance may be related to greater social/emotional disturbance in females with Turner syndrome, short stature alone does not fully account for the severity of social impairments observed in these females (Holmes, Karlsson, & Thompson, 1985). During peer interactions, females with Turner syndrome require more structure than females in a comparison group, have more difficulty understanding social cues, and show social withdrawal or anxiety (Downey, Ehrhardt, Gruen, Bell, & Morishima, 1989; Holmes *et al.*, 1985; McCauley *et al.*, 1986).

Despite the empirical support of social impairment among females with Turner or fragile X syndrome, the specificity of such impairments is as yet undetermined, as are the reasons for social difficulties. Identification of these factors may influence treatment recommendations for females with Turner or fragile X syndrome, and may have a role in developing models of social development and function.

There are several possible explanations for the social difficulties observed in females with Turner or fragile X syndrome. Social problems may be secondary to deficits with facial affective processing, language expression difficulties, or a limited repertoire of social behaviors (McCauley *et al.*, 1987; Reiss & Freund, 1990). Alternatively, problems with social interactions may be related specifically to social anxiety. Belser and Sudhalter (1995) hypothesized that anxious behaviors observed in individuals with fragile X could be related to abnormal regulation of arousal. Recent evidence shows atypical psychophysiological indices of arousal in young boys with fragile X syndrome (Roberts, Boccia, Bailey, Hatton, & Skinner, 2001), similar to atypical psychophysiological arousal observed in females with fragile X (Keysor, Mazzocco, McLeod, & Hoehn-Saric, 2001). Psychophysiological indices of arousal differ among girls with Turner or fragile X syndrome, relative to a comparison group, although to a different degree and under different conditions (Keysor *et al.*, 2001). Yet the available psychophysiological data do not include behavioral indicators of social anxiety. In the present study, behavioral markers of social anxiety were examined to explore their potential role in social functioning among females with fragile X or Turner syndrome.

Specification of Social Anxiety

According to the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV; American Psychiatric Association [APA], 1994) social anxiety is “fear of social or performance situations” (p. 411). Social anxiety may be manifested as social *interaction* anxiety (e.g., fear of talking with a stranger) or social *performance* anxiety (e.g., fear of public speaking). Although empirical validation of these and other social anxiety subtypes continues to be a topic of investigation (e.g., Herbert, Hope, & Bellack, 1992, Last, Perrin, Hersen, & Kazdin, 1992; Safren *et al.*, 1999), in the present study we examine behaviors that are consistent with the concept of social interaction anxiety.

In early childhood, avoidance of social interactions may take the form of solitary passive play (i.e., quiet exploration of objects/solitary constructive play), which is viewed as both acceptable and developmentally appropriate. It differs from reticent behavior, which is marked by prolonged looking without accompanying play and social approach-avoidance behaviors. Reticence is believed to reflect enduring temperamental characteristics and to be associated with behavioral markers of anxiety in preschool-age children (Coplan, Rubin, Fox, Calkins, & Stewart, 1994). Asendorf (1991) reported that by middle childhood, these initially differentiated behaviors (i.e., reticence and solitary passive play) merge and are an index of social anxiety. In the present study, this potential developmental confound was avoided by the exclusion of preschool-age participants.

Tools used to describe or quantify social anxiety typically include structured psychiatric interviews, clinical rating scales, and parental ratings (Bernstein, Borchardt, & Perwien, 1996). In view of the subjective nature of social anxiety, it is important to also include measures that supplement parent reports, such as direct assessment of social skills via observations of social behaviors in either naturalistic settings or role-plays of representative social situations (Bernstein *et al.*, 1995). Although analogue situations may be limited in scope and in how role-plays generalize to real-life social situations, they allow for empirically controlled observation and assessment of social skills. These methods have been successfully used in studies of social anxiety in adults (e.g., Herbert *et al.*, 1992) and very young children (e.g., Coplan *et al.*, 1994). For example, Herbert *et al.* (1992) reported that behavioral markers of social skills during 180-second simulated role-plays differentiated socially phobic persons from individuals with other diagnoses. Fydrich, Chambless, Perry, Buergener, and Beazley (1998) also provided preliminary support

for reliability and validity of behavioral markers in evaluating skills during 180-second role plays. The target behavioral markers in the present study were believed to represent trait (i.e., stable over time and settings) rather than state (i.e., passing) characteristics, which increases the likelihood of their emergence in a brief social situation. In light of paucity of research on behavioral markers of social anxiety in children, empirical background for the present study was drawn primarily from behavioral assessments of socially anxious adults. Previous behavioral assessment studies of social anxiety were often based on methods that involved global quantitative evaluation of anxiety (see Monti *et al.*, 1984 for reviews); component-like assessment based on behavior frequency counts (Monti *et al.*, 1984); or qualitative rating of behavior categories (Fydrich *et al.*, 1998). In the present study, the measures we used included frequency counts and the duration of target behaviors.

In summary, the present study is a preliminary attempt to clarify and quantify a specific aspect of social behavior—conversation initiation and maintenance—during a brief interaction. The primary aims were to investigate anxiety ratings in participants with Turner or fragile X syndrome using standardized anxiety measures (parent ratings and self-reports) and behavioral markers during a simulated role-play with adult strangers, and to examine the relations between the behavioral markers and standardized behavioral ratings. The contribution of the current study is the use of objective behavioral markers during potentially anxiety-evoking social situations; this approach is novel to studies of females with fragile X and Turner syndrome, although not completely novel to studies of social anxiety or males with fragile X (Belser & Sudhalter, 1995). We predicted that females with Turner or fragile X syndrome would demonstrate social interaction difficulties as measured by standardized anxiety scales, and that these difficulties would differ significantly from behaviors of females in a comparison group. Moreover, we predicted that behavioral markers associated with social anxiety, such as eye contact, speech duration, and body movement, would differentiate these two genetic groups from each other and from the comparison group.

METHOD

Participants

The participants in this study were enrolled in a larger project examining cognitive, social, and emotional development of children with Turner or fragile X syndrome. The sample included 29 females with

Turner syndrome, ages 7–22 years ($M = 12.92$ years, $SD = 4.35$); 21 females with fragile X syndrome, ages 7–22 years ($M = 15.53$, $SD = 4.22$); and 34 females in a comparison group, ages 6–20 years ($M = 12.60$, $SD = 3.34$). See Table I for a summary of the participants' characteristics.

All females with fragile X syndrome had the full mutation confirmed through Southern blot DNA testing, and Turner syndrome was based on a karyotype analysis. Seventy-two percent of participants with Turner syndrome were identified with 45,X karyotype (i.e., total absence of the second X chromosome) and the remainder had mosaic karyotype. The females with Turner or fragile X syndrome were recruited through support groups, newsletter announcements, and referrals from physicians. Comparison participants included nonaffected siblings of females from either of the two genetic groups and females recruited by newsletter announcements through community organizations. All participants received remuneration for participation.

Procedures

All participants individually completed a battery of psychological assessments over a 2-day period. The behavioral assessment of interest in this report typically occurred on the second day. A parent completed standardized self-reports that were used to assess behavioral, social, and emotional functioning of their children. Participants' social skills and level of anxiety were assessed with simulated social interactions, via the role-plays described below. In addition, before and after participation in the role-plays, participants were asked to complete several standardized self-report question-

naires used to assess general anxiety, social anxiety, or emotional functioning.

All participants were observed individually in a set of simulated videotaped role plays, to directly assess the participants' behaviors in potentially noxious social situations. A total of five scenarios had been employed, but only two involved initiating and maintaining a conversation. The following is the order in which these scenes were presented during the study: the first two scenarios involved reading silently or reading aloud to an examiner with whom the participant was familiar. The third and fourth scenarios involved conversing with an unfamiliar adult (conversation-based scenarios), and the fifth scenario involved presenting a speech to two familiar adults. The present report is based on analysis of the first of two conversation-based scenarios.

The rationale for selecting only the first conversation-based scenario was to examine responses of participants in a *novel*, potentially anxiety-provoking social situation, versus evaluating participants' behaviors in nonanxiety-evoking scenes (i.e., reading silently), scenarios with familiar adults (i.e., speech), or scenarios that were not novel because of prior exposure to a similar simulated social situation (i.e., the second conversation-based scenario). Also, because the focus of this study was on early initiation and maintenance of conversation with an unfamiliar person, it was important to look at behavior during the first such scenario imposed upon each participant. Evidence that social anxiety-related behaviors represent a trait versus state characteristic (Coplan *et al.*, 1994) diminishes the importance of assessing the second conversation-based scenario; however, it would be interesting to observe for consistency over time, and to do so would address

Table I. Participants' Characteristics (Group Means and Standard Deviations) for Younger (< 11 Years) and Older (> 11 Years) Subgroups

Variables	Turner syndrome		Fragile X		Comparison group	
	Younger ($n = 15$)	Older ($n = 14$)	Younger ($n = 5$)	Older ($n = 16$)	Younger ($n = 17$)	Older ($n = 17$)
Age						
<i>M</i>	9.57	16.51	10.14	17.21	9.91	15.29
<i>SD</i>	1.37	3.44	1.87	3.19	1.58	2.25
Full Scale IQ						
<i>M</i>	92.07	94.64	97.40	92.00	106.24	106.76
<i>SD</i>	11.85	16.39	15.96	16.36	13.68	15.48
Verbal IQ						
<i>M</i>	99.73	101.21	100.20	95.13	108.41	107.82
<i>SD</i>	12.04	14.67	14.97	15.28	12.36	12.24

a different set of research questions than those examined in the present study.

When the participant first entered the room for the set of role-play activities, the examiner turned on a video camera used to record each of the five role plays. Before each role play, the participant was given a brief overview of the role-play procedure and was asked if she had any questions. For the first conversation-based role play (hereafter referred to as the "target role play"), each participant was instructed to initiate and maintain a conversation with a "stranger." The participant was asked to assume the role of a neighbor, and was informed that a stranger would assume a role of a person who had just moved into the child's neighborhood. When the participant appeared ready, the examiner called for the stranger, who entered the room and sat facing the participant.

The stranger was a male or female staff member at the research center who had no prior interaction with the participant. Before involvement with the present study, each stranger was instructed to respond minimally, to use neutral affective tone when responding, and to place the burden of maintaining conversation on the participant.

Upon arrival of the stranger, the participant was asked to begin. After 105 to 120 seconds, the examiner said, "stop." All recorded videotapes were of sessions of 105 seconds to 120 seconds in duration. To secure consistency in length of interactions for all participants, only those behaviors that occurred during the initial 105-second period of role play were coded. Specifically, target behaviors described below were observed and coded during seven 15-second intervals. For each behavioral marker described below, the participants could receive a total score of 0 to 7; higher scores reflected more frequent target behavior. To receive a score of 1 during each 15-second interval, the target behavior occurred at least once.

Before coding the tapes, the first two authors of the paper viewed several practice tapes in order to reach agreement in operationalizing and coding behaviors. Training was completed after approximately 10 hours of viewing and scoring practice tapes of the participants whose scores were not included in final analyses. For analyses, all of the role-play behaviors were rated and coded randomly by the first author, who was blind to the participants' group membership.

Measures

Behavioral Markers of Anxiety

Behavioral markers of anxiety were derived on the basis of a review of behavioral assessment research.

The following eight behavioral markers were selected as potential indicators of social anxiety or social skills dysfunction: (1) Total duration of silence, defined as the total time of noninteraction during the 105 target role play. (2) Time to first utterance, defined as the amount of time elapsed after the examiner gave signal to initiate conversation until the participant first produced an utterance. (3) Number of pauses, which reflected the frequency of breaks in conversations flow during the role play; (4) Eye contact avoidance, defined as breaking eye contact with the examiner or by complete avoidance of eye contact during an interval. (5) Rigid body posture, defined as rigid or tense sitting with minimal changes in body positions. (6) Fidgeting, defined as frequent or abrupt changes in bodily position such as rocking or leaning in different directions. (7) Wringing hands, which reflected repeated movements of the upper extremities in a rapid or jerky manner. and (8) Facial movements, such as jaw clenching, biting lips, or scowling, that reflected discomfort.

Parental and Child Standardized Questionnaires

The level of anxiety or social competence of the participants was assessed with the Child Behavior Checklist (CBCL; Achenbach, 1991), a widely used standardized measure for parental assessment of children's socioemotional and behavioral functioning. The CBCL yields T scores for eight narrow-band scales. In the present study scores from the Anxiety, Withdrawal, and Social Problems scales were used in the analyses.

In addition to parental ratings, participants completed self-report measures of psychosocial functioning and general or social anxiety. The standardized anxiety measures administered to the participants included Revised Children's Manifest Anxiety Scale (RCMAS; Reynolds & Richmond, 1978) and Social Phobia and Anxiety Inventory for Children (SPAI-C; Beidel, Turner, & Morris, 1988). The RCMAS is a self-report questionnaire used to assess the subjective severity of anxiety experienced by children. In the present study the total anxiety score was used in the analyses. The SPAI-C is a self-report instrument designed for children ages 8 to 14 to measure a child's subjective ratings of somatic, cognitive, and behavioral symptoms of social anxiety. Participants over 14 years of age completed the Social Phobia and Anxiety Inventory (SPAI; Turner, Beidel, & Dancu, 1996), an adult version. However, because fewer participants (23 of 84) completed the SPAI, and because SPAI and SPAI-C scores are not interchangeable, only valid SPAI-C scores were used in analyses of social phobia scores. All participants

were instructed how to complete the forms, and younger children with lower reading levels had the forms read to them.

RESULTS

Preliminary Analyses

Although the effects of age and verbal IQ (derived from the Wechsler Intelligence Scale–Revised) were not of primary interest in this study, we carried out preliminary analyses to determine whether either factor differed across the three participant groups; or whether either factor was correlated with our behavioral measures. Two one-way analyses of variance were conducted with group membership as an independent variable and VIQ scores and age of participants as dependent variables. Significant difference between the groups emerged on IQ measures [$F(2, 81) = 5.69, p < .005$]. Participants with fragile X had lower VIQ scores ($M = 96.33, SD = 14.99$) than the comparison group ($M = 108.12, SD = 12.12$), although the group mean was well within the average range. There was no difference in VIQ score between participants with fragile X or Turner syndrome ($M = 100.44, SD = 13.15$). Females with fragile X were also significantly older ($M = 15.53$ years, $SD = 4.23$) than the comparison group ($M = 12.60$ years, $SD = 3.36$), [$F(2, 81) = 3.98, p < .05$]; however, they did not differ on age from participants with Turner syndrome ($M = 12.92$ years, $SD = 4.35$).

Because behavioral markers were not normally distributed, nonparametric tests were used for subsequent analyses. To determine if age and VIQ were related to behavioral markers of anxiety, Spearman rank correlations were computed between the eight behavioral markers and these two variables. Because data for all behavioral markers was not available for all participants, subgroups of data were used per analysis, as indicated in Table II. There was no association between VIQ scores and any of the eight behavioral markers (Table III). Therefore, VIQ scores were not addressed in subsequent analyses. In contrast, Spearman rank correlations indicated significant relations between age and three of the behavioral markers, including total amount of silence, eye contact avoidance, and fidgeting (Table III).

To address possible age effects, the total sample was divided into younger (≤ 11 years, $n = 37$) and older (> 11 years, $n = 47$) participants for selected analyses. In view of the small sample sizes, it was not possible to examine potential age by group interactions, or effects at specific age level, by subdividing the partici-

Table II. The Number of Participants in Each Group with Data for Behavioral Markers

Measures	Turner syndrome	Fragile X	Comparison group
Eye contact avoidance	18	16	20
Lip biting	21	16	21
Wringing hands	19	16	20
Fidgetiness	19	16	20
Rigidity	19	16	20
Number of pauses	21	16	21
RT to first utterance	21	16	20
Duration of silence	21	16	20

pants groups by age. Therefore, comparisons were made between the subgroups of younger and older participants across all three diagnostic groups. Mann Whitney U test results indicated that younger participants were significantly more fidgety than older participants ($U = 256, p < .05$), more frequently avoided eye contact ($U = 183, p < .05$), and were less engaged in the social interaction ($U = 183.5, p < .05$). There were no significant age-group differences for the remaining behavioral markers.

Primary Analyses

Parent and Child Ratings

Standardized behavioral ratings scales were used to assess participants' levels of anxiety as measured by parental or self-report ratings. The ratings were examined with Kruskal-Wallis tests. The only significant effect of group status found was for the CBCL social difficulties subscale ($H = 20.76, p < 0.005$). Post hoc Mann Whitney U test results showed that parents of

Table III. Correlations Between the Behavioral Markers and Age or VIQ (with Total Sample)

Measures	Age	VIQ
Eye contact avoidance ($n = 54$)	-.45 ^a	-.04
Lip biting ($n = 58$)	.097	-.19
Wringing hands ($n = 55$)	.21	-.06
Fidgetiness ($n = 55$)	-.46 ^a	-.08
Rigidity ($n = 55$)	-.11	.01
Number of pauses ($n = 58$)	-.13	-.16
RT to first utterance ($n = 57$)	-.08	-.11
Duration of silence ($n = 57$)	-.60 ^a	-.19

^a $p < .005$.

participants with Turner or fragile X syndrome perceived their children as having more social problems than parents of participants in the comparison group ($U = 159, p < .05$; $U = 173, p < .05$), respectively. Relative to the comparison sample, females with Turner or fragile X syndrome did not report experiencing more anxiety symptoms on the RCMAS, the anxiety subscale of the CBCL, or the SPAI-C ($ps > .35$). Although the mean SPAI-C score for females with Turner or fragile X syndrome did not differ from the mean score of the comparison group, their within group variability was greater than that seen in the comparison group (see Table IV).

Behavioral Markers

Ratings for three of the eight markers differed significantly among the three participant groups. Specifically, frequency of lip-biting differentiated the groups ($H = 13.58, p < 0.005$), with females in the Turner syndrome group showing less facial movement than females in the fragile X group ($U = 81, p < .05$), or comparison group ($U = 159, p < .05$). Mann Whitney U tests revealed that participants with fragile X syndrome took more time than those with Turner syndrome to initiate the role-play interactions, $U = 88.5, p < .05$, and made more hand movements, than the other groups; $U = 84, p < .05$; and $U = 94.5, p < .05$, for the Turner syndrome and comparison groups, respectively (see Table 4).

Correlations Between Behavioral Markers and Standardized Anxiety Scales

To investigate how behavioral markers were related to one another and to standardized anxiety measures, a series of Spearman rank correlations was conducted. Because of the large number of variables and relatively small sample sizes of Turner or fragile X syndrome groups, correlations between variables were examined for the total sample. The three behavioral markers that differentiated the groups (i.e., time to first utterance, wringing hands, and lip biting) were significantly correlated to each other, but not to the remaining behavioral markers (Table V). Only duration of silence was consistently correlated with most of the remaining behavioral markers.

To examine whether similar associations were evident in each of the three participant groups, correlations were carried out separately for each group using only the behavioral markers that were significantly correlated among the total group. The patterns of associations differed across the three groups. In the comparison group, duration of silence did not correlate with any behavioral markers. In the Turner and fragile X syndrome groups there were few significant associations between duration of silence and other markers (Table VI). No significant correlations emerged between any behavioral markers of anxiety and females' or parental ratings of anxiety on standardized anxiety measures (Table VII).

Table IV. Means and Standard Deviation Scores for Study Groups

Variable	Turner syndrome		Fragile X		Comparison group		
	M	SD	M	SD	M	SD	<i>p</i>
Behavioral markers							
Duration of silence	33.81	33.14	37.25	27.89	35.85	26.58	NS
Number of pauses	3.14	1.77	3.00	2.00	2.90	1.73	NS
RT to first utterance	7.71	31.52	9.94	26.72	2.15	5.67	.018 ^{a,c}
Wringing hands	.79	2.20	2.69	2.87	1.00	2.32	.010 ^{a,c}
Lip Biting	.43	.96	2.63	2.68	2.14	1.65	.001 ^{a,b}
Rigidity	1.16	2.48	1.69	2.68	1.05	1.99	NS
Fidgetiness	1.95	2.76	1.00	2.22	1.45	2.24	NS
Eye contact avoidance	5.22	1.93	5.69	2.06	5.45	2.44	NS
Standardized Checklists							
RCMAS	45.41	13.54	44.11	10.33	42.38	14.02	NS
CBCL Anxiety	55.18	7.08	54.71	7.67	52.77	5.22	NS
CBCL Withdrawal	53.82	5.84	52.82	4.43	51.35	3.65	NS
CBCL Social	61.61	8.77	57.11	9.85	52.26	5.53	.00 ^{b,c}
SPAI-C	13.76	9.91	15.01	11.33	10.08	10.25	NS

^a Significant differences between females with fragile X and Turner syndrome.

^b Significant differences between females with Turner syndrome and comparison group.

^c Significant differences between females with fragile X and comparison group.

NS = Not significant.

Table V. Intercorrelations Among the Behavioral Markers for the Total Sample ($n = 58$)

Variables	1	2	3	4	5	6	7	8
1. Eye contact avoidance	–							
2. Lip Biting	.18	–						
3. Wringing Hand	–.12	.16	–					
4. Fidgetiness	.40 ^a	.01	–.14	–				
5. Rigidity	–.01	–.03	–.10	–.28 ^b	–			
6. Number of pauses	.37 ^b	.02	–.09	.12	.06	–		
7. RT to first utterance	.24	.33 ^b	.28 ^b	.18	.17	.02	–	
8. Duration of silence	.46 ^a	.28 ^b	–.04	.40 ^a	.33 ^b	.28 ^b	.30 ^b	–

^a $p < .005$.^b $p < .05$.

DISCUSSION

The results from the present study are partially consistent with earlier evidence that parents perceive females with Turner or fragile X syndrome as having social difficulties. Among the various standardized behavioral scales examined, the only standardized rating measure that differentiated groups with a genetic syndrome from the comparison group was the CBCL Social Problems scale. The mean rating for the Turner and fragile X groups were not in the clinically significant range; but 16 (55%) of the females with Turner syndrome, and 5 (29%) of the females with fragile X syndrome had an above-average T score ($T \geq 60$) on this rating, versus 3 (10%) of the females in the comparison group. Contrary to the prediction, neither the parents of females with Turner or fragile X syndrome nor the participants themselves rated the females as more anxious, relative to comparison females, on standardized anxiety scores from the CBCL anxiety scale, RCMAS, and SPAI-C. The findings on behavioral markers were consistent with the social problems rating in that they provided partial evidence of specific social discomfort in the fragile X group.

Table VI. Correlations Between the Behavioral Markers with "Duration of Silence" in the Three Participant Groups

Measures	Turner syndrome	Fragile X	Comparison group
Eye contact syndrome	.68 ^b	.49	.26
Lip biting	–.01	.54 ^a	.13
Wringing hand	–.07	.19	–.38
Fidgetiness	.53 ^a	.48	.31
Rigidity	.28	.25	.39
Number of Pauses	.38	.11	.32
RT to first utterance	.39	.43	.14

^a $p < .005$.^b $p < .05$.

Parent Ratings and Self-Report Ratings of Child Behavior

The lack of significant findings on *parental* measures of anxiety is not surprising in view of similar findings of studies of females with Turner syndrome (e.g., Mazzocco *et al.*, 1998; Ross *et al.*, 1996; Rovet & Ireland, 1994) or fragile X (e.g., Kovar 1993; Mazzocco *et al.*, 1998). Yet the findings are inconsistent with other reports of parental endorsement of anxiety symptoms in females with Turner syndrome (McCauley *et al.*, 2001; Skuse, Percy & Stevenson, 1994) or fragile X syndrome (Lachiewicz & Dawson, 1994), relative to parents of comparison participants. Like their parents, the females with Turner or fragile X syndrome in the present study did not report greater levels of general or social anxiety than did the females from the comparison group; which is consistent with earlier null findings with females with Turner or fragile X syndrome as based on self reports (Mazzocco *et al.*, 1998; McCauley *et al.*, 2001; Ross *et al.*, 1996).

Although specific reasons for the conflicting results in the past and present investigations remain undetermined, there are several possible explanations. It is possible that, despite their medical condition, females with Turner or fragile X syndrome cope positively with their condition-related stresses. Access to social support may buffer females with Turner or fragile X syndrome from emotional dysfunction. In the present study, access to social network and coping strategies were not measured or available for analyses. Perhaps our sample was biased toward greater social support in that study participants' parents either felt that participation would be beneficial for their child or simply had the financial and social resources that allowed them to participate. An alternative explanation concerns the limitations of the measures used in our study, and the lack of sensitivity inherent in screening measures like the CBCL

Table VII. Correlations Between the Behavioral Markers and Standardized Behavioral Measures

Variables	RCMAS	CBCL Anx	CBCL Withdraw	CBCL Soc	SPAI-C
Eye contact avoidance	-.12	-.21	-.17	-.22	.14
Lip biting	.11	.01	-.00	-.21	.16
Wringing hand	.06	.17	.23	.04	-.04
Fidgetiness	.10	.03	.13	.15	.16
Rigidity	.01	-.01	-.11	.00	.09
Number of pauses	.05	-.13	-.02	.05	.27
RT to first utterance	-.11	-.10	-.01	-.13	.26
Duration of silence	.01	-.07	-.04	.01	.33

CBCL Anx = CBCL Anxiety Subscale; CBCL Withdraw = CBCL Withdrawal Subscale; CBCL Soc = CBCL Social Subscale.

anxiety scale and the RCMAS for detecting subtle group differences. This explanation is consistent with evidence that general anxiety measures lack specificity in detecting symptoms of social phobia (Herbert, Bellack, & Hope, 1991). It remains unclear how social interaction anxiety symptoms correspond to symptoms reflected by behavioral markers used in the present study. It is also possible that real group differences detectable by the behavioral markers we used were undetected because of our small sample size; or that these real differences exist only among older (or younger) females and were undetected in our sample with a wide age range. Finally, behaviors we interpreted as indicators of social anxiety may lack specificity in differentiating social anxiety from similar but qualitatively different diagnostic categories. For instance, characteristic behaviors of Avoidant Personality Disorder overlap with those identified in social anxiety, making it difficult to differentiate these two disorders. This explanation is particularly relevant for fragile X population, where high rates of Avoidant Personality Disorder have been reported (Freund, Reiss, & Abrams, 1993). Nevertheless, our findings indicate a lack of significant behavioral concerns in females with Turner or fragile X syndrome, based on standardized instruments, with the exception of higher, borderline ratings for social problems.

Specificity of a Behavioral Phenotype for Fragile X or Turner Syndrome

Among the primary findings of our study were group differences on behavioral markers measured during a brief simulated social situation. The participants with fragile X made more gestures during their role-play interactions and took more time to initiate conversation than did either of the two remaining groups. The

only group difference between the Turner and comparison group was of fewer facial movements in the Turner group. Underlying factors that contribute to the slightly different expression of discomfort in social situations in Turner or fragile X syndrome remain to be determined; and although the behavioral markers among the fragile X group are consistent with notions of discomfort during role play, the behaviors exhibited by the Turner group may not indicate discomfort but instead may reflect heightened degree of overt self-control. Although a need for self-control, behaviorally expressed as less facial movement, may reflect underlying anxiety, alternatively it may be associated with the reported inertia in females with Turner syndrome when they reach puberty that contrasts with the hyperactivity and impulsivity observed in younger prepubertal children with Turner syndrome (McCauley *et al.*, 1986). However, attention difficulties are also seen in a subset of adolescents with Turner syndrome (McCauley *et al.*, 2001). Thus it is more difficult to interpret these limited behavioral markers for the Turner group, relative to those observed in the fragile X group, but both sets of findings remain preliminary.

When considering the within-group patterns of our findings, it is interesting to note the significant findings for only some behaviors (i.e., increased fidgeting/movement of upper extremities, facial movement, and time to initiate the social interactions), but not for others (e.g., body orientation, gaze aversion, or number of pauses during the interaction). These differential patterns are somewhat surprising because all behavioral indices used in the study, including those for which no group differences emerged, were previously found to be related to level of social anxiety in adults (see Monti *et al.*, 1984 for a review). Inconsistent findings across studies may reflect how behavioral manifestations of anxiety in children differ from those

seen in adults (Perry, 1998). In the present study, although age was significantly correlated with selected behavioral markers (i.e., duration of silence, eye avoidance, and fidgeting), it was not associated with the behavioral anchors that differentiated the fragile X group from the two remaining groups. Therefore, it is unlikely that age served as a confounding variable that accounted for group differences in behavioral markers. The three behavioral markers that the younger females exhibited more frequently than the older females may reflect developmental differences in age-appropriate behaviors rather than symptoms of social anxiety. For instance, social withdrawal or avoidance of social interactions is less damaging and stigmatizing in early childhood than during adolescent years, during which the social interactions are crucial for development of self-esteem and peer acceptance (Pope, McHale, & Craighead, 1988).

Another possible explanation for these mixed results is that our frequency count behavioral assessment method might not be sensitive for detecting subtle deficits in social skills or social anxiety. A qualitative approach may have allowed detection of higher levels of anxiety in females with Turner syndrome or fragile X. Belser and Sudhalter (1995) used a qualitative method to assess anxiety-related behaviors, and they identified significantly more anxious behaviors in males with fragile X syndrome compared to a sample of males with Down syndrome. Further, in the present investigation we almost exclusively scrutinized nonverbal aspects of social skills. In contrast, Belser and Sudhalter successfully identified less competent social skills by examining both nonverbal skills and specific aspects of language and speech (e.g., topic maintenance, deviant repetitive language, etc.) during social interactions. It is noteworthy that verbal indices similar to those applied by Belser and Sudhalter differentiated socially anxious people from comparison individuals (see Monti *et al.*, 1984 for a review).

Finally, the pattern of our findings may be related to the brevity of the role play. However, earlier studies also involving short role plays (180 seconds) have been sensitive to differentiating socially anxious individuals from those without pathology in social functioning. For example, an examination of reticence in young children, which appears associated with enduring characteristics and social anxiety, suggested that anxious behaviors displayed during the initial phase of interaction remained relatively stable over the course of the entire social situation (Asendorf, 1991). Thus, capturing signs of social anxiety, if these signs reflect enduring or trait characteristics, might not be so dependent on the length of

time of an interaction as on adequately operationalizing the quality of the interactions seen among children with social anxiety.

Regardless of these possibilities and our study's limitations, our findings do indicate group differences between females with fragile X or Turner syndrome versus females in a comparison group, and between females with fragile X syndrome and females with Turner syndrome. These findings support the notion of differential behavioral phenotypes in these two disorders.

Correlations Between Behavioral Markers and Standardized Anxiety Ratings

One aim of this study was to assess the associations between the behavioral markers and standardized anxiety measures. Contrary to predictions, these associations were rarely significant. The exception was the correlation between duration of silence with six of the seven remaining behavioral variables, suggesting non-specificity of this behavioral index with social anxiety or social dysfunction. The same trend was seen in the Turner syndrome group when separate within-group analyses were conducted. The behavioral markers that differentiated the fragile X or Turner groups' behavioral presentations (i.e., time to first utterance, biting lips, wringing hands) were related to each other, but none was correlated with other behavioral markers nor with participants' or parental ratings of anxiety as measured by standardized instruments. Moreover, neither group showed significant differences on all three of these behavioral markers, and none of the three markers differed significantly for both Turner and fragile X groups.

Absence of significant correlations between the remaining behavioral markers, and the lack of associations between behavioral markers and standardized measures of anxiety may address whether such behavioral markers are pertinent in measuring social anxiety, or whether the standardized general anxiety measures are pertinent for measuring the social anxiety that may characterize the fragile X group. Alternatively, behavioral inadequacy in social situations exhibited by females with either disorder might be attributed to underlying language dysfunction or impaired discourse processing (Spinelli, de Oliveira Rocha, Giacheti, & Richieri-Costa, 1995; Sudhalter, Cohen, Silverman, & Wolf-Schein, 1990). The degree to which those explanations are supported may differ across Turner or fragile X syndrome. Emotional components may have an impact on these social skills and may interact with neuropsychological functioning.

Limitations of the Study and Directions for Future Research

The present study provides preliminary evidence of behavioral markers of social discomfort, particularly in females with fragile X syndrome; and it supports earlier reports of social difficulties in both Turner and fragile X syndromes, based on parental report. Future research is needed to establish the presence of social anxiety in females with either syndrome, and to clarify the nature of social skills or discomfort in females with either of these disorders. The role-play method used in this study involved a very limited social situation that, while informative, is limited in its generalizability. Future studies could be designed to examine more extended social interactions, or similarly brief interactions used to measure potential influences on level of social discomfort. For instance, the potential role of stranger characteristics can be assessed through role plays with same-age or same-sex peers, with and without presence of a passive observer, and in vivo interactions such as those used with younger children (Coplan *et al.*, 1994) may allow assessment of consistency or emergence of social discomfort over the course of an extended interaction. The present study examined mostly nonverbal aspects of social interaction. For a more comprehensive evaluation, operationalizing and assessing verbal indices of social anxiety along with nonverbal markers of anxiety during role play interactions should be considered in future studies.

Another important direction for future research concerns developmental effects, which are suggested by the age differences that emerged in the present study. For example, with a larger sample size, it would be possible to examine potential group or developmental differences in participants' warming up to unfamiliar adults across scenarios, and the possible interaction between developmental level and group (fragile X, Turner syndrome). When selecting the variables that assess social skills, care must be taken to identify those indices that adequately reflect socially anxious behaviors at different developmental ages.

Cognitive influences may also affect levels of social discomfort in Turner or fragile X syndrome, in different ways and to different degrees between these two groups. Accurate identification of the nature of difficulties (e.g., social phobia vs. social dysfunction vs. neurocognitive deficits) is crucial for planning the most effective intervention for these individuals. If social difficulty is related to anxiety, treatment based on anxiety reduction techniques will be the most appropriate strategy in coping

with stress. In turn, if social dysfunction or cognitive deficits (e.g., language deficits or executive function dysregulation) can account for social difficulties, then improving the impoverished repertoires of social behaviors or applying cognitive remediation would be appropriate management strategies, respectively. The limited research on cognitive components of social skills fails to support the role of cognitive ability on social skills functioning or socioemotional ratings (e.g., Mazzocco, Pennington, & Hagerman, 1994; Sobesky, Pennington, Porter, Hull, & Hagerman, 1994, respectively); although ratings on a clinical "lie scale" measure appear negatively correlated with cognitive ability among women with fragile X syndrome (Sobesky *et al.*, 1994). This latter finding addresses the limitations in accuracy of self-report measures, which in turn addresses the value of continued use of objective behavioral markers of anxiety in future research.

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Please note that the role plays used in this study were originally designed for the purpose of comparing subjective self-reports of state anxiety with parent informant-reports of a child's anxiety. The parental measures were based on their viewing the video taped role plays described in this report. The original analysis was not carried out, because the participants' self-reports were later found to be very unreliable and thus considered as invalid. The study presented in this report is based on a more objective methodology developed post hoc, by the first two authors, to address the research questions on which this paper is based. However, because the role plays were not carried out with our methodology in mind, the quality of the tapes limited our ability to code all desired points for all participants (as shown in Table II).

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