Behavioral style of young boys with fragile X syndrome

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To study the behavioral style or temperament of 45 boys, aged 47 to 88 months, with full-mutation fragile X syndrome (FXS), 102 parent ratings on the Behavioral Style Questionnaire (McDevitt and Carey 1978) were recorded. These ratings were analysed with a variety of statistical techniques. Considerable variability was evident in temperament profiles; consequently, a characteristic profile was not identified for FXS. Boys with FXS differed significantly from the reference sample on five of nine temperament dimensions. They were more active and less intense, approachable, adaptable, and persistent. No significant differences were found in distractibility, rhythmicity, mood, or sensory threshold. Only 16 of the 45 boys in the sample could be classified as easy, difficult, or slow to warm up. There was no link between severity of developmental disability and temperament ratings. This supports the theory that intelligence and temperament are separate constructs. Scores on temperament dimensions were stable over time. Our results suggest that many of the behaviors observed in boys with FXS may be related to temperament. Consequently, parent counseling and environmental modifications should be considered as first line treatment. The question of whether the behavior problems observed in boys with FXS are innate or whether they result from poorness of fit between child and environment is an important issue that needs further study.

Fragile X syndrome (FXS), first identified in 1969 and characterized molecularly in 1991 (Bell et al. 1991, Verkerk et al. 1991, Vincent et al. 1991, Yu et al. 1991), is the most common hereditary cause of mental retardation*. The cognitive impairment and behavioral characteristics of FXS result from the lack of FMR1 protein, which is believed to be necessary for normal brain development (Tassone et al. 1999). Phenotypic expression and development are highly variable in FXS. Common physical features in younger boys include large or protruding ears, hypotonia, flat feet, and hyperextensible joints. Cognitively, although some males function in the borderline to mild range, most have moderate to severe disabilities, even at young ages.

Although cognitive and language deficits in FXS are readily apparent and serve to compromise many areas of function, it is the behavioral features that interfere with successful adaptation. In addition to delayed development, especially delayed speech and language skills, boys with FXS often have short attention span, high activity level, impulsivity, lack of adaptability (similar to children diagnosed with attention-deficit–hyperactivity disorder) or autistic-like behaviors (e.g. hand flapping, gaze avoidance, tactile defensiveness). As many as 25% of males with FXS may meet the criteria for a diagnosis of autism (Turk and Graham 1997, Bailey et al. 1998). However, due to variability in presentation a definitive diagnosis is often not made until the age of 3 years. Diagnosis is based on DNA testing.

The current study examines how the behaviors frequently observed in FXS fit with the construct of temperament or behavioral style. Temperament is typically described as how behavior is expressed without regard to motivation and abilities (Carey 1986). Increasingly it is recognized that both genetic and environmental characteristics affect developmental outcome. A fundamental assumption underlying the construct of temperament is that variation in temperament style accounts for variation in behavior. This may require environmental modification to assure a 'goodness-of-fit' between the environment and an individual's behavioral style (Thomas and Chess 1977, 1989). However, most research on temperament has been couched in the psychological literature and is descriptive in nature (Keogh 1982, Bates 1987, Goldsmith et al. 1987, Strelau and Angleitner 1991, Prior 1992, Bates and Wachs 1994, Rothbart and Bates 1998). Comparatively little research examines how temperament style could help in the development of intervention for a particular individual, as suggested by the goodness-of-fit model developed by Thomas and Chess (1977, 1989) and adapted for clinical pediatric use by Carey and colleagues (Carey et al. 1985a, 1985b, 1998; Carey and McDevitt 1995; Carey and Jablow 1997).

Temperament can be conceptualized along a number of dimensions. The nine dimensions included in the Behavioral Style Questionnaire (BSQ) (McDevitt and Carey 1978) described in Table I are the most widely used and are based on the landmark New York Longitudinal Study (NYLS) (Thomas et al. 1963). Temperament has also been characterized more globally. For example, Thomas and Chess (1963) conducted a factor analysis of these nine temperament domains and identified three broad categories: easy, slow to warm up, and difficult. The child with an easy

*UK usage – learning disabilities.

temperament maintains regular feeding, sleeping, and elimination routines (high rhythmicity); is more likely to approach a novel situation with ease; and is quite adaptable when meeting the demands of the environment (high approach and adaptability). Overall, this child demonstrates a positive mood and expresses the intensity of his/her emotions in a mild manner (Thomas and Chess 1977, 1989; Carey and McDevitt 1995). The child with a slow-to-warm-up temperament is typically shy and withdrawn (Carey and McDevitt 1995), exhibits low levels of activity, adapts slowly to new environments or situations, and has a negative mood. This child also expresses the intensity of his/her emotions in a mild manner (Thomas and Chess 1977, 1989; Carey and McDevitt 1995). The difficult child maintains an irregular feeding, sleeping, and elimination routine (low rhythmicity), and is more likely to withdraw when presented with a novel situation. The difficult child adapts slowly to changes in the environment and displays an intense, negative emotional response and mood (Carey and McDevitt 1995).

In the NYLS approximately 40% of the children had an easy temperament, 15% were slow to warm up, and 10% were difficult (Carey and McDevitt 1995). The remaining 35% exhibited a combination of these qualities and did not fit into any specific category; consequently, this group was named intermediate. When Carey and colleagues (Carey and McDevitt 1978, McDevitt and Carey 1978, Hegvik et al. 1982, Fullard et al. 1984) developed their temperament ratings, they expanded the definition of intermediate into two categories: intermediate high or intermediate low (Carey and McDevitt 1995). The proportion of children in their easy (36%) and difficult (12%) categories was similar to that of the NYLS. However, the Carey sample contained fewer children who were slow to warm up (6%), and more who were rated as intermediate (46%). Of the children rated as intermediate, 32% were considered intermediate low and 14% were considered intermediate high.

Children in the intermediate high group resembled difficult children by scoring high in four to five dimensions typically related to the difficult cluster, although they scored greater than 1 SD above the mean in only one dimension (Carey 1970). Children also received a cluster rating of intermediate high when they exhibited scores greater than 1 SD above the mean in only two to three of the dimensions, rather than four or five, comprising a difficult temperament (Carey 1970).

Alternatively, children in the intermediate low group approximated those in the easy category by having only three to five dimensions represented in this cluster, with no scores 1 SD above the mean (Carey 1970). A child would also be considered intermediate low rather than easy with one to three scores on the five dimensions characterizing an easy temperament (rhythmic, approachable, adaptable, mild intensity, and positive mood), if all of these scores were greater than 1 SD above the mean (Carey 1970).

Although several studies have described temperament of children with disabilities, most have focused on Down syndrome (Baron 1972; Bridges and Cicchetti 1982; Brooks-Gunn and Lewis 1982; Rothbart and Hanson 1983; Gunn and Berry 1985a, 1985b; Huntington and Simeonsson 1987; Cunningham 1996). A brief description of these studies is provided in Table II. Collectively they suggest that: the temperament of children with disabilities is not that different from children without disabilities; temperament does not appear to be generally related to severity of disability; and temperament ratings are relatively stable over time. Extensive reviews of this literature (Huntington and Simeonsson 1993, Carey and Jablow 1997) note that the temperament of children with disabilities is consistently similar to that of typically developing children. However, considerable variability within disability groups has been observed, precluding attempts to describe a typical temperament profile for children with a specific disability. Additional studies are needed to expand this literature to other disability groups to test the generalizability of the findings mentioned above (comparability with typical children, no relation to severity of disability, and stability over time).

Children with FXS form a natural group to test these assumptions, because their behavioral style appears to be so different from that of children with Down syndrome. Although several studies in the literature on FXS have described problem behavior, temperament has not been examined. This study describes maternal ratings of the temperament of a sample of boys with FXS, aged 47 to 88 months. We collected 102 ratings of 45 boys enrolled in a prospective longitudinal study. Developmental, behavioral, and educational characteristics of young males with FXS were obtained. Using a variety of statistical techniques, our aims were to describe the temperament profile of young boys with FXS, determine how their temperament differs from that of children without disabilities, examine the relation between temperament and severity of developmental disability; and ascertain the extent to which their temperament is stable over time.

Table I: Temperament dimensions from the New York Longitudinal Study (1963)

Description	*Range*
Activity: physical movement demonstrated during daily pastimes (i.e. playing or bathing)	High (6) to Low (1)
Rhythmicity: consistency over physiological functions (i.e. sleep, hunger, and elimination)	Arrhythmic (6) to Rhythmic (1)
Approach: a tendency to advance towards a new object or depart quickly from its presence	Withdrawing (6) to Approachable (1)
Adaptability: ability to adjust or change behavior in socially desirable ways	Slow to Adapt (6) to Very Adaptable (1)
Intensity: the depth or magnitude of an emotional response expressed by an individual	Very Intense (6) to Mild Intensity (1)
Mood: the quality of an emotional reaction in either a positive or negative direction	Negative (6) to Positive (1)
Persistence/Attention Span: the extent to which a challenging task is pursued by a child	Low (6) to High (1)
Distractibility: ability to maintain attention during ongoing event disrupted by environmental stimuli	High (6) to Low (1)
Sensory Threshold: the minimal amount of sensory stimulation necessary to elicit a response	Low (6) to High (1)

Table II: Studies of temperament in children with disabilities

Authors	*Sample*	*Age*	*N*	*Instrumentation*	*Findings*
Baron (1972)	Down syndrome (DS)	6–18 mo	18	Infant Temperament Questionnaire (ITQ)	No significant differences in temperament between samples with DS and those with typical development
Carey et al. (1979)	Minimal brain dysfunction (attention-deficit–hyperactivity disorder)	36–84 mo	61	Behavioral Style Questionnaire (BSQ)	Children with ADHD were less adaptable and persistent than those with typical development. ADHD sample had higher activity levels and more negative mood. Behavioral characteristics of children with ADHD may overlap with those having a 'difficult' temperament (low adaptability and a negative mood)
Brooks-Gunn and Lewis (1982)	Down syndrome (DS), motor impairments (MI), developmental delay (DD)with no organic etiology	3–36 mo	82	Revised Infant Temperament Questionnaire (RITQ), Toddler Temperament Scale (TTS)	Differences were found at 8 mo of age. Children with DS had lower sensory threshold and activity levels than those with DD. Those with DS were rated 'easy' more often than those with MI or DD
Heffernan et al. (1982)	Neurological impairments (NI)	5–39 mo	57	Revised Infant Temperament Questionnaire (RITQ), Toddler Temperament Scale (TTS)	Similar distributions of children with NI and typical development in easy, difficult, and slow-to-warm-up categories. Children with NI had low activity, low persistence, short attention spans, high sensory thresholds, and were more withdrawn
Bridges and Cicchetti (1982)	Down syndrome (DS)	4–24 mo	74	Infant Temperament Questionnaire (ITQ)	No significant differences between children with DS and those with typical development on six of the nine domains. Children with DS has a lower sensory threshold, were less persistent and approachable. Infants with DS had delayed motor development, fewer vocalizations, and were less approachable
Gunn and Berry (1985a)	Down syndrome (DS)	Mean age, 30 mo	37	Toddler Temperament Scale (TTS)	Children with DS were more rhythmic, had a more positive mood, and were less intense than either chronological age or mental age controls. When compared to mental age controls, children with DS were more approachable and adaptable with a lower sensory threshold. When compared to chronological age controls, children with DS were less persistent and had a higher sensory threshold. Overall, 98% of the DS sample were classified as easy (68%) or intermediate low (30%)
Gunn and Berry (1985b)	Down syndrome (DS)	3–6 y	14 boys 9 girls	Behavioral Style Questionnaire (BSQ)	No significant differences in temperament ratings were found between children with DS and those with typical development
Huntington and Simeonsson (1987)	Down syndrome (DS)	3–75 mo	34[a] 29[b]	Toddler Temperament Scale (TTS)	Children with DS had temperament traits similar to those without disabilities. All children with DS did not have an easy temperament
Prior et al. (1988)	Hearing impaired (HI)	32–64 mo	26	Childhood Temperament Questionnaire (CTQ)	Children with HI were rated as more difficult than those without HI. They were also more active, less rhythmic, and less distractible
DiLavore (1991)	Autism and Down syndrome (DS)	38–95 mo	58	Behavioral Style Questionnaire (BSQ)	Children with autism were more negative, less active, less rhythmic, less approachable, less intense, less persistent, and had a higher sensory threshold than those with DS. Children with autism were more likely to be rated intermediate high/difficult than intermediate low/easy
Sanson et al. (1993)	ADHD, aggression	4–96 mo	182	Revised Infant Temperament Questionnaire (RITQ), Childhood Temperament Questionnaire (CTQ) Forms A and B	Children who were more irritable, less cooperative, and unable to adapt developed externalizing disorders more often than those who were cooperative and manageable
Keogh et al. (1997)	Developmentally delayed (DD)	36–132 mo	103	UCLA temperament scale	Children with DD rated as 'easy' tended to lose IQ points more rapidly than those with a 'difficult' temperament'

[a] 1 year olds, [b] 2 year olds.
UCLA, University of California at LA.

Method

PARTICIPANTS

The participants were 45 males with FXS in four states – North Carolina, South Carolina, Virginia, and Georgia, in the southern United States of America. The average age of enrolment in the current study was 64 months, with a range of 47 to 88 months. Developmental quotients ranged from 26 to 77, with an average of 49. Recruitment into the study, which is ongoing, began in 1994. All of the participants were diagnosed with full-mutation FXS using DNA analyses.

Subjects were recruited through genetics clinics, developmental evaluation centers, and early intervention programs. Informed consent for participation was obtained from the parents or guardians of all participants. Demographic characteristics of the children are described in Table III.

INSTRUMENTATION

Temperament assessment

The BSQ (McDevitt and Carey 1978) was used to measure behavioral style (Carey and McDevitt 1995). The scale has a high level of internal consistency (0.84) and test–retest reliability (0.89) (McDevitt and Carey 1978). Parents are asked to respond to 100 items (e.g. the child is slow to adjust to changes in household rules) on a 6-point rating scale ranging from 1 (the child almost never demonstrates a particular behavior), to 6 (the child almost always exhibits that behavior). Item ratings generate scores for the nine temperament dimensions identified by Thomas and colleagues (1963).

Developmental assessment

The Battelle Developmental Inventory (BDI) (Newborg et al. 1984) was used to measure severity of developmental disability. As the BDI covers the age range from birth to 96 months, it provides a consistent measure of development during infancy, preschool, and early elementary school years. The BDI yields an overall developmental age or quotient in addition to scores for the subdomains of personal social, adaptive, motor, communication, and cognitive development. It has a solid normative base drawn on a nationally representative sample of children with well documented reliability and validity. Additionally, it has adaptations for children with disabilities, making it particularly well suited for longitudinal studies of development with those samples (e.g. Hatton et al. 1997; Bailey et al. 1998a, b). Several independent studies have documented a high correlation between the BDI and measures of cognitive, adaptive, language, and social functioning with populations of both normally developing children and children with disabilities (McClean et al. 1987, Sexton et al. 1988, Boyd 1989, Snyder et al. 1993).

PROCEDURES

Upon entry into the study, the parents of each child were asked to complete the BSQ. At each child's first assessment, the temperament forms were discussed with the parents, and they were asked to complete the forms and return them. Subsequently, at each child's birthday, temperament forms were again mailed to both mothers and fathers. If temperament forms were not returned within 4 weeks, a follow-up letter and/or phone call was made. The data reported represent a total of 102 assessment occasions, with an average of 2.3 assessments and a range of 1 to 4 ratings per child. For children with multiple assessments, length of time between assessments ranged from 6 to 18 months. Only maternal ratings were used for this study.

The BDI was administered every 6 months by project staff with extensive training and experience in administering the measure and in working with young children with disabilities. Assessments were conducted at times and locations selected by parents as most likely to yield a representative sample of the child's competence. Most assessments were conducted in the child's home or school.

Results

A variety of statistical techniques were used to analyse the data. An alpha level of 0.01 was used as the criterion for statistical significance owing to the number of variables and the use of multiple tests.

TEMPERAMENT DIMENSIONS

We first examined the nine dimensions of temperament: activity level, rhythmicity, approach, adaptability, intensity, mood, persistence/attention span, distractibility, and sensory

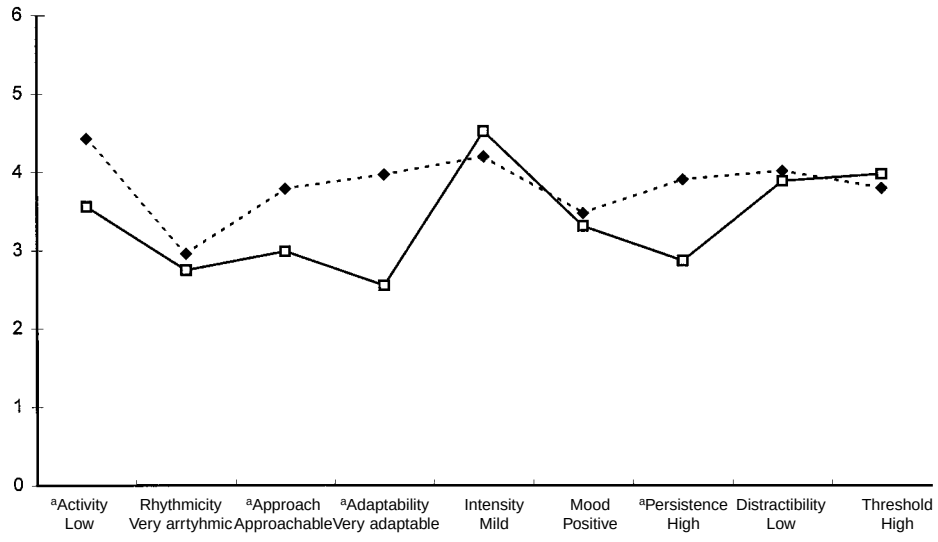

Figure 1: *Behavioral Style Questionnaire dimensions: - - -♦- - -, fragile X and —❑—, Carey Samples. [a]Significant differences in two samples at P<0.01.*

threshold. To compare the means and SDs for the sample of boys with FXS to those of the typically developing children in the Carey sample (McDevitt and Carey 1978), *t* tests with an adjusted alpha ($P<0.01$) for multiple tests were used. Significant differences were found in five of the nine dimensions. Boys with FXS were more active ($P<0.001$) and less approachable ($P<0.001$), adaptable ($P<0.001$), intense ($P<0.001$), and persistent ($P<0.001$). The means and SDs for each sample are displayed by dimension in Table IV, along with the computed values of *t*, and degrees of freedom. A graphic representation of the temperament profiles of the two samples is shown in Figure 1.

CLUSTER CATEGORIES

Only 16 of the boys fit into the three main cluster categories – easy, difficult, or slow to warm up. The remainder fell into the intermediate category with 16 in the intermediate high cluster and 13 in the intermediate low cluster. When compared to Carey's sample of typically developing children, fewer children in the FXS sample were rated as easy ($P<0.001$). Although more children with FXS were rated as difficult than were typically developing children in the Carey sample, this difference was not statistically significant. However, significantly more children in the FXS sample were rated as intermediate high (approximating aspects of the difficult category) ($P<0.005$) than in the Carey sample. There were comparable proportions of children in the intermediate low cluster and in the slow-to-warm-up cluster. The distributions of children by cluster for both the FXS and the Carey samples are displayed in Table V.

STABILITY OF TEMPERAMENT

Hierarchical linear modeling analyses (Laird and Ware, 1982, Bryk and Raudenbush 1987, Willet 1989, Burchinal et al. 1994) were conducted on each of the dimension scores to assess stability of temperament. All dimensions were expected to be stable as evidenced by a slope of zero at the mean age of assessment – 66 months. The actual slopes for each dimension were not significantly different from zero, indicating stability of the nine dimensions. The results of these analyses are presented in Table VI.

TEMPERAMENT AND LEVEL OF DISABILITY

Hierarchical linear modeling was also used to assess the relation between temperament dimensions and level of disability. Using overall scores from the BDI, we computed developmental quotients (DQs) for each child by dividing developmental age by chronological age at assessment and multiplying by 100. The scores were rank ordered by first

Table III: Description of study participants ($N=45$)

Child characteristics	*Number*
Sex	
Male	45
Ethnicity	
European–American	39
African–American	5
Latino	1
Socioeconomic status	
Public assistance[a]	16
No public assistance	29
Age at assessment	
Mean age (mo)	63.83
Range (mo)	47–88
Ratings	
Mean number of ratings	2.27
Range of ratings	1–4
Total number of ratings	102
Developmental quotients (DQ)	
Mean DQ	49
Range of DQ	26–77

[a] The family was receiving assistance (not necessarily money) from government agencies.

Table IV: Temperament dimensions, Behavioral Style Questionnaire: comparison of fragile X syndrome (FXS) and Carey samples

Dimension	*FXS mean*	*FXS SD*	*Carey mean*	*Carey SD*	*t*	*df*	*P*
Activity level	4.43	0.59	3.56	0.75	7.513	44	0.0001
Rhythmicity	2.96	0.71	2.75	0.68	1.948	44	0.1000
Approach	3.79	0.94	2.99	0.94	5.391	44	0.0001
Adaptability	3.97	0.81	2.55	0.72	12.316	44	0.0001
Intensity	4.20	0.76	4.52	0.65	3.056	44	0.0010
Mood	3.48	0.59	3.31	0.68	1.607	44	0.1000
Persistence	3.91	0.72	2.87	0.69	9.506	44	0.0001
Distractibility	4.02	0.80	3.89	0.60	1.029	44	0.2200
Threshold	3.80	0.58	3.98	0.60	1.909	44	0.1000

Table V: Temperament clusters, Behavioral Style Questionnaire: fragile X syndrome and Carey samples

Cluster	*Fragile X %*	*Carey %*	*t*	*df*	*P*
Slow to warm-up	6.7	5.7	0.254	44	0.5000
Difficult	22.2	12.6	1.498	44	0.2000
Intermediate high	35.6	12.9	3.110	44	0.0050
Intermediate low	28.9	34.6	–0.773	44	0.8000
Easy	6.6	34.3	–6.169	44	0.0001

Table VI: Hierarchical linear modeling analysis of stability of temperament dimensions over time

Dimension score	*Mean*	*t*	*df*	*P*	*Slope*	*t*	*df*	*P*
Activity	4.44	48.67	44	0.0001	0.0028	0.71	30	0.4802
Rhythmicity	3.01	32.31	44	0.0001	–0.0088	–2.48	30	0.0189
Approach	3.78	27.85	44	0.0001	0.0041	0.76	30	0.4530
Adaptability	3.94	34.25	44	0.0001	–0.0029	–0.57	30	0.5729
Intensity	4.25	39.38	44	0.0001	–0.0013	–0.31	30	0.7598
Mood	3.51	44.26	44	0.0001	0.0007	0.18	30	0.8555
Persistence	3.96	41.83	44	0.0001	–0.0087	–1.76	30	0.0881
Distractibility	4.05	37.88	44	0.0001	–0.0018	–0.38	30	0.7081
Threshold	3.78	50.89	44	0.0001	0.0058	1.37	30	0.1818

BDI assessment, last BDI assessment, and the mean for all BDI assessments during the 47 to 88 month age span. Children were grouped into the following categories: DQ < 40; DQ = 40 to 55; DQ > 55. DQs were consistent across time; consequently, we used the mean overall DQ based on all BDI assessments administered during the interval of interest. Hierarchical linear modeling analysis was used to examine the multiple assessments of each temperament dimension as predicted by time, DQ category, and their interaction. There were no significant differences in either mean level or rate of change across time for each of the dimensions by level of disability. Again, the conservative alpha level was warranted by the number of dependent variables tested.

Discussion

The lack of control groups limits the current study. Future studies are needed in which children with a broad spectrum of disability types are compared with more recent samples of typically developing children and children with disabilities, matched on both chronological age and mental age. Nonetheless, this study constitutes the first published description of temperament in the context of FXS, documents potential differences between the temperament of children with FXS and typically developing children, verifies the stability of maternal ratings of temperament over time, and is consistent with prior literature which suggests that temperament and ability are relatively independent constructs.

We should note initially the considerable variability in temperament profiles of young boys with FXS, precluding the identification of a characteristic temperament profile for FXS. However, unlike previous studies of children with developmental disabilities the temperament of boys with FXS as a group appears to be different from that of typically developing children – in this case, the reference sample for the BSQ. Only 16 fell within the three major categories of easy, difficult, or slow to warm up, which suggests that these three BSQ categories do not adequately describe the temperament of boys with FXS.

Although some of the boys were rated as easy or difficult, the largest proportion was rated as intermediate high, and boys with FXS were less likely to be rated as easy. The proportions of boys in the slow-to-warm-up and intermediate low categories were not significantly different from those of the sample of typically developing children. However, the large proportion of boys with either intermediate high or difficult temperament in our sample far exceeds that of either the Carey sample or samples of children with disabilities, which suggests that boys with FXS present a more challenging style of behavior.

Boys with FXS differed significantly on five of the nine temperament dimensions when compared with the reference sample of the BSQ. They were more active and less approachable, adaptable, intense, and persistent. This finding supports clinical and research reports of the behavior of boys with FXS (Simko et al. 1989, Freund et al. 1995, Hagerman 1996, Merenstein et al. 1996). In addition, temperament rating and severity of disability were not significantly related, which supports the theory that temperament is separate and distinct from intelligence. Boys with lower developmental quotients were not more likely to be rated as difficult, and comparable proportions of boys had easy and difficult ratings in the group with the highest developmental quotient. The mean scores on each of the nine temperament dimensions did not change over the time period from 47 to 88 months, which supports the widely accepted belief that temperament is a relatively stable construct.

Studies of the developmental (Abbeduto and Hagerman 1997, Bailey et al. 1998a), cognitive (Dykens et al. 1993, Freund et al. 1995, Wright-Talamante et al. 1996, Powell et al. 1997), and behavioral (Baumgardner et al. 1995, Merenstein et al. 1996, Turk and Graham 1997) characteristics of boys with FXS have reported distinct individual differences within the FXS population. Our findings suggest that this is also true for temperament. This supports previous reports of individual differences in temperament characteristics for children with disabilities. As in other studies of children with disabilities, considerable variability in temperament profiles makes it difficult to describe the temperament of a 'typical' boy with FXS. Considering that temperament describes individual differences in behavioral style, it would probably be inappropriate to even attempt to do so. However, unlike other studies of children with disabilities, our sample did appear to be quite different from the typically developing children who comprise the reference sample for the BSQ, suggesting that the behavioral style of these children is unique. Given the frequent descriptions of problem behavior in the literature on FXS, it is surprising that more boys were not rated as difficult. Apparently, most boys with FXS do not display the negative mood required for classification in the difficult category. However, high activity, combined with low approachability, adaptability, and persistence, must present challenges for both parents and professionals. Achieving goodness of fit between children with those characteristics and their environment, particularly school environments, is not easy (Rothbart and Jones 1998). The recent use of temperament measures and counseling as a preventive/wellness approach by health maintenance agencies in the United States of America, however, suggests that those challenges can be overcome and that adaptations can be made to achieve goodness of fit (Carey and Jablow 1997, Carey 1998).

Our results suggest that individual assessment of temperament for intervention planning might be beneficial for boys with FXS. Many of their problems seem to be related to temperament characteristics of activity, persistence, approach/withdrawal, and adaptability rather than to cognitive disability alone. Sharing results of temperament questionnaires with parents and teachers in combination with specific recommendations for classroom modifications and intervention strategies could enhance the goodness of fit between individual children and their environments. This approach is consistent with Carey's recommendations for clinical use of temperament data. Given the emphasis on family involvement in early intervention, Carey's recommendations for dealing with behavior problems via parent counseling and environmental modifications seem particularly appropriate.

Intervention studies that examine the effectiveness of maximizing the goodness of fit between child and environment and the relations among temperament, physiological measures, and clinical conditions are needed. The current study suggests that high activity level, combined with low persistence, adaptability, and approachability contribute to the problem behaviors identified by parents and teachers. It also provides insight into why many boys with FXS start

medication trials at young ages. Studies of the relation between temperament, physiology, and behavior before and after the initiation of medication regimes would provide additional insight, as well as providing empirical validation of the use of medication with this population.

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