A Process Approach to Describing Mathematics Difficulties in Girls With Turner Syndrome

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ABSTRACT. ***Objective.*** **To expand on previous reports of mathematics difficulty in girls with Turner syndrome (TS).**

Methods. **Mathematics performance was examined by evaluating the types of errors made on mathematics achievement subtests by 29 girls with TS, 26 girls with fragile X syndrome (another genetic condition associated with mathematics difficulty), and 41 girls with neither disorder. Correlations between mathematics achievement scores and measures of IQ, attention, and visuospatial skills were also examined.**

Results. **Relatively low mathematics achievement was evident in girls with TS before 10 years of age, and a higher percentage of girls with TS made operation (57%) and alignment (48%) errors on a mathematics calculations test than did girls with fragile X syndrome (19% and 14%, respectively). No group differences were found for procedural or multiplication table errors. Girls with TS attempted more "unfamiliar" problems than did girls with fragile X syndrome or girls in the comparison group. Mathematics achievement scores in girls with TS were positively correlated with Judgment of Line Orientation and Wechsler Intelligence Scale for Children–Revised Third Factor scores; these correlations differed from those in the other groups.**

Conclusions. **The qualitative group differences observed further support the concept of specificity of the TS phenotype and illustrate the importance of a process approach to assessment.** ***Pediatrics*** **1998;102:492–496;** ***Turner syndrome, mathematics, mathematics disability, fragile X syndrome.***

ABBREVIATIONS. TS, Turner syndrome; FSIQ, Full-scale IQ; RT, response time; WJ–R, Woodcock Johnson–Revised; WISC–R, Wechsler Intelligence Scale for Children–Revised; TOVA, Test of Variables of Attention; JLO, Judgment of Line Orientation; ROCF, Rey Osterrieth Complex Figure; P/O, perceptual/organizational.

Turner syndrome (TS), a common chromosomal disorder that occurs only in girls, is caused by the partial or complete absence of one X chromosome. Physical, psychosocial, and cognitive phenotypes have been described in girls with TS. This report addresses global and specific aspects of the cognitive phenotype of TS.

The concept that TS leads to a specific cognitive phenotype is supported by a variety of studies, but the degree of the effects varies and many girls with TS have average to above-average intelligence. It might be concluded from an absence of global intellectual effects that a person with TS is not affected cognitively by her condition, but a detailed assessment of the processes that underlie cognitive performance may suggest otherwise. The latter notion is examined in this article, through a preliminary evaluation of mathematics performance in girls with TS. Also illustrated is the need to base profile descriptions of performance on specific features in addition to standard scores, which are commonly used as primary, if not sole, descriptors of function.

Descriptions of the TS phenotype include a higher verbal than performance IQ[1] and generalized deficits in visuospatial tasks in the context of average Full-scale IQ (FSIQ).[2–4] The visuospatial difficulties appear to be related to poor planning and organization, a finding that implicates a potential executive function component.[1] These findings are consistent with data that show improved visuospatial performance in a group of girls with TS who received verbal mediation training.[5] In addition to visuospatial deficits, depressed mathematics performance in girls with TS is indicated by lower standard scores in mathematics achievement than in reading achievement.[6,7]

To move beyond the global description of girls with TS as having mathematics difficulties, the specificity of their mathematics skills performance can be evaluated in terms of the well documented developmental sequence of the acquisition of mathematics skills.[8,9] This can be accomplished by recording the response time (RT), strategies used, and types of errors made in solving arithmetic problems, in addition to simply observing response accuracy. With the exception of work by Rovet and colleagues,[6] much of the current literature on mathematics skills in girls with TS is based solely on standard scores from achievement and IQ tests. Assessing basic skills and performance affords a more thorough profile of cognitive components in mathematics functioning than would assessing complex tasks only, and may serve to contribute to our understanding of the TS profile.

Neuropsychological correlates of mathematics performance also contribute to understanding the processes that underlie mathematics ability or disability. Features that may lead to poor mathematics performance are poor reading,[10] visuospatial,[11] and attentional or executive function skills.[12] In view of the lack of reading deficits in girls with TS, an association between reading and arithmetic is not

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predicted. Executive function skills are deliberate, goal-oriented skills such as planning, purposeful shifts in attention, and organizational strategies. The evidence for attentional and executive function deficits in girls with TS is consistent with the findings that girls with TS have less adequate use of mathematics procedural skills and poorer retrieval of mathematical facts.[6]

In addition to examining components and correlates of mathematics function, making comparisons with other groups who have mathematics difficulties can define further the specificity of the mathematics profiles in girls with TS. The effectiveness of this approach is illustrated by efforts to specify the difficulties in social skills reported for girls with TS. Girls with fragile X syndrome, a condition that results from a mutation of the *FMR1* gene on the X chromosome, also are described as having difficulties in social skills and mathematics, even when their IQ scores are within the normal range.[13,14] Approximately 50% of girls with fragile X syndrome have mental retardation, and the remaining 50% are an important comparison group for girls with TS.

In the present report of a retrospective analysis, mathematics performance in girls with TS is compared with that in girls with fragile X syndrome and in girls with neither disorder. For some analyses, the performance of a subgroup of girls younger than 10 years was examined for addressing whether the mathematics difficulties reported in girls with TS are evident early in development. The comparisons included analyses of the errors made and the types of problems for which difficulty was observed. The findings are discussed in terms of the importance of using a process approach to describe the cognitive phenotype of girls with TS.

METHODS

Subjects

The study subjects were girls from a larger study of learning disabilities in school-aged children that was conducted at the Learning Disabilities Research Center at the Kennedy Krieger Institute. The sample in the present study consisted of 96 girls 5 through 16 years of age, including 29 with TS, 26 with fragile X syndrome, and 41 with neither syndrome, who were control subjects. Fifty-one of the subjects were younger than 10 years (17 with TS, 13 with fragile X syndrome, and 21 control subjects). There were no age differences in the overall group or in the younger group ($P > .61$). The girls with TS or fragile X syndrome had significantly lower Woodcock Johnson–Revised (WJ–R)[15] Math Cluster scores than Reading Cluster scores, which is consistent with the literature on both disorders.

Measures

Each girl had undergone an assessment battery that included the Wechsler Intelligence Scale for Children–Revised (WISC–R)[16] and four subtests of the WJ–R Test of Academic Achievement, namely, two reading (Letter–Word Identification and Passage Comprehension) and two mathematics (Math Calculations and Applied Problems) subtests. Standardized weighted composite scores, the Reading Cluster and Math Cluster scores, were derived from the two WJ–R subtests per domain. The Test of Variables of Attention (TOVA)[17] was used as a computerized evaluation of inattention and impulsivity. Two visuospatial tasks also were administered: the Judgment of Line Orientation (JLO)[18] test, which involves matching lines on the basis of angular displacement, and the Rey Osterrieth Complex Figure (ROCF) drawing task,[19] from which an "inventory score" was derived for assessing the number of details represented in each subject's reproduction of a complex design that remained in her view. Age-referenced scores were calculated for all of these tests. In addition, process-oriented measures were obtained from the Math Calculations subtest, as described below.

RESULTS

Academic Achievement Profiles

Cluster-score comparisons were made by using paired *t* tests only for girls younger than 10 years. The results were similar to those reported earlier for the entire group, with significantly lower Math Cluster scores than Reading Cluster scores in girls with TS ($t(16) = -5.686$; $P < .0001$) or fragile X syndrome ($t(12) = -5.554$; $P < .001$), but not in the control subjects ($P = .11$). The magnitude of the difference between the reading and the mathematics scores was comparable in girls with TS and girls with fragile X syndrome, in both the overall group and the younger subgroup. However, both the reading and the mathematics scores were higher in the girls with TS than in those with fragile X syndrome.

All four individual achievement subtest scores were compared in the three groups by using multivariate analysis of variance, with group status being the independent variable. Achievement score profiles varied as a function of group status ($F_{(8,178)} = 3.963$; $P < .001$). This main effect of group status remained when FSIQ was entered as a covariate ($F_{(8,176)} = 3.197$; $P < .005$) and when the sample consisted of only those girls younger than 10 years ($F_{(8,90)} = 2.864$; $P < .01$). The standard scores on each mathematics subtest were significantly lower than the standard scores on each reading subtest in the girls with TS or fragile X syndrome ($P = .0002$ to $.023$). This profile of lower mathematics scores than reading scores (by more than 5 points) was observed in the majority (≥70%) of girls with TS or fragile X syndrome, regardless of age or IQ score, and the Math Cluster score was lower than the Reading Cluster score by at least 12 points in more than half of these girls.

The mean differences in reading and mathematics scores were similar in girls with TS or fragile X syndrome (Table 1). However, this similarity in the degree of the difference does not imply a similar quality of mathematics performance. Evidence of qualitative differences between these two groups was obtained through error and correlation analyses, as described below.

Item Analyses

For item analyses, the number of girls who correctly answered a particular test item was examined as a function of group status. The age of the subject is a primary determinant of which items are administered; thus, it was important to analyze these data from individually age-matched groups of subjects. Therefore, a subset of 20 girls per group, matched for age across all three groups,

was chosen for these analyses. Each item on the Math Calculations and Applied Problems subtests was examined by using χ^2 analysis to determine the frequency of correct responses per group. No group differences were seen in this set of analyses, indicating that the groups did not differ with respect to having more difficulty on specific mathematics items or types of items.

Error Analyses

The types of errors that were made on the WJ–R Math Calculations subtest could be evaluated from the written calculations that the girls made on the record form. The types of errors were categorized by an examiner (M.M.) who was blind to the group status of each child. The categories of errors were derived from the literature on mathematics skills, and consisted of 1) operation errors (eg, subtracting instead of adding); 2) table errors (eg, $5 \times 5 = 20$); 3) calculation errors (eg, being off by 1); 4) procedural errors or "bugs," such as subtracting the smaller digit from the larger digit regardless of its position in a vertically presented problem; and 5) alignment errors. Errors that did not correspond to any of these categories and for which a cause could not be deduced were coded as "others."

Fisher's exact test was used for examining the number of subjects in 21 age-matched pairs of girls with TS or fragile X syndrome who made the different types of errors. Also examined was the ratio of the number of problems completed correctly to the number of problems attempted. The results indicate that a higher percentage of girls with TS made operation errors than did girls with fragile X syndrome (12/21 [57%] vs 4/21 [19%]; $P = .014$). The percentage of girls who made alignment errors (including improper placement of decimal points) was higher in those with TS than in those with fragile X syndrome (10/21 [48%] vs 3/21 [14%]; $P = .025$). There were no significant group differences in the percentage of girls who made table, calculation, procedural, or "other" errors. These results were not attributable to a greater number of girls with TS making errors per se, because an equivalent number of girls in each group made errors (20 of 21 and 19 of 21 in the TS and fragile X syndrome groups, respectively). However, the girls with TS made more errors (mean, 6.0 ± 3.2) than did the girls with fragile X syndrome (mean, 3.6 ± 3.1; $t(40) = -2.33$; $P < .05$).

An additional finding concerned the number of girls in each group who attempted problems that were "unfamiliar," defined as problems with an age-equivalence rating 3 years greater than the child's chronologic age. The percentage of girls who attempted unfamiliar problems was higher in the TS group than in the fragile X syndrome group (12/21 [57%] vs 4/21 [19%]; $P = .025$).

Neuropsychological–Mathematical Correlates

Qualitative differences in mathematics skills performance also were examined for all subjects, through correlations between their scores on mathematics and neuropsychological measures. Because of the nonnormal distribution of the neuropsychological scores, Spearman correlation coefficients were calculated, with α set at .01 to correct for multiple comparisons. The control group was divided into two subgroups, unaffected siblings of girls with TS or fragile X syndrome and nonsibling peers. These subgroups were differentiated because the prevalence of academic difficulty appeared to be higher in the peer subgroup, possibly because the parents of the nonsibling peers had responded to advertisements to participate in a study of learning disabilities in girls. The mean differences between FSIQ and Math Calculations or Reading Cluster scores were both slightly higher in the peer group than in the sibling group, but these differences were not statistically significant. However, an unpaired t test showed that the FSIQ scores were significantly lower in the peer group than in the sibling group ($t(39) = 4.523$; $P < .0001$), as seen in Table 1.

The neuropsychological variables were seven age-referenced normative scores, as summarized in Table 2. Each of these scores was converted to an age-referenced z score, with a positive z score always indicating a more favorable performance. The correlation results are presented in Table 3.

The correlations between the mathematics achievement and neuropsychological scores differed between the TS and fragile X syndrome groups. The JLO score was the strongest predictor of the Math Cluster score in girls with TS, but it was the weakest predictor of the Math Cluster score in girls with fragile X syndrome. However, the Perceptual/Organizational (P/O) Factor score was a strong predictor of mathematics performance in girls with fragile X syndrome, but not in those with TS.

The correlation profiles of the peer and sibling groups differed from each other, as well as from the two group profiles described above. In the peer

TABLE 1. Descriptive Scores Among Four Groups

	TS ($n = 29$)	Fragile X Syndrome ($n = 26$)	Peer Control Subjects ($n = 25$)	Sibling Control Subjects ($n = 16$)
FSIQ range	72–135	66–116	68–114	92–132
Mean (SD) FSIQ	95.0 (13.5)	85.8 (14.3)	88.9 (16.0)	109.9 (11.8)
Mean difference in mathematics and reading scores*	14.3	12.5	2.8	3.3
Mean difference in mathematics and reading scores†	18.0	18.4	6.1	1.1

* Mean across all subjects.
† Mean across subjects younger than 10 years old; $n = 17$, 13, 14, and 7, respectively.

TABLE 2. Neuropsychological Variables Included in the Analyses

Measure	Task	Basis for Score	Function Assessed
TOVA omissions	Not responding when target stimulus appears on computer screen	Percent error	Inattention
TOVA commissions	Responding to nontarget stimulus that appears, when instructed not to	Percent error	Impulsivity
JLO	Matching lines on the basis of angular displacement	Total correct/30	Two-dimensional spatial orientation
ROCF Copy–Inventory	Reproducing a complex drawing	Number of features correctly included in drawing/36	Spatial organization, spatial perception
WISC–R VC Factor	Responding to various questions presented verbally	Quality and accuracy of responses	Ability to express information and concepts verbally
WISC–R P/O Factor	Reconstructing abstract and concrete constructions, recognizing details	Speed and accuracy of responses	Aspects of spatial and organizational skills
WISC–R Third Factor	Short-term verbal memory, mental calculation	Speed and accuracy of responses	Aspects of attention, short-term memory

group, all three IQ and both visuospatial scores were positively correlated with the Math Cluster score, indicating a general tendency for a global level of performance across domains. In contrast, there were no significant correlations across the measures examined in the sibling group. This disparity between the comparison groups may have resulted from the smaller number of siblings (16) than peers, (25) but the differences were in quality, not magnitude. Regardless of the explanation for these differences, the variations in the correlation profiles show the potential variability in the processes that underlie mathematics ability and, perhaps, mathematics disability.

DISCUSSION

These preliminary data are in agreement with several other reports that girls with TS are weak in mathematics achievement relative to reading achievement. These data expand on earlier findings by showing that relatively low mathematics achievement is evident before 10 years of age and that there appears to be some specificity in this domain. A higher percentage of girls with TS made operation and alignment errors (57% and 48%, respectively) on a mathematics calculations test than did girls with fragile X syndrome (19% and 14%, respectively), another condition that is associated with mathematics difficulty. No group differences were seen for procedural or table errors. The Math Cluster scores in girls with TS were positively correlated with their JLO and WISC–R Third Factor scores, but not with their P/O Factor scores. These correlations differed from those in the fragile X syndrome, peer, and sibling groups. The primary neuropsychological features responsible for mathematics performance level cannot be determined from these correlations, but the qualitative differences in mathematics–neuropsychological associations observed further support the concept of specificity of the TS phenotype.

Another group difference that was seen in this study was that girls with TS attempted more "unfamiliar" problems than did girls with fragile X syndrome or control subjects. The possible interpretations of this result relate to the metacognitive components of mathematics skills or the impact of mathematics difficulty on a person's willingness to attempt obviously challenging problems. Perhaps the girls with TS were less aware of their own limitations than were the girls in the other groups, or perhaps they were simply more willing to attempt obviously challenging problems than were the other girls. Other factors, such as motivation and anxiety, may be responsible for these findings.

There were no group differences observed in the accuracy rates for the individual test items or item types, a finding that may be attributable to a true lack of group differences, to low statistical power, or to limitations of the WJ–R assessment. The test limitations result from the standard administration of the test, in which only those items that are between a child's basal and ceiling levels are presented, and from the design of the standardized test. For instance, the presentation of different operation problems and horizontally versus vertically presented problems is not counterbalanced in the WJ–R. The limitations illustrate the need for supplements to standardized tests when the goal of assessment is to specify a phenotype. The detailed, qualitative information needed to determine a phenotype may be obtained by using supplemental variables derived from the score on a standardized test (as was done by Rovet and colleagues[6]) or by using additional test materials.

Another variable that is important for a process-approach evaluation of mathematics performance is the RT to complete individual problems, including the problems that are solved correctly. Comparisons of a subject's performance on timed and untimed measures may be insufficient because of the potential effect on the subject's anxiety level that may result if the subject is aware of being timed. RT was not evaluated in this study because of the retrospective nature of the analyses: no RT data had been recorded as a component of the girls' assessments. In an ongoing study that includes the recording of RT, there are to date insufficient data for statistical analysis.

TABLE 3. Correlations of Math Cluster Scores with Neuropsychological Measures

	Spearman Correlation Coefficient			
	Fragile X Syndrome (n = 26)	TS (n = 29)	Peer Control Subjects (n = 25)	Sibling Control Subjects (n = 16)
TOVA omissions	.52*	.26	.45	.04
TOVA commissions	.50	.34	.16	−.03
JLO	.14	.57*	.70**	.45
ROCF Copy–Inventory	.31	.23	.64*	.32
WISC–R VC Factor	.69**	.46	.65*	.23
WISC–R P/O Factor	.51*	.20	.71**	.09
WISC–R Third Factor	.75***	.56*	.85***	.42

* $P < .01$; ** $P < .001$; *** $P < .0001$.

There is, however, anecdotal evidence of the importance of this variable for two reasons: 1) differences in RT may provide objective information about the retrieval of mathematical facts, which is an important component of mathematics performance; and 2) RT data may lead us to question the appropriateness—or accuracy—of describing a person as "unaffected" on the basis of average or above-average standard scores.

Fine-tuning the description of "mathematics difficulty" in girls with TS can provide a foundation for assessment and intervention. In view of the evidence for executive function components of the visuospatial difficulties reported in girls with TS, it is interesting to consider the possibility that executive function difficulties drive both visuospatial and mathematics difficulties, which also would lead to an observed association between the latter two domains of function. Thus, specifying the mathematics profiles in girls with TS may contribute to our understanding of the TS phenotype.

A measurable, specific cognitive feature also may serve as the focus of neurodevelopmental studies designed to address whether function is associated with the specific neuroanatomic structures or anomalies that have been described in the literature. Reiss and colleagues[20] have described subtle but consistent findings of lesser proportions of gray and white tissue in both the right and the left parietal regions, and greater proportions of gray and white tissue in the right inferior parietal–occipital region, in girls with TS than in girls without TS. However, the volumes of these regions were not significantly correlated with the psychological scores that were examined, which included the scores from the WISC–R, JLO, and ROCF. Perhaps the identification of a more specific cognitive effect will afford an opportunity to explore neurodevelopmental and psychological variables further. The identification of specific features of the mathematics profiles in girls with TS is one primary aim of our ongoing research.

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