

Original Articles

No Widespread Psychological Effect of the Fragile X Premutation in Childhood: Evidence from a Preliminary Controlled Study

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ABSTRACT. This study was designed to examine the effect of the fragile X premutation (pM) on cognitive function and behavior. Participants included 14 children (7 males, 7 females) with the fragile X pM and 14 children without the fragile X pM (and without the fragile X full mutation [fM]), each of whom was matched by age and gender with one of the participants from the pM group. The children ranged in age from 3 years, 1 month, to 17 years, 11 months. Participants were individually administered measures of intellectual functioning, academic achievement, and visual motor integration. Parent rating scales of problem behaviors were completed. Group differences were examined using nonparametric statistics. No statistically significant differences were found between the premutation and nonpremutation groups. The results from this study are consistent with the hypothesis that the premutation does not, in general, have an effect on a child's development. However, this does not preclude cases where specific factors may lead to a specific phenotype. *J Dev Behav Pediatr 22:353-359, 2001.* Index terms: *fragile X, premutation, psychological effects, FMR1.*

The discovery of the fragile X mental retardation-1 (FMR1) gene in 1991 provided new opportunities to better understand the molecular genetic influences associated with the fragile X syndrome. Of particular importance to this level of understanding was the discovery that a threshold of approximately 200 cytosine guanine guanine (CGG) trinucleotide repeats differentiated the fragile X full mutation (fM) from the fragile X premutation (pM).¹ In this report, the potential effects of the FMR1 pM on psychological development are explored. Understanding whether the pM affects development is important for practitioners who may encounter patients whose "fragile X" diagnosis is based on DNA results showing a premutation and not a full mutation. Currently, it is not known whether such a diagnosis is appropriate.

The fragile X fM is defined not only by expansion size but also by hypermethylation, which is ultimately associated with a decrease in FMR1 protein level. It is believed that this decrease in protein level is a key biological component of the fragile X syndrome,² although it is only one factor that affects the degree of involvement. Features of the syndrome include cognitive, behavioral, and physical effects, as reviewed elsewhere.³ Mental retardation is present in the vast majority of males, and in approximately 50% of females, with the fragile X fM. The behavioral features of fragile X syndrome include hyperactivity, autistic features (such as hand flapping), and social anxiety, although not all of these features are seen in every individual with the fragile X fM. The physical features most frequently observed include a long face, prominent ears, and hyperextensible joints, although the degree to which these features are present also varies across individuals.³

In contrast to the fM, individuals with the fragile X pM have a mutation that is usually unmethylated. Earlier reports indicated that individuals with the pM do not manifest

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reduced levels of FMR1 protein⁴; more recently, elevated messenger RNA has been reported for individuals with the pM,^{5,6} and reduced FMR1 protein has been reported for some persons with the pM.⁷ These biological factors, as well as other factors, may explain the variability of psychological effects reported for children with the pM.

For parents of children diagnosed with the fragile X pM, a frequently asked question concerns whether the pM will affect their child's development, and if so, to what extent. There is some evidence to suggest that both males and females with a pM, particularly a large pM, show mild physical features of the fragile X syndrome phenotype, including a high arched palate, hyperextensible joints, and ear prominence.⁸⁻¹⁰ The literature regarding effects of the pM on cognitive and psychosocial development is inconsistent. Hagerman⁸ reported on three boys with fragile X premutations in the upper range (130–210 repeats) who showed a mild version of the fragile X syndrome and concluded that the pM may have effects. More recently, Aziz and colleagues¹¹ found that six boys with the fragile X pM (55–200 repeats) or "intermediate" sized alleles (41–54 repeats) showed a range of variability in behavioral and intellectual functioning, including autistic-like behaviors and other social difficulties. Lachiewicz and colleagues¹² reported on 15 individuals with the pM who also had developmental disabilities, thus raising the question of whether individuals with the pM are at greater risk for such disabilities relative to the general population. However, no increase in prevalence of the FMR1 gene was found in a sample of 1014 clinically referred school-aged children who presented for a wide range of academic difficulties,¹³ nor in a sample of 534 clinically referred preschoolers referred for developmental delay.¹⁴ The pM was no more common in the two clinical samples than in the general population.

In addition to reports of the pM leading to cognitive effects, there is evidence that fragile X pM carriers are cognitively unaffected.¹⁵⁻¹⁸ For example, Mazzocco and colleagues^{15,16} found that female carriers of the pM did not have deficits in measures of visual-spatial skills, attention, or executive function relative to a group of females with no fragile X mutation, and similar findings were reported by Franke and colleagues.¹⁹ Mazzocco and Holden¹⁷ found that three sisters, each of whom inherited two fragile X pM alleles, did not show cognitive disability. It may be that case studies of children with the pM who appear to be affected may not be truly representative of persons within the pM group and that these cases are "coincidences."⁴ The present study was designed to further examine the potential effects associated with the fragile X pM. In particular, the goal of the study was to better understand whether the fragile X pM has an effect on a child's psychological development.

In the present study, children with the FMR1 pM were compared to children who had neither the full mutation nor the premutation. If the fragile X pM does affect a child's cognitive or psychological development, participants in the pM group would be expected to demonstrate performance deficits in the same areas as children with the fM, although to a lesser degree. On the basis of reports on

effects of the full mutation and on the basis of the hypothesis that the pM affects development, deficits in the following areas would be expected. In terms of cognitive performance, deficits in performance IQ (PIQ), mathematics subtests, and visual-motor integration tasks should emerge in the pM group, if the pM affects development. In terms of behavioral manifestations consistent with full mutation effects, differences on parent ratings would also be expected in the areas of withdrawn, anxious, or depressed behaviors, social and attention problems, stereotypes, and hyperactivity if the pM affects development. The measures selected for use in this study included measures of these skills for which a difference was expected and measures of skills for which no difference was expected, including verbal IQ, reading skills, and parent ratings on somatic complaints, thought problems, delinquent behaviors, and aggressive behaviors. The measures were administered to children in a controlled study and not from clinical samples.

METHODS

Participants

Participants included 14 children (7 males, 7 females) with the fragile X premutation (pM), and 14 children (7 males, 7 females) without a fragile X mutation (either the pM or the full mutation [fM]). Each child from the pM group was individually matched by age and gender with one of the children from the non-fragile X comparison group. Age matches were within two years for all 14 pairs of subjects, including 7 for whom the difference was one year or less. Participants ranged in age from 3 years, 1 month, to 17 years, 11 months (mean age 10 years, 4 months), and all but one of the participants were relatives

Table 1. Individuals Participating in the Study

Subject Pair	Subject Group		
	pM		No Mutation Age (yr, mo)
	Age (yr, mo)	pM Size Estimate ^a	
1	6, 11	83	6, 10
2	6, 11	83	7, 1
3	10, 9	117	10, 1
4	16, 10	105	17, 11
5	7, 8	90	7, 11
6	13, 11	80–97	13, 11
7	4, 1	147–163	3, 1
8	6, 8	52 ± 3 ^a	8, 2
9	9, 8	200	9, 10
10	4, 11	150	3, 4
11	16, 7	90	15, 2
12	9, 8	100	10, 9
13	17, 10	47–63	16, 1
14	13, 11	105–122	13, 7

pM, premutation.

^aAll pM estimates are listed as noted in Southern blot (SB) analysis report, except for pM Patient 8, whose PCR analysis report is listed in the table. The SB estimate previously reported for this patient was 58 ± 3.

(i.e., a sibling or a cousin) of children with the fragile X fM. Of the 14 children with the pM, 6 had a brother with the fM, 1 had a sister with the fM, and 6 were known to have a male cousin with the fM. The exception was a girl whose pM allele was identified when she was brought to clinical attention because of developmental delays. She had no family history of fragile X or mental retardation. Of the 14 children with no fragile X mutation, 5 had a brother with the fM, 5 had a sister with the fM, 3 had both a brother and a sister with the fM, and 1 was known to have a male cousin with the fM. Participants were identified and recruited either from a registry of families known to have fragile X syndrome (who had given prior permission to be contacted about research opportunities) or through families who contacted us through recruitment notices. Diagnoses for the fragile X pM and non-fragile X mutation status were confirmed either by obtaining a DNA report from the participant's family (1 from the pM group, 4 from the no-mutation group) or by repeating Southern blot or polymerase chain reaction (PCR) DNA analyses (13 from the pM group, 10 from the no-mutation group). For analyses using pM size, midpoint values were used if a range of sizes was reported. Age and pM data appear in Table 1.

The study protocol was approved by a Joint Committee on Clinical Investigation of Johns Hopkins University School of Medicine. Informed consent was obtained from the parents (or legal guardians) of all research participants. Families were not paid for their participation. In some cases, partial reimbursement for travel was offered, depending on the family's location of residence.

Procedure and Materials

Participants were seen at the Kennedy Krieger Institute in Baltimore, Maryland, for a 3- to 4-hour evaluation, with the exception of three children who were seen in the city in which they resided. Each participant was individually administered a test of academic achievement, a visual motor integration task, and a measure of global intellectual functioning. Each parent (or guardian) of a participant completed behavior problem questionnaires regarding the child, and biological mothers of participants were administered a measure of intellectual functioning. Mothers of all 14 of the children in the pM group and mothers of 12 of the children in the no-mutation group were themselves carriers of the pM.

Academic Achievement. Five subtests of the Woodcock Johnson Test of Academic Achievement-Revised (WJ-R)²⁰ were administered, when age appropriate. The Letter-Word Identification subtest involved having children read aloud visually presented letters and words. All participants were administered this subtest. The Passage Comprehension subtest involved reading passages silently and completing the passages by identifying missing words. The Math Calculations subtest consisted of paper and pencil mathematical problems. Twenty-five of the 28 participants were administered these two subtests. The Math Applied Problems subtest involved reading word problems and indicating the answers. Twenty-seven of the 28 participants were administered this subtest. The Word Attack subtest is a

non-word reading task used to assess phonological decoding skills. It was an age-appropriate subtest for only 24 of the 28 participants. Standard scores for all of the five subtests were based on age-referenced normative data.

Visual Motor Integration. The Beery-Buktenica Developmental Test of Visual-Motor Integration (VMI)²¹ was administered as a measure of the child's ability to integrate fine motor skills with visual-spatial perception. For this test, children were presented with a booklet of drawings in which they were asked to copy a model that remained in their view. All participants were administered this test. Standard scores for this test were based on age-referenced normative data.

Intellectual Functioning. A test of overall intellectual functioning was administered to each child. The test used depended on the child's age at the time of the evaluation. The majority of the children (8 pM, 10 non-fragile X) were administered the Wechsler Intelligence Scale for Children, Third Edition (WISC III).²² Children less than 6 years of age (2 pM, 2 non-fragile X), as well as two children over age 6, were administered the Stanford-Binet, Fourth Edition (SB-IV).²³ Participants 17 years of age (3 pM, 1 non-fragile X) were administered the Wechsler Adult Intelligence Scale, Revised (WAIS-R).²⁴ The WAIS-R was used rather than the WAIS-III because this study was completed before availability of the WAIS-III. Standard scores for all of the cognitive tests were based on age-referenced normative data.

Maternal IQ and Child's IQ. As a means to estimate heritable influences on the child's IQ score, other than the fragile X pM, the WAIS-R was administered to the mothers of participants. Cognitive testing was completed for the mothers of 24 of the 28 children in the study. The biological mothers of two children were not involved in the research, and full scale IQ information was not available for the participating biological mothers of the two remaining children. A discrepancy score was calculated between a child's IQ score and the IQ score of the child's own mother. Raw discrepancy scores were used.

Parental Ratings. Parents of child participants were asked to complete the Achenbach Child Behavior Checklist for children ages 4 to 18 years (CBCL/4-18)²⁵ and the Aberrant Behavior Checklist-Community (AbBC).²⁶ The CBCL is a parent behavior rating scale used for assessing social competence and psychopathology in children. Areas examined by the CBCL include social problems, attention problems, and withdrawn behaviors. Standard scores were based on age- and gender-referenced norms. Parents of 26 participants completed the CBCL. The parents of 2 children did not complete the questionnaire because the version used in this study was not age-appropriate for these children (child's age <4 years). The AbBC is designed for assessing maladaptive or inappropriate behaviors including hyperactivity and stereotypic behavior. Parents of 24 participants completed the AbBC, from which a total raw score was used. The four children whose parents did not complete the AbBC were seen early in the study, before the inclusion of this questionnaire. A form used to calculate familial socioeconomic status using the Hollingshead Four Factor Index²⁷ was completed by the parents of all participants.

RESULTS

Nonparametric tests were used because the sample size was insufficient for determining the normality of the data and because a preliminary examination of the data via scatterplots revealed influential data points. Group differences were examined using unpaired, Mann-Whitney U statistics, thus using all available data points. Analyses were repeated using the paired Wilcoxon Signed Rank Test to maximize statistical power of the pair-wise design. For both Mann-Whitney and Wilcoxon statistics, in cases of tied ranks, p values were adjusted for ties.

Preliminary analyses were carried out to examine possible group differences in socioeconomic status. No group difference was found ($p = .41$) based on scores from all 28 participants. Therefore, socioeconomic status was not included in the subsequent analyses. Analyses confirmed no differences in age between the two groups ($p > .94$). The lack of significant differences was also supported by the Wilcoxon test ($ps > .35$ and $.55$, respectively).

Academic Achievement, Visual Motor Integration, and Intellectual Functioning

As seen in Table 2, no statistically significant group differences were found for any of the five Woodcock Johnson Test of Academic Achievement-Revised (WJ-R) subtests that were administered ($p > .40$ for Mann-Whitney tests and $p > .42$ for Wilcoxon tests). There was no statistically significant group difference for the Beery-Buktenica Developmental Test of Visual-Motor Integration ($p = .17$ and $.13$, for the two analyses, respectively). In addition, no differences between groups emerged for full scale IQ ($p = .09$), verbal IQ ($p = .31$), or performance IQ ($p = .07$). The lack of significant differences in these three IQ scores was also supported by the Wilcoxon test ($ps = .11$,

Table 2. Group Mean and Standard Deviation for Standard Scores on Cognitive Measures^{a,b}

	pM		No Mutation	
	Mean	SD	Mean	SD
WJ-R Achievement Testing				
Letter-Word Identification	106.6	20.1	109.1	18.3
Passage Comprehension	105.2	15.5	108.1	18.0
Word Attack	109.0	12.6	113.6	26.7
Math Calculations	102.5	20.8	103.3	16.7
Math Applied Problems	99.9	16.6	104.8	17.1
Visual-Motor Integration				
VMI	90.5	20.2	97.3	14.0
IQ Scores				
FSIQ (n = 14 pairs)	98.6	16.2	108.9	14.8
VIQ (n = 11 pairs)	99.7	13.4	106.7	13.1
PIQ (n = 11 pairs)	99.7	20.3	114.4	13.3
Mom IQ – Child IQ	1.4	20.2	-4.8	11.6

pM, premutation; WJ-R, Woodcock-Johnson Test of Achievement-Revised; VMI, Beery-Buktenica Test of Visual-Motor Integration; FSIQ, full scale IQ; VIQ, verbal IQ; PIQ, performance IQ.

^aNo significant difference on any group comparison.

^bSample sizes varied across measures as specified in the "Methods" section.

Table 3. Mean and Standard Deviation Scores on Parent Ratings of Child Behavior^a

	pM		No Mutation	
	Mean	SD	Mean	SD
Child Behavior Checklist (T scores) ^b				
Withdrawn	53.4	7.1	52.4	4.9
Anxious/Depressed	51.5	3.3	51.8	4.3
Social Problems	53.6	8.3	50.7	2.1
Attention Problems	53.3	6.0	53.3	5.2
Somatic Complaints	53.2	6.0	52.6	4.6
Delinquent Behaviors	53.2	5.2	51.8	4.9
Aggressive Behaviors	52.0	3.8	51.3	2.6
Thought Problems	55.4	7.8	50.6	2.0
Aberrant Behavior Checklist (raw scores)				
Stereotypies	0.5	0.9	0.1	0.3
Hyperactivity	4.1	6.5	4.2	5.5
Irritability	1.8	3.3	1.9	1.9
Lethargic Behavior	1.5	3.1	.3	.5
Inappropriate Speech	.8	1.3	.6	1.1

pM, premutation.

^aNo significant difference on any group comparison.

^bT scores: mean = 50, SD = 10.

.79, and .20, respectively). No statistically significant difference was found between the premutation (pM) and no-mutation groups in a score reflecting the discrepancy between a child's IQ and the IQ of that child's mother ($p > .75$ and $.72$, for the Mann-Whitney and Wilcoxon tests, respectively).

Behavioral Ratings

Results for the Child Behavior Checklist (CBCL) were based on 26 participants (14 pM, 12 no-mutation). As seen in Table 3, among the CBCL ratings for withdrawn, anxious, or depressed behaviors, social problems and attention problems, there were no significant group differences ($ps > .59$ for all ratings) across both sets of analyses (unpaired and paired). Nor were there any significant group differences for the CBCL ratings of somatic complaints, delinquent behaviors, and aggressive behaviors ($ps > .50$ across both sets of analyses). Although the rating for thought problems approached significance ($p = .08$ for both analyses), the means for both groups were well within the average range as seen in Table 3. This was not an area where group differences were hypothesized. In summary, there were no significant group differences for any CBCL behavior problem ratings.

Parametric results for the Aberrant Behavior Checklist (AbBC) were based on 24 participants (10 pM, 14 no mutation). As seen in Table 3, the AbBC ratings for stereotypies ($p = .12$) and hyperactivity ($p = .63$) were not significantly different between groups; the lack of significant differences was also supported by the Wilcoxon paired test ($ps = .19$ and $.89$, respectively). The remaining AbBC ratings, including irritability, lethargic behavior, and inappropriate speech, also revealed no group differences from either set of analyses ($ps > .40$).

Premutation size and IQ. Among children with the pM, no relation was found between pM size and child's own full scale IQ score (Spearman rank coefficient = $-.088$, $p = .75$). When repeated using parametric procedures, the Pearson's coefficient was $-.131$, $p = .66$. More importantly, no relationship was found between pM size and the discrepancy between the child's full scale IQ score and the IQ score of the child's own mother (Spearman rank coefficient = $-.074$, $p = .79$). When repeated using parametric procedures, the Pearson's coefficient was $.051$, $p = .87$.

DISCUSSION

In this study, no differences in psychological scores were found between children with the fragile X mental retardation-1 (FMR1) premutation (pM) and children with no FMR1 mutation, using the cognitive and behavioral measures described earlier. These results are inconsistent with the hypothesis that the premutation affects development and support the null hypothesis that the pM does not affect a child's psychological development. Although these results are preliminary in view of the small sample size, they are an important contribution to the research and controversy regarding the pM effects. One obstacle to such research is the fact that children with the premutation are not as easily identifiable as are children with the full mutation (fM). Further evidence with larger samples is needed to address this hypothesis more definitively. Additional information of interest concerns FMR1 protein levels and cross-tissue samples in children who appear to be affected by the pM.

Understanding the potential effects of the fragile X pM is important because it may aid families and physicians in the early identification of fragile X premutation carriers and in recognizing the potential risks that may or may not be involved with this diagnosis. It is very important to examine the "effects" of the pM with respect to symptoms or features observed in the general population. In addition to considering the rates or effects seen in the general population, it is also important to consider effects seen in case studies. The examination of case studies helps to identify the variability seen among individuals with the pM. Although one cannot assume that the variation is actually linked to the pM per se, the case reports presented below help to illustrate the variability sometimes seen in clinically ascertained cases of the pM and the care with which the variability should be attributed to the premutation.

Individual Cases

Case 1 involved a girl (age 6 years, 8 months) whose pM allele was identified when she was brought to clinical attention because of mental retardation. This child had no family history of fragile X or mental retardation. She was screened as a potential candidate for the fragile X fM. She had a full scale IQ (FSIQ) score of 65 and a pM allele of 52 ± 3 repeats based on DNA PCR testing. This child was also reported, as "Case 1," by Maddalena and colleagues,²⁸ who found no evidence of a fM after looking in a second tissue type, epithelial tissue, of the child. Although inte-

resting, this case provides inconclusive evidence regarding potential pM effects. It is possible that in Case 1 the child's pM and developmental delay co-occur as a coincidence. It is also possible that her small pM, which was approximately five repeats greater than her mother's, is nonrepresentative of possible fM in her brain. Although "affected," this girl's profile was, at least in part, inconsistent with the fragile X phenotype. At age 6 she was described by her mother as having poor eye contact, difficulty staying focused, and repetitive movements and speech. However, at age 10, a psychologist's report described her as having good eye contact, strengths in visual sequencing tasks, and good peer relationships. In addition, when re-evaluated at the age of 11 years, she showed gains in her academic and cognitive scores. FMR1 protein assay collected at age 11 years revealed normal levels of FMR1 protein, indistinguishable from levels observed in controls. This case illustrates the importance of considering the circumstances under which an individual's pM is identified and of assessing potential ascertainment biases in pM study samples.

Case 2 involved a girl (age 9 years, 8 months) with a borderline FSIQ of 78 and a pM size of approximately 200 repeats. This child was classified as a pM because her results were unmethylated; however, her diagnosis is a questionable borderline pM/fM. Although no methylation was observed in this child's lymphocyte-derived DNA, hypermethylation may have been present in brain cells. No protein analysis was available for this child. Considering the fragile X phenotype, this girl's profile was also inconsistent. Based on observations from her evaluation, she was described as being very cooperative, having good eye contact, exhibiting good attention, and initiating conversations. She was also described as having no emotional or behavioral difficulties based on a questionnaire concerning behaviors associated with the fragile X syndrome.²⁹

Case 3 involved a boy (age 4 years, 1 month) with a FSIQ of 100, whose pM size fell in the range 147 to 163 repeats based on Southern blot analysis. Epithelial-derived DNA revealed an allele of similar size. A protein analysis completed for this child revealed results that were compatible with that of normal or premutation FMR1 alleles. This child was described as having poor eye contact, nervousness or anxiety, and repetitive speech, but not repetitive movements. This case supports the notion that a large pM does not necessarily lead to cognitive effects and that normal protein levels persist in a child with a pM. However, it is important to note that this child was only 4 years old at the time of evaluation, and thus if affected, a later decline in IQ is possible, and whether his behavior is related to the pM is questionable, although unlikely. Declines in IQ scores have been reported among males with fragile X between preschool and later school-age years; however, in most studies, IQ scores are well below 100, even in preschool-aged children.^{30,31}

It is interesting to note that the only child in this study who had mental retardation (Case 1) had a very small pM (52 ± 3 repeats) and that the two children with the largest pMs (Cases 2 and 3) included one child with a normal IQ score (FSIQ = 100). This and the lack of a significant correlation between pM size and IQ may indicate the lack

of prognostic information that a precise measurement of repeat number may offer for children with the pM, with respect to cognitive and behavioral function. The extent to which the case histories provide valuable information regarding overall effects of the pM is weaker. Considered alone, Cases 1 and 2 are suggestive of pM effects. Considered within the group of 14 children, and in the absence of group differences, it is difficult to determine what accounts for these two children's poorer performance. Whether "coincidence" or pM effect, what is clear is that the vast majority of the children seen in the pM group did not have cognitive difficulties.

The "lower" mean scores for children in the pM group relative to children in the no-mutation group, as seen in Table 2, must be examined closely. All mean scores, for both groups, are well within the average range. The discrepancy between a child's IQ score and the IQ score of the child's mother was comparable across the two groups. The FSIQ scores for mothers of children with pM were lower than the FSIQ scores for mothers of children with no mutation. Although not statistically significant ($p = .056$), this tendency for a group difference in maternal FSIQ may well account for the discrepancy (that was also not statistically significant) between children with the pM versus those with no mutation. All of the mothers for whom FSIQ information was available, across both groups of children, had pM alleles themselves, so it cannot be concluded that their FSIQ discrepancy was differentially affected by pM effects of mothers' IQ scores.

In summary, the results from this study do not support the hypothesis that the pM has effects on a child's cognitive or psychosocial development. The two cases suggestive of pM effects are intriguing, but they are limited in their generalizability without being considered relative to the frequency with which they occur within the overall population of pM carriers. These cases also need to be evaluated in terms of ascertainment bias, that is, in terms of who was screened for the pM and why. In this study, the most "affected" child had no family history of fragile X and had actually been screened for the fM because of mental retardation of unknown etiology. Similarly, in several studies that support pM effects, the cases of interest were clinically ascertained.^{11,12} Among the 13 nonclinically ascertained participants in this study, only one (8%) had a below-average IQ score. Finally, if these few cases of interest represent cross-tissue heterogeneity in allele size within an individual, then it may not be the pM leading to "effects." Thus, although there is no definitive explanation for the two cases included in this study, the majority of children with the pM showed no sign of psychobiological effects.

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Literary Quotes

Parental Influence on Children: Baseball at the Kearns House

The professional literature is full of data and theories about the way the thoughts, feelings, and behavior of parents do or do not have a lasting impact on the lives of their children. While the experts are arguing over the importance of parents, the general literature abounds with illustrations that might be included in any comprehensive summary.

Doris Kearns Goodwin (1943–), the popular and much respected historian and journalist, has described how she got interested in baseball under the influence of intensive instruction and a solid relationship with her father:

When I was six, my father gave me a bright-red scorebook that opened my heart to the game of baseball. After dinner on long summer nights, he would sit beside me on our small enclosed porch to hear my account of that day's Brooklyn Dodgers game. Night after night he taught me the odd collection of symbols, numbers, and letters that enable a baseball lover to record every action of the game. . . By the time I had mastered the art of scorekeeping, a lasting bond had been forged among my father, baseball, and me.

She does not seem to have been bothered that there was temporarily an element of deception in this process:

All through the summer, my father kept from me the knowledge that running box scores appeared in the daily newspapers. He never mentioned that these abbreviated histories had been a staple feature of the sports pages since the nineteenth century and were generally the first thing he and his fellow commuters turned to when they opened the *Daily News* and *Herald Tribune* in the morning. I believed that, if I did not recount the games he had missed, my father would never have been able to follow our Dodgers the proper way, day by day, play by play, inning by inning. In other words, without me, his love of baseball would be forever unfulfilled.

Her father made Doris feel important to him, helped her to develop skill at recording information, and cultivated an abiding, satisfying interest in the sport.

Doris Kearns Goodwin: *Wait Till Next Year: A Memoir*. New York, NY, Simon & Schuster, 1997, pp 13, 18

Noted by William B. Carey, M.D.