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WHAT’S KNOWN ON THIS SUBJECT: To date, studies of adaptive behavior in fragile X syndrome have focused on particular age points, either longitudinally or cross-sectionally across a broad age spectrum. Studies have shown variable patterns in adaptive behavior among people with fragile X syndrome.

WHAT THIS STUDY ADDS: This study fills a critical gap in knowledge about the profile of adaptive behavior across childhood, adolescence, and young adulthood in fragile X syndrome. This study is the first to incorporate longitudinal data from an age-matched typically developing group.

OBJECTIVE: To examine longitudinally the adaptive behavior patterns in fragile X syndrome.

METHOD: Caregivers of 275 children and adolescents with fragile X syndrome and 225 typically developing children and adolescents (2–18 years) were interviewed with the Vineland Adaptive Behavior Scales every 2 to 4 years as part of a prospective longitudinal study.

RESULTS: Standard scores of adaptive behavior in people with fragile X syndrome are marked by a significant decline over time in all domains for males and in communication for females. Socialization skills are a relative strength as compared with the other domains for males with fragile X syndrome. Females with fragile X syndrome did not show a discernible pattern of developmental strengths and weaknesses.

CONCLUSIONS: This is the first large-scale longitudinal study to show that the acquisition of adaptive behavior slows as individuals with fragile X syndrome age. It is imperative to ensure that assessments of adaptive behavior skills are part of intervention programs focusing on childhood and adolescence in this condition. Pediatrics 2014;134:315–324

KEY WORDS
fragile X syndrome, adaptive behavior, children, adolescents, Vineland

ABBREVIATIONS
ABC—Adaptive Behavior Composite
Vineland—Vineland Adaptive Behavior Scales

(Continued on last page)
Adaptive behavior, the term used to indicate a person’s ability to function independently in his or her environment, is a dynamic construct changing over the course of a person’s life and dependent on societal expectations. It is a measure of the consistency and ability to conduct a task rather than potential. In other words, it measures what a person does rather than what a person can do. Measures of adaptive behavior are particularly important in assessing people with intellectual disabilities. Not only are deficits in adaptive behavior part of the definition of intellectual disability, but adaptive behavior skills also play a pivotal role in the life success of people with intellectual disabilities. Many syndromes associated with intellectual disability have associated adaptive behavior profiles, the identification of which can be helpful as prognostic indicators and for treatment planning. Adaptive behavior is typically measured in terms of one’s ability to communicate and socialize with others, navigate the daily environment with tasks such as dressing oneself, and use coping mechanisms. As an example, on the Vineland Adaptive Behavior Scales, a common measure of adaptive behavior, children with Down syndrome tend to score lower in communication than in socialization, daily living, or motor skills, whereas children with autism or Prader–Willi syndrome show a strength in daily living skills and a weakness in socialization skills.

Fragile X syndrome, the phenotypic result of a mutation in the FMR1 gene, is the leading known inherited cause of intellectual disability and the leading known single-gene risk factor for autism spectrum disorder, affecting 1 in 4000 boys and 1 in 8000 girls. This syndrome is typically caused by an expansion of trinucleotide repeats on the fragile X mental retardation gene (FMR1) on the long arm of the X chromosome. In people with the FMR1 full mutation, the number of trinucleotide repeats (CGG) typically expands to 200 or more, which results in hypermethylation of the gene and reduction of FMR1 messenger RNA and protein production. Reduction of FMR1 protein in the brain is believed to be responsible for the cognitive–behavioral impairments found in people with fragile X syndrome.

To date, the literature addressing adaptive behavior in children with fragile X syndrome has described profiles of adaptive behavior; how adaptive behaviors change over time, and how people with fragile X syndrome differ from people with other developmental disorders. Results suggest that people with fragile X syndrome have strengths in self-help and daily living skills, with weaknesses in socialization and communication skills. Reports also suggest that there may be a change in adaptive behavior over time, although the direction of this change differs across studies and depends on whether age-equivalent scores or standard scores are examined. Some reports indicate that there is growth in adaptive behavior skills at ~10 years of age, and others suggest a decline in these abilities as people grow older. Declines in standard scores can reflect either a decline in skills or a slowed rate of growth compared with that of their same-age, typically developing peers.

In a study conducted by Hatton et al., 70 children with the FMR1 full mutation were assessed, on average 4.4 times over the course of 8 years (average interval between assessments was 12.9 months; age range 2–12 years). Age was significantly related to age-equivalent scores of adaptive behaviors in boys with fragile X syndrome without a comorbid diagnosis of autism, suggesting that there is a steady increase in adaptive skills as children get older. Other studies indicate that adaptive behavior skill acquisition slows as children with fragile X syndrome age.

For example, Fisch et al. found that retest of standard scores for both IQ and adaptive behavior decreased in nearly all members of a group of 24 boys, 4 to 15 years old, with the fragile X full mutation and also declined in most of the 13 girls with the fragile X full mutation. Declining trajectories seem to be most evident in older children and teenagers. In a longitudinal study conducted by Dykens et al., significant gains in adaptive behavior as indicated by age-equivalent scores were found in boys tested twice with the Vineland before 10 years of age, but those tested twice between 11 and 20 years of age did not show significant gains. After 11 years of age there was a mixed trajectory, with 45% showing modest gains in scores and 55% showing modest declines. The same researchers also conducted a cross-sectional analysis and found that boys aged 1 to 5 years of age showed significant gains in adaptive skills, boys 6 to 10 years of age showed moderate gains in adaptive behavior skills, and boys and young men aged 11 to 15 years and 16 to 20 years showed greater scatter and no upward trajectory in their adaptive behavior skills. Taken together, these results suggest that adaptive behavior seems to increase from ~5 to 11 years of age then reaches a plateau where it remains stable or begins to decline into late adolescence or early adulthood. However, this potential developmental profile has yet to be demonstrated with a large-scale longitudinal study of adaptive behavior beyond the age of 12 years in people with fragile X syndrome.

In terms of patterns of adaptive behavior, significant differences are regularly observed between the skill domains in fragile X syndrome. Previous research examining age-equivalent scores has suggested that daily living skills are a relative strength, whereas social skills are a relative weakness.
However, when standard scores were examined, mean scores on the socialization domain were significantly higher than either the daily living or communication domains.

In summary, studies of adaptive behavior in fragile X syndrome to date have focused on particular age points, either longitudinally or in a cross-sectional manner, across a broad age spectrum. Furthermore, many of these studies have assessed only a small number of participants. As a result, there remains a critical gap in knowledge about the actual profile of adaptive behavior skills across childhood, adolescence, and young adulthood in fragile X syndrome. The results of the study presented here address this gap and also, for the first time, incorporate longitudinal data from an age-matched typically developing control group. Understanding the development of adaptive behavior across development in fragile X syndrome is important from multiple perspectives, particularly in light of the need to identify robust end-point measures given the recent initiation of disease-specific clinical trials for people with this condition.19,20

METHOD

Participants

The participants were 275 people (186 males, 89 females) with fragile X syndrome and 225 people with typical development (122 males, 103 females). All participants with fragile X syndrome were diagnosed with the FMR1 full mutation using DNA analyses. Educational levels of the parents of participants with fragile X syndrome were also assessed. The small number of mothers who were known full mutation carriers (n = 5) had educational levels similar to those of mothers with premutation status.

Participants with fragile X syndrome were recruited through advertisements with the National Fragile X Foundation, genetics clinics, developmental evaluation centers, and early intervention programs. For a proportion of the participants between the ages of 6 and 16 years, their time 1 and Time 2 visit occurred in their home. All subsequent visits and those with the other participants were conducted at Stanford University or the University of North Carolina at Chapel Hill. Typically developing children were recruited in the local area through advertisements or were unaffected siblings of the children who had fragile X syndrome. The parents or guardians of all participants provided informed consent and received a $150 remuneration for each assessment point in which the child or family participated. The study was approved by the Stanford University and University of North Carolina internal review boards.

Measures

Adaptive Behavior Assessment

The Vineland Adaptive Behavior Scales, Interview Edition, Survey Form (the Vineland21) was used to assess adaptive behavior. The Vineland is a semi-structured interview in which the parent or close caregiver provides the responses. Each item is scored on a 3-point scale, from 0 (never performs the task), to 1 (sometimes or partially performs the task), to 2 (usually performs the task). As per Vineland manual instructions, select items could also be scored as N if the child has had no opportunity to perform the task or DK if the respondent has no knowledge of the child’s ability to perform the task. A trained clinician or research assistant conducted the Vineland interview with the primary caregiver at each assessment point. The Vineland provides standard scores and age-equivalent scores for 4 domains: Communication (receptive, expressive, and written), Daily Living Skills (personal, domestic, and community), Socialization (interpersonal relationships, play and leisure time and coping skills), and Motor Skills (gross and fine). An overall Adaptive Behavior Composite (ABC) is also derived based on these 4 domains and provides a comparison with scores of a normative population. The Vineland is normed for children between birth and 18.11 years. The Motor Skills domain provides standard scores and age equivalents for children <6 years of age and estimated standard scores for children >6 years old. The Motor Skills domain thus contributes to the overall ABC score only for children <6 years of age. The Vineland has been reported to have test–retest reliability ranging from 0.76 to 0.9322 and interrater reliability ranging from 0.62 to 0.78.22,23

Procedure

Caregivers participated in the Vineland interview approximately every 2 to 4 years as part of data collection for a longitudinal study of development and neuroimaging of people with fragile X syndrome. The mean time between assessments was 3.33 years (range 2–9 years). This interview was conducted in person along with a larger battery that was administered to both the children and parents of the children with fragile X syndrome. A group of typically developing children and their caregivers served as controls for the study. Of the typically developing participants, 60% were siblings of the participants with fragile X syndrome. These participants tested negative for any form of the fragile X mutation.

Data Analyses

To estimate trajectories of Vineland domains across fragile X and gender, we used a common growth model, also known as linear mixed or hierarchical linear model.24–26 We used maximum likelihood estimation implemented in
and males with fragile X syndrome: the males, females with fragile X syndrome, 4 groups (control females, control males, allowing for differences across the Vineland trajectories of the entire sample). We conducted the analysis assuming that data were missing at random and conditional on observed information. We administered the Vineland to a total of 500 participants. Among the total sample of 500 who participated in the study, those who were administered the Vineland at ≥1 time points were included in the analysis. Thus, 262 participants had only 1 assessment, 186 had 2 assessments, 46 had 3 assessments, and 6 participants had 4 assessments. Within the sample included in our analyses, the age ranged from 1 to 25 years. However, the data are sparse outside the age range of 6 to 18 years for females and 2 to 18 for males. Therefore, we report statistical inferences focusing only on these age ranges by gender.

A quadratic growth model was chosen (Vineland 2 = $\eta_{1i} + \eta_{2i}t + \eta_{3i}t^2 + \epsilon_{it}$) for individual $i$ at age $t$ to properly capture nonlinear developments of Vineland domains over time. Each of the 3 domains and the overall ABC were modeled separately (results reported in Tables 2 and 3). We used actual ages of participants at 4 assessment points to model the effect of age. According to the likelihood ratio test, the model fit the data significantly better in the presence of the quadratic growth parameter; whereas adding a cubic term did not further improve the model fit. Three random effects are included in the quadratic growth models to estimate Vineland trajectories of the entire sample, allowing for differences across the 4 groups (control females, control males, females with fragile X syndrome, and males with fragile X syndrome): the initial status ($\eta_{1i}$), the linear growth ($\eta_{2i}$), and the quadratic growth ($\eta_{3i}$). Individual random variation is also taken into account in the model ($\epsilon_{it}$). These 3 random effects are predicted by fragile X status and gender.

To formally test how the Vineland domains develop differently between males and females with fragile X syndrome and male and female controls separately, we used a similar quadratic growth model but using the 3 individual domains of Vineland simultaneously (results reported in Table 4). Given that 3 domains are modeled simultaneously with a smaller sample size (186 males with fragile X syndrome, 89 females with fragile X syndrome, 122 male controls, and 103 female controls), we used a random intercept model. The random intercepts of the 3 domains are allowed to be freely correlated in the model.

Previous studies that modeled overall and individual trajectories in fragile X syndrome used both standard scores and age-equivalent scores. In this study, we use standard scores to compare specific domain trajectories within group (Socialization versus Communication, Socialization versus Daily Living, Communication versus Daily Living). The use of standard scores appears better suited for such cross-domain comparisons as opposed to age-equivalent scores. As also stated in the Vineland manual, “one year’s growth has a very different meaning at different points in the age continuum and for different areas of adaptive behavior.”

**RESULTS**

Demographic information is shown in Table 1 for participants with and without fragile X syndrome. A wide range of chronological and mental ages was represented in both the participants with fragile X syndrome and control groups, and there was no significant difference between groups at the initial testing session with regard to age, gender distribution, or parental IQ. As expected, the fragile X group had significantly lower cognitive abilities than the control group.

**Estimated Trajectories**

The estimated mean Vineland trajectories based on our linear mixed model estimation are presented in Figs 1 and 2 for males and females, respectively, and overlaid onto the observed data. Tables 2, 3, and 4 show results comparing the estimated trajectories. Table 2 compares estimated Vineland scores from one age to another age (eg, 2 vs 6 years) within group. Table 3 compares groups to one another (eg, males with fragile X syndrome versus control males) at selected ages. Table 4 compares groups to one another (eg, males with fragile X syndrome versus control males) at selected ages.
compares Vineland domains (eg, Socialization versus Communication) at selected ages and from one age to another age in participants with fragile X syndrome.

Within-Group Trajectory
As can be seen in Table 2 and Figs 1 and 2, all estimated mean Vineland standard scores for males with fragile X syndrome decrease significantly over time (see Table 2, most $P$ estimates for males with fragile X syndrome are $<.001$). Vineland standard scores for females with fragile X syndrome decreased significantly only within the Communication domain (see Table 2, all $P$s for females with fragile X syndrome are $<.001$).

Between-Group Comparison at Selected Ages
Vineland standard scores for all domains were significantly lower for males with fragile X syndrome than for control males and for females with fragile X syndrome than for control females at all selected ages (see Table 3, all $P$s for males and females with fragile X syndrome are $<.001$). The discrepancy between males with fragile X syndrome and control males is greater than the discrepancy between females with fragile X syndrome and control females, as indicated by significant interaction effects (see Table 3, most $P$s $<.001$).

Comparison Between Vineland Domains for People With Fragile X
To formally test how Vineland domain scores develop differently within the fragile X syndrome groups (males
and females analyzed separately), we used a similar quadratic growth model, but with the 3 individual domains of the Vineland simultaneously. Table 4 summarizes the statistical comparisons of trajectories of the 3 Vineland domains among the males and females with fragile X syndrome. The estimated mean trajectories of the 3 domains are displayed in Fig 3 (see Figs 1 and 2 for complete data). We also compared Vineland domains for male and female controls (122 control males and 103 control females, see Supplemental Table 5 and Supplemental Figure 4).

Table 4 shows that for males with fragile X syndrome, scores on the Socialization domain were significantly higher than scores on both the Communication and Daily Living Skills domains at all selected ages (all Ps < .001). Furthermore, the decline in Socialization skills was smaller than the decline in both Communication and Daily Living Skills at all ages with the exception of 10 to 18 years, where the decline in Socialization skills was greater than the change in Daily Living Skills (P < .001). With regard to Communication and Daily Living Skills in boys with fragile X syndrome, the most meaningful difference occurred after 14 years of age, when Daily Living Skills increased while Communication Skills continued to decline (P < .001).

For females with fragile X syndrome, a more variable pattern was found. Socialization and Communication Skills were not significantly different from one another through 10 years of age. From 10 to 14 years of age there was a greater decline in
Communication Skills such that, by age 18 years of age, Communication Skills were significantly lower than Socialization Skills \((P < .05)\). The scores for Daily Living were significantly different from Socialization only at age 6 \((P < .01)\) and Communication at ages 6 and 18 \((P < .01)\).

### DISCUSSION

The results of this study demonstrate striking developmental patterns of adaptive behavioral function among children and adolescents with fragile X syndrome compared with typically developing participants. The trajectory of adaptive behavior of people with fragile X syndrome is marked by a significant decline in standard scores on all domains for males and on the Communication domain for females throughout childhood and adolescence. For males with fragile X syndrome, a relative strength throughout development is observed for Socialization Skills as compared with Communication and Daily Living Skills. For females with fragile X syndrome, a clear pattern of strengths and weaknesses across domains did not emerge, that is, the trajectories were similar across domains.

Our study is the first large-scale longitudinal investigation to show decreases...
in adaptive behavior standard scores in people with fragile X syndrome throughout childhood and adolescence. Specifically, we found that the adaptive behavior of males and females with fragile X syndrome decreases more throughout childhood than that of their same-gender, typically developing peers. Most studies to date have been cross-sectional, and those that were longitudinal were limited in sample size. Our findings partially replicate other studies that have found this domain to be a developmental strength for males with fragile X syndrome. This contrasts past studies that have found this domain to be the most aberrant in fragile X syndrome.

The discrepancy may result from higher rates of autism symptoms in other studies using different sampling procedures. Such sampling differences could have created a bias resulting in unlike research samples across studies with respect to social abilities.

We observed Daily Living Skills to increase after 14 years of age in males with fragile X syndrome. This partially replicates other studies that have found this domain to be a strength throughout childhood and adolescence. Daily Living domain scores were also observed to increase over time in females with fragile X syndrome, although, when compared at selected ages, these scores were not significantly different from those in other domains. It is possible that if our sample had included older participants, we would have observed a strength in Daily Living Skills in young adulthood. Age has previously been found to be a predictor of independence for young women with fragile X syndrome.

In addition to restricting our interpretations to selected age ranges (2–18 for males and 6–18 for females), there are other limitations to this study. Some of the initial visits were performed in the homes of the participants, so their time and ability to travel were not a limiting factor for these assessments. Most subsequent visits did require travel. Thus, anxiety and behavioral problems, if present in people with fragile X syndrome, could have deterred ongoing participation. However, issues related to attrition and the study design were mitigated by the use of mixed effect modeling, which allowed us to include participants with only 1 data point in the longitudinal analyses.

Despite this study being one of the largest to examine trajectories of adaptive behavior in people with fragile X syndrome, a larger sample would allow comparison of subgroups based on, for example, autism symptoms, cognitive functioning, and FMR1 protein levels.

### TABLE 4: Comparison of Vineland Scores Between Participants With Fragile X Based on Estimated Trajectories Using Mixed Effects Modeling

<table>
<thead>
<tr>
<th></th>
<th>Socialization Versus Communication</th>
<th>Socialization Versus Daily Living</th>
<th>Communication Versus Daily Living</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Males</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>At age 2</td>
<td>5.001</td>
<td>&lt;.001*</td>
<td>5.456</td>
</tr>
<tr>
<td>At age 6</td>
<td>8.143</td>
<td>&lt;.001*</td>
<td>12.578</td>
</tr>
<tr>
<td>At age 10</td>
<td>11.252</td>
<td>&lt;.001*</td>
<td>15.047</td>
</tr>
<tr>
<td>At age 14</td>
<td>14.269</td>
<td>&lt;.001*</td>
<td>12.862</td>
</tr>
<tr>
<td>At age 18</td>
<td>17.253</td>
<td>&lt;.001*</td>
<td>6.024</td>
</tr>
<tr>
<td>Change from age 2 to 6</td>
<td>3.142</td>
<td>.011</td>
<td>7.122</td>
</tr>
<tr>
<td>Change from age 6 to 10</td>
<td>3.089</td>
<td>&lt;.001*</td>
<td>2.469</td>
</tr>
<tr>
<td>Change from age 10 to 14</td>
<td>3.037</td>
<td>&lt;.001*</td>
<td>-2.185</td>
</tr>
<tr>
<td>Change from age 14 to 18</td>
<td>2.984</td>
<td>.034</td>
<td>-6.838</td>
</tr>
<tr>
<td><strong>Females</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>At age 6</td>
<td>-2.252</td>
<td>.488</td>
<td>9.850</td>
</tr>
<tr>
<td>At age 10</td>
<td>0.676</td>
<td>.828</td>
<td>5.777</td>
</tr>
<tr>
<td>At age 14</td>
<td>3.714</td>
<td>.270</td>
<td>1.581</td>
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<tr>
<td>At age 18</td>
<td>6.862</td>
<td>.008</td>
<td>-2.737</td>
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<tr>
<td>Change from age 6 to 10</td>
<td>2.928</td>
<td>.504</td>
<td>-4.074</td>
</tr>
<tr>
<td>Change from age 10 to 14</td>
<td>3.038</td>
<td>.301</td>
<td>-4.196</td>
</tr>
<tr>
<td>Change from age 14 to 18</td>
<td>3.148</td>
<td>.389</td>
<td>-4.318</td>
</tr>
</tbody>
</table>

For males, $N = 91$ for T1 only, $N = 83$ for T1 and T2, $N = 26$ for T1, T2, and T3, and $N = 0$ for 4 time point measures. For females, $N = 38$ for T1 only, $N = 34$ for T1 and T2, $N = 17$ for T1, T2, and T3, and $N = 0$ for 4 time point measures. *$P < .0001$. **$P < .05$. ***$P < .01$. ****$P < .001$. 

For standard scores to better capture change over time, given the consistent distribution of scores across all ability levels and ages.29,30

In examining profiles of adaptive functioning, we found a relative developmental strength for males with fragile X syndrome in Socialization compared with Communication and Daily Living Skills. Over time, Socialization Skills decreased the least as compared with the other domains. This contrasts past studies that have shown Socialization Skills to be the most aberrant in fragile X syndrome.2 This discrepancy may result from higher rates of autism symptoms in other studies using different sampling procedures. Such sampling differences could have created a bias.
CONCLUSIONS

Using a large-scale, longitudinal study we were able to elucidate trajectories of adaptive behavior across a wide age range in males and females with fragile X syndrome. The observed declines in adaptive behavior standard scores across childhood highlight the importance of educational and community programs focused on improving these skills. For example, parent training programs have been shown to have a positive impact on improving adaptive behavior. It is important to be aware of developmental periods when skills are particularly likely to diminish in fragile X syndrome so that those working with affected people can attempt to preserve behavioral sets that are most vulnerable. Understanding developmental trajectories in people with fragile X syndrome will also be of value in understanding and interpreting the effects of new treatments for this disorder. Currently, there are several ongoing clinical trials of disease-targeting medications in fragile X syndrome that use behavioral end points. Adaptive behavior could be a powerful outcome measure in such trials, particularly those with an extended trial period, to assess whether improvements in functioning generalize to functional day-to-day skills.

ACKNOWLEDGMENTS

We especially thank the many families with fragile X syndrome who devoted their precious time and energy to the project.

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females with fragile X. Neuropsychology. 2003;17(4):646–657


(Continued from first page)
An error occurred in the article by Aronson et al, titled “Variation in Care of the Febrile Young Infant <90 Days in US Pediatric Emergency Departments.” Pediatrics. 2014;134(4):667–677, doi:10.1542/peds.2014-1382. On page 669, in the Table 1 originally published, the row and column formatting was incorrect. With the exception of the top row of the table, the numbers all need to be moved up 1 row, and the left-hand column, last row heading “Median age, d (IQR)” can be deleted as it is also listed in the top row. The correct table accompanies this erratum.

doi:10.1542/peds.2014-3232


The following errors occurred in the article by Cheryl Klaiman et al, titled “Longitudinal Profiles of Adaptive Behavior in Fragile X Syndrome” published in the August 2014 issue of Pediatrics (2014;134(2):315–324; doi:10.1542/peds.2013-3990). On page 315, under the Author section, Lindsay C. Chromik was accidentally omitted. The correct authorship listing is: Cheryl Klaiman, PhD,a,b Eve-Marie Quintin, PhD,c Booil Jo, PhD,d Amy A. Lightbody, PhD,d Heather Cody Hazlett, PhD,e,f Joseph Piven, MD,e,f Scott Hall, PhD,d Lindsay C. Chromik, MS,d and Allan L. Reiss, MDg,h. Her contributions are as follows: Lindsay C. Chromik assisted with study recruitment and data collection, assisted with data analysis, reviewed article drafts, and approved the final manuscript.

doi:10.1542/peds.2014-4011


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The online version of this article, along with updated information and services, is located on the World Wide Web at:
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Supplementary Material
Supplementary material can be found at:
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References
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