Increasing Physical Activity of Children With Cystic Fibrosis: A Home-Based Family Intervention

Beverly J. Tuzin, Mary M. Mulvihill, Kristin M. Kilbourn, Deborah A. Bertran, Michael Buono, Melbourne F. Hovell, Ivan R. Harwood, and Michael J. Light

This study evaluated a home-based, parent-managed, behavioral program to increase routine physical activity of ten 7- to 14-year-old children with cystic fibrosis. In each of 3 replications of a multiple baseline design, physical activity increased only after the intervention was initiated. Eight children increased total activity 42.5% to 321%, and 2 children exercised more consistently. Study II recorded further activity increases at 6-week follow-up. Study III validated reported activity with increases of 7% to 27% in VO$_2$max, 8% and 31.6% in V$_{e}$max, and 14.2% and 20% in Wmax. Results suggest that a home-based contingency management program can increase physical activity among chronically ill children with cystic fibrosis. Future studies are needed to assess maintenance and possible health benefits.

In patients with cystic fibrosis (CF), an excess production of thick, viscous mucus leads to obstruction, fluid retention, infection, and deterioration of function in the pancreas and other organs (30) but chronic pulmonary disease accounts for 97% of mortality (29). Consequently, treatment includes clearing mucus from the lungs through chest physiotherapy (CPT), a tedious, time-consuming process with poor compliance (12, 21, 28, 33).

Exercise is an effective alternative or adjunct to CPT (27). It increases sputum production and clearance from the lungs (20, 30, 45), as well as exercise endurance, ventilatory muscle endurance, pulmonary function (3, 10, 27, 40), immune system function (22, 44), and cardiopulmonary fitness, a possible predictor of survival in CF (24, 39). Compliance is better with exercise than with CPT (1, 3, 6), but establishing an exercise routine remains a challenge.

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The principles of social learning theory (4) can be used effectively in changing behavior. Briefly, this theory is based on a reciprocal determinism: A behavior is strengthened or weakened through models and consequences. For example, physical activity can be increased by providing salient, positive consequences immediately contingent on the performance of activity. Activity goals are shaped, that is, increased, at a rate sufficiently gradual to ensure success. Then external reinforcement is faded as exercise becomes routine and associated with naturally-occurring and maintenance-enhancing reinforcers, such as performance accomplishment, enjoyment, and social interaction.

Behavioral programs have been used previously to increase adherence to an exercise regimen in children with serious medical problems. One study with hemophiliacs (15) was not particularly successful in that activity level decreased by 9-month follow-up. This decline may have been due to a flaw in the behavioral design of the program. Specified exercise was performed and monitored at home, but parental involvement was minimal, and most behavioral reinforcement occurred noncontingently at weekly hospital meetings.

Better behavioral procedures were followed during an activity program for 9- to 12-year-old children who were healthy but unfit (43). Activity was individually tailored, home-based, and parents provided salient rewards immediately contingent on performance. The result was a doubling of after-school activity, and three children maintained their activity levels for at least 12 weeks.

Behavioral procedures have been used for children with CF to increase caloric intake and weight (41, 42). Also, there have been home-based exercise programs for CF patients, which did not have behavioral components and either dictated the exercise regimen (3, 6, 10) or suffered from poor compliance (17). To date a behavioral program has not been used to increase the physical activity of children with CF. Given frequent illness episodes, determining whether a home-based exercise program can increase physical activity is especially important.

Methods

This study used three replications of a multiple baseline design (5) to examine the effects of an individually tailored, home-based, parent-managed, contingent privilege system on the routine physical activity of children with CF. The intervention was initiated in a staggered manner across children after a relatively stable pattern of activity was observed. Consequently, individual baseline periods varied in length. Studies I and II were conducted concurrently, and the interventions lasted 4 weeks. In Study II, an additional 7-day activity record was collected at 6-week follow-up. Study III replicated the previous studies, except that it also evaluated physiological correlates of physical activity and continued for 6 weeks.

Participants

While CF patients of all ages may benefit from increased physical activity, an exercise habit established during childhood may slow progression of the disease, improve ability to engage in activities of daily living (including social development), and encourage self-care. Furthermore, children may be more amenable to changing (19) and maintaining (13) exercise habits.

Ten 7- to 14-year-old medical center patients with CF and their families participated in one of three studies. The children met the following selection crite-
ria: (a) age 7 to 15 years; (b) disease of moderate severity (FEV₁ > 40% predicted, no extensive bronchiectases, esophageal varices, or pneumatoceles); (c) stability of disease for at least 4 weeks; and (d) physician approval for participation in an exercise program.

Written consent was obtained from children and parents for all components of the protocol, and the studies were approved by the Committee on Investigations Involving Human Subjects at the University of California, San Diego and by the Committee on Protection of Human Subjects, San Diego State University.

**Procedures**

Procedural consultations and record reviews were conducted in family homes during 15- to 30-min weekly (Studies I and II) or biweekly (Study III) sessions. Baseline procedures were limited to recording the children’s usual activity. The shaping procedure began at this point in Study III, as children received stickers for reporting physical activities to their parents. A full week’s compliance was reinforced with posters, fast food coupons, toys, or sports items.

In preparation for the intervention, children, parents, and an investigator selected moderate to vigorous physical activities that would be enjoyable and appropriate for the children’s ages, abilities, and family environments. These activities included bicycling, jumping rope, skating, skateboarding, swimming, soccer, running games, and walking.

Healthy children need at least 20 to 30 minutes per day of enjoyable, large muscle-based, moderate to vigorous activities (16, 35), but children with CF may need as much as 60 min of daily aerobic exercise to clear mucus from the lungs (37). Although it is safe for children with CF to increase physical activity (27, 37), disease symptoms often result in inactivity (36) and deconditioning (26, 37). Consequently, an exercise program for unfit children with CF should begin at the current level of activity and increase gradually (9). Activity increases were achieved by shaping activity levels from baseline of the previous week. Goals for activity duration and/or intensity were increased conservatively by 10 percent per week or an amount appropriate for each child’s health and confidence in meeting goals.

Consistent, rather than intermittent, activity performance was encouraged by providing reinforcement for achieving 2- to 3-day “period” goals (Studies I and II) or by permitting no more than twice the daily goal to be applied to the weekly goal (Study III). A “token economy” was employed in Studies I and II. That is, immediately contingent on reaching activity goals, “activity dollars” were awarded for later use in purchasing privileges selected by children and parents. In Study III, privileges were provided directly after activity was performed.

Privileges awarded for meeting “period” (Studies I and II) or daily (Study III) goals varied according to each child’s interests and each family’s disposition, but usually they were domestic in nature: staying up later at night, special food, telephone calls. For achieving weekly goals, privileges often were obtained outside the home: sleep-overs, community outings, a restaurant meal, a $5 toy, a children’s exercise videotape. A progress chart was posted prominently in the home both as visual reinforcement and as a cue to action. Additionally, parents were encouraged to use differential attention, the technique of praising a desired behavior in specific terms and ignoring the lack of (or undesirable) behavior.
Self-Reported Activity Measurement

The primary dependent variable was the number of after school (Studies I and II) or full day (Study III) Activity Points (AP) reported by each child. This was a subjective measure based on each child’s perception of energy expended. However, performing baseline progressive maximal exercise tests enhanced the Study III children’s ability to evaluate exercise intensity. APs were calculated by multiplying the number of minutes spent exercising by an intensity factor (see Table 1). This simplified measure was designed to facilitate understanding and use by the children.

Together, children and parents recorded active events during the day and reviewed records at night. Reliable, valid records of children’s activity have been obtained through interval recording (7) or 1-day activity recalls (31, 34). Specific, time limited behavior reported by 10- to 15-year-old children and their parents has resulted in correlations of .99 for type of exercise, .96 for duration, and .72 for exercise frequency (18).

Accuracy of self-report was enhanced through the “bogus pipeline” effect (14, 23) of the Caltrac electron activity meters worn by the children in sealed waist-pouches during waking hours. Biweekly readings were taken from the monitors, leading participants (and, initially, the investigators) to believe that self-reported activity could be objectively validated. The meters proved to be ineffective, because they were designed to monitor the duration of moderately intense movement in the vertical plane, and these children tended to engage in activities involving nonvertical acceleration at varying levels of intensity.

Fitness Measurement

In Study III, pre- and postintervention maximal exercise tests were conducted and reported as percent change. Each child exercised on an electrically braked Lode cycle ergometer for 3 min at an initial work load of 20 watts (W). As determined by individual heart rate increase, the workload was gradually increased to exhaustion. The last workload at which a participant pedaled for a full minute was considered the maximal workload (Wmax). Heart rate (HR) was recorded throughout the testing, and pulse oximetry was used to assure that oxygen saturation (SaO₂) did not fall below 85%. Expired gases were collected during maximal exertion with open

<table>
<thead>
<tr>
<th>Intensity</th>
<th>Activity description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Slow: Activities that do not need much effort and can be sustained for a long time (e.g., walking, ping-pong, bowling)</td>
</tr>
<tr>
<td>2</td>
<td>Medium: Activities that make the heart beat faster than at rest but can be sustained for awhile (e.g., fast-walking, bicycling, skating)</td>
</tr>
<tr>
<td>3</td>
<td>Fast: Activities that make the heart beat fast, cause hard breathing and sweating (e.g., jogging, active basketball, soccer)</td>
</tr>
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</table>
circuit spirometry. Maximal oxygen uptake (VO$_{2\max}$), carbon-dioxide production (VCO$_2$) and minute ventilation (V$_E$) were recorded.

**Analysis**

Unless relative stability was achieved more quickly, means and standard deviations are presented for the number of daily APs achieved during the last month of baseline. These conservative estimates of typical activity levels are compared with the APs achieved during the last 7 days of the intervention, which is the week during which activity goals had been gradually shaped to their highest levels. Evaluation is based on percent change.

**Results**

**Reported Activity**

After the interventions were initiated, 8 of the 10 children began increasing their activity, achieving 25% to 100% of weekly activity goals. Figure 1 shows the time series record of activity points achieved for children A-D, as illustrations of the within- and cross-subject variability. This shows that children differed in level and within-person variability. They also differed in the illness episodes leading to less continuous data in some cases. It is important to note for those children who experienced an illness during baseline that activity performed following the illness tended to be of a lower level than activity prior to the illness. Decisions to initiate training were made based on performance following illness episodes. Results also show that the level of exercise tended to increase, and the variability in exercise decreased with the start of the contingency program for each child. During the last week of the intervention, their reported APs were 43% to 321% over baseline (see Table 2). The 6- to 8-week follow-up of Study II recorded the APs of Children E, F, and G to be 221.9%, 102.5%, and 7.4% above baseline.

**Fitness**

At posttest, Children H, J, and K increased VO$_{2\max}$ 27%, 20%, and 7% above the pretest levels (see Table 3). Children H and J experienced increases in V$_E$ of 8% and 31.6% with accompanying increases in workload of 20% and 14.2%. There was little change in V$_E$ and no change in Wmax for Child K. SaO$_2$ levels at maximum effort were 86% to 99%, representing decreases of 1% to 8% from resting.

All children expended maximal effort during pre- and posttests, as indicated by respiratory exchange ratios ≥ 1.0. Child H also achieved 90% of his age-predicted maximal heart rate, which is unusual for children with CF, because they often reach exhaustion first (25). Although the posttest indicated that J and K’s maximal heart rates were 85% and 83% of predicted, those rates were achieved at workloads that were higher (J) or the same (K) as workloads during the pretest.

**Discussion**

Previous research suggests that physical activity improves CF symptoms, but significant physiological effects can be achieved only through long-term, consistent
behavior. Establishing a physical activity routine takes time, especially when dealing with unfit, chronically ill children and their concerned families.

In this study the children’s physical activity habits changed after the home-based, parent-managed, contingent privilege intervention was initiated. This pattern was replicated in each of the three multiple baseline designs, powerful evidence of the intervention’s effectiveness.
Table 2  Studies I, II, and III: Daily Activity Points

<table>
<thead>
<tr>
<th>Child</th>
<th>Study I</th>
<th>Study II</th>
<th>Study III</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>A</td>
<td>B</td>
<td>C</td>
</tr>
<tr>
<td>Age (yr)</td>
<td>8</td>
<td>9</td>
<td>10</td>
</tr>
<tr>
<td>Gender</td>
<td>M</td>
<td>M</td>
<td>M</td>
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</table>

Baseline

<table>
<thead>
<tr>
<th>Days</th>
<th>Study I</th>
<th></th>
<th>Study II</th>
<th></th>
<th>Study III</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>(SD)</td>
<td>M</td>
<td>(SD)</td>
<td>M</td>
</tr>
<tr>
<td>28</td>
<td>113.4</td>
<td>(107.2)</td>
<td>77.0</td>
<td>(59.2)</td>
<td>65.6</td>
</tr>
<tr>
<td>21</td>
<td>57.1</td>
<td>(94.1)</td>
<td>68.8</td>
<td>(55.4)</td>
<td>115</td>
</tr>
<tr>
<td>28</td>
<td>154.8</td>
<td>(112.3)</td>
<td>128.9</td>
<td>(90.4)</td>
<td>125</td>
</tr>
<tr>
<td>28</td>
<td>158.2</td>
<td>(137.5)</td>
<td></td>
<td></td>
<td></td>
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</tbody>
</table>

End of intervention

<table>
<thead>
<tr>
<th>Days</th>
<th>Study I</th>
<th></th>
<th>Study II</th>
<th></th>
<th>Study III</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>(SD)</td>
<td>M</td>
<td>(SD)</td>
<td>M</td>
</tr>
<tr>
<td>7</td>
<td>221.7</td>
<td>(102.3)</td>
<td>324.3</td>
<td>(131.9)</td>
<td>156</td>
</tr>
<tr>
<td>7</td>
<td>81.4</td>
<td>(30.2)</td>
<td>126.4</td>
<td>(49.4)</td>
<td>215.7</td>
</tr>
<tr>
<td>7</td>
<td>356.4</td>
<td>(93.5)</td>
<td>95</td>
<td>(112.2)</td>
<td>128.6</td>
</tr>
<tr>
<td>7</td>
<td>248.6</td>
<td>(146.4)</td>
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Change

<table>
<thead>
<tr>
<th>Study I</th>
<th>Study II</th>
<th>Study III</th>
</tr>
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<tbody>
<tr>
<td>+ 95.6%</td>
<td>+ 42.5%</td>
<td>+ 130.2%</td>
</tr>
<tr>
<td>+ 57.1%</td>
<td>+ 321%</td>
<td>- 26%</td>
</tr>
<tr>
<td>+ 138%</td>
<td>+ 82%</td>
<td>+ 2</td>
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</table>

$M$ change = 92.6% (95.4)
### Table 3  Study III: Fitness Measures, Pre- and Postintervention

<table>
<thead>
<tr>
<th>Participants</th>
<th>Age (yr)</th>
<th>Height (cm)</th>
<th>Weight (kg)</th>
<th>VO$_{2}$max (ml/kg/kg)</th>
<th>V$_{E}$max (L/min)</th>
<th>Wmax (watts)</th>
<th>HRmax (bpm)</th>
<th>SaO$_{2}$ (Resting to Wmax)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child H</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre</td>
<td>14</td>
<td>151</td>
<td>40.6</td>
<td>36.4</td>
<td>62</td>
<td>100</td>
<td>197</td>
<td>92–88%</td>
</tr>
<tr>
<td>Post</td>
<td>152</td>
<td>40.1</td>
<td>46.1</td>
<td>67</td>
<td>120</td>
<td>197</td>
<td>NC</td>
<td></td>
</tr>
<tr>
<td>Change</td>
<td></td>
<td></td>
<td>+ 27%</td>
<td>+ 8%</td>
<td>+ 20%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Child J</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre</td>
<td>10</td>
<td>138</td>
<td>29.1</td>
<td>35.0</td>
<td>38</td>
<td>70</td>
<td>181</td>
<td>97–94%</td>
</tr>
<tr>
<td>Post</td>
<td>138.5</td>
<td>30.2</td>
<td>42.1</td>
<td>50</td>
<td>80</td>
<td>178</td>
<td>NC</td>
<td></td>
</tr>
<tr>
<td>Change</td>
<td></td>
<td></td>
<td>+ 20%</td>
<td>+ 31.6%</td>
<td>+ 14.2%</td>
<td>- 1.7%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Child K</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre</td>
<td>10</td>
<td>123.5</td>
<td>25.0</td>
<td>30.8</td>
<td>34</td>
<td>50</td>
<td>169</td>
<td>95–99%</td>
</tr>
<tr>
<td>Post</td>
<td>124.5</td>
<td>25.7</td>
<td>33.0</td>
<td>33</td>
<td>50</td>
<td>174</td>
<td>NC</td>
<td>+ 3%</td>
</tr>
<tr>
<td>Change</td>
<td></td>
<td></td>
<td>+ 7%</td>
<td>- 3%</td>
<td></td>
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</table>

Note. VO$_{2}$max = maximal oxygen uptake; V$_{E}$max = maximal minute ventilation; Wmax = maximal workload; HRmax = maximal heart rate; SaO$_{2}$ = oxygen saturation.
When the intervention was terminated, 8 of the 10 children had increased the intensity, duration, or both, of their daily physical activity 42.5% to 321% above baseline. The other two children exercised with greater consistency, a healthy prerequisite for increased intensity and duration. The magnitude of change was affected by baseline levels of activity, ability, and interest; the confidence families had in increasing activity goals of unfit, chronically ill children; the degree to which progress was delayed by the periodic exacerbation of illness characteristic of chronic disease; and the extent to which families followed the behavioral protocol.

The following descriptions of participants’ experiences with the protocol demonstrate the importance of flexibility and individual tailoring when conducting a physical activity program for children with CF. Children A, C, D, and E appeared to enjoy being active, were in good health, and experienced activity increases of 57% to 321% over baseline. Child H’s health was not as good, but, at age 14, he was motivated to improve his level of fitness. He systematically increased his own goals, as planned, by about 10 percent per week. He raised his APs 138% above baseline, and his success encouraged him to undertake lengthy bicycle rides. At his age, it was sufficiently reinforcing to manage and chart his own activity goal achievement. Biweekly support was provided by the investigator, but parental involvement was minimal.

On the other hand, Children B and F were symptomatic, had low baseline activity, and limited interest in physical activity. Their activity increased 42.5% and 83.7% above baseline. The changes resulted from a slow, upward trend in both level and stability of activity, interrupted in the case of Child F by illness and hospitalization. By extending the measurement period from 4 weeks to 3 months and tailoring the program to Child F’s circumstances, his APs were 102.5% over baseline at the 6-week follow-up.

Children J and K also were not interested in becoming more active. Furthermore, their mothers had difficulty using differential attention, and privileges were awarded inconsistently, often allowing a time lapse between activity performance and reinforcement. Child J experienced episodes of illness that prevented activity, but he eventually raised his APs to 82% above baseline. Child K’s APs only increased 2%, but there were other important changes in her habits. The number of days on which Child K engaged in after school activity increased from 50% during baseline to 81% during the intervention. The median number of after school APs increased from zero to 70, while the mean increased 39%.

Initially, Child G elected to target one high intensity activity (running a homemade obstacle course). After 2 weeks, the duration of activity had increased 75% above baseline. Simultaneously, there was a precipitous drop in the medium and low intensity activities that ordinarily were performed, resulting in an overall decrease in APs. When goals were altered to include a variety of medium and low intensity activities, APs gradually increased to 26% below baseline at 4 weeks, and a behavioral reversal was observed; that is, when it was no longer specifically reinforced, the high intensity activity decreased. By 6-week follow-up, APs had increased 7.4% above baseline.

All three children (E, F, and G) in Study II reported increased activity levels at 6-week follow-up. This reflects the maintenance-enhancing value of modifying ordinary activities within routine lifestyles and developing environmental support.

In Study III, maximal exercise testing with Children H, J, and K validated self-reported activity data. APs were associated with a similarly ordered magnitude of
change in VO\textsubscript{max} and W\textsubscript{max}. In two of the three maximal exercise tests, the increases in VO\textsubscript{max} and workload suggest that even relatively modest activity changes of limited duration result in improved exercise tolerance and cardiopulmonary fitness, an important correlate of survival. The third child’s lack of significant change in all exercise test measures reflects the minimal increase in her APs.

The VO\textsubscript{max} attained by Children H and J was > 82% predicted, a level associated with lengthened survival in CF (24). These two children also increased V\textsubscript{E}\textsubscript{max}, a value related to the total amount of work accomplished. The increases in VO\textsubscript{max} and V\textsubscript{E}\textsubscript{max} were similar to or greater than previously reported (8, 10, 11, 26, 38, 39), though they were achieved at lower workloads than found in other studies of children with moderately severe CF (11, 38).

Based on the results of this study, it appears that the best candidates for a similar program would be children who enjoy being active and are in relatively good health. While children as old as 14 may be self-motivated, the success of increasing the physical activity of younger children depends on parental support (2, 32).

Alternatively, support of a behavioral protocol may be enhanced by training parents in group sessions like those effectively conducted during similarly designed nutrition programs for children with CF (41, 42). Such sessions would provide social reinforcement for following the protocol and an opportunity to discuss effective means of integrating new behaviors with current lifestyles. Additionally, the setting would be conducive to behavioral rehearsal of techniques such as differential attention.

The interpretation of the results of this study is provisional due to the small number of participants and the intervention’s limited duration. For children with chronic disease and days of debilitating illness, a longer intervention period may be needed to more firmly establish behavior change. Some of the children in this study may have been able to meet greater challenges, but these procedures provided successful experiences and positive associations with physical activity. Over time, further increases in activity duration and intensity may improve clinical symptoms and disease outcome. Long term follow-up would better evaluate this potential. Present results suggest strongly that an individually tailored contingency program, managed by parents or by a behavioral specialist, can increase physical activity among children with CF. These findings support larger clinical trials.

References


