Exercise Rehabilitation Programs for Children With Congenital Heart Disease: A Note of Caution

David J. Driscoll

Supervised exercise programs have been an important aspect of the rehabilitation process of adult patients with coronary artery disease, especially following myocardial infarction and/or coronary artery surgery (9, 10). The goals of such programs include (a) improvement of cardiovascular fitness and exercise tolerance; (b) amelioration of risk factors for coronary artery disease such as obesity, hyperglycemia, hypertension, and an adverse lipid profile; and (c) reduction of the risk of recurrent myocardial infarction and sudden death. There are insufficient scientific data available to indicate that all these goals are achievable. Certainly exercise efficiency and aerobic capacity can be increased for many patients. A reasonable exercise program, combined with appropriate dietary discretion, can reduce obesity and in turn reduce low-density lipoprotein cholesterol and perhaps increase high-density lipoprotein cholesterol. Data to suggest that an exercise rehabilitation program in and of itself can reduce the risk of recurrent myocardial infarction or sudden death are tenable at best.

Pediatric Rehabilitation Programs

There has been interest in developing exercise rehabilitation programs for children and adolescents who have had operation for congenital heart defects. Although development of these programs to some extent parallel those for adult patients with coronary artery disease, it must be recognized that the patient populations are different, the disease processes are dissimilar, and the indication for and goals for rehabilitation may be quite different. A goal of rehabilitation of adult patients is to alter the progression of coronary artery disease, an acquired disease. Obviously, the natural history of congenital cardiac defects is less likely to be affected by an exercise rehabilitation program.

If one assumes that all children who have had a cardiac operation should participate in an exercise rehabilitation program, several questions must be answered to determine whether this assumption is reasonable, scientifically sound, and cost-effective:

David J. Driscoll, M.D., is Head of the Section of Pediatric Cardiology at the Mayo Clinic, Mayo Medical School, Rochester, MN 55905.
1. Is the exercise ability/capacity of children who have had operations for congenital heart disease abnormal? Obviously the answer depends on the complexity of the congenital heart defect, the operative repair, and the operative result. In a study of 118 adult patients with pulmonary stenosis who had had pulmonary valvotomy as children or adolescents, aerobic capacity was reduced from normal but was 94% of normal. Similarly, for 74 patients who had had operation for aortic stenosis, aerobic capacity was 87% of normal. For 113 patients who had had operation to close a ventricular septal defect, aerobic capacity was 90% of predicted. Although for all three of these relatively simple congenital cardiac defects the aerobic capacity was less than published normals, it was near normal and did not contribute to long-term morbidity (4).

Patients with more complex forms of congenital heart disease, however, may have more marked reduction in aerobic capacity postoperatively. Maximum oxygen consumption (VO₂max) after repair of tetralogy of Fallot is reduced; in two studies it was 70 and 72% of predicted (7, 12). In a group of 10 patients after complete repair of pulmonary atresia and ventricular septal defect, VO₂max was 51% of predicted normal (1). Ensing et al. (5) reported that VO₂max was reduced to 59 ± 18% of predicted for 19 patients who had had repair of transposition of the great arteries. For 11 patients who had had repair of Epstein’s anomaly, VO₂max was 71.6 ± 18% of predicted (3). After the modified Fontan operation, 16 patients with tricuspid atresia and 9 patients with univentricular heart had VO₂max reduced to 48 ± 13 and 56 ± 15% of normal, respectively (2).

Although there is considerable difference in aerobic capacity of patients with repaired congenital heart defects, as a group their exercise tolerance is reduced. The reasons for this reduction are multifactorial and include chronotropic insufficiency, deconditioning that results from a sedentary life-style dictated by the severity of the cardiac defect or that is either self-imposed or imposed by health providers, reduced leg muscle mass from chronic illness, reduced ventricular function, respiratory insufficiency, and residual cardiac defects.

2. Can aerobic capacity be increased in children with congenital heart defects using an organized, supervised rehabilitation program? Certainly if deconditioning is a contributing factor to reduced aerobic capacity in children with congenital heart defects, aerobic capacity should increase if fitness is improved. Workers in the field intuitively believe this. Scientifically proving this point, however, has been more difficult. In addition, one must distinguish between increased aerobic capacity (i.e., increased VO₂max) and increased exercise efficiency (i.e., performance of work at a relatively lower submaximal VO₂ or heart rate). Ruttenberg et al. (11) reported 24 patients who had had repair of congenital heart defects and 26 age-matched controls. These subjects participated in a 9-week supervised exercise rehabilitation program. Of note, only 12 of the 27 patients and 9 of the 25 control subjects completed the entire program. Both the patient and control groups achieved a higher stage (p<0.01) of exercise at the end of the program. VO₂max increased but not significantly. Indeed, the increase in VO₂max paralleled an increase in heart rate, suggesting that the subject’s degree of effort was greater during the follow-up study than during the original study. This investigation may demonstrate an important improvement in exercise efficiency but does not demonstrate an improvement in aerobic capacity. Also, since more than half of the original participants did not complete the study, a significant ascertainment bias may have been introduced.
In a study of 26 patients who had had repair of tetralogy of Fallot or ventricular septal defect, Goldberg et al. (6) reported a significant increase in maximum working capacity following a 6-week rehabilitation program. However, VO$_2$-max did not change. Again, this indicates an improvement in exercise efficiency but not an increase in aerobic capacity.

Longmiur et al. (8) studied 60 patients after cardiac operation. The experimental group comprised 29 patients who lived near the medical center while the control group was composed of 31 patients who lived farther away. Tests were conducted to evaluate cardiovascular endurance, strength, flexibility, and coordination. A test score for these indices was determined before and after a supervised fitness rehabilitation program. In addition, test scores were determined 6 months after operation. These investigators found that the experimental group had to be subdivided into a subset who complied with the exercise program and a subset who did not. At the end of final testing, the compliant group scored higher than the noncompliant group and higher than the control group. However, the control group had not been subdivided into those who voluntarily did regular exercise and those who did not. Therefore one must question whether the final test scores reflected changes caused by the rehabilitation program or merely reflect that some patients are motivated to exercise and others are not.

The Need for Future Studies

Most investigators believe that aerobic capacity and/or exercise efficacy can be improved with an organized fitness program in children, whether or not they have heart disease and whether or not cardiac operation has been performed. The answers to several questions are unclear: (a) Are the changes of aerobic capacity and exercise efficiency that are induced by an organized exercise rehabilitation program persistent beyond the immediate period of the program? (b) Does the program change the subject's life-style and approach to exercise and fitness? (c) Are the changes in aerobic capacity and exercise efficiency a result of the exercise rehabilitation program or merely reflective of the participant selection and dropout bias?

The study to answer these important questions is complex and, to my knowledge, has not been done. The design is illustrated in Figure 1. It involves three groups of patients with repaired congenital heart defects. Obviously, the groups must be comparable in age, gender, type of congenital heart defect, and type of surgical repair. In addition, attrition of subjects from the study must be zero or near zero. Aerobic capacity and exercise efficiency are measured at the beginning of the study. This should be done at least twice to reduce the effect of acclimatization to the testing procedure which could result in a test/retest phenomenon. One group is enrolled in an organized supervised exercise program for 6 to 18 weeks; the second group is given, on one occasion, verbal instruction regarding an exercise fitness program; the third group is given no information or counseling. Aerobic capacity and exercise efficiency are again measured for all subjects 3 weeks, 3 months, and 12 months later. The major difficulty in performing this study is preventing the attrition of subjects, which is critical for removing the confounding variable of motivated and unmotivated subjects.

In addition to the above posed questions that this study would answer, there are important implications to those answers. Obviously, supervised exercise fitness
programs involve considerable manpower and expense. If these programs do not effect a sustained change in aerobic power, exercise efficiency, or quality of life, it will be difficult to justify the cost. If they effect change only in a select population but not in the majority of patients, this select population should be identified and, if indicated, entered into such a program. Appropriate but selective application of these programs would reduce the overall cost to society.

**Goals of a Pediatric Rehabilitation Program**

It is important to define the goals of a program and then determine whether those goals are met. The following would be reasonable goals for a pediatric exercise rehabilitation program:

1. To improve the quality of life: Because it is difficult to define "quality of life," it is difficult to determine important changes in the quality of life. There is strong evidence that patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect have a reasonably normal quality of life (1). Patients with other relatively simple forms of congenital heart disease (atrial septal defect, patent ductus arteriosus, coarctation of the aorta) also return to a reasonably normal quality of life after operation. Indeed, most patients with uncomplicated forms of congenital heart disease such as endocardial cushion defect and tetralogy of Fallot do well after operation.

   There indeed are some children who are so debilitated by their disease, and perhaps the consequences of operation, that a supervised exercise program may be an important adjunct to returning them to a more active life-style. Compared to all children with congenital heart defects and those having cardiac operation, this is a relatively small albeit important group. It is reasonable to target this group of patients to study the effects of exercise rehabilitation programs. However, it is probably unnecessary and not cost-effective to utilize organized
rehabilitation programs to effect changes in the quality of life for all children with congenital heart defects.

2. To reduce the incidence of sudden death in children with congenital heart disease: Although there is some evidence to suggest that an exercise rehabilitation program reduces the risk of sudden death after myocardial infarction in adults, there is no evidence that such a program alters the risk of sudden death for children with congenital heart defects, either surgically repaired or not.

3. To reduce the incidence and consequences of coronary artery disease:
The prevention of coronary artery disease is an important goal and should begin in childhood. Although this goal is perhaps less pertinent to the rehabilitation of children with congenital heart disease than is the maintenance of a healthy lifestyle for all children, it is the goal that, in adults, has been studied most thoroughly.

A sedentary life-style is a risk factor for the development of coronary artery disease. It is unclear whether this association is direct or indirect and linked by the effect of exercise on obesity, HDL cholesterol, carbohydrate metabolism, dietary habits, or lipid transport. Because the etiology of coronary artery disease is multifactorial, any program designed to reduce the incidence of coronary artery disease must be designed to eliminate or reduce all the risk factors, not only a sedentary life-style.

Summary

1. Using a supervised exercise rehabilitation program, exercise efficiency and perhaps aerobic capacity can be improved in children who have had operation for congenital heart defects. However, it is unclear whether these changes would have occurred in the absence of a supervised exercise rehabilitation program.

2. Most children return to a reasonably normal life-style and level of physical fitness after repair of congenital heart defects.

3. Supervised exercise rehabilitation programs may be useful to return selected children to an improved level of aerobic capacity and exercise efficiency.

4. There is insufficient information available at present to justify the cost of a supervised exercise rehabilitation program for all children after cardiac surgery.

5. Further studies are necessary in order to clearly define the medical efficacy, ideal population to target, and cost effectiveness of supervised exercise rehabilitation programs for children with congenital heart defects.

References


