

# Exercise in Congenital Heart Disease: A Contemporary Review of the Literature

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## Abstract

Congenital heart disease (CHD) patients tend to exercise less than their peers without heart disease. These patients often have exercise hesitancy or are not appropriately encouraged to exercise by their primary cardiologists. Regular exercise in the CHD population has been shown to increase fitness and lessen the incidence of obesity. There are positive psychosocial benefits of regular exercise including decreased mood disorders, improved academic performance, and ultimately improved quality of life. As physical activity is increasingly emphasized, a purposeful approach utilizing cardiopulmonary exercise testing for risk stratification and establishment of a baseline level of fitness is often helpful. Once exercise testing is complete, cardiopulmonary rehabilitation has been shown to improve measures of physical and mental fitness and should be considered in CHD patients in need of assistance with exercise.

**Keywords:** Congenital heart disease, exercise capacity, exercise safety

## INTRODUCTION

Survivors of congenital heart disease (CHD) are a rapidly increasing proportion of the population secondary to the improvements in surgical and medical management.<sup>[1]</sup> As these patients age and live with a chronic disease, there has been increased focus into not only survival but also quality of life through aging. In evaluating quality of life, multiple organ systems can be affected in CHD patients including cardiac, pulmonary, musculoskeletal, and neuropsychiatric, all of which exercise could potentially improve. Previously, there has been significant hesitation in allowing these patients to exercise secondary to concerns for sudden cardiac death or worsening their underlying abnormal cardiac physiology.<sup>[2]</sup> While CHD patients are often higher risk compared to the general population, there is a growing understanding that the overall physical and mental benefits to regular exercise may be greater than the low risk of exercise-induced adverse events in these patients. The main purpose of this article is to review the current research into the risks and benefits of regular physical activity with extra emphasis on the psychosocial benefits and to discuss how structured exercise testing and rehabilitation programs can be beneficial in CHD patients.

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## EXERCISE SAFETY IN CONGENITAL HEART DISEASE PATIENTS

Sedentary behavior and poor cardiopulmonary fitness is a major problem in the CHD population. As these patients age, they become at risk for acquiring atherosclerotic heart disease and obesity in addition to derangements with their underlying complex cardiac physiology and postsurgical complications.<sup>[3,4]</sup> Adults with congenital heart defects ranging from simple to complex have varying levels of exercise intolerance.<sup>[5]</sup> While the reason for this is likely multifactorial, the likely factors include physiologic limitations and excessive self-protection is likely another major factor.<sup>[2]</sup> Some of these extreme hesitations to exercise are related to the fear and anxiety that often accompany living with a chronic disease. Another etiology for this fear of exercise is the by-product of previous activity limitations placed by physicians secondary to fears of the possible proarrhythmic potential of exercise.<sup>[6]</sup> While the paradigm is shifting toward increasing physical activity in CHD patients, there remains hesitancy among some patients

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and their physicians regarding the safety and efficacy of regular exercise.

There is a growing body of evidence that exercise is safe for the majority of patients with CHD. Multiple studies have demonstrated that sudden death in the CHD population rarely occurs during exercise and instead tends to happen at rest.<sup>[7-9]</sup> Koyak *et al.* demonstrated in a large multicenter study that in 213 sudden deaths, only 10% of these occurred during exercise with the remainder occurring during rest or sleep.<sup>[7]</sup> Silka *et al.* evaluated results in the state of Oregon over a decade and found that only 15% of the 41 sudden deaths occurred during exercise.<sup>[8]</sup> These findings were further reinforced in a long-term study of patients who underwent aortic valvuloplasty for congenital aortic stenosis. In this study by Brown *et al.*, sudden death with exercise was rare and exercise restriction was not found to convey any protective benefit.<sup>[10]</sup> A similar risk profile was seen when evaluating complex CHD patients who underwent formal exercise testing in a hospital setting, which is noteworthy in that during exercise testing, these patients are often pushed to levels of exhaustion rarely reached during recreational activities. The Boston group when evaluating their cardiopulmonary exercise testing outcomes over a 3-year period showed a very low rate of adverse events requiring either test termination (0.6%) or medical intervention (0.06%) despite testing a large amount of high-risk patients.<sup>[11]</sup> Of note, the low rate of events also speaks to the importance of following the previously reported absolute and relative contraindications to cardiopulmonary exercise testing including not testing patients with active inflammation, severe systemic hypertension, or physical limitations to exercise.<sup>[12]</sup>

## EXERCISE BENEFITS IN CONGENITAL HEART DISEASE PATIENTS

While there is minimal evidence that exercise is overtly dangerous for the majority of CHD patients, there is a large body of evidence that it can be quite beneficial. The physical health benefits of exercise are important and can include obesity prevention/weight management,<sup>[2]</sup> improved heart function,<sup>[13]</sup> and increased musculoskeletal strength and aerobic fitness.<sup>[14,15]</sup> Perhaps less discussed in these patients is the potential for significant psychosocial benefits to regular exercise. The potential benefits of exercise on mood in the non-CHD population have been well studied.<sup>[16,17]</sup> The possible mechanisms for this include increased hippocampal growth,<sup>[18]</sup> improved circulation of the hypothalamic–pituitary–adrenal axis,<sup>[19]</sup> increased neurochemical receptor/hormone expression,<sup>[20]</sup> and improved cardiopulmonary endurance, with all of these highlighting the integration and involvement of multiple organ systems leading to these complex issues. On reviewing studies evaluating the exercise benefits in non-CHD patients, a large meta-analysis in noncardiac patients with depression by Kvam *et al.* showed a moderate-to-large effect on improving the symptoms of depression compared to controls, and this effect remained when exercise was

used as an adjunct therapy with antidepressants.<sup>[16]</sup> Fox *et al.* were able to demonstrate that regular exercise in high school students increased self-esteem, social integration, and academic performance.<sup>[21]</sup> In addition, exercise and youth sports participation has been shown to be a protective factor against depression and suicidal ideation in adolescents.<sup>[22]</sup> The high rate of these comorbidities in adolescent and adult CHD patients is well documented,<sup>[23,24]</sup> so any potentially modifiable factor in preventing these often overlooked but serious issues is noteworthy.

Regular exercise has been shown to modify some of these psychosocial concerns and improves the overall quality of life in patients with CHD.<sup>[25,26]</sup> Dean *et al.* evaluated responses from 174 young patients with CHD and found that ~75% of them participated in organized sports and that their participation was associated with higher quality of life, improved exercise capacity, and lower body mass index.<sup>[25]</sup> Opić *et al.* were able to show that while Dutch adults with CHD were less involved in sports than their unaffected peers, those who were active showed a higher exercise function without having a larger rate of adverse events compared to CHD patients who did not participate in sports.<sup>[26]</sup> Fredriksen *et al.* have demonstrated similar findings in that active engagement of regular physical activity in pediatric patients with moderate/severe CHD resulted in improved quality of life indexes, body mass index, and aerobic capacity.<sup>[13]</sup> Sandberg *et al.* also demonstrated similar findings in an adult population. Through evaluating 315 patients from the Swedish Registry, they were able to demonstrate that physical exercise >3 h a week was associated with the positive health status and quality of life in adult CHD survivors.<sup>[27]</sup>

## STANDARDIZED EXERCISE PROGRAMS IN CONGENITAL HEART DISEASE PATIENTS

As impaired fitness and poor exercise habits in the CHD population are common, an individualized approach to exercise training in these patients is exceedingly important. An important component of the initial assessment of CHD patients is the cardiopulmonary exercise test, which is a valuable tool to gain information on a patient's cardiopulmonary function and aerobic fitness and to determine how the cardiac and pulmonary systems respond to the stress of exercise.<sup>[28]</sup> In the cardiopulmonary exercise laboratory, the test is performed to assess functional capacity and aerobic fitness level, determine disease severity, and monitor the effects of therapies and rehabilitation.<sup>[29]</sup> There have been increasing evidence for the prognostic significance of functional capacity assessed during cardiopulmonary exercise testing in CHD patients.<sup>[30,31]</sup> This has become important for the routine management and clinical decision-making for these patients and is a major component of the surveillance for adults with CHD.<sup>[32]</sup> In addition to its use as a clinical tool, cardiopulmonary exercise testing is valuable for establishing a baseline functional capacity prior to initiating an exercise program.

Cardiopulmonary rehabilitation has been the most studied structured fitness program in CHD patients and includes exercise training, nutritional education, and psychological counseling.<sup>[33]</sup> While not uniformly standardized across centers, a structured cardiopulmonary rehabilitation program for CHD patients usually involves several steps, starting with a complete medical evaluation by a cardiologist to ensure to identify potential contraindications to exercise. Following that, a full cardiopulmonary exercise test is performed to identify the potential for dangerous arrhythmias and establish baseline fitness measurements (functional capacity and maximum heart rate and blood pressure). Once the patient is deemed safe to exercise, a structured regimen is begun that usually involves sessions 2–3 times a week averaging 30–60 min a session over 12 weeks. The goal of the session is dependent on the patient and their level of function, but generally, the intensity does not exceed 70%–80% of the peak heart rate.<sup>[34]</sup> Structured cardiopulmonary rehabilitation programs have been shown to improve body composition, endurance time, and musculoskeletal strength in CHD patients.<sup>[35,36]</sup> Wittekind *et al.* have demonstrated an increase in both maximal and submaximal oxygen consumption in Fontan patients who underwent a 12-week program training 60 min a session.<sup>[37]</sup> In addition, Bradley *et al.* have demonstrated similar findings in the tetralogy of Fallot patients utilizing a similar method for cardiopulmonary rehabilitation.<sup>[15]</sup>

In addition to improving cardiopulmonary fitness, there is evidence that cardiopulmonary rehabilitation can also reduce psychosocial stress including anxiety and depression and improve quality of life in patients with cardiac disease although there is a dearth of research specifically evaluating this in the CHD population.<sup>[38,39]</sup> Shepherd and While performed a meta-analysis that demonstrated that participation in a standardized cardiopulmonary rehabilitation program resulted in significantly improved quality of life which in turn improved vocational status in adult patients with heart disease.<sup>[39]</sup> In a non-CHD population, Lavie *et al.* were able to demonstrate a >50% improvement in depression symptoms in adults in their fourth and fifth decades of life with coronary artery disease following completion of cardiopulmonary rehabilitation.<sup>[40]</sup> Anecdotally from our local experience, these patients also socially benefit from the relationships formed between the exercise physiologists over the duration of their program although this has not been extensively studied for any disease to date. Further benefits are seen when family involvement occurs during this process as this has been shown to increase positive perceptions of exercise in CHD patients.<sup>[41]</sup> Despite the multiple benefits of structured cardiopulmonary rehabilitation, it is often underused and underutilized in the care of these patients, allowing this to be a potential area of emphasis in CHD patients.<sup>[42]</sup>

## CONCLUSION

CHD patients are surviving longer into adulthood placing the focus on physical and mental health as they age. While there

has been hesitation by both patients and their providers in the past to allow CHD patients to exercise, there is minimal evidence that exercise is excessively dangerous for the majority of CHD patients and evidence is growing that it can be quite beneficial for most of these patients. This is particularly in relation to their mental well-being, which in turns leads to higher quality of life. As exercise is emphasized more in these patients, particular care should be given to establish an individual approach emphasizing formal cardiopulmonary exercise testing and cardiopulmonary rehabilitation.

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## Conflicts of interest

There are no conflicts of interest.

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