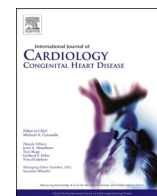




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## Exercise, prescription and training in ACHD

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### ABSTRACT

The number of adult patients with congenital heart disease (CHD) is steadily increasing and exceeds nowadays the number of children with CHD. This is due to significant advances in therapeutic possibilities that became available over the last four decades. As such, this aging population survives the CHD complications and is exposed to the traditional cardiovascular risk factors for atherosclerotic disease such as high blood pressure, elevated cholesterol levels, long-standing exposure to smoking, overweight and obesity, and a sedentary lifestyle. Consequently, it becomes important to put more emphasis on all these risk factors. A preventive strategy is central, and early encouragement of physical activity is part of this approach. A minimum of physical activity has a beneficial effect both physically and mentally. With this overview, we mainly want to emphasize the importance of preventive measures. We would like to emphasize that all individuals should receive an exercise prescription which adheres to the minimum recommendations by WHO/NICE and this advice should form the baseline. Moreover, we intend to show that physical activity can be done safely in patients with CHD and that recreational and competitive sports are feasible in many circumstances.

### 1. Introduction

The number of adult patients with congenital heart disease (CHD) is steadily increasing and exceeds nowadays the number of children with CHD [1]. This is due to significant advances in therapeutic possibilities that became available over the last four decades. As such, this aging population survives the CHD complications [2] and is exposed to the traditional cardiovascular risk factors for atherosclerotic disease such as high blood pressure, elevated cholesterol levels, long-standing exposure to smoking, overweight and obesity, and a sedentary lifestyle. Consequently, it becomes important to put more emphasis on all these risk factors. A preventive strategy is central, and early encouragement of physical activity is part of this approach. A minimum of physical activity has a beneficial effect both physically and mentally [3]. With this overview, we mainly want to emphasize the importance of preventive measures. We would like to emphasize that all individuals should receive an exercise prescription which adheres to the minimum recommendations by WHO/NICE and this advice should form the baseline. Moreover, we intend to show that physical activity can be done safely in patients with CHD and that recreational and competitive sports are feasible in many circumstances.

### 2. Cardiovascular risk factors and adult congenital heart disease

Due to surgical and non-surgical therapeutic advances life expectancy and quality of life of adults with CHD are no longer limited by the initial CHD, but by exposure to acquired cardiovascular risk factors in disease such as myocardial infarction, stroke, peripheral atherosclerosis, and progressive renal function impairment. The traditional cardiovascular risk factors for atherosclerotic disease then also occur (Fig. 1).

Studies have shown that 9–25% of CHD patients are active smokers [4,5]. Despite patients being advised to quit smoking and despite effective smoking cessation programs, smoking remains still prevalent in the CHD population.

The prevalence of systemic hypertension in CHD patients is like this of the general population where it occurs in one third of the cases. According to the underlying lesion, up to 45% of adult patients might develop *systemic hypertension* [6–8]. This is the case in patients with coarctation of the aorta where high blood pressure is more prevalent and appears earlier in life [7,8]. Particularly in these patients, blood pressure remains often high, despite the best efforts using traditional antihypertensive medication.

Lipid levels might be elevated too. The overall prevalence of

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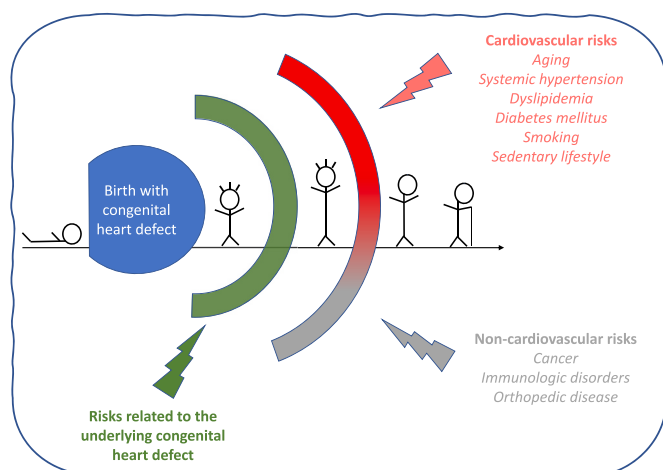


Fig. 1. Lifetime exposure to risk factors in congenital heart disease patients.

hypercholesterolemia increases with age and CHD patients are no exception [9]. Hypercholesterolemia is easily misdiagnosed if blood sampling is not done at regular time intervals. Since no symptoms are observed in the early phase of disease process secondary to hypercholesterolemia, the diagnosis might come too late and with subsequent delay in starting cholesterol-lowering medication [10]. Finally, patients might stop their medication because of perceived intolerance.

Diabetes, overweight and obesity also occur in CHD patients [9,11]. Overweight and obesity are found in relatively young patients with even complex underlying CHD (1–2/10 patients) [12]. Being overweight or obese leads not only to an unfavorable metabolic profile [13] (and consequently an increased cardiovascular risk profile) but contributes also to orthopedic complications (back, hip, and knee). Overweight and obesity are difficult to address therapeutically as they require permanent modification to a healthy lifestyle.

Finally, a *sedentary lifestyle* is also a rather common cardiovascular risk factor in CHD patients [14]. A study conducted by Dua et al. found that even up to 100% of patients with complex CHD do not achieve the minimum requirements of daily physical activity [15]. Dua et al. suggested 30 min a day of at least moderate intensity physical activity on five or more days of the week. The reasons why the minimum requirements are not achieved, are unclear. It may be that the health care professionals themselves are somewhat apprehensive [3] about encouraging a patient to engage in physical activity. In addition to that, clear guidelines were never issued and due to the phenomenon of overprotection [16], one may avoid encouraging physical activity. The patient himself may also be reluctant of being physically active. Patients may lose confidence in their body, which leads to insufficient exercise. Particularly when (sometimes harmless) symptoms appear, physical activities are omitted or advised to be omitted. Overprotection leads to a sedentary lifestyle. This is facilitated by parents that have been too protective in childhood. On the other hand, most adult CHD patients are willing to increase their physical activity levels [15].

### 3. Benefits of physical activity

Research has been done to evaluate the effect of exercise on health. Malm et al. (2019) published an extensive review on this topic [17]. Sport not only increases an individual physical condition, but has also a positive effect on overall physical and the mental health status. Physical activity leads to better blood pressure control, favorably affects the lipid profile, and promotes more efficient body weight control. A beneficial effect is also noted at biochemical and cellular level. All this might lead to a clinically significant reduction of the patient's cardiovascular risk profile.

The type of sport determines the nature of the beneficial effect to

some extent. Aerobic dynamic exercise has a better outcome on most health parameters when the effects are compared with isolated sub maximal strength exercise. In general, a volume-loaded circulation is healthier than a pressure-loaded one and this needs to be accounted for when advising on type, volume, and intensity of exercise. Even though no long-term outcome studies in CHD have been conducted, data are extrapolated from the general population to CHD patients. Besides a positive effect on the physical status, Niemann et al. showed that in patients with Fontan circulation leisure sports activity was not only associated with a better exercise capacity, but also with a better subjective healthiness and higher quality of life [18]. Ko et al. documented that more physical activity was even related to less symptoms of depression [19].

### 4. Efficiency of exercise and sports in congenital heart disease

Given that no long-term hard outcome data are available, the effects of physical activity and sport are usually measured through a surrogate endpoint. The preferred variable is the maximum oxygen consumption, which is easily obtained via a cardiopulmonary exercise test. Kempny et al. documented that depending on the underlying complexity of the CHD, maximum oxygen consumption is reduced [20]. Maximum oxygen consumption and other variables obtained from a cardiopulmonary exercise test (maximal exercise capacity, blood pressure evolution, ventilation/carbon dioxide production, ...) have predictive value for clinical outcome (mortality and/or morbidity) according to the type of CHD [8,21,22]. Physical activity, exercise, and sport improve maximum oxygen consumption in CHD patients. Most of these studies were summarized in a review paper by Tran et al. [23] Patients have better exercise capacity and feel better following exercise training or being physically active. It remains unclear whether the increase in maximal oxygen consumption will finally translate into better long-term outcome and reduced morbidity, but from a theoretical point of view, it can be assumed that cardiovascular mortality and morbidity related to atherosclerosis will decrease. To what extent cardiovascular mortality and morbidity related to the underlying CHD are influenced by exercise remains yet unclear.

### 5. Safety of sports and exercise in congenital heart disease

Concerns might arise about the safety of encouraging patients to engage in physical activity or exercise. Few reports suggest that exercise and sports are safe in CHD patients. The fear that exists in sports is that an acute event might occur including sudden cardiac death. Koyak et al. analyzed CHD patients who died suddenly [24]. Only 10% of all sudden deaths occurred during exercise. This suggests that physical activity is not a major trigger for sudden cardiac death. Opic et al. followed a cohort of patients (ranging from simple to complex) for 10 years, one part of whom exercised regularly and the other not [25]. The number of complications related to CHD in the group that exercised did not differ from the group that did not. All this suggests that there are no significant adverse effects for patients with simple and for patients with moderate/complex underlying conditions. Van Dissel et al. followed a group of patients who exercised 3 times a week for 6 months [26]. No adverse cardiovascular events occurred during and within the first 3 h after exercise. During further follow-up, some arrhythmias and musculoskeletal problems were reported. However, the arrhythmias were minor and did not occur more frequently when compared to the control group. More recently, Schuermans et al. published a meta-analysis on the effects of sports in patients with repaired tetralogy of Fallot [27]. Besides an improvement in exercise capacity, no serious adverse events could be demonstrated. All these studies suggest that physical activity in CHD patients does not lead to an increased number of complications, early mortality, or morbidity.

## 6. Type and intensity of sport

The definition of sports is always under discussion. According to the European council sport means all forms of physical activity, which through casual or organized participation, aim at expressing or improving physical fitness and mental well-being, forming social relationships, or obtaining results in competition at all levels (Recommendation No. R (92) 13 Rev of the committee of Ministers to Member states on the revised European Sports Charter).

Ultimately, physical activity can be classified into different forms. The term *physical activity* excludes the element of competition, is defined as regular exercise in daily life and physical fitness is maintained without competition. The next level is *recreational sports*. Recreational sports can be non-competitive or competitive in nature. Non-competitive sports are considered as a more intensive physical activity without competing. On the contrary, *competitive sports*, regardless of the level of achievement, is the strong desire for participants to exert themselves physically to their limits and to improve performance [28]. And even if sports are done several hours per day, if not performed at the highest (professional level) this can still be categorized as “recreational”. Table 1 summarizes the types of physical activity.

A distinction should be made between individual sports and team sports. In a team sport, one will easily tend to be highly competitive and go to extremes. Classic team sports therefore rank among competitive/athletic sports, although the sport intensity can be relatively low when at a recreational level.

In 2012 the European Society of Cardiology provided recommendations for advising physical activity and recreational sports for children with CHD [29]. Extrapolation to adults with CHD was at that time recommended because of lack of information for adult patients. Guidelines for competitive and elite sports in healthy individuals could also be consulted [30]. But, certainly in terms of elite sports, this guideline document was only applicable to 1% of adult CHD patients. A consensus paper was also available at that time and formulated advice for recreational, competitive and elite sports [31]. This consensus paper relied mainly on the anatomical diagnosis of the congenital defect so that some patients with complex conditions even with hemodynamic stability were virtually not allowed to do sports and simple anatomical conditions with complications on the other hand were more likely to be allowed to strenuous exercise despite having some risk factors.

Using these documents interchangeably led to inconsistent medical opinions and exercise advice for the same pathology with differences between centers (national and international) and even within the same CHD unit. Therefore, the former European Society of Cardiology Working Group on Grown-Up Congenital Heart Disease (currently the Working Group on Adult Congenital Heart Disease) and the European Society of Cardiology, section of Sports Cardiology, decided to produce new consensus documents for physical activity and recreational sports (2013) and for competitive sports (2020) in adults with congenital heart disease [28,32].

## 7. Physiological concept of choice of exercise

Budts et al. suggested that the static component of the physical activity in CHD patients should depend on the hemodynamic and electrical

**Table 1**  
Types of physical activity.

Physical activity	Daily exercise with no competitive component
Recreational sports	
Non-competitive	Daily exercise or exercising several hours per week with no competitive component
Competitive	Several hours per week with a competitive component, but without reaching extremes
Competitive sports	Several hours per week with a competitive component and with reaching extremes

stability of the underlying cardiac condition [32]. Five basic characteristics were assessed (ventricular morphology and function, pulmonary artery pressures, aortic dimensions, presence of arrhythmias, and oxygen saturation) and the more these variables deviate from normal, the more conservative the advice relating to intensity and volume of exercise should be, in particular to the static component of the sport. The (patho-)physiological reasons behind this concept are the following: dynamic exercise has a more beneficial effect on cardiac function and circulation than static exercise. With dynamic exercise, stroke volume and heart rate response are easier achieved and more pronounced and as a result, cardiac output increases more smoothly and consistently. It also promotes lowering systemic vascular resistance. Static exercise is more likely to lead to a pressure load on the circulation, less incremental build-up of stroke volume and cardiac output, and particularly induces the development of concentric muscular hypertrophy of the ventricle [33]. In this concept, the pre-existing pressure load of the ventricle determines the static component of the type of exercise. The next step is to choose the intensity at which to perform the effort. In a heterogeneous ACHD population, the balance between the static component of an effort and the intensity at which the effort is performed allows for an individualized exercise prescription. The algorithm is simplified in Fig. 2. The authors indicate that a thorough knowledge of the CHD is needed before formulating exercise advice. The medical and surgical history must be known, and the likely complications considered. Red flag symptoms rule out physical activity and exertion, but other symptoms per se should not be a reason to justify a sedentary lifestyle [32].

To reach an effect on the outcome of health, it is recommended to be 3–4.5 h per week physically active or to engage in a specific exercise. Each session should take at least 30 min. If starting from a deconditioned state, the intensity should be built up step by step and periodical re-assessment and follow-up are required. The position paper indicates that the follow-up frequency for physical activities with medium or low intensity is similar as the follow-up frequency that is stated in the European Society of Cardiology Grown-up Congenital Heart Disease guidelines [34], recently updated to the Adult Congenital Heart Disease guidelines [35]. For high intensity physical activities the follow-up frequency is stated by the European Society of Cardiology guidelines for competitive and elite sports [30], also recently updated [36]. An example of a case is given in text box 1.

## 8. Competitive sports

In 2020, the same authors of the position paper of physical activity in adults CHD patients released a paper focusing on competitive sports in CHD patients [28]. There was a need for this document because the new European Society of Cardiology guidelines related to competitive sports and athletic sports did not provide sufficient information for adult CHD patients. The most important update was that relative intensity is no longer included in the sport’s medical advice because in competitive sports, the athlete usually wants to go to the limit and produce a maximum effort. Team sports are also considered as competitive because intensity is difficult to restrict. The hemodynamic and electrical stability of the heart are evaluated first. It is this stability that will determine what type of sport can be done (and assuming everything is done at maximum intensity required by the respective sport). A more modern classification of sports was used and refers to endurance, mixed, power, and skills [37]. The underlying algorithm is summarized in Fig. 3. For team sports it is advised that patients participate in teams with similar physical fitness. Individuals who feel that their fitness is lower than that of their peers should be supported to find an alternative team sport [32].

It was highlighted that the suggestions made for recreational and competitive sports are not based on strong methodological studies, but rather on a rational approach to the effect of sports on underlying heart disease. It was the intention, therefore, to offer a decision platform to the health care professional for more uniform advice in adult patients with

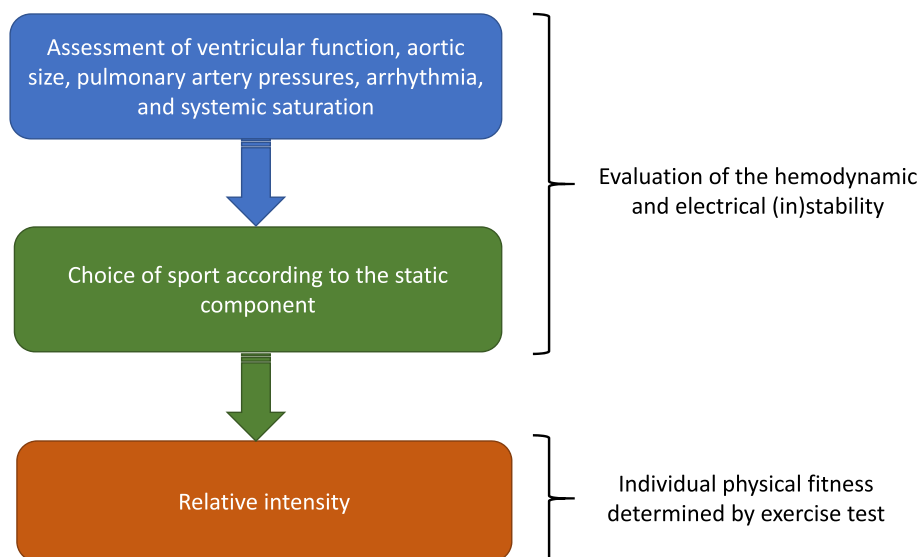


Fig. 2. Condensed decision algorithm for physical activity and recreational sports in adult congenital heart disease.

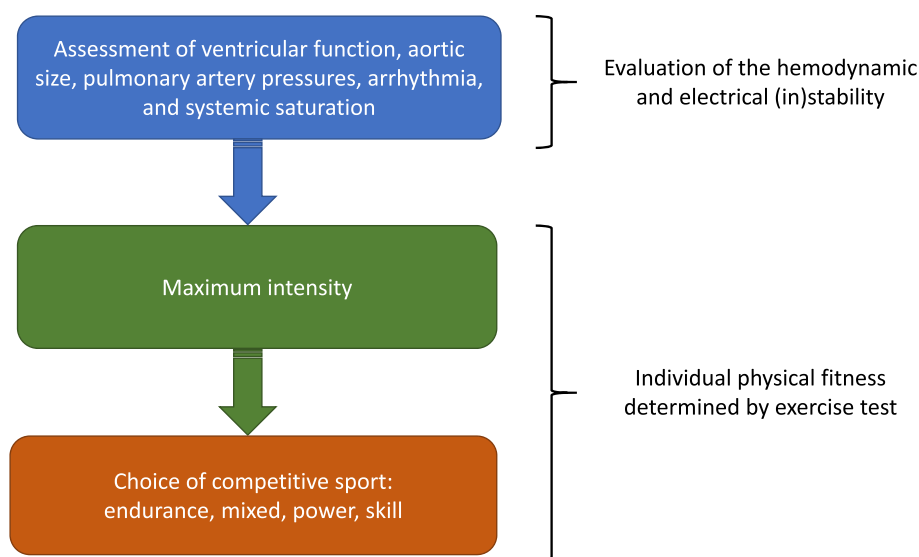


Fig. 3. Condensed decision algorithm for competitive and athletic sports in adult congenital heart disease.

CHD. An example of a case is given in text box 1.

## 9. Specific case considerations in congenital heart disease

This section deals with specific situations that may occur in a global cardiac population, consequently also in CHD patients. The topics covered in this section are general observations and suggestions that can be extrapolated to adult CHD patients.

### 9.1. Pacemaker and resynchronization therapy

The presence of a pacemaker does not preclude high intensity physical activity. However, appropriate rate adaptation is in most cases requested. Minute ventilation-based rate-response systems are preferred over accelerometer-based systems since they provide more physiological rate response [38]. Exercise testing and Holter ECG monitoring may help to program appropriate pacing rate responsiveness during exercise. However, increasing chronotropy is not always effective. Uebing et al. showed that rate-responsive pacing does not improve exercise capacity in patients with a systemic right ventricle [39]. Too fast pacing might

lead to reduced diastolic filling and consequently a decrease in stroke volume. The risk for damage to the device through body movements or contact needs to be considered.

### 9.2. Implantable cardioverter defibrillator (ICD)

Low to moderate intensity physical activities are recommended in patients with an ICD [40] and this advice can be extrapolated to CHD patients. However, a general restriction of athletes and patients with CHD to low and moderate intensity is not fully justified as the threshold for ventricular fibrillation detection can be adapted to individual and sports specific requirements. This however requires individualized and close rhythm monitoring during sports and physical activity. It is unclear how effective cardiac defibrillation is during peak and high intensity physical activities, however current studies do not point towards increased ICD malfunction or indeed fatalities [41]. Nevertheless, arrhythmias were more frequent during physical activities and there is an increased risk for inappropriate shocks. However, newer devices seem to have less inappropriate shocks because of better sensing. Also here, the risk of device damage in contact sports and through body movement



need to be considered. Sports specific considerations on matters of type of device (sub-cutaneous vs. *trans*-venous) and enhanced device protection are paramount in these scenarios.

### 9.3. Exercise at high altitude

Hypobaric hypoxia found at high altitude leads to decreased oxygen consumption, decreased cardiac output, tachycardia, hypocapnia mediated stroke volume decrease and a rise in pulmonary vascular resistance in the acclimatization phase. These features are part of the pathophysiology in many patients with CHD and hence these mechanisms can reduce oxygen tissue uptake significantly and lead to exacerbation of symptoms in patients with complex CHD. High altitude induced left ventricular dysfunction is extremely rare, however worsening of right ventricular dysfunction can occur. Myocardial ischemia is not observed in the healthy population but can present an additional problem in patients with complex and cyanotic CHD or pre-existing ischemic heart disease. Therefore, patients with cyanotic, unrepaired, or palliated complex CHD or CHD with associated pulmonary hypertension should be advised against competitive sport at moderate (1500–2500 m) or high (>2500 m) altitude and be advised they might become symptomatic even at rest [28]. There is a reported but rare risk of exacerbation of pre-existing arrhythmias at moderate and high altitudes [42], but moderate and high altitude as a direct cause for arrhythmias has not been confirmed. Air travel is well tolerated but stays at moderate and high altitude should follow an individual assessment by ECG, echocardiography and CPET.

### 9.4. Anticoagulation therapy

Anticoagulation is no contraindication for physical activity or sports in patients with CHD. However, in general it is advised against engaging in contact sports [28].

### 9.5. Wearables and remote coaching

The use of wearables has also a place in CHD patients. The purpose is twofold. First, the wearable can motivate patients to pursue a minimum physical activity status. Second, wearables can identify early detection of disease progression. However, extrapolating sensor technology and validated algorithms in structurally normal hearts to CHD must be done with caution. Tandon et al. reviewed the literature and concluded that the potential gain for patients with CHD from these technologies is immense and stated that the further developing of meaningful digital biomarkers in real-world settings is requested [43]. A potential downside is that patients might become too fixated on their health parameters, as a certain anxiety could be induced.

Several CHD patients still have professional lives which means they have less time to seek expert advice. However, it is important that when a sport-medical advice is given, the patient still undergoes an interview, a clinical examination, and some technical checks. According to general guidelines, CHD requires a systematic follow-up, and it is during the follow-up visit that sport-medical advice can be formulated. Thereafter, it may be useful to coach and adjust the patient from a distance if the motivation to exercise decreases or the type of exercise does not have the right emphasis. Remote coaching has the great advantage that someone (a person) keeps patients motivated to exercise [44–48].

## 10. Conclusions

A minimal of physical activity in all CHD patients is required. The patient population is aging and makes them more and more exposed to the common cardiovascular risk factors for atherosclerosis. Regular physical activities might counter that in the setting of primary prevention.

General exercise protocols seem to be safe (on the short term) for

patients with CHD. However, long-term results are unknown. Available consensus documents might help to counsel for personalized exercise prescription advice.

## Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

## Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ijcchd.2023.100467>.

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