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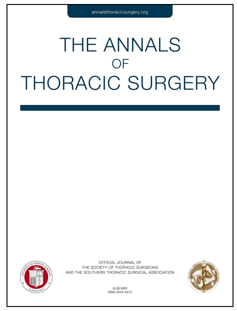

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Exercise Testing and Training in Adults with Congenital Heart Disease: A Surgical Perspective

Running Head: CHD and Exercise: A Surgical Perspective

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ABSTRACT

In the current era, the majority of children born with congenital heart disease (CHD) will survive well into adulthood due to major advances in surgical techniques as well as critical and medical care. However, reoperation and palliative surgical interventions are increasingly common in the adult CHD population. Tools to effectively risk stratify patients and therapies to improve outcomes are required to optimize the management of adult CHD patients during the pre- and post-operative period and beyond. Exercise testing is an invaluable tool to guide risk stratification. In addition, exercise training in people with CHD may decrease post-operative complications by enhancing physiological reserve and also has an important role in physical rehabilitation. This review aims to provide individualized recommendations on exercise prescription in CHD patients in the pre- and post-operative settings. The response to exercise testing and prognostic implications will also be discussed.

ABBREVIATIONS

$\%HR_{peak}$; percentage of peak heart rate

%HRR; percentage of heart rate reserve

1RM; one-repetition maximum

CHD; congenital heart disease

CPET; cardiopulmonary exercise testing

ECG; electrocardiography

HR; heart rate

HRR; heart rate reserve

MRI; magnetic resonance imaging

RER; respiratory exchange ratio

RPE; ratings of perceived exertion

VE/VCO_2; minute ventilation-carbon dioxide production

VO_2; oxygen uptake

As we enter the new era of treating and managing adults living with congenital heart disease (CHD), it is likely many will require further surgical interventions. While the contemporary CHD patient is surviving longer, many also have unfavorable comorbidities that increase the risk of adverse post-operative outcomes.[1]

Exercise testing and training have an important role in the pre- and post-operative settings for CHD patients. Exercise training is a well-established therapy in a variety of chronic conditions with important implications on clinical outcomes. Benefits including improved body composition, vascular function, cardiorespiratory fitness, muscular fitness, psychosocial health and a reduction in cardiovascular risk factors are all associated with chronic exercise training.[2, 3] Exercise prescription is more complex for CHD patients than in many other cohorts due to heterogeneous cardiac anatomy, physiological status and comorbidity, but is well tolerated when implemented appropriately. However, much of what is known is based on clinical experience, and scientific evidence remains limited. We have previously written a comprehensive review on the topic of exercise prescription for CHD patients.[4]

This review will introduce the fundamental concepts of exercise testing and prescription during the pre- and post-operative periods in CHD patients. We will also present a method for individualizing recommendations for exercise prescription based on cardiac anatomy and physiological status.

EVALUATION OF EXERCISE CAPACITY AND RESPONSE

The assessment of exercise capacity is a highly underutilized tool in the management of CHD patients. When conducted and interpreted appropriately, results from an exercise test can provide prognostic information and help guide exercise prescription. In patients with complex heart disease, exercise testing also aids as an important screening tool for cardiac arrhythmia.

Cardiopulmonary Exercise Testing Response

The gold standard measure of aerobic exercise capacity is peak oxygen uptake (VO_2), reflecting cardiorespiratory fitness assessed by cardiopulmonary exercise testing (CPET).

On average, adults living with CHD have moderately impaired exercise capacity (66% of age-predicted peak VO_2).[5] The degree of exercise impairment is comparable to acquired chronic heart failure,[6] which is concerning given the relatively young age of the CHD population. Despite depressed exercise capacity, this is not always associated with symptoms.[6] The severity of exercise intolerance can be vastly heterogeneous between and within different CHD diagnoses (Figure 1).[7]

CHD patients are frequently unable to reach their age-predicted maximum heart rate (HR) and have a reduced HR reserve (HRR; peak HR – resting HR). Chronotropic incompetence is typically defined as the inability to achieve 80% or 85% of age-predicted maximal HR (or 62% in patients on beta-blockers). The prevalence in CHD ranges from 30-60%;[8] universal definitions for chronotropic incompetence and the prognostic implications are not well established in the setting of different CHD lesions. Depressed peak HR may be related to residual scarring from surgical procedures or abnormal hemodynamic cardiac loading. The presence of right-to-left shunting can manifest as exercise-induced desaturation and can be associated with impaired exercise capacity.

Ventilatory responses during exercise are abnormal in most patients with CHD.[9] The minute ventilation-carbon dioxide production (VE/VCO_2) slope is typically elevated (ventilatory inefficiency), reflecting ventilation-perfusion mismatch and increased physiological dead space. Despite the markedly abnormal ventilatory response during exercise,[9] the majority of patients do not encroach upon their breathing reserve, even though a restrictive ventilatory impairment may exist.[10] This is likely associated with respiratory muscle weakness or chest wall abnormalities.

The 'peak' CPET results for patients who do not achieve a peak respiratory exchange ratio (RER) >1.05 or meet 'maximal' criteria should be interpreted with caution because this is suggestive of submaximal effort. In these patients, parameters derived from submaximal exercise data, including the oxygen uptake efficiency slope, VE/VCO_2 slope and the 'anaerobic threshold' may be a more reliable indicator of clinical severity.

Prognostic Utility of Cardiopulmonary Exercise Testing

CPET variables can aid in risk stratification and have important implications for prognosis.[5, 6] Diller *et al.* demonstrated CHD patients with a peak VO_2 lower than 15.5 mL/kg/min (and RER >1.0) had an almost 3-fold increased risk of death or hospitalization.[6] These findings were replicated in a large series that showed the combination of peak VO_2 and HRR was the strongest predictor of 5-year survival.[5] If criteria for an adequate test effort are not met (RER <1.0), the VE/VCO_2 slope is also a strong predictor of mortality but is limited to non-cyanotic patients.[5, 8, 9] More recently, serial CPET data have been shown to provide prognostic information; a decline in VO_2 is associated with a higher likelihood of adverse clinical outcomes.[11]

Surgical Risk Stratification

The utility of CPET for assessing pre-operative risk, morbidity, mortality and suitability for transplantation is well recognized in cardiac and non-cardiac conditions.[12] In patients with tetralogy of Fallot and pulmonary insufficiency, CPET can help guide risk stratification for pulmonary valve replacement; a peak VO_2 <20 mL/kg/min is predictive of post-operative mortality.[13] Birkey *et al.* found an association between ventilatory inefficiency and post-operative inpatient length of stay.[14] In that series, the requirement for prolonged ventilation was associated with pre-operative decreased peak VO_2 and HR response. The available evidence in non-cardiac surgery cohorts further supports using CPET as a surgical risk-stratifying tool. Higher cardiorespiratory fitness is associated with survival, reduced hospital length of stay and fewer post-operative complications.[12] High risk patients in a variety of patient cohorts have been identified using commonly proposed thresholds; peak VO_2 <15 mL/kg/min or VO_2 at 'anaerobic' threshold <9–11 mL/kg/min.[12] The latter may be particularly useful in patients where maximal exercise testing may not be appropriate.

While the landmark study by Mancini *et al.* established a robust threshold (>14 mL/kg/min) to determine appropriate timing for heart transplantation in non-CHD heart failure patients,[15] evidence does not currently exist to support the use of this threshold in adult CHD patients.[16] Nevertheless, both baseline and serial CPET can provide objective information for clinical surveillance to guide the appropriate timing of interventions. Therefore, it is recommended surgeons or cardiologists refer patients for serial and pre-operative CPET.

Exercise stress testing

When direct gas exchange measures are not available, exercise stress testing can be performed with or without imaging (echocardiography or magnetic resonance imaging; MRI) on a treadmill or bicycle with pulse oximetry, blood pressure and electrocardiography (ECG) monitoring. Exercise capacity without gas exchange can be expressed as metabolic equivalents estimated from regression equations.

Exercise stress echocardiography performed on a treadmill or cycle ergometer can provide further valuable information related to cardiac function, although the limitations of traditional two-dimensional echocardiography are further amplified in many CHD patients. Atypical cardiac chamber architecture invalidates Simpson's rule, and ventricular function is often limited to qualitative grading unless three-dimensional echocardiography is available.

Exercise stress echocardiography is most frequently used in adults for the assessment of exercise-induced ischemia. Wall motion abnormalities may be suggestive of ischemia and are more sensitive and specific than ECG changes, especially when the baseline ECG is abnormal. Stress echocardiography in the setting of CHD can also be useful to assess for dynamic outflow tract obstruction, ventricular reserve and pulmonary hypertension, but CHD echocardiographic expertise is essential to aid interpretation of the results.

Assessing muscular fitness

Impaired muscular fitness is common and is associated with reduced lung function and exercise capacity in CHD patients but is not often formally assessed.[17, 18] Strength testing is valuable to help guide exercise prescription individualized for resistance training. The practical method of assessing dynamic strength is by measuring a one-repetition maximum (1RM) and is defined as the highest load that can be lifted once with sound technique.

EFFICACY AND SAFETY OF EXERCISE TRAINING

The role of physical inactivity in chronic disease is now well-recognized, and physical activity levels in patients with CHD have been documented as being lower than the general population.[2] Despite this, formal exercise advice is not routinely provided in people with CHD due to unfounded safety concerns, and historically, exercise restriction has been encouraged.[4]

In acquired heart failure, every 6% improvement in peak VO_2 is associated with a 5% reduction in all-cause mortality or hospitalization.[19] A meta-analysis demonstrated that exercise training in adults with CHD was effective for improving aerobic capacity as expressed by peak VO_2.[20] Exercise-induced adaptations resulting in improvements in peak VO_2 may partly relate to enhanced stroke volume.[21]

Exercise training may modestly improve both diastolic and systolic function in patients with acquired heart failure.[22, 23] While the evidence in CHD is limited, in some types of CHD, preliminary studies have reported improvements in cardiac function with exercise training; One study showed an increase in stroke volume assessed by cardiac exercise MRI in Fontan patients following resistance training likely due to enhanced preload.[24] The training-related improvement in peak VO_2 may also be attributed to pulmonary, hematologic, vascular or mitochondrial changes. Evidence of enhanced skeletal muscle oxidative capacity reflected by a later onset of the 'anaerobic' threshold has been shown with exercise in CHD.[25, 26] Other adaptations from exercise training include improvements in respiratory and peripheral muscle oxygenation,[4] cardiac autonomic function,[27] vascular function,[27] skeletal muscle hypertrophy,[24] ventilatory efficiency[26] and a reduction in the occurrence of non-sustained ventricular tachycardia.[28] The feasibility of high intensity interval training has been evaluated in patients with tetralogy of Fallot and transposition of the great arteries and appeared to be well tolerated and effective in these populations.[27, 29, 30]

The efficacy and prescription of resistance training is poorly characterized. Cordina *et al.* demonstrated isolated high intensity resistance training could improve cardiac output and peak VO_2 in adult Fontan patients,[24] because increasing lower limb muscle mass through resistance training decreases venous compliance and augments peripheral muscle pump function. The effects of isolated resistance training in other CHD lesions remain unknown and require further investigation. However, given the widespread prevalence of lean mass deficits,[31-33] resistance training is likely to be beneficial.

Home-based cardiac rehabilitation programs may be a suitable alternative to traditional cardiac rehabilitation programs with improvements in peak VO_2 ranging from 1.2% to 15%, albeit with high dropout rates (15%) in studies with training durations ranging from 6 to 24 weeks.[34] While home-based programs may improve the accessibility to cardiac rehabilitation services, unsupervised sessions may come at the expense of compliance.[35]

Importantly, exercise training appears to be safe in CHD patients. Systematic reviews consistently report the absence of serious adverse events associated with exercise training.[3, 20, 34] Notably, the occurrence of severe exercise-induced arrhythmias, sudden death or other serious adverse events do not appear to be higher than that observed in acquired heart disease patients.[36] Sudden cardiac death is rare, but patients at increased risk should be screened, counseled appropriately and progressed slowly. There appears to be no elevation in serum markers of myocardial injury and hemodynamic stress after exercise interventions.[21, 29, 30] Furthermore, imaging studies show no signs of worsening ventricular function.[25, 37] Shared decision-making is encouraged, and arrhythmias should be controlled prior to commencing exercise training. It is important to recognize that most study participants are carefully selected and exclude high risk and unstable patients, so these findings should be treated with caution when extrapolating to the broader cohort of CHD patients.

Psychosocial considerations

Patients living with CHD are at increased risk for mental health issues and reduced quality of life – problems that can be aided through regular exercise. Increasingly, the harmful implications of clinician-recommended exercise restriction on perceived health and quality of life are being recognized.[38]

PRE- AND POST-OPERATIVE EXERCISE TRANING

Pre-operative exercise may increase patient eligibility for surgery and improve their ability to tolerate surgery-induced physiological stress.[39] Improving cardiorespiratory fitness and skeletal muscle reserve may attenuate the negative consequences of bed rest and facilitate post-operative recovery.

Prehabilitation (pre-operative exercise training)

The prehabilitation framework encompasses a multi-modal approach, including physical activity, nutritional support, respiratory muscle training, medical optimization and psychological support, with the aim of improving post-operative outcomes.[40] This contrasts with post-operative cardiac rehabilitation that aims to restore functional capacity to pre-operative levels and promote secondary prevention. Homeostatic imbalance induced by surgical stress is characterized by increased metabolic demand, catabolism and neuroendocrine changes. Post-operative complications appear to be interrelated with the body's ability to offset the surgical stress response. The notion of prehabilitation is to increase physiological reserve in an attempt to meet the physiological demands of surgery, and in turn, decrease the risk of post-operative complications (Figure 2),[39] although data specific to the CHD population are lacking.

Patients with CHD may be at an increased risk of surgical complications secondary to pre-existing impaired lung and respiratory muscle function,[18, 41] skeletal muscle deficiencies[31-33] and reduced physiological reserve.[6]

Prehabilitation programs generally range from four to eight weeks.[42] A minimum prehabilitation duration of four weeks is generally required to increase physiological reserve.[42] However, meaningful changes in muscle mass may require consistent training of 12 to 16 weeks. Aerobic training can induce muscle hypertrophy, although resistance training is generally more effective at increasing muscle mass. Therefore, a combined resistance and aerobic training program is considered the optimal approach to improve cardiorespiratory fitness, muscle mass and function in the pre-operative setting.

While evidence is currently limited in CHD, it would be reasonable for surgeons to refer patients for prehabilitation. In elective surgical candidates, delaying surgery to allow for a period of prehabilitation to improve physiological reserve may be considered. Furthermore, it appears that participation in prehabilitation increases the compliance to post-operative rehabilitation programs.[40]

Post-operative exercise training

In addition to promoting early post-operative ambulation and physical activity, we also recommend implementing a structured exercise program. In low risk patients, commencing aerobic exercise early (within two weeks) following cardiac surgery can improve both functional and aerobic exercise capacity with a similar adverse event rate to usual post-operative care.[43] In acquired heart disease, improvements in peak VO_2 following cardiac rehabilitation in patients who underwent coronary artery bypass grafting, heart valve replacement or percutaneous coronary interventions was inversely associated with the risk of hospitalization and mortality.[44] Preservation of sternal stability (discussed below), range of motion, post-operative pain and discomfort need to be considered. Exercises should be modified if they induce pain or excessive discomfort. Following the sternal precaution period and when sternal stability has been established, resistance exercise prescription should not be restricted beyond general recommendations for CHD patients.

Sternal Precautions

When a pain-free range of motion and ambulation is achieved, patients who undergo procedures involving sternotomy should initiate exercise as soon as possible to counteract the adverse effects of prolonged bed rest. The introduction of low intensity resistance training can occur four weeks following interventions involving a sternotomy provided there is adequate sternal stability. There is a lack of consensus regarding the timing, prescription, progression and restrictions of exercise following a median sternotomy. It is common in clinical practice for upper-limb resistance exercise to be restricted for a period of 8 to 12 weeks.[45] In addition, a weight limitation between 5lb (2.3 kg) and 10lb (4.5 kg) for up to 12 weeks post-surgery is often imposed.[45, 46] The appropriate timing of initiating upper limb exercise remains controversial, and the majority of institutions will have a center-specific sternal precaution protocol. Many CHD patients will have multiple sternotomies, which increase the risk of sternal healing complications, and an individualized conservative approach may be warranted.

EXERCISE PRESCRIPTION.

The format of exercise prescription should be designed to follow the frequency, intensity, time, type, volume and progression (FITT-VP) principles.[46] We have previously published recommendations adhering to these principles to support individualized exercise prescription based on the patient's risk classification (Table 1),[4] similar to the approach suggested by Budts *et al.*.[47] These recommendations may also be used in the pre- and post-operative settings. Post-operative exercise training following a sternotomy should also take into account sternal stability (see above). It is recommended that patients are referred to exercise professionals (e.g., exercise physiologists, physical therapists or physiotherapists) to optimize exercise prescription, resistance exercise technique and adherence. However, this should not restrict surgeons, cardiologists, nurses or other health care professionals from providing advice and promoting exercise. Even the prescription of simple incremental structured walking programs can be beneficial in CHD patients.[48]

Prior to exercise prescription, patients should be adequately screened by a CHD cardiologist for contraindications to exercise training or testing.[4] Medical investigations including; exercise testing, echocardiography, cardiac MRI and/or 24-hour Holter monitoring may be performed to aid in risk stratification. The evaluation of ventricular function, structural aortic integrity, valvular sufficiency, estimated pulmonary pressure, arrhythmia burden and the presence of outflow tract obstruction can be used to assess the patient's risk as either low, moderate or high. The patient's risk classification can help guide individualize exercise prescription recommendations in relation to the FITT-VP principles (Table 2). Most patients can safely engage in low to moderate intensity exercise in the community. In high risk patients, it is recommended for exercise to be initially undertaken with supervision in a hospital or outpatient rehabilitation medical facility. After a period of supervised exercise training and education on self-monitoring, patients can be transitioned to community or home-based programs.

It is also important to consider physical activity history to individualize exercise prescription. Patients who have previously engaged in moderate to vigorous physical activity or have experience with resistance training may potentially progress at a faster rate compared to their novice counterparts.[46] A summary of the considerations of key exercise prescription variables for CHD is shown in Table 3.

Frequency, time and mode (type)

Consistent with guidelines for healthy adults,[46] we recommend engaging in aerobic exercise at least three days per week and preferably five days per week. Resistance training should be performed at least twice weekly, either combined with aerobic exercise sessions or in isolation.

The optimal modalities and methods of exercise prescription are not well established in CHD. A combination of aerobic and resistance exercises that incorporate large muscle groups is recommended. In severely deconditioned patients, aerobic interval training may be used to increase exercise tolerance, and high intensity interval training may be trialed in a subset of low risk patients. It may also be reasonable to implement intra-set rest periods (‘cluster sets’) for patients who do not tolerate higher repetitions during resistance training, although data are lacking with regard to this approach. Incorporating intra-set rest periods may reduce the acute hemodynamic response of resistance training in cardiac patients.[49] In patients with a Fontan circulation, lower limb exercises should be a primary component of a resistance training program to augment the peripheral muscle pump.

Exercise Intensity

Exercise physiologists and clinicians with expertise in exercise physiology may utilize detailed CPET indices for exercise prescription.[4] A simpler and acceptable approach is to use the percentage of the patient's peak HR (%HR_{peak}) achieved on a symptom-limited exercise test to prescribe relative aerobic exercise intensity.[47] It is not appropriate to prescribe exercise intensity using HR methods based on the predicted maximal HR as patients rarely achieve their predicted values due to chronotropic incompetence. In the setting of CHD, the %HR_{peak} method frequently underestimates the prescribed target exercise intensity, irrespective of chronotropic response. The percentage of HRR (%HRR) method is more accurate.[50] It should be noted the linear relationship between HRR and VO_2 reserve does not hold true in patients with atrial fibrillation or atrial pacing, and other methods should be employed (discussed below).

The HRR method can be calculated using the following formula;

$$\text{Target training HR} = \%\,target\ intensity \times (peak\ HR\ - resting\ HR) + resting\ HR$$

In the setting of atypical HR kinetics, exercise-induced hypoxia and ventilatory limitation to exercise, monitoring intensity using HR may not accurately represent the physiological response. In these patients, autoregulatory methods such as ratings of perceived exertion (RPE), the talk test or symptom guided intensity likely reflect exercise physiological stress more accurately. The talk test is a validated and practical tool that can aid in prescribing exercise intensity. It identifies the work rate during exercise where a conversation cannot be maintained uninterrupted, and this demarcates moderate and vigorous intensity exercise and approximates the first ventilatory threshold.[4]

Aerobic exercise intensity categories can also be estimated using the Borg RPE scale as light (Borg 8-10), moderate (Borg 11-13), vigorous (Borg 14-16), or high (Borg ≥17) based on the numerical response associated with the scale. Both the patients and 'observer' RPE should be used to titrate and adjust training intensity, especially in patients who are not familiar with the scale. In patients with cognitive impairment, the RPE 0 to 10 category-ratio scale may be more appropriate.

Resistance exercise intensity can be prescribed as a percentage of the patients 1RM directly assessed or estimated from a multiple repetition maximum test (i.e., 5 to 10 repetition maximum). The 'load to repetition relationship' can provide an estimate of resistance exercise intensity and help guide prescription. Similar to aerobic training, an RPE scale can be used to subjectively estimate resistance exercise intensity. The Borg RPE, Borg category-ratio, OMNI-RES or 'repetitions in reserve' scale can be used, but the validity and reliability of these scales are unclear in cardiac patients.

Despite recently shifting opinion in favor of higher load and lower repetitions due to a lower acute hemodynamic load compared to lower load and higher repetitions,[51] lower intensity and higher repetitions is still recommended in high risk patients.

Volume ("dose") and progression

If possible, people living with CHD should aim to achieve a minimum total accumulated volume or "dose" equivalent to international physical activity guidelines.

Since exercise volume is the product of exercise prescription variables (i.e., FIT for aerobic exercise and sets, repetitions, load and frequency for resistance exercise), increasing any variable will result in an increase in exercise volume. Progressing frequency, duration (time), sets and repetitions are limited by time constraints. Although progressing these exercise prescription variables are recommended initially prior to increasing intensity, the increase in time required to complete the prescribed exercise volume should be considered. An interval approach combining intensity domains may be considered to increase overall exercise volume.

Muscular fitness assessments for resistance training intensity prescription are infrequently performed in clinical practice. Training load can be progressed when the prescribed repetition and load can be completed comfortably on the last set in consecutive sessions. Alternatively, the acclimatization ('titration') technique or "two for two" method is objective and easy to implement. The load can be increased when the patient can perform two repetitions of the prescribed load above the prescribed repetition range in two consecutive sessions.

FUTURE DIRECTIONS

While the currently available literature supports the implementation of exercise training as part of routine clinical care, large homogenous randomized controlled trials are limited. It is highly likely that exercise training in the pre- and post-operative period is also beneficial, CHD-specific data are needed. Many patients reside in remote or rural areas, making it difficult to attend a specialist center for cardiac rehabilitation. Therefore, research studies investigating the efficacy of telehealth exercise and lifestyle interventions are needed. Importantly, in the setting of acquired heart disease, telehealth interventions do not appear to be inferior compared to center-based cardiac rehabilitation.[52]

CONCLUSIONS

Exercise testing is a useful tool to help characterize surgical risk stratification and guide exercise prescription. Exercise training in CHD patients is beneficial and safe when prescribed appropriately and is likely to optimize pre-operative fitness as well as post-operative recovery. Surgeons, cardiologists and health care professionals should recommend exercise testing and training as standard care in CHD patients.

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Table 1 Risk classification of congenital heart disease patients to determine recommended training intensities

Risk classification	Ventricular function	Aorta	Outflow tract obstruction	Pulmonary Hypertension	Valvular function	Arrhythmias	Recommended exercise intensity
Low risk	Normal or only mild dysfunction	No coarctation or dilation	Minimal or none	No	No or mild regurgitation or stenosis	No history arrhythmias	Moderate to vigorous intensity AT and RT
Moderate risk	Moderate dysfunction	Mild coarctation or dilation	Mild	Mild	Moderate stenosis or regurgitation	History of mild arrhythmias	Low to moderate intensity AT and RT
High risk	Severe dysfunction	Moderate-severe coarctation or dilation	Moderate-severe	Moderate-severe	Severe stenosis or regurgitation	Malignant or significant arrhythmias	Low intensity AT and RT

AT; aerobic training, RT; resistance training. * If patients have factors in more than one classification, the higher risk stratification is applied. Detailed exercise training recommendations based on risk classification level are provided in Table 2. Reproduced from Tran and colleagues[4] with permission from Elsevier.

Table 2. Aerobic and resistance exercise prescription based on risk classification

Mode	Risk Classification	Intensity/relative volume load (resistance training)	Frequency	Duration (aerobic training)/Number of sets (resistance training)
Aerobic Training	Low	40 – 84% HRR or 55 – 89% HRmax 11 – 16 RPE	3 – 5 days/week	Commence at 5- 10 minutes and increase as tolerated to 30 – 60 minutes or Interval training may be employed to increase tolerance to exercise. Commencing at a work: active rest ratio of 1:2 progressing to 2:1.
	Moderate	20 – 59% HRR or 40 – 69% HRmax 8 – 13 RPE		
	High	20 – 39% HRR or 40 – 54% HRmax 8 – 10 RPE		
Resistance Training	Low	50 – 79% 1RM 1 – 3 sets, 8 – 10 repetitions ≥1 minute rest between sets	≥2 days/ week	Commence at 1 set progressing to 3 sets as tolerated. Initial supervision is recommended to provide instruction in correct lifting technique.
	Moderate	30 – 69% 1RM 1 – 3 sets, 10 – 12 repetitions ≥1 minute rest between sets		
	High	30 – 49% 1RM 1 – 3 sets, 12 – 15 repetitions ≥2 minutes rest between sets		

1RM, one-repetition maximum; Borg's RPE, rate of perceived exertion; HRmax, maximal heart rate; HRR, heart rate reserve. Exercise prescription using heart rate reserve can be calculated using the Karvonen method (prescribed exercise heart rate = % intensity × (peak HR – resting HR) + resting HR). Reproduced from Tran and colleagues[4] with permission from Elsevier.

Table 3. Special consideration for exercise prescription in congenital heart disease patients

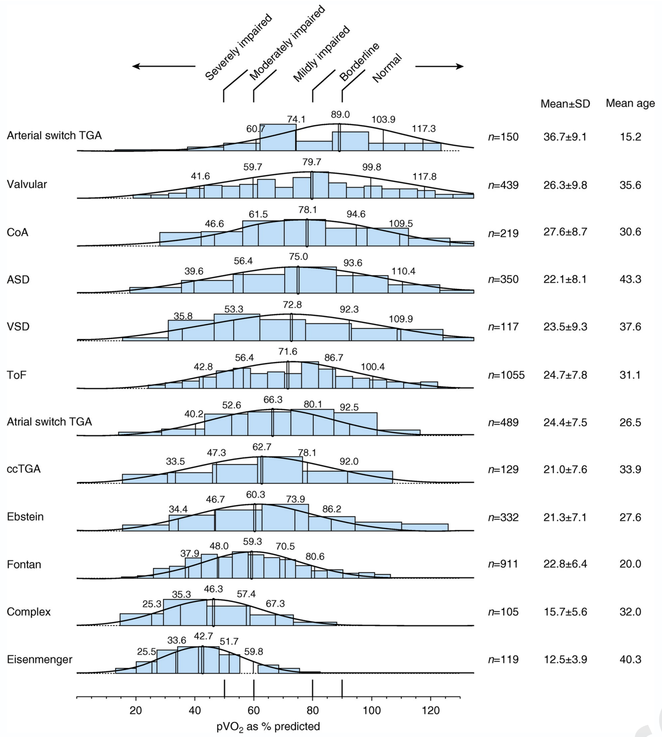
Special Considerations	
Cyanosis/hypoxemia	• Monitoring oxygen saturation is of limited utility; many patients will desaturate even during low intensity exercise • Exercise intensity can be prescribed using autoregulatory symptom-limited thresholds
Outflow tract obstruction and valvular disease	• Outflow tract obstruction may become more dynamic with exercise and increases the risk of syncope and hypotension • Valvular insufficiency predisposes the heart to chamber enlargement and arrhythmias
Pulmonary hypertension	• Patients with pulmonary hypertension have an increased risk of adverse events during vigorous-intensity exercise training • Compression of the main coronary artery from the dilation of the pulmonary artery during exercise can result in ischemia
Musculoskeletal	• Scoliosis/kyphosis is common due to multiple thoracotomies and/or sternotomies • Exercise modifications may be required due to a restricted range of motion or pain
Lung function and respiratory muscles	• Impairment in respiratory function can reduce exercise capacity • Exercise flow-volume analysis should be considered during CPET in the setting of abnormal lung or respiratory muscle function • Inspiratory muscle training can improve inspiratory muscle strength, ventilatory efficiency and may reduce post-operative pulmonary complications
Ischemia	• Patients with surgical repair and lesions involving the coronary arteries are at an increased risk of developing ischemia during exercise • Patients with stable ischemia secondary to acquired cardiovascular disease should limit exercise training intensity to 10-15 bpm below the ischemic threshold

Implantable cardioverter defibrillator and pacemakers	• Treadmills may be more useful for optimizing pacemaker rate responsiveness compared to cycle exercise • Pacemaker and implantable cardioverter defibrillator settings should be clarified, and intensity should remain below discharge thresholds to avoid inappropriate shocks • Excessive upper limb exercises should be avoided for 3-4 weeks after implantation
Psychological considerations	• Exercise self-efficacy is low and depression/anxiety is common. Psychological support to address these issues may improve compliance to exercise training
Cardiac rhythm	• Arrhythmias should be controlled prior to starting an exercise program • Chronotropic incompetence is common, and methods utilizing heart rate may not be valid in this setting

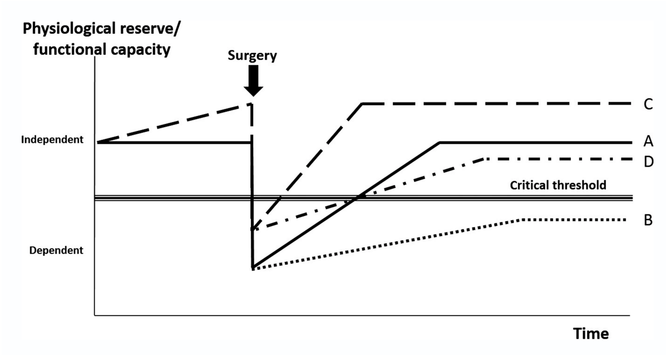
FIGURE LEGENDS

Figure 1. Peak oxygen uptake (peak VO_2) data expressed as % of predicted value. The density lines above histograms and the numbers to the right of the graph relate to all patients with a given diagnosis. The numbers above the density lines indicate %peak VO_2 values for the 10, 25, 50, 75 and 90th centile. ASD, atrial septal defect; ccTGA, congenitally corrected TGA; CoA, coarctation of aorta; Complex, complex congenital heart disease (including univentricular hearts); Ebstein, Ebstein anomaly; Eisenmenger, Eisenmenger syndrome; Fontan, patients after Fontan palliation; TGA, transposition of the great arterial; ToF, tetralogy of Fallot; Valvular, mixed collective of patients with congenital valvular heart disease; VSD, ventricular septal defect. Reproduced from Kempny and colleagues[7] by permission of Oxford University Press.

Figure 2. The prehabilitation concept. Following major a surgical intervention all patients experience an acute drop in physiological reserve/functional capacity followed by a recovery and rehabilitation phase (A). A low physiological reserve/functional capacity may increase the risk of peri□operative complications and lead to a slower, sometimes incomplete recovery (B). A prehabilitated patient may possess greater physiological reserve/functional capacity at the time of surgery, facilitating a more rapid and complete recovery (C). Crucially, in the event of a complicated recovery, prehabilitated patients may be better placed to retain their functional independence and quality of life in the longer term (D). Reproduced from Tew and colleagues[39] with permission from John Wiley & Sons, Inc.



Severely impaired
Moderately impaired
Mildly impaired
Borderline
Normal
Mean±SD
Mean age
Arterial switch TGA
60.7
74.1
89.0
103.9
117.3
n=150
36.7±9.1
15.2
Valvular
41.6
59.7
79.7
99.8
117.8
n=439
26.3±9.8
35.6
CoA
46.6
61.5
78.1
94.6
109.5
n=219
27.6±8.7
30.6
ASD
39.6
56.4
75.0
93.6
110.4
n=350
22.1±8.1
43.3
VSD
35.8
53.3
72.8
92.3
109.9
n=117
23.5±9.3
37.6
ToF
42.8
56.4
71.6
86.7
100.4
n=1055
24.7±7.8
31.1
Atrial switch TGA
40.2
52.6
66.3
80.1
92.5
n=489
24.4±7.5
26.5
ccTGA
33.5
47.3
62.7
78.1
92.0
n=129
21.0±7.6
33.9
Ebstein
34.4
46.7
60.3
73.9
86.2
n=332
21.3±7.1
27.6
Fontan
37.9
48.0
59.3
70.5
80.6
n=911
22.8±6.4
20.0
Complex
25.3
35.3
46.3
57.4
67.3
n=105
15.7±5.6
32.0
Eisenmenger
25.5
33.6
42.7
51.7
59.8
n=119
12.5±3.9
40.3
20
40
60
80
100
120
pVO2 as % predicted


Physiological reserve/
functional capacity
Surgery
Independent
Dependent
Critical threshold
C
A
D
B
Time