European Heart Journal (2013) **34**, 3669–3674
doi:10.1093/eurheartj/eht433

CURRENT OPINION

Physical activity in adolescents and adults with congenital heart defects: individualized exercise prescription†

Werner Budts[1,2]*, Mats Börjesson[3], Massimo Chessa[4], Frank van Buuren[5], Pedro Trigo Trindade[6], Domenico Corrado[7], Hein Heidbuchel[1,2], Gary Webb[8], Johan Holm[9], and Michael Papadakis[10]

[1]Cardiovascular Diseases, University Hospitals Leuven, Leuven, Belgium; [2]Department of Cardiovascular Sciences, KU Leuven, Herestraat 49, B-3000 Leuven, Belgium; [3]Swedish School of Sports and Health Science, and Karolinska University Hospital, Stockholm, Sweden; [4]Pediatric and Adult Congenital Heart Centre, IRCCS—Policlinico San Donato San Donato Milanese, Milan, Italy; [5]Heartcenter of Northrhine Westphalia University of Bochum, Bad Oeynhausen, Germany; [6]Groupe Médical du Petit-Lancy, Petit-Lancy, Switzerland; [7]Department of Cardiac, Thoracic and Vascular Sciences, University of Padua Medical School, Padova, Italy; [8]Cincinnati Children's Hospital Heart Institute, Cincinnati, OH, USA; [9]GUCH Unit, Skåne University Hospital Heart failure and Valvular heart disease, Lund, Sweden; and [10]Cardiovascular Sciences Research Centre, St George's University of London, London, UK

Received 7 January 2013; revised 15 August 2013; accepted 2 September 2013; online publish-ahead-of-print 7 November 2013

Introduction

Studies in patients with congenital heart disease (CHD) indicate that the majority of individuals participating in such programs achieve significant improvement of their exercise capacity and psychological state.[1] The challenge is to ensure safe participation in regular physical activity (PA) in order to avoid the detrimental effects associated with sedentary life style.

Why are physical activity recommendations for adolescents and adults with congenital heart disease needed?

The improved surgical techniques and clinical care of children with CHD have led to a considerable *increase in the population of patients* with CHD who reach adulthood, and the number of adults is expected to grow at a rate of 5% per year.[2] Cardiac rehabilitation programs in patients with CHD have shown improvements in peak-VO_2.[3–5] Additionally, regular physical exercise is associated with lower risk of future obesity and ischaemic heart disease.[3] Paradoxically, only a minority of CHD patients (19%) receives formal PA advice,[4] and are often encouraged towards a sedentary lifestyle as a result of overprotection,[5] and uncertainty as to which physical activities and with what intensity should be recommended.[1] This is of particular importance when one considers that children with CHD are more likely to be overweight because of physical inactivity compared with children without CHD.[6] On the other end of the spectrum, young patients may reject exercise limitations and engage in unsafe sporting practices.

Existing exercise guidelines are of limited value for the majority of patients with CHD because they focus predominantly on competitive athletes.[7] Consequently, they cover less than 1% of the CHD population and applying these recommendations to leisure time activities would be too restrictive. In addition, in all current recommendations, the decision making process is primarily based on the individual anatomic lesions. As a result formulating recommendations becomes complex and the documents are long and impractical (*Figure 1*).[8]

Aim of the recommendations

The scope of this article is to produce concise and practical recommendations relating to PA for adolescents and adults with CHD. The novel approach is to formulate clinically useful recommendations based on haemodynamic and electrophysiological parameters, instead of focusing solely on specific defects. Although the recommendations are aimed at CHD specialists, it is important to acknowledge that CHD patients with mild disease are frequently managed by non-specialists.

Methodology

Nature of recommendations

The present document is a consensus of an international panel of cardiologists with expertise in CHD, sports cardiology, and exercise

†Position article from the Working Group of Grown Up Congenital Heart Disease and the Section of Sports Cardiology of the EACPR.

The opinions expressed in this article are not necessarily those of the Editors of the *European Heart Journal* or of the European Society of Cardiology.

* Corresponding author. Tel: +32 16 344369, Fax: +32 16 344240, Email: werner.budts@uzleuven.be

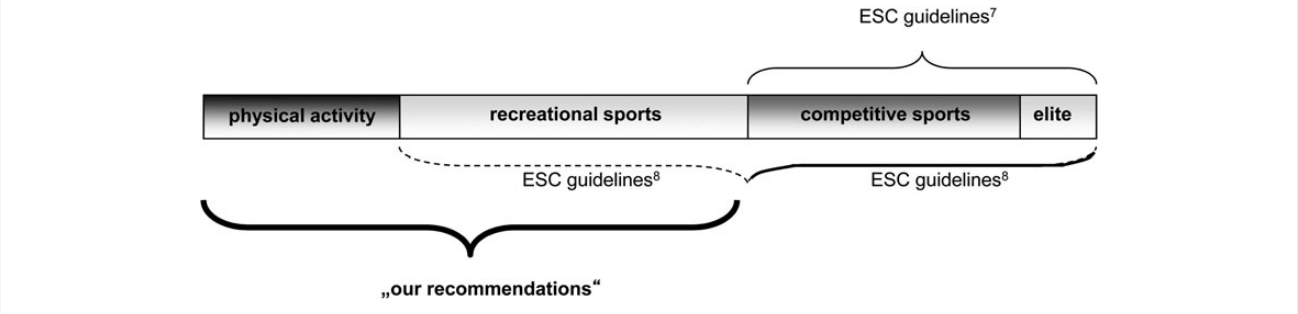

Figure 1 Available recommendations/position papers in patients with congenital heart disease.

physiology. In consideration of the scarcity of scientific data, the panel acknowledges the challenges in formulating arbitrary recommendations. Therefore, it is essential to inform patients of the potential uncertainties relating to the recommended exercise prescription regime.

Target patient population

Given that most of children with CHD have reached physical maturity by the age of 16 years, these recommendations apply to CHD patients ≥16 years. The authors concede, however, that the recommendations are not applicable to all patients with CHD and in particular not to patients with congenital rhythm or conduction disorders and with isolated congenital coronary artery anomalies.

Clinical approach to congenital heart disease

Emphasis is placed on associated haemodynamic and electrophysiological factors rather than on specific defects. All individuals are assessed by five specific parameters (*Figures 2* and *3*). This novel approach simplifies the evaluation process and at the same time allows for tailored advice since individuals with the same lesion may be affected to different degrees.

Relative intensity and classification of sports

Absolute intensity, expressed as the energy cost in metabolic equivalents[9], does not take into account the individual variations in cardiopulmonary capacity (fitness). Our exercise prescription utilizes *the* concept of relative intensity in order to provide individualized exercise prescription.

Sports are in general classified from low static to high static according to the estimated percentage of maximal voluntary contraction.[7] The static component of specific sports has been summarized and discussed by Mitchell *et al.*[10–12] For example, golf and long distance running have typical low static demands, whereas weightlifting, water skiing, and rowing have principally high static demands. However, this classification remains very arbitrary and needs interpreting with caution. Indeed, some sports involve heterogeneity with respect to static (and dynamic) cardiovascular demands in either different athletic disciplines—such as parallel bars and floor exercises in gymnastics or positions such as lineman and running back in football, or goalkeeper and mid-fielder in soccer.[12] The dynamic component in this article is replaced by *relative intensity*.

How to calculate training intensity?

Cardiopulmonary exercise testing (*CPET*) can provide parameters such as the maximal heart rate (MHR), heart rate reserve, maximal/peak oxygen consumption (peak-VO_2), first ventilatory anaerobic threshold (VAT) (also referred in the literature as the 'aerobic threshold' or 'first lactate turn point'), as well as the maximal workload.[13] To avoid

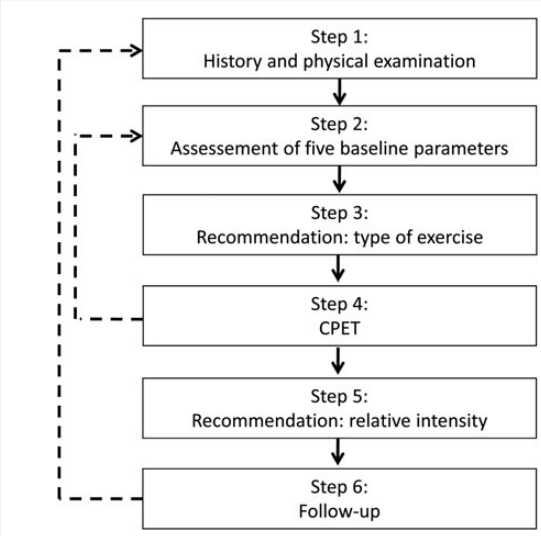

Figure 2 Flow chart depicting the six steps when evaluating adolescent and adult patients with congenital heart disease.

confusion, it is important to note that exercise is divided into three phases marked by two ventilatory thresholds; the first lactate threshold, also referred as aerobic threshold represents 40–60% of peak-VO_2 and the second lactate threshold, referred as the anaerobic threshold represents 60–90% of peak-VO_2.[14]

Objective CPET measurements allow correlation with subjective estimates such as the rate of perceived exertion (RPE) (Borg scale).[15] Individual RPE ratings correlate well with blood lactate levels and oxygen consumption.[16] Beside RPE, the authors recommend the use of a fixed percentage of the MHR which correlates well with RPE of the Borg scale (*Table 1*).[17] Monitoring of both the training heart rate and RPE is simple and can be performed by exercising individuals in the field without the need of complicated or expensive equipment. Training bursts could be allowed for patients with high intensity training, but is less applicable for patients in whom moderate or low intensity physical exercise is recommended. For CHD patients, who may have chronotropic incompetence or atrial fibrillation and in whom the use of a fixed percentage of MHR is not applicable, the exercise prescription could be based on maximal work load or VAT, as the preferred approximation of training heart rate.[9,15] However, not all patients will have a typical s-shaped heart response pattern in incremental exercise.[18–21] For

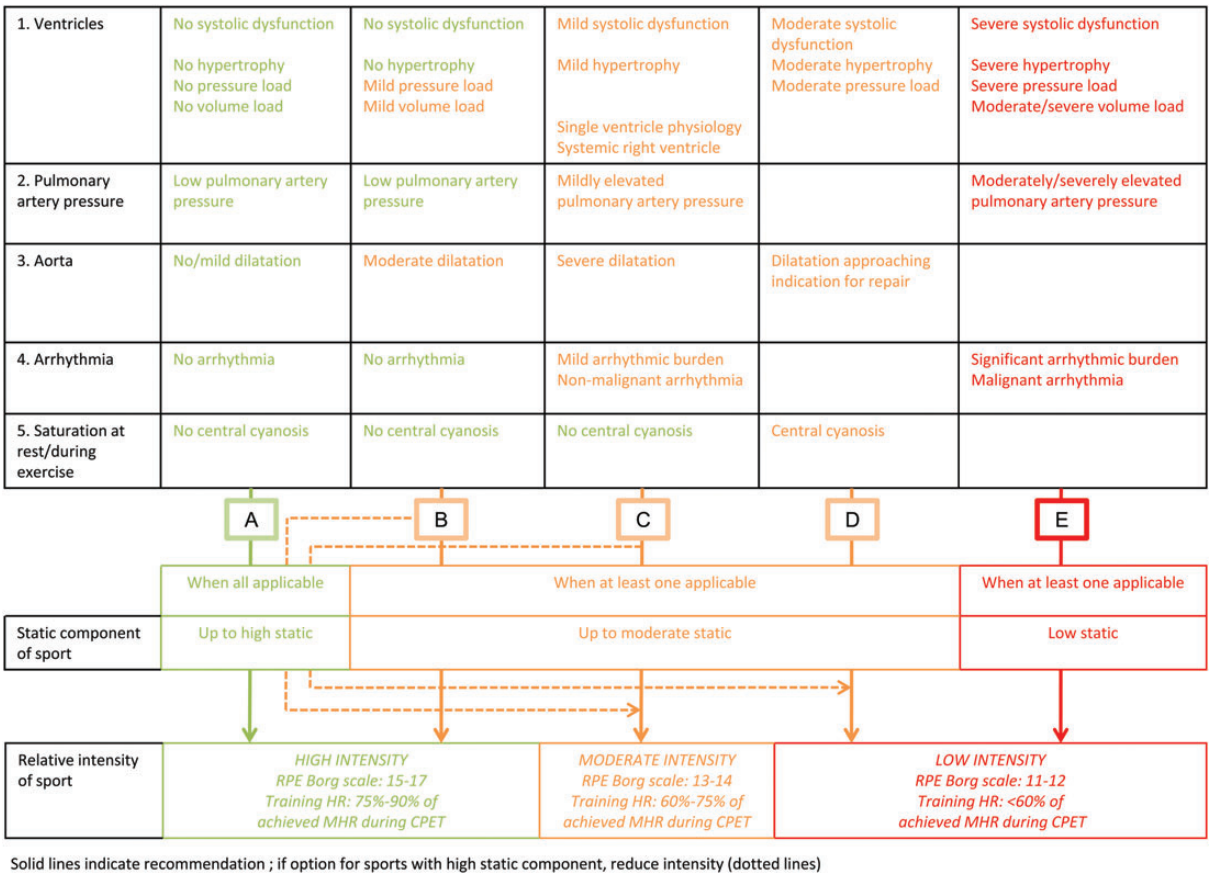

Figure 3 Flow chart depicting in detail steps 2–5. Following evaluation of the five variables and the interpretation of the CPET, an individualized recommendation can be provided (solid lines). When patients insist on sports with high static component, where it is not recommended, PA at a lower level intensity is suggested (dotted lines).

Table 1 The relation between rate of perceived exertion (Borg scale), % of maximal heart rate

RPE (Borg scale, ranging 6–20)	Subjective description of exercise intensity	Feels like	% of MHR
<10	Very light	Nothing	<35
10–11	Light	Something	35–54
12–13	Moderate	Perspiring	55–69
14–16	Hard	Sweating working	70–89
17–19	Very hard	Hard working	≥90
20	Maximal	Can't breathe anymore	100

RPE, rate of perceived exertion; MHR, maximal heart rate achieved during cardiopulmonary exercise testing.

individuals who are unable to monitor their heart rate, also the use of 'if you can talk while exercising' rule could be applied to approximate moderate relative exercise intensity.[22]

Parameters to evaluate prior to exercise prescription

Exercise is associated with an augmentation of the cardiac output, enhanced ventricular preload, and an increase in both the pulmonary and systemic arterial pressures.[23,24] Dynamic exercise imposes primarily a volume load, while static exercise produces primarily a pressure load. The physician needs to assess the potential haemodynamic repercussions, but also the potential risk for sudden cardiac death.

General recommendations

The recommended steps for comprehensive evaluation of patients with CHD in order to advise are outlined in *Figure 2*.

Step 1. History and physical examination

The physician should obtain a detailed medical and surgical history and a precise account of on-going symptoms and perform a detailed physical examination. Knowledge of the type of CHD raises awareness of potential haemodynamic and electrophysiological complications.

Table 2 Definition of variables

Variable	Definition
Ventricles	
Ventricular dysfunction	No: EF ≥55% Mild: 45% ≤EF <55% (or normal systemic right ventricle) Moderate: 30 ≤EF <45% Severe: EF <30% (or impaired systemic right ventricle)
Ventricular hypertrophy	Left ventricle: No: septal/posterior wall thickness (cm): ♂ <1.1 ♀ <1.0; LV mass (g): ♂ 88–224 ♀ 67–162 Mild: septal/posterior wall thickness (cm): ♂ 1.1–1.3 ♀ 1.0–1.2; LV mass (g): ♂ 225–258 ♀ 163–186 Moderate: septal/posterior wall thickness (cm): ♂ 1.4–1.6 ♀ 1.3–1.5; LV mass (g): ♂ 259–292 ♀ 187–210 Severe: septal/posterior wall thickness (cm): ♂ ≥1.7 ♀ ≥1.6; LV mass (g): ♂ ≥293 ♀ ≥211 Right ventricle: qualitative echocardiographic evaluation
Ventricular pressure overload	
• No pressure overload	No significant LVOT or RVOT gradient (peak systolic flow <2.6 m/s), no obstruction in great vessels
• Mild pressure overload	2.6 m/s ≤peak systolic velocity <3 m/s for LVOT and RVOT obstructions and PPS; for coarctation of the aorta, arm-leg gradient <20 mmHg
• Moderate overload	3 m/s ≤peak systolic velocity ≤4 m/s for LVOT and RVOT obstructions and PPS
• Severe pressure overload	Peak systolic velocity >4 m/s for LVOT and RVOT obstructions and PPS; for coarctation of the aorta, clinical gradient ≥20 mmHg
Ventricular volume overload	
• No volume overload	Absent/mild valve regurgitation or shunt that do not cause significant chamber dilatation (parasternal views—long axis: LVEDD: 55–63 mm; LVESD 35–42 mm; RVEDD: 30–36 mm)
• Mild volume overload	Mild: dilated right or left ventricle by severe regurgitation, however with preserved systolic function
• Moderate/severe volume overload	Significant right or left ventricular dilatation with impaired ventricular function
Ventricle physiology	Single ventricle or double ventricle Systemic left ventricle or systemic right ventricle
Pulmonary artery pressure	
• Low PAP	No PH: TVRV ≤2.8 m/s, systolic PAP ≤36 mmHg, and/or no additional echocardiographic variables suggestive of PH
• Mildly elevated PAP	Possible PH: TVRV >2.8 m/s, systolic PAP >36 mmHg, and no signs of right ventricular systolic dysfunction
• Moderately/severely elevated PAP	High probability of PH: TVRV >2.8 m/s, systolic PAP >36 mmHg, and signs of right ventricular dysfunction
Aorta	
• No/mild dilatation	Normal (≤30 mm) or borderline sizes (<35 mm) of the aorta
• Moderate dilatation	Aorta size ≥35 and <45 mm
• Severe dilatation	Aorta size ≥45 and <50 mm
• Dilatation approaching indication for repair	Aorta size ≥50 mm
Arrhythmia	
• No arrhythmias	Absence of/infrequent arrhythmias (<500/24 h) PVC if a Holter was done
• Mild arrhythmic burden/non-malignant arrhythmias	Frequent/coupled PVC and controlled atrial fibrillation/atrial flutter, which do not worsen with exercise
• Significant arrhythmic burden/potentially malignant arrhythmias	Atrial fibrillation/atrial flutter, which worsen with exercise Non-sustained ventricular arrhythmias or sustained ventricular tachycardia
Saturation at rest/during exercise	
• No central cyanosis	Absence of clinical signs; transcutaneous saturations within the range of 96–100%, at rest and during exercise

EF, ejection fraction; EDD, end-diastolic diameter; LVOT, left ventricle outflow tract; RVOT, right ventricle outflow tract; PIG, peak instantaneous gradient; PPS, peripheral pulmonary stenosis; LVEDD, left ventricle end-diastolic diameter; LVESD, left ventricle end-systolic diameter; RVEDD, right ventricle end-diastolic diameter; TVRV, tricuspid valve regurgitation velocity; PAP, pulmonary artery pressure; PVC, premature ventricular complex.

Step 2. Assessment of five baseline parameters

(1) Assessment of ventricular function:

A transthoracic echocardiogram is usually sufficient to evaluate left ventricular function. For the morphological right ventricle or univentricular heart physiology additional imaging techniques may be required, such as magnetic resonance imaging (MRI). For cut-off values, we refer to published

echocardiographic diagnostic criteria (*Table 2*).[25–27] Such values represent a generic guide rather than absolute criteria, since the authors recognize that individual centres may utilize different indices. Interpretation of echocardiographic values should always take into consideration the individual's demographics and background exercise level.[28]

(2) Assessment of pulmonary artery pressure (PAP):

Pulmonary artery pressure is obtained by the tricuspid valve regurgitation velocities (TVRV). We suggest using the diagnostic criteria of Galie *et al.*,[29] where pulmonary hypertension (PH) is considered to be excluded when TVRV $\leq$ 2.8 m/s, and no additional echocardiographic variables suggestive of PH are present. In case of doubt, right heart catheterization should be performed.[29]

(3) Assessment of the aorta:

Although aortic diameter should ideally be indexed, for simplification purposes and in line with recent guidelines for the diagnosis and management of patients with thoracic aortic disease,[30] the authors suggest to use absolute numbers. Inadequate measurement of the aortic diameter by echocardiography may require additional imaging by computerized tomography or MRI.

(4) Assessment of arrhythmia:

The evaluation of CHD patients with arrhythmias includes personal history inquiring for the presence of palpitations, presyncope or syncope, and unexplained weakness. Besides a 12-lead ECG, the evaluation may include 24-h Holter monitoring. When initial testing fails to demonstrate any arrhythmia, further investigations may be required. *Table 2* summarizes the degree of severity in arrhythmia.

(5) Assessment of saturation at rest/during exercise:

In patients with a potential right-to-left shunt, transcutaneous arterial saturation at rest should be recorded. Central cyanosis is largely excluded when transcutaneous saturations are $>$95%, at rest, and during exercise (*Table 2*). Low arterial saturation due to pulmonary disease must not be forgotten. Evaluation with spirometry should be included in patients with reduced arterial saturation without a cardiovascular explanation.

Step 3. Decision on type of exercise

The physician should assess each parameter (1–5) in turn, assign the individual patient in a specific route, and decide on the static component of the exercise (*Figure 3*). When at least one of the parameters is outside the conventional normal limits, moderate or low statics sport is recommended (solid lines, *Figure 3*). This does not necessarily exclude sports with a high static component, but it is then recommended to exercise at reduced intensity, as discussed in step 5 (dotted lines, *Figure 3*).

Step 4. Cardiopulmonary exercise testing

The results of the exercise test will help determine the relative intensity. When CPET is not available, regular exercise testing without measurement of expired air might be an alternative. The following parameters need extra attention for individualized advice.

(1) Peak VO_2, MHR, Borg scale:

The fitness levels of patients with CHD are decreased when compared with normal subjects, as shown by a lower peak VO_2.[31] Importantly, VO_2 max has been found to be one of the best predictors of morbidity and mortality in patients with CHD.[32–34] The Borg scale will assist the patient to get a feeling as at what level he/she should exercise.

(2) Transcutaneous saturation or arterial blood gasses:

When desaturation is suspected during PA, continuous measurement of saturation is needed when performing CPET. If desaturation is present during exercise, the patient has to re-enter the algorithm at step 2 (*Figure 2*).

(3) Detection of arrhythmia or conduction disorder during exercise:

Recently, Koyak *et al.*[35] demonstrated that exercise-induced arrhythmias increase the risk of sudden death by 6.6-fold. In case of the detection of arrhythmia, the patient has to re-enter the algorithm at step 2 (*Figure 2*). When treatment is established, repeat assessment of all five variables is recommended.

(4) Blood pressure response to exercise:

A normal blood pressure response during exercise includes a rise in systolic blood pressure by $\geq$25 mmHg. Diastolic blood pressure is usually associated with a small drop[36] and an increase of $>$10 mmHg during exercise or recovery is abnormal. Individual centres may use their own criteria to determine 'abnormal blood pressure response'.

Step 5. Recommendation on relative intensity

After interpretation of the CPET, an individualized recommendation can be provided (*Figure 3*). In the interest of encouraging participation in regular exercise of as many individuals with CHD as possible, the authors wish to provide patients with a wide range of sporting disciplines. As such, we accept that some patients may wish to participate in PA classified as high static, despite a recommendation for a medium or low static activity. In these cases, we suggest a compromise in the relative intensity of exercise, starting at a lower intensity level, compared with the recommended one. However, some types of sports cannot be practiced at reduced intensity and then preferred to be discouraged.

For the frequency and duration of each exercise session, a combined minimum of 3–4.5 h of physical activities per week is recommended; the minimum minutes per session should be 30 min (for daily or almost daily PA). Although no data are available in the literature for CHD patients, we hypothesize that habitual training will be necessary to achieve a long-term clinical benefit. For team sports, the intensity of PA is difficult to restrict for the individual. Patients should be encouraged to participate in teams (or with peers) of similar physical fitness, i.e. when someone want to play football, he plays with colleagues some of which may be a bit more or less physically fit compared with the patient. That is where support groups may also come handy. Patients should monitor their symptoms (Borg scale) and heart rate to ensure they do not exceed the recommendations, at least for prolonged period of times, particularly for start-stop sports like football. Individuals who feel that their fitness is lower than that of their peers should be supported to either find an alternative team or sport.

Step 6. Follow-up

New symptoms should prompt discontinuation of the patient's exercise regime and re-evaluation. For physical activities of medium and low intensity, we recommend follow-up as frequently as suggested by the ESC GUCH guidelines.[7,37] In patients with regular high intensity PA, follow-up is recommended by the ESC guidelines for competitive and elite sports.[7] We recommend going through the PA algorithm at each routine visit. As the performance of the patient changes, the degree of PA should be adapted.

Specific recommendations

Individuals with permanent pacemaker or resynchronization therapy

Pacemaker therapy *per se* does not preclude high intensity PA. Modern *pacemakers* provide bi-ventricular synchronization to optimize systemic ventricular ejection. Implantation of the leads is often performed epicardially during surgical corrective interventions, allowing for *optimal positioning* on both ventricles to achieve adequate resynchronization. *Programming of the pacemaker* needs to assure appropriate rate adaptation during exercise. Persistent sinus rhythm can be used for tracking. Minute ventilation-based rate-response systems may be 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. It should be emphasized, however, that chronotropic incompetence in CHD patients is often a symptom of ventricular dysfunction or ischaemia, and is difficult to adjust by rate responsive pacing. This was highlighted by the study of Uebing *et al.*[39] where rate responsive pacing in patients with a systemic right ventricle did not improve either the right ventricular haemodynamics or exercise capacity.

Individuals with implantable cardioverter defibrillator

For patients with an *implantable cardioverter defibrillator (ICD),* only low to moderate intensity PA is recommended.[40] It remains unclear how effective cardiac defibrillation is during peak PA and the ICD cannot be regarded as a substitute for activity restraints based on the underlying cardiovascular disorder. A recently presented US-EU international registry (Boston, Heart Rhythm Society Meeting, 2012) showed that fatalities in ICD recipients, who continued to perform competitive or high-intensity sports, were rare. There was also *no report of increased lead malfunctions*. Nevertheless, the Registry confirmed that *arrhythmias were more frequent during PA* and there was a clear increase of *inappropriate shocks during sports*. Finally, *the risk for damage* to the device through body movements or contact needs to be considered.

Limitations

These recommendations aim to provide the platform for more uniform advice between health care professionals in order to avoid confusion between non-expert physicians and patients, alike. They are, however, based on limited clinical evidence, underscoring the urgent need for physiologic studies regarding mechanisms of central and peripheral effects of various forms of exercise training in specific populations, as well as the acute and chronic benefits and risks of different training regimes. It would therefore be prudent to apply this document with caution and tailor advice to individual patients. Additionally, because of the variety of CHD with different residua and sequelae, not all patients will fit into the algorithm. Therefore, the physician has to take into consideration the individual anatomical lesion in order to assess potential complications at an early stage.

Conclusion

We envisage that our recommendations will result in more unified protocols throughout Europe and offer the required reassurance to both patients and physicians who manage patients with adult CHD to encourage these patients to enjoy a physically active life style. Such an individualized exercise prescription might offer a platform with the aim to maximize benefit for health and minimize the cardiovascular risk. Finally, it could be used as a tool for future research to increase the level of evidence.

Funding

The authors received a writing grant from the European Society of Cardiology.

Conflict of interest: none declared.

References

1. Dua JS, Cooper AR, Fox KR, Graham Stuart A. Exercise training in adults with congenital heart disease: feasibility and benefits. *Int J Cardiol* 2010;**138**:196–205.
2. Deanfield J, Thaulow E, Warnes C, Webb G, Kolbel F, Hoffman A, Sorenson K, Kaemmer H, Thilen U, Bink-Boelkens M, Iserin L, Daliento L, Silove E, Redington A, Vouhe P, Priori S, Alonso MA, Blanc JJ, Budaj A, Cowie M, Deckers J, Fernandez Burgos E, Lekakis J, Lindahl B, Mazzotta G, Morais J, Oto A, Smiseth O, Trappe HJ, Klein W, Blomstrom-Lundqvist C, de Backer G, Hradec J, Mazzotta G, Parkhomenko A, Presbitero P, Torbicki A. Management of grown up congenital heart disease. *Eur Heart J* 2003;**24**:1035–1084.
3. Ten Harkel AD, Takken T. Exercise testing and prescription in patients with congenital heart disease. *Int J Pediatr* 2010;**2010**:791980.
4. Lunt D, Briffa T, Briffa NK, Ramsay J. Physical activity levels of adolescents with congenital heart disease. *Aust J Physiother* 2003;**49**:43–50.
5. Reybrouck T, Mertens L. Physical performance and physical activity in grown-up congenital heart disease. *Eur J Cardiovasc Prev Rehabil* 2005;**12**:498–502.
6. Pinto NM, Marino BS, Wernovsky G, de Ferranti SD, Walsh AZ, Laronde M, Hyland K, Dunn SO Jr, Cohen MS. Obesity is a common comorbidity in children with congenital and acquired heart disease. *Pediatrics* 2007;**120**:e1157–e1164.
7. Pelliccia A, Fagard R, Bjornstad HH, Anastassakis A, Arbustini E, Assanelli D, Biffi A, Borjesson M, Carre F, Corrado D, Delise P, Dorwarth U, Hirth A, Heidbuchel H, Hoffmann E, Mellwig KP, Panhuyzen-Goedkoop N, Pisani A, Solberg EE, van-Buuren F, Vanhees L, Blomstrom-Lundqvist C, Deligiannis A, Dugmore D, Glikson M, Hoff PI, Hoffmann A, Hoffmann E, Horstkotte D, Nordrehaug JE, Oudhof J, McKenna WJ, Penco M, Priori S, Reybrouck T, Senden J, Spataro A, Thiene G. Recommendations for competitive sports participation in athletes with cardiovascular disease: a consensus document from the Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. *Eur Heart J* 2005;**26**:1422–1445.
8. Takken T, Giardini A, Reybrouck T, Gewillig M, Hovels-Gurich HH, Longmuir PE, McCrindle BW, Paridon SM, Hager A. Recommendations for physical activity, recreation sport, and exercise training in paediatric patients with congenital heart disease: a report from the Exercise, Basic & Translational Research Section of the European Association of Cardiovascular Prevention and Rehabilitation, the European Congenital Heart and Lung Exercise Group, and the Association for European Paediatric Cardiology. *Eur J Prevent Cardiol* 2012;**19**:1034–1065.

9. Vanhees L, Rauch B, Piepoli M, vanBUuren F, Takken T, Borjesson M, Bjarnason-Wehrens B, Doherty P, Dugmore D, Halle M. Importance of characteristics and modalities of physical activity and exercise in the management of cardiovascular health in individuals with cardiovascular disease (part iii). *Eur J Prev Cardiol* 2012; epub ahead of publication.
10. Mitchell JH, Maron BJ, Epstein SE. 16th Bethesda Conference: cardiovascular abnormalities in the athlete: recommendations regarding eligibility for competition. October 3–5, 1984. *J Am Coll Cardiol* 1985;**6**:1186–1232.
11. Mitchell JH, Haskell WL, Raven PB. Classification of sports. *J Am Coll Cardiol* 1994;**24**: 864–866.
12. Mitchell JH, Haskell W, Snell P, Van Camp SP. Task force 8: classification of sports. *J Am Coll Cardiol* 2005;**45**:1364–1367.
13. Mezzani A, Agostini P, Cohen-Solal A, Corra U, Jegier A, Kouidi E *et al.* Standards for the use of cardiopulmonary exercise testing for the functional evaluation of cardiac patients: a report from the exercise physiology section of the European association for cardiovascular prevention and rehabilitation. *Eur J Cardiovasc Prev Rehabil* 2009; **16**:249–267.
14. Binder RK, Wonisch M, Corra U, Cohen-Solal A, Vanhees L, Saner H, Schmid JP. Methodological approach to the first and second lactate threshold in incremental cardiopulmonary exercise testing. *Eur J Cardiovasc Prev Rehabil* 2008;**15**:726–734.
15. Borg G. Perceived exertion as an indicator of somatic stress. *Scand J Rehabil Med* 1970;**2**:92–98.
16. Stoudemire NM, Wideman L, Pass KA, McGinnes CL, Gaesser GA, Weltman A. The validity of regulating blood lactate concentration during running by ratings of perceived exertion. *Med Sci Sports Exerc* 1996;**28**:490–495.
17. Nieman DC. Exercise prescription. *Exercise Testing and Prescription*. New York: McGraw-Hill; 2003, p230–279.
18. Hofmann P, Von Duvillard SP, Seibert FJ, Pokan R, Wonisch M, Lemura LM, Schwaberger G. %HRmax target heart rate is dependent on heart rate performance curve deflection. *Med Sci Sports Exerc* 2001;**33**:1726–1731.
19. Pokan R, Hofmann P, Von Duvillard SP, Schumacher M, Gasser R, Zweiker R, Fruhwald FM, Eber B, Smekal G, Bachl N, Schmid P. Parasympathetic receptor blockade and the heart rate performance curve. *Med Sci Sports Exerc* 1998;**30**:229–233.
20. Tabet JY, Meurin P, Ben Driss A, Thabut G, Weber H, Renaud N, Odjinkem N, Solal AC. Determination of exercise training heart rate in patients on beta-blockers after myocardial infarction. *Eur J Cardiovasc Prev Rehabil* 2006;**13**:538–543.
21. Koike A, Itoh H, Taniguchi K, Hiroe M. Detecting abnormalities in left ventricular function during exercise by respiratory measurement. *Circulation* 1989;**80**: 1737–1746.
22. Vanhees L, Stevens A. Exercise intensity: a matter of measuring or of talking?. *J Cardiopulm Rehabil* 2006;**26**:78–79.
23. Stickland MK, Welsh RC, Petersen SR, Tyberg JV, Anderson WD, Jones RL, Taylor DA, Bouffard M, Haykowsky MJ. Does fitness level modulate the cardiovascular hemodynamic response to exercise?. *J Appl Physiol* 2006;**100**:1895–1901.
24. Tolle JJ, Waxman AB, Van Horn TL, Pappagianopoulos PP, Systrom DM. Exercise-induced pulmonary arterial hypertension. *Circulation* 2008;**118**: 2183–2189.
25. Baumgartner H, Hung J, Bermejo J, Chambers JB, Evangelista A, Griffin BP, Iung B, Otto CM, Pellikka PA, Quinones M. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. *Eur J Echocardiogr* 2009;**10**:1–25.
26. Lang RM, Bierig M, Devereux RB, Flachskampf FA, Foster E, Pellikka PA, Picard MH, Roman MJ, Seward J, Shanewise J, Solomon S, Spencer KT, St John Sutton M, Stewart W. Recommendations for chamber quantification. *Eur J Echocardiogr* 2006;**7**:79–108.
27. Vahanian A, Alfieri O, Andreotti F, Antunes MJ, Baron-Esquivias G, Baumgartner H, Borger MA, Carrel TP, De Bonis M, Evangelista A, Falk V, Iung B, Lancellotti P, Pierard L, Price S, Schafers HJ, Schuler G, Stepinska J, Swedberg K, Takkenberg J, Von Oppell UO, Windecker S, Zamorano JL, Zembala M. Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 2012;**33**:2451–2496.
28. Papadakis M, Carre F, Kervio G, Rawlins J, Panoulas VF, Chandra N, Basavarajaiah S, Carby L, Fonseca T, Sharma S. The prevalence, distribution, and clinical outcomes of electrocardiographic repolarization patterns in male athletes of African/Afro-Caribbean origin. *Eur Heart J* 2011;**32**:2304–2313.
29. Galie N, Hoeper MM, Humbert M, Torbicki A, Vachiery JL, Barbera JA, Beghetti M, Corris P, Gaine S, Gibbs JS, Gomez-Sanchez MA, Jondeau G, Klepetko W, Opitz C, Peacock A, Rubin L, Zellweger M, Simonneau G. Guidelines for the diagnosis and treatment of pulmonary hypertension: the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). *Eur Heart J* 2009;**30**:2493–2537.
30. Hiratzka LF, Bakris GL, Beckman JA, Bersin RM, Carr VF, Casey DE Jr., Eagle KA, Hermann LK, Isselbacher EM, Kazerooni EA, Kouchoukos NT, Lytle BW, Milewicz DM, Reich DL, Sen S, Shinn JA, Svensson LG, Williams DM. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with Thoracic Aortic Disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. *Circulation* 2010;**121**:e266–e369.
31. Fredriksen PM, Ingjer F, Nystad W, Thaulow E. A comparison of VO2(peak) between patients with congenital heart disease and healthy subjects, all aged 8–17 years. *Eur J Appl Physiol Occup Physiol* 1999;**80**:409–416.
32. Giardini A, Specchia S, Tacy TA, Coutsoumbas G, Gargiulo G, Donti A, Formigari R, Bonvicini M, Picchio FM. Usefulness of cardiopulmonary exercise to predict long-term prognosis in adults with repaired tetralogy of fallot. *Am J Cardiol* 2007;**99**: 1462–1467.
33. Giardini A, Hager A, Lammers AE, Derrick G, Muller J, Diller GP, Dimopoulos K, Odendaal D, Gargiulo G, Picchio FM, Gatzoulis MA. Ventilatory efficiency and aerobic capacity predict event-free survival in adults with atrial repair for complete transposition of the great arteries. *J Am Coll Cardiol* 2009;**53**:1548–1555.
34. Inuzuka R, Diller GP, Borgia F, Benson L, Tay EL, Alonso-Gonzalez R, Silva M, Charalambides M, Swan L, Dimopoulos K, Gatzoulis MA. Comprehensive use of cardiopulmonary exercise testing identifies adults with congenital heart disease at increased mortality risk in the medium term. *Circulation* 2012;**125**:250–259.
35. Koyak Z, Harris L, de Groot JR, Silversides CK, Oechslin EN, Bouma BJ, Budts W, Zwinderman AH, Van Gelder IC, Mulder BJ. Sudden cardiac death in adult congenital heart disease. *Circulation* 2012;**126**:1944–1954.
36. Daida H, Allison TG, Squires RW, Miller TD, Gau GT. Peak exercise blood pressure stratified by age and gender in apparently healthy subjects. *Mayo Clin Proc* 1996;**71**: 445–452.
37. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJ, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E. ESC guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J* 2010;**31**:2915–2957.
38. Alt E, Combs W, Willhaus R, Condie C, Bambl E, Fotuhi P, Pache J, Schomig A. A comparative study of activity and dual sensor: activity and minute ventilation pacing responses to ascending and descending stairs. *Pacing Clin Electrophysiol* 1998;**21**: 1862–1868.
39. Uebing A, Diller GP, Li W, Maskell M, Dimopoulos K, Gatzoulis MA. Optimised rate-responsive pacing does not improve either right ventricular haemodynamics or exercise capacity in adults with a systemic right ventricle. *Cardiol Young* 2010;**20**: 485–494.
40. Heidbuchel H, Corrado D, Biffi A, Hoffmann E, Panhuyzen-Goedkoop N, Hoogsteen J, Delise P, Hoff PI, Pelliccia A. Recommendations for participation in leisure-time physical activity and competitive sports of patients with arrhythmias and potentially arrhythmogenic conditions. Part II: ventricular arrhythmias, channelopathies and implantable defibrillators. *Eur J Cardiovasc Prev Rehabil* 2006;**13**: 676–686.