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Abstract

All children have a natural need to move, play, and perform activities. Physical activity is necessary for optimal physical, emotional, and psychosocial development for healthy children as well as children with congenital heart disease (CHD). In this paper we provide recommendations for physical activity, recreational sport, and exercise training in children and adolescents with CHD. In general, children with CHD should be advised to comply with public health recommendations of daily participation in 60 min or more of moderate-to-vigorous physical activity that is developmentally appropriate and enjoyable and involves a variety of activities. While all patients with CHD can participate and benefit from physical activity and exercise, those with specific lesions or complications may require counselling regarding precautions and recommendations.

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Introduction

All children have a natural need to move, play, and perform activities. Physical activity is necessary for the optimal physical, emotional, and psychosocial development of healthy children as well as children with congenital heart disease (CHD). However, many parents, patients, caregivers, educators, physicians, and other health professionals have questions regarding appropriate levels of physical activity for children and adolescents with CHD.

Recent recommendations for sport participation in patients with CHD^{1–3} have focused mainly on eligibility criteria for competitive sports. The current recommendations will extend to leisure sports, physical activity, and even exercise training programmes with the focus on promotion of physical activity and exercise, while identifying circumstances for precautions and specific guidance and counselling. It will provide recommendations for clinical practice.

Defining physical activity and exercise

Many different types of physical activity and exercise can be performed in many different settings. Table 1 first provides definitions for the different types of

exercise (physical activity, leisure sports, competitive sports, and exercise training).

Physical performance depends on five basic functions: strength, endurance, skills/coordination, speed, and flexibility. Regular exercise can improve all of them. As strength and endurance are more closely related to the cardiovascular system, this paper mainly focuses on these two functions.

All physical activities can be characterized according to their static (need for strength) and dynamic (need for endurance) components, as reviewed by Mitchell et al.⁴ and shown in Figure 1.

In addition to the static/dynamic classification, some physical activities might be associated with increased risk of injury if loss of consciousness occurs (like swimming or horse riding) and some other of physical activities have a high possibility of body collision or trauma that could have serious consequences, like severe bleeding, aortic dissection, device damage, or embolization.

Changing physical fitness

In contrast to physical activity, physical fitness is the ability of a participant to perform physical activity and exercise. It depends on habitual variables like the amount of regular physical activity, but also on

Table 1. Definitions of exercise terminology

Term	Definition
Physical activity	Any active bodily movements resulting from muscle contraction that increases the metabolic rate above the resting level. In general vigorous activity is defined as activities that increase metabolic rate to more than six times the resting rate [>6 metabolic equivalent of task (MET)]. Moderate activity is defined as an activity within 3–6 MET.
Leisure sport	Recreational physical activities without pressure to play, continue to play, or play at a higher intensity than is desired by the participant. The activity or sport may be organized or informal, and may be ad hoc or structured for competition between participants or teams. However, any participant may stop participating or may lower the intensity of their participation at any time, without perceived pressure or coercion from self or others to continue.
Competitive sport	Organized, competitive, and skilful physical activities inside fixed rules of commitments and fair play. It involves pressure to train or play, or continue to train/play, at a high intensity regardless of whether that intensity is desired by or recommended for the participant. The source of the coercion or pressure may be from the participant, other participants, coaches, or spectators.
Exercise training	Specialized, planned methods and programmes of physical activity used to increase a subject's physical activity capacity (habilitation or rehabilitation), performance, or fitness. The exercise training may or may not occur under the supervision of an exercise professional.
Sedentary lifestyle	A lifestyle with no or marginally regular physical activity or exercise. It is usually characterized by excessive time spent in sedentary pursuits, such as watching TV, using the computer, and playing video games.

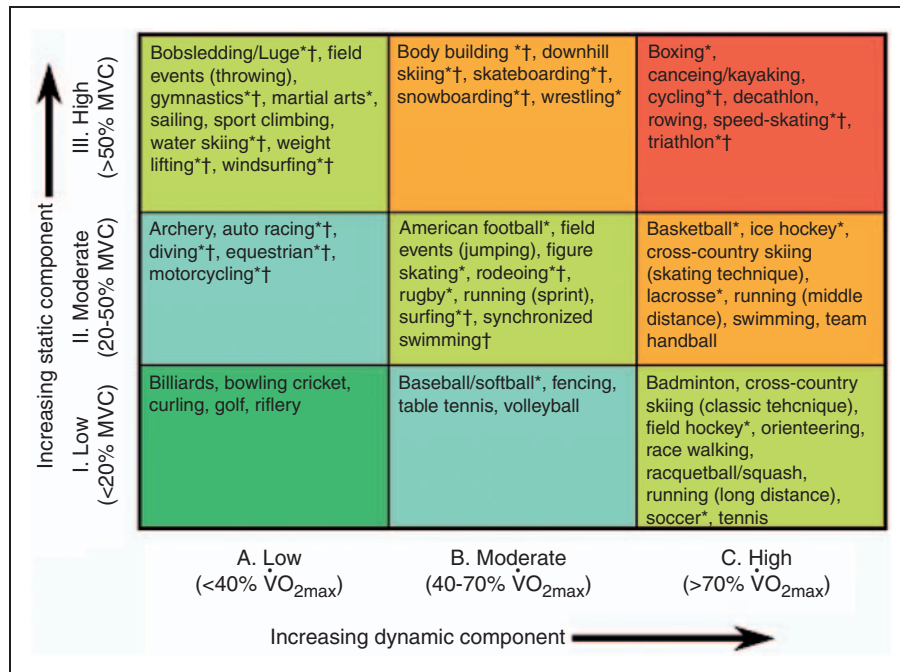


Figure 1. Classification of organized sports and exercise. This classification is based on peak static and dynamic components achieved during participation. It should be noted, however, that higher values may be reached during training or competition. The increasing dynamic component is defined in terms of the estimated percentage of maximal oxygen uptake ($\dot{V}O_{2max}$) achieved and results in increasing cardiac output. The increasing static component is related to the estimated percentage of maximal voluntary contraction (MVC) reached and results in increasing blood pressure load. The lowest total cardiovascular demands (cardiac output and blood pressure) are shown in green and the highest in red. Blue, yellow, and orange depict low moderate, moderate, and high moderate total cardiovascular demands. *Risk of bodily collision. †Increased risk if syncope occurs. (From Mitchell et al.,⁴ reproduced with permission from Elsevier).

Table 2. Classification of exercise training intensity concerning endurance exercise based on maximal oxygen uptake ($\dot{V}O_{2max}$), heart rate (HR) reserve, or maximal heart rate in children

Intensity	% $\dot{V}O_{2max}$	% HR _{res}	% HR _{max}	Type
Very light	<20	<29	<54	Active recovery training
Light	20–39	29	54	Rehabilitative training
Moderate	40–59	47	66	Endurance training
Heavy	60–84	65	77	High-intensity endurance training
Very heavy	>85	>87	>91	Interval training

Modified from Pollock et al.⁵ Carvalho and Mezzani,⁶ Hui and Chan.⁷

bodily variables like overweight or chronic disease state, as well as on genetic components.

Endurance exercise is the most common way to improve cardiovascular fitness. The benefits of exercise are dependent on the dose of exercise. The dose of exercise can be described using the so-called FITT factors, where FITT stands for Frequency, Intensity, Time, and Type of activity. In addition, the duration of a programme (week/months) is an important factor.

The intensity of an endurance exercise programme is usually described based on the percentage of a

participant's maximal oxygen uptake capacity or oxygen consumption ($\dot{V}O_{2max}$). In Table 2, the intensity levels for endurance exercise ranging from very light to very heavy are described, as well as the corresponding percentage of heart rate reserve and maximal heart rate.^{5–7}

For muscle strength or resistance exercise, the intensity is classified as the ratio between muscle contraction and maximum voluntary contraction (MVC), as shown in Table 3. Strength exercise with a low intensity will not result in an improvement in cardiovascular fitness

($\dot{V}O_{2max}$) or muscle strength (MVC). Training with a very high intensity increases the risk of injury, especially in children.

Training models for strength and endurance training in childhood are quite similar to those in adults. However, they should be considered more a theoretical construct than a real training programme, as children often lose interest in strict training plans or overly structured activities and will not participate.

In general, all exercise training and sport sessions should start with a 10–15-min dynamic warm-up period followed by 20–60 min of exercise training. Finally, a 10-min cool-down period with less intensive activities and stretching should end the exercise training session. Between the training sessions there must be enough time to recover (12–48 h depending on intensity and condition of the child).

Particularly in children, it is important to take into account the age of the target group when the aim is to establish an exercise training programme or recommendations for physical activity, exercise, or the promotion of sport participation. In preschool children, the

development of skills and even difficult coordinative tasks is of utmost importance. Prepubertal children are usually bored by 'adult type' endurance training (e.g. stationary cycling) with continuous intensity. Strength training in prepubertal children is only recommended under the close supervision of a conscientious trainer or paediatric physical therapist and should be mainly aimed at improving skills and technique and not at lifting high weights.⁸ A playful mixture of different activity tasks focusing mainly on skills/coordination, but also on speed and carefully on flexibility and strength, is the goal in this age group. Training effects in the prepubertal age group can predominantly be found in improved efficiency of the muscles and improved neuronal control of the muscle and not in an increase in muscle mass. In prepubertal children, muscle growth is limited by low levels of sex hormones. Moreover, it is during this period that physical activity habits for later life are established.

For pubertal children, other components of exercise play a more important role. Social components like integration into a team or compliance to fixed rules are important. After puberty, training can be similar to adults', focusing especially on strength (for muscle mass) and endurance (for aerobic capacity), but not forgetting the already-mentioned and very important other aspects of sport. For all age groups, it is one of the most essential tasks of parents, teachers, and trainers to avoid a sedentary life style during childhood and to convince the child that participation in physical activity is fun.

The general recommendations following the FITT principle for physical activity participation and exercise training in healthy children and adolescents are shown in Table 4. Interval training can be used alternatively with aerobic training in healthy children.⁹

Table 3. Classification of exercise training intensity concerning strength exercise based on maximal voluntary contraction (MVC, one repetition maximum)

Intensity	% MVC	Effects on strength
Low	<20	Virtually none
Moderate	20–50	Rehabilitative
High	50–70	Optimal to increase muscle strength
Very high	>70	Optimal to increase muscle mass

Table 4. General recommendations following the FITT principle for physical activity participation and exercise training in healthy children and adolescents

FITT	Cardiovascular (aerobic) training	Interval training	Muscle strength (resistance) training
Frequency	≥3 times/week	≥3 times/week	2–3 times/week
Intensity	Moderate-to-heavy exercise ($\dot{V}O_{2peak}$ 40–85%)	3–5 min of light-to-moderate baseline exercise ($\dot{V}O_{2peak}$ 20 to 59%) interrupted 6–8 times by 1–3 min bouts of very intense exercise ($\dot{V}O_{2peak}$ >85%)	High (50–70% MVC)
Time	20–60 min	In total 20–60 min	2–3 min per muscle group (about 8–20 repetitions), in total ≥30 min
Type	Running, jumping, cycling, swimming, football	Running, jumping, cycling, swimming	Push-ups, sit-ups/crunches, pull-ups, handgrips, squats, climbing, martial arts, rowing

MVC, maximal voluntary contraction; $\dot{V}O_2$, oxygen uptake or oxygen consumption; Interval training can be used alternatively with aerobic training in healthy children.⁹

Reduced fitness of children with CHD

Many children with CHD have reduced exercise capacity and a reduced level of physical activity. A significantly reduced aerobic capacity in patients with CHD is worrisome, as health outcome and survival may be directly related to exercise capacity.¹⁰ Exercise capacity is an important predictor of health outcome and survival in adult patients with cardiovascular disease¹⁰ and children with pulmonary disease,¹¹ as well as in healthy children.^{10,12} This relation of exercise capacity and survival as well as morbidity has also been shown in patients with congenital heart disease,¹³ especially in those with repaired tetralogy of Fallot,¹⁴ transposition of the great arteries after atrial redirection,¹⁵ and with Fontan circulation.¹⁶

Other potential benefits of regular physical activity and exercise in patients with CHD include the adoption of a healthy lifestyle. In the long term, physical activity and exercise may be beneficial to prevent atherosclerotic cardiovascular disease, dyslipidaemia, obesity, hypertension, osteoporosis, and type 2 diabetes, which are frequently observed in sedentary individuals.¹⁷ Furthermore, increased physical activity in childhood

is associated with improved gross motor performance and positive emotional, social, and intellectual development.¹⁸

The reduced physical activity levels that have been noted among children with CHD are probably not caused by physiological factors (such as reduced exercise capacity), since several studies have shown that the relationship between exercise capacity and physical activity is low. Rather, it has been suggested that over-restriction of children with CHD by parents, caregivers, healthcare providers, educators, and sports trainers might be important^{19,20} and related to misperceptions regarding the relative risks versus benefits of participation.

In adults with heart disease, exercise rehabilitation programmes are mostly targeted towards patients with coronary artery disease²¹ and more recently also towards patients with chronic heart failure.²² In the sections on specific congenital heart lesions, we outline whether and how the general physical activity and exercise recommendations for healthy children should be modified for children with a typical lesions and treatment history. The recommendations for the most common congenital heart defects are summarized in Table 5.

Table 5. Summary of physical activity recommendations for common congenital heart defects

	Cardiorespiratory	Musculoskeletal	Other
Healthy children	60 mins/day MVPA Participation in competitive sport, leisure sport and physical activity unrestricted	2–3 days/week Intensity unrestricted within safe limits for injury prevention	<2-h screen time
Septal defects	Like healthy	Like healthy	Light activity for 3–6 months after device placement or until sternotomy heals
Aortic stenosis			Exercise testing is recommended in order to objectively document normal heart function with exertion.
Mild	Like healthy	Like healthy	
Moderate	Limit to moderate intensity at competitive sport only	Limit to moderate intensity only	
Aortic regurgitation (mild to moderate)	Like healthy	Like healthy	Rule out left ventricular or aortic dilatation and arrhythmia.
Bicuspid aortic valve (isolated)	Like healthy	Avoid very high intensity	
Coarctation of aorta	Like healthy	Limit to low or moderate intensity only	Exercise test to rule out abnormal BP response to exertion
Aortic dilatation or aneurysm (stable)			
Mild	Like healthy	Avoid very high intensity	
Moderate	Limit to low and moderate intensity competitive sport only	Limit to low intensity only	

(continued)

Table 5. Continued

	Cardiorespiratory	Musculoskeletal	Other
Pulmonary stenosis			Exercise test may be useful
<30 mmHg gradient	Like healthy	Like healthy	
30–50 mmHg gradient	Limit to low or moderate intensity competitive sport only	Limit to low or moderate intensity only	
Tetralogy of Fallot (without significant regurgitation)	Like healthy	Like healthy	Exercise test and Holter monitor are recommended
Pulmonary or tricuspid valve regurgitation (significant)	Limit to low-to-moderate intensity competitive sport only if right ventricular dysfunction	Limit to low-to-moderate intensity competitive sport only if right ventricular dysfunction	
Ebstein anomaly (without significant regurgitation)	Like healthy	Like healthy	
Transposition of the great arteries (arterial switch, without significant residuals)	Limit to low-to-moderate intensity competitive sport only	Limit to low-to-moderate intensity competitive sport only	Exercise test and Holter monitor recommended
Transposition of the Great Arteries (congenitally corrected)	Limit to low or moderate intensity competitive sport only	Limit to low-to-moderate intensity only	Should be individualized depending on clinical status
Functional single ventricle (Fontan)	Limit to low-to-moderate intensity competitive sport only	Limit to low-intensity competitive sport only	Exercise test and Holter monitor are recommended Contact limitations if on anticoagulants
Implanted devices (pacemaker, defibrillator)	Like healthy	Like healthy	Avoid activities with risk of contact to device or leads
Eisenmenger or pulmonary hypertension	Individualize recommendations based on clinical status and exercise response	Individualize recommendations based on clinical status and exercise response	Exercise test assessment is recommended
Heart transplantation	Individualize recommendations for competitive sport	Individualize recommendations for competitive sport	Exercise test assessment is recommended

For all lesions, unless individual circumstances indicate precautions, participation in leisure sport and cardiorespiratory and musculoskeletal physical activity is unrestricted and should be promoted consistently. In addition, all patients should be recommended to limit sedentary pursuits (i.e. screen time) to no more than 2 h/day. MVPA: moderate or vigorous physical activity.

Physical activity in children with CHD

Sufficient levels of physical activity are just as important for children with CHD as they are important for healthy children. However, children with CHD might be less active than peers because of overrestriction by parents and educators.²³ The early work of Bergman and Stamm showed that many children with ‘innocent’ heart murmurs were restricted by parents and

educators in their participation in physical activities.²³ More than 40 years later, the issue of ‘overprotection’ remains. Historically, the cause of restrictive attitudes has been assumed to be related to misperceptions regarding the risks versus benefits of participation. However, recent research has suggested that parent uncertainty about appropriate choices of physical activity may play a role since there is typically no agreement between parents, medical records, and cardiologists

regarding the physical activity restrictions appropriate for a child with complex CHD.²⁴

Several recent studies have shown that many children with CHD participate in less physical activity than recommended by public health guidelines.^{25–27} When a direct comparison with healthy peers was made, boys with CHD in particular demonstrated lower levels of physical activity.^{26–28} In addition, children with CHD with an activity restriction are at increased relative risk of becoming overweight (2.5) or obese (6.1).²⁹ When using self-report questionnaires, many children with CHD overestimate their exercise ability and physical activity level.^{30,31} Furthermore, a significant proportion of children with CHD believe that they can participate in physical activities that are more intense than those actually recommended by the cardiologist.³²

In a study of young adults with CHD, several with severe lesions, it was observed that only 33% of patients requested information regarding safe/appropriate levels of exercise.³³

The most frequently mentioned reason for not inquiring about physical activity restrictions was the assumption that all types of exercise were safe to perform.³³ Of those patients who received advice, 30% were restricted from some types of exercise, while 19% received recommendations to perform more exercise.³³ Recently a brief and simple instrument has been developed to screen for knowledge about appropriate and safe levels of physical activity in children with CHD.³⁴ The self-completed questionnaire with five questions about knowledge of parents and patients concerning physical activity, lifestyle issues, and heart condition is easy to administer and can help to identify those patients who need more information regarding appropriate and safe levels of physical activity.³⁴

It would appear that in children with CHD, physical activity level is not related to exercise capacity (e.g. peak).¹⁹ McCrindle et al.¹⁹ found no association between physical activity levels, measured using an accelerometer, and $\dot{V}O_{2\text{peak}}$ in 147 7–18-year-old patients after Fontan repair.

However, Arvidsson et al.²⁵ found a significant correlation between physical activity level and $\dot{V}O_{2\text{peak}}$ in 9–11-year-old girls with CHD and 14–16-year-old boys and girls with CHD, the relationship did not occur in 9–11-year-old boys with CHD. However, similar to the results of McCrindle et al., total time spent being physically active was not associated with $\dot{V}O_{2\text{peak}}$ in all age groups (9–11 and 14–16-year-olds).²⁵

A large study in 194 children with CHD reported significantly lower values for motor development compared to healthy children.³⁵ Almost 60% of the children with CHD reported moderate or severe disturbances in motor development. Reduced motor development was observed even in children with mild uncorrected lesions, and in

children without residual lesions after previous surgery. The authors suggested that this reduction was probably caused by overrestriction of physical activity from parents, healthcare providers, educators, and others.

In summary, many children with CHD are not meeting current public health physical activity guidelines. In many instances, physical activity levels are not related to exercise capacity, suggesting a behavioural influence (e.g. overrestriction or uncertainty) on physical activity.

Physical activity promotion

To our knowledge there is only one published study³⁶ about the promotion of physical activity in patients with CHD, and it was conducted in adult subjects. In 61 patients with CHD (with various diagnoses), Dua et al. found that a 10-week physical activity promotion programme, consisting of walking at home 5 days/week, was safe and effective in improving exercise capacity (Bruce treadmill time), self-efficacy, and self-perception as well as daily activity in patients with NYHA class.^{1–4,36} However, no control group was used, nor was there a follow up of the patients after the programme had been discontinued. Therefore, the long-term effects of the programme as well as the 'placebo effect' of an exercise test and intervention (including twice weekly telephone calls) are unknown.

Recommendations for clinical practice

Clinicians should be aware of the detrimental effects of inactivity and sedentary behaviour and should promote physical activity in children with CHD. Advice regarding appropriate levels of physical activity should be implemented in clinical consultations.

Children with CHD should comply with public health recommendations¹⁷ for daily participation in ≥ 60 min of moderate-to-vigorous physical activity that is developmentally appropriate and enjoyable, and involves a variety of activities. Moreover, they should perform < 2 h/day of sedentary activities such as watching TV, using a computer, and playing video games. (Class I, level of evidence B)

Physical activity counselling should be a priority during every visit with a healthcare professional. The importance of daily activity should be emphasized and encouraged at all clinic visits. Healthcare professionals should encourage the patient (and their caregivers) to ask questions about their physical activity levels and should provide specific counselling to encourage physical activity, particularly during winter months or inclement weather. (Class I, level of evidence C)

Clinicians should provide written recommendations (including restrictions and permissions) to both primary care providers as well as parents and patients regarding appropriate levels of physical activity and exercise. This

has to be repeated at school entry to reassure teachers about the safety of common school activities and school sports. (Class I, level of evidence C)

Physical activity participation should be assessed regularly. For children, assessments of motor skills should be completed until the child has achieved all of the basic motor skills (typically about 10 years of age). The basic motor skills include not only walking and running, but more sophisticated movement patterns (e.g. hopping, jumping, leaping) as well as object manipulation skills (e.g. throwing, catching, kicking). Involvement of a physical activity expert may be indicated. Objective measurements of the patient's daily physical activity (versus self-report or proxy-report) should be considered and patients who do not achieve the recommended level of daily moderate-to-vigorous activity should be offered additional counselling or training interventions until an adequate level of physical activity is achieved and maintained. (Class I, level of evidence C).

Specific congenital cardiac lesions

Isolated shunt defects (atrial septal defect, ventricular septal defect, and patent arterial duct)

General considerations. When exercise capacity is measured formally by exercise testing with measurements of gas exchange, a suboptimal or reduced exercise capacity is found in a considerable number of patients.^{37,38} This might be caused in some of them by a low level pulmonary arterial hypertension during exercise.³⁹ In most of them a haemodynamic burden is absent and physical inactivity seems the main reason for reduced exercise capacity. Some degree of training may be beneficial for this patient group.

Atrial septal defect. In patients with atrial septal defect (ASD), blood is shunted from the left to the right atrium during diastole, when the better compliance of the right ventricle lowers right atrial pressure more effectively than it does on the left side. In small defects, the total amount of blood is negligible. In larger defects, this leads to a volume overload of the right ventricle and might cause pulmonary hypertension during exercise.³⁹ Many patients will have a mildly reduced exercise capacity. After closure, patients will improve their exercise capacity and many will normalize it.⁴⁰ Residual pulmonary hypertension or atrial arrhythmias are very rare during childhood. Therefore, no limitation exists for physical exercise or sports activities for this patient group. This holds true for leisure sports and other physical activities.

After interventional device ASD closure, the patient should resume light sport activities as soon as the

puncture site at the groin healed completely (10–14 days after intervention). Contact sports should be avoided for 6 months till the device is completely covered by the endocardium, and the risk of dislodgement is minimal.

After surgical patch closure, there are no restrictions beyond those after the healing of the sternotomy wound.

Ventricular septal defect. In a patient with a small VSD, only a small amount of blood will be shunted from the left to the right ventricle. During exercise, the shunt will remain relatively small. Haemodynamic studies performed during graded exercise on this patient group showed a higher pulmonary than systemic blood flow. However, the relative shunt fraction (shunt flow/systemic blood flow) decreased with increasing exercise intensity.⁴¹ Values of systemic cardiac output during exercise were lower than in healthy subjects. Exercise performance was assessed by measurements of maximal endurance time and the maximal work rate on a bicycle ergometer. Exercise studies during submaximal testing with measurements of gas exchange showed reduced values for ventilatory anaerobic threshold in about half the patients in a consecutive series of 43 patients with native VSD (mean 86 ± 12 % of normal).⁴² The same was found in patients with a surgically closed VSD (mean 86 ± 12 % of normal).³⁸ In this patient group, the only variable that correlated with a decreased level of exercise capacity was a lower-than-normal level of physical activity during daily life. Therefore, these patients need to be encouraged to perform the recommended level of daily physical activities.

Children with a ventricular septal defect (repaired or un-repaired) should be encouraged to be active and to adopt a healthy lifestyle. If exercise capacity is abnormal, children with a VSD should be advised to engage in a physical training programme. After interventional or surgical VSD closure, the same recommendations apply as after ASD closure.

Previous postoperative exercise studies undertaken in small groups of children (including those with an ASD or VSD) have shown an improvement in maximal work rate on a bicycle ergometer, without any change in $\dot{V}O_{2peak}$.⁴³ This shows that mechanical efficiency for exercise is improved, which means that more work is performed at a same percentage of the $\dot{V}O_{2peak}$.

Patent arterial duct. For patients with a patent arterial duct (PDA), there is a similar haemodynamic situation as in patients with VSD. Nowadays, PDA closure is performed percutaneously. The same recommendation applies as after percutaneous ASD closure.

Recommendations for clinical practice

Leisure sport and daily physical activities (recreational activities). As long as there are no exercise-induced

arrhythmias, resting myocardial dysfunction, or pulmonary arterial hypertension, there is no reason for these children not to participate normally in physical exercise without restrictions. (Class I, level of evidence C)

Competitive sport. No limitation exists for competitive sports activities. Limitations for competitive sports are only in patients with pulmonary arterial hypertension. Information on exercise recommendations for these patients can be found in the section 'Cyanotic patients/Eisenmenger'. (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. Children with ASD/VSD and a reduced exercise capacity would benefit from the same exercise training programme recommended in healthy children. (Class I, level of evidence C)

Aortic stenosis (AS)

Congenital aortic stenosis can be classified in subvalvular, valvular, and supra-valvular stenosis. Subvalvular aortic stenosis is caused by a septal muscular ridge (hypertrophic obstructive cardiomyopathy), a tunnel-like fibromuscular stenosis, a fibromuscular ring, or a discrete membranous subaortic stenosis. Aortic stenosis can be part of a more complex heart defect and be associated with a ventricular septal defect with posterior and left malalignment of the outlet septum, with multilevel left heart obstruction (Shone complex) or small left heart structures (hypoplastic left heart syndrome).

The group of valvular aortic stenoses consists of both mucoid-degenerated thickened aortic valve cusps and thin cusps laterally fused at the commissures level. A common form is the total fusion of two cusps (bicuspid aortic valve).

Supra-valvular aortic stenosis usually appears as an hourglass-like stenosis at the sinotubular junction which extends to the proximal ascending aorta and is frequently seen in Williams Beuren syndrome or in familial supra-aortic stenosis.⁴⁴

Recommendations for clinical practice

Preparticipation screening. Medical history is essential to differentiate asymptomatic from symptomatic patients. Patients reporting symptoms like syncope, dizziness, angina, or disproportionate dyspnoea at exercise are at special risk for sudden cardiac death, and surgical or interventional treatment should be considered. With echocardiography, the exact anatomical location of the stenosis can be defined and the degree quantified as mild (mean instantaneous gradient <25 mmHg), moderate (25–40 mmHg), or severe (>40 mmHg).⁴⁵ An exercise test in asymptomatic patients with

moderate or severe stenosis might be necessary to unveil exercise-inducible symptoms, especially when patients describe themselves as asymptomatic.

Leisure sport and daily physical activities, competitive sport. For patients with hypertrophic obstructive cardiomyopathy, the reader is referred to the recommendation on congenital arrhythmogenic diseases.⁴⁶

For valvular aortic stenosis, there are epidemiological data on the incidence of sudden death during sport activity. About 2–4% of all young athletes with sudden cardiac deaths are found to have valvular aortic stenosis.^{47,48} Syncope, dizziness, angina pectoris, or disproportionate dyspnoea, as well as a pathological exercise test results, are precursors of sudden cardiac deaths in the elderly.^{49,50} Prospective data in children, adolescents, and young adults are missing, as balloon valvuloplasty is performed very liberally with good results and little adverse effects.^{51,52} One has to refer to the recommendations for adults with aortic stenosis,^{3,53} despite the fact that these recommendations are mainly based on studies in elderly with calcified aortic stenosis.

For the other forms of congenital aortic stenosis, specific studies on sport and exercise are rare. Therefore, one also has to extrapolate from the recommendations for valvular aortic stenosis,^{3,53} despite the pathophysiology of these lesions during exercise being very different (dynamic increase of subvalvular stenosis during exercise with reduced coronary perfusion, versus altered coronary blood supply and possible coronary artery involvement in supra-valvular aortic stenosis).

Children with mild AS can participate in all types of recreational/competitive sports, but should undergo serial evaluation of AS severity on at least an annual basis.⁵⁴ (Class I, level of evidence C)

Children with moderate AS can engage in low-to-moderate intensity dynamic and static competitive sports if exercise testing to at least the level of the sports activity demonstrates satisfactory exercise capacity without symptoms, ST-segment depression, or ventricular tachyarrhythmias, and with normal blood pressure response.⁵⁴ (Class I, level of evidence C)

Children with severe AS or symptomatic patients with moderate AS should only engage in low-intensity leisure sport and physical activity depending on symptoms and the findings at the exercise test. They should not engage in competitive sports.⁵⁴ (Class I, level of evidence C)

Children with mild or moderate aortic regurgitation should not be withheld from any kind of physical activity as long as no left ventricular (LV) dilatation, aortic dilatation/aneurysm, or arrhythmia can be detected. (Class I, level of evidence C)

Exercise training as a preventive/therapeutic intervention. All training concepts in patients with

aortic stenosis were developed with the aim of preventing calcified aortic valve stenosis. They cannot be directly transposed to the different types of congenital aortic stenoses. Physiologically, it is unknown whether exercise training is able to improve or at least is able to stop the progression of the stenosis or restenosis. So at the moment, no recommendation can be made regarding therapeutic exercise training. However, there are some data from individual patients enrolled in training studies after surgical correction of CHD that show that physical training after surgical resection of the stenosis is helpful to reach full exercise capacity more quickly.^{55–57}

Bicuspid aortic valve syndrome

A bicuspid aortic valve (BAV) is an aortic valve that opens only in two commissures. Most commonly, the commissure between right and left coronary cusps (right–left fusion) and, less often, that between right and noncoronary cusps (right–nonfusion) are fused by a raphe resulting in two asymmetrical cusps. Fusion of the left coronary and the noncoronary cusp and ‘true’ symmetrically bicuspid valve without recognizable raphe are rare. Especially the right–left fusion BAV is associated with other congenital heart defects like coarctation or malalignment ventricular septal defect, whereas on long-term follow up, right–nonfusion BAV is prone to become regurgitant or stenotic.⁵⁸

Bicuspid aortic valves are accompanied with intrinsic histological aortic wall anomalies including cystic media necrosis and elastic fibre degradation which are similar, but less extensive, than those seen in Marfan syndrome.^{59–61} Patients are prone to develop dilatation of the aortic root (right–left fusion) or of the ascending aorta/aortic arch (right–noncoronary cusp fusion).^{62,63} Because of the complexity of the findings, the name ‘bicuspid aortic valve syndrome’ was coined.⁶⁴

However, several congenital defects of the aorta (BAV, coarctation, enhanced aortic stiffness, idiopathic aortic aneurysm) seem to be linked to a common autosomal dominant genetic anomaly with incomplete penetrance and variable expression.^{61,65}

Recommendations for clinical practice

Preparticipation screening. Echocardiography will define the diagnosis and confirm/exclude concomitant findings, including valve function. Medical history, physical examination with blood pressure measurement at all four limbs, and electrocardiogram (ECG) will also give clues to these accompanying defects. In case of aortic valve dysfunction and coarctation, an exercise test, or in case of moderate or severe aortic dilatation

magnetic resonance imaging (MRI) or helical computed tomography (CT), will complete the assessment.

Leisure sport and daily physical activities, competitive sport. If aortic stenosis, aortic regurgitation, aortic aneurysm, and coarctation are ruled out, patients with BAV can participate in all sport activities. However, a recent study showed, that patients with BAV who undergo regular training showed a progression in LV diameters as well as aortic diameters.⁶⁶ Additionally, the risk of events rises with the progression of valvular dysfunction.⁶⁷ Therefore, close surveillance with yearly echocardiography is warranted. Furthermore, strength training with a very high static component (i.e. weight lifting) enhances aortic stiffness⁶⁸ and dilatation⁶⁹ and should be avoided. (Class IIa, level of evidence C)

If aortic stenosis or regurgitation is present, physical activities must be curtailed as suggested by the guidelines for the management of valvular heart disease.⁴⁶

Patients with mild aortic dilatation must stay at close surveillance with at least yearly echocardiography as dilatation may become progressive with any kind of training.⁶⁶ Patients with stable moderate aortic aneurysm (40–45 mm in adults and their equivalents in children) may participate in low static and low or moderate dynamic competitive sports, additionally excluding sports with body collision and trauma.^{45,70} (Class IIa, level of evidence C)

Patients with progressive or large aortic aneurysm (>45 mm in adults or equivalent in children) must be consulted individually. They must limit their sports activities to low static/low dynamic activities or in more severe cases referred to surgery. (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. Longitudinal studies in subjects with BAV are limited to competitive athletes and they show slow progression in LV and ascending aortic diameters with regular training compared to those athletes with tricuspid aortic valve.⁶⁶ However, no data is available on strict endurance training programmes. Several studies carried out in patients without aortic valve malfunction have shown that various kinds of endurance training are able to improve the elastic properties of the aorta.^{71–75} Even though there is no definitive proof in patients with BAV, it could be argued that moderate endurance training should be promoted in patients with BAV after aortic stenosis, aortic regurgitation, aortic aneurysm, and coarctation are ruled out. Therefore, endurance training recommendations should follow those proposed for healthy children. Interval training should be omitted and resistance training should be limited to low or moderate intensities involving small muscle groups separately. (Class IIa, level of evidence C)

Aortic coarctation syndrome (COA)

Coarctation of the aorta is defined as a significant narrowing at the aortic isthmus, which is most commonly located after the origin of the left subclavian artery at the insertion of the arterial ligament. It is caused by ductal tissue extending into the aortic wall and restricting the lumen of the aorta after birth. Another hypothesis of coarctation origin is that diminished blood flow through the aortic arch in fetal life might cause underdevelopment of the aortic arch. This might explain why coarctation is often seen in patients with a BAV, a subvalvular, valvular, or supra-valvular aortic stenosis, hypoplastic left heart syndrome, mitral valve stenosis, or complex congenital heart defects with one of those entities.

Aortic coarctation is nowadays mainly divided into isolated and complex coarctation. However, even those classified as isolated coarctation often show minor additional anatomical abnormalities like BAV, parachute mitral valve, or elongation of the distal transverse aortic arch with displacement of the left subclavian artery more distally in the course of the aortic arch. In all patients, histological abnormalities with disarray and depletion of elastic fibres are seen in the media of the elastic arteries distal as well as proximal to the coarctation site. Changes are not confined to the aorta, since intracranial arterial aneurysms can be found in 10% of patients.⁷⁶ 'Coarctation syndrome' was established to describe a systemic arterial vessel disease with the stenosis at the aortic isthmus as the most prominent feature.^{77,78} However, even that name misses the large overlap with BAV syndrome,^{61,65} as well as the overlap with several defects with hypoplastic left heart structures.^{79–82}

In the long term, cardiovascular morbidity and mortality is increased in untreated subjects⁸³ and it is raised even in surgically well 'repaired' patients. Coronary artery disease is the leading cause of death after coarctation repair.⁸⁴ Stroke, heart failure, ruptured aortic and cerebral aneurysms, and sudden cardiac death occur even after early repair.⁸⁵ An elevated blood pressure seems to play a central role in these long-term complications.⁸⁶

Exercise capacity is reduced^{13,37} despite a good morphological result without residual stenosis.⁸⁷ Some authors advocate that this is the result of a lack of exercise in childhood due to overprotective parents or physicians.³⁵ On the other hand, there is a more impaired exercise capacity after coarctation repair than after surgical repair of other minor congenital heart defects.⁸⁸ Possible mechanisms include the presence of vascular dysfunction related to reduced aortic elasticity, decreased peripheral endothelium-dependent and -independent arteriolar function,⁸⁹ and enhanced

baroreceptor reflexes.^{90,91} The most striking finding at exercise in many coarctation patients is a disproportionate rise in systolic blood pressure,⁸⁷ caused both by minor restenosis and by aortic wall stiffening. This is particularly important because exercise-induced hypertension is usually associated with persistent hypertension at follow up in patients with repaired coarctation.

Recommendations for clinical practice

Preparticipation screening. Medical history should focus on previous surgical or interventional procedures as well as on blood pressure history and medication. Physical examination with blood pressure measurement at all four limbs gives first clues to the presence or extent of restenosis. ECG might show signs of left hypertrophy. Echocardiography can demonstrate left heart hypertrophy and could reveal intracardiac associated defects. Echocardiographic depiction of the aortic arch is rarely possible. CW-Doppler of the aortic isthmus should only be analysed concerning diastolic forward flow, which is a sign of restenosis. Gradient calculation from peak velocity at the end of the aortic arch is not predictive of restenosis.⁹² With the suspicion of restenosis and to exclude aneurysm at the coarctation site or in the ascending thoracic aorta, MRI or helical CT should be performed. However, it is of doubt whether an incidence of mostly small cranial aneurysm warrants a cranial MRI or cranial CT in every patient.

Leisure sport and daily physical activities, competitive sport. With the lack of any studies on sports and physical training in coarctation patients both before and after repair, it is only possible to extrapolate from the recommendations for arterial hypertension and from those for BAV. Isolated coarctation patients without significant residual gradient at the coarctation site (<20 mmHg), without excessive rise in systolic blood pressure at exercise (>3 standard deviations above reference)⁸⁷ and without aortic or cerebral aneurysm can participate in all kind of sport activities except those with a very high static component. (Class IIa, level of evidence C)

Patients with aortic restenosis should be referred to catheter intervention or surgery, as well as patients with large aneurysm. (Class I, level of evidence C)

For patients with small aneurysm, see the section on BAV syndrome.

Exercise training as a preventive/therapeutic intervention. With the lack of studies on the safety and efficacy of exercise training in coarctation patients, one has to go back to the exercise

physiological principles. A low-to-moderate endurance training has an favourable impact on arterial hypertension, arterial stiffness,^{71,72} and endothelial function,^{93,94} as well as on coronary artery disease^{95,96} and stroke⁹⁷ prevention. This implicates that an active life style and even aerobic endurance training should be promoted in coarctation patient as primary prevention against typical long-term sequels like in healthy children. Interval training should be omitted and resistance training limited to low or moderate intensities in small muscle groups separately. (Class IIa, level of evidence C)

Regular medical surveillance is necessary to rule out complications.

Pulmonary stenosis

General considerations. Pulmonary valve stenosis (PS) is the most common form of isolated right ventricular (RV) outflow tract obstruction. It is usually caused by fused or dysplastic cusps and is mainly congenital. The degree of RV outflow obstruction is highly variable, with most patients having mild or modest degrees of obstruction and presenting only with a cardiac murmur and normal ECG and exercise tolerance. In a minority of patients, the degree of obstruction might be sufficient to promote RV hypertrophy and to produce symptoms, usually in the form of early fatigue during exercise⁹⁸ or atrial arrhythmias related to right atrial dilation. Although three decades ago surgical valvotomy was the treatment of choice, nowadays most patients are amenable of percutaneous balloon relief of the obstruction with excellent results in terms of early significant residual obstruction.⁹⁹ However, a small minority of patients might present with small pulmonary valve annulus and might therefore still require a full surgical valvotomy or transvalvular patch repair.

Medium- and long-term follow-up studies have shown favourable results with either surgical or balloon valvuloplasty in terms of freedom from reintervention, cardiac function, and exercise capacity.^{100,101} However, a minority of patients can develop significant pulmonary regurgitation after neonatal or paediatric intervention, and might be candidates for pulmonary valve replacement.¹⁰² This patient subgroup can present with mildly reduced exercise capacity.

In PS, RV systolic pressure and oxygen demand are increased at rest and more so with exercise. Exercise tolerance in children and adults with mild valvular PS is nearly normal, but is diminished in those with moderate and severe stenosis, indicating impaired ability to sustain adequate cardiac output. Following relief of stenosis, cardiac performance readily improves in children, but it can remain abnormal in adults. This appears to be related to postoperative resolution of RV hypertrophy in children, whereas myocardial

fibrosis may explain the slow and reduced improvement in adults.¹⁰³

PS, whether native or residual, is classified based on the peak transvalvular gradient as mild (gradient <30 mmHg), moderate (gradient between 30 and 50 mmHg), and severe (gradient >50 mmHg).

Recommendations for clinical practice

Leisure sport and daily physical activities (recreational activities). Patients with native mild PS usually have no RV hypertrophy and a normal exercise tolerance. These patients should have no restriction in the type and level of physical activities. The same applies to patients with treated PS who have mild residual PS and only mild pulmonary regurgitation. (Class I, level of evidence C)

Patients with treated PS who have moderate residual PS and moderate or higher pulmonary regurgitation and a normal ECG should have no restriction in the type and intensity of physical activity. Annual reassessment of the underlying condition by ECG should be performed to confirm the stability of the condition. (Class I, level of evidence C)

Patients with isolated residual significant pulmonary valve regurgitation, especially if associated with RV dilation, should follow the recommendations given for healthy children (Table 2) if RV systolic function assessed by two-dimensional ECG or MRI is normal. If RV function is abnormal, they should have their activity levels restricted to those activities with mild cardiovascular involvement. (Class I, level of evidence C)

Patients with significant pulmonary regurgitation who have received a transvalvular patch (with or without extension to RV infundibulum) or a valvotomy should have exercise restrictions similar to that of patients with tetralogy of Fallot. (Class I, level of evidence C)

Patients with significant PS (pressure gradient >50 mmHg), either native or post-treatment, usually have various degrees of RV hypertrophy and might present RV systolic dysfunction. Usually such patients will present symptoms in the form of right-sided heart failure and exercise intolerance. These patients should be referred for treatment at any age they present and mild physical activity can be recommended only in asymptomatic patients on an individual basis. (Class I, level of evidence C)

Competitive sport. Patients with treated or untreated PS and with a peak gradient <30 mmHg, normal RV function and size, no more than mild regurgitation, normal ECG, and only mild RV hypertrophy can participate in all competitive sports if symptoms are

not present. Re-evaluation is necessary every 12 months.³ (Class I, level of evidence C)

Patients with treated or untreated PS and a peak gradient between 30 and 50 mmHg and no significant pulmonary regurgitation, normal RV size and function, normal ECG, and only mild RV hypertrophy can participate in moderate intensity aerobic and mild anaerobic sports. Re-evaluation is necessary every 6 months.³ (Class I, level of evidence C)

Patients with low residual peak systolic gradient and associated significant pulmonary regurgitation and abnormal RV size can participate in low-intensity aerobic competitive sports if they are asymptomatic and have normal RV systolic function. (Class I, level of evidence C)

MRI might be useful for the further stratification of such patients.¹

Patients with a peak systolic gradient >50 mmHg can participate in low-intensity competitive sports after 3–6 months of treatment of the PS.^{1,3} (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. There are no reported data on the safety and efficacy of exercise training in patients with native and treated PS.

Asymptomatic patients with isolated mild and moderate PS, with or without associated pulmonary regurgitation and normal RV size and function, should follow the same recommendations as for healthy subjects. (Class I, level of evidence C)

Patients with less than moderate PS who have moderate or severe pulmonary regurgitation and RV dilation but who are asymptomatic and have normal RV systolic function should be involved in an exercise training programme limited to moderate endurance or resistive intensity. (Class IIa, level of evidence C) Patients with pulmonary regurgitation and abnormal RV size and function might benefit from surgical or percutaneous intervention. It is important to assess this group of patients with MRI and repeated 24-h ECG monitoring to exclude the presence of arrhythmias. If treatment is not warranted, the exercise training programme should be limited to low endurance and low resistive intensities. Exercise training programmes in this group should be continued lifelong and should be performed in hospital environments. Frequent re-evaluation of such patients every 6–12 months is of paramount importance. (Class I, level of evidence C)

In patients with sustained exercise-induced atrial or ventricular arrhythmias, exercise training might be associated with a risk of sudden death during exercise. Patients with sustained exercise-induced arrhythmias should be refrained from participating in exercise training programmes. Patients with exercise-induced

nonsustained but repetitive arrhythmias (frequent ectopic beats, isolated, or as bigeminy, monomorphic, or polymorphic) can participate in exercise training programmes with low endurance and low resistive intensities if 24-h ECG monitoring does not show any sustained arrhythmia. (Class I, level of evidence C)

Patients with significant PV stenosis should be referred for treatment before participating in an exercise training programmes. (Class I, level of evidence C)

Tetralogy of Fallot

General considerations. Tetralogy of Fallot (ToF) is the most common form of cyanotic congenital defect. Surgical repair is achieved by patch closure of the ventricular septal defect and enlarging the RV outflow tract through an infundibular patch, which sometimes need to be extended beyond the pulmonary annulus in patients with significant valvular and supra-valvular PS. This leads to three important long-term consequences of repaired ToF, which are the presence of significant RV outflow tract restenosis and/or significant pulmonary regurgitation, as well as the occurrence of cardiac arrhythmias, predominantly of ventricular origin related to re-entry in the area of the infundibular patch.

Several reports have assessed the exercise capacity in ToF patients. These reports emphasize that several residual lesions can cause a reduced exercise tolerance in patients with repaired ToF.

Residual PS after direct or conduit repair of ToF has been associated with reduced exercise capacity and increased ventilatory response to exercise, as a consequence of increased afterload to the RV with abnormal cardiac output response to exercise.¹⁰⁴ Presence of residual pulmonary artery branch stenosis and presence of aortopulmonary collateral vessels have also been associated with abnormal ventilatory response to exercise, as a consequence of ventilation/perfusion mismatch.^{105,106}

Presence of residual pulmonary valve regurgitation is possibly the most common residual defect after complete repair of ToF. There is evidence that long-standing significant pulmonary regurgitation causes, as a consequence of increased RV volume overload, progressive RV dilation. Its effect on RV function is however less clear. Indeed, even though long-standing significant pulmonary regurgitation has clearly been associated with reduced RV systolic function, it is common experience that some patients with significant pulmonary regurgitation maintain a similar RV ejection fraction at serial testing. There is evidence that, in presence of normal RV systolic function, the effect of isolated significant pulmonary regurgitation on exercise capacity is limited.¹⁰⁷ A possible explanation for this finding is that pulmonary regurgitation seems to

be of reduced importance during exercise, as a consequence of shortening of the diastole and decrease in the pulmonary vascular resistance.¹⁰⁸ However, patients with significant pulmonary regurgitation who also have a reduced RV systolic function have the lowest exercise capacity, particularly if they also have a reduced LV systolic function. Recent observations also confirm the predominant impact of reduced RV function of exercise capacity in repaired ToF.¹⁰⁹

Even though in recent years most children received complete surgical repair in the first year of life, there is a large number of adults who required palliative procedures and subsequently underwent delayed complete repair. Patients with late repair frequently developed more important degrees of RV hypertrophy with reduced compliance of the right ventricle, leading to diastolic RV dysfunction after complete repair. Even though such condition is associated with an higher early postoperative morbidity related to prolonged intensive care stay and low cardiac output syndrome,¹¹⁰ it has been associated with improved exercise capacity at long term, possibly from its influence on pulmonary regurgitation.^{111,112}

Finally, a subgroup of patients with repaired ToF will present, or will be at risk of, sustained potentially life-threatening ventricular arrhythmias. Factors which are known to be associated with an increased risk of ventricular arrhythmias are an older age at repair, increased RV systolic pressure, the presence of RV dilation and dysfunction (usually as a consequence of significant pulmonary regurgitation), the presence of LV systolic dysfunction, and the presence of a broad QRS (duration >180 msec).^{112–115}

Recommendations for clinical practice

Leisure sport and daily physical activities (recreational activities). Asymptomatic patients with nonsignificant pulmonary regurgitation and normal/mild RV dilation, with normal RV pressure and systolic function and who do not have atrial or ventricular arrhythmias at ECG monitoring or exercise testing should be encouraged to engage in physical activities to the same extent and intensity of the general population (Table 2). (Class I, level of evidence C)

Asymptomatic patients with nonsignificant pulmonary regurgitation and only moderate RV dilation but normal RV function and those with <mild tricuspid valve regurgitation and no arrhythmia documented at ECG monitoring or exercise test should follow the recommendations of healthy children. (Class I, level of evidence C)

Patients with atrial or ventricular arrhythmias, which are successfully controlled by medical treatment or radiofrequency ablation, can follow the

recommendations of healthy children as well. (Class I, level of evidence C)

Asymptomatic patients with significant pulmonary regurgitation who have moderate or significant RV dilation but normal RV systolic function and no arrhythmias documented at 24-h ECG monitoring or exercise test should also follow the recommendations of healthy children. The condition should be reviewed every 6 months. (Class I, level of evidence C)

Asymptomatic subjects with significant pulmonary regurgitation and significant RV dilation and abnormal RV function can be allowed to participate in mild-intensity aerobic activities after serial testing has shown a stable condition and no arrhythmias is documented at 24-h ECG monitoring or exercise test. (Class I, level of evidence C)

Patients who are symptomatic from mild or moderate efforts, those with more than moderate residual RV outflow obstruction, those with abnormal LV function, those with signs of increased RV pressures (RV systolic pressure >2/3 of systolic blood pressure) or increased transpulmonary gradient, those with a significant residual intracardiac shunt (pulmonary/systemic output $\geq 1.5:1$) not amenable of correction, and those with documented sustained atrial or ventricular arrhythmias which are not controlled by medical therapy or radiofrequency ablation should have their physical exercise restricted to low-intensity activities that are preferably aerobic. (Class I, level of evidence C)

Competitive sports. Patients with an excellent repair who have a normal or near-normal RV pressure, no or only mild RV volume overload, no significant residual shunt, and no atrial or ventricular arrhythmias on ambulatory ECG monitoring or exercise testing can participate in all sports of any intensity.¹ ESC guidelines limit intensity to moderate aerobic and anaerobic activities.³ (Class I, level of evidence C)

Patients with residual pulmonary regurgitation and increased RV volumes, increased RV systolic pressure (RV systolic pressure $\geq 50\%$ of LV systolic pressure), or atrial or ventricular arrhythmias should participate only in low-intensity competitive sports.¹ (Class I, level of evidence C)

Exercise training. Several cohort studies and a small, randomized study have assessed the safety and efficacy of endurance exercise training in patients with repaired ToF.

In a small, randomized study Therrien et al.¹¹⁶ analysed the effect of an individualized aerobic exercise training programme in a group of 18 stable adult patients with repaired ToF. The training protocol consisted of hospital-based sessions (40 min of duration once a week with an intensity of 60–85% of $\dot{V}O_{2peak}$)

and home-based sessions (at least 30 min of brisk walk twice a week with an intensity of 60–85% of peak heart rate) for a period of 12 weeks. Analysis of each patient's home exercise diary showed that seven of the nine patients enrolled in the training group completed the minimum required number of training sessions. The study, which included eight patients with significant pulmonary regurgitation and various degrees of RV systolic dysfunction (but excluded patients with documented ventricular arrhythmias), showed that, compared to the control group, patients enrolled in this moderate workload training programme had significantly improved their exercise capacity at the end of the study.

The available evidence suggests that, in order to achieve a meaningful improvement of $\dot{V}O_{2\text{peak}}$, a heavy intensity exercise training programme is required (70–85% of $\dot{V}O_{2\text{peak}}$). Regarding the duration of the exercise training programme, the available data suggest that at least three training sessions/week with duration between 30 and 40 min are necessary to obtain an improvement of $\dot{V}O_{2\text{peak}}$ after 3 months of training. Of note, the data available suggest the possibility that compliance with the exercise training protocol might be higher in the pediatric age group, and possibly higher physical activity levels are maintained in pediatric patients outside of the training protocol.⁵⁶ The optimal arrangement, especially in those patients with residual hemodynamic lesions, consists in the combination of hospital- and home-based training sessions.

In conclusion, all children and young adults with repaired ToF who are asymptomatic or mildly symptomatic and who do not have sustained atrial or ventricular arrhythmias during routine ambulatory ECG monitoring or exercise testing (either spontaneously or after medical/electrophysiological intervention) should be involved in an exercise training programme. (Class I, level of evidence B)

Patients without RV outflow obstruction and those with nonsignificant pulmonary regurgitation and normal RV size and function should follow the recommendations for the general population (Table 2) if they are asymptomatic and have no documented arrhythmias at ECG monitoring and exercise testing. (Class I, level of evidence B)

Asymptomatic patients with significant pulmonary regurgitation and RV dilation, those with mild/moderate RV dysfunction, and those with atrial or ventricular arrhythmias controlled by medical therapy or radiofrequency ablation should follow the recommendations for the general population (Table 2) but limited to aerobic exercise training and only if LV systolic function is normal. (Class I, level of evidence B)

Patients with mild LV dysfunction should follow the recommendations for the general population (Table 2). (Class I, level of evidence C)

Patients with documented sustained atrial or ventricular arrhythmias (either spontaneous or exercise induced) that are not controlled by medical therapy or radiofrequency ablation, those with RV hypertension (RV pressure >2/3 of systolic arterial blood pressure), and those with symptoms for mild-to-moderate levels of exercise should be excluded from exercise training programmes at present since the safety profile is unknown. (Class I, level of evidence C)

Ebstein anomaly

General considerations. Ebstein anomaly is characterized by an abnormality of the tricuspid valve in which the septal leaflet and often the mural leaflet attachments are atypically displaced from the atrioventricular ring. The commissure between the two leaflets is the point of maximal displacement and is at the posterior border of the ventricular septum. The anterosuperior leaflet is usually malformed, excessively large, and abnormally attached or adherent to the endocardium of the RV-free wall. Thus the inlet portion of the RV is usually integrated functionally with the right atrium (atrialized portion), while the other, including the trabecular and outlet portions of the RV, constitute the functional, and often small, RV. Varying degrees of tricuspid regurgitation result from the abnormal tricuspid morphology, with consequent further atrial enlargement. Either an atrial septal defect or a patent foramen ovale is present in over one-third of hearts, with subsequent potential right–left shunting. Associated cardiac defects are commonly observed in the form of ventricular septal defects, abnormalities of the RV outflow tract, or abnormal cardiac loop. In addition, the apical displacement of the septal tricuspid valve leaflet is associated with discontinuity of the central fibrous body, creating a potential substrate for multiple accessory atrioventricular pathways.

The natural history and the functional status of patients with Ebstein anomaly is quite variable, as a consequence of the severity of underlying tricuspid valve abnormality and regurgitation and presence of associated defects, ranging from patients with severe neonatal heart failure and cyanosis to asymptomatic adult patients.

Few studies have assessed the exercise capacity and its determinants in children and adults with Ebstein anomaly. MacLellan-Tobert et al.¹¹⁷ studied exercise capacity in 117 patients with a similar representation of children and adults. They observed that, besides having a reduced $\dot{V}O_{2\text{peak}}$ compared to normal, patients who had received an operation (in the form of tricuspid

valve plasty or replacement, and atrial septal defect closure if present preoperatively) had a higher exercise capacity. They observed that there was a correlation between $\dot{V}O_{2peak}$ and both resting oxygen saturation and younger age at test in the preoperative group, whereas peak was related to younger age and smaller cardiothoracic ratio in patients who had received an intervention. Cyanosis at rest was frequent in their preoperative cohort at rest (average 82%), and was particularly evident at peak exercise (average 75%). Of note, only a very small minority of postoperative patients had a normal exercise capacity in this study (average $\dot{V}O_{2peak}$ 64% of predicted), with no patient in the preoperative group (natural history) having normal exercise capacity (average $\dot{V}O_{2peak}$ 50% of predicted). Another important finding is that patients with Ebstein anomaly have an abnormal heart rate response to exercise, likely to be the expression of sinus node dysfunction, both pre- and postoperatively.

A subsequent smaller study by Trojarska et al.¹¹⁸ has shown that there is an inverse relationship between exercise capacity and the anatomical and functional severity of Ebstein anomaly, suggesting that reduced RV stroke volume and possibly abnormal ventricular–ventricular interaction can play a role in the reported reduced exercise capacity. Of note, the patients studied by Trojarska et al. represented adult survivors in natural history with normal arterial saturation ($SpO_2\%$) values at rest (average $SpO_2\%$ 97%). The same study showed that patients with Ebstein anomaly have an excessive ventilatory response to exercise, likely related to a ventilation–perfusion mismatch consequent to pulmonary hypoperfusion and right–left shunting at the atrial level.

Recommendations for clinical practice

Leisure sport and daily physical activities (recreational activities). Non-cyanotic, asymptomatic patients who have a reasonably developed RV with no more than mild tricuspid regurgitation, normal LV systolic function, and no arrhythmias demonstrated at 24-h ECG monitoring, should have no restriction to physical activity. (Class I, level of evidence C)

Asymptomatic patients who have moderate tricuspid regurgitation, normal arterial saturation, and supraventricular arrhythmias (paroxysmal, associated or not to Wolff-Parkinson-White syndrome (WPW)) which are controlled by medical/interventional treatment can be allowed low-level dynamic, moderate-level static physical activities. (Class I, level of evidence C)

Patients with symptoms during exercise or at rest, those with significantly dilated right atrium and RV,

those with LV systolic dysfunction, those with severe tricuspid regurgitation, and those with chronic atrial arrhythmias (atrial fibrillation) or repetitive ventricular arrhythmias should be precluded from undertaking physical exercise of any level. (Class I, level of evidence C)

Competitive sport. Patients with mild forms of Ebstein anomaly without cyanosis, normal RV size, normal LV systolic function, and no evidence of atrial or ventricular arrhythmias can participate in all sports.¹ (Class I, level of evidence C)

Patients with moderate tricuspid valve regurgitation can participate in low-intensity competitive sports if they are asymptomatic and there is no evidence of arrhythmias on 24-h ECG monitoring.¹ (Class I, level of evidence C)

Patients with severe Ebstein anomaly are precluded from all competitive sport participation. However, 3 months after surgical repair, low-intensity competitive sports can be permitted if the patient is asymptomatic, tricuspid regurgitation is absent or mild, right atrial size is not severely increased, and no atrial or ventricular arrhythmias are documented on 24-h ECG monitoring or exercise test. Selected patients with an excellent haemodynamic result may be permitted additional participation on an individual basis after 3 months from repair.¹ (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. No data exists on the effect and safety of exercise training in patients with Ebstein anomaly.

Non-cyanotic, asymptomatic patients who have a reasonably developed RV with no more than mild tricuspid regurgitation and normal LV systolic function and no arrhythmias demonstrated at 24-h ECG monitoring should follow the recommendations for the general population (Table 2). In these patients, a fully outpatient setting is appropriate. (Class I, level of evidence C)

Those patients with symptoms for moderate levels of exercise or who have subcyanosis or cyanosis at rest, those with moderate tricuspid valve regurgitation, and those with supraventricular arrhythmias (paroxysmal, associated or not to WPW) that are controlled by medications would probably benefit from an exercise training programme of moderate endurance and moderate strength training. This exercise training programme should be first established in an ambulatory setting with additional sessions performed in the community/home setting. Frequent re-assessment of these patients involved in a training programme is mandatory to exclude the presence of physical deterioration related to exercise. (Class I, level of evidence C)

Transposition of the great arteries (TGA)

General considerations. More than two decades ago, intra-atrial 'physiological' repair for TGA according to Senning or Mustard was replaced by the neonatal arterial switch operation (asoTGA) as the surgical procedure of choice for anatomical correction of TGA with or without VSD. A follow-up period of 20–25 years is now available, demonstrating excellent intermediate and long-term general and cardiac health status in children, adolescents and young adults. Freedom from reintervention has been found in >90% of patients at mean age 17 years.¹¹⁹ However, in a group of young adults, 17% were recently detected with at least one clinically significant cardiac lesion such as ventricular or valvular dysfunction or arrhythmia.¹²⁰ Increasing reintervention rates with age for sub- and supravalvular pulmonary stenoses, possible development of significant neo-aortic valve insufficiency as well as morphology and function of the re-implanted coronary arteries remain a matter of caution and concern.

Exercise capacity after asoTGA. Mid- and long-term exercise performance in children and adolescents^{119,121–129} and in young adults,^{120,130} as assessed by cardiopulmonary exercise test on treadmill or cycle, has been studied in several recent studies^{119,121–126} with results in the lower normal or subnormal range for $\dot{V}O_{2peak}$ (average 87%, range 79–113% of predicted normal) and for peak heart rate (HR_{peak} ; average 95%, range 93–97% of predicted normal). A longitudinal cohort study over 20 years^{119,127–129} has shown continuously normal exercise capacity and normal exercise ECG as well as $\dot{V}O_{2peak}$ and HR_{peak} in most patients, but reduced physical activity patterns compared to healthy peers (averaged 39 min of moderate-to-vigorous activity on 2.6 days/week at mean age 17 years). Significant RV outflow tract obstruction¹²⁶ and abnormal pulmonary flow distribution due to pulmonary artery branch obstruction or hypoplasia as well as reduced LV stroke volume (data not shown) have been evaluated as independent risk factors for reduced $\dot{V}O_{2peak}$.^{120,130} Diminished coronary flow reserve during exercise is a matter of controversial discussion;^{131–133} negative effects of increased RV pressure and myocardial hypertrophy are assumed.

Recommendations for clinical practice

Preparticipation screening. Prior to admission to any sport programme, children after asoTGA should be evaluated by a paediatric cardiologist with respect to: the exact medical history, NYHA functional classification, physical examination, ECG, Holter ECG, and (tissue-Doppler) echocardiography, cardiopulmonary

exercise test, and cardiac MRI (routinely after pubertal growth to assess ventricular function, coronary artery morphology and delayed enhancement, aortic root size and function, RV outflow tract and pulmonary perfusion).

In absence of any of the above-mentioned complications, standard ECG and echocardiography should be repeated every 2 years, as also cardiopulmonary exercise test and Holter ECG, according to the intensity of exercise participation.

Leisure sport and daily physical activities (recreational activities). Children and adolescents without residual lesions (normal left and right ventricular function, no atrial or ventricular tachyarrhythmia) and a normal exercise test result including exercise ECG can participate in all kinds of leisure sports, recreational physical activities, and school sport classes without any restriction.^{2,134} (Class I, level of evidence C)

Children with minor residual lesions (small residual VSD, mild stenosis or insufficiency of the neo-aortic and neo-pulmonary valves, mild supravalvular PS, singular supra- or ventricular ectopic beats) but normal exercise test including exercise ECG can also participate in unrestricted leisure and school sport activities. (Class I, level of evidence C)

Children with haemodynamically significant residual lesions (left or right ventricular dysfunction, ventricular hypertrophy, or dilation; RV outflow tract stenosis with its subvalvular, valvular and supravalvular sites with an echocardiographical gradient of >30 mm Hg;¹²⁶ more than mild neo-aortic insufficiency; supra-ventricular or ventricular arrhythmia) can take part in leisure sport activities following the individual advice of the paediatric cardiologist. In case of normal exercise testing including exercise ECG, participation in leisure and school sport to voluntary exhaustion is allowed. High static load exercise should be avoided.⁴ (Class I, level of evidence C)

Competitive sport. Children and adolescents without or with minor remaining findings (see above) and normal exercise test including exercise ECG can participate in all categories of competitive sport.^{2–4,134,135} However, activities with a combined high dynamic and high static component are not recommended.^{3,4} (Class I, level of evidence C)

Children with haemodynamically significant remaining findings (see above) are generally not advised to take part in competitive sports. According to the individual advice of the paediatric cardiologist, and provided that their exercise test including exercise ECG is normal, they might participate in sports with low-to-moderate static and dynamic load. (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. Though the majority of children and adolescents after asoTGA presents without remaining haemodynamic lesions, subnormal exercise capacity may be prevalent predominantly due to parental overprotection^{134,136} and reduced physical activity patterns¹²⁹ (Hövels-Gürich et al., unpublished). In order to prevent or treat psychomotor and psychosocial deficiencies and to reduce atherosclerosis lifestyle risk factors¹³⁷ such as a sedentary lifestyle and overweight, at least 60 min of daily moderate or vigorous activity have been advised.¹³⁸ A structured training programme should be identical with that in unconditioned healthy children (Table 2). (Class I, level of evidence C)

Congenitally corrected transposition of the great arteries (ccTGA)

General considerations. CcTGA is a rare lesion where the left atrium is connected to the right ventricle and then to the aorta, whereas the right atrium is connected to the left ventricle and consecutively to the pulmonary artery (PA), leaving the right ventricle as the systemic ventricle. Associated abnormalities such as a VSD, LV outflow tract (LVOT) obstruction, and anomalies of the left-sided systemic tricuspid valve are present in >90%. There is increasing risk of developing considerable systemic AV-valve regurgitation, consecutive systemic RV dysfunction and heart failure, and supraventricular tachycardia as well as spontaneous complete heart block with age. Conventional surgery aiming at physiological repair (e.g. AV-valve replacement, VSD-closure, LVOT-PA conduit) has been disappointing. Novel surgical approaches aiming at restoration of normal AV and VA connections (double switch) seem more promising.¹³⁹

On the other hand, there is an unknown number of patients with ccTGA that live a normal life without suspicion of a cardiac defect.

Exercise capacity in ccTGA patients. The extent of the concomitant lesions is predominantly responsible for the grade of exercise intolerance. Literature on exercise performance is scarce: one small study in children¹⁴⁰ and a few studies in adults with isolated ccTGA^{141–144} report on markedly diminished $\dot{V}O_{2peak}$ (average 59%, range 30–84% of predicted normal). Chronotropic incompetence and impaired stroke volume response of the systemic RV are thought to be the prevalent causes. RV dysfunction has been found associated with increased RV filling pressure on tissue Doppler imaging,¹⁴² with reduced coronary flow reserve on positron emission tomography,¹⁴⁵ and with myocardial fibrosis on cardiac MRI.¹⁴² $\dot{V}O_{2peak}$ and exercise cardiac index have been found comparable between patients with

asymptomatic isolated ccTGA and those after intra-atrial repair for simple TGA;^{143,146} the capacity to increase RV stroke volume during exercise has only been demonstrated in ccTGA, possibly due to the absence of a rigid atrial baffle.¹⁴⁶ B-type natriuretic peptide (BNP) can be high in some patients and the increase of BNP correlates with deterioration of clinical status and decreasing exercise capacity.¹⁴⁷

Recommendations for clinical practice

Preparticipation screening. Before participation in any sportive activity, children with ccTGA must be extensively evaluated by a paediatric cardiologist with respect to: exact medical history, NYHA classification, physical cardiological examination, BNP assessment, standard ECG, Holter ECG, echocardiography, exercise test, and in selected cases cardiac MRI (assessment of ventricular function, delayed enhancement). At least yearly re-evaluation is suggested.

Leisure sport and daily physical activities (recreational activities). Children and adolescents with ccTGA are generally recommended to take part in playful leisure exercise. Playing with friends and siblings does hardly bear significant risks from overexertion, as children are able to limit their level of activity by self-chosen breaks. Recommended types of exercise include interval training at low-to-moderate intensity, e.g. ball games and running games, or low-to-moderate endurance exercise training. Types of sports with a high static component are not appropriate.¹³⁴ If possible, participation in school sports without competitive pressure should be achieved. (Class I, level of evidence C)

Competitive sport. Asymptomatic patients with isolated ccTGA without concomitant cardiac lesions and absence of systemic ventricular dilation or dysfunction and atrial or ventricular tachyarrhythmia, who have a normal exercise response in terms of workload, heart rate, ECG, and blood pressure response, might be eligible for competitive sport with low-to-moderate dynamic and low-to-moderate static activity. Periodic re-evaluation is necessary. (Class I, level of evidence C)

The vast majority of children and adolescents with ccTGA is not recommended to participate in competitive sports because of the abnormal responses to exercise related to the systemic position of the right ventricle, the risk of increasing AV valve regurgitation and cardiac arrhythmia.^{2,3,135} (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. Systematic physical training has been demonstrated to increase endurance and workload

achieved at a given HR_{peak} and $\dot{V}O_{2peak}$ in patients with systemic right ventricle.¹⁴⁸ Children with ccTGA can profit from participation in so-called children's heart sport groups, providing the opportunity to be physically active ruled by a sports therapist under medical supervision. By that, additional psychomotor and psychosocial deficiencies can be successfully treated.¹³⁴ Asymptomatic children and adolescents with isolated ccTGA can undergo a structured training programme based on the individual exercise capacity and adapted to levels of moderate-to-high dynamic and low-to-moderate static load. (Class I, level of evidence C)

Children after the Fontan procedure

General considerations. The Fontan procedure is used to palliate a variety of complex congenital heart defects that are not amenable to biventricular repair. Through a series of procedures, the caval veins are connected directly to the pulmonary arteries so that the functional single ventricle remains to support the systemic circulation. Although the Fontan procedure has enabled survival with a functional single ventricle, there are a variety of sequels and complications that result in altered exercise physiology. In a review of the exercise training literature after Fontan, Takken et al.¹⁴⁹ indicated that there is wide variability in $\dot{V}O_{2peak}$ among these patients, but most studies have reported an average exercise capacity that is reduced compared to healthy peers. The theoretical explanations provided for the reduced exercise capacity range widely and include the presence of chronotropic incompetence, diminished peak stroke volume due to reduced pulmonary venous diastolic return, the effects of chronic ventricular volume overload, impaired ventricular systolic contractile function, and altered pulmonary blood flow or respiratory gas exchange. More recently, it has been suggested that decreased or altered skeletal muscle function may also contribute to the reduced maximal exercise capacity of these children.¹⁵⁰

The influence of patient management factors on exercise capacity. The many factors that may influence exercise capacity will be different for each patient who has had the Fontan procedure. Therefore, each patient's exercise capacity must be assessed in light of their unique physiology, anatomy, and surgical history. At the most basic level, long-term outcomes are influenced by the morphological structure of the ventricle, the energy efficiency of the cavopulmonary connection, and the presence/absence of aortopulmonary collateral arteries.¹⁵¹ Exercise capacity will also be influenced by complications associated with the surgical procedures, such as arrhythmias or thromboembolic events. Finally, exercise capacity may be influenced by

longer-term sequels, such as myocardial dysfunction or damage to the pulmonary vascular bed.¹⁵¹ Whether or not the patient's exercise capacity is reduced, many of the ongoing management practices for these patients may also have an impact on their physical activity and exercise participation. Patients who have implanted devices (pacemakers or internal cardiac defibrillators) and those who require anticoagulation medication should not participate in activities that might result in damage to the device or bleeding injuries. Some of these patients have congenital and acquired central nervous system abnormalities, resulting in delays of neurodevelopment and cognitive impairments, particularly in relation to visual motor integration,¹⁵¹ which also will influence their exercise capacity and physical activity participation.

Recommendations for clinical practice

Leisure sport and daily physical activities (recreational activities). In spite of the known limitations regarding exercise capacity and physiology, a large cross-sectional study of children and adolescents after Fontan procedure found no relationship between exercise capacity and physical activity participation.¹⁹ Although initially surprising, a closer examination of the difference between physical activity and maximal exercise capacity offers an explanation of this result. Activities that can be performed for an extended period of time typically require 40–60% of maximal capacity. Activities that require 60–80% of maximal capacity produce a training effect (i.e. increased fitness). On average, those patients who have had the Fontan procedure have approximately 70% of the exercise capacity of their healthy peers.¹⁴⁹ Therefore, despite the limitations for maximal exercise, the Fontan procedure typically provides sufficient capacity to allow these patients to participate in the types of physical activity that are typical of daily life. Recent research¹⁵² suggests that higher levels of moderate-to-vigorous physical activity are associated with taking antithrombotic medication, warm weather seasons of the year, male gender, and better motor skills. While the influence of season and gender are well recognized, these results suggest that perhaps the specific physical activity counselling provided to children taking antithrombotic medication may encourage increased levels of daily physical activity by emphasizing lifetime activities such as biking, hiking, and swimming.

The following recommendation can be made regarding exercise participation in this group:

1. Patients should be encouraged to perform moderate-to-vigorous activity on a daily basis in accordance with public health recommendations for optimal health (typically ≥ 60 min/day). Healthcare professionals

should be strong advocates for the importance of daily physical activity so that patients who have the Fontan procedure can obtain the many benefits associated with physical activity (e.g. optimal growth and development, peer socialization, establishment of healthy lifestyle habits). (Class I, level of evidence C)

2. Patients involved in formal or organized physical activity programmes should have a full cardiopulmonary exercise test to provide specific information regarding their exercise capacity. The baseline assessment should also include assessments for arrhythmias and ventricular and valvular function. Formal or organized physical activity should not be restricted unless the patient has an implanted device, is taking antithrombotic medication, or restrictions are warranted based on the results of the baseline assessment. (Class I, level of evidence C)

3. Patients not involved in formal or organized physical activity programmes should not be restricted and should be actively encouraged to participate in a wide variety of physical activities. (Class I, level of evidence C)

4. Patients (and their caregivers) should be taught to monitor their activity intensity. No restriction of physical activity is necessary as long as the patient is able to rest as needed. Healthcare professionals should ensure that the patient (and their caregivers in the case of a child) understands the specific signs and symptoms that signal the need for rest. Typically such signs and symptoms may include palpitations, chest pain, tachycardia, dyspnoea that is disproportionate to the exercise effort, nausea, and presyncope. However, the cardiologist responsible for each patient should determine the specific criteria for a temporary cessation of physical activity based on the individual's current status and treatment history. Patients should not participate in physical activity settings if anyone, including the patient, caregiver, coach, other participants, or spectators, can reasonably be expected to encourage the patient to continue exercising in spite of signs or symptoms indicating that a rest period is required. (Class I, level of evidence C)

5. Patients with implanted devices or taking antithrombotic medication should be limited to sport activities with low risk of impact or trauma. (Class I, level of evidence C)

Competitive sport. 1. Single patients can engage in low-to-moderate dynamic and low static competitive sports on an individual medical decision if they have no symptoms on exercise and normal ventricular function and oxygen saturation. (Class I, level of evidence C)

2. Patients with implanted devices or taking antithrombotic medication should be limited to sports with

low risk of impact or trauma. (Class I, level of evidence C)

Exercise training as preventive/therapeutic intervention. The six studies reviewed previously¹⁴⁹ demonstrate that, after the Fontan procedure, patients are able to improve their exercise capacity through training. Each of these studies^{150,153–157} involved a small number of participants ($n=1–16$) performing aerobic exercise training, with or without resistance training, in accordance with the FITT principle. These studies demonstrate that patients who have the Fontan procedure can significantly increase their $\dot{V}O_{2peak}$ through systematic exercise training. Recommendations for exercise training are as for the general population (Table 2).

However, since exercise capacity is not related to objective measures of physical activity in these patients,¹⁹ whether such training can increase the daily moderate-to-vigorous physical activity recommended for optimal health benefits¹⁵⁸ is currently unknown. A preliminary report (available as abstract only) suggests that home-based physical activity counselling can increase moderate-to-vigorous physical activity participation after the Fontan procedure.¹⁵⁹

Cyanotic patients/Eisenmenger syndrome

General considerations. Cyanosis in general is caused by shunting of deoxyhaemoglobin into the arterial system. In several lung diseases this is caused by anatomical intrapulmonary arteriovenous fistulae or by a functional shunt through the capillaries, when they are not able to increase oxygen content in the blood properly. In cyanotic congenital heart defects there are several other mechanisms for right to left shunting. Most isolated shunts in congenital heart defects are seen within the same level of the systemic and pulmonary circulation like atrial septal defect, ventricular septal defect, aortopulmonary window, or persistent arterial duct. Initially there is a left to right shunt with pulmonary recirculation of oxygenated blood. Only if stenotic or atretic structures downstream to the lungs cause a rise in the prestenotic pressure, right to left shunting and cyanosis can appear. In the long term, patients with isolated shunts can develop progressing pulmonary arteriolar remodelling which will increase pulmonary vascular resistance and lead to shunt reversal and cyanosis. Last but not least, many patients with complex congenital heart defects, discordant connections of vessels and/or cardiac chambers may have a complete mixing of the systemic and pulmonary venous return (parallel circulation instead of serial circulation) which leads to cyanosis.

Nowadays, most of the CHD are corrected by surgery or catheter intervention early in childhood. However, there are still some cyanotic patients surviving into adulthood without surgery¹⁶⁰ or with palliative surgery, because pulmonary vascular disease is already established.

It is important to note that there are almost no published data on exercise participation in this patient group. There are only small observational studies on exercise physiology. Most publications in that field are summaries and recommendations without data supporting them. So one is prone to transfer data from patients with similar pathophysiological entities. Especially in patients with pulmonary vascular disease, there are data from patients with idiopathic pulmonary arterial hypertension (iPAH). However, these available data must be handled with care as they resemble only some findings of the disease (i.e. increased pulmonary vascular resistance) and not the complete haemodynamic situation.

Recommendations for clinical practice

Preparticipation screening. The exact knowledge of the medical history, the sequence of interventions as well as a history of arrhythmia or existence of syncope-like symptoms is essential to give advice to these patients. Current imaging with echocardiography as well as data from a MRI or helical CT are necessary to answer all anatomical details that might have an influence on exercise physiology. Also recent haemodynamic data from a left and right heart catheterization should be available. To rule out dangerous arrhythmia, at least a current resting ECG and a 24-h Holter ECG are mandatory.

Additionally, a cardiopulmonary exercise test must be performed. These data can be supplemented by a stress echocardiography, stress MRI, or even stress testing in the catheter laboratory.

As all these patients are rather unique, exercise counselling should only be performed by congenital cardiologists, experienced in both the haemodynamics of these patients and in sports cardiology.¹⁶¹

Leisure sport and daily physical activities (recreational activities). Every congenital cardiologist has experienced pulmonary hypertensive crises in the postoperative management of patients with pulmonary hypertension.¹⁶² Despite the fear to provoke a pulmonary hypertensive crisis, no adverse events during laboratory exercise tests are published.^{163–165} Also, none of the 61 deaths of patients with Eisenmenger syndrome reported by Daliendo et al.¹⁶⁶ were related to sport or exercise participation.

This is in contrast to patients with iPAH or PAH from corrected CHD without current shunt. The lack of shunt causes the right heart to fail, the systemic cardiac output to drop, and the patient to develop a syncope. Therefore, leisure sport and physical activity in general should only be allowed as long as no symptoms appear and cyanosis remains acceptable. However, inadequate dyspnoea or even slight signs of cerebral malperfusion like dizziness or discomfort must be avoided. Patients with PAH without shunt should avoid any excessive physical activity that leads to distressing symptoms.¹⁶⁷ (Class I, level of evidence C)

Competitive sport. The 36th Bethesda recommendation allows competitive sport in all fields only with a peak systolic pulmonary arterial pressure of ≤ 30 mmHg.¹ All other patients with PAH should be individually counselled.

For patients with un-operated cyanotic defects, the recent Bethesda recommendation for participation in competitive sport allows low dynamic/low static disciplines.¹ For palliated cyanotic patients participation in competitive sports is only allowed in low dynamic/low static disciplines, if arterial oxygen saturation remains above 80%, tachyarrhythmia with symptoms of impaired consciousness are not present and ventricular function is not or only mildly impaired.

Exercise training as preventive/therapeutic intervention. Regular exercise training improves endothelial function in the systemic vessels even in severe heart failure patients¹⁶⁸ and improves survival.¹⁶⁹ An animal model,¹⁷⁰ trying to translate these improvements to the pulmonary vessels, failed to show a positive effect of exercise training in pulmonary hypertensive rats.

However, recently patients with idiopathic or chronic thromboembolic pulmonary hypertension¹⁷¹ profited from exercise training in respect to an increased 6-min walking distance, $\dot{V}O_{2\text{peak}}$, ventilatory anaerobic threshold, and peak work load. Additionally, quality of life improved. No adverse events were reported. Nevertheless, no change in the invasive haemodynamics could be found. It has, therefore, been hypothesized that exercise training may mainly result in an improvement of the peripheral exercising muscles, with no change in the pulmonary vascular bed.¹⁷² However, even in stable PAH patients the therapeutic range is rather small and the proof for a better survival is still missing. After this human study was published, a re-evaluation of the rat model showed that rats with stable PAH tolerated exercise training well and profited in terms of an increased exercise capacity and myocardial capillary density, whereas rats with progressive PAH had leukocyte lung infiltration,

Table 6. Recommendations following the FITT principle for exercise training in children with cyanosis/Eisenmenger (Class IIb, level of evidence C)

FITT	Interval training	Muscle (resistance) training	Endurance training	Passive physical therapy
Frequency	3–7 times a week	3–5 times a week	3–7 times a week	3–7 times a week
Intensity	Interval training with baseline at moderate intensity (40–59% of $\dot{V}O_{2peak}$) and short intervals (30 sec) of endurance level (up to 85% $\dot{V}O_{2peak}$)	Low intensity (i.e. 500–1000 g Dumbbells), 10 repetitions for separate small muscle group	Moderate intensity (40–59% of $\dot{V}O_{2peak}$)	Mainly passive
Time	10–30 min	In total up to 30 min strength training	60 min	30 min
Type	Bicycle	Dumbbell training.	Walking	stretching, breathing technique, body perception/relaxation

Distressing symptoms especially those of excessive dyspnoea or cerebral malperfusion must be avoided; SpO₂ must be monitored and should be >80% throughout exercise; The patient must be continuously supervised by a physician or experienced physiotherapist (adapted from Mereles et al.¹⁷¹)

increased PAH, and shortened survival.¹⁷³ This study outlines the importance of a stable situation before starting an exercise programme in PAH patients.

For patients with cyanotic CHD or Eisenmenger syndrome, there is only a nonrandomized controlled trial with four patients in the exercise group.¹⁷⁴ In summary this study showed that low-grade exercise training similar to that study in iPAH and thromboembolic PAH,¹⁷¹ with 24-min interval bicycle training (base 10–25 Watt, peaks 30 sec and 20–50 Watt) after a 10-min warm-up, for twice a week over 3 months could be performed without complications in these four patients. But no objectively measurable improvements were seen beyond those in functional class.

With the lack of data and having physiology in mind, one can only speculate that the risk of syncope might be smaller in patients with cyanotic heart disease and persisting right to left shunt than in iPAH patients as the shunt secures LV filling and systemic cardiac output. But there is doubt, whether the progressive cyanosis during exercise hampers the positive effects of exercise and whether improvements in systemic vascular function even worsens cyanosis. At the moment, rehabilitative exercise training in these patients should be limited to scientific studies in centres of congenital cardiology.

Patients with cyanotic congenital heart disease must curtail physical activity within symptom limits. Patients have to avoid excessive physical activity

that leads to distressing symptoms, especially symptoms of excessive dyspnoea or signs of cerebral malperfusion (i.e. discomfort, dizziness). (Class I, level of evidence C)

Rehabilitative low-intensity exercise training might be considered in patients:

1. in a stable haemodynamic situation,
2. with an arterial oxygen saturation >80% during exercise,
3. without symptoms of excessive dyspnoea or cerebral malperfusion at training,
4. under close surveillance of a paediatric/congenital cardiologist,
5. preferably included in a scientific protocol. (Class IIb, level of evidence C)

The recommendations following the FITT principle for exercise training in children with cyanosis/Eisenmenger are shown in Table 6.

Heart transplanted children

General considerations. Over the last 25 years, with introduction of the current immunosuppressant regimen, paediatric solid organ transplantation has increased dramatically. Heart, lung, and heart and lung transplantation increased steadily in numbers

from the latter half of the 1980s. The numbers have subsequently levelled in the USA at approximately 350–400/year reflecting limitations of donor availability. Long-term survival has generally been quite good and mirrors results in adult populations. Recent reports of actuarial survival at 15 and 20 years have been as high as 80 and 53%, respectively.¹⁷⁵

Amazingly, there are very little data on exercise performance in children and adolescents following heart transplantation. There are even less data available on the effects of exercise training and cardiac rehabilitation for this population. Recommendations for either recreational or competitive sports participation are nil. The reason for the paucity of data include a relatively small number of subjects compared to the adult transplant population, difficulties with exercise rehabilitation programmes for small children, and the mistaken belief that many of these children have no need for such programmes. What little data that are available dispel all of these notions. Unfortunately we cannot, as is so often the case, draw on studies from the much larger adult transplant population to help fill our knowledge gap. This is because there is an equal paucity of evidence-based recommendation for sports and exercise participation in this population. However, data from the adult population provide useful information on the nature of exercise limitations and the effects of training and rehabilitation programmes that may be extended to the paediatric population.

Exercise capacity in the heart transplant population. Exercise capacity as measured by both aerobic capacity and musculoskeletal strength is significantly decreased in the paediatric and adult populations following heart transplantation. The causes of these limitations are multifactorial. The few reports of exercise testing in paediatric heart transplant patients have reported aerobic capacities, as measured by $\dot{V}O_{2peak}$, of 50–60% of healthy age- and sex-matched peers.^{176–178} These values are not significantly different from those reported in the adult population.^{179–181} The reasons for this finding appear to be due to both central and peripheral factors combining to impair aerobic capacity. Especially in early post-transplant periods, stroke volume is limited. This may be due to systolic impairment but more importantly to diastolic dysfunction with high cardiac filling pressures. Diastolic abnormalities persist and even late after transplant an inability to maintain adequate ventricular preload appears to limit cardiac output.

Abnormalities of autonomic innervation and function also impact on cardiac output during exercise. At least initially there is a loss of autonomic

innervations to the heart. This significantly decreases chronotropic reserve as well as blunts the time course of the chronotropic response. There is some evidence for re-innervation and improved chronotropy late after transplant in some patients or as a response to cardiac training as will be discussed below. In addition to the cardiac effects, autonomic tone is abnormal in the peripheral vasculature. Brachial reactivity is impaired and systemic vascular resistance is increased.^{182–185}

Limitations of the peripheral exercising musculature are most likely at least as important as central mechanisms in limiting aerobic capacity. Following heart transplant, skeletal muscle mass is often reduced by 20% of normal. Capillary density is also significantly decreased. This may reflect the marked deconditioning in these patients that occurs prior to transplantation but may also be the result of immunosuppressant therapy. These changes result in an impaired ability of the exercising muscle to extract oxygen. Muscle strength is significantly impaired especially in the early transplant period. In addition to muscle mass, bone demineralization is a frequent finding. This may result in stress and compression fractures. Ongoing immunosuppressant medications may continue to exacerbate the problem of demineralization.^{186,187}

Serial studies of exercise performance following paediatric heart transplants are limited. Recent studies of Davis et al.¹⁷⁶ and Dipchand et al.¹⁷⁸ are conflicting. Both show early improvement in aerobic capacity and working capacity. Davis saw a decline after about 3 years of improvement while Dipchand's population remained steady with some patients showing a decrease associated with the onset of graft vasculopathy. The reason for these findings are unclear but are probably the combined improvement of systolic and especially diastolic function in the immediate post-transplant period as well as the longer-term improvement in musculoskeletal conditioning even in the absence of formal rehabilitation. In addition, improved chronotropy suggests at least some patients benefit from autonomic re-innervation of the donor heart.

Recommendations for clinical practice

Leisure sport and daily physical activities (recreational activities). There are no evidence-based studies assessing the types, safety, or benefit of sports and exercise participation in children following heart transplantation. In a very small interview study of mixed solid organ transplant patients, Olausson et al.¹⁸⁸ reported that most expressed the opinion that they lived normal lives. Ross et al.¹⁷⁵ reported on the long-term survival

of a cohort of paediatric heart transplantation patients at 10 and 20 years post transplant. They state that 'physical rehabilitation and return to normal lifestyle has been nearly 100%'. They cite two anecdotal cases of patients participating in vigorous competitive sports. In an editorial, Fricker¹⁷⁹ listed a series of recommendations for paediatric heart transplantation patients (Class I, level of evidence C):

1. Exercise should be encouraged not discouraged.
2. All patients should be in a monitored rehabilitation programme within 3 months following transplantation.
3. There should be a return to age-appropriate activities, including physical education class within 6 months after transplantation.
4. Endurance activities will be better tolerated than intermittent high-intensity activity.
5. Participation in competitive sports should be individualized with detailed yearly re-evaluation of participation.

Competitive sport. There is an equal lack of data in the adult population regarding competitive sports participation following heart transplantation. There are a number of case reports and small studies showing that heart transplant recipients can train and compete often quite successfully in vigorous athletic activity. However, the numbers in these studies are too small to generalize to the entire transplant population regarding the safety and benefits of such training. Given these limitations, the recommendations from the 36th Bethesda Conference on Eligibility Recommendations for Competitive Athletes with Cardiovascular Abnormalities are probably appropriate.¹⁸⁰ (Class I, level of evidence C):

1. Because of special issues involved with transplant patient management, decisions as to the feasibility of athletic competition for cardiac transplant recipients should be made in conjunction with the patient's transplant cardiologist.
2. Athletes with no coronary luminal narrowing, no exercise-induced ischaemia, and normal exercise capacity for age can generally participate in all competitive sports as appropriate for their exercise capacity.
3. Athletes with coronary luminal narrowing should be risk stratified as outlined in the recommendations by Thompson et al.¹⁸⁰

In the absence of any better data, these recommendations are probably a good basis, at least, for the evaluation of the adolescent population as well.

Exercise training as preventive/therapeutic intervention. From the limited exercise testing data in the paediatric population as well as the significantly more extensive adult data, it is clear that the majority of patients with heart transplants would benefit from a structured exercise rehabilitation programme. (Class I, level of evidence B for adult studies, and level C for paediatric studies). Studies of rehabilitation programmes in adults have shown $\dot{V}O_{2peak}$, muscle strength, and endurance as well as bone mineral density can all be improved by formal exercise programmes both in the immediate post-transplant period and even years after transplant.^{181,182,186,189,190} The improvement in $\dot{V}O_{2peak}$ may average anywhere from 20 to 50% depending on the individual study. Studies that assessed the mechanisms by which $\dot{V}O_{2peak}$ was improved with exercise training showed that both central and peripheral factors were involved. The increased ability of the exercising muscles to more effectively extract oxygen and widen arterial/venous oxygen content difference appeared to be at least as important as improvement in cardiac output in the changes seen in adult transplant patients following an exercise rehabilitation programme.

The structure of the adult rehabilitation programmes is all fairly similar. They are generally 12–16 weeks in duration with sessions a minimum of 3–5 times/week. They include both endurance training as well as light resistance training. Although home-based programmes can show some improvement, it is important to note that all studies that examined supervised versus home programmes found significantly superior results with supervised programmes. Long-term programmes of up to a year have demonstrated sustained improvements in $\dot{V}O_{2peak}$ and quality of life.^{181,189,190}

There is an extreme lack of data regarding the benefits of exercise rehabilitation programmes in paediatric heart transplant patients. There are no significant data regarding the efficacy of rehabilitation in the immediate post-transplant period in children and minimal data on those who are remote from the time of transplant. The lack of studies on this population may be the result of several factors. These include difficulties in providing appropriate rehabilitation facilities for a structured supervised programme as well as poor compliance and willingness to participate in either a supervised or a home-based programme. In addition, the perception that these patients are doing well and do not require any formal rehabilitation programme may also be a significant factor. The only study in paediatric heart transplant patients is a relatively recent study by Patel et al.¹⁷⁷ This study was a quite small ($n=11$) home-based programme. It was 12 weeks in duration and included both endurance and strength training. The time from transplant was an average of 5.26 years. There was an approximately 15% increase in $\dot{V}O_{2peak}$

as well as improvement in strength for most major muscle groups.

A suggested training programme should be similar to that in healthy unconditioned children. However, resistance training should be limited to moderate intensities.¹⁹¹

Conclusions and final remarks

In this paper recommendations for exercise participation in children and adolescents with CHD are provided. In general, children with CHD should be advised to comply with public health recommendations of daily participation in 60 min or more of moderate-to-vigorous physical activity that is developmentally appropriate and enjoyable and involves a variety of activities.

As can be appreciated from the low levels of evidence (mostly level C), most of the provided recommendations are based on expert opinion or, in a few cases, on small studies. This shows that further research is needed in this area. Studies with a more rigorous research design (e.g. larger sample size, control group, blinded observers, attractive and controlled exercise training programmes) and a longer follow up are indicated. In addition, the effect of exercise training on quality of life, peripheral muscle function, and physical activity levels needs to be evaluated.

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