CHAPTER 29

Exercise, physical activity, and congenital heart disease

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Introduction

Exercise testing in adult cardiac patients has mainly focused on ischaemic heart disease. The results of exercise testing with ECG monitoring are often helpful in diagnosing the presence of significant coronary artery disease. In children with heart disease, the type of pathology is different. Ischaemic heart disease is very rare. The majority of patients will present with congenital heart defects that will affect exercise capacity. In patients with congenital heart disease, exercise tests are frequently performed to measure exercise function or to assess abnormalities of cardiac rhythm. The risk of exercise testing is very low in the paediatric age group.[1] Findings from exercise testing show a reduced exercise capacity in many children with congenital heart disease and exercise responses differ between patients with different underlying heart defects. Results from exercise testing can provide insight into the clinical condition of the child and help decision making regarding patient management. However, exercise testing not only provides information about the heart, but also about the level of daily physical activity (PA). In general, PA has been found to be reduced in children with congenital heart disease, and below recommended guidelines. This has resulted in a shift in research attention from evaluating exercise responses of children with congenital heart disease towards investigating means to improve PA and exercise capacity for health.

Commonly used parameters to assess exercise performance and aerobic exercise function in children with cardiac disease

In exercise physiology, aerobic exercise performance is traditionally assessed by determination of the *maximal oxygen uptake* ($\dot{V}O_2$ max). This reflects the highest level of oxygen which does not further increase, despite an increase in exercise intensity. In paediatric exercise testing the $\dot{V}O_2$ max is frequently assessed by means of an incremental exercise test until voluntary exhaustion. However, although the measurement of $\dot{V}O_2$ max is useful, since it gives information about maximal exercise tolerance, the physiological definition of $\dot{V}O_2$ max is not always met in children. Only about 50% of children are able to reach such a plateau after repeated exercise tests.[2] Many children are not motivated to exercise to that point of exhaustion.[2] Therefore, other criteria should be used to confirm a maximal exercise effort such as i) a respiratory gas exchange ratio ($\dot{V}CO/\dot{V}O_2$) > 1.10, ii) a peak heart rate (HR) which is close to 200 beats·min^{-1} and, iii) the subjective appearance of exhaustion. When one or more of these criteria are met, it is assumed that the peak $\dot{V}O_2$ is a maximal value. (Interested readers are referred to Chapter 12 for further discussion of peak $\dot{V}O_2$ and $\dot{V}O_2$ max during paediatric exercise testing). As shown in Figure 29.1a, peak $\dot{V}O_2$ is often reduced in children with congenital heart disease.

Additionally, the evaluation of the HR max provides important information. Some patients with congenital heart disease present with chronotropic limitation or are unable to raise their HR as expected according to the increase in workload.[3] Because of that, the HR max cannot always be used as a criterion for evaluating the maximal character of an exercise test in congenital heart disease.[2] Figure 29.1b shows average peak HR values for children with various congenital heart defects.

The *oxygen pulse* equals stroke volume (SV) times arteriovenous oxygen difference and is calculated by dividing $\dot{V}O_2$ by HR. In healthy subjects, the oxygen pulse continuously increases during submaximal exercise. Furthermore, the oxygen pulse has been used as a surrogate for SV during exercise and has been shown to correlate with SV during submaximal exercise.[4–7] The response of the oxygen pulse to exercise in children can be assumed to be somewhat different from the response in adults, but also in children, this parameter has been proposed as an indirect measure of SV.[8] Due to the child's smaller heart and total blood volume, SV is lower both at rest and during exercise.[9] A child can compensate for this by a higher HR response to submaximal exercise in comparison with adults. Oxygen uptake is further increased by a higher increase in arterial-mixed venous oxygen difference during submaximal exercise in children compared to adults. When the oxygen pulse is used as an indicative parameter for the evaluation of SV during graded exercise, the arterial-mixed venous oxygen difference is considered to remain fairly constant. Therefore, in children with congenital heart defects, the course of the oxygen pulse during an incremental exercise test gives an idea of the changes in SV in response to the increased workload and can provide interesting information regarding the patients' actual clinical condition.

In healthy children and adults, the blood remains fully oxygenated, even during maximal exercise. However, in congenital heart disease, the evaluation of *oxygen saturation during exercise* can provide clinicians with important information. When patients desaturate, this happens in a steady, progressive manner with the lowest numbers observed in the first minute after the load is removed.

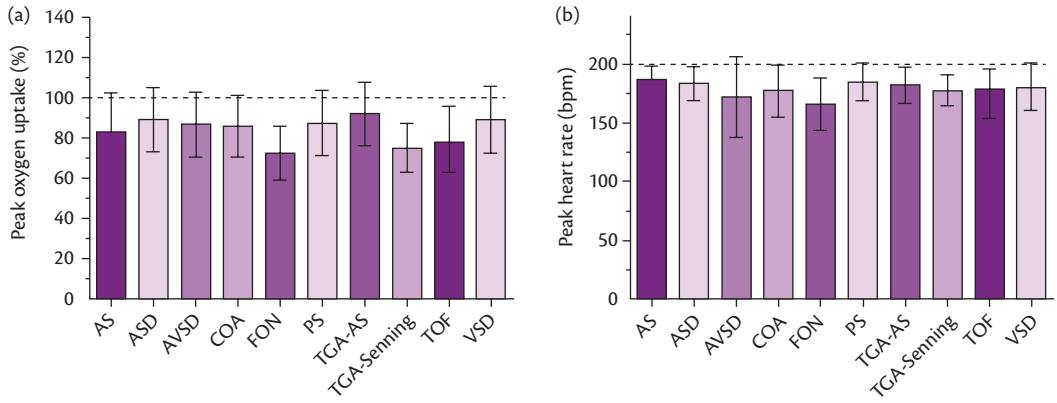

Figure 29.1 Peak oxygen uptake (a) and peak heart rate (b) in children with various congenital heart diseases.
AS, invasively treated aortic valve stenosis; ASD, surgically treated atrial septal defect; AVSD, surgically treated atrioventricular septal defect; COA, surgically treated aortic coarctation; FON, univentricular heart repaired with Fontan operation; PS, invasively treated pulmonary valve stenosis; TGA-AS, transposition of the great arteries surgically treated with the arterial switch operation; TGA-Senning, transposition of the great arteries surgically treated with the senning operation; TOF, surgically corrected tetralogy of Fallot; VSD, surgically treated ventricular septal defect. Dotted line represents peak $\dot{V}O_2$ of healthy children.

Because maximal exercise tests may have several drawbacks in the paediatric population, clinical investigators have tried to define exercise parameters that are independent from reaching maximal effort. In the past, *heart rate response to exercise* has frequently been used to assess cardiovascular exercise performance.[10,11] However, in patients with congenital heart disease several drawbacks exist, as many patients may show a relative bradycardia during exercise, which is not associated with a high value for $\dot{V}O_2$ max, as should theoretically be expected. Therefore, the use of the HR response to exercise in the assessment of cardiovascular exercise performance can be misleading in patients with congenital heart disease, and cannot be considered to be a valid determinant of aerobic fitness.[12]

A more sensitive assessment of aerobic exercise function can be obtained by analysis of gas exchange. Therefore, considerable attention has been focused on the determination of the *ventilatory anaerobic threshold* in children, which is a very useful and reproducible indicator of aerobic exercise function in the paediatric age group.[13,14] This parameter reflects the highest exercise intensity at which a disproportionate increase in CO_2 elimination ($\dot{V}CO_2$) is found relative to $\dot{V}O_2$.[14,15] (Interested readers are referred to Chapter 10 and Chapter 12 for more detailed discussions of anaerobic and ventilator thresholds).

More recently, newer concepts have been developed to assess dynamic changes of respiratory gas exchange during exercise in patients with congenital heart disease.

The study of the steepness of the slope of $\dot{V}CO_2$ *versus* $\dot{V}O_2$ *above the ventilatory anaerobic threshold*, has been found to be a very sensitive and reproducible index for the assessment of cardiovascular exercise function in patients with congenital heart disease.[15]

Another objective and effort-independent parameter is the *oxygen uptake efficiency slope* (OUES), which was introduced as a measure of exercise capacity by Baba *et al.*[16] The OUES represents the slope of the semilog plot of minute ventilation ($\dot{V}_E$) versus $\dot{V}O_2$ (see Figure 29.2). Thus OUES provides an estimation of the efficiency of $\dot{V}_E$ with respect to $\dot{V}O_2$, steeper slopes indicate a larger exercise capacity. The OUES has been shown to correlate highly with peak $\dot{V}O_2$ and to linearly increase with age during childhood and into adolescence.[17]

Also the $\dot{V}_E / \dot{V}CO_2$ *-slope* is supposed to be independent of achieving maximal exertion. This measure represents the slope of the regression line between $\dot{V}_E$ and CO_2 production. It provides information regarding ventilatory efficiency during exercise. A high $\dot{V}_E/\dot{V}CO_2$ slope observed in patients with repaired, noncyanotic congenital heart disease can often indicate hypoperfusion due to an impaired cardiac output response to incremental exercise.[18] An inefficient ventilatory response to carbon dioxide will limit the exercise performance, as it indicates wasted ventilation. This ventilatory inefficiency has been related to worsening of the clinical condition of cardiac patients, as well as of mortality.[19,20]

The $\dot{V}O_2$ *versus exercise intensity* slope is a valid measurement of oxygen flow to the exercising tissues. Calculation of this slope can document limited oxygen flow to working muscles in congenital heart disease. As such a reduced slope of oxygen uptake versus exercise intensity constitutes another factor limiting exercise capacity as it reflects impaired oxygen delivery to the exercising tissues.[21]

In order to be able to answer important questions concerning the normality of exercise responses in patients, the results can be compared with normal reference values. It is recommended that each laboratory carefully chooses the normal values to which they compare the exercise tolerance, because geographical differences may

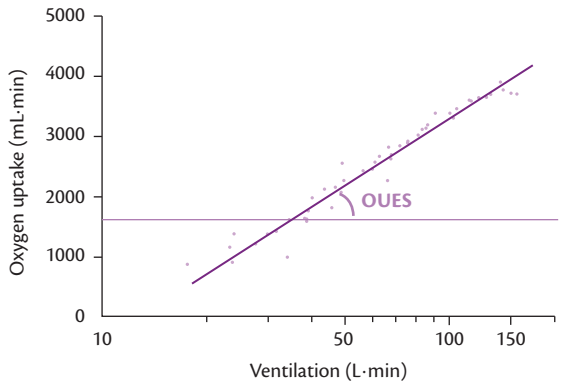

Figure 29.2 The oxygen uptake efficiency slope.
OUES, oxygen uptake efficiency slope.

influence the results of aerobic exercise performance. Reference values may also change over time and should be regularly updated and validated.

Cardiorespiratory response to exercise in specific congenital heart defects

Left-to-right shunts

Atrial septal defect

Children with atrial septal defect (ASD) usually have a normal or near normal exercise capacity. These children can attain normal or near normal values for $\dot{V}O_2$ max.[1,22] A number of haemodynamic abnormalities to exercise have been documented. The increase of cardiac output during exercise may be smaller than normal. Maximal HR response has been found to be lower than normal. In those who underwent surgical closure of the ASD, the age at surgery has been shown to influence exercise performance. In a consecutive series of 50 patients with ASD or ventricular septum defect evaluated in our laboratory, the ventilatory anaerobic threshold (as an estimate of aerobic exercise performance) was at the lower limit of normal (89 ± 14.4% of normal).[23] When studying the exercise response in children who underwent surgical closure of an ASD, a normal value was found in children who underwent surgery before five years of age, whereas a significantly lower value was found in children operated on after that age.[24]

In general, abnormalities detected in children either with unoperated or surgically closed ASD are usually minor and do not result in major limitations in exercise performance. Unless arrhythmia is a complication, these children should be encouraged to perform physical exercise and to participate in all sports at all levels. Exercise testing is generally indicated if symptoms of arrhythmia or dyspnoea on exercise are reported.

Ventricular septal defect

A small ventricular septal defect (VSD) will transmit only a small amount of blood from the left to the right side of the heart. Also during exercise, the shunt will remain small. Haemodynamic studies in this patient group showed that during graded exercise, patients with a VSD had a higher pulmonary circulation than systemic circulation, as could be expected. However, the relative shunt fraction decreased with increasing exercise intensity.[25] Subnormal values for cardiac output were found in this patient group. Studies during submaximal exercise testing, using gas exchange measurements, showed suboptimal values for ventilatory anaerobic threshold in a consecutive series of 43 patients with an unoperated VSD, evaluated in our laboratory. This value averaged 90 ± 15.3% of normal and was below the lower limit of the 95% confidence interval.[23] In this patient group, the decreased level of exercise capacity was correlated with decreased habitual PA. Finally, in a group of 18 patients who underwent surgical closure of a large VSD with pulmonary hypertension before age one year, the value for aerobic exercise performance was at the lower limit of normal (92 ± 17% of normal).[26] In a retrospective study about quality of life, Meijboom *et al.* [27] reported a normal exercise capacity in 84% of the patient group who underwent surgical closure of a VSD (n = 109). Also Binkhorst *et al.*[28] reported that children with patent or surgically closed VSDs have a normal exercise capacity, despite a mild chronotropic limitation in the latter. This shows that surgical correction of a congenital heart defect early in life can normalize the child's exercise performance.

Patent ductus arteriosus

Similarly, in patients with patent ductus arteriosus, results of exercise testing will generally be normal if the size of the shunt is moderate or small. These subjects will ordinarily be asymptomatic. In most conditions, these defects will be closed surgically or percutaneously, at an age when exercise testing is not feasible. Exercise testing will add little to the routine clinical evaluation of these patients.[1]

Valvular heart lesions

Aortic stenosis

Exercise testing in patients with aortic stenosis may show ST segment changes on the ECG, reflecting ischaemia, a drop in blood pressure, or an inadequate rise in blood pressure with increasing exercise intensity and eventually arrhythmia during exercise testing. The major haemodynamic determinant of ST segment changes during exercise is the inadequate oxygen delivery to the left ventricle. After relief of the gradient by surgery or balloon dilation, improvement of ST segment changes on the ECG during exercise has been reported.[29] A critical aortic stenosis can be identified by clinical findings and confirmed by echo-Doppler examination and eventually by cardiac catheterization.

During exercise testing, most of the patients show a reduced aerobic exercise performance which may be improved after surgery.[29] This may be related both to the inability of the cardiac output to increase adequately during exercise and also due to the effect of a medically imposed restriction of heavy PA and competitive sports. Sudden cardiac death (SCD) during exercise has been reported and has been ascribed to malignant arrhythmias. Fortunately, this is unlikely to happen in asymptomatic patients who present with mild to moderate stenosis.[22] In our experience, SCD has never occurred in follow up in over 200 patients with mild to moderate aortic stenosis (Doppler gradient < 60 mmHg). It allows them to perform up to moderate exercise, including recreational sports.

Pulmonary valve stenosis

Similarly to aortic stenosis, pulmonary valve stenosis results in a right ventricular overload. This may lead to a diminished pulmonary flow. In mild cases (gradients < 30 mmHg) normal or near normal values for ventilatory threshold have been found.[30]

During exercise the transvalvular pressure gradient in pulmonary stenosis may increase during graded exercise testing.[1] In mild cases (gradients < 30 mmHg), values for ventilatory anaerobic threshold have been found to be at the lower limit of normal.[30] In cases with moderate to severe pulmonary stenosis, right ventricular pressures may rise considerably during exercise, which may limit exercise capacity.[31] In patients with mild to moderate pulmonary stenosis (Doppler > 50 mmHg), relief of the stenosis results in an improvement of exercise tolerance. However, exercise performance may be limited in cases with severe pulmonary incompetence.

Cyanotic heart disease

Tetralogy of Fallot

Children who have undergone surgical repair of Tetralogy of Fallot (ToF), and who are felt postoperatively to have good results (no residual VSD and a pressure gradient between right ventricle and

pulmonary artery below 20 mmHg), are generally asymptomatic at rest. However, a variety of abnormalities may be brought out by intensive exercise.[32] These include a high right ventricular pressure with values as high as 100 mmHg during maximal exercise, caused by a pressure gradient between right ventricle and pulmonary artery, a blunted increase in SV and HR, and the appearance of ventricular arrhythmias.

Despite these abnormalities, children who underwent total surgical repair for ToF are usually well during daily life. However, formal exercise testing has repeatedly shown subnormal values for $\dot{V}O_2$ max and also for ventilatory anaerobic threshold in this patient group.[15,33,34] Moreover, some individuals may reach normal values. Furthermore, after training, patients with this type of pathology can significantly increase maximal exercise capacity.[35] When the adequacy of the oxygen transport during exercise in patients with ToF repair was assessed by calculation of the slope of $\dot{V}O_2$ vs exercise intensity, reduced values have been found in patients after repair of ToF.[21] This was associated with increased values for the physiological dead space ventilation during exercise or the slope of V_E vs $\dot{V}CO_2$.[36] This is mostly attributed to significant residual haemodynamic abnormalities, such as severe pulmonary regurgitation and right ventricular dysfunction.

Postoperative ToF patients may have ventricular ectopy during exercise (exercise-induced arrhythmia). Exercise-induced ventricular arrhythmias are mainly seen in patients with late repair and poor right ventricular function.[37] Patients with important residual haemodynamic abnormalities such as those previously mentioned are at risk for cardiovascular events.[20]

Transposition of the great arteries

In simple transposition of the great arteries (TGA), the aorta arises from the right ventricle, while the pulmonary artery originates from the left ventricle. This results in severe cyanosis, as desaturated systemic venous blood is pumped in the systemic circulation, while the pulmonary venous return is pumped via left atrium and left ventricle in the lungs. Since this blood is already fully oxygenated, no more oxygen will be added to the blood.

The surgical approach to TGA from the late 1960s to early 1980s involved baffling or rerouting the systemic venous return (from the superior and inferior vena cava) to the mitral valve and left ventricle (atrial switch procedures, also referred to as Mustard or Senning procedure). Exercise testing following the atrial switch procedures has shown a variety of abnormalities even in patients who were asymptomatic at rest.[15,29,38] Also a variety of arrhythmias have been documented during exercise testing (junctional rhythm, premature atrial contractions, premature ventricular contractions).

Currently the arterial switch operation is the preferred surgical technique for transposition of the great arteries. Normal or near-normal values for exercise performance and normal ST on ECG have been reported in this patient group.[39,40] With earlier techniques coronary problems early and late after surgery were common. Improved surgical techniques appear to have resolved these problems.

Fontan circulation

In tricuspid atresia, there is a congenital absence of the tricuspid valve. In a Fontan circulation the caval veins are currently connected directly on the pulmonary arteries, bypassing the right heart. This means that there is no effective right heart pump. Although the survival and also exercise performance of these patients improve dramatically, most of these subjects still have a limited exercise tolerance.[41–43] The circulatory output after Fontan is primordially regulated by the pulmonary vasculature, which limits ventricular preload. The ventricule usually 'will pump whatever it gets'. Therefore, in the healthy Fontan patient, it may be the absence of a sub-pulmonary pump that limits normal increases in pulmonary pressures, trans-pulmonary flow requirements, and cardiac output that are required during exercise.[44]

The HR response is usually blunted in patients with Fontan circulation, and due to this, the term chronotropic incompetence is frequently incorrectly used. Using this definition depends on the comparison between patients and normal subjects regarding chronotropic incompetence: as a proportion of maximal exercise capacity, or in absolute values. Fontan patients have the reputation of having chronotropic impairment when HR is expressed relative to maximal normal value, but in absolute values they are even faster than controls. The term 'incompetence' suggests that increasing the HR would improve exercise tolerance; not only is this incorrect, but it is also dangerous. Heart rate response to exercise is indeed impaired in Fontan patients in comparison to healthy controls, but it is appropriate for the level of ventricular preload. A very fast HR with a limited preload would result in a decreased SV with hypotension, syncope, and eventually death. The lower oxygen saturation in the arterial blood sometimes seen in Fontan patients during exercise rather may be due to residual venous or atrial shunting.[45]

Fontan patients have a decline of exercise tolerance with increasing pulmonary vascular resistance and increasing ventricular end diastolic pressure; this downward slope may be slowed by regular exercise and may be very important for prolonging the favourable haemodynamic result in these patients. This is especially so as the pulmonary circulation in current Fontan circuits is characterized by absent pulsatility and limited increase in regular flow and pressure; this may result in increasing pulmonary vascular resistance.

Rhythm disturbances and conduction defects

Congenital complete atrioventricular block

In congenital complete atrioventricular block, the atrial rate increases normally during exercise, but ventricular rate does not accelerate adequately. In some cases, these patients may develop dizziness and syncope. Exercise testing in these patients shows subnormal values for peak $\dot{V}O_2$ or ventilatory anaerobic threshold and even for the increase of $\dot{V}O_2$ versus exercise intensity.[46] This results from the lack of acceleration of HR during exercise, one of the major components in increasing cardiac output and therefore oxygen delivery to the exercising tissues. In some cases with severe bradycardia and syncope, a pacemaker is inserted. These children should avoid competitive sports and physical activities where there is a danger of body collision as the wiring system that connects the pacemaker with the heart is easily damaged.

In the paediatric population, the frequency and significance of arrhythmia differs from adults.[46] As a general rule, the assessment of cardiac arrhythmia during exercise is useful in the management of these patients. If arrhythmia disappears with increasing exercise intensity, the prognosis of this type of arrhythmia is usually benign.

Congenital complete atrioventricular block after surgery

The anatomic structure of the sinus node is vulnerable to damage following cardiac surgery. Damage of the sinus node has been observed after surgical procedures that require extensive

manipulations and sutures in the atria. Specific defects include D-transposition of the great arteries, repaired by atrial baffling procedures.[47] Fortunately, surgically acquired complete atrioventricular block is relatively uncommon, even with extensive surgery in the atria (e.g. D-transposition of the great arteries). In surgically acquired atrioventricular block there is usually no escape rhythm. Safety pacing is recommended.[48]

Habitual physical activity in children with congenital heart disease

Different methods have been applied in assessing the daily level of PA in paediatric patients. Physical activity can be assessed by self-report or objective measures. However, PA is difficult to assess and interpret, because no method is completely accurate.[49] (Interested readers are referred to Chapter 21 where PA assessment is extensively reviewed). However, despite their shortcomings, PA assessments are of value in children and adolescents with congenital heart disease. They can provide complementary information to exercise testing, which is useful in the interpretation of the exercise test results and in the formulation of PA advice to the patient.

It has been reported that children with congenital heart disease have reduced levels of PA when compared to healthy children and have PA levels below what is recommended.[50–52] However, results are conflicting in relation to PA.[53] Low levels of PA may be lifestyle-related in part, because patients with congenital heart disease have a potential to improve their physical fitness by PA similar to that of their healthy peers. Indeed, a relation between PA and exercise capacity also exists in children with congenital heart disease.[54]

Natural evolution of aerobic exercise performance and daily level of physical activity in children with congenital heart disease

It is a normal evolution to see the exercise capacity change during life. However, data regarding the progression of exercise intolerance with age in patients with congenital heart disease are scarce. Nevertheless, it is possible that the first signs of an impaired heart function are demonstrated by an exercise capacity that decreases faster than normal.[55]

To study the natural evolution of aerobic exercise performance during medium-term follow-up in patients with congenital heart disease, exercise performance tests were compared in patients who underwent exercise testing at least twice with a time interval of about three years. In our laboratory, serial exercise testing documented that cardiovascular exercise performance declines progressively in children with medically imposed restriction of intensive PA, such as aortic stenosis and in children with residual hemodynamic lesions. In the other patient groups we investigated, which were unoperated ventricular septal defect, pulmonary stenosis, and surgically closed ventricular septal defect, exercise performance remained stable during medium-term follow-up.[30] Moreover, the results of this study showed that at the initial evaluation all patients were in class I of the New York Heart Association (NYHA). At reassessment, about three years later, all patients remained in NYHA class I, except for two patients with a Fontan circulation, who belonged to class II and III at re-evaluation.[30] The daily level of PA, assessed by a standardized questionnaire, was significantly lower both at the first and second evaluation in patients with aortic stenosis, surgical repair of Tetralogy of Fallot, and Fontan repair, compared to the other patient groups. These subnormal values for the daily PA level were associated with a significant decrease of aerobic exercise function at re-assessment.[30]

We also performed a first longitudinal study in children and adolescents with atrial switch operation and concluded that in the overall group, the exercise performance remained stable during a follow-up of 3.5 ± 2 years, but that in individual patients, a decreasing exercise capacity was correlated with the development of haemodynamic lesions.[56] In a later study only investigating patients with Senning repair for complete transposition of the great arteries, we demonstrated that peak $\dot{V}O_2$ and peak oxygen pulse decreased faster with age than in healthy controls. This decline is most obvious during childhood and adolescence, and in female patients, when compared to young adults. In Figure 29.3, the decline in peak $\dot{V}O_2$ as a percentage of predicted values is plotted for children and adolescents with various congenital heart defects. The fact that the progressive decline is most obvious during childhood and adolescence suggests the impossibility to increase SV to the same extent as healthy peers during growth.[57] Moreover, a faster decline in peak oxygen pulse was related to a decrease in right ventricular contractility in this patient group.[57]

Fernandes *et al.*[58] found similar results in patients with a Fontan circulation in which a decline was particularly present in patients younger than 18 years; in adults this decline of exercise capacity tended to be similar to that of a normal population. Also Giardini *et al.*[59] showed a faster decline in exercise capacity compared to healthy controls in patients with a Fontan circulation, and this decline was stronger in patients with a morphologic right ventricle.

These data show the combined effect of the heart defect, the residual haemodynamic lesions, and hypoactivity on the evaluation of aerobic exercise performance in these groups of patients. In children and adolescents with aortic stenosis, significantly lower values

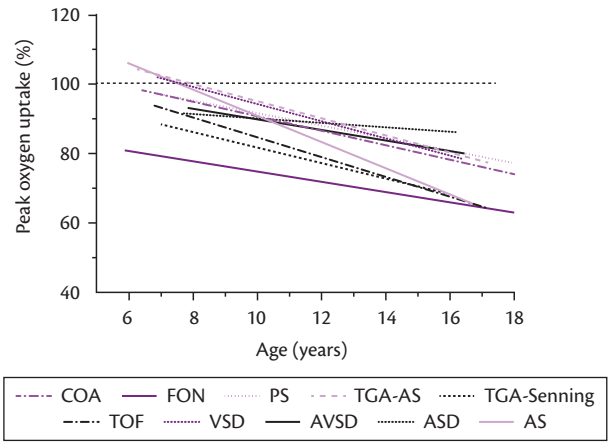

Figure 29.3 Changes in peak oxygen uptake with age.
AS, invasively treated aortic valve stenosis; ASD, surgically treated atrial septal defect; AVSD, surgically treated atrioventricular septal defect; COA, surgically treated aortic coarctation; FON, univentricular heart repaired with Fontan operation; PS, invasively treated pulmonary valve stenosis; TGA-AS, transposition of the great arteries surgically treated with the arterial switch operation; TGA-Senning, transposition of the great arteries surgically treated with the senning operation; TOF, surgically corrected tetralogy of Fallot; VSD, surgically treated ventricular septal defect. Dotted line represents peak $\dot{V}O_2$ of healthy children.

for aerobic exercise performance were found both at first and second assessment. Similar data have been reported by Driscoll *et al.*[60]

The results of these studies show that the suboptimal aerobic exercise performance in children and adolescents with congenital heart disease are to some extent attributable to residual haemodynamic lesions after corrective surgery of the defect, and also to some degree of hypoactivity resulting from overprotection of the parents and the environment. However, in some patients there may also be an increase of the severity of the disease, which may prevent the individual from performing the same amount of physical exercise as their healthy peers. Therefore, except for some cases with severe disease progression and/or medically imposed restriction of intensive dynamic or static physical exercise, children and adolescents with congenital heart disease and their parents should be strongly encouraged to be more active, and to prevent the deleterious effect of physical deconditioning.

Exercise recommendations and rehabilitation of children with congenital heart disease

Nowadays most children with congenital heart disease are encouraged to be fully active and to participate in all recreational sport activities, even after corrective surgery. These recommendations are based on clinical findings that have shown that physical exercise in children with congenital heart disease has beneficial effects on physical, psychological, and social levels of both children and their parents/carers. In the majority of cases these children do not need to participate in a formal rehabilitation programme, but they should be encouraged to participate in recreational physical activities in leisure time and at school. Even after corrective surgery, a formal rehabilitation programme is mostly restricted to the hospitalization period and consists mainly of chest physiotherapy (breathing exercises) and early mobilization. As soon as the children are discharged from the hospital, they are encouraged to resume their normal physical activities at home. Unfortunately, despite this encouragement, many of these children are often (inappropriately) considered to be very fragile, resulting in overprotective behaviour from parents, and greater restriction from sports activities than is necessary. Unfortunately, greater inactivity and consequent deconditioning usually follow. With this in mind, performing regular physical exercise may be challenging for children with congenital heart disease, and encouraging an active lifestyle in paramount.[52,61,62]

A more formal paediatric cardiac rehabilitation programme might partially reverse the effects of physical inactivity and deconditioning, thereby improving exercise function. Controlled and clinical exercise studies in patients with congenital heart disease have shown that maximal exercise capacity can be improved following a period of physical training.[35] However, $\dot{V}O_2$ max did not improve in all studied subjects. The improvement of maximal exercise performance (assessed during cycle ergometry) without an increase in peak $\dot{V}O_2$ represents an improved mechanical efficiency during exercise. This may be beneficial for the patients, since the same level of exercise will be perceived as easier to perform and will induce less dyspnoea.[35]

Next to the benefit of an increased exercise and functional capacity, an exercise programme might also induce favourable benefits on the social level. It allows children with congenital heart disease to initiate exercise in a safe environment. An exercise programme might thus increase both parents' and children's level of confidence with regard to exercise; quite often there is also a positive affect regarding social interaction behaviours and well-being of the children. Indeed, playing or being active together with healthy peers may also help reduce children's feelings of being 'different'.

In the majority of cases of congenital heart disease there are only a few contraindications for physical exercise for both operated and non-operated cardiac defects.[37] Cumulative medical experience has shown that the potential risk of physical exercise in patients with congenital heart disease is very low.[35] In fact, only a few heart defects have been associated with sudden cardiac death during sports participation, which are mainly hypertrophic cardiomyopathy, severe aortic stenosis, congenital anomalies of the coronary arteries, Marfan's syndrome, and myocarditis. Fortunately, these anomalies represent only a small percentage of the total number of congenital heart defects in which sport participation is allowed. Since children, especially those who perform competitive sports, may be exposed to high levels of physiological stress, specific and detailed recommendations have been formulated for children with congenital heart disease.[22,37,63] These recommendations can be used for counselling children and adolescents with congenital heart disease.

Table 29.1 provides an overview of conditions requiring restrictions of heavy physical exercise and competitive sports, including moderate to severe aortic stenosis, left to right shunts with pulmonary hypertension, hypertrophic cardiomyopathy, pulmonary hypertension, and arrhythmia that worsens during exercise.[22,37,63,64] However, the final decision to allow the child with congenital heart disease to participate in physical exercise should always be based on a full cardiological examination. Regardless, as a general rule, cardiopulmonary exercise testing is advised in children with congenital heart defects before participation in sports activities is allowed.

Table 29.1 Exercise restrictions for the most common types of congenital heart disease

No restrictions	Avoid high-intensity static sports	Avoid high-intensity static and dynamic sports
Mild AS	COA	Moderate-severe AS
ASD		FON
VSD		PS 30–50 mmHg gradient
AVSD		TGA-AS
PS < 30mmHg gradient		PAH-CHD
TOF		HCMP
		Exercise induced arrhythmia

AS, aortic valve stenosis; ASD, atrial septal defect; AVSD, atrio-ventricular septal defect; COA, aortic coarctation; FON, univentricular heart repaired with Fontan operation; PS, pulmonary valve stenosis; TGA-AS, transposition of the great arteries surgically treated with the arterial switch operation; TOF, Tetralogy of Fallot; VSD, ventricular septal defect, PAH, pulmonary arterial hypertension associated with congenital heart disease; HCMP, hypertrophic cardiomyopathy.

Conclusions

As a large proportion of patients with congenital heart disease are in the care of paediatric specialists, exercise testing equipment and exercise protocols have to be adapted for children. Functional performance should be assessed by performing exercise testing with measurement of gas exchange. In some groups of patients with congenital heart disease, suboptimal values have been found for aerobic exercise capacity. These values can be ascribed to haemodynamic dysfunction or residual haemodynamic lesions after surgery (e.g. in transposition of the great arteries, Tetralogy of Fallot, Fontan repair for univentricular heart). In other types of pathologies, medically imposed restriction of intensive physical exercise or competitive sports may determine to some extent a subnormal value of exercise performance. Finally, in some other types of congenital heart disease without overt haemodynamic dysfunction (e.g. ventricular septal defect or atrial septal defect, with normal pressures in the pulmonary circulation) a suboptimal value for aerobic exercise capacity is often related to reduced levels of PA in daily life. Exercise interventions to increase PA have been shown to generally be safe and beneficial in increasing exercise capacity in children with congenital heart disease, although further research in this area is necessary. Therefore, except for some cases with medically imposed restriction of intensive physical exercise, most patients are encouraged to be fully active during leisure time and to participate in all types of physical exercise at school.

Summary

- In children with congenital heart disease, maximal exercise testing is frequently performed to measure cardiorespiratory function and to assess abnormalities of cardiac rhythm.
- Both maximal and submaximal exercise parameters need to be taken into account in order to evaluate the child's clinical and physical condition.
- A reduced exercise capacity is common in children with congenital heart disease. This relates not only to the cardiac condition, but often also to a low level of physical activity in daily life.
- Serial exercise testing is particularly useful in children with congenital heart disease as it is possible that the first signs of an impaired heart function are demonstrated by an exercise capacity that decreases faster than normal.
- Exercise training interventions to increase physical activity have been shown to generally be safe and beneficial in increasing exercise capacity in children with congenital heart disease.
- Except for some cases with medically imposed restriction of intensive physical exercise, most patients are encouraged to be fully active during leisure time and to participate in all types of physical exercise at school.
- The final decision to allow the child with congenital heart disease to participate in physical exercise should always be based on a full cardiological examination along with cardiopulmonary exercise testing.

References

1. Gibbons RJ, Balady GJ, Bricker JT, *et al.* ACC/AHA 2002 guideline update for exercise testing: summary article. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee to Update the 1997 Exercise Testing Guidelines). *J Am Coll Cardiol*. 2002; 16; 408: 1531–1540.
2. Rowland TW. Aerobic exercise testing protocols. In: Rowland TW (ed.) *Pediatric laboratory exercise testing. Clinical guidelines.* Champaign, IL: Human Kinetics; 1993: 19–41.
3. Reybrouck T, Vangesselen S, Gewillig M. Impaired chronotropic response to exercise in children with repaired cyanotic congenital heart disease. *Acta Cardiol*. 2009; 646: 723–727.
4. Whipp BJ, Higgenbotham MB, Cobb FC. Estimating exercise stroke volume from asymptotic oxygen pulse in humans. *J Appl Physiol*. 1996; 816: 2674–2679.
5. Jones S, Elliott PM, Sharma S, McKenna WJ, Whipp BJ. Cardiopulmonary responses to exercise in patients with hypertrophic cardiomyopathy. *Heart*. 1998; 801: 60–67.
6. Hsi WL, Wong PL, Lai JS. Submaximal oxygen pulse divided by body weight during incremental exercise test. *Am J Phys Med Rehabil*. 1997; 764: 297–303.
7. Klainman E, Fink G, Lebzelter J, Krelbaumm T, Kramer MR. The relationship between left ventricular function assessed by multigated radionuclide test and cardiopulmonary exercise test in patients with ischemic heart disease. *Chest*. 2002; 1213: 841–845.
8. Bar-Or O, Rowland TW. Cardiovascular heart diseases. In: Robertson LD (ed.) *Pediatric exercise medicine: from physiological principles to health care application*. Leeds: Human Kinetics; 2004. p. 177–213.
9. Wilmore JH, Costill DL. *Children and adolescents in sport and exercise. Physiology of sport and exercise*. Leeds: Human Kinetics; 2004.
10. Adams FH, Linde LM. Physical working capacity: an index of cardiac fitness. *Chest*. 1961; 39: 577–578.
11. Adams FH, Duffie ERJr. Physical working capacity of children with heart disease. *Lancet*. 1961; 81: 493–496.
12. Washington RL, Bricker JT, Alpert BS, *et al.* Guidelines for exercise testing in the pediatric age group. From the Committee on Atherosclerosis and Hypertension in Children, Council on Cardiovascular Disease in the Young, the American Heart Association. *Circulation*. 1994; 904: 2166–2179.
13. Reybrouck T, Weymans M, Stijns H, Knops J, Van der Hauwaert L. Ventilatory anaerobic threshold in healthy children. Age and sex differences. *Eur J Appl Physiol Occup Physiol*. 1985; 543: 278–284.
14. Wasserman K. The anaerobic threshold measurement to evaluate exercise performance. *Am Rev Respir Dis*. 1984; 129(2 Pt 2): S35–S40.
15. Reybrouck T, Mertens L, Kalis N, *et al.* Dynamics of respiratory gas exchange during exercise after correction of congenital heart disease. *J Appl Physiol*. 1996; 802: 458–463.
16. Baba R, Nagashima M, Goto M, *et al.* Oxygen uptake efficiency slope: a new index of cardiorespiratory functional reserve derived from the relation between oxygen uptake and minute ventilation during incremental exercise. *J Am Coll Cardiol*. 1996; 15; 286: 1567–1572.
17. Akkerman M, van Brussel M, Bongers BC, Hulzebos EH, Helders PJ, Takken T. Oxygen uptake efficiency slope in healthy children. *Pediatr Exerc Sci*. 2010; 223: 431–441.
18. Mezzani A, Giordano A, Moussa NB, *et al.* Hemodynamic, not ventilatory, inefficiency is associated with high VE/VCO2 slope in repaired, noncyanotic congenital heart disease. *Int J Cardiol*. 2015; 191: 132–137.
19. Rausch CM, Taylor AL, Ross H, Sillau S, Ivy DD. Ventilatory efficiency slope correlates with functional capacity, outcomes, and disease severity in pediatric patients with pulmonary hypertension. *Int J Cardiol*. 2013; 1696: 445–448.
20. Muller J, Hager A, Diller GP, *et al.* Peak oxygen uptake, ventilatory efficiency and QRS-duration predict event free survival in patients late after surgical repair of tetralogy of Fallot. *Int J Cardiol*. 2015; 196: 158–164.
21. Reybrouck T, Mertens L, Brusselle S, *et al.* Oxygen uptake versus exercise intensity: a new concept in assessing cardiovascular exercise function in patients with congenital heart disease. *Heart*. 2000; 841: 46–52.

22. Hirth A, Reybrouck T, Bjarnason-Wehrens B, Lawrenz W, Hoffmann A. Recommendations for participation in competitive and leisure sports in patients with congenital heart disease: a consensus document. *Eur J Cardiovasc Prev Rehabil*. 2006; 133: 293–299.
23. Reybrouck T, Weymans M, Stijns H, Van der Hauwaert LG. Ventilatory anaerobic threshold for evaluating exercise performance in children with congenital left-to-right intracardiac shunt. *Pediatr Cardiol*. 1986; 71: 19–24.
24. Reybrouck T, Bisschop A, Dumoulin M, Van der Hauwaert LG. Cardiorespiratory exercise capacity after surgical closure of atrial septal defect is influenced by the age at surgery. *Am Heart J*. 1991; 122(4 Pt 1): 1073–1078.
25. Bendien C, Bossina KK, Buurma AE, *et al*. Hemodynamic effects of dynamic exercise in children and adolescents with moderate-to-small ventricular septal defects. *Circulation*. 1984; 706: 929–934.
26. Reybrouck T, Mertens L, Schulze-Neick I, *et al*. Ventilatory inefficiency for carbon dioxide during exercise in patients with pulmonary hypertension. *Clin Physiol*. 1998; 184: 337–344.
27. Meijboom F, Szatmari A, Utens E, *et al*. Long-term follow-up after surgical closure of ventricular septal defect in infancy and childhood. *J Am Coll Cardiol*. 1994; 245: 1358–1364.
28. Binkhorst M, van de Belt T, de Hoog M, van Dijk A, Schokking M, Hopman M. Exercise capacity and participation of children with a ventricular septal defect. *Am J Cardiol*. 2008; 1028: 1079–1084.
29. Pianosi PT, Driscoll DJ. Exercise testing. In: Allen HE, Driscoll D, Shaddy RE, Feltes TF (eds.) *Moss' heart disease in infants, children and adolescents*. Philadelphia: Wolters Kluwer—Lippincott Wiliams & Wilkins; 2008. p. 81–94.
30. Reybrouck T, Rogers R, Weymans M, *et al*. Serial cardiorespiratory exercise testing in patients with congenital heart disease. *Eur J Pediatr*. 1995;154(10): 801–806.
31. Rowland TW. Congenital obstructive and valvular heart disease. In: Goldberg B (ed.) *Sports and exercise for children with chronic health conditions*. Leeds: Human Kinetics; 1995. p. 183–207.
32. Fahey JT. Congenital Heart Disease—Shunt lesions and cyanotic heart disease. In: Goldberg B (ed.) *Sports and exercise in children with chronic health conditions*. Leeds: Human Kinetics; 1995. p. 208–224.
33. Reybrouck T, Weymans M, Stijns H, Van der Hauwaert LG. Exercise testing after correction of tetralogy of Fallot: the fallacy of a reduced heart rate response. *Am Heart J*. 1986; 1125: 998–1003.
34. Luijnenburg SE, De Koning WB, Romeih S, *et al*. Exercise capacity and ventricular function in patients treated for isolated pulmonary valve stenosis or tetralogy of Fallot. *Int J Cardiol*. 2012; 1583: 359–363.
35. Duppen N, Takken T, Hopman MT, *et al*. Systematic review of the effects of physical exercise training programmes in children and young adults with congenital heart disease. *Int J Cardiol*. 2013; 1683: 1779–1787.
36. Reybrouck T, Boshoff D, Vanhees L, Defoor J, Gewillig M. Ventilatory response to exercise in patients after correction of cyanotic congenital heart disease: relation with clinical outcome after surgery. *Heart*. 2004; 902: 215–216.
37. Takken T, Giardini A, Reybrouck T, *et al*. Recommendations for physical activity, recreation sport, and exercise training in paediatric patients with congenital heart disease: a report from the Exercise, Basic and Translational Research Section of the European Association of Cardiovascular Prevention and Rehabilitation, the European Congenital Heart and Lung Exercise Group, and the Association for European Paediatric Cardiology. *Eur J Prev Cardiol*. 2012; 195: 1034–1065.
38. Reybrouck T, Gewillig M, Dumoulin M, Van der Hauwaert LG. Cardiorespiratory exercise performance after Senning operation for transposition of the great arteries. *Brit Heart J*. 1993; 702: 175–179.
39. Reybrouck T, Eyskens B, Mertens L, Defoor J, Daenen W, Gewillig M. Cardiorespiratory exercise function after the arterial switch operation for transposition of the great arteries. *Eur Heart J*. 2001; 2212: 1052–1059.
40. Muller J, Hess J, Horer J, Hager A. Persistent superior exercise performance and quality of life long-term after arterial switch operation compared to that after atrial redirection. *Int J Cardiol*. 2013; 1662: 381–384.
41. Gewillig M. The Fontan circulation. *Heart*. 2005; 916: 839–846.
42. Gewillig MH, Lundstrom UR, Bull C, Wyse RK, Deanfield JE. Exercise responses in patients with congenital heart disease after Fontan repair: patterns and determinants of performance. *J Am Coll Cardiol*. 1990; 156: 1424–1432.
43. Diller GP, Dimopoulos K, Okonko D, *et al*. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. *Circulation*. 2005; 1126: 828–835.
44. La Gerche A, Gewillig M. What limits cardiac performance during exercise in normal subjects and in healthy fontan patients? *Int J Pediatr*. 2010; 10: 1–8.
45. Gewillig MH, Lundstrom UR, Bull C, Wyse RK, Deanfield JE. Exercise responses in patients with congenital heart disease after Fontan repair: patterns and determinants of performance. *J Am Coll Cardiol*. 1990; 156: 1424–1432.
46. Reybrouck T, Vanden Eynde B, Dumoulin M, Van der Hauwaert LG. Cardiorespiratory response to exercise in congenital complete atrioventricular block. *Am J Cardiol*. 1989; 6414: 896–899.
47. Paul MH, Wessel HU. Exercise studies in patients with transposition of the great arteries after atrial repair operations (Mustard/Senning): a review. *Pediatr Cardiol*. 1999; 201: 49–55.
48. Eliasson H, Sonesson SE, Salomonsson S, Skog A, Wahren-Herlenius M, Gadler F. Outcome in young patients with isolated complete atrioventricular block and permanent pacemaker treatment: A nationwide study of 127 patients. *Heart Rhythm*. 2015; 1211: 2278–2284.
49. Warren JM, Ekelund U, Besson H, Mezzani A, Geladas N, Vanhees L. Assessment of physical activity—a review of methodologies with reference to epidemiological research: a report of the exercise physiology section of the European Association of Cardiovascular Prevention and Rehabilitation. *Eur J Cardiovasc Prev Rehabil*. 2010; 172: 127–139.
50. Arvidsson D, Slinde F, Hulthen L, Sunnegardh J. Physical activity, sports participation and aerobic fitness in children who have undergone surgery for congenital heart defects. *Acta Paediatr*. 2009; 989: 1475–1482.
51. De Bleser L, Budts W, Sluysmans T, *et al*. Self-reported physical activities in patients after the Mustard or Senning operation: comparison with healthy control subjects. *Eur J Cardiovasc Nurs*. 2007; 63: 247–251.
52. Massin MM, Hovels-Gurich HH, Gerard P, Seghaye MC. Physical activity patterns of children after neonatal arterial switch operation. *Ann Thorac Surg*. 2006; 812: 665–670.
53. Ewalt LA, Danduran MJ, Strath SJ, Moerchen V, Swartz AM. Objectively assessed physical activity and sedentary behaviour does not differ between children and adolescents with and without a congenital heart defect: a pilot examination. *Cardiol Young*. 2012; 221: 34–41.
54. Weymans M, Reybrouck T. Habitual level of physical activity and cardiorespiratory endurance capacity in children. *Eur J Appl Physiol Occup Physiol*. 1989; 588: 803–807.
55. Fredriksen PM, Veldtman G, Hechter S, *et al*. Aerobic capacity in adults with various congenital heart diseases. *Am J Cardiol*. 2001; 873: 310–314.
56. Reybrouck T, Mertens L, Brown S, Eyskens B, Daenen W, Gewillig M. Long-term assessment and serial evaluation of cardiorespiratory exercise performance and cardiac function in patients with atrial switch operation for complete transposition. *Cardiol Young*. 2001; 111: 17–24.
57. Buys R, Budts W, Reybrouck T, Gewillig M, Vanhees L. Serial exercise testing in children, adolescents and young adults with Senning repair for transposition of the great arteries. *BMC Cardiovasc Disord*. 2012; 12: 88.
58. Fernandes SM, McElhinney DB, Khairy P, Graham DA, Landzberg MJ, Rhodes J. Serial cardiopulmonary exercise testing in patients with previous Fontan surgery. *Pediatr Cardiol*. 2010; 312: 175–180.

59. Giardini A, Hager A, Pace NC, Picchio FM. Natural history of exercise capacity after the Fontan operation: a longitudinal study. *Ann Thorac Surg*. 2008; 853: 818–821.
60. Driscoll DJ, Wolfe RR, Gersony WM, *et al*. Cardiorespiratory responses to exercise of patients with aortic stenosis, pulmonary stenosis, and ventricular septal defect. *Circulation*. 1993; 87(2 Suppl): I102–I113.
61. Longmuir PE, Brothers JA, de Ferranti SD, *et al*. Promotion of physical activity for children and adults with congenital heart disease: a scientific statement from the American Heart Association. *Circulation*. 2013; 12721: 2147–2159.
62. Muller J, Hess J, Hager A. Daily physical activity in adults with congenital heart disease is positively correlated with exercise capacity but not with quality of life. *Clin Res Cardiol*. 2012; 1011: 55–61.
63. Graham TP, Jr., Bricker JT, James FW, Strong WB. 26th Bethesda conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. Task Force 1: congenital heart disease. *Med Sci Sports Exerc*. 1994; 26(Suppl): S246–S253.
64. Pelliccia A, Fagard R, Bjornstad HH, *et al*. Recommendations for competitive sports participation in athletes with cardiovascular disease: a consensus document from the Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. *Eur Heart J*. 2005; 2614: 1422–1445.