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EUROPEAN SOCIETY OF CARDIOLOGY

Position Paper

Recommendations for participation in competitive and leisure sports in patients with congenital heart disease: a consensus document

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Received 20 March 2006 **Accepted** 27 March 2006

Background **Physical activity is important for patients with congenital heart disease. The aim of this paper is to provide a consensus document for participation in competitive or leisure sport activity in children and adults with congenital heart disease.**

Methods **The recommendations are based on expert consensus meetings, personal experience of the contributing authors and an updated review of the literature regarding exercise performance and risk stratification in patients with congenital heart disease.**

Results **Physical performance and exercise tolerance is close to normal in patients with simple lesions with successful repair or no need for therapy. Most patients with complex lesions have some degree of residual disease, making them less suitable for participation in competitive sport.**

Conclusion **Regular exercise at recommended levels can be performed and should be encouraged in all patients with congenital heart disease. Many can attend sports with no restrictions. Special concern should be given to those patients with a significant ventricular dysfunction or recent history or risk of arrhythmia.** ***Eur J Cardiovasc Prev Rehabil*** **13:293–299**

European Journal of Cardiovascular Prevention and Rehabilitation 2006, **13**:293–299

Keywords: congenital heart disease, recommendations, exercise, sport

Conflict of interest: none.

A short version of the recommendations regarding competitive sports in patients with congenital heart disease was included/published in the paper by Pelliccia *et al.* 'Recommendations for competitive sports participation in athletes with cardiovascular disease', *Eur Heart J 2005;* **26**:1422–1445.

Introduction

Numerous scientific reports have documented the positive effects of physical activity on public health in general [1–3] and on cardiovascular health in particular [4]. Futhermore, patients with congenital heart disease (CHD) seem to benefit from regular physical activity [5,6]. Regarding the cardiovascular long-term health effects of high intensity, competitive sport very little is known both in healthy people and in those with CHD [7].

Overprotection is common in children with CHD [8]. The resulting sedentary lifestyle leads to diminished physical work capacity and places them at risk for early development of cardiovascular disease and other illnesses associated with physical inactivity [9]. Children have a natural need for motor activities and this should not be interrupted or discouraged. Perception and motor activities in children with CHD are catalysts not only for the

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1741-8267

child's physical development but also for the emotional, psychosocial and cognitive skill [10].

As regards adolescents, we know that physical activity in youth is a major predictor of maintained fitness throughout life [11]. International recommendations suggest at least 60 min of moderate to vigorous physical activity daily [12]. On the other hand not all adolescents with CHD are eligible for competitive sport. If the adolescent needs to be restricted in his or her physical activity, information about this should be given at an early adolescent stage (10–12 years), allowing both the child and the parents to adapt to the new rules before things get too serious.

The purpose of this review is to give the reader a comprehensive, though practical, tool to guide patients with all relevant CHD, including both children and adults, in leisure and competitive sport participation. The recommendations represent the consensus document of an international expert panel appointed by the European Society of Cardiology. The recommendations are based on published scientific evidence, if available, and on the personal experience of the contributing authors.

Evaluation/risk stratification

If the patient is seen on a regular basis and is well known to the physician, advice regarding leisure sport activity can be given during routine visits. In those attending competitive sports, the level of preparticipation screening should be extensive and up to date (Table 1).

Table 1 Eligibility for non-restricted participation in competitive sports in congenital heart disease patients

Eligible	Not eligible
I Surgical procedure	Uncorrected or palliative corrected
Fully corrected (anatomically)	Significant lesions not operated
	Univentricular hearts
	Mustard, Senning or Rastelli corrected TGA
	Arterio-pulmonal shunts
II Medical history	
Satisfactory	Abnormal
NYHA class I	Symptoms of severe palpitations/syncope
	Exercise-induced symptoms (dyspnoea, angina, palpitations, syncope)
	NYHA class II or higher
III Physical examination	
Satisfactory	Abnormal
	Hypertension
	Hepatomegaly, raised venous pressure
IV ECG/Holter	
Satisfactory	Abnormal
	Ischemia (coronary anomaly, TGA-switch)
	QRS-duration (Fallot)
	Significant hypertrophy
	Significant arrhythmia
V Morphology/haemodynamic	
Satisfactory	Abnormal
	Significant rest-lesion
	Mean transvalvular gradient of aorta $\geq$ 20 mmHg
	Peak transvalvular gradient of the pulmonary artery of $>$50 mmHg
	Significant hypertrophy
	Significant myocardial dysfunction
	Pulmonary hypertension
VI Maximal ergospirometry	
Satisfactory	Abnormal
Values within normal range	Chest pain or syncope
	Significant arrhythmia
	Ischemia on ECG

TGA, transposition of the great arteries; ECG, electrocardiogram.

The surgical reports and the latest diagnostic testing are the most important part of the medical record.

We recommend a questionnaire to describe the symptomatic status according to the criteria of the New York Heart Association (NYHA).

Physical examination should include the resting blood pressure (BP).

The standard 12-lead electrocardiogram (ECG) should be searched for signs of hypertrophy, changes in repolarization and for arrhythmias.

Echocardiography should assess known morphological and haemodynamic problems in that particular defect. If this is not feasible the echo should be repeated or substituted by more suitable investigations.

Magnetic resonance imaging (MRI) should be performed on a regular basis in patients with coarctation of the aorta. MRI can also be necessary in patients with a poor echocardiographic window or to supplement right ventricular studies. Normally it is not requested to decide upon participation in leisure sport activity.

Heart catheterization and angiocardiogram is not routinely used in preparticipating evaluation of athletes with CHD.

If arrhythmia is suspected or common in that specific lesion 24-h Holter monitoring should have been performed within the last 6 months.

Maximal ergospirometry best evaluates the work capacity and the cardiopulmonary reserves. We prefer treadmill, especially in children, but also bicycle ergospirometry provides the necessary data and may have advantages if frequent BP measurements are important. The exercise protocol used should be standardized, aiming at exercise duration of between 6 and 10 min [13]. It should give information about maximal oxygen uptake ($M\dot{V}o_2$), expressed in millilitres per kilogram per minute, maximal heart rate and recovery heart rate at 2 min, respiratory exchange ratio at peak exercise and also describe $\dot{V}o_2$ at the ventilatory anaerobic threshold (VAT) including the method of VAT definition [14]. The oxygen uptake

efficiency slope can be used in patients not capable or willing to attain maximal exertion [15,16]. Measurement of exercise BP and pulse oxymetry are difficult. If indicated (see specific lesions) we recommend to focus on the start and end of the exercise test.

Follow-up

All patients with CHD should be followed on a regular basis according to guidelines given elsewhere [17]. There is no need for specific follow-up in patients only participating in leisure sport activities. Adolescents and adults participating in competitive sport need a structured reassessment every year [18].

Classification of sports

Sports are usually classified as isotonic/dynamic or isometric/static and to the level of intensity. Our group previously reported an overview of the commonest sports in Europe in respect to the classification above [18]. In general static exercise causes mainly pressure overload and can be difficult to control and is therefore less suitable than dynamic exercise in patients with CHD.

Types of congenital heart disease

Simple shunt lesions

Atrial septal defect and patent foramen ovale

An atrial septal defect (ASD) represents a direct communication between the left and the right atrium, allowing shunting of blood between the two chambers. A significant ASD is defined with a Qp/Qs ratio of over 1.5, which also is the indication for elective surgical or percutaneous closure. Shunting over a patent foramen ovale (PFO), with its flap valve, is only possible when the pressure in the right atria exceeds the left atrial pressure.

In non-significant ASD or PFO and if surgical or transcatheter closure of a significant ASD is performed early in life, exercise performance within the normal range is expected [19]. Successfully corrected significant ASD with no residual pulmonary hypertension also has good exercise tolerance with exercise capacity close to normal [20,21]. For exercise recommendations see Table 2.

Ventricular septal defect

A ventricular septal defect (VSD) represents a direct communication anywhere along the interventricular septum. A significant VSD is defined with a Qp/Qs ratio of over 1.5, which also is the indication for elective surgical or percutaneous closure.

Patients with a significant VSD become symptomatic early in life and will either undergo repair or develop Eisenmenger syndrome (see later). The natural history of non-significant VSD shows a low normal or just below normal exercise performance [22]. Exercise studies in successfully repaired VSD show a high incidence of abnormal exercise haemodynamics, but the overall exercise capacity and tolerance should be expected to be close to normal [23]. Again, for exercise recommendations see Table 2.

Table 2 Recommendations for sport participation in congenital heart diseases

Lesion	Recommendation
ASD (closed or non-significant or PFO)	No restrictions Scuba diving should be avoided in those with a remaining shunt, due to the risk of paradoxical embolism
VSD (closed or non-significant)	No restrictions
PDA (closed or non-significant)	No restrictions
AVSD (successfully repaired)	No restrictions
Moderate MVR	Low to moderate dynamic and static sports
PAPVC/TAPVC (successfully repaired)	No restrictions
Pulmonary stenosis (mild)	No restrictions
Moderate	Low to moderate dynamic and static sports
Aortic stenosis (mild)	Low to moderate dynamic and static sports
Moderate	Low dynamic and static sports No competitive sport if left ventricular dysfunction or symptoms
CoA (successfully repaired)	No restrictions[a]
TOF (successfully repaired)	Low to moderate dynamic and static sports[a]
Residual disease	Low dynamic and static sports[a]
TGA	
asoTGA (successfully repaired)	No restrictions
iarTGA, ccTGA	Low to moderate dynamic and low static sports[b]
Ebstein anomaly	Low to moderate dynamic and low static sports[b]
Univentricular hearts/Fontan circulation	Low to moderate dynamic and low static sports[b]
Eisenmenger's syndrome	Low dynamic sports[b]
Congenital coronary artery anomalies	No restrictions
Successfully repaired	

For definitions, risk stratification and follow-up see text. ASD, atrial septal defect; PFO, patent foramen ovale; VSD, ventricular septal defect; PDA, persistent ductus arteriosus; AVSD, atrioventricular septal defect; MVR, mitral valve regurgitation; PAPVC/TAPVC, partial or total anomalous pulmonary venous connection; CoA, coarctation of the aorta; TOF, tetralogy of Fallot; TGA, transposition of the great arteries; aso, arterial switch operation; iar, intra-atrial repair; cc, congenitally corrected. [a]Those with conduit, interposed graft or on anticoagulant drugs should avoid sports with the risk of bodily collision. [b]No competitive sport.

Persistent ductus arteriosus

Persistent ductus arteriosus (PDA) is a remnant of fetal life, representing a communication between the aorta and the pulmonary artery.

Patients with closed PDA without residual pulmonary hypertension are expected to have an excellent physical tolerance with normal exercise capacity [24]. These patients need no regular follow-up. See Table 2 for recommendations.

Atrioventricular septal defect

In atrioventricular septal defect (AVSD) there is a common atrioventricular junction, guarded by a common

atrioventricular valve with shunting at both the atrial and ventricular level.

Exercise performance and tolerance in successfully repaired AVSD is normally good [25,26]. Echocardiogram and exercise ECG within the preceding 12 months is recommended to look for significant mitral valve regurgitation, left ventricular function and arrhythmias [27]. Exercise recommendations are provided in Table 2.

Total or partial anomalous pulmonary venous connection

Total anomalous pulmonary venous connection is present when all, and partial anomalous pulmonary venous connection when one or more but not all pulmonary veins are abnormally drained, either to the right atrium or at some venous level above or below the diaphragm.

If successfully repaired, long-term prognosis is essentially good. Available studies are few, but exercise tolerance seems to be good and exercise capacity is close to normal [28]. The chronotropic response is slightly impaired [29,30]. Table 2 provides exercise recommendations.

Simple obstructive lesions

Pulmonary stenosis

A typical pulmonary stenosis is non-dysplastic and caused by fusion of the valve leaflets. In adults calcification may be present. Mild pulmonary stenosis is defined as a peak transvalvular gradient of less then 50 mmHg, moderate stenosis as 50 mmHg or greater but less than 80 mmHg and severe as 80 mmHg or greater or a right ventricular pressure greater than two-thirds of the systemic systolic pressure.

Exercise capacity and tolerance is quite extensively investigated in children and adults with pulmonary stenosis, both before and after intervention. In mild pulmonary stenosis exercise performance should be expected to be close to normal. Both children and adults with moderate and severe pulmonary stenosis have moderate impaired physical tolerance, due to low cardiac output [31]. Especially children improve their exercise tolerance after intervention, emphasizing the disfavourable course of long-standing, significant pulmonary stenosis [32,33]. For exercise recommendations see Table 2.

Aortic stenosis

Congenital aortic stenosis is a result of abnormal development of the aortic valve commissures and a bicuspid valve is the most common morphologic finding. Aortic stenoses are classified as mild (mean aortic valve gradient ≤ 20 mmHg), moderate (mean aortic valve gradient between 21 and 49 mmHg) or severe (mean aortic valve gradient ≥ 50 mmHg).

Exercise tolerance and capacity is moderately impaired in those with severe aortic stenosis and will improve after surgery [34]. Patients with mild or moderate aortic stenosis have either normal or slightly subnormal exercise capacity, the latter probably due to medically imposed restrictions. Sudden cardiac death during exercise, probably due to fatal arrhythmias, is associated with aortic stenosis [35]. This is very unlikely to happen in asymptomatic patients, however, with mild or moderate aortic stenosis. Exercise testing to look for limiting symptoms seems valuable in predicting the need for surgical intervention in asymptomatic patients and should be performed regularly because of the progressive nature of this lesion [36,37]. Exercise recommendations are listed in Table 2.

Coarctation of the aorta

Coarctation of the aorta is either a localized stenosis or a longer hypoplastic segment of the aorta most commonly located at the junction of the distal aortic arch and the descending aorta.

Patients with successfully repaired coarctation of the aorta have no residual obstruction (gradient between the upper and lower limbs < 21 mmHg) or hypertension and exercise tolerance and capacity are normal [38]. Truly 'successful repair', however, is rather seldom. Most follow-up studies on repaired coarctation of the aorta find residual hypertension at rest, on ambulatory BP monitoring or an abnormal peak exercise BP response [39,40]. In adults a peak BP response above 230 mmHg should be regarded as abnormal, as in children it is necessary to take the child's age and height in account using diagrams given elsewhere [41].

For exercise recommendation see Table 2. If the criteria for hypertension at rest or by ambulatory BP monitoring are fulfilled, the patient should be guided accordingly [18]. The clinical impact of an abnormal exercise BP response remains unclear. We recommend these patients to avoid high static and high dynamic sports.

Complex lesions

Tetralogy of Fallot

The anatomic features of tetralogy of Fallot consist of subpulmonary infundibular stenosis, VSD, overriding of the aorta and right ventricle hypertrophy. The treatment of choice is total correction in early childhood. Unoperated or shunt palliated patients are rare since late repair can be recommended in most cases [17].

Exercise studies in tetralogy of Fallot conclude with mild to moderate subnormal exercise capacity [42,43]. Studies also show that an active lifestyle and training result in better performance [44,45]. Patients with successful repair are asymptomatic, have no more than mild pulmonary

regurgitation, near normal biventricular function and M$\dot{V}o_2$ above 80% of predicted [46]. Exercise-induced ventricular arrhythmias are mainly seen in patients with late repair and poor right ventricular performance [47]. Supraventricular arrhythmias have also been reported [48]. Fatal cases are very rare, however, in asymptomatic patients. Risk stratification should describe the degree of pulmonary regurgitation, residual right ventricular outflow tract obstruction, right ventricular function, history of arrhythmias and age at repair [49,50]. Exercise recommendations are listed in Table 2.

Transposition of the great arteries

In transposition the great arteries (TGA) the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. Until the mid-1980s intra-atrial repair of TGA (iarTGA) according to Senning or Mustard was the treatment of choice. After this operation the right ventricle remains the systemic ventricle. Today the arterial switch operation of TGA (asoTGA) is preferred. The advantage with this operation is that the left ventricle becomes the systemic ventricle. If TGA is combined with a large subaortic VSD, the Rastelli operation is performed, also leaving the left ventricle as the systemic ventricle.

Patients successfully corrected with asoTGA have normal exercise capacity and normal exercise-ECG on short-term follow-up [51]. Serial exercise testing is recommended in those few patients with intramural coronary arteries, who are at increased risk of ischemic heart disease [52]. Patients with iarTGA have moderately impaired physical performance, with M$\dot{V}o_2$ ranging from 25 to 35 ml/kg per min in most studies. The major limiting factors are right ventricular dysfunction, tricuspid regurgitation, impaired chronotropic response and atrial arrhythmia [53,54].

Patients with iarTGA are not eligible for competitive sport, but benefit from regular low to moderate intense physical activity. See also Table 2 for further exercise recommendations.

Congenitally corrected transposition of the great arteries

In congenitally corrected TGA (ccTGA) the atria are in normal position, but the left atrium is connected to the morphological right ventricle and vice versa. The ventricles are connected to the 'wrong' great arteries, leaving the right ventricle as the systemic ventricle. In up to 80% of cases ccTGA is combined with other lesions. VSD, pulmonary abnormalities and tricuspid valve abnormalities are the most common.

Literature on exercise performance in ccTGA is scarce. Few adult patients are asymptomatic and most patients have severe impairment of their exercise capacity due associated lesions, right ventricle dysfunction, tricuspid valve regurgitation and arrhythmia [55–57]. Table 2 lists exercise recommendations.

Ebstein anomaly

Ebstein anomaly is a malformation of the tricuspid valve with apical displacement of the valve resulting in a small right ventricle. It is often combined with right ventricular outflow tract obstruction.

Most patients have severe impairment of exercise capacity with M$\dot{V}o_2$ below 25 ml/kg per min [58,59]. Surgery may improve physical performance [60]. Evaluation should include heart size on chest radiograph, degree of right ventricular atrialization and tricuspid regurgitation and documentation of arrhythmias, all determinants for exercise tolerance. Exercise recommendations are shown in Table 2.

Univentricular hearts/Fontan circulation

Single ventricle function is most commonly a result of double inlet ventricles, atrioventricular atresia or hypoplastic left or right heart syndrome. With only one pumping chamber the systemic venous return is passive, bypassing the heart.

Patients with Fontan circulation all have moderate to severe impaired exercise capacity [61–63]. M$\dot{V}o_2$ is in the range of 15–30 ml/kg per min in most studies. This is due to reduced cardiac output response to exercise, abnormal heart rate response both during exercise and in the recovery phase, lower oxygen saturation due to shunting at different levels and abnormal ventilatory response [64–66]. Although arrhythmias are common in these patients, exercise has not been shown to trigger fatal episodes [67]. As regards exercise recommendation, these patients are not eligible for competitive sport. They benefit from regular light and moderate strenuous sport. See also Table 2.

Eisenmenger's syndrome

Patients with Eisenmenger's syndrome have pulmonary hypertension in combination with a right-to-left or bidirectional shunt. All patients have severe impairment of exercise tolerance [68]. Some studies have shown improved exercise performance after pharmacological intervention [69,70]. Any kind of strenuous exercise is prohibited (Table 2).

Congenital coronary artery anomalies

Congenital coronary artery anomalies (CCAAs) describe a coronary artery arising from an ectopic position off the aorta. Sudden cardiac death may be the first symptom in CCAA [71]. Chest pain, breathlessness, syncope or dizziness during exercise are clinical signs of CCAA

[72]. Exercise ECG and echocardiography are useful tools to diagnose CCAA. Multi-slice computed tomography and magnetic resonance coronary angiogram also seem reliable non-invasive tools to diagnose CCAA. Patients with successfully repaired CCAA are expected to have normal exercise tolerance [73]. For exercise recommendations see Table 2.

Conclusion

Prepubertal children with CHD need no restrictions in their physical activity. Regular exercise at a recommended level can be performed and should be encouraged at all ages in all patients with CHD. Not all patients, however, are eligible for competitive sport. We acknowledge the difficulties in formulating arbitrary exercise recommendations, particularly for certain CHDs in which scientific evidence is scarce. Therefore, caution is needed in applying the present document and efforts should be made to tailor precise advice to each individual.

Acknowledgements

We gratefully acknowledge the advice and comments of Benedicte Eyskens, MD, Department of Pediatric Cardiology, University Hospital Gasthuisberg, 3000 Leuven, Belgium.

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