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FIMS Position Statement

Congenital heart disease and sports

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by

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INTRODUCTION

Nowadays, congenital heart diseases are usually diagnosed at a young age and, whenever possible, corrected surgically at that stage. This implies that a growing number of patients with sometimes very complex (corrected) anomalies reach adulthood and might present to their cardiologist with questions regarding their ability to perform sports or heavy exercise (Fredriksen 2001, Kitchiner 1996, Swan 2000). In this context, it has to be pointed out that their physical capacity may not be adequate to meet the requirements of such an exercise (Fratellone 1994).

Large scale studies in young patients with congenital heart diseases are scarce, due to the low incidence of these conditions. For this reason, recommendations for exercise in those patients are mainly derived from pathophysiological considerations or experience with analogous, acquired heart diseases (Graham 1994, Kaminer 1995, Slansky1993). It remains however questionable whether arrhythmias have the same significance in congenital cardiac malformations as in acquired anomalies or if compensation mechanisms are of similar value in both conditions (Pelech 1986, Garson 1985).

For this reason, the continuous, individual monitoring of the malformation in every patient seems to be of crucial importance: This includes regular checks for arrhythmias, syncopal events, pulmonary hypertension, cardiac dysfunction and compensation mechanisms. In children, recommendations on exercise capacity should consider the self-discipline of the young patient together with the social environment (school, parents). Furthermore, it is important to monitor children with congenital heart diseases at least on a yearly basis, as rapid changes in their cardiac haemodynamics might occur due to growth (Fritsch 1994, Immer 1994).

Many cardiac malformations in children are not common and sometimes very complex. Therefore, this Review focuses on the more frequent (>4 % of all heart diseases) congenital cardiac abnormalities (Schuhmacher, 2001).

SEPTAL AND VASCULAR DEFECTS

Untreated Atrial Septal Defect (ASD)

The most commonly diagnosed type of this malformation is the defect of the ostium secundum (ASD II) with a connection between the two atriae in the middle or upper part of the septum. In 25% of the patients, this disorder is associated with anomalous pulmonary venous connection. Most patients, especially children, seldom display clinical symptoms and usually, operative repair is performed in early childhood, before strenuous physical tasks are performed. In patients that have not been operated, the volume of the left-to-right shunt should be evaluated through echocardiography. A unilateral enlargement of the right atrium is usually indicative of a shunt superior to 40%.

Smaller ASD with shunts below 30% or a pulmonary-systemic flow ratio of < 1,5 do not represent major right ventricular volume loads. Larger defects with higher shunt volumes might, however, lead to a significant right ventricular overload with subsequent pulmonary hypertension. These cases should be operated (Singer 2001).
The significance of a persistent foramen ovale for the prevalence of paradoxical emboli, especially during Valsalva-manoeuvre, as often observed during static exercise such as weightlifting, has yet to be determined. There are no reports on higher incidences of such events in athletes. Nevertheless, activities under altered environmental pressure conditions, such as diving or exercise at high-altitude, might be at a slightly higher risk for this complication.

***RECOMMENDATIONS**:*

- *In patients with small ASD without significant shunt volumes no restrictions for physical activities apply.*

- *In patients with signs of beginning pulmonary hypertension and/or significant right-to-left shunts, light physical activity can be tolerated. If other, impairing conditions of the cardiovascular system such as arrhythmias are associated with the ASD, only light dynamic, health-orientated exercise should be recommended after careful individual evaluation.*

Corrected Atrial Septal Defect (ASD)

ASD with significant shunt volumes are usually closed surgically or through catheterisation techniques shortly after diagnosis. After correction of such defects, no further sequel has to be expected. However, a higher incidence of supraventricular arrhythmias in operated ASD patients has been reported. The incidence of such episodes was clearly related to the age of the patients at the date of the corrective surgery with the highest incidence of arrhythmias in patients that had been operated in preadolescence or later (Reybrouk 1991, 1995).

Before patients with history of corrected ASD start exercising on a regular basis, a basic cardiac examination should therefore be performed including (Stress-) ECG, Doppler-Echocardiography and a chest X-Ray, especially, if a significant shunt or pulmonary hypertension was present prior to the surgical intervention.

***RECOMMENDATIONS**:*

- *For patients with successfully operated ASD and no signs of pulmonary hypertension, significant arrhythmias or cardiac dysfunctions, no restrictions for physical activities apply (6 months after surgery). In patients with apparent pulmonary hypertension, persistent right-to-left-shunt and/or signs of myocardial dysfunction, light physical activity can be tolerated. If other, impairing conditions of the cardiovascular system such as arrhythmias are associated, only light dynamic, health-orientated exercise should be tolerated after careful individual evaluation.*

Untreated Ventricular Septal Defect (VSD)

Defects of the ventricular septum are found in various locations and are sometimes associated with complex anomalies. Defects should be classified according to the magnitude of their shunt volume: Small VSD (left-to-right shunt < 30%), moderate VSD (left-to-right shunt 30- 50%) and large VSD (left-to-right shunt > 50%) (Gabriel 2002, Schumacher, 2001). The clinical symptoms are mainly depending on the amount of shunted blood which is influenced by the pressure and resistance of the pulmonary vasculature. The severity of an apparent VSD can often be diagnosed upon clinical examination, however, Doppler-Echocardiography is recommended to objectify the clinical findings. In cases of doubt or for further investigation of other malformations associated with the VSD, invasive diagnostic procedures might be necessary.

RECOMMENDATIONS

- *In patients with small VSD without significant shunt volumes and without signs of right ventricular enlargement, no restrictions for physical activities apply.*
- *Patients with moderate VSD (shunt 30-50%) should only perform light physical activity, depending on the cardiac adaptation to the shunt.*
- *Large defects, usually associated with a marked increase in pulmonary vascular pressures should avoid any physical exertion exceeding the every-day routine. Furthermore, this applies to patients with medium size defects associated to other anomalies: In these cases an individual evaluation is recommended.*

Corrected Ventricular Septal Defect (VSD)

After surgical or interventional closure of a VSD, a complete cardiac examination with

ECG, stress-ECG, Holter Report and Doppler-Echocardiography should be performed to exclude the presence of a persisting defect, pulmonary hypertension or significant cardiac arrhythmias. Invasive diagnostic procedures might be necessary for this issue.

RECOMMENDATIONS

- *For patients with successful closure of a VSD and no signs of pulmonary hypertension, significant arrhythmias or cardiac dysfunctions, no restrictions for physical activities apply (6 months after surgery). Control examinations every 6 months are recommended.*
- *In patients with persisting, medium-size VSD or persisting pulmonary hypertension, only light sporting activity should be allowed.*
- *Patients with large persisting defects, severe pulmonary hypertension or complex arrhythmias should avoid any physical exertion exceeding the every-day routine.*
- *Light dynamic, health-orientated exercise might be tolerated after careful individual evaluation.*

Untreated Patent Ductus Arteriosus

The patent ducus arteriosus (PDA) can be classified as small, moderate and large, depending on its size (Schuhmacher, 2001). The small PDA is characterized by the absence of clinical symptoms and normal cardiac dimensions. Moderate and large PDA show increased cardiac dimensions and high blood pressure amplitudes, corresponding to the left-to-right shunt. Pulmonary hypertension is frequently observed. However, a PDA at an adult age is seldom observed, as most defects are corrected surgically shortly after diagnosis.

RECOMMENDATIONS

- *For patients with small PDA no restrictions for physical activities apply.*
- *Patients with moderate or large PDA and signs of left ventricular enlargement should only perform light exercise. Patients with large PDA and severe pulmonary hypertension should refrain from exercise and consider operative treatment of the malformation.*
- *In patients with PDA and other, associated abnormalities, the exercise capacity is limited by the main malformation. Light dynamic, health-orientated exercise might be tolerated after careful individual evaluation.*

Corrected Patent Ductus Arteriosus

Successful surgical correction of this malformation is achieved if after the surgical intervention none of the clinical symptoms of PDA persist and normal cardiac dimensions, no residual shunt and no signs of pulmonary hypertension or complex arrhythmias are found.

RECOMMENDATIONS

- *For patients with successful closure of an isolated PDA and no signs of pulmonary hypertension, significant arrhythmias or cardiac dysfunctions, no restrictions for physical activities apply (3 months after surgery).*
- *In patients with persisting symptoms or persisting pulmonary hypertension, the severity of these conditions determine the physical exercise capacity. In general, light, health oriented sporting activity should be allowed.*

DEXTROCARDIAL OBSTRUCTIONS

Untreated Pulmonic Stenosis

Isolated pulmonic stenosis is the most prevalent type of dextrocardial obstruction. Pulmonic stenosis (PS) can be of various severity and is usually evaluated using Doppler-Echocardiography. Method-dependant differences to the invasive measurements in determination of the pressure gradient across the defective valve with this method should be considered. Pressure gradients between the right ventricle and the pulmonary artery below 25 mmHg are without clinical significance. Gradients between 25-49 mmHg characterise light stenosis. Transvalvular gradients between 50 and 79 mmHg indicate moderate, pressure gradients superior to 80 mmHg severe pulmonic stenosis (Nugent 1997,

Schumacher 2001). The consequence of persistent PS is an increase in right ventricular pressure. Other criteria for the evaluation of exercise capacity in PS are dextrocardial dimensions, exercise associated dyspnea, faints, arrhythmias and other ECG-abnormalities. PS can be associated with other cardiac malformations, especially ASD.

***RECOMMENDATIONS*:**

- *For PS patients with pressure gradients below 49 mmHg and no other pathological findings, no restrictions for physical activities apply. Control examinations should be performed on a yearly base.*
- *Patients with PS and pressure gradients above 50 mmHg should only perform light exercise and consider interventional treatment of their condition (Valvuloplasty, Valvulotomy).*
- *In patients with severe PS and pressure gradients above 80 mmHg or moderate PS with additional pathologies, only light dynamic, health-orientated exercise should be recommended after careful individual evaluation.*

Pulmonic Stenosis (Corrected)

After interventional correction of PS through Valvulotomy or Valvuloplasty, the valvular situation should be carefully re-evaluated:

RECOMMENDATIONS:

- *For patients with residual pressure gradients below 49 mmHg and no other pathological findings, no restrictions for physical activities apply. However, patients should refrain from exercise 1 month after valvuloplasty and 3 months after vallotomy. Control examinations must be performed on a regular basis.*
- *In patients with persistent systolic pressure gradients over 50 mmHg, the recommendations for untreated PS apply. Especially right ventricular enlargement and significant arrhythmias might further impair exercise capacity. Light dynamic, health-orientated exercise might be tolerated after careful individual evaluation.*

Sinistrocardial Obstructions

Aortic Stenosis

Congenital stenosis of the aortic orifice can be caused by agenesia, malformations, incongruence or commissural fusion of the valvular cusps or the presence of a bicuspid valve. Furthermore, malformations in the valvular ring might lead to severe aortic stenosis. In 20-30% of all cases, dysplastic or bicuspid aortic valves present a significant regurgitation in addition to the stenosis. The valvular aortic stenosis is frequently associated with other cardiac malformations, mainly aortic coarctation or patent ductus arteriosus.

It is important to monitor children with aortic stenosis on a regular basis, as rapid changes in the severity of their condition can occur during growth. A feature of the young patient is the higher incidence of sudden cardiac death, especially in severe cases, where exercise might trigger potential fatal outcomes.

For the classification of aortic stenosis in view of exercise capacity, a mean systolic gradient across the valve of < 25 mmHg ($A > 1,0\ cm^2$) is considered to reflect light, gradients between 25 and 50 mmHg moderate ($A = 0,5\text{-}1,0\ cm^2$) and pressure gradients above 50 mmHg ($A < 0,5\ cm^2$) severe obstruction of the aortic valve (Carabello 2001, Graham 1994). It has to be pointed out that the classification of aortic stenosis in view of valve replacement therapy follows a slightly different approach (Schumacher 2001). The suggested estimation of physical exercise capacity might be used for both, sub- and supravalvular aortic stenosis.

After interventional therapy through valvotomy or catheterisation techniques, a complete re-evaluation of the cardiac status is necessary: Very often after these treatments, a residual stenosis or, in contrast, a worsened aortic regurgitation remains. Otherwise, criteria suitable for

acquired aortic stenosis or regurgitation apply.

***RECOMMENDATIONS**:*

- *In view of exercise capacity, congenital aortic stenosis should be treated like acquired aortic stenosis.*
- *For patients with light, asymptomatic aortic stenosis no restrictions for physical activities apply Symptoms, especially syncopal episodes, reduce exercise capacity based on individual evaluation.*
- *Patients with moderate aortic stenosis should only be active at low intensities.*
- *Severe aortic stenosis is, even if asymptomatic, not suited for any physical activity. The indication for an invasive correction of the malformation should be checked on a regular basis.*
- *Persisting aortic stenosis after Valvulotomie or interventional catheterisation techniques are to be considered in the same way as the untreated malformation in view of exercise tolerance (Calzolari 2001).*

Untreated Aortic Coarctation

Isolated stenosis of the aortic isthmus (AIS) causes a pressure increase in the prestenotic vasculature, depending on the severity of the malformation. In addition to the severity, other, associated cardiac malformations have to be considered. Most patients, especially at a young age, are asymptomatic. The severity of the AIS is defined by the pressure gradient across the narrowness, comparable to the classification of valvular aortic stenosis. At a gradient of > 50 mmHg between upper and lower body or persistent prestenotic arterial hypertension, operative intervention is required.

RECOMMENDATIONS:

- *For patients with mild, asymptomatic AIS, systolic pressure gradients<20 mmHg between upper and lower body and without pathological hypertension in the upper body, no restrictions for physical activities apply*
- *In patients with pressure gradients >20 mmHg at rest or pathological peripheral blood pressure readings, light dynamic, health-orientated exercise might be tolerated.*

Aortic Coarctation (corrected)

In a majority of patients, aortic coarctation is corrected surgically during childhood. However, even after this treatment, pathological pressure gradients, cardiac hypertrophy or peripheral, prestenotic hypertension can persist. In this context, the stress blood pressure regulation is of major importance. It is important to monitor young patients with history of this malformation on a regular basis, as rapid changes in the severity of their condition can occur during growth.

RECOMMENDATIONS:

- *For patients with fully corrected aortic coarctation without remaining pressure gradient or associated malformations, no restrictions for physical activities apply (6 months after intervention).*
- *If a significant pressure gradient remains, the recommendations for the untreated AIS should be followed. Accompanying malformations or cardiac adaptations such as aortic dilatation or aneurysmatic dysplasia of the stenotic area are reducing exercise capacity. Light dynamic, health-orientated exercise might be tolerated after careful individual evaluation.*

PULMONARY HYPERTENSION

Pulmonary Hypertension (PH) can be a sequel of any left-to-right shunt in congenital heart diseases. Furthermore, this condition can be idiopathic or the outcome of acquired diseases. In all cases, a high resistance in the pulmonary vasculature causes a right ventricular volume overload with enhanced risk of sudden cardiac death under exercise conditions. In most cases, PH progresses slowly towards an irreversible stage. The duration of this process is, however, highly variable. Patients which have been operated on cardiac malformations with apparent PH should be screened on a regular base for persistence of the PH after the intervention.

***RECOMMENDATIONS**:*

- *For patients with systolic pulmonary pressures below 40 mmHg and no signs of right ventricular adaptations or arrhythmias, no restrictions for physical activities apply.*
- *If pulmonary pressures exceed 40 mmHg and if no cyanosis at rest or during exercise is observed, light, health-orientated exercise might be tolerated if no other adaptive processes related to the PH are apparent.*
- *If at rest or during exercise, a cyanosis is present, the pulmonary systolic pressure at rest exceeds 40 mmHg or other pathological findings associated to the PH become apparent, patients should refrain from physical activity. Light dynamic, health-orientated exercise might be tolerated after careful individual evaluation.*

REFERENCES

1. Calzolari A, Giordano U, Di Giacinto B, et al. Exercise and sports participation after surgery for congenital heart disease: the European perspective. Ital Heart J 2: 736-9, 2001.
2. Carabello BA, Crawford FA. Valvular heart disease. New Engl J Med 337, 32-41, 2001.
3. Faratellone PM, Steinfeld L, Coplan NL .Exercise and congenital heart disease. Am Heart J 127: 1676-80, 1994.
4. Fredriksen PM, Veldtman G, Hechter S, et al. Aerobic capacity in adults with various congenital heart diseases. Am J Cardiol 87: 310-4, 2001.
5. Fritsch J, Winter UJ, Kaemmerer H, et al. Cardiopulmonary capacity of patients with congenital heart defects in childhood, adolescence and adulthood. Z Kardiol Suppl 3: 131-9, 1994.
6. Gabriel HM, Heger M, Innerhofer P, et al. Long-term outcome of patients with ventricular septal defect considered not to require surgical closure during childhood. J Am Coll Cardiol 39: 1066-71, 2002.
7. Garson A, McNamara DG. Sudden death in a pediatric cardiology population (1958-1983): Relation to prior arrhythmias. J Am Coll Cardiol 5: 134 B-137B, 1985.
8. Graham TP, Bricker T, James FW, et al. Congenital heart disease. Med Sci Sports Exerc S 246-S 253, 1994
9. Immer FF, Haefeli-Bleuer B, Seiler A, et al. Congenital heart defects: prevalence and course during the school years (8 to 16 years). Schweiz Med Wochenschr 124: 893-9, 1994.
10. Kaminer SJ, Hixon RL, Strong WB. Evaluation and recommendations for participation in athletics for children with heart disease. Curr Opin Pediatr 7: 595-600, 1995.
11. Kitchiner D. Physical activities in patients with congenital heart disease. Heart 76: 6-7, 1996.
12. Nugent EW, Freedom RM, Nora JJ, et al. Clinical course in pulmonary stenosis. Circulation 56 (Suppl) :I 38 – I 47, 1977.
13. Pelech AN, Kartdihardjo W, Balfe JA, et al. Exercise in children before and after coarctectomy: hemodynamic, echocardiographic and biochemical assessment. Am Heart J 112: 1263-1270, 1986.
14. Reybrouck T, Rogers R, Weymans M, et al. Serial cardiorespiratory exercise testing in patients with congenital heart disease. Eur J Pediatr 154: 801-6, 1995.
15. Reybrouck T, Bisschop A, Dumoulin M, et al. Cardiorespiratory exercise capacity after surgical closure of atrial septal defect is influenced by the age at surgery. Am Heart J 122: 1073-1078, 1991.
16. Schumacher G. Pulmonalstenose: In: Schumacher G, Hess J, Bühlmeyer K (Hrsg). Klinische Kinderkardiologie. Springer-Verlag, S 239-251, 2001.
17. Schumacher G, Hess J, Bühlmeyer K (Hrsg). Klinische Kinderkardiologie. Springer-Verlag, 2001.
18. Sklansky MS, Bricker JT. Guidelines for exercise and sports participation in children and adolescents with congenital heart disease. Prog Pediatr Cardiol 2: 55-66, 1993.
19. Singer H, Hofbeck M. Betreuung herzoperierter Jugendlicher und junger Erwachsener, In: Schuhmacher G, Hess J, Bühlmeyer G (Hrsg), Klinische

Kinderkardiologie S 555-562. Springer-Verlag, 2001.
20. Swan L, Hillis WS. Exercise prescription in adults with congenital heart disease: along way to go. Heart 83: 685-7, 2000.