Physical activity participation in youth with surgically corrected congenital heart disease: Devising guidelines so Johnny can participate

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While most youth with congenital heart disease have simple structural lesions and near-normal activity tolerance and functioning, youth with complex structural defects may be affected by significant morbidity. Efforts are being made to consider the impact of exercise and physical activity on quality of life. The benefits of physical activity are well documented; however, the levels of participation and self-efficacy for activity remain low, thus increasing the risk of obesity and cardiovascular morbidity. Rehabilitation is a novel and emerging area, and decisions regarding advocacy versus restriction of physical activity may pose a challenge for both cardiologists and nonspecialists. The present article outlines the rationale for physical activity recommendations in youth with surgically corrected congenital heart disease and the psychosocial determinants of participation. Clinical recommendations and future directions are proposed.

Key Words: Chronic illness; Congenital heart disease; Physical activity; Quality of life; Youth

THE CHANGING SCOPE OF PAEDIATRIC CARDIOLOGY IN NORTH AMERICA

One in 100 North American infants is born with congenital heart disease (CHD) each year. CHD encompasses a range of developmental defects along a continuum from mild lesions that spontaneously resolve without intervention, to severe and complex defects that are associated with life-long residual sequelae (1-3). Advances in diagnosis and management have altered the prognosis for patients with CHD; patients with simple structural lesions who have had successful repair are expected to have near-normal lives and functioning (2,4-6).

However, some youth with more complex structural lesions require closer evaluation and monitoring. These youth may experience a significant 'burden of disease' and a range of detrimental physical and psychosocial effects, including reduced exercise capacity and physical functioning, depression, anxiety, behavioural and conduct disorders, covert fears and low self-efficacy for physical activity. These difficulties persist and tend to 'track' to adulthood. Young adults with CHD may display increased dependence,

La participation à l'activité physique des jeunes ayant une cardiopathie congénitale corrigée par voie chirurgicale : Préparer des lignes directrices pour que Johnny puisse participer

La plupart des jeunes atteints d'une cardiopathie congénitale ont des lésions structurelles simples et une tolérance à l'activité ainsi qu'un fonctionnement quasi-normaux, mais les jeunes atteints d'anomalies structurelles complexes peuvent présenter d'importantes morbidités. On s'efforce d'évaluer les effets de l'exercice et de l'activité physique sur la qualité de vie. Les bienfaits de l'activité physique sont bien documentés, mais le taux de participation et d'auto-efficacité de l'activité demeure faible, ce qui accroît le risque d'obésité et de morbidité cardiovasculaire. La réadaptation est un secteur en émergence, et les décisions relatives aux prises de position par rapport à la restriction de l'activité physique peuvent représenter un défi à la fois pour les cardiologues et les non-spécialistes. Le présent article expose les principes des recommandations d'activité physique chez les jeunes ayant une cardiopathie congénitale corrigée par voie chirurgicale et les déterminants psychosociaux de la participation. Des recommandations cliniques et de futures orientations sont proposées.

difficulties with social and sexual relationships, and sub-optimal educational and occupational achievement (4,7-10). Academic failure and low IQ scores have also been reported in this group, especially if residual neurological complications are present (11). The 'burden of disease' affects families and caregivers; the mothers of youth with serious CHD experience caregiver distress, and encounter psychological and financial burdens related to their child's health (11,12).

The management of and advocacy for children with CHD is necessarily broad and comprehensive, and professionals increasingly consider health to be a multidetermined construct. Efforts are being made to reduce morbidity in children and youth across a range of chronic disease (10,13). Many are exploring the clinical efficacy of physical activity as a tool to enhance functional health status and health-related quality of life.

BENEFITS AND RISKS OF EXERCISE

Studies have demonstrated the cardiovascular, musculoskeletal and functional benefits associated with exercise for

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Figure 1) Classification: Dynamic and static components of popular physical activities. Max O₂ Estimated per cent of maximal oxygen consumption, which results in increased cardiac output; MVC Estimated per cent of maximal voluntary contraction, which results in increased blood pressure load. Reproduced from reference 30

youth with surgically repaired CHD (14). Supervised and incremental programs are associated with increased maximal oxygen consumption, peak oxygen consumption, peak oxygen pulse, physical activity levels, exercise tolerance, total treadmill walking time, heart rate recovery, walking distance, power output and oxygenation of respiratory muscles (5,14-20). In addition to increased muscular endurance, flexibility and coordination, exercise is associated with a reduction in cholesterol and total body fat percentage (14).

In addition to numerous physiological and functional benefits, exercise is associated with enhanced self-esteem, confidence, initiative, quality of life and perceptions of skill improvement (14,20). Conversely, there is an inverse relationship between poor health perceptions and chronic inactivity (8). The poor general health in paediatric Fontan patients is related to reduced physical functioning (10).

While physical activity and exercise participation are beneficial and should be encouraged for most youth with surgically corrected CHD, it is important to balance the numerous benefits of physical activity with a brief discussion of the slight risk of sudden cardiac death during activity in a subset of these patient groups. Youth with some structural defects, such as severe aortic valve stenosis or repaired tetralogy of Fallot, may encounter an increased risk of sudden death due to cardiac rhythm disturbances (21). Moreover, while a discussion of nonstructural cardiac conditions, such as cardiomyopathy, myocarditis and rhythm disorders such as long QT syndrome, are beyond the scope of the present article, primary care providers should be aware of the risk of sudden cardiac death during exercise and the activity recommendations for these groups (21). In addition, some CHD patients require long-term systemic anticoagulation, and physical activity should be advocated with appropriate precautions against injury that might result in bleeding complications (22).

BARRIERS TO PARTICIPATION

Despite the benefits of exercise, most youth with CHD are less active than healthy, age-matched peers and they do not derive the benefits of vigorous participation (23,24). This puts them at risk of becoming sedentary adults and acquiring latent chronic diseases (24). Cognitive processes appear to mediate the relationship between CHD and reduced participation. Studies (7,25) indicate that youth with CHD display poor self-efficacy toward physical activity, and that low self-efficacy may be a more important predictor of participation than the severity of the disease. Self-efficacy is a belief in one's ability to execute a particular task across a variety of contexts, and refers to perceived confidence. It is a powerful predictor of behaviour, and is influenced by physiological arousal, perceptions of mastery, vicarious learning and verbal persuasion. Adherence to recommendations may be more likely if patients believe in their ability to successfully undertake physical activity (26). Moreover, social and contextual barriers, such as social stigma, exclusion by teachers and peers in physical education, parental overprotection, liability concerns, low socioeconomic status, and lack of access to safe and enjoyable physical activity programs, may also impede participation for some youth with CHD (27).

DYNAMIC AND STATIC EXERCISE

It is important to consider whether the particular activity is dynamic or static (isometric). Dynamic activities, such as distance running or cross-country skiing, are associated with a change in muscle length from the forces exerted. Although these activities are associated with an increase in hemodynamic volume load, no significant pressure load is exerted on the heart. In contrast, while static activities such as weight lifting or disc throwing are not associated with a significant change in muscle length, an intramuscular force is generated. Thus, even in the absence of an increase in heart rate or hemodynamic volume load, these activities are associated with a pressure load on the heart (28,29). Primary care providers are advised to use the static/dynamic classification of the activity (Figure 1), as well as knowledge of the anatomy and physiology of the CHD to make appropriate recommendations (29,30).

EXERCISE RECOMMENDATIONS

In the absence of published guidelines for physical activity participation in youth with CHD, recommendations in the following section have been extrapolated from consensus statements on sport participation in CHD. Health care providers should be cognizant of this limitation in the existing literature base.

Children with left to right shunts, such as atrial septal defects and ventricular septal defects, as well as patent ductus arteriosus, constitute the largest group of patients with CHD. Repaired, asymptomatic patients with no residual shunts, and those with small unrepaired or residual shunts and normal pressure in the pulmonary artery, can safely engage in unrestricted recreational and competitive activities. Until repair, it is advisable to restrict symptomatic

patients with elevated pressures in the pulmonary artery from competitive activities (29).

Patients with obstructive lesions, such as aortic valve stenosis, pulmonary valve stenosis and coarctation of aorta, may experience fatigue and dyspnea on activity if the degree of obstruction is severe and limits exercise-induced augmentation of cardiac output. For patients with severe aortic valve stenosis who present with dyspnea, chest pain, syncope or important left ventricular hypertrophy, participation in competitive activities may be associated with an increased risk of sudden cardiac death (22,29). Until valvotomy or valve replacement has been undertaken to relieve the obstruction, it is advisable for both symptomatic and asymptomatic youth with moderate to severe stenosis to be restricted from strenuous competitive activities, particularly static exercise (29). If the stenosis is mild with a low-pressure gradient, asymptomatic repaired patients can safely participate in strenuous activities (29).

In the current era, the majority of patients with complex cyanotic heart conditions, such as tetralogy of Fallot, double outlet right ventricle and transposition of the great arteries (TGA), have undergone surgical repair or palliation. Thus, they demonstrate a good prognosis and moderate natural history, and liberal recommendations for both recreational and competitive activities may be advised. Patients who have undergone the atrial switch operation for TGA are now adults and, increasingly, they are a rare group. They have subnormal exercise capacity and more activity limitations. Due to psychosocial barriers, they tend to 'downgrade' their physical activity expectations (31). The majority of patients with TGA who are surgically repaired with an arterial switch operation can safely participate in recreational and competitive activities (32).

Unrepaired patients with severe complex CHD may experience intolerance to certain physical activities and increased hypoxemia on exertion. Eisenmenger's physiology with pulmonary hypertension is a complication associated predominantly with unrepaired, high-volume shunt lesions. This group may experience severe activity intolerance and self-imposed limitations. There is an important risk of sudden cardiac death during intense activities, and involvement in competitive activities should be avoided. Individualized physical activity programs for patients with Eisenmenger's syndrome may be permitted, provided that they are asymptomatic, free of arrhythmias and ventricular dysfunction, and have an oxygen saturation greater than 80% (29).

For children with univentricular hearts who have undergone the three-stage palliative Fontan procedure, a single ventricle is responsible for systemic circulation. These youth may have reduced exercise capacity and cardiac output, and may be unlikely to sustain involvement in strenuous activities (3,10,23,33). However, physical activity training can increase the exercise capacity of Fontan patients (3). After repair, and in the absence of significant symptoms, these patients may benefit from involvement in low-intensity or recreational activities (34).

For a detailed and comprehensive list of recommendations and restrictions for highly competitive sport participation, reference should be made to the report by the American College of Cardiology (30,34). Specific recommendations for patients with arrhythmias, myocarditis, hypertrophic cardiomyopathy and Marfan syndrome are beyond the scope of the present article. For these conditions, readers can refer to the reports by Maron et al (35) and Zipes et al (28) for comprehensive competitive sport guidelines. Recreational activity guidelines (22) for children with genetic cardiovascular conditions are also available.

CONCLUSION

Most patients with CHD have undergone successful repair and enjoy near-optimal functioning and well being. Some youth with more complex CHD may be affected by ongoing morbidity, and this has directed attention toward enhancing functional health status and quality of life. Physical activity has been proposed as a therapeutic mechanism for enhancing quality of life in paediatric patients with complex CHD. Despite the known physical, functional and psychosocial benefits of physical activity participation, the majority of these youth display low levels and intensity of participation.

In addition to known anatomical and functional limitations, low self-perceptions, social barriers and parental anxiety may mediate the relationship between CHD and reduced physical activity participation in some patients. Informed by the patient's diagnosis and surgical history, devising physical activity recommendations may be a promising and effective way in which to encourage young cardiac patients to safely participate in and enjoy life-long patterns of habitual, healthy physical activity.

Given the current state of knowledge, this process may be associated with particular challenges for health care providers. Well-established physical activity consensus statements should be devised, especially for noncompetitive sports participation. Informed by such guidelines, proposing recommendations for physical activity in youth with CHD involves research, shared decision making and carefully incorporated clinical judgment. The range of activities that a participant with a particular cardiac lesion can safely engage in must be evaluated, and the numerous benefits of participation should be balanced with the slight risks. By minimizing risk and maximizing quality of life, advocacy for physical activity may be considered as a crucial and routine component of patient management, and a catalyst by which to facilitate the attainment of a 'normal' childhood.

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