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Promotion of Physical Activity for Children and Adults With Congenital Heart Disease A Scientific Statement From the American Heart Association

Patricia E. Longmuir, PhD; Julie A. Brothers, MD; Sarah D. de Ferranti, MD, MPH; Laura L. Hayman, PhD, RN, FAHA; George F. Van Hare, MD; G. Paul Matherne, MD, MBA, FAHA; Christopher K. Davis, MD, PhD; Elizabeth A. Joy, MD, MPH, FACS; Brian W. McCrindle, MD, MPH, FAHA, Chair; on behalf of the American Heart Association Atherosclerosis, Hypertension and Obesity in Youth Committee of the Council on Cardiovascular Disease in the Young

The American Heart Association recognizes the importance of physically active lifestyles to the health and well-being of children and adults with congenital heart defects. Counseling of patients with congenital heart defects should emphasize the importance of daily physical activity and decreasing sedentary behavior as appropriate for the patient's clinical status. The suggested practices are based on relevant research regarding the benefits of physical activity for healthy children and adults, because research on physical activity among patients with congenital heart defects is lacking. There is no evidence regarding whether or not there is a need to restrict recreational physical activity among patients with congenital heart defects, apart from those with rhythm disorders. It is important to recognize that most patients with congenital heart defects are relatively sedentary and that the physical and psychosocial health benefits of physical activity are important for this population, which is at risk for exercise intolerance, obesity, and psychosocial morbidities. Therefore, counseling to encourage daily participation in appropriate physical activity should be a core component of every patient encounter.

Physically active lifestyles are important for children and adults living with congenital heart disease (CHD). The health benefits of physical activity accrue during activities of moderate intensity. Although higher intensity activity is required for changes to cardiorespiratory or musculoskeletal fitness, research demonstrates that activities of more moderate intensity provide significant health benefits¹⁻⁴ even if there are no

changes in measures of cardiorespiratory or musculoskeletal fitness. As such, it is important to recognize that this scientific statement clearly distinguishes physical activity from exercise or fitness. Physical activity is a broader concept that incorporates all types of physical movement, not solely those designated as exercise (planned, repetitive physical activity designed to increase fitness). Given that health benefits will accrue from physical activity of moderate intensity, the health benefits of a physically active lifestyle are within reach of nearly all patients with CHD. Healthcare professionals must tailor recommendations to each patient's clinical status, but the focus should be on the health benefits of physical activity⁵ and promoting appropriate participation for all patients. Children and adults with CHD should be encouraged to achieve recommended levels of physical activity.⁶⁻⁸ Very few patients with CHD will have disease that significantly impacts physical activity with family and friends. Similarly, only a very few diagnoses (such as risk of ventricular arrhythmia) necessitate activity restrictions. For most, physical activity can be unlimited and should be strongly promoted. Physical activity counseling should be part of every patient interaction, whether or not the patient's clinical status justifies activity restrictions.

Historically, cardiopulmonary exercise testing primarily has been used to evaluate morbidity and mortality in patients with CHD, with evidence accumulating of a link between exercise capacity and midterm outcomes.⁹ It is also helpful for examining changes in cardiovascular physiology during exercise

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and to evaluate the individual's risk for exercise-related morbidities. Knowledge of the patient's submaximal or maximal exercise performance may be useful in assessing the patient's ability to engage in recreational sport and physical activity and to reassure the patient and family about the patient's ability to be physically active. Subsequent testing can reinforce the exercise benefits obtained as the patient increases his or her physical activity participation. However, the scope of this statement is to describe best practices for healthcare providers promoting physically active lifestyles to children and adults with CHD.

Review of Current Guidelines

Current physical activity guidelines⁶⁻⁸ recommend that healthy adults perform muscle-strengthening activities ≥ 2 days per week and accumulate 75 minutes of vigorous to 150 minutes of moderate activity per week, with each activity session lasting ≥ 10 minutes. For children, ≥ 60 minutes of daily physical activity is recommended, with periods of vigorous activity performed at least 3 days per week. Muscle- and bone-strengthening activities are recommended at least 3 days per week. In contrast to more familiar aerobic activity, high-impact and anaerobic burst exercises such as jumping are most conducive to this goal. Sedentary behavior guidelines are also available for children and youth.⁶ Sedentary transport, extended sitting, and time indoors should be limited throughout the day. Screen time should be no more than 2 hours per day for children ≥ 5 years of age. Children < 3 years should have no screen time.

Guidelines specific to CHD, which are seldom evidence based, are available for adolescents or adults involved in intensive training for competitive sport and for individuals of all ages with genetic cardiac disorders or arrhythmias. In practice, the lesion-specific CHD recommendations for competitive sport from the 36th Bethesda Conference¹⁰ are commonly referenced; however, these recommendations apply only to the adolescent or adult athlete "who participates in an organized team or individual sport that requires regular competition against others as a central component, places a high premium on excellence and achievement, and requires some form of systematic (and usually intense) training." Highly competitive sport environments typically involve near-maximal training intensities. The European Society of Cardiology has also published recommendations for competitive sport in adolescent/adult athletes with cardiovascular disease.¹¹ Despite their clearly stated competitive focus, these recommendations often are inappropriately referenced for preadolescent children or for recreational sport, physical education, or unstructured physical activity at all ages.

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 published "Recommendations for physical activity, recreation sport, and exercise training in paediatric patients with congenital heart disease."¹² Based on a nonsystematic review of available data, the main point of the publication was to promote rather than restrict physical activity, an important paradigm shift. Restriction of physical activity or recreational sport is limited by individual clinical characteristics or

basic risks. Previous recommendations specific to adults with CHD¹³ encouraged dynamic rather than static exercise and emphasized that the physical and psychological health benefits of recreational physical activity have often been underappreciated. Rather than promoting one type of physical activity over another, it is important to recognize that the effects of dynamic and static exercise differ, and the health and fitness benefits they provide are also different.

In 2004, the American Heart Association published recommendations for unstructured physical activity, physical education, and recreational sport for young patients with genetic cardiovascular disease,¹⁴ primarily cardiomyopathy, long-QT syndrome, arrhythmogenic right ventricular dysplasia, and Marfan syndrome. The authors indicate that arrhythmia-related events are more common in genetic cardiovascular diseases, and events may occur more often with exertion, which makes precautions or restrictions for some higher-risk physical activities a reasonable approach. The authors provide a list of common sports; rate each as high, moderate, or low intensity; and then provide a grade on a scale of 0 to 5 for recommended participation (0-1, not advised/discouraged; 4-5, probably permitted). Casual play of children in the school yard and patients with structural CHD are excluded from these recommendations, largely on the basis of expert opinion. Two publications from the Heart Rhythm Society provide physical activity recommendations for individuals with supraventricular¹⁵ and ventricular¹⁶ arrhythmias, including patients with pacemakers and defibrillators. These recommendations are not specific to individuals with CHD but may be helpful for those patients who also have arrhythmias.

Benefits of Physical Activity

The numerous benefits of frequent and sustained physical activity, as well as the risks of inactivity for people with and without heart disease, have been documented extensively.¹⁷⁻²³ The benefits include well-described physiological improvements in skeletal muscle function, vascular health, immune system function, obesity prevention,²⁴ and complex psychological, cognitive and social function.²⁵ Emerging science also suggests that the health risks associated with inactivity, such as hypertension, diabetes, obesity, depression, cancer, and atherosclerotic cardiovascular disease, occur independent of the individual's physical fitness¹⁷ or compliance with daily physical activity recommendations.²¹ This indicates that patients who exercise vigorously for the recommended 60 minutes per day but also have prolonged sedentary periods will continue to be at increased risk.¹⁸

As a group, patients with CHD have lower levels of daily physical activity²⁶⁻²⁹ and a higher prevalence of obesity and other cardiovascular risk factors³⁰ than healthy children. Given the high survival rates for most CHD patients well into adulthood, these risk factors for acquired heart disease are becoming increasingly important, as is the need for higher levels of lifelong physical activity levels to combat them.

Physical Activity Assessment

Assessments of habitual physical activity should be part of the ongoing health evaluation of children and adults with CHD.

Table 1. Examples of Health-Related Fitness Protocols

Health-Related Fitness Dimension	Examples of Assessment Protocols
Cardiorespiratory fitness	20-m shuttle run, 15-m shuttle run, modified Canadian Aerobic Fitness Test
Flexibility	Sit and reach, back-saver sit and reach
Muscular strength	Handgrip strength, standing broad jump, vertical jump, Bosco jump protocol
Muscular endurance	Plank static hold, partial curl-up, curl-up, bent-arm hang, push-up
Body composition	Waist circumference, body mass index, waist-to-hip ratio, sum of skinfolds

Understanding a patient's risk for sedentary lifestyle morbidities entails more than cardiopulmonary exercise capacity. Physical literacy is an individual's capacity to attain and maintain the active lifestyle associated with positive health outcomes. Assessments of physical literacy evaluate current behavior, motor skill, and activity knowledge and motivation in addition to measures of health-related fitness.³¹ Motor skill delays are associated with sedentary lifestyles,³² and gross, fine, and visual motor function delays are common among children with CHD.^{33–35} Traditionally, it has been assumed that the motor delays that occur in children with CHD are related to early perioperative morbidity; however, the association of sedentary lifestyles with motor skill delays in children without a comparable medical history suggests that perioperative morbidities may not be the only reason for the prevalence of these delays in children with CHD. Although outcomes from cardiopulmonary exercise testing are linked to mortality, health-related fitness measures (Table 1) are associated with measures of health and daily living of more immediate concern to patients, including functional independence, quality of life, mobility disability,^{36–38} and features of the metabolic syndrome.³⁶ Protocols for assessment of exercise capacity,³⁹ motor skill,^{40,41} and health-related fitness^{37,42–44} are well established.

From a clinical perspective, the initial focus should be habitual activity behavior. Objective assessments of habitual

physical activity use accelerometers, pedometers, or heart rate monitors.⁴⁵ The pros and cons for these objective activity measures are summarized in Table 2. Although doubly labeled water is the “gold standard” for assessment of free-living physical activity,⁴⁶ it is very invasive and expensive and requires specialized techniques not suitable for clinical practice.

Free-living physical activity changes throughout the day and from day to day, so continuous assessment of physical activity during all waking hours is recommended.⁴⁷ Adults should be monitored for 2 weekdays and 2 weekend days,⁴⁸ with children monitored for 1 week.⁴⁹ The steps or activity recorded on the monitor should be invisible to the patient. Accelerometer measures of daily physical activity are valid for healthy individuals^{45,50,51} and those with CHD^{50,52} and are associated with New York Heart Association classification.⁵² Pedometers can also be used to encourage increased physical activity by having the number of steps taken displayed to the user. Current recommendations for optimal health⁵³ are for adults to achieve 10 000 steps per day and for children to achieve 12 000 steps per day.⁵⁴

Self-reports of physical activity significantly overestimate the quantity and intensity of activity performed,^{46,55} with reported activity often exceeding total daily time.⁵⁶ Therefore, use of self-reports is recommended only when more objective measures cannot be obtained. The Habitual Activity Estimation Scale was developed specifically for self-reported physical activity among clinical populations.⁵⁷ It requires the respondent (self or proxy) to indicate the percentage of time spent in sedentary, light, moderate, and vigorous activity for each period of the day. Because total activity reported must equal 100%, the reported activity cannot exceed the actual time available. Valid and reliable questionnaires can also be used to assess activity attitudes and preferences,⁵⁸ and these have been shown to predict future exercise behavior.⁵⁹ The frequency of neurocognitive, developmental, and learning disabilities among individuals with CHD, potential language barriers, and the need for parent proxy reports for younger children must also be considered when self-report instruments are used.

Table 2. Pros and Cons of Objective Measures of Daily Physical Activity

Assessment	Pro	Con
Heart rate monitor	Noninvasive Measures for full day over multiple days Relatively inexpensive	Heart rate changes may be unrelated to physical activity Not suitable for bathing or aquatic activities Requires a known relationship between heart rate response and physical activity work load
Pedometer	Noninvasive Relatively inexpensive Visibility of measurements to user varies between models	Only provides total activity per time period Measurement accuracy varies with cost Not suitable for bathing or aquatic activities Technical limitations to the detection and measurement of certain types of activity vary, depending on the model used and its placement on the body
Accelerometer	Noninvasive Activity intensity and duration Measures for full day over multiple days Shock and water resistant, with some models being suitable for aquatic activities Suitable for children and adults Measurements are not visible to user Recording of activity cannot be altered by user	Technical limitations to the detection and measurement of certain types of activity vary, depending on the model used and its placement on the body Initial cost of purchase is relatively higher than pedometers or heart rate monitors

Physical Activity Promotion

Daily physical activity is a cornerstone of cardiovascular health promotion that is influenced by many factors (patient, the patient's family, and contextual factors including school, work, and community).⁶⁰ Promoting physical activity to the patient with CHD is an important role for healthcare providers and requires a balance between the patient's activity interests and clinical assessment. Unfortunately, healthcare providers often indicate that they do not have the knowledge, skills, resources, or time needed to implement the extensive literature on physical activity promotion⁶¹⁻⁶³ or to counsel patients about physical activity.⁶⁴ These perceived limits support the literature that demonstrates that physical activity advice provided during routine primary care consultations does not effectively change objectively measured physical activity behavior,^{65,66} although there is a small positive effect on patient self-reports of activity participation.⁶⁶ In contrast, self-directed exercise that is undertaken with professional guidance and consistent professional support can result in behavior changes that are sustained over the long term.⁶⁷

Given the benefits of professional support and the limited resources of healthcare providers, it is important that healthcare providers be prepared to refer patients to physical activity professionals whenever the required physical activity counseling exceeds the healthcare provider's resources or scope of practice. Healthcare providers should provide patients with referrals to specific physical activity counseling resources, in the same way that they refer patients for dietary advice or therapy. Referrals should be only to individuals or organizations that hold widely recognized certifications in physical activity counseling and exercise prescription. Physical activity certifications should meet the same standards as other health professions, including mandatory undergraduate training, national examination of knowledge and competencies, and continuous education requirements. Advanced certifications offered by the American College of Sports Medicine and the Canadian Society for Exercise Physiology meet these criteria and include requirements for experience with cardiac populations.

Whether or not the patient will be referred to a physical activity professional, 3 aspects key to physical activity can be identified:

- The patient's readiness and/or ability to change behavior
- Physical activity dimensions important to or desired by the patient
- Clinical considerations relevant to the patient's physical activity

These key aspects will be addressed in greater detail in the following subsections.

Readiness/Ability to Change

Effective behavior change counseling (with children or adults) enables the individual to identify personally relevant goals and use intrinsic motivation to develop and implement action plans that are meaningful and appropriate (eg, motivational interviewing).⁶⁸ By identifying and building on the concerns of the patient, the clinician can bring to the fore the patient's intrinsic motivation for physical activity. The creation of a plan that

is consistent with patient/family values greatly increases the likelihood of sustained behavior change. This patient-centered approach also manages the resistance or defensiveness that is often created by a more prescriptive counseling style.

Successful behavior change depends on the "readiness for change" (ie, stage of change)^{69,70} of the patient and his or her significant others. The healthcare provider's task is to motivate and support behavior change through effective patient-centered counseling. Stage of change theory⁷¹ describes 5 stages that represent a continuum of behavior change, from precontemplation (the person is unaware of the need for change) to maintenance (the desired behavior has been achieved for ≥6 months). Motivational interviewing can be used to develop the patient's intrinsic motivation toward behavior change linked to the patient's beliefs and values through the use of non-judgmental, open-ended questions and reflective listening.^{68,70} Motivational interviewing skills are best developed through interactive learning. The "RULE" (Resist, Understand, Listen, Empower) acronym summarizes the 4 guiding principles:

1. Resist the "righting reflex," the natural tendency to overtly support the "good" or desired behavior. Although intended to be helpful, such comments often turn the patient's focus toward justification of why the negative behavior continues. Example: Healthcare professional (HP): "Increasing your daily physical activity would give you more energy and help to control your weight. It also helps to relieve stress." Patient (P): "I find I am really tired after I exercise, and it's even more stressful trying to fit one more thing into my already hectic schedule."
2. Understand and explore the patient's own motivation for change and perception of the current situation. Example: HP: "I know a lot of people feel really tired after they exercise. Some people like that feeling, but for others it makes it really hard to do all of the other things that have to be done that day. How long does that tired feeling last for you? What type of exercise are you doing that makes you so tired?"
3. Listen with empathy and attend to verbal and nonverbal communication from the patient as sustaining (or changing) the behavior is described. Example: P: "By the time I pick up my daughter from daycare and get home, I'm already rushed to make dinner. I don't have time to take her out to play, and by the time dinner is finished, it's dark." HP: "It sounds like your schedule is really packed, and it would be very difficult for you to make time to play outside with your daughter on weekdays. Can you tell me more about where you live? Are there places in your home or outside where your daughter could play while you are making dinner?"
4. Empower the patient, encouraging hope and optimism that they can impact their own health by changing adverse health behaviors. Example: P: "Every year I make a resolution to get fit, but my actions never live up to my enthusiasm." HP: "It's really hard to change our behavior. Old habits die hard, as they say. But I don't think that is a reflection of what you can achieve. I think it's more that you haven't had the best guidance in developing your plan to change. I know that if we help you to create a plan that you want to do, and that is really

Table 3. Important Individual and Activity Influences on Physical Activity

Important Activity Demands	Important Participant Abilities
<p>Dynamic and static exercise intensity</p> <p>Dynamic exercise is joint movement through relatively small forces within the muscle.⁷³ Static exercise develops relatively large intramuscular forces with little joint movement.⁷³ Energetic swimming would be primarily dynamic, whereas climbing on the monkey bars would have a greater static component (sport examples in Figure).</p> <p>CHD-related safety considerations</p> <p>Player/equipment collisions may injure the individual or damage implanted devices.⁷³ Consider the implications of a syncopal episode⁷³ (“Clinical Considerations Relevant to Physical Activity”).</p>	<p>Intrinsic motivation and beliefs about physical activity</p> <p>Activity guided by intrinsic motivation is typically 40% to 60% of maximal capacity to avoid fatigue. Fitness training occurs at 60% to 80% of maximal effort. More specific activity guidelines may be required for those highly motivated to excel.</p> <p>Perceptions regarding supports for and barriers to physical activity</p> <p>Effective physical activity counseling is based on the patient’s perceived supports for and barriers to activity. The significance of specific activities for the patient and family is also important. Activities that are a high priority for the patient/family can enhance motivation, whereas restriction of important activities is usually unsuccessful.</p>
<p>Authority of supervision</p> <p>Degree of oversight/influence on participation by someone in a position of authority.</p>	

CHD indicates congenital heart disease.

realistic for your lifestyle and all of your responsibilities, that you will be successful in making the changes that you want to see.”

Physical Activity Dimensions

Once a person with CHD indicates a desire to increase participation in physical activity, the next step is to determine the types of activities that should be encouraged (Figure). All types of physical activity (whether sport or activities of daily living) can be characterized by 6 attributes⁷²:

- Mobility (moving from place to place, moving the body in space)
- Object manipulation (control of equipment needed for the activity)
- Cognitive function (memory, understanding, relationships, etc)
- Behavior and social skills (appropriate behavior, interaction with others)
- Communication and perception (information to/from environment/others)
- Fitness (emotional and physiological)

Successful participation occurs when the attributes of the activity match the participant’s abilities and interests. Physical activity promotion and counseling should focus on increasing the overlap between attributes of the desired activity and the abilities of the person with CHD. In the setting of CHD, there are also important influences that may have to be considered depending on the individual’s clinical status. The most common of these influencing factors are summarized in Table 3. It is important to recognize that each of these activity- or participant-influencing factors may have either a positive or negative influence. An assessment of each dimension in conjunction with relevant clinical considerations will achieve a reasoned approach to promoting physical activity.

Among the influencing factors summarized in Table 3, the authority of supervision requires a brief discussion, because it is often unrecognized or is misinterpreted as being solely the impact of excessive demands by others. However, the full scope of the authority of supervision has 2 aspects:

- The source of control over the participant’s level of effort. Who ultimately controls the intensity of activity for the participant? Will the participant have complete control over the intensity of participation, or will participation be controlled by others in part or in whole (eg, coach, teammates, spectators, officials, sponsors)?
- Whether control by that source is sufficient to ensure that the patient remains within medically necessary activity restrictions. Will the need to decrease intensity or stop participation be recognized? If it is recognized, will it be respected and consistently acted upon?

An understanding of the authority of supervision is important when one counsels individuals with CHD in the setting of medically necessary activity restrictions. Although a clear explanation (preferably with specific examples) will ensure that the patient understands the required restrictions, it is the authority of supervision that will determine whether those restrictions are implemented consistently. Will the patient’s own intrinsic motivation (eg, competitive nature) lead to signs and symptoms or restrictions being ignored? Will a coach responsible for supervising the activity intensity consistently implement the required restrictions, even if it decreases the likelihood of winning? Will a teacher actually be able to determine when the restrictions have been exceeded (eg, students going on a cross-country run will be out of sight of the teacher for most of the activity)? These questions are just a few examples that illustrate the importance of understanding the source and effectiveness of the controls over the patient’s activity participation when activity restrictions are medically necessary.

Clinical Considerations Relevant to Physical Activity

The third component of providing effective and appropriate physical activity counseling is to consider the unique clinical status of the individual with CHD. Few, if any, clinical studies support the need for physical activity restriction or, conversely, establish the safety of all forms of physical activity for patients with CHD. Among those who urge caution,

the potential for sudden death is cited as the primary concern. However, the most common causes of sudden death during sport participation are either not related to CHD (eg familial hypertrophic cardiomyopathy, commotio cordis, myocarditis, dilated cardiomyopathy, long-QT syndrome) or are conditions that manifest with sudden death as the first symptom (coronary artery anomalies).⁷⁴ Although sudden death occurs in patients with repaired CHD (eg aortic stenosis, Mustard or Senning procedure), these events are seldom tied to physical activity or sport.⁷⁵ For example, sudden death events after aortic balloon valvuloplasty are unaffected by physical activity restrictions.⁷⁶ Other authors emphasize the low rate of arrhythmias with exercise in patients with known cardiac conditions.¹³ On the basis of these limited data, there appears to be a negligible risk of physical activity–related sudden death among most children or adults with CHD.

Although the risk of physical activity–related sudden death is sufficiently low for children or adults with CHD, there are some morbidities that influence the optimal activity type or intensity. Healthcare providers should consider the individual's current clinical status and encourage activities that will enable safe and enjoyable participation. If patients with CHD have arrhythmias, the recommendations for physical activity published by the Heart Rhythm Society^{15,16} should apply. Morbidities that may influence activity choices and counseling for patients with CHD include ventricular dysfunction, aortic dilation, syncope, hypoxia, anticoagulation, and device implantation.

It is important to recognize that each patient might have multiple morbidities. For example, a patient with tetralogy of Fallot might have significant right ventricular dysfunction and an implanted device. Each issue would inform the counseling provided to encourage appropriate physical activity participation. Healthcare providers should use standard diagnostic tests, including ECG, echocardiography, and cardiopulmonary exercise testing, to assess the physical activity–related capacity of each patient. An overview of the clinical assessment of CHD patients before recommendation of physical activity is provided in the next section. In the absence of actual evidence to better define physical activity–related risk, the approach to eligibility is necessarily somewhat arbitrary and must be individualized. Counseling to ensure patients accurately understand physical activity benefits, as well as risks, is essential.

Ventricular Dysfunction

Patients with significant ventricular dysfunction should be counseled to enjoy a wide range of recreational sport and physical activity opportunities, while limiting their involvement in competitive sport. The assumption is that significant ventricular dysfunction may lead to ventricular fibrillation during participation in intense competitive sport participation.⁷⁷ Although there is a lack of empirical data, it would seem reasonable to encourage patients such as those with coronary artery compression or insufficiency, important pulmonary hypertension, or severe valve obstruction to participate in noncompetitive activities that have low to moderate dynamic and static components (Figure). Recommended activities would include walking, bowling, cricket, bocce,

curling, baseball/softball, golf, or tai chi. For patients whose ventricular dysfunction is associated with arrhythmia, the recommendations of the Heart Rhythm Society should be followed.^{15,16}

Aortic Dilation in the Absence of Systemic Connective Tissue Disorders

Dilation of the aortic root and ascending aorta can occur in structurally normal hearts and in various forms of CHD, most notably in conotruncal defects and bicuspid aortic valve. There are common pathological features found in different structural defects, including abnormalities of transforming growth factor- β signaling.⁷⁸ Isolated aortic dilation often presents a dilemma for the physician with regard to recommendations for physical activity. Extreme dilation (diameter >99th percentile) is likely pathological⁷⁹ and can result in aortic aneurysm and dissection.⁸⁰ The risk of dissection is largely related to the size of the aorta, and general criteria for prophylactic aortic root/ascending aorta replacement do exist for adult patients.^{80,81} Before this intervention, recommendations for physical activity must account for the risk of dissection. Wall stress on the aorta is proportional to blood pressure, and blood pressure during exercise is generally related to the intensity of the exercise. Although dynamic exercise typically results in a larger increase in cardiac output, whereas static exercise results in greater afterload, this wall stress–to–blood pressure relationship with the risk of dissection is true for both static and dynamic exercise. Thus, when counseling these patients in regard to physical activity, limiting the intensity of the activity, regardless of type, is recommended. Activities of moderate intensity are generally safe. If patients desire participation in resistance/static-type exercises such as weight lifting, recommendations should be focused on lifting weights well below the 1-repetition maximum and on the use of appropriate technique, such as avoidance of the Valsalva maneuver during repetitions.

Syncope

Exertional syncope may be a concern for some CHD patients, such as those with valvular obstruction, arrhythmias, pulmonary hypertension, or sinus node dysfunction; however, empirical data to support or refute this assumption are lacking. Clinicians should aggressively investigate any documented syncope during exertion in CHD patients. Even if such investigation determines that there is no disease beyond the CHD diagnosis, the investigation results can inform counseling and activity promotion with the patient and significant others. Patients at risk for syncope, whether or not the episodes are triggered by or associated with exertion, should be counseled to participate in activities that would not endanger themselves or others should an episode occur. Patients should be encouraged to participate in activities such as walking, racquet sports, soccer, baseball/softball, table tennis, dancing, tai chi, yoga, bowling, and cricket. Activities such as horseback riding, gymnastics, rock climbing, or scuba diving should be avoided. Activities such as swimming, bike riding, ice skating, or skiing may be undertaken with supervision, unless episodes of syncope are common.

Increasing Static Component ↑	III. High (>50% MVC)	Bobsledding/Luge*†, Field events (throwing), Gymnastics*†, Martial arts*, Sailing, Sport climbing, Water skiing*†, Weight lifting*†, Windsurfing*†	Body building*†, Downhill skiing*†, Skateboarding*†, Snowboarding*†, Wrestling*	Boxing*, Canoeing/Kayaking, Cycling*†, Decathlon, Rowing, Speed-skating*†, Triathlon*†
	II. Moderate (20-50% MVC)	Archery, Auto racing*†, Diving*†, Equestrian*†, Motorcycling*†	American football*, Field events (jumping), Figure skating*, Rodeoing*†, Rugby*, Running (sprint), Surfing*†, Synchronized swimming†	Basketball*, Ice hockey*, Cross-country skiing (skating technique), Lacrosse*, Running (middle distance), Swimming, Team handball
	I. Low (<20% MVC)	Billiards, Bowling, Cricket, Curling, Golf, Riflery	Baseball/Softball*, Fencing, Table tennis, Volleyball	Badminton, Cross-country skiing (classic technique), Field hockey*, Orienteering, Race walking, Racquetball/Squash, Running (long distance), Soccer*, Tennis
		A. Low (<40% Max O ₂)	B. Moderate (40-70% Max O ₂)	C. High (>70% Max O ₂)
		Increasing Dynamic Component →		

Figure. Classification of sports by degree of peak dynamic and static components achieved during competition. Max O₂ indicates maximal oxygen uptake (reflects cardiac output demands); and MVC, maximal voluntary contraction (reflects blood pressure load). *Risk of bodily collision. †Increased risk if syncope occurs. Green represents the lowest overall cardiovascular demands; blue, low moderate; yellow, moderate; orange, high moderate; and red, the highest. Reprinted from Mitchell et al.^{81a} Copyright © 2005, with permission from Elsevier.

Hypoxia

Some CHD patients, such as those with elevated pulmonary vascular resistance and intracardiac shunting, may experience increasing cyanosis with effort. Exertion-related hypoxia generally does not require activity restriction, because the increasing hypoxia will usually restrict activity to an appropriate level. It is important to encourage patients at risk for exertion-related hypoxia to be physically active within comfortable limits, with the assumption that the authority of supervision and the individual's approach will allow self-regulation of activity intensity. The "talk test" (ie, activity at an intensity that permits easy conversation with others) may be a helpful tool for patients to guide their own activity intensity. The potential impact of altitude on oxygen saturation should also be considered for patients living in or visiting mountainous regions.

Anticoagulation

Anticoagulation poses a small risk of bleeding injury⁸² that is generally unrelated to the type of physical activity selected. Activities such as walking, jogging, swimming, cross-country skiing, or bike riding are unlikely to result in significant body impact and should be encouraged. Individuals taking anticoagulation medication should avoid activities in which body impacts are an intentional aspect of the sport, such as tackle football, combative martial arts, boxing, and ice hockey with body checking. Bleeding history, current medication status and level of anticoagulation, and patient skill and comfort in a particular sport should be considered for activities in which unintentional impacts may occur (eg, volleyball, basketball, baseball, diving, and skiing).

Device Implantation

For patients with implanted devices (ie, pacemakers and implantable cardioverter-defibrillators), the primary considerations when encouraging physical activity participation are

protection of the device and device leads and the prevention of soft tissue injuries from impact to the device. Activities that involve a high likelihood of direct blows to the chest or device, such as tackle football, boxing, and ice hockey with body checking, should be discouraged. Some authors⁸³ have suggested that implantable cardioverter-defibrillators may offer insufficient protection during physical activity, should an arrhythmia that requires a shock occur, because the reliability of these devices has not been established during high levels of sympathetic activation. This view has been questioned, however, with only 10% of physicians surveyed⁸⁴ encouraging their patients with implantable cardioverter-defibrillators to limit their participation to low-intensity physical activity, such as golf. Currently, there are no published data regarding physical activity recommendations for individuals with other types of implanted devices (eg, right ventricle-to-pulmonary artery conduits, stents, valves), and research to document the physical activity in these populations is recommended.

Clinical Assessment of CHD Patients Before Recommendation of Physical Activity

A thorough assessment of a patient's individual activity level, tolerance to exercise, and general approach to physical activity and fitness begins with a detailed history. It is important to elicit the presence of exertional symptoms such as angina, excessive dyspnea, palpitations, dizziness, and syncope (previous sections). Even in the absence of such symptoms, there are many diseases that require objective assessment of the exercising patient before a particular physical activity is recommended. Formal exercise testing is recommended for most of these patients to better refine any particular patient's risk of exercising and to benchmark his or her current level of submaximal fitness as it relates to successful physical activity participation. This may also have the additional benefit of reassuring both patients and their families that if

Table 4. Variables Typically Measured During a Cardiopulmonary Exercise Test

\dot{V}_{O_2} max	The highest amount of oxygen that the body can consume during maximal exercise. The achievement of \dot{V}_{O_2} max is recognized when there is no increase in heart rate or oxygen consumption although the exercise workload is increased or when the ratio of $\dot{V}_{CO_2}:\dot{V}_{O_2}$ exceeds 1.1. \dot{V}_{O_2} max may be reported in liters per minute for exercise modes that are independent of body weight (eg, stationary cycling) or in liters per minute per kilogram of body weight for exercise modalities that require the participant to support their own body weight (eg, treadmill walking/jogging). The measurement of \dot{V}_{O_2} max is typically feasible only in highly trained or highly motivated individuals (eg, athletes).*
\dot{V}_{O_2} peak	The highest amount of oxygen consumed during a maximal exercise test to the limit of voluntary effort. This is the most common variable used to report aerobic exercise capacity. \dot{V}_{O_2} peak is the outcome of an exercise test that is terminated because the participant is unable or unwilling to continue. \dot{V}_{O_2} peak may or may not be equal to \dot{V}_{O_2} max, depending on whether the maximal oxygen transport capacity of the body is achieved before or after the limit of voluntary effort. \dot{V}_{O_2} peak may be reported in liters per minute for exercise modes that are independent of body weight (eg, stationary cycling) or in liters per minute per kilogram of body weight for exercise modalities that require the participant to support their own body weight (eg, treadmill walking/jogging).*
Ventilatory anaerobic threshold	The point at which \dot{V}_{CO_2} and \dot{V}_E begin to increase out of proportion to \dot{V}_{O_2} . This is a measure of submaximal work rate that is sensitive to the effects of exercise training. It has the advantage of not being affected by patient motivation and does not require a maximal effort.*
Ventilatory efficiency	The ratio and/or slope of \dot{V}_E/\dot{V}_{CO_2} , otherwise known as ventilatory equivalent of CO_2 . This is a physiologically important variable that is decreased (ie, the slope is greater) in patients with heart failure, cyanosis, and pulmonary disease. A higher value indicates that more breathing effort must be used to eliminate the same amount of CO_2 .*
Work rate	A surrogate measure of muscle power and exercise capacity. Typically measured in watts for exercise performed on a cycle or arm ergometer.
Treadmill time	This is a surrogate measure of exercise capacity when a treadmill protocol is used. Means and standard deviations of treadmill times for various age groups have been published.
Heart rate profile	This is composed of resting heart rate, peak heart rate, heart rate reserve (peak rate minus resting rate), and heart rate recovery. Resting heart rate and heart rate recovery in particular can be improved with improved fitness.
Blood pressure	Arm blood pressure responses to exercise are measured during a test and can reveal hypertension not otherwise discovered with resting values alone.
Pulmonary function	Standard pulmonary function tests using spirometry can be performed to reveal obstructive, restrictive, or mixed pulmonary disease at baseline or as a result of exercise.
Oxygen saturation	Reliable oxygen saturation monitoring can be helpful in patients with resting desaturation and those at risk for desaturation during exercise (eg, those with fenestrated Fontan circuits).

\dot{V}_{CO_2} indicates carbon dioxide production; \dot{V}_E , minute ventilation; and \dot{V}_{O_2} , oxygen consumption.

*Requires measurement of exhaled O_2 and CO_2 during the test.

no significant risk factors are identified, even intense exercise can be enjoyed safely, even by those CHD patients who have a potentially limited maximum exercise capacity. Furthermore, conveying the results of an exercise test provides an opportunity to ensure that patients understand that daily physical activity and increased fitness do not require a maximal effort, but rather that fitness benefits are best gained with exercise that allows conversation (ie, the talk test). Ensuring that patients understand that maximum exercise capacity is unrelated to daily physical activity and fitness may provide patients with the realization that their suboptimal fitness levels can be improved through an increase in their physical activity levels. Table 4 lists many of the clinically important variables measured during an exercise test. Table 5 includes risk factors

of exercise that may be uncovered during testing that can be used for clinical recommendations for the type, frequency, and intensity of physical activity.

Physical Activity Counseling Practices

Suggested practices for physical activity counseling among patients with CHD are summarized below. Suggested practices are described rather than formal recommendations made because of the paucity of scientific data specific to physical activity among patients with CHD. Currently, the body of knowledge for optimal type or quantity of physical activity and methods for activity assessment or counseling is limited to healthy populations. It is recommended that funding agencies prioritize support for research to develop scientific

Table 5. Exercise Risk Factors Evaluated During a Cardiopulmonary Exercise Test

1. Abnormal hemodynamic response to exercise: Either a progressive drop in blood pressure (a normal response is an increase in systolic pressure and little or no change in diastolic pressure) or excessive rise in blood pressure (>250 mmHg systolic), especially when it results in symptoms. Patients with coarctation of the aorta, repaired or unrepaired, may have normal resting right arm blood pressure but significantly elevated pressure during exercise.⁸⁵
2. Abnormal heart rate profile: A blunted peak heart rate, lower heart rate reserve, and blunted heart rate recovery are associated with poorer overall prognosis in various forms of congenital heart disease.⁸⁶
3. Arrhythmias: Supraventricular and ventricular tachycardia, ventricular fibrillation, and all forms of heart block can manifest during exercise testing.
4. Myocardial ischemia: On electrocardiography during exercise, significant ST depression of ≥ 2 mm can signify ischemia induced by the extra metabolic demands of exercise.
5. Reduced ventilatory efficiency: An increase in the \dot{V}_E/\dot{V}_{CO_2} slope is associated with heart failure and an increased risk of sudden death in some congenital heart disease populations.^{87,88}

\dot{V}_{CO_2} indicates carbon dioxide production; and \dot{V}_E , minute ventilation.

evidence regarding physical activity among children and adults with CHD. Until sufficient high-quality research data are available, specific evidence-based recommendations cannot be provided.

Counseling Practices to Promote Physical Activity

Physical activity counseling can be provided by physicians, nurses, exercise science professionals, or other allied health professionals with appropriate training. For patients with congenital heart defects, it is important for the health professional to accomplish the following:

- Promote physical activity based on general population guidelines⁶⁻⁸ for patients who have had a successful repair of their congenital heart defect without residual sequelae.
- Ask about and encourage patients and families at every visit to meet the recommended amount of daily physical activity.
- Ensure that patients and families understand there are health risks that occur with prolonged sedentary periods, which can occur even when the patient meets the daily physical activity guidelines.
- Have the patient (or parent) report the patient's typical activity behavior during clinic wait times so the results can be reviewed and discussed to increase awareness of discrepancies between actual and recommended activities. The Habitual Activity Estimation Scale⁵⁷ is a simple method of asking about the proportion of time spent in sedentary, light, moderate, or vigorous activity for each portion of the day.
- Explain the physical activity implications of all medication or treatment changes, including the implications of discontinuing a previous treatment (eg, when anticoagulation therapy is discontinued), identified changes in cardiac function, or when previous activity restrictions (eg, immediately after sternotomy) are no longer required.
- Assess the patient's stage of change, by comparing self-reported need for change to measurements of physical activity participation, when assessing exercise capacity or physical activity participation.
- Encourage patients to identify realistic and measurable goals when counseling them to change their physical activity behavior; avoid prescribing behavior changes not identified and prioritized by the patient.
- Counsel about physical activity using patient-centered communication and recognize the patient's readiness for change; refer patients to professionals experienced in counseling physical activity behavior change, when available.
- Counsel patients to build activity time throughout their day so that they accumulate 30 minutes (adults) to 60 minutes (children) of moderate to vigorous activity most days of the week; children are intermittently active, and most adults find it difficult to clear uninterrupted time for physical activity.
- Acknowledge the time constraints that dominate current lifestyles and encourage patients to develop specific plans for building physical activity into a typical day.

- Encourage children to participate in school-based physical activity (physical education, recess, after school sports, etc) and provide parents with information so they can effectively communicate their child's ability to participate to school officials.
- Emphasize the health benefits of light or moderate activity and breaking up extended periods of sedentary time with light activity.
- Advise parents to strictly limit screen time, and emphasize screen time limits at each clinic visit so that children have no screen time before 3 years of age and children ≥ 5 years have no more than 2 hours per day.⁶

Counseling Practice When Activity Restrictions Are Required

- Promote physical activity to all patients, even when activity restrictions are medically necessary, so that patients and significant others are encouraged to enjoy the types of physical activity that are appropriate.
- Ensure that patients and families understand the reason for activity restrictions and provide examples of activities that would not be restricted so that they can make appropriate decisions regarding activity opportunities that were not specifically discussed during the visit.
- Avoid general statements such as "no body contact" or "no competitive sports," which patients and families find difficult to interpret.
- Provide the patient and family with an opportunity to ask questions or propose "*what if's*" for how new or unexpected activity opportunities should be assessed (eg, "What if my child is at a sleepover and the children are imitating wrestling? What if the teacher divides the students into 'teams' for a 'tournament' in gym class?").
- Ask the patient about any physical activity experiences of concern at each clinic visit (either before participation or in hindsight).
- Assess and monitor exercise response in patients with physical activity-related risk every 3 to 5 years, or more often for those whose risk may increase, using exercise assessment protocols that mimic the physical activities that the patient wishes to perform, to demonstrate the patient's capacity for physical activity, inform activity counseling, and provide updated information on exercise capacity changes.
- Teach patients who need to monitor their activity intensity to use the "talk test," because people who can talk comfortably will limit their activity intensity to 60% to 80% of maximum.
- For patients with ventricular arrhythmias, encourage activities with low- and moderate-intensity dynamic and static components.
- If episodes of syncope are of concern, encourage activities that are not associated with a significant injury risk if the patient should experience an episode of syncope during participation, such as walking, jogging, cross-country skiing, racquet sports, golf, and noncombative martial arts.
- For patients with exertion-related hypoxia, encourage physical activity at an intensity that does not worsen the hypoxia to dangerous levels.

- Counsel patients receiving anticoagulation regarding the degree of impact avoidance required, particularly in relation to activities with unintentional or less forceful impact (eg, basketball, figure skating, gymnastics).
- For patients with aortic dilation, encourage light- to moderate-intensity dynamic exercise and the use of the talk test to monitor the level of effort.

Practices for Physical Activity Assessment

- Monitoring of physical activity participation by use of objective measurements is reasonable, because pedometers and accelerometers are inexpensive and widely available and can be used for comparison of patient results to population data (eg, National Health and Nutrition Examination Survey,⁸⁹ Canadian Health Measures Survey⁹⁰), the overreporting bias of self-reports is significant, and accurate information about current participation can inform activity counseling.
- Monitor motor skill development at key time points, such as toddler, preschool, school entry, and transition to middle school, to identify patients who require a referral for remedial support and to optimize the opportunity for successful participation in physical activity with family and friends.
- Monitor health-related fitness every 3 to 5 years, through the use of measures of submaximal aerobic capacity, muscular strength and endurance, flexibility, back strength, and body composition, because comparison of results to population data (eg, Canadian Health Measures Survey⁹¹) can inform patient counseling and identify patients who do not have adequate fitness levels for activities of daily living and safe participation in desired types of physical activity.

Healthcare Provider Training

- Healthcare providers caring for patients with CHD should understand the physical and psychosocial benefits of regular physical activity and be prepared to discuss those benefits with their patients.
- Physical activity counseling training curricula should be developed and implemented so that trainees gain knowledge and experience related to physical activity promotion and counseling, in addition to cardiopulmonary exercise testing, and healthcare providers can acquire and maintain physical activity knowledge.
- Consider referral to a physical activity specialist for specific patient counseling, because specialists (certified through the American College of Sports Medicine, the Canadian Society for Exercise Physiology, or equivalent) will have more in-depth knowledge, expertise, and experience in achieving substantial changes to actual physical activity behavior, are skilled at motivating initial and continued participation, and have extensive knowledge of the physiological impact of specific activities within a variety of settings.
- Healthcare providers should be educated regarding the professional scope of practice of those with training in

exercise or physical activity science (eg, kinesiologists, physical educators, or exercise specialists) and recognize that the knowledge required for exercise prescription or physical activity counseling is typically not included in medical or rehabilitation curricula.

Policy and Advocacy

- Healthcare providers should advocate for multilevel policy changes designed to increase physical activity opportunities, both within the daycare/school/workplace and in the community, so that physically active lifestyles are readily available for children, adolescents, and adults.
- Advocate for community design initiatives that increase physical activity opportunities, such as active transportation options, safe walking facilities, daily school physical activity, employment activity incentives, and sidewalk connectivity.
- Design cardiac clinic environments to promote active play by providing activity-promoting toys (eg, blocks, cars, wire tracks with beads to push, whiteboard paint on walls for drawings), removing televisions/video games/fragile decorations, and clearly conveying the health benefits/risks of active/sedentary behavior, respectively.

Research Recommendations

As stated at the outset, these recommendations are based primarily on expert opinion because of the lack of relevant research-specific physical activity among individuals with CHD. The following research recommendations are provided to encourage the development of an evidence-based body of knowledge:

- Amount (dose, duration, and intensity) of physical activity participation necessary to optimize physical and psychosocial health outcomes in children and youth whose CHD alters the known physical activity-to-health outcome relationship
- Amount (dose, duration, and intensity) of physical activity required to enhance fitness in children and youth with CHD who cannot follow the FITT (Frequency, Intensity, Time, Type of activity) training principles
- Effective and efficient strategies to increase physical activity knowledge and awareness of its importance among CHD patients and their families
- Longitudinal studies to examine growth, development, and health benefits of varying levels of physical activity participation for patients with CHD
- Optimal timing and format (eg, provider counseling, e-health) of approaches and strategies for promoting physical activity participation in children and adults with CHD
- Evaluation of physical activity counseling training curricula and referral strategies, with emphasis on cost and quality outcomes
- Optimal methods (and timing) for assessing and monitoring adverse responses to physical activity participation in children and adults with CHD
- Describing the adverse physical and psychosocial health consequences of sedentary behavior in children and adults with CHD

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*Modest.

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