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47.1 Introduction

Children participate in sports all over the world and should have a pre-participation physical evaluation (PPE) before the season begins.

The 2010 consensus guidelines suggest a PPE for children.

The primary goal is to maximize safe participation for all, identify medical problems with risks of life-threatening complications during participation (e.g., hypertrophic cardiomyopathy) and conditions that require a treatment plan before or during participation (e.g., hypertension), rehabilitate old musculoskeletal injuries, treat conditions that interfere with performance (e.g., exercise-induced bronchospasm), and remove unnecessary restrictions on participation.

The PPE should take place 4–6 weeks before the season starts, permitting time to evaluate and treat medical problems and/or rehabilitate musculoskeletal injuries before sports participation.

Most children with chronic medical conditions can participate in a sport at some level after appropriate evaluation and/or treatment. There are some exceptions like cervical spine stenosis and they cannot participate in contact sports.

Sudden death in the young athlete occurs with prevalence of between 1:100,000. The risk of sudden death is disproportionately higher in males. The median age was 17 years.

47.2 Cardiovascular Conditions

Cardiovascular conditions causing sudden death in young athletes include:

- Hypertrophic cardiomyopathy
- Coronary artery anomalies
- Commotio cordis
- Myocarditis
- Aortic rupture (Marfan syndrome)
- Arrhythmogenic right ventricular hypertrophy
- Long QT syndrome
- Wolff-Parkinson-White syndrome
- Aortic stenosis

Most athletes who die suddenly have no symptoms of life-threatening cardiovascular disease, and the PPE is not efficient in detecting them. However, athletes suspected of having these conditions on the basis of historical or physical findings must not participate until further evaluation by a cardiologist.

Sudden cardiac death during exercise in patients with mitral valve prolapse is rare. *Athletes with mitral valve prolapse can participate in all competitive sports unless the following exist:*

- A history of syncope documented to be arrhythmogenic in origin
- A family history of sudden death associated with mitral valve prolapse
- Repetitive forms of supraventricular and ventricular arrhythmias, particularly if exaggerated by exercise
- Moderate to marked mitral regurgitation
- Prior embolic event
- Uncontrolled stage 2 hypertension – It is recommended that uncontrolled stage 2 hypertension (systolic and/or diastolic blood pressure [BP] ≥ 99 th percentile plus 5 mmHg) or end-organ damage (e.g., retinal, renal, or cardiac changes) requires exclusion from sports participation and highly static activities until it is better controlled
- Fever – Children and adolescents with fever should be restricted from participation because fever may accompany myocarditis or other infections that can make exercise dangerous

47.3 Other Relevant Conditions

In addition to cardiovascular abnormalities, numerous other medical conditions should be identified before clearance for sports participation because the conditions are associated with increased risk of adverse outcome or injury if left untreated. Examples of these conditions include:

- Exercise-induced bronchoconstriction (EIB) occurs in athletes at a prevalence similar to that of the general population (9–15%), yet it may be unrecognized in the young athlete. Pulmonary disease accounts for 2% of sudden death in sports. EIB can be treated with pre-exercise medication in most patients and is not a reason to avoid exercise. The use of post-exercise spirometry in the routine PPE is not recommended
- Eating disorders (e.g., anorexia nervosa, bulimia nervosa) can manifest as excess exercise and malnutrition. Persistent exercise in the malnourished amenorrheic female athlete can cause short- and long-term consequences
- Some form of regular exercise is likely to be beneficial in most children and adolescents with diabetes mellitus. However, modifications in the pre-exercise insulin dose and additional glucose monitoring are necessary
- Athletes at risk of heat illness should follow guidelines for appropriate clothing, fluid intake, heat acclimatization, adjustment of activity level for heat and humidity levels, and timing of practices

Whether or not they are at risk of heat illness, athletes should consume fluid 2 h before prolonged exercise and every 20 min during activity. Electrolyte replacement drinks (i.e., sports drinks) are recommended after the first hour of prolonged exercise. The volume of intake varies according to the athlete's weight:

- 40 kg–500 mL, 2 h before prolonged exercise and 150 mL every 20 min during activity
- 60 kg–750 mL, 2 h before prolonged exercise and 250 mL every 20 min during activity

- Concussion – There is no evidence-based guideline regarding return to play for child athletes (i.e., 5- to 12-year-olds) following sports-related concussion. For the young athlete with repeated concussions, the decision to return to contact sports should be based on the number of concussions, the mechanism of previous concussions, the duration of recovery, and the time in between injuries
- Musculoskeletal injuries – Identifying and fully rehabilitating old musculoskeletal injuries have the greatest yield for identifying problems that will interfere with subsequent performance because injuries are common among athletes. Players with injuries to an extremity are more likely to injure that extremity during the season than an extremity that has not been injured. Proper rehabilitation can lead to lower injury rates
- Adolescent athletes with Osgood-Schlatter disease report stopping training and sports participation for months, and this may be with clinician advice. However, Osgood-Schlatter disease is a common problem and, although painful, should result in little if any restriction from sports activity when managed appropriately
- Obesity is a risk factor for heat injury, and exercise is an important component of obesity management, and restricting exercise is contraindicated in this setting

Incremental aerobic exercise tests are performed in children and adolescents for a variety of reasons. The primary indication is to provide the clinician with information about a young patient's physical working capacity. *The information gained from an aerobic exercise test is helpful in determining:*

The athlete's history of previous injury should draw the clinician's attention to assess for residual effects. In addition to providing a plan for rehabilitating strength, endurance, and proprioceptive and flexibility deficits, the clinician should provide the athlete with a plan for returning to play. The athlete is at risk of re-injury and delayed recovery if he or she returns to competition too soon. Training errors, such as too-rapid increases in pace, distance, repetitions, or weight/resistance, are the most common factor in overuse injuries.

Another goal of the PPE is to remove unnecessary restriction on participation in sports or an exercise program because they are believed to have cardiac disease. As examples:

- One study of the morbidity of cardiac non-disease identified 93 seventh- to ninth-grade students who had "something wrong with their hearts" according to school records. After pediatric cardiology evaluation, 75 of these 93 students (81%) were found to have no cardiac disease, yet 30 of these 75 students (40%) had activity restrictions ranging from being homebound to being able to participate in physical education classes but not competitive sports

- Whether a patient can perform daily activities within his or her functional capacity
- Whether he or she is responding appropriately to an exercise intervention program
- Whether chronic disease progression is affecting the patient's physical capacity

Contraindications – Exercise testing can be performed in most children. However, it is contraindicated in children with certain medical conditions. As a general rule, the exercise test should begin at a low workload so that the child becomes accustomed to the exercise and surroundings. In some cases, he or she may need to practice before beginning the test.

Exercise testing protocols may be continuous or discontinuous. When comparing results of two tests performed on an individual patient (e.g., pre- and post-exercise training), it is important to perform both tests using the same protocol and exercise modality.

- Continuous – Test protocols are usually continuous (i.e., without rest periods) and have either ramped or incremental stages
- In discontinuous exercise protocols, children are permitted to rest between stages. As an example,

each exercise stage might last 2–3 min, with 1–2 min of rest in between. Discontinuous protocols also may be more appropriate for children who are unfit and have low exercise tolerance

The average maximum heart rate in children and adolescents is considered to be 200 bpm with a wide range of individual values. It may vary by 5–10 bpm within an individual child performing different protocols. Most researchers accept an exercise test to be a maximal effort if the child’s maximum heart rate is greater than 95% of predicted HR_{max} (i.e., HR ≥190 bpm).

- Exercise-induced bronchoconstriction (EIB) affects up to 80% of individuals who have asthma

Exercise testing is a useful tool in the diagnosis of EIB and for evaluating the exercise capacity and cardiopulmonary response to exercise in the child with asthma. Children with EIB commonly present with post-exercise coughing and chest pain; wheezing and dyspnea also may be present. The maximum heart rate criterion is not a good indicator of effort intensity in children with EIB.

- Cystic fibrosis – Compared with those with sedentary lifestyles, children, adolescents, and young adults with cystic fibrosis (CF) who exercise regularly may recover more quickly from acute illnesses. In addition, the use of exercise as an adjunct treatment to clear mucus in CF patients may result in fewer respiratory infections

In one cohort study, 109 CF patients age 7–35 years underwent pulmonary function and exercise testing and then were followed for 8 years. Survival rates were greatest among patients with the highest levels of aerobic fitness (83%, 51%, and 28% among those with VO_{2peak} ≥82%, 59–81%, and ≤58% of predicted, respectively). Patients with higher levels of aerobic fitness were more than three times likely to survive than those with lower levels of aerobic fitness after adjustment for other risk factors.

- Idiopathic pulmonary arterial hypertension is considered by some to be a contraindication to maximal exercise testing in children, and it is not performed

Submaximal exercise testing also may be a valuable tool for assessing the prognosis and treatment of children with idiopathic pulmonary arterial hypertension. In many clinics, the 6-min walk test is given in lieu of maximal testing.

- Children and adolescents who have arthritis of any type may be less physically active than their healthy peers. Reasons for inactivity include chronic joint pain and stiffness, reduced strength, synovitis, and/or joint deformity

Children and adolescents who have arthritis appear to have decreased aerobic capacity for a variety of reasons. In one comparison study of aerobic capacity and workload completed by children with juvenile idiopathic arthritis (JIA, formerly juvenile rheumatoid arthritis, JRA) and healthy children during cycle ergometer exercise, children with JIA had a significantly lower VO_{2peak} (33.0 mL/kg per min versus 46.9 mL/kg per min). No direct relationship was found between functional aerobic capacity and disease severity in the affected children. The authors speculated that the lower VO_{2peak} values in children with JIA appear to be caused by either mechanical inefficiency or hypoactivity.

- In children who have neuromuscular disease, exercise performance is usually limited by decreased muscle function rather than cardiorespiratory capacity. Exercise testing of these patients can provide a quantitative assessment of the child’s condition, the improvement in economy of locomotion after surgical treatment, and the potential effects of exercise stress

Contraindications for exercise testing in children and adolescents

Acute inflammatory cardiac disease (e.g., pericarditis, myocarditis, acute rheumatic heart disease)
Uncontrolled heart failure
Acute myocardial infarction
Acute pulmonary disease (e.g., acute asthma, pneumonia)
Severe systemic hypertension (e.g., blood pressure greater than 240/120 mmHg)
Acute renal disease (e.g., acute glomerulonephritis)
Acute hepatitis (within 3 months after onset)
Drug overdose affecting cardiorespiratory response to exercise (e.g., digitalis toxicity, salicylism, quinidine toxicity)
Severe aortic stenosis
Severe pulmonary stenosis

Serious ventricular dysrhythmia, especially when associated with significant cardiac disease
Coronary arterial diseases (anomalous left coronary artery, homozygous hypercholesterolemia, Kawasaki disease [acute phase])
Severe pulmonary vascular disease
Metabolic disorders (glycogenolysis types I and V)
Hemorrhagic diseases
Orthostatic hypotension

Adapted from Washington RL, Bricker JT, Alpert BS, et al. Guidelines for exercise testing in the pediatric age group. From the Committee on Atherosclerosis and Hypertension in Children, Council on Cardiovascular Disease in the Young, the American Heart Association. *Circulation* 1994; 90:2166 and James FW. Exercise testing in children and young adults: an overview. *Cardiovasc Clin* 1978; 9:187

Indications for terminating pediatric exercise testing before reaching maximal voluntary capacity level

The onset of serious cardiac arrhythmias (e.g., ventricular tachycardia, supraventricular tachycardia)
Any appearance of potential hazard to the patient
Failure of electrocardiographic monitoring system
Symptoms such as pain, headache, dizziness, or syncope, precipitated by exercise
Segmental ST depression or elevation ≥ 3 mm during exercise
Arrhythmia (over 25% of beats) precipitated or aggravated by exercise
Recognized types of intracardiac block precipitated by exercise

The European Society of Cardiology (ESC) has proposed guidelines for pre-participation screening for young athletes planning to begin competitive sports, which includes a standard 12-lead electrocardiogram (ECG), based upon a national screening program that has been in effect in Italy since 1982.

The following recommendations were made:

- An initial complete personal and family history and physical examination should be performed before beginning training and competition
- The evaluation should be performed by a clinician with specific training, medical skill, and cultural background to identify clinical symptoms and signs associated with cardiovascular diseases associated with sudden cardiac death (SCD). In Italy, clinicians primarily responsible for these examinations are trained in postgraduate sports medicine programs full

time for 4 years and work in sports medical centers dedicated to periodic evaluation of athletes

- Screening evaluations should be repeated at least every 2 years
- A 12-lead ECG should be obtained (seeking evidence of a standardized list of abnormalities). If a specific diagnosis is considered, more detailed ECG review may be helpful
- Patients with abnormal findings on history, physical examination, family history, or ECG are referred for further testing, such as echocardiography, ambulatory monitoring, exercise treadmill testing, or cardiac magnetic resonance imaging (MRI)

The potential advantage of the ECG is most commonly attributed to its ability to detect hypertrophic cardiomyopathy, in which the ECG is abnormal in up 95% of patients.

The routine use of ECG screening is the risk of false-positive results. The prevalence of such findings was addressed in a series of 32,652 Italian subjects who underwent routine pre-participation screening that included an ECG. The prevalence of markedly abnormal ECG patterns suggestive of significant structural heart disease was <5%. However, these results cannot be generalized to other countries. In addition to the potential impact of genetic differences, the nature of pre-participation screening is unique in Italy, where it is performed by trained and licensed sports medicine specialists who practice in dedicated sports clinics.

Over 8 years, four athletes were found to have borderline left ventricular hypertrophy (LVH) (13 mm). One was later confirmed to have hypertrophic cardiomyopathy (HCM) by genetic analysis and a second was considered to have possible HCM. In addition, 12 athletes were diagnosed with other cardiac structural abnormalities including mitral valve prolapse, myocarditis, Marfan syndrome, arrhythmogenic right ventricular cardiomyopathy, and bicuspid aortic valves. The screening ECG also can detect arrhythmogenic right ventricular cardiomyopathy, long QT syndrome, and Brugada syndrome.

Sudden cardiac death (SCD) associated with athletic activity is a rare but devastating event. Victims are usually young and apparently healthy, but many have underlying cardiovascular disease

that is not diagnosed until after the event. As a result, there is great interest in detecting such abnormalities early and then defining appropriate activity restrictions for affected individuals to minimize the risk of SCD.

The majority of SCD events in athletes are due to malignant arrhythmias, usually sustained ventricular tachycardia (VT) degenerating into ventricular fibrillation (VF), or primary VF itself. Although definitions vary, “young” often refers to high school and college athletes, but applies in general to individuals under age 35 in whom SCD is usually due to congenital heart disease. Older, or “masters,” athletes include individuals over age 35, in whom SCD is most commonly due to coronary heart disease (CHD).

In general, *patients with known genetic disorders that predispose to SCD* (e.g., hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, Marfan syndrome, long QT syndrome) should avoid recreational activities with the following characteristics:

- “Burst” exertion, involving rapid acceleration and deceleration, as is common in sprints, basketball, tennis, and football. Activities with stable energy expenditure, such as jogging, biking on level terrain, and lap swimming, are preferred
- Extreme environmental conditions (temperature, humidity, and altitude) that impact blood volume and electrolytes
- Systematic and progressive training focused on achieving higher levels of conditioning and excellence

Patients with unusual or high-risk clinical features may require greater restriction. These features include a history of syncope or pre-syncope, prior cardiac surgery, prior arrhythmic episodes, or an implantable cardioverter-defibrillator (ICD). It is widely acknowledged that SCD is the leading medical cause of death in athletes, although its exact incidence remains unclear.

An overall incidence of 1:50,000 per year in young athletes is a reasonable estimate based on existing information from retrospective cohort

studies and prospective observational and cross-sectional studies. Male athletes are consistently found to be at greater risk, and there appears to be a disproportionately higher risk among male African-American athletes.

Structural heart disease can increase the risk for SCD by one or more of the following mechanisms:

- Ventricular tachyarrhythmias (most common cause)
- Bradyarrhythmia or asystole
- Syncope
- Dissection of the great vessels, as in patients with Marfan syndrome

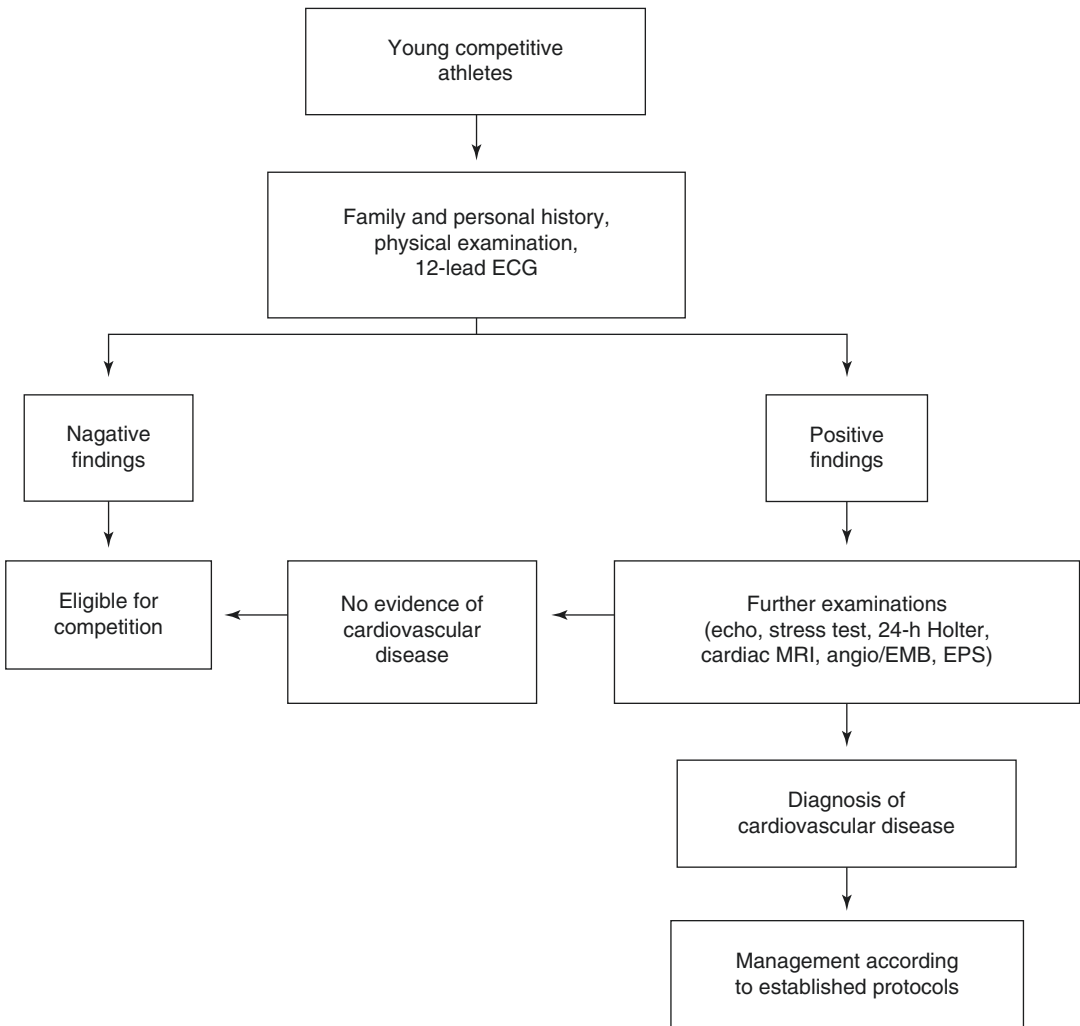
Hypertrophic cardiomyopathy (HCM) is a relatively common disease, occurring in 0.16–0.29% of individuals in the general population (one in 350–625). *Congenital coronary artery abnormalities* were found in 12–33% of young athletes with SCD. The most common anomalies associated with SCD are the origin of the left main coronary artery from the right sinus of Valsalva and the origin of the right coronary artery from the left coronary sinus. Athletes with Marfan syndrome, familial aortic aneurysm or dissection, or congenital bicuspid aortic valve with any degree of ascending aortic enlargement should not participate in sports that involve the potential for bodily collision.

Myocarditis was present in 6–7% of cases of SCD in competitive athletes. Active myocarditis is associated with atrial and ventricular tachyarrhythmias, and bradyarrhythmias.

The incidence of SCD among competitive athletes is actually quite low, estimated to be between 1 per 50,000 athletes and 1 per 300,000 athletes.

Sudden cardiac death associated with athletic activity is a rare but devastating event. Victims are usually young and apparently healthy, but many have underlying cardiovascular disease that is not diagnosed until after the event. As a result, there is great interest in detecting such abnormalities early and then defining appropriate activity restrictions for affected individuals to minimize the risk of SCD.

Flow diagram illustrating the proposed screening protocol for young competitive athletes



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