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REVIEW

The role of exercise testing in pediatric cardiology

Intérêt du test d'effort en cardiologie pédiatrique

Martial M. Massin

Division of Pediatric Cardiology, Queen Fabiola Children's University Hospital, Free University of Brussels (ULB), 15, avenue J.J.-Crocq, 1020 Brussels, Belgium

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Child

Summary Exercise testing for cardiac disease in children differs in many aspects from the tests performed in adults; their cardiovascular response to exercise presents different characteristics, which are essential for the interpretation of hemodynamic data. Moreover, diseases that are associated with myocardial ischemia are very rare in young patients, and the main indications for exercise testing are evaluation of exercise capacity and identification of exercise-induced arrhythmias. This article describes the specificity of exercise testing in pediatric cardiology, in terms of techniques, indications and interpretation of data.

MOTS CLÉS
Cardiologie pédiatrique ;
Test d'effort ;
Cardiopathies congénitales ;
Arythmies ;
Enfant

Résumé Le test d'effort chez les enfants cardiaques diffère par de nombreux aspects de celui réalisé chez l'adulte ; leur réponse cardiovasculaire à l'effort présente des caractéristiques dont la connaissance est essentielle à l'interprétation des données hémodynamiques. De plus, les maladies ischémiques sont exceptionnelles chez les jeunes patients et les principales indications du test d'effort sont une évaluation de la capacité physique et l'identification d'arythmies induites par l'exercice physique. Cet article décrit les spécificités techniques, les indications et l'interprétation des données en cardiologie pédiatrique.

Abbreviations: ECG, electrocardiogram; LQTS, long QT syndrome; PCO_2, partial pressure of carbon dioxide; PO_2, partial pressure of oxygen; SVT, supraventricular tachycardia; VAT, ventilatory anaerobic threshold; VE, minute ventilation; VCO_2, carbon dioxide production; VO_2, oxygen consumption; VT, ventricular tachycardia.

E-mail address: martial.massin@huderf.be

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Background

Standardized exercise testing has almost become a routine procedure in the care of children with cardiac disease [1]. Impairment of functional capacity is usual in this population and may be the result of the primary cardiac problem, its treatment or hypoactivity. The test is often conducted to provide objective information about exercise capacity, to identify abnormal responses to exercise, to make management decisions, to assess the efficacy of medical and surgical interventions, to evaluate exercise-related adverse events, to define individual safety limits, to instill confidence in child and family, and to motivate patients to engage in physical activity, resulting in improved patient outcomes.

Procedure

An analysis of the possible gain in information expected from the test versus the potential harm induced is mandatory before a test is performed, even if adverse reactions are rare [2]. An exercise test should not be carried out if the patient has an acute infection, acute exacerbation of a chronic disease, or any other unstable health condition that may pose an additional transient risk. Severe obstructive diseases, pulmonary hypertension, severe heart failure or certain arrhythmias warrant special consideration. An experienced physician and emergency equipment should always be present during the test.

According to international recommendations [1], the test should be terminated when diagnostic findings have been established and further testing will not yield any additional information, when monitoring equipment fails, when signs or symptoms indicate that further testing may compromise the patient's well-being, and when extreme fatigue or other symptoms of insufficient cardiac output are associated with decrease or failure of heart rate to increase with increasing workload. Other criteria are progressive fall in systolic blood pressure with increasing workload, systolic hypertension > 250 mmHg, diastolic hypertension > 125 mmHg, intolerable dyspnoea or tachycardia, progressive fall in oxygen saturation < 90% or a 10-point drop from resting saturation in a symptomatic patient, 3 mm flat or downward sloping ST-segment depression, triggering of atrioventricular block or QTc lengthening > 500 ms and increasing ventricular ectopy with increasing workload, including a triplet.

Defining that an effort was maximal may sometimes be difficult, but is very important to determine maximal exercise data. A plateau of oxygen uptake (i.e. an increase during the final completed stage of an incremental exercise test of < 2 mL/kg/min for a 5—10% increase in exercise intensity or of < 2 standard deviations of the average increase in oxygen uptake during the preceding stages) occurs in half of the children [3]. In other cases, the investigators have to rely on their impression that a maximal effort has occurred and on indicators such as heart rate > 200/min on a treadmill or > 195/min on a bicycle, breathing reserve < 40% or a respiratory exchange ratio of $\geq$ 1 on a treadmill or 1.05 on a bicycle [4].

Exercise testing in children, especially sick young children, is more challenging than in adolescents and adults. Treadmill exercise testing is possible in children from the age of 3 years, but a harness with a rope and one extra-staff member may be necessary to safeguard the child during the test. Nevertheless, owing to the strong resemblance to daily activities, the larger muscle mass involved and the lower probability of leg muscle fatigue, a treadmill test may be especially advantageous in very small or very sick children, as well as increasing the likelihood of the cardiorespiratory system being the limiting factor. Because cycle ergometry requires that the subject maintains a cycle cadence with increasing workloads (especially with a mechanically braked cycle ergometer that cannot vary resistance to keep workload constant over a range of pedal speeds) and because the patient must be big enough to reach pedals, exercise testing on a bicycle is hardly possible before the age of 6 years. Bicycle testing is portable and less expensive than treadmill testing, and accurate determination of the workload is easy. Moreover, the upper body is more stable on a cycle ergometer than on a treadmill, so a bicycle is preferred when the underlying condition requires accurate blood pressure assessment or ischemia detection on an electrocardiogram (ECG). However, many subjects may not be accustomed to cycling and premature muscular fatigue during bicycle testing may prevent them from reaching maximal effort.

Numerous exercise protocols have been used in children. Most laboratories use continuous graded protocols requiring a maximal effort, even if important data can be obtained during submaximal exercise [4]. The choice of protocol depends on the age and body size of the child to be tested, the measurements planned and the equipment available. Hemodynamic and gas exchange responses are better, and determination of the ventilatory threshold is easier in protocols with shorter stage durations, the so-called ramp protocols, whereas a steady state for more physiological functions (heart rate and oxygen uptake) requires exercise stages of $\geq$ 3 minutes. In general, the total exercise duration should be kept to 6—12 minutes in children, to avoid premature muscle fatigue and lack of attention and motivation. For treadmill exercise testing, most laboratories use the Bruce treadmill protocol [5,6], in which increase in work rate is accomplished by increasing speed and grade every 3 minutes. Disadvantages of the Bruce protocol are large interstage increments in work that can make estimation of maximal values less accurate, and intermediate stages than can be either run or walked, resulting in different oxygen costs. Alternatives, with a slower increase in workload, are more appropriate for unfit patients; an example is the Balke protocol, in which the speed of the belt is held constant and the increase in work rate is accomplished only by increasing the slope. Different protocols are also used for cycle ergometry [7]. When an electronically braked cycle ergometer is available, a continuous ramp protocol is usually preferred. Increments in workload can be increased by 5 to 20 W/min, depending on height, weight or body surface area [8], or can be standardized by 0.25 W/kg/min [9]. Additional procedures are used to answer specific questions, such as the Wingate test protocol to determine anaerobic power, step-like increases of workload to assess oxygen uptake kinetics, or a 4—10-minute intense exercise bout to induce bronchoconstriction.

Exercise testing with ECG monitoring, but without gas analysis, is sufficient when only abnormal blood pressure

Table 1 Main cardiopulmonary exercise testing variables.

Variable	Interpretation	Indications
Peak VO_2	↓ if cardiopulmonary dysfunction or deconditioning	CHD, CM, PAH, potential transplant recipients, complete atrioventricular block
Maximal heart rate	↓ if chronotropic insufficiency	Operated CHD, LQTS, transplanted heart
	↓ if efficacy of beta-blockade	CHD/CM with heart failure, arrhythmia
ECG	Exercise-induced arrhythmia	CHD, primary arrhythmia
	Ischemic changes	Kawasaki disease, coronary arterial anomalies (congenital or post-repair in Ross or arterial switch operation)
	Other exercise-induced changes	LQTS, Brugada, WPW, pacing system
Oxygen pulse	↓ if limited stroke volume adaptation	CHD, TCPC, CM
Oxygen saturation	↓ if pulmonary disease, intracardiac or intrapulmonary shunts	CHD
Blood pressure	↓ if cardiac dysfunction	CHD, CM (hypertrophic CM++), PAH
	↑ if arterial hypertension	Aortic coarctation
VAT	↓ if cardiopulmonary dysfunction or deconditioning	CHD, CM, rehabilitation programs
VE/VO_2, VE/VCO_2 slopes	↑ if ventilatory inefficiency (ventilation/perfusion mismatch)	CHD with heart failure or right-to-left shunt, operated Fallot, PAH, potential transplant recipients
Pulmonary function tests	↓ if co-existent pulmonary problems	CHD with multiple thoracotomies, right-to-left shunt or ventilation/perfusion mismatch

CHD: congenital heart disease, CM: cardiomyopathy, ECG: electrocardiogram; LQTS: long QT syndrome; PAH: pulmonary arterial hypertension; TCPC: total cavopulmonary connection; VAT; ventilatory anaerobic threshold; VE: minute ventilation; VCO_2: carbon dioxide production; VO_2: oxygen consumption; WPW: Wolff-Parkinson-White syndrome.

response, electrocardiographic changes or arterial oxygen desaturation must be detected. Exercise testing is also performed without gas analysis when the patient cannot tolerate the mouthpiece or the facemask (e.g. very young or very stressed children). Endurance time for treadmill protocols [6] and peak work rate for bicycle protocols are then used as an index of exercise capacity. However, their usefulness is limited: the normal range for these variables is quite broad in children, they are influenced by factors unrelated to the cardiopulmonary system, optimal effort cannot be ascertained, and little information is provided regarding the cause of exercise intolerance. Therefore, exercise testing usually includes an expiratory gas analysis and variables relevant to the assessment of cardiopulmonary adaptation to exercise are then calculated from breath-by-breath estimates of oxygen consumption (VO_2), carbon dioxide production (VCO_2), minute ventilation (VE) and end-tidal partial pressures of oxygen (PO_2) and carbon dioxide (PCO_2).

Outcome variables

Peak oxygen uptake (peak VO_2) is generally seen as the gold-standard variable for assessing aerobic capacity and cardiac function (Table 1). Peak VO_2, calculated as the average VO_2 over the last 30 seconds of peak exercise, should be used instead of maximal oxygen uptake, as half of untrained children and most heart failure patients do not achieve a plateau phase [4]. Owing to recruitment of more muscle groups, children usually achieve ± 10% higher peak VO_2 values on a treadmill than on a bicycle [10,11], and ± 20% higher peak values during upright cycling than during supine cycling [11], but no difference is noted between similar protocols [11]. Peak VO_2 is usually expressed in mL/min/kg body mass [12,13]. This expression underestimates the aerobic capacity of overweight and taller subjects.

Maximal workload (in watts) can easily be determined on a bicycle, but for a treadmill test it may be only estimated from the maximum speed and incline. It is important to use

the same protocol to evaluate the trend in achievement of a given patient because the maximal power depends on the exercise protocol chosen, as long stages lead to a muscle fatigue at lower workloads compared with a protocol with shorter stages. Maximal power increases with age and, after puberty, the increase is greater in males than in females. Maximal power is commonly around 3.0—3.5 W/kg in healthy boys and 2.5—3.5 W/kg in healthy girls.

A 12-lead ECG should be used during exercise testing. The limb electrodes must be placed supraclavicularly, near the shoulders, and suprailiac. Resting heart rate and heart rate at a given workload decrease with growth, whereas peak heart rate is independent of age and sex in children, but depends on the mode of exercise, testing on a treadmill leading to higher maximal heart rates than on a bicycle. An increase in heart rate during exercise is delayed in athletes, but is more rapid than normal in deconditioned children or in those with heart failure. The decline in heart rate during recovery is much faster in the youngest children [13]. Heart rate response in cardiac patients must be interpreted very carefully because many have chronotropic incompetence [4]. An ECG can also detect exercise-induced arrhythmias and ischemic or QTc changes.

The oxygen pulse, which is the VO_2 per heartbeat, represents the product of stroke volume and arteriovenous oxygen extraction. This last variable changes predictably during exercise because, at peak exercise, oxygen extraction is maximized and mixed venous oxygen saturation varies little across a wide spectrum of cardiac function, so that oxygen pulse can be used as a surrogate marker for stroke volume. As it is difficult to measure cardiac output accurately by non-invasive methods in children, oxygen pulse is often used to assess ventricular function during exercise in children. Interpretation is difficult, however, when oxygen extraction is abnormal (e.g. in case of anaemia, polycythaemia, hypoxemia and metabolic or muscular disorders). Stroke volume has a higher relative contribution to cardiac output during the initial stages of exercise and the O_2 pulse kinetics profile is hyperbolic. A reduced rise in O_2 pulse is noted during exercise when stroke volume adaptation is limited (e.g. in case of ventricular dysfunction, obstructive cardiac or vascular disease or valvular regurgitation). The oxygen pulse is also decreased in Fontan and total cavopulmonary connection patients [14], probably because of the limited ability of the passively perfused pulmonary vascular bed to accommodate the high rate of blood flow during severe exercise. By comparison, oxygen pulse is usually normal at peak exercise in case of chronic aortic regurgitation because the fall in systemic vascular resistance during exercise lessens the severity of the regurgitation, and the dilated left ventricle helps to maintain forward stroke volume. Other ratios have been proposed, such as the VO_2 efficiency slope [15], which analyses the relationship of VO_2 versus the logarithm of VE and indicates the status of systemic and pulmonary perfusion, or work efficiency, which analyses the relationship of VO_2 versus work rate [16] or, during graded treadmill exercise, versus estimated exercise intensity [17].

Oxygen saturation is measured by pulse oximetry using a finger or forehead sensor. Oximeters may have difficulty tracking the signal at peak exercise because they are motion sensitive. Exercise-induced arterial desaturation usually occurs in patients with pulmonary diseases or relevant intrapulmonary or intracardiac shunts.

Blood pressure measurement is essential if patients with cardiac diseases or suspected exercise-induced cardiovascular symptoms are tested. During exercise, systolic blood pressure increases whereas diastolic blood pressure remains relatively constant [18]. A lack of increase or even a decrease in blood pressure may be related to cardiac dysfunction, although a drop can also occur in healthy children [2]. An increase in diastolic blood pressure during exercise may indicate hypertension.

The ventilatory anaerobic threshold (VAT) is an indirect indicator of the onset of metabolic acidosis during graded exercise; it is usually defined as the VO_2 immediately below the exercise intensity at which pulmonary ventilation increases disproportionally relative to VO_2. VAT is determined by a sudden increase in the ventilatory equivalent for O_2 (VE/VO_2) without an increase in the ventilatory equivalent for CO_2 (VE/VCO_2), a sudden change in the slope of the regression line between VO_2 and VCO_2 (V-slope method) or by changes in the respiratory exchange ratio. In any case, it is recommended to use 10—15-second data averages instead of breath-by-breath data, and to plot gas exchange data over VO_2 rather than over time [19]. VAT is expressed as a percentage of peak VO_2, decreases with age [20], suggesting an increase in lactic acid anaerobic capacity during growth, and is lower than normal in children with cardiac disease [4,21]. VAT cannot be determined in all individuals from a single exercise test, and analysis of the same data set may reveal some interobserver variability. The anaerobic threshold is determined from a series of blood lactate measurements during an incremental exercise test. The work rate at which the lactate-over-work rate curve steepens considerably defines the anaerobic threshold. However, it is rarely performed in children because it usually does not add relevant information and because blood samplings are not well tolerated by young patients.

Ventilatory equivalents are often used to detect ventilatory inefficiency. The ventilatory equivalent for oxygen (VE/VO_2) and the ventilatory slope (VE/VCO_2) have a higher slope in cardiac patients. Pulmonary blood flow maldistribution and consequent ventilation/perfusion mismatch are probably the main factors that underlie this elevation; they result from pulmonary vascular obstructive disease in pulmonary hypertension, residual peripheral pulmonary stenoses in operated tetralogy of Fallot and elevated pulmonary capillary wedge pressure in heart failure. The ventilatory slope is also elevated in case of right-to-left shunt because the respiratory drive is increased by the CO_2-rich systemic venous blood entering the systemic arterial circulation.

Many cardiac patients also have co-existent pulmonary problems, which can contribute to exercise intolerance; they tend to have mild-to-moderate restrictive changes resulting in smaller lung volumes and flow rates, leading to a $\pm$ 25% reduction in ventilatory capacity on average [22]. A restrictive lung disease due to intrathoracic fibrosis or restriction of chest wall motion must be especially suspected in patients who have had multiple thoracotomies. Therefore, in cardiac patients with exercise intolerance, pulmonary function must be assessed by baseline spirometry, measurement of maximal voluntary ventilation (forced

expiratory volume in 1 second multiplied by 40) and breathing reserve (percentage of maximal voluntary ventilation that is not used at peak exercise and is normally ± 30%), and exercise tidal flow-volume loops [4,22]. The analysis of exercise end-tidal PCO_2 is also useful; lower values will be observed in case of ventilation/perfusion mismatch (the air from alveoli with high ventilation/perfusion ratio dilutes air from other alveoli) and right-to-left shunt (PCO_2 of the blood returning from the lung is reduced to compensate for the shunted hypercapnic blood to systemic circulation), whereas higher values will be noted in case of airway obstruction or hypoventilation due to obesity. When exercise-induced bronchospasm is suspected in patients with chest pain, exercise testing with pre- and postexercise pulmonary function before and after giving an inhaled beta-agonist, can be useful.

Exercise testing in congenital heart disease

The main reason for exercise testing in congenital heart disease is to assess physical capacity, to obtain objective information about the functional status of the heart; this can be used to determine whether complaints have a cardiac cause, to provide recommendations for physical activity [23], to give indications for treatment (heart failure therapy, surgery, transcatheter intervention) and to evaluate the success of interventions [24].

Heart disease may adversely affect hemodynamic adaptations to exercise in a variety of ways. Lack of a pulmonary ventricle in palliated univentricular heart, ventricular dysfunction, valvular disease, shunts, obstructions, abnormal pulmonary vasculature and associated pulmonary and musculoskeletal disorders may impair the cardiopulmonary response to exercise.

Heart rate dynamics during exercise are often abnormal in operated congenital heart disease. A lower peak heart rate is usually defined as chronotropic incompetence when < 80% of the predicted value is reached during exercise testing. This is influenced by the surgery itself and is usually not observed after transcatheter therapy [25]. Cardiac denervation with lack of sympathetic reinnervation and damage to the sinus node or its blood supply, leading to sinus node dysfunction, may play a role; it is more frequent after palliation of univentricular heart or atrial switch [4,26] and, logically, after cardiac transplantation. As the increase in cardiac output is only obtained by an increase in heart rate beyond moderate intensity exercise, chronotropic impairment may reduce exercise capacity.

Exercise evaluation is recommended before individualized exercise prescription in at least the following conditions [27]: closed atrial or ventricular septal defect with persisting pulmonary hypertension, myocardial dysfunction, symptomatic tachyarrhythmias or significant heart block; moderate aortic stenosis; untreated mild coarctation of the aorta or repaired aortic coarctation, even with good results; atrial [17] or arterial switch operation [28—30] for transposition of the great arteries; congenitally corrected transposition of the great arteries; repaired tetralogy of Fallot [17] and other repaired cyanotic congenital heart defects, including Fontan and total cavopulmonary connections [4]; Ebstein's anomaly; and repaired congenital coronary artery anomalies.

Regular physical activity is associated with many health benefits in the general population but also in cardiac patients. Overprotection is common in these children. The resulting sedentary lifestyle leads to diminish physical work capacity and places the children at risk of early development of adult cardiovascular disease and other illnesses associated with inactivity. Physical fitness in patients who have undergone an apparently successful correction is not only related to cardiac function but also to strength, coordination, flexibility, muscular endurance and motivation. Exercise testing can instill confidence in children and parents. In detrained young patients, the provision of a postoperative rehabilitation program will allow them to improve their level of physical activity within a few months and the benefits are maintained for at least a few years. Hospital programs (but also home programs) have been developed [31]. However, the literature on pediatric cardiac rehabilitation programs is poor and the most effective strategies are still to be elucidated. Although patients with serious heart disease are advised to avoid high-intensity competitive sports, many will benefit from participation in physical activities in a less competitive, recreational environment. Most guidelines are of limited value for the majority of the patients because they focus predominantly on competitive sports and base the decision-making process on the individual anatomical lesions. A novel approach is to formulate clinically useful recommendations relating to physical activity, based on hemodynamic and electrophysiological variables [23]. In such an approach, cardiopulmonary exercise testing can help to provide an individualized recommendation after interpretation of peak VO_2, maximal heart rate, Borg scale, blood pressure response to exercise, transcutaneous saturation during exercise and detection of rhythm and conduction disorders during exercise [23].

Many factors contribute to the decision to transplant patients with congenital heart disease. Rigorous assessment enables the most appropriate candidates to be selected for cardiac transplantation. To stratify the level of risk for outcome of transplantation and to classify the patients as either urgent or standard depending on their prevailing clinical condition, various investigations are undertaken, including metabolic exercise testing. Peak VO_2, autonomic response to exercise and ventilatory equivalents contribute to the overall evaluation of the condition of the potential transplant recipient; however, their value is limited, as numerous exercise protocols are used [32].

Exercise testing in acquired heart disease and cardiomyopathies

The severity of dilated myocardiopathies may be evaluated by observing effort tolerance, blood pressure response, electrocardiographic alterations and arrhythmias. Exercise testing, with determination of VAT and peak VO_2, is one of the most important tests for evaluating the severity of cardiac insufficiency, disease progression and the effectiveness of treatments. Metabolic exercise testing may also provide

diagnostic information in cardiomyopathy caused by primary metabolic abnormalities, such as mitochondrial disease, by detecting severe acidaemia. Peak VO_2 has been shown to be linked with outcome in ambulatory children with dilated cardiomyopathy; those with a value $\leq$ 62% of predicted have a 2-year increased rate of death or clinical deterioration and require urgent listing for heart transplantation [33]. Peak VO_2 is also helpful in determining the timing of heart transplantation.

All patients with hypertrophic cardiomyopathy should undergo an evaluation in which their risk of sudden death is assessed. The highest risk has been associated, among other factors, with abnormal blood pressure response during upright exercise that is attenuated or hypotensive, indicative of hemodynamic instability. Therefore, exercise testing is recommended on an annual basis in that condition.

Exercise testing is commonly used for the objective assessment of exercise capacity in pulmonary arterial hypertension. Peak VO_2 and peak systolic arterial pressure seem to have the best predictive value for outcome. While the results of the 6-minute walking test and metabolic exercise testing do correlate in pulmonary arterial hypertension, exercise testing has failed to confirm improvements observed with the walking test in clinical trials. Lack of standardization and insufficient expertise in performing exercise testing are usually considered to be the main reasons for this discrepancy.

The non-invasive follow-up of patients with a history of Kawasaki disease is not clearly defined. Usually, an exercise ECG is considered to be desirable at the time of final evaluation, 5 years after the onset of Kawasaki disease, in patients who have had no or transient dilatation of coronary arteries in the acute phase of the disease. For patients who have had coronary aneurysms at 30 days after the onset of Kawasaki disease, an exercise ECG should be performed at each follow-up visit at ages when it is feasible.

Exercise testing in arrhythmia

The most common reason for exercise testing in pediatric cardiology is to clarify exercise-related symptoms that have occurred either during or immediately after exercise. Exercise testing enables the identification and evaluation of arrhythmias in children.

Congenital long QT syndrome

Prolongation of the QT interval in repeated 12-lead ECGs is the hallmark of this syndrome. However, one-fifth to one-fourth of patients with confirmed long QT syndrome (LQTS) have a normal range QTc [34]. In some LQTS patients, exercise testing may reveal chronotropic incompetence, T wave alternans, ventricular tachyarrhythmias or paradoxical behavior of the QT interval under stress (increasing rather than decreasing). As measurement of the QT interval during exercise is difficult, the use of provocative tests for QT measurement in the recovery phase of exercise testing has been proposed to unmask LQTS patients with a normal resting ECG [35]. QTc is measured at 3 minutes of recovery and an increase of $\geq$ 30 ms is usually considered to be significant. The clinical use of this test in uncertain cases requires more extensive validation [36]. When testing for the efficacy of beta-blockade in LQTS patients, the goal of the evaluation is to determine if there is blunting of the heart rate response and suppression of dysrhythmia at maximal exercise.

Brugada syndrome

Brugada syndrome is definitively diagnosed when a type I ST-segment elevation is observed either spontaneously or after intravenous administration of a sodium channel-blocking agent in at least one right precordial lead [36]. However, many diseases and conditions can lead to a Brugada-like ECG abnormality. In asymptomatic subjects displaying a type I ECG, some findings are considered to support the diagnosis of Brugada syndrome [36]. Among them, we find attenuation of ST-segment elevation at peak of exercise stress test, followed by its appearance during the recovery phase [37,38]. However, in selected patients (usually with an SCN5A mutation), ST-segment elevation might become more evident during exercise [37].

Catecholaminergic polymorphic ventricular tachycardia

Exercise stress test is the most reliable way to establish the diagnosis of catecholaminergic polymorphic ventricular tachycardia (VT). This arrhythmia is diagnosed in young patients in the presence of a structurally normal heart, a normal resting ECG and unexplained exercise-induced bidirectional VT or polymorphic ventricular premature beats or VT [36]. When patients start exercising, monomorphic premature ventricular beats appear when the sinus rate exceeds an individual threshold rate (usually between 110 and 130 beats per minute), and may be followed, as the heart rate increases, by polymorphic premature ventricular beats and bidirectional or polymorphic VT (Figs. 1 and 2). When the test is discontinued, arrhythmias gradually disappear (Fig. 2). Exercise tests should be repeated periodically to find the adequate dosage of the beta-blocker medication and to ensure that the degree of sinus tachycardia that precedes the onset of arrhythmias is known, so that it can be avoided as much as possible in daily life. Moreover, the presence of couplets or more successive ventricular premature beats during exercise testing seems to be significantly associated with future arrhythmic events, suggesting intensification of treatment (flecainide, implantable cardiac defibrillator, left cardiac sympathetic denervation) in these patients [39].

Arrhythmogenic right ventricular dysplasia

Exercise-provoked arrhythmias may develop in arrhythmogenic right ventricular dysplasia. The typical arrhythmia is a monomorphic VT with a left bundle branch block pattern.

Other forms of ventricular tachycardia

The usefulness of exercise testing in patients with VT is variable, according to the cause of the tachycardia. In some forms, such as right ventricular outflow tract tachycardia in a normal heart, VT may be reproducibly induced during

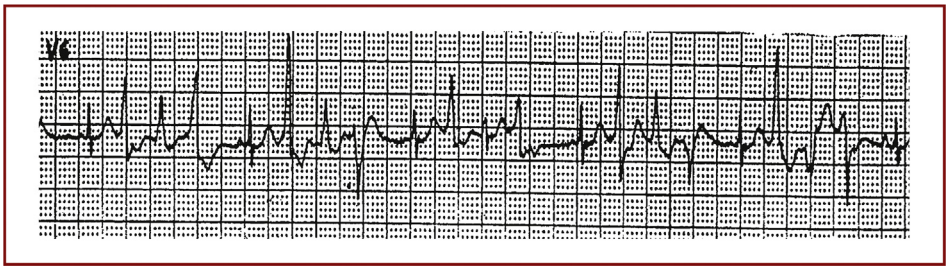

Figure 1. Exercise-induced polymorphic ventricular beats in catecholaminergic polymorphic ventricular tachycardia.

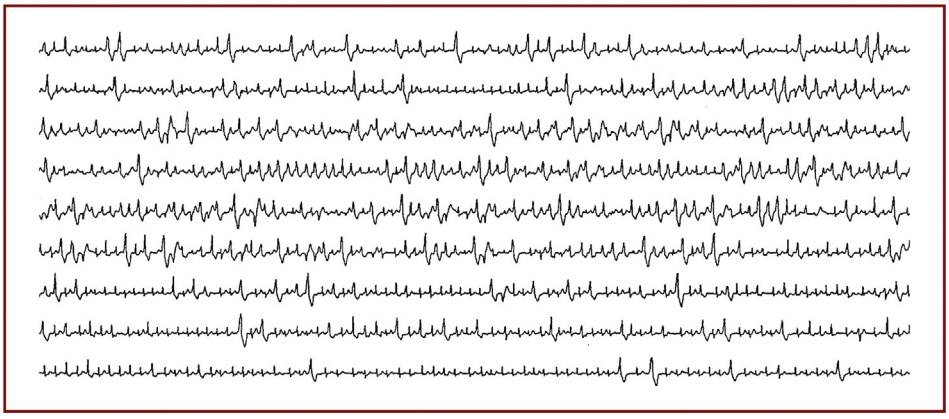

Figure 2. Typical sequence of exercise-induced ventricular arrhythmia in catecholaminergic polymorphic ventricular tachycardia.

exercise testing. However, in most forms, the reproducibility is limited.

Supraventricular tachycardia

Frequently, patients report the onset of supraventricular tachycardia (SVT) symptoms during exertion; exercise testing could therefore serve as an ideal non-invasive provocative test for SVT induction in those cases, and for evaluation of the adequacy of response to treatment in children with an SVT found during exercise before therapy. However, the diagnostic yield of exercise testing in patients with suspected SVT is limited, even among those with exercise-related events (Fig. 3). Nevertheless, exercise testing can still be useful in defining accessory pathway conduction characteristics among patients with ventricular pre-excitation. This evaluation is recommended to help gauge the risk of developing rapid ventricular response during atrial arrhythmias, especially atrial fibrillation, because some rare pediatric cases of sudden death have been reported as the possible first event of that abnormality. In children with a delta wave on a resting ECG, exercise testing may aid in the evaluation of the refractory period of the accessory pathway (Fig. 4). Abrupt loss of pre-excitation during exercise suggests a longer antegrade refractory period in the accessory pathway than in the atrioventricular node. However, this response may be difficult to recognise, because the adrenergic state speeds conduction in the atrioventricular node and therefore reduces the area of myocardium that is stimulated prematurely from the accessory pathway. Patients with total QRS normalization during exercise and no tachycardia symptoms require no electrophysiological study and may be released for the practice of physical activity.

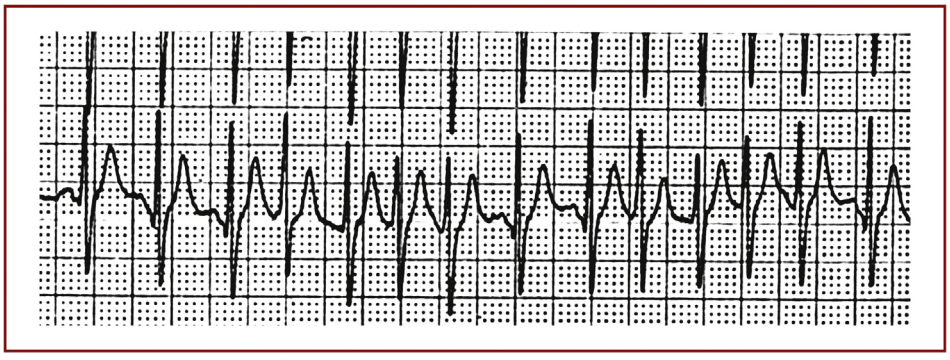

Figure 3. Postexercise atrioventricular nodal re-entry tachycardia.

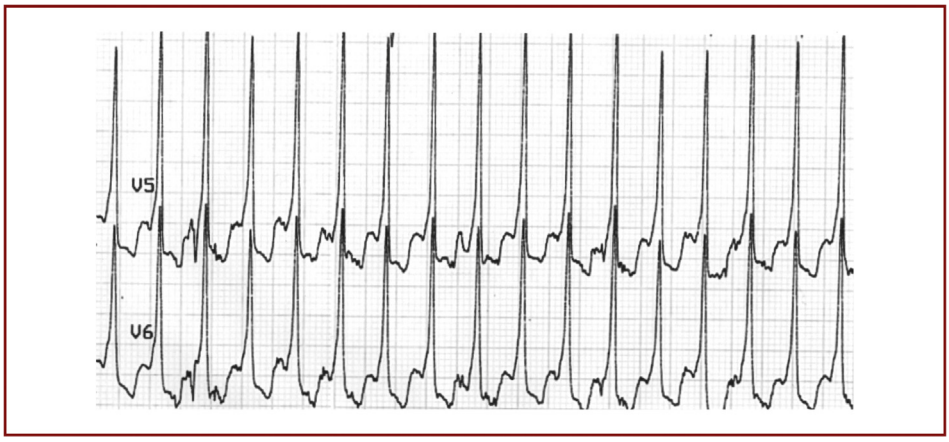

Figure 4. Delta wave persisting at the end of exercise testing (heart rate 201 beats per minute) in Wolff-Parkinson-White syndrome.

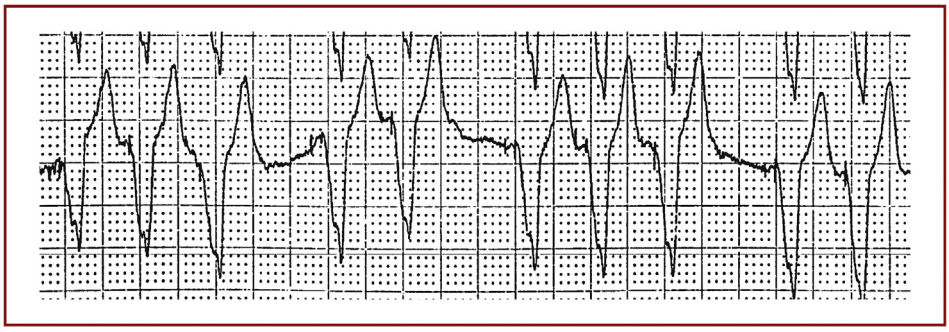

Figure 5. Exercise-related electronic Wenckebach phenomenon of a dual-chamber pacemaker.

Conduction disorders and pacing

Exercise testing may distinguish resting bradycardia with normal chronotropic response from sinus node dysfunction. The proposal has been made to obtain an objective evaluation of exercise tolerance in children with complete atrioventricular block before giving the indication for pacing. In patients with cardiac pacemakers, exercise testing may be required in the evaluation of the quality of atrial sensing and pacing rate responsiveness during exercise (Fig. 5), whereas, in patients with implantable cardiac defibrillators, it is useful to assess the risk of sinus rate crossover.

Conclusion

The acquisition and interpretation of exercise testing in children with cardiac disease present clinicians with some unique challenges, but the collected information provides unique insights into the patient's hemodynamic status and prognosis. Therefore, exercise testing plays a major role in the continuum of care for children with cardiac disease.

Disclosure of interest

The author declares that he has no conflicts of interest concerning this article.

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