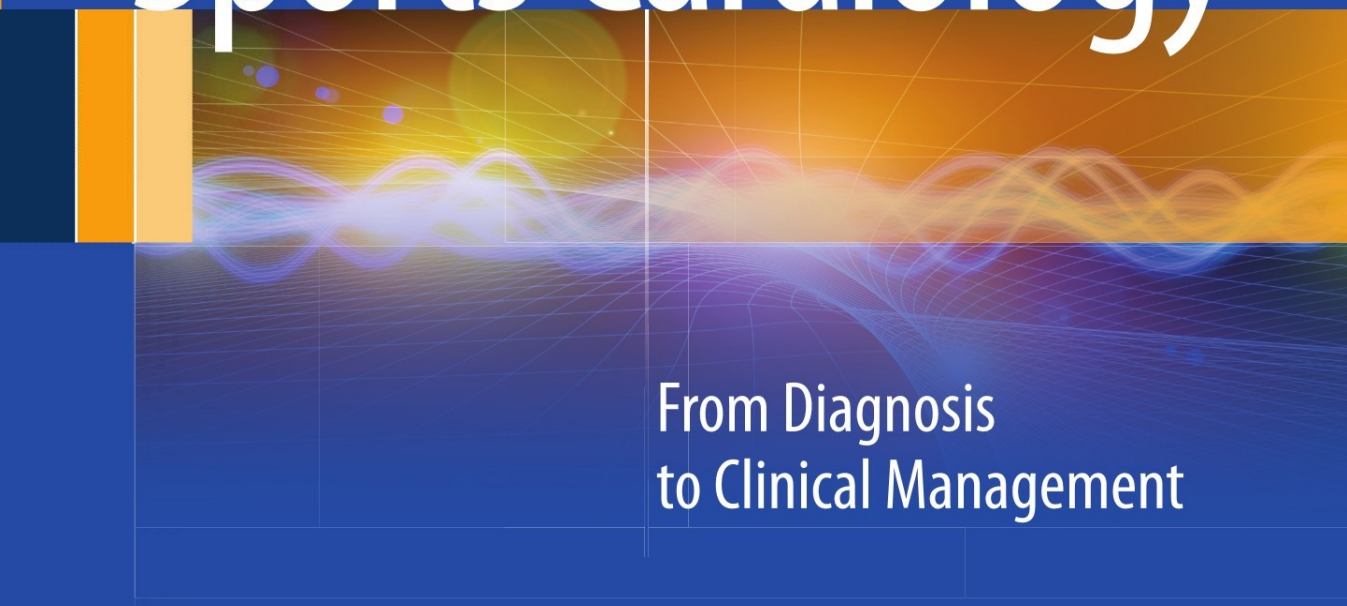
Massimo Fioranelli
Gaetano Frajese
Editors

Sports Cardiology



From Diagnosis to Clinical Management

Forewords by
Jacob Shani
Moussa Mansour

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I dedicate this book to all my colleagues, who are excellent cardiologists; their professional and personal commitment has enabled the development of the Mater Dei Heart Center:

Maria Bianchi, Caterina Bisceglia, Valentina Boccadamo, Fiorella Caranci, Giovanna Giubilato, Carlo Gonnella, Roberto Leo, Antonio Lucifero, Enrica Mariano, Augusto Mazzetti, Francesco Perna, Emiliano Pica, Maddalena Piro and Vittoria Rizzello

Massimo Fioranelli

Foreword

The field of sports cardiology gained notoriety over the last few years when several well-known athletes have succumbed to sudden cardiac death. The media attention to these events has inflamed the controversy of whether better screening would have prevented any of the deaths or whether the time has come for nations and health organization to agree on an universal approach.

Unfortunately there is no universal approach to screen athletes or the general population. There is no accepted method of screening people of different ages that want to participate in athletic activity. The challenges to the physicians who have to decide whether to approve or deny the participation and the right to compete are great.

The International Olympic Committee (IOC) has recommended, but not mandated, that all countries screen their athletes to minimize the risk of sudden death. According to the IOC, if the twelve lead ECG raises the suspicion of an underlining serious problem in an athlete, further testing should be performed. In many screening programs, the work up also includes an echocardiogram. By contrast, the American Heart Association, (AHA) and the American College of Cardiology (ACC) do not recommend the use of ECG for cardiovascular screening of athletes at any level. The Canadian Heart and Stroke Foundation also has no screening recommendations for athletes nor does the Canadian Academy of Sports and Exercise Medicine.

At the far end of the spectrum is the Italian model, in which individuals are not allowed to participate in minor sports unless they have had a family history taken, physical examination and twelve lead ECG. In the United States and Canada the sports medicine community had not embraced mandatory ECG screening because of concerns about cost, access, and risk of false positives, that leave some healthy kids on the side lines. Instead North American doctors typically rely on information from family history and physical exam to guide decisions.

In this volume the Editors managed to put together a textbook and a reference for physicians and people who have interest in sports medicine. To my knowledge there is no other textbooks such as this. All the issues that involve cardiac activity and sports from arrhythmia and sudden death to mechanical and hemodynamic issues are addressed.

This book is unique, because it is concise, accurate and extremely helpful in making decisions. Both the general practitioner as well as the cardiologist will find this textbook a useful resource and reference when they deal with

patients with or without cardiac issues, who want to participate in athletic activity. Through the collective efforts of all the Authors and the Editors, they were able to catalog the knowledge of this very important branch of medicine that impacts enormously the quality of life of the general populations, and athletes in particular.

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Foreword

Earlier this year and within the span of few weeks, two European football players, Fabrice Muamba and Piermario Morosini sustained sudden cardiac arrests, sending shock waves across the athletic and medical communities. These and other similar events over recent years have fueled an increased interest in sports cardiology. This field has been overlooked for years, which resulted in a poor understanding of the prevention and management of cardiac conditions among athletes.

This book, edited by Massimo Fioranelli and Gaetano Frajese, is a comprehensive practical guideline to sports cardiology. The areas covered include all aspects of this field, from history and physical examination to advanced imaging. The chapters are concise and clear, making this book an important tool for practicing cardiologists who will find themselves increasingly faced with situations requiring proficiency in the prevention, diagnosis and treatment of cardiac diseases in athletes.

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Preface

Physical activity and health are now inseparable terms; regular exercise, like a pharmacological intervention, is a powerful tool of prevention and treatment for many cardiovascular diseases. In addition, the psychophysical well-being that comes from sport drives many individuals, including the elderly, to initiate and continue the practice of many sports.

The athlete's cardiovascular adaptation is sometimes a thin line between physiology and pathology, and often makes it difficult to assess the individual risk.

Whenever an athlete dies on the playing fields, cardiologists are called upon to answer the usual questions: what was the cause of death, and why was not possible to prevent it?

This whole issue revolves around the professional competence of the modern cardiologist and the limitations of current screening procedures.

If this is an objective difficulty, amplified by the media's widespread reporting of just a few fatal accidents, there are also clinical settings with less media attention but which are not free from reflection or from medical and legal implications.

The constant evolution of knowledge in the field of cardiovascular pathophysiology and the stressful pace of the work force the clinical cardiologist and the sports physician to rapidly and continuously update their knowledge.

As with other specializations, and maybe more so, the modern cardiologist has a variety of skills that make it even more important to integrate news with everyday clinical practice; we therefore require tools that can assist doctors in making clinical decisions.

This book is developed for these requirements and is characterized on the one hand by a systematic review of indications for modern non-invasive and invasive diagnosis, and on the other hand on the clinical approach to most cardiovascular problems that affect athletes.

Particular care has been take to cover modern interventional therapies ranging from coronary artery interventions to issues related to closure of the patent foramen ovale, precautions to observe in patients with antiarrhythmic devices, and safety precautions for sporting activity in patients with congenital heart disease.

I hope that this book can help the modern sports physicians in the daily task of solving clinical problems.

Massimo Fioranelli

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Part I

Cadiovascular Diagnostics

Medical History and Physical Examination in Sports Medicine

1

Milena Leo, Francesco Perna, Roberto Leo and Massimo Fioranelli

1.1 Introduction

Medical history and physical examination are recognized worldwide as the milestone of athletes' preparticipation cardiovascular screening. They are defined as the systematic medical practice of evaluating large populations of athletes before sports participation in order to identify (or raise suspicion of) those abnormalities that might undergo disease progression or sudden cardiac death.

Although this screening process traditionally involves the evaluation of several organs, it mainly focuses on cardiovascular diseases and especially on sudden death, a tragic event bearing a considerable impact on the lay and medical communities, as well as on the general population, because athletes are usually young and considered the healthiest part of society [1].

Intense athletic training and competition act as a trigger to increase the risk of sudden cardiac death or disease progression in susceptible athletes with underlying heart disease (although quantification of that risk remains elusive). Most cardiac sudden deaths in young athletes occur during sports training or competition. Therefore, a relationship has been drawn between intense physical activity and arrhythmic sudden death. The risk of sudden death, however, appears to be unrelated to the level of athletic competition (i.e., high school, college, or professional). Indeed, early identification of relevant diseases may well prevent some instances of sudden death after temporary or permanent withdrawal from sports or targeted therapeutic interventions [2,3].

To date, there is universal agreement about the need to screen athletes for suspected heart disease before participation in competitive sport, but recommendations about the methods (European Society of Cardiology (ESC), International Olympic Committee (IOC), American Heart Association (AHA) preparticipation screening scientific statement) and the actual clinical practice vary greatly [4-6]. Noninvasive tests, such as electrocardiogram or echocardiogram, could raise the chances of detecting cardiovascular abnormalities, such as hypertrophic cardiomyopathy (HCM). However, according to some authors, such screening proposals would be hard to achieve and too expensive for most schools. This is further hampered by the fact that sudden cardiac death is an infrequent event in athletes (1:200,000) and that only a small proportion of those participating in team sports are at risk from misdiagnosed cardiovascular disease. The causes of sudden death during sporting activity are strictly related to the participants' age. Atherosclerotic coronary artery disease accounts for the vast majority of fatalities in adults (age > 35 years), while cardiomyopathies, such as HCM or arrhythmogenic right ventricular cardiomyopathy/dysplasia, have been consistently observed as the leading cause of cardiac arrest in younger athletes. Other, less common cardiovascular causes for sport-related sudden death in young athletes include premature atherosclerotic coronary artery disease, congenital coronary anomalies, myocarditis, dilated cardiomyopathy, mitral valve prolapse, conduction system diseases, Wolff-Parkinson-White (WPW) syndrome, ion channel

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diseases (long QT syndrome, catecholaminergic polymorphic ventricular tachycardia, Brugada syndrome), spontaneous aortic rupture complicating Marfan's syndrome or bicuspid aortic valve [7].

Therefore, personal and family history and physical examination may be good starting points in athletes' screening for the prevention of sudden cardiac death [8].

1.2 Family History

Most cases of non-traumatic sudden death in athletes are genetically determined diseases with an autosomal dominant pattern of inheritance, and therefore the importance of family history in identifying affected athletes should be emphasized.

According to the AHA statements, ESC recommendations and COCIS (Italian) guidelines for preparticipation screening [4,6,9], the main topics to be investigated are:

- premature, sudden and unexpected death (before the age of 50 due to heart disease, in at least 1 relative);
- disability from heart disease in a close relative (need for pacemaker or cardioverter-defibrillator implantation, cardiac surgery or transplantation, myocardial infarction before 55 years in men or 65 years in women);
- family history of hypertrophic or dilated cardiomyopathy, long-QT syndrome or other ion channel diseases, Marfan's syndrome, or clinically relevant arrhythmias.

1.3 Personal Medical History

Personal medical history is essential to define the athlete's cardiovascular risk and to guide the subsequent physical examination.

For this purpose, the most important topics to be investigated are:

- presence of ischemic risk factors such as diabetes mellitus, hypertension, dyslipidemia, smoking;
- history of cardiovascular diseases, including transient ischemic attacks, stroke, peripheral vascular disease and peripheral edema;
- history of previous accidents, especially with chest contusion;
- medications or drug abuse;
- prior identification of heart murmurs or elevated blood pressure values;
- typical cardiac symptoms, such as chest pain, tightness or discomfort, dyspnea, palpitations and syncope, as described below in detail.

It is important to correlate symptoms to the patient's baseline level of physical activity in order to better assess his/her real condition and to identify the best type of management [4,6,9].

1.3.1 Chest Pain

Chest pain is a very common symptom, but is difficult to evaluate. Many diseases can lead to chest pain:

- cardiac diseases: stable angina, acute coronary syndromes (ACS), coronary vasospasm (Prinzmetal's angina), hypertrophic cardiomyopathy, aortic stenosis, arrhythmias, aortic dissection, mitral valve disease, pericarditis;
- respiratory diseases: pneumothorax, pulmonary embolism, pneumonia, pleurisy, lung cancer;
- musculoskeletal diseases: costochondritis, Tietze's syndrome, trauma, rib pain (including fracture, bone metastases, osteoporosis), radicular pain, nonspecific musculoskeletal pain (e.g. fibromyalgia);
- breast disease;
- gastrointestinal diseases: gastro-oesophageal reflux disease, oesophageal rupture, oesophageal spasm, peptic ulcer disease, cholecystitis, pancreatitis, gastritis;
- skin diseases: herpes zoster infection;
- psychiatric diseases: e.g. anxiety, depression, panic attack;
- others: sickle cell crisis, diabetic mononeuritis, tabes dorsalis.

The nature of pain (weight, burning, stabbing, discomfort), as well as its location (middle of the chest, upper abdomen, neck, jaw, left arm, left shoulder), radiation (neck, jaw, back, left or right arm), frequency, intensity, duration, aggravating and relieving factors, correlation with exercise, associated symptoms (e.g. nausea and/or vomiting,

sweating, dizziness, and palpitations) are useful features to observe in order to clarify the underlying cause.

Common causes of chest pain in athletes are:

- panic attacks, especially in women, typically associated with palpitations, tingling limbs, dizziness, shortness of breath, sometimes before or soon after stressful activity. A positive screening for previous panic attacks is highly sensitive to diagnosis of actual panic disorder but should not preclude a complete cardiac examination;
- gastro-oesophageal reflux disease, often due to excessive swallowing of air during exercise, or consumption of fizzy drinks or meals that are rich in fat;
- myopericarditis: to be suspected in the case of recent fever or flu, presence of burning, sometimes oppressive, chest pain, which varies with a change in body position or breathing;
- cocaine abuse;
- pleuropericarditis, typically involving posterior, stabbing chest pain, which varies depending on breathing, and is associated with fever or cough;
- spontaneous pneumothorax, typically occurring during exercise with acute and intense dyspnea, common in tall athletes, with marfanoid habitus;
- ischemic heart disease, typically induced by exercise and attenuated while resting, and often associated with icy sweating and general discomfort. It should be suspected in particular in adult athletes.

Less common, but not less important causes of chest pain in athletes are hypertrophic cardiomyopathy, aortic stenosis, coronary artery abnormalities such as myocardial bridge: all those diseases present with effort-related chest pain, dyspnea, or recurrent syncope. In these cases physical examination and additional tests, such as ECG and echocardiogram, are crucial for correct diagnosis.

1.3.2 Dyspnea

Dyspnea is the subjective discomfort of experiencing one's own breathing. It is a normal symptom on heavy exertion, even in well trained athletes, but it can suggest underlying pathological conditions if it occurs at rest or during ordinary physical activity.

Many diseases can cause it: respiratory diseases such as asthma, pneumothorax, chronic obstructive pulmonary disease, pneumonia, interstitial lung disease, cardiac diseases such as ischemia, congestive heart failure, psychogenic causes, neurological diseases. Treatment typically depends on the underlying cause.

Depending on how it was triggered, it may be classified as acute dyspnea, i.e. severe shortness of breath lasting several minutes or hours, or chronic dyspnea, which lasts for several weeks or months.

It may be further distinguished as inspiratory, expiratory or mixed dyspnea, according to the corresponding phase of the respiratory cycle.

According to the *New York Heart Association* (NYHA), dyspnea may be classified into four categories on the basis of the extent of the subject's functional limitation:

- class I: the subject has no dyspnea;
- class II: dyspnea appears during moderate exertion, with only slight limitation during ordinary activity;
- class III: dyspnea is associated with mild exertion, with marked limitation even during less-than-ordinary activity so that the patient feels comfortable only at rest;
- class IV: dyspnea is also present at rest, with severe limitation of daily activities.

Common causes of dyspnea in athletes are:

- panic attacks: dyspnea typically occurs at rest in this case, but not during exercise;
- exercise asthma, with prevalent expiratory dyspnea due to bronchospasm; it is not related to the level of physical activity; predisposing factors are cold, wet weather, and family or personal history of allergies;
- exercise laryngospasm, which is common in women, and is typically resolved by reassurance and anxiolytics;
- spontaneous pneumothorax, which is more common in tall athletes, with marfanoid habitus, typically occurring together with acute and intense dyspnea during exercise.

Other important causes, even though less frequent, are hypertrophic cardiomyopathy, aortic

stenosis, ischemic heart disease (especially in adult athletes), and all other cardiac diseases that lead to congestive heart failure.

1.3.3 Palpitations

Palpitations involve an unpleasant awareness of one's own heartbeat. When describing palpitations, patients use terms such as skipping, racing, fluttering, pounding or stopping of the heart. Palpitations may result from an irregular heartbeat, from rapid acceleration or slowing of the heart, or from increased forcefulness of cardiac contraction. Such perceptions also depend on how patients experience their own body sensations. They are a common symptom, both in people with healthy and diseased hearts.

On the basis of the clinical presentation (means of onset and interruption, duration, frequency of episodes, correlation with physical activity, associated symptoms) it is possible to distinguish between three forms of palpitations: anxiety palpitations, extrasystolic palpitations and palpitations due to prolonged supraventricular or ventricular arrhythmias.

Anxiety palpitations are frequent in athletes, especially in women, before and during competitions: they are typically described as a form of agony, associated with a slightly higher than normal heart rate, with a gradual start and end, and no association with any organic disease.

Extrasystolic palpitations are described as an intermittent perception of a missing heartbeat or heart drop that lasts a few seconds but can recur more or less frequently in a day. Both premature supraventricular and ventricular complexes can cause this symptom. Extrasystolic palpitations are usually a benign condition, but, if frequent, they can be related to cardiac diseases such as ischemic heart disease, hypertrophic cardiomyopathy, arrhythmogenic right ventricular disease, Brugada syndrome, long QT syndrome or mitral valve prolapse. Additional tests may therefore be required.

Tachyarrhythmias other than premature complexes generally cause a prolonged feeling of accelerated, regular or irregular heartbeat that can be well tolerated or rather associated with dyspnea, dizziness, light-headedness and chest pain. The onset and end of tachyarrhythmia can be more or less abrupt. Moreover, tachyarrhythmias can be related to physical activity or they can take place at rest, and they can arise both in normal hearts (typically atrioventricular nodal re-entrant tachycardia, WPW syndrome, some cases of atrial fibrillation, or right ventricular outflow tract tachycardia) and in diseased hearts (ischemic heart disease, hypertrophic cardiomyopathy, other cardiomyopathies, arrhythmogenic right ventricular disease, Brugada syndrome, QT long syndrome or mitral valve prolapse). Furthermore, gastrointestinal diseases such as hiatal hernia, thyroid dysfunction, anemic status, coffee and other stimulant abuse, or smoking may be predisposing factors for arrhythmias. In such cases additional tests are required as well.

1.3.4 Syncope and Presyncope

Syncope is a transient loss of consciousness and postural tone characterized by rapid onset, short duration and spontaneous recovery due to global cerebral hypoperfusion that is most often the result of hypotension. Many forms of syncope are preceded by a prodromal state, known as *presyncope*, characterized by lightheadedness, dizziness, temporary loss of vision or hearing, pain, nausea, abdominal discomfort, weakness, sweating, palpitations and other phenomena.

Syncope is a common condition in both the general population and athletes.

Neurally-mediated syncope is the most common type of syncope observed among athletes. It is characterized by peripheral vasodilation, decreased blood pressure, along with bradycardia, and is caused by an increase in the parasympathetic tone and concomitant inhibition of sympathetic output. It generally occurs in susceptible people under physically stressful conditions (strenuous effort, dehydration, hunger or hypoglycemia).

However, cardiogenic syncope must be investigated. It is due to left ventricular ejection obstruction (such as in aortic stenosis or obstructive hypertrophic cardiomyopathy), arrhythmias, pulmonary embolism or hypertension, aortic dissection, con-

genital coronary anomalies.

Differential diagnosis is important in order to identify and treat the underlying cause.

Furthermore, regardless of its cause, syncope may result in physical injury such as head trauma: it is therefore important to clarify the circumstances of onset and the association with physical activity in order to avoid accidents during exertion.

1.4 Physical Examination

Important information about athletes' preparticipation cardiovascular evaluation comes from a careful and precise physical examination. It includes a general inspection of the patient, measurement of arterial blood pressure in both arms and one or both lower extremities, examination of the arterial pulse and dynamic cardiac auscultation. Based on the results of this inexpensive evaluation, a definite diagnosis can often be made. Selected non-invasive and invasive tests are therefore performed only when appropriate.

1.4.1 General Inspection

Visual inspection of the athlete may provide important information.

First of all, it gives the physician an idea about the general health status of the athlete, his/her physical development and habitus.

Furthermore, careful observation of the athlete may provide clues for the diagnosis of underlying heart diseases.

Xanthoma, xanthelasma or arcus senilis may be signs of familial hyperlipidemia and therefore of premature coronary artery disease.

Edema, cyanosis and breathlessness, though rare in athletes, may suggest heart failure.

Observation of the skin, thorax and extremities may provide clues for the diagnosis of congenital heart diseases, which are often unknown in young athletes.

Bilateral prominence of the anterior chest with bulging of the upper two-thirds of the sternum is common in children with a large ventricular septal defects; a unilateral bulge at the fourth and fifth intercostal spaces at the lower left sternal border is found in adults with ventricular septal defect.

Furthermore, clubbed fingers or cyanosis of the skin or nails suggest congenital heart disease with right-to-left blood shunt.

Underdeveloped musculature in the lower extremities compared to the upper extremities can occur in patients suffering from coarctation of the aorta.

Among those syndromes associated with congenital heart disease, special attention must be paid to *Marfan's syndrome*. It is an autosomal dominant disorder suggested by skeletal features such as increased height, long fingers (*arachnodactyly*), lax joints, kyphoscoliosis, pectus excavatum or carinatum, an elongated face, high-arched palate and flat feet, bilateral subluxation of the lens, severe myopia and blue sclera. Patients with Marfan's syndrome usually have mitral valve prolapse, mitral regurgitation, calcified mitral annulus, mitral chord rupture, dilated aortic root, prolapse of the aortic cusps or aortic dissection.

Patients with *Ehlers-Danlos syndrome* have hyperextensible joints and hyperelastic and friable skin often associated with arterial dilation and rupture, aortic regurgitation or mitral valve prolapse.

1.4.2 Arterial Blood Pressure Measurement

Arterial blood pressure measurement is fundamental because the prevalence of hypertension is growing among athletes, thus carrying a higher cardiovascular risk.

When measuring blood pressure, care should be taken to:

- allow the athlete to sit quietly for several minutes;
- carry out at least two measurements 1-2 minutes apart;
- perform measurements in both arms to detect possible differences due to supravalvular aortic stenosis or subclavian steal syndrome;
- carry out measurements in both arms and at least one leg to detect possible differences caused by peripheral vascular disease or, especially in

younger athletes, coarctation of the aorta.

For practical reasons, hypertensive patients are categorized according to precise thresholds as indicated by ESC and AHA guidelines. However, the real threshold for defining hypertension must be considered flexibly, based on the overall cardiovascular risk of each individual.

It is important to remember that many factors may contribute to variations in blood pressure during daily activities: body posture, status of cerebral or gastrointestinal activity, emotional or painful stimuli, environmental factors such as temperature and noise levels, smoking, coffee, alcohol and other drugs with direct or neurally mediated vasomotor properties.

Furthermore, isotonic exercise in both the supine and upright positions produces a moderate increase in blood pressure; sustained isometric muscular contractions produce an abrupt increase in systolic, mean and diastolic blood pressure that depends on the strength of the contraction. This may have important implications for physical activity restrictions in hypertensive patients.

1.4.3 Arterial Pulses Palpation

Palpation of peripheral arterial pulses is a basic and important element of an athlete's physical examination for several reasons.

First of all, palpation of the radial pulse allows for quick evaluation of the patient's heart rate and rhythm.

Secondly, simultaneous palpation of both the radial pulse and femoral pulse is useful as screening for coarctation of the aorta. If the femoral pulse is reduced or delayed with respect to the radial pulse, further evaluation is needed.

Moreover, evaluation of arterial pulse characteristics may indicate the presence of important heart diseases. A normal arterial pulse has a smooth rapid upstroke, a smooth peak, and a more gradual downstroke. Hyperkinetic arterial pulses are common in athletes with marked bradycardia and an extremely large stroke volume. In contrast, hypokinetic arterial pulse is less frequent and indicates a condition of diminished stroke volume, for example hypovolemia, left ventricular failure, mitral or aortic valve stenosis. The finding of a *bisferiens* pulse, with a waveform characterized by two positive waves during systole, suggests the diagnosis of hypertrophic cardiomyopathy or, less frequently, aortic regurgitation with or without aortic stenosis. The *parvus et tardus* pulse, a small pulse with a delayed systolic peak, is typical of aortic stenosis. A *pulsus alternans*, characterized by regular alternation of the systolic peak in pressure pulses, indicates severe depression of left ventricular performance. Conditions such as asthma, obstructive airways disease and cardiac tamponade are often associated with a *paradoxical pulse*, defined as a marked decrease in pulse amplitude or a decrease in systolic arterial pressure by more than 10 mmHg during normal quiet inspiration.

1.4.4 Cardiac Auscultation

Cardiac assessment should be performed in a quiet setting to facilitate auscultation. The assessment should include a brief visual inspection of the precordium to search for asymmetry and abnormal impulses. Precordial palpation can detect thrills, abnormal apical impulse location and parasternal heave.

The physician should then carefully auscultate for heart sounds, heart murmurs and extracardiac sounds.

The four *primary areas of cardiac auscultation* are: 1) the primary and secondary aortic areas in the second right interspace and the third left interspace adjacent to the sternum, respectively; 2) the pulmonary area in the second left interspace; 3) the tricuspid area in the fourth and fifth interspaces adjacent to the left sternal border; 4) the mitral area at the cardiac apex. Furthermore, in case of heart murmurs, cardiac auscultation should be enlarged to additional areas (such as the neck, posterior thorax, right sternal border and epigastrium) to which it typically radiates.

It is important to adopt a systematic approach to auscultation: in each area one should first listen to the heart sounds, then heart murmurs during systole and diastole respectively, trying to record the time of appearance, the pitch and other characteristics.

The patient should be auscultated in the left lat-

eral decubitus position and, when possible, also while standing, squatting and during and after the *Valsalva manoeuvre* and the *handgrip manoeuvre*, in order to assess sound and murmur changes according to preloading and afterloading variations.

Moreover, if there are diagnostic clues in the medical history as well as in the examination of arterial, venous and cardiac pulzations, the auscultatory counterparts should be pursued diligently.

1.4.4.1 Heart Sounds

The heart cycle is physiologically marked by two (or three) heart sounds; a fourth heart sound may be present in pathological conditions.

The *first heart sound (S1)* is produced by the closure of the mitral valve (M1) followed after only 20 to 30 ms by the closure of the tricuspid valve (T1), so that at the apex in a normal subject only a single sound is usually heard. Wide splitting of S1 because of a delay in T1 can be a sign of *Ebstein's anomaly* of the tricuspid valve, often misdiagnosed in children, right bundle branch block, or ectopic beats originating from the left ventricle. Similarly, ectopic beats originating from the right ventricle, mitral valve stenosis, or left atrial mixoma causing mitral obstruction, will produce reversed splitting of S1 (T1, M1) due to a delay in mitral valve closure and left ventricular contraction. S1 intensity is higher in the case of tachycardia and lower in the case of bradycardia or I grade atrioventricular block, which is often found in athletes.

The *second heart sound (S2)* involves closure of the aortic valve (A2) followed by closure of the pulmonary valve (P2). Inspiration causes increased blood flow into the pulmonary vascular bed, delaying the closure of the pulmonary valve slightly. This inspiratory widening of S2 (*physiological split*) is generally a reassuring sign during the athlete's cardiac examination. A *fixed split* of S2 (because of a P2 delay over 40 ms during both inspiration and expiration) can be a sign of congenital heart disease with pulmonary hyperflow such as an atrial septal defect: in this case cardiac auscultation in the standing position is useful because it can modify S2 by reducing venous blood return. A *paradoxical split* (i.e. narrowing on inspiration) can be a sign of severe aortic stenosis, hypertrophic cardiomyopathy or left bundle branch block.

S1 is separated from S2 by a small pause corresponding to the systole, while S2 is separated from S1 by a large pause corresponding to the diastole.

The *third heart sound (S3)* is related to the early diastolic filling of the ventricles. It is a common benign event in children, adolescents and young adults, but it is rarely present in adults aged over 40. It is also common in endurance athletes with left ventricular dilation and bradycardia. S3 can be heard at the apex in the left lateral decubitus position soon after S2; it generally disappears during tachycardia and while standing.

The *forth heart sound (S4)* is related to the late diastolic filling of the ventricles due to atrial contraction. It is always an abnormal finding: it may be heard in the presence of reduced ventricular compliance (such as severe ventricular hypertrophy for aortic stenosis, hypertrophic or hypertensive cardiomiopathy, or ischemic heart disease) or excessively rapid late diastolic filling (secondary to hyperkinetic states or acute atrioventricular valve incompetence) or arrhythmias such as heart block. S4 is best heard at the apex impulse with the patient turned in the left lateral position, occurring shortly before S1.

Other pathological heart sounds are non-ejection clicks and opening snaps.

Non-ejection clicks are mid-systolic sounds produced by prolapse of the mitral or tricuspid valve and are often associated with a systolic regurgitant murmur. The sound has a sharp, high-frequency clicking quality and may be confined to the apex or widely transmitted on the precordium. There may be a single mid-systolic click, or multiple clicks, probably as a result of different leaflets prolapsing in different times. The timing of the click varies considerably with postural changes and this may be helpful in differentiating non-ejection clicks from early ejection sounds, a split S2 or a S3. In general, manoeuvres that decrease LV volume such as sitting or standing or the strain of the Valsalva manoeuvre move the click closer to S1, while manoeuvres that increase LV volume move the click towards S2.

Opening snaps are crisp, sharp sounds typically produced by the opening of a thickened and deformed stenotic atrioventricular valve. It can be

heard in the mid-precordial location, usually most clearly in the area extending from the left sternal border to the apex. It immediately precedes the diastolic rumble of valvular stenosis.

1.4.4.2 Heart Murmurs and Extra-cardiac Sounds

Cardiac murmurs are prolonged series of auditory vibrations produced by blood turbulence through cardiac structures or great vessels because of a high flow rate, forward flow through a constricted or irregular orifice, backward flow through an incompetent valve, septal defects or patent ductus arteriosus.

Useful features to examine in order to characterize heart murmurs are:

- the *timing* in the cardiac cycle, in order to distinguish between systolic (falling between S1 and S2), diastolic (falling between S2 and S1) or continuous murmurs: there is seldom any difficulty distinguishing between systole and diastole since systole is considerably shorter with normal heart rates; in the case of rapid heart rates, simultaneous palpation of the lower right carotid artery or identification of second heart sound, the louder sounds at the base are useful in properly locating murmurs in the cardiac cycle;
- the intensity (*loudness*), which is related to the velocity of blood flow across the area of murmur production and to the transmission characteristics of the tissue between the source of the murmur and the stethoscope. For systolic murmurs, the loudness is generally graded from 1 to 6 as described by Freeman and Levine (Table 1.1);
- the frequency (*pitch*), which is directly related to the flow velocity and to the pressure head that drives the blood across the murmur-producing area: this is categorized as high, medium or low;
- the configuration (*shape*), which is determined by the intensity over time; it enables crescendo, decrescendo, crescendo-decrescendo (diamond-shaped) or plateau murmurs to be defined;
- the *quality*, which is described as, for example, blowing, harsh, rumbling or musical;
- the *location* and sites of maximum intensity in the primary cardiac areas, radiation due to the direction of blood flow in other thoracic areas (right of the sternum, epigastrium, posterior thorax or neck).

Table 1.1 Classification of the intensity of heart murmurs as described by Freedman and Levine

Intensity	Characteristics
Grade 1	Just audible with a good stethoscope in a quiet room
Grade 2	Quiet but readily audible with a stethoscope
Grade 3	Easily heard with a stethoscope
Grade 4	A loud, obvious murmur with a palpable thrill
Grade 5	Very loud, heard not only over the pericardium but also elsewhere in the body
Grade 6	Heard with the stethoscope off chest

Many cardiac diseases can produce murmurs: detailed analysis of murmur characteristics is a precious tool for differential diagnosis. The most common cardiac conditions associated with heart murmurs in athletes, especially adults, are reported in Table 1.2.

Moreover, murmurs may occur in the absence of physiological and structural abnormalities in the cardiovascular system when, in early systole, peak flow velocity across the left or right ventricle outflow tract exceeds the murmur threshold, the so-called *innocent murmurs*. They are quite common in young athletes. These murmurs are rigorously systolic, of short duration, almost always less than grade 3 in intensity and vary considerably from one examination to another, with the body position and the level of physical activity. They are best heard at the left sternal border on the third or fourth interspace and are not associated with a thrill or with radiation to the carotid arteries or axillae. Hyperkinetic states such as fever or hyperthyroidism, may increase their intensity. For a murmur to be considered innocent, the examination of the cardiovascular system, and if possible an echocardiogram, should reveal no abnormalities.

Among *extracardiac sounds*, pericardial friction sounds may be heard. They are produced by pericardial leaflets rubbing each other due to inflammation of the pericardial sac. These friction sounds are very high pitched, leathery and scratchy in nature. They seem close to the heart and are best heard with the patient leaning forward or in the knee-chest position, holding breath after forced expiration. The pericardial rub may have three

Table 1.2 Characteristics of heart murmurs in various heart diseases

Disease	Timing	Quality	Shape	Location	Radiation	Other findings
Aortic stenosis	Mid-systolic	Harsh	Diamond	1^{st}-2^{nd} right intercostal spaces	Carotid arteries and cardiac apex (Gallivardin phenomenon)	- intensity not correlated to stenosis gravity and increased by increased cardiac output - single S2 or paradoxical splitting of S2 (A2 is absent or late) - S4 (S3) often heard - *parvus et tardus* pulse - systolic ejection click often precedes the murmur
Hypertrophic cardiomyopathy	Mid-systolic	Harsh	Diamond	Lower left sternal edge	Apex, not carotid arteries	- intensity related to intraventricular gradient (increased by decrease in LV preload and afterload or increase in LV contractility): useful stand-squat-stand position and leg raising manoeuvres - normal S2 - S4 always present - bisferiens pulse
Mitral regurgitation	Holosystolic (mid-late-systolic in acute MRs)	Blowing	Plateau	Apex	Axilla	- intensity is not influenced by respiration - short rumble - loud S3 from LV
Tricuspid regurgitation	Holosystolic (mid-late-systolic in acute TRs)	Soft	Plateau	Lower left sternal border	-	- JVP shows a prominent v wave with a rapid y descent - intensity increases during inspiration (Carvallo's sign) - short rumble - loud S3 from RV
Ventricular septal defect	Systolic	Harsh	Plateau	4^{th}, 5^{th} and 6^{th} intercostal spaces along the sternal border	-	- intensity not influenced by respiration - wide physiological splitting of S2 - S4 often present in case of a large shunt

(cont. →)

Table 1.2 Characteristics of heart murmurs in various heart diseases *(continued)*

Disease	Timing	Quality	Shape	Location	Radiation	Other findings
Mitral stenosis	Diastolic	Rumble	Decrescendo-crescendo	Apex	-	- poor correlation between the intensity of the murmur and the severity of the obstruction - loud S1 - mitral opening snap often precedes the murmur
Aortic regurgitation	Diastolic	Blowing	Decrescendo	1^{st}-2^{nd} right interspace (in case of aortic dilatation) 3^{rd}-4^{th} left interspace (in case of valvular disease)	Apex	- intensity increased by handgrip - Austin-Flint murmur (late-diastolic rumble due to partial mitral valve closure induced by increased LVEDP) often heard in severe AR - S3 often present - Corrigan's pulse (rapid rise and quick collapse) - Musset's sign (bobbing of the head with each heartbeat) - Traube's sign (gunshot sound heard over the femoral artery)
Atrial septal defect	Mid-systolic	Scratchy	Diamond	Left upper sternal border	-	- fixed splitting of S2 - low to medium pitched early diastolic murmur over the lower sternal border (because of increased diastolic flow across the tricuspid valve) often present in case of large shunts - cyanosis and clubbing often present in case of right-to-left shunting

components during the intervals of the cardiac cycle when the heart has the greatest excursions within the pericardial sac: at the time of atrial systole, at the time of ventricular contraction and during rapid early diastolic filling. Pericardial friction sounds are common in cases of pericarditis, a common cause of chest pain and dyspnea in young athletes after seasonal influenza [10-12].

References

1. Maron BJ (2003) Sudden death in young athletes. N Engl J Med 349:1064–1075
2. Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO (1996) Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. JAMA 276:199–204
3. Corrado D, Basso C, Pavei A, Michieli P, Schiavon M, Thiene G (2006) Trends in sudden cardiovascular death in young competitive athletes after implementation of a preparticipation screening program. JAMA 296:1593–1601
4. Corrado D, Pelliccia A, Bjørnstad HH, Vanhees L, Biffi A, Borjesson M et al (2005) Cardiovascular preparticipation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol: consensus statement of the Study Group of Sport Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Eur Heart J 26:516 –524
5. IOC Medical Commission, International Olympic Committee (2006) Sudden Cardiovascular Death in Sport: Lausanne Recommendations: Preparticipation Cardiovascular Screening. European Journal of Cardiovascular Prevention & Rehabilitation 13:859–875
6. Maron B, Thompson P, Ackerman M, Balady G, Berger S, Cohen D et al (2007) Recommendations and Considerations Related to Preparticipation Screening for Cardiovascular Abnormalities in Competitive Athletes: 2007 AHA Scientific Statement Update Circulation 115:1643–1655
7. Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO (2009) Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980–2006. Circulation 119:1085–1092
8. Giese EA, O'Connor FG, Brennan FH, Depenbrock PJ, Oriscello RG (2007) The athletic preparticipation evaluation: cardiovascular assessment. Am Fam Physician 1;75(7):1008–14
9. Santilli G, Casasco M, et al, Comitato Organizzativo Cardiologico per l'Idoneità allo Sport (COCIS) (2010) Protocolli cardiologici per il giudizio di idoneità allo sport agonistico. Medicina dello Sport Vol 63:1–137
10. Fuster V, Wayne Alexander R, O'Rourke R (2010) Hurst's The Heart 13th edition. McGraw-Hill, New York
11. Brukner P, Khan K (2010) Clinical sports medicine. McGraw-Hill, New York
12. Bell C (2008) Cardiovascular Physiology in exercise and sport. Churchill Livingstone, Philadelphia

Electrocardiographic Signal Analysis

2

Francesco Perna

2.1 ECG Analysis

Despite the well-known benefits of regular physical exercise on the cardiovascular system, there is a small population of young athletes (aged under 35 years) who carry an elevated risk of sudden cardiac death (SCD) during exercise. The annual incidence of SCD in young athletes is about 0.5/100,000 per year in the United States and 2.1/100,000 in Italy, and it occurs in more than 90% of cases during or immediately after physical exercise. Sudden deaths are mostly due to cardiac abnormalities that might be identified, provided that appropriate screening of subjects who are candidate to agonistic sports is carried out. Such a psychologically overwhelming problem as the death of a young athlete during a sport competition led to the development of an effective cardiovascular screening program in Italy, which achieved a 89% reduction of the incidence of SCD over 26 years [1]. On the other hand, there are currently no diagnostic tools that can definitely recognize subjects who are potentially at risk of SCD, especially among people with an apparently normal heart. The aim of this chapter is to describe the main instrumental examinations that are used for non-invasive cardiovascular diagnosis in the athletes, and some non-invasive arrhythmic risk markers that are used in cardiology.

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2.2 Electrocardiogram

Twelve-lead electrocardiogram (ECG) is currently included, along with the medical history and a physical examination, in the preparticipation cardiovascular screening protocol that is used in Italy. It is worth noting that various potentially life-threatening heart diseases can be diagnosed or at least suspected by surface ECG analysis. Therefore, several consensus groups strongly recommend inclusion of ECG in the screening programs.

2.2.1 Cardiomyopathies

ECG is usually abnormal in subjects with cardiomyopathies, in particular those affected by *hypertrophic cardiomyopathy* (particularly with the obstructive form of the disease) (Fig. 2.1). The most common ECG abnormalities for this disease are:

- signs of left ventricular hypertrophy, in particular Sokolow-Lyon index (R in V5 + S in V1) greater than 35 mm;
- ventricular repolarization abnormalities with signs of ventricular overload (ST segment depression, T wave inversion, or giant negative T waves in the apical variant of the disease);
- left axis deviation;
- pathological (narrow but deep) Q waves in the lateral (I, aVL, V5, V6) or sometimes inferior (II, III, aVF) leads.

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is often suspected when the surface ECG shows:

- inverted T waves in V1-V3 or beyond in indi-

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viduals ≥ 14 years of age in the absence of right bundle branch block;
- epsilon wave (reproducible low-amplitude signals between end of the QRS complex to the onset of the T wave) in the right precordial leads (V1 to V3);
- non-sustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis.

T wave inversion in the right precordial leads is common in children aged under 14 years, so this feature is not specific enough to diagnose ARVC in this subgroup. Additional minor criteria to be recognized in ECG, SAECG and Holter ECG, along with non-ECG criteria, can be found in the new task force consensus document [2].

ECG is almost always pathological in *dilated cardiomyopathy*, even though such abnormalities are not specific in the majority of cases; the most common ECG features in this disease are left ventricular hypertrophy, low QRS voltages, left bundle branch block, supraventricular or ventricular premature beats, repolarization abnormalities and non-sinus rhythm.

Myocarditis accounts for 5% of cases of SCD in athletes (up to 20% among young military recruits). There are no specific ECG anomalies in acute myocarditis, and it can sometimes mimic acute myocardial ischemia. The most common traits are: ST segment elevation, T wave inversion, widespread ST segment depression and pathological Q waves.

2.2.2 Ion Channel Diseases

Congenital long QT syndrome (LQTS) is a potentially life-threatening pathological condition that could be identified or suspected by ECG. It accounts for about 1-2% of cases of SCD in athletes, and the underlying arrhythmia is often a polymorphous ventricular tachycardia. This pathology is characterized by a prolonged QT interval on surface ECG. The QT interval should be estimated as a mean value derived from 3-5 cardiac cycles, usually in leads II, V5 and V6, and is measured from the beginning of the earliest onset of the QRS complex to the end of the T-wave. Since the QT interval inversely correlates to the heart rate, it is usually corrected for the heart rate itself using the Bazett formula: $QTc + QT/\sqrt{RR}$. This formula is deemed to be acceptable when the heart rate is between 60 and 100 beats per minute (bpm). As a rule, QTc interval is deemed as pathological if it is greater than 450 ms in men and 470 ms in women. T wave morphology may be altered in patients with different genotypes of congenital LQTS: LQTS1 is characterized by broad-based T-waves, LQTS2 by low-amplitude notched T waves, LQTS3 by late-onset peaked or biphasic T-waves.

Short QT syndrome (SQTS) is an extremely rare inherited ion channel disease that has only recently described. It is characterized by a QTc interval constantly equal or less than 320 ms, T wave morphology abnormalities in the precordial

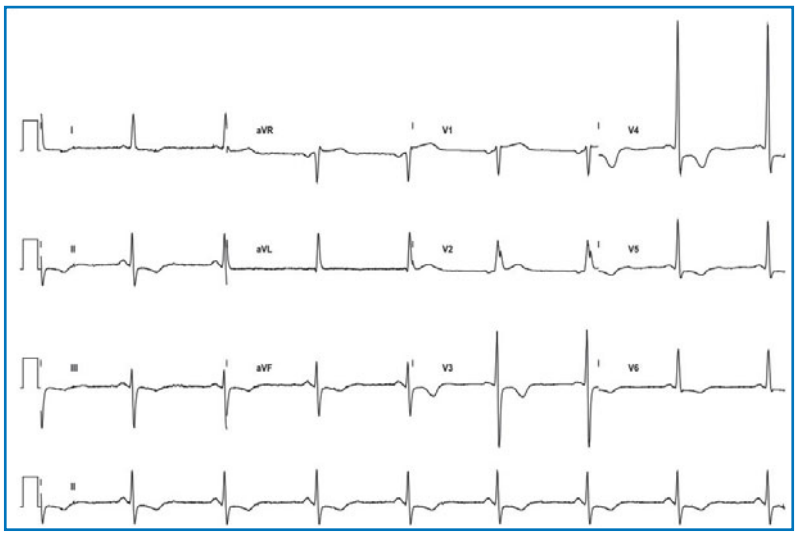

Fig. 2.1 ECG of a patient with hypertrophic cardiomyopathy

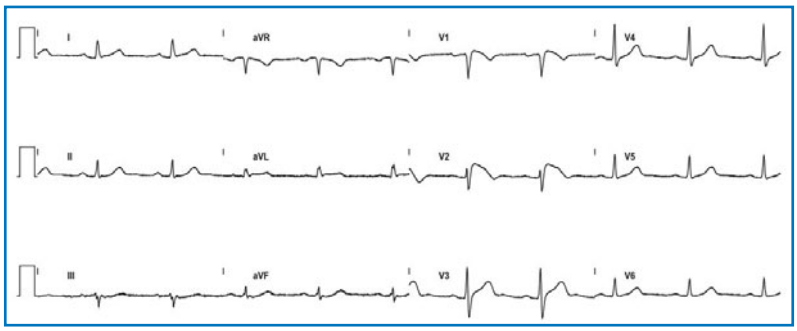

Fig. 2.2 Type 1 Brugada ECG pattern

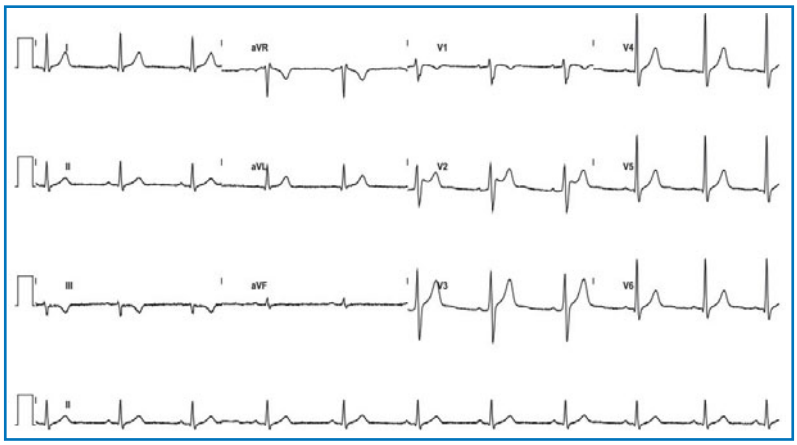

Fig. 2.3 Type 2 Brugada ECG pattern

leads and high incidence of atrial fibrillation, syncope and sudden cardiac death in the absence of structural heart disease. To date, few patients affected by this syndrome have been identified worldwide, including both familiar and sporadic cases [3].

Brugada syndrome is characterized by an ECG pattern of ST segment elevation in V1-V3 associated with documentation of ventricular tachycardia (often polymorphous), ventricular fibrillation, family history of SCD at a young age (< 45 years), otherwise unexplained syncope, nocturnal agonal respiration, in subjects with otherwise normal heart. There are three ST elevation patterns in the right precordial leads (V1-V3) that may suggest the presence of this syndrome:

- type 1 (coved type): marked J point and ST segment elevation ≥ 2 mm or 0.2 mV at its peak, followed by a negative T wave, with no isoelectric separation (Fig. 2.2);
- type 2: high early ST segment elevation ST (≥ 2 mm at the J point) giving rise to a gradually descending ST-segment elevation (remaining ≥ 1 mm above the baseline), followed by a positive or biphasic T-wave that results in a saddle-back configuration (Fig. 2.3);
- type 3: ST elevation in the right precordial leads < 1 mm with a saddle-back or coved configuration.

These descriptions are based on the correct placement of the right precordial leads. However, in patients with a high clinical suspicion of Brugada syndrome who have a normal baseline ECG, alternative placement of the right precordial leads in a superior intercostal space (or, in selected cases, rightward displacement) may disclose a Brugada-like pattern. It should be stressed that ST segment alterations are often dynamic, and a typical ECG pattern may be unmasked by several clinical conditions such as a febrile state or pharmacologic agents. Intravenous administration of class I antiarrhythmic agents (sodium channel blockers) such as flecainide, propaphenone and ajmaline may unmask a Brugada pattern, and this is commonly used as a provocative test when a high clinical suspicion of Brugada syndrome is present.