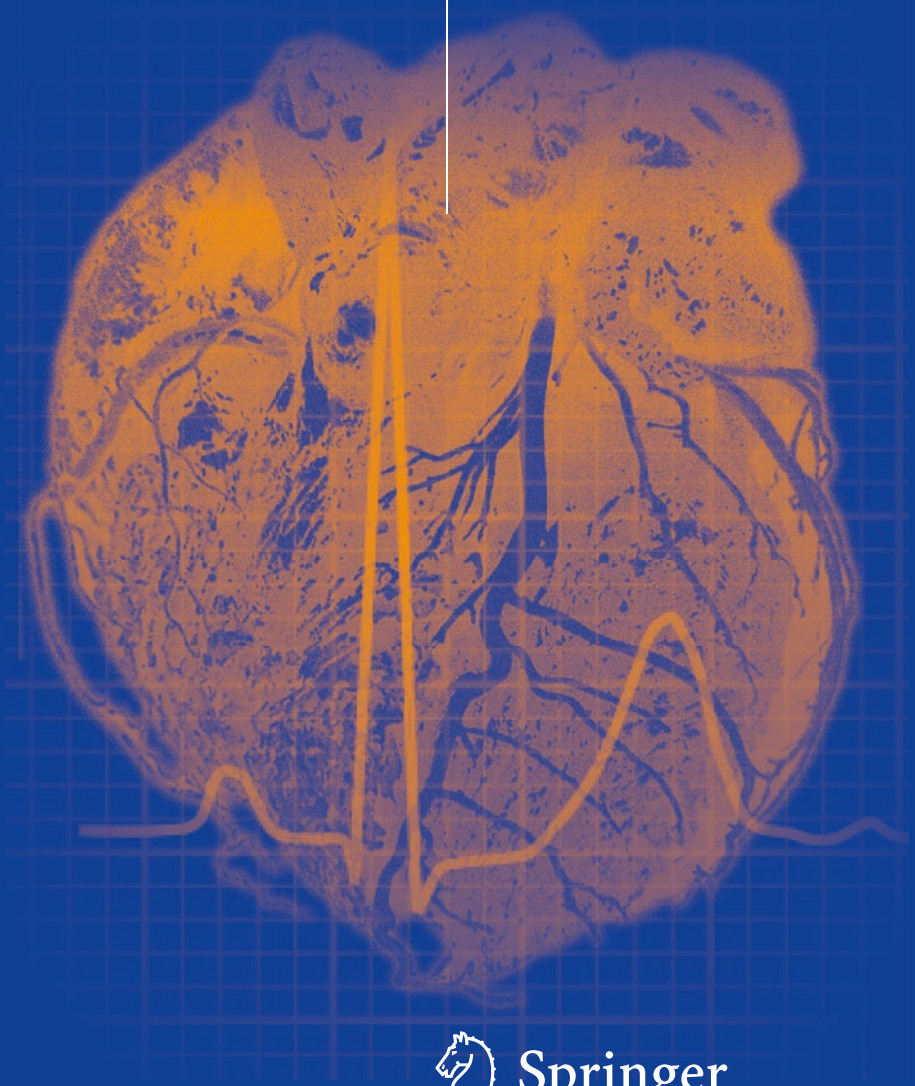
Gabriele Bronzetti

Atlas of Pediatric and Youth ECG



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To Angelita, Francesco, and Chiara, for all your heartbeats that I wasn't there 4

Foreword I

One of the most remarkable transformations in life is the growth of an infant into a mature adult. Human infants are helpless when born and have one of the longest periods of dependence on their parents of any creature. Every organ of the body goes through a constant process of growth and development ultimately culminating in the emergence of a mature adult. Among the organs that change significantly in the process of growth is the human heart. The development of electrocardiography as a method of recording the electrical activity of the heart gave medicine dramatic new insights into both normal cardiac physiology and its pathophysiology. These initial investigations focused almost exclusively on adult men, with little appreciation of the ways in which electrocardiogram may differ in women and the developing child. The emergence of pediatric cardiology as a distinct discipline has emphasized the need to establish the unique electrocardiographic characteristics of heart in child and adolescent in health and in response to illness. While there have been several attempts at producing comprehensive texts to explore the wide range of electrocardiographic findings in children and adolescents, the subject has not received the attention that it so richly deserves. The current book by Gabriele Bronzetti admirably helps fill this need. Using the insights and experiences gained over a career spent tending to the cardiac needs of children and adolescents, he has produced a remarkable atlas of electrocardiograms that cover the entire spectrum of disorders found in clinical practice. The book provides a rich and invaluable resource for those who wish to explore the fascinating and complex world of ECG findings in younger patients. One of the book's strengths is the meticulous attention to a detailed analysis of each ECG presented, as well as the clarity of the recordings themselves. Bronzetti's work will prove an indispensable resource for anyone interested in the complex and fascinating world of pediatric electrocardiography.

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Foreword II

"A doctor walks into a bar and says to the bartender…"

No, this is not a joke. In many ways it was a pivotal moment of Dr. Bronzetti's life. The rub: Gabriele Bronzetti wasn't the doctor; he was the bartender. Thus began (not even arguably) the most important of Dr. Bronzetti's many passions: his love for his amazing wife Angelita, who he eventually followed into the study and practice of medicine. Perhaps someday she'll tell me the story of what drew her as a young neonatologist into that bar on that day.

Dr. Bronzetti's second passion, which also may have had its origins that day, is for medicine and eventually pediatric cardiology and its heart rhythm disorders. His third passion is for teaching, as is aptly demonstrated in this book on pediatric electrocardiography. His next passion (of which I am aware) is for the arts and written word, which is what makes this *Atlas of Pediatric and Youth ECG* uniquely special. His final passion is for his own children, but also extends to the patients and families that he cares for so expertly.

I met Gabriele Bronzetti when he came to train with me in pediatric electrophysiology at The Hospital for Sick Children, Toronto, in 2004. Gabriele was adored by staff and patients alike and excelled in both learning our discipline and teaching it to the core fellows, residents, and students within our program. His appreciation for the fine arts, as well as other important aspects of life, became obvious to me one evening when he invited me home for dinner. With two of my younger and more boisterous children in tow, I arrived at his home to realize he was staying in the Toronto residence of a renowned art expert, and I spent the evening envisioning and avoiding priceless pieces being destroyed by my offspring. As dinner was being prepared, I heard what to a non-Italian sounded like the most passionate of arguments coming from Gabriele and Angelita in the kitchen. I eventually realized this was all about whether the pasta was done just right. It is with this background that I introduce you to Dr. Bronzetti, passionate physician, teacher, husband, and father, whose grasp of language and the written word, whether in English or Italian, defines the word "art."

Electrocardiography is to a great extent part of the art of medicine. While the statistical norms have been finely calculated for adults and children by giants such as Drs. Pentti Rautaharju and André Davignon, the application of electrocardiography to diseases has primarily relied on translation of empiric observations into defined set of rules. Thus, we "interpret" the results of an ECG. Since only normal values are well described, this can lead the casual observer to consider an ECG as the "sedimentation rate" of the heart. It's great if it is normal, but if not, you'd better do a battery of other tests to find the cause of the abnormality. This simplistic approach is hazardous, as the ECG contains many measurements, some of which, when defined at the 98th percentile, may be statistically abnormal in any individual. On the other hand, certain patterns can be recognized for specific cardiac lesions and can provide the astute clinician with a likely diagnosis while awaiting more definitive studies such as echocardiography. These classic findings in congenital heart disease are beautifully described and illustrated in Chap. 18 of this atlas.

In the first chapter, Dr. Bronzetti provides philosophical approaches to the evaluation of the ECG, invoking classics from children's books to weighty novels, and reviews the indications for electrocardiography. The second chapter introduces the reader to basic concepts of electrocardiography, this time using classic sculptures, and provides further philosophy on the

reporting of ECGs. Chapters 3 and 5 introduce the reader to the development of electrocardiographic waves (both normal and pathological) and their ontogeny through childhood, this time with some biblical references for interest. Chapter 4 takes a magical tour of the "numerology" of ECGs, including calculation of rate and axis.

Chapter 6 defines normal ECGs and normal variants. Those entrusted with ECG interpretation, hopefully also provided with context, should remember to highlight findings deserving further assessment and minimize variations that might lead to unnecessary investigation or worry. Chapters 7 through 9 highlight ECG findings of metabolic derangements, artifacts and devices, and bradycardias and conduction delays, respectively. Each of these chapters is beautifully illustrated with dozens of high-quality electrocardiograms demonstrating these specific findings.

Electrocardiography remains the mainstay for diagnosis of cardiac rhythm disorders. Chapters 10, 11, and 12 describe the electrocardiography of preexcitation syndromes, supraventricular tachycardia, and ventricular tachycardia. Given our renewed understanding of the implication of preexcitation in children, the reader is appropriately referred to the 2012 Guideline document. The electrocardiography of SVT subtypes is described, an information which may be crucial in guiding long-term management. Ventricular tachycardia, albeit rare in children, is described in detail due to the importance of determining its etiology.

As might be expected from an Italian cardiologist, the electrocardiography of cardiomyopathies is beautifully described and differentiated. Similarly, the identification and differentiation of channelopathies are well described and illustrated. Finally, Dr. Bronzetti rounds out with chapters on cardiac tumors, pulmonary hypertension, and ischemia.

This atlas provides beautiful illustrations, current references, and a philosophy and style that is a pleasure to read. I congratulate Dr. Bronzetti and "whole-heartedly" recommend his work to you, the reader.

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Preface

One of the most fascinating and mysterious aspects of an ECG is the wealth of information hiding behind a seemingly innocent tracing. The ECG is a technically simple examination that produces familiar writing on a piece of paper, in a language that has stayed the same since it was invented more than a century ago. While echocardiography now uses machines that produce images that even 20 years ago were unimaginable, an ECG performed today displays the same signs seen by the founding fathers of electrocardiography, from Einthoven onward.

In the space of a century, we have discovered the many diseases that can lie concealed between the waves, from channelopathies to cardiomyopathies, from arrhythmic syndromes to congenital heart diseases. In the meantime, the ECG has been supported but not replaced by the new imaging techniques and has earned a precise role in screening programs for various conditions, in particular in sports preparticipation screening for prevention of sudden cardiac death, a program that is already strongly supported in Italy but which is now spreading throughout the world. Without getting into any discussion, it is known that the "Italian" position considers the ECG to be a cost-effective method. The ECG is generally satisfactory in terms of specificity and sensitivity; in other words, a normal ECG reassures the cardiologist and the families of young sports players. The most important thing, however, is to recognize when an ECG is normal, avoiding false-positive results and all related implications in terms of additional tests and examinations as well as the risk of discouraging sports activities. Sports activities aside, ECG still plays a key role in the diagnosis of arrhythmias and ischemic syndromes. Nevertheless, the interpretation and deductive analysis of an ECG is no easy task, but requires curiosity, clinical experience, and culture. There are no shortcuts, and the quick routes on a cardiac map are usually accessory pathways and should not be followed. The ECG seizes and stops the anatomy of an instant, a hermeneutic process even more difficult in babies and in the young where the heart is changing day by day. This continuous alteration during developmental age takes the cardiologist by surprise: in the attempt to keep up with the new methods and the enormous number of publications, he or she has been obliged to neglect electrocardiographic semeiotics and now struggles to interpret the tracings of a newborn, of a youngster with normal variants, or of a 60-year-old who has had cardiac surgery and suffers from hidden congenital heart disease. One of the particular features of this book is that all the tracings come from the same tertiary center. They were selected from thousands of tracings that I have personally collected over the last 20 years and which allowed me to make or confirm a diagnosis. The part for electrophysiology experts is kept to a minimum in order to leave room for examples and tracings that everyone working in the medical sector can understand. The ECGs that a pediatric cardiologist sees in a lifetime, the ones that a pediatrician imagines in his or her patients and that a cardiologist of adult patients fears when he or she is on night duty, are all included and commented on. This atlas can be also of interest to other players in the field of medicine – anesthetists, students, and nurses – who may be the first to see the mysterious pink paper emerging. No one should feel left out since, as Tolstoy would say, all normal ECGs look alike, but every ECG is in its own way abnormal.

Bologna, Italy — Gabriele Bronzetti

Acknowledgements

"I would like to thank all the Maestros we have had and would like to have had and all the young doctors that actively contributed to the writing of this book and whose thirst for knowledge made it necessary.

My thanks also to Susan West for improving my English here and there (her words, not mine!)."

Special thanks for the invaluable collaboration of Tanya Bodnar, Maurizio Brighenti, Cristina Ciuca, Alessandro Corzani, Cinzia D'Angelo, Junio Massimiliano Durante, Elisabetta Mariucci, and Monica Urru.

A special mention in memory of Gabriele Cristiani who contributed fundamentally to the construction of some images.

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Abbreviations

ACAOS	Origin of the coronary artery from the opposite sinus
AF	Atrial fibrillation
Aflutter	Atrial flutter
ALCAPA	Anomalous left coronary artery from pulmonary artery
ARCAPA	Anomalous right coronary artery from pulmonary artery
ARVC	Arrhythmogenic right ventricular cardiomyopathy
ASD	Atrial septal defect
AV	Atrioventricular
AVB	Atrioventricular block
AVNRT	Atrioventricular nodal reentrant tachycardia
AVRT	Atrioventricular reentrant tachycardia
BAV	Bicuspid aortic valve
BER	Benign early repolarization
BS	Brugada syndrome
CAVC	Complete atrioventricular canal
CCTGA	Congenitally corrected transposition of great arteries
CPVT	Catecholaminergic polymorphic ventricular tachycardia
DCM	Dilated cardiomiopathy
EAT	Ectopic atrial tachycardia
ECG	Electrocardiogram
ECMO	Extracorporeal membrane oxygenation
ERP	Early repolarization pattern
HCM	Hypertrophic cardiomiopathy
HR	Heart rate
JET	Junctional ectopic tachycardia
LBBB	Left bundle branch block
LGL	Lown-Ganong-Levine syndrome
LQTS	Long QT syndrome
LVH	Left ventricular hypertrophy
PAC	Premature atrial contraction
PAPVR	Partial anomalous pulmonary venous return
PMK	Pacemaker
PJRT	Permanent junctional reciprocating tachycardia
PSVT	Paroxysmal supraventricular tachycardia
PVC	Premature ventricular contraction
QTc	Corrected QT
RBBB	Right bundle branch block

RVH	Right ventricular hypertrophy
RVOTVT	Right ventricular outflow tract tachycardia
SVT	Supraventricular tachycardia
TAPVR	Total anomalous pulmonary venous return
TGA	Transposition of great arteries
VSD	Ventricular septal defect
VT	Ventricular tachycardia
WPW	Wolff-Parkinson-White

Introduction

1

The real voyage of discovery consists not in seeking new landscapes, but in having new eyes

Marcel Proust

1.1 A Literary Premise to a Practical Book

Is another book on the electrocardiogram (ECG) really necessary if there are already so many? One could venture a literary answer, stating that those who know Anna Karenina should not ignore Madame Bovary. We do not know and will never cease to talk about the heart enough, the heart that follows the rules and the heart that is forced to betray them because of congenital or acquired reasons, because of hemodynamic or electrical reasons. More prosaically, one can add that among the many published texts, only a few are dedicated to children and the young.

This book would like to share a simple, fresh look at a set of more or less orderly signs on pink paper, which reveals and hides a treasure chest of 12 leads in 12 interminable seconds. The ECG is crystallized time like a Jurassic butterfly in a drop of amber. Faced with the pink paper, any person may feel like an entomologist watching an insect during its metamorphosis and a scientist, like an artist, who does not invent but discovers. The truth is in the detail: almost everything is in the almost nothing of a small or hidden wave. Physicians should seize tiny clues and as Proust says "we must break the spell that holds things captive, bring them down to us and prevent them from falling forever into emptiness." We must learn to see the detail that reflects or deflects from *normality*, sure only of its vast and elusive nature. We have to dig up the past, stop the present, and foresee the future using various arts and cultures, even the "intermittence of the heart." These are arrhythmias of Proustian memory, revealing beats that open up time with the ephemeral and irrevocable grace of a butterfly. If growing up means becoming what we already are, we can only really know the ECG of an adult by looking at the ECG of the baby that the adult once was.

Ferdinand Céline was a French physician who became a famous writer. He said that adults are children gone bad. So even the doctor with little or no specialization in pediatrics eventually feels the need to know what the patient's ECG was like before the deterioration. In "The Metamorphosis" by Franz Kafka, Gregor Samsa becomes an insect and dies unrecognized even by his family. Culture and science, on the other hand, should serve to discover the man hiding in an insect.

To start off on the right foot, we should rely on Tolstoy, and citing the opening words of his "Anna Karenina," we have to recognize that *all normal ECGs look alike but every ECG is abnormal in its own way*.

1.2 Why Does Everything Begin with P?

There are mathematical explanations dating back to Descartes that put the P at the beginning of the ECG glossary, and not the A as would be more intuitive [1]. However, if the smallest wave, the one that first comes out of the heart and breaks through the skin to land on the pink paper, is called P, there must be other reasons. Many childhood heroes begin with P: "Le Petit Prince (The Little Prince)," to tell us that what is essential is invisible to the eye and can only be seen with the heart. "Pinocchio," to remind us that the ECG is sincere but does not always tell the truth. "Peter Pan," with his desire never to grow up, just like the P wave. Le Petit Poucet or Pollicino (Tom Thumb), the cunning P, able to disguise and hide. The Dickensian Philip Pirrip "Pip" of "Great Expectations." "Pippi Longstocking" by Astrid Lindgren, herself an orphan, an example of power to the imagination. The "Pink Panther" of seraphic guile. Schulz's cartoon strip "Peanuts," with the gang including the dynamic "Peppermint Patty." Not to mention Walt Disney's "humanized" dog Pluto, an example of candor and levity. Then we have the dreamy Pimpa, with its four paws firmly planted in the clouds. Finally, a sign of the times is the very current "Peppa Pig," which sums up the ECG with porky pragmatism. In fact, pork is pink and cheap and can be used up to the very last bit. In German culture there is the "Eierlegende Wollmilchsau," a mythic pig that lays eggs like a chicken, gives milk like a cow, wool like a sheep, and ends up in ham and chops, practically like an ECG, which sometimes acts as an echocardiogram, a catheterization, and an MRI all in one.

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1.3 ECG Waves: An Entomological Vision

On an ECG and on the palm of a hand, the body of a beat can be dissected into three parts like a beetle (from the Greek "en temnein," cut into pieces) (Table 1.1, Fig. 1.1), namely, the head, the P wave and PR segment, where everything starts; the chest, the QRS complex, where everything moves; and the abdomen, the ST-T segment, restoring ionic balance and where everything returns to baseline status. We start off on our campaign with a butterfly net of graph paper to capture the enchanting winged creature and study it; the subjects of our research could be at different stages of development. In insect metamorphosis there are four stages: egg, larva, pupa, adult insect (the latter also called imago, or imagine). The ECG is the only image, the only visible phase of the electrical activity of the heart. More precisely, we can recognize four ECG stages: neonatal age up to 1 month, early infancy up to 3 years, second and third periods of infancy up to 12 years, and adolescence and adulthood after the age of 12 years.

The ECG approach cannot ignore the knowledge of its diachronic development, the real objective of the pages that follow.

Childhood is endless and over the years, and in the absence of disease, the P waves remain surprisingly unchanged, while the T and R fly. To condense the ineffable concept, having cited Tolstoy we must mention Nabokov. The Russian writer was not only a great expert on butterflies;

Table 1.1 ECG's dissection and deduction

What to look	Finding	Suspected diagnosis
Atrial wave		
Rhythm	A-V dissociation, bradycardia A-V dissociation, tachycardia	III degree AVB JET (narrow QRS)—VT (wide QRS)
P wave morphology	Negative P wave D_2-D_3-aVF, tachycardia Abnormal P wave axis	Incessant arrhythmia (PJRT, AT) Ectopic atrial rhythm, atrial isomerism, dextrocardia
	Peaked P wave >2.5 mm P duration >120 ms	PS, TA, PAIVS, Ebstein, PEX, RCM Mitral steno/insufficiency, HCM, RCM
PR interval		
	Short PR—Delta wave Long PR	Metabolic (Pompe, Danon, Fabry, PRKGA 2), mitochondrial (LHON, MELAS), TS, Duchenne-NMD, pre-excitation AVC, CCTGA, RF, KD, NMD, Brugada, HCM
QRS interval		
Electrical axis	Left axis deviation Right axis deviation (extreme)	AVC, TA, ostium primum ASD, VSD, CCTGA, Steinert Noonan syndrome, severe RVH
Q waves	Q in right precordial leads (always pathologic) Anterolateral, inferior pathologic Q waves Absent Q wave in V6	Severe RVH, CCTGV, univentricular heart ALCAPA, Duchenne,-NMD CCTGV, LVH/RVH, normal variant
Delta wave	Delta wave + short PR	Pre-excitation/WPW, Ebstein, CCTGV, HCM, TS
Epsilon wave	Small positive wave in the end of QRS	ARVC
J wave	Slurring/notching of terminal part of QRS	Hypothermia, early repolarization (Brugada)
Bundle branch block	SR—VT (RBBB—LBBB morphology)	ASD/RV overload, idiopathic VT, myocarditis, neoP, CMPs, normal variant, PM
Repolarization (S-T, T, QT, U)		
	↓ S-T right precordial leads (left) 0–7 days ↓ S-T right precordial leads >7 days Positive T wave in V1: 7 days–7 years of age Negative T wave V2–V4 < 12–14 years Negative T wave >14 aa ↓ left precordial-inferior leads ↑ S-T multiple leads ↑ S-T V1–V3 Prominent U waves (>50% of T wave)	PPHN RVH RV overload Juvenile pattern Overload, ischemia, electrical memory, neoplasms, CMPs ALCAPA, CMPs Pericarditis, myocarditis, MI, BER, PNX Brugada, BER Hypokalemia, long QT, bradycardia

ASD atrial septal defect, *ARVC* arrhythmogenic right ventricular cardiomyopathy, *AT* atrial tachycardia, *AVB* atrioventricular block, *AVC* atrioventricular canal, *CCTGA* congenitally corrected transposition of great arteries, *BER* benign early repolarization, *CMPs* cardiomyopathies, *HCM* hypertrophic cardiomyopathy, *JET* junctional ectopic tachycardia, *LVH* left ventricular hypertrophy, *KD* Kawasaki disease, *LHON* Leber's hereditary optic neuropathy, *MELAS* myopathy, encephalopathy, lactic acidosis, and stroke-like, *MI* myocardial infarction, *NMD* neuromuscular diseases, *neoP* neoplasms, *PAIVS* pulmonary atresia with intact ventricular septum, *PEX* pectus excavatum, *PKRGA 2* protein kinase AMP-activated non-catalytic subunit gamma 2, *PJRT* permanent junctional reciprocating tachycardia, *PMK* pacemaker, *PNX* pneumothorax, *PPHN* persistent pulmonary hypertension of the newborn, *PS* pulmonary stenosis, *RCM* restrictive cardiomyopathy, *RF* rheumatic fever, *RVH* right ventricular hypertrophy, *SR* sinus rhythm, *TA* tricuspid atresia, *TS* tuberous sclerosis, *VSD* ventricular septal defect, *VT* ventricular tachycardia

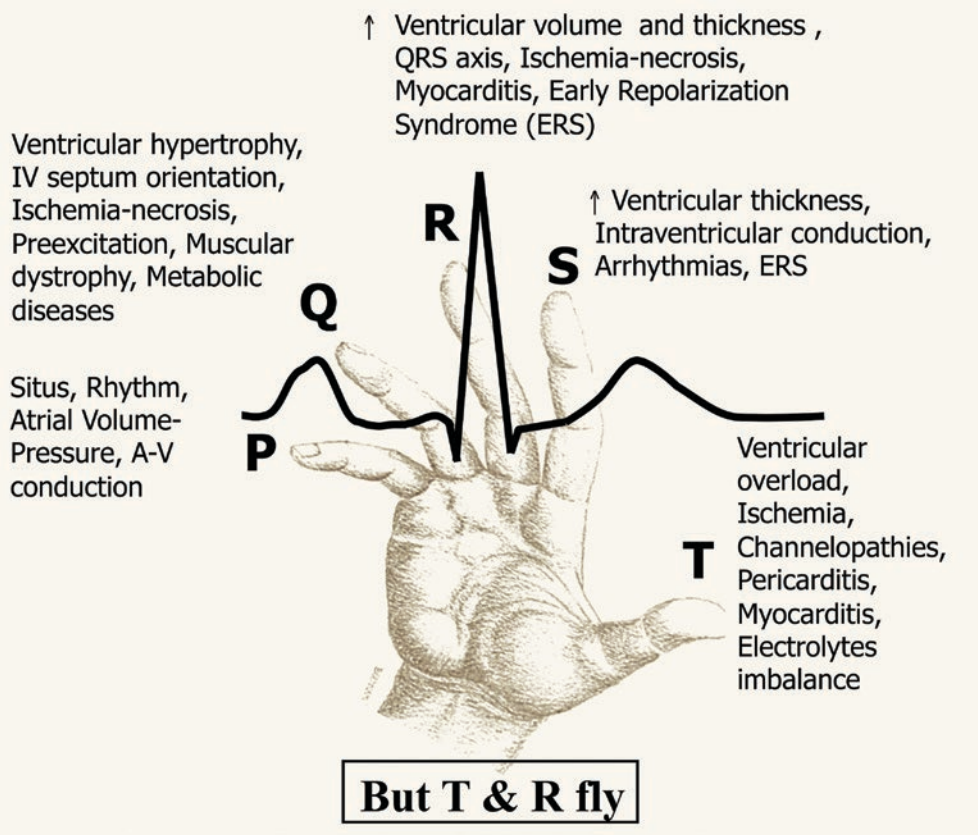

Fig. 1.1 The ECG in the palm of the hand and at your fingertips

he was also able to write masterpieces directly in his adopted language, English. The P remains the same, but T and R fly. In a word, BUT T & R FLY.

Immediacy and low cost are the features that make the ECG absolutely modern and, combined with a skillful "imagination," able to be ahead of the more expensive cardiac imaging. In emergency situations such as arrhythmias or chest pain, when it's just you and the patient and you cannot delay the diagnosis by asking a colleague or performing another examination, the ECG remains an unavoidable tool of bearable lightness (Table 1.2).

The book is addressed to everyone, students, nurses, and doctors (mindful cardiologists, inquisitive pediatricians, and alert anesthesiologists) more or less familiar with young and older patients. We have carefully avoided an introductory section on electrophysiology, not essential to the understanding of the text and which in any case can be found in infinite variations, both in the attic and on the web. The book collects the essence of exams carried out in a reference center over the past 20 years. At this point the good teacher would admonish her/his pupils with "mind your p's and q's" which, in the ECG's world, is literally the most appropriate advice I can give to the reader.

Table 1.2 Common indications for pediatric ECGs

Screening	Neonatal (nonmandatory), mother with systemic lupus erythematosus, sports
Symptoms	Palpitations, syncope, chest pain, congestive heart failure, cyanosis, epilepsy
Conditions	Rheumatic disease, Kawasaki disease, pericarditis, myocarditis, hypertension, neuromuscular disease, electrolyte imbalance, infectious disease (Lyme disease, etc.)
Family history	Congenital heart disease, cardiomyopathy, primary electrical diseases, sudden death
Therapy	Antiarrhythmic, pacemakers, anticancer, diuretic, QT-prolonging drugs, poisoning

Reference

1. Hurst JW. Naming of the waves in the ECG, with a brief account of their genesis. Circulation. 1998;98:1937–42.

Suggested Reading

Barrier JM. Hollywood cartoons: American animation in its golden age. Oxford: Oxford University Press; 1999. isbn:978-0-19-503759-3

Proust M. À la recherche du temps perdu. Bernard Grasset et Gallimard; 1913.

Tolstoy L. Anna Karenina. The Russian Messenger; 1877, isbn: 978-1-84749-059-9.

Basic Concepts of Electrocardiography

2

We did not start from the action potential, and we have deliberately not got bogged down in vectorcardiographic analysis. So we can say that Willem Einthoven is our *notable absentee*. The "child" vision requires an intuitive approach which nevertheless seeks to translate the deep, unconscious electrophysiology into a concept of surface: the vector and its symbol, the arrow shot by ions that travel across the membrane proteins.

The activity of the heart creates an electric field that can be detected by surface electrodes. This electrical activity may be assimilated to a dipole, a system consisting of two electric charges of equal and opposite sign and separated by a constant distance in time. When the atria (or ventricles) undergo depolarization, the wave that spreads across the muscle takes many different directions simultaneously and can be represented by arrows. Each arrow represents a different individual vector, and the set of these individual vectors can be summarized by the mean (or resulting) electrical vector. It is best to keep in mind that the forces generated in opposite directions can neutralize each other. The most intuitive way to read the ECG is to think of this vector (which applies to every wave from P to T) as an arrow that we see from our vantage points and that is the standard 12-lead ECG. The direction of this arrow relative to the electrode determines the magnitude of the recorded voltage (the voltage of course also depends on the mass of the cardiac tissue involved). When the arrow is approaching, the electrode which sees the tip will record a positive voltage and then a positive wave; conversely, when the arrow moves away from the electrode and this sees the bottom, it will react with a negative voltage and a negative wave (Fig. 2.1).

The arrow that we imagine on the ECG goes a long way and travels across a sky called *conduction system*. This skyway begins in the pale cells of the *sinoatrial node*, located in the upper right atrium where the superior vena cava ends. Hence, the impulse that arises from spontaneous depolarization regulated by the autonomic nervous system and by circulating hormones will go toward the left atrium through the muscle cells but also past the Bachman bundle and toward the ventricles with internodal pathways and through the working myocardium. The AV junction consists of a proximal part, the AV node, and a distal part, the bundle of His. From the His bundle, the two main branches will finally start and will end in the ventricles to the Purkinje fibers. Now ventricles are activated and the electromechanical coupling can move the blood. The main ventricular vector can be divided in three parts, taking into account that the right ventricle starts to depolarize shortly after the left ventricle: the first part represents the resultant force of the septum (vector 1), moving from the left toward the right and triggering the q wave in V6 (and lead I and aVL); the second represents the activation of the free wall of ventricles (vector 2); the third represents the activation of the postero-basal aspects of ventricles (vector 3).

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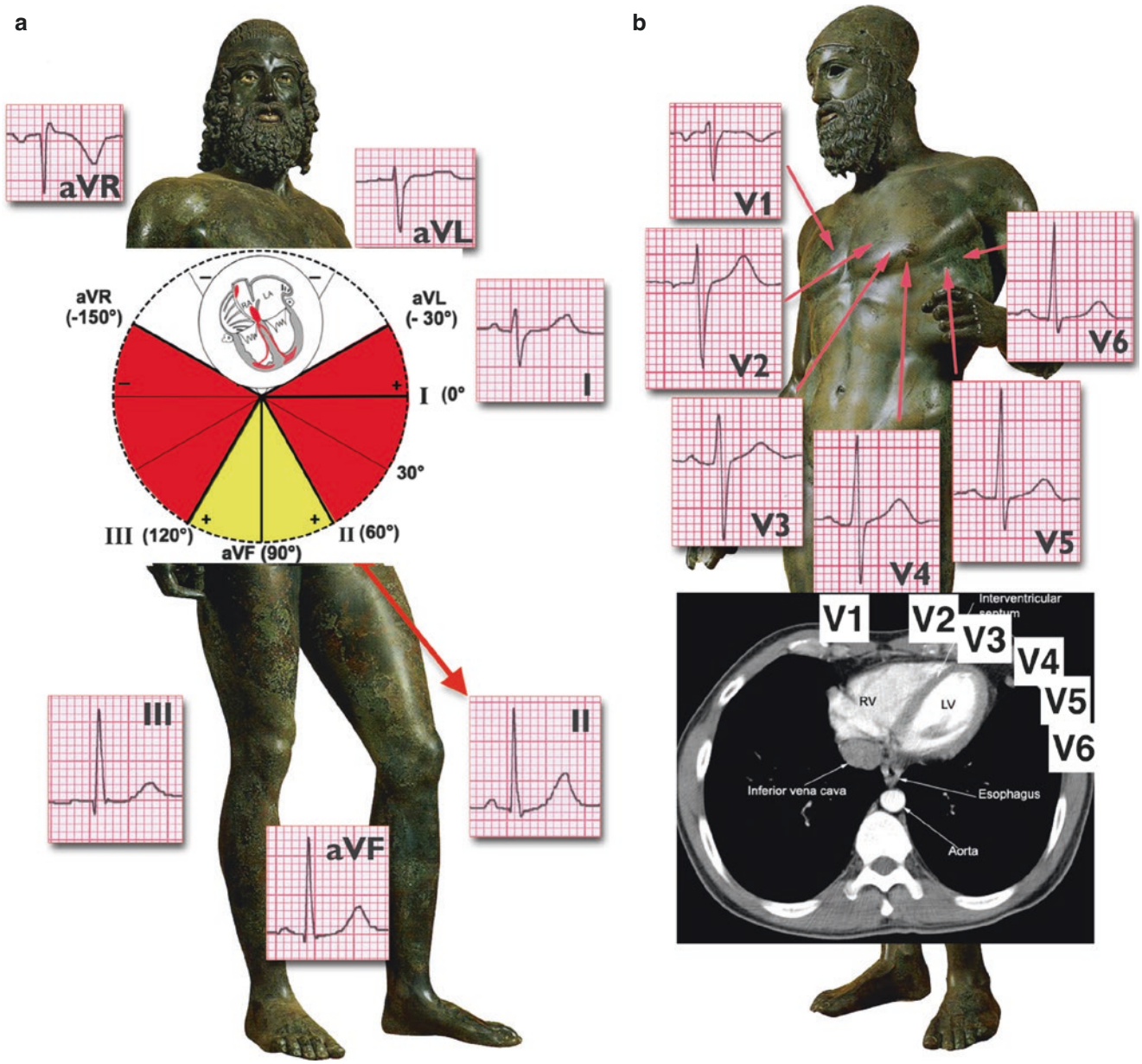

Fig. 2.1 The Riace Bronzes as models for a 12-lead ECG in a 12-year-old boy. **Bronze (a)** the one with the more restless style. Frontal plane, standard limb leads (I, II, III), and augmented unipolar (aVR, aVL, aVF); like the style of the statue, these leads are sensitive to movements and artifacts. **Bronze (b)** horizontal plane, precordial leads V1 (fourth intercostal space, right parasternal), V2 (fourth intercostal space left parasternal), V3 (equidistant between V2 and V4), V4 (fifth intercostal space midclavicular line), V5 (fifth intercostal space anterior axillary), V6 (fifth intercostal space, axillary media); like the style of the statue, these leads are more stable and relaxed.

2.1 Technical Aspects

Before moving on to atrium and ventricular dissection, it is worth remembering some technical aspects. You should always check that paper speed and calibration are standard and thus respectively 25 mm/s and 10 mm = 1 mV. An increase in speed or amplitude may be required when greater precision in estimation of intervals or small amplitude waves is important. True dextrocardia must be distinguished from misplacement of the limb leads (the so-called nurse's dextrocardia).

When faced with an improbable ECG, you have to question the correctness of lead placement, sometimes poorly positioned due to staff carelessness, thoracic deformity, dressings, etc. [1, 2] The surface ECG is a simple technique, but that does not mean it can be performed superficially. Just like cooking, it is the simplest recipes that require perfection of the few necessary ingredients. Every laboratory should comply with the standard placement of the electrodes, and nurses should be skilled in being able to put children of all ages at their ease, resorting to various arts ranging from soap bubbles to smartphones.

In remote-viewed tracings, whether sent by fax or other means, you have to make sure that the relationship with the original is 1:1; even small differences of scale can lead to blunders, for example in the calculation of the QT interval (if you don't know the scale, you can count the little boxes that at standard paper speed correspond to 40 ms). Sometimes there are funny artifacts: the oscillatory ventilation often used in neonatal intensive care can simulate drug-resistant flutter (diagnosis and therapy are electric, just switch off the oscillator) [3]. Even hiccups can create bizarre waves resembling extrasystoles which are neither supraventricular nor ventricular, but simply underventricular or diaphragmatic. Congenital heart disease may require nonstandard leads such as V3R, V4R, and V7: the first two increase the sensitivity of the right overload [4], while V4R (positioned on the right midclavicular fifth intercostal space) can be an excellent alternative to V1–V2 which cannot be applied due to dressings or for other reasons. With ECGs that are "RBBB like" or with an "atypical" RBBB, in family members of patients with Brugada syndrome, or in the case of unexplained syncope, in addition to the ECG standard you should do an ECG with right precordial leads (V1 and V2) moved high in the third and in the second intercostal space. In the same conditions, it is also useful to perform an ECG in the course of fever as it is known that hyperthermia can unmask the Brugada pattern. Modern electrocardiographs have sample rate and bandwidth higher, respectively, than 500 samples/s and 250 Hz – they write very well but they don't read quite so well. Automatic diagnosis may be incomplete, petulant, or catastrophic and is not always reliable, especially when the machines do not know they are dealing with a child and see infarction instead of infants [5]. "Manual" diagnosis should avoid as much as possible scary terms like "block," "delayed," "nonspecific alteration," and "anomaly" which are not much help. When these terms are unavoidable, they should be followed by conclusions that are as reassuring as possible. You should also avoid defensive adverbs and phrases such as *likely* and *cannot be excluded* followed by a dozen differential diagnoses. The perfect report consists of a single word: "N o r m a l." A broken clock tells the correct time at least twice a day. Cardiologists who are too far forward or too far back never get it right. For all the time and amplitude parameters, refer to the normal value tables; the Davignon and Rijnbeek tables are still useful [6, 7] (Table 2.1).

Table 2.1 Values of normal ECG parameters according to age

	HR	QRS axis	QTc*		PR	R wave*		S wave*		Q wave*	
	(bpm)	(degree)	M	F	(ms)	V1	V6	V1	V6	III	V6
0–3 day	90–155	60–170	480	480	70–150	27	12	21	10	3	3
3–7 day	90–155	65–165	480	480	80–140	24	12	17	10	3	3
7–30 day	95–180	65–160	440	440	70–130	21	17	11	10	4	4
1–3 month	95–180	30–120	440	440	70–130	(12) 20	(15) 22	13	7	5	3
3–6 month	95–180	5–105	440	440	70–150	(13) 22	(16) 27	17	10	7	4
6–12 month	95–170	5–100	440	440	70–160	(11) 20	(17) 27	18	7	8	6
1–3 year	90–150	5–100	440	450	80–160	(10) 21	(18) 29	21	7	7	6
3–5 year	70–135	5–100	440	450	80–160	(9) 18	(19) 31	22	5	5	6
5–8 year	65–132	5–140	450	460	90–160	(6) 15	(20) 29	23	4	6	7
8–12 year	60–130	5–110	450	460	90–170	(5) 11	(21) 32	25	4	7	7
12–16 year	60–120	5–130	450	460	90–180	(5) 11	(20) 30	22	4	7	7

( ) = median; * = 98° centile

References

1. Dickinson DF. The normal ECG in childhood and adolescence. Heart. 2005;91:1626–30.
2. Sharieff GQ, Rao SO. The pediatric ECG. Emerg Med Clin North Am. 2006;24:195–208.
3. Bronzetti G, Canzi A, Picchio FM, Boriani G. Fluttering waves in electrocardiograms recorded in neonatal intensive care unit. Int J Cardiol. 2003;92:299–301.
4. Akula R, Hasan SP, Alhassen M, Mujahid H, Amegashie E. Right-sided EKG in pulmonary embolism. J Natl Med Assoc. 2003;95:714–7.
5. Kligfield P, Gettes LS, Bailey JJ, et al. Recommendations for the standardization and interpretation of the electrocardiogram: part I: the electrocardiogram and its technology: a scientific statement from the American Heart Association electrocardiography and arrhythmias committee, council on clinical cardiology; the American College of Cardiology Foundation; and the Heart Rhythm Society: endorsed by the International Society for Computerized Electrocardiology. Circulation. 2007;115:1306–24.
6. Davignon A, Rautaharju P, Boisselle E, Soumis F, Mégélas M, Choquette A. Normal ECG standards for infants and children. Pediatr Cardiol. 1980;1:123–31.
7. Rijnbeek PR, Witsenburg M, Schrama E, Hess J, Kors JA. New normal limits for the paediatric electrocardiogram. Eur Heart J. 2001;22:702–11.

Suggested Reading

Bronzetti G, Bonvicini M. Pediatric ECG: chance, limits and necessity. G Ital Cardiol (Rome). 2014;15:720.

Chou T-C, Knilans TK. Electrocardiography in clinical practice - adult and pediatric, 4th edition. Saunders.

Goodacre S, McLeod K. ABC of clinical electrocardiography: Paediatric electrocardiography. BMJ. 2002 Jun 8;324:1382–5.

Price A, Kaski J. How to use...the paediatric ECG. Arch Dis Child Educ Pract Ed. 2014;99:53–60.

The Waves During Developmental Age

3

In the various stages of development, the ECG changes constantly, pursued with cognitive anxiety by the cardiologist. It is a horizon slipping away in front of us, an ontology of waves that knows no end because when it ends, the ECG can no longer be recorded. The handwriting of the heart depends on position, relative mass of the ventricles, physique, thoracic deformity, lung inflation, pneumothorax, and impedance of tissues. Intracellular accumulations increase the voltage and thus the amplitude of the waves on the paper, while extracellular accumulations or fibrosis depresses them. And so a newborn with a 21 g heart can generate a QRS complex as wide as that of an adult with a heart 10–15 times heavier; a young person with anorexia can show widespread low voltage typical of a devastating myocarditis; a massive myocardial hypertrophy with fibrosis can have lower voltage than a heart fivefold less thick or heavy (Table 3.1). The entomologist recognizes the age of bees through sight and smell: the young bee is hairier and more colorful and emits different Nasonov pheromones from an adult bee. To infer the age of a child, we could choose lead V1, look at it, and smell it, leading to enlightening diagnostic inspirations. You need intuition, a nose, and fragrant pink paper is much better suited than an aseptic computer screen. In addition to V1, lead II is also a formidable lead due to dissection of the P wave at baseline and during arrhythmias, to the atrial and ventricular electrical axis calculation, and to the accurate estimation of the QT interval.

Having to settle for just three leads—for intellectual challenge or need—you could supplement the abovementioned couple with a left precordial such as V5 or V6. In the majority of cases, 3 out of apostles 12 are sufficient to preach the electric *word*. Among the ancillary leads, aVR could be considered the most useless, given its persistent negativity. Only at the height of an ischemic calvary, such as proximal obstruction of the left coronary artery, does the ST segment of aVR rise to a sardonic positivity when its 11 colleagues become dramatically negative. In children, these scenarios are fortunately extremely rare (coronary artery dissection, acute thrombosis of an aneurysm in Kawasaki disease, compression of the left main branch of the left coronary artery by the pulmonary artery as can happen in ACAOS or in primary pulmonary hypertension or during balloon angioplasty of the pulmonary artery); in the section that examines the ST segment, the specific ischemic aspects of the first two decades of life will be discussed.

Table 3.1 Normal pediatric ECG findings

• Marked respiratory sinus arrhythmia (↑ isp↓ esp), physiologic low atrial rhythm
• Short PR and narrow ventricular complex
• Physiologic I degree and II degree AVB (Mobitz 1)
• Atrial and ventricular extrasystoles
• Axis deviation, QRS axis >90°
• Precordial leads mimicking dextrocardia in neonates (lead I is crucial)
• Prominent infero-lateral Q waves
• The repolarization is more important than voltage to diagnose ventricular hypertrophy
• Early repolarization pattern
• Negative T waves in V1–V4 (up to 12 years of age)
• Notched T wave in right precordial leads (V2–V3), prominent U wave

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3.1 P Wave

The P wave is fundamental, and unlike other waves, the normal values do not change significantly at different ages. *But T & R fly*. In sinus rhythm, the P wave is the sequential activation of the right and left atrium. It can be monophasic, but it is not uncommon for it to be notched or biphasic. The right atrial vector points down and ahead, while the left atrial vector points to the rear, to the left and down (Fig. 3.1). The resulting axis is in the left lower quadrant (0–90°), and the lead that sees it best is lead II, in addition to V1. The P wave can be +/− biphasic in all three inferior leads, while in aVL, it can be biphasic −/+, positive or negative. It must not, however, be negative in lead I; in this case, ectopic rhythm, malposition of the electrodes or dextrocardia should be suspected.

For all age groups, the amplitude limit is 2.5 mm, apart from newborns/infants where it extends to 3 mm up to 6 months. Especially in the very young, left atrial hypertrophy can be missed when a time limit of 120 ms is employed, given the smaller size of the atria and the greater atrial conduction velocity.

The diagnosis of atrial enlargement is valid only with ECG in sinus rhythm, as atrial ectopic beats may create an infinite spectrum of amplitude and duration of P waves [1]. Atrial enlargements are a result of a pressure overload (outflow obstruction or ventricular hypertension) or volume (insufficiency of atrioventricular valves [AV] or shunt) of the atria.

In right atrial enlargement (Fig. 3.2), in sinus rhythm, and in situs solitus, there is an increase in voltage of the initial atrial vector, while the voltage of the left atrium is unaffected; since the P wave is the sum of the right and left components and the left follows the right, there is no appreciable prolongation of P wave duration in right atrial enlargement:

- The P wave is high and pointed in the limb leads II, III, and aVF, and the voltage is increased and greater than 2.5 mm (3 mm in infants).
- The P wave in V1 may remain unchanged or may be pointed or diphasic (with a predominant positive component).

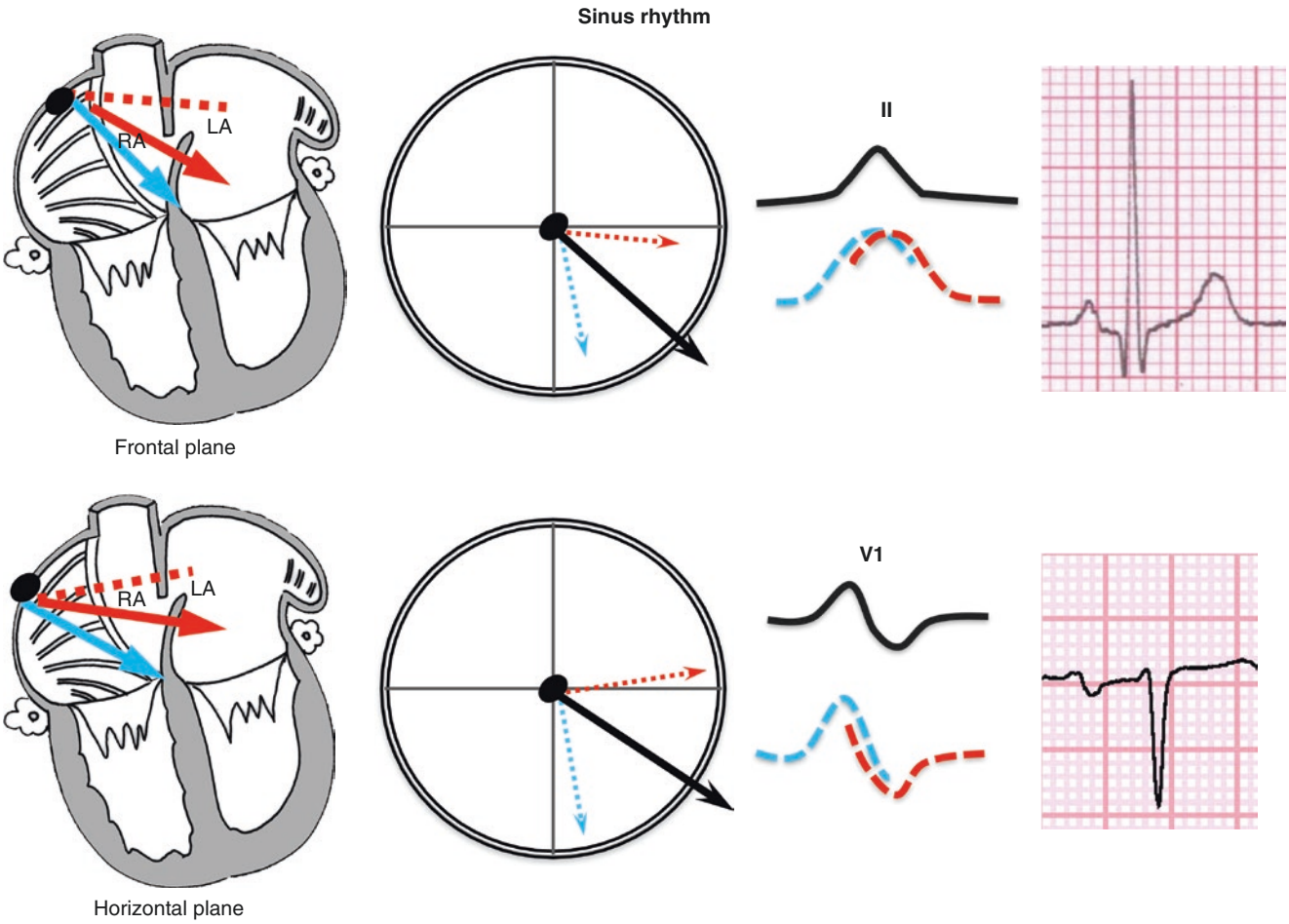

Fig. 3.1 Depolarization of the normal atrium

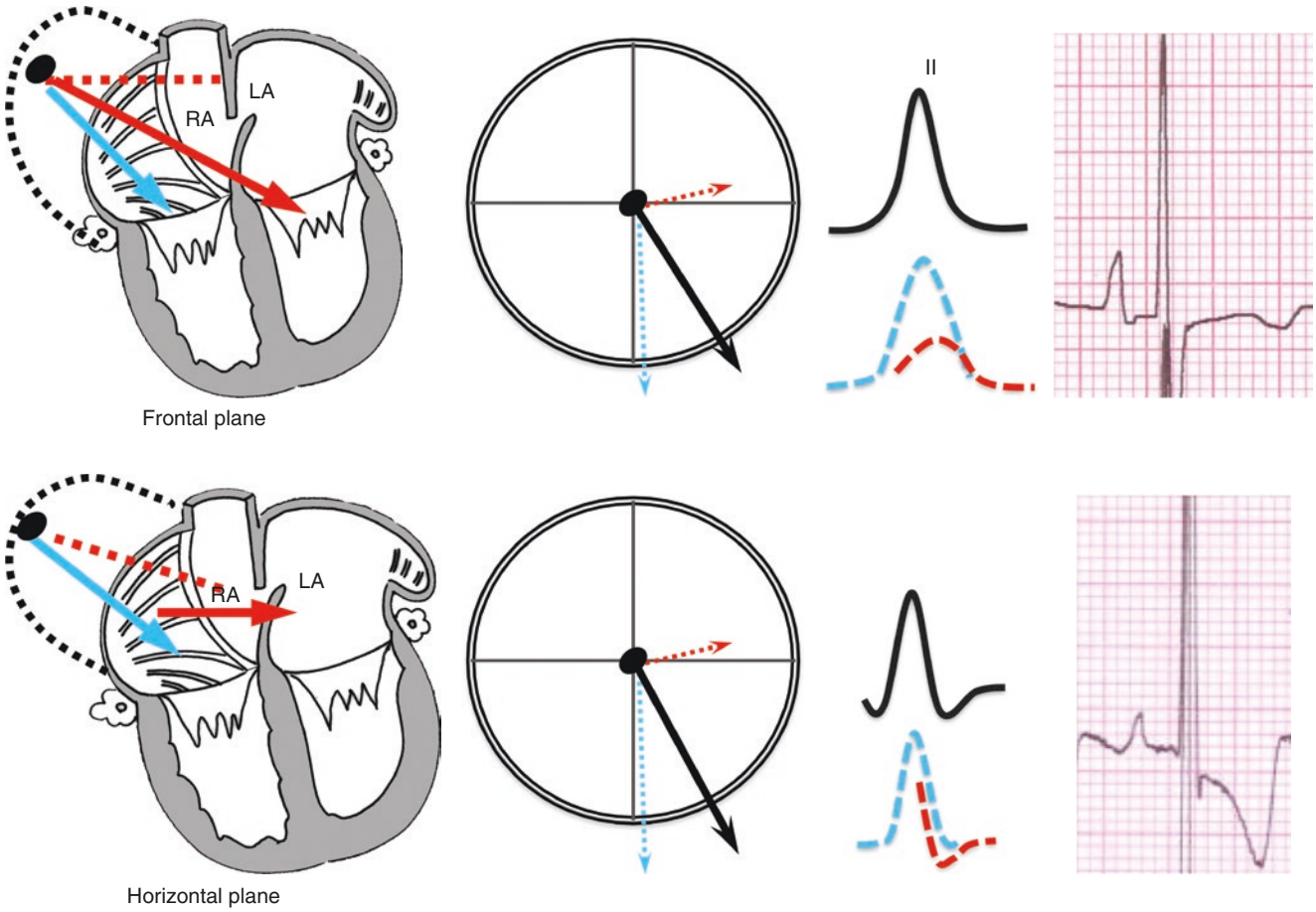

Fig. 3.2 Right atrial enlargement

In left atrial enlargement (Fig. 3.3), you can see an increase of the voltage and the duration of the second component of atrial activation. Especially in smaller infants, the P wave distortion can be particularly evident in V1. The main criteria of left atrial enlargement are:

- Bicuspid P wave (M shaped) with the second component increased in amplitude and lasting more than 90 ms in children and 110 ms in young adults, clearly visible in leads II, III, and aVF.
- In V1, the portion of the wave relative to the left atrium (the terminal part) is more electronegative (>1 mm).

Chest deformities such as pectus excavatum can also alter the morphology of P waves, especially in the right precordial leads. This can sometimes be bizarre, as in pathological conditions marked by right atrial enlargement, for instance, Ebstein's anomaly or pulmonary atresia with intact ventricular septum. Staying with V1, the negative component that is attributed to the left atrial overload has a low specificity and, if there are no clinical, pathological murmurs or other signs of left ventricular hypertrophy, is therefore not pathognomonic.

The polarity of the P wave can reveal the visceral-atrial situs: in situs solitus and levocardia, the axes of P and QRS are concordant in the lower left quadrant, while in situs inversus and dextrocardia, they point down and to the right (atrial situs usually follows the visceral situs) (Figs. 3.4, 3.5, and 3.6).

It is therefore a good sign that in levocardia, the right atrium is on the right and in dextrocardia it is on the left: in fact, the "squinting" between the P wave and the QRS axes shows a discrepancy between situs and heart position, such as dextrocardia in situs solitus (Table 3.2; Fig. 3.7). This can be an important clue as this pattern may be an indicator of complex heart defects not obvious in dextrocardia with situs inversus. When faced with abnormal P wave polarity within the framework of complex congenital heart disease, you should search for a heterotaxy condition, namely, ambiguous sites: the sinus node is a "right-hand" structure, so a right atrial isomerism can have two symmetrically located and competitive sinus nodes (Fig. 3.8). Conversely, with a left isomerism, the sinus node may be missing and replaced

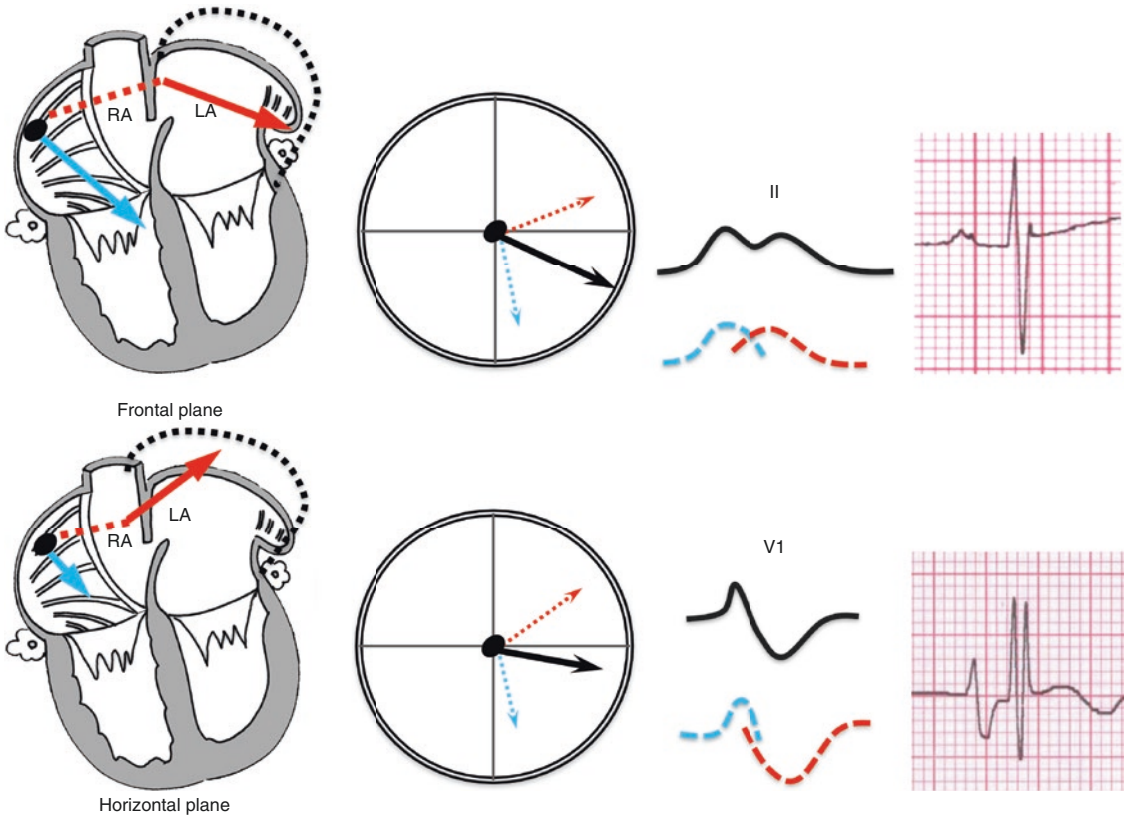

Fig. 3.3 Left atrial enlargement

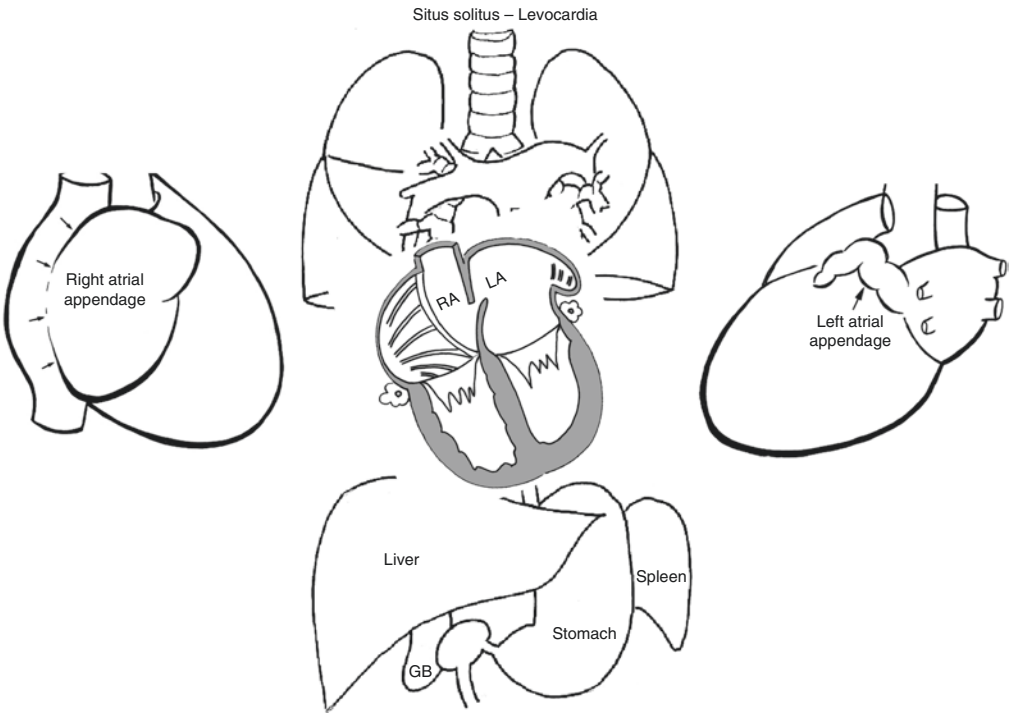

Fig. 3.4 Cardiac and visceral situs

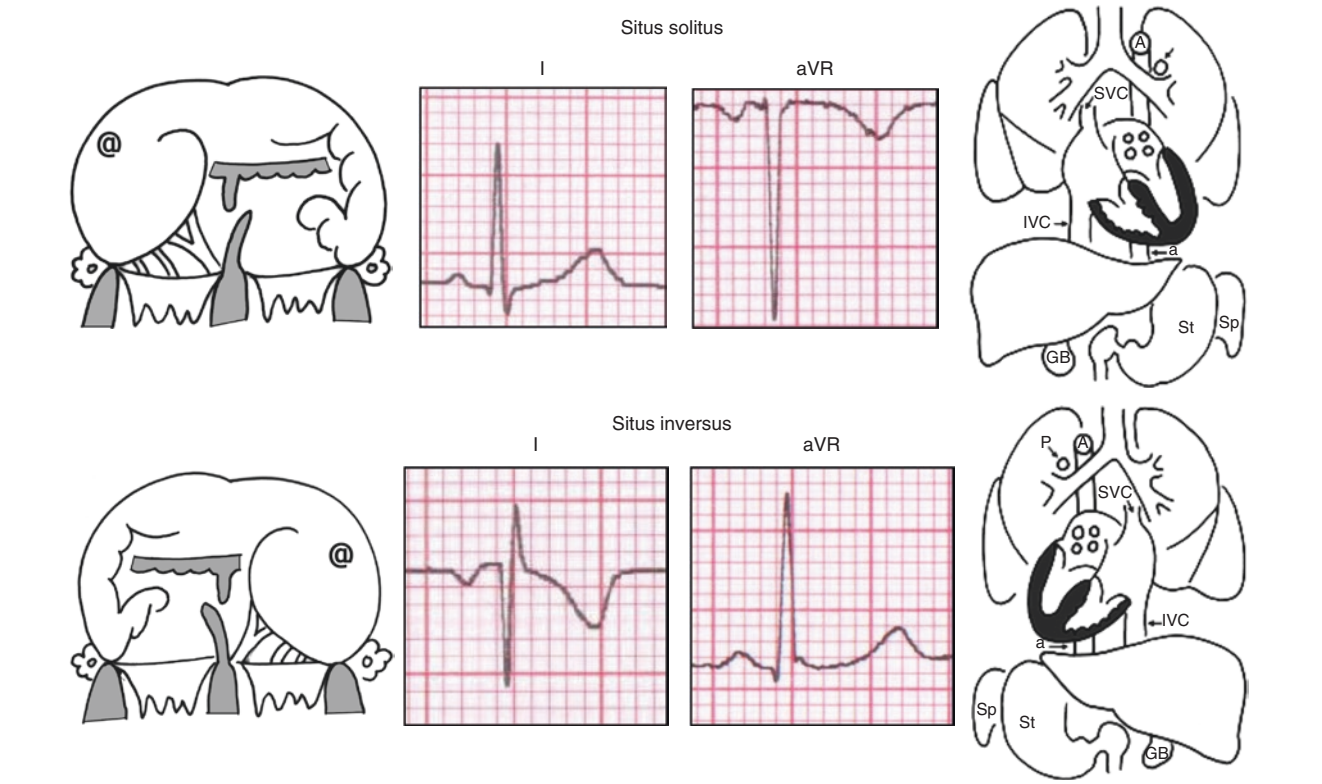

Fig. 3.5 ECG in situs solitus and situs inversus

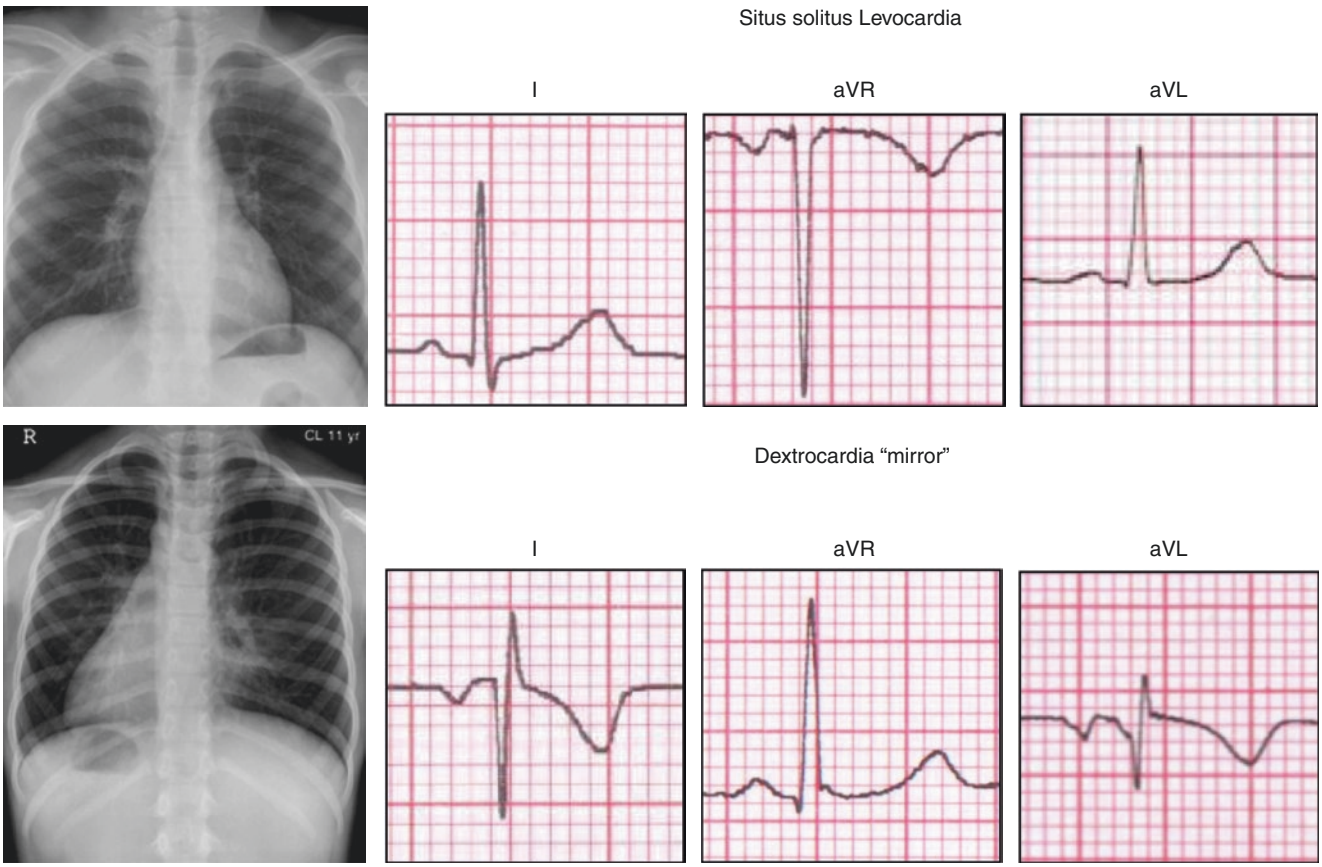

Fig. 3.6 ECG in levocardia and dextrocardia

Table 3.2 Clinical-instrumental discordances

Finding	Discrepancy	Deduction
Nrl QRS axis	Situs inversus	Complex CHD
RA enlargement	Nrl RV/small	PAIVS, Ebstein, TA
LA enlargement	Nrl LV on ECG	MS, AS, HCM, RCM
Northwest axis	Absent RV overload	Noonan, Steinert
Biatrial enlargement	Nrl ventricles	RCM
Low voltage in limb leads	Nrl precordial voltages	Myocarditis, ARVC
Low voltages	Asthenic habitus	Eating disorders
Low voltages	Athletic habitus	ARVC, DCM, HCM
Low voltages	Nrl or ↑ cardiac wall thickness	Amyloidosis, myocarditis, ARVC, CP
Left axis deviation	Newborn period	Preterm birth, TA, AVC, PAIVS, Steinert, Ebstein
Nrl QRS	Discordant T wave	Overload, ischemia, pre-excitation, electric memory
Negative T waves V4–V6	Apparently normal echo	Apical HCM, papillary muscle hypertrophy, LVNC
RVH	Nrl pulmonary pressures	CCTGA, systemic RV (Mustard/Senning)
Intermittent pre-excitation	Long PR or AV Block	CCTGA
RBBB	Left axis deviation	LVH, ToF, Ebstein
Pseudonecrosis Q waves High R waves in V1–V2	Nrl ST-T No other signs of RVH	Duchenne disease

CHD congenital heart disease, *RA* right atrial, *RV* right ventricular, *PAIVS* pulmonary atresia with intact ventricular septum, *TA* tricuspid atresia, *LA* left atrial, *LV* left ventricular, *Nrl* normal, *MS* mitral stenosis, *AS* aortic stenosis, *HCM* hypertrophic cardiomyopathy, *RCM* restrictive cardiomyopathy, *ARVC* arrhythmogenic right ventricular cardiomyopathy, *RVH* right ventricular hypertrophy, *TR* tricuspid regurgitation, *CCTGA* congenitally corrected transposition of the great arteries, *AVB* atrioventricular block, *RBBB* right bundle branch block, *LVH* left ventricular hypertrophy, *ToF* tetralogy of Fallot, *CP* constrictive pericarditis, *LVNC* left ventricular non-compaction cardiomyopathy

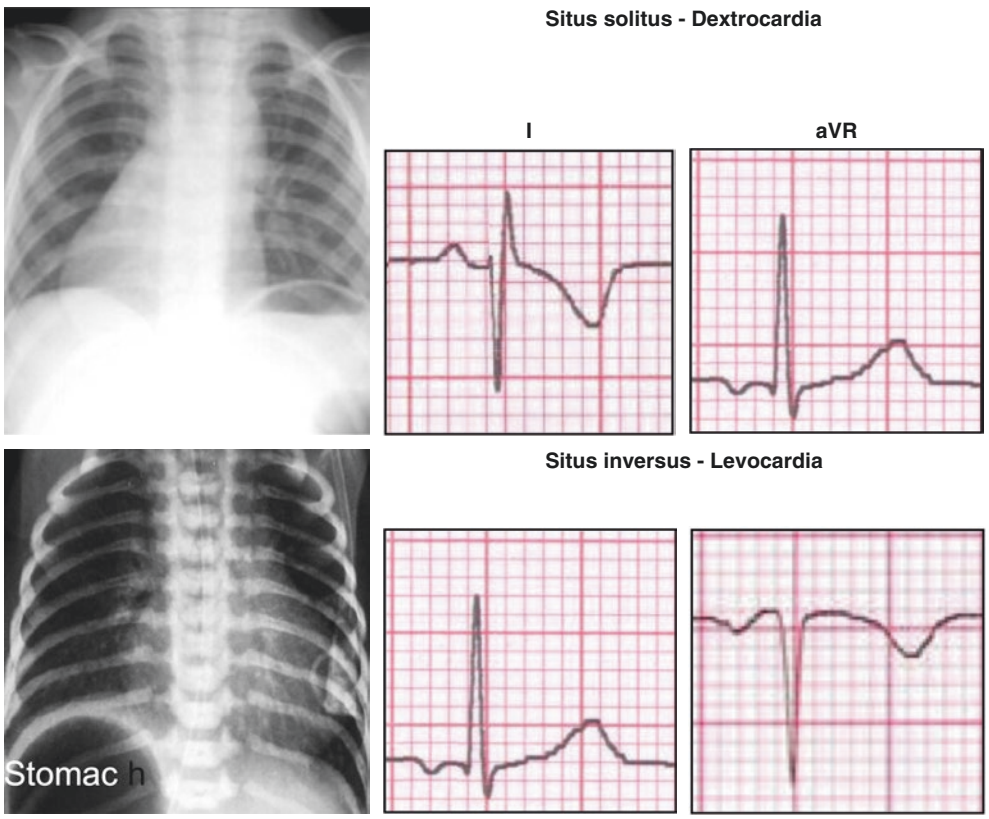

Fig. 3.7 Discordance between situs and cardiac apex

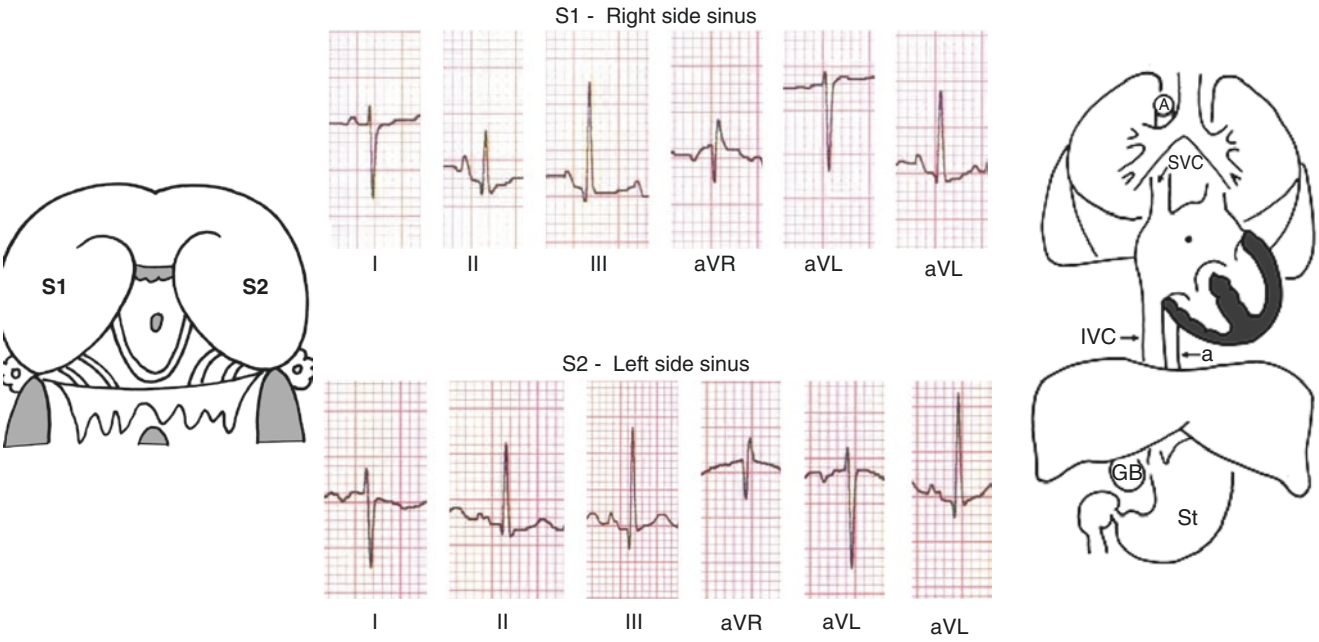

Fig. 3.8 Right isomerism

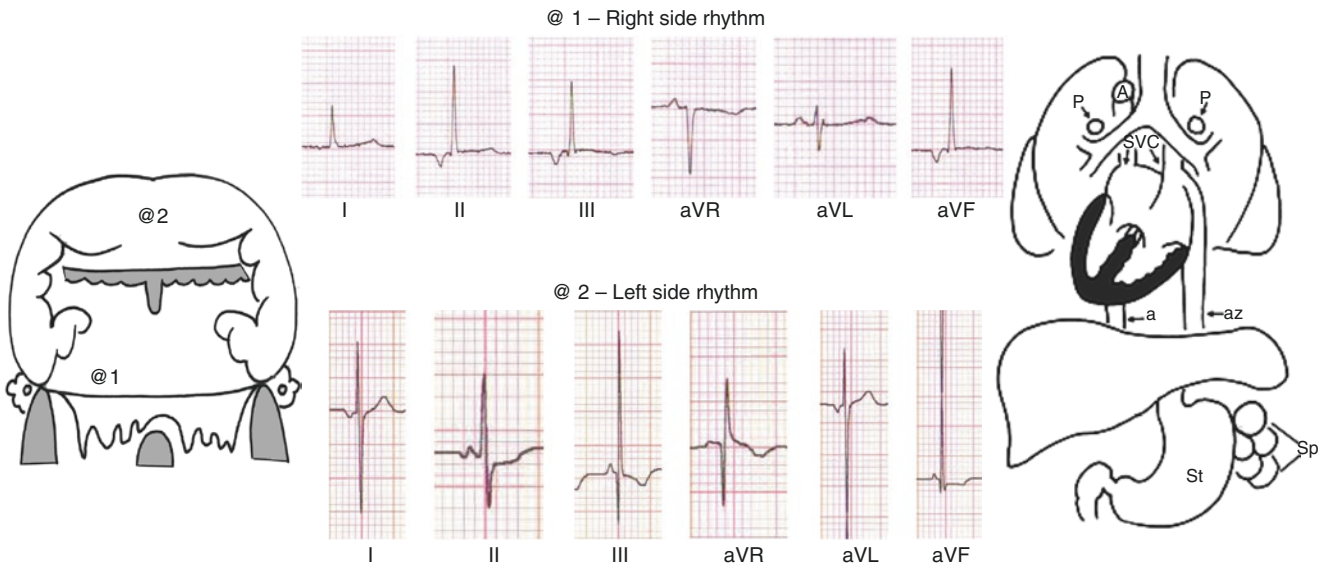

Fig. 3.9 Left isomerism

by ectopic and migrant pacemakers (Fig. 3.9). There may also be two AV nodes in the right isomerism, the so-called twin AV node, prone to the unusual node-to-node reentry, distinctive because of the possibility of finding two supraventricular tachycardias in the same patient, each with a different QRS morphology [2], since both AV nodes can be traveled alternately in an anterograde or retrograde direction (Fig. 3.10).

Sinus or phasic (respiratory) arrhythmia is common and normal; during inspiration, the heart rate increases. By definition, the morphology of the P wave should not change in the two phases; however, even in respiratory periodicity, it is possible to observe two distinct P wave morphologies. Due to the augmented vagal tone during expiration, the sinus node can be slightly inhibited with the escape of alternative atrial foci, located close to the sinus or more distally in the lower right atrium (the so-called coronary sinus rhythm); the P wave morphology therefore changes accordingly.

Note that the lower the heart rate, the more acceptable a physiological dualism of the atrial rhythm, while at high

frequencies, the hypothesis of automatic atrial tachycardia should always be considered. A low rhythm of the coronary sinus is not rare, and in this case, the atrial vector follows a direction from bottom to top, and the inferior leads will record a negative P wave. The pathological significance therefore depends on the heart rate, and according to age and circadian persistence, you must rule out an automatic atrial tachycardia. The low atrial rhythm is more common in the presence of dilated coronary sinus, usually due to a persistent left superior vena cava (PLSVC) draining into it. This vascular anomaly can be found in various congenital heart diseases, from a simple bicuspid aortic valve to the more complex such as tetralogy of Fallot. Therefore, when faced with a low atrial rhythm, the patient's medical history and objective examination must be more scrupulous than normal, and in the event of doubt, an echocardiogram is also warranted (Fig. 3.11).

Regarding heart rate, the myth of the declining rate from fetus to old age (heart rate decreases from womb to tomb) should be debunked since the cardiac sympathetic nervous system can mature after the parasympathetic nervous system, justifying a certain bradycardia (Table 1.2). So, during examination, a newborn can have a heart rate of around 80 bpm and a 6 year-old child a rate of up to 120 bpm. Consistently low heart rates and those unresponsive to stimuli or activities (a heart rate of 60–80 bpm of a baby while feeding or crying is unacceptable) should trigger a warning of long QT with 2:1 AVB (Fig. 3.12a), blocked bigeminal PAC (Fig. 3.12b), and congenital atrioventricular block (AVB)—associated or not with structural heart disease (Fig. 3.12c). Neonatal bradycardia may be due not only to primarily cardiac situations, such as neurological conditions (hypoxia, intracranial hypertension) or endocrine-metabolic conditions (hypothyroidism). Channelopathies such as Brugada syndrome or related to ion channels other than those of sodium may occur with undue sinus bradycardia. In general, fever increases the HR by about 10 beats per Celsius degree, and physiological sinus tachycardia is expected not to exceed 220 bpm. This concept only partially helps in the management of the newborn/infant that reaches the emergency room in a critical condition and tachycardia. In such cases, the differential diagnosis includes sepsis, heart failure, and paroxysmal tachycardia.

The chapter on arrhythmias will go into more detail regarding the morphology and rate of P waves. It will be the hunt for something ephemeral, because in a certain way, the P wave is a *femme* who runs away and hides in the T or in the QRS with sophisticated mimicry. It's a breathtaking thriller, the rallying cry being "Cherchez la P."

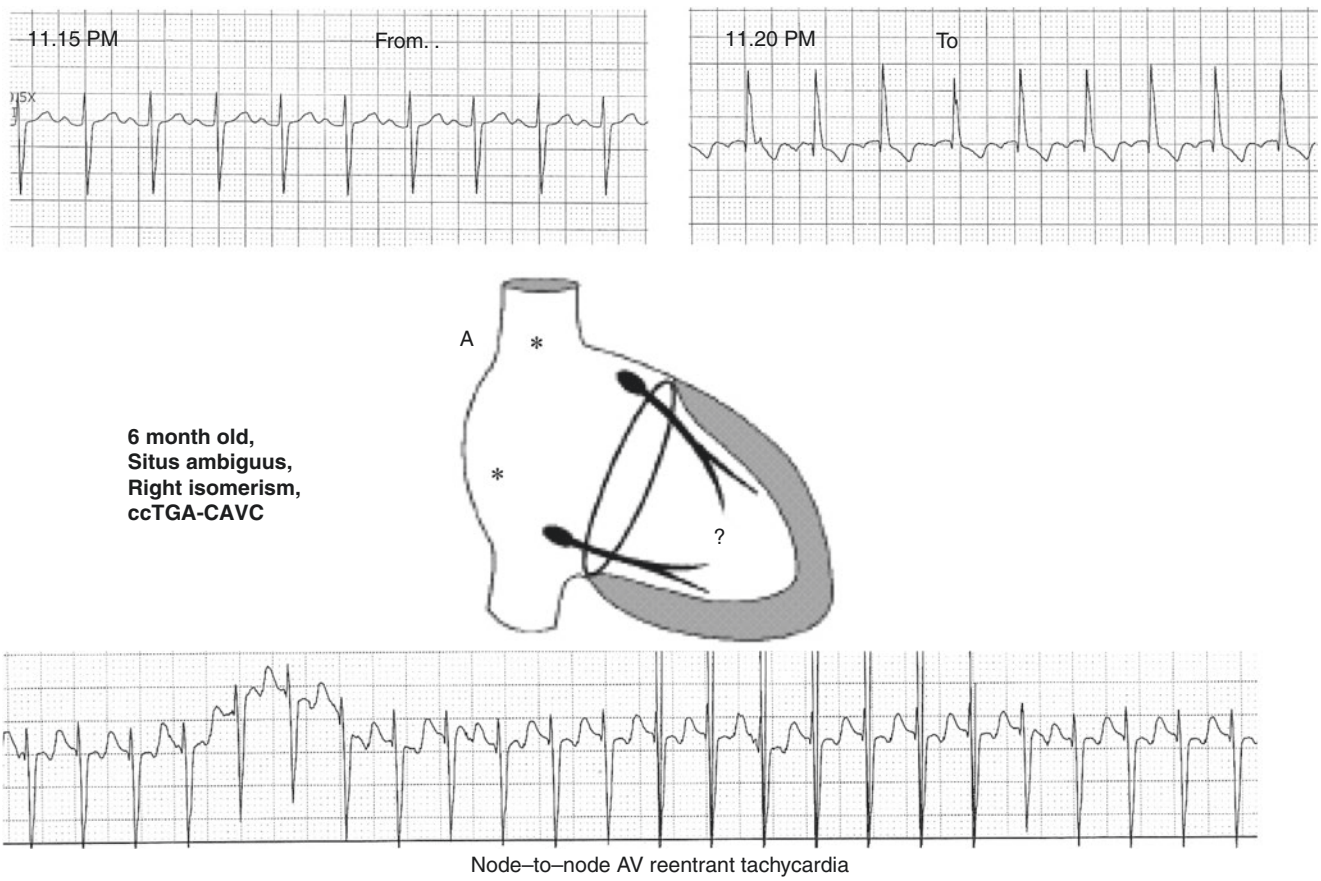

Fig. 3.10 Twin AV node in complex congenital heart disease

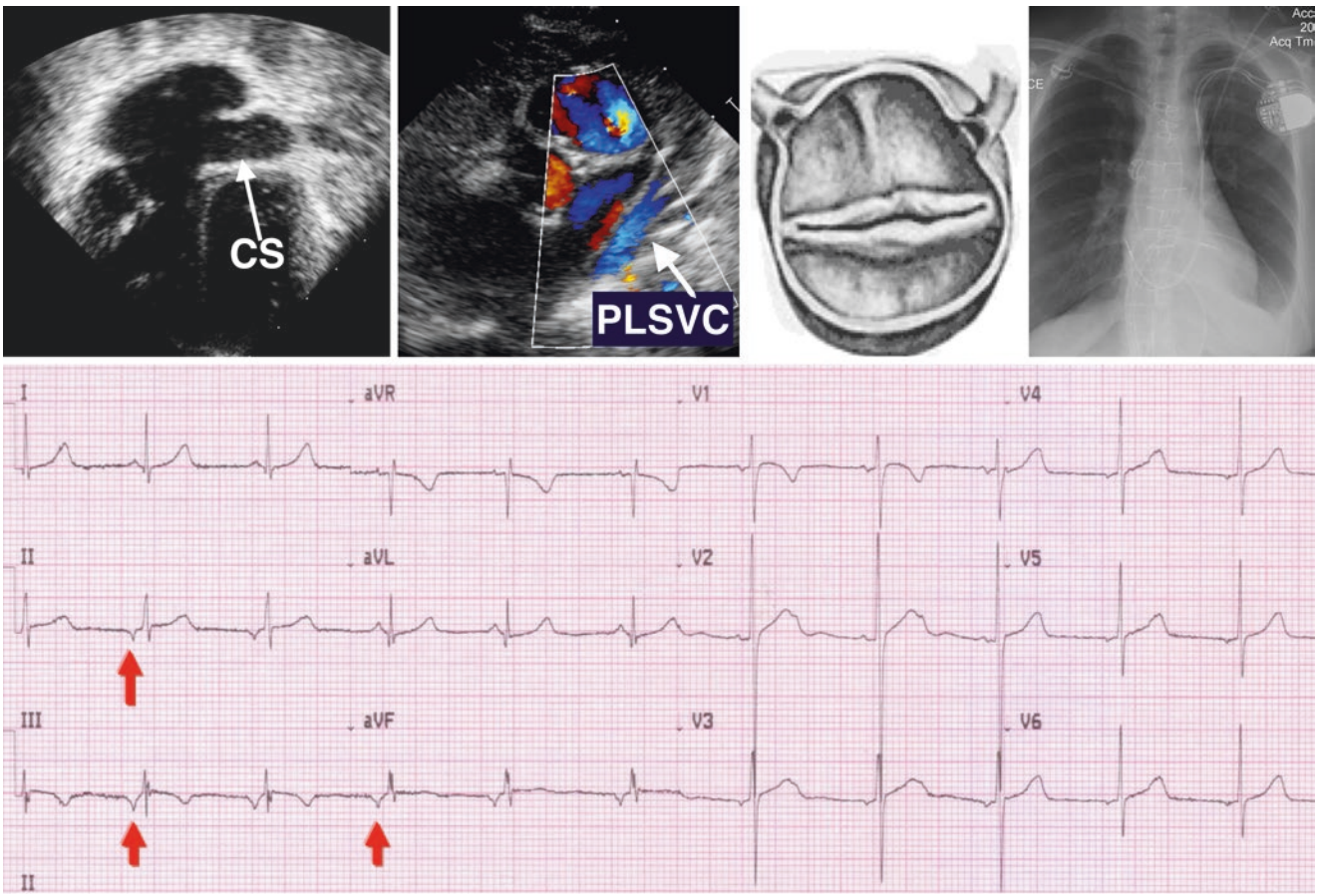

Fig. 3.11 Persistent left superior vena cava. The chest X ray shows the unusual course of the PM catheters via a PLSVC

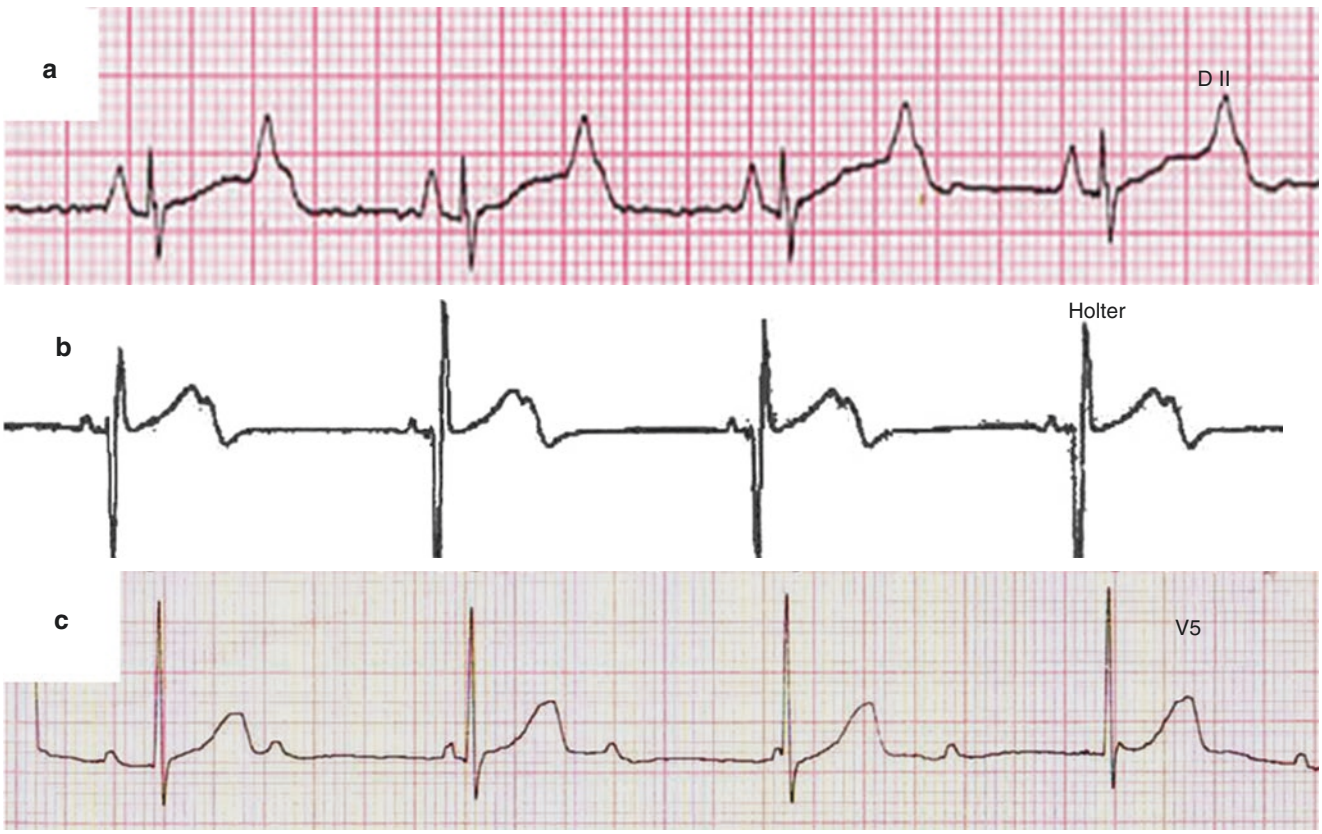

Fig. 3.12 Severe neonatal bradycardia

3.2 PR Interval

The PR interval depends on age and on the autonomic nervous system, and therefore a newborn usually but not necessarily has a shorter PR than a child's PR. To make a diagnosis of pre-excitation, you should see a clear delta wave. Although pre-excitation is in the great majority of cases an isolated condition, it sometimes accompanies congenital heart disease and storage diseases, a concept which recommends an echocardiogram (Fig. 3.13). The accessory pathway can itself be defined as the smallest congenital heart disease: it is an anatomically distinct structure—although invisible to the naked eye—that can be ablated permanently with a transcatheter procedure. Before the advent of radio-frequency catheter ablation (RFCA), surgery was the only way to permanently remove an accessory pathway, like any congenital heart disease.

Pre-excitation syndromes include Mahaim fibers, meaning complex AV connections (atrio-fascicular, atrioventricular nodal, etc.), where the resting ECG may show a normal PR, a nuanced pre-excitation, and an rS pattern in lead III.

A short PR interval is also compatible with physiological and harmless low atrial rhythm (where the rhythm arises in a site close to the AV node) or severe storage disease and hypertrophic cardiomyopathy phenotype (Pompe, Danon, and Fabry disease, mitochondrial disease, PKRG2 etc.). In the newborn, the normal PR ranges from 80 to 150 ms, in children and adolescents from 120 to 200 ms. Ten percent of young people can have a PR at the upper limit for their age in the absence of heart disease; an intermittent second-degree AV block can be found in 5% of the young, especially in athletes, and is physiological during the night, while if it occurs in the waking state or during activity, it requires investigation.

At the normal rate for a child (100–150 bpm), the PR interval can range between 80 and 120 ms but occasionally can even reach 150 ms to 180 ms. During 24-h Holter

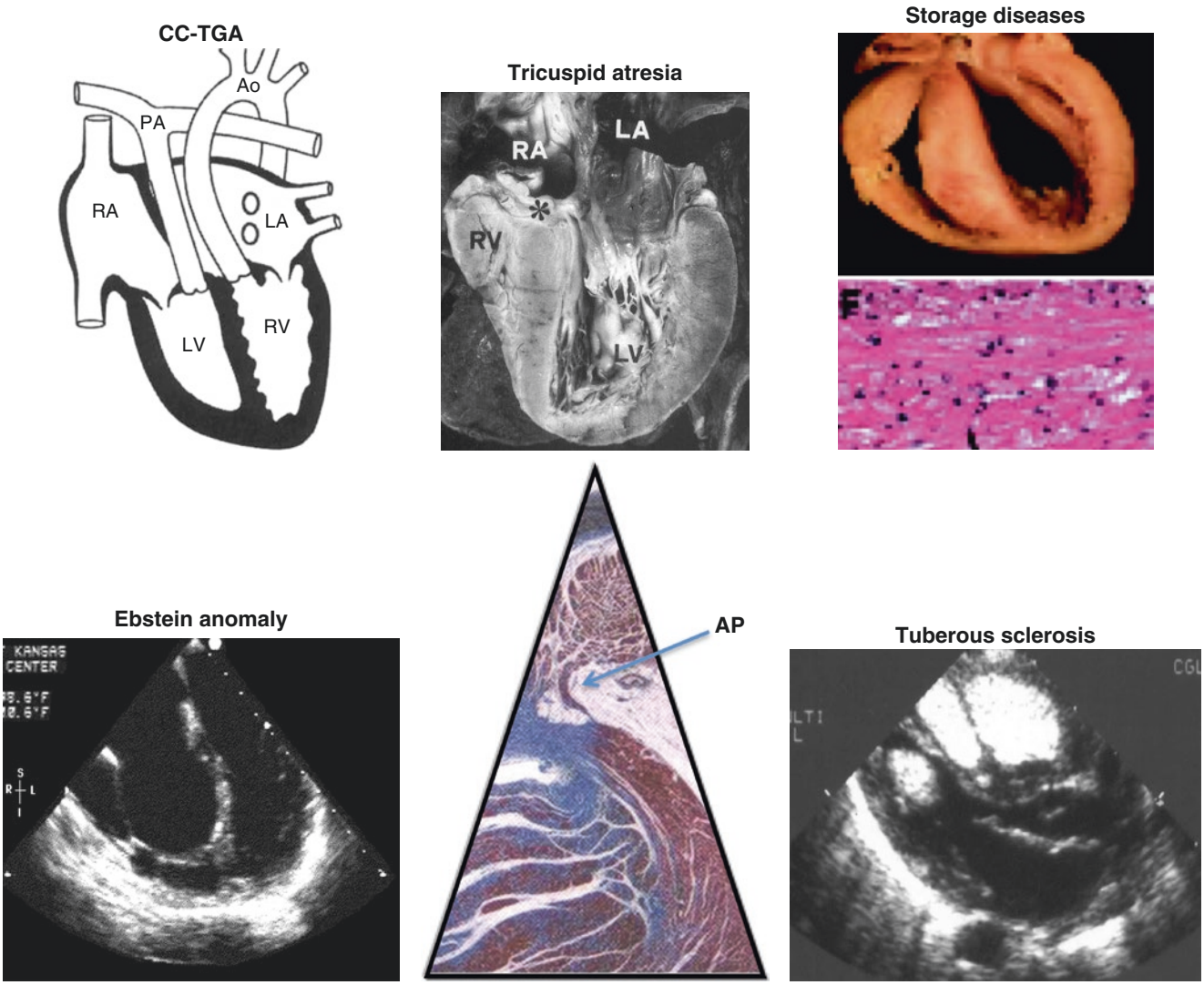

Fig. 3.13 CHD and WPW

monitoring, it is not uncommon to see values around 200 ms. An enhanced vagal tone may explain this phenomenon in the presence of concomitant bradycardia; conversely, in the case of sinus tachycardia or during exercise, the lengthening of the PR cannot be justified by the vagal tone (Table 2.1).

A long PR associated with right bundle branch block (RBBB) is suggestive of ostium primum-type atrial septal defect (ASD) in the setting of endocardial cushion defects (atrioventricular canal) but can also be compatible with channelopathies such as Brugada syndrome. However, in causing first-degree AVB, benign causes are much more numerous than malignant ones, and medical history and context are important. First- and second-degree AVBs, up to complete block, can be explained by electrolyte imbalance and enhanced vagal tone but are nonetheless compatible with congenitally corrected transposition of the great vessels (CCTGA or L-TGA), atrioventricular canal (AVC), neuromuscular diseases, rheumatic and Kawasaki disease, borreliosis, and laminopathies. AVB 2:1 is typical of congenital long QT syndrome. In fact, prolonged repolarization can inhibit AV 1:1 conduction; the beta-blocker used in this setting resolves the block in a seemingly paradoxical way, reducing the sinus rate and allowing the 1:1 conduction, but without shortening the QT interval duration (Table 3.3). Even bradycardia due to bigeminal atrial extrasystole with "blocked" atrial impulses can surprisingly be treated with beta-blockers (Fig. 3.12b). In pregnant women with systemic lupus erythematosus, Sjögren's syndrome, or other immunopathies, the fetal AV node can be damaged by the maternal antibodies, and when this happens, it usually results in complete AVB and very rarely in a lesser degree of AVB. Among the causes of acquired PR lengthening, we should not forget rheumatic disease, which represented a Jones minor criterion (now downgraded to a sign suggestive of recent streptococcal infection) [3]. A transient QT prolongation can also be very occasionally seen in the course of rheumatic disease.

Table 3.3 Atrioventricular block (AVB)

	Finding	Setting
First-degree AVB	• Newborn: Nrl PR 70–160 ms • Childhood and adolescence: Nrl PR 80–180 ms	Nrl variant; ostium primum ASD; inlet VSD; Ebstein; CCTGV; myocarditis[a]; NMD; electrolyte imbalances; hypothyroidism
Second-degree AVB	• **Mobitz 1** (Luciani-Wenckebach): progressive ↑ PR up to blocked atrial beat (P) • **Mobitz 2**: periodic block of atrial pulse (P)[b]	• Nrl in children if at rest or during sleep; athlete's heart • Nrl only if temporary (sleep, i.e., the first days post-cardiac transplantation)
Third-degree AVB	• AV dissociation	Congenital[c]; postsurgical; CHD; VSD/ASD s/p percutaneous closure; myocarditis; RF; MD; NMD; Lev-Lenègre disease

NrL normal, *ASD* atrial septal defect, *VSD* ventricular septal defect, *CCTGA* congenitally corrected transposition of the great arteries, *NMD* neuromuscular diseases (Duchenne, Steinert, Kearns-Sayre, Emery-Dreifuss, laminopathies), *CHD* congenital heart diseases (20% in CCTGA), *RF* rheumatic fever, *MD* metabolic diseases (carnitine deficiency, glycogenosis)

[a]Rheumatic, viral, Lyme disease, Chagas

[b]If AVB 2:1, you cannot tell if type 1 or 2; advanced AVB if ≥3:1

[c]Maternal lupus, maternal systemic sclerosis, maternal connective tissue disease

3.3 Delta Wave

It has already been said that in children, the PR interval can be very short and that to make diagnosis of pre-excitation, it is necessary to see delta waves, indicative signs of abnormal atrioventricular connection. A short PR without a clear delta wave is sometimes attributed to the Lown-Ganong-Levine syndrome but only when supraventricular tachycardia (SVT) is present. For some cardiologists, LGL does not exist (or exists as an enigmatic and eponymous palindromic acronym similar to Wolff-Parkinson-White [WPW] syndrome) and is only the appearance of accelerated conduction in the nodal pathway. There are leads, especially the lower ones, and in particular lead III, where the initial QRS is sometime slurred, creating the impression of a delta wave. This "pseudo-delta" aspect in a single lead or in less than three leads should not be reported as pre-excitation and should be seriously taken into account only in the case of frank clinical episodes of paroxysmal tachycardia.

Initially, Rosenbaum [4] distinctly divided WPW patterns into type A and type B. The first is caused by accessory pathways located on the left and shows positive delta waves and concordantly positive QRS in all precordial leads, with V1–V3 reminiscent of the right bundle branch block (RBBB). Type B, on the contrary, is due to accessory pathways located on the right and is shown with negative delta waves and negative or nearly negative QRS in the right precordial leads, while the rest of the precordial leads can suggest a left bundle branch block (LBBB). The rule is—as in ventricular tachycardia (VT)—that everything that arises in a ventricle reveals itself with the appearance of an opposite side branch block (e.g., if the accessory pathways or the ventricular focus of a VT is located in the right ventricle, the ECG will show an LBBB and vice versa).

There are labyrinthine algorithms to infer the location of the accessory pathway based on delta wave silhouette; this noninvasive divination is not at present mandatory because in the ablation setting, even the most reliable prediction can be proved wrong and does not avoid the need for scrupulous mapping which not infrequently reveals surprises, for example, multiple accessory pathways: the electrophysiologist should be prepared for any location of the abnormal connection, right and left, and this is particularly true in adults with straightforward indications for RFCA. In children, instead, knowing in advance that a pathway is on the left can be encouraging, indicating the improbability of a complication such as AVB. It does, however, give you time to decide whether to use the transeptal or retrograde aortic approach; the suspicion of an anteroseptal pathway could indicate a procedure with cryoenergy or even postponing the intervention to a less risky age. Moreover, the finding of a positive delta wave in aVR, especially if associated with a negative delta wave in lead II, is suggestive of a posteroseptal accessory pathway, near the coronary sinus or in the afferent veins [5]. These rather infrequent connections have also been described in pediatric patients and are characterized by a high rate of recurrence of tachycardia after ablation.

3.4 QRS Complex

The QRS axis in the frontal plane represents the mean vector of ventricular depolarization. From the newborn to the elderly, there is a ventricular axis shift progressively from right to left (Fig. 3.14). This notion is so true that in the infant, a marked left axis deviation is pathognomonic of structural heart disease such as AV canal or tricuspid atresia. In adults, the normal axis is between −30° and +110°. The QRS axis guides the diagnosis of specific congenital heart disease and can corroborate a diagnosis of ventricular overload, when it is too oriented toward the cardiac chamber suspected of being hypertrophic or dilated (Table 3.4). In hypertrophic cardiomyopathy, for example, a left axis deviation may be the first and the only ECG clue. Interestingly, in Noonan syndrome, the right axis deviation may have nothing to do with pulmonary stenosis and thus of right ventricular hypertrophy, and the strain of the left ventricle can anticipate the echocardiographic recognition of the ventricle hypertrophy typical of this genetic condition (this is a case of electric premonition, instead of the better known and less disturbing electrical memory). It is also possible in Noonan syndrome to see dominant S waves in all precordial leads. A personal observation requiring confirmation is the prevalence of marked right (or left) axis deviation in children with myotonic dystrophy of Steinert, regardless of the presence of echocardiographic abnormalities.

The QRS axis on the horizontal plane is described by identifying the transition from V1 to V6 or the one where the R wave exceeds the S wave. The electrical axis is stable at around the age of 12 and will then change only for pathological reasons; after the age of 3 years, the R/S ratio in V1–V2 is

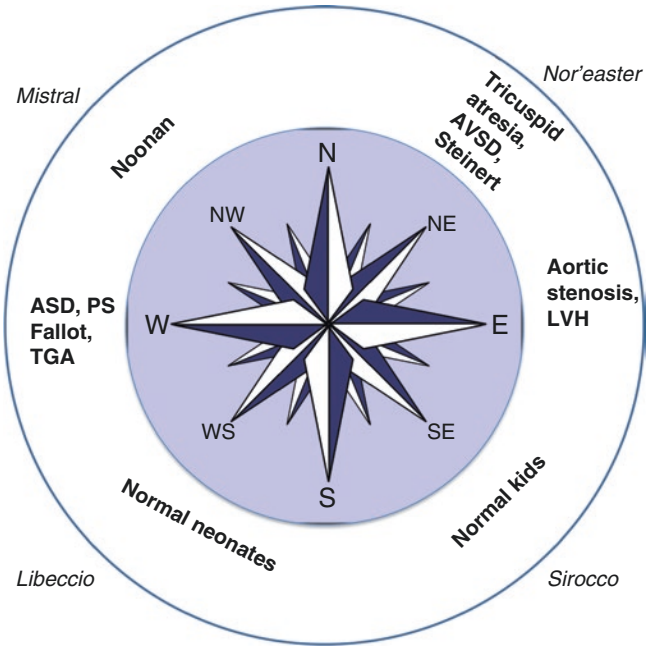

Fig. 3.14 The Rose of the Winds of the cardiac axis

Table 3.4 ECG synopsis of the congenital heart diseases

	Anatomy	Signs
ASD	RA and RV dilatation	RBBB, negative T waves V1–V5 (overshoot) right axis dev., "crochetage"
VSD-PDA	Biventricular hypertrophy	Katz-Wachtel sign, LAE
AVC	Abnormal conduction system	LAH, RBBB, I° degree AVB, LAE
PS	RVH, RA dilatation	Peaked R V1–V3, deep S in V6, positive T V1 (7 days to 7 years), right axis dev.
AS-AoC	LVH, LA dilatation	LV overload, LV strain, LAE
Fallot TGA PA + VSD	RVH, VSD; cyanosis; postsurgical extensive scarring	Peaked R waves in right precordial leads Deep S waves in left precordial leads Inferior Q waves (postsurgical RBBB, VT)
Ebstein	Massive RA-RV dilatation—anomalous AV pathways	Polyphasic RBBB, delta wave, massive RA enlargement
CCTGA	Ventricular and conduction system inversion	Q wave in V1, absent Q wave in V6, AVB, delta wave, LAE
TA	Abnormal conduction system	LAH, left electrical prevalence
PAIVS	RV hypoplasia, RA dilatation	Small R wave in right precordial leads, RA enlargement, left electrical prevalence
ALCAPA	Anomalous left coronary artery from the pulmonary artery, anterolateral ischemia	Q wave + anteroseptal and lateral ST elevation / depression
HLHS	RVH and RV dilatation	Right electrical prevalence Poor left electrical forces
UVH left type	LVH	Peaked R waves in left precordial leads, diffuse isodiphasism
Metabolic diseases	LVH, pre-excitation	LV or biventricular overload, short PR, delta wave
Mustard/Senning/Fontan	Transposition, UV, extensive scarring, systemic right ventricle, fibrosis Single ventricle, atriomegaly, extensive scarring, fibrosis	Sinus node dysfunction Atrial flutter, AF, VT

ASD atrial septal defect, *VSD* ventricular septal defect, *PDA* patent ductus arteriosus, *AVC* atrioventricular canal, *PS* pulmonary stenosis, *AS* aortic stenosis, *AoC* aortic coarctation, *TGA* transposition of the great arteries, *PA + VSD* pulmonary atresia with ventricular septal defect, *CCTGA* congenital transposition of the great arteries, *PAIVS* pulmonary atresia with intact ventricular septum, *ALCAPA* anomalous left coronary artery from the pulmonary artery, *HLHS* hypoplastic left heart syndrome, *UVH* univentricular heart, *Mustard/Senning* eponyms of transposition of the great vessel palliations, *Fontan* eponym of palliation for single ventricle physiology CHD, *RA* right atrium, *RV* right ventricle, *LAE* left atrium enlargement, *LAH* left anterior hemiblock, *RBBB* right bundle branch block, *AVB* atrioventricular block, *RVH* right ventricular hypertrophy, *LVH* left ventricular hypertrophy

usually less than 1, and if not, we must consider dextrocardia, right ventricular hypertrophy, Duchenne muscular dystrophy, ventricular pre-excitation, hypertrophic cardiomyopathy, etc. This last disease can also show up with strange R wave regressions from V2 to V6 (the "arrow sign"), a jump of R from V1 to V2, and the absence of Q in V6 [6]. If you can exclude the abovementioned conditions, including a posterior myocardial infarction (very rare in the young), the presence of dominant R waves in the right precordial leads can be defined as a physiological counterclockwise rotation which can affect some 20% of children up to 8 years and 10% of adolescents under 16 years; an R/S > 1 in V2 is located in less than 1% of normal adults. On the contrary, in clockwise rotation, the transition moves toward the left precordial leads, with poor progression of the R wave from V1 to V6. This pattern can coexist with left axis deviation in the frontal plane, from mild to frank left anterior hemiblock. This adult pattern may also be defined as a normal variant but only after excluding pathological conditions, first of all left ventricular hypertrophy. In ventricular pre-excitation syndromes, transition of the QRS in the precordial leads can help to infer the site of electrical bypass. An early transition (in the right precordial leads V1–V2) indicates left-sided accessory pathways—the delta wave and the QRS point in the opposite direction to the site of the connection—while a late transition (from V3 to V6) suggests right accessory pathways. Also in the case of outflow tract tachycardia, an LBBB with transition before V2 suggests the rarer form arising from left ventricular outflow tract (or aortic cusp) (LVOT), instead of the well-known form originating from the right ventricular outflow tract (RVOT).

Unusual heart rotations and thoracic deformity may also warrant abnormal Q waves in the absence of any clinical evidence or pathological imaging. Moreover, a bizarre axis must always be confirmed by a repeat ECG to exclude malpositioning of the limb leads.

In general, the ECG is not particularly sensitive and specific in the recognition of cardiac hypertrophy due to pressure overload, especially in the left ventricle. In fact, using only the voltage criteria, for left ventricular hypertrophy, the sensitivity of the ECG is below 35%, while the specificity is about 85%. In hypertrophic cardiomyopathy, instead, the ECG sensitivity increases because it is not only the quality of the

hypertrophy that counts but also its quality, as demonstrated by certain mutations of troponin T where despite normal cardiac thicknesses on echo, the ECG is already visibly altered.

There are also hypertrophic cardiomyopathies that do not meet the voltage criteria for hypertrophy due to the presence of fibrosis or to enlargement of the QRS, which tends to reduce the peak voltage (the area of the R "triangle" remains the same). Volume overload may be seen on the ECG before the pressure overload, and sometimes severe aortic regurgitation is seen better than a similarly severe aortic stenosis; this is explained by the fact that the radius of the left ventricle has more impact than the wall thickness in the calculation of ventricular mass. In general, repolarization is more important than depolarization in diagnosing overload; the R wave can jump off the page in healthy and thin young subjects, while there are severe aortic stenosis that do not fulfill the Sokolow criteria for left ventricular hypertrophy [7] (Table 3.5). In the case of overload, the right ventricle is more eloquent; putting together different signs (R and T in V1, S in V6, right axis deviation, right atrial enlargement), you achieve an acceptable sensitivity and specificity in the diagnosis of hypertrophy or specific congenital heart disease (Table 2.1). The full-term newborn shows right prevalence with prominent R waves in the right precordial leads and reciprocal S waves in the left precordial leads. In the anatomical sense, right dominance is reached at the 31st week of pregnancy, and it is not surprising to see a left ventricular dominance in premature infants 14.

The Q wave, dreaded in adults for its ischemic significance, is more tolerated in children where, on the contrary, its absence in leads where it is usually normal to see it is worrying (see lead III and V6). Sometimes, the absence of a Q wave surreptitiously reveals pre-excitation or congenital heart disease or is the only clue of an incomplete LBBB. The Q wave can be quite pronounced, up to 6–8 mm, especially in lead III and left precordial leads. As in adults, its extent in the limb leads may decrease during deep inspiration.

The normal Q wave lasts less than 30 ms and covers less than 25% of the area of the R wave that follows. A Q wave in the aforementioned limits is normal in aVL, while it is less common although not definitely pathological in lead I. An inappropriate presence should trigger a suspicion of ventricular overload; deep and narrow Q waves in the inferior leads are compatible with volume overload (the diastolic overload described by Cabrera) but can also indicate forms not yet severe of overload pressure (hypertrophic cardiomyopathy!), coronary anomalies, and muscular dystrophy.

In hypertrophic cardiomyopathy, it is rare to see pathological or pseudo-infarct Q waves not accompanied by altered ST-T segments; on the contrary, in muscular dystrophy, there

Table 3.5 Ventricular hypertrophy criteria

	ECG findings	98° Centile
RVH	• qR in V1, V4R, V3R • rSR' (if R' > 98° or double the r) • R in V1 > 98° • R in V4R > 98° • R in aVR > 10 mm • S in I or V6 > 10 mm[a] • Positive T wave in V1 between 7 days and 7 years[b] • RV overload pattern • Intrinsicoid deflection in V1, V4 > 0.03 s • Right axis deviation (QRS axis > 160°) • R V1 + SV 5 > 10.5 mm and right axis deviation >120°(Seattle criteria for athletes)	1–3 days 27 mm; 3–7 days 24 mm; 7–30 days 21 mm ≤ 1 month 12 mm; 1 month–1 year 10 mm
LVH	• R in V6 > 98° [R in V6 > V5; wide R in inferior leads] • S in V1 > 98° • Negative and asymmetrical T wave in V5–V6 ± ST-T depression after 48 h of life • Inferior, lateral, and left precordial pronounced Q waves[c] • Secondary: left axis dev., intrinsicoid deflection delay, LA enlargement[d]	1 day 12 mm; 2 weeks 16 mm; 6 month 22 mm; 1 year 23 mm; 10 years 25 mm; 1 day 20 mm; 1 week 10 mm; 6 month 18 mm; 1 year 21 mm
BVH	• RV + LV hypertrophy criteria • Midprecordial lead voltages >98° • Katz-Wachtel sign[e] • Sokolow-Lyon criteria • Cornell criteria	1 day to 6 month R + S > 52 mm; after 1 year R + S > 55 mm S V1 + R V5 or V6 > 35 mm (adults) R aVL + S V 2 > 28 mm in male, >20 mm in female (adults)

RVH right ventricular hypertrophy, *LVH* left ventricular hypertrophy, *BVH* biventricular hypertrophy

[a]Sensitive but not specific (d.d. LVH, left anterior hemiblock)

[b]Up to 10 years: it means overload, not necessarily hypertrophy

[c]Absence of Q wave in V6-I if severe septal hypertrophy

[d]Negative terminal portion of P wave with duration and amplitude >1 mm in V1, P wave duration in II > 0.12 s; [e]wide and isodiphasic RS complex, initially described for large VSD

can be abnormal Q waves in the inferior and lateral leads in the absence of consistent repolarization abnormalities. The absence of the Q wave where it is expected—for example, in V6—can raise the suspicion of L-TGA or left ventricle hypertrophy and may even overshadow ventricular pre-excitation, especially if there are clinical facts such as paroxysmal arrhythmias. In addition, an absent Q wave in V6 is compatible with right ventricle overload, since in this case, the heart is parallel to the frontal plane. In heart disease with pulmonary hypoperfusion, the R wave in V6 is a good indicator of the development of the left ventricle and thus of the degree of pulmonary obstruction, to which it is inversely proportional. Although the prevalence of the right ventricle ends at about 6 months of age, the ECG at this time does not yet have an adult pattern due to the vertical position of the heart favoring the midprecordial leads (V3–V5). It should be remembered that Black people usually show higher voltages in the precordial leads, and the QRS axis derives from the sum of the electrical forces of the two ventricles. In biventricular hypertrophy, the electrical forces can cancel each other out, but when the biventricular hypertrophy is significant, the so-called sign of Katz-Wachtel might be encountered. By this we mean the wide isodiphasism of the midprecordial leads V3–V5. The sum of R + D > 60 mm in V4 is quite specific, as can be seen in extensive ventricular septal defect (VSD).

Children may present a very narrow QRS, and from birth to adolescence, the average QRS duration increases from 50 to 80 ms. The rsR' or rSR' with relatively narrow QRS pattern may be a normal variant and can be visible in up to 20% of young people, but when R' exceeds normal limits (18 mm by 1 year of age and 10 mm thereafter, ±axis >120° after 3 months), it is advisable to rule out an overload of the right ventricle with an echocardiogram (i.e., ASD). Likewise, in these cases, we can talk about bundle branch block even before getting to 120 ms of the QRS duration, the limit considered to be "adult." For psychological reasons (patients and parents read the ECG as the last undisputable word), you should be very cautious before writing the word "block" in a report, even in its attenuated version of incomplete (Fig. 3.15). A true RBBB with wide S waves in leads I V6 should make you think of the existence of congenital heart disease like ASD or other types of volume or pressure overload of the

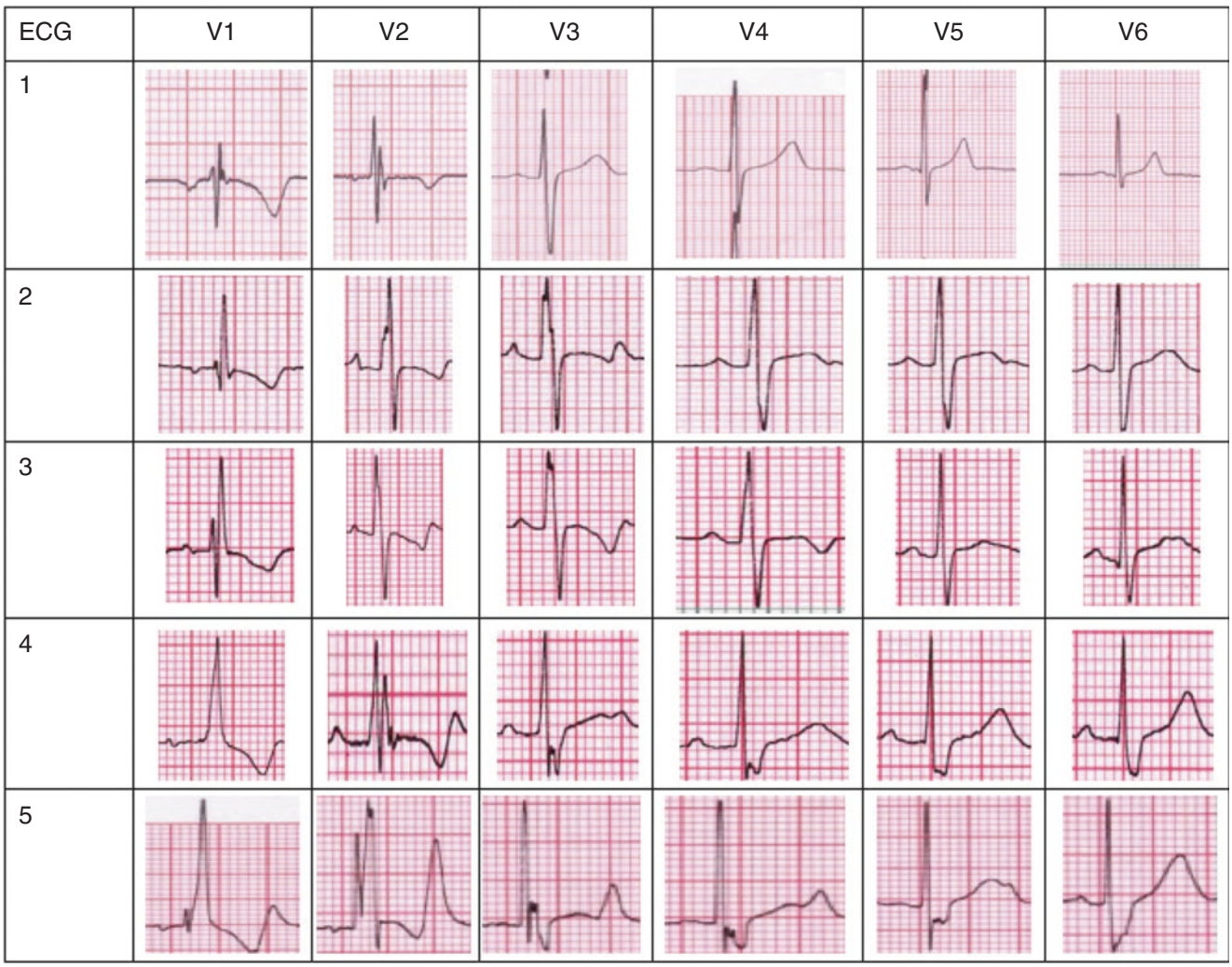

Fig. 3.15 Different degrees of right bundle branch delay and block

right ventricle (e.g., pre-tricuspid shunts such as anomalous pulmonary venous return, tricuspid or lung insufficiency, pulmonary stenosis, etc.). The so-called "crochetage" alludes to the notch in the upper part of the R wave in the inferior leads, which when associated with RBBB increases the diagnostic accuracy for ASD and raises the suspicion of an overload of the right ventricle volume even without RBBB [8] (sensitivity of 70% and specificity of 90% if the sign is present in all three inferior leads). The crochetage disappears after surgery in 35% of cases, even where the RBBB remains, confirming that its diagnostic value is supplementary and not redundant. A complete RBBB is almost always present after surgery of tetralogy of Fallot (in these cases, the QRS is always very wide but may be more gradual depending on the extent of the residual and progressive right ventricular dilatation) but can also be possible after closure of a subaortic VSD or be an incidental finding in subjects with healthy hearts. The finding of a complete RBBB justifies an echocardiogram, as does a clinical history that excludes a family disturbance (e.g., Lev-Lenègre disease).

An atypical RBBB, a variation of the palindrome "rsr" (nonclassic, atypical V6, etc.), from minimal to more complex changes in V1 such as polyphasic QRS can bring to mind Ebstein's disease; nevertheless, "strange" RBBBs should raise the suspicion of Brugada syndrome (especially in combination with a long PR), arrhythmogenic right ventricular cardiomyopathy (ARVC), and also the possibility of an Osborn wave in hypothermia, accidental or iatrogenic. In such cases, it is possible to see a thermo-dependent long QT.

An LBBB should never be considered normal as it can suggest many pathological conditions, particularly cardiomyopathies (but also cardiac tumors); it requires instrumental tests and follow-up. A very early appearance is particularly suspicious, while the analysis of large series of young adults shows more favorable outcome that can be compatible with excellent prognosis. It is worth repeating that before defining a pediatric ECG as showing "LBBB," ventricular pre-excitation must definitely be excluded. In the case of an oversight, we would be in good company: even the fathers of ventricular pre-excitation—Wolff, Parkinson, and White—initially described ventricular pre-excitation as a form of bundle branch block [9]. The wave that closes the QRS curtain has a negative deflection; the S wave comes from the depolarization of the right ventricular outflow tract and the posterior basal portion of the interventricular septum heading to the right and above. In adults, the "S1 S2 S3" pattern is noteworthy, and although seen even in 20% of healthy subjects, it can reveal right ventricular hypertrophy, especially in combination with RSR' framework in V1. While the S wave in the right precordial leads represents the left forces, the S wave in V6 can be considered a mirror of the R wave in V1; it shows good sensitivity and acceptable specificity in the diagnosis of right ventricular hypertrophy and is especially useful when lead V1 is not readable because of dressings and thoracic deformity.

3.5 J Wave

The J point is the ECG's no man's land, the space-time of transition from depolarization to repolarization. In theory, this junction (J means junction) should not be seen, being the imperceptible edge of fusion between the QRS and ST, the ECG's horizon. When, on the other hand, it deviates from baseline, heterogeneous situations may be involved: ischemia, pericarditis, bundle branch block, drugs, electrolyte imbalance, and early repolarization.

No man's land as it was called, troubled, and seismic, if you think about how dramatically fever (Brugada) or hypothermia (Osborn Wave) can shake things up. Today, among adult cardiologists, the J wave has become problematic due to the emergence, alongside the Brugada pattern, of ominous implications of sudden death and elevated J point in the right precordial leads and inferior-lateral leads ("J wave syndromes," early repolarization syndromes). Sometimes we squabble about whether the elevated J wave is attributed to late depolarization rather than early repolarization: the children's world seems to be spared similar diatribes. For here early repolarization preserves its historically capitalized benign significance and can be recognized by (1) ST-segment elevated detachment from 1 to 4 mm; (2) "notch," with reverse polarity to the R wave, or "slurring" of the terminal part of the R wave with consensual polarity to the R wave; (3) upward concave ST segment; (4) large T wave, wide and symmetrical; and (5) preference for precordial leads V2–V6. This so-called juvenile pattern prevails in athletes and can be found in up to 50% of them. Two variations have been described: the Caucasian variations, with elevated concave upward ST, and the Afro-Caribbean variation, with upward convex ST with an initial stretch which differs from Brugada type 1, where the elevated ST crashes down to sink into a negative T. Prominent J waves are described in specific clinical conditions such as frostbite or states like hypothermic neuroprotection; the wave is known under various names, ranging from "Camel sign" and "late delta wave" to "Osborn wave" (Fig. 3.16). Due to the diffusion of hypothermia in neonatal intensive care units, this rare ECG pattern can already be observed at a very early age.

This pattern and Brugada syndrome can also be encountered in children, although with reduced incidence compared with adults [10]. It seems that this depends on the hormone dependency of the syndrome, and in fact in childhood, there are neither the deaths nor the overwhelmingly male dominance typical of adults [11]. It is known that in apparently normal cases, hyperthermia may reveal a Brugada pattern similar to Ic class drugs used in drug testing. Fever accompanied by atypical bundle branch block can become embarrassing [12]. The prognostic significance of this phenomenon is unknown; when dealing with suspicious ECGs, an ECG recording during hyperthermia may be advisable although what to do in the case of any positivity is

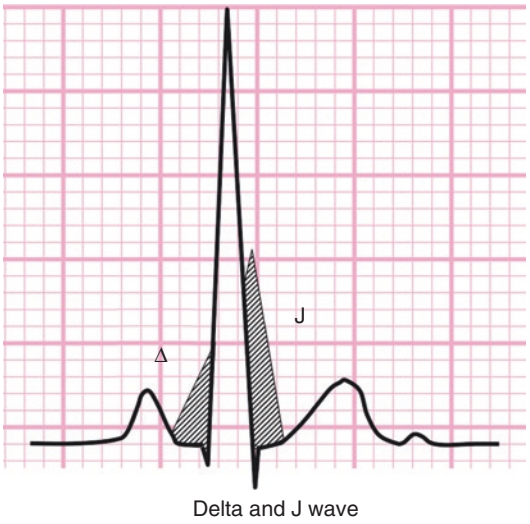

Fig. 3.16 Delta and J waves

far from certain (genetic testing?). As with adults, it may be useful to move the right precordial leads one or two intercostal spaces upward (second and third intercostal space in addition to the standard fourth space), a move that should unmask a false Brugada.

3.6 Epsilon Wave

The epsilon wave (EW) can be found in the first portion of the ST segment and may range from a small ripple to a reasonably sized wave. The EW is due to ventricular late potentials (Fig. 3.17). While the delta wave represents the early excitement of myocytes due to an accessory pathway, the epsilon wave represents the late excitement of a stunted and severely diseased myocardium, hence the name, in strict Greek succession to emphasize the nature of post-excitation opposite to pre-excitation. The epsilon wave is very specific for ARVC but has poor sensitivity and is rarer in the child than in the adults [13]. In this case, the preferred leads are the precordial ones from V1 to V4. This wave can also be seen in other situations with serious involvement of the right ventricle such as myocardial infarction, infiltrative disease, and sarcoidosis.

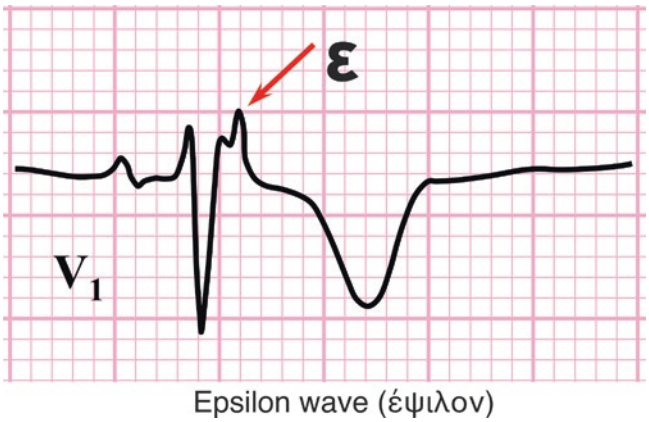

Fig. 3.17 Epsilon wave

3.7 ST Segment

The ST segment is located between the J wave and the beginning of the T wave. Regarding the displacement from the isoelectric line, ST depressions are in general pathological more often than ST elevations (see tracings in the iconographic section). The PR segment should be used as the reference baseline, but even if this precaution is taken, it is possible to confuse acute pericarditis with the juvenile early repolarization pattern; considering the many admissions to the emergency room of children and young people with chest pain, you will understand how sometimes it is difficult not to request an echocardiogram or biochemical examinations (at the risk of finding a high troponin value). In these cases, the PR segment analysis may be crucial. In fact, in classic acute pericarditis, the vector of the ST segment is facing the front, to the left and down giving rise to a diffuse concave elevation that is present in all leads except aVR and V1. There is also a depression of the PR segment, indicating an atrial subepicardial lesion, which may be present in all leads except in aVR, where it is sometimes even possible to note an elevation of the PR segment (the sign of the "knuckle") [14].

Pericarditis apart, in children, there are various conditions in which the ST segment can be altered: coronary anomalies, cardiomyopathies, electrolyte imbalances, pre-excitation, myopericarditis, tumors, overload, and drugs. Ischemic syndromes are rare and congenital, such as the anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) and the anomalous origin of the coronary artery from the opposite Valsalva's sinus (ACAOS) [15]. Among acquired coronary diseases, the commonest is represented by Kawasaki disease complicated by thrombosis of coronary aneurysms. Coronary fistulas connected with the pulmonary artery or the heart chambers may very rarely cause ischemia, only when they are very large.

An ALCAPA becomes symptomatic in the first months of life with heart failure and ECG abnormalities suggestive of acute or subacute myocardial infarction; in rare cases, the disease occurs in adulthood where it can presents with a normal ECG and sudden onset arrhythmia. An ACAOS is also compatible with a flawless resting ECG (even with an exercise ECG) and yet occurs at a young age with acute infarction or malignant ventricular arrhythmia. The ischemic mechanism of this condition is the constriction of the aberrant coronary artery, which runs between the aorta and the pulmonary trunk. Moreover, there may be an abnormal coronary ostium, segments with acute angles, and endothelial abnormalities that contribute to the occlusive process. A similar ischemic process may occur in pulmonary hypertension due to a dilated and hypertensive pulmonary artery which can compress the left main trunk of the left coronary artery, even when this originates regularly from the left Valsalva's sinus. In Kawasaki disease, the pre-infarction ECG is usually silent, and acute ischemia can occur only after thrombosis of a large aneurysm.

In the newborn, ST-segment depressions of 1–2 mm are considered normal, especially in the precordial leads, the ones where the T wave changes rapidly in the first weeks of life. Also in the early days of life, an elevation of the ST segment is acceptable in the inferior and left precordial leads (ST depression is less acceptable and in any case only in the first week) with negative T waves. It must be said that "sinister" diseases such as aortic coarctation may appear in the first days of life presenting with an overload of the right ventricle, having taken on the weight of the obstruction during fetal life.

Regarding the allocation of ventricular hypertrophy, after the first week, the ST-segment depression is a not very sensitive sign, but it is specific, much more useful than voltage amplitude.

3.8 T Wave

If there is an incontrovertible law in the heart, it is that the T wave always follows the R wave, as in the word *heart*. While it changes in accordance with cardiac development, at the earliest age, the T wave has a scenic point of observation in V1. Here, in the first week of life, it is capable of very rapid flights and can be positive or negative with impunity.

But from age 7 days to age 7 years (approximate numbers useful to remember), its positivity in V1 unmasks right ventricular overload with excellent sensitivity and specificity, long before the appearance of a pathological R wave or even a Q wave. So, the motto "in V1 from seven days to seven years the T wave never lies" is true. In the other precordial leads up to V4, the T wave can be negative up to age 10–12 years (up to 10% of children) in accordance with the so-called juvenile pattern, and then in V2–V6, the T resurfaces; if this is not the case, cardiomyopathy must be excluded—see ARVC—before designating it as a variant of normal. In fact, a negative T wave in leads other than V1 can also be found in 10% of young people under the age of 14 years but only in 1–2% thereafter. In the inferior-lateral leads, this percentage is even smaller and always requires investigation. An inverted T wave in the anterolateral lead combined with ST depression can reveal overload, myopathy, and, more rarely, ischemia. Accentuation of the negative-positive diphasism from V1 to V4 can be suggestive of volume overload of the right ventricle, typically in ASD [16]. In the presence of pre-excitation by accessory pathways, ST and T alteration has no prognostic significance (those who depolarize badly can repolarize badly, a precept that applies from pre-excitation to bundle branch block, through pacemakers or other temporary rhythms). An isolated negative T wave in lead III is a common finding, with or without accompaniment of the neighboring and often sympathetic aVF, but if in addition to this couple the T wave in the more detached lead II is also negative, then the picture becomes less normal. Negative T waves in the inferior leads are sometimes indicative of mitral valve prolapse, some say even before it is justified by the left ventricular overload caused by mitral regurgitation. In fact, it proves that in mitral valve prolapse, regardless of the valve function, there may be fibrosis of the postero-basal aspect of the left ventricle, which can justify certain electrocardiographic changes and also the typical arrhythmias of the mitral valve prolapse syndrome. A negative T isolated in aVL not accompanied by the same changes in V5–V6 and lead I or pathological Q waves may also be insignificant. To raise the suspicion of overload, it is not just the negative polarity of the T wave and its being discordant from QRS (always pathological except for bundle branch block) that is important but also its depth, geometry, and duration (Table 3.6). In the right precordial lead, a diphasic negative-positive T wave (overshooting) is suggestive of right ventricular volume overload, as in the case of ASD or anomalous pulmonary venous return. We call this picture the butterfly sign (see below); the amplitude of the voltage of this bipolarity and its extension toward the left precordial lead is correlated with the degree of the shunt.

Moreover, if in the same beat the T wave apex in V2–V3 is later than the apex of the T wave in the left precordial leads V5–V6 (we call this split of the T wave), the sensitivity for ASD increases [16].

Nearly 20% of children have a notched T wave in V2–V3 [6], which can be confused with a T wave on which a P wave is inscribed and can raise the suspicion of atrial arrhythmia with a 2:1 atrioventricular ratio, but looking at the limb leads, it is possible to clarify this false interpretation.

After 12 years of age, a negative T wave in V4 should never be accepted as normal. In addition to the polarity of the T, the area, the duration, and the variability are also important. In the first days of life, negative T waves are tolerated in the lateral leads. Likewise, at other ages too, small negative T waves in the inferior-lateral leads which normalize with posture and exercise can be considered normal, although this behavior is often not enough to avoid diagnostic tests such as ultrasound or magnetic resonance. Conversely, wide and deep negative T waves are always suspicious: when present in the left precordial leads, in the absence of symptoms and with normal auscultation, the first thing to exclude is hypertrophic cardiomyopathy and in particular the apical type, which can occasionally not be seen on an echocardiogram. Non-compaction cardiomyopathy (LVNC) can also occur in this

Table 3.6 Cause of deep T wave inversion

• Apical hypertrophic cardiomyopathy
• Solitary papillary muscle hypertrophy
• Left ventricular non-compaction cardiomyopathy
• Complete atrioventricular block (V2–V4)
• Severe right ventricular hypertrophy
• Non-Q infarction
• Acute abdomen (acute pancreatitis)
• Cocaine
• Subarachnoid hemorrhage—intracranial hypertension
• Cardiac electrical memory (post-ventricular tachycardia, post-pacemaker, intermittent pre-excitation)
• Cardiac tumor
• WPW

way. Negative and wide T waves in precordial leads can be seen in all cardiomyopathies, especially ARVC when limited to V1–V4. In complete AVB, it is not unusual to see giant T waves here (the so-called proximity phenomenon, due to increases in diastolic volume), a situation that needs to be monitored carefully because it could portend current or imminent bradycardia-dependent QT prolongation at risk of torsades de pointes. There are, on the other hand, situations (pre-excitation, pacemaker stimulation, paroxysmal tachycardia) in which T wave negativity transiently follows the event of altered excitatory dynamics: in these cases, the electric memory is called into question, a phenomenon that has no prognostic implications. Much more fearful is electric premonition, i.e., the appearance of pathological negative T waves in the absence of any clinical or instrumental warning sign, which could announce a cardiomyopathy is still in the larval stage (see certain particularly electrically unstable mutations). Among the noncardiac causes of deep negative T waves, intracranial hemorrhage is worthy of attention (Table 3.6).

3.9 U Wave

Small or even invisible, the U wave is named in alphabetical order, but the letter U also stands for "unknown." Its electrogenesis is in fact a matter of speculation. It is thought to be due to postpotentials of the ventricular myocardium and/or to the repolarization of Purkinje fibers. It usually has the same polarity as the T but also remains positive in leads in which the T is physiologically negative (precordial leads in young people, leads III). Normally it can be up to a third as wide as the T which precedes it; in children or during bradycardia, it is often prominent and especially increases in the course of hypokalemia. In the right precordial leads, it is often difficult to distinguish the U wave from a notch of the T wave, this being one of the reasons why the QT interval should never be measured in these leads [1].

3.10 QT Interval

The saying *in cauda venenum* (the poison is in the tail) is a perfect fit for the pitfalls of the last part of the cardiac cycle. The QT interval embodies repolarization, the delicate charging of the electrical device operated by an infinite variety of ion channels that may be lacking either genetically (autosomal dominant or recessive conditions) or because of acquired causes such as hypothermia, hypokalemia, or drugs (for an updated list, see www.QTdrugs.org). The anomaly of the QT—or rather the lack of homogeneity—is the basis of life-threatening electrical instability. Although the QT represents the most famous interval of the last 20 years, it is surprising that its correct measurement is still unknown to many [17], even among electrophysiologists. Here is the simple recipe for a correct QT interval measurement. Ingredients are a baseline standard 12-lead ECG (not a Holter device reading because the rapid heart rate changes can distort measurement), a ruler, and a calculator. Take lead II or V5 with the U wave partially skimmed off the T wave (the U wave should be taken into account only when it is more than half as high as the T). Take an average of 3 beats, excluding short cycles in relative bradycardia, as well as long post-tachycardia cycles, and then draw the tangent of the descending limb of the T wave, and consider the intersection with the isoelectric line as the end of the T wave. The web and various applications contain many formulas for QT correction, but the Bazett formula is the one most commonly adopted. Although modern electrocardiographs are very reliable, it is always better to do manual calculations using the tangent method [18]. A clearly separate U wave should not be counted, as opposed to when it is too merged with the T or more than half as wide as the T wave. As already mentioned, it can be difficult to distinguish a T wave notch from a U wave.

The correction of the QT interval is the mathematical reduction of a nonlinear phenomenon, and there is no perfect formula. More than 20 formulas have been proposed, but the Bazett formula is the one that is most common and easy to apply. It works sufficiently well in a field of frequencies between 50 and 120 bpm; however, for higher heart rate values, the Bazett formula tends to overestimate the QTc, while for lower heart rates, it tends to overcorrect and thus to underestimate it, in which cases it is better to use the formula of Fredericia. This phenomenon should be kept in mind especially in extreme bradycardia (as in total AVB) complicated by a long QT interval, in which the underestimation of repolarization threatens to disregard the risk of torsades de pointes. The QT in the infant is physiologically long (the physiological sinus tachycardia of neonatal age does overestimate the QT with the Bazett formula). A long QT syndrome at this age can present with sinus bradycardia and with profound bradycardia due to 2:1 AV block. Vice versa, "structural" AV blocks can

become complicated with bradycardia-dependent long QT, based on a prolongation of repolarization by functional remodeling of ion channels. A particularly short QT (QTc < 320 ms) should make you suspect the very rare short QT syndrome, especially when the T waves, particularly in the precordial leads, are high and sharp. Before pronouncing a binding diagnosis, bear in mind that congenital long QT, T wave, and U wave may be affected by electrolyte imbalance and drugs, although the electrical stability of the neonatal heart compensates for this influence. For several reasons, including the concept of critical mass, the heart of a newborn can also be surprisingly stable even in the case of very severe hypokalemia or other insults; nevertheless, this heart is subject to the laws of chaos of any complex system that could foresee the unpredictable catastrophic development of minor or imperceptible events: the butterfly effect. Finally, in the case of wide QRS, the ventricular depolarization must be separated from the calculation of the QT. A classic example is the patient with operated tetralogy of Fallot, who almost always has an RBBB with wide QRS and where you will thus have to consider the JT interval (normal JTc <360 ms).

References

1. Chou TC, Knilans TK. Electrocardiography in clinical practice: adult and pediatric. 4th ed. Philadelphia: WB Saunders; 1996. p. 37–53.
2. Saul JP, Walsh EP, Triedman JK. Mechanisms and therapy of complex arrhythmias in pediatric patients. J Cardiovasc Electrophysiol. 1995;6:1129–48.
3. WHO Expert Consultation on Rheumatic Fever and Rheumatic Heart Disease. Rheumatic fever and rheumatic heart disease: report of a WHO Expert Consultation, Geneva, 29 October–1 November 2001. WHO technical report series 923. Geneva: WHO, 2004.
4. Rosenbaum FR, Hecht HH, Wilson FN, Johnston FD. The potential variations of the thorax and the esophagus in anomalous excitation (Wolff-Parkinson-White syndrome). Am Heart J. 1945;29:281–326.
5. Takahashi A, Shah DC, Jais P, Hocini M, Clementy J, Haissaguerre M. Specific electrocardiographic features of manifest coronary vein posteroseptal accessory pathways. J Cardiovasc Electrophysiol. 1998;9:1015–25.
6. Oreto G, Luzza F, Donato A, Carbone V, Satullo G, Calabrò MP. L'elettrocardiogramma - un mosaico a 12 tessere. Centro Scientifico Editore: Torino; 2009.
7. Rivenes SM, Colan SD, Easley KA, et al. Usefulness of the pediatric electrocardiogram in detecting left ventricular hypertrophy: results from the prospective pediatric pulmonary and cardiovascular complications of vertically transmitted HIV infection (P2C2 HIV) multicenter study. Am Heart J. 2003;145:716–23.
8. Heller J, Hagège AA, Besse B, Desnos M, Marie FN, Guerot C. "Crochetage" (notch) on R wave in inferior limb leads: a new independent electrocardiographic sign of atrial septal defect. J Am Coll Cardiol. 1996;27:877–82.
9. Wolff L, Parkinson J, White PD. Bundle-branch block with short P-R interval in healthy young people prone to paroxysmal tachycardia. Am Heart J. 1930;5:685–704.
10. Probst V, Denjoy I, Meregalli PG, et al. Clinical aspects and prognosis of Brugada syndrome in children. Circulation. 2007;115:2042–8.
11. Ezaki K, Nakagawa M, Taniguchi Y, et al. Gender differences in the ST segment: effect of androgen-deprivation therapy and possible role of testosterone. Circ J. 2010;74:2448–54.
12. Baruteau AE, Baruteau J, Mabo P. The differential diagnosis of primary electrical diseases from seizures in childhood. Cardiol Young. 2010;20:111–2.
13. Hamilton RM. The diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) in children. In: Redington AN, Van Arsdell GS, Anderson RH, editors. Congenital diseases in the right heart. London: Springer-Verlag; 2009. p. 131–7.
14. Spodick DH. Acute pericarditis: current concepts and practice. JAMA. 2003;289:1150–3.
15. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation. 2007;115:1296–305.
16. Wang MX, Wu GF, Gu JL, et al. Defective T wave combined with incomplete right bundle branch block: a new electrocardiographic index for diagnosing atrial septal defect. Chin Med J (Engl). 2012;125:1057–62.
17. Viskin S, Rosovski U, Sands AJ, et al. Inaccurate electrocardiographic interpretation of long QT: the majority of physicians cannot recognize a long QT when they see one. Heart Rhythm. 2005;2:569–74.
18. Tyl B, Azzam S, Blanco N, Wheeler W. Improvement and limitation of the reliability of automated QT measurement by recent algorithms. J Electrocardiol. 2011;44:320–5.

Suggested Reading

Allen HD, Shaddy RE, Penny DJ, Feltes TF, Cetta F. Moss & Adams' heart disease in infants, children, and adolescents, including the fetus and young adult. 9th ed. Philadelphia: Lippincott Williams & Wilkins (LWW). isbn:978–1–496-30024-9

Bronzetti G, Mariucci E, Bonvicini M, Picchio FM. The ECG in pediatric patients: what the cardiologist needs to know. G Ital Cardiol (Rome). 2011;12:408–18.

Schwartz PJ, Garson A Jr, Paul T, Stramba-Badiale M, Vetter VL, Wren C, European Society of Cardiology. Guidelines for the interpretation of the neonatal electrocardiogram. A task force of the European society of cardiology. Eur Heart J. 2002;23:1329–44.

Arrhythmetics and the Magic Numbers in Cardiology

4

4.1 Heart Rate Calculation

4.1.1 In the Case of Regular Rhythm

1. Identify a QRS complex that falls on a vertical line bordering a large square; the HR is calculated by dividing **300** by the number of large squares that separate it from the next QRS complex.
2. By dividing **60,000** (milliseconds in 1 min) by the interval—in milliseconds—between two P waves (atrial rate) or between two QRS complexes (ventricular rate) (Fig. 4.1).

4.1.2 In the case of fast heart rate

1. The HR is obtained by dividing **1500** by the small squares between two waves.

In the standard ECG, the paper speed is 25 mm/s: a large square of graph paper measures 200 ms, and in 1 min there are 300 large squares (40 ms for each millimeter). If the two complexes are separated by only one large square, the HR will be 300 bpm; if there are two squares, the HR will be 150 bpm, three squares 100 bpm, four squares 75 bpm, five squares 60 bpm, and so on (Figs. 4.2 and 4.3).

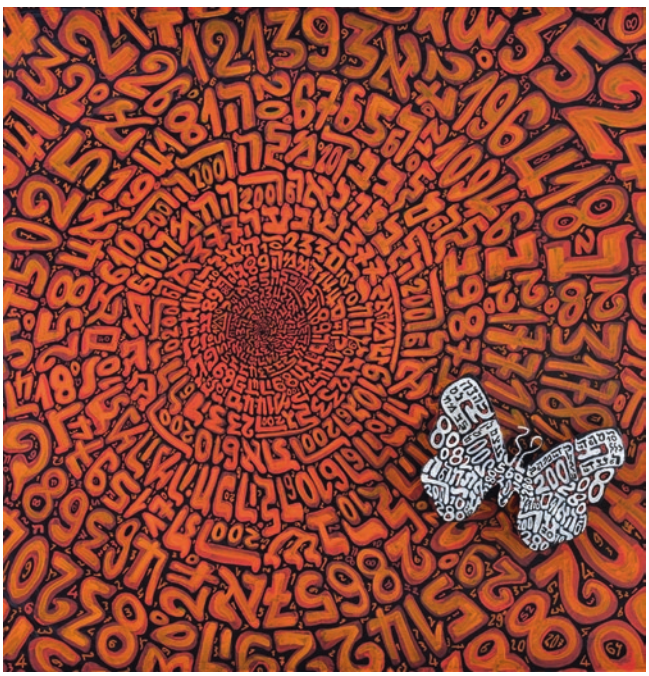

Fig. 4.1 Tobia Ravà. Existential codes

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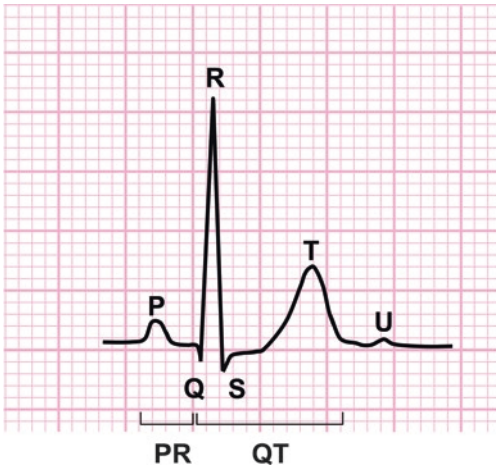

Fig. 4.2 The ideal PQRST complex and the seven questions (Portable Questionnaire Resolvable Simply Trying)

1. Is the paper speed 25 mm/s and the amplitude 10 mm/1 mV? For a fax or non-original copy: is the scale 1:1, i.e., do 10 small boxes of graph paper correspond to 10 mm?
2. Is the P wave ahead of the QRS? What polarity does P have in lead II? How tall is it? How long does it last? Is it definitely sinus? What is the atrial rate?
3. Is the PR normal? Can you exclude delta waves everywhere?
4. Are there as many QRS complexes as P waves? What is the direction of the electrical axis? Is the aVR as it should be (mostly negative apart from in the neonate), and can you exclude lead malpositioning? Is there a Q wave in V6? Is the amplitude of the R waves consistent with age and body weight?
5. Can the elevated ST segment in the precordial leads be explained by the early repolarization pattern?
6. Is the QT normal by applying the method of the tangent? At a glance, does the T wave end before the "center line" between two QRS complexes at the HR above 70 bpm? If so, the QT is normal.
7. Is the T wave negative in the precordial leads in a patient aged >12 years? This is not normal. Is the T wave negative in leads III and aVF? It could be normal.

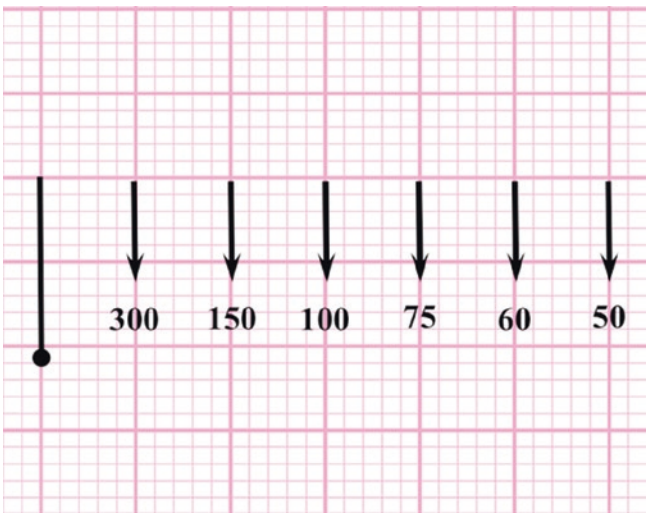

Fig. 4.3 Heart rate at a glance

4.1.3 In the Case of Irregular Rhythm

1. The HR is calculated by counting the number of QRS complexes in **6 s** (equivalent to 30 large squares or 15 cm of paper) and multiplying it by 10.
   Example: 21 QRS complexes in 30 large squares, the ventricular rhythm is 210 bpm.

4.2 QRS Axis

The ECG leads can be represented as axes lying on the same frontal plane, with the heart positioned at the center of it. The electrical activity of the heart can be reduced to a resultant vector which represents the instantaneous mean cardiac axis (hexaxial reference system) (Fig. 3.3). On the hexaxial reference system:

- For simplicity the normal heart axis is between 0° and 90° (normal values vary according to age).
- Right axis deviation: the cardiac axis moves to the right (more positive than 90°, between 90° and 180°).
- Left axis deviation: the cardiac axis moves to the left (more negative axis than 0, between 0° and −90°).

For electrical axis calculation, a practical method may be to find the lead in the frontal plane with an isoelectric QRS (the algebraic sum of wave approaches zero): on the hexaxial reference system, the QRS axis will be perpendicular to this lead, directed toward one of the two leads which are located at right angles to the previous one and precisely toward the lead in which the positive component is predominant. In the absence of a perfectly isoelectric lead, it is necessary to make corrections of 15°. For example, if the "almost" isoelectric lead is aVF with R > S pattern and positive lead I, the cardiac axis will not be exactly at 0° (between the left two quadrants) but in the lower left quadrant at +15°; in the same way, in the case of aVF with R < S pattern and positive lead I, the cardiac axis will be not 0° but in the upper left quadrant at −15°. Another method is based on analysis of leads I and aVF, which in the hexaxial system are arranged perpendicularly (Figs. 4.4 and 4.5).

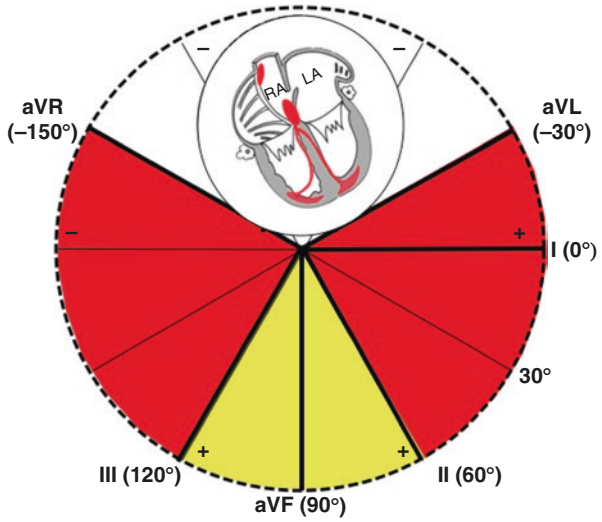

Fig. 4.4 The hexaxial system. The wing span of normality

4.3 QT Estimation at a Glance

We already talked about the correct measurement of the QT (*the cutest method to test the QT? Cut the T with the tangent.* Note that in the last sentence, every word contains a T, as well as every tangent sword contains the QT") (Chap. 3). Well, is it possible to roughly estimate the length of QTc at a glance, without a ruler, without a calculator, and without a web connection? At an HR of 60 bpm, it's easy, because only in this case is the measured QT the same as the corrected one (see the Bazett formula). Above and below 60 bpm, the QTc will be, respectively, higher and lower than the measured QT. With good approximation and at HR above 70 bpm, the QT interval can be defined as normal if the T wave ends before the vertical line equidistant from the two R waves that include it; when the HR is greater than 80 bpm, it may be normal for the QT interval to exceed the center line, while

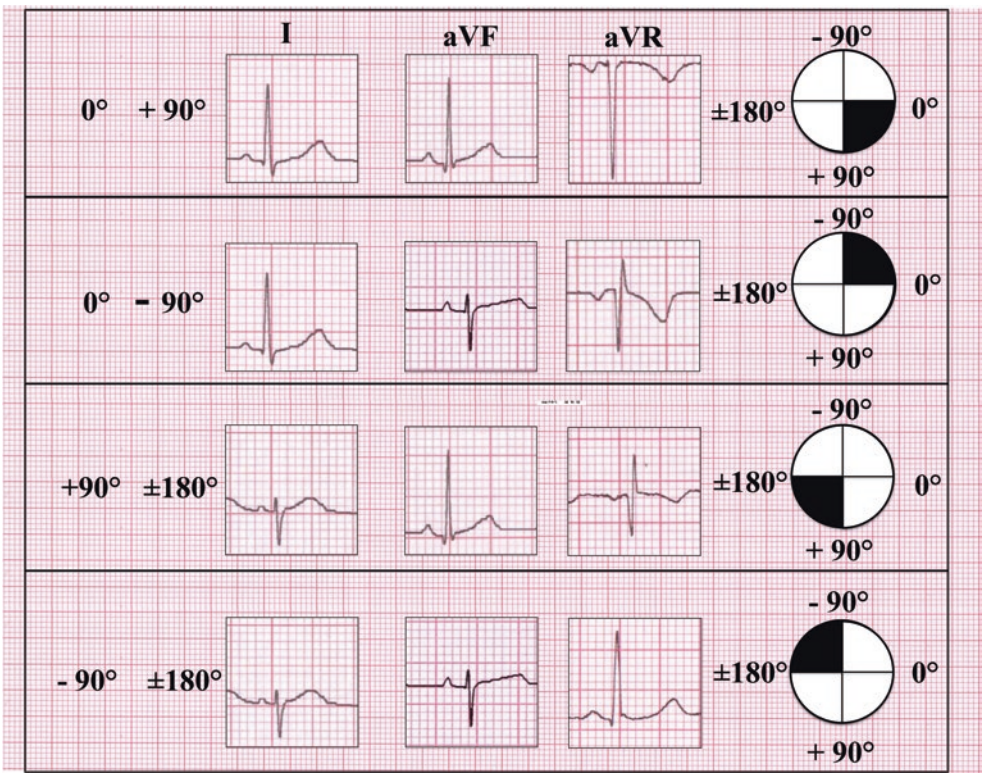

Fig. 4.5 Quadrants and cardiac axis

HR below 70 even a long QT ends before the center line, and so you have to make a precise calculation. More mathematically, the QT is normal when at 70 bpm the value does not exceed 400 ms; for each increase of 10 beats, the measured QT interval should be 20 ms shorter (so at 80 bpm, it should not exceed 380 ms, at 90 bpm 360 ms, 100 bpm 340 ms, and so on).

4.3.1 Bazett Formula

QTc is the QT interval corrected by the HR: the numerator is the measured QT interval in millisecond, and the denominator is the RR interval in seconds (obtained by dividing 60 by the HR). As previously mentioned, this formula is convenient and commonly used but also fairly inaccurate. Several methods have been proposed, and the approach based on regression like the one shown below seems the most accurate (Table 4.1).

Bazett formula [1]:

$$\mathrm{QTc} = \frac{\mathrm{QT}}{\sqrt{\mathrm{RR}}}.$$

Fridericia formula [2]:

$$\mathrm{QTc} = \frac{\mathrm{QT}}{3\sqrt{\mathrm{RR}}}.$$

Sagie o Framingham formula [3]:
$\mathrm{QTc} = \mathrm{QT} + 0.154 \times (1000 - \mathrm{RR})$

Nomogram formula [4]:
$\mathrm{QTc} = \mathrm{QT} + 0.116 \times (1000 - \mathrm{RR})$ if RR >1000 msec;
$\mathrm{QTc} = \mathrm{QT} + 0.156 \times (1000 - \mathrm{RR})$ if 600 < RR <1000 msec;
$\mathrm{QTc} = \mathrm{QT} + 0.384 \times (1000 - \mathrm{RR})$ if RR < 600

Table 4.1 Magic numbers in cardiology

Number	Cardiac world	Noncardiac world
440	• "Old" upper limit of normal QTc interval	• 440 Hz, the frequency of the note La and the tuning fork, on which all musical instruments are tuned • It is a Harshad number (a positive integer divisible by the sum of its digits): 440: 8 = 55
330	• Lower limit of QT interval, below which it is possible to suspect a short QT	• Maximum speed of the Lamborghini Murcielago • Harshad number: 330:6 = 55
220	• HR beyond which PSVT is suspected in infants • Limit from which to subtract the patient's age to determine the maximum HR during exercise • A SPERRI <220 ms appears more frequently in patients with WPW who suffered cardiac arrest	• 220 Volt is the domestic electrical voltage in Italy; • Harshad number: 220:4 = 55
110	• QRS duration ≥110 ms in V1–V3 is an ECG criterion for ARVD • >110° = criterion for right axis deviation	• Maximum degree mark in Italy • Harshad number: 110:2 = 55
110 × Φ	• 180 ms = duration of QRS beyond which the risk of TV increases in patients with TDF • 180 ms = upper limit of the PR interval in children	• Maximum score in darts • Harshad number: 180:9 = 20
55	• ECG criterion for ARVD: an interval >55 ms between the nadir and the upstroke of S wave in V1–V3 • Correlates with the severity of the disease and with the induction of TV in the EPS • Limit in milliseconds from the beginning and nadir of the S wave; in wide complex tachycardia, it is useful to differentiate VTs from PSVTs conducted with aberrancy (ms) • Upper limit of the HV interval in the EPS (ms)	• Tenth number of the Fibonacci sequence

ARVD arrhythmogenic right ventricular dysplasia, *AF* atrial fibrillation, *Nadir* the most negative point of the S wave on the ECG, *EPS* electrophysiological study, *SPERRI* shortest preexcited RR interval (used to define the properties of electrical conduction of the accessory pathway in patients with WPW during AF), *TOF* tetralogy of Fallot, *PSVT* paroxysmal supraventricular tachycardia, *VT* ventricular tachycardia, *Upstroke* terminal part of the S wave that is equivalent to the attainment of the baseline, *WPW* Wolff-Parkinson-White, *Φ* golden number that corresponds to 1.6180339

References

1. Bazett HC. An analysis of time relations of the electrocardiogram. Heart. 1920;7:353–70.
2. Fridericia LS. Die Systolendauer im Elektrokardiogramm bei normalen Menschen und bei Herzkranken. (The duration of systole in the electrocardiogram of normal subjects and of patients with heart disease). Acta Med Scand. 1920;53:469.
3. Sagie A, Larson MG, Goldberg RJ, Bengtson JR, Levy D. An improved method for adjusting the QT interval for heart rate (the Framingham heart study). Am J Cardiol. 1992;70:797–801.
4. Karjalainen J, Viitasalo M, Mänttäri M, Manninen V. Relation between QT intervals and heart rates from 40 to 120 beats/min in rest electrocardiograms of men and a simple method to adjust QT interval values. J Am Coll Cardiol. 1994;23(7):1547–53.

Suggested Reading

Baumert M, Porta A, Vos MA, Malik M, Couderc JP, Laguna P, Piccirillo G, Smith GL, Tereshchenko LG, Volders PG. QT interval variability in body surface ECG: measurement, physiological basis, and clinical value: position statement and consensus guidance endorsed by the European heart rhythm association jointly with the ESC working group on cardiac cellular electrophysiology. Europace. 2016;18:925–44.

Priori SG, Wilde AA, Horie M, Cho Y, Behr ER, Berul C, Blom N, Brugada J, Chiang CE, Huikuri H, Kannankeril P, Krahn A, Leenhardt A, Moss A, Schwartz PJ, Shimizu W, Tomaselli G, Tracy C. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes: document endorsed by HRS, EHRA, and APHRS in May 2013 and by ACCF, AHA, PACES, and AEPC in June 2013. Heart Rhythm. 2013;10:1932–63.

Schwartz PJ, Ackerman MJ. The long QT syndrome: a transatlantic clinical approach to diagnosis and therapy. Eur Heart J. 2013;34:3109–16.

ECG Metamorphosis: From the Newborn to Adulthood

5

5.1 The Newborn Infant

There is one thing that neonatologists and one of the greatest Italian poets, Giacomo Leopardi, agree on: the first 48 h of man are the most dangerous—man is born with labor, and the risk of death is birth [1, 2]. Even the easiest childbirth involves dramatic adaptations. Dystocia, asphyxia, diaphragmatic hernia, and infectious, toxic, metabolic, or respiratory agents affect the pulmonary circulation, opposing the physiological decline in arterial resistance. The right ventricle is the first to pay the hypoxic-ischemic bill, but in severe cases the left is not much better off. We are talking in these cases of persistent pulmonary hypertension or neonatal transient ischemia. There are mild forms with slight cyanosis and tricuspid regurgitation and lethal forms with severe global dysfunction. The ECG reveals, in order, right overload beyond the norm or diffuse ischemia. Likewise, therapy ranges from just a whiff of oxygen to extracorporeal membrane oxygenation (ECMO); the latter drastic resolution is nowadays often obviated by sildenafil and nitric oxide [3].

Does anyone remember the controversy about neonatal long QT screening that inflamed the 1990s? Only the members of the lined-up troops remain, skeptics versus incurable optimists [4, 5]. Now each country behaves in its own way, with a trend to a historical opposition between "Americans" and "Italians."

The first would never do an ECG; the second do it to everyone. According to the Italians, the ECG is a valuable tool because it sees not only diseased ion channels but also canal diseases (partial, intermediate, and complete atrioventricular canal defect).

Complete neonatal AVB affects one newborn in 20,000 and is always pathological. In a third of cases, it is associated with structural heart disease (CCTGA, CAVC, Ebstein's anomaly); in others it is idiopathic or autoimmune as in children of mothers with SLE. Complete AVB is usually recognized during ultrasound fetal examination.

There are serious heart diseases that can escape a fetal ultrasound and can manifest dramatically in the first month of life, due to the closure of the ductus arteriosus. Aortic coarctation stands out in this group, and an ECG could contribute to the diagnosis, when palpation of femoral pulses is not enough.

In summary, if for some reason you come across a neonatal ECG, you should assess three main aspects: P waves and HR, QRS axis, and repolarization. The parameters of normality of all these points have already been described. Even a quick glance should never miss a left axis deviation: if associated with long PR and RBBB, you have to consider AVC (partial or complete defect); if associated with normal PR and too "adult" precordial leads, without the expected right ventricular prevalence typical of infants, then tricuspid atresia must be considered. As far as repolarization is concerned, not only is the quantity (in the neonate the QT is normal up to 490 ms) important but also the quality. You must take into account that especially in the precordial leads, finding slightly elevated ST traits and small negative T waves can be normal in the first week of life, much more than finding a depressed ST and negative, large, and symmetrical T waves (Figs. 5.1 and 5.2).

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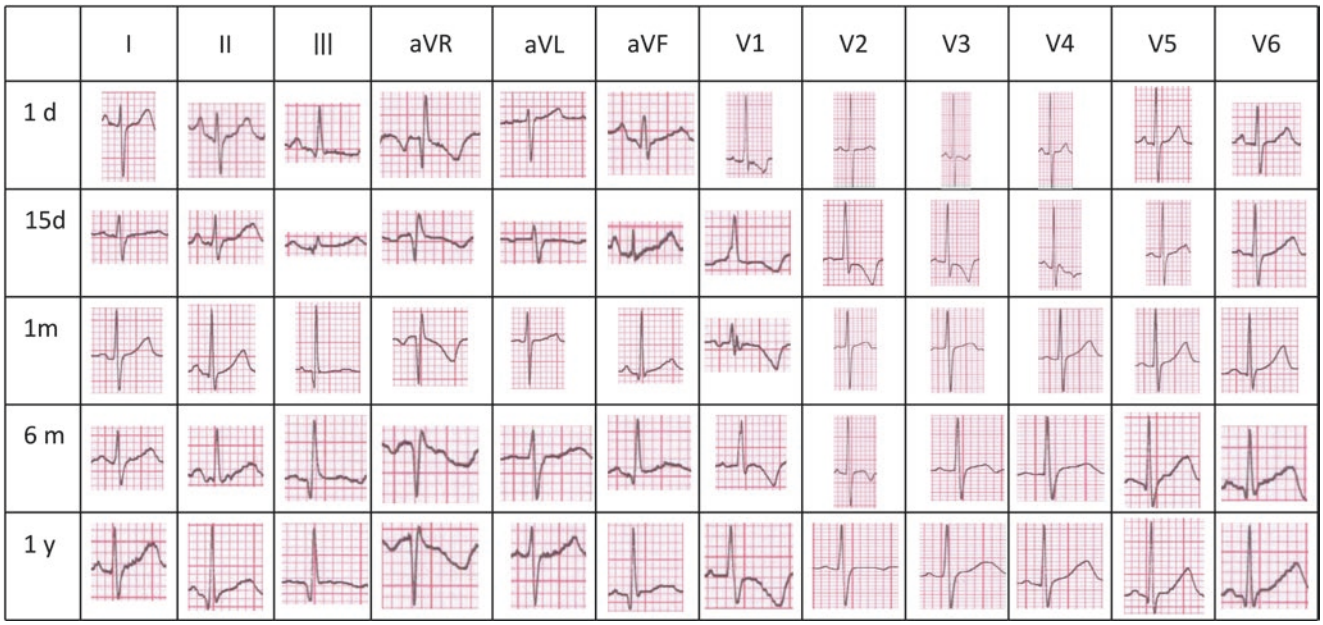


Fig. 5.1 Normal ECG through the first year of life

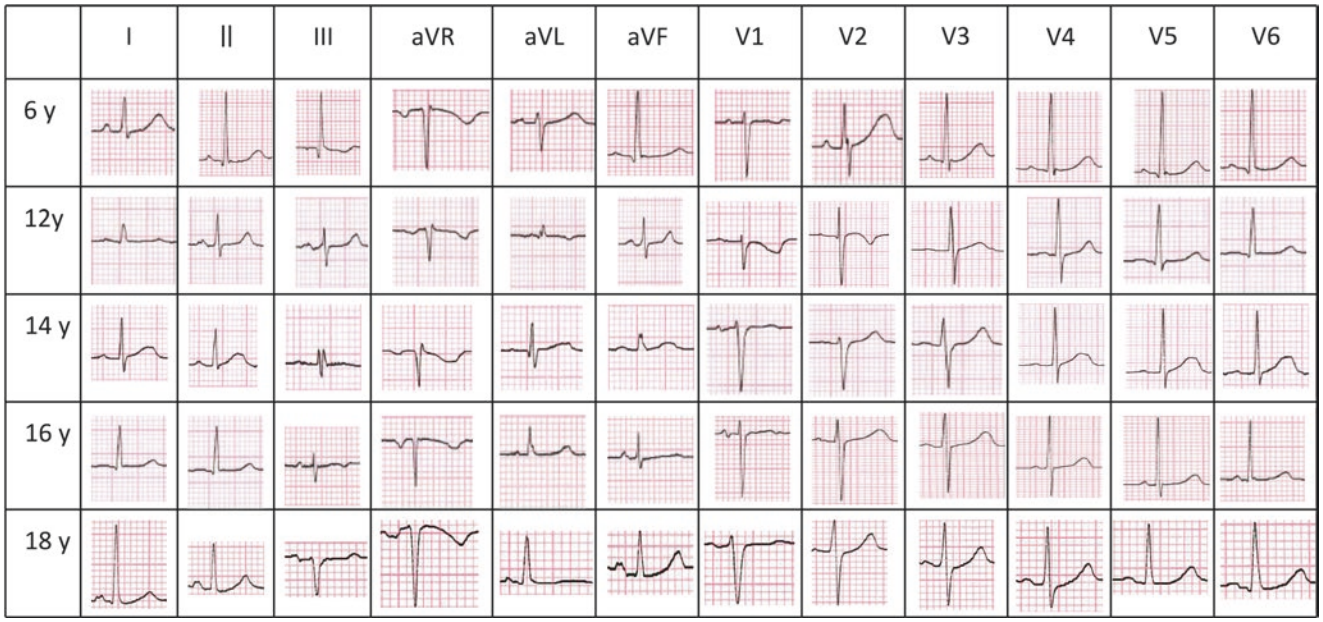


Fig. 5.2 Normal ECG after 6 years of life

5.2 The Child-Kid

During the developmental age, the normal ECG and all its variants may already be tricky enough, but it may also present with even more disturbing scenarios (Fig. 4.1). Syncope and chest pain are timeless dilemmas. But even before symptoms that are alarming, the mere case of a youngster who wants to take up a sport opens unresolved issues. In emergency conditions, together with history and physical examination, the ECG has an acceptable sensitivity. Myocarditis is a great pretender (in an ominous group made up of aortic dissection, pulmonary embolism, endocarditis, hypertrophic cardiomyopathy), and yet in the young it does not enter into competitive diagnosis with coronary atherosclerotic disease but with congenital coronary anomalies. Widespread low voltages—<5 mm in the limb leads and <10 mm in the precordials—together with ST-segment changes, are suggestive of myocarditis. You can also see Q waves of necrosis in

the absence of adequate alterations of the ST segment and vice versa. Among congenital anomalies of coronary arteries, ALCAPA may become symptomatic during the first months of life. In fact, in the case of early heart failure, the ECG should always be checked for signs of ischemia (Q waves in DI and aVL, ST trait alteration) diagnostic of this rare but operable cardiopathy. On the contrary, with ACAOS (origin of a coronary artery from the opposite sinus)—with an interarterial course or not—the disease can be silent for many years, and the baseline ECG is always normal but can suddenly become symptomatic at a young age (usually after 10 years of age) with syncope, chest pain, or life-threatening arrhythmias during exercise. ACAOS differs from myocarditis even by a sudden mutability of the tracings during critical coronary obstruction. A normal ECG at rest and dramatic symptoms during exercise are traits that ACAOS shares with a purely electric disease, catecholaminergic polymorphic ventricular tachycardia (CPVT).

Still on the subject of coronary accidents, we have to mention the thrombosis of a coronary artery aneurysm in the setting of Kawasaki disease as an extremely rare cause of syncope or sudden death.

So, ACAOS and CPVT are therefore potentially lethal diseases, the first hemodynamic and the second electric, that can be hidden in the resting ECG. Alongside this ominous couple, we can also see the long QT, sometimes marked by variability such that a patient can alternate between a pathological ECG and other normal ones. In all these situations, the exercise ECG becomes a key test when a clinical suspicion is present—a familial ominous syncope on exertion—even in the presence of normal resting ECG. Pain or alteration of the ST segment (ACAOS), ventricular arrhythmias adrenergic dependent (CPVT) or altered QT hysteresis (long QT) may reinforce the suspicion and indicate the need for further investigations.

Exercise testing is also useful in situations like manifest arrhythmic preexcitation and ventricular extrasystoles. In the first, the disappearance of the preexcitation during physiological sinus tachycardia is to be hoped for, so as to prescribe, in the least invasive way possible, a long refractory period of the accessory pathway and with it a low risk of arrhythmia (defined as the inability of the accessory pathway to quickly conduct a possible atrial fibrillation to the ventricles). In the second, we expect a suppression of the ventricular ectopy by the growing sinus rhythm, an encouraging but not definitely exculpatory result, to show that the arrhythmia is not adrenergic dependent and does not get worse at higher heart rates.

References

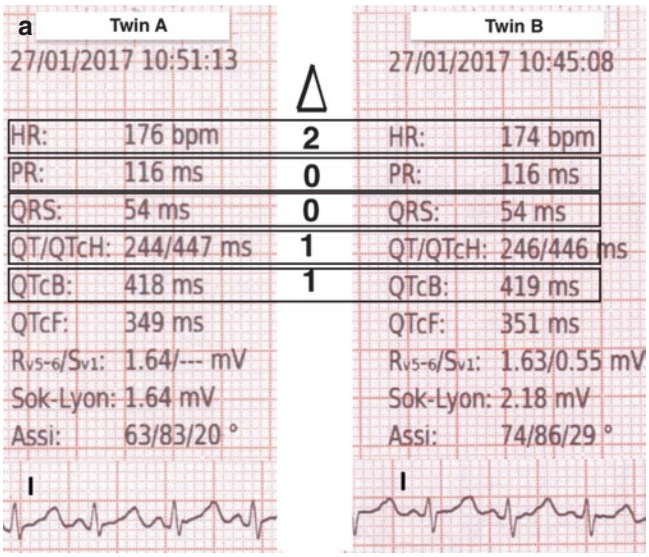
1. Bahl R, Qazi S, Darmstadt GL, Martines J. Why is continuum of care from home to health facilities essential to improve perinatal survival? Semin Perinatol. 2010;34:477–85.
2. Leopardi G. Canto notturno di un pastore errante dell'Asia. Can Underwrit.
3. Lapointe A, Barrington KJ. Pulmonary hypertension and the asphyxiated newborn. J Pediatr. 2011;158(2 Suppl):e19–24.
4. Schwartz PJ, Stramba-Badiale M, Segantini A, et al. Prolongation of the QT interval and the sudden infant death syndrome. N Engl J Med. 1998;338:1709–14.
5. Zupancic JA, Triedman JK, Alexander M, Walsh EP, Richardson DK, Berul CI. Cost-effectiveness and implications of newborn screening for prolongation of QT interval for the prevention of sudden infant death syndrome. J Pediatr. 2000;136:481–9.

Suggested Reading

Brockmeier K, Nazal R, Sreeram N. The electrocardiogram of the neonate and infant. J Electrocardiol. 2016;49:814–6.

O'Connor M, McDaniel N, Brady WJ. The pediatric electrocardiogram. Part I: age-related interpretation. Am J Emerg Med. 2008 Feb;26:221–8.

Schwartz PJ, Garson A Jr, Paul T, Stramba-Badiale M, Vetter VL, Wren C, European Society of Cardiology. Guidelines for the interpretation of the neonatal electrocardiogram. A task force of the European society of cardiology. Eur Heart J. 2002;23:1329–44.



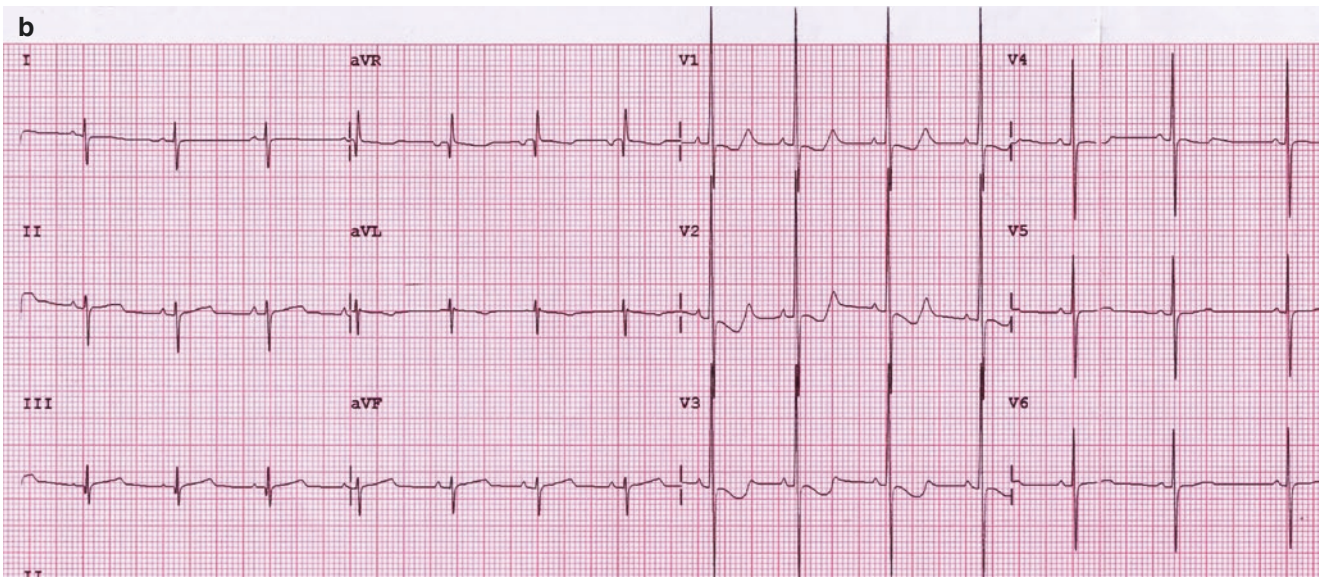


Fig. 6.1 (**a**) Two identical twins (female) moderately preterm, 2 kg weight. The parameters are identical: PR and QRS duration are the same and the QTc delta is only 1 ms. This could be nice evidence that ECG is a reliable diagnostic tool. (**b**) Normal newborn. See the right prevalence in the precordial leads normal for age. R wave in V1 is 25 mm and S wave in V6 is 11 mm, both at the upper limit. The ST segment is mildly depressed up to V4 but not pathologic. Low amplitude, flat or slightly negative T waves are normal in the first week of life. P axis tends to the second quadrant. The ventricular mean vector is oriented to the right side as expected in a newborn

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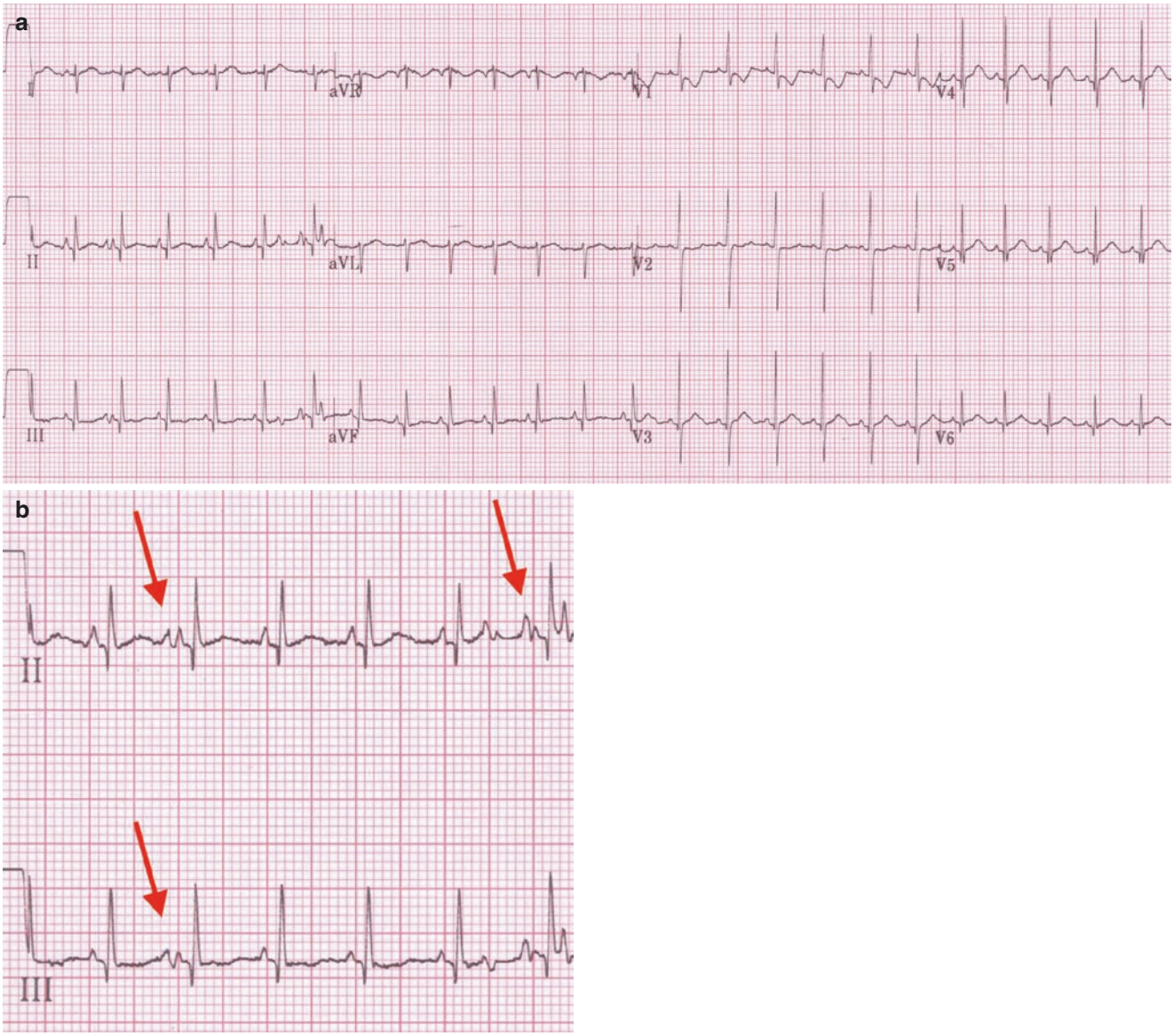

Fig. 6.2 (**a**) One-week-old female, HR 150 bpm, QRS axis 105°. (**b**) Close-up: there are artifacts in many leads (movement or hiccups). These waves do not alter the P and R sequence and hence must be an extracardiac phenomenon

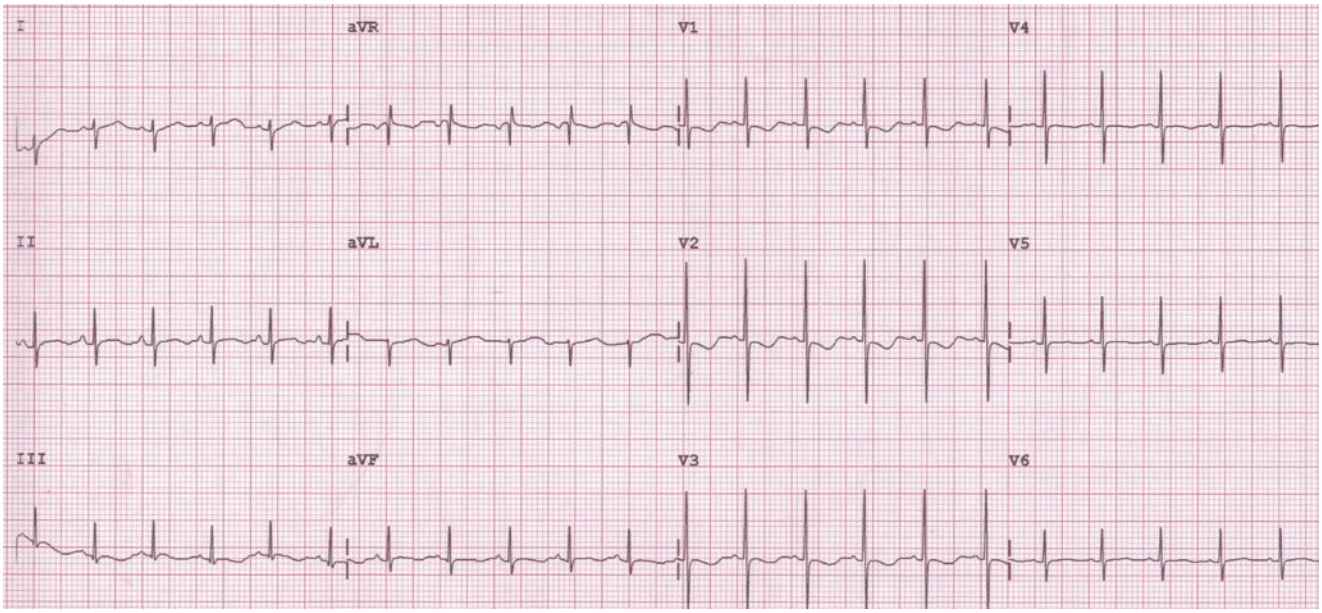

Fig. 6.3 Male neonate four day old; note the non-specific ST–T changes, normal for age

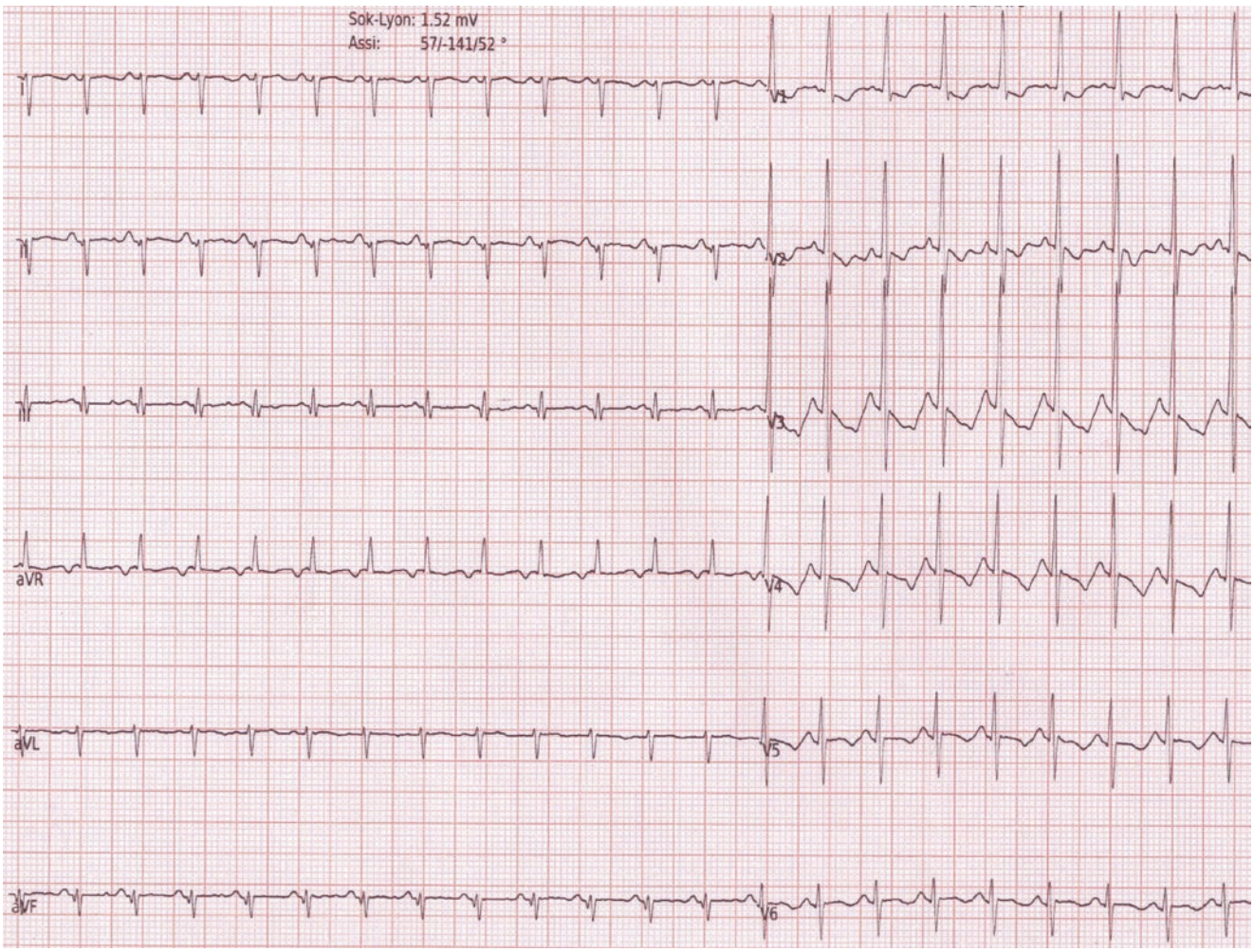

Fig. 6.4 One-week-old boy with mild pulmonary hypertension due to dystocia. ST–T changes are diffuse and more than normal in the precordial leads

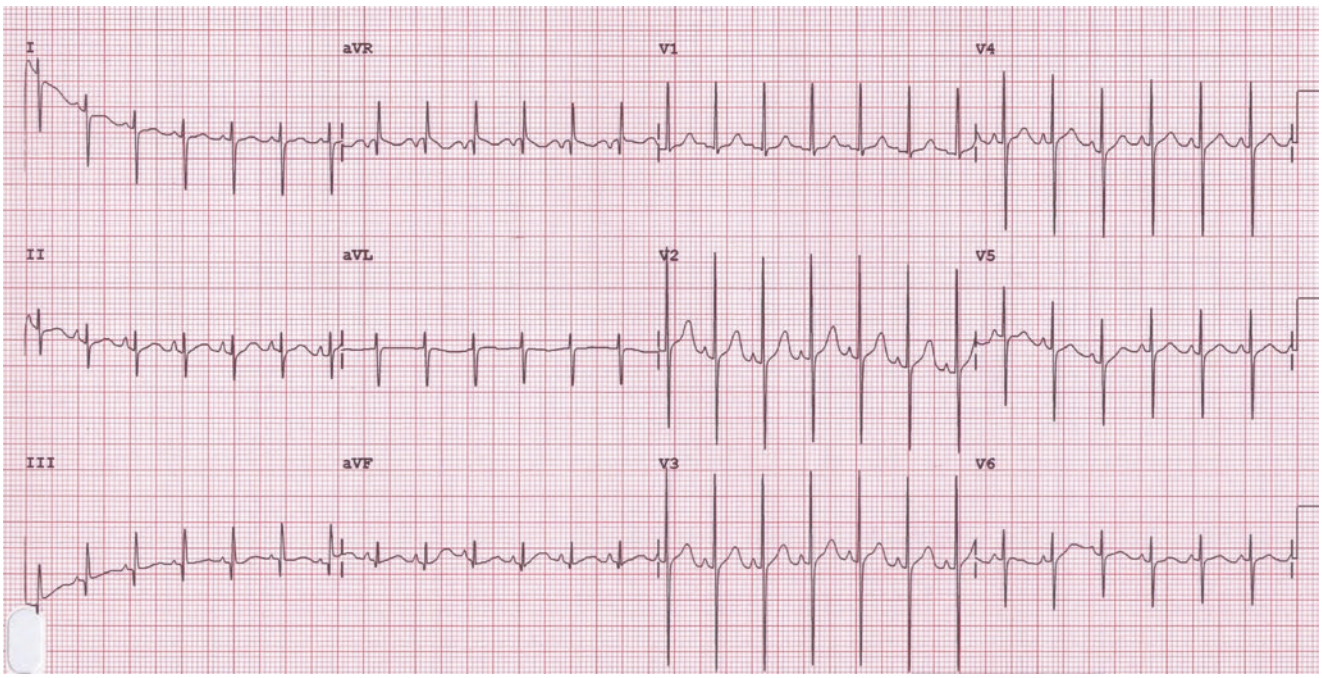

Fig. 6.5 Three-day-old female. The right axis deviation is normal. There is a positive T wave in V1; till the first week of life, a positive T wave in V1 is considered non-pathologic; on the contrary from 7 days to 7 years, a positive T wave in V1 is a sign of right ventricle overload (pressure > volume)

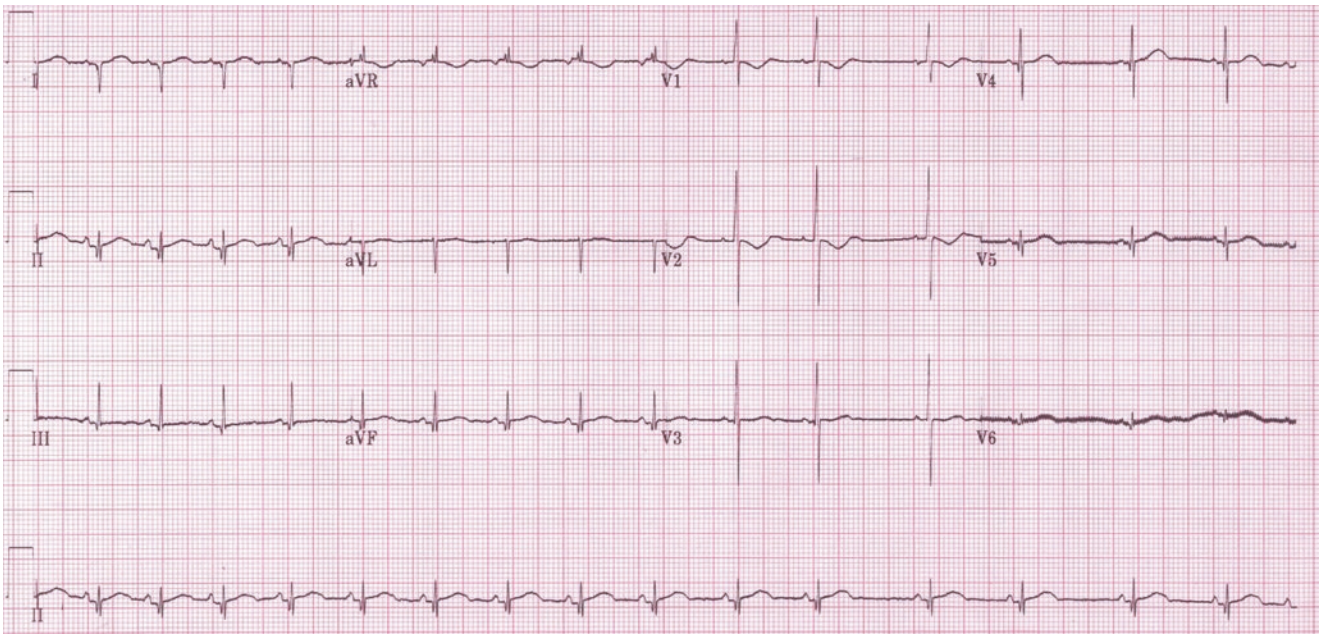

Fig. 6.6 Male, 3 day old with sinus bradycardia. A relative bradycardia that responds to stimuli should not be considered pathological. There is a marked right axis deviation and the left forces are poorly represented, as expected in a newborn

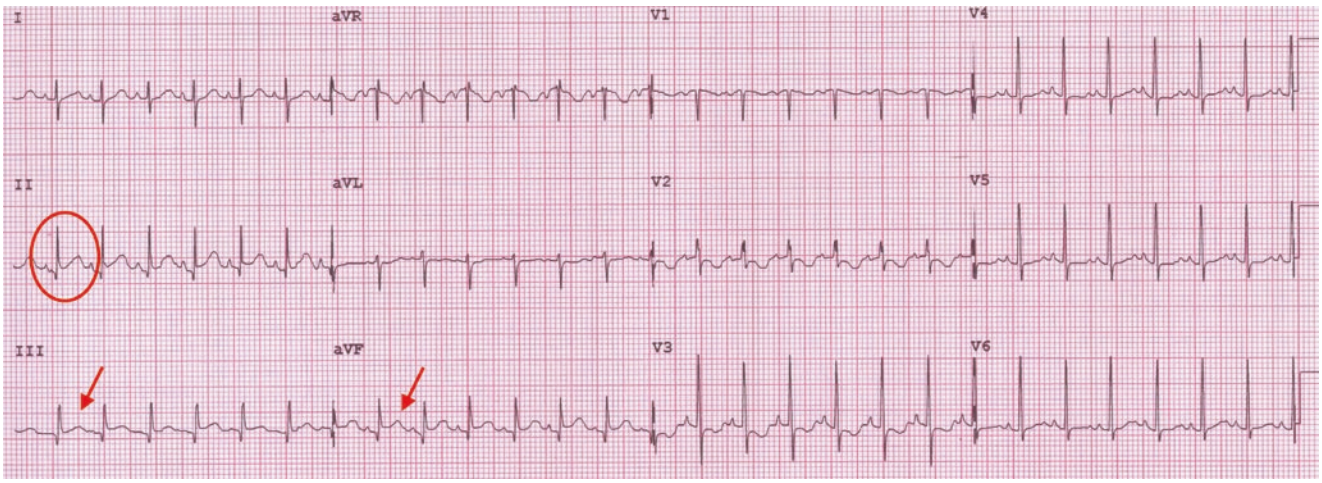

Fig. 6.7 Preterm infant. A left ventricular prevalence can be noted. The QRS axis is vertical in lead I and is isoelectric and then about 90°. The absence of the expected right ventricle prevalence is typical of a preterm baby (the more severe the prematurity, the more pronounced the left prevalence)

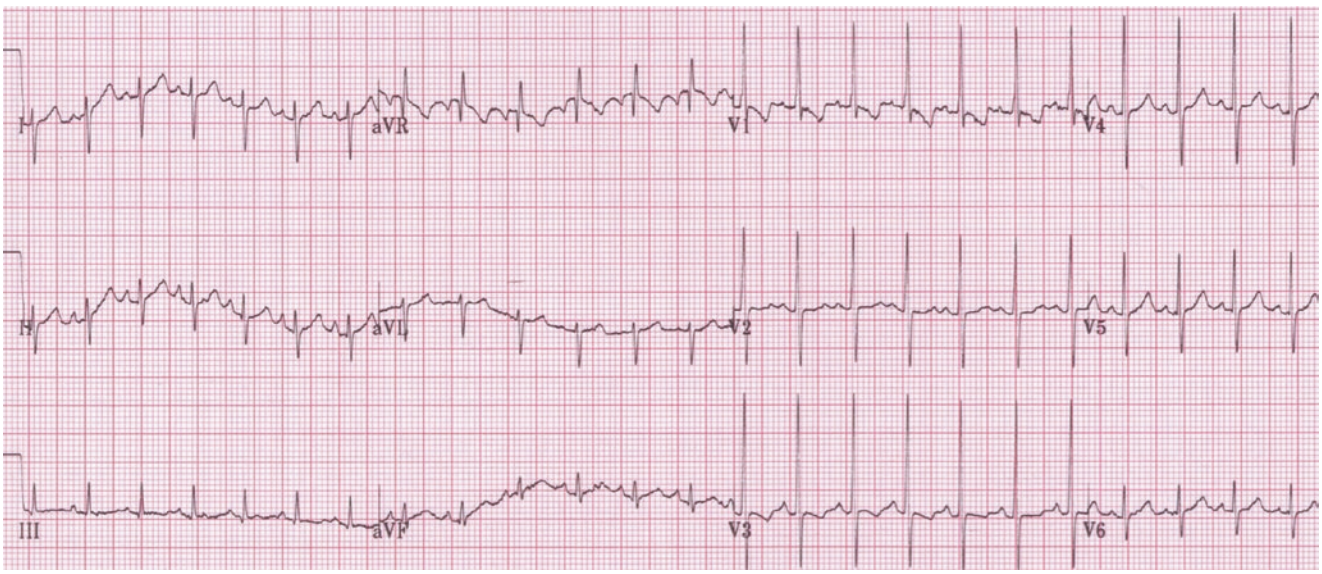

Fig. 6.8 Fifteen-day-old boy. P wave in DII with a regular axis in the second left quadrant; HR of 150 b/min normal for the age. The QRS axis is deviated to the right (150°) but as an isolated feature is not pathological. In the precordial leads, you can see a right ventricular dominance with "left forces" rightly still behind. In the right precordial leads, the ST segment is mildly depressed (horizontal-descendent non-pathologic change)

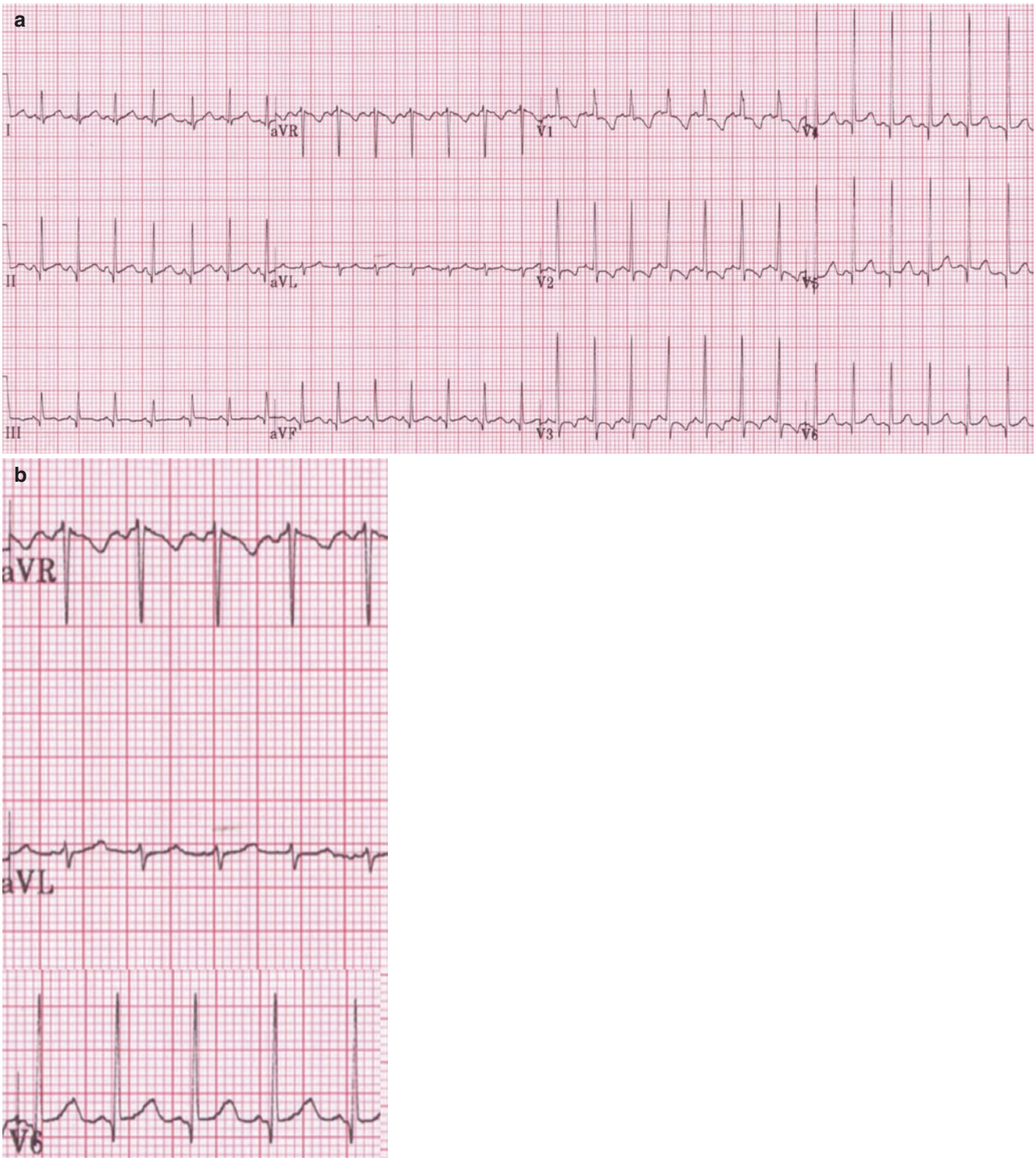

Fig. 6.9 (**a**) One-month-old female. HR around 150 bpm; aVL is the most isoelectric lead placing the ventricular axis around 60°.The right prevalence vanishes and left precordial leads begin to be pronounced. (**b**) Close-up

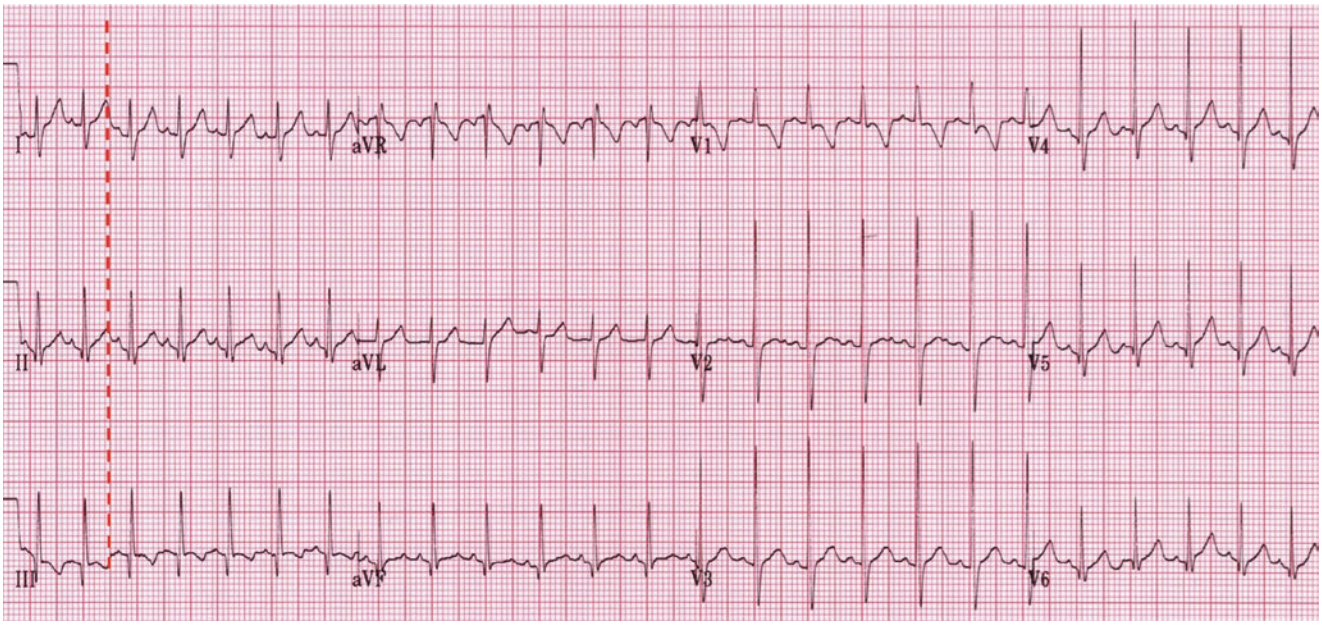

Fig. 6.10 Two-month-old baby boy. Normal sinus rhythm for age (> 150 bpm), QRS axis around 80°, normal. The T in V1 is negative, then normal; in lead III there is a negative T, not pathological. The QTc interval is 415 ms; at a glance the T ends just after the point equidistant from the R waves among which the T is included. At 150 bpm that describes a normal QT

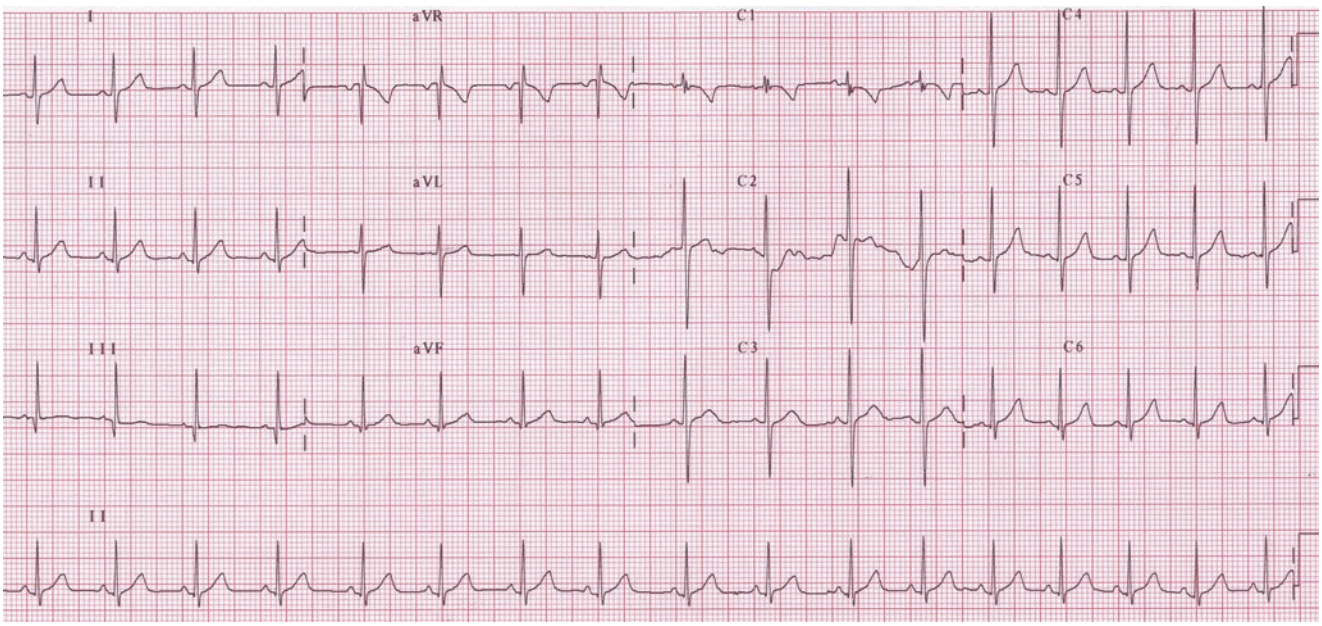

Fig. 6.11 Three-month-old female. Axes P, R, and T normally point to the second left quadrant (between 0° and 90°) consistent with age. The QRS in V1 can be defined as "mild delay of right intraventricular conduction" and not a RBBB

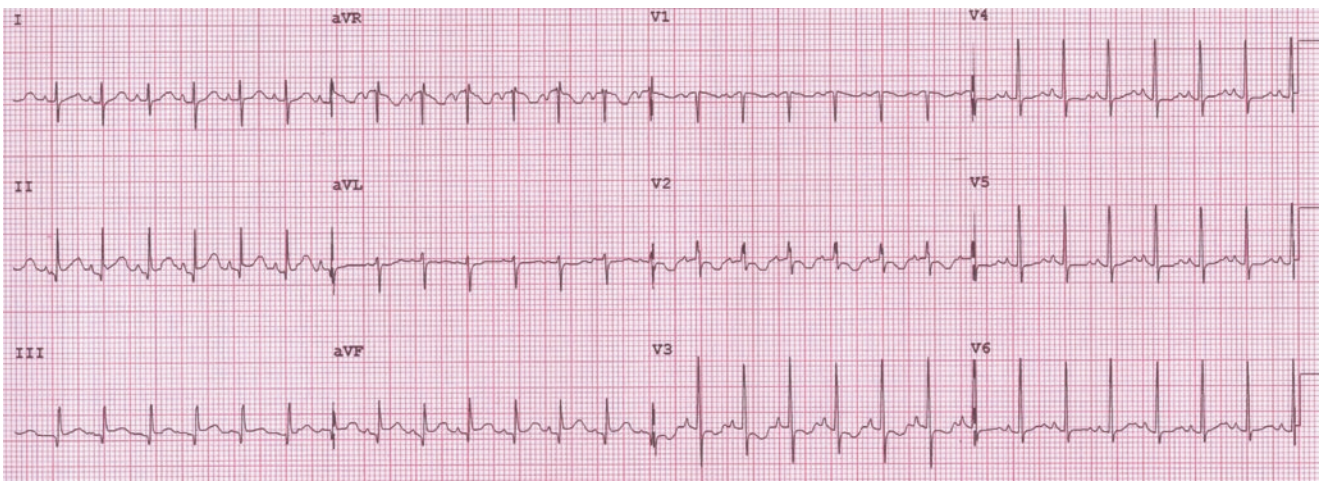

Fig. 6.12 One-month-old female. A left ventricle dominance can be noted. The patient was a severe preterm with systemic hypertension (see Fig. 6.7)

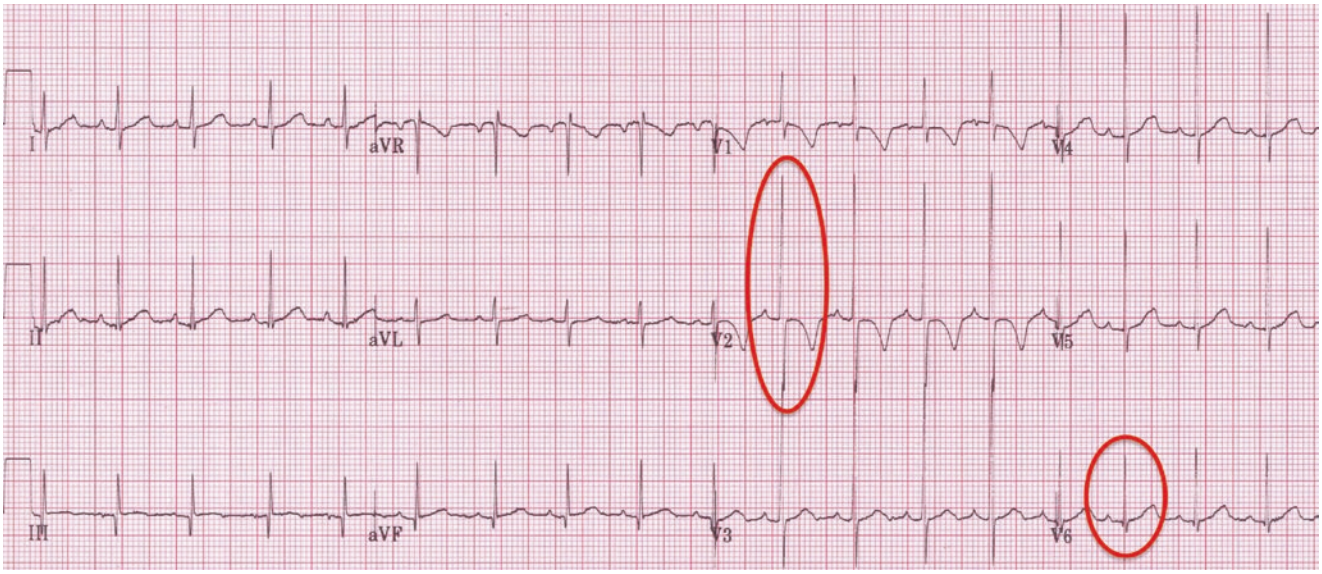

Fig. 6.13 Six-month-old female. In the frontal plane the most isoelectric leads are I and aVL, a fact makes it possible to place the QRS axis around 60° and 90° (second quadrant, physiological for age). The generous R wave in V2 is normal, and also the absence of a deep S wave in V6 excludes right ventricular hypertrophy

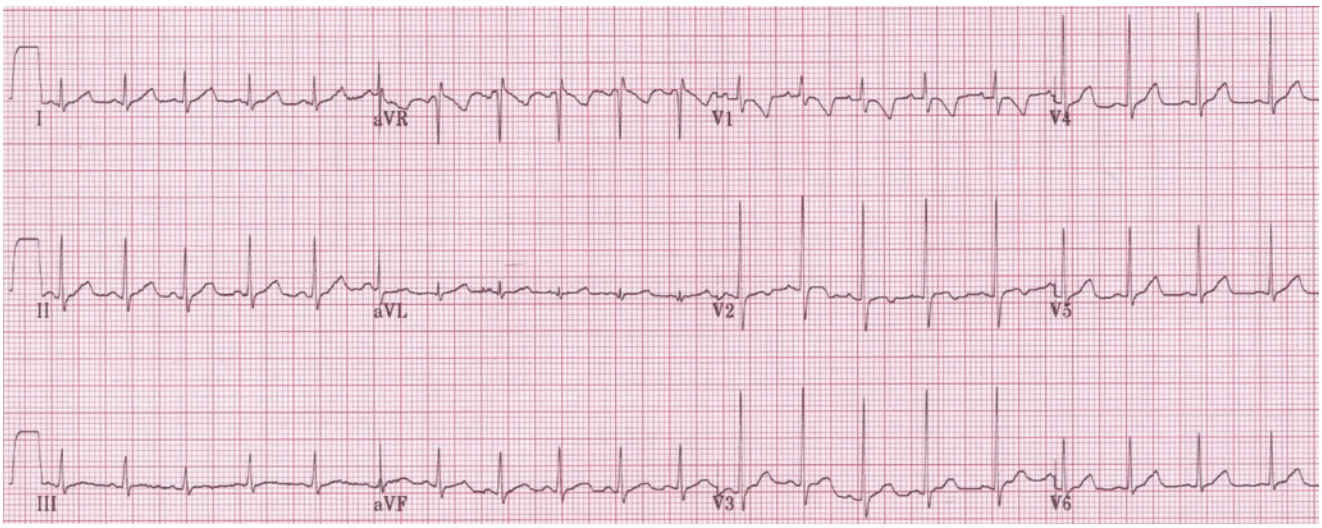

Fig. 6.14 Nine-month-old male, normal

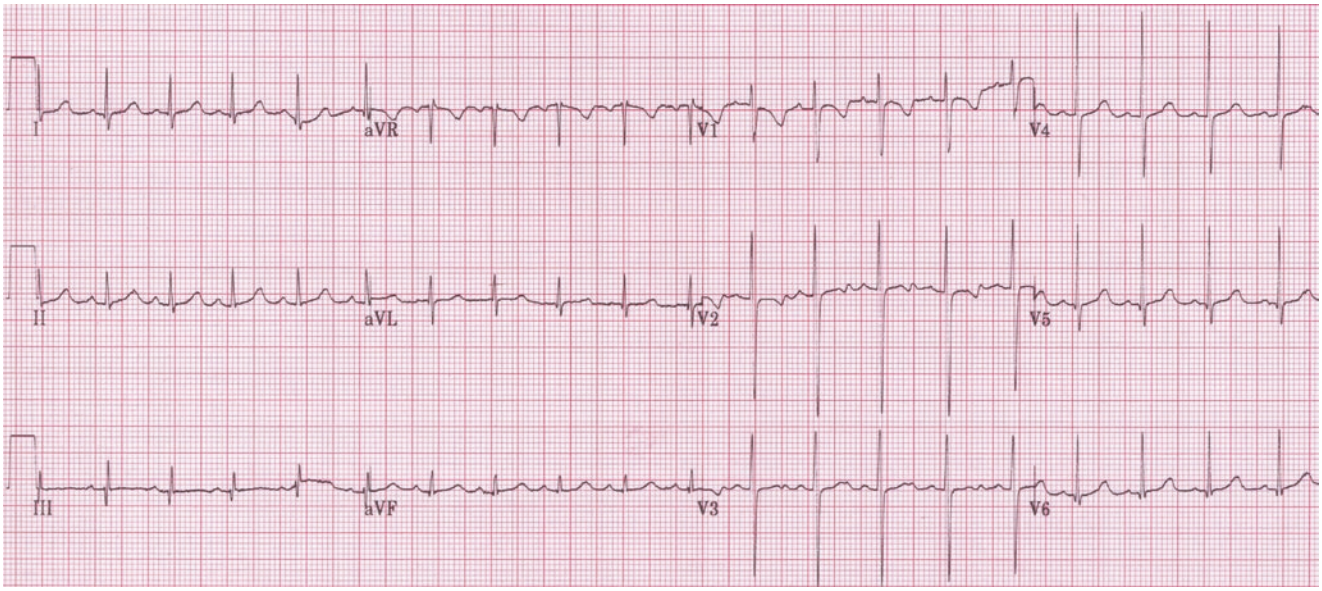

Fig. 6.15 One-year-old boy. Note the notch in the right precordial leads

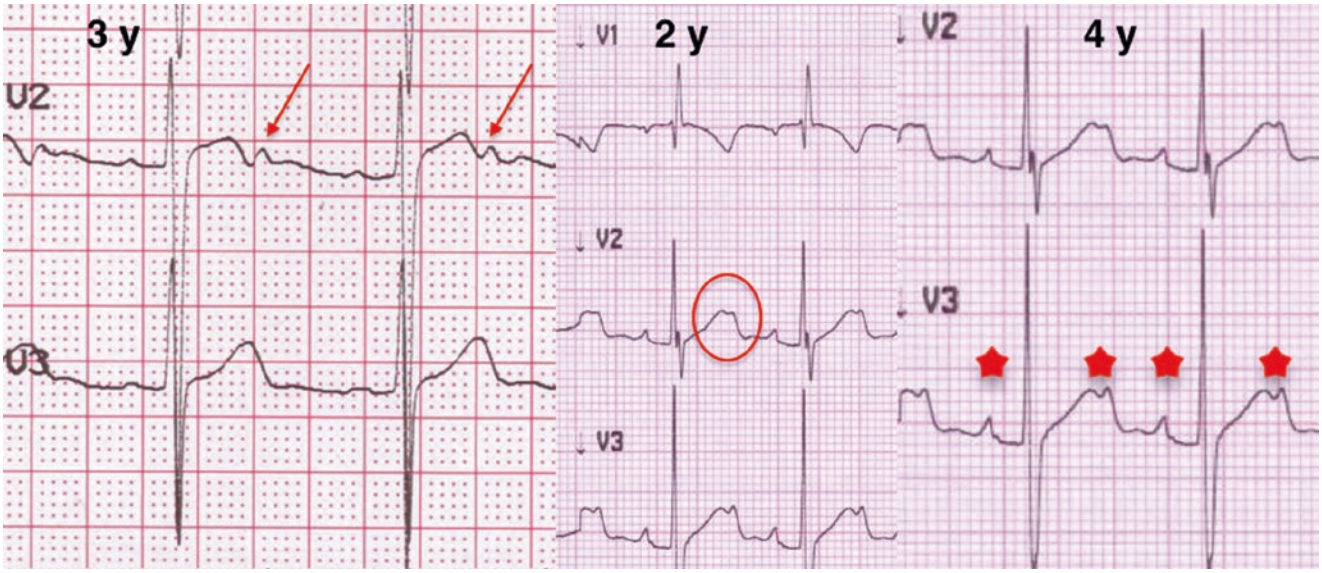

Fig. 6.16 Other examples of right precordial notched T. The notch is normal but can simulate a blocked P

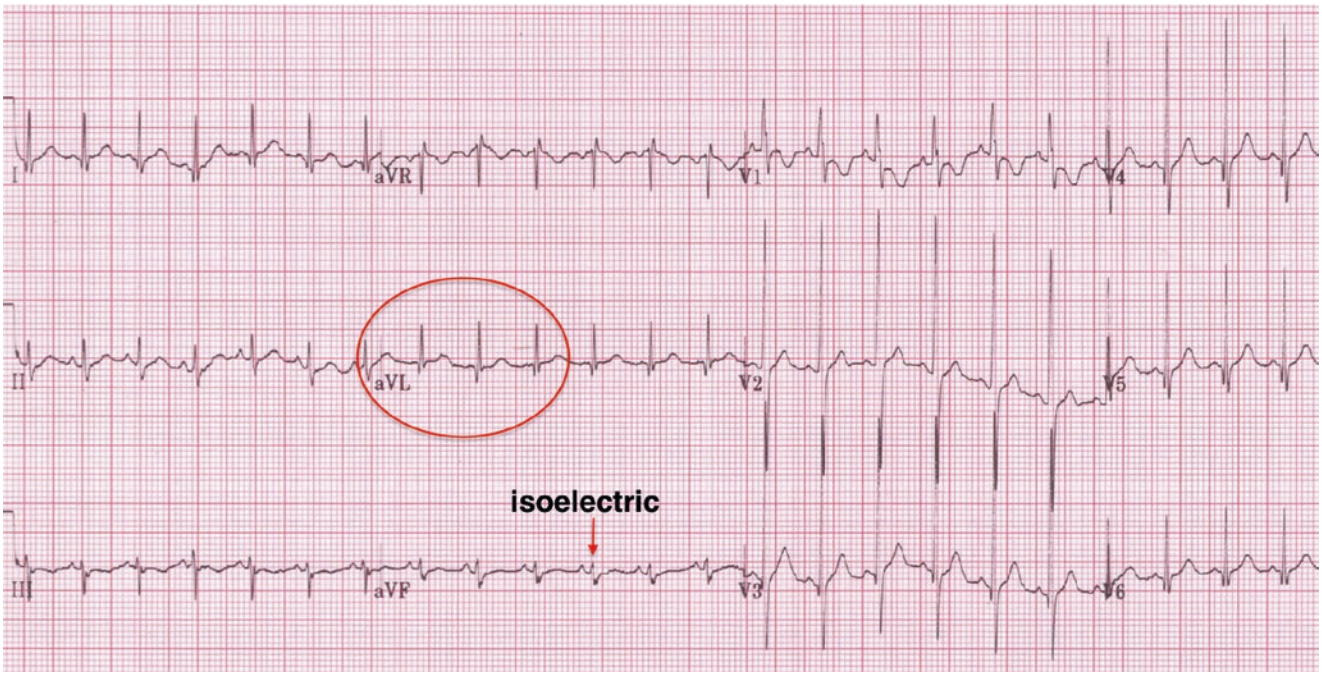

Fig. 6.17 The most isoelectric lead is aVF, placing the QRS axis between the first and second quadrant (between 0°and −15°). Right side voltages are well represented but within normal limits

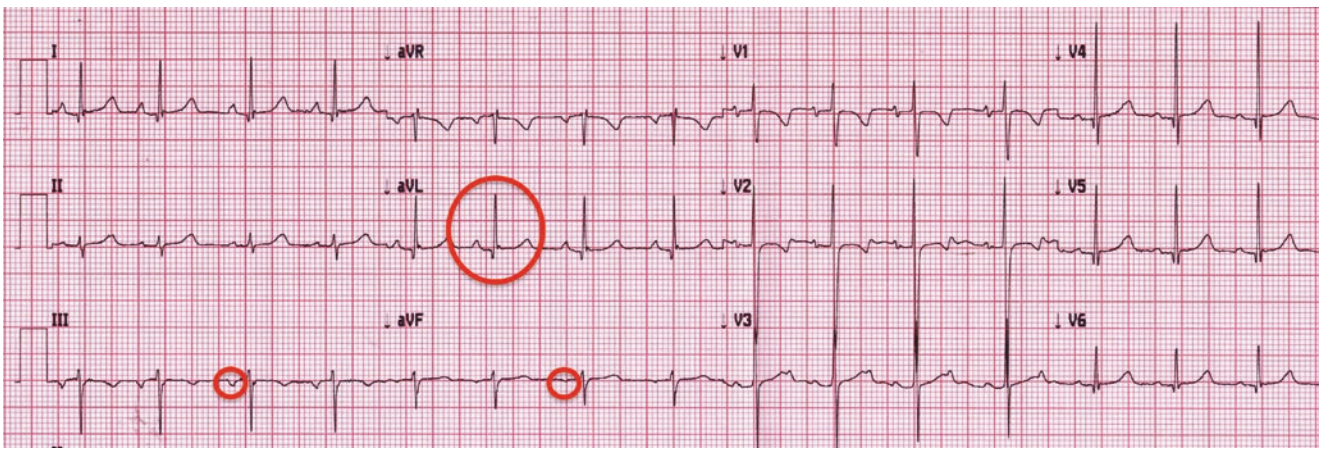

Fig. 6.18 Three-year-old male. The rhythm is not exactly sinus: as you can see, the P wave is negative in lead III and in aVF suggesting the origin of the rhythm in the low right atrium, a normal variant. There is a left axis deviation (−25°). The echocardiogram was normal

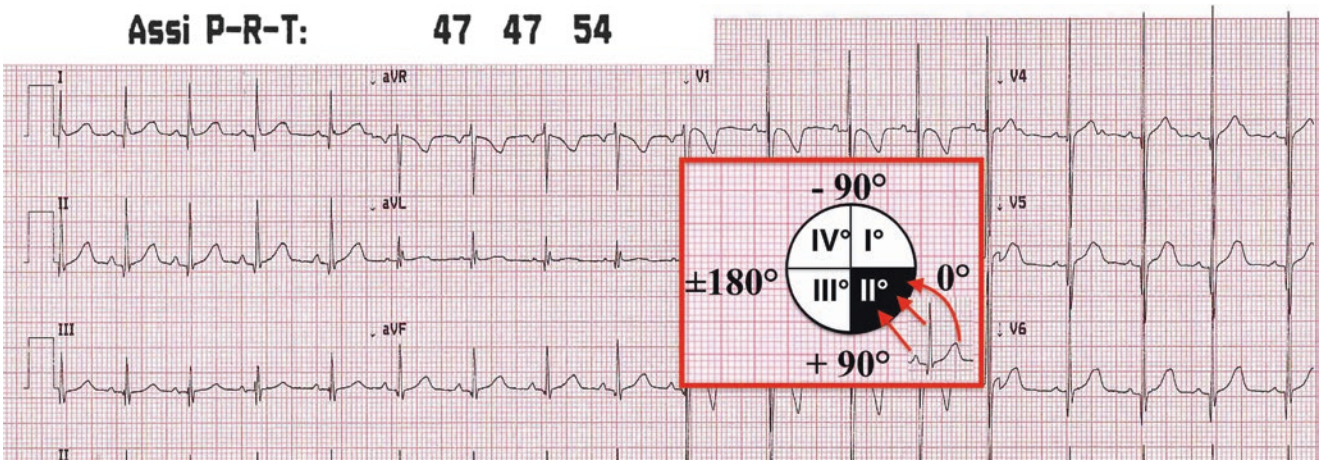

Fig. 6.19 The automatic diagnosis of the ECG machine sentenced "biventricular hypertrophy." Actually, the P, R, and T axes fall off in the second quadrant, coinciding approximately at 50°. The QRS is very narrow, about 80 ms, and the voltage of the precordial leads (which deceived the ECG machine!) is within the limits

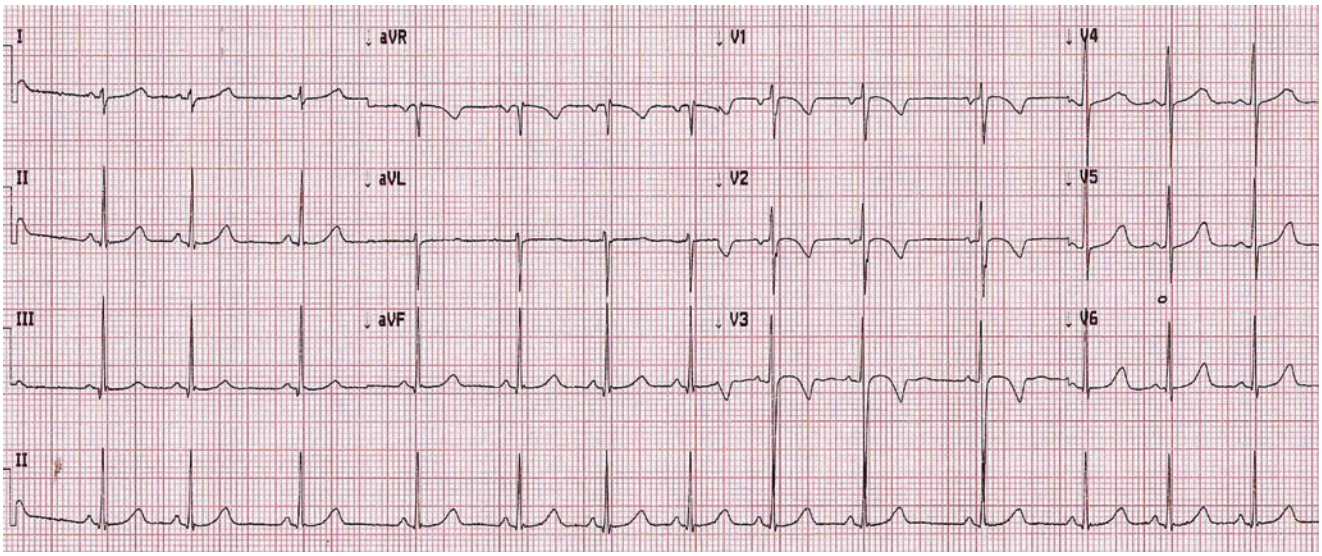

Fig. 6.20 Five-year-old male. The QRS axis is about 90° and therefore lead I is isoelectric. To be a front lead with a 90° angle with respect to the ventricular vector not only involves the isoelectric wave in opposite polarity but also deflections of low amplitude. This explains why in children with the QRS axis between 60° and 90°, leads aVL and I may have low voltages. This is not enough to talk about "low voltages" that needs to be confirmed in different leads, which in fact does not happen here

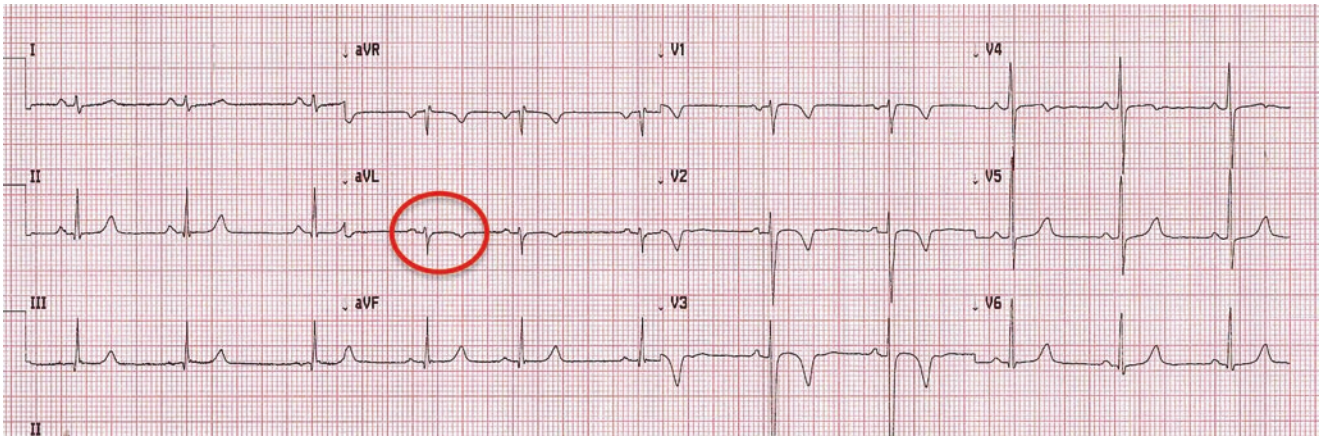

Fig. 6.21 Six-year-old male. The negative T wave in aVL only is non-pathologic

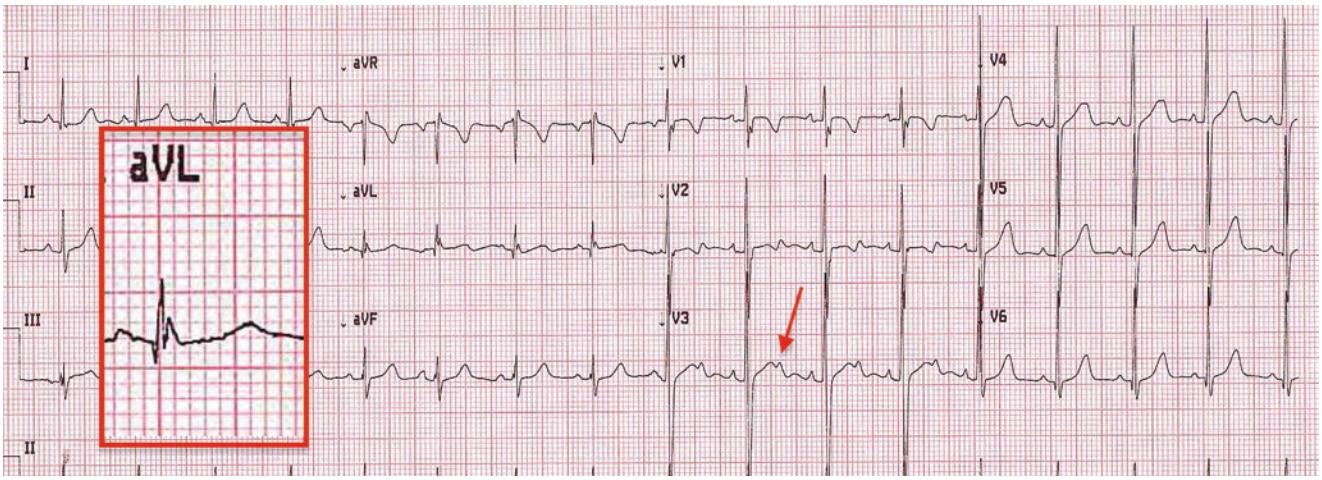

Fig. 6.22 Seven-year-old female. The small and narrow Q wave in aVL is non-pathologic. Note the notched T in V3

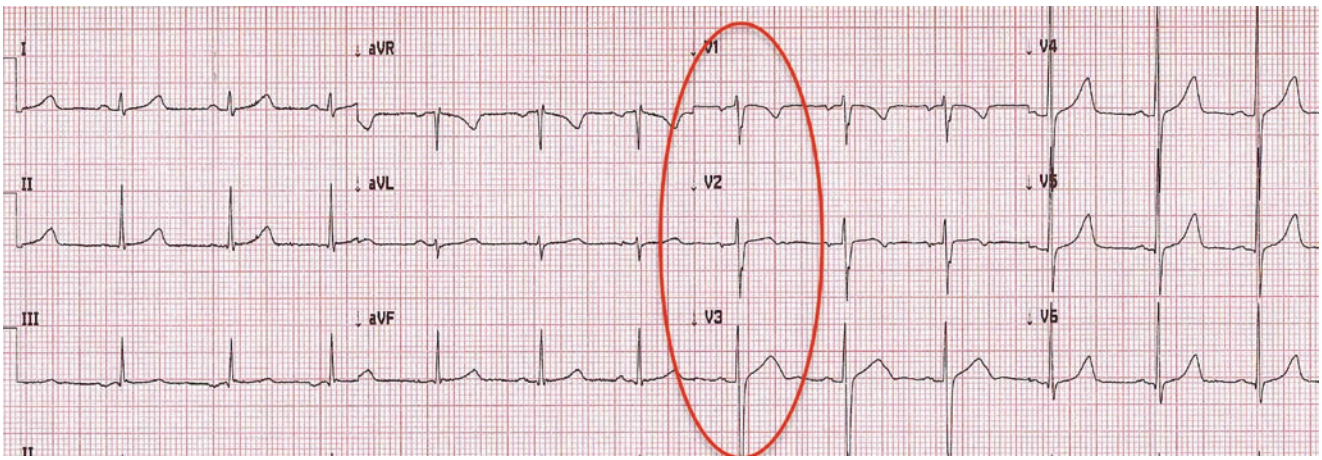

Fig. 6.23 Eight-year-old male. The "juvenile pattern" with negative T waves in all precordial leads is now evolving to the adult pattern. The T wave is positive in V3 and biphasic in V2

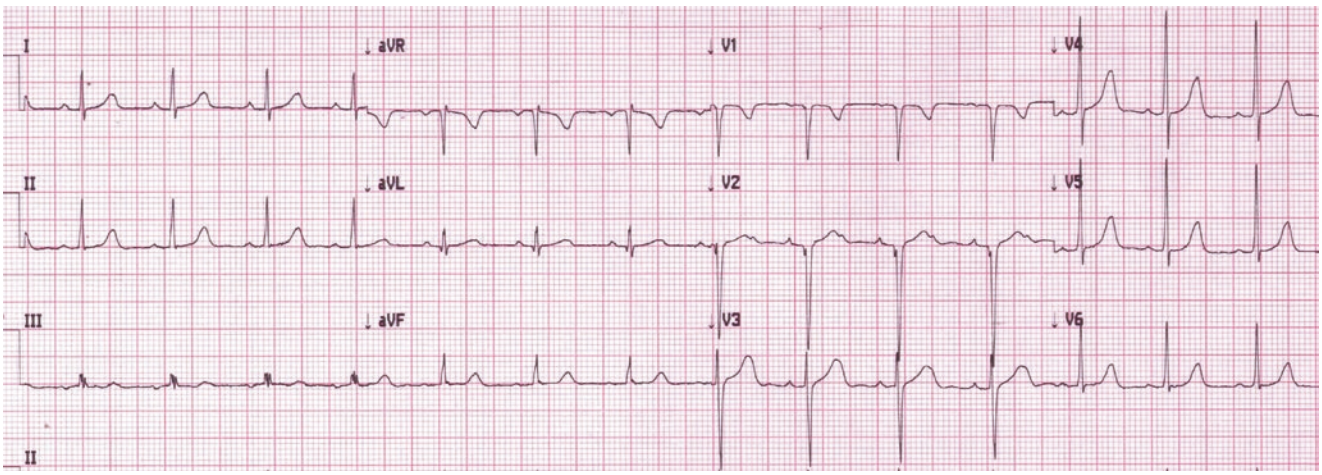

Fig. 6.24 Nine-year-old female. There is a poor progression of the R wave in the right precordial leads. In the absence of other signs, this can be attributed to a physiological clockwise rotation of the heart on the horizontal plane

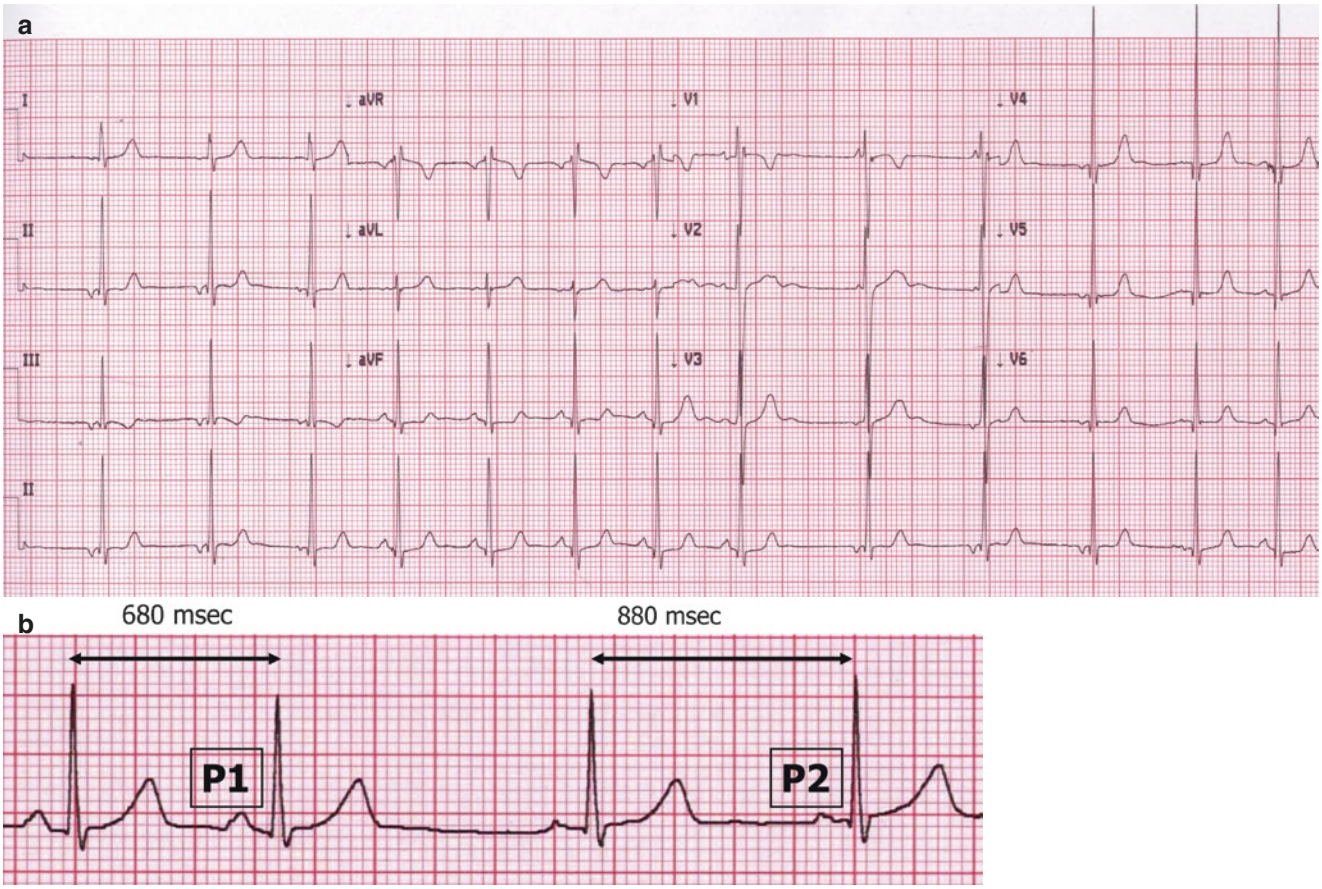

Fig. 6.25 (**a**) Eight-year-old male patient referred for "arrhythmia." The first two beats are part of the short cycle, in opposition to the third and fourth forming part of the long-phasic arrhythmia cycle. (**b**) Close-up. The P waves of the two cycles are slightly different: P1sinus, P2 parasinus, more caudal than the first. That diversity can still be considered as part of phasic respiratory arrhythmia

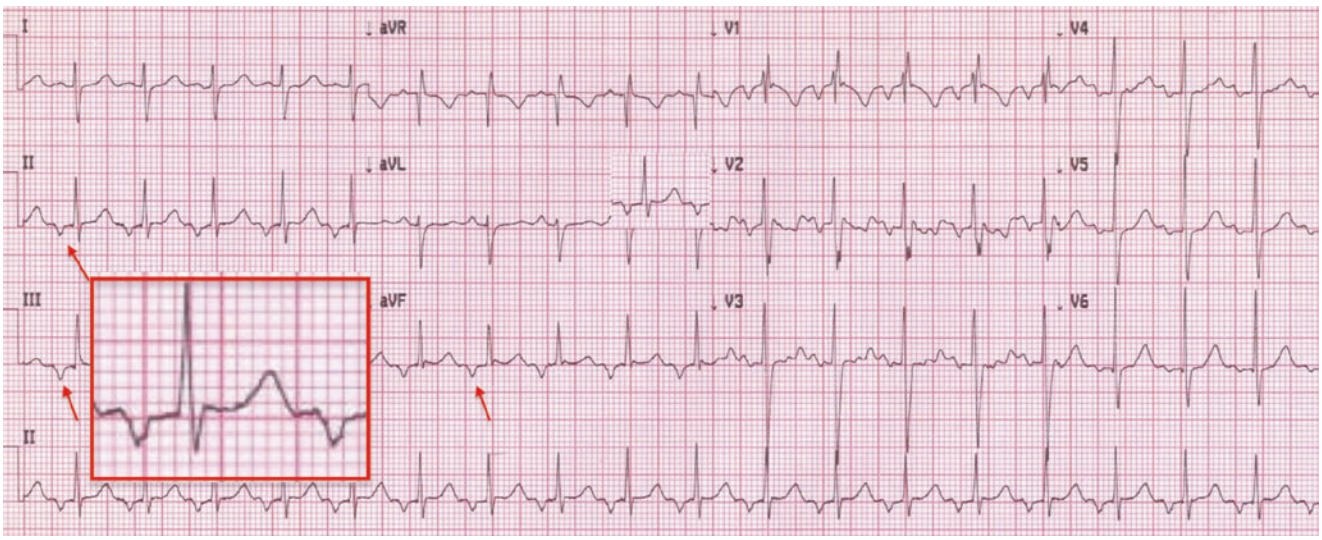

Fig. 6.26 Nine-year-old male, bicuspid aortic valve and LPSVC. The low atrial rhythm, also called coronary sinus rhythm (arrows indicate the negative P), may be considered benign and physiological. At times it is caused by dilation of the coronary sinus, a result of drainage in it of the left superior vena cava. The LPSVC may be a normal variant (0.1–0.5%), but may be associated with heart disease from bicuspid aortic valve to tetralogy of Fallot (in this case up to 30%). Therefore a low atrial rhythm does not always require an echocardiogram, but in terms of history and objectivity, it could recommend it

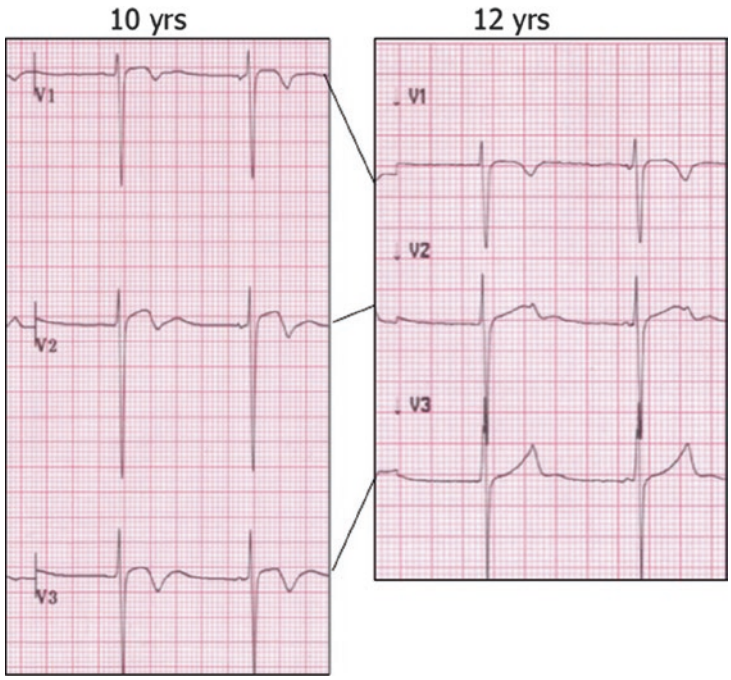

Fig. 6.27 Transition from juvenile to adult pattern in the precordial leads of a boy

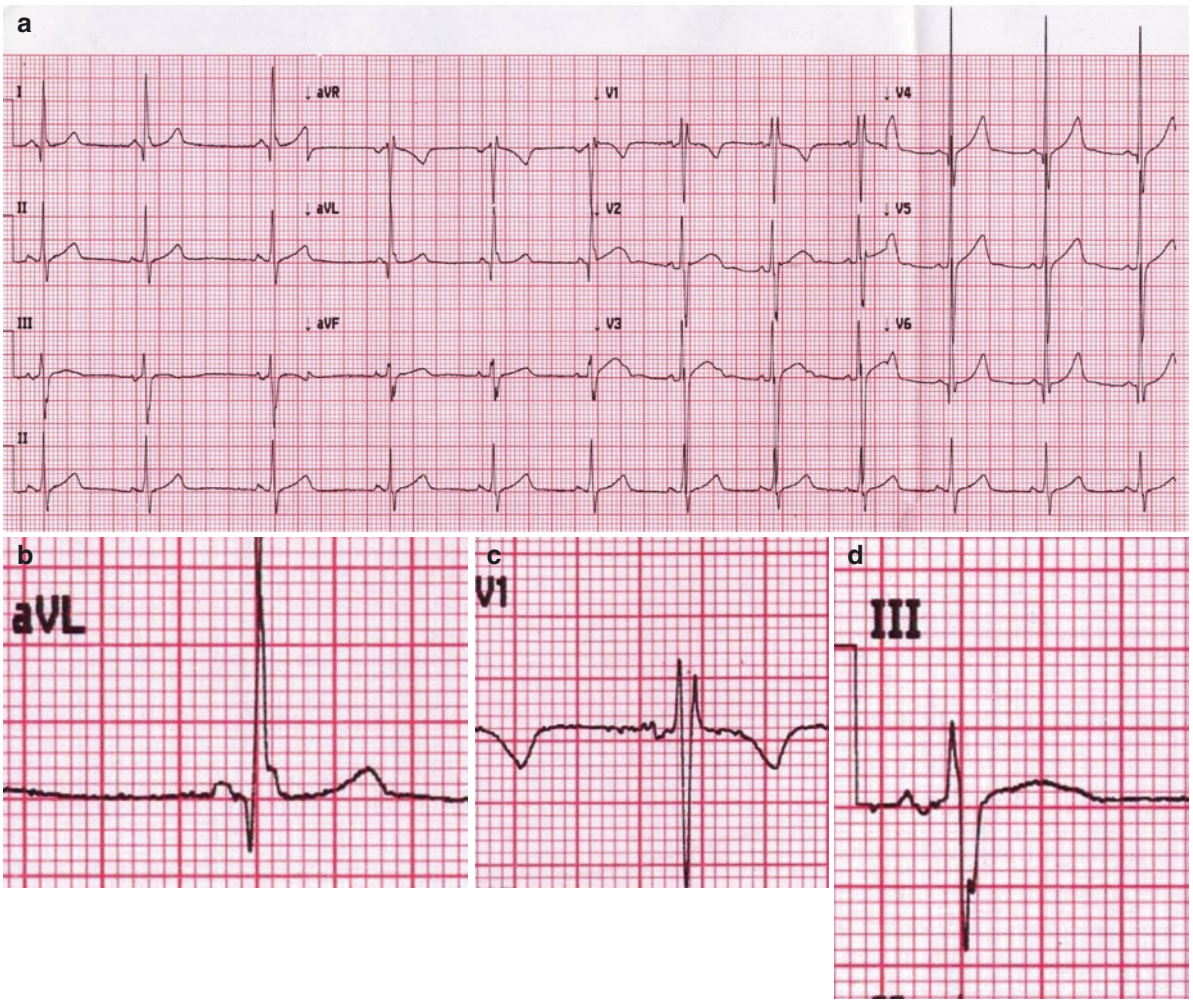

Fig. 6.28 (**a**) Twelve-year-old boy. The QRS axis is between the first and the second left quadrant, about 0°. Q waves in leads I, aVL (**b**), and V6 are physiological (3.5 mm high and last 20 ms). The initial part of R wave in leads II and III (**c**) is slurred, but there are no delta waves, although the PR is relatively short. In V1 (**d**) there is a conduction delay but not enough to merit being called RBBB; in the other precordial leads, you can see a "notch" of T, of no pathological significance

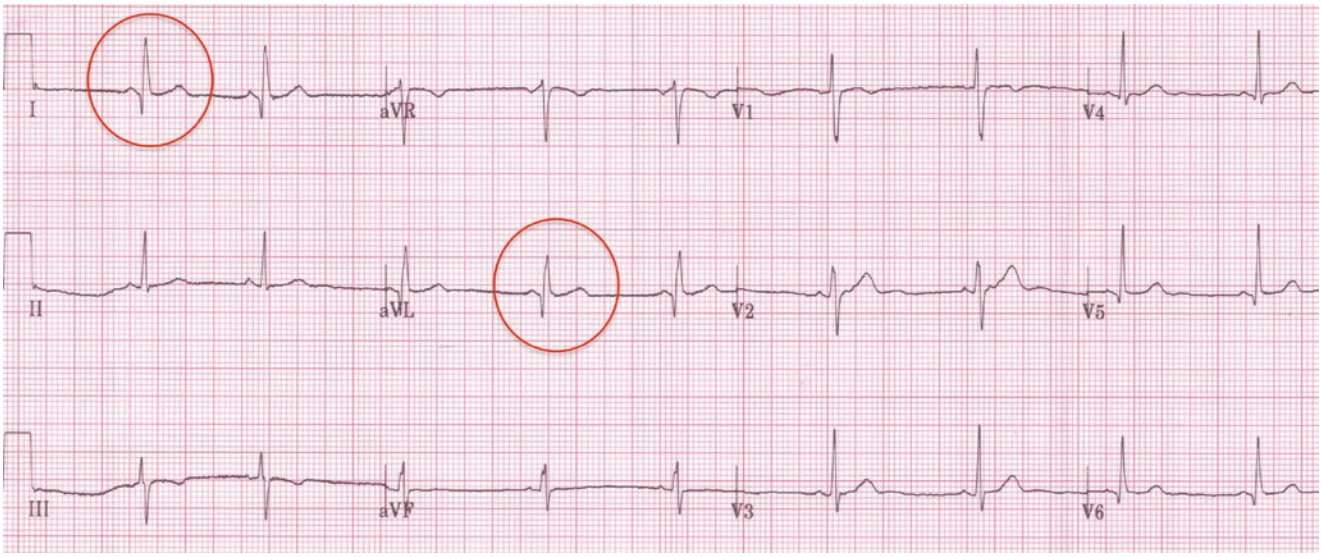

Fig. 6.29 Eighteen-year-old female. Adult pattern. The Q wave in aVL is more than 25% of the R wave. However it is narrow (20 ms in duration) and not worrying

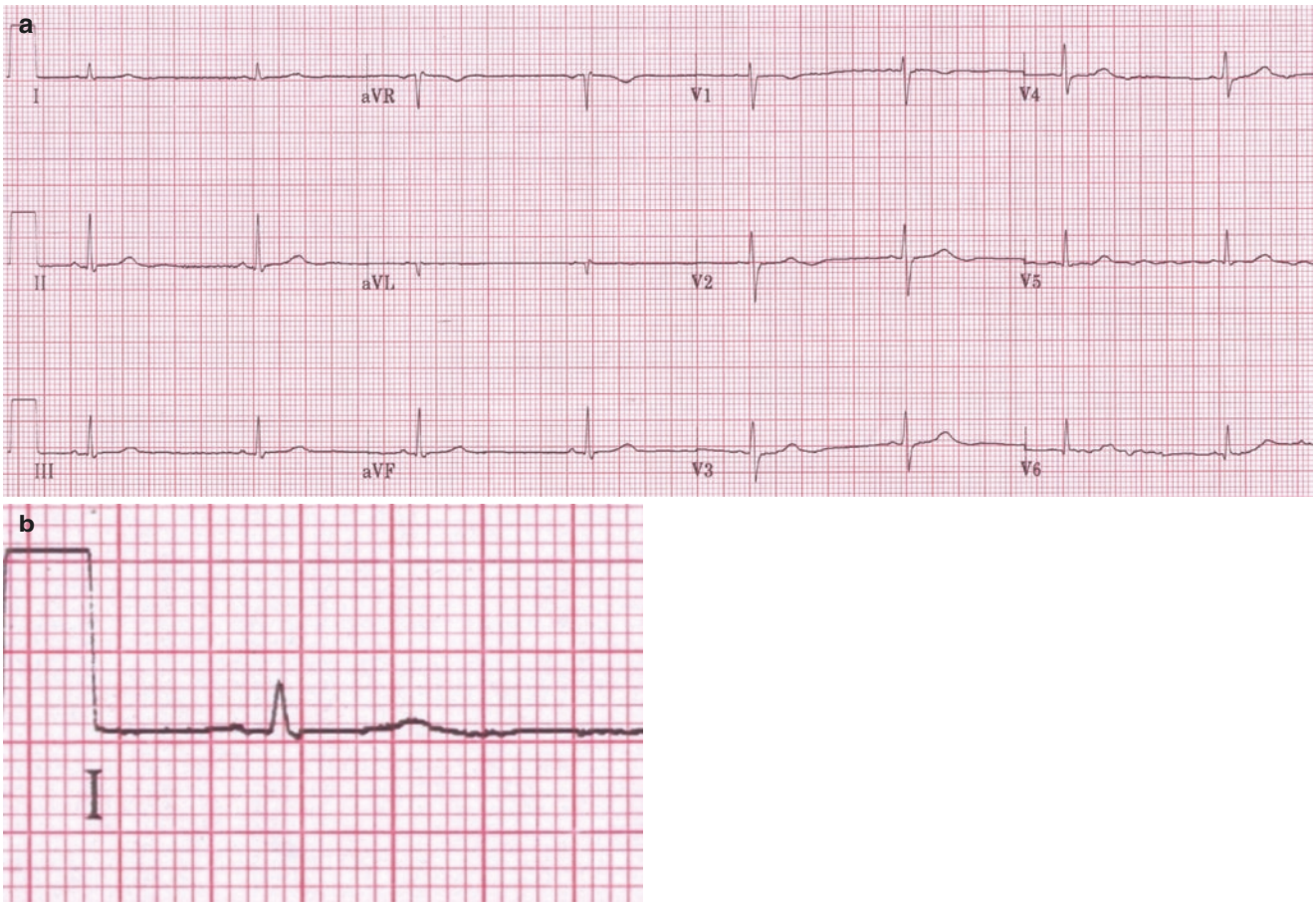

Fig. 6.30 (**a**) Sixteen-year-old girl. Bradycardia, low voltages, and repolarization abnormalities – from non-specific to QT prolongation – are distinctive features of anorexia and prolonged fasting. The heart goes into a sort of lethargy. (**b**) Close-up

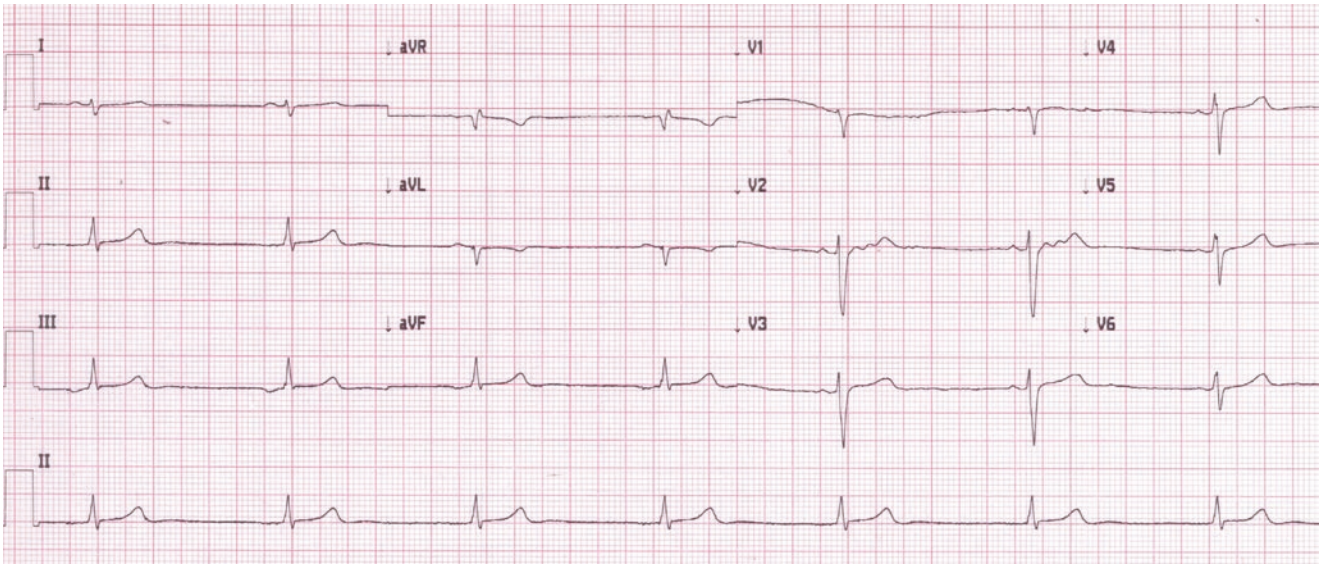

Fig. 6.31 Fifteen-year-old male. Anorexia. Low right atrium rhythm

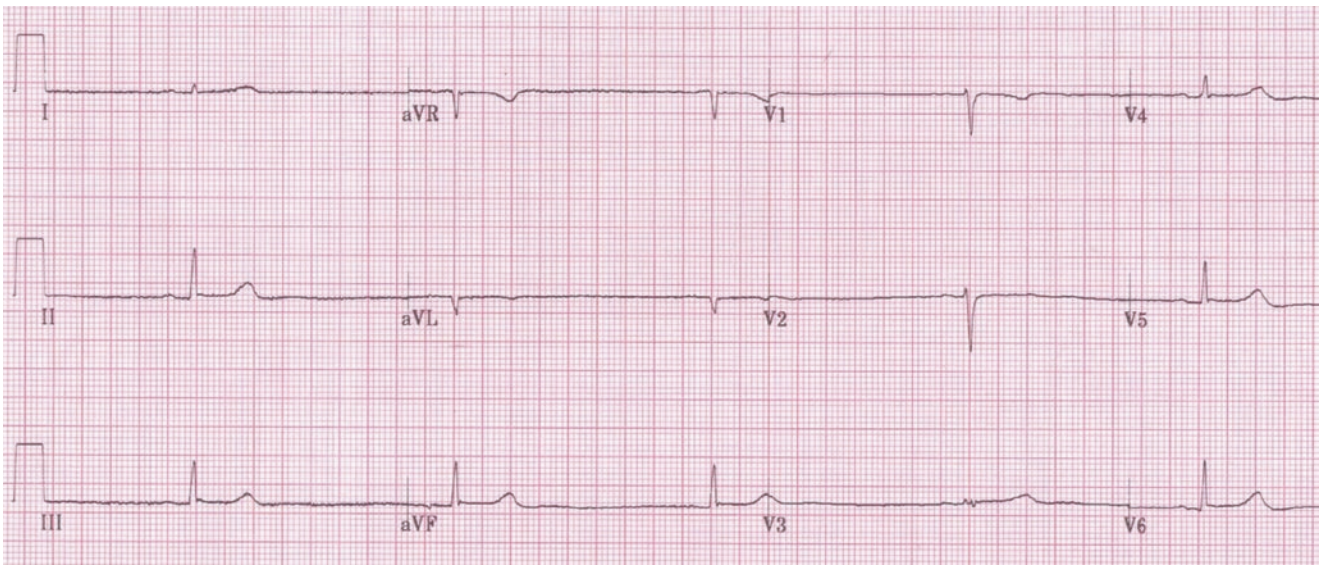

Fig. 6.32 Sixteen-year-old female. Anorexia. The body weight is 30 kg

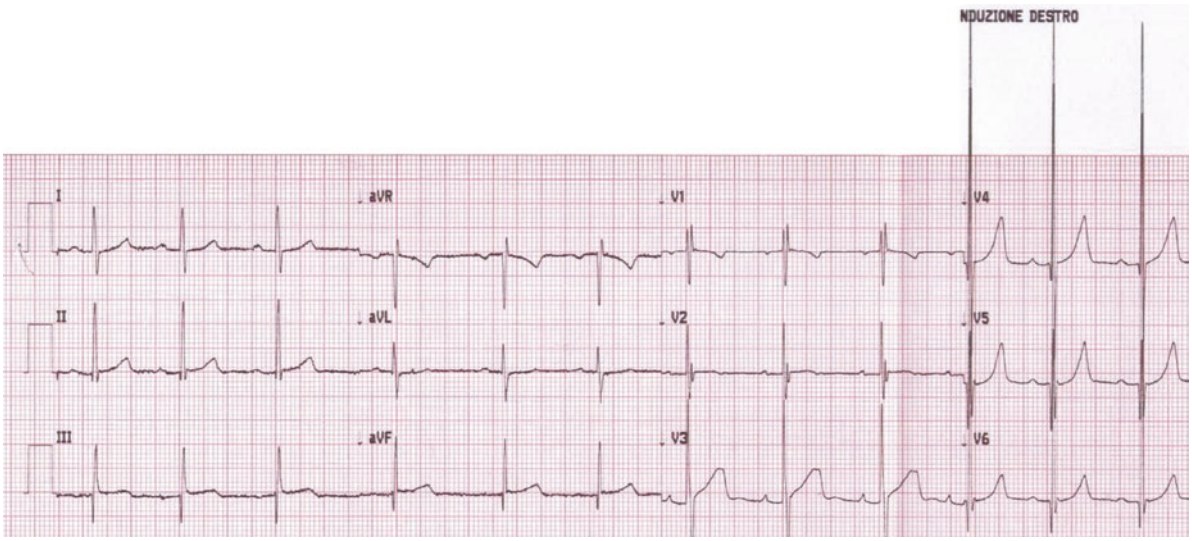

Fig. 6.33 Sixteen-year-old, weight 30 kg. This boy has the same age and weight as the previous case (Fig. 6.32) but the ECG is completely different!

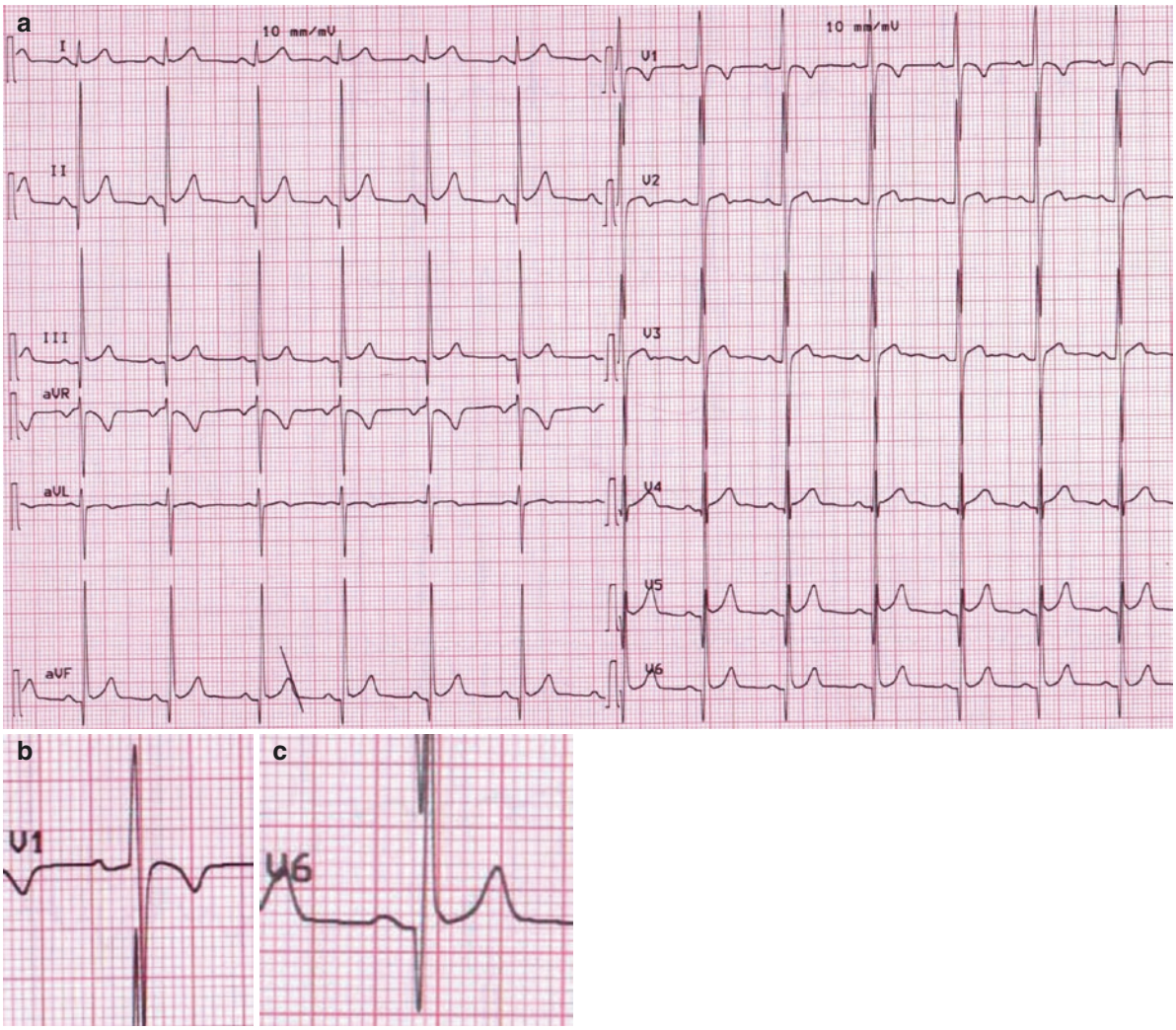

Fig. 6.34 (**a**) Twelve years old. In the right precordial leads, the R/S ratio is high (**b**), along with deep Q waves in the left precordial leads (**c**) suggesting the pattern of Duchenne dystrophy. The cardiological and neurological examinations were normal. It is a normal variant

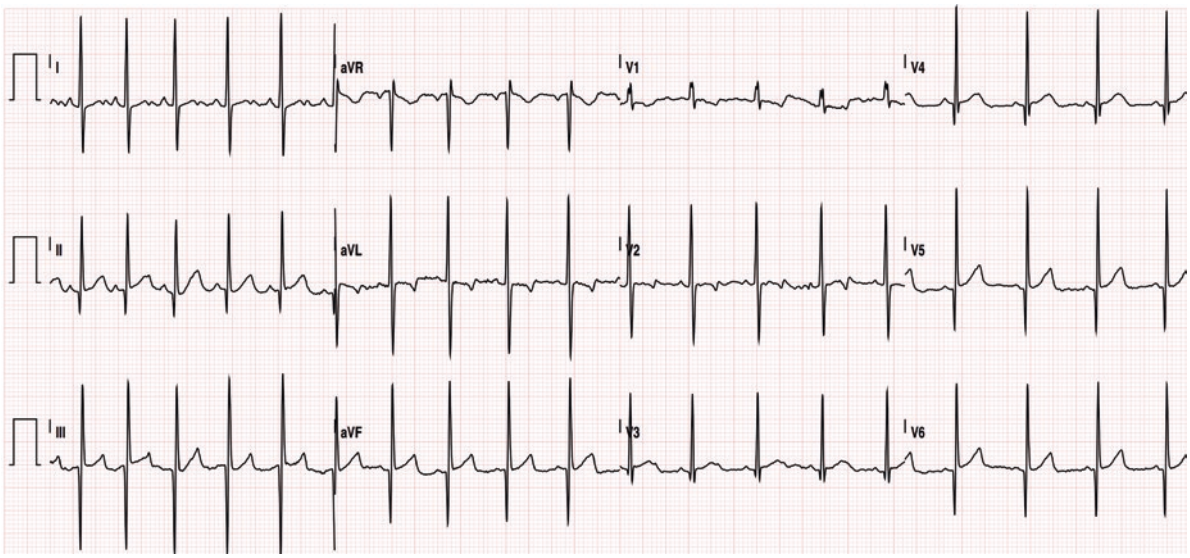

Fig. 6.35 One-year-old boy. Impressively deep Q waves. A full work-up was negative. However, it is reassuring not to find Q waves in aVL and I, a feature that otherwise suggests ALCAPA

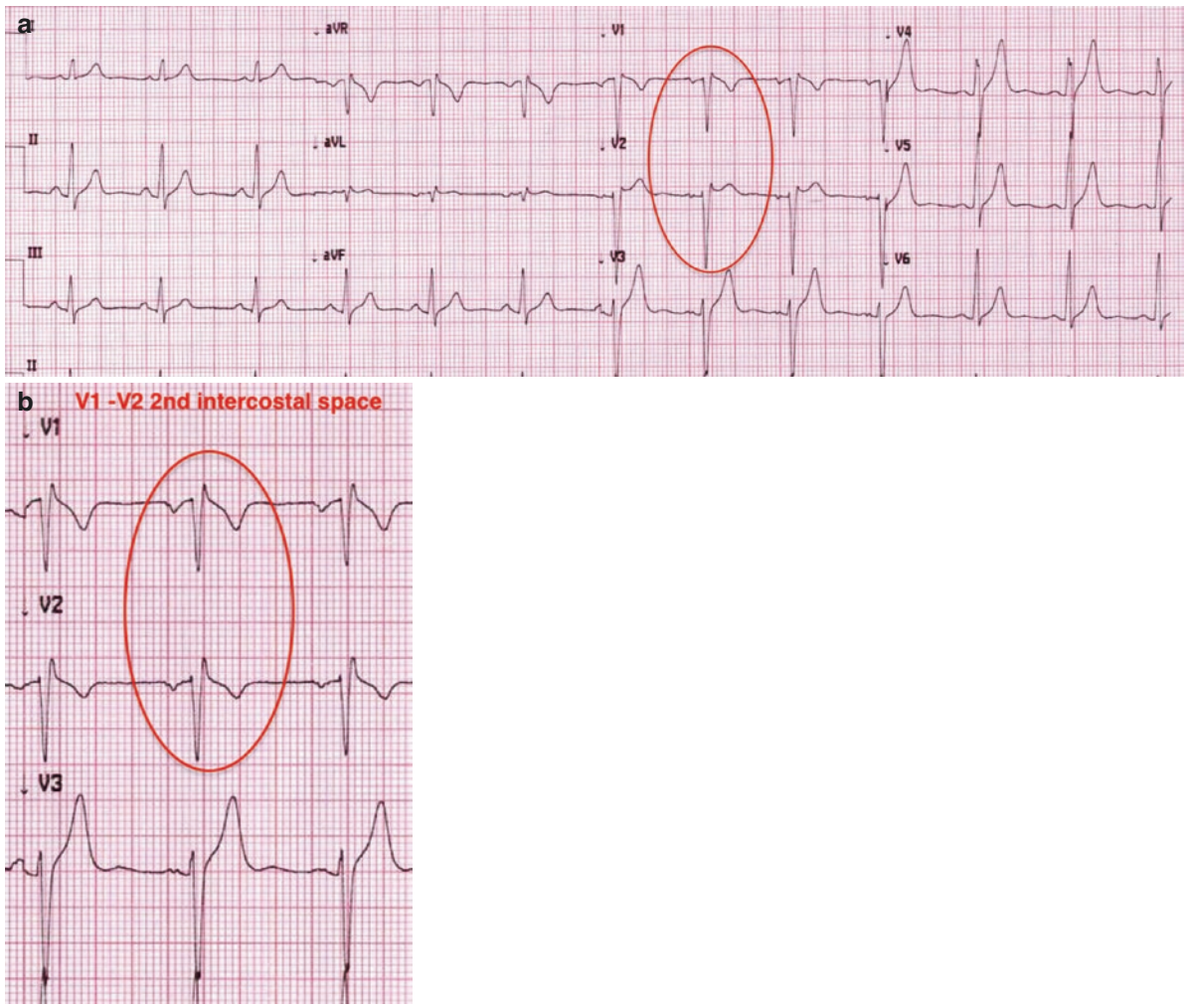

Fig. 6.36 (**a**) Twelve-year-old boy. The screening ECG in the absence of symptoms or family history raised the suspicion of Brugada pattern. (**b**) Moving the right precordial leads to the third and the second intercostal space, the result is reassuring, more suggestive of incomplete RBBB

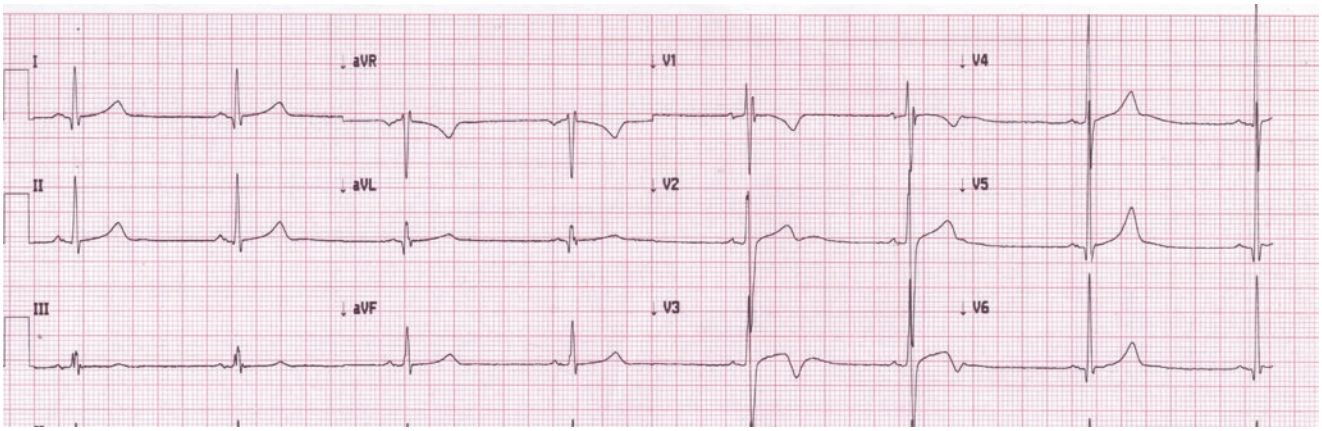

Fig. 6.37 Sixteen-year-old male. Mild right intraventricular conduction delay and normal variant pattern in V2-V3

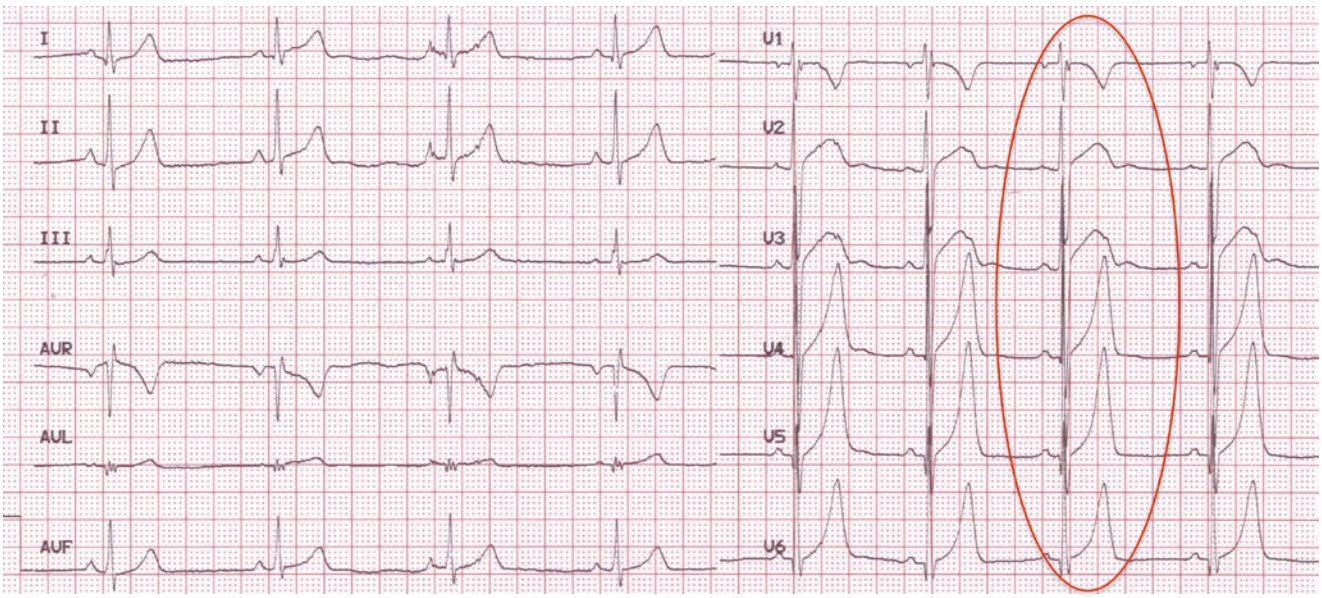

Fig. 6.38 Seventeen-year-old girl, so-called "vagotonic" normal variant. Sinus bradycardia and tall T waves in the precordial leads can be noted

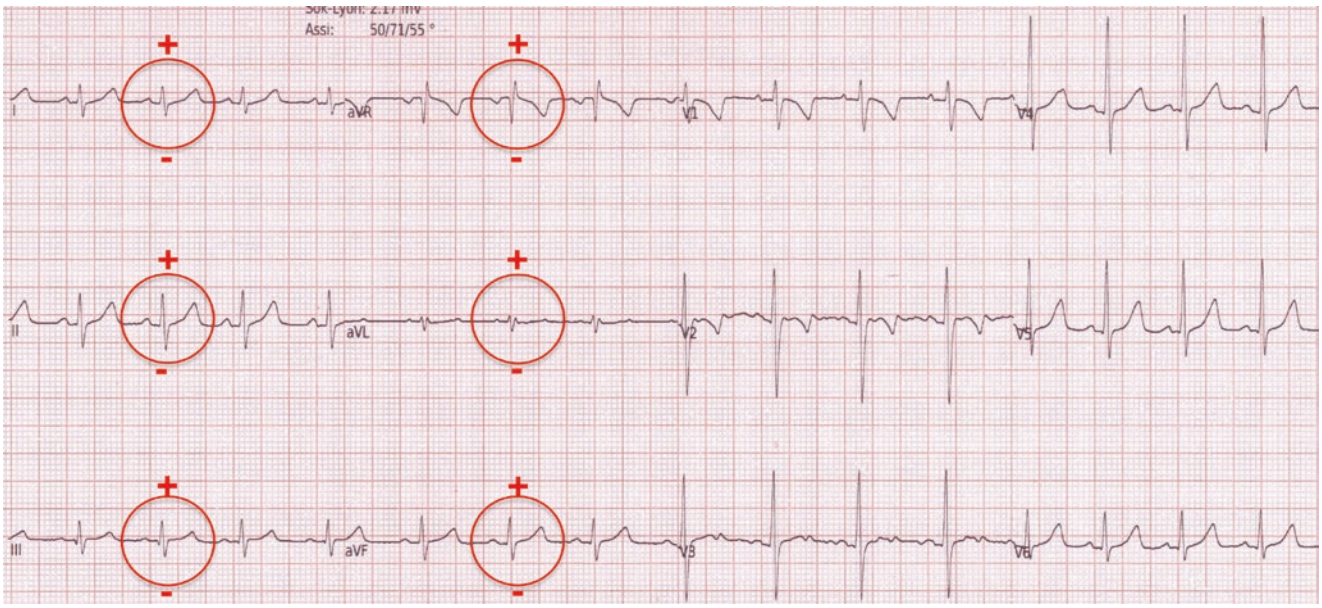

Fig. 6.39 Eleven-year-old male. In the frontal plane all the leads are isoelectric. In this case it is correct to answer "indeterminable QRS axis"

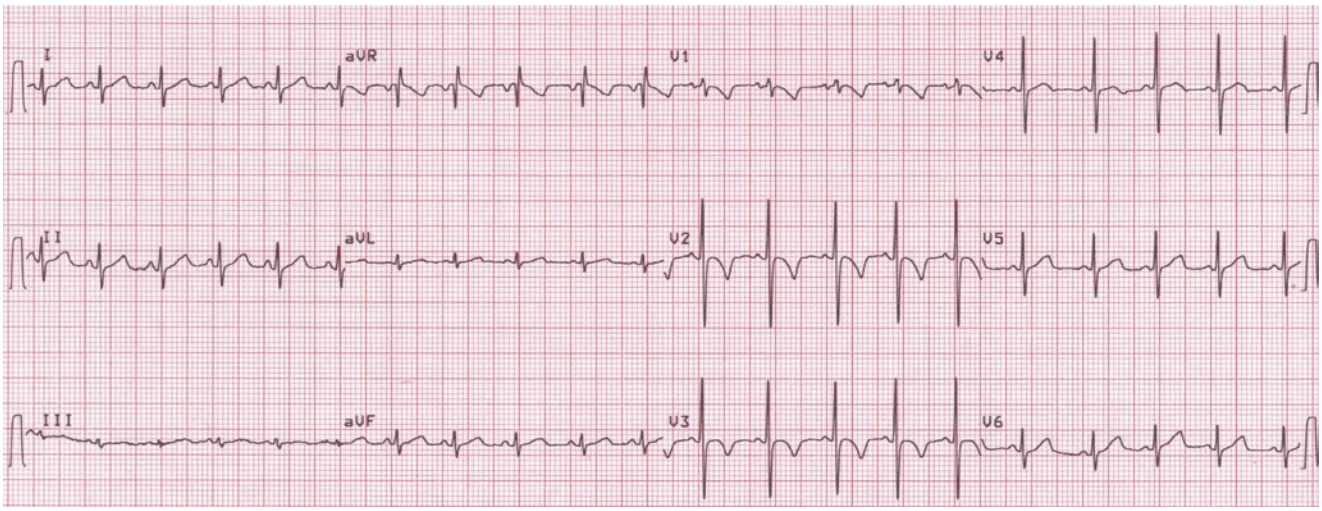

Fig. 6.40 Seven-month-old girl. This is a rare case of "complete isoelectricity." All leads are isoelectric. The echocardiogram was normal

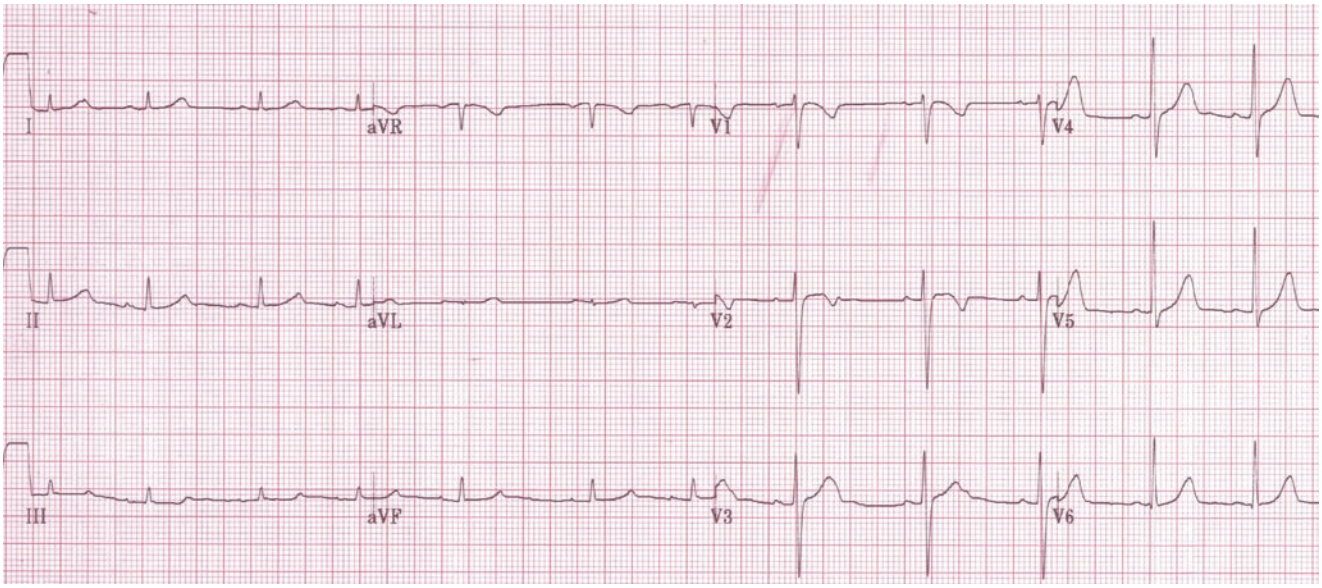

Fig. 6.41 Seven-year-old male. Low voltages in the frontal leads. This pattern is aspecific compatible with a normal variant, cardiomyopathy, myxedema, and anorexia. In this case, the patient was affected by rickets

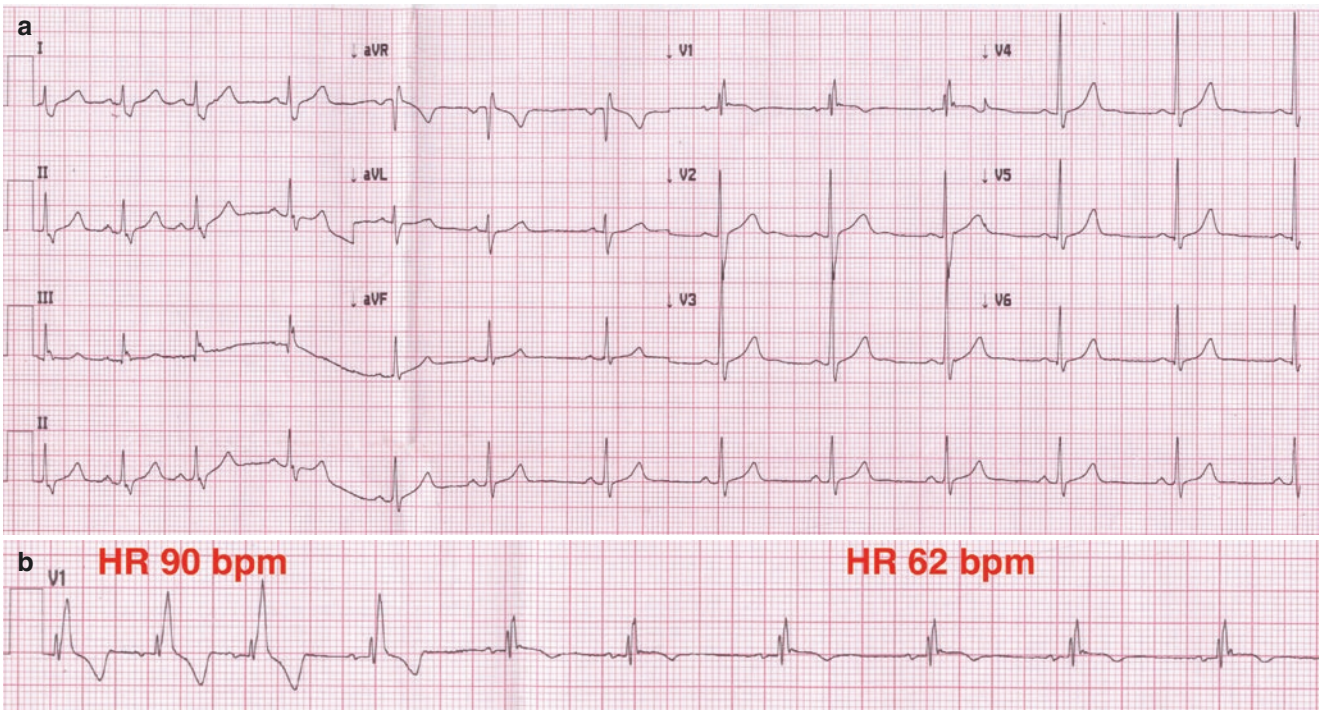

Fig. 6.42 (**a**) Thirteen-year-old male. An incomplete RBBB can be noted. (**b**) Close-up, the RBBB is not stable and depends on HR

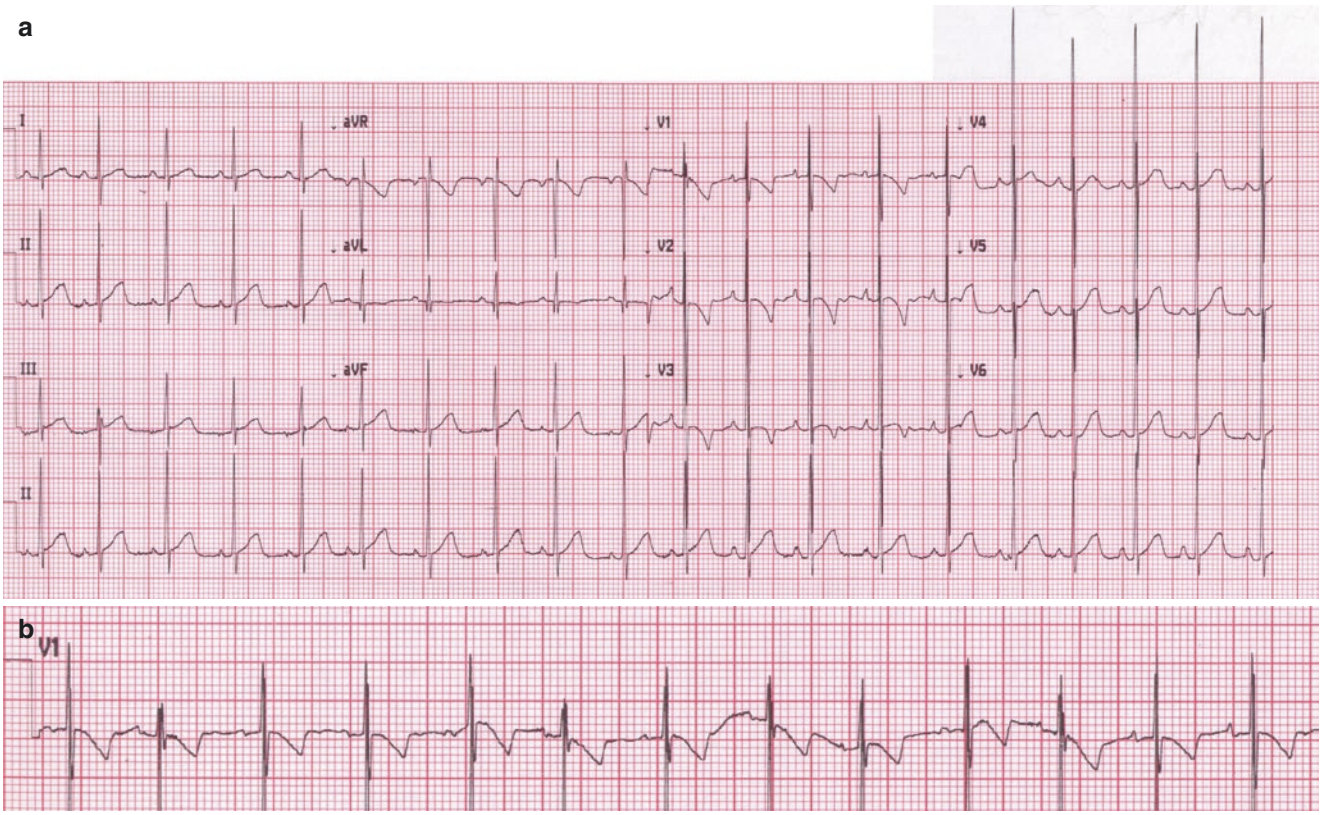

Fig. 6.43 (**a**, **b**). Five-year-old boy. The difference in R wave voltage is due to a breathing artifact

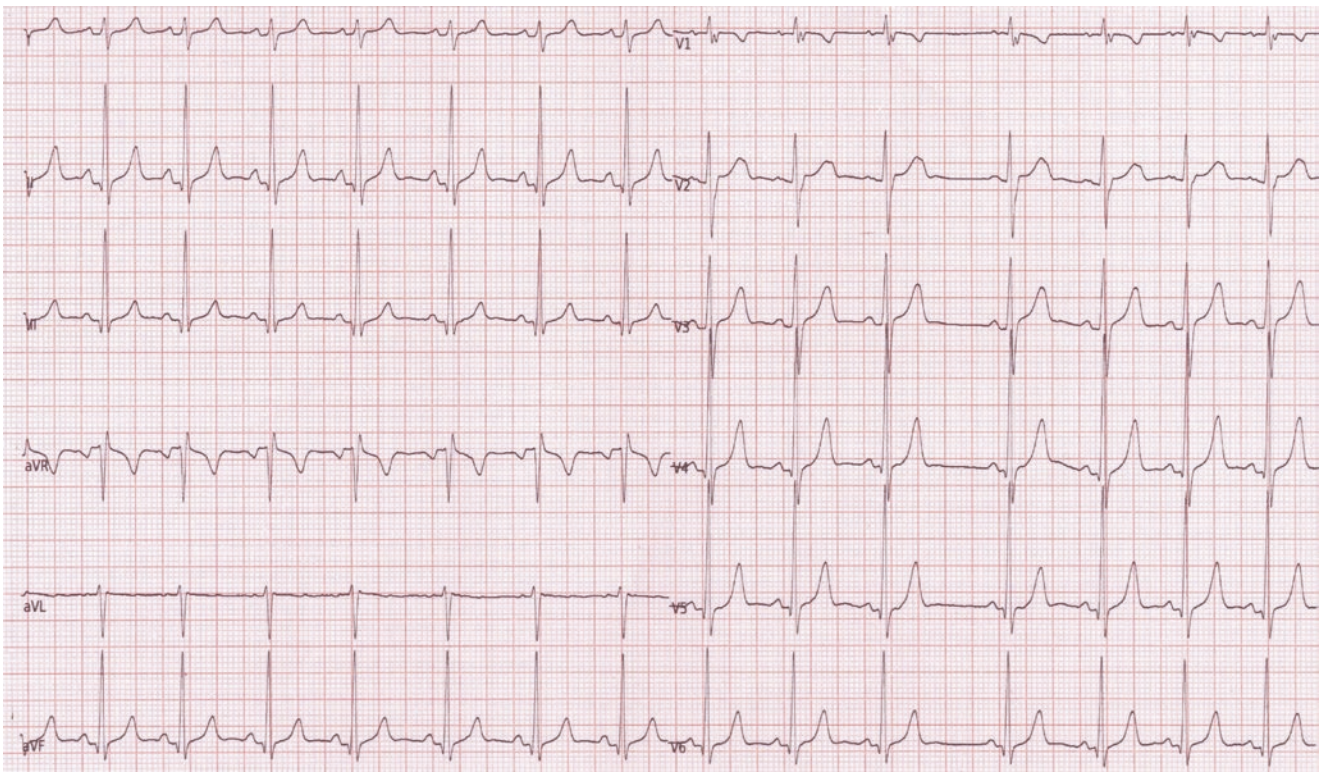

Fig. 6.44 Six-year-old boy. Lead I is isoelectric, placing the QRS axis at 90°, the so-called "vertical heart." Normal ECG

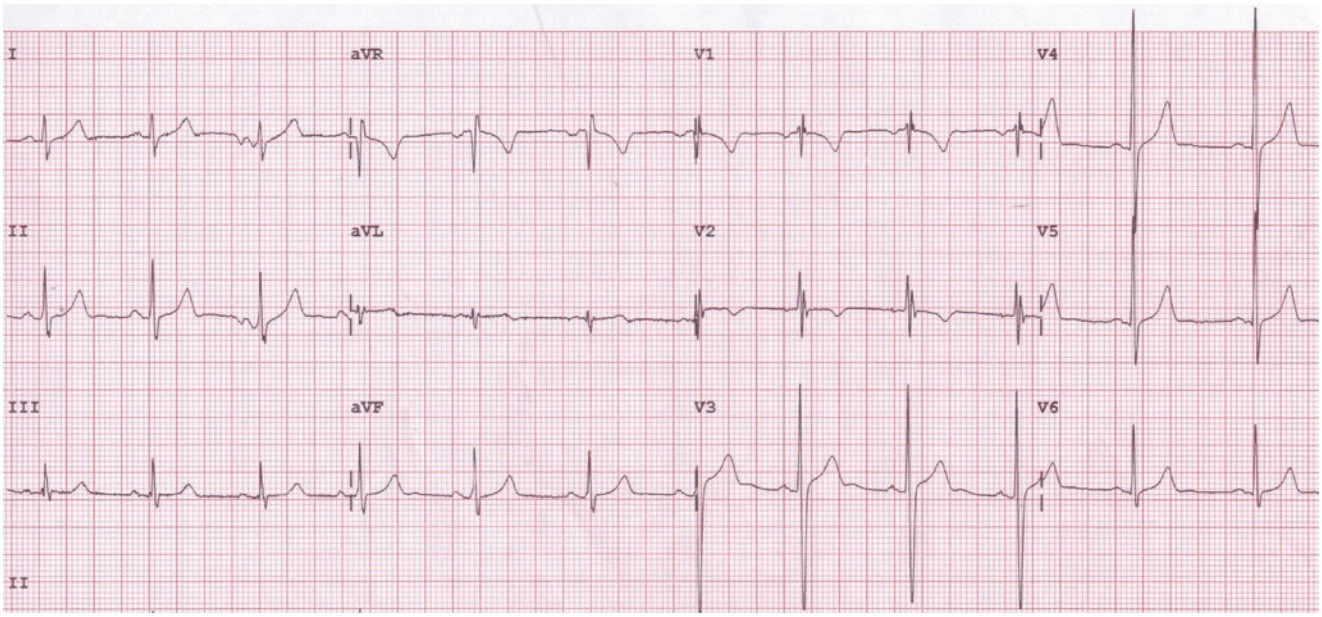

Fig. 6.45 Seven-year-old boy. Right intraventricular conduction delay; vertical heart. Normal ECG

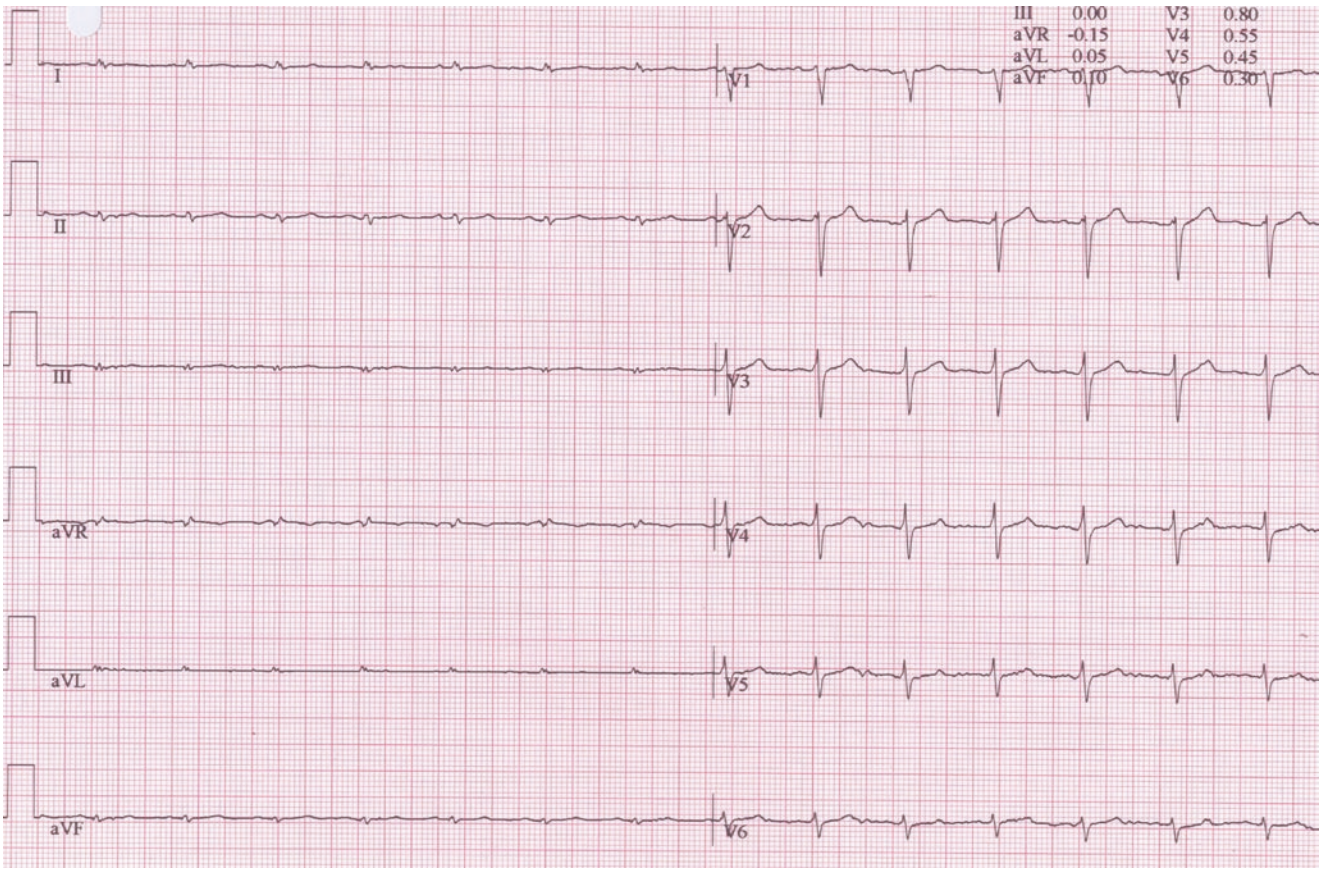

Fig. 6.46 Twelve-year-old boy. In this obese youngster, the voltages are lower than expected. Low voltages are described in obesity, but in this case the low voltages are impressive and merit follow-up to rule out an underlying cardiomyopathy or storage disease (i.e., amyloidosis)

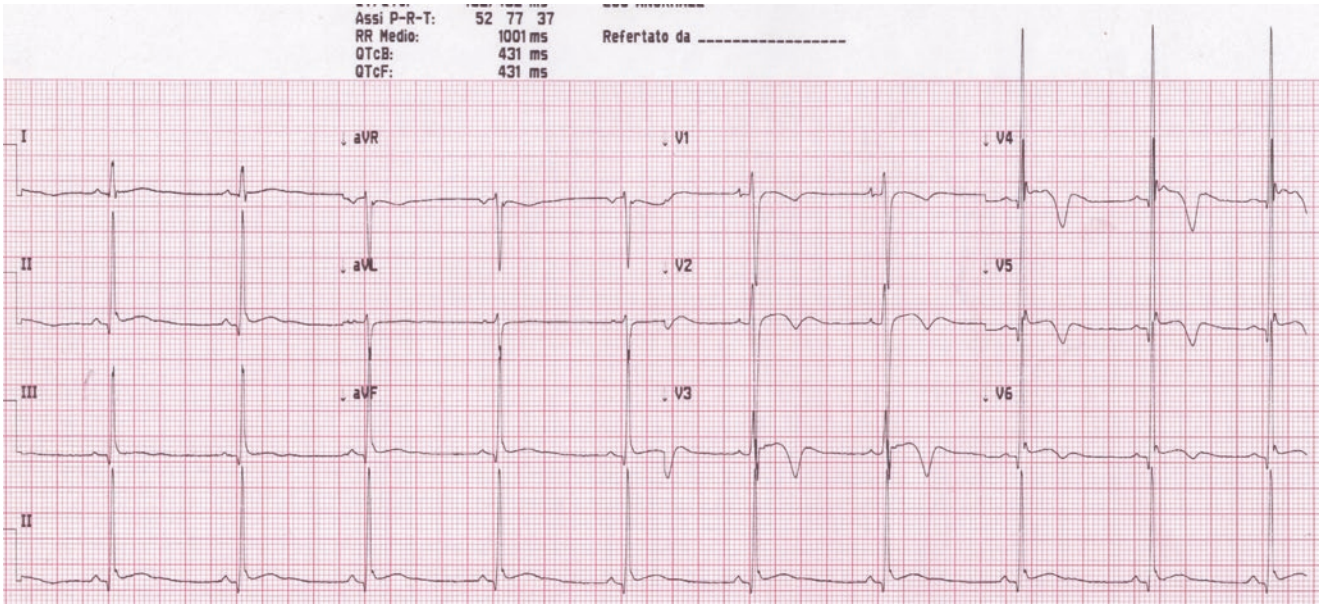

Fig. 6.47 Fifteen-year-old black athlete. Asymptomatic, negative family history, normal echocardiogram. T wave inversions in leads V1-V4 represent an ethnic variant of "athlete's heart." T wave inversions in the lateral leads may represent the expression of a cardiomyopathy and deserve further evaluation and follow-up

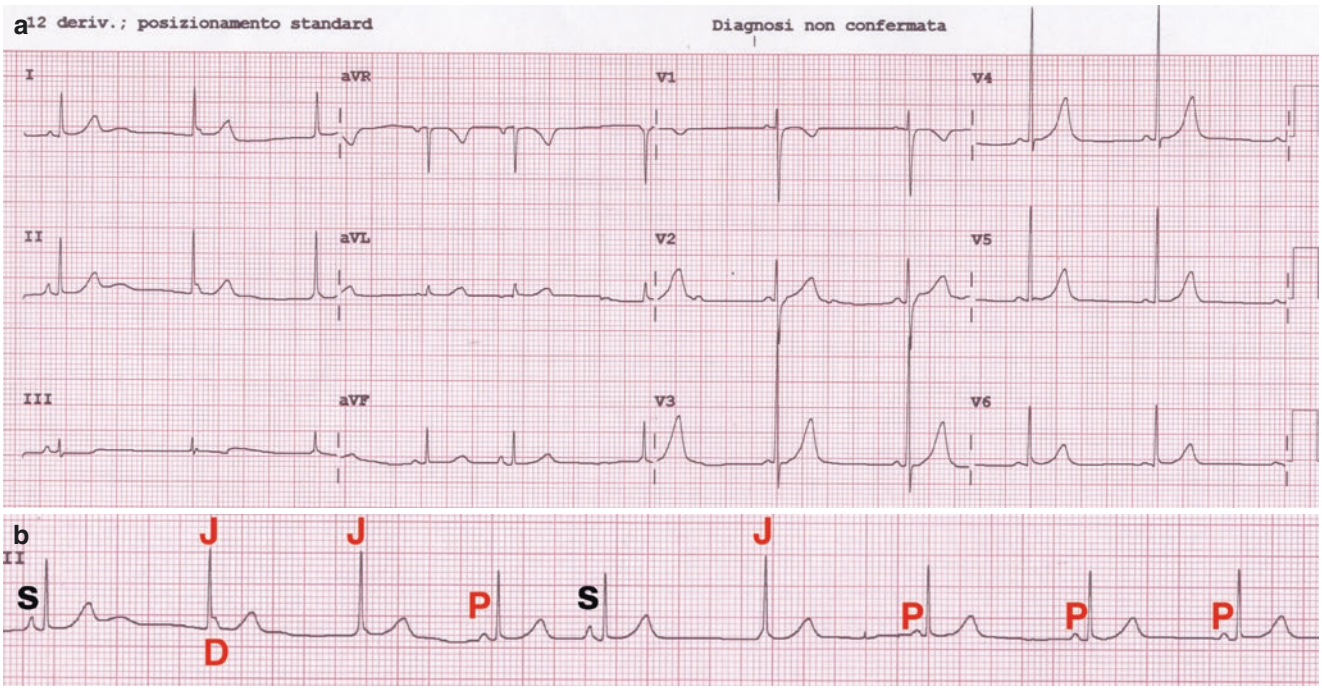

Fig. 6.48 (**a**) Nine-year-old boy. Physiological junctional rhythm. (**b**) Close-up: *S* sinus beat, *D* dissociated *P* wave, *J* junctional beat, *P* parasinusal beat

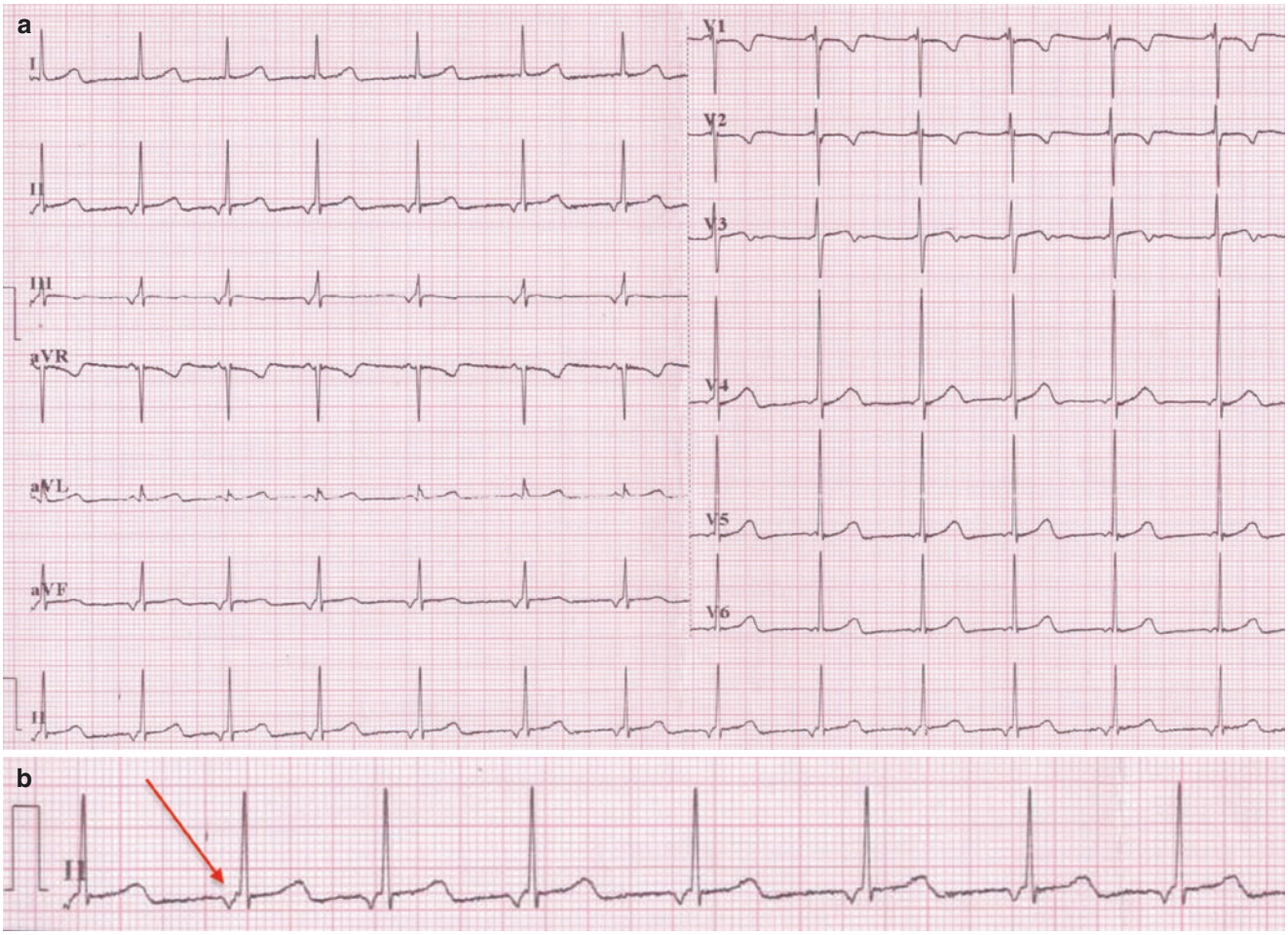

Fig. 6.49 (**a**) Six-year-old girl referred with the diagnosis of WPW. The PR interval is short, not because of pre-excitation but just because the rhythm arises in the low right atrium or coronary sinus, closer to the AV node, then shortening the PR interval. (**b**) Close-up

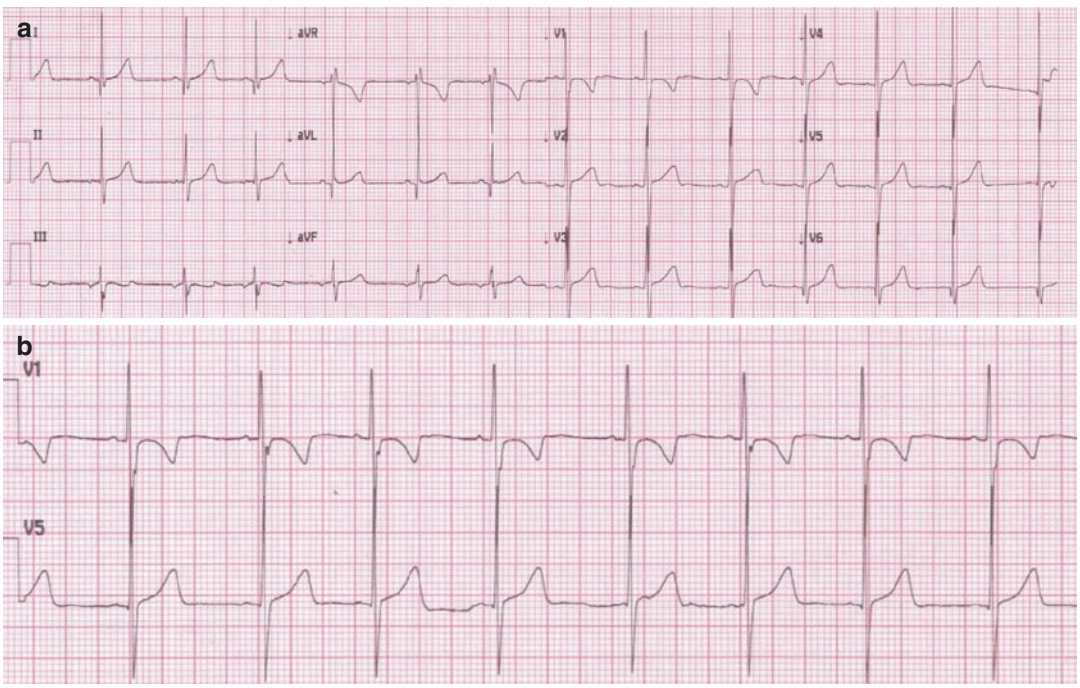

Fig. 6.50 (**a**) Nine-year-old boy. Another case similar to the previous one. (**b**) Close-up

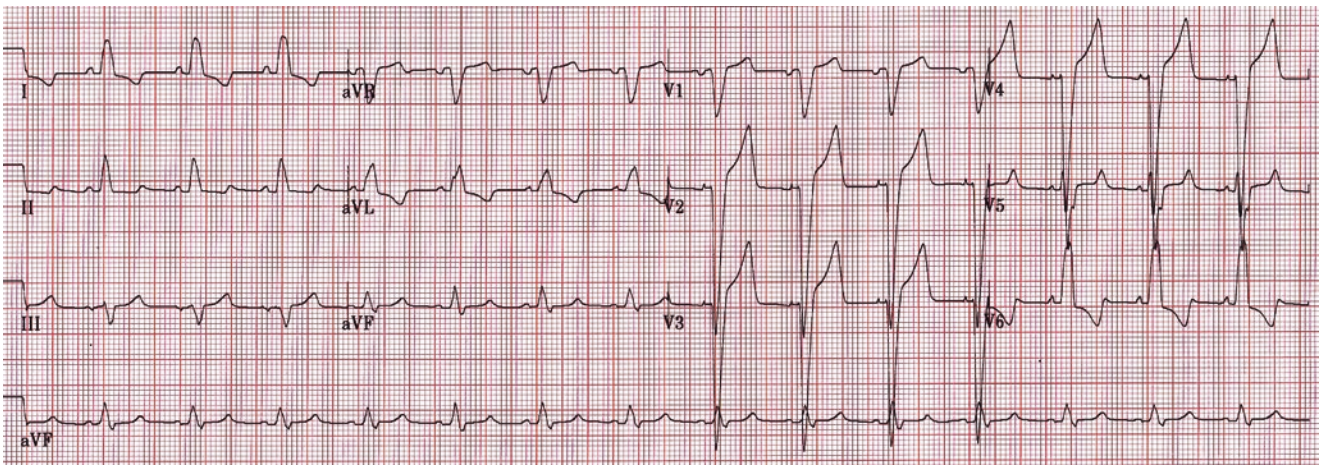

Fig. 6.51 Eight-year-old boy without heart disease. The ECG shows short PR interval and RBBB. An idiopathic RBBB is not so rare in the young. An LBBB as in this case is very rare. A follow-up is warranted and also a pre-excitation syndrome has to be ruled out

Drugs, Electrolyte Abnormalities, and Metabolic Factors

7

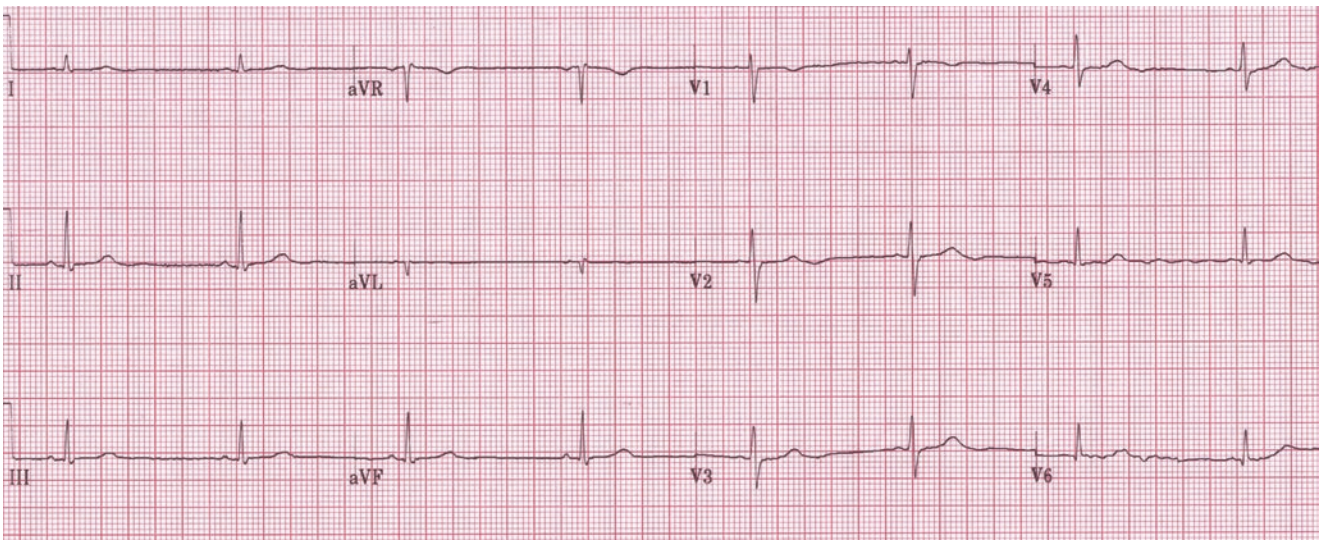

Fig. 7.1 Sixteen-year-old girl. Mental anorexia. Bradycardia and low voltages are typical. QT prolongation has been described, usually secondary to electrolyte imbalance (hypokalemia)

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G. Bronzetti, *Atlas of Pediatric and Youth ECG*, https://doi.org/10.1007/978-3-319-57102-7_7

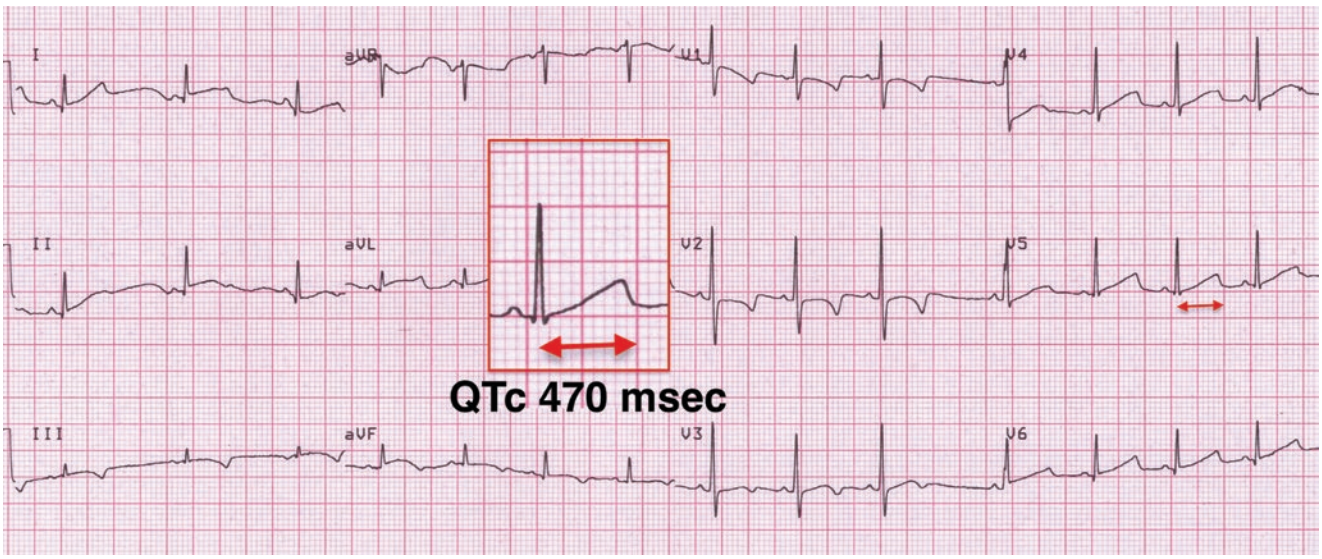

Fig. 7.2 Three years old. Diabetes, hypoglycemic spell. Note the mild QT prolongation

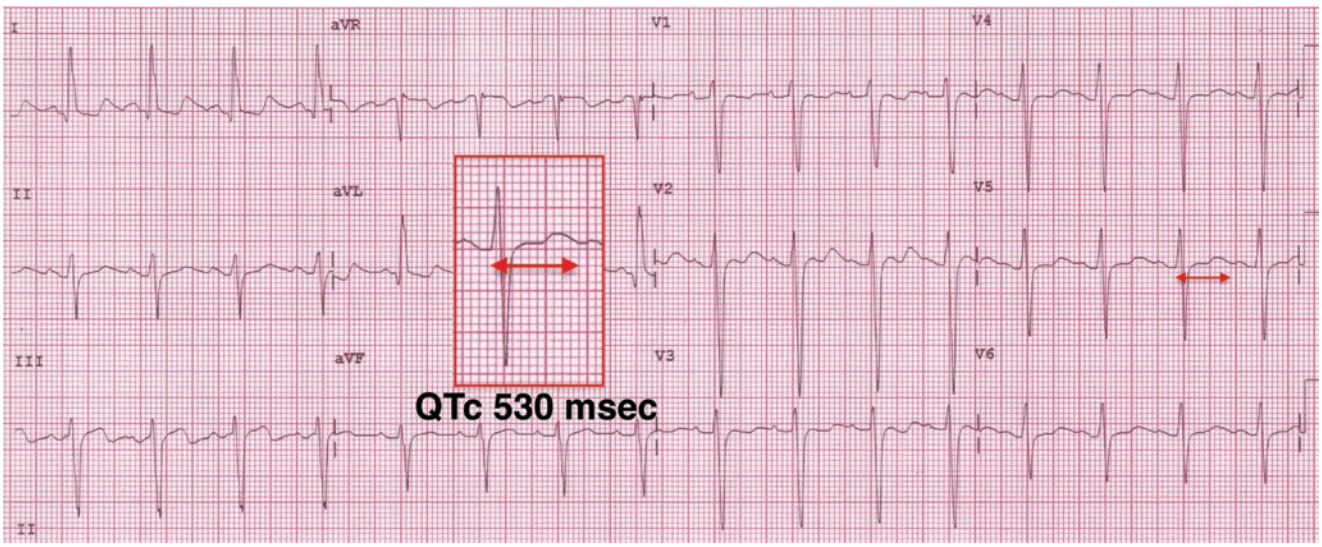

Fig. 7.3 Ten years old, diabetes onset. As in the previous case, there is a QT prolongation, severe in this case. A mild QRS enlargement can be noted

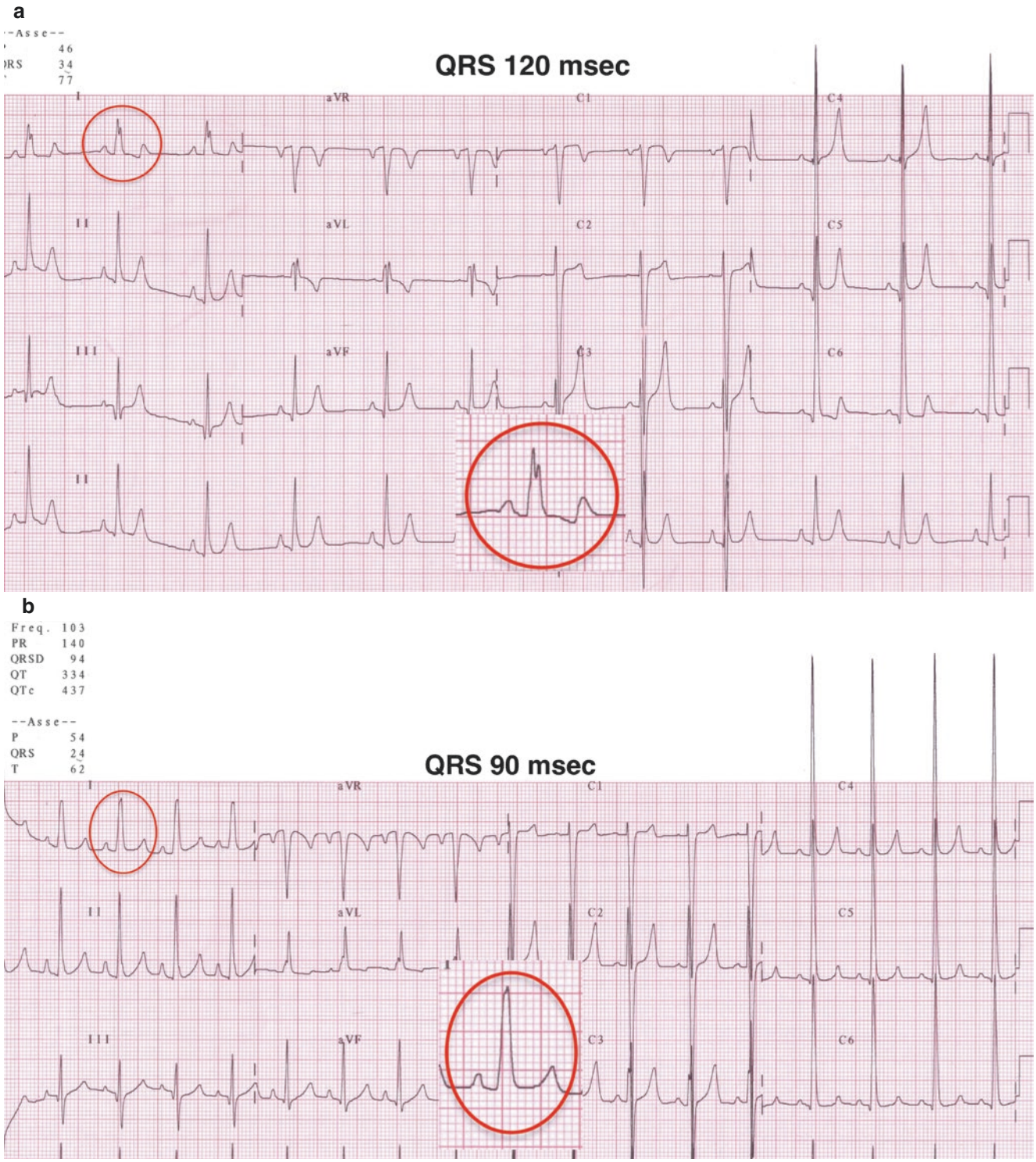

Fig. 7.4 Ten years old, carnitine deficit with cardiomyopathy (LVEF 35%). (**a**) Before treatment the QRS lasts 120 ms. (**b**) After carnitine supplementation, the QRS tends to normal

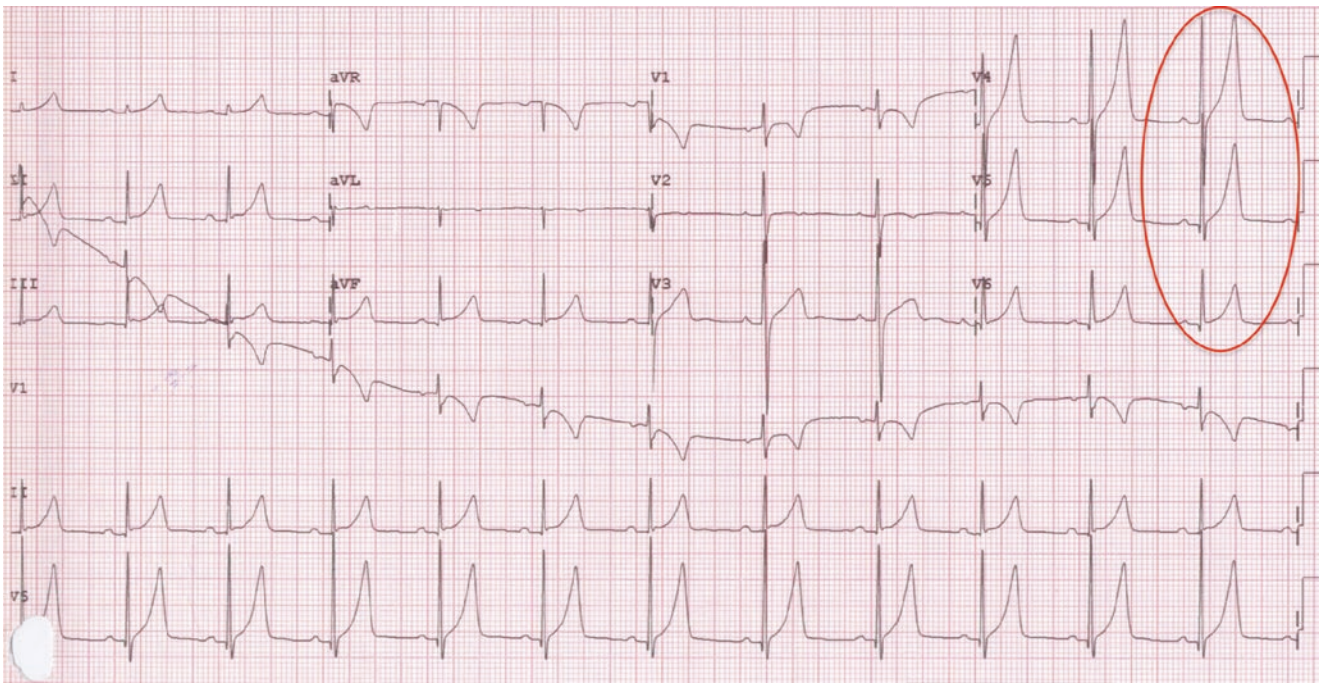

Fig. 7.5 Seven-year-old male. Renal insufficiency. Hyperkalemia (serum K 6.5 mmm). Note the tall and peaked T wave in the precordial leads

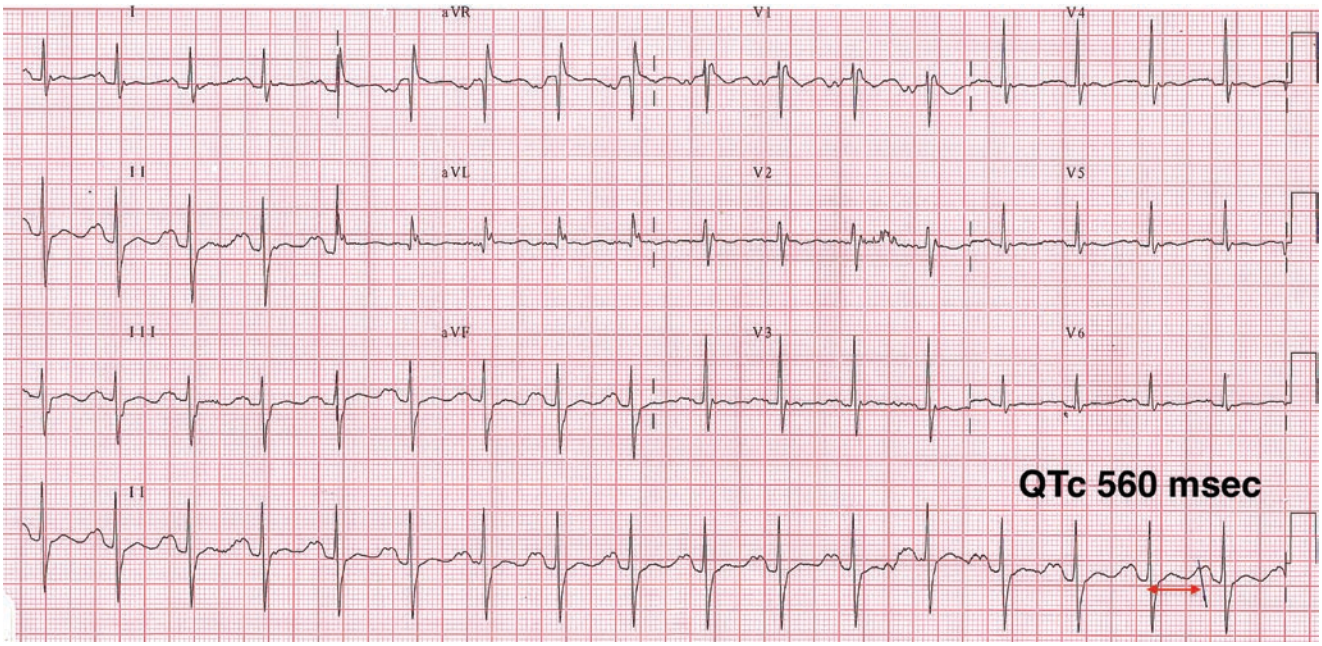

Fig. 7.6 Six-year-old girl with repaired VSD and aortic coarctation. Severe hypokalemia secondary to diuretic treatment. A severe QT prolongation can be noted

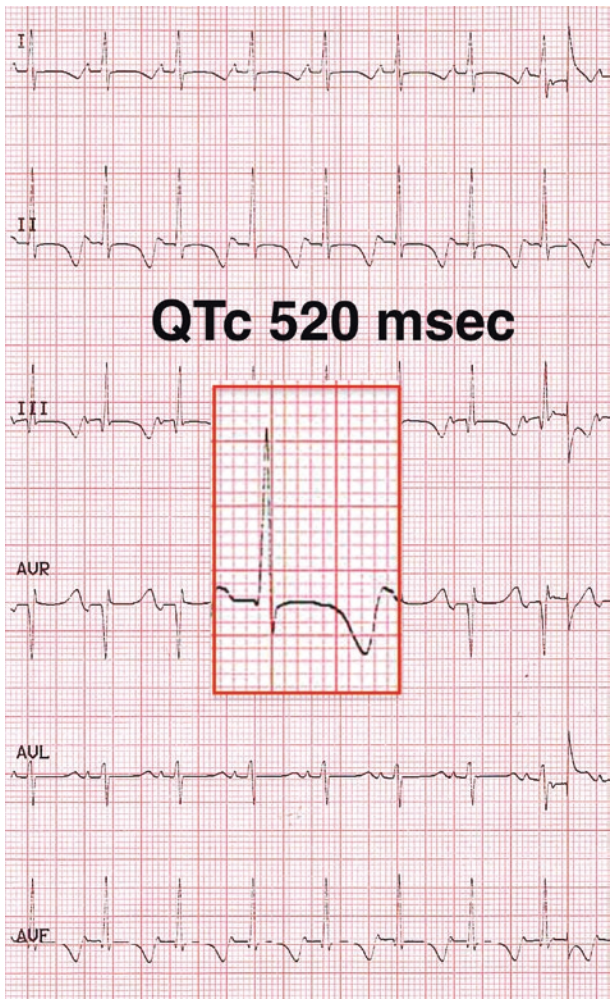

Fig. 7.7 Eight-year-old male. Hypocalcemia. Diffuse and extensive ST-T changes. A severe QT prolongation can be noted

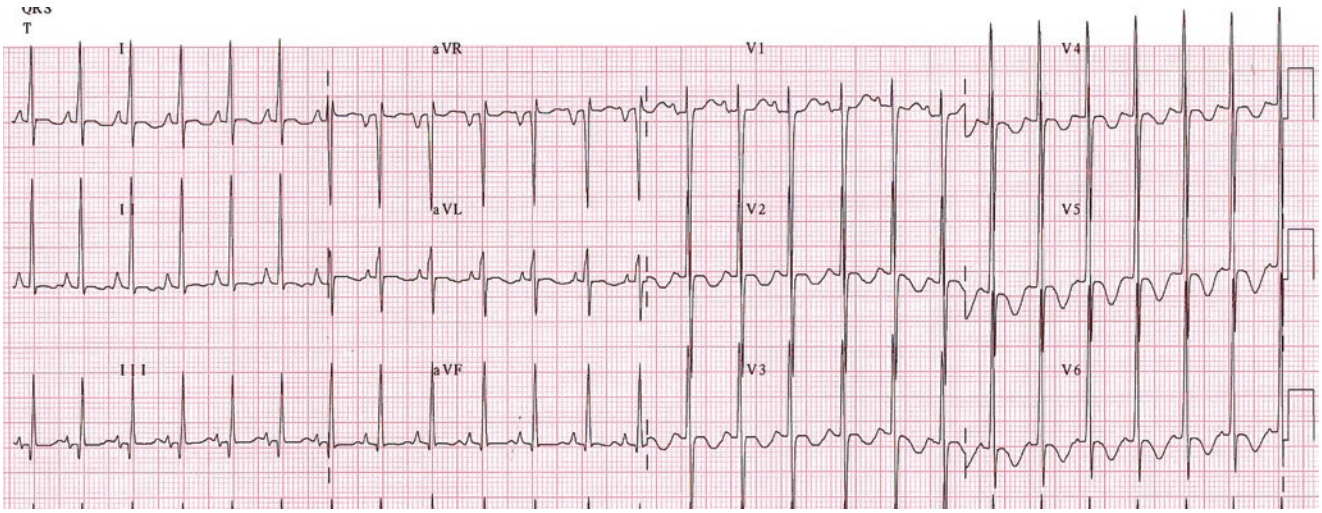

Fig. 7.8 Three-year-old female. Dilated cardiomyopathy and hypocalcemia secondary to diuretic treatment. As in the previous case, severe repolarization abnormalities can be noted (QTc 515 ms)

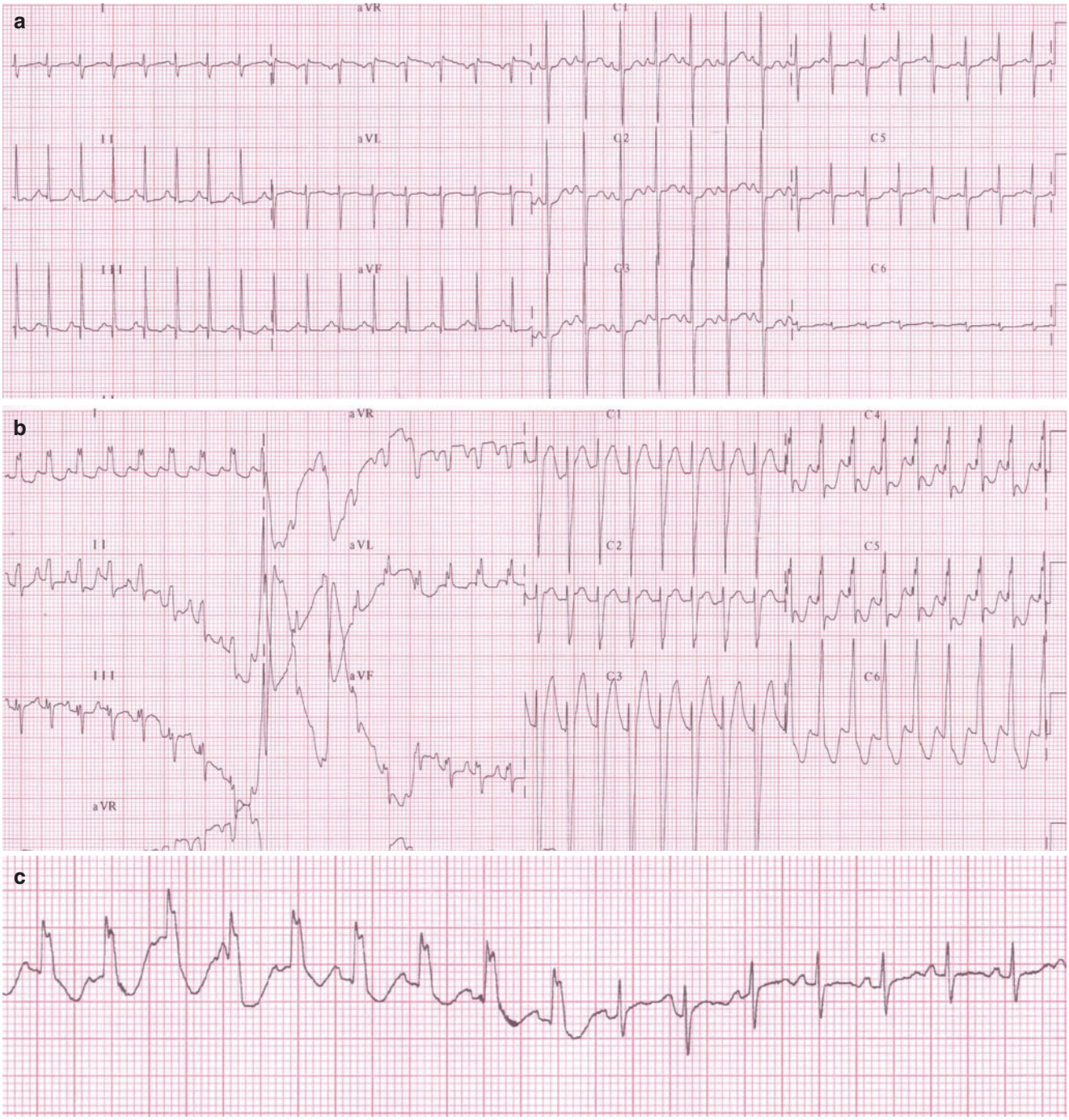

Fig. 7.9 Neonate with truncus arteriosus waiting for surgery. (**a**) Basal ECG. (**b**) Functional LBBB due to hypercalcemia (oversupplementation). (**c**) Close-up of the transition from LBBB to normal

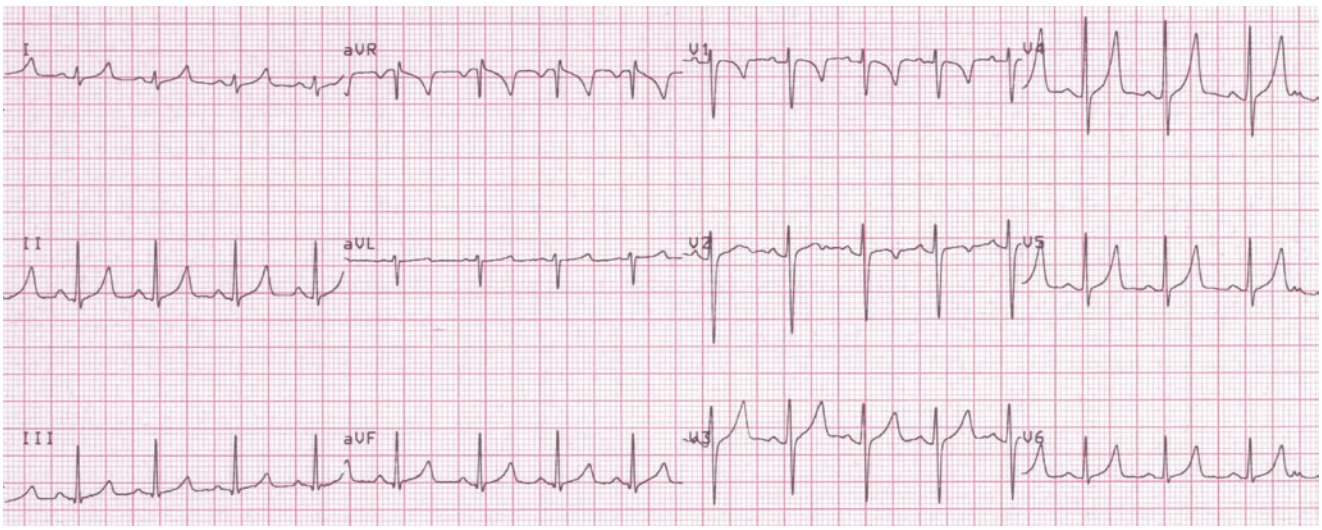

Fig. 7.10 Seven years old. Another case of hyperkalemia (oversupplementation). Note the tall and peaked T waves

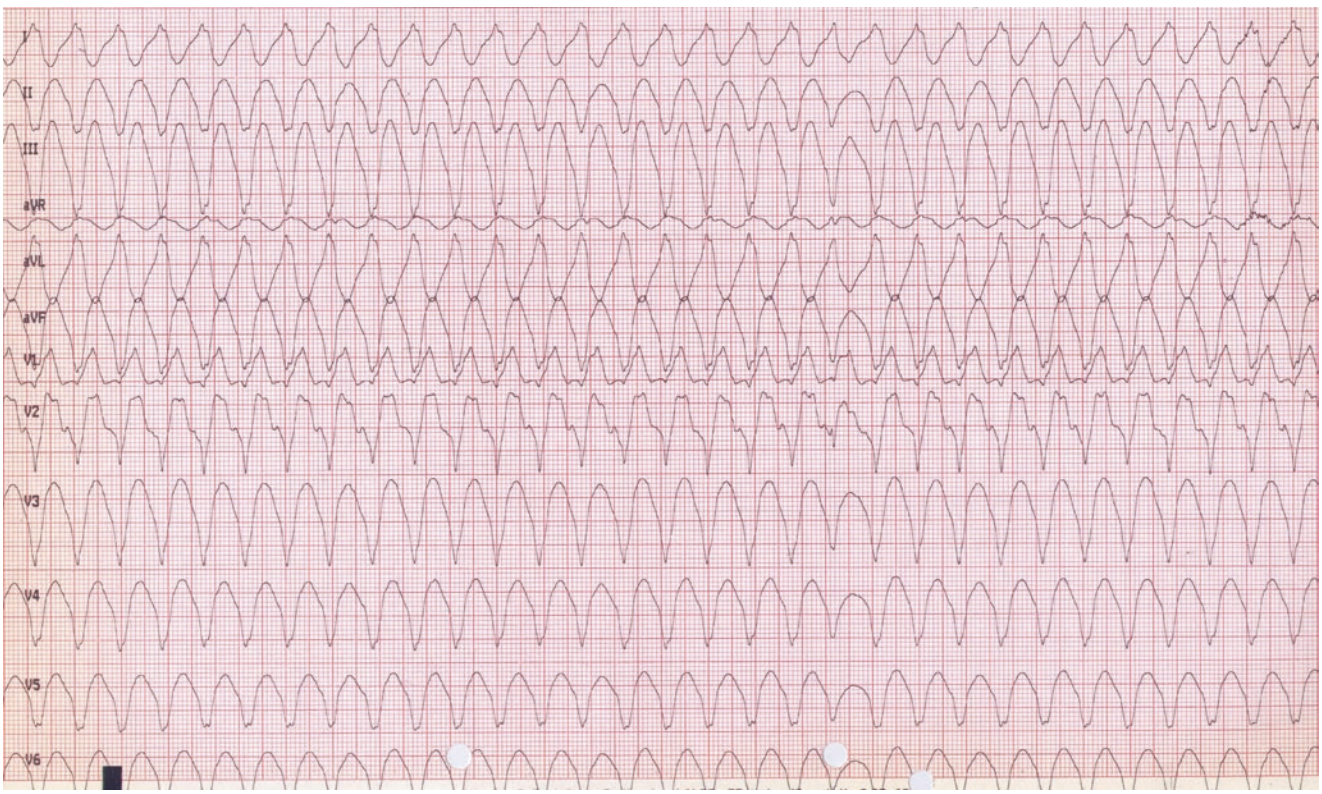

Fig. 7.11 Neonate admitted in ER in shock and severe acidosis. The QRS are extremely wide

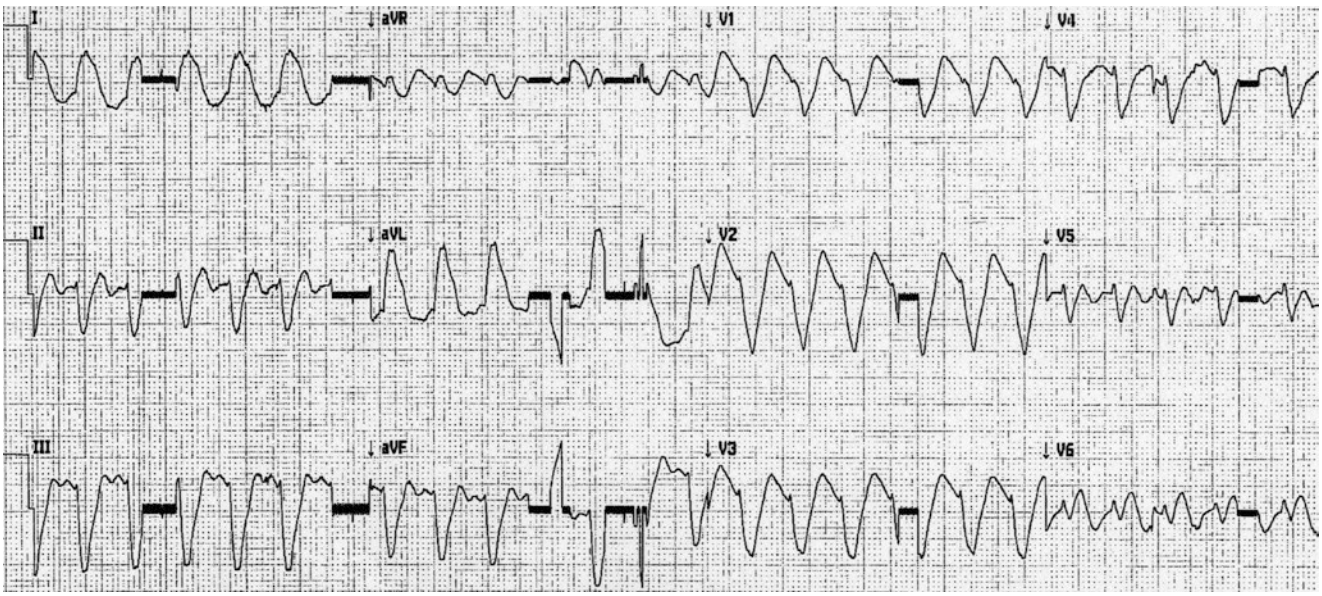

Fig. 7.12 As in the previous case, this is a neonate with severe metabolic disease and acidosis. Note the wide, agonic QRS typical of decompensated metabolic disease

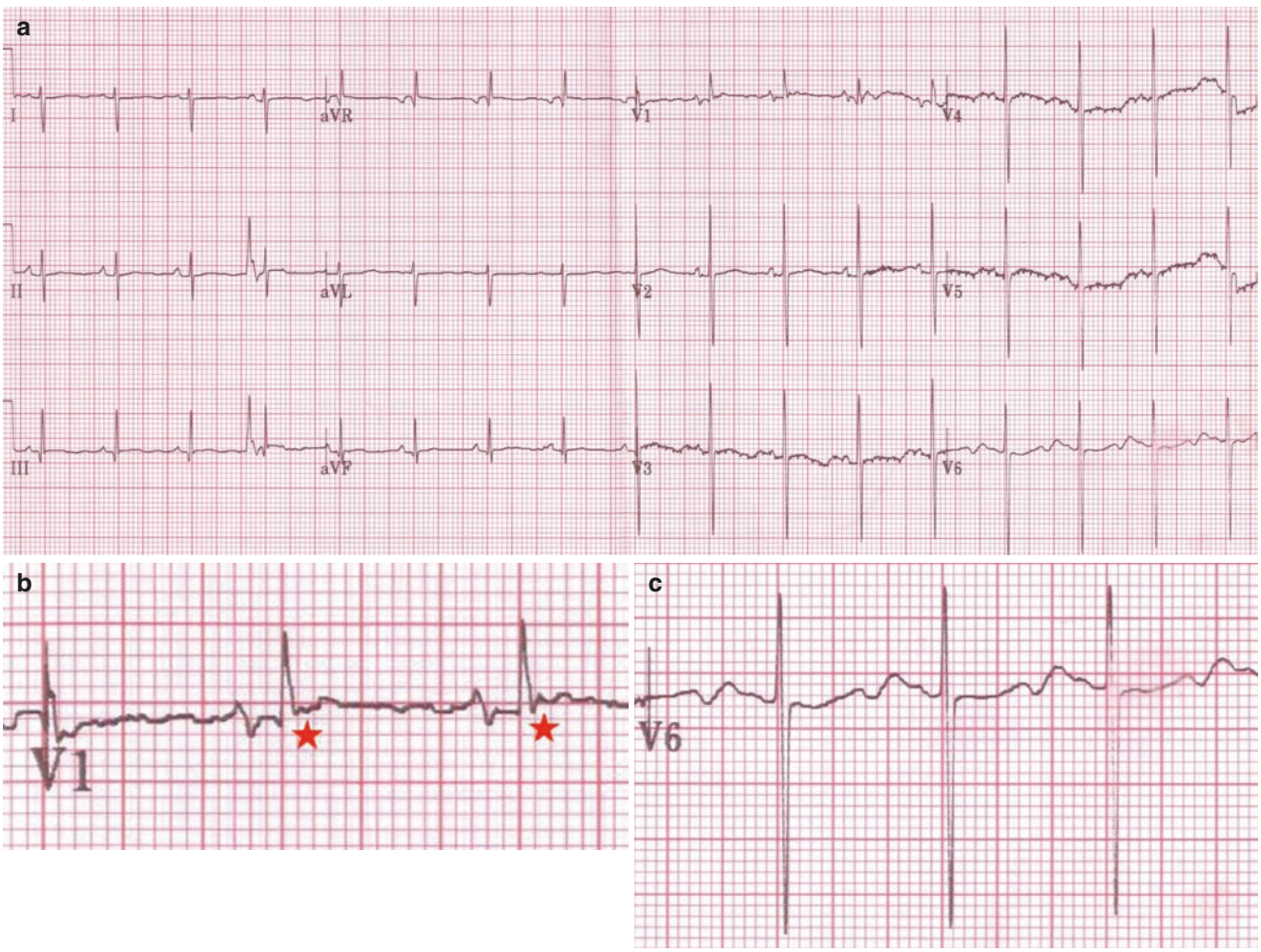

Fig. 7.13 (**a**) Newborn on hypothermia treatment for hypoxic brain suffering. Sinus rhythm, normal HR. (**b**) In V1 a notched J point can be noted. This is the equivalent of the Osborn wave ("camel" wave) described in hypothermia as well as in severe hypercalcemia or significant brain damage. (c) There is also QT prolongation, another consequence of hypothermia

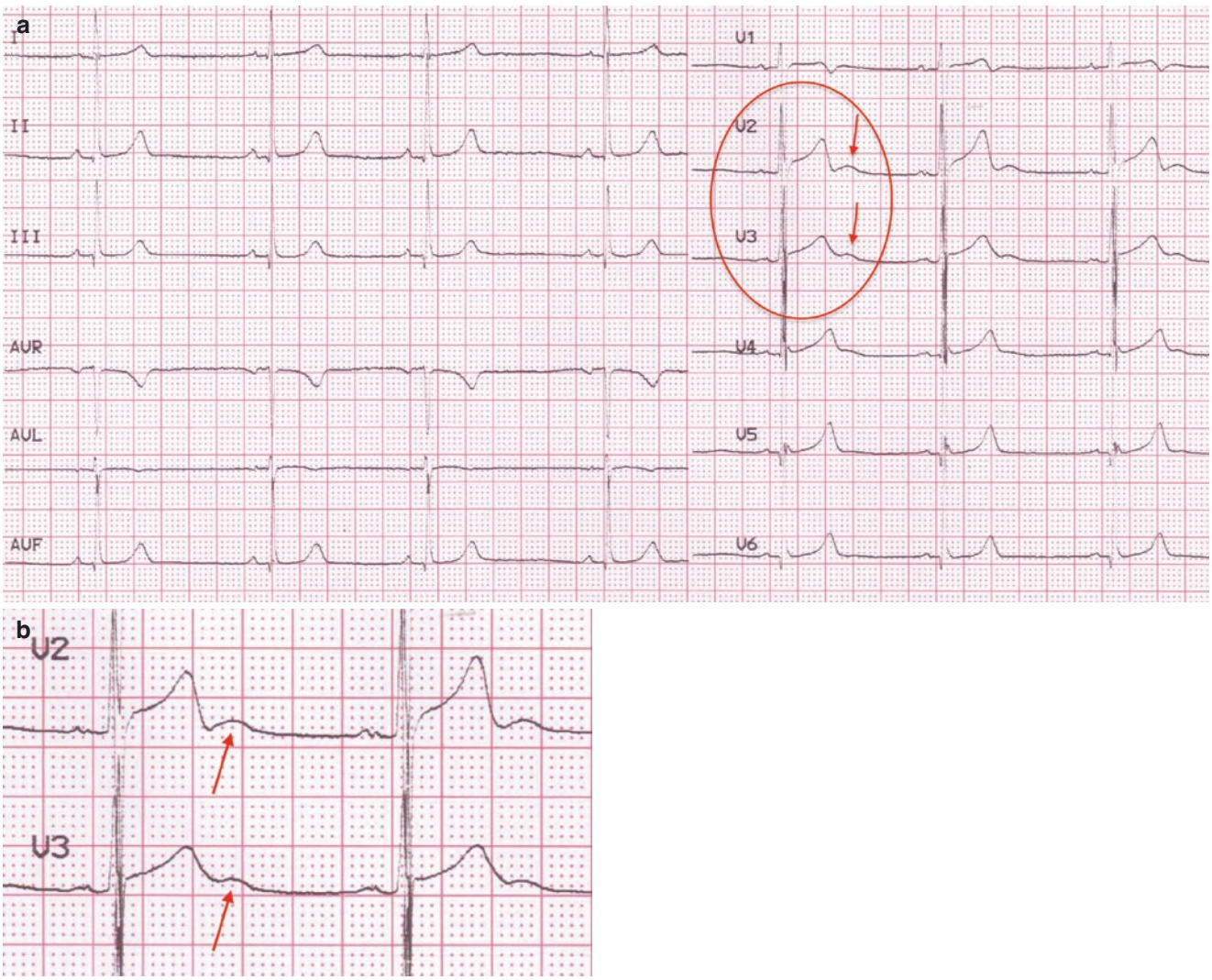

Fig. 7.14 (**a**) Twelve-year-old male. Dysentery and hypokalemia. Note the U wave. (**b**) Close-up

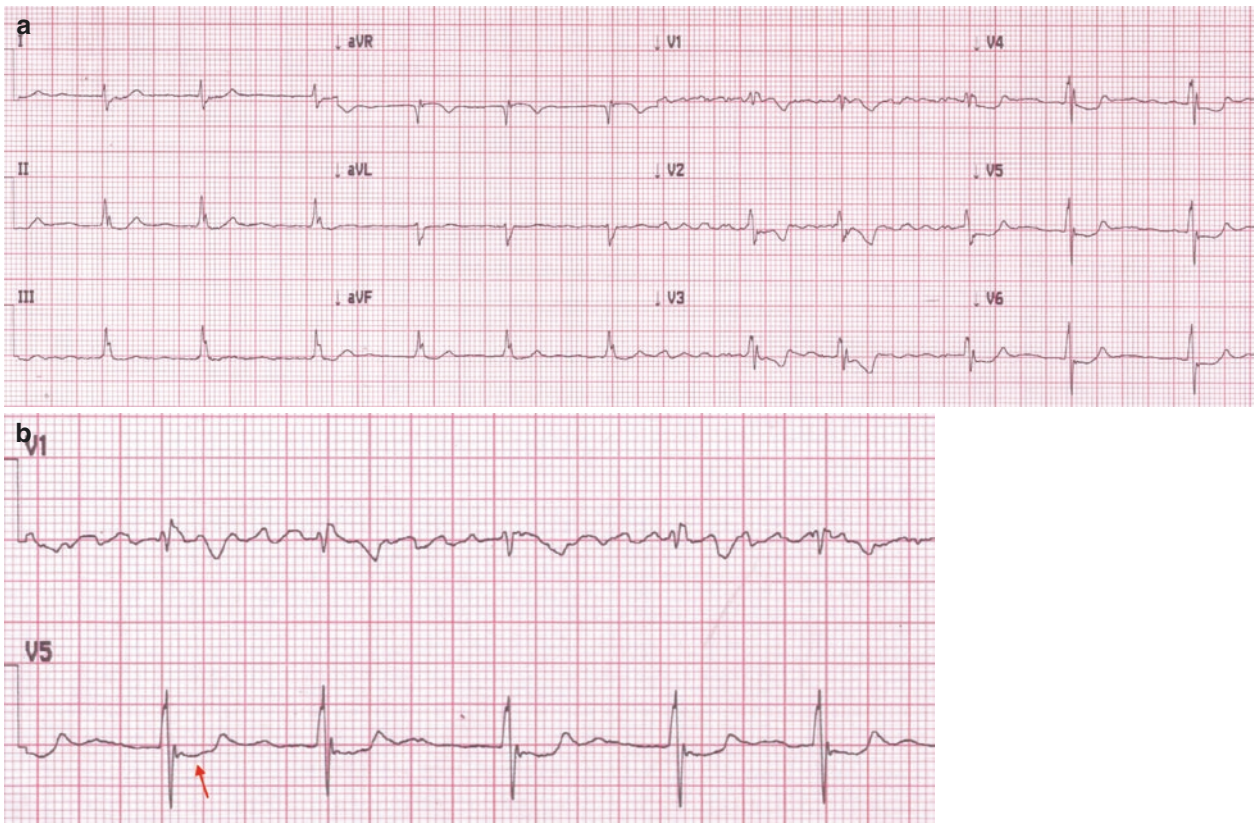

Fig. 7.15 (**a**) Twenty-year-old female. Large ASD which underwent late surgery. The massive dilatation of right-sided chamber is the cause of chronic atrial fibrillation. The patient is on digoxin to slow the ventricular rate. (**b**) Close-up. The downsloping ST depression with a characteristic "sagging" can be noted, the so-called digoxin effect. The sagging resembles a cycloid brachistochrone

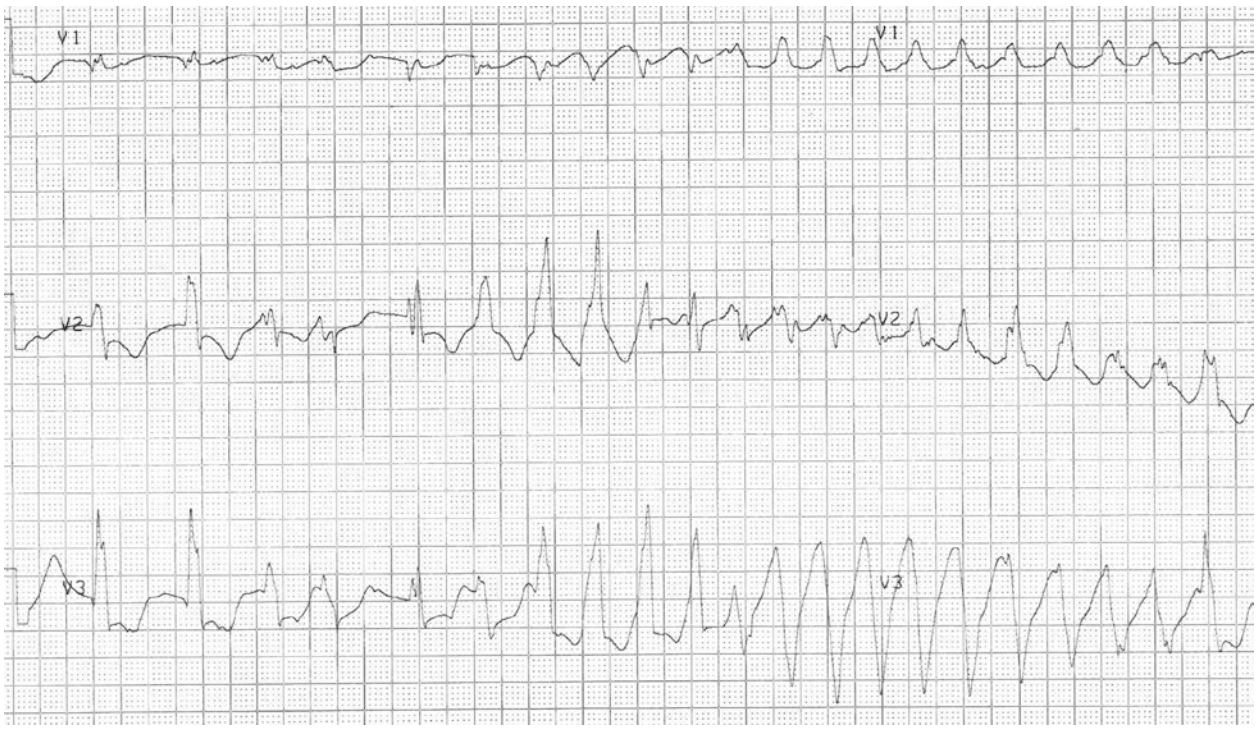

Fig. 7.16 Twenty-year-old female, tetralogy of Fallot, in the immediate postoperative course for right outflow homograft implant. On amiodarone for treating AF, she developed QT prolongation and a run of torsade de pointes. Amiodarone was stopped and VVI pacing was started. Amiodarone, like other class III drugs in the Vaughan-Williams classification (sotalol), affects the repolarization, predominantly blocking the potassium channels. In case of class III drugs overdosage or intoxication, the antidote is potassium chloride

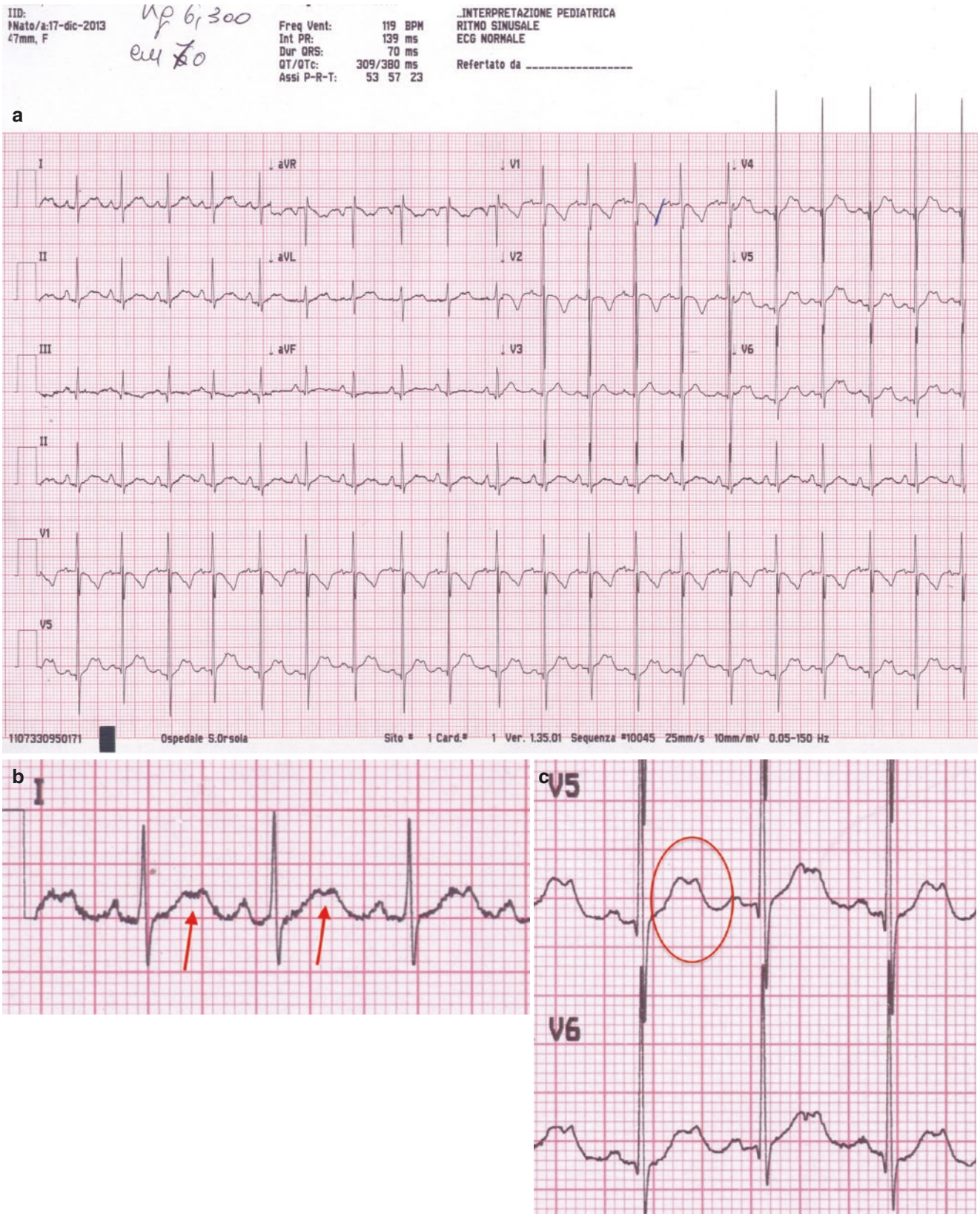

Fig. 7.17 (**a**) Eight-month-old male on amiodarone for JET. A notched, bicuspid T wave can be noted, despite a not prolonged QT interval. (**b, c**) Close-up

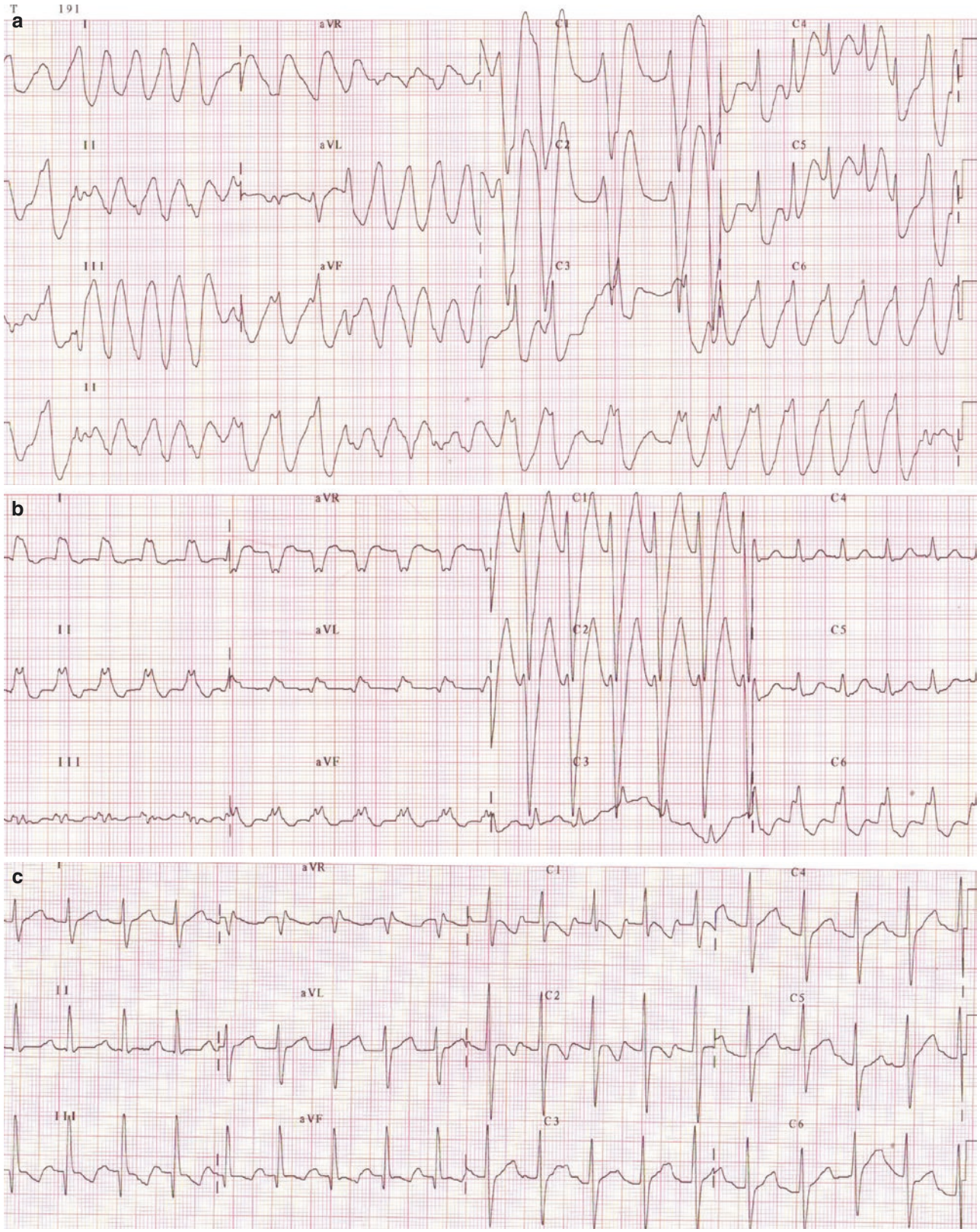

Fig. 7.18 Two-year-old female on flecainide for PSVT prophylaxis. (**a**) She was admitted to the ER with severe acidosis, soporous, and with impressively wide complex ventricular tachycardia on ECG. A flecainide intoxication caused by erroneous galenic preparation was suspected and confirmed by serum flecainide check, revealing a concentration twice the upper limit. (**b**) Sodium bicarbonate infusion was started with quick rhythm conversion and narrowing of the QRS until complete normalization (**c**). We mentioned the usefulness of the potassium in case of class III poisoning by flecainide; sodium bicarbonate is an excellent antidote to flecainide (do not forget that flecainide is a sodium channel blocker)

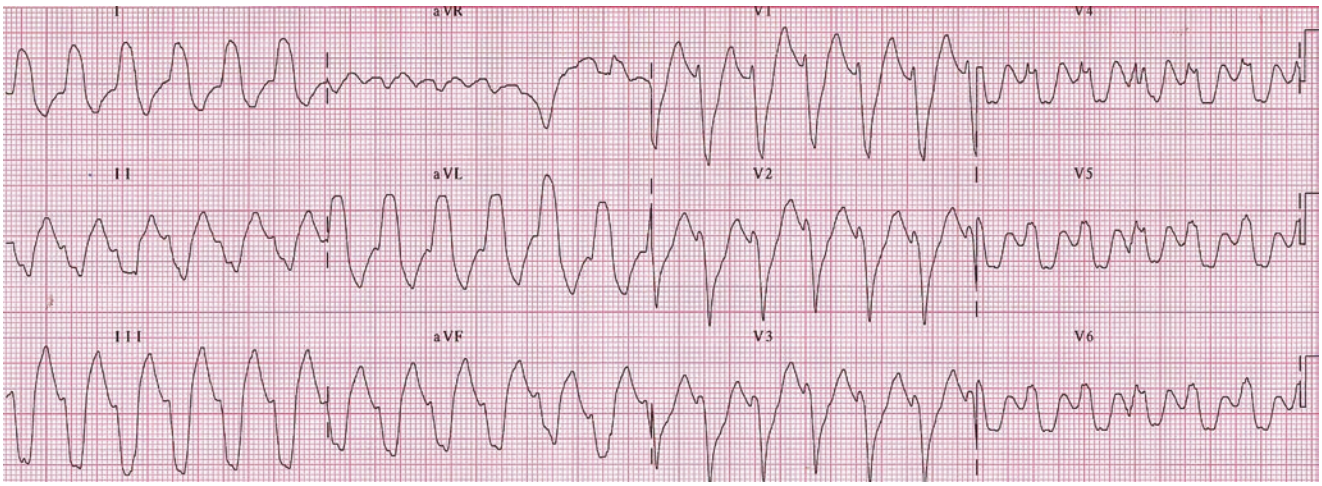

Fig. 7.19 Three-year-old male on flecainide for PSVT prophylaxis. Accidental poisoning occurred (the baby ingested a handful of pills). As in the previous figure, you can see the very wide ventricular complex

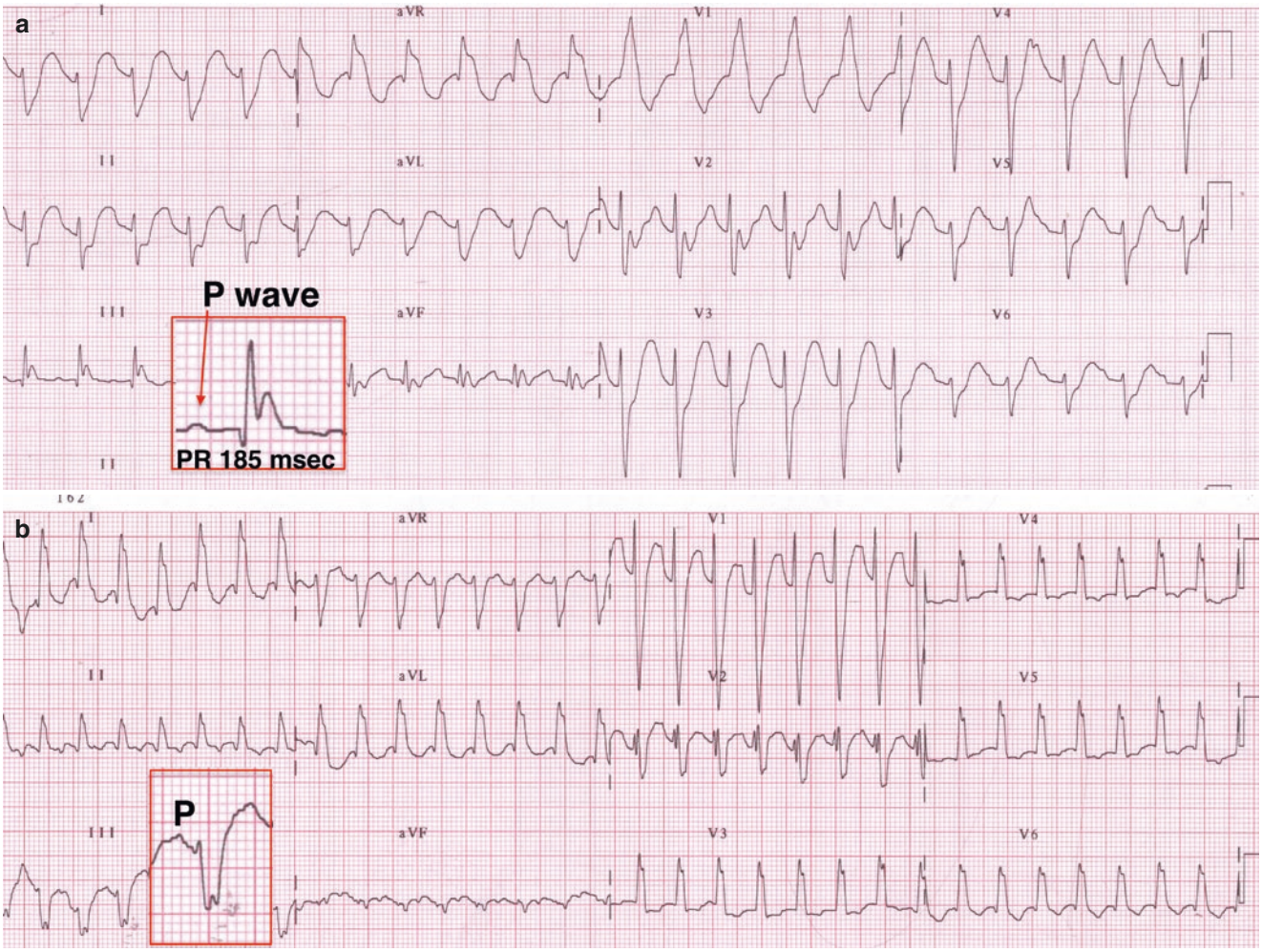

Fig. 7.20 Three-month-old boy on propafenone. (**a**) Sinus rhythm with RBBB. First-degree AV block. (**b**) Sinus rhythm with LBBB. Like other class IC antiarrhythmic drugs, propafenone acts depressing the phase 0 of the action potential without affecting repolarization, slowing the conduction in fast-response tissues. AV conduction blocks and widening of the QRS are common effects and could be a warning to adjust the posology

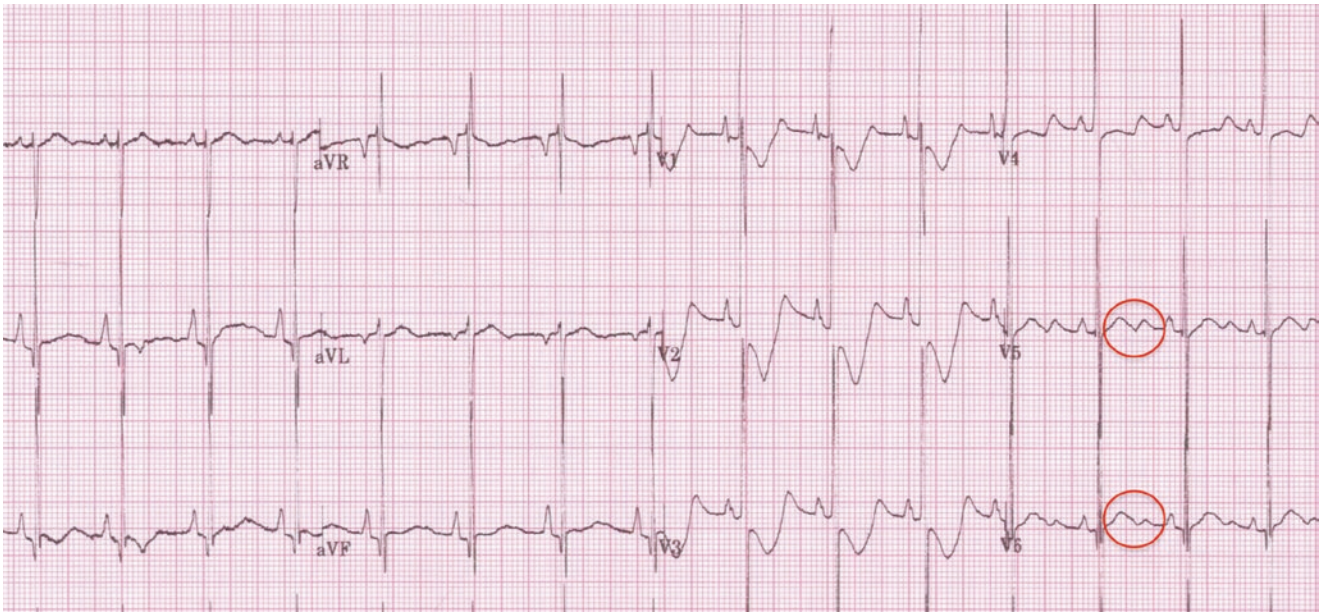

Fig. 7.21 Newborn on hypothermic treatment for brain injury. Long QT and U wave can be noted, along with marked change of ST segment especially in the right precordial leads

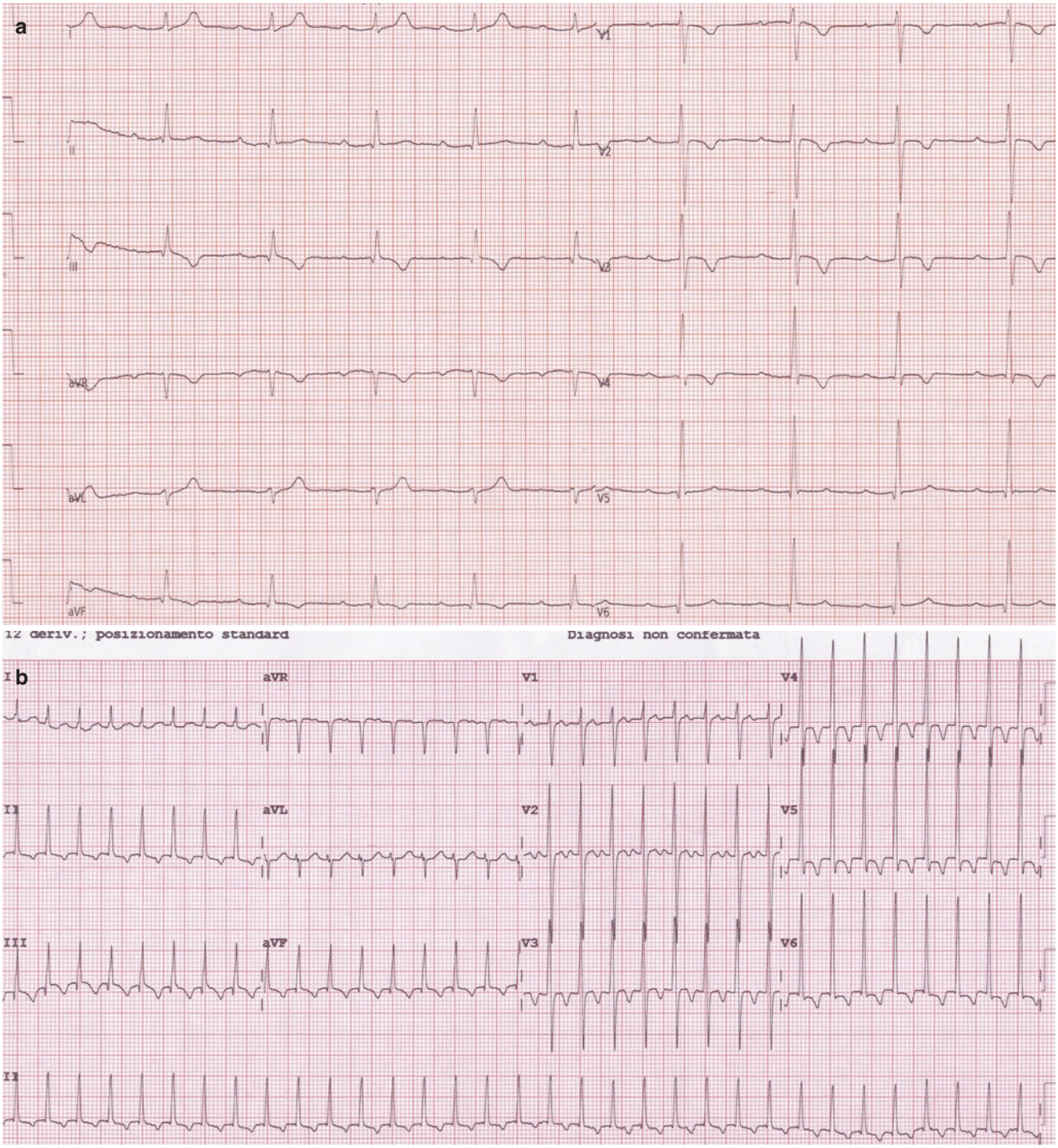

Fig. 7.22 (**a**) 11 year old male with transfusion-dependent thalassaemia. A major complication is heart failure due to accumulation of iron within heart muscle cells. Arrhythmias are common, from AV block to tachycardias. Here you can see a first degree AV block. (**b**) The same patient during an episode of paroxysmal supraventricular tachycardia, EAT

Suggested Reading

Current Annual Report. National poison data system. The American Association of Poison Control. 2014. http://www.aapcc.org/. Accessed 20 Aug 2014.

Hoffman RS. Fluid, electrolyte, and acid-base principles. In: Goldfrank L, et al., editors. Goldfrank's toxicologic emergencies. 6th ed. Stamford: Appleton and Lange; 1998. p. 243.

Oreto G, Luzza F, Donato A, Carbone V, Satullo G, Calabrò MP. L'elettrocardiogramma—un mosaico a 12 tessere. Centro Scientifico Editore: Torino; 2009.

Artifacts and Devices

8

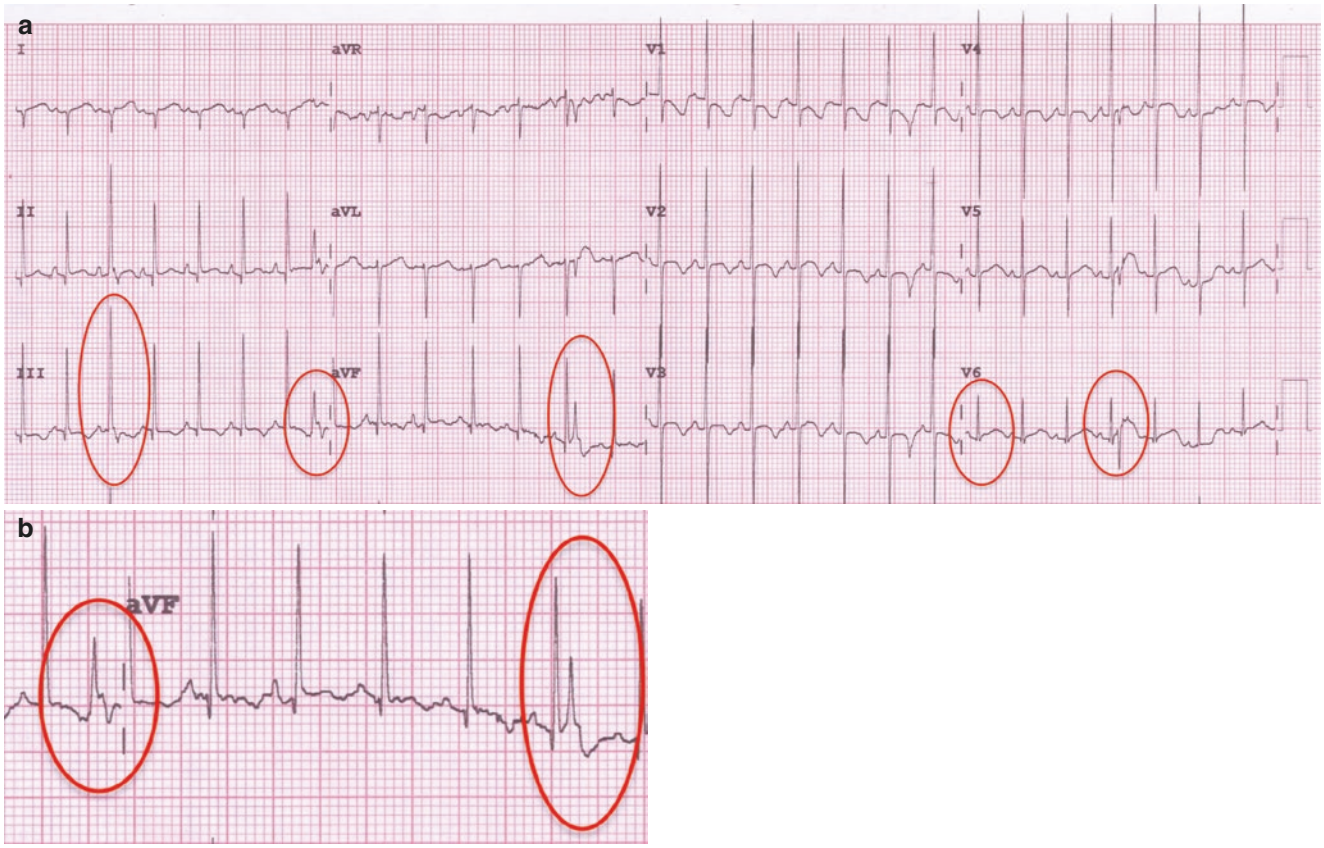

Fig. 8.1 (**a**) Two-month-old baby boy with hiccups. The first abnormal QRS complex simulates a PVC; the others are more clearly artifacts due to hiccups. So they are neither supraventricular nor ventricular beats. They are simply subventricular. Phrenic, if you prefer. (**b**) Close-up

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G. Bronzetti, *Atlas of Pediatric and Youth ECG*, https://doi.org/10.1007/978-3-319-57102-7_8

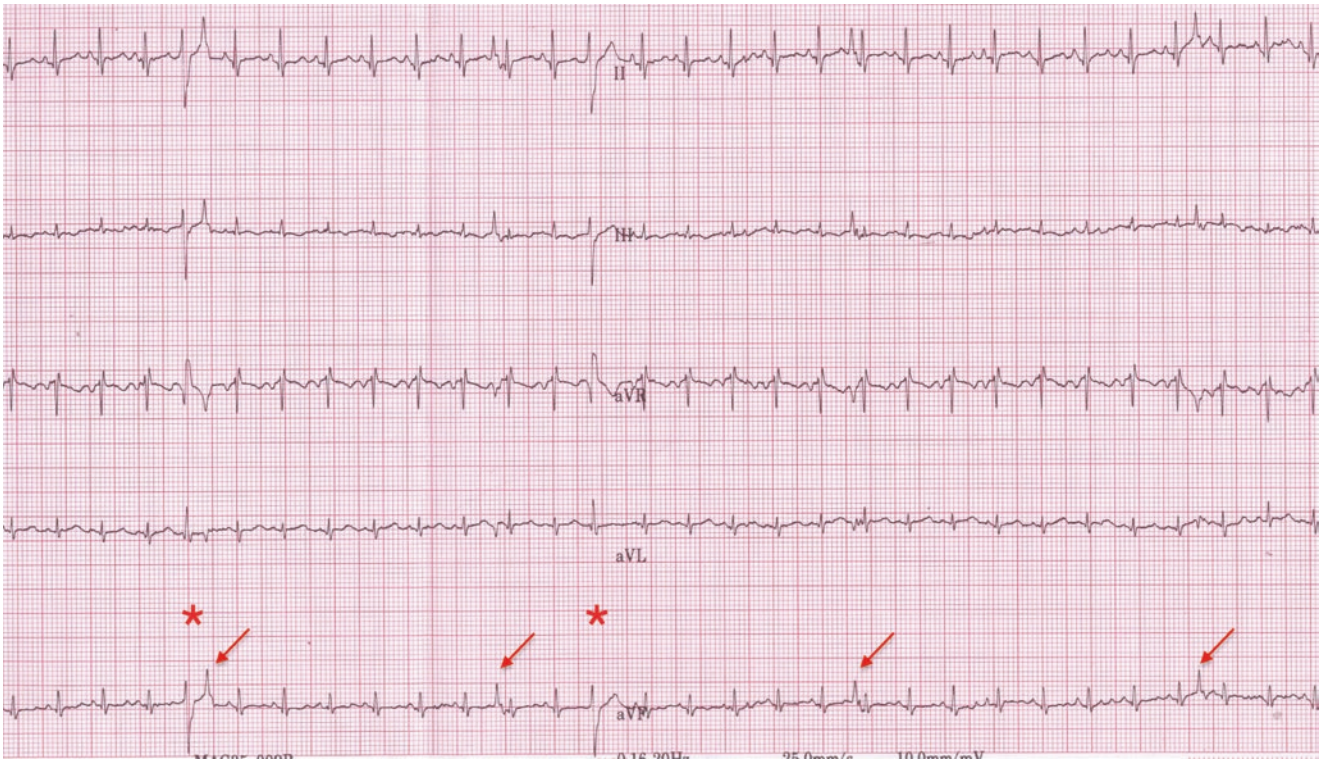


Fig. 8.2 As in the previous figure, the ECG shows an infant with hiccups. Unlike the previous case, there are also real PVCs (*asterisk*). The "subventricular" beats are quite rhythmic (*arrows*)

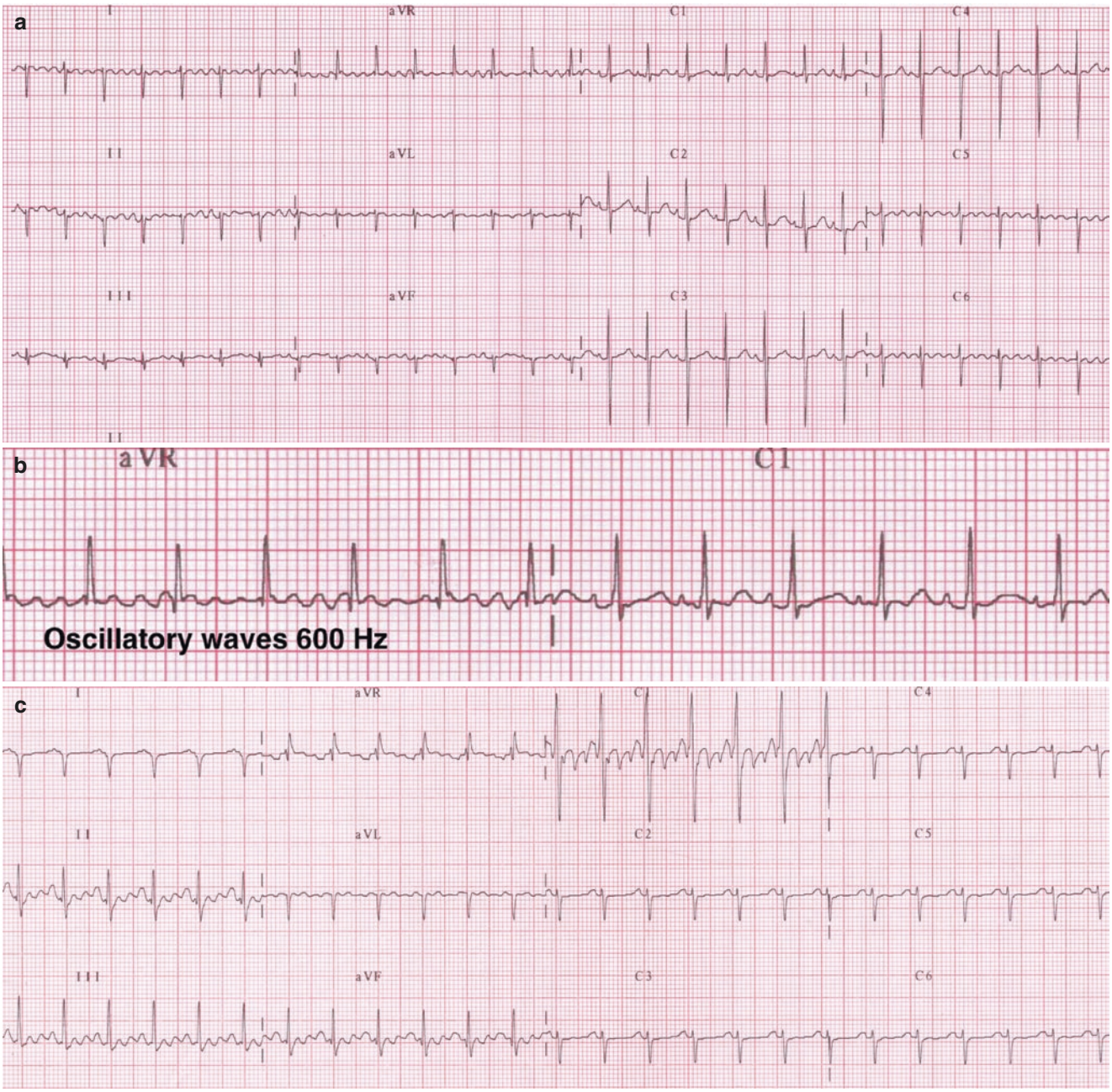


Fig. 8.3 (**a**) Infant on high-frequency oscillatory ventilation (HFOV) for diaphragmatic hernia. Some leads are consistent with atrial flutter with 3:1 or 2:1 A:V ratio. Diagnosis (and therapy) is easy: turn off the HFOV just a moment. (**b**) Close-up. (**c**) Another similar case

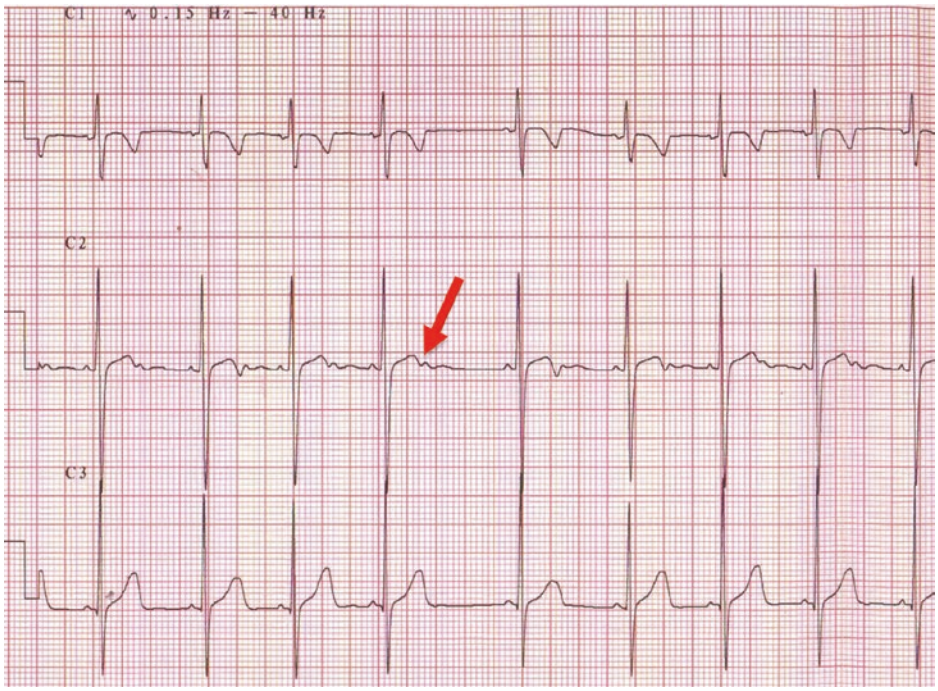


Fig. 8.4 Nine-year-old boy. The notched T wave in V3 may simulate a blocked P wave. The irregular rhythm due to the sinus respiratory arrhythmias can corroborate this deception

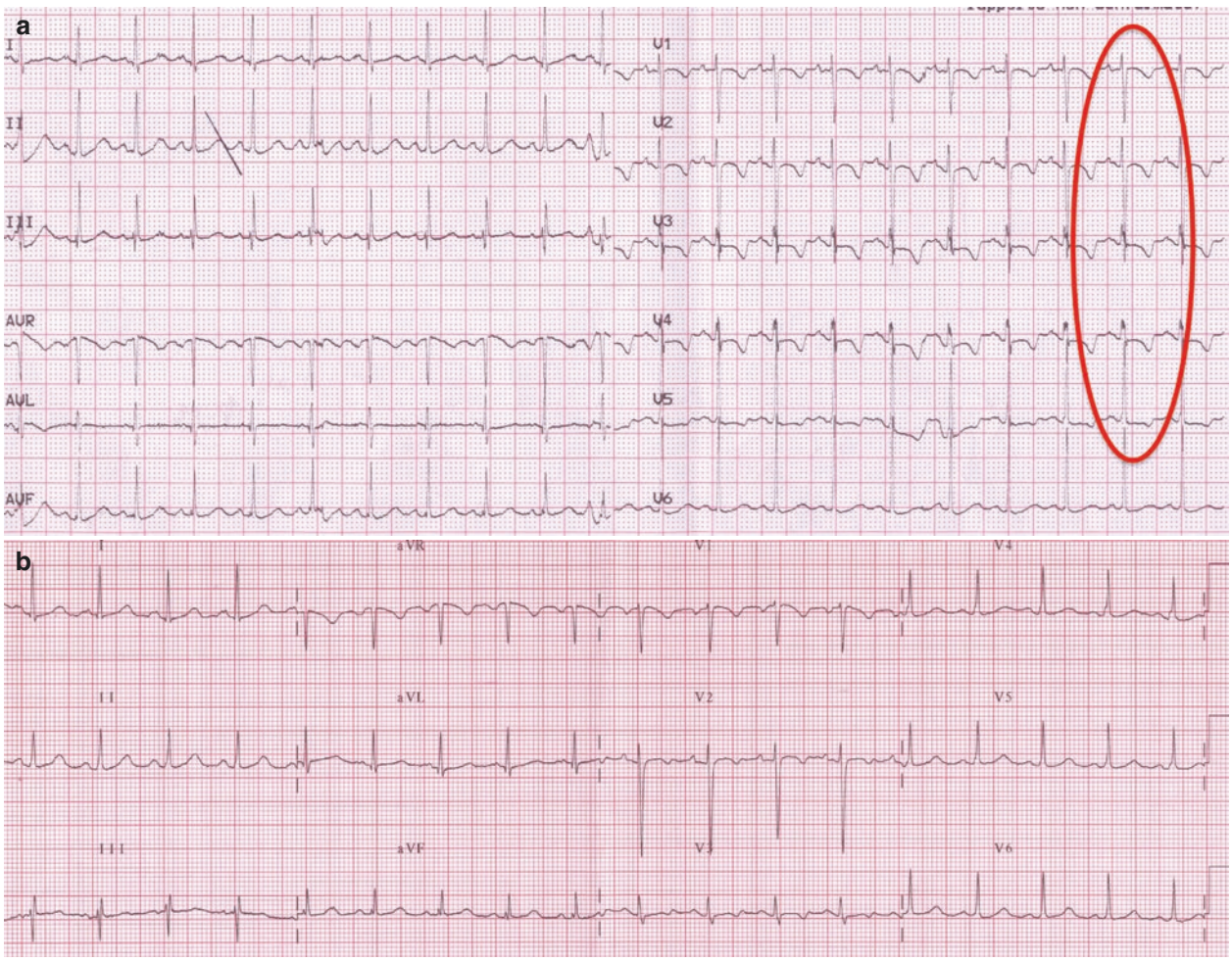


Fig. 8.5 Five-year-old girl admitted with breathing difficulties. (**a**) The ECG shows pathological ST-T changes in the precordial leads. On questioning the nurse, it emerged that the ECG was taken with the girl almost orthostatic in the arms of the mother. (**b**) The ECG was then repeated in a standard manner and was normal. As a rule, faced with a puzzling or pathologic ECG, it is always wise to enquire about the environment and the situation in which the ECG was recorded

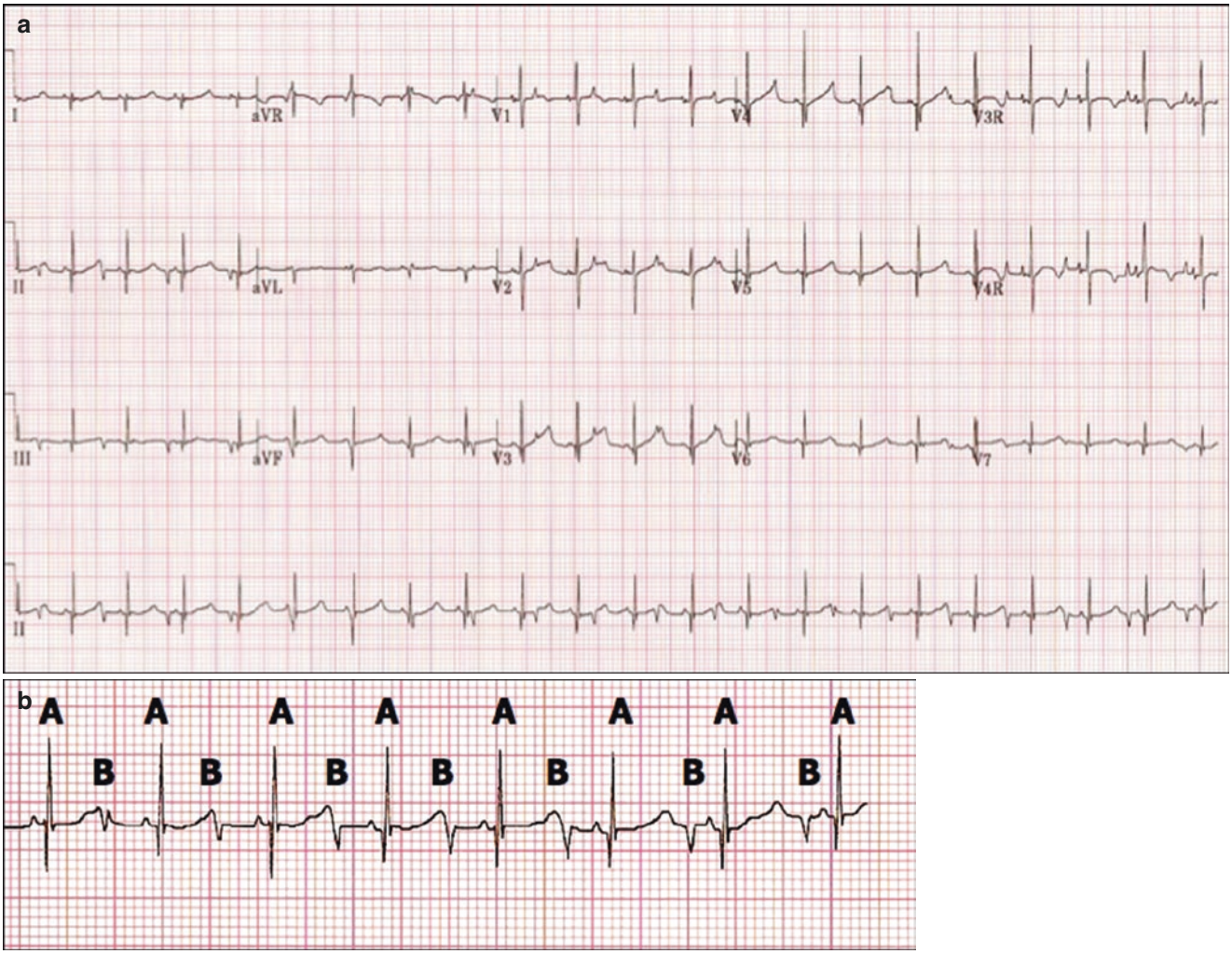


Fig. 8.6 (**a**) One month old. At a glance, it seems an artifact from hiccups. (**b**) A and B are two different QRS complexes in the "same body." It is a case of conjoined twins, joined at the abdomen

Fig. 8.7 (**a**) Eleven-year-old boy. A case of heart transplant. (**b**) The tiny waves (*arrows*) are the P waves of the remnant of the "old" sick heart

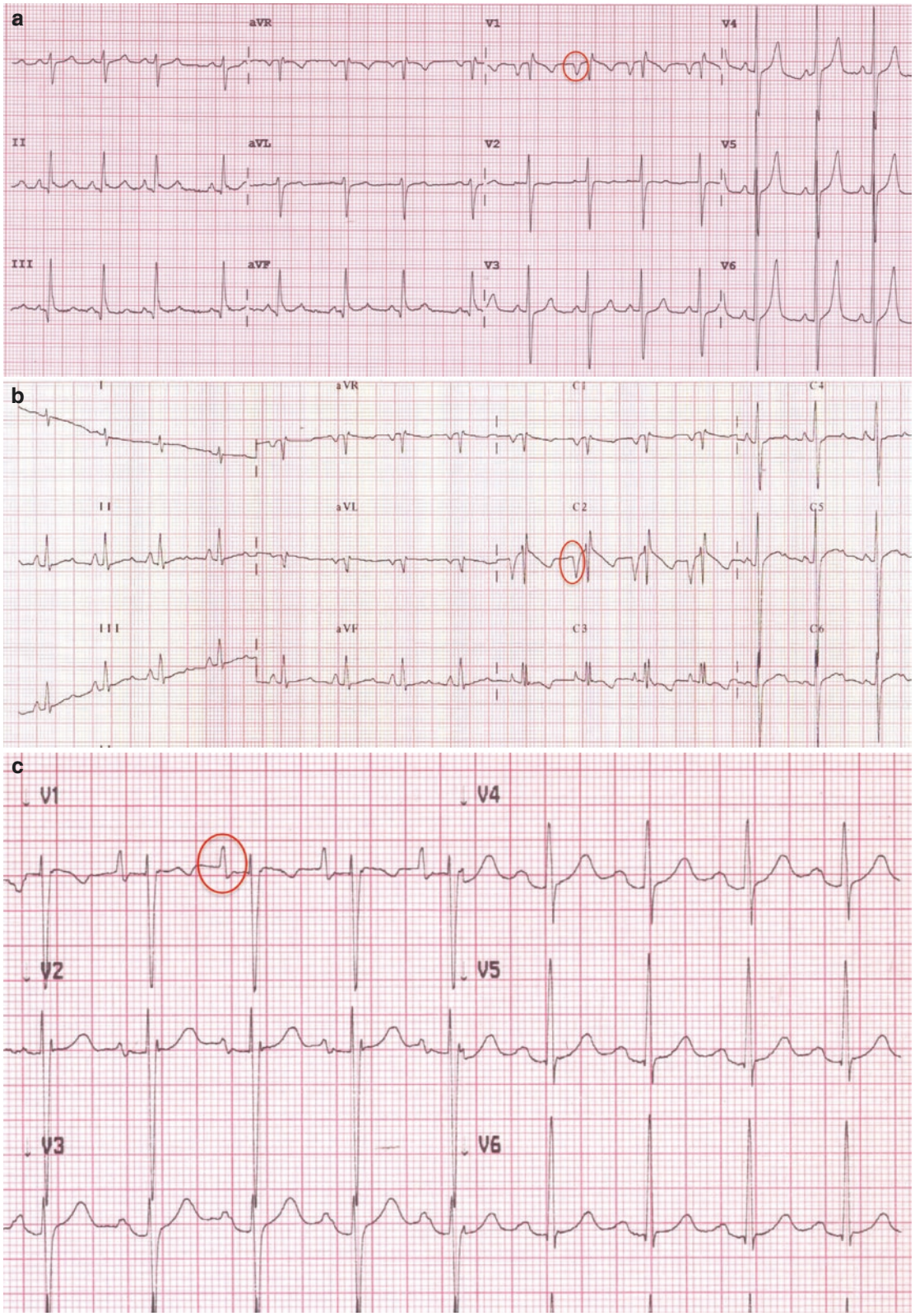


Fig. 8.8 (**a–c**). Three cases of adolescents with pectus excavatum (PEX). Many different features of this condition have been described, from RBBB to unusual rotation in the horizontal plane. In these cases, pseudo-pathological P waves can be noted. In (**b**) the P wave is very bizarre and meets the criteria for significant left atrial enlargement. Once again, it is wise, when faced with "odd" features, to ask about chest deformities or other conditions know to alter the ECG

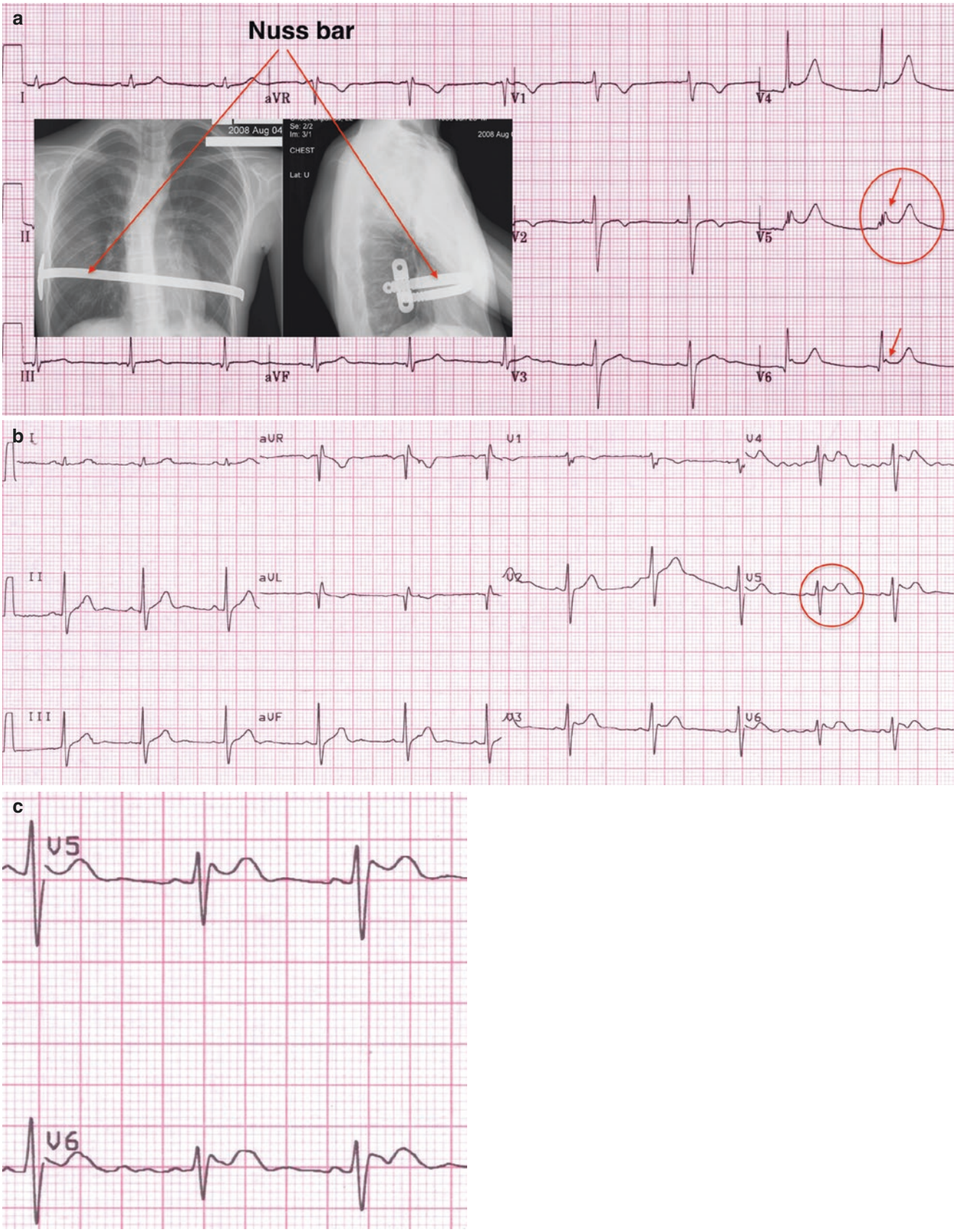


Fig. 8.9 (**a**, **b**) Two patients who underwent the Nuss procedure for PEX. The stainless steel bar placed in the chest produces this interesting artifact. In the precordial leads from V3 to V6, it is possible to find small R waves, concave ST elevation, and large J waves. (**c**) Close-up: a sort of Brugada on the left side! The "wrong" Brugada, the bartender said

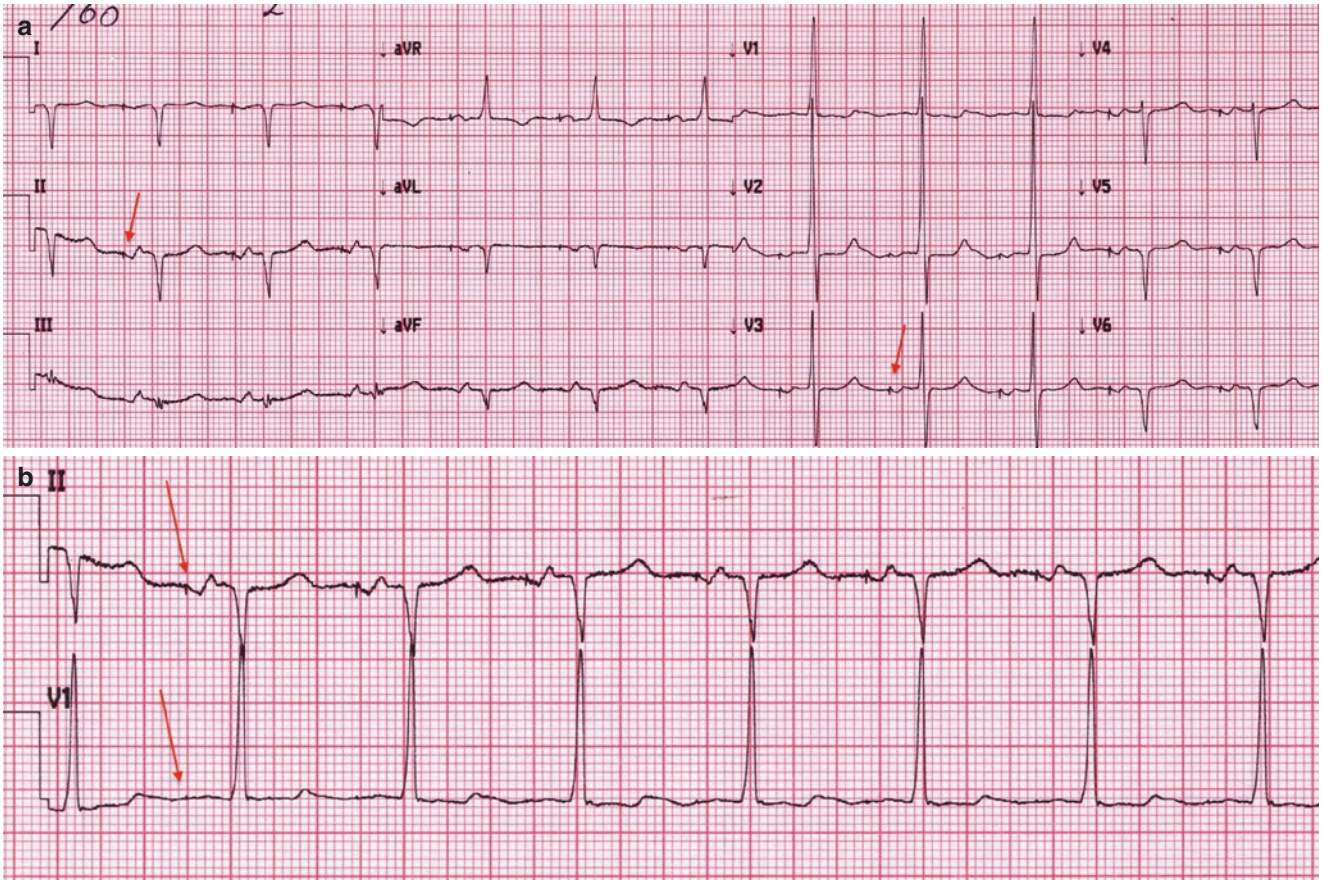


Fig. 8.10 (**a**) Twenty years old with TGA who underwent Senning operation. The sinoatrial dysfunction (extensive atrial scar) was treated with an AAI PMK. (**b**) Close-up: the bipolar spike is very small

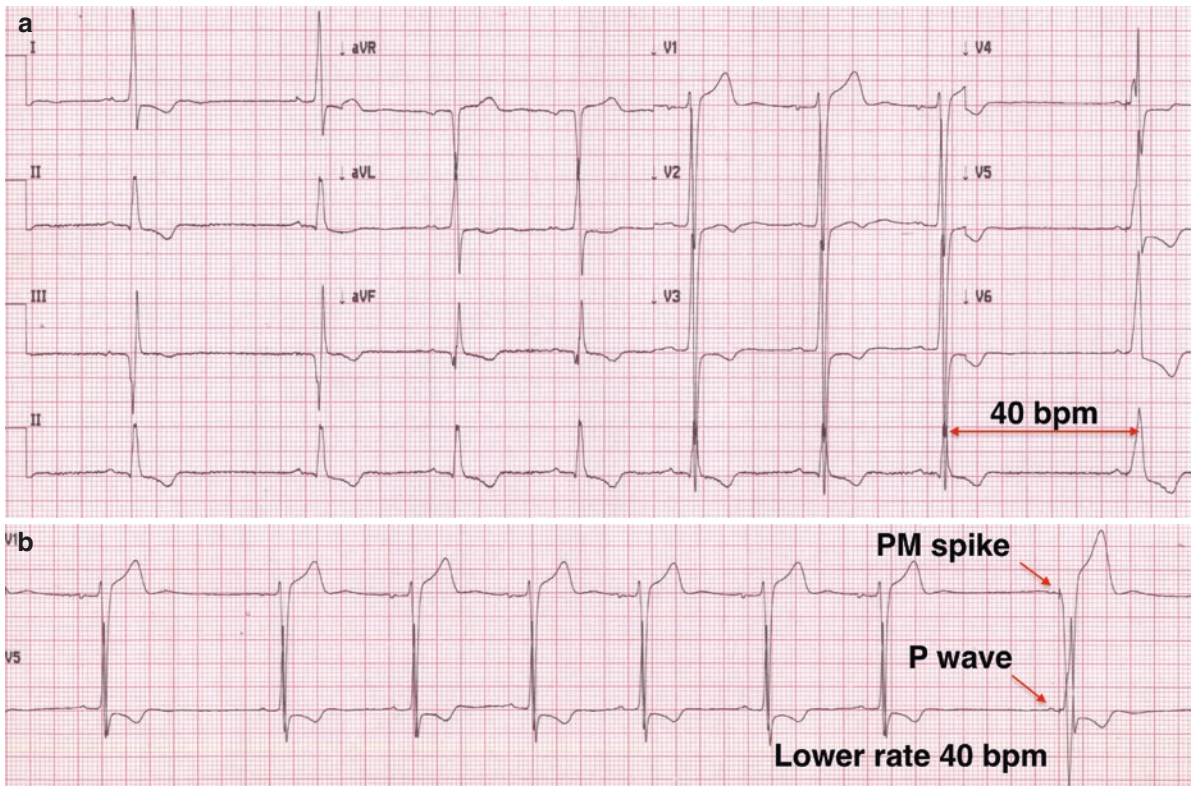


Fig. 8.11 (**a**) Sixteen-year-old male. HCM underwent an ICD implant as secondary prophylaxis after a cardiac arrest. The ICD also works as a VVI PM. After a sinus arrest, the device, that has no atrial sensing, releases a ventricular spike despite a late P wave. (**b**) Close-up

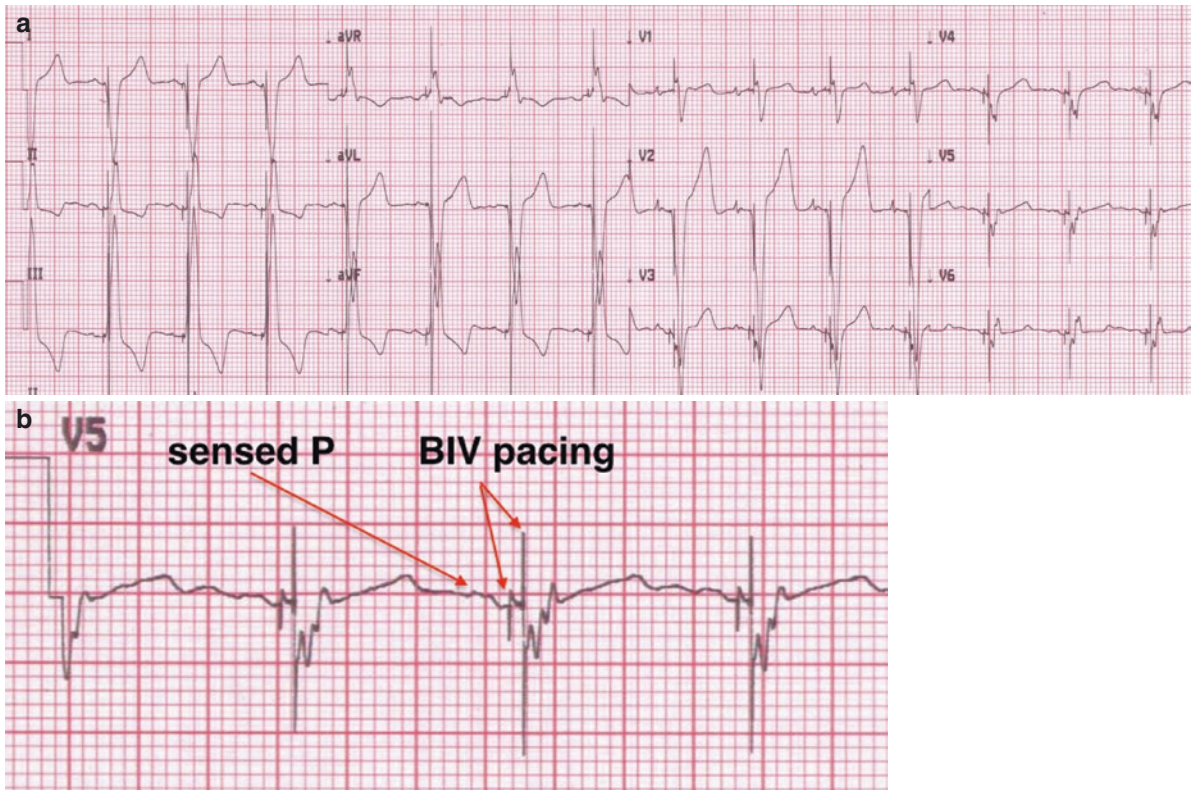


Fig. 8.12 (**a**) Ten-year-old female, neonatal complete AVB treated by pacing since birth. She developed pacemaker syndrome with severe left ventricular impairment. A biventricular pacing system (cardiac resynchronization therapy) was implanted. The PMK works in DDD: the atrial activity is sensed and followed by a double ventricular spike. (**b**) Close-up

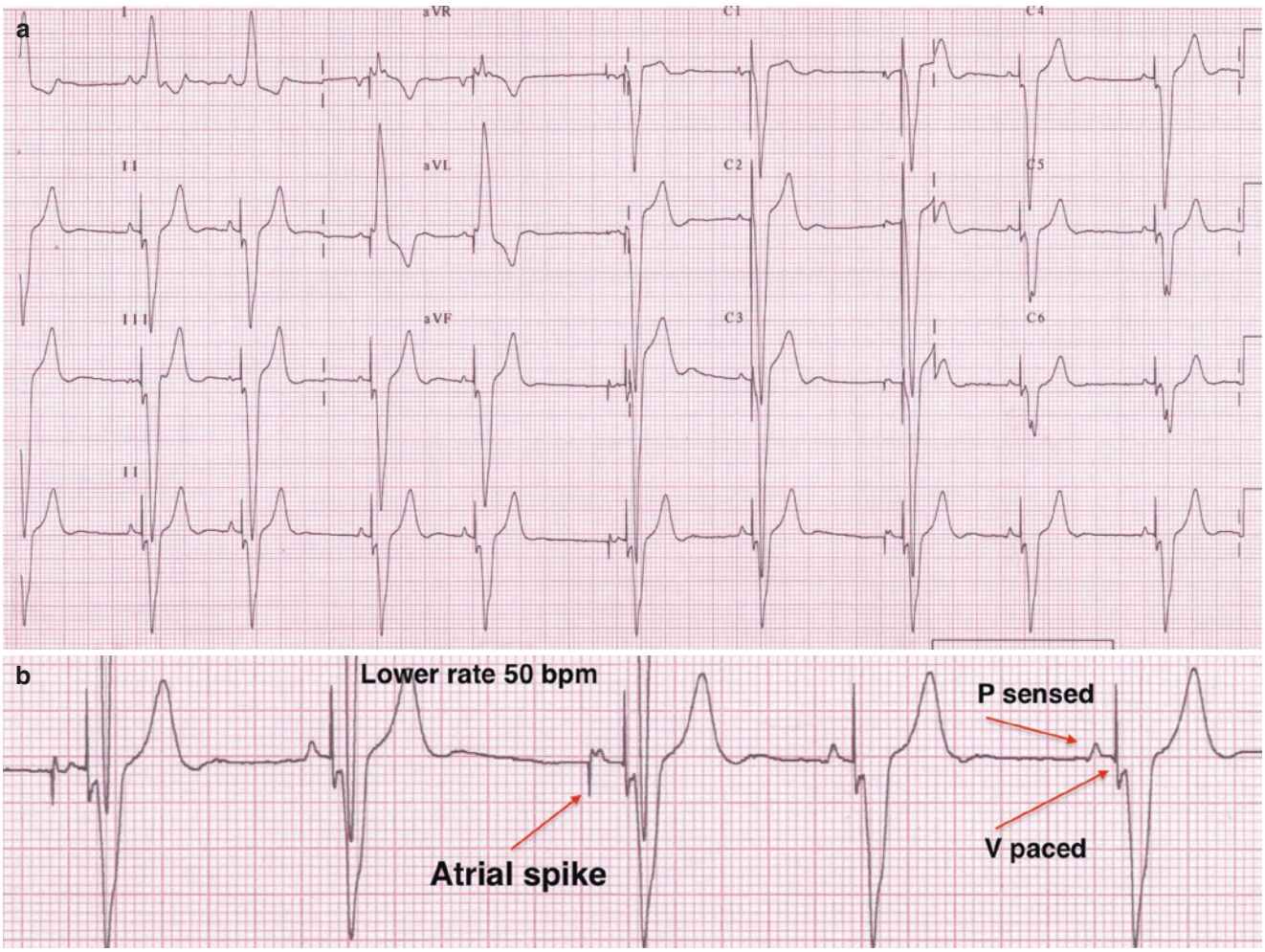


Fig. 8.13 (**a**) Two-year-old boy affected by complete AVB, treated with epicardial DDD PMK. The lower rate is set at 50 bpm. When the spontaneous atrial rate is >50 bpm, the PM works in atrial sensing – ventricular pacing. (**b**) When the atrial rate is <50 bpm works, the PM stimulates both atrium and ventricle

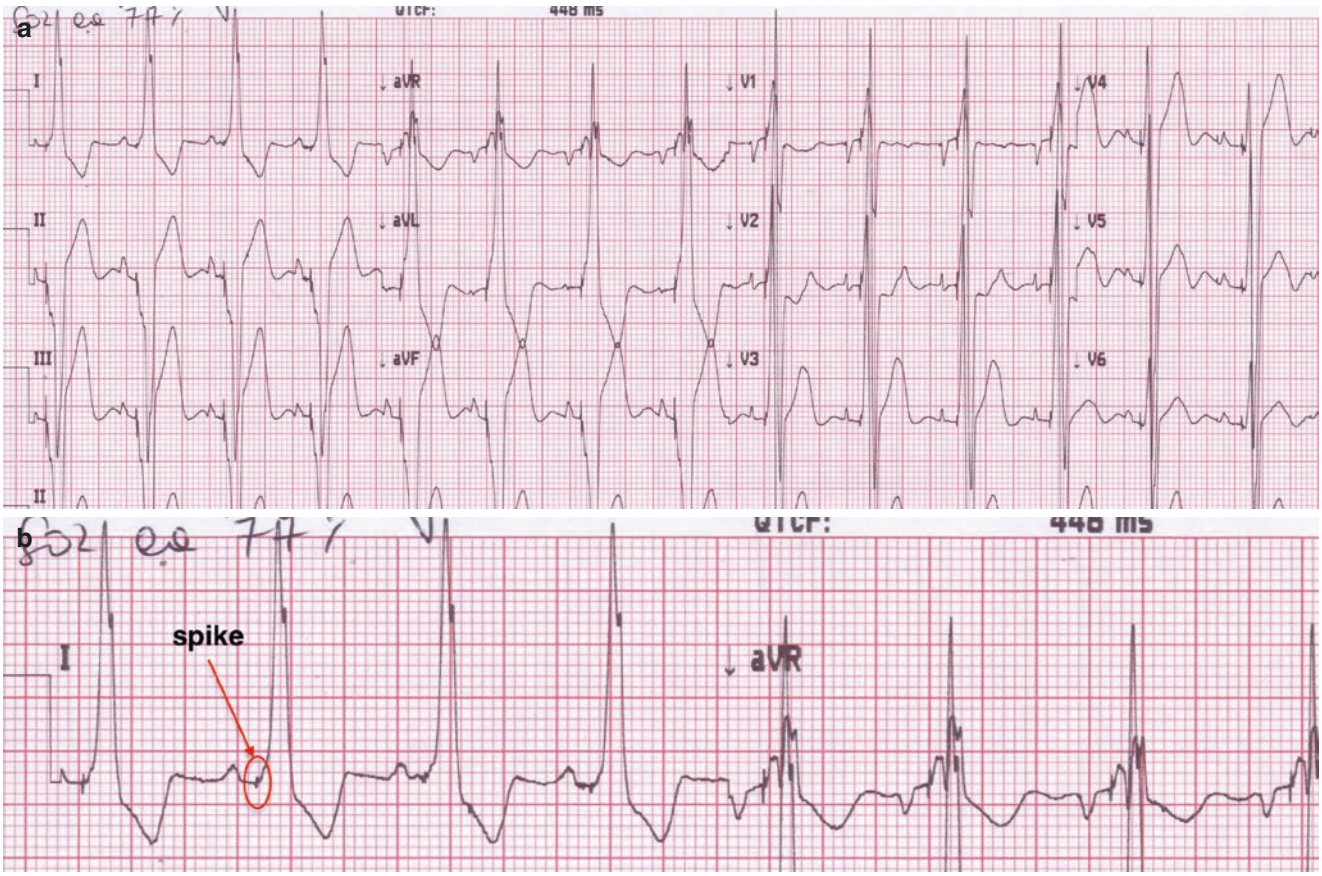


Fig. 8.14 (**a**) Eight-year-old boy with complex congenital heart disease (single ventricle palliated with Fontan operation) and complete AVB, Treated with epicardial DDD PMK. Due to the catheter working in bipolar modality, the spike is very small, sometimes difficult to see. (**b**) Close-up

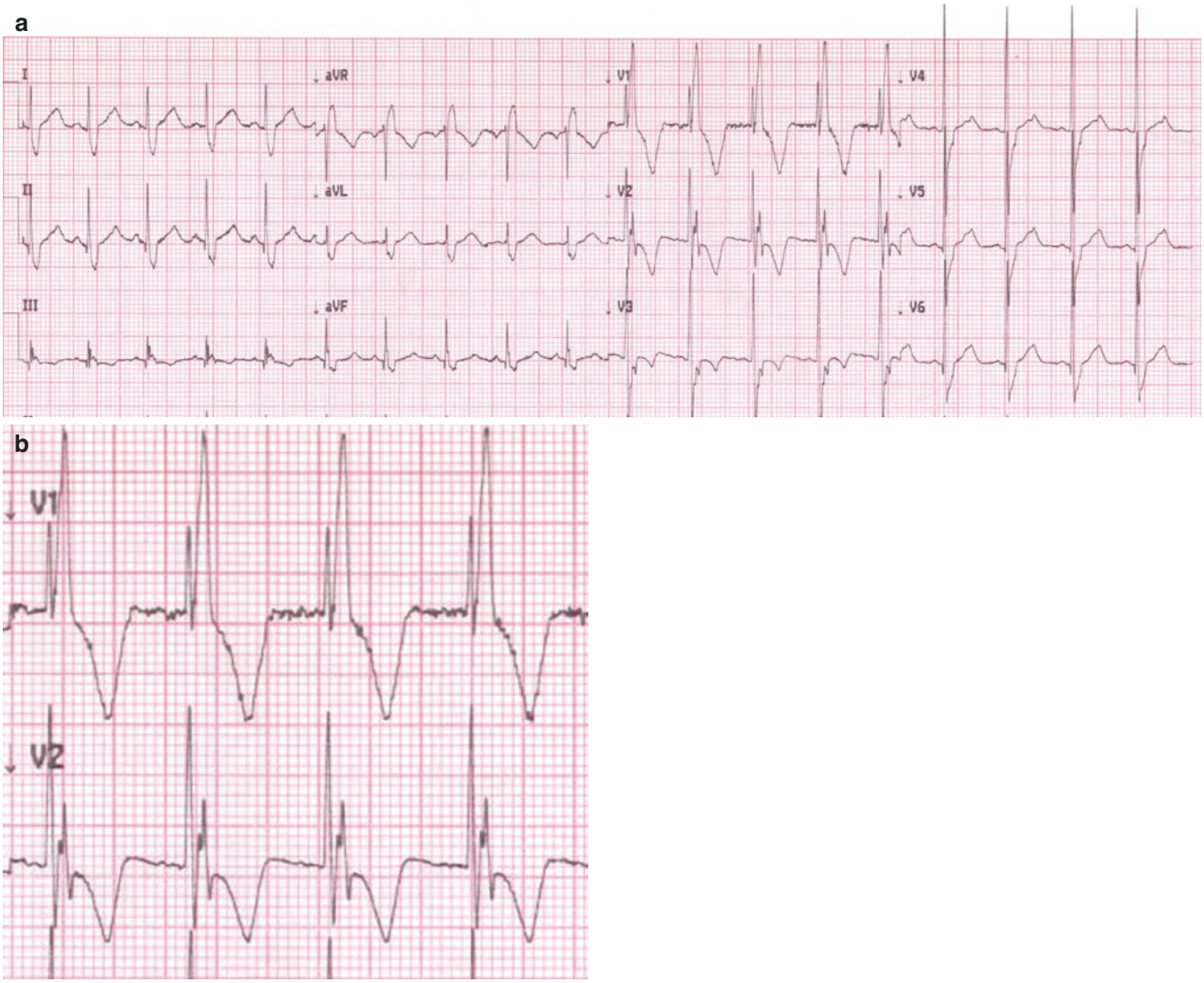


Fig. 8.15 (**a**) Ten-year-old boy. No cardiac disease. Screening ECG found an RBBB. The initial part of the QRS simulated a PMK spike. The ECG machine was deceived, leading to an incorrect automatic report. (**b**) Close-up

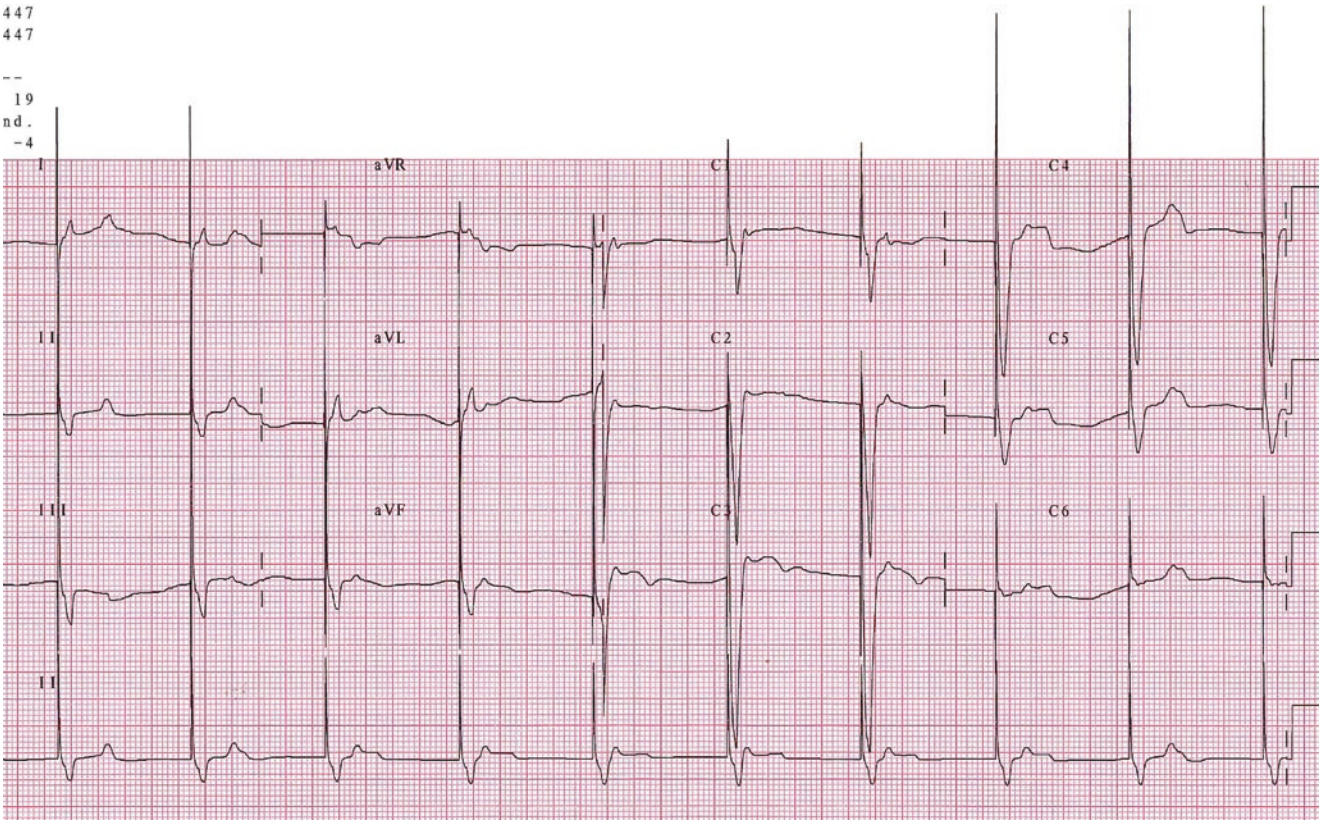


Fig. 8.16 Six-year-old boy with familial complete AVB. Transvenous DDD PMK. A prominent spike can be noted

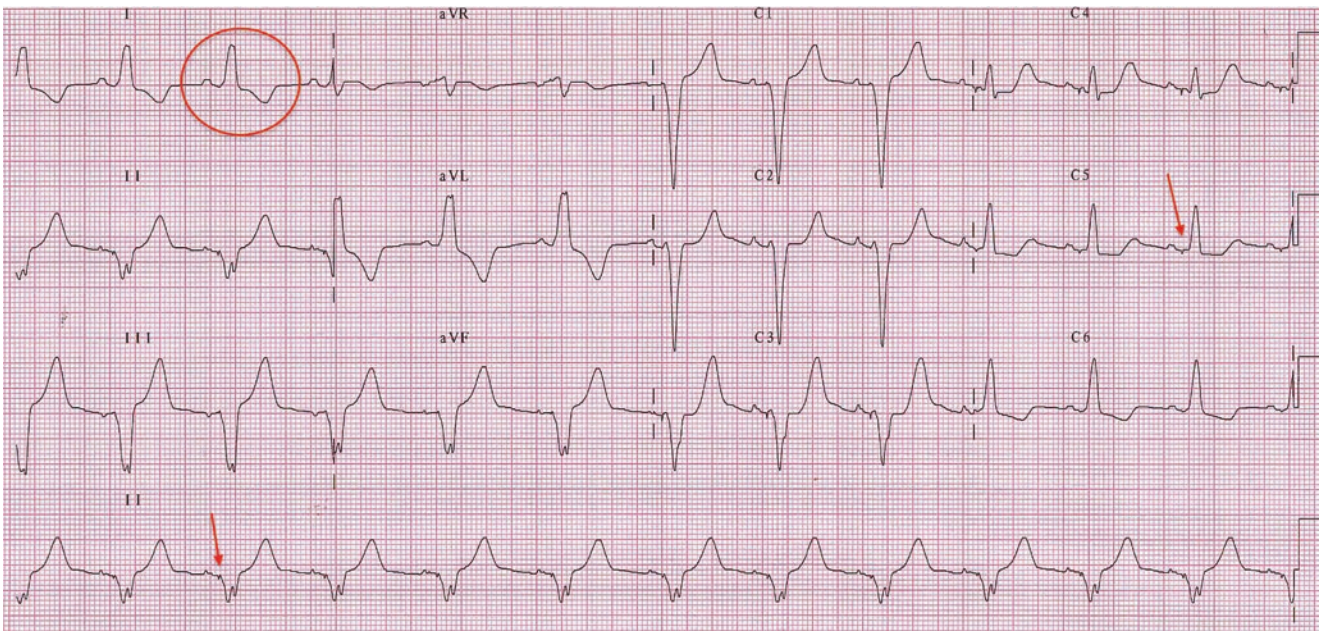


Fig. 8.17 Another case of DDD pacemaker in an 8-year-old girl with complete AVB. The bipolar leads make the spikes almost invisible (see circle in lead I)

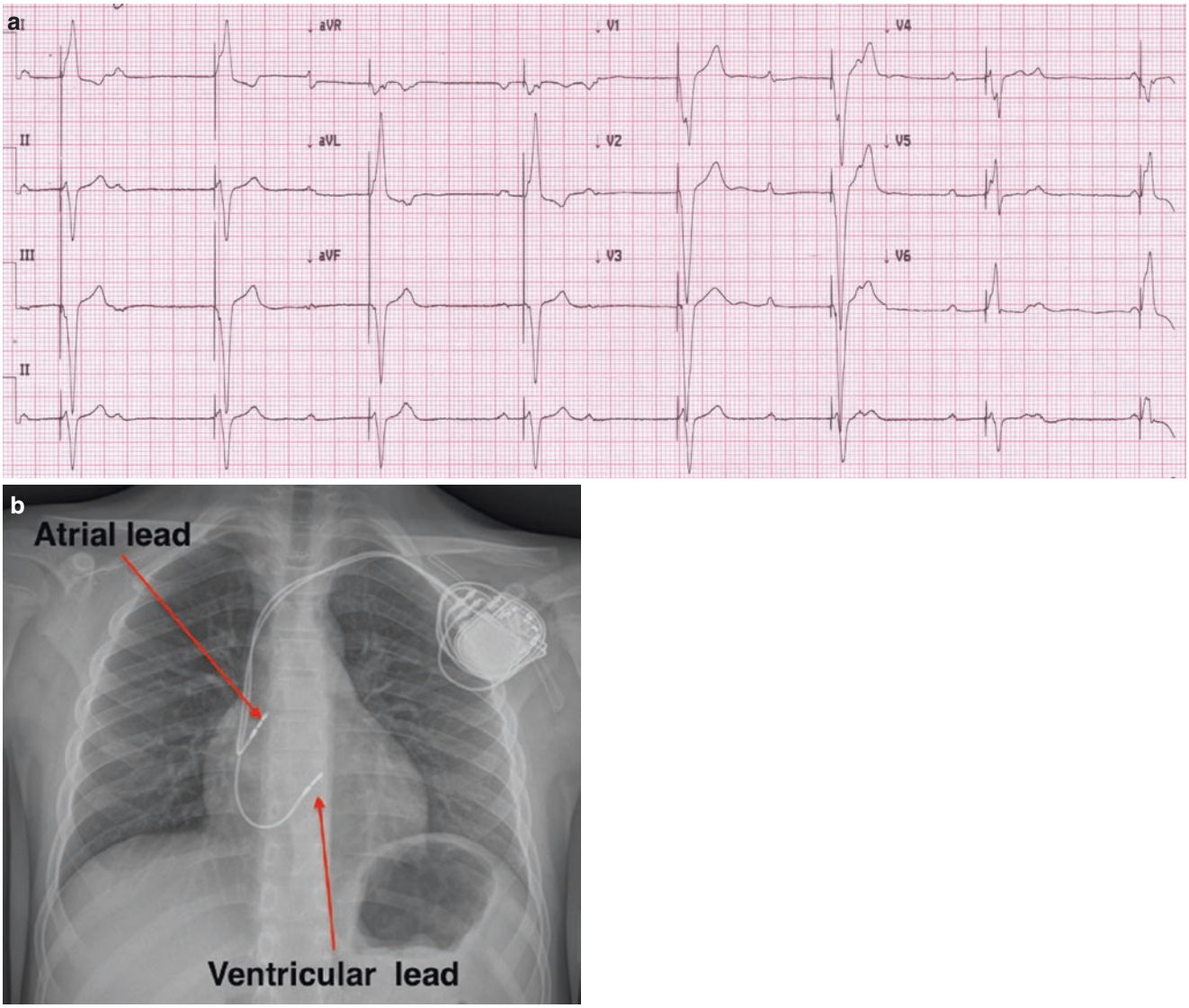


Fig. 8.18 (**a**) Ten-year-old boy with PMK DDD implanted for complete AVB. The right ventricular lead was placed in the low aspect of the interventricular septum. The resultant QRS has of course an LBBB morphology. (**b**) Chest X ray

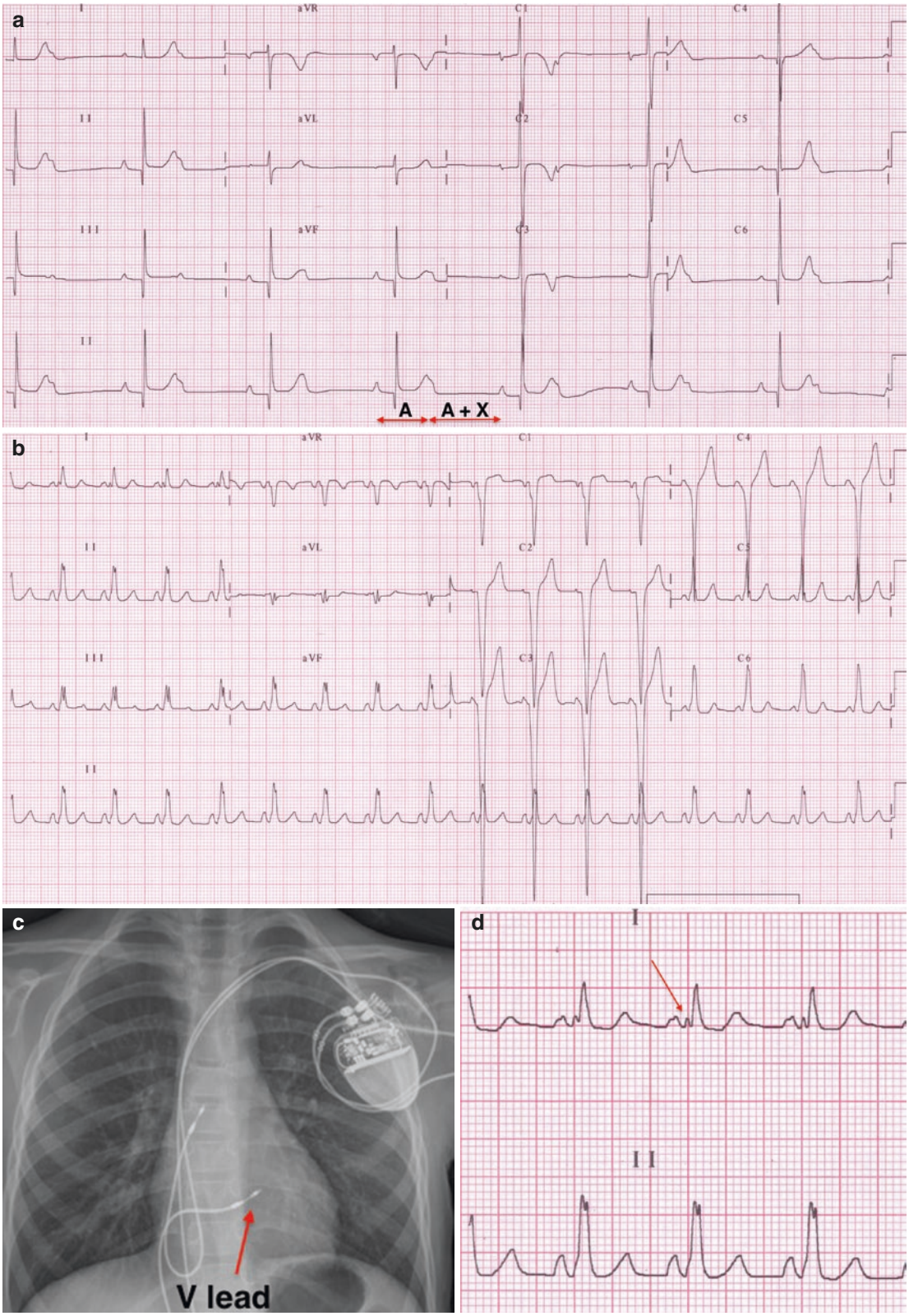


Fig. 8.19 (**a**) Ten-year-old boy affected by complete AVB. A ventriculophasic sinus arrhythmia can be noted. This is a particular phenomenon commonly observed in patients with complete atrioventricular (AV) block. Typically, the P-P intervals which contain a QRS complex are shorter than the PP intervals which do not contain it. (**b**) A transvenous DDD PMK was implanted. The paced ECG simulates ventricular pre-excitation: short PR interval and wide QRS. (**c**) The reason is a selective pacing of the anterosuperior aspect of the interventricular septum that simulates the ventricular end of a right anterior accessory pathway. (**d**) The spike mimics a delta wave (many cardiologists were deceived by this ECG, most of them reporting "WPW"). A funny fate for the boy, from too much late excitation to much early excitation

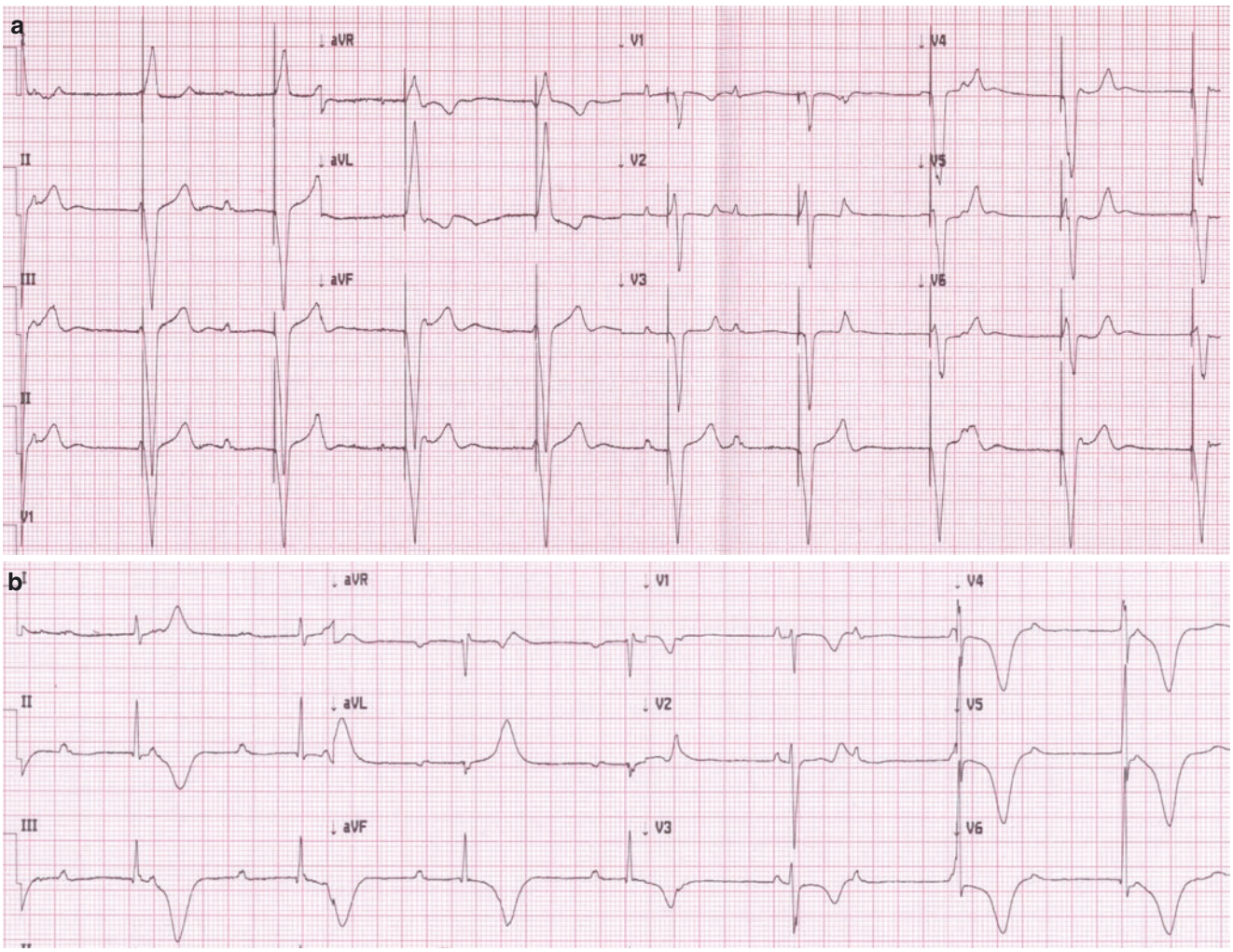


Fig. 8.20 (**a**) Nine-year-old female implanted with a VVI PMK for complete AVB. The lower rate is 55, with hysteresis of 5 (the PM paces at 55 bpm but intervenes only if the spontaneous heart rate is below 50 bpm). (**b**) The spontaneous junctional beats show large negative T waves: this is a form of electric memory

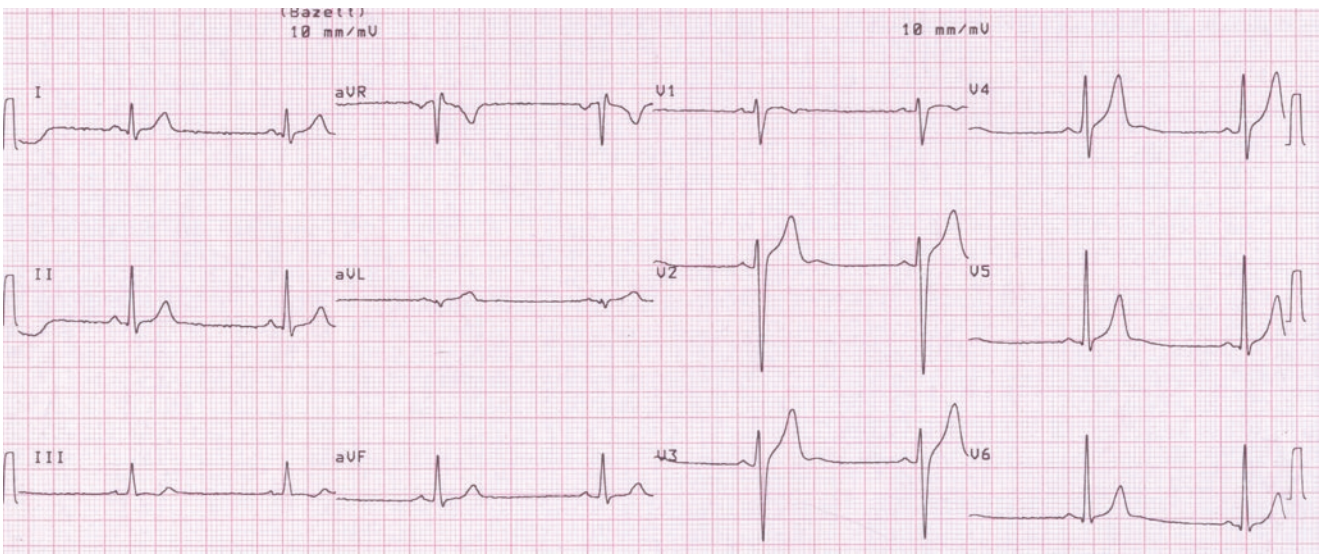


Fig. 8.21 Fourteen-year-old female with epilepsy and history of PMK. In this case, the bradycardia is the consequence and not the cause of the PMK. She was implanted with a vagal PMK, a pacing system that stimulates the X nerve, a non-pharmacological option for some forms of epilepsy

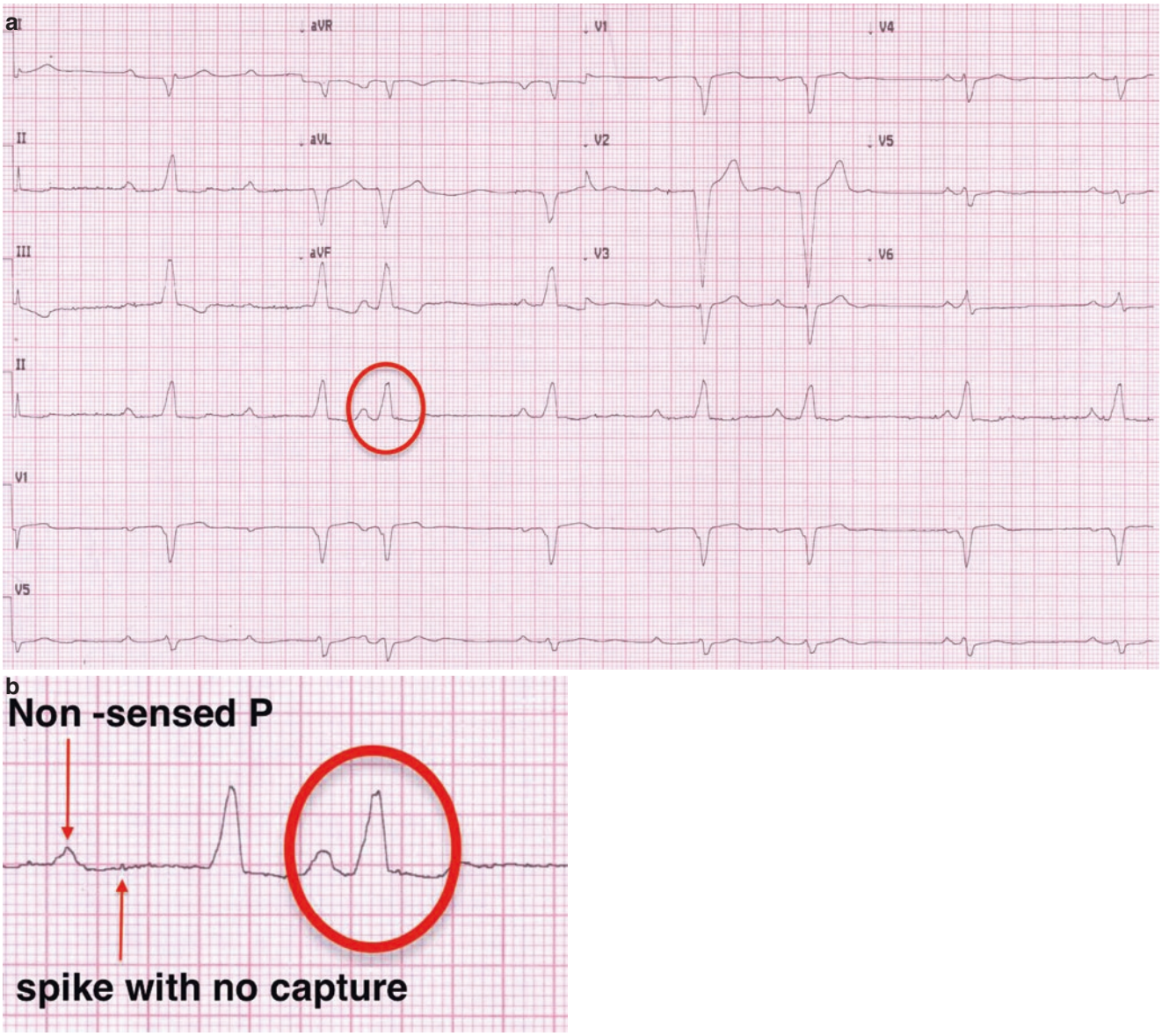


Fig. 8.22 (**a**) Seventeen-year-old girl with DDD PMK for acquired idiopathic complete AVB. The PMK spikes are almost invisible (bipolar leads). (**b**) Close-up. The irregular rhythm is due to a sensing deficit of the atrial leads. The spontaneous atrial activity is non-sensed and then the PMK delivers atrial spikes in the atrial refractory period. Only in one beat the atria correctly sensed (circle) and appropriately followed by a ventricular spike. The atrial lead has been found unstable and has been fixed

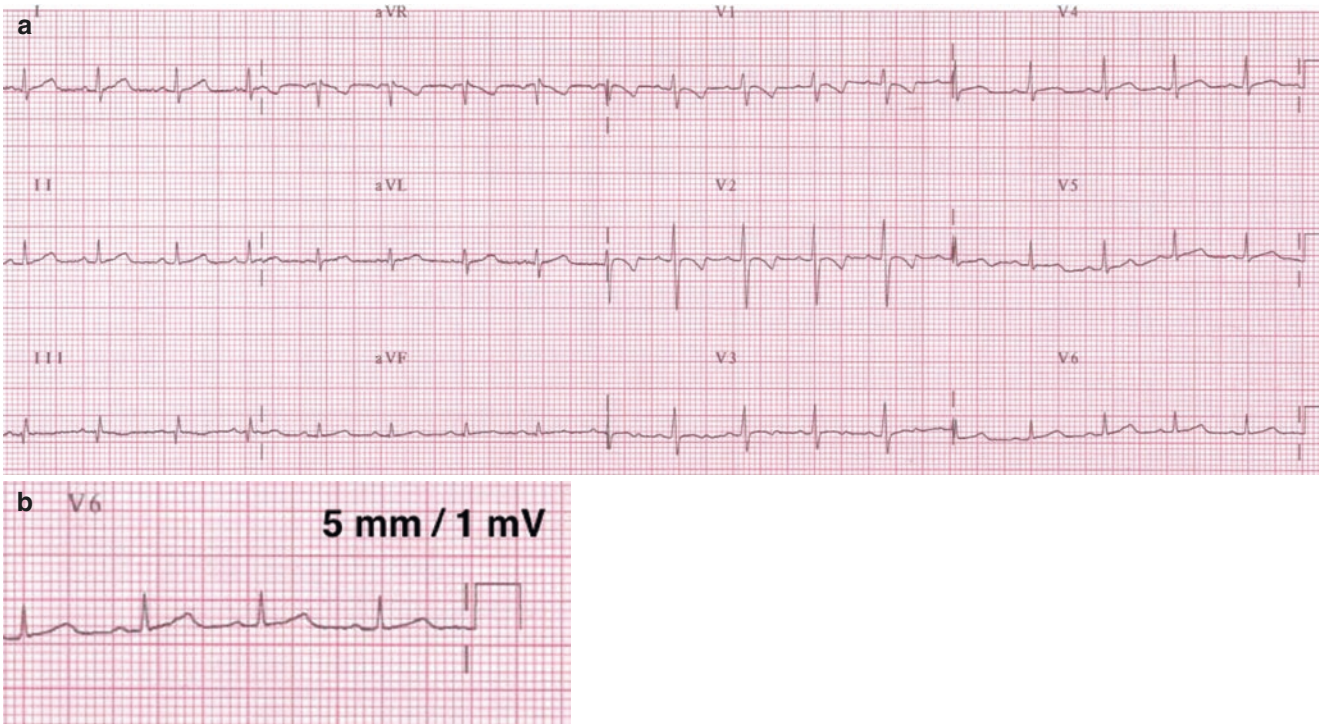


Fig. 8.23 (**a**) Ten-year-old boy admitted with elevated creatine kinase and diagnosis of myositis. The ECG raised the suspicion of myocarditis, due to the diffuse low QRS voltages. (**b**) Paying more attention, it is possible to note a half then standard calibration, 5 mm/1 mV

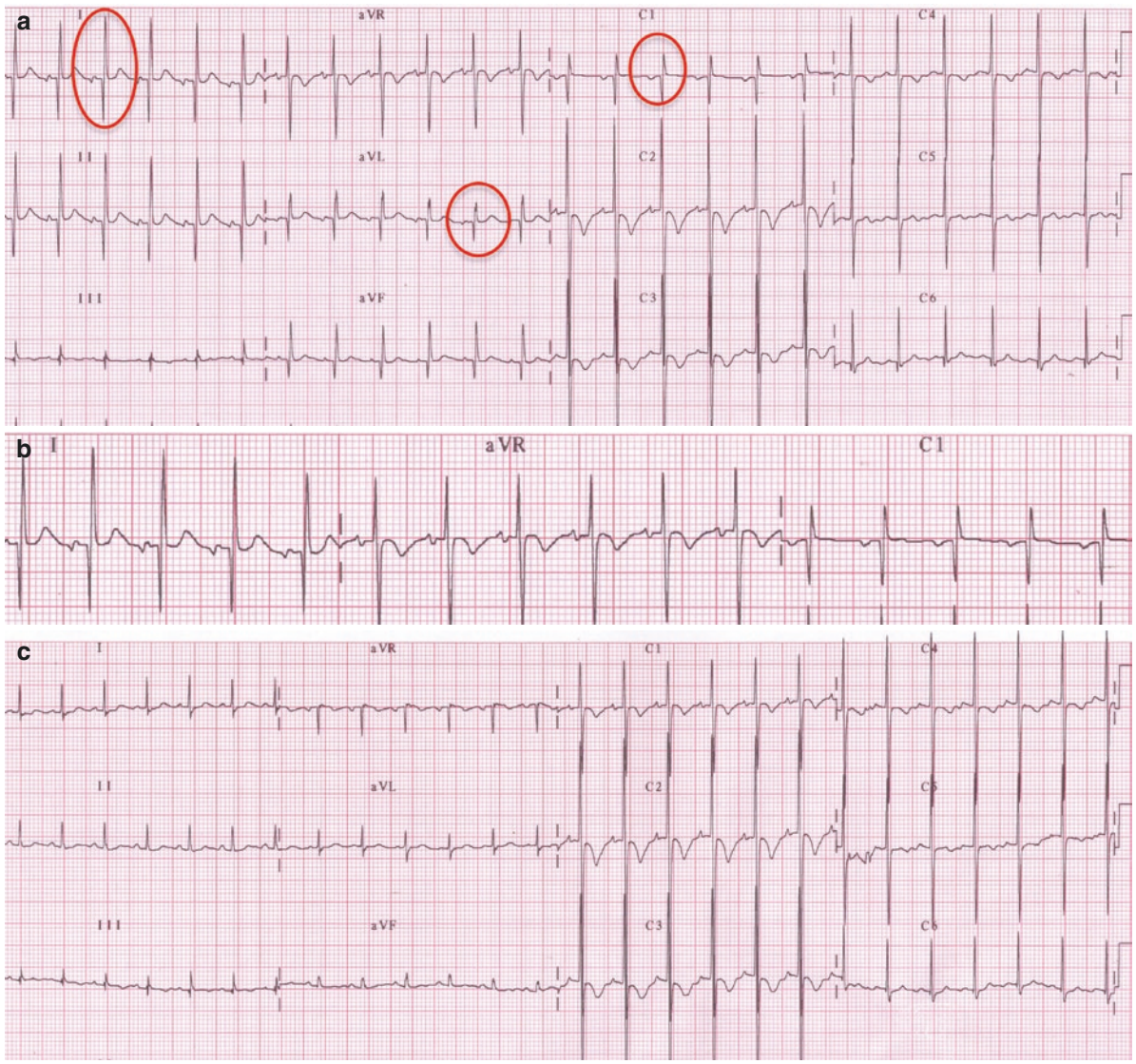


Fig. 8.24 Seven-year-old girl admitted for chest pain. (**a**) The first ECG was suggestive for ischemia and raised the suspicion of ALCAPA or myocarditis; note also the unusual aVR. (**b**) Close-up. (**c**) A repeated ECG was normal; a lead misplacement was discovered; the right arm and V1 electrodes were inverted (both are *red*)

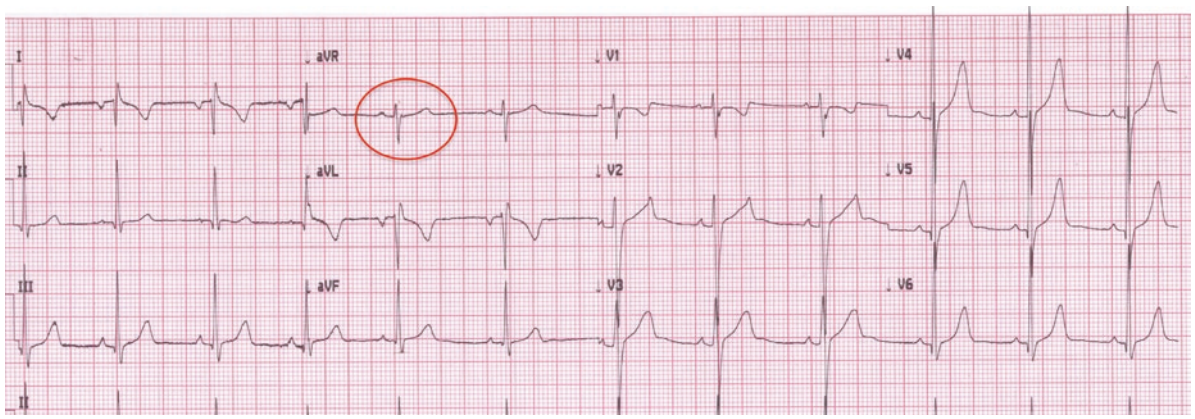


Fig. 8.25 Twelve-year-old boy. The "classic" lead misplacement, the inversion of the right and left arms. If the machine is no longer available, it will be enough to auscultate the patient to confirm the misplacement and to rule out dextrocardia. The stethoscope, as well as aVR, tells the truth

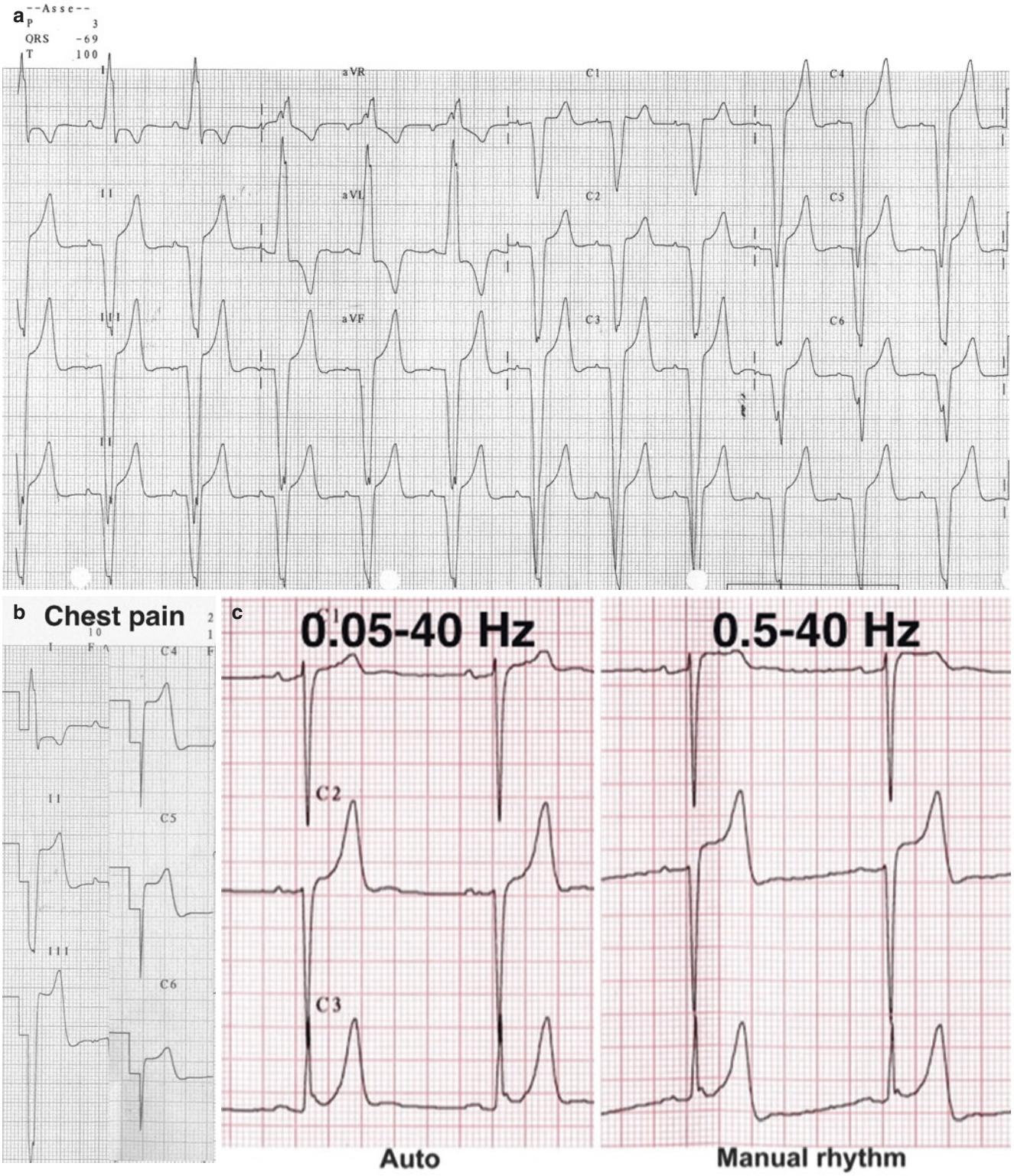


Fig. 8.26 Seventeen-year-old girl with severe obstructive HCM who underwent Morrow myectomy. (**a**) Basal ECG; (**b**) during chest pain, the ECG showed an impressive ST change, suggestive of acute subepicardial ischemia. Many hypotheses can be proposed in this setting; one can be a myocardial bridge. (**c**) The truth was an artifact due to the manual mode of the ECG recording that in some machine has a different filter. The filter at 0.5–40 Hz can create this kind of artifact, while the filter at 0.05–40 prevents it

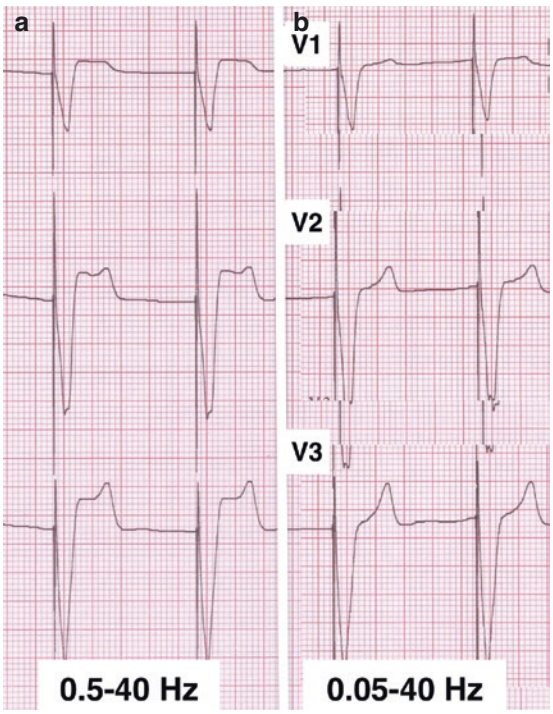


Fig. 8.27 As in the previous figure, in a 30-year-old woman with complex CHD who underwent Fontan palliation and PMK implant, the same phenomenon occurred. She was admitted for chest pain. (**a**) Usually in PMK carrier, the ECG is not reliable to judge the ST segment, but in this case the ST elevation in V2–V3 during chest pain is worrying. (**b**) The repeated tracing with the correct 0.05–40 Hz filtering is reassuring

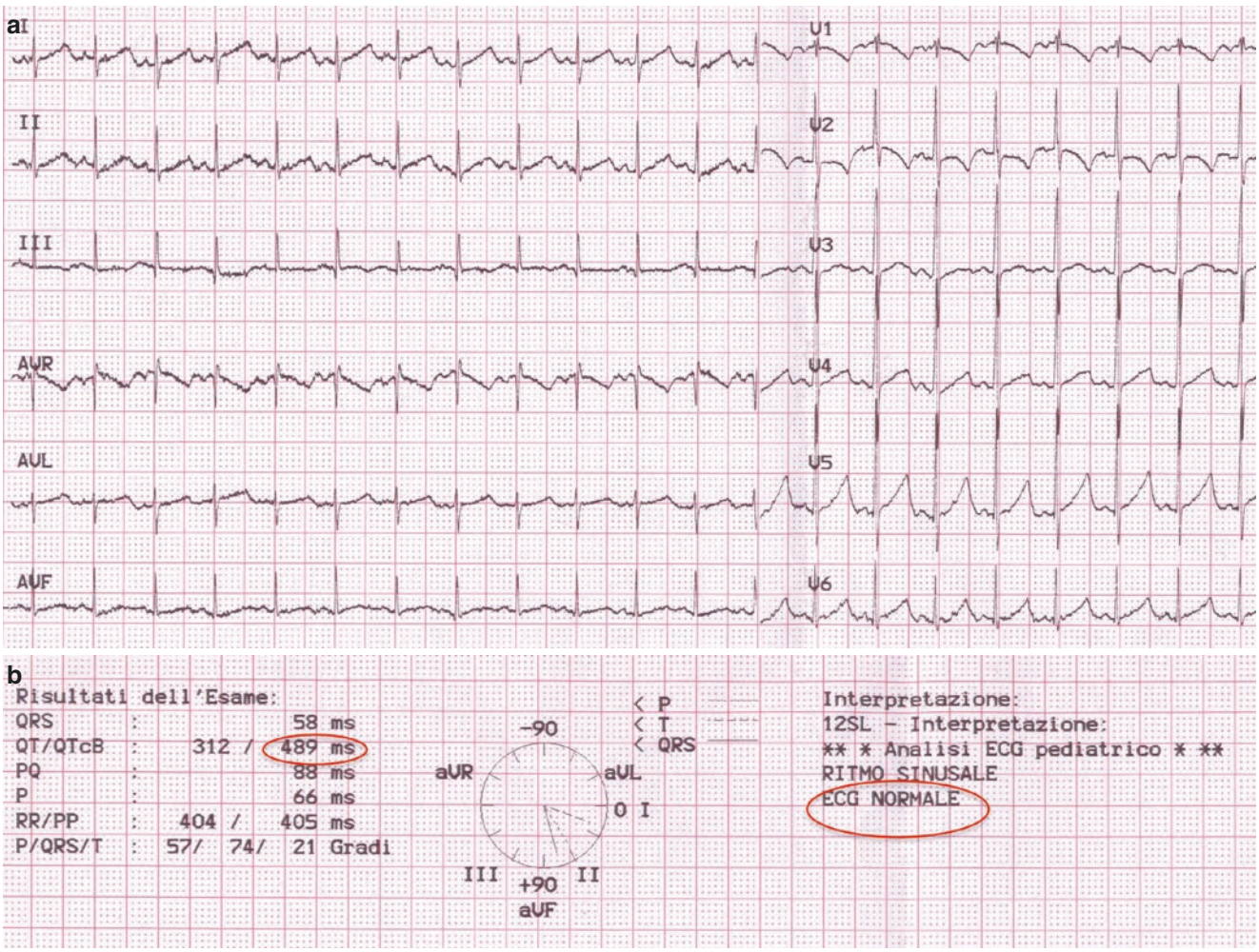


Fig. 8.28 (**a**) Six months old. The QT is prolonged (QTc 490 ms). The machine correctly measures the QTc but does not report it, answering "Normal ECG." (**b**) Close-up

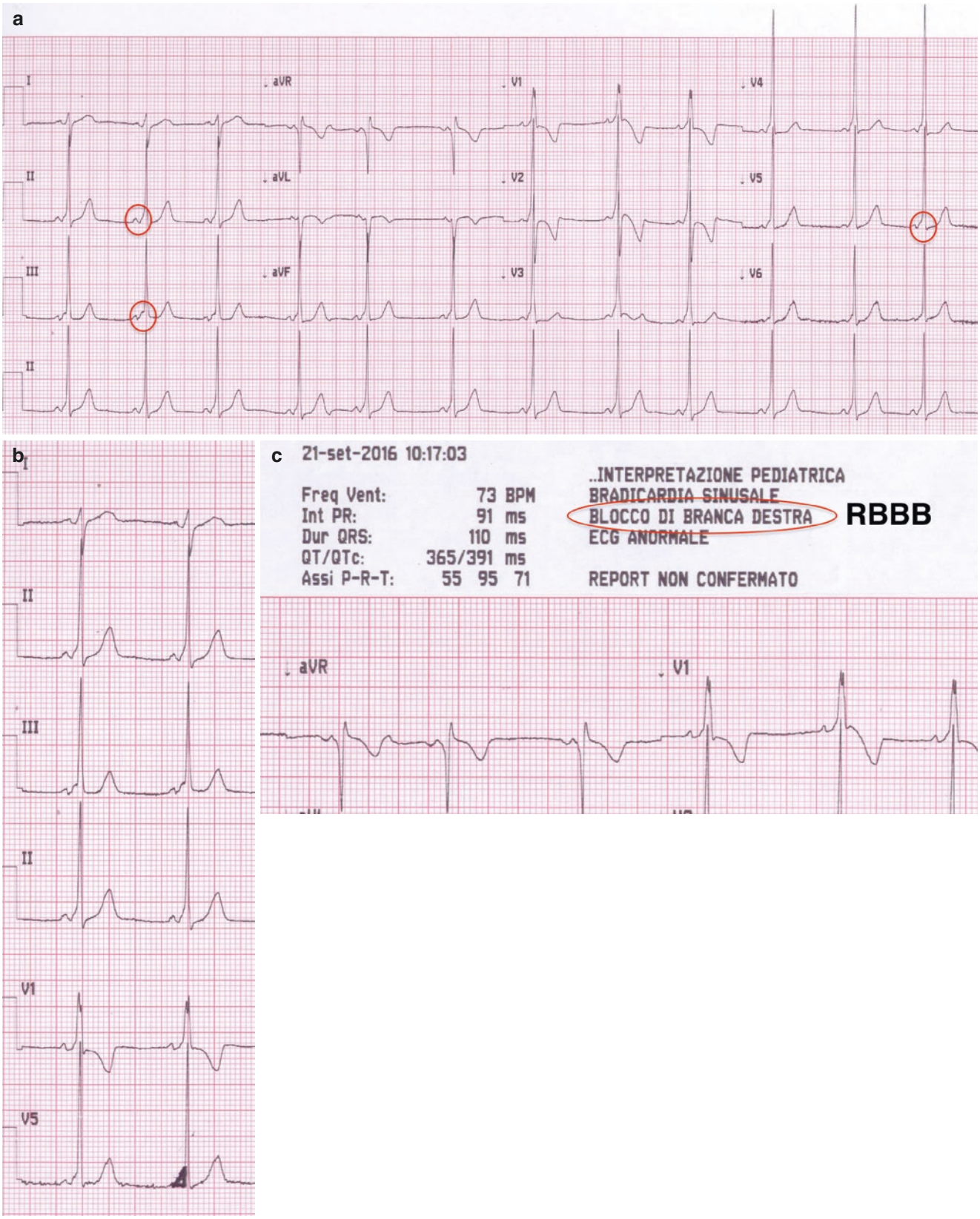


Fig. 8.29 (**a**) Thirteen-year-old boy. The automatic answer does not report the WPW pattern. Instead, the report mentions an RBBB. (**b**, **c**) Close-up

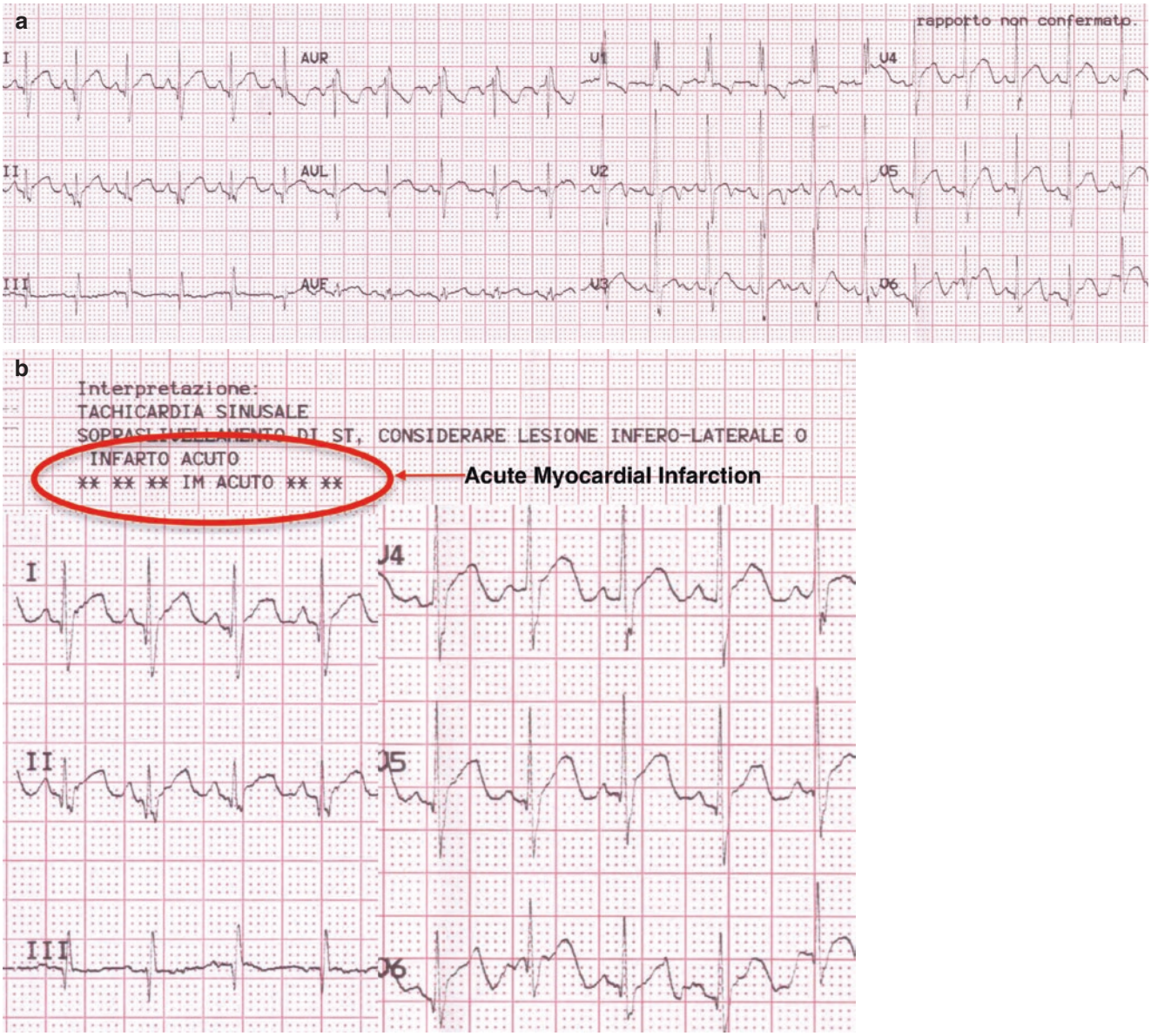


Fig. 8.30 (**a**) One-year-old boy with normal ST elevation. The ECG machine misunderstands the normal ST elevation for ischemia. (**b**) Close-up. The machine sees infarction instead of infants

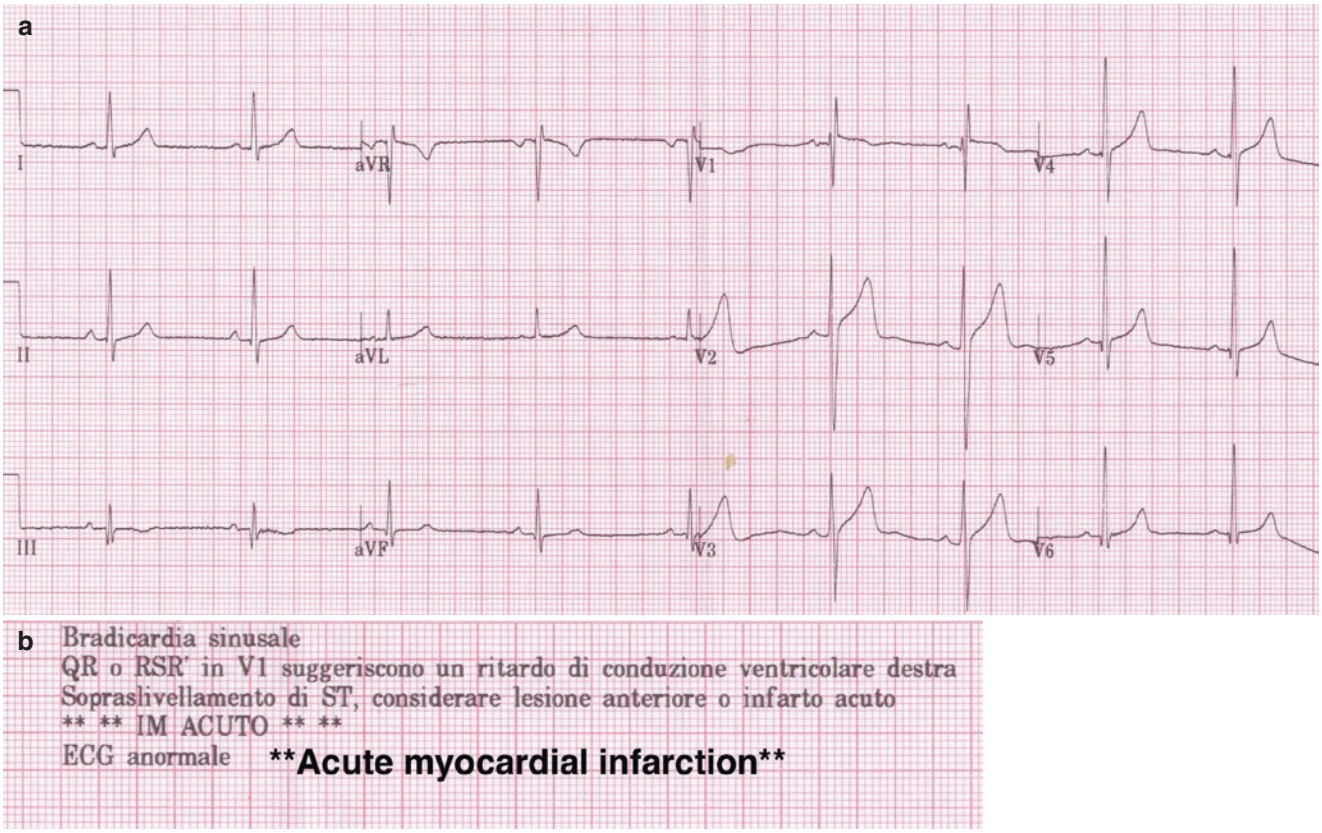


Fig. 8.31 (**a**) As in the previous case, in this 16-year-old boy, the juvenile pattern has been misinterpreted as myocardial infarction. (**b**) Close-up

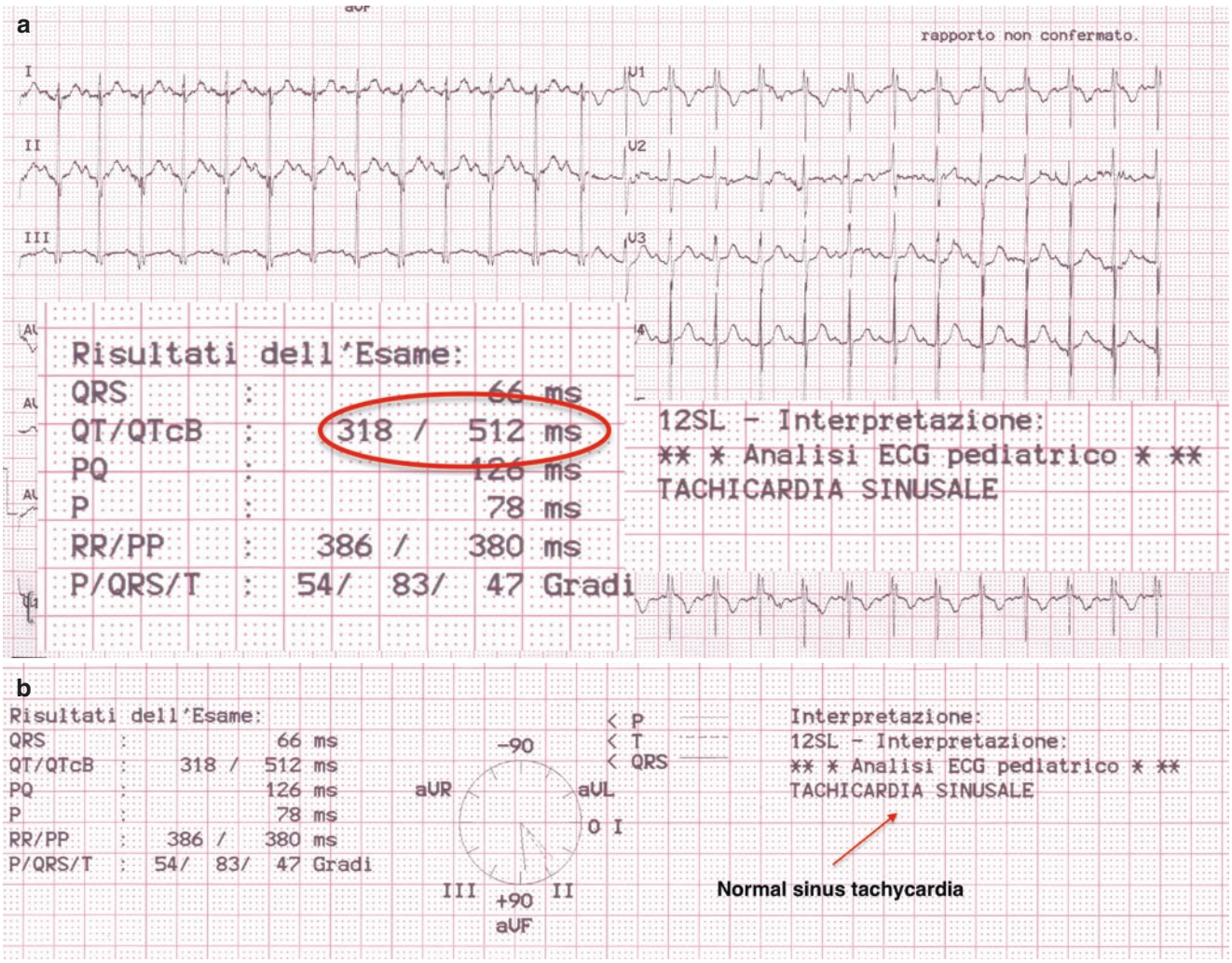


Fig. 8.32 (**a**) Another case of wrong QT calculation by the machine in a 1-year-old boy. The real QTc tends to be short, 370 ms. (**b**) Close-up. The machine answers "QTc 512" but luckily forgets to mention the diagnosis of long QT in the final report. Two mistakes that cancel each other, two negatives make a positive in math

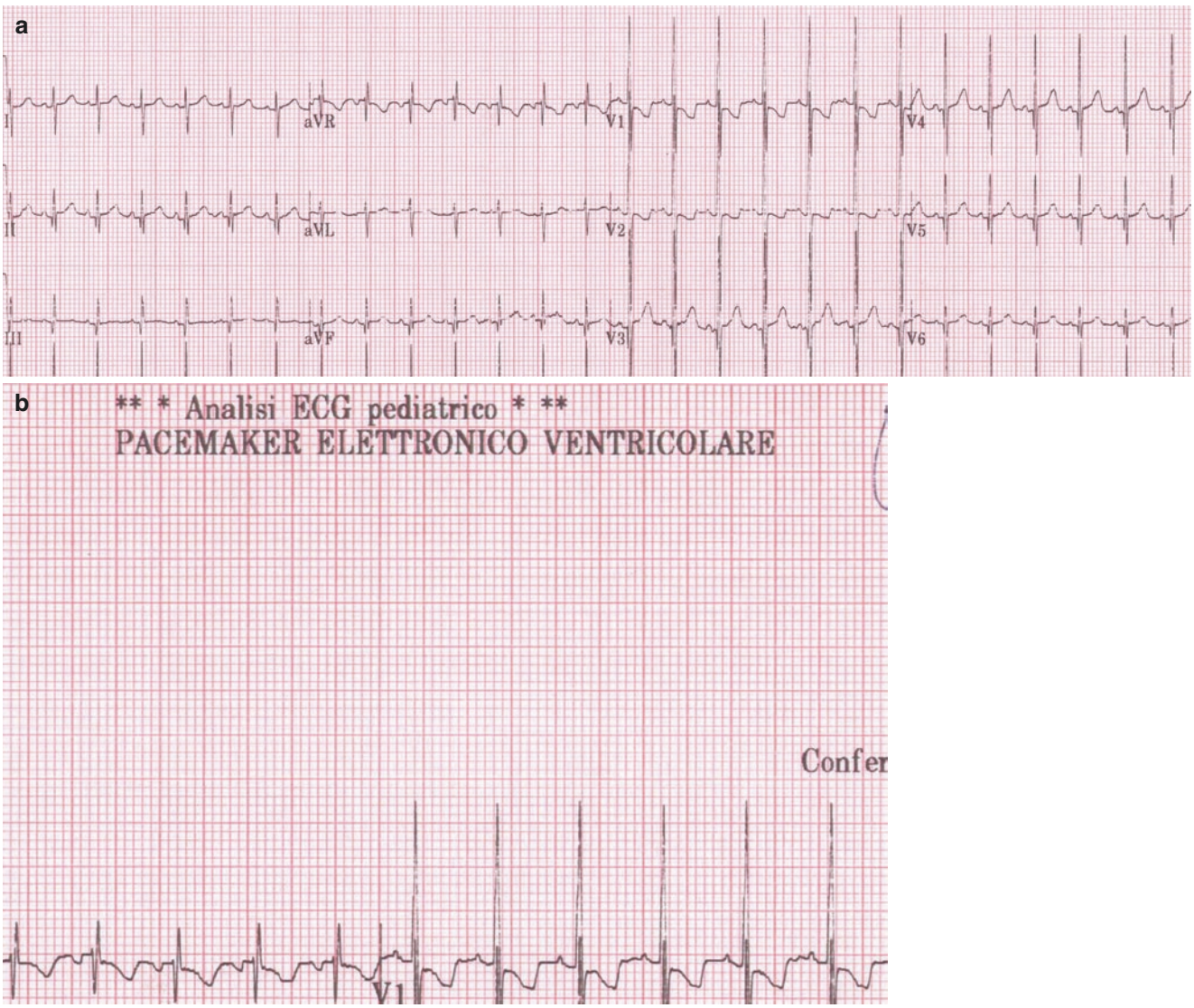


Fig. 8.33 (**a**) A neonate was believed to be a PM recipient by the ECG machine. (**b**) Close-up. The reasons are the very narrow QRS, misunderstood for spike

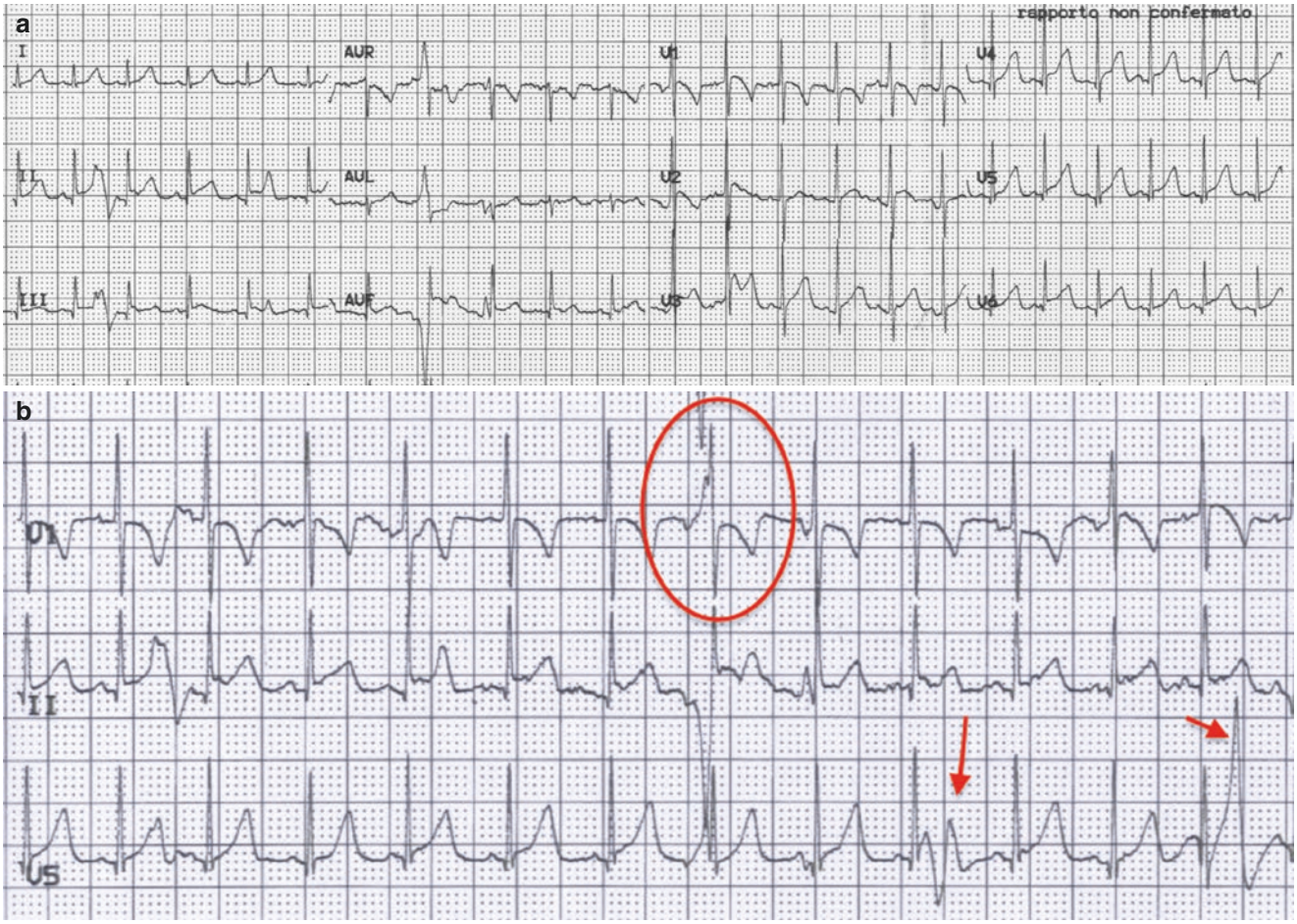


Fig. 8.34 (**a**) Fifteen-year-old boy admitted for racing heart. The movement created artifacts that raised the suspicion of delta wave in a setting of intermittent pre-excitation. (**b**) Actually it is clearly a pseudo-delta wave (*circle*) as the rough artifacts that follow showed (*arrows*)

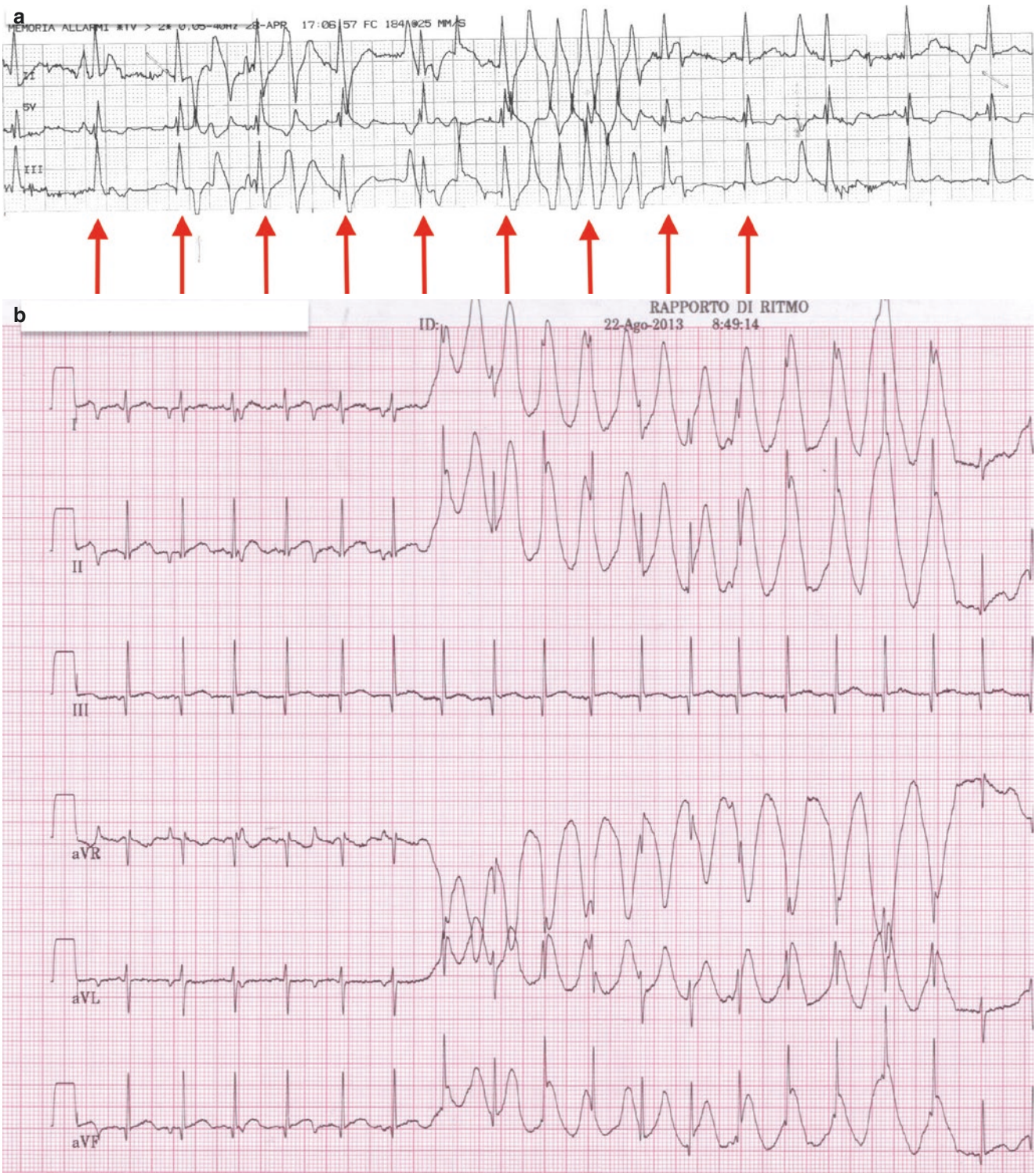


Fig. 8.35 Three different cases of pseudo-VT due to movement or vibratory artifacts. (**a**) Monitor strip in the ward. (**b**) Basal ECG. (**c**) Holter monitoring. In all cases, under the worrying wide complex tachycardia, when paying attention, it is possible to find a normal sinus rhythm

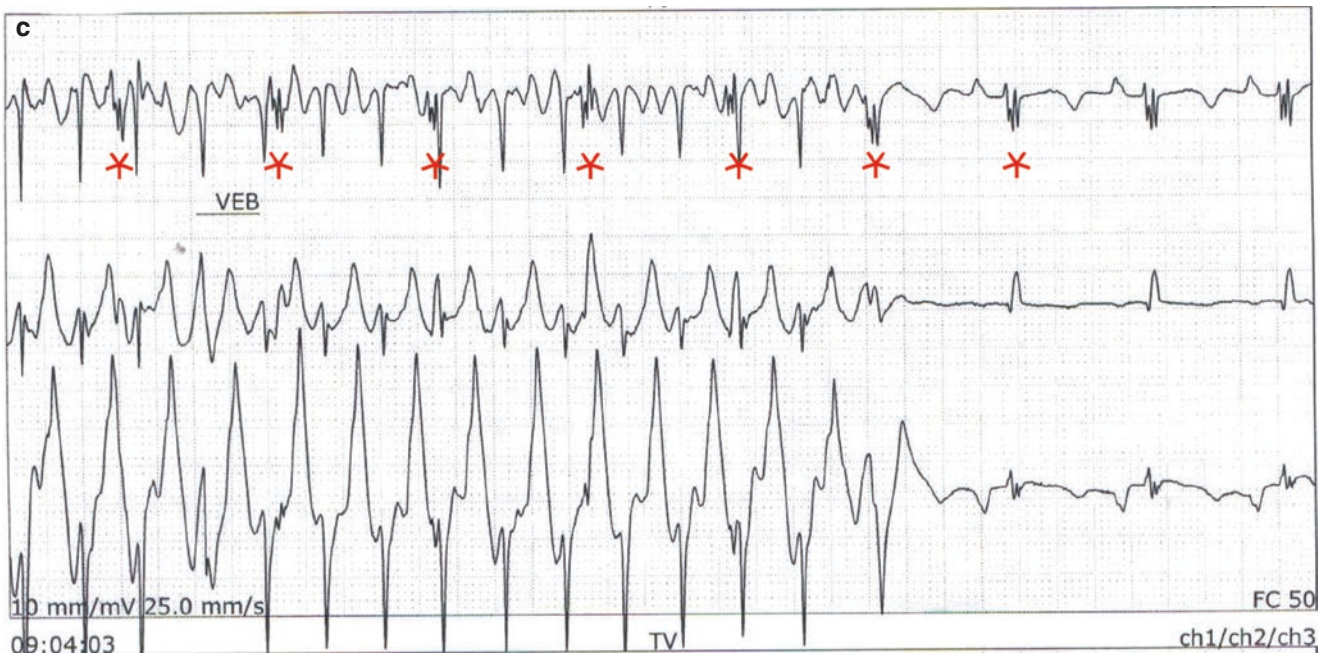


Fig. 8.35 (continued)

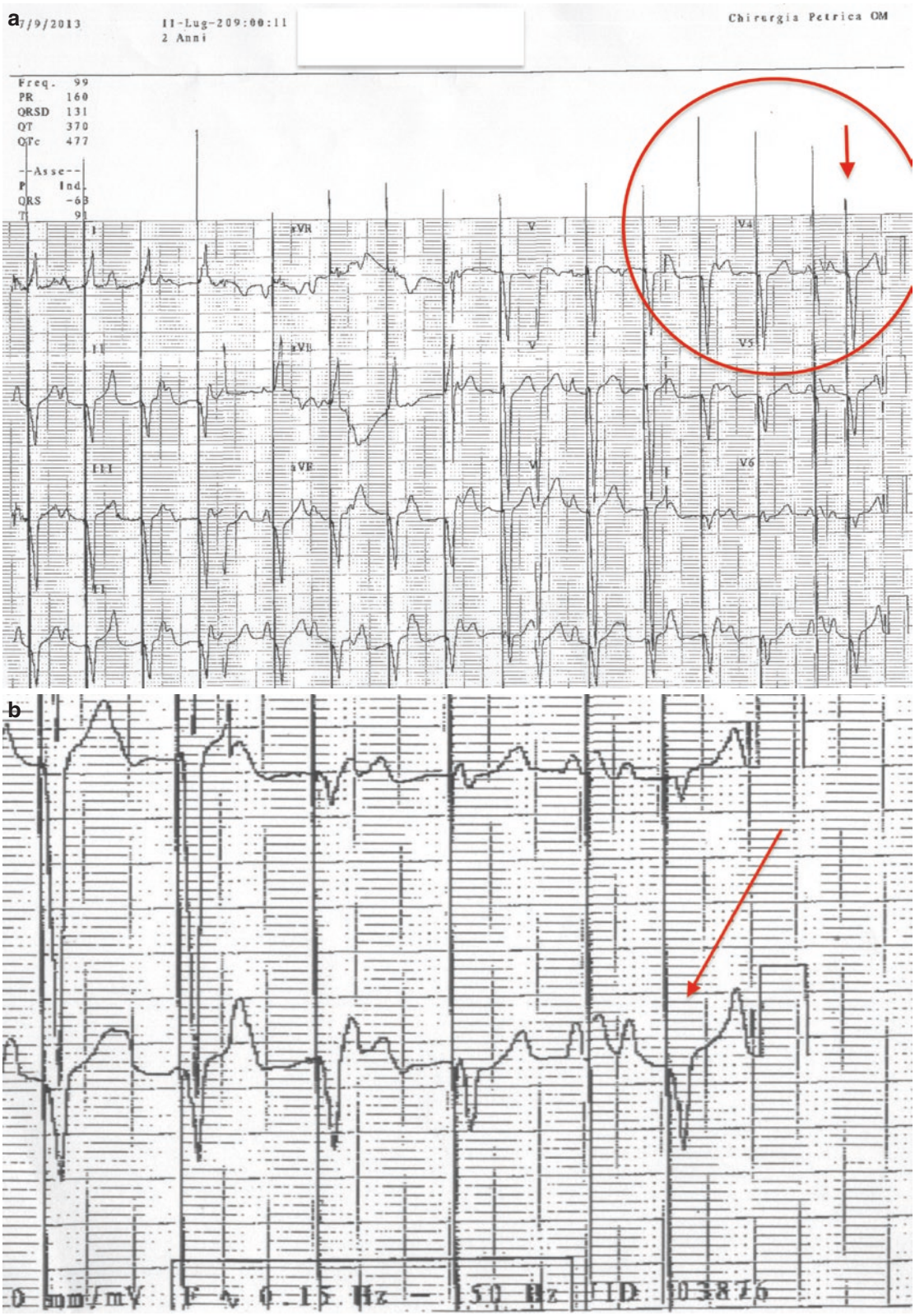
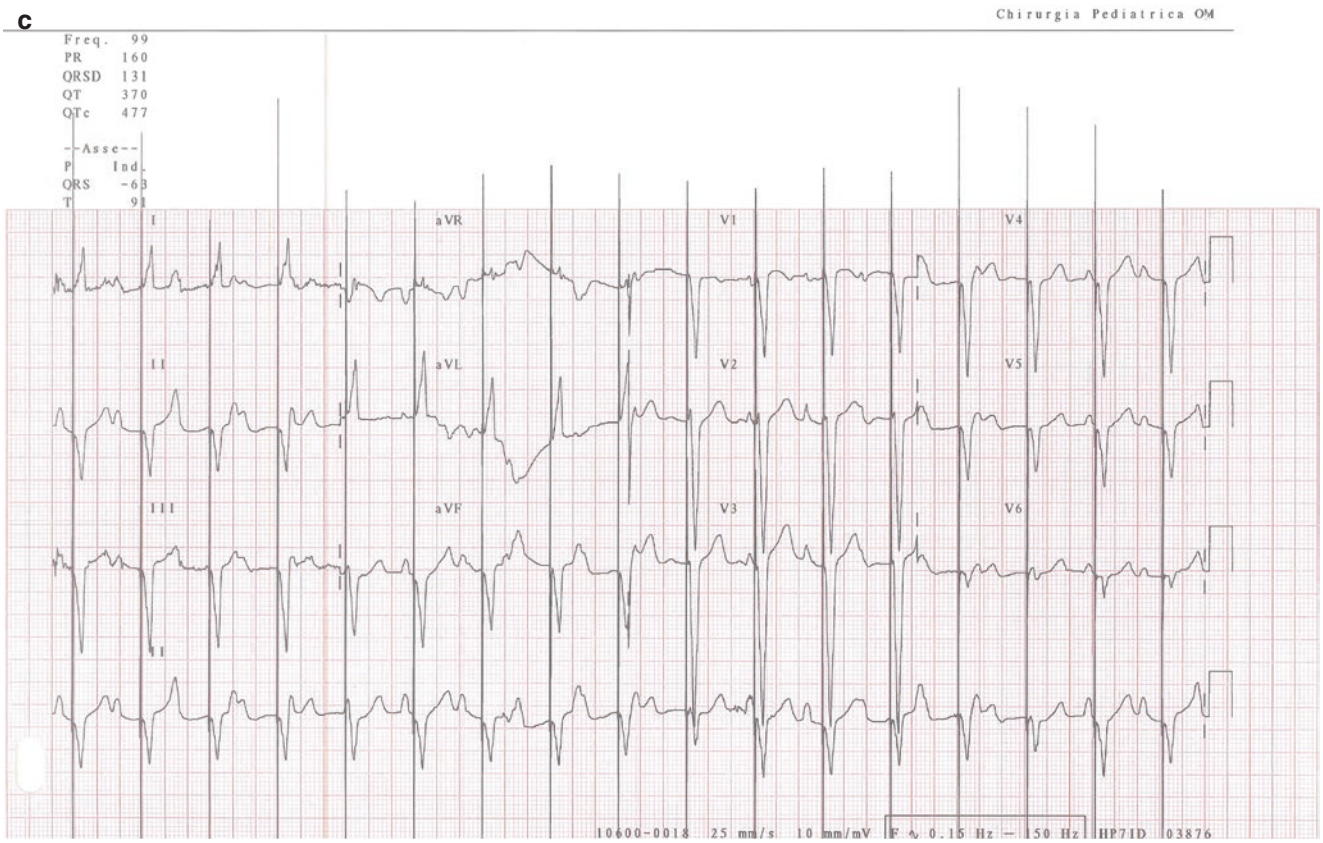


Fig. 8.36 Two-year-old female with CCTGA and AVB on VVI PM. She was admitted to the ER because of a pre-syncopal episode. The ECG was sent by fax. (**a**) The anticipated paced beat (*circle*) raised the suspicion of PM malfunction as sensing deficit, as a possible cause of inappropriate stimulation and potential life-threatening arrhythmias; (**b**) close-up. (**c**) The original ECG revealed the truth. (**d**) It was an artifact due to the fax scrolling defect that shortened the distance between two beats by a few millimeters, creating the false impression of PM malfunction . The error was confirmed from the header of the fax in which the words present the same scrolling issue



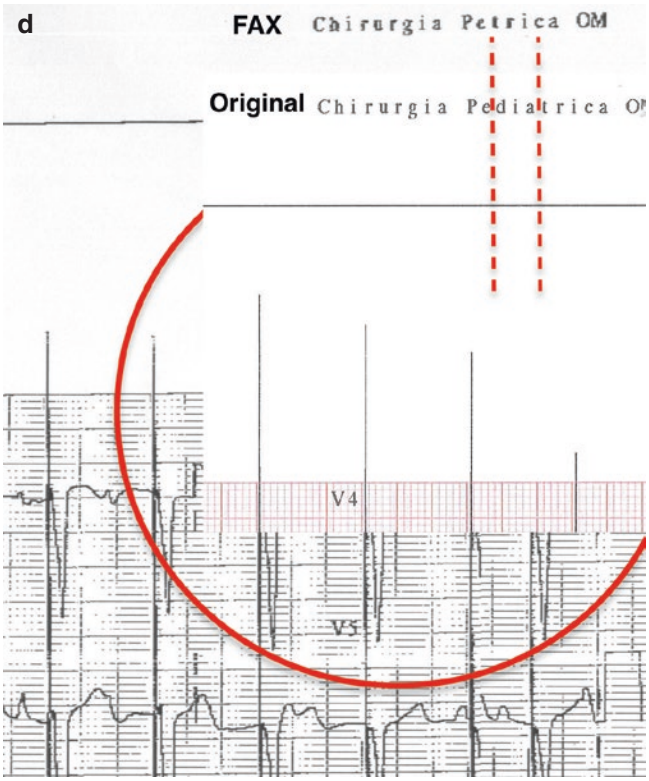


Fig. 8.36 (continued)

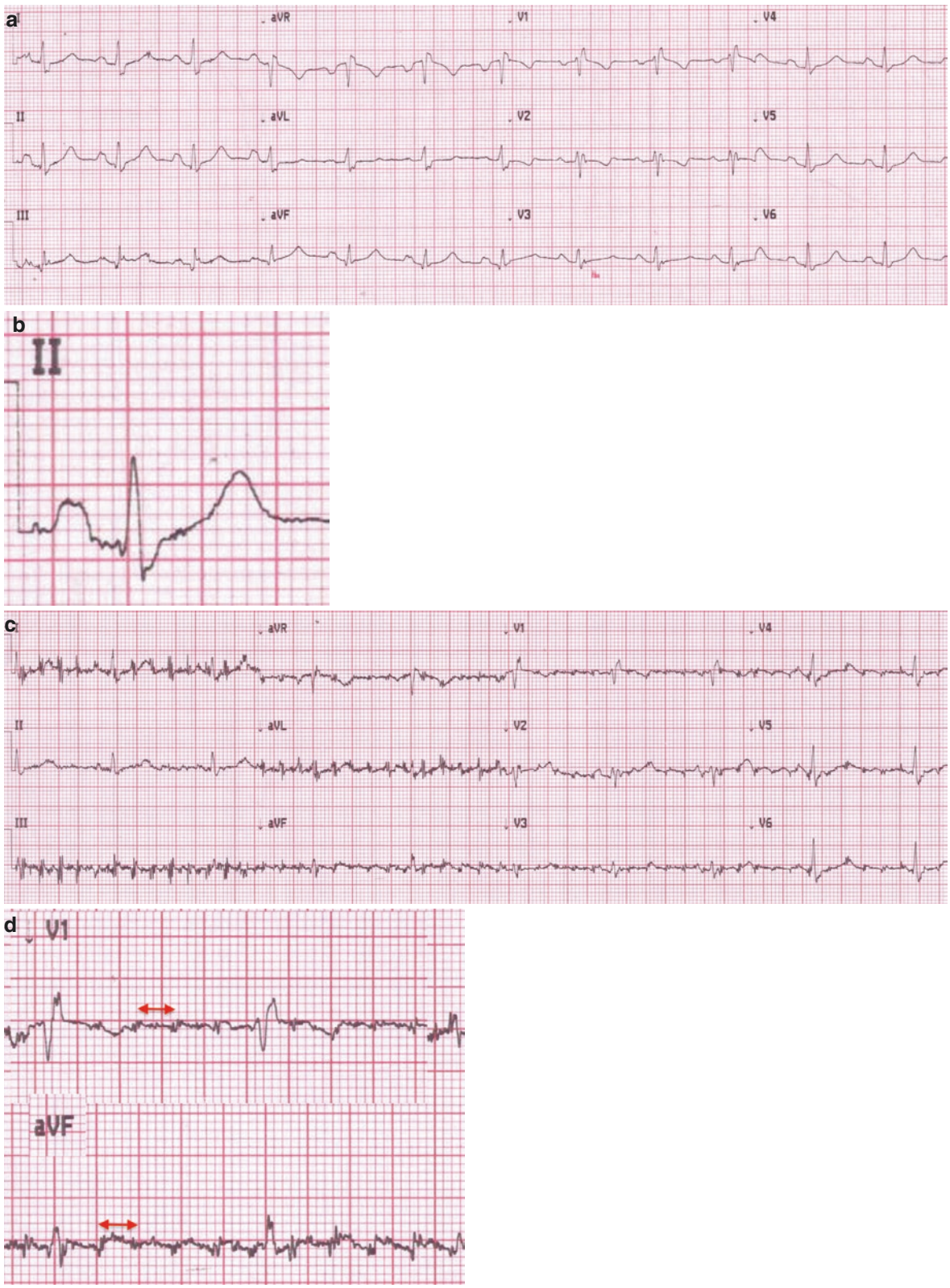


Fig. 8.37 (**a**) Fifty-year-old woman, carrier of a moderate form of EA symptomatic for palpitation on treatment with atenolol. (**b**) Close-up. Note that the P wave is larger than the QRS, consistently with the severely enlarged right atrium. (**c**) Because of severe depression, she started to take antidepressants. The ECG done to measure the QT interval raised the suspicion of atrial flutter, not uncommon in EA. (**d**) In fact, "atrial" waves have a rate of 300 bpm, while the ventricular waves are around 60 bpm, a ratio consistent with atrial flutter 5:1. Actually the "atrial" waves are artifacts due to tremor. The parkinsonism is a known side effect of neuroleptics and antidepressants

Suggested Reading

Oreto G, Luzza F, Donato A, Carbone V, Satullo G, Calabrò MP. L'elettrocardiogramma—un mosaico a 12 tessere. Centro Scientifico Editore: Torino; 2009.

Stanger P, Lister G, Silverman NH, Hoffman JI. Electrocardiograph monitor artifacts in a neonatal intensive care unit. Pediatrics. 1977;60:689–95.

Patel SI, Souter MJ. Equipment-related electrocardiographic artifacts: causes, characteristics, consequences, and correction. Anesthesiology. 2008;108:138–48.

Bradycardias and Conduction Disorders

9

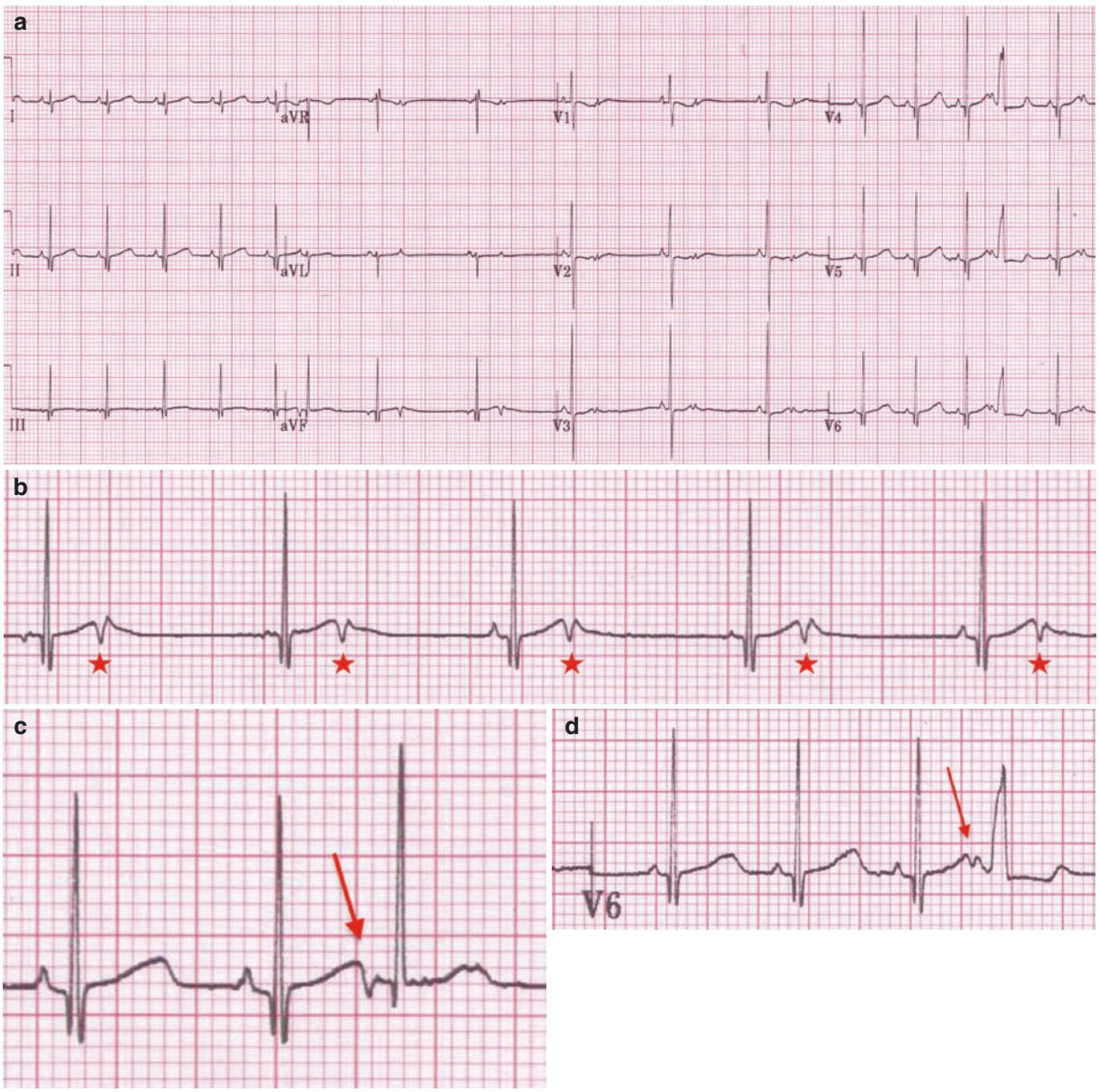


Fig. 9.1 (**a**) Neonate referred for bradycardia. (**b**) The cause of the bradycardia was ectopic atrial beats (*asterisks*). These beats are anticipated and then blocked. (**c**) Sometimes they are conducted, with narrow and (**d**) wide complex with LBBB aberrancy

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G. Bronzetti, *Atlas of Pediatric and Youth ECG*, https://doi.org/10.1007/978-3-319-57102-7_9

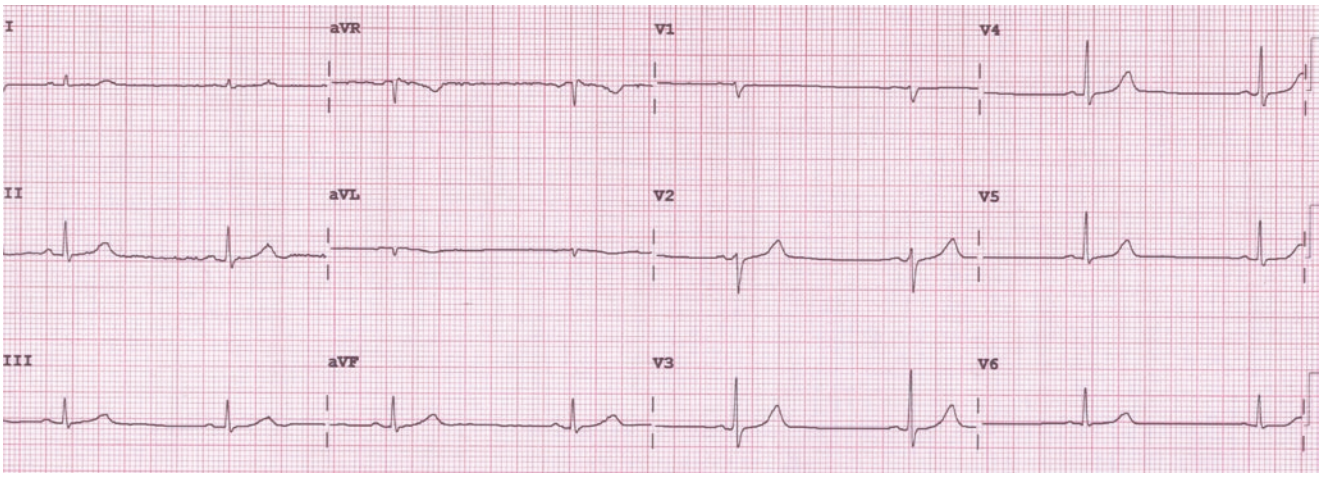

Fig. 9.2 Fourteen-year-old female with anorexia. Due to the shortage of food, the body a sort of lethargy, witnessed also by the low amplitude of the QRS

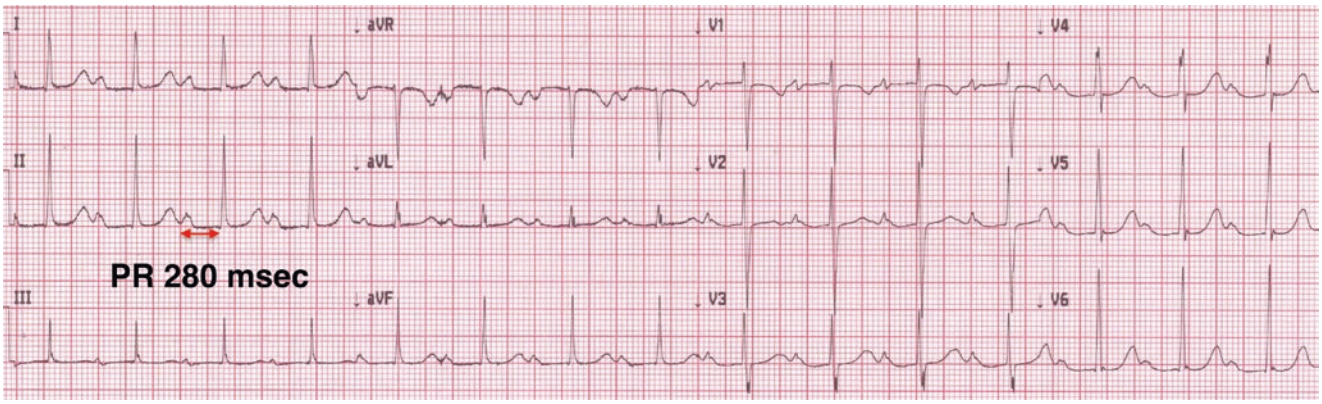

Fig. 9.3 Eight-year-old male with acute rheumatic fever complicated by mitral regurgitation and first-degree AV block

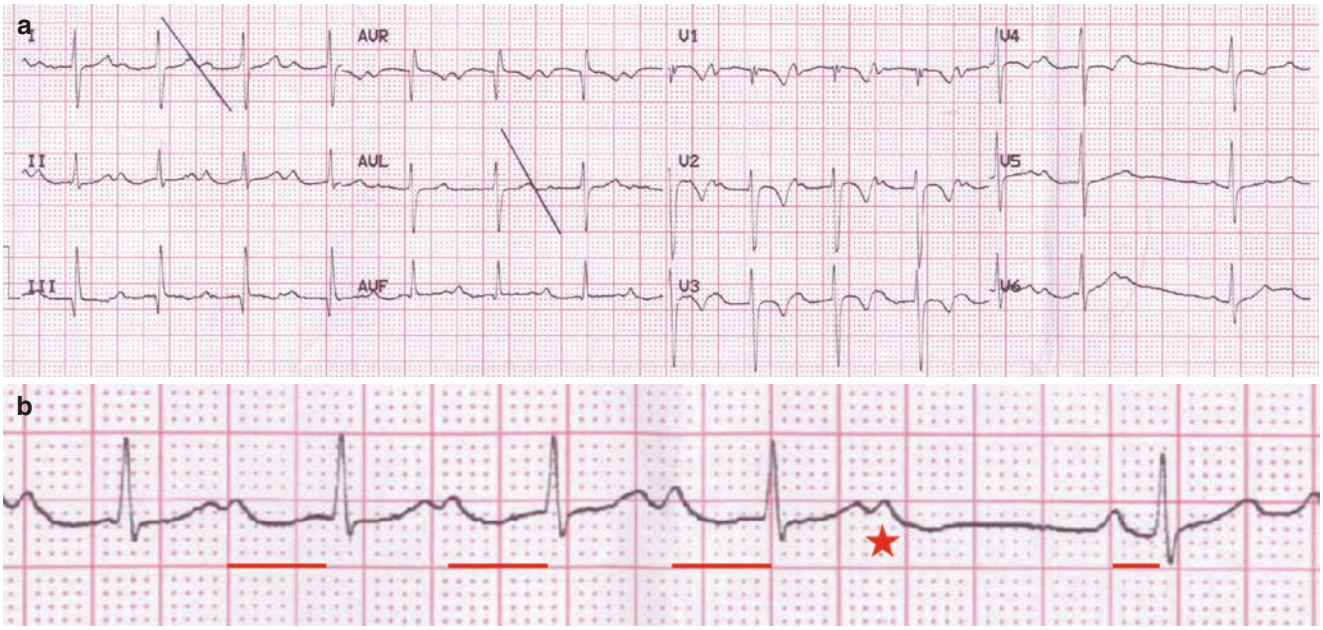

Fig. 9.4 (**a**) Two-year-old girl. Idiopathic second-degree AV block Mobitz 1 (Luciani-Wenckebach). (**b**) Close-up

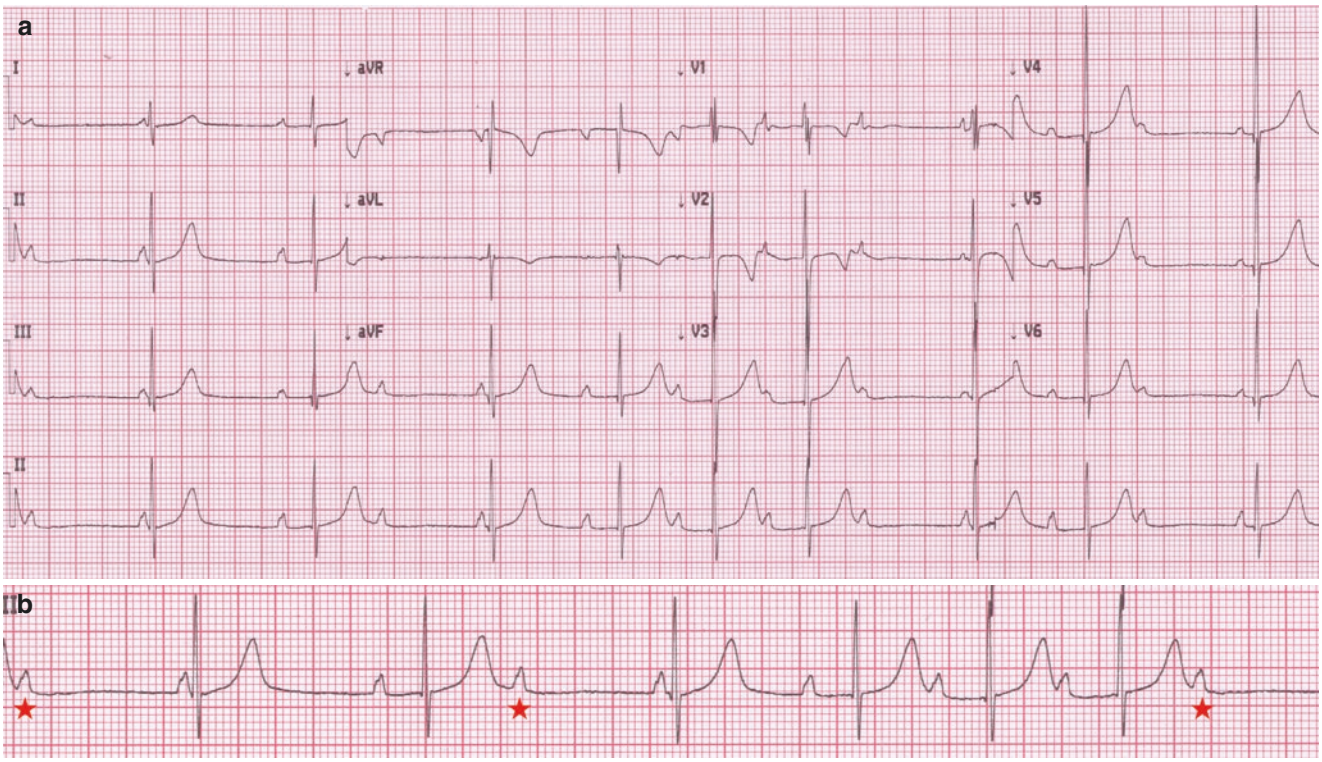

Fig. 9.5 (**a**) As in the previous case, a second-degree AV block Mobitz 1 (Luciani-Wenckebach) can be noted in a 4-year-old male. (**b**) Close-up

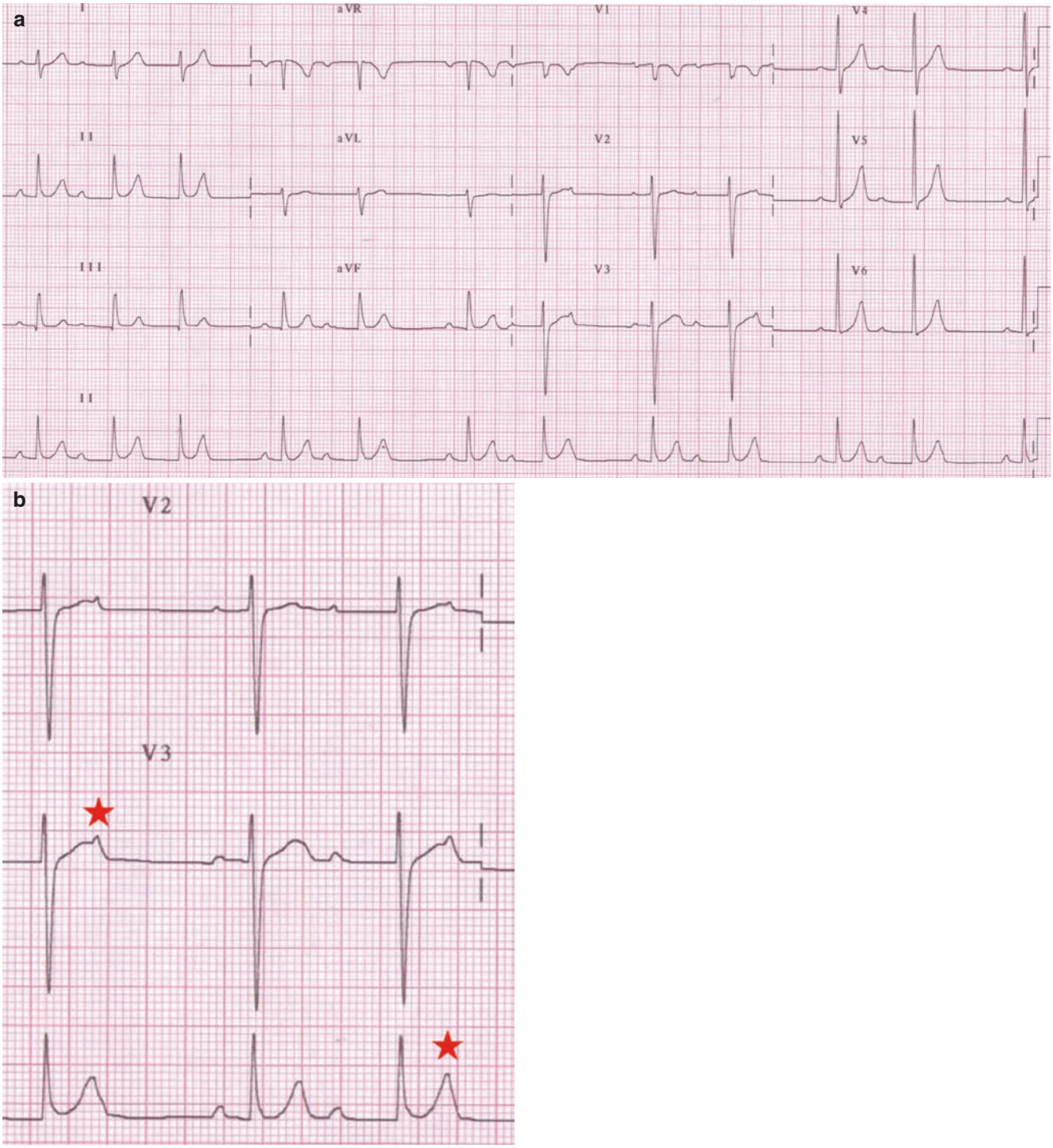

Fig. 9.6 (**a**) Fourteen-year-old boy; see the previous case. (**b**) Close-up (*asterisk* indicates the nonconducted P waves)

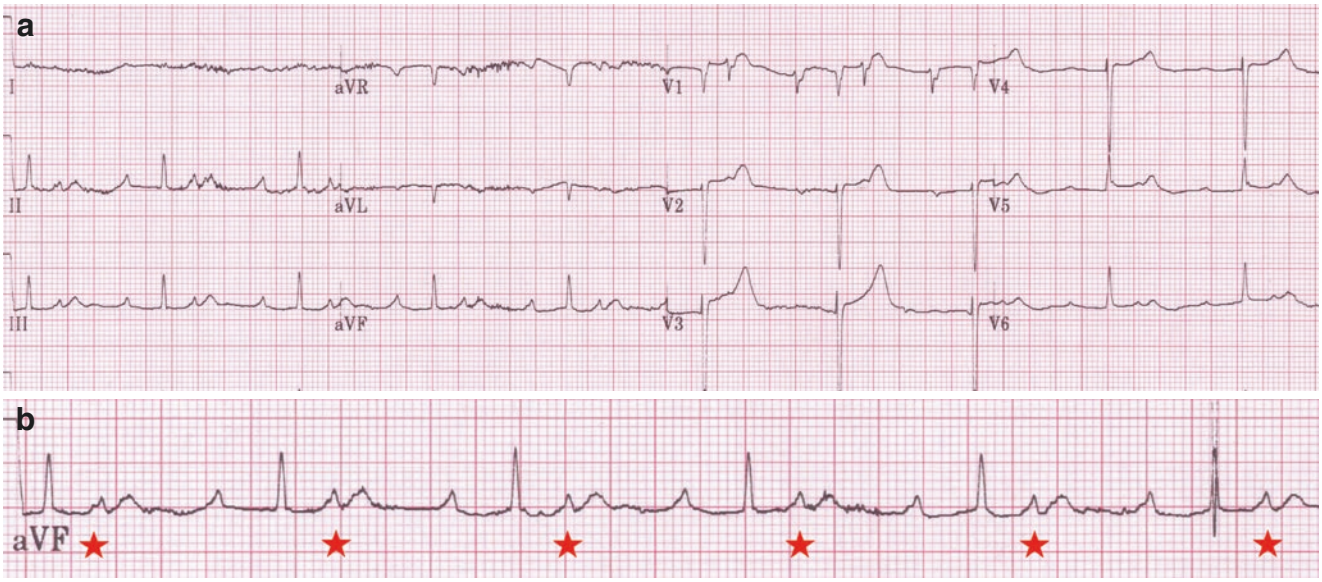

Fig. 9.7 (**a**) Twenty-year-old female, postresection of muscular subaortic stenosis (Morrow operation). She developed a second-degree AV block Mobitz 2. (**b**) Close-up. During isoprenaline induced sinus tachycardia

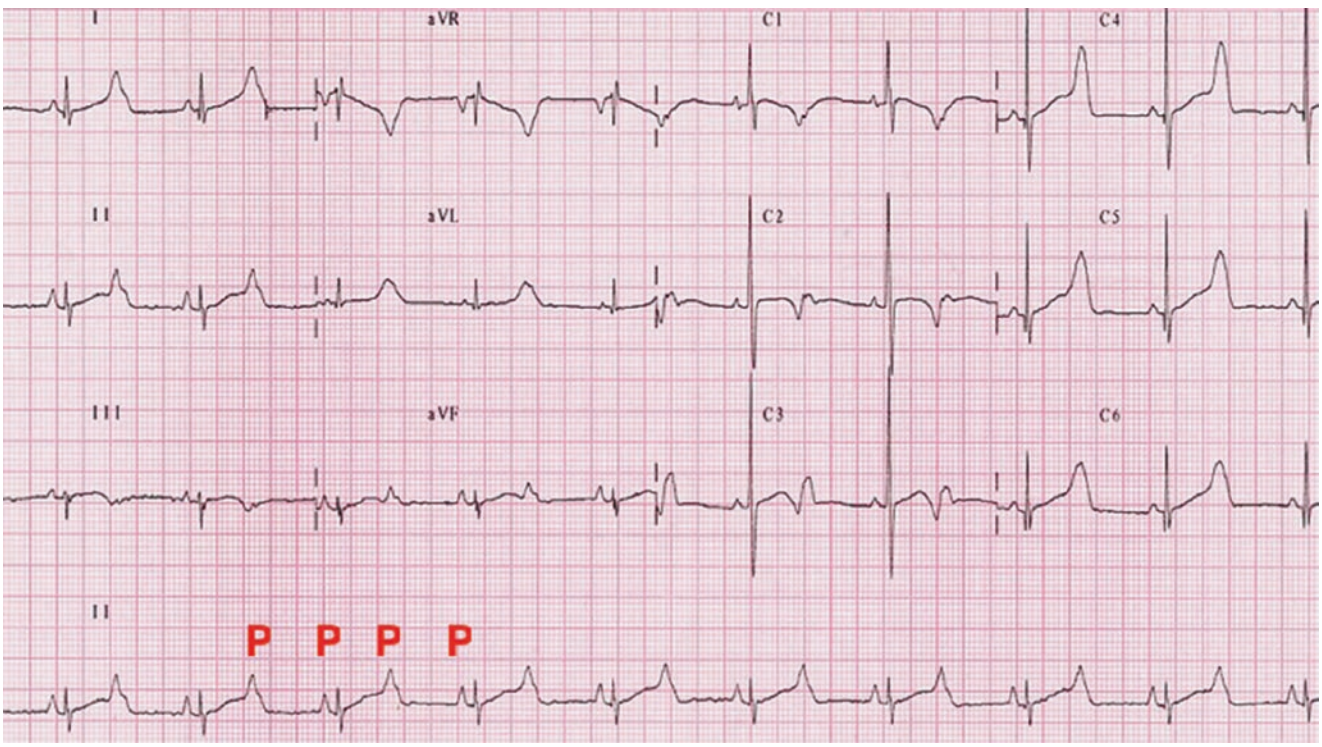

Fig. 9.8 Six-month-old with 2:1 AV block. The patient is affected by LQTS 3, and in this setting the 2:1 AV block is a form of functional block (the AV node is intact)

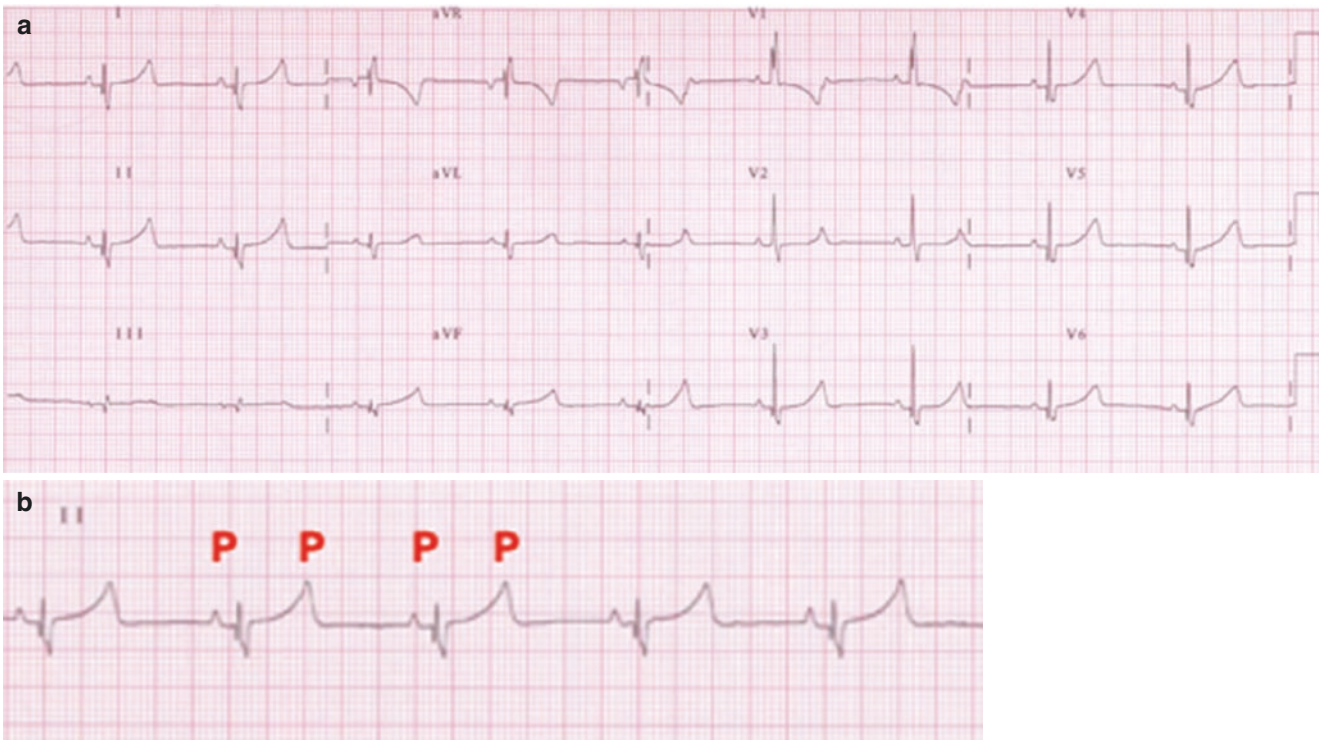

Fig. 9.9 (**a**) Neonate with 2:1 AV block, mildly prolonged QT; the genetic testing for LQTS was negative. (**b**) Close-up. Note the ventriculophasic dissociation

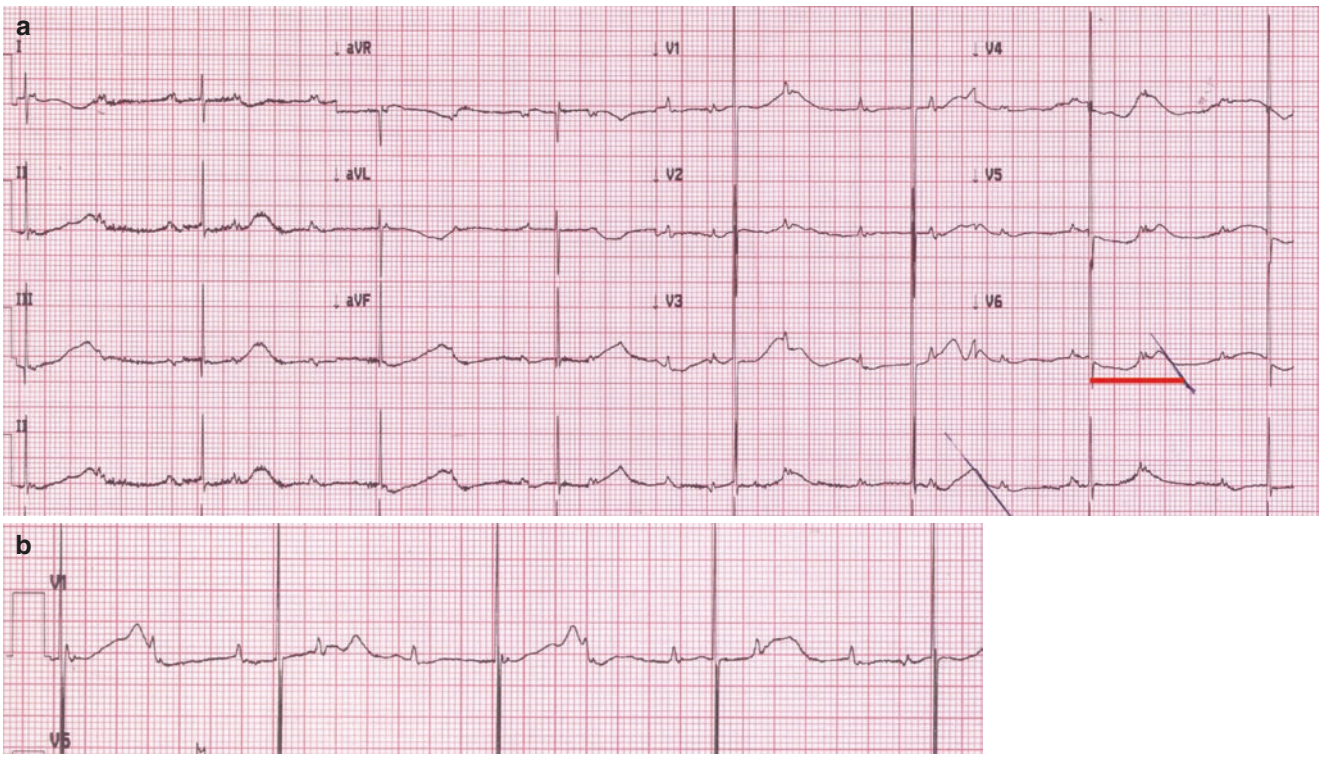

Fig. 9.10 (**a**) Neonate with complete AVB. There is an example of bradycardia-dependent QT prolongation. The patient was implanted and the QT returned to normal. (**b**) Close-up

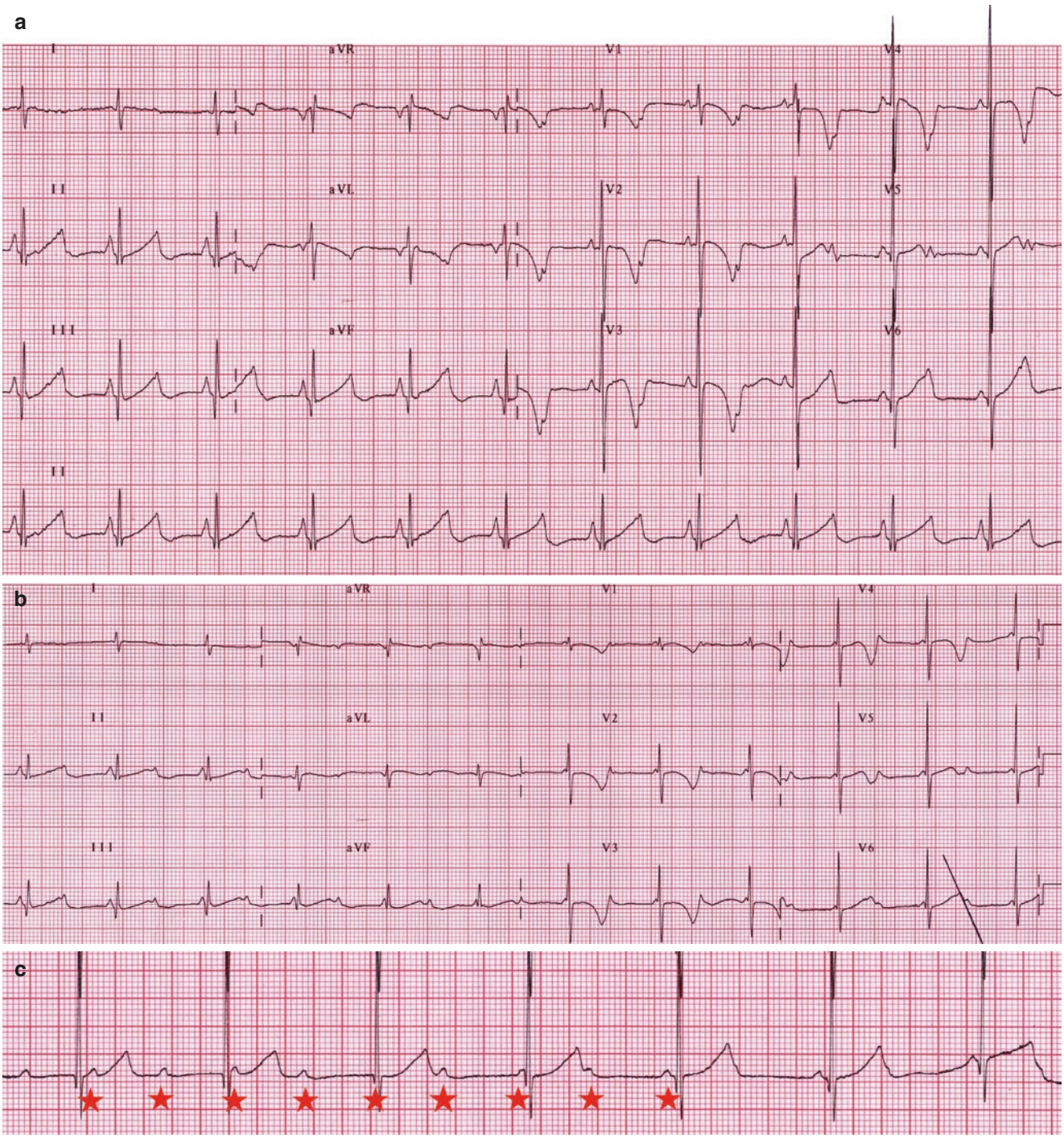

Fig. 9.11 Two-year-old boy with bradycardia. (**a**) The first ECG seems just to show sinus rhythm, but in the second one (**b**) the P waves are clearly dissociated from the QRS. (**c**) Close-up. A short strip can fail in recognizing a complete AV block, as in this tracing where one P is apparently in a correct position in front of the QRS, and the other is buried in the T waves. Deep and wide T waves are common in complete AVB

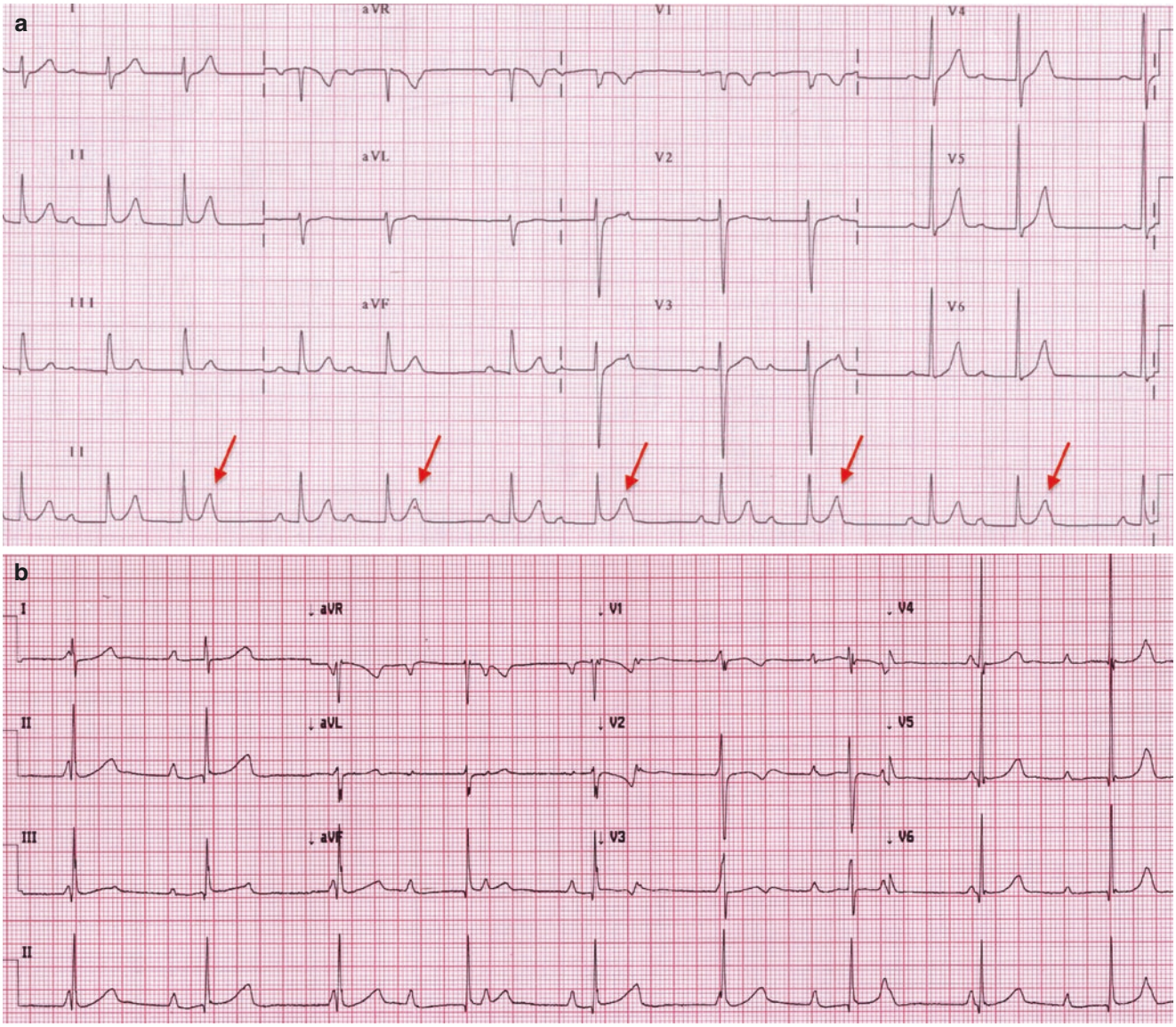

Fig. 9.12 (**a**) Six-year-old female with second-degree AVB Luciani-Wenckebach type (progressive prolongation of PR interval up to a "blocked" P wave), evolved in 2 years into a complete AVB; *arrows* indicate the nonconducted P wave. (**b**) Complete AVB

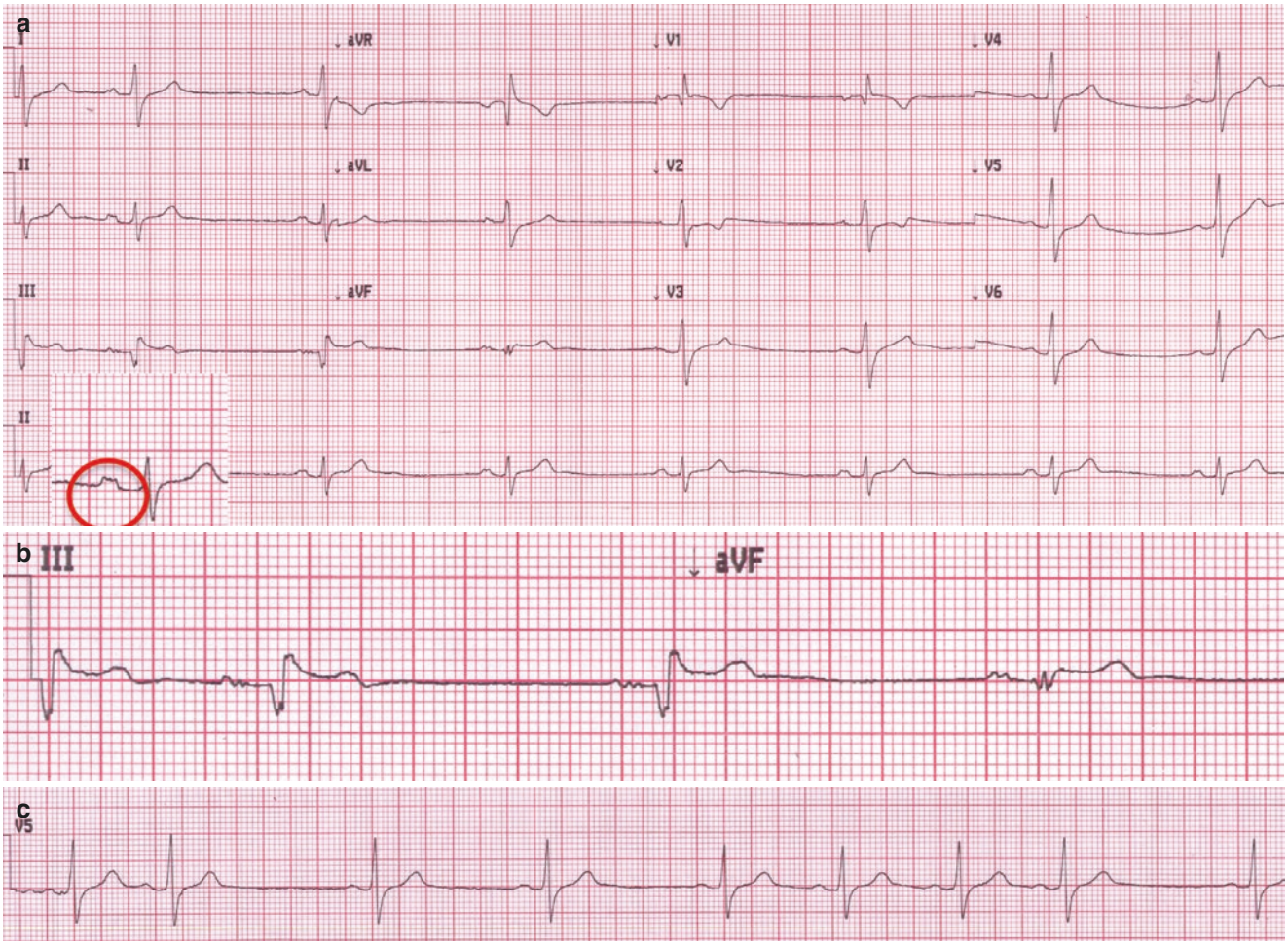

Fig. 9.13 (**a**) Ten-year-old female with Brugada syndrome (SCN5A mutation). Note the BS traits are not merely in the right precordial leads where they are usually expected, but in the inferior leads (lead III and aVF). Note also a reciprocal—and sometimes independent from R wave in V1—S wave in V6. (**b**, **c**) Close-up. This case like others underscores that the presence of a large S wave in V6 is not reassuring and rather in some cases it strengthens the suspicion of BS. Long PR and sinoatrial block are common in BS

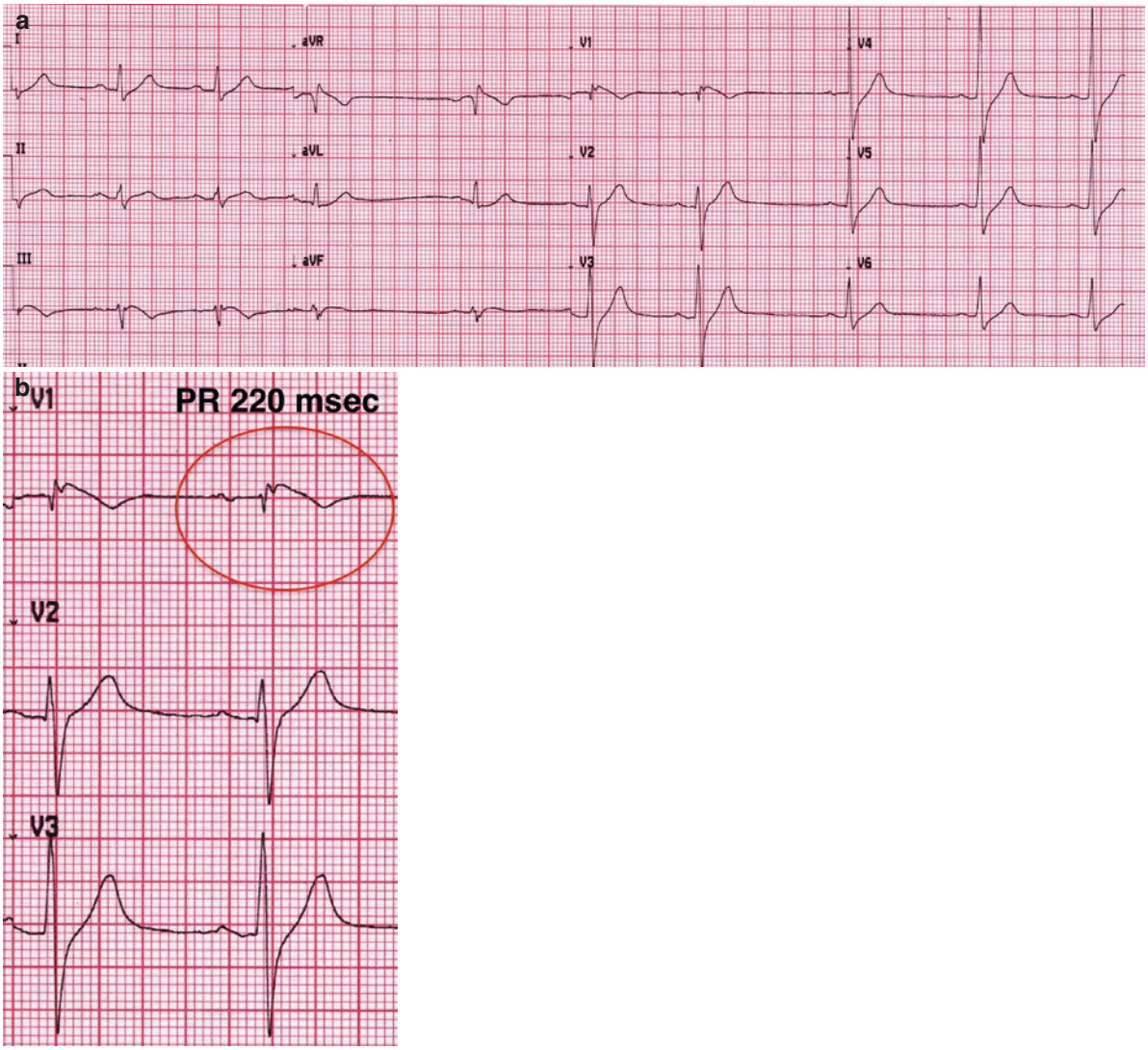

Fig. 9.14 (**a**) BS in a 9-year-old boy. (**b**) Along the BS features in the right precordial leads, a long PR can be noted (PR 220 ms)

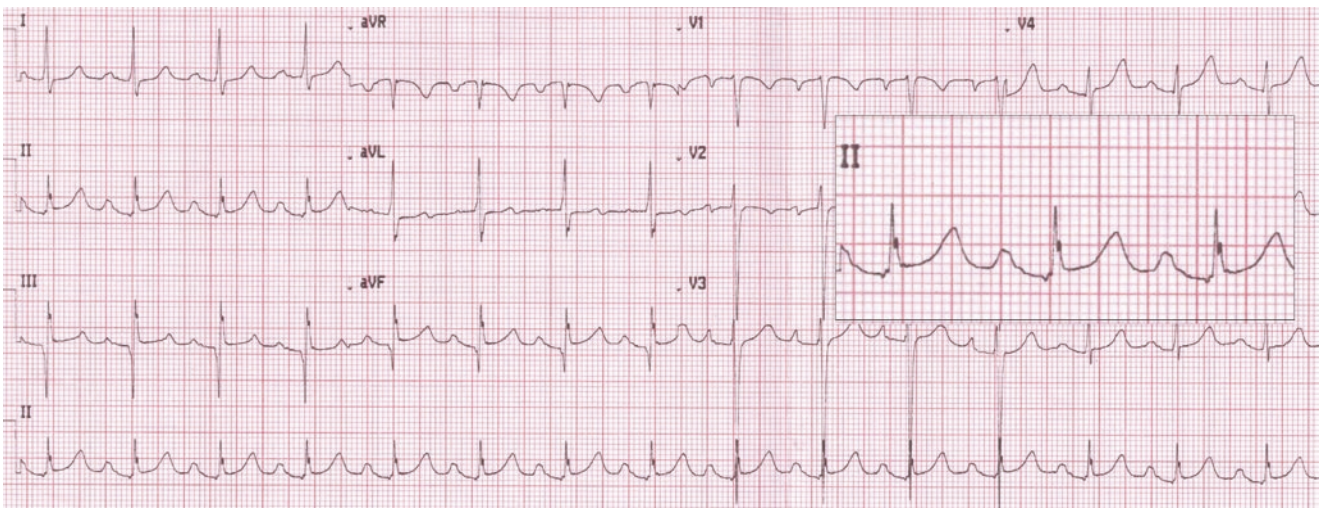

Fig. 9.15 Fifteen-year-old female CCTGA. A first-degree AVB is present (PR 230 ms). CCTGA is susceptible to AV block (see text), either congenital or later in adulthood. Other CHD are exposed to post-operative conduction disturbances, as, for instance, Fontan, Mustard, or Senning operation

Suggested Reading

Murray LE, Smith AH, Flack EC, Crum K, Owen J, Kannankeril PJ. Genotypic and phenotypic predictors of complete heart block and recovery of conduction after surgical repair of congenital heart disease. Heart Rhythm. 2016;14(3):402–9. doi:10.1016/j.hrthm.2016.11.010.

Brugada J, Blom N, Sarquella-Brugada G, et al. European Heart Rhythm Association; Association for European Paediatric and Congenital Cardiology. Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement. Europace. 2013;15(9):1337–82. doi:10.1093/europace/eut082.

Baruteau AE, Pass RH, Thambo JB, Behaghel A, Le Pennec S, Perdreau E, Combes N, Liberman L, McLeod CJ. Congenital and childhood atrioventricular blocks: pathophysiology and contemporary management. Eur J Pediatr. 2016;175:1235–48.

Pre-excitation Syndromes

10

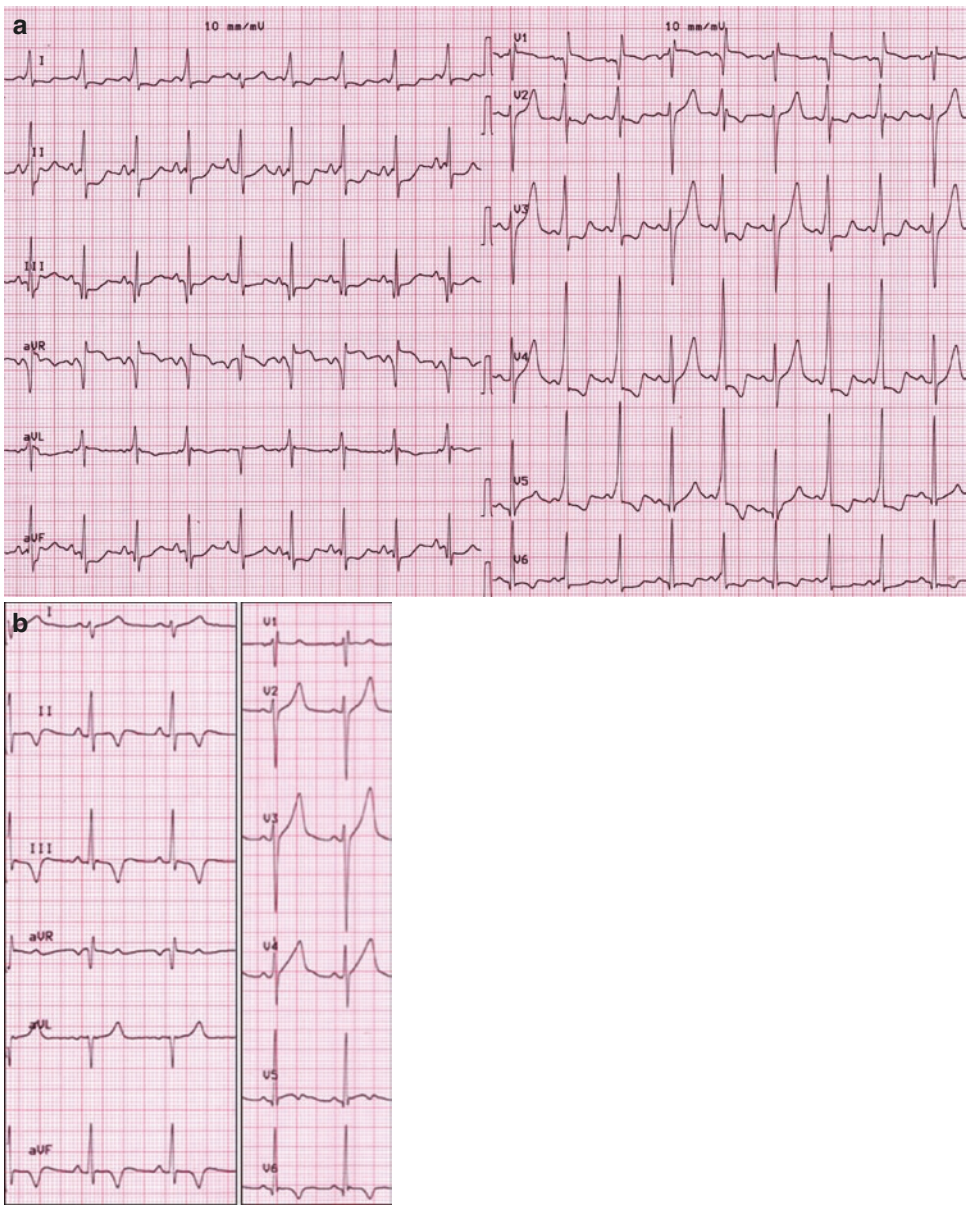

Fig. 10.1 (**a**) Twelve-year-old male, asymptomatic, diagnosed as "WPW" during sports participation screening. There is an intermittent pre-excitation. (**b**) In the upright position, the slight increase in HR abolishes the pre-excitation (due to a right posterior AP), denoting a bypass tract with long refractory period and therefore little risk of life-threatening arrhythmias (see atrial fibrillation conducted along the pathway with high ventricular rate); all leads are delta-free. The negative T waves in the inferior and lateral leads are an "electric memory" phenomenon. The stress test was done for arrhythmic risk stratification and not for ischemia. It is well recognized that pre-excitation may affect the repolarization making the assessment of ischemic changes of the ST segment

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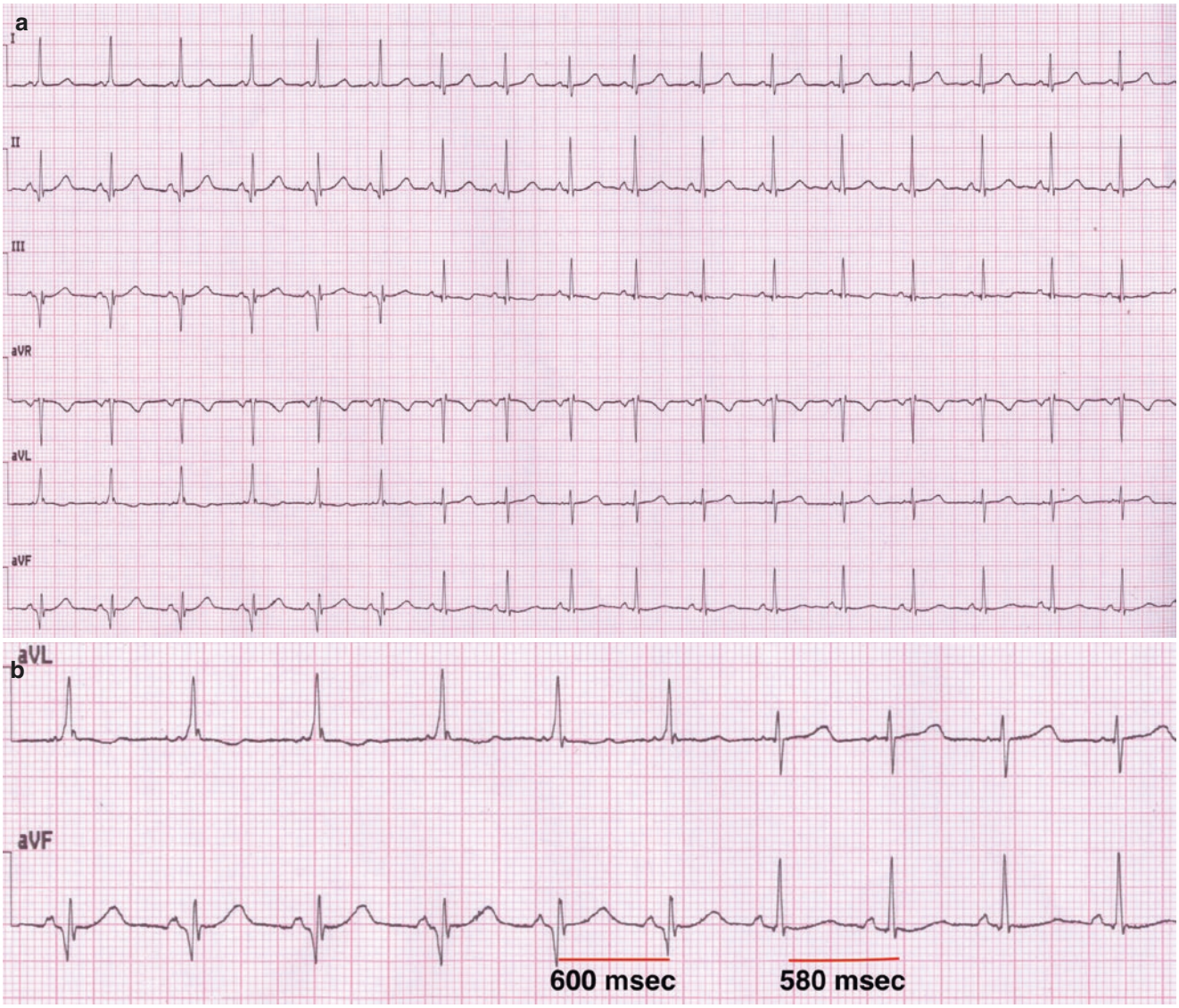

Fig. 10.2 (**a**) Another example of intermittent pre-excitation (IP) in a 13-year-old boy. The pre-excitation is HR dependent. In IP the delta wave usually disappears at faster HR (shorter R–R cycle), a behavior that on the conduction properties of the accessory pathways (AP) regarding the risk of life-threatening arrhythmias. (**b**) Close-up

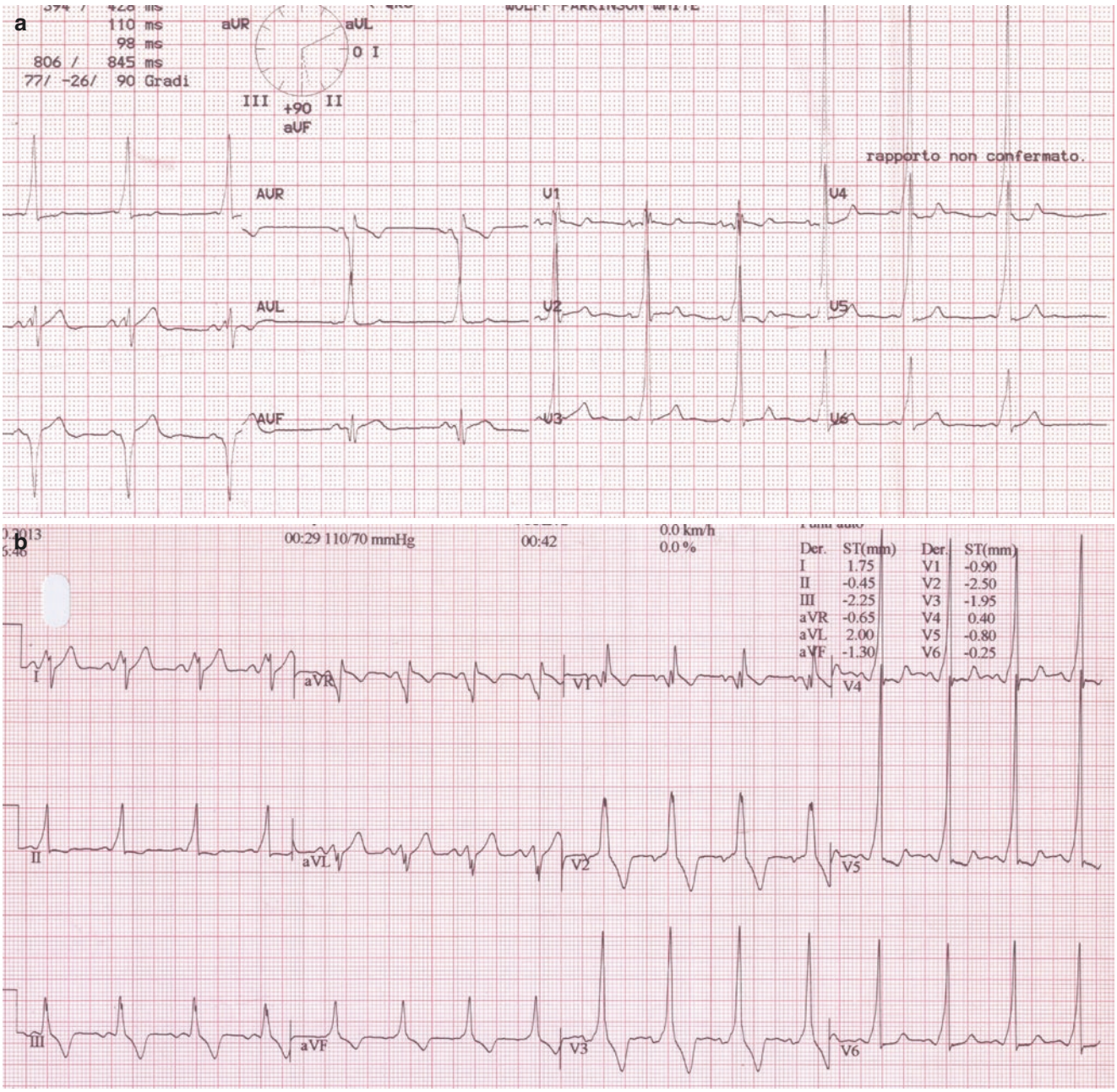

Fig. 10.3 Thirteen-year-old male with multiple APs proven during the endocavitary electrophysiological study (EPS). (**a**) Right inferoposterior AP. (**b**) Right anteroseptal AP

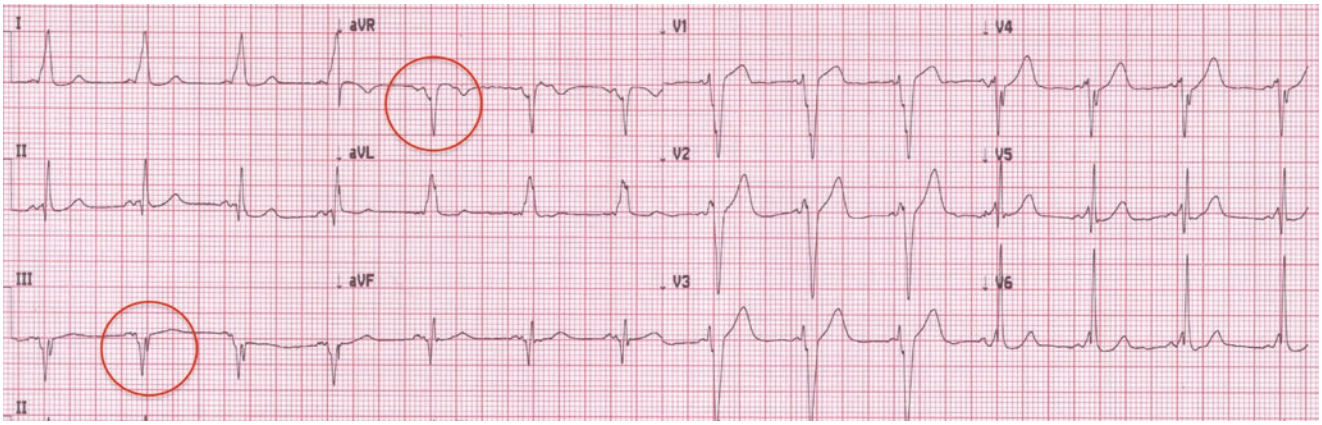

Fig. 10.4 Sixteen-year-old female with right lateral AP who underwent radiofrequency catheter ablation (RFCA). Delta is negative in leads III and aVR. The vector of AP runs away from *right* to *left*. In lead I and in the precordial leads, the pattern is LBBB-like, consistent with the AP site

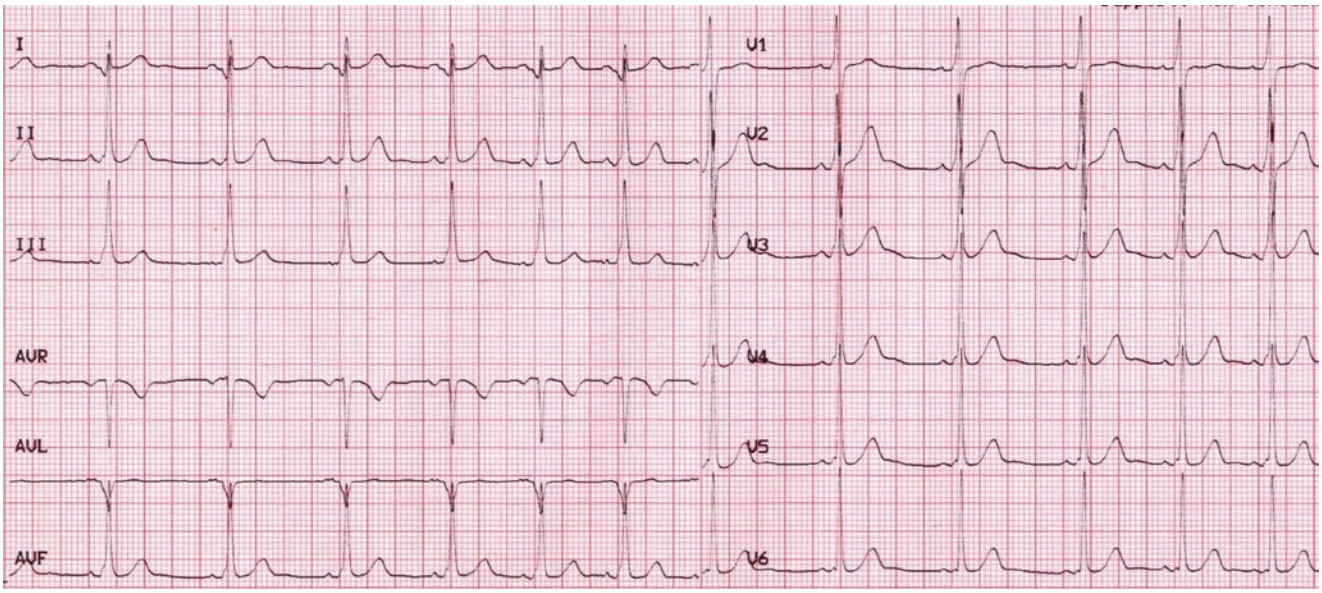

Fig. 10.5 Ten-year-old male with left lateral AP, the opposite site to the previous case. Here too, the ECG is fairly coherent, showing a negative delta wave in aVL and lead I, due to the vectors that fly off from *left* to *right*. Among APs, the left-sided APs are more common

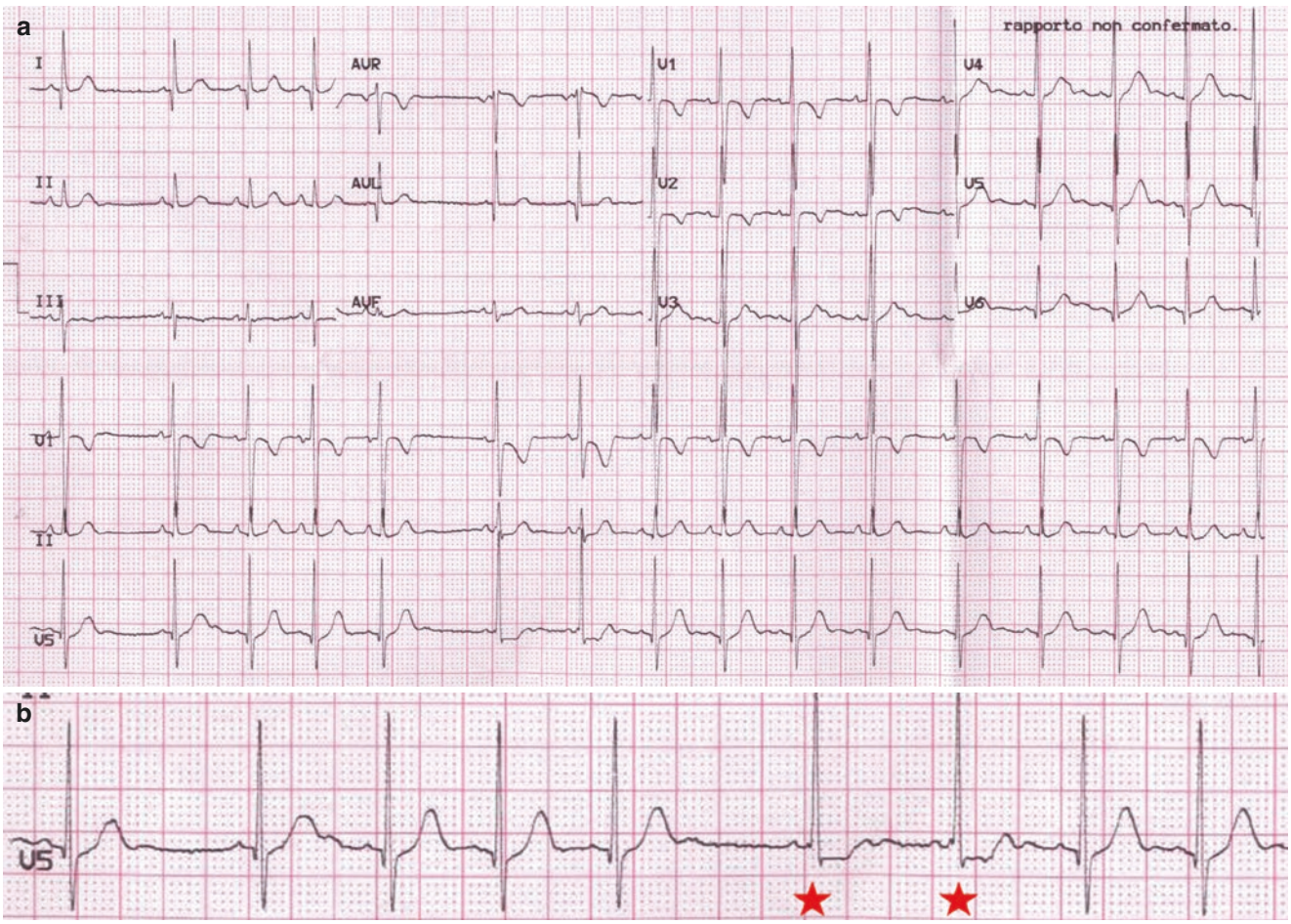

Fig. 10.6 (**a**) Another case of IP in a 6-year-old female. (**b**) Close-up: "just two beats of" pre-excitation

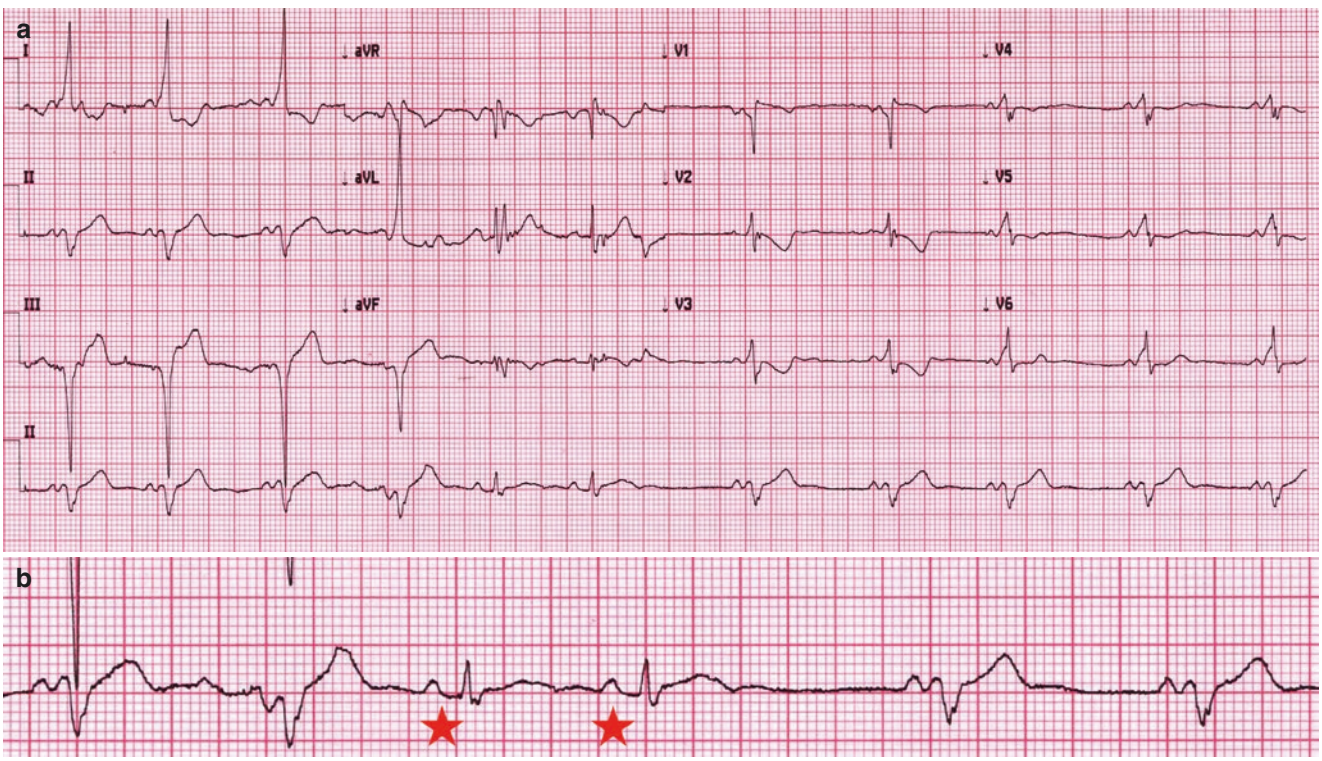

Fig. 10.7 (**a**) Again an IP in a 10-year-old male. (**b**) Close-up: this time, "just two beats off " pre-excitation

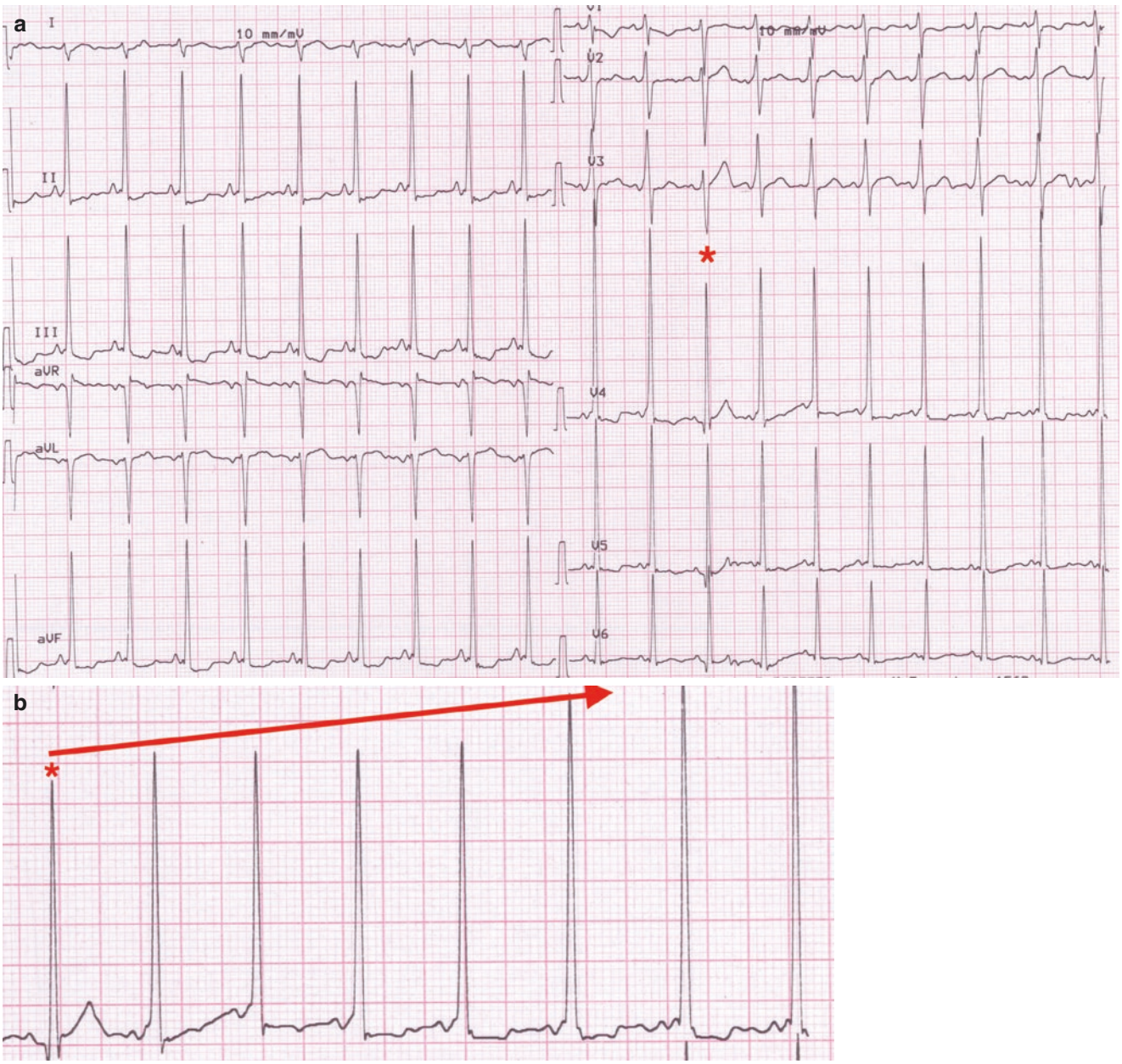

Fig. 10.8 (**a**) Eight-year-old male with incidental finding of intermittent pre-excitation due to a right postero-septal AP. Almost all QRS are pre-excited (the one with asterisk is not pre-excited), but the morphology varies with the degree of pre-excitation, which in turn depends on the cycle length (P–P interval or delta-delta!). There is a sort of "concertina effect." (**b**) Close-up

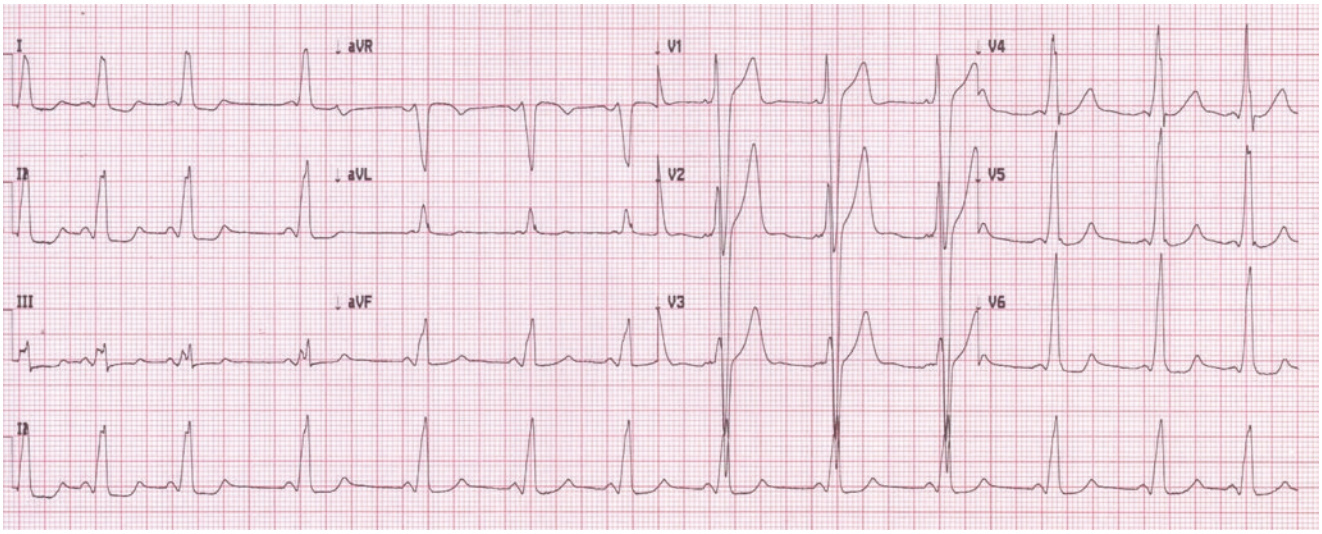

Fig. 10.9 Thirteen-year-old female. Right anteroseptal AP

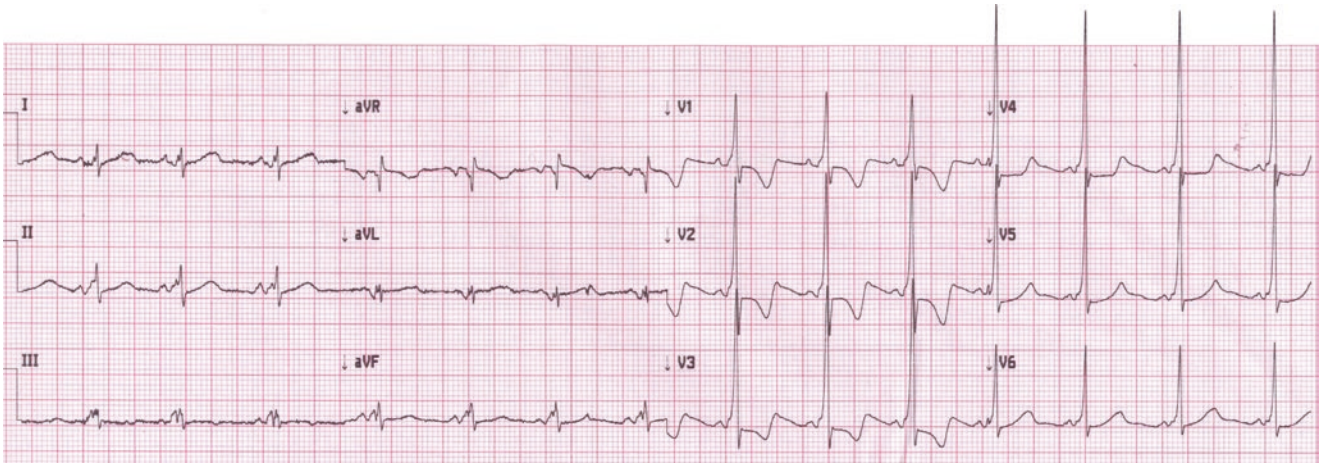

Fig. 10.10 Eleven-year-old male. Left lateral AP

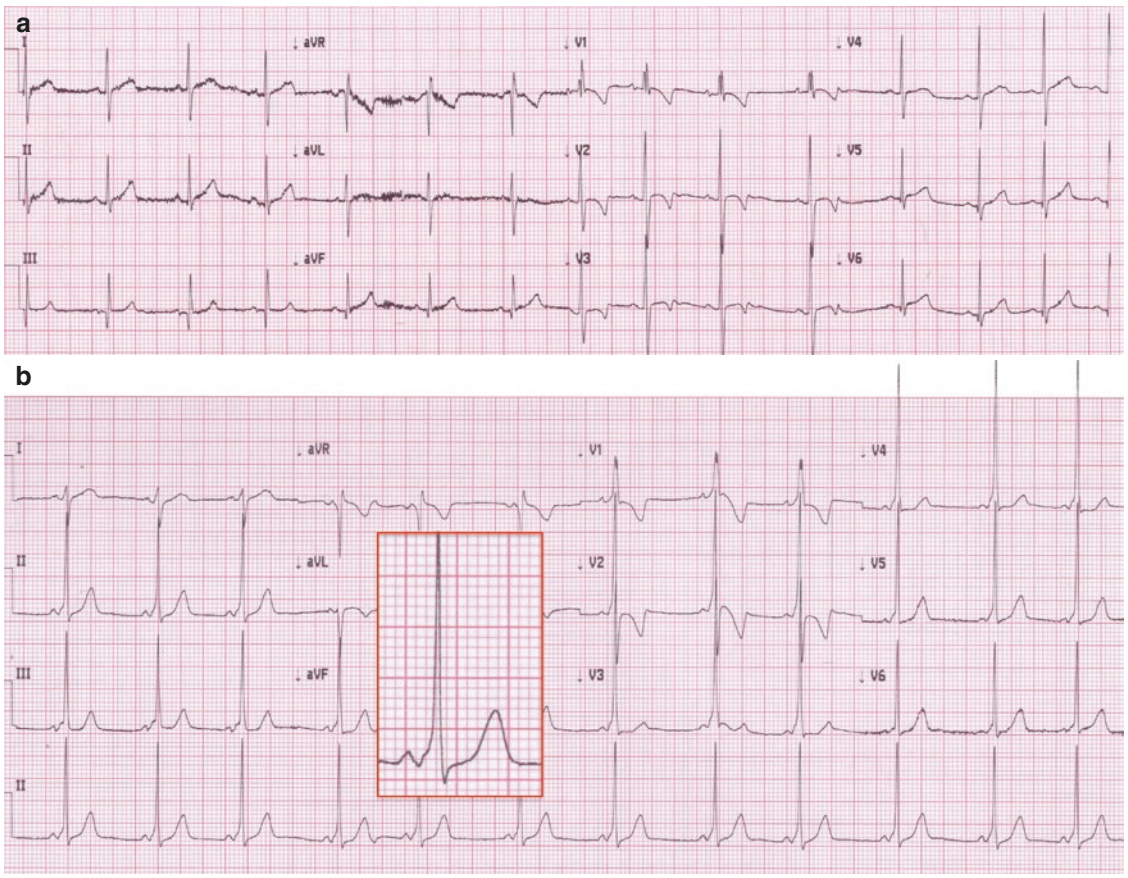

Fig. 10.11 A case of neonatal SVT. (**a**) In the first year of life, he was symptomatic but not pre-excited. (**b**) At 2 years of life he was pre-excited but not symptomatic. This means that in the first year of life the AP was able to conduct only retrogradely; in other words it was occult or concealed

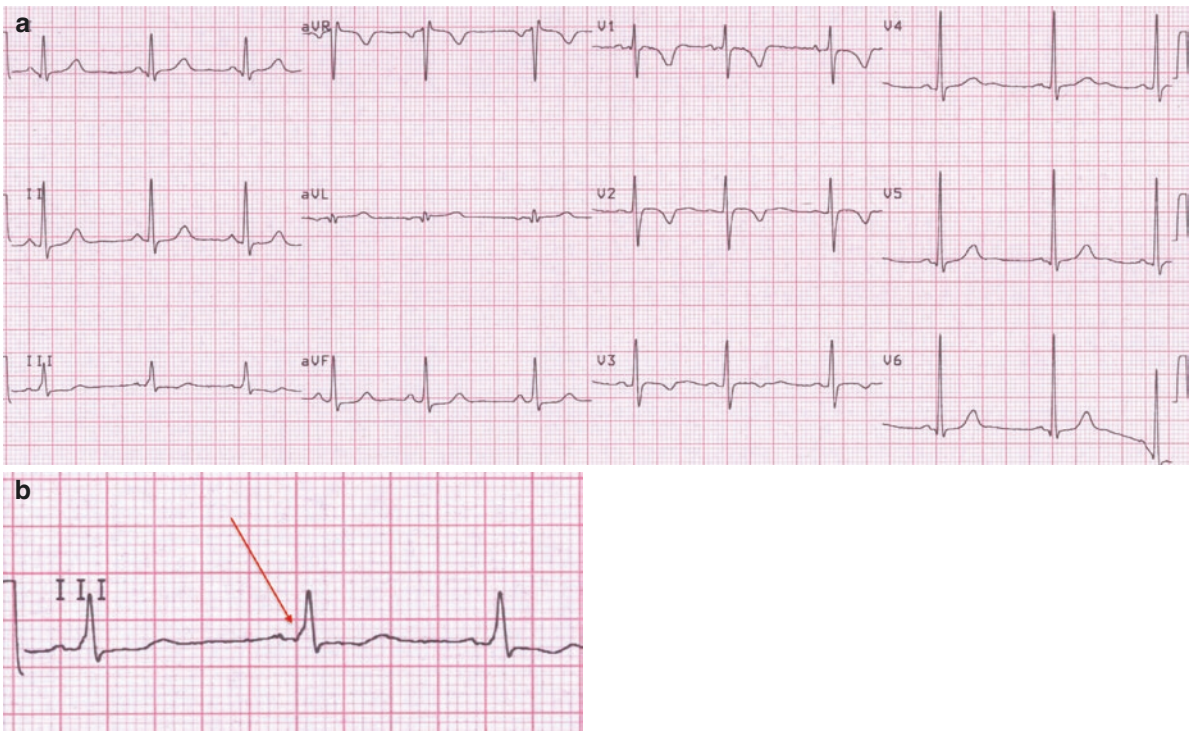

Fig. 10.12 (**a**) Ten-year-old male. The R wave in lead III is slurred, raising the suspicion of pre-excitation. The remaining leads are normal and PR interval is reassuring. (**b**) Close-up. It is a pseudo-delta: one swallow does not make a summer

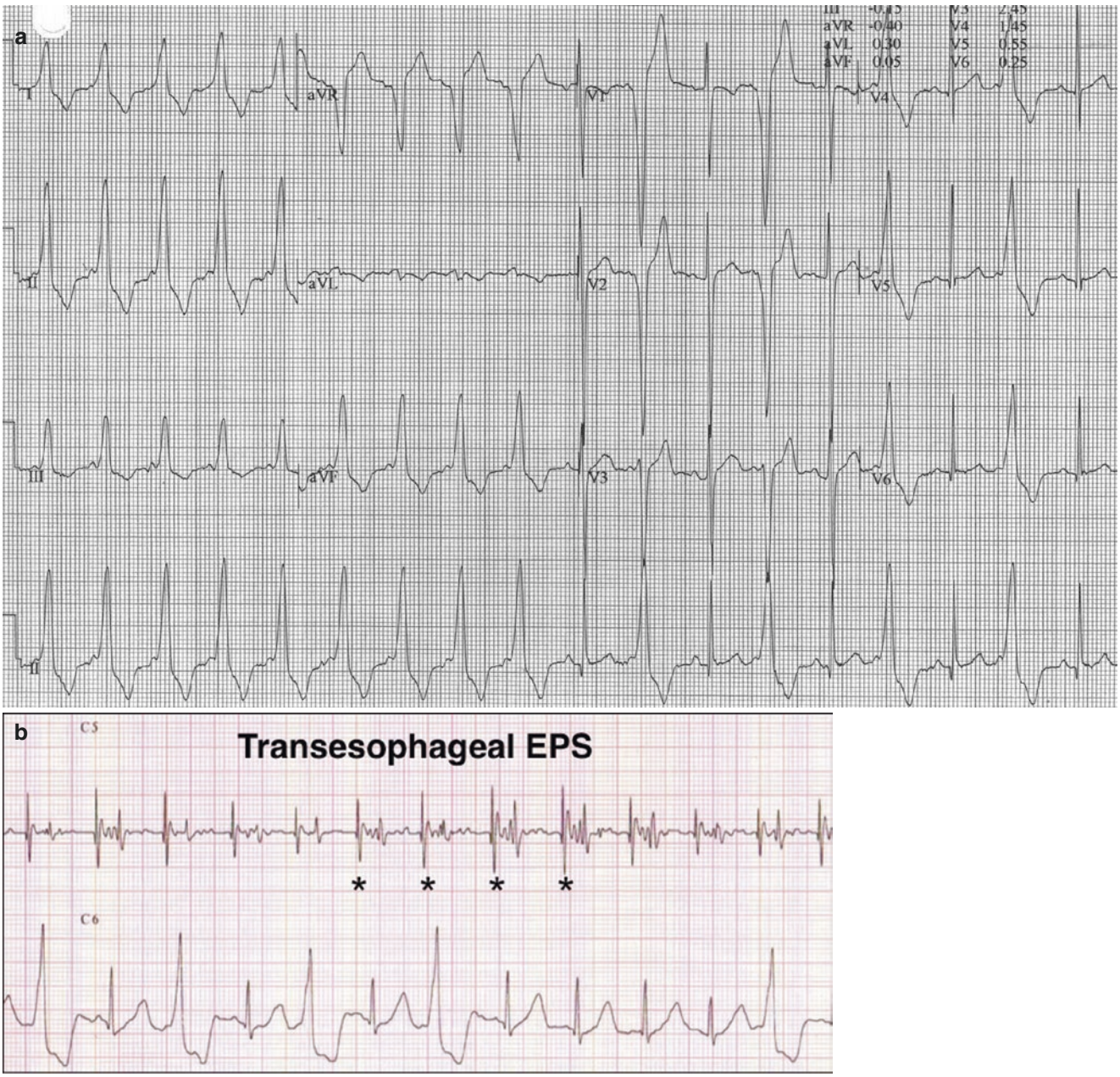

Fig. 10.13 Twelve-year-old male referred for RVOT. (**a**) The tracing shows wide complexes with LBBB morphology and vertical axis, suggestive of right infundibular origin (RVOT VT). (**b**) The transesophageal electrophysiologic study revealed ventricular pre-excitation: all the ventricular complexes, both narrow and wide, are preceded by atrial deflection. The P wave in the surface ECG is fused with the R wave, while in the esophageal tracing it can be clearly distinguished from the ventricular wave which it constantly precedes (*asterisk*). The intermittent pre-excitation implies a long refractory period of the accessory pathway and represents a low risk of major arrhythmias like pre-excited atrial fibrillation with possible degeneration into ventricular fibrillation

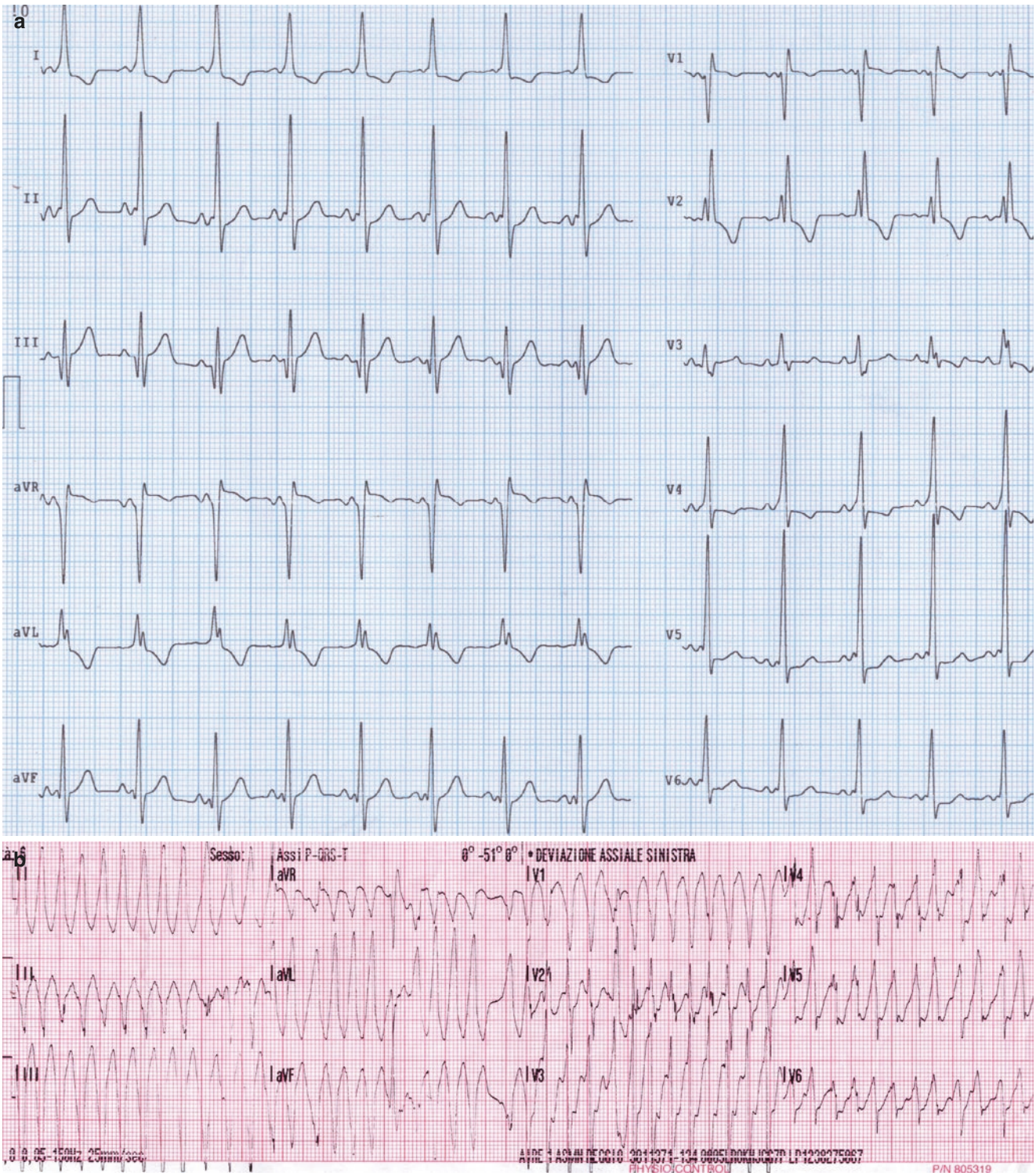

Fig. 10.14 (**a**) Eight-year-old male with Ebstein's anomaly (EA) with right posterior AP. (**b**) Episode of AF conducted to the ventricle by the AP. In EA it is not unusual to find multiple APs

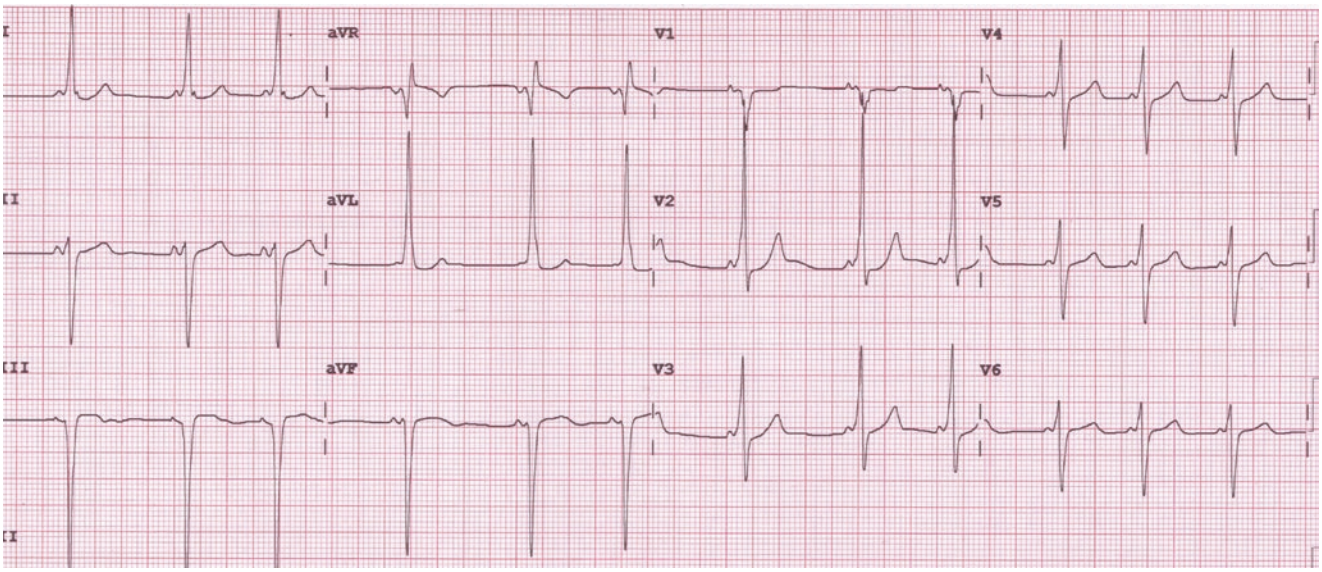

Fig. 10.15 Eleven-year-old male. Left postero-septal AP

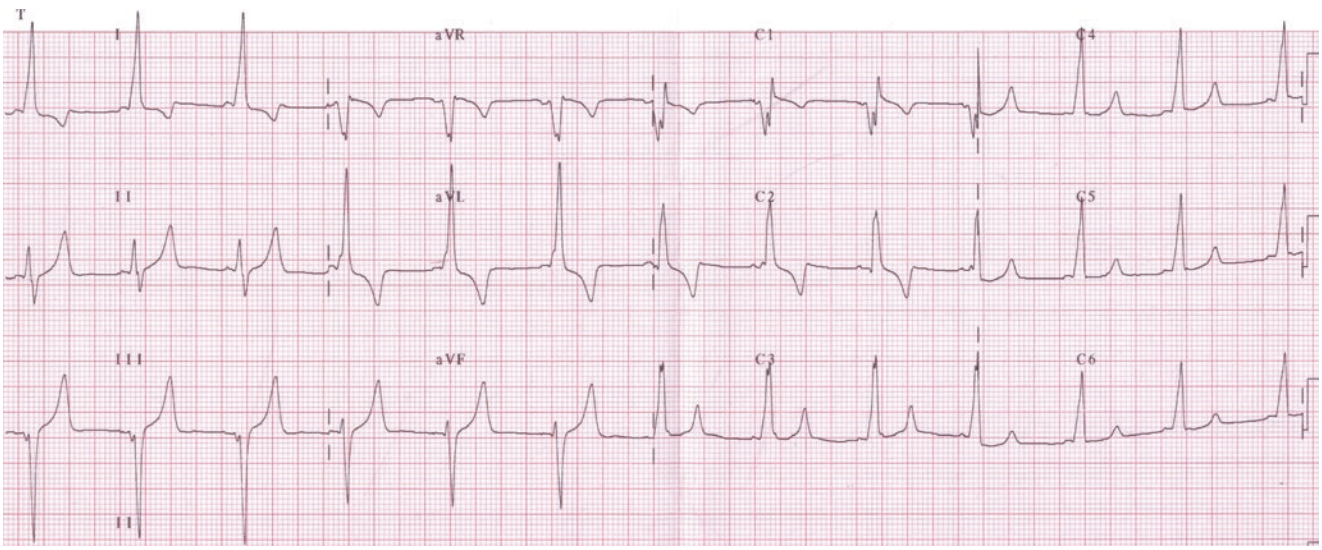

Fig. 10.16 Ten-year-old female. Right postero-septal AP

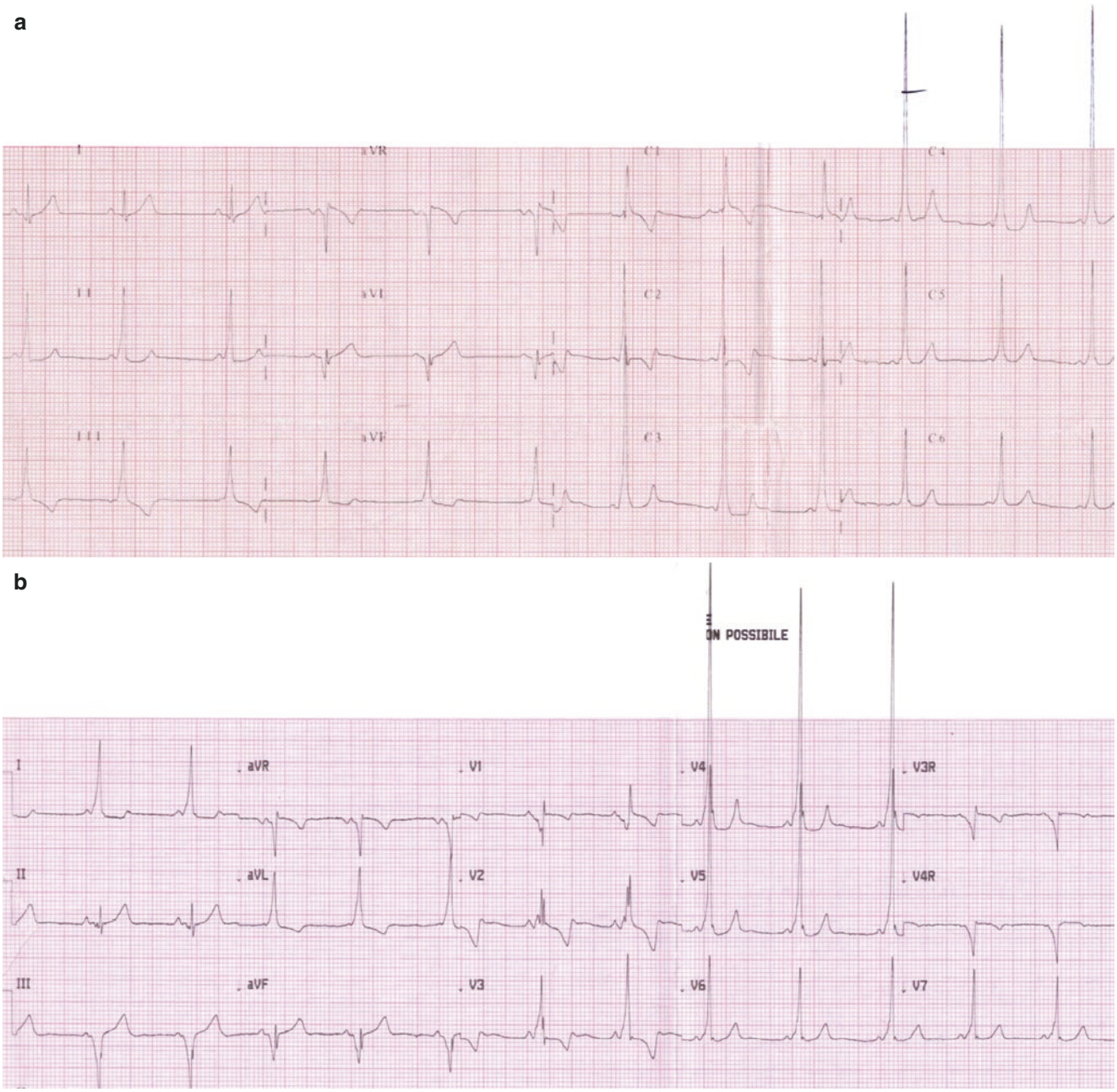

Fig. 10.17 Sixteen-year-old male with EA and double accessory pathway. (**a**) Midseptal AP. (**b**) Right postero-septal AP

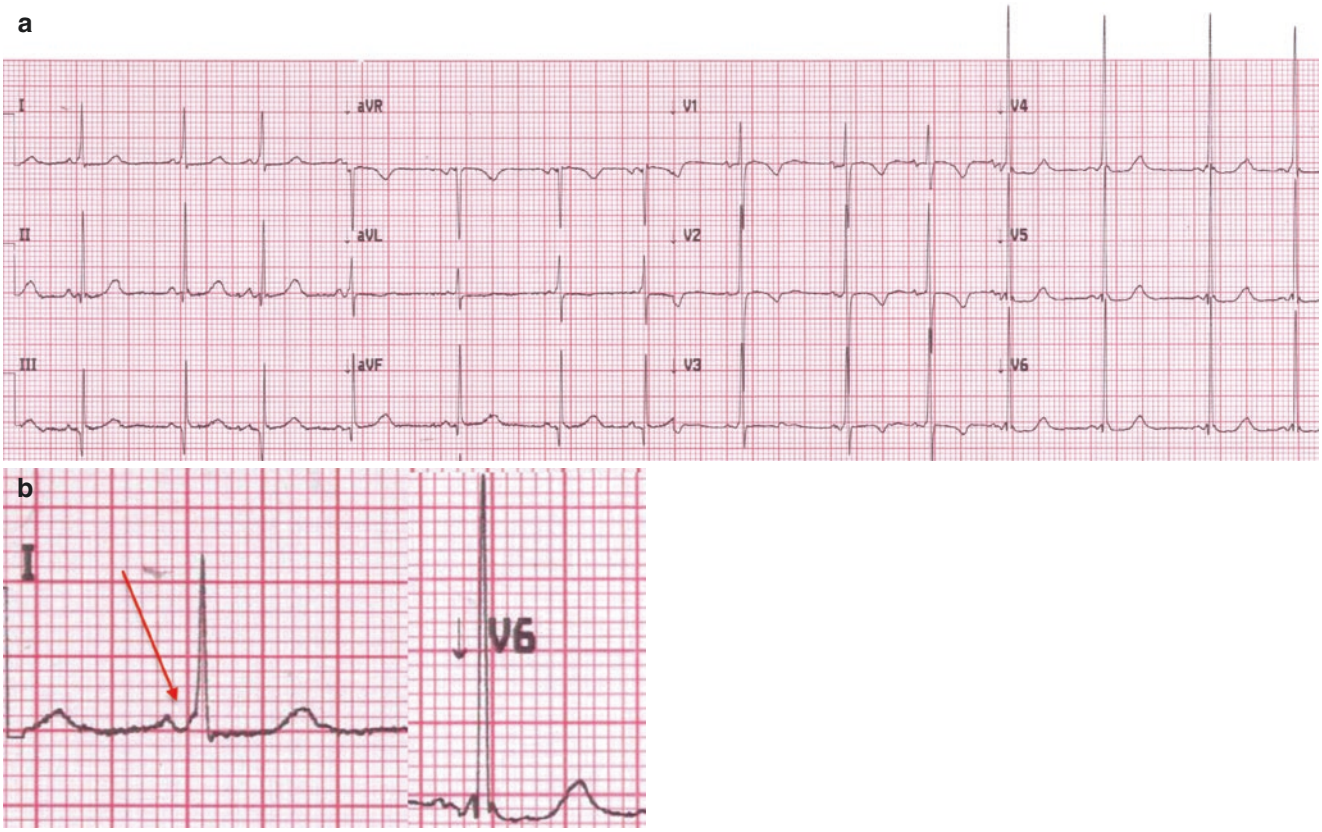

Fig. 10.18 (**a**) This 10-year-old very active (soccer) male was disqualified with diagnosis of HCM. The LV septum was 11 mm thick (diastole); the "slurred" QRS and the ST changes were believed due to HCM. The adenosine test confirmed an AP and a more careful echo was normal. (**b**) Close-up

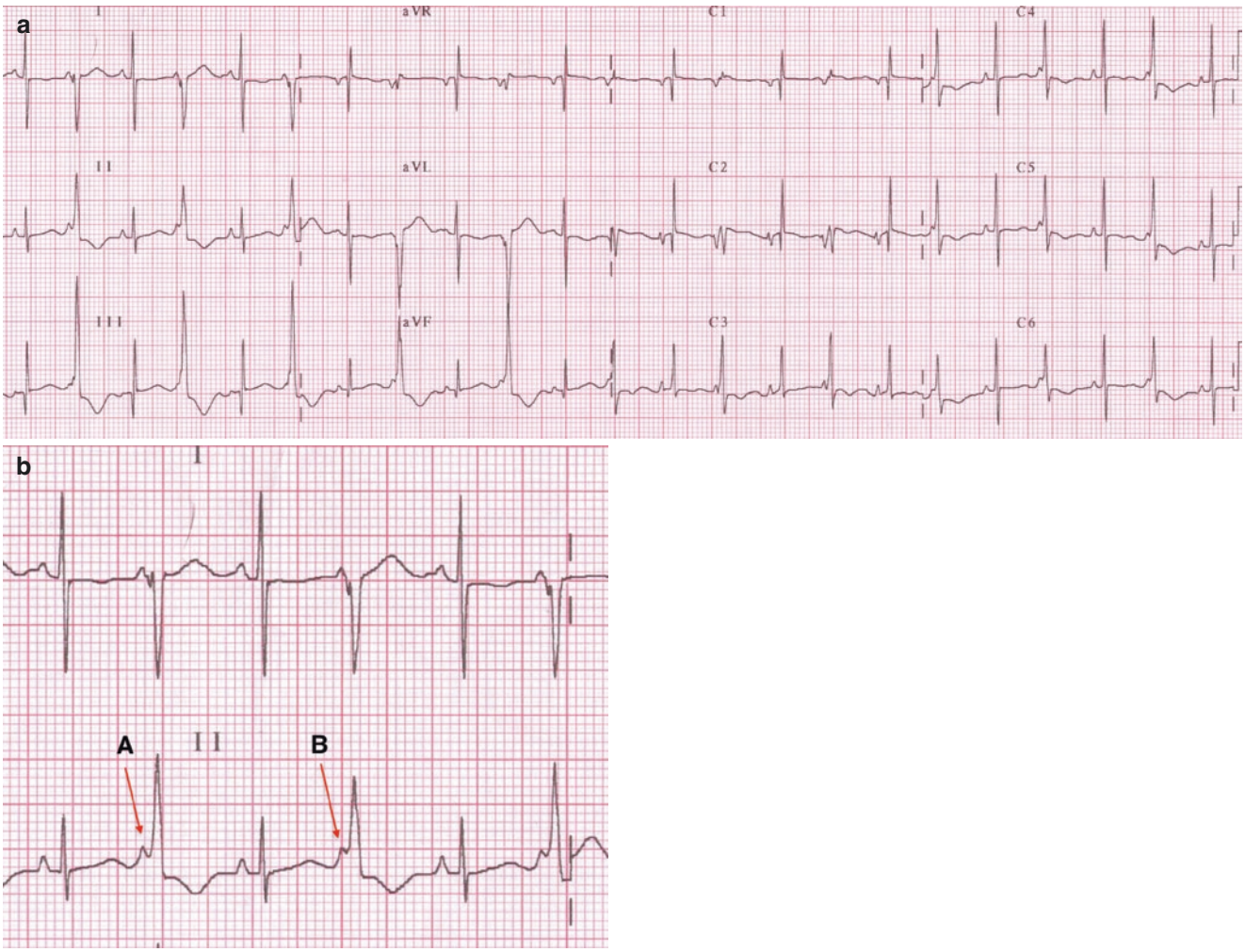

Fig. 10.19 (**a**) Neonate with diaphragmatic hernia. The alternation between wide and narrow QRS is consistent with both pre-excitation and ventricular bigeminism. (**b**) The degree of pre-excitation may be slightly different, as in the complex A and B. Subsequently, with prolonged monitoring, an accessory pathway was demonstrated (note that in the "wide" QRS, the rapid intrinsicoid deflection rules out a ventricular ectopy, a rule working especially in adults)

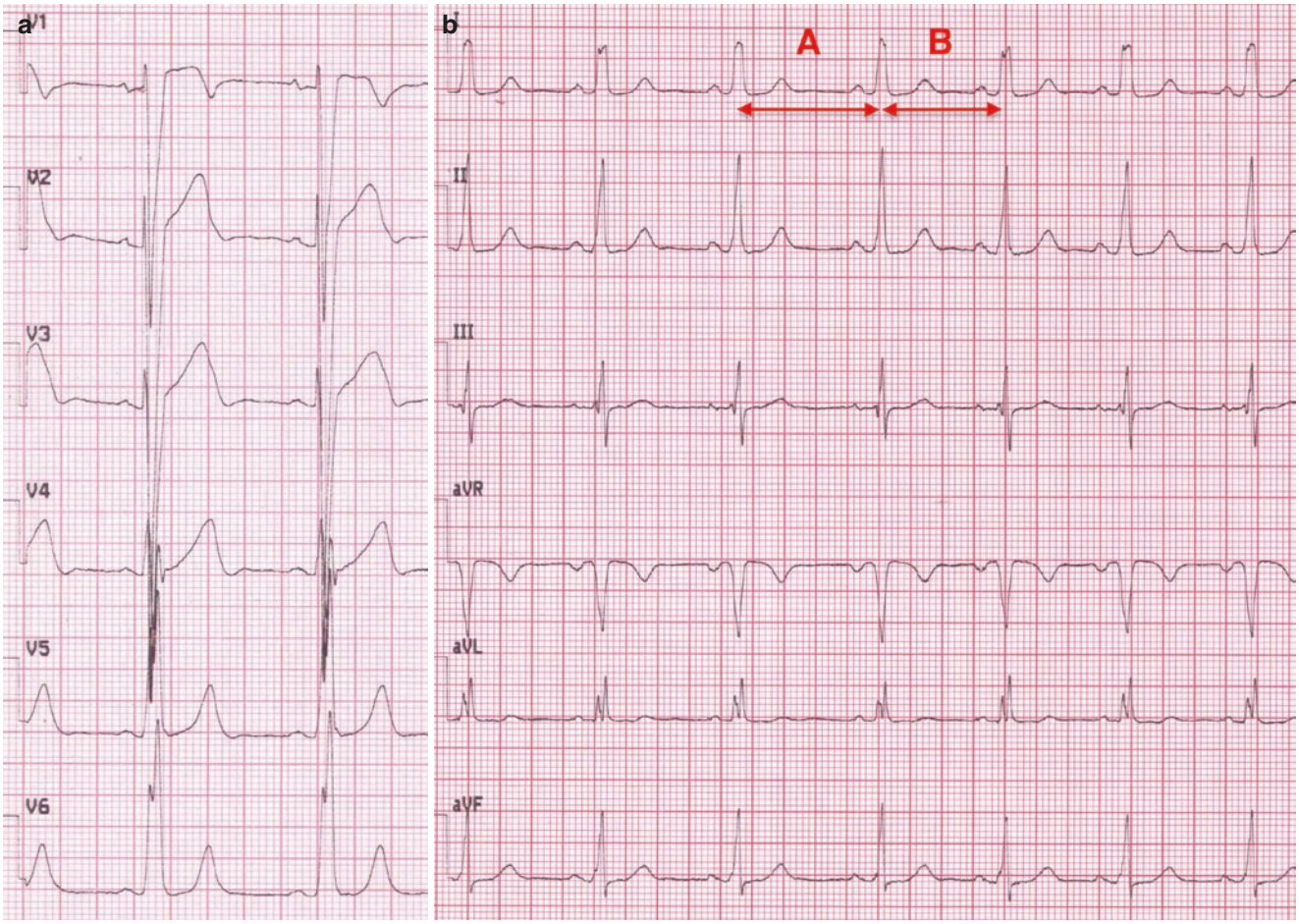

Fig. 10.20 (**a**) Thirteen-year-old patient with Mahaim fibers. In sinus rhythm there is no clear pre-excitation, but the QRS has a typical LBBB pattern; (**b**) the longer the R–R cycle, the more the degree of LBBB increases. This behavior is not what is expected in a simple HR-dependent LBBB, but the opposite, and is thus consistent with Mahaim connection as in this case the EPS demonstrated

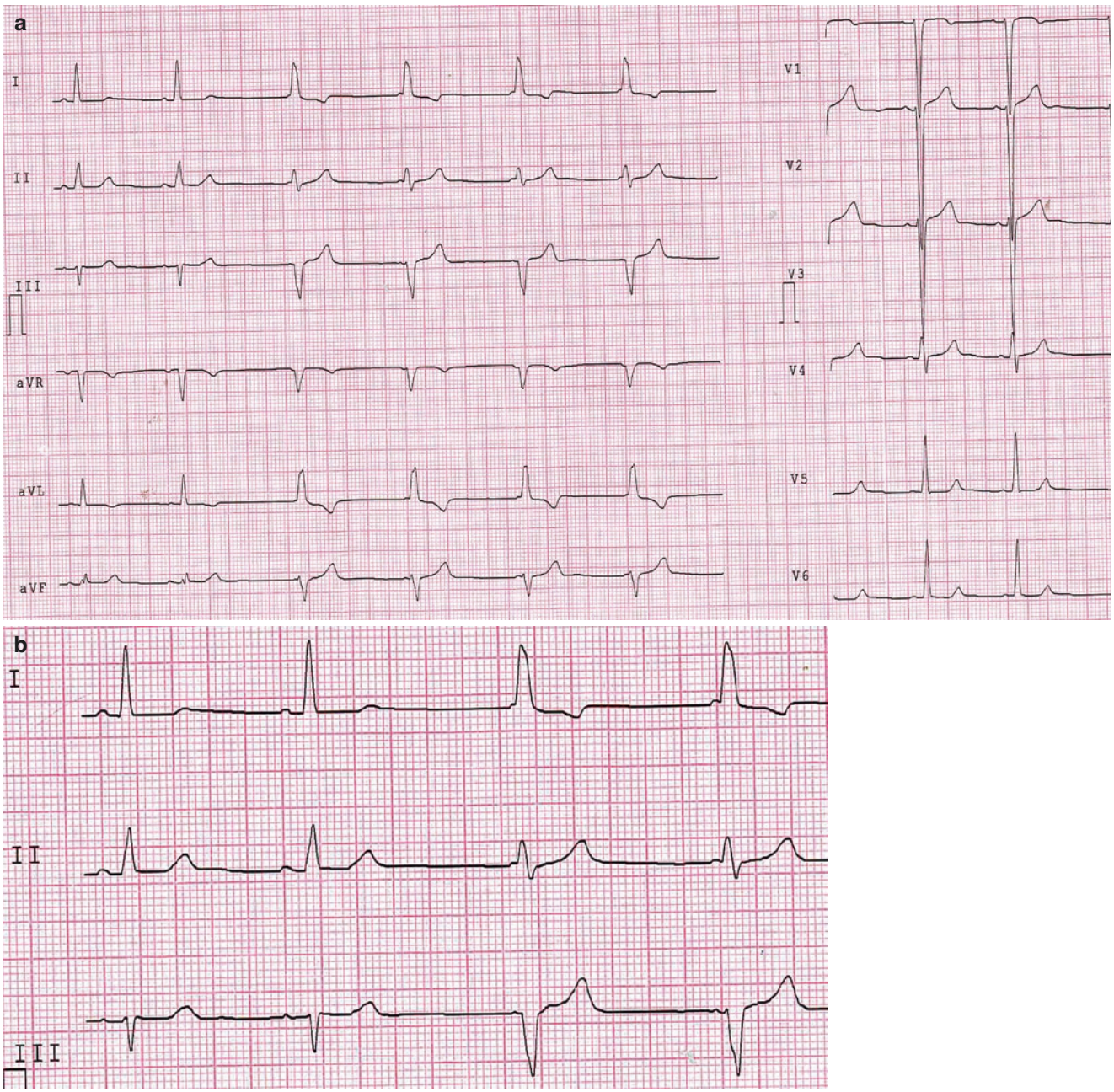

Fig. 10.21 (**a**) Fifteen-year-old male. Another case of Mahaim connection confirmed by endocavitary electrophysiological study (EPS). The ECG shows the typical features of rS in lead III; there is also a delta wave with very short PR which is rare in Mahaim. As in the previous case, the degree of pre-excitation is HR dependent. (**b**) Close-up

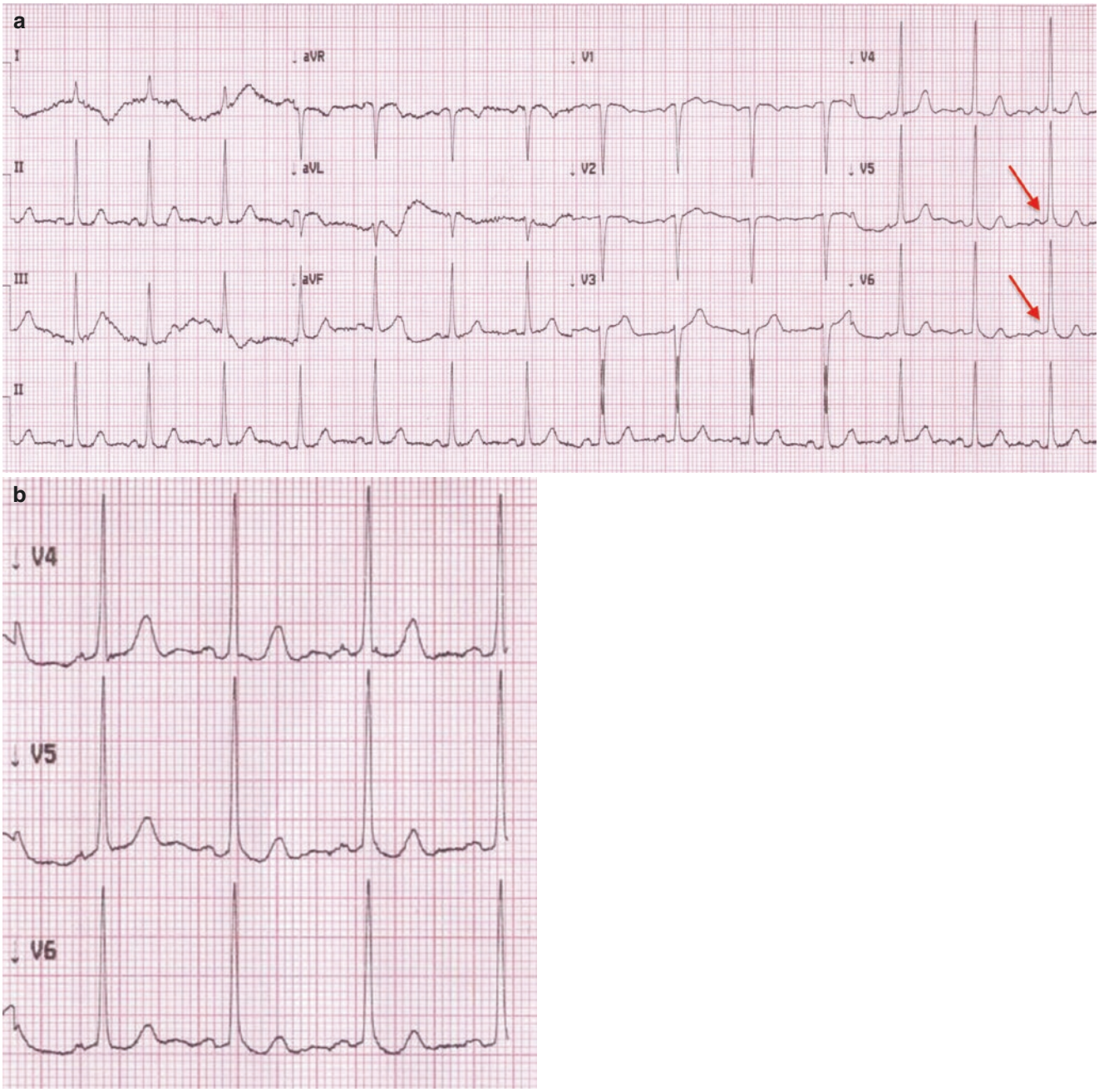

Fig. 10.22 (**a**) Fourteen-year-old male. Pre-excitation in a Bethlem syndrome (autosomal dominant myopathy, a form of muscular dystrophy caused by a mutation of gene coding for type VI collagen). Also in Duchenne dystrophy there is a relative prevalence of APs. (**b**) Close-up

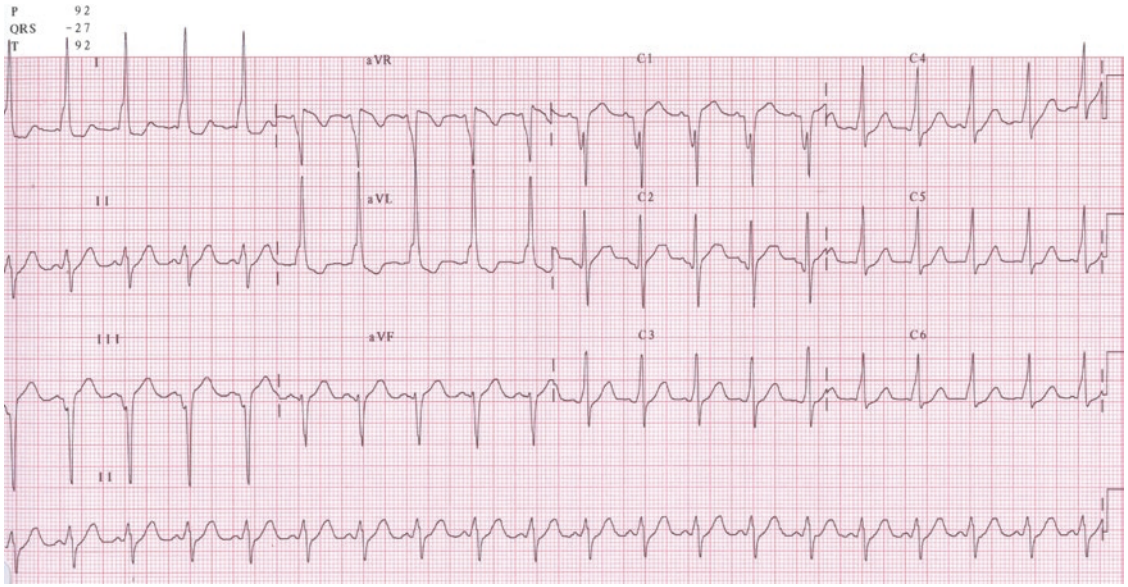

Fig. 10.23 Seventeen-year-old female. Right free wall AP

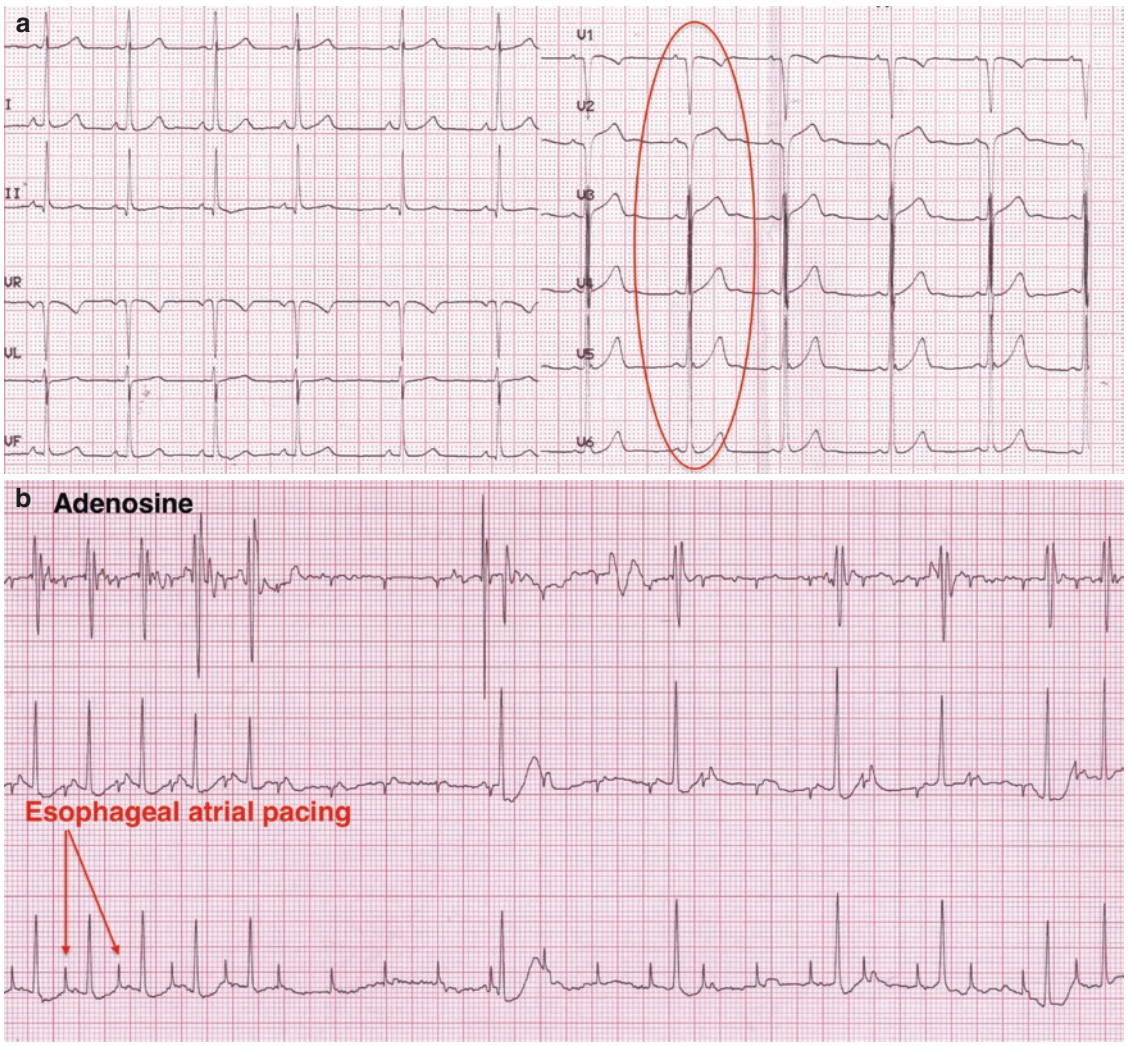

Fig. 10.24 (**a**) Twelve-year-old soccer player. The poor progression of the R wave in the right precordial leads and the absence of Q in V6 raised the issue of pre-excitation. (**b**) The adenosine test during esophageal pacing demonstrated a drug-induced AV block; considering that APs are usually insensitive to adenosine, this is strong evidence that no APs are present. Interestingly, the girl's father had an ECG very similar to his daughter's

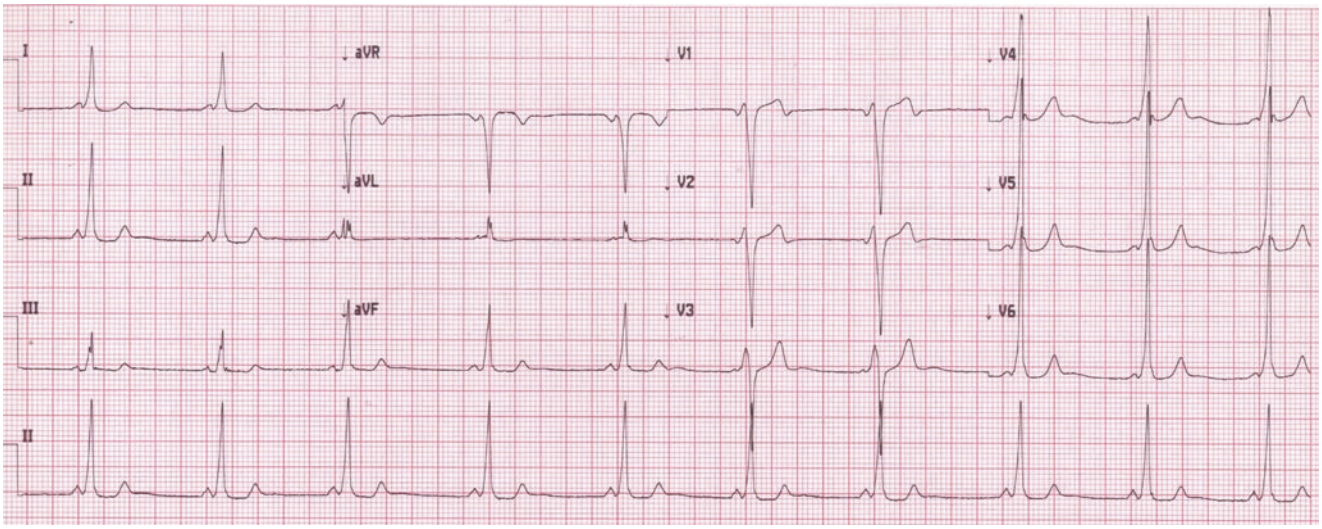

Fig. 10.25 Fourteen-year-old male. The WPW pattern is consistent with parahissian AP which was confirmed on EPS study

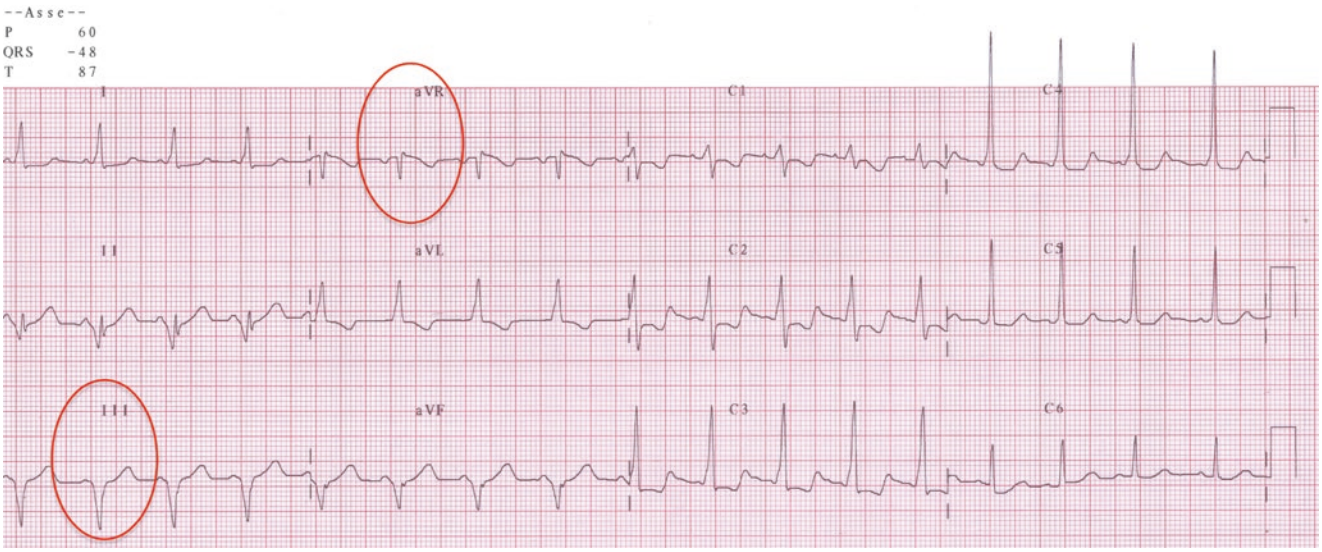

Fig. 10.26 Sixteen-year-old male. Left-sided posterolateral AP

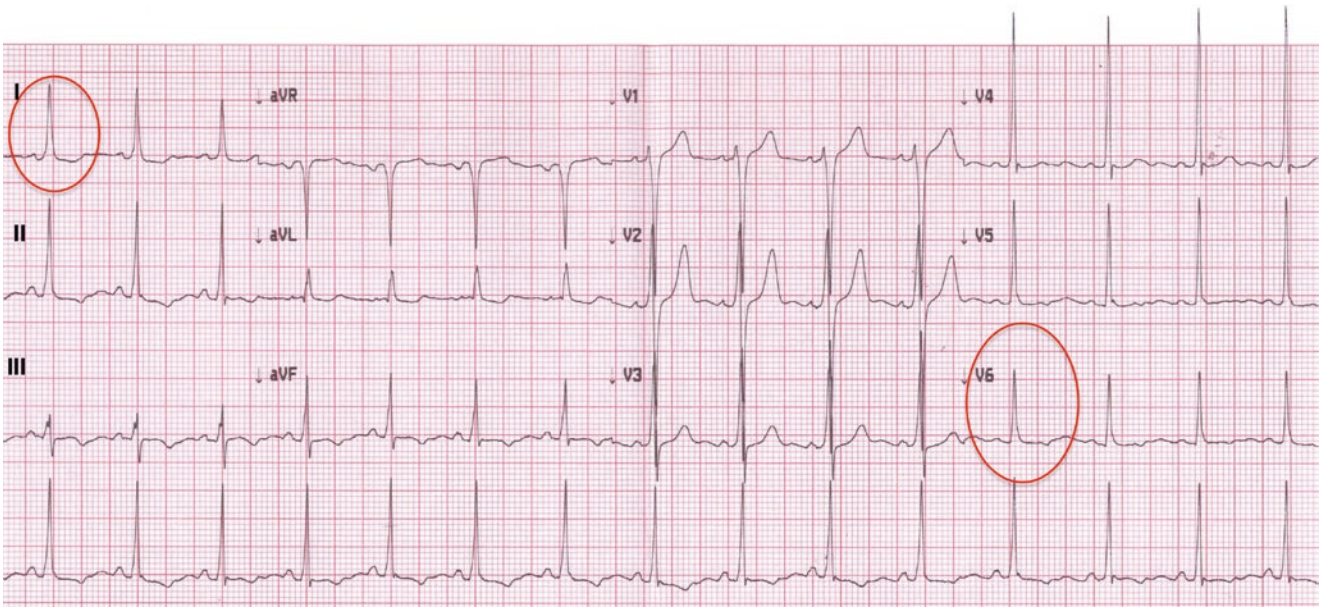

Fig. 10.27 Thirteen-year-old female with MVP and mild mitral regurgitation. The ST-T changes were attributed to the MVP. The PR is not that short but there is evidence of a delta wave which was confirmed on exercise test (at heart rate >150 bpm, delta wave and ST-T changes disappear)

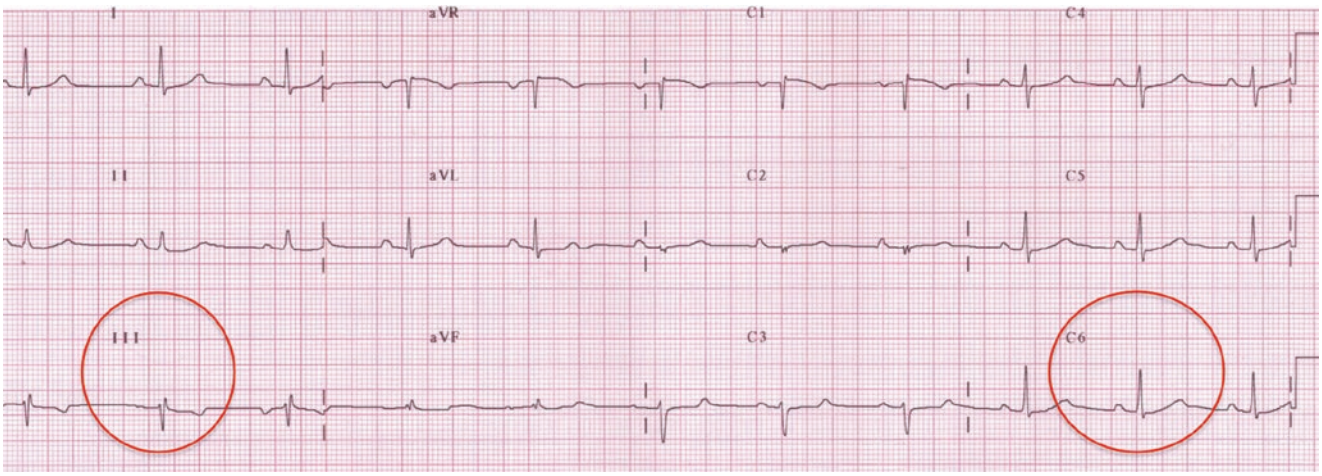

Fig. 10.28 Seven-year-old male. Ebstein's anomaly with severe tricuspid regurgitation and Mahaim fibers. The PR interval is prolonged; there are Mahaim features such as rS in lead III and absence of Q wave in V6. The low voltages, especially in leads II, aVF, and V2, can refer to initial fibrosis

Suggested Reading

Cohen MI, Triedman JK, Cannon BC, Davis AM, Drago F, Janousek J, Klein GJ, Law IH, Morady FJ, Paul T, Perry JC, Sanatani S, Tanel RE. PACES/HRS expert consensus statement on the management of the asymptomatic young patient with a Wolff-Parkinson-White (WPW, ventricular preexcitation) electrocardiographic pattern: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology Foundation (ACCF), the American Heart Association (AHA), the American Academy of Pediatrics (AAP), and the Canadian Heart Rhythm Society (CHRS). Heart Rhythm. 2012;9:1006–24.

Al-Khatib SM, Arshad A, Balk EM, Das SR, Hsu JC, Joglar JA, Page RL. Risk stratification for arrhythmic events in patients with asymptomatic pre-excitation: A systematic review for the 2015 ACC/AHA/HRS guideline for the management of adult patients with supraventricular tachycardia: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. Heart Rhythm. 2016;13:e222–37.

Brugada J, Blom N, Sarquella-Brugada G, et al. European Heart Rhythm Association; Association for European Paediatric and Congenital Cardiology. Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement. Europace. 2013;15(9):1337–82. doi:10.1093/europace/eut082.

Supraventricular Tachycardia

11

In young people, premature atrial contractions (PACs) can be frequent. Sometimes they are not transmitted to the ventricles or are conducted with aberrancy and therefore appear in the ECG with a wide QRS. PACs are normally a benign situation except when they trigger a reentry tachycardia. We should keep the number 220 in mind, quite an electrical number (220 V is the domestic voltage in most countries), as an upper limit for a sinus tachycardia. The reentry form is the most frequent mechanism of supraventricular tachycardia (SVT) in all ages. When SVT is diagnosed during fetal life, antiarrhythmic drugs are given to the mother in order to delay delivery. Atrial flutter is known as a typical fetal and newborn arrhythmia that could be fatal if misunderstood but which is extremely docile once treated with current synchronized cardioversion, with a low recurrence risk. In newborns, SVT is normally caused by an accessory pathway and is conducted between 250–300 bpm, while in children older than 6 years, nodal reentry SVT could appear with a frequency around 200 bpm (Fig. 11.1) [1]. The diagnosis of SVT in an infant is not easy inasmuch as it is frequently confounded with sepsis, heart failure, and decompensated ductus-dependent congenital heart diseases (i.e., aortic coarctation). In addition to its therapeutic efficacy, adenosine plays a diagnostic role, which is why it is named *diagnosine*, since adenosine makes it possible to discriminate between tachycardia depending on the atrioventricular node (AV node) and tachycardia not depending on the AV node including ventricular tachycardia (VT). Since atrial fibrillation (AF) is uncommon in the pediatric population, this means that adenosine is safe even in preexcitation syndromes (Fig. 11.2). A particularly unique arrhythmia is Coumel's tachycardia (also known as permanent junctional reciprocating tachycardia (PJRT), an inappropriate definition because it is not a junctional tachycardia but an AV orthodromic reentry due to a slow conducting para junctional accessory pathway), treacherous for its heart rate, not that fast, that could be interpreted as normal in a routine clinical evaluation, but nevertheless capable of causing tachycardiomyopathy if not discovered and treated [2]. Automatic SVT, including incessant SVT, can appear at any age sometimes after flu or surgical procedures. Multifocal atrial tachycardia is typical during acute viral bronchiolitis (especially associated with respiratory syncytial virus) or during Costello syndrome [3].

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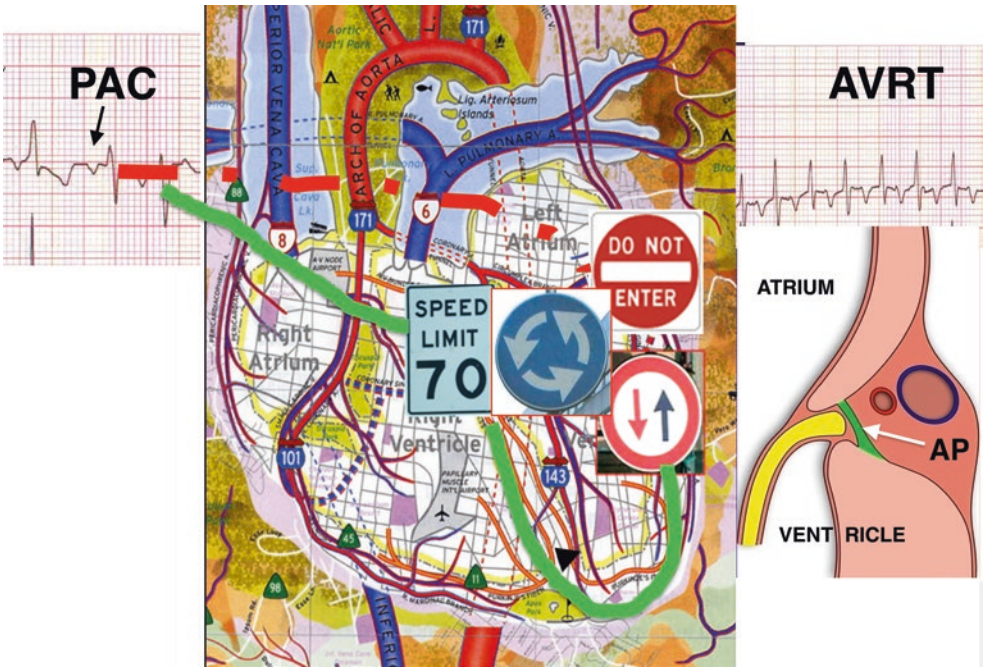

Fig. 11.1 Schematic "road map" of reentrant SVT. In a context of pre-excitation, a PAC finds the AP refractory while the AV node – which has a shorter refractory period – is capable of anterograde conduction. Once in the ventricle, the wave front can travel to the atrium via the AP, which in the meantime has had time to recover excitability. This retrograde conduction reexcites the atrium and may perpetuate the reentry

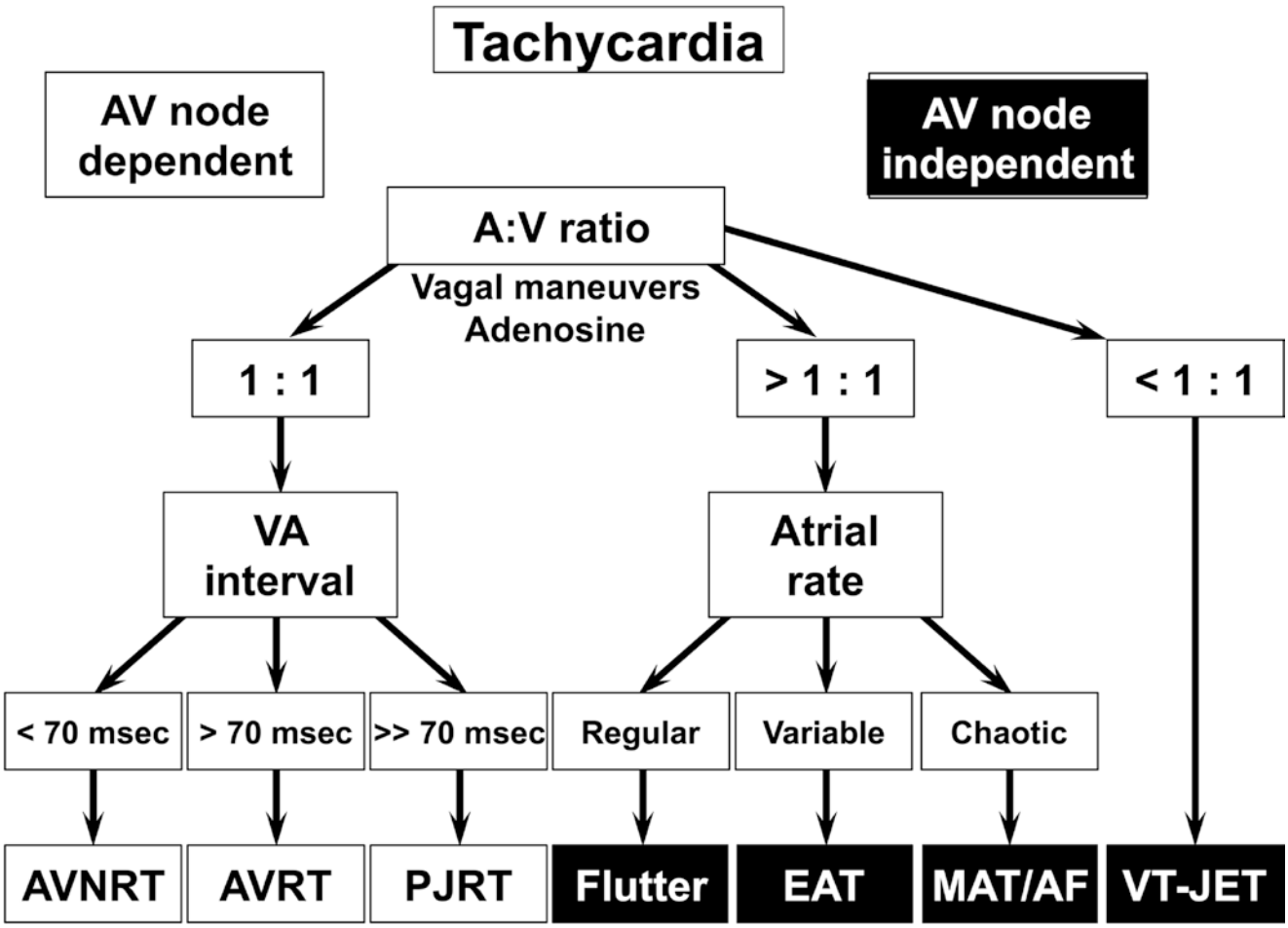

Fig. 11.2 Diagnostic algorithm for tachycardia. Modified with permission from [1]

11.1 ECG Diagnosis

The 12-lead ECG is crucial and normally helps in identifying the type of SVT. Things to be checked are heart rate, P waves (morphology, axis, the relationship between P waves and the QRS complex, PR and RP intervals), QRS complex, starting and finishing pattern of tachycardia, and the response to vagal maneuvers and to drugs (Figs. 11.2 and 11.3) [1]. If the baseline ECG with normal sinus rhythm presents a ventricular pre-excitation (short PR interval, delta wave, wide QRS, ST-T changes), the diagnosis of an atrioventricular reentry tachycardia is obvious, while the presence of a long PR interval should suggest a nodal reentry (dual AV nodal pathways and baseline conduction along the slow pathway), and in the case of Mahaim fibers, a normal PR interval, a pseudo-pre-excitation, and a pattern rS in DIII may be present [4].

When a newborn baby arrives in the emergency room in poor clinical conditions and the 12-lead ECG shows sinus rhythm with large P waves, keep in mind the possibility of a just ceased SVT or VT with AV dissociation. STVs are normally characterized by a narrow QRS complex; however, a wide QRS complex could be present in SVT conducted with aberrancy, Mahaim tachycardia, or antidromic AVRT or VT (AV dissociation, fusion beats, capture beats, concordance throughout the chest leads) (Figs. 11.4 and 11.5). Mahaim fibers connect the anterior wall of the right atria to the right bundle branch, to the fascicles, or to the right distal ventricle and possess decremental conduction capacity; they lead to antidromic arrhythmia with an LBBB morphology. Mahaim arrhythmia is very rare, and Mahaim fibers are frequently a bystander of other arrhythmic mechanisms [1].

Recognizing P waves is not easy at a fast heart rate, mostly because they can be hidden in the QRS complex or in the T wave. When they are visible with an RP interval less than the PR interval, the most likely diagnosis is atrioventricular reentry by an accessory pathway (AVRT), since atria activation follows ventricular activation usually with an interval exceeding 70 ms. In this case, the P wave can be hidden in the ST segment or embedded in the T wave ("camel hump," notch) [5].

On the contrary, in the case of typical atrioventricular nodal reentrant tachycardia (AVNRT) slow-fast type, atrial and ventricular activation is almost simultaneous, and P waves are therefore hidden in QRS waves or may form the final part of the QRS complex, creating pseudo S waves in leads II, III, and aVF or pseudo R waves in V1. To confirm the diagnosis, these appendices should not be evident during sinus rhythm. The relationship between the PR and RP intervals is very important: the RP is virtual or extremely short in the common AVNRT (slow-fast type) and junctional ectopic tachycardia (JET); the RP interval is long in sinus tachycardia, sinoatrial reentrant tachycardia, atrial ectopic tachycardia

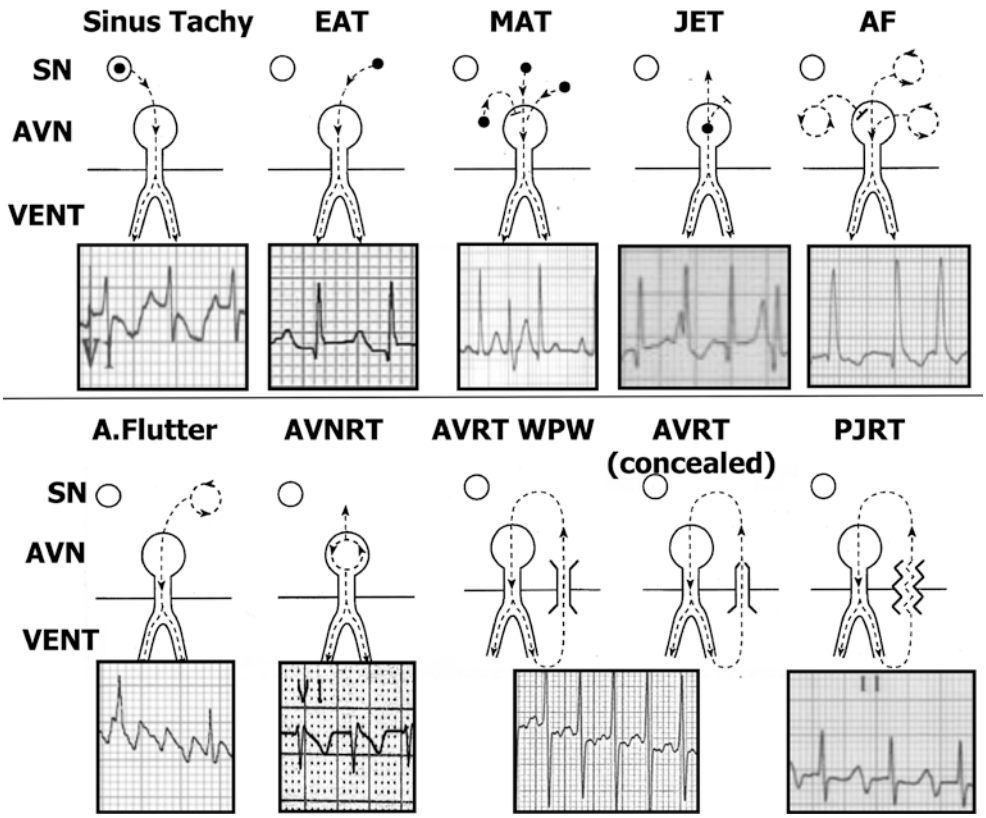

Fig. 11.3 SVT synopsis. Modified with permission from [1]

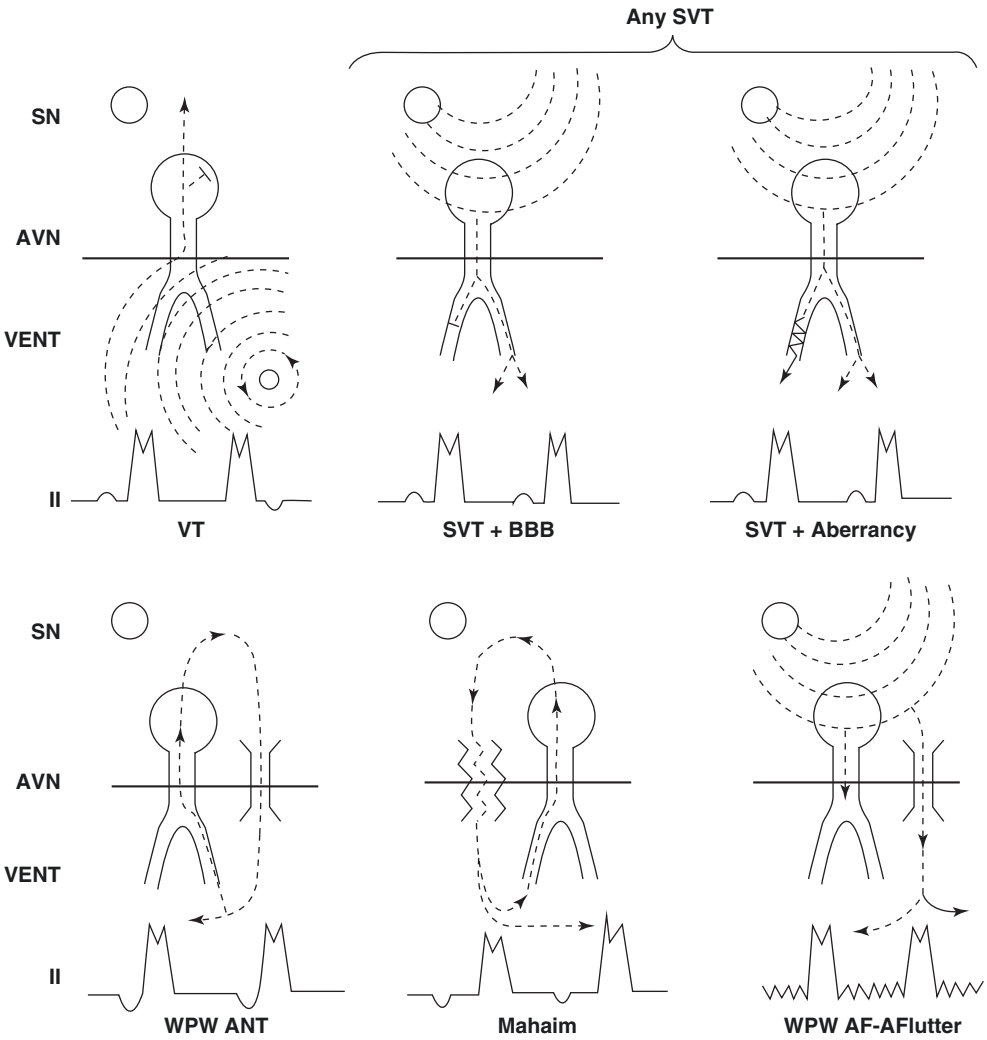

Fig. 11.4 Wide complex tachycardia. Modified with permission from [1]

(AET), atrial flutter, AVNRT (fast-slow or slow-slow), and permanent junctional reciprocating tachycardia (PJRT). In the AVRT using an AP, the RP interval depends on the location of the bypass tract but, in most cases, is >70 ms. If the P wave is visible, in addition to the relationship with the QRS complex, its morphology and axis could be important in order to identify the direction of atrial activation. In the commonest forms of SVT (AVRT and AVNRT), the atria are depolarized from the junction to the top (concentric caudo-cranial activation), leading to negative P waves in the inferior leads. Therefore, in the case of tachycardia and positive P waves in leads II, III, and aVF, the diagnosis of AVNRT and AVNRT can be excluded. PJRT and atypical AVNRT are characterized by a normal or slightly prolonged PR interval with negative P waves in the inferior leads and positive P waves in aVR and aVL. The P wave polarity in lead I has a diagnostic value (positive is typical of AVNRT, negative of PJRT) as well as other clinical and electrophysiological features (AVNRT is usually paroxysmal and starts with ectopic beats, while PJRT is incessant and begins with a critical shortening of the sinusal cycle). Atrial ectopic tachycardia (AET) arises in an ectopic focus which can be single or multiple or congenital or acquired. The P wave axis helps to identify the origin of the arrhythmia either near to the "crista terminalis" or to the right pulmonary veins or parasinusal: in that case, the P is pseudo-normal and an arrhythmia should be suspected due to a heart rate inappropriate for the age or physiological state. In patients with AET, a first-degree AV block could be present; second-degree Mobitz 1 AVB either spontaneous or induced could be helpful for the diagnosis. AETS are normally non-responsive to adenosine; in exceptional cases the drug could slow or block the arrhythmia if the mechanism is the triggered activity.

JET is the only SVT with AV dissociation and sinus P wave. The P wave can be simultaneous, just before or just after the QRS complex. P waves can be negative in the inferior leads as a result of junctional origin and retroconduction to the atria. AV dissociation could cause atrial dilatation which can be inferred from giant P waves. Postoperative JET can be easily diagnosed using the epicardial wires.

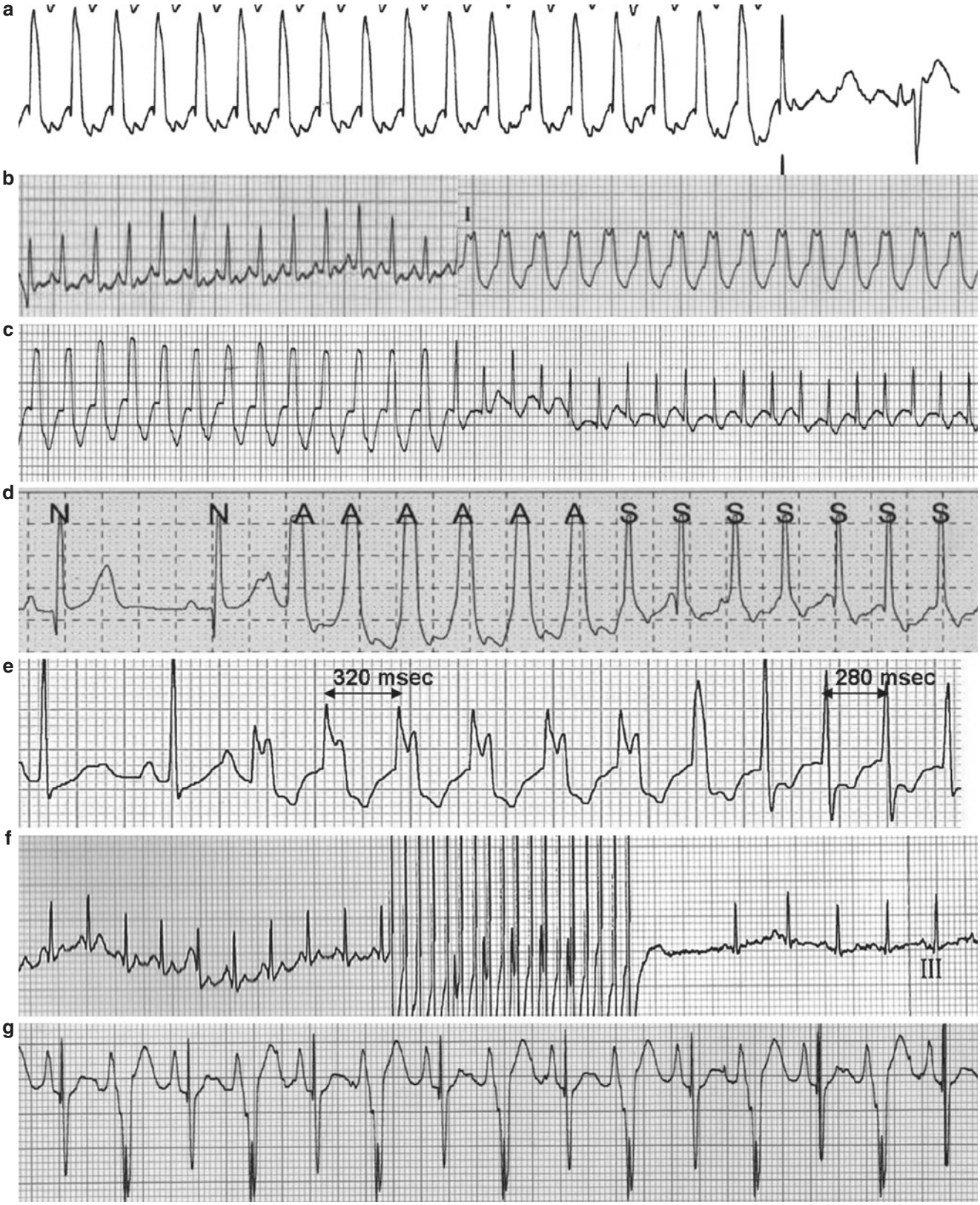

Fig. 11.5 Seven examples of SVT. (**a**) AVRT conducted with LBBB: the P wave is clearly visible; adenosine is effective in terminating the SVT. The last beat of the AVRT is not aberrated. (**b**) Adenosine transforms a narrow complex PSVT in a wide complex PSVT, due to sustained aberrancy (linking phenomenon). (**c**) Adenosine transforms a PSVT conducted with linking in a narrow complex PSVT, by interrupting the linking. (**d**) Spontaneous transition from wide complex (LBBB aberration) to narrow complex. (**e**) (See (**d**)) The transition to narrow complex PSVT significantly shortens the cycle length of the arrhythmias demonstrating that the accessory pathway is to the left, ipsilateral to the branch with transient functional block. (**f**) Esophageal overdrive of neonatal atrial flutter. (**g**) Intermittent pre-excitation in a newborn: the pre-excited beats show a negative delta wave

Another important diagnostic trick in differentiating SVT is the mode of onset and offset. If the arrhythmia begins with a premature ventricular contraction (PVC), the most likely diagnosis is AVRT since the ventricles are included in the mechanism of the arrhythmia, while AVNRT is less likely considering that the PVC rarely penetrates the AV node retrogradely, stopping the arrhythmia [6–9].

Sometimes, the ECG evaluation cannot lead to the diagnosis, and the response to drugs or to vagal maneuvers can be extremely helpful. Adenosine can stop the vast majority of AV reentrant arrhythmias. On the contrary, in automatic/ectopic tachycardia, adenosine is able to slow the ventricular rate by means of AV node block, but not to terminate the arrhythmia. The same can be said of atrial flutter in which adenosine may have a great diagnostic value [10].

The clinical presentation can help to distinguish reentrant from automatic tachycardia. In a neonate the most likely tachycardia is the reentry form due to an accessory pathway, AVRT, while in an older child, an incessant tachycardia could be automatic (AET or JET) with the exception of PJRT.

A newborn with an SVT usually has a heart rate >220 bpm, and a reentry SVT will often have a heart rate >250 bpm. The diagnosis can be confirmed taking into account the response to drugs or to vagal maneuvers, hence keeping in mind the 220 rule and the 30 rule. If the heart rate is between 220 and 250 bpm, even though the P waves seem sinusal, the most likely mechanism is an automaticity, even more so if the HR changes by a range of 30 bpm during 30 min of observation (reentrant arrhythmias have a quite stable HR and are less sensitive to adrenergic stimulation).

The changing and paroxysmal nature of SVT makes diagnosis with a standard ECG difficult and requires extended monitoring either in the hospital or in the clinic. Several ways of monitoring are available: from the classic Holter ECG to the devices provided by modern cardiology-based telemedicine (event recorder, loop recorder, etc.). The "old" Holter is rarely able to capture the fleeting SVT and is often poorly tolerated by children. Hence, telemedicine is becoming of paramount importance to make the monitoring of arrhythmias in the pediatric population more and more personalized (Fig. 11.6). Telemedicine is evolving at a very brisk pace; in addition to hospital-provided remote services, the number of apps to monitor heart rate and arrhythmia is growing on an almost daily basis, turning our mobiles into true "smheartphones" [11]. Even social networks are proving valuable in sharing clinical cases in real time (Fig. 11.7).

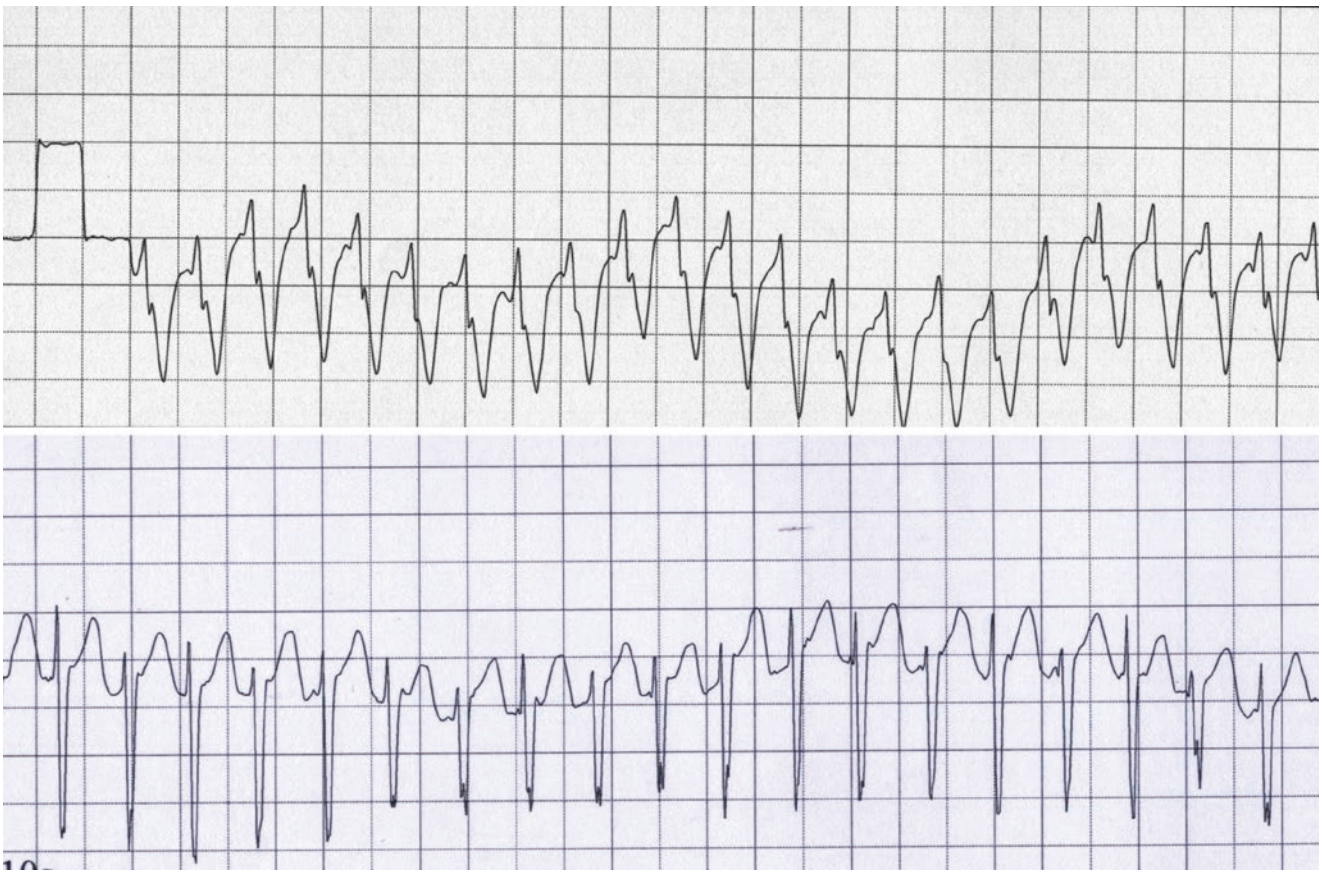

Fig. 11.6 Two cases of VT (wide complex tachycardia) and SVT (narrow complex tachycardia) diagnosed with an event recorder

Fig. 11.7 An example of chat operating in Italy to share interesting or tricky pediatric ECGs

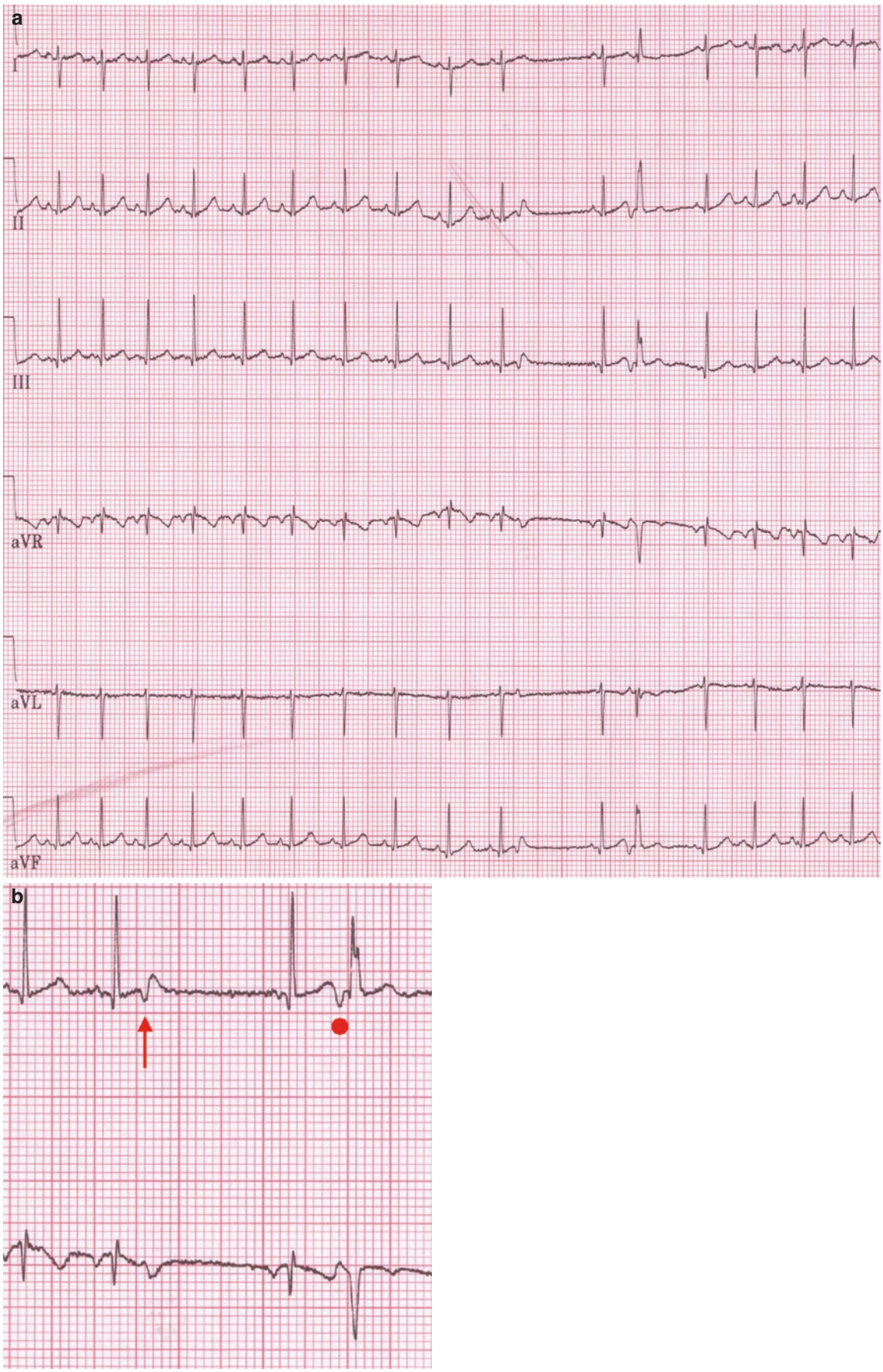

Fig. 11.8 (**a**) Premature atrial contraction (PAC) in a 2-year-old boy. (**b**) Close-up. While one PAC is blocked in the AV node (*arrow*), the following one is conducted with functional bundle branch block, so-called aberrancy (*dot*)

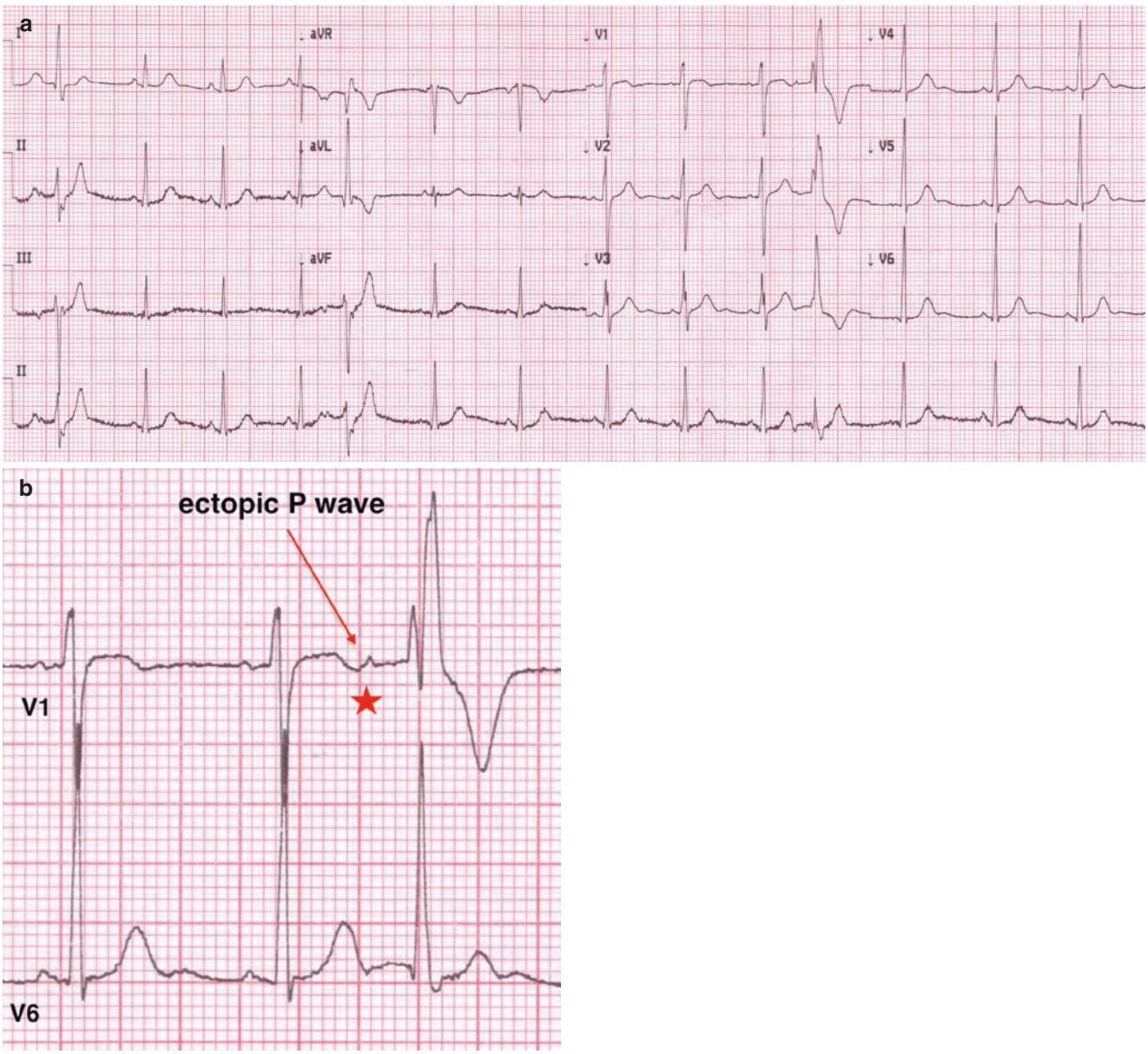

Fig. 11.9 Eight-year-old boy. (**a**) PACs conducted with RBBB aberrancy. (**b**) Close-up

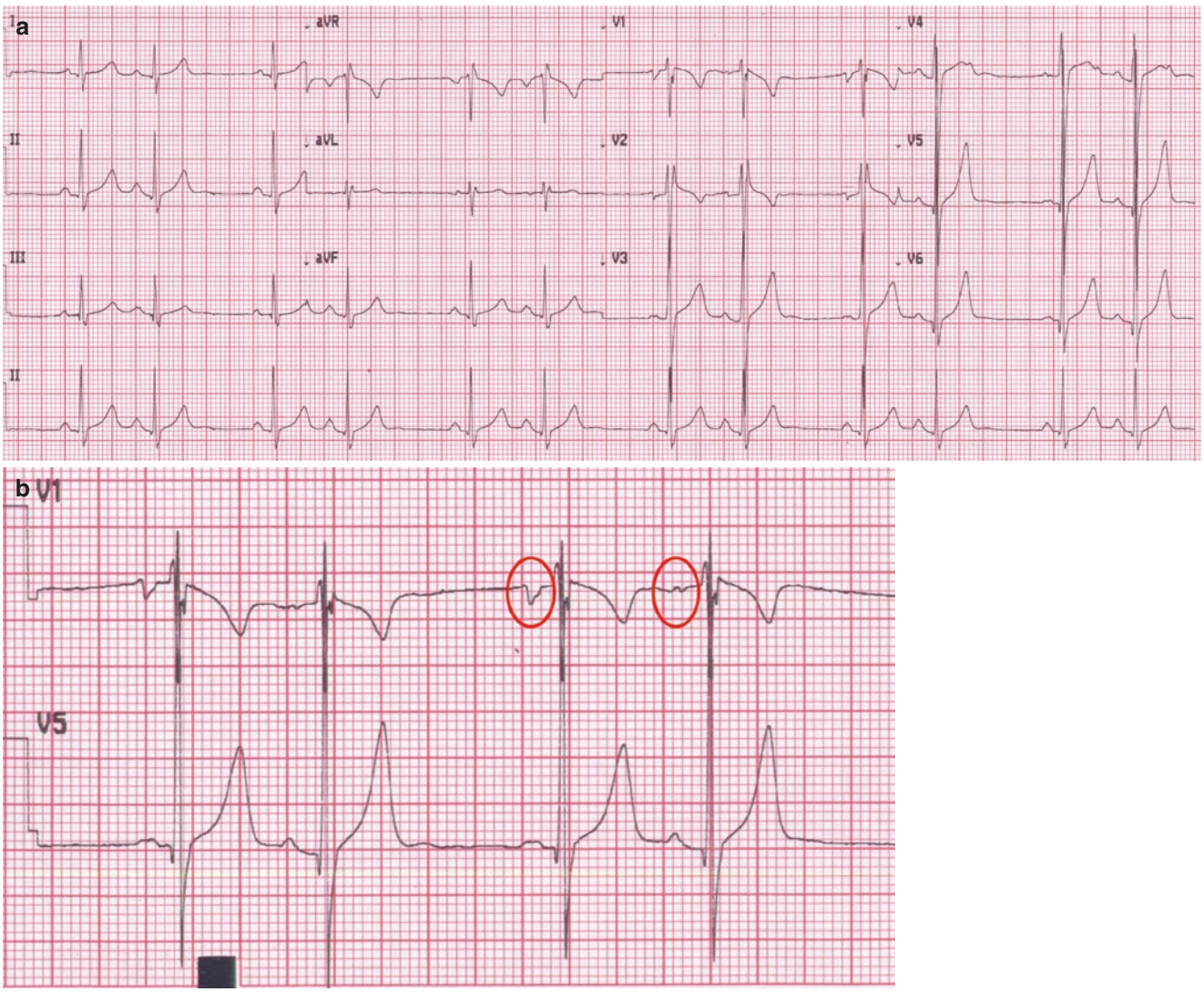

Fig. 11.10 (**a**) Eight-year-old male. PACs in bigeminism. (**b**) Close-up: the anticipated atrial beat is clearly different from the sinus P

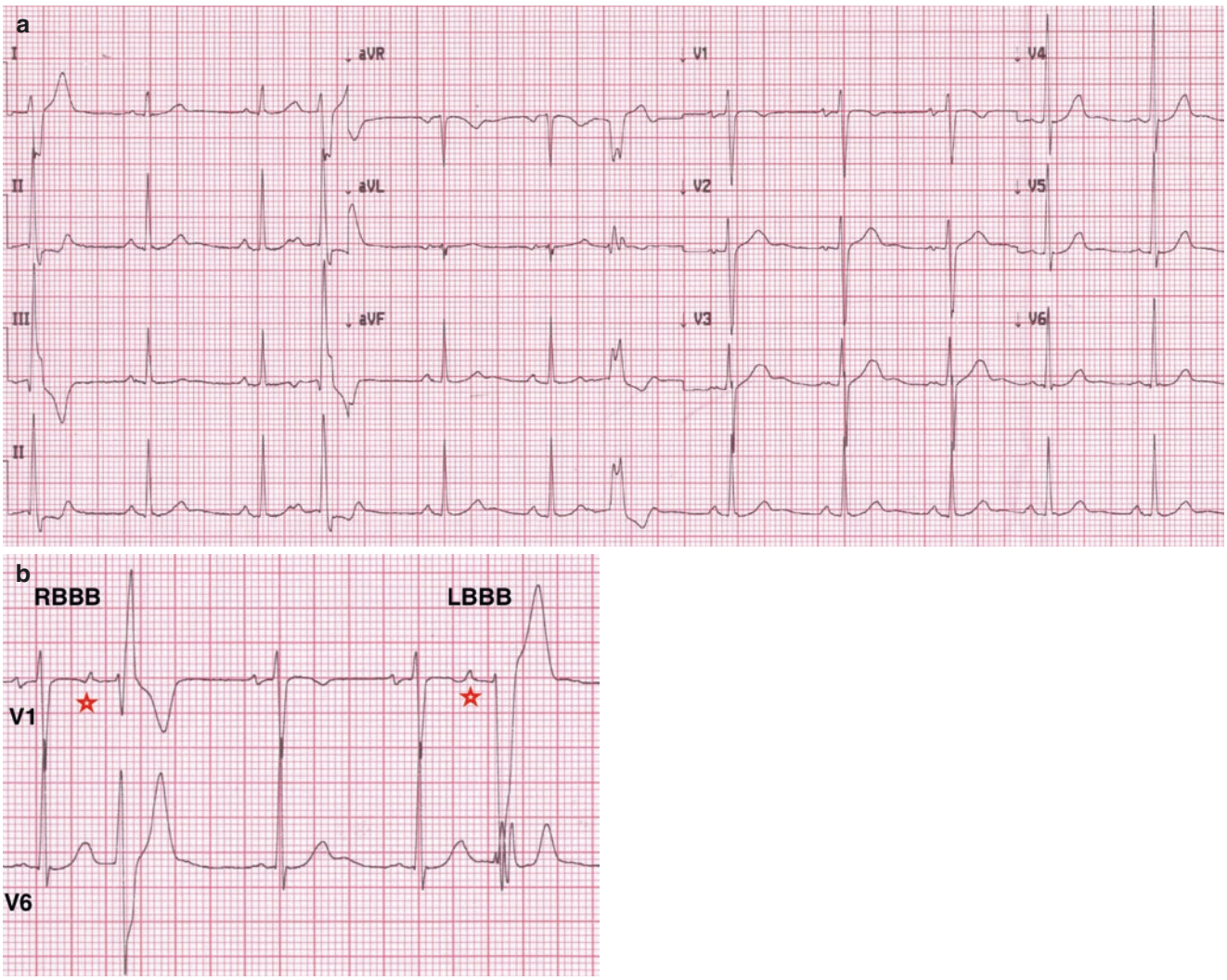

Fig. 11.11 (**a**) Ten-year-old female. PACs conducted with aberrancy. (**b**) Close-up: one PAC is conducted with RBBB, the following one with LBBB. These "wide" beats are occasionally misunderstood for PVCs

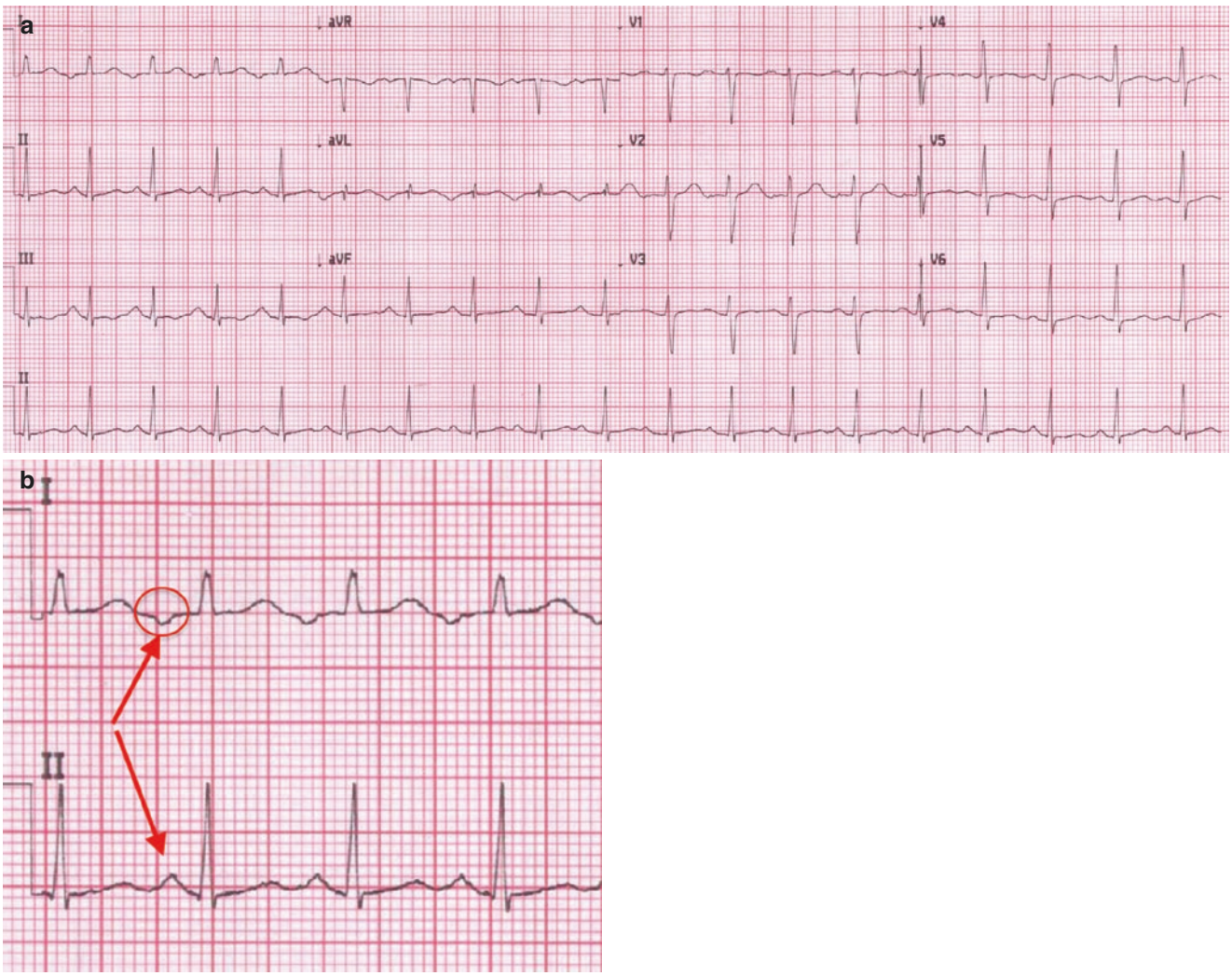

Fig. 11.12 (**a**) Twelve-year-old female. This apparently seems just a sinus tachycardia; the P wave in lead II looks like a sinus P. Paying more attention, the P wave is negative in leads I and aVL (**b**). So the atrial vector is running away from leads I and aVL. In fact, the EPS found an ectopic atrial tachycardia (EAT) arising from the left atrial appendages. The patient successfully underwent radiofrequency catheter ablation

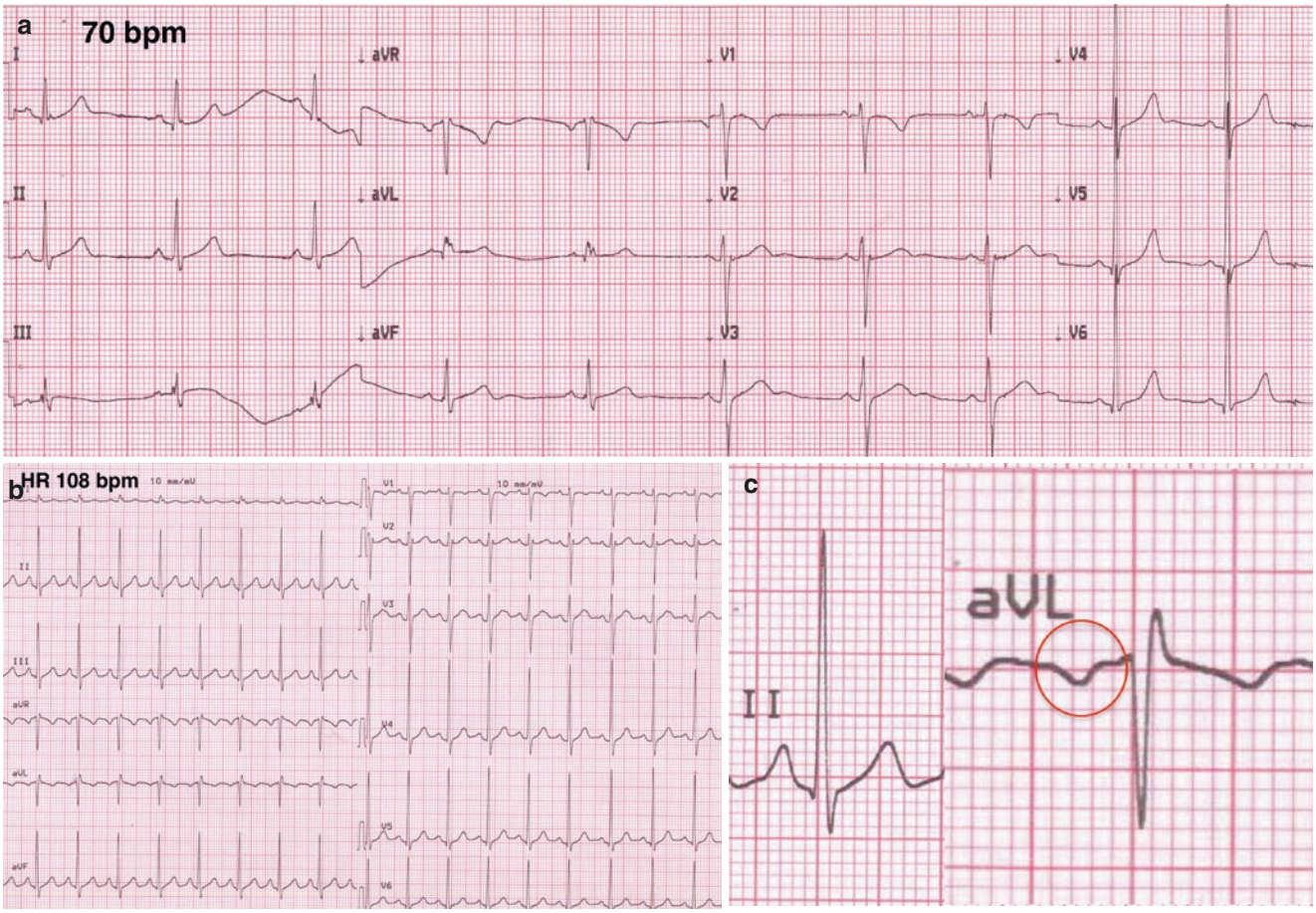

Fig. 11.13 (**a**) Eleven-year-old male soccer player. Normal sinus rhythm 70 bpm. (**b**) The young man became symptomatic due to palpitations, and the ECG revealed an apparent sinus rhythm. The HR was not too fast but stable, almost incessant; a careful analysis found that aVL was negative. This finding rules out a sinus tachycardia and also a sinoatrial reentry tachycardia. The most probable diagnosis is EAT arising in the upper left atrium, very similar to the previous case. As a rule, when the HR, even not that fast, is inappropriate for the age or the situation, an incessant arrhythmia should be excluded; if the QRS is identical to the sinus rhythm, pay attention to the P morphology in all leads. (**c**) Close-up

Fig. 11.14 (**a**) Ten-year-old male with complex CHD (palliated single ventricle physiology). (**b**) Close-up. Run of non-sustained EAT with different degrees of aberrancy. Star = ectopic P wave

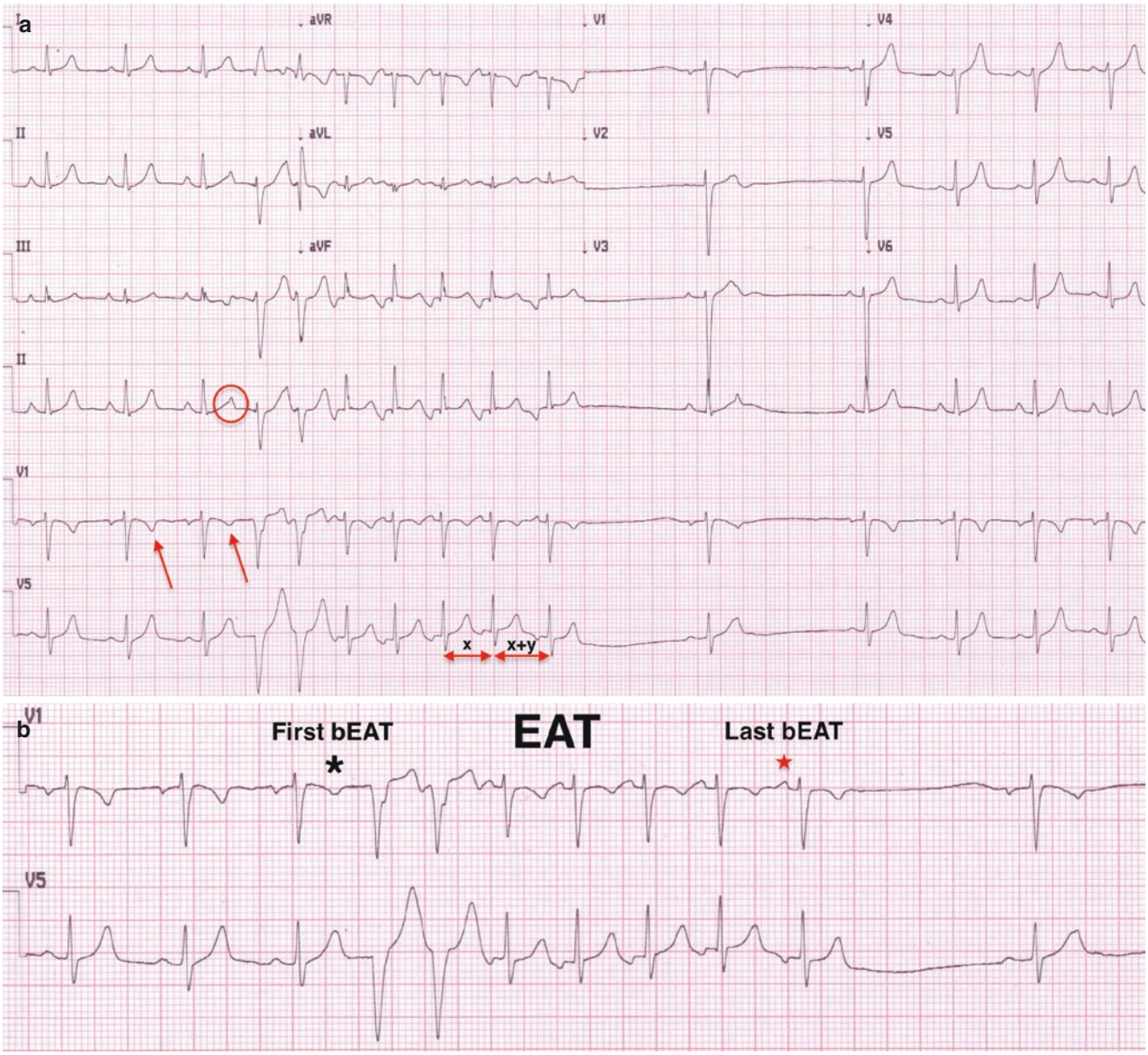

Fig. 11.15 (**a**) Twenty-year-old male with a run of EAT. The first ectopic beat is buried in the T wave: the motto "for one beat the T may eat a bit of EAT" is true (*circle* and *arrows*). There is a cool-down cycle, longer than the previous one, just before the run expires. However, the P waves morphology and the behavior of the arrhythmia is equally compatible with the so-called PJRT (due to an AP with decremental conduction), initiated in this case by a PAC. The following sinus beats are slow due to the overdrive suppression that the EAT made on the sinoatrial cells. (**b**) Close-up

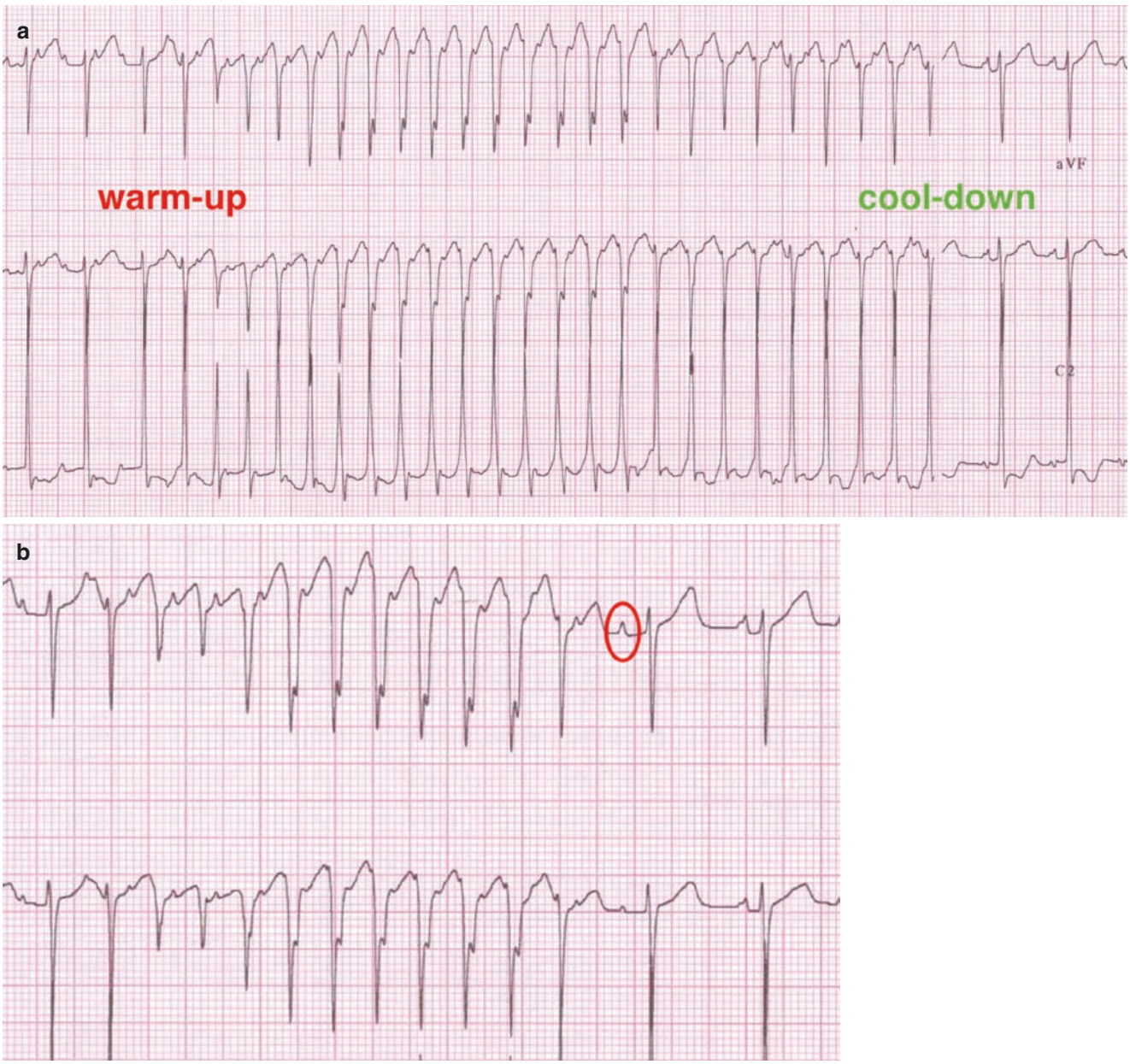

Fig. 11.16 (**a**) Two-year-old male with pulmonary stenosis and EAT. Examples of warm-up and cooldown. At the beginning of the run, a 2:1 A/V conduction ratio can be noticed. These arrhythmias' mode of beginning and ending is typical of automatic or triggered tachycardia. (**b**) Close-up

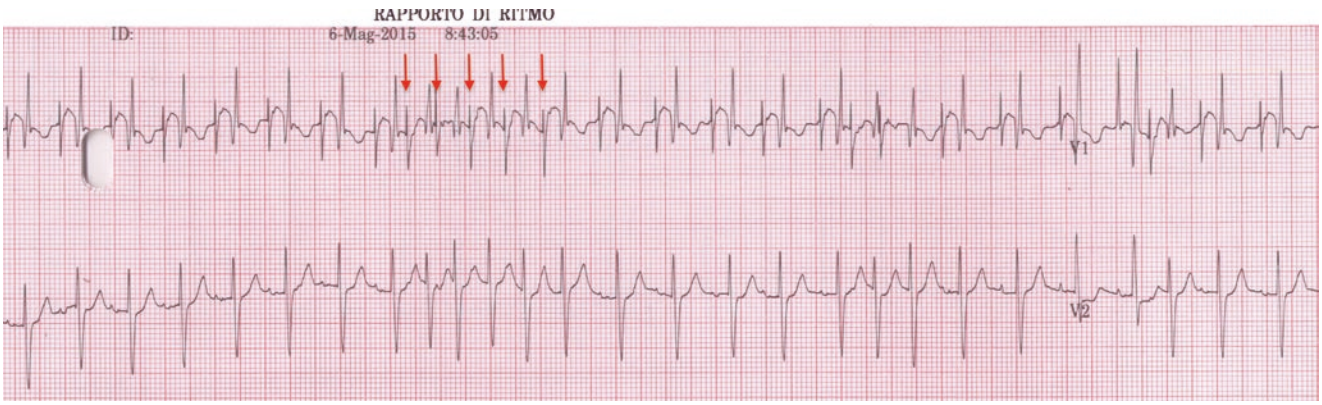

Fig. 11.17 Eight-year-old early postoperative course of VSD closure. Run of EAT; the atrial wires left by the surgeon permit an epicardial signal (*arrows*); the atrial premature contractions are clearly visible. As in the previous case, the burst ends with a cooling down

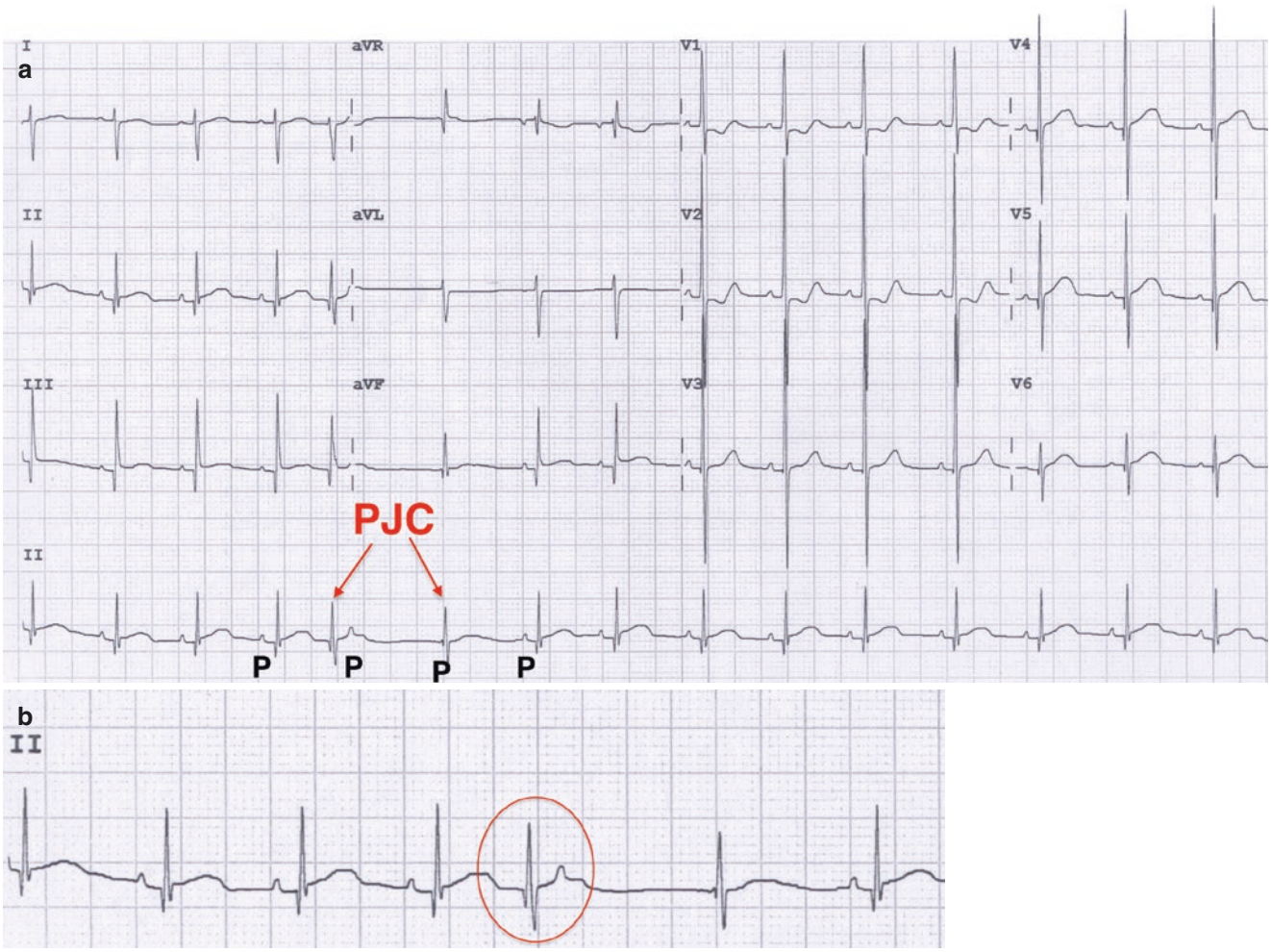

Fig. 11.18 (**a**) Five-year-old male with junctional premature contraction (JPC). The dissociation between the narrow complex – identical to the sinus QRS – and the P wave is clear. The P wave following the first JPC is positive in the inferior leads and therefore must be a dissociated sinus beat because a rectroconducted P wave should be negative in the inferior leads. (**b**) Close-up

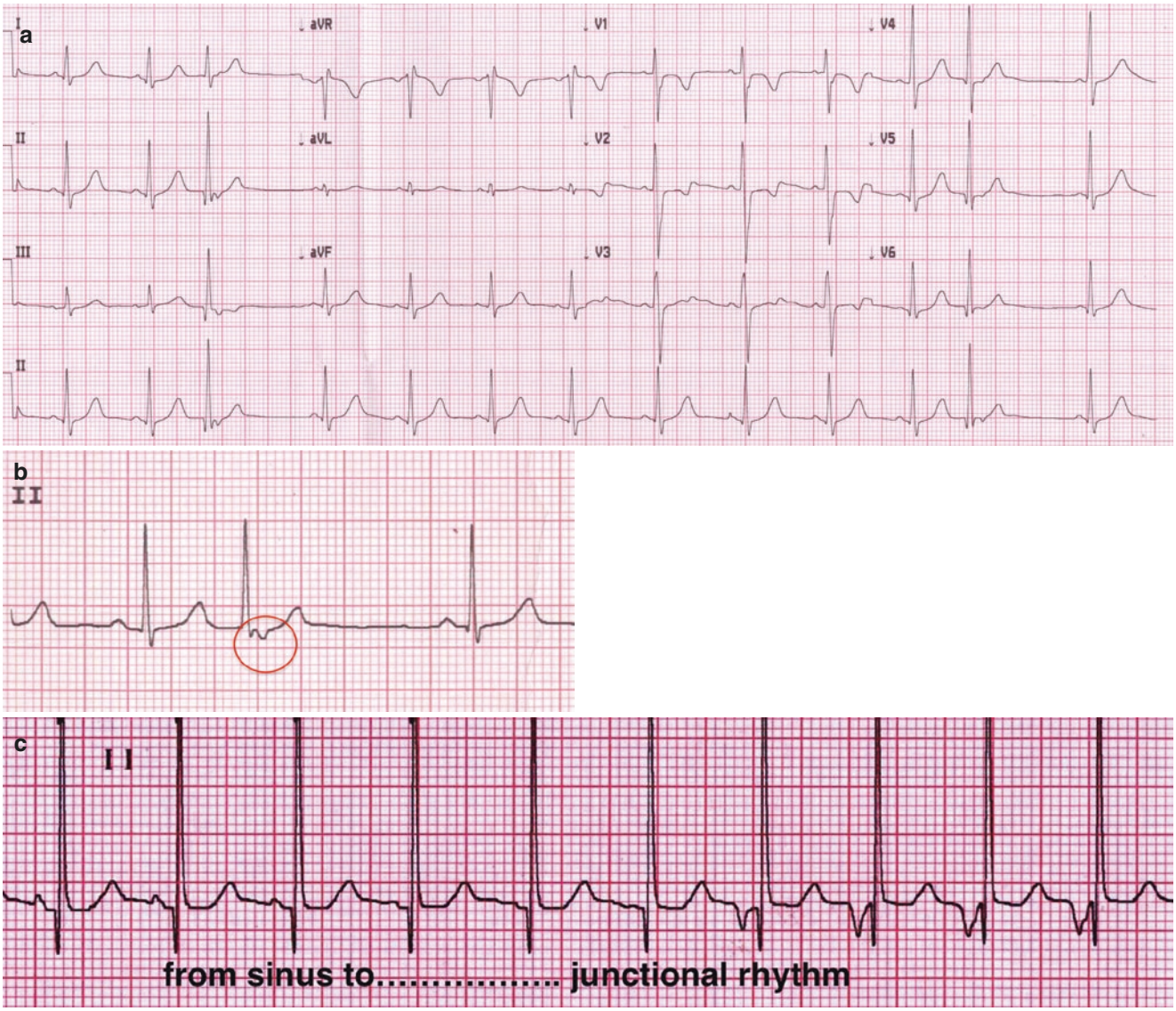

Fig. 11.19 (**a**), (**b**) Eight-year-old female with JPC. The P waves following the JPC are negative in the inferior leads and then must be retroconducted. (**c**) An example of low right atrium/junctional ectopic isorhythmic competitive rhythm; a shift from positive, isoelectric, and finally negative P waves can be noted

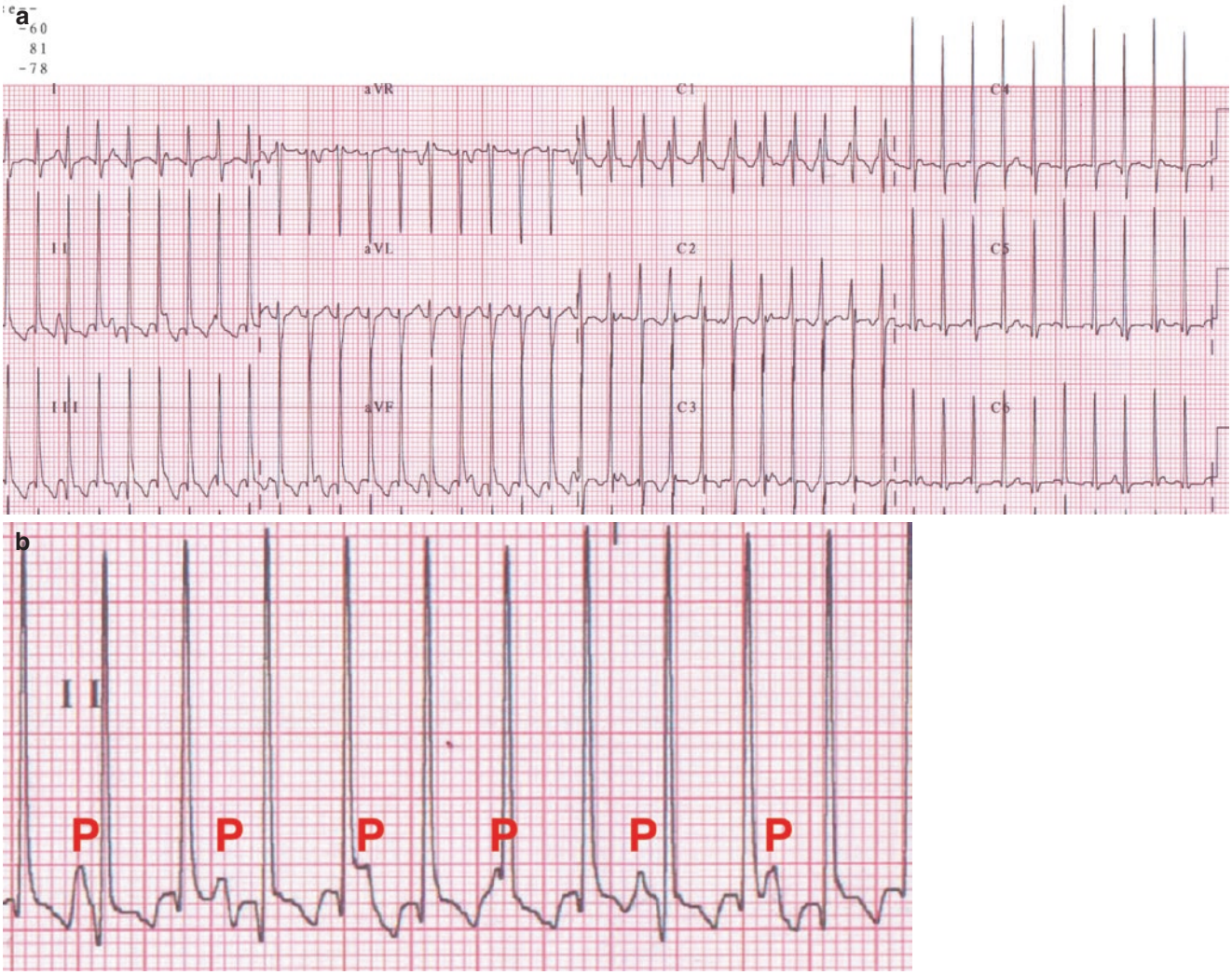

Fig. 11.20 (**a**) Two-year-old male admitted for heart failure. Large P waves are dissociated from the QRS and show atrial enlargement (J). The ventricular rate is 280 bpm. This is a JET, potentially lethal arrhythmia for fast and relentless character. Unlike a PJRT, which is usually slower and may evolve slowly to tachycardiomyopathy, an unrecognized JET can quickly lead to shock. (**b**) Close-up

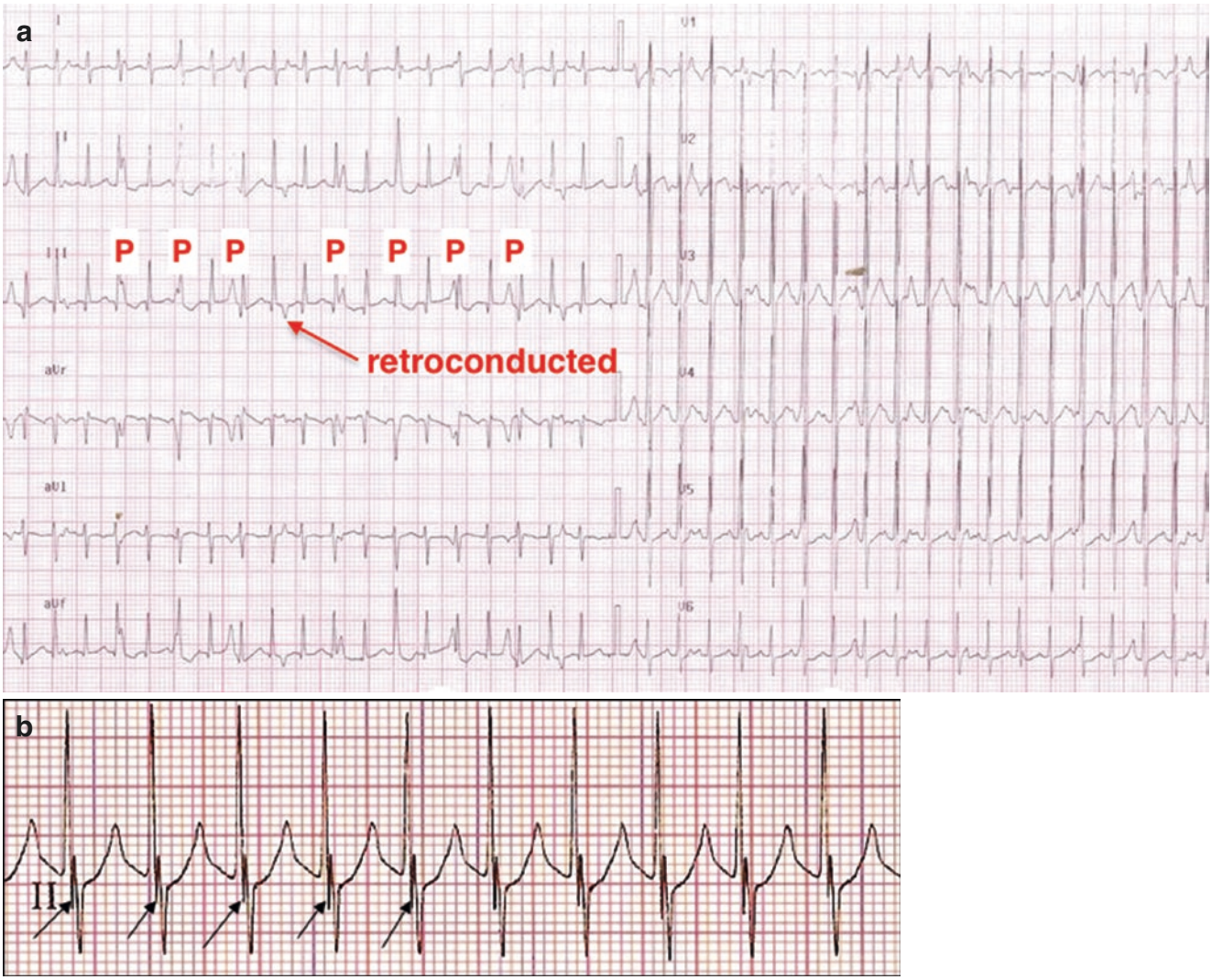

Fig. 11.21 (**a**) Another example of congenital JET. P waves are dissociated from the QRS (P); A single retroconducted P wave can be noticed (*arrow*). (**b**) An example of postoperative JET. The epicardial electrodes make it possible to record the atrial signal (*arrow*). The atrial wave is superimposed on the QRS and it is difficult to see it on the surface ECG

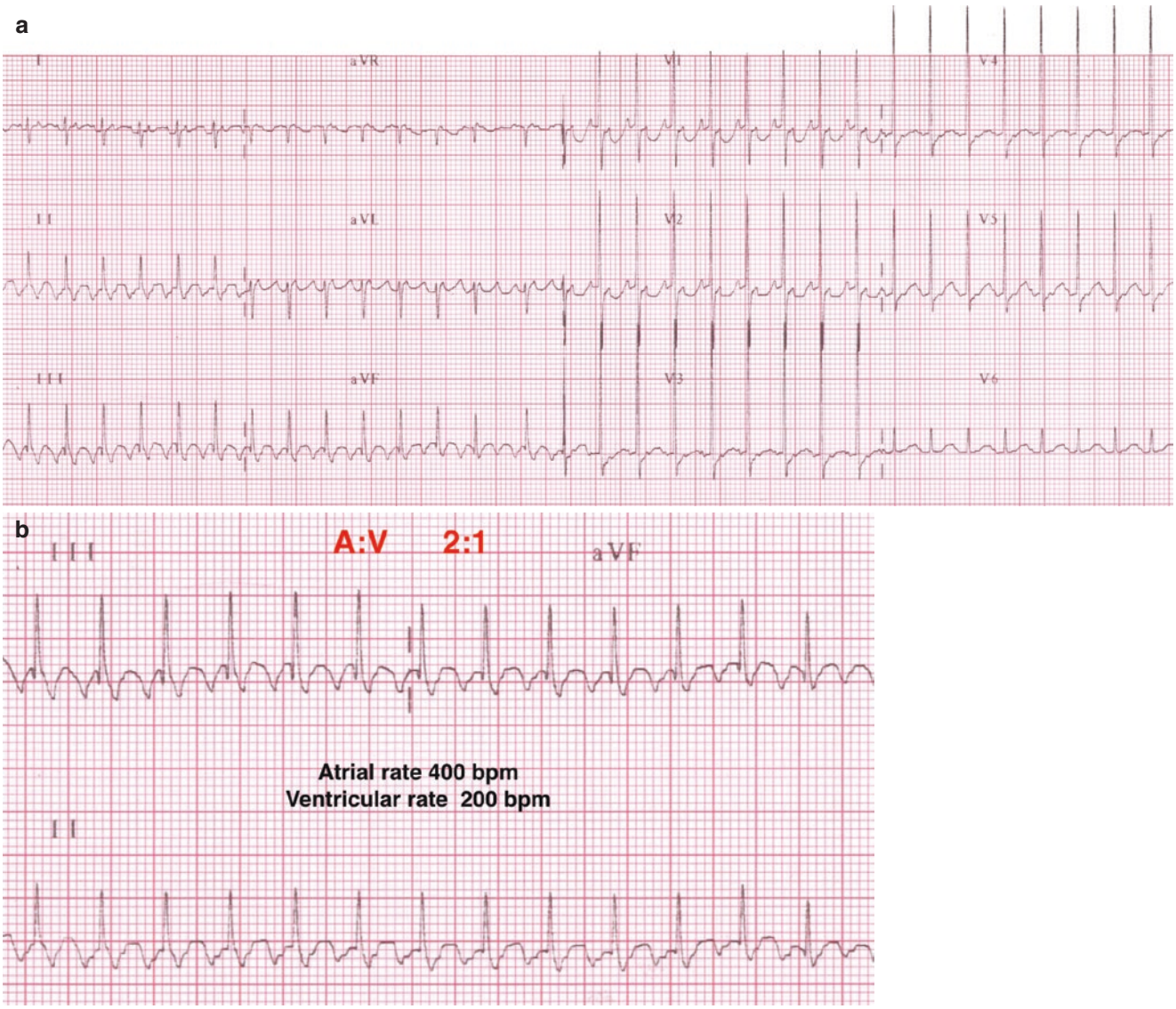

Fig. 11.22 (**a**) Neonatal atrial flutter. (**b**) Close-up: the A/V ratio is 2:1. The saw tooth F wave can be noted

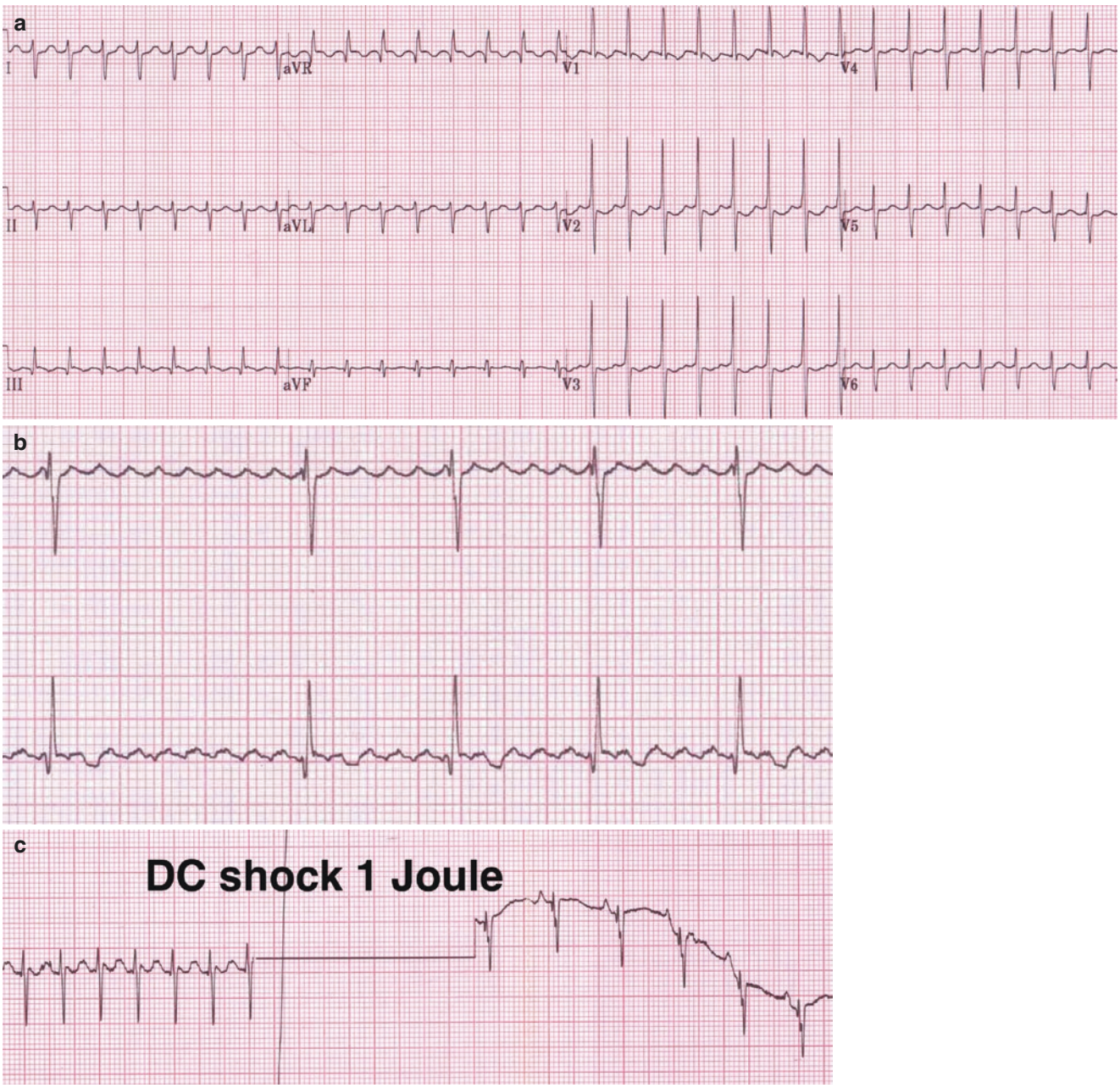

Fig. 11.23 (**a**) Neonate, 2 kg in weight, with atrial flutter. The ventricular rate is 200 (100 beats/kg), but in this case the P/F waves are not easy to see. (**b**) The diving reflex (ice pack) unmasked the F waves. (**c**) The DC shock was effective and cheap (1J only). A neonate with an HR not that fast, around 200 bpm, without a clear P wave should be suspected for atrial flutter. Due to the mass and the anatomy of a neonate's right atrium, the atrial rate of an atrial flutter is around 380–450 bpm, making 1:1 AV conduction impossible, while the 2:1 conduction is possible and common. An orthodromic reentrant tachycardia (the most common SVT of the neonatal age) is usually faster, in a range of 250–300 bpm. To discriminate between an EAT and an atrial flutter, the vagal maneuver or adenosine is very useful

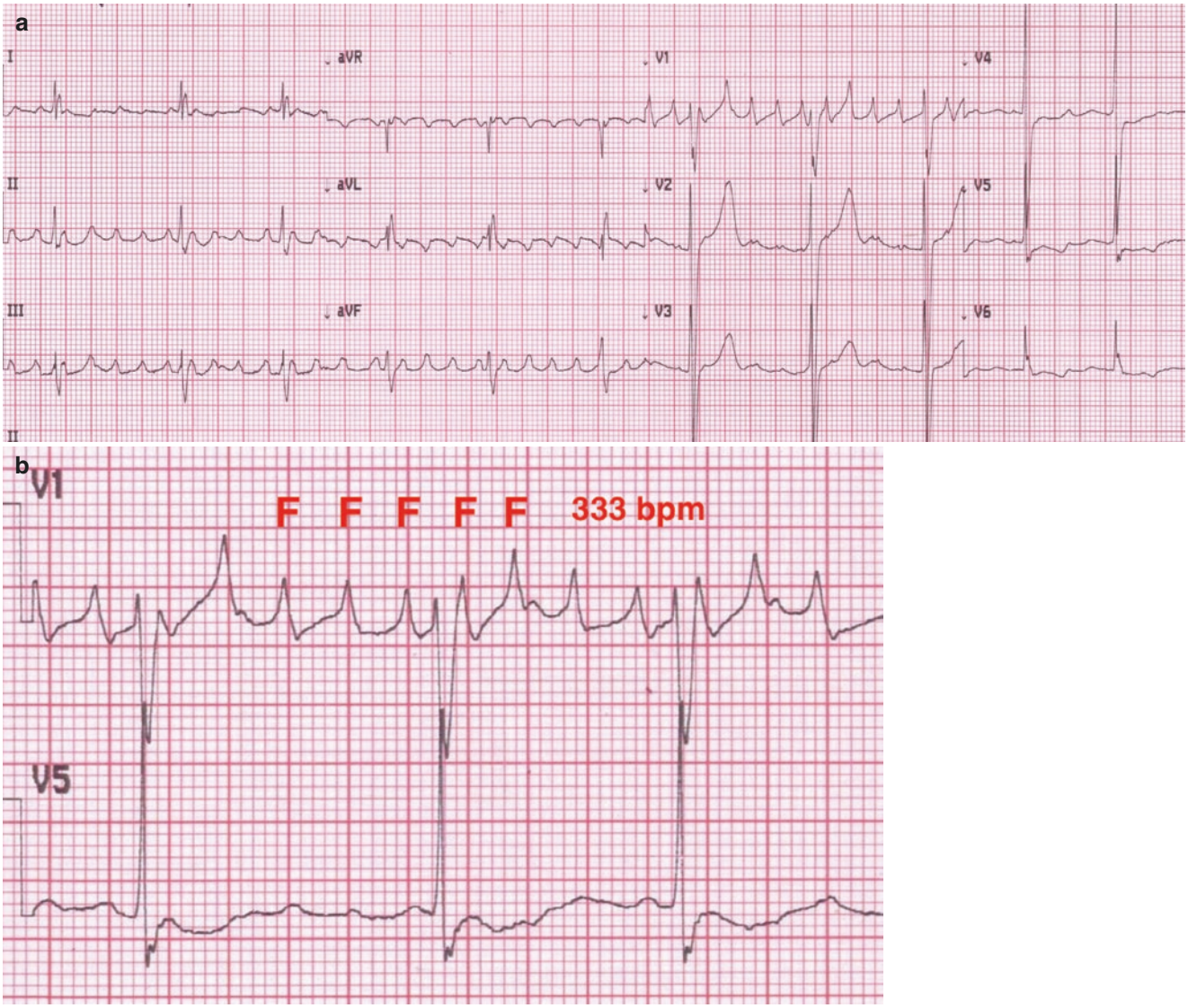

Fig. 11.24 (**a**) Twelve-year-old male with restrictive cardiomyopathy and chronic atrial flutter on treatment with beta blocker. The saw tooth waves are typically best seen in inferior leads and V1. The atrial rate is about 330 bpm. The prevalent conduction is 4:1. (**b**) Close-up

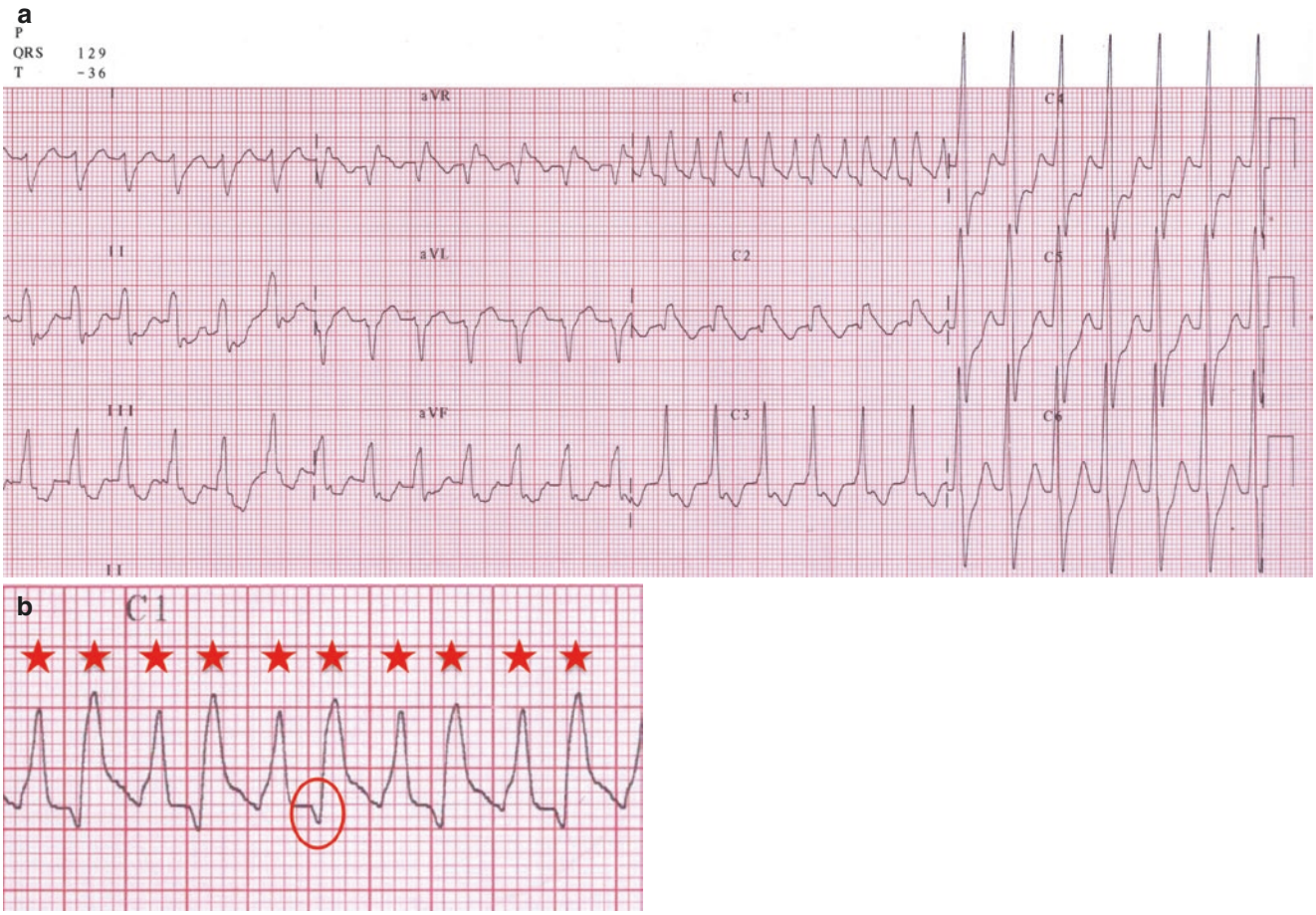

Fig. 11.25 (**a**) Thirty-year-old female, with large ASD and pulmonary hypertension in the early postoperative course of ASD closure surgery. There is a typical atrial flutter (Type I, counterclockwise reentry). The atrial rate (F waves) is typical for an adult atrial flutter, with 2:1 A/V conduction. A Q wave in V1 is present (*circle*), indicative of severe RVH. (**b**) Close-up

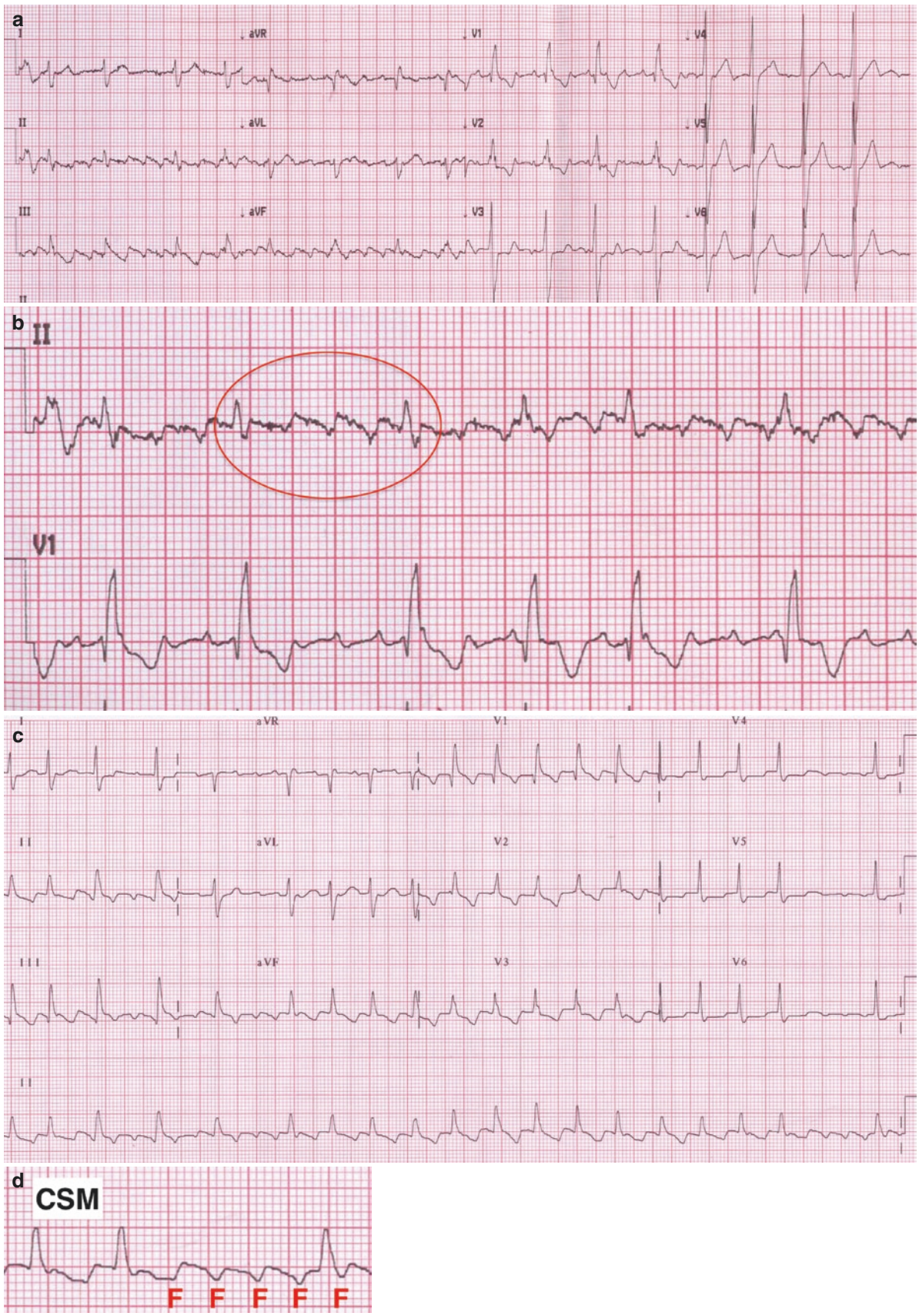

Fig. 11.26 (**a**), (**b**). Atrial flutter in a 16-year-old male tetralogy of Fallot repaired in infancy. Type I A, a typical incisional arrhythmia clockwise reentry. Classic atrial flutter F waves 300 bpm. (**c**) Incisional flutter type I in a 60 years old female who underwent ASD repair. (**d**) The carotid sinus massage unmasked the F waves

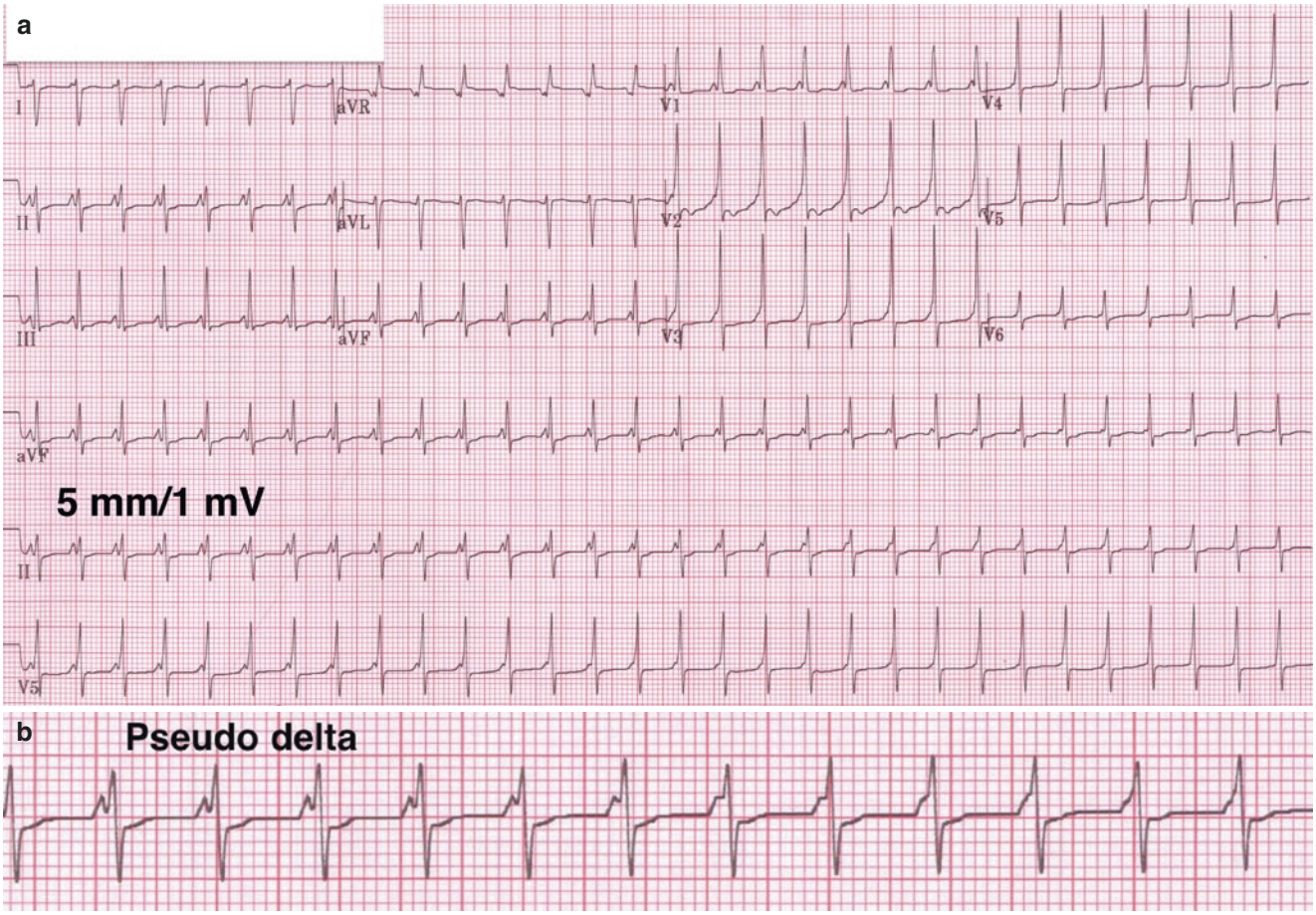

Fig. 11.27 (**a**) One-month-old baby boy with SVT. The HR is 185 bpm. The apparent sinus tachycardia with ventricular pre-excitation (pseudo delta) is caused by the isorhythmic overlap between the JET and the dissociated sinus tachycardia, the latter due to the JET-induced heart failure. Note the atrial enlargement typical of JET and other incessant arrhythmias with AV dissociation. (**b**) Close-up

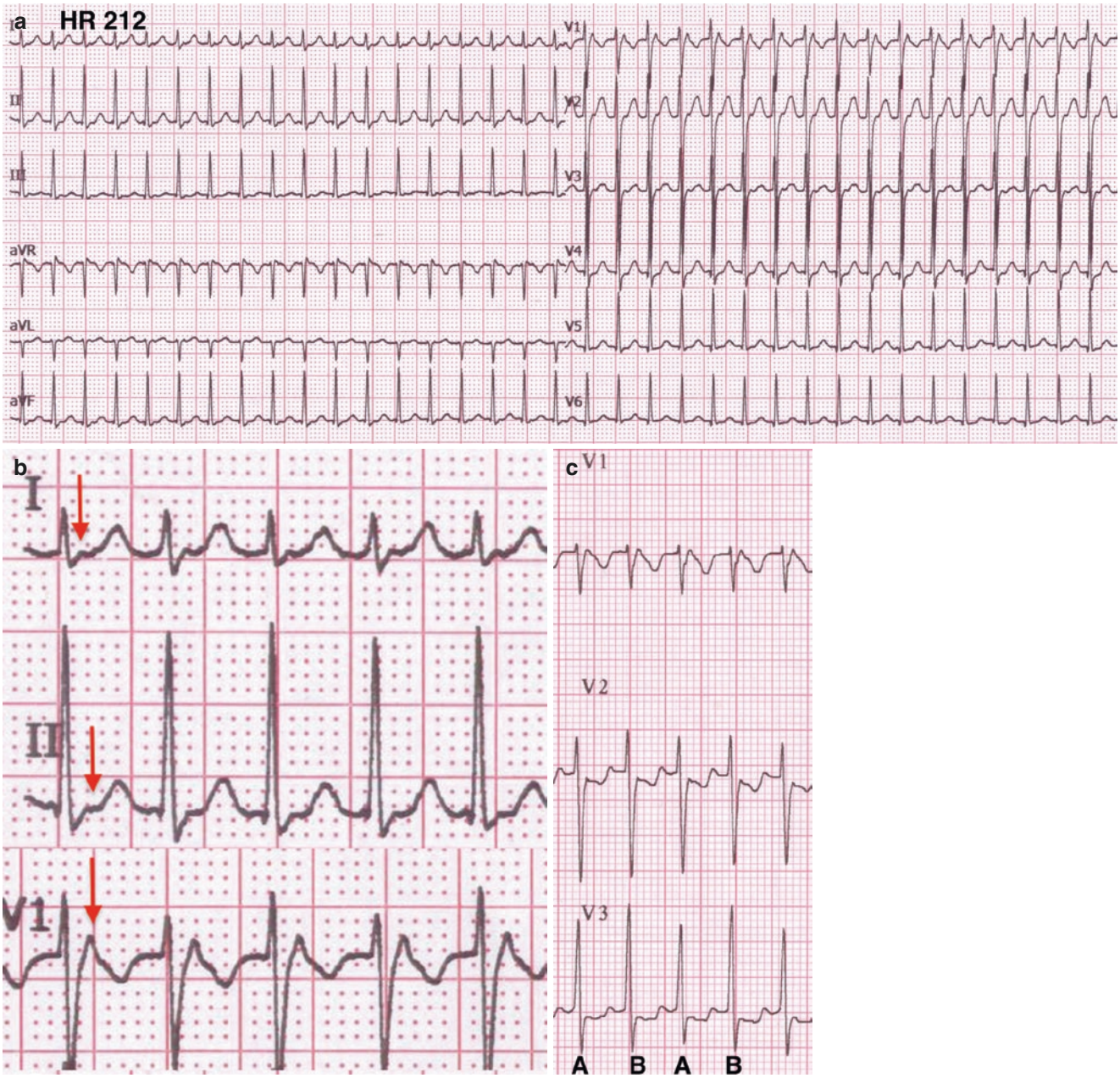

Fig. 11.28 (**a**) Eight-year-old male with AVNRT. There is a palindromic HR, 212, as befits a reentrant tachycardia. The RP interval is <80 ms, as is clear in V1 (pseudo r'). (**b**) Close-up. (**c**) Another example of pseudo r' in AVNRT

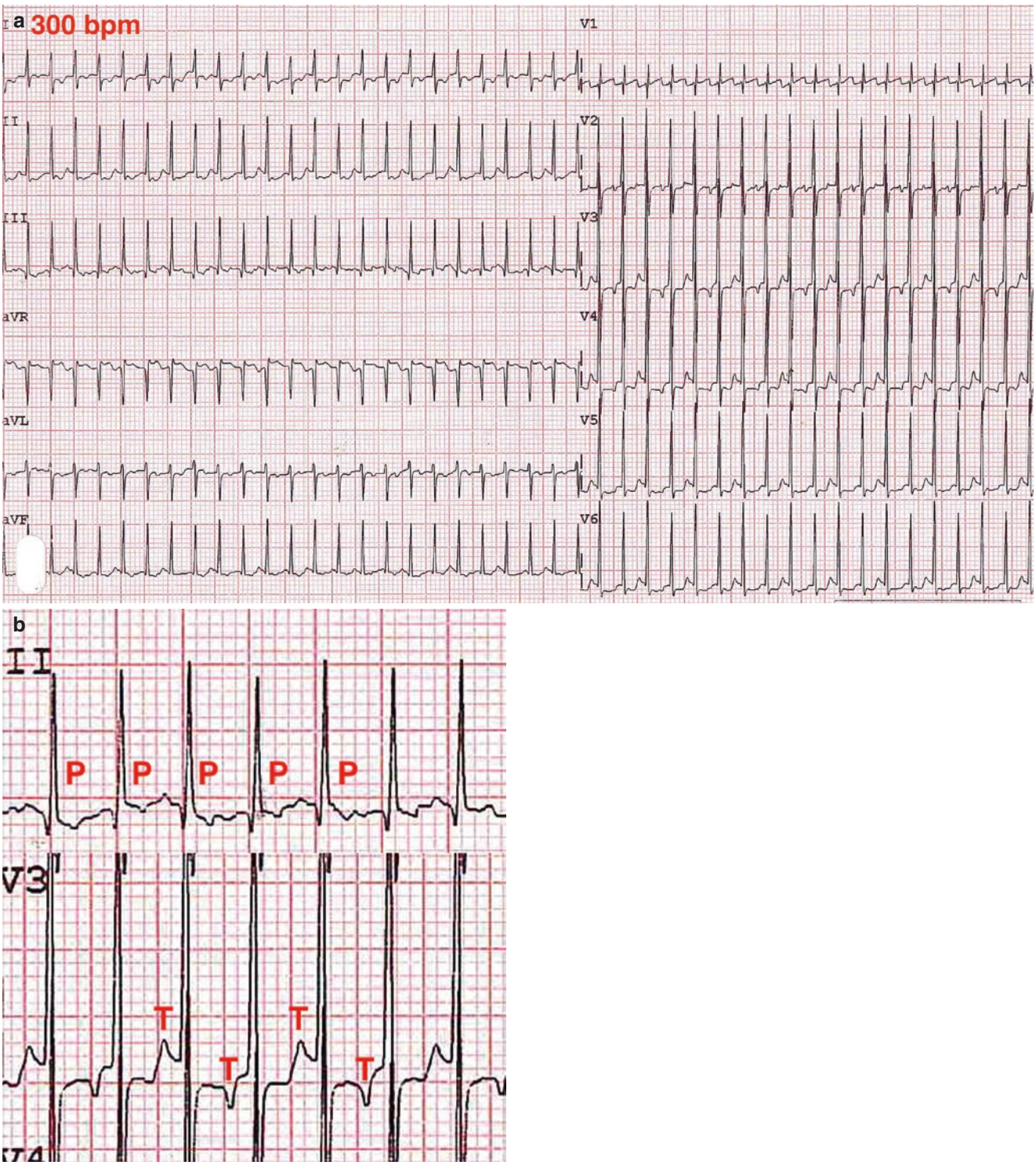

Fig. 11.29 (**a**) Three-month-old female. Typical orthodromic reentry from an occult accessory pathway (normal basal ECG, without pre-excitation). The arrhythmia has an HR of about 300 bpm. A P wave with RP < PR is clearly seen in lead III. The RP interval is 80 ms, too short for misunderstanding the P for the T wave. In front of the R wave in V3, there is a wave with an apparent sinus position (or compatible with automatic atrial arrhythmia). It is actually a T wave which presents the phenomenon of electrical alternance (positive/negative). (**b**) Close-up

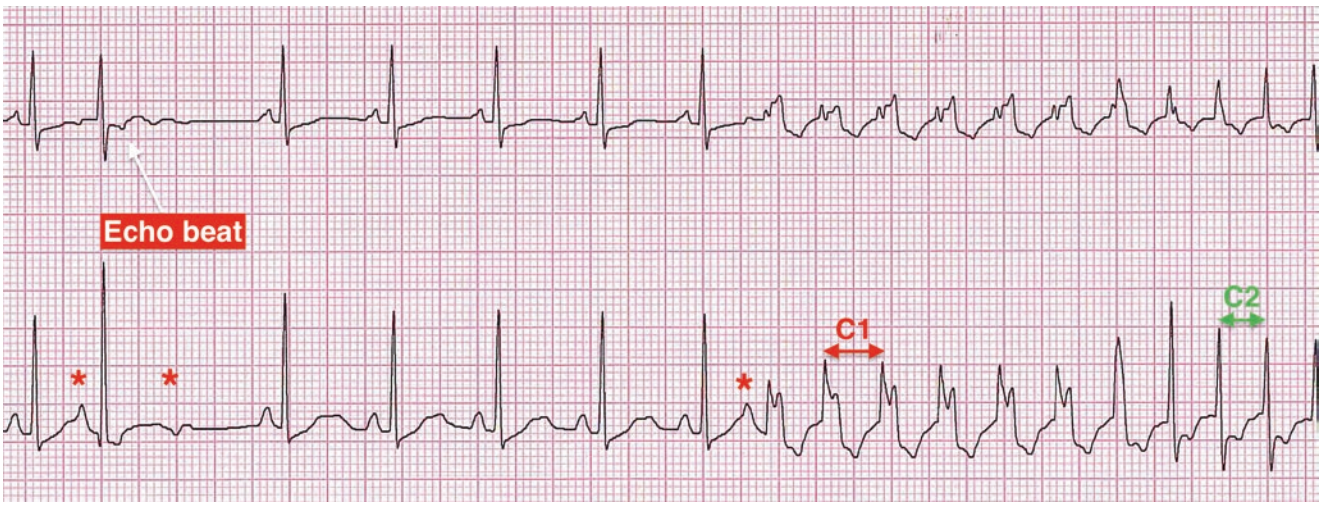

Fig. 11.30 Same patient Fig. 11.29. The arrhythmia recurred after adenosine, owing to frequent ectopic beats that trigger the reentry. In the third beat from the left, a PAC (*) intervenes but PSVT does not start (there is only an atrial echo beat), even with the next PAC (*) which does not pass from the node or the accessory pathway. A third PAC triggers a reentrant tachycardia with wide complex tachycardia in the first six beats owing to a type LBBB aberrant conduction; the QRS then narrows, after a mid-width hybrid beat. Furthermore, the cycle of the narrowest ventricular complexes is shorter, so that the arrhythmia is more rapid (C2). This aspect is compatible with a left accessory pathway. In a reentrant PSVT supported by an accessory pathway that has the same side as an "aberrant" branch, the additional time due to conduction delay will be added to the arrhythmia cycle. An arrhythmia which manifests firstly with a wide and then with a narrow complex is almost always the same arrhythmia which runs through a "cold" branch in the beginning, and it is conducted with aberrancy. When the aberrancy is sustained, we call this "linking phenomenon." There are rarely two different arrhythmias overlapping [12]

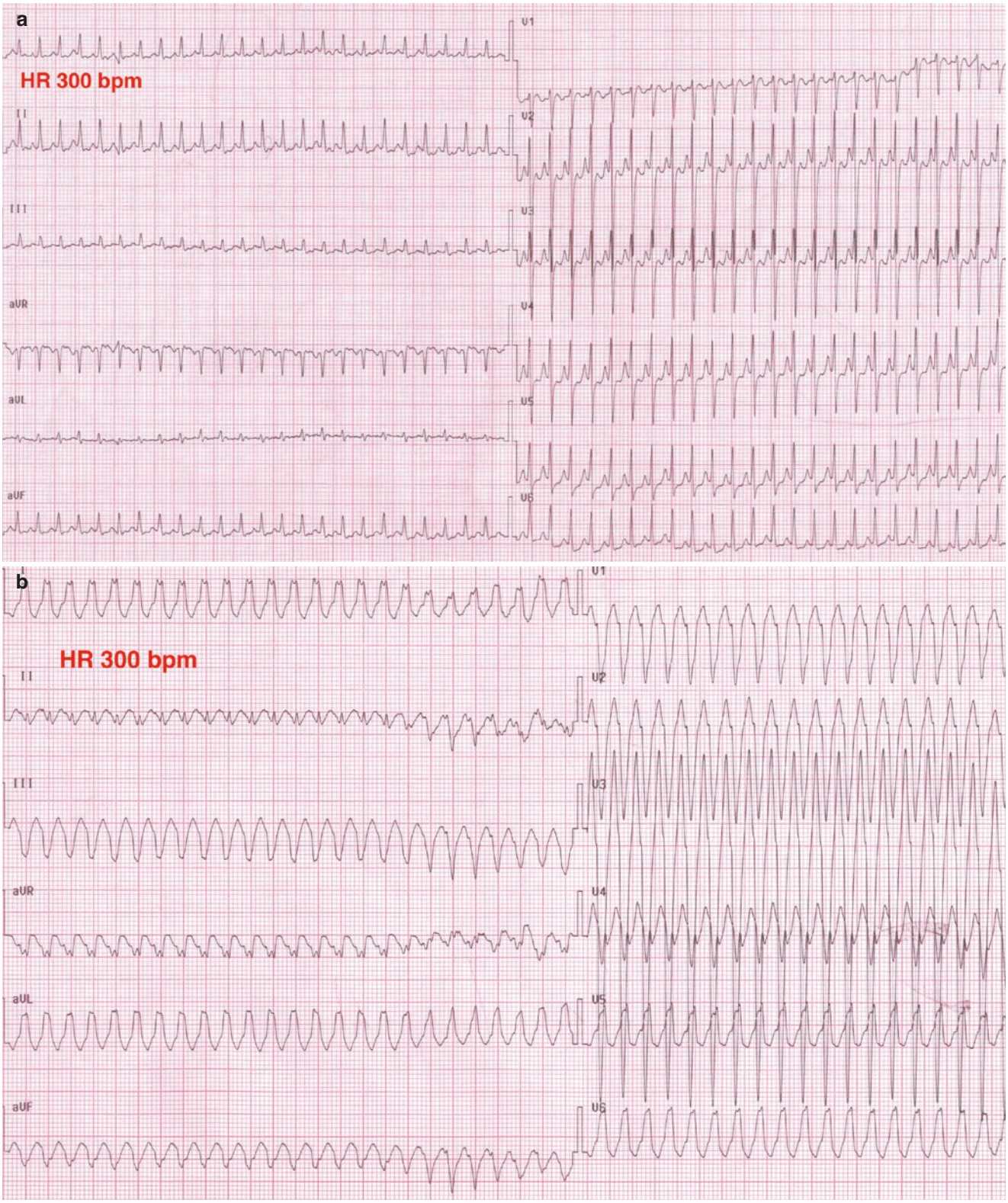

Fig. 11.31 (**a**) Six-month-old baby boy with PSVT. The narrow complex tachycardia resolves after an adenosine bolus. (**b**) Immediately after the conversion to sinus rhythm, a wide complex tachycardia starts. It is not a VT or an antidromic AVRT but the same initial PSVT with aberrant conduction (linking phenomenon). The HR is the same as the narrow one, 300 bpm, and there is an LBBB. For the same reason as the previous case, if the LBBB aberrancy does not affect the cycle length, the accessory pathway should be right sided or septal, anything but left sided

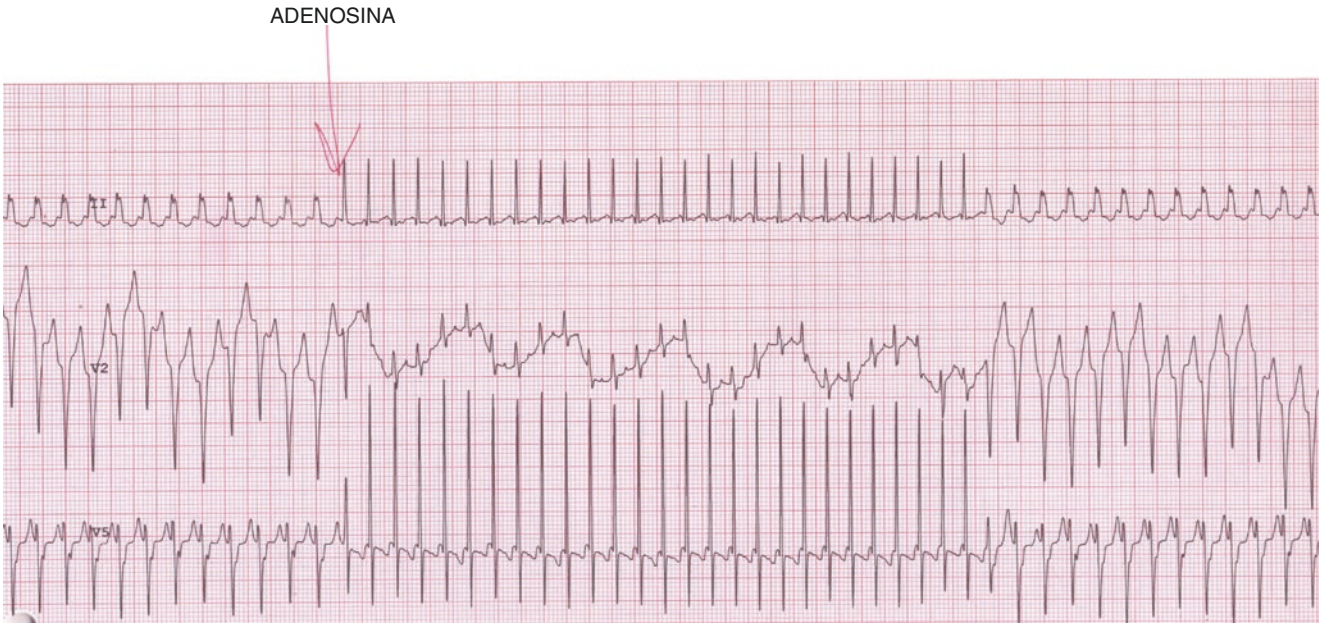

Fig. 11.32 A case similar to the previous one with wide-narrow-wide complexes shift during adenosine bolus

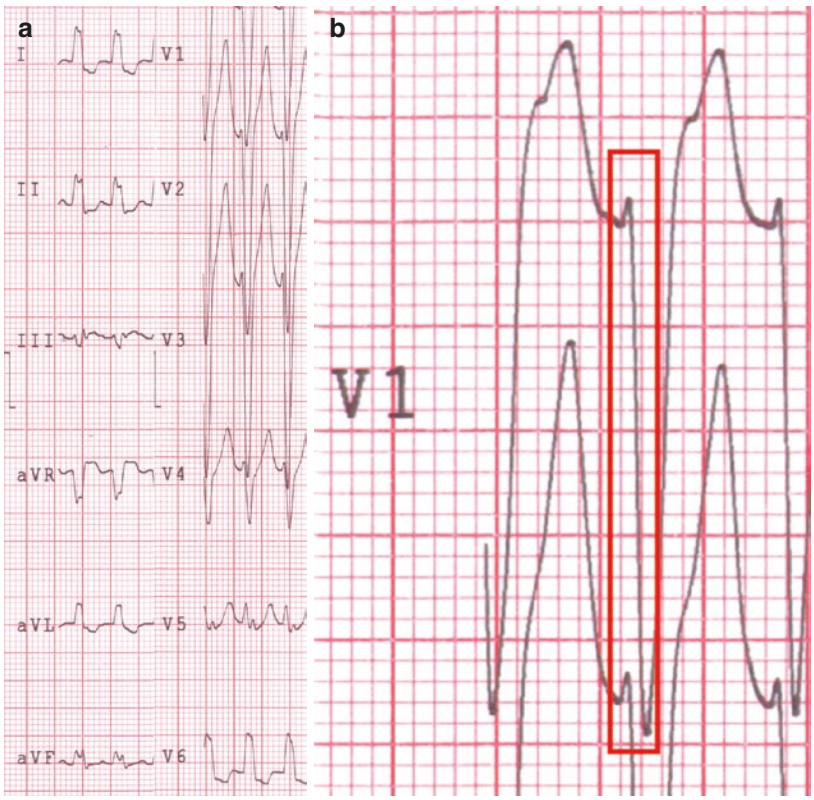

Fig. 11.33 (**a**) Ten-year-old male, first episode of tachycardia. Wide complex arrhythmia: 215 bpm, LBBB pattern. No dissociation, fusion, or retrograde conductions are seen. The differential diagnosis between a PSVT with aberrant conduction and a VT has to be purely morphological (clinically unresponsiveness to vagal maneuvers). The classic LBBB appearance directs us toward a PSVT with aberrant conduction. The rapid fall of the S wave in V1 (less than 1 mm from the nadir, less than 40 ms) further confirms this diagnosis. (**b**) Close-up. The rapid intrinsicoid deflection also rules out an antidromic AVRT

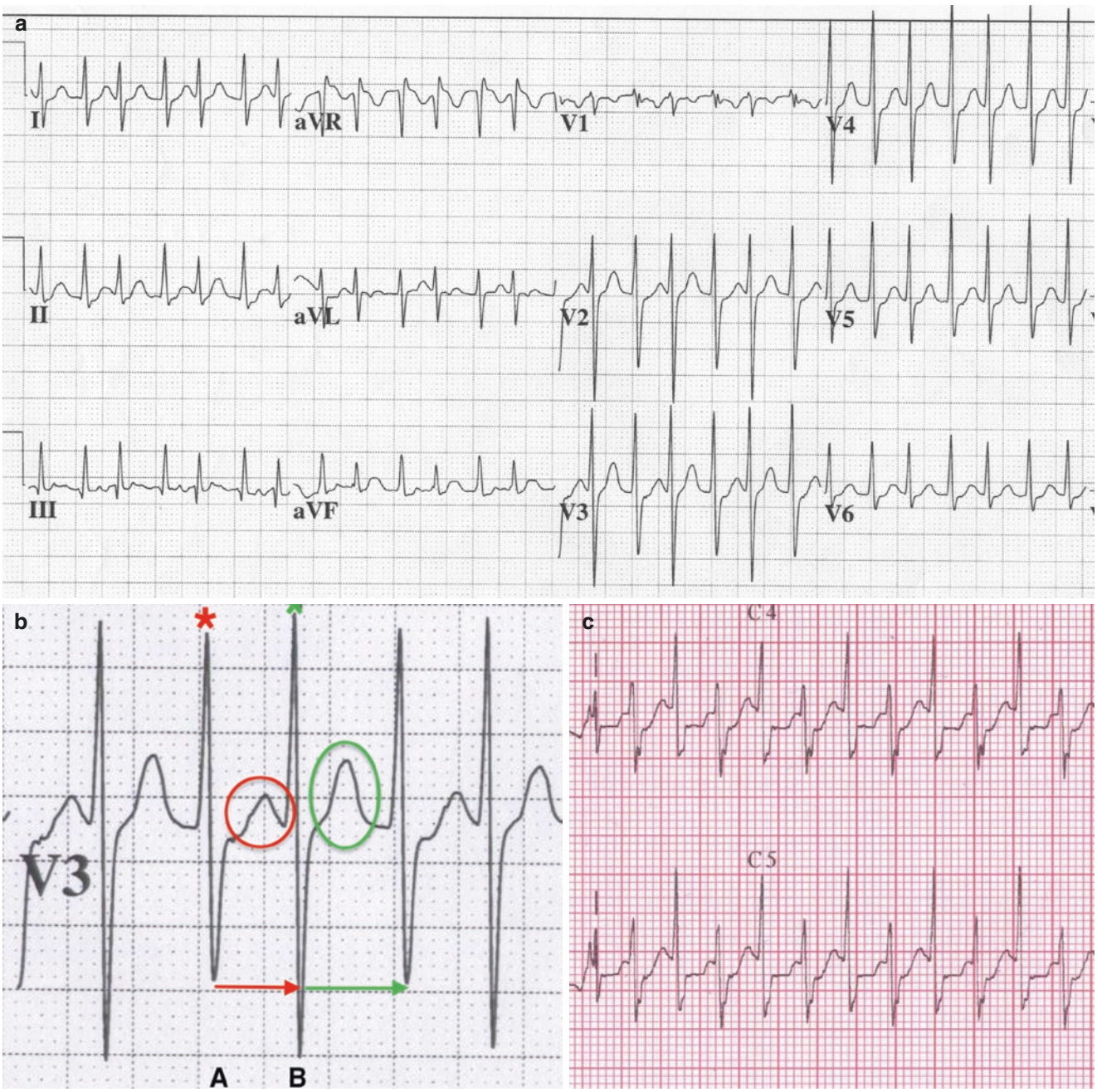

Fig. 11.34 (**a**) Eleven-year-old male with anteroseptal AP and paroxysmal AVRT. (**b**) There is a multiple alternance: QRS morphology, cycle length and T wave are beat-to-beat alternating. The reason is an intermittent aberrancy of conduction; another explanation may be an anterograde conduction alternating between the fast and the slow AV nodal pathway. (**c**) Another example of alternance

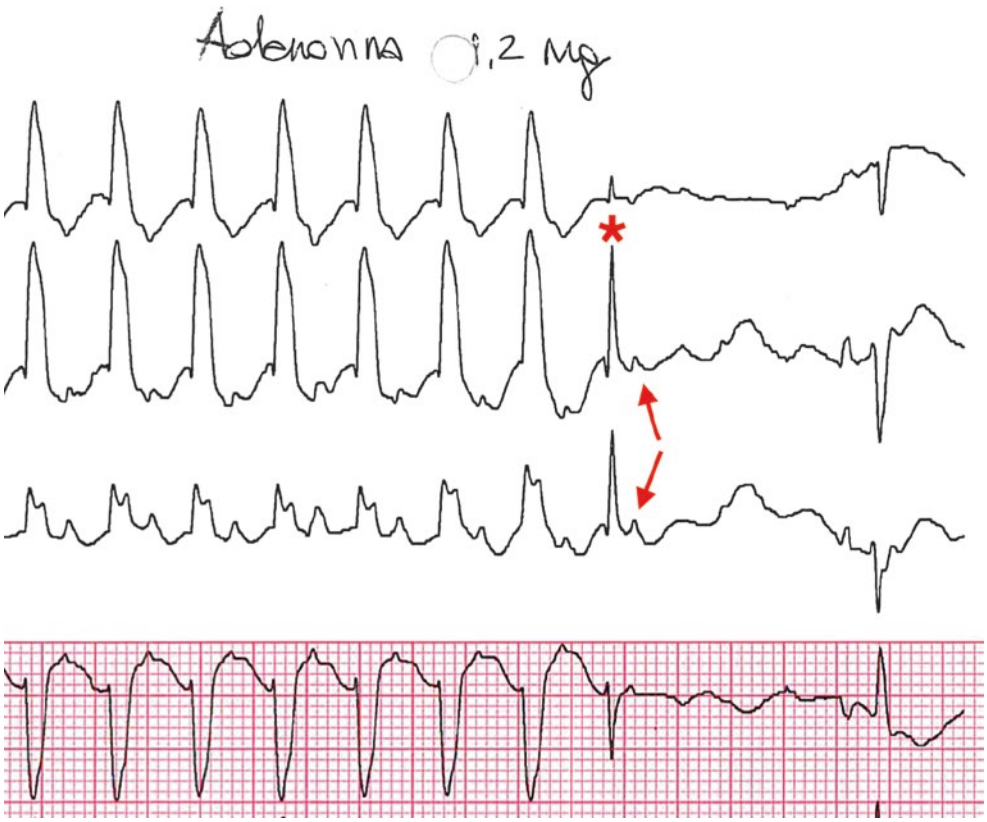

Fig. 11.35 Another case of AVRT in an infant with wide complex tachycardia due to linking phenomenon. Before adenosine stops the arrhythmia (the arrows mark the agonic P wave which will crash in the "drunk" AV node), there is a narrow complex (*). So adenosine is able to interrupt the linking phenomenon, and at the same time it, is able to induce it (see Fig. 11.25), probably working on the differential refractoriness of slow and fast AV nodal pathways

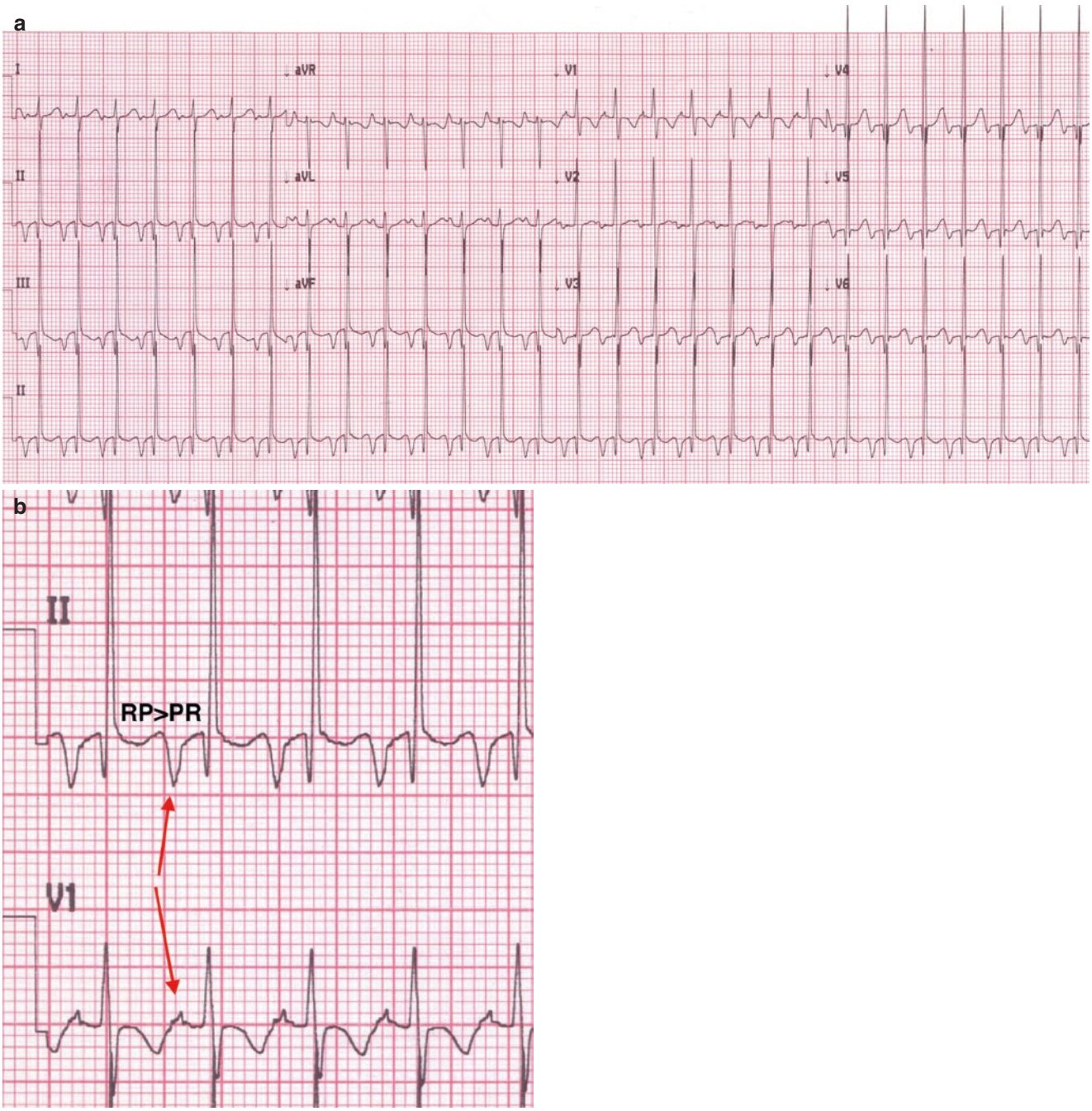

Fig. 11.36 (**a**) Six-year-old male. Typical morphology of P (negative waves in the inferior leads, positive in aVR and aVL) in an apparently "sinus" position, not too fast (170 b/min), and relentless character are the typical features of PJRT otherwise called "Coumel." (**b**) Close-up. In this orthodromic reentrant tachycardia, the V > A conduction through a slow accessory pathway causes an RP > PR interval

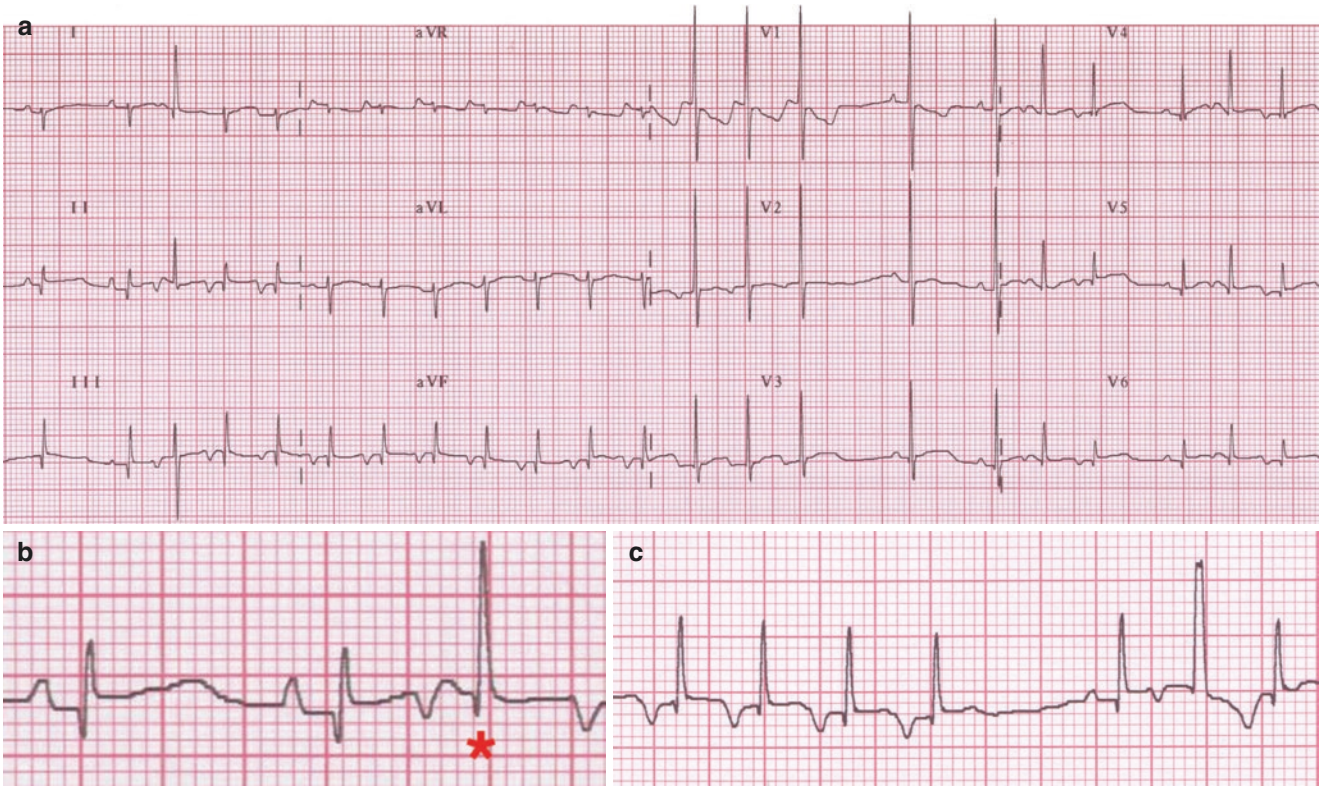

Fig. 11.37 (**a**) Another example of PJRT. This tachycardia is incessant by nature. After a short sinus rhythm, the arrhythmia starts with a relatively wide QRS complexes (*), owing to the aberrant conduction caused by the "relaxation" of branches during the long cycle (Ashman phenomenon). (**b**), (**c**) Close-up [13, 14]

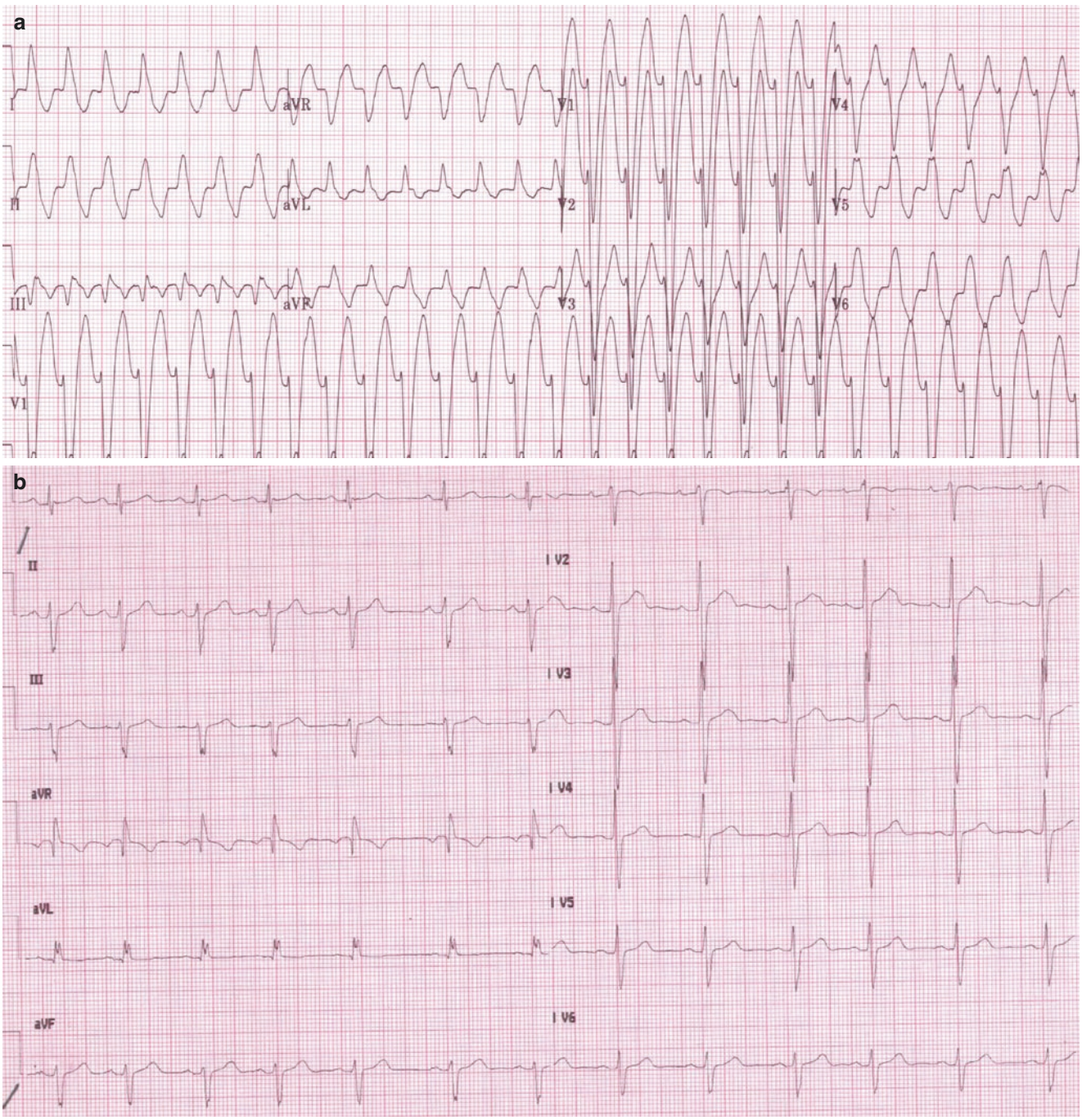

Fig. 11.38 (**a**) Thirteen-year-old male with wide complex tachycardia with LBBB morphology. Many differential diagnoses can be contemplated, from VT, to aberrancy, to preexistent LBBB. (**b**) The young man was a carrier of Mahaim fibers. The basal ECG reveals the suggestive features of rS pattern in lead III and the absence of Q waves in V6. Mahaim tachycardia is a form of antidromic tachycardia, and because of the right-sided atriofascicular connection, and although it is rare, it should always be taken into consideration in the case of wide complex tachycardia with LBBB pattern

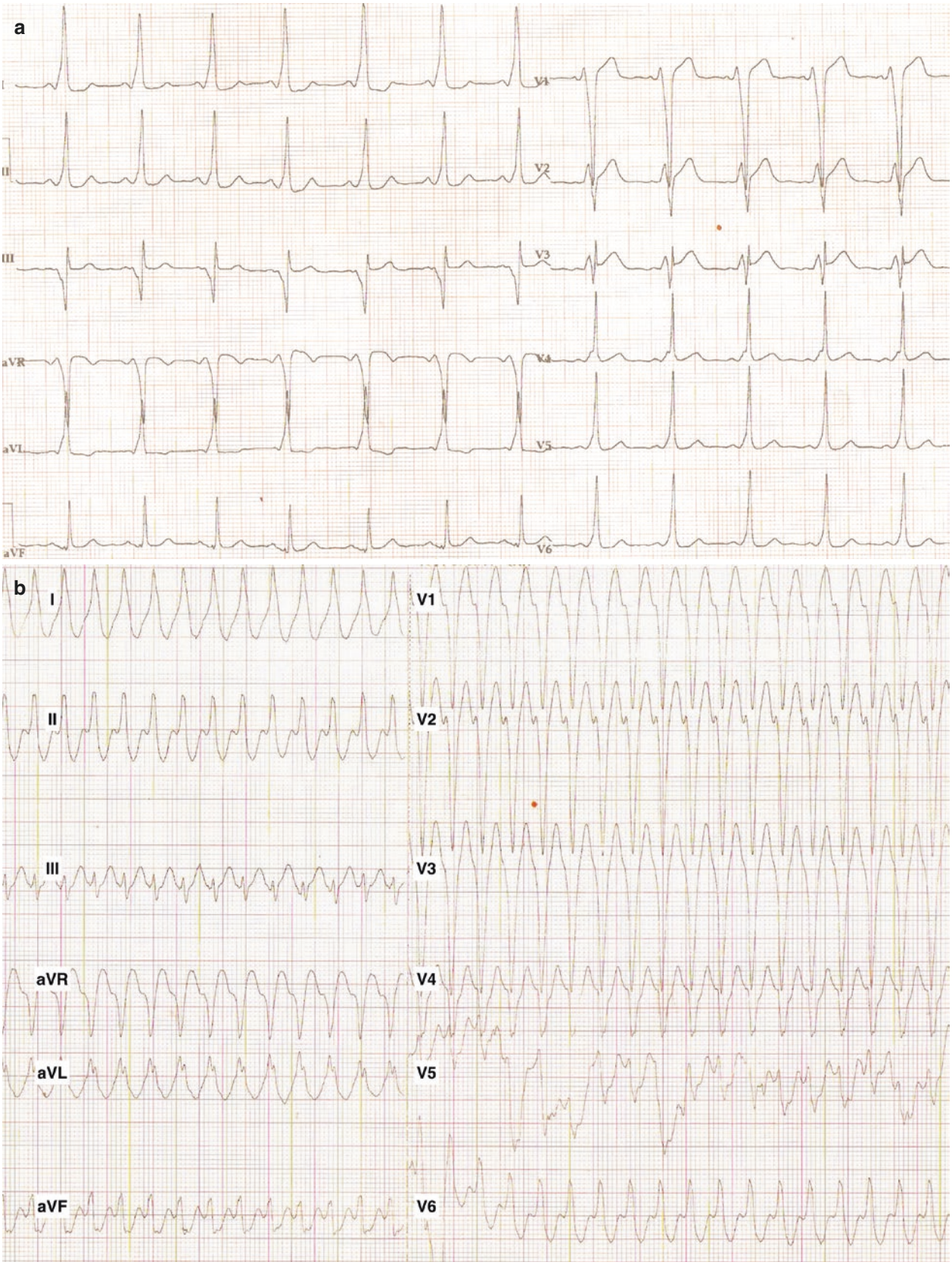

Fig. 11.39 (**a**) eleven-year-old male with WPW syndrome. He is a carrier of a right posterior AP. (**b**) The clinical SVT shows wide complexes and the QRS are similar to the pre-excited sinus beats. This is an example of the rare antidromic AVRT (the wavefront runs from atrium to ventricle trough the accessory pathway and go back to the atrium trough the AV node), which represents less than 5% of AVRTs (courtesy of Dr Enrico Rosati)

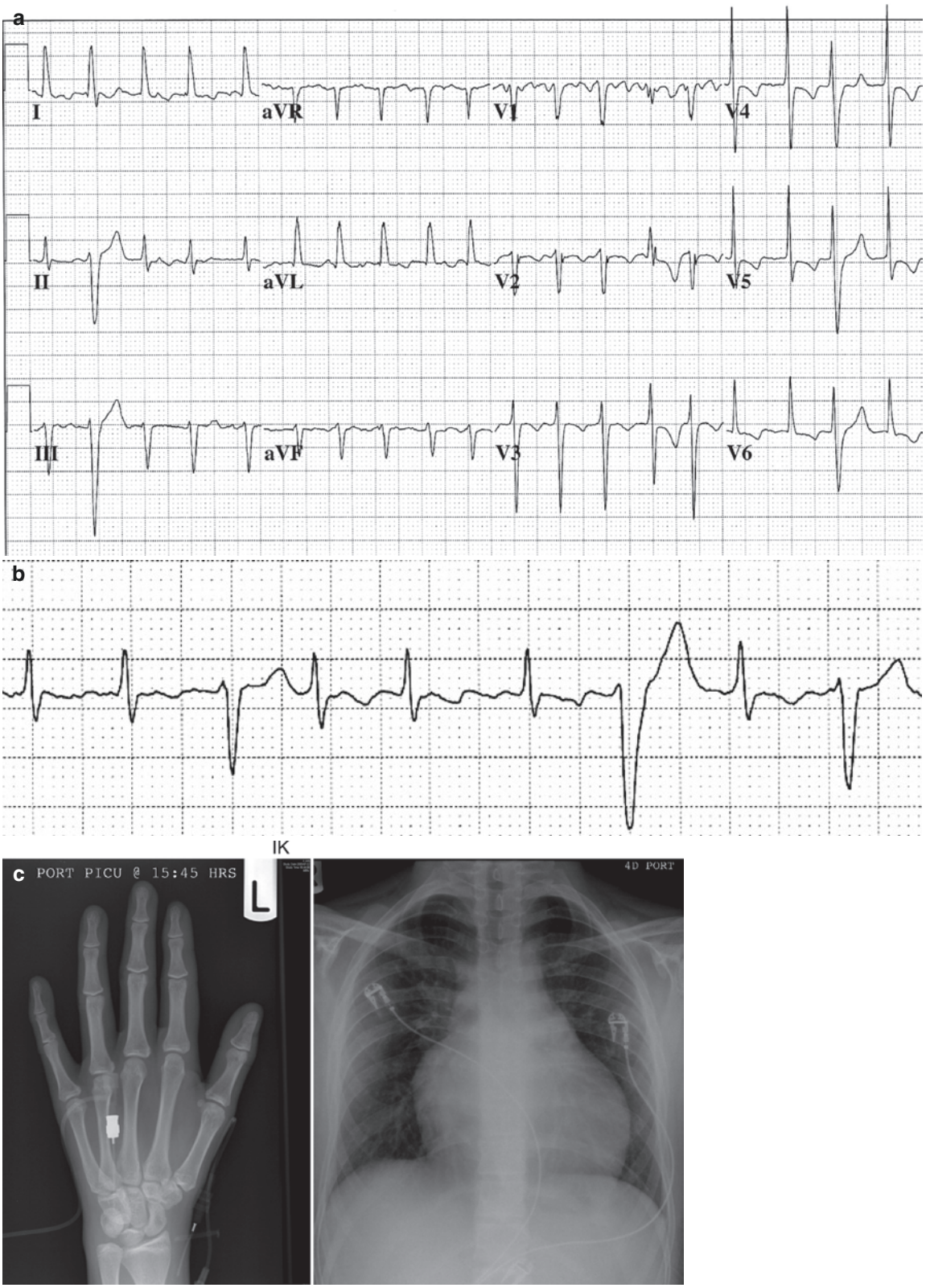

Fig. 11.40 Four-year-old male with a family history of dilated cardiomyopathy and life-threatening arrhythmias. Some family members were diagnosed as carriers of heart-hand syndrome, an association of CHD, DCM, and limb deformities. The HHS is genetically heterogeneous; in this case a mutation in the LMNA gene encoding A-type lamins was found. Very early AF should always be considered carefully, looking for laminopathy or channelopathies. (**a**) The ECG shows an atrial fibrillation: no P waves are recognizable. (**b**) Close-up: Ashman phenomenon. (**c**) Chest and hand X ray

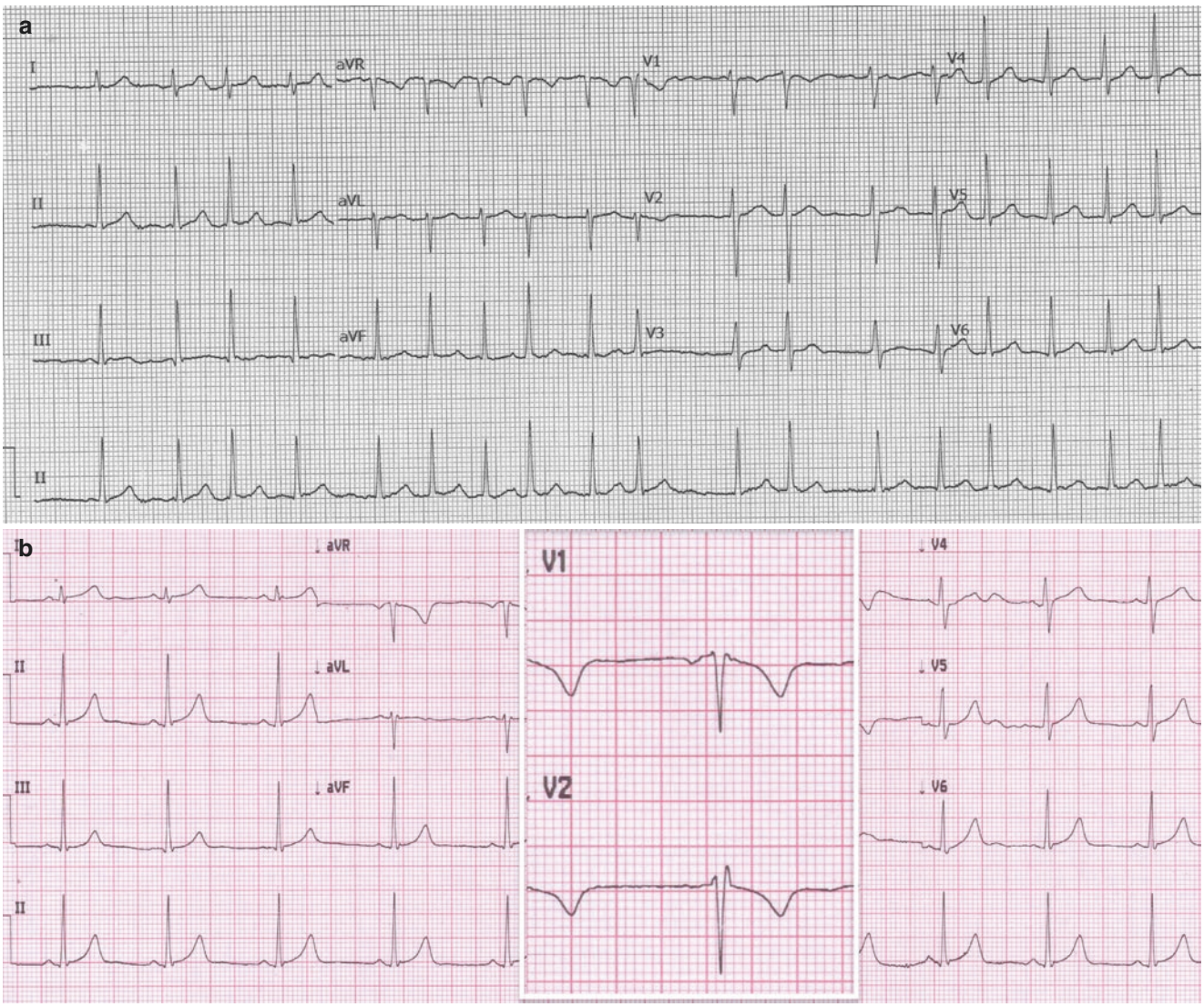

Fig. 11.41 Sixteen-year-old female with family history of early AF (before 20 years of age). (**a**) First episode of AF; (**b**) the baseline ECG is normal, but the follow-up will be tight especially on the precordial leads, knowing the association between Brugada syndrome and AF

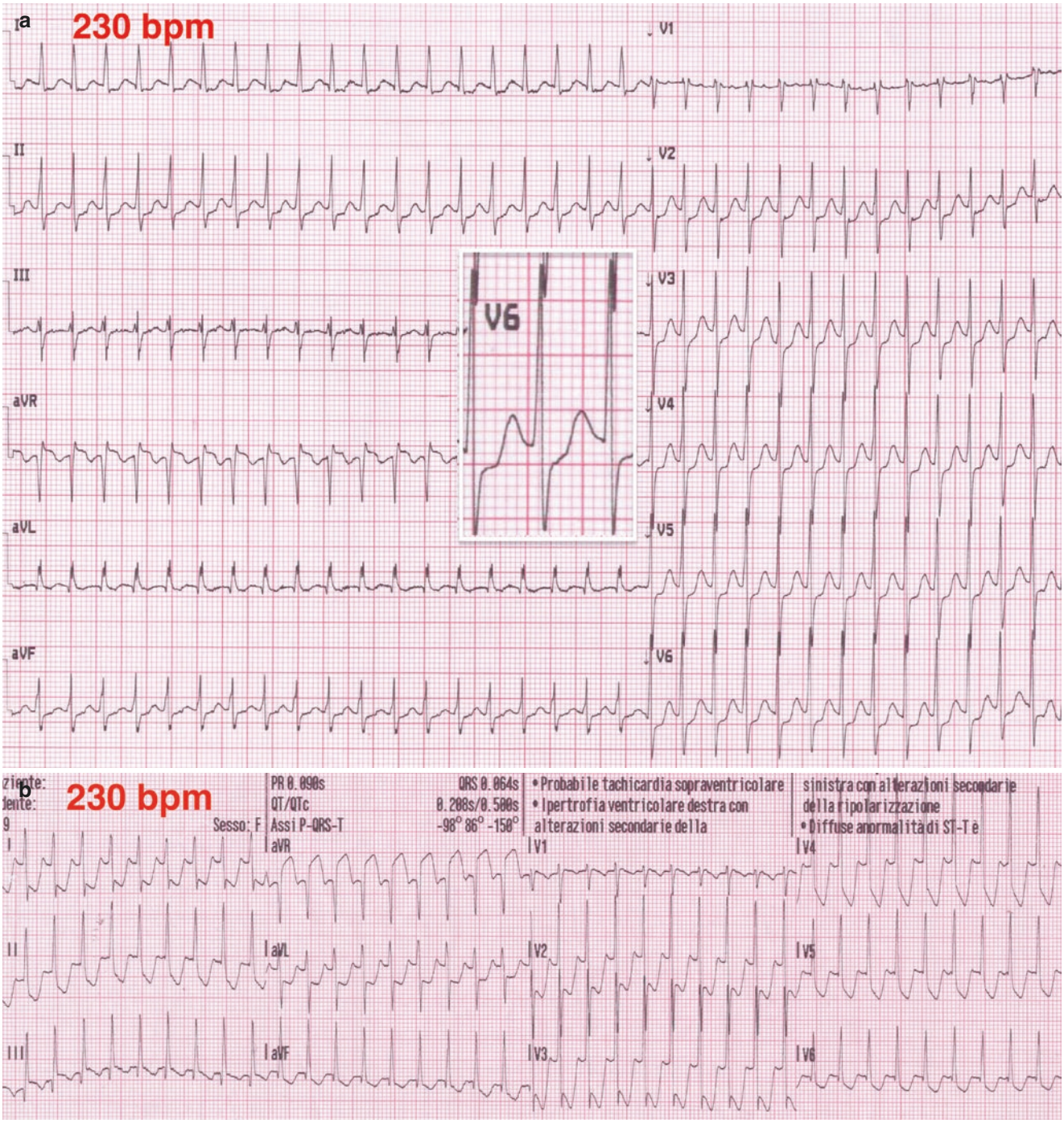

Fig. 11.42 Two females, 7 years (**a**) and 9 years (**b**) old, with SVT. Both SVTs are characterized by ischemic ST changes. In the second case, the patient complained of chest pain. Nonspecific ST-T changes are well described in SVT. Here it seems a real transient subendocardial ischemia due to the fast rate, even in the absence of coronary heart disease as was proven in this case with a CT scan. Myocardial injury markers were negative, and soon after the PSVT the ECG was normal

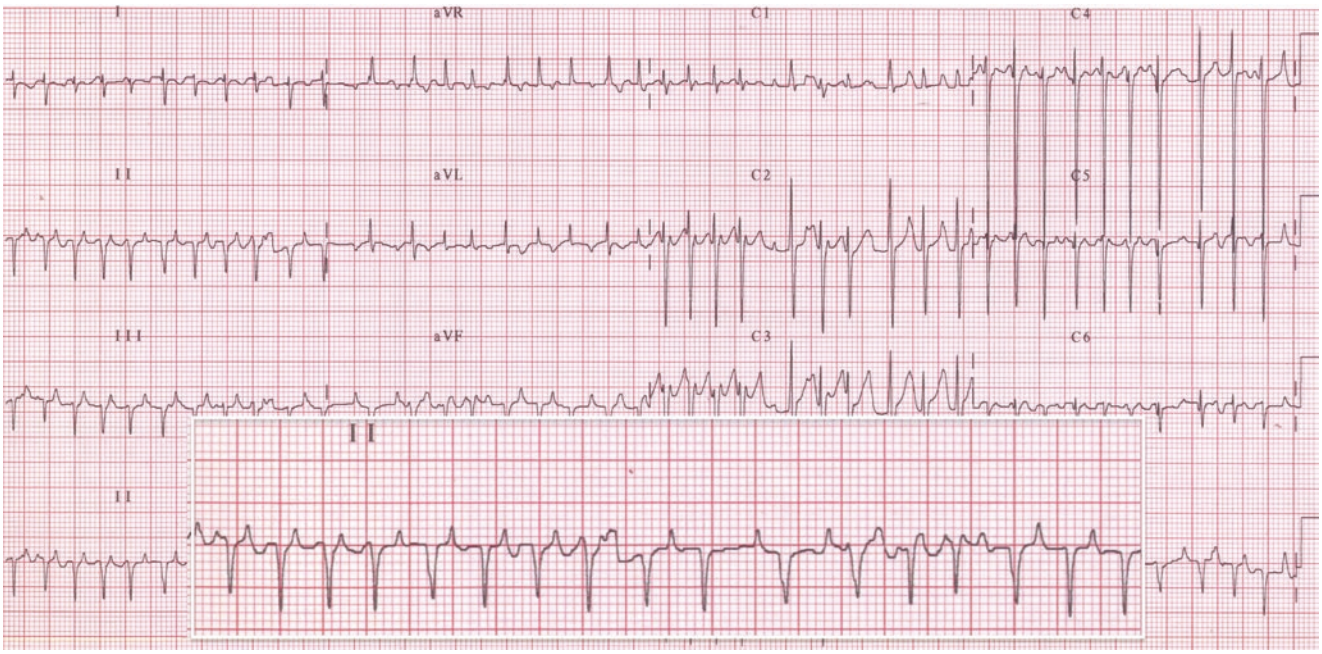

Fig. 11.43 Three-year-old female with Costello syndrome, HCM, and aortic coarctation. Multifocal atrial tachycardia, at least three foci of EAT

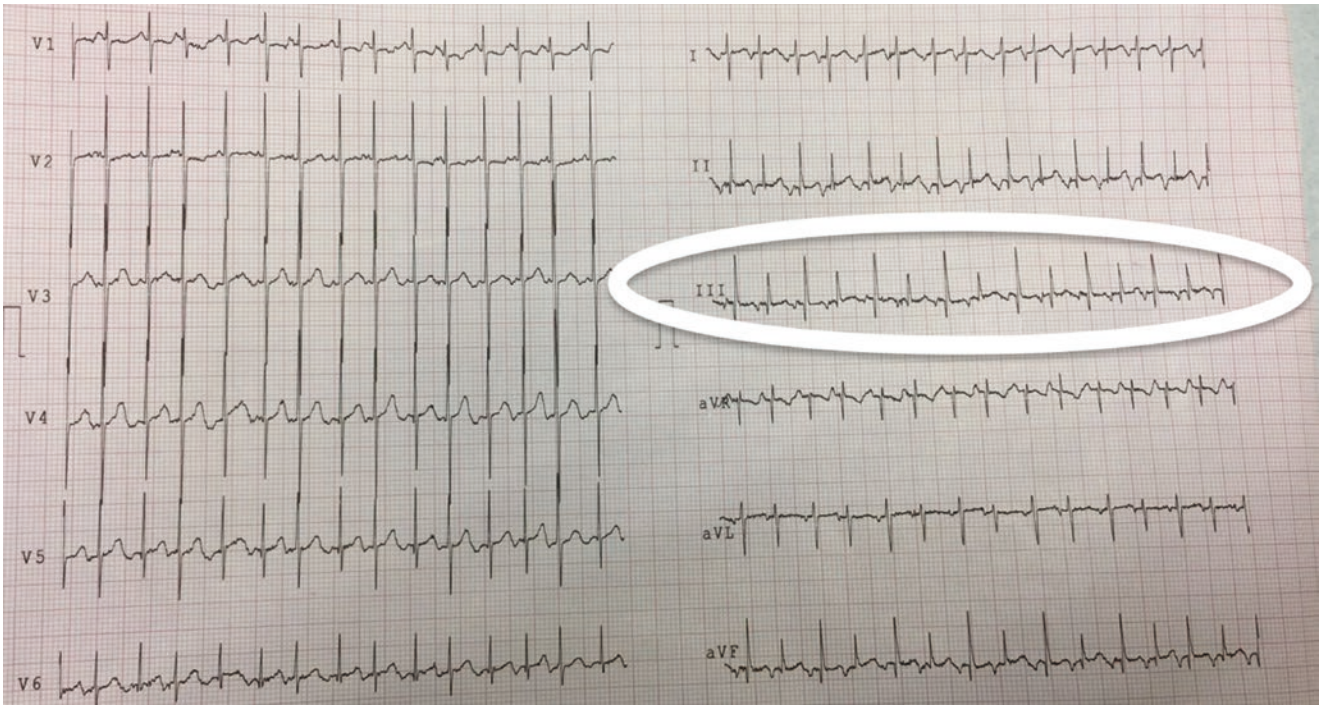

Fig. 11.44 Two-year-old boy with tachycardia during pneumonia. The doctor on duty in ER was a pediatrician puzzled by the HR, 210 bpm, in part justified by the fever, but incessant. The adult cardiologist agreed with sinus tachycardia. The ECG was shared in the chat "What's a P," the diagnosis of PJRT was made, and the baby was successfully treated. Note that despite the poor image quality, some subtle features as QRS voltage and cycle length alternance (*circle*) still remain readable

References

1. Walsh EP, Saul JP, Triedman JK. Cardiac arrhythmias in children and young adults with congenital heart disease. Philadelphia: Lippincott Williams & Wilkins; 2001.
2. Critelli G. Recognizing and managing permanent junctional reciprocating tachycardia in the catheter ablation era. J Cardiovasc Electrophysiol. 1997;8:226–36.
3. Bronzetti G, Mariucci E, Cervi E, et al. Supraventricular tachycardia in children. G Ital Cardiol. 2013;14:597–612.
4. Sternick EB, Timmermans C, Sosa E, et al. The electrocardiogram during sinus rhythm and tachycardia in patients with Mahaim fibers: the importance of an "rS" pattern in lead III. J Am Coll Cardiol. 2004;44:1626–35.
5. Trohman RG. Supraventricular tachycardia: implications for the intensivist. Crit Care Med. 2000;28(10 Suppl):N129–35.
6. Ganz LI, Friedman PL. Supraventricular tachycardia. N Engl J Med. 1995;332:162–73.
7. Obel OA, Camm AJ. Supraventricular tachycardia. ECG diagnosis and anatomy. Eur Heart J. 1997;18(Suppl C):C2–11.
8. Xie B, Thakur RK, Shah CP, Hoon VK. Clinical differentiation of narrow QRS complex tachycardias. Emerg Med Clin North Am. 1998;16:295–330.
9. Josephson ME, Wellens HJ. Differential diagnosis of supraventricular tachycardia. Cardiol Clin. 1990;8:411–42.
10. Vignati G, Annoni G. Characterization of supraventricular tachycardia in infants: clinical and instrumental diagnosis. Curr Pharm Des. 2008;14:729–35.
11. Schlechte EA, Boramanand N, Funk M. Supraventricular tachycardia in the pediatric primary care setting: age-related presentation, diagnosis, and management. J Pediatr Health Care. 2008;22:289–99.
12. Kerr CR, et al. Changes in ventriculoatrial intervals with bundle branch block aberration during reciprocating tachycardia in patients with accessory atrioventricular pathways. Circulation. 1982;66:196–201.
13. Schamroth L, Jacobs ML. A study in intracardiac conduction with special reference to the Ashman phenomenon. Heart Lung. 1982;11:381–2.
14. Calabrò MP, Cerrito M, Luzza F, Oreto G. Supraventricular tachycardia in infants: epidemiology and clinical management. Curr Pharm Des. 2008;14:723–8.

Suggested Reading

Brugada J, Blom N, Sarquella-Brugada G, et al. European Heart Rhythm Association; Association for European Paediatric and Congenital Cardiology. Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement. Europace. 2013;15(9):1337–82. doi:10.1093/europace/eut082.

Kang KT, Etheridge SP, Kantoch MJ, Tisma-Dupanovic S, Bradley DJ, Balaji S, Hamilton RM, Singh AK, Cannon BC, Schaffer MS, Potts JE, Sanatani S. Current management of focal atrial tachycardia in children: a multicenter experience. Circ Arrhythm Electrophysiol. 2014;7(4):664–70.

Jaeggi E, Öhman A. Fetal and neonatal arrhythmias. Clin Perinatol. 2016;43:99–112.

Salerno JC, Seslar SP. Supraventricular tachycardia. Arch Pediatr Adolesc Med. 2009;163:268–74.

Ventricular Arrhythmias

12

12.1 Idiopathic Ventricular Tachycardia (IVT) and PVCs

We have already talked about the electrical stability of a young heart (e.g., severe hypokalemia could be unnoticed in a newborn while the same plasma level of potassium could be lethal for an adult). VT is infrequent in children and represents less than 1% of clinical arrhythmias, while benign PVCs are much more frequent. Up to 20% of children may present benign PVCs in the first year of life. PVCs in the newborn and in the young should be judged according to morphology and not according to number. Twenty four thousand monomorphic, isolated PVCs are less worrying than 24 polymorphic or repetitive PVCs. The presence of isolated PVCs in infancy is not worrying; on the contrary, usually the sooner they start, the sooner they end. Much more worrying are PVCs starting in a "healthy" teenager, since several causes such as myocarditis or cardiomyopathy need to be ruled out. According to some authors, PVCs originating in the left ventricle with an RBBB morphology may disappear during childhood, while PVCs originating from the right ventricle with an LBBB morphology tend to persist.

Again, a VT with LBBB morphology and thus arising in the right ventricle, once ARVC has been ruled out, is usually more benign than the VT originating in the left ventricle. In fact, the RV is the well-known site of benign arrhythmias, mainly represented by RVOT VT. To differentiate VT from SVT clinically, there is no cutoff for the heart rate before the ECG reading, but in the newborn and the nursling, VT is usually slower than SVT. Moreover, in small children, an accelerated idioventricular rhythm (AIVR) alternating with a regular SR is not a rare event among VTs.

In pediatric age, ventricular tachycardia (VT) is extremely rare. While a cardiologist sees 1000 VTs frequently associated with a myocardial infarction or a previous myocardial infarction, a pediatric cardiologist sees ten VTs all caused by different situations (Table 12.1). In childhood, VT can have a narrow QRS complex. All signs currently used in cardiology to diagnose a VT are valid: AV dissociation, VA retroconduction, and the presence of capture and fusion beats. If the aspect of the QRS complex is similar to a classic bundle branch block, we are usually more likely to be dealing with an SVT conducted with aberrancy; furthermore, the knowledge of the different types of youth VT is mandatory (RVOT VT has an LBBB morphology with vertical axis, while Belhassen's VT has an RBBB associated with a left anterior fascicular block) (Fig. 12.1). Adenosine can be used as a diagnostic tool, considering that a low rate of VT is responsive to adenosine administration (less than 1%). Furthermore, it is important to establish if there is a structural cardiac disease or if VT appears in the absence of a structural heart disease. The young patient without known cardiac disease normally presents an IVT. The most common is the RVOT VT; moreover, the right ventricle is the site of different types of IVT such as moderator band VT, tricuspid annulus VT, and papillary muscle VT. The left ventricle is the site of the idiopathic left ventricle tachycardia (ILVT) originating from the left posterior hemifascicle (also known as Belhassen's VT or verapamil-sensitive VT) [1]. There are also other forms of left-sided IVT, such as left ventricular outflow tachycardia (LVOT) with an LBBB morphology like RVOT but with a rapid transition in the precordial leads (R > S in V1 and V2), mitral annulus VT, great cardiac vein VT, left anterior hemifascicle VT, papillary muscle VT, and left ventricular summit VT. Nonidiopathic VT could be the clinical manifestation of myocarditis, which should always be considered either as an acute presentation or a previous undiagnosed myocarditis with fibrosis, cardiomyopathies, or cardiac tumors (hamartomas and cardiac tumors should be considered in young children with incessant VT).

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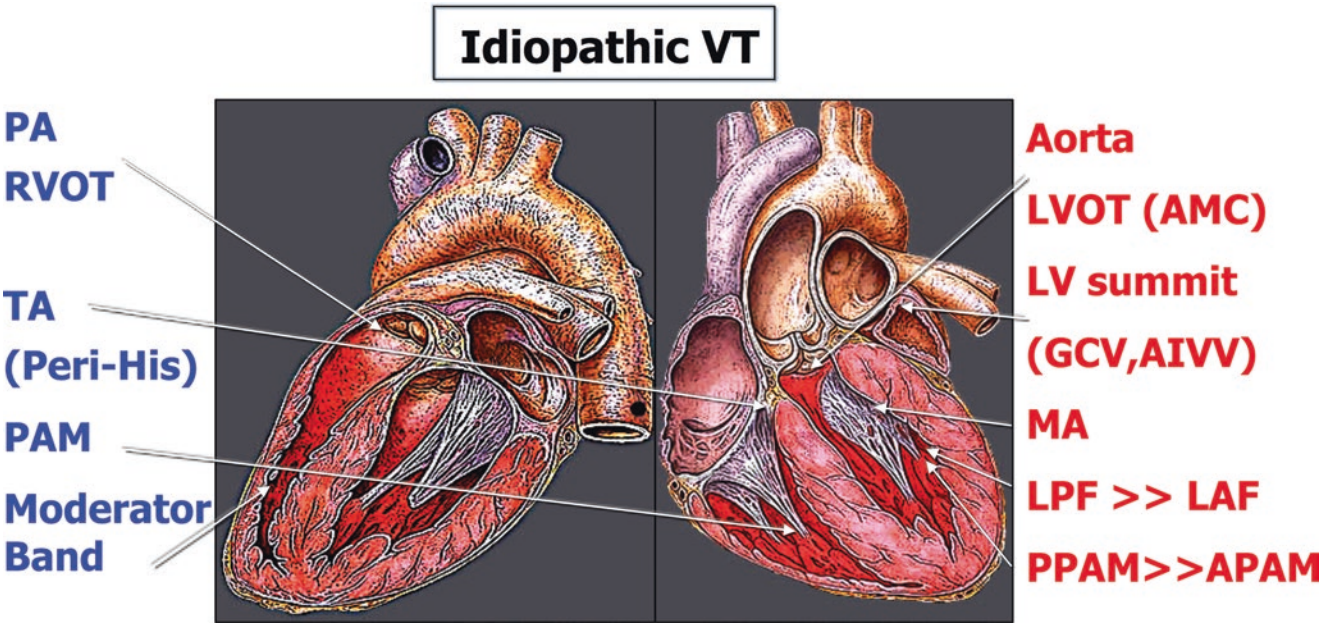


Fig. 12.1 Forms of idiopathic VT. Legend: *AIVV*, anterior interventricular vein; *AMC*, aorto-mitral continuity; *APAM*, anterolateral papillary muscle; *GCV*, great cardiac vein; *LAF*, left anterior fascicle; *LPF*, left posterior fascicle; *LV*, left ventricle; *LVOT*, LV outflow tract; *MA*, mitral annulus; *MCV*, middle cardiac vein; *PA*, pulmonary artery; *PAM*, papillary muscle; *PPAM*, posteromedial papillary muscle; *RV*, right ventricle; *RVOT*, RV outflow tract; *TA*, tricuspid annulus. Modified from Heikenwaelder Hugo, heikenwaelder@aon.at, www.heikenwaelder.at via Wikimedia Commons under CC BY-SA 2.5

Table 12.1 Etiologies of ventricular tachycardia

Congenital heart diseases	**Toxic**
Tetralogy of Fallot Mustard/Senning Ebstein anomaly GUCH Mitral valve prolapse	Drugs Digitalis Antiarrhythmics Cocaine-street drugs
Cardiomyopathies	**Ischemic**
HCM ARVC DCM Neuromuscular diseases	Kawasaki disease Coronary artery anomalies Postsurgical
Primary electrical diseases	**Cancer**
LQTS-SQTS Brugada syndrome CPVT	Rhabdomyomas Hamartomas Fibroid cancer
Infectious	**Metabolic**
Myocarditis Rheumatic fever	Hypoxia Acidosis Electrolyte imbalances

ARVC, arrhythmogenic right ventricular cardiomyopathy; *DCM*, dilated cardiomyopathy; *HCM*, hypertrophic cardiomyopathy; *CPVT*, catecholaminergic polymorphic ventricular tachycardia; *GUCH*, grown-up congenital heart disease; *LQTS*, long QT syndrome; *SQTS*, short QT syndrome

12.2 VT Associated with Structural Heart Disease

Among CHD, tetralogy of Fallot is the most affected by VT in the late follow-up. Mustard or Senning repairs may be complicated by VT due to a failing systemic right ventricle [2]. The incidence of VT in patients with tetralogy of Fallot is 4%, while the sudden death incidence 20 years after the surgical correction is 2% [3]. Risk factors for sudden death are surgical correction consisting in annular patch, severe pulmonary regurgitation, left ventricular systolic dysfunction, and QRS duration >180 ms. Other congenital heart diseases associated with VT are coronary anomalies, Ebstein or Ebstein-like severe tricuspid valve dysplasia, univentricular heart with severe impairment of systolic function, and patients with Eisenmenger syndrome. Coronary anomalies associated with VT are ALCAPA and ACAOS. The clinical presentation of ALCAPA is left ventricular dysfunction in the nursling, while in late childhood it could be diagnosed following a VT/FV starting after physical exercise or emotional stress; it should be noted that the rest ECG is rarely normal. On the contrary, ACAOS patients have a normal ECG.

The most common cause of VT and sudden death (SD) in young patients is hypertrophic cardiomyopathy [4]. The prevalence of HCM is 1:500; except for the mutation of sarcomeric proteins transmitted in the autosomic dominant way during childhood, the hypertrophic phenotype is shared by several syndromes such as Pompe disease, Danon disease, Noonan syndrome, and Friedreich ataxia and other neuromuscular syndromes such as Anderson-Fabry disease, Kearns-Sayre syndrome, Barth syndrome, etc. In these patients, association with short PR interval and pre-excitation is frequent.

Another cause of VT and SD is ARVC, where VT typically has an LBBB morphology. ARVC could be difficult to diagnose at the beginning of the disease, and differential diagnosis with benign RVOT could be problematic. Typical ECG patterns such as negative T waves in the right precordial leads could be fleeting and transient, while the epsilon wave, a pathognomonic sign, appears when the disease is quite advanced and has a low sensitivity. As said before, the distinction between ARVC and RVOT is crucial; the first is an evolving disease associated with a high risk of SD, while the second is a benign condition with a nonnegligible rate of spontaneous resolution and an RFCA success rate >90%.

12.3 VT Without Evidence of Structural Heart Disease

Long QT syndrome is the paradigm of channelopathies; nevertheless, the opposite condition of short QT syndrome recently entered the large family of channelopathies [5]. It is important to acknowledge that "electrical" diseases are associated with circadian or longer ECG variation and faced with a borderline ECG it is important to collect a detailed medical and family history. In addition, the cardiologist should (1) perform the ECG recording at different times and body temperatures, (2) analyze the family's ECGs, and (3) perform an exercise test and a Holter ECG (the last one is important in young children who may not be able to perform an exercise test). Furthermore, long QT syndrome diagnosis is based on a probabilistic evaluation that includes instrumental evaluation, clinical examination, and molecular exams (Schwartz score).

Brugada syndrome is a channelopathy inherited in an autosomal dominant manner, characterized by the presence of ST elevation in the right precordial leads. Up to now, 12 mutations have been described; mutation of the SCN5A gene with a downregulation of the sodium channel is the most frequent mutation found in 15–20% of cases, while the mutation of CACNA1c expressed with the loss of calcium channel function has been described in 7–10% of cases. The resting ECG may present a long PR interval, which is rare in young patients and therefore could be an important clue.

Hence, the right ventricle outflow tract is the focus of most arrhythmias and those purely "electrical" such as RVOT VT and, on the other hand, those associated with a structural alteration such as tetralogy of Fallot or ARVC. The odd vulnerability of RVOT is due to genetic characteristics, embryological development (crista neuralis), and, finally, anatomic features of this area, which is involved in several congenital heart diseases (Fig. 12.2). The left ventricle has its own "Bermuda Triangle" too: the postero-basal aspect. In fact, this is a site involved in different situations with significant arrhythmic expressions such as mitral valve prolapse, Duchenne dystrophy, nonischemic fibrosis, myocarditis, and ARVD extended to the left ventricle (Fig. 12.3).

Finally, catecholaminergic polymorphic ventricular tachycardia (CPVT) is an extremely rare cause of VT or FV. Clinical suspicion should be based on a history of syncope during physical effort or emotional stress in the presence of a normal resting ECG or with a short PR interval. The presence of polymorphic or irregularly shaped ventricular arrhythmias during ECG exercise stress testing could help the diagnosis. CPVT is caused by cardiac ryanodine receptor type 2 (RyR2) or calsequestrin 2 (CASQ2) gene mutations. The RyR 2 mutation is responsible for almost 60% of CPTV while the second one is more rare; in 25% of patients the cause is unknown.

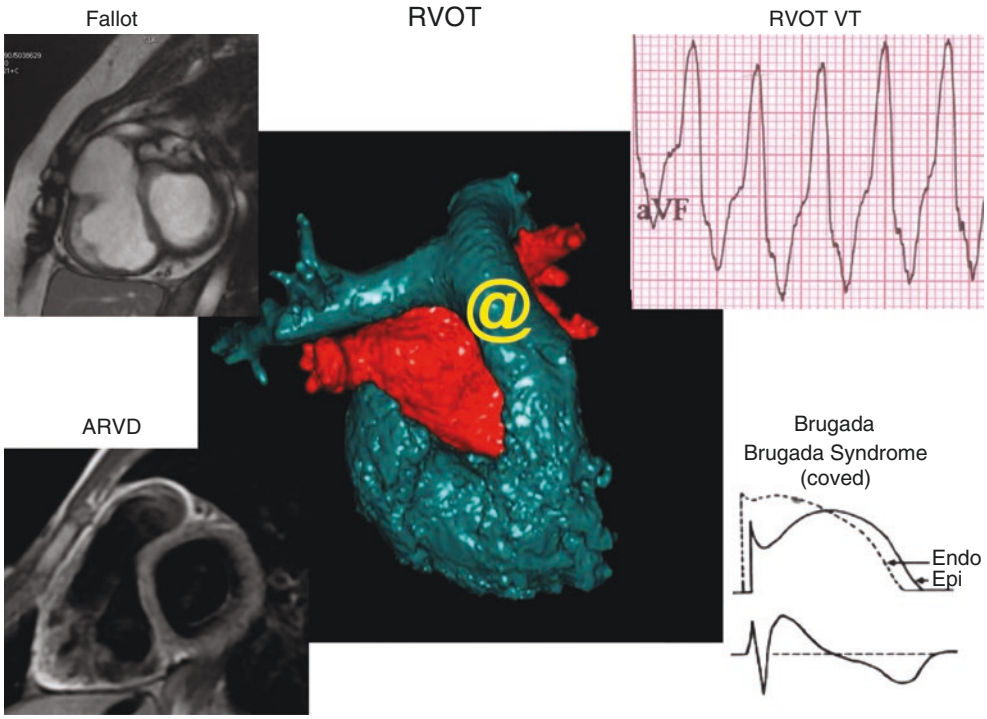


Fig. 12.2 The RVOT as a site of macrostructural and microstructural (or electric) arrhythmogenic diseases

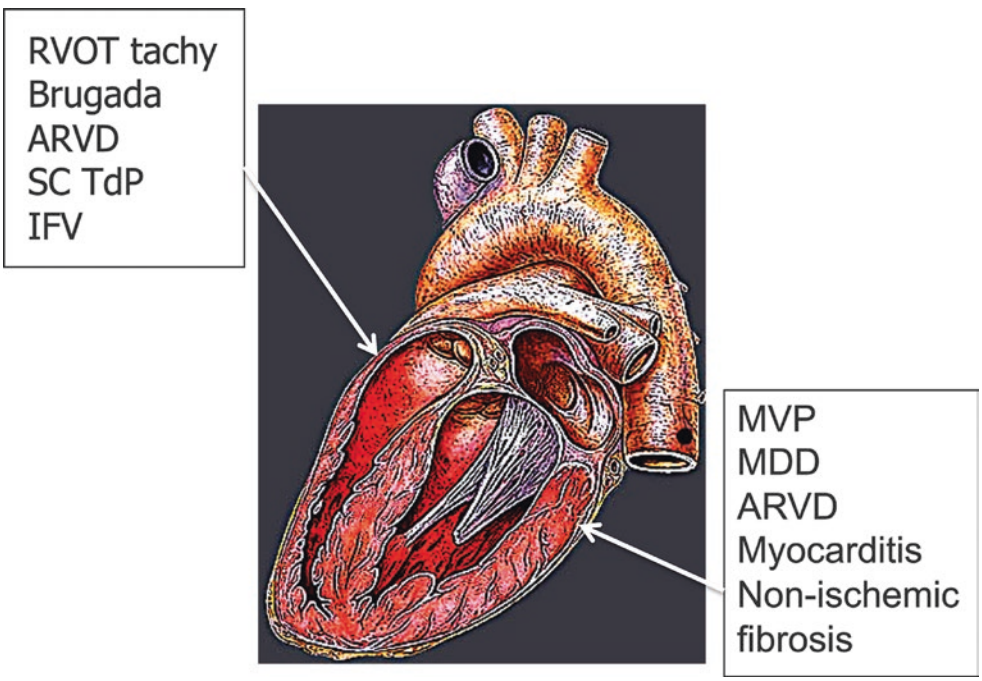


Fig. 12.3 Right- and left-sided arrhythmias. Legend: *SC TdP*, short-coupled torsades de pointes; *IVF*, idiopathic ventricular fibrillation; *MVP*, mitral valve prolapse; *MDD*, muscular Duchenne dystrophy. Modified from Heikenwaelder Hugo, heikenwaelder@aon.at, www.heikenwaelder.at via Wikimedia Commons under CC BY-SA 2.5

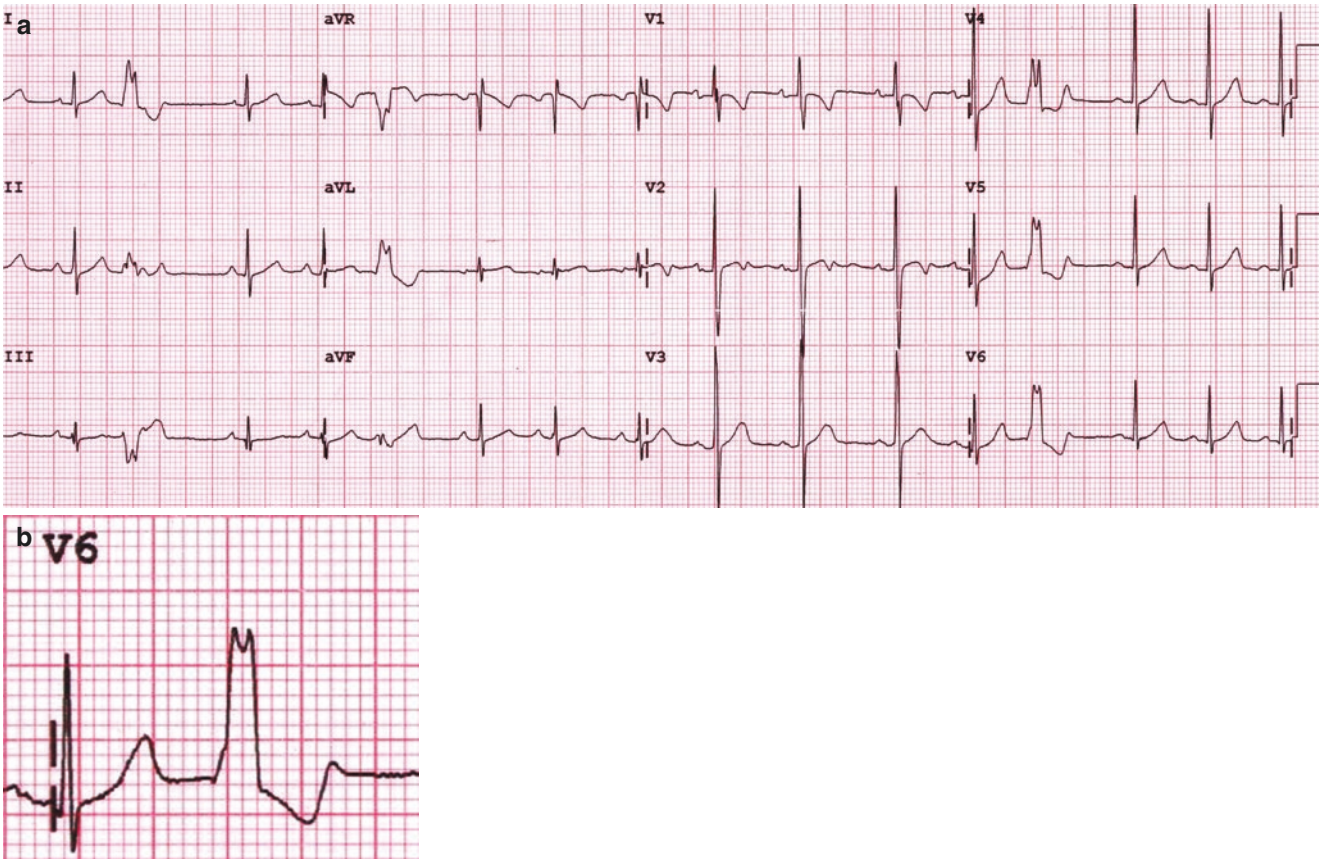


Fig. 12.4 (**a**) Eight-year-old male with frequent monomorphic PVCs with LBBB and left axis deviation. The patient is asymptomatic; echocardiogram is normal. The family history is negative and the workup will be complete with an exercise test, with no need for an MRI. (**b**) Close-up

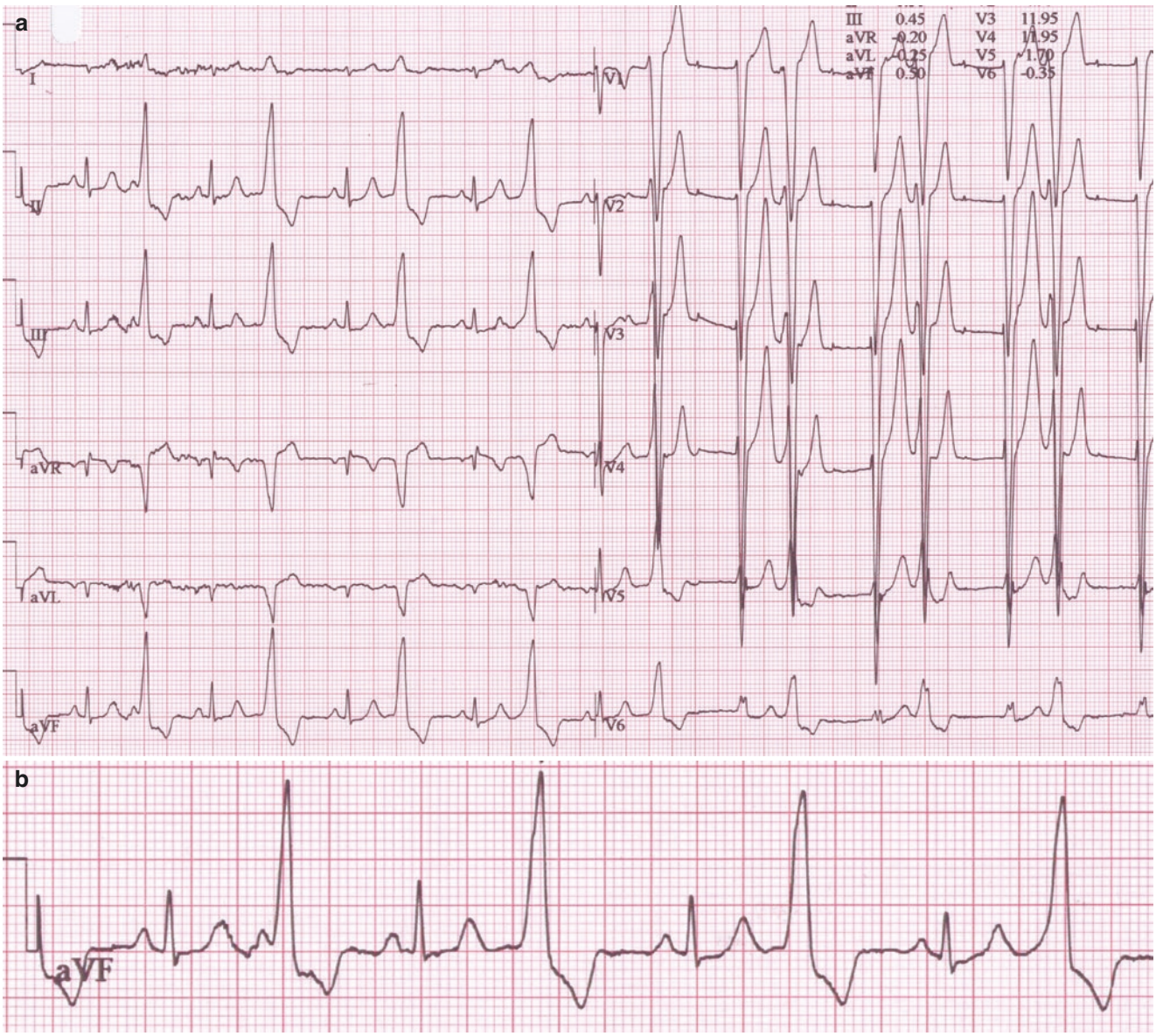


Fig. 12.5 (**a**), (**b**). Eleven-year-old female with bigeminal PVC. The LBBB and vertical axis denote an origin from the right ventricular outflow tract, the most common site of idiopathic PVC, and ventricular tachycardia

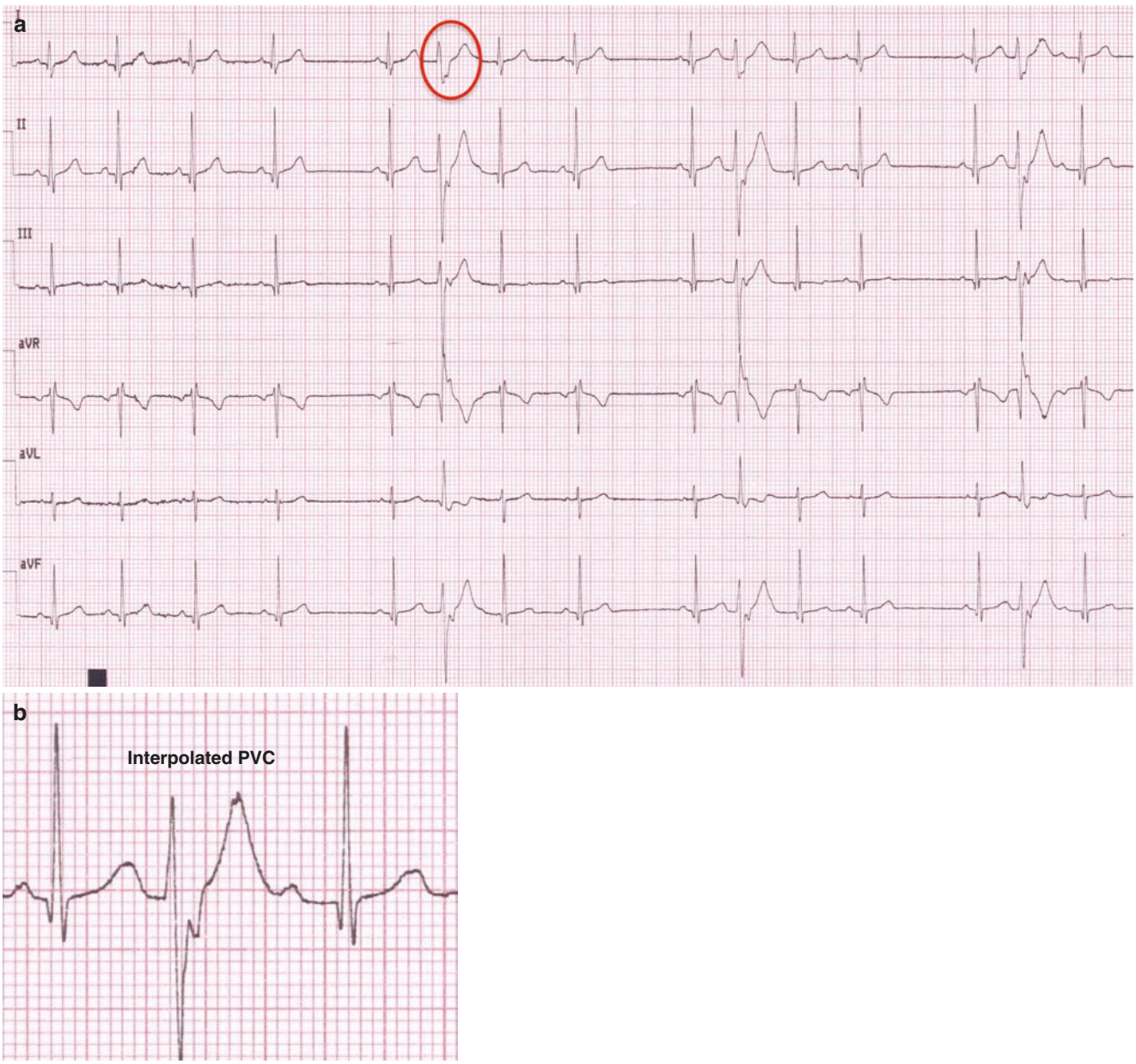


Fig. 12.6 (**a**) Eight-year-old boy with PVCs. RBBB morphology (in the absence of precordial leads, the QRS morphology of RS in lead I is consistent with RBBB) and superior axis deviation (the QRS vector is directed toward aVR and aVL). The origin can be located in the lower aspect of the left ventricle, most likely in the left posterior fascicle, a well-known site of idiopathic PVCs and tachycardia. These PVCs are perfectly interpolated (**b**), and there is no compensatory pause

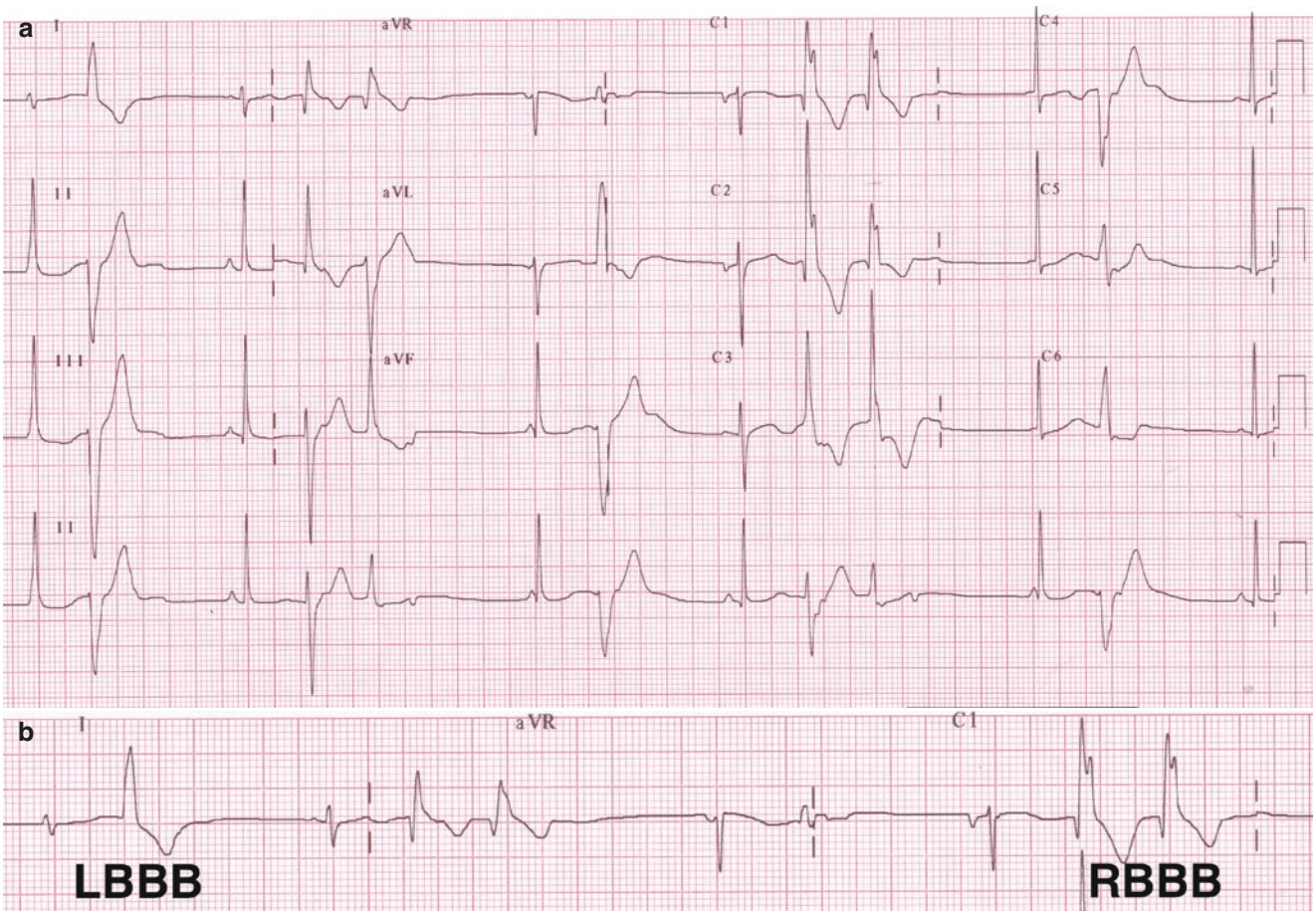


Fig. 12.7 (**a**)10-Year-old male admitted for chorea. There are polymorphic PVCs and couplets with LBBB and RBBB. (**b**) The patient also showed prominent U waves and long QT. These electrocardiographic aspects, along with somatic and neurological signs, are suggestive for Andersen–Tawil syndrome, also known as LQTS 7 (genetic testing ongoing)

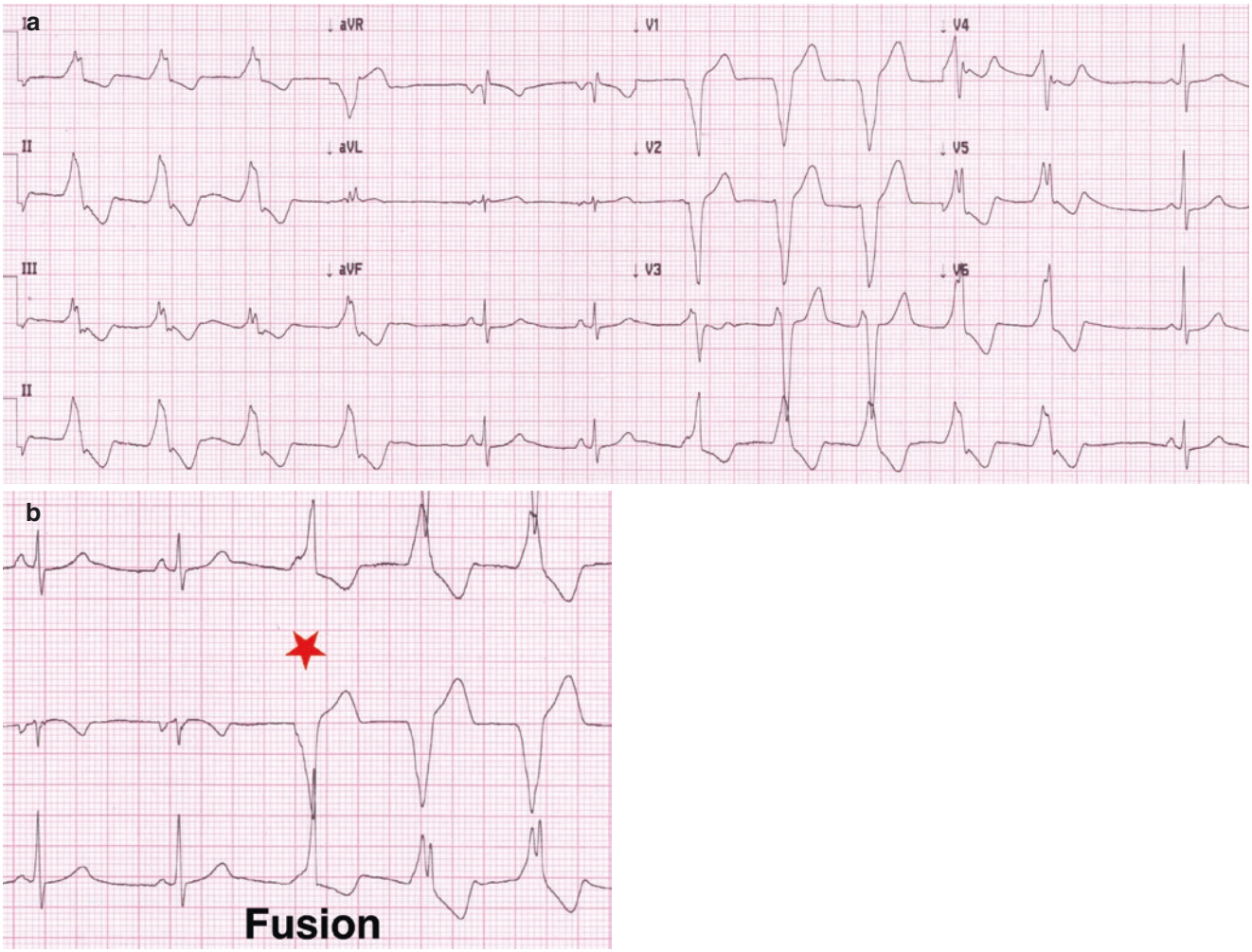


Fig. 12.8 (**a**) Twelve-year-old female with incidental finding of wide complex QRS. There is alternance between sinus/narrow beats and ventricular/wide beats with LBBB morphology and vertical axis (RVOT origin). This idioventricular rhythm is competitive with the sinus rhythm at physiological HR. (**b**) A fusion between a sinus beat and the ventricular beat can be noted. The exercise test confirmed that the idioventricular rhythm is suppressed by sinus tachycardia (overdrive suppression)

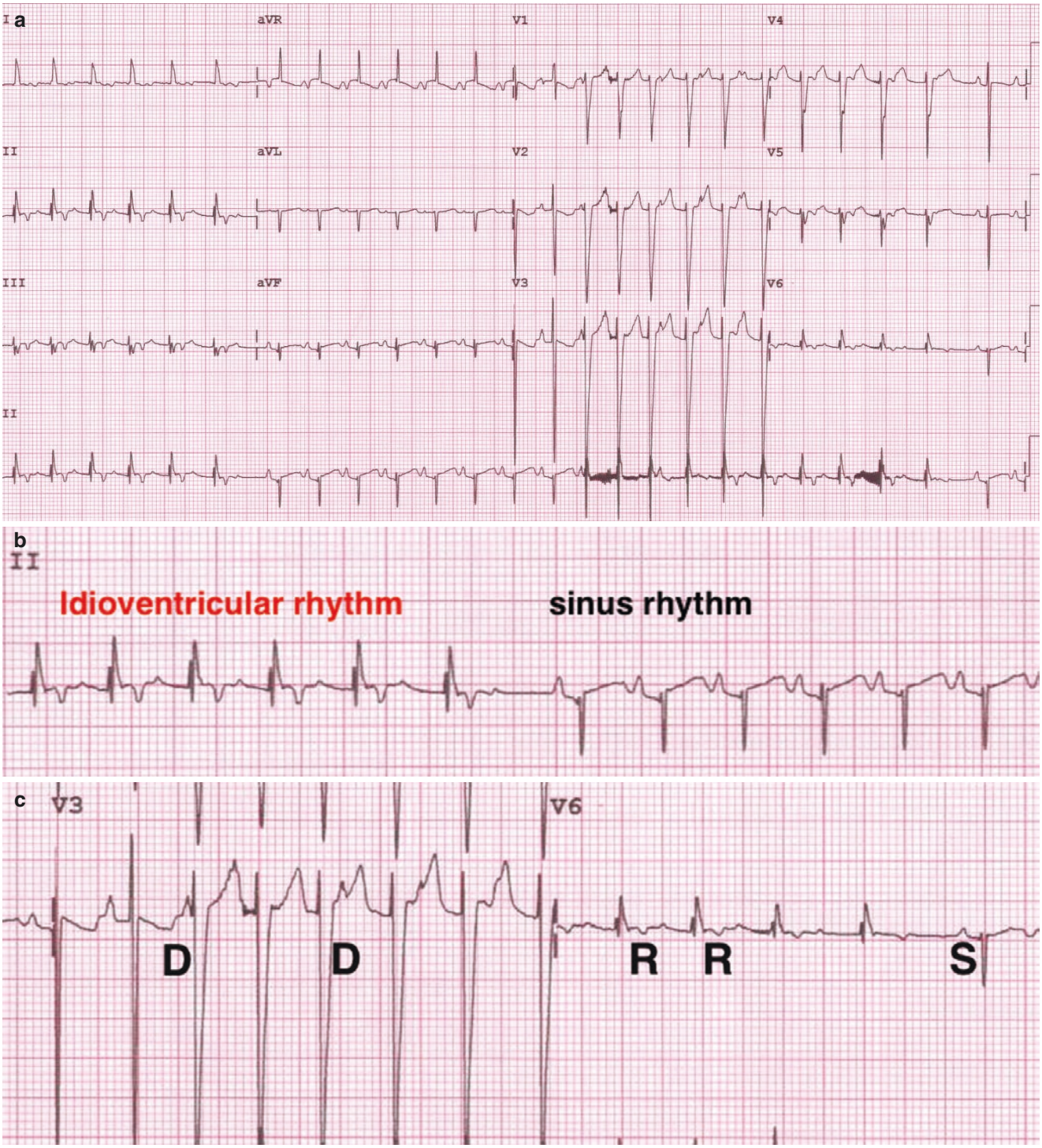


Fig. 12.9 Neonate female. (**a**) There is an alternance between slow VT (also called accelerated idioventricular rhythm) and sinus rhythm at a slightly slower HR. (**b**) Close-up, retroconducted P waves are seen, negative in the inferior leads. (**c**) At the restart of the arrhythmia, the P waves appear dissociated (D) and retroconducted (R), eventually returning to sinus rhythm (S). The arrhythmia was treated with sotalol for the first year of age and then stopped without relapses. As can be noted, in infants and small children, the ventricular ectopy may present narrow QRS, recognizable from the sinus beat only according to morphology. Another cause of non-sinus neither junctional narrow QRS is a fascicular origin

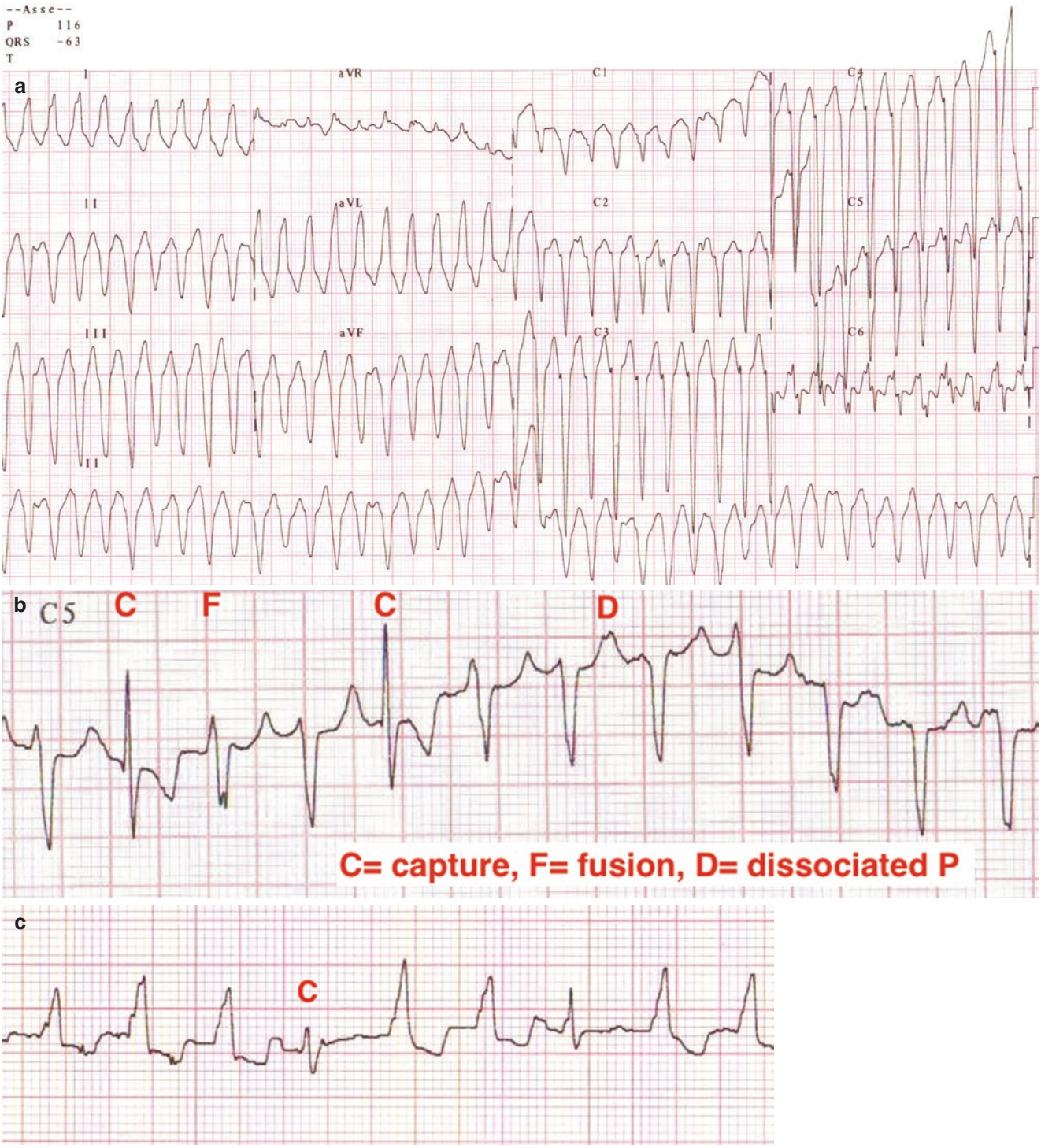


Fig. 12.10 (**a**) Seven-month-old boy admitted for heart failure. The almost absolute negative concordance in the precordial leads (only V6 shows an R wave) and the "slow" falling down of the S wave in V1 (delayed intrinsicoid deflection) helped to identify the ventricular nature of the arrhythmia. (**b**) The VT is confirmed by fusion phenomena (F), characterized by a profile of the QRS which is a hybrid between the ventricular ectopic beat and the sinus beat; the dissociation (D) where it is clear that the P wave is found randomly in front of the R wave which follows it, without being the cause. (**c**) Close-up. An example of capture (C) where a sinus beat is able to pace for a while. The cardiac imaging was negative as was the biopsy (cardiomyopathies, tumors, and myocarditis were ruled out). Amiodarone and flecainide were continued for 6 years. At treatment interruption, morpho-functional normality and the absence of relapses were documented

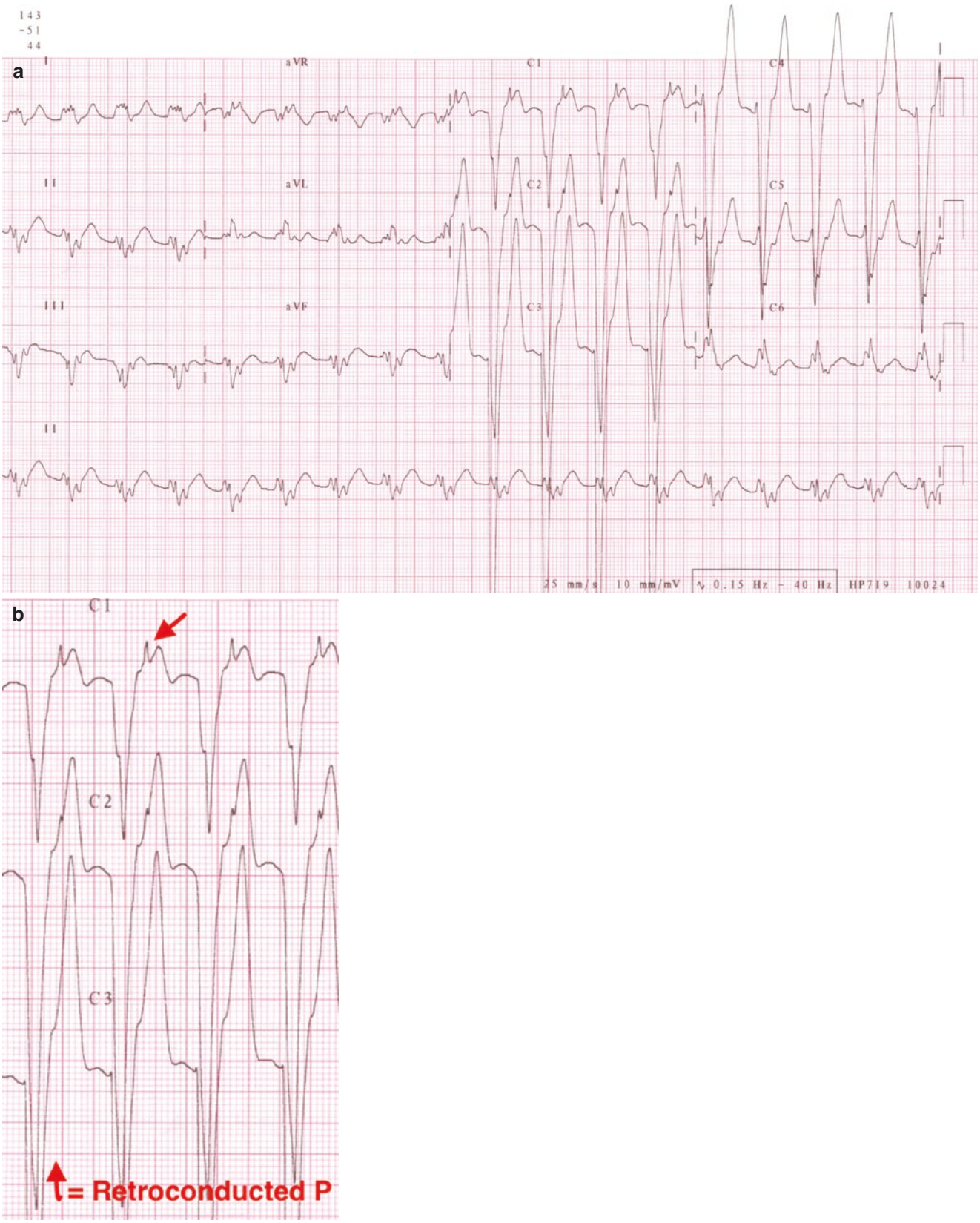
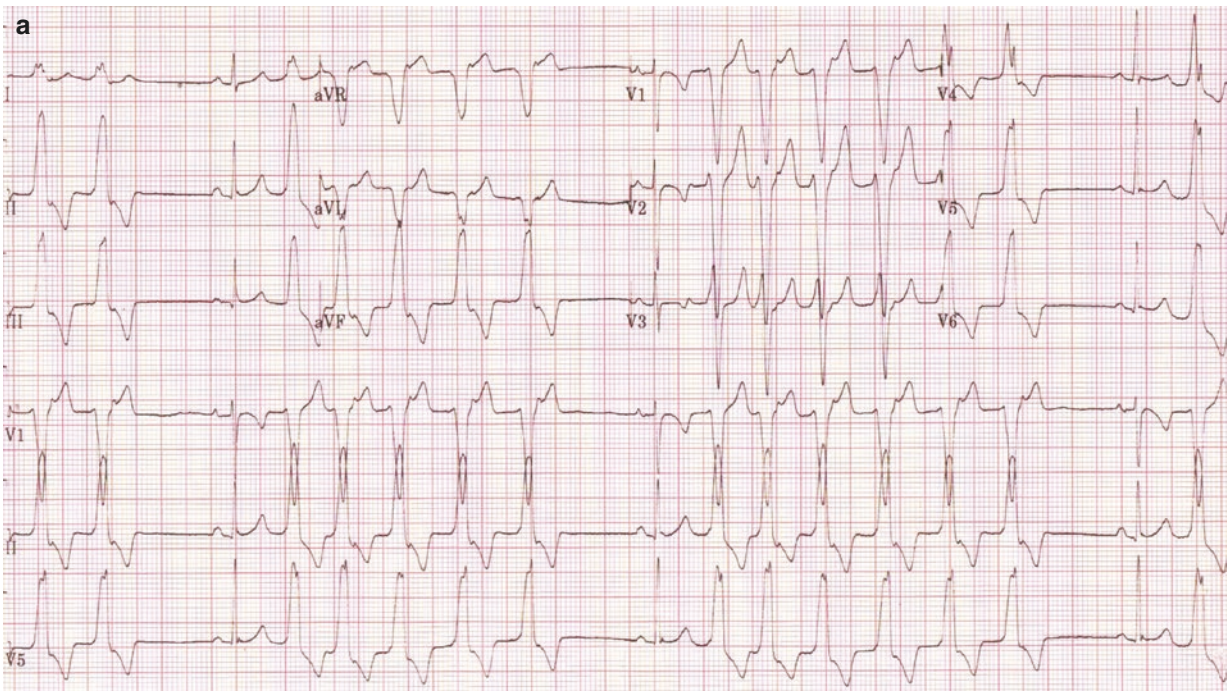


Fig. 12.11 (**a**) Twelve-year-old male with incessant tachycardia and tachycardiomyopathy. There is an LBBB with superior axis deviation. (**b**) Close-up. Retroconducted P waves can be noted. The EPS found a VT originating from the moderator band, successfully ablated with RFCA



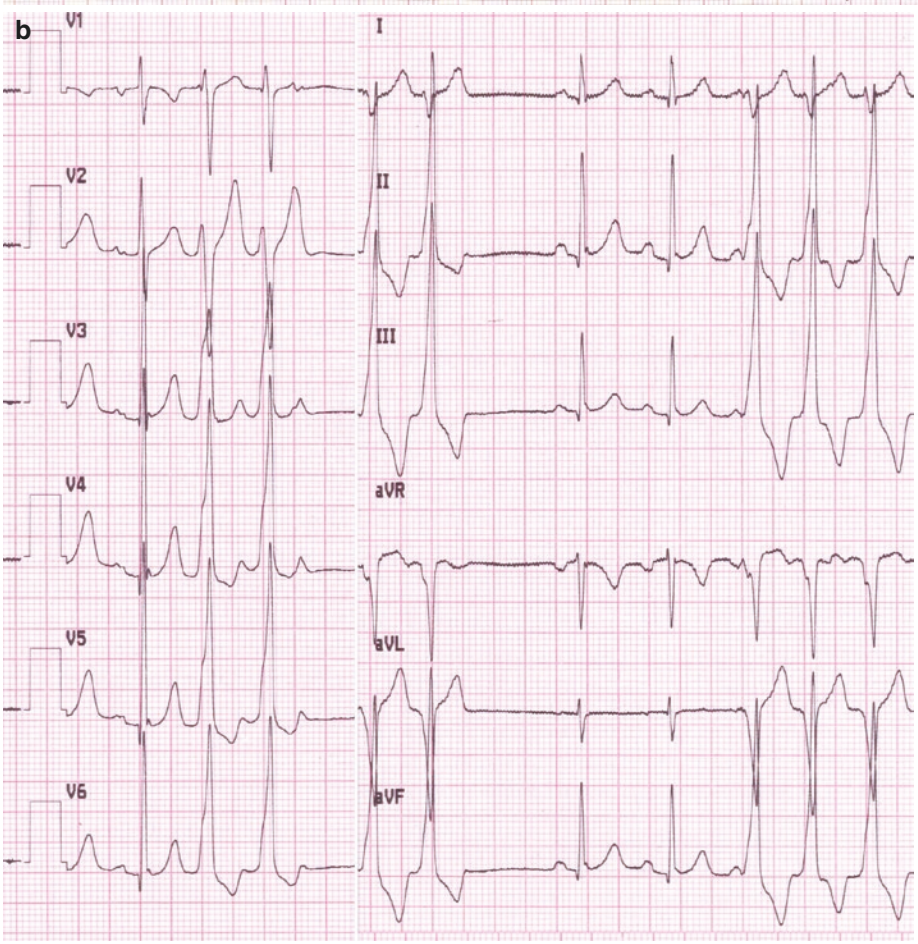
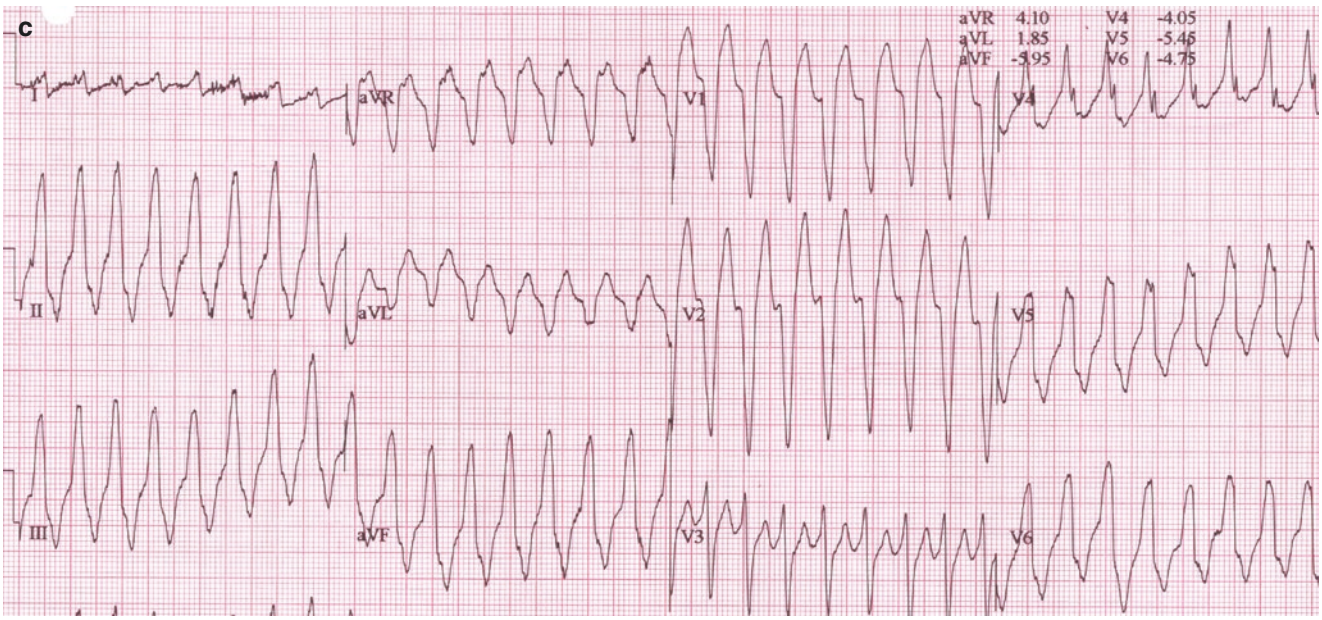


Fig. 12.12 (**a**) Sixteen-year-old female. Non-sustained ventricular tachycardia phases with LBBB appearance and vertical axis can be seen. This is the classic presentation of RVOT tachycardia, typical of youth, with a female predominance. The rate of the VT is not high, at about 150 bpm. During the stress test the arrhythmia was already suppressed at a sinus rate of 180 bpm. This behavior emphasizes the benign nature of the arrhythmia. The arrhythmia completely disappeared 3 years later. (**b**) Thirteen-year-old girl with RVOT VT. (**c**) Eighteen-year-old female with a more aggressive form of RVOT VT; during the exercise test the VT rate rose to 200 bpm. The patient was successfully ablated



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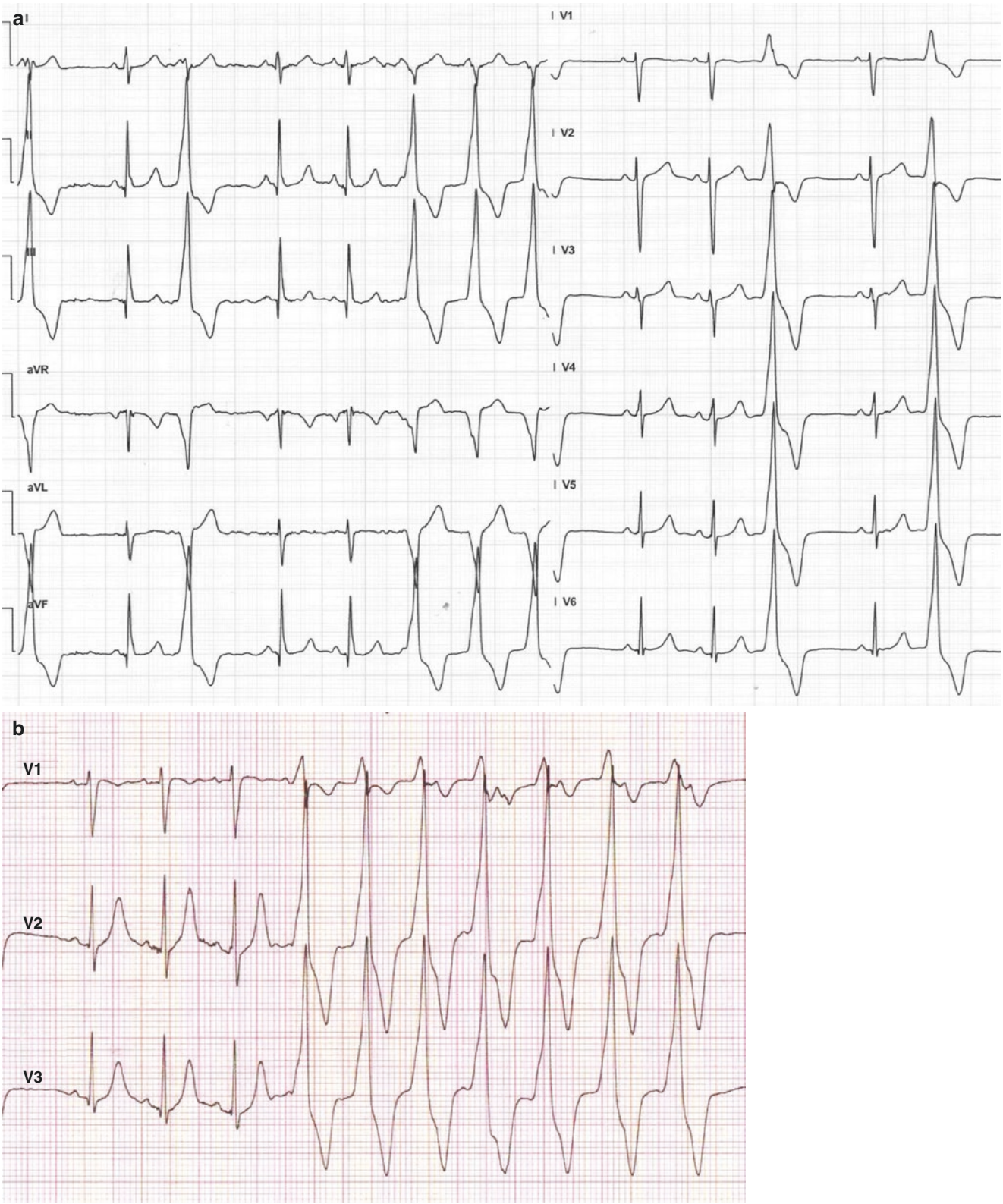


Fig. 12.13 (**a**) Five-year-old male with idiopathic tachycardia. There is an LBBB morphology but the transition (R > S) in the precordial leads is early. The EPS confirmed a left ventricular outflow tract VT (LVOT VT). (**b**) Close-up

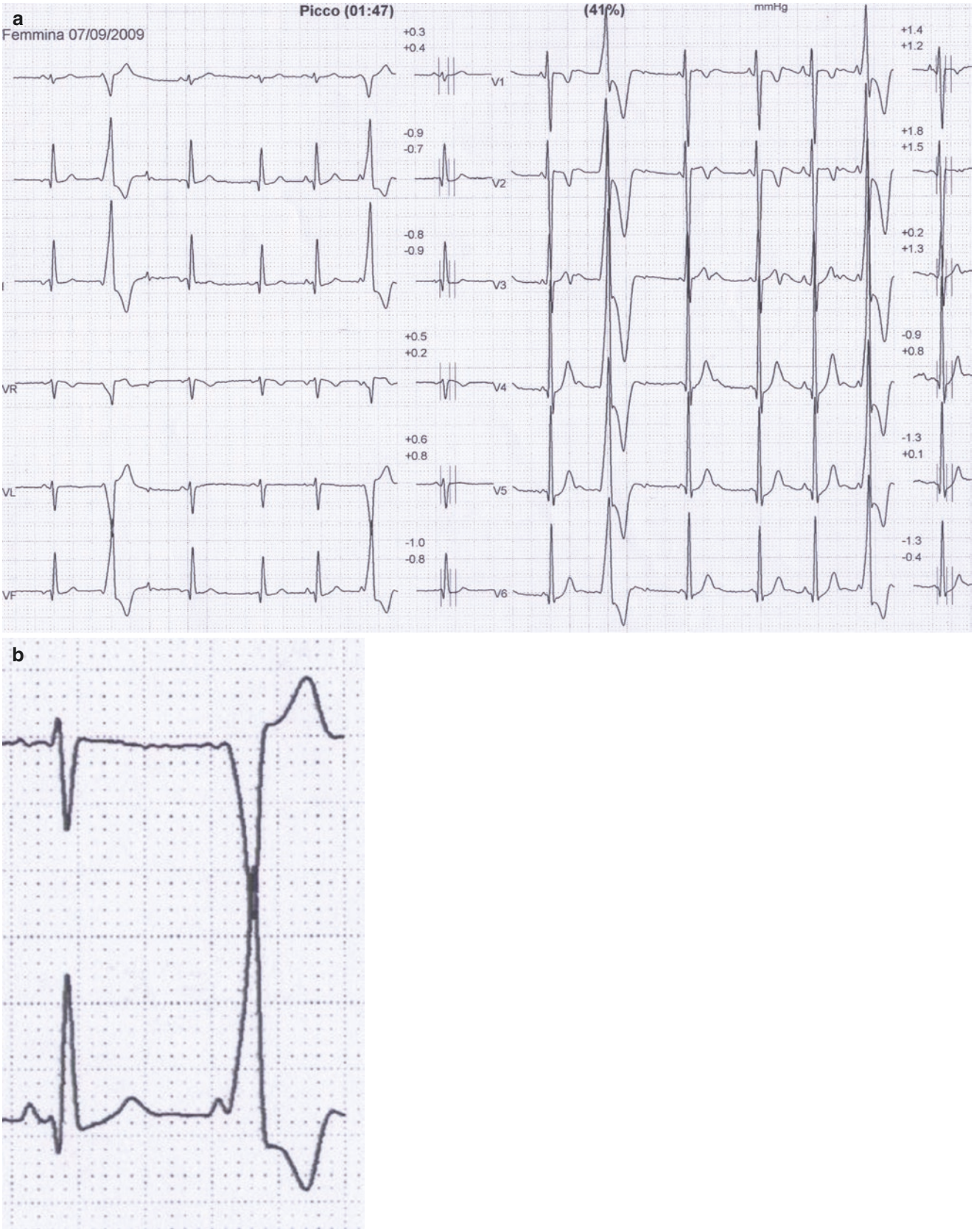
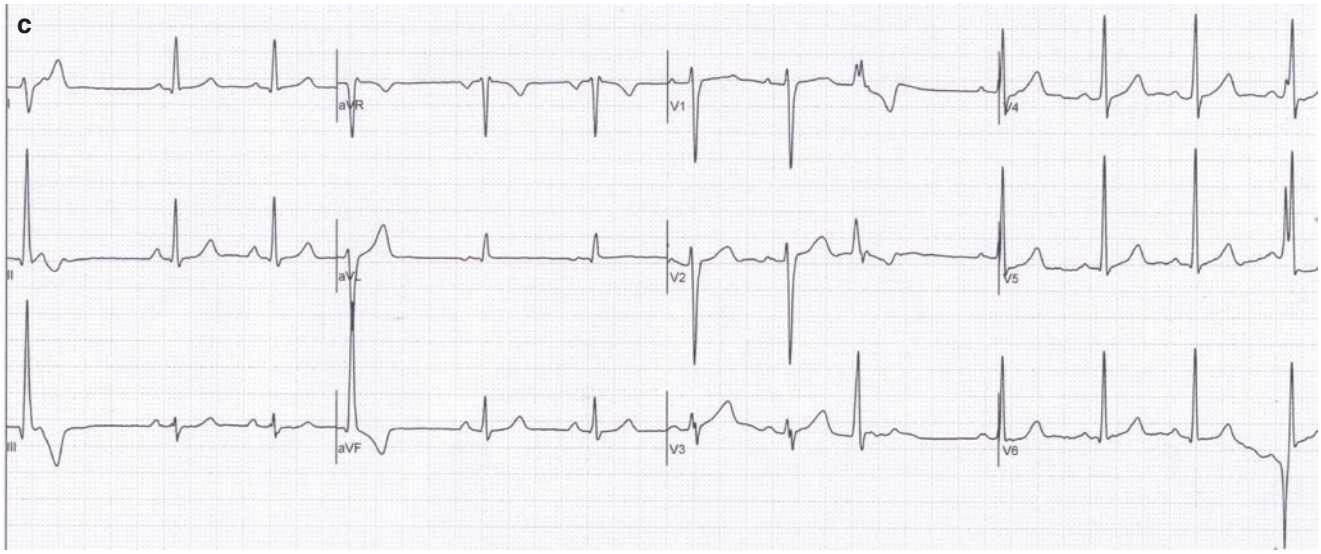


Fig. 12.14 (**a**) Ten-year-old female with idiopathic PVCs. (**b**) Close-up. As in the previous case, there is an LBBB morphology with early transition in the precordial leads. The EPS found a right aortic coronary cusp origin. Note the "pseudo-delta." (**c**) A 16-year-old female with mitral annulus PVC



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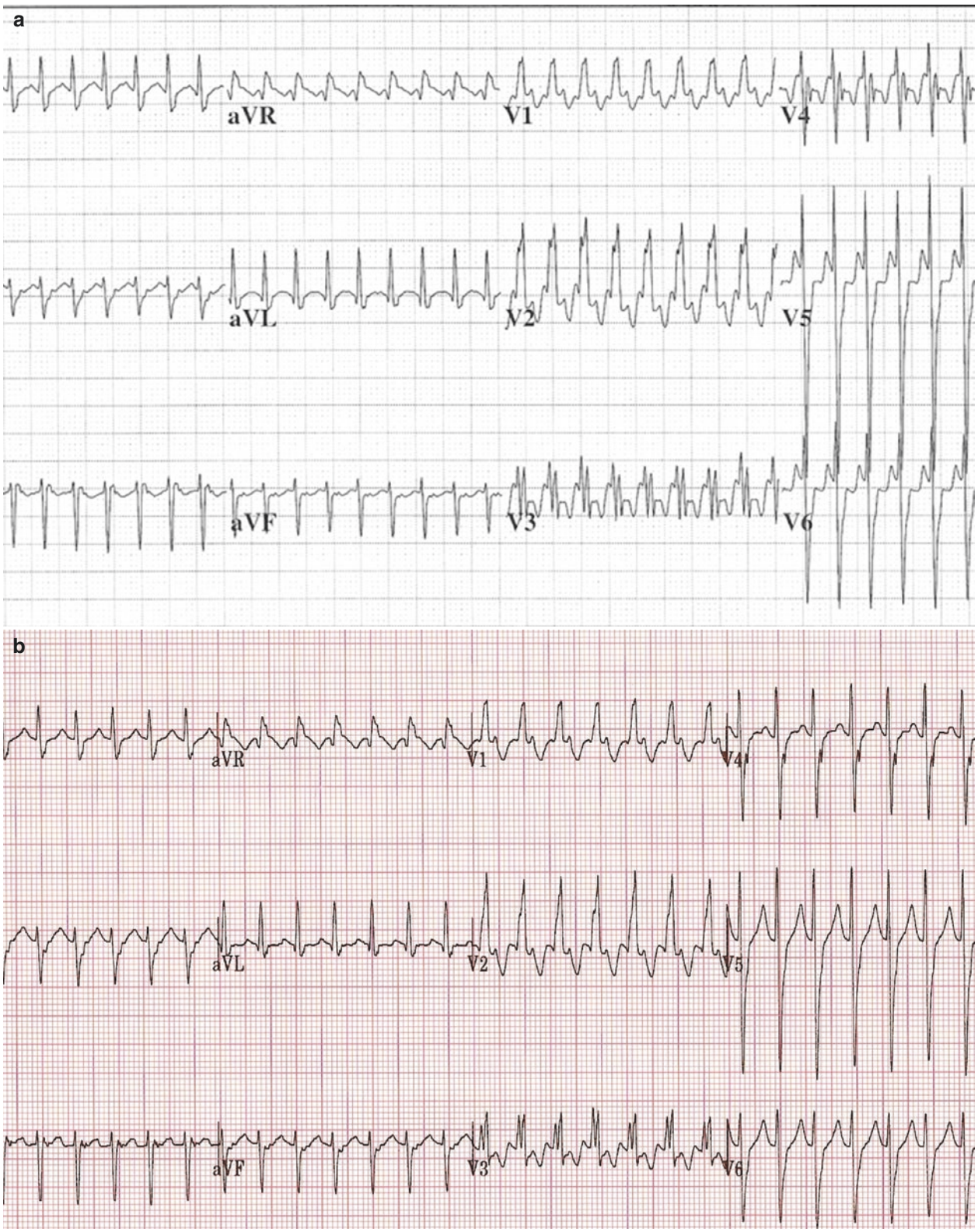
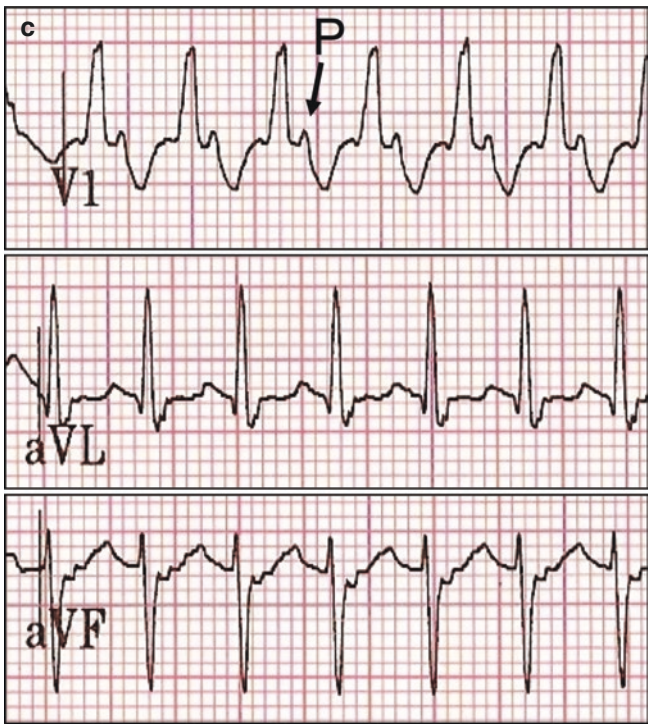


Fig. 12.15 (**a**) Eight-year-old boy with neonatal arrhythmia in prophylactic therapy with amiodarone from the first months of life. (**b**) He relapsed 2 years after drug interruption at 6 years of age. (**c**) Close-up. The HR is 210 bpm, relatively narrow QRS complex (duration 80 ms), with RBBB and left axis deviation. In V1 there is a positive peaked wave (*arrow*) which has no ventricular relevance: it is the retroconducted P wave. The aVL lead is positive and the inferior leads show rS waves, consistent with left axis deviation. This is the typical presentation of fascicular ventricular tachycardia (ILVT, idiopathic left ventricular tachycardia otherwise called "verapamil sensitive" or Belhassen tachycardia). The site of origin (posterior hemifascicle of the left bundle branch) is consistent with the typical ECG presentation and makes the arrhythmia susceptible to radiofrequency ablation



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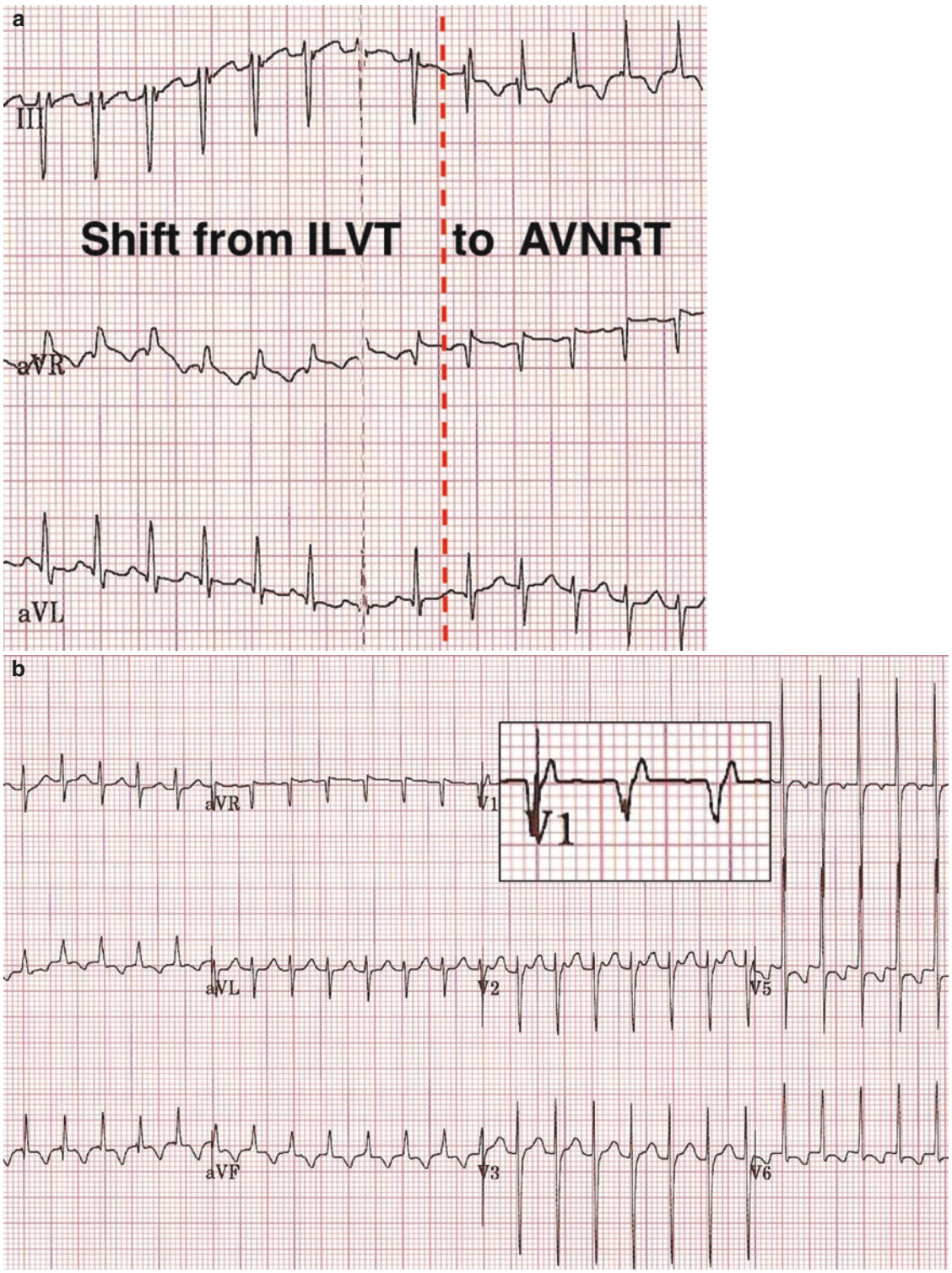

Fig. 12.16 (**a**) The same patient as previous figure. When you see narrow complexes following wide complexes (or at least different) without any apparent continuity, it is normally the same tachycardia initially with aberrant conduction (linking) which then resolves. The coexistence of two tachycardias is an exceptional event. In this case, we see the ILVT of previous ECGs which changes toward an imperceptibly faster one, with narrower complex and with different electrical axis tachycardia 1 min after the bolus of adenosine (at the emergency room, it was interpreted as PSVT and initially treated as such). (**b**) Thus the second arrhythmia is a nodal reentry (AVNRT) induced by adenosine which, by surpassing it in speed, suppressed the preexisting arrhythmia (see figure below). (**c**) The electrophysiological study reproduced the mechanism of dual arrhythmia and confirmed a nodal reentrant tachycardia ("slow-fast") induced by adenosine, owing to its differential effect in nodal duality conduction. All antiarrhythmic agents may be proarrhythmic, including adenosine, which is "proarrhythmically" antiarrhythmic in this case, replacing a VT with a PSVT. In addition, we can see the retroconducted P wave in V1 (pseudo R). (**d**) After the restoration of sinus rhythm, the widespread negative T waves can be interpreted as "electric memory" [6]

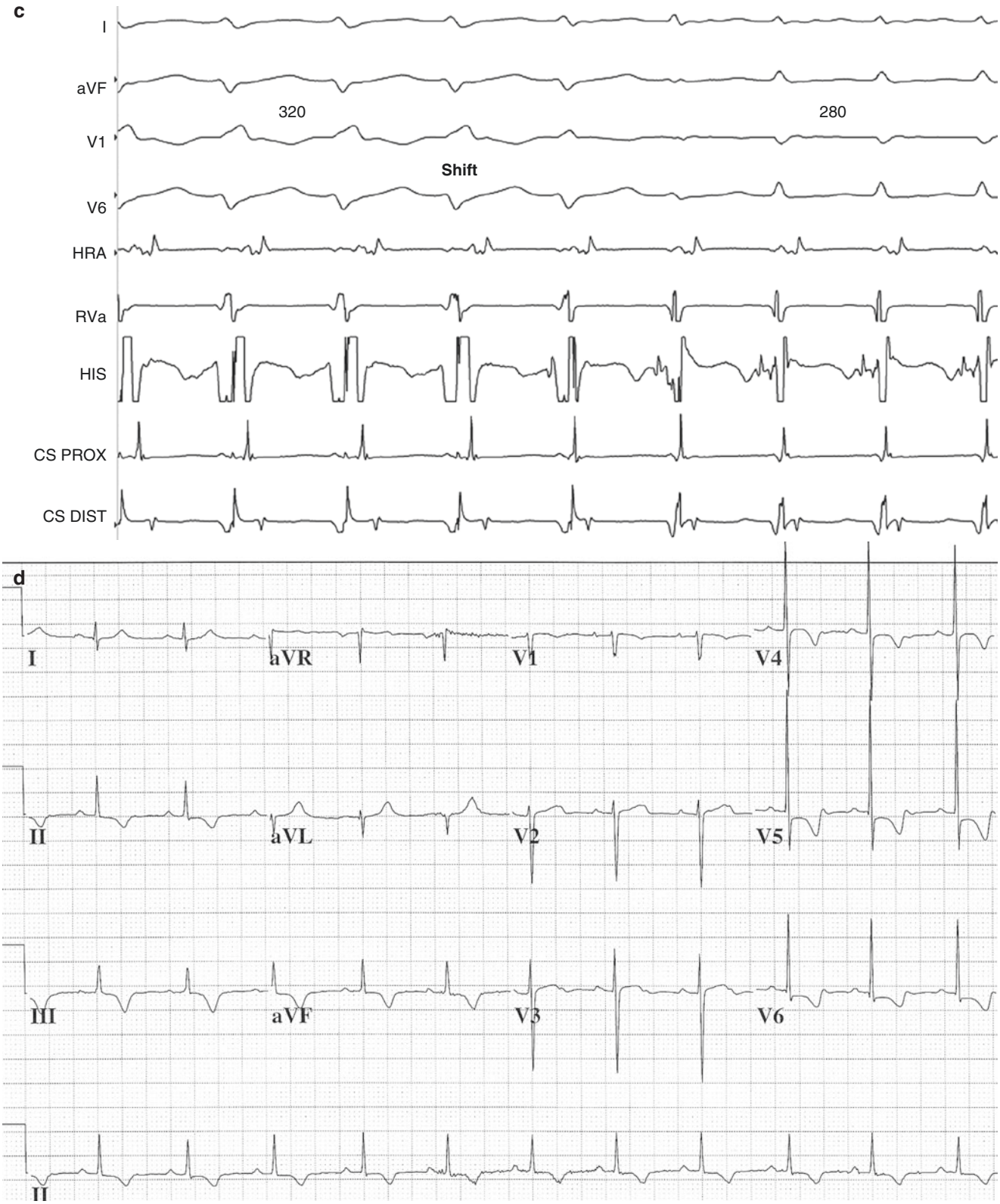

Fig. 12.16 (continued)

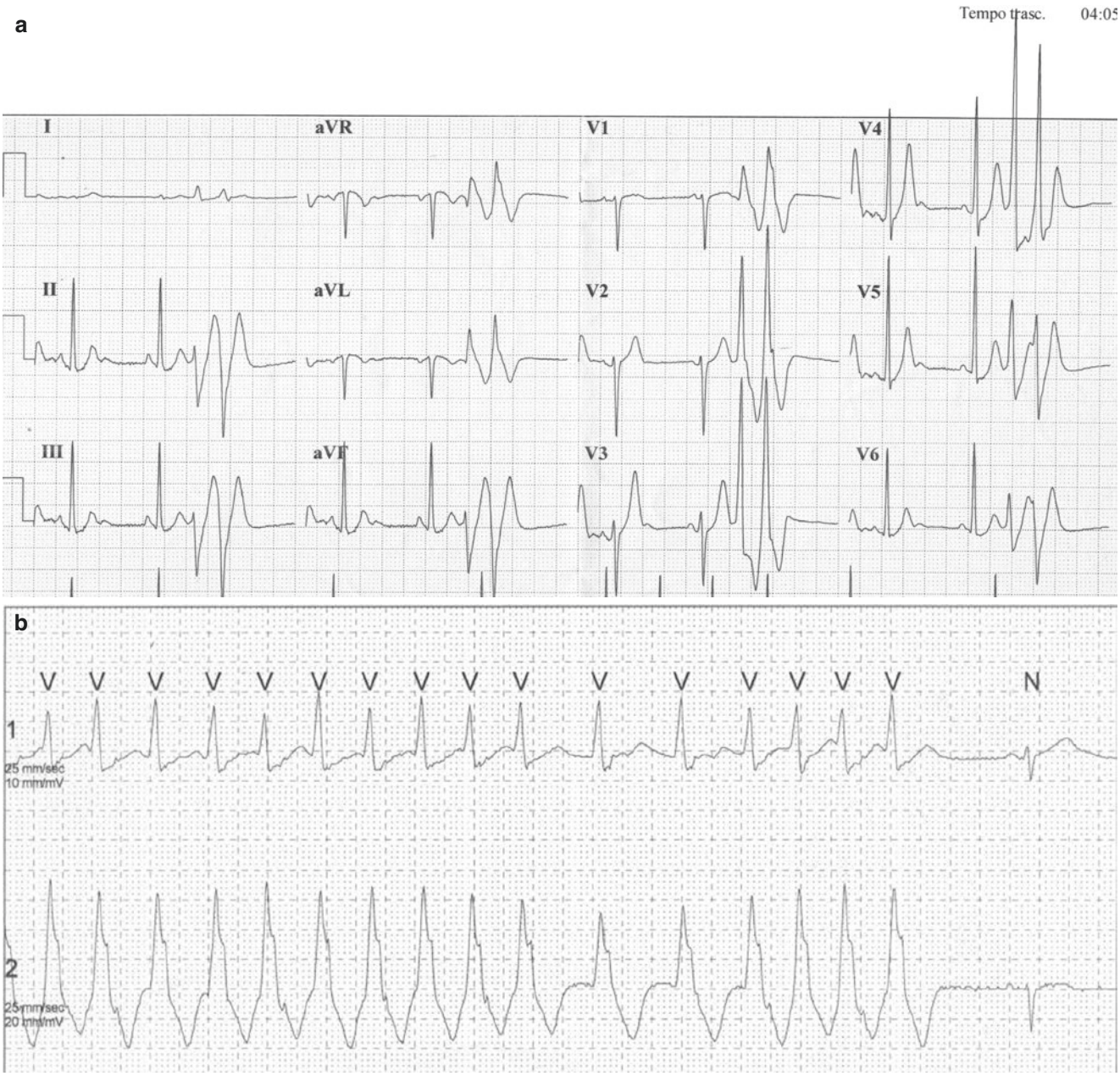

Fig. 12.17 (**a**) Sixteen-year-old girl with MVP and incidental finding of PVCs at rest. There are PVCs with RBBB morphologies and superior axis deviation in the frontal plane, indicating an origin from the papillary muscle or from the postero-basal wall close to the mitral anulus. (**b**) Holter documented a non-sustained VT. Fibrosis can be the substrate of this malignant form of MVP and correlates with ventricular arrhythmias [7]

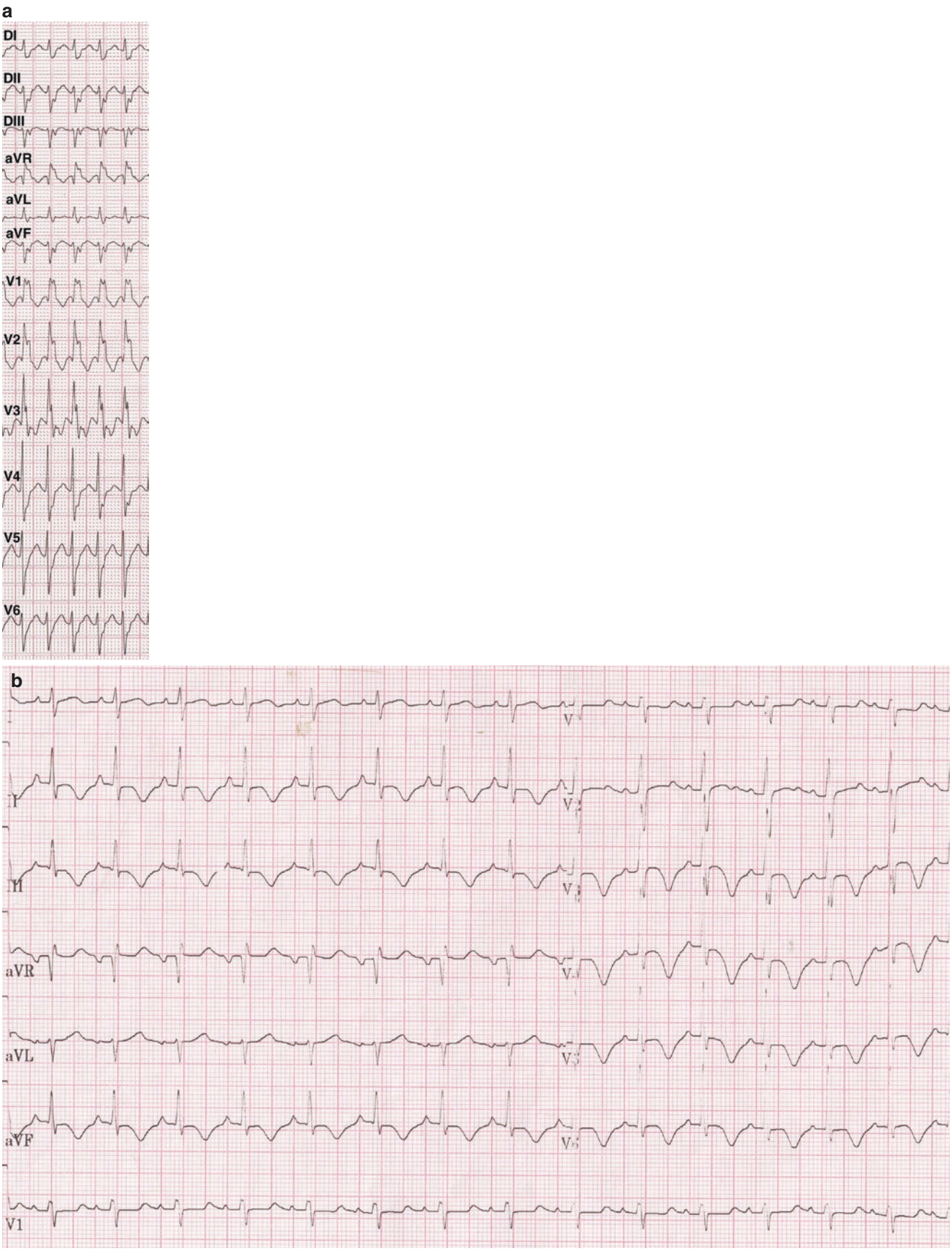

Fig. 12.18 (**a**) Another example of ILVT in a 15-year-old boy. The typical morphology RBBB and left axis deviation can be noted. (**b**) Verapamil was effective. In sinus rhythm there are widespread negative T waves and QT prolongation: this is the electric memory, common after a sustained VT

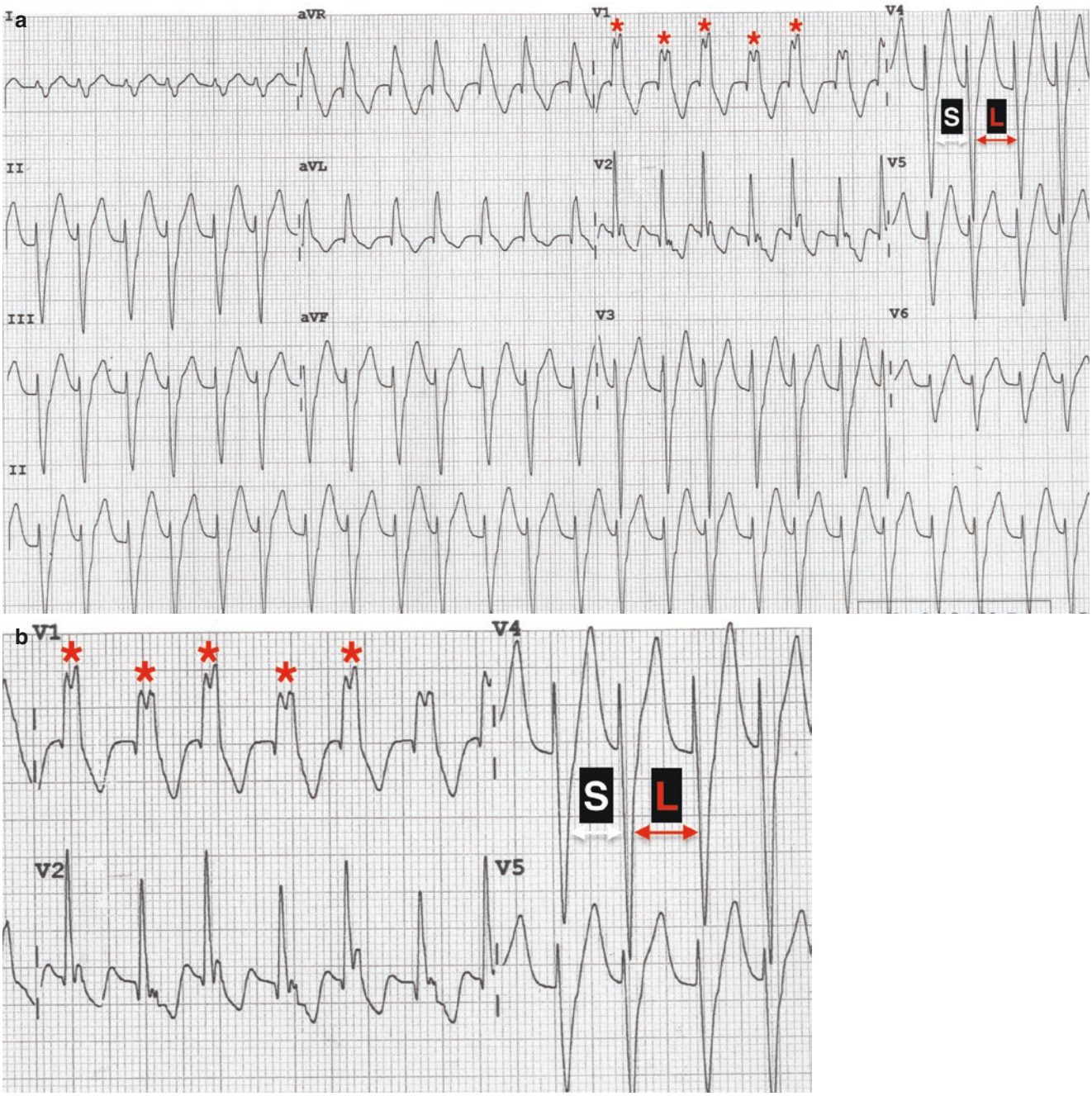

Fig. 12.19 (**a**) Eighteen-year-old male with ILVT. A subtle alternance between amplitude duration and cycle length of QRS can be noted, especially in V1. (**b**) Close-up. L = long cycle; S = short cycle

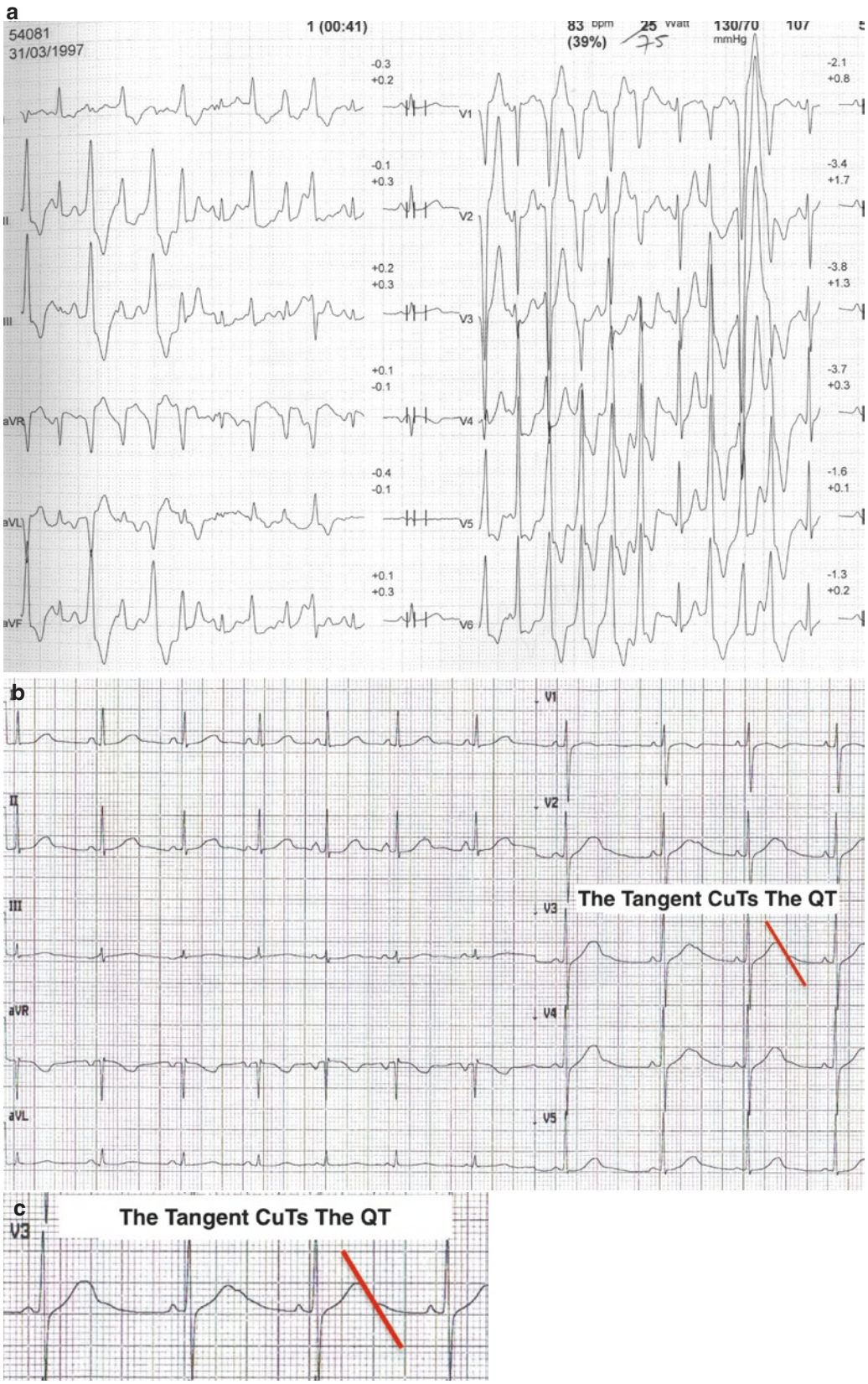

Fig. 12.20 (**a**) Thirteen-year-old male with syncopal episodes during effort. The exercise test found polymorphic VT that was defined catecholaminergic VT. (**b**) Resting ECG: initially the boy was believed a LQTS patient, but cutting the T wave, the QTc returned to normal range (note that in the sentence *the tangent cuts the QT, every word contains a T, as well as every sword* contains *the QT*). (**c**) Close-up. Genetic testing is negative. Currently he is on treatment with beta blocker and flecainide

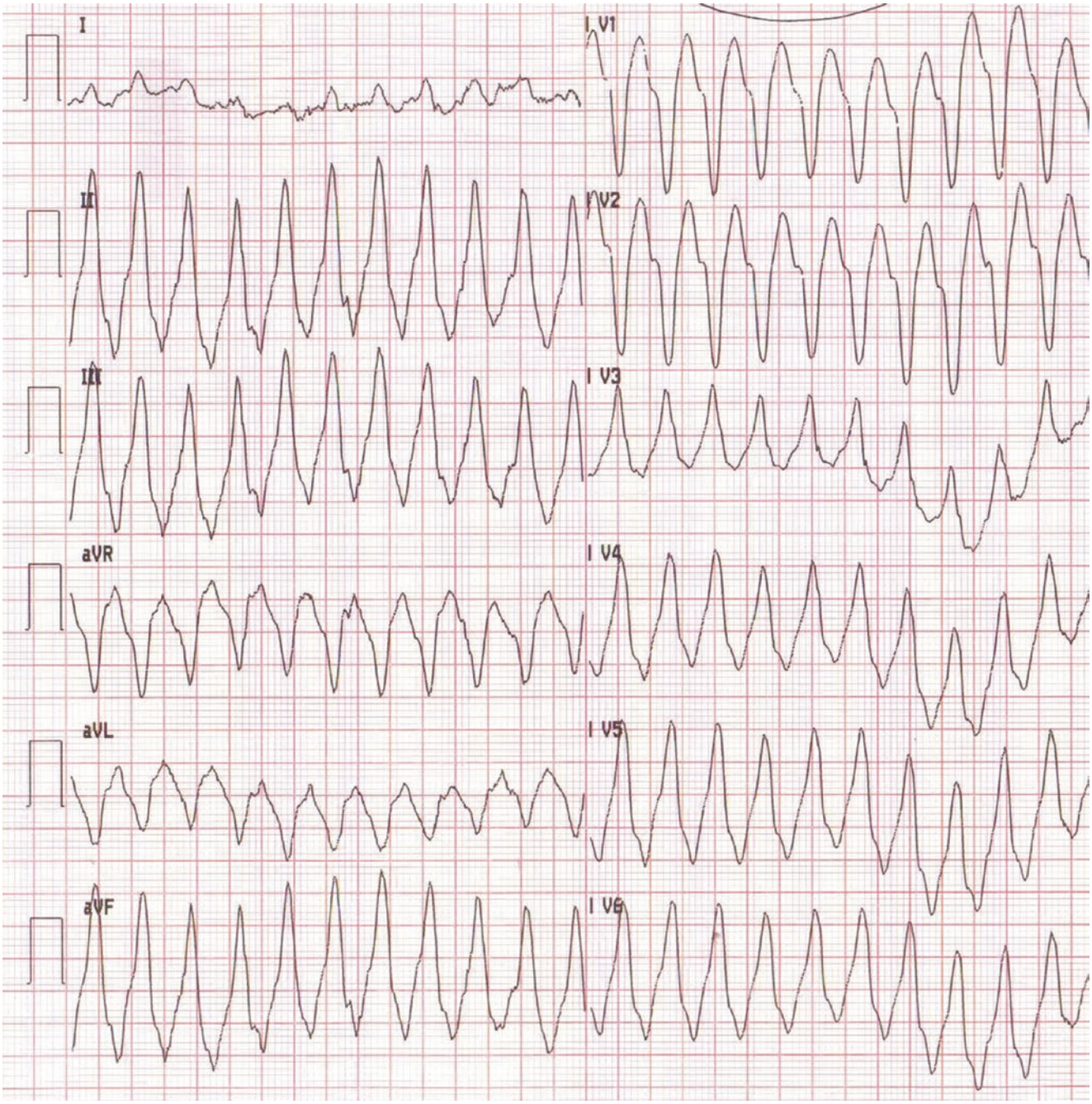

Fig. 12.21 Fourteen-year-old female with LVNC cardiomyopathy symptomatic for palpitation. During the exercise test it was possible to reproduce a wide complex tachycardia. In this case, although the echocardiography and the cardiac MRI localized the disease only to the left ventricle, the arrhythmia is compatible with an origin from the right ventricle (LBBB and vertical axis), in other words RVOT VT. Therefore, any apparently unilateral cardiomyopathy can be shared by both ventricles. Thus an apparently right cardiomyopathy (e.g., ARVC) does not rule out contralateral involvement and left-sided arrhythmias; likewise a right-sided arrhythmia (e.g., RVOT) does not exclude left heart disease (as a non-compaction cardiomyopathy in this case)

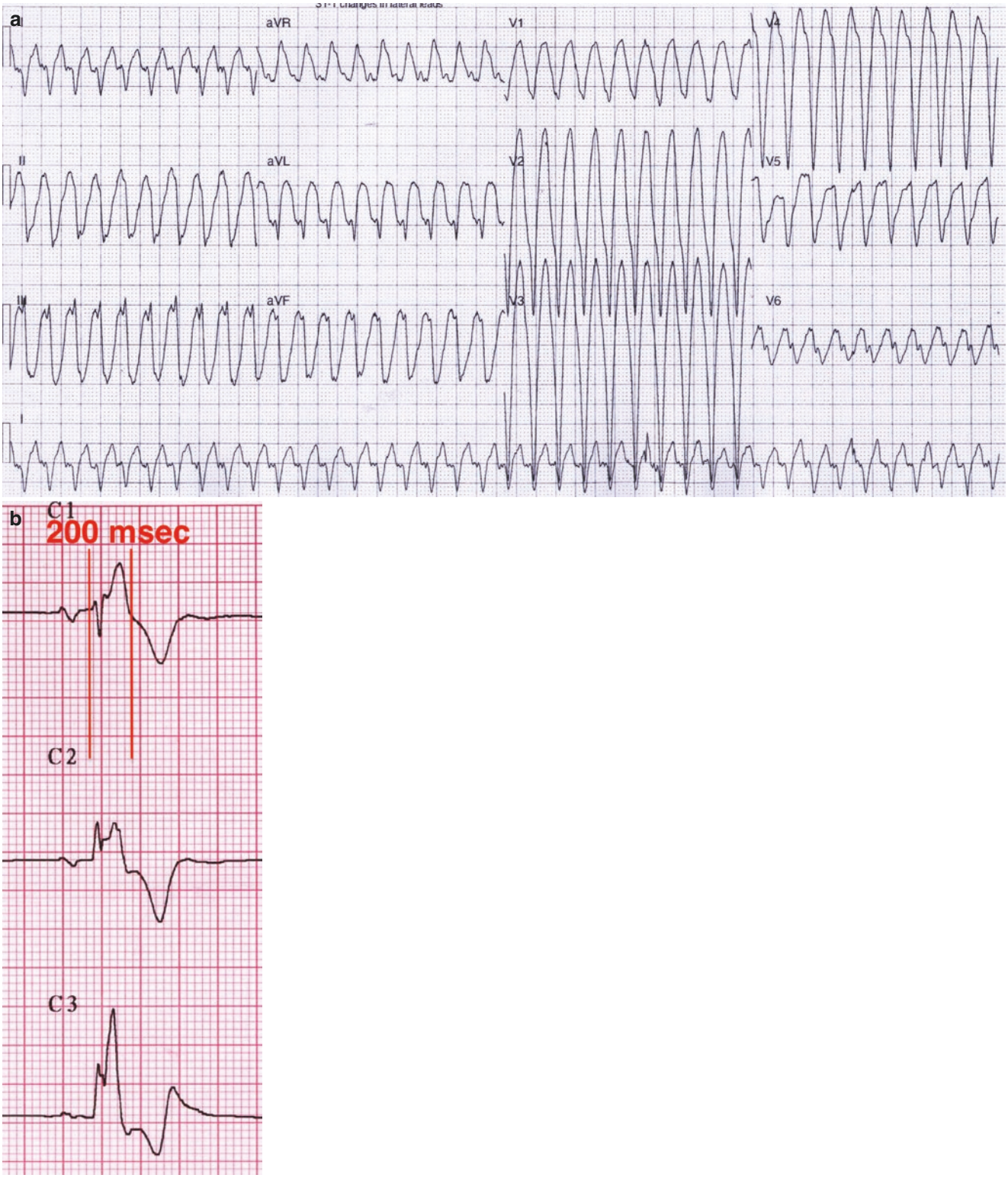

Fig. 12.22 (**a**) Thirty-five-year-old male with repaired tetralogy of Fallot. Wide complex tachycardia, HR 235 bpm, negative precordial concordance, overall LBBB-like morphology, mostly positive aVR; VT diagnosis is overt even without clear AV dissociation or presence of other criteria. The pharmacological cardioversion was effective. The patient underwent surgical correction of tetralogy of Fallot in infancy without important hemodynamic sequelae. (**b**) The basal ECG showed the typical RBBB with QRS duration of 200 ms. In operated tetralogy of Fallot, a good correlation between QRS duration >180 ms and the risk of severe ventricular arrhythmias has been demonstrated. Typically, we find classic incisional arrhythmias originating in the septum and in the right outflow tract, sites of patches and sutures

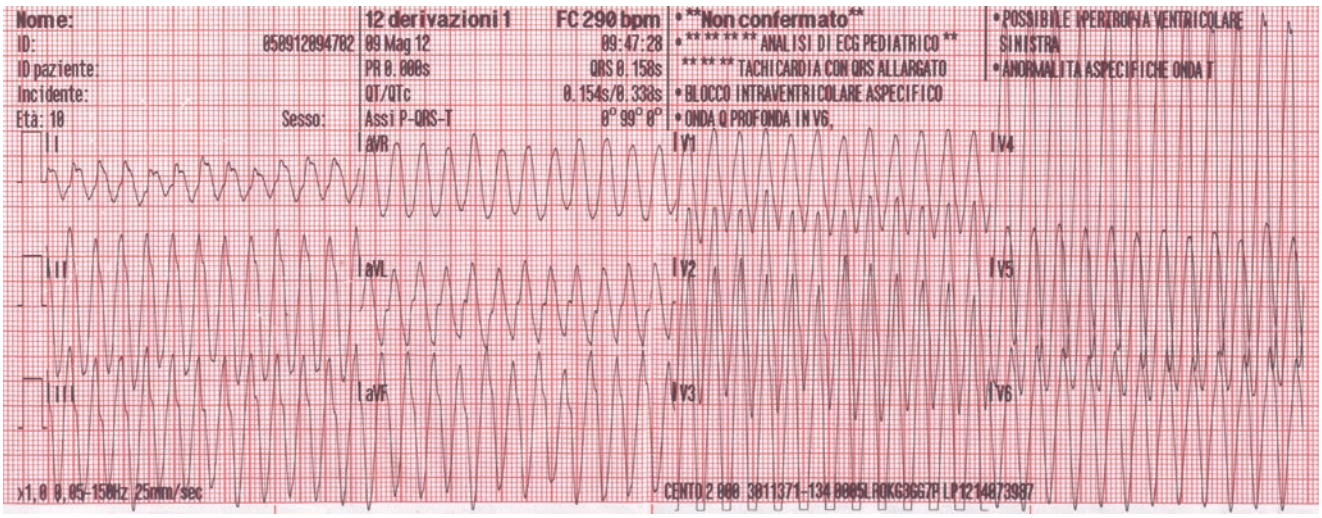

Fig. 12.23 Sixteen-year-old male with severe neonatal cerebral hypoxia. Wide complex tachycardia, HR 290 bpm. The differential diagnosis must take VT or antidromic AVRT into consideration. The negative concordance of the precordial leads is more suggestive of VT. The adenosine was not effective and the patient was successfully treated with DC shock. Despite a nondiagnostic echocardiogram, the heart should have suffered the same ischemia as the brain and may have some area of fibrosis as a substrate for a VT like that

Fig. 12.24 (**a**) Forty-year-old female in the postoperative course of complex CHD: there are PVCs and after a sequence with short-long-short cycle length, there is a torsades de pointes start. (**b**) Ventricular fibrillation in an 11-year-old male with complete AVB and bradycardia-dependent QT prolongation, waiting for PMK replacement

References

1. Pfammatter JP, Paul T. Idiopathic ventricular tachycardia in infancy and childhood: a multicenter study on clinical profile and outcome. Working Group on Dysrhythmias and Electrophysiology of the Association for European Pediatric Cardiology. J Am Coll Cardiol. 1999;33:2067–72.
2. Engelfriet P, Boersma E, Oechslin E, et al. The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period. The Euro Heart Survey on adult congenital heart disease. Eur Heart J. 2005;26:2325–33.
3. Gatzoulis MA, Balaji S, Webber SA, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet. 2000;356:975–81.
4. Tester DJ, Ackerman MJ. Cardiomyopathic and channelopathic causes of sudden unexplained death in infants and children. Annu Rev Med. 2009;60:69–84.
5. Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes: document endorsed by HRS, EHRA, and APHRS in May 2013 and by ACCF, AHA, PACES, and AEPC in June 2013. Heart Rhythm. 2013;10:1932–63.
6. Bronzetti G, Chiu C, Stephenson EA, et al. Simultaneous wide and narrow QRS complex tachycardias in a child: what is the mechanism? J Cardiovasc Electrophysiol. 2006;17:560–2.
7. Basso C, Perazzolo M, Rizzo S. Arrhythmic mitral valve prolapse and sudden cardiac death. Circulation. 2015;132(7):556–66.

Suggested Reading

Alexander ME, Berul CI. Ventricular arrhythmias: when to worry. Pediatr Cardiol. 2000;21:532–41.

Priori SG, Blomström-Lundqvist C, Mazzanti A, Blom N, Borggrefe M, Camm J, Elliott PM, Fitzsimons D, Hatala R, Hindricks G, Kirchhof P, Kjeldsen K, Kuck KH, Hernandez-Madrid A, Nikolaou N, Norekvål TM, Spaulding C, Van Veldhuisen DJ, Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC). 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC) Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). Europace. 2015;17:1601–87.

Crosson JE, Callans DJ, Bradley DJ, Dubin A, Epstein M, Etheridge S, Papez A, Phillips JR, Rhodes LA, Saul P, Stephenson E, Stevenson W, Zimmerman F. PACES/HRS expert consensus statement on the evaluation and management of ventricular arrhythmias in the child with a structurally normal heart. Heart Rhythm. 2014;11:e55–78.

Cardiomyopathies and Myocarditis

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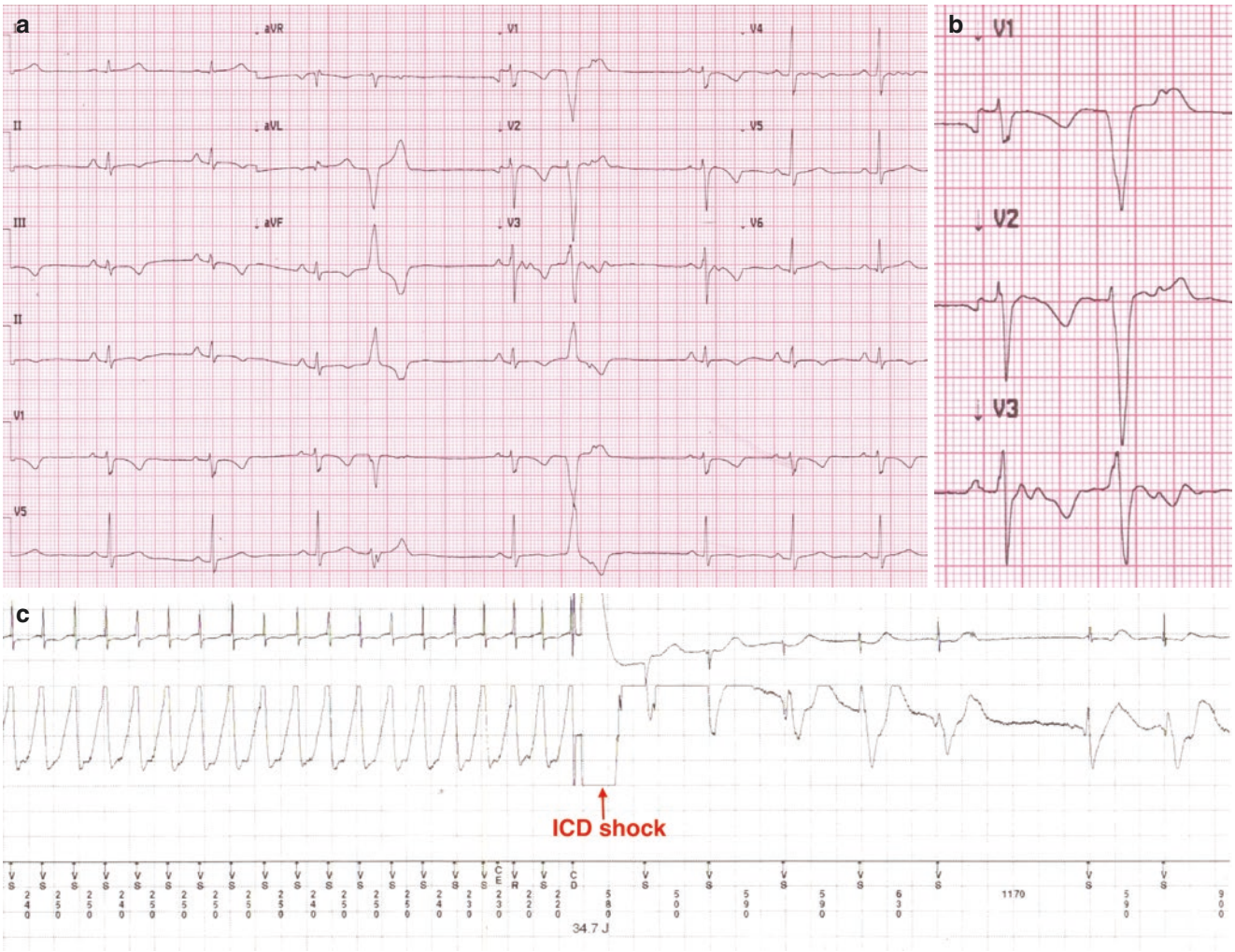

Fig. 13.1 (**a**) Fourteen-year-old boy with incidental finding of abnormal right precordial leads and PVCs. (**b**) Polymorphic PVCs with LBBB morphology (arising from RV). MRI and US found signs of ARVC. (**c**) the patient had a syncopal episode soon after the diagnosis of ARVC and was implanted with an ICD. After 3 months the device discharged a sustained VT

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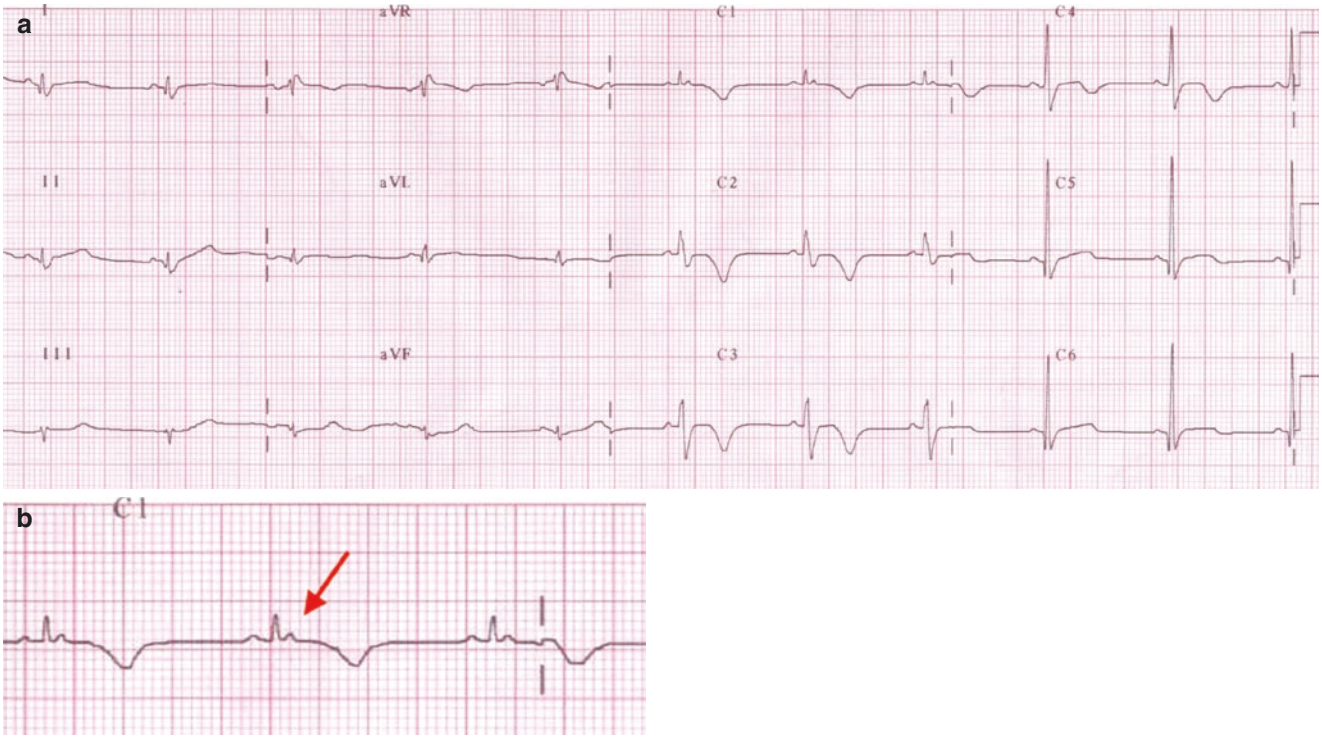

Fig. 13.2 (**a**) Thirteen-year-old male with ARVC. The abnormal T wave in right precordial leads can be noted. The low voltage in the limb leads is also suggestive of ARVC. (**b**) Epsilon wave (*arrow*)

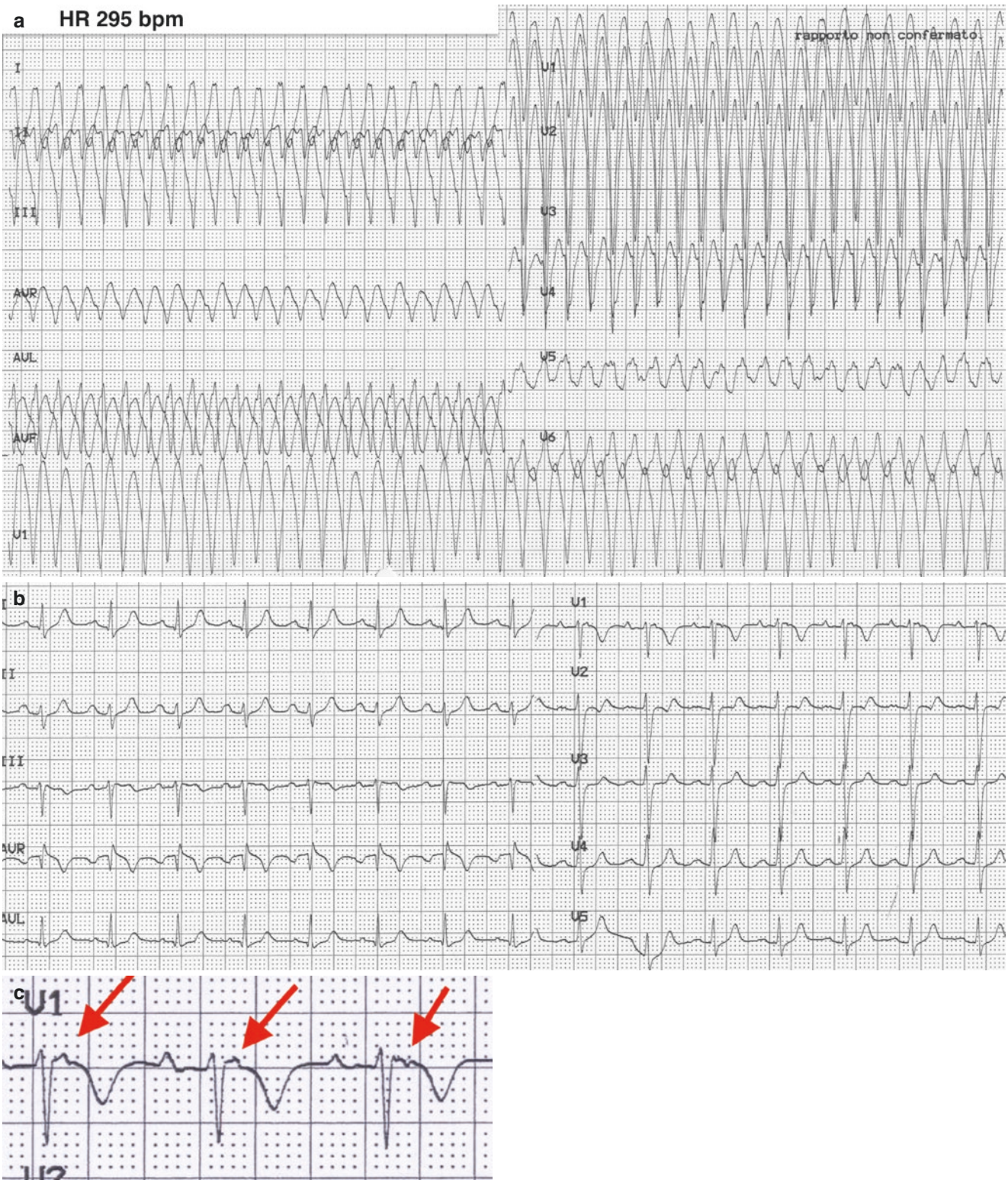

Fig. 13.3 (**a**) Fifteen-year-old male with ARVC. Fast VT as a first manifestation of the disease, (**b**) baseline ECG, (**c**) epsilon wave in V1

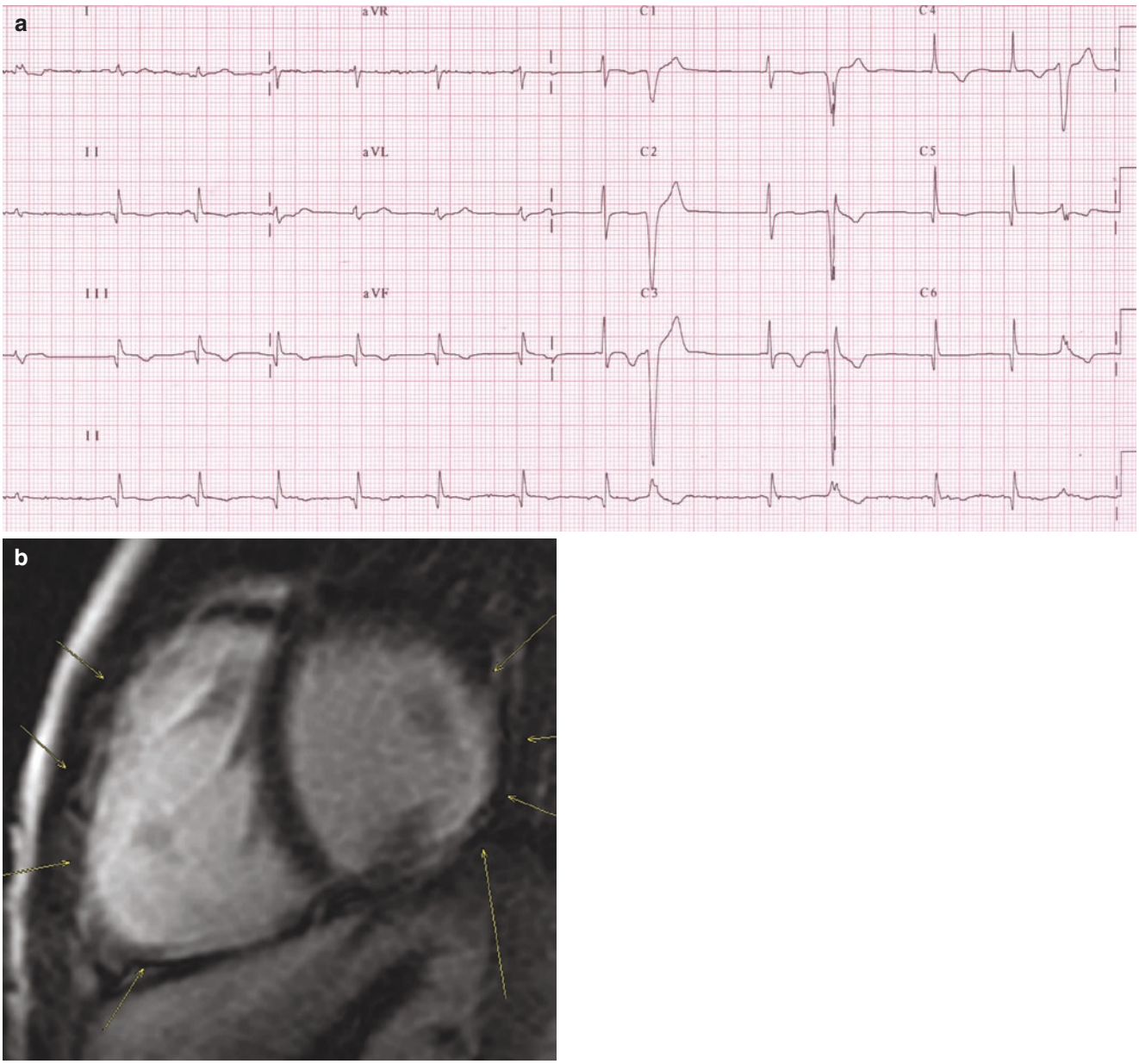

Fig. 13.4 (**a**) Thirteen-year-old male with ARVC with left ventricular extension. Diffuse low voltages, pathologic Q waves (inferior leads), and ST–T changes, PVCs can be noticed. (**b**) The MRI showed severe biventricular fibrosis

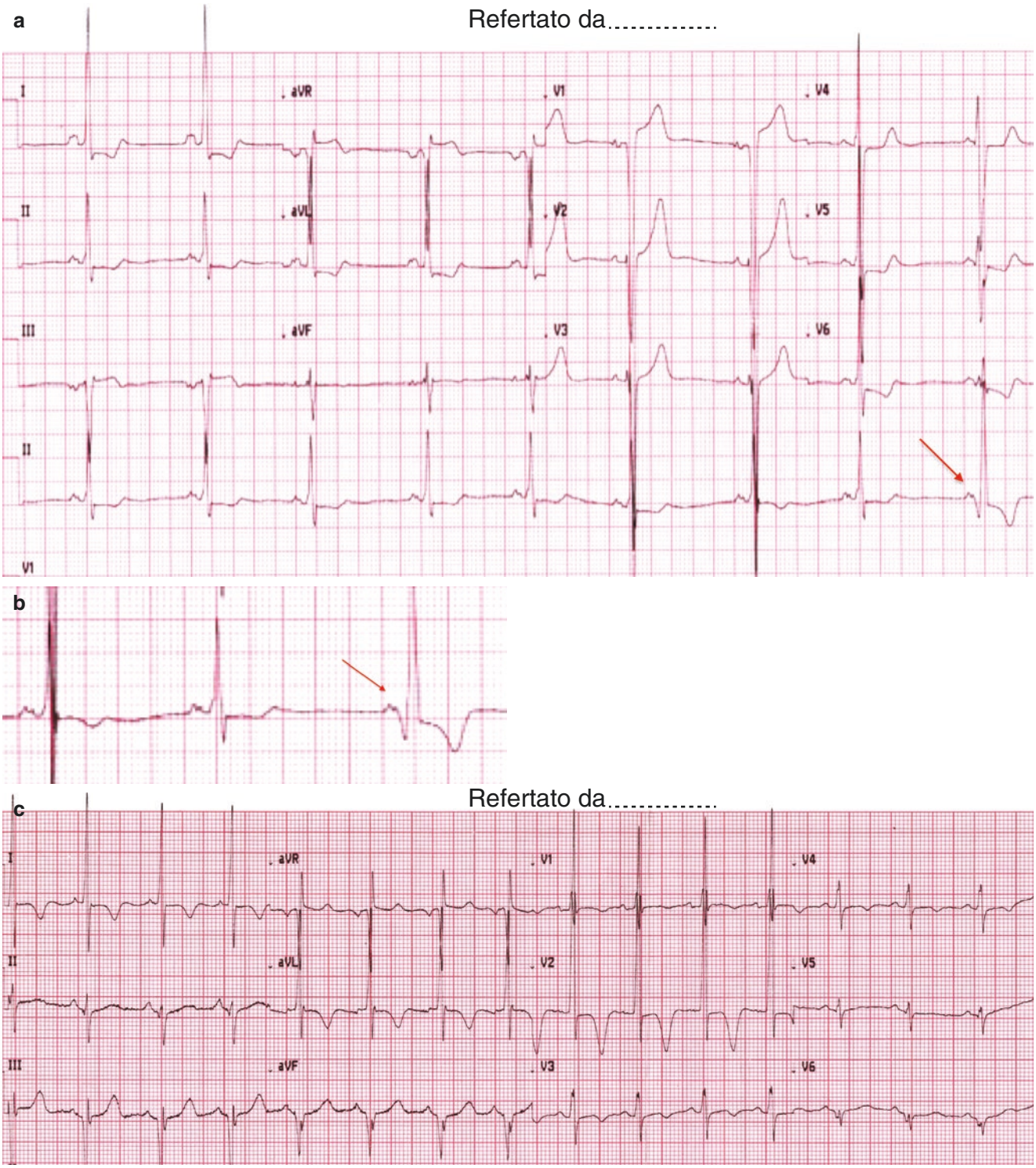

Fig. 13.5 (**a**) Fifteen-year-old girl with HCM associated with ventricular preexcitation. The S–T T changes are secondary to both conditions. (**b**) Close-up. The last beat (*arrow*) is different in both depolarization and repolarization, compatible with a fusion beat (PVC) or conduction trough a second accessory pathway. (**c**) Sixteen-year-old girl with massive septal HCM. The *pseudodelta* QRS slurring can be noticed

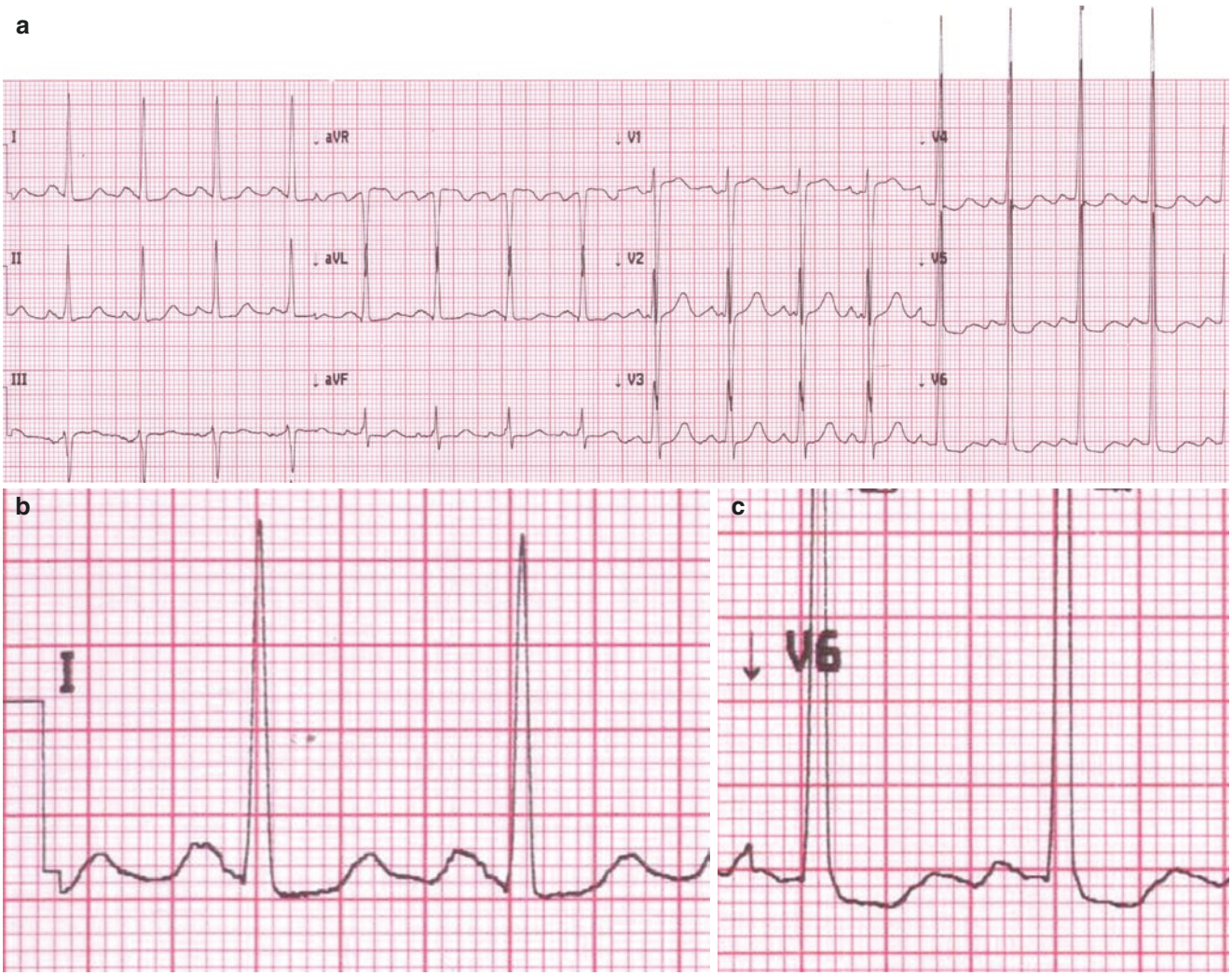

Fig. 13.6 (**a**) DCM in a 9-year-old female. (**b**, **c**) Close-up. Note the diffuse ST–T changes

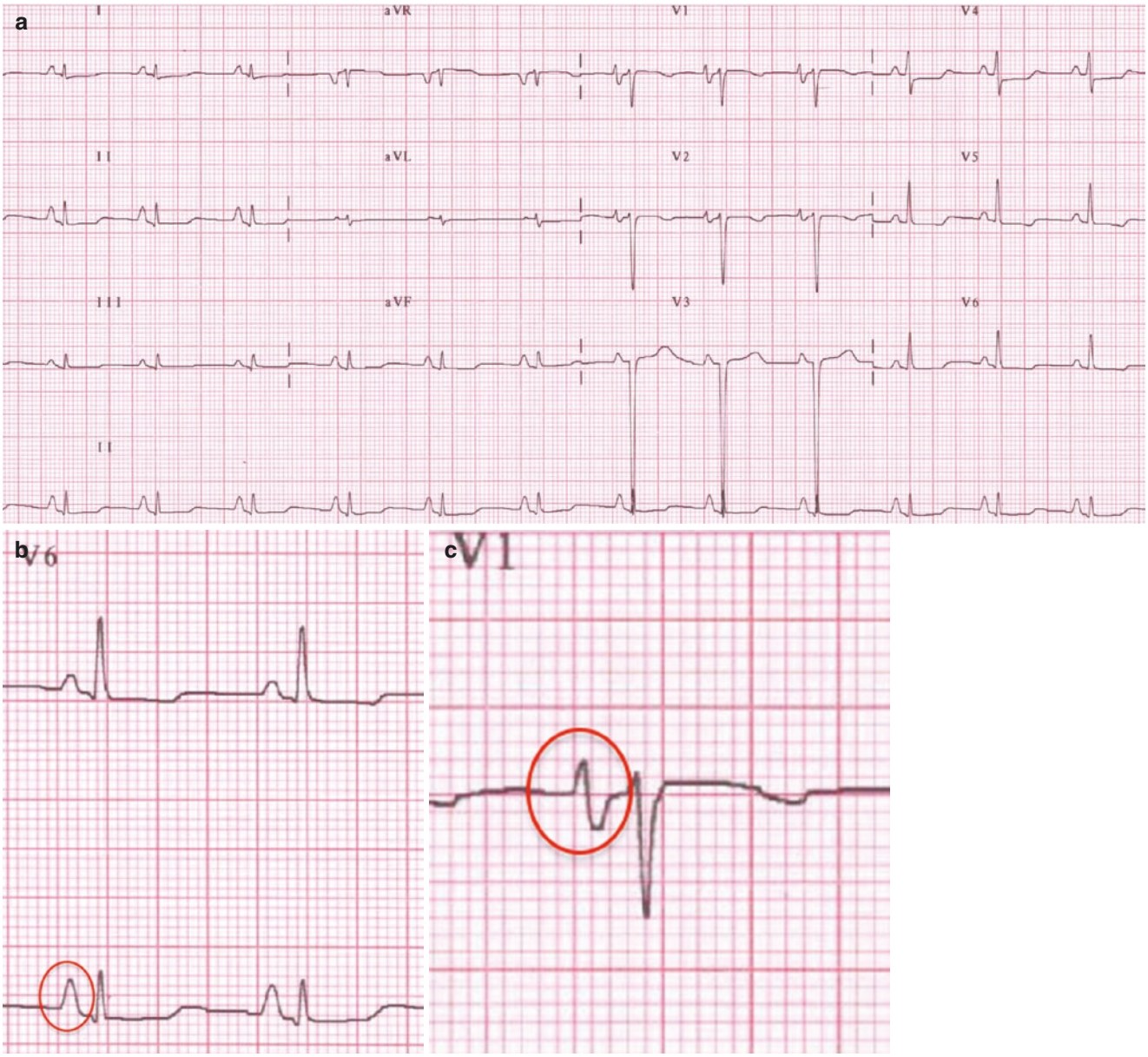

Fig. 13.7 (**a**) DCM secondary to anthracycline therapy for lymphoma. Once again, limb leads low voltages and ST–T changes are suggestive of CMP. (**b**, **c**) Close-up. The atrial enlargement indicates high filling pressure

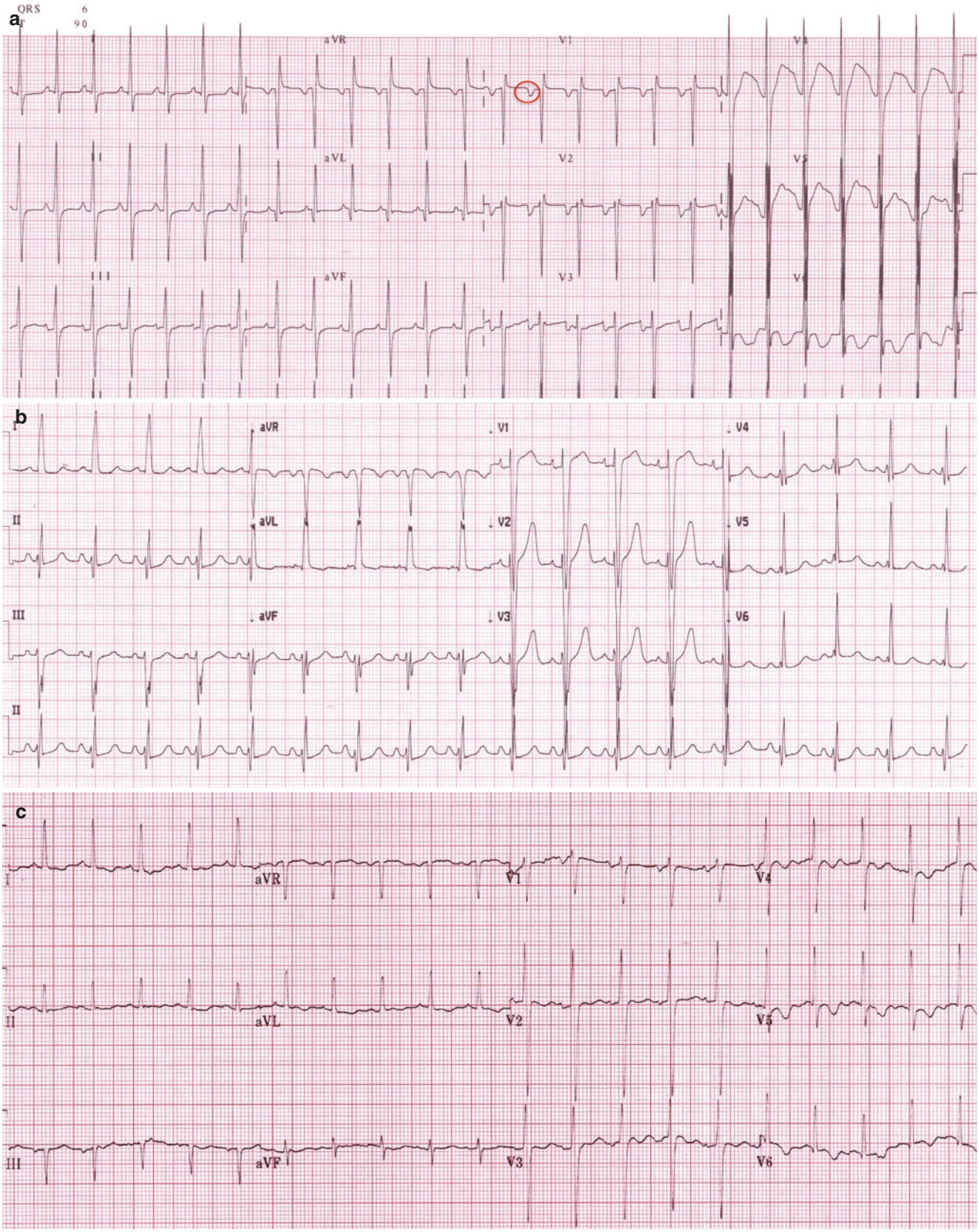

Fig. 13.8 Left ventricular non compaction cardiomyopathy (LVNC). (**a**) Two-year-old male: note the atrial enlargement and ST–T changes. (**b**) Thirteen-year-old female: note the QRS enlargement for intraventricular conduction delay (incomplete LBBB). (**c**) Twenty-two-year-old male with trisomy 13. Note the ST–T changes

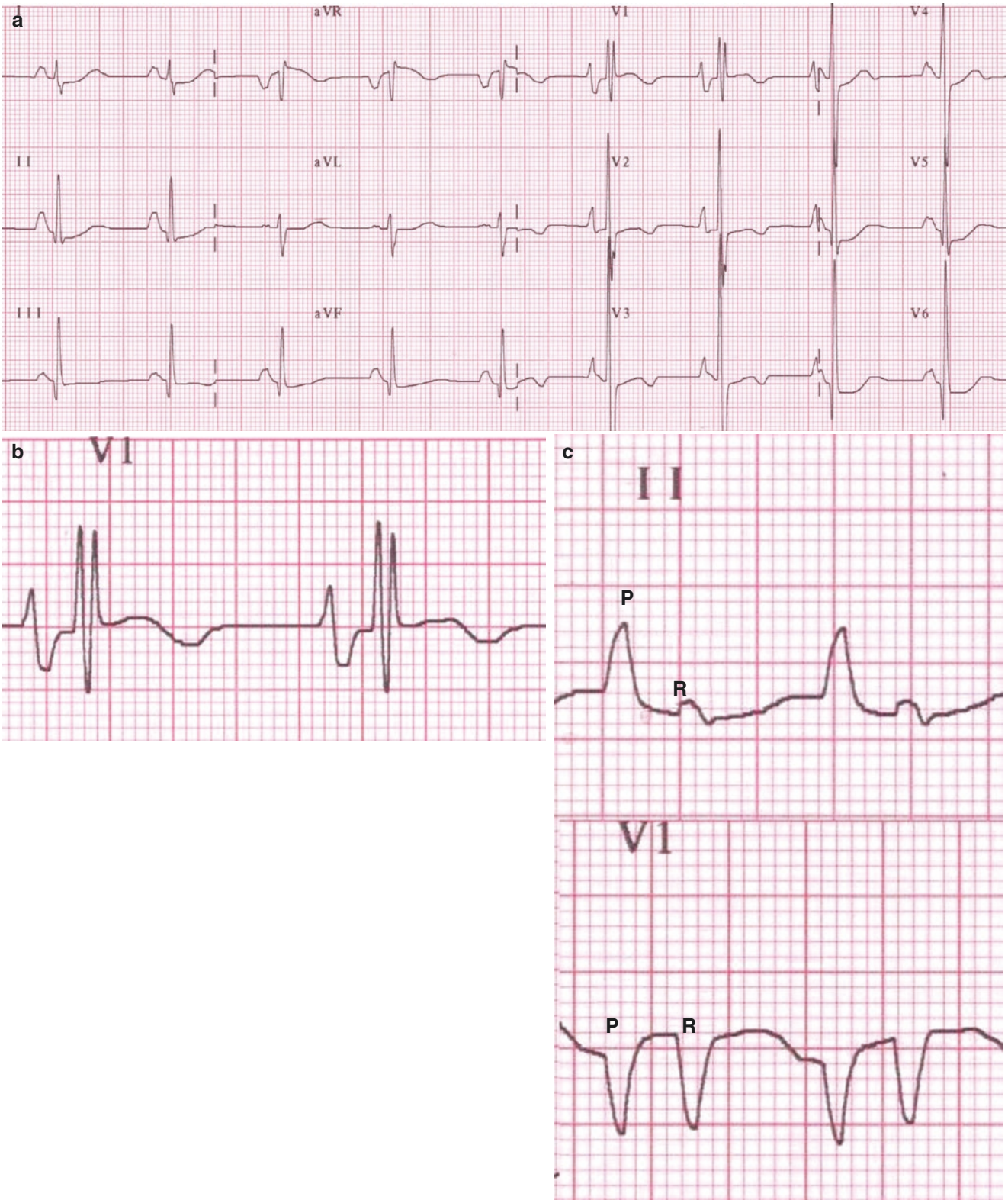

Fig. 13.9 (**a**) Ten-year-old male with restrictive CMP. (**b**) close-up, note the atrial enlargement. (**c**) Restrictive CMP in a 14 year old male. Note the massive biatrial enlargement; in some leads the P wave is bigger than the R wave

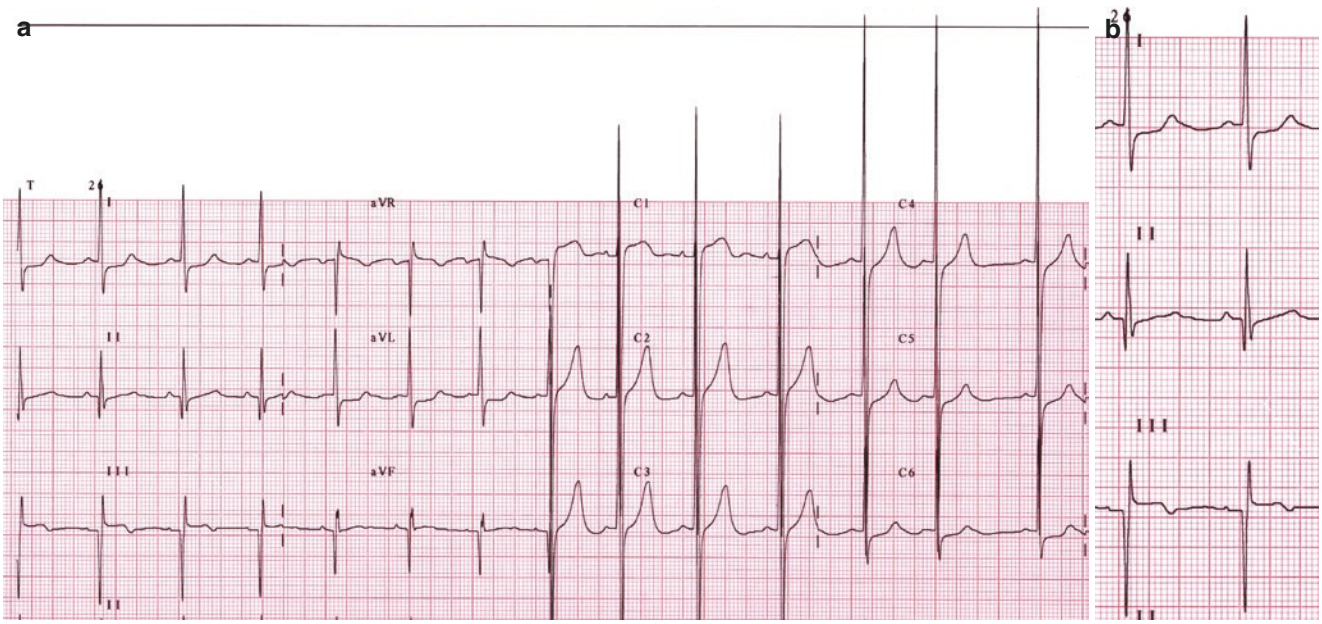

Fig. 13.10 (**a**) Eleven-year-old male with HCM. (**b**) Close-up. Huge Q waves in the inferior leads can be noted

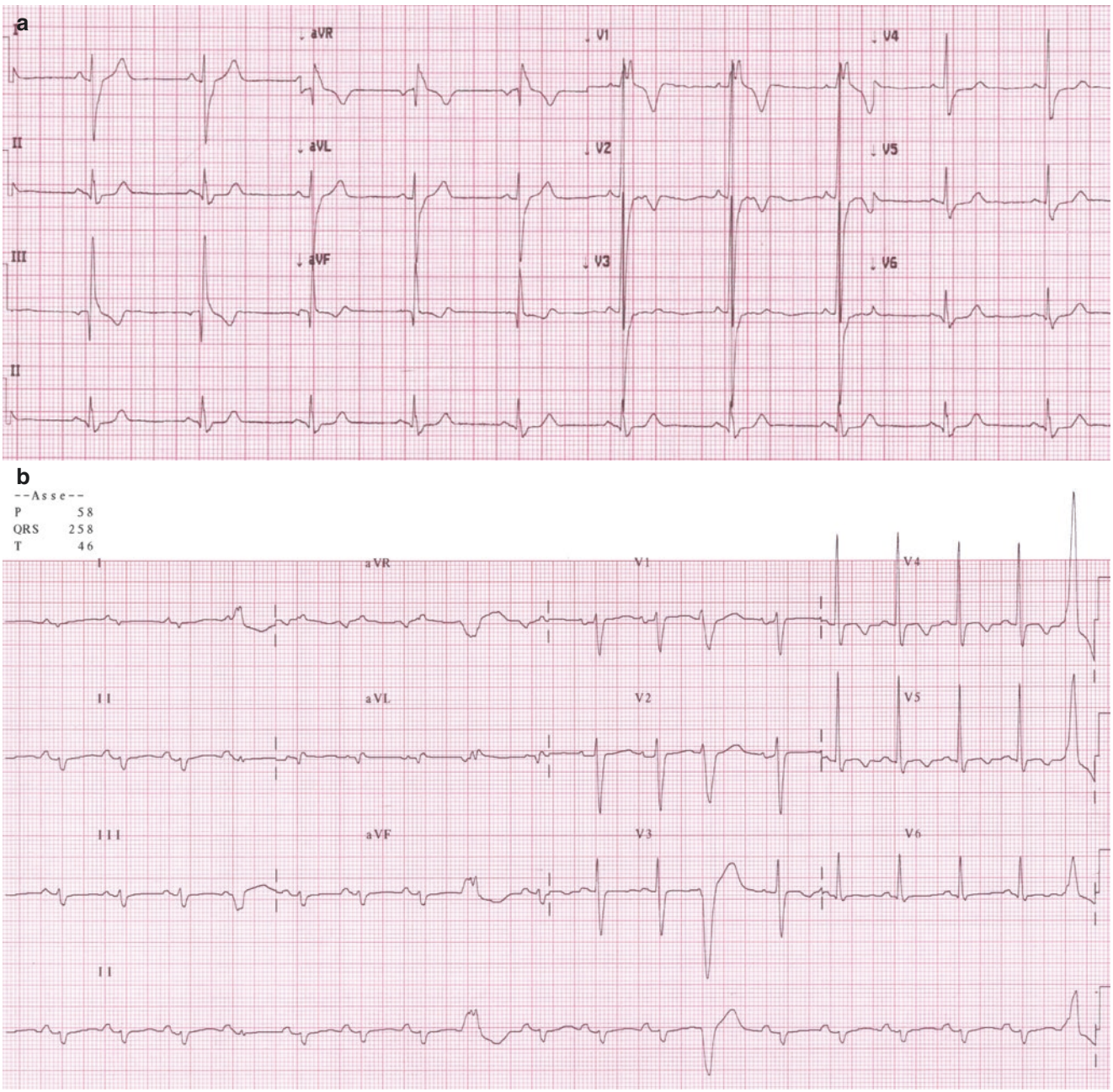

Fig. 13.11 HCM in two young 20-year-old women. (**a**) On echo the ventricular septum is 30 mm thick. The voltages are not as can be expected in a massive hypertrophy like this. There is the arrow sign in the precordial leads (R wave decreasing from V2 to V6. (**b**) Massive hypertrophy on echo but the voltages in limb leads are poor. The MRI showed severe fibrosis, one of the reasons which explain the discrepancy between thickness and ECG voltages

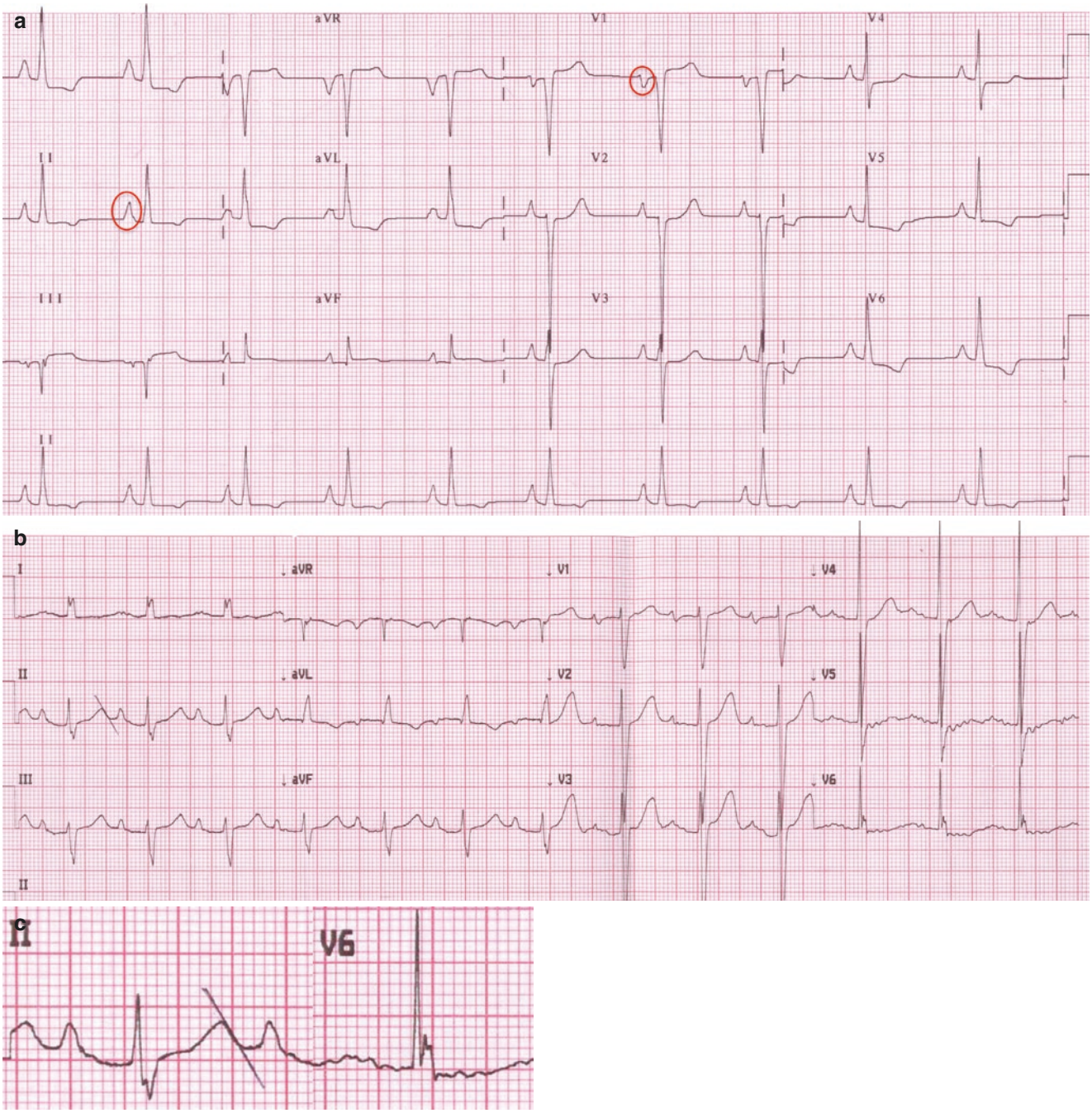

Fig. 13.12 (**a**) Twelve-year-old male with HCM and restrictive physiology. The systolic function is intact, but the patient showed diastolic heart failure. The atrial enlargement can be noted. (**b**) Fifteen-year-old male with restrictive neuropathic HCM. (**c**) Close-up. A long PR and incomplete LBBB can be noticed

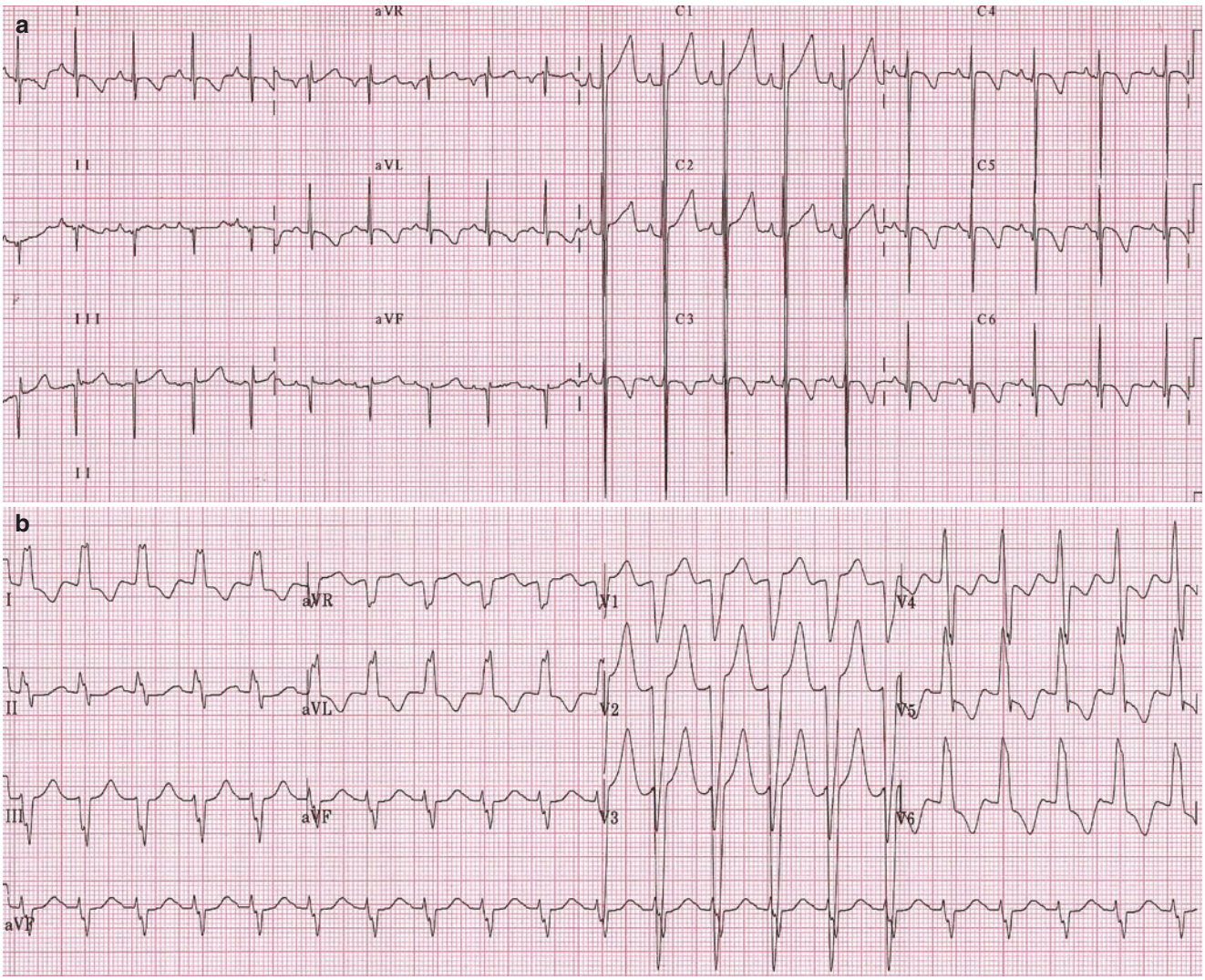

Fig. 13.13 (**a**) Neonatal Noonan syndrome with non-obstructive HCM. Note the left axis deviation and the strain pattern. (**b**) An intermittent LBBB was documented

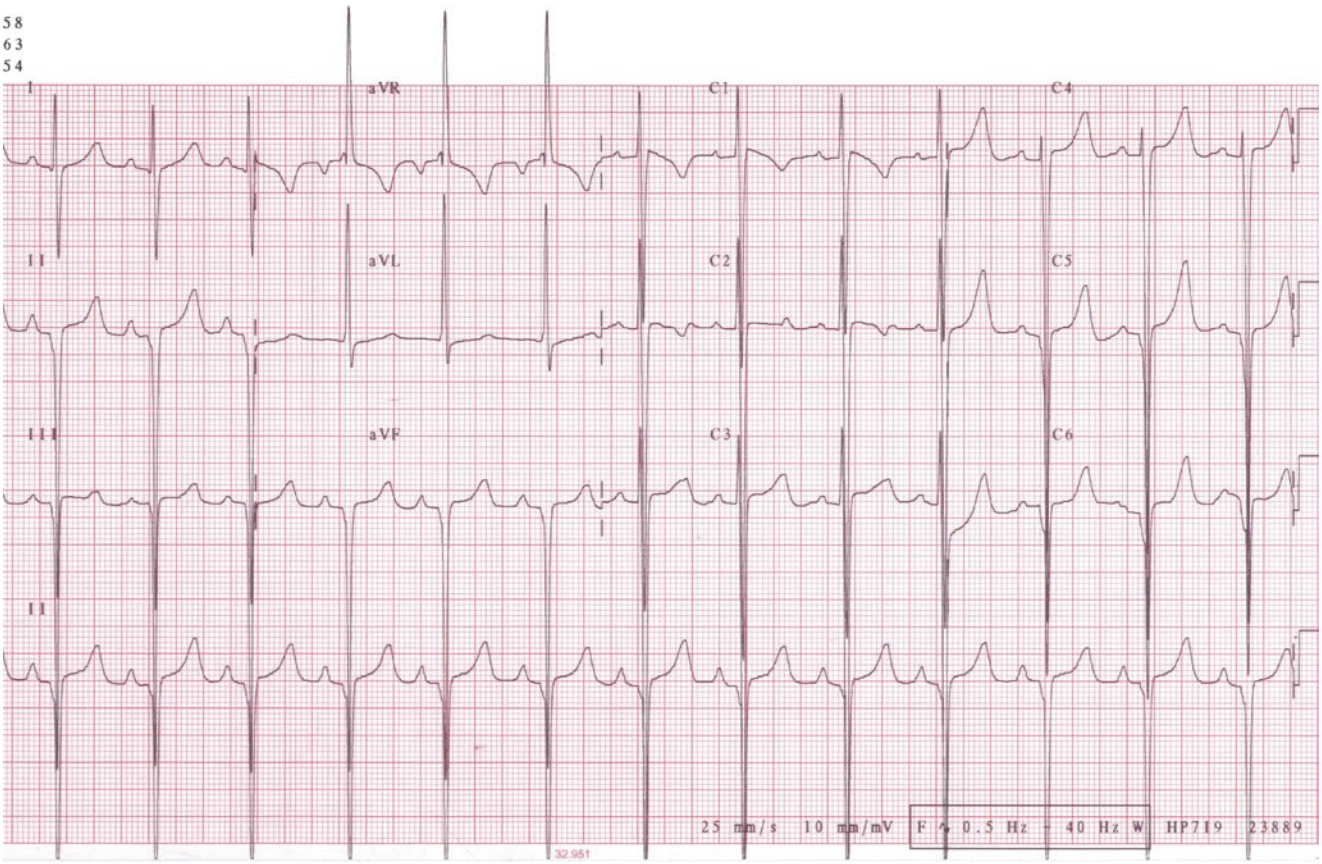

Fig. 13.14 Eight-year-old male. Noonan syndrome with obstructive HCM. The pathognomonic northwest axis deviation can be noted

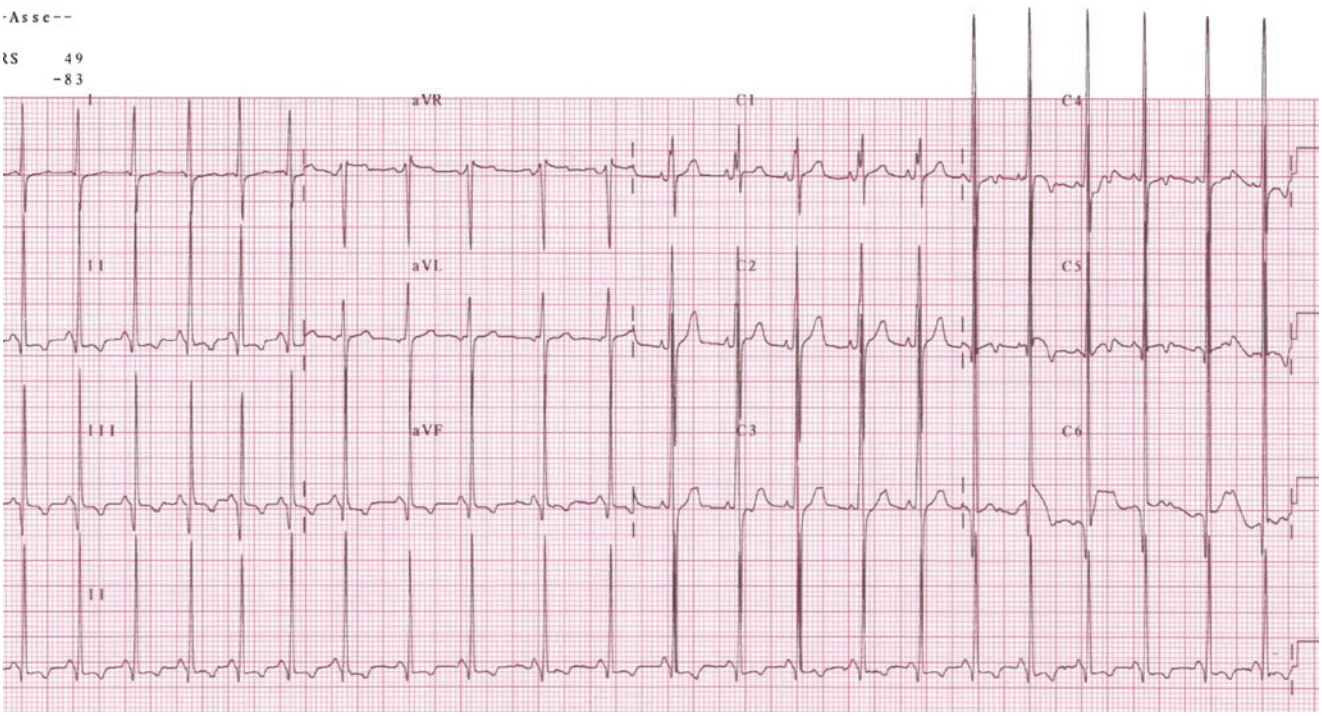

Fig. 13.15 One-year-old male with Pompe disease. The distinctive features of biventricular hypertrophy and short PR can be noted

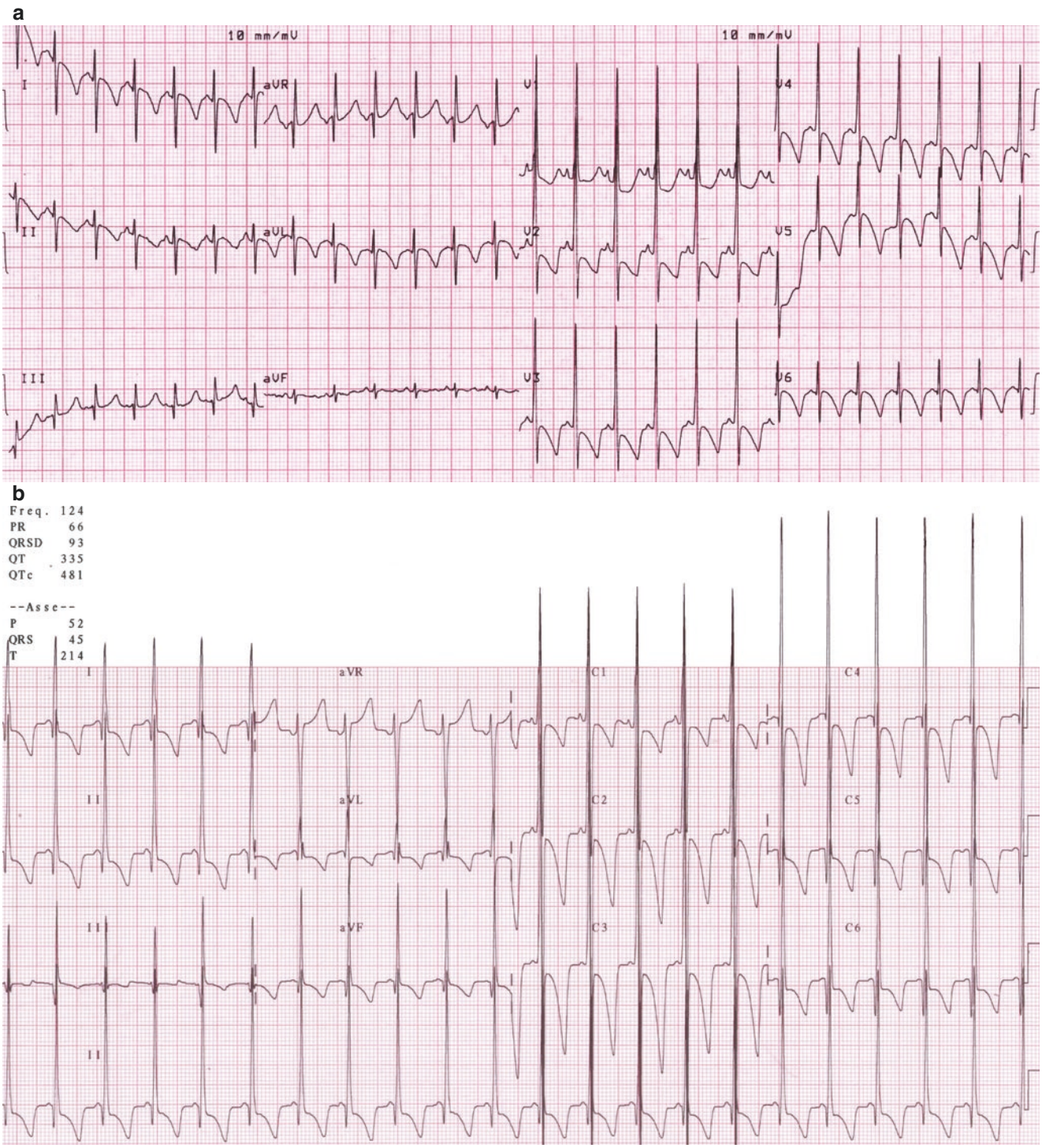

Fig. 13.16 Pompe disease. (**a**) Two-week-old male. The substitutive enzymatic therapy normalized echo and ECG; (**b**) 2-month-old female. The two different patients have very similar ECGs

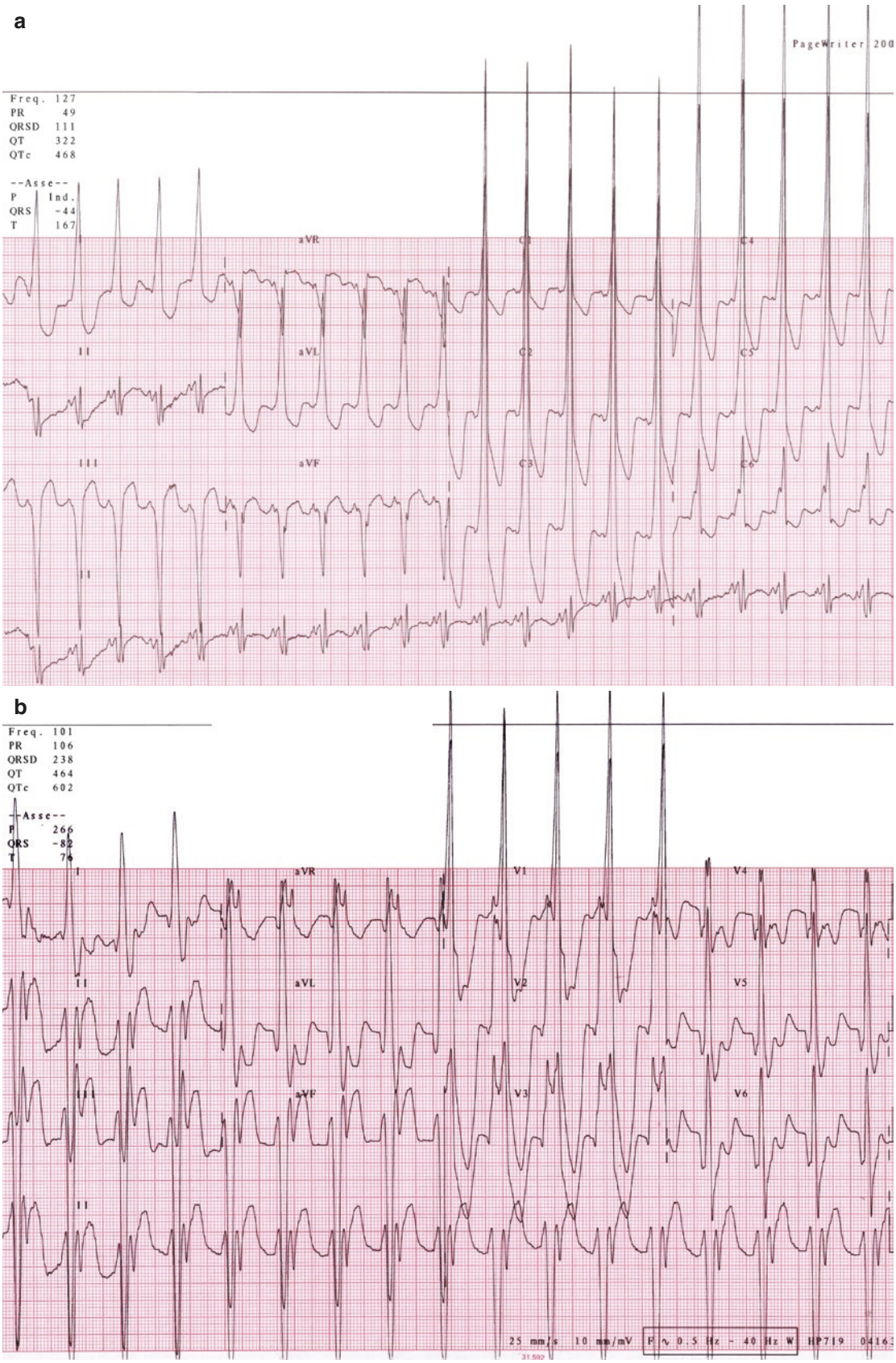

Fig. 13.17 Danon disease (x-linked storage disease due to mutation of LAMP2). (**a**) Six-year-old with severe arrhythmic expression who underwent ICD implant; (**b**) 8-year-old; (**c**) the ECGs are similar and peculiar for enormous voltages and Q wave (pseudonecrosis). As in Pompe disease, a short PR interval can be noted

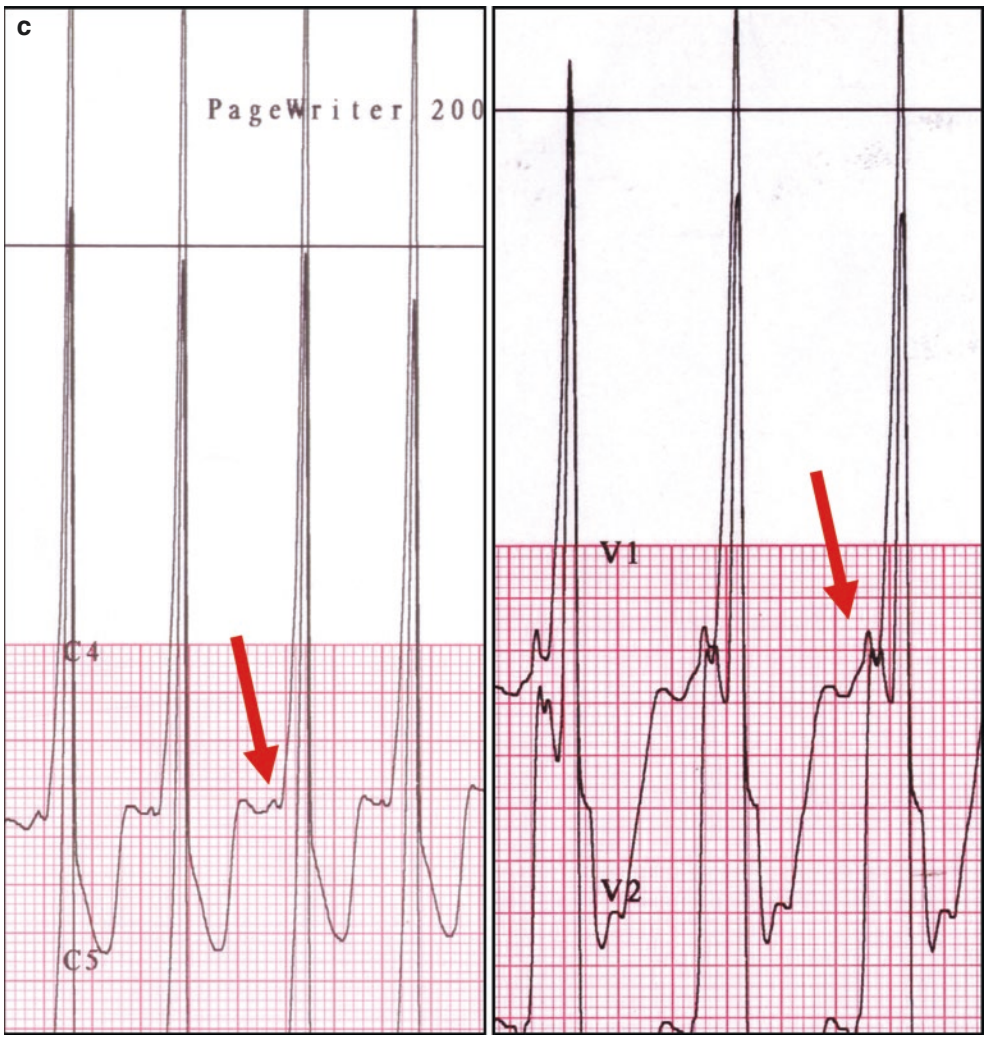

Fig. 13.17 (continued)

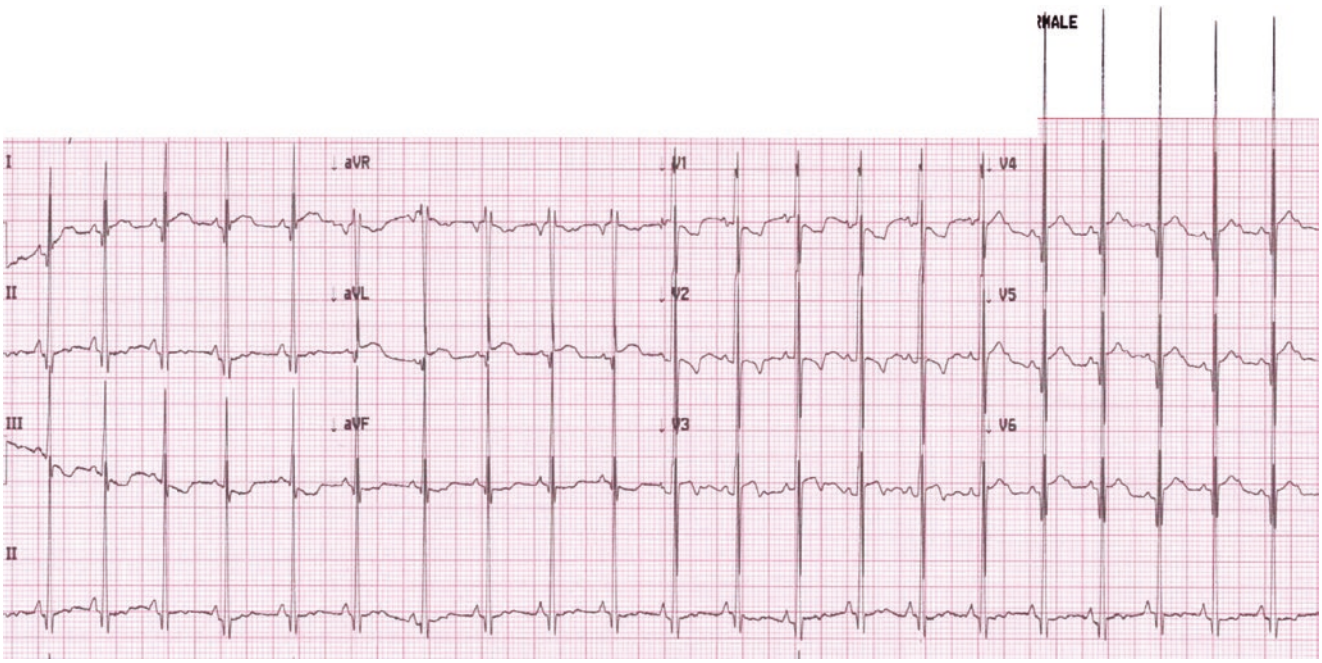

Fig. 13.18 Seven-month-old with mitochondrial disease. Biventricular hypertrophy

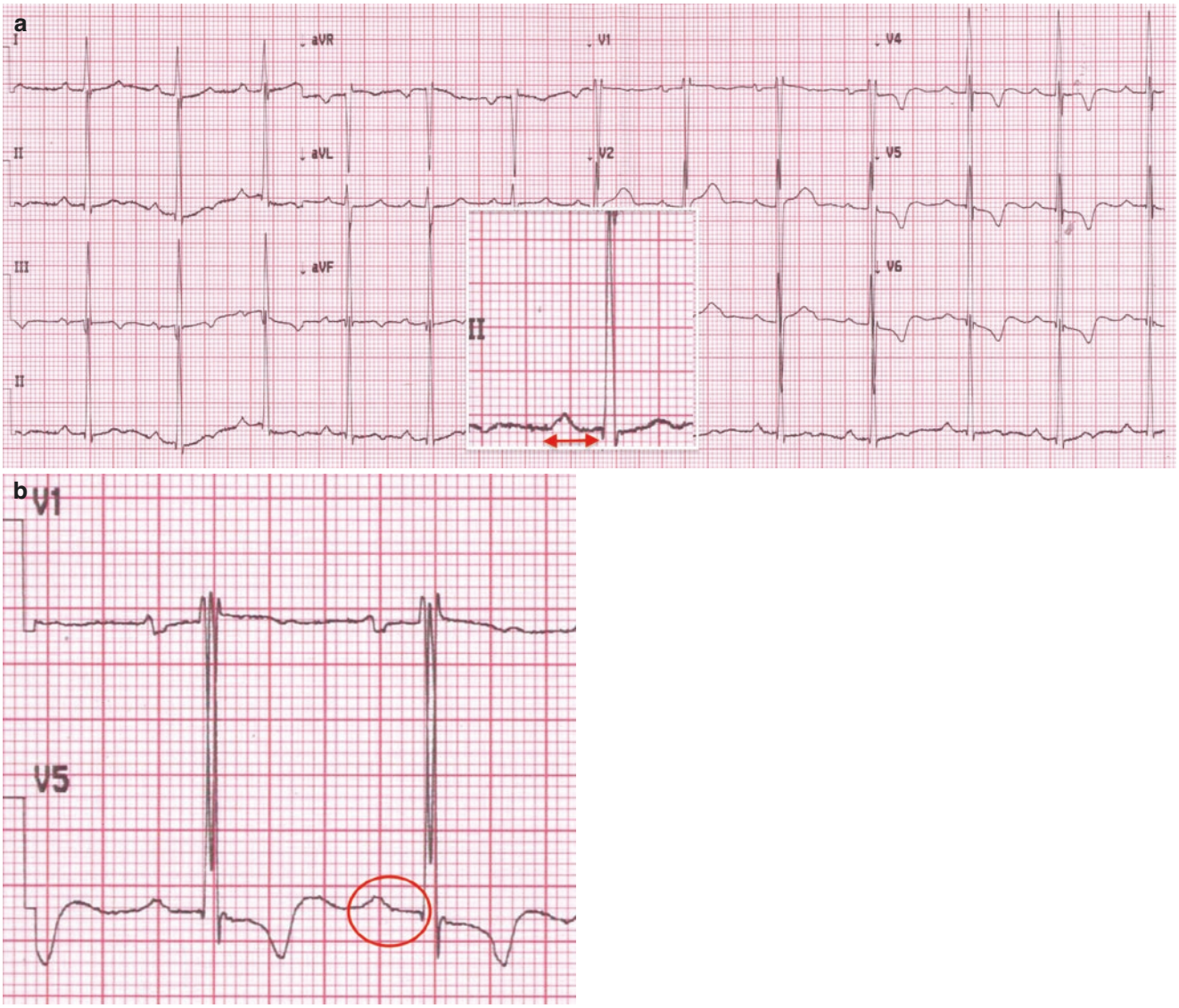

Fig. 13.19 (**a**) Fifteen-year-old male with HCM phenotype with long PR. A PRKAG2-HCM was found. Aside from HCM with short PR, due to association with WPW or in storage disease like Pompe or Danon and PRKAG2 too, there are forms of HCM with PR prolongation. In this case the AV node dysfunction prevailed on the pre-excitation. The finding of a long PR together with left ventricular overload should suggest hypertrophic HCM and laminopathies. Sarcomeric mutations are described with this pattern. It is also necessary to exclude neuromuscular diseases such as Steinert's disease and mitochondrial diseases such as Kearns-Sayre. The negative T wave limited to the precordial leads is characteristic of the apical forms and is also found in the case of isolated papillary muscles hypertrophy of the mitral valve. (**b**) Close-up

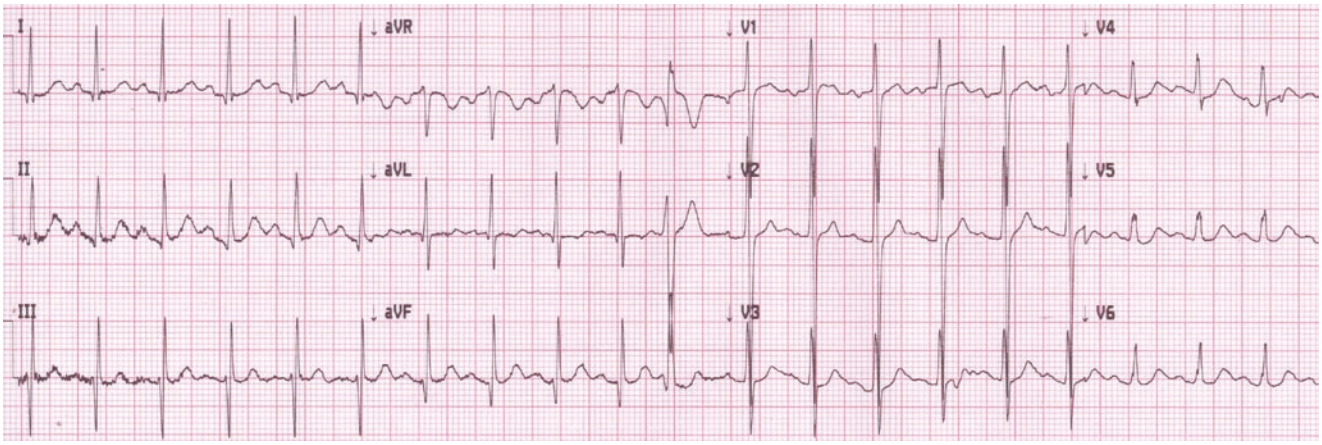

Fig. 13.20 Thirteen-year-old male with HCM. On the echocardiogram the most affected aspect is the septum with reverse septal curvature morphology. On the echocardiogram the maximum thickness is 25 mm. As in other forms of HCM with septal involvement, there is the "arrow" sign: the R wave is decreasing from V2 to V6. Note that the voltages are not impressive or at least not as can be expected knowing the massive hypertrophy present on the echocardiogram

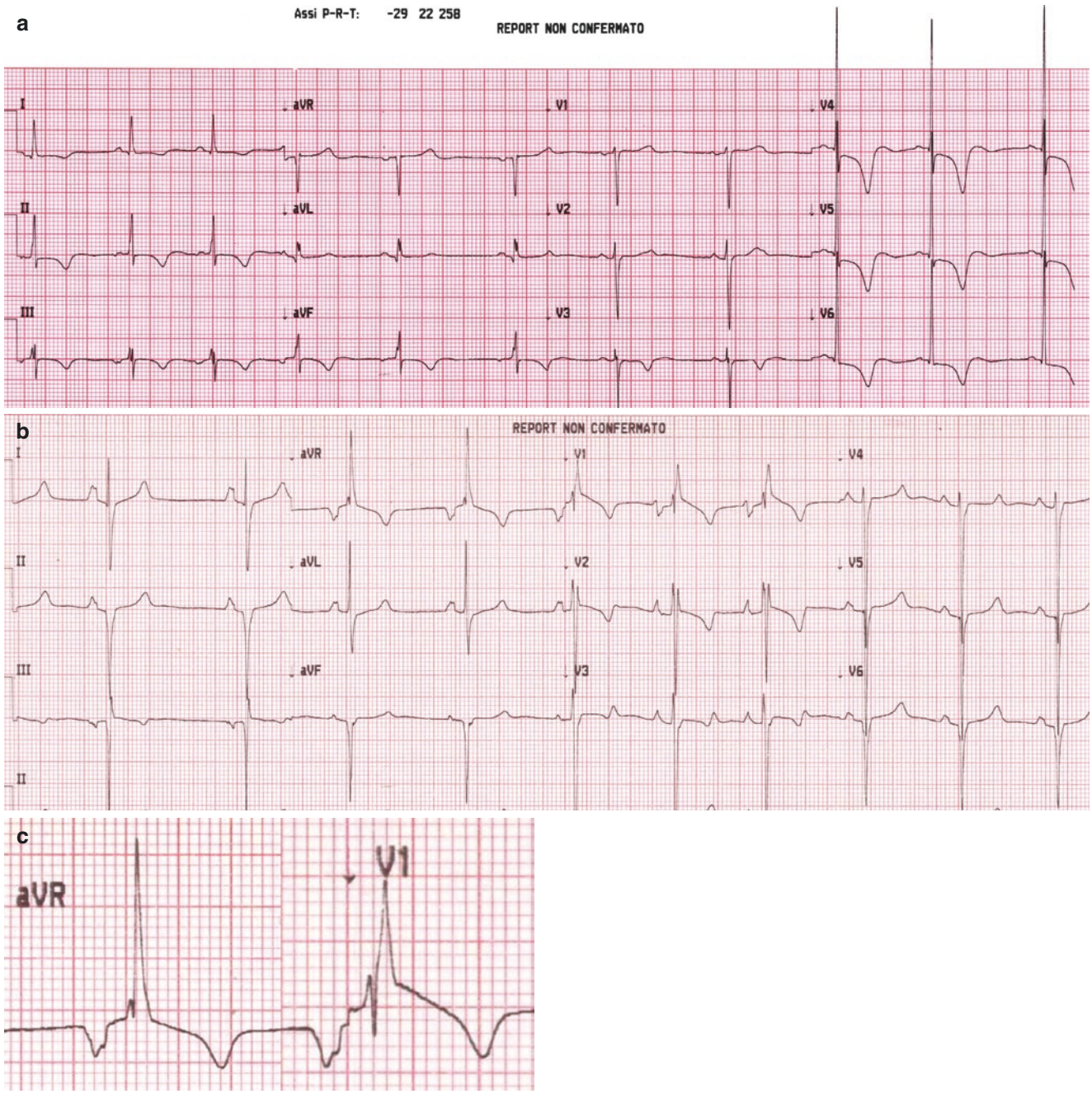

Fig. 13.21 (**a**) Thirteen-year-old male with Noonan syndrome and apical form of HCM; note the deep negative T waves in the midprecordial and left precordial leads. (**b**) Sixteen-year-old female with Noonan syndrome and concentric HCM. Note also the typical northwest axis and the atrial enlargement. (**c**) Close-up

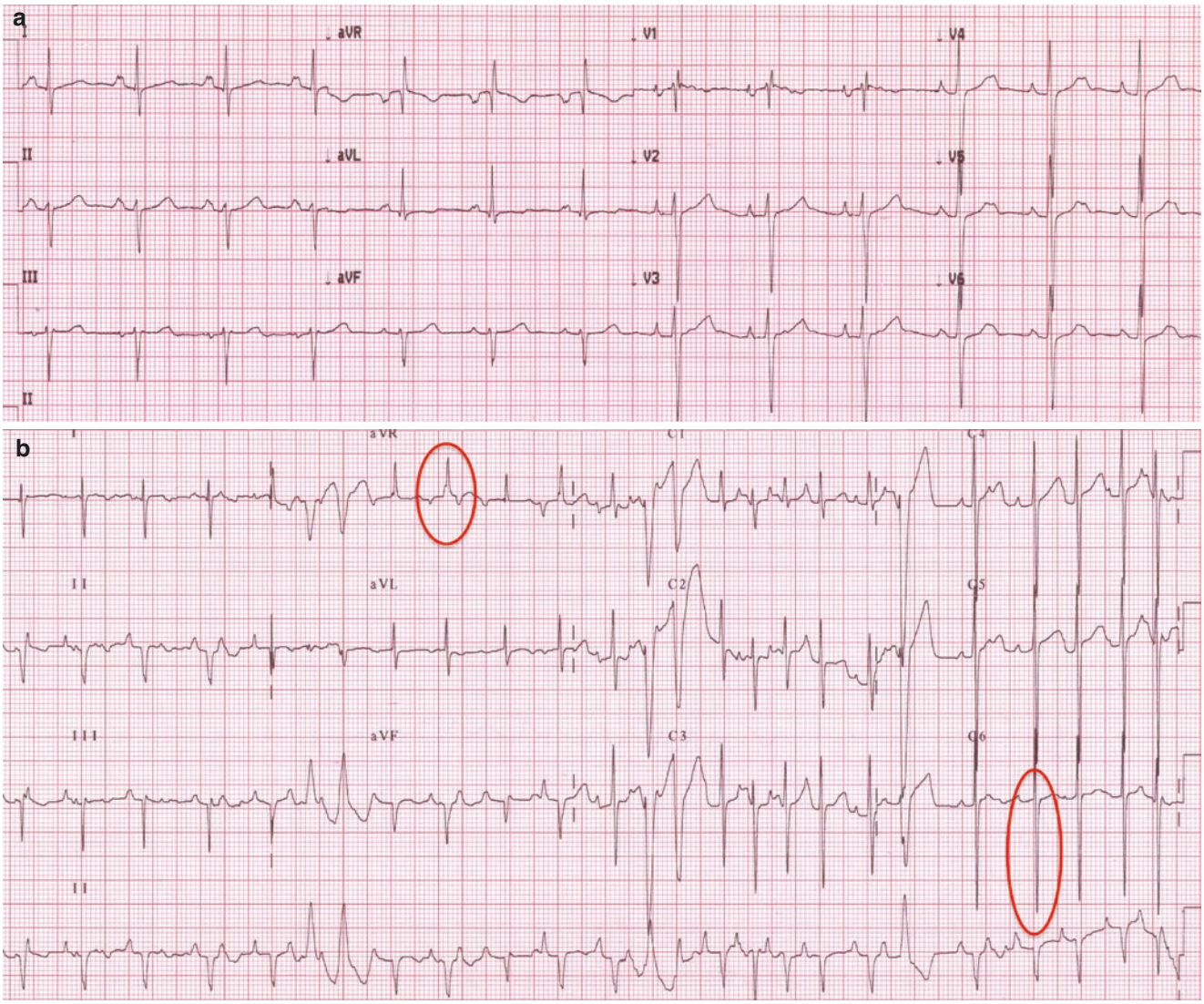

Fig. 13.22 (**a**) Two-year-old male with Noonan syndrome-HCM. (**b**) Burst of non-sustained EAT; some beats are conducted with aberrancy. Note the northwest axis and the large S in V6, the "S" of Noonan

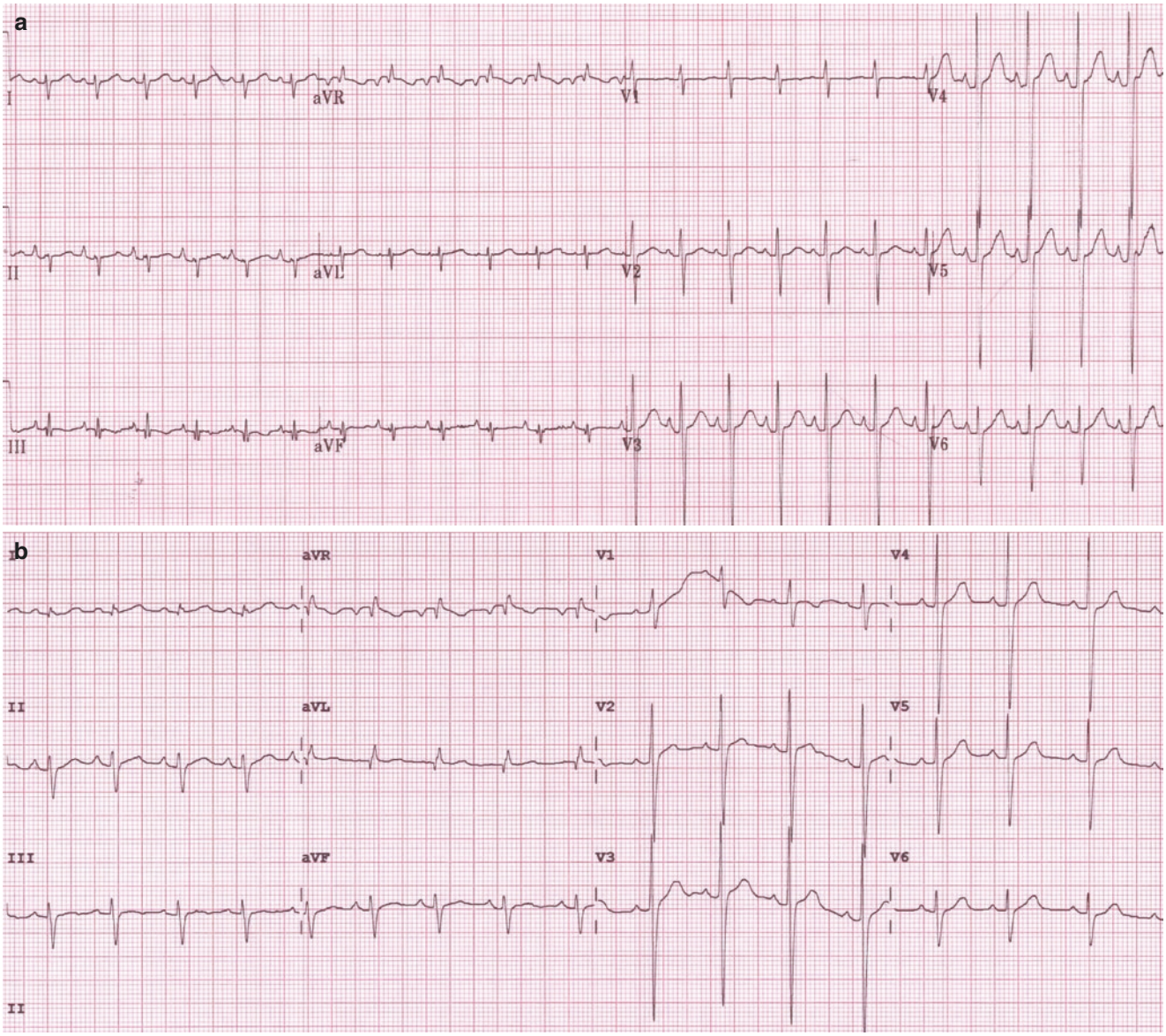

Fig. 13.23 Steinert myotonic dystrophy. (**a**) Neonate. Despite the absence of detectable heart disease there are features similar to Noonan syndrome, such as extreme axis deviation and large S in V6. (**b**) Two-year-old male with the abovementioned features

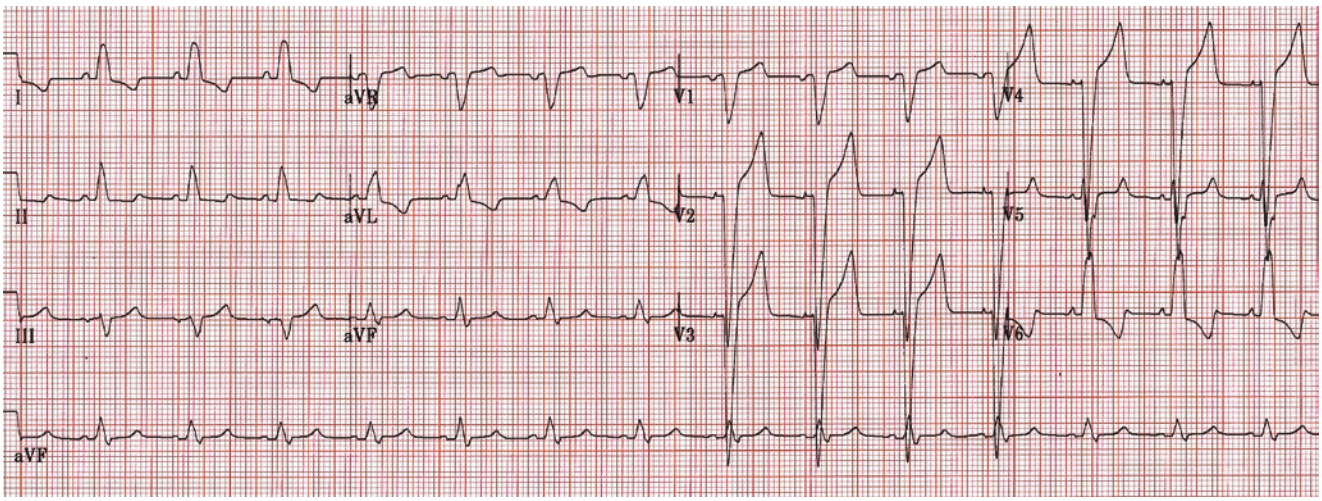

Fig. 13.24 Fourteen-year-old female. Septal HCM after Morrow myectomy. A LBBB developed

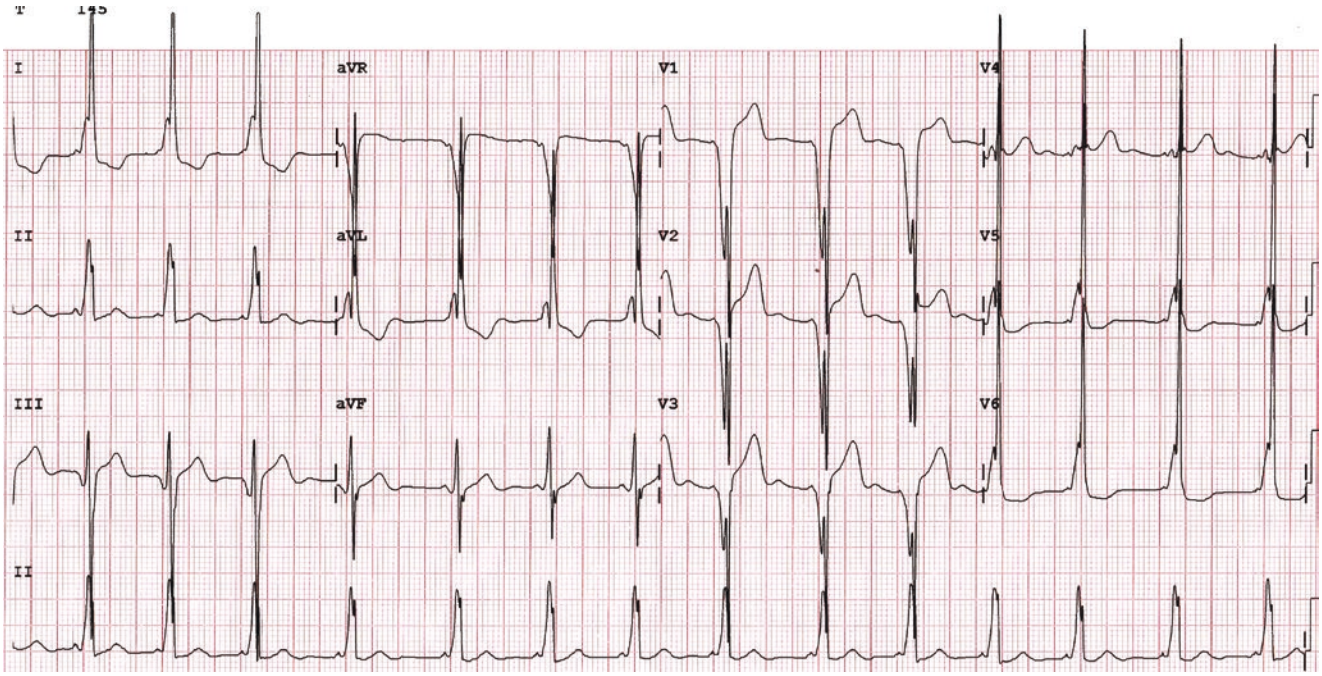

Fig. 13.25 Fifteen-year-old male with Duchenne dystrophy (MD). A distinctive trait of DMD may be a short PR interval. In this case there is a true delta wave with ventricular pre-excitation, as seen in other CMPs

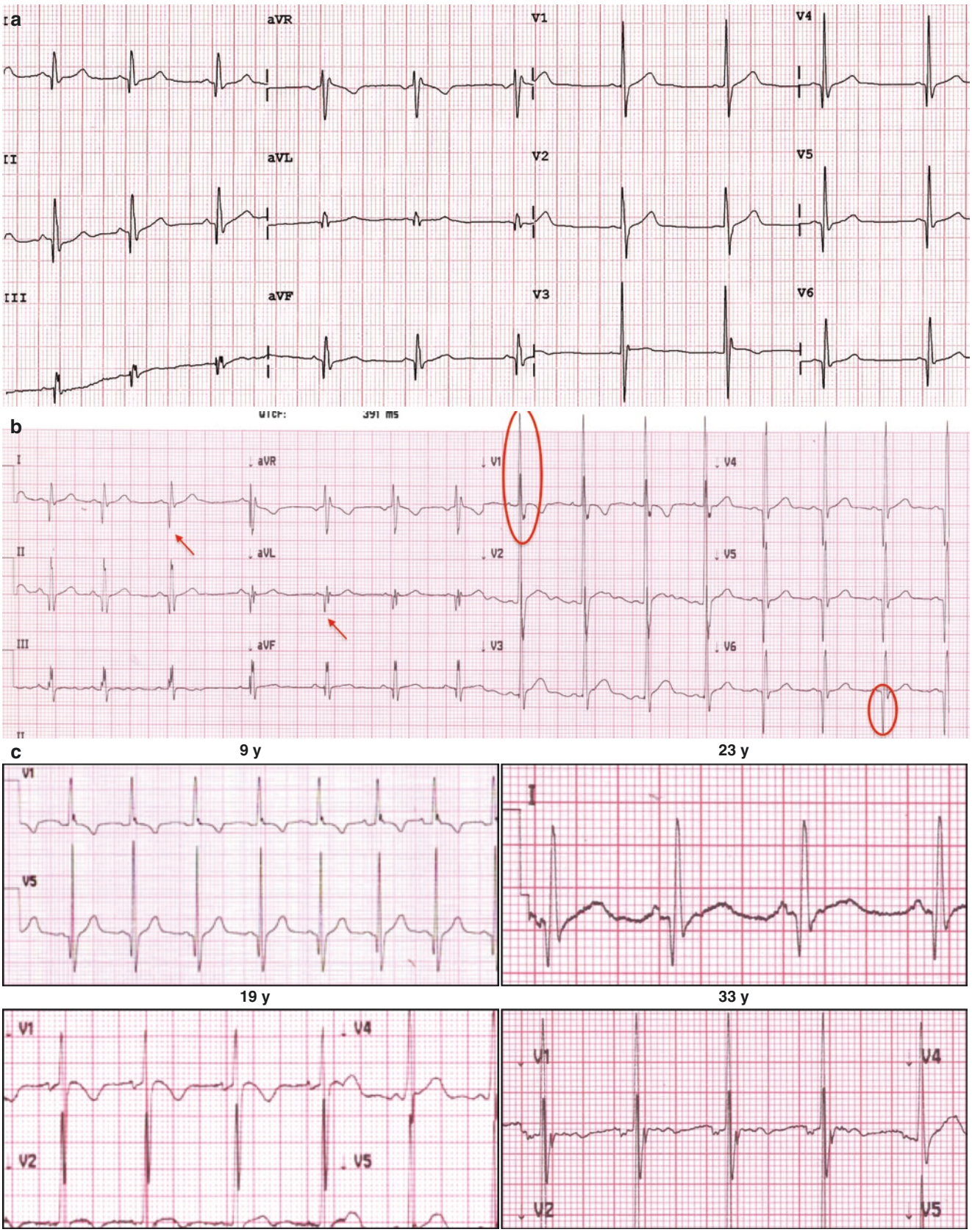

Fig. 13.26 Duchenne dystrophy. (**a**) Fourteen-year-old, note the peculiar traits like the tall R wave in the right precordial leads (sign of postero-basal necrosis), the short PR, and infero-lateral Q waves. (**b**) Eleven-year-old, note the large Q wave in V6. (**c**) DMD at different ages. (**d**) Eight-year-old boy with Becker dystrophy: the R/S ratio in V1 is >1

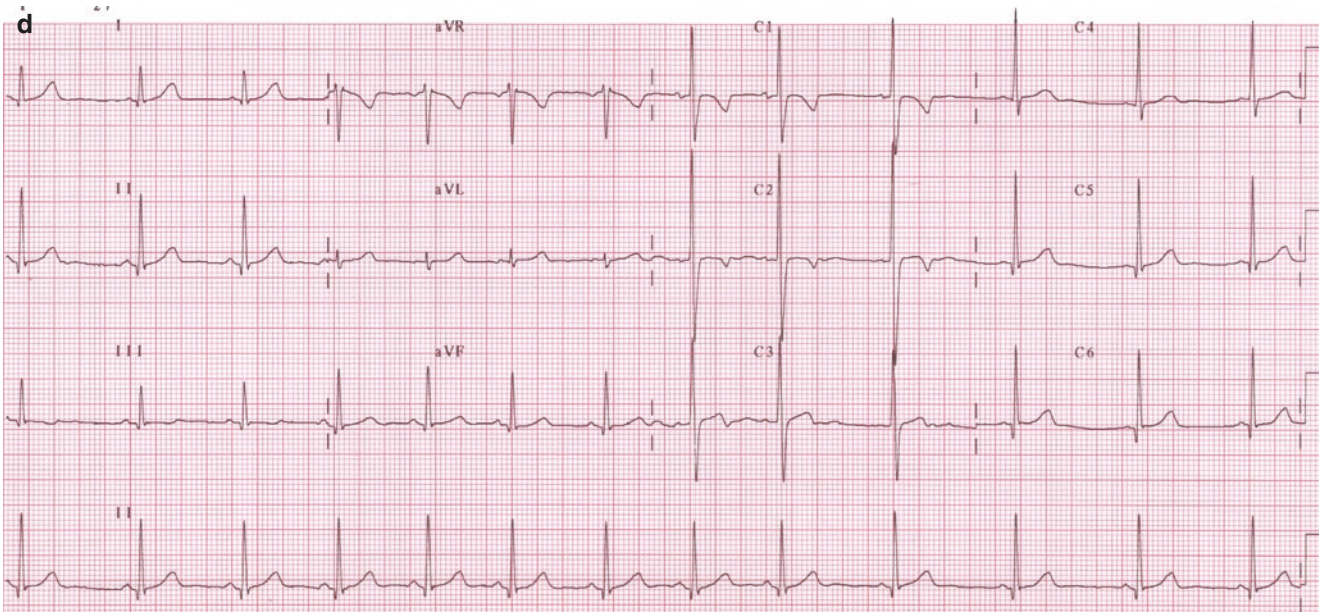

Fig. 13.26 (continued)

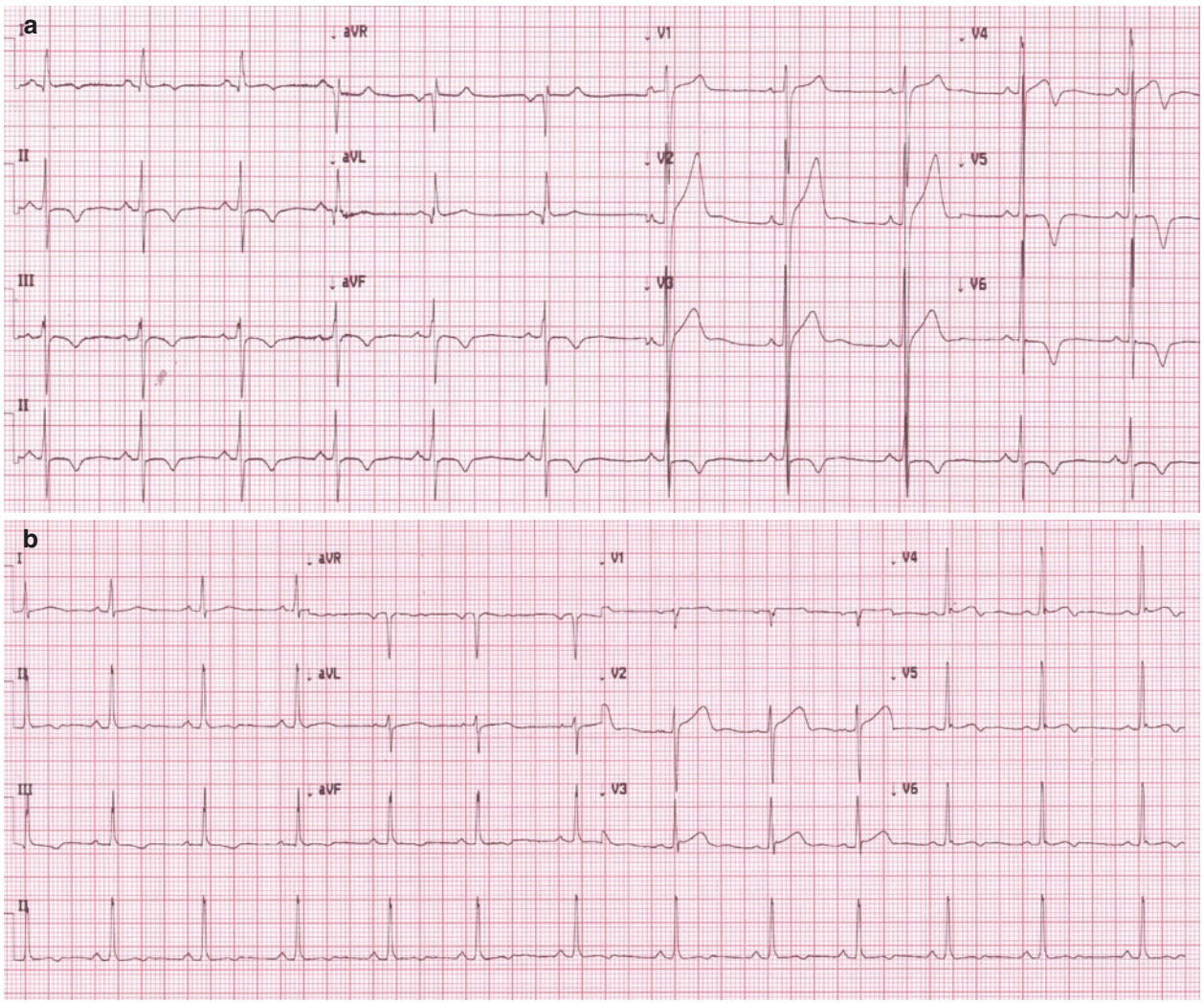

Fig. 13.27 Two cases of Friedreich ataxia and HCM. (**a**) Thirteen-year-old male with HCM, apical form. (**b**). Seventeen-year-old female with concentric form

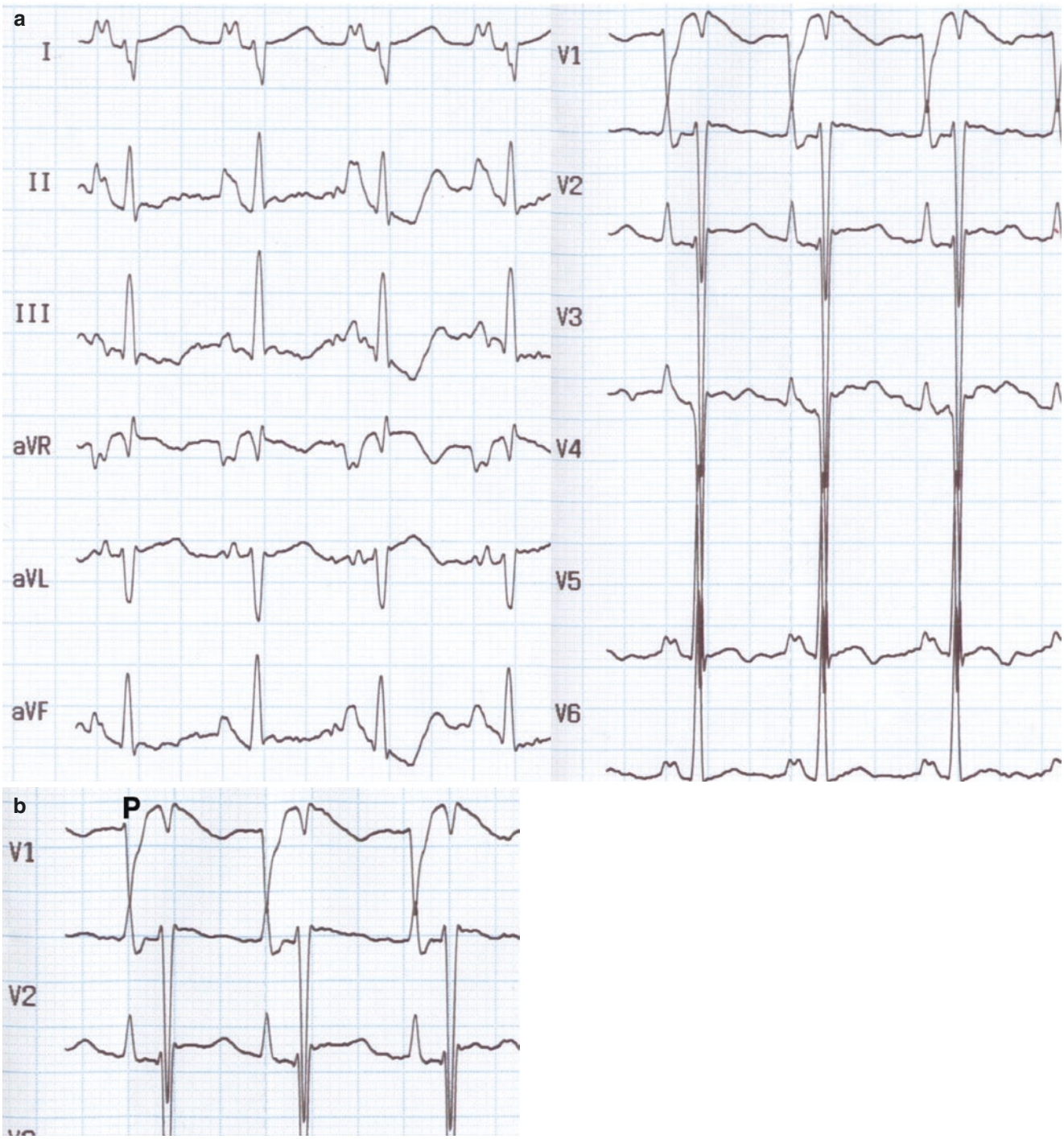

Fig. 13.28 (**a**) DMD with LVNC phenotype and restrictive physiology. (**b**) Note the gigantic P wave

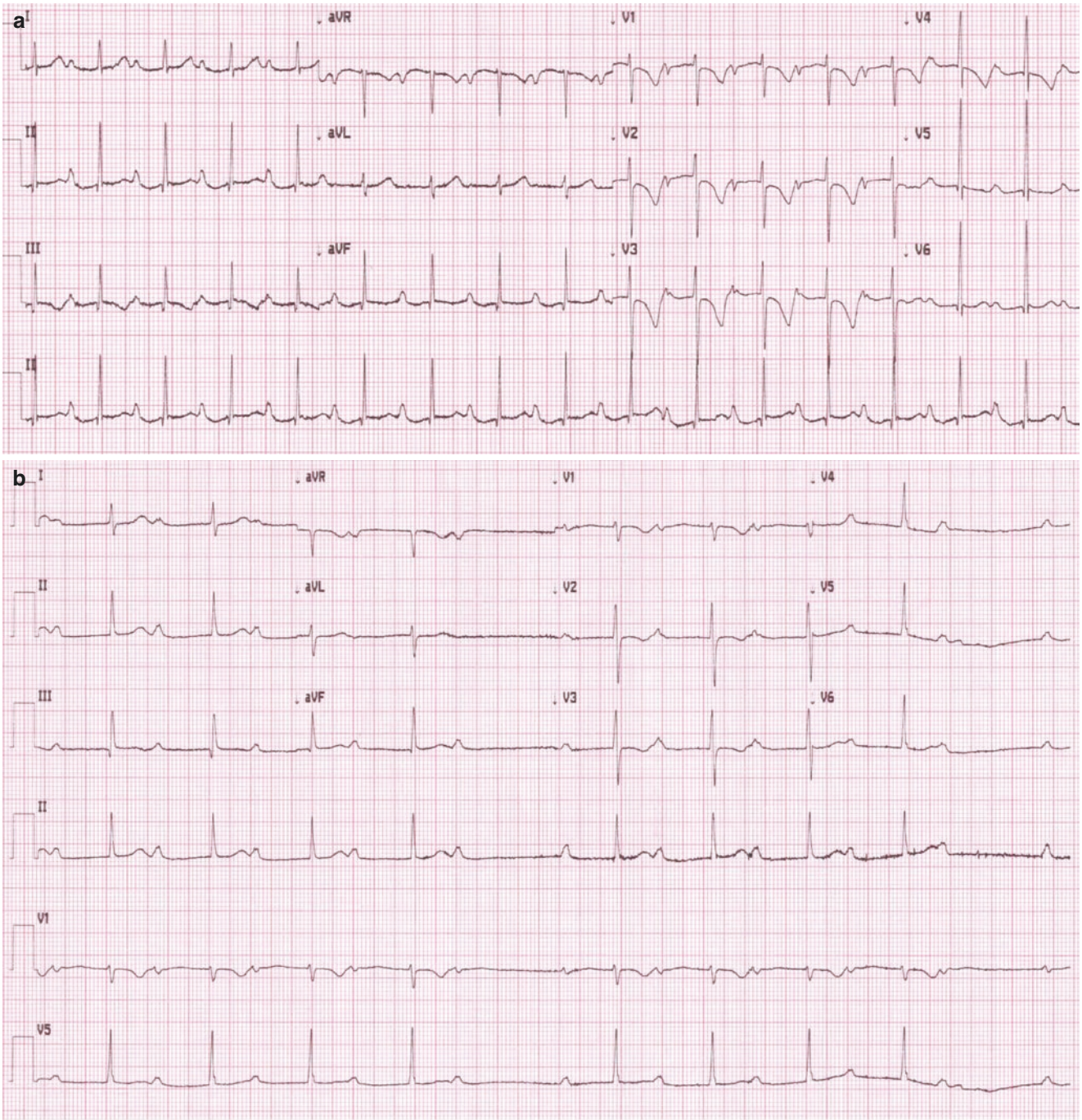

Fig. 13.29 (**a**) Two-year-old male. (**b**) Eight-year-old female with rheumatic fever. Carditis is actually a form of myocarditis with special predilection for the conduction system. In these cases, there is respectively a first (**a**)- and a second (**b**)- degree AV block

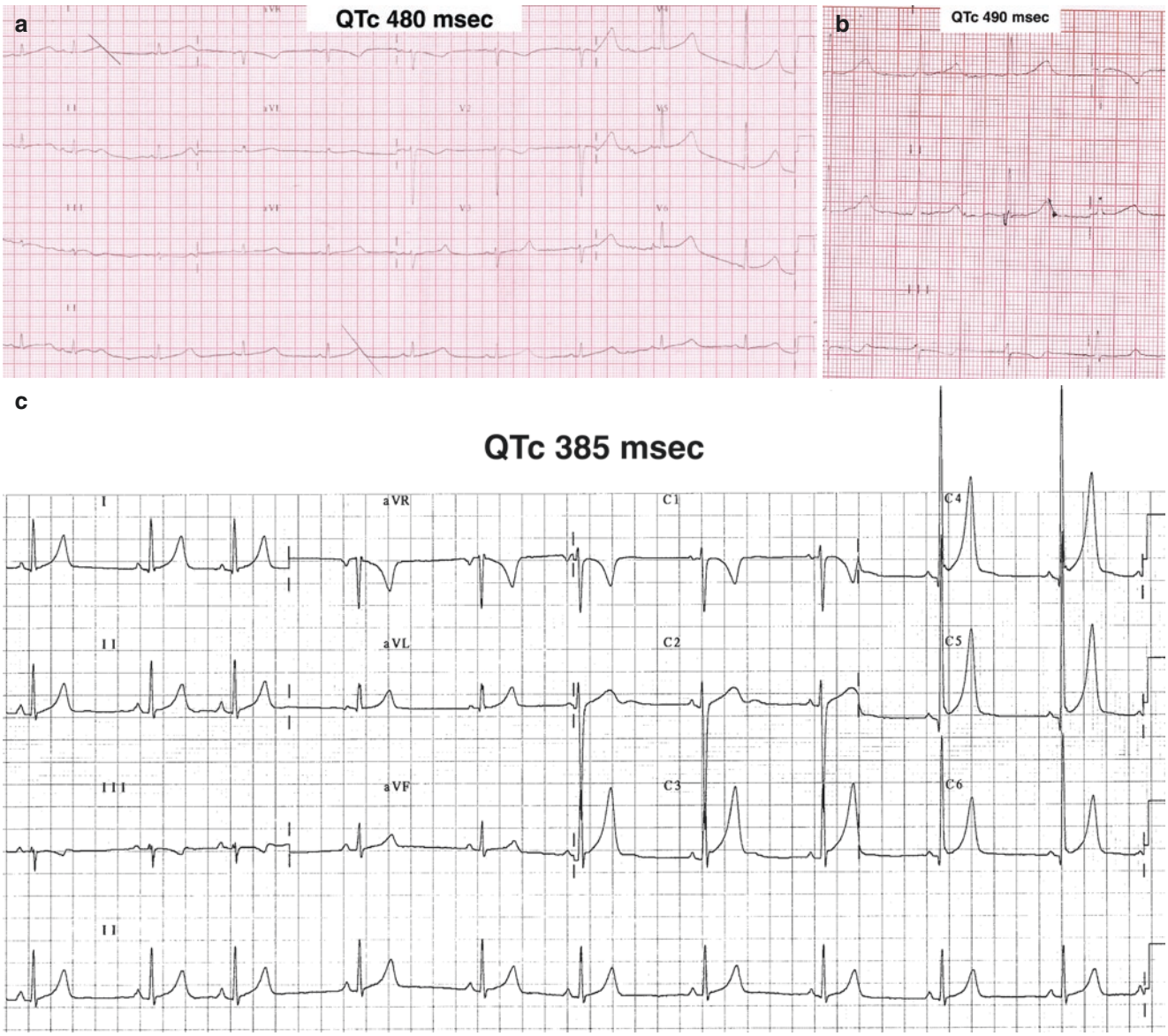

Fig. 13.30 (**a**) Eleven-year-old male with rheumatic fever. The patient manifested QT prolongation in the absence of drug treatment affecting the repolarization. (**b**) Close-up. Genetic testing for LQTS was negative. Once the acute phase of RF resolved, the QT returned to normal (**c**). So RF can impact on heart electricity in many ways, prolonging PR and QT interval. However this last feature is very rare

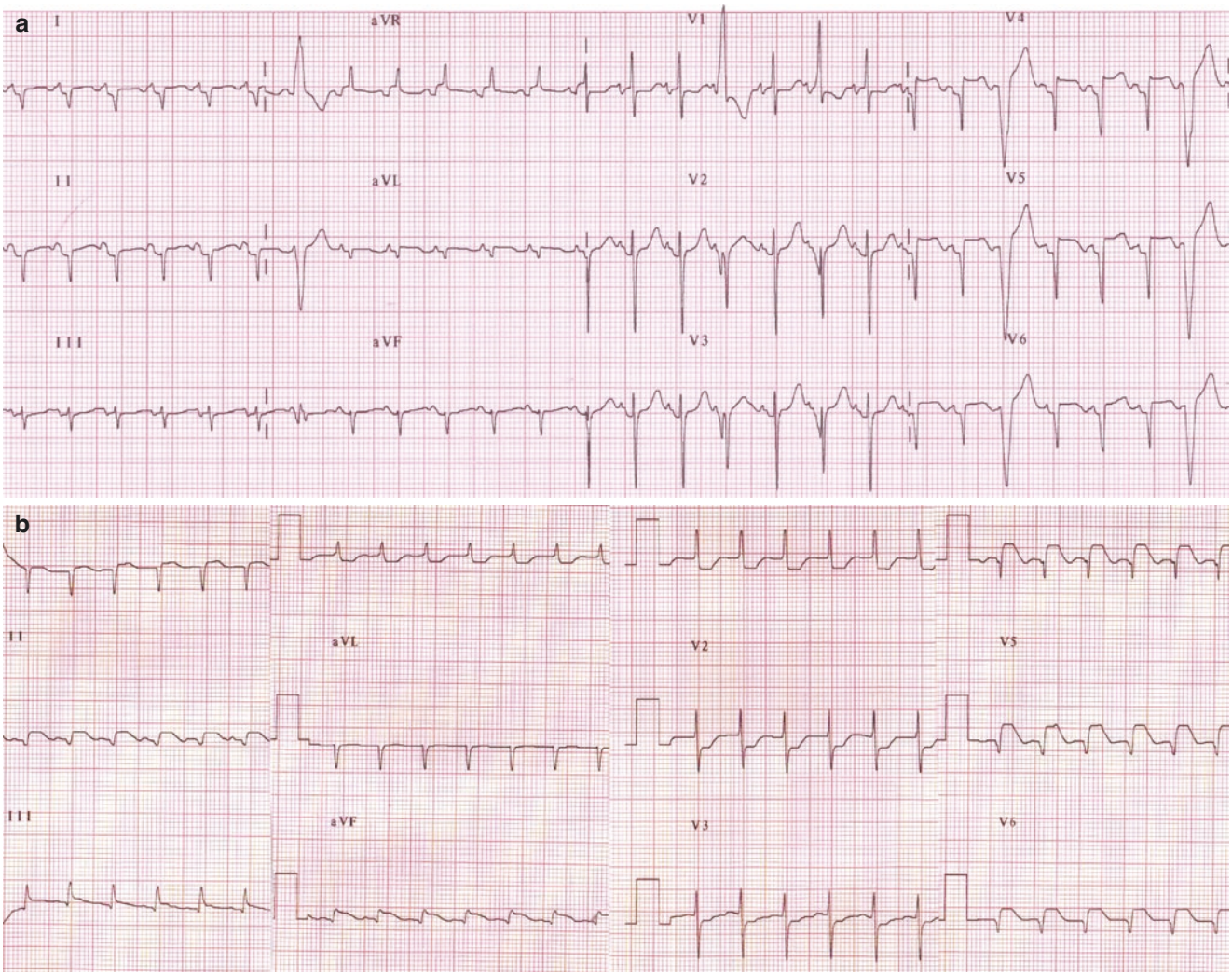

Fig. 13.31 Neonates with myocarditis. (**a**) Diffuse low voltages, truncated R wave in the precordial leads, and ST elevation. Note the PVC's (**b**) as in the previous figure, there are features of loss of muscle and subepicardial damage

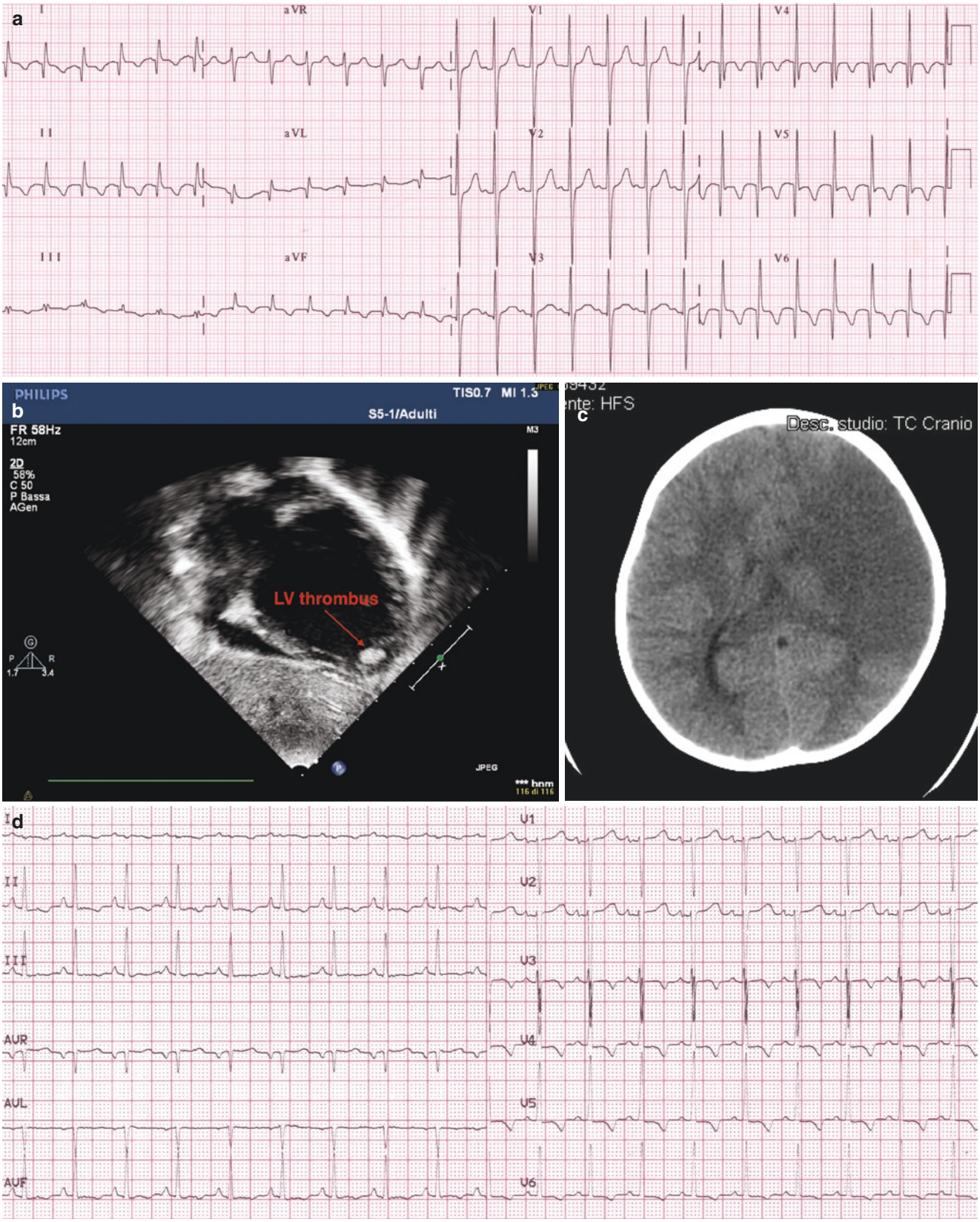

Fig. 13.32 Myocarditis in children. (**a**) Two-year-old female admitted for breathing difficulties. Note the sinus tachycardia and the ST–T changes (**b**) The US showed diffuse hypokinesia with an LVEF of 35%, complicated by an apical thrombus which caused a stroke (**c**). (**d**) Seven-year-old female admitted for fatigue during influenza. On the echocardiogram the LVEF was 40; note the ST changes

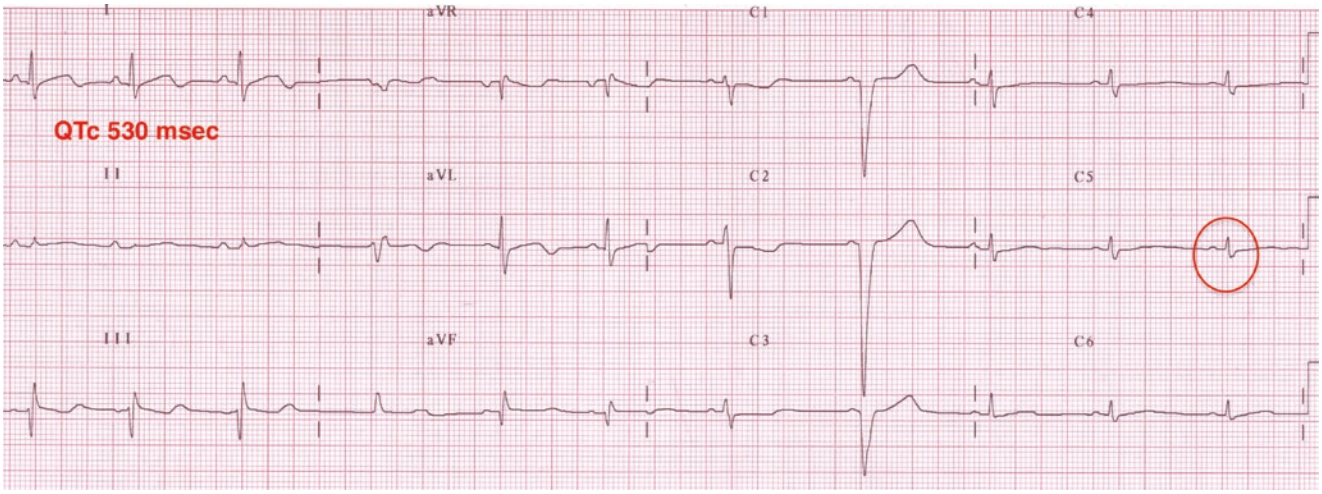

Fig. 13.33 Thirteen-year-old male with myocarditis diagnosed after a syncopal episode. Note the low voltages, the long QTc, and the PVC. Likely a ventricular arrhythmia was the cause of syncope. Unlike neonates which are electrically stable during myocarditis, after 10 years of age myocarditis may have a strong arrhythmic connotation

Suggested Reading

Moak JP, Kaski JP. Hypertrophic cardiomyopathy in children. Heart. 2012;98:1044–54.

Jefferies JL, Towbin JA. Dilated cardiomyopathy. Lancet. 2010;375:752–62.

Rapezzi C, Arbustini E, Caforio AL, Charron P, Gimeno-Blanes J, Heliö T, Linhart A, Mogensen J, Pinto Y, Ristic A, Seggewiss H, Sinagra G, Tavazzi L, Elliott PM. Diagnostic work-up in cardiomyopathies: bridging the gap between clinical phenotypes and final diagnosis. A position statement from the ESC working group on myocardial and pericardial diseases. Eur Heart J. 2013;34:1448–58.

Weintraub RG, Semsarian C. Paediatric cardiomyopathy: getting to the heart of the matter. Lancet. 2013;382:1866–7.

Biagini E, Pazzi C, Olivotto I, et al. Usefulness of electrocardiographic patterns at presentation to predict long-term risk of cardiac death in patients with hypertrophic cardiomyopathy. Am J Cardiol. 2016;118(3):432–9.

Cardiac Tumors and Pericardial Disease

14

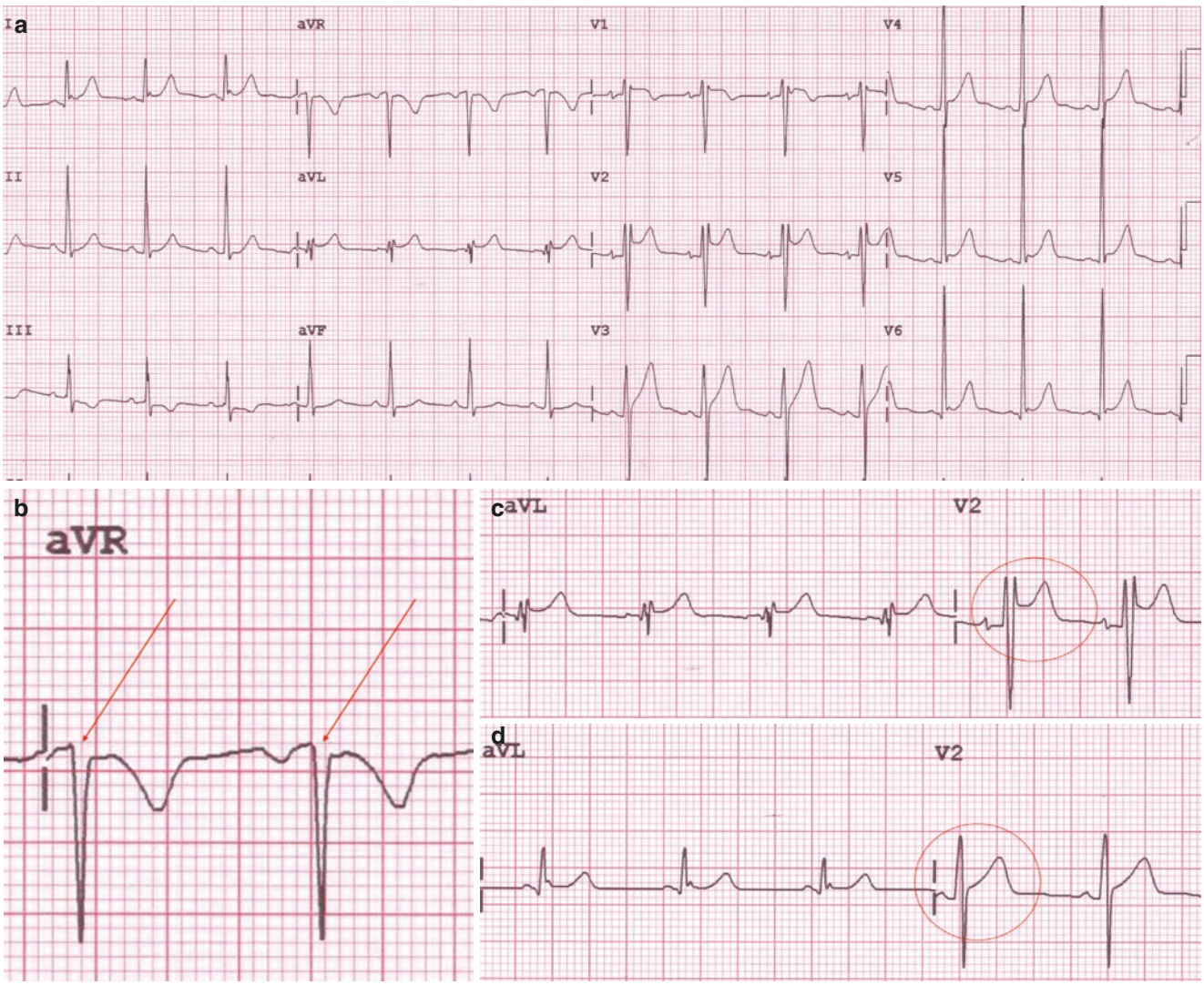

Fig. 14.1 (**a**) Twelve-year-old female with chest pain suggestive of pericarditis. (**b**, **c**) There is widespread concave ST elevation and PR depression. Reciprocal ST depression and PR elevation in lead aVR (with or without V1). As an ancillary but aspecific sign, there can be sinus tachycardia due to pain or pericardial effusion. (**d**) With healing, the ECG returned to normal

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G. Bronzetti, *Atlas of Pediatric and Youth ECG*, https://doi.org/10.1007/978-3-319-57102-7_14

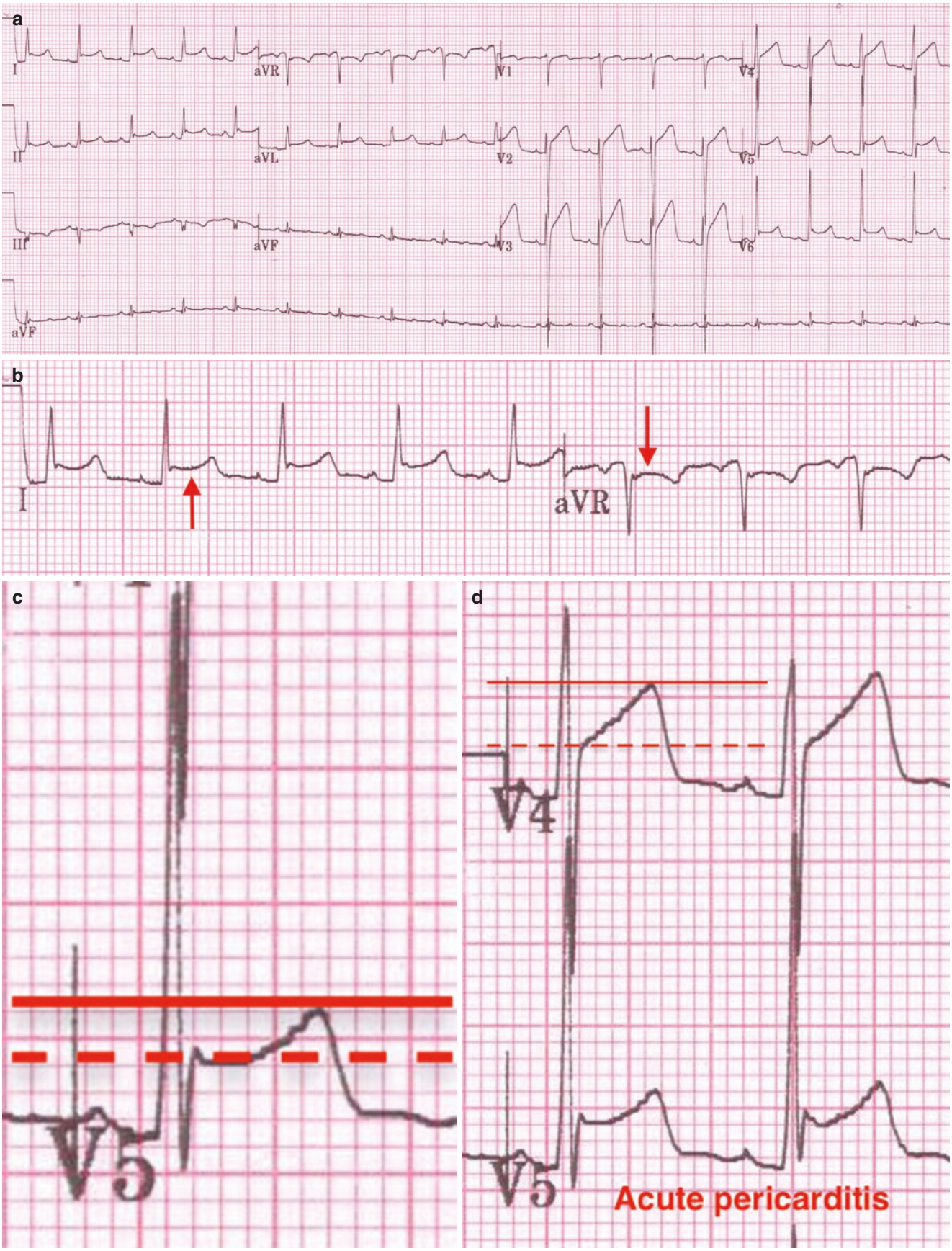


Fig. 14.2 (**a**) Nine-year-old male with typical manifestation of pericarditis. (**b**) Widespread concave ST elevation and PR elevation in aVR. (**c**) The height of the ST segment elevation in the J point is compared to the amplitude of the T wave in V5: a ratio of >0.25 suggests pericarditis; a ratio of <0.25 suggests BER. (**d**) In some leads as in V4, the pattern looks like BER but the ratio is >0.25

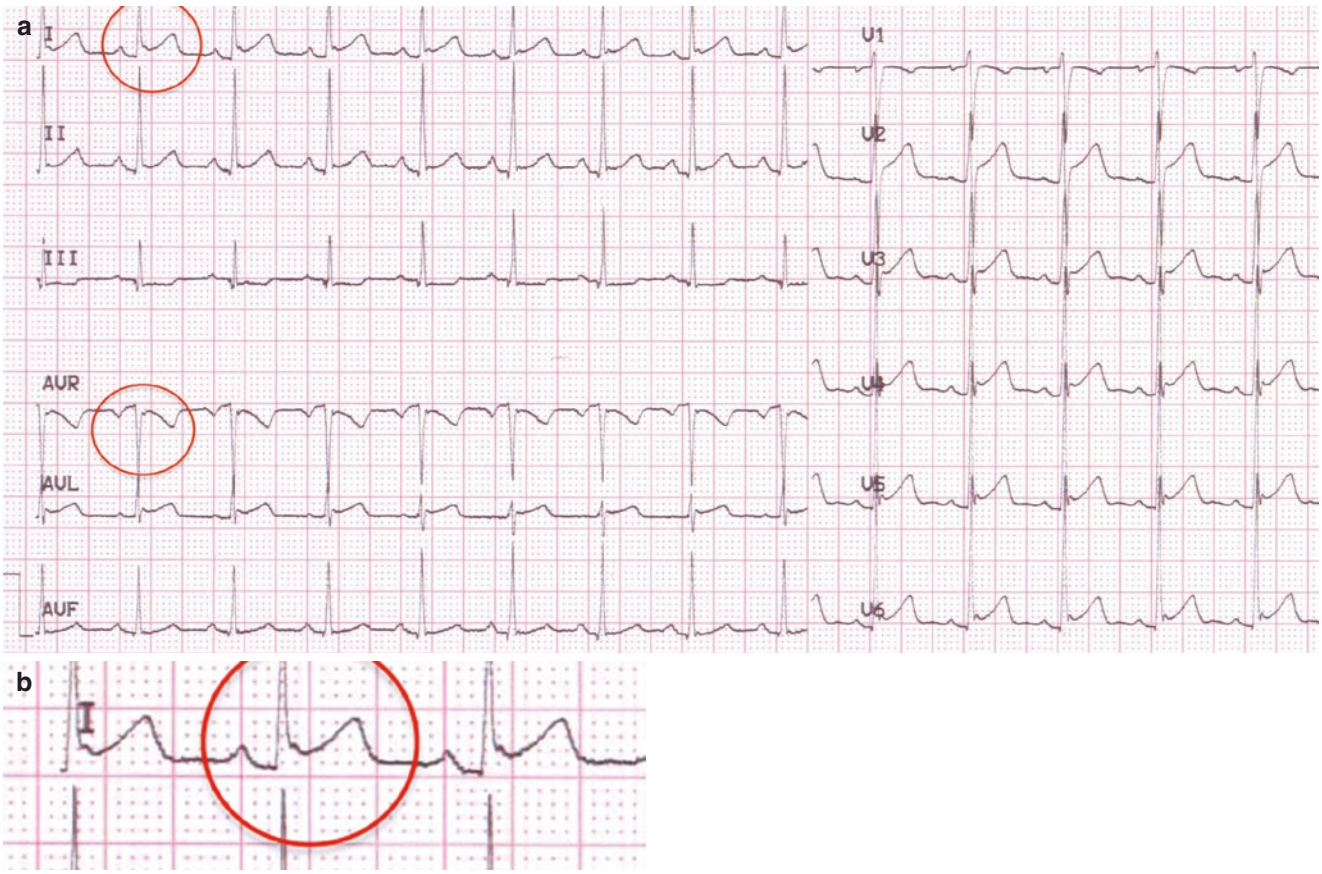


Fig. 14.3 (**a**) Ten-year-old male with acute pericarditis and mild pericardial effusion. ESR and CRP are elevated. Only leads I and aVR are suggestive, but not pathognomonic. (**b**) Close-up. The diagnosis of pericarditis remains a clinical and laboratory diagnosis

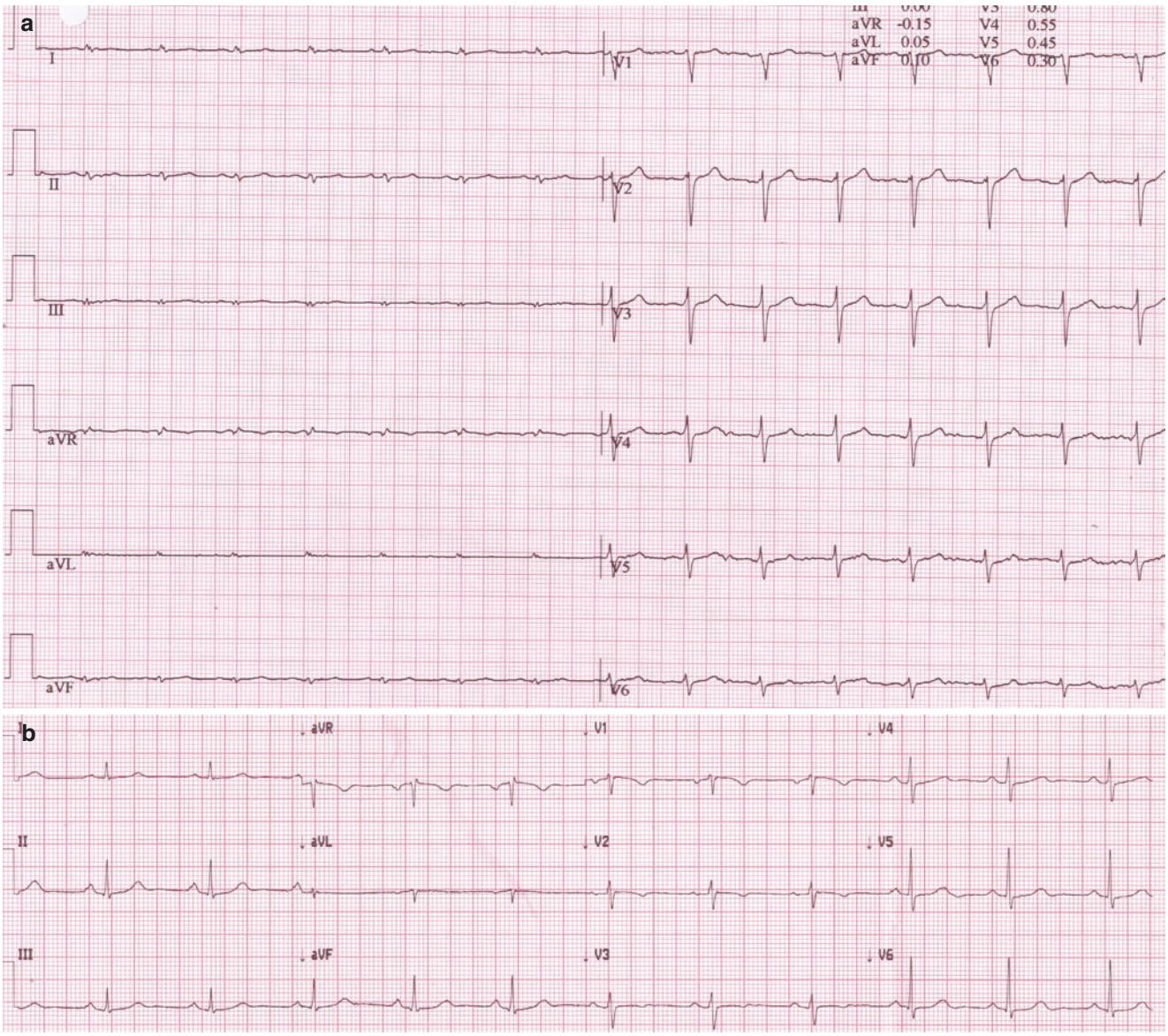
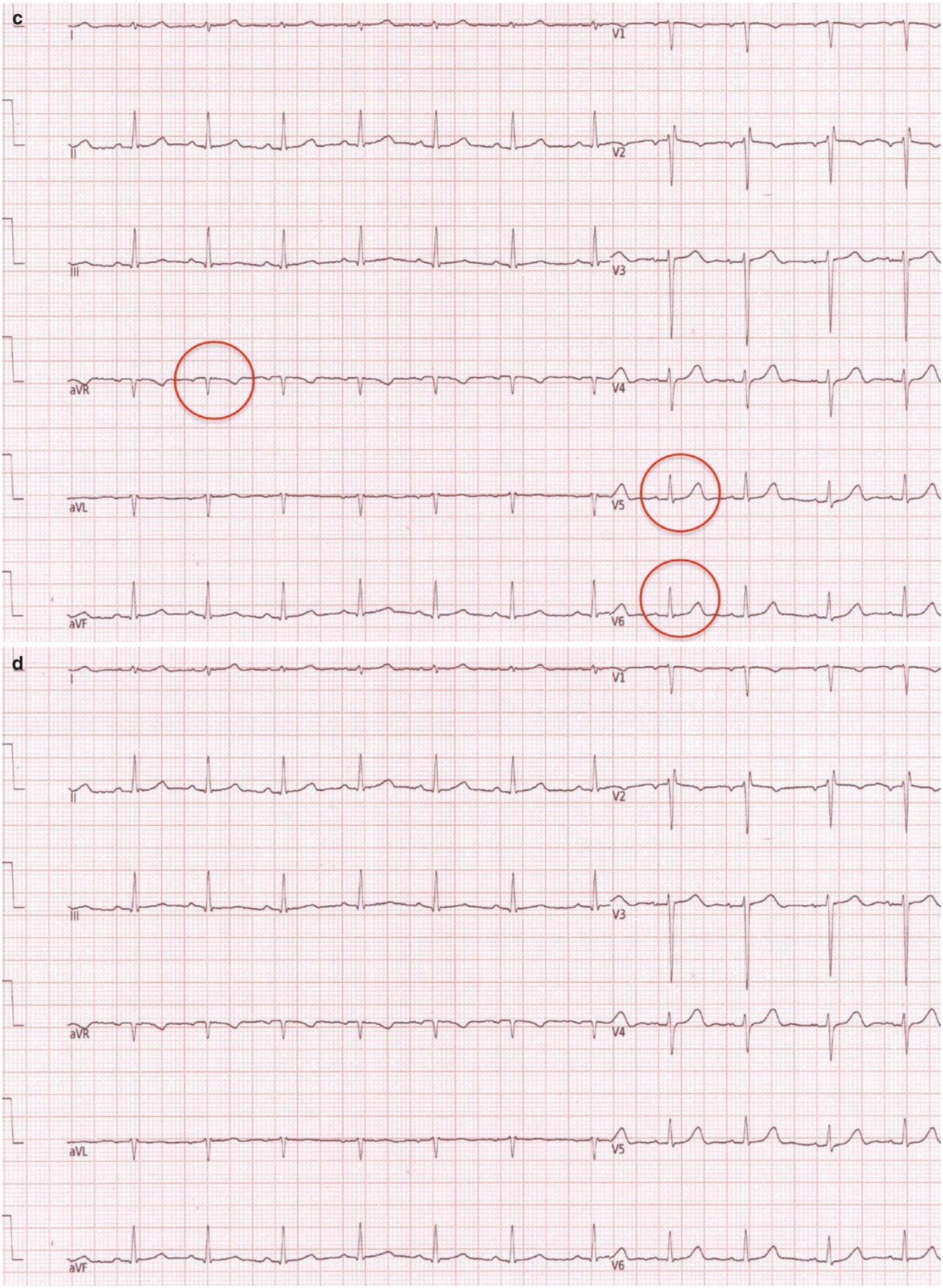


Fig. 14.4 Low voltages in pericardial effusion. (**a**) Fourteen-year-old male with severe pericardial effusion. (**b**) Twenty-year-old woman with incidental finding of moderate to severe pericardial effusion. (**c**) Fifteen-year-old girl with severe pericardial and pleural effusion. (**d**) Eight-year-old male with pericardial effusion



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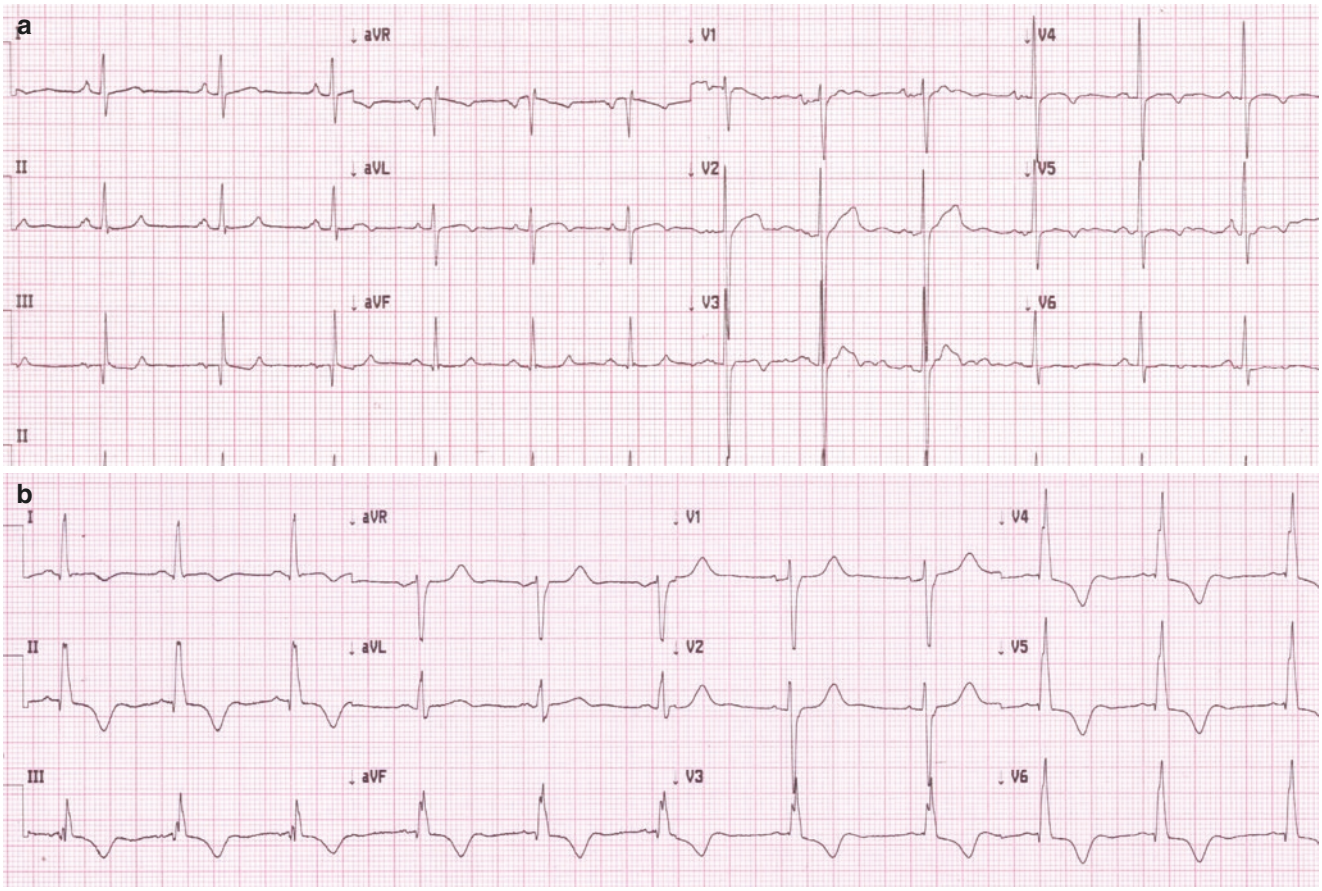


Fig. 14.5 (**a**) Ten-year-old male with incidental finding of ST changes in the left precordial leads. Echocardiogram and MRI confirmed a small benign tumor (fibroma) in the apical aspect of the left ventricle. (**b**) Sixteen-year-old female with large fibroma in the apex of the left ventricle (3 × 3 cm). There are deep negative T waves as in apical HCM. When a large tumor is located in the right or left ventricular outflow tract signs of RVH or LVH can be found

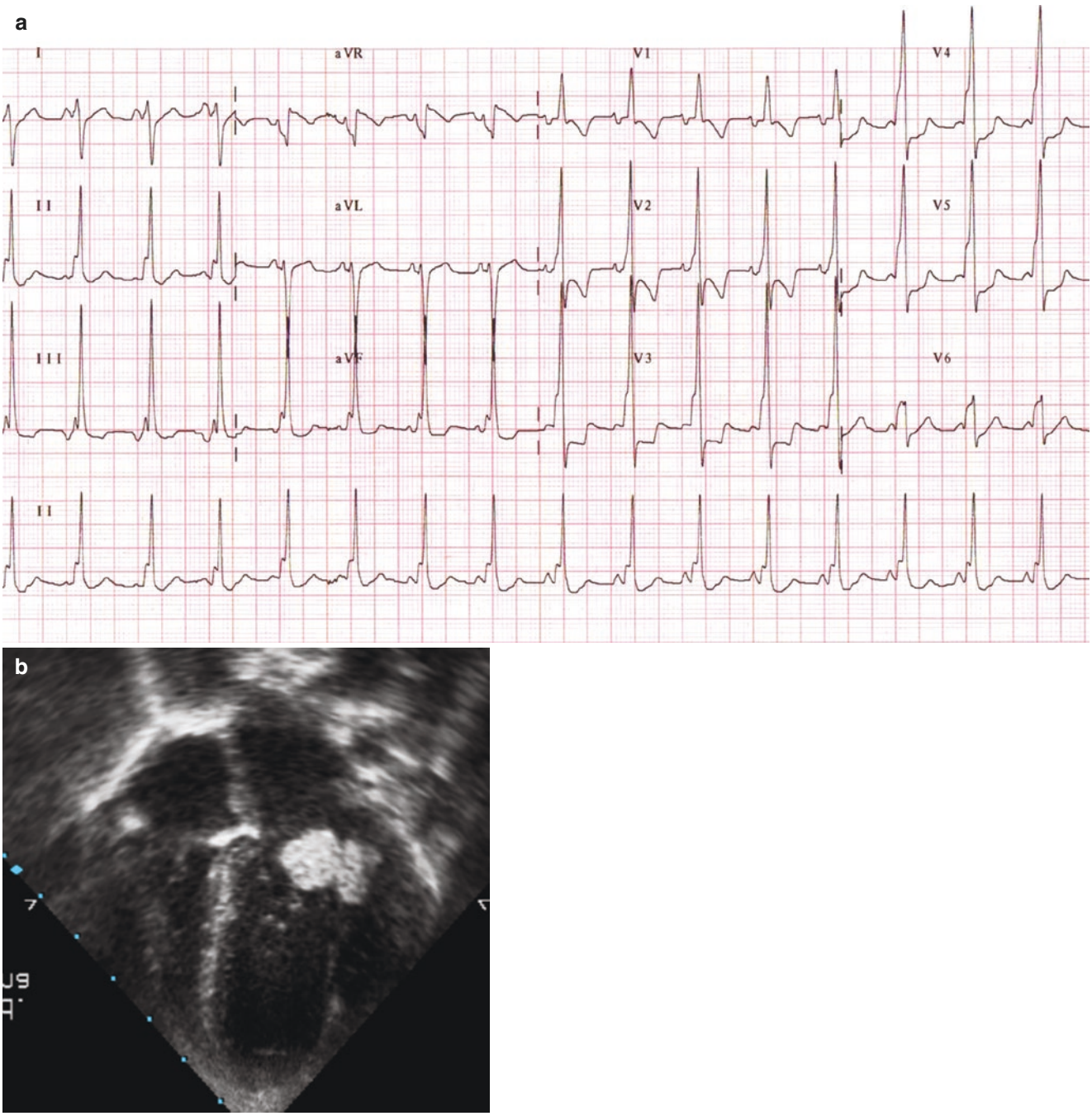


Fig. 14.6 (**a**) Neonate with tuberous sclerosis (TS) and WPW. Baseline ECG with pre-excitation due to a left accessory pathway. (**b**) Echocardiogram: the apical four-chamber view shows two large rhabdomyomas involving the mitral valve and the left AV ring. As it is easy to infer, this anatomy explains the connection between the atrium and the ventricle which acts as a large accessory pathway. Rhabdomyomas are the most common primary cardiac tumors in childhood and are often associated with TS

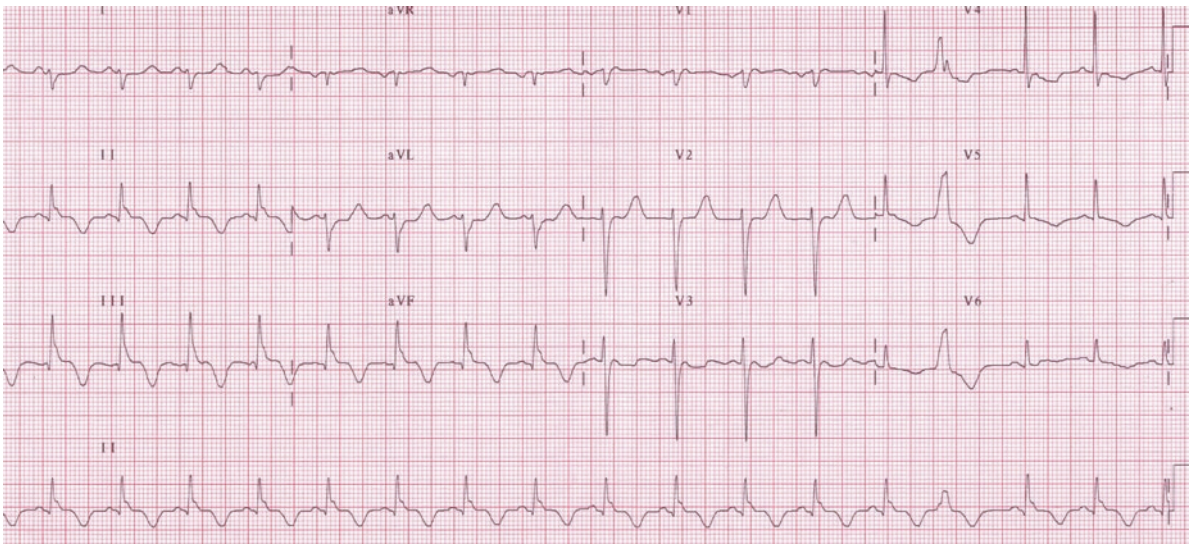
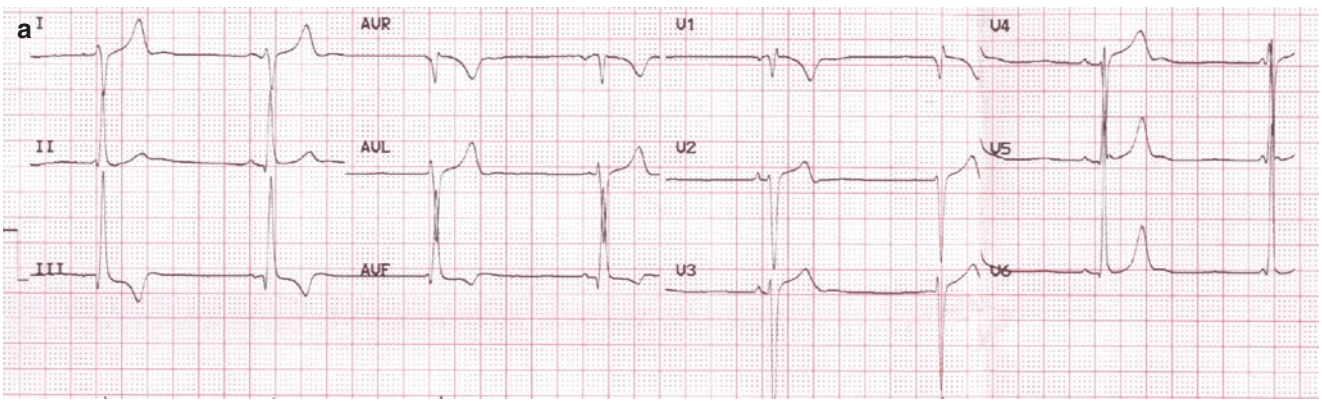


Fig. 14.7 Thirteen-year-old male with acute lymphoblastic leukemia and cardiac infiltration. The patient was admitted for cardiac tamponade. The echocardiogram showed a severe impairment of the ventricular function. Severe and diffuse ST-T changes can be noted



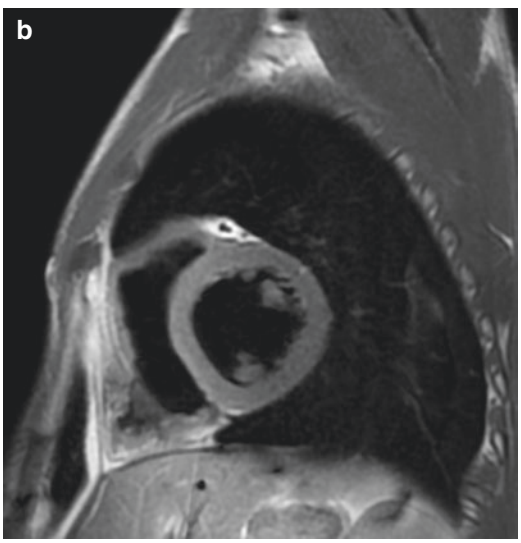


Fig. 14.8 (**a**) Eighteen-year-old male. This is a young athlete, as shown by the marked sinus bradycardia alternating with junctional rhythm and early repolarization. There is a right axis deviation (QRS axis about 110°) in the frontal plane and a clockwise rotation in the horizontal plane (poor growth precordial R waves); T waves in aVF and lead III are negative but non-specific. On the echocardiogram, it was impossible to obtain "normal" images from standard windows and a bipartite aspect of the right ventricle was noted. The first diagnosis was diverticulum of the right ventricle, confirmed by tomography. On MRI, instead, the picture was explained by a partial agenesis of the pericardium especially of the left ventricle. (**b**) MRI, sagittal short axis projection. The absence of the pericardium allows the lung to interpose itself above and below the heart. The left "naked" ventricle appears to float in the lung. This diagnosis is compatible with the ECG (right axis deviation, poor growth of precordial R, T wave abnormalities) and with the unusual rotation of the heart on echo

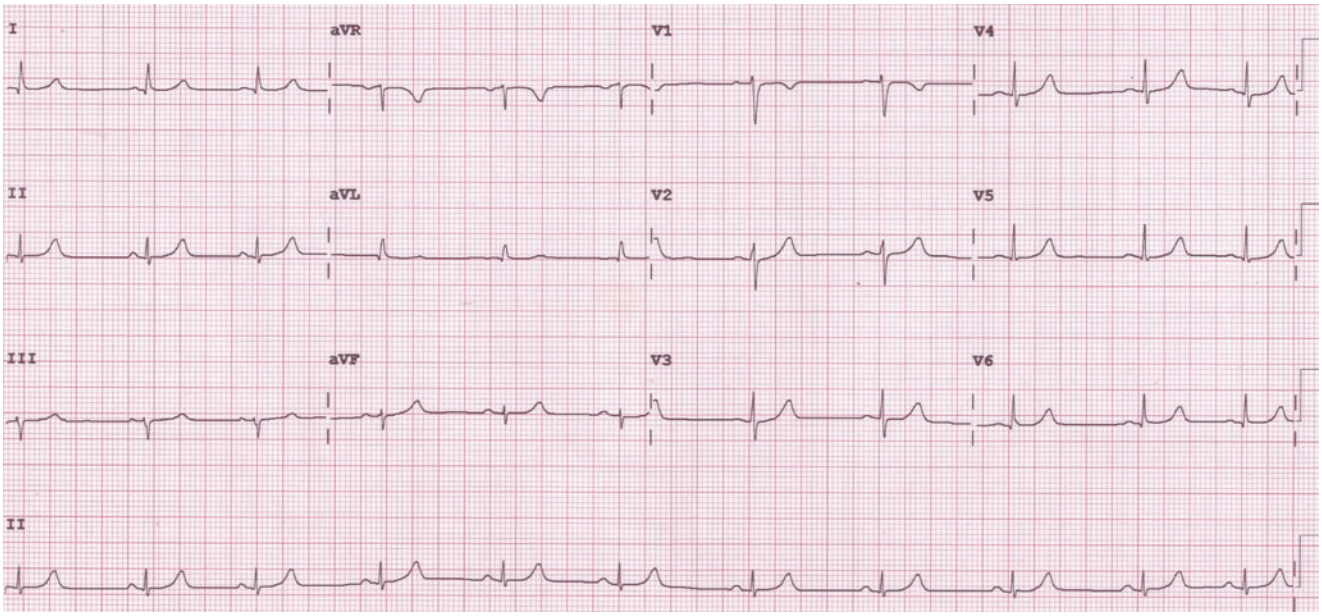


Fig. 14.9 Eighteen-year-old female with neurofibromatosis type 1 (NF1). Several cardiac abnormalities are described in NF1, such as ASD, mitral abnormalities, aortic coarctation, left ventricle hypertrophy. In this case, the echocardiogram was normal. On the ECG diffuse low voltages can be noted. According to our observation in young adult patients with NF1 there is a tendency to low voltages, but we do not have a plausible explanation for this

Suggested Reading

Careddu L, Oppido G, Petridis FD, Liberi R, Ragni L, Pacini D, Pace Napoleone C, Angeli E, Gargiulo G. Primary cardiac tumours in the paediatric population. Multimed Man Cardiothorac Surg. 2013;2013:mmt013.

Elias MD, Glatz AC, O'Connor MJ, Schachtner S, Ravishankar C, Mascio CE, Cohen MS. Prevalence and risk factors for pericardial effusions requiring readmission after pediatric cardiac surgery. Pediatr Cardiol. 2017;38(3):484–94.

Adler Y, Charron P, Imazio M, Badano L, Barón-Esquivias G, Bogaert J, Brucato A, Gueret P, Klingel K, Lionis C, Maisch B, Mayosi B, Pavie A, Ristić AD, Sabaté Tenas M, Seferovic P, Swedberg K, Tomkowski W, Achenbach S, Agewall S, Al-Attar N, Angel Ferrer J, Arad M, Asteggiano R, Bueno H, Caforio AL, Carerj S, Ceconi C, Evangelista A, Flachskampf F, Giannakoulas G, Gielen S, Habib G, Kolh P, Lambrinou E, Lancellotti P, Lazaros G, Linhart A, Meurin P, Nieman K, Piepoli MF, Price S, Roos-Hesselink J, Roubille F, Ruschitzka F, Sagristà Sauleda J, Sousa-Uva M, Uwe Voigt J, Luis Zamorano J; European Society of Cardiology (ESC). 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: The Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC) Endorsed by: The European Association for Cardio-Thoracic Surgery (EACTS). Eur Heart J. 2015;36:2921–64.

Pulmonary Hypertension and Acquired Valvular Disease

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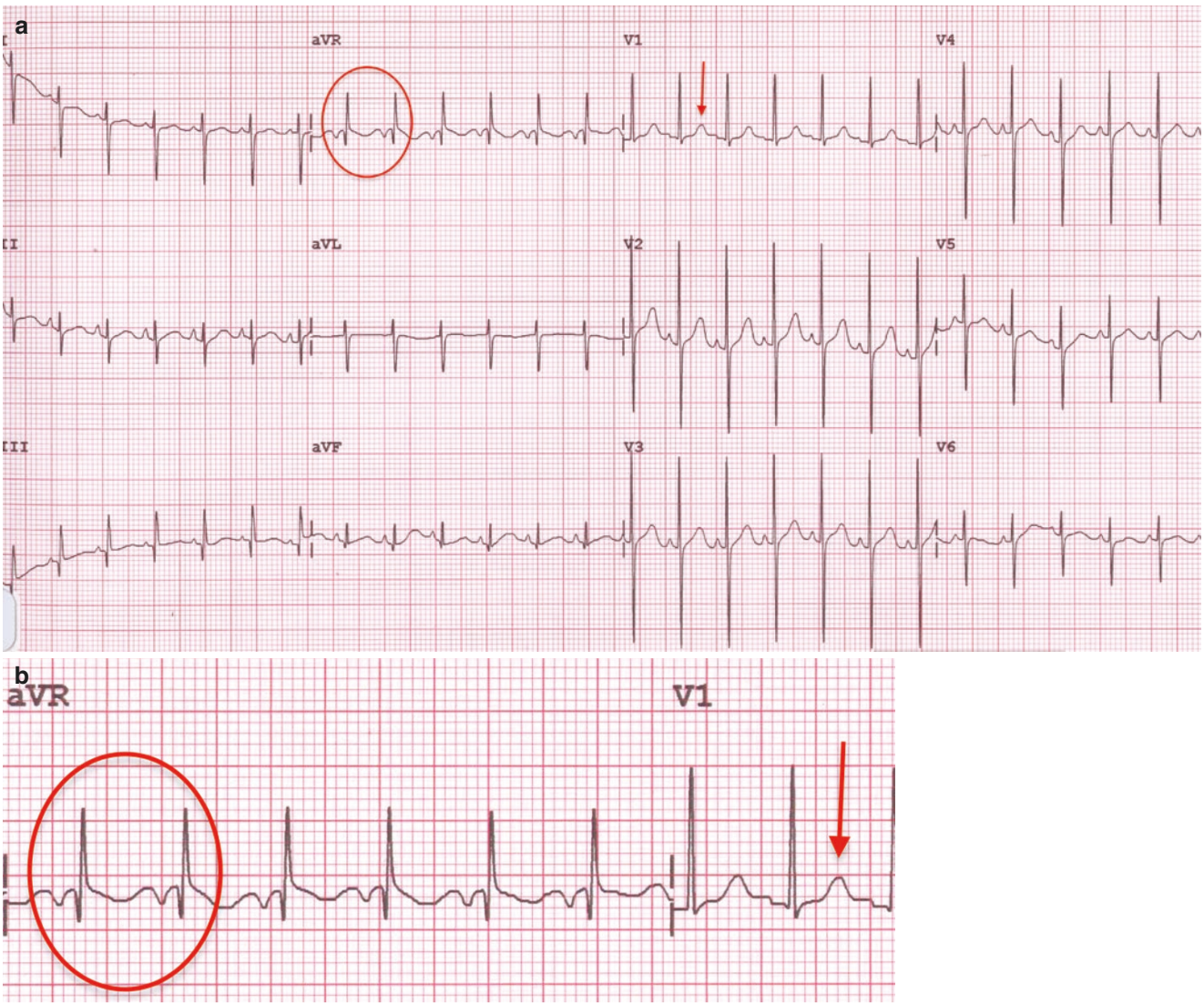

Fig. 15.1 (**a**) One-month-old baby boy with mild pulmonary hypertension. Note the right axis deviation (dominant R wave in aVR) and the positive T wave in V1. (**b**) Close-up

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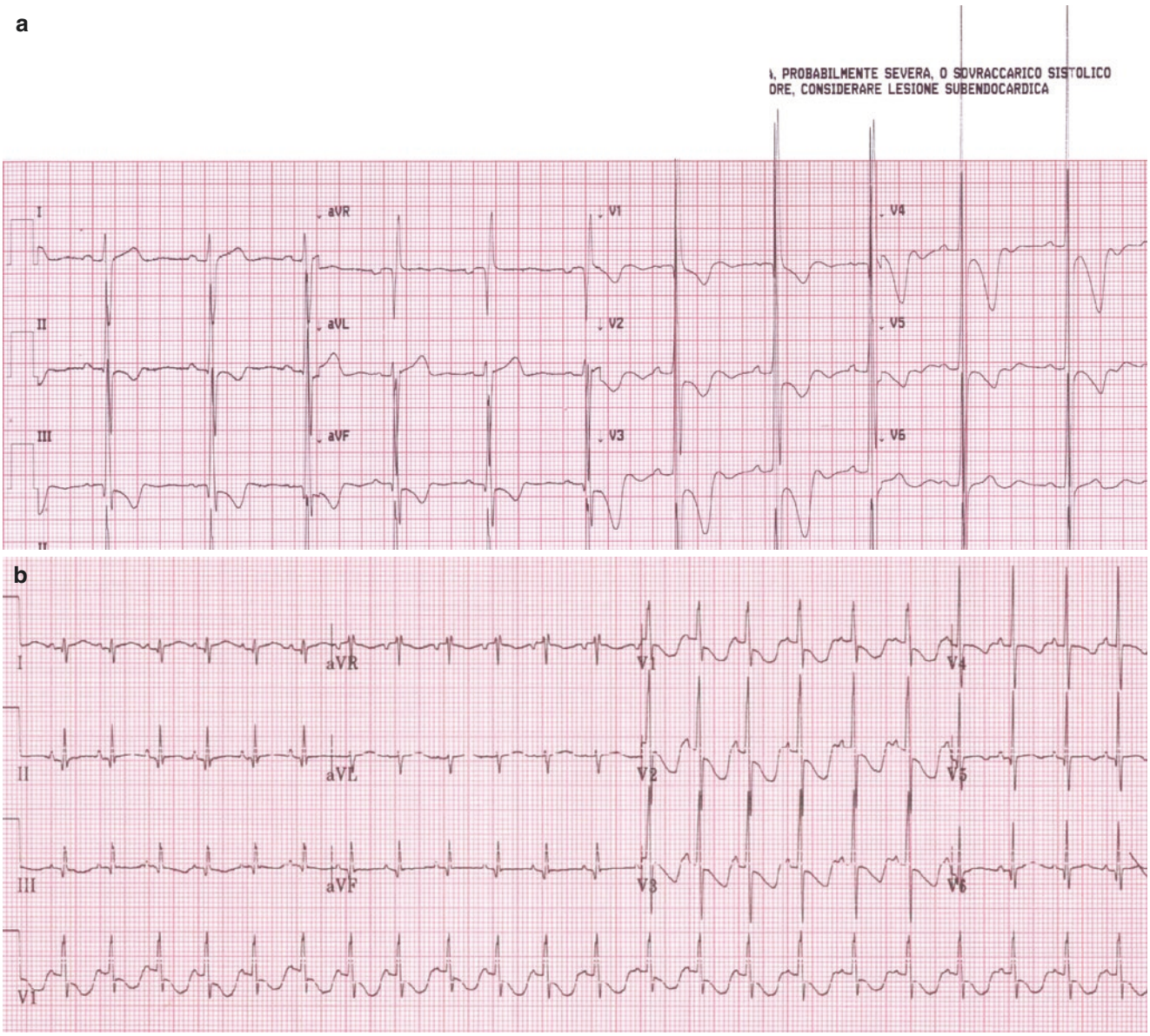


Fig. 15.2 (**a**) Ten-year-old male with severe primitive pulmonary hypertension (PPH). Tall R waves in the right precordial leads and reciprocal deep S waves in the left precordial leads can be noticed. According to the Chou classification, this is type A of right ventricular hypertrophy (RVH) due to pressure overload. (**b**) Ten-year-old male with a large ASD and mild pulmonary hypertension. Note the right intraventricular conduction delay and the ST changes in right precordial leads. According to Chou, this is type B of RVH, due to volume overload. RVH type C is described in chronic lung disease: in this case there are no classic signs of RVH and a pattern like rS in V1–V2 and RS in V5–V6 can be found, due to right ventricle pressure overload, the shift of the heart and to the emphysematous lung

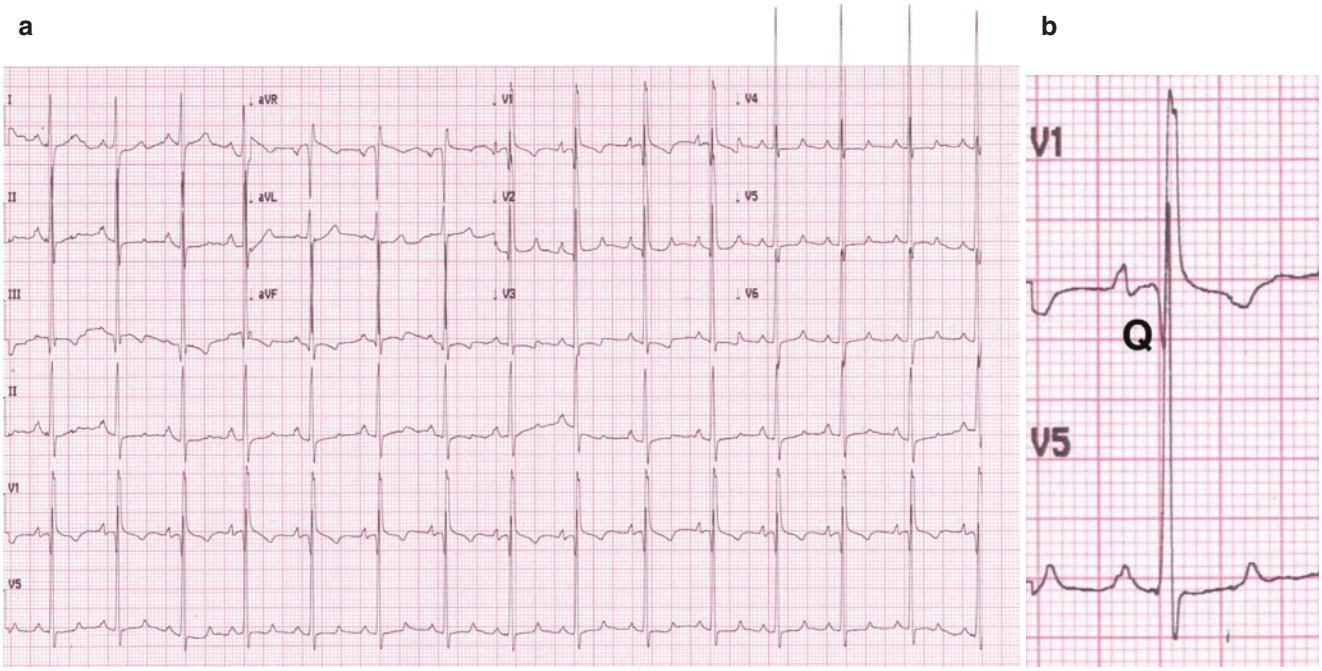


Fig. 15.3 (**a**) Thirteen-year-old female who underwent surgery for diaphragmatic hernia at neonatal age. She was left with severe pulmonary hypertension on pharmacological treatment. (**b**) The severe RVH is indicated by the Q wave in V1, as well as by the high right precordial R wave (RVH type A)

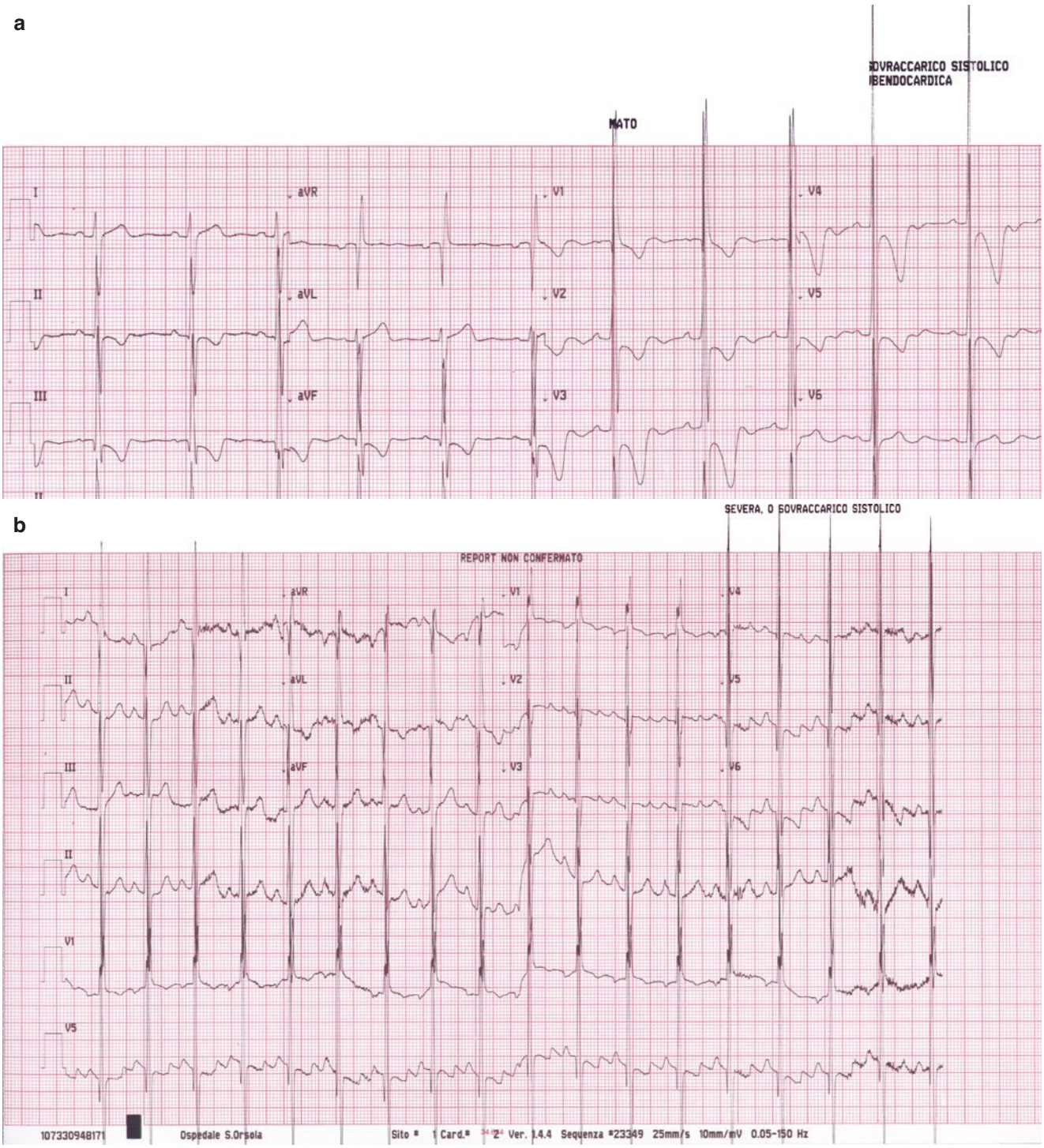
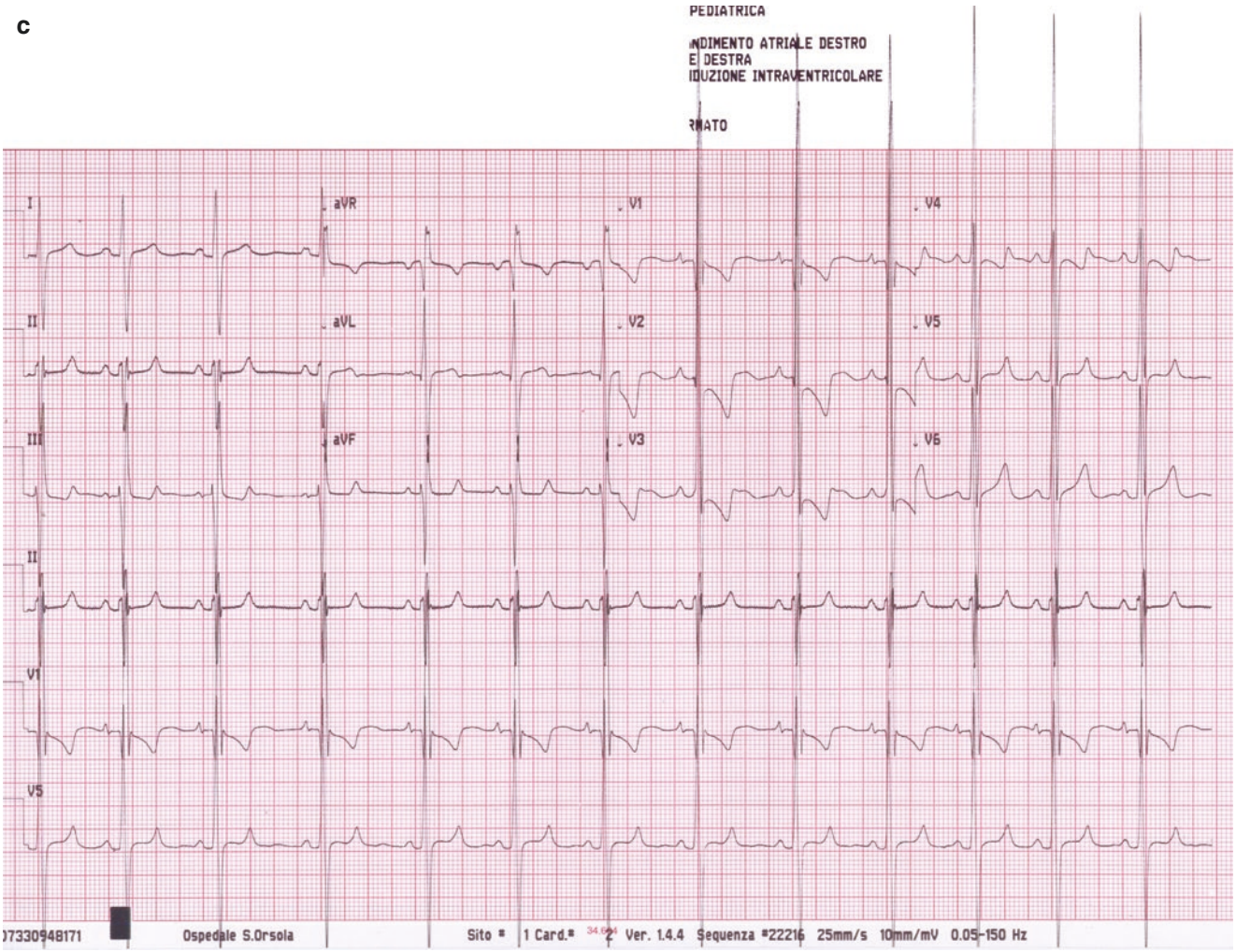


Fig. 15.4 Eisenmenger syndrome. (**a**) Ten-year-old female with unrepaired VSD (late diagnosis). A biventricular hypertrophy can be noted. (**b**) Seven-year-old male with unrepaired CAVC (late diagnosis). Note the left axis deviation. (**c**) Fifteen-year-old female with unrepaired CAVC (late diagnosis). Note the left axis deviation and the biventricular hypertrophy.



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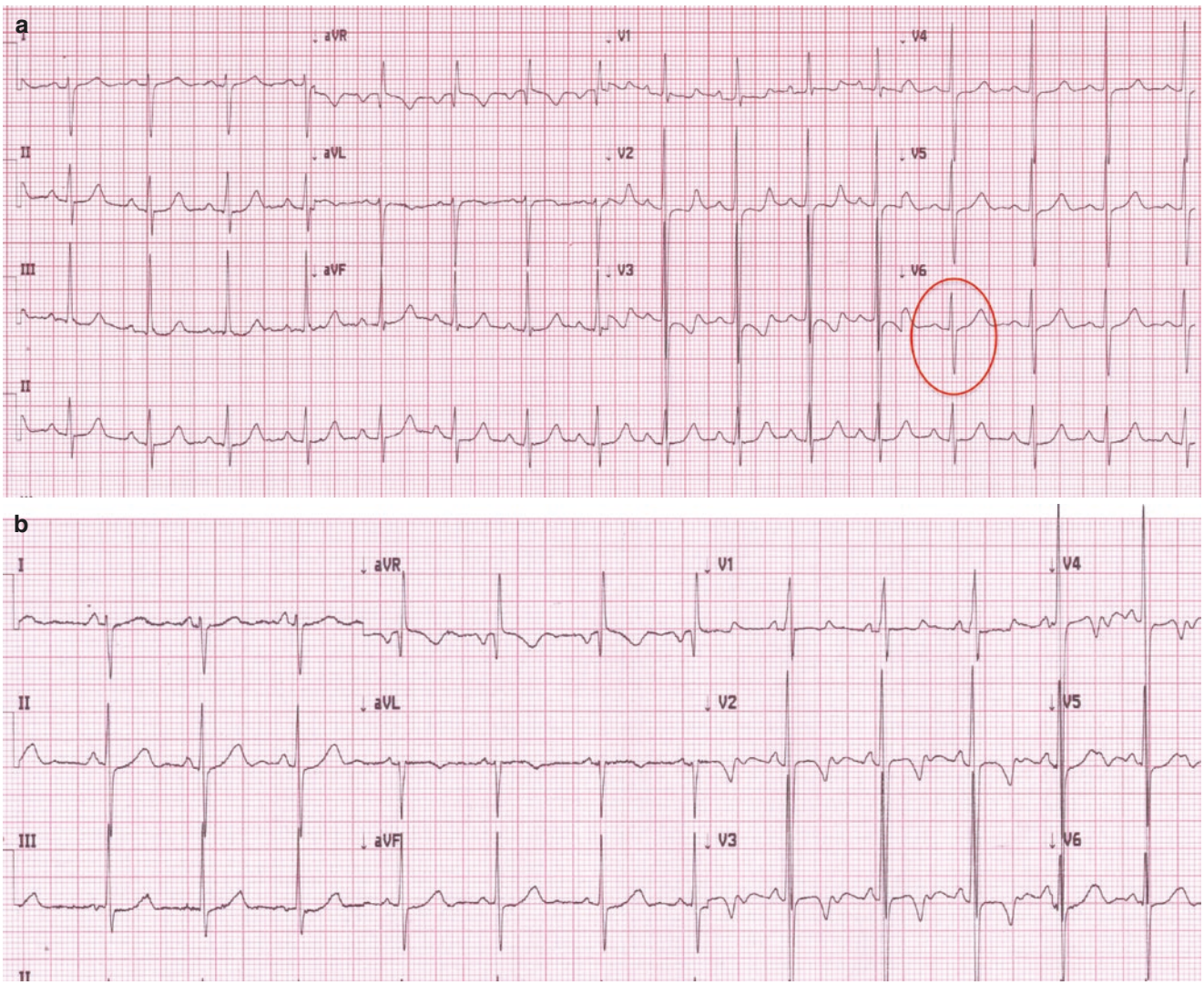

Fig. 15.5 (**a**) Nine-year-old male with moderate PPH, the right ventricular pressure is subsystemic (RVSP 70 mmHg). Note the positive T wave in V1 and the S in V6. (**b**) As in the previous case this is a 6-year-old female with moderate form of PPH on pharmacological treatment

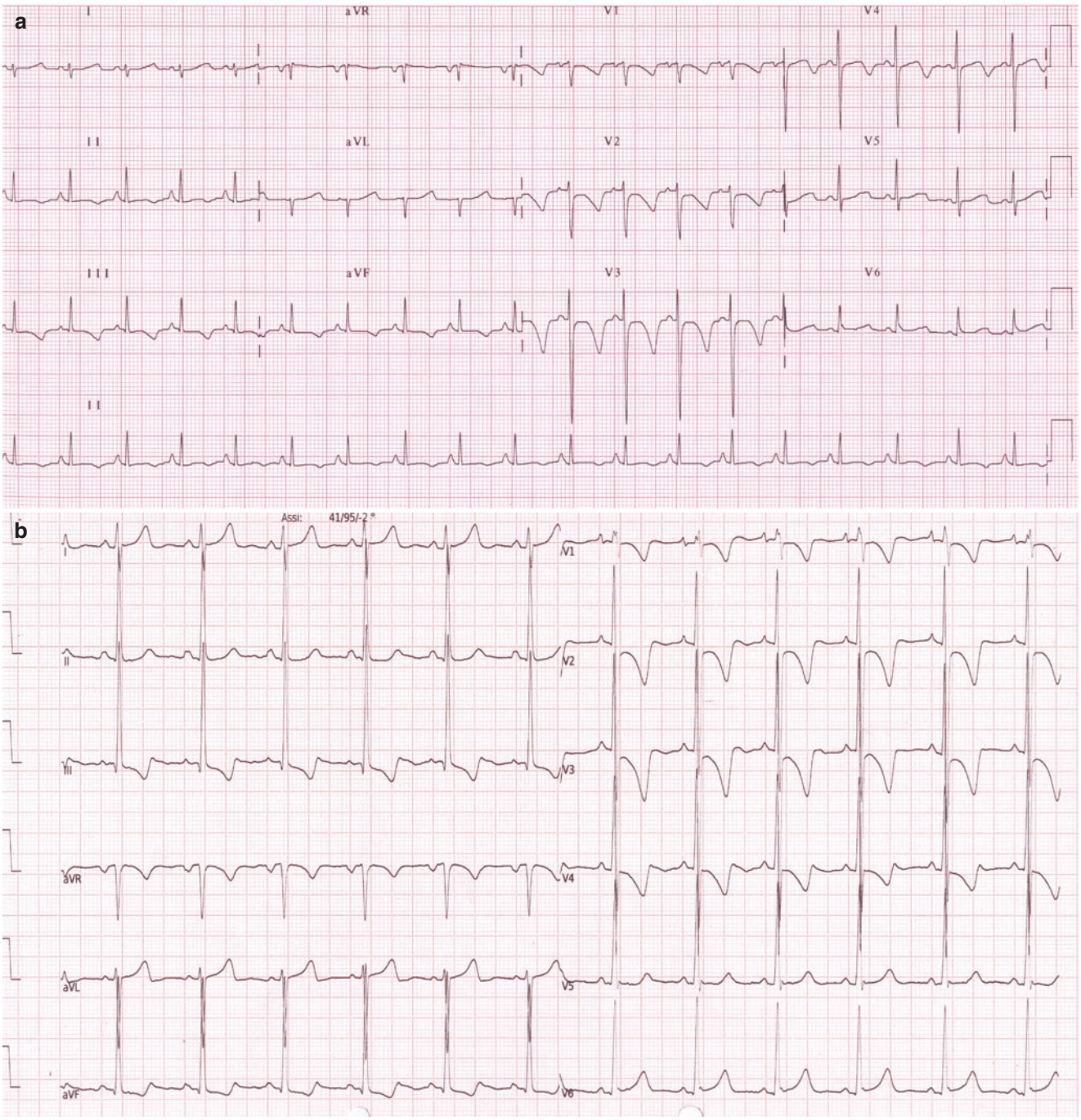

Fig. 15.6 (**a**) Six-year-old female with pulmonary hypertension in post-repaired left side diaphragmatic hernia. The RVH patter is atypical, type C like, probably because of the heart shifting and the lung hypoplasia. (**b**) Nine-year-old female with moyamoya syndrome and RVH due to peripheral pulmonary stenosis. Also in this case the RVH is atypical and the diagnosis is guided by a marked ST change in the precordial leads and by ancillary signs such as right axis deviation or right atrium enlargement

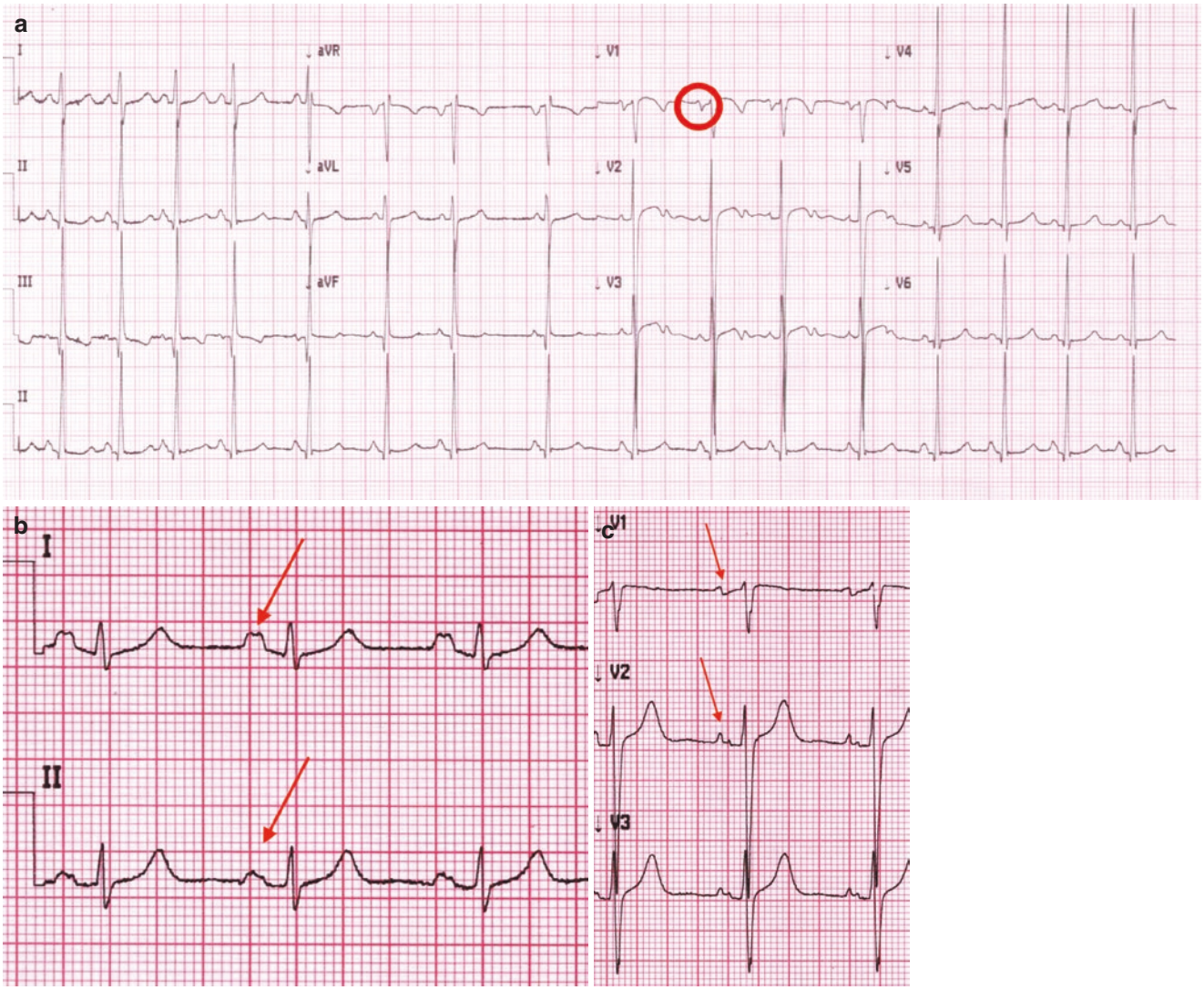

Fig. 15.7 (**a**) Nine-year-old male with moderate post rheumatic mitral stenosis (mean transmitral diastolic gradient 10 mmHg). Note the negative deflection in V1, denoting the left atrial enlargement. (**b**) As in the previous case, an 11-year-old male with mitral stenosis due to rheumatic fever. The P wave is bicuspid in lead II but does not meet the "adult" criteria for left atrial enlargement (P wave duration >0.12 s). (**c**) Close-up of the precordial leads

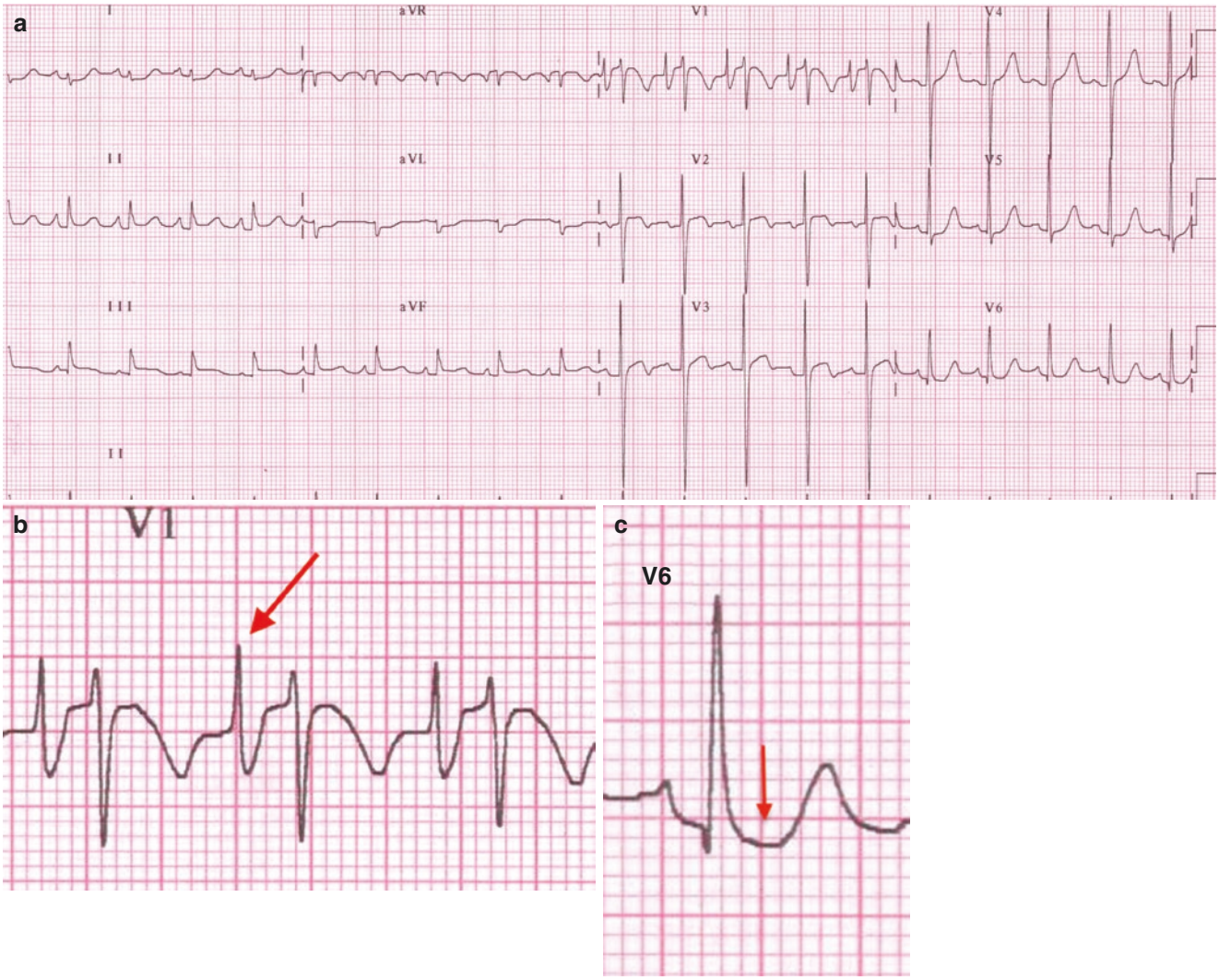

Fig. 15.8 (**a**) 13-year-old African female with severe mitral stenosis and moderate pulmonary hypertension (post rheumatic) on treatment with furosemide and digoxin. There is no apparent ventricular overload. (**b**) Note the right and left atrial enlargement. (**c**) A possible digitalis effect on ST segment in V6 can be noticed

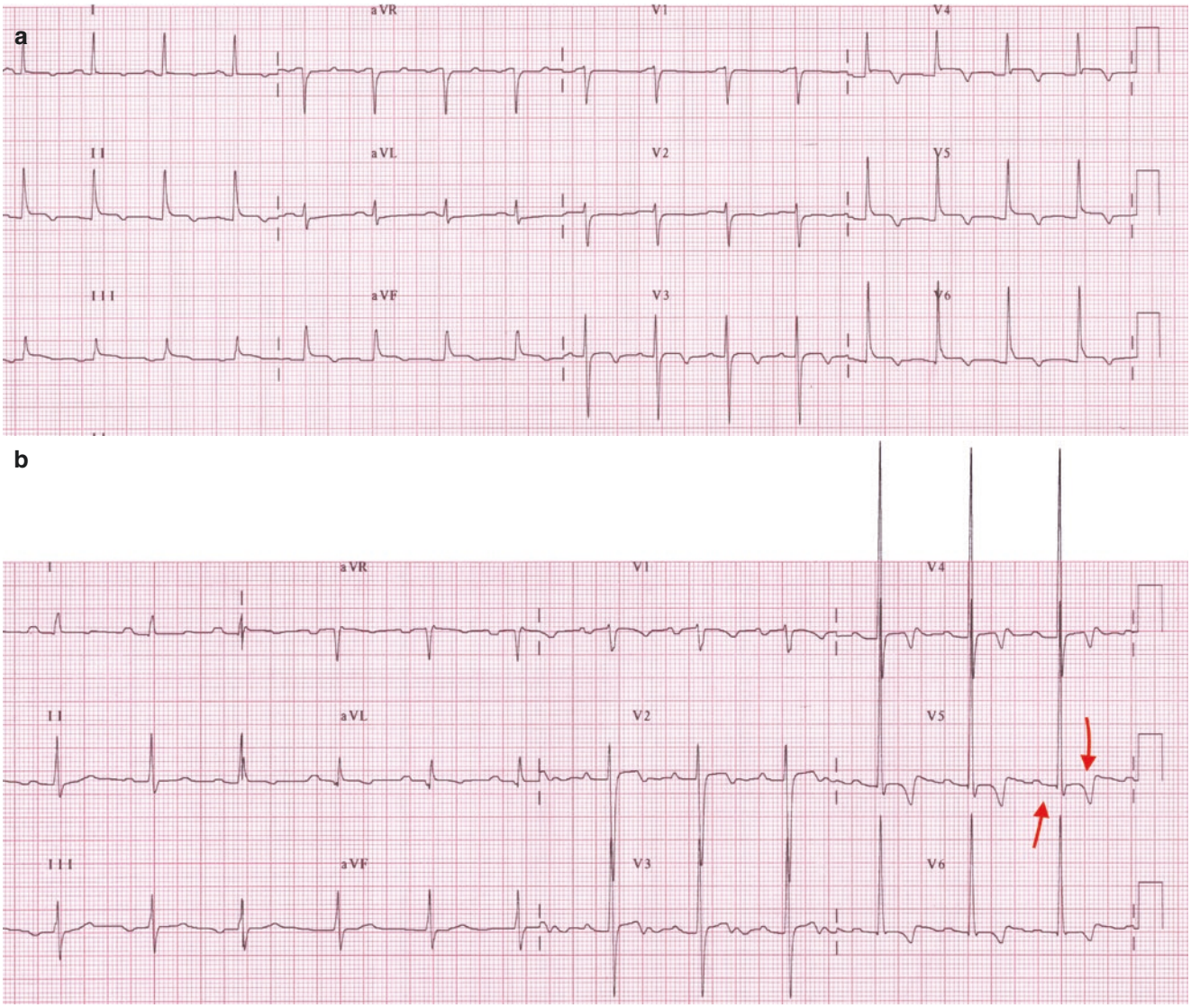

Fig. 15.9 Mitral insufficiency in subacute rheumatic fever. (**a**) Fifteen-year-old female, widespread ST-T changes but no other signs of ventricular overload. (**b**) Nine-year-old female, note the T changes as well as the PR prolongation.

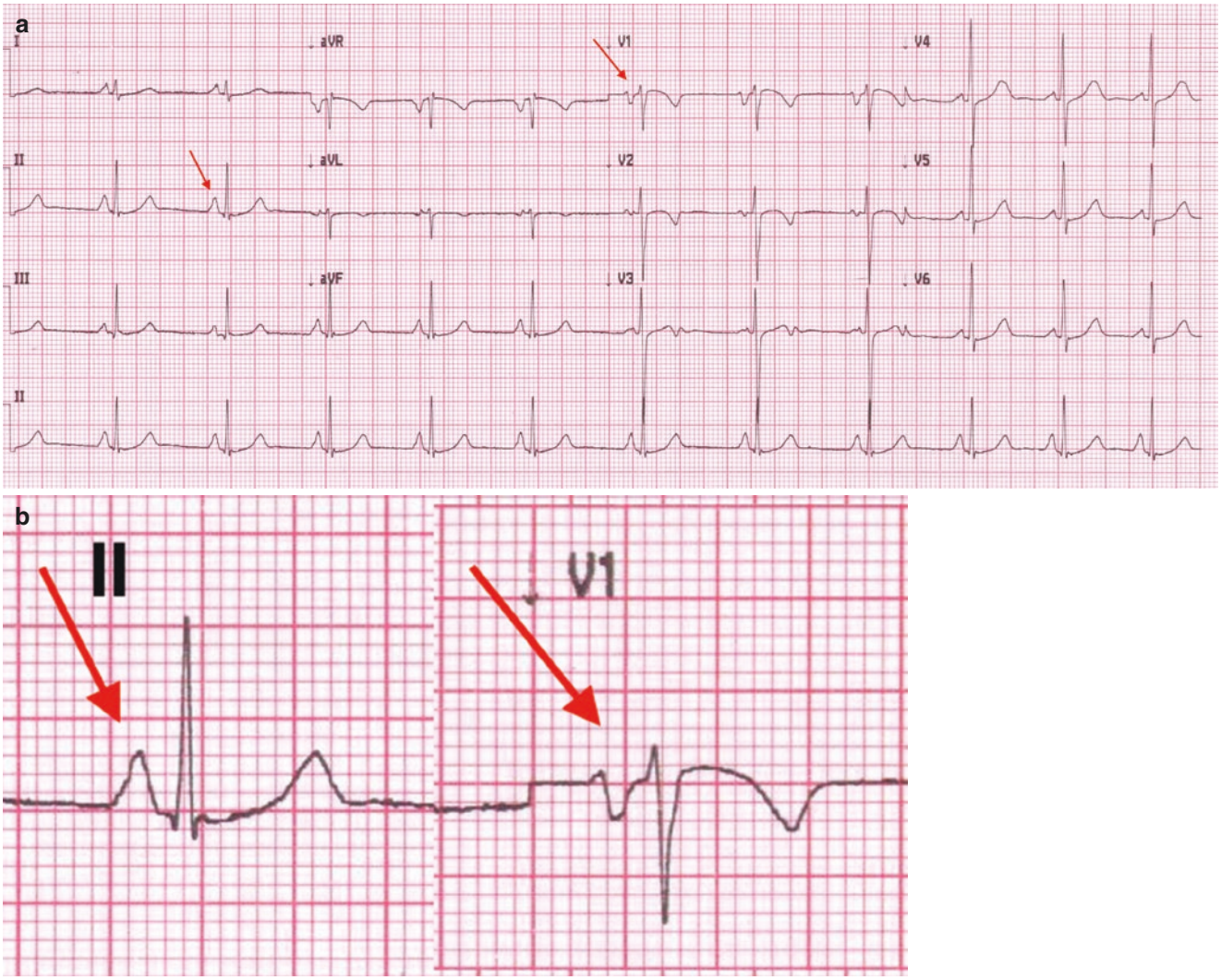

Fig. 15.10 (**a**) Nine-year-old male with moderate mitral steno-insufficiency with moderate pulmonary hypertension. (**b**) Close-up. Note the right (lead II) and left (lead V1) atrial enlargement

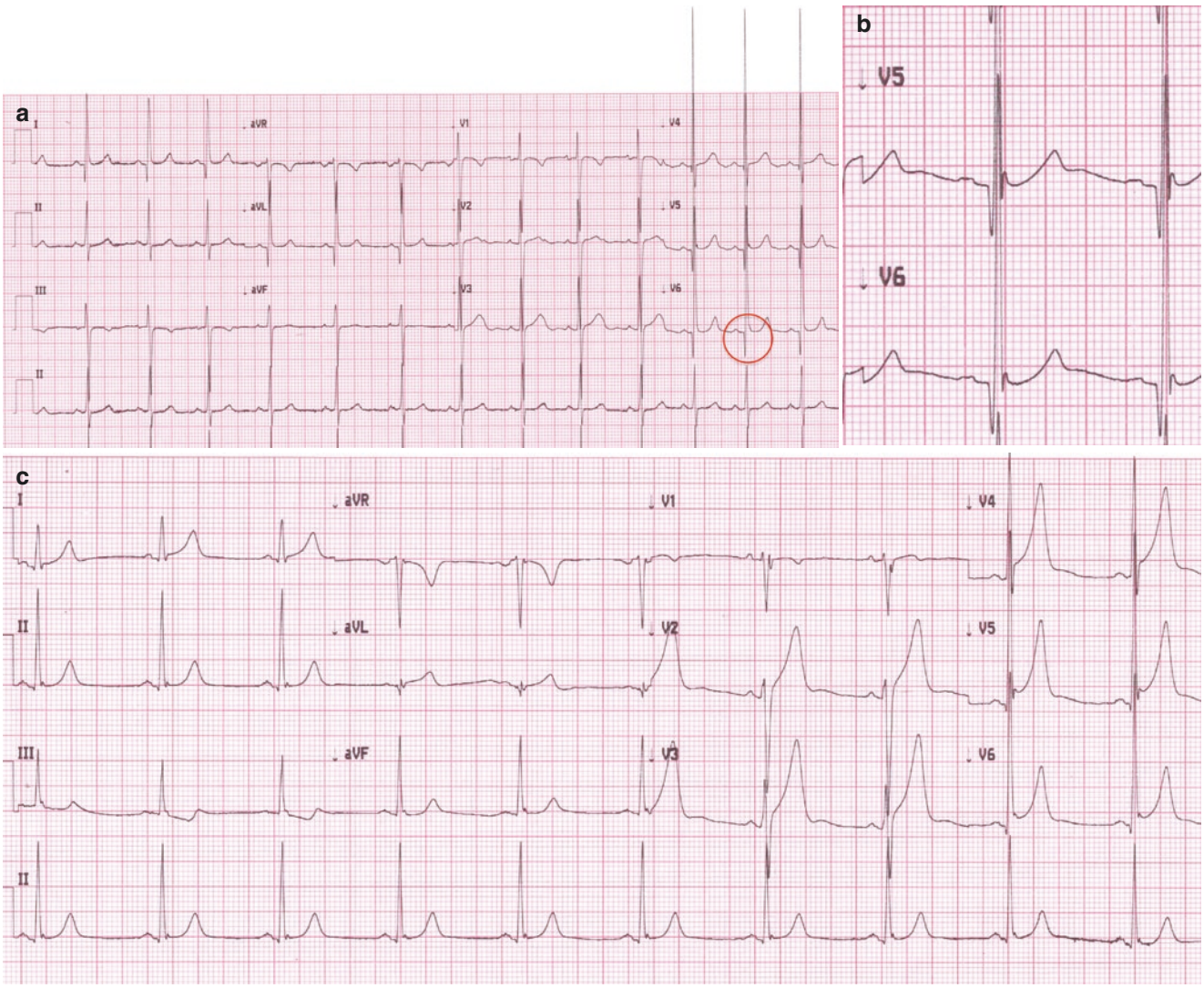

Fig. 15.11 Post rheumatic aortic insufficiency. (**a**) Fifteen-year-old male. The voltages of the R waves meet the criteria for LVH; in V6 there are deep Q waves (Cabrera sign). (**b**) Close-up. Another case of severe aortic regurgitation and deep Q waves in left precordial leads. (**c**) The sensitivity and specificity of the ECG in the left ventricle volume overload is usually poor. Tall T waves in the left precordial leads are described as a sign of volume overload, as it is possible to see in this 20-year-old male with severe aortic regurgitation

Suggested Reading

Beghetti M, Lador F. Editorial: pediatric pulmonary hypertension. Front Pediatr. 2015;3:105.

Marijon E, Mirabel M, Celermajer DS, Jouven X. Rheumatic heart disease. Lancet. 2012;379:953–64.

Maher KO, Tweddell JS. Aortic and mitral valve disease and left ventricular dysfunction in children. Pediatr Crit Care Med. 2016;17(8 Suppl 1):S131–9.

Channelopathies

16

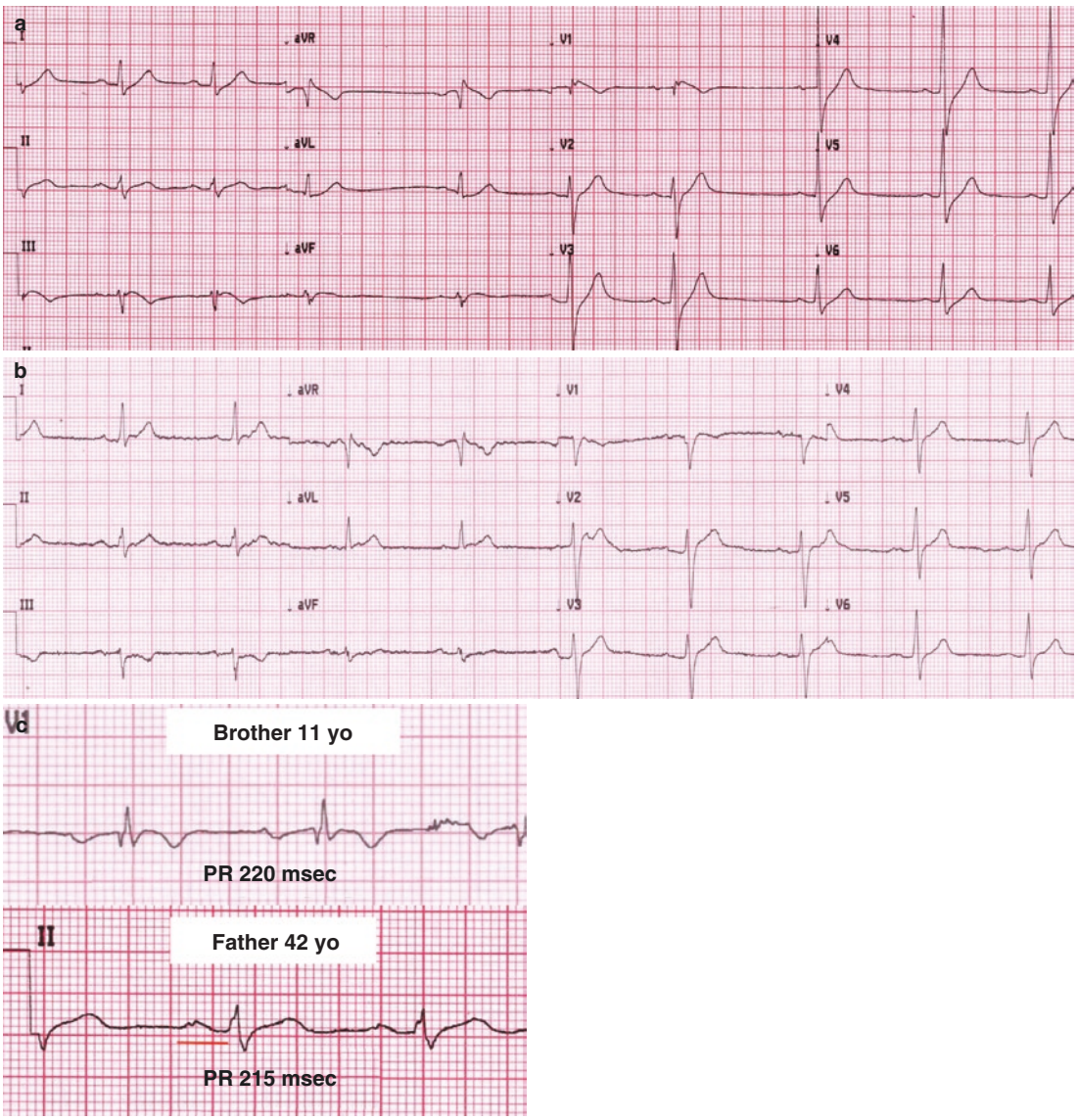

Fig. 16.1 Family with Brugada due to SCN5A mutation. All members are asymptomatic. (**a**) 9-year-old male. A saddleback pattern can be noted in V1 (Brugada type 2). Note also the first-degree AV block: PR is 215 ms, identical to the father (**b**) and similar to the brother (**c**). Regardless of the pattern in the right precordial leads, in young family members of Brugada syndrome, a first-degree AV block is highly suggestive of the disease. Also the left axis deviation should be considered an ancillary sign in Brugada patient

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G. Bronzetti, *Atlas of Pediatric and Youth ECG*, https://doi.org/10.1007/978-3-319-57102-7_16

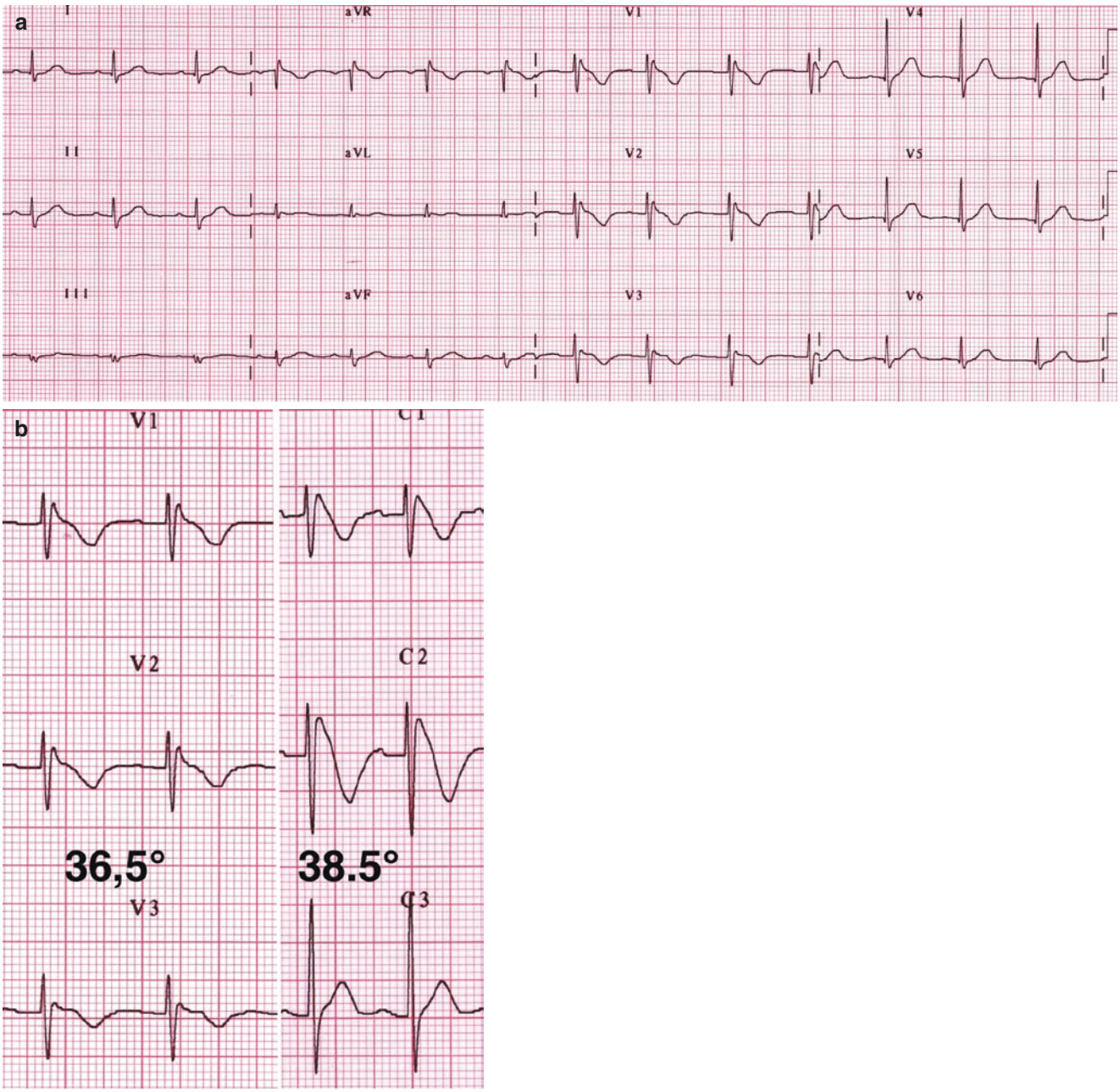


Fig. 16.2 (**a**) An 8-year-old girl admitted for pneumonia. There is a suggestive Brugada pattern. (**b**) During fever the right precordial leads are even more suggestive. The fever is known for its ability to unmask the Brugada pattern (type 1, coved). The genetic test was negative, as in more than 65% of people with Brugada pattern

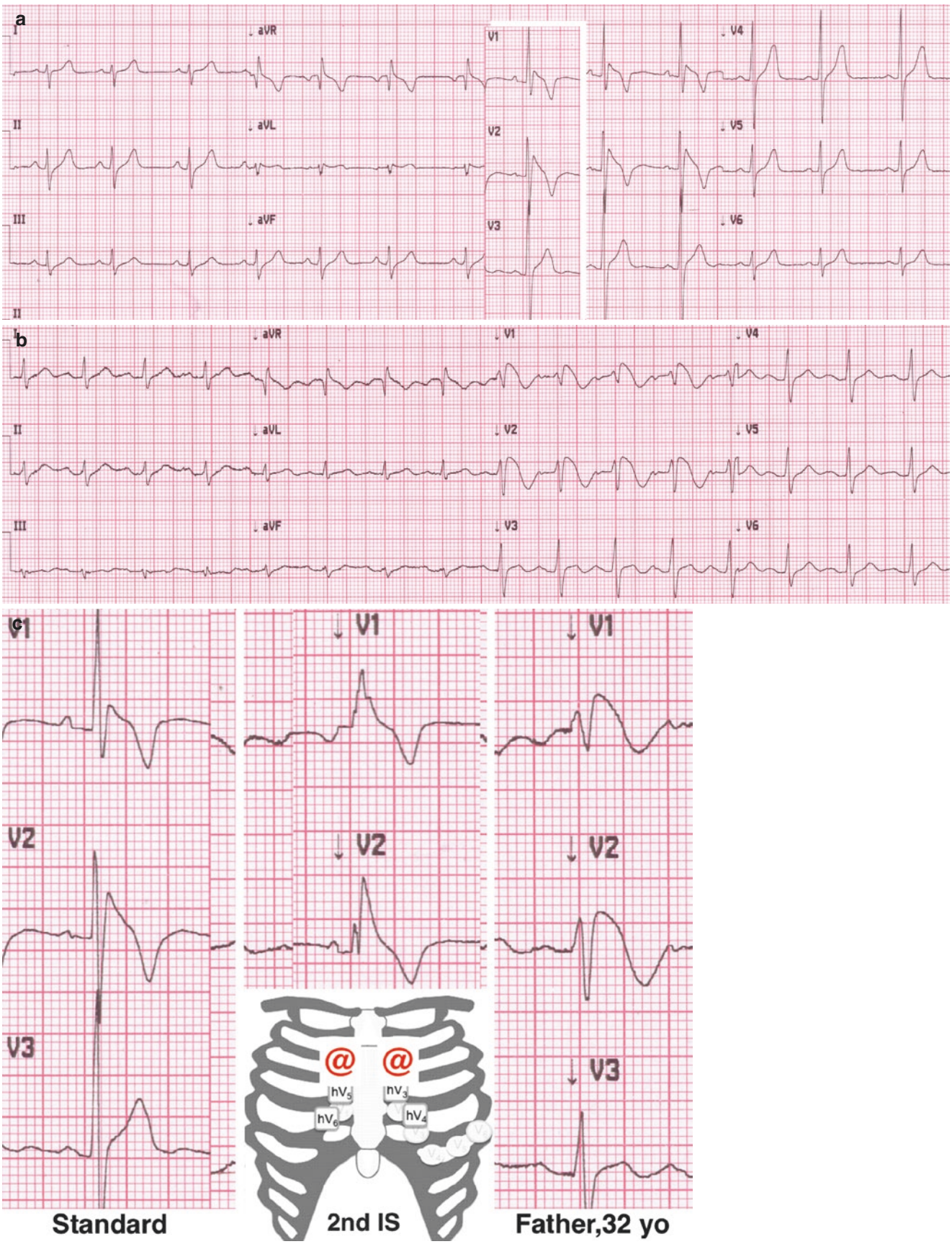


Fig. 16.3 (**a**) An 11-year-old male asymptomatic with pre-sport participation ECG. The right precordial leads present an atypical RBBB. (**b**) We did the ECG of the father (first ECG in his life), and the result was impressive, with a didactic Brugada pattern type 1 (Coved, ST elevation with at least 2 mm of elevation in at least 1 right precordial lead). (**c**) Moving V1 and V2 in the upper intercostal space (up to the second), the Brugada pattern is even more clear

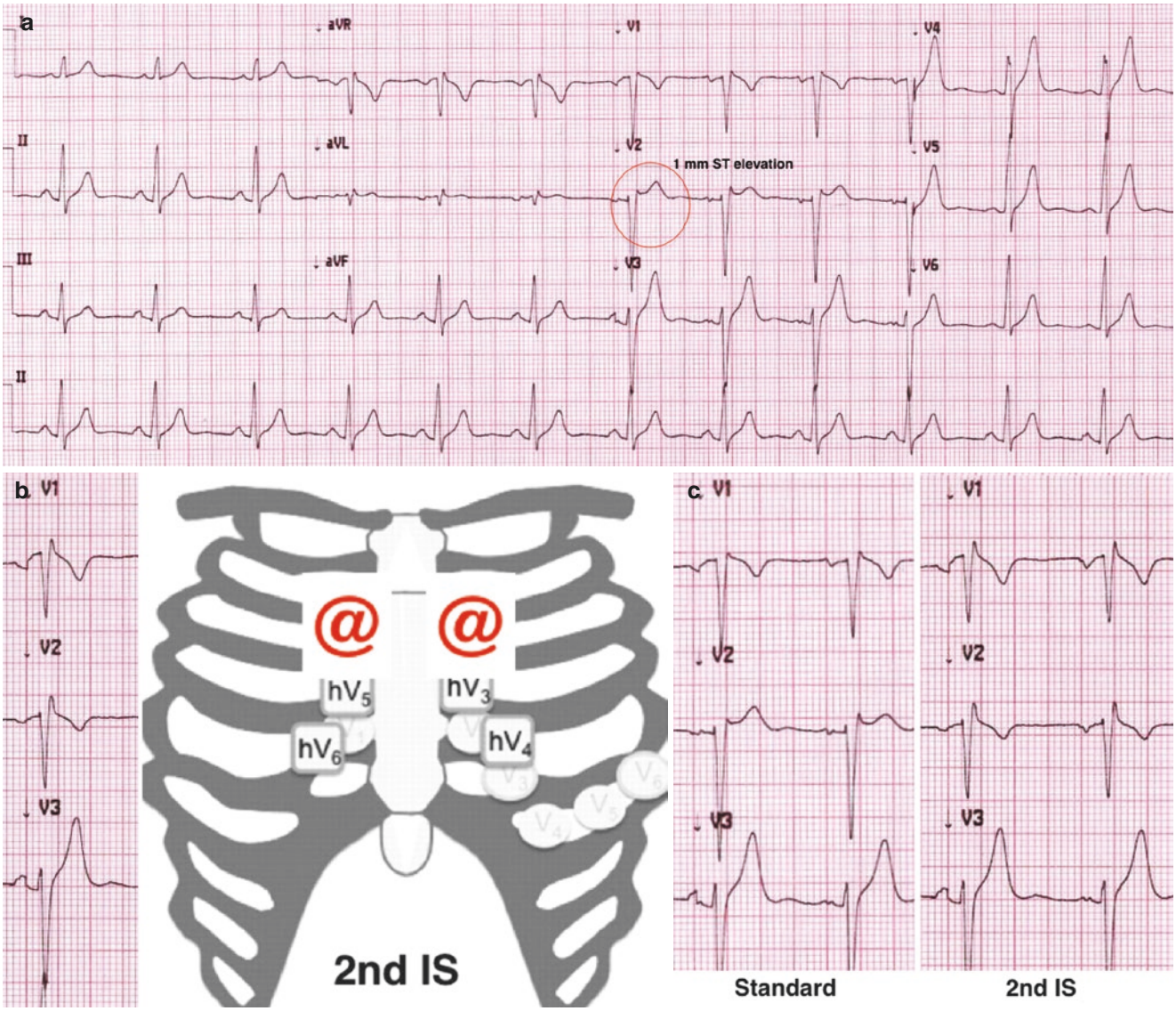


Fig. 16.4 (**a**) A 10-year-old asymptomatic male referred for Brugada pattern type 2 (saddleback ST elevation in right precordial leads). (**b**, **c**) Moving V1–V2 to the second intercostal space, the picture is that of the usual benign right conduction delay. All the family members have a normal ECG, and no genetic tests were performed

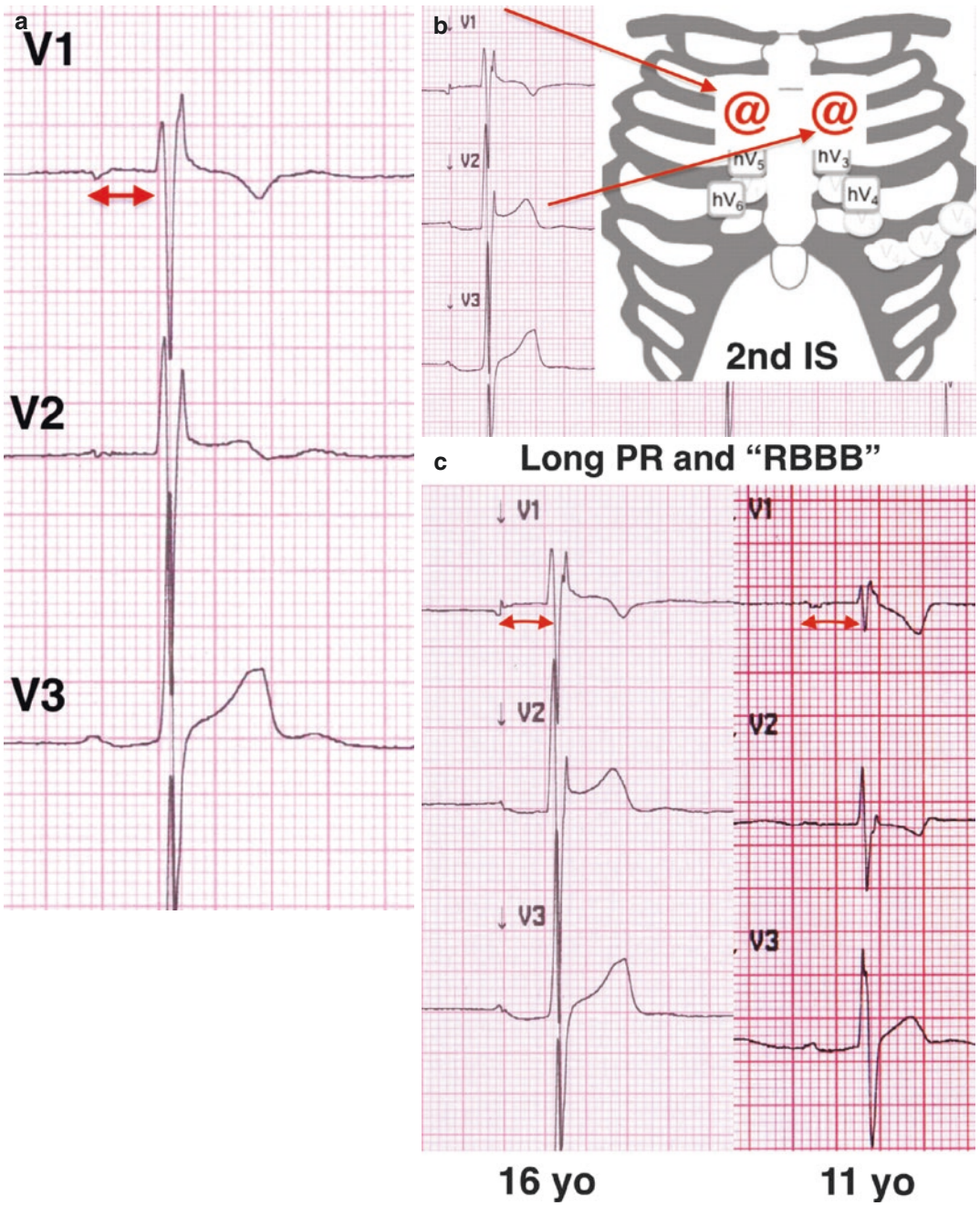


Fig. 16.5 (**a**) Standard right precordial leads of a 16-year-old male with profound sinus bradycardia and first-degree AV block. (**b**) Moving V1–V2 to the second intercostal space, a saddleback pattern in V2 can be noted. (**c**) Synopsis with another 11-year-old male, both are followed for bradycardia and long PR interval. In such cases, a red flag for Brugada or other channelopathies has to be raised

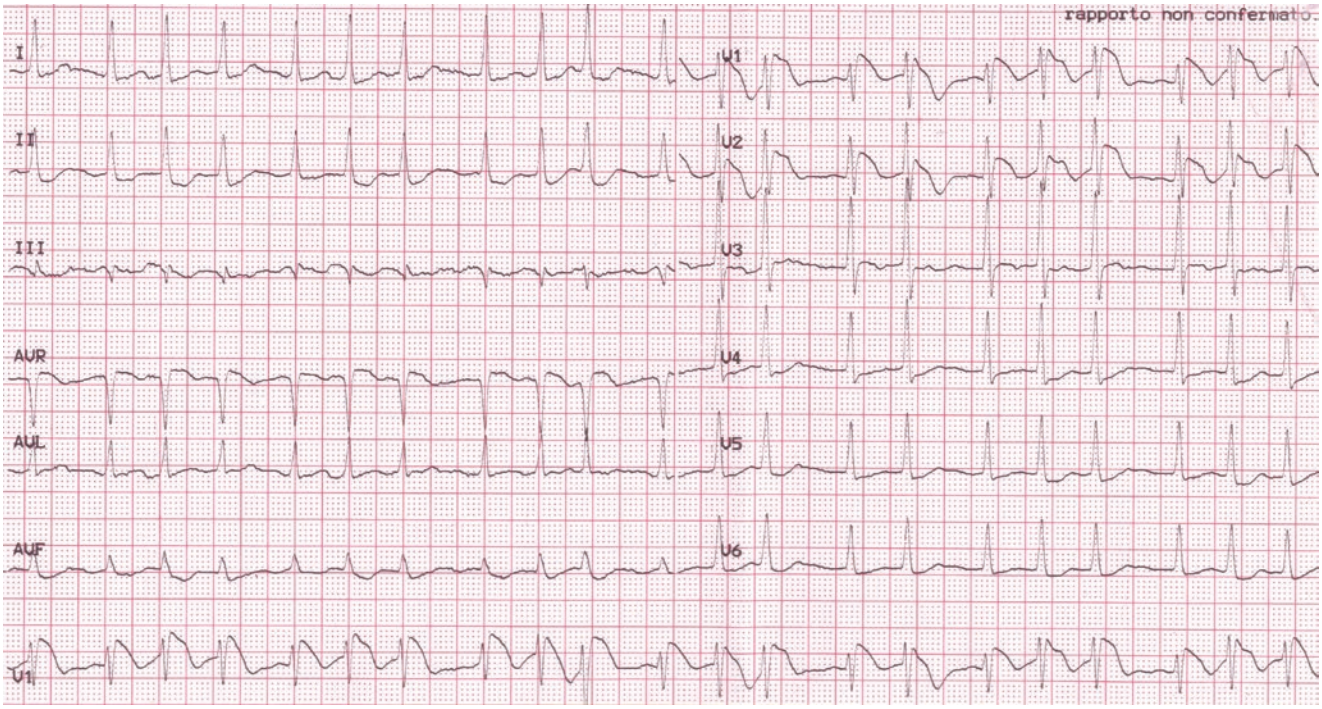


Fig. 16.6 A 22-year-old woman with Brugada syndrome and atrial fibrillation. As we said for bradycardia, unusual tachycardia must be seen with suspicion in children and young persons. AF in the young may be idiopathic, but channelopathies should be excluded

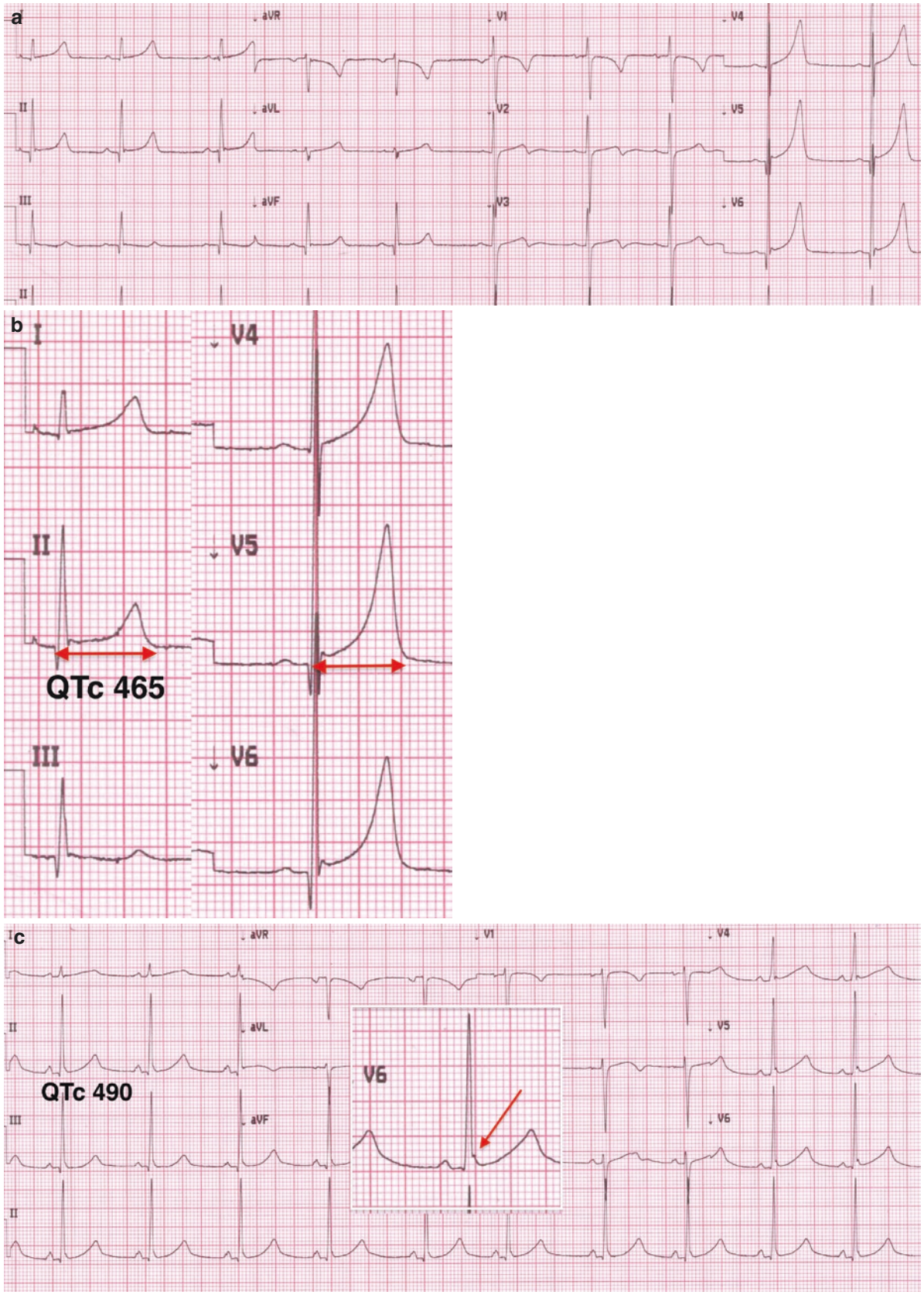
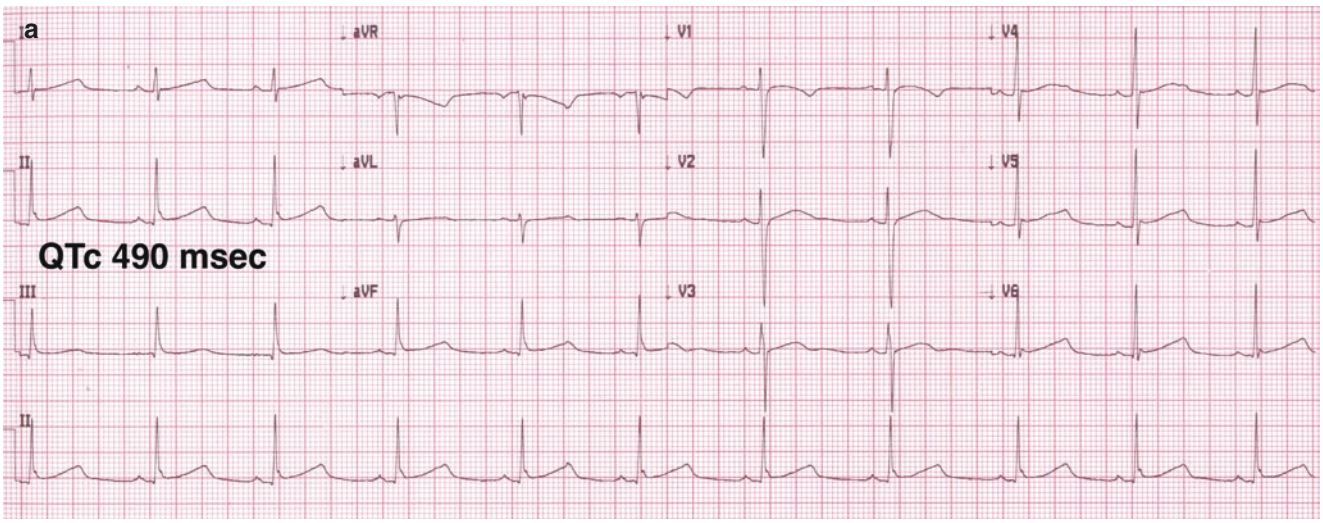


Fig. 16.7 (**a**) A 12-year-old male. The QT interval corrected by the Bazett is 465 ms. The patient is a LQT1 carrier due to mutation of the potassium channel-related gene. The T wave in V5 has a wide base and is tall; this can be the characteristic ECG phenotype of LQT1 but also compatible with the normal pattern for the age. (**b**) Close-up. (**c**) A 20-year-old female with LQT1 syndrome. A J wave due to benign early repolarization (*arrow*) can be noted: so an early repolarization is not incompatible with the *late* repolarization. Both patients are on treatment with nadolol



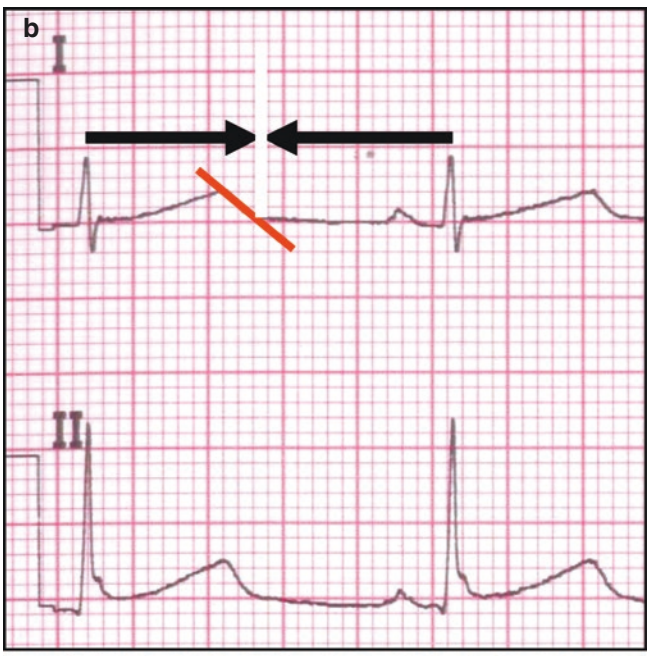


Fig. 16.8 (**a**) An11-year-old girl with LQT2 (loss of function of KCNH2). QTc is 490 ms. Although bradycardic due to therapy with nadolol, the T ends beyond the "center line" between the two R waves, denoting a visually elongated QT. The T wave in V5 is of relatively low amplitude and has a small notch in its proximal branch. (**b**) Close-up

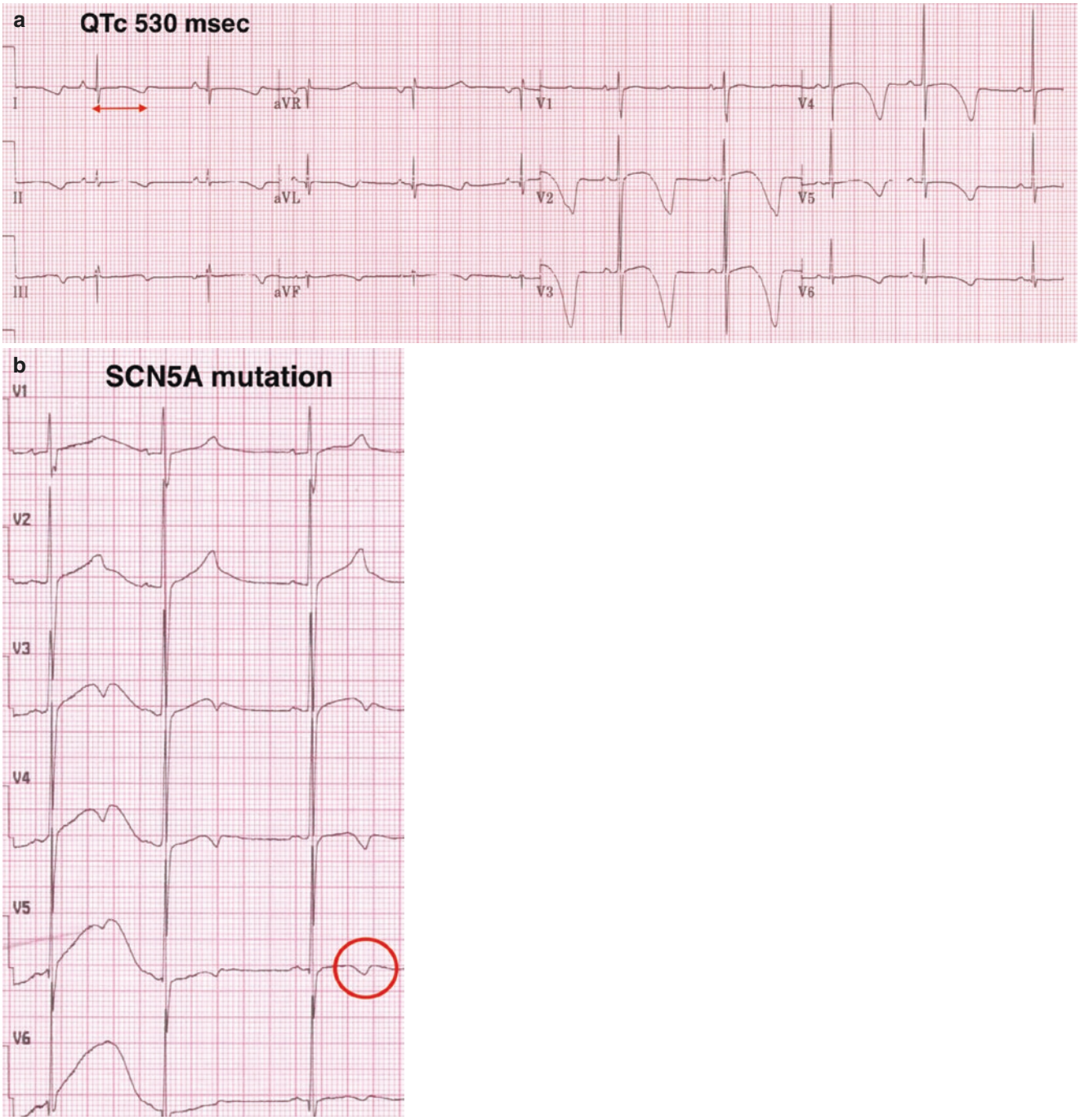


Fig. 16.9 (**a**) A 9-year-old male with LQT2. The QT is 530 ms, but widespread and marked ST changes are present. (**b**) A 12-year-old male with LQT3 (SCN5A mutation, gain of function of sodium channel). In the left precordial leads, the T waves are biphasic. Abnormal T waves are more characteristic of LQT2 and LQT3, compared with LQT1

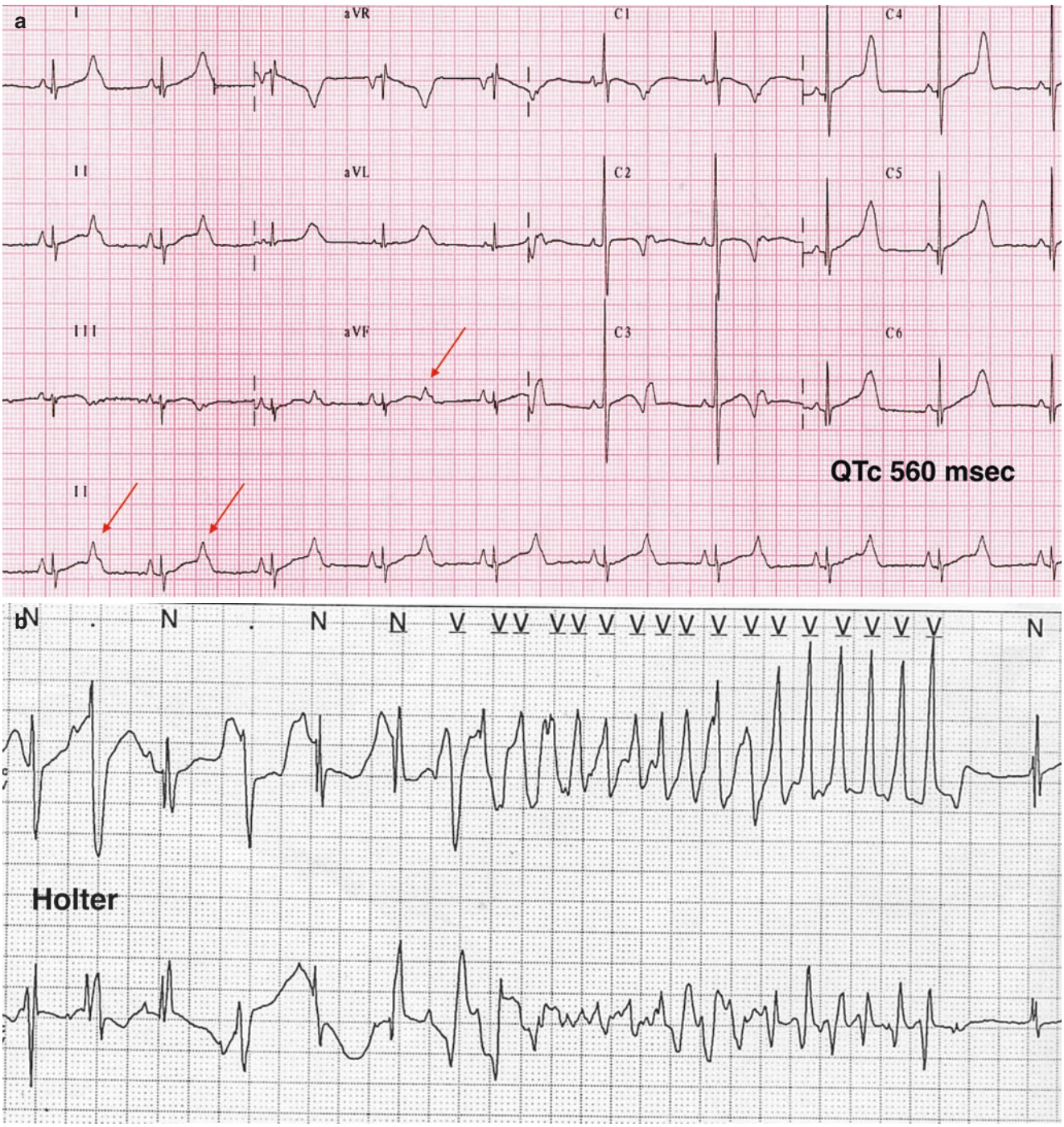
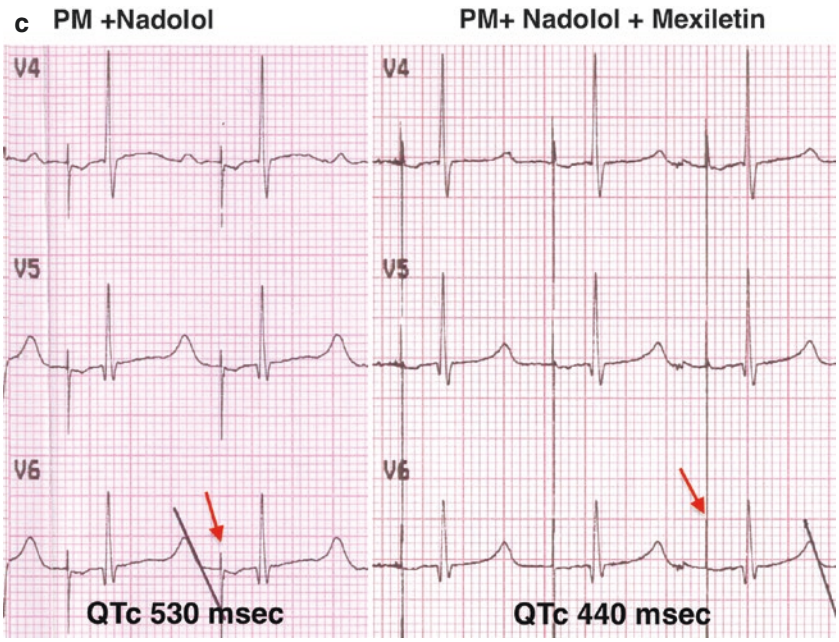


Fig. 16.10 (**a**) A 6-month-old boy presented with apparent life-threatening event (ALTE). The ECG shows 2:1 AV block, and the P waves are buried in the "late" T waves. The genetic testing was positive for LQT3 due to SCN5A mutation. (**b**) The Holter monitoring found a torsades de pointes. The patient received an epicardial dual chamber PMK and beta-blocker treatment with nadolol. (**c**) On adding oral mexiletine, the QT shortened dramatically (arrow = atrial spike)

Fig. 16.10 (continued)



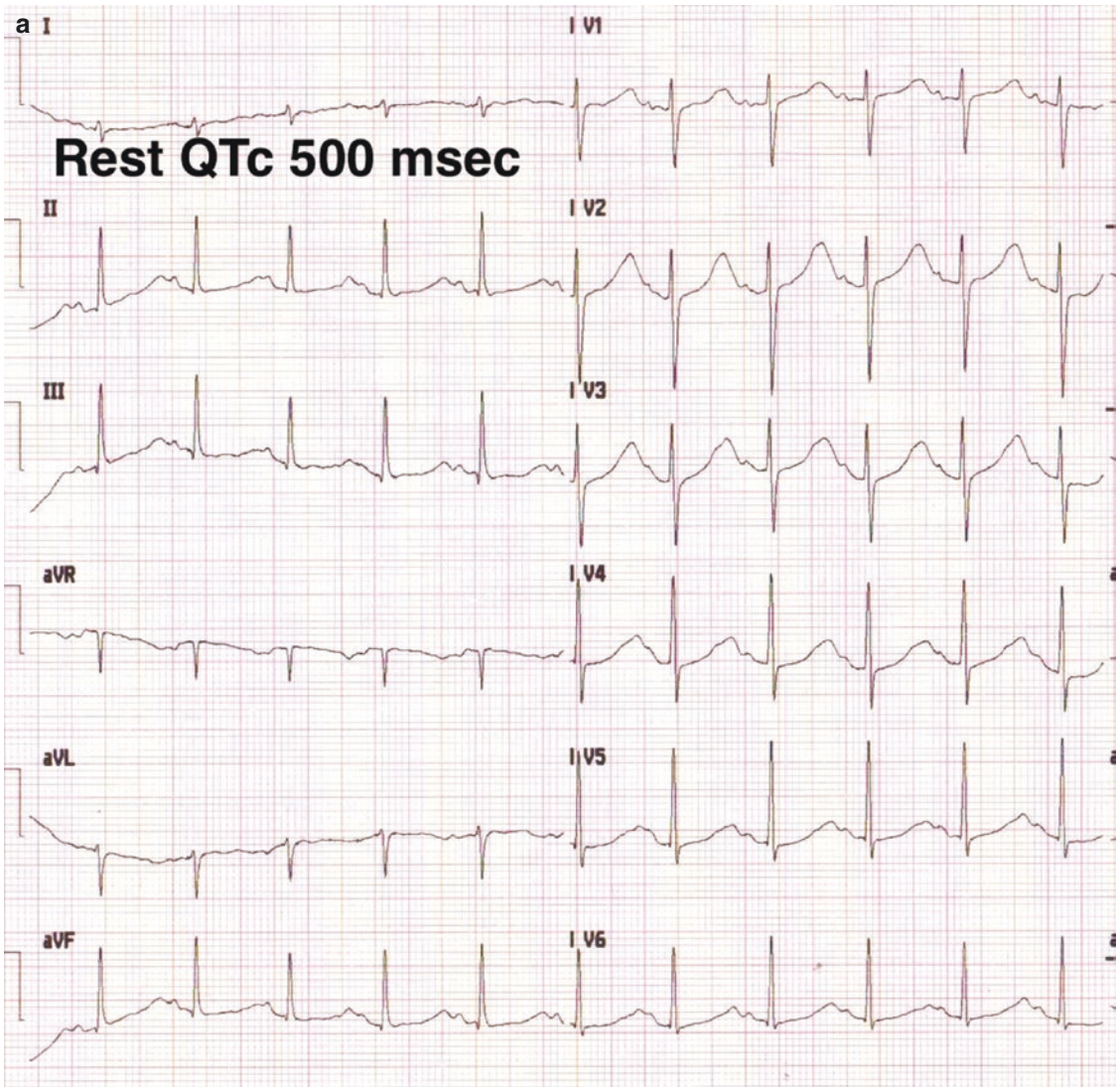
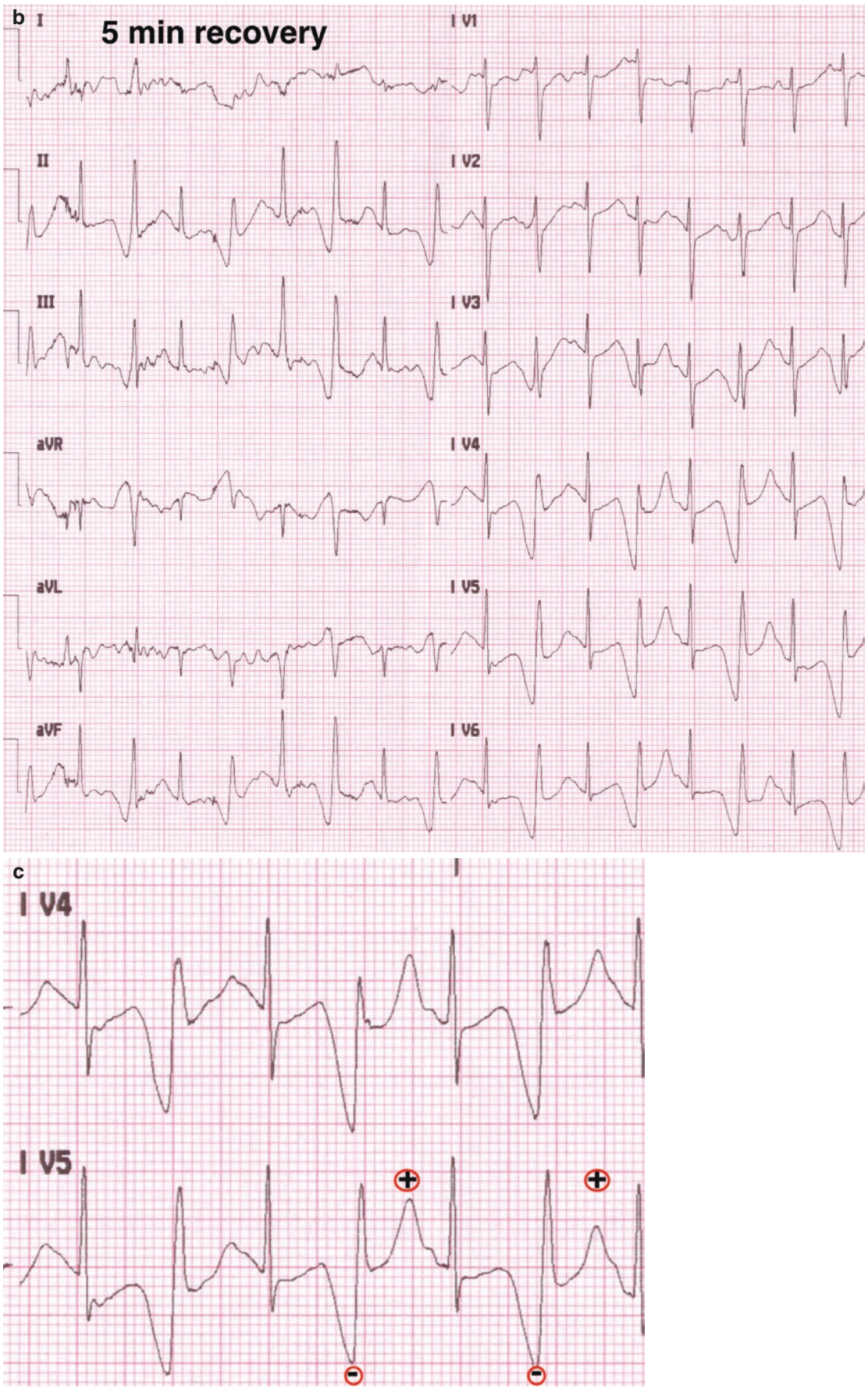


Fig. 16.11 A 16-year-old female with LQT1 syndrome. (**a**) Baseline ECG, (**b**) recovery phase of the exercise test. Note the T wave alternans, a prognostic marker in LQTS. In fact, it denotes the instability of repolarization. (**c**) Close-up



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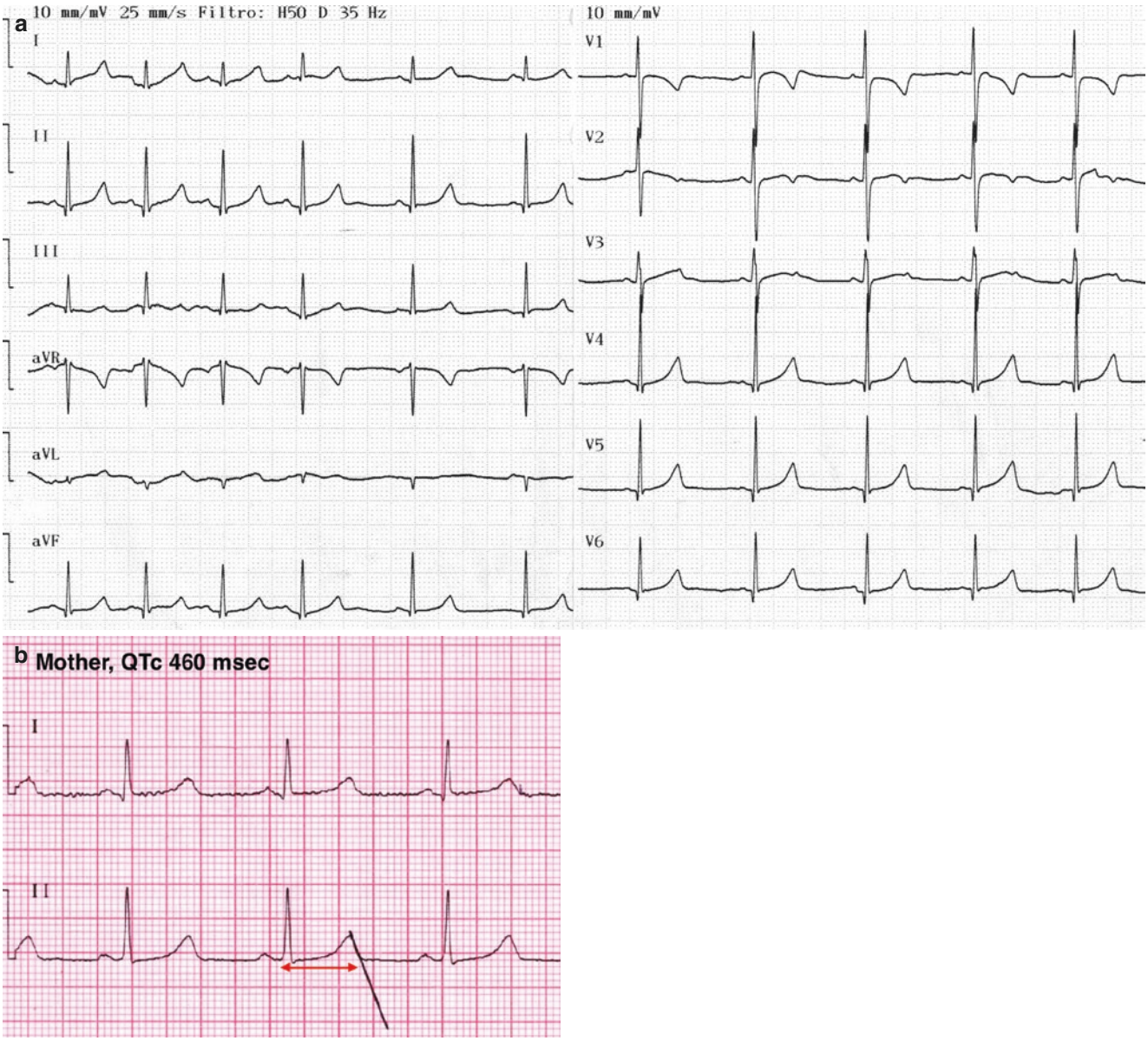


Fig. 16.12 (**a**) A 10-year-old female with syncope during effort. The baseline QTc is 460 ms. There was a strong family history for sudden death in the mother's family. The genetic testing found multiple mutations, two on the gene SCN5A and one on gene KCNQ1. Despite this shattering family and genetic history, the baseline QT is not that prolonged, even in the mother. (**b**) This case underscores the importance of history. In fact, an isolated QTc of 460 msec in a female should be considered normal

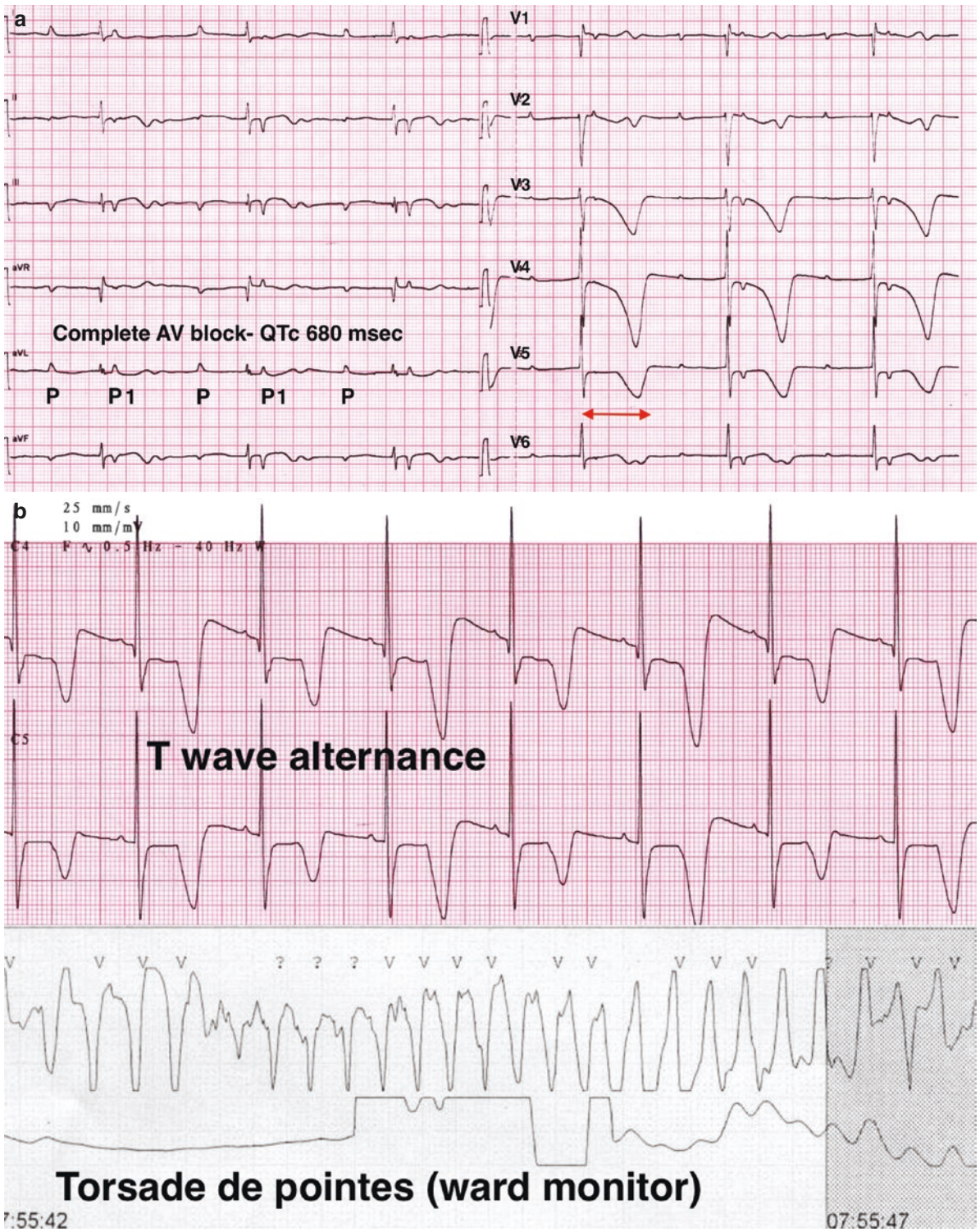
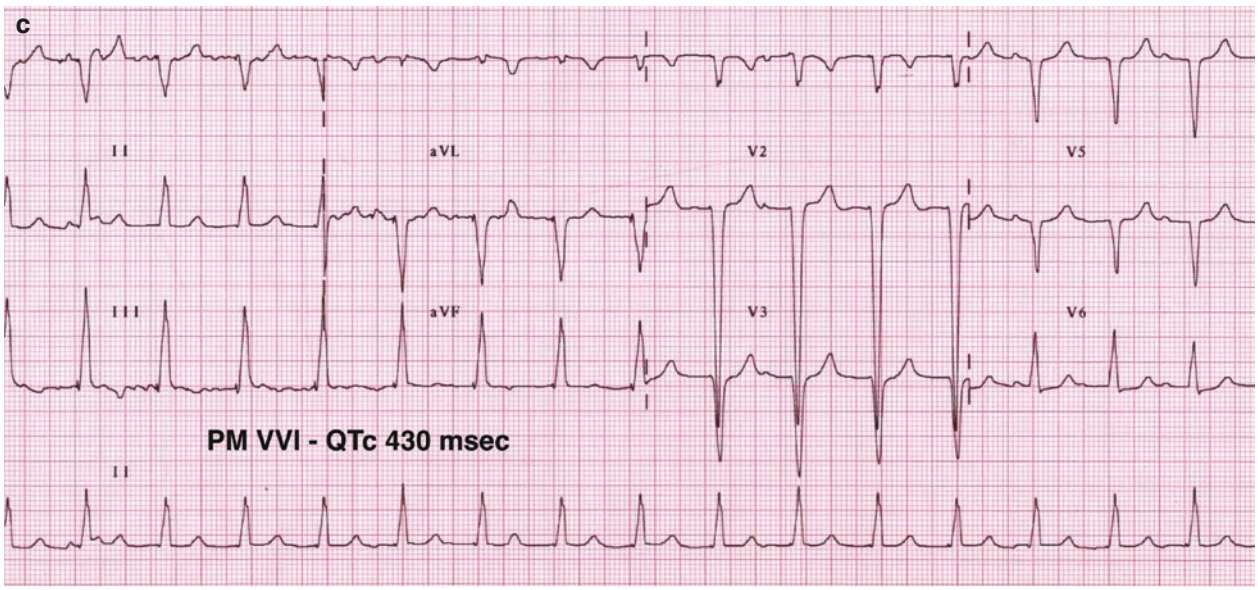


Fig. 16.13 An 8-year-old female, subaortic VSD repaired at 6 months of age. She had two syncopal episodes at home. (**a**) ECG in the ER where a total AV block was detected. A ventriculophasic dissociation is present: the atrial interval P-P 1 (the one which embraces the QRS) is shorter than the P1-P without QRS inside. The QTc interval is 680 ms (bear in mind that the Bazett formula tends to overcorrect in the case of very slow HR). Note the wide T waves. On the ward, treatment with isoproterenol increased the heart rate and shortened the QT interval. However, alarming phenomena were recorded, the T wave alternans. (**b**) Continuous monitoring: after repeated R on T phenomena, a torsades de pointes with spontaneous resolution occurred. (**c**) A VVI pacemaker was implanted and the QT returned to normal. This case is an example of bradycardia-dependent long QT syndrome, a form of acquired LQTS probably multifactorial and based on a genetic substrate different from the classic LQTS (Romano-Ward)



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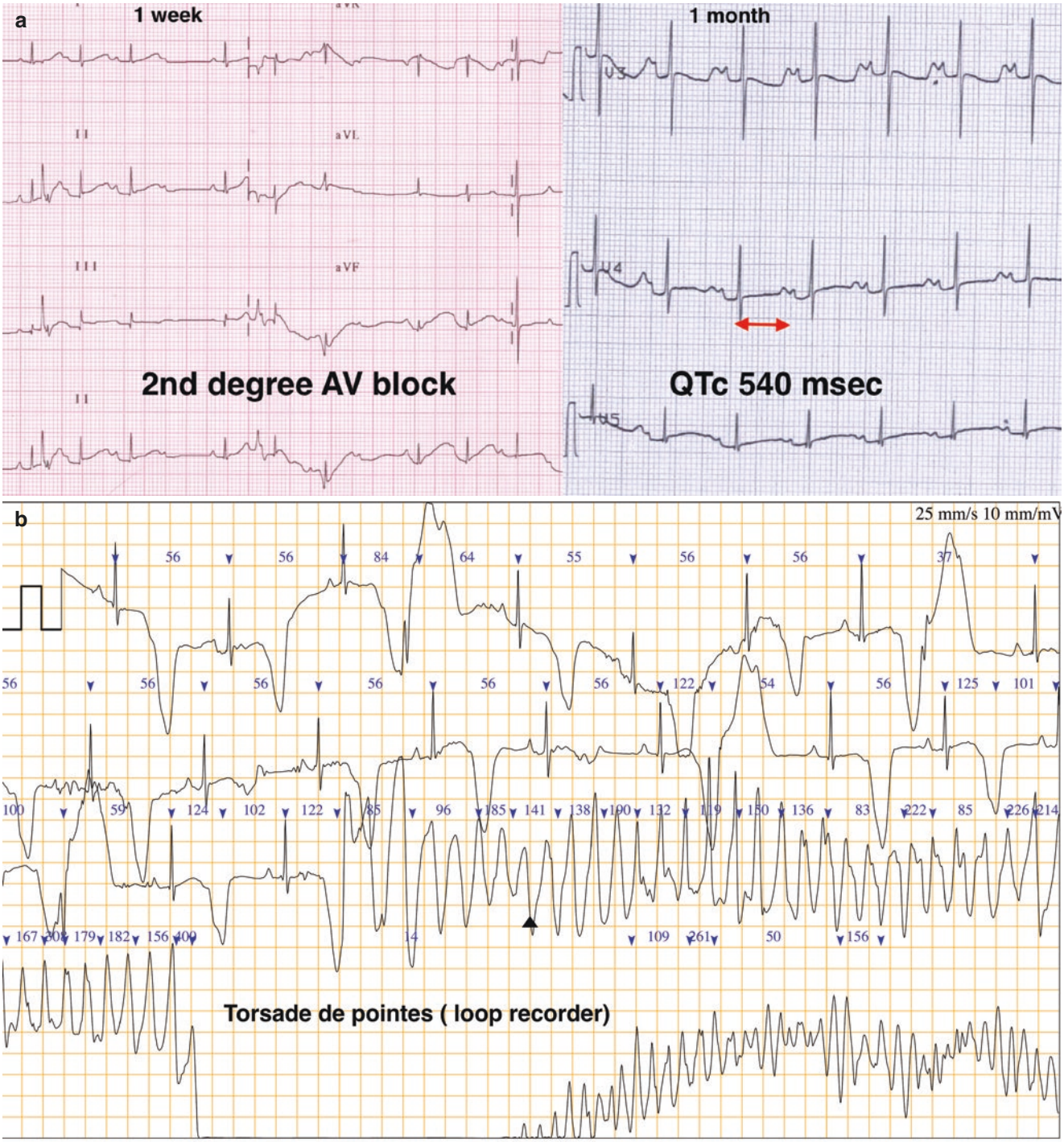

Fig. 16.14 Neonate in a family with history of pacemaker implantation in youth. (**a**) First- and second-degree AV block and LQT. (**b**) While wearing a loop recorder, a torsade de pointes occurred. After pacemaker implantation, the QT returned to normal. As in the previous figure, this is another case of bradycardia-dependent LQTS

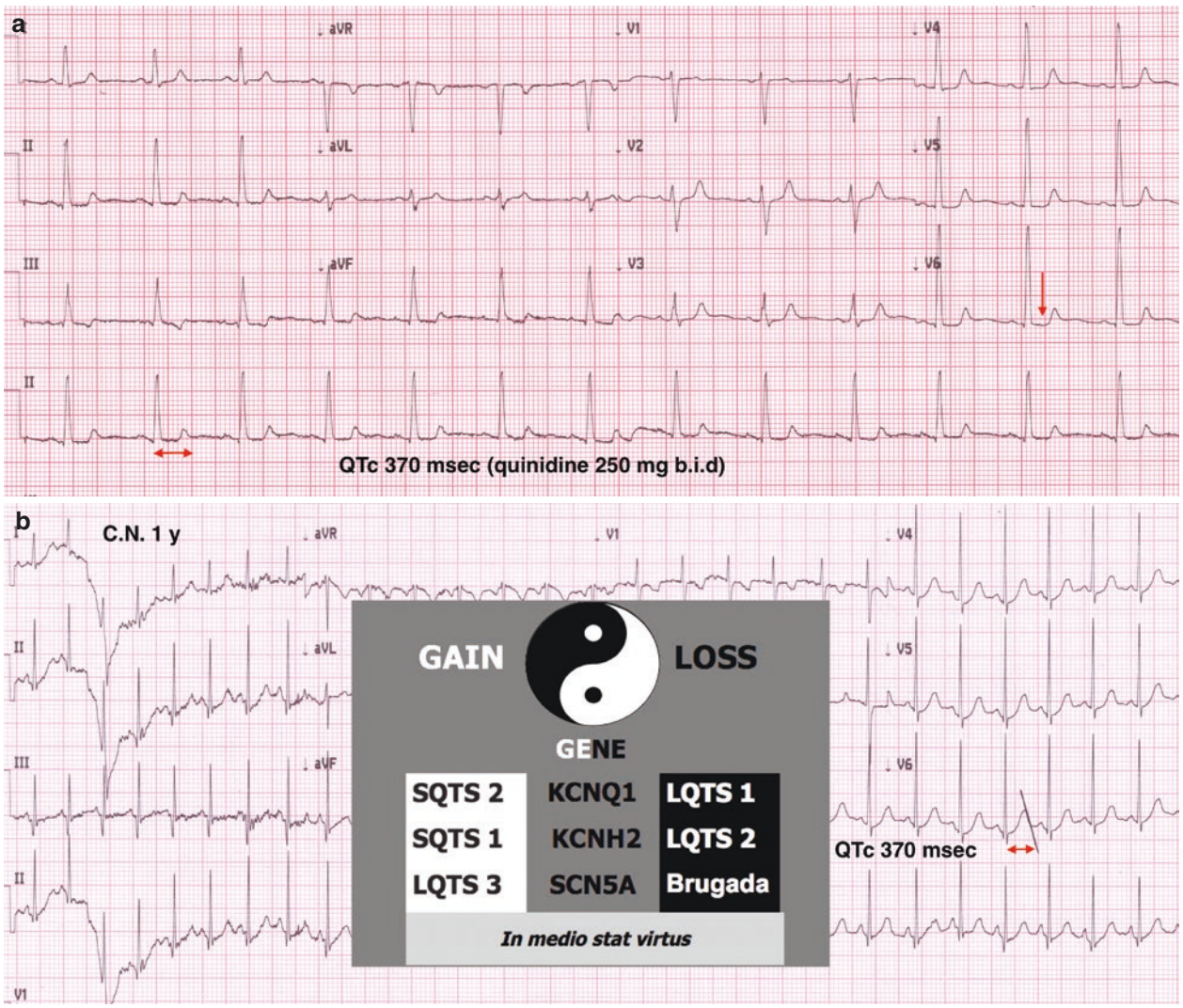

Fig. 16.15 (**a**) A 30-year-old woman with short QT syndrome (SQT1) due to KCNH2 mutation, on treatment with oral quinidine. (**b**) The neonate daughter has a short, but not too short QT (QTc 370 ms), and the genetic testing is negative. Most cases of SQTS are due to a gain of function of potassium channels

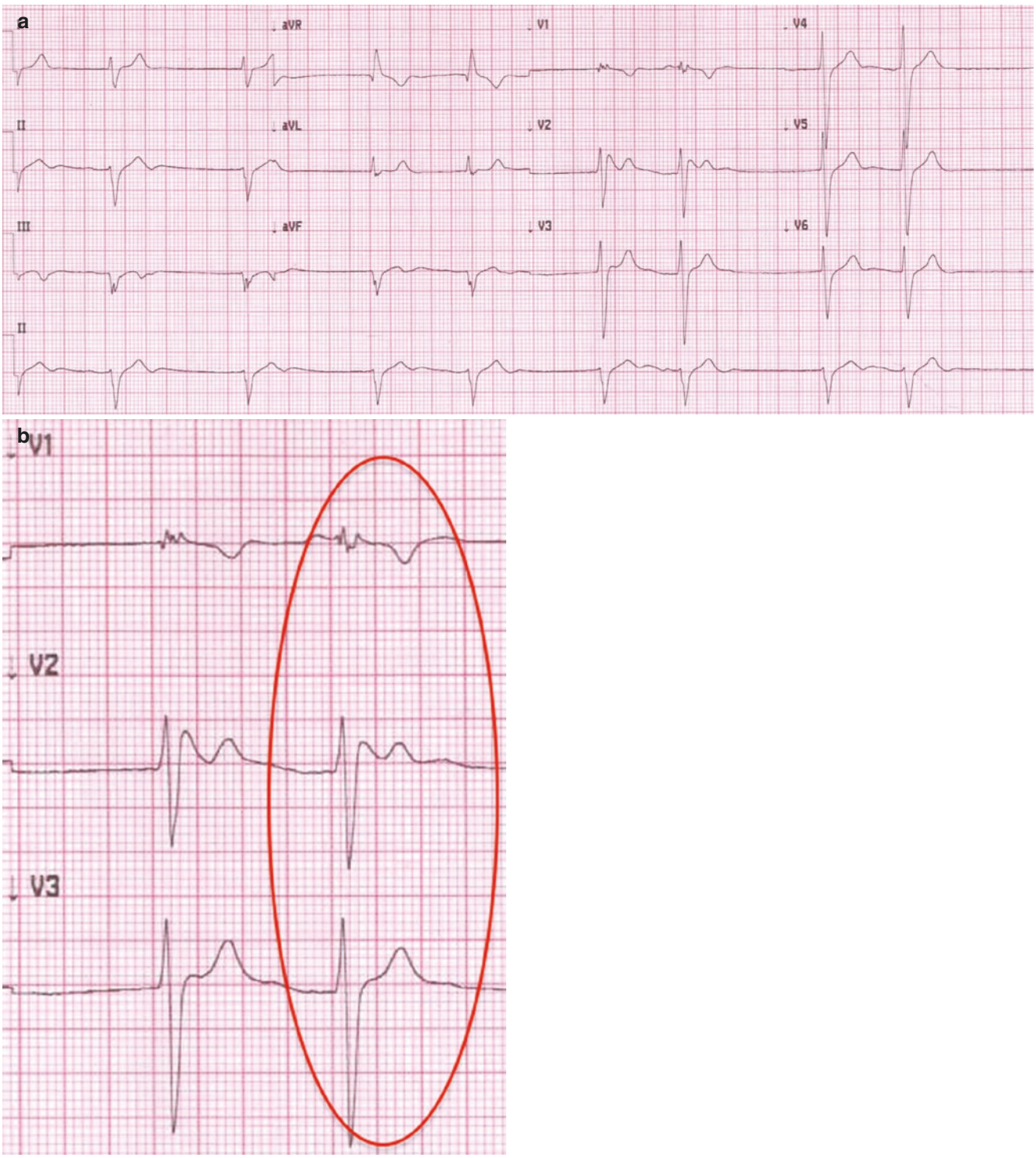

Fig. 16.16 A 24-year-old male followed since infancy for sick sinus syndrome. Although the boy was asymptomatic, Holter monitoring showed frequent pauses of sinus arrest, episodes of first-degree AV block, junctional rhythm, and paroxysmal AF. He underwent pacemaker implantation at 8 years. (**a**) There is sinus arrest and junctional escape rhythm. The genetic test found two novel mutations in the SCN5A gene, one inherited from the mother and one from the father. Neither variant seems by itself sufficient to cause a phenotype, supporting the recessive inheritance. (**b**) The right precordial leads are Brugada-like (type 2), another disease due to defective mutation of SCN5A

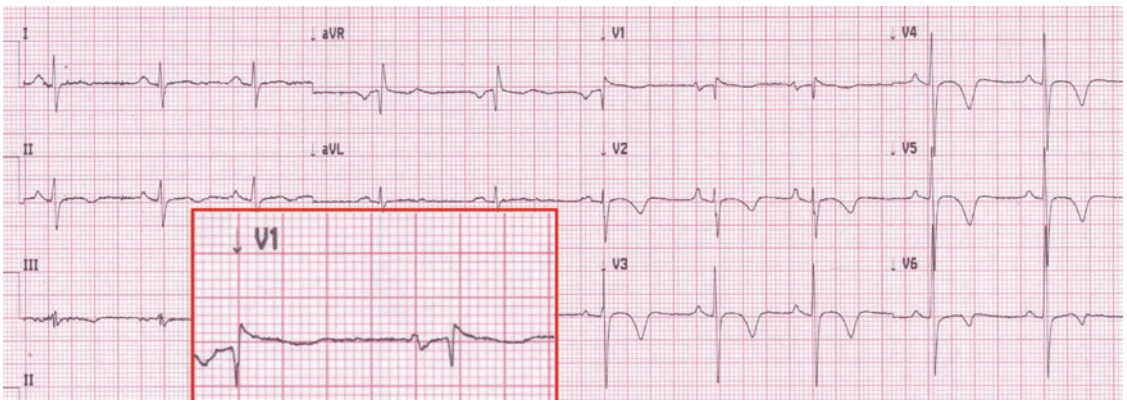

Fig. 16.17 A 3-year-old boy with recurrent paroxysmal atrial flutter. There is sinus bradycardia; in V1 the J point is elevated and in V6 the S wave is deep, as in the previous case (large S in V6 wave is not specific but recurrent in Brugada pattern). Genetic testing is ongoing (SCN5A, LMNA, Cx40, EMD), but there is a strong suspicion of a mutation of the SCN5A. As mentioned above, these S waves are apparently the reciprocal of the R wave in V1 and believed by many to be benign, whereas they could be a sign of Brugada syndrome. Another sign of BS, absent in this case, is the ST depression in the inferior leads. In fact, in BS the pathological vector that causes the ST elevation in the right precordial leads is the same as the one that causes the ST depression in the inferior leads, making these inferior leads the true reciprocal

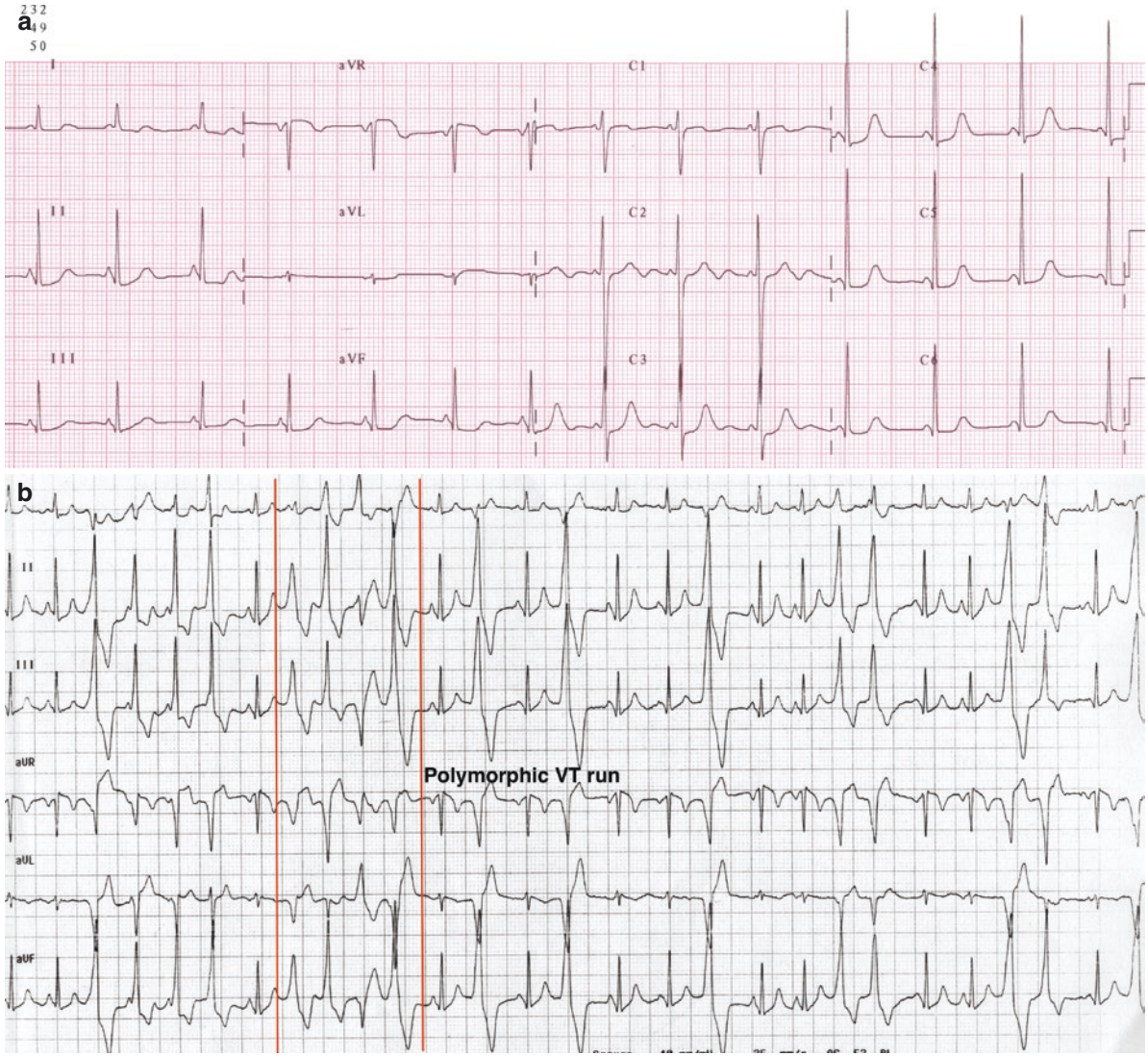

Fig. 16.18 (**a**) A 9-year-old girl with palpitation during effort. The ECG showed short PR but no delta wave suggestive of pre-excitation. (**b**) The exercise test induced run of VT. Genetic testing is in progress. The association of short PR and adrenergic-dependent ventricular arrhythmias is suggestive of catecholaminergic polymorphic ventricular tachycardia (CPVT). Care should be taken to not misinterpret short PR and wide complex tachycardia for pre-excitation syndrome!

Suggested Reading

Fernández-Falgueras A, Sarquella-Brugada G, Brugada J, Brugada R, Campuzano O. Cardiac Channelopathies and sudden death: recent clinical and genetic advances. Biology (Basel). 2017;6. pii: E7.

Lieve KV, Wilde AA. Inherited ion channel diseases: a brief review. Europace. 2015;17(Suppl 2):ii1–6.

Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes: document endorsed by HRS, EHRA, and APHRS in May 2013 and by ACCF, AHA, PACES, and AEPC in June 2013. Heart Rhythm. 2013;10:1932–63.

Ischemic Disease

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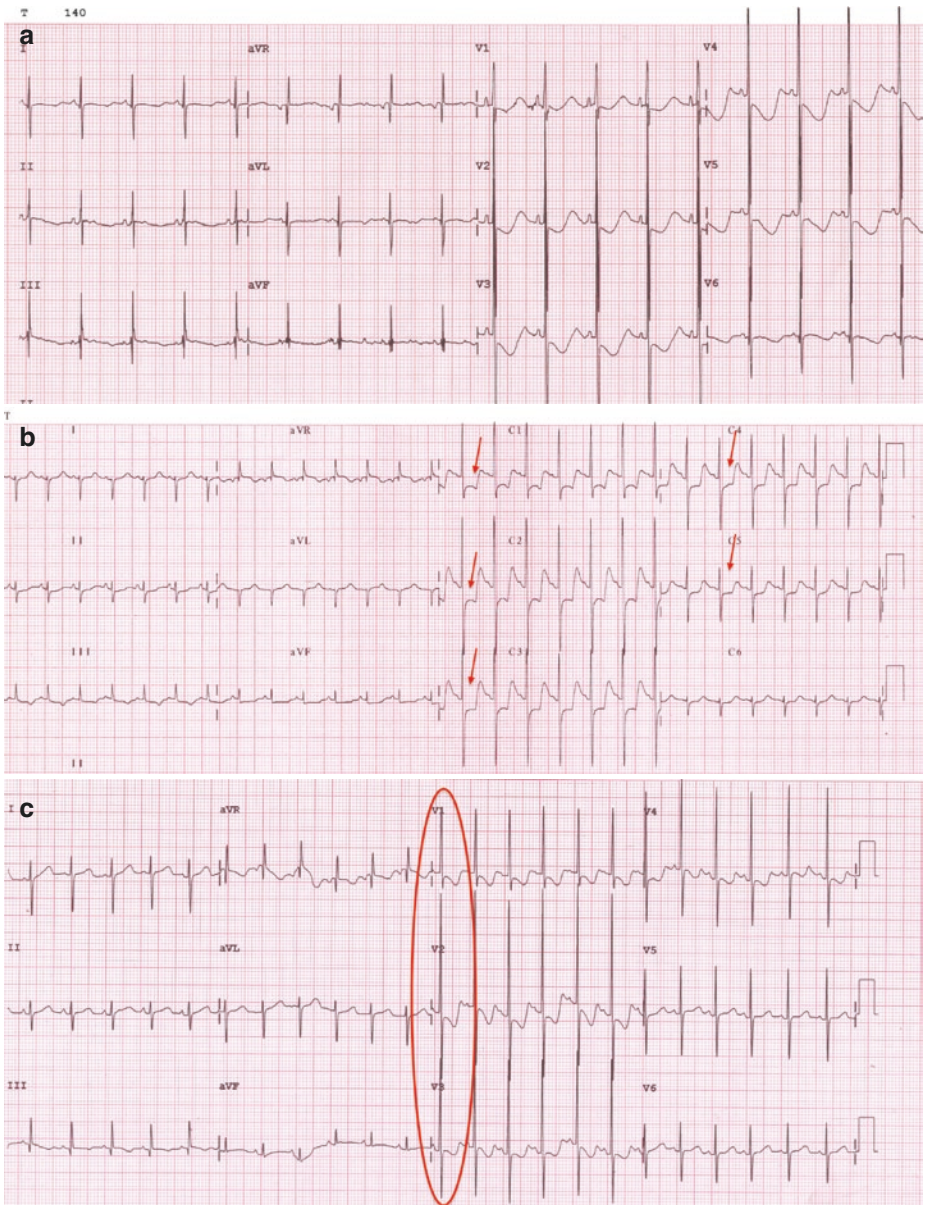

Fig. 17.1 Persistent pulmonary hypertension of the newborn. Many etiologies—from dystocia to sepsis—can lead to this condition also known as transient myocardial ischemia of the newborn infant. The clinical presentation may vary from mild oxygen dependency to need for ECMO. (**a**) Severe ischemia with widespread ST changes. (**b**) Moderate ischemia, the ST depression is less diffuse than case a. (**c**) Mild PPHN. (**d**) Usually the normalization takes a few days. Although this is the pathophysiology of almost all ischemic ECGs in the newborn, the echocardiogram has to rule out a congenital coronary anomaly

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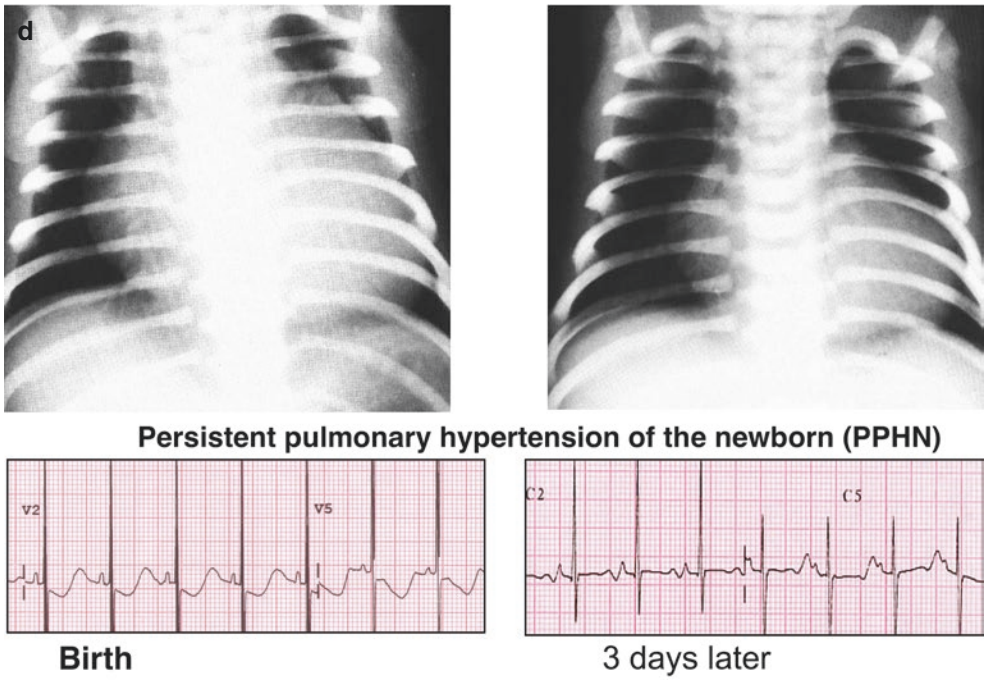

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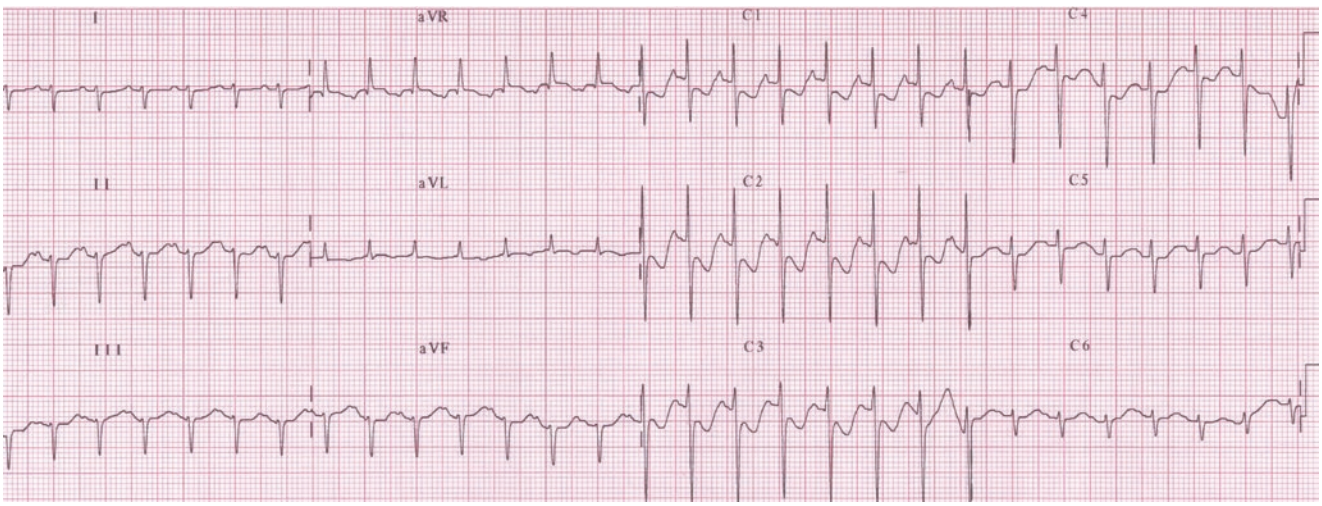

Fig. 17.2 Slightly preterm baby boy with dystocia. Initially this was believed to be a PPHN patient: history, echo, and ECG were consistent with such diagnosis. A second echo unmasked an ALCAPA. Because of the pulmonary hypertension, the ALCAPA pathophysiology had no time to develop, and the ECG is not yet typical of ALCAPA. Once again, pay attention to not miss an overlap between PPHN and ALCAPA

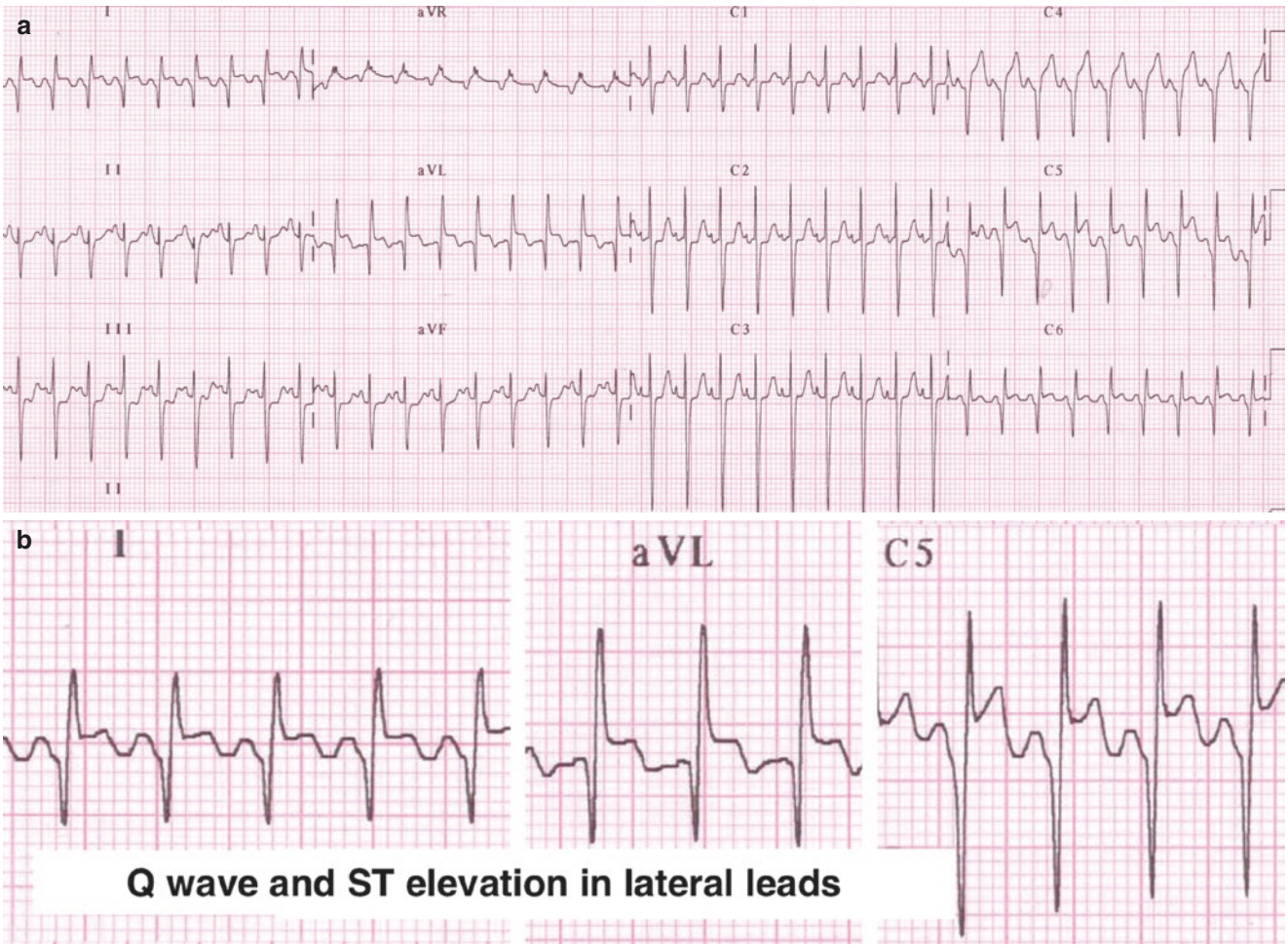

Fig. 17.3 (**a**) A 1-month-old boy with diaphragmatic hernia (DH) and ALCAPA. He underwent DH repair on the third day of life and subsequently developed heart failure. Initially the ST changes were attributed to the pulmonary hypertension typical of DH. Later, ALCAPA was suspected because the signs of ischemia worsened (Q waves and ST elevation, necrosis, and subepicardial ischemia) precisely when pulmonary pressure decreased, a behavior that makes sense in the ALCAPA pathophysiology: the decompressed pulmonary artery "steals" blood from the myocardium through the wrong left coronary artery. (**b**) Close-up, note the large Q and poor R wave

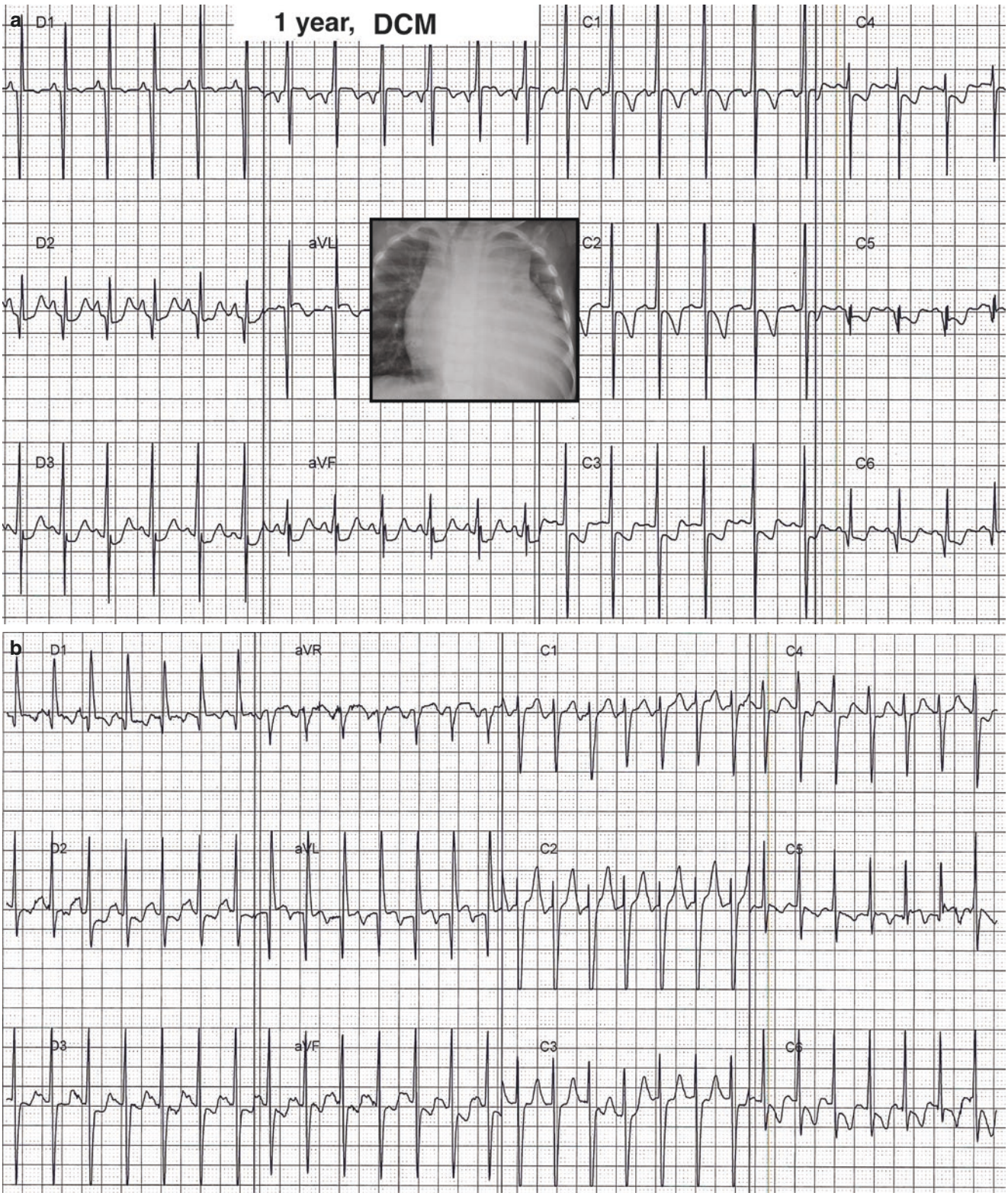

Fig. 17.4 Another two cases of ALCAPA. The two patients, both 1-year-olds, were followed as DCM patients, close to being screened for heart transplant. A more careful echo raised the suspicion of ALCAPA. (**a**) The huge Q waves in limb left lateral leads can be noted, along the widespread ST changes. This is not the classic ECG of DCM! (**b**) Again a large Q wave in aVL and widespread ST changes (courtesy of Cristina Pedron)

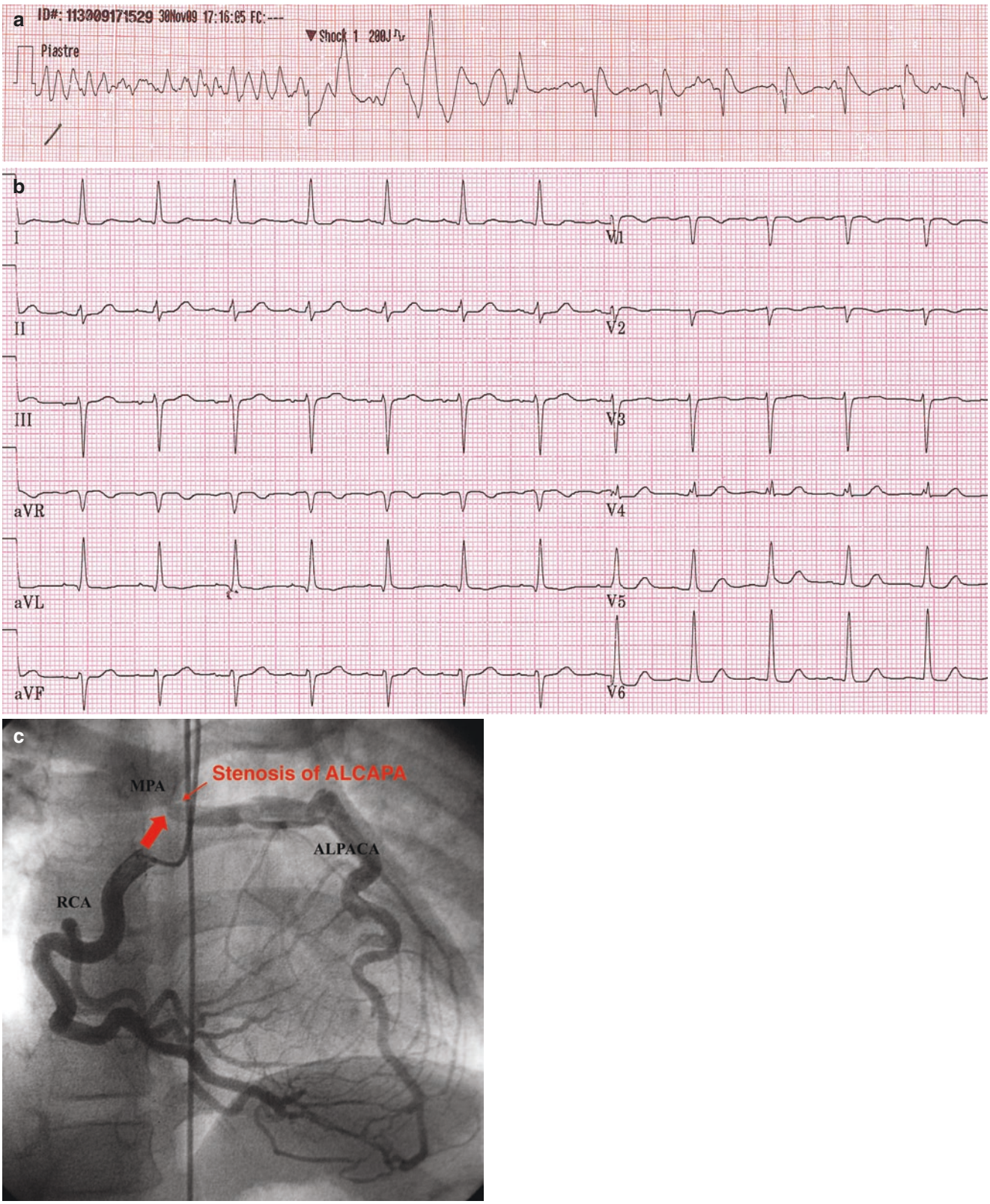

Fig. 17.5 (**a**) VF of a 32-year-old woman—mother of two children—while carrying a heavy box of apples. DC shock was effective. (**b**) The baseline ECG was not diagnostic (poor progression of R waves in the precordial leads and mild ST changes). (**c**) The diagnosis was ALCAPA. She had been protected for a long time by the proximal stenosis of the abnormal coronary artery arising from the pulmonary artery. This "pathology in the pathology" prevented the stealing of blood which is one of the mainstays in the pathophysiology of ALCAPA. Nevertheless, over the years, some silent and subtle ischemia and consequent fibrosis must have been at work, preparing the substrate for the VF

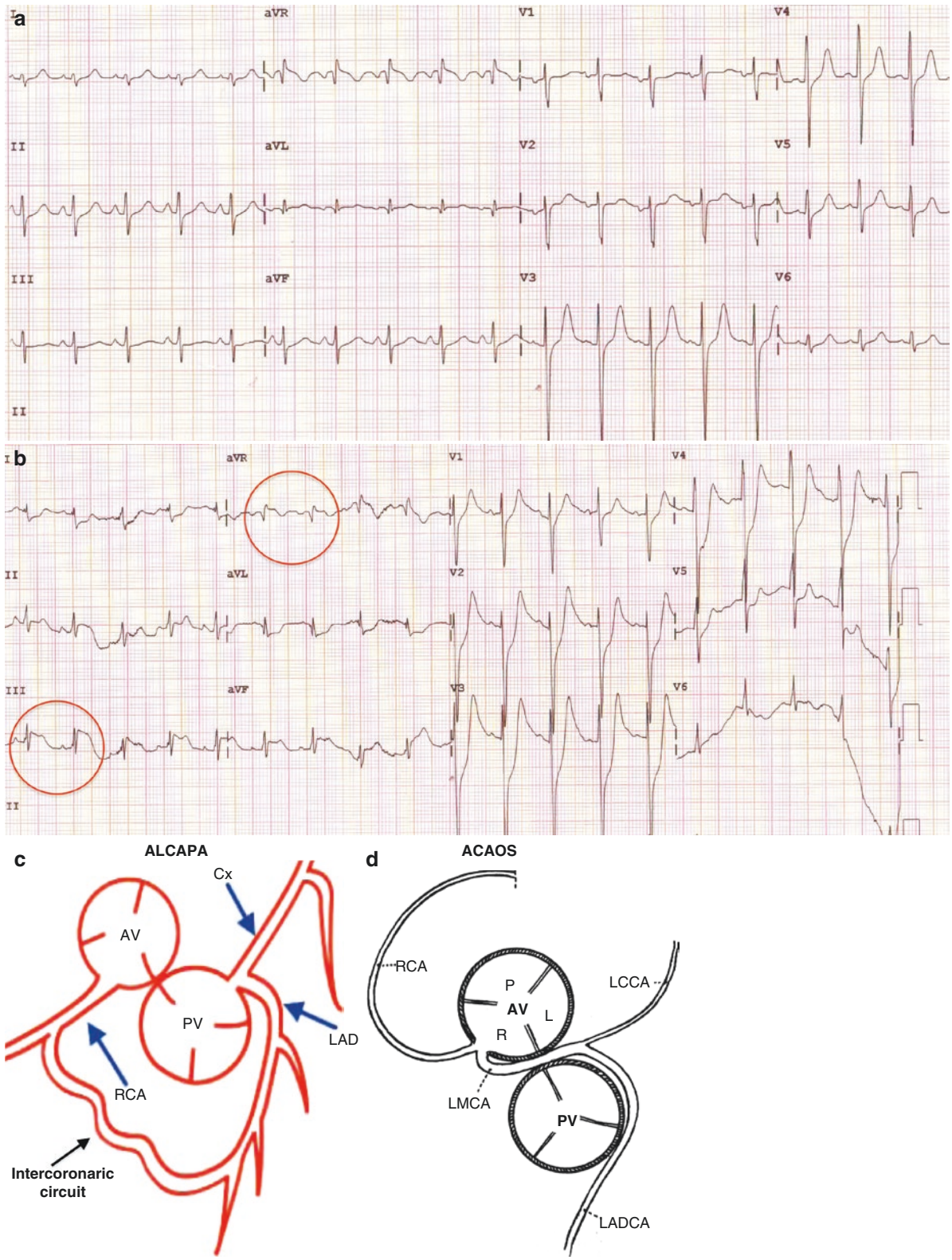

Fig. 17.6 A 13-year-old male basketball player resuscitated from VF that occurred few minutes after a match. The patient was admitted in Glasgow Coma Scale 2. (**a**) ECG on arrival, not remarkable. (**b**) 3 h later during the toilette of the tube, he presented a severe drop in blood pressure. The ECG showed marked ST change consistent with proximal coronary obstruction (left main syndrome). The echo confirmed an ACAOS. The left coronary artery was crushed between the aorta and pulmonary artery, and although it was patent on admission, during hypotension it collapsed again. (**c**) ALCAPA and ACAOS pathophysiology comparison. While in the first the ischemia is current and chronic (loss of supply and blood steal), and consequently the resting ECG is almost always abnormal, in the second the ischemia is potential and acute (left main compression), and therefore the resting ECG (and almost always the exercise test) is normal

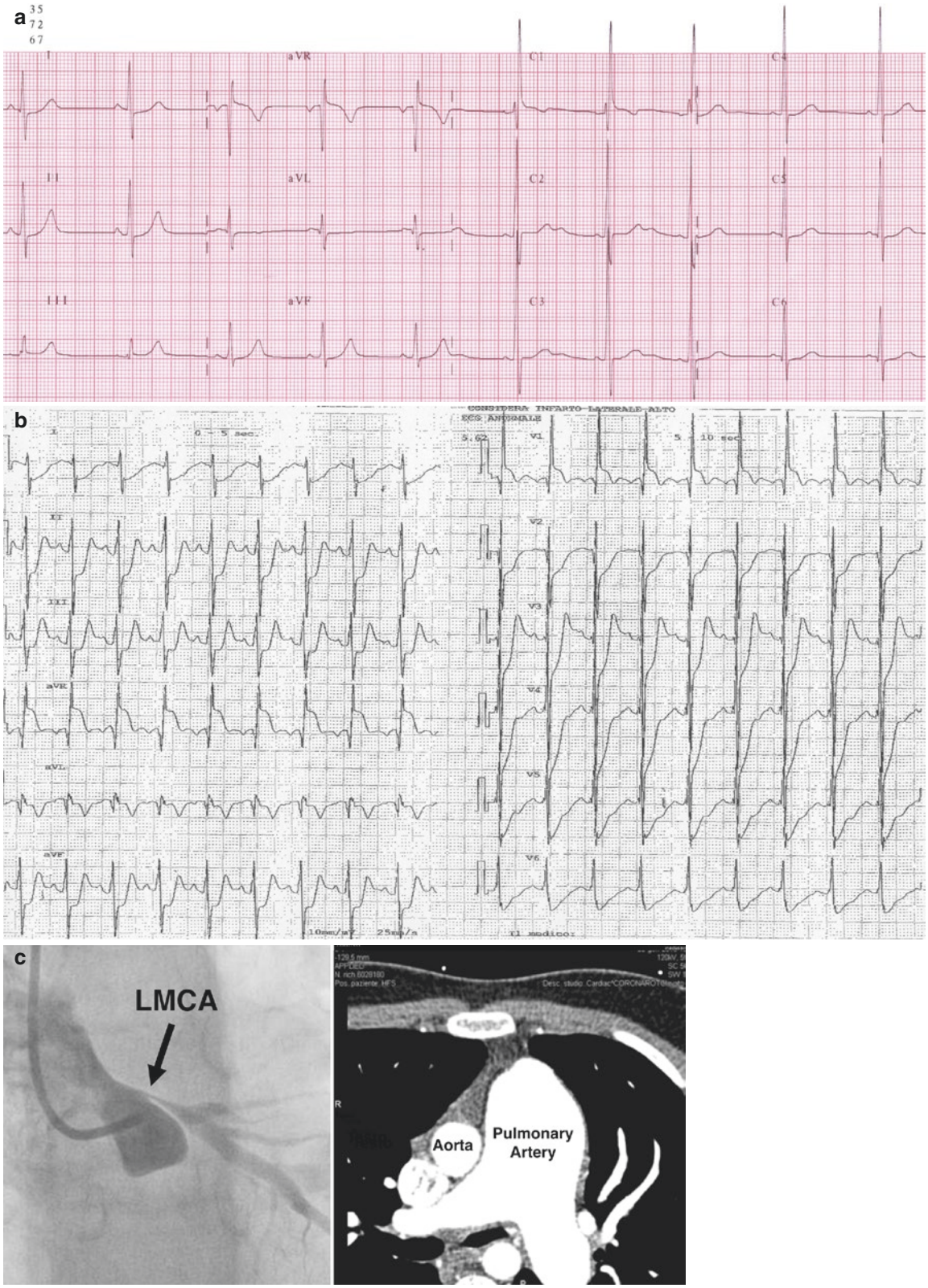

Fig. 17.7 (**a**) Resting ECG of a 14-year-old male who underwent ASD and pulmonary valve stenosis repair in infancy. There was no severe residual disease, but the pulmonary trunk was very dilated. (**b**) ECG just after a syncopal episode during exercise. Note the widespread ST depression in all leads but aVR where the ST is elevated, a picture suggestive of left main obstruction. (**c**) The ischemic event was caused by the severely enlarged pulmonary artery as proved by the coronary angiography/catheterization (*left*) and CT scan (*right*). This is a kind of coronary compression different from ACAOS. Such an ECG picture may occur also in the setting of Kawasaki disease if a thrombus occludes an aneurysm of the LMCA or LAD or in case of myocardial bridge (with or without HCM)

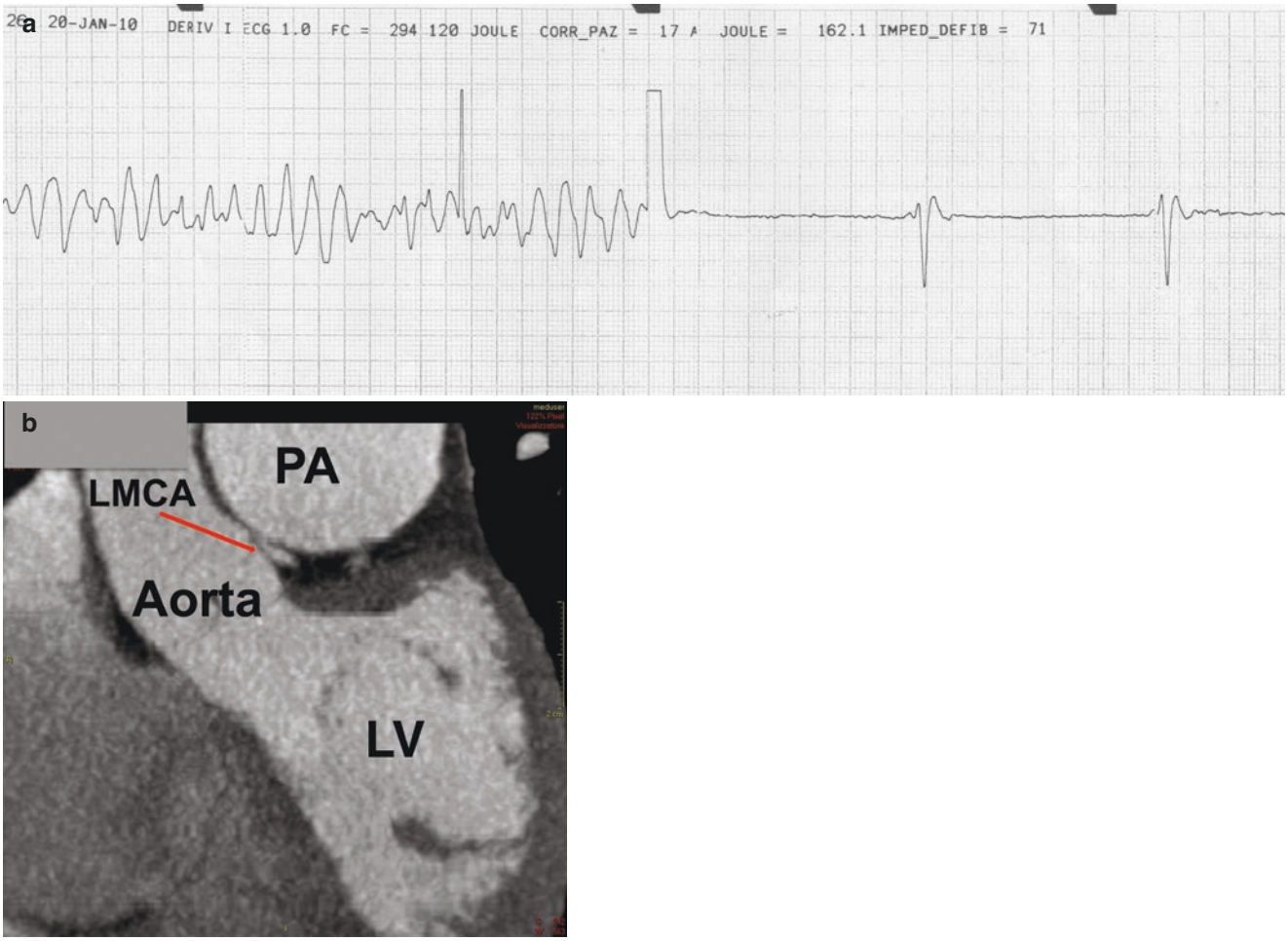

Fig. 17.8 A 13-year-old male with PPH. (**a**) VF occurred during the 6 min walking test. (**b**) Resting ECG, MRI showed the dilated pulmonary artery compressing the left main coronary artery (LMCA). (LV, left ventricle). This is another kind of coronary artery compression. The patient successfully underwent LMCA stenting

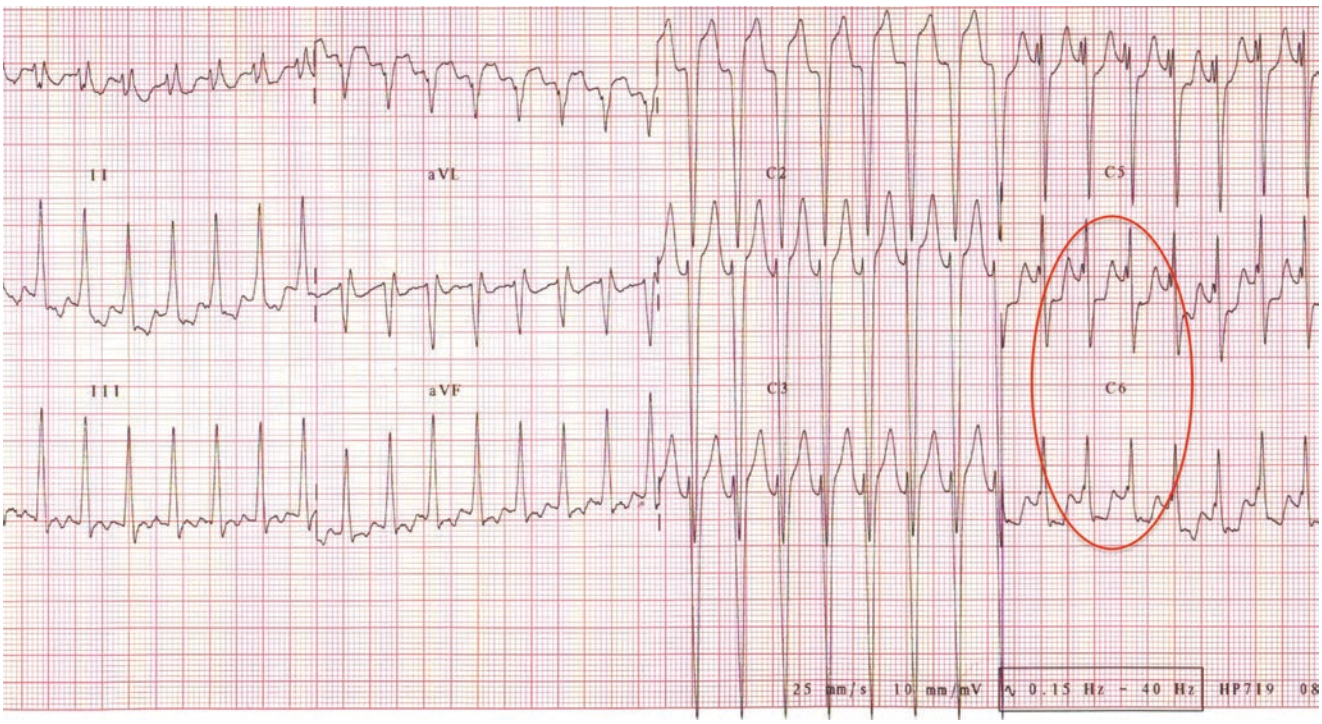

Fig. 17.9 A 15-year-old female with complex CHD and severe cyanosis (resting saturation 65%, hematocrit 66%). During a paroxysmal EAT, the fast HR (187 bpm) in addition to cyanosis, high hematocrit and low cardiac output concurred to trigger severe ischemia as can be noted from the widespread ST change

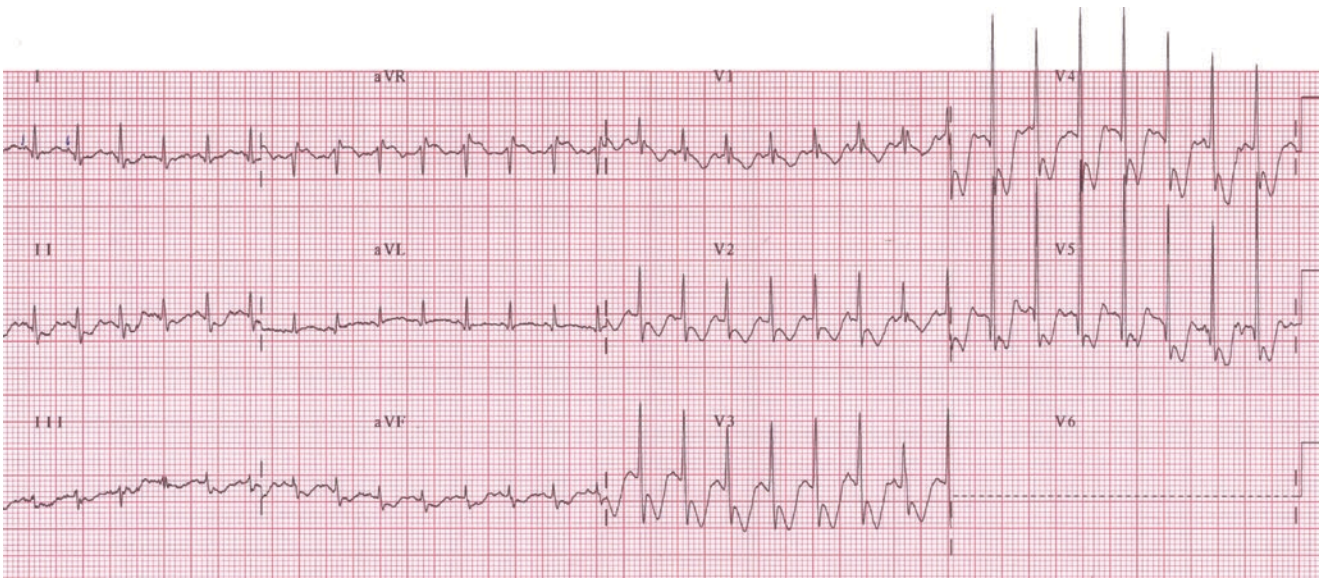

Fig. 17.10 An 8-year-old female with sickle cell anemia. ECG during chest pain. There is sinus tachycardia and precordial ST depression. The ischemia is due to vaso-occlusive crisis (a combination of abnormal hemoglobin structure and function, erythrocyte density, endothelial dysfunction, microvascular tone, inflammatory mediators, and coagulation)

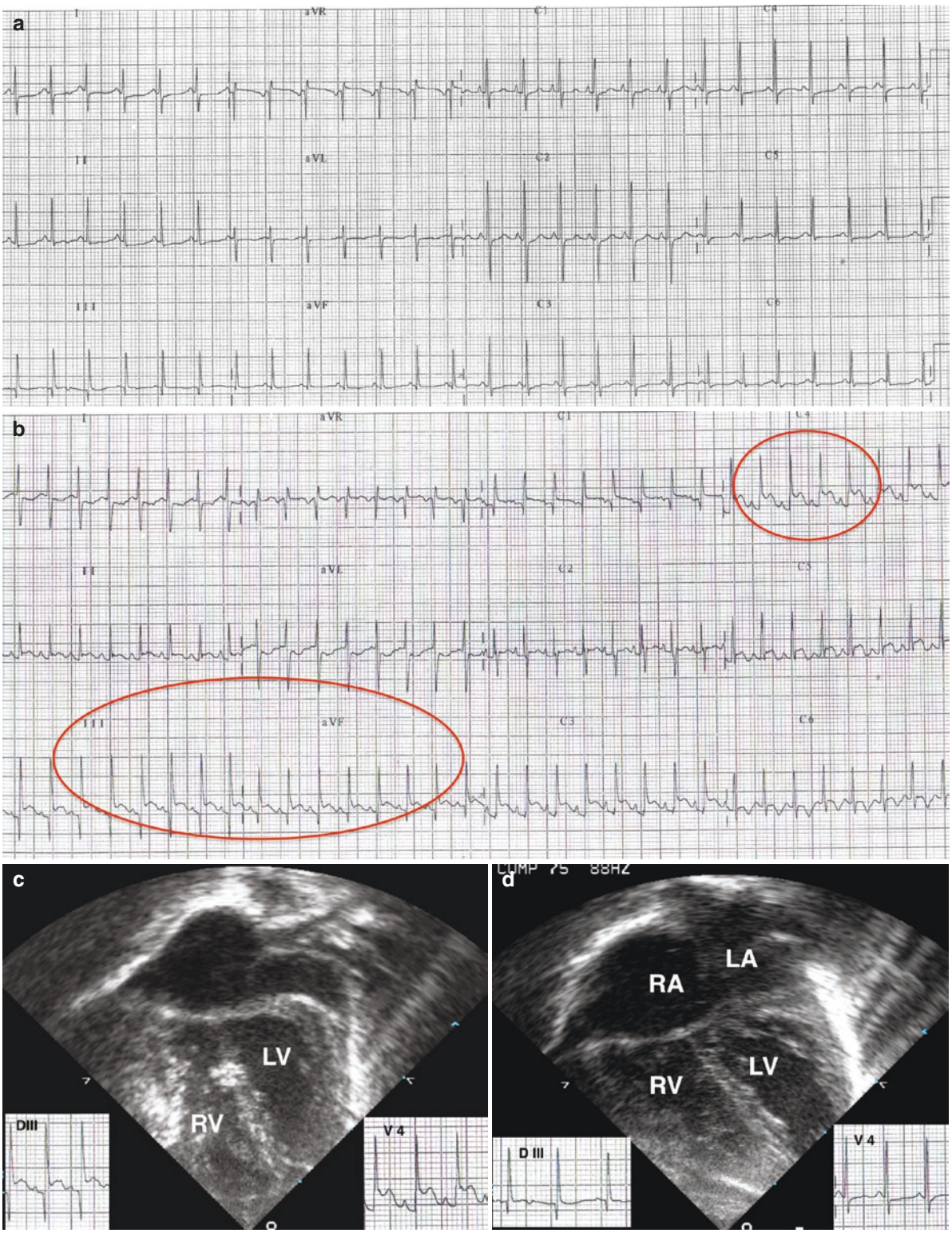

Fig. 17.11 (**a**) A 3-month-old boy with complex CHD (single ventricle heart) in the early postoperative course of palliation (Glenn's procedure). (**b**) The patient had a central venous line, and air embolism occurred. Suddenly the baby boy started to inconsolably cry. (**c**) The ST changes are concomitant with the "white" on the echo (the bright white represents the air-embolized myocardium). (**d**) A few minutes later, both ECG and echo normalized. This is an example of air embolism in the coronary arteries. It can occur during heart catheterization and in patient with venous line and right-to-left shunting

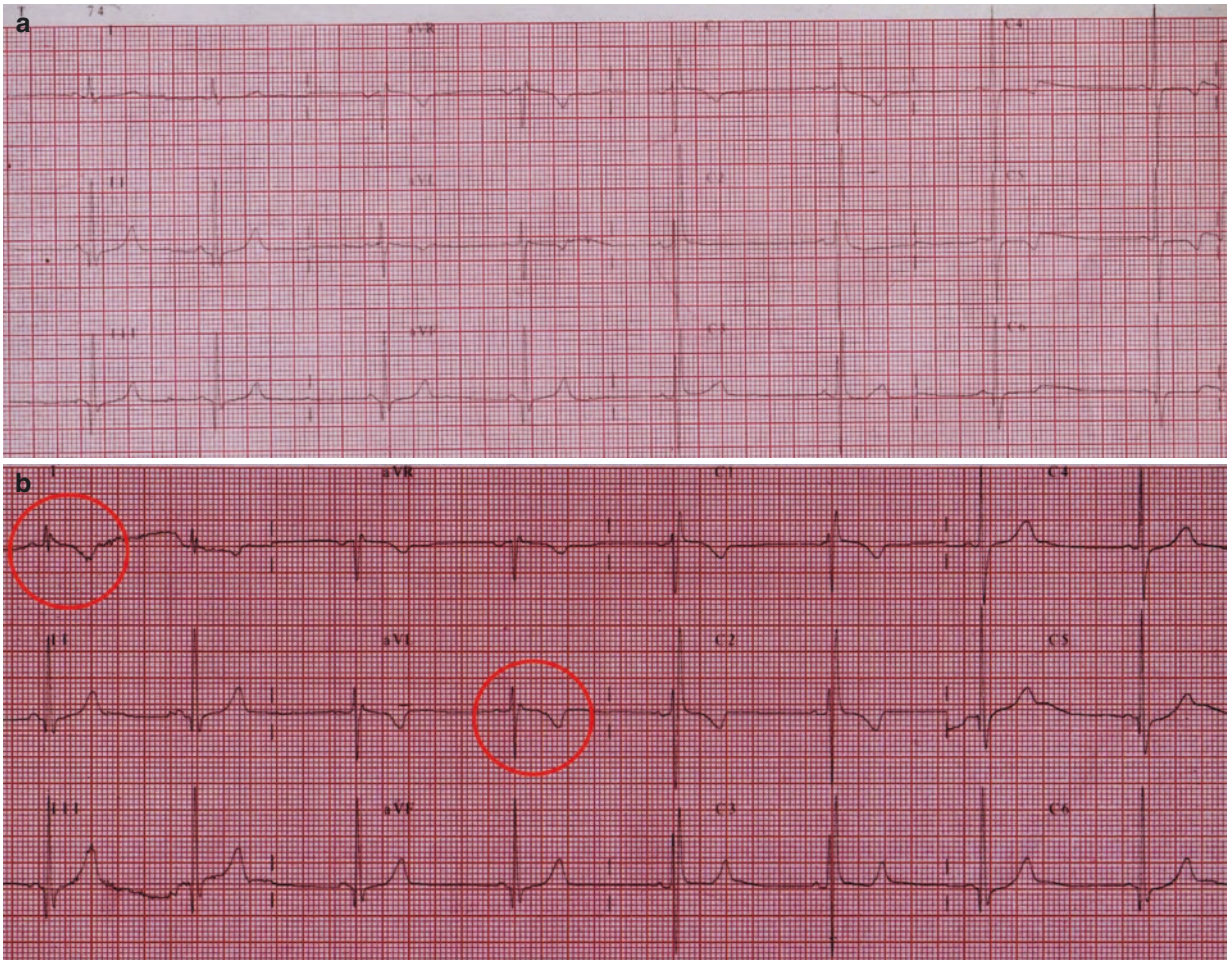

Fig. 17.12 (**a**) A 10-year-old male with TGA who underwent "switch operation" (Jatene procedure) in the first days of life. He developed a moderate stenosis of the left descending anterior coronary artery, a well-described sequelae of switch operation. In the resting ECG, there are ST changes in the precordial leads. (**b**) During the dobutamine test, although the heart rate remains unchanged, the ST segment shows ischemic change. Negative T waves in leads I and aVL along with pseudonormalization of ST changes in the precordial leads can be noted

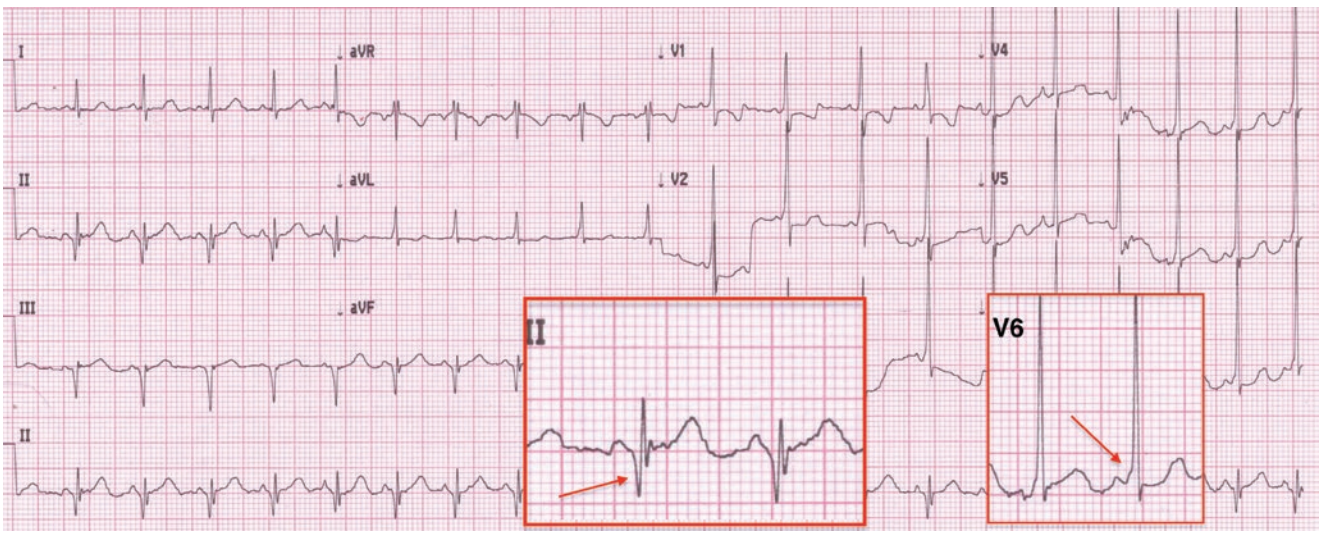

Fig. 17.13 A 3-year-old boy with ventricular pre-excitation (WPW). In the inferior leads the q waves can raise the suspicion of ischemia. The "Q" waves in the inferior leads are actually "delta waves," as precordial leads can confirm. It is important to rule out pre-excitation in the case of the presence of significant Q waves where they are expected not to be present (lead I, aVL, V1, inferior leads) and when the Q waves are absent where they are expected to be present (V6). Once WPW has been excluded, it is possible to focus the attention on ischemia (lead I, aVL), RVH (V1), and CCTGA (V6)

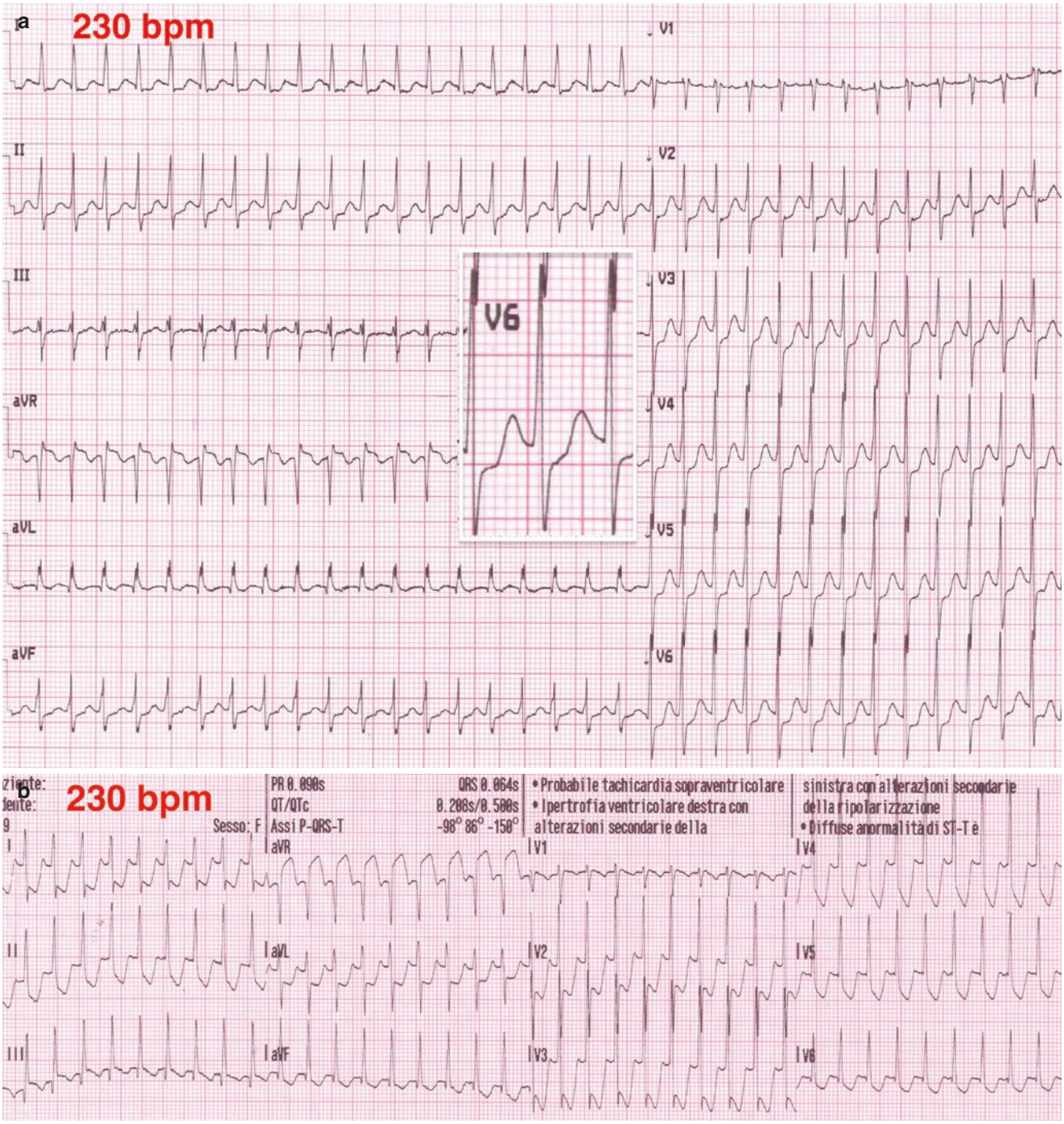

Fig. 17.14 Two females with SVT, 7 (**a**) and 9 (**b**) years old. Both SVTs are characterized by ischemic ST changes. In the second case (**b**), the patient complained of chest pain. Aspecific (nonspecific) ST–T changes are well described in SVT. Often the P buried in the S-T trait is the cause. Here it seems a real transitory subendocardial ischemia due to the fast rate, even in the absence of coronary heart disease as was proven in this case with a CT scan. Myocardial injury markers were negative, and soon after the PSVT, the ECG was normal

Suggested Reading

Justino H, Khairy P. Congenital heart disease and coronary atherosclerosis: a looming concern? Can J Cardiol. 2013;29:757–8.

Berenson GS, Srinivasan SR, Bao W, Newman WP 3rd, Tracy RE, Wattigney WA. Association between multiple cardiovascular risk factors and atherosclerosis in children and young adults. The Bogalusa heart study. N Engl J Med. 1998;338:1650–6.

Takahashi M. Cardiac ischemia in pediatric patients. Pediatr Clin N Am. 2010;57:1261–80.

Angelini P. Coronary artery anomalies. An entity in search of an identity. Circulation. 2007;115:1296–305.

Congenital Heart Disease

18

The value of electrocardiography for diagnosing congenital heart disease has decreased considerably since the introduction of fetal ultrasound as routine antenatal screening. In the past, brilliant cardiologists used to deduce the diagnosis of tetralogy of Fallot and the relative degree of oxygen desaturation by judging the S wave in lead V6. Nowadays, an accomplished gynecologist, who suddenly has lots of ultrasound equipment and no ECG experience, can grapple quite successfully with a cardiologic diagnosis. So, univentricular hearts and complex cardiac defects are now expected to be known before the ECG is performed. Nevertheless, some conditions left undetected by antenatal screening come to light in early infancy or even later in life, following a symptomatic or random ECG recording. Right ventricular hypertrophy (RVH) beyond the expected prevalence for the age, "adult" conduction intervals, and abnormal axis deviations could all suggest an underlying CHD (Tables 1.1, 3.4, 18.1, 18.2, and 18.3): left axis deviation of at least −30° is suggestive of atrioventricular canal (AVC) in a "rosy pink" newborn or toddler—which is certain in trisomy 21—or of tricuspid atresia (TA) in a cyanotic baby [1]. The ECG is quite informative in the case of complex cardiac defects and right ventricular overload, while it is much less sensitive in detecting left ventricular overload. Surprisingly, the ECG could be perfect in the transposition of the great arteries (TGA)—a CHD with an absolute ductus dependency—while an asymptomatic CCTGA may leave a number of signs on the ECG even in the neonatal period: the former has a clear clinical presentation with no need of an ECG; the latter could occasionally be encountered in an ECG recording performed due to detection of a "not-quite-innocent" heart murmur or bradycardia. Post-tricuspid shunts, such as ventricular septal defect (VSD) and patent ductus arteriosus (PDA), initially cause left ventricular overload, and, if untreated, can eventually lead to Eisenmenger syndrome with predominant right ventricular hypertrophy. Ebstein anomaly, CCTGV, and isomerism-associated arrhythmias are related to specific cardiac conduction system anomalies. Children who undergo congenital heart treatment procedures are at increased risk of various hyper- and hypokinetic postoperative "incisional" arrhythmias, e.g., atrial reentry or atypical flutter (atrial septal defect, Mustard/Senning correction for the transposition of the great vessels, Fontan procedure for univentricular heart) and ventricular tachycardia (tetralogy of Fallot) [2].

A short overview of the major congenital heart defects is presented below. Some of them, in view of a certain degree of hemodynamic compensation (patent ductus, mild oxygen desaturation or shunt), could be missed inadvertently during initial clinical evaluation or encountered by chance or following symptoms in an ECG carried out as an initial cardiac diagnostic technique.

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G. Bronzetti, *Atlas of Pediatric and Youth ECG*, https://doi.org/10.1007/978-3-319-57102-7_18

Table 18.1 ECG synopsis of all CHD

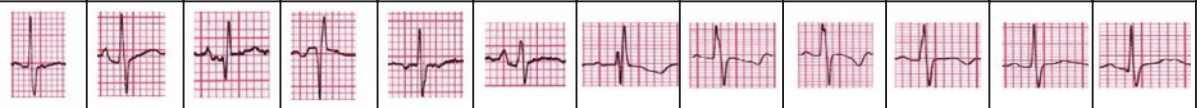
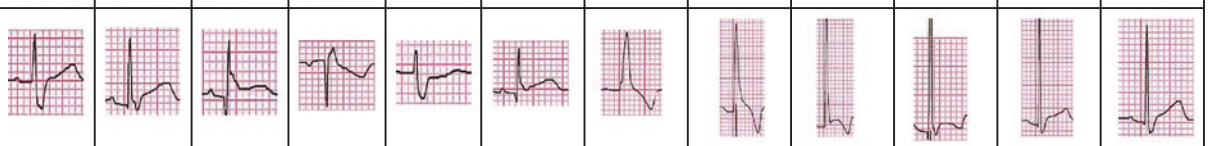
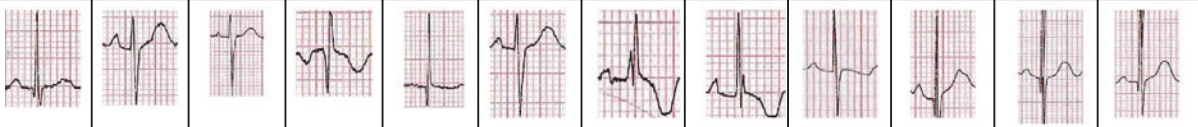
	I	II	III	aVR	aVL	aVF	V1	V2	V3	V4	V5	V6
ASD												
VSD												
PDA												
AVSD												
PoS												

Table 18.2 ECG synopsis of all CHD

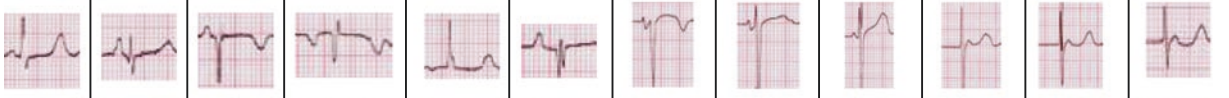
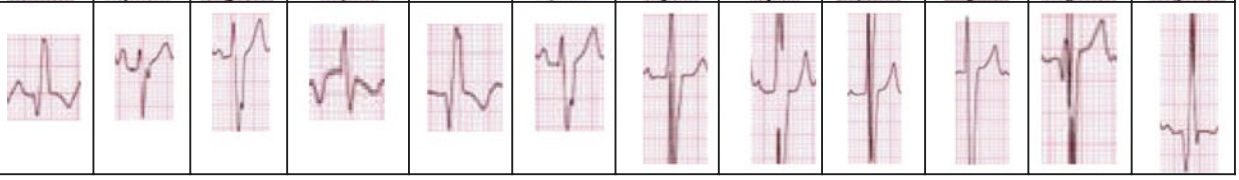
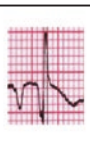
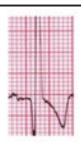
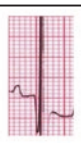
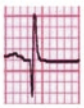
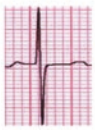
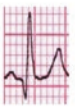
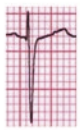
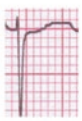
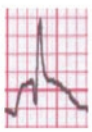
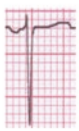
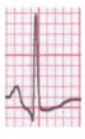
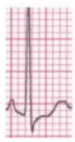
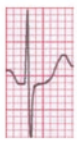
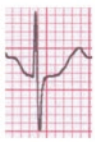
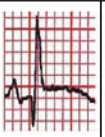
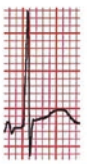
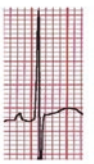
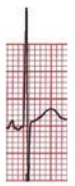
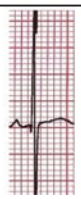
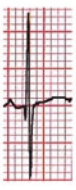
	I	II	III	aVR	aVL	aVF	V1	V2	V3	V4	V5	V6
Eb A												
CC TGA												
TA												
PAIS												
ALC APA												

Table 18.3 ECG synopsis of all CHD

	I	II	DIII	aVR	aVL	aVF	V1	V2	V3	V4	V5	V6
AoS												
Ao Co												
ToF												
TGA												
PA+ VSD												

18.1 Cardiac Defects with Left-to-Right Pre-tricuspid Shunts (Atrial Septal Defects and Anomalous Pulmonary Venous Return)

In patients with small atrial septal defects (ASD) and no evidence of significant right-sided chamber dilatation, the ECG is usually normal, unless it is an *ostium primum* associated with a partial AV canal (left axis deviation in *ostium primum* vs right axis deviation in *ostium secundum*). With a significant left-to-right shunt, the ECG changes are proportionate to the degree of the right ventricle dilatation [3].

The *rsr'* or *rsR'* QRS morphology in the right precordial leads is a frequent finding, with a sluggish and polyphasic aspect of the r' wave, secondary to right ventricle volume overload and dilatation (unfortunately rsR pattern is a not so rare finding even in young with normal heart, but the R wave amplitude counts). Starting from late childhood, an indented aspect ("crochetage") of the R wave could be observed in the inferior leads [4]. The above electrocardiographic abnormalities are reversible, with a possible normalization of the ECG immediately after surgical closure or transcatheter occlusion with devices.

The QRS axis is usually vertical, but in the presence of significant blood shunting and elevated right ventricular systolic pressure, a rightward QRS axis and some right ventricular conduction delay (RBBB) are frequently observed. Atrial fibrillation/atypical atrial flutter and IART secondary to atrial enlargement and fibrosis may complicate the clinical picture after the second decade of life.

Adult patients with coexisting cardiovascular problems (systemic arterial hypertension, heart valve disorders, fascicular blocks) may exhibit a left QRS axis deviation, which is also characteristic of a complete or partial atrioventricular canal defect (AVC) and Holt-Oram syndrome. In major shunts, the ventricular repolarization pattern could be modified to the same extent as depolarization: as previously mentioned, negative-positive diphasism of the *T* wave (overshoot) in the right precordial leads is suggestive of right ventricular volume overload ("butterfly sign"). The *PR* interval of a classic *ostium secundum* ASD is normal, while its prolongation and first-degree heart block may occur in ASD *ostium primum* or *sinus venosus type*. The ECG changes in patients with *anomalous pulmonary venous return*, and dilated right chambers correspond to those outlined above for ASDs. On some occasions, these patients may suffer from sinus bradycardia and ectopic atrial rhythms: in fact, *anomalous pulmonary venous return* may be coupled with anomalous systemic venous drainage, such as a left-sided superior vena cava

(LSVC) which drains into the right atrium via the coronary sinus. Both venous anomalies may cause low atrial rhythm or ectopic atrial rhythms. Sinus node dysfunction is a common postoperative complication, its frequency being inversely related to the child's age at the time of corrective surgery. AV conduction disorders which may progress to a complete heart block are also reported in rare cases of ASD repair (e.g., sinus venosus).

18.2 Cardiac Defects with Left-to-Right Post-tricuspid Shunts (Ventricular Septal Defect and Patent Ductus Arteriosus)

As mentioned earlier for ASD, the ECG is usually normal in the case of minor ventricular septal defects (VSD) with no evidence of left chamber dilatation. With conspicuous right-to-left shunts, the ECG changes are secondary to left ventricular dilatation and hypertrophy, left atrial enlargement, and/or right ventricular pressure overload, which develop due to pulmonary arterial hypertension or infundibular pulmonary stenosis. Toddlers with a large VSD or PDA exhibit biventricular hypertrophy and equiphasic QRS complexes in the mid-precordium, known as the Katz-Wachtel phenomenon [5]. Older patients can satisfy voltage criteria for left ventricular hypertrophy. Left atrial hypertrophy or prolonged atrial depolarization and left axis deviation are often present as well. Right ventricular pressure overload is associated with pulmonary arterial hypertension or infundibular pulmonary stenosis. Most patients who have undergone repair of a subaortic ventricular septal defect develop a postoperative RBBB (much less often an LBBB which is, instead, a complication of myomectomy according to Morrow or correction of a subaortic stenosis).

18.3 Partial, Intermediate, and Complete Atrioventricular Canal

The ECG patterns ranging from left axis deviation to left anterior hemiblock reflect the underlying particular anatomy of the conduction system (the inferior displacement of the AV node and the His bundle, hypoplastic anterior fascicle of the left bundle) and the right and/or left ventricular dilatation due to significant shunting [6]. In patients with a partial (i.e., *ostium primum* ASD with mitral valve anomaly) or intermediate (with an associated restrictive type VSD) AV canal, apart from the left anterior fascicular block, an RBBB can be especially observed in the case of a conspicuous left-to-right shunting with right ventricular dilatation. Moreover, in patients with a complete AV canal (i.e., *ostium primum* ASD and a large VSD) and left-to-right shunting with biventricular dilatation (often aggravated by atrioventricular valve incompetence), a left ventricular hypertrophy or biventricular hypertrophy can complicate the above-described ECG pattern. A first-degree AV block is a frequent finding, while an advanced or complete heart block might represent a postoperative complication, especially in large inlet VSDs.

18.4 Right-Sided Obstructive Lesions (Pulmonary Stenosis, Tetralogy of Fallot, Pulmonary Atresia with VSD, and Pulmonary Atresia with Intact Ventricular Septum)

In patients with pulmonary stenosis, the ECG changes reflect the degree of right ventricular hypertrophy. The amplitude of the *R* wave in lead V1 has demonstrated a good correlation with the pressure gradient across the pulmonary valve [7]. Common electrocardiographic indications that are used in diagnosing right ventricular hypertrophy are a high and monophasic *R* wave in V1, rightward deviation of the mean QRS axis in the frontal plane, deep *q* waves in the inferior leads, and deep *S* waves in V5–V6. Upright *T* waves in V1 are associated with increased right ventricular pressure in children aged 7 days to 7 years of life. Right atrial enlargement, if present, represents an additional diagnostic tool for right ventricular hypertrophy. Extreme axis deviation, also known as "northwest axis," is a characteristic finding, which is very frequent in patients with Noonan syndrome, even in the absence of significant pulmonary stenosis. There are no specific ECG changes for diagnosing tetralogy of Fallot, although the above signs of right ventricular hypertrophy are usually present. Moreover, a correlation between the degree of cyanosis and *R/S* voltage ratio in left precordial leads has been described: in "rosy pink" Fallot infants, the *R/S* voltage ratio is normal for age, while in a "cyanotic" Fallot, deep *S* waves can be observed in the left precordial leads with an *r/S* ratio < 1.

In pulmonary atresia with VSD, the ECG changes are indistinguishable from Fallot tetralogy. In the case of a large VSD, right ventricular pressures are usually equal than their systemic equivalents. On the contrary, in pulmonary atresia with intact ventricular septum, the right-sided pressures are higher than the left-sided ones, and the electric representation of the right ventricle has an inverse correlation with its degree of hypoplasia. In this case, the congenital anomaly affects the right heart as a whole, and the atretic pulmonary valve may produce no ECG changes, in view of the absent electrical predominance of the hypoplastic right ventricle which, nevertheless, is subjected to a consistent pressure overload. Right atrial enlargement, as a consequence of severe or massive tricuspid insufficiency, will guide the diagnosis.

In patients with tetralogy of Fallot who undergo corrective surgery, an RBBB is a common, almost invariable postoperative complication, usually with a quite wide QRS complex. The QRS duration may increase with time, in parallel with right ventricular dilatation secondary to pulmonary insufficiency, and represents a risk factor for ventricular arrhythmias (significant when QRS is longer than 180 ms).

18.5 Left-Sided Obstructive Lesions (Aortic Stenosis and Coarctation of the Aorta)

In young adults, the ECG sensitivity for diagnosing left ventricular hypertrophy is low, even in the presence of a severe aortic stenosis or coarctation of the aorta, while during the neonatal period and in early infancy, it is much more accurate and useful.

QRS axis deviation and prominent *Q* waves are some of the known ECG signs denoting left ventricular hypertrophy of at least moderate degree, while high voltage in the left leads is a more sensitive but less specific criterion [8]. In the neonatal coarctation of the aorta, the ECG signs of right ventricular overload may precede those of left overload. In terms of overload, generally the repolarization alterations are more specific and sensitive than depolarization ones.

18.6 Tricuspid Atresia

TA is the most common form of univentricular heart, actually the prototype of left univentricular heart. In the absence of a tricuspid valvular plane, the conducting system dislocates in the posterior and inferior direction (the AV node and its branches), similar to what happens in AV canal defect. A left axis deviation or a left fascicular hemiblock is thus typical ECG finding in these patients, with a quite certain diagnosis in a cyanotic infant.

18.7 Tricuspid Valve Dysplasia and Ebstein's Anomaly

The ECG usually shows right atrial involvement with prolonged *PR* interval and unusual *RBBB*-polyphasic QRS complexes in lead V1. Right atrial involvement and PR interval prolongation are both consequences of atrial dilatation, with a subsequent atrial conduction delay. Polyphasic RBBB is due to a ventricular conduction delay in view of the "atrialization" of the upper segment of the right ventricle [9]. Negative *T* waves in the right precordial leads and prominent *Q* waves in the inferior quadrants are common. The latter is a consequence of right ventricular wall and/or septum thickening and fibrotic changes [10]. Additionally, there is an association with ventricular preexcitation in up to 25% of cases due to the presence of accessory pathways like Kent or Mahaim, often multiple and only exceptionally left-sided [11]. These patients do not usually show a characteristic polyphasic QRS complex in V1 but a short PR interval with a delta wave. In Mahaim-type accessory pathway, a normal or prolonged PR interval with an initial slurring of the QRS complex and anterior pseudonecrosis (poor R wave progression in the anterior leads) may be observed.

18.8 Transposition of the Great Arteries (TGA)

The ECG at birth may be completely normal in the event of a simple transposition of the great vessels, without an associated VSD, pulmonary stenosis, or coarctation of the aorta. It may sometimes display right ventricular overload (as the aorta arises from the right ventricle), aggravated by systemic vascular resistance and pressure.

18.9 Congenitally Corrected Transposition of the Great Arteries (CCTGA)

Due to the mirror-imaged arrangement of the ventricles that we could simplify by calling it ventricular inversion, the activation of the interventricular septum will proceed from right to left and not, as is usually the case, from left to right. Consequently, the ECG assumes an aspect of anterior pseudonecrosis with Q waves present in the right precordial leads and absent in the lateral leads [12]. In the frontal plane left axis deviation can be frequent and positive anterior T waves are also present. In addition, since the interventricular septum activation progresses from the bottom up, there are usually deep *q* waves in the inferior leads as well, most evident in lead III.

A high-grade AV block is common in these patients, already present at birth or with later presentation. Estimates foresee an annual incidence of about 2% and a prevalence in adults of about 22% [13]. Arrhythmias and conduction disorders are due to the anterior dislocation of the AV node and the bundle of His, the course of the latter usually being oblong and disrupted, often running close to a VSD border (thus, a frequent postoperative AV block); the natural history of CCTGV is also characterized by a frequent histopathologic finding of a fibrotic bundle of His [14]. The morphologically right ventricle in CCTGV has a tricuspid valve that is often dysplastic even to the degree of Ebstein's anomaly; the latter two cardiac defects could harbor accessory pathways and have a similar ECG expression both in terms of pre-excitation (accessory pathways) and post-excitation (conducting disturbances), similarly to storage disease as Fabry's disease and PRKAG2 mutation.

18.10 Congenital Coronary Artery Anomalies

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is the most common anomaly of the coronary origin. Right coronary artery anomalous origin from the pulmonary artery (ARCAPA) is a possible, but usually asymptomatic, condition in view of the minor hemodynamic significance of its blood supply compared with the left coronary artery. ALCAPA becomes symptomatic from the very first months of life, presenting on ECG

with anterior and/or lateral wall necrosis with associated abnormal deep and wide *q* waves: as pulmonary arterial pressure drops soon after birth, the flow in the left coronary artery decreases and then reverses, resulting in the "coronary steal" phenomenon, in which a left-to-right shunt leads to abnormal left ventricular perfusion and ischemia. The extent of acquired collateral circulation between the two main coronary arteries (the right one is usually dilated!) during the critical period determines the extent of myocardial ischemia. In the rare cases of additional anatomical defects—i.e., stenotic origin of the left coronary artery—the above mechanism could be limited, causing mild symptoms and normal ECGs up to adult age. So, like ACAOS and coronary artery fistulae, ALCAPA and other coronary artery anomalies should be kept in mind for differential diagnosis in case of syncope or major arrhythmias precipitated by emotional or physical stress [15, 16]. However, ACAOS differs significantly from ALCAPA. The basic electrocardiogram in ACAOS is, in fact, always normal, as is the stress electrocardiogram. Symptoms occur on exertion, a situation in which the coronary artery that has an abnormal course between the aorta and the pulmonary artery can be crushed. The diagnosis of ACAOS is therefore eminently echocardiographic.

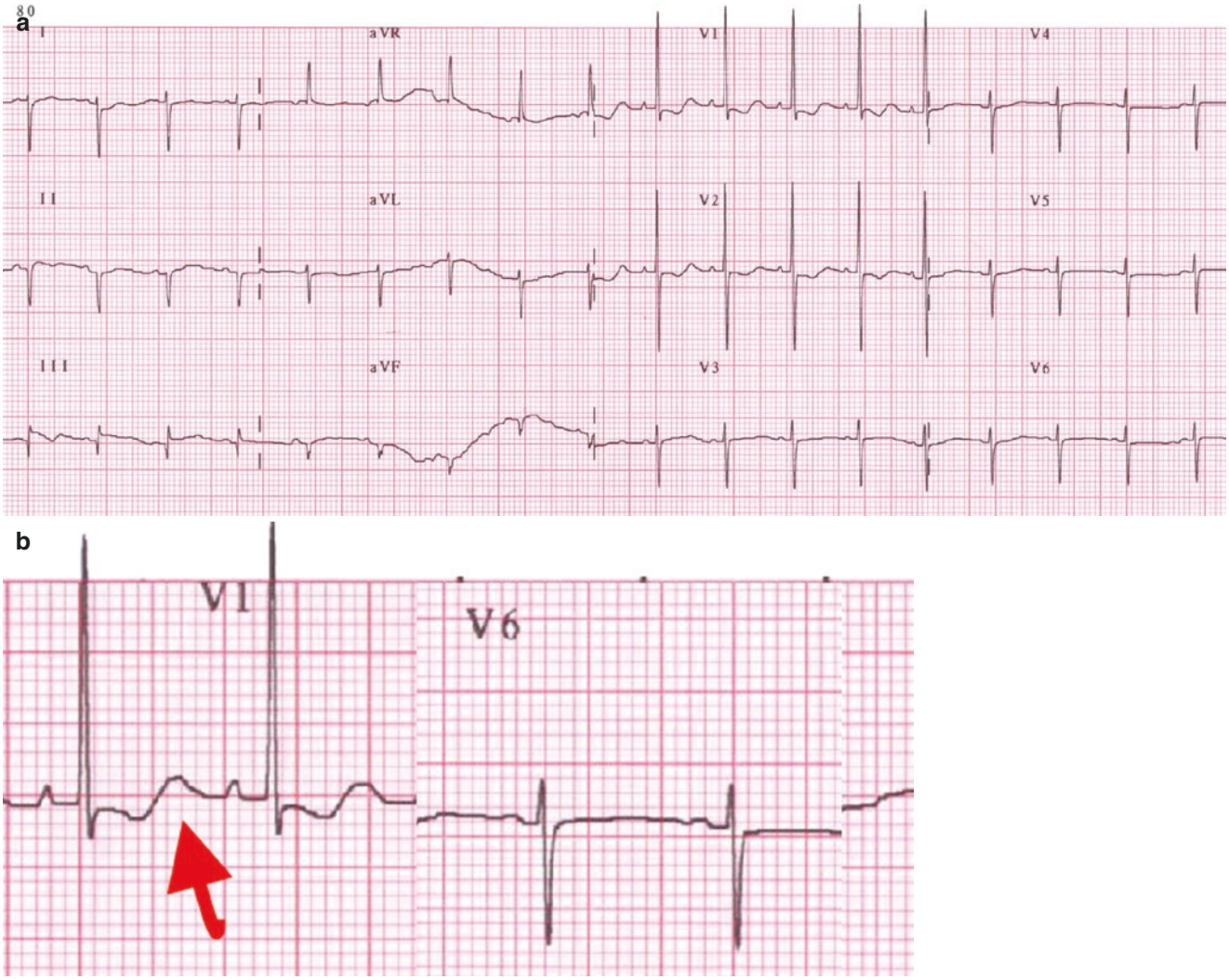


Fig. 18.1 (**a**) Ten-day-old baby girl. Sinus rhythm, P wave does not suggest overload. The QRS shows right axis deviation, standing at about 200°. The R wave in V1 is at the upper limit but it is almost monophasic, and S V6 are at the upper limit too, not frankly pathological. Left voltages are not much represented, a normal pattern for a neonate. (**b**) Close-up. The most interesting aspect of the repolarization is in V1: the T wave is diphasic, with prevalence of the positive terminal component. After the first week of life and up to 7 years of age, the positive T in V1 should be regarded as systolic right ventricular overload. The patient had moderate valvular pulmonary stenosis (PS)

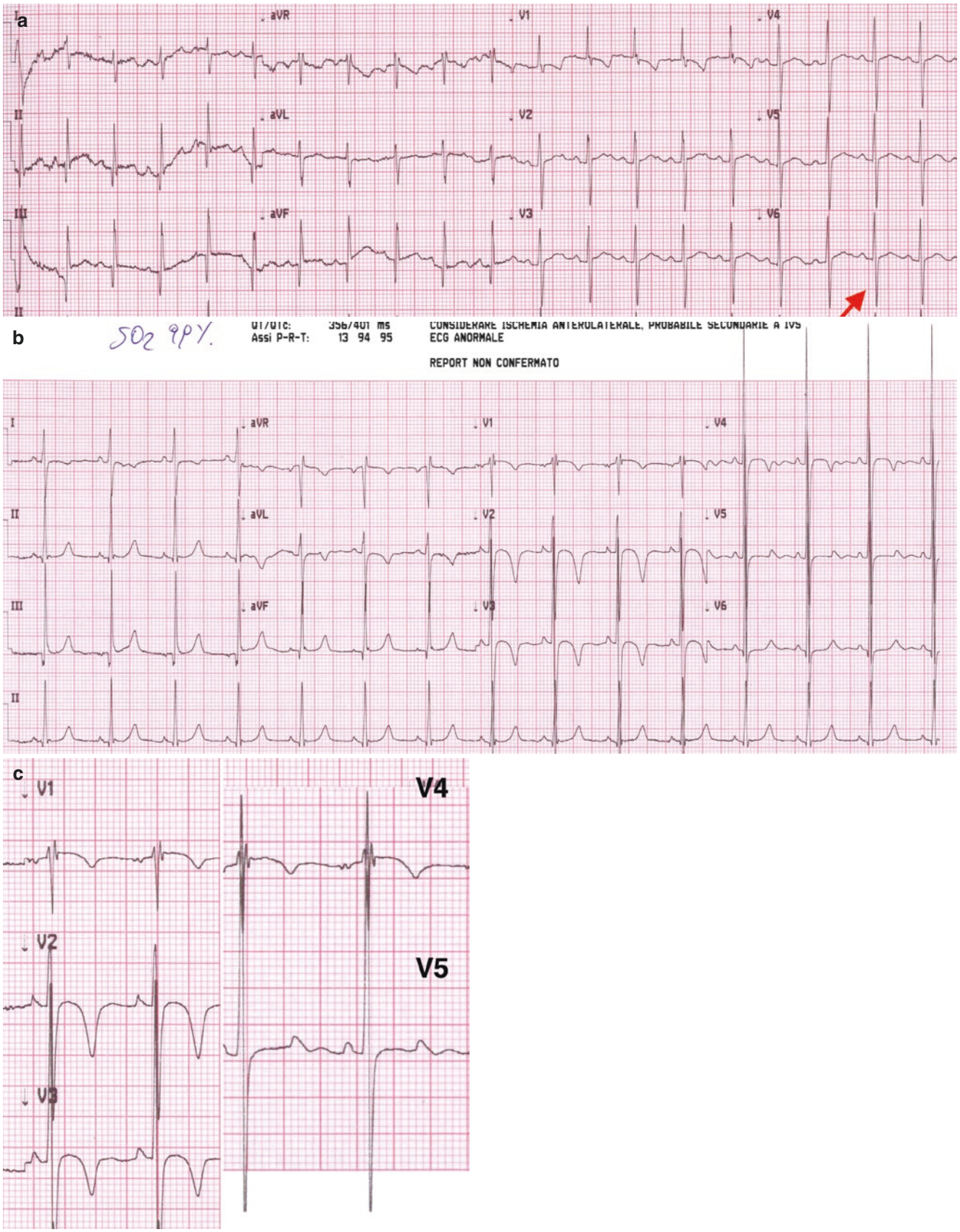


Fig. 18.2 (**a**) Two-year-old male with moderate PS. (**b**) Similar case, 6-year-old male with moderate PS. On echo the gradient through the pulmonary valve was 70 mmHg. The right axis deviation is not impressive, 100°, as well as the R wave in V1. In both cases, the deep S wave in V6 reveals the pressure overload of the right ventricle despite the silence of the other leads. (**c**) Close-up

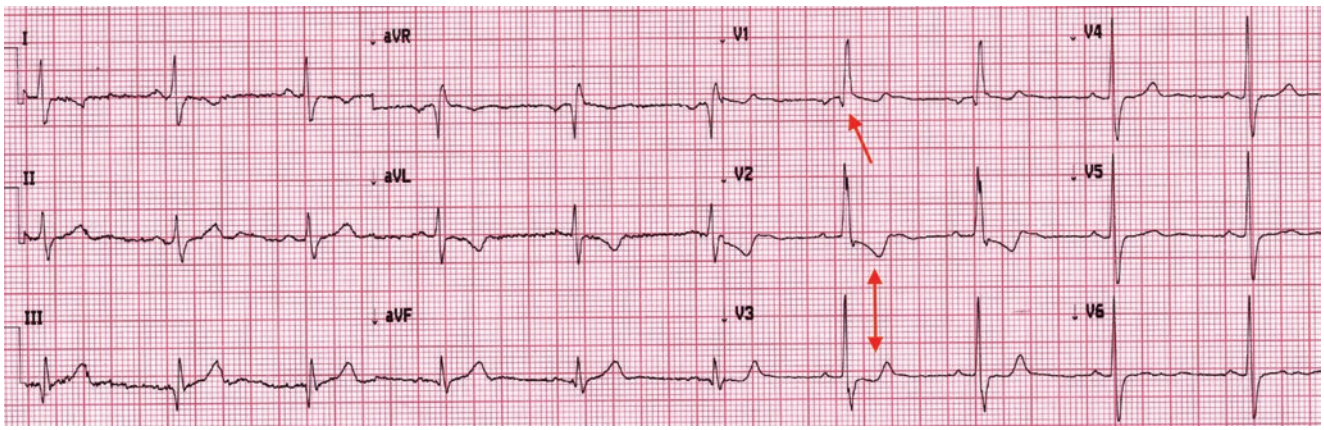


Fig. 18.3 Thirty-year-old female with severe PS. Note the qR in V1, a specific sign of severe RVH, along with ST depression

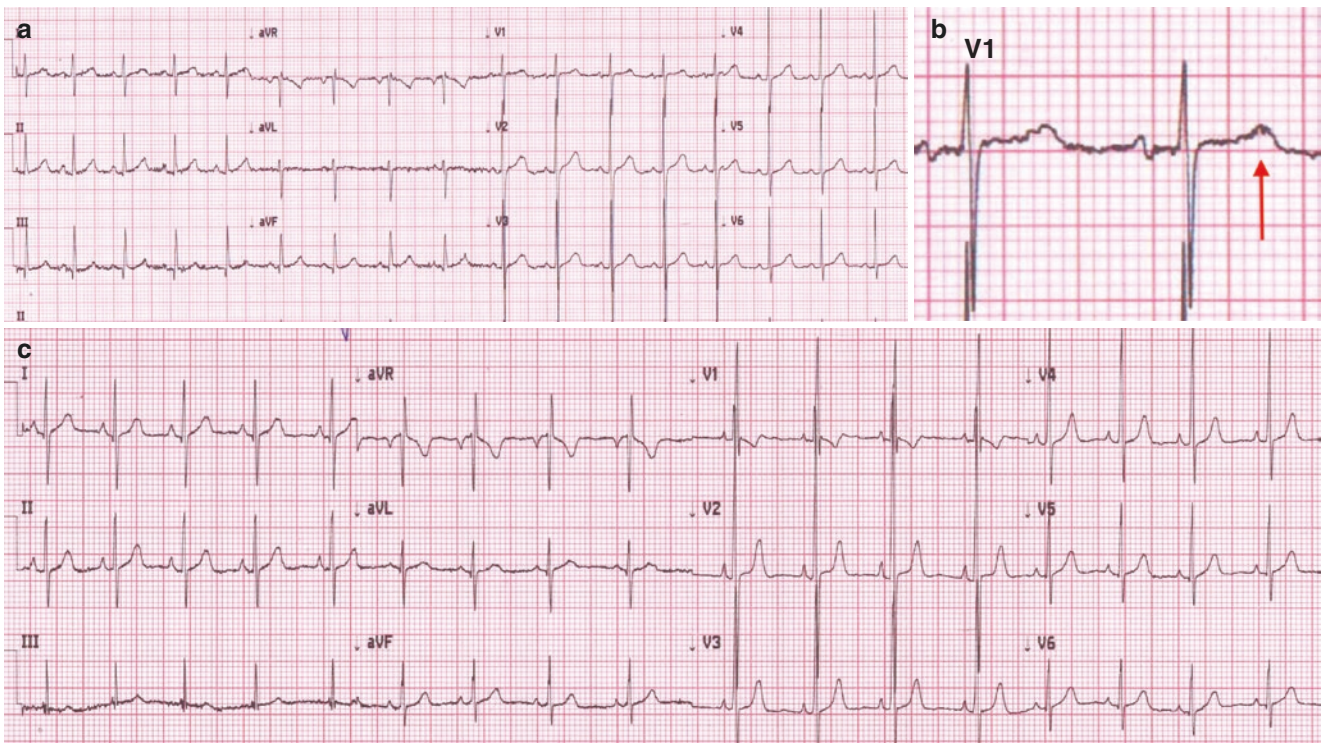


Fig. 18.4 (**a**) Six-month-old male with VSD who underwent banding of the pulmonary artery. (**b**) Despite the severe RVH (RV systolic pressure being 110 mmHg), the only sign of right pressure overload is the positive T in V1. (**c**) Eight-month-old female with Alagille syndrome and moderate stenosis of the right and left main pulmonary branches (RSVP 70): in V1 note the monophasic tall R wave and the biphasic T wave. *From 7 days to 7 years, the T in V1 never lies*

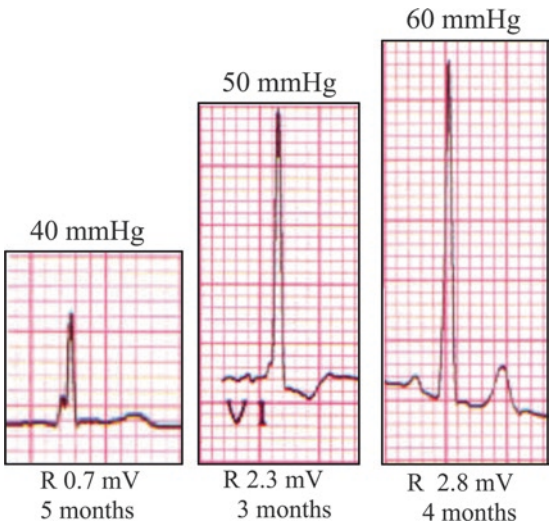


Fig. 18.5 Correlation between transpulmonary valve gradient during catheterization and ECG. There is a good correlation between the amplitude of the R wave in V1 and the degree of PS, especially for significant stenosis. For mild stenosis, the T wave is more eloquent, positive in V1 when it should be negative. In severe forms, Q waves in V1 can be found, always pathological, or marked repolarization abnormalities like ST depression

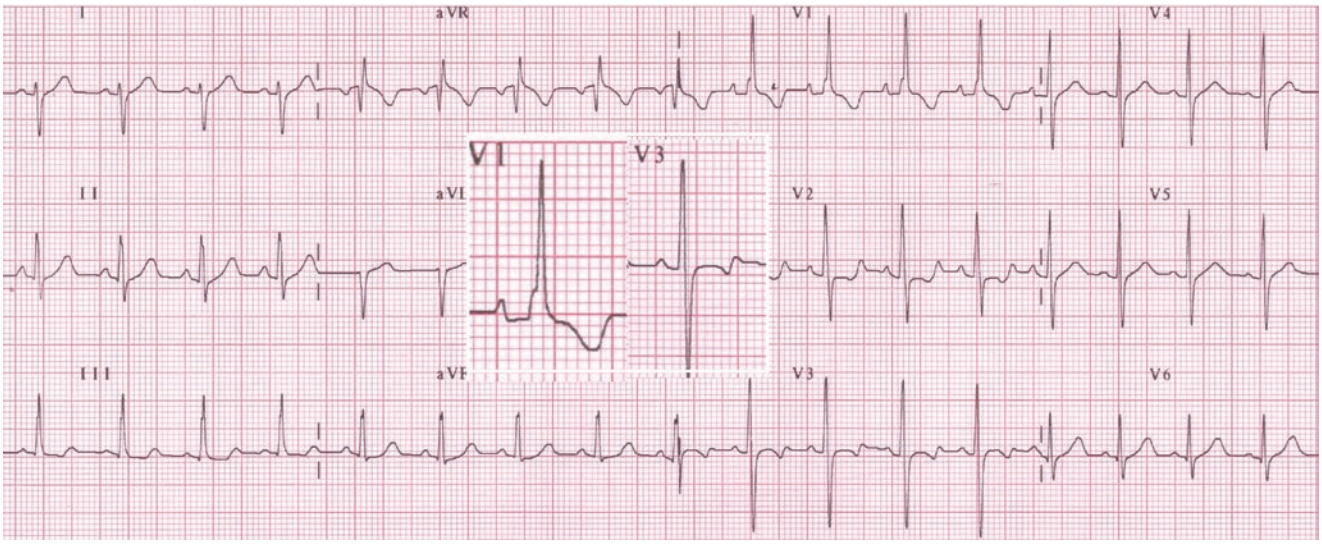


Fig. 18.6 Seven-year-old male with moderate ASD type II (*ostium secundum*). There is a tall, monophasic, and initially slurred R wave in V1. In V3 an "overshoot" pattern can be seen (inverted T wave showing terminal positivity), a pattern described in RV overload

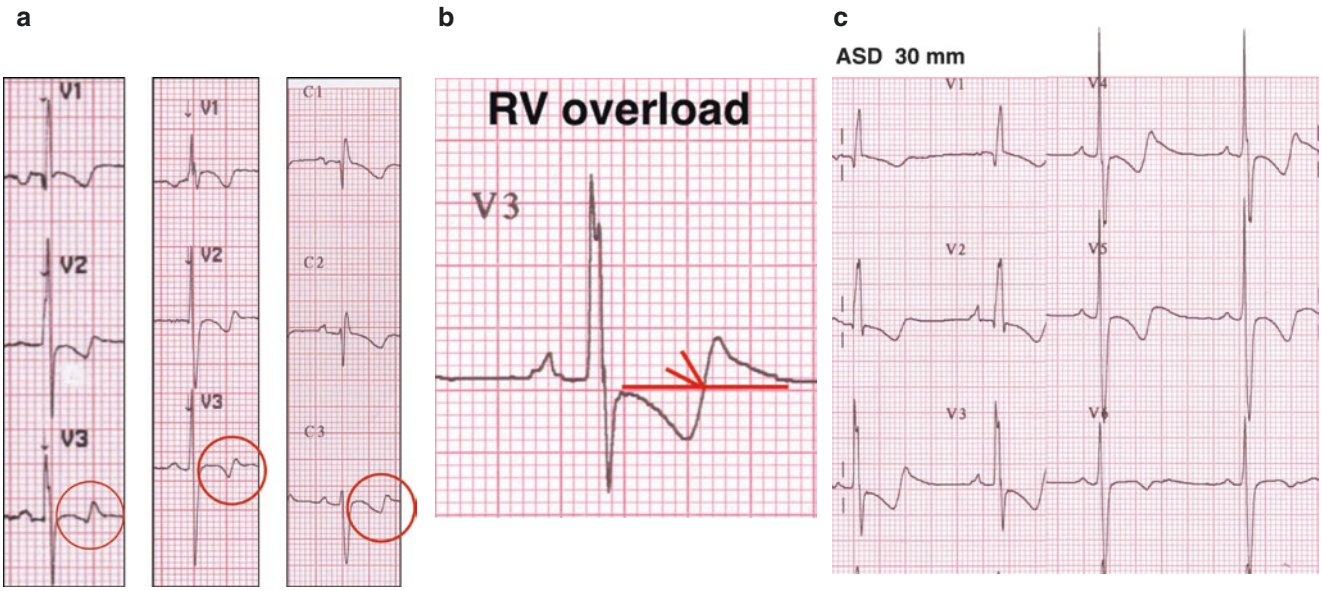


Fig. 18.7 (**a**) Three cases of ASD type II (diameter > 10 mm). In the right precordial leads, the biphasic T wave—negative/positive (overshoot)—is a sensitive and specific sign of right ventricular volume overload (caused in most cases by ASD), especially if extended up to V4–V5. (**b**) In the larger ASD, the biphasic T waves tend to be in symmetry. Drawing a line from the start of the negative component to the end of the positive one, you get a figure which, if not pathognomonic, is at least entomological. This is the *butterfly sign*. (**c**) Nineteen-year-old female with very large ASD type II, diameter 30 mm. The larger the ASD, the more butterfly signs extend to the left precordial leads

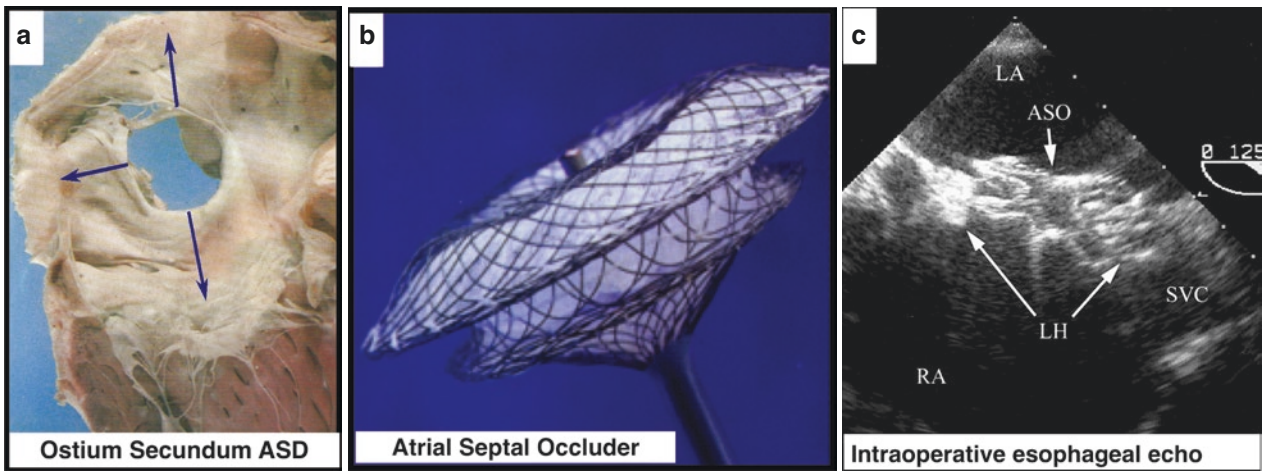


Fig. 18.8 In most cases, type II ASD can benefit from the percutaneous closure with devices of various shapes. It is curious that in the Anglo-Saxon world, these devices are called **butterflies.** So the butterfly sign leads to another … butterfly. (**a**) Gross anatomy of ASD type II; (**b**) the "Amplatzer" TM septal occluder; (**c**) transesophageal echo view of percutaneous ASD closure with Amplatzer septal occluder

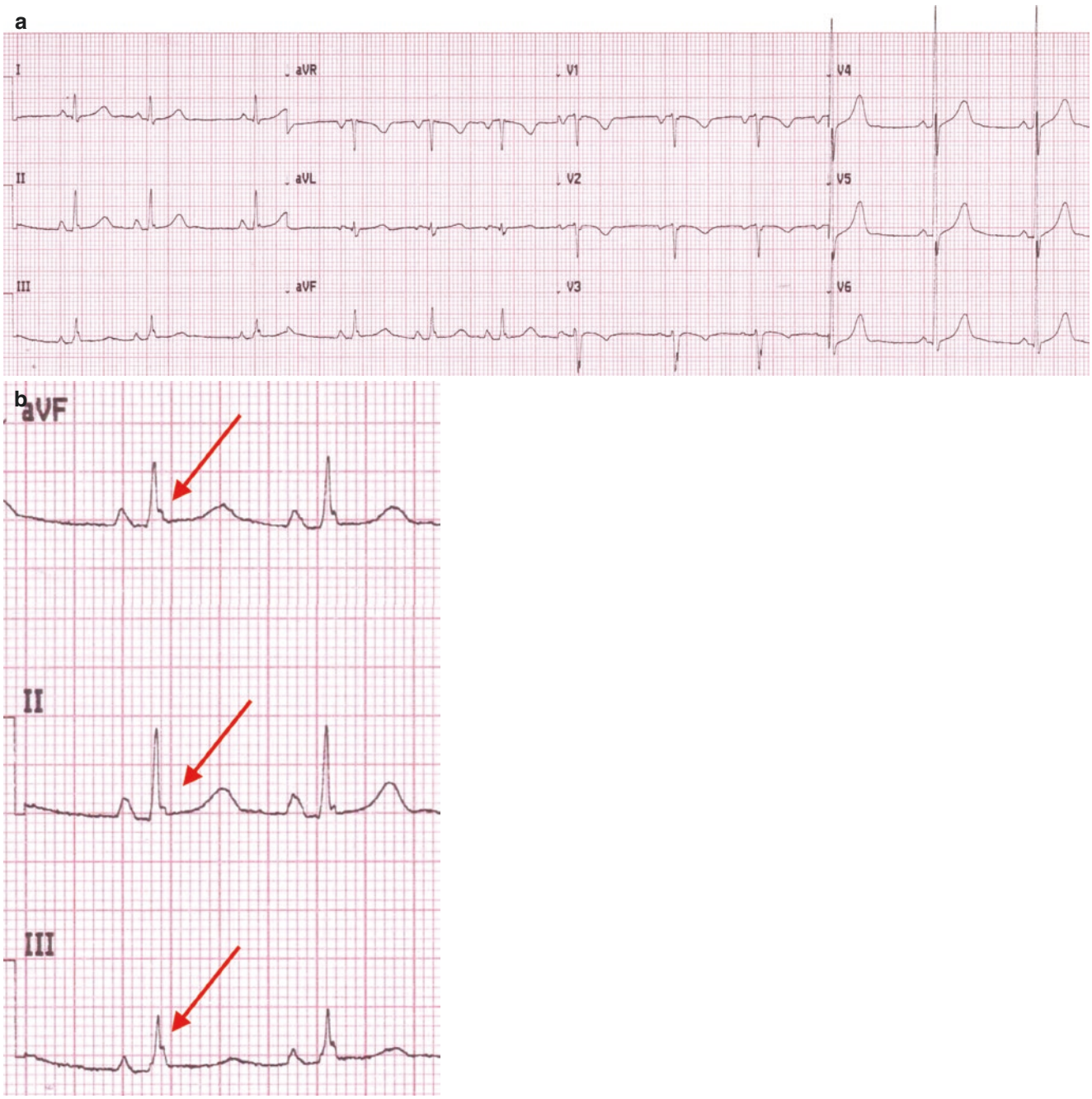


Fig. 18.9 (**a**) Ten-year-old male with ASD type II 8 mm of diameter. The ECG does not suggest a specific diagnosis, in the face of a pathological murmur and a diagnostic echo. Only the inferior leads have a "notch." This sign has been related to volume overload of the right ventricle, more often in ASD, and baptized with the French term "crochetage." In this case, the right precordial leads, usually eloquent in ASD, are silent and the "crochetage" is the only sign of right overload. (**b**) Close-up

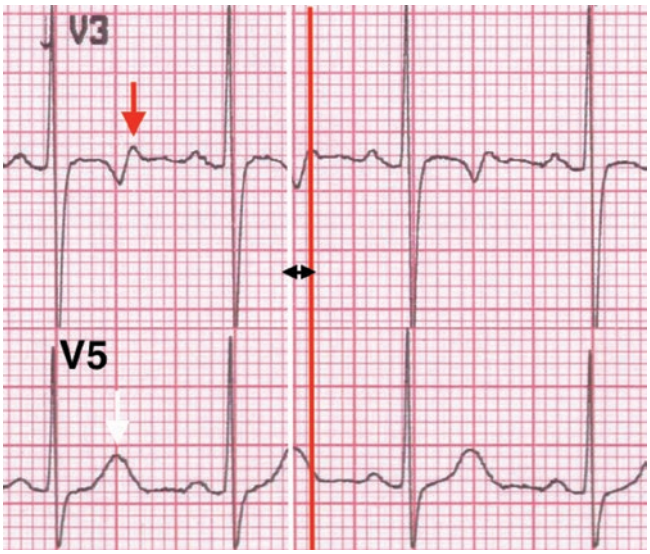


Fig. 18.10 An example of a large ASD. The overshoot in the right precordial leads (*red arrow*) reaches its peak later, compared with the T peak in the left precordial leads (*white arrow*). This is a quite specific sign of right ventricular overload. This splitting (distance between *red* and *white line*) is proportional to the degree of right ventricular overload and to the splitting of the second tone that might be found on auscultation. So you might hear and see double, a double split (*ASD as an acronym of Auscult-See-Double)*

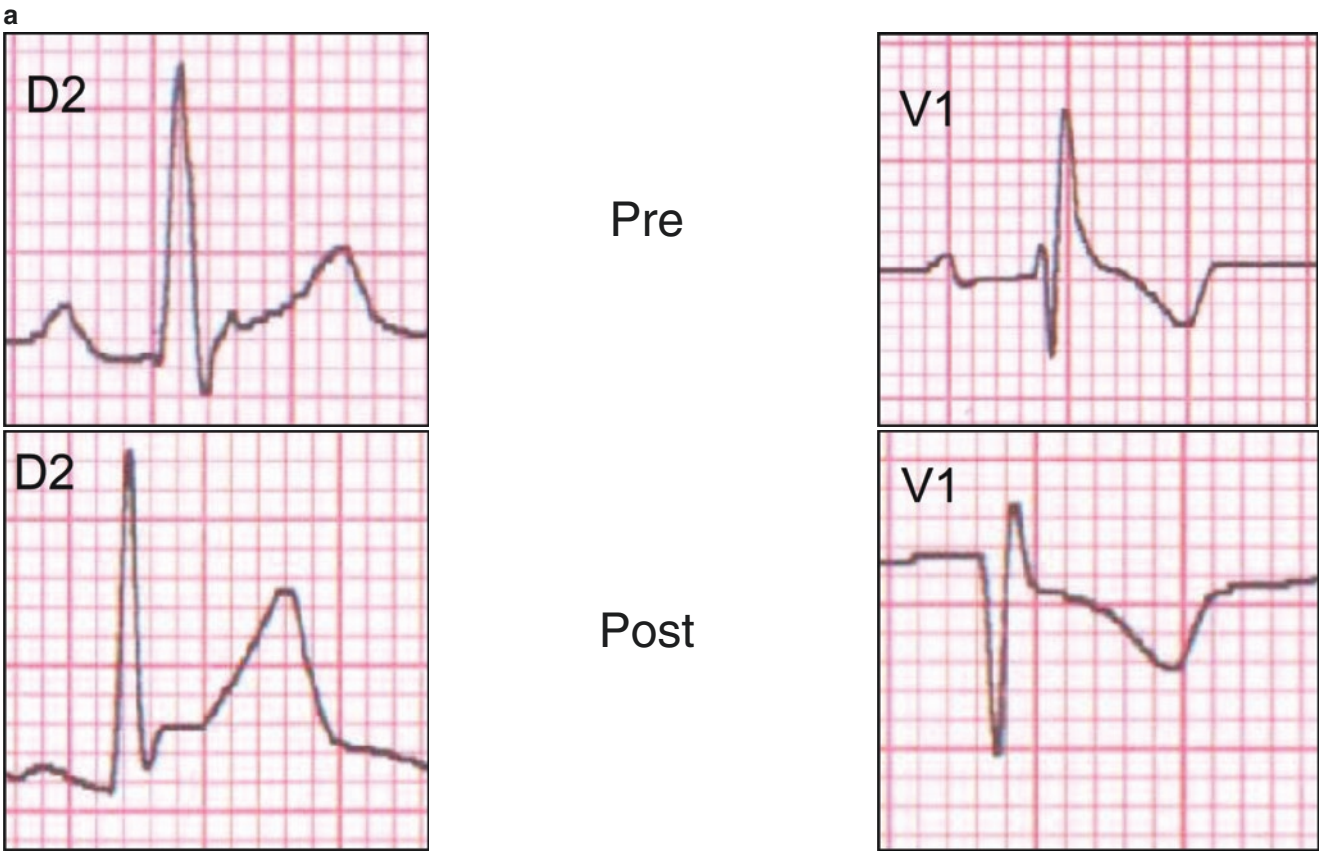
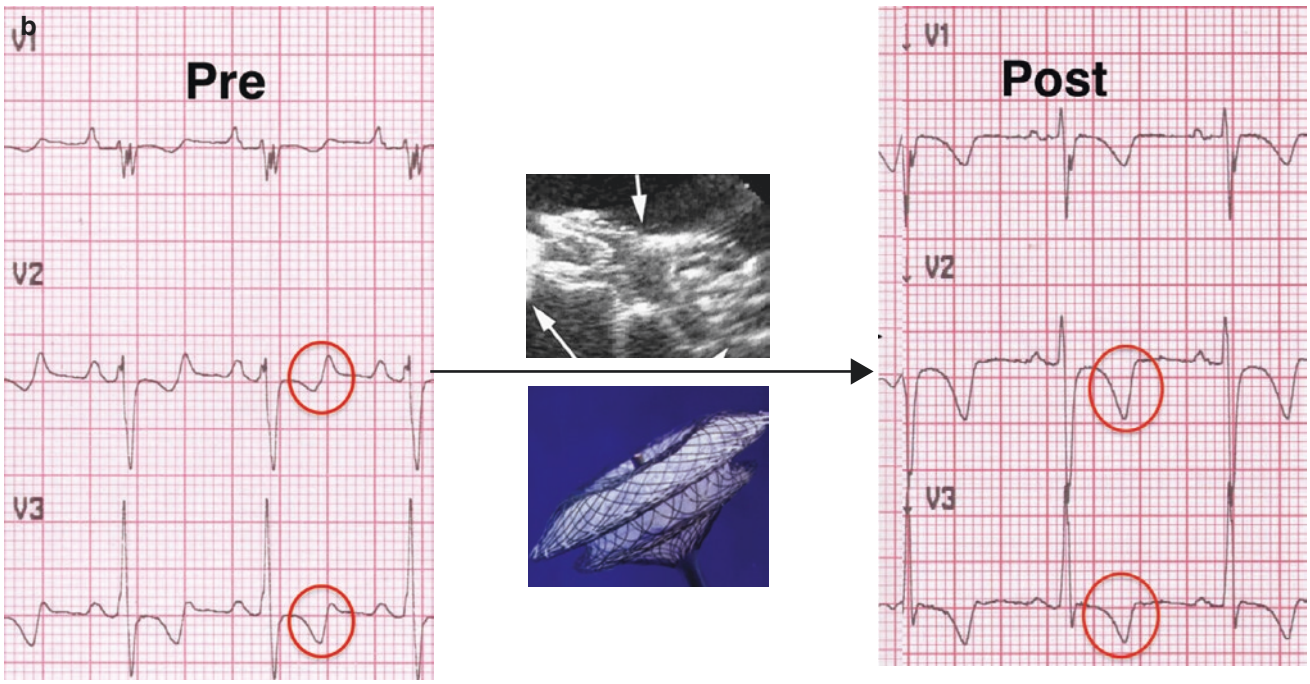


Fig. 18.11 (**a**) Eleven-year-old male pre- and post partial pulmonary anomalous venous return repair (PPAVR). Before the correction, the "crochetage" in DII and the broad R' in V1 can be seen. After the repair the QRS in lead II shrinks and the "crochetage" disappears; the R 'in V1 is also reduced. (**b**) Nine-year-old female. After the ASD closure, the butterfly sign disappears and the QRS in V1-V2 is less splintered



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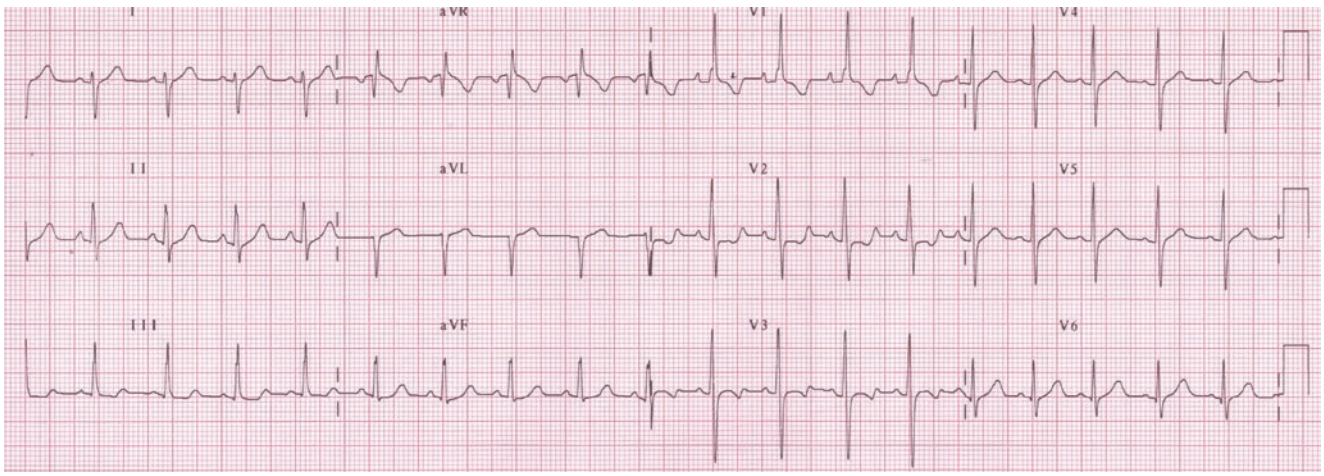


Fig. 18.12 Four-year-old male with partial anomalous pulmonary venous return (PAPVR). The first echocardiogram was nondiagnostic (PPAVR is tricky on echo because the atrial septum can be intact), but the monophasic and slurred R wave in V1 along with the overshoot forced us to perform a second, diagnostic ultrasound scan

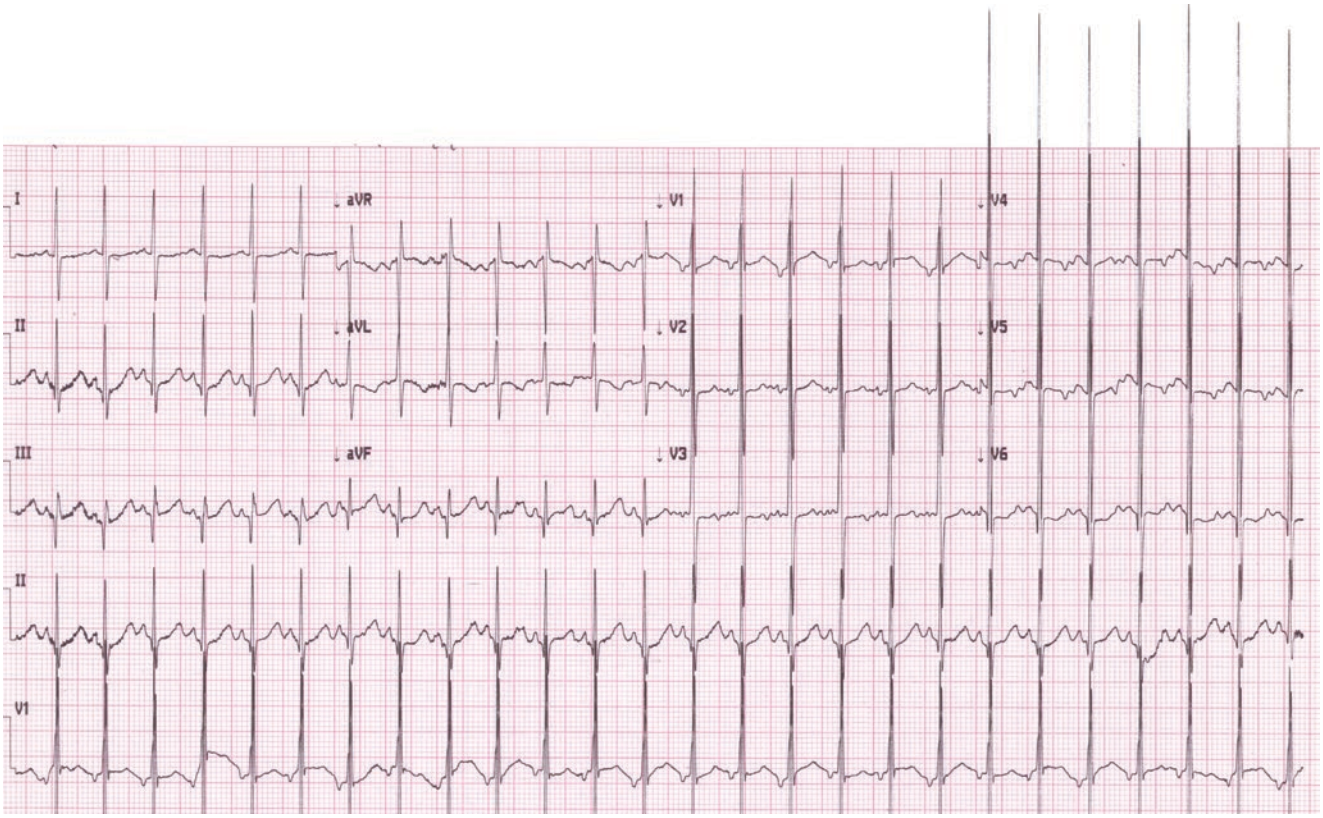


Fig. 18.13 2-month-old baby boy with failure to thrive due to a large subaortic VSD. Signs of biventricular hypertrophy and widespread high voltages of QRS, especially in the precordial leads. These large biphasic QRS complexes from V2–V5 (symmetric R/S in the midprecordial leads) are called Katz-Wachtel sign. This kind of overload is shared by other types of post-tricuspid shunt such as patent ductus arteriosus, arteriovenous fistula, coronary fistula, AV malformation, etc.

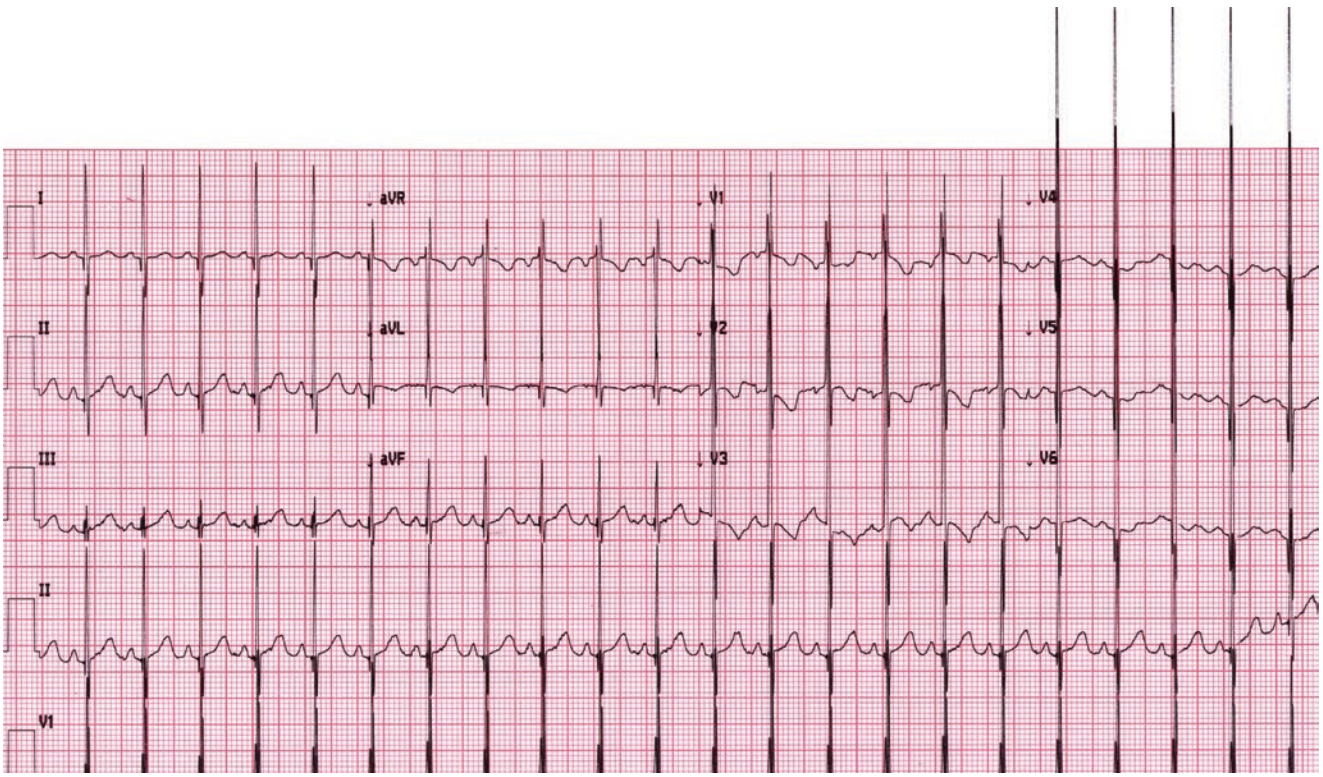


Fig. 18.14 Three-month-old baby boy with large ductus arteriosus. Biventricular overload (and Katz-Wachtel signs) can be noted

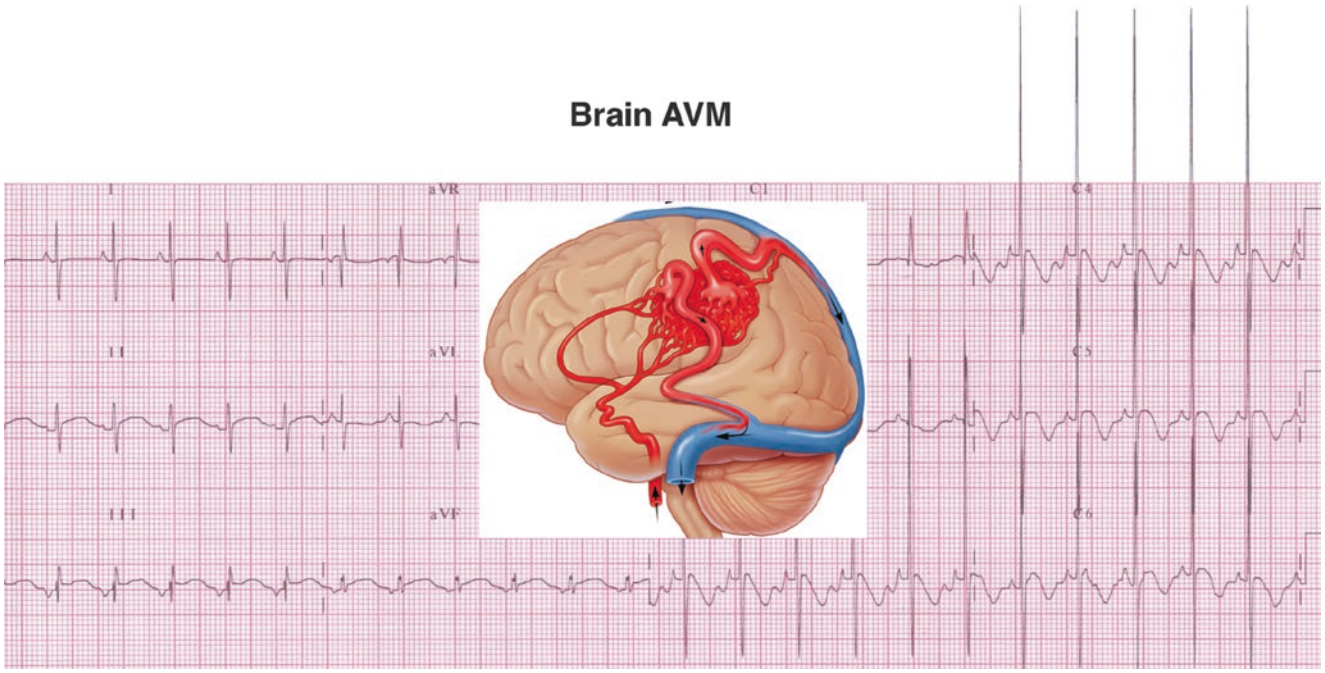


Fig. 18.15 Five-month-old female with massive cerebral arteriovenous malformation (brain AVM). A biventricular overload with ST change can be noted. The ECG is compatible with a large VSD or patent ductus of Botallo. The echo ruled out CHD but found dilated sections and normal biventricular function. The condition was that of high output heart failure, as it is possible to find in anemia and hyperthyroidism. In this case, the auscultation found a systolic-diastolic murmur in the head, leading to the diagnosis of cerebral arteriovenous malformation

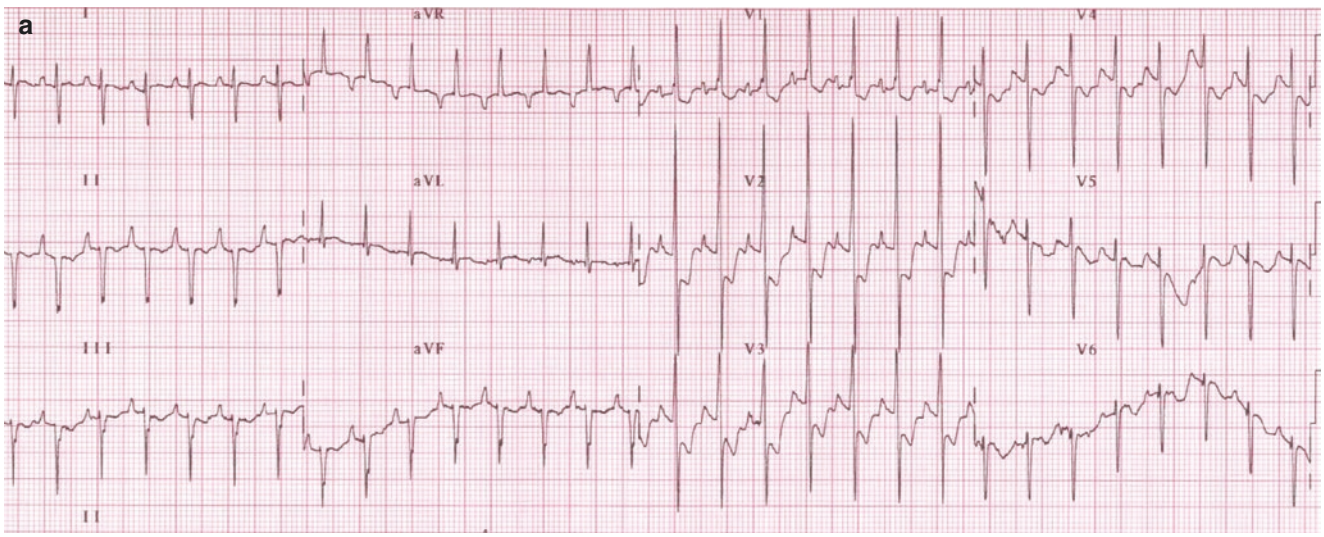
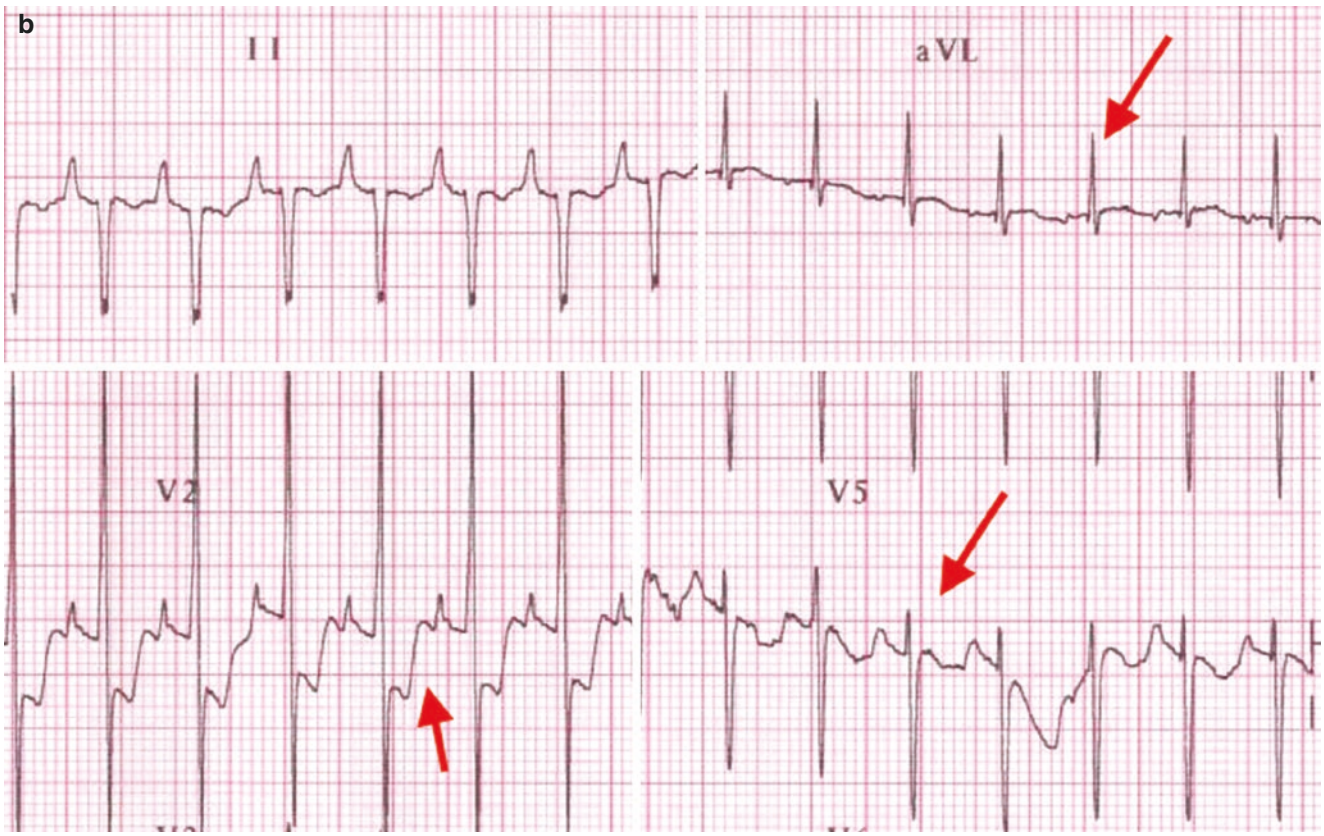


Fig. 18.16 (**a**) One-month-old female admitted to ER for severe heart failure (shock). The echo found a CAVC and aortic coarctation (AC); this last dramatically showed up soon after the Botallo's closure. There is a left anterior hemiblock which in neonates is almost pathognomonic of CAVC. The right ventricle overload and ischemia is due to the pulmonary hypertension and to AC; the left precordial leads are poorly represented (the short R waves are only partly due to left anterior hemiblock). Such a right dominance is not surprising as also the right ventricle has the task of bearing the weight of AC in fetal life. Also a right atrial enlargement can be noted. (**b**) Close-up



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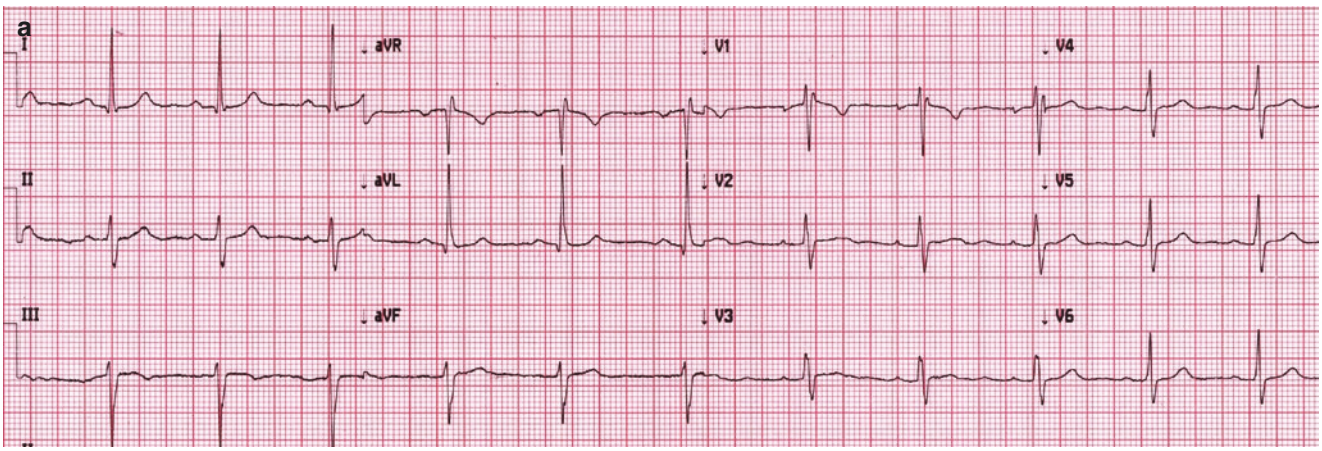
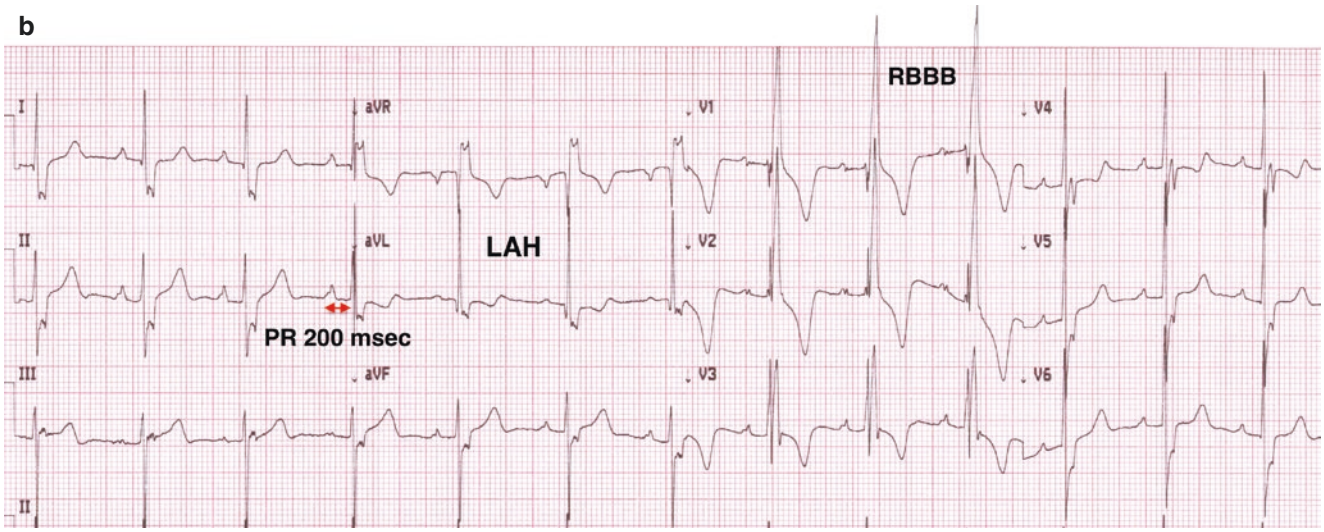


Fig. 18.17 (**a**) Three-year-old male, partial AV canal defect (type I *ostium primum* ASD and mitral cleft). LAH and mild right conduction delay can be noted. (**b**) Eight-month-old female with intermediate AV canal (restrictive VSD). All the features of AV canal are present: first-degree AV block; LAH; RBBB



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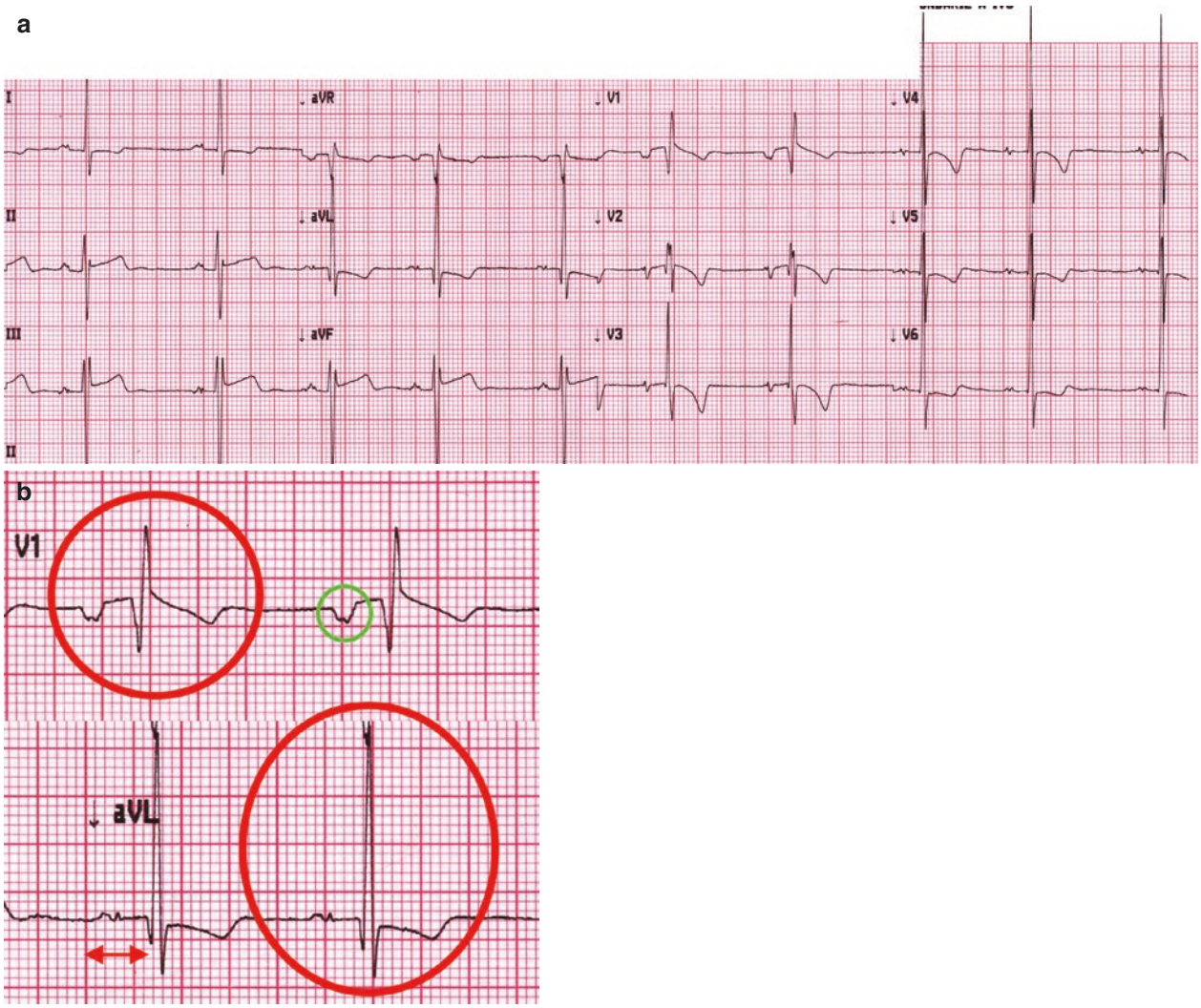


Fig. 18.18 (**a**) Two-year-old male trisomy 21 and complete AVC in the early postoperative course of complete repair. As in the previous figure, there are all the typical aspects of the CAVC: LAH, long PR interval, RBBB. (**b**) Close-up: note also the left atrial enlargement and the ST depression in the left precordial leads, due to the severe regurgitation of the left AV valve (which is not exactly a mitral valve)

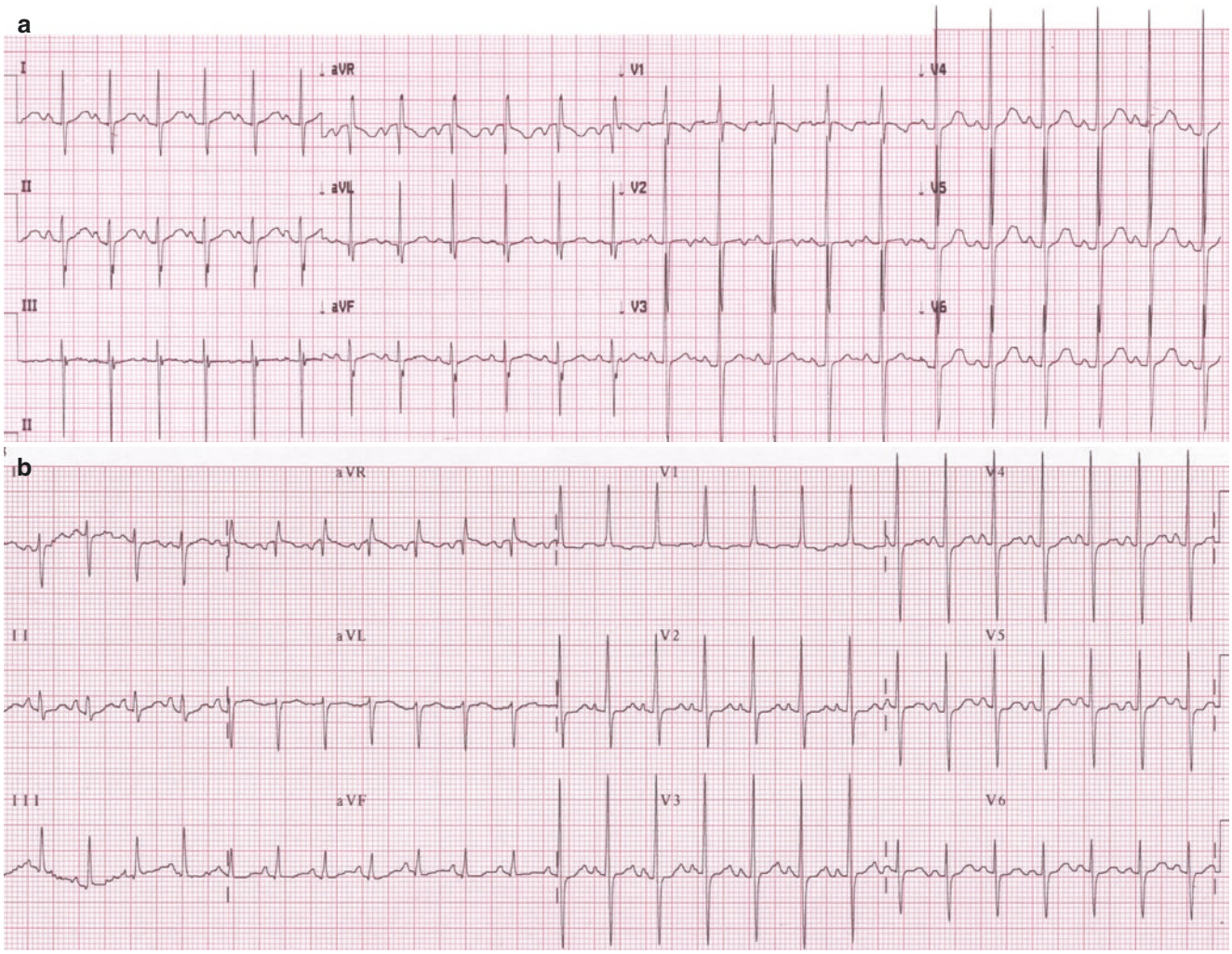


Fig. 18.19 (**a**) Three-month-old boy with intermediate AVC. There is mitral regurgitation through the cleft. The VSD is restrictive by definition, but the left-to-right shunt is significant as can be inferred from the Katz-Wachtel sign. (**b**) 20 day old male with total anomalous pulmonary venous return (TAPVR). Monophasic R wave in V1 and large S wave in V6 can be noticed

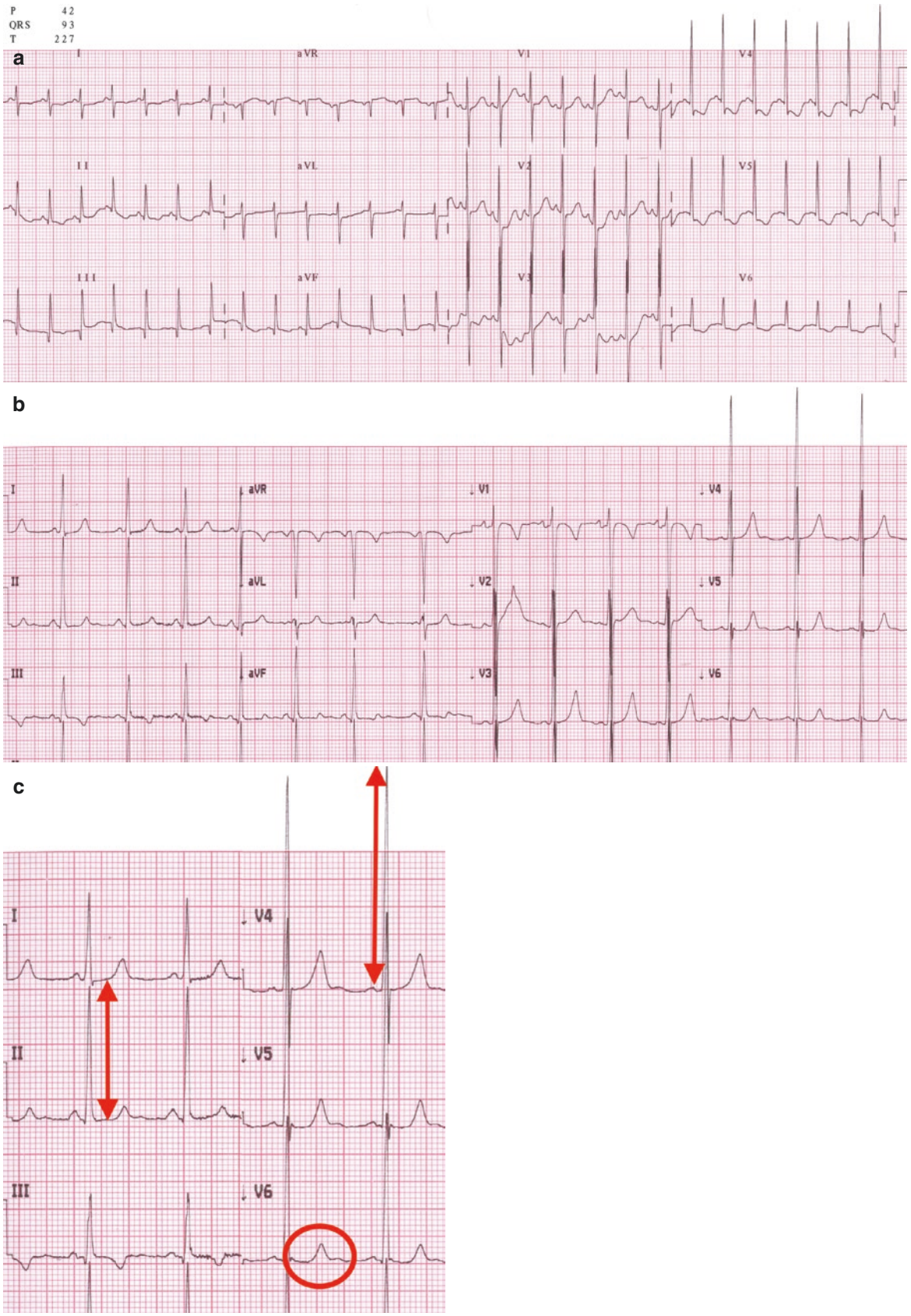


Fig. 18.20 (**a**) Neonate with bicuspid aortic valve (BAV) and severe aortic stenosis. There is a widespread ST depression due to the left ventricle overload. The positive T wave in V1 is normal in the first week of life. (**b**) Ten-year-old male with BAV and severe steno-insufficiency; the voltages meet the criteria for LVH and a slightly downsloping ST segment can be noted (**c**)

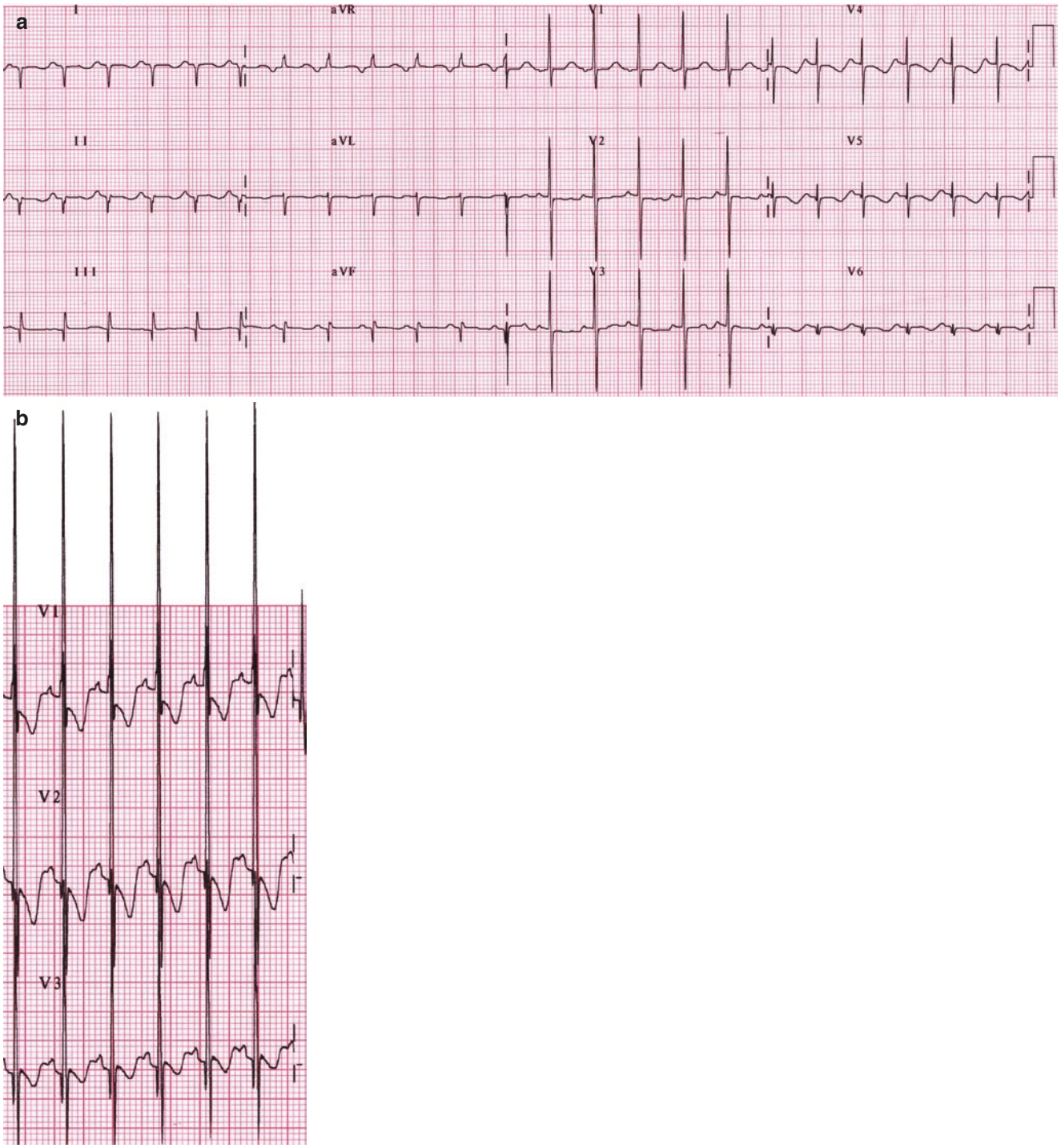


Fig. 18.21 (**a**) One-month-old baby boy with aortic coarctation: the voltages are not significant while there are diffuse T changes, including the positive T wave in V1; as in other ductus-dependent left-side obstruction, the right ventricle is not indifferent to the overload. (**b**) In this case of neonatal coarctation, the right precordial leads denote the severe right ventricle involvement and left-side forces are poorly represented

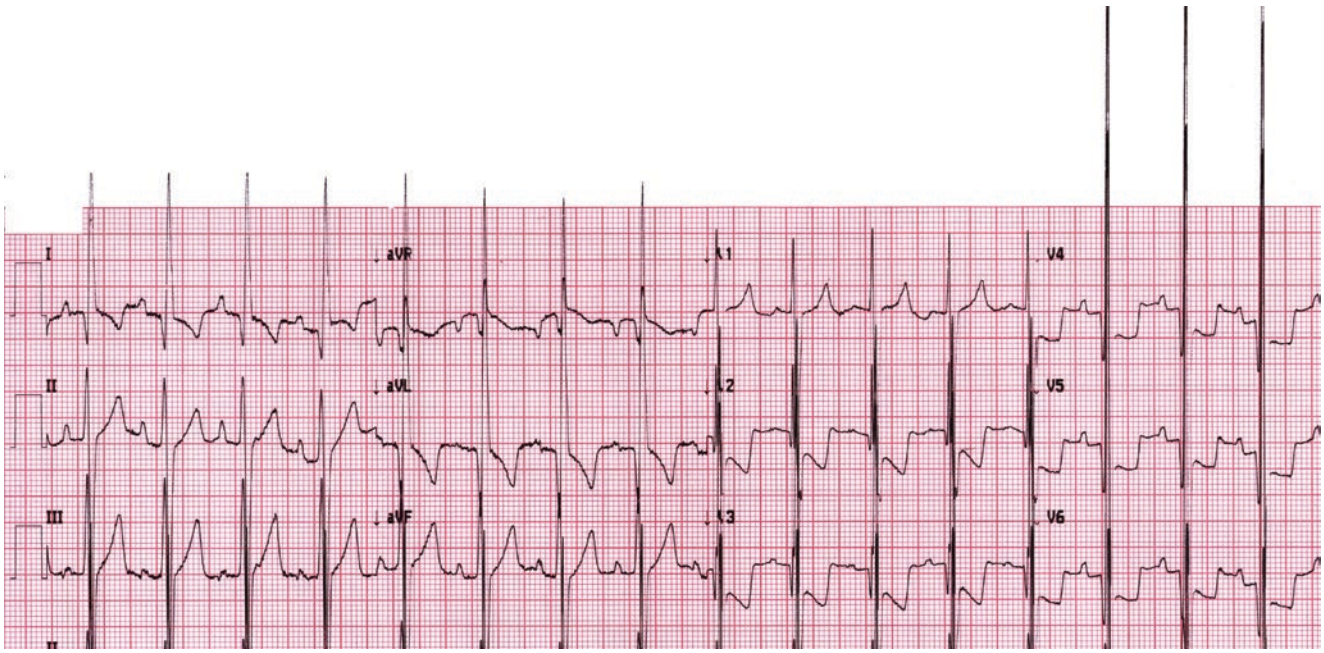


Fig. 18.22 Nine-year-old female with BAV and severe stenosis. The typical ST-T changes in LVH consist of ST depression with upward convexity and T wave inversion in the left precordial leads, with reciprocal change in right precordial leads. In children and young adults, the voltage criteria are not useful due to the normally higher voltage of QRS. That is why repolarization is more important than depolarization in judging the LVH. In this case, there is an impressive amplitude of the R wave, but the ST depression is the actual diagnostic clue

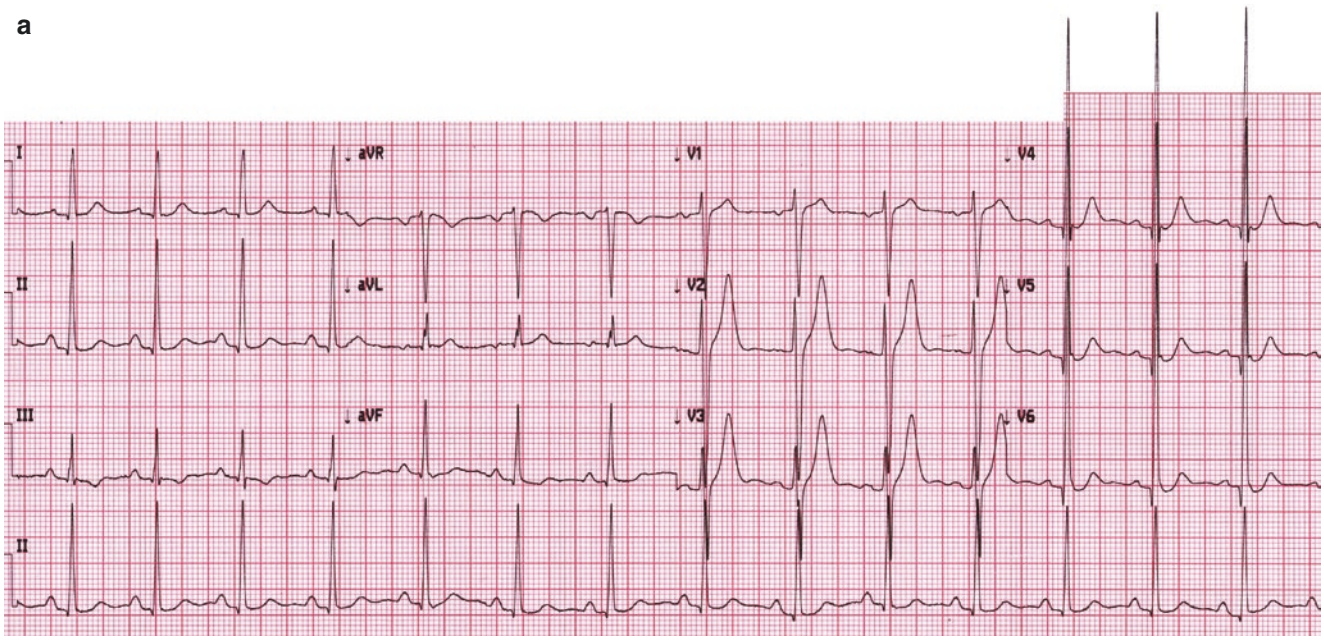
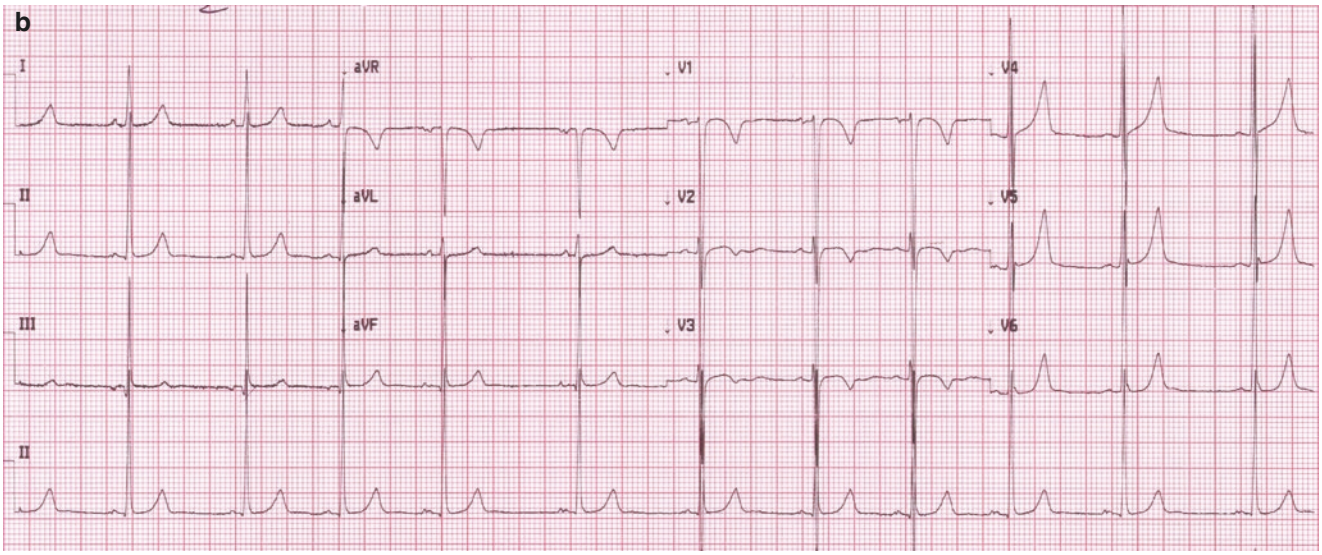


Fig. 18.23 As already stated, the diagnosis of LVH using voltage criteria is disappointing in the young. (**a**) Sixteen-year-old male with aortic stenosis; the peak systolic gradient across the valve is 60 mmHg. In the precordial leads, the voltage criteria are met, but they are aspecific in the young. (**b**) Nineteen-year-old male with BAV and aortic stenosis with a peak gradient of 75 mmHg. The voltage criteria (Sokolow-Lyon) are met also in the limb leads: S aVR > 14 mm; R aVF > 20 mm; precordial largest R + precordial largest S > 45. The repolarization is not affected, because the negative T waves in the precordial leads can be considered a normal variant. In this case, the auscultation of a systolic harsh murmur and the ECG can guess the diagnosis before the ultrasound scan



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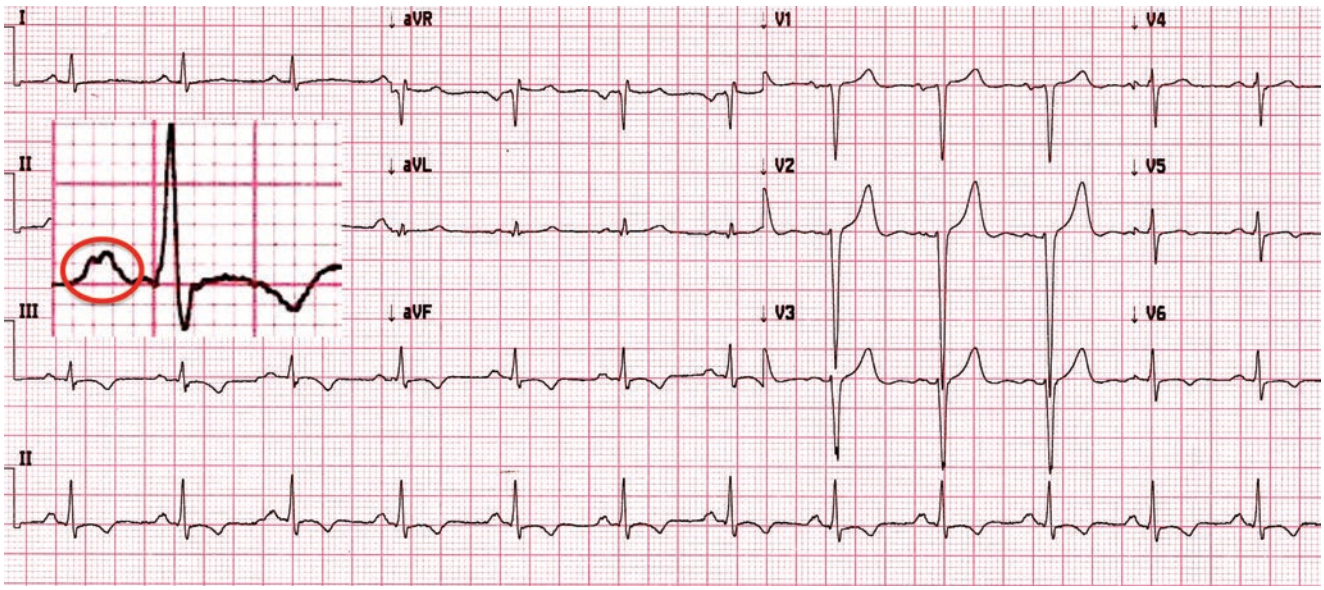


Fig. 18.24 Twenty-two-year-old obese male with BAV and severe aortic stenosis. The obesity dampens the amplitude of the voltages, especially in the left precordial leads. However, the ST-T changes in the inferior and in the precordial leads are suggestive of LVH. The left atrial enlargement, see lead II, in an ancillary sign of LVH

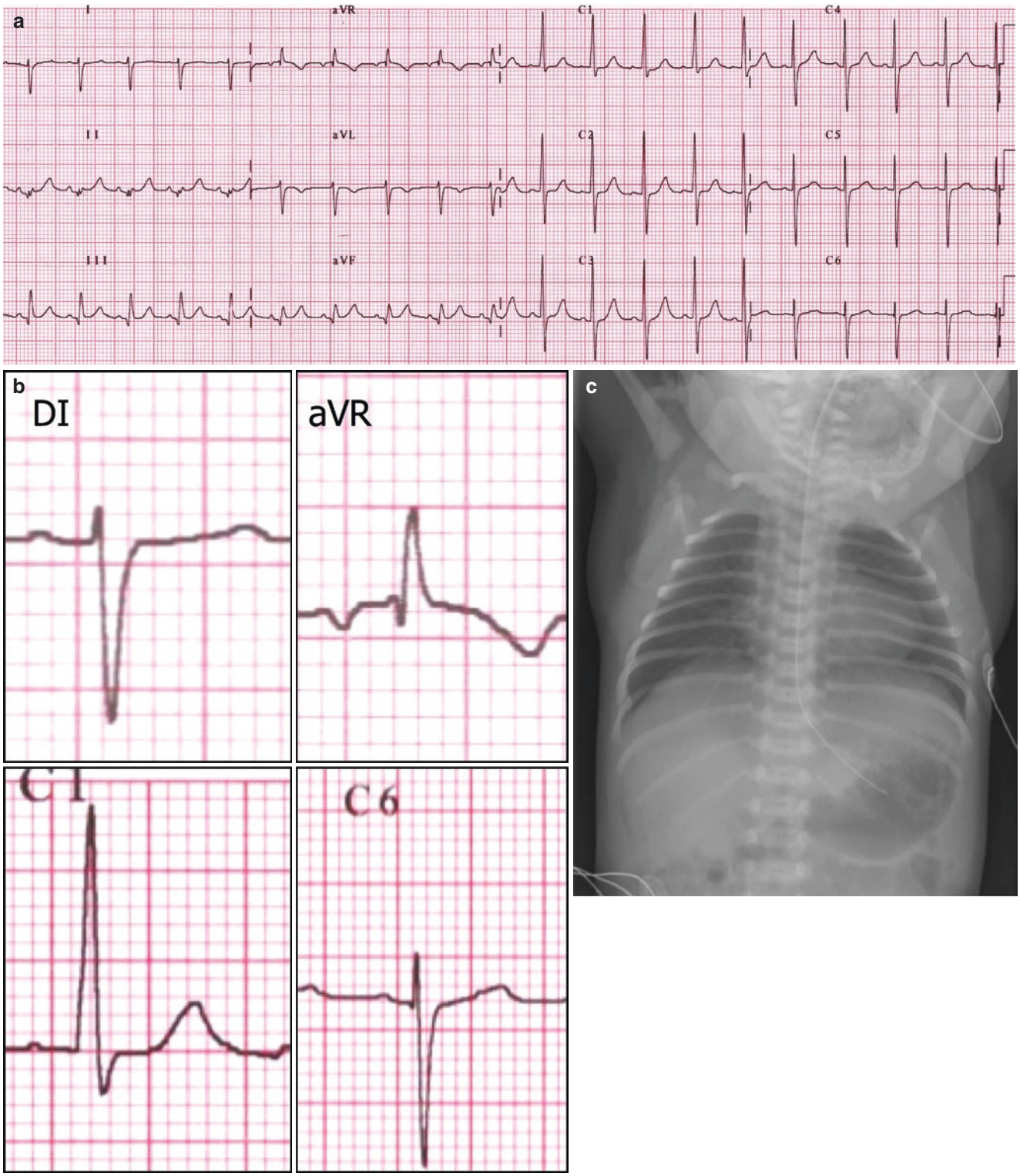


Fig. 18.25 (**a**) Five-month-old female with tetralogy of Fallot and moderate cyanosis. Right axis deviation, QRS axis 135°. The right ventricular hypertrophy is never extreme because the right ventricle is relieved by the shunt to the left ventricle (of course at the price of cyanosis). (**b**) The left ventricle, however, is poorly represented (small R and large S in V6), because despite the presence of VSD in tetralogy of Fallot, there is a decreased pulmonary blood flow. (**c**) Chest X-ray showed the classic shape "cor en sabot," socket-like heart and "dry" lungs

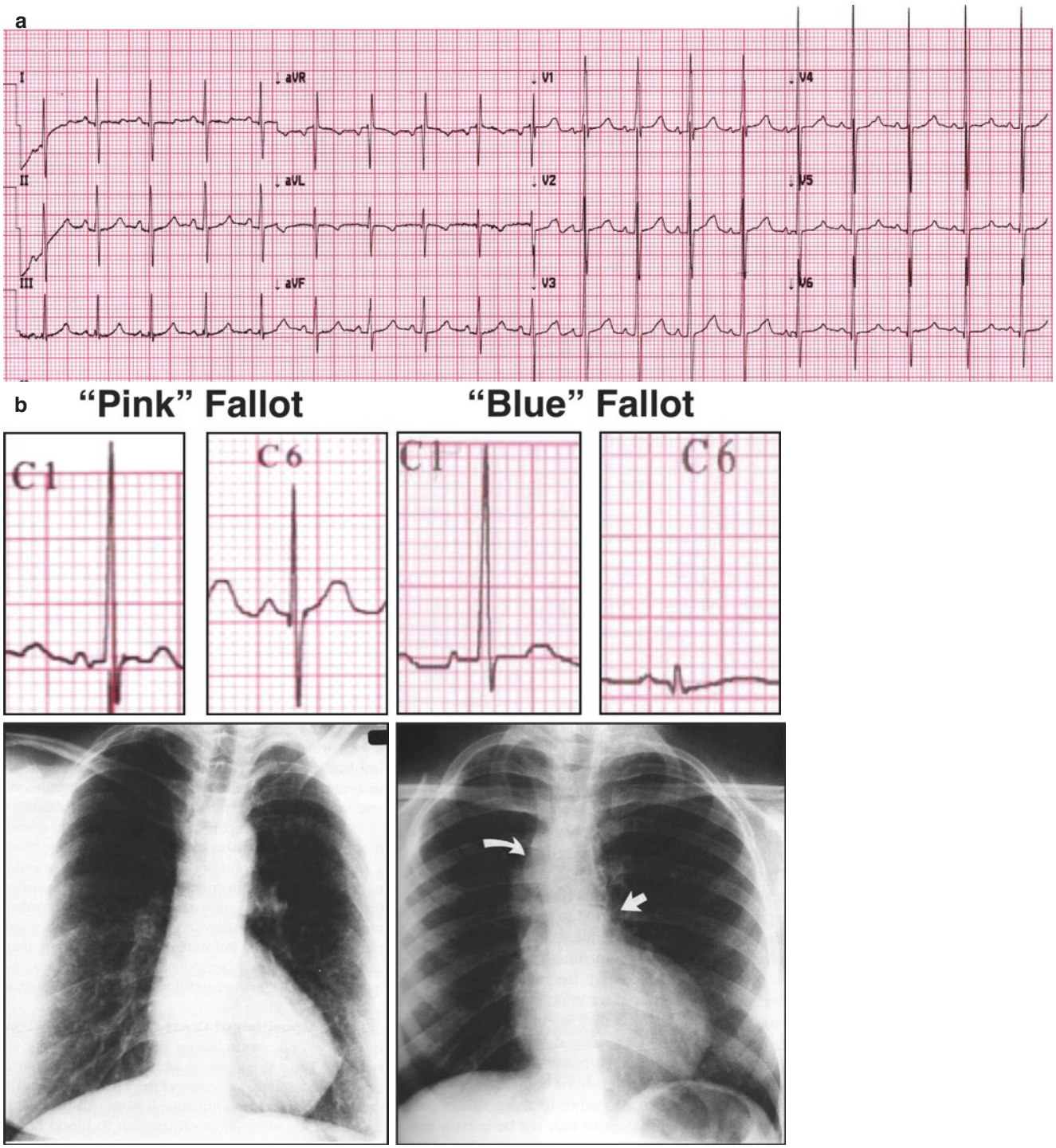


Fig. 18.26 (**a**) Six-month-old Fallot with mild obstruction, so-called pink Fallot. The R wave in V6 is a marker of pulmonary blood flow and in this case is nicely represented. (**b**) The "pink Fallot" looks like a VSD. In contrast, a severe obstruction will show poor left voltage and "dry" lungs

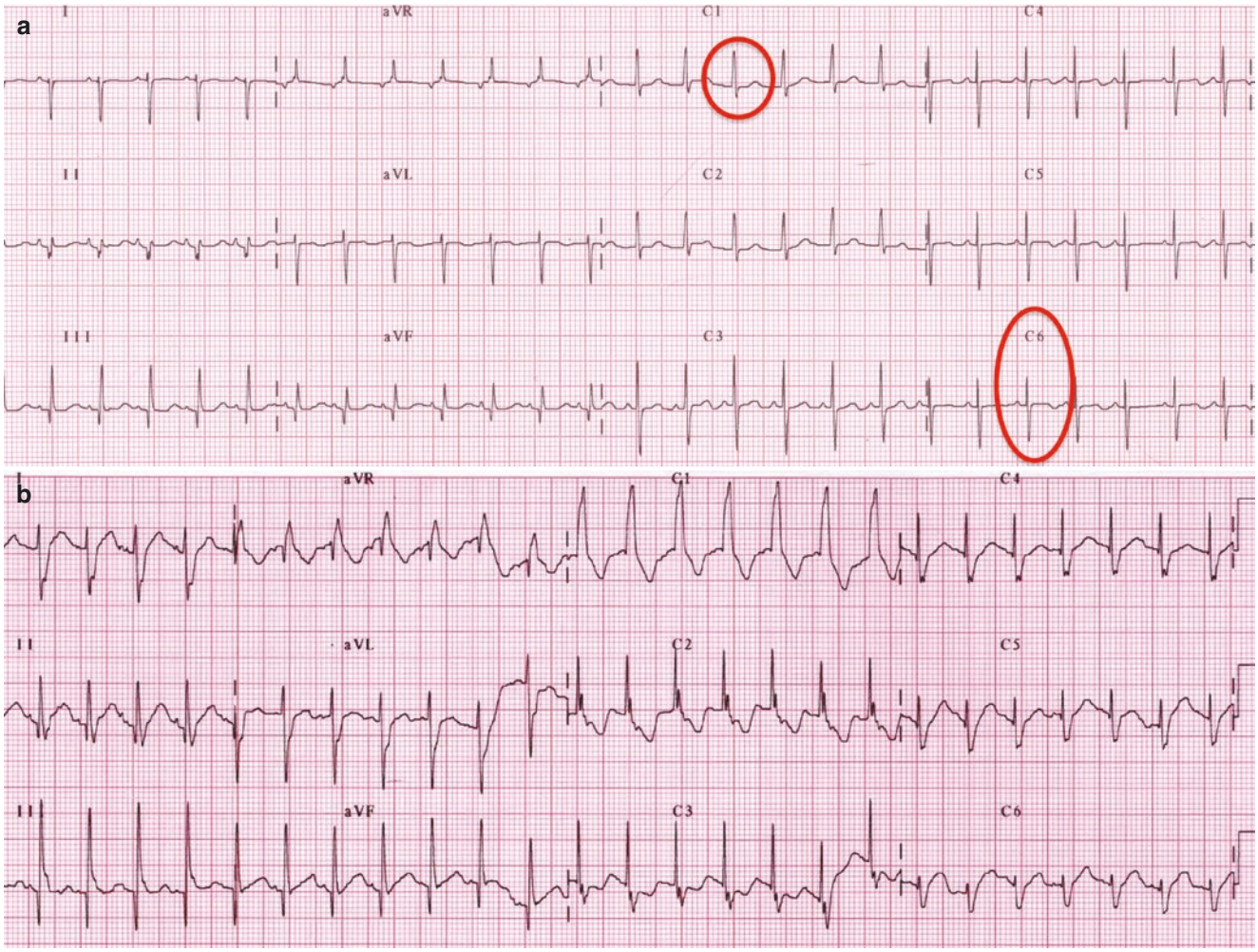


Fig. 18.27 (**a**) Pre-op ECG of eight-month-old Fallot with severe cyanosis on treatment with propranolol. Right axis deviation, positive T wave in V1, and scarcely represented left ventricle are consistent with the history (**b**) Post op ECG: RBBB is usual after tetralogy of Fallot repair

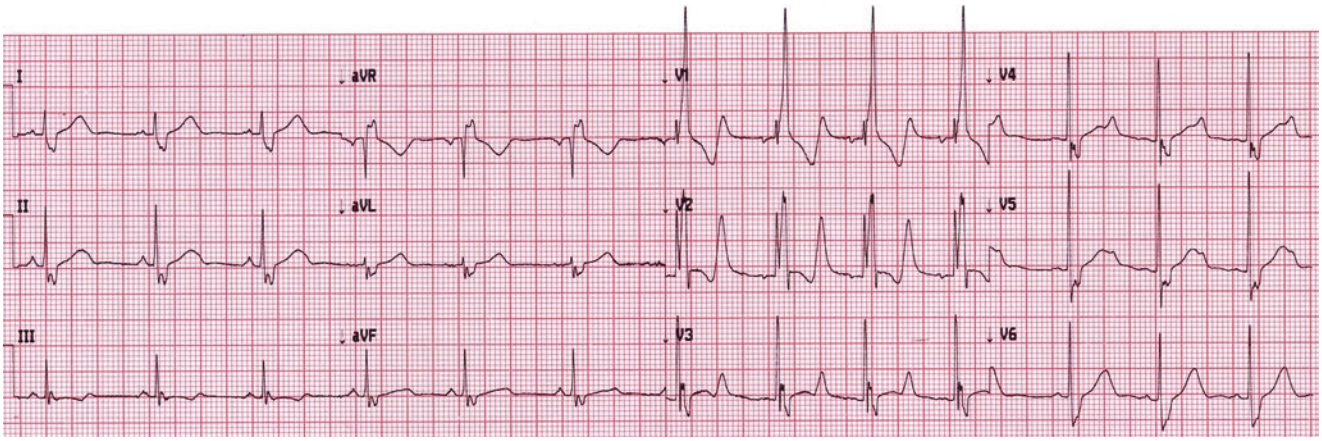


Fig. 18.28 Nine-year-old tetralogy of Fallot who underwent complete repair at 6 months of life. The RBBB is not a concern because is almost invariably present after ToF repair. Note that in V1, the first R wave is very narrow (spikelike), so narrow that the ECG machine interpretation reported "pacemaker rhythm, RBBB." The machine is almost perfect but *it* is unaware that in 99.9% of pacemaker carriers, the ventricular lead is in the right ventricle and the ECG will show a LBBB

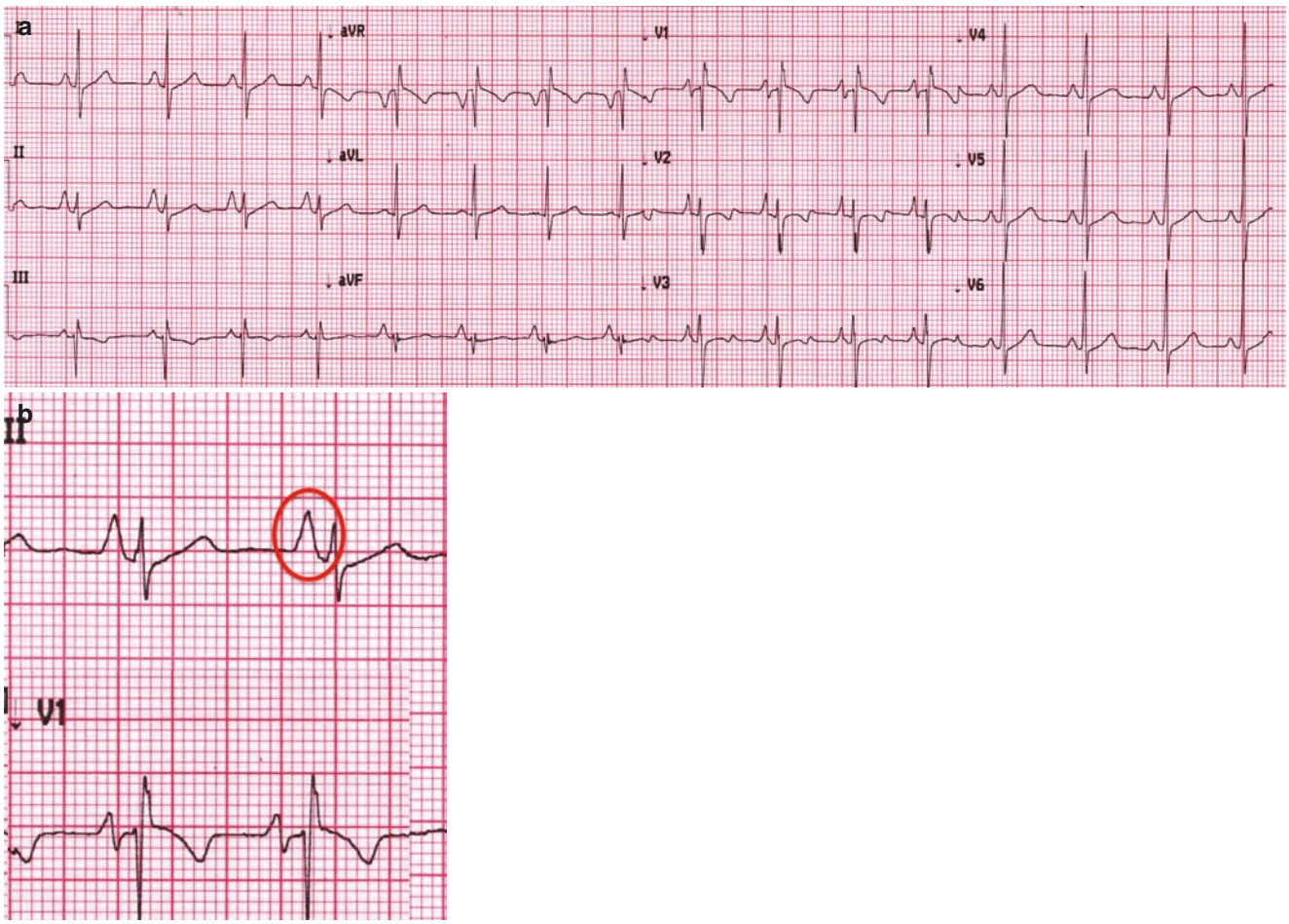


Fig. 18.29 (**a**) Six-year-old boy with pulmonary atresia with intact ventricular septum (PAIVS). Large ASD, no aortopulmonary shunt, oxygen saturation 88%. Right atrium enlargement and right ventricle hypoplasia are typical of PAIVS and are perfectly addressed by this ECG (**b**)

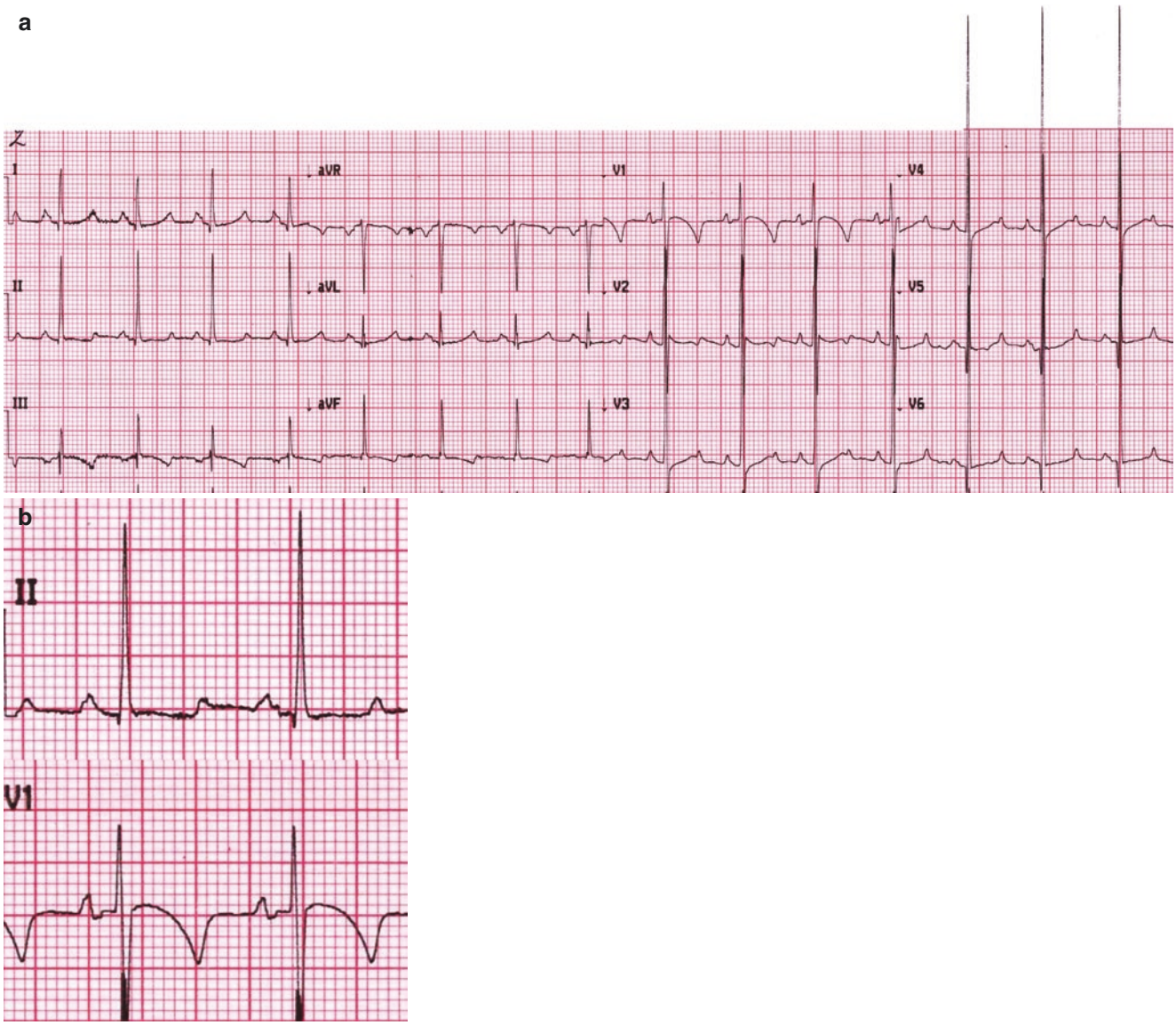


Fig. 18.30 (**a**) Two-year-old boy with PAIVS. ASD is closed, there is a Blalock-Taussig shunt, and the oxygen saturation is 94%. Even in this case, as in a condition much different from a pathophysiological standpoint, the tetralogy of Fallot, the left ventricle voltages are proportional to the oxygen saturation. The BT shunt is responsible for the pulmonary blood flow. (**b**) Close-up

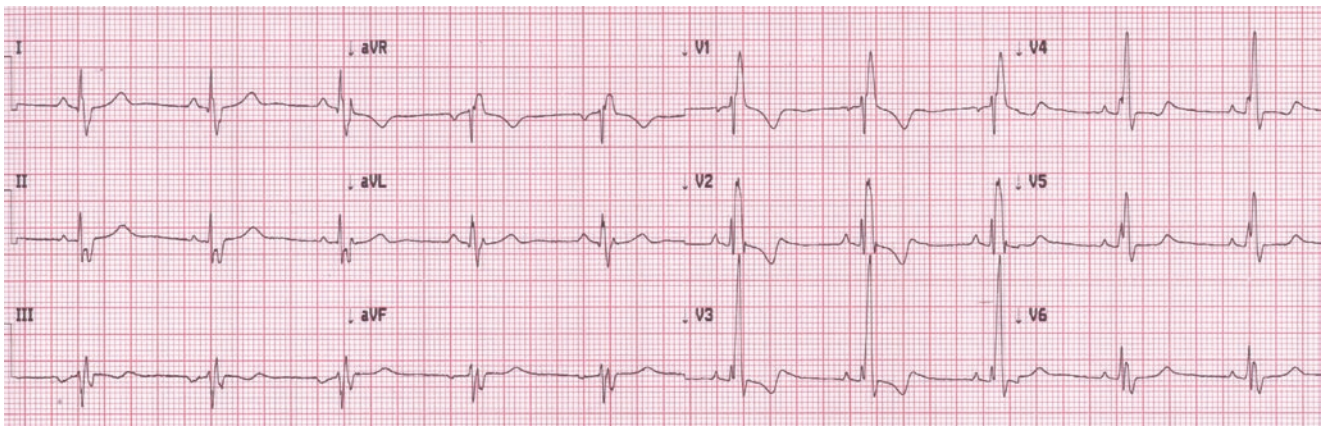


Fig. 18.31 Sixteen-year-old female with pulmonary atresia with VSD who underwent repair in infancy. PAVSD is an extreme form of Fallot (complete disconnection between the right ventricle and the pulmonary artery). The ECG with the RBBB is the same as a Fallot

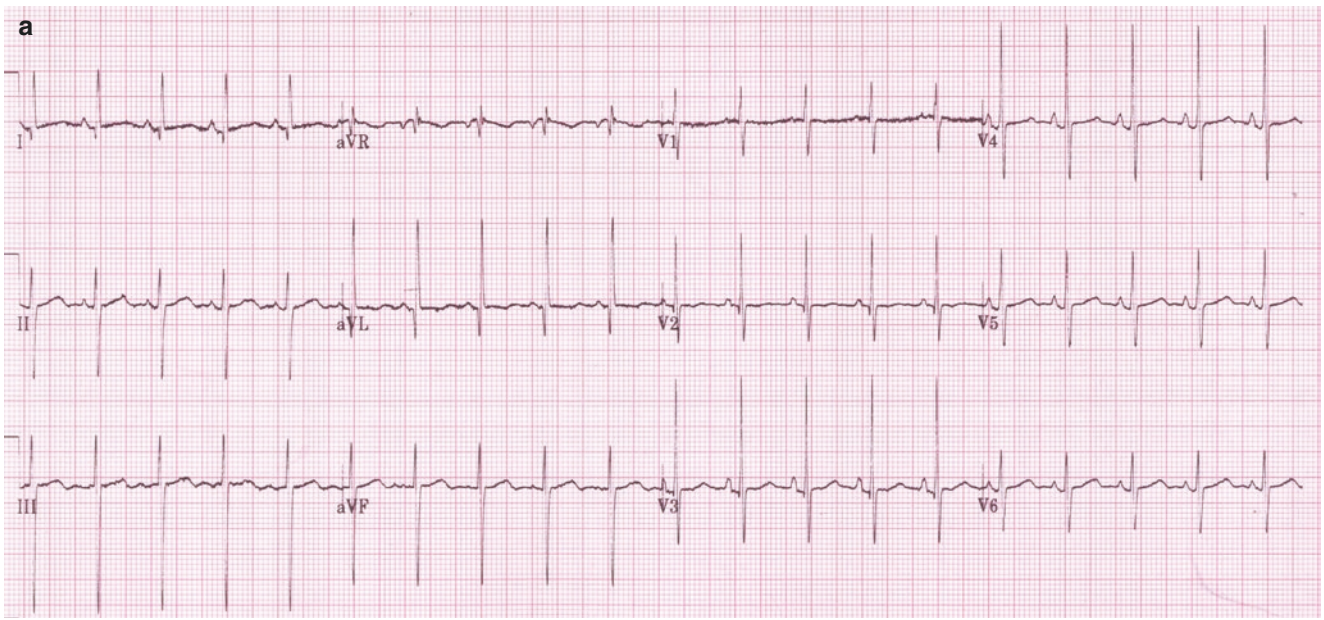
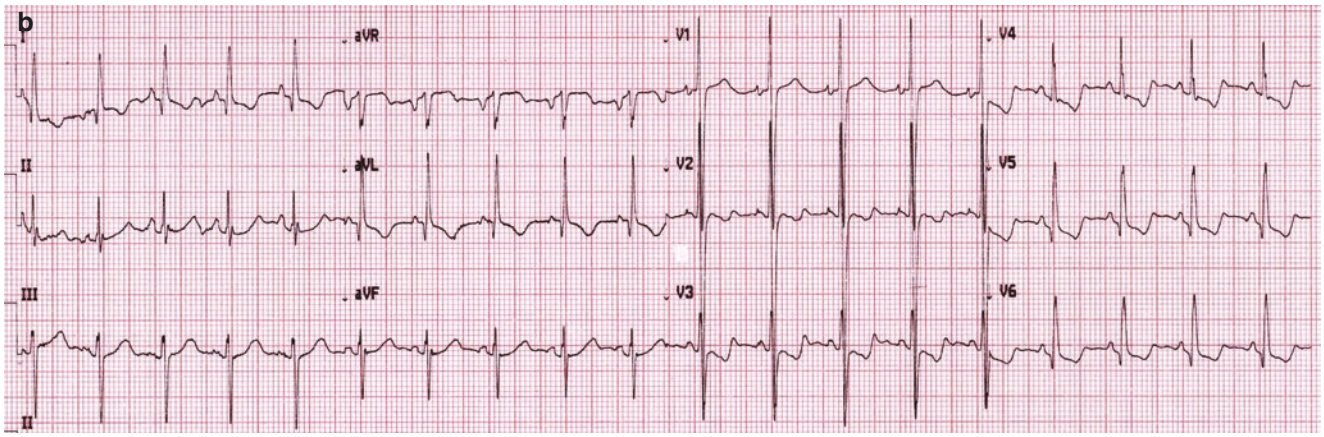
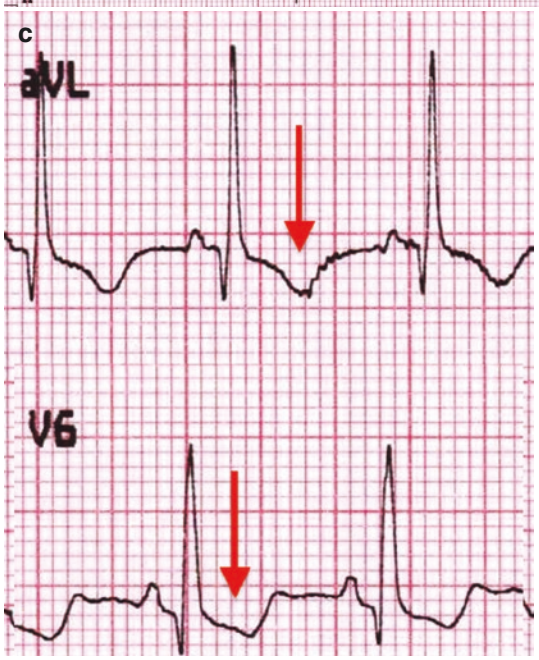


Fig. 18.32 (**a**) Neonate 7-day-old male with tricuspid atresia (TA), the commonest form of single ventricle. Cyanosis and LAH on ECG allow the diagnosis before ultrasound. (**b**) TA in a 5-year-old male palliated with BT shunt and Glenn. LAH is present, the right ventricle is poorly represented, and there is left ventricle overload/strain (**c**)





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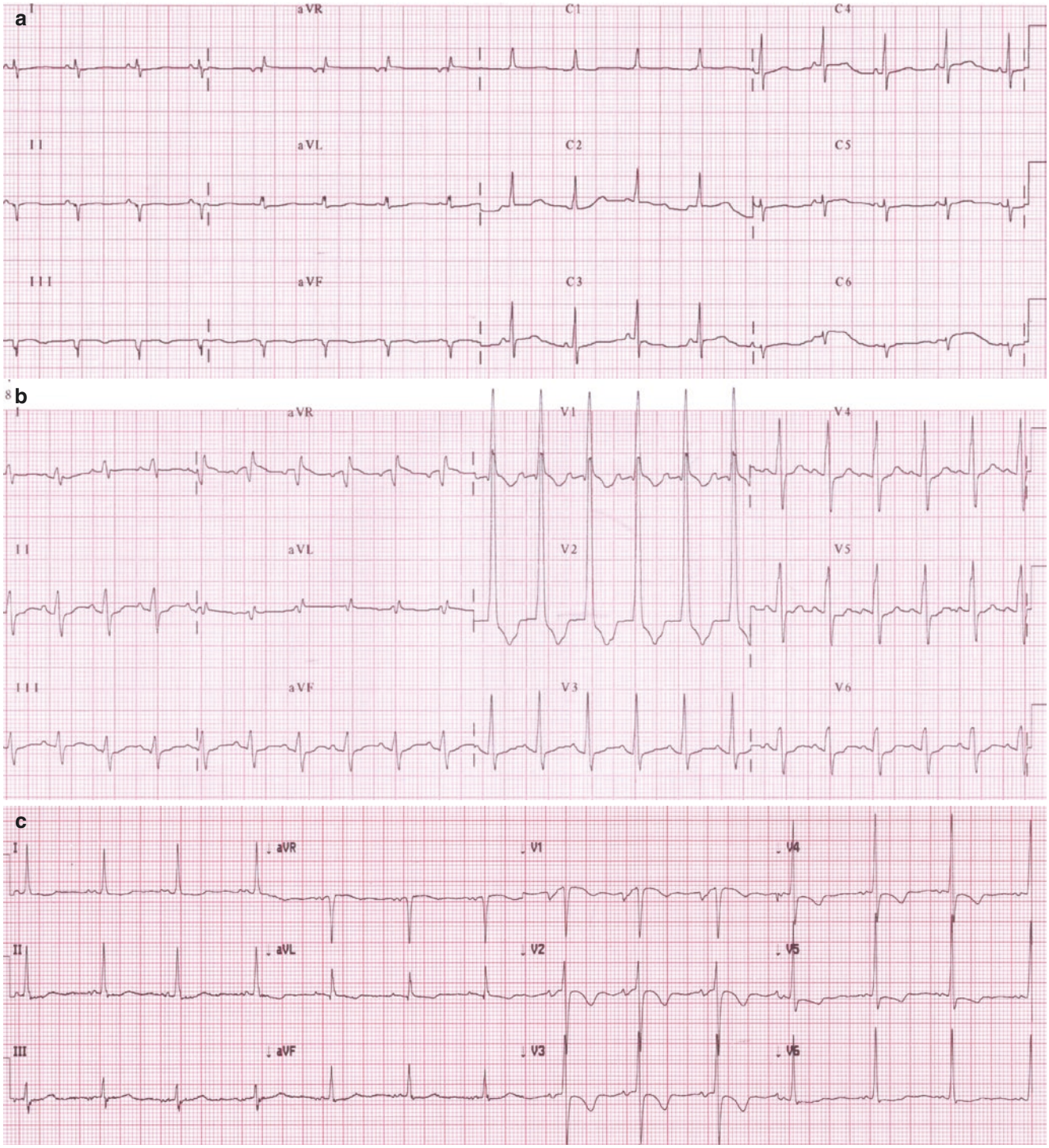


Fig. 18.33 (**a**) HLHS newborn baby boy. Widespread low voltages of QRS. In particular, the left forces are scarcely represented. (**b**) Ten-year-old HLHS after Norwood palliation. The left ventricle is atretic and there are only right ventricle forces. (**c**) Single ventricle due to right ventricle hypoplasia. Unlike the previous case, the left forces predominate here and there is left ventricle strain

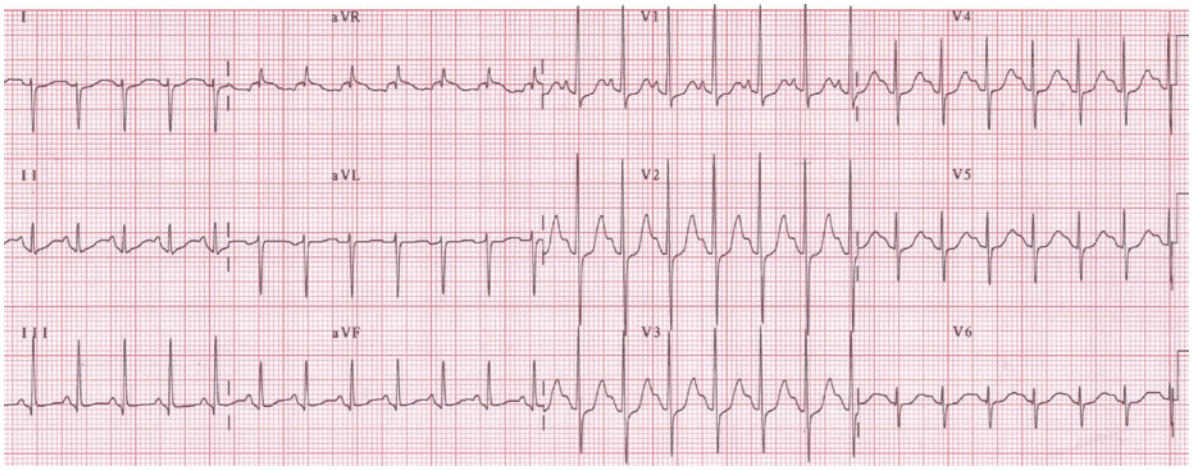


Fig. 18.34 One-week-old female with TGA "simple" (no VSD or PS are associated). On the first day of life, the ECG is usually silent. TGA may sometimes show disproportionate right axis deviation or RVH. In this case, the RVH is suspected from the positive T wave in V1

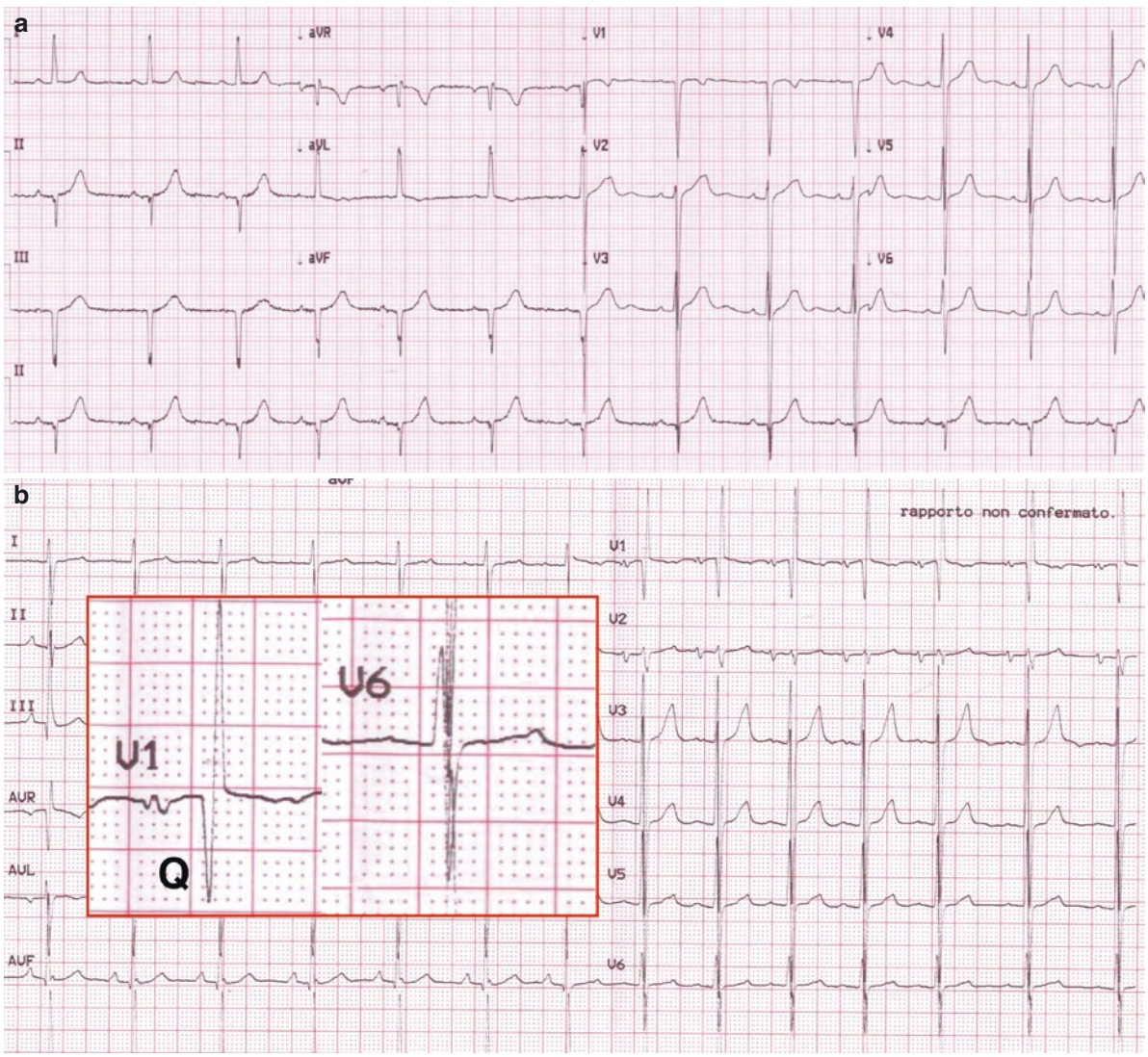


Fig. 18.35 (**a**) Ten-year-old male with CCTGA. Normally, the ventricular excitation starts at the middle third of the left aspect of the interventricular septum and spreads rightward, causing the Q wave in V6. That is why in CCTGA, where there is a ventricular inversion, the Q wave in V6 may be absent as in this case, while it may be present in V1. (**b**) Six-year-old female with CCTGA

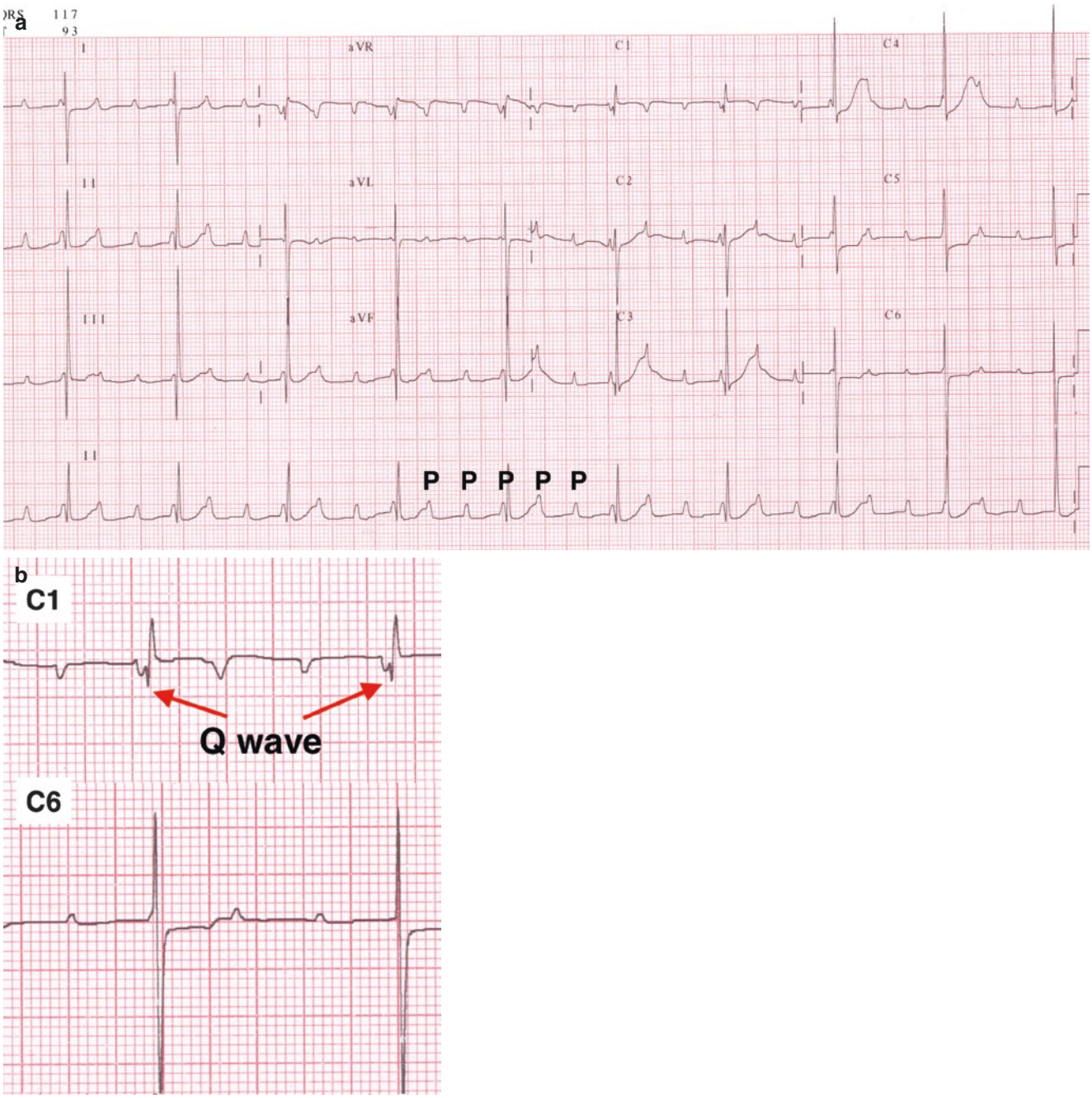


Fig. 18.36 (**a**) CCTGA in a neonate female. All the abovementioned traits are present (Q in V1, absence of Q in V6). A complete AV block can be noted, a not rare complication in CCTGA due to the abnormally elongated and tortuous course of the conduction system. (**b**) Close-up: in V1 a Q wave fused with the dissociated P wave can be noted

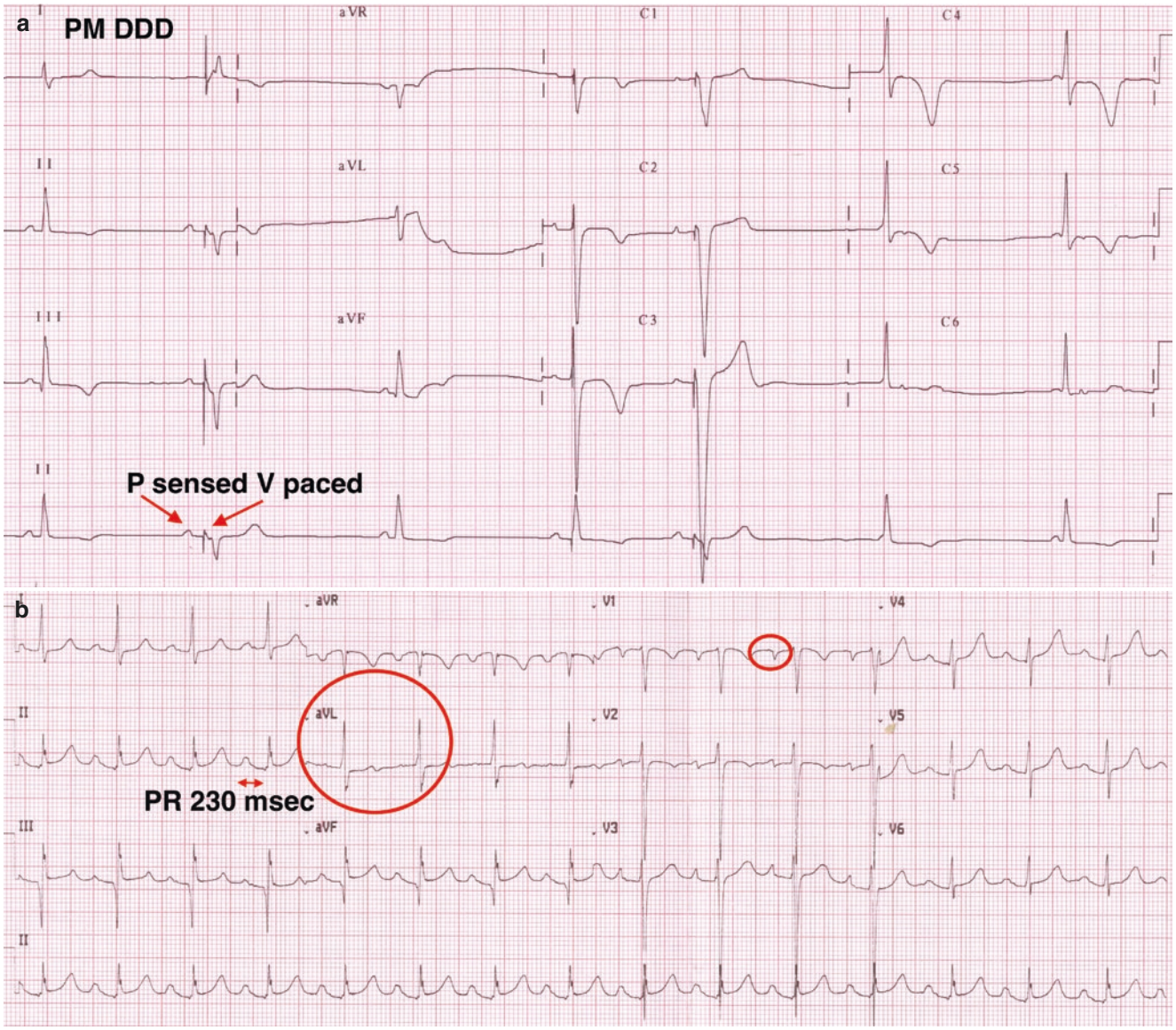


Fig. 18.37 (**a**) Twenty-one-year-old female with CCTGA and complete AVB who underwent PM implantation. The PM is working nicely in DDD mode, sensing the spontaneous P wave and tracking the ventricle. (**b**) Sixteen-year-old male with CCTGA and first-degree AV block. There is also a left atrial enlargement due to the moderate regurgitation of the systemic AV valve, which anatomically is a tricuspid valve

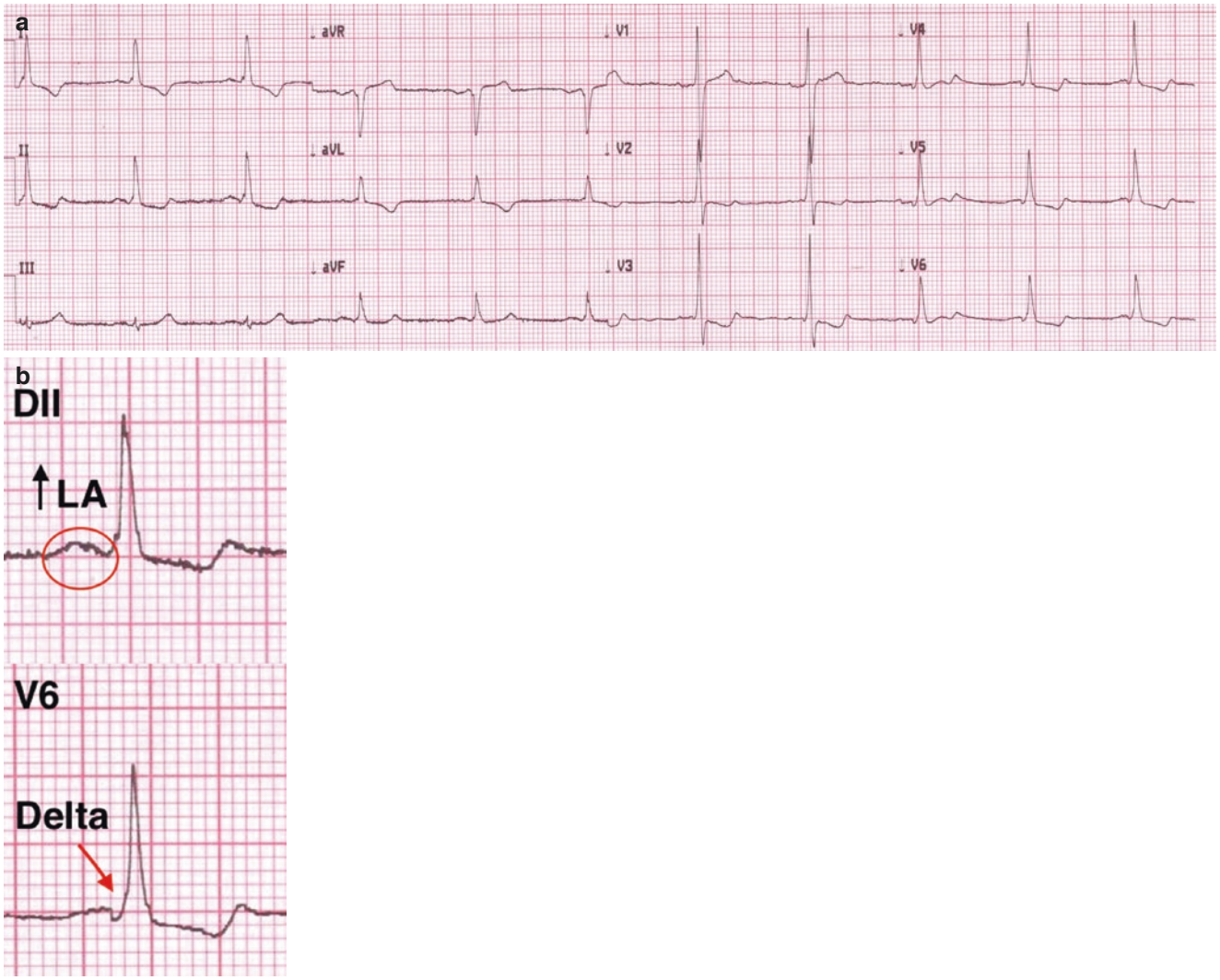


Fig. 18.38 (**a**) In CCTGA the systemic AV valve is a tricuspid valve, simply because the systemic/subaortic ventricle is a right ventricle. This disrupted anatomy is sometimes associated with an Ebstein-like tricuspid valve, a good substrate for accessory pathways. (**b**) This fifty-year-old woman presents a left-sided accessory pathway. The ST changes are secondary to the pre-excitation. A left atrial enlargement can be noted, due to the insufficiency of the Ebstein-like AV valve. CCTGA is the rare condition in which it is possible to find an AV block—"post-excitation"—and an accessory pathway ("pre-excitation") in the same patient

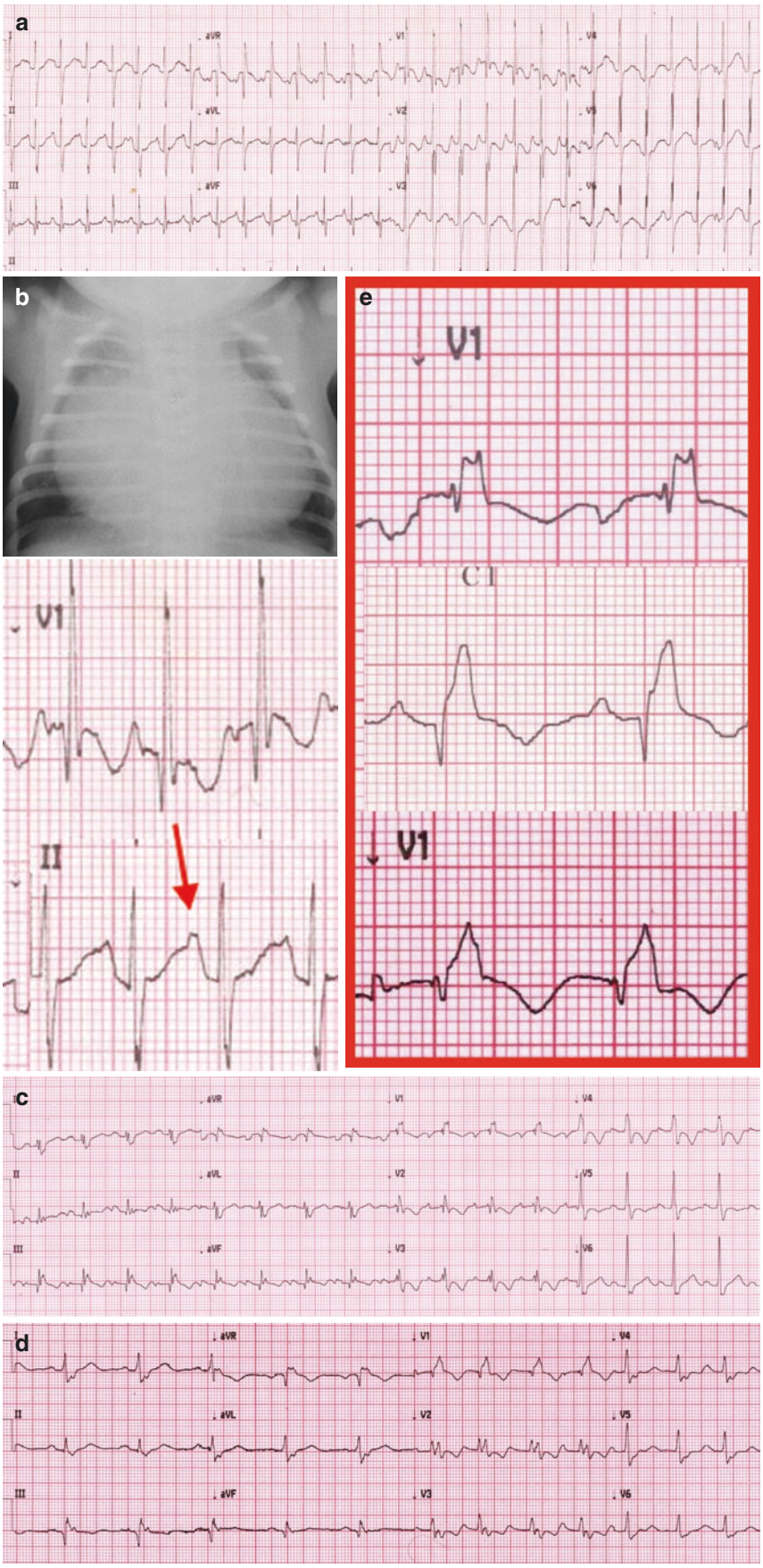
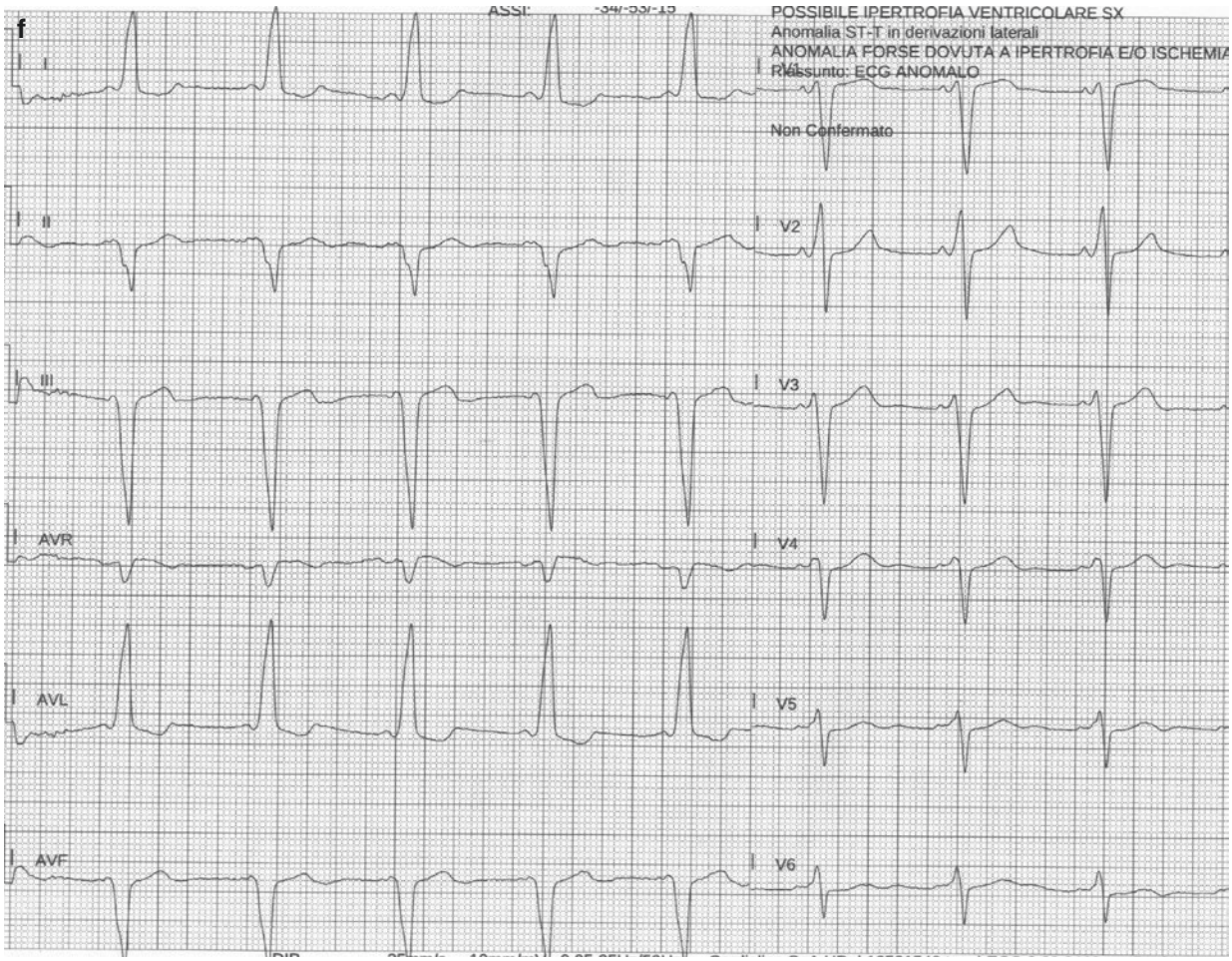
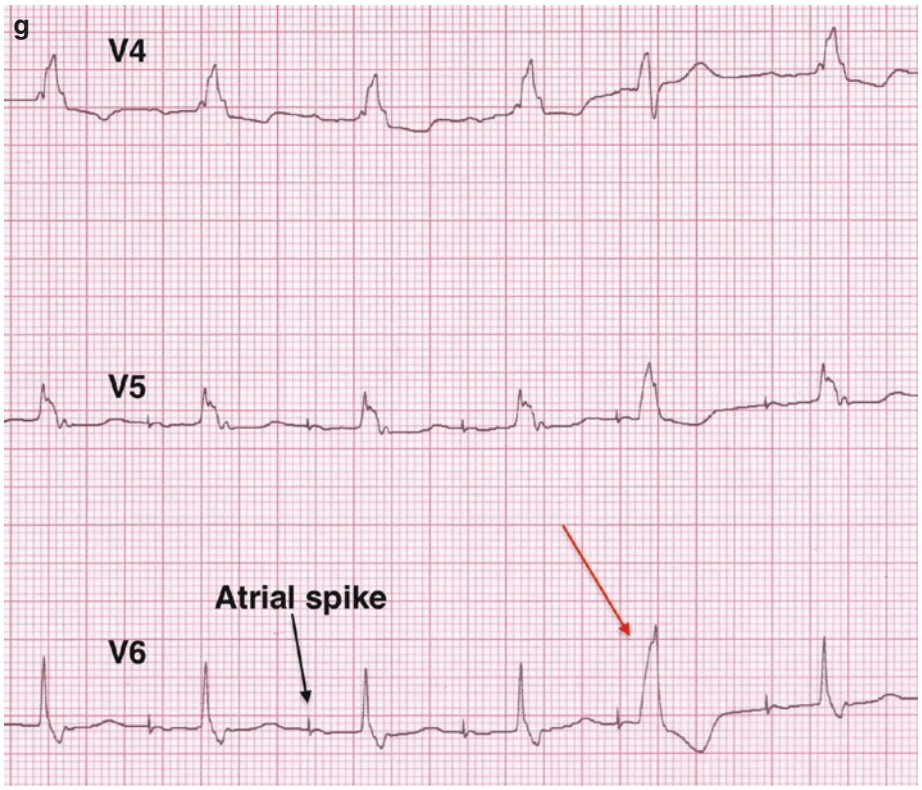


Fig. 18.39 Ebstein's anomaly (EA). As in the previous cases, in EA it is possible to find clues related to conduction disturbances and accessory pathways Mahaim fibers included, even in the same patient. As common traits, it is possible to find low voltages and polyphasic RBBB, mainly related to the anatomical disruption and to the fibrosis. Because of the ventricular atrialization and the tricuspid regurgitation, the enlarged right atrium may show large P waves. (**a**–**b**) Neonate with EA: note the RBBB and right atrial enlargement. (**c**) Fourteen-year-old female with EA: first-degree AV block. (**d**) Thirty-two-year-old male with EA. First-degree AV block and low voltages (**e**). As a rule, faced with an unusual or funny RBBB (fragmented and with low amplitude), an EA should always be ruled out. (**f**) Eight-year-old male with EA and WPW due to a right AP; the pre-excitation shadows all the typical traits of EA. (**g**) Forty-year-old male with EA with temporary atrial pacing: all but one atrial beats are conducted with long PR interval; only one (arrow) show a short PR and LBBB due to the conduction via Mahaim fibers (atrio-fascicular bypass)





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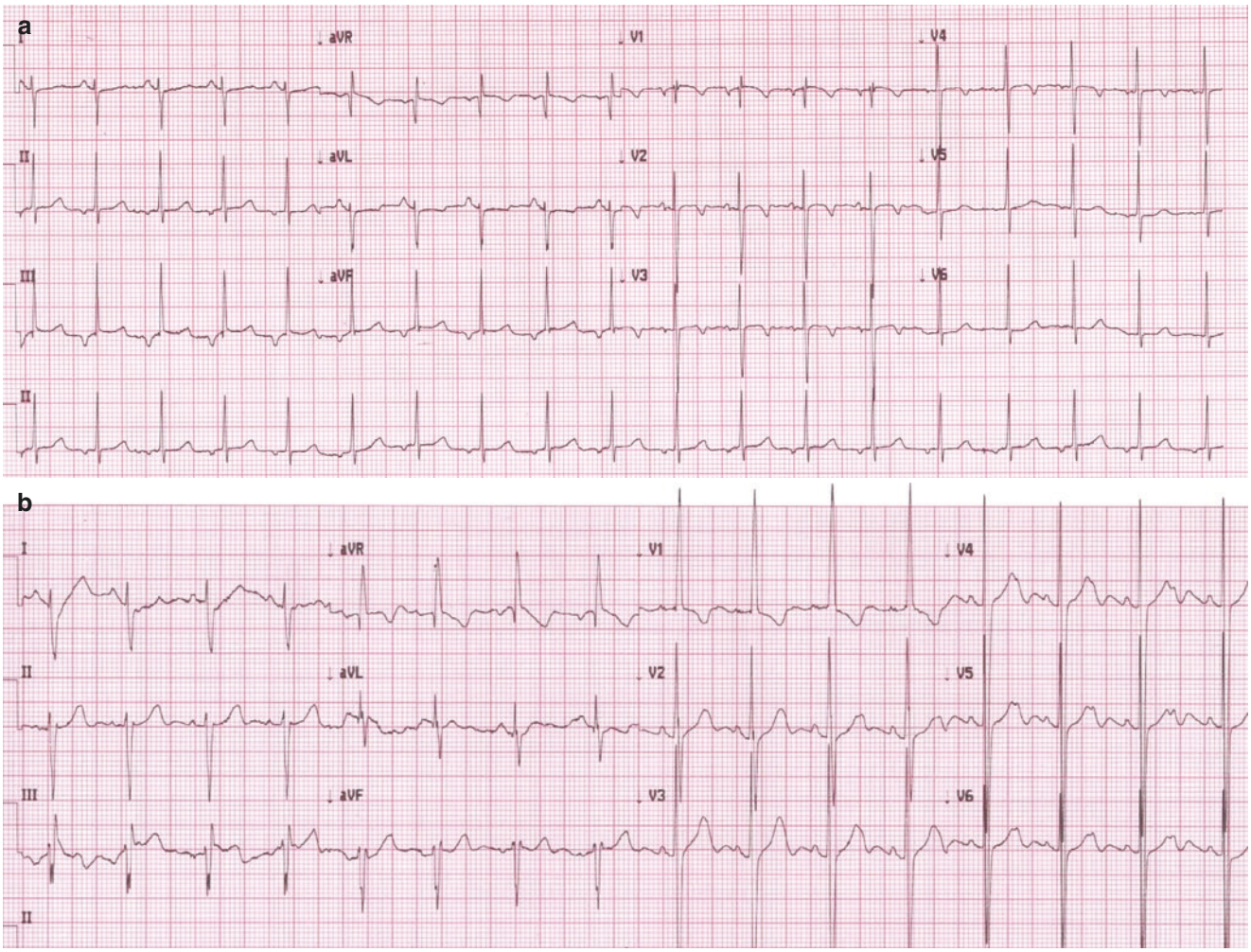

Fig. 18.40 Among heterotaxy syndromes, the left atrial isomerism (LAI) is not always related to complex CHD. (**a**) Two-year-old male with left isomerism and no CHD. The only clue of LAI is the ectopic atrial rhythm (low-right atrium) due to the absence of the sinus node (see text). (**b**) Three-year-old male with LAI and DORV. An RVH can be noted. The P morphology denotes an ectopic rhythm

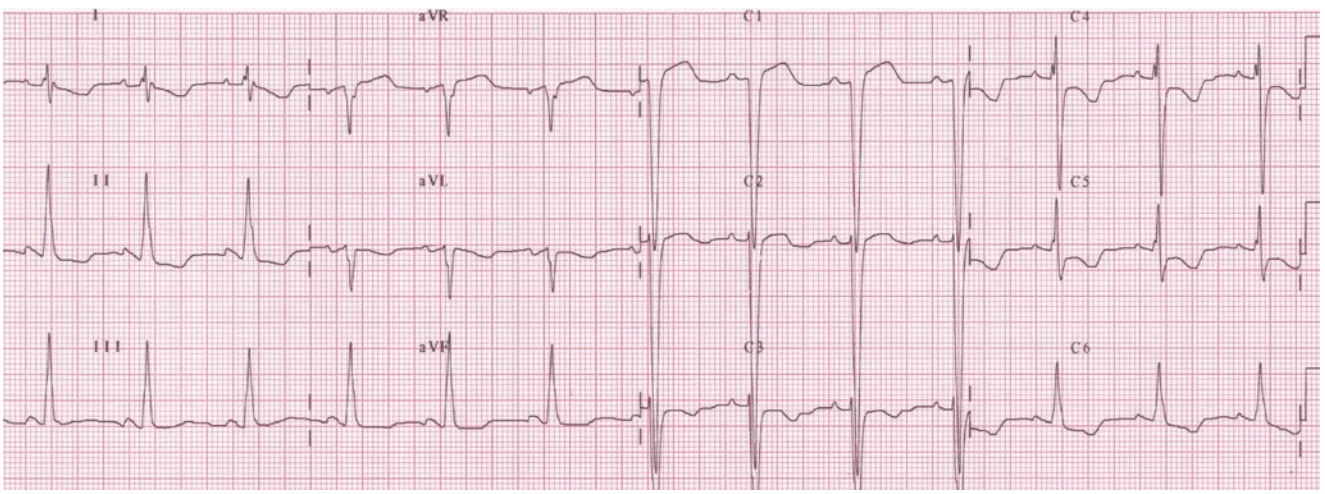

Fig. 18.41 In the right atrial isomerism (RAI), a CHD is always present, usually complex. Fifteen-year-old female with RAI and complex cyanotic CHD (DORV, TAPVR, aortic coarctation) who underwent palliation. There is a sinus rhythm. Note the widespread and impressive ST changes

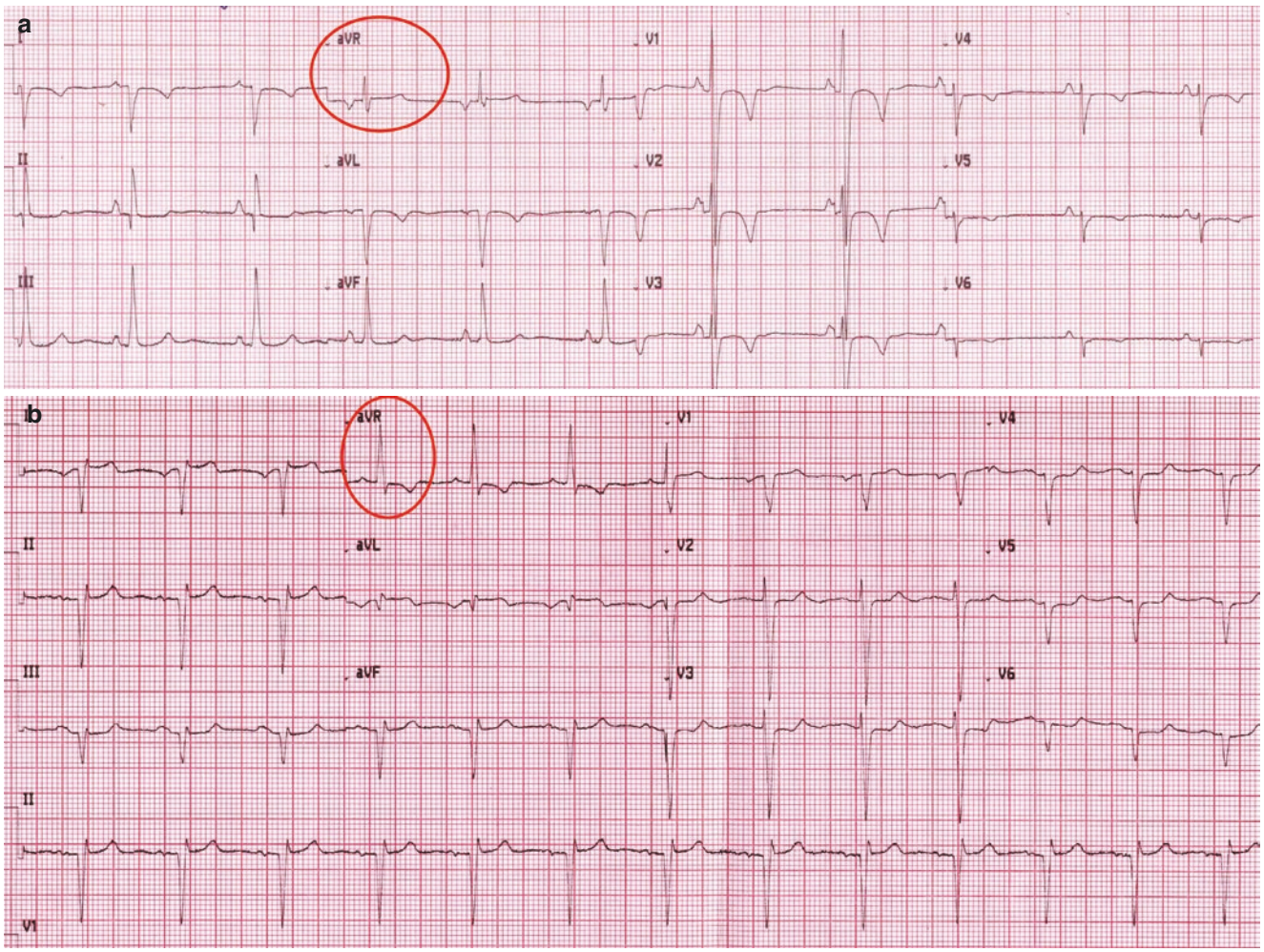

Fig. 18.42 (**a**) Twelve-year-old male with situs solitus, dextrocardia, and single ventricle who underwent "Fontan" palliation. The situs solitus can be inferred from the normal P wave. Conversely, the QRS complexes are poorly represented in the left precordial leads raising the suspicion of dextrocardia (**b**) Twenty-year-old female with situs inversus, dextrocardia, and VSD. In this case, the positive P wave in aVR (while it is positive in leads I and aVL) is diagnostic of situs inversus (once the possibility of lead misplacement has been ruled out)

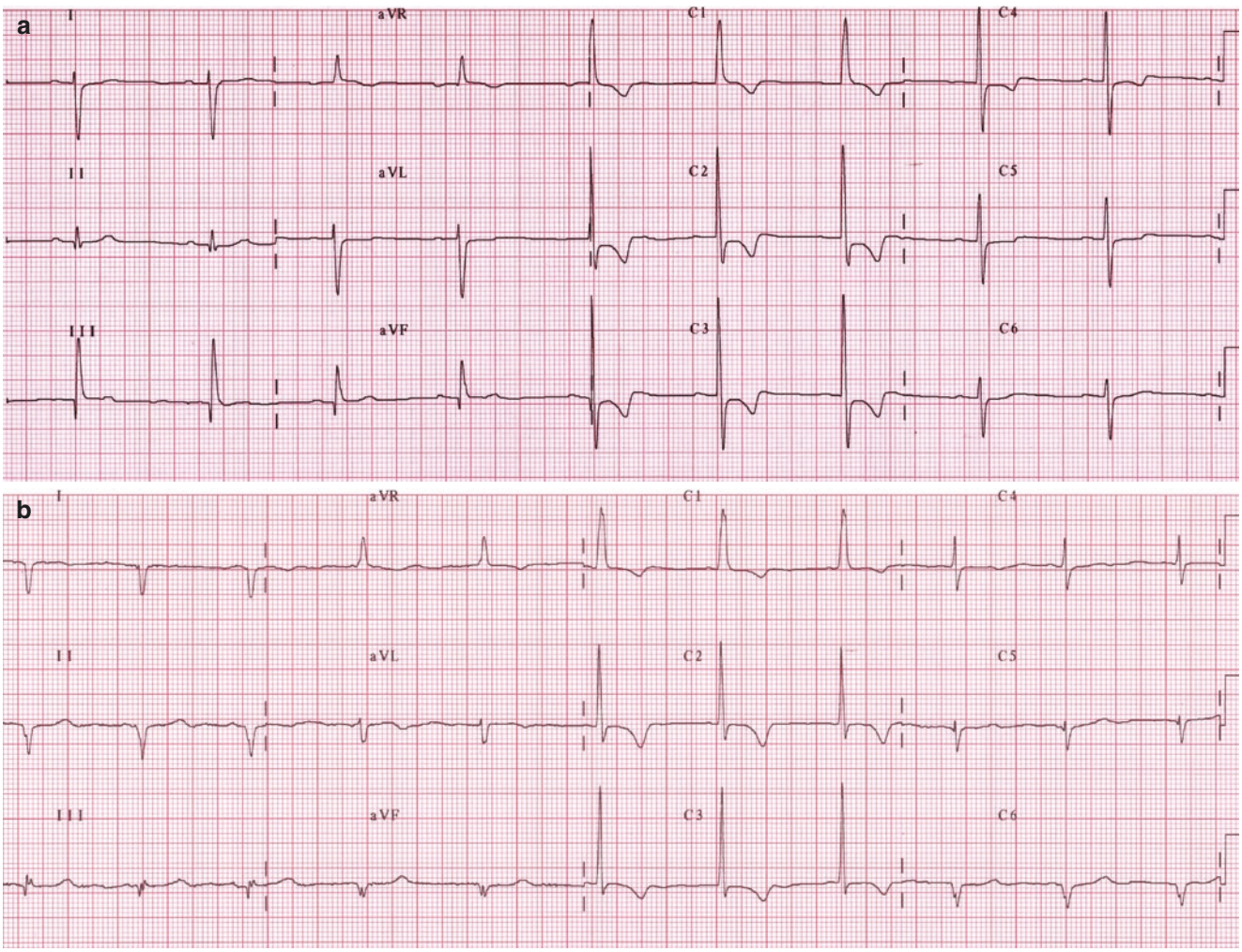

Fig. 18.43 (**a**) Thirty-four-year-old man with TGA who underwent a Senning operation. (**b**) Thirty-five-year-old woman who underwent a Mustard operation. The two operations developed in the late 1950s are very similar and consist in the detransposition at the level of the atrium. In the Senning operation, to achieve the venous baffle, an origami-style cutting and folding of the atrial tissue occurred; in the "simpler" Mustard operation, the baffles are created with a pantaloon-shaped patch. With such repairs, the surgeon diverts the systemic venous return to the left ventricle and the pulmonary venous returns to the right ventricle. So the circulation is restored but the right ventricle remains connected with the aorta and is then systemic. The ECG shows the hypertrophy and the dominance of the right ventricle. Until two decades ago, Mustard and Senning were the only options for TGA, before the advent of the arterial switch operation in the 1980s (Jatene procedure)

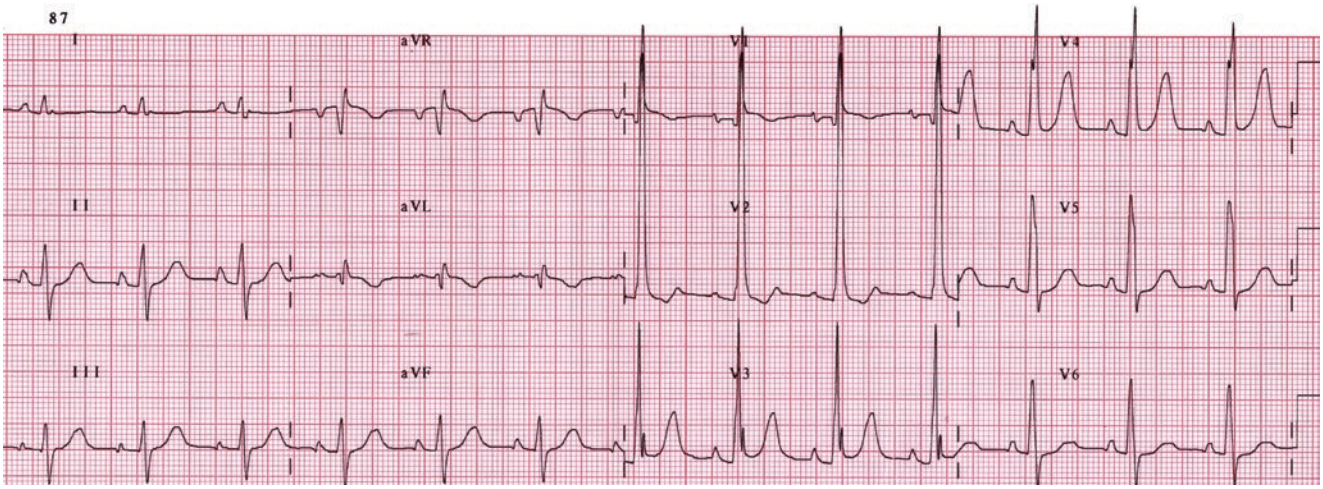

Fig. 18.44 Twelve-year-old male with single ventricle (right type) palliated with Fontan-like operation (total cavopulmonary connection, TCPC). TCPC is the third and last step which follows the neonatal BT shunt and the subsequent bidirectional Glenn operation (superior vena cava connected to the right pulmonary artery). A qR complex in V1 and large R in the right precordial leads can be noted

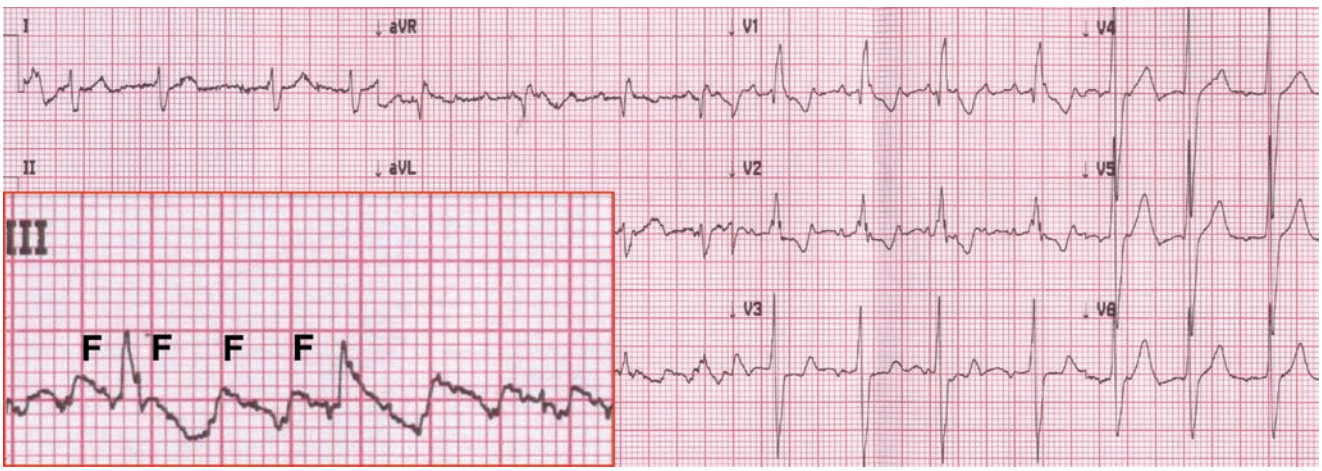

Fig. 18.45 Twenty-eight-year-old female with single ventricle who underwent a Fontan operation. The "old" Fontan (atrial-pulmonary connection) operation involved massive atrial scars prone to early arrhythmias such as intra-atrial reentrant tachycardia (IART), atrial flutter, and AF. In this case, the ECG shows an atrial flutter with variable ventricular rate. The F waves are clearly visible in the zoom window

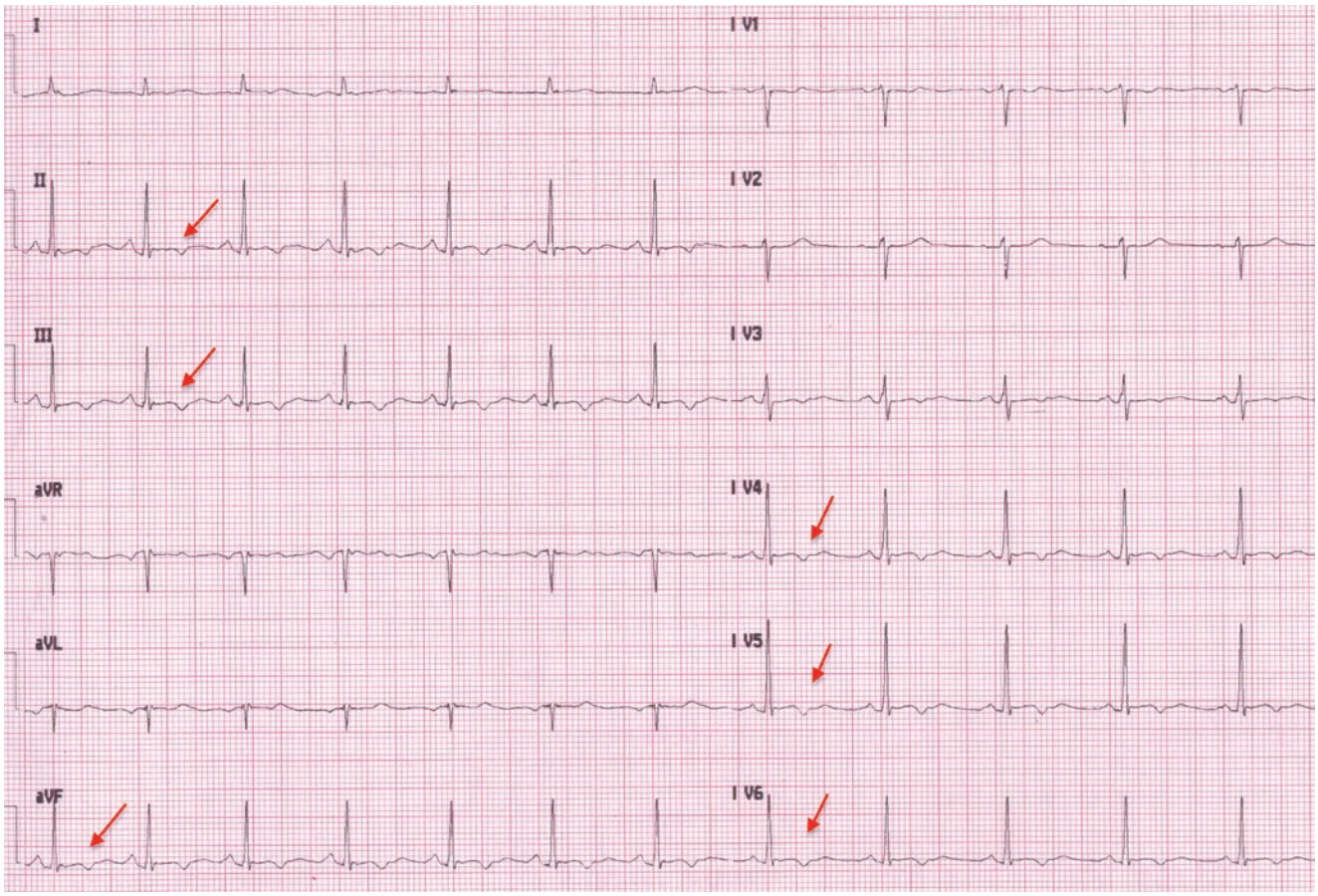

Fig. 18.46 Fourteen-year-old girl with MVP. The echo showed mild mitral insufficiency and normal left ventricle. The discrepancy between the widespread ST changes (well described in MVP) and the apparently normal ventricle on echocardiogram should suggest a form of malignant MVP which has to be excluded with cardiac MR [17]

References

1. Shashi V, Berry MN, Covitz W. A combination of physical examination and ECG detects the majority of hemodynamically significant heart defects in neonates with down syndrome. Am J Med Genet. 2002;108:205–8.
2. Khairy P, Marelli AJ. Clinical use of electrocardiography in adults with congenital heart disease. Circulation. 2007;116:2734–46.
3. Sung RJ, Tamer DM, Agha AS, et al. Etiology of the electrocardiographic pattern of "incomplete right bundle branch block" in atrial septal defect: an electrophysiologic study. J Pediatr. 1975;87(6 Pt 2):1182–6.
4. Rodriguez-Alvarez A, Martinez De Rodriguez G, Goggans AM, et al. The vectorcardiographic equivalent of the "crochetage" of the QRS of the electrocardiogram in atrial septal defect of the *ostium secundum* type: preliminary report. Am Heart J. 1959;58:388–94.
5. Katz LN, Wachtel H. The diphasic QRS type of electrocardiogram in congenital heart disease. Am Heart J. 1937;13:22–4.
6. Borkon AM, Pieroni DR, Varghese PJ, Ho CS, Rowe RD. The superior QRS axis in *ostium primum* ASD: a proposed mechanism. Am Heart J. 1975;90:215–21.
7. Rasmussen K, Sorland SJ. Prediction of right ventricular systolic pressure in pulmonary stenosis from combined vectorcardiographic data. Am Heart J. 1973;86:318–28.
8. Glancy DL, Morrow AG, Simon AL, Roberts WC. Juxtaductal aortic coarctation. Analysis of 84 patients studied hemodynamically, angiographically, and morphologically after age 1 year. Am J Cardiol. 1983;51:537–51.
9. Hernandez FA, Rochkind R, Cooper HR. The intracavitary electrocardiogram in the diagnosis of Ebstein's anomaly. Am J Cardiol. 1958;1:181–90.
10. Bialostozky D, Medrano GA, Munoz L, Contreras R. Vectorcardiographic study and anatomic observations in 21 cases of Ebstein's malformation of the tricuspid valve. Am J Cardiol. 1972;30:354–61.
11. Ho SY, Goltz D, McCarthy K, et al. The atrioventricular junctions in Ebstein malformation. Heart. 2000;83:444–9.
12. Victorica BE, Miller BL, Gessner IH. Electrocardiogram and vectorcardiogram in ventricular inversion (corrected transposition). Am Heart J. 1973;86:733–44.
13. Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. Circulation. 1983;67:1374–7.
14. Gillette PC, Busch U, Mullins CE, McNamara DG. Electrophysiologic studies in patients with ventricular inversion and "corrected transposition". Circulation. 1979;60:939–45.
15. Perry LW, Scott LP. Anomalous left coronary artery from pulmonary artery: report of 11 cases—review of indications for and results of surgery. Circulation. 1970;41:1043–52.

16. Burch GE, Depasquale NP. The anomalous left coronary artery: an experiment of nature. Am J Med. 1964;37:159–61.
17. Lancellotti P, Garbi M. Malignant mitral valve prolapse. Substrates to ventricular remodeling and arrhythmias. Circ Cardiovasc Imaging. 2016;9:e005248.

Suggested Reading

Triedman JK, Newburger JW. Trends in congenital heart disease: the next decade. Circulation. 2016;133:2716–33.
Khairy P, Van Hare GF, Balaji S, Berul CI, Cecchin F, Cohen MI, Daniels CJ, Deal BJ, Dearani JA, Nd G, Dubin AM, Harris L, Janousek J, Kanter RJ, Karpawich PP, Perry JC, Seslar SP, Shah MJ, Silka MJ, Triedman JK, Walsh EP, Warnes CA. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: developed in partnership between the pediatric and congenital electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European heart rhythm association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Can J Cardiol. 2014;30:e1–e63.
Bronzetti G, Mariucci E, Bonvicini M, Picchio FM. The ECG in pediatric patients: what the cardiologist needs to know. G Ital Cardiol (Rome). 2011;12:408–18.

The Athlete's Heart

19

Regular exercise activity, especially the aerobic type, is responsible for structural and functional cardiac remodeling with a variety of associated ECG patterns, known as *athlete's heart* [1, 2].

The ECG patterns that are encountered in the heart of the athlete could be divided into two types: exercise related (normal variants, proportional to the type, and intensity of physical exercise) and non-exercise related (abnormal patterns) (Table 19.1).

A short summary of the major ECG changes in healthy athletes aged 14–35 years is provided below:

- Up to 80% of trained athletes have at least one of the following alterations: 1) sinus bradycardia and atrial or junctional escape rhythms, 2) first-degree atrioventricular block, 3) high voltages (left ventricular hypertrophy pattern), 4) early repolarization pattern, and 5) incomplete RBBB.
- The above ECG changes should be distinguished from those less common ones which are not exercise related. The latter are present in less than 5% of the athletes and should prompt further investigation. They are: (1) wave T inversion (> 1 mm in at least two contiguous leads), (2) ST-segment depression (> 0.5 mm in at least two contiguous leads), (3) pathologic Q waves, (4) atrial enlargement, (5) left axis deviation/left anterior fascicular block and right axis deviation, (6) signs of left ventricular hypertrophy (LVH), (7) complete RBBB or complete LBBB, (8) long or short QT, (9) ventricular pre-excitation, (10) Brugada-like pattern, and (11) tachyarrhythmias [3, 4].

Table 19.1 The athlete's ECG

Normal findings	Pathological findings
Sinus bradycardia	Marked sinus bradycardia[a]
Respiratory sinus arrhythmia	Atrial tachyarrhythmias
Ectopic atrial rhythm/junctional	PVCs[b]/ventricular arrhythmias
I/II degree AVB Mobitz 1	Ventricular pre-excitation
Incomplete RBBB	II degree AVB Mobitz 2/III degree AVB
Convex ST elevation con inverted T waves in right precordial leads (V1–V4)[d]	Complete LBBB/intraventricular conduction delay with QRS > 140 ms
Benign early repolarization	LVH + no voltage-related signs[e]/RVH
Isolated voltage criteria for LVH[e]	ST depression ≥0.5 mm in two or more leads
	Brugada-like pattern[f]
	Long/short QT interval[g]

AVB, atrioventricular block; *RBBB*, right bundle branch block; *LVH*, left ventricular hypertrophy; *VEBs*, ventricular ectopic beats; *LBBB*, left bundle branch block; *RVH*, right ventricular hypertrophy [R-V1 and S-V6 > 98° centile + right axis deviation (axis > 120°)]

[a]HR < 30/min or pause ≥3 sec

[b]≥ 2 PVCs/10 sec registration

[c]Short PR (childhood and adolescence <80 ms) + delta wave

[d]African ethnicity

[e]Sokolow-Lyon voltage criteria (S-V1 and R-V6 > 98th centile, poor specificity and sensitivity in children) without left axis deviation (−30° < axis < −90°), left atrial enlargement, strain-type ST alterations (negative and asymmetrical T waves ± ST depression, specific sign in the pediatric age), inverted T waves or pathological Q waves [deep >3 mm or length > 40 ms in two or more leads (but lead III and aVR)]

[f]ST elevation (coved, convex, saddle shaped) anterior precordial leads (V1–V3) ± inverted T waves in two or more leads (V1–V3)

[g]Age-based reference nomograms (2-1)

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19.1 Exercise-Related ECG Changes

Sinus bradycardia is quite common in the athlete's heart in view of the vagal hypertonia. Its severity could be directly proportional to the degree of training, although there is wide interindividual variability according to the achieved training level due to genetic influences. A slow heartbeat with rates around 40–45 bpm is considered normal in trained athletes, and in highly trained athletes, the heart rate could be as low as 35 bpm in physiological conditions. At rest or during sleep, pauses of up to 3 s can normally occur.

Changes in AV conduction are common in view of the increased parasympathetic and decreased sympathetic tones. First-degree AV block is present in approximately 35% of athletes, and Wenckebach second-degree (Mobitz 1) AV block is encountered in up to 10% of athletes. The above-described changes in AV conduction can often be reduced or eliminated by hyperventilation or physical exercise (an exercise stress test could be helpful to demonstrate this). If symptoms occur (dizziness, presyncope, or syncope), regardless of the existing ECG pattern or evidence of Mobitz 2 second-degree or third-degree AV block, they should be promptly investigated.

Signs of LVH (without other sings of ventricular overload) can be seen in up to 40–60% of the athletes.

The presence of additional minor ECG abnormalities (e.g., left atrial enlargement, left axis deviation, ST-T alterations or pathologic Q waves) can warrant further testing with an echocardiography.

Early repolarization pattern *(*ERP*)* is a common ECG variant. It is present in 1–5% of the general population and is very common in young athletes, with a prevalence rate of 25–40% (up to 50–80% in highly trained athletes, mostly in leads V2–V5). The ERP pattern is more frequent among young black adults, especially males. In black athletes, ST-segment elevation associated with T wave inversion in leads V1–V4 could be considered a normal variant. The ERP is characterized by an upward concave elevation ≥0.1 mV (up to 4 mm) of the J point in at least two contiguous leads, often with associated tall T waves. It is manifested by an apparent slowing of the inscription of the waveform at the end of the QRS that merges with the ST segment of the complex ("slurring") or as an end-QRS notch that occurs on the downslope of an R wave ("notching"). It is due to hypervagotonia and bradycardia and is usually attenuated or vanishes in the presence of tachycardia. Moreover, the J- point/ST-segment elevation often disappears with detraining.

The prevalence of incomplete RBBB (QRS < 120 ms) is approximately 35–50% in athletes, compared to 5–10% in young healthy adults. This pattern is more common in endurance sports, which tend to increase right ventricular mass and volume. A fixed splitting of the second heart sound (S2) in the above situation warrants further echocardiographic evaluation in order to rule out a possible atrial septal defect (ASD) or partial pulmonary anomalous venous return (PPAVR). An ECG pattern with a T wave inversion that extends out to V4 in patients aged >12 years could be explicative of cardiomyopathy as ARVC or HCM. A "nonclassic" morphology of RBBB warrants the use of V1 and V2 leads recorded in the second and third intercostal spaces to rule out a Brugada pattern.

19.2 Non-exercise-Related ECG Changes

A T wave inversion >1 mm in at least two contiguous leads is not rare: it can be found in approximately 2–3% of a young healthy population. Nevertheless, its presence in leads other than III, aVR, and V1 may reflect an underlying ischemic heart disease, volume or pressure overload (aortic valve disease, systemic hypertension, congenital heart disease), or a cardiomyopathy (especially HCM and ARVC) and thus should prompt further diagnostic evaluation (echocardiography, ECG Holter, or, if necessary, MRI). Inverted T waves in young athletes may represent the first and only sign of an underlying cardiomyopathy, in the absence of any other features and before structural changes in the heart can be detected on echocardiography. There are HCM-related mutations in which the ECG changes precede the echocardiographic phenotype; on the other hand, there are cardiomyopathy and CHD that can be overlooked from a not careful echocardiographic evaluation (i.e., apical cardiomyopathy, ACAOS, or other coronary anomalies).

An *ST-segment depression* > 0.5 mm in two or more leads is rare and is normally associated with a T wave inversion (overload/strain).

The Q waves are defined pathological if their depth exceeds 25% of the depth of the associated R wave or their duration is >40 ms in two contiguous leads (except D3 and aVR). As a rule, pathological Q waves caused by hypertrophic cardiomyopathy are more than 3 mm deep and/or with a duration of >40 ms.

A right atrial enlargement is defined by a P wave amplitude >2.5 mm in any lead. Even taller T waves can often be

seen in young people (up to 15%); they are less frequent after the age of 18–20 years (approximately 1%).

A left atrial enlargement is characterized by a negative component of the T wave >40 ms in leads V1 or V2, with the amplitude of its negative component >1 mm and an overall duration of the P wave >120 ms. According to the above-mentioned criteria, it is present in up to 12–14% of children and 1–4% of people aged over 18–20 years.

The mean frontal plane *QRS axis is rightward in children* and tends to normalize in adolescents. A normal range of QRS axis is considered between −30° and +120°. A QRS axis of less than −30° should be considered abnormal and needs further evaluation, as well as the presence of a left anterior fascicular block. A right axis deviation is frequently encountered in young people (up to 15–20% of cases), while it is quite rare after the age of 20 and in the latter case may reflect an underlying respiratory disorder or pulmonary vascular disease. A left axis deviation might be seen in young healthy adults (e.g., 8% of healthy airplane pilots in their thirties and forties).

The ECG changes indicative of right ventricular hypertrophy are not so frequent in athletes and young people >16 years of age (0.5%). They are characterized by right axis deviation and the following voltage criteria: R wave in V1 > 7 mm with R/S ratio > 1 in V1 or the Sokolow-Lyon criteria for the right ventricle (R wave in V1 + S wave in V5 or V6 > 10.5 mm). Right ventricular hypertrophy is often associated with repolarization abnormalities in the right precordial leads and/or right atrial enlargement. Its differential diagnosis includes RBBB, previous posterior myocardial infarction, and congenital heart disease, as well as with a normal variant which is due to the apical rotation of the heart in a horizontal plane.

Complete bundle branch blocks (RBBB, LBBB with QRS > 120 ms) and hemiblocks are quite rare before the age of 40 (0.4–1% of athletes); similarly, the prevalence of left anterior hemiblock is low (0.5–1% in young people). A combination of RBBB + left anterior hemiblock or trifascicular blocks is also infrequent. Complete heart blocks should be considered pathologic until proved otherwise, as they can reveal a previous or latent cardiac disease (e.g., ischemia, cardiomyopathy, hypertensive heart disease, myocarditis, Chagas disease, sarcoidosis, corrected CHD) and warrant further work-up with echocardiography, exercise stress test, ECG Holter, or MRI. If the results are normal, the prognosis will be excellent, but a cautious follow-up of the patient is still required.

The upper limit of QTc, historically established and universally accepted, is 440 ms, with age and gender dependent deviations. Nevertheless, in view of its statistical and demographic nature, this limit is reached by a considerable number of healthy people (>2%), while the prevalence of long QT syndrome is <1/2500. It is thus of vital importance to adopt less restrictive upper limits of QT, especially in a sports-related setting, in order to reduce the number of false-positive results with their dramatic consequences in terms of additional evaluations and a lifetime ban from sports. A series of recent studies with a large number of athletes enrolled have tried to redefine existing reference ranges (not only for QTc but also for axis deviation and atrial enlargement) taking into account its impact on sensitivity and specificity [5]. Their results have demonstrated a marked advantage for specificity without detecting any important negative effect on sensitivity. So, in view of the above considerations, in the newly published guidelines, the upper limit of QTc has been set at 470 ms in males and 480 ms in females [6]. In athletes with severe bradycardia a possible underestimation (by overcorrection) of QTc calculated with Bazett's formula should be taken into consideration, and according many the Fredericia's formula is more accurate.

The ECG definition of the short QT syndrome (SQTS) is not clear, due to the rarity of this "young" entity. QTc less <340 ms warrants further evaluation, and QTc interval ≤ 320 ms is considered diagnostic, regardless the result of the genetic test.

Brugada-like ECG pattern is characterized by a J elevation >2 mm with a descending ST segment followed by a negative T wave in at least two of the three right precordial leads (V1–V3). As the ERP pattern is quite common among athletes, the suspicion of Brugada is inevitable in a considerable number of cases. If in doubt, it is advisable to shift the right precordial leads to a higher position (up to the second intercostal space above normal). The ECG changes rapidly, even in a couple of hours. It should be kept in mind that hyperthermia, hypokalemia, and vagal hypertonia induced by prolonged physical activity are all potential triggers for tachyarrhythmias in Brugada syndrome.

The presence of *tachyarrhythmias* (SVT, AF, Aflutter) should be considered pathologic, as well as ventricular couplets, triplets, or nonsustained VT. The new Seattle criteria consider the presence of at least two PVC on a ten-second ECG rhythm strip abnormal.

The definition of pre-excitation requires a distinct delta wave in addition to a short PR interval.

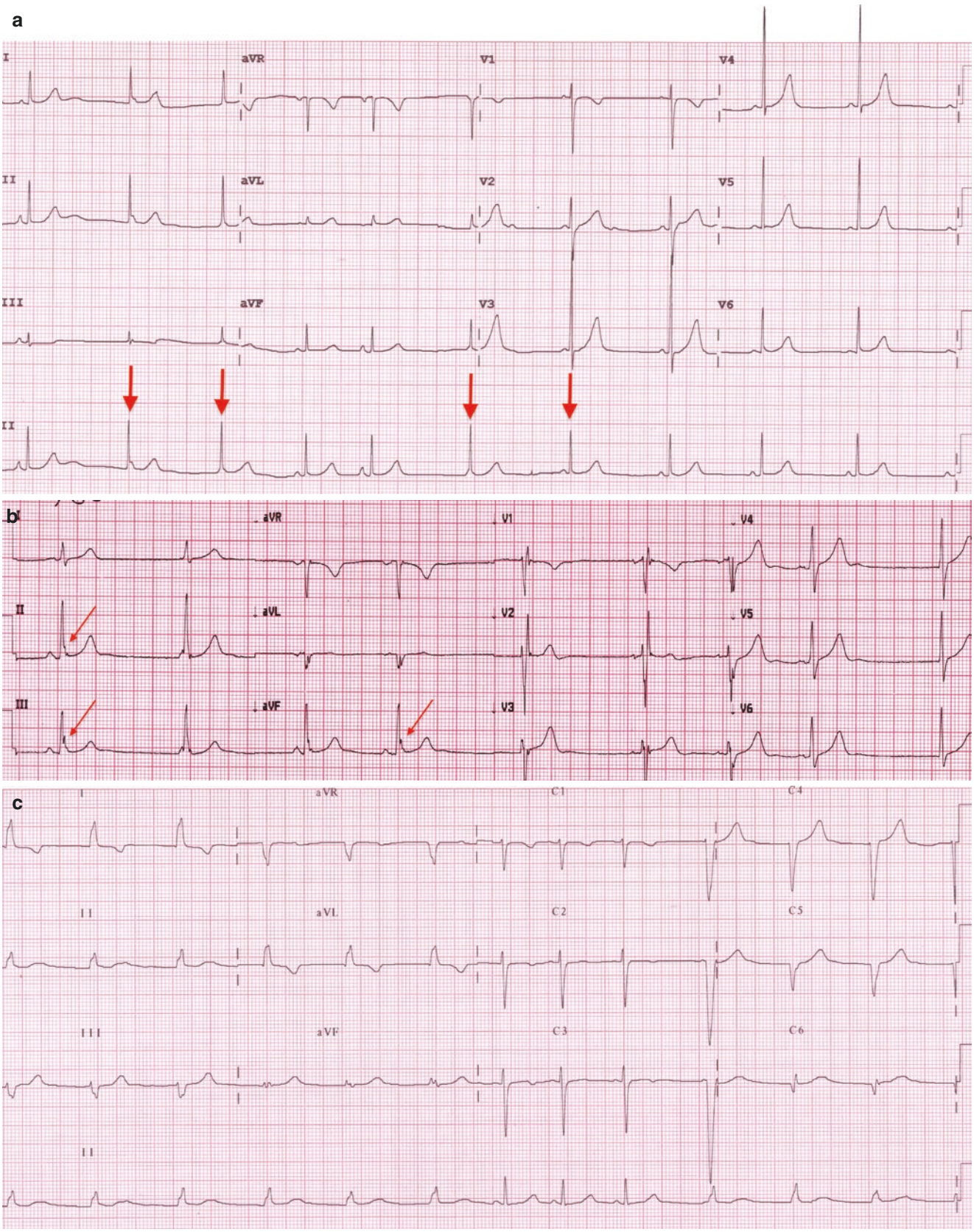

Fig. 19.1 (**a**) 12-year-old male soccer player. Sinus bradycardia, phasic arrhythmia, and junctional escape rhythm. Dissociated P can be seen (*arrows*). (**b**) As in the previous figure, this is a 17-year-old trained athlete. Note the sinus bradycardia, the junctional beats, the incomplete RBBB, and the benign early repolarization pattern in the inferior leads (BER, *arrow*). (**c**) 16-year-old trained bicycle racer. Note the phasic arrhythmia; during the bradycardic phase, there is an idioventricular escape rhythm with LBBB morphology

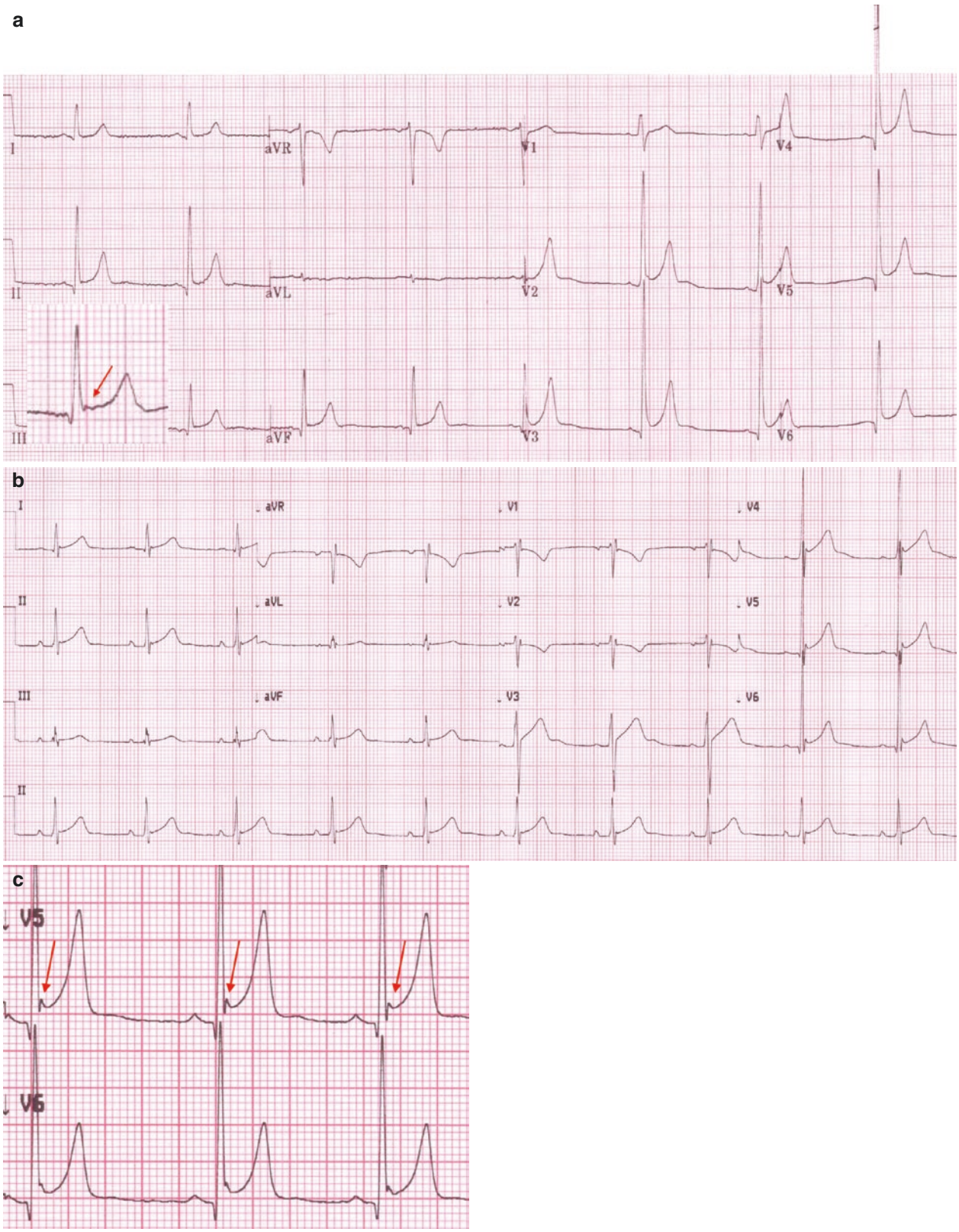

Fig. 19.2 (**a**) 15-year-old well-trained male. The BER pattern can be noted. The features typical of BER are concave ST elevation, especially in the precordial leads (V2–5), notching or slurring of the J point, ST elevation less than 25% of the T wave height in the precordial leads, and large and peaked nonsymmetrical T waves. (**b**) As in the previous figure, there is a clear BER. In V5 a "fish hook," J point can be noted. As a rule, BER is typical of young people <40 years old. To differentiate BER from STEMI, by definition, the ST elevation and J point become less visible as the heart rate increases. (**c**) Close-up

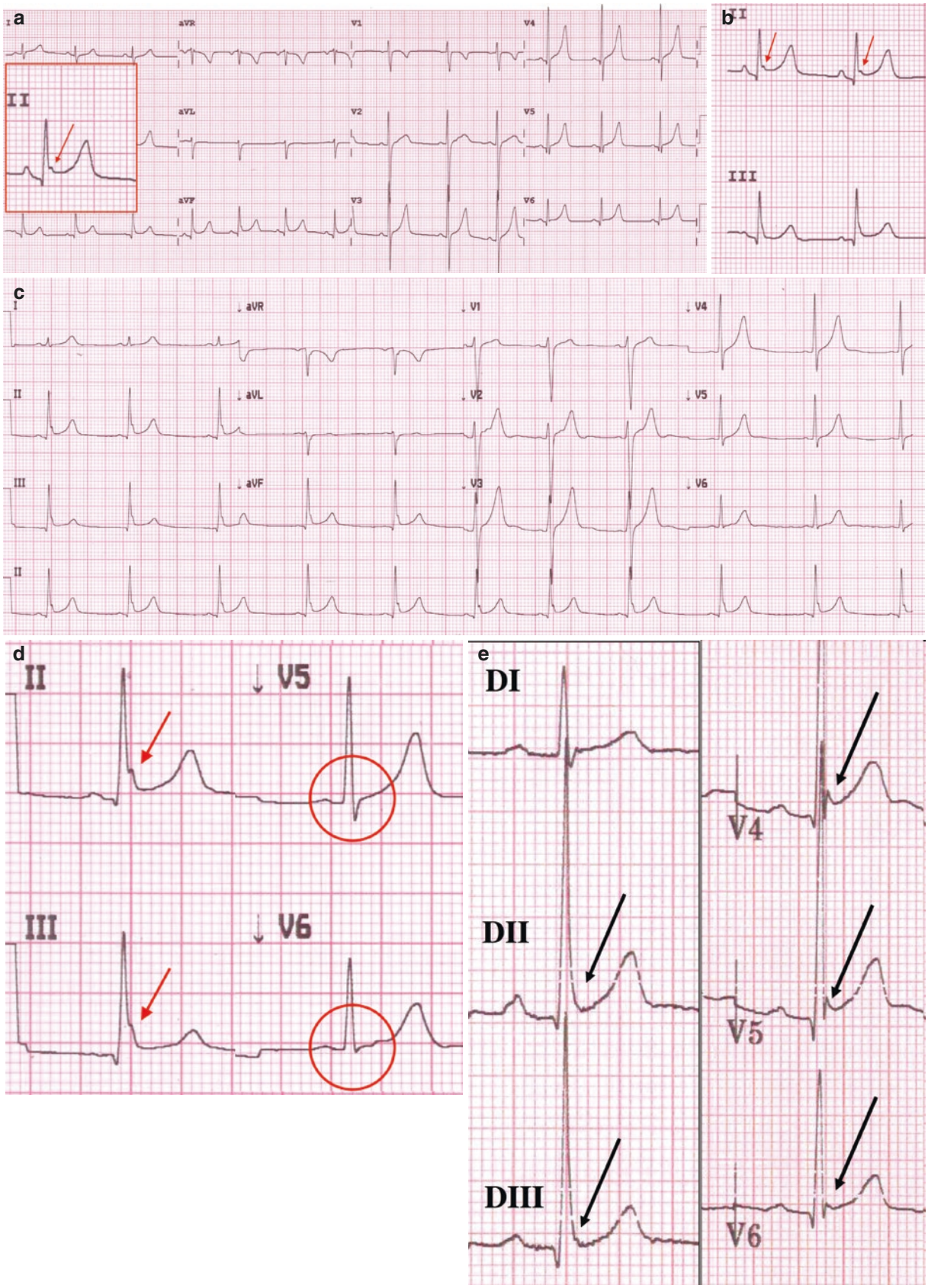

Fig. 19.3 (**a**) 8-year-old female with BER and J point notching in the inferior leads. (**b**) Close-up. (**c**) 18-year-old male. (**d**) The BER is more prominent in the inferior leads than in the precordial leads. (**e**) In this case the BER is more evident in the precordial leads than in the inferior leads (see Fig. 19.2)

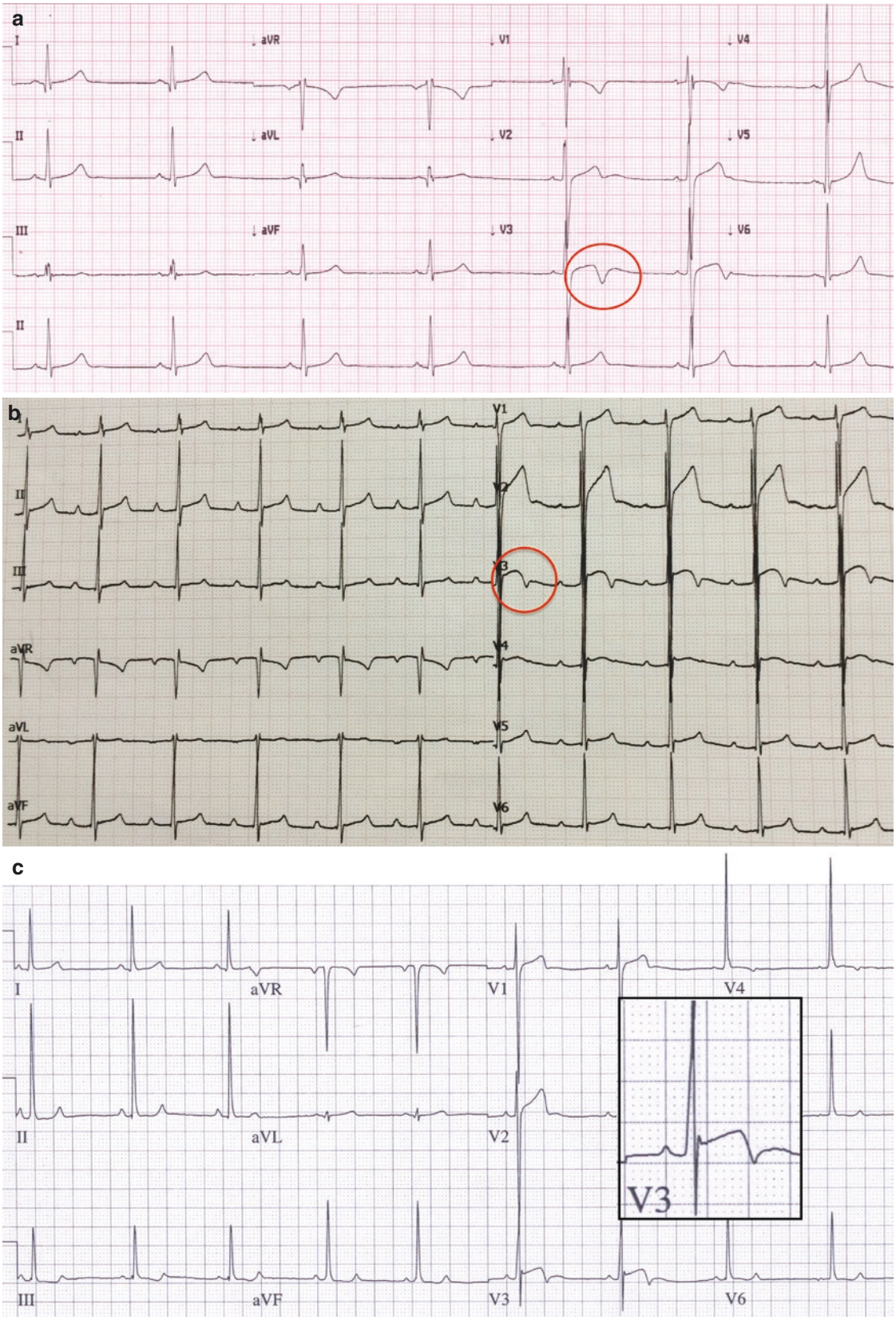

Fig. 19.4 (**a**) 12-year-old black male soccer player. (**b**) 16-year-old black basketball player. (**c**) 17-year-old black basketball player. They all show the black/African variant repolarization changes in V3, characterized by domed ST-segment elevation and T wave inversion, which differs from the Brugada pattern in which the high takeoff and downsloping of the ST segment precedes inverted T waves. In this benign pattern, the "domed" ST elevation followed by T wave inversion in leads V1–V4 is considered a normal variant. More than 60% of black athletes show ST-segment elevation and up to 25% show T wave inversions

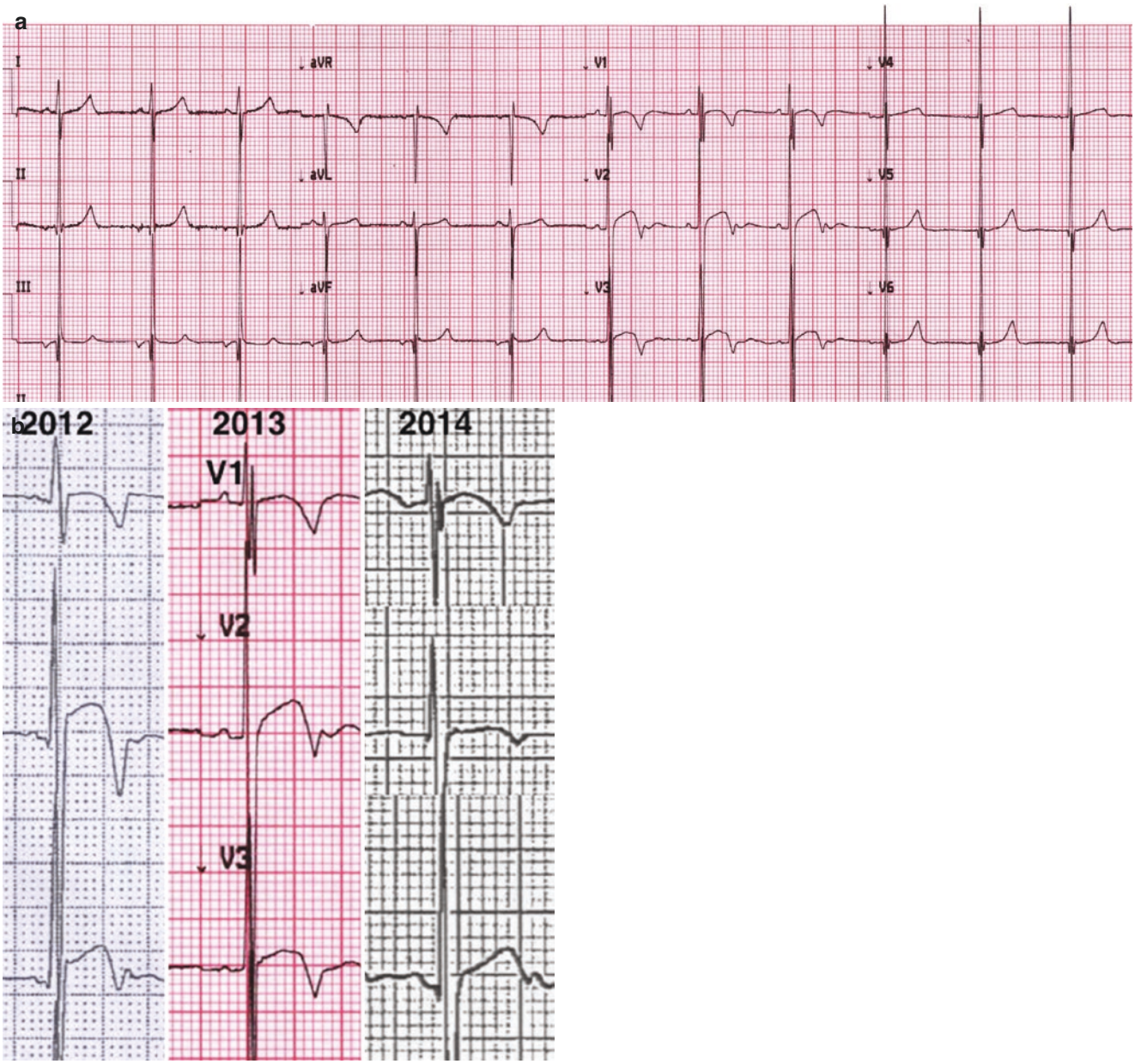

Fig. 19.5 (**a**) 14-year-old male swimmer. (**b**) The domed ST elevation followed by T wave inversion tends to be less prominent 2 years later

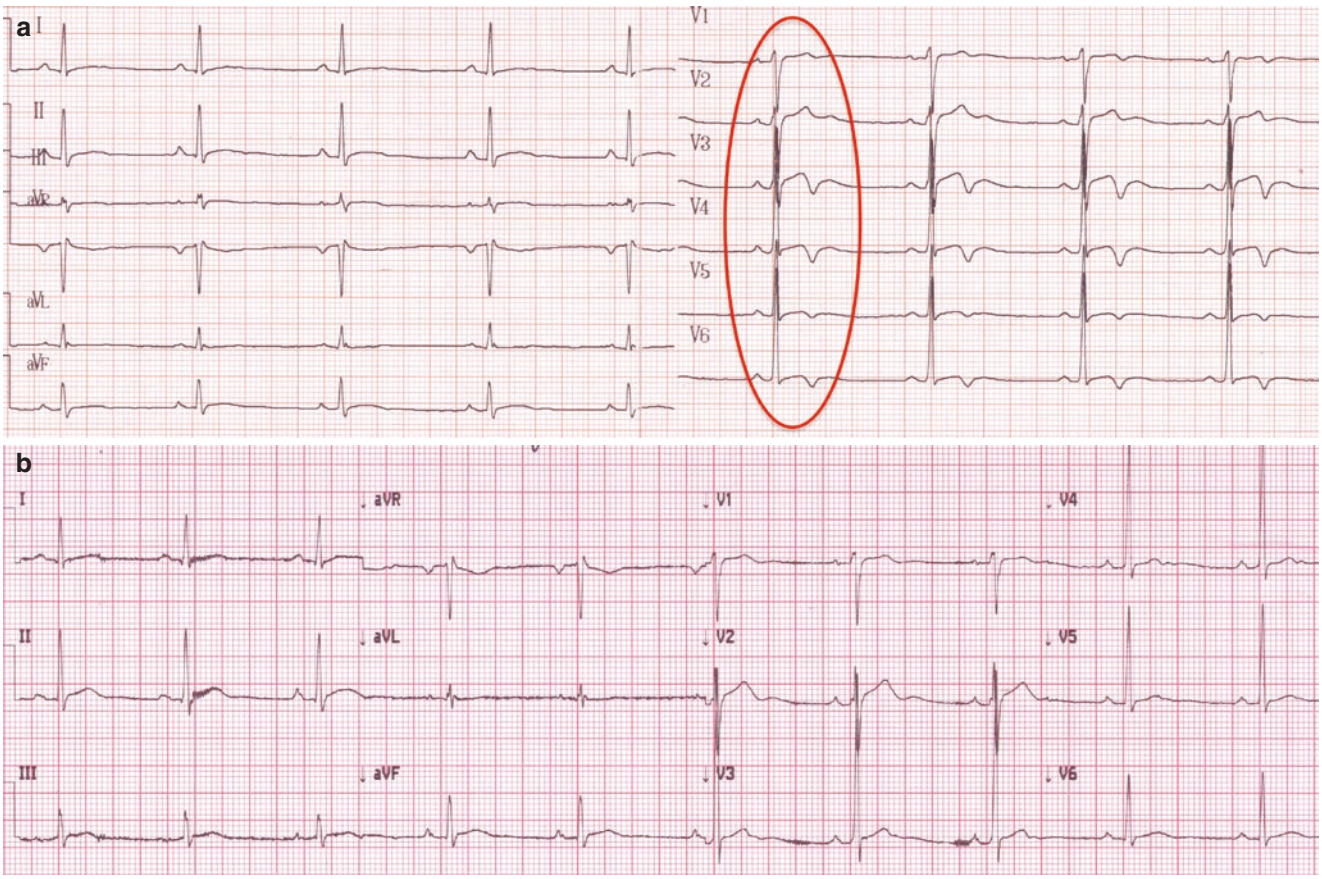

Fig. 19.6 (**a**) 15-year-old male basketball player. The domed ST followed by negative T wave is accepted as a benign variant until V4, while negative T waves in the left precordial leads are less desirable. However, in this case the pattern is not stable and tends to normalize, as the ECG recorded 6 months later shows. (**b**) This variability has to be considered as favorable but nevertheless warrants a follow-up

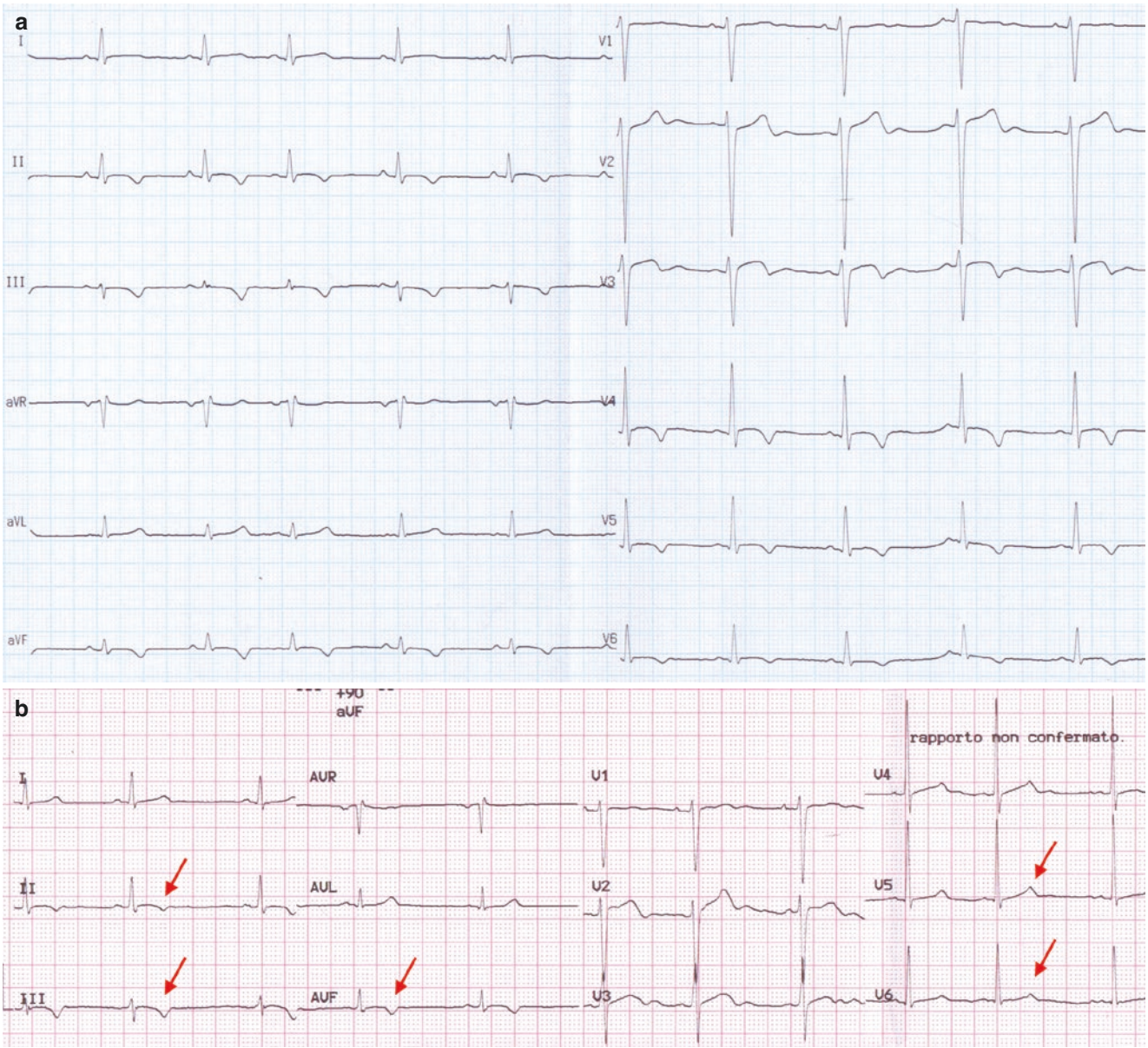

Fig. 19.7 (**a**) 14-year-old male soccer player. As in the previous case, there are widespread ST changes. In the precordial leads, until V4 the picture looks like the normal variant described above. On the contrary, the left precordial leads and the inferior leads show negative T waves in the absence of ST-segment elevation prior to T wave inversion. Also in this case, there is a partial normalization 1 year later (**b**) when the ECG shows negative T waves only in the inferior leads. In the young, negative T waves in the absence of the "benign" domed ST elevation should be regarded with suspicion, and a cardiomyopathy should be ruled out

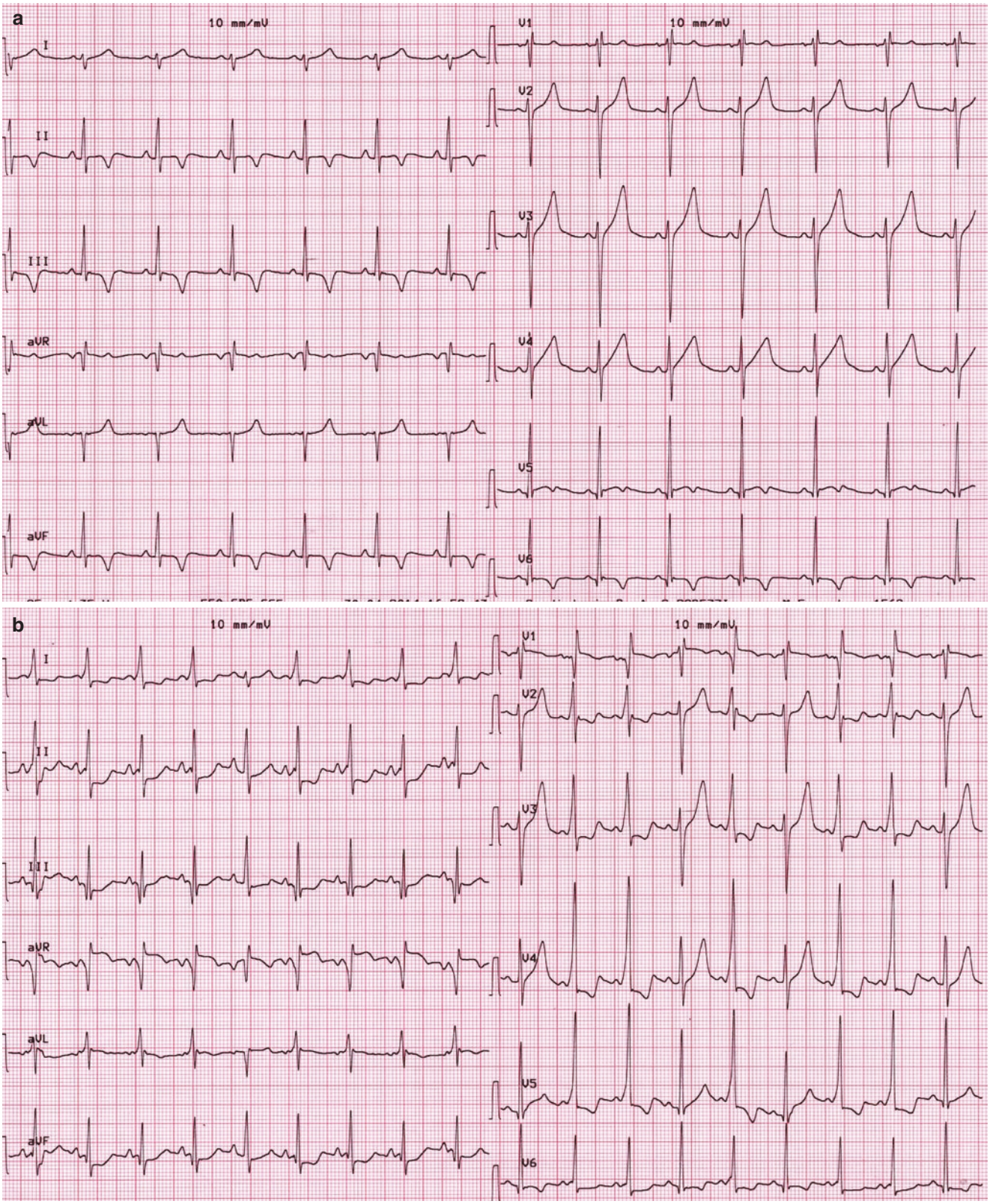

Fig. 19.8 (**a**) 14-year-old male soccer player disqualified due to widespread negative T wave. (**b**) Another ECG revealed a clear intermittent pre-excitation. This last ECG explains the first, which may be interpreted as electrical memory (T wave memory)

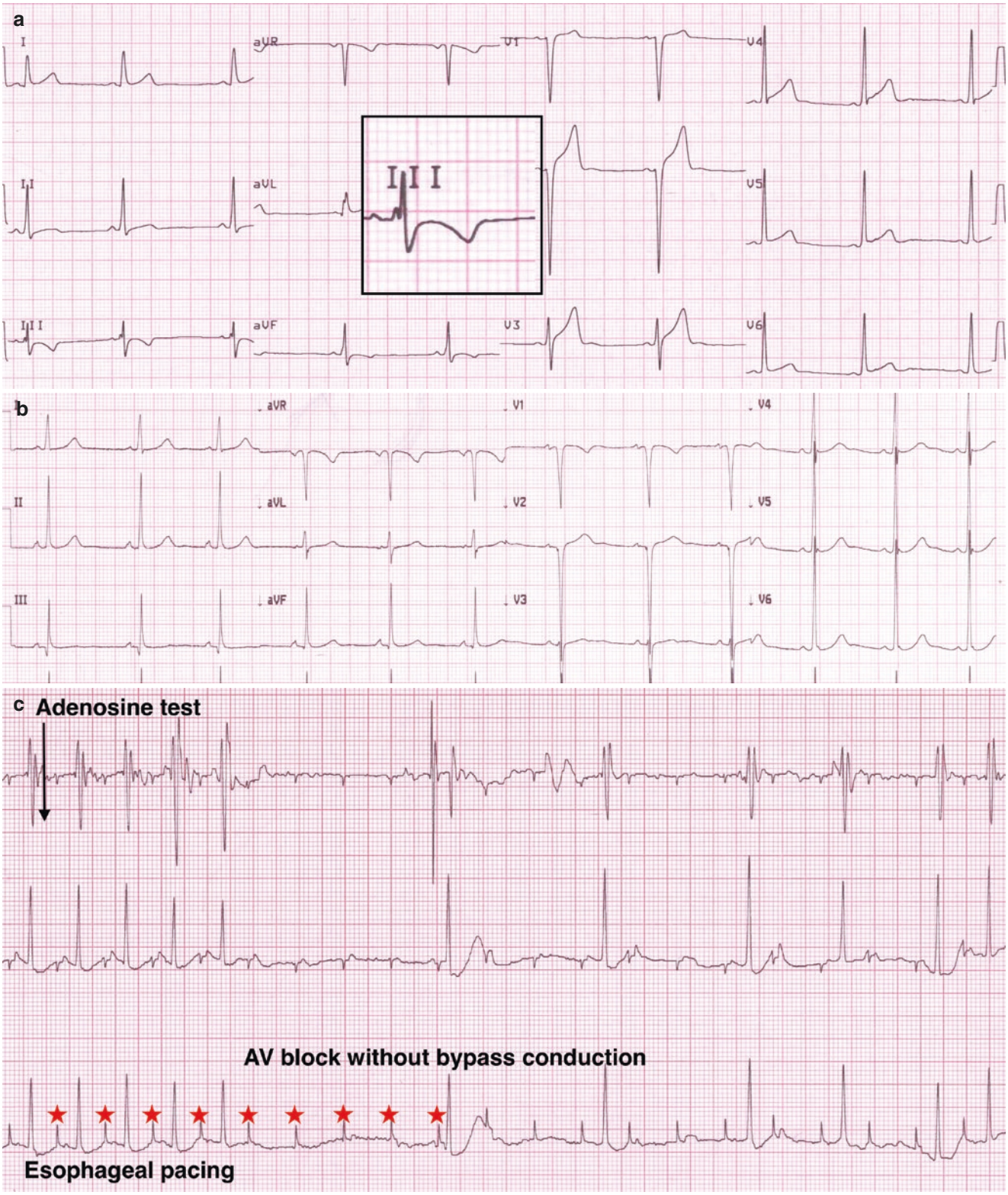

Fig. 19.9 (**a**) 12-year-old soccer player disqualified due to ventricular pre-excitation. The PR interval is 110 ms. There is no clear delta wave, and in lead III the initial part of QRS is slurred. The exercise test was negative and the boy returned to competition. (**b**) 14-year-old gymnast referred for suspected pre-excitation. From V1 to V3, the R wave is almost absent and from V4 to V6 the initial QRS is slurred. The adenosine test was negative (**c**); no accessory pathways were detected. The ECG may be interpreted as normal variant due to clockwise rotation in the horizontal plane. The girl's father has an identical ECG (stars = atrial spikes)

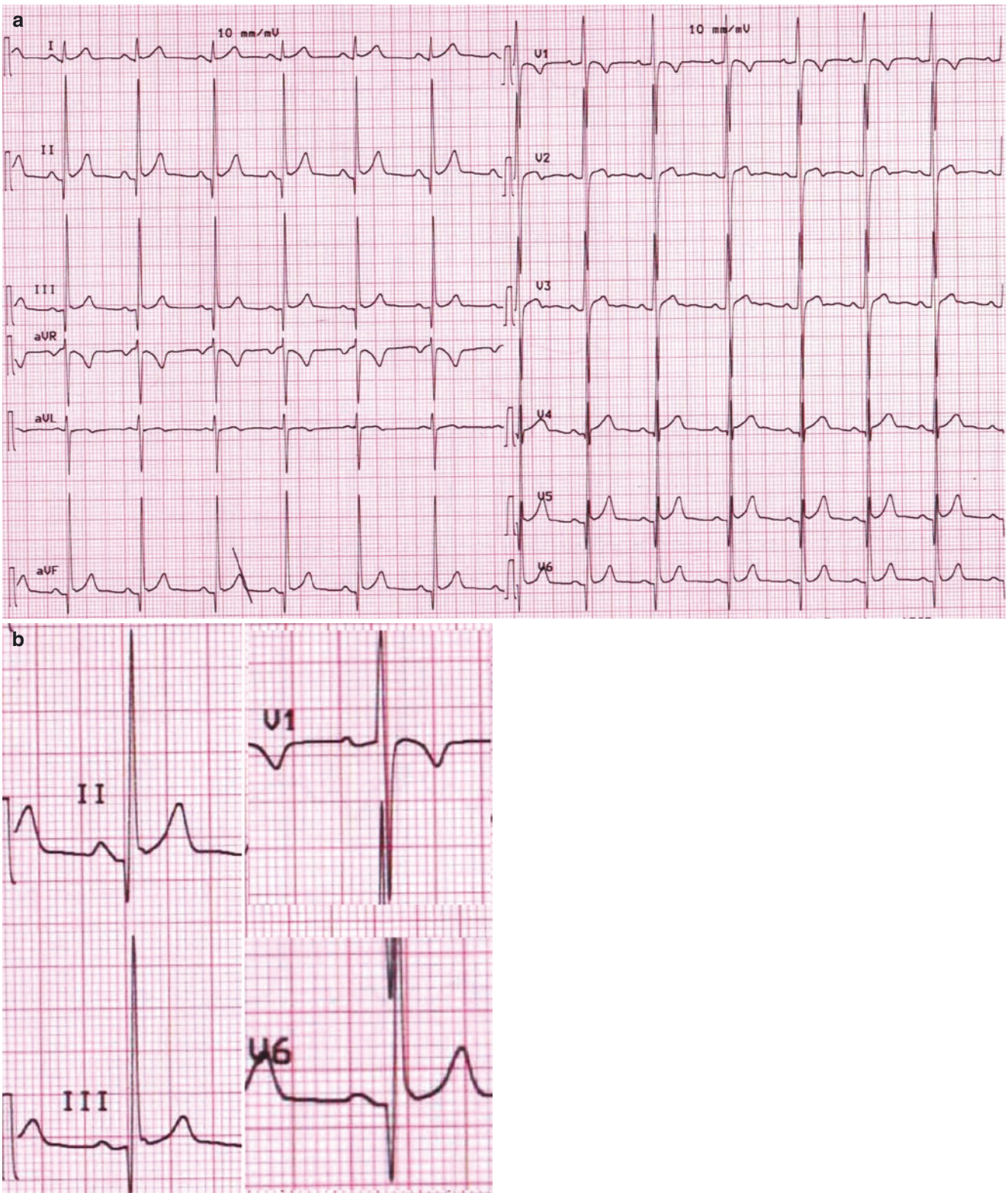

Fig. 19.10 (**a**) 15-year-old soccer player referred for abnormal ECG. The tall R wave in V1 and the prominent Q waves in the inferolateral leads raised the suspicion of neuromuscular disease. The neurological work-up was negative. Echocardiogram and exercise test were both negative: in particular, there was no RVH or Duchenne features. The picture can be interpreted as a counterclockwise rotation in the horizontal plane. (**b**) Close-up. Q waves are less than 25% of R waves

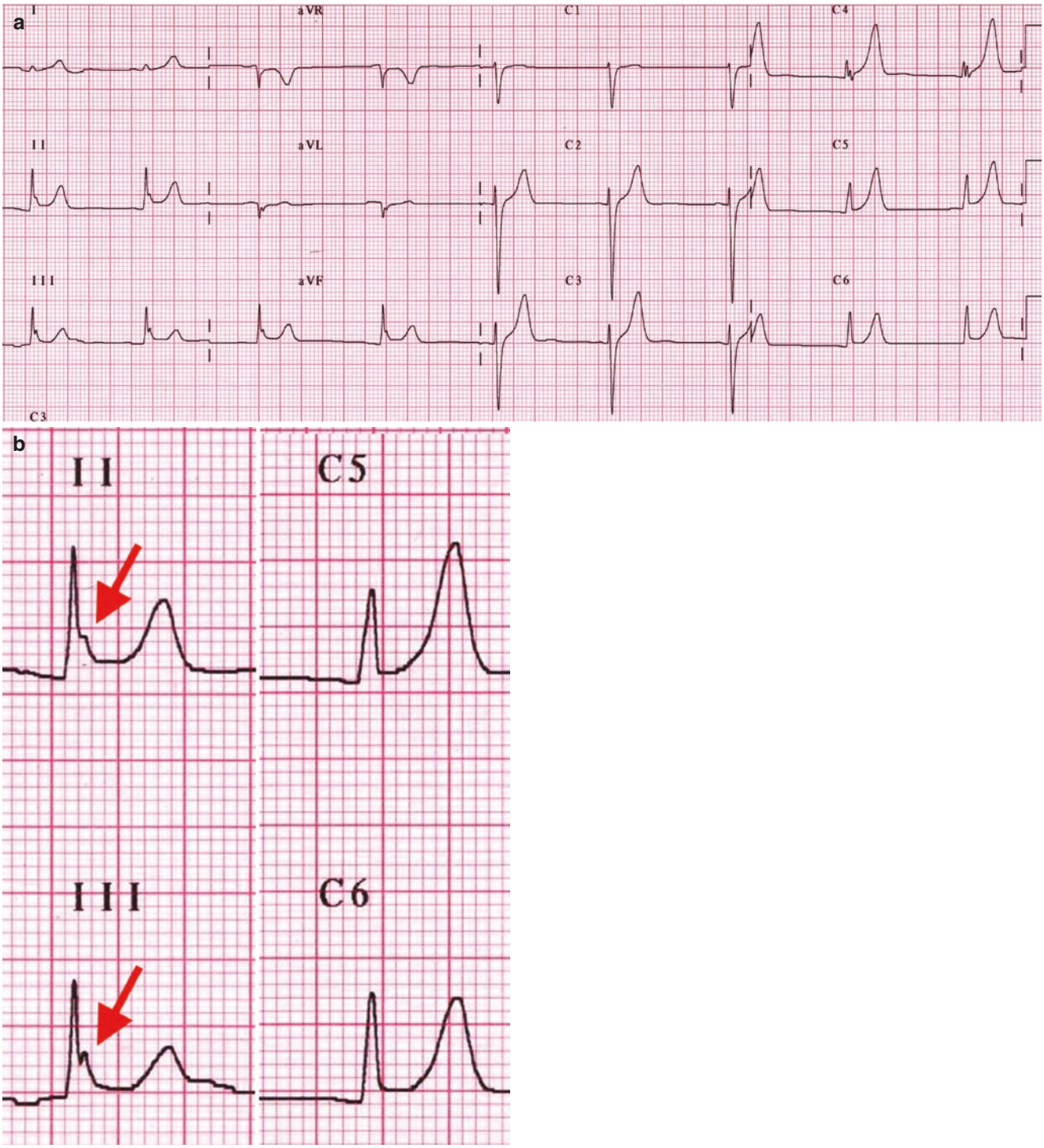

Fig. 19.11 (**a**) 17-year-old male swimmer. The precordial leads are strange, with the trait of clockwise rotation and low voltages. The echocardiogram explained the puzzling ECG: in fact, to obtain the standard long axis view and the apical view, we had to shift the echo windows further down than normal, due to the cardiac shift secondary to the extra large lungs. (**b**) Close-up. The BER pattern can be noted. Interestingly, the T wave seems unaffected by the shift

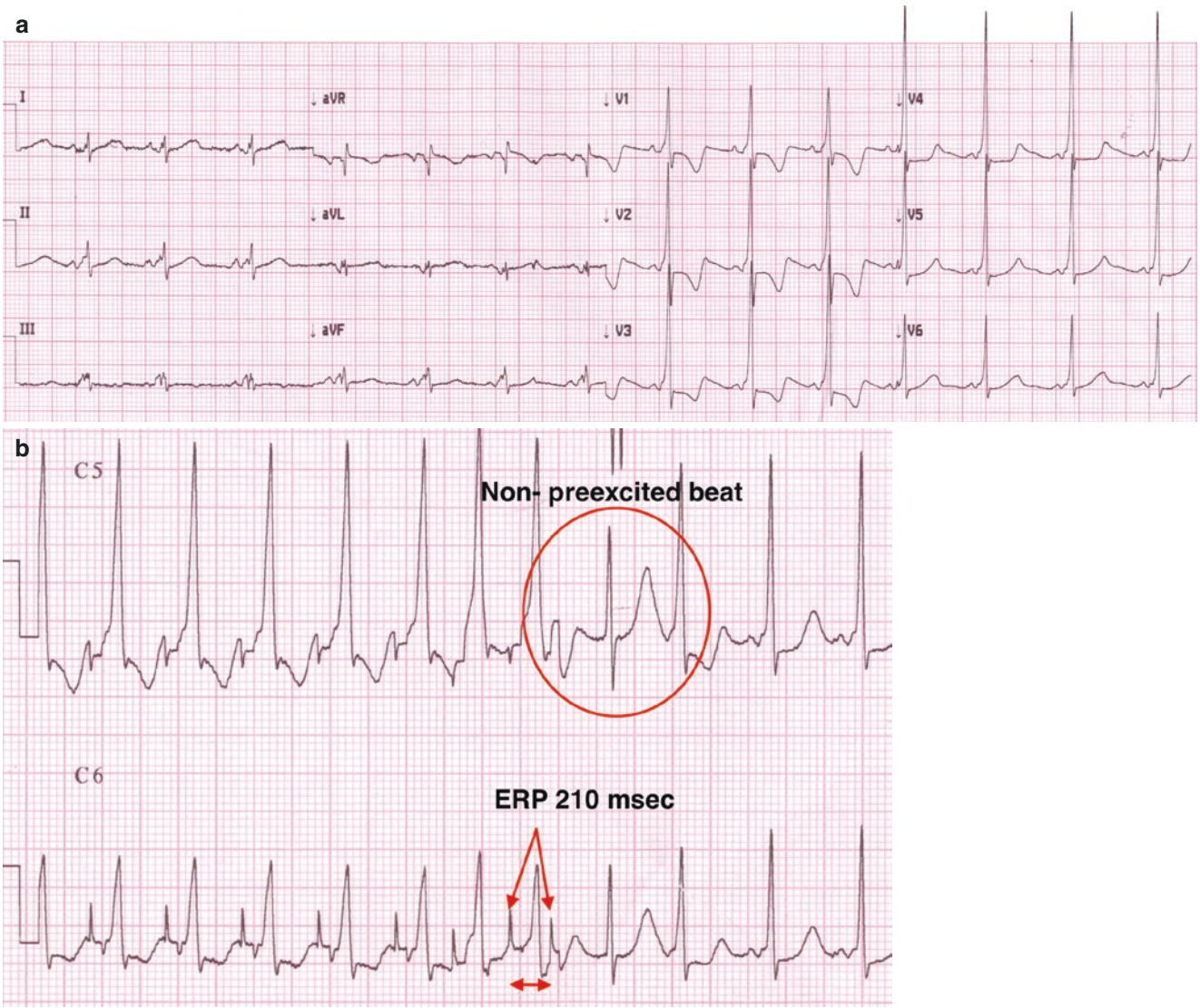

Fig. 19.12 (**a**) 12-year-old boy with left lateral accessory pathways. (**b**) The transesophageal electrophysiologic study was able to stratify the patient at low risk for life-threatening arrhythmias. The tracing shows a programmed atrial stimulation with a basic drive cycle length of 400 ms during isoproterenol infusion. Introducing extrastimuli with a decreasing coupling interval, the AP's effective refractory period (ERP) was 210 ms. This is the interval in which the extrastimulus is conducted to the ventricle only via the AV node, the AP being refractory. The delta wave disappears, the QRS shrinks, and the boy can run

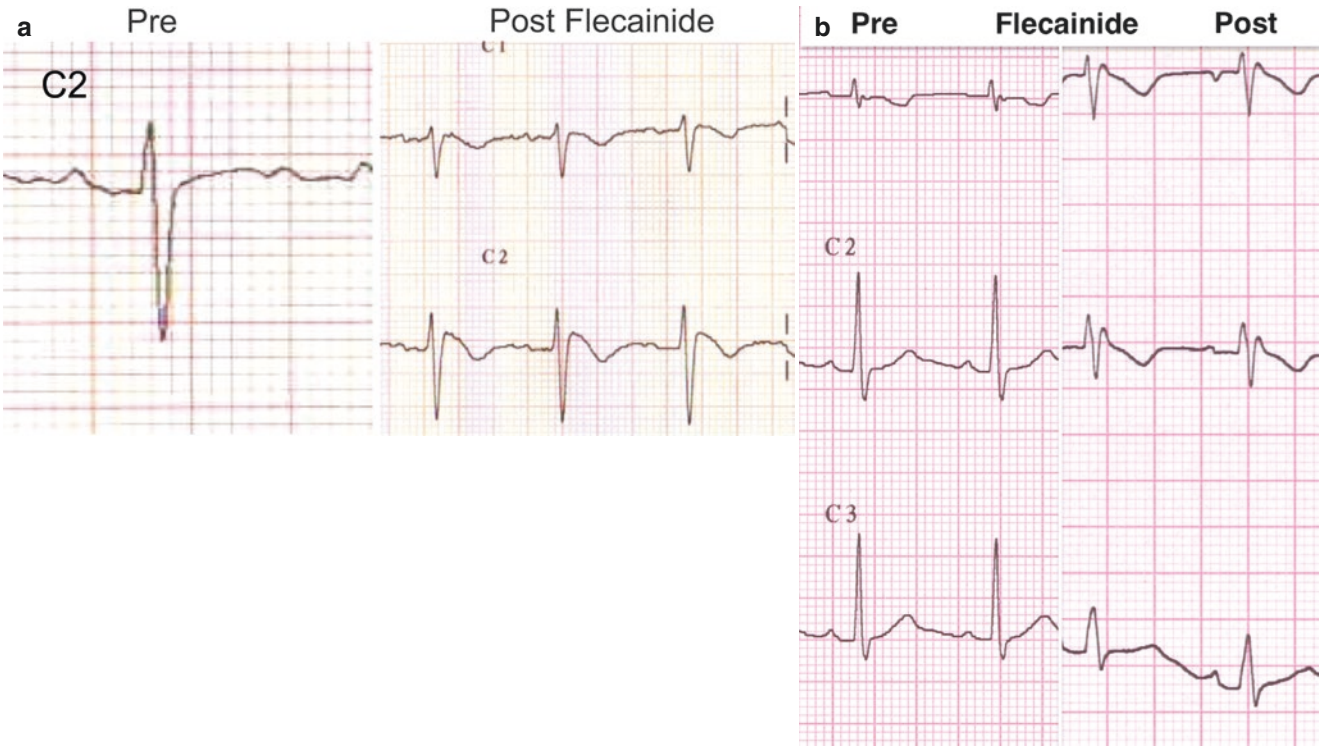

Fig. 19.13 Two young pregnant women in the third trimester. Pregnancy is in fact a 9-month-long sports match, potentially swollen by many dangers. One of these can be the pharmacological treatment of the fetus trough the mother. (**a**) 25-year-old woman carrying a fetus with atrial flutter on treatment with oral flecainide. The ECG on flecainide shows a pattern which if not diagnostic is at least suggestive of Brugada. The genetic test for the Brugada-related mutations was negative. (**b**) As in the previous case, this is a 30-year-old pregnant woman on flecainide to treat the fetal PSVT. In this case, the genetic testing was positive for SCN5A mutation

References

1. Uberoi A, Stein R, Perez MV, et al. Interpretation of the electrocardiogram of young athletes. Circulation. 2011;124:746–57.
2. Trivax JE, McCullough PA. Phidippides cardiomyopathy: a review and case illustration. Clin Cardiol. 2012;35:69–73.
3. Corrado D, Pelliccia A, Heidbuchel H, et al. Section of sports cardiology, European Association of Cardiovascular Prevention and Rehabilitation. Recommendations for interpretation of 12-lead electrocardiogram in the athlete. Eur Heart J. 2010;31:243–59.
4. Drezner JA, Ackerman MJ, Anderson J, et al. Electrocardiographic interpretation in athletes: the "Seattle criteria". Br J Sports Med. 2013;47:122–4.
5. Gati S, Sheikh N, Ghani S, et al. Should axis deviation or atrial enlargement be categorised as abnormal in young athletes? The athlete's electrocardiogram: time for re-appraisal of markers of pathology. Eur Heart J. 2013;34:3641–8.
6. Brosnan M, La Gerche A, Kalman J, et al. The Seattle criteria increase the specificity of preparticipation ECG screening among elite athletes. Br J Sports Med. 2014;48:1144–50.

Suggested Reading

Zorzi A, ElMaghawry M, Corrado D. Evolving interpretation of the athlete's electrocardiogram: from European Society of Cardiology and Stanford criteria, to Seattle criteria and beyond. J Electrocardiol. 2015;48:283–91.

Drezner JA, Ackerman MJ, Anderson J, et al. Electrocardiographic interpretation in athletes: the "Seattle criteria". Br J Sports Med. 2013;47:122–4.

Brosnan M, La Gerche A, Kalman J, et al. The Seattle criteria increase the specificity of preparticipation ECG screening among elite athletes. Br J Sports Med. 2014;48:1144–50.

Conclusions

20

More than a century after its birth, and in the era of demanding tests,—fast and silent like computed tomography or slow and loud like magnetic resonance—the ECG retains its "original lightness," a bearable lightness. The ECG flies and finally lands on the patient's shoulder without the gravity of other tools but with formidable diagnostic yield, able to shake the shoulders of the most skeptical cardiologist.

Rarely can other disciplines in science and medicine make use of a test which is both economic and at the same time so wide ranging. The investment does not end in a pink sheet and will have fulfilled the eventful cultural ground traveled by those who have to read these "light" exams. This book aims to alleviate the task of these professionals of the heart, whether frugal editors of reports or romantic and visionary exegetes of the pink sheet. Of course, it is a surface ECG, but only shallow people do not judge by appearances. "We are such stuff as dreams are made on, and our little life is rounded with a sleep." So it's up to us to interpret them. To those who have had the patience and goodwill to get this far, we wish sweet dreams or rather pink ones!

Among the many images of this book, the first is the ECG of a neonate, and the last is the ECG of a pregnant woman. The image of a woman "daughter of her son" will close this book. It is, in fact, hard to imagine a more evolutionary situation. Mary's halo reflects the checkered floor to remind us that even the most horizontal pink and measurable image can have a vertical significance beyond our wildest imagination.

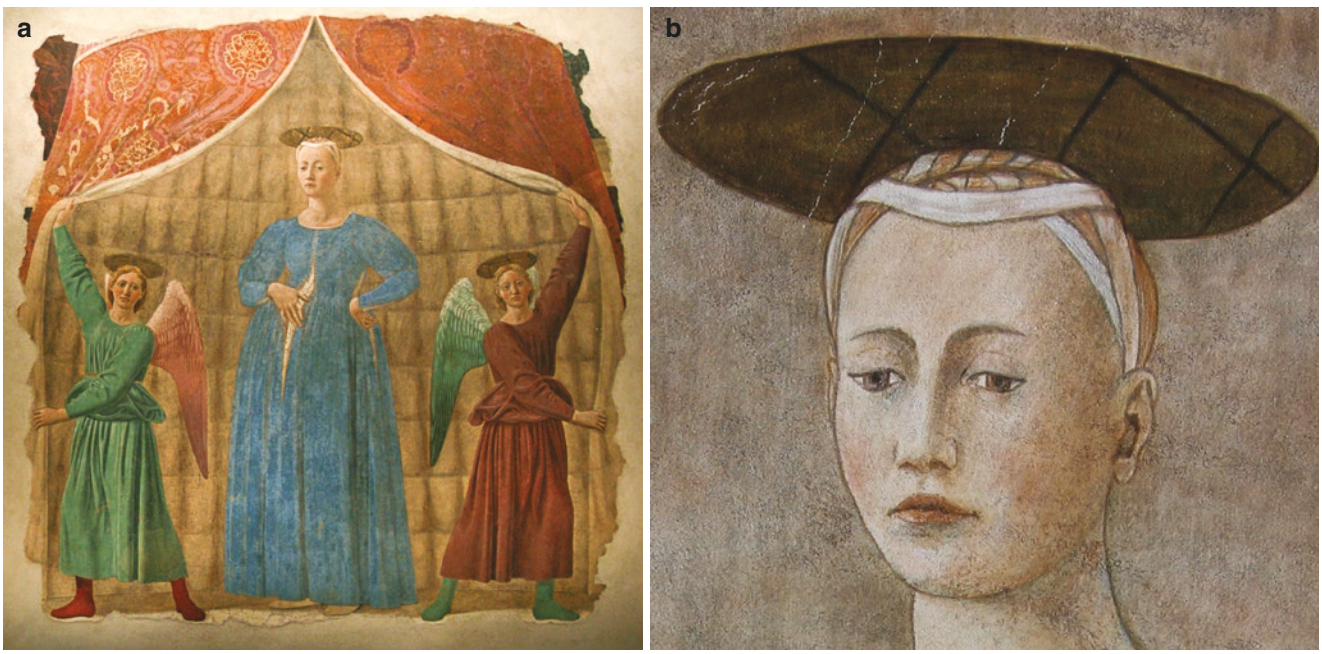

Fig. 20.1 (**a**) "Madonna del Parto," Piero della Francesca (Fresco, Monterchi, Italy) (**b**) detail. Mary's halo

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