ASYMPTOMATIC ELITE ATHLETE SOCCER PLAYER WITH DOUBTFUL HEART DISEASE

FUTEBOLISTA ATLETA DE ELITE ASSINTOMÁTICO COM SUSPEITA DE DOENÇA CARDIACA

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Jovem atleta de elite, caucasiano, futebolista profissional (zagueiro), 23 anos, segundo grau completo, natural Blumenau Estado de Santa Catarina e morador de Santo André São Paulo Brazil. Veio ao nosso núcleo de Saúde no Esporte da Faculdade de Medicina do ABC (FMABC) para avaliação periódica obrigatória. Refere que realiza treinos regulares seis vezes por semana de aproximadamente 1hora e 30minutos por dia de caráter competitivo.Assintomático. Nega alergias, cirurgias previas, vícios ou drogas ilícitas. História familiar negativa sem mortes súbitas em parentes jovens de primeiro grau

Exame físico: corado, eupnéico 74Kg, 1,84 m, PA: 130/80, FC: 66bpm, circunferência abdominal: 84cm. Exame cardiovascular, pulmonar e outros NDN.

O atleta trazia em mãos uma declaração do InCor de SP datada de dezembro de 2006 onde constaba que já havia realizado exames cardiológicos tendo interrompido os treinamentos por 4 meses decorrente da presença de pausas sinusais noturnas assintomáticas prolongadas (de até 7 segundos) registradas em repetidos exames de Holter.

O ECG desta data mostrava alterações da repolarização ventricular em parede antero-septal e ondas S profundas de V_2 a V_4.

Ecocardiograma: discreta hipertrofia septal (septo 13mm) e de parede posterior(12mm) simétrica sem obstrução na via de saída, FE VE: (68%), cálculo de massa normal (133,77g/m2) e todas as câmaras de dimensões normais.

Teste ergométrico: normal

Holter repetidos mostraram eventual aparecimento de bloqueio AV de segundo grau tipo Wenckebach e pausas noturnas assintomáticas por vezes numerosas de sendo a maior de 7 segundos. Todas ocorridas durante a noite. Eventual ritmo de RIVA (Ritmo Idioventricular Acelerado)

ECG-AR: Ausência de potenciais tardios.

Ressonância Magnética Cardíaca realizada em janeiro de 2007 onde consta mínima acentuação do trabeculado do terço distal da parede lateral e inferolateral distal do VE considerada muito provavelmente como uma variação anatômica normal. Estudo com perfusão miocárdica normal.

Young elite athlete, Caucasian, professional soccer player (defender), 23 years old, second degree in primary school complete. He was born in Blumenau, State of Santa Catarina, and lives in Santo André, São Paulo, Brazil. He came to our Center of Health and Sports of the School of Medicine of ABC (FMABC) for the compulsory periodical evaluation. He states that he undergoes a regular training six times a week, of approximately 1 hour and 30 minutes per day, of a competitive nature. Asymptomatic. He denies having allergies, prior surgeries, or vices. Negative family history, without sudden cardiac deaths in a young, first-degree relative.

Physical examination: flushed, eupneic, 74 Kg, 1.84 m of height, BP 130/80,HR 66 bpm, abdominal circumference 84 cm. Cardiovascular and pulmonary examination, and others, nothing worth mentioning.

The athlete brought a statement dated from December 2006, where it said that he had undergone cardiological examinations and had interrupted his training for 4 months due to the presence of night, asymptomatic, and prolonged sinus pauses (of up to 7 seconds) recorded in Holter.

The ECG from this date showed ventricular repolarization alterations in the antero-septal wall and deep S wave from V2 through V4.

Echocardiogram: discrete septal hypertrophy (septum 13 mm) and posterior wall (12 mm), that was symmetrical without obstruction in the outflow tract, EF (68%), normal mass estimation (133.77 g/m2) and all chambers with normal dimensions.

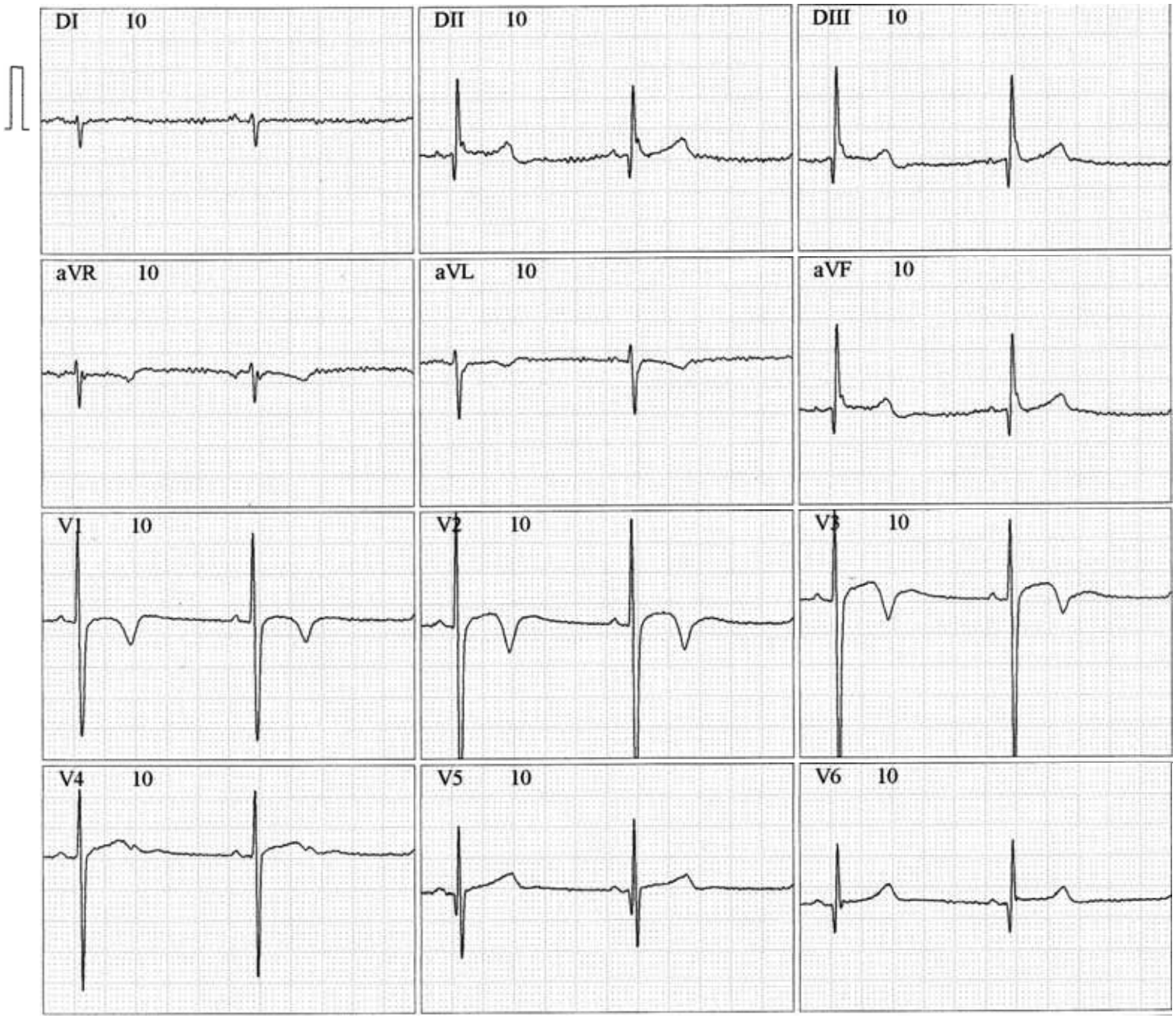
Treadmill exercise testing: Normal.

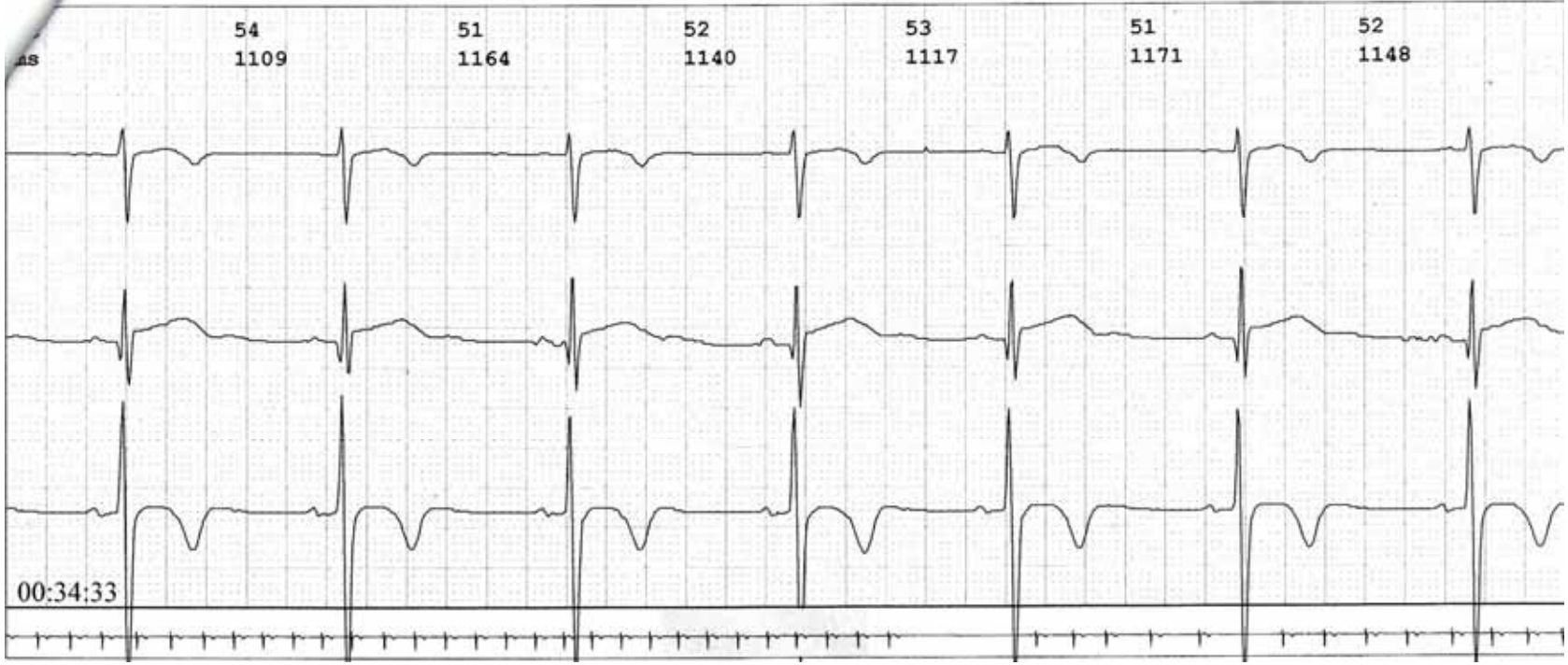
Repeated Holters: showed possible appearance of AV block, of the Wenckebach type, and asymptomatic night pauses, at times numerous and greater than 7 seconds. All occurring during the night. Possible accelerated idioventricular rhythm (AIVR).

High-resolution ECG(SAECG) : absence of late potentials.

Cardiac magnetic resonance conducted on January 2007, where there is a minimal emphasis of the trabeculated distal third of the lateral wall an distal inferolateral wall of the LV, considered very likely as a normal anatomic variation. Normal study with myocardial perfusion.

Name: M.G.; Gender: M.; Age: 23 yo.; Ethnic Group: Caucasian.;
Profession: Soccer player.; Weight: 79Kg.; Height: 1,84m.; Date: 12/14/2010

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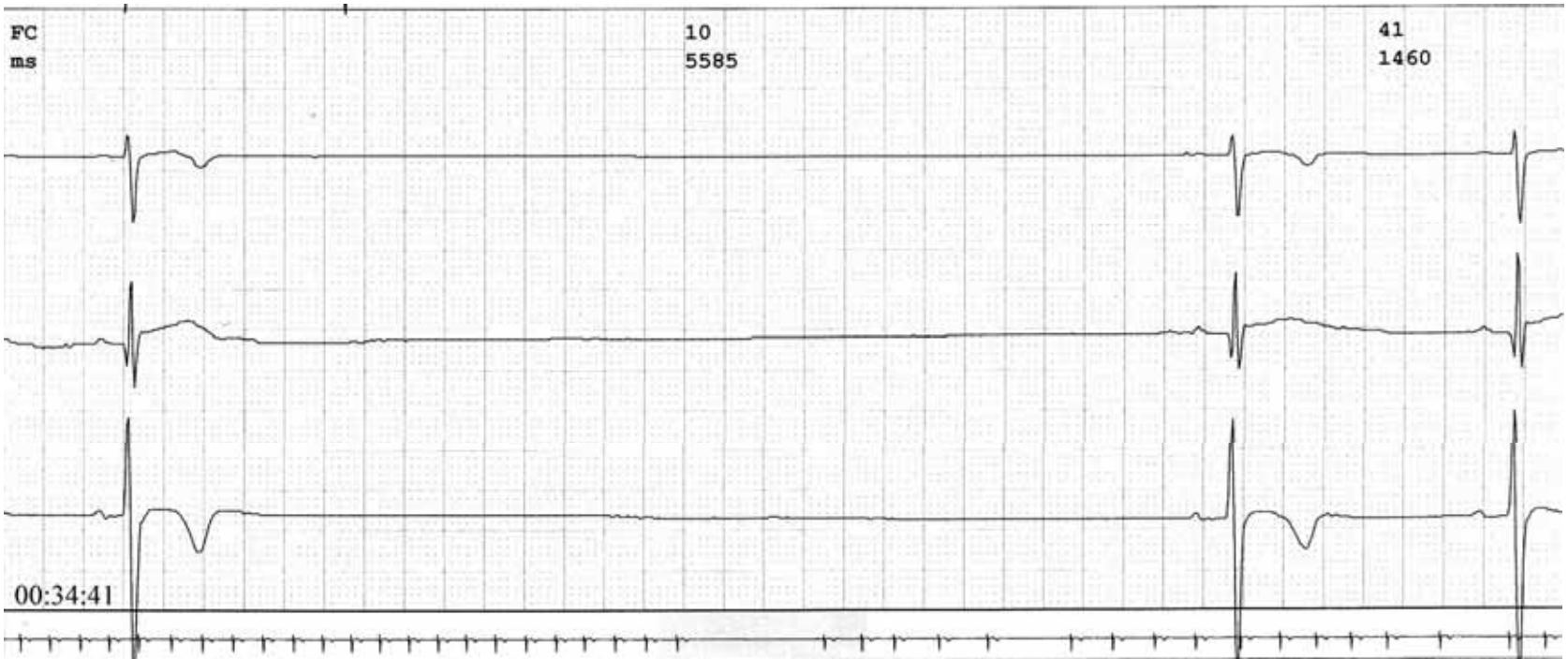
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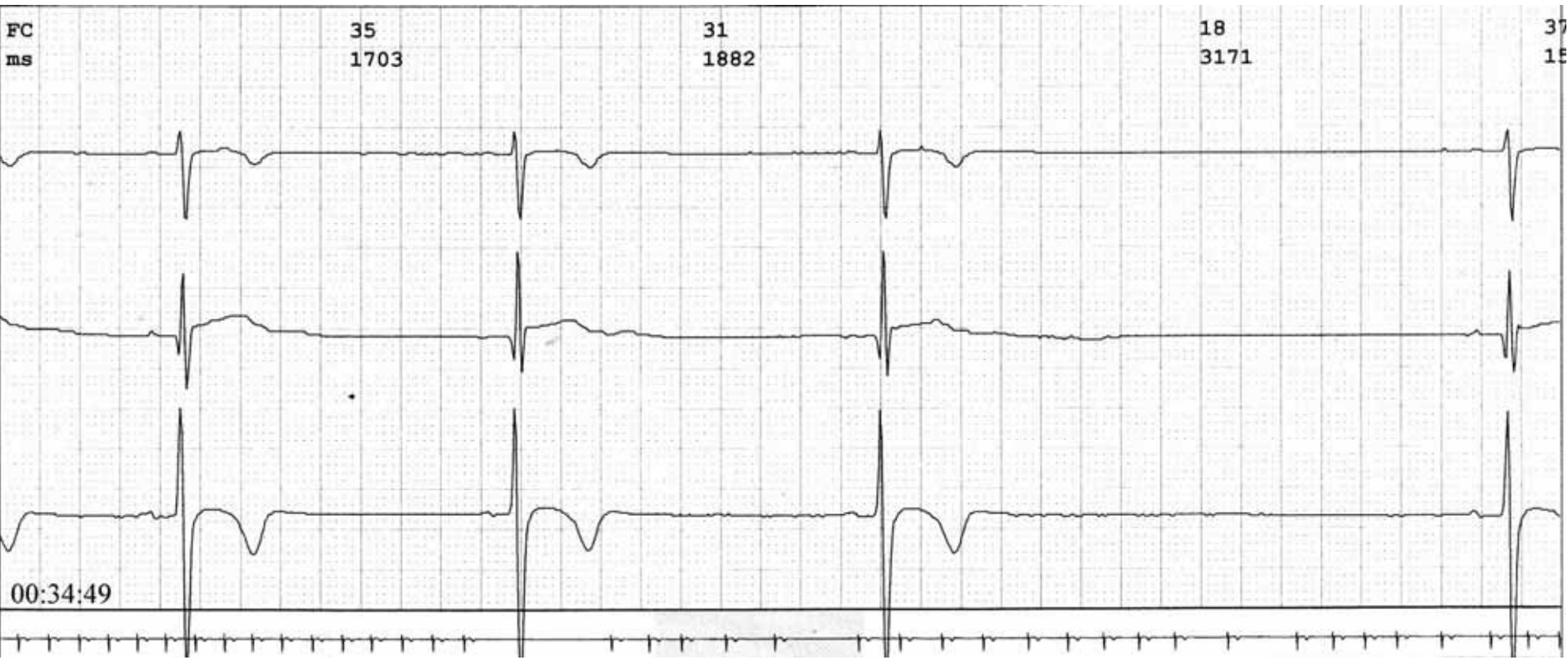
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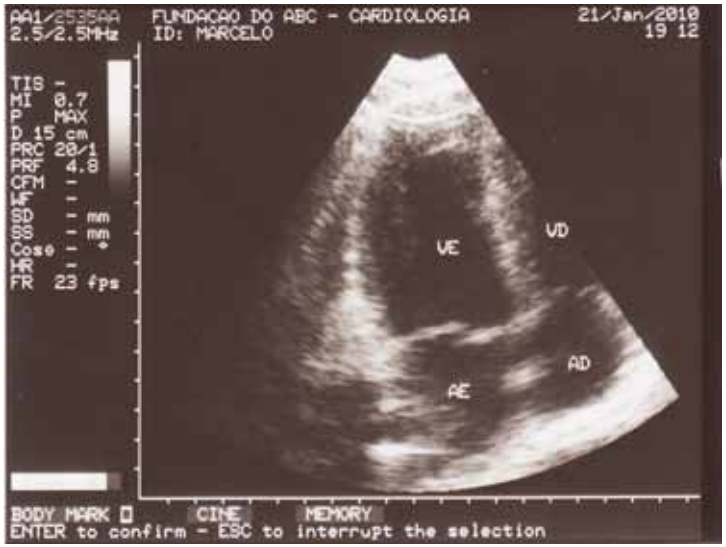
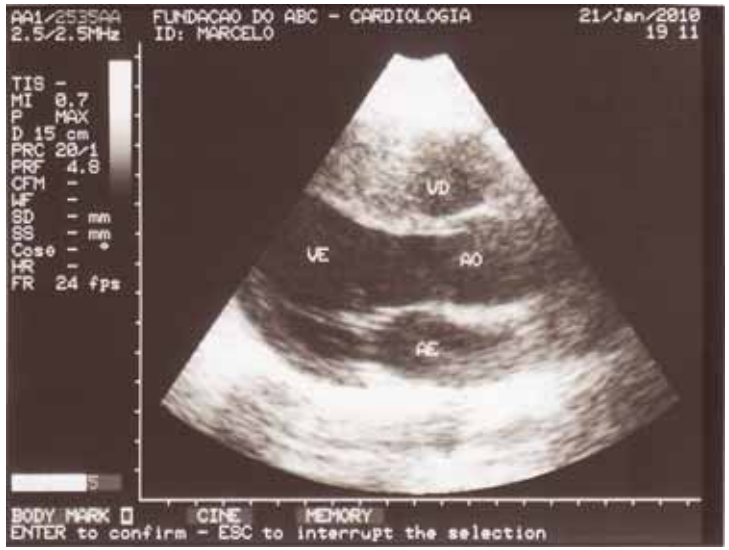
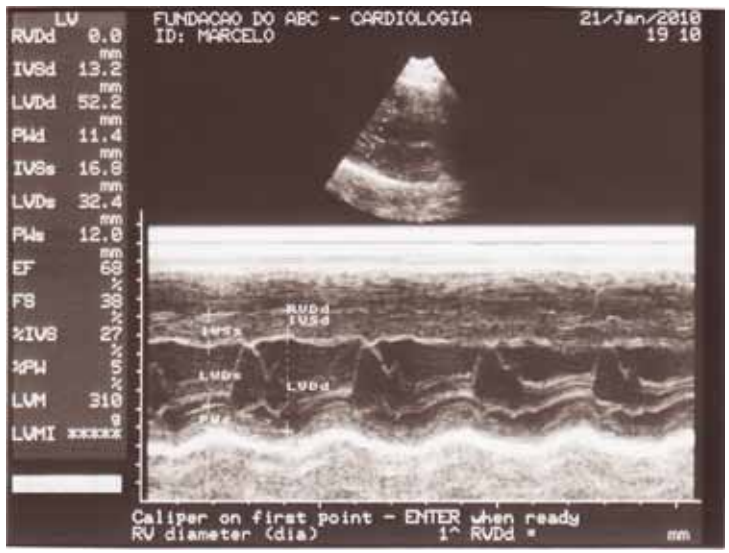




La medición del septum en diástole se hizo a nivel de válvula mitral, cuando deberia hacerce por debajo de los bordes libres de la misma y en eje corto. Con este proceder equivocado se obtienen septos falsamente hipertróficos. Este septo no mide mas de 12 mm y además no le veo el aspecto de vidrio esmerilado caracteristico de MCH.

The measurement of the septum in diastole was made at the mitral valve level, when should JOINING below the free edges of it and in short axis.



This wrong procedure are obtained falsely hypertrophic septum. This septum is no more than 12 mm and also do not see the ground-glass appearance characteristic of HCM.

Juan José Sirena MD commentaries Santiago del Estero Argentina.

Estimados colegas y Andrés
Mi opinion: En el contexto clinico del joven deportista asintomático, sin antecedentes familiares, con ergometria normal e hipertrofia ventricular izquierda concéntrica diagnosticada por el ecocardiograma las alteraciones encontradas en el ECG y Holter las interpreto asi: 1- Sindrome de repolarización precóz (supradesnivel del punto J y del segmento ST en las precordiales derechas). 2- Disfunción sinoauricular extrinseca por hipertono vagal con disminución de la población de los receptores beta (beta bloqueo fisiológico por sobreentrenamiento) que explica la bradicardia sinusal y las pausa nocturnas de 6-7 segundos. Recomendaria para confirmar y liberarlo para la práctica deportiva: Cardio-estimulación trans-esofágica pre y post-atropina para evaluar la función del nódulo sinusal y punto Wenchebach que seguramente seran normales. En síntesis: Sindrome cardioritmológico del atleta
Afectuosamente Juan Sirena PD: tengo casos parecidos que poximamente enviaré al foro

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Dear colleagues and Andrés
My opinion: In the clinical setting of asymptomatic young athlete with no family history, normal exercise testing and left symmetrical ventricular hypertrophy diagnosed by echocardiogram, the alterations found in the ECG and Holter I interpret them as follows: 1 Early repolarization pattern (J-point elevation and ST segment in right precordial leads) 2 Sinoatrial dysfunction by extrinsic vagal hypertonus and decreased population of beta receptors (beta blocking physiological overtraining status) that shows sinus bradycardia and nocturnal pause of 6-7 seconds. I would recommend: Cardio transesophageal stimulation pre-and post-atropine to assess sinus node function and will surely point normal Wenchebach. Summary: cardio rhythmologic athlete syndrome
Affectionately
Juan Sirena MD
PS: I have similar cases to send to the forum soon.

Estimado Andrés,

Este sí es un caso desafiante!. Desde ya le has realizado todas las evaluaciones que clarifican el cuadro. Desde mi punto de vista hay varios elementos a tener en cuenta:

El paciente es asintomático: Buen pronóstico. Diferente sería si tuviera síntomas.

No tiene antecedentes familiares de MSC o cardiopatía hereditaria: Otro dato a favor de la "benignidad".

Los deportistas entrenados hasta el 80% tienen alteraciones en el ECG. La repolarización precoz es la regla más que la excepción, 50-80 % de los deportistas la presentan. Esto es debido a la hipervagotonía, que es un fenómeno reversible y se reduce o desaparece con el de-acondicionamiento. En estos casos la bradicardia aumenta el supradesnivel del ST y la taquicardia o el isoproterenol disminuye o la hace desaparecer[1]. A pesar que se ha demostrado en un grupo seleccionado que la repolarización precoz podría ser un predictor de FV[2], los datos disponibles, aún no apoyan que en la población general asintomática o atletas con esta alteración sea predictiva de alto riesgo de FV. La repolarización precoz hay que diferenciarla del SBr, que en este caso no tiene. .

Las ondas T negativas en las precordiales derechas es un fenómeno que puede acompañar a la repolarización precoz. Recientemente se ha demostrado que la presencia de T negativas podría estar indicando una cardiopatía subyacente aún sin una cardiopatía demostrable. En estos casos seguimiento clínico, ECGs seriados y ECO frecuentes es una recomendación a seguir[3]. En este caso la RNM descarta la displasia del VD.

1. **Bianco M, Bria S, Gianfelici A, Sanna N, Palmieri V, Zeppilli P. Does early repolarization in the athlete have analogies with the Brugada syndrome? Eur Heart J 2001;22:504–510.**
2. **Haï¨ssaguerre M, Derval N, Sacher F, et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med 2008;358:2016–2023.**
3. **Pelliccia A, Di Paolo FM, Quattrini FM, Basso C, Culasso F, Popoli G, De Luca R, Spataro A, Biffi A, Thiene G, Maron BJ. Outcomes in athletes with marked ECG repolarization abnormalities. N Engl J Med 2008;358:152–161.**

Frecuentemente el ECG muestra signos de HVI que refleja el remodelamiento fisiológico con engrosamiento de la pared y de la cavidad. Demostrado en este caso por una HVI leve.
La bradicardia es el resultado de cambios en el SNA. La bradicardia sinusal y los paros sinusales > 3 seg debe ser distinguido de la enfermedad del nódulo. Esto puede ser excluido por demostrar que no hay síntomas, que la FC se normaliza con el ejercicio y esta bradicardia revierte con el de-acondicionamiento. En este caso, a pesar de presentar pausas de hasta 7 seg, durante el sueño (gran hipertonía vagal) considero que no tiene enfermedad del nódulo sinusal. Igualmente el fenómeno Wenckebach que presenta.
Por lo tanto, creo que lo que le pasa a este paciente es bastante fisiológico y lo dejaría continuar con su actividad. Eso sí, el seguro de mala praxis al día, por las dudas vió, uno nunca sabe donde el diablo mete la cola.
Saludos,
Oscar Pellizón MD.

Dear Andres, This is a challenging case! Of course you made all the evaluations to clarify the situation. From my point of view, there are several elements to take into account.
The patient is asymptomatic: good prognosis. It would be different if he had any symptoms.
There is no family history of SCD or inherited heart disease: another data in favor of a “benign” outcome. Up to an 80% of trained sportsmen have ECG alterations. Early repolarization is a rule more than an exception, 50-80% of sportsmen have it. This is due to hypervagotonia, which is a reversible phenomenon and is reduced or disappears with deconditioning. In these cases, bradycardia increases ST elevation and tachycardia or isoproterenol decreases it or makes it disappear(1). In spite of what has been proven in a selected group, i.e. that early repolarization could be a predictor of VF(2), the data available still do not support that in the general asymptomatic population or athletes with this alteration, it could be a high risk predictor for VF. Early repolarization should be differentiated from BrS, which is absent in this case.

Negative T waves in the right precordial leads is a phenomenon that could accompany early repolarization. Recently, it has been proven that the presence of negative T waves could indicate an underlying heart disease, still without demonstrable heart disease. In these cases, clinical follow up, serial ECGs, and frequent echos, are a recommendation to follow(3). In this case, NMR rules out RV dysplasia. Frequently, ECG shows LVH signs that reflect physiological remodeling with wall and chamber thickening. Proven in this case by mild LVH. Bradycardia is the result of changes in the ANS. Sinus bradycardia and sinus arrests > 3 sec should be differentiated from node disease. This could be ruled out by showing there are no symptoms, that HR becomes normal with exercise, and that this bradycardia reverts with deconditioning. In this case, in spite of presenting pauses of up to 7 sec during sleep (great vagal hypertonia), I think that there is no sinus node disease. The same for the Wenckebach phenomenon he presents. Therefore, I think that what is going on with this patient is quite physiological, and I would let him continue with his activity. But of course, keep the malpractice insurance updated, just in case you know. One never knows when the devil may play his tricks.

Regards, Oscar Pellizón MD.

1.Bianco M, Bria S, Gianfelici A, Sanna N, Palmieri V, Zeppilli P. Does early repolarization in the athlete have analogies with the Brugada syndrome? Eur Heart J 2001;22:504–510.

2.Haı̈ssaguerre M, Derval N, Sacher F, et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med 2008;358:2016–2023.

3.Pelliccia A, Di Paolo FM, Quattrini FM, Basso C, Culasso F, Popoli G, De Luca R, Spataro A, Biffi A, Thiene G, Maron BJ. Outcomes in athletes with marked ECG repolarization abnormalities. N Engl J Med 2008;358:152–161.

Analisis del joven futbolista Antes todo quiero decir que este caso es desde el punto de vista electrocardiografico una hIpertrofia asimétrica. Porque ? porque en las hipertrofias asimétricas los complejos QRS son cancelados como en los ECG normales Vamos al análisis de este caso: El corazón está horizontalizado (ondas P a la izquierda, positivas en I y aVL, con el eje del QRS hacia abajo y a la derecha). El segmento ST está elevado en las derivaciones inferiores y ligeramenenta deprimido en I aVL lo que dificulta el diagnóstico de repolarización precóz. Se observan ondas Q profundas en V_5-V_6 por hipertrofia septal que dirige el primer vector hacia la derecha. Las ondas S, profundas > 30mm (estan cortadas) sugieren que los potenciales han sido desviados hacia atrás, muy probablemente por hipertrofia septal posterior. Las ondas T negativas de V_1 a V_4 , son patológicas y probablemente consecuencia de hipertrofia septal (desde el septo alto hasta el septo bajo), planteando el diagnóstico diferential entre isquemia crónica y el patrón juvenil de T. En muchos casos de ondas T invertidas de las hipertrofias ventriculares, el aumento de la frecuencia cardiaca puede ocasionar pseudo normalización de las mismas. porque la taquicardias al aumentar la secreción adrenérgica e produce un acortamiento selectivo del intervalo QT de la camada epicárdica de la pared ventricular ocasionando positivización de onda T precordial. Durante el Holter o la prueba de esfuerzo este fenómeno suele observarse porque la concentración de receptores de adenosina a1 es 17 veces mayor en el epicardio que en el endocardio. Estos receptores a1 aumentan la concentración de K+ dependiente de ATP que acorta el potencial de acción del epicardio defendiendo a esta camada del efecto destructor de la epinefrina. Este fenomeno biológico es bien conocido en las isquemias agudas, hiperkalemia, hipercalcemia, y en el género masculino por acción de los andrógenos.

Con respecto al Holter no consigo ver la hora del registro, pero si es de noche puede ser efecto vagotónico durante la fase REM. Si estos paros sinusales se tornan muy frecuentes, puede acarrear transtonos acumulativos de las funciones cognitivas cerebrales.Conducta: interrupción de las actividades deportivas por 6 meses. Si después de este tiempo la hipertrofia revierte, lo liberaria para la práctica de deporte.

Un fraternal abrazo

Samuel Sclarovsky

Analysis of young soccer player

Before all I think that this case from the electrocardiograph standpoint has an asymmetrical hypertrophy. Why asymmetrical hypertrophy?. Because the QRS complexes are canceled, as in the normal ECGLet's go ahead to the analysis of this case: The heart has an horizontal position: P waves on the left, (positive in I and aVL) with the QRS axis down and to right. The ST elevation in the inferior leads, and minimally depressed in I and aVL (difficult the diagnosis of early repolarization). Precordial leads show deep Q waves in left leads V_5-V_6 consequence of septal hypertrophy because the first vector is directed to the right. S waves, deep > 30mm (they are cut) suggest that the potential was shifted back, most likely by posterior septal hypertrophy. Negative T waves from V_1 to V_4, are pathological and probably secondary to septal hypertrophy (septum from the high to the low septum), raising the differential diagnosis between chronic ischemia and juvenile T pattern. In many cases of inverted T waves in ventricular hypertrophy, increased heart rate may cause pseudo normalization of them because the tachycardia by increasing adrenergic secretion produces a selective shortening of the QT interval of the epicardial layer of ventricular wall causing positive polarity of the precordial T waves. During the Holter or exercise testing this phenomenon is usually observed because the concentration of adenosine A1 receptors is 17 times greater in the epicardium than the endocardium. These receptors a1 increase the concentration of K+ ATP-dependent shortening of action potential of epicardium defending this layer of the destructive effect of epinephrine. This biological phenomenon is well known in acute ischemia, hyperkalemia, hypercalcemia, and male genderby androgen action.

In reference to the Holter monitoring, I can not see the time of registration, but if it is night, may be increased parasympathetic tone during REM phase. If these sinus arrest are very frequent, may result in cumulative brain cognitive functions upset.

Conduct: interruption of sporting activities by 6 months. If after this time reverses hypertrophy, release him for the practice of sport.

Hug

Samuel Sclarowski

Dear Andrés,

The ECG with the very thin and deep Q waves strongly suggests HCM. Biphasic T waves may not be specific. The echo shows a moderate LVH. If this morphological abnormality does not vanish after 2 or 3 months off sports, HCM will be the final diagnosis. Genotyping may help (MYBPC3.....). What about the first degree family ECG and echo? Bradycardia is an epiphenomenom.

Thank you again for sharing with us this very tough case. Screening for cardiac diseases in elite athletes should begin very early before the age of full professionnal involvement.

Kind regards,

Prof Philippe Chevalier M.D. Ph.D. from Lyon France

philippe.chevalier@chu-lyon.fr

Querido Andrés,

El ECG con ondas Q muy finas y profundas sugiere fuertemente HCM. T bifásicas pueden no ser específicas. El eco muestra una HVI moderada. Si esta hipertrofia no desaparece después de 2 o 3 meses de vacaciones deportivas, HCM será el diagnóstico final.

El genotipado puede ayudar (MYBPC3. ....). ¿Tienen los ECGs y ECOs de los familiares de primer grado?

La bradicardia es un epifenómeno.

Gracias de nuevo por compartir con nosotros este caso muy difícil.

La detección de enfermedades cardiacas en los deportistas de élite debe comenzar muy temprano antes de la edad de la participación como profesional.

Un cordial saludo,

Phillippe

Recommendations for competitive sports participation in athletes with cardiovascular disease[1]:
Asymptomatic sinus bradycardia, sinus bradyarrhythmia, wandering pacemaker (PM), and sinus pauses are common in young athletes. A number of studies suggest that these arrhythmias in athletes are the consequence of increased vagal tone and withdrawal of sympathetic tone. Occasionally, marked sinus bradycardia (i.e. _40 b.p.m.) at rest, or sinus pauses >3 s can be found in asymptomatic, well-trained endurance athletes. These arrhythmias are usually benign, and evaluation may be limited to history, physical examination, and ECG. Treatment is usually not necessary. If marked bradycardia is associated with symptoms, such as lightheadedness, pre-syncope/syncope, or exertional fatigue, 24 h Holter monitoring and exercise testing are recommended. If structural heart disease is suspected, echocardiography (or another imaging technique) is mandatory. In selected cases, a deconditioning period of 1–2 months could be useful to clarify the clinical significance of extreme bradyarrhythmias.
The prevalence of first-degree AV block or second-degree AV block of the Wenckebach type (Mobitz type I) is high. The AV block typically occurs during sleep or at rest. In asymptomatic athletes without structural heart disease (as assessed by echocardiography) and with resolution of the AV block during exercise (as assessed by 24 h Holter monitoring and/or exercise testing), no further investigations and no therapy are indicated.
Rarely, Mobitz type II or third-degree AV block may be observed in athletes, which require a more comprehensive clinical and diagnostic evaluation. If these findings are associated with symptoms or with structural heart disease, PM implantation is recommended.
Dr. Guillermo Acosta Hospital San Juan Bautista Catamarca, Argentina.

1. **Pelliccia A, Fagard R, Bjørnstad HH, Anastassakis A, Arbustini E, Assanelli D.et al. Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology; Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Eur Heart J. 2005 Jul;26:1422-1445.**

Recomendações para a prática desportiva competitiva em atletas com doença cardiovascular[1]: Bradicardia sinusal assintomática, bradiarritmia sinusal, marcapasso mutável e pausas são comuns em atletas jovens. Vários estudos sugerem que estas arritmias em atletas são a consequência de um aumento do tônus vagal e diminuição concomitante do tônus simpático. Ocasionalmente, observa-se marcada bradicardia sinusal em repouso(< 40 bpm), ou pausas sinusais> 3 segundos. Quando encontradas em atletas de endurance ou sobretreinados assintomáticos são geralmente benignos. A avaliação deve ser limitada a história pessoal e familiar, exame físico e ECG. Não é necessário tratamento. Se a bradicardia acentuada está associada a sintomas, como tonturas, pre-síncope/síncope ou fadiga ao exercício indica-se um Holter de 24hrs e teste ergométrico. Se uma doença cardíaca estrutural é suspeita, o ecocardiograma (ou outra técnica de imagem) é obrigatório.

Em casos selecionados, um período de 1-2 meses de descondicionamento pode ser útil para esclarecer o significado clínico de bradiarritmias extremas ou paradas sinusais longas.

A prevalência de bloqueio AV de primeiro grau ou de bloqueio AV de segundo grau do tipo Wenckebach (Mobitz tipo I) é alta e ocorre normalmente durante o sono ou em repouso.

Em atletas assintomáticos e sem cardiopatia estrutural (avaliada por ecocardiografia) e com resolução do bloqueio AV perante o exercício (avaliada pelo monitoramento 24hrs Holter e/ou teste ergométrico), não mais investigações e nenhuma terapia é indicada.

Raramente, bloqueio AV tipo Mobitz II ou de terceiro grau pode ser observado em atletas, e sua exigem uma avaliação clínica mais abrangente. Se estes estão associados a sintomas e doença cardíaca estrutural, a implantação de um Marca-passos é recomendada.

Dr. Guillermo Acosta Hospital San Juan Bautista de Catamarca, na Argentina.

1. **Pelliccia A, Fagard R, Bjørnstad HH, Anastassakis A, Arbustini E, Assanelli D.et al. Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology; Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Eur Heart J. 2005 Jul;26:1422-1445.**

FINAL CONCLUSIONS

CONCLUSÕES FINAIS

ECG DIAGNOSIS

1. Rhythm Sinus bradychardia
2. HR: 51bpm
3. QRS axis: + 100º. Vertical axis.
4. **Notch or slurring contour of terminal portion of the QRS complex on inferior leads**
5. Relatively deep but narrow q waves in the left precordial leads
6. Prominent R wave in V_1 and concomitant deep and narrow Q waves in left precordial leads V_5-V_6. (hypertrophy of first middle septal vector) SEPTAL HYPERTROPHY.
7. Early repolarization pattern: **J point and ST segment elevation concave to the top in inferior leads followed by concordant T wave. At least two adjacent precordial leads show ST segment elevation, with values ≥1 mm (2mm).** Negative asymmetrical T waves from V_1 to V_3 and bifid T wave on V_4 lead. In Spanish study[1], 26 athletes with negative T waves ≥ 2mm in three or more ECG leads at rest were studied. All athletes had ECG at rest simulating myocardial ischemia or "pseudoischemia" with a tendency to normalize during exercise. All have no evidence of heart disease and no adverse effects on the follow-up Myocardial perfusion studies were normal in the studied athletes. Antimyosin studies showed mild and diffuse myocardial radiotracer uptake in 15 athletes (68%). No adverse clinical events were observed in the follow-up. These results suggest that marked repolarization abnormalities have no clinical or pathological implications in athletes and should, therefore, not preclude physical training or participation in sporting events.

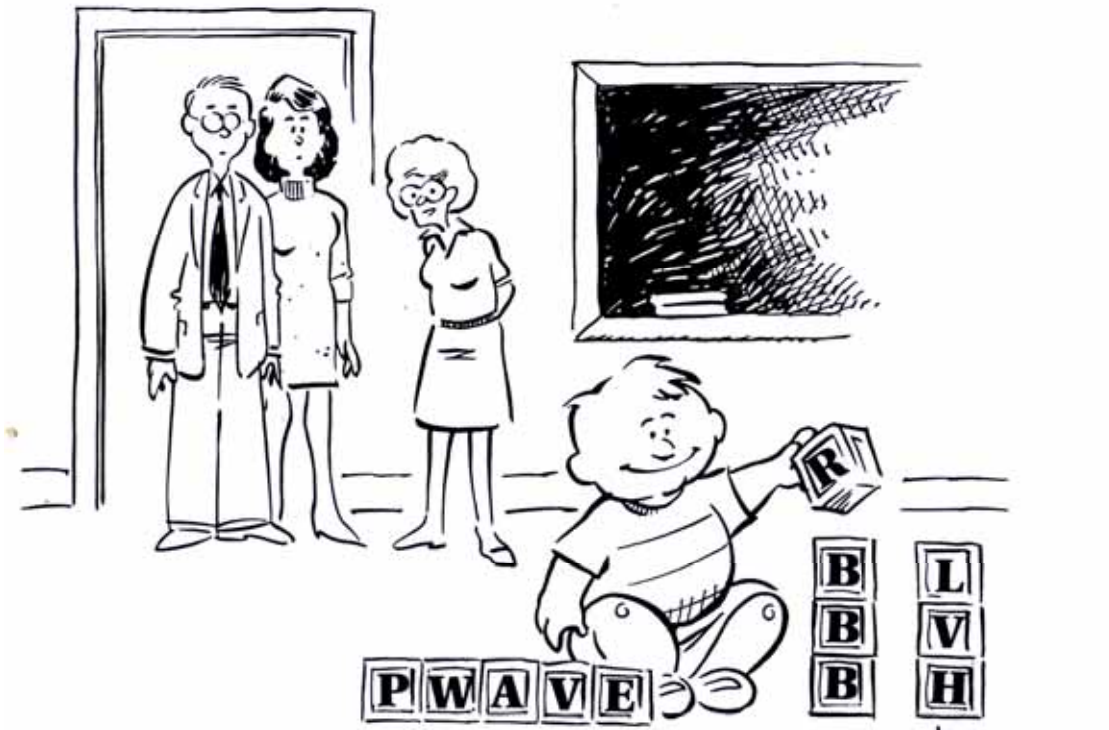
The red color letters are frequent components of early repolarization pattern. This pattern is very Frequent in athlete´s heart.

1. **Serra-Grima R, Estorch M, Carrió I, Subirana M, Bernà L, Prat T. Marked ventricular repolarization abnormalities in highly trained athletes' electrocardiograms: clinical and prognostic implications.JAm Coll Cardiol. 2000 Oct;36:1310-1316.**

ECG DIAGNÓSTICO
Ritmo: bradicardia sinusal
FC: 51bpm
SÂQRS (eixo do QRS): + 100º. O eixo vertical: Entalhe ou borramento no contorno da porção terminal do R nas derivações inferiores. Ondas Q relativamente profundas porem estreitas em precordiais esquerdas V5 e V6. Onda R proeminente em V1 e concomitante Q profundas em precordiais esquerdas V5-V6. (Assinalando hipertrofia do septo médio: vetor septal aumentado)
Padrão de repolarização precoce: elevação ponto J e do segmento ST de concavidade superior nas derivações inferiores seguido por ondas T positivas concordantes em pelo menos duas derivações precordiais adjacentes ≥ 1 mm.
Ondas T negativas assimétricas de V1 a V3 e bífidas em V4. *Comentários: em um estudo espanhol com 26 atletas que apresentavam ondas T negativas ≥ 2mm em três ou mais derivações no ECG de repouso. Estas ondas T simulavam isquemia miocárdica ou "pseudo-isquemia" com tendência a normalização durante o exercício. Nenhum mostrou evidência de doença cardíaca ou teve eventos adversos no seguimento, e o estudo de perfussão miocárdica foi normal.*
Estes resultados assinalam que alterações de repolarização importantes da onda T não têm implicações patológicas em atletas e portanto, não deve impedir-se o treinamento físico ou a participação competitiva em eventos esportivos.

As letras de cor vermelha são freqüentes componentes do padrão de repolarização precoce. Este padrão é muito freqüente no coração do atleta.

1. **Serra-Grima R, Estorch M, Carrió I, Subirana M, Bernà L, Prat T. Marked ventricular repolarization abnormalities in highly trained athletes' electrocardiograms: clinical and prognostic implications.JAm Coll Cardiol. 2000 Oct;36:1310-1316.**



"Our Pre-School Career Aptitude Tests indicate little Mark will make a very fine cardiologist".

"Nosso teste pré-escolar de aptidão profissional indica que o pequeno Mark será um excelente cardiologista."

In a study of 1282 professional football players, frequent and complex VT unassociated with structural heart disease and no adverse follow-up were recorded in 355 trained athletes[1].

Em um estudo realizado em 1282 jogadores de futebol profissional, observou-se freqüentes e complexas TV sem associação com cardiopatia estrutural e sem eventos adversos no seguimento. Estas TV complexas foram registradas em 355 atletas treinados.[1]

1. **Choo JK, Abernethy WB 3rd, Hutter AM Jr. Electrocardiographic observations in professional football players. Am J Cardiol. 2002 Jul 15;90(2):198-200.**

The resting ECG is an essential tool in modern cardiological practice and its recording is part of the basic cardiovascular examination. Nevertheless, it may be a cause of concern even in asymptomatic individuals when it is not entirely normal. We present the case of a young sports soccer player with a doubtfull resting ECG and the consequent dilemmas in relation to eligibility for competitive athletic activity. The heart continuously adapts to adjust its output to a continuum of pathophysiological situations ensuring adequate blood distribution. These situations range from high performance in well-trained sportsman to failure in a variety of cardiac syndromes. Changes in the concentration of intracellular Ca^{2+} are crucial. They have immediate and late effects that can be oversimplified as follows. Immediate effects result from abrupt and large variations in Ca^{2+} triggering contraction after binding to the contractile proteins. These variations are involved in the process known to as excitation–contraction coupling. In contrast, the late effects involve a process that is, by analogy, referred to as excitation–transcription coupling. This process involves activation of gene expression by Ca^{2+}. In this scheme, specific and localised elevations of Ca^{2+} can be converted into changes in gene expression with long-term effects on the adaptation of the heart to a sustained stimulus. There is emerging evidence of an extraordinary diversity of responses, depending on the location, intensity, and duration of Ca^{2+} signals that can be activated during pathology. Whereas alterations of cellular and molecular mechanisms underlying chronic pathology are relatively well defined, the initial changes and their hierarchy are unknown. However, the actual picture suggests promising perspectives for new therapeutic interventions on old targets or new strategies. Markers of overtraining do not parallel a decrease in performance and should be interpreted with caution. The document takes note of the 25-year Italian experience on systematic pre-participation screening of competitive athletes and focuses on relevant issues, mostly regarding the relative risk, causes, and prevalence of sudden death in athletes; the efficacy, feasibility, and cost-effectiveness of population-based pre-participation cardiovascular screening; the key role of 12-lead ECG for identification of cardiovascular diseases such as cardiomyopathies and channelopathies at risk of sudden death during sports; and the potential of preventing fatal events.

The main purpose of the consensus document was to reinforce the principle of the need for pre-participation medical clearance of all young athletes involved in organized sports programmes, on the basis of

i) The proven efficacy of systematic screening by 12-lead ECG (in addition to history and physical examination) to identify hypertrophic cardiomyopathy-the leading cause of sports-related sudden death-and to prevent athletic field fatalities
ii) The potential screening ability in detecting other lethal cardiovascular diseases presenting with ECG abnormalities.

The consensus document recommends the implementation of a common European screening protocol essentially based on 12-lead ECG[1].

Of 33,735 athletes, 22 were found to have HCM and were disqualified (3,5%of the disqualified conditions). In addition 238 were disqualified for rhythm and conduction abnormalities, 168 for systemic hypertension, 133 for valvular disease, and 60 for various others disease. A follow-up of this study , published in 2006, has shown that the incidence of SD in athletes has decreased from 3.5 per 100,000 person-years in 1979 to 1980 to 0,4 per 100.000 person-year in 2003-2004. The death rate in nonathletes has remained relatively stable at around 0.8 per 100.000 person-years, suggesting that the screening process reduced the incidence of SCD in athletes[1].

1. **Corrado D, Pelliccia A, Bjørnstad HH, Vanhees L, Biffi A, Borjesson M, et al. Cardiovascular pre-participation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol. Consensus Statement of the Study Group of Sport Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Eur Heart J. 2005 Mar;26:516-524.**
2. **Corrado D, Basso C, Pavei A, Michieli P, Schiavon M, Thiene G. Trends in sudden cardiovascular death in young competitive athletes after implementation of a preparticipation screening program.JAMA. 2006 Oct 4;296:1593-601.**

According to the statements from the International Cardiological Committees on Eligibility for Sports, athletes with a clinical diagnosis of HCM should be excluded from most competitive sports, with the possible exception of those of low intensity.

Clinical distinctions between physiological athlete's heart and pathological conditions such as HCM have critical implications especially for trained athletes. Even if the standard two-dimensional echocardiography represents an irreplaceable method in the evaluation of cardiac adaptations to physical exercise, the data currently available suggest the usefulness of Doppler myocardial imaging (DMI) in the assessment of the myocardial systolic and diastolic function of the athlete's heart.

The combined use of standard two-dimensional echocardiography and Doppler myocardial imaging DMI may be taken into account for a valid, non-invasive and easily repeatable evaluation of both physiological and pathological ventricular hypertrophy, and in selecting a subgroup of HCM patients at higher risk of cardiac events.

In particular, DMI analysis in the trained individual has demonstrated an interesting opportunity for:

1) The differential diagnosis from pathological left ventricular hypertrophy due to HCM;
2) The prediction of cardiac performance during physical effort;
3) The evaluation of bi-ventricular interaction;
4) The analysis of myocardial adaptations to various training protocols; and
5) The early identification of specific genotypes associated with cardiomyopathies.

1. **Caso P, D'Andrea A, Caso I, Severino S, Calabrò P, Allocca F, et al. The athlete's heart and hypertrophic cardiomyopathy: two conditions which may be misdiagnosed and coexistent. Which parameters should be analysed to distinguish one disease from the other? J Cardiovasc Med (Hagerstown). 2006 Apr;7:257-266.**

THE PREPARTICIPATION SCREENING PROGRAMME

1. History:

- Personal: Personal history of murmur or congenital heart disease in childhood, matternal rubella, exposition to toxics used or enviromental, dizziness or syncope during or after exercise, syncope, palpitations during or after exercise may be a sign of arrhythmia, intolerance to exercise, precordialgia, dyspnea: Excessive/progressive dyspnea may indicate valvular diseases, pulmonary disease, or structural anomalies may indicate the presence of: HCM, dromotropic disorder, MVP, aortic stenosis or arrhythmia Precordialgia intra- or post-strain may indicate early coronary atherosclerosis. Ask questions about the current or past use of legal (tobacco, alcohol) and illegal drugs. Recent history of virus infection may lead to symptoms compatible to myocarditis;
- Family history Ask questions about SD in first-degree relatives under 45 years old Ask questions about the knowledge in the family about HCM, LQTS, Marfan-type somatic habit, sindactily, etc

2. Physical examination: Anthropometrical evaluation: weight, height, BP and percentage of body fat; identification and characterization (intensity, location and time of cycle) of murmurs and arrhythmias, standing and in supine position; Recognize phenotypes: e.g. Marfand, Noonan and Holt-Oram syndrome, supravalvular aortic stenosis, Williams syndrome. Measurement of BP in superior and inferior limbs, and assessment of femoral, radial and foot pulses to exclude Aorta coarctation.Auscultation must be performed in decubitus and standing to identify murmurs influenced by dynamic obstruction in the LV outflow tract; detection of extracardiac clicks and sounds; Muscle-skeletal aptitude. Try to detect medical conditions or skeletal muscles that may predispose to injuries or diseases during a competition.
3. Resting 12-lead ECG

1. Hevia AC, Fernández MM, Palacio JM, Martín EH, Castro MG, Reguero JJ. ECG as a part of the preparticipation screening programme: an old and still present international dilemma. Br J Sports Med. 2010 Jul 15. [Epub ahead of print]

MOST FREQUENT CHARACTERISTICS OF ECG IN ATHLETES

1) RHYTHM: Sinus, junctional or rarely ventricular. Variable pacemaker or rhythm of left atrium. Junctional rhythm is present in 0.31% (in the general population in 0.02%). Phasic or respiratory sinus arrhythmia: present in 60% (in the population in athletes in 2.4%).
Long sinus pauses: they are frequent (> 3 seconds);
2) HEART RATE: Sinus bradycardia in more than 50% of the cases. HR of 30 to 40 bpm in rest are not rare. In highly trained athletes, there are descriptions of HR of 25 bpm. ETIOLOGY: vagal hypertone, decrease in resting sympathetic tone and intrinsic component of bradycardia;
P WAVE: Increase of voltage and notches are described;
4) PR INTERVAL: 1st degree AV block: 5% and 30% (in non athletes, 0.65%). When the PR interval does not reach the value as a criterion for 1st degree AV block, it is relatively prolonged. The PR interval normalizes or even gets smaller after exercise;
5) 2nd degree AV block:
a) Mobitz Type I or Wenckebach: it is observed in 10% (in non athletes < 1 in 30,000 or 0.003%), and it disappears invariably during exercise and atropine;
b) Mobitz Type II; AV dissociation; Complete or 3rd degree AV block: 5 each 12,000 athletes.
6) SAQRS or QRS axis: tendency to vertical position;
7) Presence of voltage criteria for LVH: SV1+RV5 > 35mm (Sokolow and Lyon index);
8) Possible pattern of RVH: RV1+SV5 >10.5 mm between 18% and 69% of the cases. RVH manifests by a diastolic pattern translated by minimal degrees of IRBBB. The IRBBB is observed in 15% of athletes; Absence of progression of increase of voltage of r or R wave with QR pattern from V1 to V3: pattern of pseudo infarction in anterior wall.
Pattern of Complete RBBB(rare);

ST SEGMENT

Pattern of Early Repolarization Variant or Early repolarization pattern

It is described in four patterns:

a) J point and ST segment elevation followed by peaked T wave from V4 to V6 and in the inferior wall (2.4% to 44%);

b) J point and ST segment depression (rare);

c) J point and ST segment elevation followed by inverted T wave;

d) Disappearance of ST segment elevation after exercise.

T WAVE

Juvenile pattern of T wave.

Inverted and asymmetrical T wave in left leads: I, aVL, V5 and V6, secondary to physiologic LVH.

Negative or biphasic T waves from V1 to V3 and/or in the inferior wall.

Frequent “normalization” of T wave before strain. This type of response is not observed in HCM or in CAD.

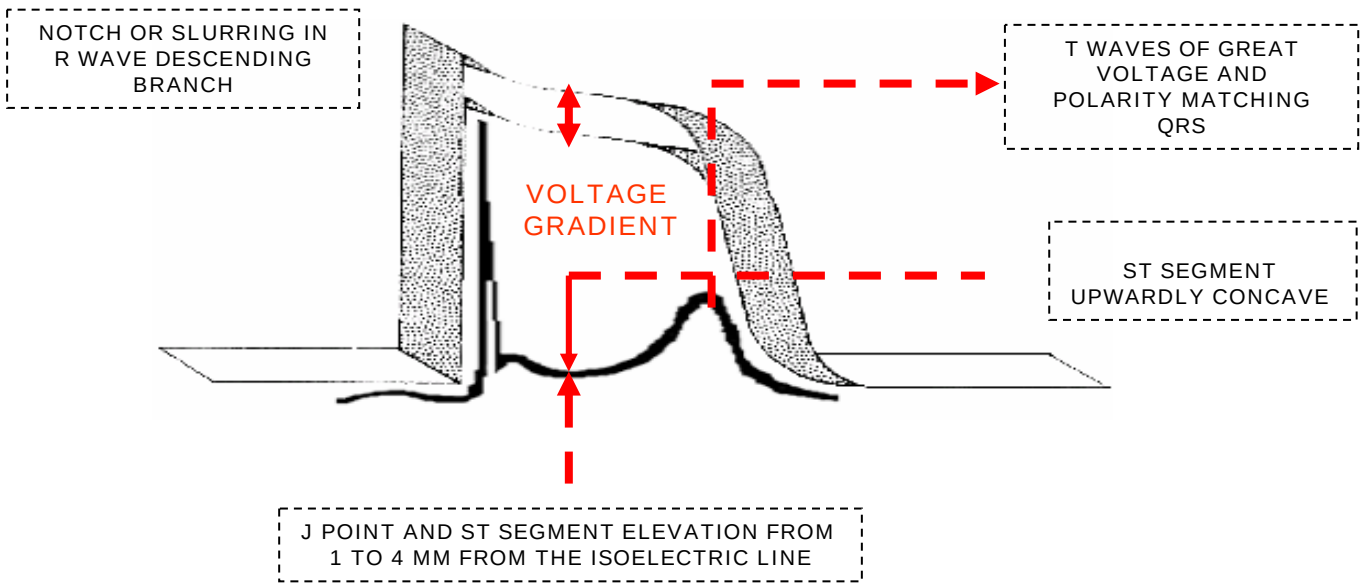
Myocardial perfusion imaging associated to exercise stress test always negative.

Characteristic reversion of ECG “alterations” in cases of interruption of competitive activity;

U WAVE

After the T wave, a rounded deflection is observed mostly in precordial leads V3 and V4, called U wave. Usually, it has the same polarity as T wave. It is always, in normal cases, positive in D1, D2 and from V2 through V6. Its amplitude is inversely proportional to HR, BEING GREATER IN ATHLETES with bradycardia, and smaller in children with tachycardia.

In ERP, there is a voltage gradient, however, no dispersion of duration of action potentials in ventricular wall thickness. For this reason, these patients showed ST segment elevation with no tendency to develop arrhythmias. HR: sinus bradicardia is frequent; QRSaxis, ST segment and T wave, are oriented in the same direction in the FP;
Deep and narrow Q waves followed by R wave of great voltage in left precordial leads;
Notch or slurring of R wave descending branch;
Transition area in precordial leads of sudden occurrence;



J point and ST segment elevation, usually < 2 mm (exceptionally it may be > 5 mm) of superior concavity in middle and/or left precordial leads and possibly in inferior leads; Possible reduction in J point and ST segment elevation by sympathetic action and sympathomimetic drugs;
Absence of reciprocal or mirror image (exception in VR lead); Symmetrical T waves, with great width and polarity matching QRS;

SUMMARY ECG/SAECG/VCG IN ATHLETES

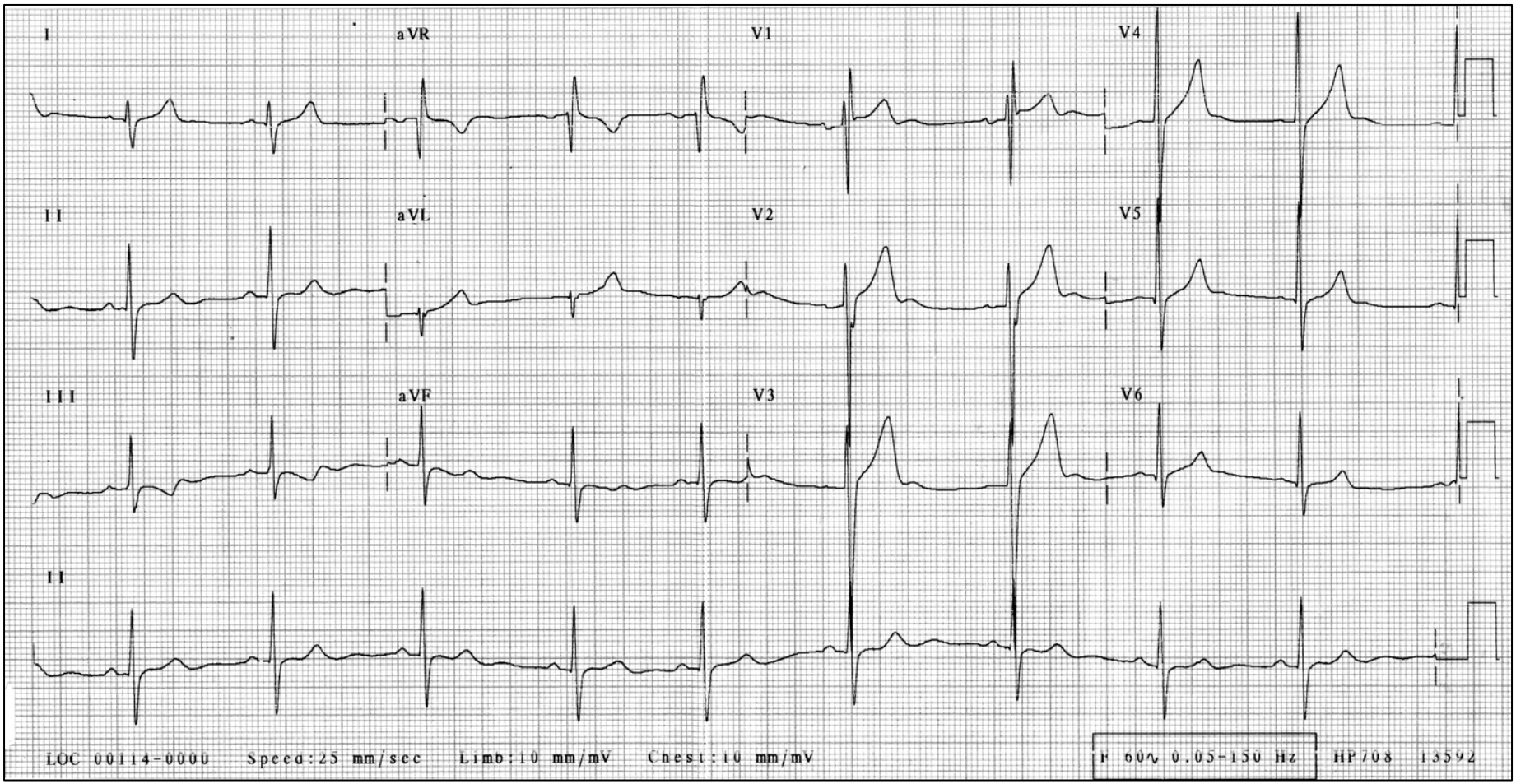
1) Sinus bradycardia.
2) Sinus arrhythmia.
3) P wave with notches and of greater voltage.
4) First degree AV block: 6% to 36%.
5) 2nd degree AV block, Wenckebach type: Mobitz Type I (0.125% to 10%).
6) IRBBB or end conduction delay.
7) Voltage or axis criterion for RVE.
8) Voltage criterion for LVE.
9) J point and ST segment elevation or depression.
10) QT interval in the superior borderline of normality.
11) T wave of increased voltage, peaked and inverted.
12) Atrial fibrillation and flutter[1].
13) Junctional rhythm.
14) SAECG: Presence of late potentials in 10% of the cases against 1.4% in the population of athletes[1];
15) VCG: Increase of anterior forces in almost all cases;Dislocation of QRS loop to the front and left in the HPT loop not matching QRS loop;

1) **Furlanello, et al. J Cardiovasc Electrophysiol 1998;9 (Suppl 8); S63-S68.**
2) **Borbola, J & Denes, P. Late potentials in patents with sustained ventricular tachycardia. In: El-Sherif, N.; Turitto, G (eds). High-Resolution Electrocardiography. Mount Kisco (NY):Futura, 495-520, 1992**

ARRHYTHMIAS IN THE HEARTS OF ATHLETES AND COMPARATIVE INCIDENCE WITH THE GENERAL POPULATION

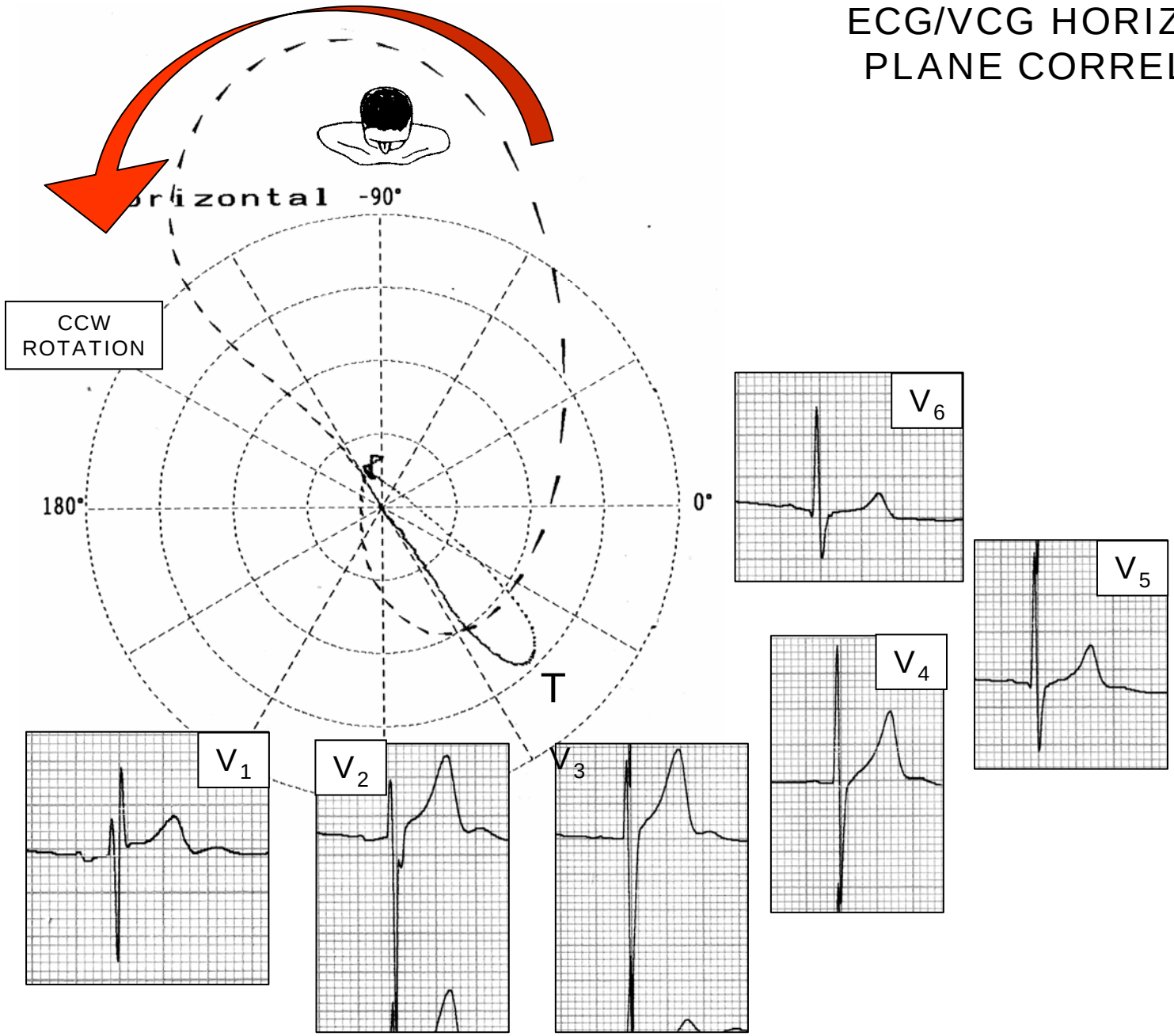
ARRHYTHMIA	GENERAL POPULATION %	ATHLETES %
Sinus Bradycardia	23.7	50-85
Sinus Arrhythmia	2.4-20	13.5-69
Atrial Variable Pacemaker	NOT AVAILABLE	7.4-19
1st degree AV block	0.65	6-33
Mobitz Type 1	0.003	0.125-10
Mobitz Type II	0.003	NOT REPORTED
3rd degree AV block	0.0002	0.017
Junctional Rhythm	0.06	0.31-7.0

Name: ASF Sex: Male Age: 18 yo. Race: Afro-Descendent
Weight: 97 Kg Height:1,93 m Biotype: Asthenic Date: 07/30/2008
Professional soccer player: forward

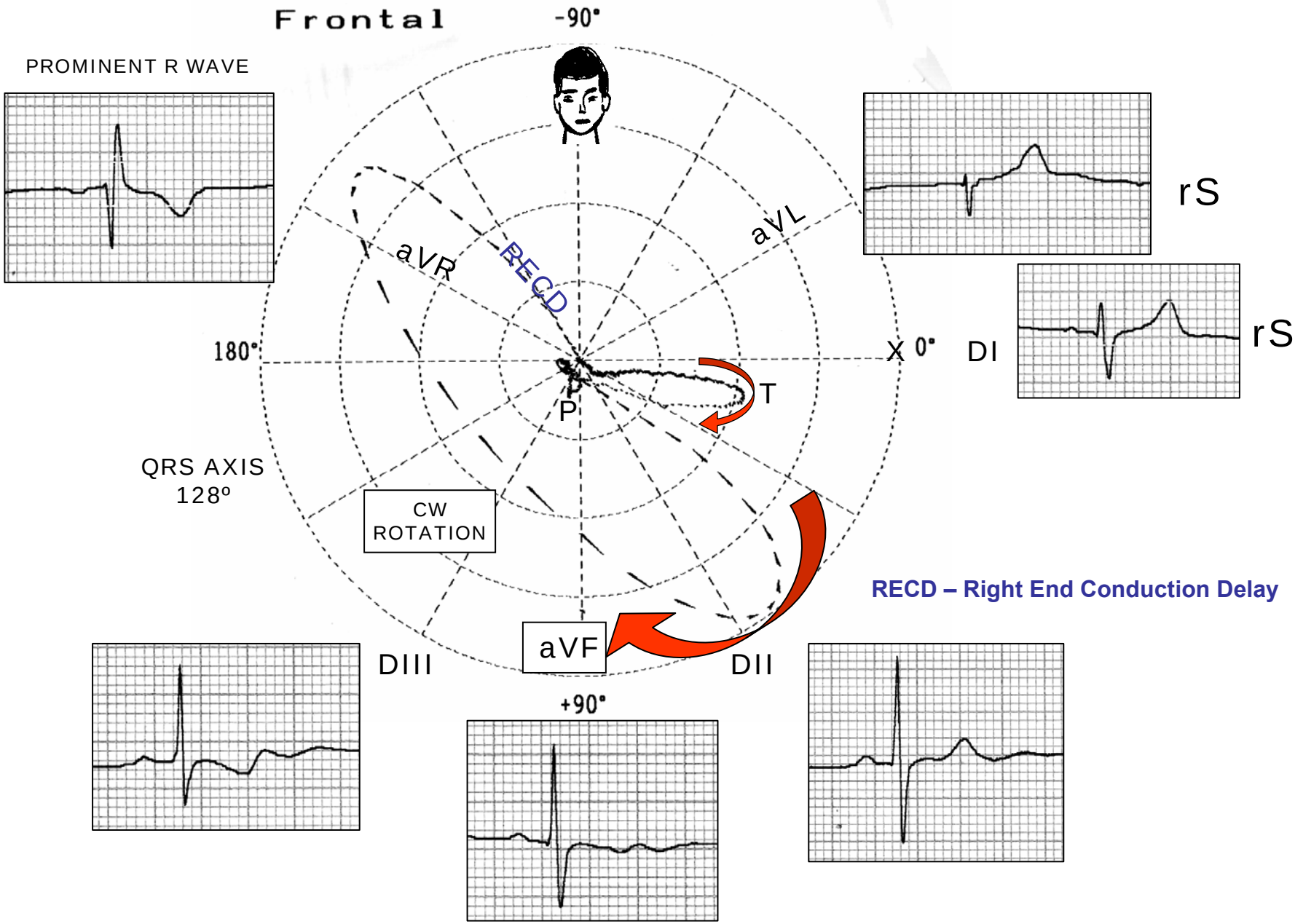


Clinical Diagnosis: Familiar hypertension (BP: 150x105mmHg). Recent diagnosis.
ECG diagnosis: HR: 58bpm, P axis: 66bpm, PR interval: 168ms, QRSd: 97 ms, QRS axis: +128º, QT: 406ms, QTc: 399ms, T axis: +5º.
Which is the diagnosis ?

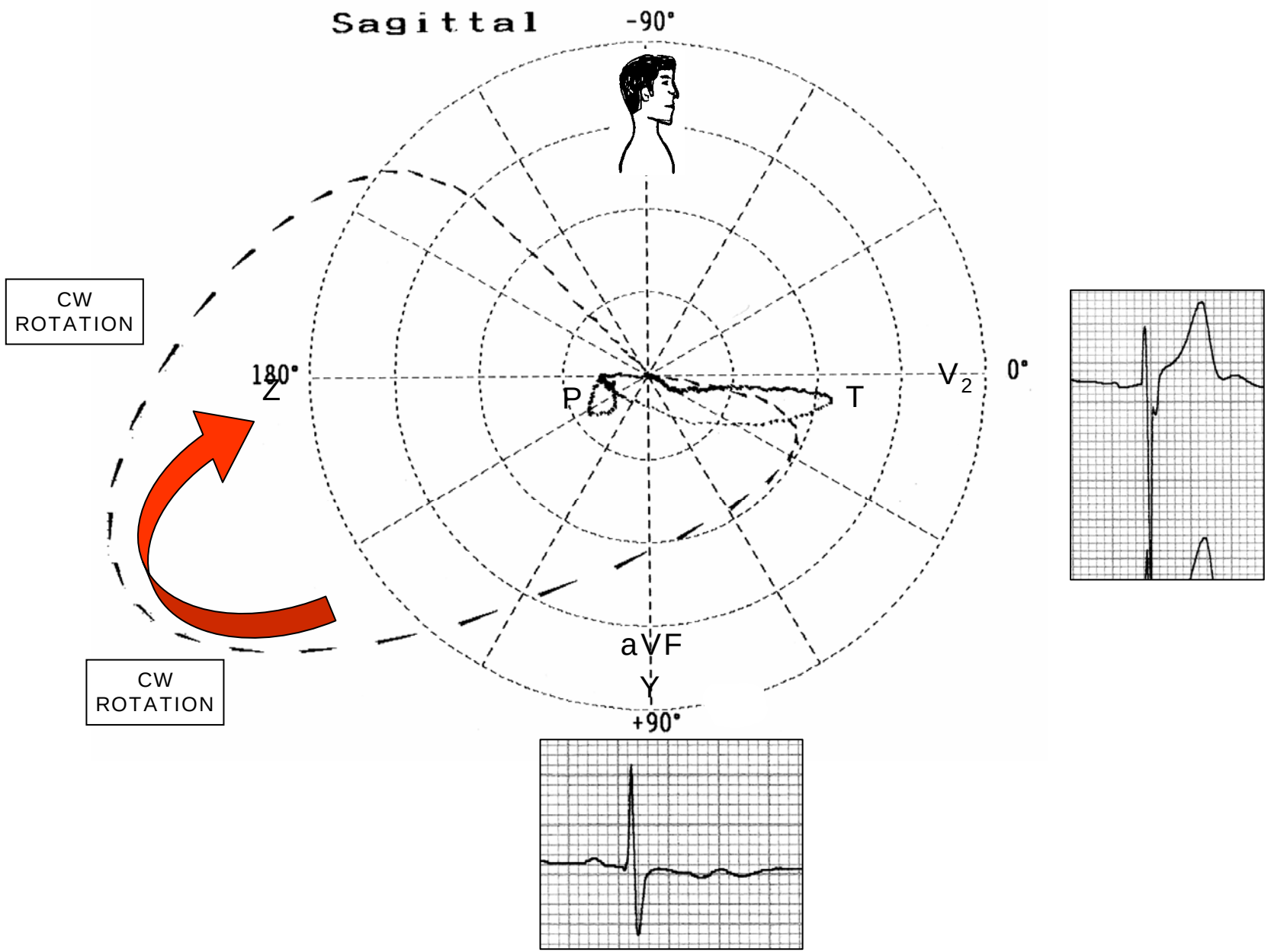
ECG/VCG HORIZONTAL PLANE CORRELATION



ECG/VCG FRONTAL PLANE CORRELATION

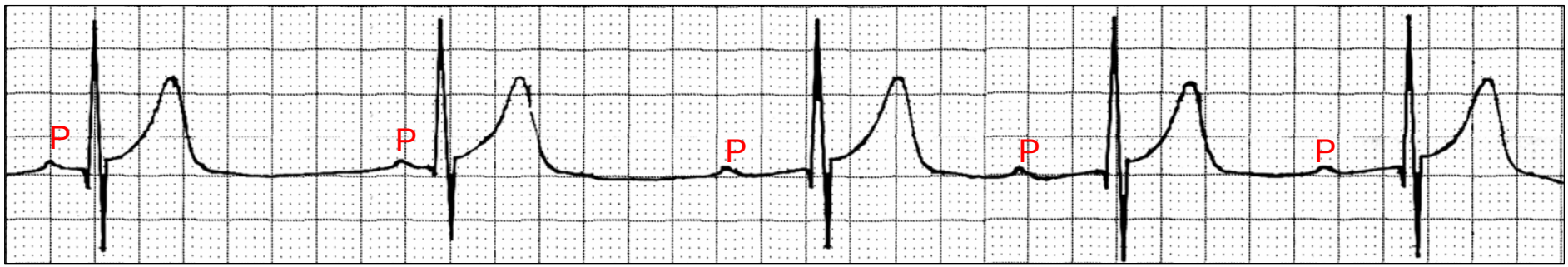


ECG/VCG RIGHT SAGITTAL PLANE CORRELATION



HOLTER RECORDING
1ST DEGREE AV BLOCK

Name: B . C. **Sex:** Male **Age:** 22 **Race:** Black **Weight:** 74 Kg.
Height: 1.82 m **Biotype:** Athletic **Date:** 01/04/2002
Time: 2:50:12 AM Patient sleeping. **Profession:** Marathon runner



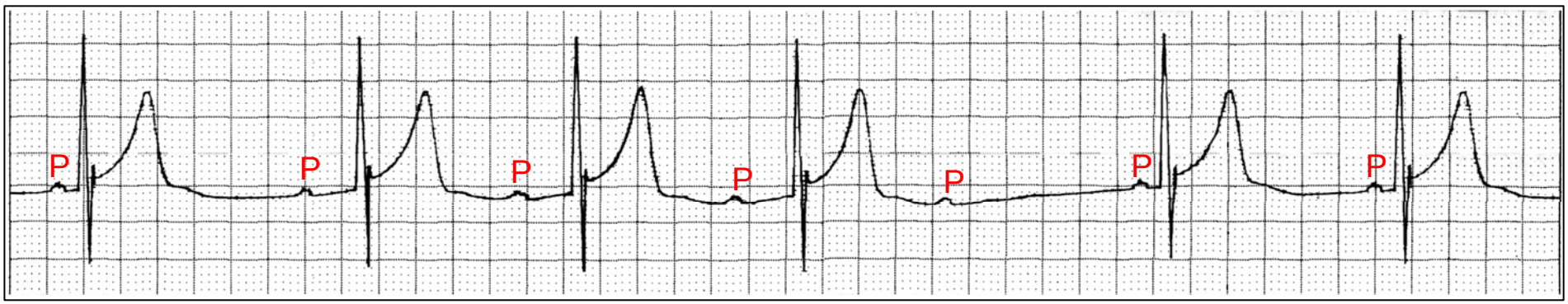
Heart rate of 38 bpm.
1st degree AV block usually observed for a few seconds, as in this case, where it is present only in the three last beats.

1st degree AV block is observed in average between 10% and 33% of athletes (1), generally very briefly. In non-athletes it is around 0.65%.

1) Smith WG, et al. Br Heart J 1964:469-476.

HOLTER RECORDING

Name: A . S. **Sex:** Male **Age:** 26 **Race:** Black **Weight:** 64 Kg.
Height: 1.68 m **Biotype:** Athletic **Date:** 05/01/2003
Time: 3:42:30 AM Patient sleeping. **Profession:** long distance runner



Gradual prolongation of PR interval until the 5th P wave is not conducted: 2nd degree AV block; Wenckebach or Mobitz Type I.

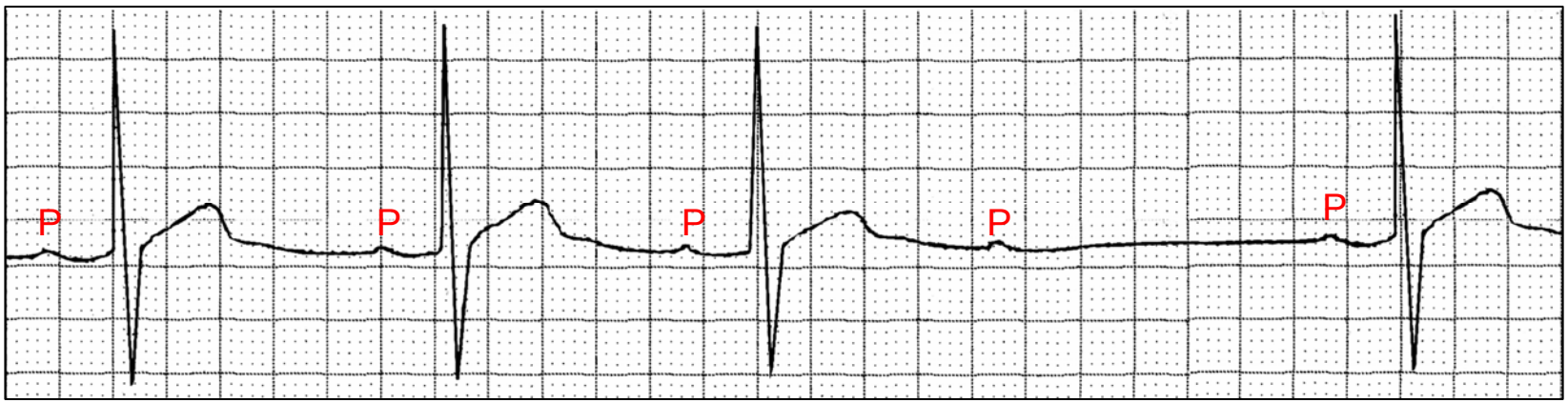
This modality of dromotropic disorder is observed in more than a 20% of elite athletes (1). In the general population, 2nd degree AV block Type I & II is observed and 1 each 30,000 people or 0.003 %

1) Viitasalo MT, et al. Br Heart J. 1982;47:213-220.

HOLTER RECORDING

2ND DEGREE AV BLOCK, MOBITZ TYPE II WITH NARROW QRS

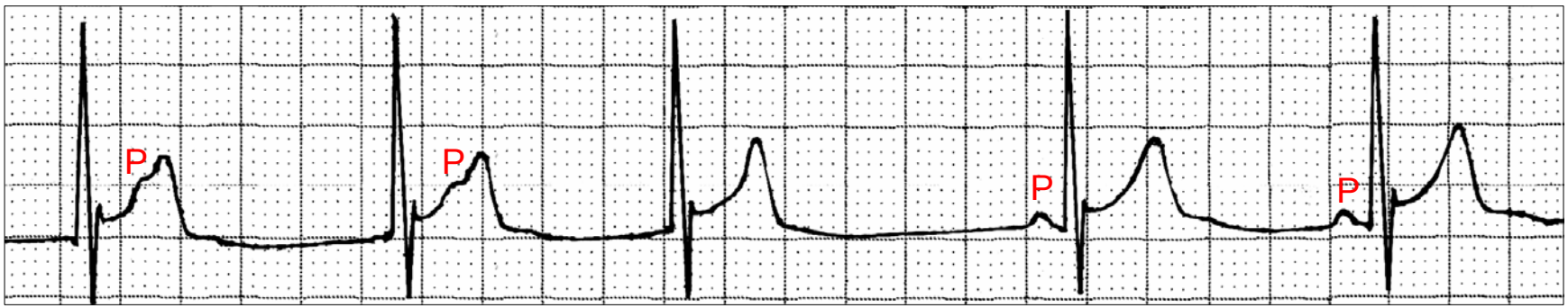
Name: E . J. **Sex:** Male **Age:** 26 **Race:** White **Weight:** 70 Kg.
Height: 1.72 m **Biotype:** Athletic **Date:** 25/01/2001
Time: 1:52:10 AM Patient sleeping **Profession:** Long distance runner



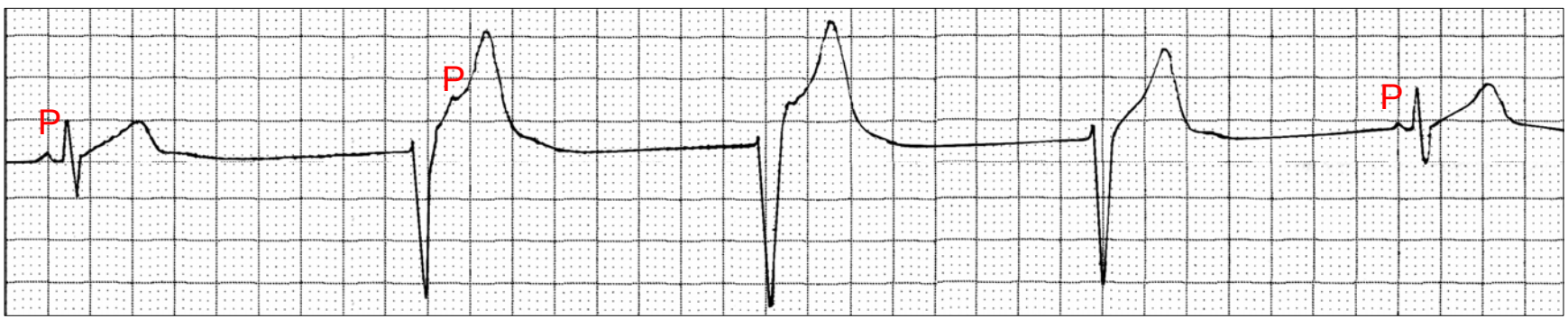
PR interval remains constant until a P wave is not conducted. This type of block is observed in 7% of the cases in athletes of enduro. Fixed or constant PR interval: it does not exist, progressive prolongation of PR, with the block occurring suddenly. In general, 2nd degree AV block type II with narrow QRS is observed in 35% of the cases and in the remaining 65%, the QRS is long.

HOLTER RECORDING

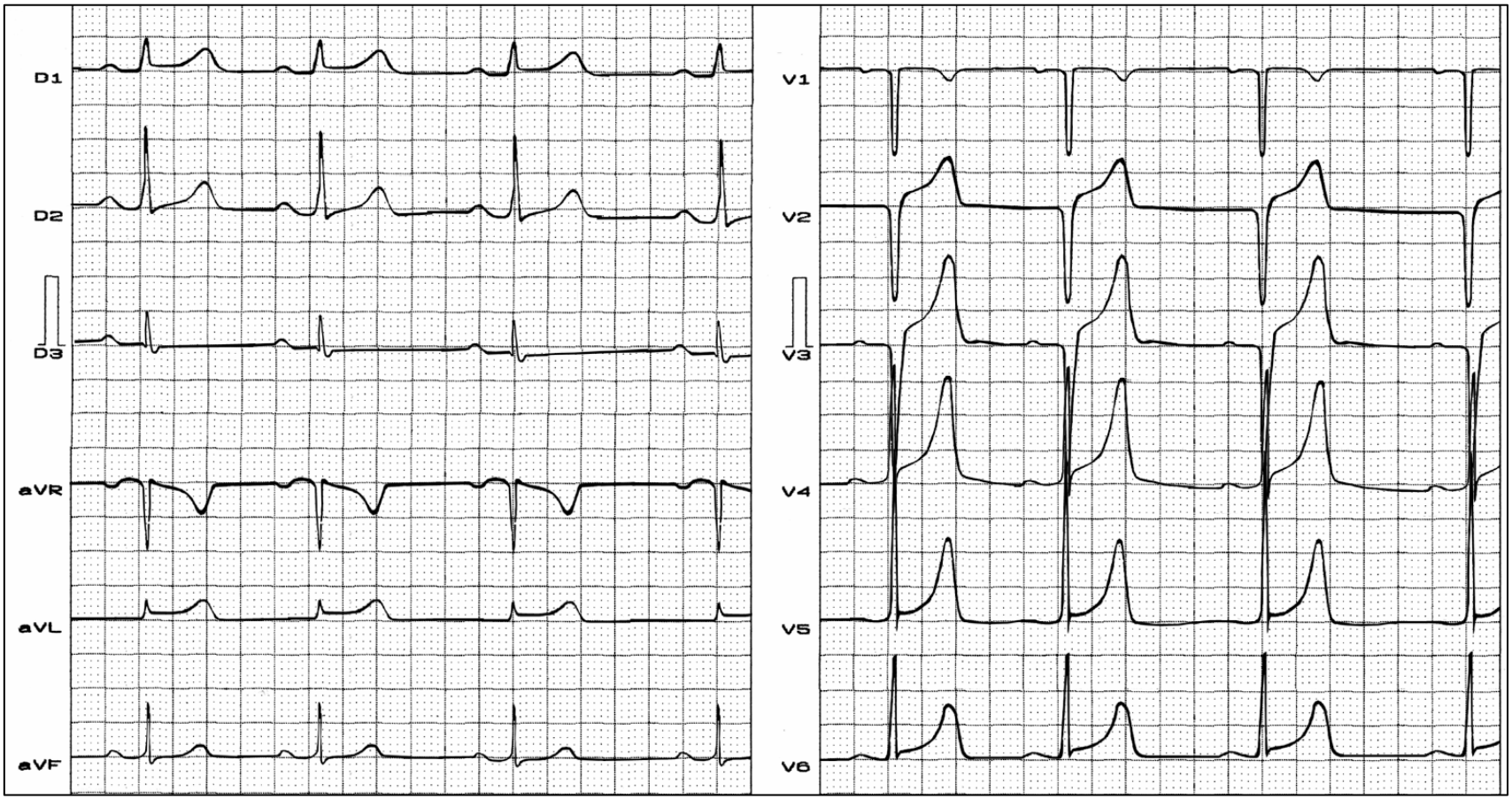
ATRIOVENTRICULAR DISSOCIATION (DISSOCIATION BY INTERFERENCE) WITH JUNCTIONAL SCAPE RHYTHM



ATRIOVENTRICULAR DISSOCIATION (DISSOCIATION BY INTERFERENCE) WITH SCAPE VENTRICULAR RHYTHM

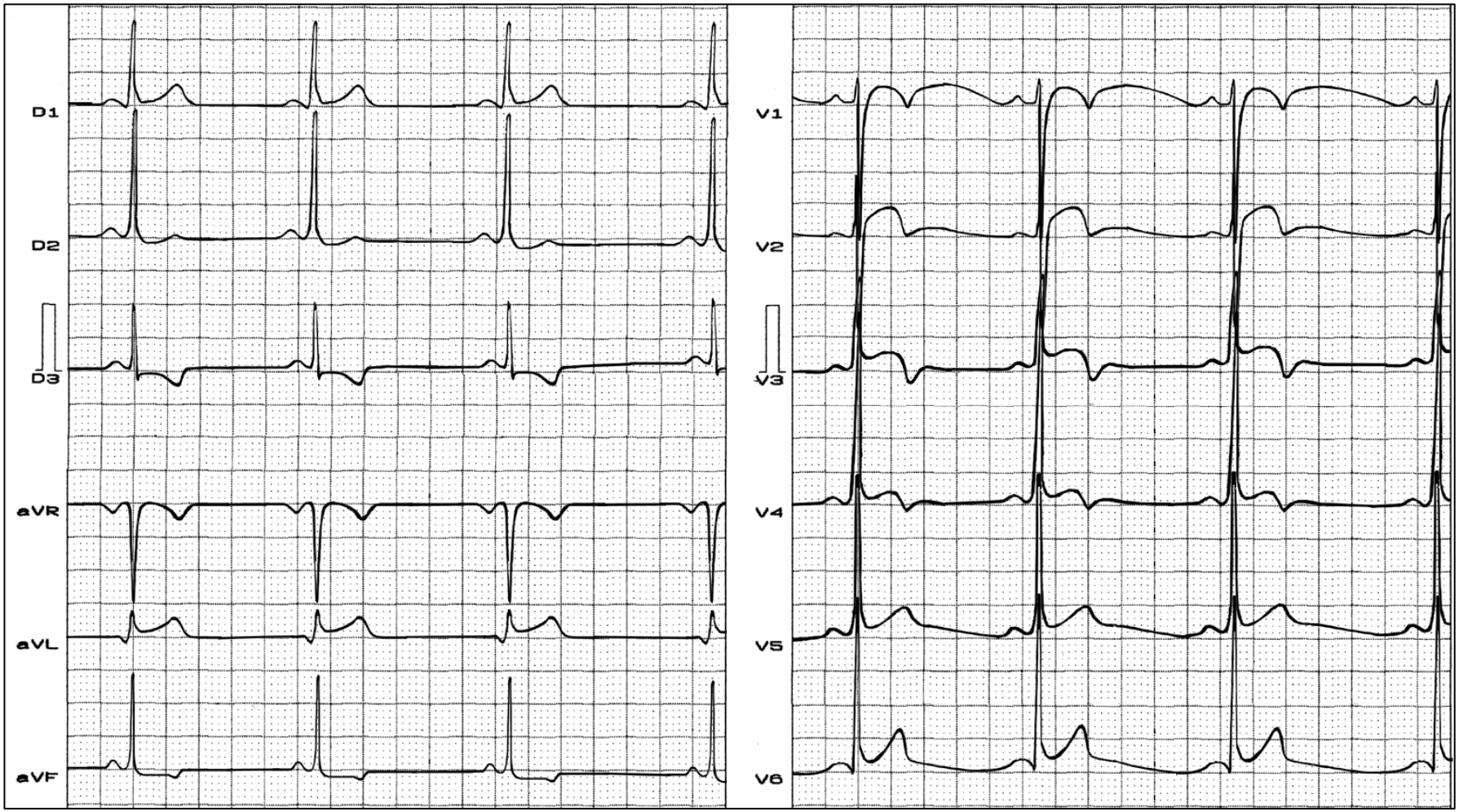


Name: BCA **Age:** 22y **Sex:** Male **Race:** Black **Weight:** 82 kg
Height: 1.96m **Biotype:** Athletic Profession: professional basketball player **Date:** 2/09/2001



CLINICAL DIAGNOSIS: athlete's heart. Normal variant.
ECG DIAGNOSIS: sinus rhythm; HR: between 50 bpm and 57 bpm: phasic or respiratory sinus bradyarrhythmia; QS from V1 to V3: pattern of pseudo infarction in antero-septal wall. Peaked T waves from V3 to V6. Normal X-rays of chest and echocardiogram.

Name: BCW **Age:** 24y **Sex:** Male **Race:** Black **Weight:** 86 kg
Height: 2.02 m **Biotype:** Asthenic **Profession:** professional basketball player. **Date:** 05/01/1999



CLINICAL DIAGNOSIS: healthy patient. Tracing obtained in a periodical evaluation.
ECG DIAGNOSIS: sinus bradycardia, phasic sinus arrhythmia. Positive voltage criterion for LVE. SV_1 or V_2+RV_5 or V_6 >35 mm (Index of Sokolow Lyon). ST segment elevation from V_2 to V_6 and with negative T from V_1 to V_4. Early repolarization, pattern of pseudo injury and anterior subepicardial ischemia. Normal chest X-rays and echocardiogram.

CLASSIFICATION OF ABNORMALITIES OF THE ATHLETE'S ECG

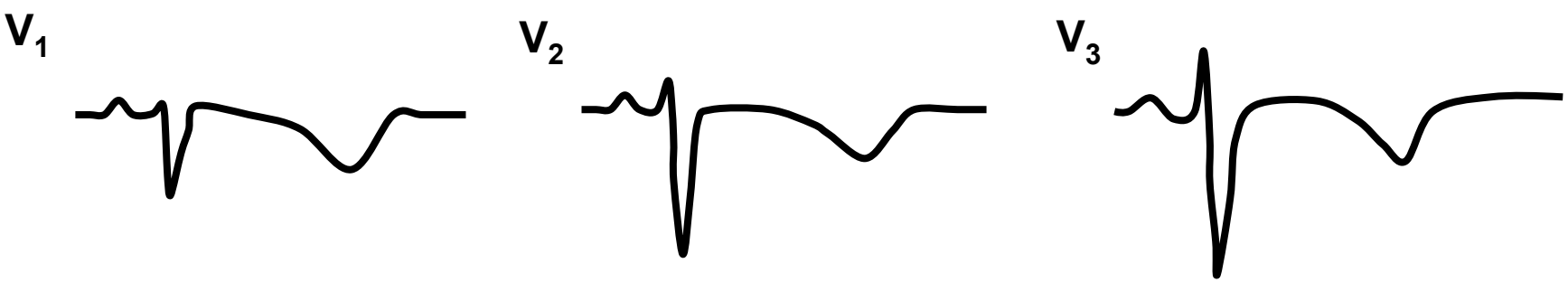
TRAINING-RELATED ECG CHANGES	UNCOMMON AND TRAINING-UNRELATED ECG CHANGES <5%
Sinus bradycardia (high prevalence)	Marked bradycardia associated with symptoms, such as lightheadedness, pre-syncope/syncope, or exertional fatigue,
Sinus phasic arrhythmia (high prevalence)	Symptomatic sinus pauses >3 s Convulsion, syncope, near syncope lightheadedness.
Asymptomatic sinus pauses >3 s Mainly nocturnal or at rest	QT-segment depression
First- degree AV block (high prevalence)	Pathological Q waves
Mobitz type II (Wenckebach type) second-degree AV block (high prevalence)	Left atrial Enlargement
Increased P wave amplitude ajnd notching	LAFB or extreme left axis deviation
Incomplete RBBB	Right axis deviation/LPFB
Early repolarization pattern with notching or slurring of the terminal QRS complex.	

1. Modified from Corrado D. 12-lead ECG in the athlete: physiological versus pathological abnormalities. Br J Sports Med 2009;43:669-676

CLASSIFICATION OF ABNORMALITIES OF THE ATHLETE'S ECG

TRAINING-RELATED ECG CHANGES	UNCOMMON AND TRAINING-UNRELATED ECG CHANGES <5%
Early repolarization pattern with notching or slurring of the terminal QRS complex.	Right axis deviation/LPFB
Isolated QRS voltage criteria for left ventricular hypertrophy Positive Sokolow Lyon >35mm	Mobitz type II or third-degree AV block may be observed
Right ventricular hypertrophy criteria	Ventricular pre-excitation
Junctional escape complexes or Junctional escape rhythym	Complete RBBB/LBBB
	Long/short QT interval
	Brugada phenocopyes
	Epsilon waves
	f-QRS

Deep inverted T - wave from V_1 to V_3 in adults without incomplete or complete RBBB is not significant in elite athletes with exception with suspect of ARVD/C. In normal, young patients, there is usually positive T polarity in V_1; however, it may flatten and nearly always has a positive polarity in V_2. In absence of Complete RBBB in patients >12 years old, negative T wave from V_1 to V_3 is a sign with great value for diagnosis of ARVD/C. In symptomatic patients carriers of ARVD, the ECG generally shows T wave inversion in V_1 and V_2, which may reach up to V_6[1].



1. **Fontaine G, Tsezana R, Lazarus A, Lascault G, Tonet J, Frank R. Repolarization and intraventricular conduction disorders in arrhythmogenic right ventricular dysplasia Ann Cardiol Angeiol (Paris). 1994 Jan;43:5-10.**

Exertional pain in the chest and exertional syncope should prompt an extensive evaluation. Clearance for participation in sports should be curtailed until a complete evaluation has ruled out the presence of any of the following disorders[1]:

1. Cardiomyopathies: mainly HCM and ARVD/C
2. Channelopathies, without apparent heart disease
3. Congenital coronary arterial anomaly:

Anomalous origin of coronary arteries (AOCA)

- *Anomalous origin of the left coronary artery from the pulmonary artery Bland-White-Garland (BWG) syndrome*
- *Anomalous aortic origin of a coronary artery from the wrong aortic sinus of valsalva*
- *Atresia of the left main coronary artery and others miscelaneous*
- *Myocardial bridges*
- *Coronary arterial fistulas*.

The advent of state-of-the-art modalities of imaging seems, at times, to have supplanted the ECG in making the diagnosis of potentially serious coronary artery abnormalities, especially in asymptomatic mainly the three-dimensional coronary RN angiography[2] and ECG-gated multidetector computed tomography scanner, and reconstructed 3-dimentional images of the heart[3]. However, as is also the case for a detailed history and physical examination, the ECG provides a potentially insightful look at the coronary arteries. Furthermore, the past decade has witnessed an increase in the use of the ECG as a screening tool in the assessment of the risk of SCD in athletes in high school.

1. **Cohen M, Berger S. The electrocardiogram as an adjunct in diagnosing congenital coronary arterial anomalies.Cardiol Young. 2010 Dec;20 Suppl 3:59-67.**
2. **Clemente A, Del Borrello M, Greco P, Mannella P, Di Gregorio F, Romano S, et al. Anomalous origin of the coronary arteries in children: diagnostic role of three-dimensional coronary MR angiography. Clin Imaging. 2010 Sep-Oct;34:337-343.**

Congenital Coronary Arterial Anomaly

OTHER DENOMINATIONS:

ALCAPA syndrome: English acronym for Anomalous Left Coronary Artery Arising from the Pulmonary Artery The ALCAPA syndrome may be the result of:

1) Abnormal tronco-conal separation between the aorta and pulmonary arteries;

2) Persistance of pulmonary sinus of Valsalva along with aortic sinus of Valsalva, which spring from coronary arteries.

ALCAB: Abnormal Left Coronary Arterial Branching.

ALMCA: Acronym for Anomalous origins of the Left Main Coronary Artery.

Epidemiology

0.25-0.5% or 4 per 1,000 of all congenital heart diseases[1].

Congenital anomalies of coronary arteries occur in 0.2-1.2% of the general population and cause 12% of sudden cardiac deaths related to sports and 1.2% of deaths not related to sports[2].

The annual risk of SCD in athletes ranges between 5 and 10 per 1,000,000[3].

Of the carriers of the BWG syndrome, only 10% reach an adult age[4].

As most of the patients are asymptomatic, the diagnosis is usually made post-mortem, and as the risk of SCD is high, aggressive surgical treatment is indicated, in association to close follow up[5].

1. **Pfannschmidt J, Ruskowski H, de Vivie ER. Bland-White-Garland syndrome. Clinical aspects, diagnosis, therapy. Klin Padiatr. 1992 Sep-Oct;204:328-334.**
2. **von Kodolitsch Y, et al. Z Kardiol. 2005; 94:1-13.**
3. **Halawa B. Pol Merkuriusz Lek. 2004; 16:5-7.**
4. **Kreutzer U, et al. Z Kardiol 1998; 87:560-565.**
5. **Davis JA, et al. J Am Coll Cardiol. 2001; 37:593-597.**

The clinical diagnosis of ALCAPA is still a challenge for pediatricians and pediatric cardiologists[1].
The clinical and pathomorphological aspects may be grouped in 2 types[2]:
a) Infantile;
b) Adult.
The infantile type is observed in absence of enough collateral circulation, which explains the bad prognosis in natural history in patients without intervention and pattern of myocardial infarction in ECG. The entity should be suspected in infants in the face of unexplained cardiomegaly[3].
The ALCAPA syndrome may result in myocardial infarction, heart failure and possibly death during the early infantile period[4].
In this type, two subtypes stand out:
Severe symptoms with death before a year;
-Early disease followed by improvement.

Adult type: characterized by absence of early symptoms.
In this case, collateral circulation is present appropriately or almost, with normal or almost normal ECG.
In this context, coronary insufficiency usually manifests with strain, which explains sudden deaths in young athletes.

1. Birk E, et al .Isr Med Assoc J. 2000;2:111-114.
2. Pfannschmidt J, et al. Padiatr. 1992; 204:328-334.
3. el Habbal MM, et al. Br Heart J. 1988;60:90-92.
4. Fierens C, et al. Heart. 2000; 83:E2.

THE ECG

The ECG is the most helpful resource in clinical diagnosis, especially in symptomatic infants, and the VCG is useful not only for the diagnosis, but also for the follow up from the prognostic point of view. In children and in adutls, the ECG/VCG may be normal or almost normal[1], nevertheless, two elements characterize the ECG in this entity:

Rhythm: Sinus. There may be recurrent AF. A tendency to severe arrhythmias has been described, post-acute infarction and physical strain, which results in arrhythmic sudden cardiac death[2]. A study indicates the possibility of SCD by polymorphic VT secondary to reperfusion[3].

In pediatric patients, VT/VF was associated to a high rate of mortality (approximately 80%) in patients with recent or remote MI[4]. Sudden cardiac death may occur during the practice of exercises in teenagers.

HR: frequent sinus tachycardia in cases with heart failure.

P wave: normal in most cases. In the presence of left ventricular failure with increase in *LV end diastolic pressure* or mitral failure by ischemia or antero-lateral papillary muscle infarction, it may originate electrocardiographic pattern of LAE. In infants, sweating and dyspnea secondary to dilated cardiomyopathy are described, secondary to anomalous origin of coronary artery[5].

1. **Askenazi J, Nadas AS. Anomalous left coronary artery originating from the pulmonary artery. Report on 15 cases. Circulation. 1975 Jun;51:976-987.**
2. **Ho J, Jevon G, Sanatani S.. Anomalous origin of the left coronary artery with diffuse coronary hypoplasia resulting in sudden death. Can J Cardiol. 2005 May 1;21:529-531.**
3. **Saeed M, Gabara R, Strasberg B, Kusniec J, Rosanio S, Ware DL, Birnbaum Y. Reperfusion-related polymorphic ventricular tachycardia as a possible mechanism of sudden death in patients with anomalous coronary arteries. Am J Med Sci. 2005 Jun;329:327-329.**
4. **Johnsrude CL, Towbin JA, Cecchin F, Perry JC. Postinfarction ventricular arrhythmias in children. Am Heart J. 1995 Jun;129:1171-1177.**
5. **Roest AA, Filippini LH, Van Unnik-Treurniet RA, Blom NA. Tachypnoea and dyspnoea in two infants due to dilated cardiomyopathy associated with an anomalous origin of the left coronary artery. Ned Tijdschr Geneeskd. 2004 Dec 4;148:2451-2456.**

QRS axis : it may be normal; however there are cases –particularly in adults- with a tendency to extreme shift to the left. The cause of the shift is controversial. It has been proposed that it may be the consequence of selective hypertrophy of the LV postero-basal wall without irrigation involvement (irrigated by the right coronary artery). The anterior and lateral walls, irrigated by the LCA, are thin and possibly fibrotic.

Electrical position: usually horizontal.

LVH pattern: as a consequence of replication of myocytes, predisposed by chronic hypoxia. More observed in adults.

QRS duration: there usually is mild increase for the age, not reaching values compatible to block. The average in children younger than one year old, is 70 ms, and in older, 90 ms.

Deep S waves from V1 to V3: this sign should be due to RV postero-basal hypertrophy, which increases the negative component of QRS complexes located in the anterior opposite region (V1 to V3) (resulting in deeper S waves in antero-septal wall rS complexes: V1 to V3).

Amputation of R waves in V2 and V3 or pattern of myocardial infarction: present in the cases where collateral circulation from the RCA is insufficient (90% of the cases). More frequent in babies.

Pattern of myocardial infarction: present in the cases where collateral circulation from the RCA is insufficient (90% of the cases). More frequent in babies. Q waves of necrosis is present in 88.8% of the cases[1] frequently in the anterior, antero-septal, antero-apical or antero-lateral (apico-lateral) wall: V5, V6, I and aVL, mainly in infants. They are characteristically deeper, but not wide. Q wave with depth ≥ 3 mm and ≥ 30 ms with QR pattern in at least 1 of the following leads: I, aVL, and from V5 to V7, and absence of Q wave in inferior wall leads, which is considered typical and highly sensitive[2]. Pattern of myocardial infarction: present in the cases where collateral circulation, from the RCA, is insufficient (90% of the cases). More frequent in infants.

1. **Buziashvili Iul, Abdullaev FZ, Dvinianinova NB, Sopromadze DL. Electrocardiographic characteristics of an anomalous origin of the left coronary artery from the pulmonary trunk. Kardiologiia. 1988 Aug;28:59-63.**
2. **Johnsrude CL, Perry JC, Cecchin F, Smith EO, Fraley K, Friedman RA, et al. Differentiating anomalous left main coronary artery originating from the pulmonary artery in infants from myocarditis and dilated cardiomyopathy by electrocardiogram. Am J Cardiol. 1995 Jan 1;75:71-74.**

Ventricular repolarization: frequent ST segment elevation in the acute phase of infarction or later, possibly related to the formation of a ventricular aneurysm, accompanied by ischemic T waves. The appearance of an unexpected ST segment elevation during a surgical act of a different nature has been described. In the cases in which this atypical electrocardiographic manifestation occurs during elective surgery for other causes, the authors advise ruling out the possibility of the presence of anomalous origin of the coronary artery[1].

QT and QTc interval: QTc interval prolongation has been described, which returns to normal levels after surgical correction[2].

Ventricular arrhythmias: in athletes, potentially fatal ventricular arrhythmias generally occur as a consequence of structural heart disease, as HCM, ARVD, and the different forms of anomalous origin of coronary arteries[3].

Three pathophysiologic mechanisms have been proposed to explain the origin of ventricular arrhythmias:

1) Local ischemia caused by short circuit;

2) Reentry circuit that originates in the peripheral region of the area involved by necrosis;

3) Electrical instability secondary to endocardial fibrosis[4].

There is a description in literature of paroxysmal AV block complicated with syncope, which requires permanent pacemaker implantation[5].

1. **Alfirevic A, Mossad E, Niezgoda J. Unexpected ST segment changes in children--a case report. Paediatr Anaesth. 2005 Jan;15:63-67.**
2. **Glaser J, Rosenman D, Balkin J, Zion MM, Yakirevich V, Vidne B. Anomalous origin of the left coronary artery from the pulmonary artery: new electrocardiographic, echocardiographic and surgical observations. J Cardiovasc Surg (Torino). 1986 May-Jun;27:347-350.**
3. **Link MS, Homoud MK, Wang PJ, Estes NA 3rd. Cardiac arrhythmias in the athlete: the evolving role of electrophysiology. Curr Sports Med Rep. 2002 Apr;1:75-85.**
4. **Chang RR, Allada V. Electrocardiographic and echocardiographic features that distinguish anomalous origin of the left coronary artery from pulmonary artery from idiopathic dilated cardiomyopathy. Pediatr Cardiol. 2001 Jan-Feb;22:3-10.**
5. **Nishimura N, Bando S, Yamamoto H, Nishikado A, Akiyama K, Mori H. A case of anomalous origin of right coronary artery from the left sinus of Valsalva with ventricular aneurysm ventricular tachycardia and paroxysmal A-V block. Kokyu To Junkan. 1989 Jan;37:97-100.**

The pool of scientific evidence supporting the efficacy and cost-effectiveness of ECG screening for athletes is growing. However, feasibility and practical concerns regarding false-positive results, cost-effectiveness, physician infrastructure, and health care resources for large-scale implementation of ECG screening still exist[1]. The majority of SCD cases are caused by an underlying abnormality that potentially may be identified on cardiovascular screening, depending on the specific abnormality and the content of the cardiovascular screening applied. Indeed, today, cardiac screening is universally recommended by the European Society of Cardiology (ESC) and American Heart Association (AHA) and required by the Fédération Internationale de Football Association (FIFA) and Union of European Football Associations (UEFA). Pre-participation examination is by consensus understood to include personal history and physical examination; controversy exists regarding the usefulness and appropriateness of screening using resting 12-lead ECG. The ESC recommends screening includes personal history, physical examination, and 12-lead resting ECG, whereas recommendations from the AHA includes only personal history and physical examination. There is firm scientific ground to state that the sensitivity of screening with ECG is vastly superior to, and the cost-effectiveness significantly better than, screening without ECG. Cardiac screening of elite athletes with personal history, physical examination, and ECG is cost-effective also in comparison with other well-accepted procedures of modern health care. Newly published recommendations for the interpretation of the ECG in athletes and future studies on ECGs in athletes of different ethnicity, gender, and age may further increase the specificity of ECG in cardiac screening, refining the screening procedure and lowering the costs for additional follow-up testing. Cardiac screening without ECG is not cost-effective and may be only marginally better than no screening at all and at a considerable higher cost. The current evidence, suggests that the ECG should be mandatory in pre-participation screening of athletes[2].

1. **Drezner J, Corrado D. Is there evidence for recommending electrocardiogram as part of the pre-participation examination? Clin J Sport Med. 2011 Jan;21:18-24.**
2. **Borjesson M, Dellborg M.Is there evidence for mandating electrocardiogram as part of the pre-participation examination? Clin J Sport Med. 2011 Jan;21:13-17.**

THE CAUSES OF ARRHYTHMIC SUDDEN DEATH IN YOUNG ATHLETES (AVERAGE AGE: 17 YEARS)

I) ENTITIES WITH STRUCTURAL HEART DISEASE (98%)

1) Hypertrophic cardiomyopathy (HCM) whether in its obstructive form or in its non-obstructive form (36%);
2) Congenital anomalies of coronary arteries with increase of ventricular mass (19%);
3) Tumors or cardiac masses (10%);
4) Aorta rupture due to Marfan syndrome (5%). Mutation in the gene in fibrillin-1 (FBN1), in chromosome 15q21.1 and Marfan-like syndrome with no eye anomalies, mapped in chromosome 3p24;
5) Arrhythmogenic right ventricular dysplasia/cardiomyopathy (3%). Prevalence of 1 in 15,000;
6) Early atherosclerotic coronary artery disease (2%) by familial hypocholesterolemia and dominant mixed hyperlipidemia by alteration in chromosome 6;
7) Mitral valve prolapse syndrome (MVPS) (2%);
8) Myocarditis (2%)
9) Familial arrhythmogenic syndrome: ventricular and tachyarrhythmia association (syndrome of Wolff-Parkinson-White), progressive disease of conduction system and cardiac hypertrophy by involvement of regulatory subunit gamma-2 (PRKAG2) of AMP- activated by protein kinase[1];
10) Aortic stenosis.

1. **Gollob MH, et al. Curr Opin Cardiol. 2002;17:229-234**

THE CAUSES OF ARRHYTHMIC SUDDEN DEATH IN YOUNG ATHLETES (AVERAGE AGE: 17 YEARS)

II) ENTITIES WITHOUT STRUCTURAL HEART DISEASE (2%);

1) Drug abuse, e.g. anabolic agents,
2) Ventricular pre-excitation of the Wolff-Parkinson-White syndrome type, with anomalous pathway of short refractory period, not detected previously;
3) Cardiac concussion or commotio cordis;
4) Channelopathies or primary electrical diseases.

A) OF THE SARCOLEMMA OR EXTERNAL CHANNELOPATHIES:

1) Congenital long QT syndrome
2) Congenital short QT syndrome
2) Brugada syndrome;
3) hereditary progressive cardiac conduction defect Lenègre's
4) Idiopathic ventricular fibrillation (IVF) "Haıssaguerre syndorme";
5) Mixed forms or with overlapped phenotypic aspects:
   5a) BrS and LQT3;
   5b) BrS and Lenègre disease;
   5c) BrS and sinus node dysfunction;
   5d) BrS, LQTS and progressive conduction disorder;
6) Some sudden unexpected nocturnal death syndromes (SUNDS)= BrS;
7) Some sudden infant death syndromes (SIDS).

B) OF THE CHANNELS OF THE ENDOPLASMIC RETICULUM OR INTRACELLULAR CHANNELOPATHIES:

1) Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT).

DIFFERENCES BETWEEN PHYSIOLOGICAL VENTRICULAR HYPERTROPHY OF THE ATHLETE AND THE PATHOLOGICAL ONE (VENTRICULAR REMODELING)

	PHYSIOLOGICAL VENTRICULAR HYPERTROPHY	PATHOLOGICAL VENTRICULAR HYPERTROPHY VENTRICULAR REMODELING
Location:	Symmetrical, however, it may be asymmetrical.	Asymmetrical, however, it may be symmetrical.
Relative Ischemia:	Absent.	Present.
Myocitic/Non-myocitic Component Relationship:	Maintained.	Loss of balance in favor of the non-myocitic component (fibrosis).
Energetic Cycle:	Aerobiosis.	Anaerobiosis.
Renin-angiotensin-aldosterone Mechanism	Normal.	Increased.
Norepinephrine	Normal.	Increased.

DIFFERENCES BETWEEN PHYSIOLOGICAL LVH OF THE ATHLETE AND THE PATHOLOGICAL VENTRICULAR REMODELING

	PHYSIOLOGICAL LVH	PATHOLOGICAL LVH REMODELING
Atrial Natriuretic Peptide	**Normal.**	**It may be increased.**
Pump Function	**Normal.**	**Depressed.**
Heart Rate:	**Tendency to sinus bradychardia by vagotony.**	**Frequent tachycarrhythmia and sympathotony.**
LV Pd2:	**Normal.**	**Increased.**
Pulmonary Artery Pressure And Central Venous Pressure:	**Normal.**	**It may be increased.**
ANS:	**Parasympathetic predominance.**	**Sympathetic predominance.**
Curve Of Dissociation of Hb:	**Deviation to the right.**	**Deviation to the left.**
Echocardiogram:	**Proportional growth between the diameter and the thickness of walls. Normal LA.**	**Loss of walls thickness/diameter ratio. Increased LA.**

DIFFERENCES BETWEEN PHYSIOLOGICAL LVH OF ATHLETES AND HCM WHEN BOTH PRESENT WALL THICKNESS BETWEEN 13 mm & 15 mm

The concentric or symmetrical form of HCM (5%), may be confused with the athlete's heart with physiological hypertrophy of its walls, since the increase is not asymmetrical. For the differential diagnosis, the following criteria could be used:

	ATHLETE	**HCM**
Bizarre ECG pattern of LVE	No.	Yes.
LV cavity < 45 mm	No.	Yes.
LV cavity > 55 mm	Yes.	No.
LAE	No.	Yes.
Female Gender	Negative.	Positive.
Decrease Of Hypertrophy With Less Physical Training	Positive.	Negative.
Family History Or Provable Genetic Mutation	Negative.	Positive.

NON-INVASIVE SUPPLEMENTARY TESTS

1) **Electrocardiogram (ECG/EKG), QT dispersion, QRS duration (QRSD) or QRS complex duration, fragmented QRS (f-QRS), QT interval,**
2) **Vectorcardiogram (VCG)**
3) **Ergometer Test**
4) **Cardiopulmonary Metabolic Exercise Testing or Cardiopulmonary Exercise Metabolic Testing (CMET), Ergometer test with spirometery.**
5) **Echocardiogram M-module, two-dimensional with transthoracic Doppler.**
6) **Transesophageal or Biplanar Echocardiogram**
7) **Three-dimensional Echocardiogram (RT3DE) Real-Time Three-Dimensional (3D) Echocardiography,**
8) **Intracardiac Echocardiogram.**
9) **High resolution ECG, Signal-averaged ECG(SAECG)/Signal-averaged P wave analysis**
10) **Ambulatory electrocardiography monitoring, Ambulatory Electrocardiography Recorders (AECG) Holter monitoring.**
11) **Heart rate variability**
12) **Heart rate turbulence**
13) **Baroreflex sensitivity**
14) **Microvolt T wave alternans (TWA): Microvolt T-Wave Alternans**
15) **Body-Surface Potential Maps (BSPM)**
16) **Nuclear Magnetic Resonance, MRI: Magnetic Resonance Imaging, CMRI: Cardiac Magnetic Resonance Imaging, MDE-MRI: Delayed-Enhancement Magnetic Resonance Imaging (Gadodiamide MDE-MRI);**
17) **Ultra-Fast Computed Tomography (UFCT) or Electron-Beam Tomography;**
18) **Radioisotopic Ventriculography. Radionuclide Angiography or Radionuclide Ventriculography**

INVASIVE SUPPLEMENTARY TESTS

1. **Programmed Electrophysiological Stimulation (PES).**
2. **Cineangiography, coronary angiography, and ventricular angiography.**
3. **Endomyocardial biopsy(EMB) and RVEMB: Right Ventricle Endomyocardial Biopsy**