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Guidelines

Cardiovascular pre-participation screening in young athletes: Recommendations of the Association of European Paediatric Cardiology

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Abstract Sudden death in young competitive athletes can be avoided by implementation of pre-participation screening programmes. A screening programme should be performed only by trained physicians and should include the athlete's personal and family history, physical examination results, and the readings from a 12-lead-electrocardiogram. The athlete should undergo this screening programme every second year to detect progressive diseases. In addition, the programme should include detailed instructions to the athletes to pause training during infections in order to prevent sudden death due to myocarditis.

Keywords: Pre-participation screening; sudden cardiac death; electrocardiogram; prevention; young athletes

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Sudden death in young athletes is a rare but dramatic event with an incidence <1–3/100,000 athletes.[1–3] These deaths are mainly caused by cardiovascular diseases, with cardiomyopathies and coronary anomalies being the leading causes[2–4] (Fig 1). Corrado et al demonstrated that the early identification of cardiovascular diseases through a nationwide screening programme could prevent athletes from suffering sudden cardiac death. Screening could lead to a decrease of 89% in such deaths[6] (Fig 2). Screening programmes are recommended by several medical and sports associations, such as the European Society of Cardiology, the American Heart Association, the International Olympic Committee, and the American Society for Sports Medicine.[5,7–9] No international paediatric society, however, appears to have made any recommendations in this regard. Although screening programmes are universally recommended, the protocols are not uniform: the difference in most programmes being the inclusion or not of 12-lead electrocardiogram screening. The major concerns against electrocardiogram screening are the additional costs and the number of false-positive results.[5] The implementation of electrocardiogram guidelines by Corrado et al[10] and Drezner et al[11], and special training for the physicians involved, has led to a reduction in the number of false-positive results.[12] Underlining the importance of a standardised screening programme by trained physicians, Thünenkötter et al performed pre-participation examinations in all athletes of the FIFA World Cup 2006, and demonstrated that 1% of these athletes, who play in the best leagues all over the world, had findings indicating the presence of cardiovascular diseases.[13]

The Working Group for Preventive Cardiology of the Association of European Paediatric Cardiology decided to support the recommendation of the

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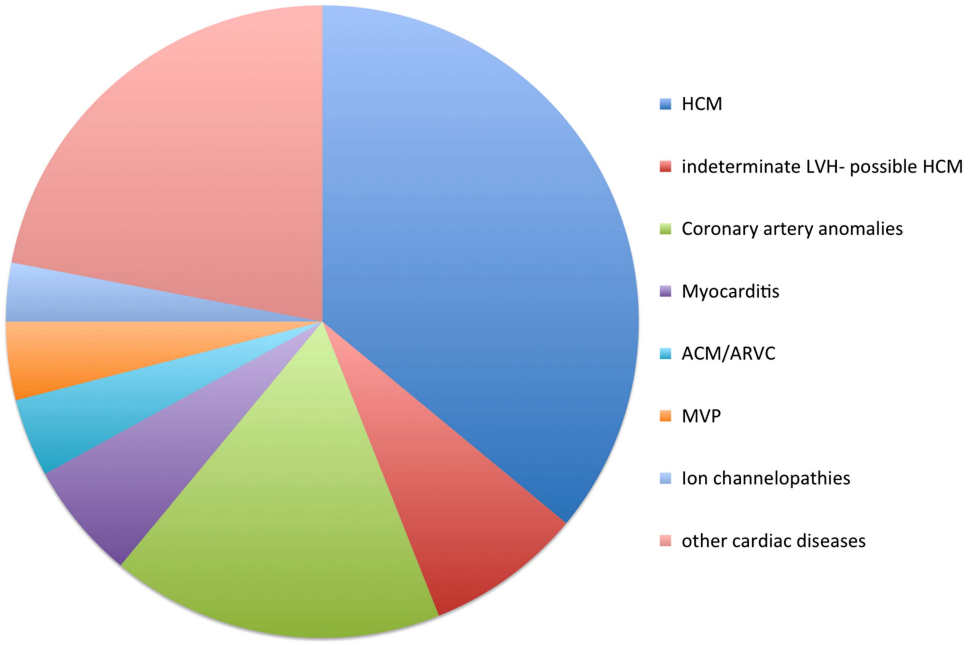

Figure 1.
Cardiovascular reasons for sudden cardiac death in athletes (modified Maron et al[5]). ACM/ARVC = arrhythmogenic right ventricular cardiomyopathy; HCM = hypertrophic cardiomyopathy; LVH = left ventricular hypertrophy; MVP = mitral valve prolapse.

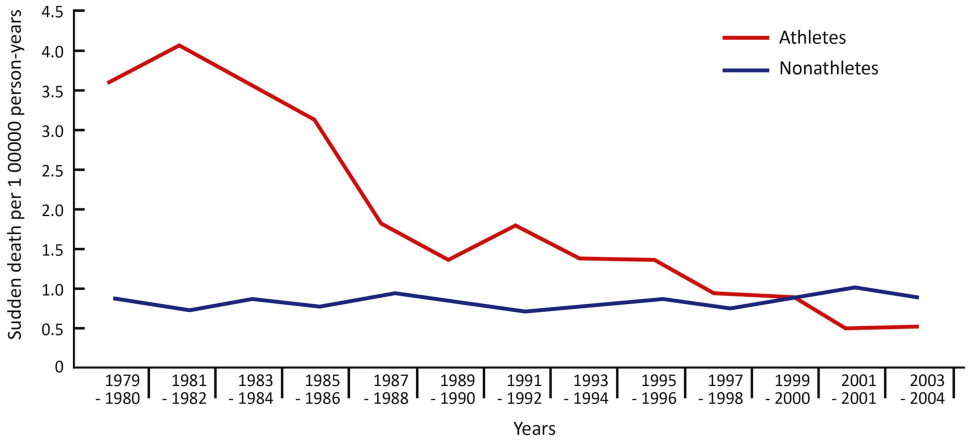

Figure 2.
Annual incidences of sudden cardiac death in Italy among screened and unscreened athletes (modified Corrado et al[6]).

European Society of Cardiology,[7] with special focus on the necessity for a 12-lead electrocardiogram to facilitate the timely detection of cardiovascular abnormalities in young athletes. This could result in proper diagnosis and therapy after detection, and reduce the risk for sudden cardiac death. The study, moreover, wishes to emphasise the importance of proper patient education to prevent sudden cardiac death due to myocarditis (Fig 3).

Causes of sudden cardiac death

Maron et al[3] investigated the sudden death of 1866 athletes in North America. Of this total, 56% of the deaths were due to cardiovascular diseases, 22% due to blunt trauma, 3% due to commotio cordis, and 10% due to miscellaneous causes. The leading cause of sudden cardiac death in this study was hypertrophic cardiomyopathy, as seen in 36% of the athletes, followed by coronary anomalies (17%), myocarditis (6%), and arrhythmogenic right-ventricular cardiomyopathy and mitral valve prolapse (4%) (Fig 1).

After the implementation of a nationwide screening programme in Italy, the leading cause of sudden cardiac death was arrhythmogenic right-ventricular cardiomyopathy, which was seen in 22% of those surveyed. Hypertrophic cardiomyopathy as a cause of sudden cardiac death was found in 2% of the athletes,

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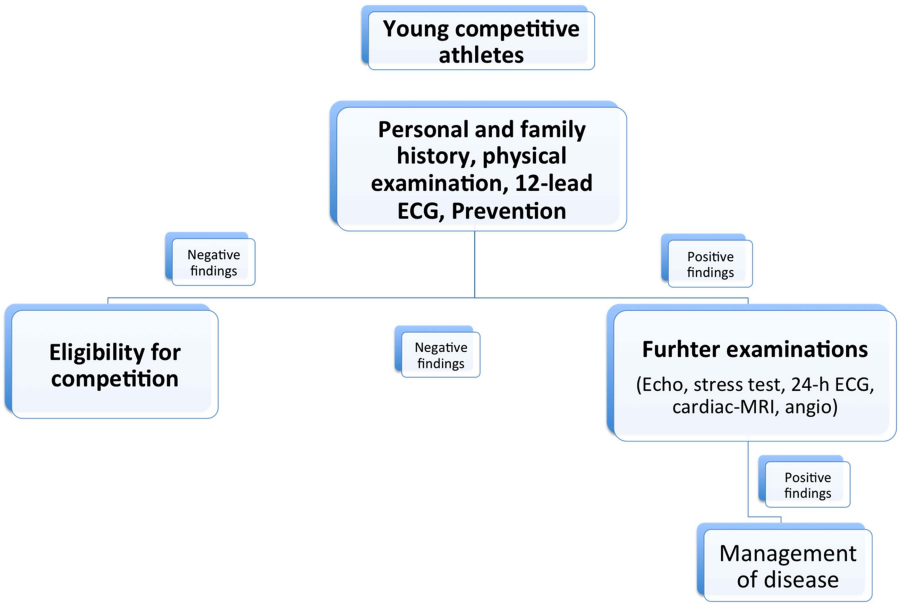

Figure 3.
New Association of European Paediatric Cardiology guidelines for pre-participation screening in young competitive athletes. ECG = electrocardiogram.

whereas the incidence of hypertrophic cardiomyopathy in non-athletes who were not screened was 9.5%. This discrepancy is explained by the unique exposure of Italian athletes to this systemic cardiovascular screening programme.[4,6] Table 1 shows the causes of sudden cardiac death after the implementation of the screening programme in screened athletes and unscreened non-athletes.

It is very important to note that some diseases can show a negative history and absent symptoms on physical examination in children. In particular, progressive diseases such as arrhythmogenic right-ventricular cardiomyopathy, usually starting during late adolescence, and heterogeneous diseases such as mitral-valve prolapse, which can appear in mild-to-severe forms, often have no symptoms in children, emphasising the importance of follow-up examinations.[14,15]

It is not the purpose of this paper to discuss the necessity of screening the entire population aged 12–25 years for cardiovascular disease, as this might not be as effective as screening in young competitive athletes alone.[16]

Recommendations for a cardiovascular screening programme

Nearly all cardiological and sports associations recommend that a screening programme should include a family and personal history and a physical examination, but they continue to challenge the need for a routine 12-lead electrocardiogram.[5,7,8] The screening should be performed on athletes when they start their competitive activity, and should be repeated routinely every 1–3 years.[7,9,14,15]

Table 1. Causes of sudden cardiac death in screened athletes and unscreened non-athletes in veneto region of Italy from 1979 to 1996 (modified Corrado et al[7]).

	Athletes (n = 49) (n (%))	Non-athletes (n = 220) (n (%))
ARVC/ACM	11 (22.4)	18 (8.2)
Atherosclerotic coronary artery disease	9 (18.5)	36 (16.4)
Anomalous origin of coronary artery	6 (12.2)	1 (0.4)
Conduction system pathology	4 (8.2)	20 (9)
MVP	5 (10.2)	21 (9.5)
HCM	1 (2)	16 (7.3)
Myocarditis	3 (6.1)	19 (8.6)

ACM/ARVC = arrhythmogenic right ventricular cardiomyopathy; HCM = hypertrophic cardiomyopathy; MVP = mitral valve prolapse

A great number of conditions causing sudden cardiac death are genetically determined, indicating the importance of *family history*. According to the guidelines of the European Society of Cardiology, athletes are considered to be at risk for sudden cardiac death when their close relatives had experienced a premature heart attack or a sudden cardiac death (<55 years of age in men, <65 years in women).[7,16] The presence of a disabling cardiovascular disease, such as cardiomyopathy, Marfan Syndrome, long QT syndrome, Brugada Syndrome, severe arrhythmia, or coronary artery disease, is also indicative of being at

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Table 2. Classification of ECG abnormalities in athletes (modified Drezner et al. Br J Sports Med 2017)

International consensus standards for ECG interpretation in athletes: definitions of ECG criteria

Abnormal ECG findings in athletes

These ECG findings are unrelated to regular training or expected physiological adaptation to exercise, may suggest the presence of pathological cardiovascular disease and require further diagnostic investigation.

ECG abnormality	definition
T wave inversion	≥ 1 mm in depth in two or more contiguous leads; excludes leads aVR, III and V1
► Anterior	► V2–V4 – excludes: black athletes with J-point elevation and convex ST segment elevation followed by TWI in V2–V4; athletes < age 16 with TWI in V1–V3; and biphasic T waves in only V3
► Lateral	► I and AVL, V5 and/or V6 (only one lead of TWI required in V5 or V6)
► Inferolateral	► II and aVF, V5–V6, I and AVL
► Inferior	► II and aVF
ST segment depression	≥ 0.5 mm in depth in two or more contiguous leads
Pathological Q waves	Q/R ratio ≥ 0.25 or ≥ 40 ms in duration in two or more leads (excluding III and aVR)
Complete left bundle branch block	QRS ≥ 120 ms, predominantly negative QRS complex in lead V1 (QS or rS) and upright notched or slurred R wave in leads I and V6
Profound non-specific intraventricular conduction delay	Any QRS duration ≥ 140 ms
Epsilon wave	Distinct low amplitude signal (small positive de ection or notch) between the end of the QRS complex and onset of the T wave in leads V1–V3
Ventricular pre-excitation	PR interval < 120 ms with a delta wave (slurred upstroke in the QRS complex) and wide QRS (≥ 120 ms)
Prolonged QT interval	QTc ≥ 470 ms (male) QTc ≥ 480 ms (female) QTc ≥ 500 ms (marked QT prolongation)
Brugada type 1 pattern	Coved pattern: initial ST elevation ≥ 2 mm (high take-off) with downsloping ST segment elevation followed by a negative symmetric T wave in ≥ 1 leads in V1–V3
Profound sinus bradycardia	< 30 beats per minute or sinus pauses ≥ 3 s
Profound 1° atrioventricular block	≥ 400 ms
Mobitz type II 2° atrioventricular block	Intermittently non-conducted P waves with a xed PR interval
3° atrioventricular block	Complete heart block
Atrial tachyarrhythmias	Supraventricular tachycardia, atrial brillation, atrial utter
Premature ventricular contractions	≥ 2 premature ventricular contractions per 10 s tracing
Ventricular arrhythmias	Couplets, triplets and non-sustained ventricular tachycardia

Borderline ECG findings in athletes

These ECG findings in isolation likely do not represent pathological cardiovascular disease in athletes, but the presence of two or more borderline ndings may warrant additional investigation until further data become available.

ECG abnormality	definition
Left axis deviation	−30° to −90°
Left atrial enlargement	Prolonged P wave duration of > 120 ms in leads I or II with negative portion of the P wave ≥ 1 mm in depth and ≥ 40 ms in duration in lead V1
Right axis deviation	> 120°
Right atrial enlargement	P wave ≥ 2.5 mm in II, III or aVF
Complete right bundle branch block	rSR' pattern in lead V1 and an S wave wider than R wave in lead V6 with QRS duration ≥ 120 ms

normal ECG findings in athletes

These training-related ECG alterations are physiological adaptations to regular exercise, considered normal variants in athletes and do not require further evaluation in asymptomatic athletes with no signi cant family history.

normal ECG finding	definition
Increased QRS voltage	Isolated QRS voltage criteria for left (SV1 + RV5 or RV6 > 3.5 mV) or right ventricular hypertrophy (RV1 + SV5 or SV6 > 1.1 mV)
Incomplete right bundle branch block	rSR' pattern in lead V1 and a qRS pattern in lead V6 with QRS duration < 120 ms
Early repolarisation	J point elevation, ST elevation, J waves or terminal QRS slurring in the inferior and/or lateral leads
Black athlete repolarisation variant	J-point elevation and convex ('domed') ST segment elevation followed by T wave inversion in leads V1–V4 in black athletes
Juvenile T wave pattern	T wave inversion V1–V3 in athletes less than age less than 16
Sinus bradycardia	≥ 30 bpm
Sinus arrhythmia	Heart rate variation with respiration: rate increases during inspiration and decreases during expiration
Ectopic atrial rhythm	P waves are a different morphology compared with the sinus P wave, such as negative P waves in the inferior leads ('low atrial rhythm')
Junctional escape rhythm	QRS rate is faster than the resting P wave or sinus rate and typically less than 100 beats/min with narrow QRS complex unless the baseline QRS is conducted with aberrancy
1° atrioventricular block	PR interval 200–400 ms
Mobitz type I (Wenckebach) 2° atrioventricular block	PR interval progressively lengthens until there is a non-conducted P wave with no QRS complex; the rst PR interval after the dropped beat is shorter than the last conducted PR interval

risk. *Personal history* is considered positive in case of a family history of exertional chest pain, syncope or near-syncope, irregular heartbeat or palpitations, unexplained shortness of breath, or fatigue during exercise.[5,7,16]

Physical examination is considered positive if a heart murmur – any diastolic and systolic grade >2/6, mid- or end-systolic click, second single or widely split or fixed heart sound – is detected. A diminished or delayed femoral pulse, an irregular heart rhythm, or

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elevated blood pressure readings over the 95th percentile also indicate positive risk.[5,7,16,17] Other positive findings may be musculoskeletal or ocular abnormalities suggestive of Marfan Syndrome.[7,16]

The American Heart Association still recommends a screening programme that includes only personal and family history and physical examinations.[5] In contrast, the European Society of Cardiology, the International Olympic Committee, several European countries, and several professional sporting leagues recommend cardiovascular pre-participation screening including a *12-lead-electrogradiogram*.[7,8] This procedure enhances the probability of detecting cardiovascular diseases in athletes.[10,12,18] The most common concern regarding the electrocardiogram is the high incidence of a false-positive reading, which can result in further cardiovascular evaluation or restriction from sport.[5] After the recommendation of interpretation criteria for a 12-lead electrocardiogram by Corrado et al, the proportion of abnormal electrocardiogram readings decreased from 16 to 9%.[10,19] These criteria have recently been revised by an international committee[20] (Table 2).

Electrocardiogram patterns in hypertrophic cardiomyopathy are shown in up to 95% of the patients studied.[21] The nationwide screening programme in Italy that included a 12-lead electrocardiogram resulted in a significant reduction of sudden cardiac death, showing a 77% higher sensitivity for detecting hypertrophic cardiomyopathy than history or physical examination.[6] Drezner et al showed an improvement in electrocardiogram interpretations by primary care and cardiology physicians from a baseline of 74/85%–91/96% after they used an electrocardiogram interpretation tool.[12] Asif et al summarised in their reviews that the cost-effectiveness of a screening programme that includes an electrocardiogram cannot be doubted[18,22] (Table 3).

Pre-participation examination should be performed by doctors trained in examining athletes and in reading their electrocardiogram.

An important part of a screening programme is the importance of reducing the incidence of sudden cardiac death due to myocarditis: the risk for sudden cardiac death due to myocarditis is about 6–12%.[3,6,23] It is important to note that the clinical presentation of myocarditis with an asymptomatic course up to sudden cardiac death complicates both diagnosis and prevention. Weber et al showed that, in myocarditis, the risk for sudden cardiac death does not correlate with the intensity of inflammation.[24] In addition, exercise in patients suffering from myocarditis enhances the viral disease, worsens cardiomyopathy, and aggravates the autoimmune process against the heart tissue.[25,26]

The best method of prevention would be a detailed education of young athletes during the pre-participation examination, explaining to them the need to pause exercise and competition during an infection to reduce

Table 3. Cost-effectiveness models for cardiovascular screening (modified Asif et al[18]).

Study	Type of study	FP rate	Prevalence of disease	Cost	Comments
Wheeler (2010)	Decision-analysis model	H, P, ECG = 5%	Overall risk of SCD 2,4/100,000 athletes per year	ICE of adding ECG to H and P is $ 42,000/ly saved	ECG alone is cost-effective. The addition of ECG to baseline practices saved 2.1 life-years per 1000 athletes screened and focussed H + P and ECG saved 2.6 life-years per 1000 athletes screened
Leslie (2012)	Simulation model screening for HCM, WPW, LQTS at age 14	FP rate 15%	2/1000	$91,000/ly	High FP rates and relatively low prevalence of disease. Alternative assumptions for HCM prevalence and mortality for WPW would reduce CE to <$50,000/ly gained
Schoenbaum (2012)	Decision-analysis model	FP rate of ECG 5%, H, P, and ECG 9%	1/1000	ICE of ECG alone $37,700/QALY; H, P, ECG $68,800/QALY	Low prevalence of disease used in study. If 1.45/1000 were used H, P, and ECG would have an ICE of <$50,000/ly gained

CE = cost-effectiveness; ECG = electrocardiogram; FP = false-positive; HCM = hypertrophic cardiomyopathy; ICE = incremental cost-effectiveness; LQTS = long QT-syndrome; LY = life-year; QALY = quality-adjusted life-year; SCD = sudden cardiac death; WPW = Wolf–Parkinson–White

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the risk for sudden cardiac death. The education should also include spreading awareness about the need to recognise red flag symptoms and the importance of new family history.

Conclusion

This manuscript posits that these recommendations are the first by an international paediatric society. The cardiovascular pre-participation screening of young athletes should include personal and family history, physical examination, and a 12-lead electrocardiogram. Trained physicians using the Electrocardiogram Criteria should perform the screening programme. The screening programme should include instructing athletes to suspend exercise during infections and recognise red flag signs. The screening should be performed before the start of competitive sports and should be repeated every second year to detect progressive diseases.

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Conflicts of Interest

None.

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