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Sports participation and inherited cardiac conditions:

Hypertrophic cardiomyopathy and balancing the risk

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Take Home Messages

- High intensity exercise is known to increase risk of cardiac arrest in some ICC, as a result exercise has been traditionally advised against.
- Studies show support for moderate intensity exercise in terms of mental health and cardiovascular disease risk.
- Advances in treatment hopefully means athletes participating with better outcomes and reduced risk in the future.
- Risk assessment and quality of life are vital and a balanced approach with shared decision making is essential and supported by the latest ESC guidance.

Introduction

The occurrence of a sudden cardiac arrest (SCA) in sport is a traumatic event which has gained attention over recent years with incidents involving several high profile athletes. A large post-mortem follow-up study demonstrated that cardiac disease represents 15% of deaths in college age athletes(1). Inherited cardiac conditions (ICC) are the more frequent cause in athletes of a younger age (under 35) suffering sudden cardiac death(2). Exercise has been treated with high levels of caution in relation to cardiac conditions and the European Society of Cardiology (ESC) has provided nuanced guidance in more recent years as research has progressed(3,4).

How do does HCM increase risk of SCA

Patients with ICC have an anomalous substrate that when combined with an environmental condition such as illness, medication or that of intense exercise can increase the risk of SCA(3,5,6). Possible causative mechanisms include via ventricular arrhythmia (5,6)or structural defect in hypertrophic cardiomyopathy (HCM) possibly via increase in left ventricular outflow tract obstruction (LVOTO)(5,7)

Identification and screening

Cardiac screening is used by organisations throughout the world to assess for underlying cardiac defects, which subsequently aids risk-stratification for participation in sport. The degree of

screening varies between sports and country. For example, Cardiac Risk in the Young provide cardiac screening in the United Kingdom via focused cardiac history and electrocardiogram as a minimum with echocardiography utilised in elite athletes(8). This process has to balance financial viability with pick-up rate and occurrence of false positive results(8).

Impact of underlying ICC on a patient and risk stratification

The latest ESC guidance sets clear advice on approaches to risk management and shared decision making regarding advice toward sports of different intensities(4). Regarding physiological impact, HCM in athletes has been shown to have result in a reduced VO_2 max on exercise testing(9). In contrast the RESET HCM trial showed that moderate exercise does improve VO_2 max in non-athletes without adverse effect(3,10). A study of 1660 patients supported this with a composite end point of syncope, ICD shock, cardiac arrest and death showing no difference between vigorous activity and a sedentary lifestyle(11). In this context vigorous activity was classed over one year, for a subject to have exercised for 60 hours or more to a level of 6 metabolic equivalents (METs)(11).

These studies suggest that continuation of exercise can reduce VO2 max in athletes however moderate-vigorous exercise is low risk and is of benefit to non-athletes in terms of cardiovascular fitness.

Table 1. Exercise recommendation and study findings in HCM

Evidence/Trial	Intensity of exercise	No. patients	Adverse events
ESC 2020 – class IIb recommendation(4)	High intensity exercise (for low risk individuals)	-	-
RESET-HCM(10)	Moderate	136	2% – symptomatic NSVT
Lampert et al(11)	Vigorous (6 METs >60 hours per year	1660	4.6% vs. 4.7% (vigorous – reached composite end-point
Wasserstrum et al(12)	Moderate (cardiac rehabilitation)	32	9% - stopped due to symptoms

(Abbreviations) HCM – Hypertrophic cardiomyopathy, NSVT – non-sustained ventricular tachycardia, composite end-point = syncope, ICD shock, cardiac arrest and death

Mental health and exercise

Exercise is recognised to improve cardiovascular risk and benefit mental health which is supported at moderate intensity(13). Studies have demonstrated the detrimental impact of HCM on patients' mental health notably with higher rates of depression and anxiety with reduced quality of life linked to HCM symptoms(14). Alongside the assumed benefit of improvement in psychological wellbeing, is the possibility of overtraining and concept of arrhythmia caused by exercise(3,13). Under-reporting of symptoms due to concern for being disallowed from participation must also be considered(9). Sports participation is of utmost importance to some and central to both their identity and income(15). The most recent ESC guidance demonstrates clearer personalisation of care and emphasis on the patient/doctor discussion around participation in sport(3,4). See Figure 1 for a depiction of an ICC patient journey.

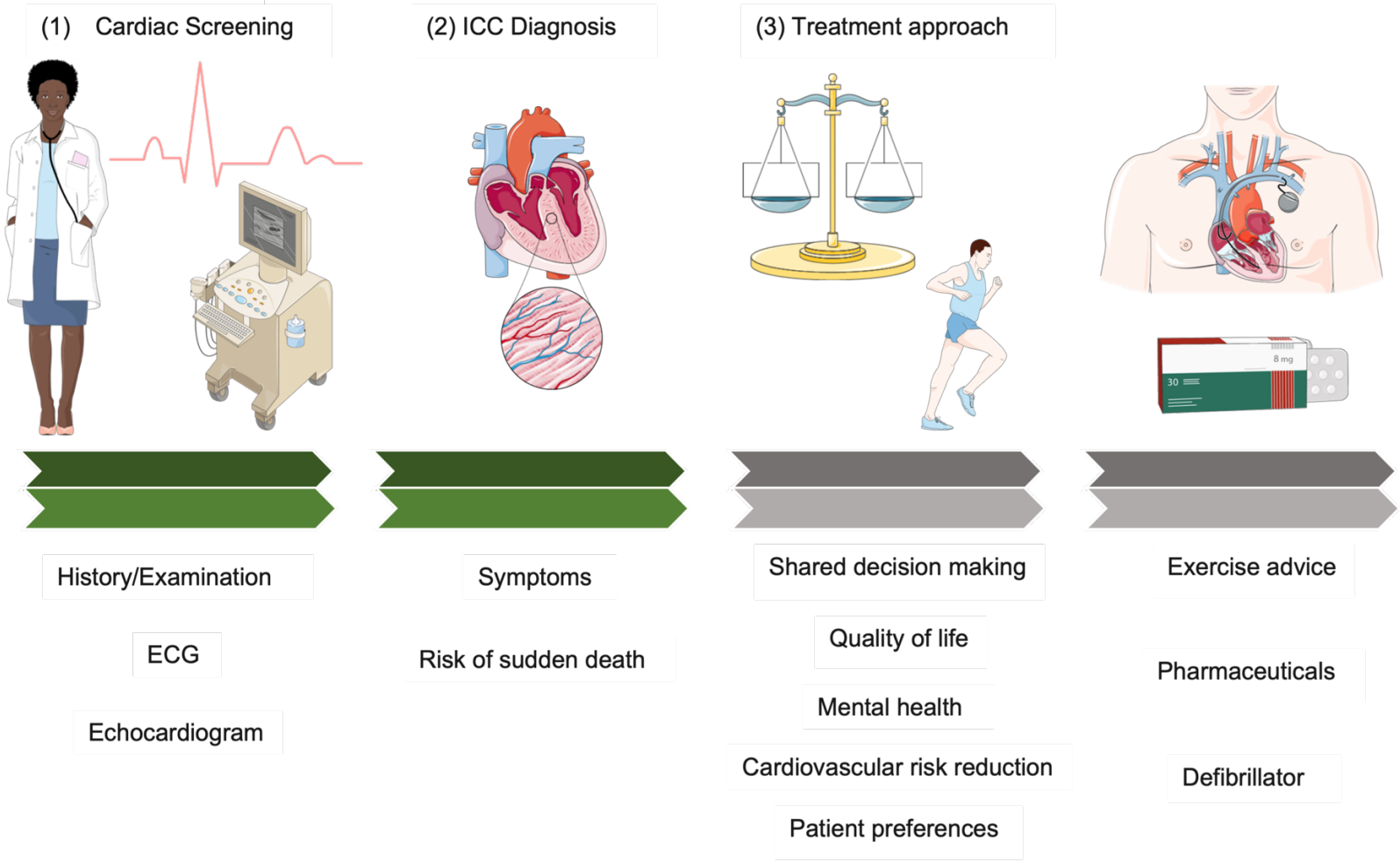

Figure 1. Process of ICC identification, diagnosis, consideration of risk, patient perspective and management plan. ICC, Inherited cardiac condition.

Current interventions in ICC

Pharmacological management includes beta blockers and disopyramide as well as atrial fibrillation and heart failure management as required(16). Myectomy can be used in severe cases or if refractory, cardiac transplantation is sometimes required(16). For athletes requiring an implantable cardiac defibrillator, a subcutaneous approach is viewed by some as the preferred option compared to a transvenous device due to the potential for pacemaker lead complications secondary to physical movement(17).

Looking to the future

Mavacamtem has recently been approved in the UK for use in symptomatic HCM(18) with the EXPLORER trial demonstrating an improvement in symptoms as well as VO_2 max and LVOT gradient(19). Could this improve the risk profile and lead to changes in exercise advice?

Conclusions

Exercise prescription in ICC is a changing field as we understand more about the outcomes of these patients. Screening plays an important part in helping to identify patients in order to prevent SCA. Once identified, open and frank discussions are essential to support a way forward for the patient and clinician where risk can be freely discussed to maximise both physical and mental health. Longer term follow-up and advancement of therapies may help to support more flexibility in sports participation advice.

Disclosures

No conflicts of interest

References

[1] Harmon KG, Asif IM, Maleszewski JJ, Owens DS, Prutkin JM, Salerno JC, *et al.* Incidence, cause, and comparative frequency of sudden cardiac death in national collegiate athletic association athletes a decade in review. *Circulation* 2015;132:10–19. https://doi.org/10.1161/CIRCULATIONAHA.115.015431.

[2] Wasfy MM, Hutter AM, Weiner RB. Sudden Cardiac Death in Athletes. *Methodist Debakey Cardiovasc J* 2016;12:76. https://doi.org/10.14797/mdcj-12-2-76.

[3] Semsarian C, Gray B, Haugaa KH, Lampert R, Sharma S, Kovacic JC. Athletic Activity for Patients With Hypertrophic Cardiomyopathy and Other Inherited Cardiovascular Diseases: JACC Focus Seminar 3/4. *J Am Coll Cardiol* 2022;80:1268–1283. https://doi.org/10.1016/j.jacc.2022.07.013.

[4] Pelliccia A, Sharma S, Gati S, Bäck M, Börjesson M, Caselli S, *et al.* 2020 ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease. *Eur Heart J* 2021;42:17–96. https://doi.org/10.1093/eurheartj/ehaa605.

[5] Basso C, Rizzo S, Carturan E, Pilichou K, Thiene G. Cardiac arrest at rest and during sport activity: Causes and prevention. *European Heart Journal, Supplement* 2021;22:E20–E24. https://doi.org/10.1093/EURHEARTJ/SUAA052.
[6] Hammond-Haley M, Patel RS, Providência R, Lambiase PD. Exercise restrictions for patients with inherited cardiac conditions: Current guidelines, challenges and limitations. *Int J Cardiol* 2016;209:234–241. https://doi.org/10.1016/j.ijcard.2016.02.023.
[7] Elliott P, Gimeno J, Tomé M, Shah J, Ward D, Thaman R, *et al.* Left ventricular outflow tract obstruction and sudden death risk in patients with hypertrophic cardiomyopathy. *Eur Heart J* 2006;27:1933–1941. https://doi.org/10.1093/eurheartj/ehl041.
[8] Sharma S, Merghani A, Gati S. Cardiac screening of young athletes prior to participation in sports: Difficulties in detecting the fatally flawed among the fabulously fit. *JAMA Intern Med* 2015;175:125–127. https://doi.org/10.1001/jamainternmed.2014.6023.
[9] Newman DB, Garmany R, Contreras AM, Bos JM, Johnson JN, Geske JB, *et al.* Cardiopulmonary Exercise Testing in Athletes With Hypertrophic Cardiomyopathy. *American Journal of Cardiology* 2023;189:49–55. https://doi.org/10.1016/j.amjcard.2022.11.008.
[10] Saberi S, Wheeler M, Bragg-Gresham J, Hornsby W, Agarwal PP, Attili A, *et al.* Effect of moderate-intensity exercise training on peak oxygen consumption in patients with hypertrophic cardiomyopathy a randomized clinical trial. *JAMA - Journal of the American Medical Association* 2017;317:1349–1357. https://doi.org/10.1001/jama.2017.2503.
[11] Lampert R, Ackerman MJ, Marino BS, Burg M, Ainsworth B, Salberg L, *et al.* Vigorous Exercise in Patients With Hypertrophic Cardiomyopathy. *JAMA Cardiol* 2023;8:595–605. https://doi.org/10.1001/jamacardio.2023.1042.
[12] Wasserstrum Y, Barbarova I, Lotan D, Kuperstein R, Shechter M, Freimark D, *et al.* Efficacy and safety of exercise rehabilitation in patients with hypertrophic cardiomyopathy. *J Cardiol* 2019;74:466–472. https://doi.org/10.1016/j.jjcc.2019.04.013.
[13] Aurélio M, Peluso M, Helena L, Guerra De Andrade S. *PHYSICAL ACTIVITY AND MENTAL HEALTH: THE ASSOCIATION BETWEEN EXERCISE AND MOOD*. Vol 60.; 2005.
[14] Cox S, O'Donoghue AC, McKenna WJ, Steptoe A. Health related quality of life and psychological wellbeing in patients with hypertrophic cardiomyopathy. *Heart* 1997;78:182–187. https://doi.org/10.1136/hrt.78.2.182.
[15] Drezner JA, Malhotra A, Prutkin JM, Papadakis M, Harmon KG, Asif IM, *et al.* Return to play with hypertrophic cardiomyopathy: Are we moving too fast? A critical review. *Br J Sports Med* 2021;55:1041–1047. https://doi.org/10.1136/bjsports-2020-102921.
[16] Arbelo E, Protonotarios A, Gimeno JR, Arbustini E, Barriales-Villa R, Basso C, *et al.* 2023 ESC Guidelines for the management of cardiomyopathies: Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC). *Eur Heart J* 2023;44:3503–3626. https://doi.org/10.1093/eurheartj/ehad194.
[17] Dahm JB, Hansen C. Subcutaneous Intracardiac Defibrillator (S-ICD): The Better ICD-Alternative in Athletes with Arrhythmogenic Risks? *Dtsch Z Sportmed* 2022;73:81–86. https://doi.org/10.5960/dzsm.2021.522.
[18] NICE (National Institute for Health and Care Excellence). Mavacamten for treating symptomatic obstructive hypertrophic cardiomyopathy. Technology appraisal guidance [TA913]. Available at https://www.nice.org.uk/guidance/ta913. Accessed January 16, 2024.
[19] Olivotto I, Oreziak A, Barriales-Villa R, Abraham TP, Masri A, Garcia-Pavia P, *et al.* Mavacamten for treatment of symptomatic obstructive hypertrophic cardiomyopathy (EXPLORER-HCM): a randomised, double-blind, placebo-controlled, phase 3 trial. *The Lancet* 2020;396:759–769. https://doi.org/10.1016/S0140-6736(20)31792-X.