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ON MY MIND

Return to Play With Genetic Heart Disease: The Importance of Developing a Personalized Emergency Action Plan

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Sudden cardiac death (SCD), while rare, is the most common cause of death for athletes during sports. Historically, athletes diagnosed with genetic heart diseases were subjected to blanket restrictions on sport participation because of theorized risks of exercise. More recently, observational evidence is emerging showing that when managed in expert settings with guideline based risk-stratification and management, athletes with many inherited cardiac conditions have low rates of breakthrough cardiac events.[1–3] The prospective multicenter observational study, LIVE-HCM (Lifestyle and Exercise in Hypertrophic Cardiomyopathy), demonstrated no increased risk of events in individuals with HCM who exercised at a vigorous intensity compared with nonvigorous exercisers, with event-rates of 15.9 and 15.3 per 1000 person-years, respectively.[4] Similar results were also seen in several smaller studies of athletes with HCM, where continued participation in competitive sports was not associated with excess of serious events. Long QT syndrome (LQTS) is another condition with data demonstrating low risk of exercise-induced arrhythmias in optimally treated patients. The prospective multicenter LIVE-LQT demonstrated low overall event rates in LQTS patients with contemporary management, and no difference in event-rate between vigorous and nonvigorous exercisers (8.3 vs 8.6 per 1000 person-years). Small studies of LQTS patients continuing to participate in sports show similarly low event-rates. Even individuals with defibrillator implants, who are by definition *high risk*, have not shown serious adverse events in competitive athletes or in those participating in high-risk sports, such as surfing.[1] For other conditions, such as arrhythmogenic cardiomyopathy (particularly *PKP-2*–mediated disease), there is a increased risk of heart failure, ventricular arrhythmias, and sudden death for individuals continuing to exercise, which highlights the importance of expert assessment and personalized risk stratification within a specialized multidisciplinary clinic.[1,3] Based on these data, recent professional society statements from European Society of Cardiology (2020), American Heart Association/American College of Cardiology (2024), and Heart Rhythm Society (2024) have evolved; now, participation in sports for many athletes with genetic heart disease "*may be considered*" (2b) or "*is reasonable*" (2a).[1,2]

Shared decision-making forms an increasingly important part of the expert consultation for athletes with genetic heart disease.[1–3,5] Shared decision-making involves a personalized discussion and evaluation of risk and risk tolerance, reviewing the best evidence available, and limitations of that data, understanding uncertainties and any controversies, and a discussion of values and preferences of the athlete.[1,5] Shared decision-making models of care empower athletes to make an informed decision about their care as part of a patient-centered care model. Shared decision-making incorporates all stakeholders in the athlete's care including the athlete, their family, their school or club, and often the sport's governing bodies.

As the focus moves away from a binary clearance model and toward facilitating return-to-play for athletes as part of a shared decision-making model, the importance of logistic and practical aspects to mitigate potential risk is emerging. The concept of emergency action planning for the population at large is well described. It is

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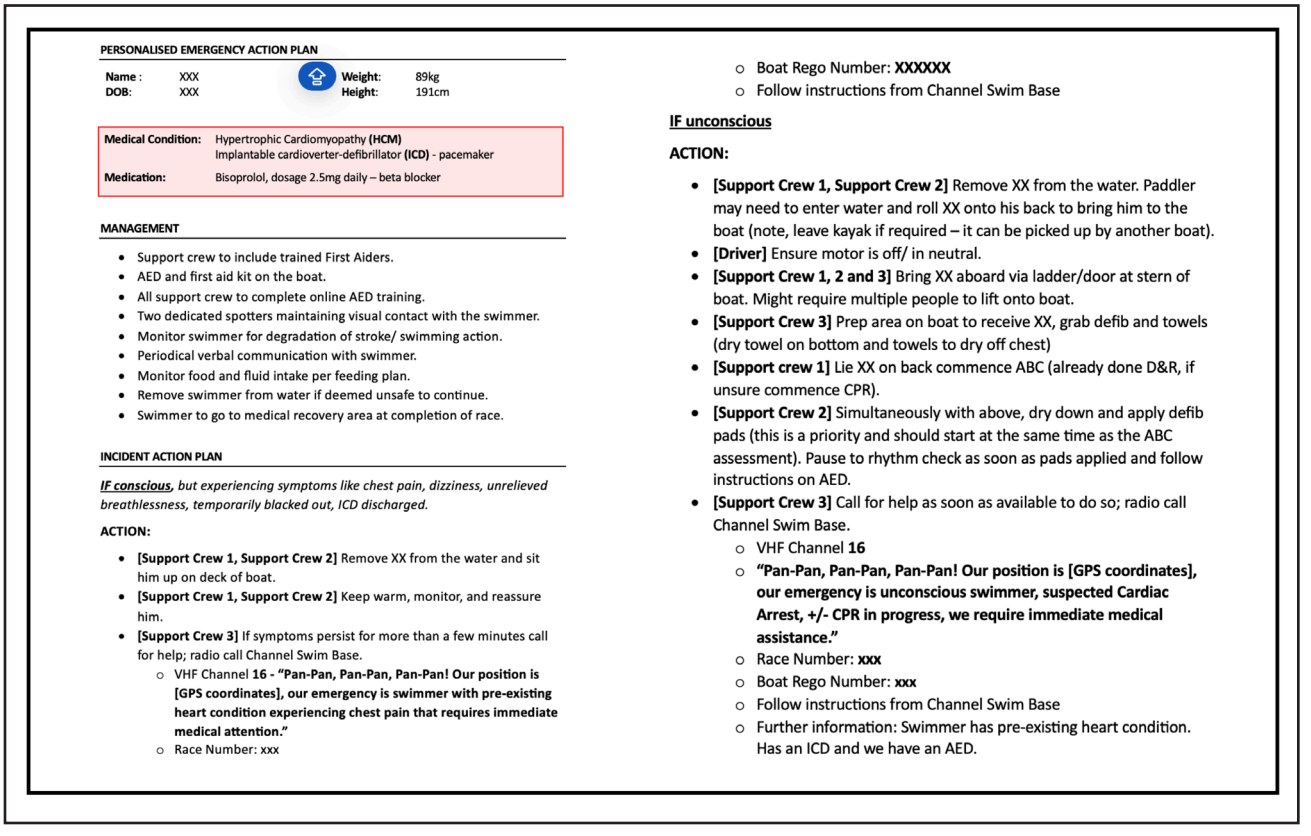
recommended that clubs, schools, and sporting organizations develop emergency action plans (EAP) in the event of sudden cardiac arrest (SCA) for all team-members, to include education on recognition of signs of SCA, CPR training, access to early defibrillation with automated external defibrillators on site, and clear plans for dispatch and transport to medical facilities for further management.

While less well recognized than population-focused EAPs, the concept of a personalized EAP (pEAP) is emerging for athletes who are working toward returning to play and have a known SCA risk—carrying genetic heart disease.[1] While exercise may not increase the risk of SCA for some CV disease, such as HCM or LQTS on optimal therapy, these individuals still carry a risk of SCA, which may occur during exercise.[4] The pEAP is of particular importance when the athlete may be placed in circumstances, such as collision sports or sports in or on water, where an arrhythmic event may place the athlete at higher risk than an event occurring at other times.

This document should be prepared jointly by the athlete and the cardiologist, with input from school/club where relevant. The pEAP should include both standard EAP practices, with the additional inclusion of personalized plans tailored to the specific sport and clinical situation for that athlete, with the goal of successful management of the worst-case scenario for the athlete. Situations which have not been carefully planned or thought out can lead to critical time delays and may be the difference between a successful or unsuccessful resuscitation. We propose that implementation of a pEAP is a key part of facilitating return to play for athletes with genetic heart disease who have elected to return to sports after a comprehensive shared decision making process. A comprehensive pEAP for an HCM patient planning an open water swim, for example, should outline critical elements that may be encountered by the athlete when returning to play (see Figure). First, precautionary measures should be outlined (eg, monitoring, training of support crew). Next, potential risks and adverse events based on the sport and clinical condition should be identified (eg, loss of consciousness in the water). Finally, response to these including the specific prescribed roles and action for all personnel required to act in case of an emergency should be outlined.

As data emerges showing that athletes can participate without excessive risk, as well as the recent, more supportive return-to-play guidelines for some genetic heart diseases, development of pathways to minimize risk



PERSONALISED EMERGENCY ACTION PLAN

Name : XXX **DOB:** XXX **Weight:** 89kg **Height:** 191cm

Medical Condition: Hypertrophic Cardiomyopathy **(HCM)**
Implantable cardioverter-defibrillator **(ICD)** - pacemaker

Medication: Bisoprolol, dosage 2.5mg daily – beta blocker

MANAGEMENT

- Support crew to include trained First Aiders.
- AED and first aid kit on the boat.
- All support crew to complete online AED training.
- Two dedicated spotters maintaining visual contact with the swimmer.
- Monitor swimmer for degradation of stroke/ swimming action.
- Periodical verbal communication with swimmer.
- Monitor food and fluid intake per feeding plan.
- Remove swimmer from water if deemed unsafe to continue.
- Swimmer to go to medical recovery area at completion of race.

INCIDENT ACTION PLAN

***IF conscious**, but experiencing symptoms like chest pain, dizziness, unrelieved breathlessness, temporarily blacked out, ICD discharged.*

ACTION:

- **[Support Crew 1, Support Crew 2]** Remove XX from the water and sit him up on deck of boat.
- **[Support Crew 1, Support Crew 2]** Keep warm, monitor, and reassure him.
- **[Support Crew 3]** If symptoms persist for more than a few minutes call for help; radio call Channel Swim Base.
  - VHF Channel **16 - "Pan-Pan, Pan-Pan, Pan-Pan! Our position is [GPS coordinates], our emergency is swimmer with pre-existing heart condition experiencing chest pain that requires immediate medical attention."**
  - Race Number: xxx
  - Boat Rego Number: **XXXXXX**
  - Follow instructions from Channel Swim Base

IF unconscious

ACTION:

- **[Support Crew 1, Support Crew 2]** Remove XX from the water. Paddler may need to enter water and roll XX onto his back to bring him to the boat (note, leave kayak if required – it can be picked up by another boat).
- **[Driver]** Ensure motor is off/ in neutral.
- **[Support Crew 1, 2 and 3]** Bring XX aboard via ladder/door at stern of boat. Might require multiple people to lift onto boat.
- **[Support Crew 3]** Prep area on boat to receive XX, grab defib and towels (dry towel on bottom and towels to dry off chest)
- **[Support crew 1]** Lie XX on back commence ABC (already done D&R, if unsure commence CPR).
- **[Support Crew 2]** Simultaneously with above, dry down and apply defib pads (this is a priority and should start at the same time as the ABC assessment). Pause to rhythm check as soon as pads applied and follow instructions on AED.
- **[Support Crew 3]** Call for help as soon as available to do so; radio call Channel Swim Base.
  - VHF Channel **16**
  - **"Pan-Pan, Pan-Pan, Pan-Pan! Our position is [GPS coordinates], our emergency is unconscious swimmer, suspected Cardiac Arrest, +/- CPR in progress, we require immediate medical assistance."**
  - Race Number: **xxx**
  - Boat Rego Number: **xxx**
  - Follow instructions from Channel Swim Base
  - Further information: Swimmer has pre-existing heart condition. Has an ICD and we have an AED.

Figure. Example pEAP for patient with HCM.
XX's replace identifiable information in this example. pEAP for a patient with HCM (age, 47 years) who has elected to participate in a ≈20-km open-water ocean swimming event after comprehensive assessment and discussion in shared decision-making model of care. AED indicates automated external defibrillator; CPR, cardiopulmonary resuscitation; HCM, hypertrophic cardiomyopathy; ICD, implanted cardioverter-defibrillator; pEAP, personalized emergency action plan; and VHF, very high frequency.

through careful personalized risk assessment and management plans, including the development of pEAPs, are critical.[1] For athletes who elect to continue sports despite risk, the process of developing a pEAP is a critical part of the shared-decision making model of care, and may take some time. Genetic heart diseases do carry a risk of SCA/SCD, but with appropriate risk-assessment, treatment, and pEAP, exercise may not increase the risk of SCA for some, beyond the risk of event during activities of daily living. By developing a pEAP together and ensuring all stakeholders, including athlete, family, and club/association, are well informed of potential risks and outcomes, the worst-case scenario is clearly highlighted. Relatedly, the pEAP also helps to emphasize the seriousness of the risks; indeed, some athletes may modify their decision to return to certain sports on development. The development of a pEAP is a critical part of the shared decision-making model of care, and takes time, with "worst-case scenario" situations clearly described and the patient and stakeholders, well-informed of all potential risks and outcomes.

ARTICLE INFORMATION

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