

# The Management of Athletes with Congenital Heart Disease



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## KEYWORDS

- Sudden cardiac death • Congenital heart disease • Congenital coronary anomalies
- Kawasaki disease • Sports cardiology

## KEY POINTS

- Sudden cardiac death is a devastating event and efforts should be made to its prevention by careful evaluation of athletes, especially pertaining to underlying cardiovascular conditions.
- Congenital heart defects affect about 1% of the population and it is of utmost importance the basic knowledge of common defects for the evaluation of athletes.
- Repaired congenital heart defects restoring normal physiology usually allow exercise activities and sports participation in most individuals.
- Unrepaired or palliated congenital heart defects usually pose at least some degree of limitations on the ability to participate in sports and athletics activities.
- Coronary anomalies, either congenital or acquired, are an important cause of sudden cardiac events during sports and exercise participation.

Gone are the days when we ran outside freely, climbed trees, rode bikes, and ran as long as humanly possible without having been prescreened by a health practitioner. The impact of sudden loss of basketball greats Reggie Lewis and Len Bias, volleyball Olympians such as Flo Hyman, countless world soccer players clipped of life mid-sprint, karate competitors heel-struck in the chest falling dramatically to their death is undeniable. Although rare, sudden cardiac death (SCD) is devastating to families and communities, and society dictates avoidance of any circumstances predisposing to the loss of human life during exertion.

SCD can only be “sudden” when there are no predictive factors, herein is the difficulty in prevention. Some of the more common risk factors for SCD have been described, although the true incidence and prevalence worldwide are difficult to ascertain given

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the complexity in defining the denominator (defined population in which SCD occurs). The most common final pathway leading to sudden cardiac arrest is ventricular tachycardia/fibrillation, which might be managed by the presence of an automated external defibrillator at all sports and exercise activities, along with trained experts in cardiopulmonary resuscitation. The exceptions are those with known connective tissue abnormalities (Marfan and Loews-Dietz syndromes, for example) leading to sudden aortic dissection.

Screening of athletes before sports participation by trained professionals is useful in identifying individuals who carry the known risk factors for SCD. Overrestriction may be almost as harmful as underrestriction of athletes, however. For example, patients with congenital heart disease (CHD) have been and continue to be overrestricted in their activities because there remains a paucity of data as to the safety of exercise in this population. There is a burgeoning population of children and adult survivors with CHD that may be inappropriately restricted by health care providers who have no or few data to advise them, parents who fear for their vulnerable children, or patients who self-restrict themselves for lack of appropriate counseling and not knowing their true limitations (or lack of).

Sports medicine specialists are tasked with providing individual exercise prescriptions for exercising individuals with and without CHD, and these are currently outlined in the 36th Bethesda guidelines.<sup>1</sup> There is precedent in the Special Olympics and para-Olympians. Sports medicine specialists should be able to identify those at overwhelming risk for adverse events surrounding vigorous activity and direct appropriate evaluation by the specialist (ie, cardiologist). Equally importantly, they should be able to coach individuals to improve performance and quality of life with exercise.

## EXERCISE AS MEDICINE

How does exercise figure into the mental and emotional well-being, quality of life, societal productivity, and general well-being of people with CHD? Can exercise attenuate the natural history of heart failure described in adolescents and adults after surgery for CHD<sup>2</sup>? Competitive and recreational sports participation may improve the quality of life and exercise capacity as well as lower body mass index in patients with mild, moderate, or severe CHD.<sup>3</sup> In one population of patients with a variety of complex congenital heart lesions, 50% participated in competitive sports, 25% were recreational athletes, and 29% of the severely affected congenital heart patients participated in competitive sports despite recommendations against such activities by the 36th Bethesda guidelines,<sup>4</sup> indicating the perceived benefits of exercise far outweighed the reported risks in the population studied. The job of sports medicine professionals is to help patients balance the risk:benefit ratio. The neurocognitive benefits of exercise are myriad and not to be underestimated in a population that is at risk for neurocognitive delays due to the effects of their disease, underlying genetics, or complexity of necessary interventions.

The positive effects of exercise on the myocardium, including enhanced myocardial contractility, improved diastolic filling due to improved myocardial relaxation, better compliance, decreased filling pressure, and improved atrial compliance, all contributing to increases in stroke volume and cardiac output, have been well documented,<sup>5-7</sup> although less well documented in the myocardium of patients with CHD that has been manipulated by surgical or catheter interventions or affected by residual lesions.

Despite the latest observation that there is an increase in coronary calcium scoring in elite runners, the cardioprotective effects of exercise are well demonstrated. Exercise acts directly on coronary blood flow by increasing myocardial oxygen demand,

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