Sudden cardiac death during exercise in patients with congenital heart disease: the exercise paradox and the challenge of appropriate counselling

Gerhard-Paul Diller* and Helmut Baumgartner

Division of Adult Congenital and Valvular Heart Disease, Department of Cardiovascular Medicine University Hospital of Münster, Münster, Germany.

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This editorial refers to ‘Sudden unexpected death in children with congenital heart defects’ by J. Jortveit et al., on page 621.

The population of patients with congenital heart disease (CHD) is growing rapidly. It is well appreciated by congenital cardiologists that the majority of CHD patients are not cured, and sequelae as well as late complications are prevalent.¹ One of the major causes of mortality in this population is sudden, mostly arrhythmic, death.² While not being the leading cause of mortality in the majority of CHD subgroups, sudden cardiac death (SCD) remains a particularly emotive subject inducing profound reactions from medical professionals, families, and society. This is due the fact that death occurs unexpectedly, often in patients who are otherwise well and lead a relatively normal life. Risk stratification has, therefore, been a major focus of CHD research at many institutions. Despite decades of effort, identifying CHD patients at particularly high risk of SCD and those who would most likely benefit from the implantation of defibrillator-cardioverters (ICDs) remains challenging. While different risk factors and risk scores have been developed, their value for identifying patients at risk is still limited.³–⁵ On the other hand, implantation of ICDs in young CHD patients is associated with high morbidity and risk of inadequate shocks, ultimately also impacting on the quality of life in this population.⁶

Sudden death can occur both at rest and during exercise. However, it has been well appreciated that exercise is associated with an increased risk of SCD during sporting activity.⁷,⁸ The current study by Jortveit et al.⁹ is an important contribution to the literature on SCD and exercise in CHD. It is particularly relevant as it focuses on children with the condition. Due to the continuing decline in CHD-related mortality (as a consequence of improved interventional and surgical techniques), more and more CHD patients reach older age. As a consequence, increasing numbers of patients will succumb to acquired cardiovascular disease and stroke.¹⁰ Arteriosclerotic disease manifests itself mainly in middle-aged or elderly men and post-menopausal women. However, its roots lie decades earlier, before the onset of overt symptoms or complications of the disease. In fact, atherosclerosis often develops from childhood. Childhood adiposity represents an increasing problem in most western societies, and is a risk factor for later complications.¹¹,¹² Early interventions to reduce cardiovascular risk factors are thus required, and regular exercise is accepted as a key intervention to promote cardiovascular health. Unfortunately, CHD patients are often advised against physical exercise and generally overprotected. Not surprisingly, a recent study found that these patients had a lower level of physical activity compared with healthy peers.¹³ One reason for advising CHD patients against physical activity might be the uncertainty regarding the individual risk of SCD. While previous studies in adults with CHD have reported this risk to be relatively low, it was still relevant. Koyak and co-workers investigated the risk of SCD in a multinational registry of 25 790 adult patients with CHD. They reported on 1189 deaths using a case–control study design.¹⁴ While 19% of the overall deaths were sudden, only 10% of those occurred in association with exercise. Therefore, the overall proportion of sudden deaths during exercise was ~1.5%. These relatively low rates are also supported by another Dutch study, reporting that only ~8% of SCDs were exercise related.¹⁵ Although reassuring, the SCD rate is still substantial in this adult population compared with the general population.

Based on a large and unique national Norwegian registry, including all ~940 000 live births in the country between 1994 and 2009,
linked to a CHD register and the Norwegian Cause of Death Registry, Jortveit et al. identified children experiencing sudden unexpected death unrelated to cardiac surgery. Overall, only 19 children (0.2%) died suddenly and unexpectedly, and only a subset of these due to cardiac reasons. Reassuringly, none of the children died during physical activity. However, two children were fortunate enough to survive sports-related cardiac arrest. Limitations of the study include the limited depth of clinical information inherent to such nationwide registers based largely on administrative data and the fact that the level of physical exercise cannot be quantified. When directly comparing the results of the study of Jortveit et al. with those of previous adult studies one should consider that, beyond the obvious age difference, surgical and interventional techniques have evolved over time. Therefore, unlike adult CHD patients under follow-up at grown-up congenital centres, contemporary children with CHD may have benefited from the wide availability of timely, state-of-the-art repair, which may in turn impact on the rate of complications, including the propensity for SCD.

Despite the transiently increased risk of SCD associated with exercise, numerous studies have established unequivocally that regular exercise is related to improved long-term prognosis and a reduced risk of cardiovascular and non-cardiac death. The magnitude of risk reduction is substantial and—in general—comparable with that expected from medical statin therapy. Even moderate regular physical activity (of 30 min duration, five times a week) leads to a reduction of 30–40% in cardiovascular risk, while regular high levels of exercise are associated with a 70% reduction in age-adjusted all-cause mortality in men and with a 80% reduction in women. This dose-related beneficial effect of exercise is supported by other studies linking weekly running time to decreasing death rates related to cardiovascular disease, but also cancer and stroke. Exercise prescription is not straightforward, however, due to the heterogeneity of CHD and the fact that current risk scores are far from perfect. As a consequence, understanding of CHD is required when informing patients and parents about risks and benefits of physical activity. Beyond clinical experience, a structured approach to the patient is recommended taking into account recognized risk factors for arrhythmias, complications, and death in this population. Both patient-specific risk and the nature and intensity of the intended sport should be considered. The Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology has recently presented a position paper and expert opinion on this topic, which may aid clinicians in clearing patients for exercise. As illustrated in Figure 1, it calls for the assessment of ventricular function, and exclusion of aortic dilatation, pulmonary hypertension, and cyanosis before allowing strenuous exercise. In addition, a history of (especially exercise-related) arrhythmias should be excluded. Based on these factors, the optimal level of exercise intensity and the safety of isometric exercise should be estimated. If in doubt, supervised exercise testing should be performed to assess objectively physical capacity and exclude malignant exercise-induced arrhythmias. The relative static and dynamic components of various athletic activities have been delineated in a study by Mitchell et al.

Taking together the available evidence, it should be highlighted that exercise training is beneficial for healthy individuals as well as most patients with acquired cardiovascular conditions. While published experience in CHD patients is more limited, it is likely that CHD patients also benefit from regular exercise. The available

Figure 1 Trade-off between transiently increased risk of sudden cardiac death (SCD) associated with exercise and the long-term mortality benefits of exercise in patients with congenital heart disease (CHD). CV, cardiovascular. Modified and based on Chugh and Weiss and Budts et al.
data and experience from cardiopulmonary exercise testing in a large number (thousands) of adult CHD patients also suggests a low risk of complications and exercise-induced SCD in this population. Jortveit et al. This notion is supported by the study of Jortveit et al., in this issue of the journal, suggesting a near 0% risk in children with CHD. As a consequence, children and adults with CHD should not be categorically discouraged from regular physical activity or from taking part in non-competitive sports. However, patients still stand to benefit from a tailored approach, with individual exercise prescription based on clinical experience, exclusion of high risk features, and a supervised exercise test in selected cases. In addition, the nature and intensity of exercise should be adapted to the individual patient. This approach not only should reduce the risk of SCD but also reassures the patient, parents, and the instructors supervising sporting activities.

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**References**


