



Editorial

Sarcopenia: An unrecognized, but important factor for adults with congenital heart disease

Susanne J. Maurer, Oktay Tutarel*

Department of Congenital Heart Disease and Paediatric Cardiology, German Heart Centre Munich, Technical University of Munich, Munich, Germany

ARTICLE INFO

Article history:

Received 28 July 2019

Accepted 12 August 2019

Available online 14 August 2019

Sarcopenia was initially defined as a loss of skeletal muscle of the limbs two standard deviations below the mean of a healthy young reference group [1]. Current evidence suggests, that it is not limited to this finding, but is also associated with a loss of muscle strength [2]. This has led to a more detailed concept that includes muscle strength (grip strength) or function (walking speed or distance) [2]. Based on this, a number of revised definitions of sarcopenia have been proposed which include both factors, a loss of muscle mass as well as a loss of muscle function [1–3]. Sarcopenia is not rare, and its prevalence is increasing with age, affecting up to 50% of octogenarians [1,2]. Furthermore, it is not only a common finding in elderly people, but also increasingly encountered in patients with chronic illnesses, for example chronic heart failure. Possible explanations include an abnormal energy metabolism coupled with mitochondrial dysfunction, as well as a change of myofibers [1]. Moreover, malnutrition, systemic inflammation, and oxidative stress are also encountered, which pave the way for sarcopenia [4]. This is even true in younger patients. In a recent study in 55 patients with non-ischemic dilated cardiomyopathy and a mean age of 37 years, 47% met criteria for muscle wasting [5]. Furthermore, muscle wasting was associated with lower left ventricular ejection fraction, lower 6-min walk distance, higher New York Heart Association functional class, and higher hospitalization rate [5].

In a substantial proportion of patients with adult congenital heart disease (ACHD), neurohormonal activation is present, that bears the hallmarks of chronic heart failure, relating to symptom severity and ventricular dysfunction [6]. Additionally, respiratory

and skeletal muscle weakness is also common in young adults with complex congenital heart defects (CHD) and resembles that found in older adults with advanced heart failure from acquired heart disease [7]. Therefore, it is reasonable to assume that sarcopenia is also frequent in the ACHD population. In the current issue, Sandberg and colleagues report their findings in 73 ACHD patients with more complex disease from three centers in Sweden [8]. The cohort included mainly patients with a Fontan circulation, Tetralogy of Fallot, and after an atrial switch for transposition of the great arteries. While in one center all patients were eligible, in the remaining two centers only convenience samples were included. They were compared with age- and sex-matched controls. A comparison with a population based healthy reference cohort might have added strength to the study. Furthermore, the definition used for sarcopenia was unfortunately based only on the body composition without including measures of muscle function. Nonetheless, according to the used definition 47% of male and 59% of female ACHD patients fulfilled the criteria of sarcopenia, compared to 10% and 9%, respectively, in the control group. These numbers are higher than those reported in a recent Japanese study, where 14% of male, and 18% of female ACHD patients were found to be sarcopenic [4]. The composition of cohorts in both studies differed, which might explain the differences. Still the higher number of female patients affected is noteworthy. Interestingly, in Sandberg and colleagues study, sarcopenia was not associated with systemic ventricular function (SVF), while the majority of the patients had none or only mild impairment of SVF [8]. The same multiple causative factors, which are encountered in patients with acquired cardiovascular disease, are probably also responsible for this high prevalence of sarcopenia in ACHD patients. In addition, there is the well-recognized lifelong detraining that many patients with CHD have since childhood as a result of reduced levels of physical activity [9].

The important question is, if sarcopenia is associated with a worse outcome in the ACHD population. Considering for example Fontan patients for whom skeletal and inspiratory muscle function plays an important role for venous return, such an association seems reasonable, but still needs proof. Furthermore, it is unclear if sarcopenia is a symptom indicating those patients that are more severely affected or a direct cause for the worse outcome. In other words, what comes first: sarcopenia or deterioration of the CHD? Further research is necessary to delineate this relationship. Finally, if sarcopenia is associated with a worse outcome, which intervention would be able to reverse it and would this lead to an improved

* Corresponding author at: Department of Congenital Heart Disease and Paediatric Cardiology, German Heart Centre Munich, Technical University of Munich, Lazarettstr. 36, 80636 Munich, Germany.

E-mail address: oktay.tutarel@tum.de (O. Tutarel).

outcome? Exercise training, nutritive supplements as well as various pharmaceutical interventions have been suggested as treatment options for sarcopenia in patients with acquired cardiovascular disease [1]. Their long-term effect is unclear. Data for ACHD patients is scarce. Recently, Shiina and colleagues reported some positive effects of resistance training in a small pilot study [4]. Further research is required.

Sandberg and colleagues study raises important questions and is a valuable addition to our knowledge. It emphasizes that we need to focus more on the extracardiac effects and manifestations that CHD may cause, and not only on the treatment of the CHD itself.

Declaration of competing interest

The authors report no relationships that could be construed as a conflict of interest.

References

[1] J. Springer, J.I. Springer, S.D. Anker, Muscle wasting and sarcopenia in heart

- failure and beyond: update 2017, *ESC Heart Fail.* 4 (2017) 492–498.
- [2] J.E. Morley, S.D. Anker, S. von Haehling, Prevalence, incidence, and clinical impact of sarcopenia: facts, numbers, and epidemiology-update 2014, *J. Cachexia. Sarcopenia Muscle* 5 (2014) 253–259.
- [3] Y. Kinugasa, K. Yamamoto, The challenge of frailty and sarcopenia in heart failure with preserved ejection fraction, *Heart* 103 (2017) 184–189.
- [4] Y. Shiina, N. Matsumoto, D. Okamura, Y. Takahashi, Y. Kijima, T. Fukuda, et al., Sarcopenia in adults with congenital heart disease: nutritional status, dietary intake, and resistance training, *J. Cardiol.* 74 (2019) 84–89.
- [5] M. Hajahmadi, S. Shemshadi, E. Khalilipour, A. Amin, S. Taghavi, M. Maleki, et al., Muscle wasting in young patients with dilated cardiomyopathy, *J. Cachexia. Sarcopenia Muscle* 8 (2017) 542–548.
- [6] A.P. Bolger, R. Sharma, W. Li, M. Leenarts, P.R. Kalra, M. Kemp, et al., Neurohormonal activation and the chronic heart failure syndrome in adults with congenital heart disease, *Circulation.* 106 (2002) 92–99.
- [7] M. Greutmann, T.L. Le, D. Tobler, P. Biaggi, E.N. Oechslin, C.K. Silversides, et al., Generalised muscle weakness in young adults with congenital heart disease, *Heart.* 97 (2011) 1164–1168.
- [8] C. Sandberg, K. Johansson, C. Christersson, J. Hlebowicz, U. Thilen, B. Johansson, Sarcopenia is common in adults with complex congenital heart disease, *Int. J. Cardiol.* (2019), <https://doi.org/10.1016/j.ijcard.2019.06.011>.
- [9] A. Giardini, Generalised myopathy in young adults with congenital heart disease, *Heart* 97 (2011) 1115–1116.