International Journal of Cardiology 278 (2019) 91–92

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Editorial

Advanced care planning in adult congenital heart disease: A life-long journey

Margarita Brida *

Adult Congenital Heart Centre and National Centre for Pulmonary Hypertension, Royal Brompton Hospital, London, UK
National Heart and Lung Institute, Imperial College, London, UK
Division for Adult Congenital Heart Disease, Department of Cardiovascular Medicine, University Hospital Centre Zagreb, Croatia

ARTICLE INFO

Article history:
Received 22 November 2018
Accepted 30 November 2018
Available online 4 December 2018

Adults born with congenital heart disease (CHD) represent a remarkable success story in the field of cardiology and cardiac surgery. In the 1950s, survival of children born with CHD was only around 15%. Due to the advances in pediatric cardiology, innovations in surgical and interventional procedures, more than 90% of children nowadays born with CHD are expected to reach adulthood [1]. However, behind this success story many challenges remain. Although the majority of these patients had operation(s) in childhood they are rarely cured, but rather palliated and therefore need to live with a chronic disease. Along with the ongoing morbidities such as heart failure and arrhythmias, reduced exercise capacity, and the need for further electrophysiological, interventional, or surgical procedures, mortality is also increased in this population of patients [2]. This is especially true for patients with more complex CHD that require life-long medical care in order to have a satisfactory life quality. Indeed, CHD is a challenge of a lifetime and proper education of patients regarding their underlying cardiac condition and life-long health behavior is vital in order for them to share responsibility for their life. The education should begin in adolescence with structured transition programmes [3].

Troost et al. provided an excellent overview as well as their own tertiary centre expertise on the importance of such life-long approach with advanced care planning in adult CHD. The authors' emphasis is on two main life periods: transition from pediatric care to adulthood and palliation with end of life care [4].

A well-established transition programme in the expert CHD centre is mandatory. It ensures that the patient in the vulnerable adolescent age is not lost to follow-up and continues to receive optimal and timely medical care, as well as learning about his underlying cardiac condition. As the authors have explicitly stressed, during this transition phase it is important, depending on the patient maturity, for him to obtain an understanding of his heart condition and potential future complications. If the patient remains stable with an uneventful course during regular clinical follow-ups, pointing out once more potential risks is unnecessary and potentially harmful, as it may lead to psychological issues [4]. The fine balance between disease information and its emotional impact always needs to be considered. The long-lasting relationship formed between the adult CHD physician and the patient represents solid ground for such communication.

The second important life period, especially in the subgroup of complex and very ill patients with refractory heart failure, is a stage where medicine has exhausted its options and patients are inevitably confronted with end of life issues. These terminally ill patients may be of young age, hence difficulties in coping with disabling symptoms, psychological distress, and life expectances might be even more pronounced in comparison to the older population with acquired heart failure. Of particular importance, in this setting, is early introduction of specialized palliative care. Its beneficial influence has been shown in acquired end stage heart failure patients and also oncological patients. An intriguing study conducted amongst patients with metastatic lung cancer showed that early palliative care led to significant improvements in quality of life and mood. It also showed that those patients that received less aggressive treatment but more palliative care had longer survival rates [5] (Fig. 1). The etiology of this improvement in survival is not clear, though presumably a number of factors are contributing that relate to the alleviation of suffering from physical pain and the avoidance of depression. The message is very clear that early intervention with palliative care in terminally ill patients should be part of the comprehensive care plan. Palliative care needs to be delivered timely and by a dedicated team of trained health care professionals. This is enforced in PAL-HF (palliative heart failure) trial showing improvement in health-related quality of life in end-stage HF patients [6].

In conclusion, the number of adults with CHD will continue to grow, including patients with more complex CHD. Healthcare providers need to be aware of this unique group of adults with CHD who are in need of

DOI of original article: https://doi.org/10.1016/j.ijcard.2018.11.027.
* Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK.
E-mail address: margarita.brida@icloud.com.

https://doi.org/10.1016/j.ijcard.2018.11.136

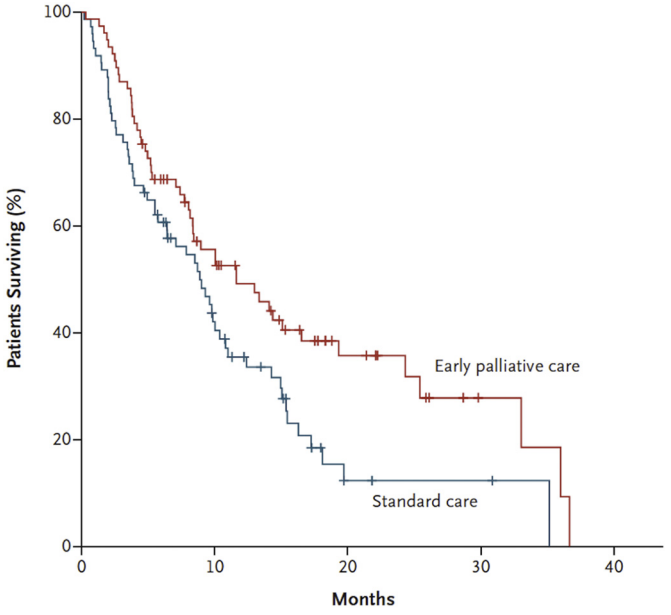

Fig. 1. Kaplan–Meier estimates of survival according to metastatic lung cancer study group. Despite receiving less aggressive end-of-life care, patients in the palliative care group had significantly longer survival than those in the standard care group (median survival, 11.6 vs. 8.9 months; $P = 0.02$). After adjustment for age, sex, and baseline Eastern Cooperative Oncology Group performance status, the group assignment remained a significant predictor of survival (hazard ratio for death in the standard care group, 1.70; 95% CI, 1.14 to 2.54; $P = 0.01$).
Reprinted with permission from Temel et al. [5].

life-long specialist care. Beyond addressing cardiac issues, health care systems needs to provide for the individual patient as a whole, including education and psychological support, as these are closely interrelated to quality of life and outcome.

Conflict of interest

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

Acknowledgment and affiliation

M.B. was the recipient of the International Training and Research Fellowship EMAH Stiftung Karla VÖLLM, Krefeld, Germany.

References

[1] P. Moons, L. Bovijn, W. Budts, A. Belmans, M. Gewillig, Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium, Circulation 122 (22) (2010) 2264–2272.
[2] G.P. Diller, A. Kempny, R. Alonso-Gonzalez, L. Swan, A. Uebing, W. Li, et al., Survival prospects and circumstances of death in contemporary adult congenital heart disease patients under follow-up at a large tertiary centre, Circulation 132 (22) (2015) 2118–2125.
[3] M.A. Gatzoulis, Adult congenital heart disease: education, education, education, Nat. Clin. Pract. Cardiovasc. Med. 3 (1) (2006) 2–3.
[4] E. Troost, L. Roggen, E. Goossens, P. Moons, P. De Meester, A. Van De Bruaene, et al., Advanced care planning in adult congenital heart disease: transitioning from repair to palliation and end-of-life care, Int. J. Cardiol. (2018), https://doi.org/10.1016/j.ijcard.2018.10.078 pii: S0167-5273(18)34021-X [Epub ahead of print].
[5] J.S. Temel, J.A. Greer, A. Muzikansky, E.R. Gallagher, S. Admane, V.A. Jackson, et al., Early palliative care for patients with metastatic non-small-cell lung cancer, N. Engl. J. Med. 363 (8) (2010) 733–742.
[6] J.G. Rogers, C.B. Patel, R.J. Mentz, B.B. Granger, K.E. Steinhauser, M. Fiuzat, et al., Palliative care in heart failure: the PAL-HF randomized, controlled clinical trial, J. Am. Coll. Cardiol. 70 (3) (2017) 331–341.