ESC European Society of Cardiology

European Heart Journal (2025) **46**, 3515–3516
https://doi.org/10.1093/eurheartj/ehaf498

CardioPulse

Global Spotlights

Increased longevity in adults with congenital heart disease and the need for 'extended transition' of care

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A new population of adolescents and young adults has emerged over the last decades, surviving congenital heart disease (CHD) thanks to early surgery and improvements in medical care, whose numbers now exceed those of paediatric patients. The acronym 'GUCH' (Grown Up Congenital Heart), proposed by Jane Somerville, has been widely used in Europe to define this growing population, now superseded by 'adult CHD' (ACHD). Due to their improved longevity, it is common to encounter ACHD patients who are 50 years of age or older, a scenario hard to predict in the early days of CHD management. These novel opportunities, however, also imply unique challenges as the prevalence of arrhythmias and heart failure, the most common complications in adults with CHD, increases with age. Surgeries and interventional procedures performed in childhood, despite indisputable benefits in survival and quality of life, rarely represent a 'cure'. Residual defects and/or late sequelae are common and require timely interventions, following the indications provided in guidelines and consensus documents.[1,2] Furthermore, ACHD individuals are disproportionately exposed to acquired conditions such as coronary heart disease, due to myriad reasons including coronary artery anomalies (both congenital and secondary to surgery) and chronic inflammation. While managing these conditions in older patients falls under the responsibility of adult cardiologists, the rarity and peculiarity of ACHD complications often require skills and cultural background that are exclusive to paediatric cardiologists/ACHD specialists. Thus, there is an emerging unmet need mandating a shift in management programmes, which are only beginning to gain attention in the literature.[1] As we still struggle to provide appropriate transition from paediatric to adult care and a non-negligible number of patients with CHD are lost to follow-up after adolescence, we must now consider the emerging need for structural transition programmes which may span beyond early adulthood. Hence, we propose the concept of an 'extended transition' of care combining multidisciplinary skills to manage long-term CHD complications and promote patient empowerment in cardiovascular prevention (*Figure 1*).

Biological age and frailty in ACHD

A discrepancy between biological and chronological age is evident in patients with complex and moderately complex ACHD individuals, translating into premature aging and increased morbidity and mortality.[3,4] In the most serious conditions, such as Fontan or Eisenmenger syndrome, hard events may be anticipated by decades compared with the general population.[5] Furthermore, extracardiac involvement and early neurological decline contribute significantly to premature senescence. More accurately, we can say that ACHD purports *frailty*, a complex condition characterized by a decrease of physiological reserves and by a weakened response to stressors.[6]

Multiple reasons have been advanced to explain premature aging and frailty in ACHD, including increased unique complications such as protein-losing enteropathy and Fontan-associated liver disease, increased susceptibility to traditional risk factors such as diabetes and hypertension, an inflammatory response to chronic stress, and genetic predisposition to extracardiac disorders (from malignancies to neurological damage). Interestingly, acquired cardiac and extracardiac problems represent the main determinant of mortality in the elderly ACHD population. Understanding the complex interaction between congenital, genetic and acquired conditions is key to developing correct treatment strategies. Likewise, implementing the multidimensional assessment of frailty, now a cornerstone of geriatric medicine and a mandatory step e.g. for Transcatheter Aortic Valve Implantation eligibility, is desirable in ACHD irrespective of age.

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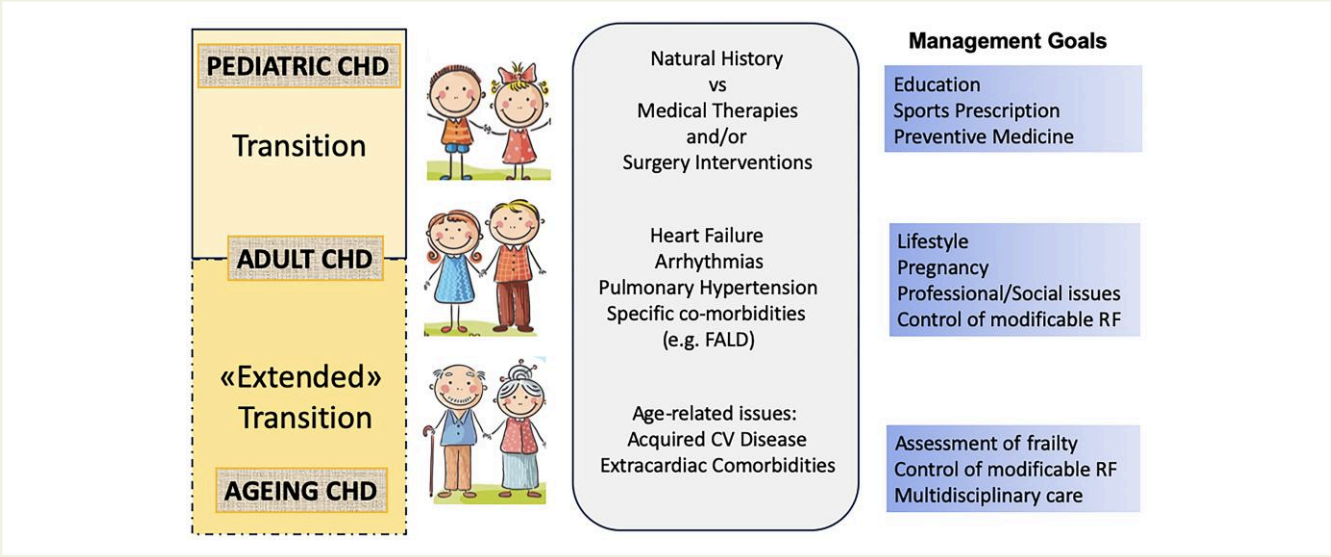

Figure 1 The concept of an 'extended' transition of care in ACHD, issues and stakeholders; CHD, congenital heart disease; CV, cardiovascular; FALD, Fontan-associated liver disease; RF, risk factor

The need for an 'extended transition' of care

Transition of care in paediatric medicine is classically defined as 'the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented health care systems'.[7] As such, it is typically considered concluded by the end of adolescence, after the handover from the paediatric to the adult cardiology team. However, even in best case scenarios, major unmet needs remain. First and foremost, the understanding and management of complications specific to ACHD is not part of the general cardiological expertise and patients are often faced with limited confidence or neglect. For this reason, many continue to seek the help of their physician in the paediatric setting, making the transition process futile. Furthermore, the control of cardiovascular risk factors and inappropriate behaviours, including a sedentary lifestyle, is rarely considered a priority in this population. This is due to the ingrained but erroneous belief that, due to their reduced life expectancy, ACHD patients do not depend on aggressive management of co-morbidities—a bias shared by cancer survivors before the advent of cardio-oncology. Overcoming these limitations represents an urgent target of future strategies for transition, which should extend well beyond adolescence. Necessary actions include the personalized prescription of exercise and a timely correction of modifiable cardiovascular risk factors, to be pursued with the same commitment as the correction of structural congenital defects. Importantly, frailty assessment should be incorporated into decision-making protocols in older ACHD adults, possibly by dedicated tools and scores. Caring for patients facing the long-term interplay of CHD and acquired disease not only requires a collaboration between adult and paediatric/ACHD cardiologists, but a whole team including, among others, geriatric and internal medicine physicians, hepatologists and neurologists. Ideally, dedicated multidisciplinary clinics should be implemented, at least in tertiary referral centres, along the model of other, rare systemic conditions.[8] In an era when multiple rare conditions compete for healthcare resources, and even less ambitious transition programmes, limited to adolescence, struggle to take off,[7] the concept of an extended, multidisciplinary transition of care is challenging. With our ACHD patients, however, we seem to be already late on our schedule.

Declarations

Disclosure of Interest

All authors declare no disclosure of interest for this contribution.

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