The success and failure of the adult congenital heart

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This special edition of *Heart Failure Reviews* focuses on myocardial dysfunction, or “heart failure,” in the setting of adult congenital heart disease (ACHD). However, to understand better the “failure” of the heart with ACHD, it is deeply informative to understand the successes in the field of congenital heart disease. Much like atherosclerotic disease [1], ACHD is a story that begins in childhood. Knowing from whence the field has come and where it is currently informs our understanding of ACHD in critical ways that are not the case for adults with atherosclerotic heart disease and myocardial dysfunction. As such, this introduction aims to provide some context for the excellent reviews that follow.

Congenital heart defects (CHDs) are the most common structurally related group of birth defects in humans, occurring in approximately 1% of all live births [2], and are the leading cause of neonatal and infant death due to congenital issues [3]. In spite of this disease magnitude, advancements in the care of CHDs did not occur until the 1940s, a relatively short time ago. In fact, of those children born with CHD in 1940, only 20% survived to adulthood [4]. By 1960, the survival had doubled to 40%, and by 1980, survival to adulthood was 90%.

The field of congenital heart disease care began with and remains a symbiotic relationship between pediatric cardiologists and congenital heart surgeons. It was Helen Taussig who saw the potential for using Alfred Blalock and Vivien Thomas’ subclavian-to-pulmonary artery shunt as a therapy for tetralogy of Fallot, and with that, the first truly palliative congenital heart surgery was born in 1944 [5]. Between that surgery and the 1990s, the field ushered in the first open heart surgery, cardiopulmonary bypass, intracardiac baffling for transposition of the great arteries, the Fontan procedure for tricuspid atresia, the arterial switch operation, and the Norwood procedure [5]. As

surgical innovations abated, advancements in medical therapies and care accelerated into the 1990s, and by the early 2000s, eight patients with CHDs became the first to receive a transcatheter valve replacement [6].

As a result of the great success within the field, as of 2014, there were more than 1.5 million adults with CHDs in the USA [7], and that number is increasing by an estimated 5% per year [8], putting a reasonable estimate of the current population at 2 million adults. This ever-increasing population of adults with CHDs requires ongoing cardiac care. While it was previously believed that simple CHDs did not warrant ongoing care in adulthood, emerging evidence demonstrates clearly that even simple CHDs confer impacts on long-term cardiovascular risk [9]. However, the great success of the field has drastically outpaced our ability to develop the legion of cardiologists with expertise in ACHD to care for this population. While there is now an American Board of Internal Medicine subspecialty board certification in ACHD, as of June 2019, prior to the 2019 administration of the exam, there were only 302 cardiologists in the USA who have been certified [10], and a significant number of those who have been do not practice ACHD care as a primary focus—varying numbers are electrophysiologists, intensivists, and interventionalists who do not provide “general” ACHD care. Currently, there are 18 ACGME-accredited, 2-year fellowship programs, 16 of which are participating in the Match [11]. In 2019, 55% (5/9) of the programs that participated in the Match filled their positions [12]. Thus, an exceptionally small number of trainees are pursuing training in ACHD, and this has been the case since the inception of the board examination.

Owing to the severe lack of board-certified ACHD cardiologists with currently limited hope of reinforcements in the form of trainees pursuing ACHD, if all 302 of us who are board certified focus solely on ACHD, there would be approximately 6600 patients per cardiologist. That is a number that is more than twice the number of patients Marelli and colleagues have estimated would require a regional ACHD center [13], one comprised of multiple cardiologists. Assuming a given person works 5 days per week during 48 weeks of a given year, that would necessitate each of us seeing 28 ACHD patients per day, if all patients were only seen once annually.

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While advanced practice providers are an integral part of what we do, it is clear that for the foreseeable future, if ACHD patients are to receive cardiological care, a lot of them will have to be cared for by non-ACHD-trained cardiologists.

With the reality of the lack of ACHD cardiologists in mind, this special edition of *Heart Failure Reviews* is meant to serve as a reference for all cardiologists caring for patients with ACHD. The goal has been to provide the congenital context for subgroups of CHDs and practical approaches to caring for them. Drs. Vaikunth and Lui begin the edition by covering general principles of heart failure in ACHD [14], and this is built upon by a review of the general approach to heart failure management to patients with ACHD [15]. Following on, Mueller et al. cover heart failure in the largest such group of patients with ACHD, that being tetralogy of Fallot [16]. The intricacies of heart failure in the setting of the adult with Ebstein anomaly [17] is then addressed before the edition turns attention to the systemic right ventricle [18, 19] and ultimately to perhaps the most challenging group, those with a failing Fontan circulation [20]. The value of exercise testing in the assessment of heart failure in ACHD is then reviewed by Burstein et al. [21]. Finally, the edition culminates with the role of cardiac resynchronization and implantable defibrillators in patients with ACHD [22].

Heart failure in patients with ACHD is to be expected at some point. As I have said to many trainees and colleagues, “Heart failure is a part of all congenital heart disease; it’s just a matter of where they are on the spectrum.” To this end, it is hoped that this edition of *Heart Failure Reviews* will provide useful information that will help improve the care of the growing population of adults with CHDs.

Compliance with ethical standards

Conflict of interest The author declares that he has no conflict of interest.

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