

# Opportunities for training to advance the care for adults with congenital heart disease with advanced circulatory failure

Christopher R. Broda MD 

Department of Pediatrics, Section of Pediatric and Adult Congenital Cardiology, Texas Children's Hospital, Baylor College of Medicine, Houston, Texas

## Correspondence

Christopher R. Broda, Department of Pediatrics, Section of Pediatric and Adult Congenital Cardiology, Texas Children's Hospital, Baylor College of Medicine, 6621 Fannin Street, Suite 19345-C, Houston, TX 77030.

Email: broda@bcm.edu

## Abstract

Heart failure is an emerging issue with important implications in adult patients with congenital heart disease. Practitioners with expertise in both adult congenital heart disease and heart failure are needed to manage this growing and often complex population. In the United States, the optimal training pathway to enable practitioners to best care for these patients is ill-defined. This article explores possibilities and issues that interested trainees may encounter during their training experience.

## KEYWORDS

adult congenital heart disease, fellowship training, heart failure, heart transplantation, mechanical circulatory support

## 1 | ADULT CONGENITAL HEART DISEASE WORKFORCE

Much effort and consideration has been given to designing and implementing appropriate physician training to engender germane skills to care for patients with adult congenital heart disease (ACHD), a population which is growing in both size and complexity.<sup>1</sup> In the United States, the current training pathway allows for physicians who have completed either a pediatric or adult cardiology fellowship to pursue a two-year fellowship which allows one to become eligible to take the Adult Congenital Heart Disease Certification Exam offered by the American Board of Internal Medicine (ABIM). With an estimated population of well over one million individuals and only ~300 board-certified ACHD specialists,<sup>2,3</sup> the need for “generalists” in ACHD is paramount. However, it is very likely that subspecialization within the ACHD specialty is additionally necessary to optimize care for patients. ACHD-specific expertise is needed in focused areas including surgery, cardiac catheterization, electrophysiology, critical care, and advanced heart failure.<sup>4</sup> Despite the present and future needs in these disciplines, there are no clear pathways for trainees to gain a specific knowledge and skill set for ACHD subspecialty training. While all the aforementioned experts are necessary to improve the care and outlook for ACHD patients, perhaps one of the most pressing issues is the need to address the significant problem of heart failure in the ACHD population.

## 2 | THE NEED FOR ADULT CONGENITAL ADVANCED CIRCULATORY FAILURE SPECIALISTS

The importance of training specialists with proficiency in advanced heart failure for ACHD patients is difficult to overstate because heart failure is the leading cause of death in the ACHD population and responsible for ~12% of the hospital admissions in patients with complex congenital heart disease.<sup>5,6</sup> The gravity of the problem of heart failure in the ACHD population is well recognized by ACHD providers and researchers; recently, heart failure was listed as a high-priority research area by the National Heart, Lung and Blood Institute/Adult Congenital Heart Association Working Group.<sup>7</sup>

Despite the commonly used phrase, it might be more accurate to describe ACHD patients suffering from intractable symptoms related to their cardiovascular anatomy as “circulatory failure” rather than “heart failure.” The term heart failure implies impairments in the cardiac structure or function; however, many individuals with ACHD may have end-stage disease with multi-organ dysfunction related to abnormalities of the circulatory system and yet the classic etiologies of heart failure such as reduced systolic function of the left ventricle and/or significant (mitral) valve regurgitation are absent.

Advanced circulatory failure (ACF) in ACHD patients is often due to a myriad of different symptoms instigated by a sequelae of altered physiologies. Symptomatology differs by disease processes

including pulmonary hypertension/Eisenmenger's syndrome and chronic cyanosis; systemic right ventricular failure after an atrial switch operation or physiologically palliated congenitally corrected transposition of the great arteries; and left, right, or biventricular failure from unusual mechanisms in the tetralogy of Fallot population.<sup>8-11</sup> Single ventricle patients palliated to Fontan circulation can develop end-stage multi-organ dysfunction including hepatopathy, cyanosis, or hyperammonemia resulting from the development of veno-venous collaterals, circulatory inefficiency from a heavy aortopulmonary collateral burden, poorly understood chronic renal disease, development of micro-arteriovenous malformations in the pulmonary circulation from inadequate delivery of hepatic factor, protein losing enteropathy, plastic bronchitis, incessant arrhythmias, and systolic and/or diastolic heart failure.<sup>12-15</sup> Traditional means of treating heart failure may not be applicable in these populations.

Mechanical circulatory support (MCS) devices such as ventricular assist devices (VADs) have revolutionized the treatment for patients with traditional advanced heart failure. However, the use of VADs in ACHD patients is a rare occurrence at <1% of all implants, despite relatively similar outcomes compared to non-ACHD patients.<sup>16</sup> This may be due to the widely held beliefs that ACHD patients may be poor VAD candidates due to the presence of scar tissue, abundant collateral vessels, restrictive anatomy, and/or limited venous access. Still, the number of implants of VADs in ACHD patients appears to be steadily increasing over time. Opportunities for research and clinical application are ripe for mechanical support in ACHD patients as this growing population promises to place increased pressure to already burdened organ transplantation programs.<sup>17</sup> It is estimated that 10-20% of CHD patients will eventually require heart or heart-lung transplant and ACHD patients account for ~3% of all listings for heart transplant.<sup>18,19</sup> Evidence suggests that ACHD patients are more likely than non-ACHD patients to die or worsen before a heart can be obtained. After cardiac transplantation, ACHD patients seem to have higher mortality in the immediate postoperative period but those who survive have better long-term outcomes compared to non-ACHD patients.<sup>20</sup> Some believe, as ACF care for ACHD is improved, these differences in adverse outcomes in the peri-transplant period will be attenuated.

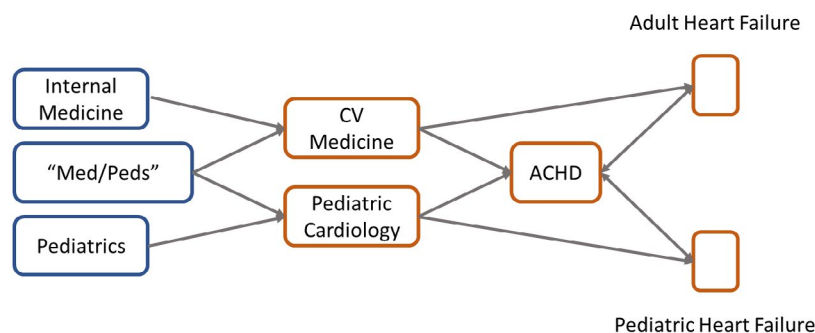
Given these clear differences in circulatory failure between the ACHD and non-ACHD patients, it is likely that an erudite expertise in the congenital condition is important for optimal patient management.

Because of this, for physicians who desire to take care of this heterogeneous and underserved patient population, focused training in ACHD-specific ACF would be ideal.

### 3 | OBSTACLES FOR TRAINING IN ADULT CONGENITAL ADVANCED CIRCULATORY FAILURE

In the United States, to become a practicing heart failure specialist at an adult hospital generally requires one to obtain an Advanced Heart Failure and Transplant Cardiology board certification offered by the ABIM. This certification requires completion of a one-year advanced heart failure fellowship, a prerequisite of which is completion of a cardiovascular medicine fellowship (Figure 1). Physicians who choose to complete a pediatric cardiology fellowship, rather than adult cardiovascular medicine fellowship, are not eligible to obtain the ABIM board certification in Advanced Heart Failure and Transplant Cardiology. There is no similar board certification for pediatric advanced heart failure and transplantation practitioners; generally, pediatric HF physicians will have completed a one-year pediatric advanced heart failure fellowship after completion of pediatric cardiology (Figure 1).

The current training pathway to become an ACHD cardiologist can already be intimidating for a prospective trainee with a minimum time commitment of 8 or 9 years depending on the chosen pathway (Figure 1). With the majority of medical school graduates carrying a significant debt burden and other financial and societal demands, there is intense pressure on physicians-in-training to minimize the duration of training.<sup>21</sup> Compounding the problem of generating ACHD specialists, physicians who choose to train along the "adult pathway" with an internal medicine residency and cardiovascular medicine fellowship could potentially expect an overall lower salary as an ACHD specialist compared to a general adult cardiologist.<sup>22</sup> Because eligibility for pediatric and adult advanced heart failure fellowships significantly differs, the "optimal" training pathway may be to complete a medicine/pediatrics residency, cardiovascular disease fellowship and/or pediatric cardiology fellowship, an ACHD fellowship, and adult and pediatric heart failure fellowships. Adding any additional time to training after ACHD fellowship for ACF sub-specialization is likely a difficult sell to trainees.



**FIGURE 1** Adult congenital heart disease and heart failure training pathways. Blue box—residency; Orange box—fellowship. Sizes of boxes are relative to the duration of the training

Given the pressure to reduce training time, a potentially attractive option for interested trainees may be to focus on ACHD within a three-year cardiology fellowship and advanced heart failure fellowship. Previously, one possible pathway that had been suggested to become an ACF specialist is to include additional ACHD experiences in a cardiovascular medicine fellowship and attempt to focus a traditional adult heart failure fellowship on the care of the ACHD patient.<sup>23</sup> The American College of Cardiology's Core Cardiovascular Training Statement (COCATS) on Training in Heart Failure does specifically mention congenital heart disease in 2 of the 16 Core Competency Components in medical knowledge required for Level III Training.<sup>24</sup> However, there are no additional specific recommendations regarding the type and number of experiences necessary to provide a clinician with adequate expertise to manage ACHD patients with circulatory failure. In this training pathway, the trainee does not engage in a formal ACHD fellowship and, therefore, may not have a substantial clinical experience caring for the myriad of pathophysiologies seen in the ACHD population. Likely, completion of an ACHD fellowship would be the most comprehensive option for one to gain a germane experience for ACHD ACF care. The relevance of the equivalent pediatric training pathway (completion of pediatric cardiology then pediatric heart failure fellowship) is uncertain as the vast majority of ACHD heart failure care occurs at adult hospitals and employment opportunities/credentialing may be limited for pediatric heart failure physicians without ABIM board certification in ACHD or heart failure at adult institutions.<sup>25</sup>

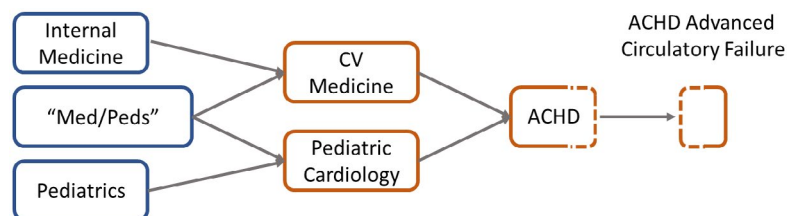
Perhaps the requirements of the advanced heart failure fellowship can be met while a fellow is engaged in the two-year ACHD fellowship. The COCATS recommendations for ACHD training explicitly mention fellows should gain experience with consultation related to outpatient management, hospital admissions, and consideration for advanced therapy and transplantation for ACHD patients with advanced heart failure and pulmonary hypertension.<sup>1</sup> Within the current ACHD fellowship training recommendations, there is some time allotted to focus on a particular subspecialty. Completion of an advanced heart failure fellowship while pursuing an ACHD fellowship would be an enticing avenue of training for physicians. However, it is unclear if the current training paradigm is able to provide sufficient time within the 2-year ACHD fellowship to meet equivalent Level III Training expertise provided by a dedicated 12-month advanced heart failure training experience.

#### 4 | FELLOWSHIP TRAINING IN ACHD-SPECIFIC ADVANCED CIRCULATORY FAILURE, MECHANICAL CIRCULATORY SUPPORT, AND ORGAN TRANSPLANTATION

A potential solution to meet the need for ACF care in the ACHD population may be to dedicate additional time training in ACHD-specific advanced circulatory failure, MCS, and organ transplantation which would begin during or after the two-year ACHD fellowship (Figure 2). Amenable ACHD fellowship programs could offer trainees elective/research time (up to 6 months in some recommendations<sup>26</sup>) to focus on advanced heart failure medicine which could minimize the additional time needed to gain the relevant experience needed to care for the ACF ACHD patients. Trainees would have ample exposure to the outpatient and inpatient management of ACHD patients with advanced circulatory failure, including hemodynamic assessment, medical management as well as evaluation and selection for cardiac transplantation and/or durable MCS devices. This would necessitate training at a high-volume center, with known low number of ACHD patients who receive cardiac transplantation and/or mechanical support; the training (at this point in time) would likely require supplementary experience in traditional adult and/or pediatric congenital-specific heart failure management. There could be potential for lower volume ACHD training programs to partner with willing centers with more mature heart failure/ACHD ACF experience to allow "externship" opportunities for interested trainees. A dedicated "ACHD Advanced Circulatory Failure Fellowship" would enable a directed effort to both study and contribute to this evolving field. Ideally, at the completion of training, the physician would obtain, or be eligible for, a certification in advanced heart failure, transplantation, and mechanical circulatory support.

An alternative strategy would be for institutions to allow for a dedicated individual to focus his or her practice specifically on the care of ACHD patients with circulatory failure, empowering them with the financial resources, appropriate personnel, and protected time needed to care for these patients and advance the field.

Certainly, the answer to meet the growing need of expertise in advanced circulatory failure in the ACHD population is not straightforward. Moving the needle on this issue will require a substantial interest from trainees, strong mentorship, institutional buy-in, and appropriate recognition from governing bodies. Governmental, industry, and/or philanthropic funding of grant opportunities for



**FIGURE 2** Proposed training pathway for adult congenital advanced circulatory failure subspecialization. Blue box—residency; Orange box—fellowship. Sizes of boxes are relative to the duration of the training. Dashed line represents areas of potential overlap

inclined trainees may spur the development of this much needed expertise. Undoubtedly, the subspecialty of ACHD ACF holds substantial opportunity to build a prosperous and fulfilling academic career caring for interesting, complex, and underserved patients.

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## ORCID

Christopher R. Broda  <https://orcid.org/0000-0001-6844-2348>

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