Advanced Heart Failure in the ACHD Population
Finding the Fellows’ Role in a Growing Field

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“By failing to prepare, you are preparing to fail.”
—Benjamin Franklin (1)

Preparation for every scenario in the rapidly changing field of cardiovascular medicine may not be possible. However, flexibility and a proactive approach to clinical training provides fellows with the best preparation to handle challenges as the field evolves. There is a growing need to care for congenital heart disease patients experiencing the symptoms of and dying of heart failure (HF), and this paper seeks to outline the challenges and opportunities for fellows-in-training who wish to partake in the growing field of treating these patients.

The adult congenital heart disease (ACHD) population is a developing segment within cardiology. Until several years ago, there were no Accreditation Council for Graduate Medical Education-accredited ACHD training programs. In 2015, the first American Board of Internal Medicine ACHD board certification examination was offered. The leaders within the field have moved to prepare trainees for the influx of pediatric patients who will transition to adulthood. As a result of this dedicated training, more specific guideline-driven care is being delivered, which should positively affect the care these patients receive. Successful training programs in this area will need to possess vision, flexibility, and a program champion.

In response to the advancement in care delivery, an increasingly important piece of the ACHD puzzle has emerged—the development of advanced HF, which is now the leading cause of death in patients with ACHD (2). Despite a large number of high-volume transplant centers, the number of congenital patients undergoing transplantation remains low, with much of the recent data coming from single-center experiences (3,4). According to the Scientific Registry of Transplant Recipients, from June 2015 to July 2016, the top 10 centers by volume completed 662 cardiac transplants. Of those, 34 (5.1%) were ACHD patients (5). Furthermore, transplant waitlist mortality or delisting due to worsening clinical status is unfortunately more common in ACHD patients who are listed as status 1A for cardiac transplantation (6). Plus, there are limited experiences with mechanical circulatory support (MCS) in the ACHD population, which potentially reduces therapeutic options for these patients (7). According to a recent abstract, from June 2006 to June 2015, there were 14,746 patients entered into the INTERMACS (Interagency Registry for Mechanically Assisted Circulatory Support) database. Of those, ventricular assist device support was identified in only 111 ACHD patients, and survival was not significantly different between ACHD and non-ACHD patients (8).

Although there are a number of possible explanations regarding the etiology of the ACHD patient who decompensates, such as timing, comorbidities, and complications of surgery, perhaps we are just not as prepared as we should be from a training standpoint. The recent statement from the American Heart Association regarding this issue noted the following: “Heart transplant specialists face the challenge of determining eligibility for advanced heart failure treatments among an increasingly complex population of congenital heart disease patients in whom guidelines for heart transplant and mechanical
circulatory support are scant” (9). Identification of weaknesses is the first step. Next, cardiovascular disease fellows should be trained to become leaders in the field of advanced HF specific to ACHD patients.

One obvious challenge is that under the current 1 year of advanced HF/transplant training, there is no required ACHD or pediatric HF training. Currently, the general cardiovascular disease (CVD) fellowship requires only a brief exposure to ACHD, which is clearly insufficient. How can we expect improvement in HF and transplantation outcomes in these patients if the trainee’s first exposure to a failing Fontan patient is as a newly minted HF attending physician? Are these providers considering a ventricular assist device in a systemic right ventricle or focused on the hemodynamic consequences of an atrial arrhythmia in a patient post-Mustard or Senning procedure? It seems that the current model does not give advanced HF fellows the adequate tools or training required to succeed in taking care of a failing congenital patient. Conversely, are ACHD fellowships providing enough access to advanced therapy training for which “standard” HF patients are candidates? Clearly, both scenarios need improvements.

Fortunately, program leadership has designed a fellowship dedicated to my goal of being an HF cardiologist specializing in advanced therapeutic options for failing congenital patients. This was done by dedicating the third year of my general CVD fellowship to integral congenital heart disease (CHD) paradigms. One-half of the third year of my fellowship was spent at our affiliated children’s hospital learning pediatric echocardiography, catheterization, cardiac critical care, and HF. The remaining 6 months were spent focused on ACHD and pulmonary hypertension. My fourth year is a formal advanced HF year, with a dedicated focus on congenital patients with progressive HF. For other fellows considering a similar path, it is important to decide one’s ultimate career focus—whether transplantation, MCS, pulmonary hypertension, or research, among others—within the ACHD population. This could help drive allocation of training time.

A plan such as this requires 4 major aspects: vision, flexibility, collaboration, and a program champion. Program directors and section leaders must possess the vision to recognize the need to train expert physicians to care for a growing and complicated ACHD patient population. Second, program leadership should provide the flexibility that allows for “out of the box” training and learning. Third, collaboration between ACHD/HF/pediatrics as well as the program leadership is a must to coordinate, fund, and educate trainees. This collaboration may be the most complicated component. The burden lies heavily on the trainee to identify and connect with dedicated mentors within the various groups to help guide both clinical and research questions. Then, in addition to program directors, a true champion for the plan must be identified to support the trainee. In my case, that person was a leader in the HF/transplant section, but within other institutions that champion might reside in the ACHD or pediatric communities.

Opportunities such as this are feasible at other centers as well. There are several programs that have robust ACHD, HF/transplant, and pediatric cardiac programs in which collaboration across the fields would provide exceptional training. For centers without all 4 necessary aspects, partnering with other institutions should be considered for additional training opportunities.

All CVD and advanced HF fellows also need more dedicated cross training. Could there be a way to allow trainees to be fully trained in both specialties without lengthening fellowship? As the guidelines are currently written, boarding in general cardiology, ACHD, and HF/transplant would require a minimum 6 years after residency. Should trainees be expected to spend 6 years of fellowship in addition to the 3 to 4 years of mandatory residency time? In addition, time must also be allocated for academic endeavors and publishing. There are many areas within this nascent field that are ripe for research. Currently, the ACHD field lags behind HF in regard to proven medical therapy in this population. Plus, timing and appropriateness of MCS and candidacy and listing for transplantation (i.e., heart alone, heart/lung, or heart/liver) are not well defined.

Additional educational opportunities come in the form of conferences. For those interested in this discipline, there are a growing number of sessions at major cardiovascular meetings that are dedicated to this growing field, as well as continuing medical education opportunities. Attending meetings dedicated to HF/transplant and ACHD has allowed me to interact with leaders who have not only provided mentorship, but also proven invaluable in generating research questions and collaborations.
The answers to all of these questions are neither obvious nor straightforward. Due to the relatively recent emergence of this field, much of the current practice regarding transplant/MCS in ACHD is art based on case series and single-center reviews. In time, with coordinated and collaborative training and research, the field will hopefully move from a nuanced art form to strong data-driven care, which will further improve outcomes in a very complicated, young, and underserved population.

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REFERENCES

RESPONSE: Bridging the Gap

Cross-Disciplinary Training in Adult Congenital Heart Disease

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I sincerely thank Dr. Menachem for sharing his valuable experience and insights, and for his empowered commitment to the highest standards of training in the care of adults with congenital heart disease (CHD). Indeed, major mortality reductions have resulted in a dramatic demographic shift in the population with CHD, with adults now outnumbering children by a ratio of 2:1. Projected estimates indicate that the population of adults with CHD will continue to grow until a steady state is reached in approximately 30 to 35 years (1). We are challenged to plan accordingly to meet workforce requirements in providing qualified comprehensive care.

To this point, 2015 was a landmark year. The American Board of Internal Medicine introduced the first certification examination in North America, thereby officially recognizing adult CHD as a bona fide subspecialty of cardiology. The curriculum was structured for pediatric or adult cardiologists as a 2-year fellowship program within recognized training centers. Inspired by the U.S. example, Canada initiated a process toward recognizing adult CHD as an area of focused competence via a Diploma of the Royal College of Physicians and Surgeons of Canada. In Europe, the working group on grown-up congenital heart disease stipulated training requirements and recommended that examinations be organized by national adult and pediatric cardiology societies, with certification by the European Society of Cardiology. To date, national examinations have been developed by Germany, the United Kingdom, and the Netherlands.

Dr. Menachem has raised the important concern of how to best prepare for complex yet foreseeable issues that plague adults with CHD, yet involve expertise beyond the scope of standard adult CHD training. These may include advanced heart failure therapies and transplantation, arrhythmia and hemodynamic interventions, specialized imaging, high-risk pregnancy, pulmonary hypertension, genetic counseling, and end-of-life care. Although each regional and supraregional adult CHD center must grapple...
with optimizing care within the constraints of available resources, a multidisciplinary team-based approach is indispensable. A growing body of evidence supports the notion that outcomes are superior when care is provided by individuals with CHD-specific expertise.

Although there is no “one size fits all” solution to delivering the highest quality care, I fully support the cross-disciplinary training approach advocated by Dr. Menachem. I would argue that it is critically needed. In addition to acquiring the desired competencies, such training fosters opportunities for research and innovation. Dr. Menachem’s fellowship program was diligently crafted to include a year of formal advanced heart failure training preceded by a year dedicated to pediatric and adult CHD. Other fellows may pursue variants such as formal board-eligible training in adult CHD supplemented by ample exposure to advanced heart failure therapies. In general, adult CHD programs are highly committed to their clientele and supportive of processes to improve delivery of care. Some already integrate opportunities for subspecialty exposure by offering a core adult CHD training program with various subspecialty pathways, or by partitioning training into an adult CHD component and another subspecialty module. Regardless of the chosen path, cross-disciplinary training requires dedication, flexibility, collaboration across fields, vision, and supportive mentorship, as witnessed by Dr. Menachem’s experience.

REFERENCE