

## A Rising Tide of Adults with Congenital Heart Disease: What Can be Done?

Anushree Agarwal<sup>1\*</sup> and Zian H Tseng<sup>2</sup>

<sup>1</sup>Assistant Professor of Medicine, Division of Cardiology, San Francisco, California, USA

<sup>2</sup>Murray Davis Endowed Professor, Associate Professor of Medicine in Residence, Cardiac Electrophysiology Section, Cardiology Division, University of California, San Francisco, California, USA

**\*Corresponding Author:** Anushree Agarwal, Assistant Professor of Medicine, Division of Cardiology, San Francisco, California, USA.

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Congenital heart disease (CHD) is the most common type of birth defect and over the last three decades, adults with CHD (ACHD) have become one of the fastest growing populations of adults with chronic heart disease [1,2]. Owing to the extraordinary advances in cardiac surgery, intensive care, and noninvasive diagnosis over the last six decades [3-5]. Approximately 85% of infants born with cardiovascular anomalies can now expect to reach adulthood, and with continued improvement in surgical techniques, this is expected to increase further in the next few decades [6,7]. Adults now account for two thirds of patients with both complex and noncomplex CHD in North America [4], and in a large UK population-based study more than half of the CHD patients were aged  $\geq 20$  years [5]. Overall, it is estimated that  $\sim 1.6$  million adults in United States are now living with CHD, a figure that is expected to increase by about 5 - 10% annually [3,8,9]. Therefore, the profile of this patient population will change over the next few decades, not only due to advancing age, but also improved survival of patients with complex anomalies.

Health care for the increasing ACHD population requires careful planning for their specialized needs and is more expensive than adults without CHD. Recent research has shown increases in the total number of emergency department (ED) visits, hospitalizations (especially for heart failure), mean hospital charges over time, and readmissions [10-12]. Adults with CHD also have worse perioperative outcomes for non-cardiac surgery than non-CHD patients [11,13-17]. Additionally, adults with CHD frequently have neurological/psychological issues, pulmonary limitations, hepatic abnormalities, renal dysfunction, hematologic problems, and endocrinopathies. Their abnormal cardiac physiology also often creates special challenges regarding pregnancy and delivery [18]. Common age-related acquired comorbidities such as hypertension [19], and among ACHD patients  $> 60$  years old, coronary artery disease (CAD), depression and dementia may be predictors of mortality and health resource utilization [20-23]. Recent studies suggest that mortality among ACHD patients stems primarily from non-CHD cardiovascular diseases, respiratory disease, and cancer [21,24,25]. Increased risk for arrhythmias is also emerging as a common source of morbidity in the ACHD population [19]. Hence we are seeing a rising tide of a challenging population with health challenges related both to sequelae of CHD and aging.

Quality of care data suggest that to bridge gaps in care and improve outcomes, ACHD patients benefit from regular outpatient visits at specialized ACHD centers [26-28]. Referral to specialized-ACHD care is independently associated with a significant mortality reduction [29,30]. Overall, these patients require a comprehensive team approach that involves not only frequent regular outpatient evaluations with specialists in CHD but also care from providers with experience managing the interaction between CHD and typical adult-onset comorbidities [31,32]. The specialized center often serves as the medical "home" for this population of patients and visits are considered a measure of quality of CHD care.

Unfortunately, there have been significant gaps in access to care for these patients, and particularly, patients transitioning from pediatric to adult specialists often suffer fragmented and inconsistent care, which may coincide with loss of insurance [27,33,34]. Data from

the Netherlands shows that adults with CHD have 3.6-fold higher odds of facing barriers to obtaining insurance and increased premiums [35]. To meet these challenges, tremendous efforts have been made globally to improve the access to care and understand the clinical needs of these patients, and specifically in domains of clinical, education, research, and community collaborations. Accredited training opportunities are now available for pediatric and adult cardiologists; multicenter research collaboration have been created like the Alliance for Adult Research in Congenital Cardiology; and other patient/provider collaborative organizations such as the Adult Congenital Heart Association and the International Society of Adult Congenital Heart Disease have initiated major global initiatives. Funding for research through various federal and non-federal organizations has also improved, especially in the US [36].

Despite all these efforts, much work still needs to be done to meet the needs of this population. We need to plan early to develop the infrastructure to meet the challenges of caring for the rapidly increasing population. Educational efforts should be developed to involve and enable these patients and their families to seek early and continued specialized care and be involved in their medical decision-making, especially during their transition into adulthood. A multicenter registry of specific lesions would facilitate research and implementation of care advances and “big data” approaches can be employed to obtain accurate population-based estimates of the rate of comorbidities and health care utilization to help understand how care should be structured, to guide, for example, specific training in acquired age-related comorbidities. These estimates will also provide evidence for ACHD care providers to develop and implement clinical care guidelines and to select specific processes they need to follow. The basic health care utilization and cost data at the population level should be created to help policymakers allocate resources appropriately to provide the most cost-efficient care that improves clinical outcomes.

As the growing ACHD population is expected to continue to live longer and face challenges of acquired conditions, providers who may not be familiar yet with the management of these patients will see them more frequently in outpatient and inpatient settings. As such, increased overall awareness in the community of the needs of the ACHD population will improve care and help policymakers efficiently and appropriately allocate resources for this care. The onus of continuing the success stories of the surgical and technical innovations now relies on providing the optimal, efficient care to the rapidly growing and aging ACHD population aimed at improving their quality of care and longevity.

### Conflicts of Interest

None of the authors have any potential conflicts of interest, including related consultancies, shareholdings and funding grants.

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