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Review

Adults with congenital heart disease – Facing morbidities and uncertain early mortality☆

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ABSTRACT

In the current era, most individuals born with congenital heart disease will reach adulthood. There are now more adults than children living with congenital heart disease in developed countries, and the population continues to expand. Although the life-saving accomplishments achieved in pediatric cardiology are to be extolled, longer-term sequelae also warrant close attention. Adults with moderate or complex disease continue to face cardiac and noncardiac comorbidities as well as a shorted life expectancy. This article begins with a summary of common comorbidities and patterns of mortality faced by adults with congenital heart disease. This is followed by an overview of the broader impact of these issues on the lives of patients, as well as a call for interdisciplinary and comprehensive care to optimize patient outcomes.

1. Introduction

The landscape of individuals living with congenital heart disease has shifted in recent decades. Although diagnoses typically occur in the fetal and pediatric settings, the majority of patients now survive past the age of 18 years, thus contributing to the current environment in which approximately two-thirds of patients with congenital heart disease in developed nations are adults [1]. There are an estimated 1.5 million adults in the United States with congenital heart disease [1]. In Europe, the estimate is 1.8 million, and in Canada, > 160,000 [1,2]. Adults aged 60 years and older are thought to comprise about 10% of adults with congenital heart disease. The increasing prevalence of young- and middle-aged adults with congenital heart disease will likely translate into a growing geriatric population of increasing complexity [3,4]. Nonstandard definitions of qualifying pathologies, non-uniform screening methods, and geographic variability all play roles in generating accurate estimates for global congenital heart disease epidemiology [5]. However, it is estimated that there are 13 million adult survivors who are followed at > 15,000 hospitals around the world [6].

Although the life-saving accomplishments provided by diagnostic, surgical and interventional advances in pediatric care are to be lauded, the longer-term sequelae of congenital heart disease warrant close attention. Adults with congenital heart disease, particularly those with moderate or complex disease, face significant cardiac and noncardiac comorbidities and a shorted life expectancy. In this article, we (i) review common comorbidities faced by adults with congenital heart disease, (ii) describe patterns of mortality, (iii) provide insight into the unique considerations faced by adults living with congenital heart disease, and (iv) emphasize an approach of interdisciplinary and comprehensive care to optimize patient outcomes.

2. Comorbidities in Adult Congenital Heart Disease

As the population of adults with congenital heart disease grows, it is becoming increasingly important for providers to understand not only the cardiac sequelae of their disease, but also the noncardiac problems they face in adulthood [7]. Nearly all organ systems are affected by congenital heart disease, and specific populations, such as patients with Fontan repairs, are at higher risk of complications [7]. Comorbidities may arise as a consequence of medical treatment, prior intervention, or the underlying heart defect. In one recent study, more than half of adults with congenital heart disease had at least one comorbidity, and 23% had two or more; those with multiple comorbidities were less likely to work full time and had worse survival [8]. Further, the number of patients with more than two noncardiac comorbidities associated with a hospitalization almost doubled between 1998 and 2010 [9].

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2.1. Cardiovascular Comorbidities

Heart-related issues are the leading comorbidities in adults with congenital heart disease, particularly heart failure, arrhythmias including sudden cardiac death, and coronary artery disease [8]. Therefore, screening and coordinated care of these health concerns are critical.

Heart failure remains the leading cause of death in adults with congenital heart disease [10]. Multiple mechanisms exist for heart failure in this group: ventricular volume or pressure overload, inability of the systemic ventricle to meet metabolic needs, variable pulmonary blood flow and vascular integrity, and abnormalities of myocardial tissue and the chest wall [10]. Given these unique and complex contributions to heart failure, management guidelines for acquired heart disease are not easily extrapolated to congenital heart disease [5]. Exercise intolerance is recognized as a common symptom of heart failure, and is associated with hospitalizations and mortality [4,5]. However, it is important to note that the recognition and significance of exercise intolerance is challenging in this population. Functional symptoms are notoriously underreported by this group as a result of lifelong adaption to a chronic condition [5,10]. Significantly, it has been suggested that adults with congenital heart disease who also develop heart failure are particularly vulnerable to psychosocial and quality of life impairment [11].

Arrhythmias are the most common long-term complication in adults with congenital heart disease, and a leading cause of hospitalization [12]. These increase with age and lesion complexity. All types of ventricular and atrial arrhythmias may be encountered in adults with congenital heart disease, with multiple types often coexisting. Ventricular arrhythmias are up to 100 times more common than in the general population, and are a leading cause of sudden death. Re-entrant tachyarrhythmias and automatic foci may be related to abnormal anatomy as well as surgical alterations, as is nodal dysfunction, which often leads to pacemaker implantation [5,12]. Arrhythmias, especially sudden cardiac death, are of particular concern in patients with repaired tetralogy of Fallot. They are common and not benign in this population, associated with increased heart failure, reoperation, stroke, and death [13]. Atrial arrhythmias also predispose patients to embolic stroke, alongside other causes of blood stasis and increased blood viscosity. Ischemic stroke rates among adults with congenital heart disease have been reported as 9 to 12 times higher (< 55 years old) and 2 to 4 times higher (55–64-year-olds) compared to the general population [14]. The incidence of hemorrhagic stroke has also been shown to be 5 to 6 times higher (< 55 years) and 2 to 3 times higher (55–64-year-olds). Further, the broader impact of atrial fibrillation includes impaired quality of life [15].

Coronary artery disease in adults with congenital heart disease is thought to have a similar overall prevalence as in the general population, and has been flagged as an independent predictor of mortality in this group [4]. However, the relative risk of developing coronary artery disease is linked to the type of congenital heart disease. It is more commonly seen at younger ages, and in coarctation of the aorta, anomalous coronary arteries, and in surgical repairs involving coronary transfer [16]. In one study conducted to characterize coronary bypass surgery in adults with congenital heart disease, the mean age at surgery was 66 years, and common congenital heart disease lesions were atrial and septal defects and tetralogy of Fallot. Hospital mortality was 2%, with freedom from reintervention 88% at 5 years and 82% at 10 years [16]. The absence of symptoms due to innervation disruption at prior surgery or autonomic dysfunction, difficult electrocardiogram interpretation, and difficulties with stress test performance and interpretation make diagnosis of coronary disease challenging in this population. Therefore, a higher index of suspicion for screening and aggressive risk factor modification may be warranted [16].

While not necessarily a comorbidity, but rather a result of aging with repaired congenital heart disease, repeat cardiac surgery is common. Approximately 20% of adult congenital heart disease admissions involve cardiac surgery, for either new diagnoses or repeat interventions [17]. Even with operative mortality as low as 1–2% (such as in pulmonary valve surgery), significant cumulative risk is present since it is not uncommon for patients to require multiple interventions [13]. It is likely not surprising that overall healthcare resource utilization is high. Rates of emergency room visits, hospital admissions, and cardiac interventions are several-fold greater than in the general population [18,19]. In multiple studies, more than half of patients have emergency room contact or inpatient stays, and a significant portion of these include critical care stays. Patients with severe congenital heart disease have higher rates for all contact compared to those patients with less complex lesions, independent of age, gender, comorbidity, and time in study [18]. During surgical admissions, providers must address not only surgical issues, but also those of acquired comorbidities and advancing age [17]. In two separate studies, 10% of adult congenital heart disease surgery admissions were considered high resource use admissions (greater than 90th percentile for hospital charges), yet accounted for over 30% of total charges [17,20]. These admissions had significantly longer median lengths of stay (over two weeks compared to less than one) and higher inpatient mortality rates.

In summary, living with congenital heart disease involves more than coping with the original cardiac diagnosis and pediatric intervention. During adulthood, patients with congenital heart disease face several cardiac comorbidities that entail symptom burden, increased hospitalizations and outpatient clinic appointments, and quality of life impairment. Thus, the potential impact on the lives of adult patients (including family and work commitments) cannot be overstated.

2.2. Non-cardiovascular Comorbidities

Comorbidities beyond the cardiovascular system are increasingly important as this population moves through adulthood. A recent scientific statement from the American Heart Association provides a comprehensive review of noncardiac comorbidities [7]. Typical events such as pregnancy and noncardiac surgery often require special care [21]. Diseases of the lungs, kidneys, and liver, as well as diabetes and cancer, deserve particular attention.

Lung disease is pervasive in adults with congenital heart disease. Reasons include diaphragmatic weakness, restrictive thoracic cage, and previous thoracotomy/sternotomy, as well as abnormalities in lung parenchyma, lung hypoplasia due to altered blood flow, and amiodarone-induced toxicity [22,23]. Pulmonary arterial hypertension is prevalent in 4–10% of adults with congenital heart disease and is associated with exercise intolerance and heart failure. It arises most commonly in the setting of left-to-right shunts causing increased pulmonary blood flow, which leads to increased pulmonary vascular resistance and vascular remodeling. It may persist even after defect closure, and is an important cause of morbidity and mortality [5,23]. Adults with congenital heart disease not uncommonly have abnormal spirometry, with low forced vital capacity and forced expiratory volume measurements. Restrictive lung disease is especially common in patients with Fontan repairs and patients who have had multiple sternotomies. Cardiopulmonary exercise testing is useful as an objective measure of exercise capacity [22]. Improving exercise capacity through aerobic training is considered safe and is worthwhile, particularly given the long-term benefits of physical activity on quality of life, morbidity and mortality [7,24].

The prevalence of renal dysfunction in adults with congenital heart disease is far beyond that of the general population: 18-fold higher in noncyanotic patients and 35-fold higher in cyanotic patients [25]. Despite their young age, nearly half of adults with congenital heart disease have mild renal dysfunction, and the rate of decline in renal function may be more rapid [22]. In patients with moderate or severe lesions, renal dysfunction is present in 1 in 5, and up to almost three fourths of patients with Eisenmenger Syndrome. Moderately or severe impairment

in glomerular filtration rate may carry a 3-fold higher risk of mortality [25]. Mechanisms for renal dysfunction include low cardiac output and stimulation of the adrenergic and renin-angiotensin-aldosterone systems. Chronic hypoxia with secondary erythrocytosis and increased blood viscosity may lead to increased oncotic pressure in the renal tubules. Risk factors for cardiovascular disease such as hypertension and diabetes pre-dispose to renal dysfunction. Surgeries requiring cardiopulmonary bypass and nephrotoxic exposures such as contrast are also important causes of renal dysfunction in this population [22]. Resultant renal vasoconstriction and sodium and water retention promote remodeling and worse cardiac function, similar to in acquired heart failure. Renal dysfunction also aggravates hypertension and anemia, both important considerations in heart failure [22,25]. Not surprisingly, end-stage renal disease is known to have a negative impact on health-related quality of life [26].

The significance of hepatic disease, particularly in the Fontan population, is rapidly gaining recognition. However, the prevalence is poorly characterized and difficult to estimate, possibly as a result of the myriad of causes and manifestations of disease [7]. Symptoms are often sub-clinical until the late stage, and noninvasive imaging poorly estimates the degree of fibrosis [27]. Congestive hepatopathy from right heart failure, infections/hepatitis, transfusions, and medications (such as those used for pulmonary hypertension) are important causes of liver disease. Prolonged episodes of hypoxia and low cardiac output, elevated central venous pressure, or thrombosis of hepatic vessels also contribute, particularly in the Fontan circulation [22]. Importantly, liver disease leads to coagulopathy and immunodeficiency, and the development of cirrhosis or hepatocellular carcinoma [22,28], all of which impact heart transplant candidacy. In recognition of the dual focus on quantity and quality of life, prompt treatment of the complications of liver disease has been associated with improved health-related quality of life [29].

Diabetes mellitus is associated with micro- and macrovascular complications and consequently a shorter life expectancy in those affected compared to the general population. Sedentary lifestyle and obesity are important contributions to its development. Adults with congenital heart disease may be more sedentary due to physical limitations or activity restrictions recommended earlier in life. They may also be more at risk of being overweight as a result of differing nutritional strategies in infancy [30]. Development of diabetes in adults with congenital heart disease is not uncommon, with prevalence of 4–8% in one study. Those with diabetes and congenital heart disease experience a higher mortality than those without congenital heart disease [31]. Metabolic syndrome, an important precursor to diabetes characterized by obesity, dyslipidemia, insulin resistance, and hypertension, is also common in adults with congenital heart disease, 15% versus 7.4%. In one study, there were similar rates of obesity, but more dyslipidemia and elevated fasting glucose in adults with congenital heart disease. These cardiovascular risk factors are associated with increased mortality, a 2-fold risk of atherosclerotic disease, and a 5-fold risk of developing diabetes [30].

The annual incidence of age-related malignancy is rising among adults with congenital heart disease, with cancer identified as the fourth leading cause of death after heart failure, pneumonia, and sudden cardiac death [32]. The prevalence of cancer in adults with congenital heart disease is up to 2 times higher than in the general population, as evidenced by multiple studies; this may be due to genetic links or congenital heart disease-related surgeries, as well as long-term exposure to radiation during imaging and interventions [7]. Alarmingly, screening rates for malignancies such as breast, cervical, and colon cancer, are lower than in the general population. In one study, adults with simple, moderate, and complex congenital heart disease had 58%, 37%, and 44% adequate cancer screening respectively, which decreased as NYHA class increased [33]. Barriers to screening include lack of health insurance or education and not having a primary care provider. In addition, a focus on the underlying cardiac disease may divert attention from cancer surveillance [33].

Most individuals with congenital heart disease identify as being a “cardiac patient.” However, as they progress through adulthood, the involvement of other organ systems and noncardiac comorbidities (and additional clinic visits and consultations with medical sub-specialists) adds to the complexity of symptomatology and medical care. Patients are also confronted with the impact of one or more comorbidities on health-related quality of life and mortality.

3. Mortality

Although most people born with congenital heart disease now survive to adulthood, this fact does not imply a normal life expectancy, and life expectancy predictions vary widely by the population under study. In 2010, Khairy et al. reported an overall median age at death of 75 years, although 23 years was observed in patients with severe lesions [34]. In this same year, Zomer et al. reported a median age at death of 48 years [35], and in 2016, Engelings et al. reported an average age at death of 40 years [36]. It is not uncommon for patients with repaired tetralogy of Fallot to survive into their seventh and eighth decades [37]. However, adverse events occur in about half of patients by age 50 [13]. In contrast, adults with Fontan repairs are at high risk of early death. One study reported freedom from failure, transplant, or death of 70% at 20 years after Fontan, with approximately 50% experiencing a major adverse event within 15 years of surgery [38]. In the elderly, relative survival prospects are similar to the general population, mainly due to the predominance of simple lesions but also related to balanced physiology or superior resiliency in this subgroup who has already survived so far into adulthood [3,4].

For most subgroups, progressive heart failure remains the predominant cause of death. However, arrhythmia and sudden cardiac death also warrant consideration [4,35,36]. Malignancy, pneumonia and other organ failure are leading noncardiac causes of death [32,35]. It is these noncardiac conditions that more strongly drive mortality in the elderly [3,32].

Diller et al. introduced the concept of equivalent age, whereby mortality rates by congenital heart disease lesion are compared to those for the general population. For example, they concluded that a 40-year-old patient with a Fontan repair in their patient series had roughly the same 5-year mortality rate as a 75-year-old without congenital heart disease [32]. Although medical advances that extend lives are ongoing, the implication of this is that many adults with complex congenital heart disease may in some ways be considered “geriatric” years (or even decades) before their peers. Many patients face a known shortened life expectancy, and a priority should be to determine the most effective ways to balance optimism and realism among patients, families and health care providers.

4. The Broader Impact of Living With Congenital Heart Disease

4.1. Neurocognitive, Psychosocial and Lifestyle Considerations

Although the focus in clinical practice and research is usually physical comorbidities, the neurocognitive and psychosocial issues faced by adults with congenital heart disease are not inconsequential. Children with congenital heart disease are known to be at elevated risk of developmental disorders and disabilities, as well as executive function deficits [39,40]. These neurocognitive challenges do not disappear in adulthood, and in fact, difficulties with executive functions (e.g., organizing and planning behaviors) may become even more pronounced in the less structured adult environment [41]. Adults with congenital heart disease, especially with more complex lesions, face lower educational attainment and higher rates of unemployment [42–44]. The heart-brain axis serves as an important source of neurodevelopmental and neurocognitive impairment, with multiple opportunities for adult exposures that can lead to cumulative neurologic

impairment [45]. Factors that may contribute to cognitive decline in the adult setting include heart failure, atrial fibrillation, coronary artery disease, cardiac surgery and critical illness [41,45]. The presence of neurocognitive difficulties among adults with congenital heart disease impacts education and employment opportunities and abilities. This also impacts patients' abilities to navigate the complex nature of cardiac and noncardiac comorbidities, particularly adherence with multiple health behaviors (including attendance at an increasing number of medical appointments, complicated medication regimens, and diet and physical activity recommendations).

Authors of a meta-analysis concluded that self-reported emotional symptoms among adolescents and adults with congenital heart disease were comparable to those of healthy peers or normative data [46]. The authors, however, noted significant heterogeneity across studies. North American studies that utilized structured psychiatric interviews observed that mood and/or anxiety disorders were present in approximately one third of adults with congenital heart disease [47–49]. The prevalence of post-traumatic stress disorder in this patient population has also begun to receive attention [50,51]. It is interesting to note that European studies suggest more favorable emotional functioning compared to American studies, which may be related to healthcare access issues [6]. In order to understand the potential for psychosocial impairment in adulthood, a multifactorial approach is required [52,53]. Social challenges faced by many individuals with congenital heart disease throughout the lifespan include “feeling different” and difficulties with peer and romantic relationships [49,54–56]. Other health-related challenges include managing the transition from pediatric to adult care, treatment decision-making, and the adjustment to implanted cardiac devices and declining health [7,57].

The nature of congenital heart disease and its comorbidities leads to important lifestyle considerations. Educational and career counseling is important to maximize achievement in these domains. [24,42–44,56] Documentation of diagnoses and limitations are important to provide employers and insurance companies with the latest knowledge relevant to this group. Contraception and pregnancy planning also require a proactive approach, particularly for women with complex disease in whom pregnancy carries risks for both mother and fetus [58].

In addition to these direct effects, congenital heart disease also has the potential to indirectly impact many facets of a person's life. For example, although tattoos and piercings are generally considered safe, endocarditis surveillance is important [24]. For these reasons, patient education at the time of transition to adulthood includes not only cardiac-specific information, but unique considerations for lifestyle matters such as educational and vocational planning, contraception and family planning, physical activity, and alcohol and substance use [59].

In summary, when considering the broader impact of living with congenital heart disease, it becomes clear that the effects extend beyond physical manifestations. Many adult patients face neurocognitive and psychosocial challenges and are confronted with more complex decision-making regarding lifestyle matters that are relatively simple for their healthy peers. Despite these unique challenges, the coping skills and resilience of patients deserves attention [60,61]. For example, a qualitative study of 7 patients (aged 17–32 years) with Fontan physiology revealed three main themes: happiness over being me, focusing on possibilities, and being committed to life [62]. Thus, providers caring for adults with congenital heart disease are urged to be transparent by addressing common challenges, while also acknowledging the resilience and impressive coping strategies and positive views on life expressed by many patients.

4.2. Patient Considerations Regarding Life Expectancy and Advance Care Planning

As previously stated, adults with congenital heart disease of moderate or great complexity face a shortened life expectancy compared to healthy peers. It is thus important to balance hope with realism, which has been described as “hoping for the best but preparing for the worst.” [63] The results of a 2006 study, however, found that 85% of adolescents and young adults with moderate or complex congenital heart disease expected to live longer than was estimated based on mortality data available at that time [64]. Specifically, patients with congenital heart disease of moderate complexity expected to live to age 77 ± 10 years, and those with complex disease expected to live to age 70 ± 13 years. This study revealed a significant lack of education and/or awareness of mortality risks among adolescents and young adults. It is unknown whether this would be different in a contemporary cohort and/or among middle-aged or older adults.

Nonetheless, there is emerging evidence to suggest that adults with congenital heart disease seek information about life expectancy and wish to discuss advance care planning. The results of two North American studies suggest that between 61 and 70% of adults with congenital heart disease want information about the life expectancy of patients with their type of heart condition [65,66]. Of adults with congenital heart disease surveyed in outpatient clinics, 78% reported that they wanted their providers to initiate end-of-life discussions, ideally prior to being diagnosed or symptomatic with life-threatening complications [67]. In another study the median rating for the importance of discussing advance care planning on a 0 to 10 scale was 7 [66]. Unfortunately, research among adults with congenital heart disease in the outpatient setting has revealed that few patients (1–13%) recall previous advance care planning discussions with providers and a minority (5–21%) have completed advance directives [65–67]. A survey of 48 North American adult congenital heart disease providers revealed that the most common barriers to advance care planning discussions were (i) challenges estimating life expectancy (87%), and (ii) the belief that patients are not ready for these discussions if their anticipated life expectancies exceed 5 years (63%) or 10 years (79%) [68]. As we do not currently know whether advance care planning or end of life care preferences and practices vary according to cultural or religious practices within the congenital heart disease population, international research into these matters is advised.

There are multiple reasons why some patients with moderate or complex disease might be unaware of their shortened life expectancy, including a paucity of clear information provided by pediatric and/or adult providers and a reluctance on the part of some patients to accept information that is provided to them. What is evident, however, is that many patients in the adult setting express interest in initiating these important discussions over time.

5. The Importance of Interdisciplinary and Comprehensive Care

In the context of a growing population with complex and costly care needs, we must ensure that reducing disease burden means targeting overall well-being alongside survival [69–71]. Significant hidden morbidity and suffering are likely to be encountered as these patients navigate the previously-uncharted territory of living longer, and they should be empowered to achieve the best quality of life possible [69].

It has been proposed that comprehensive care in congenital heart disease begins at birth (or even earlier for those diagnosed in the fetal period) [68]. Within this model of care, the journey of living with congenital heart disease typically includes multiple interventions throughout childhood and adolescence, during which time conversations about long-term expectations are typically held with parents. In adulthood, there is often a plateau of stability before the onset of adverse cardiac events and functional decline. At this time, the focus appropriately shifts to advance care planning, now directly with the patient.

Throughout their lives, the quality of life and psychosocial well-being of individuals with congenital heart disease merits clinical attention. Although most adults with congenital heart disease with anxiety and/or mood disturbance do not receive treatment [47–49], patients are known to be interested in opportunities for peer support and

Table 1
Challenges and opportunities for comprehensive care of adults with congenital heart disease.

Challenge	Opportunities
Cardiac morbidities	Educate patients regarding the most common cardiac comorbidities faced by patients with their specific diagnosis in order to ensure prompt medical attention when required. Inform patients that congenital heart disease does not preclude coronary artery disease and encourage the adoption of heart-healthy behaviors (e.g., diet and physical activity) to reduce risk. Encourage regular surveillance for cardiac conditions according to guideline recommendations.
Noncardiac morbidities	Educate parents of children with congenital heart disease (and later patients themselves as they reach adolescence and young adulthood) about the important of maintaining a healthy weight. Identify other specialists within an institution (or community) with the expertise and ability to closely collaborate (e.g., a hepatologist to share in the care of patients with Fontan repairs and liver disease). Encourage guideline- and lesion-directed screening for conditions such as diabetes, renal disease, and malignancy.
Neurocognitive, psychosocial and lifestyle considerations	Provide educational and vocational guidance beginning in the pediatric setting. Offer a comprehensive transition education program that includes broader lifestyle matters based upon patients' specific diagnostic, treatment, and functional considerations. Recognize that neurocognitive difficulties present challenges with adherence to complex health care regimens and modify communication strategies accordingly. Improve access to specialized mental health care by integrating mental health professionals within adult congenital heart disease teams and/or identifying community providers to whom to refer patients.
Reduced life expectancy and advance care planning	Invite discussions about care preferences early in the physician-patient relationship. Re-address advance care planning at regular intervals and at changes in clinical status. Introduce the completion of advance directives as standard practice. Liaise with palliative care specialists to improve the quality of end-of-life experiences for patients and families.

mental health treatment [72,73]. In order to address the psychosocial challenges faced by adults with congenital heart disease, “Four A's” have been proposed: (1) ask about specific challenges and difficult situations, (2) advise about certain challenges and how they might be addressed, (3) assist with education and problem-solving, and (4) arrange referrals to mental health professionals as indicated [52,53]. Ideally, mental health professionals (e.g., psychologists, psychiatrists, clinical social workers) would be integrated within adult congenital heart disease teams [52,74]. When this is not possible, medical teams are encouraged to closely collaborate with mental health professionals within one's institution as well as the broader patient geographic catchment area.

Although advance care planning ideally begins before the detection of life-threatening complications [67], such discussion becomes critical at the time of significant decline. Advance directives have advantages for patients, their loved ones, and providers as they allow patients to specify their preferences for end-of-life care and identify who they wish to make decisions on their behalf should they be unable to do so. Guidelines for the care of both adolescents and adults with congenital heart disease emphasize the importance of advance care planning to occur early in the disease course [21,57]. It should be noted that in order to be effective, provider initiation of end-of-life and advance care planning discussions requires a unique skill set in which not all providers may feel comfortable or adequately trained. Among surveyed adult congenital heart disease providers, the vast majority wanted more information and resources about end-of-life communication strategies (89%) and advance directives (79%) [68] Recommendations to facilitate advance care planning have been offered; [63] these include normalizing discussions, incorporating an Ask-Tell-Ask approach, providing a range when asked about prognoses, and acknowledging one's own emotional reactions. Further, information and training in advance care planning skills is clearly indicated in continuing education initiatives.

In addition to recommendations about advance care planning, guidelines from multiple professional organizations recommend that adults with congenital heart disease be seen at least once in a center that specializes in this type of care [21,75]. A significant reduction in mortality has been shown concurrent with a parallel increase in referral to these centers, independent of age, sex, and comorbidity, and especially in complex congenital heart disease [76]. In order to provide comprehensive care across patients' lifespans, an interdisciplinary approach is required. Just as adult congenital heart disease providers must collaborate with other specialists (e.g., nephrologists, hepatologists, endocrinologists, and obstetricians) to manage comorbidities and extend lives, it is necessary to collaborate with mental health professionals and palliative care specialists to optimize patient quality of life and end of life care. It takes an entire community of health professionals to attend to the complex needs of adult survivors of congenital heart disease.

6. Conclusions

Adult congenital heart disease is a multi-systemic health condition with the potential for significant burden on the daily lives of patients and their families. Effective management requires an interdisciplinary approach to address cardiac and noncardiac comorbidities and attend to the broader implications of living (and dying) with a chronic health condition. Table 1 contains recommendations for education and integrated care that may assist providers in addressing the challenges faced by these patients. As this unique group continues to grow and differentiate, management of both physical and psychosocial matters is key to maximizing quality of life and long-term outcomes.

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