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Transition to Adult Congenital Heart Disease Care: A Review

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The population of adults with congenital heart disease (ACHD) has grown due to recent advances in surgical procedures. The survival rate to adulthood is now more than 95%. This review identifies current recommendations and status of ACHD management and treatment in the United States by examining comprehensive guidelines for management and transition and comparing them to the current state of the science. Successful transition from pediatric to adult care begins during the adolescent years, and prepares patients for management at an ACHD regional center utilizing multidisciplinary teams of ACHD specialists. Advocacy and research needs for the ACHD population persist.

ADVANCEMENTS IN SURGICAL procedures for congenital heart disease (CHD) over the past 40 years have resulted in an increased survival rate to adulthood from 25% to more than 95% (Warnes, 2005). There has been an increase in the median age of patients with complex CHD, as well as an increase in the number of adults with CHD (ACHD) (Marelli, Mackie, Ionescu-Ittu, Rahme, & Pilote, 2007). The American College of Cardiology (ACC) estimated the total number of adults living with CHD in the United States (U.S.) to be approximately 800,000 in 2001, and other investigators have placed their more recent estimates above 1 million (Hoffman, Kaplan, & Liberthson, 2004; Warnes et al., 2008).

The significant growth of the ACHD population has created a new population of adult chronic care patients and subset of adult cardiology patients. Strides have been made in the past 20 years to develop a system for transitioning the CHD pediatric population into adult care, but still no formal nation-wide system exists in the U.S. (Webb, 2010).

Successful transition programs to adult care with other pediatric chronic disease populations, such as hemophilia, diabetes mellitus, spina bifida, cystic fibrosis, juvenile rheumatoid arthritis, and sickle cell disease, begin the transition process 5 to 7 years before the young adult is to transfer to primary care with the adult provider (Fernandes et al., 2012; Peter, Forke, Ginsburg, & Schwarz, 2009; van Staa, Jedeloo, van Meeteren, & Latour, 2011). A purposeful transition allows for development of self-management and communication skills in the adolescent patient, parent preparation for allowing the patient to assume responsibility for his/her care, and pediatric and adult practitioner organization of transfer of care (van Staa et al., 2011). Although planned transition remains a shared goal of pediatric cardiologists, Fernandes et al. (2012) found that clinical instability associated with comorbidities of adulthood prompt transition to ACHD care more often than age.

The purpose of this review is to identify current recommendations and status of ACHD management and treatment in the U.S. The review encompasses the historical evolution of ACHD management and treatment over the past 2 decades by examining the most comprehensive guidelines for management and transition, and by showing the current state of the science. Healthcare utilization of the ACHD population is examined, and healthcare needs specific to the ACHD population are then considered and contrasted to the

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needs of the pediatric CHD population. Finally, ACHD centers nationwide and worldwide are discussed, and opportunities for nursing involvement are highlighted. Future research and interventional needs are noted throughout.

Literature Search

A literature search was conducted on PubMed for articles on ACHD. Key search terms and phrases included combinations of 'adult congenital heart disease', 'ACHD', 'pediatric transition to adult care', and 'ACHD regional center'. Research articles published since 2006 were reviewed by title and excluded if content appeared to be clinically based. A review of abstracts on the remaining articles resulted in retention of 20 articles. Citations in these articles were also screened, adding another 26 sources.

Recommendations

The ACC dedicated its annual meeting in Bethesda, Maryland, in 2000 to addressing the needs of adults with congenital heart disease (Webb & Williams, 2001). Although 14 years have passed since the conference, the recommendations stemming from this conference are currently the most comprehensive guidelines for management and transition of ACHD, and many of the goals remain unmet (Webb, 2010).

Participants of the 32nd Bethesda Conference delivered five Task Force reports, each focusing on an aspect of ACHD needs. Specific areas examined by each Task Force report are shown in Table 1. The first report served to define and quantify the ACHD population in the U.S., categorizing disease population as complex, moderate, or simple lesions (Warnes et al., 2001). Tables 2, 3, and 4 differentiate among complex, moderate, and simple cardiac lesions according to the committee's designation. Task Forces 2–5 focused on special health care needs of ACHD, including unique medical needs, contraception, pregnancy, exercise tolerance, rehabilitation, and psychosocial issues; the workforce and levels of training needed to treat the growing ACHD population; structures of health care systems to most effectively treat the ACHD population, specifically regional ACHD centers with specialized teams of staff; and access to healthcare and advocacy need (Child et al., 2001; Foster et al., 2001; Landzberg et al., 2001; Skorton et al., 2001).

Clinical practice guidelines set forth by the ACC/American Heart Association (AHA) Task Force in 2008 include the 32nd Bethesda Conference committee recommendations regarding organization of and access to regional ACHD centers, specialized education for cardiovascular specialists in all fields for the treatment and management of ACHD, ACHD population advocacy, and the psychosocial needs of the ACHD population (Warnes et al., 2008).

Provider Education

Participants of the 32nd Bethesda Conference called for 3 levels of training in ACHD. Level 1 entails basic exposure with didactic educational materials to enable the provider to competently recognize and facilitate referral of patients with CHD. This level includes training of all medical cardiology fellows and should include material on CHD anatomy, physiology, pathology, genetics, natural history, clinical presentation, and management. Level 1 trainees should also be able to evaluate CHD results of electrocardiography, echocardiography, and cardiac catheterization (Child et al., 2001).

Achievement of Level 2 and Level 3 training requires the same training as Level 1 with the addition of 1 to 2 years of hands-on ACHD training. Among the competencies for upper level training are medical, surgical, and postoperative management of CHD; experience with clinical research methodology; and psychosocial aspects of adolescence and

Table 1 2000 Bethesda Conference Task Force Reports.

Task Force 1
- Profile of CHD in adult life
- Definition and quantification of the ACHD population
- Disease categorization of cardiac lesions
Task Force 2
- Unique medical needs of ACHD
- Contraception
- Pregnancy
- Exercise tolerance
- Rehabilitation
- Psychosocial issues
Task Force 3
- Provider workforce description
- Educational requirements of providers
Task Force 4
- Organization of health care systems
- Regional ACHD centers
- Specialized healthcare teams
Task Force 5
- Access to care
- Advocacy needs

(Webb & Williams, 2001).

Table 2 Complex Categorization Cardiac Lesions.

Conduits, valved or nonvalved
Cyanotic congenital heart (all forms)
Double-outlet ventricle
Eisenmenger syndrome
Fontan procedure
Mitral atresia
Single ventricle (double inlet or outlet, common or primitive)
Pulmonary atresia (all forms)
Pulmonary vascular obstructive diseases
Transposition of the great arteries
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of atrioventricular or ventriculoarterial connection not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

(Webb & Williams, 2001).