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FOREWORD

Adult congenital heart disease

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“Without balloon septostomy or surgical intervention, 50% of children with transposition will die within the first month and 90% within the first year ... prior to the advent of surgery, a third (of patients with tetralogy of Fallot) died in infancy” [1].

These statistics, from a 1978 edition of the well-established textbook, *‘The Heart’*, edited by JW Hurst, reflected the medical knowledge of the day [1]. Approximately 0.8% of live births have congenital heart disease (CHD); without surgery or intervention, most children died. Following surgery of some defects, especially the more simple ones, survival to adulthood was to be expected, although no real statistics were available. A decade later, in the 1989 edition of *‘Moss’ Heart Disease in Infants, Children, and Adolescents’*, a more optimistic tone

of survival appeared, and along with it, the issues of concern for continued patient care [2].

Adult CHD (ACHD), as a specialty, was not a major concern in either pediatric or internal medicine cardiology training programs for decades. Pediatric cardiologists assumed that patients would simply transition to internal medicine and internists, with training in acquired heart disease, assumed that these pediatric congenital heart specialists had the knowledge to care for

all CHD patients, even into adulthood. However, the flood gates of patient numbers had not yet opened. Foresight among some prompted concerns of who is best suited to provide medical, surgical and psychological care. As a result, a focus shift has occurred over the past decade, with training programs evolving continuity, 'cradle-to-cane-to-grave', congenital heart programs with collaboration between pediatric and internal medicine cardiologists. At present, with statistics showing already over 1 million individuals with ACHD in the USA, or one in every 150 people, and increasing by thousands yearly, specialty training and board certification have evolved.

ACHD patients have distinct problems that are different from same-age individuals with acquired heart disease. The eventual onset of comparable age-related acquired issues such as coronary artery disease, hypertension and diabetes will further add to and compound ACHD patient issues. A repaired congenital heart is not synonymous with a normal heart. Residual surgical scars can be the nidus for arrhythmias. Years of hypoxia cause adverse myocellular histopathologic changes. Risk of sudden cardiac death can be increased. Reversed ventricular morphologies can portend early heart failure. Conduits and valves may require revisions. Decades of cardiac pacing can adversely effect myocardial function. Pregnancy carries additional risks. Psychological and social issues,

as well as employment and insurability, play major roles in an ACHD patient's outlook on life.

The field of ACHD is one of evolution in knowledge and interventions. At present, diverse technologies are evolving. As will be presented in this review, prevention of sudden cardiac death and improved heart-failure management by use of implanted cardiac devices has evolved. Implantable devices provide options to prevent arrhythmogenic death as well as to 'remodel' altered cellular architecture. However, with predictable eventual myocardial deterioration, heart transplantation becomes an eventual option among many ACHD patients, especially those with reversed ventricular anatomies or functional single ventricles.

Financial & competing interests disclosure

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References

1. Hurst JW, Logue RB, Schlant RC, Wenger NK. *The Heart*. McGraw Hill, NY, USA (1978).
2. Adams FH, Emmanouilides GC, Riemenschneider TA. *Moss' Heart Disease in Infants, Children, and Adolescents*. Lippincott Williams & Wilkins, PA, USA (1989)