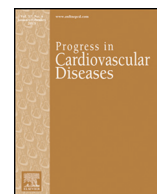




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Editorial

Adult Congenital Heart Disease Care Provision: the Need for a Jack/Jackie of All Trades☆☆☆



“Is it better to know a little about a lot, or a lot about a little?” has often been subjected to philosophical debate. Though generally not applied to medical specialization, the statement nicely contrasts what is required from the generalist compared to the specialist. The lines get blurred, however, when applied to the newly formed specialty of caring for adults with congenital heart disease (CHD; ACHD), where complexity applies to a variety of organ systems beyond just the cardiovascular issues. Barely 5 decades ago, the infant with CHD had a 50% chance of surviving their first year of life and only a 15% chance of surviving to adulthood.¹ Now it is expected that over 90% of infants with CHD will survive into their adult years, some with near-normal life expectancies. CHD runs a large gamut, from simple shunt lesions, to highly complicated lesions, such as the functional single ventricle with heterotaxy, where the normal relationship of abdominal organs and the heart cannot be relied upon.² While a simple shunt may or may not require repair, and often can be corrected percutaneously or through a minimally invasive operative technique; the hypoplastic left heart requires a series of complicated procedures, and patients endure substantial morbidity and mortality risk along the way.³ More importantly the sequelae stemming from exposure to congenitally and then surgically altered, abnormal anatomy can affect a multitude of different organ systems and greatly impact quality of life.⁴

Because of the depth and breadth of lesions and their manifestations, the ACHD specialist must not only understand the anatomy of each heart lesion and its spectrum of severity, but also the varied physiological effects on multiple organ systems, how to manage them and when to refer to specialists that are familiar with and can assist with further management of their sequelae. Little over a decade ago the first set of clinical practice guidelines in ACHD were published.⁵ Recently these were updated, a daunting task owing to the great number of interval publications. The field of ACHD remains in its infancy, however, and there remains much to do to improve the health of this rapidly expanding patient population. Current guidelines recommend regular involvement by an ACHD specialist in the routine care of all but the simplest of CHD lesions.⁶ Management in specialized centers of ACHD appears to improve patient survival,⁷ but this still requires recognition and then appropriate referral. It also requires a specialized center to be in proximity to the patient; though the number of such centers is increasing,⁸ it still remains inadequate. Ideal clinical practice should also include a suitable model of care transition from pediatric to ACHD care, one in which the patient understands the importance of the

process, does not feel abandoned by their previous provider and feels confident in their new care team.⁹

ACHD classification has been recently revised to include both anatomic and physiological distinctions,⁶ thus facilitating the determination of which services these patients may require. Careful clinical evaluation including cardiopulmonary exercise testing and multimodality imaging forms the foundation of further patient phenotyping. Procedural care, be it surgical or transcatheter, should only be performed by specialists with familiarity and/or specific training in ACHD.⁶ In the case of surgery, mortality benefits have been demonstrated for this practice.¹⁰ It is also prudent that providers adequately prepare patients for their extended futures, ensuring healthy lifestyle interventions – eating properly, avoiding tobacco and other drugs and encouraging adequate physical activity. This is one area patients have some degree of control over, unlike their CHD diagnosis. Psychological and neurological health must also be emphasized, with the recognition that patients with more complicated forms of CHD requiring repeated surgical intervention are at greater risk for these sequelae.¹¹ Finally, discussions about sexual activity and reproduction is not the norm for most non-CHD cardiologists. Patients with CHD benefit from discussions regarding pregnancy prevention (including for women, the type and form of contraceptive that is preferred) and pre-pregnancy planning, including fetal and maternal risk assessment to ensure the safest pregnancy outcome.¹²

In this issue I have asked several of my terrific colleagues in ACHD (some of whom are well-known experts in the field and others who are rising stars) to tackle challenging topics in CHD in order to summarize our established clinical approaches and the direction in which the field is heading. This is by no means meant to be a compendium of current ACHD care, but simply a starting point from which to begin understanding this highly varied and challenging patient population. I remain indebted to my colleagues and mentors, but most of all to my patients for teaching me so much about this ever growing and advancing field of practice.

Statement of Conflict of Interest

The author declares no conflict of interest.

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Abbreviations and Acronyms: ACHD, adult congenital heart disease; CHD, congenital heart disease.

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