Specialized Adult Congenital Heart Care Saves Lives

Running title: Webb et al.; Specialized ACHD Care Saves Lives

Gary Webb, MD1; Michael J. Landzberg, MD2; Curt J. Daniels, MD3

1Cincinnati Adolescent and Adult Congenital Heart Disease Program, The Heart Institute at Cincinnati Children's Hospital Medical Center, University of Cincinnati Medical School, Cincinnati, OH; 2Boston Adult Congenital Heart (BACH) Group, Depts of Cardiology, Boston Children’s Hospital and Brigham and Women’s Hospital, Harvard Medical School, Boston, MA; 3Columbus Ohio Adult Congenital Heart Disease Program, Nationwide Children's Hospital, Columbus, The Ohio State College of Medicine, Columbus, OH

Address for Correspondence:
Gary Webb, MD
Cincinnati Children's Hospital
3333 Burnet Avenue
Cincinnati, OH 45229
Tel: 513-803-1777
Fax: 513-803-1778
E-mail: gary.webb@cchmc.org

Journal Subject Codes: Ethics and policy:[100] Health policy and outcome research, Etiology:[8] Epidemiology, Cardiovascular (CV) surgery:[41] Pediatric and congenital heart disease, including cardiovascular surgery

Key words: Editorial, adult congenital heart disease, survival, outcomes research, guideline
For more than half a century growing numbers of practitioners and clinician-scientists have created systems for and practiced healthcare delivery within centers of expertise for young and older adults with congenital heart disease (ACHD). To date, this has been driven largely by energy, enthusiasm, and shared mission without supportive data demonstrating the benefit of such ACHD care programs in the lives of patients and families served by this developing field. Now, for the first time, Marelli and colleagues present in this issue of Circulation good evidence that there is a survival advantage to adult congenital heart patients being cared for by ACHD specialists\(^1\). This confirms what those working in this field had long assumed and hoped, but to date could not substantiate.

The authors utilized their own extensively validated province-wide administrative longitudinal database that has been highly effective in demonstrating the demography and epidemiology of ACHD care in Quebec as a model for North America. Their study included data from 1990 to 2005. They performed an elegant combination of time-series analysis together with case control studies that included year-dependent survival as well as center-dependent survival so as to determine mortality trends within a highly monitored and geographically stable group of adults with CHD. A number of important findings emerged. Specialized ACHD care was independently associated with reduced mortality. Among patients who died, just under half died of cardiovascular diseases. Other causes of death included neoplasms (~18%), respiratory disease (~9%), and diseases of other systems) (~31%). Patients with severe CHD, with a higher expected mortality rate, benefited more than did patients with less severe forms of CHD, but all groups showed reduced mortality.

The study period included the presentation and publication of the first set of ACHD cardiovascular care guidelines, sponsored by the Canadian Cardiovascular Society\(^2\).\(^3\). Thus, the
authors were able to assess the potential impact of these guidelines on patient referral to one of
the 3 specialized ACHD centers in the province of Québec, and to relate patient mortality to
these events. They found that mortality among ACHD patients rose slowly from 1990 to 2000,
and then fell during last 5 years of the study. From 1990 to 1997, there was only a slight increase
in the number of patients seen at ACHD centers in Québec. This increased quite sharply
beginning in 1997, three years before the mortality rate for this population began to decline.

The authors suggest multiple reasons why increasing patient referrals to ACHD centers of
expert care might improve outcomes. Team members within such a system of healthcare
delivery have a special interest, training, and experience with ACHD patients, and as such, may
be more likely to utilize practice guidelines, reduce variability in care and maximize the use of
medical advances than other practitioners who may be less familiar with this information. They
also discuss the “practice makes perfect” hypothesis to explain how such higher volume
specialized centers allow the accumulation of skills and knowledge, the acquisition of
specialized equipment and the development of service models that have been demonstrated in
other cardiovascular patient groups to improve outcomes.

Sophisticated readers may recognize additional and important confounders to the study.
Receipt of care within ACHD specialized care centers may be associated with better transition
and transfer of care from pediatric providers (not analyzed in the current study). If true, the
association with improved outcomes may reflect the success of lifelong coordinated congenital
heart disease care rather than simply adult care. Patient motivation and attention to good health
care principles may influence outcomes and may drive particular adults with CHD to specialized
care centers. As well, the current study demonstrates that the majority of deaths in ACHD
centers of expertise were coded as due to non-cardiac and other causes. This intriguing finding
suggests that, in addition to the direct benefits of specialized cardiovascular ACHD care, improved survival in such patients may reflect the impact of a broad range of healthcare services available at such centers. While these influences may carry important distinction for future study and healthcare refinement, the data presented underscore the fact that specialized ACHD care centers have now truly come of age.

What should we do with this information? This study reinforces recommendations from America\textsuperscript{5}, Canada\textsuperscript{6-8}, and Europe\textsuperscript{9} that adult patients with moderate or complex CHD be followed regularly in specialized ACHD centers, in collaboration with their other providers. Patients and families should be informed of this survival advantage, and encouraged to attend centralized specialized ACHD facilities available to them. Physicians of all types and especially cardiologists should be informed of the survival advantage and encouraged to share or refer appropriate patients to centers dedicated to coordinated ACHD care. Likewise such expert centers appear to have an obligation to communicate well with local providers, to disseminate best practices and care guidelines, and to promulgate optimal quality and innovation.

Efforts to build excellence in ACHD care need to be redoubled, making very timely two ongoing efforts. The first is the development of an accreditation process and criteria for US ACHD clinics, coordinated by patient advocacy within the Adult Congenital Heart Association (ACHA). ACHA will build a roadmap of standards based on expert consensus and care guidelines, including expertise in staffing and services with an emphasis on collaboration, in the hopes that these will lead to a strengthening network of specialized ACHD care programs and improved quality of care in the United States\textsuperscript{10-12}. It is estimated by Marelli and colleagues\textsuperscript{13} that the US will require at least 150 ACHD centers to care for the 50\% of the ACHD population with moderate and severe forms of CHD. The implications for ACHD program building are far
reaching and now have clear and concise data to support this approach.

The second important initiative aims to increase the number of trained and certified ACHD subspecialty cardiologists. Recent collaboration between the American Board of Pediatrics and the American Board of Internal Medicine has led to the creation of a subspecialty certification in ACHD, with the first qualifying examination scheduled for 2015. This will require a greater number of dedicated fellowship training programs for ACHD cardiologists. To this end, ACGME agreed in 2013, to accredit fellowship programs in the subspecialty of ACHD. With these accomplishments we anticipate greater numbers of both pediatric and internal medicine cardiologists will seek training opportunities, and more academic institutions will provide fellowship positions in the advanced care of adults with CHD, thereby increasing the workforce that will be required.

The results from the current study provide legitimacy to prior expert guideline statements that can continue to form the basis for the continued development of systems of ACHD care delivery in the United States and elsewhere. The belief by specialized ACHD cardiologists that has led them to advance such care is now supported by data. The imperative now becomes the achievement of changes in practice that builds a specialized healthcare delivery system to engender fiscally responsible and patient-centered quality care in an atmosphere of collaboration and coordination within the multidisciplinary team of specialized healthcare providers needed for the many and varied needs of ACHD patients. Fortunately, the time to move forward on behalf of these patients is now.

**Conflict of Interest Disclosures:** None.
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Circulation. published online March 3, 2014;
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2014 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

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http://circ.ahajournals.org/content/early/2014/03/03/CIRCULATIONAHA.114.009049

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